A DICTIONARY OF NEUROLOGICAL SIGNS

CLINICAL NEUROSEMIOLOGY

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To my parents

Lexicographer: a writer of dictionaries; a harmless drudge, that busies himself in ... detailing the signification of words.

– Samuel Johnson. *A Dictionary of the English Language.* 1755

Lexicographer: a pestilent fellow who, under the pretence of recording some particular stage in the development of a language, does what he can to arrest its growth, stiffen its flexibility, and mechanize its methods.

– Ambrose Bierce. *The Devil's Dictionary.* 1911

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FOREWORD

Neurology has always been a discipline in which careful physical examination is paramount. The rich vocabulary of neurology replete with eponyms attests to this historically. The decline in the importance of the examination has long been predicted with the advent of more detailed neuroimaging. However, neuroimaging has often provided a surfeit of information from which salient features have to be identified, dependent upon the neurological examination. A dictionary of neurological signs has a secure future.

A dictionary should be informative but unless it is unwieldy, it cannot be comprehensive, nor is that claimed here. Andrew Larner has decided sensibly to include key features of the history as well as the examination. There is no doubt that some features of the history can strike one with the force of a physical sign. There are entries for "palinopsia" and "environmental tilt" both of which can only be elicited from the history and yet which have considerable significance. There is also an entry for the "head turning sign" observed during the history taking itself as well as the majority of entries relating to details of the physical examination.

This book is directed to students and will be valuable to medical students, trainee neurologists, and professions allied to medicine. Neurologists often speak in shorthand and so entries such as "absence" and "freezing" are sensible and helpful. For the more mature student, there are the less usual as well as common eponyms to entice one to read further than the entry which took you first to the dictionary.

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PREFACE

In writing a book devoted to neurological signs and their meaning, it is not my intention to undervalue in any way the skill of neurological history taking. This remains the key element of the doctor-patient encounter both in the neurological clinic and on the ward, and is clearly crucial in order to formulate diagnostic hypotheses, guide clinical examination, and help decide on the nature of the pathological process (if one is present). However, having sat through several thousand neurological consultations, I do not subscribe to the view that all one need do is listen carefully and the patient will "tell you the diagnosis", although this may happen on rare (and often memorable) occasions. Clearly, history taking is not simply a passive recording of symptoms ("what the patient complains of"), but also an active process of seeking information of possible diagnostic significance through appropriate questions; this might be called the "historical examination". This latter facet of history taking, much the more difficult skill to learn, may disclose certain neurological signs which are not available to *physical* examination (principally in the sensory domain, but also intermittent motor phenomena). Hence, my use of the term "sign" in this book is a broad one, encompassing not only findings in physical examination (its traditional use) but also from focused history taking. My operational definition of sign is therefore simply a "signifier", in the sense of phenomena of semiologic value, giving information as to anatomical location and/or pathological cause.

Most neurological textbooks adopt an approach which is either symptom-based, beginning with what the patient complains of and then offering a structured differential diagnosis; or disease-based, assuming that a diagnosis has already been established. Although such texts are of great value, it seems to me that this does leave a place for a book devoted to neurological signs. Signs, elicited in either the historical or neurological examination, bridge the gap between the patient's symptoms, and the selection of appropriate investigations to confirm or refute the examiner's diagnostic formulations and thus establish a diagnosis.

Although it has been mooted whether the dramatic technological advances in neurological practice, for example in neuroimaging, might render neurological examination redundant, others maintain the central importance of neurological examination in patient management.[1,2] It will come as little surprise to the reader that I am emphatically of the latter persuasion. However, this book does not aim to be a handbook of neurological examination technique (one reason for the absence of pictures), or neurological investigation, many excellent examples of which already exist. Rather, it seeks to elucidate the interpretation of neurological signs ("neurosemiology"): their anatomical, physiological, and pathological significance

(where these are known). It should be added quickly that this is not to suggest that neurological signs are peculiarly objective (as some systems of clerking might suggest): as with all clinical observations, neurological signs are subject to both inter- and intra-observer variation and are biased by prior knowledge of the history and other examination findings.[3-5] As with other elements of clinical examination, relatively little study of the accuracy and precision of neurological signs has been undertaken; a methodology to remedy this situation has been suggested.[6] It is hoped that the current work might encourage more such studies. To those who might suggest that, in an age of molecular genetics, such an undertaking is passé, and rather nineteenth-century in its outlook, I would argue that precision in the definition of clinical signs is of relevance if meaningful genotype/phenotype correlations are to be established.

An attempt has been made to structure the entries in this volume in the following way:

- a definition of the sign, or the common usage of the term (subtypes italicised);
- a brief account of the clinical technique required to elicit the sign;
- a description of other neurological signs which may accompany the index sign (cross referenced as appropriate).

Where known, there is appended:

- a brief account of the neuroanatomical basis of the sign;
- an explanation, where possible, of the pathophysiological and/or pharmacological basis of the sign;
- the neuropathological basis of sign;
- a differential diagnosis of the commonest clinical diseases causing or associated with the sign (bulleted);
- brief details of specific treatments of these disorders, if available.

Using this schema, it will hopefully prove possible to integrate clinical phenomenology with the underlying neuroscience (anatomy, physiology, and pathology) in an accessible manner which will facilitate assimilation by the reader. Clearly not all these factors are known or applicable for every sign, and hence definitions vary quite considerably in length, the longer entries generally being for signs of greater clinical importance. Salient references from the primary and secondary literature are given, particularly for the more uncommon signs, for those wishing to pursue topics further. Entries are cross-referenced to other relevant signs.

Clearly such an undertaking cannot hope to be (and does not claim to be) comprehensive, such is the diversity of neurological function. Moreover, the limitations of my personal clinical experience means that selections are inevitably somewhat arbitrary, precluding (at the very least!) inclusion of signs familiar in paediatric neurological practice. Dermatological signs of potential neurological relevance have also been largely overlooked, and after much consideration "bruit" has been omitted. Nonetheless, it is hoped that this book will be of use to all students of neurology, both undergraduate and postgraduate, both dedicated neurology trainees and those required, perhaps against their personal inclinations, to develop some familiarity with neurology for examination purposes (*e.g.* candidates for the MRCP). It may also serve as a book of reference for more experienced clinicians. Since the majority of patients with neurological symptoms and signs in the United Kingdom are currently seen by general practitioners and general physicians, a situation which is likely to persist for some time, if not indefinitely,[7] it is very much hoped that these groups will also find the book of use, as indeed may members of ancillary professions: nursing, physiotherapy, speech and language therapy, occupational therapy, radiography.

The definitions given are not conceived of as in any way immutable. Language, after all, is plastic with respect to meaning and usage, and my aim is certainly not to "fix" the language. Nor do I suppose, despite my indebtedness to many distinguished colleagues, that I have been free from errors, all of which are my own doing. I shall be happy to hear from those who find errors, disagree with my suggested definitions, or feel that important signs have been omitted.

REFERENCES

1. Ziegler DK. Is the neurologic examination becoming obsolete? *Neurology* 1985; **35**: 559
2. Caplan LR. *The effective clinical neurologist.* Oxford: Blackwell Scientific 1990
3. Stam J, van Crevel H. Reliability of the clinical and electromyographic examination of tendon reflexes. *Journal of Neurology* 1990; **237**: 427-31
4. Maher J, Reilly M, Daly L, Hutchinson M. Plantar power: reproducibility of the plantar response. *BMJ* 1992; **304**: 482
5. Hansen M, Sindrup SH, Christensen PB, *et al.* Interobserver variation in the evaluation of neurological signs: observer dependent factors. *Acta Neurologica Scandinavica* 1994; **90**: 145-9
6. McAlister FA, Straus SE, Sackett DL, on behalf of the CARE-COAD1 Group. Why we need large, simple studies of the clinical examination: the problem and a proposed solution. *Lancet* 1999; **354**: 1721-4
7. *Neurology in the United Kingdom: Towards 2000 and beyond.* London: Association of British Neurologists 1997

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Thanks are also due to Philippa, Thomas and Elizabeth for their forbearance, because, willy-nilly, "The life of every family is conditioned by the work of its elders; think of a doctor's house, or a writer's ..." (Rumer Godden, *The River,* 1946).

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A

Abadie's Sign
Abadie's sign is the absence or diminution of pain sensation when exerting deep pressure on the Achilles tendon by squeezing. This is a frequent finding in the tabes dorsalis variant of neurosyphilis, *i.e.* with dorsal column disease.
[Cross References: ARGYLL ROBERTSON PUPIL]

Abdominal Reflexes
Both superficial and deep abdominal reflexes are described, of which the superficial (cutaneous) reflexes are the more commonly elicited. A wooden stick or pin is used to scratch the abdominal wall, from the flank to the midline, parallel to the line of the dermatomal strips, in upper (supraumbilical), middle (umbilical), and lower (infraumbilical) areas. The manoeuvre is best performed at the end of expiration when the abdominal muscles are relaxed, since the reflexes may be lost with muscle tensing; to avoid this, patients should lie supine with their arms by their sides.

Superficial abdominal reflexes are lost in a number of circumstances:

- normal old age
- obesity
- after abdominal surgery
- after multiple pregnancies
- in acute abdominal disorders (Rosenbach's sign).

However, absence of all superficial abdominal reflexes may be of localising value for corticospinal pathway damage (upper motor neurone lesions) above T6. Lesions at or below T10 lead to selective loss of the lower reflexes with the upper and middle reflexes intact, in which case Beevor's sign may also be present. All abdominal reflexes are preserved with lesions below T12.

Abdominal reflexes are said to be lost early in multiple sclerosis, but late in motor neurone disease; this may be helpful, particularly when differentiating the progressive lateral sclerosis variant of motor neurone disease from multiple sclerosis.
[Cross References: BEEVOR'S SIGN; UPPER MOTOR NEURONE SYNDROME]

Abducent Nerve Palsy
Abducent (VI) nerve palsy causes a selective weakness of the lateral rectus muscle resulting in impaired abduction of the eye, manifest clinically as diplopia on lateral gaze, or on shifting gaze from a near to a distant object. This may be due to:

- Microinfarction in the abducent nerve, due to hypertension, diabetes mellitus;
- Raised intracranial pressure: a "false localising sign", caused by stretching of the nerve over the ridge of the petrous temporal bone;
- Nuclear pontine lesions (congenital, *e.g.* Duane retraction syndrome, Möbius syndrome).

Isolated weakness of the lateral rectus muscle may also occur in myasthenia gravis. In order not to overlook this fact, and miss a potentially treatable condition, it is probably better to label isolated abduction failure as "lateral rectus palsy", rather than abducent nerve palsy, until the aetiological diagnosis is established.
[Cross References: DIPLOPIA; "FALSE LOCALISING SIGNS"]

Absence
An absence, or absence attack, is a brief interruption of awareness of epileptic origin. This is a barely noticeable suspension of speech or attentiveness, without postictal confusion or awareness that an attack has occurred. Idiopathic generalized epilepsy of absence type (petit mal; absence epilepsy) is exclusive to childhood, and is associated with 3 Hz spike and slow wave EEG abnormalities. Absence of this type may be confused with a more obvious distancing or "glazing over", possibly with associated automatisms such as lip smacking, due to a complex partial seizure of temporal lobe origin ("atypical absence").

Ethosuximide and/or valproate are the treatments of choice for idiopathic generalized absence epilepsy, whereas carbamazepine, sodium valproate, or lamotrigine is first choice for complex partial seizures.
[Cross References: AUTOMATISM; SEIZURE]

Abulia
Abulia (aboulia) is a "syndrome of hypofunction", characterized by lack of initiative, spontaneity and drive (aspontaneity), apathy, slowness of thought (bradyphrenia), and blunting of emotional responses and response to external stimuli. It may be confused with the psychomotor retardation of depression and is sometimes labelled as "pseudodepression". More plausibly, abulia has been thought of as a minor or partial form of akinetic mutism. There may also be some clinical overlap with catatonia.

Abulia may result from frontal lobe damage, most particularly that involving the frontal convexity, and has also been reported with focal lesions of the caudate nucleus. As with akinetic mutism, it is likely that lesions anywhere in the "centromedial core" of the brain, from frontal lobes to brainstem, may produce this picture.

Pathologically, abulia may be observed in:

- Parkinson's disease, sometimes as a forerunner of a frontal lobe dementia;
- Anterior cerebral artery infarcts causing basal forebrain damage.

- Bhatia KP, Marsden CD. The behavioural and motor consequences of focal lesions of the basal ganglia in man. *Brain* 1994; **117**: 859-76
- Fisher CM. Abulia. In: Bogousslavsky J, Caplan L (eds.). *Stroke syndromes.* Cambridge: CUP 1995:182-7
[Cross References: AKINETIC MUTISM; APATHY; BRADYPHRENIA; CATATONIA; FRONTAL LOBE SYNDROMES; PSYCHOMOTOR RETARDATION]

Acalculia
Acalculia, or dyscalculia, is difficulty in performing simple mental arithmetic. This depends on two processes, number processing and calculation; a deficit confined to the latter process is termed anarithmetria.

Acalculia may be classified as:

Primary - a specific deficit in arithmetical tasks, more severe than any other co-existing cognitive dysfunction;
Secondary - in the context of impairments of language (aphasia, alexia or agraphia for numbers), attention, memory, and space perception (*e.g.* neglect). Acalculia may occur in association with alexia, agraphia, finger agnosia, right-left disorientation and difficulty spelling words as part of the Gerstmann syndrome with lesions of the dominant parietal lobe.
Secondary acalculia is the commoner variety.

Preservation of calculation skills in the face of total language dissolution (production and comprehension) has been reported in a patient with focal left temporal lobe atrophy probably due to Pick's disease. Isolated acalculia may be seen with lesions of:

- dominant (left) parietal/temporal/occipital cortex, especially involving the angular gyrus (Brodmann areas 39 and 40);
- medial frontal lobe (impaired problem solving ability?);
- subcortical structures (caudate nucleus, putamen, internal capsule).

Impairments may be remarkably focal, for example one operation (*e.g.* subtraction) may be preserved whilst all others are impaired.
In patients with mild to moderate Alzheimer's disease with dyscalculia but no attentional or language impairments, cerebral glucose metabolism was found to be impaired in the left inferior parietal lobule and inferior temporal gyrus.

- Benson DF, Ardila A. *Aphasia:a clinical perspective.* New York: OUP 1996:235-51
- Hirono N, Mori E, Ishii K, *et al.* Regional metabolism: associations with dyscalculia in Alzheimer's disease. *Journal of Neurology, Neurosurgery and Psychiatry* 1998; **65**: 913-6
- Lampl Y, Eshel Y, Gilad R, Sarova-Pinhas I. Selective acalculia with sparing of the subtraction process in a patient with a left parietotemporal hemorrhage. *Neurology* 1994; **44:** 1759-61
- Rossor M, Warrington EK, Cipolotti L. The isolation of calculation skills. *Journal of Neurology* **1995**; 242: 78-81
[Cross References: AGRAPHIA; ALEXIA; APHASIA; GERSTMANN SYNDROME; NEGLECT]

Accommodation Reflex
- see PUPILLARY REFLEXES

Achilles Reflex
- see REFLEXES

Achromatopsia
Achromatopsia, or dyschromatopsia, is an inability or impaired ability to see colours which may be ophthalmological or neurological in origin. Achromatopsia may be congenital or acquired; only in the latter case does the patient complain of impaired colour vision.

Achromatopsia is most conveniently tested for clinically using pseudoisochromatic figures (*e.g.* Ishihara plates), although these were specifically designed for detecting congenital colour blindness and test the red-green channel more than blue-yellow. Sorting colours according to hue, for example with the Farnsworth-Munsell 100 Hue test, is more quantitative, but more time consuming. Difficulty performing these tests does not always reflect achromatopsia (see Pseudoachromatopsia).

Probably the commonest cause of achromatopsia is inherited "colour blindness", of which several types are recognized: in monochromats only one of the three cone photoreceptor classes is affected, in dichromats two; anomalous sensitivity to specific wavelengths of light may also occur (anomalous trichromat). These inherited dyschromatopsias are binocular, symmetrical, and do not change with time.

Acquired achromatopsia may result from damage to the optic nerve or the cerebral cortex. Unlike inherited conditions, these deficits are noticeable (patients describe the world as looking "grey" or "washed out") and may be confined to only part of the visual field (*e.g.* hemiachromatopsia).

Optic neuritis typically impairs colour vision (red-green > blue-yellow) and this defect may persist whilst other features of the acute inflammation (impaired visual acuity, central scotoma) remit.

Cerebral achromatopsia results from cortical damage (most usually infarction) to the inferior occipitotemporal area. Area V4 of the visual cortex, which is devoted to colour processing, is in the occipitotemporal (fusiform) and lingual gyri. Unilateral lesions may produce a homonymous hemiachromatopsia. Lesions in this region may also produce prosopagnosia, alexia, and visual field defects, either a peripheral scitoma which is always in the upper visual field, or a superior quadrantanopia, reflecting damage to the inferior limb of the calcarine sulcus in addition to the adjacent fusiform gyrus. Transient achromatopsia in the context of vertebrobasilar ischaemia has been reported.

The differential diagnosis of achromatopsia encompasses colour agnosia, a loss of colour knowledge despite intact perception; and colour anomia, an inability to name colours despite intact perception.

- Orrell RW, James-Galton M, Stevens JM, Rossor MN. Cerebral achromatopsia as a presentation of Trousseau's syndrome. *Postgraduate Medical Journal* 1995; **71**: 44-6
- Zeki S. A century of cerebral achromatopsia. *Brain* 1990; **113**: 1721-77
[Cross References:AGNOSIA; ALEXIA; ANOMIA; PROSOPAGNOSIA; PSEUDOACHROMATOPSIA;QUADRANTANOPIA, QUADRANTANOPSIA; SCOTOMA]

Action Dystonia
- see DYSTONIA

Action Myoclonus
- see MYOCLONUS

Adiadochokinesia
- see DYSDIADOCHOKINESIA

Adie's Syndrome
Also: Adie's Tonic Pupil
- see HOLMES-ADIE PUPIL, HOLMES-ADIE SYNDROME

Afferent Pupillary Defect (APD)
An afferent pupillary defect (APD), or relative afferent pupillary defect (RAPD), is an abnormal pupillary response in which the normally equal direct and consensual pupillary reflexes are asymmetric, the direct response being less than the consensual; this may be particularly evident using the "swinging flashlight" test, in which the two pupils are alternately illuminated every 2-3 seconds in a darkened room. Quickly moving the light to the diseased side may produce pupillary dilatation (Marcus Gunn pupil). Subjectively patients may note that the light stimulus seems less bright in the affected eye.

APD suggests an asymmetric optic nerve pathology, such as optic neuritis or tumour, causing a conduction defect; indeed this is the most sensitive sign of optic nerve pathology. Although visual acuity may also be impaired in the affected eye, and the disc appear abnormal on fundoscopy, this is not necessarily the case. Since APD depends on asymmetry of optic nerve conduction, no defect may be observed if both optic nerves are affected.

APD has also been described with lesions of the retina, optic chiasm, optic tract, brachium of the superior colliculus and pretectal nucleus (in the latter two situations without visual impairment).
[Cross References: AMBLYOPIA; MARCUS GUNN PUPIL, MARCUS GUNN SIGN; PUPILLARY REFLEXES]

Ageusia
Ageusia or hypogeusia is a loss or impairment of the sense of taste (gustation). This may be tested by application to each half of the protruded tongue the four fundamental tastes (sweet, sour, bitter, and salt).

Isolated ageusia is most commonly encountered as a transient feature associated with coryzal illnesses of the upper respiratory tract, as with anosmia. Indeed, many complaints of loss of taste are in fact due to anosmia, since olfactory sense is responsible for the discrimination of many flavours.

Neurological disorders may also account for ageusia. Afferent taste fibres run in the facial (VII) and glossopharyngeal (IX) cranial nerves, from taste buds in the anterior two-thirds and posterior one-third of the tongue respectively. Central processes run in the solitary tract in the brainstem and terminate in its nucleus (nucleus tractus solitarius), the rostral part of which is sometimes called the gustatory nucleus. Fibres then run to the ventral posterior nucleus of the thalamus, then to the cortical area for taste adjacent to the general sensory area for the tongue (insular region).

Lesions of the facial nerve proximal to departure of the chorda tympani branch in the mastoid (vertical) segment of the nerve (*i.e.* proximal to emergence of the facial nerve from the stylomastoid foramen), can lead to ipsilateral impairment of taste sensation over the anterior two-thirds of the tongue, along with ipsilateral lower motor neurone facial weakness (*e.g.* in Bell's palsy), with or without hyperacusis. Lesions of glossopharyngeal nerve causing impaired taste over the posterior one-third of the tongue

usually occur in association with ipsilateral lesions of the other lower cranial nerves (X, XI, XII; jugular foramen syndrome) and hence may be associated with dysphonia, dysphagia, depressed gag reflex, vocal cord paresis, anaesthesia of soft palate, uvula, pharynx and larynx, and weakness of trapezius and sternocleidomastoid.

Ageusia as an isolated symptom of neurological disease is extremely rare, but has been described with focal central nervous system lesions (infarct, tumour, demyelination) affecting the nucleus of the tractus solitarius (gustatory nucleus) and/or thalamus, and with bilateral insular lesions.

Anosmia and dysgeusia have also been reported following acute zinc loss.

- Finelli PF, Mair RG. Disturbances of taste and smell. In: Bradley WG, Daroff RB, Fenichel GM, Marsden CD (eds.). *Neurology in Clinical Practice.* Boston: Butterworth Heinemann 2000 (3rd edition): 263-9
- Hepburn AL, Lanham JG. Sudden-onset ageusia in the antiphospholipid syndrome. *Journal of the Royal Society of Medicine* 1998; **91**: 640-1
[Cross References: ANOSMIA; BELL'S PALSY; DYSGEUSIA; FACIAL PARESIS; HYPERACUSIS]

Agnosia

Agnosia is a deficit of higher sensory (most often visual) processing causing impaired recognition; the term means literally "absence of knowledge", but its precise clinical definition continues to be a subject of debate. Lissauer (1890) originally conceived of two kinds of agnosia:

Apperceptive - in which there is a defect of complex (higher order) perceptual processes;

Associative - in which perception is thought to be intact but there is a defect in giving meaning to the percept by linking its content with previously encoded percepts; this has been described as "a normal percept that has somehow been stripped of its meaning", or "perception without knowledge".

These deficits should not be explicable by a concurrent intellectual impairment, disorder of attention, or by an inability to name or describe verbally the stimulus (anomia). As a corollary of this last point, there should be no language disorder (aphasia) for the diagnosis of agnosia.

Intact perception is sometimes used as a *sine qua non* for the diagnosis of agnosia, in which case it may be questioned whether apperceptive agnosia is truly agnosia. However, others retain this category, not least because the supposition that perception is normal in associative visual agnosia is probably not true. Moreover, the possibility that some agnosias are in fact higher order perceptual deficits remains: examples include some types of visual and tactile recognition of form or shape (*e.g.* agraphognosia; astereognosis; dysmorphopsia); some authorities label these phenomena "pseudoagnosias". The difficulty with definition perhaps reflects the continuing problem of defining perception at the physiological level.

Theoretically, agnosias can occur in any sensory modality, but some authorities believe that the only unequivocal examples are in the visual and auditory domains (*e.g.* prosopagnosia, and pure word deafness, respectively). Nonetheless, many other "agnosias" have been described, although their clinical definition may lie outwith

some operational criteria for agnosia. With the passage of time, agnosic defects merge into anterograde amnesia (failure to learn new information).

Anatomically, agnosias generally reflect dysfunction at the level of the association cortex, although they can on occasion result from thalamic pathology. Some may be of localizing value. The neuropsychological mechanisms underpinning these phenomena are often poorly understood.

- Damasio AR, Eslinger P. The agnosias. In: *Diseases of the Nervous System: Clinical Neurobiology.* (Asbury AK, McKhann GM, McDonald WI, eds.). London: Heinemann 1986: 839-47
- Farah MJ. *Visual agnosia: disorders of object recognition and what they tell us about normal vision.* Cambridge: MIT Press 1995

[Cross References: AGRAPHOGNOSIA; ALEXIA; AMNESIA; ANOSOGNOSIA; ASOMATOGNOSIA; ASTEREOGNOSIS; AUTOTOPAGNOSIA; DYSMORPHOPSIA; FINGER AGNOSIA; PROSOPAGNOSIA; PURE WORD DEAFNESS; SIMULTANAGNOSIA; VISUAL AGNOSIA]

Agrammatism

Agrammatism is a reduction or loss in the production or comprehension of the syntactic elements of language, for example articles, prepositions, conjunctions, verb endings (*i.e.* the non-substantive components of language), whereas nouns and verbs are relatively spared. Despite this impoverishment of language, or "telegraphic speech", meaning is often still conveyed because of the high information content of verbs and nouns. Agrammatism is encountered in Broca's type of non-fluent aphasia, associated with lesions of the posterior inferior part of the frontal lobe of the dominant hemisphere (Broca's area). Agrammatic speech may also be dysprosodic.

[Cross References: APHASIA; DYSPROSODY]

Agraphaesthesia

Agraphaesthesia, dysgraphaesthesia, or graphanaesthesia, is a loss or impairment of the ability to recognize letters or numbers traced on the skin, *i.e.* of graphaesthesia. Whether this is a perceptual deficit or a tactile agnosia ("agraphognosia") remains a subject of debate. It occurs with damage to the somatosensory parietal cortex.

[Cross References: AGNOSIA]

Agraphia

Agraphia or dysgraphia is a loss or disturbance of the ability to write or spell. Since writing depends not only on preserved language function but also on motor, visuospatial and kinaesthetic function, many factors may lead to dysfunction, *e.g.*

Central, aphasic, or linguistic dysgraphias - these are usually associated with aphasia and alexia, and the deficits mirror those seen in the Broca/anterior and Wernicke/posterior types of aphasia; oral spelling is impaired. From the linguistic viewpoint, two types of paragraphia may be distinguished, *viz.,*

> *surface/lexical/semantic dysgraphia:* misspelling of irregular words, pro-ducing phonologically plausible errors (*e.g.* simtums for symptoms); this is seen with left temporoparietal lesions, *e.g.* Alzheimer's disease, Pick's disease;

deep/phonological dysgraphia: inability to spell unfamiliar words and non-words; semantic errors; seen with extensive left hemisphere damage.

Mechanical agraphia - impaired motor control, due to paresis (as in dominant parietal damage), dyspraxia (may be accompanied by ideomotor limb apraxia), dyskinesia (hypokinetic or hyperkinetic), or dystonia; oral spelling may be spared.

Neglect (spatial) dysgraphia - associated with other neglect phenomena consequent upon a non-dominant hemisphere lesion; there may be missing out or misspelling of the left side of words (paragraphia); oral spelling may be spared.

Pure agraphia - a rare syndrome in which oral language, reading and praxis are normal.

A syndrome of agraphia, alexia, acalculia, finger agnosia, right-left disorientation and difficulty spelling words (Gerstmann syndrome) may be seen with dominant parietal lobe pathologies.

Writing disturbance due to abnormal mechanics of writing is the most sensitive language abnormality in delirium, possibly because of its dependence on multiple functions.

- Benson DF, Ardila A. *Aphasia: a clinical perspective. New* York: OUP 1996:212-34
[Cross References: ALEXIA; APHASIA; APRAXIA; BROCA'S APHASIA; GERSTMANN SYNDROME; HYPERGRAPHIA; MACROGRAPHIA; MICROGRAPHIA; NEGLECT; WERNICKE'S APHASIA]

Agraphognosia

- see AGRAPHAESTHESIA

Akathisia

Akathisia is a feeling of inner restlessness, often associated with restless movements of a continuous and often purposeless nature (*e.g.* rocking to and fro, repeatedly crossing and uncrossing the legs, standing up and sitting down, pacing up and down). Voluntary suppression of the movements may exacerbate inner tension or anxiety.

Recognized associations include Parkinson's disease and neuroleptic medication (acute or tardive side effect), suggesting that dopamine depletion may contribute to pathophysiology; dopamine depleting agents (*e.g.* tetrabenazine, reserpine) may cause akathisia.

Treatment by reduction or cessation of neuroleptic therapy may help, but can exacerbate coexistent psychosis. Centrally acting β-blockers such as propranolol may also help, as may anticholinergic agents, amantadine, clonazepam, and clonidine.

-Sachdev P. *Akathisia and restless legs.* Cambridge: CUP 1995
[Cross References: PARKINSONISM; TIC]

Akinesia

Akinesia is an inability or, more usually in clinical practice, a reduced or delayed ability to initiate voluntary movement (the latter may be better termed hypokinesia). This cannot be attributed to motor unit or pyramidal system dysfunction. Reflexive motor activity may be preserved (*kinesis paradoxica*). There may be concurrent slowness of movement (bradykinesia) and reduction in amplitude of movements

(hypometria). Akinesia may co-exist with any of the other clinical features of extrapyramidal system disease, particularly rigidity, but the presence of akinesia is regarded as an absolute requirement for the diagnosis of parkinsonism. Hemiakinesia may be a feature of motor neglect of one side of the body (possibly a motor equivalent of sensory extinction). Bilateral akinesia with mutism (akinetic mutism) may occur if pathology is bilateral.

Neuroanatomically, akinesia is a feature of disorders affecting:

- frontal-subcortical structures, *e.g.* the medial convexity subtype of frontal lobe syndrome;
- basal ganglia;
- ventral thalamus;
- limbic system (anterior cingulate gyrus).

Neurophysiologically, akinesia is associated with loss of dopamine projections from the substantia nigra to the putamen.

Pathological processes underpinning akinesia include:

- neurodegeneration, *e.g.* Parkinson's disease, Steele-Richardson-Olszewski syndrome, striatonigral degeneration; it may occur late in Pick's disease and Alzheimer's disease;
- hydrocephalus;
- neoplasia, *e.g.* butterfly glioma of the frontal lobes;
- cerebrovascular disease.

Akinesia resulting from nigrostriatal dopamine depletion (*i.e.* idiopathic Parkinson's disease) may respond to treatment with levodopa or dopamine agonists. However, many parkinsonian/akinetic-rigid syndromes show no or only partial response to these agents.

[Cross References: AKINETIC MUTISM; BRADYKINESIA; EXTINCTION; FRONTAL LOBE SYNDROMES; HEMIAKINESIA; HYPOKINESIA; HYPOMETRIA; *KINESIS PARADOXICA;* NEGLECT; PARKINSONISM]

Akinetic Mutism

Akinetic mutism is a "syndrome of negatives", characterized by lack of voluntary movement (akinesia), absence of speech (mutism), lack of response to question and command, but with normal alertness and sleep-wake cycles (*cf.* coma). Blinking (spontaneous and to threat) is preserved. Frontal release signs, such as grasping and sucking, may be present, as may double incontinence, but there is a relative paucity of upper motor neurone signs affecting either side of the body, suggesting relatively preserved descending pathways. Abulia has been characterized as a lesser form of akinetic mutism.

Pathologically, akinetic mutism is associated with bilateral lesions of the "centromedial core" of the brain, at any point from frontal lobes to brainstem, which interrupt reticular-cortical or limbic-cortical pathways but which spare corticospinal pathways:

- anterior cingulate cortex (medial frontal region);
- paramedian reticular formation, posterior diencephalon, hypothalamus
- subacute communicating hydrocephalus

Other structures (*e.g.* globus pallidus) have been implicated but without pathological evidence.

These pathologies may be vascular, neoplastic, or structural (hydrocephalus). Akinetic mutism may be the final state common to the end-stages of a number of neurodegenerative pathologies.

Occasionally, treatment of the cause may improve akinetic mutism (*e.g.* relieving hydrocephalus). Agents such as dopamine agonists (*e.g.* bromocriptine) and ephedrine have also been tried.

- Cairns H. Disturbances of consciousness with lesions of the brain stem and diencephalon. *Brain* 1952; **75**: 109-46

[Cross References: ABULIA; AKINESIA; BLINK REFLEX; COMA; FRONTAL LOBE SYNDROMES; FRONTAL RELEASE SIGNS; GRASP REFLEX; LOCKED-IN SYNDROME; MUTISM]

Akinetic Rigid Syndrome

- see PARKINSONISM

Akinetopsia

Akinetopsia is a specific inability to see objects in motion, the perception of other visual attributes such as colour, form, and depth, remaining intact (statokinetic dissociation is known as Riddoch's phenomenon); the syndrome may also be called cerebral visual motion blindness. Such cases, although exceptionally rare, demonstrate the separateness of movement vision, as do cases in which motion vision is selectively spared in a scotomatous area (Riddoch's syndrome).

Akinetopsia reflects a lesion selective to area V5 of the visual cortex. It may be associated with acalculia and aphasia.

- Zihl J, Von Cramon D, Mai N. Selective disturbance of movement vision after bilateral brain damage. *Brain* 1983; **106**: 313-40
- Zeki S. Cerebral akinetopsia (cerebral visual motion blindness). *Brain* 1991; **114:** 811-24

[Cross References: ACALCULIA; APHASIA; RIDDOCH'S PHENOMENON]

Alexia

Alexia or dyslexia is a disorder of reading. Alexia may be categorised as:

Peripheral - a defect of perception or decoding written script (other language functions often intact); or

Central - a breakdown in deriving meaning (other language functions are often also affected).

Peripheral alexias include:

Alexia without agraphia, also known as pure alexia or pure word blindness. This is the archetypal peripheral alexia. Patients lose the ability to recognise written words quickly and easily; they seem unable to process all the elements of a written word in parallel. They can still access meaning but adopt a laborious letter-by-letter strategy for reading, with a marked word-length effect (*i.e.* greater difficulty reading longer

words). Patients with pure alexia may be able to identify and name individual letters, but some cannot manage even this ("global alexia"). Strikingly the patient can write at normal speed (*i.e.* no agraphia) but is then unable to read what they have just written. Alexia without agraphia often coexists with a right homonymous hemianopia, and colour anomia or impaired colour perception (achromatopsia); this may be only in one hemifield, classically right (hemiachromatopsia). Pure alexia has been characterized by some authors as a limited form of associative visual agnosia or ventral simultanagnosia.

Hemianopic alexia occurs when a right homonymous hemianopia encroaches into central vision. Patients tend to be slower with text than single words as they cannot plan rightward reading saccades.

Neglect alexia (or hemiparalexia) results from failure to read either the beginning or end of a word (more commonly the former) in the absence of a hemianopia, due to hemispatial neglect.

The various forms of peripheral alexia may coexist; following a stroke, patients may present with global alexia which evolves to a pure alexia over the following weeks. Pure alexia is caused by damage to the left occipito-temporal junction, its afferents from early mesial visual areas, or its efferents to the medial temporal lobe. Global alexia usually occurs when there is additional damage to the splenium or white matter above the occipital horn of the lateral ventricle. Hemianopic alexia is usually associated with infarction in the territory of the posterior cerebral artery damaging geniculostriate fibres or area VI itself, but can be caused by any lesion outside the occipital lobe that causes a macular splitting homonymous field defect. Neglect alexia is usually caused by occipito-parietal lesions, right sided lesions causing left neglect alexia.

Central (linguistic) alexias include:

Patients with aphasia who often have coexistent difficulties with reading (reading aloud and/or comprehending written text) and writing (alexia with agraphia, such patients may have a complete or partial Gerstmann syndrome, the so-called "third alexia" of Benson). The reading problem parallels the language problem; thus in Broca's aphasia reading is laboured with particular problems reading function words (of, at) and verb inflections (-ing, -ed); in Wernicke's aphasia numerous paraphasic errors are made.

From the linguistic viewpoint, two types of paralexia (substitution in reading) may be distinguished:

Surface dyslexia - in which there are regularization errors with exception words (*e.g.* pint pronounced to rhyme with mint); this may be seen with left temporoparietal infarction, temporal lobe Pick's disease, late Alzheimer's disease.

Deep dyslexia - the inability to translate orthography to phonology, manifesting as an inability to read plausible non-words, and semantic errors related to word meaning rather than sound (*e.g.* sister read as uncle); visual errors are also common (*e.g.* sacred read as scared). Deep dyslexia is seen with extensive left hemisphere damage.

- Benson DF, Ardila A. *Aphasia: a clinical perspective.* New York: OUP 1996: 180-211

- Binder JR, Mohr JP. The topography of callosal reading pathways: a case control analysis. *Brain* 1992; **115:** 1807-26
- Farah MJ. *Visual agnosia: disorders of object recognition and what they tell us about normal vision.* Cambridge: MIT Press 1995
[Cross References: ACHROMATOPSIA; ACALCULIA; AGNOSIA; AGRAPHIA; APHASIA; BROCA'S APHASIA; GERSTMANN SYNDROME; MACULA SPARING, MACULA SPLITTING; NEGLECT; PROSOPAGNOSIA; SIMULTANAGNOSIA; VISUAL AGNOSIA; WERNICKE'S APHASIA]

"Alice in Wonderland Syndrome"
- see METAMORPHOPSIA

Alien Grasp Reflex
The term alien grasp reflex has been used to describe a grasp reflex occurring in full consciousness, which the patient could anticipate but perceived as alien (*i.e.* not modified by will); it occurred in the absence of other abnormal movements. These phenomena were associated with an intrinsic tumour of the right (non-dominant) frontal lobe. It was suggested that the grasp reflex and alien hand syndromes are not separate entities but part of the spectrum of frontal lobe dysfunction, the term "alien grasp reflex" attempting to emphasize the overlap.

- Silva MT, Howard RS, Kartsounis LD, Ross Russell RW. The alien grasp reflex. *European Neurology* 1996; **36**: 55-6
[Cross References: ALIEN LIMB; GRASP REFLEX]

Alien Limb
An alien limb is one which manifests slow, involuntary, wandering (levitating), quasi-purposive movements, most usually the arm but occasionally the leg. An arm so affected may show apraxic difficulties in performing even the simplest tasks, and may be described by the patient as uncooperative or having "a mind of its own" (*le main étranger).* This phenomenon is often associated with a prominent grasp reflex, forced groping, intermanual conflict, and magnetic movements (*q.v.*) of the hand.

Different types of alien hand have been described, reflecting the differing anatomical locations of underlying lesions. A *callosal* type, characterized primarily by intermanual conflict, has been differentiated from a *frontal* type which shows features of environmental dependency such as forced grasping and groping and utilization behaviour. A rarer *sensory* or posterior variant has been contrasted with these anterior or motor types of alien limb, resulting from a combination of cerebellar, optic, and sensory ataxia. A paroxysmal alien hand has been described, probably related to seizures of frontomedial origin.

Recognized pathological associations of alien limb include:

- corticobasal (ganglionic) degeneration
- corpus callosum tumours, haemorrhage
- medial frontal cortex infarction (territory of the anterior cerebral artery)
- trauma and haemorrhage affecting both corpus callosum and medial frontal area
- Alzheimer's disease (very rare)

- posterior cerebral artery occlusion (sensory variant)
- following commissurotomy (corpus callosotomy alone insufficient).

Functional imaging studies in corticobasal degeneration, along with the evidence from focal vascular lesions, suggest that damage to and/or hypometabolism of the medial frontal cortex (Brodmann area 32) and the supplementary motor area (Brodmann area 6) are associated with alien limb phenomena. More generally, it seems that these areas are involved in the execution of learned motor programs, and damage thereto may lead to the release of learned motor programs from voluntary control.

- Ay H, Buonanno FS, Price BH, Le DA, Koroshetz WJ. Sensory alien hand syndrome: case report and review of the literature. *Journal of Neurology, Neurosurgery and Psychiatry* 1998; **65:** 366-9
- Feinberg TE, Schindler RJ, Flanagan NG, Haber LD. Two alien hand syndromes. *Neurology* 1992; **42**: 19-24
- Sawle GV, Brooks DJ, Marsden CD, Frackowiak RSJ. Corticobasal degeneration: a unique pattern of regional cortical oxygen hypometabolism and striatal fluorodopa uptake demonstrated by positron emission tomography. *Brain* 1991; **114:** 541-56
[Cross References: ALIEN GRASP REFLEX; APRAXIA; ARM LEVITATION; ATAXIA; FORCED GROPING; GRASP REFLEX; INTERMANUAL CONFLICT; MAGNETIC MOVEMENTS; UTILIZATION BEHAVIOUR]

Alloaesthesia

Alloaesthesia (allesthesia, alloesthesia) is the condition in which a sensory stimulus given to one side of the body is perceived at the corresponding area on the other side after a half-second delay. The trunk and proximal limbs are affected more often than the face or distal limbs. This phenomenon may also be termed allochiria. Visual alloaesthesia, the transposition of an object seen in one visual field to the contralateral visual field, is also described, for example in "top of the basilar" syndrome.

Tactile alloaesthesia may be seen in the acute stage of right putaminal haemorrhage (but seldom in right thalamic haemorrhage) and occasionally with anterolateral spinal cord lesions.

The mechanism is uncertain: some consider it a disturbance within sensory pathways, others that it is a sensory response to neglect.

- Kawamura M, Hirayama K, Shinohara Y, Watanabe Y, Sugishita M. Alloaesthesia. *Brain* 1987; **110:** 225-36
[Cross References: ALLOKINESIA; NEGLECT]

Allochiria

- see ALLOAESTHESIA

Allodynia

Allodynia is the elicitation of pain by light mechanical stimuli (such as touch or light pressure) which do not normally provoke pain (*cf.* hyperalgesia), *i.e.* this is a positive sensory phenomenon. Examples of allodynia include the trigger points of trigeminal neuralgia, the affected skin in areas of causalgia, and some peripheral neuropathies; it may also be provoked, paradoxically, by prolonged morphine use.

Various pathogenetic mechanisms are considered possible, including sensitization (lower threshold, hyperexcitability) of peripheral cutaneous nociceptive fibres (in which neurotrophins may play a role); ephaptic transmission ("cross-talk") between large and small (nociceptive) fibres; and abnormal central processing.

The treatment of neuropathic pain is typically with anticonvulsants such as carbamazepine and gabapentin. Interruption of sympathetic outflow, for example with regional guanethidine blocks, may sometimes help, but relapse may occur.

[Cross References: HYPERALGESIA; HYPERPATHIA]

Allokinesia

Allokinesia is a motor response in the wrong limb, *i.e.* contralateral to the side of intended movement, or in the wrong direction. It may be a motor system counterpart of alloaesthesia, and is seen with right hemisphere lesions as part of a neglect syndrome.

[Cross References: ALLOAESTHESIA; NEGLECT]

Alternate Cover Test

- see COVER TEST, COVER-UNCOVER TEST

Alternating Sequences Test

- see APRAXIA; FRONTAL LOBE SYNDROMES

Altitudinal Field Defect

Altitudinal visual field defects respect the horizontal meridian, and are characteristic of (but not exclusive to) disease in the distribution of the central retinal artery. Central vision may be preserved (macula sparing) because the blood supply of the macula often comes from the cilioretinal arteries.

Causes of altitudinal visual field defects include:

- Monocular:
  - Central retinal artery occlusion (CRAO)
  - Acute ischaemic optic neuropathy (AION)
  - Retinal detachment
  - Choroiditis
  - Glaucoma
  - Chronic atrophic papilloedema
- Bilateral:
  - Sequential CRAO, AION
  - Bilateral occipital lesions

[Cross References: MACULA SPARING and SPLITTING; QUADRANTANOPIA]

Amaurosis

Amaurosis is visual loss, with the implication that this is not due to refractive error or intrinsic ocular disease. The term is most often used in the context of amaurosis fugax, a transient monocular blindness, which is most often due to embolism from a stenotic ipsilateral internal carotid artery. Systemic lupus erythematosus and the antiphospholipid antibody syndrome are also recognised causes.

Amblyopia
Amblyopia refers to poor visual acuity, most usually in the context of a "lazy eye", in which the poor acuity results from the failure of the eye to establish normal cortical representation of visual input during the critical period of visual maturation (between the ages of six months and three years). This may result from:

- strabismus
- uncorrected refractive error
- stimulus deprivation

Amblyopic eyes may demonstrate a relative afferent pupillary defect, and sometimes latent nystagmus.

Amblyopia may not become apparent until adulthood when the patient suddenly becomes aware of unilateral poor vision. The finding of a latent strabismus (heterophoria) may be a clue to the fact that such visual loss is long-standing.

The bilateral simultaneous development of central or centrocaecal scotomas in chronic alcoholics has often been referred to as tobacco-alcohol amblyopia, although probably better termed nutritional optic neuropathy.

[Cross References: AFFERENT PUPILLARY DEFECT; ESOTROPIA; HETEROPHORIA; NYSTAGMUS; SCOTOMA]

Amimia
- see HYPOMIMIA

Amnesia
Amnesia is an impairment of episodic memory, or memory for personally experienced events. This is a component of long-term (as opposed to working) memory which is distinct from memory for facts (semantic memory), in that episodic memory is unique to the individual whereas semantic memory encompasses knowledge held in common by members of a cultural or linguistic group. Episodic memory generally accords with the lay perception of memory. A precise clinical definition for amnesia has not been demarcated, perhaps reflecting the heterogeneity of the syndrome.

Amnesia may be retrograde (for events already experienced) or anterograde (for newly experienced events). Retrograde amnesia may show a temporal gradient, with distant events being better recalled than more recent ones, relating to the duration of anterograde amnesia. Amnesia may be acute and transient or chronic and persistent. In a pure amnesic syndrome, intelligence and attention are normal and skill acquisition (procedural memory) is preserved. Amnesia may occur as one feature of more widespread cognitive impairments, *e.g.* in Alzheimer's disease.

The neuroanatomical substrate of episodic memory is a distributed system in the medial temporal lobe and diencephalon surrounding the 3rd ventricle (the circuit of Papez) comprising the entorhinal area of the parahippocampal gyrus, perforant and alvear paths, hippocampus, fimbria/fornix, mammillary bodies, mammillothalamic tract, anterior thalamic nuclei, internal capsule, cingulate gyrus, and cingulum. Basal forebrain structures (septal nucleus, diagonal band nucleus of Broca, nucleus basalis of Meynert) are also involved.

Classification of amnesic syndromes into subtypes has been proposed, since lesions in different areas produce different deficits reflecting functional subdivision within the system; thus left temporal lesions produce problems in the verbal domain, right sided lesions affect non-verbal/visual memory. A distinction between medial temporal pathology (*e.g.* hippocampus), leading to difficulty encoding new memories (anterograde amnesia and temporally limited retrograde amnesia), and diencephalic pathology (*e.g.* Korsakoff's syndrome), which causes difficulty retrieving previously acquired memories (extensive retrograde amnesia) with diminished insight and a tendency to confabulation, has been suggested, but overlap may occur. A frontal amnesia has also been suggested, although impaired attentional mechanisms may contribute.

Various psychometric tests of episodic memory are available. These include the Wechsler Memory Score (WMS-R), the Recognition Memory Test which has both verbal (words) and visual (faces) subdivisions, the Rey Auditory Verbal Learning Test (immediate and delayed free recall of a random word list), and the Rey-Osterreith Complex Figure (non-verbal memory). Retrograde memory may be assessed with a structured autobiographical interview, and with the Famous Faces Test.

There are many causes of amnesia including:

Acute/transient:
- Transient global amnesia
- Transient epileptic amnesia
- Closed head injury
- Drugs

Chronic/persistent:
- Sequel of herpes simplex encephalitis
- Alzheimer's disease (may show isolated amnesia in early disease)
- Limbic (paraneoplastic) encephalitis
- Hypoxic brain injury
- Temporal lobectomy (bilateral; or unilateral with previous contralateral injury, usually birth asphyxia)
- Bilateral posterior cerebral artery occlusion
- Korsakoff's syndrome
- Bilateral thalamic infarction
- Third ventricle tumour, cyst

Few of these are amenable to treatment. Functional or psychogenic amnesia involves failure to recall basic autobiographical details such as name.

- Hodges JR, Greene JDW: Disorders of memory. In: Kennard C (ed.). *Recent advances in clinical neurology 8.* Edinburgh: Churchill Livingstone 1995, pp 151-69
- O'Connor M, Verfaellie M, Cermak LS. Clinical differentiation of amnesic subtypes. In: Baddeley AD, Wilson BA, Watts FN (eds.). *Handbook of memory disorders.* Chichester: John Wiley 1995: 53-80
- Parkin AJ (ed.). *Case studies in the neuropsychology of memory.* Hove: Psychology Press 1997

[Cross References: CONFABULATION; DEMENTIA]

Amusia

Amusia is a loss of the ability to appreciate music; this may be receptive, or expressive (*e.g.* loss of ability to sing, whistle); tests for the evaluation of amusia have been described. Clearly a premorbid appreciation of music is a *sine qua non* for the diagnosis, and most cases reported occur in trained musicians.

Amusia may occur in the context of more widespread cognitive dysfunction, such as aphasia and agnosia. It has been found in association with pure word deafness. Isolated amusia has been reported in the context of focal cerebral atrophy affecting the non-dominant temporal lobe. However, functional studies have failed to show strong hemispheric specificity for music perception, but suggest a cross-hemispheric fragmented neural substrate.

- Confavreux C, Croisile B, Garassus P, Aimard G, Trillet M. Progressive amusia and aprosody. *Archives of Neurology* 1992; **49**: 971-6
- Schuppert M, Münte TF, Wieringa BM, Altenmüller E. Receptive amusia: evidence for cross-hemispheric neural networks underlying music processing strategies. *Brain* 2000; **123**: 546-59
- Wertheim N. The amusias. In: Vinken PJ, Bruyn GW (eds.). *Handbook of clinical neurology, Vol. 4: Disorders of speech, perception, and symbolic behaviour.* Amsterdam: North-Holland Publishing, 1969: 195-206
[Cross References: PURE WORD DEAFNESS]

Amyotrophy

Amyotrophy is a thinning or wasting (atrophy) of musculature with attendant weakness. This may result from involvement of:

- Lower motor neurones (in which case fasciculations may also be present):
  amyotrophic lateral sclerosis
  benign focal amyotrophy/monomelic amyotrophy
  disinhibition-dementia-parkinsonism-amyotrophy complex (DDPAC)
  amyotrophic Creutzfeldt-Jakob disease
  "asthmatic amyotrophy" (Hopkins' syndrome)
- Nerve roots:
  diabetic amyotrophy (polyradiculopathy, especially L2-L4)
- Plexus:
  Neuralgic amyotrophy (Parsonage-Turner syndrome)

Hence although the term implies neurogenic (as opposed to myogenic) muscle wasting, its use is non-specific with respect to neuroanatomical implications.
[Cross References: FASCICULATION; NEUROPATHY; PLEXOPATHY; RADICULOPATHY; WASTING]

Anaesthesia

Anaesthesia (anesthesia) is a complete loss of sensation; hypoaesthesia (hypaesthesia, hypesthesia) is a diminution of sensation (*i.e.* negative sensory phenomena). Anaesthesia may involve all sensory modalities (global anaesthesia, as in general surgical anaesthesia) or be selective (*e.g.* thermoanaesthesia, analgesia). Regional patterns of anaesthesia are described, *e.g.* "glove-and-stocking anaesthesia" in peripheral neuro-

pathies, "saddle anaesthesia" involving S3-5 dermatomes resulting from a cauda equina syndrome.

Anaesthesia is most often encountered after resection or lysis of a peripheral nerve segment, whereas paraesthesia or dysaesthesia reflect damage to a nerve which is still in contact with the cell body.

Anaesthesia dolorosa is a persistent unpleasant pain (*i.e.* a positive sensory phenomenon) which may be experienced in the distribution of a resected nerve, *e.g.* following treatment for trigeminal neuralgia. This may respond to various medications, including tricyclic antidepressants, carbamazepine, and selective serotonin reuptake inhibitors.

[Cross References: ANALGESIA; DYSAESTHESIA; NEUROPATHY; PARAESTHESIA]

Analgesia

Analgesia or hypoalgesia is a complete loss or diminution of pain sensation, or the absence of a pain response to a normally painful stimulus. These negative sensory phenomena may occur as one component of total sensory loss (anaesthesia) or in isolation. Consequences of analgesia include the development of neuropathic ulcers, burns, Charcot joints, even painless mutilation or amputation.

Analgesia may occur in:

- peripheral nerve lesions, *e.g.* hereditary sensory and autonomic neuropathies (HSAN), leprosy;
- central spinal cord lesions which pick off the decussating fibres of the spinothalamic pathway in the ventral funiculus (with corresponding thermoanaesthesia), *e.g.* syringomyelia;
- cortical lesions, *e.g.* medial frontal lobe syndrome (akinetic type).

Congenital syndromes of insensitivity to pain were once regarded as a central pain asymbolia (*e.g.* Osuntokun's syndrome), but on further follow-up some have turned out to be variants of HSAN.

- Larner AJ, Moss J, Rossi ML, Anderson M. Congenital insensitivity to pain: a 20 year follow up. *Journal of Neurology, Neurosurgery and Psychiatry* 1994; **57**: 973-4

[Cross References: ANAESTHESIA; FRONTAL LOBE SYNDROMES]

Anarithmetria

- see ACALCULIA

Anarthria

Anarthria is the complete inability to articulate words (*cf.* dysarthria). This is most commonly seen in bulbar motor neurone disease.

A motor disorder of speech production with preserved comprehension of spoken and written language has been termed pure anarthria; this syndrome has also been called aphemia, phonetic disintegration, apraxic dysarthria, cortical dysarthria, verbal apraxia, subcortical motor aphasia, pure motor aphasia, and small or mini Broca's aphasia. It reflects damage in the left frontal operculum, but sparing Broca's area.

A pure progressive anarthria may result from focal degeneration affecting the frontal operculum bilaterally (so-called Foix-Chavany-Marie syndrome).

- Lecours AR, Lhermitte F. The "pure" form of the phonetic disintegration syndrome (pure anarthria): anatomo-clinical report of a single case. *Brain and Language* 1976; **3**:88-113.
[Cross References: APHEMIA; DYSARTHRIA]

Anhidrosis
Anhidrosis, or hypohidrosis, is a loss or lack of sweating. This may be due to primary autonomic failure, or to pathology within the posterior hypothalamus ("sympathetic area"); if unilateral, an ipsilateral Horner's syndrome may be seen. Anhidrosis may occur in other CNS disorders, including multisystem atrophy, Parkinson's disease, multiple sclerosis, and below a spinal cord lesion. Localised or generalised anhidrosis may be seen in Holmes-Adie syndrome.
[Cross References: HOLMES-ADIE PUPIL, HOLMES-ADIE SYNDROME; HORNER'S SYNDROME; HYPERHIDROSIS]

Anismus
Anismus is paradoxical contraction of the external anal sphincter during attempted defaecation, leading to faecal retention and a complaint of constipation. This may occur as an idiopathic condition in isolation, or as a feature of the off periods of Parkinson's disease. It is thought to represent a focal dystonia, and may be helped by local injections of botulinum toxin.

- Jost WH, Muller-Lobeck H, Merkle W. Involuntary contractions of the striated anal sphincters as a cause of constipation: report of a case. *Diseases of the Colon and Rectum* 1998; **41**: 258-60
[Cross References: DYSTONIA]

Anisocoria
Anisocoria is an inequality of pupil size. This may be physiological, in which case the inequality is usually mild and does not vary with degree of ambient illumination; or pathological, with many possible causes:

- Affected pupil constricted (miosis; oculosympathetic paresis):
  - Horner's syndrome
  - Argyll Robertson pupil
  - Cluster headache
- Affected pupil dilated (mydriasis; oculoparasympathetic paresis):
  - Holmes-Adie pupil (vermiform movements of the pupil margin may be visible with a slit-lamp)
  - Oculomotor (III) nerve palsy (outflow from Edinger-Westphal nucleus)
  - Mydriatic agents (phenylephrine, tropicamide)
  - Anticholinergic agents (*e.g.* asthma inhaler accidentally puffed into one eye)

[Cross References: ARGYLL ROBERTSON PUPIL; HOLMES-ADIE PUPIL; MIOSIS; MYDRIASIS]

Anomia

Anomia or dysnomia is a deficit in naming or word-finding. This may be detected as abrupt cut-offs in spontaneous speech with circumlocutions and/or paraphasic substitutions. Formal tests of naming are also available (*e.g.* Graded Naming Test). Patients may be able to point to named objects despite being unable to name them, suggesting a problem in word retrieval but with preserved comprehension.

Category-specific anomias have been described, *e.g.* for colour (*cf.* achromatopsia). Anomia occurs with pathologies affecting the left temporoparietal area, but since it occurs in all varieties of aphasia is of little precise localizing or diagnostic value. The term anomic aphasia is reserved for unusual cases in which a naming problem overshadows all other deficits. Anomia may often be seen as a residual deficit following recovery from other types of aphasia, as an early feature of Alzheimer's disease, or with any dominant hemisphere space-occupying lesion.

- Benson DF, Ardila *A.Aphasia: a clinical perspective.* New York:OUP 1996: 252-61
[Cross References: APHASIA; CIRCUMLOCUTION; PARAPHASIA]

Anosmia

Anosmia is the inability to perceive smells due to damage to the olfactory pathways (olfactory neuroepithelium, olfactory nerves, rhinencephalon). Olfaction may be tested with kits containing specific odours (*e.g.* clove, turpentine); each nostril should be separately tested. Unilateral anosmia may be due to pressure on the olfactory bulb or tract, *e.g.* due to a subfrontal meningioma.

Anosmia may be congenital (*e.g.* Kallman's syndrome, hypogonadotrophic hypogonadism, a disorder of neuronal migration) or, much more commonly, acquired. Rhinological disease (allergic rhinitis, coryza) is by far the commonest cause; this may also account for the impaired sense of smell in smokers. Head trauma is the commonest neurological cause, due to shearing off of the olfactory fibres as they pass through the cribriform plate. Recovery is possible due to the capacity for neuronal and axonal regeneration within the olfactory pathways. Olfactory dysfunction is also described in Alzheimer's disease and Parkinson's disease. Patients with depression may also complain of impaired sense of smell.

- Finelli PF, Mair RG. Disturbances of taste and smell. In: Bradley WG, Daroff RB, Fenichel GM, Marsden CD (eds.). *Neurology in Clinical Practice.* Boston: Butterworth Heinemann 2000 (3rd edition): 263-9
[Cross References: AGEUSIA; DYSGEUSIA; PAROSMIA]

Anosodiaphoria

Babinski (1914) used the term anosodiaphoria to describe a disorder of body schema in which patients verbally acknowledge a clinical problem (*e.g.* hemiparesis) but fail to be concerned by it. Anosodiaphoria usually follows a stage of anosognosia. *La belle indifférence* describes a similar lack of concern for acknowledged disabilities which are psychogenic.

[Cross References: ANOSOGNOSIA; *BELLE INDIFFÉRENCE*]

Anosognosia

Anosognosia is a patient's unawareness or denial of his or her illness. The term was first used by von Monakow (1885) and has been used to describe denial of blindness (Anton's syndrome), deafness, hemiplegia (Babinski), hemianopia, aphasia, and amnesia. Some authorities would question whether this unawareness is a true agnosia, or rather a defect of higher level cognitive integration (*i.e.* perception).

Anosognosia with hemiplegia most commonly follows right hemisphere injury (parietal and temporal lobes) and may be associated with left hemi-neglect and left-sided hemianopia; it is also described with right thalamic and basal ganglia lesions. Many patients with posterior aphasia (Wernicke type) are unaware that their output is incomprehensible or jargon, possibly through a failure to monitor their own output. Cerebrovascular disease is the commonest cause of anosognosia.

The neuropsychological mechanism of anosognosia is unclear: the hypothesis that it might be accounted for by personal neglect (asomatognosia), which is also more frequently observed after right hemisphere lesions, would seem to have been disproved experimentally by studies using selective hemisphere anaesthesia in which the two may be dissociated. Moreover, the two may be dissociated clinically. In Alzheimer's disease, anosognosia is related to memory dysfunction but not to frontal lobe features. At a practical level, anosognosia may lead to profound difficulties with neurorehabilitation.

- McGlynn S, Schacter DL. Unawareness of deficits in neuropsychological syndromes. *Journal of Clinical and Experimental Neuropsychology* 1989; **11**: 143-205
- Starkstein SE, Fedorof JP, Price TR, Leiguarda R, Robinson RG. Anosognosia in patients with cerebrovascular lesions: a study of causative factors. *Stroke* 1992; **23**: 1446-53.

[Cross References: AGNOSIA; ANOSODIAPHORIA; ASOMATOGNOSIA; CORTICAL BLINDNESS; EXTINCTION; JARGON APHASIA; NEGLECT]

Antecollis

Antecollis (anterocollis) is forward flexion of the neck. It may be a feature of multisystem atrophy (*cf.* retrocollis in Steele-Richardson-Olszewski syndrome), a sustained dystonic posture in advanced Parkinson's disease, and, unusually, in spasmodic torticollis.

[Cross References: RETROCOLLIS; TORTICOLLIS]

Anton's Syndrome

- see ANOSOGNOSIA; CONFABULATION; CORTICAL BLINDNESS

Apallic Syndrome

- see VEGETATIVE STATE

Apathy

Apathy is a neurobehavioural disorder characterized by a lack of interest in environmental stimuli, manifest as listlessness, paucity of spontaneous movement (akinesia) or speech (mutism), and lack of initiative, spontaneity and drive. These are all features of the abulic state, and it has been suggested that apathy and abulia represent

different points on a continuum of motivational and emotional deficit, abulia being at the more severe end. The diminished motivation of apathy should not be attributable to impaired level of consciousness, emotional distress, or cognitive impairment although it may coexist with the latter, as in Alzheimer's disease.

Apathy may be observed in diseases affecting frontal-subcortical structures, for example in the frontal lobe syndrome affecting the frontal convexity, or following multiple vascular insults to paramedian diencephalic structures (thalamus, subthalamus, posterior lateral hypothalamus, mesencephalon) or the posterior limb of the internal capsule; there may be associated cognitive impairment of the so-called "subcortical" type in these situations. Apathy is also described following amphetamine or cocaine withdrawal, in neuroleptic-induced akinesia and in psychotic depression.

- Marin RS. Differential diagnosis and classification of apathy. *American Journal of Psychiatry* 1990; **147**: 22-30
- Mega MS, Cummings JL, Fiorello T, Gornbein J. The spectrum of behavioral changes in Alzheimer's disease. *Neurology* 1996; **46**: 130-5.
- Starkstein SE, Fedorof JP, Price TR, Leiguarda R, Robinson RG. Apathy following cerebrovascular lesions. *Stroke* 1993; **24**: 1625-30
[Cross References: ABULIA; AKINETIC MUTISM; DEMENTIA; FRONTAL LOBE SYNDROMES]

Aphasia

Aphasia, or dysphasia, is an acquired loss or impairment of language (as opposed to speech) function. Language may be defined as the complex system of symbols used for communication (including reading and writing), encompassing various linguistic components (*viz.* phonology, semantic/lexical, syntax), all of which are dependent on dominant hemisphere integrity. Non-linguistic components of language (emotion, inflection, cadence), collectively known as prosody, may have contributions from both hemispheres. Language is distinguished from speech (oral communication), disorders of which are termed dysarthria or anarthria. Dysarthria and aphasia may coexist but are usually separable.

Clinical assessment of aphasia requires analysis of the following features, through listening to patient's spontaneous speech as well as asking patient to read and repeat:

- FLUENCY: is output effortful, laboured, with agrammatism and dysprosody (non-fluent); or flowing, with paraphasias and neologisms (fluent)?
- COMPREHENSION: spared or impaired?
- REPETITION: preserved or impaired?
- NAMING: preserved or impaired?
- READING: alexia?
- WRITING: agraphia?

These features allow definition of various types of aphasia (see Table and specific entries). For example, motor aphasias are characterized by non-fluent verbal output, with intact or largely unimpaired comprehension, whereas sensory aphasias demonstrate fluent verbal output, often with paraphasias, sometimes jargon, with impaired comprehension. Conduction aphasia is marked by relatively normal spontaneous

speech (perhaps with some paraphasic errors) but a profound deficit of repetition. In transcortical motor aphasia spontaneous output is impaired but repetition is intact.

Aphasias most commonly follow a cerebrovascular event: the specific type of aphasia may change with time following the event, and discrepancies may be observed between classically defined clinicoanatomical syndromes and the findings of everyday practice. Aphasia may also occur with space-occupying lesions and in neurodegenerative disorders, often with other cognitive impairments (*e.g.* Alzheimer's disease) but sometimes in isolation (primary progressive aphasia).

	Broca	**Wernicke**	**Conduction**	**Transcortical: Motor/Sensory**
Fluency	⇩⇩	N	N	⇩/N
Comprehension	N	⇩⇩	N	N/⇩
Repetition	⇩	⇩	⇩⇩	N/N
Naming	⇩	⇩	⇩	N?/N?
Reading	⇩	⇩	⇩	N?/N?
Writing	⇩	⇩	⇩	N?/N?

- Albert ML. Treatment of aphasia. *Archives of Neurology* 1998; **55**: 1417-9
- Benson DF, Ardila A. *Aphasia: a clinical perspective.* New York: OUP 1996
- Damasio AR. Aphasia. *New England Journal of Medicine* 1992; **326**: 531-9
- Mohr JP. Acquired language disorders. In: Asbury AK, McKhann GM, McDonald WI (eds.). *Diseases of the Nervous System: Clinical Neurobiology.* London: Heinemann 1986:816-27
- Willmes K, Poeck K. To what extent can aphasic syndromes be localized? *Brain* 1993; **116**: 1527-40

[Cross References: AGRAMMATISM; AGRAPHIA; ALEXIA; ANOMIA; APROSODIA, APROSODY; BROCA'S APHASIA; CIRCUMLOCUTION; CONDUCTION APHASIA; CONDUIT D'APPROCHE; DYSARTHRIA; DYSPROSODY; JARGON APHASIA; NEOLOGISM; OPTIC APHASIA; PARAPHASIA; TRANSCORTICAL APHASIA; WERNICKE'S APHASIA]

Aphemia

Aphemia is a motor disorder of speech production with preserved comprehension of spoken and written language. This syndrome has also been called phonetic disintegration, pure anarthria, apraxic dysarthria, cortical dysarthria, verbal apraxia, subcortical motor aphasia, pure motor aphasia, small or mini Broca's aphasia, and kinetic speech production disorder, reflecting the differing views as to the nature of the underlying disorder (aphasia, dysarthria, apraxia). Aphemia probably encompasses at least some cases of the "foreign accent syndrome", in which altered speech production and/or prosody makes speech output sound foreign. Such conditions may stand between pure disorders of speech (*i.e.* dysarthrias) and of language (*i.e.* aphasias). They usually reflect damage in the left frontal operculum, but sparing Broca's area.

- Lecours AR, Lhermitte F. The "pure" form of the phonetic disintegration syndrome (pure anarthria): anatomo-clinical report of a single case. *Brain and Language* 1976; **3**:88-113.
- Schiff HB, Alexander MP, Naeser MA, Galaburda AM. Aphemia: clinical-anatomic correlations. *Archives of Neurology* 1983; **40**: 720-7
[Cross References: ANARTHRIA; APHASIA; DYSARTHRIA; DYSPROSODY; SPEECH APRAXIA]

Aphonia

Aphonia is loss of the sound of the voice, necessitating mouthing or whispering of words. As for dysphonia, this most frequently follows laryngeal inflammation, although it may follow bilateral recurrent laryngeal nerve palsy. Dystonia of the abductor muscles of the larynx can result in aphonic segments of speech (spasmodic aphonia, or abductor laryngeal dystonia); this may be diagnosed by asking the patient to keep talking and then hearing the voice fade away to nothing; patients may comment that they cannot hold any prolonged conversation. Aphonia of functional or hysterical origin is well recognised.

Aphonia should be differentiated from mutism, in which patients make no effort to speak, and anarthria in which there is a failure of articulation.
[Cross References: ANARTHRIA; DYSPHONIA; MUTISM]

Apraxia

Apraxia or dyspraxia is a disorder of movement characterized by the inability to perform a voluntary motor act despite an intact motor system (*i.e.* no ataxia, weakness) and without impairment in level of consciousness. Automatic/reflex actions are preserved, hence there is a voluntary-automatic dissociation which some authorities see as critical to the definition of apraxia. Different types of apraxia have been delineated, the standard classification being that of Liepmann (1900):

Ideational apraxia, conceptual apraxia - a deficit in the conception of a movement; this frequently interferes with daily motor activities and is not facilitated by the use of objects; there is often an associated aphasia;

Ideomotor apraxia (IMA) - a disturbance in the selection of elements that constitute a movement (*e.g.* pantomiming the use of tools); in contrast to ideational apraxia, this is a "clinical" disorder inasmuch as it does not greatly interfere with everyday activities; moreover, use of objects may facilitate movement; it may often be manifest as the phenomenon of using body parts as objects, *e.g.* in demonstrating how to use a toothbrush, how to hammer a nail, *etc*., a body part is used to represent the object (finger used as toothbrush, fist as hammer);

Limb-kinetic, or melokinetic apraxia – slowness, clumsiness, awkwardness in using a limb, with a temporal decomposition of movement.

Apraxia may also be defined anatomically:

Parietal (posterior): ideational and ideomotor apraxia are seen with unilateral lesions of the inferior parietal lobule (most usually of the left hemisphere), or premotor area of the frontal lobe (Brodmann areas 6 and 8);

Frontal (anterior): unilateral lesions of the supplementary motor area are associated with impairment in tasks requiring bimanual co-ordination, leading to difficulties with alternating hand movements, drawing alternating patterns (*e.g.* m n m n m n in joined up writing: alternating sequences test, Luria figures). This may be associated with the presence of a grasp reflex and alien limb phenomena (limb-kinetic type of apraxia).

Difficulties with the clinical definition of apraxia persist, as for the agnosias. For example, "dressing apraxia" and "constructional apraxia" are now considered visuospatial problems rather than true apraxias. Likewise, some cases labelled as eyelid apraxia or gait apraxia are not true ideational apraxias. The exact nosological status of speech apraxia also remains tendentious.

- Freund H-J. The apraxias. In: C Kennard (ed.). *Recent Advances in Clinical Neurology 8.* Edinburgh, Churchill Livingstone 1995:29-49
- Pramstaller PP, Marsden CD. The basal ganglia and apraxia. *Brain* 1996;**119**:319-40
[Cross References: ALIEN LIMB; EYELID APRAXIA; FORCED GROPING; FRONTAL LOBE SYNDROMES; GAIT APRAXIA; GRASP REFLEX; SPEECH APRAXIA]

Aprosexia

Aprosexia is a syndrome of psychomotor inefficiency characterized by complaints of forgetting conversations as soon as they are finished, material just read, instructions just given, and difficulty keeping the mind on a specific task. These difficulties, into which the patient has insight, and often bitterly complains of, are commonly encountered in the memory clinic. They represent a disturbance of attention or concentration rather than being a harbinger of dementia, as demonstrated by normality on formal psychometric tests. Concurrent sleep disturbance, irritability, and low mood are common and may reflect an underlying affective disorder (anxiety, depression) which merits specific treatment.
[Cross References: ATTENTION; DEMENTIA]

Aprosodia, Aprosody

Aprosodia or aprosody is an absence of speech melody, intonation, cadence, rhythm, and accentuations (*i.e.* the non-linguistic aspects of language) which convey or imply emotion and attitude, or of the ability to comprehend such cues. Aprosodia may be classified, in a manner analogous to the aphasias, as:

Sensory (posterior) - impaired comprehension of the emotional overtones of spoken language or emotional gesturing; this may be associated with visual extinction and anosognosia, reflecting a right posterior temporoparietal region pathology;
Expressive/Motor (anterior) - an inability to produce emotional overtones.

- Ross ED. The aprosodias. Functional-anatomic organization of the affective components of language in the right hemisphere. *Archives of Neurology* 1981; **38**: 561-9
[Cross References: ANOSOGNOSIA; APHASIA; DYSPROSODY; VISUAL EXTINCTION]

Areflexia

Areflexia is an absence or a loss of tendon reflexes. This may be physiological, in that some individuals never demonstrate tendon reflexes; or pathological, reflecting an anatomical interruption or physiological dysfunction at any point along the monosynaptic reflex pathway which is the neuroanatomical substrate of phasic stretch reflexes. Sudden tendon stretch, as produced by a sharp blow from a tendon hammer, activates muscle spindle Ia afferents which pass to the ventral horn of the spinal cord, there activating α-motor neurones, the efferent limb of the reflex, so completing the monosynaptic arc. Hence, although reflexes are typically regarded as part of the examination of the motor system, reflex loss may also occur in "sensory" disorders, affecting the Ia afferents from the muscle spindle. It is often possible to "hear" that reflexes are absent from the thud of tendon hammer on tendon.

Areflexia is most often encountered in disorders of peripheral nerves (hence a feature of the lower motor neurone syndrome), in axonal neuropathies due to anatomical disconnection of the reflex arc, and in demyelinating neuropathies (Guillain-Barré syndrome, chronic inflammatory demyelinating polyneuropathy) due to functional disconnection. Areflexia may also occur in neuromuscular junction disorders, such as the Lambert-Eaton myasthenic syndrome, in which the reflexes may be "restored" following muscular contraction (facilitation). Transient areflexia may be seen in central nervous system disorders such as cataplexy, and in acute spinal cord syndromes ("spinal shock", *e.g.* acute compression, acute inflammatory myelopathy). [Cross References: CATAPLEXY; FACILITATION; HYPOREFLEXIA; LOWER MOTOR NEURONE SYNDROME; REFLEXES]

Argyll Robertson Pupil (ARP)

The Argyll Robertson pupil is small (miosis) and irregular. It fails to react to light (reflex iridoplegia), but does constrict to accomodation (when the eyes converge), *i.e.* there is light-near pupillary dissociation (ARP = **a**ccommodation **r**eaction **p**reserved). Since the light reflex is lost, testing for the accommodation reaction may be performed with the pupil directly illuminated: this can make it easier to see the response to accommodation, which is often difficult to observe when the pupil is small or in individuals with a dark iris. There may be an incomplete response to mydriatic drugs. Although pupil involvement is usually bilateral, it is often asymmetric, causing anisocoria.

The Argyll Robertson pupil was originally described in the context of neurosyphilis, especially tabes dorsalis. If this pathological diagnosis is suspected, a helpful clinical concomitant is the associated loss of deep pain sensation, as assessed, for example, by vigorously squeezing the Achilles tendon (Abadie's sign). There are, however, a number of recognized causes of ARP besides neurosyphilis, including:

- multiple sclerosis
- encephalitis
- diabetes mellitus
- syringobulbia
- sarcoidosis
- Lyme disease
- pinealoma

- herpes zoster
- Charcot-Marie Tooth disease
- Dejerine-Sottas hypertrophic neuropathy

Miosis and pupil irregularity are inconstant findings in some of these situations, in which case the term pseudo-Argyll Robertson pupil may be preferred.

The neuroanatomical substrate of the Argyll Robertson pupil is uncertain. A lesion in the tectum of the (rostral) midbrain proximal to the oculomotor nuclei has been claimed; in multiple sclerosis and sarcoidosis, magnetic resonance imaging has shown a lesion in the periaqueductal grey matter at the level of the Edinger-Westphal nucleus, but these cases lacked miosis and may be classified as pseudo-Argyll Robertson pupil. Some authorities think a partial oculomotor (III) nerve palsy or a lesion of the ciliary ganglion is more likely.

- Argyll Robertson D. Four cases of spinal myosis: with remarks on the action of light on the pupil. *Edinburgh Medical Journal* 1869; **15**: 487-93
- Dacso CC, Bortz DL. Significance of the Argyll Robertson pupil in clinical medicine. *American Journal of Medicine* 1989; **86**: 199-202
[Cross References: ABADIE'S SIGN; ANISOCORIA; LIGHT-NEAR PUPILLARY DISSOCIATION; MIOSIS; PSEUDO-ARGYLL ROBERTSON PUPIL]

Arm Levitation

Spontaneous arm levitation is one behaviour displayed by an alien limb, indicative of parietal lobe pathology. It is most often seen in corticobasal (ganglionic) degeneration, but a few cases with pathologically confirmed Steele-Richardson-Olszewski syndrome (progressive supranuclear palsy) have been reported.
[Cross Reference: ALIEN LIMB]

Asomatognosia

Asomatognosia is a lack of regard for a part, or parts, of the body, most typically failure to acknowledge the existence of a hemiplegic left arm. Asomatognosia may be verbal (denial of limb ownership) or non-verbal (failure to dress or wash limb). All patients with asomatognosia have hemispatial neglect (usually left), hence this would seem to be a precondition for the development of asomatognosia; indeed, for some authorities asomatognosia is synonymous with personal neglect.

The anatomical correlate of asomatognosia is damage to the right supramarginal gyrus and posterior corona radiata (most commonly due to a cerebrovascular event); cases with right thalamic lesions have also been reported. The predilection of asomatognosia for the left side of the body may simply be a reflection of the aphasic problems associated with left sided lesions that might be expected to produce asomatognosia for the right side.

Asomatognosia is related to anosognosia (unawareness or denial of illness) but the two are dissociable on clinical and experimental grounds. Some authorities consider asomatognosia as a form of confabulation.

- Feinberg TE, Haber LD, Leeds NE. Verbal asomatognosia. *Neurology* 1990; **40**: 1391-4
[Cross References: ANOSOGNOSIA; CONFABULATION; NEGLECT]

Astasia-Abasia

Astasia-abasia is a disorder of gait characterized by impaired balance (disequilibrium), wide base, shortened stride, start/turn hesitation and freezing. The term has no standardized definition and hence may mean different things to different observers; it has also been used to describe a psychogenic disorder of inability to stand or walk despite normal leg strength when lying or sitting. Modern clinical classifications of gait disorders subsume astasia-abasia under the categories of subcortical disequilibrium and frontal disequilibrium, *i.e.* gait disorders with prominent disequilibrium or impaired postural control. A transient inability to sit or stand despite normal limb strength may be seen after an acute thalamic lesion (thalamic astasia).

- Nutt JG, Marsden CD, Thompson PD. Human walking and higher-level gait disorders, particularly in the elderly. *Neurology* 1993; **43**: 268-79

Astereognosis

Astereognosis is the failure to recognize an object (*e.g.* key, coin) palpated by the hand with the eyes closed, despite intact primary sensory modalities. Description of qualities such as the size, shape and texture of the object may be possible. Hence, this is a failure of higher order (*i.e.* cortical) processing and is associated with lesions of the posterior parietal lobe (post central gyrus) association cortex. There may be associated impairments of two-point discrimination and graphaesthesia (cortical sensory syndrome).

Some authorities recommend *stereoanaesthesia* or *stereohypaesthesia* as more appropriate terms for this phenomenon, to emphasize that this may be a disorder of perception rather than a true agnosia (for a similar debate in the visual domain, see Dysmorphopsia).

[Cross References: AGNOSIA; DYSMORPHOPSIA; GRAPHAESTHESIA; TWO-POINT DISCRIMINATION]

Asterixis

Asterixis is a sudden, brief, arrhythmic lapse of sustained posture due to involuntary interruption in muscle contraction. It is most easily demonstrated by observing the dorsiflexed hands with arms outstretched (*i.e.* the motion to indicate "stop"), lapses being seen as flicking or flapping movements of the hands. Movement is associated with EMG silence in anti-gravity muscles for 35-200 ms. These features distinguish asterixis from tremor and myoclonus; the phenomenon has previously been described as negative myoclonus or negative tremor.

Asterixis has been described ipsilateral to lesions of the pons or medulla, and contralateral to lesions of the midbrain, thalamus, primary motor cortex and parietal lobe.

Recognised causes of asterixis include:

- hepatic encephalopathy
- hypercapnia
- uraemia
- drug-induced, *e.g.* anticonvulsants, levodopa
- thalamic lesions (haemorrhage, thalamotomy)

[Cross References: ENCEPHALOPATHY; MYOCLONUS; TREMOR]

Asynergia

Asynergia or dyssynergia is a lack or impairment of synergy of sequential muscular contraction in the performance of complex movements, such that they seem to become broken up into their constituent parts, so called decomposition of movement. This may be evident when performing rapid alternating hand movements. Dyssynergy of speech may also occur, a phenomenon sometimes termed scanning speech (*q.v.*) or scanning dysarthria. This is typically seen in cerebellar syndromes, most often those affecting the cerebellar hemispheres, and may coexist with ataxia, dysmetria, and dysdiadochokinesia.

[Cross References: ATAXIA; CEREBELLAR SYNDROMES; DYSARTHRIA; DYSDIADOCHOKINESIA; DYSMETRIA; SCANNING SPEECH]

Ataxia

Ataxia or dystaxia is a lack of co-ordination (rate, range, timing, direction, force) of voluntary motor acts, impairing their smooth performance. This most often refers to a cerebellar problem, but sensory ataxia, optic ataxia, and frontal ataxia are also described, so it is probably best to qualify ataxia rather than to use the word in isolation.

In *cerebellar ataxia,* defective timing of agonist and antagonist muscle contraction (asynergia) produces jerking, staggering, inaccurate movements (decomposition of movement), which may manifest as intention tremor, dysmetria (past pointing), dysdiadochokinesia, ataxic dysarthria (sometimes known as scanning speech, although this also has other connotations), excessive rebound phenomenon, macrographia, head tremor (titubation), gait ataxia, and abnormal eye movements (nystagmus, square-wave jerks, saccadic intrusions). There may be concurrent limb hypotonia. Cerebellar hemisphere lesions cause ipsilateral limb ataxia (hemiataxia) whereas midline cerebellar lesions involving the vermis produce selective truncal and gait ataxia.

Sensory ataxia results from impaired proprioception and may be seen in disease of the dorsal (posterior) columns of the spinal cord (hence "spinal ataxia"), sensory neuropathies, and neuronopathies affecting the dorsal root ganglia. It is markedly exacerbated by removal of visual cues (*e.g.* in Romberg's sign), unlike the situation with cerebellar ataxia, and also leads to pseudoathetosis.

Optic ataxia is misreaching for visually presented targets, with dysmetria, due to a parieto-occipital lesion, as seen in Balint's syndrome.

"*Frontal ataxia*" is similar to cerebellar ataxia, but results from lesions of the contralateral frontal cortex or frontopontine fibres. These fibres run in the cortico-pontocerebellar tract, synapsing in the pons before passing through the middle cerebellar peduncle to the contralateral cerebellar hemisphere.

Triple ataxia, the rare concurrence of cerebellar, sensory and optic types of ataxia, may be associated with an alien limb phenomenon (sensory type).

There are many causes of cerebellar ataxia, including:

Inherited:

- Autosomal recessive: Friedreich's ataxia
- Autosomal dominant: clinically ADCA types I, II, and III, now reclassified genetically as spinocerebellar ataxias types 1-7

- Episodic ataxias: channelopathies involving potassium (type 1) and calcium (type 2) channels
- Mitochondrial disorders
- Huntingdon's disease
- Dentatorubropallidoluysian atrophy (DRPLA)
- Inherited prion diseases, especially Gerstmann-Straussler-Scheinker (GSS) syndrome

Acquired:

- Cerebrovascular events (infarct, haemorrhage): usually cause hemiataxia; postanoxic cerebellar ataxia
- Inflammatory: demyelination: multiple sclerosis, Miller Fisher variant of Guillain-Barré syndrome, central pontine myelinolysis
- Inflammatory: infection: cerebellitis with Epstein-Barr virus; encephalitis with *Mycoplasma;* HIV
- Neoplasia: tumours, paraneoplastic syndromes
- Neurodegeneration: one variant of multisystem atrophy (MSA-C); prion diseases
- (Brownell-Oppenheimer variant of sporadic Creutzfeldt-Jakob disease, kuru)
- Drugs/toxins, *e.g.* alcohol, phenytoin
- Metabolic: vitamin E deficiency, thiamine deficiency (Wernicke's encephalopathy), hypothyroidism (debatable)

- Wood NW, Harding AE. Ataxic disorders. In: Bradley WG, Daroff RB, Fenichel GM, Marsden CD (eds.). *Neurology in Clinical Practice: Principles of Diagnosis and Management.* Boston: Butterworth-Heinemann 2000 (3rd edition): 309-17

[Cross References: ALIEN LIMB; ASYNERGIA; BALINT'S SYNDROME; CEREBELLAR SYNDROMES; DYSARTHRIA; DYSDIADOCHOKINESIA; DYSMETRIA; HEAD TREMOR; HEMIATAXIA; HYPOTONIA; MACROGRAPHIA; NYSTAGMUS; OPTIC ATAXIA; PROPRIOCEPTION; PSEUDOATHETOSIS; REBOUND PHENOMENON; ROMBERGISM, ROMBERG'S SIGN; SACCADIC INTRUSION; SCANNING SPEECH; SQUARE-WAVE JERKS; TREMOR]

Ataxic Hemiparesis

Ataxic hemiparesis is a syndrome of ipsilateral hemiataxia and hemiparesis, the latter affecting the leg more severely (crural paresis); there may be additional dysarthria, nystagmus, and paraesthesia. This syndrome is caused by lacunar infarction in the contralateral basis pons at the junction of the upper third and lower two-thirds; it may also be seen with infarcts in the contralateral thalamocapsular region, posterior limb of the internal capsule (anterior choroidal artery syndrome), red nucleus, and the paracentral region (anterior cerebral artery territory).

- Fisher CM. Ataxic hemiparesis. A pathologic study. *Archives of Neurology* 1978; **35**: 126-8

[Cross References: ATAXIA; HEMIATAXIA; HEMIPARESIS; PSEUDOCHOREOATHETOSIS]

Ataxic Nystagmus

- see INTERNUCLEAR OPHTHALMOPLEGIA; NYSTAGMUS

Athetosis

Athetosis is an involuntary movement disorder characterized by slow, sinuous, purposeless, writhing movements, often more evident in the distal limbs. Athetosis often co-exists with the more flowing, dance-like movements of chorea, in which case the movement disorder may be described as choreoathetosis. Indeed the term athetosis is now little used except in the context of athetoid cerebral palsy. Athetoid-like movements of the outstretched hands may also been seen in the presence of sensory ataxia (impaired proprioception) and are known as pseudoathetosis or pseudochoreoathetosis.

Choreoathetoid movements result from disorders of the basal ganglia.

[Cross References: CHOREA, CHOREOATHETOSIS; PSEUDOATHETOSIS; PSEUDOCHOREOATHETOSIS]

Atrophy

Atrophy is a wasting or thinning of tissues. The term is often applied to wasted muscles, usually in the context of lower motor neurone pathology (in which case it is synonymous with amyotrophy), but also with disuse. It may also be applied to other tissues, such as subcutaneous tissue (as in hemifacial atrophy).

[Cross References: AMYOTROPHY; HEMIFACIAL ATROPHY; LOWER MOTOR NEURONE SYNDROME; WASTING]

Attention

Attention is a distributed cognitive function, important for the operation of many other cognitive domains; the terms concentration, vigilance, and persistence may be used synonymously with attention. Distinction may be made between different types of attention, *viz.*:

- *Sustained*
- *Selective*
- *Divided/executive function.*

Impairment of attentional mechanisms may lead to distractability (with a resulting complaint of poor memory, better termed aprosexia, *q.v.*). disorientation in time and place, perceptual problems, and behavioural problems (*e.g.* disinhibition), as in the cardinal disorder of attention, delirium (*q.v.*).

The anatomical substrates of attention encompass the ascending reticular activating system of the brainstem, the thalamus, and the prefrontal (multimodal association) cerebral cortex (especially on the right). Damage to any of these areas may cause impaired attention.

Attention may be tested in a variety of ways, all of which are essentially looking for a defect in working memory ("short term memory"):

- Orientation in time/place;
- Digit span forwards/backwards;
- Reciting months of the year backwards, counting back from 30 to 1;
- Serial sevens (serial subtraction of 7 from 100, = 93, 86, 79, 72, 65).

In the presence of severe attentional disorder (as in delirium) it is difficult to make any meaningful assessment of other cognitive domains.

Besides delirium, attentional impairments may be seen following head injury, and in ostensibly "alert" patients, *e.g.* with Alzheimer's disease (the dysexecutive syndrome of impaired divided attention).

- Morris RG. Attentional and executive dysfunction. In: Morris RG (ed.). *The cognitive neuropsychology of Alzheimer-type dementia.* Oxford: OUP 1996: 49-70
- Perry RJ, Hodges JR. Attention and executive deficits in Alzheimer's disease: a critical review. *Brain* 1999; 122: 383-404
[Cross References: APROSEXIA; DELIRIUM; DEMENTIA; DISINHIBITION; FRONTAL LOBE SYNDROMES; PSEUDODEMENTIA]

Aura

An aura is a brief feeling or sensation, lasting seconds to minutes, occurring immediately before the onset of a paroxysmal neurological event such as an epileptic seizure or a migraine attack (migraine with aura, "classical migraine"), "warning" of its imminent presentation, although auras may also occur in isolation.

An aura indicates the focal onset of neurological dysfunction, and may be entirely sensory, such as the fortification spectra (teichopsia) of migraine, or more complex, such as the *déjà vu or jamais vu* auras of focal-onset epilepsy, indicative of temporal lobe and limbic onset respectively. An olfactory aura or parosmia may occur in seizures of medial temporal lobe origin (uncus); simple auditory phenomena may indicate an origin in the superior temporal gyrus. Perhaps the commonest aura in temporal lobe epilepsy is the rising epigastic sensation. Visual auras may occur with seizures of occipital or temporal origin; complex hallucinations and a "tunnel vision" phenomenon are exclusive to seizures of anteromedial temporal and occipitotemporal origin, whereas elementary hallucinations, illusions, and visual loss are common to both occipital and temporal lobe seizures.

- Bien CG, Benninger FO, Urbach H, Schramm J, Kurthen M, Elger CE. Localizing value of epileptic visual auras. *Brain* 2000; **123**: 244-53
- Palmini A, Gloor P. The localising value of auras in partial seizures. *Neurology* 1992; **42**: 801-8
[Cross References: *DÉJÀ VU;* FORTIFICATION SPECTRA; HALLUCINATION; ILLUSION; *JAMAIS VU;* PAROSMIA; "TUNNEL VISION"]

Automatism

Automatisms are automatic, involuntary, behaviours (*i.e.* the patient acts like an automaton) occurring during the state of impaired consciousness during or shortly after an epileptic seizure. There is usually amnesia for the event. Automatisms occur in about one-third of patients with complex partial seizures, most commonly those of temporal or frontal lobe origin. Although there are qualitative differences between the automatisms seen in seizures arising from these sites, they are not of sufficient specificity to be of reliable diagnostic value; bizarre automatisms are more likely to be frontal.

Automatisms may take various forms:

Oro-facial movements: for example lip smacking, chewing and swallowing movements, salivation (especially temporal lobe origin);

Gestural: hand fumbling, foot shuffling, tidying, or more complex actions such as undressing; upper limb movements are said to be more suggestive of temporal lobe origin, lower limbs movements (kicking, cycling) of frontal lobe origin;

Ambulatory: walking or running around (cursive seizures);

Emotional: laughing and, more rarely, crying (gelastic and dacrystic seizures, respectively, although crying may also be a feature of non-epileptic seizures), fear, anger;

Verbal: humming, whistling, grunting, speaking incoherently; vocalization is common in frontal lobe automatisms.

- Delgado-Escueto AV, Bascal FE, Treiman DM. Complex partial seizures on closed circuit television and EEGs: a study of 691 attacks in 79 patients. *Annals of Neurology* 1982; **11**: 292-300

[Cross References: ABSENCE; AURA; SEIZURE]

Autotopagnosia

Autotopagnosia, or somatotopagnosia, is a rare disorder of body schema characterized by inability to identify parts of the body, either to verbal command or by imitation; this is sometimes localized but at worst involves all parts of the body.

This may be a form of category-specific anomia with maximum difficulty for naming body parts. Finger agnosia and right-left disorientation are partial forms of autotopagnosia, all of which are most often seen following cerebrovascular events involving the left parietal area.

[Cross References: AGNOSIA; FINGER AGNOSIA; GERSTMANN SYNDROME; RIGHT-LEFT DISORIENTATION]

B

Babinski's Sign
Babinski's sign is a polysynaptic reflex consisting of an extensor movement (dorsiflexion) of the big toe on eliciting the plantar response, due to contraction of extensor hallucis longus, with or without fanning (abduction) of the other toes (fan sign; *signe de I'éventail*); toe abduction is neither necessary nor sufficient for Babinski's sign to be present. There may be simultaneous contraction of other limb flexor muscles, consistent with the notion that Babinski's sign forms part of a flexion synergy (withdrawal) of the leg. The use of the term "negative Babinski sign" to indicate the normal finding of a downgoing (flexor; plantar flexion) big toe is incorrect, "flexor plantar response" being the appropriate appellation.

The plantar response is most commonly performed by stroking the sole of the foot, although many other variants are described (*e.g.* Chaddock's sign, Oppenheim's sign, *q.v.*).

Babinski's sign is normal in infants with immature (unmyelinated) corticospinal tracts; persistence beyond three years of age, or re-emergence in adult life, is pathological. In this context, Babinski's sign is considered a reliable ("hard") sign of corticospinal (pyramidal) tract dysfunction (upper motor neurone pathology), and may coexist with other signs of upper motor neurone dysfunction (*e.g.* weakness in a so-called pyramidal distribution, spasticity, hyperreflexia). However, if weakness of extensor hallucis longus is one of the features of upper motor neurone dysfunction, or from any other cause, Babinski's sign may be unexpectedly absent although expected. In the presence of extrapyramidal signs, it is important to distinguish Babinski's sign from a striatal toe (spontaneous upgoing plantar).

- Van Gijn J. *The Babinski sign: a centenary.* Utrecht: Universiteit Utrecht, 1996

[Cross References: CHADDOCK'S SIGN; HYPERREFLEXIA; OPPENHEIM'S SIGN; PARKINSONISM; PLANTAR RESPONSE; SPASTICITY; STRIATAL TOE; UPPER MOTOR NEURONE SYNDROME; WEAKNESS]

Babinski's Trunk-Thigh Test
Babinski's trunk-thigh test is suggested to be of use in distinguishing organic from functional paraplegia and hemiplegia (the latter in conjunction with Hoover's sign). The recumbent patient is asked to sit up with the arms folded on the front of the chest. In organic hemiplegia there is involuntary flexion of the paretic leg; in paraplegia both legs are involuntarily raised. In functional paraplegic weakness neither leg is raised, and in functional hemiplegia only the normal leg is raised.
[Cross References: HEMIPLEGIA; HOOVER'S SIGN; PARAPLEGIA]

Balint's Syndrome
Balint's syndrome (first described in 1909) consists of the triad of:
1) "*psychic paralysis of gaze*", an inability to direct voluntary eye movements to visual targets, probably a form of oculomotor apraxia;

2) *optic ataxia*;
3) *simultanagnosia* (dorsal type).

Loss of spontaneous blinking has also been reported.

The commonest cause of Balint's syndrome is bilateral occipital or parieto-occipital infarctions, but it has also been described as a migrainous phenomenon and in Marchiafava-Bignami disease with pathology affecting the corpus callosum.

It is thought to reflect functional disconnection between higher order visual cortical regions and the frontal eye fields, with sparing of the primary visual cortex.

- Damasio A. Disorders of complex visual processing. Agnosia, achromatopsia, Balint's syndrome and related difficulties of orientation and attention. In: Mesulam M-M (ed.). *Principles of behavioral neurology.* Philadelphia: Davis 1985: 259-88
[Cross References: BLINKING; OCULAR APRAXIA; OPTIC ATAXIA; SIMULTANAGNOSIA]

Ballism, Ballismus

Ballism or ballismus is a hyperkinetic involuntary movement disorder characterized by wild, flinging, throwing movements of a limb; it most usually involves one half of the body (*hemiballismus*), although it can sometimes involve a single extremity (*monoballismus*) or both halves of the body (*paraballismus*). These movements are often continuous during wakefulness but cease during sleep. Hemiballismus may be associated with limb hypotonia. Clinical and pathophysiological studies suggest that ballism is a severe form of chorea. It is most commonly associated with lesions of the contralateral subthalamic nucleus.
[Cross References: CHOREA; HEMIBALLISMUS; HYPOTONIA].

Battle's Sign

Battle's sign is a haematoma overlying the mastoid process, which indicates an underlying basilar skull fracture which extends into the mastoid portion of the temporal bone. It appears 48-72 hours after the trauma which causes the fracture.

Beevor's Sign

Beevor's sign is an upward movement of the umbilicus in a supine patient attempting to flex the head onto the chest against resistance (the examiner's hand). It indicates a lesion involving lower abdominal muscles but sparing the upper ones. It may be seen with thoracic cord tumours at or below T10 (cutaneous abdominal reflexes are absent below the lesion) and has also been reported in syringomyelia and primary muscle disease affecting abdominal muscles (*e.g.* facioscapulohumeral muscular dystrophy).

- Tashiro K. Charles Edward Beevor (1854-1908) and Beevor's sign. In: Rose FC (ed.). *A short history of neurology: the British contribution 1660-1910.* Oxford: Butterworth Heinemann 1999: 222-5
[Cross References: ABDOMINAL REFLEXES]

Belle Indifférence

La belle indifférence refers to the seeming lack of concern in the presence of serious symptoms in an hysterical patient, who may show exaggerated emotional reactions in other ways.

Patients with organic lesions may also demonstrate a lack of concern for their disabilities, either due to a disorder of body schema (anosodiaphoria) or due to incongruence of mood (typically in frontal lobe syndromes, sometimes seen in multiple sclerosis).
[Cross References: ANOSODIAPHORIA; FRONTAL LOBE SYNDROMES]

Bell's Palsy
Bell's palsy is an idiopathic peripheral (lower motor neurone) facial weakness (prosopoplegia). It is thought to result from viral inflammation of the facial (VII) nerve. Other causes of lower motor neurone facial paresis (*q.v.*) may need to be excluded before a diagnosis of Bell's palsy can be made.

In the majority of patients with Bell's palsy (idiopathic facial paresis), spontaneous recovery occurs over three weeks to two months. Poorer prognosis is associated with older age (over 40) and if no recovery is seen within four weeks of onset. The place of steroid treatment remains uncertain: one meta-analysis suggests a small benefit for early treatment.

- Williamson IG, Whelan TR. The clinical problem of Bell's palsy: is treatment with steroids effective? *British Journal of General Practice* 1996; **46**: 743-7

[Cross References: BELL'S PHENOMENON; FACIAL PARESIS; LOWER MOTOR NEURONE SYNDROME]

Bell's Phenomenon
Bell's phenomenon is reflex upward, and slightly outward, deviation of the eyes in response to forced closure, or attempted closure, of the eyelids. This is a synkinesis of central origin involving superior rectus and inferior oblique muscles. It may be very evident in a patient with Bell's palsy (idiopathic facial nerve paralysis) attempting to close the paretic eyelid. The reflex indicates intact nuclear and infranuclear mechanisms of upward gaze, and hence that any defect of upgaze is supranuclear. However, in making this interpretation it should be remembered that perhaps 10-15% of the normal population do not show a Bell's phenomenon.
[Cross References: BELL'S PALSY; GAZE PALSY; SUPRANUCLEAR GAZE PALSY; SYNKINESIS]

Benediction Hand
Median nerve lesions in the axilla or upper arm lead to impaired flexion of the index (complete) and middle (partial) fingers on attempting to make a fist, due to weakness of flexor digitorum profundus. The resulting posture is likened to that of a priest saying benediction.

A somewhat similar, but not identical, appearance may occur with ulnar nerve lesions: hyperextension of the metacarpophalangeal joints of the ring and little fingers with slight flexion at the interphalangeal joints. The index and middle fingers are less affected because of the intact innervation of their lumbrical muscles (median nerve).
[Cross References: CLAW HAND]

Bent Spine Syndrome
- see CAMPTOCORMIA

Bielschowsky's Sign, Bielschowsky's Test

Bielschowsky's sign is head tilt towards the shoulder, typically to the side contralateral to a trochlear (IV) nerve lesion. The intorsion of the unaffected eye brought about by the head tilt compensates for the double vision caused by the unopposed extorsion of the affected eye.

Bielschowsky's (head tilt) test consists of the examiner tipping the patient's head from shoulder to shoulder to see if this improves or exacerbates double vision, as will be the case when the head is respectively tilted away from or towards the affected side in a unilateral trochlear (IV) nerve lesion. The test is usually negative in a skew deviation causing vertical divergence of the eyes.

[Cross References: DIPLOPIA; SKEW DEVIATION]

Blepharoptosis

- see PTOSIS

Blepharospasm

Blepharospasm is a focal dystonia of the orbicularis oculi resulting in repeated involuntary forced eyelid closure, with failure of voluntary opening. It may be sufficiently severe to result in functional blindness. The condition typically begins in the sixth decade of life, and is commoner in women than men. Blepharospasm may occur in isolation or in combination with other involuntary movements which may be dystonic (orobuccolingual dystonia or Meige syndrome; limb dystonia) or dyspraxic (eyelid apraxia).

Blepharospasm is usually idiopathic but may be associated with lesions (usually infarction) of the rostral brainstem, diencephalon, and striatum; it has been occasionally reported with thalamic lesions. The pathophysiological mechanism(s) underlying blepharospasm are not understood, but it may reflect dopaminergic pathway disruption causing disinhibition of brainstem reflexes.

Local injections of botulinum toxin into orbicularis oculi are the treatment of choice, the majority of patients deriving benefit and requesting further injection. Failure to respond to botulinum toxin may be due to concurrent eyelid apraxia or dopaminergic therapy with levodopa.

- Grandas F, Elston J, Quinn N, Marsden CD. Blepharospasm: a review of 264 patients. *Journal of Neurology, Neurosurgery and Psychiatry* 1988; **51**: 767-72

[Cross References: BLINKING; DYSTONIA; EYELID APRAXIA; GAPING; YAWNING]

Blind Spot

The blind spot is defined anatomically as the point on the retina at which axons from the retinal ganglion cells enter the optic nerve; since this area is devoid of photoreceptors there is a physiological blind spot. This area may be mapped clinically by confrontation with the examiner's blind spot, or mechanically. Enlargement of the blind spot (peripapillary scotoma) is observed with raised intracranial pressure causing papilloedema: this may be helpful in differentiating papilloedema from other causes of disc swelling such as optic neuritis, in which a central scotoma is the commonest field defect. Enlargement of the blind spot may also be a feature of

peripapillary retinal disorders including big blind spot syndrome.
[Cross References: DISC SWELLING; PAPILLOEDEMA; SCOTOMA]

Blinking
The involuntary blink rate is decreased in Steele-Richardson-Olszewski syndrome; this may also be the case in other causes of parkinsonism, but possibly not in idiopathic Parkinson's disease. It is thought to reflect impaired central dopaminergic activity. Loss of spontaneous blinking has also been reported in Balint's syndrome.
In patients with impaired consciousness, the presence of involuntary blinking implies an intact pontine reticular formation; absence suggests structural or metabolic dysfunction of the reticular formation.
[Cross References: BALINT'S SYNDROME; CORNEAL REFLEX; PARKINSONISM; SIGHING; YAWNING]

Blink Reflex
The blink reflex consists of bilateral reflex contraction of the orbicularis oculi muscles. This may be induced by:

~ Mechanical stimulus, *e.g.* percussion over the supraorbital ridge; this quickly habituates with repetitive stimulation in normal individuals (*cf.* glabellar tap reflex); in unconscious patients with closed eyes, stroking the eyelashes may induce contraction of orbicularis oculi ("eyelash reflex");
~ Sudden visual stimulus approaching the eyes (menace reflex, threat reflex, visuopalpebral reflex): this should be done unexpectedly since the reflex can be voluntarily suppressed. Failure to respond to a stimulus moving into the temporal field of vision may indicate a hemianopic defect in patients unable to comply with standard confrontation visual field testing. Care should be taken to avoid generating air currents with the hand movement as this may stimulate the corneal reflex which may simulate the visuopalpebral reflex.
~ Sudden loud sounds (acousticopalpebral reflex).

The final common (efferent) pathway for these responses is the facial nerve nucleus and facial (VII) nerve, the afferent limbs being the trigeminal (V), optic (II), and auditory (VIII) nerves respectively.
Electrophysiological study of the blink reflex may demonstrate peripheral or central lesions of the trigeminal (V) nerve (afferent) or facial (VII) nerve (efferent). Recently it has been reported that in the evaluation of sensory neuronopathy the finding of an abnormal blink relfex favours a non-paraneoplastic aetiology, since the blink reflex is normal in paraneoplastic sensory neuronopathies.

- Auger RG, Windebank AJ, Lucchinetti CF, Chalk CH. Role of the blink reflex in the evaluation of sensory neuronopathy. *Neurology* 1999; **53**: 407-8
[Cross References: CORNEAL REFLEX; GLABELLAR TAP REFLEX]

Body Part As Object
- see APRAXIA

Bouche de Tapir
Patients with facioscapulohumeral (FSH) dystrophy have a peculiar and characteristic facies, with puckering of the lips when attempting to whistle. The pouting quality of the mouth, unlike that seen with other types of bilateral (neurogenic) facial weakness, has been likened to the face of the tapir (*Tapirus* sp.).
[Cross References: FACIAL PARESIS]

Bovine Cough
A bovine cough lacks the explosive character of a normal voluntary cough. It may result from injury to the distal part of the vagus nerve, particularly the recurrent laryngeal branches which innervate all the muscles of the larynx (with the exception of cricothyroid) with resultant vocal cord paresis. Because of its longer intrathoracic course, the left recurrent laryngeal nerve is more often involved. A bovine cough may be heard in patients with tumours of the upper lobes of the lung (Pancoast tumour) due to recurrent laryngeal nerve palsy. Bovine cough may also result from any cause of bulbar weakness, such as motor neurone disease, Guillain-Barré syndrome, and bulbar myopathies.

- Arcasoy SM, Jett JR. Superior pulmonary sulcus tumors and Pancoast's syndrome. *New England Journal of Medicine* 1997; **337**: 1370-6
[Cross References: BULBAR PALSY]

Bradykinesia
Bradykinesia is a slowness in the performance of voluntary movements, one of the typical signs of parkinsonian syndromes, in which situation it is often accompanied by difficulty in the initiation of movement (akinesia, hypokinesia) and reduced amplitude of movement (hypometria) which may increase with rapid repetitive movements (fatigue). It may be overcome by reflexive movements or in moments of intense emotion (*kinesis paradoxica*). Bradykinesia in parkinsonian syndromes reflects dopamine depletion in the basal ganglia. It may be improved by levodopa and by dopaminergic agents, less so by anticholinergic agents.
Slowness of voluntary movement may also be seen with psychomotor retardation, frontal lobe lesions producing abulia, and in the condition of obsessive slowness.
[Cross References: ABULIA; AKINESIA; FATIGUE; HYPOKINESIA; HYPOMETRIA; *KINESIS PARADOXICA;* PARKINSONISM; PSYCHOMOTOR RETARDATION]

Bradylalia
Bradylalia is a slowness of speech, typically seen in the frontal-subcortical types of cognitive impairment, with or without extrapyramidal features, or in depression.
[Cross References: PALILALIA]

Bradyphrenia
Bradyphrenia is a slowness of thought, typically seen in frontal-subcortical types of cognitive impairment, *e.g.* Steele-Richardson-Olszewski syndrome, vascular dementia. Such patients typically answer questions correctly but with long response times.
[Cross References: ABULIA; DEMENTIA]

Broca's Aphasia

Broca's aphasia is the classic "expressive aphasia", in distinction to the "receptive aphasia" of Wernicke; however, there are problems with this simple classification, since Broca's aphasics may show comprehension problems with complex material, particularly in relation to syntax.

Considering each of the features suggested for the clinical classification of aphasias (see Aphasia), Broca's aphasia is characterized by:

- FLUENCY: slow, laboured, effortful speech (non-fluent) with phonemic paraphasias, agrammatism, and dysprosody; the patient knows what s/he wants to say and usually recognises the paraphasic errors (*i.e.* patients can "self-monitor");
- COMPREHENSION: comprehension for simple material is preserved;
- REPETITION: impaired;
- NAMING: impaired (anomia), may be helped by phonemic or contextual cueing (*cf*. Wernicke's aphasia);
- READING: alexia with laboured oral reading, especially of function words and verb inflections. Silent reading may also be impaired (deep dyslexia) as reflected by poor text comprehension;
- WRITING: similarly affected.

The syndrome may emerge during recovery from a global aphasia. Broca's aphasia is sometimes associated with a right hemiparesis, especially affecting the arm and face; there may also be bucco-lingual-facial dyspraxia. Depression may be a concurrent feature.

Classically Broca's aphasia is associated with a vascular lesion of the third frontal gyrus in the inferior frontal lobe (Broca's area), but in practice such a circumscribed lesion is seldom seen. More commonly there is infarction in the perisylvian region affecting the insula and operculum (Brodmann areas 44 and 45), which may include underlying white matter and the basal ganglia (territory of the superior branch of the middle cerebral artery).

The term small Broca's aphasia, or mini-Broca's aphasia, or Broca's area aphasia, is reserved for a more circumscribed clinical and neuroanatomical deficit than Broca's aphasia, wherein the damage is restricted to Broca's area or its subjacent white matter. There is a mild and transient aphasia or anomia which may share some of the characteristics of aphemia/phonetic disintegration (*i.e.* a motor disorder of speech production with preserved comprehension of spoken and written language).

- Mohr JP, Pessin MS, Finkelstein S, Funkenstein HH, Duncan GW, Davis KR. Broca aphasia: pathologic and clinical. *Neurology* 1978; **28**: 311-24

[Cross References: AGRAMMATISM; AGRAPHIA; ALEXIA; APHASIA; APHEMIA; DYSPROSODY; PARAPHASIA; WERNICKE'S APHASIA]

Brown-Séquard Syndrome

The Brown-Séquard syndrome is the consequence of anatomical or, more usually, functional hemisection of the spinal cord, producing the following pattern of clinical findings:

MOTOR:

Ipsilateral spastic weakness, due to corticospinal tract involvement;

Segmental lower motor neurone signs at the level of the lesion, due to root and/or anterior horn cell involvement.
SENSORY:
A dissociated sensory loss, *i.e.*:
Ipsilateral loss of proprioception, due to dorsal column involvement;
Contralateral loss of pain and temperature sensation, due to crossed spinothalamic tract involvement.

Spinal cord lesions producing this syndrome may be either extramedullary (*e.g.* prolapsed cervical intervertebral disc, extrinsic spinal cord tumour) or intramedullary (*e.g.* multiple sclerosis, intrinsic spinal cord tumour); the former group is said to be the more common.

- Aminoff MJ. *Brown-Séquard.A visionary of science*.New York:Raven 1993: 112-31
- Engelhardt P, Trostdorf E. Zur Differentialdiagnose des Brown-Séquard-Syndroms. *Nervenarzt* 1997; **48**: 45-9
- Tattersall R, Turner B. Brown-Séquard and his syndrome. *Lancet* 2000; **356**: 61-3
[Cross References: DISSOCIATED SENSORY LOSS; MYELOPATHY; PROPRIOCEPTION; SPASTICITY; WEAKNESS]

Brudzinski's (Neck) Sign
Brudzinski described a number of signs, but the one most often used in clinical practice is the neck sign, which is sometimes evident in cases of meningeal irritation, for example due to meningitis. Passive flexion of the neck to bring the head onto the chest is accompanied by flexion of the thighs and legs. As with nuchal rigidity and Kernig's sign, Brudzinski's sign may be absent in elderly or immunosuppressed patients with meningeal irritation.
[Cross References: KERNIG'S SIGN; MENINGISM; NUCHAL RIGIDITY]

Bruxism
Bruxism is forcible grinding or gnashing of the teeth. This is common in children, and as a parasomnia, occurring in 5-20% of the population during non-REM sleep. It may also occur in encephalopathic disorders (*e.g.* hepatic encephalopathy). If necessary, a rubber device or bite may be worn in the mouth to protect the teeth.

- Glaros AG, Rao SM. Bruxism: a critical review. *Psychological Bulletin* 1977; **84**: 767-81
[Cross References: ENCEPHALOPATHY]

Buccofacial Dyspraxia
- see OROFACIAL DYSPRAXIA

Bulbar Palsy
Bulbar palsy is a lower motor neurone syndrome of weakness of bulbar muscles, which may be differentiated from an upper motor neurone bulbar weakness (pseudobulbar palsy).

Clinical features of bulbar palsy include:

~ Dysarthria of flaccid/nasal type;
~ Dysphonia;
~ Dysphagia, often with nasal regurgitation;
~ Weak ("bovine") cough; risk of aspiration;
+/- Wasted, fasciculating tongue;
+/- absent jaw jerk;
+/- absent gag reflex.

Bulbar palsy is usually neurogenic, due to:

- Brainstem disorders affecting cranial nerve motor nuclei (intrinsic):
  motor neurone disease (which may also cause a pseudobulbar palsy)
  poliomyelitis
  glioma
  syringobulbia
- Cranial nerve lesions outside the brainstem (may be associated sensory signs):
  Infiltration by carcinoma, granuloma
- Neuromuscular junction transmission defect:
  myasthenia gravis.

A myogenic bulbar palsy may be seen in oculopharyngeal muscular dystrophy, inclusion body myositis, or polymyositis.
[Cross References: BOVINE COUGH; DYSARTHRIA; DYSPHAGIA; DYSPHONIA; FASCICULATION; GAG REFLEX; JAW JERK; LOWER MOTOR NEURONE SYNDROME; PSEUDOBULBAR PALSY; UPPER MOTOR NEURONE SYNDROME]

Buphthalmos
Buphthalmos, or ox-eye, consists of a large and bulging eye caused by raised intra-ocular pressure due to congenital or secondary glaucoma. This is one of the ophthalmological features of Sturge-Weber syndrome.

C

Cacosmia
- see PAROSMIA

Calf Hypertrophy
Calf enlargement has many causes; it may reflect true hypertrophy (enlargement of muscle fibres) or, more commonly, pseudohypertrophy, due to infiltration with tissue elements other than muscle.

Hypertrophy may be due to neuromuscular disorders producing:

- chronic partial denervation, *e.g.*:
  radiculopathy;
  peripheral neuropathy;
  spinal muscular atrophy;
  following paralytic poliomyelitis;
- continuous muscle activity *e.g.*:
  myotonia congenita;
  Isaac's syndrome (neuromyotonia);
  generalised myokymia.

Pseudohypertrophy may be due to:

- Dystrophinopathies (Duchenne muscular dystrophy, Becker dystrophy), due to excess connective tissue.
- Infection/inflammation: myositis
- Infiltration: amyloidosis, tumour, cysticercosis

- Wilson H, Kidd D, Howard RS, Williams AJ, Spencer GT. Calf hypertrophy following paralytic poliomyelitis. *Postgraduate Medical Journal* 2000; **76**: 179-81
[Cross References: GOWERS SIGN; MYOKYMIA; MYOTONIA; NEUROMYOTONIA]

Caloric Testing
Caloric tests examine the oculovestibular responses and are used to identify vestibular pathology in the assessment of dizziness/vertigo. Each labyrinth may be separately assessed by irrigating each outer ear, with the head flexed to 30 degrees to allow maximum stimulation of the horizontal semicircular canals, using water 7°C above and below body temperature (*i.e.* 30 and 44°C) for 30-40 seconds. Induced nystagmus is then timed both with and without visual fixation (in the dark, Frenzel glasses). This method is cheap but has poor patient acceptability.

Normally, the eyes show conjugate deviation towards the ear irrigated with cold water, with corrective nystagmus in the opposite direction; with warm water the opposite pattern is seen. Dysconjugate responses suggest brainstem damage or

depression. In coma the deviation may be present but without corrective saccades, even at a time when the oculocephalic responses elicited by the doll's head manoeuvre (vestibulo-ocular reflexes) are lost. As coma deepens even the caloric reflex is lost as brainstem involvement progresses.

A reduced duration of induced nystagmus is seen with canal paresis; enhancement of the nystagmus with removal of visual fixation suggests this is peripheral in origin (labyrinthine, vestibulocochlear nerve), whereas no enhancement suggests a central lesion.

- Rudge P, Bronstein AM. Investigations of disorders of balance. In: Hughes RAC (ed.). *Neurological Investigations.* London: BMJ Publishing 1997: 283-314
[Cross References: COMA; NYSTAGMUS; OCULOCEPHALIC RESPONSE; VERTIGO; VESTIBULO-OCULAR REFLEXES]

Camptocormia

Camptocormia, or "bent spine syndrome", was first described as a psychiatric phenomenon in men facing armed conflict (a "war neurosis"). It has subsequently been realised that reducible lumbar kyphosis may also result from neurological disorders, including muscle disease (paravertebral myopathy, nemaline myopathy), Parkinson's disease, dystonia, and, possibly, as a paraneoplastic phenomenon. It may be related in some instances to dropped head syndrome.

- Djaldetti R, Mosberg-Galili R, Sroka H, Merims D, Melamed E. Camptocormia (bent spine) in patients with Parkinson's disease: characterisation and possible pathogenesis of an unusual phenomenon. *Movement Disorders* 1999; **14**: 443-7
[Cross References: DROPPED HEAD SYNDROME; DYSTONIA]

Camptodactyly

Camptodactyly (literally "bent finger") is a flexion deformity at the proximal interphalangeal joint, especially affecting the little finger, usually unilaterally. It is not accompanied by any sensory or motor signs. It may occur as part of a developmental disorder or be isolated; familial cases are also described.

It is important to differentiate Camptodactyly, a non-neurogenic cause of clawing, from neurological diagnoses such as:

- ulnar neuropathy
- C8/T1 radiculopathy
- cervical rib
- syringomyelia

- Llewelyn JG, Winer J, Evans DM, Finnegan T, Thomas PK. "Camptodactyly": a non-neurogenic cause of clawing of the fingers. *Journal of Neurology, Neurosurgery and Psychiatry* 1999; **66**: 268-9
[Cross References: CLAW HAND]

Carphologia

Carphologia, or floccillation, is an aimless plucking at clothing, as if picking off pieces of thread. This may sometimes be seen in psychiatric illness, delirium,

Alzheimer's disease, or vascular dementia particularly affecting the frontal lobe.
[Cross References: DELIRIUM; DEMENTIA]

Carpopedal Spasm

- see *MAIN D'ACCOUCHEUR*

Cataplexy

Cataplexy is a sudden loss of lower limb tone which may lead to falls (drop attacks) without loss of consciousness, usually lasting less than 1 minute. Attacks may be precipitated by strong emotion (laughter, anger, surprise). During an attack there is electrical silence in anti-gravity muscles, which are consequently hypotonic, and transient areflexia.

Cataplexy may occur as part of the syndrome of narcolepsy (excessive and inappropriate daytime somnolence, hypnagogic hallucinations, sleep paralysis); or as symptomatic cataplexy in certain neurological diseases including brainstem lesions, Von Economo's disease (postencephalitic parkinsonism), Niemann-Pick disease, and Nome's disease.
[Cross References: AREFLEXIA; HYPERSOMNOLENCE; HYPOTONIA]

Catatonia

Catalonia is a clinical syndrome, first described by Kahlbaum (1874), characterized by a state of unresponsiveness with maintained but immobile body posture (sitting, standing; *cf.* stupor), mutism, and refusal to eat or drink, with or without staring, grimacing, limb rigidity, maintained abnormal postures (waxy flexibility or *flexibilitas cerea*), negativism, echophenomena (imitation behaviour), stereotypy, and urinary incontinence or retention. After recovery patients are often able to recall events which occurred during the catatonic state (*cf.* stupor).

"Lethal catatonia", in which accompanying fever and collapse lead to death, was described in the 1930's, and seems to resemble neuroleptic malignant syndrome.

Although initially thought to be exclusively a feature of psychiatric disease (schizophrenia, affective disorder), it is now recognised that organic (structural or metabolic disease) may underlie catatonia. Catatonia of psychiatric origin often responds to lorazepam.

Catatonia may be confused clinically with abulia.

- Muqit MMK, Rakshi JS, Shakir RA, Larner AJ. Catatonia or abulia? A difficult differential diagnosis. *Movement Disorders* 2001; **16:** 360-2.
- Rosebush PI, Hildebrand A, Furlong B, Mazurek M. Catatonic syndrome on a general psychiatric ward: frequency, clinical presentation and response to lorazepam. *Journal of Clinical Psychiatry* 1990; **51:** 357-62
[Cross References: ABULIA; AKINETIC MUTISM; IMITATION BEHAVIOUR; MUTISM; NEGATIVISM; RIGIDITY; STEREOTYPY; STUPOR]

Cauda Equina Syndrome

A cauda equina syndrome results from a pathological process affecting the spinal roots below the level of L2 where the spinal cord terminates (*i.e.* it is a syndrome of multiple radiculopathy). Depending on precisely which roots are affected, this may

produce symmetrical or asymmetrical sensory impairment in the buttocks and backs of the thighs, radicular pain, and lower motor neurone type weakness of the foot and/or toes (even a flail foot). Weakness of hip flexion (L1) does not occur, and this may be useful in differentiating a cauda equina syndrome from a conus lesion which may otherwise produce similar features. Sphincters may also be involved, resulting in incontinence, or, in the case of large central disc herniation at L4/L5 or L5/S1, acute urinary retention.

Causes of a cauda equina syndrome include:

- Central disc protrusion;
- Tumour;
- Inflammatory disease, *e.g.* sarcoidosis;
- Ankylosing spondylitis (rare).

[Cross References: FOOT DROP; INCONTINENCE; RADICULOPATHY; URINARY RETENTION]

Central Scotoma, Centrocaecal Scotoma

- see SCOTOMA

Cerebellar Syndromes

Differing clinical pictures may be seen with pathology in differing parts of the cerebellum. Broadly speaking, a midline cerebellar syndrome (involving vermis) may be distinguished from a hemispheric cerebellar syndrome (involving the hemispheres). Their clinical characteristics are:

Midline cerebellar syndrome: gait ataxia but with little or no limb ataxia, hypotonia, nystagmus (because the vestibulocerebellum is spared), or dysarthria; causes include alcoholic cerebellar degeneration, tumour of the midline (*e.g.* medulloblastoma), paraneoplastic cerebellar degeneration;

Hemispheric cerebellar syndrome: limb ataxia (*e.g.* finger-nose, heel-shin ataxia), dysdiadochokinesia, dysmetria, dysarthria, nystagmus; usual causes are infarcts, haemorrhages, demyelination, and tumours.

Pancerebellar syndrome: affecting all parts of the cerebellum, and showing a combination of the above signs (*e.g.* cerebellar degenerations).

- Holmes G. The Croonian lectures on the clinical symptoms of cerebellar disease and their interpretation. *Lancet* 1922: **i**: 1177-82; 1231-7; **ii**: 59-65; 111-5

[Cross References: ASYNERGIA; ATAXIA; DYSARTHRIA; DYSDIADOCHOKINESIA; DYSMETRIA; HEMIATAXIA; HYPOTONIA; NYSTAGMUS]

Cerebellopontine Angle Syndrome

Lesions of the Cerebellopontine angle produce a constellation of ipsilateral signs:

- Depressed corneal reflex (early);
- Lower motor neurone facial (VII) weakness;
- Sensorineural hearing loss (VIII);
- Hemiataxia.

The commonest causes of this syndrome are acoustic neuroma (schwannoma) or meningioma; occasional causes include dermoids, epidermoids, and chordoma.
[Cross References: CORNEAL REFLEX; FACIAL PARESIS; HEMIATAXIA]

Chaddock's Sign
Chaddock's sign, or the external malleolar sign, is a variant method for eliciting the plantar response, by application of a stimulus in a circular direction around the external malleolus, or the lateral aspect of the foot, moving from heel to little toe. Extension of the hallux (upgoing plantar response, Babinski's sign) is pathological, indicating corticospinal tract (upper motor neurone) pathology. The development of Babinski's sign always predates that of Chaddock's sign.

- Chaddock CG. A preliminary communication concerning a new diagnostic nervous sign. *Interstate Medical Journal* 1911; **18**: 742-6
- Van Gijn J. *The Babinski sign: a centenary.* Utrecht: Universiteit Utrecht, 1996
[Cross References: BABINSKI'S SIGN; OPPENHEIM'S SIGN; PLANTAR RESPONSE; UPPER MOTOR NEURONE SYNDROME]

Charcot Joint
A Charcot joint is the result of a destructive arthropathy seen following repeated injury to an anaesthetic joint in patients with impaired or absent pain sensation. There is progressive destruction of articular surfaces with disintegration and reorganisation of joint structure. Although the destruction is painless, the Charcot joint itself may be painful.
Charcot joints may be seen in:

- tabes dorsalis (hips, knees, ankles)
- syringomyelia (elbow)
- hereditary sensory (and autonomic) neuropathies (HSAN, "congenital insensitivity to pain"; ankles)
- leprosy
- diabetes mellitus.

[Cross References: ANALGESIA]

Charles Bonnet Syndrome
- see HALLUCINATIONS

Chasm
- see YAWNING

Cherry Red Spot at the Macula
- see MACULOPATHY

Cheyne-Stokes Breathing
- see PERIODIC RESPIRATION

Chorea, Choreoathetosis

Chorea is an involuntary movement disorder characterized by jerky, restless, purposeless movements (literally dance-like) which tend to flit from one part of the body to another in a rather unpredictable way, giving rise to a fidgety appearance. There may also be athetoid movements (slow, sinuous, writhing), jointly referred to as Choreoathetosis. Severe proximal choreiform movements of large amplitude are referred to as ballism or ballismus. When, as is often the case, such movements are confined to one side of the body they are referred to as hemichorea-hemiballismus. There may be concurrent abnormal muscle tone, either hypotonia or rigidity.

The pathophysiology of chorea (as for ballismus) is unknown; movements may be associated with lesions of the contralateral subthalamic nucleus, caudate nucleus, putamen, and thalamus.

Causes of chorea and Choreoathetosis include:

- Huntington's disease (HD)
- Levodopa therapy of Parkinson's disease
- Systemic lupus erythematosus (SLE)
- Chorea gravidarum
- Sydenham's chorea (post-infectious, rheumatic chorea, St Vitus dance)
- Neuroacanthocytosis
- Polycythaemia rubra vera (hyperviscosity)
- CNS tumour
- Paroxysmal dyskinesias: paroxysmal kinesigenic Choreoathetosis (PKC), paroxysmal dystonic Choreoathetosis (PDC)
- Hyperosmolality (hyperglycaemia, hypernatraemia)
- Dentatorubropallidoluysian atrophy (DRPLA)
- Multiple sclerosis (rare)
- Hyperthyroidism
- Variant Creutzfeldt-Jakob disease
- "Senile chorea" (diagnosis of exclusion, especially of HD)

Where treatment is necessary, antidopaminergic agents such as dopamine receptor antagonists (*e.g.* neuroleptics, sulpiride, risperidone) and dopamine depleting agents (*e.g.* tetrabenazine, reserpine) may help, although they may cause parkinsonism, akathisia, neuroleptic malignant syndrome, and sedation. Chronic neuroleptic use may also cause chorea, but these movements are repetitive and predictable, unlike "classic" chorea.

[Cross References: ATHETOSIS; BALLISM, BALLISMUS; DYSKINESIA; HYPOTONIA; PSEUDOCHOREOATHETOSIS; RIGIDITY]

Chvostek's Sign

Chvostek's sign is the contraction of facial muscles provoked by lightly tapping over the facial nerve as it crosses the zygomatic arch. Chvostek's sign is observed in hypocalcaemic states, such as hypoparathyroidism and the respiratory alkalosis associated with hyperventilation. There may be concurrent posturing of the hand, known as *main d'accoucheur* for its resemblance to the posture adopted for manual delivery of a baby.

The pathophysiology of this mechanosensitivity of nerve fibres is uncertain, but is probably related to increased discharges in central pathways. Although hypocalcaemia might be expected to impair neuromuscular junction transmission and excitation-contraction coupling (since Ca^{2+} ions are required for these processes) this does not in fact occur.
[Cross References: *MAIN D'ACCOUCHEUR;* SPASM]

Ciliospinal Response
The ciliospinal response consists of rapid bilateral pupillary dilatation and palpebral elevation in response to a painful stimulus in the mantle area, for example pinching the skin of the neck.

- Reeves AG, Posner JB. The ciliospinal response in man. *Neurology* 1969; **19**: 1145-52
[Cross References: PUPILLARY REFLEXES]

Circumlocution
Circumlocution refers to:

- A discourse that wanders from the point, only eventually to return to the original subject matter, as seen in fluent aphasias;
- A response to word-finding difficulties, as in early Alzheimer's disease or non-fluent aphasias: in response to familiar pictures, patients may comment that the name is on the tip-of-the-tongue but they cannot access it, and therefore give alternatives, *e.g.* "gardener's friend" or "beetle" for ladybird.

[Cross References: ANOMIA; APHASIA; DEMENTIA]

Clasp-Knife Phenomenon
The clasp-knife phenomenon is the name sometimes applied to the sudden "give" encountered when passively moving a markedly spastic limb. Since the clasp-knife phenomenon is a feature of spasticity, the term "clasp-knife rigidity" is best eschewed to avoid confusion.
[Cross References: RIGIDITY; SPASTICITY]

Claw Foot
Claw foot, or *pied en griffe,* is an abnormal posture of the foot, occurring when weakness and atrophy of the intrinsic foot muscles allows long flexors and extensors to act unopposed, producing shortening of the foot, heightening of the arch, flexion of the distal phalanges and dorsiflexion of the proximal phalanges (*cf.* pes cavus). This may occur in chronic neuropathies of early onset which involve motor fibres, such as hereditary motor and sensory neuropathies types I and II.
[Cross References: PES CAVUS]

Claw Hand
Claw hand, or *main en griffe,* is an abnormal posture of the hand with hyperextension at the metacarpophalangeal joints (5^{th}, 4^{th}, and, to a lesser extent, 3^{rd} finger) and flexion at the interphalangeal joints. This results from ulnar nerve lesions above the

elbow, or injury to the lower part of the brachial plexus (Dejerine-Klumpke type), producing wasting and weakness of hypothenar muscles, interossei, and ulnar (medial) lumbricals, allowing the long finger extensors and flexors to act unopposed.
[Cross References: BENEDICTION HAND; CAMPTODACTYLY]

Clonus
Clonus is rhythmic, involuntary, repetitive, muscular contraction and relaxation. It may be induced by sudden passive stretching of a muscle or tendon, most usually the Achilles tendon (ankle clonus) or patella (patellar clonus). Ankle clonus is best elicited by holding the relaxed leg underneath the moderately flexed knee, then quickly dorsiflexing the ankle and holding it dorsiflexed. A few beats of clonus is within normal limits but sustained clonus is pathological.

Clonus reflects hyperactivity of muscle stretch reflexes and may result from self re-excitation. It is a feature of upper motor neurone disorders affecting the corticospinal (pyramidal) system. Patients with disease of the corticospinal tracts may describe clonus as a rhythmic jerking of the foot, for example when using the foot pedals of a car.

Clonus may also be observed as part of a generalized (primary or secondary) epileptic seizure, either in isolation (clonic seizure) or much more commonly following a tonic phase (tonic-clonic seizure). The clonic movements usually involve all four limbs and decrease in frequency and increase in amplitude over about 30-60 seconds as the attack progresses. Rather different "clonic" movements may occur in non-epileptic seizures.
[Cross References: MYOCLONUS; SEIZURE; UPPER MOTOR NEURONE SYNDROME]

Cogan's (Lid Twitch) Sign
Cogan's sign is a twitching of the upper eyelid seen a moment after the eyes are moved from downgaze to the primary position. Twitches may also be seen with eye closure after sustained upgaze. These phenomena are said to be characteristic signs of ocular myasthenia.

(Cogan's sign should not be confused with Cogan's syndrome, an autoimmune disorder of episodic vertigo, tinnitus, hearing loss and interstitial keratitis; and the oculomotor apraxia of Cogan, a congenital lack of lateral gaze.)

- Cogan DG. Myasthenia gravis: a review of the disease and a description of lid twitch as a characteristic sign. *Archives of Ophthalmology* 1965; **74**: 217-21
[Cross References: FATIGUE; ICE PACK TEST; OCULAR APRAXIA]

Cogwheeling, Cogwheel Phenomenon, Cogwheel Rigidity
- see RIGIDITY

Collier's Sign
Collier's sign ("posterior fossa stare") is elevation and retraction of the upper eyelids. This may be seen with upper dorsal midbrain supranuclear lesions, *e.g.* "top of the basilar syndrome", Parinaud's syndrome, and may be accompanied by paralysis of vertical gaze (especially upgaze) and light-near pupillary dissociation.

- Collier J. Nuclear ophthalmoplegia with special reference to retraction of the lids and ptosis and to lesions of the posterior commissure. *Brain* 1927; **50**: 488-98
[Cross References: LID RETRACTION; LIGHT-NEAR PUPILLARY DISSOCIATION]

Colour Anomia
- see ACHROMATOPSIA; ANOMIA

Coma
Coma is a state of unresponsiveness, with eyes closed, from which a patient cannot be roused by verbal or mechanical stimuli. It represents a greater degree of impairment of consciousness than stupor or obtundation, all three forming part of a continuum, rather than discrete stages, between being alert and comatose. This lack of precision prompts some authorities to prefer the description of the individual aspects of neurological function in unconscious patients, such as eye movements, limb movements, vocalization, and response to stimuli, since this conveys more information than the use of terms such as coma, stupor or obtundation, or the use of a lumped "score", such as the Glasgow Coma Scale.

These signs should be documented serially to assess any progression of coma, since assessment of the depth of coma may be made by observing changes in eye movements and response to central noxious stimuli: roving eye movements are lost before oculocephalic responses; caloric responses are last to go. The switch from flexor to extensor posturing (decorticate *vs.* decerebrate rigidity) also indicates increasing depth of coma.

There are many causes of coma, including:

- Drugs/toxins;
- Metabolic causes;
- Infections;
- Epilepsy;
- Vascular insults (haemorrhage, infarction).

A number of behavioural states may be mistaken for coma, including abulia, akinetic mutism, catatonia, and the locked-in syndrome.

- Bates D. Medical coma. In: Hughes RAC (ed.). *Neurological emergencies.* London: BMJ Publishing 1997 (2nd edition): 1-28
- Plum F, Posner JB. *The diagnosis of stupor and coma.* Philadelphia: FA Davis 1980 (3rd edition)
- Teasdale G, Jennett B. Assessment of coma and impaired consciousness: a practical scale. *Lancet* 1974; **2**: 81-4
- Young GB, Ropper AH, Bolton CF (eds.). *Coma and impaired consciousness: a clinical perspective.* New York: McGraw-Hill 1998
[Cross References: ABULIA; AKINETIC MUTISM; CALORIC TESTING; CATATONIA; DECEREBRATE RIGIDITY; DECORTICATE RIGIDITY; LOCKED-IN SYNDROME; OBTUNDATION; OCULOCEPHALIC RESPONSE; ROVING EYE MOVEMENTS; STUPOR; VEGETATIVE STATE; VESTIBULO-OCULAR REFLEXES]

Conduction Aphasia

Conduction aphasia is defined as a fluent aphasia with paraphasic errors (especially literal) in which, in its "pure" form, there is a dissociation between relatively preserved auditory and visual comprehension of language and impaired repetition (in which the phenomenon of *conduit d'approche* may occur) and naming. Reading comprehension is good or normal, and is better than reading aloud which is impaired by paraphasic errors.

Conduction aphasia was traditionally explained as due to a disconnection between sensory (Wernicke) and motor (Broca) areas for language, involving the arcuate fasciculus in the supramarginal gyrus. Certainly the brain damage (usually infarction) associated with conduction aphasia most commonly involves the left parietal lobe (lower postcentral and supramarginal gyri) and the insula, but it is variable, and the cortical injury may be responsible for the clinical picture.

Conduction aphasia is most often seen during recovery from Wernicke's aphasia, and clinically there is often evidence of some impairment of comprehension. If isolated, the prognosis for conduction aphasia is good.

[Cross References: ANOMIA; APHASIA; BROCA'S APHASIA; *CONDUIT D'APPROCHE*; PARAPHASIA; TRANSCORTICAL APHASIA; WERNICKE'S APHASIA]

Conduit d'approche

Conduit d'approche, or "homing-in" behaviour, is a verbal output phenomenon applied to patients with conduction aphasia attempting to repeat a target word, in which multiple phonemic approximations of the word are presented, with gradual improvement until the target word is achieved. This phenomenon suggests that an acoustic image of the target word is preserved in this condition. A similar phenomenon may be observed in patients with optic aphasia attempting to name a visual stimulus.

A similar behaviour is seen in so-called speech apraxia, in which patients repeatedly approximate to the desired output before reaching it.

[Cross References: CONDUCTION APHASIA; OPTIC APHASIA; SPEECH APRAXIA]

Confabulation

Confabulation is the falsification of memory occurring in clear consciousness in association with amnesia of organic origin, *i.e.* paramnesias are related as true events. However, most amnesic patients, suffering from medial temporal lobe/hippocampal lesions do not confabulate, and poor memory alone cannot explain confabulation. Concurrent hypothalamic/diencephalic and basal forebrain/frontal cortex lesions may be required to develop this syndrome.

Confabulating patients often give a fairly coherent and entirely plausible account of events or experiences, sometimes in response to the examiner's suggestion. Confabulations may be classified as:

- Momentary; or
- Fantastic: these may be of almost delusional intensity.

Confabulation is a classic feature of the Wernicke-Korsakoff syndrome, but is in fact rarely seen. It may also occur in cortical blindness (Anton's syndrome).

- BerlyneN. Confabulation. *British Journal of Psychiatry* 1972; **120**: 31-9
- Downes JJ, Mayes AR. How bad memories can sometimes lead to fantastic beliefs and strange visions. In: Campbell R, Conway MA (eds.). *Broken memories: case studies in memory impairment.* Oxford: Blackwell 1995:115-23
[Cross References: AMNESIA; ASOMATOGNOSIA; CORTICAL BLINDNESS; DELUSION; PARAMNESIA]

Consensual Light Reflex
- see PUPILLARY REFLEXES

Constructional Apraxia
- see APRAXIA

Contracture
The term contracture may be used to describe:

- a prolonged painful muscle spasm with EMG silence, as observed in myotonia and paramyotonia;
- more commonly, an acquired restriction of joint mobility, which may be due to a variety of factors, including prolonged muscle spasticity with or without muscle fibrosis (*i.e.* there is pathological muscle shortening), and ligamentous restrictions. This often occurs in the context of limb immobilisation or inactivity, for example in a flexed posture. Injections of botulinum toxin to abolish muscle spasticity may be required to assess whether there is concurrent ligamentous restriction, and thus to plan optimum treatment, which may involve surgery.

Contractures of muscular origin may be seen in conditions such as Emery-Dreifuss disease (especially elbow, Achilles tendon, posterior part of neck) and Duchenne muscular dystrophy.
[Cross References: MYOTONIA; PARAMYOTONIA; PARAPLEGIA; SPASM; SPASTICITY]

Coprolalia
Coprolalia is the involuntary utterance of expletives or other obscene language. This is a complex vocal tic most characteristically seen in Gilles de la Tourette syndrome although it actually occurs in less than half of affected individuals. Other disease associations are:

- Lesch-Nyhan syndrome
- postencephalitic parkinsonism
- neuroacanthocytosis
- cingulate cortical seizures

The pathophysiology of coprolalia is unknown but may be related to frontal (cingulate and orbitofrontal) dysfunction, for which there is some evidence in Gilles de la Tourette syndrome.
[Cross References: TIC]

Copropraxia
Copropraxia is a complex motor tic comprising obscene gesturing, sometimes seen in Gilles de la Tourette syndrome.
[Cross References: TIC]

Corneal Reflex
The corneal reflex consists of a bilateral blink response elicited by touching the cornea lightly, for example with a piece of cotton wool. As well as observing whether the patient blinks, the examiner should also ask whether the stimulus was felt: a difference in corneal sensitivity may be the earliest abnormality in this reflex. Synkinetic jaw movement may also be observed (see Corneomandibular Reflex).

The afferent limb of the corneal reflex is via the trigeminal (V) nerve, the efferent limb via the facial (VII) nerve to orbicularis oculi. Trigeminal nerve lesions cause both ipsilateral and contralateral corneal reflex loss. Cerebral hemisphere (but not thalamic) lesions causing hemiparesis and hemisensory loss may also be associated with a decreased corneal reflex. Reflex impairment may be an early sign of a cerebellopontine angle lesion, which may also cause ipsilateral lower motor neurone type facial (VII) weakness and ipsilateral sensorineural hearing impairment (VIII). The corneal reflex has a high threshold in comatose patients, and is usually preserved until late (unless coma is due to drug overdose), in which case its loss is a poor prognostic sign.
[Cross References: BLINK REFLEX; COMA; CEREBELLOPONTINE ANGLE SYNDROME; CORNEOMANDIBULAR REFLEX; FACIAL PARESIS]

Corneomandibular Reflex
The corneomandibular reflex consists of anterolateral jaw movement following corneal stimulation.

It has been observed in about three-quarters of patients with motor neurone disease (MND) who display no other pathological reflexes, a frequency much higher than that seen in patients with stroke causing hemiparesis or pseudobulbar palsy, and it is therefore suggested as a sensitive indicator of upper motor neurone involvement in MND.

- Okuda B, Kodama N, Kawabata K, Tachibana H, Sugita M. Corneomandibular reflex in ALS. *Neurology* 1999; **52**: 1699-701
[Cross References: CORNEAL REFLEX; PSEUDOBULBAR PALSY]

Cortical Blindness
Cortical blindness is loss of vision due to bilateral visual cortical damage (usually hypoxic-ischaemic in origin), or bilateral subcortical lesions affecting the optic radiations. A small central field around the fixation point may be spared (macula sparing). Pupillary reflexes are presereved but optokinetic nystagmus cannot be elicited.

Cortical blindness may result from:

- bilateral (sequential or simultaneous) posterior cerebral artery occlusion;
- "top of the basilar syndrome";

- migraine;
- cerebral anoxia;
- bacterial endocarditis;
- Wegener's granulomatosis;
- following coronary or cerebral angiography (may be transient);
- epilepsy (transient)
- cyclosporin therapy, *e.g.* following organ transplantation.

If acute in onset (*i.e.* vascular), cortical blindness may ultimately evolve to prosopagnosia via visual object agnosia.

Patients with cortical blindness may deny their visual defect (Anton's syndrome, visual anosognosia) and may confabulate about what they "see".

[Cross References: ANOSOGNOSIA; CONFABULATION; MACULA SPARING, MACULA SPLITTING; OPTOKINETIC NYSTAGMUS (OKN), OPTOKINETIC RESPONSE; PROSOPAGNOSIA; PUPILLARY REFLEXES; VISUAL AGNOSIA]

Coup de Sabre

Coup de sabre is a localized form of scleroderma manifest as a linear, atrophic lesion on the forehead which may be mistaken for a scar. This lesion may be associated with hemifacial atrophy and epilepsy, and neuroimaging may show hemiatrophy and intracranial calcification. Whether these changes reflect inflammation or a neurocutaneous syndrome is not known.

- Duyff RF, Vos J. A "scar" and epilepsy: coup de sabre. *Journal of Neurology, Neurosurgery and Psychiatry* 1998; **65**: 568

[Cross References: HEMIFACIAL ATROPHY]

Cover Test, Cover-Uncover Test

The simple cover and cover-uncover tests may be used to demonstrate manifest and latent strabismus (heterotropia and heterophoria) respectively.

The cover test demonstrates tropias: the uncovered eye is forced to adopt fixation; any movement therefore represents a manifest strabismus (heterotropia).

The cover-uncover test demonstrates phorias: any movement of the covered eye to re-establish fixation as it is uncovered represents a latent strabismus (heterophoria).

The alternate cover test, in which the hand or occluder moves back and forth between the eyes, repeatedly breaking and re-establishing fixation, is more dissociating and therefore helpful in demonstrating whether or not there is strabismus. It should be performed in the nine cardinal positions of gaze to determine the direction that elicits maximal deviation. However, it does not distinguish between tropias and phorias, for which the cover and cover-uncover tests are required.

[Cross References: HETEROPHORIA; HETEROTROPIA]

Cramp

- see SPASM; STIFFNESS

Cremasteric Reflex

The Cremasteric reflex is a superficial or cutaneous reflex consisting of contraction of the cremaster muscle causing elevation of the testicle, following stimulation of the skin of the upper inner aspect of the thigh from above downwards (*i.e.* the L1, L2 dermatomes, via the ilioinguinal and genitofemoral nerves).

The Cremasteric reflex is lost when the corticospinal pathways are damaged (as for the superficial abdominal reflexes), or following lesions of the genitofemoral nerve. It may also be absent in elderly men, or in patients with hydrocele, varicocele, orchitis or epididymitis.

[Cross References: ABDOMINAL REFLEXES; REFLEXES]

Crocodile Tears

Crocodile tears reflect inappropriate unilateral lacrimation during eating, such that tears may spill down the face (epiphora). This autonomic synkinesis is a striking but rare consequence of aberrant reinnervation of the facial (VII) nerve, usually after a Bell's palsy, when fibres originally supplying the salivary glands are re-routed to the lacrimal gland via the greater superficial petrosal nerve.

[Cross References: BELL'S PALSY; EPIPHORA; SYNKINESIS]

Crying

- see AUTOMATISM; PATHOLOGICAL CRYING

D

Dalrymple's Sign
Dalrymple's sign is increased width of the palpebral fissure, often seen in hyperthyroidism.
[Cross References: LID RETRACTION]

Dazzle
Dazzle is a painless intolerance of the eyes to bright light (*cf.* photophobia). It may be peripheral in origin (retinal disease; opacities within cornea, lens, vitreous); or central (lesions anywhere from optic nerve to occipitotemporal region).
[Cross References: PHOTOPHOBIA]

Decerebrate Rigidity
Decerebrate rigidity is a posture observed in comatose patients in which there is extension and pronation of the upper extremities, extension of the legs, and plantar flexion of the feet (= extensor posturing), which is taken to be an exaggeration of the normal standing position. Stimulation with painful stimuli may induce opisthotonos, hyperextension and hyperpronation of the upper limbs.

Decerebrate rigidity occurs in severe metabolic disorders of the upper brainstem (anoxia/ischaemia, trauma, structural lesions, drug-induced). A similar picture was first observed by Sherrington (1898) following section of the brainstem of cats at the collicular level, below the red nuclei, such that the vestibular nuclei were intact. The action of the vestibular nuclei, unchecked by higher centres, may be responsible for the profound extensor tone.

Decerebrate rigidity indicates a deeper level of coma than decorticate rigidity; the transition from the latter to the former is associated with a worsening of prognosis.
[Cross References: COMA; DECORTICATE RIGIDITY; OPISTHOTONOS]

Decomposition of Movement
- see ASYNERGIA

Decorticate Rigidity
Decorticate rigidity is a posture observed in comatose patients in which there is adduction of the shoulders and arms, and flexion of the elbows and wrists (= flexor posturing). The lesion responsible for decorticate rigidity is higher than that causing decerebrate rigidity, often being diffuse cerebral hemisphere/diencephalic disease, although, despite the name, it may occur with upper brainstem lesions. Common causes are anoxia/ischaemia, trauma, and drugs.
[Cross References: COMA; DECEREBRATE RIGIDITY]

Déjà Vu

Déjà vu is an inappropriate sensation of familiarity with ones surroundings, or of having lived through this moment before, despite this not being the case. It occurs as a complex aura of focal onset epilepsy. *Déjà vu* is indicative of temporal lobe onset, and is said by some authors to be the only epileptic aura of reliable lateralising significance (right).

[Cross References: AURA; *JAMAIS VU*]

Delirium

Delirium is a neurobehavioural syndrome of which the cardinal feature is a deficit of attention, the ability to focus on specific stimuli; it may also be characterized as an "acute confusional state", "acute organic reaction", or "acute brain syndrome". Although there may be a concurrent alteration in level of awareness, ranging from lethargy to hypervigilance, delirium is not primarily a disorder of arousal or alertness (*cf.* coma, stupor, obtundation). Other features commonly observed in delirium include:

- impaired cognitive function: disorientation in time and place;
- perceptual disorders: illusions, hallucinations;
- behavioural disturbances: agitation, restlessness, aggression, wandering; these may occur as a consequence of perceptual problems;
- language: rambling, incoherent speech; logorrhoea;
- altered sleep-wake cycle: "sundowning" (restlessness and confusion at night);
- tendency to marked fluctuations in alertness/activity, with occasional lucid intervals;
- delusions: often persecutory.

Hence this abnormal mental state shows considerable clinical heterogeneity.

The course of delirium is usually brief (seldom more than a few days, often only hours). On recovery the patient may have no recollection of events, although islands of recall may be preserved, corresponding with lucid intervals (a useful, if retrospective, diagnostic feature).

Delirium is often contrasted with dementia, a "chronic brain syndrome", in which attention is relatively preserved, the onset is insidious rather than acute, the course is stable over the day rather than fluctuating, and which lasts months to years. However, it should be noted that in the elderly delirium is often superimposed on dementia, perhaps reflecting impaired cerebral reserve.

Although the pathophysiology of delirium is not well understood, recognised causes, or risk factors for the development, of delirium include:

- Metabolic disorders (hepatic failure, uraemia, porphyria)
- Drugs/toxins/alcohol, especially withdrawal from (*e.g.* delirium tremens)
- Infection: primary CNS (encephalitis, meningitis), or systemic (urinary tract, chest, septicaemia)
- Head injury
- Cerebrovascular disease
- Inflammatory disorders (*e.g.* collagen vascular disease)
- Epilepsy (*e.g.* some forms of status)

These merit treatment in their own right, and investigations should be tailored to identify these aetiological factors. The EEG may show non-specific slowing in delirium, the degree of which is said to correlate with the degree of impairment, and reverses with resolution of delirium.

It is suggested that optimal nursing of delirious patients should aim at environmental modulation to avoid both under- and over-stimulation; a side room is probably best.

- Taylor D, Lewis S. Delirium. In: Hughes RAC (ed.). *Neurological Emergencies.* London: BMJ Publishing 1997 (2nd edition): 76-101

[Cross References: AGRAPHIA; ATTENTION; COMA; DELUSION; DEMENTIA; HALLUCINATION; ILLUSION; LOGORRHOEA; OBTUNDATION; STUPOR; "SUNDOWNING"]

Delusion

A delusion is a fixed false belief, not amenable to reason (*i.e.* held despite evidence to the contrary), and not culturally sanctioned. There are a number of common forms of delusion, including:

~ persecutory (paranoia);
~ reference: important events or people being influenced by patients thoughts, ideas;
~ grandiose/expansive: particularly in mania;
~ guilt/worthlessness: particularly in depression;
~ hypochondria;
~ thought broadcast and thought insertion;
~ control by an external agency.

Specific, named, delusions are those of:

Capgras: the "delusion of doubles", a familiar person or place thought to be an impostor, or double; resembles reduplicative paramnesia;

Fregoli: a familiar person is identified in other people, even though they bear no resemblance; may be seen in schizophrenia;

De Clérambault (erotomania): the belief (usually of a single woman) that a famous person is secretly in love with her ("hope"), followed by the belief that that person is persecuting her ("resentment"); may be seen in schizophrenia.

Delusions are a feature of primary psychiatric disease (psychoses such as schizophrenia; neuroses such as depression), but may also be encountered in neurological disease with secondary psychiatric features ("organic psychiatry"), *e.g.* delirium, and dementing syndromes such as Alzheimer's disease, Lewy body dementia.

[Cross References: DELIRIUM; DEMENTIA; HALLUCINATION; ILLUSION; REDUPLICATIVE PARAMNESIA]

Dementia

Dementia is a loss of intellectual (cognitive) functions sufficient to interfere with social and occupational functioning. Cognition encompasses multiple functions including memory, language, perception, attentional mechanisms and executive function (planning, reasoning). These elements may be affected selectively or globally: older

definitions of dementia requiring global cognitive decline have now been superseded. Amnesia may or may not, depending on the classification system used, be a *sine qua non* for the diagnosis of dementia. Attentional mechanisms are largely preserved, certainly in comparison with delirium, a condition which precludes meaningful neuropsychological assessment because of profound attentional deficits. Although commoner in the elderly, dementia can also occur in the presenium, and in children who may lose cognitive skills as a result of hereditary metabolic disorders. Failure to develop cognitive skills is termed learning disability. Multiple neuropsychological tests are available to test different areas of cognition.

The heterogeneity of dementia is further exemplified by the fact that it may be acute or insidious in onset, and its course may be progressive, stable, or, in some instances, reversible ("dysmentia"). A distinction is drawn by some authors between cortical and subcortical dementia: in the former the pathology is predominantly cortical and neuropsychological findings are characterized by amnesia, agnosia, apraxia, and aphasia (*e.g.* Alzheimer's disease); in the latter pathology is predominantly frontal-subcortical and neuropsychological deficits include psychomotor retardation, attentional deficits, with relative preservation of memory and language; movement disorders may also be apparent (*e.g.* Huntington's disease, Steele-Richardson-Olszewski syndrome). However, not all authors subscribe to this distinction, and considerable overlap may be observed.

Cognitive deficits also occur in affective disorders such as depression, usually as a consequence of impaired attentional mechanisms; this is often labelled as pseudodementia since it is potentially reversible with treatment of the underlying affective disorder. It may be difficult to differentiate dementia originating from depressive or degenerative disease, since depression may also be a feature of degenerative disease. Impaired attentional mechanisms account for the common complaint of not recalling conversations or instructions immediately after they happen (aprosexia). Behavioural abnormalities are common in dementias due to degenerative brain disease, and may require treatment in their own right.

Causes of dementia include:

- Neurodegenerative disease: Alzheimer's disease, frontotemporal dementia (encompassing Pick's disease), dementia with Lewy bodies, Huntington's disease, Steele-Richardson-Olszewski syndrome, corticobasal degeneration, prion disease, Down's syndrome, dementia pugilistica.
- Vascular disease: focal strategic infarcts (*e.g.* paramedian thalamic infarction), multiple infarcts, Binswanger's disease.
- Inflammatory disorders: multiple sclerosis.
- Infection: HIV dementia, neurosyphilis, Whipple's disease.
- Metabolic causes: Wernicke-Korsakoff syndrome, vitamin B_{12} deficiency, hypothyroidism, hyperparathyroidism/hypercalcaemia, leucodystrophies.
- Structural disease: normal pressure hydrocephalus, tumours.

Cognitive dysfunction may be identified in many other neurological illnesses.

Investigation of patients with dementia aims to identify its particular cause; because of the possibility of progression, reversible causes are regularly sought though rare. Specific treatments for dementia are few: cholinesterase inhibitors have been licensed

for the treatment of mild to moderate Alzheimer's disease and may find a role in other conditions, such as dementia with Lewy bodies.

- Burns A, Levy R (eds.). *Dementia.* London: Chapman & Hall 1995
- Chiu E, Gustafson L, Ames D, Folstein MF (eds.). *Cerebrovascular disease and dementia: pathology, neuropsychiatry and management.* London: Martin Dunitz 2000
- Cummings JL, Benson DF. *Dementia: a clinical approach.* Boston: Butterworth Heinemann 1992 (2nd edition)
- Doran M. Diagnosis of presenile dementia. *British Journal of Hospital Medicine* 1997; **58**: 105-10
- Growdon JH, Rossor MN (eds.). *The dementias.* Boston: Butterworth Heinemann 1998
- Kertesz A, Munoz DG (eds.). *Pick's disease and Pick complex.* New York: Wiley-Liss 1998
- McCarthy RA, Warrington EK. *Cognitive neuropsychology: a clinical introduction.* San Diego: Academic 1990
- Snowden JS, Neary D, Mann DMA. *Fronto-temporal lobar degeneration: fronto-temporal dementia, progressive aphasia, semantic dementia.* New York: Churchill Livingstone 1996
- Spreen O, Strauss E. *A compendium of neuropsychological tests: administration, norms, and commentary.* New York: Oxford University Press 1998 (2nd edition)
[Cross References: AGNOSIA; AMNESIA; APHASIA; APRAXIA; APROSEXIA; ATTENTION; DELIRIUM; DYSMENTIA; PSEUDODEMENTIA; PSYCHOMOTOR RETARDATION]

De Musset's Sign
- see HEAD TREMOR

Diaphoresis
Diaphoresis is sweating. This may be physiological, as in sympathetic activation (*e.g.* during hypotension, hypoglycaemia), or pathological (hyperhidrosis, *q.v.*). Diaphoresis may be seen delirium tremens, or may be induced by certain drugs, *e.g.* cholinesterase inhibitors.
[Cross References: HYPERHIDROSIS]

Digital Reflex
- see HOFFMANN'S SIGN, TRÖMNER'S SIGN

Diplopia
Diplopia is double vision, *viz.,* seeing two images of a single object. The spatial and temporal characteristics of the diplopia may help to ascertain its cause.

Diplopia may be monocular, in which case ocular causes are most likely (although it may be cortical or functional in origin), or binocular, implying a divergence of the visual axes of the two eyes. With binocular diplopia, it is of great importance to ask

the patient whether the images are separated horizontally, vertically, or obliquely (tilted), since this may indicate the extraocular muscle(s) most likely to be affected. Whether the two images are separate or overlapping is important when trying to ascertain the direction of maximum diplopia.

The experience of diplopia may be confined to, or particularly noticeable during, the performance of particular activities, reflecting the effect of gaze direction; for example, diplopia experienced on coming downstairs may reflect a trochlear (IV) nerve palsy; or only on looking to the left may reflect a left abducens (VI) nerve palsy. Double vision experienced on looking at a distant object after looking down (*e.g.* reading) may occur with bilateral abducens (VI) nerve palsies. The effect of gaze direction on diplopia should always be sought, since images are most separated when looking in the direction of a paretic muscle. Conversely, diplopia resulting from the breakdown of a latent tendency for the visual axes to deviate (latent strabismus, quint) results in diplopia in all directions of gaze.

Examination of the eye movements should include asking the patient to look at a target, such as a pen, in the various directions of gaze (versions) to ascertain where diplopia is maximum. Ductions are tested monocularly with the opposite eye covered. Then, each eye may be alternately covered to try to demonstrate which of the two images is the false one, namely that from the non-fixing eye. The false image is also the most peripheral image. Thus in a left abducens (VI) nerve palsy, diplopia is maximum on left lateral gaze; when the normal right eye is covered the inner image disappears; the non-fixing left eye is responsible for the remaining false image, which is the more peripheral and which disappears when the left eye is covered.

Other clues to the cause of diplopia include ptosis (unilateral: oculomotor (III) nerve palsy; bilateral: myasthenia gravis), and head tilt or turn (*e.g.* turn to the right suggests a weak right lateral rectus muscle suggesting a right abducens (VI) nerve palsy; tilt to the left shoulder suggests a right trochlear (IV) nerve palsy, = Bielschowsky's sign).

Manifest squints (heterotropia) are obvious but seldom a cause of diplopia if long-standing. Latent squints may be detected using the cover-uncover test, when the shift in fixation of the eyes indicates an imbalance in the visual axes; this may account for diplopia if the normal compensation breaks down. This produces diplopia in all directions of gaze (comitant). Patients may with an effort be able to fuse the two images.

Transient diplopia (minutes to hours) suggests the possibility of myasthenia gravis. There are many causes of persistent diplopia, including the breakdown of a latent strabismus, development of oculomotor (III), trochlear (IV) or abducent (VI) nerve palsy (singly or in combination), orbital myopathy (thyroid), and mass lesions of the orbit (tumour, pseudotumour).

Divergence of the visual axes or ophthalmoplegia without diplopia suggests a long-standing problem, such as amblyopia or chronic progressive external ophthalmoplegia. Some eye movement disorders are striking for the lack of associated diplopia, *e.g.* internuclear ophthalmoplegia.

[Cross References: ABDUCENT NERVE PALSY; AMBLYOPIA; BIELSCHOWSKY'S SIGN, BIELSCHOWSKY'S TEST; COVER TEST, COVER-UNCOVER TEST; HETEROPHORIA; HETEROTROPIA; INTERNUCLEAR OPHTHALMOPLEGIA; OCULOMOTOR NERVE PALSY]

Disc Swelling

Swelling or oedema of the optic nerve head may be visualized by ophthalmoscopy. It produces haziness of the nerve fibre layer obscuring the underlying vessels; there may also be haemorrhages and loss of spontaneous venous pulsation. Disc swelling due to oedema must be distinguished from pseudopapilloedema, elevation of the optic disc not due to oedema, in which the nerve fibre layer is clearly seen.

Disc swelling may be due to raised intracranial pressure (papilloedema, *q.v.*), or local inflammation of the optic nerve (papillitis), and may be associated with marked impairment of vision, for example in optic neuritis, or be without specific visual complaint (as may be the case in papilloedema). The history, visual acuity and visual fields may help determine the cause of disc swelling.

Recognised causes of disc swelling include:

Unilateral:

- optic neuritis;
- acute ischaemic optic neuropathy (arteritic, non-arteritic)
- orbital compressive lesions, *e.g.* optic nerve sheath meningioma (Foster Kennedy syndrome)
- Graves ophthalmopathy (through compression of retinal veins by myositis)
- Central retinal vein occlusion
- Infiltration: carcinoma, lymphoma, granuloma
- Raised intracranial pressure (papilloedema; more usually bilateral)

Bilateral:

- raised intracranial pressure (papilloedema)
- malignant hypertension
- hypercapnia
- high CSF protein, as in Guillain-Barré syndrome
- any of the unilateral causes

[Cross References: FOSTER KENNEDY SYNDROME; PAPILLOEDEMA; PSEUDOPAPILLOEDEMA; VENOUS PULSATION]

Disinhibition

Disinhibited behaviour is impulsive, showing poor judgement and insight; it may transgress normal cultural or social bounds. The disinhibited patient may be inappropriately jocular (*witzelsucht*), distractible (impaired attentional mechanisms), and show emotional lability. A Disinhibition Scale encompassing various domains (motor, intellectual, instinctive, affective, sensitive) has been described.

Disinhibition is a feature of frontal lobe, particularly orbitofrontal, dysfunction. This may be due to neurodegenerative disorders (frontotemporal dementia, Alzheimer's disease), mass lesions, or be a feature of epileptic seizures.

[Cross References: ATTENTION; EMOTIONAL LABILITY; FRONTAL LOBE SYNDROMES; *WITZELSUCHT*]

Dissociated Sensory Loss

Dissociated sensory loss refers to impairment of selected sensory modalities with preservation, or sparing, of others. It is usually an indication of an intramedullary

spinal cord lesion. For example, a focal central cord pathology such as syringomyelia will, in the early stages, selectively involve decussating fibres of the spinothalamic pathway within the ventral commissure, thus impairing pain and temperature sensation, often in a suspended, "vest-like" or cuirass distribution, whilst the dorsal columns are spared, leaving proprioception intact. The anterior spinal artery syndrome also leaves the dorsal columns intact. Conversely, pathologies confined, largely or exclusively, to the dorsal columns (classically tabes dorsalis and subacute combined degeneration of the cord from vitamin B_{12} deficiency, but probably most commonly seen with compressive cervical myelopathy) impair proprioception, sometimes sufficient to produce pseudoathetosis or sensory ataxia, whilst pain and temperature sensation is preserved. A double dissociation of sensory modalities on opposite sides of the trunk is seen in the Brown-Séquard syndrome.

Small fibre peripheral neuropathies may selective affect the fibres which transmit pain and temperature sensation, leading to a glove-and-stocking impairment to these modalities. Neuropathic (Charcot) joints and skin ulceration may occur in this situation; tendon reflexes may be preserved.

[Cross References: ANALGESIA; ATAXIA; BROWN-SÉQUARD SYNDROME; CHARCOT JOINT; MYELOPATHY; PROPRIOCEPTION; PSEUDOATHETOSIS]

Dix-Hallpike Positioning Test

- see HALLPIKE MANOEUVRE, HALLPIKE TEST

Doll's Head Manoeuvre

- see BELL'S PHENOMENON; CALORIC TESTING; OCULOCEPHALIC RESPONSE; SUPRANUCLEAR GAZE PALSY; VESTIBULO-OCULAR REFLEXES

"Dorsal Guttering"

Dorsal guttering refers to the marked prominence of the extensor tendons on the dorsal surface of the hand when intrinsic hand muscles (especially interossei) are wasted, as may occur in an ulnar nerve lesion, a lower brachial plexus lesion, or a T1 root lesion. Benign extramedullary tumours at the foramen magnum may also produce this picture (a "false localising sign"). In many elderly people the extensor tendons are prominent in the absence of significant muscle wasting.

[Cross References: WASTING]

Dressing Apraxia

- see APRAXIA

Drooling

- see SIALORRHOEA

Dropped Head Syndrome

Dropped head syndrome (head droop or head drop) refers to forward flexion of the head on the neck, such that the chin falls on to the chest (*cf.* antecollis) and the head

cannot be voluntarily extended. This syndrome has a wide variety of causes including:

Neuromuscular disorders causing axial truncal muscle weakness, especially of upper thoracic and paraspinous muscles:

- myasthenia gravis;
- motor neurone disease;
- polymyositis;
- Guillain-Barré syndrome, chronic inflammatory demyelinating polyneuropathy;
- "dropped head syndrome", or "isolated neck extensor myopathy", a condition of uncertain aetiology but which may on occasion be steroid-responsive ("bent spine syndrome" or camptocormia may be a related form of axial myopathy).

Extrapyramidal disorders:

- Parkinson's disease;
- Multiple system atrophy.

- Katz JS, Wolfe GI, Burns DK, Bryan WW, Fleckenstein JL, Barohn RJ. Isolated neck extensor myopathy. A common cause of dropped head syndrome. *Neurology* 1996; **46**: 917-21

- Oerlemans WGH, de Visser M. Dropped head syndrome and bent spine syndrome: two separate entities or different manifestations of axial myopathy? *Journal of Neurology, Neurosurgery and Psychiatry* 1998; **65**: 258-9

- Rose MR, Levin KH, Griggs RC. The dropped head plus syndrome: quantitation of response to corticosteroids. *Muscle Nerve* 1999; **22**: 115-8

[Cross References: ANTECOLLIS; CAMPTOCORMIA; MYOPATHY]

Dynamic Aphasia

Dynamic aphasia refers to an aphasia characterized by difficulty initiating speech output, ascribed to executive dysfunction. There is a reduction in spontaneous speech, but on formal testing no paraphasias, minimal anomia, preserved repetition and automatic speech. "Incorporational echolalia", when the patient uses the examiner's question to help form an answer, may be observed. Dynamic aphasia may be conceptualised as a lesser version of transcortical motor aphasia, and may be seen with lesions of dorsolateral prefrontal cortex ("frontal aphasia").

- Esmonde T, Giles E, Xuereb J, Hodges J. Progressive supranuclear palsy presenting with dynamic aphasia. *Journal of Neurology, Neurosurgery and Psychiatry* 1996; **60**: 403-10

- Robinson G, Blair J, Cipolotti L. Dynamic aphasia: an inability to select between competing verbal responses. *Brain* 1998; 121: 77-89

[Cross References: ECHOLALIA; TRANSCORTICAL APHASIA]

Dysaesthesia

Dysaesthesia is an unpleasant, abnormal or unfamiliar, sensation, often with a burning and/or "electrical" quality. Some authorities reserve the term for provoked positive sensory phenomena, as opposed to spontaneous sensations (paraesthesia). Dysaesthesia differs from paraesthesia in its unpleasant quality, but may overlap in some

respects with allodynia, hyperalgesia and hyperpathia (the latter phenomena are provoked by stimuli, either non-noxious or noxious).

There are many causes of dysaesthesia, both peripheral (including small fibre neuropathies, neuroma, nerve trauma) and central (*e.g.* spinal multiple sclerosis).

Dysaesthetic sensations may be helped by agents such as carbamazepine, amitriptyline, and gabapentin.

[Cross References: ALLODYNIA; HYPERALGESIA; HYPERPATHIA; PARAESTHESIA]

Dysarthria

Dysarthria is a motor speech disorder of neurological origin (*cf.* dysphonia due to primary laryngeal pathology), causing impaired motor control (articulation) of the speech musculature. There is no language disturbance (*cf.* aphasia, although the two often coexist).

There are various syndromes of dysarthria, which have been classified as follows:

Flaccid/Nasal dysarthria: hypernasal, breathy, whining output, as in bulbar palsy, myasthenia gravis;

Spastic dysarthria: slow, strained ("strangled") output, monotonous; as in pseudobulbar palsy; may coexist with Broca's aphasia;

Ataxic/cerebellar dysarthria: altered rhythm of speech, uneven irregular output, slurred speech (as if inebriated), improper stresses; seen in acute cerebellar damage due to asynergia of speech muscle contractions (*cf.* scanning speech);

Hypokinetic dysarthria: monotonic pitch and volume, as in parkinsonism;

Hyperkinetic dysarthria: several varieties are described, including choreiform (as in Huntington's disease), dystonic (as in tardive dyskinesia, and other dystonic syndromes), tremulous (tremor syndromes), and the dysarthria with vocal tics (including coprolalia) in Gilles de la Tourette syndrome;

Mixed dysarthria: combination of any of above.

- Darley FL, Aronson AE, Brown JR. *Motor speech disorders.* Philadelphia: Saunders 1975

[Cross References: ANARTHRIA; APHASIA; ASYNERGIA; BROCA'S APHASIA; BULBAR PALSY; COPROLALIA; DYSPHONIA; FATIGUE; LOWER MOTOR NEURONE SYNDROME; PARKINSONISM; PSEUDOBULBAR PALSY; SCANNING SPEECH; UPPER MOTOR NEURONE SYNDROME]

Dyscalculia

- see ACALCULIA

Dyschromatopsia

- see ACHROMATOPSIA

Dysdiadochokinesia

Dysdiadochokinesia or adiadochokinesia is a difficulty in performing rapid alternating movements, for example pronation/supination of the arms, tapping alternately with the palm and dorsum of the hand, tapping the foot on the floor.

Dysdiadochokinesia is a sign of cerebellar dysfunction, especially hemisphere disease, and may be seen in association with asynergia, ataxia, dysmetria, and excessive rebound phenomenon. It may reflect the impaired checking response seen in cerebellar disease. Dysdiadochokinesia may also be seen with disease of the frontal lobes or basal ganglia.
[Cross References: ASYNERGIA; ATAXIA; CEREBELLAR SYNDROMES; DYSMETRIA; REBOUND PHENOMENON]

Dysgeusia
Dysgeusia is a complaint of distorted taste perception. It may occur along with anosmia as a feature of upper respiratory tract infections, and has also been described with various drug therapies, in psychiatric disease, and as a feature of zinc deficiency.

- Henkin RI, Patten BM, Pe RK, Bronzert DA. A syndrome of acute zinc loss. Cerebellar dysfunction, mental changes, anorexia and taste and smell dysfunction. *Archives of Neurology* 1975; **32**: 745-51
[Cross References: AGEUSIA; ANOSMIA]

Dysgraphaesthesia
- see AGRAPHOGNOSIA; GRAPHAESTHESIA

Dysgraphia
- see AGRAPHIA

Dyskinesia
Dyskinesia may be used as a general term for excessive involuntary movements, encompassing tremor, myoclonus, chorea, athetosis, tics, stereotypies, and hyperekplexia. The term may be qualified to describe a number of other syndromes of excessive movement, *e.g.*:

Drug-induced dyskinesia: fluid, restless, fidgety movements seen in patients with Parkinson's disease after several years of levodopa therapy, and often described according to their relationship to timing of tablets (*e.g.* peak dose, diphasic), although others are unpredictable (yo-yo-ing); in MPTP-induced parkinsonism, dyskinesias tend to occur early, hence it may be the depth of dopamine deficiency rather than chronicity of treatment which is the key determinant; reduction in overall levodopa use (increased frequency of smaller doses, controlled-release preparations, addition of dopamine agonists) may reduce these effects; amantadine is sometimes helpful.
Paroxysmal dyskinesias, i.e. paroxysmal kinesigenic choreoathetosis/dystonia (PKC; usually responds to carbamazepine), and paroxysmal non-kinesigenic dystonia/choreoathetosis (PDC; does not respond to carbamazepine);
Focal dyskinesias, e.g. orofacial dyskinesia, belly-dancer's dyskinesia; tardive dyskinesia: an involuntary movement disorder developing after years on neuroleptic medications, not necessarily remitting with drug withdrawal; tetrabenazine may help.

- Fahn S. The paroxysmal dyskinesias. In: Marsden CD, Fahn S (eds.). *Movement disorders 3.* Oxford: Butterworth-Heinemann 1994, pp 310-45

[Cross References: ATHETOSIS; CHOREA, CHOREOATHETOSIS; DYSTONIA; HYPEREKPLEXIA; MYOCLONUS; PARKINSONISM; STEREOTYPY; TIC; YO-YO-ING]

Dyslexia
Dyslexia is difficulty or impairment in reading, usually applied to developmental abnormalities of reading ability. A loss of previously acquired reading ability is probably better termed alexia.
[Cross References: ALEXIA]

Dysmentia
The term dysmentia has been suggested as an alternative to dementia, to emphasize the possibility of treating and preventing cognitive decline.
- Chiu E. What's in a name: dementia or dysmentia? *International Journal of Geriatric Psychiatry* 1994; **9**: 1-4
[Cross References: DEMENTIA]

Dysmetria
Dysmetria, or past-pointing, is a disturbance in the control of range of movement in voluntary muscular action, and is one feature of the impaired checking response seen in cerebellar lesions (especially hemisphere lesions). Saccadic dysmetria may also be evident: hypometria (undershoot) is common in parkinsonism, hypermetria (overshoot) is more typical of cerebellar disease.
In cerebellar disorders, dysmetria reflects the asynergia of co-ordinated muscular contraction.
[Cross References: ASYNERGIA; CEREBELLAR SYNDROMES; DYSDIADOCHOKINESIA; PARKINSONISM; REBOUND PHENOMENON; SACCADES]

Dysmorphopsia
The term dysmorphopsia has been proposed for impaired vision for shapes, a visual recognition defect in which visual acuity, colour vision, tactile recognition and visually-guided reaching movements are intact. These phenomena have been associated with bilateral lateral occipital cortical damage (*e.g.* after carbon monoxide poisoning) and are thought to reflect a selective loss of the magnocellular visual pathway. Whether this condition is an agnosia for shape or visual form, or a perceptual problem ("pseudoagnosia"), remains a subject of debate and the term dysmorphopsia has been suggested as a compromise between the different strands of thought.

- Milner AD, Perrett DI, Johnston RS, *et al.* Perception and action in "visual form agnosia". *Brain* 1991; **114:** 405-428
[Cross References: AGNOSIA; VISUAL AGNOSIA]

Dysnomia
- see ANOMIA

Dysphagia

Dysphagia is difficulty swallowing. This may have local gastroenterological causes (tumour, peptic ulceration/stricture) or neurological causes due to pathology occurring anywhere from cerebral cortex to muscle. Neurological control of swallowing is bilaterally represented and so unilateral upper motor neurone lesions may cause only transient problems, *e.g.* poststroke dysphagia is common, but there is evidence of cortical reorganization (neuroplasticity) underpinning recovery. Bilateral upper motor neurone lesions cause persistent difficulties.

Dysphagia of neurological origin may be accompanied by dysphonia, palatal droop, and depressed or exaggerated gag reflex.

Causes of dysphagia include:

- Upper motor neurone pathology: pseudobulbar palsy, *e.g.* motor neurone disease, bilateral cerebrovascular disease, multiple sclerosis;
- Lower motor neurone pathology: bulbar palsy, isolated vagus (X) nerve palsy, jugular foramen syndrome;
- Autonomic neuropathy, *e.g.* Chagas disease, Riley Day syndrome;
- Neuromuscular junction pathology: myasthenia gravis;
- Muscular pathology: polymyositis, oculopharyngeal muscular dystrophy.

Difficulty swallowing may on occasion be functional in origin (globus hystericus).

If swallowing is compromised with a risk of aspiration, feeding may need to be undertaken via nasogastric tube, percutaneous gastrostomy or jejunostomy placed endoscopically (PEG or PEJ), or even parenterally.

- Hamdy S, Aziz Q, Rothwell JC, Singh KD, Barlow J, Hughes DG, Tallis RC, Thompson DG. The cortical topography of human swallowing musculature in health and disease. *Nature Medicine* 1996; **2**: 1217-24

[Cross References: BULBAR PALSY; DYSPHONIA; GAG REFLEX; JUGULAR FORAMEN SYNDROME; PSEUDOBULBAR PALSY]

Dysphasia

- see APHASIA

Dysphonia

Dysphonia is a disorder of the volume, pitch or quality of the voice resulting from dysfunction of the larynx, *i.e.* a disorder of phonation or sound generation. Hence this is a motor speech disorder and could be considered as a dysarthria if of neurological origin.

Dysphonia manifests as hoarseness, or a whispering quality to the voice, which at its extreme results in complete loss of the voice (aphonia). Causes include:

- infection (laryngitis);
- structural abnormalities, *e.g.* polyp, nodule, papilloma of vocal cord;
- neurological causes:

  focal dystonic syndrome: spasmodic dysphonia or laryngeal dystonia (either abductor or adductor); the voice may have a strained and harsh quality, with

low volume and pitch, vocal tremor, and irregularly distributed stoppages; with continuing speech, or if holding a single note, the voice may fade away entirely. These syndromes may be amenable to treatment with botulinum toxin;
flaccid dysphonia, due to superior laryngeal nerve or vagus nerve palsy, bulbar palsy.

- Whurr R, Lorch M, Fontana H, Brookes G, Lees A, Marsden CD. The use of botulinum toxin in the treatment of adductor spasmodic dysphonia. *Journal of Neurology, Neurosurgery and Psychiatry* 1993; **56**: 526-30
[Cross References: APHONIA; BULBAR PALSY; DYSARTHRIA; DYSTONIA; HYPOPHONIA; VOCAL TREMOR, VOICE TREMOR]

Dyspraxia
Dyspraxia is difficulty or impairment in the performance of a voluntary motor act despite an intact motor system and level of consciousness. This may be developmental in origin ("clumsy child"), but in adult practice reflects a loss of function (hence apraxia is a better term).
[Cross References: APRAXIA]

Dysprosody
Dysprosodic speech lacks the melody, intonation, cadence, rhythm, accentuations (*i.e.* the non-linguistic aspects of language) which convey or imply emotion and attitude. This is sometimes known as speech dyspraxia.
Dysprosody may occur with:

~ Anterior left hemisphere damage (with linguistic aspects of aphasia such as agrammatism);
~ Right sided lesions: "emotional dysprosody"; this may also apply to the interpretation of non-linguistic aspects of language.

- Monrad-Krohn GH. Dysprosody or altered "melody of language". *Brain* 1947; **70**: 405-15
[Cross References: APHASIA; APHEMIA; APROSODIA, APROSODY; FISHER'S SIGN]

Dyssynergia
- see ASYNERGIA

Dystaxia
- see ATAXIA

Dystonia
Dystonia, a term first used by Oppenheim in 1911, is a motor syndrome of sustained involuntary muscle contractions causing twisting and repetitive movements and/or abnormal postures. Dystonic movements may initially appear with voluntary movement of the affected part ("action dystonia") but may eventually occur with

voluntary movement elsewhere in the body ("overflow"). The severity of dystonia may be reduced by sensory tricks (*geste antagoniste*), using tactile or proprioceptive stimuli to lessen or eliminate posturing; this feature is unique to dystonia. Dystonia may develop after muscle fatiguing activity, and focal dystonic patients show more rapid fatigue than normals.

Dystonic disorders may be classified according to:

~ *age of onset* (the most significant predictor of prognosis: worse with earlier onset);
~ *distribution* (focal, segmental, multifocal, generalised, hemidystonia);
~ *aetiology:* primary/idiopathic *vs.* secondary/symptomatic.

Primary/idiopathic dystonias include:

- Primary torsion dystonia (idiopathic torsion dystonia)
- Severe generalized dystonia (dystonia musculorum deformans)
- Segemental, multifocal and focal dystonias (*e.g.* torticollis, blepharospasm, writer's cramp)
- Dopa-responsive dystonia (DRD; Segawa's syndrome)
- Myoclonic dystonia

The differential diagnosis of secondary/symptomatic dystonia is broad (more than 40 known causes), including:

- Heredodegenerative disorders: Wilson's disease, Huntington's disease, Hallevorden-Spatz disease, mitochondrial disorders, X-linked dystonia-parkinsonism (lubag)
- Paroxysmal dystonias/dyskinesias: paroxysmal kinesigenic choreoathetosis/ dystonia (PKC; usually responds to carbamazepine), and paroxysmal non-kinesigenic dystonia/choreoathetosis (PDC; does not respond to carbamazepine)
- Metachromatic leukodystrophy
- Gangliosidoses (GM1, GM2)
- Perinatal cerebral injury
- Encephalitis
- Head trauma
- Multiple sclerosis
- Drugs/toxins, *e.g.* antipsychotic, antiemetic, and antidepressant drugs (often relieved within 20 minutes by intramuscular biperiden 5mg or procyclidine 5mg)
- Psychogenic

Appropriate investigations to exclude these symptomatic causes (especially Wilson's disease) are appropriate.

The pathogenesis of dystonia is poorly understood. Different mechanism may apply in different conditions. Peripheral focal dystonias such as torticollis and writer's cramp have been suggested to result from abnormal afferent information relayed from "stiff" muscle spindles.

From a therapeutic point of view, one of the key questions relates to response to levodopa: dopa-responsive dystonia (DRD) responds very well to levodopa (and response fluctuations do not develop over time; *cf.* Parkinson's disease). Other treat-

ments which are sometimes helpful include anticholinergics, dopamine antagonists, dopamine agonists, and baclofen. Botulinum toxin may be very helpful in some focal dystonias (*e.g.* blepharospasm).

- Fahn S, Marsden CD, Calne DB. Classification and investigation of dystonia. In: Marsden CD, Fahn S (eds.). *Movement Disorders 2.* London, Butterworth 1987: 332-58
- Fahn S, Marsden CD. The treatment of dystonia. In: Marsden CD, Fahn S (eds.). *Movement Disorders 2.* London, Butterworth 1987: 359-82
- Grunewald RA, Yoneda Y, Shipman JM, Sagar HJ. Idiopathic focal dystonia: a disorder of muscle spindle afferent processing? *Brain* 1997; **120:** 2179-85
- Van Harten PN, Hoek HW, Kahn RS. Acute dystonia induced by drug treatment. *BMJ* 1999; **319:** 623-6
- Warner TT, Jarman P. The molecular genetics of the dystonias. *Journal of Neurology, Neurosurgery and Psychiatry* 1998; **64:** 427-9

[Cross References: ANISMUS; BLEPHAROSPASM; DYSPHONIA; EYELID APRAXIA; FATIGUE; GAPING; *GESTE ANTAGONISTE*; HEMIDYSTONIA; TORTICOLLIS; WRITER'S CRAMP]

E

Echolalia
Echolalia is the involuntary automatic repetition of an interviewer's speech. This may be observed in a variety of clinical situations:

- Transcortical sensory aphasia, usually as a result of a vascular lesion of the left hemisphere but an analogous situation may be encountered in Alzheimer's disease; "incorporational echolalia", when the patient uses the examiner's question to help form an answer, may be observed as a feature of "dynamic aphasia" which bears resemblance to transcortical motor aphasia, but may result from a frontal lesion;
- Gilles de la Tourette syndrome, as a complex vocal tic, along with coprolalia;
- Alzheimer's disease, Pick's disease, as a symptom of dementia;
- Schizophrenia, as a catatonic symptom;
- Early infantile autism, mental retardation, as a reflection of pathological mental development;
- As a feature of imitation behaviour with frontal lobe lesions;
- Normal children at a particular stage of language acquisition.

[Cross References: APHASIA; COPROLALIA; DYNAMIC APHASIA; IMITATION BEHAVIOUR; JARGON APHASIA; LOGORRHOEA; PALILALIA; TRANSCORTICAL APHASIA]

Echophenomena
- see IMITATION BEHAVIOUR

Echopraxia
Echopraxia is the involuntary automatic imitation of an interviewer's movements. This may be observed as a feature of apraxic syndromes such as corticobasal degeneration, as a complex motor tic in Gilles de la Tourette syndrome, and in frontal lobe disorders (imitation behaviour).
[Cross References: COPROPRAXIA; IMITATION BEHAVIOUR; TIC]

Emotional Lability
Emotional lability, or moria, is an inappropriate expression of emotion, for example uncontrollable ("uninhibited") laughter or crying in situations which although funny or sad are not particularly so. Hence, unlike pathological laughter and crying (*q.v.*) there is congruence of mood and affect. There may be sudden and unpredictable shifts from one extreme to the other. This neurobehavioural state reflects frontal lobe (especially orbitofrontal) lesions, often vascular in origin, and may coexist with disinhibited behaviour and a pseudobulbar palsy ("pseudobulbar affect"); it is

commoner in vascular dementia than Alzheimer's disease. It may also be seen in delirium and in psychiatric disorders (mania).
[Cross References: DELIRIUM; DISINHIBITION; FRONTAL LOBE SYNDROMES; PATHOLOGICAL CRYING; PATHOLOGICAL LAUGHTER; PSEUDOBULBAR PALSY; *WITZELSUCHT*]

Encephalopathy
Encephalopathy is a general term referring to any acute or chronic diffuse disturbance of brain function. Characteristic features include:

- an altered level of consciousness, which may range from drowsiness to a failure of selective attention, to hypervigilance;

with or without:

- disordered perception, memory (*i.e.* cognitive deficits);
- convulsions;
- headache;
- abnormal movements: tremor, myoclonus, asterixis;
- focal neurological deficits (less common).

Clearly these features overlap with those of delirium.
As with terms such as coma and stupor, it is probably better to give a description of the patients clinical state rather than use a term that is open to variable interpretation. Although the term is sometimes reserved for metabolic causes of diffuse brain dysfunction, this usage is not universal. Recognised causes of encephalopathy include:

- Metabolic disorders: hypoxia/ischaemia, hypoglycaemia; organ failure, electrolyte disturbances, hypertension;
- drugs/toxins;
- brain inflammation/infection (*e.g.* encephalitis);
- miscellaneous conditions, *e.g.* Alzheimer's disease, Creutzfeldt-Jakob disease.

[Cross References: ASTERIXIS; COMA; DELIRIUM; MYOCLONUS; STUPOR; TREMOR]

Enophthalmos
Enophthalmos is an inward displacement of the eyeball (sinking or withdrawal) within the socket (*cf.* exophthalmos). It is classically described as one of the cardinal features of Horner's syndrome (along with miosis, ptosis, and anhidrosis) but is seldom actually measured. Enophthalmos may also occur in dehydration (probably the commonest cause), orbital trauma (*e.g.* orbital floor fracture), senile orbital fat atrophy, hemifacial atrophy, and orbital tumour causing tethering and posterior traction on the eyeball.
[Cross References: ANHIDROSIS; EXOPHTHALMOS; HEMIFACIAL ATROPHY; HORNER'S SYNDROME; MIOSIS; PTOSIS]

Entomopia
Entomopia (literally "insect eye") is the name given to a grid-like pattern of multiple copies of the same image; hence, this is a type of polyopia. This phenomenon has been reported in migraine; its pathogenesis is uncertain.

- Lopez JR, Adornato BT, Hoyt WF. "Entomopia": a remarkable case of cerebral polyopia. *Neurology* 1993; **43**: 2145-6
[Cross References: POLYOPIA]

Environmental Dependency
- see IMITATION BEHAVIOUR; UTILIZATION BEHAVIOUR

Environmental Tilt
Environmental tilt, also known as tortopia, is the sensation that visual space is tilted on its side or even upside down ("floor-on-ceiling" phenomenon, "upside-down" reversal of vision). This may last seconds to minutes. The temptation to dismiss such bizarre symptoms as functional should be resisted, since environmental tilt is presumed to reflect damage to cerebellar connections and central vestibular-otolith connections. It has been reported in the following situations:

- lateral medullary syndrome of Wallenberg
- transient ischaemic attacks in basilar artery territory
- demyelinating disease
- head injury
- encephalitis
- following third ventriculostomy for hydrocephalus

[Cross References: LATERAL MEDULLARY SYNDROME; VERTIGO; VESTIBULO-OCULAR REFLEXES]

Epiphora
Epiphora is overflow of tears down the cheek. This may be due to a blocked nasolacrimal duct, or irritation to the cornea causing increased lacrimation, but it may also be neurological in origin, *e.g.* due to the sagging of the lower eyelid in peripheral facial (VII) nerve (Bell's) palsy, or the "crocodile tears" following aberrant facial nerve regeneration.
[Cross References: BELL'S PALSY; CROCODILE TEARS]

Epley Manoeuvre
- see HALLPIKE MANEOUVRE, HALLPIKE TEST; VERTIGO

Esophoria
Esophoria is a variety of heterophoria in which there is a tendency for the visual axes to deviate inward (latent convergent strabismus). Clinically this may be observed using the cover-uncover test as an outward movement of the covered eye as it is uncovered.

Esophoria may occur in individuals with hyperopia (long-sightedness).
[Cross References: COVER TEST, COVER-UNCOVER TEST; EXOPHORIA; HETEROPHORIA]

Esotropia
Esotropia is a variety of heterotropia in which there is manifest inward turning of the visual axis of one eye; the term is synonymous with convergent strabismus. It may be demonstrated using the cover test as an outward movement of the eye which is forced to assume fixation by occlusion of the other eye.

Esotropia may be associated with congenital latent nystagmus (*i.e.* nystagmus appearing when one eye is covered) in the presence of amblyopia; the slow phase in the viewing eye is towards the nose.

Acute esotropia has been described following contralateral thalamic infarction.

[Cross References: AMBLYOPIA; COVER TEST, COVER-UNCOVER TEST; EXOTROPIA; HETEROTROPIA; NYSTAGMUS]

Exophoria
Exophoria is a variety of heterophoria in which there is a tendency for the visual axes to deviate outward (latent divergent strabismus). Clinically this may be observed in the cover-uncover test as an inward movement of the covered eye as it is uncovered. Exophoria may occur in individuals with myopia.

[Cross References: COVER TEST, COVER-UNCOVER TEST; ESOPHORIA; HETEROPHORIA]

Exophthalmos
Exophthalmos is forward displacement of the eyeball, a definition which overlaps with proptosis, as do the causes, the commonest of which is dysthyroid eye disease (Graves' disease).

[Cross References: PROPTOSIS]

Exotropia
Exotropia is a variety of heterotropia in which there is manifest outward turning of the visual axis of an eye; the term is synonymous with divergent strabismus. It may be demonstrated using the cover test as an inward movement of the eye which is forced to assume fixation by occlusion of the other eye.

[Cross References: COVER TEST, COVER-UNCOVER TEST; ESOTROPIA; HETEROTROPIA]

Extensor Posturing
- see DECEREBRATE RIGIDITY

External Ophthalmoplegia
- see OPHTHALMOPARESIS, OPHTHALMOPLEGIA

Extinction
Extinction is the failure to respond to a novel or meaningful sensory stimulus on one side when a homologous stimulus is given simultaneously to the contralateral side (*i.e.* double simultaneous stimulation); it is sometimes called "suppression". The stimuli may be visual, auditory, or tactile, *e.g.* asking the patient to say which hand is touched when the eyes are shut; it is important to show that the patient responds

appropriately to each hand being touched individually, but then neglects one side when both are touched simultaneously.

More subtle defects may be tested using simultaneous bilateral heterologous (asymmetrical) stimuli, although it has been shown that some normal individuals may show extinction in this situation.

A motor form of extinction has been postulated, manifesting as increased limb akinesia when the contralateral limb is used simultaneously.

The presence of extinction is one of the behavioural manifestations of neglect, and most usually follows non-dominant (right) hemisphere lesions.

There is evidence for physiological interhemispheric rivalry or competition in detecting stimuli from both hemifields, which may account for the emergence of extinction following brain injury.

- Fink GR, Driver J, Rorden C, Baldeweg T, Dolan RJ. Neural consequences of competing stimuli in both visual hemifields: a physiological basis for visual extinction. *Annals of Neurology* 2000; **47**: 440-6

[Cross References: AKINESIA; HEMIAKINESIA; NEGLECT; VISUAL EXTINCTION]

Extrapyramidal Signs

- see PARKINSONISM

Eyelid Apraxia

Eyelid apraxia is an inability to open the eyelids at will, although they may open spontaneously at other times (*i.e.* voluntary-automatic dissociation). The term has been criticised on the grounds that this may not always be a true "apraxia", in which case the term "levator inhibition" may be preferred: the open eyelid position is normally maintained by tonic activity of the levator palpebrae superioris. Clinically there is no visible contraction of orbicularis oculi, which distinguishes eyelid apraxia from blepharospasm (however, perhaps paradoxically, the majority of cases of eyelid apraxia occur in association with blepharospasm). Electrophysiological studies do in fact show abnormal muscle contraction in the pre-tarsal portion of orbicularis oculi, which has prompted the suggestion that "focal eyelid dystonia" may be a more appropriate term.

Although the phenomenon may occur in isolation, associations have been reported with:

- Steele Richardson Olszewski syndrome (progressive supranuclear palsy);
- Parkinson's disease;
- Huntington's disease;
- multiple system atrophy;
- MPTP intoxication;
- motor neurone disease;
- acute phase of nondominant hemisphere cerebrovascular event;
- Wilson's disease;
- neuroacanthocytosis.

The precise neuroanatomical substrate is unknown but the association with basal ganglia disorders points to involvement of this region. The underlying mechanisms may be heterogeneous including involuntary inhibition of levator palpebrae superioris.

- Boghen D. Apraxia of eyelid opening: a review. *Neurology* 1997; **48**: 1491-4
[Cross References: APRAXIA; BLEPHAROSPASM; DYSTONIA]

F

"Face-Hand Test"
The "face-hand test" may be useful if hemiparesis or upper limb monoparesis is suspected to be psychogenic: the examiner lifts the paretic hand directly over the patient's face and drops it: in organic weakness the hand will hit the face, whereas patients with functional weakness avoid this.
[Cross References: BABINSKI'S TRUNK-THIGH TEST; HOOVER'S SIGN]

Facial Paresis
Facial paresis (prosopoplegia) may result from:

~ central (upper motor neurone) or peripheral (lower motor neurone; facial nerve) lesions;
~ neuromuscular junction transmission disorders;
~ primary disease of muscle (*i.e.* myogenic).

Facial paresis is clinically heterogeneous which may be helpful with lesion localization.

Facial paresis due to peripheral facial (VII) nerve origin causes ipsilateral weakness of frontalis, orbicularis oculi, buccinator, orbicularis oris and platysma. Clinically this produces:

- drooping of the side of the face with loss of the nasolabial fold;
- widening of the palpebral fissure with failure of lid closure (lagophthalmos);
- eversion of the lower lid with excessive tearing (epiphora);
- inability to raise the eyebrow, close the eye, frown, blow out the cheek, show the teeth, laugh, and whistle;
- there may be dribbling of saliva from the paretic side of the mouth;
- corneal reflex is depressed (efferent limb of reflex arc affected).

Depending on the precise location of the facial nerve injury, there may also be paralysis of the stapedius muscle in the middle ear, causing sounds to seem abnormally loud (especially low tones: hyperacusis), and impairment of taste sensation on the anterior two-thirds of the tongue if the chorda tympani is affected (ageusia, hypogeusia). Lesions within the facial canal distal to the meatal segment cause both hyperacusis and ageusia; lesions in the facial canal between the nerve to stapedius and the chorda tympani cause ageusia but no hyperacusis; lesions distal to the chorda tympani cause neither ageusia nor hyperacusis (*i.e.* facial motor paralysis only). Lesions of the cerebellopontine angle cause ipsilateral hearing impairment and corneal reflex depression (afferent limb of reflex arc affected) in addition to facial weakness.

In upper motor neurone facial weakness ("central facial palsy"), the ability to raise the eyebrow is preserved due to bilateral supranuclear connections to the upper part of the face. A dissociation between volitional and emotional facial movements may also occur: emotional facial palsy refers to the absence of emotional facial movement but

with preserved volitional movements which may be seen with frontal lobe (especially non-dominant hemisphere) precentral lesions (as in abulia, Fisher's sign). Volitional paresis without emotional paresis may occur when corticobulbar fibres are interrupted (precentral gyrus, internal capsule, cerebral peduncle, upper pons).

Causes of lower motor neurone facial paresis include:

- Bell's palsy: idiopathic lower motor neurone facial weakness, assumed to result from a viral neuritis;
- herpes zoster (Ramsey Hunt syndrome);
- diabetes mellitus;
- Lyme disease (borreliosis, Bannwarth's disease);
- sarcoidosis;
- leukaemic infiltration, lymphoma;
- HIV seroconversion;
- neoplastic compression (*e.g.* cerebellopontine angle tumour; rare);
- facial nerve neuroma;
- myasthenia gravis.

These conditions may need to be differentiated from Bell's palsy.

Causes of recurrent lower motor neurone facial paresis include:

- diabetes mellitus;
- Lyme disease (borreliosis, Bannwarth's disease);
- Sarcoidosis;
- Leukaemia, lymphoma.

Causes of upper motor neurone facial paresis include:

Unilateral:

- Hemisphere infarct (with hemiparesis);
- Lacunar infarct (facio-brachial weakness, +/- dysphasia);
- Space occupying lesions: intrinsic tumour, metastasis, abscess.

Bilateral:

- Motor neurone disease;
- Diffuse cerebrovascular disease;
- Pontine infarct (locked-in syndrome).

Myogenic facial paresis may be seen in facioscapulohumeral (FSH) dystrophy, myotonic dystrophy, mitochondrial disorders.

- Borod JC, Koff E, Lorch MP, Nicholas M, Welkowitz J. Emotional and non-emotional facial behaviour in patients with unilateral brain damage. *Journal of Neurology, Neurosurgery and Psychiatry* 1988; **51**: 826-32

[Cross References: ABULIA; AGEUSIA; BELL'S PALSY; BELL'S PHENOMENON; *BOUCHE DE TAPIR;* CEREBELLOPONTINE ANGLE SYNDROME; CORNEAL REFLEX; EPIPHORA; FISHER'S SIGN; HYPERACUSIS; LAGOPHTHALMOS; LOCKED-IN SYNDROME; LOWER MOTOR NEURONE SYNDROME; PSEUDOBULBAR PALSY; UPPER MOTOR NEURONE SYNDROME]

Facilitation

Facilitation is an increase in muscle strength following repeated contraction. Clinically, facilitation can be demonstrated by the appearance of tendon-reflexes after prolonged (*ca.* 30 seconds) forced maximal contractions against resistance, *e.g.* the biceps jerk after elbow flexion, knee jerk after knee extension.

This phenomenon of post-tetanic potentiation is most commonly seen in Lambert-Eaton Myasthenic Syndrome (LEMS), a disorder of neuromuscular junction transmission associated with presence of autoantibodies directed against presynaptic voltage-gated calcium ion (Ca^{2+}) channels (VGCC). The mechanism is thought to be related to an increased build up of Ca^{2+} ions within the presynaptic terminal with repetitive firing of axonal action potentials, partially overcoming the VGCC antibody-mediated ion channel blockade, and leading to release of increasing quanta of acetylcholine.

[Cross References: FATIGUE]

"False Localising Signs"

Neurological signs may be described as "false localising" when their appearance reflects pathology distant from the expected anatomical locus. The classic example is abducent nerve palsy (unilateral or bilateral) in the context of raised intracranial pressure, caused by stretching of the nerve over the ridge of the petrous temporal bone. Other cranial nerve palsies (*e.g.* VII) have also been recorded in cases of idiopathic intracranial hypertension. Shift of intracranial contents as a consequence of raised pressure may also cause an oculomotor (III) nerve palsy, contralateral hemiparesis, and even ipsilateral hemiparesis due to herniation of the temporal lobe through tentorial notch successively compressing the oculomotor nerve, ipsilateral and then contralateral cerebral peduncles.

Spinal cord lesions may also be false localising, with pathology located many (up to 10) segments above the level of the neurological signs.

- Davie C, Kennedy P, Katifi HA. Seventh nerve palsy as a false localising sign. *Journal of Neurology, Neurosurgery and Psychiatry* 1992; **55**: 510-1
- Jamieson DRS, Teasdale E, Willison HJ. False localising signs in the spinal cord. *BMJ* 1996; **312:** 243-4

[Cross References: ABDUCENT NERVE PALSY; URINARY RETENTION]

Fan Sign (*Signe de l'éventail*)

- see BABINSKI'S SIGN

Fasciculation

Fasciculations are rapid, flickering, twitching, involuntary movements within a muscle belly resulting from spontaneous activation of a bundle, or fasciculus, of muscle fibres (*i.e.* a motor unit), which do not move the joint. Fasciculations may also be induced by lightly tapping over a partially denervated muscle belly. The term was formerly used synonymously with fibrillation, but the latter term is now reserved for contraction of a single muscle fibre, or a group of fibres smaller than a motor unit.

Brief and localized fasciculations can be a normal finding (*e.g.* in the intrinsic foot muscles, especially abductor hallucis, and gastrocnemius, but not tibialis anterior), particularly if unaccompanied by other neurological symptoms and signs (wasting,

weakness, sensory disturbance, sphincter dysfunction). Persistent fasciculations most usually reflect a pathological process involving the lower motor neurones in the anterior (ventral) horn of the spinal cord and/or in brainstem motor nuclei. However, fasciculations are not pathognomonic of lower motor neurone pathology since they can on rare occasions be seen in upper motor neurone pathology.

The pathophysiological mechanism of fasciculations is thought to be spontaneous discharge from motor nerves, but the site of origin of this discharge is uncertain. Although ectopic neural discharge from anywhere along the lower motor neurone from cell body to nerve terminal could produce fasciculation, the commonly encountered assumption that it originates in the anterior horn cell body is not supported by the available evidence, which points to a more distal origin in the intramuscular nerve terminals. In addition, denervation of muscle fibres may lead to nerve fibre sprouting (axonal and collateral) and enlargement of motor units which makes fasciculations more obvious.

Fasciculations may be seen in:

- Motor neurone disease with lower motor neurone involvement (*i.e.* progressive muscular atrophy, progressive bulbar atrophy variants)
- Spinal muscular atrophy
- Cervical radiculopathy (restricted to myotomal distribution)
- Multifocal motor neuropathy with conduction block
- Benign fasciculation syndrome - typically only after exercise and without muscle atrophy or weakness
- Cramp fasciculation syndrome
- Kennedy's syndrome (X-linked bulbospinal neuronopathy)
- Almost any lower motor neurone disease, especially compression
- Metabolic causes: thyrotoxicosis, tetany, after acetylcholinesterase inhibitors, anaesthetic muscle relaxants.

- Blexrud MD, Windebank AJ, Daube JR. Long-term follow-up of 121 patients with benign fasciculations. *Annals of Neurology* 1993; **34**: 622-5
- Desai J, Swash M. Fasciculations: what do we know of their significance? *Journal of the Neurological Sciences* 1997; **152 (suppl1):** S43-8
- Layzer RB. The origin of muscle fasciculations and cramps. *Muscle Nerve* 1994; **17**: 1243-9

[Cross References: CALF HYPERTROPHY; FIBRILLATION; LOWER MOTOR NEURONE SYNDROME]

Fatigue

The term fatigue may be used in different contexts to refer to both a sign and a symptom.

The sign of fatigue consists of a reduction in muscle strength with repeated muscular contraction. This most characteristically occurs in disorders of neuromuscular junction transmission (*e.g.* myasthenia gravis), but it may also be observed in disorders of muscle (*e.g.* myopathy, polymyositis) and neurogenic atrophy (*e.g.* motor neurone disease). In myasthenia gravis, fatigue may be elicited in the extraocular muscles by prolonged upgaze; in bulbar muscles by prolonged counting or speech; and in limb

muscles by repeated contraction, especially of proximal muscles (*e.g.* shoulder abduction). Fatigue in myasthenia gravis is understood as a decline in the amount of acetylcholine released from motor nerve terminals with successive impulses, along with a reduced number of functional acetylcholine receptors (AChR) at the motor end-plates, due to binding of AChR antibodies and/or complement mediated destruction of the postsynaptic folds.

Clearly this definition differs from what patients describe as "fatigue"; generally this implies a tiredness or unwillingness to make voluntary muscle contractions, but there is no specific clinical definition. In multiple sclerosis (MS) and chronic fatigue syndromes, a prominent complaint of disabling fatigue may be made when neurological examination reveals little or no neurological deficit. This type of fatigue may be evaluated with various instruments (*e.g.* the Krupp Fatigue Severity Score). The cause of fatigue in MS is unknown: one hypothesis is based on the observation that demyelinated axons show a frequency-dependent conduction block. Interestingly 3,4-diaminopyridine, which has been tried as a treatment for fatigue in MS, has been found to improve subjective symptoms of fatigue but with little impact on these suggested neurophysiological parameters of fatigue. Amantadine has also been used for fatigue in MS.

A gradual decline in the amplitude and speed of initiation of voluntary movements (hypometria, hypokinesia), as seen in disorders of the basal ganglia, especially Parkinson's disease, may also be described as fatigue (*e.g.* "slow" micrographia may be ascribed to fatigue). Steele-Richardson-Olszewski syndrome is notable for lack of fatigue.

- Sheean GL, Murray NMF, Rothwell JC, Miller DH, Thompson AJ. An open-labelled clinical and electrophysiological study of 3,4-diaminopyridine in the treatment of fatigue in multiple sclerosis. *Brain* 1998; **121**: 967-75

[Cross References: DYSTONIA; HYPOKINESIA; HYPOMETRIA; MICROGRAPHIA; WEAKNESS]

Festinant Gait, Festination

Festinant gait or festination is a gait disorder characterized by rapid short steps due to inadequate maintenance of the body's centre of gravity over the legs. To avoid falling and to maintain balance the patient must "chase" the centre of gravity, leading to an increasing speed of gait and a tendency to fall forward when walking (propulsion). A similar phenomenon may be observed if patient is pulled backwards (retropulsion).

Festination is common in idiopathic Parkinson's disease, where it may be related to the flexed posture and impaired postural reflexes commonly seen in these patients; it is less common in symptomatic causes of parkinsonism.

[Cross References: PARKINSONISM; POSTURAL REFLEXES]

Fibrillation

Fibrillation was previously synonymous with fasciculation, but the term is now reserved for the spontaneous contraction of a single muscle fibre, or a group of fibres smaller than a motor unit, hence this is more appropriately regarded as an electrophysiological sign without clinical correlate.

[Cross References: FASCICULATION]

Finger Agnosia
Finger agnosia is a type of tactile agnosia, in which there is inability to identify which finger has been touched when the eyes are closed, despite knowing that a finger has been touched; or inability to point to or move a finger when it is named; or inability to name the fingers This is a disorder of body schema, and may be regarded as a partial form of autotopagnosia.

Finger agnosia is most commonly observed with lesions of the dominant parietal lobe. It may occur in association with acalculia, agraphia, and right-left disorientation, with or without alexia and difficulty spelling words (Gerstmann syndrome). Isolated cases in association with left corticosubcortical posterior parietal infarction have been reported; since this causes no functional deficit, it may be commoner than reported.

- Delia Sala S, Spinnler H. Finger agnosia: fiction or reality? *Archives of Neurology* 1994; **51**: 448-50
[Cross References: AGNOSIA; AUTOTOPAGNOSIA; GERSTMANN SYNDROME]

Fisher's Sign
Fisher's sign is the paucity of facial expression conveying emotional states or attitudes (emotional facial paresis). It follows nondominant (right) hemisphere lesions and may accompany emotional dysprosody of speech.
[Cross References: DYSPROSODY; FACIAL PARESIS]

Flaccidity
Flaccidity is a floppiness which implies a loss of normal muscular tone (hypotonia). This may occur transiently after acute lesions of the corticospinal tracts (flaccid paraparesis), before the development of spasticity, or as a result of lower motor neurone syndromes.
[Cross References: HYPOTONIA; LOWER MOTOR NEURONE SYNDROME]

Flail Arm
Flail arm refers to a severe and symmetric wasting and weakness of the arms without significant functional involvement of other regions, seen in one variant of motor neurone disease ("flail arm syndrome", also known as Vulpian-Bernhart's form). Men are much more frequently affected than women; this group may show improved survival. Alternative designations for this syndrome include amyotrophic brachial diplegia, dangling arm syndrome, and neurogenic man-in-a-barrel syndrome.

- Hu MTM, Ellis CM, Al-Chalabi A, Leigh PN, Shaw CE. Flail arm syndrome: a distinctive variant of amyotrophic lateral sclerosis. *Journal of Neurology, Neurosurgery and Psychiatry* 1998; **65**: 950-1
[Cross References: AMYOTROPHY; "MAN-IN-A-BARREL"]

Flail Foot
- see CAUDA EQUINA SYNDROME; FOOT DROP

***Flexibilitas Cerea* (Waxy Flexibility)**
- see CATATONIA

Flexor Posturing
- see DECORTICATE RIGIDITY

Floccillation
- see CARPHOLOGIA

Foot Drop
Foot drop, often manifest as the foot dragging during the swing phase of the gait, causing tripping and/or falls, may be due to upper or lower motor neurone lesions, which can be distinguished clinically. *Stiff foot drop,* with upper motor neurone lesions, leads to a circumducting gait; it may be possible to see or hear the foot dragging or scuffing along the floor, and this may cause excessive wear on the point of the shoe. There will be other upper motor neurone signs (hemiparesis; spasticity, clonus, hyperreflexia, Babinski's sign). *Floppy foot drop,* with lower motor neurone lesions, leads to a stepping gait (steppage) to try to lift the foot clear of the floor, and a slapping sound on planting the foot. At worst, there is a flail foot in which both the dorsiflexors and the plantar flexors of the foot are weak (*e.g.* in high sciatic nerve or sacral plexus lesions). Other lower motor neurone signs may be present (hypotonia, areflexia or hyporeflexia).
[Cross References: CAUDA EQUINA SYNDROME; HEMIPARESIS; LOWER MOTOR NEURONE SYNDROME; STEPPAGE, STEPPING GAIT; UPPER MOTOR NEURONE SYNDROME]

Foot Grasping
- see GRASP REFLEX

Forced Grasping
- see GRASP REFLEX

Forced Groping
Forced groping describes involuntary movements of a hand in search of an object or item which has touched it or brushed against it; the hand may follow the object around if it is moved (magnetic movements). There may be an accompanying grasp reflex. This type of behaviour may be seen with an alien limb in the context of corticobasal degeneration. Forced groping may be conceptualised as an exploratory reflex which is "released" in the context of frontal lobe pathology, as in utilization behaviour.
[Cross References: ALIEN LIMB; GRASP REFLEX; MAGNETIC MOVEMENTS; UTILIZATION BEHAVIOUR]

Foreign Accent Syndrome
- see APHEMIA

Formication
- see PARAESTHESIA; TINEL'S SIGN

Fortification Spectra
Fortification spectra, or teichopsia, are visual hallucinations, more complex than flashes of light (photopsia) or scintillations, occurring during the aura of migraine with aura ("classical migraine"), and thought to represent spreading depression (of possible ischaemic origin) in the occipital cortex. The appearance is not simply that of battlements, but a radial array akin to the design of medieval castles.
[Cross References: AURA; HALLUCINATION; PHOTOPSIA]

Foster Kennedy Syndrome
The Foster Kennedy syndrome consists of optic atrophy in one eye with optic disc oedema in the other, due to a tumour compressing one optic nerve (to produce atrophy) and causing raised intracranial pressure (to produce papilloedema). A pseudo-Foster Kennedy syndrome is described in consecutive acute ischaemic optic neuropathy.

- Kennedy F. Retrobulbar neuritis as an exact diagnostic sign of certain tumors and abscesses in the frontal lobe. *American Journal of Medical Science* 1911; **142**:355-68.
[Cross References: OPTIC ATROPHY; PAPILLOEDEMA]

Freezing
Freezing is the sudden inability in a patient with parkinsonism to move or to walk, *i.e.* gait failure, as though the patient were turned to ice or the feet were nailed to the floor. This is one of the unpredictable motor fluctuations in late Parkinson's disease which may lead to falls (usually forward onto the knees) and injury. It may occur in confined spaces (*e.g.* doorways), when trying to turn, or when trying to do two things at once. It is not seen in the early years of levodopa therapy.

Two variants are encountered, occurring either during an off period or wearing off period, or randomly, *i.e.* unrelated to drug dosage or timing.

Treatment strategies include use of dopaminergic agents and, anecdotally, L-threodops, but these agents are not reliably helpful, particularly in random freezing. Use of visual targets (real or imagined) may help, *e.g.* stepping over a line.

Freezing may also occur in multiple system atrophy, and has also been reported as an isolated phenomenon.
[Cross References: PARKINSONISM]

Froment's Sign
Froment has two eponymous signs:

- Activated rigidity or synkinesis;
- In an ulnar nerve lesion, flexion of the distal phalanx of the thumb (flexor pollicis longus, innervated by the median nerve) when attempting to squeeze a sheet of paper between the thumb and the index finger, as a compensation for the weakness of thumb adduction (adductor pollicis, innervated by the ulnar nerve). The term is also sometimes used for weakness of little finger adduction, evident when trying to grip a piece of paper between the ring and little finger.

[Cross References: RIGIDITY; SYNKINESIS]

Frontal Lobe Syndromes

The frontal lobes of the brain have enlarged greatly during phylogeny; their diverse connections with the basal ganglia, basal forebrain, and cerebellum, as well as other cortical areas, reflect their multiple motor and behavioural functions. Damage to the frontal lobes may produce a variety of clinical signs, most frequently changes in behaviour. Such changes may easily be overlooked with the traditional neurological examination, although complained of by patient's relatives, and hence specific bedside tests of frontal lobe function should be utilized, for example:

- Verbal fluency: *e.g.* letter/phonemic (F,A,S); category/semantic (animals, foods).
- Proverb interpretation: *e.g.* "Make hay while the sun shines"; "Too many cooks spoil the broth"; interpretation tends to be concrete in frontal lobe disorders.
- Cognitive estimates: *e.g.* height of Post Office Tower, length of a man's spine, distance from London to Edinburgh; may be grossly abnormal or inappropriate.
- Copying motor sequences, to assess motor programming ability: *e.g.* Luria fist-edge- palm test (three step motor sequence with hand).
- Alternating sequence tests: *e.g.* alternating finger flexion/extension out of phase in two hands, repeatedly writing m n m n m n (also used as tests of praxis, which may be affected with frontal lobe pathology); swapping a coin from hand to hand behind back in a predictable pattern and asking patient which hand the coin is in.
- Set-shifting or go/no go tests, in which an alternating pattern is suddenly changed, *e.g.* changing the previously predictable (left/right) pattern of coin hidden in clenched hand swapped over behind back; rhythmic tapping with pen on a surface (I tap once, you tap twice; I tap twice, you tap once); tests of response inhibition (ask patient to clap three times, s/he does so multiple times).

A useful clinico-anatomical classification of frontal lobe syndromes which reflects the functional subdivisions of the frontal lobes is as follows:

Orbitofrontal Syndrome (disinhibited):
disinhibited behaviour (including sexual disinhibition), impulsivity
inappropriate affect, *witzelsucht,* euphoria
emotional lability (moria)
lack of judgement, insight
distractibility, lack of sustained attention; hypermetamorphosis
motor perseverations are not a striking feature

Frontal Convexity Syndrome (apathetic):
apathy; abulia, indifference
motor perseveration
difficulty set-shifting, stimulus boundedness
reduced verbal fluency
deficient motor programming, *e.g.* three step hand sequence, rhythmical tapping (go/no-go test)

Medial Frontal Syndrome (akinetic):
little spontaneous movement, bradykinesia, hypokinesia
sparse verbal output (akinetic mutism)
incontinence

sensorimotor signs in lower limbs
indifference to pain

Overlap between these regional syndromes may occur.

A "*dysexecutive syndrome*" has also been defined, consisting of difficulty planning, adapting to changing environmental demands (impaired cognitive flexibility, *e.g.* in set-shifting tests), and directing attentional resources. This may be seen with dorsolateral (prefrontal) damage.

These frontal lobe syndromes may be accompanied by various neurological signs (frontal release signs or primitive reflexes). Other phenomena associated with frontal lobe pathology include imitation behaviours (echophenomena) and, less frequently, utilization behaviour, features of the environmental dependency syndrome.

- Parkin AJ. *Explorations in cognitive neuropsychology.* Hove: Psychology Press 1996: 220-42
-Trimble MR. *Biological psychiatry.* Chichester: Wiley 1996: 147-56
[Cross References: ABULIA; AKTNESIA; AKINETIC MUTISM; APATHY; ATTENTION; DISINHIBITION; EMOTIONAL LABILITY; FRONTAL RELEASE SIGNS; HYPERMETAMORPHOSIS; HYPERORALITY; HYPERPHAGIA; HYPERSEXUALITY; INCONTINENCE; PERSEVERATION; UTILIZATION BEHAVIOUR; *WITZELSUCHT*]

Frontal Release Signs

Frontal release signs are a constellation of clinical signs evident in the presence of diffuse frontal lobe pathology, usually of vascular or degenerative aetiology. As these responses are present during infancy but disappear during childhood, the term "primitive reflex" may be used. Some are of little value (*e.g.* palmomental reflex). These signs include:

- grasp reflex
- pout reflex (also sometimes known as snout reflex)
- palmomental reflex
- rooting reflex (turning of the head towards a tactile stimulus on the face)
- corneomandibular reflex

Concurrent clinical findings may include dementia, gait disorder (frontal gait, *marche à petit pas*), urinary incontinence, akinetic mutism and *gegenhalten.* Common causes of these findings are diffuse cerebrovascular disease and motor neurone disease.
[Cross References: CORNEOMANDIBULAR REFLEX; *GEGENHALTEN;* GRASP REFLEX; *MARCHE À PETIT PAS;* POUT REFLEX; PALMOMENTAL REFLEX; ROOTING REFLEX]

Fugue

Fugue is a state characterized by loss of personal memory and wandering away from normal surroundings. This may occur in the context of:

- Hysteria;
- Epilepsy;
- Depression (associated with suicide);

- Alcoholism.

Some patients with frontotemporal dementia may spend the day walking long distances, and may be found a long way from home, unable to give an account of themselves, and aggressive if challenged; generally they are able to find their way home (spared topographical memory) despite their other cognitive deficits.
[Cross References: DEMENTIA; SEIZURES]

Funnel Vision
- see "TUNNEL VISION"

G

Gag Reflex

The gag reflex is elicited by touching the posterior pharyngeal wall, tonsillar area, or the base of the tongue, with the tip of a thin wooden ("orange") stick; depressing the tongue with a wooden spatula, and the use of a torch for illumination of the posterior pharynx, may be required to get a good view. There is a palatal response (palatal reflex), consisting of upward movement of the soft palate with ipsilateral deviation of the uvula; and a pharyngeal response (pharyngeal reflex or gag reflex) consisting of visible contraction of the pharyngeal wall. Lesser responses include medial movement, tensing, or corrugation of the pharyngeal wall. In addition there may be head withdrawal, eye watering, coughing, and retching. Hence there is variability of response in different individuals. Some studies claim the reflex is absent in many normal individuals, especially with increasing age, without evident functional impairment; whereas others find it in all healthy individuals, although variable stimulus intensity is required to elicit it.

The afferent limb of the reflex arc is the glossopharyngeal (IX) nerve, the efferent limb in the glossopharyngeal and vagus (X) nerves. Hence individual or combined lesions of the glossopharyngeal and vagus nerves depress the gag reflex, as in neurogenic bulbar palsy.

Dysphagia is common after a stroke, and the gag reflex is often performed to assess the integrity of swallowing. Some argue that absence of the reflex, since it may be normal in elderly individuals, does not predict aspiration and is of little diagnostic value, whereas pharyngeal sensation (feeling the stimulus at the back of the pharynx) is rarely absent in normals and is a better predictor of the absence of aspiration. Others find that even a brisk pharyngeal response in motor neurone disease may be associated with impaired swallowing. Hence the value of the gag reflex remains debatable. A videoswallow may be a better technique to assess the integrity of swallowing.

- Davies AE, Kidd D, Stone SP, MacMahon J. Pharyngeal sensation and gag reflex in healthy subjects. Lancet 1995; **345**: 487-8
- Hughes TAT, Wiles CM. Palatal and pharyngeal reflexes in health and motor neuron disease. *Journal of Neurology, Neurosurgery and Psychiatry* 1996; **61**: 96-8
[Cross References: BULBAR PALSY; DYSPHAGIA]

Gait Apraxia

Gait apraxia is a name given to an inability to walk despite intact motor systems and sensorium. Patients with gait apraxia are often hesitant, seemingly unable to lift their feet from the floor (magnetic gait), or put one foot in front of the other. Arms may be held out at the sides to balance for fear of falling; their fear may be so great that they sit in a chair gripping its sides. These phenomena may be observed with lesions of the frontal lobe and white matter connections, with or without basal ganglia involvement,

for example in diffuse cerebrovascular disease and normal pressure hydrocephalus. A syndrome of isolated gait apraxia has been described with focal degeneration of the medial frontal lobes. In modern classifications of gait disorders, gait apraxia is subsumed into the categories of frontal gait disorder, frontal disequilibrium, and isolated gait ignition failure.

Gait apraxia is an important diagnosis to establish since those afflicted generally respond poorly, if at all, to physiotherapy; moreover, because both patient and therapist often become frustrated because of lack of progress, this form of treatment is often best avoided.

- Nutt JG, Marsden CD, Thompson PD. Human walking and higher-level gait disorders, particularly in the elderly. *Neurology* 1993; **43**: 268-79
- Rossor MN, Tyrrell PJ, Warrington EK, Thompson PD, Marsden CD, Lantos P. Progressive frontal gait disturbance with atypical Alzheimer's disease and corticobasal degeneration. *Journal of Neurology, Neurosurgery and Psychiatry* 1999; **67**: 345-52
[Cross References: APRAXIA]

Ganser Phenomenon

The Ganser phenomenon, or *vorbeireden,* consists of giving approximate answers to questions, which can at times verge on the absurd (Q: "How many legs does a cow have?"; A:"Three"). This may occur in psychiatric disease such as depression, schizophrenia, and malingering, and sometimes in neurological disease (head injury, epilepsy). A Ganser syndrome of hallucinations, conversion disorder, cognitive disorientation and approximate answers is also described but of uncertain nosological validity.

Gaping

Gaping, or involuntary opening of mouth, may occur as a dystonia of the motor trigeminal nerve, also known as Brueghel syndrome after that artist's painting *De Gaper* ("Yawning man", *ca.* 1558) which is said to illustrate a typical case. Afflicted individuals may also demonstrate paroxysmal hyperpnoea and upbeating nystagmus, suggesting a brainstem (possibly pontine) localization of pathology. The condition should be distinguished from other cranial dystonias with blepharospasm (Meige syndrome).

- Gilbert GJ. Brueghel syndrome: its distinction from Meige syndrome. *Neurology* 1996; **46**: 1767-9
[Cross References: BLEPHAROSPASM; DYSTONIA; NYSTAGMUS]

Gaze Palsy

Gaze palsy is a general term for any impairment or limitation in conjugate (yoked) eye movements. This may be supranuclear, nuclear, or infranuclear in origin. Preservation of the vestibulo-ocular reflexes may help differentiate supranuclear gaze palsies from nuclear/infranucelar causes.
[Cross References: SUPRANUCLEAR GAZE PALSY; LOCKED-IN SYNDROME; VESTIBULO-OCULAR REFLEXES]

Gegenhalten

Gegenhalten, or paratonia, or paratonic rigidity, is a resistance to passive movement of a limb changing its posture or position, which is evident in both flexor and extensor muscles (as in rigidity, but not spasticity), which seems to increase further with attempts to get the patient to relax, such that there is a resistance to any applied movement (however, this is not a form of impaired muscle relaxation akin to myotonia and paramyotonia). For instance, when lifting the legs by placing the hands under the knees, the legs may be held extended at the knees despite encouragement on the part of the examiner for the patient to flex the knees.

Gegenhalten is a sign of bilateral frontal lobe dysfunction, especially mesial cortex and superior convexity (premotor cortex, area 6). It is not uncommon in elderly individuals with diffuse frontal lobe cerebrovascular disease.

[Cross References: FRONTAL RELEASE SIGNS; MYOTONIA; PARAMYOTONIA; RIGIDITY; SPASTICITY]

Gerstmann Syndrome

The Gerstmann syndrome consists of acalculia, agraphia (of central type), finger agnosia, and right-left disorientation; there may also be alexia and difficulty spelling words but these are not necessary parts of the syndrome. Gerstmann syndrome occurs with lesions of angular gyrus and supramarginal gyrus of the dominant hemisphere (left posterior parietotemporal region), for example infarction in the territory of the middle cerebral artery.

All the signs do fractionate or dissociate, *i.e.* they are not causally related, or representative of a unitary neuropsychological function, as once suggested. Nonetheless the Gerstmann syndrome remains useful for the purposes of clinical localisation.

- Benton AL. Gerstmann's syndrome. *Archives of Neurology* 1992; **49**: 445-7

[Cross References: ACALCULIA; AGRAPHIA; ALEXIA; FINGER AGNOSIA; RIGHT-LEFT DISORIENTATION]

Geste Antagoniste

Geste antagoniste is a sensory "trick" which alleviates (and is characteristic of) dystonia. *Geste antagoniste* consists of a tactile or proprioceptive stimulus which is learned by the patient, for example touching the chin, face or neck to overcome torticollis, or singing to inhibit blepharospasm. The mechanism is unknown: although afferent feedback from the periphery may be relevant, it is also possible that concurrent motor output to generate the trick movement may be the key element, in which case "sensory trick" is a misnomer.

[Cross References: DYSTONIA]

"Give-Way" Weakness

- see SPASTICITY; WEAKNESS

Glabellar Tap Reflex

The glabellar tap reflex (also known as Myerson's sign) is elicited by repeated tapping with a finger on the forehead, preferably with irregular cadence, whilst observing the eyelids blink (*i.e.* blink relfex). Usually, blinking in response to

tapping habituates quickly, but in extrapyramidal disorders it may not do so. This sign was once thought useful for the diagnosis of idiopathic Parkinson's disease but in fact it is fairly non-specific, occurring in many akinetic-rigid disorders.
[Cross References: BLINK REFLEX; PARKINSONISM]

"Glove and Stocking" Sensory Loss
Sensory loss, to all or selected modalities, confined to the distal parts of the limbs ("glove and stocking") implies the presence of a peripheral neuropathy. If so, motor signs (distal weakness, reflex diminution or loss) may also be present.
[Cross References: NEUROPATHY]

Gowers Sign
Gowers sign is a characteristic manoeuvre used by patients with proximal lower limb and trunk weakness to rise from the ground. From the lying position, the patient rolls to the kneeling position, pushes on the ground with extended forearms to lift the rump and straighten the legs, then uses the hands on the knees to push up the trunk ("climbing up oneself").

This sign was originally described by Gowers in the context of Duchenne muscular dystrophy but may be seen in other causes of proximal leg and trunk weakness, *e.g.* Becker muscular dystrophy, spinal muscular atrophy.

Graefe's Sign
- see VON GRAEFE'S SIGN

Graphaesthesia
Graphaesthesia is the ability to identify numbers or letters written or traced on the skin, first described by Head in 1920. Loss of this ability (agraphaesthesia, dysgraphaesthesia, or graphanaesthesia; sometimes referred to as agraphognosia) is typically observed with parietal lobe lesions, for example in conditions such as corticobasal degeneration. Such a cortical sensory syndrome may also cause astereognosis and impaired two-point discrimination.
[Cross References: AGRAPHAESTHESIA; ASTEREOGNOSIS; TWO-POINT DISCRIMINATION]

Graphanaesthesia
- see AGRAPHAESTHESIA

Graphospasm
- see WRITER'S CRAMP

Grasp Reflex
The grasp reflex consists of progressive forced closure of the hand (contraction of flexor and adductor muscles) when tactile stimulation (*e.g.* the examiner's hand) is moved slowly, exerting pressure, across the palm in an upward direction. Once established, the patient is unable to release the grip (forced grasping), allowing the examiner to pull the arm away. There may also be accompanying groping move-

ments of the hand, once touched, in search of the examiner's hand or clothing (forced groping, magnetic movement). Although categorized a reflex, it may sometimes be accessible to modification by will (so-called alien grasp reflex). It is usually bilateral, even with unilateral pathology. Foot grasping (*i.e.* flexion and adduction of the toes and curling of the sole in response to pressure on the sole), may coexist, as may other frontal release signs (*e.g.* pout reflex, palmomental reflex, *gegenhalten*).

The grasp reflex may be categorized as a frontal release sign (or primitive reflex), since it is most commonly associated with lesion(s) in frontal lobes or deep nuclei and subcortical white matter. Clinicoradiological correlations suggest the cingulate gyrus is the structure most commonly involved, followed by the supplementary motor area.

- De Renzi E, Barbieri C. The incidence of the grasp reflex following hemispheric lesion and its relation to frontal damage. *Brain* 1992; **115:** 293-313

[Cross References: AKINETIC MUTISM; ALIEN GRASP REFLEX; FRONTAL RELEASE SIGNS]

H

Hallpike Manoeuvre, Hallpike Test

The Hallpike manoeuvre (Nylen-Bárány manoeuvre, positioning manoeuvre, Dix-Hallpike positioning test) is a test to induce (or to modify) nystagmus by stimulating the otolith organs of the inner ear. It consists of briskly tilting the patient's head backwards to 30-45° below the horizontal ("head hanging position") and turning it 45° to one side or the other. Prior to performing the manoeuvre, the examiner should warn the patient that s/he may feel "giddy", and to keep their eyes open throughout, since the development of nystagmus with the symptoms of vertigo is the observation of interest to the examiner. With a peripheral lesion (*e.g.* benign paroxysmal positional vertigo, diseases of the labyrinth), nausea, vomiting and rotational-vertical nystagmus occur several seconds after the manoeuvre and then rapidly fatigue (usually < 30 seconds), only to recur when the patient is returned to the upright position, with the nystagmus now in the opposite direction. Repetition of the manoeuvre (if the patient can be persuaded to undergo it) causes less severe symptoms (habituation). This is the diagnostic test for benign paroxysmal positional vertigo. Central lesions (disorders of the vestibular connections) tend to produce isolated nystagmus which does not fatigue or habituate with repetition.

Caloric testing may be required to elicit further the causes of dizziness.

- Dix MR, Hallpike CS. The pathology, symptomatology and diagnosis of certain common disorders of the vestibular system. *Proceedings of the Royal Society of Medicine* 1952; **45:** 341-54
- Lanska DJ, Remler B. Benign paroxysmal positioning vertigo: classic descriptions, origins of the provocative positioning technique, and conceptual developments. *Neurology* 1997; **48**: 1167-77

[Cross References: CALORIC TESTING; NYSTAGMUS; VERTIGO]

Hallucination

An hallucination is a perception in the absence of adequate peripheral stimulus (*cf.* illusion). Such perceptions are substantial, constant, occur in objective space, and are usually not accompanied by insight. They most usually occur in the visual and auditory domains. Visual hallucinations may range in complexity from simple spots or flashes of light (photopsia, scintillation), through more complex patterns (fortification spectra, epileptic aura), to fully formed objects or individuals.

Visual hallucinations may be normal, especially when falling asleep. There are many other causes including both psychiatric and neurological disease, including:

- delirium (especially hyperalert delirium);
- withdrawal states (*e.g.* delirium tremens; hypnotics, anxiolytics);
- overdose (*e.g.* anticholinergic drugs);

- neurodegenerative disease: dementia with Lewy bodies > Alzheimer's disease (these may be associated with cholinergic depletion, and improved with acetylcholinesterase inhibitor drugs); treated idiopathic Parkinson's disease.
- narcolepsy-cataplexy;
- peduncular hallucinosis;
- migraine coma;
- Charles Bonnet syndrome (visual hallucinations of the blind);
- schizophrenia;
- epilepsy.

Different mechanisms may account for visual hallucinations in different conditions: defective visual input and processing may occur in visual pathway lesions, whereas epilepsy may have a direct irritative effect on brain function; visual hallucinations associated with brainstem lesions may result from neurotransmitter abnormalities (cholinergic, serotonergic).

- Barodawala S, Mulley GP. Visual hallucinations. *Journal of the Royal College of Physicians of London* 1997; **31**: 42-8
- Manford M, Andermann F. Complex visual hallucinations. Clinical and neurobiological insights. *Brain* 1998; **121**: 1819-40
[Cross References: DELIRIUM; FORTIFICATION SPECTRA; ILLUSION; PHOTOPSIA]

Head Droop, Head Drop
- see DROPPED HEAD SYNDROME

Head Impulse Test
The head impulse test assesses the vestibulo-ocular reflex. It consists of rapid turning of the head to one side by about 15 degrees; it must be sufficiently rapid to ensure that smooth pursuit eye movements do not compensate for head turning. The examiner observes ability of the subject to maintain fixation on a distant target; if the vestibulo-ocular reflex is intact fixation is maintained. If the vestibulo-ocular reflex is impaired, then an easily visible saccade back to the target occurs at the end of the movement.

This test is recommended in patients suffering a first attack of acute spontaneous vertigo; it is invariably positive in acute unilateral vestibular neuritis (peripheral vestibulopathy).

- Halmagyi GM, Curthoys IS. A clinical sign of canal paresis. *Archives of Neurology* 1988; **45**: 737-9
[Cross References: VESTIBULO-OCULAR REFLEXES]

Head Thrust
- see OCULAR APRAXIA

Head Tremor
Head tremor may be characterized as "yes-yes" (nodding, *tremblement affirmatif*) when predominantly in the vertical plane, or "no-no" (side-to-side, *tremblement*

negatif) when predominantly in the horizontal plane. Head tremor may occur in isolation or with evidence of tremor elsewhere (*e.g.* postural limb tremor, vocal tremor, in essential tremor), or dystonia (*e.g.* torticollis). In essential tremor the head movements are often intermittent, "yes-yes", and of frequency about 7 Hz; dystonic head tremor is often jerky and disorganized, with a frequency of less than 5 Hz. Cerebellum and brainstem disease such as multiple sclerosis can also produce head tremor (or titubation). Head tremor is an exceptionally rare symptom of Parkinson's disease. It may also be seen as a consequence of aortic valve regurgitation (De Musset's sign).

Treatment of head tremor varies with cause:

- essential tremor: propranolol, primidone
- dystonic tremor: anticholinergics, propranolol, botulinum toxin injections
- cerebellar tremor: isoniazid.

[Cross References: DYSTONIA; TREMOR]

"Head Turning Sign"

It is often observed that patients who are cognitively impaired turn their head towards their spouse, partner, or carer to seek assistance when asked to give a history of their problems, or during tests of neuropsychological function. It is a non-specific sign of cognitive impairment;many a normal husband manifests it in the presence of his wife.

Hemeralopia

Hemeralopia, or day blindness, is worsening of vision in bright light (*cf.* nyctalopia). This phenomenon may reflect severe impairment of blood flow to the eye, such that photostressing the macula by exposure to bright light is followed by only slow regeneration of the bleached photopigments.

If due to retinal ischaemia, hemeralopia may be accompanied by neovascularization of the retina. Impoverished perfusion pressure may be demonstrated by pressing on the eyeball (*e.g.* with the thumb) during ophthalmoscopy ("ophthalmodynamometry") and observing the collapse of retinal arteries: thumb pressure greater than diastolic retinal artery pressure causes intermittent collapse; thumb pressure greater than systolic pressure leads to a cessation of pulsation.

Hemeralopia may also occur in retinal diseases such as cone dystrophies.

[Cross References: NYCTALOPIA]

Hemiachromatopsia

- see ACHROMATOPSIA; ALEXIA

Hemiakinesia

Hemiakinesia is akinesia or hypokinesia (inability or difficulty initiating movement) confined to one side of the body. Although hemiakinesia is the norm at the onset of idiopathic Parkinson's disease ("hemiparkinsonism"), persistent hemiakinesia should prompt a re-evaluation of this diagnosis. Corticobasal degeneration often remains unilateral; a search for structural lesions of the basal ganglia should also be undertaken. Hemiakinesia may also indicate motor neglect, usually with right-sided lesions.

Lesions of the basal ganglia, ventral ("motor") thalamus, limbic system, and frontal lobes may cause hemiakinesia.
[Cross References: AKINESIA; EXTINCTION; HEMIPARKINSONISM; HYPOKINESIA; NEGLECT]

Hemianopia

Hemianopia (hemianopsia) is a defect of one half of the visual field. Hemianopic defects may be congruent (homonymous) or non-congruent (heteronymous), and may be detected by standard confrontational testing of the visual fields or automatically (*e.g.* Goldman perimetry). These tests of the visual fields are an extension of the tests for visual acuity which assess areas away from the fovea. Because of the strict topographic arrangement of pathways within the visual system, particular abnormalities of the visual fields give a very precise indication of the likely site of pathology.

Homonymous hemianopia reflects a post-chiasmal lesion. It is important to assess whether the vertical meridian of a homonymous hemianopia cuts through the macula (macula splitting), implying a lesion of the optic radiation; or spares the macula (macula sparing), suggesting an occipital cortical lesion. Incongruous defects may be found with lesions of the optic tract. Commonly homonymous hemianopia results from cerebrovascular disease causing occipital lobe infarction, or intraparnechymal tumour, but it may be a "false localising sign" due to raised intracranial pressure, temporal lobe herniation causing posterior cerebral artery compromise.

Heteronymous defects reflect a chiasmal lesion. The commonest of these is a bitemporal hemianopia due to chiasmal compression, for example by a pituitary lesion or craniopharyngioma. Binasal defects are rare, suggesting lateral compression of the chiasm, for example from bilateral carotid artery aneurysms; binasal hemianopia is also described with optic nerve head lesions. Unilateral temporal hemianopia may result from a lesion anterior to the chiasm which selectively affects only the ipsilateral crossing nasal fibres (junctional scotoma of Traquair).
[Cross References: "FALSE LOCALISING SIGN"; MACULA SPARING, MACULA SPLITTING; QUADRANTANOPIA; SCOTOMA]

Hemiataxia

Hemiataxia is ataxia confined to one half of the body. The vast majority of isolated hemiataxic syndromes reflect a lesion of the ipsilateral cerebellar hemisphere, but on occasion supratentorial lesions may cause hemiataxia (posterior limb of the internal capsule, thalamus). However, in almost all of these cases hemiataxia coexists with ipsilateral hemiparesis (ataxic hemiparesis, *q.v.*), hemisensory disturbance (hemiataxia-hypesthesia), or both.

- Luijckx G-J, Boiten J, Lodder J, Heurs-van Raak L, Wilmink J. Isolated hemiataxia after supratentorial brain infarction. *Journal of Neurology, Neurosurgery and Psychiatry* 1994; **57**: 742-4
[Cross References: ATAXIA; ATAXIC HEMIPARESIS; CEREBELLAR SYNDROMES; CEREBELLOPONTINE ANGLE SYNDROME; LATERAL MEDULLARY SYNDROME]

Hemiballismus

Hemiballismus is unilateral ballismus, an involuntary hyperkinetic movement disorder in which there are large amplitude, vigorous ("flinging") irregular movements. Hemiballismus overlaps clinically with hemichorea ("violent chorea"); the term *hemiballismus-hemichorea* is sometimes used to reflect this overlap. Hemiballismic limbs may show a loss of normal muscular tone (hypotonia).

Anatomically, hemiballismus is most often associated with lesions of the contralateral subthalamic nucleus of Luys or its efferent pathways, although there are occasional reports of its occurrence with lesions of the caudate, putamen, globus pallidus, lentiform nucleus, thalamus, and precentral gyrus; and even with ipsilateral lesions. Pathologically, vascular events (ischaemia, haemorrhage) are the commonest association but hemiballismus has also been reported with space-occupying lesions (tumour, arteriovenous malformation), inflammation (encephalitis, systemic lupus erythematosus, post-streptococcal), demyelination, metabolic causes (hyperosmolal non-ketotic hyperglycaemia), infection (toxoplasmosis in AIDS), drugs (oral contraceptives, phenytoin, levodopa, neuroleptics) and head trauma.

Pathophysiologically, hemiballismus is though to result from reduced conduction through the direct pathway within the basal ganglia-thalamo-cortical motor circuit (as are other hyperkinetic involuntary movements, such as choreoathetosis). Removal of excitation from the globus pallidus following damage to the efferent subthalamic-pallidal pathways disinhibits the ventral anterior and ventral lateral thalamic nuclei which receive pallidal projections and which in turn project to the motor cortex.

Hemiballismus of vascular origin usually improves spontaneously, but drug treatment with neuroleptics (haloperidol, pimozide, sulpiride) may be helpful. Other drugs which are sometimes helpful include tetrabenazine, reserpine, clonazepam, clozapine, and sodium valproate.

- Albin RL, Young AB, Penney JB. The functional anatomy of basal ganglia disorders. *Trends in Neurosciences* 1989; **12**: 366-75
- Lee MS, Marsden CD. Movement disorders following lesions of the thalamus or subthalamic region. *Movement Disorders* 1994; **9**: 493-507
- Martin JP. Hemichorea resulting from a local lesion of the brain. (The syndrome of the body of Luys.) *Brain* 1927; **50**: 637-51

[Cross References: BALLISM, BALLISMUS; CHOREA; HEMICHOREA; HYPOTONIA]

Hemichorea

Hemichorea is unilateral chorea, an involuntary movement disorder which overlaps with hemiballismus, and with which it shares similar pathophysiology and aetio-logy. It may replace hemiballismus during recovery from a contralateral subthalamic lesion.

[Cross References: CHOREA; HEMIBALLISMUS]

Hemidystonia

Hemidystonia is dystonia affecting the whole of one side of the body, a pattern which mandates structural brain imaging because of the chance of finding a causative lesion (vascular, neoplastic), which is greater than with other patterns of dystonia (focal, segmental, multifocal, generalized). Such a lesion most often affects the contralateral

putamen or its afferent or efferent connections.

- Marsden CD, Obeso JA, Zaranz JJ, Lang AE. The anatomical basis of symptomatic hemidystonia. *Brain* 1985; **108**: 461-83
[Cross References: DYSTONIA]

Hemifacial Atrophy
Hemifacial atrophy is thinning of subcutaneous tissues on one side of the face; it may also involve muscle and bone (causing enophthalmos), and sometimes brain, in which case neurological features (hemiparesis, hemianopia, focal seizures, cognitive impairment) may also be present. The clinical heterogeneity of hemifacial atrophy probably reflects a pathogenetic heterogeneity. The syndrome, sometimes referred to as Parry-Romberg syndrome, may result from maldevelopment of autonomic innervation or vascular supply, or as an acquired feature following trauma or a consequence of linear scleroderma (morphoea), in which case a *coup de sabre* may be seen.

- Larner AJ, Bennison DP. Some observations on the aetiology of hemifacial atrophy ("Parry-Romberg syndrome"). *Journal of Neurology, Neurosurgery and Psychiatry* 1993; **56**: 1035-6
[Cross References: *COUP DE SABRE;* ENOPHTHALMOS; HEMIANOPIA; HEMIPARESIS]

Hemifacial Spasm
Hemifacial spasm is an involuntary dyskinetic (not dystonic) movement disorder consisting of contractions of muscles on one side of the face, sometimes triggered by eating or speaking, and exacerbated by fatigue or emotion. The movements give a twitching appearance to the eye or side of the mouth, sometimes described as a pulling sensation. Patients often find this embarrassing because it attracts attention. The movements may continue during sleep.

The movements often result from compression of the facial (VII) nerve, usually at the root entry zone, often by a tortuous anterior or posterior inferior cerebellar artery. Other causes include intrapontine lesions (*e.g.* demyelination), following a Bell's palsy, and mass lesions (tumour, arteriovenous malformation) located anywhere from the facial nucleus to the stylomastoid foramen.

Structural lesions may be amenable to surgical resection. For idiopathic hemifacial spasm, or patients declining surgery, botulinum toxin injections are the treatment of choice.
[Cross References: BELL'S PALSY; DYSKINESIA]

Hemiparesis
Hemiparesis is a weakness affecting one side of the body, less severe than a hemiplegia. Characteristically this affects the extensor muscles of the upper limb more than flexors, and the flexors of the leg more than extensors ("pyramidal" distribution of weakness), producing the classic hemiparetic/hemiplegic posture with flexed arm and extended leg, the latter permitting standing and a circumducting gait. Hemiparesis results from damage (most usually vascular) to the corticospinal pathways anywhere from motor cortex to the cervical spine. Accompanying signs

may give clues as to localisation, the main possibilities being hemisphere, brainstem, or cervical cord. Hemisphere lesions may also cause hemisensory impairment, hemianopia, aphasia, agnosia or apraxia; headache, and incomplete unilateral ptosis, may sometimes feature. Spatial neglect, with or without anosognosia, may also occur, particularly with right sided lesions producing a left hemiparesis. Pure motor hemiparesis may be seen with lesions of the internal capsule, corona radiata, and basal pons (lacunar infarct), in which case the face and arm are affected more than the leg (faciobrachial syndrome). Brainstem lesions may produce diplopia, ophthalmoplegia, nystagmus, ataxia, and crossed facial sensory loss or weakness in addition to hemiparesis. Spinal lesions are more likely to show bilateral long tract signs (*e.g.* bilateral Babinksi's sign) and may have accompanying spinal or root pain, sphincter disturbance, and a sensory or motor level.

Hemiparesis is most usually a consequence of a vascular event (cerebral infarction). Tumour may cause a progressive hemiparesis (although meningiomas may produce transient "stroke-like" events). Hemiparetic multiple sclerosis is rare but well described. Transient hemiparesis may be observed as an ictal phenomenon (Todd's paresis), or in familial hemiplegic migraine which is associated with mutations in a voltage-gated Ca^{2+} ion channel gene.

[Cross References: AGNOSIA; ANOSOGNOSIA; APHASIA; APRAXIA; BABINSKI'S SIGN; "FALSE LOCALISING SIGNS"; HEMIANOPIA; NEGLECT; PTOSIS; UPPER MOTOR NEURONE SYNDROME; WEAKNESS]

Hemiparkinsonism

Hemiparkinsonism describes the finding of parkinsonian signs restricted to one side of the body, most usually akinesia, in which case the term hemiakinesia may be used. Idiopathic Parkinson's disease may present with exclusively or predominantly unilateral features (indeed, lack of asymmetry at onset may argue against this diagnosis) but persistent hemiparkinsonism, particularly if unresponsive to adequate doses of levodopa, should alert the clinician to other possible diagnoses, including corticobasal degeneration or structural lesions.

[Cross References: HEMIAKINESIA; PARKINSONISM].

Hemiplegia

Hemiplegia is a complete weakness affecting one side of the body, *i.e.* a clinically more severe picture than a hemiparesis.

[Cross References: HEMIPARESIS; WEAKNESS]

Hertwig-Magendie Sign

- see SKEW DEVIATION

Heterochromia Iridis

- see HORNER'S SYNDROME

Heterophoria

Heterophoria is a generic term for a latent tendency to imbalance of the ocular axes (latent strabismus; *cf.* heterotropia). This may be clinically demonstrated using the cover-uncover test: if there is movement of the covered eye as it is uncovered and

takes up fixation, this reflects a phoria. Phorias may be in the horizontal (esophoria, exophoria) or vertical plane (hyperphoria, hypophoria).

- Shaunak S, O'Sullivan E, Kennard C. Eye movements. In: Hughes RAC (ed.). *Neurological Investigations.* London: BMJ Publishing 1997: 253-82
[Cross References: COVER TEST, COVER-UNCOVER TEST; ESOPHORIA; EXOPHORIA; HETEROTROPIA; HYPERPHORIA; HYPOPHORIA]

Heterotropia

Heterotropia is a generic term for manifest deviation of the eyes (manifest strabismus; *cf.* heterophoria), synonymous with squint. This may be obvious; an amblyopic eye, with poor visual acuity and fixation, may become deviated. Sometimes it may be more subtle, coming to attention only with the patient's complaint of diplopia. Using the alternate cover test, in which binocular fixation is not permitted, an imbalance in the visual axes may be demonstrated, but this will not distinguish between heterotropia and heterophoria; to make this distinction the cover test is required: if the uncovered eye moves to adopt fixation then heterotropia is confirmed. Tropias may be in the horizontal (esotropia, exotropia) or vertical plane (hypertropia, hypotropia).

- Shaunak S, O'Sullivan E, Kennard C. Eye movements. In: Hughes RAC (ed.). *Neurological Investigations.* London: BMJ Publishing 1997: 253-82
[Cross References: AMBLYOPIA; COVER TEST, COVER-UNCOVER TEST; ESOTROPIA; EXOTROPIA; HETEROPHORIA; HYPERTROPIA; HYPOTROPIA]

Hiccups

A hiccup (hiccough) is a brief burst of inspiratory activity involving the diaphragm and the inspiratory intercostal muscles with reciprocal inhibition of expiratory intercostal muscles. The sound ("hic") and discomfort result from glottic closure immediately after the onset of diaphragmatic contraction. This may be characterized as a physiological form of myoclonus (or singulus).

Most episodes are self-limited, but prolonged or intractable hiccuping should prompt the search for a structural or functional cause, either gastroenterological or neurological; hiccuping is seldom the only abnormality if the cause is neurological since it usually reflects pathology within the medulla or affecting the afferent and efferent nerves of the respiratory muscles. Medullary causes include:

- infarction (posterior inferior cerebellar artery territory);
- tumour;
- abscess;
- tuberculoma;
- syrinx;
- haematoma;
- demyelination;
- CNS infection, *e.g.* viral encephalitis.

Treatment should be aimed at the underlying cause. If none is identified, physical measures to stop the hiccups such as rebreathing may then be tried. Of the many various pharmacotherapies tried, the best are probably baclofen and chlorpromazine.

- Fetter M, Kennard C. Hiccup. In: Brandt T, Caplan LR, Dichgans J, Diener HC, Kennard C (eds.). *Neurological disorders: course and treatment.* San Diego: Academic Press 1996: 145-8
- Howard RS. Persistent hiccups. *BMJ* 1992; **305**: 1237-8
[Cross References: MYOCLONUS]

Hippus
Hippus is excessive pupillary unrest, *i.e.* rhythmic contraction and dilatation of the pupil. It may reflect an imbalance between afferent pupillary sympathetic and parasympathetic autonomic activity. Hippus may be a normal phenomenon; it may be observed during recovery from an oculomotor (III) nerve palsy, but otherwise is of no localizing significance.

Hoffmann's Sign
Hoffmann's sign or reflex is a digital reflex consisting of flexion of the thumb and index finger in response to snapping the distal phalanx of the middle finger. Although sometimes a normal finding, it may be indicative of a corticospinal tract lesion above C5 or C6.
[Cross References: TRÖMNER'S SIGN; UPPER MOTOR NEURONE SYNDROME]

Hoffmann-Tinel Sign
- see TINEL'S SIGN

Holmes-Adie Pupil, Holmes-Adie Syndrome
The Holmes-Adie, or tonic, pupil is an enlarged pupil which, in a darkened environment, is unresponsive to a phasic light stimulus, but may respond slowly to a tonic light stimulus. Reaction to accommodation is preserved, hence this is one of the causes of light-near pupillary dissociation (*q.* v.). A Holmes-Adie pupil is usually unilateral, and hence a cause of anisocoria.

Holmes-Adie pupil may be associated with other neurological features (Holmes-Adie syndrome). These include loss of lower limb tendon reflexes (especially ankle jerks); impaired corneal sensation; chronic cough; and localised or generalised anhidrosis, sometimes with hyperhidrosis (Ross's syndrome). Holmes-Adie syndrome is much commoner in women than men.

Pathophysiologically Holmes-Adie pupil results from a peripheral lesion and shows denervation supersensitivity, constricting with application of dilute (0.2%) pilocarpine (*cf.* pseudo-Argyll Robertson pupil).
[Cross References: ANHIDROSIS; ANISOCORIA; HYPERHIDROSIS; LIGHT-NEAR PUPILLARY DISSOCIATION; PSEUDO-ARGYLL ROBERTSON PUPIL]

Hoover's Sign
Hoover's sign may be used to help differentiate organic from functional hemiplegia or monoplegia. It is based on the fact that when a recumbent patient attempts to lift one leg, downward pressure is felt under the heel of the other leg, a normal synkinetic movement. The finding of this normal synkinetic movement, detected when the heel

of the supposedly paralysed leg presses down on the examiner's palm, constitutes Hoover's sign: no increase in pressure is felt beneath the heel of a paralysed leg in an organic hemiplegia.

In addition, the synkinetic hip extension movement is accentuated when attempting to raise a contralateral paretic leg, whereas in functional weakness it is abolished.

[Cross References: BABINSKI'S TRUNK-THIGH TEST; "FACE-HAND TEST"; SYNKINESIS]

Horner's Syndrome

Horner's syndrome results from impairment of ocular sympathetic innervation and is defined by a constellation of clinical findings, most usually occurring unilaterally, *viz.:*

- partial ptosis, due to weakness of Müller's muscle;
- miosis, due to the unopposed action of the sphincter pupillae muscle, innervated by the parasympathetic nervous system;
- anhidrosis, a loss of sweating (if the lesion is distal to the superior cervical ganglion);
- enophthalmos, retraction of the eyeball (though this is seldom measured).

The first two mentioned signs are usually the most evident and bring the patient to medical attention; the latter two are usually less evident or absent.

Additional features which may be seen include:

- heterochromia iridis, different colour of the iris (if the lesion is congenital);
- elevation of the inferior eyelid due to a weak inferior tarsal muscle ("reverse ptosis", or "upside-down ptosis").

The sympathetic innervation of the eye consists of a long, three neurone, pathway, extending from the diencephalon down to the cervicothoracic spinal cord, then back up to the eye via the superior cervical ganglion and the internal carotid artery, and the ophthalmic division of the trigeminal (V) nerve. A wide variety of pathological processes, spread across a large area, may cause a Horner's syndrome, although many examples remain idiopathic. Causes include:

- brainstem/cervical cord disease (vascular, demyelination, syringomyelia);
- Pancoast tumour;
- malignant cervical lymph nodes;
- carotid aneurysm, carotid artery dissection;
- involvement of T1 fibres, *e.g* in T1 radiculopathy, or lower trunk brachial plexopathy;
- cluster headache;
- congenital.

[Cross References: ANHIDROSIS; ANISOCORIA; ENOPHTHALMOS; MIOSIS; PLEXOPATHY; PTOSIS; RADICULOPATHY]

Hyperacusis

Hyperacusis is an abnormal loudness of sounds, especially low tones, due to paralysis of the stapedius muscle which functions normally to damp conduction across the ossi-

cular chain of the middle ear. This most commonly occurs with lower motor neurone facial (VII) nerve (Bell's) palsy, located proximal to the nerve to stapedius. Ageusia may also be present if the chorda tympani branch of the facial nerve is involved.

Hyperacusis may occassionally occur with central (brainstem) lesions.

[Cross References: AGEUSIA; BELL'S PALSY; FACIAL PARESIS]

Hyperaesthesia

Hyperaesthesia is increased sensitivity to sensory stimulation of any modality, *e.g.* pain (hyperalgesia), touch.

[Cross References: ANAESTHESIA]

Hyperalgesia

Hyperalgesia is the perception of increased intensity of pain from a stimulus which is normally painful (*cf.* allodynia). Paradoxically this may sometimes be induced by morphine.

[Cross References: ALLODYNIA; DYSAESTHESIA; HYPERPATHIA]

Hyperekplexia

Hyperekplexia (literally, to jump excessively) is an involuntary movement disorder in which there is a pathologically exaggerated startle response, usually to sudden unexpected auditory stimuli, but sometimes also to tactile (especially trigeminal) and visual stimuli. The startle response is a sudden shock-like movement which consists of eye blink, grimace, abduction of the arms, and flexion of the neck, trunk, elbows, hips, and knees. The muscular jerk of startle satisfies the definition of myoclonus. Ideally for hyperekplexia to be diagnosed there should be a physiological demonstration of exaggerated startle response, but this criterion is seldom adequately fulfilled.

Hyperekplexia syndromes may be classified as:

Idiopathic (the majority);

Hereditary/familial: an autosomal dominant disorder with muscular hypertonia in infancy, leg jerks and gait disorder. Familial cases have been associated with mutations in the α_1 subunit of the inhibitory glycine receptor gene;

Symptomatic:

- perinatal ischaemic-hypoxic encephalopathy
- brainstem lesions (encephalitis, haemorrhage)
- thalamic lesions (inflammation, vascular)
- drugs (cocaine, amphetamines)
- Gilles de la Tourette syndrome

Attacks may respond to the GABA agonist clonazepam.

- Matsumoto J, Hallett M. Startle syndromes. In: Marsden CD, Fahn S (eds.) *Movement disorders 3.* Boston: Butterworth 1994: 418-33

- Shiang R, Ryan SG, Zhu Y-Z, *et al.* Mutational analysis of familial and sporadic hyperekplexia. *Annals of Neurology* 1995; **38**: 85-91

[Cross References: INCONTINENCE; MYOCLONUS]

Hypergraphia
Hypergraphia is excessive writing. This may be seen as part of the interictal psychosis which may develop in patients with complex partial seizures from a temporal lobe (especially non-dominant hemisphere) focus, or with other non-dominant temporal lobe lesions (vascular, neoplastic) or psychiatric disorders (schizophrenia). Hypergraphia is a feature of Geschwind's syndrome, along with hyperreligiosity and hyposexuality.

- Benson DF. The Geschwind syndrome. *Advances in Neurology* 1991; **55**: 411-21
[Cross References: HYPERRELIGIOSITY; HYPOSEXUALITY]

Hyperhidrosis
Hyperhidrosis is excessive (unphysiological) sweating. This may be "essential" (*i.e.* without obvious cause), or seen as a feature of Parkinson's disease, or occurring in a band above a spinal cord injury. Localised hyperhidrosis caused by food (gustatory sweating) may result from aberrant connections between nerve fibres supplying sweat glands and salivary glands. Other causes of hyperhidrosis include mercury poisoning, phaeochromocytoma, and tetanus. Transient hyperhidrosis contralateral to a large cerebral infarct in the absence of autonomic dysfunction has also been described. Regional syndromes of hyperhidrosis (hands, feet, axillae) are also described.

Treatment is difficult. Symptoms may be helped (but not abolished) by low dose anticholinergic drugs, clonidine or propantheline. For focal syndromes, botulinum toxin injections or sympathectomy may be helpful.

- Collin J, Whatling P. Treating hyperhidrosis. *BMJ* 2000; **320**: 1221-2
- Labar DR, Mohr JP, Nichols FT, Tatemichi TK. Unilateral hyperhidrosis after cerebral infarction. *Neurology* 1988; **38**: 1679-82
[Cross References: ANHIDROSIS; DIAPHORESIS; HOLMES-ADIE PUPIL, HOLMES-ADIE SYNDROME]

Hyperkinesia
Hyperkinesia indicates a disorder characterized by excessive movement, such as ballism, or chorea, or the speech disorders occurring with them.
[Cross References: BALLISM, BALLISMUS; CHOREA, CHOREOATHETOSIS; DYSARTHRIA]

Hypermetamorphosis
Hypermetamorphosis is an overattention to external stimuli. Patients with hypermetamorphosis may explore compulsively and touch everything in their environment. This is one element of the environmental dependency syndrome and may be associated with other forms of utilization behaviour, imitation behaviour (echolalia, echopraxia) and frontal release signs such as the grasp reflex. It occurs with severe frontal lobe damage and may be observed following recovery from herpes simplex encephalitis and in frontal lobe dementias including Pick's disease. Bitemporal lobectomy may also result in hypermetamorphosis, as a feature of the Klüver-Bucy syndrome.
[Cross References: ECHOLALIA; ECHOPRAXIA; FRONTAL RELEASE SIGNS; GRASP REFLEX; IMITATION BEHAVIOUR; KLÜVER-BUCY SYNDROME; UTILIZATION BEHAVIOUR]

Hypermetria
- see DYSMETRIA

Hyperorality
Hyperorality is a neurobehavioural abnormality consisting of drinking more than usual, eating excessively, eating anything in sight, and putting objects inappropriately into the mouth. It is a feature of frontal lobe pathology. It is one element of the Klüver-Bucy syndrome, along with hypersexuality.
[Cross References: KLÜVER-BUCY SYNDROME]

Hyperpathia
Hyperpathia is an overreaction to cutaneous sensory stimuli such as light touch or hot and cold stimuli, especially repetitive stimuli, such that they cause unpleasant sensations, often of a burning painful type. The term thus overlaps to some extent with hyperalgesia (although the initial stimulus need not be painful itself) and dysaesthesia. There is an accompanying diminution of sensibility due to raising of the sensory threshold (*cf.* allodynia), and the pain is not stimulus-bound (*i.e.* spreads beyond the area of stimulation).

Hyperpathia is a feature of thalamic lesions, and hence tends to involve the whole of one side of the body following a unilateral lesion such as a cerebral haemorrhage or thrombosis. Generalized hyperpathia may also be seen in variant Creutzfeldt-Jakob disease, in which posterior thalamic (pulvinar) lesions are said to be a characteristic radiological finding.
[Cross References: ALLODYNIA; DYSAESTHESIA; HYPERALGESIA]

Hyperphagia
Hyperphagia is increased or excessive eating. Binge eating, particularly of sweet things, is one of the neurobehavioural disturbances seen in certain of the fronto-temporal dementias. Hyperphagia may be one feature of a more general tendency to put things in the mouth (hyperorality), for example in the Klüver-Bucy syndrome.
[Cross References: HYPERORALITY; KLÜVER-BUCY SYNDROME]

Hyperphoria
Hyperphoria is a variety of heterophoria in which there is a latent upward deviation of the visual axis of one eye. Using the cover-uncover test, this may be observed clinically as the downward movement of the eye as it is uncovered.
[Cross References: COVER TEST, COVER-UNCOVER TEST HYPOPHORIA; HETEROPHORIA]

Hyperreflexia
Hyperreflexia is an exaggerated briskness of the tendon reflexes. This may be physiological in an anxious patient (reflexes often denoted ++), or pathological in the context of corticospinal pathway pathology (upper motor neurone syndrome, often denoted +++). It is often difficult to distinguish normal from pathological brisk reflexes. Hyperreflexia (including a jaw jerk) in isolation cannot be used to diagnose an upper motor neurone syndrome, and asymmetry of reflexes is a soft sign. On the

other hand, upgoing plantar responses are a hard sign of upper motor neurone pathology; other accompanying signs (weakness, sustained clonus, absent abdominal reflexes) also indicate abnormality.

Hyperreflexia reflects an increased gain in the stretch reflex; this may be due to impaired descending inhibitory inputs to the monosynaptic reflex arc. Rarely pathological hyperreflexia may occur in the absence of spasticity, suggesting different neuroanatomical substrates underlying these phenomena.

"Hyperreflexia" of the bladder detrusor muscle may be a cause of urinary urge incontinence.

- Sherman SJ, Koshland GF, Laguna JF. Hyper-reflexia without spasticity after unilateral infarct of the medullary pyramid. *Journal of the Neurological Sciences* 2000; **175**: 145-55

[Cross References: ABDOMINAL REFLEXES; CLONUS; INCONTINENCE; JAW JERK; REFLEXES; UPPER MOTOR NEURONE SYNDROME; SPASTICITY; WEAKNESS]

Hyperreligiosity

Hyperreligiosity is a neurobehavioural symptom, manifest as sudden religious conversion, or increased and unswerving orthodoxy in devotion to religious rituals. It may be encountered along with hypergraphia and hyposexuality as a feature of Geschwind's syndrome. It has also been observed in some patients with fronto-temporal dementia; the finding is cross-cultural, having been described in Christians, Moslems, and Sikhs.

- Benson DF. The Geschwind syndrome. *Advances in Neurology* 1991; 55: 411-21

[Cross References: HYPERGRAPHIA]

Hypersexuality

Hypersexuality is a pathological increase in sexual drive and activity. Recognised causes include bilateral temporal lobe damage, as in the Klüver-Bucy syndrome, septal damage, and drug-treatment in Parkinson's disease. Hypersexuality is also a feature of the Kleine-Levin syndrome. Sexual disinhibition may be a feature of frontal lobe syndromes, particularly of the orbitofrontal cortex.

[Cross References: DISINHIBITION; FRONTAL LOBE SYNDROMES; KLÜVER-BUCY SYNDROME]

Hypersomnolence

Hypersomnolence is characterized by excessive daytime sleepiness, with a tendency to fall asleep at inappropriate times and places, for example during meals, telephone conversations, at the wheel of a car. Causes include:

- Narcolepsy (may be accompanied by other features such as sleep paralysis, hypnagogic hallucinations, cataplexy);
- Midbrain lesions
- Idiopathic CNS hypersomnia;

- Kleine-Levin syndrome;
- Nocturnal hypoventilation, due to:
  - sleep apnoea syndrome (obstructive sleep apnoea; Pickwickian syndrome)
  - chest wall anomalies
  - neuromuscular and myopathic disorders affecting the respiratory muscles, especially the diaphragm, for example:
    - motor neurone disease
    - myotonic dystrophy
    - metabolic myopathies, *e.g.* acid maltase deficiency
    - mitochondrial disorders
- Drugs.

Nocturnal hypoventilation causes arterial oxygen desaturation which may lead to disturbed sleep and morning headaches. Clinical signs may include a bounding hyperdynamic circulation and sometimes papilloedema, as well as features of underlying neuromuscular disease. Cognitive impairment (slowing) may be one of the presenting signs of obstructive sleep apnoea. Investigations may reveal a raised haematocrit and early morning hypoxia. Sleep studies confirm nocturnal hypoventilation with dips in arterial oxygen saturation. Treatment is with nocturnal intermittent positive pressure ventilation.
[Cross References: ASTERIXIS; CATAPLEXY; PAPILLOEDEMA; PARADOXICAL DIAPHRAGM MOVEMENT]

Hyperthermia
Body temperature is usually regulated within narrow limits through the co-ordinating actions of a centre for temperature control ("thermostat"), located in the hypothalamus (anterior-preoptic area), and effector mechanisms (shivering, sweating, panting, vasoconstriction, vasodilation), controlled by pathways located in or running through the posterior hypothalamus and peripherally in the autonomic nervous system. Lesions of the anterior hypothalamus (*e.g.* trauma, ischaemia, inflammation, tumour) may result in hyperthermia (*cf.* hypothermia).
Other causes of hyperthermia include:

- Infection: bacteria,viruses (pyrogens, *e.g.* interleukin-1);
- Malignant hyperthermia;
- Neuroleptic malignant syndrome (hyperthermia, rigidity, autonomic dysfunction);
- Heatstroke.

[Cross References: HYPOTHERMIA]

Hypertonia, Hypertonus
Hypertonia of hypertonus is an exaggeration of normal muscular tone, usually implying spasticity of corticospinal pathway origin rather than extrapyramidal (leadpipe) rigidity.
[Cross References: CLASP-KNIFE PHENOMENON; HYPERREFLEXIA; RIGIDITY; SPASITICITY; UPPER MOTOR NEURONE SYNDROME]

Hypertropia
Hypertropia is a variety of heterotropia in which there is manifest upward vertical deviation of the visual axis of one eye. Using the cover test, this manifests as downward movement of the uncovered eye. Depending on the affected eye, this finding is often described as a "left-over right" or "right-over left".
[Cross References: COVER TEST, COVER-UNCOVER TEST HYPOTROPIA; HETEROTROPIA]

Hypoaesthesia
Hypoaesthesia (hypaesthesia, hypesthesia) is decreased sensitivity to sensory stimulation of any modality, *e.g.* pain (hypoalgesia), touch.
[Cross References: ANAESTHESIA]

Hypoalgesia
Hypoalgesia is a decreased sensation of pain to a normally painful stimulus.
[Cross References: ANALGESIA]

Hypogeusia
- see AGEUSIA

Hypohidrosis
- see ANHIDROSIS

Hypokinesia
Hypokinesia is a delay in the initiation of voluntary movements, which at worst may progress to an inability to initiate voluntary movement (akinesia). It may often coexist with bradykinesia and hypometria, and is a feature of disorders of the basal ganglia (akinetic-rigid or parkinsonian syndromes), for example:

- Parkinson's disease;
- Multiple system atrophy;
- Steele-Richardson-Olszewski syndrome;
- some variants of prion disease.

[Cross References: AKINESIA; BRADYKINESIA; PARKINSONISM]

Hypometria
Hypometria is a reduction in the amplitude of voluntary movements. It may be demonstrated by asking a patient to make repeated, large amplitude, opposition movements of thumb and forefinger, or tapping movements of the foot on the floor. A gradual decline in amplitude (which may be referred to as fatiguability; *cf.* fatigue) denotes hypometria. Voluntary saccadic eye movements may also show a "step", as a correcting additional saccade compensates for the undershoot (hypometria) of the original movement.

Hypometria is a feature of parkinsonian syndromes such as idiopathic Parkinson's disease.
[Cross References: AKINESIA; BRADYKINESIA; DYSMETRIA; FATIGUE; HYPOKINESIA; PARKINSONISM; SACCADES]

Hypomimia
Hypomimia, or amimia, is a deficit or absence of expression by gesture or mimicry. This is usually most obvious as a lack of facial expressive mobility ("mask-like fades").

This is a feature of frontal-subcortical disease, *e.g.* basal ganglia disease producing akinetic-rigid or parkinsonian syndromes, and frontal lobe lesions (especially of the non-dominant hemisphere).
[Cross References: FACIAL PARESIS; PARKINSONISM]

Hypophonia
Hypophonia is a quiet voice, as in hypokinetic dysarthria. It is often a feature of parkinsonian syndromes (*e.g.* idiopathic Parkinson's disease, multiple system atrophy), and may occur early in Steele-Richardson-Olszewski syndrome. In isolation, other causes of dysphonia may need to be considered.
[Cross References: DYSARTHRIA; DYSPHONIA; PARKINSONISM]

Hypophoria
Hypophoria is a variety of heterophoria in which there is a latent downward deviation of the visual axis of one eye. Using the cover-uncover test, this may be observed clinically as the upward movement of the eye as it is uncovered.
[Cross References: COVER TEST, COVER-UNCOVER TEST; HYPOPHORIA; HETEROPHORIA]

Hyporeflexia
Hyporeflexia is a diminution of tendon reflexes, short of their total absence (areflexia). This may be physiological, as with the diminution of the ankle jerks with normal aging; or pathological, most usually as a feature of a peripheral lesion. Although frequently characterized as a feature of a lower motor neurone syndrome, the pathology underlying hyporeflexia may occur anywhere along the monosynaptic reflex arc, including the sensory afferent fibre and dorsal root ganglion as well as the motor efferent fibre, and/or the spinal cord synapse.

Hyporeflexia may also accompany central lesions, particularly with involvement of the mesencephalic and upper pontine reticular formation. Hyporeflexia is an accompaniment of hemiballismus, and may also be noted in brainstem encephalitis (Bickerstaff's encephalitis), in which presence of a peripheral nerve disorder is debated. Hyporeflexia is not a feature of myasthenia gravis but may occur in Lambert-Eaton myasthenic syndrome (*cf.* facilitation); it is not seen in most muscle diseases unless they are advanced.
[Cross References: AREFLEXIA; FACILITATION; LOWER MOTOR NEURONE SYNDROME; REFLEXES]

Hyposexuality
Hyposexuality is a lack of sexual drive, interest, or activity. It may be associated with many diseases, physical or psychiatric, and/or medications which affect the central nervous system. Along with hypergraphia and hyperreligiosity, hyposexuality is one of the defining features of the Geschwind syndrome.

- Benson DF. The Geschwind syndrome. *Advances in Neurology* 1991; **55**: 411-21
[Cross References: HYPERGRAPHIA; HYPERRELIGIOSITY]

Hypothermia
Hypothalamic damage, particularly in the posterior region, can lead to hypothermia (*cf*. hyperthermia) or poikilothermia (body temperature varying with ambient temperature, as in reptiles). There are many pathological causes:

- tumour
- trauma
- infarct
- haemorrhage
- sarcoidosis
- Wernicke's encephalopathy
- fat embolism
- histiocytosis X
- multiple sclerosis (rare)

A rare syndrome of paroxysmal or periodic hypothermia has been described, and labelled as diencephalic epilepsy. Non-neurological causes of hypothermia are more common, including hypothyroidism, hypopituitarism, hypoglycaemia, and drug overdose.

- Thomas DJ, Green ID. Periodic hypothermia. *BMJ* 1973; **2**: 696-7
[Cross References: HYPERTHERMIA]

Hypotonia, Hypotonus
Hypotonia (hypotonus) is a diminution or loss of normal muscular tone, causing floppiness of the limbs. This is particularly associated with peripheral nerve lesions, as well as lesions of the cerebellum and certain basal ganglia disorders such as hemiballismus-hemichorea.
[Cross References: ATAXIA; FLACCIDITY; HEMIBALLISMUS; HYPERTONIA]

Hypotropia
Hypotropia is a variety of heterotropia in which there is manifest downward vertical deviation of the visual axis of one eye. Using the cover test, this manifests as upward movement of the uncovered eye. Depending on the affected eye, this finding is often described as a "left-over-right" or "right-over-left".
[Cross References: COVER TEST, COVER-UNCOVER TEST HYPERTROPIA; HETEROTROPIA]

I

Ice Pack Test
The ice pack test is performed by holding an ice cube, wrapped in a towel or a surgical glove, over the levator palpebrae superioris muscle of a ptotic eye for 2-10 minutes. Improvement of ptosis is said to be specific for myasthenia gravis: cold improves transmission at the neuromuscular junction (myasthenic patients often improve in cold as opposed to hot weather). This phenomenon is not observed in other causes of ptosis. Whether this observation is also applicable to myasthenic diplopia has yet to be determined.

- Larner AJ, Thomas DJ. Can myasthenia gravis be diagnosed with the "ice pack test"? A cautionary note. *Postgraduate Medical Journal* 2000; **76**: 162-3
[Cross References: FATIGUE]

Ideational Apraxia
- see APRAXIA

Ideomotor Apraxia (IMA)
- see APRAXIA

Illusion
An illusion is a misinterpretation of a perception (*cf.* delusion, hallucination). Illusions occur in normal people when they are tired, inattentive, in conditions of poor illumination, or if there is sensory impairment. They also occur in disease states such as delirium, and psychiatric disorders (affective disorders, schizophrenia).
[Cross References: DELUSION; HALLUCINATION]

Imitation Behaviour
Imitation behaviour is the reproduction by the patient of gestures (echopraxia) and/or utterances (echolalia) made by the examiner in front of the patient; these "echophenomena" are made by the patient without preliminary instructions to do so. They are consistent and have a compulsive quality to them, perhaps triggered by the equivocal nature of the situation. There may be accompanying primitive reflexes, particularly the grasp reflex, and sometimes utilization behaviour.

Imitation behaviour occurs with frontal lobe damage; originally mediobasal disease was thought the anatomical correlate, but more recent studies suggest upper medial and lateral frontal cortex. Certainly imitation behaviour never occurs with retrorolandic cortical lesions.

A distinction has been drawn between "naïve" imitation behaviour, which ceases after direct instruction from examiner not to imitate his/her gestures, which may be seen

in some normal individuals; and "obstinate" imitation behaviour which continues despite instruction to stop; the latter is said to be exclusive to frontotemporal dementia.

- De Renzi E, Cavalleri F, Facchini S. Imitation and utilisation behaviour. *Journal of Neurology, Neurosurgery and Psychiatry* 1996; **61**: 396-400
- Lhermitte F, Pillon B, Serdaru M. Human autonomy and the frontal lobes. Part I: imitation and utilization behaviour: a neuropsychological study of 75 patients. *Annals of Neurology* 1986; **19**: 326-34
- Shimomura T, Mori E. Obstinate imitation behaviour in differentiation of frontotemporal dementia from Alzheimer's disease. *Lancet* 1998; **352**: 623-4
[Cross References: ECHOLALIA; ECHOPRAXIA; GRASP REFLEX; UTILIZATION BEHAVIOUR]

Imitation Synkinesis
- see MIRROR MOVEMENTS

Impersistence
Impersistence is an inability to sustain simple motor acts, such as conjugate gaze, eye closure, protrusion of the tongue, or keeping the mouth open. It is most commonly seen with lesions affecting the right hemisphere, especially central and frontal mesial regions, and may occur in association with left hemiplegia, neglect, anosognosia, hemianopia, and sensory loss. These patients may also manifest perseveration, echolalia and echopraxia.

Impersistence is most often observed following vascular events but may also be seen in Alzheimer's disease and frontal lobe dementias, and metabolic encephalopathies. Impersistence of tongue protrusion and hand grip may be seen in Huntington's disease. Neuropsychologically, impersistence may be related to mechanisms of directed attention which are needed to sustain motor activity.

- Fisher M. Left hemiplegia and motor impersistence. *Journal of Nervous and Mental Disease* 1956; **123**: 201-18
- Kertesz A, Nicholson I, Cancelliere A, Kassa K, Black SE. Motor impersistence: a right-hemisphere syndrome. *Neurology* 1985; **35**: 662-6
[Cross References: ANOSOGNOSIA, ECHOLALIA; ECHOPRAXIA; HEMIANOPIA; MILKMAID'S GRIP; NEGLECT; PERSEVERATION; TROMBONE TONGUE]

Inattention
- see NEGLECT

Incontinence
Urinary incontinence may result from neurological disease. Neurological pathways subserving the appropriate control of micturition encompass the medial frontal lobes, a micturition centre in the dorsal tegmentum of the pons, spinal cord pathways, Onuf's nucleus in the spinal cord segments S2-S4, the cauda equina, and the pudendal nerves. Thus the anatomical differential diagnosis of incontinence is broad. Moreover incontinence may be due to inappropriate bladder emptying or a consequence of loss

of awareness of bladder fullness with secondary overflow. Other features of the history and/or examination may give useful pointers as to localisation. Incontinence of neurological origin is often accompanied by other neurological signs, especially if associated with spinal cord pathology (see Myelopathy). The pontine micturition centre lies close to the medial longitudinal fasciculus and local disease may cause an internuclear ophthalmoplegia. However, other signs may be absent in disease of the frontal lobe or cauda equina.

Causes of urinary incontinence include:

- Idiopathic generalised epilepsy with tonic-clonic seizures; the differential diagnosis of "loss of consciousness with incontinence" also encompasses syncopal attacks with or without secondary anoxic convulsions, non-epileptic attacks, and hyperekplexia;
- Frontal lobe lesions: frontal lobe dementia; normal pressure hydrocephalus;
- Spinal cord pathways: urge incontinence of multiple sclerosis; loss of awareness of bladder fullness with retention of urine and overflow in tabes dorsalis;
- Sacral spinal cord injury; degeneration of the sacral anterior horn cells in Onuf's nucleus (multiple system atrophy);
- Cauda equina syndrome; tethered cord syndrome (associated with spinal dysraphism);
- Pelvic floor injury.

Neurogenic incontinence may be associated with urgency, which results from associated abrupt increases in detrusor pressure (detrusor hyperreflexia); this may be helped by anticholinergic medication (*e.g.* oxybutinin). In addition there may be incomplete bladder emptying, which is usually asymptomatic, due to detrusor sphincter dyssynergia; for post-micturition residual volumes of greater than 100 ml (assessed by in-out catheterisation or ultrasonography), this is best treated by clean intermittent self-catheterisation.

- Fowler CJ. Investigaion of the neurogenic bladder. In: Hughes RAC (ed.). *Neurological Investigations.* London: BMJ Publishing 1997: 397-414

[Cross References: CAUDA EQUINA SYNDROME; DEMENTIA; FRONTAL LOBE SYNDROMES; HYPEREKPLEXIA; INTERNUCLEAR OPHTHALMOPLEGIA; MYELOPATHY; SEIZURES; URINARY RETENTION]

Intermanual Conflict

Intermanual conflict is a behaviour exhibited by an alien hand (*le main étranger*) in which it reaches across involuntarily to interfere with the voluntary activities of the contralateral (normal) hand. This behaviour is more characteristic of the callosal, rather than the frontal type, of alien limb. It is most often encountered in patients with corticobasal degeneration.

[Cross References: ALIEN LIMB]

Internal Ophthalmoplegia

- see OPHTHALMOPARESIS, OPHTHALMOPLEGIA

Internuclear Ophthalmoplegia (INO)

An internuclear Ophthalmoplegia, or medial longitudinal fasciculus syndrome, consists of ipsilateral weakness of eye adduction with contralateral nystagmus of the abducting eye (ataxic or dissociated nystagmus), but with preserved convergence. This may be obvious with pursuit eye movements, but is better seen when testing reflexive saccades or optokinetic responses when the adducting eye is seen to "lag" behind the abducting eye. INO may be asymptomatic or, rarely, may cause diplopia, oscillopsia, or a skew deviation.

INO may be unilateral or bilateral. The commonest cause by far is demyelination, particularly in young patients, but other causes include cerebrovascular disease (particularly older patients), Wernicke-Korsakoff syndrome, encephalitis, trauma, and paraneoplasia.

A similar clinical picture may be observed with more distal pathology, and referred to as a pseudo-internuclear Ophthalmoplegia (*q.v.*), especially in myasthenia gravis.

- Zee DS. Internuclear Ophthalmoplegia: pathophysiology and diagnosis. In: Büttner U, Brandt Th. *Ocular motor disorders of the brain stem.* London: Baillière Tindall, 1992: 455-70

[Cross References: DIPLOPIA; ONE-AND-A-HALF SYNDROME; OPTOKINETIC NYSTAGMUS, OPTOKINETIC RESPONSE; OSCILLOPSIA; PSEUDO-INTERNUCLEAR OPHTHALMOPLEGIA; SACCADES; SKEW DEVIATION]

Intrusion

An intrusion is an inappropriate recurrence of a response (verbal, motor) to a preceding test or procedure, reflecting inattention in dementing disorders or delirium. These phenomena overlap with the recurrent type of perseveration.

The term intrusion is also used to describe inappropriate saccadic eye movements which interfere with macular fixation.

[Cross References: DELIRIUM; DEMENTIA; PERSEVERATION; SACCADIC INTRUSION]

Inverse Marcus Gunn Phenomenon

- see JAW WINKING; PTOSIS

Inverted Reflexes

A phasic tendon stretch reflex is said to be inverted when the movement elicited is opposite to that normally seen, *e.g.* extension of the elbow rather than flexion when eliciting the supinator (brachioradialis) jerk; or flexion (hamstring contraction) rather than extension of the knee when tapping the patellar tendon.

The finding of inverted reflexes may reflect dual pathology, but more usually reflects a single lesion which simultaneously affects a root or roots, interrupting the local reflex arc, and the spinal cord, damaging corticospinal (pyramidal tract) pathways which supply segments below the reflex arc. Hence, an inverted supinator jerk is indicative of a lesion at C5/6, and an inverted knee jerk indicates interruption

of the L2/3/4 reflex arcs, with concurrent damage to pathways descending to levels below these segments.

- Boyle RS, Shakir RA, Weir AI, McInnes A. Inverted knee jerk: a neglected localising sign in spinal cord disease. *Journal of Neurology, Neurosurgery and Psychiatry* 1979; **42**: 1005-7
[Cross References: REFLEXES]

Iridoplegia
- see OPHTHALMOPARESIS, OPHTHALMOPLEGIA

J

Jacksonian March
Jacksonian march is the sequential spread of a simple partial seizure to involve other body parts, for example jerking may spread from one hand up the arm, to the ipsilateral side of the face. It may culminate in a secondary generalised seizure. The pathophysiological implication is of electrical disturbance spreading through the homunculus of the motor cortex. A sensory equivalent is rare.
[Cross References: SEIZURES]

Jactitation
Jactitation is literally "throwing about", implying restlessness. Although the term is sometimes used to refer to jerking or convulsion of epileptic origin, it is essentially non-specific and may also refer to the restlessness seen in acute illness, high fever, and exhaustion. It differs from the restlessness implied by akathisia.
[Cross References: AKATHISIA; SEIZURES]

Jamais Vu
Jamais vu is a complex aura of focal onset epilepsy in which there is a sensation of strangeness about visual stimuli that are in fact familiar, as though they had never been seen before (*cf. déjà vu*). This is suggestive of seizure onset in the limbic system, but is not lateralising (*cf. déjà vu*).
[Cross References: AURA; *DEJÀ VU*]

Jargon Aphasia
Jargon aphasia is a fluent aphasia characterized by a jumbled, unintelligible and meaningless output, with multiple paraphasias and neologisms, and sometimes echolalia (as in transcortical sensory aphasia). There may be a pressure of speech (logorrhoea).

There is debate as to whether jargon aphasia is simply a primary Wernicke/posterior/ sensory type of aphasia with failure to self-monitor speech output, or whether additional deficits (*e.g.* pure word deafness, intellectual impairment) are also required. Others suggest that jargon aphasia represents aphasia and anosognosia, leading to confabulation and reduplicative paramnesia.
[Cross References: ANOSOGNOSIA; APHASIA; CONFABULATION; ECHOLALIA; LOGORRHOEA; PURE WORD DEAFNESS; REDUPLICATIVE PARAMNESIA; TRANSCORTICAL APHASIA; WERNICKE'S APHASIA]

Jaw Jerk
The jaw jerk, or masseter reflex, is contraction of the masseter and temporalis muscles in response to a tap on the jaw with the mouth held slightly open. Both the afferent and efferent limbs of the arc run in the mandibular division of the trigeminal (V) nerve, connecting centrally with the mesencephalic nucleus of the trigeminal

nerve. The reflex is highly reproducible; there is a linear correlation between age and reflex latency, and a negative correlation between age and reflex amplitude.

Interruption of the reflex arc leads to a diminished or absent jaw jerk as in bulbar palsy (although an absent jaw jerk may be a normal finding, particularly in the elderly). Bilateral supranuclear lesions cause a brisk jaw jerk, as in pseudobulbar palsy.

- Fitzek S, Fitzek C, Hopf HC. Normative values of the masseter reflex (myotatic masseter reflex). *Journal of Neurology* 2000; **247 (suppl3)**: 176-7 (abstract 724)
[Cross References: BULBAR PALSY; PSEUDOBULBAR PALSY; REFLEXES]

Jaw Winking

Jaw winking, also known as the Marcus Gunn phenomenon, is a widening of a congenital ptosis when a patient is chewing, swallowing, or opening the jaw (*i.e.* trigemino-oculomotor synkinesis). It is believed to result from aberrant innervation of the pterygoid muscles and levator palpebrae.

Eyelid closure on opening of the jaw (inverse Marcus Gunn phenomenon) is also described, as the Marin-Amat syndrome, thought to be due to aberrant facial (VII) nerve regeneration.

- Rana PVS, Wadia RS. The Marin-Amat syndrome: an unusual facial synkinesia. *Journal of Neurology, Neurosurgery and Psychiatry* 1985; **48**: 939-41
[Cross Reference: PTOSIS; SYNKINESIS]

Jendrassik's Manoeuvre

Jendrassik's manoeuvre is used to enhance or bring out absent or depressed tendon reflexes by contraction of distant muscle groups, *e.g.* clenching teeth, or making a fist, interlocking fingers and pulling the hands against one another. If previously absent reflexes are then elicited, this may be denoted +/-. Co-contraction increases the gain in the monosynaptic reflex arc, as distinct from facilitation or post-tetanic potentiation which is seen in Lambert-Eaton myasthenic syndrome following tetanic contraction of muscles involved in the reflex..

- Jendrassik E. Ueber allgemeine Localisation der Reflexe. *Deutshce Archiv fur Klinische Medicin* 1894; **52**: 569-600
[Cross Reference: FACILITATION; REFLEXES]

Joint Position Sense

- see PROPRIOCEPTION

Jugular Foramen Syndrome

The glossopharyngeal (IX), vagus (X), and accessory (XI) cranial nerves may be damaged by lesions at or around the jugular foramen, producing a jugular foramen syndrome (or Vernet's syndrome). This produces:

- Ipsilateral weakness and atrophy of sternocleidomastoid and trapezius due to accessory nerve involvement (atrophy may be the more evident, hence the importance of feeling the muscle bellies);

- Dysphagia, dysphonia, palatal droop, impaired gag reflex; ipsilateral reduced taste sensation on the posterior one third of the tongue, and anaesthesia of the posterior one third of the tongue, soft palate, pharynx, larynx and uvula, due to glossopharyngeal and vagus nerve involvement.

Causes of the jugular foramen syndrome include:

- Skull base trauma/fracture;
- Glomus jugulare tumour;
- Inflammatory/infective collection at the skull base
- Ischaemia.

The differential diagnosis includes retropharyngeal or retroparotid space occupying lesions, which may in addition involve the hypoglossal nerve (XII; Collet-Sicard syndrome) and the sympathetic chain with or without the facial nerve (VII; Villaret's syndrome).
[Cross Reference: DYSPHAGIA; DYSPHONIA; GAG REFLEX]

Junctional Scotoma
Also known as ***Junctional Scotoma of Traquair***
- see SCOTOMA

K

Kayser-Fleischer Rings
Kayser-Fleischer rings are deposits of copper, seen as a brownish discoloration, in Descemet's membrane. Although often visible to the naked eye (difficult in people with a brown iris), they are best seen with a slit-lamp examination. Since they are a highly reliable sign of intracerebral copper deposition in Wilson's disease (hepatolenticular degeneration) everyone suspected of this diagnosis (*i.e.* with parkinsonism or dystonia presenting before age 50) should have a slit-lamp examination (as well as copper and caeruloplasmin measured). Very occasionally cases of neurological Wilson's disease without Kayser-Fleischer rings have been reported.

- Finelli PF. Kayser-Fleischer ring: Hepatolenticular degeneration (Wilson's disease). *Neurology* 1995; **45**: 1261-2
[Cross Reference: DYSTONIA; PARKINSONISM]

Kernig's Sign
Kernig's sign is pain in the lower back (and also sometimes the neck) and resistance to movement with passive extension of the knee on the flexed thigh in a recumbent patient. It is indicative of meningeal mechanosensitivity due to inflammation, either infective (meningitis) or chemical (subarachnoid haemorrhage), in which case it may coexist with nuchal rigidity and Brudzinski's neck sign. If unilateral it may indicate irritation of the lumbosacral nerve roots from a ruptured intervertebral disc (in which case Lasègue's sign may also be present).
[Cross Reference: BRUDZINSKI'S (NECK) SIGN; LASÈGUE'S SIGN; NUCHAL RIGIDITY]

Kinesis Paradoxica
Kinesis paradoxica is the brief but remarkably rapid and effective movement sometimes observed in patients with Parkinson's disease or post-encephalitic parkinsonism, despite the poverty and slowness of spontaneous movement (akinesia, hypokinesia; bradykinesia) seen in these conditions. It often occurs in response to alarm, excitement or emotion (*e.g.* in response to a genuinely funny joke).
[Cross References: AKINESIA; BRADYKINESIA; HYPOKINESIA; PARKINSONISM]

Klazomania
Klazomania was the term applied to the motor and vocal tics seen in encephalitis lethargica (von Economo's disease), an observation which helped to promote the idea that such tics were of neurological, rather than psychiatric, origin in other situations as well, such as Gilles de la Tourette syndrome.
[Cross References: PARAKINESIA; TIC]

Klüver-Bucy Syndrome
The Klüver-Bucy syndrome consists of a variety of neurobehavioural changes, originally observed following bilateral temporal lobectomy (especially anterior tip) in monkeys, but subsequently described in man. The characteristic features, some or all of which may be present, are:

- visual agnosia (*e.g.* misrecognition of others);
- hyperorality;
- hyperphagia, binge eating;
- hypermetamorphosis;
- hypersexuality;
- emotional changes: apathy; loss of fear, rage reactions.

Causes described in man include:

- following bilateral temporal lobectomy;
- Pick's disease;
- Alzheimer's disease: especially hyperorality and hyperphagia, but it is rare to have all features;
- post-ictal, in a patient with a previous unilateral temporal lobectomy.

- Anson JA, Kuhlman DT. Post-ictal Klüver-Bucy syndrome after temporal lobectomy. *Journal of Neurology, Neurosurgery and Psychiatry* 1993; **56**: 311-3
- Klüver H, Bucy P. Preliminary analysis of functions of the temporal lobes in monkeys. *Archives of Neurology and Psychiatry* 1939; **42**: 979-1000.
[Cross References: APATHY; HYPERMETAMORPHOSIS; HYPERORALITY; HYPERPHAGIA; HYPERSEXUALITY; VISUAL AGNOSIA]

Knee Tremor
A characteristic tremor of the patellae, sometimes known as knee bobbing, juddering, or quivering, is seen in the condition of primary orthostatic tremor (POT). It is due to rapid rhythmic contractions of the leg muscles on standing; these dampen or subside on walking, leaning against a wall, or being lifted off the ground, with disappearance of the knee tremor. Auscultation with the diaphragm of a stethoscope over the lower limb muscles reveals a regular thumping sound, likened to the sound of a distant helicopter. EMG studies show pathognomonic synchronous activity in the leg muscles with a frequency of 14-18Hz, thought to be generated by a central oscillator (peripheral loading does not alter tremor frequency).
A number of drugs may be helpful in POT, including phenobarbitone, primidone, clonazepam, and levodopa, but not propranolol (*cf.* essential tremor).

- Heilman KM. Orthostatic tremor. *Archives of Neurology* 1984; **41**: 880-1
- Brown P. New clinical sign for orthostatic tremor. *Lancet* 1995; **346**: 306-7
[Cross References: TREMOR]

Körber-Salus-Elschnig Syndrome
- see NYSTAGMUS

Kyphoscoliosis

Kyphoscoliosis is a twisting of the spine, in both the anteroposterior (kyphosis) and lateral (scoliosis) planes. Although such deformity is often primarily orthopaedic in origin, it may also be a consequence of neurological disease which causes weakness of paraspinal muscles. Recognised neurological causes include Friedreich's ataxia and neurofibromatosis. Stiff person syndrome may produce a characteristic hyperlordotic spine. Some degree of scoliosis occurs in virtually patients suffering from paralytic poliomyelitis before the pubertal growth spurt.

[Cross References: CAMPTOCORMIA; STIFFNESS]

L

Lagophthalmos
Lagophthalmos is an inability to close the eyelid in a peripheral facial (VII) nerve paralysis producing a widening of the palpebral fissure.
[Cross References: BELL'S PALSY; FACIAL PARESIS]

Lasègue's Sign
Lasègue's sign is pain along the course of the sciatic nerve induced by stretching of the nerve, achieved by flexing the thigh at the hip while the leg is extended at the knee ("straight leg raising"). This is similar to the manoeuvre used in Kernig's sign (gradual extension of knee with thigh flexed at hip). Both indicate irritation of the lower lumbosacral nerve roots and/or meninges. Various modifications of Lasègue's sign have been described.
[Cross References: KERNIG'S SIGN]

Lateral Medullary Syndrome
The lateral medullary syndrome (or Wallenberg's syndrome, after the neurologist who described it in 1895) results from damage (usually infarction) of the posterolateral medulla with or without involvement of the inferior cerebellum, producing the following clinical features:

- Nausea, vomiting, vertigo, oscillopsia (involvement of vestibular nuclei);
- Contralateral hypoalgesia, thermoanaesthesia (spinothalamic tract);
- Ipsilateral facial hypoalgesia, thermoanaesthesia, + facial pain (trigeminal spinal nucleus and tract);
- Horner's syndrome (descending sympathetic tract), +/- ipsilateral hypohidrosis of the body;
- Ipsilateral ataxia of limbs (olivocerebellar/spinocerebellar fibres, inferior cerebellum);
- Dysphagia, dysphonia, impaired gag reflex;
- +/- eye movement disorders, including nystagmus, abnormalities of ocular alignment (skew deviation, ocular tilt reaction, environmental tilt), smooth pursuit and gaze holding, and saccades;
- +/- hiccup.

Infarction due to vertebral artery occlusion (occasionally posterior inferior cerebellar artery) or dissection is the commonest cause of lateral medullary syndrome, although tumour, demyelination, and trauma are also recognised causes.

- Fisher CM, Karnes W, Kubik C. Lateral medullary infarction: the pattern of vascular occlusion. *Journal of Neuropathology and Experimental Neurology* 1961; **20**: 103-13
- Pearce JMS. Wallenberg's syndrome. *Journal of Neurology, Neurosurgery and Psychiatry* 2000; **68**: 570

- Sacco RL, Freddo L, Bello JA, Odel JG, Onesti ST, Mohr JP. Wallenberg's lateral medullary syndrome. Clinical-magnetic resonance imaging correlations. *Archives of Neurology* 1993; **50**: 609-14
[Cross References: ANAESTHESIA; DYSPHAGIA; DYSPHONIA; ENVIRONMENTAL TILT; GAG REFLEX; HEMIATAXIA; HICCUP; HORNER'S SYNDROME; HYPOALGESIA; HYPOHIDROSIS; MEDIAL MEDULLARY SYNDROME; NYSTAGMUS; OCULAR TILT REACTION; OSCILLOPSIA; SACCADES; SKEW DEVIATION; VERTIGO]

Lateral Rectus Palsy
- see ABDUCENT NERVE PALSY

Laterocollis
Laterocollis is a lateral head tilt; this may be seen in 10-15% of patients with torticollis.
[Cross References: TORTICOLLIS]

Laughter
- see AUTOMATISM; PATHOLOGICAL LAUGHTER

Leadpipe Rigidity
- see RIGIDITY

Levator Inhibition
- see EYELID APRAXIA

Lhermitte's Sign
Lhermitte's sign, or the "barber's chair syndrome", is a painless but unpleasant tingling or electric shock-like sensation spreading instantaneously down the arms and legs following neck flexion (active or passive). It is associated with pathology within the cervical spinal cord. Although most commonly encountered (and originally described in) demyelination, it is not pathognomonic of this condition, and has been described with other local pathologies such as:

- subacute combined degeneration of the cord (vitamin B_{12} deficiency)
- traumatic or compressive cervical myelopathy (*e.g.* cervical spondylosis)
- epidural/subdural/intraparenchymal tumour
- radiation myelitis
- pyridoxine toxicity
- inflammation, *e.g.* systemic lupus erythematosus, Behçet's disease
- cervical herpes zoster myelitis
- cavernous angioma of the cervical cord

Pathophysiologically, this movement-induced symptom may reflect the exquisite mechanosensitivity of axons which are demyelinated, or damaged in some other way. A "motor equivalent" of Lhermitte's sign, McArdle's sign, has been described.

- Lhermitte J, Bollack J, Nicolas M. Les douleurs à type de décharge electrique consécutives à la flexion cèphalique dans la sclérose en plaques: un case de forme sensitive de la sclérose multiple. *Revue Neurologique* 1924; **39:** 56-62
- Smith KJ. Conduction properties of central demyelinated axons: the generation of symptoms in demyelinating disease. In: Bostock H, Kirkwood PA, Pullen AH (eds.). *The neurobiology of disease: contributions from neuroscience to clinical neurology.* Cambridge, CUP 1996: 95-117
[Cross References: McARDLE'S SIGN; MYELOPATHY]

Lid Lag
Lid lag is present if a band of sclera is visible between the upper eyelid and the corneal limbus on attempted downgaze (*cf.* lid retraction), seen for example in thyroid eye disease (von Graefe's sign), Steele-Richardson-Olszewski syndrome, and Guillain-Barré syndrome.
[Cross References: LID RETRACTION; VON GRAEFE'S SIGN]

Lid Retraction
Lid retraction is present if a band of sclera is visible between the upper eyelid and the corneal limbus in the primary position (*cf.* lid lag). This should be distinguished from contralateral ptosis. Recognised causes of lid retraction include:

- Overactivity of levator palpebrae superioris: dorsal mesencephalic lesion (Collier's sign), ptosis of contralateral lid, paradoxical lid retraction with jaw movement (jaw winking, Marcus Gunn phenomenon);
- Overactivity of Müller's muscle: irritative oculosympathetic lesions (Claude-Bernard syndrome);
- Contracture of the levator muscle: thyroid eye disease (Dalrymple's sign), myotonic syndromes, aberrant oculomotor (III) nerve regeneration (pseudo-von Graefe's sign);
- Cicatricial retraction of the lid, *e.g* following trauma.

[Cross References: COLLIER'S SIGN; CONTRACTURE; DALRYMPLE'S SIGN; JAW WINKING; LID LAG; PSEUDO-VON GRAEFE'S SIGN; PTOSIS; STELLWAG'S SIGN; SUNSET SIGN]

Light-Near Pupillary Dissociation
Light-near pupillary dissociation refers to the loss of pupillary light reflexes, whilst the convergence-accommodation reaction is preserved (see Pupillary Reflexes). This dissociation may be seen in a variety of clinical circumstances:

- Argyll Robertson pupil: small irregular pupils with reduced reaction to light, typically seen in neurosyphilis; the absence of miosis and/or pupillary irregularity has been referred to as pseudo-Argyll Robertson pupil, which may occur with sarcoidosis, diabetes, and aberrant regeneration of the oculomotor (III) nerve;
- Holmes-Adie pupil: dilated pupil showing strong but slow reaction to accommodation but minimal reaction to light (tonic > phasic);

- Parinaud's syndrome (dorsal rostral midbrain syndrome): due to a lesion at the level of the posterior commissure, and characterized by vertical gaze palsy, lid retraction (Collier's sign) or ptosis, and large regular pupils responding to accommodation but not light.

[Cross References: ARGYLL ROBERTSON PUPIL; COLLIER"S SIGN; HOLMES-ADIE PUPIL, HOLMES-ADIE SYNDROME; LID RETRACTION; PARINAUD'S SYNDROME; PSEUDO-ARGYLL ROBERTSON PUPIL; PUPILLARY REFLEXES]

Light Reflex
- see PUPILLARY REFLEXES

Locked-In Syndrome
The locked-in syndrome results from de-efferentation, such that a patient is awake, self-ventilating and alert, but unable to speak or move; vertical eye movements and blinking are usually preserved, affording a channel for simple (yes/no) communication.

The most common cause of the locked-in syndrome is basilar artery thrombosis causing ventral pontine infarction (both pathological laughter and pathological crying have on occasion been reported to herald this event). Other pathologies include pontine haemorrhage and central pontine myelinolysis. Bilateral ventral midbrain and internal capsule infarcts can produce a similar picture.

The locked-in syndrome may be mistaken for abulia, akinetic mutism, coma, and catatonia.

- Feldman MH. Physiological observations in a chronic case of locked in syndrome. *Neurology* 1971; **21**: 459-78

[Cross References: ABULIA; AKINETIC MUTISM; BLINKING; COMA; CATATONIA; PATHOLOGICAL CRYING, PATHOLOGICAL LAUGHTER]

Lockjaw
- see TRISMUS

Logoclonia
Logoclonia is the tendency for a patient to repeat the final syllable of a word when speaking; hence it is one of the reiterative speech disorders (*cf.* palilalia, echolalia); it may be described as the festinating repetition of individual phonemes. Logoclonia is an indicator of bilateral brain injury, usually involving subcortical structures, and may be seen in late stages of dementia of Alzheimer type (but not in semantic dementia).
[Cross References: ECHOLALIA; FESTINATION, FESTINANT GAIT; PALILALIA]

Logopenia
Logopenia is a reduced rate of language production, due especially to word finding pauses, but with preserved phrase length and syntactically complete language, seen in aphasic syndromes.
[Cross References: APHASIA]

Logorrhoea
Logorrhoea is literally a flow of speech, or pressure of speech, denoting an excessive verbal output; content is often irrelevant, disconnected and difficult to interpret. Superficially this resembles the output in the Wernicke/posterior type of aphasia but syntax and morphology are intact, rhythm and articulation is usually normal, and paraphasias and neologisms are few. Moreover comprehension is better than anticipated in a Wernicke type of aphasia. Patients may be unaware of their impaired output (anosognosia) due to a failure of self-monitoring.

Logorrhoea may be observed in subcortical (thalamic) aphasia, usually following recovery from lesions (usually haemorrhage) to the anterolateral nuclei. Similar speech output may be observed in psychiatric disorders such as mania and schizophrenia.

- Damasio AR. Aphasia. *New England Journal of Medicine* 1992; **326**: 531-9
[Cross References: APHASIA; DELIRIUM; ECHOLALIA; JARGON APHASIA; WERNICKE'S APHASIA]

Long Tract Signs
- see UPPER MOTOR NEURONE SYNDROME

Lower Motor Neurone Syndrome
A lower motor neurone (LMN) syndrome constitutes a constellation of motor signs resulting from damage to lower motor neurone pathways, *i.e.* from anterior horn cell distally, encompassing the motor roots, nerve plexuses, peripheral nerves, and neuromuscular junction. Following the standard order of neurological examination of the motor system, the signs include:

- APPEARANCE - muscle wasting; fasciculations or fibrillations may be observed or induced, particularly if the pathology is at the level of the anterior horn cell;
- TONE - reduced tone (flaccidity, hypotonus);
- POWER - weakness, often marked; depending on the precise pathological process, weakness often affects both flexor and extensor muscles equally (although this is not always the case);
- CO-ORDINATION - depending on the degree of weakness, it may not be possible to comment on the integrity or otherwise of co-ordination in LMN syndromes; in a pure LMN syndrome co-ordination will be normal;
- REFLEXES - depressed (hyporeflexia) or absent (areflexia); plantar responses are flexor.

It is often possible to draw a clinical distinction between motor symptoms resulting from lower or upper motor neurone pathology and hence to formulate a differential diagnosis and direct investigations accordingly. Sensory features may also be present in LMN syndromes if the pathology affects sensory as well as motor roots, or both motor and sensory fibres in peripheral nerves.
[Cross References: AREFLEXIA; FASCICULATION; FIBRILLATION; FLACCIDITY; HYPOREFLEXIA; HYPOTONIA, HYPOTONUS; NEUROPATHY; UPPER MOTOR NEURONE SYNDROME; WEAKNESS]

M

Macrographia
Macrographia is abnormally large handwriting. It may be seen in cerebellar disease, possibly as a reflection of the kinetic tremor and/or the impaired checking response seen therein (*cf.* micrographia).
[Cross References: MICROGRAPHIA; TREMOR]

Macropsia
- see METAMORPHOPSIA

Macro-Square-Wave Jerks
- see SQUARE-WAVE JERKS

Macula Sparing, Macula Splitting
Macula sparing is a feature of an homonymous hemianopia in which central vision is intact, due to damage confined to the occipital cortex without involving the occipital pole. This may occur because anastomoses between the middle and posterior cerebral arteries maintain that part of area 17 necessary for central vision after occlusion of the posterior cerebral artery.

Cortical blindness due to bilateral (sequential or simultaneous) posterior cerebral artery occlusion may leave a small central field around the fixation point intact, also known as macula sparing.

Macula splitting, an homonymous hemianopia which cuts through the vertical meridian of the macula, occurs with lesions of the optic radiation.
[Cross References: CORTICAL BLINDNESS; HEMIANOPIA]

Maculopathy
Maculopathy is any process affecting the macula, with changes observable on ophthalmoscopy. These processes may produce a central or ring scotoma and visual failure. Common causes include:

- Diabetes mellitus: Oedema and hard exudates at the macula are a common cause of visual impairment, especially in non-insulin dependent diabetes mellitus.
- Hypertension: abnormal vascular permeability around the fovea may produce a macular star.
- Drug-induced: *e.g.* "bull's-eye" maculopathy of chloroquine
- "Cherry red spot at the macula": this appearnce may occur in sialidosis ("cherry red spot-myoclonus syndrome") and gangliosidoses (*e.g.* Tay-Sachs disease).

[Cross References: RETINOPATHY]

Magnetic Movements
Movements may be described as magnetic in varying contexts:

- the following or tracking movements of an alien hand in corticobasal degeneration, reaching out to touch or grasp the examiner's hand;
- a hesitant gait (ignition failure), with seeming inability to lift the feet ("stuck to the floor") in gait apraxia.

[Cross References: ALIEN LIMB; FORCED GROPING; GAIT APRAXIA; GRASP REFLEX]

Main d'accoucheur

Main d'accoucheur, or carpopedal spasm, is a posture of the hand with wrist flexion in which the muscles are rigid and painful. This tetanic posture may develop in acute hypocalcaemia (induced by hyperventilation, for instance) or hypomagnesaemia, reflecting muscle hyperexcitability. Development of *main d'accoucheur* within 4 minutes of inflation of a sphygmomanometer cuff (Trousseau's sign) indicates latent tetany. Mechanosensitivity of nerves may also be present elsewhere (Chvostek's sign).

Main d'accoucheur is so called because of its resemblance to the posture of the hand adopted for the manual delivery of a baby ("obstetrical hand").

[Cross References: CHVOSTEK'S SIGN]

Main en griffe

- see CLAW HAND

Main étranger

- see ALIEN LIMB

"Man-in-a-Barrel"

"Man-in-a-barrel" is a clinical syndrome of brachial diplegia with preserved muscle strength in the legs. This most usually occurs as a result of bilateral borderzone infarcts in the territories between the anterior and middle cerebral arteries ("watershed infarction"). This may be as a consequence of cerebral hypoperfusion (*e.g.* during cardiac arrest, cardiac surgery), in which case the prognosis is poor. The clinical picture has also been reported with cerebral metastases. Acute central cervical cord lesions may also produce a "man-in-a-barrel" syndrome, for example after severe hyperextension injury, or after unilateral vertebral artery dissection causing anterior cervical spinal cord infarction. This may follow a transient quadriplegia, and considerable recovery is possible.

A neurogenic main-in-a-barrel syndrome has been reported ("flail arm syndrome"), which is a variant of motor neurone disease.

- Mohr JP. Distal field infarction. *Neurology* 1969; **19**: 279 (abstract GS7)

[Cross References: FLAIL ARM; QUADRIPARESIS, QUADRIPLEGIA]

Marche à petit pas

Marche à petit pas is a disorder of gait characterized by impairments of balance, gait ignition, and locomotion, particularly there is shortened stride (literally *marche à petit pas*) and a variably wide base. This gait disorder is often associated with dementia, frontal release signs, and urinary incontinence, and sometimes with apraxia,

parkinsonism, and pyramidal signs. This constellation of clinical signs reflects underlying pathology in the frontal lobe and subjacent white matter, most usually of vascular origin. Modern clinical classifications of gait disorders have subsumed *marche à petit pas* into the category of frontal gait disorder.

- Nutt JG, Marsden CD, Thompson PD. Human walking and higher-level gait disorders, particularly in the elderly. *Neurology* 1993; **43:** 268-79
[Cross References: APRAXIA; DEMENTIA; FRONTAL RELEASE SIGNS; PARKINSONISM]

Marcus Gunn Phenomenon
- see JAW WINKING

Marcus Gunn Pupil, Marcus Gunn Sign
The Marcus Gunn pupil or sign is the adaptation of the pupillary light reflex to persistent stimulation, that is, a dilatation of the pupil is observed with continuing stimulation with incident light. This is indicative of an afferent pathway defect, such as retrobulbar neuritis. A modification of this observation, the swinging flashlight sign or test (originally described by Levitan), compares the direct and consensual pupillary light reflexes in one eye; the speed of swing is found by trial and error. Normally the responses are equal but in the presence of an afferent conduction defect an inequality is manifest as pupillary dilatation. The test is known to be unreliable in the presence of bilateral afferent defects of light conduction.

- Pearce JMS. The Marcus Gunn pupil. *Journal of Neurology, Neurosurgery and Psychiatry* 1996; **61:** 520
- Thompson HS, Corbett JJ. Swinging flashlight test. *Neurology* 1989; **39:** 154-6
[Cross References: AFFERENT PUPILLARY DEFECT; PUPILLARY REFLEXES]

Mask-like Facies
The poverty of spontaneous facial expression seen in extrapyramidal disorders such as idiopathic Parkinson's disease is sometimes described as mask-like; hypomimia is the appropriate technical term.
[Cross References: HYPOMIMIA; PARKINSONISM]

Masseter Reflex
- see JAW JERK

McArdle's Sign
McArdle's sign is a reduction in lower limb strength, increased lower limb stiffness and impaired mobility following neck flexion. The difference may best be appreciated by comparing leg strength in full neck extension and full neck flexion. The sign was initially described in multiple sclerosis but may occur in other myelopathic conditions, present at any point between the foramen magnum and the lower thoracic cord. The mechanism is presumed to be stretch-induced conduction block in demyelinated plaques in the corticospinal tracts. McArdle's sign may be envisaged as the motor equivalent of Lhermitte's sign.

- O'Neill JH, Mills KR, Murray NMF. McArdle's sign in multiple sclerosis. *Journal of Neurology, Neurosurgery and Psychiatry* 1987; **50:** 1691-3
- McArdle MJ. McArdle's sign in multiple sclerosis. *Journal of Neurology, Neurosurgery and Psychiatry* 1988; **51:** 1110
[Cross References: LHERMITTE'S SIGN; MYELOPATHY]

Medial Medullary Syndrome
The medial medullary syndrome, or Dejerine's anterior bulbar syndrome, results from damage to the medial medulla, most usually infarction as a consequence of anterior spinal artery or vertebral artery occlusion. The clinical picture is of:

- Ipsilateral tongue paresis and atrophy, fasciculations (hypoglossal nerve involvement);
- Contralateral hemiplegia with sparing of the face (pyramid);
- Contralateral loss of position and vibration sense (medial lemniscus) with pain and temperature sensation spared;
- +/- upbeat nystagmus (?nucleus intercalatus of Staderini).

- Hirose G, Ogasawara T, Shirakawa T, *et al.* Primary position upbeat nystagmus due to unilateral medial medullary infarction. *Annals of Neurology* 1998; **43:** 403-6
- Sawada H, Seriu N, Udaka F, Kameyama M. Magnetic resonance imaging of medial medullary infarction. *Stroke* 1990; **21:** 963-6
[Cross References: HEMIPLEGIA; FASCICULATION; LATERAL MEDULLARY SYNDROME; NYSTAGMUS]

Menace Reflex
- see BLINK REFLEX

Meningism
Meningism (meningismus, nuchal rigidity) is a stiffness or discomfort on passive movement (especially flexion) of the neck in the presence of meningeal irritation (*e.g.* infective meningitis, subarachnoid haemorrhage). A number of other, eponymous, signs of meningeal irritation have been described, of which the best known are those of Kernig and Brudzinski.

Meningism is not synonymous with meningitis, since it may occur in acute systemic pyrexial illnesses (pneumonia, bronchitis), especially in children. Moreover, meningism may be absent despite the presence of meningitis in the elderly and those receiving immunosuppression.
[Cross References: BRUDZINSKI'S (NECK) SIGN; KERNIG'S SIGN; NUCHAL RIGIDITY]

Metamorphopsia
Metamorphopsia is an illusory visual phenomenon characterized by objects appearing distorted or misshapen in form: these may include distortions of personal body image, spatial relationships, and size (objects being smaller [micropsia] or larger [macropsia] than normal): the "Alice in Wonderland syndrome". Metamorphopsias are most often transient and episodic, as for example during migraine, seizures, psychotropic drug

abuse, and petechial intraparenchymal haemorrhages; or, rarely, more long-lasting or permanent, following brain infarction (most commonly involving the occipito-parietal or temporo-parietal cortex: lesions on the right are more likely than those on the left to give metamorphopsia). Retinal disease causing displacement of photoreceptors may produce metamorphopsia, *e.g.* micropsia due to receptor separation in retinal oedema, macropsia due to receptor approximation in retinal scarring. Occasional cases of metamorphopsia have been reported with lesions of the optic chiasm, optic radiation, and retrosplenial region. Indeed, it seems that metamorphopsia may occur with pathology at any point along the visual pathway from retina to cortex, with differing patterns:

- retinal lesions - ipsilateral monocular;
- chiasmal lesions - bitemporal;
- occipitoparietal lesions - contralateral homonymous.

Metamorphopsia may be associated with visual hallucinations.

- Shiga K, Makino M, Ueda Y, Nakajima K. Metamorphopsia and visual hallucinations restricted to the right visual hemifield after a left putaminal haemorrhage. *Journal of Neurology Neurosurgery and Psychiatry* 1996; **61:** 420-1
[Cross References: HALLUCINATION; ILLUSION; MICROPSIA]

Micrographia

Micrographia is small handwriting. It is most often recognised in association with the extrapyramidal features of idiopathic Parkinson's disease (indeed it may be the presenting sign), but may occasionally occur with other parkinsonian syndromes (*e.g.* Steele-Richardson-Olszewski syndrome) or in isolation with focal lesions of the midbrain or basal ganglia.

A distinction may be drawn between "slow" micrographia, in which letters become progressively smaller as writing proceeds, as in Parkinson's disease (this may reflect fatigue, a gradual decline in the amplitude and speed of initiation of voluntary movements); and "fast" micrographia, in which letters are small throughout (*e.g.* Steele-Richardson-Olszewski syndrome) in which fatigue is not evident.

There is a poor correlation between micrographia and the side, severity or duration of classical parkinsonian features, and its response to levodopa preparations is very variable. These observations, along with reports of isolated micrographia with cortical lesions demonstrated by neuroimaging, suggest that the anatomical basis of micrographia may be at the level of the cortex (dominant parietal lobe) rather than the basal ganglia.

- McLennan JE, Nakano K, Tyler HR, Schwab RS. Micrographia in Parkinson's disease. *Journal of the Neurological Sciences* 1972; **15:** 141-52
- Scolding NJ, Lees AJ. Micrographia associated with a parietal lobe lesion in multiple sclerosis. *Journal of Neurology, Neurosurgery and Psychiatry* 1994; **57:** 739-41
[Cross References: FATIGUE; PARKINSONISM]

Micropsia
Micropsia, or "Lilliput sight", is the underestimation of the size of a normally recognized object. It is the commonest form of metamorphopsia, and often occurs with lesions of the right temporo-parietal cortex; retinal oedema may also cause micropsia. The entirely subjective nature of the disorder may account for the relative rarity of reports.

- Ceriani F, Gentileschi V, Muggia S, Spinnler H. Seeing objects smaller than they are: micropsia following right temporo-parietal infarction. *Cortex* 1998; **34**: 131-8
[Cross References: METAMORPHOPSIA]

Milkmaid's Grip
Milkmaid's grip is the descriptive term applied to the inability to maintain a firm grip (*e.g.* of the examiner's fingers), detected as an alternating squeezing and releasing (as required for successful milking by hand). Seen in Huntington's disease, this may reflect a combintion of chorea and motor impersistence.
[Cross References: CHOREA; IMPERSISTENCE; TROMBONE TONGUE]

Miosis
Miosis is an abnormally small pupil. Causes include:

- Oculosympathetic paresis of whatever cause, *e.g.* Horner's syndrome, pontine haemorrhage;
- Drug-induced: *e.g.* opiates.

If only one pupil appears small (anisocoria), it is important to distinguish miosis from contralateral mydriasis, when a different differential will apply.
[Cross References: ANISOCORIA; ARGYLL ROBERTSON PUPIL; HORNER'S SYNDROME; MYDRIASIS]

Mirror Movements
Mirror movements are involuntary movements of one side of the body that accompany and "mirror" (reflect) intentional movements on the opposite side (this is also known as imitation synkinesis). They are usually symmetrical and most often seen when using distal muscles of the upper limb. Mirror movements are frequently present in young children but prevalence decreases with age. Persistence of mirror movements into adult life ("congenital mirror movements") is pathological, as is acquisition in adult life. These movements are uncommon after acquired brain lesions with no relationship to specific anatomical areas.

Congenital mirror movements are associated with skeletal developmental abnormalities, especially of the atlanto-occipital region, such as Klippel-Feil syndrome. They are also seen in 85% of patients with X-linked Kallmann syndrome (hypogonadotrophic hypogonadism and anosmia). Acquired mirror movements have been described following thalamic lesions, and in association with spastic paraparesis, extrapyramidal disorders, Friedriech's ataxia, and phenylketonuria.

There is some neurophysiological evidence from patients with X-linked Kallmann syndrome for the existence of an ipsilateral corticospinal pathway, consistent with other evidence that this condition is primarily a disorder of axonal guidance during

development. Concurrent activity within ipsilateral and contralateral corticospinal pathways may explain mirroring of movements. Alternatively, a failure of transcallosal inhibition, acquired at the time of myelination of these pathways, may contribute to the genesis of mirror movements. Loss of joint position sense following thalamic lesions may be of relevance. A deficit of sustained attention has also been postulated as the cause of mirror movements.

- Mayston MJ, Harrison LM, Quinton R, Stephens JA, Krams M, Bouloux P-MG. Mirror movements in X-linked Kallmann's syndrome. I. A neurophysiological study. *Brain* 1997; **120:** 1199-216

[Cross References: ANOSMIA; ATTENTION; PROPRIOCEPTION; SYNKINESIS]

"Mirror Sign"

The term "mirror sign" has been applied to the phenomenon of misrecognition of self as another when seen in a mirror. This may occur in Alzheimer's disease and frontotemporal dementia, and is associated with impaired cognition, confabulation, and prefrontal dysfunction. Some authors believe "the phenomenon of the mirror" to be an extreme example of prosopagnosia, but other studies have not found an association.

- Caixeta LF, Caramelli P, Bahia V, Buchpiguel CA, Nitrini R. Clinical and neuroanatomical correlates of the mirror sign in frontotemporal dementia and Alzheimer's disease. *Neurobiology of Aging* 2000; **21(suppl1):** S217 (abstract 988)

[Cross References: CONFABULATION; "PICTURE SIGN"; PROSOPAGNOSIA]

Mitbewegungen

- see SYNKINESIA, SYNKINESIS

Mitgehen

- see NEGATIVISM

Monoballismus

Monoballismus is ballism affecting a single limb.

[Cross References: BALLISM, BALLISMUS; HEMIBALLISMUS]

Monomelia

- see MONOPARESIS, MONOPLEGIA

Mononeuritis Multiplex, Mononeuropathy Multiplex

- see NEUROPATHY

Mononeuropathy

- see NEUROPATHY

Monoparesis, Monoplegia

Monoparesis is weakness, monoplegia complete weakness ("paralysis"), of a single limb. Monoparesis of the arm or leg of upper motor neurone type is usually cortical

in origin, although may unusually arise from a cord lesion (leg more frequently than arm). Hoover's sign and Babinski's trunk-thigh test may be helpful in deciding whether monoparetic/monoplegic leg weakness is of non-organic origin; the "face-hand test" in arm weakness.

Peripheral disorders can sometimes present exclusively with single limb weakness, such as monomelic motor neurone disease, multifocal motor neuropathy with conduction block, and Guillain-Barré syndrome.

[Cross References: BABINSKI'S TRUNK-THIGH TEST; "FACE-HAND TEST"; HEMIPARESIS; HOOVER'S SIGN]

Monotonia

Monotonia is a restricted range of speech inflection, occurring with hypophonia as part of the hypokinetic dysarthria observed in parkinsonism.

[Cross References: DYSARTHRIA; HYPOPHONIA; PARKINSONISM]

Moria

- see EMOTIONAL LABILITY

Mutism

Mutism is absence of speech output. This may result from psychiatric disease (schizophrenia, affective disorders, with or without catatonia), or neurological disease, for example:

- akinetic mutism;
- dementia syndromes, especially frontal lobe dementia, late stages of primary progressive aphasia;
- encephalopathy (toxic/drug-induced/metabolic);
- damage to Broca's area, supplementary motor area; severe pseudobulbar palsy, bilateral thalamic damage;
- cerebellar mutism: rare, following midline cerebellar surgery in children;
- bilateral vocal cord paralysis (although this may be better termed aphonia);
- psychogenic.

In neurological disorders there may be difficulty initiating movements, completing motor sequences, or inhibition of appropriate responses.

- Altshuler LL, Cummings JL, Mills MJ. Mutism: review, differential diagnosis and report of 22 cases. *American Journal ofPsychiatry* 1986; **143:** 1409-1414 (erratum: *American Journal ofPsychiatry* 1987; **144:** 542)
- Ersahin Y, Mutluer S, Cagli S, Duman Y. Cerebellar mutism: report of seven cases and review of the literature. *Neurosurgery* 1996; **38:** 60-6

[Cross References: AKINETIC MUTISM; APHONIA; CATATONIA; DEMENTIA; ENCEPHALOPATHY; PSEUDOBULBAR PALSY]

Mydriasis

Mydriasis is an abnormally large pupil, due to dilatation. Causes include:

- Oculoparasympathetic paresis, from lesions at the Edinger-Westphal nucleus or anywhere along the course of the oculomotor (III) nerve;

- Tonic enlargement of the pupil (Holmes-Adie pupil);
- Sympathomimetic drugs, *e.g.* adrenaline.

If only one pupil appears large (anisocoria), it is important to distinguish mydriasis from contralateral miosis, when a different differential will apply (*e.g.* Horner's syndrome).
[Cross References: ANISOCORIA; HOLMES- ADIE PUPIL, HOLMES-ADIE SYNDROME; HORNER'S SYNDROME; MIOSIS]

Myelopathy
A myelopathy is a disorder of the spinal cord. Such disorders may be further characterized according to whether the responsible lesion lies within or outside the spinal cord: intrinsic or intramedullary lesions are always intradural; extrinsic or extramedullary lesions may be intradural or extradural. It may be possible to differentiate intramedullary from extramedullary lesions on clinical grounds, although this distinction is never absolute because of clinical overlap.
Clinical features of Extrinsic/Extramedullary Myelopathy:

- motor - sequential spastic paraparesis below the level of the lesion; upper motor neurone (UMN) signs occur early; lower motor neurone (LMN) signs are unusual and have a segmental (radicular) distribution if present;
- sensory - symptoms of pain may be radicular (*e.g.* secondary to a neurofibroma) or verterbal (*e.g.* secondary to neoplastic or inflammatory processes); sensory signs are not usually marked until the later stages, and all modalities are often involved. A Brown-Séquard syndrome may be commoner in extrinsic than intrinsic myelopathies;
- sphincters - may have bladder urgency, impotence.

Pathologies commonly causing extrinsic myelopathy include:

- prolapsed disc, osteophyte bar;
- tumour (primary, secondary);
- arteriovenous malformation/haematoma;
- abscess.

Clinical features of Intrinsic/Intramedulary Myelopathy include the following, dependent on the extent to which the cord is involved: some pathologies have a predilection for posterior columns, central cord, *etc.*

- motor - LMN signs may be prominent and diffuse; UMN signs tend to occur late (spastic paraparesis below level of lesion). A combination of UMN and LMN signs is much more likely to reflect intrinsic than extrinsic pathology;
- sensory - symptoms of central (funicular) pain may occur; dissociated sensory loss (spinothalamic > dorsal column involvement, or *vice versa*), suspended sensory loss, and sacral sparing are characteristic of intramedullary lesions; a Brown-Séquard syndrome may occur. Vibratory sensibility is more often affected than proprioception;
- sphincters - bladder involvement common, often early and slow to recover.

Pathologies commonly causing intrinsic myelopathy include:

- multiple sclerosis or other inflammatory process causing transverse myelitis (complete or partial), *e.g.* viral infection, HTLV-1 infection, tabes dorsalis;
- tumour (primary, secondary);
- syringomyelia;
- infarction, *e.g.* anterior spinal artery syndrome;
- metabolic causes: vitamin B_{12} deficiency producing subacute combined degeneration of the cord..

MR imaging of the cord is often helpful in defining the cause of myelopathy.

- Johnston RA. Acute spinal cord compression. In: Hughes RAC (ed.). *Neurological Emergencies.* London: BMJ Publishing 1997 (2nd edition): 272-94
- Tartaglino LM, Flanders AE, Rapoport RJ. Intramedullary causes of myelopathy. *Seminars in Ultrasound, CT, and MRI* 1994; **15**: 158-88
[Cross References: BROWN-SÉQUARD SYNDROME; LOWER MOTOR NEURONE SYNDROME; PARAPARESIS; PROPRIOCEPTION; SACRAL SPARING; UPPER MOTOR NEURONE SYNDROME; VIBRATION]

Myerson's Sign

- see GLABELLAR TAP REFLEX

Myoclonus

Myoclonus is involuntary, "shock-like", muscle jerking, rhythmic or irregular, arising in the central nervous system (CNS). This may be focal, multifocal, or generalised. Multiple irregular asynchronous myoclonic jerks may be termed polymyoclonus. Myoclonus may be characterized in several ways:

Clinical classification (by observation, examination):
- ~ *spontaneous;*
- ~ *action* - following voluntary action; may be elicited by asking patient to reach out to touch the examiner's hand;
- ~ *reflex, stimulus-sensitive* - jerks produced by somaesthetic stimulation of a limb.

Anatomical/pathophysiological classification (by electrophysiological recordings):
- ~ *cortical;*
- ~ *subcortical/reticular;*
- ~ *propriospinal/segmental.*

Aetiological classification:
- ~ *Physiological, e.g.* "sleep starts" (hypnic jerks);
- ~ *Essential*: in the absence of any other abnormality of the CNS;
- ~ *Epileptic*: as a manifestation of idiopathic epilepsy;
- ~ *Symptomatic*: of other neurological diseases, of which there are many, including:
  - anoxic brain injury (Lance-Adams syndrome);
  - vascular lesions;
  - neoplasia;
  - encephalopathies: especially of metabolic origin, but also toxic, viral, paraneoplastic, mitochondrial;

- degenerations: basal ganglia, spinocerebellar;
- malabsorption syndromes (coeliac disease, Whipple's disease);
- storage disorders, *e.g.* Lafora body disease, Tay-Sachs disease, sialidosis;
- dementias: Alzheimer's disease (usually late), Creutzfeldt-Jakob disease (usually early).

The clinical differential diagnosis of myoclonus includes chorea, tic, tremor, and certain peripheral nerve disorders (fasciculation, myokymia).

Drugs useful in the treatment of myoclonus include clonazepam, sodium valproate, primidone, and piracetam. These may need to be given in combination to suppress severe action myoclonus.

Brief lapses of muscle contraction with loss of posture may also be seen; since these are in some ways the converse of myoclonus they have in the past been labelled negative myoclonus, but the term asterixis is now preferred.

- Marsden CD, Hallett M, Fahn S. The nosology and pathophysiology of myoclonus. In: Marsden CD, Fahn S (eds.). *Movement Disorders.* London, Buttenworth 1982: 196-248

- Obeso JA, Artieda J, Rothwell JC, Day B, Thompson P, Marsden CD. The treatment of severe action myoclonus. *Brain* 1989; **112:** 765-77

[Cross References: ASTERIXIS; CHOREA; FASCICULATION; HICCUPS; MYOKYMIA; PALATAL MYOCLONUS; TIC; TREMOR]

Myokymia

Myokymia is an involuntary, spontaneous, wave-like, undulating, flickering movement within a muscle (*cf.* fasciculation); it may be likened to a "bag of worms". Electrophysiologically this corresponds to regular groups of motor unit discharges, of peripheral nerve origin. Myokymia is thus related to neuromyotonia and stiffness, since there may be concurrent impairment of muscle relaxation and a complaint of muscle cramps.

A syndrome of superior oblique myokymia is described, often following superior oblique palsy, which produces a microtremor of the eye and causes oscillopsia or transient diplopia. Facial myokymia is a rare facial dyskinesia, possibly related to disinhibition of the facial (VII) nerve nucleus by focal pontine lesions (tumour, demyelination).

- Thompson PD. Stiff people. In: Marsden CD, Fahn S (eds.). *Movement disorders 3.* Boston: Butterworth 1994:373-405

[Cross References: FASCICULATION; MYOTONIA; NEUROMYOTONIA; STIFFNESS]

Myopathy

The term myopathy means a primary disorder of muscle causing wasting and/or weakness in the absence of sensory abnormalities. Clinically, myopathic processes need to be differentiated from neuropathies, particularly anterior horn cell diseases and motor neuropathies, and neuromuscular junction disorders. Generally in primary

muscle disease, there are no fasciculations, reflexes are lost late, and phenomena such as fatigue and facilitation do not occur.

Myopathies may be subdivided according to the clinical pattern of weakness, and/or their aetiology:

Proximal: affecting shoulder abductors, hip flexors predominantly:

- Inflammatory: polymyositis, dermatomyositis;
- Progressive muscular dystrophies: Duchenne, Becker, limb-girdle, facioscapulo-humeral (FSH);
- Metabolic: acid-maltase deficiency; thyroid dysfunction, Cushing's syndrome;
- Non-metastatic feature of malignant disease.

Distal: an unusual pattern for myopathy, which needs to be differentiated from distal polyneuropathy:

- Myotonic dystrophy;
- Miyoshi dystrophy;
- Desmin myopathy.

Bulbar palsy (*q.v.*).
Facial paresis (*q.v.*).
Diaphragm weakness:

- acid-maltase deficiency;
- acute polymyositis;
- neuralgic amyotrophy.

Axial myopathy:

- Camptocormia ("bent spine syndrome");
- Dropped head syndrome.

- Swash M. Dropped-head and bent-spine syndromes: axial myopathies? *Lancet* 1998; **352:** 758

[Cross References: ATROPHY; BULBAR PALSY; CAMPTO-CORMIA; DROPPED HEAD SYNDROME; FACIAL PARESIS; FATIGUE; GOWERS SIGN; HEAD DROOP, HEAD DROP; PARADOXICAL DIAPHRAGM MOVEMENT; WASTING; WEAKNESS]

Myorhythmia

Myorhythmia is an involuntary movement disorder characterized by rhythmic contraction (1-2 Hz) of either the oculomasticatory or oculo-facial skeletal muscles. Characteristically there is also convergent-divergent pendular nystagmus with synchronous rhythmic movement of the mouth, tongue, jaw and sometimes proximal and distal skeletal muscles. The movements are continuous and persist during sleep.

Although very rare, oculomasticatory myorhythmia is of diagnostic importance since it is pathognomonic for Whipple's disease of the nervous system. The neurological manifestations of Whipple's disease are protean, and include dementia, ataxia, supranuclear ophthalmoplegia (with sparing of the pupils), seizures, myoclonus, nystagmus and psychosis.

Whipple's disease is caused by the bacterium *Tropheryma whipplei.* Treatment is with antibiotics, usually a two week intravenous course of trimethoprim-sulpha-

methoxazole or ceftriazone followed by oral treatment for one year; valproate may be helpful for involuntary movements which do not respond to antibiotics.

- Simpson DA, Wishnow R, Gargulinski RB, Pawlak AM. Oculofacial-skeletal myorhythmia in central nervous system Whipple's disease: additonal case and review of the literature. *Movement Disorders* 1995; **10:** 195-200
- Anderson M. Neurology of Whipple's disease. *Journal of Neurology, Neurosurgery and Psychiatry* 2000; **68:** 2-5

[Cross References: ATAXIA; DEMENTIA; MYOCLONUS; NYSTAGMUS]

Myotonia

Myotonia is a stiffness of muscles with inability to relax after voluntary contraction (action myotonia), or induced by electrical or mechanical (*e.g.* percussion myotonia) excitation. The phenomenon is often described by patients as "cramp" or stiffness. This is a reflection of primary muscle disease (*i.e.* myogenic; *cf.* neuromyotonia, neurogenic muscle stiffness), which persists after peripheral nerve or neuromuscular junction blockade.

Electrophysiology reveals myotonic discharges, with prolonged twitch relaxation phase, which may be provoked by movement, percussion, and electrical stimulation of muscle; discharges typically wax and wane.

A similar clinical phenomenon of slow muscle relaxation may be observed in other circumstances, for example hypothyroidism, but without the characteristic EMG findings of myotonia, hence this is labelled as pseudomyotonia. Paramyotonia is myotonia exacerbated by cold and exertion (paradoxical myotonia).

Recognised causes of myotonia include:

- myotonic dystrophy, or myotonia dystrophica;
- hyperkalaemic periodic paralysis;
- myotonia congenita (autosomal dominant Thomsen's disease, autosomal recessive Becker's myotonia);
- K^+-aggravated myotonia;
- Schwartz-Jampel syndrome (chondrodystrophic myotonia);
- proximal myotonic myopathy (PROMM)

Mutations in genes encoding voltage-gated ion channels have been identified in some of the inherited myotonias, hence these are channelopathies: skeletal muscle voltage-gated Na^+ channel mutations have been found in K^+-aggravated myotonia, and also paramyotonia congenita and hyperkalaemic periodic paralysis. Chloride (Cl^-) channel mutations have been identified in myotonia congenita. These latter conditions respond best to mexilitene.

[Cross References: NEUROMYOTONIA; PARAMYOTONIA; PERCUSSION MYOTONIA; PSEUDOMYOTONIA; STIFFNESS; WOLTMAN'S SIGN]

N

Narcolepsy
- see HYPERSOMNOLENCE

Negative Myoclonus
- see ASTERIXIS

Negative Tremor
- see ASTERIXIS

Negativism
Negativism is a motor sign of mental disorder, usually schizophrenia, consisting of the patient doing the opposite of what is asked and actively resisting efforts to persuade compliance. Movement of a limb in response to application of pressure despite the patient having been told to resist (*mitgehen*) is one element of negativism. It may also be a feature of catatonia. The similarity of some of these features to *gegenhalten* suggests the possibility of frontal lobe dysfunction as the underlying cause.
[Cross References: CATATONIA; *GEGENHALTEN*]

Neglect
Neglect is a failure to orient toward, respond to, or report novel or meaningful stimuli. If failure to respond can be attributed to concurrent sensory or motor deficits (*e.g.* hemiparesis, hemianopia, visuospatial deficits) neglect is not present.
Neglect can involve stimuli in the extrapersonal environment (*e.g.* visual neglect) or personal space (*e.g.* personal neglect or asomatognosia). Neglect of contralateral hemispace may also be called unilateral spatial neglect, inattention, or hemineglect. Lesser degrees of neglect may be manifest as extinction (double simultaneous stimulation). Motor neglect may be evident as hemiakinesia, hypokinesia, or motor impersistence. Alloaesthesia and allokinesia may also be features of neglect.
Neglect may be obvious (*e.g.* patient not dressing one side of the body), but is sometimes more subtle, in which case it may be tested for using various simple tests:

- Cancellation tests, *e.g.* stars (unstructured array), letters (structured array);
- Figure copying, *e.g.* Rey-Osterreith figure;
- Line bisection, numbering a clock face;
- Drawing from memory.

Neglect is commoner after right rather than left brain damage (usually of vascular origin). Marked degrees of neglect may seriously hamper attempts at neurorehabilitation.

- Bowen A, McKenna K, Tallis RC. Reasons for variability in the reported rate of occurrence of unilateral spatial neglect after stroke. *Stroke* 1999; **30**: 1196-202

- Parkin AJ. *Explorations in cognitive neuropsychology.* Hove: Psychology Press 1996:90-109
[Cross References: ALLOAESTHESIA; ALLOKINESIA; ASOMATOGNOSIA; EXTINCTION; HEMIAKINESIA; HYPOKINESIA; IMPERSISTENCE]

Negro's Sign
Negro has two eponymous signs:

- Cogwheel (jerky) type of rigidity in basal ganglia disorders;
- In both peripheral and central facial paralysis, the eyeball deviates outward and elevates more than normal when the patient attempts to look up due to overaction of the inferior oblique and superior rectus muscles, respectively.

[Cross References: BELL'S PALSY; FACIAL PARESIS; PARKINSONISM; RIGIDITY]

Neologism
A neologism is a non-word approximating to a real word, produced in spontaneous speech; it is thought to result from an inability to organize phonemes appropriately. Hence, this is a type of literal or phonemic paraphasia encountered in aphasic syndromes, most usually those resulting from left superior temporal lobe damage (Wernicke type).
[Cross References: APHASIA; PARAPHASIA; WERNICKE'S APHASIA]

Nerve Thickening
The characterization of a peripheral neuropathy should always include examination to see if any nerves are thickened. Good places to feel for nerve thickening include the elbow (ulnar nerve), anatomical snuff box (superficial radial nerves), and head of the fibula (common peroneal nerve). Nerve thickening may be noted in:

- amyloid polyneuropathy
- leprosy
- tomaculous neuropathy
- hereditary neuropathy with liability to pressure palsies

[Cross References: NEUROPATHY]

Neuromyotonia
Neuromyotonia is neurogenic muscle stiffness (*cf.* myotonia, myogenic muscle stiffness) which reflects continuous muscle activity. Clinically this is manifest as muscle cramps and stiffness, particularly during and after muscle contraction, and as muscular activity at rest (myokymia, fasciculations). Tendon areflexia and abnormal postures of hands and feet may also be observed.

A syndrome of ocular neuromyotonia has been described in which spasms of the extraocular muscles cause a transient heterophoria and diplopia.

Physiologically neuromyotonia is characterized by continuous motor unit and muscle fibre activity which is due to peripheral nerve hyperexcitability; it is abolished by curare (*cf.* myotonia). Neuromyotonia may be associated with autoantibodies directed against presynaptic voltage-gated $\mathbf{K}^+$ channels. Around 20% of patients have

an underlying small-cell lung cancer or thymoma, suggesting a paraneoplastic aetiology in these patients. Neuromyotonia has also been associated with mutations within the voltage-gated K^+ ion channel gene

Carbamazepine and phenytoin may help the stiffness and areflexia.

- Browne DL, Gancher ST, Nutt JG, *et al.* Episodic ataxia-myokymia syndrome is associated with a point mutation in the human potassium channel gene, KCNA1. *Nature Genetics* 1994; **8**: 136-40
- Ezra E, Spalton D, Sanders MD, Graham EM, Plant GT. Ocular neuromyotonia. *British Journal of Ophthalmology* 1996; **80**: 350-5
- Hart IK, Waters C, Vincent A, *et al.* Autoantibodies detected to expressed K^+ channels are implicated in neuromyotonia. *Annals of Neurology* 1997; **48**: 238-46
- Isaacs H. A syndrome of continuous muscle-fibre activity. *Journal of Neurology, Neurosurgery and Psychiatry* 1961; **24**: 319-25

[Cross References: FASCICULATION; MYOKYMIA; MYOTONIA; PARAMYOTONIA; PSEUDOMYOTONIA; STIFFNESS]

Neuronopathy

- see NEUROPATHY

Neuropathy

Neuropathies are disorders of peripheral nerves. Various clinical patterns of peripheral nerve involvement may be seen:

~ *Mononeuropathy* - sensory and/or motor involvement in the distribution of a single nerve;
~ *Mononeuropathy multiplex* - simultaneous involvement of two or more nerves, usually in different parts of the body; if due to inflammatory disease (as is often the case) this may be described as mononeuritis multiplex;
~ *Polyneuropathy* - a widespread process, predominantly affecting the distal parts of nerves; may be predominantly sensory ("glove and stocking" sensory loss) or motor, with or without concomitant autonomic involvement.

These clinical patterns may need to be differentiated in practice from disorders affecting the neuronal cell bodies in the ventral (anterior) horns of the spinal cord or dorsal root ganglia (motor and sensory neuronopathies, respectively); and disorders of the nerve roots (radiculopathy) and plexuses (plexopathy). Clinical signs resulting from neuropathies are of lower motor neurone type (wasting, weakness, reflex diminution or loss).

The causes of neuropathy are legion. Mononeuropathies often result from local compression (entrapment neuropathy), trauma, or diabetes. Mononeuropathy multiplex often reflects intrinsic inflammation (*e.g.* polyarteritis nodosa, Churg-Strauss syndrome, systemic lupus erythematosus, rheumatoid arthritis, Sjögren's syndrome, cryoglobulinaemia, isolated PNS vasculitis). Polyneuropathies may have genetic, infective, inflammatory, toxic, nutritional, and endocrine aetiologies. Many neuropathies, particularly polyneuropathies in the elderly, remain idiopathic, despite intensive investigation.

Sensory neuronopathies have a rather more limited differential diagnosis, including:

- Paraneoplasia: anti-Hu antibody syndrome (although a similar syndrome, presumed paraneoplastic, may occur in the absence of these antibodies);
- Sjögren's syndrome;
- Associated with anti-GD1b ganglioside antibodies
- CIDP;
- HIV.

- Staal A, van Gijn J, Spaans F. *Mononeuropathies: examination, diagnosis and treatment.* London: WB Saunders 1999
- Stewart JD. *Focal peripheral neuropathies.* Philadelphia: Lippincott Williams & Wilkins 2000 (3rd edition)
[Cross References: AMYOTROPHY; LOWER MOTOR NEURONE SYNDROME; PLEXOPATHY; RADICULOPATHY; WASTING; WEAKNESS]

Nuchal Rigidity
Nuchal rigidity is neck stiffness, and is usually synonymous with meningism, in which case other signs of meningeal irritation are usually present (Kernig's sign, Brudzinski's neck sign). If these other signs are absent, then isolated nuchal rigidity may suggest a foraminal pressure cone. It may also occur in syndromes causing predominantly axial (as opposed to limb) rigidity (*e.g.* Steele-Richardson-Olszewski syndrome). Resistance to passive neck movement is also seen in intubated patients.
[Cross References: BRUDZINSKI'S (NECK) SIGN; KERNIG'S SIGN; MENINGISM; PARKINSONISM]

Nuchocephalic Reflex
The nuchocephalic reflex is present in infants and children up to the age of 4 years: on rapid turning of the shoulders of a standing subject to either left or right (eyes closed to avoid fixation) there is bilateral contraction of the cervical musculature so that the head is held in the original position. Beyond about four years of age the reflex is inhibited, such that the head is actively turned in the direction of shoulder movement after a time lag of about half a second. If the reflex is present in adults (*i.e.* disinhibited), it is claimed to be a "regressive" sign, indicative of diffuse cerebral dysfunction.

- Jenkyn LR, Walsh DB, Walsh BT, Culver CM, Reeves AG. The nuchocephalic reflex. *Journal of Neurology, Neurosurgery and Psychiatry* 1975; **38**: 561-6

Nyctalopia
Nyctalopia, or night blindness, is an impairment of visual acuity specific to scotopic vision, implying a loss or impairment of rod photoreceptor function. Patients may spontaneously complain of a disparity between daytime and nocturnal vision, in which case acuity should be measured in different ambient illumination.
Nyctalopia may be a feature of:

- Retinitis pigmentosa;
- Vitamin A deficiency;
- Cancer-associated retinopathy: most commonly associated with small-cell lung cancer (anti-recoverin antibodies may be detected), though gynaecological

malignancy and melanoma have also been associated (with anti-bipolar retinal cell antibodies in the latter).

[Cross References: HEMERALOPIA; RETINITIS PIGMENTOSA]

Nylen-Bárány Manoeuvre

- see HALLPIKE MANOEUVRE

Nystagmoid Jerks

- see NYSTAGMUS

Nystagmus

Nystagmus, or talantropia, is an involuntary bilateral oscillation of the eyeballs (very occasionally it is unilateral, *e.g.* in internuclear ophthalmoplegia), of which many varieties are described. This may be:

Physiological:

- Optokinetic nystagmus (OKN; *e.g.* looking out of a moving railway carriage);
- Induced by vestibular stimuli (*e.g.* merry-go-round; caloric testing);
- Nystagmoid jerks: in extremes of lateral or vertical gaze (end-point nystagmus, a form of gaze-evoked nystagmus);

Pathological: this may be classified according to direction, waveform, anatomy/aetiology, or clinical frequency (common, rare). When describing nystagmus, it is necessary to make observations in the nine cardinal positions of gaze for the direction, amplitude and beat frequency of nystagmus. Nystagmus may be abortive or sustained in duration. The intensity of jerk nystagmus may be classified by a scale of three degrees:

~ 1st degree - present when looking in the direction of the fast phase;
~ 2nd degree - present in the neutral position;
~ 3rd degree - present when looking in the direction of the slow phase (*i.e.* present in all directions of gaze).

It is important to distinguish nystagmus from other involuntary eye movements such as square-wave jerks, ocular flutter, and opsoclonus (*q.v.*).

Directional classification of nystagmus:

~ Horizontal (common)
~ Vertical (rare):

Downbeat: seen with structural lesions of the cervico-medullary junction, midline cerebellum and floor of the 4th ventricle, but also with more diffuse cerebellar disease;

Upbeat: of less localising value than downbeat, upbeat nystagmus may occur with pontomesencephalic, pontomedullary, and even caudal medullary lesions (infarct, inflammation); bow-tie nystagmus is probably a variant of upbeat nystagmus.

~ Torsional: usually accompanies horizontal nystagmus of peripheral vestibular (labyrinthine) origin.

Waveform classification of nystagmus:

~ Jerk nystagmus, characterized by a slow drift of the eyes in one direction (slow phase) followed by a rapid, corrective, saccadic movement in the opposite direction (fast phase); the direction of jerk nystagmus is named by the direction of the fast phase, but it is the character of the slow phase, the pathological part of the process, which is more eloquent regarding anatomical correlation (*vide infra);*
~ Pendular or undulatory nystagmus, in which the movements of the eyes are more or less equal in amplitude and velocity (sinusoidal oscillations) about a central (null) point. This is often congenital, may be conjugate or disconjugate (sometimes monocular), but is not related to concurrent internuclear ophthalmoplegia or asymmetry of visual acuity.

When studied using oculography, the slow phase of jerk nystagmus may show a uniform velocity ("saw-toothed"), indicative of imbalance in vestibulo-ocular reflex activity. A slow phase with exponentially decreasing velocity (negative exponential slow phase) is ascribed to "leakiness" of a hypothetical neural integrator, a structure which converts eye or head velocity signals into approximations of eye or head position signals (thought to lie in interstitial nucleus of Cajal in the midbrain for vertical eye movements, and in the nucleus propositus hypoglossi for horizontal movements). A slow phase with exponentially increasing velocity (high-gain instability, runaway movements) may be seen in congenital or acquired pendular nystagmus. The pathophysiology of acquired pendular nystagmus is thought to be deafferentation of the inferior olive by lesions of the red nucleus, central tegmental tract, or medial vestibular nucleus.

Anatomical/aetiological classification of nystagmus:
~ Peripheral Vestibular: unidirectional (directed to side opposite lesion), and more pronounced when looking in direction of the fast phase (*i.e.* 1st degree), usually with a rotatory component and associated with vertigo. Tends to fatigue, and usually transient. Nystagmus of peripheral vestibular origin is typically reduced by fixation (hence these patients hold their heads still) and enhanced by removal of visual fixation (in the dark, with Frenzel's lenses).
~ Central Vestibular: unidirectional or multidirectional, 1st, 2nd or 3rd degree; typically sustained and persistent. There may be other signs of central pathology (*e.g.* cerebellar signs, upper motor neurone signs). Not affected by removal of visual fixation.
~ Cerebellar/brainstem: commonly gaze-evoked due to a failure of gaze-holding mechanisms. It may be unidirectional with unilateral cerebellar lesion (*e.g.* vascular disease) in which case it typically occurs when the eyes are looking in the direction of the lesion (*cf.* peripheral vestibular nystagmus); multidirectional nystagmus of cerebellar origin may occur in multiple sclerosis, drug/toxin exposure, cerebellar degenerations.
~ Congenital: usually horizontal, pendular type nystagmus; worse with fixation, attention, anxiety. It may appear with blindness of childhood onset, or be acquired with neurological disease (multiple sclerosis, mitochondrial disease, Whipple's disease, Pelizaeus-Merzbacher disease).

Other forms of nystagmus include:
~ *Ataxic/Dissociated:* in abducting >> adducting eye, as in internuclear opthalmoplegia and pseudo-internuclear ophthalmoplegia.

~ *Periodic Alternating*: primary position nystagmus, almost always in the horizontal plane, which stops and then reverses direction every minute or so; 4-5 minutes observation may be required to see the whole cycle; localising value similar to downbeat nystagmus.

~ *Convergence-retraction* (Körber-Salus-Elschnig syndrome): adducting saccades (medial rectus contraction), occurring spontaneously or on attempted upgaze, often accompanied by retraction of the eyes into the orbits, associated with mesencephalic lesions of the pretectal region (*e.g.* pinealoma).

~ *See-saw*: a disconjugate cyclic movement of the eyes, comprising elevation and intorsion of one eye while the other eye falls and extorts, followed by reversal of these movements; may be congenital (*e.g.* with albinism, retinitis pigmentosa) or acquired (mesodiencephalic or lateral medullary lesions, *e.g.* brainstem stroke, head trauma, syringobulbia).

Many pathologies may cause nystagmus, the commonest being demyelination, vascular disease, tumour, neurodegenerative disorders of cerebellum and/or brainstem, metabolic causes (*e.g.* Wernicke-Korsakoff's syndrome), paraneoplasia, drugs (alcohol, phenytoin, barbiturates, sedative-hypnotic drugs), toxins, and epilepsy. Treatment of nystagmus is usually that of the underlying cause, where possible. Pendular nystagmus may respond to anticholinesterases, consistent with its being a result of cholinergic dysfunction. Periodic alternating nystagmus responds to baclofen, hence the importance of making this diagnosis. See-saw nystagmus may respond to baclofen, clonazepam, or alcohol.

- Leigh RJ, Zee DS. *The Neurology of Eye Movements.* New York: OUP 1999 (3rd edition)
- Lopez LI, Bronstein AM, Gresty MA, Du Boulay EPG, Rudge P. Clinical and MRI correlates in 27 patients with acquired pendular nystagmus. *Brain* 1996; **119**: 465-72
- Lueck CJ. A simple guide to nystagmus. *Hospital Medicine* 2000; **61:** 544-6, 548-9
- Rudge P, Bronstein AM. Investigations of disorders of balance. In: Hughes RAC (ed.). *Neurological Investigations.* London, BMJ Publishing 1997: 283-314

[Cross References: CALORIC TESTING; HALLPIKE MANOEUVRE; INTER-NUCLEAR OPHTHALMOPLEGIA; MYORHYTHMIA; OPTOKINETIC NYS-TAGMUS (OKN), OPTOKINETIC RESPONSE; OPSOCLONUS; OSCILLOPSIA; PALATAL MYOCLONUS; PSEUDO-INTERNUCLEAR OPHTHALMOPLEGIA; SPASMUS NUTANS; SQUARE-WAVE JERKS; VERTIGO]

O

Obscurations
Obscurations are transient losses ("greying out") of vision lasting a few seconds, occurring in the context of raised intracranial pressure (ICP), and especially associated with activities known to elevate ICP (coughing, sneezing, bending down, straining at stool) and relieved by their cessation. These symptoms are thought to reflect critical compromise of optic nerve head perfusion and are invariably associated with the finding of papilloedema. Obscurations mandate urgent investigation and treatment to prevent permanent visual loss.
[Cross References: PAPILLOEDEMA]

Obtundation
Obtundation is a state of altered consciousness characterized by reduced alertness and a lessened interest in the environment, sometimes described as psychomotor retardation or torpor. An increased proportion of time is spent asleep and the patient is drowsy when awake. Obtundation is less severe impairment of consciousness than stupor.
[Cross References: COMA; PSYCHOMOTOR RETARDATION; STUPOR]

Ocular Apraxia
Ocular apraxia (ocular motor apraxia) is a disorder of voluntary saccade initiation; reflexive saccades and spontaneous eye movements are preserved. Ocular apraxia may be overcome by using dynamic head thrusting, with or without blinking (to suppress vestibulo-ocular reflexes): the desired fixation point is achieved through reflex contraversive tonic eye movements to the midposition following the overshoot of the eyes caused by the head thrust.

The anatomical substrate of ocular apraxia is not certain. Ocular apraxia may occur as a congenital syndrome (in the horizontal plane only: Cogan's syndrome), or may be acquired in ataxia telangiectasia (Louis-Bar syndrome), Niemann-Pick disease (mainly vertical plane affected), and Gaucher's disease (horizontal plane only).
[Cross References: SACCADES]

Ocular Bobbing
Ocular bobbing refers to intermittent abnormal vertical eye movements, usually conjugate, consisting of a fast downward movement followed by a slow return to the initial horizontal eye position. The sign has no precise localizing value, but is most commonly associated with intrinsic pontine lesions, *e.g.* infarct, haemorrhage, tumour, central pontine myelinolysis. It has also been described in encephalitis, Creutzfeldt-Jakob disease, and toxic encephalopathies. Its pathophysiology is uncertain but may involve mesencephalic and medullary burst neurone centres.

Variations on the theme include:

~ Inverse ocular bobbing (also known as fast upward ocular bobbing, ocular dipping): slow downward movement, fast return;

~ Reverse ocular bobbing: fast upward movement, slow return to midposition;
~ Converse ocular bobbing (also known as slow upward ocular bobbing, reverse ocular dipping): slow upward movement, fast down.

- Fisher CM. Ocular bobbing. *Archives of Neurology* 1964; **11**: 543-6.
- Bosch EP, Kennedy SS, Aschenbrener CA. Ocular bobbing: the myth of its localizing value. *Neurology* 1975; **25**: 949-53
[Cross References: OCULAR DIPPING]

Ocular Dipping
Ocular dipping, or inverse ocular bobbing, consists of a slow spontaneous downward eye movement with a fast return to the midposition. This may be observed in anoxic coma or following prolonged status epilepticus and is thought to be a marker of diffuse, rather than focal, brain damage.
Reverse ocular dipping (slow upward ocular bobbing) consists of a slow upward movement followed by a fast return to the midposition.

- Stark JR, Masucci EF, Kurtzke JF. Ocular dipping. *Neurology* 1984; **34**: 391-3
[Cross References: OCULAR BOBBING]

Ocular Flutter
Ocular flutter is an eye movement disorder characterized by involuntary bursts of back-to-back horizontal saccades without an intersaccadic interval (*cf.* square-wave jerks). Ocular flutter may be accurately diagnosed with oculography. (For further details of aetiology and pathogenesis, see Opsoclonus.)
[Cross References: OPSOCLONUS; SACCADES; SACCADIC INTRUSION; SQUARE-WAVE JERKS]

Ocular Tilt Reaction
The ocular tilt reaction is a synkinesis consisting of lateral head tilt, skew deviation (hypotropia), and ocular torsion, all to one side, due to a disorder of torsional vestibulo-ocular reflexes perhaps as a consequence of imbalance of otolith (and posterior semicircular canal?) inputs. This may be a tonic phenomenon (as in the lateral medullary syndrome) or paroxysmal (multiple sclerosis). Different mechanisms may provoke this reaction, which reflects pathology in graviceptive pathways anywhere from utricle and semicircular canal to contralateral rostral midbrain (medial longitudinal fasciculus, interstitial nucleus of Cajal) via ponto-medullary vestibular nuclei. Different patterns of skew deviation may help to localise the responsible lesion.

- Brandt Th, Dieterich M. Different types of skew deviation. *Journal of Neurology, Neurosurgery and Psychiatry* 1991; **54**: 549-50
[Cross References: HYPOTROPIA; SKEW DEVIATION; SYNKINESIS; TULLIO PHENOMENON; VESTIBULO-OCULAR REFLEXES]

Oculocephalic Response
Oculocephalic responses are most commonly elicited in unconscious patients; the head is passively rotated in the horizontal or vertical plane (doll's head maneouvre)

and the eye movements are observed. Conjugate eye movement in a direction opposite to that in which the head is turned is indicative of an intact brainstem (intact vestibulo-ocular reflexes). With pontine lesions, the oculocephalic responses may be lost, after roving eye movements but before caloric responses disappear.
[Cross References: CALORIC TESTING; COMA; ROVING EYE MOVEMENTS; VESTIBULO-OCULAR REFLEXES]

Oculogyric Crisis
Oculogyric crisis is an acute dystonia of the ocular muscles, usually causing upward and lateral displacement of the eye. It is often accompanied by a disorder of attention (obsessive, persistent thoughts), with or without dystonic or dyskinetic movements. It occurs particularly with symptomatic (secondary) as opposed to idiopathic (primary) dystonias, for example post-encephalitic and neuroleptic-induced dystonia, the latter now being the commonest cause. This is usually an acute effect but may on occasion be seen as a consequence of chronic therapy (tardive oculogyric crisis).

Treatment of acute neuroleptic-induced dystonia is with either parenteral benzodiazepine or an anticholinergic agent (procyclidine, benztropine, trihexyphenidyl).
[Cross References: DYSKINESIA; DYSTONIA]

Oculomotor Nerve Palsy
Oculomotor (III) nerve palsy produces:

- Ptosis: weakness of levator palpebrae superioris, +/- Müller's muscle;
- Mydriasis: impaired parasympathetic outflow to the pupil ("internal ophthalmoplegia");
- Diplopia: weakness of medial rectus, inferior rectus, superior rectus, and inferior oblique muscles causing the eye to point "down and out" (external ophthalmoplegia; the presence of intorsion confirms integrity of superior oblique muscle/trochlear (IV) nerve function.

Changes may be complete or partial. It is suggested that individual fascicles within the oculomotor nerve are arranged topographically, with superior division lesions affecting superior rectus and levator palpebrae superiorirs, medial lesions affecting medial rectus.

Oculomotor nerve palsies may be distinguished as "pupil involving" or "pupil sparing" (*q.v.*), the former implying a "surgical", the latter a "medical" cause, but this distinction only holds for complete palsies. Incomplete palsies are more likely to be of "surgical" origin (e.g posterior communicating artery aneurysm). Imaging is the appropriate management if in doubt. Transtentorial herniation due to raised intracranial pressure may cause an oculomotor nerve palsy due to stretching of the nerve, a "false localising sign".
[Cross References: DIPLOPIA; "FALSE LOCALISING SIGNS"; MYDRIASIS; OPHTHALMOPARESIS, OPHTHALMOPLEGIA; PTOSIS; PUPIL SPARING]

Oculovestibular Response
- see CALORIC TESTING

One-And-A-Half Syndrome

The one-and-a-half syndrome consists of an ipsilateral horizontal gaze palsy and an ipsilateral internuclear ophthalmoplegia, such that the only preserved horizontal eye movement is abduction in one eye; vertical movements and convergence are spared. This results from a brainstem lesion which involves both the abducens (VI) nerve nucleus or paramedian pontine reticular formation, causing ipsilateral horizontal gaze palsy, and the adjacent medial longitudinal fasciculus, causing internuclear ophthalmoplegia.

In young patients this is most often due to demyelination, in the elderly to brainstem ischaemia. Myasthenia gravis may cause a pseudo-one-and-a-half syndrome.

- Wall M, Wray SH. The one-and-a-half syndrome. A unilateral disorder of the pontine tegmentum: a study of 20 cases and a review of the literature. *Neurology* 1983; **33**: 971-80

[Cross References: GAZE PALSY; INTERNUCLEAR OPHTHALMOPLEGIA]

Ophthalmoparesis, Ophthalmoplegia

Ophthalmoparesis is a paralysis, ophthalmoplegia a weakness or limitation, of eye movements. Causes may be central (CNS pathways), or peripheral (cranial nerve nuclei, cranial nerves, neuromuscular junction, extraocular muscles).

A distinction is sometimes drawn between *external ophthalmoplegia,* weakness of the extraocular muscles of central, neuromuscular, or myopathic origin (*e.g.* chronic progressive external ophthalmoplegia (CPEO), a mitochondrial disorder); and *internal ophthalmoplegia,* fixity of the pupil (iridoplegia) and ciliary apparatus. Hence in an oculomotor (III) nerve palsy there may be both internal and external ophthalmoplegia.

[Cross References: DIPLOPIA; INTERNUCLEAR OPHTHALMOPLEGIA; MIOSIS; MYDRIASIS; OCULOMOTOR NERVE PALSY; PUPIL SPARING]

Opisthotonos

Opisthotonos is an abnormal posture consisting of arching of the back and extension of the limbs such that the body may be supported just on the head and ankles. Opisthotonos may be seen in:

- Coma; decerebrate rigidity
- basilar meningitis
- hydrocephalus
- structural lesions of the posterior fossa
- acute drug- (neuroleptic-) induced dystonic reaction; or chronic feature of tardive dystonia
- tetanus
- pseudoseizures

As in decerebrate rigidity, Opisthotonos may reflect unopposed extensor tone from the intact vestibular nuclei released from supratentorial control.

[Cross References: COMA; DECEREBRATE RIGIDITY]

Oppenheim's Sign

Oppenheim's sign is a variant method for eliciting the plantar respone, by application of heavy pressure to the anterior surface of the tibia, for example with the thumb, moving from patella to ankle. Extension of the hallux (upgoing plantar response, Babinski's sign) is pathological. Like Chaddock's sign, Oppenheim's sign always postdates the development of Babinski's sign as a reliable indicator of corticospinal pathway (upper motor neurone) pathology.

- Van Gijn J. *The Babinski sign: a centenary.* Utrecht: Universiteit Utrecht, 1996

[Cross References: BABINSKI'S SIGN; CHADDOCK'S SIGN; PLANTAR RESPONSE; UPPER MOTOR NEURONE SYNDROME]

Opsoclonus

Opsoclonus, or saccadomania, is an eye movement disorder characterized by involuntary bursts of polydirectional saccades (sometimes with a horizontal preference) without an intersaccadic interval (*cf.* square-wave jerks). Like ocular flutter, opsoclonus may be accurately diagnosed with oculography.

Opsoclonus reflects mesencephalic or cerebellar disease affecting the omnipause cells which exert tonic inhibition of the burst neurones which generate saccades.

Causes of opsoclonus include:

- paraneoplasia: in children with neuroblastoma (Kinsbourne's syndrome); in adults the opsoclonus-myoclonus syndrome is most commonly associated with small-cell lung cancer but it may also occur in association with breast cancer in which case onconeural antibodies (anti-Ri, or type 2 anti-neuronal nuclear antibodies [ANNA-2]) may be detected in serum and CSF;
- postinfectious: a monophasic disorder following respiratory or gastrointestinal infection;
- intraparenchymal (especially mesencephalic) lesions, *e.g.* tumour, demyelination, sarcoidosis, metabolic/toxic encephalopathy.

Postinfectious opsoclonus generally remits spontaneously. Of the paraneoplastic disorders, opsoclonus associated with lung and breast tumours persists and the patients decline from their underlying illness; neuroblastoma associated opsoclonus may be steroid responsive.

[Cross References: OCULAR FLUTTER; SACCADIC INTRUSION; SQUARE-WAVE JERKS]

Optic Aphasia

Optic aphasia is a visual modality-specific naming disorder. It has sometimes been grouped with associative visual agnosia, but these patients are not agnosic since they can demonstrate recognition of visually-presented stimuli by means other than naming, *e.g.* gesture. Moreover, these patients are not handicapped by their deficit in everyday life, whereas agnosic patients are often functionally blind. Objects that are semantically related can be appropriately sorted, indicating intact semantics. This is not simply anomia, since the deficit is specific to visual stimuli; objects presented in tactile modality, by sound, or by spoken definition can be named. Naming errors are often semantic, and perseverations (*"conduit d'approche"*) are common. Perception

is intact, evidenced by the ability to draw accurately objects which cannot be named. Reading is poorly performed.

Optic aphasia is associated with unilateral lesions of the left occipital cortex and subjacent white matter.

The neuropsychological explanation of optic aphasia is unclear. It may be a mild type of associative visual agnosia, despite the differences.

- Farah MJ. *Visual agnosia: disorders of object recognition and what they tell us about normal vision.* Cambridge: MIT Press 1995
- Lhermitte F, Beauvois MF. A visual-speech disconnexion syndrome: report of a case with optic aphasia, agnosic alexia and colour agnosia. *Brain* 1973; **96**: 695-714
[Cross References: ANOMIA; *CONDUITD'APPROCHE;* VISUAL AGNOSIA]

Optic Ataxia

Optic ataxia is impaired voluntary reaching for a visually presented target, with misdirection and dysmetria. It may resemble cerebellar ataxia. Tactile search with the palm and fingers may be undertaken in searching for an object, using somatosensory cues to compensate for the lack of visual information.

Optic ataxia occurs with lesions of the superior parietal lobule; the primary visual cortex is intact. It is one feature, along with psychic paralysis of gaze ("sticky fixation") and simultanagnosia (visual disorientation), of Balint's syndrome in which there is some evidence for parieto-occiptial (and possibly frontal) lobe dysfunction (disconnection).

[Cross References: ATAXIA; BALINT'S SYNDROME; DYSMETRIA; SIMULTANAGNOSIA]

Optic Atrophy

Optic atrophy is pallor of the optic nerve head as visualized by ophthalmoscopy. The temporal disc may appear pale in a normal fundus, so that optic atrophy can only be confidently diagnosed when there is also nasal pallor.

Optic atrophy may be the consequence of any optic neuropathy which causes optic nerve damage leading to gliotic change of the optic nerve head. The appearance of optic atrophy is non-specific with respect to aetiology. Common causes include previous optic neuritis and chronic papilloedema, but retinal lesions, optic chiasm and optic tract pathologies can all produce optic atrophy (*e.g.* inherited optic neuropathies, tobacco-alcohol amblyopia; vitamin $\mathbf{B}_{12}$ deficiency).

[Cross References: DISC SWELLING; PAPILLOEDEMA]

Optokinetic Nystagmus (OKN)

Also known as **Optokinetic Response**

Optokinetic nystagmus (OKN) is familiar to anyone who has watched a railway passenger observing passing telegraph poles from the window of a moving train: OKN is an involuntary rhythmic eye movement induced by observing moving stimuli. In clinical practice a striped drum serves to test both visual pursuit and saccades. Rotation of the stripe to the left produces leftward pursuit, followed by a compensatory saccade to the right, followed by pursuit to the left of the next stripe, with another compensatory saccade, and so on. Hence, OKN is a physiological nystagmus.

Parietal hemisphere lesions (vascular or neoplastic) typically impair OKN. Testing for OKN may be useful in patients with suspected hysterical visual loss, since OKN cannot occur unless visual function is present; the response is lost in blindness. An internuclear ophthalmoplegia may be made more evident by testing OKN.
[Cross References: CORTICAL BLINDNESS; INTERNUCLEAR OPHTHALMOPLEGIA; NYSTAGMUS; SACCADE; VESTIBULO-OCULAR REFLEXES]

Orofacial Dyspraxia
Orofacial dyspraxia, or buccofacial dyspraxia, is an inability to make voluntary, learned, movements with the orofacial musculature, such as blowing out a match, kissing, licking the lips.

Recognised causes of orofacial dyspraxia include:

- transient accompaniment of Broca's aphasia, conduction aphasia, and transcortical motor aphasia of cerebrovascular origin;
- trauma to pre-Rolandic area just above the Sylvian fissure;
- in some patients with primary progressive aphasia; a related but distinct condition of "progresive loss of speech output with orofacial dyspraxia" has been described.

Clinical and imaging studies show a strong correlation between orofacial dyspraxia and lesions in the frontal operculum; it may also occur with subcortical lesions involving periventricular and/or peristriatal white matter as well as the basal ganglia.

- Tyrrell PJ, Kartsounis LD, Frackowiak RSJ, Findley LJ, Rossor MN. Progressive loss of speech output and orofacial dyspraxia associated with frontal lobe hypometabolism. *Journal of Neurology, Neurosurgery and Psychiatry* 1991; **54**: 351-7
[Cross References: APRAXIA]

Orthostatic Hypotension
Orthostatic hypotension or postural hypotension is the finding of a persistent drop in blood pressure on standing, defined as a greater than 20 mmHg fall in systolic pressure and/or a 5 mmHg fall in diastolic pressure one minute after a change from the supine to the upright position. Normally there is a drop in blood pressure of lesser magnitude on standing but this is usually quickly compensated. To demonstrate orthostatic hypotension, it may be necessary to measure blood pressure not only on immediate standing but also after two to ten minutes, since the fall may be delayed. Measuring blood pressure automatically on a tilt table is also helpful in diagnosing orthostatic hypotension.

Symptoms which may be associated with orthostatic hypotension include exercise-induced or postprandial light-headedness, transient visual loss, blackouts, and pain in a "coathanger" distribution across the shoulders. There may be supine hypertension and reversal of the normal circadian blood pressure rhythm (normally lower at night), with increased frequency of micturition at night. Other features of autonomic dysfunction include dry eyes and dry mouth (xerophthalmia, xerostomia), a tendency to constipation, and lack of penile erections.
Orthostatic hypotension may be found in:

- pure autonomic neuropathy;

- neurodegenerative disorders such as multiple system atrophy, Parkinson's disease, dementia with Lewy bodies;
- phaeochromocytoma;
- other causes of autonomic neuropathy (*e.g.* Guillain-Barré syndrome, amyloidosis).

However, the commonest cause of orthostatic hypotension in hospital practice is probably dehydration or overzealous treatment with anti-hypertensive or diuretic agents.

Treatments for pure autonomic failure encompass both non-pharmacological approaches (*e.g.* increased salt intake, head-up bed tilt, wearing a G-suit) and pharmacological therapies, including fludrocortisone, ephedrine, and midodrine.

- Mathias CJ, Kimber JR. Treatment of postural hypotension. *Journal of Neurology, Neurosurgery and Psychiatry* 1998; **65**: 285-9
[Cross References: NEUROPATHY; PARKINSONISM; XEROPHTHALMIA, XEROSTOMIA]

Oscillopsia

Oscillopsia is an illusory movement of the environment due to excessive slip of images on the retina ("retinal slip") during active or passive head movement, producing a complaint of blurring, jumping, or oscillation of the visual representation of the environment. Oscillopsia is most often due to acquired bilateral loss of vestibular function (loss of the vestibulo-ocular reflexes). Other recognised causes of oscillopsia include:

- acquired nystagmus;
- superior oblique myokymia;
- other ocular oscillations.

Oscillopsia does not occur in congenital nystagmus, nor in opsoclonus, presumably due to the operation of the visual suppression mechanism which normally operates during saccadic eye movements.

Oscillopsia may be treated with clonazepam; if due to acquired pendular nystagmus anticholinesteraes or alcohol may help.

- Leigh RJ. Oscillopsia: impaired vision during motion in the absence of the vestibulo-ocular reflex. *Journal of Neurology, Neurosurgery and Psychiatry* 1998; **65**: 808
[Cross References: MYOKYMIA; NYSTAGMUS; OPSOCLONUS; VESTIBULO-OCULAR REFLEXES]

Overflow

- see DYSTONIA; SYNKINESIA, SYNKINESIS

P

Palatal Myoclonus
Palatal myoclonus, also known as palatal tremor, is a focal myoclonic syndrome characterized by rhythmic, unilateral or bilateral, palatal contractions which continue during sleep. This may be asymptomatic, or there may be a clicking sound in the inner ear (especially in essential palatal myoclonus). There may be associated contractions of external ocular muscles (oculopalatal myoclonus), larynx, neck, diaphragm, trunk, and limbs, which may bring the palatal myoclonus to attention. Palatal myoclonus may be accompanied by pendular nystagmus and oscillopsia.

Palatal myoclonus is associated with hypertrophy of the inferior olivary nucleus, evident radiologically and pathologically. This is a consequence of a lesion in the dentato-olivary pathway which leads to transsynaptic degeneration and hypermetaboilsm of the olivary nucleus. Although many cases are essential/idiopathic, recognised symptomatic causes of palatal myoclonus include vascular lesions, trauma, neoplasia, demyelination, epilepsy and, rarely, adult-onset Alexander's disease.

Drug treatment of palatal myoclonus is often unsuccessful, although reports of benefit with 5-hydroxytryptophan, carbamazepine, sodium valproate, clonazepam, baclofen, and even sumatriptan have appeared. Botulinum toxin injections may also help.
[Cross References: MYOCLONUS; NYSTAGMUS; OSCILLOPSIA; TINNITUS]

Palatal Reflex
- see GAG REFLEX

Palilalia
Palilalia is a disorder of articulation characterized by the involuntary repetition of syllables within a word, whole words, or phrases (hence a reiterative speech disorder); the term stutter may be used for repetition of single syllables, and the term palilogia has sometimes been used for the repetition of phrases to distinguish this from palilalia. These phenomena may be encountered in:

- Parkinson's disease (along with bradylalia, slowness of speech)
- Steele-Richardson-Olszewski syndrome
- Gilles de la Tourette syndrome (along with vocal and motor tics)
- Pick's disease, as part of the so-called PES syndrome (palilalia, echolalia, stereotypy) or the PEMA syndrome (palilalia, echolalia, mutism, amimia)
- late stages of Alzheimer's disease
- postencephalitic parkinsonism (von Economo's disease)
- Fahr's disease (bilateral basal ganglia calcification)
- as a normal finding in children below the age of about six

In pathological states, palilalia may reflect difficulty in set shifting, as seen in frontal lobe (frontal convexity) syndromes.

[Cross References: BRADYLALIA; ECHOLALIA; FRONTAL LOBE SYNDROMES; HYPOMIMIA; MUTISM; PARKINSONISM; STEREOTYPY; STUTTER; TIC]

Palinopsia

Palinopsia is an illusory visual phenomenon characterized by the persistence or recurrence of visual images immediately after the stimulus has been removed, hence visual perseveration. This is distinct from the physiological after-image. It may be associated with polyopia. The description of the symptom may lead to it being mistaken for diplopia (pseudodiplopia).

Palinopsia occurs most frequently in the context of a left homonymous hemianopia, secondary to right occipitotemporal or occipitoparietal lesions: these may be vascular, neoplastic, metabolic, ictal, or drug- or toxin-induced (*e.g.* carbon monoxide poisoning). It has also been described with retinal and optic nerve disease, and in normal individuals.

- Michel EN, Troost BT. Palinopsia: cerebral localization with computed tomography. *Neurology* 1980; **30**: 887-9
- Pomeranz HD, Lessell S. Palinopsia and polyopia in the absence of drugs or cerebral disease. *Neurology* 2000; **54**: 855-9

[Cross References: HEMIANOPIA; ILLUSION; PERSEVERATION; POLYOPIA]

Palmomental Reflex

The palmomental reflex consists of contraction of the mentalis muscle induced by stroking the ipsilateral palm with a blunt object. It is observed in about one quarter of normal adults, and is very common in the normal elderly, but may indicate damage to the contralateral paracentral cortex or its connections. Hence, it may be considered a frontal release sign or primitive reflex, but is less specific than the grasp reflex.

[Cross References: FRONTAL RELEASE SIGNS]

Papilloedema

Papilloedema is swelling (oedema) of the optic nerve head due to raised intracranial pressure (*cf.* other causes of disc swelling, which may cause pseudopapilloedema).

A number of stages of papilloedema are described: in the acute stage, the only findings may be oedema at the superior and inferior poles of the disc, absence of spontaneous venous pulsation, and enlargement of the blind spot. As papilloedema progresses the whole disc is involved and splinter heamorrhages may be evident at the disc margin. These early stages may be asymptomatic, or may be associated with transient losses of vision (obscurations), often provoked by activities or movements which further raise intracranial pressure. Enlargement of the blind spot and constriction of the visual field may be evident, but visual acuity is often unimpaired (*cf.* disc swelling due to papillitis). Chronic papilloedema produces gliosis of the optic nerve head and eventually optic atrophy ("sequential optic atrophy") with nerve fibre damage and permanent visual field defects.

[Cross References: BLIND SPOT; DISC SWELLING; OBSCURATIONS; OPTIC ATROPHY; PSEUDOPAPILLOEDEMA; SCOTOMA; VENOUS PULSATION]

Paraballismus
- see BALLISM, BALLISMUS; HEMIBALLISMUS

Paradoxical Diaphragm Movement
The normal movement of the diaphragm (*i.e.* down in inspiration, causing outward abdominal wall movement) may be reversed (paradoxical) in conditions which cause diaphragm weakness (*i.e.* inward abdominal wall movement on inspiration), *e.g.* Guillain-Barré syndrome, acid-maltase deficiency, phrenic nerve injury. This may be detectable clinically or by X-ray screening of the diaphragm; vital capacity is lower when lying compared to standing. Paradoxical diaphragm movement is a potentially alarming sign since it may indicate incipient respiratory failure.
[Cross References: MYOPATHY]

Paraesthesia (CHECK)
Paraesthesia is an abnormal sensation, often described as a tingling sensation, or likened to "pins and needles" or electricity, pricking, or even crawling (formication), *i.e.* positive sensory symptoms. The sensation is not pleasant but nor is it painful (*cf.* dysaesthesia). Some patients may describe this sensation as "numbness" or "deadness", in which case care needs to be taken to differentiate it from anaesthesia (*i.e.* a negative phenomenon). Some authorities reserve the term for spontaneous rather than evoked positive sensory phenomena, as a distinction from dysaesthesia.

Paraesthesia is a feature of neuropathy, and may occur in the distribution of a compressed or entrapped nerve, perhaps reflecting the mechanosensitivity of nerves in this situation (*e.g.* Phalen's sign, Tinel's sign). Paraesthesia is a more reliable indicator of the diagnosis of neuropathy than pain. Paraesthesia may also be provoked by hyperventilation (especially perioral, hands and feet [acroparaesthesia]). Central lesions may also produce paraesthesia (*e.g.* Lhermitte's sign).
[Cross References: ANAESTHESIA; DYSAESTHESIA; LHERMITTE'S SIGN; PHALEN'S SIGN; TINEL'S SIGN]

Paragrammatism
- see WERNICKE'S APHASIA

Paragraphia
- see AGRAPHIA

Parakinesia
Parakinesia has been used to describe:

- a volitional purposeful act performed to camouflage or draw attention away from an involuntary movement, such as chorea;
- strange movements of psychogenic origin. It should be remembered that many movements thought in years gone by to conform to this definition have subsequently been recognised to have an organic basis (*e.g.* klazomania).

[Cross References: CHOREA, CHOREOATHETOSIS; DYSKINESIA; KLAZOMANIA]

Paralexia
- see ALEXIA

Paralysis
Paralysis is a total loss of power to move a body part; equivalent to the suffix -plegia. The use of the word has not been entirely consistent, for example *paralysis agitans* originally used by Parkinson to describe the disease which now bears his name.

The periodic paralyses are a group of conditions characterized by episodic muscular weakness and stiffness (myotonia) associated with mutations in the skeletal muscle voltage-gated sodium and calcium ion channel genes (channelopathies).
[Cross References: MYOTONIA; PLEGIA]

Paramnesia
Paramnesia is a false memory. This may be neurological or psychiatric in origin. Relation of paramnesias as the truth occurs in confabulation.
[Cross References: AMNESIA; CONFABULATION; REDUPLICATIVE PARAMNESIA]

Paramyotonia
Paramyotonia is similar to myotonia in that muscle does not relax normally following contraction (voluntary, percussion), which may prompt a complaint of muscle aching or stiffness, but differs in that repetitive muscle use (*e.g.* exercise) leads to an increased delay in relaxation (worsening stiffness). For example, if a patient with paramyotonia is asked to squeeze the eyelids shut repeatedly, voluntary reopening may initially be possible, but after several attempts the eyes may remain closed for a minute or so. Paramyotonia particularly affects the face and forearms. This type of muscle stiffness may also be sensitive to temperature, being made worse by cooling which may provoke muscle weakness. Weakness may outlast exposure to cold by several hours. In this situation the resting muscle membrane potential is reduced from around -80 mV to -40 mV, at which point muscle fibres are inexcitable (contracture).

Paramyotonia congenita is a channelopathy affecting the α-subunit of the sodium channel. Mutations in the same gene have been documented in hyperkalaemic periodic paralysis and K^+-aggravated myotonia.

- Davies NP, Eunson LH, Gregory RP, Mills KR, Morrison PJ, Hanna MG. Clinical, electrophysiological, and molecular genetic studies in a new family with paramyotonia. *Journal of Neurology, Neurosurgery and Psychiatry* 2000; **68**: 504-7 [erratum: *Journal of Neurology, Neurosurgery and Psychiatry* 2000; **69**: 139]
- Ebers GC, George AL, Barchi RL, *et al.* Paramyotonia congenita and hyperkalaemic periodic paralysis are linked to the adult muscle sodium channel gene. *Annals of Neurology* 1991; **30**: 810-6
[Cross References: CONTRACTURE; MYOTONIA; PARALYSIS]

Paraparesis
Paraparesis is a weakness of the lower limbs, short of complete weakness (paraplegia, *q.v.*). This may result from lesions anywhere from cerebral cortex (frontal, parasagittal lesions) to peripheral nerves, producing either an upper motor neurone (spastic) or

lower motor neurone (flaccid) picture. A spinal cord lesion (myelopathy) is probably the commonest cause.
[Cross References: FLACCIDITY; MYELOPATHY; PARAPLEGIA; SPASTICITY]

Paraphasia
Paraphasias are a feature of aphasias (disorders of language), particularly (but not exclusively) fluent aphasias resulting from posterior dominant temporal lobe lesions (*cf.* anterior lesions which tend to produce non-fluent aphasias with agrammatism). Paraphasias may be described as:

~ *literal/phonemic* - impaired phonology (*i.e.* sound based), approximations to real words, or neologisms (*e.g.* "sitter" for "sister"); they may be encountered in Broca's aphasia and conduction aphasia, when the patient may recognise them to be errors, as well as in Wernicke's aphasia;
~ *semantic/categoric* - a failure of the semantic system, *i.e.* understanding the referential meaning of words (*e.g.* "orange" for "apple"); may be observed in Wernicke's aphasia.

Patient's with Wernicke's aphasia often seem unaware of both literal and semantic paraphasias due to a failure of self-monitoring of output.
[Cross References: APHASIA; BROCA'S APHASIA; CONDUCTION APHASIA; JARGON APHASIA; NEOLOGISM; WERNICKE'S APHASIA]

Paraplegia
Paraplegia is a total weakness (paralysis) of the lower limbs (*cf.* paraparesis). This may result from lower motor neurone lesions involving multiple nerve roots and/or peripheral nerves (e.g. paraparetic Guillain-Barré syndrome) producing a flaccid, areflexic paraplegia; but more commonly it is due to upper motor neurone lesions interrupting corticospinal pathways (corticospinal tract, vestibulospinal tract, reticulospinal tracts, and other extrapyramidal pathways), most usually in the spinal cord. The latter may acutely produce a flaccid areflexic picture ("spinal shock"), but later this develops into an upper motor neurone syndrome (hypertonia, clonus, hyperreflexia, loss of superficial reflexes [*e.g.* abdominal, cremasteric reflexes] and Babinski's sign) with possible lower motor neurone signs at the level of the lesion; bladder involvement is common (urinary retention). Because of the difficulty in distinguishing whether an acute paraplegia is of LMN or UMN origin, imaging to exclude potentially reversible cord compression is mandatory.
Recognised causes of paraplegia of upper motor neurone origin include:

- traumatic section of the cord;
- cord compression;
- inflammatory lesions: acute transverse myelitis of viral origin, multiple sclerosis, neuromyelitis optica (Devic's syndrome);
- ischaemic lesions; anterior spinal artery syndrome, venous infarction of the cord.

In paraplegia of upper motor neurone origin, enhanced flexion defence reflexes ("flexor spasms") may occur, producing hip and knee flexion, ankle and toe

dorsiflexion. Eventually such flexor responses may become a fixed flexion deformity with secondary contractures ("paraplegia in flexion"). Prevention of this situation may be possible by avoiding spasms, which are often provoked by skin irritation or ulceration, constipation, bladder infection, and poor nutrition. Physiotherapy and treatments such as baclofen, dantrolene and tizanidine may be used; botulinum toxin injections may be helpful for focal spasticity.

"Paraplegia in extension", with extension at the hip and knee, may be seen with incomplete or high spinal cord lesions.

- Johnston RA. Acute spinal cord compression. In: Hughes RAC (ed.). *Neurological Emergencies.* London: BMJ Publishing 1997 (2nd edition): 272-94
- Passmore AP, Taylor IC, McConnell JG. Acute Guillain-Barré syndrome presenting as acute spinal cord compression in an elderly woman. *Journal ofthe Royal Society of Medicine* 1990; **83**: 333-4

[Cross References: ABDOMINAL REFLEXES; AREFLEXIA; BABINSKI'S SIGN; CLONUS; CONTRACTURE; CREMASTERIC REFLEX; FLACCIDITY; HYPER-REFLEXIA; HYPERTONIA, HYPERTONUS; LOWER MOTOR NEURONE SYN-DROME; MYELOPATHY; PARAPARESIS; SPASTICITY; UPPER MOTOR NEURONE SYNDROME; URINARY RETENTION]

Paratonia

- see *GEGENHALTEN*

Paresis

Paresis denotes a weakness which is less than total paralysis (-plegia), which may be of upper or lower motor neurone origin. Various prefixes denote the location of such weakness, *e.g.* hemiparesis, monoparesis, ophthalmoparesis, paraparesis, quadripar-esis (*q.v.*).

Since localised pain may prevent strong muscular exertion, apparent weakness in such circumstances may be labelled "algesic pseudoparesis".

[Cross References: LOWER MOTOR NEURONE SYNDROME; UPPER MOTOR NEURONE SYNDROME; WEAKNESS]

Parinaud's Syndrome

Parinaud's syndrome, or the dorsal midbrain syndrome, consists of paralysis of vertical gaze, especially upgaze, with or without mydriasis, loss of pupillary light reflexes (light-near dissociation), loss of convergence, convergence or retraction nystagmus, and lid retraction (Collier's sign).

This results from dorsal midbrain lesions, such as pineal tumours, which affect the pretectum and posterior commissure and so interfere with conjugate eye movements in the vertical plane.

- Parinaud H. Paralysie des mouvements associés des yeux. *Archives de Neurologie Paris* 1883; **5**: 145-72

[Cross References: COLLIER'S SIGN; LIGHT-NEAR PUPILLARY DISSOCIA-TION; NYSTAGMUS; SUPRANUCLEAR GAZE PALSY]

Parkinsonism

Parkinsonism is a clinical syndrome characterized by the presence of some or all of the following features; there is overlap with so-called akinetic-rigid syndromes in which these features predominate:

- akinesia, hypokinesia (*sine qua non*)
- rigidity: consistent (leadpipe) or jerky (cogwheeling; Negro's sign)
- bradykinesia
- hypometria
- tremor, usually at rest, of frequency 3.5-7.0 Hz, "pill-rolling" type; there may sometimes be an additional action component to the tremor, and very occasionally there is exclusively an action tremor
- stooped posture: forward flexion of trunk, flexion of knees, elbows; "simian posture"
- impaired postural reflexes, with or without a history of falls
- mask-like facies, poverty of facial expression (hypomimia)
- reduced blink rate (this may be a particular feature of the Steele-Richardson-Olszewski syndrome)
- hypophonic, monotonic voice (hypokinetic dysarthria)
- widened palpebral fissure (Stellwag's sign)
- seborrhoea
- sialorrhoea
- festinant (shuffling) gait
- micrographia
- dystonic postures, *e.g.* striatal toe
- apraxia
- akathisia
- cognitive impairment (usually of frontal-subcortical type)
- autonomic dysfunction, especially orthostatic hypotension

Conventionally parkinsonism is viewed as a disorder of the extrapyramidal system producing "extrapyramidal signs", although this term has limitations: despite the fact that some of the cardinal features of parkinsonism (bradykinesia, rigidity, postural instability, tremor) result from pathology in the basal ganglia, particularly affecting dopaminergic pathways, other features may reflect cortical involvement, at least in part (*e.g.* apraxia, micrographia).

The incidence of parkinsonism increases dramatically with age; it is also associated with an increased risk of death, particularly in the presence of a gait disturbance.

The differential diagnosis of parkinsonism is broad, and includes:

- idiopathic Parkinson's disease
- multiple system atrophy
- Steele-Richardson-Olszewski syndrome (also known as progressive supranuclear palsy, although there are other causes of a progressive supranuclear palsy)
- corticobasal degeneration
- drug-induced parkinsonism (*e.g.* neuroleptics, MPTP)

- toxin-induced parkinsonism (*e.g.* manganese, carbon monoxide)
- Wilson's disease (hepatolenticular degeneration)
- dementia with Lewy bodies
- neuroleptic malignant syndrome
- normal pressure hydrocephalus
- "arteriosclerotic parkinsonism", resulting from multiple subcortical infarcts
- Huntington's disease, especially juvenile onset
- post-encephalitic parkinsonism (encephalitis lethargica, Von Economo's disease)
- dementia pugilistica, post-traumatic parkinsonism
- systemic lupus erythematosus
- Sjögren's disease
- Hypoparathyroidism
- Parkinsonism-dementia complex of Guam

Obsessive slowness also enters the differential diagnosis but typical parkinsonian features (akinesia, rigidity) are not present in this condition.

It is crucial not to miss the diagnosis of Wilson's disease, although rare, since in the early stages this disorder is reversible with copper chelation therapy; hence copper and caeruloplasmin should be checked in all patients with young-onset (under age 50) parkinsonism (and dystonia).

Response to levodopa therapy is only reliably seen in idiopathic Parkinson's disease, although some patients with multiple system atrophy or Steele-Richardson-Olszewski syndrome may benefit. The features particularly responsive in Parkinson's disease are bradykinesia and rigidity; tremor is less reliably helped.

- Bennett DA, Beckett LA, Murray AM, *et al.* Prevalence of parkinsonian signs and associated mortality in a community population of older people. *New England Journal of Medicine* 1996; **334:** 71-6
- Gardner-Thorpe C. *James Parkinson 1755-1824.* Exeter: A Wheaton & Co. Ltd 1987 [includes facsimile of Parkinson's book on the shaking palsy]
- Litvan I, Agid Y, Calne D, *et al.* Clinical research criteria for the diagnosis of progressive supranuclear palsy (Steele-Richardson-Olszewski syndrome): report of the NINDS-SPSP international workshop. *Neurology* 1996; **47**: 1-9
- Oertel WH, Quinn NP. Parkinsonism. In: Brandt T, Caplan LR, Dichgans J, Diener HC, Kennard C (eds.). *Neurological disorders: course and treatment.* San Diego: Academic Press 1996:715-72

[Cross References: BRADYKINESIA; DYSARTHRIA; DYSPRAXIA; DYSTONIA; HYPOKINESIA; HYPOMIMIA; HYPOPHONIA; MASK-LIKE FACIES; MICROGRAPHIA; ORTHOSTATIC HYPOTENSION; POSTURAL REFLEXES; RIGIDITY; SEBORRHOEA; SIALORRHOEA; STRIATAL TOE; SUPRANUCLEAR GAZE PALSY; TREMOR]

Parosmia

Parosmia is a false smell, *i.e.* the subjective sensation of a smell which does not exist. Such smells are usually unpleasant (cacosmia), and may be difficult for the patient to define. Causes include purulent nasal infections or sinusitis, and partial recovery

following transection of olfactory nerve fibres after head injury. Transient parosmia may presage seizures of temporal lobe cortical origin (olfactory aura), particularly involving the medial (uncal) region.
[Cross References: AURA; SEIZURES]

Parry-Romberg Syndrome
- see HEMIFACIAL ATROPHY

Past-pointing
- see DYSMETRIA

Patellar Reflex
- see REFLEXES

Pathological Crying/Laughter
Pathological laughter and pathological crying (PLC) have been defined as reflecting an incongruence of mood (subjective feeling) and expression or affect ("objective", observed), such that patients laugh involuntarily though not happy, or cry though not sad. There may be a sense that the patient is struggling against these displays of emotion (*cf.* emotional lability). These are ascribed to a loss (release) of the normal inhibition of the motor component of facial expression *(i.e.* cortical-subcortical disinhibition). PLC may occur in the context of a pseudobulbar palsy ("pseudobulbar affect") but not invariably so. PLC differs from emotional lability in that there is congruence of mood and affect in the latter, although sudden fluctuations and exaggerated emotional expression are common to both, suggesting a degree of overlap.
PLC may occur in:

- Multiple sclerosis: crying > laughing; related to intellectual impairment (more extensive brain involvement, but not brainstem);
- Alzheimer's disease;
- Stroke: PLC may be the harbinger of brainstem stroke;
- Motor neurone disease;
- Head injury;
- Gelastic epilepsy.

Effective treatments for PLC include:

- Amitriptyline
- Levodopa
- Amantadine
- Serotonin reuptake inhibitors: fluoxetine, citalopram.

- Feinstein A. *The clinical neuropsychiatry of multiple sclerosis.* Cambridge: Cambridge University Press 1999: 65-79
- Robinson RG. *The clinical neuropsychiatry of stroke: cognitive, behavioral and emotional disorders following vascular brain injury.* Cambridge: Cambridge University Press 1998: 455-71
[Cross References: AUTOMATISM; EMOTIONAL LABILITY; PSEUDOBULBAR PALSY]

Pelvic Thrusting
Pelvic thrusting may be a feature of seizures of frontal lobe origin; occasionally it may occur in temporal lobe seizures. Pelvic thrusting also occurs in pseudoseizures, particularly those of the "thrashing" variety.

- Geyer JD, Payne TA, Drury I. The value of pelvic thrusting in the diagnosis of seizures and pseudoseizures. *Neurology* 2000; **54**: 227-9
[Cross References: AUTOMATISM; SEIZURE]

Percussion Myotonia
Percussion myotonia is the myotonic response of a muscle to a mechanical stimulus, *e.g.* when struck with a tendon hammer. For example, a blow to the thenar eminence may produce involuntary and sustained flexion of the thumb. This response, which may be seen in myotonic dystrophy, reflects the impaired muscle relaxation which characterises myotonia.
[Cross References: MYOTONIA]

Periodic Respiration
Periodic respiration is a cyclical waxing and waning of the depth and rate of breathing (Cheyne Stokes breathing), over about two minutes. It may be observed in unconscious patients with lesions of the deep cerebral hemispheres, diencephalon, or upper pons. Prolonged circulatory time and hypoxaemia may also provoke this form of respiration, but with a shorter cycle.
[Cross References: COMA]

Perseveration
Perseveration is inappropriate, repeated or recurrent, motor behaviour or speech. A number of varieties have been described, associated with lesions in different areas of the brain:

~ "*Stuck-in-sef":* inappropriate maintenance of a current category or framework; thought to reflect a deficit in executive function; associated with frontal lobe (especially frontal convexity) damage, which is associated with an inert, apathetic pattern of behaviour (rather than the disinhibited pattern associated with orbitofrontal damage);
~ "*Recurrent":* unintentional repetition of a previous response to a subsequent stimulus; thought to represent an abnormal post-facilitation of a memory trace; associated with posterior left (dominant) hemisphere damage; commonly seen in aphasics, Alzheimer's disease; this overlaps with "intrusions";
~ "*Continuous":* inappropriate prolongation or repetition of a current behaviour without interruption; thought to represent a deficit of motor output; associated with basal ganglia damage.

Sensory perseveration is also described, *e.g.* palinopsia in the visual system.

- Sandson J, Albert ML. Varieties of perseveration. *Neuropsychologia* 1984; **22**: 715-32
[Cross References: APHASIA; FRONTAL LOBE SYNDROMES; INTRUSION; PALINOPSIA]

Pes Cavus
Pes cavus is a high-arched foot due to equinus (plantar flexion) deformity of the first ray, with secondary changes in the other rays (*i.e.* deformity is more evident on the medial side of the foot in most cases). This may be due to imbalance of muscular forces during development (*e.g.* strong peroneus longus, weak peroneus brevis and tibialis anterior, although the precise pattern may differ with cause), which may be a consequence of neurological disease. This may be genetic, *e.g.* hereditary motor and sensory neuropathy (HMSN, Charcot-Marie-Tooth syndrome), hereditary spastic paraparesis, Friedreich's ataxia, Marfan's syndrome; or due to an early neurological insult, *e.g.* cerebral palsy, paralytic poliomyelitis. Familial pes cavus without other neurological signs also occurs.

Surgical treatment of pes cavus may be necessary, especially if there are secondary deformities causing pain, skin breakdown, or gait problems.
[Cross References: CLAW FOOT]

Phalen's Sign
Phalen's sign is present when tingling (paraesthesia) is experienced in the distribution of the median nerve when the wrist is held in forced flexion (90° for 30-60 seconds); patients may volunteer that they experience such symptoms when carrying heavy items such as shopping bags. Hyperextension of the wrist ("reverse Phalen's manoeuvre") may also reproduce symptoms. These are signs of compression of the median nerve at the wrist (carpal tunnel syndrome). Like other provocative tests (*e.g.* Tinel's sign), the sensitivity of Phalen's sign for this diagnosis is not high compared to electrophysiological testing.

The pathophysiology of Phalen's sign is probably the lower threshold of injured nerves to mechanical stimuli, as for Tinel's sign and Lhermitte's sign.

- Heller L, Ring H, Costeff H, Solzi P. Evaluation of Tinel's and Phalen's signs in the diagnosis of the carpal tunnel syndrome. *European Neurology* 1986; **25**: 40-2
[Cross References: LHERMITTE'S SIGN; PARAESTHESIA; TINEL'S SIGN]

Pharyngeal Reflex
- see GAG REFLEX

Phonagnosia
Phonagnosia is an inability to recognise familiar voices in the absence of hearing impairment; the patient can recognise and understand words and sentences (*cf.* pure word deafness). Phonagnosia is the equivalent in the auditory domain of prosopagnosia in the visual domain.
[Cross References: AGNOSIA; PROSOPAGNOSIA; PURE WORD DEAFNESS]

Phonetic Disintegration
- see APHEMIA

Phonophobia
Phonophobia is a dislike, or fear, of sounds, especially loud sounds, often experienced during a migraine headache.
[Cross References: HYPERACUSIS]

Phosphene
Phosphenes are percepts in one modality induced by an inappropriate stimulus, *e.g.* when pressure is applied to the eyeball, the mechanical stimulus may induce the perception of light. Flashes of light when the eyes are moved have been reported in optic neuritis, presumably reflecting the increased mechanosensitivity of the demyelinated optic nerve; this is suggested to be the visual equivalent of Lhermitte's sign. Noise induced visual phosphenes have also been reported.

- Davis FA, Bergen D, Schauf C, McDonald I, Deutsch W. Movement phosphenes in optic neuritis: a new clinical sign. *Neurology* 1976; **26**: 1100-4
- Lessell JB, Cohen MM. Phosphenes induced by sound. *Neurology* 1979; **29**: 1524-6
[Cross References: LHERMITTE'S SIGN; SYNAESTHESIA]

Photophobia
Photophobia is a dislike, or fear, of light, often experienced with meningitis and other causes of meningeal irritation, and during a migraine headache.
[Cross References: DAZZLE; MENINGISM]

Photopsia
Photopsias are simple visual hallucinations consisting of flashes of light which often occur with a visual field defect. They suggest dysfunction in the inferomedial occipital lobe, such as migraine or an epileptogenic lesion.
[Cross References: AURA; HALLUCINATION]

"Picture Sign"
The "picture sign" is present when a patient believes that individuals seen on the television screen are actually present in the home; indeed they may emerge from the television into the room. This may occur as part of the cognitive disturbance of Alzheimer's disease or dementia with Lewy bodies, or as part of a psychotic disorder. Like the "mirror sign", the "picture sign" is a misidentification phenomenon.
[Cross References: "MIRROR SIGN"]

Pied en griffe
- see CLAW FOOT

"Pinch Sign"
The "pinch sign" is an inability to make a small circle by approximating the distal phalanges of the thumb and index finger, due to weakness of flexor digitorum profundus in the index finger and flexor pollicis longus in the thumb as a consequence of median nerve lesions in the forearm, *e.g.* anterior interosseous syndrome. This results in a pinching posture of thumb and index finger. The "straight thumb sign" may also be present (*q.v.*).
[Cross References: FROMENT'S SIGN; "STRAIGHT THUMB SIGN"]

Plantar Response
The plantar response is most commonly elicited by stroking the sole of the foot with a blunt object The first response of the hallux is the critical observation, which may be

facilitated by having ones line of vision directly above the axis of the toe. The normal response after maturation of the corticospinal tracts (*i.e.* after about three years of age) is for the big toe to flex. An extensor response of the big toe in an adult (Babinski's sign), with or without fanning (abduction) of the other toes (fan sign, *signe de l'éventail),* is a reliable sign of upper motor neurone pathology. Use of the term "negative Babinski's sign" or "negative Babinski response" to mean "flexor plantar response" is incorrect and should not be used. This normal plantar response is a superficial cutaneous reflex, analogous to abdominal and cremasteric reflexes, whereas the pathological response is often accompanied by activity in other flexor muscles. In some individuals the toes do not move at all, in which case the response is labelled as "mute" or absent. Assessment of the response may be confounded by withdrawal of the foot in ticklish individuals. Differentiation from the striatal toe seen in parkinsonian syndromes is also important.

The plantar response may be elicited in a variety of other ways which are not in routine clinical use. Of these, perhaps the most frequently used are Chaddock's sign (application of a stimulus in a circular direction around the external malleolus, or the lateral aspect of the foot from heel to little toe) and Oppenheim's sign (application of heavy pressure to the anterior surface of the tibia from patella to ankle). These may be helpful in ticklish patients who object to having their feet stroked. If upgoing, they suggest a spread of the "receptive field" of the reflex. Babinski's sign is the first to occur in the presence of upper motor neurone pathology.

It is often difficult to form a definite judgement on the plantar response and reproducibility is also questionable. A study of 24 experienced clinicians invited to examine plantar responses "blind" found that the interobserver percentage agreement beyond chance was on average only 16.7% (95% confidence interval [CI] 0.4-33%); intraobserver percentage agreement was a little better (average 59.6%; CI 39.6-79.6%). There remains a persistent belief, particularly amongst trainees, that an experienced neurologist can make the plantar response go which ever way s/he chooses.

- Maher J, Reilly M, Daly L, Hutchinson M. Plantar power: reproducibility of the plantar response. *BMJ* 1992; **304**: 482
- Van Gijn J. *The Babinski sign: a centenary.* Utrecht: Universiteit Utrecht, 1996
[Cross References: ABDOMINAL REFLEXES; BABINSKI'S SIGN; CHADDOCK'S SIGN; CREMASTERIC REFLEX; OPPENHEIM'S SIGN; STRIATAL TOE, UPPER MOTOR NEURONE SYNDROME]

Plegia

Plegia means stillness, implying a complete weakness (or paralysis in common parlance), as in monoplegia, diplegia, ophthalmoplegia, paraplegia, quadriplegia. Hence plegia is a more severe weakness than paresis.
[Cross References: PARESIS]

Plexopathy

Lesions confined to the brachial, lumbar, or sacral plexi may produce a constellation of motor and sensory signs (weakness, reflex diminution or loss, sensory loss) which cannot be ascribed to single or multiple roots (radiculopathy) or peripheral nerves (neuropathy). Lesions may involve the whole plexus (panplexopathy):

~ *Brachial:C5*-T1*;*
~ *Lumbar:* L2-L4;
~ *Sacral:* L5-S3;

or be partial, *e.g.* upper trunk of brachial plexus (C5-C6), producing "waiter's tip" posture (as for C5/C6 root avulsion); lower trunk of brachial plexus (C8-T1; as for C8/T1 root avulsion).

Electrophysiological studies may be helpful in distinguishing plexopathy from radiculopathy: sensory nerve action potentials are reduced or absent in the former, and EMG shows sparing of paraspinal muscles (*cf.* radiculopathy).

Recognised causes of brachial plexopathy include:

- Trauma: Upper plexus: Dejerine-Klumpke paralysis ("waiter's tip" posture); Lower plexus: Erb-Duchenne paralysis (claw hand).
- Inflammation/Idiopathic: brachial neuritis, neuralgic amyotrophy.
- Malignant infiltration, *e.g.* carcinoma of lung (Pancoast), breast, +/- Horner's syndrome; pain a significant symptom.
- Post-radiation (*e.g.* after radiotherapy for malignant breast cancer with axillary spread).
- Tomaculous neuropathy.
- Hereditary neuropathy with liability to pressure palsies (HNLPP).
- Neurogenic thoracic outlet syndrome (rare): cervical rib or C7 transverse process or fibrous band compressing the lower trunk. May be surgically remediable.

Recognised causes of lumbosacral plexopathy include:

- Compression; *e.g.* iliopsoas haematoma (anticoagulation, haemophilia), abscess (tuberculosis); abdominal aortic aneurysm; pregnancy (fetal head in the second stage of labour).
- Neoplasia (direct spread > metastasis).
- Trauma (rare; *cf.* brachial plexopathy).
- Radiation.
- Vasculitis (mononeuritis multiplex much commoner).
- Idiopathic.

Imaging with MRI is superior to CT for defining structural causes of plexopathy.

- Chad DF. Nerve root and plexus disorders. In: Bogousslavsky J, Fisher M (Eds). *Textbook of Neurology.* Boston: Butterworth-Heinemann 1998:491-506
- Taylor BV, Kimmel DW, Krecke KN, Cascino TL. Magnetic resonance imaging in cancer-related lumbosacral plexopathy. *Mayo Clinic Proceedings* 1997; **72**: 823-9

[Cross References: AMYOTROPHY; CLAW HAND; HORNER'S SYNDROME; NERVE THICKENING; NEUROPATHY; RADICULOPATHY]

Polymyoclonus

- see MYOCLONUS

Polyneuropathy

- see NEUROPATHY

Polyopia

Polyopia, or polyopsia, is a visual illusory phenomenon in which a single target is seen as multiple images, most usually double but sometimes higher multiples (*e.g.* entomopia), persisting when looking away from the object. This may be likened to "echoes" of the image, and eye movement may produce a trailing effect. Polyopia may be related to palinopsia.

Polyopia is associated with occipital and occipito-parietal lesions, bilateral or confined to the non-dominant hemisphere, and with drug abuse. It has also been described in disease of the retina and optic nerve, and in normal individuals.

The pathophysiology is unknown; suggestions include a defect of visual fixation or of visual integration; the latter may reflect pure occipital cortical dysfunction.

- Pomeranz HD, Lessell S. Palinopsia and polyopia in the absence of drugs or cerebral disease. *Neurology* 2000; **54**: 855-9

[Cross References: ENTOMOPIA; PALINOPSIA]

Positioning Manoeuvre

- see HALLPIKE MANOEUVRE, HALLPIKE TEST

Post-tetanic Potentiation

- see FACILITATION

Postural Hypotension

- see ORTHOSTATIC HYPOTENSION

Postural Reflexes

Postures such as standing are largely reflex in origin, dependent upon involuntary muscle contraction in anti-gravity muscles. Interference with such reflex activity impairs normal standing. Postural and righting reflexes depend on the integration of labyrinthine, proprioceptive, exteroceptive, and visual stimuli, mostly in the brainstem but also involving cerebral cortex. However, abnormalities in these reflexes are of relatively little diagnostic value except in infants.

One exception is extrapyramidal disease (parkinsonism, Huntington's disease, but not idiopathic dystonia) in which impairment or loss of postural reflexes may be observed. Sudden passive tilting of the body, for example by the examiner pulling on both shoulders when positioned behind a patient who is standing comfortably ("pull test") may provoke repetitive steps backwards (retropulsion, festination) or even *en bloc* falling, due to failure of reflex muscle contraction necessary to maintain equilibrium. Pushing the patient may likewise provoke propulsion or festination, but this manoeuvre is less safe since the examiner will not be placed to catch the patient should they begin to topple over.

[Cross References: DYSTONIA; FESTINANT GAIT, FESTINATION; PARKINSONISM; PROPRIOCEPTION]

Pout Reflex

The pout reflex, or snout reflex, consists of a pouting movement of the lips when lightly tapped with a finger or tendon hammer. Its presence is indicative of impaired

corticobulbar projections, most usually due to cerebrovascular disease or motor neurone disease.
[Cross References: FRONTAL RELEASE SIGNS; PRIMITIVE REFLEXES]

Presbycusis
Presbycusis is a progressive sensorineural hearing loss, especially for high frequencies, developing with increasing age, which may reduce speech discrimination. It is thought to be due to age-related attrition of hair cells in the organ of Corti and/or spiral ganglion neurones.

Presbyopia
Presbyopia is progressive far-sightedness which is increasingly common with increasing age, thought to be due to an age-related impairment of accommodation.

Priapism
Priapism is an unintended, sustained, and usually painful erection of the penis unrelated to sexual activity. It may occur with intramedullary spinal cord lesions (*e.g.* multiple sclerosis) which damage the lumbosacral erection centres. There are also non-neurological causes, such as haematological conditions (sickle cell anaemia, polycythaemia rubra vera) which may cause intrapenile thromboses.

Primitive Reflexes
Reflexes which are normally found in infancy but which disappear with brain maturation during childhood may be labelled as "primitive reflexes" if they re-emerge in adulthood as a consequence of pathological states. Many of these reflexes are seen with frontal lobe pathology (*e.g.* grasp, pout/snout, palmomental, rooting, corneomandibular) and hence may also be known as "frontal release signs". However, the term "primitive reflex" could equally apply to Babinski's sign which is not necessarily frontal in origin.
[Cross References: BABINSKI'S SIGN; CORNEOMANDIBULAR REFLEX; FRONTAL RELEASE SIGNS; GRASP REFLEX; PALMOMENTAL REFLEX; POUT REFLEX; ROOTING REFLEX]

Proprioception
Proprioception sensation, or joint position sense, is knowledge about ones position in space, originating from sensory receptors in skin, muscle, and viscera. Proprioceptive information is carried within the dorsal columns of the spinal cord (more reliably so than vibration sensation, though not necessarily exclusively). Lesions affecting this part of the cord, particularly in the cervical region (*e.g.* subacute combined degeneration of the cord due to vitamin $\mathbf{B}_{12}$ deficiency, tabes dorsalis), lead to impairments of proprioception with sparing of spinothalamic sensations (pin-prick, temperature) producing a dissociated sensory loss. Impairment of proprioception leads to sensory ataxia which may manifest clinically as pseudoathetosis or pseudochoreoathetosis and a positive Romberg's sign.

[Cross References: ATAXIA; DISSOCIATED SENSORY LOSS; PSEUDOATHETOSIS; PSEUDOCHOREOATHETOSIS; ROMBERGISM, ROMBERG'S SIGN; VIBRATION]

Proptosis

Proptosis is forward displacement of the eyeball, an exaggerated degree of exophthalmos. This may be assessed clinically by standing directly behind the patient and gradually tipping the head back, observing when the globe of the eyeball first comes into view; this is most useful for asymmetric proptosis. An exophthalmometer may be used to measure proptosis. Once established, it is crucial to determine whether the proptosis is axial or non-axial. Axial proptosis reflects increased pressure within or transmitted through the cone of extraocular muscles (*e.g.* thyroid ophthalmopathy, cavernous sinus thrombosis), whereas non-axial proptosis suggests pressure from an orbital mass outside the cone of muscles (*e.g.* orbital lymphoma, pseudotumour, mucocele). Pulsatile axial proptosis may be seen in carotico-cavernous fistula, in which case there may be a bruit audible by auscultation over the eye.

Dedicated orbital CT or MRI, the latter with fat-suppression sequences and intravenous gadolinium contrast, may be required to detect intraorbital masses.

[Cross References: EXOPHTHALMOS]

Propulsion

- see FESTINANT GAIT, FESTINATION; POSTURAL REFLEXES

Prosopagnosia

Prosopagnosia is a form of visual agnosia characterized by an inability to recognize previously known human faces or equivalent stimuli (hence, a retrograde defect) and to learn new ones (anterograde defect). As with more pervasive visual agnosia (*q.v.*), this may be:

~ *apperceptive:* due to faulty perceptual analysis of faces; or
~ *associative:* a semantic defect in recognition.

Familiar individuals may be recognized by their voices or clothing; hence, the defect may be one of visually triggered episodic memory. It is important to note that the defect is not limited solely to faces; it may encompass animals ("zooagnosia"), or cars.

Prosopagnosia is often found in association with a visual field defect, most often a left superior quadrantanopia or even hemianopia, although for the diagnosis of prosopagnosia to be made this should not be sufficient to produce a perceptual deficit. Alexia and achromatopsia may also be present, depending on the exact extent of the underlying lesion.

Anatomically, prosopagnosia occurs most often in association with bilateral occipito-temporal lesions involving the inferior and mesial visual association cortices in the lingual and fusiform gyri, sometimes with subjacent white matter. Unilateral non-dominant (right) hemisphere lesions have occasionally been associated with prosopagnosia, and a syndrome of progressive prosopagnosia associated with selective focal atrophy of the right temporal lobe has been reported. Involvement of the

periventricular region on the left side may explain accompanying alexia, and disconnection of the inferior visual association cortex (area V4) may explain achromatopsia.

Pathological causes of prosopagnosia include:

- cerebrovascular disease: this is by far the commonest cause;
- tumour, *e.g.* glioma, extending from one hemisphere to the other via the splenium of the corpus callosum;
- epilepsy (paroxysmal prosopagnosia), due to bilateral foci or spread from one occipital focus to the contralateral hemisphere;
- right temporal lobe atrophy;
- herpes simplex encephalitis, usually as part of an extensive amnesic syndrome (although memory impairment may put this outwith the operational criteria for an agnosia);
- rare cases of developmental prosopagnosia have been described.

- Evans JJ, Heggs AJ, Antoun N, Hodges JR. Progressive prosopagnosia associated with selective right temporal lobe atrophy. A new syndrome? *Brain* 1995; **118**: 1-13
- Farah MJ. *Visual agnosia: disorders of object recognition and what they tell us about normal vision.* Cambridge: MIT Press 1995

[Cross References: ACHROMATOPSIA; AGNOSIA; ALEXIA; HEMIANOPIA; PHONAGNOSIA; QUADRANTANOPIA; VISUAL AGNOSIA; ZOOAGNOSIA]

Prosopoplegia

- see BELL'S PALSY; FACIAL PARESIS

Pseudoachromatopsia

Pseudoachromatopsia is failure on tests of colour vision (*e.g.* pseudoisochromatic plates) not due to central or peripheral achromatopsia, but for example due to visual neglect.

[Cross References: ACHROMATOPSIA; NEGLECT]

Pseudo-Argyll Robertson Pupil

A pseudo-Argyll Robertson pupil shows light-near dissociation of pupillary reactions, but, unlike the "true" Argyll Robertson pupil, there is no miosis or pupil irregularity. Indeed the pupil may be dilated (mydriasis) and resemble a Holmes-Adie pupil. The latter may be differentiated on the basis of its response to dilute (0.2%) pilocarpine: Holmes-Adie pupil results from a peripheral lesion and shows denervation supersensitivity constricting with dilute pilocarpine, whereas the pseudo-Argyll Robertson pupil results from a central lesion and does not respond.

Pseudo-Argyll Robertson pupil has been reported in:

- diabetes mellitus
- multiple sclerosis
- Wernicke's encephalopathy
- sarcoidosis
- tumour

- haemorrhage
- aberrant oculomotor (III) nerve regeneration
- spinocerebellar ataxia type 1 (SCA1)

[Cross References: ARGYLL ROBERTSON PUPIL; HOLMES-ADIE PUPIL; MIOSIS; MYDRIASIS]

Pseudoathetosis

Pseudoathetosis is the name given to athetoid-like movements, most usually of the outstretched fingers and hands, resulting from sensory ataxia (impaired proprioception); it is made worse with the eyes closed. There may also be chorea-like movements (see Pseudochoreoathetosis). Causes include any interruption to the anatomical pathway mediating proprioception, most often lesions in the dorsal cervical cord (*e.g.* multiple sclerosis, subacute combined degeneration of the cord due to vitamin B_{12} deficiency), but also lesions of the large (myelinated) peripheral nerve fibres, and of the parietal lobe.

[Cross References: ATHETOSIS; CHOREA, CHOREOATHETOSIS; PROPRIOCEPTION; PSEUDOCHOREOATHETOSIS]

Pseudobulbar Affect

- see EMOTIONAL LABILITY; PATHOLOGICAL CRYING, PATHOLOGICAL LAUGHTER; PSEUDOBULBAR PALSY

Pseudobulbar Palsy

Pseudobulbar palsy describes bilateral upper motor neurone lesions affecting fibres passing to the cranial nerve nuclei (*cf.* bulbar palsy). This leads to a variety of clinical problems, including:

- difficulty with speech (spastic dysarthria, dysphonia);
- difficulty with swallowing (dysphagia);
- brisk jaw jerk and pout reflex; there may be trismus;
- slow, spastic, tongue movements;
- gag reflex may be depressed or exaggerated.

There may be associated emotional lability, or pathological laughter and crying ("pseudobulbar affect"), and a gait disorder with *marche à petit pas.* There are otherwise few signs in the limbs, aside from brisk reflexes and upgoing plantar responses (Babinski's sign).

Causes of pseudobulbar palsy include:

- Motor neurone disease (in which there may be coincident bulbar palsy)
- Multiple sclerosis
- Bilateral internal capsule lacunar infarctions, widespread small vessel disease (Binswanger's disease)
- Congenital childhood suprabulbar palsy (Worster-Drought syndrome; perisylvian syndrome).

[Cross References: BABINSKI'S SIGN; BULBAR PALSY; DYSARTHRIA; DYSPHAGIA; DYSPHONIA; EMOTIONAL LABILITY; GAG REFLEX; JAW JERK;

MARCHE À PETIT PAS; PATHOLOGICAL CRYING, PATHOLOGICAL LAUGHTER; TRISMUS; UPPER MOTOR NEURONE SYNDROME]

Pseudochoreoathetosis

Pseudochoreoathetosis is the name given to choreoathetoid type involuntary movements, including dystonic movements, which result from a loss or impairment of proprioception. These may be observed with lesions anywhere along the proprioceptive pathways, including parietal cortex, thalamus (may be associated with ataxic hemiparesis and hemihypoaesthesia), spinal cord, dorsal root ganglia (neuronopathy), and mononeuropathy.

- Sharp FR, Rando TA, Greenberg SA, Brown L, Sagar SM. Pseudochoreoathetosis. Movements associated with loss of proprioception. *Archives of Neurology* 1994; **51:** 1103-9

[Cross References: ATAXIC HEMIPARESIS; CHOREA, CHOREOATHETOSIS; DYSTONIA; PROPRIOCEPTION; PSEUDOATHETOSIS]

Pseudodementia

Pseudodementia is a label given to cognitive impairments resulting from affective disorders, most commonly anxiety and depression; the terms "dementia syndrome of depression" and "depression-related cognitive dysfunction" have also been used. The pattern of cognitive deficits in individuals with depression most closely resembles that seen in so-called subcortical dementia, with bradyphrenia, attentional and executive deficits. In addition there may be evident lack of effort and application, "don't know" answers, approximate answers (Ganser phenomenon, *vorbereiden*), and evidence of mood disturbance (tearfulness). Memory loss for recent and distant events may be equally severe (*cf.* temporal gradient of memory loss in dementia, *e.g.* due to Alzheimer's disease).

The recognition of pseudodementia is important since the deficits are often reversible with appropriate treatment with anti-depressants. However, it should be borne in mind that depression is sometimes the presenting symptom of an underlying neurodegenerative dementing disorder such as Alzheimer's disease.

- Kiloh L. Pseudodementia. *Acta Psychiatrica Scandinavica* 1961; **37**: 336-51

[Cross References: ATTENTION; BRADYPHRENIA; DEMENTIA; GANSER PHENOMENON]

Pseudodiplopia

- see PALINOPSIA

Pseudohypertrophy

- see CALF HYPERTROPHY

Pseudo-Internuclear Ophthalmoplegia (Pseudo-INO)

Pseudo-internuclear Ophthalmoplegia is a disorder of eye movements with impaired adduction in one eye and horizontal nystagmus in the abducting eye (*i.e.* signs as in an internuclear Ophthalmoplegia) but without an intrinsic brainstem lesion. This sign

may be seen in myasthenia gravis (a diagnosis which is always worthy of consideration in a patient with an "isolated INO") due to extraocular muscle weakness, Guillain-Barré syndrome, Miller Fisher syndrome, thyroid ophthalmopathy, and orbital pseudotumour.
[Cross References: INTERNUCLEAR OPHTHALMOPLEGIA; ONE-AND-A-HALF SYNDROME]

Pseudomyotonia
The term pseudomyotonia has been used to described the clinical appearance of myotonia (slow muscular relaxation after contraction) in the absence of myotonic discharges on electromyography. Pseudomyotonia is most commonly observed as the slow-relaxing or "hung-up" tendon reflexes (Woltman's sign) of hypothyroidism, although other causes are described.

Pseudomyotonia has also been used to describe difficulty opening the hand in cervical osteoarthritis, although muscle relaxation is normal; finger flexion on attempted extension was explained as aberrant regeneration of the C7 root.

The term pseudomyotonia has also been used to describe neuromyotonia and myokymia (as, for example, in Isaacs syndrome) to distinguish it from myotonia.

- Satoyoshi E, Doi Y, Kinoshita M. Pseudomyotonia in cervial root lesions with myelopathy. A sign of the misdirection of regenerating nerve. *Archives of Neurology* 1972; **27**: 307-13
[Cross References: MYOTONIA; NEUROMYOTONIA; WOLTMAN'S SIGN]

Pseudopapilloedema
Pseudopapilloedema is the name given to elevation of the optic disc that is not due to oedema (*i.e.* intracranial pressure is not raised). There may or may not be visible drusen (hyaline bodies). In distinction to oedematous disc swelling, the nerve fibre layer is not hazy and the underlying vessels are not obscured; however, spontaneous venous pulsation is usually absent, and haemorrhages may be seen, so these are not reliable distinguishing features. Visual acuity is usually normal, but visual field defects (most commonly in the inferior nasal field) may be found.
[Cross References: DISC SWELLING; PAPILLOEDMEA; VENOUS PULSATION]

Pseudoparesis
- see PARESIS; WEAKNESS

Pseudo-von Graefe's Sign
Pseudo-von Graefe's sign is seen when there is retraction or elevation of the upper eyelid (*cf.* Von Graefe's sign), medial rotation of the eye, and pupillary constriction on attempted downgaze or adduction. This constellation of findings is said to be a lid-gaze synkinesis following aberrant regeneration after an oculomotor (III) nerve palsy.
[Cross References: LID RETRACTION; SYNKINESIS; VON GRAEFE'S SIGN]

Psychomotor Retardation
Psychomotor retardation is a slowness of thought (bradyphrenia) and movement seen in psychiatric disorders, particularly depression. It may be confused with the akinesia

of parkinsonism, and abulic or catatonic states. Psychomotor retardation may also be a feature of the "subcortical" type of dementia, or of impairments of arousal (obtundation).
[Cross References: ABULIA; AKINESIA; CATATONIA; DEMENTIA; OBTUNDATION; PARKINSONISM]

Ptosis
Ptosis (blepharoptosis) is the name given to a drooping eyelid; this may be congenital or acquired, partial or complete, unilateral or bilateral, fixed or variable, isolated or accompanied by other signs (*e.g.* miosis in a Horner's syndrome; diplopia in myasthenia gravis; mydriasis and downward and outward deviation of the eye in an oculomotor (III) nerve palsy).

Ptosis may result from brainstem disease involving the oculomotor (III) nerve, pathology anywhere along the oculosympathetic autonomic pathway causing a Horner's syndrome, or in cortical disease (infarction) reflecting hemispheric control of the eyelid (probably bilateral).

When considering the cause of ptosis, the differential diagnosis is broad, including:

- Supranuclear lesion:
  Hemiparesis - due to cortical infarct; ptosis usually ipsilateral, incomplete;
  Duane syndrome - ptosis on eye adduction, due to supranuclear levator inhibition; usually with family history.
- Oculomotor (III) nerve:
  Hypertension, diabetes - ptosis often complete;
  Compressive lesion (*e.g.* posterior communicating artery aneurysm) - usually incomplete; ptosis may be present with subarachnoid haemorrhage;
  Oculomotor-trigeminal synkinesis: ptosis on jaw opening (inverse Marcus Gunn phenomenon).
- Neuromuscular junction:
  Myasthenia gravis - ptosis variable, bilateral or unilateral;
  Excessive botulinum toxin given for blepharospasm.
- Muscular causes:
  Mitochondrial disease, myotonic dystrophy, oculopharyngeal muscular dystrophy - ptosis usually bilateral.
- Local causes:
  Trauma, thyroid eye disease.

- Caplan LR. Ptosis. *Journal of Neurology, Neurosurgery and Psychiatry* 1914; **37**:1-7
[Cross References: BLEPHAROSPASM; DIPLOPIA; HORNER'S SYNDROME; JAW WINKING; MIOSIS; MYDRIASIS; PUPIL SPARING; SYNKINESIS]

Ptyalism
- see SIALORRHOEA

Pulfrich Phenomenon
The Pulfrich phenomenon is the observation by a patient with unilateral optic neuritis that a pendulum swung from side to side appears to traverse a curved trajectory, as a consequence of conduction slowing in the demyelinated optic nerve.

- Rushton D. Use of the Pulfrich pendulum for detecting abnormal delay in the visual pathways in multiple sclerosis. *Brain* 1975; **98**: 283-96
[Cross References: AFFERENT PUPILLARY DEFECT; PHOSPHENE]

"Pull Test"
- see POSTURAL REFLEXES

Pupillary Reflexes
Two pupillary reflexes are routinely examined in clinical practice:

~ *Light reflex:* the eye is illuminated directly and the reaction (constriction) observed; the *consensual light reflex* is observed by illuminating the contralateral eye. In an eye with poor acuity, a relative afferent pupillary defect may be observed using the "swinging flashlight test". The afferent pathway subserving the light reflex is optic nerve to thalamus, brainstem, and Edinger-Westphal nucleus, with the efferent limb (pupillomotor parasympathetic fibres) in the oculomotor (III) nerve. The contralateral (consensual) responses reflect fibres crossing the midline in the optic chiasm and in the posterior commissure at the level of the rostral brainstem.
~ *Accommodation reflex:* this is most conveniently examined by asking the patient to look into the distance, then focus on a near object (sufficiently close to necessitate convergence of the visual axes) when pupil constriction should occur (accommodation-convergence synkinesis). The afferent pathways subserving this response are less certain than for the light reflex, and may involve the occipital cortex, although the final (efferent) pathway via Edinger-Westphal nucleus and oculomotor nerve is common to both accommodation and light reflexes.

In comatose patients, fixed dilated pupils may be observed with central diencephalic herniation, whereas midbrain lesions produce fixed midposition pupils.
A dissociation between the light and accommodation reactions (light-near pupillary dissociation, *q.v.*) may be observed.
[Cross References: AFFERENT PUPILLARY DEFECT; ARGYLL ROBERTSON PUPIL; CILIOSPINAL RESPONSE; CORTICAL BLINDNESS; LIGHT-NEAR PUPILLARY DISSOCIATION]

Pupil Sparing
Oculomotor (III) nerve lesions may be pupil sparing (normal response to light) or pupil-involving (mydriasis, loss of light reflex). The latter situation usually implies a "surgical" cause of oculomotor palsy (*e.g.* posterior communicating artery aneurysm), especially if extraocular muscle function is relatively preserved. Pupil sparing suggests a "medical" cause (*e.g.* diabetes mellitus, hypertension) especially if the palsy is otherwise complete (complete ptosis, eye deviated downwards and outwards). This disparity arises because pupillomotor fibres run on the outside of the nerve, and are relatively spared by ischaemia but are vulnerable to external compression. However, the distinction is not absolute; imaging for an aneurysm (spiral CT, MRA, angiography) is necessary if the clinical scenario leaves room for doubt.
[Cross References: OPHTHALMOPARESIS, OPHTHALMOPLEGIA; PTOSIS; PUPILLARY REFLEXES]

Pure Word Blindness
- see ALEXIA

Pure Word Deafness

Pure word deafness is a rare condition characterized by an inability to comprehend and discriminate spoken language, despite adequate hearing as measured by audiometry, and with preserved spontaneous speech, reading, reading comprehension, and writing. Lip reading may assist in the understanding of others who sometimes seem to the patient as though they are speaking in a foreign language. There may be associated amusia, depending on the precise location of cerebral damage.

Pure word deafness has been variously conceptualised as a form of auditory agnosia or a subcortical sensory aphasia.

Pure word deafness is most commonly associated with bilateral lesions of the temporal cortex or subcortical lesions whose anatomical effect is to damage the primary auditory cortex or isolate it through lesions of the auditory radiation. Very rarely pure word deafness has been associated with bilateral brainstem lesions at the level of the inferior colliculi.

- Meyer B, Kral T, Zentner J. Pure word deafness after resection of a tectal plate glioma with preservation of wave V of brain stem auditory evoked potentials. *Journal of Neurology, Neurosurgery and Psychiatry* 1996; **61:** 423-4
- Tanaka Y, Yamadori A, Mori E. Pure word deafness following bilateral lesions. A psychophysical analysis. *Brain* 1987; **110:** 381-403
[Cross References: AGNOSIA; AMUSIA; APHASIA]

Pyramidal Signs

Also known as **Pyramidal Weakness**
- see HEMIPARESIS; UPPER MOTOR NEURONE SYNDROME; WEAKNESS

Q

Quadrantanopia
Quadrantanopia (quadrantanopsia), a defect in one quarter of the visual field, suggests an optic radiation lesion. Temporal lobe pathology damaging Meyer's loop typically produces a superior homonymous quadrantanopia. Parietal lobe lesions may produce inferior quadrantic defects. Damage to extrastriate visual cortex (areas V2 and V3) has also been suggested to cause quadrantanopia; concurrent central achromatopsia favours this notion.

- Horton JC, Hoyt WF. Quadrantic visual field defects. A hallmark of lesions in extrastriate (V2/V3) cortex. *Brain* 1991; **114**: 1703-18
[Cross References: ACHROMATOPSIA; HEMIANOPIA]

Quadriparesis, Quadriplegia
Quadriparesis and quadriplegia refer to weakness (partial or total, respectively) of all four limbs which may be of upper motor neurone or, less commonly, lower motor neurone type (*e.g.* in Guillain-Barré syndrome). As with hemiplegia, upper motor neurone quadriplegia may result from lesions of the corticospinal pathways anywhere from motor cortex to cervical cord via the brainstem, but is most commonly seen with brainstem and upper cervical cord lesions. In such circumstances, respiration may be affected.
[Cross References: HEMIPARESIS; PARAPLEGIA]

R

Rabbit Syndrome
Rabbit syndrome is a rest tremor of the perioral and nasal muscles, which may occur with both antipsychotic drug therapy and idiopathic Parkinson's disease. It is therefore presumably related to dopamine deficiency. Drug-induced rabbit syndrome may remit with drug withdrawal.
[Cross References: PARKINSONISM]

"Raccoon Eyes"
"Raccoon eyes" refers to an appearance of bilateral periorbital ecchymosis, appearing 48-72 hours after an anterior basal skull fracture.

Radiculopathy
A radiculopathy is a disorder of nerve roots, causing pain in a radicular distribution, paraesthesia, sensory diminution or loss in the corresponding dermatome, and lower motor neurone type weakness with reflex diminution or loss in the corresponding myotome. Radiculopathies may be single or multiple (polyradiculopathy, *e.g.* cauda equina syndrome).

Electrophysiological studies may be helpful in distinguishing radiculopathy from a neuropathy or plexopathy: sensory nerve action potentials are normal for intrathecal root lesions, and EMG shows involvement of paraspinal muscles.

Causes of radiculopathy include:

- Compression: disc protrusion: cervical (especially C6, C7), lumbar (L5, S1) >>> thoracic; bony metastases; spondylolisthesis; fracture; infection.
- Root avulsion, *e.g.* C5/C6, "waiter's tip" posture; C8/T1, claw hand +/- Horner's syndrome.
- Diabetic polyradiculopathy: thoraco-abdominal, lumbosacral (= diabetic amyotrophy, also known as diabetic lumbar sacral plexopathy, proximal diabetic neuropathy; especially involves L2-L4).
- Neoplasia: with meningeal symptoms, due to spread from carcinoma of breast or lung, melanoma, non-Hodgkin's lymphoma, leukaemia.
- Infection: HIV (CMV late in the course), *Borrelia,* syphilis (tabes dorsalis), herpes zoster (thoracic > cervical > lumbosacral; sensory >> motor).
- Demyelination: Guillain-Barré syndrome, chronic inflammatory demyelinating polyradiculopathy (CIDP).

- Chad DF. Nerve root and plexus disorders. In: Bogousslavsky J, Fisher M (Eds). *Textbook of Neurology.* Boston: Butterworth-Heinemann 1998: 491-506
[Cross References: CAUDA EQUINA SYNDROME; NEUROPATHY; PARAESTHESIA; PLEXOPATHY; REFLEXES; WEAKNESS]

Rebound Phenomenon

This is one feature of the impaired checking response seen in cerebellar disease, along with dysdiadochokinesia and macrographia. It may be demonstrated by observing an overshoot of the outstretched arms when they are released suddenly after being pressed down by the examiner.

[Cross References: ASYNERGIA; ATAXIA; DYSDIADOCHOKINESIA; DYSMETRIA; MACROGRAPHIA]

Recruitment

Recruitment, or loudness recruitment, is the phenomenon of abnormally rapid growth of loudness with increase in sound intensity, which is encountered in patients with sensorineural (especially cochlear sensory) hearing loss. Thus patients have difficulty with sounds of low to moderate intensity ("Speak up, doctor") but intense sounds are uncomfortably loud ("There's no need to shout, doctor!"). Speech discrimination is relatively unimpaired in conductive hearing loss.

"Recruitment" may also be used to refer to pathological "spread" of tendon reflexes, implying broadening of their receptive field.

[Cross References: REFLEXES]

Reduplicative Paramnesia

Reduplicative paramnesia is a delusion in which patients believe familiar places, objects, individuals or events to be duplicated. The syndrome is probably heterogeneous and bears some resemblance to the Capgras delusion described in psychiatry.

Reduplicative paramnesia is commoner with right (non-dominant) hemisphere damage; frontal, temporal and limbic system damage has been implicated. This may occur transiently as a consequence of cerebrovascular disease, following head trauma, or even after migraine attacks, or more persistently in the context of neurodegenerative disease (Alzheimer's disease).

- Benson DF, Gardner H, Meadows JC. Reduplicative paramnesia. *Neurology* 1976; **26**: 147-51

[Cross References: DELUSION; PARAMNESIA]

Reflexes

Reflex action (sensory stimulus provoking involuntary motor response) is a useful way of assessing the integrity of neurological function, since disease in the afferent limb, synapse, or efferent limb of the reflex arc may lead to dysfunction, as may changes in inputs from higher centres.

Different types of reflex may be distinguished. Muscle tendon reflexes may be either tonic (in response to a static applied force: "stretch reflex") or phasic (in response to a brief applied force, for example a blow from a tendon hammer to the muscle tendon). The latter are of particular use in clinical work because of their localising value (see Table). However, there are none between T2 and T12, and thus for localisation one is dependent on sensory findings, or occasionally cutaneous (skin or superficial) reflexes, such as the abdominal reflexes.

Reflex	Root Value
Jaw jerk	Trigeminal (V) nerve
Supinator (Brachioradialis)	C5, C6
Biceps	C5, C6
Triceps	C7
Finger flexion (Digital)	C8, T1
Abdominal	T7 to T12
Cremasteric	L1
Knee (Patellar)	L3, L4
Hamstring	L5, S1
Ankle (Achilles)	(L5) S1 (S2)

Tendon reflex responses are usually graded on a five point scale:

- absent (areflexia; as in lower motor neurone syndromes, such as peripheral nerve or anterior horn cell disorders; or acute upper motor neurone syndromes, *e.g.* "spinal shock")

+/- present only with reinforcement (Jendrassik's manoeuvre)

+ normal

++ brisk normal

+++ pathologically brisk (hyperreflexia, as in upper motor neurone syndromes)

Reflex "spread", or "recruitment", for example a finger jerk when eliciting the supinator or biceps jerk, is suggestive of corticospinal pathway (upper motor neurone) pathology, producing an enlarged receptive field for the reflex response; concurrent disruption of the local reflex arc may result in inverted reflexes (*q.v.*).

Reflex responses may vary according to the degree of patient relaxation or anxiety (pre-contraction). Moreover, there is interobserver variation in the assessment of tendon reflexes (as with all clinical signs): a biasing effect of prior knowledge upon reflex assessment has been recorded.

- Stam J, van Crevel H. Reliability of the clinical and electromyographic examination of tendon reflexes. *Journal of Neurology* 1990; **237**: 427-31

[Cross References: AREFLEXIA; FACILITATION; FRONTAL RELEASE SIGNS; HYPERREFLEXIA; HYPOREFLEXIA; INVERTED REFLEXES; JENDRASSIK'S MANOEUVRE; LOWER MOTOR NEURONE SYNDROME; PRIMITIVE REFLEXES; PUPILLARY REFLEXES; UPPER MOTOR NEURONE SYNDROME; see also specific (named) reflexes]

Relative Afferent Pupillary Defect (RAPD)

- see AFFERENT PUPILLARY DEFECT

Retinitis Pigmentosa

Retinitis pigmentosa is a generic name for an inherited retinal degeneration, characterized clinically by typical appearances on ophthalmoscopy, with peripheral

pigmentation of "bone-spicule" type, arteriolar attenuation, and eventually unmasking of choroidal vessels and optic atrophy. This process may be asymptomatic in its early stages, but may later be a cause of nyctalopia (night blindness), and produce a mid-peripheral ring scotoma on visual field testing.

A variety of genetic causes of isolated retinitis pigmentosa have been partially characterized:

- autosomal recessive: linked to chromosome 1q
- X-linked:Xp11,Xp21
- autosomal dominant: 3q, 6p, 8

In most cases, patients with retinitis pigmentosa have no associated systemic or extraocular abnormalities, but there are a number of multisystem disorders in which it occurs:

- Abetalipoproteinaemia (Bassen-Kornzweig syndrome)
- Kearns-Sayre syndrome, mitochondrial disorders in general
- Lawrence-Moon-Bardet-Biedl syndrome
- Refsum's disease
- Usher's syndrome.

[Cross References: NYCTALOPIA; OPTIC ATROPHY; SCOTOMA]

Retinopathy

Retinopathy is any pathological process affecting the retina, with changes observable on ophthalmoscopy; dilatation of the pupil aids observation of the peripheral retina. Common causes include:

- Diabetes mellitus: various abnormalities may occur, in both insulin-dependent (IDDM) and non-insulin dependent (NIDDM) patients. "Background" diabetic retinopathy is manifest as microaneuryms, dot and blot haemorrhages, hard exudates, and diffuse retinal oedema, all of which may be asymptomatic. Oedema and hard exudates at the macula are a common cause of visual impairment. Proliferative retinopathy is characterized by neovascularisation of the disc due to retinal hypoxia, typically in IDDM, with the risk of vitreous haemorrhage, traction retinal detachment and irreversible visual loss. Laser treatment of new vessels is the treatment of choice.
- Hypertension: hypertensive retinopathy may cause arteriolar constriction, with the development of cotton-wool spots; and abnormal vascular permeability causing flame-shaped haemorrhages, retinal oedema and hard exudates; around the fovea, the latter may produce a macular star. Optic disc swelling may be seen in malignant hypertension. Arteriosclerosis, thickening of vessel walls with prolonged hypertension, may cause changes at arteriovenous crossings ("AV nipping"). Systemic hypertension is associated with an increased risk of branch retinal vein and central retinal artery occlusion.
- Drug-induced, *e.g.* antimalarials (chloroquine); chlorpromazine.
- Retinitis pigmentosa (*q.v.*).
- Cancer-associated retinopathy: arteriolar narrowing, optic atrophy.

An electroretinogram (ERG) may be helpful in confirming the presence of a retinopathic disorder.
[Cross References: MACULOPATHY; RETINITIS PIGMENTOSA]

Retrocollis
Retrocollis is an extended posture of the neck. Steele-Richardson-Olszewski syndrome (progressive supranuclear palsy, PSP) is commonly associated with retrocollis (*cf.* antecollis in multisystem atrophy), and it may also be a feature of torticollis.
[Cross References: ANTECOLLIS; PARKINSONISM; TORTICOLLIS]

Retropulsion
- see FESTINANT GAIT, FESTINATION; POSTURAL REFLEXES

"Reverse Ptosis"
- see HORNER'S SYNDROME; PTOSIS

Revilliod's Sign
Revilliod's sign is an acquired inability to wink, a sign of corticobulbar disease (possibly an early sign).

Riddoch's Phenomenon
Riddoch's phenomenon is the dissociation of the perception of static and kinetic visual stimuli (statokinetic dissociation) which may occur with occipital or anterior visual pathway lesions.

- Zeki S, Ffytche DH. The Riddoch syndrome: insights into the neurobiology of conscious vision. *Brain* 1998; **121**: 25-45
[Cross References: AKINETOPSIA; VISUAL AGNOSIA]

Right-Left Disorientation
Right-left disorientation is an inability to say whether a part of the body is right or left, or to use a named body part to command. It may occur in association with acalculia, agraphia, and finger agnosia (the Gerstmann syndrome), indicating a posterior parietal dominant hemisphere lesion.
[Cross References: ACALCULIA; AGRAPHIA; AUTOTOPAGNOSIA; FINGER AGNOSIA; GERSTMANN SYNDROME]

Rigidity
Rigidity is an increased resistance to passive movement of a joint which is constant throughout the range of joint displacement and not related to the speed of joint movement; resistance is present in both agonist and antagonist muscles. In these particulars, rigidity differs from spasticity. It also needs to be differentiated from stiffness.
Rigidity may be:

~ *consistent:* "leadpipe rigidity"; or
~ *jerky:* "cogwheeling, cogwheel phenomenon, cogwheel rigidity" or Negro's sign,

when a rhythmic fluctuation (*i.e.* tremor), like a ratchet or cogwheel, is superimposed on the background of sustained rigidity.

The presence of rigidity may be made more obvious by reinforcing manoeuvres (*e.g.* clenching and relaxing the contralateral fist, performing mental arithmetic), a finding variously known as activated rigidity, or Froment's sign, or synkinesis (but note that both Froment's sign and synkinesis have other meanings too). However, this may occur in some normal subjects; it is most helpful in the diagnosis of Parkinson's disease if unilateral. Rigidity may also be demonstrated using Wartenberg's swing test(*q.v.*).

Rigidity is a feature of parkinsonism and may co-exist with any of the other clinical features of extrapyramidal system disease, but particularly akinesia; both are associated with loss of dopamine projections from the substantia nigra to the putamen. Rigidity is a feature of pathology within the basal ganglia.

The pathophysiology of rigidity is thought to relate to overactivity of tonic stretch reflexes in the spinal cord due to excessive supraspinal drive to spinal cord **α-motor** neurones following loss of descending inhibition as a result of basal ganglia dysfunction. In other words, there is a change in the sensitivity of the spinal interneurones which control **α-motor** neurones due to defective supraspinal control. Hence rigidity is a positive or release symptom, reflecting the operation of intact suprasegmental centres. The physiological correlate of this is the increased EMG activity found in rigid muscles with increased 1A afferent fibre activity, suggesting maintained **α-γ** linkage. In support of this, pyramidotomy has in the past been shown to produce some relief of rigidity.

Rigidity in Parkinson's disease may be lessened by treatment with levodopa preparations. The techniques of modern stereotactic neurosurgery may also be helpful, particularly stimulation of the subthalamic nucleus, although both thalamotomy and pallidotomy may also have an effect.

The term rigidity may also be used to describe:

- posturing associated with coma: decorticate or decerebrate, flexor and extensor posturing respectively;
- a lack of mental flexibility, particularly evident in patients with frontal lobe dysfunction.

- Cantello R, Gianelli M, Civardi C, Mutani R. Pathophysiology of Parkinson's disease rigidity: role of corticospinal motor projections. *Advances in Neurology* 1996; **69:** 129-33

[Cross References: DECEREBRATE RIGIDITY; DECORTICATE RIGIDITY; FROMENT'S SIGN; FRONTAL LOBE SYNDROMES; PARKINSONISM; STIFFNESS; SYNKINESIS; TREMOR; WARTENBERG'S SWING TEST]

Rinne's Test

Rinne's test is one of the tuning fork tests (512 Hz fork preferred), which is used to define whether there is a conductive element to hearing loss. The patient is asked to compare the loudness of a vibrating tuning fork held at the external auditory meatus (air conduction; AC) with the fork held against the mastoid process (bone conduction; BC); masking of the other ear, for example by rubbing the tragus, is advised.

Normally air conduction is louder (AC > BC); if bone conduction sounds louder (BC > AC), then this is indicative of a conductive hearing loss. In sensorineural hearing loss, AC and BC are diminished to a similar extent, and air conduction remains louder (AC > BC).
[Cross References: WEBER'S TEST]

Risus Sardonicus
Risus sardonicus ("sardonic smile") due to spasm of the facial musculature is a classic feature of the neuromuscular syndrome of tetanus, now exceptionally rarely seen in the West. Risus sardonicus may also occur in the context of dystonia, more usually symptomatic (secondary) than idiopathic (primary) dystonia.
[Cross References: DYSTONIA; SPASM]

"Rocket Sign"
The so-called "rocket sign" is a toppling backwards, after jumping to the feet from the sitting position, due to postural instability, seen in Steele-Richardson-Olszewski syndrome and ascribed to frontal lobe dysfunction. A history of falls due to postural instability in the first year after disease onset is one of the mandatory inclusion criteria for the diagnosis of Steele-Richardson-Olszewski syndrome.
[Cross References: PARKINSONISM; "WHEELCHAIR SIGN"]

Roger's Sign
Roger's sign, or the numb chin syndrome, is an isolated neuropathy affecting the mental branch of the mandibular division of the trigeminal (V) nerve, causing pain, swelling, and numbness of the lower lip, chin and mucous membrane inside the lip. This is usually a sign of metastatic spread of cancer to the jaw. Hypoaesthesia involving the cheek, upper lip, upper incisors and gingiva, due to involvement of the infraorbital portion of the maxillary division of the trigeminal nerve ("numb cheek syndrome") is also often an ominous sign, resulting from recurrence of squamous cell carcinoma of the face infiltrating the nerve.

- Campbell WW Jr. The numb cheek syndrome: a sign of infraorbital neuropathy. *Neurology* 1986; **36**: 421-3
- Roger H, Paillas J. Le signe du mentonnier (parasthésie et anesthésie unilatérale) révélateur d'un processus néoplasique métastatique. *Revue Neurologique (Paris)* 1937; **2**: 751-2

Rombergism, Romberg's Sign
Romberg's sign is adjudged present (or positive) when there is a dramatic increase in unsteadiness, sometimes with falls, after eye closure in a patient standing comfortably. (Before asking the patient to close his/her eyes, it is advisable to position ones arms in such a way as to be able to catch the patient should they begin to fall.) Patients may fall forward immediately on eye closure ("sink sign"). These phenomena result from sensory ataxia (*i.e.* loss of proprioception from the feet), which occurs most commonly with posterior column spinal cord disease: Romberg's sign may be seen in tabes dorsalis.

A modest increase in sway on closing the eyes may be seen in normal subjects, and patients with cerebellar ataxia, frontal lobe ataxia, and vestibular disorders; on occasion these too may produce an increase in sway sufficient to cause falls (*i.e.* the test is not specific). Posturography is an attempt to quantify the Romberg test.
[Cross References: ATAXIA; PROPRIOCEPTION]

Rooting Reflex
The rooting reflex is a turning of the head towards a tactile stimulus on the face or an object approaching the mouth, a normal response in infants which is lost during childhood. Its presence in adults is indicative of diffuse premotor frontal disease, this being a primitive reflex or frontal release sign.
[Cross References: FRONTAL RELEASE SIGNS; PRIMITIVE REFLEXES]

Rosenbach's Sign
- see ABDOMINAL REFLEXES

Roving Eye Movements
Roving eye movements consist of slow drifting movements of the eyes from side to side; the eyelids are closed and there may be slight divergence of the ocular axes. Roving eye movements may be seen in normal sleep, but also in comatose patients in whom they are indicative of an intact brainstem (*e.g.* the early diencephalic stage of central herniation) but are otherwise non-localising. As coma deepens, roving eye movements are lost before the movements provoked by the oculocephalic (doll's head) manoeuvre (oculocephalic reflexes, vestibulo-ocular reflexes), or the caloric tests.
[Cross References: CALORIC TESTING; COMA; VESTIBULO-OCULAR REFLEXES]

S

Saccades

Saccades are rapid, ballistic, yoked movements of the eyes which bring the gaze to a new location in visual space. These movements may be performed voluntarily (tested clinically by asking the patient to "Look to your left", *etc.*) or reflexively, *i.e.* in response to an object of potential interest within the visual field (tested clinically by asking the patient to shift gaze from one of examiner's hands to another). Internuclear ophthalmoplegia may be revealed when testing saccadic eye movements.

A number of parameters may be observed, including latency of saccade onset, saccadic amplitude, and saccadic velocity. An anti-saccadic task (*i.e.* suppression of saccades to a novel visual stimulus) may be used to assess ease of saccade suppression. Of these, saccadic velocity is the most important in terms of localization value, since it depends on burst neurones in the brainstem (paramedian pontine reticular formation for horizontal saccades, rostral interstitial nucleus of the medial longitudinal fasciculus for vertical saccades). Latency involves cortical and basal ganglia circuits; anti-saccades involve frontal lobe structures; and amplitude involves basal ganglia and cerebellar circuits (saccadic hypometria, with a subsequent correctional saccade, may be seen in extrapyramidal disorders such as Parkinson's disease; saccadic hypermetria or overshoot may be seen in cerebellar disorders). Difficulty in initiating saccades may be described as ocular (motor) apraxia. Anti-saccades may be poorly suppressed in Huntington's disease.

Assessment of saccadic velocity may be of particular diagnostic use in parkinsonian syndromes. In Steele-Richardson-Olszewski syndrome (progressive supranuclear palsy) slowing of vertical saccades is an early sign (suggesting brainstem involvement; horizontal saccades may be affected later), whereas vertical saccades are affected late (if at all) in corticobasal degeneration, in which condition increased saccade latency is the more typical finding, perhaps reflective of cortical involvement.

- Leigh RJ, Riley DE. Eye movements in parkinsonism: it's saccadic speed that counts. *Neurology* 2000; **54**: 1018-9

[Cross References: INTERNUCLEAR OPHTHALMOPLEGIA; OCULAR APRAXIA; OCULAR FLUTTER; OPSOCLONUS; PARKINSONISM; SACCADIC INTRUSION; SQUARE-WAVE JERKS]

Saccadic Intrusion

Saccadic intrusions are inappropriate saccades which interfere with visual fixation (static or during motor pursuit). Several types of saccadic intrusion are described, including ocular flutter, opsoclonus, and square-wave jerks.

[Cross References: OCULAR FLUTTER; OPSOCLONUS; SACCADES; SQUARE-WAVE JERKS]

Saccadomania
- see OPSOCLONUS

Sacral Sparing
Sacral sparing is the preservation of pain and temperature sensation in sacral dermatomes. This is a late, unusual, but diagnostic feature of an intrinsic (intramedullary) spinal cord lesion. Spastic paraparesis below the level of the lesion due to corticospinal tract involvement is invariably present by this stage.

Sacral sparing is explained by the lamination of fibres within the spinothalamic tract: ventrolateral fibres (of sacral origin) are involved later than the dorsomedial (cervical, thoracic) fibres by an expanding central intramedullary lesion (*e.g.* glioma, ependymoma, syringomyelia).

Although sacral sparing is rare, sacral sensation should always be checked in any patient with a spastic paraparesis.

[Cross References: DISSOCIATED SENSORY LOSS; MYELOPATHY; PARAPARESIS]

Saturday Night Palsy
- see WRIST DROP

Scanning Speech
Scanning speech is a motor speech disorder (*i.e.* a dysarthria) comprising slow, deliberate, dysprosodic, monotonic verbal output. It may be confused with non-fluent aphasia (Broca's aphasia).

Scanning speech was originally considered a feature of cerebellar disease in multiple sclerosis (after Charcot), and the term is often used with this implication. However, cerebellar disease typically produces an ataxic dysarthria which is somewhat different to scanning speech (variable intonation, interruption between syllables, "explosive" speech). Scanning speech correlates with midbrain lesions, often after recovery from prolonged coma.

[Cross References: ASYNERGIA; APHASIA; BROCA'S APHASIA; CEREBELLAR SYNDROMES; DYSARTHRIA]

Scotoma
A scotoma is a localised area of impaired vision within an otherwise normal visual field: this may be mapped clinically by confrontation testing, or automatically. In addition to the peripheral field, the central field should also be tested, with the target object moved around the fixation point. A central scotoma may be picked up in this way, or a more complex defect such as a centrocaecal scotoma in which both the macula and the blind spot are involved. Infarction of the occipital pole will produce a central visual loss, as will optic nerve inflammation. Scotomata may be absolute (no perception of form or light) or relative (preservation of form, loss of colour).

A scotoma may be physiological, as in the blind spot, or pathological, reflecting pathology anywhere from retina and choroid to visual cortex. The exact pattern may have localising value due to the retinotopic arrangement of fibres in the visual pathways: any unilateral area of restricted loss implies a pre-chiasmatic lesion

(choroid, retina, optic nerve), although lesions of the anterior calcarine cortex can produce a contralateral monocular temporal crescent. Bilateral homonymous scotomata are post-chiasmal in origin; bilateral heteronymous scotomata may be seen with chiasmal lesions.

Various types of scotoma may be detected:

- Central scotoma: occupying the macula, due to involvement of the macula or the papillomacular bundle; the typical (but not exclusive) finding in optic neuritis, also seen with disease of the macula, optic nerve compression, Leber's hereditary optic neuropathy. A unilateral central scotoma with a contralateral superior temporal defect may be seen with lesions at the anterior angle of the chiasm, damaging the ipsilateral optic nerve plus the crossing loop of fibres (Wilbrand's knee) originating from the inferonasal portion of the contralateral eye: this may be termed a junctional scotoma, but should not be confused with the junctional scotoma of Traquair (*vide infra*);
- Caecocentral or centrocaecal scotoma: involving both the macula and the blind spot; seen in optic nerve disease, *e.g.* Leber's hereditary optic neuropathy, toxic/nutritional optic neuropathy, sometimes in optic neuritis;
- Arcuate scotoma: retinal or optic nerve disease: glaucoma, acute ischaemic optic neuropathy, drusen;
- Annular or ring scotoma: retinitis pigmentosa, cancer-associated retinopathy;
- Junctional scotoma of Traquair: a monocular temporal scotoma, sometimes even hemianopia, seen with optic nerve involvement sufficiently close to the chiasm to involve only ipsilateral crossing nasal axons (subserving the temporal visual field) but sparing nasal axons crossing from the contralateral eye;
- Peripapillary scotoma: this describes enlargement of the blind spot.

- Trobe JD, Acosta PC, Krischer JP, Trick GL. Confrontation visual field techniques in detection of anterior visual pathway lesions. *Annals of Neurology* 1981; **10**:28-34
[Cross References: ALTITUDINAL FIELD DEFECT; BLIND SPOT; HEMIANOPIA; MACULOPATHY; QUADRANTANOPIA; RETINITIS PIGMENTOSA; RETINOPATHY]

"Scratch Test"

The "scratch test", or "direction of scratch" test, examines perception of the direction (up or down) of a scratch applied to the anterior shin (for example, with the sharp margin of a paper clip); it has been claimed as a reliable test of posterior column function of the spinal cord. Errors in this test correlate with central conduction times and vibration perception threshold.

- Hankey GJ, Edis R. The utility of testing tactile perception of direction of scratch as a sensitive clinical sign of posterior column dysfunction in spinal cord disorders. *Journal of Neurology, Neurosurgery and Psychiatry* 1989; **52**: 395-8
- Motoi Y, Matsumoto H, Kaneshige Y, Chiba S. A reappraisal of "direction of scratch" test: using somatosensory evoked potentials and vibration perception. *Journal of Neurology, Neurosurgery and Psychiatry* 1992; **55**: 509-10
[Cross References: PROPRIOCEPTION; VIBRATION]

Seborrhoea
Seborrhoea is a greasiness of the skin which may occur in extrapyramidal disorders, particularly Parkinson's disease.
[Cross References: PARKINSONISM]

Self-mutilation
Self injury to the point of mutilation, especially around the mouth, may be seen in certain neurological conditions, such as Lesch-Nyhan syndrome, Gilles de la Tourette syndrome, and neuroacanthocytosis.

Seizures
Seizures are sudden, paroxysmal episodes of neurological dysfunction with or without impairment of consciousness, which may be epileptic (*i.e.* due to abnormal synchronous electrical activity within the brain, either focally or generally) or non-epileptic in origin ("pseudoseizures", non-epileptic attack disorder). The two varieties may co-exist. Seizure morphology may be helpful in establishing aetiology and/or focus of onset.

Epileptic:
Idiopathic generalised: tonic-clonic (grand mal); absence attack (petit mal);
Partial: simple (no impairment of Consciousness), for example jerking of one arm, which may spread sequentially to other body parts (jacksonian march); or complex, in which there is impairment or loss of consciousness: may be associated with specific aura (olfactory, *déjà vu, jamais vu*) and/or automatisms (motor, *e.g.* cursive; or emotional, *e.g.* gelastic, dacrystic); limb posturing and pelvic thrusting may be seen in frontal lobe epilepsy. Secondary generalisation of seizures of partial onset may occur. Investigation of partial seizures to exclude a symptomatic cause is recommended (MR imaging, EEG). Some are amenbale to surgical intervention. Otherwise, as for idiopathic generalised epilepsies, various anti-epileptic medications are available. Partial seizures may prove more resistant to treatment than generalised seizures.
Non-epileptic:
long lasting, thrashing, pelvic thrusting, carpet burns, may have incontinence; past history of physical or sexual abuse. Best treated with psychological approaches, or drug treatment of underlying affective disorders; anti-epileptic medications are best avoided.

The differentiation of epileptic from non-epileptic seizures may be difficult; it is sometimes helpful to see a video recording of the attacks.

- Duncan JS, Shorvon SD, Fish DR. *Clinical epilepsy.* New York: Churchill Livingstone 1995: 25-101
[Cross References: ABSENCE; AURA; AUTOMATISM; *DÉJÀ VU;* INCONTINENCE; JACKSONIAN MARCH; *JAMAIS VU;* PELVIC THRUSTING]

Sialorrhoea
Sialorrhoea (drooling) is excessive salivation, possibly due to excess flow of saliva but more likely secondary to a reduced frequency of swallowing (*e.g.* in parkinsonian

syndromes) or difficulty swallowing (*e.g.* motor neurone disease, developmental perisylvian syndrome).

Metallic poisonings (mercury, bismuth, lead) may also produce marked salivation (ptyalism).

If troublesome, treatment of sialorrhoea with anticholinergic agents may be tried (atropine, hyoscine), although they may cause confusion in Parkinson's disease. In extreme cases, irradiation of the salivary glands has been used. Recently, the use of intraparotid injections of botulinum toxin has been found useful.

- Bhatia KP, Münchau A, Brown P. Botulinum toxin is a useful treatment in excessive drooling of saliva. *Journal of Neurology, Neurosurgery and Psychiatry* 1999; **67**: 697
[Cross References: BULBAR PALSY; PARKINSONISM]

Sighing

Occasional deep involuntary sighs may occur in multiple system atrophy. Sighing is also a feature, along with yawning, of the early (diencephalic) stage of central herniation of the brainstem with an otherwise normal respiratory pattern. Sudden inspiratory or expiratory sighs are said to be a feature of the hyperkinetic choreiform dysarthria characteristically seen in choreiform disorders such as Huntington's disease.

- Quinn N. Multiple system atrophy. In: Marsden CD, Fahn S (eds.). *Movement disorders 3.* Boston: Butterworth 1994: 262-81
[Cross References: BLINKING; DYSARTHRIA; YAWNING]

Simian Posture

- see PARKINSONISM

Simultanagnosia

Simultanagnosia is impaired perception of multi-element or multipart visual displays, such that pictures are described in a piecemeal manner. Recognition of single objects is preserved; this is likened to having a fragment or island of clear vision which may shift from region to region

Two types of Simultanagnosia are described:

~ *dorsal:* an attentional limitation preventing more than one object being seen at a time; although superficially similar to apperceptive visual agnosia, with which it has sometimes been classified, patients with dorsal Simultanagnosia can recognise objects quickly and accurately, but unattended objects are not seen. There may be inability to localise stimuli even when they are seen, manifest as visual disorientation. Reading is severely impaired. Patients may grope, as though blind. Dorsal Simultanagnosia is associated with bilateral posterior parieto-occipital lesions;

~ *ventral:* a limitation in the number of objects which can be recognised in unit time (*i.e.* no primary recognition problem in that individual shapes can be recognised). Ventral Simultanagnosia is most evident during reading which is severely impaired and empirically this may be the same impairment as seen in pure alexia; otherwise deficits may not be evident, unlike dorsal Simultanagnosia. Ventral Simultanagnosia may be a form of associative visual agnosia. It is associated with left inferior temporo-occipital cortical lesions.

- Coslett HB, Saffran E. Simultanagnosia: to see but not two see. *Brain* 1991; **114**: 1523-45
- Farah MJ. *Visual agnosia: disorders of object recognition and what they tell us about normal vision.* Cambridge: MIT Press 1995
[Cross References: AGNOSIA; ALEXIA; BALINT'S SYNDROME; VISUAL AGNOSIA; VISUAL DISORIENTATION]

Singulus
- see HICCUPS

"Sink Sign"
- see ROMBERG'S SIGN, ROMBERGISM

Skew Deviation
Skew deviation, or the Hertwig-Magendie sign, refers to a vertical misalignment of the eyes of supranuclear origin (*cf.* hypertropia, hypotropia, due to oculomotor nerve palsy or extraocular muscle disease). The commonest associated lesions are in the brainstem tegmentum, anywhere between the diencephalon and medulla. The precise pattern of skew deviation, with ocular tilt reaction, may be of localising value. In this situation Bielschowsky's head tilt test is usually negative.

- Brandt Th, Dieterich M. Different types of skew deviation. *Journal of Neurology, Neurosurgery and Psychiatry* 1991; **54:** 549-50
[Cross References: BIELSCHOWSKY'S SIGN, BIELSCHOWSKY'S TEST; HYPERTROPIA; HYPOTROPIA; OCULAR TILT REACTION; TULLIO PHENOMENON]

Snarling Facies
Patients with profound weakness of facial musculature may appear to snarl when asked to smile, or may seem peculiarly unamused by an examiner's attempted witticisms. This may be seen in myasthenia gravis ("myasthenic snarl") and other causes of facial weakness, such as facioscapulohumeral dystrophy.

Snout Reflex
- see POUT REFLEX

Somatotopagnosia
- see AUTOTOPAGNOSIA

Spasm
The word spasm implies a sudden, involuntary, muscle contraction, which may be painful. For example, flexor spasms in patients paraplegic due to upper motor neurone lesions are sudden contractions of the flexor musculature, particularly of the legs, either spontaneous or triggered by light touch. Hemifacial spasm is an involuntary contraction of facial musculature.

Spasm may also refer to a tetanic muscle contraction (tetany), as seen in hypocalcaemic states (*e.g. main d'accoucheur*), tetanus (*e.g.* risus sardonicus), or

tonic spasms of various muscles (*e.g.* jaw musculature, trismus) which may be dystonic or spastic in origin.

Patients may use the word spasm differently, *e.g.* to denote paroxysmal sensory phenomena, or even seizures. Infantile seizures consisting of brief flexion of the trunk and limbs (salaam or jackknife seizures) may be known as spasms.

- Rowland LP. Cramps, spasms, and muscle stiffness. *Revue Neurologique (Paris)* 1985; **141**; 261-73

[Cross References: CONTRACTURE; DYSTONIA; HEMIFACIAL SPASM; *MAIN D'ACCOUCHEUR;* PARAPLEGIA; RISUS SARDONICUS; SEIZURES; TRISMUS]

Spasmus Nutans

Spasmus nutans is the clinical triad of head nodding, anomalous head postures, and nystagmoid eye movements seen in children aged between 1 and 8. This is usually a benign condition.

[Cross References: NYSTAGMUS]

Spastic Catch

- see SPASTICITY

Spasticity

Spasticity is an increased resistance to the passive movement of a joint due to abnormally high muscle tone, which varies with the amplitude and speed of displacement of a joint (*cf.* rigidity). The excessive resistance evident at the extremes of joint displacement may suddenly give way, a phenomenon known as clasp-knife (or, confusingly, clasp-knife rigidity). Spasticity may vary in degree from mild, (*e.g.* a spastic catch on supination/pronation of the forearm), to extreme (*e.g.* immobile limbs in fixed flexion with secondary contractures and painful spasms: paraplegia in flexion).

The amount and pattern of spasticity depends on the location of the lesion and tends to be greater with spinal cord than cortical lesions. Scales to quantitate spasticity are available (Ashworth, modified Ashworth, Wartenberg pendulum test) but have shortcomings. Spasticity may also vary in distribution: for lesions above the spinal cord it typically affects the arm flexors and the leg extensors to a greater extent (hemiparetic posture).

Spasticity is a clinical feature of upper motor neurone syndromes, which may be accompanied by both positive (hypertonus, clonus, limb hyperreflexia, Babinski's sign, flexor or extensor spasms) and negative phenomena (weakness in a pyramidal distribution, motor underactivity): the latter may be more significant determinants of disability. Slow, laboured speech, with slow voluntary tongue movements, may be referred to as spastic dyasrthria, which may occur in the context of a pseudobulbar palsy.

The pathogenesis of spasticity has traditionally been ascribed to damage to the corticospinal and/or corticobulbar pathways at any level from cerebral cortex to spinal cord. However, various lines of evidence (*e.g.* the failure of pyramidotomy to produce spasticity in animals, rare human cases of isolated pyramid infarction causing

hyperreflexia and weakness without spasticity) has led to the implication of other motor tracts in the genesis of spasticity, *viz.:*

- the dorsal reticulospinal tract, which lies in the lateral funiculus of the cord and hence is often damaged concurrently with the adjacent lateral corticospinal tract (*e.g.* in MS, which seems to have a predilection for the lateral funiculus); this descending pathway has an inhibitory effect on stretch reflexes which is under cortical control;
- the medial reticulospinal tract and vestibulospinal tracts which are not under cortical control and whose excitatory effects on extensor tone may remain unopposed.

Physiologically, spasticity has been characterized as an exaggeration of the muscle stretch reflexes, with reduced threshold (hyperexcitable □-motor neurones) and abnormal reflex transmission (increased gain). The role of neurotransmitters (glutamate, glycine, catecholamines, serotonin) in the pathogenesis of spasticity is unclear, but the efficacy of baclofen (a $GABA_B$ agonist) and benzodiazepines suggest impaired GABAergic transmission may contribute, perhaps through a loss of presynaptic inhibition mediated by interneurones or the inhibition of glutamate release.

Treatment of severe spasticity, for example in multiple sclerosis, often requires a multidisciplinary approach. Urinary infection, constipation, skin ulceration and pain can all exacerbate spasticity, as may inappropriate posture; appropriate management of these features may ameliorate spasticity. Drugs which may be useful include baclofen, dantrolene (a blocker of muscle excitation-contraction coupling), and tizanidine (□$_2$-adrenoreceptor agonist). Intrathecal baclofen may also be of benefit in selected cases, and for focal spasticity injections of botulinum toxin may be appropriate. For painful immobile spastic legs with reflex spasms and double incontinence, irreversible nerve injury with intrathecal phenol may be advocated to relieve symptoms. The place of cannabinoids has yet to be determined but is currently under investigation.

- Brown P. Pathophysiology of spasticity. *Journal of Neurology, Neurosurgery and Psychiatry* 1994; **57**: 773-7
- Lance JW. Symposium synopsis. In: *Spasticity: disordered motor control.* (Feldman RG, Young RR, Koella WP, eds.) Chicago: Year Book Medical Publishers 1980: 485-94
- Spasticity: current and future management. *Hospital Medicine* 1998; **59**: 61-70
[Cross References: BABINSKI'S SIGN; CLASP-KNIFE PHENOMENON; CLONUS; CONTRACTURE; DYSARTHRIA; HYPERREFLEXIA; HYPERTONUS; PARAPLEGIA; PSEUDOBULBAR PALSY; REFLEXES; SPASM; UPPER MOTOR NEURONE SYNDROME; WEAKNESS]

Speech Apraxia

Speech apraxia is one of the labels applied to a disorder of communication characterized by slow speech tempo ("groping for words"), impaired articulation, and dysprosody, with relatively intact language function and no dysgraphia. More errors occur with increasing articulatory complexity (consonant clusters *vs.* single consonants). Automatic or reactive speech (*e.g.* expletives, cliches) is without error. This,

or a very similar, constellation of features has also been known as cortical dysarthria, aphemia, or phonetic disintegration. There may be associated orofacial apraxia.

Speech apraxia is associated with inferior frontal dominant (left) hemisphere damage in the region of the lower motor cortex or frontal operculum; it has been claimed that involvement of the anterior insula is specific for speech apraxia.

The exact nosological status of this entity remains in some doubt. The syndrome is thought to reflect disturbances of planning articulatory and phonatory functions, but is most often encountered as part of a non-fluent aphasia.

- Dronkers NF. A new brain region for coordinating speech articulation. *Nature* 1996; **384**: 159-61

[Cross References: APHASIA; APHEMIA; APRAXIA]

Spinal Mass Reflex

The spinal mass reflex is involuntary flexion of the trunk in a comatose patient, such that they appear to be attempting to sit up.

[Cross References: COMA]

Square-Wave Jerks

Square-wave jerks reflect an instability of ocular fixation, consisting of small saccades which interrupt fixation, moving the eye away from the primary position and then returning; in other words, they represent a disorder of saccadic eye movements in which there is a saccadic interval (of about 200 msec; *cf.* ocular flutter, opsoclonus). Very frequent square-wave jerks may be termed square-wave oscillations. Very obvious square-wave jerks (amplitude > 7°) are termed macro-square-wave jerks.

Square-wave jerks are often best appreciated at ophthalmoscopy. Their name derives from the appearance they produce on electro-oculographic recordings.

Although square-wave jerks may be normal in elderly individuals, they may be indicative of disease of the cerebellum or brainstem, *e.g.* Huntington's disease, Parkinson's disease, Steele-Richardson-Olszewski syndrome, cerebellar degeneration.

[Cross References: NYSTAGMUS; OCULAR FLUTTER; OPSOCLONUS; SACCADIC INTRUSION]

Square-Wave Oscillations

- see SQUARE-WAVE JERKS

Squint

- see HETEROTROPIA

Stellwag's Sign

Stellwag's sign is a widening of the palpebral fissure due to upper eyelid retraction. Along with a reduced blink rate, this creates a very typical staring, "astonished", facies. The clinical phenomena of Stellwag's sign overlap with those labelled as the sunset sign.

Stellwag's sign is seen in Steele-Richardson-Olszewski syndrome, and thyroid eye disease.

[Cross References: BLINKING; LID LAG; LID RETRACTION; SUNSET SIGN]

Steppage, Stepping Gait

Steppage or stepping gait occurs with a lower motor neurone type of foot drop ("floppy" foot drop), *e.g.* due to a common peroneal nerve palsy, peripheral neuropathies. Because of the weakness of foot dorsiflexion (weak tibialis anterior) there is compensatory overaction of hip and knee flexors during the swing phase of walking to ensure the foot clears the ground. In the strike phase, there is a characteristic slapping down of the foot, again a consequence of weak ankle dorsiflexion. Proprioceptive loss, as in dorsal column spinal disease, may also lead to a gait characterized by high lifting of the feet, and also stomping down of the foot in the strike phase.

The pattern of gait with upper motor neurone foot drop ("stiff" foot drop), *e.g.* due to a corticospinal tract lesion, is quite different, with the foot being dragged, sometimes with circumduction of the leg. This may leads to falls as a consequence of tripping over the foot, especially on up-hill gradients, and a characteristic pattern of wear on the point of the shoe.

[Cross References: FOOT DROP; LOWER MOTOR NEURONE SYNDROME; PROPRIOCEPTION; ROMBERGISM, ROMBERG'S SIGN; UPPER MOTOR NEURONE SYNDROME]

Stereoanaesthesia

- see ASTEREOGNOSIS

Stereohypaesthesia

- see ASTEREOGNOSIS

Stereotypy

Stereotypies may be defined as regular repeated movements, which are voluntary but not apparently goal-directed, and which may be carried out in a uniform pattern for long periods of time (*cf.* tic). Whole areas of the body may be involved by Stereotypies and hence this movement is more complex than a tic.

Stereotypies are common in patients with learning disability and schizophrenia. The term has also been used to describe movements associated with chronic neuroleptic use. Very characteristic manual Stereotypies (washing, rubbing movements: "hand washing") may be seen in Rett's disease.

Verbal Stereotypies are reiterated words or syllables produced by patients with profound non-fluent aphasia.

- Jankovic J. Stereotypies. In: Marsden CD, Fahn S (eds.). *Movement disorders 3.* Boston: Butterworth 1994: 503-17
- Lees AJ. *Tics and related disorders.* Edinburgh: Churchill Livingstone 1985

[Cross References: APHASIA; BROCA'S APHASIA; TIC]

Stiffness

Stiffness of muscles occurs as a feature of all pyramidal and extrapyramidal disorders (as spasticity and rigidity, respectively), but the term stiffness is usually reserved for disorders in which stiffness is the principal symptom due to continuous motor unit activity within muscles. This may be primarily of muscular origin (myotonia) or of

neural origin (myokymia, neuromyotonia). Accompanying signs may prove helpful in diagnosis, such as slow muscle relaxation (myotonia), percussion irritability of muscle, and spontaneous and exertional muscle spasms. Hyperlordotic posture is typical of stiff man/stiff person syndrome. Stiffness must be differentiated from both rigidity and spasticity.
Recognised causes of stiffness include:

- Stiff man/stiff person syndrome
- Stiff limb syndrome
- Progressive encephalomyelitis with rigidity
- Neuromyotonia (Isaac's syndrome; armadillo syndrome)
- Schwartz-Jampel syndrome (chondrodystrophic myotonia)
- Tetanus
- Strychnine poisoning

The stiff man/stiff person syndrome is probably autoimmune in its pathogenesis since it is strongly associated with insulin-dependent diabetes mellitus and the presence of antibodies to glutamic acid decarboxylase (anti-GAD antibodies), the enzyme in the synthetic pathway of GABA.

- Barker R, Revesz T, Thorn M, Marsden CD, Brown P. Review of 23 patients affected by the stiff man syndrome: clinical subdivision into stiff trunk (man) syndrome, stiff limb syndrome, and progressive encephalomyelitis with rigidity. *Journal of Neurology, Neurosurgery and Psychiatry* 1998; **65**: 633-40
- Scolding NJ. Stiff man syndrome. In: Scolding NJ (ed.). *Immunological and inflammatory disorders of the central nervous system.* Oxford: Butterworth Heinemann 1999: 139-46
- Thompson PD. Stiff people. In: Marsden CD, Fahn S (eds.). *Movement disorders 3.* Boston: Butterworth 1994: 373-405
[Cross References: MYOKYMIA; MYOTONIA; NEUROMYOTONIA; PARAMYOTONIA; RIGIDITY; SPASTICITY]

Stomping
- see STEPPAGE, STEPPING GAIT

Strabismus
- see HETEROPHORIA; HETEROTROPIA

Straight Leg Raising
- see LASEGUE'S SIGN

"Straight Thumb Sign"
Median nerve lesions in the forearm cause weakness of flexor pollicis longus, which normally flexes the distal phalanx of the thumb. Hence the thumb remains straight when the patient attempts to grasp something or make a fist. The "pinch sign" may also be present.

- Cherington M. Anterior interosseous nerve syndrome straight thumb sign. *Neurology* 1977; **27**: 800-1
[Cross References: "PINCH SIGN"]

Striatal Toe
Striatal toe refers to the tonic extension of the hallux which is seen in dystonic syndromes, and as a feature of extrapyramidal disorders.
Striatal toe may be confused with Babinski's sign (extensor plantar response), the principal difference being that the latter is elicited by stimulation whereas the former is a tonic response.

- Winkler AS, Reuter I, Harwood G, Clough C, Chaudhuri KR. The frequency and significance of "striatal toe" in parkinsonism. *Journal of Neurology, Neurosurgery and Psychiatry* 1999; **66**: 271
[Cross References: BABINSKI'S SIGN; PARKINSONISM]

Stupor
Stupor is a state of altered consciousness characterized by deep sleep or unresponsiveness, susceptible to arousal only by vigorous and/or repeated stimuli, with lapse back into unresponsiveness when the stimulus stops. Stupor is a less severe impairment of conscious level than coma, but worse than obtundation (torpor). It is suggestive of diffuse cerebral dysfunction, *e.g.* drug-induced.
[Cross References: COMA; DELIRIUM; ENCEPHALOPATHY; OBTUNDATION]

Stutter
Stutter, one of the reiterative speech disorders, is usually a developmental problem, but may be acquired in aphasia with unilateral or bilateral hemisphere lesions (*e.g.* vascular damage, trauma, Alzheimer's disease, Parkinson's disease, Steele-Richardson-Olszewski syndrome). Unlike developmental stutter, acquired stutter may be evident throughout sentences, rather than just at the beginning. Furthermore, developmental stutter tends to occur more with plosives (phonemes where the flow of air is temporarily blocked and suddenly released, as in 'p', 'b'), whereas acquired stutter is said to affect all speech sounds fairly equally. Cessation of developmental stutter following bilateral thalamic infarction in adult life has been reported.

- Fleet WS, Heilman KM. Acquired stuttering from a right hemisphere lesion in a right-hander. *Neurology* 1985; **35**: 1343-6
- Muroi A, Hirayama K, Tanno Y, Shimizu S, Watanabe T, Yamamoto T. Cessation of stuttering after bilateral thalamic infarction. *Neurology* 1999; **53**: 890-1
[Cross References: APHASIA; ECHOLALIA; PALILALIA]

"Sundowning"
"Sundowning" is increased confusion in the late afternoon, evening, and night-time, which may be seen in patients with delirium, and sometimes dementia. EEG recordings in delirious patients may show disordered circadian rhythms, the probable physiological correlate of "sundowning".
[Cross References: DELIRIUM; DEMENTIA]

Sunset Sign
The sunset sign consists of retraction of the upper eyelids, downturning of the eyes, with paralysis of upgaze, which results in a staring appearance. This may be seen in untreated hydrocephalus in children, but also progressive supranuclear palsy (Steele-Richardson-Olszewski syndrome; Stellwag's sign).
[Cross References: LID RETRACTION; STELLWAG'S SIGN]

Suppression
- see EXTINCTION

Supranuclear Gaze Palsy
A supranuclear gaze palsy results from pathology located above the nuclei of the nerves supplying the extraocular muscles. Voluntary gaze is impaired while the integrity of the oculomotor nuclei and infranuclear connections may be demonstrated by the preservation of the vestibulo-ocular reflexes (VOR), overcoming the ophthalmoplegia, at least in the early stages (*e.g.* the supranuclear gaze palsy in the vertical plane in Steele-Richardson-Olszewski syndrome).
Supranuclear gaze palsies may be:

Horizontal:
Hemisphere (frontal) lesion: eyes deviated to the side of the lesion, or in the case of an irritative (*e.g.* epileptic) focus away from the side of the lesion;
Paramedian pontine reticular formation: eyes deviated to contralateral side.

Vertical:
Brainstem compression/distortion;
Dorsal upper midbrain (*e.g.* rostral interstitial nucleus of the median longitudinal fasciculus; pineal lesion causing Parinaud's syndrome).

Causes of supranuclear gaze palsy include:

- Steele-Richardson-Olszewski syndrome (progressive supranuclear palsy, PSP);
- Creutzfeldt-Jakob disease;
- Corticobasal degeneration;
- Progressive subcortical gliosis of Neumann;
- Adult-onset Niemann-Pick disease;
- Gaucher's disease.

- Lees AJ. The Steele-Richardson-Olszewski syndrome (progressive supranuclear palsy). In: Marsden CD, Fahn S (eds.). *Movement Disorders 2.*London, Butterworth 1987: 272-87
[Cross References: GAZE PALSY; PARKINSONISM; VESTIBULO-OCULAR REFLEXES]

Suspended Sensory Loss
- see DISSOCIATED SENSORY LOSS; MYELOPATHY

Swinging Flashlight Sign

- see AFFERENT PUPILLARY DEFECT; MARCUS GUNN PUPIL, MARCUS GUNN SIGN

Synaesthesia

Synaesthesia is a perceptual experience in one sensory modality following stimulation of another sensory modality. The most commonly encountered example is colour-word Synaesthesia ("coloured hearing"), *i.e.* experiencing a colour on hearing a particular word. Known synaesthetes include the composers Messaien and Scriabin, and the author Nabokov.

Neuropsychologically this phenomenon, which occurs in a small percentage of the normal population, has been conceptualized as a break down of modularity. Functional imaging studies of colour-word synaesthetes show activation of visual associative areas of cortex (but not primary visual cortex), as well as perisylvian language areas, when listening to words which evoke the experience of colour.

- Baron-Cohen S, Harrison JE (eds.). *Synaesthesia: classic and contemporary readings.* Oxford: Blackwell 1997

- Paulesu E, Harrison J, Baron-Cohen S, *et al.* The physiology of coloured hearing: a PET activation study of colour-word Synaesthesia. *Brain* 1995; **118:** 661-76

[Cross References: PHOSPHENE]

Synkinesia, Synkinesis

Synkinesis refers to involuntary movements which accompany or are associated with certain voluntary movements (*mitbewegungen,* motor overflow). These may be normal, for example the swinging of the arms when walking, or abnormal, *e.g.* involuntary contraction of orbicularis oculi when opening the mouth (the Marin-Amat syndrome: inverse Marcus Gunn phenomenon), acquired after lower motor neurone facial (VII) nerve palsies and presumed to reflect aberrant reinnervation. Crocodile tears (lacrimation when salivating), another reflection of aberrant reinnervation following a lower motor neurone facial nerve palsy, may also fall under this rubric, although there is no movement *per se* (autonomic synkinesis). Aberrant regeneration of the oculomotor (III) nerve can lead to pseudo-von Graefe's sign.

Abnormal synkinesis may be useful is assessing whether weakness is organic or functional (*cf.* Hoover's sign).

Synkinesis may also refer to the aggravation of limb rigidity detected when performing movements in the opposite limb (*e.g.* clenching and relaxing the fist); this is also known as activated rigidity, or Froment's sign.

[Cross References: CROCODILE TEARS; FROMENT'S SIGN; HOOVER'S SIGN; JAW WINKING; PSEUDO-VON GRAEFE'S SIGN; RIGIDITY]

T

Talantropia
- see NYSTAGMUS

Tandem Gait
Tandem gait is the ability to walk along a straight line placing one foot directly in front of the other, heel to toe, which may be likened to walking a tightrope. In ataxic disorders (midline cerebellum or loss of proprioception) in which axial coordination is most affected, the ability to tandem walk is impaired, reflecting the tendency of such patients to compensate for their incoordination by developing a broad based gait.
[Cross References: ATAXIA; CEREBELLAR SYNDROMES]

Teichopsia
- see FORTIFICATION SPECTRA

Telegraphic Speech
- see AGRAMMATISM

Temporal Desaturation
Temporal desaturation refers to an impairment in perception of red targets confined to the temporal hemifield. This may be the earliest indication of a developing temporal field defect, as in a bitemporal hemianopia due to a chiasmal lesion, or a monocular temporal field defect (junctional scotoma of Traquair) due to a distal ipsilateral optic nerve lesion.
[Cross References: HEMIANOPIA; SCOTOMA]

Tetanus, Tetany
- see *MAIND'ACCOUCHEUR*; RISUS SARDONICUS; SPASM

Tetraparesis, Tetraplegia
- see QUADRIPARESIS, QUADRIPLEGIA

Threat Reflex
- see BLINK REFLEX

Tic
A tic is an abrupt, jerky repetitive movement involving discrete muscle groups (hence a less complex movement than a stereotypy). Vocal tics are also described. Tics vary in intensity, lack rhythmicity, and are relatively easy to imitate. They may be temporarily suppressed by will power but this is usually accompanied by a growing inner tension or restlessness, only relieved by the performance of the movement.

The pathophysiology of tics is uncertain; the belief that Gilles de la Tourette syndrome was a disorder of the basal ganglia has now been superseded by evidence of dysfunction within the cingulate and orbitofrontal cortex, perhaps related to excessive endorphin release.

The aetiological differential diagnosis of tic includes:

- idiopathic;
- Gilles de la Tourette syndrome;
- tics related to structural brain damage;
- drug-induced tics

Treatment of tics is most usually with dopamine antagonists (haloperidol, sulpiride) and opioid antagonists (naltrexone); clonidine (central α_2 adrenergic receptor antagonist) and tetrabenazine (dopamine-depleting agent) have also been reported to be beneficial on occasion.

The word tic has also been used to describe the paroxysmal, lancinating pains of trigeminal neuralgia (tic douloureux).

- Lees AJ. *Tics and related disorders.* Edinburgh: Churchill Livingstone 1985
- Weeks RA, Turjanski N, Brooks DJ. Tourette's syndrome: a disorder of cingulate and orbitofrontal function? *Quarterly Journal of Medicine* 1996; **89:** 401-8
[Cross References: KLAZOMANIA; STEREOTYPY]

"Tie Sign"

- see VISUAL DISORIENTATION

Tinel's Sign (Hoffmann-Tinel Sign)

Tinel's sign (Hoffmann-Tinel sign) is present when tingling (paraesthesia) is experienced when tapping lightly with a finger or a tendon hammer over a compressed or regenerating peripheral nerve. The tingling (Tinel's "sign of formication") is present in the cutaneous distribution of the damaged nerve ("peripheral reference"). Although originally described in the context of peripheral nerve regeneration after injury, Tinel's sign may also be helpful in diagnosing focal entrapment neuropathy such as carpal tunnel syndrome. However, it is a "soft" sign; like other provocative tests for carpal tunnel syndrome (*e.g.* Phalen's sign) it is not as reliable for diagnostic purposes as EMG. One study found a specificity of 59-77%, and sensitivity of 60-67%.

A "motor Tinel sign" has been described, consisting of motor EMG activity and jerking of muscles evoked by manipulation of an entrapped nerve trunk.

The neurophysiological basis of Tinel's sign is presumed to be the lower threshold of regenerating or injured (demyelinated) nerves to mechanical stimuli, which permits ectopic generation of orthodromic action potentials, as in Lhermitte's sign.

- Heller L, Ring H, Costeff H, Solzi P. Evaluation of Tinel's and Phalen's signs in the diagnosis of the carpal tunnel syndrome. *European Neurology* 1986; **25**: 40-2
- Montagna P. Motor Tinel sign: a new localising sign in entrapment neuropathy. *Muscle Nerve* 1994; **17**: 1493-4

- Smith KJ. Conduction properties of central demyelinated axons: the generation of symptoms in demyelinating disease. In: Bostock H, Kirkwood PA, Pullen AH (eds.). *The neurobiology of disease: contributions from neuroscience to clinical neurology.* Cambridge, CUP 1996: 95-117
[Cross References: LHERMITTE'S SIGN, PHALEN'S SIGN]

Tinnitus
Tinnitus is hearing non-environmental sounds in the ear. This is most usually a subjective phenomenon (*i.e.* heard only by the sufferer) accompanying either conductive or sensorineural hearing loss. However, in about one-fifth of sufferers, tinnitus is objective (*i.e.* heard also by an observer). This may result from:

- vascular causes: *e.g.* arteriovenous malformation, fistula; carotid or vertebrobasilar bruit;
- mechanical causes: *e.g.* palatal myoclonus.

The common causes of subjective tinnitus are:

- middle/inner ear disease: cochlear hydrops (Menière's disease), presbycusis, acoustic tumour;
- pulsatile: normal heartbeat, glomus jugulare tumour, raised intracranial pressure, cervical/intracranial aneurysm, arteriovenous malformation.

[Cross References: PALATAL MYOCLONUS]

"Tip-Of-The-Tongue" Phenomenon
- see CIRCUMLOCUTION

Titubation
- see HEAD TREMOR

Todd's Paresis
Todd's paresis (Todd's paralysis) is a transient localised weakness (usually hemiparesis), lasting seconds to minutes (exceptionally 24 to 48 hours), observed following a focal motor seizure or Jacksonian seizure originating in the central motor strip, or febrile convulsion. The phenomenon was first described by RB Todd in 1854. The pattern and duration of post-ictal signs is quite heterogeneous. Aphasia is also described. A postictal "paralytic" conjugate ocular deviation may be observed after adversive seizures.

- Rolak LA, Rutecki P, Ashizawa T, Harati Y. Clinical features of Todd's post-epileptic paralysis. *Journal of Neurology, Neurosurgery and Psychiatry* 1992; **55:** 63-4
[Cross References: HEMIPARESIS; SEIZURES]

Torpor
- see OBTUNDATION

Torticollis

Torticollis (wryneck, nuchal dystonia) is an involuntary movement disorder characterized by cervical dystonia, causing contraction of neck musculature (especially sternocleidomastoid, trapezius, and splenius capitis). In the majority of cases (> 50%) this produces rotation, but laterocollis, retrocollis, tremulous ("no-no") and complex (*i.e.* variable) forms are seen; antecollis is unusual. Contractions are usually unilateral, may be associated with local pain, and may be relieved by a "sensory trick" (*geste antagoniste*).

Causes of torticollis include:

- Idiopathic (the majority)
- Secondary to acquired cervical spine abnormalities, trauma
- Cervical spinal tumour
- Tardive effect of neuroleptics

The treatment of choice is botulinum toxin injections into affected muscles, which helps up to 70-80% of patients, but need to be repeated every three months or so.

[Cross References: ANTECOLLIS; DYSTONIA; *GESTE ANTAGONISTE;* LATEROCOLLIS; RETROCOLLIS]

Tortopia

- see ENVIRONMENTAL TILT

Transcortical Aphasia

Transcortical aphasia is defined as an aphasia in which there is a dissociation between preserved repetition (*cf.* conduction aphasia) and impaired fluency (transcortical motor aphasia, TCMA) or comprehension (transcortical sensory aphasia, TCSA). TCMA shows similarities with Broca's aphasia; TCSA with Wernicke's aphasia. TCMA is associated with infarction in the supplementary motor area, superior to Broca's area; TCSA with infarction in the parieto-temporal border zone. Hence, some authorities prefer to label these conditions as extrasylvian aphasic syndromes, to distinguish them from the perisylvian aphasic syndromes (Broca, Wernicke, conduction); moreover, these syndromes are not "transcortical" in any literal sense.

Dynamic aphasia (*q.v.*) may be a lesser version of TCMA, in which there are no paraphasias and minimal anomia, preserved repetition and automatic speech, but reduced spontaneous speech. This may be associated with lesions of dorsolateral prefrontal cortex ("frontal aphasia") in the context of frontal lobe degeneration. There may be incorporational echolalia, when the patient uses the examiner's question to help form an answer.

- Boatman D, Gordon B, Hart J, Seines O, Miglioretti D, Lenz F. Transcortical sensory aphasia: revisited and revised. *Brain* 2000; **123**: 1634-42

[Cross References: APHASIA; BROCA'S APHASIA; CONDUCTION APHASIA; DYNAMIC APHASIA; ECHOLALIA; PARAPHASIA; WERNICKE'S APHASIA]

Tremblement affirmatif, Tremblement negatif

- see HEAD TREMOR

Tremor

Tremor is an involuntary movement, roughly rhythmic and sinusoidal, although some tremors (*e.g.* dystonic) are irregular in amplitude and periodicity. Tremors may be classified clinically:

~ *Rest tremor*: present when a limb is supported against gravity and there is no voluntary muscle activation, *e.g.* the 3.5-7 Hz "pill-rolling" hand tremor of Parkinson's disease; midbrain/rubral tremor.

~ *Action tremor:* present during any voluntary muscle contraction; various subtypes are recognised:

- *Postural tremor*: present during voluntary maintenance of a posture opposed by gravity, *e.g.* arm tremor of essential tremor; 6Hz postural tremor sometimes seen in Parkinson's disease, which may predate emergence of akinesia/rigidity/rest tremor; modest postural tremor of cerebellar disease; some drug-induced tremors (including alcohol withdrawal, delirium tremens); tremor of IgM paraproteinaemic neuropathy;
- *Kinetic tremor:* present with movement, often with an exacerbation at the end of a goal-directed movement (intention tremor), *e.g.* cerebellar/midbrain tremor (3-5Hz);
- *Task-specific tremor:* evident only during the performance of a highly-skilled activity, *e.g.* primary writing tremor;

~ *Isometric tremor:* present when voluntary muscle contraction is opposed by a stationary object, *e.g.* primary orthostatic tremor (14-18Hz).

~ *Psychogenic tremors:* these are difficult to classify, with changing characteristics; the frequency with which such tremors are observed varies greatly between different clinics.

EMG may be useful for determining tremor frequency, but is only diagnostic in primary orthostatic tremor.

Various treatments are available for tremor, with variable efficacy. Essential tremor often responds to alcohol, and this is a reasonable treatment (previous anxieties that such a recommendation would lead to alcoholism seem unjustified); alternatives include propranolol, primidone, alprazolam, flunarizine, and nicardipine. In Parkinson's disease, tremor is less reliably responsive to levodopa preparations than akinesia and rigidity; anticholinergics such as benzhexol may be more helpful (but may cause confusion). Primary orthostatic tremor has been reported to respond to clonazepam, primidone, and levodopa. Cerebellar tremor is often treated with isoniazid, but seldom with marked benefit, likewise limb weights; stereotactic surgery may be the optimum treatment if preliminary data are confirmed.

- Bain PG, Findley LJ. *Assessing Tremor Severity.* London: Smith-Gordon 1993
- Deuschl G, Bain P, Brin M and an Ad Hoc Scientific Committee. Consensus statement of the Movement Disorder Society on tremor. *Movement Disorders* 1998; **13(suppl3):** 2-23
- Findley LJ, Koller WC (eds.). *Handbook of Tremor Disorders.* New York: Marcel Dekker, Inc. 1995

[Cross References: HEAD TREMOR; KNEE TREMOR; PARKINSONISM; VOCAL TREMOR, VOICE TREMOR]

Trendelenburg's Sign
Trendelenburg's sign is tilting of the pelvis toward the side of the unaffected raised leg in a unilateral superior gluteal nerve lesion.

Trismus
Trismus is an inability to open the jaw due to tonic spasm of the masticatory muscles (principally masseter and temporalis) effecting forced jaw closure ("lockjaw"): This may be due to dystonia of the jaw muscles (*e.g.* drug-induced dystonic reaction), other neuromuscular diseases (polymyositis, tetanus, nemaline myopathy, trauma to the muscles of mastication, rabies, strychnine poisoning), infection in the pterygomandibular space, and central disorders (brainstem encephalopathy, multiple sclerosis, pseudobulbar palsy).

- Lai MM, Howard RS. Pseudobulbar palsy associated with trismus. *Postgraduate Medical Journal* 1994; **70**: 823-4
[Cross References: DYSTONIA; PSEUDOBULBAR PALSY]

Trombone Tongue
Trombone tongue refers to an irregular involuntary darting of the tongue in and out of the mouth when the patient is requested to keep the tongue protruded. This sign may be seen in choreiform movement disorders such as Huntington's disease and neuroacanthocytosis.
[Cross References: CHOREA; IMPERSISTENCE; MILKMAID'S GRIP]

Trömner's Sign
Trömner's sign is flexion of the thumb and index finger in response to tapping or flicking the volar surface of the distal phalanx of the middle finger held partially flexed between the examiner's finger and thumb. This is an alternative method for eliciting the finger flexor response to Hoffmann's sign ("snapping" the distal phalanx). As in the latter, it is suggestive of a corticospinal tract (upper motor neurone) lesion above C5 or C6, especially if unilateral, although it may be observed in some normal individuals.
[Cross References: HOFFMANN'S SIGN; UPPER MOTOR NEURONE SYNDROME]

Trousseau's Sign
- see *MAIN D'ACCOUCHEUR*

Tullio Phenomenon
The Tullio phenomenon is the experience of vestibular symptoms and signs (vertigo, nystagmus, oscillopsia, postural imbalance, ocular tilt reaction, +/- skew deviation) on exposure to high intensity acoustic stimuli, presumed to be due to hyperexcitability of the normal vestibular response to sound, causing pathological stimulation of the semicircular canals and/or otoliths. This unusual phenomenon may be associated

with perilymph leaks or a defect in the capsule forming the roof of the anterior semicircular canal. The sound sensitivity is probably at the level of the receptors rather than the vestibular nerve.

- Watson SRD, Halmagyi GM, Colebatch JG. Vestibular hypersensitivity to sound (Tullio phenomenon). Structural and functional assessment. *Neurology* 2000; **54:** 722-8

[Cross References: NYSTAGMUS; OCULAR TILT REACTION; OSCILLOPSIA; SKEW DEVIATION; VERTIGO]

"Tunnel Vision"

A complaint of "tunnel vision" may indicate constriction of the visual field. This may be observed, for example, with enlargement of the blind spot and papilloedema as a consequence of raised intracranial pressure, in which case the field enlarges the further away from the eye the target is held (funnel vision). In non-organic visual impairment, by contrast, the visual field stays the same size with more distant targets (tunnel vision). A tunnel vision phenomenon has also been described as part of the aura of seizures of anteromedial temporal and occipitotemporal origin.

[Cross References: AURA; BLIND SPOT; HEMIANOPIA; PAPILLOEDEMA]

Two-point Discrimination

Two-point discrimination is the ability to discriminate two adjacent point stimuli (*e.g.* using a pair of calipers) as two rather than one. The minimum detectable distance between the points (acuity) is smaller in the skin of the finger tips (*i.e.* greater acuity) than, say, the skin on the back of the trunk. Impairments of two-point discrimination may occur with dorsal column spinal cord lesions, in which proprioception (and possibly vibration) is also impaired. Cortical parietal lobe lesions may produce a cortical sensory syndrome of astereognosis, agraphaesthesia, and impaired two-point discrimination.

[Cross References: ASTEREOGNOSIS; GRAPHAESTHESIA; PROPRIOCEPTION; VIBRATION]

U

Uhthoff's Phenomenon

Uhthoff's phenomenon (symptom) is the worsening of visual acuity ("amblyopia" in Uhthoff's 1890 description) with exercise in optic neuritis, reflecting the temperature sensitivity of demyelinated axons (*i.e.* reduced safety factor for faithful transmission of action potentials). The term has subsequently been applied to exercise and/or temperature related symptoms in other demyelinated pathways. It has also been described in the context of other optic nerve diseases, including Leber's hereditary optic neuropathy, sarcoidosis and tumour.

Evidence suggesting that Uhthoff's phenomenon is associated with an increased incidence of recurrent optic neuritis, and may be a prognostic indicator for the development of multiple sclerosis, has been presented.

- Guthrie TC, Nelson DA. Influence of temperature changes on multiple sclerosis: critical review of mechanisms and research potential. *Journal of the Neurological Sciences* 1995; **129**: 1-8
- Scholl GB, Song HS, Wray SH. Uhthoff's symptom in optic neuritis: relationship to magnetic resonance imaging and development of multiple sclerosis. *Annals of Neurology* 1991; **30**: 180-4
- Selhorst JB, Saul RF. Uhthoff and his symptom. *Journal of Neuro-Ophthalmology* 1995; **15**: 63-9 (erratum: *Journal of Neuro-Ophthalmology* 1995; **15**: 264)

[Cross References: LHERMITTE'S SIGN; PHOSPHENE]

Unterburger's Sign

Unterburger's test examines the integrity of vestibulospinal connections and attempts to define the side of a vestibular lesion. The patient is asked to march on the spot with the eyes closed (*i.e.* proprioceptive and visual cues are removed); the patient will rotate to the side of a unilateral vestibular lesion (Unterburger's sign). The test is not very useful, particularly in chronic, progressive, or partially compensated vestibular lesions.

[Cross References: PROPRIOCEPTION; VERTIGO]

Urgency

- see INCONTINENCE

Urinary Retention

Although urinary retention is often urological in origin (*e.g.* prostatic hypertrophy) or be a side effect of drugs *(e.g.* anticholinergics), it may have neurological causes. It may be a sign of acute spinal cord compression, with or without other signs in the lower limbs, or of acute cauda equina compression, for example with a central L1 disc herniation. Sometimes the level of the pathology is several segments above that

expected on the basis of the ("false localising") neurological signs. Loss of awareness of bladder fullness may lead to retention of urine with overflow.

A syndrome of urinary retention in young women has been described, associated with myotonic-like activity on sphincter EMG; this condition may be associated with polycystic ovary disease and is best treated with clean intermittent self-catheterisation.

- Fowler CJ. Investigaion of the neurogenic bladder. In: Hughes RAC (ed.). *Neurological Investigations.* London: BMJ Publishing 1997: 397-414
- Jamieson DRS, Teasdale E, Willison HJ. False localising signs in the spinal cord. *BMJ* 1996; **312:** 243-4
- Johnston RA. Acute spinal cord compression. In: Hughes RAC (ed.). *Neurological Emergencies.* London: BMJ Publishing 1997 (2nd edition): 272-94

[Cross References: CAUDA EQUINA SYNDROME; "FALSE LOCALISING SIGNS"; INCONTINENCE; MYELOPATHY; PARAPLEGIA; RADICULOPATHY]

Upper Motor Neurone Syndrome

An upper motor neurone (UMN) syndrome constitutes a constellation of motor signs resulting from damage to upper motor neurone pathways, *i.e.* proximal to the anterior horn cell. These may be termed "pyramidal signs", but since there are several descending motor pathways (*e.g.* corticospinal, reticulospinal, vestibulospinal), of which the pyramidal or corticospinal pathway is just one, "upper motor neurone syndrome" is preferable. "Long tract signs" may be a more accurate term, but it is often used interchangeably with "pyramidal signs". The syndrome may be variable in its clinical features but common elements, following the standard order of neurological examination of the motor system, include:

- APPEARANCE - usually normal, but there may be wasting in chronic UMN syndromes, but this is usually not as evident as in lower motor neurone syndromes; contractures may be evident in chronically spastic limbs;
- TONE - hypertonus, with spasticity, clasp-knife phenomenon and sustained clonus;
- POWER - weakness, often in a so-called pyramidal distribution (*i.e.* affecting extensors more than flexors in the upper limb, and flexors more than extensors in the lower limb); despite its clinical utility, the term pyramidal is, however, a misnomer (see Weakness);
- CO-ORDINATION - depending on the degree of weakness, it may not be possible to comment on the integrity of co-ordination in UMN syndromes; in a pure UMN syndrome co-ordination will be normal, but syndromes with both ataxia and UMN features do occur (*e.g.* spinocerebellar syndromes, ataxic hemiparesis syndromes);
- REFLEXES - limb hyperreflexia, sometimes with additional reflexes indicative of corticospinal tract involvement (Hoffmann's sign, Trömner's sign, crossed adductor reflex); an extensor plantar response (Babinski's sign); cutaneous reflexes (abdominal, cremasteric) are lost.

The most reliable ("hardest") signs of UMN syndrome are increased tone, clonus, and upgoing plantar responses.

These features help to differentiate UMN from LMN syndromes, although clinically the distinction is not always easy to make: a "pyramidal" pattern of weakness may occur in LMN syndromes (*e.g.* Guillain-Barré syndrome) and acute UMN syndromes may cause flaccidity and areflexia (*e.g.* "spinal shock").
[Cross References: ABDOMINAL REFLEXES; ATAXIC HEMIPARESIS; BABINSKI'S SIGN; CLASP-KNIFE; CLONUS; CONTRACTURE; CREMASTERIC REFLEX; HOFFMANN'S SIGN; HYPERREFLEXIA; HYPERTONIA, HYPERTONUS; LOWER MOTOR NEURONE SYNDROME; PSEUDOBULABR PALSY; SPASTICITY; TRÖMNER'S SIGN; UPPER MOTOR NEURONE SYNDROME; WEAKNESS]

Utilization Behaviour
Utilization behaviour is a disturbed response to external stimuli, a component of the environmental dependency syndrome, in which seeing an object implies that it should be used. Two forms of utilization behaviour are described:

~ *induced,* when an item is given to the patient or their attention is directed to it, *e.g.* handing them a pair of spectacles which they put on, followed by a second pair, which are put on over the first pair;
~ *incidental,* or spontaneous, when the patient uses an object in their environment without their attention being specifically directed towards it.

Another element of the environmental dependency syndrome which coexists with utilization behaviour is imitation behaviour *(e.g.* echolalia, echopraxia); primitive reflexes and hypermetamorphosis may also be observed.
Utilization behaviour is associated with lesions of the frontal lobe, affecting the inferior medial area bilaterally. It has also been reported following paramedian thalamic infarction.

- De Renzi E, Cavalleri F, Facchini S. Imitation and utilisation behaviour. *Journal of Neurology, Neurosurgery and Psychiatry* 1996; **61**: 396-400
- Lhermitte F, Pillon B, Serdaru M. Human autonomy and the frontal lobes. Part I: imitation and utilization behaviour: a neuropsychological study of 75 patients. *Annals ofNeurology* 1986; **19**: 326-34
- Shallice T, Burgess PW, Schon F, Baxter DM. The origins of utilization behaviour. *Brain* 1989;**112**: 1587-98
[Cross References: ECHOLALIA; ECHOPRAXIA; FRONTAL LOBE SYNDROMES; HYPERMETAMORPHOSIS; IMITATION BEHAVIOUR]

V

Valsalva Manoeuvre
The Valsalva manoeuvre is a simple test of autonomically-mediated cardiovascular reflexes, comprising forced expiration against resistance ("straining"), followed by release of the resistance and completion of expiration. The first phase produces impaired cardiac filling due to impaired venous return as a consequence of elevated intrathoracic pressure, with a fall in cardiac output and blood pressure, inducing peripheral vasoconstriction (sympathetic pathways) to maintain blood pressure. The second phase causes a transient overshoot in blood pressure as the restored cardiac output is ejected into a constricted circulation, followed by reflex slowing of heart rate.

In autonomic (sympathetic) dysfunction, reflex vasoconstriction, blood pressure overshoot and bradycardia do not occur. The latter may be conveniently assessed by measuring R-R intervals in a prolonged ECG recording, an R-R interval ratio between the straining and release phases of less than 1.1 suggesting impaired baroreceptor response.
[Cross References: ORTHOSTATIC HYPOTENSION]

Vegetative State
The vegetative state is a clinical syndrome in which cognitive function is lost, due to neocortical damage (hence no awareness, response, speech), whilst vegetative (autonomic, respiratory) function is preserved due to intact brainstem centres. Primitive postural and reflex limb movements may also be observed. The syndrome, also known as neocortical death and the apallic syndrome, may be seen after extensive ischaemic-hypoxic brain injury, for example following resuscitation after cardiac arrest, and needs to be distinguished from coma, akinetic mutism, and the locked-in syndrome. If persistent, the prognosis is poor, but occasional reports of very late recovery have appeared.

- Wade DT, Johnston C. The permanent vegetative state: practical guidelines on diagnosis and management. *BMJ* 1999; **319**: 841-4
[Cross References: AKINETIC MUTISM; COMA; LOCKED-IN SYNDROME]

Venous Pulsation
Venous pulsation is evident in the normal retina when observed with an ophthalmoscope, particularly at the margin of the disc. It is sometimes difficult to see, and may be more obvious in the recumbent position (because of higher pressure within the retinal veins). Venous pulsation is lost when intracranial pressure rises above venous pressure, hence this may be an early sign of impending papilloedema; however, venous pulsation may also be absent in pseudopapilloedema.
[Cross References: PAPILLOEDEMA; PSEUDOPAPILLOEDEMA]

Vernet's Syndrome
- see JUGULAR FORAMEN SYNDROME

Vertigo
Vertigo is an illusion of movement, a sense of rotation or of tilt, causing a feeling of imbalance or dysequilibrium. It is a subtype of "dizziness', to be distinguished from the light-headedness of general medical conditions (vasovagal attacks, presyncope, cardiac dysrhythmias). Vertigo is often triggered by head movement and there may be associated autonomic features (sweating, pallor, nausea, vomiting). Pathophysiologically, vertigo reflects an asymmetry of signalling anywhere in the central or peripheral vestibular pathways. Clinically it may be possible to draw a distinction between central and peripheral lesions: in the latter there may be concurrent hearing loss and tinnitus (reflecting vestibulocochlear (VIII) nerve involvement). Facial weakness (VII) and ipsilateral ataxia suggest a cerebellopontine angle lesion; diplopia, bulbar dysfunction and long tract signs are suggestive of a central pathology. Peripheral vertigo tends to compensate rapidly and completely with disappearance of nystagmus after a few days, whereas central lesions compensate slowly and nystagmus persists. The clinical pattern of vertigo may gives clues as to underlying diagnosis:

Vertigo	Peripheral	Central
Acute	Labyrinthitis	
Prolonged, spontaneous	Otomastoiditis; Vestibular neur(on)itis; Labyrinthine concussion; Isolated labyrinthine infarct; Vestibular nerve section; Drug-induced	Brainstem/cerebellum haemorrhage/infarct/ demyelination
Recurrent, episodic	Menière's disease (endolymphatic hydrops); Autoimmune inner ear disease (isolated, systemic); Perilymph fistula; Migraine (rare) Epilepsy (rare)	Vertebrobasilar ischaemia (with associated features)
Positional	Benign paroxysmal positional vertigo (BPPV)	4th ventricle lesions: multiple sclerosis Chiari malformation Brainstem/cerebellar tumours Spinocerebellar atrophy
Chronic	Vestibular decompensation/failure	Neurological disorder Psychogenic

All patients with vertigo should have a Hallpike manoeuvre performed during the examination.

Specific treatments are available for certain of these conditions. A brief course of a vestibular sedative (cinnarizine, Serc) is appropriate in the acute phase, but exercises to "rehabilitate" the semicircular canals should be begun as soon as possible in peripheral causes. In BPPV, most patients respond to the Epley manoeuvre to reposition the otoconia which are thought to cause the condition (canalolithiasis). Cawthorne-Cooksey exercises are helpful in vestibular decompensation or failure.

- Baloh RW. Vertigo. *Lancet* 1998**; 352**: 1841-6
- Luxon LM. Vertigo: new approaches to diagnosis and management. *British Journal of Hospital Medicine* 1996; **56**: 519-20, 537-41
[Cross References: ATAXIA; CALORIC TESTING; FACIAL PARESIS; HALLPIKE MANEOUVRE, HALLPIKE TEST; ILLUSION; NYSTAGMUS]

Vestibulo-ocular Reflexes

The vestibulo-ocular reflex (VOR) is a physiological mechanism which generates eye rotations that compensate for head movements, especially during locomotion, so stabilizing the retinal image on the fovea. VORs depend upon the integrity of the connections between the semicircular canals of the vestibular system (afferent limb of reflex arc) and oculomotor nuclei in the brainstem (efferent limb). Loss of vestibular function, as in acute bilateral vestibular failure, causes gaze instability due to loss of VORs, causing the symptom of oscillopsia (*q.v.*) when the head moves. As well as vestibular input, compensatory eye rotations may also be generated in response to visual information (pursuit-optokinetic eye movements) and neck proprioceptive information; anticipatory eye movements may also help stabilize the retinal image.

VORs are difficult to assess in conscious patients because of concurrent pursuit-optokinetic eye movements, and because rotation of the head through large angles in conscious patients leads to interruption of VORs by vestibular nystagmus in the opposite direction (optokinetic nystagmus). The head impulse test (*q.v.*) may be used to test VORs in conscious patients, for example those with vertigo in whom vestibular failure is suspected.

VORs are also useful in assessing whether ophthalmoplegia results from a supranuclear or infranuclear disorder, since in the former the restriction of eye movement may be overcome, at least in the early stages, by the intact VOR, *e.g.* the supranuclear gaze palsy in the vertical plane in Steele-Richardson-Olszewski syndrome.

In unconscious patients, VORs may be tested by rotating the head and looking for contraversive conjugate eye movements (oculocephalic responses, doll's head eye movements); VORs are lost in brainstem death.

Another important element of VOR assessment is VOR suppression, tested by asking the patient to fixate on their thumbs with arms held outstretched whilst rotating at the trunk or sitting in a swivel chair. VOR suppression can also be assessed during caloric testing: when the nystagmus ceases with fixation, removal of the fixation point (*e.g.* with Frenzel's glasses) will lead to recurrence of nystagmus in normals but not in those with reduced or absent VOR suppression. VOR suppression is impaired (presence of nystagmus even with slow head movements) in cerebellar and brainstem disease.

- Leigh RJ, Brandt T. A reevaluation of the vestibulo-ocular reflex: new ideas of its purpose, properties, neural substrate, and disorders. *Neurology* 1993; **43**: 1288-95
- Rudge P, Bronstein AM. Investigations of disorders of balance. In: Hughes RAC (ed.). *Neurological Investigations.* London: BMJ Publishing 1997: 283-314
[Cross References: CALORIC TESTING; COMA; HALLPIKE MANEOUVRE, HALLPIKE TEST; HEAD IMPULSE TEST; OCULAR TILT REACTION; OCULOCEPHALIC RESPONSE; OPTOKINETIC NYSTAGMUS (OKN), OPTOKINETIC RESPONSE; OSCILLOPSIA; SUPRANUCLEAR GAZE PALSY; VERTIGO]

Vibration

Vibratory sensibility represents a temporal modulation of tactile sense; the elevation of vibration to a "sensory modality" is not justified. Vibratory sensibility is easily tested using a tuning fork (128 Hz). This assesses the integrity of rapidly adapting mechanoreceptors (Pacinian corpuscles) and their peripheral and central connections; the former consist of large afferent fibres, the latter of ascending projections in both the dorsal and lateral columns. The notion of "posterior column signs" (impaired vibration and proprioception) is not substantiated, although common in clinical neurological parlance (and textbooks). Instances of dissociation of vibratory sensibility and proprioception are well recognised, for instance the former is usually more impaired with intramedullary myelopathies.

- Calne DB, Pallis CA. Vibratory sense: a critical review. *Brain* 1966; **89:** 723-46
[Cross References: MYELOPATHY; PROPRIOCEPTION; TWO-POINT DISCRIMINATION]

Visual Agnosia

Visual agnosia is a disorder of visual object recognition. According to Lissauer (1890), this may be divided into two types:

~ *apperceptive visual agnosia*: a defect of higher order visual perception leading to impaired shape recognition, manifested as difficulty copying shapes or matching shapes, despite preserved primary visual capacities, including visual acuity and fields (adequate to achieve recognition), brightness discrimination, colour vision and motion perception (indeed motion may facilitate shape perception; see Riddoch's phenomenon). Reading is performed with great difficulty, with a "slavish" tracing of letters which is easily derailed by any irrelevant lines; such patients may appear blind.
~ *associative visual agnosia*: an impairment of visual object recognition thought not to be due to a perceptual deficit, since copying shapes of unrecognised objects is good. The scope of this impairment may vary, some patients being limited to a failure to recognise faces (prosopagnosia) or visually presented words (pure alexia, pure word blindness).

Visually agnosic patients can recognise objects presented to other sensory modalities. Clinically, apperceptive visual agnosia lies between cortical blindness and associative visual agnosia.

Apperceptive visual agnosia results from diffuse posterior brain damage; associative visual agnosia has been reported with lesions in a variety of locations, usually ventral

temporal and occipital regions, usually bilateral but occasionally unilateral. Pathological causes include cerebrovascular disease, tumour, advanced degenerative dementia, and carbon monoxide poisoning.

A related syndrome which has on occasion been labelled as apperceptive visual agnosia is simultanagnosia (*q.v.*), the inability to recognise more than one object at a time, particularly the dorsal variant. Associative visual agnosia has sometimes been confused with optic aphasia (*q.v.*).

- Farah MJ. *Visual agnosia: disorders of object recognition and what they tell us about normal vision.* Cambridge: MIT Press 1995

[Cross References: AGNOSIA; ALEXIA; CORTICAL BLINDNESS; OPTIC APHASIA; PROSOPAGNOSIA; RIDDOCH'S PHENOMENON; SIMULTANAGNOSIA]

Visual Disorientation

Visual disorientation refers to the inability to perceive more than a fragment of the visual field at any one time; it is sometimes characterized as a shifting fragment or island of clear vision. Visual disorientation is secondary to, and an inevitable consequence of, the attentional disorder of dorsal simultanagnosia (*q.v.*), in which the inability to attend two separate loci leads to impaired localization.

Visual disorientation may be demonstrated by sitting directly opposite the patient and asking them, whilst looking at the bridge of the examiner's nose, to reach for the examiner's hand held up in the peripheral field of vision. Once contact is made with the hand, the examiner holds up the other hand in a different part of the field of vision. Individuals with visual disorientation will find it hard to see the hand and will grope for it, sometimes mistakenly grasping the examiner's clothing ("tie sign") or face.

Visual disorientation may be a feature of Alzheimer's disease; indeed, sometimes it may be the presenting feature, but there are usually signs of more generalized cognitive problems (*e.g.* impairment of episodic memory).

- Farah MJ. *Visual agnosia: disorders of object recognition and what they tell us about normal vision.* Cambridge: MIT Press 1995

[Cross References: SIMULTANAGNOSIA; VISUAL AGNOSIA]

Visual Extinction

Visual extinction is the failure to respond to a novel or meaningful visual stimulus on one side when a homologous stimulus is given simultaneously to the contralateral side (*i.e.* double simultaneous stimulation), despite the ability to perceive each stimulus when presented singly.

[Cross References: EXTINCTION; NEGLECT]

Vocal Tremor, Voice Tremor

Vocal or voice tremor is a shaking, quivering, or quavering of the voice. It may be heard in:

- Essential tremor;
- Cerebellar disorders;

- Spasmodic dysphonia/laryngeal dystonia;
- Parkinson's disease;
- Motor neurone disease.

The pathophysiology is uncertain but may relate to rhythmic contractions of the cricothyroid and rectus abdominis muscles.
[Cross References: DYSPHONIA; TREMOR]

Von Graefe's Sign
Von Graefe's sign is the retarded descent of the upper eyelid during movement of the eye from the primary position to downgaze, the lid "follows" the eye. This may be termed "lid lag", although some authorities reserve this term for a static situation in which the lid is higher than the globe on downgaze. Von Graefe's sign is seen in thyroid ophthalmopathy.
[Cross References: PSEUDO-VON GRAEFE'S SIGN]

Vorbereiden
- see GANSER PHENOMENON

W

"Waiter's Tip" Posture
- see PLEXOPATHY; RADICULOPATHY

Wallenberg's Syndrome
- see LATERAL MEDULLARY SYNDROME; OCULAR TILT REACTION

Wartenberg's Swing Test
Wartenberg's swing test is used to assess limb and trunk rigidity (*cf.* Wartenberg's pendulum test, used to measure spasticity, *q.v.*). With the patient standing, the examiner holds the shoulders and gently shakes backwards and forwards, the two sides out of phase. Normally the passive arm swing induced by this movement will be out of phase with the trunk movements, but in rigidity the limbs and trunk tend to move *en bloc*. Passive swinging of the wrist or elbow joint may also be performed to assess rigidity.
[Cross References: PARKINSONISM; RIGIDITY; SPASTICITY]

Wasting
Wasting refers to a thinning of the musculature, also known as atrophy or, if of neurogenic origin, amyotrophy.

Wasting may be a consequence of disorders of:

- muscle (myopathies, dystrophies);
- peripheral nerve (more so in axonal than demyelinating peripheral neuropathies);
- anterior horn cells (*e.g.* motor neurone disease).

Wasting may occur in chronic upper motor neurone syndromes (*e.g.* chronic hemiplegia) but is not as evident as in lower motor neurone syndromes where wasting may appear acutely (over a few weeks).

Wasting may also be seen in general medical disorders associated with a profound catabolic state, *e.g.* cancer cachexia, uncontrolled heart failure, liver cirrhosis, renal failure.
[Cross References: AMYOTROPHY; ATROPHY; LOWER MOTOR NEURONE SYNDROME; UPPER MOTOR NEURONE SYNDROME]

Weakness
Weakness is an objective loss of muscle strength. This is conveniently quantified or rated using the MRC grading system:

5 = normal power
4 = active movement against gravity and resistance
3 = active movement against gravity
2 = active movement with gravity eliminated

1 = flicker or trace of contraction
0 = no contraction (paralysis).

However, this is not a linear scale; grade 4 often becomes subdivided into 4-, 4, and 4+ (or even 5-) according to the increasing degree of resistance which the examiner must apply to overcome activity. It is also important to assess what effort the patient is making to comply with the testing; "apparent weakness" may be shorthand for lack of patient effort, sudden "giving way" of muscle contraction being a possible indicator of this. Non-uniform resistance may also be due to pain (algesic pseudo-paresis). Testing records only the best forced maximal contraction, and should not develop into an unseemly trial of strength between patient and examiner. Accepting all these difficulties, it should be acknowledged that the grading of weakness, like all clinical observations, is subject to some degree of observer bias.

The pattern of muscle weakness may suggest its anatomical origin. So-called "pyramidal weakness" (*i.e.* affecting upper limb extensors more than flexors, and lower limb flexors more than extensors), suggests an upper motor neurone lesion (corticospinal pathways). However, there is no evidence that pure lesions of the pyramidal tracts produce this picture: pyramidotomy in the monkey results in a deficit in fine finger movements, but without weakness. Moreover, a similar pattern of weakness may be observed in lower motor neurone disorders such as Guillain-Barré syndrome. Coexistent wasting suggests muscle weakness is of lower motor neurone origin, especially if acute, although wasting may occur in long-standing upper motor neurone lesions. Weakness with minimal or no muscle wasting may be non-organic, but may be seen in conditions such as multifocal motor neuropathy with conduction block.

- *Aids to the Examination of the Peripheral Nervous System.* London: HMSO 1976
[Cross References: HYPERREFLEXIA; LOWER MOTOR NEURONE SYNDROME; UPPER MOTOR NEURONE SYNDROME; WASTING]

Weber's Test

Weber's test is one of the tuning fork tests, which may be used to confirm a conductive component in unilateral or asymmetric hearing loss. The vibrating tuning fork is put on the middle of the forehead and the patient asked in which ear it is heard; this depends entirely upon bone conduction (BC). Hence the sound localises to the side of a conductive hearing loss (where bone conduction is greater than air conduction, BC > AC), and away from the side of a sensorineural loss.
[Cross References: RINNE'S TEST]

Wernicke's Aphasia

Wernicke's aphasia is the classical "receptive aphasia", in distinction to the "expressive aphasia" of Broca, although this classification is problematic since there are concurrent "expressive" problems in Wernicke'a aphasia. Considering each of the features suggested for the clinical classification of aphasias (see Aphasia), Wernicke's aphasia is characterized by:

- FLUENCY: fluent speech with phonemic and semantic paraphasias and paragrammatism; "empty speech" with few verbs and nouns; prosody usually

preserved; at worst, flowing speech (logorrhoea) devoid of semantic meaning (jargon aphasia, semantic aphasia); automatic speech is often better preserved than spontaneous, *e.g.* counting, days of week, overlearned phrases ("I'm fine");
- COMPREHENSION: impaired auditory comprehension (*sine qua non;* "word deafness");
- REPETITION: impaired;
- NAMING: severely impaired (anomia) and not aided by cueing (*cf.* Broca's aphasia);
- READING: usually impaired with numerous paralexic errors;
- WRITING: similarly affected.

There may be associated anxiety, with or without agitation and paranoia.

Neuroanatomically, Wernicke's aphasia is associated with pathology (usually infarction) in the left (dominant) posterior superior temporal lobe (Wernicke's area, posterior two-thirds of the superior temporal gyrus, auditory association cortex; Brodmann area 22). Damage may be more extensive, involving Brodmann areas 37, 39 and 40. A correlation exists between the size of the lesion and the extent of the aphasia. There may be concurrent auditory agnosia. A similar picture may be seen with infarcts of the head of the left caudate nucleus and left thalamic nuclei.

[Cross References: AGNOSIA; AGRAPHIA; ALEXIA; ANOMIA; APHASIA; BROCA'S APHASIA; JARGON APHASIA; LOGORRHOEA; PARAPHASIA; TRANSCORTICAL APHASIA]

"Wheelchair Sign"

The so-called "wheelchair sign" has been applied to parkinsonian patients who take to using a wheelchair early in the course of their disease. Such patients almost certainly do not have idiopathic Parkinson's disease; early falls are a particular feature in Steele-Richardson-Olszewski syndrome (progressive supranuclear palsy).

[Cross References: PARKINSONISM; "ROCKET SIGN"]

Winging of the Scapula

Winging of the scapula is a failure to hold the medial border of the scapula against the rib cage when pushing forward with the hands. It is most easily observed by asking the patient to push or press against a wall or the examiner's hand whilst observing the scapula which lifts away from the chest wall.

Winging of the scapula is a consequence of weakness of the serratus anterior muscle, usually due to a neuropathy of the long thoracic nerve of Bell, but sometimes as a consequence of brachial plexus injury or root injury. Weakness of trapezius may also cause winging of the upper part of the scapula on abduction of the arm.

Witzelsucht

Witzelsucht refers to excessive and inappropriate facetiousness or jocularity, a term coined in the 1890's for one of the personality changes observed following frontal (especially orbitofrontal) lobe injury.

[Cross References: EMOTIONAL LABILITY; FRONTAL LOBE SYNDROMES]

Woltman's Sign

Woltman's sign denotes slow-relaxing, or "hung up", tendon reflexes. These are most commonly seen in the context of untreated hypothyroidism, but have also been recorded in other situations, including treatment with β-blockers, diabetes mellitus, and complete heart block. The phenomenon is sometimes labelled pseudomyotonia because of its superficial resemblance to the slow muscle relaxation of myotonia, but electrophysiological testing does not show myotonic discharges.

The mechanisms underlying Woltman's sign are uncertain: changes in basal metabolic rate and in muscle fibre types (selective loss of fast twitch fibres) have been suggested.

- Larner AJ. Normalisation of slow-relaxing tendon reflexes (Woltman's sign) after cardiac pacing for complete heart block. *British Journal of Clinical Practice* 1995; **49**: 331-2

[Cross References: MYOTONIA; PSEUDOMYOTONIA]

Wrist Drop

Wrist drop describes a hand hanging in flexion due to weakness of wrist extension. This results from radial nerve palsy, either in the axilla or spiral groove of the humerus ("Saturday night palsy"). Distal lesions affecting branches of the posterior interosseous branch of the radial nerve may produce more circumscribed deformity, such as weak extension of metacarpophalangeal joints ("drop finger", "drop thumb").

Writer's Cramp

Writer's cramp, or graphospasm, is a focal dystonia of the hand in which dystonic posturing is induced specifically by writing: this is the commonest task-specific dystonia. The involuntary movements may eventually make it impossible to write with the dominant hand; learning to write with the opposite hand may only be a partial solution, since it too may become affected. Muscle fatigue may make writing more legible. Writer's cramp is much commoner than primary writing tremor as a cause of difficulty writing.

Botulinum toxin injections may be of benefit if relatively few muscles are affected. There may be an associated carpal tunnel syndrome.

- Sheehy MP, Marsden CD. Writer's cramp – a focal dystonia. *Brain* 1982; **105**: 461-80

[Cross References: DYSTONIA; FATIGUE; TREMOR]

Wry Neck

- see TORTICOLLIS

X

Xanthopsia
Xanthopsia is a visual disturbance characterized by excessive perception of yellow colours (literally "yellow vision"). It may be caused by a number of drugs including digoxin (especially if levels are toxic), thiazides (especially chlorothiazide), sulphomaides, and barbiturates. The mechanism is uncertain.

It has been suggested that Vincent van Gogh (1853-1890) suffered from xanthopsia as a consequence of digitalis toxicity, accounting for the bright yellows in many of his later canvases.

Xerophthalmia, Xerostomia
Xerophthalmia, dryness of the eyes, and xerostomia, dryness of the mouth, due to impaired secretion from the lacrimal glands and the salivary glands respectively, often occur together. This may be a feature of autonomic dysfunction, for example in Lambert Eaton myasthenic syndrome, or due to autoimmune disorders such as Sjögren's syndrome.
[Cross References: ORTHOSTATIC HYPOTENSION]

Y

Yawning
Yawning is an arousal reflex generated in the brainstem reticular formation which is thought to counteract brain hypoxia; it may precede vasovagal syncope. Excessive or pathological yawning (chasm) is compulsive, repetitive yawning not triggered by physiological stimuli such as fatigue or boredom. Known causes of chasm include:

- encephalitis;
- seizures;
- multiple sclerosis;
- tumours of the **4th** ventricle;
- electroconvulsive therapy;
- drugs (valproate, imipramine);
- neuroleptic withdrawal.

It may represent a disturbance of dopaminergic transmission. Levodopa may help.

- Leonhardt M, Abele M, Klockgether T, Dichgans J, Weller M. Pathological yawning (chasm) associated with periodic leg movements in sleep: cure by levodopa. *Journal of Neurology* 1999; **246:** 621-2
[Cross References: SIGHING]

Yo-yo-ing
Yo-yo-ing is a form of dyskinesia experienced by patients with idiopathic Parkinson's disease who have been treated for several years with levodopa preparations, in which there are sudden and unpredictable swings between hypokinesia/akinesia ("off "state; freezing) and severe hyperkinesia ("on" state), sometimes known as the "on-off phenomenon". Yo-yo-ing is difficult to treat: approaches include dose fractionation, improved drug absorption, or use of dopaminergic agonists with concurrent reduction in levodopa dosage.
[Cross References: AKINESIA; DYSKINESIA; HYPOKINESIA]

Z

Zooagnosia
The term zooagnosia has been used to describe a difficulty in recognising animal faces. This may be observed as a component of prosopagnosia. In one case, this deficit seemed to persist despite improvement in human face recognition, suggesting the possibility of separate systems for animal and human face recognition; however, the evidence is not compelling.

- Assal G, Favre C, Anderes J. Nonrecogntion of familiar animals by a farmer: zooagnosia or prosopagnosia for animals. *Revue Neurologique (Paris)* 1984; **140:** 580-4
[Cross References: PROSOPAGNOSIA]