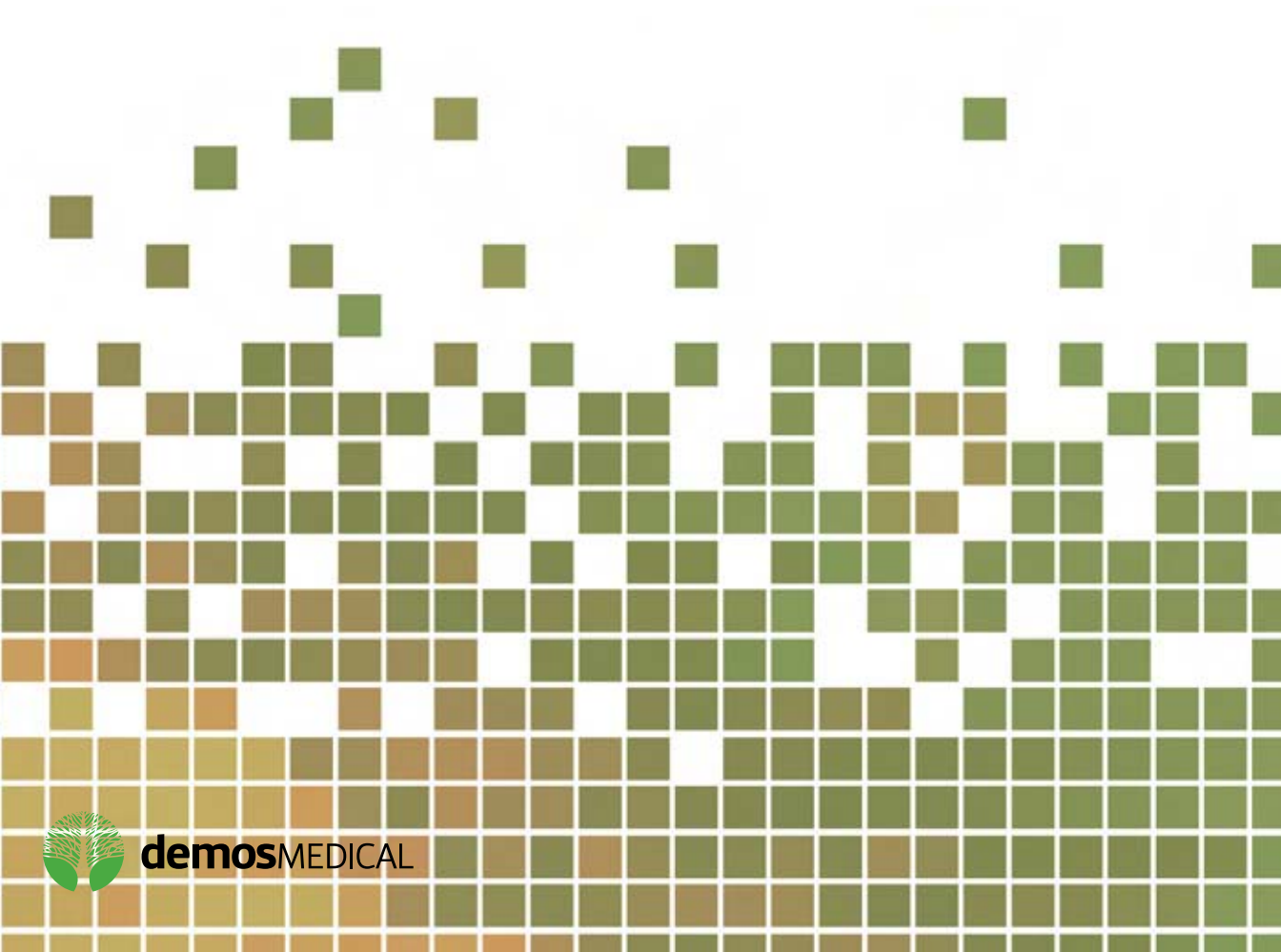
The Dystonia Patient

Michael S. Okun

A Guide to Practical Management



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A Guide to Practical Management

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For Tyler and Samantha, and for Tyler's Hope for a Dystonia Cure

While we search for the cure, may we strive to deliver and inspire the most empathetic and comprehensive care possible for all the dystonia sufferers in the world.

All royalties from this book will be donated to

Tyler's Hope for a Dystonia Cure
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Preface

Although it is unknown how many people in the world actually suffer from dystonia, the numbers of those diagnosed with the syndrome seem to increase each time the issue is reexamined. When I was in training, my mentors used to say Parkinson's disease is ten times more common than dystonia. Each year that has passed, however, there has been a reduction in this ratio among the experts, with some even quoting 3–4:1. Although many people have never heard of dystonia, it is common. As a practicing movement disorders expert, it has always been troubling to me that as we identify more dystonia cases, we have as a field offered little guidance for the best multidisciplinary and interdisciplinary techniques, and we have not established the best care paradigms for these patients. Challenged by Rick Staab, the CEO and founder of Tyler's Hope for a Dystonia Cure and a dystonia dad, we have responded by writing *The Dystonia Patient: A Guide to Practical Management.*

The book is designed to be comprehensive and includes chapters covering the multidisciplinary and interdisciplinary treatment of the dystonia patient. Each chapter is accompanied by practical pearls that physicians and healthcare providers should keep in mind when attempting to provide the best possible comprehensive care for patients. The book also features lists of resources and websites for patients, families, and medical teams. The content spans children to adults, as well as medical/behavioral and surgical approaches to the dystonia patient. Our underlying motive in writing the book was to drive the construction of multidisciplinary and interdisciplinary care teams and to address the need for best care in dystonia patients. We also hope that as care teams form, they will enhance the treatment of the dystonia family.

The chapters are divided into main topic areas that address the comprehensive care for the dystonia patient. Morishita and colleagues review the medical as well as surgical approaches to dystonia treatment. Dr. Rodriguez follows this chapter by reviewing botulinum toxin therapy for dystonia. Romrell, Greenhut, and colleagues then explore the uses of nurse practitioners/physician

assistants as well as social workers for the dystonia interdisciplinary team. Speech and swallowing as well as physical and occupational therapy are then addressed by Plowman-Prine, Myers, and Gardner-Smith in what includes a comprehensive review of current approaches to these common problem areas. Reckess, Ward, and colleagues then discuss the role of psychology and psychiatry in the treatment of the dystonia patient. Finally, Zeilman reviews the complex topic of deep brain stimulation programming in dystonia.

We hope that by providing this resource we can in some small way catalyze the formation of multidisciplinary and interdisciplinary teams for the care of the dystonia patient. This book, written by the dystonia team at the University of Florida Movement Disorders Center, hopefully will capture the imagination of others and inspire professionals to develop teams aimed at the practical and comprehensive care of the dystonia patient.

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The Dystonia Patient

1

Medical and Surgical Approaches to Dystonia

Takashi Morishita
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Dystonia is a neurologic syndrome characterized by involuntary muscle contractions of opposing muscles that may result in twisting and repetitive movements or abnormal postures. Partly because of its rich expression and its variable course, dystonia is frequently not recognized or is misdiagnosed. The prevalence of generalized primary torsion dystonia in Rochester, Minnesota, was reported to be 3.4 per 100,000 population, and focal dystonia incidence was reported as 30 per 100,000 population (1). Although recent studies have revealed the genetics of various forms of dystonia and its pathophysiology, the assessment of various therapeutic interventions has been problematic because of the heterogeneous nature of the disorder (2). In addition, patients have not consistently responded to one type of therapy; therefore, multiple strategies including oral medications, chemodenervaton, and surgical treatments have been required. In recent years, much progress has been made in surgical treatment, especially deep brain stimulation (DBS). This chapter will discuss the clinical features and review the literature concerning medical and surgical approaches to dystonia. In the section on surgical procedures, we will focus on DBS and provide pearls for clinicians. A separate chapter in the text will cover botulinum toxin treatment of dystonia.

Phenomenology

The main features of dystonia may be summarized as follows:

1. Muscle contractions are sustained at the peak of movement for a relatively long duration, unlike chorea or myoclonus, in which the involuntary movements are brief.
2. Both agonist and antagonist muscles of a body part simultaneously contract, which may result in twisting or abnormal movements of the affected body part.
3. The same muscle groups are generally involved, unlike chorea, in which the involuntary movements may be random and involve different muscle groups.

Besides these main features of dystonia, several characteristics are unique to this disease. Dystonic symptoms are generally aggravated by voluntary movements (action dystonia) as well as by fatigue and stress. Abnormal dystonic movements that appear only during certain actions may be termed "task-specific dystonia"—an example is writer's cramp. Primary dystonia usually begins with a single part of the body (focal dystonia) after which it may gradually generalize, with the spread most often to contiguous body parts. Patients with younger-onset focal dystonia tend to progress to generalized forms of dystonia (3). As the dystonic syndrome evolves, even nonspecific voluntary action can bring out dystonia, and eventually actions in other parts of the body can induce dystonic movements of the primarily affected body part—this is termed "overflow dystonia." In addition, children and adolescents with primary or secondary dystonia rarely develop a sudden and marked increase in the severity of dystonia, and the phenomenon in these cases may be termed "dystonic storm."

Dystonia can present with tremor (dystonic tremor) or myoclonus (dystonia-myoclonus). Dystonic tremor associated with cervical dystonia can usually be distinguished from essential tremor as the dystonic tremor may be less regular or rhythmic than is seen in essential tremor. Dystonic tremor is usually associated with a head tilt and chin deviation and does not require maintenance of the posture for activation of muscles.

One of the most characteristic features of dystonia is that the abnormal movement may be ameliorated by "sensory tricks" (geste antagoniste)—lightly touching the affected body part can often reduce muscle contractions/dystonic postures. Besides sensory tricks, certain voluntary activities can relieve dystonia, and these are sometimes termed "paradoxical dystonia" (4). This phenomenon

is most often seen in patients with facial dystonia when they are speaking or chewing, and it may alleviate their blepharospasm. Other alleviating factors that have been reported include sleep, hypnosis, and relaxation.

Interestingly, pain is not very common in dystonia with the exception of cervical dystonia, where in up to 75% of patients pain can be seen (5). It is unknown why cervical dystonia has pain associated, but factors might include larger muscle mass or larger numbers of pain receptors (5). Another study has suggested that pain may derive from central as well as myofascial origin, especially since there is a gap between the severity and/or duration of motor signs and the pain (6).

Classification

Dystonia may be classified by 1) age at onset, 2) distribution of affected body regions, 3) etiology, and 4) genetics. Regarding the age at onset, early-onset dystonia may be classified as a group of dystonias with onset before the age of 26 years. Classification based on age is clinically useful in predicting the outcome of dystonia, because the earlier the age at onset, the more likely symptoms will be severe. Based on the distribution, dystonia may be classified into four categories; focal, segmental, multifocal, and generalized dystonia. "Focal," "segmental," "multifocal," and "generalized" dystonia can be defined as affecting a single body part, one or more contiguous body parts, two or more noncontiguous body parts, or the entire body. "Hemidystonia" is sometimes included as multifocal dystonia, but as a syndrome it is characteristic and different (Table 1.1).

TABLE 1.1 Classification of the Dystonias by Age and Distribution

AGE OF ONSET:	
Early-onset dystonia: <26 yr	
Late-onset dystonia: >26 yr	
DISTRIBUTION	DEFINITION
Focal	Single body region (e.g., blepharospasm, oromandibular dystonia, spasmodic dysphonia, cervical dystonia, task-specific dystonias)
Segmental	Contiguous body region (Meige's syndrome, camptocormia)
Multifocal	Two or more noncontiguous body parts
Generalized	Entire body

TABLE 1.2 Causes of the Dystonia by Etiology

PRIMARY DYSTONIA	
Early-onset limb dystonia (DYT1)	
Mixed dystonia (DYT6, DYT13)	
Late-onset craniocervical dystonia (DYT7)	
SECONDARY DYSTONIA	
Dystonia-plus	Dopa-responsive dystonia GTP cyclohydrolase-1 mutations (DYT5a) Tyrosine hydroxylase mutations (DYT5b) Other biopterin deficiencies Dopamine agonist-responsive dystonia (aromatic acid decarboxylase deficiency) Myoclonus-dystonia (DYT11) Rapid-onset dystonia parkinsonism (DYT12)
Heredogenerative dystonias	Autosomal dominant Huntington's disease Machado-Joseph's disease (SCA3) Dentatorubralpallidoluysian atrophy (DRPLA) Autosomal recessive Wilson's disease GM1 and GM2 gangliosidosis Metachromatic leukodystrophy Homocystinuria Hartnup disease Glutaric acidemia Methylmalonic aciduria Hallervorden-Spatz disease Dystonic lipidosis Ceroid-lipofuscinosis Ataxia-telangiectasia Neuroacanthocytosis Intraneuronal inclusion disease Juvenile parkinsonism (Parkin) X-Linked recessive Lubag (X-linked dystonia-parkinsonism or DYT3) Lesch-Nyhan syndrome Mitochondrial MERRF MELAS Leber's disease
Perinatal cerebral injury with kernicterus	Athetoid cerebral palsy
Infection	Viral encephalitis Encephalitis lethargica Reye's syndrome Subacute sclerosing panencephalitis Creutzfeldt-Jakob disease HIV infection

TABLE 1.2 *(continued)*

SECONDARY DYSTONIA	
Drugs	Levodopa and dopamine agonists, dopamine receptor–blocking agents, fenfluramine, anticonvulsants, flecainide, ergots, some calcium channel blockers
Toxins	Manganese, carbon monoxide, carbon disulfide cyanide, methanol, disulfiram, 3-nitroproprionic acid, wasp sting toxin
Metabolic	Hypoparathyroidism
Brain/brainstem lesions	Paraneoplastic brainstem encephalitis, primary antiphospholipid syndrome, ischemic injury central pontine myelinolysis, multiple sclerosis tumors, arteriovenous malformation (AVM), trauma, surgery (thalamotomy)
Spinal cord lesions	Syringomyelia, trauma, surgery
Peripheral lesions	Lumbar stenosis, trauma, electrical injury, complex regional pain syndrome (CRPS)
Unknown etiology	Parkinson's disease Corticobasal degeneration Multiple system atrophy Progressive supranuclear palsy

Historically, dystonia is largely classified into two groups: primary (idiopathic) and secondary (symptomatic) (Table 1.2). While the term "secondary" seems to be straightforward, the definition of "primary" dystonia has been changing as the genetics of various forms of dystonia have been revealed. In this context, the term "primary torsion dystonia" (PTD) has been proposed to replace primary dystonia, and the following three clinical criteria should be employed: 1) dystonia as the sole abnormality directly attributable to the condition, 2) no laboratory or imaging abnormalities to suggest an acquired or degenerative cause for dystonia and no dramatic response to levodopa to suggest dopa-responsive dystonia, and 3) historical information failing to implicate a known acquired or environmental cause of dystonia (7).

Fifteen subtypes of dystonia have been classified based on the genetics to date, and they have been designated dystonia types (DYT) 1–15 by the Human Genome Organization/Genome Database (HUGO/GDB) (Table 1.3) (2,8–23). However, this genetic classification is problematic since it includes various etiologic types of dystonia. Six of these 15 dystonias are primary forms (DYT1, 2, 4, 6, 7, and 13), and the others are mixed with secondary-like dystonias including dystonia-plus syndromes. While a chromosomal location has been identified for 13 types of dystonia, only 5 mutated genes have been identified (DYT-1, 5a, 5b, 11, and 12). The other 2 subtypes (DYT2 and 4) have been assigned on the basis of clinical description alone.

TABLE 1.3 Genetic Classification of the Dystonias

TYPE	CHROMOSOME/GENE	INHERITANCE	AGE AT ONSET	FEATURES
DYT1	9q34/torsin A; deletion of one pair of GAG triplets	AD; penetrance rate 30–40%	Mean age 12.5, early onset (<26 years old testing is useful)	1/2000 in the Ashkenazi Jewish population; commercial testing is available, limbs affected first; pure dystonia, MRI normal
DYT2		AR	Early onset	Described in Spanish gypsies
DYT3	Xq13.1	XR	Mean 35 yr; adult-onset	"Lubag," Filipino males, dystonia-parkinsonism
DYT4		AD	13–37 yr	Single Australian family, "whispering dysphonia" family
DYT5a, DRD, Segawa disease	14q22.1/GTP cyclohydrolase-1	AD	Childhood	Usually limb-onset, dramatic response to levodopa
DYT5b, DRD	11p/TH	AR	Infant	Infantile parkinsonism
DYT6	8p21-q22	AD	Mean 19 yr	Mixed type; in the Mennonite/Amish populations; site of onset in arm/cranial > leg/neck; usually remains as upper body
DYT7	18p	AD	28–70 yr	Familial torticollis in a northwestern German family; occasional arm involvement
DYT8, paroxysmal dyskinesias	2q33-q35/ Myofibril-logenesis regulator 1	AD	Variable, early childfood, adolescence	Paroxysmal nonkinesogenic dyskinesia
DYT9	1p21	AD	2–15 yr	Paroxysmal dyskinesia with spasticity
DYT10, PKC, PKD	16p11.2-q12.1	AD	6–16 yr	Paroxysmal kinesogenic dyskinesia
DYT11	7q21-q23/ epsilon-sargoglycan	AD	Variable	Myoclonus-dystonia syndrome; alcohol responsive
DYT12	19q/ATP1A3	AD	Childhood, adolescent	Rapid-onset dystonia parkinsonism
DYT13	1p36.13-p36.32	AD	5 yr to adults	Single Italian family with cervical dystonia

TABLE 1.3 *(continued)*

TYPE	CHROMOSOME/GENE	INHERITANCE	AGE AT ONSET	FEATURES
DYT14	14q14	AD	Childhood	Dopamine-responsive dystonia
DYT15	18p11	AD	Childhood to adlescent	Myoclonus-dystonia syndrome, alcohol responsive

AD, autosomal dominant; AR, autosomal recessive; DRD, dopa-responsive dystonia; TH, tyrosine hydroxilase; XR, X-linked recessive.

Oral Medication

Levodopa has been recognized as an effective treatment option since the discovery of dopa-responsive dystonia (DRD) (24). For generalized dystonia beginning in childhood or adolescence, a trial of levodopa is helpful to rule out DRD, especially if the patient is still young (25). Only small doses of levodopa are required to improve dystonic symptoms in patients with DRD. If levodopa is given with carbidopa, a response can frequently occur with a dose of less than 300 mg/day (26).A therapeutic trial should last at least one month, with a maximum dose of up to 1000 mg/day. If this treatment is successful, it should be maintained at the lowest possible dose (27). The most frequently reported side effect is nausea. Sinemet must not be abruptly discontinued, as this may lead to a life-threatening condition called neuroleptic malignant syndrome.

If levodopa is ineffective, anticholinergics should be tried as the next step in therapy (27). Trihexyphenidyl or benztropine are the most commonly used anticholinergics for dystonia, but others such as ethopropazine hydrochloride have been used (2). One prospective double-blind crossover study of 31 patients with torsion dystonia found that 22 (71%) had a clinically significant response to trihexyphenidyl, and 42% continued to show a considerable or dramatic benefit even after a mean follow-up of 2.4 years (28). Trihexyphenidyl in high dosages (up to 120 mg/day) is effective for the treatment of segmental and generalized dystonia in young patients (29). Surprisingly, children seem to tolerate higher doses when compared to adults. One study reported that a minimum of 40 mg/day of trihexyphenidyl was required for a clinical response (30). Trihexyphenidyl is started with a dose of 1–2 mg/day and gradually titrated up to the maximum tolerated dose through 4–6 weeks of treatment. Patients with a short duration of dystonia—especially within the first 5 years—tend to have the best response (31). Side effects may include memory loss, dry mouth, confusion,

hallucinations, exacerbation of acute angle glaucoma, and sedation. However, if increased very slowly, young patients may be able to tolerate high doses (32).

If anticholinergics are not tolerated or not helpful, baclofen, a γ-aminobutyric acid (GABA) agonist, should be considered (and can be added to the anticholinergic) (2,26). Although baclofen seems to be less effective than anticholinergics, it may result in a dramatic response in children. In a retrospective study, dramatic improvement in symptoms, especially in gait, was found in 29% of 31 children and adolescents with idiopathic dystonia using doses ranging from 40 to 180 mg/day (33). The side effects included sedation, weakness, and memory loss. Baclofen must not be abruptly discontinued because sudden withdrawal can cause seizures. Our own clinical experience has revealed benefit, but not to the magnitude reported in some of these studies.

Benzodiazepines can be a supplementary medication for patients on anticholinergics or baclofen, or they can be used in monotherapy. Clonazepam is usually the most commonly used benzodiazepine because of its long half-life. Jankovic and Ford reported that clonazepam was useful in some patients with blepharospasm (34). No controlled trial has evaluated this therapeutic approach. We have found it clinically useful to combine anticholinergics, baclofen, and benzodiazepines into a cocktail.

Tetrabenazine's mechanism of action is as an inhibitor of vesicular monoamine transporter 2 (VMAT2), and it leads to depletion of dopamine and other monoamines (norepinephrine and serotonin) in the central nervous system (35). Tetrabenazine has been used for Huntington's disease by several controlled trials, and recently in Tourette syndrome (36). Although there have not been controlled trials concerning its efficacy in dystonia, there are several positive reports (35,37,38). In spite of being a dopamine depleter, tetrabenazine rarely results in tardive dyskinesia. In fact, tetrabenazine has been used for the treatment of tardive dyskinesia (35). Patients are usually treated with 25–75 mg/day in divided doses. Tetrabenazine can be combined with anticholinergics and other medications (37,39). Side effects reported include drowsiness, parkinsonism, depression, and akathisia. Our own personal experience with tetrabenazine for dystonia has been disappointing, but it may prove efficacious in specific subtypes and in specific doses for certain forms of dystonia.

Anecdotal treatment with other oral medications including tizanidine and dopamine antagonists has been reported (40,41). In general, dopamine antagonists are not recommended because of acute and tardive side effects, although in select cases of secondary dystonia they may be effective (particularly the atypical class) (26) (Table 1.4).

TABLE 1.4 Common Medications Used for the Symptomatic Treatment of Dystonia

MEDICATION	TYPICAL STARTING DOSE (MG/DAY)	TYPICAL THERAPEUTIC DOSE (MG/DAY)	COMMENTS
Carbidopa/levodopa	25/100	Up to 800	To be given 3 times per day; always try levodopa first, especially if young, to rule out DRD—requires only low dose
Trihexyphenidyl	1–2	Up to 120	In divided doses; if increased very slowly, young patients are able to tolerate high doses
Benztropine	0.5–1	Up to 8	Watch for anticholinergic side effects
Baclofen	5–10	Up to 120	GABA agonist; do not abruptly discontinue (risk of seizures)
Clonazepam	0.5–1	Up to 5	
Tetrabenazine	25	Up to 75	
Tizanidine	2	24	Unlike baclofen, minimal risk of seizures with abrupt discontinuation

DRD, dopamine-responsive dystonia.

A common approach to the medical treatment of dystonia is to start with a trial of sinemet. Following this trial, sinemet can either be continued or slowly weaned. Next, a trial of slowly escalating doses of an anticholinergic is used. Once the highest tolerated dose is achieved, a muscle relaxant can be added. Following the addition of the muscle relaxant at maximally tolerated doses, a benzodiazepine may be added. Clinicians have empirically argued that a "cocktail" may have synergistic effects, although controlled trials have not been published on this approach.

Surgical Treatment Options for Dystonia

Surgical treatment options for dystonia include peripheral nerve procedures as well as central nervous system (CNS) approaches. Peripheral denervation surgeries including intradural sectioning of the cervical nerve roots (rhizotomy), extradural sectioning of the posterior primary divisions of the cervical nerve roots (ramisectomy), and myotomy, which may be indicated for patients with focal

and segmental dystonia, especially cervical dystonia. Because of the complex nature of cervical dystonia, those three procedures should be customized for each patient. Bertrand and Molina-Negro reported that 97 (87%) of 111 patients had total or marked relief of symptoms after selective peripheral denervation (42). However, the efficacy on activities of daily living has been controversial, as has been the methodology of the study. Krauss et al. demonstrated significant improvements not only in dystonia, but also in occupational work and activities of daily living after peripheral denervation (43). On the other hand, Ford et al. reported that surgical intervention did not return patients to their occupation (44). Adverse effects included paresthesias in the posterior cervical region, worsening of dysphagia, and paralysis of muscles. Most experts in the field have been gravitating away from peripheral surgeries to address dystonia and are now favoring brain lesion surgery or deep brain stimulation in cases where medication and botulinum toxin fall short.

Intraspinal procedures for dystonia are considered high risk and may result in spinal cord injuries such as tetraplegia, Brown-Sequard syndrome, or monoplegia. Peripheral denervation can also be applied for blepharospasm, but few data are available. Additionally, there have been reports concerning myectomy for blepharospasm (45,46). The procedure is currently not widely recommended because of the risk of exposure keratitis, facial droop, and postoperative swelling and scarring (27).

Intrathecal baclofen (ITB) is another surgical procedure that continuously delivers specific doses of baclofen into the intrathecal space. Since Narayan et al. first demonstrated the efficacy of ITB in 1991 in a report of an 18-year-old man with severe cervical and truncal dystonia (47), subsequent studies have shown the benefits of ITB (48–53). Walker et al. used a bolus injection of ITB as a screening test for implantation of the pump (53). They selected 14 patients following positive screening tests. Five of 14 patients had improvement on the Burke-Fahn-Marsden dystonia rating scale, and only 2 patients had a clear clinical benefit. On the other hand, Hou et al. reported 10 patients who underwent implantation of ITB pump and performed bolus injection of ITB in all cases (48). They found that patients without an initial response to bolus baclofen injection still benefited from continuous infusion. In addition, Hou et al. also found that ITB was more effective for generalized or segmental dystonia, especially in patients with spastic dystonia involving the lower body and trunk. The therapeutic response to ITB has been reported to wane over time, but Albright et al. demonstrated that 92% of 72 participants implanted with a

baclofen pump retained their responses to ITB during a median follow-up of 29 months (52). Surgical complications included cerebrospinal fluid (CSF) leaks, infections, and catheter problems, and the rate of these issues was reported to be as high as 20–38% (48,50–53).

CNS surgical procedures have been in development at least since Spiegel and Wycis introduced stereotactic neurosurgical techniques for the treatment of movement disorders (54). CNS surgery has been applied to dystonia based at least partially on the finding in the 1950s that ablative surgery improved dystonia in Parkinson's disease (PD) (55). In the early era of stereotactic neurosurgery, thalamotomy was employed more frequently than pallidotomy. One study of 17 patients with dystonia revealed moderate improvement in 8 (47%) patients (56). Another prospective study of 56 patients who underwent thalamotomy for generalized dystonia showed that only 34% of patients had 50% improvement in the symptoms (57). In this study the ventral intermediate (Vim) and posterior ventral oral nuclei (Vop) of the thalamus were targeted, and the procedure ameliorated the symptoms in the distal limbs, but not the axial symptoms. The complications of bilateral thalamotomy have included dysarthria, hemiparesis, pseudobulbar palsy, ataxia, paresthesias, and personality changes. The biggest problem with early thalamotomy studies has been that localization of the target was not meticulous, and outcomes were not measured with standardized validated scales.

Based on studies with animal models (58) and the knowledge that lesioning the globus pallidus internus (GPi) was very effective for alleviating drug-induced dyskinesia as well as for addressing dystonia in PD (59), pallidotomy was then applied for the treatment of dystonia. Yoshor et al. compared the long-term outcomes of thalamotomy and pallidotomy using the global outcome scale (GOS) (60). In the study, pallidotomy revealed significantly higher mean GOS score in the patients with primary dystonia compared to thalamotomy, and they also concluded that pallidotomy was a more effective treatment option for primary dystonia than thalamotomy. Ondo et al. reported that 6 of 8 patients with primary and secondary generalized dystonia had marked improvement, and the Burke-Fahn-Marsden Dystonia Rating Scale (BFMDRS) scores decreased by 59% (61,62). Lin et al., however, reported only 13% improvement in BFMDRS following 12 months of follow-up in a study of 18 cases of secondary dystonia addressed with lesion therapy (63). This discrepancy in the literature highlights the heterogeneous nature of the dystonias and the importance of patient selection and screening. Transient complications

including lethargy and weakness, as well as persistent complications of dysarthria, have also been reported in lesion studies (60,61).

Most specialists with an adequate ability to provide DBS when patients can reliably attend follow-up visits, opt for DBS over lesioning therapies. Much of this shift in the field has been because of the benefits of reversibility, lower risk with bilateral procedures, and the initial positive results of clinical trials (64). In addition, the stimulation parameters can be customized for each patient. Although most reports concerning the outcomes following DBS have been small open-label nonblinded studies, DBS's beneficial effect has continued in selected cases. DBS can be employed for generalized or segmental dystonia, and electrodes can be implanted in the GPi bilaterally (although other targets have been used). The DBS outcomes reported have been highly variable, and we will discuss the impact of the etiology of the dystonia and patient selection on clinical outcome in other sections of this chapter. The most serious complications of DBS include hardware problems, infections, and intracranial hemorrhages. Details on complications of DBS are also provided later in this chapter.

Patient Selection and Screening for DBS

Potential DBS candidates should be referred to experienced teams for a complete multidisciplinary or interdisciplinary screening. The DBS team is typically composed of a neurologist specializing in movement disorders, a stereotactic neurosurgeon, a neuropsychologist, a psychiatrist, and in some cases physical, occupational, and speech therapists (65). Preoperative evaluation includes reviewing history, cognition, mood, and motor function tests. There are no formal age criteria for DBS. The risks and benefits should be discussed for each individual. Underlying medical comorbidities such as cardiac, pulmonary, and other conditions should be evaluated as they can increase the overall risk of surgery. Hypertension should be aggressively treated prior to surgery to avoid potential hemorrhage associated with microelectrode recording (65).

Prerequisite conditions for DBS include an accurate diagnosis, a medication-refractory state, and the exclusion of medication-related side effects affecting the clinical scenario. Potentially important historical points include birth, developmental, and medication history; toxins such as cyanide, manganese, and carbon monoxide; family history, and trauma. Metabolic disorders such as Wilson's disease, glutaric aciduria, propionic acidemia, and methylmalonic aciduria should be excluded. Iron deposition disease, if suspected, should be

adequately screened for, but it should be noted that DBS has been effective in a subset of these patients, particularly those with PANK2 mutations (66). Weakness on examination caused by primary disease such as trauma or stroke will not improve postoperatively despite any improvement of dystonic symptoms following surgery. GPi-DBS may be effective for DYT-1–positive generalized dystonia and also primary generalized dystonia (67,68). Therefore, genetic diagnosis is usually performed for the DYT-1 allele in dystonia patients under the age of 26 as its presence may predict a favorable outcome (69).

Although GPi-DBS does not seem to dramatically affect the cognition and mood of patients with dystonia (68), preoperative psychological and psychiatric evaluation is recommended (65,70). Worsened preoperative anxiety may impact the outcome of the surgery (especially patient satisfaction) even if the surgery is successful in improving motor symptoms. In addition, recently there have been reports of suicide in patients undergoing GPi-DBS (71). Adequate evaluation and treatment of preexisting psychiatric conditions is therefore strongly recommended prior to DBS.

Motor functions may be evaluated with the Unified Dystonia Rating Scale (UDRS), BFMDRS, and Toronto Western Spasmodic Torticollis Rating Scale (TWSTRS). The UDRS and BFMDRS are used for dystonia, and TWSTRS is used specifically for the assessment of cervical dystonia. The Extrapyramidal Symptom Rating Scale (ESRS) can be applied for tardive dyskinesia and the assessment of the efficacy of GPi-DBS (72,73). Joint contractures, bony deformities, or fixed dystonic posture should be elicited, as they usually will not respond to DBS surgery. These features are thus potentially unfavorable characteristics for DBS, and their presence should be discussed with patients and families prior to intervention (70).

Imaging of the brain with magnetic resonance imaging (MRI) or computed tomography (CT) should also be routinely performed to rule out dystonia secondary to a structural lesion and to verify the integrity of targets. The outcomes of DBS for secondary dystonia are highly variable. Primary generalized dystonia and primary dystonias in general seem to respond more favorably than secondary dystonia to intervention with GPi-DBS (74). The best candidates for surgery are thus considered to be the patients with primary dystonia, and this is an important point when discussing outcomes with patients (65,70). Following a thorough preoperative assessment including historical, clinical, psychiatric, and radiographic information, the indications for DBS should be discussed for each individual patient in a multidisciplinary or interdisciplinary meeting and then discussed with patients and families.

The Surgical Procedure

A high-resolution, volumetric brain MRI is usually obtained on or immediately prior to the day of surgery. If MRI-CT fusion is utilized on the morning of surgery, a stereotactic head ring is applied to the patient under local anesthesia. A stereotactic head CT scan is then obtained with the localizer attached to the ring. The CT image is fused to the MRI image so that localization of the subcortical brain structures can be mapped out in coordinate space. If an MRI-compatible head ring is available, some centers opt for using just the MRI. Additionally, many centers have been experimenting with frameless technologies for movement disorders (75). Teams providing DBS should use the technology with which they are most comfortable.

The target point for the tip of the electrode is usually selected utilizing a combination of direct and indirect targeting. In direct targeting, the GPi and the surrounding structures may be visualized. Gross estimations are made of the location of the sensorimotor areas of GPi, which are located in the posterolateral and ventral regions of the structure. The trajectory of the lead should run in the direction of the dorsolateral border of the optic tract in the hope that it will pass through the sensorimotor areas of the pallidum. The Schaltenbrand-Wahren or Schaltenbrand-Bailey atlas can also be superimposed on the image to confirm the target. The anterior commissure (AC), the posterior commissure (PC), and a midline plane are identified to anchor the Cartesian coordinate system. Typical coordinates for GPi of dystonia may range from 18 to 22 mm lateral, 4 to 5 mm inferior, and 1 to 4 mm anterior to the midcommissural point. These coordinates are widely variable, and some dystonia patients seem to have smaller brains when compared to Parkinson's disease (76). The next step for the surgeon is choosing an entry point that will avoid sulci, blood vessels, and, if possible, the ventricles. Indirect targeting may be performed simply by punching in a standard set of coordinates. Most surgeons prefer a combination of indirect targeting that can then be modified with direct visualization.

After injecting a local anesthetic, the skin is incised and a burr hole is fashioned at the entry point. Most centers use microelectrode recording to physiologically refine the target, but a few use only macrostimulation. The microelectrode, when used, is inserted and passed toward the target in the brain. Microelectrode recording (MER) can help to precisely identify neuronal structures. With MER, the borders of GPi and sensorimotor cells can be identified.

There are characteristic firing patterns that may be encountered, but one should keep in mind that in dystonia the physiology of cells may depend on the patient position (worsened with action, so lying down on the OR table may diminish recording) and the type of dystonia. MER tracts are usually started 30–50 mm above the target. The microelectrode typically passes through the striatum, globus pallidus externa (GPe), and GPi in that order. The striatum is usually quiet with little activity, but in one case of blepharospasm and craniofacial dystonia it was reported hyperactive (77). Neuronal activity may be encountered in the GPe, but it is difficult to differentiate GPi from GPe as in general they are both slower than what is seen in Parkinson's and they therefore may resemble each other. Pauses between structures or border cells can also help in their differentiation. Starr et al. reported that while GPi cells have a markedly higher mean firing rate (96 ± 23 Hz) than GPe (52 ± 18 Hz) in patients with PD, the firing rates are nearly identical (55 ± 22 and 53 ± 23 Hz, respectively) in patients with dystonia. He further emphasized the existence of "high-frequency bursting cells" and the absence of "pauser cells" as features of GPi (78). The MER tract typically ends with elicitation of visual evoked potentials by flashing a light into the eyes or providing microstimulation. Depending on the plane and location of the MER pass, practitioners may or may not elicit visual responses, and this can serve as a clue to where they may be within the target. The MER tract information can then be superimposed on the MRI image or atlas to determine more precisely the position of the target. There are many ways to perform MER, including single tract verification, multiple pass mapping, and a Ben Gunn approach, and they have not been systematically compared for outcome. The goal of MER is to define the sensorimotor region of GPi and to estimate the locations of the dorsal/ventral borders of GPi and the approximate location of the internal capsule (located posterior and medial). MER penetrations are usually not made greater than 2 mm deep to the pallidum to avoid injuring a vessel in the choroidal fissure (79).

Some patients, particularly children, cannot tolerate the awake procedure and therefore may require sedation. For optimal preservation of neuronal firing characteristics, propofol and inhalational agents may be avoided (76,78,80). Sanghera et al. recently reported that the discharge pattern of GPi under general anesthesia with desflurane tended to be more irregular than seen in the awake patient (80). As an alternative, dexmedetomidine may allow for useful physiologic MER recordings with mild sedation, although this has not been tested on a large scale (81). Children with dystonia may require sedation to tolerate the

procedure. Similarly, adults with severe dystonia who may pull out of the head frame or induce a frameshift may also require appropriate sedation.

The DBS lead is thus implanted based on stereotactic imaging and MER findings. Macrostimulation may then be performed to detect thresholds for beneficial and adverse effects and to confirm the lead location. A temporary pulse generator can then be attached to the DBS electrode, and stimulation of all four contacts can be performed. Threshold levels of macrostimulation can be correlated with the distance between DBS lead and surrounding structures, especially the optic tract and internal capsule (78,82). If the lead is placed optimally, a motor side effect may be commonly elicited at low voltage (the voltage ranges are widely variable depending on where the contact is within the structure) with bipolar stimulation at a pulse width of 90 μsec and a frequency of 185 Hz (83). Based on capsular motor findings, the optimal lead placement can be adjusted (if needed). This is a critical part of the procedure as the wider pulse widths and higher amplitudes of DBS in dystonia require that the lead not be placed too close to the internal capsule.

The pulse generator can be implanted on the same day or 2–4 weeks later, depending on the surgeon's preference. The pulse generator implantation is usually performed under general anesthesia. In the procedure, the DBS lead is connected to extension cable and the pulse generator. The pathway of the cable is tunneled through the subgaleal space under the scalp and then goes under the fascia of the neck muscles into the subclavicular area. The pulse generator is then implanted and secured in the subclavicular area.

Programming

Postoperative programming for dystonia is perhaps the most difficult of any DBS procedure. Favorable results may take weeks or even months, and the range of effective parameters reported have been highly variable and seem to depend on the individual patient (84,85). The precise effect of each parameter is still unclear. Vercueil et al. tried to detect the effects of pulse width variation in a double-blind controlled study of patients with primary generalized dystonia, but no difference between short and long pulse width was found (86). Programming strategies in dystonia are different from that in Parkinson's disease and must be tailored to the patient. If there is great difficulty in programming, referral to an experienced center should be considered as the lead may be misplaced or the target selection suboptimal. Details on DBS programming are provided in a separate chapter.

Outcomes of DBS in Dystonia

Primary Dystonia

Two important trials have provided class 1 evidence concerning the efficacy of bilateral GPi-DBS for the treatment of primary generalized dystonia, and another trial addressed the treatment of primary cervical dystonia (87–89). Most cases reported were part of small open-label uncontrolled studies. Outcomes of representative literature concerning GPi-DBS for primary dystonia are summarized in Table 1.5 (68,74,78,87–107).

Vidailhet et al. reported a prospective, randomized double-blind multicenter study of bilateral GPi-DBS in 22 patients with primary generalized dystonia. Patients were evaluated preoperatively and at 3, 6, and 12 months postoperatively. A mean improvement of 54% in the BFMDRS movement score and 44% in the BFMDRS disability score were seen following 12 months with chronic stimulation. At 3 months, patients underwent a double-blind evaluation in the presence and absence of neurostimulation on alternating days. Neurostimulation resulted in a statistically significant mean improvement of 29% in the BFMDRS movement score, compared with the unstimulated condition. Recently published data have shown that the beneficial effect has been sustained for 3 years and has also confirmed that bilateral GPi DBS has an acceptable safety profile for dystonia (68).

Another prospective, randomized double-blind multicenter study showed the efficacy of chronic GPi simulation in a series of 40 patients with primary segmental and primary generalized dystonia (88). In the study, the patients were randomly assigned to receive either neurostimulation or sham stimulation for 3 months. At 3 months, the neurostimulation group had a mean improvement of 39.9% in BFMDRS movement scores and 38% in BFMDRS disability scores, while the sham stimulation group had 4.9% and 11% improvement, respectively. In addition, a mean improvement of 45% in BFMDRS movement scores and 41% in BFMDRS disability scores were seen following 6 months of chronic stimulation.

Kiss et al. reported a prospective single-blind study of bilateral GPi-DBS in 10 patients with cervical dystonia. This study showed a mean improvement of 44, 64, and 65% in TWSTRS severity, disability, and pain scores, respectively, at 12 months after surgery (89). Hung et al. reported long-term results of 10 patients with cervical dystonia treated with DBS for up to 5 years, and the mean improvement was 54.8, 52.1, and 50.5% in TWSTRS severity,

TABLE 1.5 Outcomes of GPi-DBS for Primary Dystonia

REF.	AUTHOR	YEAR	STUDY	ETIOLOGY	N	FU PERIODS	SCALE (SUBSCALE)	OUTCOME (% IMPROVEMENT)
90	Vercueil et al.	2001	Case series	Primary generalized	1	12 mo	BFMDRS (m/d)	67/81
				Primary generalized	1	6 mo	BFMDRS (m/d)	70/50
				Primary DYT1+	1	12 mo	BFMDRS (m/d)	86/86
				Primary DYT1-	1	24 mo	BFMDRS (m/d)	41/43
				Cranial-cervical	1	6 mo	BFMDRS (m/d)	66/66
91	Krauss et al.	2002	Case series	Cervical	5	20 mo	TWSTRS (s/d/p)	62/69/50
92	Bereznai et al.	2002	Case series	Segmental	3	3–12 mo	BFMDRS (m)	72.5
				Primary DTY1+	1	3–12 mo	Tsui scale	45
				Cervical	1	3–12 mo	NA	Improved
				Meige syndrome	1	3–12 mo	NA	Improved
93	Yianni et al.	2003	Case series	Primary DYT1+	2	12 mo	BFMDRS (m)	85
				Primary DYT1-	11		BFMDRS (m)	46
				Cervical	7		TWSTRS (s/d/p)	50/38/43
94	Cif et al.	2003	Case series	Primary DYT1+	15	26.6 mo	BFMDRS (m/d)	71/63
				Primary DYT1-	17		BFMDRS (m/d)	74/49
95	Krauss et al.	2003	Case series	Primary DYT1-	2	24 mo	BFMDRS (m)	73
96	Kupsch et al.	2003	Case series	Primary DYT1+	1	3–12 mo	BFMDRS (m)	22
				Primary DYT1-	3			50
				Segmental	1			41
97	Katayama et al.	2003	Case series	Primary	5	6 mo	BFMDRS (m)	51–92
98	Coubes et al.	2004	Case series	Primary DYT1+	17	24 mo	BFMDRS (m)	83
				Primary DYT1-	14	24 mo	BFMDRS (m)	75
99	Vayssiere et al.	2004	Case series	Primary generalized	19	NA	BFMDRS	>80
74	Eltahawy et al.	2004	Case series	Primary DYT1+	1	6 mo	BFMDRS (m)	25

				Primary DYT1-	1			21
				Cervical	3			57
100	Krause et al.	2004	Case series	Primary DYT1+	4	12–66 mo	BFMDRS (m)	53
				Primary DYT1-	6		BFMDRS (m)	32
				Cervical	1		BFMDRS (m)	0
87	Vidailhet et al.*	2005	Prospective, randomized, double-blind multicenter study	Primary DYT1+	7	12 mo	BFMDRS (m/d)	53/45.6
				Primary DTY1-	15		BFMDRS (m/d)	55.4/45
107	Bittar et al.	2005	Case series	Primary generalized	6	24 mo	BFMDRS (m/d)	46/39
				DYT1+	2			
				DYT1-	4			
				Cervical	6		TWSTRS (s/d/p)	58/62/58
101	Zorzi et al.	2005	Case series	Primary DYT1+	1	4 mo	BFMDRS (m/d)	70/45
				Primary DYT1-	8	19.1 mo	BFMDRS (m/d)	32/37
102	Diamond et al.	2006	Case series	Primary generalized	10	27.5 mo	UDRS	15.3
				DYT1+	5			
				DYT-	5			
88	Kupsch et al.*	2006	Prospective, randomized, double-blind multicenter study	Primary generalized and segmental	40	6 mo	BFMDRS (m/d)	45/41
				DYT1+	6			
				DYT1-	27			
				Unknown	7			
78	Starr et al.	2006	Case series	Primary DYT1+	6	13.2 mo	BFMDRS (m)	59
				Primary DYT1-	1	NA	BFMDRS (m)	NA
				Segmental	3	21.7 mo	BFMDRS (m)	47
				Meige's syndrome	1	9 mo	BFMDRS (m)	90
				Generalized*	2	10.5 mo	BFMDRS (m)	12
103	Hung et al.	2007	Case series	Cervical	10	12–67 mo	TWSTRS (s/d/p)	54.8/52.1/50.5

(*continued*)

TABLE 1.5 (*continued*)

REF.	AUTHOR	YEAR	STUDY	ETIOLOGY	N	FU PERIODS	SCALE (SUBSCALE)	OUTCOME (% IMPROVEMENT)
104	Alterman et al.	2007	Case series	Primary generalized DYT1+ DYT1-	15 12 3	12 mo	BFMDRS (m/d)	89/75
105	Tisch et al.	2007	Case series	Primary generalized DYT1+ DYT1-	15 7 8	6 mo	BFMDRS (m/d)	69.5/58
106	Ostrem et al.	2007	Case series	Meige syndrome	6	6 mo	BFMDRS (m/d) TWSTRS (t)	72/38 54
89	Kiss et al.*	2007	Prospective, randomized, single-blind multicenter study	Primary cervical	10	12 mo	TWSTRS (s/d/p)	43/64/65
141	Grips et al.	2007	Case series	Segmental	8	NA	UDRS BFMDRS GDS	55.7 60.6 66.5
68	Vidailhet et al.	2007	Prospective randomized, multicenter study	Primary generalized DYT1+ DYT1-	22 7 15	NA	BFMDRS (m/d)	58/46
115	Loher et al.	2008	Case series	Generalized Cervical Meige's syndrome	2 4 1	3 yr 3 yr 3 yr	BFMDRS (m/d) TWSTRS (s/d/p) BFMDRS eye/ mouth/speech	66/61 29/60/47 92/75/33

AIMS, abnormal involuntary movement scale; BFMDRS, Burke-Fahn-Marsden Dystonia Rating Scale; DYT1+, positive for DYT1 gene mutation; DYT-, negative for DYT1 gene mutation; FU, follow-up; GDS, Global Dystonia Scale; NA, not available; TWSTRS, Toronto Western Spasmodic Torticollis Rating Scale; UDRS, Unified Dystonia Rating Scale.

Subscales and scoring: d = disability; m = movement; p = pain; s = severity.

*Class I evidence study.

disability, and pain scores, respectively (103). Finally, Ostrem et al. reported a series of 6 patients with cranial-cervical dystonia (Meige's syndrome), and their results revealed a mean improvement of 71% in BFMDRS movement scores.

Secondary Dystonia

As discussed above, the surgical outcomes of secondary dystonia are highly variable, and the determination of the best candidates is still under investigation. However, there have been several reports demonstrating favorable results of GPi-DBS for the treatment of secondary dystonia. Castelnau et al. reported a series of 6 patients with pantothenate kinase–associated neurodegeneration (PKAN), and their data showed a mean improvement of 75% in BFMDRS movement scores with up to 42 months of follow-up (66). Three other reports have also revealed favorable results (78,100,108).

Although the number of reports has been limited, it is well known that tardive dystonia may respond well to GPi-DBS. A prospective double-blind multicenter study in 10 patients with medically refractory tardive dyskinesia recently showed the efficacy of GPi-DBS (73). In the trial, patients were evaluated on two consecutive days in a double-blind fashion. This trial showed a 50% significant mean improvement following 6 months of stimulation. Other reports have also demonstrated some improvement in BFMDRS scores (range 35–75%) (109).

Regarding dystonia-plus syndrome, there have been several reports that GPi-DBS improved both dystonic and myoclonic features in myoclonic dystonia (93,110,111). A single case report also demonstrated that GPi-DBS was effective in Lubag syndrome (DYT3) (112). Finally, there has been one case report that GPi-DBS did not improve rapid-onset dystonia-parkinsonism syndrome (113).

The etiology of secondary causes of dystonia such as postanoxic, post-trauma, or cerebral palsy can be extremely variable, and the response to DBS can also be variable (74,78,93,95,101,110,114–117). There have been case reports concerning the application of DBS to multiple sclerosis (93), bilateral striatal necrosis (114), Lesch-Nyhan syndrome (118), glutaric aciduria (74), bilateral basal ganglia calcification (101), and plexus neuropathy (115) (Table 1.6).

TABLE 1.6 Outcomes of GPi-DBS for Secondary Dystonia

REF.	AUTHOR	YEAR	ETIOLOGY	N	FU PERIODS	SCALE (SUBSCALE)	OUTCOME (% IMPROVEMENT)
119	Trottenberg et al.	2001	Tardive dystonia	1	6 mo	BFMDRS (m)	73
						AIMS	54
114	Vercueil et al.	2001	Postanoxic	1	18 mo	BFMDRS (m/d)	3/16
			Bilateral striatal necrosis	1	36 mo	BFMDRS (m/d)	0/0
			Posttraumatic	1	12 mo	BFMDRS (m/d)	72/60
116	Chang et al.	2002	Posttraumatic	1	12 mo	NA	Improved
93	Yianni et al.	2003	Myoclonic dystonia	1	24 mo	AIMS	74
			Myoclonic dystonia	1	4 mo	AIMS	22
			Myoclonic dystonia	1	4 mo	AIMS	65
			MS w/ spasmodic torticollis	1	10 mo	BFMDRS	19
			Tardive dystonia	1	12 mo	BFMDRS	31
			Posttraumatic	1	12 mo	AIMS	0
94	Cif et al.	2003	Secondary dystonia	21	23.1 mo	BFMDRS (m/d)	31/7
95	Krauss et al.	2003	Infantile cerebral palsy	4	24 mo	BFMDRS (m)	18.9
118	Taira et al.	2003	Lesch-Nyhan syndrome	1	24 mo	BFMDRS (m/d)	33/50
74	Eltahawy et al.	2004	Postencephalitic	1	6 mo	BFMDRS (m)	0
			Glutaric aciduria	1		BFMDRS (m)	12
			Huntington's disease	1		BFMDRS (m)	17
			Tardive dyskinesia	1		BFMDRS (m)	35
100	Krause et al.	2004	Tardive dystonia	1	NA	NA	NA
			Tardive dystonia	1	30 mo	BFMDRS (m)	0
			Tardive dystonia	1	42 mo	BFMDRS (m)	0
			Perinatal hypoxia	1	48 mo	BFMDRS (m)	11
			PKAN	1	30 mo	BFMDRS (m)	50
			Posttraumatic	1	12 mo	BFMDRS (m)	16

108	Umemura et al.	2004	PKAN	1	3 mo	BFMDRS (m)	80
110	Cif et al.	2004	Myoclonic dystonia	1	20 mo	BFMDRS (m/d)	84/89
101	Zorzi et al.	2005	Bil basal ganglia calcification	1	50 mo	BFMDRS (m/d)	64/71
			Cerebral palsy	1	28 mo	BFMDRS (m/d)	33/22
			Encephalopathy	1	15 mo	BFMDRS (m/d)	0/0
66	Castelnau et al.	2005	PKAN	6	21 mo	BFMDRS (m/d)	75/53
111	Magarinos-Ascone et al.	2005	Myoclonic dystonia	1	24 mo	BFMDRS (m/d)	48/79
78	Starr et al.	2006	Tardive dystonia	4	20 mo	BFMDRS (m)	60
			Posttraumatic	1	32 mo	BFMDRS (m)	8
			Cerebral Palsy	1	33 mo	BFMDRS (m)	38
			PKAN	1	12 mo	BFMDRS (m)	80
102	Diamond et al.	2006	Basal ganglia hemorrhage	1	104 days	UDRS	22
112	Evidente et al.	2007	Lubag syndrome (DYT3)	1	12 mo	BFMDRS (m/d)	71/62
73	Damier et al.*	2007	Tardive dyskinesia	10	6 mo	ESRS	50
115	Loher et al.	2008	Posttraumatic	1	10 yr	NA	Improved
			Plexus neuropathy	1	10 yr	Frequency of attack	Improved

AIMS, Abnormal Involuntary Movement Scale; BFMDRS, Burke-Fahn-Marsden Dystonia Rating Scale; ESRS, Extrapyramidal Symptoms Rating Scale; FU, follow-up; MS, multiple sclerosis; NA, not available; PKAN, pantothenate kinase–associated neurodegeneration; TWSTRS, Toronto Western Spasmodic Torticollis Rating Scale. Subscales and scoring: d = disability; m = movement; p = pain; s = severity.
*Class I evidence study.

TABLE 1.7 Outcomes of STN and Thalamic DBS for Dystonia

	AUTHOR	YEAR	ETIOLOGY	N	TARGET	FU PERIODS	SCALE (SUBSCALE)	OUTCOME (% IMPROVEMENT)
125	Detante et al.	2004	Primary generalized	1	Bil STN	3 mo	NA	No improvement
			PKAN	3		3 mo		
126	Chou et al.	2005	Cervical dystonia and ET	1	Bil STN	6 mo	TWSTRS (s/d)	79/100
127	Zhang et al.	2006	Tardive dystonia	1	Bil STN	3 mo	BFMDRS (m)	91.9
			Antiemetics	1	Bil STN	3 mo	BFMDRS (m)	90.6
			Neonatal anoxia	2	Bil STN	6 mo	BFMDRS (m)	Did poorly
			Lesion in lentiform nuclei	1	Bil STN			90.8
			Neonatal jaundice	1	Bil STN			Did poorly
			Posttraumatic thal infarct	1	Bil STN			Did poorly
			Neonatal anoxia and jaundice	1	Unil STN & GPi			Did poorly
			No cause	1	Unil STN			Did poorly
128	Kleiner-Fisman et al.	2007	Segmental-major cervical	1	Bil STN	12 mo	BFMDRS (m/d)	21/50
							TWSTRS (s/d/p)	26/26/61
			Segmental-major cervical	1	Bil STN		BFMDRS (m/d)	NA/NA
							TWSTRS (s/d/p)	43/69/16
			Segmental-major cervical	1	Bil STN		BFMDRS (m/d)	-11/-21
							TWSTRS (s/d/p)	-8/11/-20
			Primary generalized	1	Bil STN		BFMDRS (m/d)	72/40
							TWSTRS (s/d/p)	26/88/100
129	Sun et al.	2007	Primary generalized	2	Bil STN	6-42 mo	BFMDRS	76-100
			Tardive dyskinesia	2				
130	Novak et al.	2008	Primary generalized	1	Bil STN	29 mo	BFMDRS	36
122	Sellal et al.	1993	Posttrauma	1	Unil VPL	NA	NA	Dramatically improved

119	Trottenberg et al.	2001	Tardive dystonia	1	Bil Vim & GPi	6 mo	BFMDRS AIMS	Vim stimulation did not improve the symptoms
114	Vercueil et al.	2001	Primary generalized	1	Unil VLp	36 mo	GOS	3
			Primary generalized	1	Bil VLp	6 mo	GOS	2
			Primary multifocal	1	Bil VLp	60 mo	GOS	2
			Postanoxic	1	Bil VLp	96 mo	GOS	0
			Posttrauma	1	Bil VLp	72 mo	BFMDRS (m/d)	28/28
			PKAN	1	Bil VLp	132 mo	GOS	0
			PKAN	1	Bil VLp	120 mo	BFMDRS (m/d)	26/no improvement
			Poststroke	1	Bil VLp	36 mo	BFMDRS (m/d)	31/no improvement
131	Loher et al.	2001	PNKD	1	Unil Vim	4 yr	NA	Improved
115	Loher et al.	2008	Plexus neuropathy	1	Unil Vim	9 yr	Frequency of attack	Improved
120	Fukaya et al.	2007	Writer's cramp	5	Unil Vo-Vim & ipsilateral GPi	2 yr	BFMDRS (hand writing scale)	91
121	Goto et al.	2008	Writer's cramp	1	Unil Vo & ipsilateral GPi	3 yr	NA	Improved

BFMDRS, Burke-Fahn-Marsden Dystonia Rating Scale; ET, essential tremor; FU, follow-up; GOS, Global Functional Outcome Scale; NA, not available; PKAN, pantothenate kinase–associated neurodegeneration; STN, subthalamic nucleus; TWSTRS, Toronto Western Spasmodic Torticollis Rating Scale; VLp, ventralateralposterior nucleus; VPL, ventroposterolateral nucleus.
Subscales and scoring: d = disability; m = movement; p = pain; s = severity.

Subthalamic Nucleus and Thalamic DBS

Although GPi is the most established target for generalized and segmental dystonia to date, the best target for dystonia is unknown. Vim (VLp) thalamus has been employed as a target, and the results of early series of thalamic stimulation for generalized and segmental dystonia have been disappointing (114). However, optimal targets for well-selected individual patients may bring favorable results in the future, and a few reports have demonstrated improvement with thalamic DBS in select patients (120–122). Sellal et al. applied ventroposterolateral nucleus (VPL) stimulation for the treatment of posttraumatic hemidystonia based on the finding that superficial sensory stimulation reduced the dystonic movement and demonstrated dramatic improvement (122). Fukaya et al. reported that Vim-Vo stimulation was more effective for the treatment of writer's cramp than GPi-DBS, and a case report demonstrated the efficacy of Vo-DBS (120,121). The fact that thalamotomy was effective on writer's cramp supported the possibility the thalamic DBS may be a promising treatment option, and we await more complete studies (123,124). In addition, other reports have shown the efficacy of chronic subthalamic nucleus stimulation (STN-DBS) for generalized and segmental dystonia (125–130). All targets other than GPi remain interesting but highly experimental, with CM thalamus also suggested as a future location for stimulation. The results of alternative targets are summarized in Table 1.7.

Complications of DBS

Because DBS is an elective procedure, a careful analysis of the risks and benefits is necessary for patients and clinicians in order to make an informed decision. For this reason, a thorough understanding of the range and likelihood of adverse events (AEs) associated with DBS is paramount. The range of reported DBS-related AEs varies widely, both in terms of rates (from 0 to >40% [132,133]) and categories of events (Table 1.8). The wide variation of reported DBS AE rates has been attributed to the experience level of the implanting center (134); although this may account for some of the variation, the methodology with which AEs are tracked and reported is a more likely an important contributor to this disparity. In fact, a recent meta-analysis designed to ascertain AE prevalence in DBS concluded that it could not be completed in an accurate fashion, primarily because there was an absence of standardized reporting (135) and the fact that AE rates are likely underreported in the literature (136).

TABLE 1.8 Range of Adverse Events Reported in the Literature

RELATED TO STIMULATION	RELATED TO SURGERY	RELATED TO HARDWARE
Temporary paresthesias, 16–81%	Headache, 4–24%	Lead replacement NOS, 20%
Permanent paresthesias, 6–16%	Asymptomatic hemorrhage, 2.5–8%	Erosion 10–12%
1Dysarthria, 2–36%	Symptomatic hemorrhage, 2.5–5%	Infection, 1.8–10%
Disequilibrium, 2.7–23%	Pain, 25–38%	Skin irritation, 10%
Gait disorders, 0–23%	Seizure, 1.7–2.5%	Wire breakage, 2–10%
Dystonia, 0.9–16%	Lead misplacement, 4.5–10%	Lead migration, 4–9%
Mild paresis, 4–16%	Subcutaneous hematoma, 3–5%	Extension replacement, 8%
Increased salivation, 0.8–16%	Cardiovascular, 4.2%	Intermittent stimulation, 5%
Hypophonia, 2.6–11%	Paresis, 5%	Loss of effect NOS, 10–25%
Bone fracture (also surgery-related), 10–11%	Stroke, 3.7%	IPG malfunction, 1.6–5%
Depression, 2.6–18%	Cardiac ischemia, 2%	
Sleepiness, 2–11%	Venous infarct, 1%	
Tremor rebound, 36%		
Incoordination, 33%		
Dysphagia, 24%		
Asthenia, 18%		
Altered mental status/thinking, 2–16%		
Insomnia, speech disorder, 13%		
Accidental injury, bradykinesia, hallucinations, 11%		
Dizziness, facial weakness, nausea 2.6%		
Diplopia, 2%		

Source: Refs. 129,134–144.

Despite these obstacles, much can still be said about DBS AEs. They are usually categorized as related to surgery, hardware, or stimulation. Surgical side effects are usually reported to be low. When noted, deaths and cardiovascular events have been considered to be unrelated to the surgical procedure (132,133,137). Hardware-related AEs, which increase with length of follow-up, can be as high as 27% (138) at 5 years. Overall, stimulation-related AEs occur in 10–42.5% of patients (132,138), and they are more

commonly encountered in bilaterally implanted patients (52%) than in those implanted unilaterally (31%) (132). The side effects of bilateral systems are more persistent and often do not respond to reprogramming (138). The most commonly reported stimulation-related AEs are paresthesias, dysarthria, and gait disorders and disequilibrium. They are frequently viewed as mild and tolerable or amenable to reprogramming (132,137–140). In addition to those listed in Table 1.8, other AEs may include suicide, suicidal ideation, word-finding difficulties, hydrocephalus, cerebrospinal fluid leak, mania, hypomania, air embolus, anxiety, incontinence, orthostasis/syncope, pain, and visual problems. Although recent attention focused on AEs in DBS may make the procedure appear daunting, it turns out that preliminary data reveal that quality of life, motor, and subjective patient-oriented outcomes do not significantly differ when comparing DBS patients with and without AEs (136).

Dystonia patients treated with DBS may be prone to specific AEs. They are often implanted bilaterally, which, as mentioned above, may predispose them to stimulation-related side effects that are persistent and recalcitrant to reprogramming. For GPi, the internal capsule and optic tract are nearby structures, and therefore pulling or visual changes may occur. Furthermore, as more severe forms of dystonia present at younger ages, implanting hardware in children can present special problems. For one, their smaller body sizes are less accommodating to larger IPGs such as the Kinetra, and they may have previously been implanted with baclofen pumps that may limit surgical choices. Also, with growth their DBS leads may be pulled out of the subcortical target as the skull expands, necessitating intracranial lead reimplantation. For patients with cervical dystonia, the dystonic posturing can also cause lead migration, leading to the necessity for intracranial reimplantation. Lead migration and fracture may be more common in dystonia.

Pearls for Practitioners Performing or Managing Dystonia Surgery Patients

1. Potential candidates for DBS surgery should be referred to experienced multidisciplinary/interdisciplinary teams.
2. Hypertension should be aggressively treated prior to surgery to avoid potential hemorrhage associated with microelectrode recording.

3 DBS does not have to be performed bilaterally, and procedures do not have to be performed in one sitting.
4. Patients with primary generalized, cervical dystonia or tardive dyskinesia seem to have the best response to GPi-DBS. In contrast, outcomes of DBS for secondary dystonia are highly variable.
5. Surgery should be performed before contracture or bony changes occur, especially in children, as they do not usually respond to DBS.
6. Preoperative psychological or psychiatric comorbidities may result in a bad DBS outcome. Preoperative neuropsychological and psychiatric evaluation is recommended. If any psychological problems are identified, they should be addressed prior to surgery.
7. Unrealistic patient expectations can result in a DBS failure. Realistic preoperative education regarding reasonable and unreasonable expectations should be clearly established.
8. Patients and families must be willing to agree to multiple programming sessions over many months, as the range of effective DBS programming parameters may be highly variable for each patient.
9. The goal of surgery is not to decrease the medications. Medication adjustment should be performed depending on the individual situation. Abrupt cessation of medications can result in side effects.
10. Symptom rebound can signify a lead fracture or battery failure.

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2

The Treatment of Dystonia with Botulinum Toxins

Ramon L. Rodríguez

The treatment of dystonia can present challenges even for the most experienced practitioner. The available oral therapies often fail to provide clinically significant improvement in a large subset of patients, or alternatively these therapies are associated with intolerable systemic side effects that may limit their use. The introduction of botulinum toxin as a therapeutic option for patients with dystonia has provided the opportunity to treat patients who have previously not been able to tolerate pharmacotherapy or who have failed other multidisciplinary therapeutic modalities.

Botulinum toxin (BTX) has the distinct advantage of application to focal troublesome areas. Only areas affected by dystonia will receive the treatment, which will minimize the occurrence of systemic and other side effects. Experienced practitioners can now deliver targeted treatment to specific muscles and muscle groups, improving posture and frequently minimizing the pain associated with dystonia. In this chapter we will briefly review the history of the medical application of botulinum toxins and discuss their role in the treatment of dystonia.

History

Botulinum toxin is the most potent neurotoxin used to date by humans (1). It is a large complex protein formed by the anaerobic, spore-forming bacillus *Clostridium botulinum*. The bacilli form spores that germinate, reproduce, and produce toxin under appropriate conditions. Following ingestion of the toxin, there can be systemic spread and eventual interruption of the normal neuromuscular transmission. This occurs by prevention of the release of the neurotransmitter acetylcholine (Ach) from the presynaptic nerve terminal

(Figure 2.1). This process will ultimately result in paralysis or in the symptom of muscle weakness. In its generalized form (not caused by focal injections), botulism will result. Generalized botulism is a condition characterized by progressive muscle weakness resulting from systemic spread of toxin, and it can involve the breathing muscles, resulting in pulmonary arrest and death if not treated immediately.

The first description of botulism was made in 1822 by a German physician, Justinus Kerner, who referred to the disease as "sausage poisoning" and even suggested the potential of the toxin applied as a therapeutic modality (2). In 1895, Emille Pierre van Ermengen was able to isolate the bacillus from infected food served at a funeral where several attendees died of botulinum poisoning (2). During World War II Edward Schantz prepared crystalline forms of BTX A, and in 1981 Alan Scott (an ophthalmologist) utilized BTX A for the treatment of strabismus. Following many clinical successes, BTX was then used to treat disorders of the facial nerve, blepharospasm, cervical dystonia, and eventually other conditions associated with muscle hyperactivity (2).

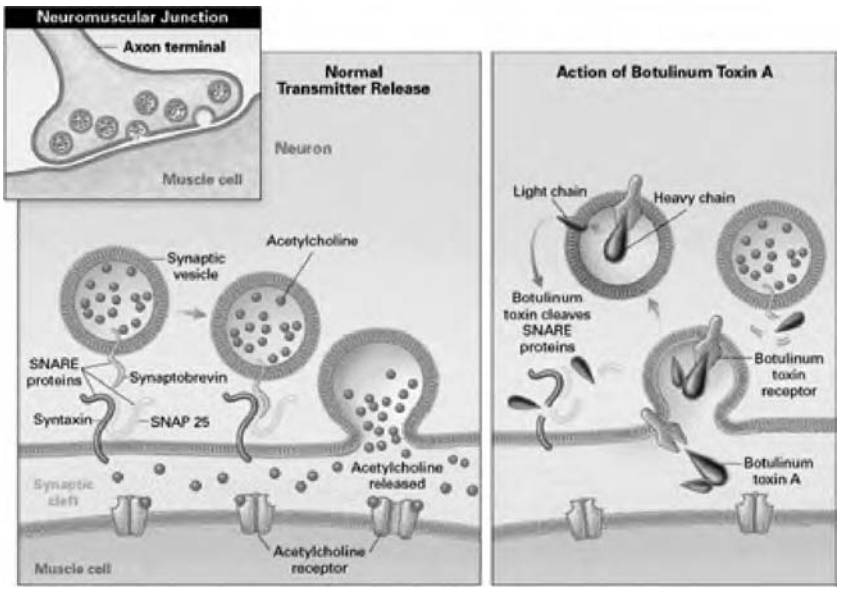

FIGURE 2.1 Acetylcholine in nerve terminals is packaged in vesicles. Normally, vesicle membranes fuse with those of the nerve terminals, releasing the transmitter into the synaptic cleft. The process is mediated by a series of proteins collectively called the SNARE proteins. Botulinum toxin, taken up into vesicles, cleaves the SNARE proteins, preventing assembly of the fusion complex and thus blocking the release of acetylcholine. (From Rowland LP. Stroke, spasticity, and botulinum toxin. N Engl J Med 2002;347:382–383.)

Pharmacology of BTX

There are seven serotypes of BTX, and they are classified as A, B, C, D, E, F, and G. A, B, and F are associated with human disease. The toxin in these cases is produced as a single inactive polypeptide. However, enzymatic nicking of the polypeptide by bacterial proteases can result in the formation of a protein consisting of a light and a heavy chain, united by a disulfide bond (1). The heavy chain mediates internalization of the toxin into the presynaptic terminal, while the light chain is associated with the toxin's effects. The light chain seems to work as an endopeptidase, cleaving the proteins responsible for membrane fusion (SNARE complex) and the proteins necessary for docking of the Ach-containing vesicles into the presynaptic membrane (for eventual of release of Ach in the neuromuscular junction). BTX A, C, and E cleave SNAP 25, while toxins B, D, F, and G cleave VAMP or synaptobrevin; BTX C also cleaves syntaxin in addition to SNAP 25 (1).

Mechanism of Action

The main action of botulinum toxin is to interrupt neuromuscular transmission and to denervate muscle. The process consists of three steps: binding, internalization, and neuromuscular blockade. During binding, the heavy chain irreversibly binds to the presynaptic cholinergic receptors. During the second step, the toxin is internalized by endocytosis; the heavy and light chains are then separated and the light chain translocates into the neuronal cytosol. The last step, neuromuscular blockade, is accomplished by cleaving the proteins responsible for membrane fusion and docking of the Ach-containing vesicles in the presynaptic terminal to the SNARE complex. Each serotype has a different protein through which it exerts action. By this process, Ach is prevented from release into the neuromuscular junction, resulting in paralysis. Evidence suggests that once this occurs, collateral sprouting and upregulation of the nicotinic receptors occurs. Eventually, the original nerve–muscle connection is restored, collateral sprouting regresses, and the synaptic junction returns to the preinjection state (3,4).

Immunogenicity of Botulinum Toxin

Botulinum toxin administration can trigger an immune response resulting in the creation of neutralizing antibodies that can render the toxin ineffective for future injection sessions. It is believed that certain measures and precautions

TABLE 2.1 Recommended Dosages for Frontalis Antibody Test

FORMULATION	DOSE
Botox	15–20 units divided in 2 places
Myobloc	1000 units divided in 2 places

Source: Refs 7,8.

can minimize the possibility of this response, including 1) using the lowest effective dosage, 2) avoiding booster injections, and 3) not injecting patients prior to a 3-month postinjection waiting period (1). Finally, it has also been suggested that the larger the neurotoxin complex, the greater the possibility of development of neutralizing antibodies (5).

Once neutralizing antibody formation is suspected in an individual patient, it may be corroborated by taking a few simple steps. The gold standard is the mouse neutralization assay (6), where antibodies from human serum are administered to mice who have also received toxin. The neutralizing antibodies protect the mice from the lethal effects of the toxin, the neutralization is quantified, and the amount of resistance is reported. This test is expensive and may require a long turnaround time for laboratories to run the assay. A more practical approach is the frontalis antibody test, where a predetermined dosage of botulinum toxin is injected into the frontalis muscle and the patient is assessed 2 weeks following injection. If the patient is able to move his or her eyebrows symmetrically, this may suggest resistance (7). Table 2.1 details the recommended dosages for the frontalis test.

Treatment with Botulinum Toxin

In the United States, botulinum toxin type A was first approved for the treatment of strabismus, blepharospasms, and hemifacial spasm in 1989 and eventually received approval for the treatment of glabellar lines/wrinkles, cervical dystonia, and hyperhydrosis. BTX B is currently indicated for the treatment of cervical dystonia, although many practitioners use it off-label for other types of dystonias. Other conditions that may be addressed with botulinum toxin include spasticity (9), sialorrhea (10), oromandibular and other focal dystonias (11), achalasia (12), tics (13), palatal myoclonus (14), tremors (15,16), headaches (17), and myofascial pain (18). Other formulations of botulinum toxin are available in other countries (Table 2.2). Here we will focus primarily on the treatment of dystonia.

TABLE 2.2 Different Formulas of Botulinum Toxin

FORMULA	TOXIN TYPE	MANUFACTURER
Botox	A	Allergan
Myobloc/Neurobloc	B	Solstice Neurosciences
Dysport	A	Ipsen
Xeomin	A	Merz Pharma
Prosigne	A	Lanzhou Institute of Biological Products (China)
Reloxin	A	Inamed
Meditoxin (Neuronox)	A	Medy-tox Inc.

The Injection Process

The injection process is considered by most practitioners to be relatively simple and straightforward. Botulinum toxin type A (Botox) requires reconstitution with preservative-free normal saline. The dilution, which should be decided by the injector, is commonly formulated into 1 or 2cc. Botulinum toxin type B (Myobloc) arrives in the clinic in a reconstituted form and needs no further preparation (except in particular situations, where a practitioner deems a further dilution is necessary). Insulin syringes are appropriate for the procedure, but other syringes can also be utilized. Alcohol, gauze pads, and 27G and 30G needles are the usual things required by most injectors (Figure 2.2). Whether electromyogram (EMG) is required for the injection is a decision that is up to the practitioner. Multiple reports document successful treatment in cervical dystonia with and without the use of EMG. A study by Comella et al. revealed that there was no difference in the number of patients that reported improvement with and without EMG guidance, but the group treated with EMG guidance reported a more marked improvement (19). The pros and cons of using EMG are presented in Table 2.3 (20,21).

Focal Dystonias

Cervical Dystonia

Cervical dystonia is the most commonly occurring focal dystonia (22). Botulinum toxin types A and B have been approved for the treatment of cervical dystonia in the United States since 2000. Botulinum toxin has been shown

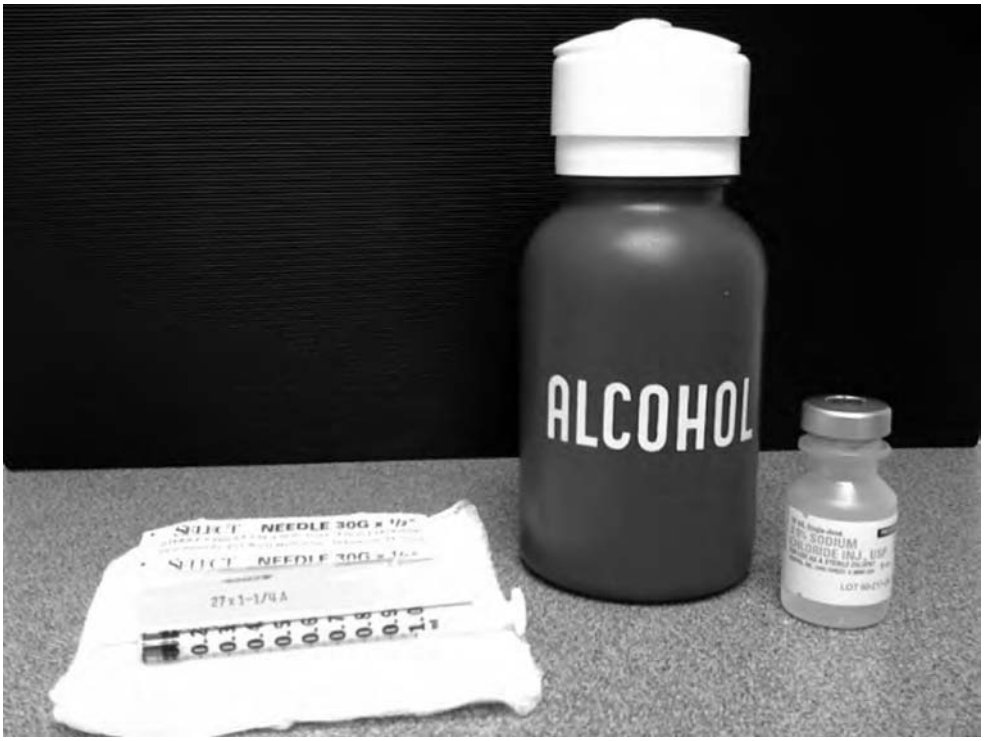

FIGURE 2.2 Tools for injection.

TABLE 2.3 Pros and Cons Concerning the Use of EMG Guidance for the Injection of Botulinum Toxin

PROS (SUPPORTING ARGUMENTS)	CONS (OPPOSING ARGUMENTS)
EMG guidance may improve the clinical effect of injections	Need for additional equipment
Deeper muscles difficult to access	More painful/time consuming/expensive
Further damage with poorly targeted injections	EMG is redundant if the muscle is contracting/hypertrophied
	Cannot confirm exact anatomic location of the EMG needle tip
	No differentiation between contractions produced by agonist versus antagonist muscles
	Needle EMG may be misleading
	Risk of spread to adjacent, uninvolved, and unintended muscles
	Injection treatment without needle EMG has been shown effective

Source: Refs. 20, 21.

to improve both the abnormal posture as well as the pain associated with cervical dystonia (23–27).

The most important goals prior to initiation of botulinum toxin treatment in cervical dystonia are to review the expectations with patients, define the most involved muscles, and choose the most appropriate dosage (25). Cervical dystonia can present in many ways, and having knowledge of the muscles involved for each subtype and/or presentation can be essential. The most common presentations include torticollis (turning of head), laterocollis (head tilts to one side), anterocollis (forward flexion of head), and retrocollis (head extension). Frequently a combination of these forms is observed. Tables 2.4 and 2.5 contain the primary and secondary muscles involved for each type of dystonia and the recommended injection dosages (Figure 2.3). Only muscles judged to be involved in posture or movement are initially targeted for injection. One common theme for injecting patients with cervical dystonia is that it is not always necessary to inject every neck and shoulder muscle group. Caution should be observed when both sternocleidomastoid

TABLE 2.4 Common Muscles Involved in Cervical Dystonia

POSTURE	PRIMARY INVOLVED MUSCULATURE	SECONDARY INVOLVED MUSCULATURE
Torticollis	Ipsilateral splenius capitis	Ipsilateral semispinalis
	Contralateral sternocleidomastoid	Ipsilateral longisimus
	Splenius cervicis	Trapezius
	Levator scapulae	
	Inferior oblique longus capitis	
Laterocollis	Ipsilateral splenius capitis	Splenius cervicis
	Ipsilateral scalene complex	
	Levator scapulae	
	Longissimus	
Anterocollis	Bilateral sternocleidomastoid	Bilateral scalene complex
Retrocollis	Bilateral splenius capitis	Bilateral splenius cervicis
	Bilateral semispinalis capitis	
	Bilateral longissimus	

Muscles compiled from extensive review of the medical literature and *www.wemove.org* (28,29). Other dosages are possible depending on individual experience.

TABLE 2.5 Recommended Injection Dosing for Botulinum Toxins Type A and Type B in Cervical Dystonia

MUSCLE	BOTULINUM TYPE A (BOTOX)	BOTULINUM TYPE B (MYOBLOC)
Sternocleidomastoid	15–75	1000–3000
Splenius Cervicis	20–60	1000–3000
Levator Scapulae	25–100	1000–4000
Splenius capitis	50–150	1000–3000
Scalene complex	15–50	1000–3000
Longissimus	50–150	1000–5000
Semispinalis capitis	50–150	1000–5000
Trapezius	50–100	1000–5000

Dosages compiled from extensive review of the medical literature, *www.wemove.org*, author's experience, and botulinum toxin injection guide (28–30). Other dosages are possible depending on individual experience.

TRAP: Trapezius
SPC: Splenius capitis
SCM: Sternocleidomastoid
LS: Levator scapulae
SCAL: Scalene complex

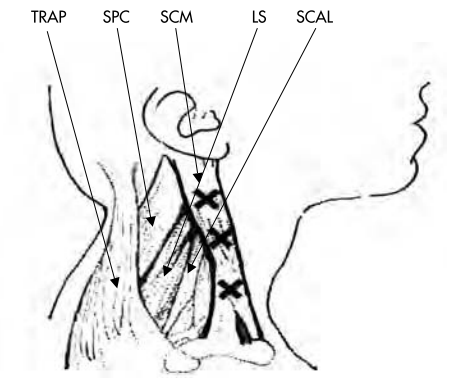

FIGURE 2.3 Frequently injected muscles in cervical dystonia. (From Fernandez HH et al. A Practical Approach to Movement Disorders. New York: Demos Medical Publishing, 2007.)

muscles are injected as dysphagia can occur and is more common with bilateral injections (3).

Blepharospasm

The benefit of botulinum toxin in the treatment of dystonia has been shown in multiple controlled trials (31–36). Blepharospasm is one indication in which it has shown consistent benefit. Blepharospasm usually consists of involuntary, clonic and sometime tonic, forced closure of the eyelids. Botulinum toxin type A has been approved in the United States for the treatment of this condition since 1989. The target muscle to be injected is

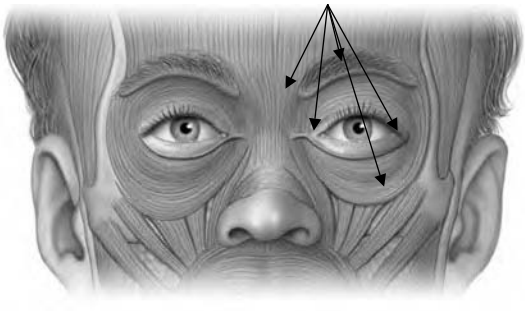

FIGURE 2.4 Recommended injection sites in the orbicularis oculi for the treatment of blepharospasm. (From Fernandez HH et al. A Practical Approach to Movement Disorders. New York: Demos Medical Publishing, 2007.)

TABLE 2.6 Recommended Muscles and Dosages for Injection in Blepharospasm

MUSCLES	BOTULINUM TOXIN A	BOTULINUM TOXIN B (NO FDA INDICATION)
Orbicularis oculi	10–20 units (2.5-5 units per site)	250–1000 units (125–250 units per site)
Frontalis	10–20 (5 units per site)	500–1000 (250 units per site)
Corrugators	2.5–5	250–750
Procerus	2.5–5	125–500

Dosages compiled from extensive review of the medical literature, *www.wemove.org*, author's experience, and botulinum toxin injection guide (30). Other dosages are possible depending on individual experience.

the orbicularis oculi, but some patients may also benefit from injection of the frontalis, corrugators, and procerus (Figure 2.4). When injecting the orbicularis oculi, it is recommended that the pretarsal fibers be targeted (37). Table 2.6 gives information on common recommended dose ranges for specific muscles. Caution should be observed when injecting the upper eyelid in the pretarsal area, and clinicians should attempt to avoid injecting the midline in order to minimize the risk of ptosis.

Oromandibular Dystonia

Oromandibular dystonia can present as jaw opening, jaw closing, jaw deviation, and lingual and orofacial dystonia. The benefit of botulinum toxin for the treatment of oromandibular dystonia has been documented in multiple case reports and open-label studies (35,38–40). Its treatment depends largely on the phenomenology of the specific movement disorder in an individual patient. The specific movement disorder will guide the muscle selection. Table 2.7 shows the muscle involved in each type of dystonia and the recommended injection dosages.

TABLE 2.7 Recommended Muscles and Injection Dosages for the Treatment of Oromandibular Dystonia

PHENOMENOLOGY	INVOLVED MUSCULATURE	BOTULINUM TOXIN A	BOTULINUM TOXIN B
Jaw closing Dystonia	Masseters	25–100	1000–3000
	Temporalis	20–60	1000–3000
Jaw Opening Dystonia	Lateral pterygoids	20–100	1000–3000
	Digastrics	10–20	250–750
	Omohyoid	10–20	500–1000
	Genohyoid	10–20	500–1000
Lingual dystonia	Genioglossus	25–50	500–1000
	Hypoglossus	25–50	500–1000
Jaw deviation	Lateral pterygoids (contralateral to side of deviation)	20–100	1000–3000
Orofacial dystonia (dyskinesias)	Orbicularis oris	1.75–7.5	250–500
	Mentalis	2.5–5	125–250
	Platysma	5–20	500–2500
	Nasalis	2.5–5	125–250
	Levator labii superioris	1.75–5.0	125–250
	Depressor labii	2.5–5	125–250
	Risorius	1–5	125–250
	Zygomaticus	1.75–12.5	125–500

Dosages compiled from extensive review of the medical literature, *www.wemove.org*, author's experience, and botulinum toxin injection guide (28–30). Other dosages are possible depending on individual experience.

Laryngeal Dystonia

Laryngeal dystonia, also known as spasmodic dysphonia, is largely the result of involuntary movements of the vocal folds that result in dysphonia. Multiple open-label studies and case reports have shown the benefits of botulinum toxin for treatment of this disorder (41–44). There are two types of dysphonia: adductor dystonia, producing a strangled voice, and abductor dysphonia, resulting in whispering voice (45). The musculature involved and recommended injection dosing are presented in Table 2.8.

TABLE 2.8 Recommended Musculature and Dosing for Laryngeal Dystonia

DYSTONIA	MUSCLES INVOLVED	BOTULINUM TOXIN A	BOTULINUM TOXIN B
Adductor dysphonia	Thyroarytenoid	1.25–10 (unilateral)	50–750*
Abductor dysphonia	Cricoarytenoid	0.5–6.25	Unknown

Dosing obtained from review of multiple sources including medical literature (41,46,48), author's experience, and *www.wemove.org*.
*Based on case reports and published open-label studies (46–48).

Focal Dystonia of the Limbs

Upper Limbs

Focal dystonia of the limbs can present in the upper extremities as writer's cramp, arm adduction, internal rotation, pronated arm, flexed elbow, flexion of the wrist, extension of the wrist, thumb in palm, clenched fist, intrinsic muscles involved plus hand muscles, extended fingers, and several other unique presentations. Multiple case reports and open-label studies have revealed the benefits of botulinum toxin for the treatment of these conditions (49–54). A combination of these presentations can be observed in the same patient, and the goal for therapy is to find the patterns that are most disabling and to address them by injecting overactive musculature. This specific type of dystonia is particularly disabling when it occurs in the dominant hand. Muscle selection requires careful evaluation both at rest and following exertion. It is important to ask the patient about tightening in particular areas and to discern what the extremity is "trying to do" as well as whether there is an occurrence of tremor. Sometimes with writer's cramp, having the patient demonstrate the movement will reveal the involved musculature in the contralateral (thought to be unaffected) extremity, and this information may help guide injection. The types of upper extremity presentations along with the mostly likely involved musculature are presented in Table 2.9.

Lower Limbs

Focal dystonia of the limbs can also present predominantly in the lower extremities with plantar flexion, foot inversion, equinovarus foot deformity, toe extension, toe curling, and other unique presentations. Botulinum toxin has been used to address these abnormalities (55,56). A combination of these presentations can sometimes be observed in an individual patient, and the goal for therapy is to target the patterns of movement that are most disabling to the patient. This type

TABLE 2.9 Presentation, Musculature Involved, and Dosing Recommendations for Treatment of Focal Dystonia of the Upper Limb

PHENOMENOLOGY	MUSCLES	BOTULINUM TOXIN TYPE A	BOTULINUM TOXIN TYPE B
Writer's cramp	Adductor digiti minimi	2.5–25	125–250
	Adductor pollicis	5–25	500–1500
	Dorsal interoseus	7.5–25	250–500
	Extensor carpi radialis	10–40	500–1500
	Extensor carpi ulnaris	10–40	500–1500
	Extensor digitorium communis	10–30	500–1500
	Extensor indices	2.5–25	500–1000
	Extensor pollicis longus	5–15	250–750
	Flexor carpi radialis	15–50	500–1500
	Flexor carpi ulnaris	15–50	500–1500
	Flexor digitorium profundus	15–40	250–1500
	Flexor digitorium superficialis	15–40	250–1500
	Flexor pollicis brevis	2.5–5	125–250
	Flexor pollicis longus	5–25	500–1500
	Lumbricales	2.5–20	125–1250
	Opponens pollicis	5–35	125–250
	Pronator quadratus	10–35	500–2500
	Pronator teres	10–35	500–2500
Adducted arm	Latissimus dorsi	65–125	2500–5000
Internally rotated	Pectoralis major	75–160	2500–5000
	Teres major	50–100	1000–3000
	Trapezius	50–140	
Flexed elbow	Brachioradialis	25–90	1,000–3,000
	Brachialis	25–75	1,000–3,000
	Biceps	25–175	2,500–5,000
Pronated forearm	Pronator quadratus	10–35	1,000–2500
	Pronator teres	10–35	1000–2500
Flexed wrist	Flexor carpi radialis	15–50	1,000–3000
	Flexor carpi ulnaris	15–50	1000–3000

TABLE 2.9 (*continued*)

PHENOMENOLOGY	MUSCLES	BOTULINUM TOXIN TYPE A	BOTULINUM TOXIN TYPE B
Extended wrist	Extensor carpi radialis	10–40	1000–3000
	Extensor carpi ulnaris	10–40	1000–3000
Thumb in palm	Adductor pollicis	5–25	500–2500
	Flexor pollicis longus	5–25	500–2500
	Thenar group	5–25	500–2500
Clenched fist	Flexor digitorium profundus	15–40	750–3000
	Flexor digitorium superficialis	15–40	750–3000
Intrinsic plus hand	Dorsal interosseus	7.5–25 (muscle group, 2.5/muscle)	375–1250
	Lumbricales	2.5–20 (muscle group)	125–1000

Dosages compiled from extensive review of the medical literature, *www.wemove.org*, author's experience, and the botulinum toxin injection guide (28–30). Other dosages are possible depending on individual experience.

of dystonia can be particularly disabling because it may affect mobility. Muscle selection requires careful evaluation at rest and at exertion. It is important to ask the patient about tightening in particular areas and what the extremity seems to naturally be "trying to do." Observation with the shoes on and off may be necessary as this may reveal the predominant abnormality that must be targeted for success. The types of lower extremity dystonia along with the most commonly involved musculature are presented in Table 2.10.

The Child with Dystonia

Botulinum toxin treatment has been used successfully in the pediatric population in cases of both dystonia and spasticity (10,57–65). Dose modification is necessary when treating the child with dystonia. Dose adjustment is recommended based on weight, muscle bulk, severity of dystonia, and expected function of the extremity (66). Also, dose adjustment should be performed in subsequent injection sessions based on a patient's response and duration of response based on the previous injection. It is difficult to make a particular recommendation when it comes to dosing in children, as there is large variability when it comes to size,

TABLE 2.10 Presentation, Musculature Involved, and Dosing Recommendations for the Treatment of Focal Dystonia of the Lower Limb

PHENOMENOLOGY	MUSCULATURE INVOLVED	BOTULINUM TYPE A	BOTULINUM TYPE B
Plantar flexion/foot inversion	Gastrocnemius	50–200	2500–10,000
	Soleus	50–100	2500–5000
	Tibialis posterior	50–200	2500–5000
Plantar flexion/foot eversion	Peroneus longus	35–85	2500–5000
	Peroneus brevis	40–70	2500–5000
Toe extension	Extensor hallucis longus	20–100	2000–4000
Equinovarus foot	Flexor digitorium brevis	30–80	2500–5000
	Flexor digitorium longus	50–100	2500–5000
	Flexor/extensor hallucis longus	20–100	2000–4000
	Gastrocnemius	50–200	2500–10,000
	Soleus	50–100	2500–5000
	Tibialis anterior	50–200	2500–5000
	Tibialis posterior	50–200	2500–7500
Toe curling	Flexor digitorium brevis	25–100	2500–5000

Dosages compiled from extensive review of the medical literature, author's experience, *www.wemove.org*, and botulinum toxin injection guide (28–30). Other dosages are possible depending on individual experience.

weight, and disease severity. There have been multiple reports of complications in children injected with botulinum toxin, but most of these reports have been in patients injected for spasticity, a condition that usually requires higher dosages of medication (67,68). It is safe to stay either at the lowest range of the recommended dosages or below, based on the judgment of the physician. The total recommended maximum body dose per visit is usually the lesser of 16 units/kg or 400 units for Botox (69). Anxiolitics and topical anesthetics can be beneficial and make the process more comfortable for the child.

Complications of Therapy with Botulinum Toxins

Botulinum toxin has a very favorable side effect-to-benefit ratio. The potential complications of treatment depend primarily on the area of the body being addressed by injection. Patients treated for cervical dystonia may develop

excessive weakness, dysphagia, dry mouth, bruising, and pain (23,24,26,70). The incidence of dry mouth may be more apparent with botulinum toxin type B, as it seems to have a higher affinity for the autonomic nervous system (26,71,72). Flu-like symptoms may be observed and are likely to be related to interactions with the protein complex (73). Those treated for limb dystonia may develop excessive weakness of the involved extremity. Patients treated for blepharospasm may develop ptosis, double vision, and dry eyes (31,33,37). Clinical observation of distant spread of toxin is rare but has been documented by the use of an EMG (74).

Role of the Multidisciplinary/Interdisciplinary Team

Collaboration from various specialties is very helpful for improving care and for maximizing the clinical outcome for individual patients. Specialties that can improve the care of patients with dystonia included physical and occupational therapy, psychology, psychiatry, and social work services. The team approach allows a thorough evaluation and expert advice from multiple specialists and helps to develop treatment goals and assess response to therapy. For example, the upper limb dystonia patient may benefit from, in addition to botulinum toxin therapy, physical/occupational therapy to assess and retrain proper limb motor function as well as psychology and social work services to reintegrate into family and work environments. Additionally, the occupational therapist can make recommendations regarding assistive devices.

Ten Practical Points in the Treatment of Dystonia with Botulinum Toxin

1. Involve all the members of the multidisciplinary team to tailor the treatment plan.
2. Discuss the expectations and potential side effects with patient and family members.
3. Botulinum toxin is not a cure for dystonia.
4. Botulinum toxin can improve posture and reduce pain.
5. Review the anatomy before injecting.
6. Treatment requires repeated injections.
7. Avoid boosters injections.

8. Bilateral subcutaneous mastectomy injections increases risk of dysphagia.
9. EMG may be helpful for guidance.
10. Dosage and muscle selection are the most important factors for success.

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3

The Role of the Nurse Practitioner and Physician Assistant in the Management of Dystonia

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The nurse practitioner (NP) and physician assistant (PA) play similar and complementary roles in providing extensive and quality care to patients. During the 1960s, NP and PA training programs were created in response to a shortage of physicians, especially in medically underserved and economically deprived communities (1). The formal PA role was initiated in the 1960s by physicians at Duke University who recognized that a group of combat medics from Vietnam were skilled and particularly helpful, but lacked formal education. This was the genesis of the first PA education program, and the purpose was to train these medics in a program similar to programs initiated for the training of physicians during World War II. Similar to the first PA program, the first NP educational program was initiated in response to physician shortages in the 1960s. Henry K. Silver, a pediatrician, and Loretta C. Ford, a nursing professor, co-founded the first formal program at the University of Colorado (2).

Education

Historically, PA and NP tracks began as certificate programs, but they have since evolved and grown into graduate training, including the granting of master's degrees. NPs are registered nurses who have graduated from accredited schools and are required to pass a certification exam. The NP primary care tracks (family, adult, pediatrics, geriatrics, and women's health tracks) account for 84% of graduates, with specialty tracks such as neonatology, oncology, psychiatry/mental health, and emergency medicine accounting for the remainder.

NP education emphasizes health assessment, encompassing diagnosis and treatment, and is considered an extension of nursing practice. The average length of education is 26 months (range 15–36 months). NPs are now required to become board certified through a comprehensive written examination before they may practice (3).

PAs have varied backgrounds and may lack medical education and/or experience prior to starting training. Almost all PA programs are primary care–oriented, but two programs emphasize surgery and one emphasizes pediatrics. The average length of education is 27 months, with a range of 20–36 months. PAs are also required to be board certified (3). PA programs are shorter than NP programs when considering that NPs must complete nursing school prior to registering for the additional NP curriculum. Most PA programs are an intensive 2-year experience, with the PA students rotating with medical students.

Scope of Practice

In the outpatient or clinic setting, the PA or NP usually obtains the complete medical history and performs a thorough physical and neurologic examination. They provide care in collaboration with and under the supervision of a physician. NP practice parameters vary state-to-state, with some locales having independent practice NPs (not requiring any physician involvement) and some having a collaborative agreement with a participating physician (4). Clinical professional activities and scope of practice are regulated by state licensing boards (3). In most states NPs work in collaboration with a physician and are professionally autonomous. In contrast, all PAs are required to work under the delegated authority of a physician. Most states do allow practice at a distance from the supervising physician as long as some form of communication is maintained (3).

Areas of Practice and Efficiency

Approximately 85% of NPs and 50% of PAs practice in a primary care setting, including family practice, internal medicine, pediatrics, and women's health. Others may, however, work in surgery, orthopedics, emergency medicine, and other medical specialties (2).

A comparison of the productivity of PAs/NPs and physicians in internal medicine, family medicine, obstetrics and gynecology, pediatrics, and orthopedics

revealed that PAs/NPs were able to evaluate 10% more patients annually in the ambulatory setting when compared to physicians. The productivity, based on number of patients seen per hour, was the same when comparing all three types of providers (NPs, PAs, and physicians) (3). Patients expressed that they were satisfied with their medical care and did not distinguish preferences based on provider type. PAs, NPs, and physicians in primary care are generally viewed similarly regardless of patient characteristics (3).

The Use of NPs and PAs in Neurology and Neurosurgery

Neurology and neurosurgery have effectively utilized both NPs and PAs in the management of patients. The need for NPs and PAs has increased over time as the number of patients has exceeded the capacity of neurologists and neurosurgeons. The use of NPs and PAs in this specialty area has been expanding. One randomized trial looked at whether a trained outreach nurse practitioner could provide quality botulinum toxin injections in dystonia patients at home versus utilizing a hospital outpatient clinic. The results revealed that the trained outreach nurse practitioner provided a service that was as good as, and in certain aspects of practice better than that which was provided by a hospital outpatient clinic. The nurse practitioner provided a more flexible, safe, and cost-effective service for this client group (5). Movement disorders practices, including the diagnosis and treatment of dystonia, have been increasing their utilization of NPs and PAs to meet an almost impossible demand for services.

Role of the NP and PA in the Care of Dystonia Patients

The NP and PA function in a similar way in the care of dystonia patients. Patients with dystonia often require an interdisciplinary or multidisciplinary approach. The NP and PA can assist in the coordination of care for the dystonia patient. They assess and identify both the medical and social needs of the dystonia patient and thus initiate the necessary medical and social services. Specialties that the dystonia patient may require include physical therapy, occupational therapy, communicative disorders (speech and swallowing), psychiatry, psychology, and social work. The NP and PA often provide the dystonia patient and family with the resources for education about dystonia as well as the provision of contact information for support groups, advocacy groups, and dystonia foundations. There are few support groups for dystonia

TABLE 3.1 Websites

Benign Essential Blepharospasm Research Foundation	*www.blepharospasm.org*
The Canadian Movement Disorder Group, Dystonia	*www.cmdg.org/Movement_/dystonia/dystonia.htm*
Care 4 Dystonia	*www.carefordystonia.org*
Dystonia Medical Research Foundation	*www.dystonia-foundation.org*
The Dystonia Society	*www.dystonia.org.uk/index.html*
The Movement Disorder Society	*www.movementdisorders.org*
National Institute of Health, Fact Sheet on Dystonia	*www.nih.gov/about/researchresultsforthepublic/Dystonia.pdf*
National Spasmodic Dysphonia Association	*www.dysphonia.org*
National Spasmodic Torticollis Association	*www.torticollis.org*
PBS: Twisted	*www.pbs.org/independentlens/twisted/*
Tyler's Hope for a Dystonia Cure	*www.tylershope.org*
University of Florida, Movement Disorders Center	*www.mdc.mbi.ufl.edu*
WE MOVE (Worldwide Education & Awareness for Movement Disorders)	*www.wemove.org*

patients nationwide. This may be due in part to the diversity of dystonic syndromes, making each patient more unique than in other diseases. The Internet provides a valuable resource for referral of patients to information and support. (Table 3.1) There are even chat rooms available for patients and family members to ask questions and share experiences.

The NP/PA can also provide specialized and tailored services for the dystonia patient. These services may include baclofen pump programming/refilling, botulinum toxin administration, and deep brain stimulator programming. All of these services have a strong element of education for the patient and caregiver and provide for the continued successful management of the patient.

Examining the Patient with Dystonia

The NP or PA first obtains the individualized and relevant elements of the history for each dystonia patient. The date of diagnosis is determined, and the

person who made the diagnosis is identified (Table 3.2)—primary care physician, general neurologist, or movement disorder specialist. A description of the first symptom is then sought, which may include the age of onset and time interval since onset. History of head trauma, stroke, or family history of dystonia is particularly relevant. Details of the area of involvement as well as a description of dystonic position and accompanying features including duration, involvement of a single body part (focal dystonia), or involvement of multiple body parts should be determined. Situations, activities, or specific positions in which abnormal movements occur, including action, specific tasks, or overflow from one body part to another, should be ascertained. Factors that

TABLE 3.2 Examining the Patient with Dystonia

HISTORY

- Date of diagnosis
- Specialty of diagnosing clinician
- Date of first symptom
- Description of first symptom
- History of head trauma, stroke
- Family history
- Detail of area of involvement
  - Focal or multifocal
  - Age of onset
  - Situations in which movement occurs
  - "Overflow": dystonic movements occur in the the affected limb during voluntary movement of an unaffected one
  - Factors that exacerbate symptoms
  - Factors that alleviate symptoms
  - Tremor or myoclonus

MEDICATIONS

- Duration of therapy
- Maximum dose achieved
- Intervals
- Effectiveness
- Side effects

SOCIAL SUPPORT

BENEFITS FROM THIRD-PARTY PAYER

exacerbate or alleviate dystonia should then be reviewed; these include stress, fatigue, and sleep deprivation. Sleep, hypnosis, relaxation therapy, and geste antagonists (relief by a trick) are factors that may improve or reduce muscle contractions in particular patients. Dystonia may also have accompanying tremor (rhythmical shaking) or myoclonus (sudden jerks). The history should include a review of medications and their doses and effectiveness for specific symptoms of dystonia. Key factors to uncover may include duration of therapy, maximum dose achieved, intervals between doses (e.g., 3 or 4 hours; three times a day with meals), and finally the effectiveness of each medication. Medication effectiveness should include the degree of benefit achieved. Did the patient note a slight, moderate, significant, or noticeable response to therapy? Medication review should include a discussion of side effects, especially those prompting discontinuation of therapy. Interestingly, children are usually able to tolerate much higher doses of anticholinergic medications than are adults. Examples of side effects commonly including mental clouding, blurred vision, xerostomia, urinary retention, constipation with anticholinergics, and somnolence/confusion may occur with benzodiazepines and/or muscle relaxants. Somnolence, nausea, or symptomatic orthostasis (often manifested by dizziness on standing) may occur with use of carbidopa/levodopa or tricyclic antidepressants (e.g., amitriptyline, nortriptyline) (Table 3.3). PAs and NPs should seek a full discussion of the social history with particular attention to individual living circumstances and the social support network. Who is the caregiver, and does the caregiver have adequate support? It should also be determined if respite care is available or may be required. Finally, benefits available from third-party payers with regard to prescription medications should be sought and assistance provided. Knowledge of co-payment for drugs versus out-of-pocket expense can guide therapeutic choices and may be enormously helpful for families.

Clinical Pearls for the PA and NP

The first visit with a dystonic patient can be quite lengthy and time consuming. If the diagnosis of dystonia is made during this visit, be prepared to discuss the implications of the diagnosis. Similar to Parkinson's disease, patients and families generally need more social support and education than medical support at initial diagnosis. If genetic testing for dystonia is considered, pre- and posttest counseling may be indicated. All information usually cannot be

TABLE 3.3 Common Medications and Side Effects

MEDICATION	SIDE EFFECTS
Antidepressant—TCA	Sedation, orthostatic hypotension, weight gain, insomnia, dry mouth, blurred vision, urinary retention, constipation, confusion, sexual dysfunction
Antidepressant—SSRI	Dizziness, tremor, anorexia, nausea, sexual dysfunction, sweating, orthostatic hypotension
Antidepressant—mixed	Nausea, headache, dry mouth, dizziness, insomnia
Anticholinergics	Confusion/mental clouding, blurred vision, dry mouth, urinary retention, constipation
Muscle relaxants	Weakness, fatigue, dizziness, dry mouth, confusion, headache
Benzodiazepines	Somnolence, confusion, headache, ataxia, dizziness
Carbidopa/levodopa	Nausea, orthostatic hypotension, dizziness, confusion, hallucinations
Tetrabenazine	Drowsiness, fatigue, nausea, parkinsonism, depression, akasthisia, orthostatic hypotension
Clozaril	Agranulocytosis, drowsiness/sedation, confusion, dizziness, headache
Medications for erectile dysfunction	Headache, flushing, dyspepsia, diarrhea, nasal congestion, dizziness

provided in a single visit, and patients should be encouraged to participate in a dystonia support group if one is available. The importance of seeking and verifying information from a reputable and reliable source must be emphasized. Encourage activity and exercise with both the patient and family.

Dealing with a challenging dystonia patient can be taxing. The value of diplomacy cannot be underestimated. In some interactions, changing from open-ended questions to direct queries may be helpful. Ask the patient: "If I were to help you with only one symptom, what is most important to you?" This single question may provide insight into what is most bothersome for an individual sufferer. Physicians often concentrate on the physical and neurologic examination without asking about the functional state, an area in which the PA/NP excels. Asking about marital life, fatigue, sleep, and depression can often uncover treatable aspects of the disease that the interdisciplinary or multidisciplinary effort can address (Table 3.4).

TABLE 3.4 Clinical Pearls for the PA/NP

- Be prepared for lengthy first visit
- Provide pre- and postgenetic counseling if indicated
- Encourage participation in support groups
- Emphasize seeking information from reliable sources
- Encourage activity and exercise
- Ask what symptom is most important to him/her
- Ask about marital life, fatigue, depression, apathy, and sleep

Coordinating the Care of a Dystonia Patient

The NP/PA can assist in the coordination of care, which may include referrals to interdisciplinary/multidisciplinary team members (Table 3.5). It may be helpful for the NP/PA to create an available directory with contact information for members of the multidisciplinary team. The directory should be easily accessible by the practitioner as well as by other staff members. The NP/PA may also lead weekly or biweekly interdisciplinary meetings along with the physician to discuss and tailor management of difficult patients.

The NP/PA usually manages a dedicated phone number with voice mail for medical questions. The outgoing message should identify the NP/PA who will be receiving the call. It should also include contact information for new and return patients so that misdirected calls can easily be rerouted to appropriate administrative staff. The message should request that the caller state his or her name, spell it, and include identifying information such as date of birth, medical record number, or Social Security number in order to ensure proper and efficient identification. The caller should be directed to include a phone number at which the patient or representative can be reached. The PA/NP should indicate on the outgoing message that if it is an emergency, the patient or caregiver should call 911. The PA/NP should provide a timeline for returning calls, (within 24 hours, 2 working days, etc.). In addition, establish a dedicated time each day to return calls. There are many reasons why a patient or caregiver may contact a PA/NP or physician (Table 3.5).

The NP/PA may consider using e-mail as a mode of correspondence with patients. Some practitioners find it a quick and efficient way answer many questions. An e-mail consent form would need to be provided to and signed by the patient. A consent form for using e-mail that is hospital and state approved

TABLE 3.5 Common Reasons for Patient Calls

- Discussion of medications and side effects
- Adjustments to medications
- Prescription refills
  - Request that patient contact his/her pharmacist to submit a fax request for refill to the secure fax machine in the office
  - Encourage patients to contact their pharmacist or the clinician's office when the last refill is provided; this will reduce the number of last minute refill requests
- Request for appointment for reevaluation
- Refer patient to primary care physician or emergency department for non–dystonia-related symptoms

should be utilized. Some dystonia patients have difficulty with dysphonia and dysarthia, making e-mail the ideal way to facilitate communication. By utilizing e-mail, communication for both the dystonia patient and practitioner can be made quicker and easier, especially for simple tasks such as medication refills.

Depending upon the structure of a practice, the NP/PA can usually see return and urgent patients more efficiently than new patients. The NP/PA also provides teaching and education to the patient and caregiver and to students in the clinic. This service can include reviewing medication schedules, as well as titration and adjustments based on side effects of medications. The NP/PA can determine what referrals are required for the comprehensive care of the dystonia patient (Table 3.6).

Support groups and community services can be helpful for the dystonia patient and caregiver. The NP/PA can help facilitate participation in a support group. Many groups provide "breakout sessions" for caregivers, which provide an opportunity to share experiences and learn from others suffering in a similar situation. The NP/PA should question the caregiver to ensure adequate support is available. In appropriate situations, the NP/PA should contact a social worker to determine whether community resources may be available for respite care or alternatively for home assistance to provide an often much-needed break for the caregiver.

The outpatient social worker can assist the NP/PA in providing information regarding resources available in the community for respite care as well as home care. Information regarding financial assistance may also be available through the social worker. In cases where placement in an assisted living facility or nursing home may be indicated, the social worker can assist with

TABLE 3.6 Coordinating the Multidisciplinary Care of Dystonia Patients

- Provide a dedicated phone number for medical questions
- Establish dedicated time each day to return calls
- Consider using e-mail as mode of correspondence with patients
- NP/PA can see urgent or return patients
- Provide teaching and education to patient, caregivers, as well as students and other health professionals
- Review medication schedules, titration, and side effects
- Provide information on support groups or community services
- Refer to social worker for evaluation of community resources
- Lifestyle counseling including exercise and stress reduction
- Provide a directory with contact information
- Refer to physical therapy and occupational therapy if indicated
- Refer to speech language pathologist
- Refer for neurosurgery, psychiatry, MRI and neuropsychological evaluation if a surgical candidate
- Refer to psychology and psychiatry as indicated
- Suggest the possibility of using legal counsel for wills, etc.

placement issues and may also be able to assist the family with obtaining financial assistance for long-term care.

The dystonia patient and caregiver may also require lifestyle counseling. This may include discussing stress reduction and activities of daily living. Suggesting and facilitating the use of assistive devices may be helpful and reduce fractures. Assistive devices might enable the person with dystonia to continue leisure activities he or she enjoys.

Physical therapy for stretching, range of motion, and gait and balance training may be beneficial for select dystonia patients. The NP/PA should consider the option of home physical therapy for home-bound patients. Many local area agencies provide home services. In addition to physical therapy, the dystonia patient may require occupational therapy. Occupational therapy may assist with activities of daily living. As an example, a person with writer's cramp (focal dystonia) may benefit from an orthotic. Additionally, a sufferer receiving botulinum toxin injections may synergistically benefit from occupational therapy following each injection.

Some dystonia patients have difficulty with speech and/or swallowing. Such difficulties may include dysarthria, dysphonia, and dysphagia. These

patients may benefit from a referral to a speech language pathology specialist. For patients with dysphagia or difficulty maintaining weight, a referral to a registered dietician or nutritionist may be indicated. In cases of dysphagia that result in difficulty with maintaining adequate oral nutritional intake, the option of placement of a feeding tube should be considered and discussed.

Some patients may have difficulty coping with their disease or alternatively have difficulty with depression. For these patients a referral to a psychologist and psychiatrist may be neccesary. The person with dystonia may need assistance with coping with his or her diagnosis and its impact on his or her daily life. In addition, the parents of a child with dystonia or the spouse of an adult may also be in need of the services of the psychologist. Couples or family counseling may be indicated more often than practitioners realize, and these services should not be underutilized. The strain of caring for a child with dystonia can be difficult on any marriage.

If a patient is a potential candidate for surgical intervention, then the NP/PA should refer the patient to psychology for a neuropsychological evaluation to identify any underlying cognitive deficits and to establish a baseline that can be followed postoperatively. An MRI is obtained to evaluate for any abnormalities and to aid in surgical planning. The patient is also referred to a psychiatrist to evaluate for any underlying anxiety or depression that may be untreated in addition to assessing whether the patient is emotionally ready to undergo a surgical procedure while awake. A consultation is also forwarded to the neurosurgeon, who will meet with the patient to assess his or her surgical candidacy and once again review the potential risks as well as expectations for deep brain stimulation.

The NP/PA may also recommend that the patient or caregiver obtain legal counsel when addressing future care. Decisions regarding wills are best made when one is healthy. The clinician should provide examples of a living will/advance directive. The clinician should also encourage the patient to consider his or her choices for end-of-life issues and discuss them openly with family members. If needed, the clinician should encourage patients to consult with appropriate legal professionals to discuss legal issues as well as healthcare surrogate and power of attorney. An attorney can also assist with Social Security Disability/Medicare coverage issues as needed.

When placing referrals the NP/PA should request that office personnel confirm the accuracy of contact information. In addition, up-to-date insurance information should be sought as many referrals require preauthorization.

Programming and Refilling Baclofen Pumps

Intrathecal baclofen therapy (ITB) is a treatment option in children with generalized spasticity as well as dystonia. ITB was first introduced in the late 1980s, and it has been used in dystonia for approximately 10 years. An advantage of ITB over oral baclofen is that there are fewer systemic side effects. ITB is most effective in the treatment of secondary dystonia. The goals of treatment should be identified prior to considering baclofen pump placement, and indications may include moderate to severe generalized dystonia "that is impeding care, causing discomfort, or impeding function" (6).

Multidisciplinary teams are important in evaluating candidacy for ITB. Responsiveness to intrathecal baclofen is evaluated prior to pump implantation by bolus lumbar injections or by a screening trial with a continuous infusion. Pumps are placed under general anesthesia during a procedure which typically lasts less than 2 hours. A number of complications may occur with ITB pump implants, including cerebrospinal fluid leakage, catheter-related problems, and infections (6,7).

An early side effect is a temporary inability to urinate, which may last 2–3 days. Constipation commonly occurs as a chronic side effect of ITB. When an overdose of ITB occurs, symptoms include hypotonia, decreased alertness, respiratory depression, bradycardia, and coma (6,7). If respiration is not supported, death may occur. The pump may in these cases be temporarily turned off, and supportive care administered until improvement occurs (6,7).

A more common and potentially life-threatening situation is baclofen withdrawal. It is important for patients receiving ITB to have oral baclofen available. Withdrawal can result in an increase in tone, alterations in mental status, pruritis, and agitation. Fever, tachycardia, rigidity, and painful spasms of muscles may occur in moderate withdrawal. In rare cases severe withdrawal seizures, delirium hallucinations, rhabdomyolysis, multiple organ system failure, and death have occurred. It is critical to provide accurate and immediate treatment in an emergency room or intensive care setting to avoid complications and death (6).

A programmer with experience in ITB use is critical, as there are a number of side effects that may occur from either overdosing or withdrawal from ITB, as well as potential malfunction of the pump or catheter, which may require more investigation. Fine-tuning the dose is necessary, and different

modes can be used, including the simple continuous mode, which is used when initiating ITB. Complex continuous mode as well as flex programming may be used later in the course of therapy. Timing of refills is dependent upon several factors, including daily dose, concentration of baclofen, and reservoir size.

Ten Pearls/Practical Tips for the NP/PA

1. The first visit can be time-consuming. Allocate a sufficient interval in the schedule for evaluation.
2. Ask the patient: "What is the most important thing you would like addressed today?" What is most important to the patient may not be readily apparent to the clinician.
3. Remember to ask the family members and caregivers how they are doing. Their well-being is important in providing care for the patient.
4. Utilize all members of the multidisciplinary/interdisciplinary team when addressing the needs of the patient and the family.
5. When genetic testing is indicated, provide appropriate counseling pre- and posttesting.
6. Consider all potential therapies, including oral medications, botulinum toxin injection, physical, occupational, and speech therapy, as well as ITB and DBS.
7. Utilize the resources for counseling, including social work, psychology, and psychiatry, for patient and family members.
8. Provide a list of resources for support groups and websites.
9. Encourage exercise and activity.
10. Utilize the method best suited for answering questions from patients and family members, whether e-mail or phone.

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4

Dystonia from a Social Work Perspective

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Gregory McGann

Imagine that your body is out of your control. You cannot force it do any action that you request it to do. For some strange reason your body twists and turns in odd contortions that are often both painful and awkward. Your mind is clear but your brain cannot seem to receive the proper signals to communicate with the various parts of your body. It is as if your body has decided to stop cooperating with your brain. Tasks that you once performed without thinking twice such as walking, talking, eating, or getting dressed may now require assistance to accomplish. You feel your cherished independence slipping away. Perhaps even worse, imagine you are a parent watching this happen to your child.

This scenario is commonly encountered in persons diagnosed with dystonia, a neurologic movement disorder characterized by involuntary muscle co-contractions, which may force body parts into abnormal, and sometimes painful, movements or postures. Dystonia can be localized and can affect any part of the body including the eyelids, face, vocal chords, or any of the extremities. It can also be generalized, simultaneously affecting the limbs, trunk, and other major body areas (1). Dystonia strikes 500,000 Americans, one third of whom may be children. The disease is chronic, and its prognosis in individual cases may be difficult to predict. There is no cure (2).

In this chapter we will discuss the specific contributions social workers may make to an interdisciplinary/multidisciplinary team for patients, families, and caregivers dealing with dystonia. Social work has evolved to encompass a wide range of skills, including bio-psychosocial assessment, education, communication, advocacy, counseling, and case management.

Each specific skill may offer a valuable perspective to the interdisciplinary/multidisciplinary team as well as to the patient, the family, and the patient's support system.

The diagnosis of any serious, chronic illness may typically be associated with a constellation of psychosocial issues for the patient, family, and caregivers. Some of these issues can present immediately at the time of diagnosis and continue throughout the illness, while others may develop gradually over the duration of the illness. Some problems may resolve over a period of time. It is important to ask about and be aware of a patient's own assessment of his or her *current* experiences and feelings on an ongoing basis. With the consent of the patient, it is also important to ask family members and caregivers for an assessment of patient functioning.

Patients may react to the news of a serious illness in various ways, and some reactions may be more constructive than others. As Falvo notes, "how individuals view their condition, its causes and its consequences greatly affects what they do in the face of it" (3). Some patients express relief at finally hearing a diagnosis that may explain long-standing symptoms, but others may find the words devastating and immediately life altering. Others, when confronted with bad news, may react with denial. Whatever the initial reaction, suddenly having to face the management of any chronic illness often exposes the patient and family to multiple new stressors. This can include depression, grief, false hopes, financial pressures, the disruption of established family roles and difficulty adhering to complicated treatment regimens over the long term. All of the above can put a great deal of strain on interpersonal relationships. For caregivers, depression, fatigue, and burnout are common risks. Elizabeth Holtzman, who writes a blog on chronic illnesses, has noted that the newly diagnosed often feel as if "a new identity has been thrust upon them, and they must learn a new autonomy" (4).

Ideally, the social worker is paired with the patient and the family as soon as possible following the diagnosis. In this situation it is useful to follow the patients' cues on the degree and frequency of support they desire. It may take a while for the patient to digest the information given to them. Initially, simply acknowledging the diagnosis may be a sufficient step toward coping. Other patients and families may want additional information right away about the diagnosis or how to access community resources or concrete services.

The Social Work Assessment

A diagnosis of dystonia presents a unique emotional assault upon the patient and those who will care for them. The origin of dystonia may seem capricious. The illness can be transmitted genetically, or it may result from an infection, reaction to a medication, an accident, or other trauma (immediately or delayed). In many cases there is no obvious cause. Symptoms vary widely and may be dramatic and, in contrast to other chronic illnesses, are often publicly evident. When the mouth, tongue, or vocal chords are affected, speech can be impaired. Although there is typically no cognitive impairment in many dystonia subtypes, some patients are mistakenly identified as developmentally delayed. Dystonia is frequently misdiagnosed, and this can result in delays in receiving effective treatment regimens. In a survey by the Dystonia Medical Research Foundation administered to patients diagnosed with paroxysmal dystonia or dyskinesia, patients reported having seen an average of six doctors prior to receiving a correct diagnosis. Nearly 70% of the patients in the Dystonia Foundation survey were diagnosed incorrectly, usually with a psychiatric condition. An incorrect diagnosis of a psychogenic disorder left these patients feeling overwhelming angry and frustrated (5).

Each member of the interdisciplinary team in the healthcare setting will make an assessment within the context of their discipline. The social work assessment should be of the whole person and take into consideration culture, native language, educational levels, family relationships, social supports, coping mechanisms, emotional strengths and deficits, preexisting stressors, mental health and substance abuse history, financial status, living situation, and other environmental issues. The social work assessment is a critical piece of the overall treatment plan and can help the team incorporate family and cultural values and mores into the treatment plan.

With the historic perspective of "beginning where the client is," social workers are uniquely positioned to support the patient as well as their family and caregivers from diagnosis throughout the course of the illness. Each transition may necessitate a fresh assessment.

As part of assessing the whole person, the social worker should find ways to encourage the patient to "tell his own unique story" and allow him to tell it from his own perspective. Docherty and McColl encourage the use of narrative to prompt patients to share experiences, noting that such patient accounts can be "infused with character, embedded in context and enriched by history" (6).

Such accounts can familiarize the team about their patient in ways much richer than standard intake forms or checklists. Other patients, however, may be better engaged by asking open-ended questions, such as the following:

- How are you feeling after hearing the news of the diagnosis?
- Have you had other diagnoses before this one?
- Do you trust the staff of this facility to treat your illness?
- Who is your family? Who makes up your support system?
- Do you work? Will your employer be supportive of you over the course of a chronic illness?
- Do you have health insurance? Do you understand your plan? Will you be able to access and afford the healthcare and medications you need?
- How do you anticipate this diagnosis will affect your life?
- How will you get to and from medical appointments?

Social workers are trained to be sensitive and respectful of cultural, ethnic, and religious beliefs and may play an important role in sensitizing the rest of the team to these issues and how they may affect treatment (7).

Education/Communication

Another area where the education and training of the social worker can be of particular value to the larger team is helping to facilitate a more complete appreciation of what the person with dystonia (and his or her significant others) understands concerning the diagnosis and what he or she is currently experiencing both physically and *emotionally*. A recent issue of *Dystonia Dialogue* noted that "the vocabulary used to talk about dystonia is often confusing. . . . [E]ven getting an answer for the question 'What form of dystonia do I have?' is not always straightforward" (8).

It is imperative that social workers seek education from all members of the multidisciplinary/interdisciplinary team to familiarize themselves with the terminology of the illness as well as treatment options. Social workers are often responsible for interpreting this information in ways that patients can more easily comprehend. Improving a patient's understanding can be an important step in improving adherence.

Considerable research indicates that the quality of the patient–doctor relationship can influence the success of treatment. The 2008 *Almanac of Chronic Disease* notes: "Many adults with serious chronic diseases feel they

do not receive adequate care for their conditions" (9). The American Medical Association recognized that many physicians have failed to take notice of or overestimated the medical literacy of their patients and have taken measures to educate members about including an assessment of medical literacy as part of the intake process (10). Often the doctor and patient may seem to speak different languages. In an oft-cited study of patient–doctor communication patterns, Mishler noted that doctors used "medical terminology, objective descriptions of physical symptoms and classification within a reductionist-biomedical model," while patients wanted to have "a non-technical discourse about the subjective experience of illness within the context of social relationships and the patient's everyday world" (11). Other studies have indicated that good doctor–patient communication may be more difficult when providers and patients are from different racial or ethnic groups or have different socioeconomic status. For example, nonwhite Americans often receive a different level of treatment for chronic conditions (12). Social workers, with their historical focus on the marginalized and underserved, have a unique perspective to help bridge these gaps between patients and providers.

Children as young as 6 years (and occasionally even younger) can be diagnosed with dystonia, and special skill may be required to help both the child and their family understand what is happening. Hiding disturbing medical news from children is not helpful, as research and experience has shown that "the chronically ill child has some sense of illness severity, even without medical explanations" (13). Families will have different ways of discussing these issues with their children and may rely on healthcare providers to assist them. Mussatto noted that "professionals providing health care have an opportunity to influence how children and families interpret and adapt to these challenges" (14).

As part of the family system, siblings of diagnosed children may have needs that are overlooked. Siblings may feel relatively unseen as more of the parent's attention becomes focused on the child diagnosed with an illness. Siblings may worry that they will also "become victims of the disease." Because many of the symptoms of dystonia may appear odd or dramatic, self-image may be an ongoing concern for children and their siblings, particularly as they become adolescents. Parents, some of whom will already be overburdened, may need guidance about an increasing need for their children to interact with children untouched by the illness.

Richard Cabot, believed to be the first medical social worker, recognized a need to serve as a "translator" between physicians and patients. To this day, social workers continue to fill this critical role. This includes situations in which a true language barrier exists. In a primarily English-speaking facility, patients who do not use English as their primary language may need a translator, or a healthcare professional who speaks their native language, to ensure effective communication. It is important to be aware that a family member, because of a possible tendency to leave out upsetting information or an inability to understand medical terminology, may not be the ideal person to translate for the patient. It is also inappropriate to utilize children as translators since this thrusts them into an uncomfortable role reversal of communicating about more mature adult matters with the parent. To determine what patients perceive, the social worker may ask questions such as:

- Do you understand what the doctor has just said to you about your diagnosis?
- How comfortable are you asking your doctor questions?
- Do you understand the written materials you have been provided?
- How is this diagnosis going to affect you and the rest of your family?

By exploring such questions the social worker can help the team have a more complete understanding of the level of awareness and comprehension of the patient and the support system around them.

Advocacy

Establishing a model for social work in a healthcare setting, the Cornell Center Department of Social Work noted that social workers should directly represent patients and their rights in the healthcare environment (14). Accessing the modern American healthcare system is often unreasonably difficult and can be fraught with frustration. Too often patients may feel subservient to their healthcare providers or lost in large systems that overwhelm them by sheer size and complexity. Determining even basic information such as when or how to schedule appointments, where clinics and labs are located, why certain lab tests are needed, how to determine which services are covered by insurance, where to submit claims, and why doctors need to ask for prior authorizations or exceptions for drug coverage can be daunting. A social work advocate can provide tremendous relief working with systems that are complex with

complicated rules and excessive bureaucracy. Patients who have to figure out these systems in times of ill health and other stressors may feel helpless and ultimately give up. For these patients, social workers may have to periodically assume the role of navigators through complicated mazes of healthcare providers, insurance companies, and other large bureaucratic systems. While social workers normally strive to empower clients, there are times that taking the lead in these situations can be a great stress reliever for overburdened patients or families. Being an assertive advocate when patients become overwhelmed can help build a strong therapeutic alliance.

By virtue of their extensive training, social workers are particularly aware of power differences that exist between the patient and other professionals, agencies, and hierarchical systems that may need to be accessed in the course of an illness. Social workers can use their own professional power to advocate for those without a voice. Knowing the patient's rights and educating patients and families about these rights in a medical setting (patient's bill of rights), in the school system (Individuals with Disabilities Education Act), and/or in the work setting (Americans with Disabilities Act) serves as a valuable contribution to overall patient functioning. It is also important that social workers make the rest of the team aware of the difficulties patients and their families experience as they try to access information and resources.

Helping people in need and addressing social problems and social injustice are part of the social work code of ethics. Social workers collaborate as part of the healthcare team to accomplish identified goals for an individual patient (15). Social workers recognize that there exist central relationships between patients and larger systems. "Relationships between and among people are an important vehicle for change. Social workers engage people as partners in the helping process. Social workers seek to strengthen relationships among people in a purposeful effort to promote, restore, maintain and enhance the well being of the individuals, families, social groups, organizations and communities" (16).

On the macro level, social workers need to be aware of legislation or other initiatives that affect funding and access to healthcare. Helping families and the rest of the multidisciplinary/interdisciplinary team appreciate the relationship between these issues and quality care can enhance long-range planning for current and future patients.

At times social work advocacy for the patient may appear to be in conflict with the rest of the healthcare team. Respecting a patient's right to

self-determination remains, however, a fundamental value of social work. When patients are not ready to pursue certain treatments or wish to delay or discontinue treatment recommended by their doctors, social workers should strive to understand the reasons behind these decisions, support the patient where possible, and facilitate communication between the patient and the rest of the team. Sometimes patients may need additional information or explanation of the treatment plan before obtaining the comfort level necessary to proceed. Other major patient concerns such as financial issues or anxiety about outcomes or side effects may require further advocacy or education. In other instances, resistance from the family or caregiver may have to be addressed before the patient becomes more compliant with the treatment team.

A social worker's primary responsibility is to the welfare of the client, but difficult decisions may arise in situations where the patient declining treatment is impaired or incapacitated. Situations with these components may require ethics consult or peer consultation to clarify capacity.

Again, open-ended questions can help to determine how to best advocate for patients. Some possible questions include:

- How can we collaborate with and empower you and your family?
- Are you having difficulty accessing services? What barriers are you encountering, and how can we help you overcome those?
- Are your caregivers taking care of themselves? Do your caregivers need more support?
- Do you and your family feel heard by your treatment team? And conversely, does the treatment team feel heard by the patient and his or her family or caregiver(s)?

Counseling

Receiving the news of a diagnosis can affect patients and their families in a variety of positive and negative ways. A National Institutes of Health publication, "Coping with Chronic Illness," lists shock, denial, confusion, fear, avoidance, anger, grief, and guilt as common reactions (18). LeMaistre noted that "the experience of serious illness has been approached in two ways: (1) a gloomy perspective of resignation, self denial, and helplessness, or a Pollyanna approach that denies altogether that there has been a real trauma" (19). Beattie, while acknowledging that an emotional reaction to

news of a diagnosis of a chronic illness is natural, has noted that for some, the experience is similar to symptoms of posttraumatic stress disorder, where the trauma is reexperienced through dreams, nightmares, or intrusive thoughts (20).

Some patients express relief at finally learning the reason for their symptoms. Patients who are relieved by a definitive explanation for troubling symptoms may have begun to doubt themselves or question their own mental health. Frequently, though, this sense of relief is short-lived as the reality of how their life change begins to settle in.

Several studies have documented the psychological impact of chronic illness on patients and their support systems. One study noted that up to one third of individuals with a serious medical condition experienced symptoms of depression (21). Another study showed that in newly diagnosed multiple sclerosis patients, nearly half of the patients and their partners had high levels of anxiety and distress (22). When these concerns are not adequately addressed, a cycle of worsening symptoms—of both the illness and the depression—may result.

It should be noted that there exist "conflicting data about the impact of chronic illness on the mental health of children" (23). Pao et al. noted that assessing psychiatric illness in children is often difficult because of "physical symptoms that interfere with diagnostic measures." Pao has also noted that this may lead to both overdiagnosis and underdiagnosis in children (24). The care team should therefore always explore the feelings of the patient. Edwards et al. additionally found that adolescents with disabilities rated themselves lower on quality-of-life scales and scales of depressive symptoms than adolescents without a disability (25), and this should be considered in younger dystonia patients. Burke and Elliot noted that "characteristics of the child not associated with health status are also important determinants of psychological status in pediatric illness." They have stressed the need for a model that considers the "illness parameters, characteristics of the child and environmental factors." They cite other studies that have confirmed that children with a brain-related illness have more psychological or behavior disorders than do children with other illnesses, but not all children with chronic illness seem to develop depressed symptoms (26). A common theme in a Dystonia Medical Research Foundation publication (written by young people), "I Will Gallop Instead of Run, I Will Type Instead of Write," underscores the importance of having a positive attitude when dealing with and developing coping mechanisms for illness symptoms (27).

Interdisciplinary teams in a movement disorder clinic setting often include psychiatry and/or psychology practitioners, and these professionals should be actively involved in the regular care of patients. Anti-anxiety medications or antidepressants may be necessary to help patients deal with the diagnosis and ongoing course of dystonia. Medications may not be enough or may be contraindicated for specific conditions. Some antidepressants, dopamine blockers, or anticonvulsants have been linked to causing or worsening dystonia symptoms.

Grohol, in a meta-analysis of the effectiveness of antidepressants by themselves or in combination with psychotherapy has noted that "combined treatment of psychotherapy and medication is the usual and preferred treatment of choice for depression" (28). When giving support, the social worker can stress the synergistic combination of both approaches.

As primary providers of mental health services, social workers should be utilized to provide ongoing counseling to the family system, especially in situations where the patient may be reluctant to add another doctor (psychiatrist or psychologist) to his or her increasingly crowded life. It is important that the counselor be knowledgeable and comfortable in dealing with chronic illness. It is essential to recognize the grieving process the patient and their families may be experiencing as they deal with the losses associated with previous functioning. Pacing is very important. Be aware that many patients cannot fully begin the problem-solving process until they have sufficiently grieved their losses.

A unique perspective that social work brings to a multidisciplinary/interdisciplinary team is the examination of the impact of the illness not only on the patient, but also on the family. An essential part of the social work role is to provide understanding and support to family members and significant others who may inadvertently be placed in the role of caregiver. In addition to the patient assessment, the social worker should also explore the motivation, abilities, and obligations of the caregiver. For example, a spouse or a parent of a patient may already be taking care of an elderly parent. It can be especially challenging when the person in the role of family caregiver is diagnosed and consequently becomes a patient as well. Dystonia in some cases can be genetically transmitted, and more than one child may become symptomatic, further burdening parental caregivers.

Illness and stress have a huge impact on the family system. Intimacy issues may develop between the patient and his or her partner or between the parents of a diagnosed child. This in turn may increase conflict. These issues are often

difficult for families to talk about. Seeking out counseling and practical resources (respite care) would be important to explore and encourage.

Support groups can be beneficial for some patients and caregivers, and social workers are often natural facilitators for support groups. A downside of support groups has been the difficulties people have in accessing them. Patients may have difficulty traveling, and caregivers may not be able to leave the patient for the requisite amount of time. Newer generations may find that online resources, support groups, and chat rooms are easier to access and more useful.

Often the treatment patients receive can affect them both physically and emotionally. Botulinum toxin injections, for instance, can take up to 2 weeks to take effect and may improve functioning for 6–8 weeks but then begin to wear off. One patient described it as being on an emotional and physical roller coaster every 3 months. On a more practical note, this patient learned to schedule the activities she most needed to accomplish during the peak time of the medication effectiveness. During other times of the cycle, she required more counseling support. She found that help and understanding in an informal support group she had created of other women who also had a chronic illness.

Professionals should follow the patient's lead regarding counseling but be aware that some families will be reluctant to request such help. The challenge in social work services is to identify the barriers that prevent patients from achieving their goals and help discover what actions the patient and family can undertake so that they can take back control of their lives. Occasionally patients need to be reminded that they are much more than a disease. Their medical diagnosis, while an integral part of their lives, does not define their totality overall as a person. Healthcare professionals may periodically need to remind each other of this fact as well.

When helping patients and families deal with their counseling needs, social workers should ask questions such as:

- Would you be interested in a support group you can attend face-to-face or online to help for problem solving issues etc?
- If not, is establishing a website to communicate with others a feasible alternative?
- Would you be interested in getting involved with fundraising for researching a cure?
- Who are you, outside of your diagnosis?

- What needs to happen so that caregivers can get back to some of the activities they enjoyed before the diagnosis?
- Is there an assistive device that could help patients get back to activities they enjoyed before? Alternatively, is there a new activity or interest they can pursue?

Case Management

Social work expertise in the area of case management in healthcare is typically utilized with inpatient hospitalizations, but these skills have equally important application during outpatient clinic visits (and in some cases can prevent hospitalization). Definitions of case management (sometimes called care management) are widely variable depending upon the profession involved. Social work case management is a method of providing services whereby a social worker collaboratively assesses the needs of the client and the client's family, when appropriate, and arranges, coordinates, monitors, evaluates, and advocates for a package of multiple services to meet the specific client's complex needs. Distinct from other forms of case management, social workers address both the individual client's bio-psychosocial status as well as the state of the social system. It is important to distinguish true case management from managed care—some insurance carriers use the terms interchangeably. The focus of managed care is on cost containment, whereas the focus of case management is on the best and most appropriate treatment plan for the patient (29).

Any social worker assisting with case management planning must be knowledgeable about all available resources and involve the patient to the greatest extent possible in all phases of planning. The ultimate goal of case management is to optimize patient functioning. Good case management contributes to improved continuity of care while maximizing efficient and cost-effective interventions.

A major resource for any social worker with this population is the Dystonia Medical Research Foundation. Patients should be aware of this resource early in their disease course. In the survey cited earlier in this chapter, 100% of the respondents reported that their physician failed to mention the Dystonia Medical Research Foundation as a resource (30). The Dystonia Foundation provides a wide variety of educational materials that explain the different types of dystonia and include literature written for children ages 8–12 and another specifically for teen-aged patients. Each booklet includes lists of excellent resources (disabled sports groups, travel venues, camps, and adapted

driving). The Dystonia Medical Research Foundation also offers a catalogue of videos, CDs, tapes, booklets, and the magazine *Dystonia Dialogue*. Other pamphlets include dealing with caregiver stress and strategies for applying for Social Security Disability benefits (for adults and children). There is also a guidebook for parents on the educational rights of disabled children and how to obtain school-based services (how to set up an Individualized Education Plan [IEP] or 504 plan). This kit provides clear information that will arm parents with core tools and suggestions on how to get the school to respond to a child with special needs. The school system is an essential resource that parents will need to know how to access when their child is diagnosed. The Dystonia Medical Research Foundation also has information on assistive devices, a website for possible funding sources, and a used medical equipment exchange. There is also the Foundation's "Operation Friendship" program, which attempts to match members to meet or talk by phone.

Summary

"The hardest thing to find out is that it's permanent. You're not going to die from it . . . but you're going to live with it for the rest of your life" (31).

"Professional social work seeks to enhance adaptations among clients and the systems within which they are embedded" (32).

"The Profession, (Social work), attempts to function in an environment of obstructive administrative 'systems,' severe financial restrictions and conflicting demands" (33).

The first two sentiments reflect the long-term struggle faced by patients diagnosed with dystonia and the focus of social workers as part of a multidisciplinary or interdisciplinary team. The third quote reminds us of the institutional barriers social workers face as they strive to provide the support necessary for patients and caregivers along the journey.

Each patient will engage his or her specific dystonia and accompanying symptoms on his or her own terms. Just as the location and severity of patient symptoms will vary, so will their needs, their inner strengths, coping skills, and family supports. One important constant in the patient's journey should be the support of a skilled interdisciplinary team where the primary focus is on the patient's well-being as well as the needs of their caregivers and families. Each discipline will bring specific training and skills, and none will have the complete answer.

The unique contribution of the social work profession is to recognize the strengths and needs of each patient, family, and caregiver relative to the larger systems that impact their lives and to work toward achieving and enhancing optimal adaptations. Each social work intervention should be designed to support the patient in a way that recognizes the totality of his or her life.

Ten Practical Tips for Social Workers

1. You need to be able to communicate with your team. Educate yourself about dystonia, know the terminology, and understand treatment options.
2. Do a thorough psychosocial assessment as soon as possible with the patient and family so you know their situation. Reassess periodically.
3. Honor the social work dictum: Begin where the patient is.
4. Know the resources. Begin with having written materials and website information on hand from the Dystonia Medical Research Foundation.
5. Remember that dystonia affects the body, not the cognition, personality, creativity, imagination, or sense of humor.
6. If you do not have time to do the necessary counseling yourself, then refer to a therapist who understands the impact of chronic illness.
7. Be aware of power imbalances. When necessary, network and advocate for the patient and family to help them obtain what they need.
8. When resources are inadequate, educate the team and public about unmet needs and work collectively to try to develop new ones.
9. Be aware of the emotional and physical health of the caregivers. Provide burnout-prevention strategies.
10. Respect the boundaries of the team, the patient, the family, and yourself. Liberally apply burnout-prevention strategies for yourself.

Important Websites and Phone Numbers

www.dystonia-foundation.org
1-800-377-3978 (U.S.)
1-800-361-8601 (Canada)
www.wemove.org (U.S.)
wemove@wemove.org (Canada)
www.care4dystonia.org

www.tylershope.org (a father's blog about his son, and a charity raising money to cure dystonia)
Botox Reimbursement Hotline:
www.BOTOX.com/reimbursement (U.S.)
1-800-530-6680 (U.S.)
www.allergan.ca (Canada)
1-877-255-3746 (Canada)
www.socialsecurity.gov.
1-800-772-1213
www.abledata.com (assistive devices)
1-800-227-0216 (U.S.)
www.abilities-ca
1-416-923-1885 (Canada)
www.exceptionalparent.com (publishes Exceptional Parent Magazine)
1-201-634-6550
www.needymeds.com (lists all drugs offered under drug company patient assistance programs and phone numbers and applications to apply for help)
888-445-4588

Resources for Caregivers

www.familycaregiving101.org
www.caregiver.com
www.thefamilycaregiver.org
www.agingcare.com
www.4therapy.com

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5

Speech and Swallowing Disorders in Dystonia

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John C. Rosenbek
Harrison N. Jones

This chapter discusses the role of the speech-language pathologist (SLP) in the evaluation and treatment of patients with dystonia. It is divided into three parts. The first two sections will discuss swallowing and speech function in patients with dystonia, providing definitions of normal and abnormal function, incidence data, signs and symptoms of abnormalities, evaluation strategies, and treatment guidelines. The chapter will end with ten practical tips for the SLP working with this patient population. For definitions of types of dystonia discussed in this chapter, readers are directed to the medical and surgical approaches chapter in the book.

Dystonia and Swallowing

Definitions of Normal and Abnormal Swallowing

Normal swallowing is defined as the efficient and safe transfer of liquid, including saliva, and food from the mouth into the stomach. Abnormal swallowing, or dysphagia, is defined as inefficient or unsafe transfer of material from the mouth to the stomach. Swallowing is a highly patterned response capable of variable degrees of volitional control, which has traditionally, though perhaps a bit artificially, been divided into three stages: oral, pharyngeal, and esophageal. Abnormalities can occur throughout any of the three stages of swallowing and are discussed in this traditional manner. Oropharyngeal dysphagia is inefficient or unsafe transfer of material from the mouth through the pharynx and into the upper portion of the esophagus, while esophageal dysphagia is inefficient or absent movement of material through the esophagus and into the stomach (1).

Incidence of Dysphagia in Dystonia

Dysphagia and feeding difficulties can occur across several forms of dystonia in both children and adults. Dystonias that affect the musculature involved in swallowing, such as cervical, lingual, jaw, oromandibular (OMD), laryngeal, and generalized dystonia involving the respiratory mechanism, represent such forms. Reported incidences of dysphagia in patients with dystonia vary widely, likely due to the locus of involvement, severity, etiology, and patient sampling methods (2). Table 5.1 provides a summary of the literature on swallowing in dystonia, including reported incidences, swallowing signs and symptoms, treatments, and outcomes.

Swallowing Signs and Symptoms in Dystonia

Cervical Dystonia

Swallowing function in patients with cervical dystonia (also known as spasmodic torticollis) has been studied more extensively than in any other form of dystonia. Cervical dystonia is the most commonly encountered dystonia type, and this condition is characterized by abnormal head and neck postures brought about by involuntary, sustained, or repetitive neck muscle contractions (3). In some series the videofluoroscopic swallowing examination (VFSE) reveals dysphagia in about half of subjects (4–6). The most common swallowing signs include a pharyngeal swallow delay (3,7,8), postswallow vallecular residue (8,9), and decreased tongue base retraction (3). Postswallow residue, including vallecular retention, is seen in Figures 5.1 and 5.2. Swallowing symptoms reported by patients include discomfort during swallowing (10). Of clinical note, several studies have reported that patients did not complain of any difficulty swallowing, though instrumental examination revealed dysphagia (3,5,8). The SLP should always therefore be vigilant for subclinical swallowing abnormalities, even in the absence of complaints of dysphagia.

Several possible mechanisms have been proposed for dysphagia in cervical dystonia. First, a "mechanical dysphagia" may be caused by the abnormal posture of the neck (e.g., rotated or tilted head or dystonically overactive neck musculature) leading to asymmetry during swallowing (3). Second, it has been proposed that a neurogenic cause may lead to a pharyngeal swallowing delay (3,7). Munchau et al. (3) postulated that abnormalities of pharyngeal sensory processing may have been responsible for this finding.

TABLE 5.1 Summary of Literature Reporting Swallowing Abnormalities in Dystonia

REF.	DYSTONIA TYPE (N)	EVALUATION METHOD	DYSPHAGIA INCIDENCE	SWALLOWING SIGNS AND SYMPTOMS IN INDIVIDUALS WITH DYSTONIA	OUTCOMES OF TREATMENT
11	Focal (2) General (1) OMD (1)	Chewing patterns via surface EMG of: -anterior temporalis -masseter muscles -anterior belly diagastric	N/A	• Involuntary "munching" in focal dystonia patients • Patients with focal dystonia reported pain with chewing • Distorted, slow chewing pattern with abrupt rhythmic changes due to repeated blockages in jaw opening or closing phases	N/A
9	Cervical (18)	CSE VFSE	11% pre† 44% post† 22% pre† 72% post†	• Postswallow pharyngeal residue • Difficulties managing solids • Two patients required a liquid diet due to severe amounts of residue that they were unable to clear • Distal esophageal abnormalities	Botulinum toxin injections: • Found to worsen swallow signs in this group of patients • Patients were reported to use multiple swallows to clear residue
7	Cervical (25 patients) (25 controls)	CSE and patient report Electrophysiological -submental EMG -larynx piezoelectric sensor	36%† 72%†	• Pharyngeal swallow delay based on submental EMG • Laryngeal relocation time prolonged • Cricopharyngeal sphincter muscle hyperreflexive in some patients	• Patients reported special head and neck maneuvers such as holding or fixing the head while swallowing to be particularly useful

(*continued*)

TABLE 5.1 (*continued*)

REF.	DYSTONIA TYPE (N)	EVALUATION METHOD	DYSPHAGIA INCIDENCE	SWALLOWING SIGNS AND SYMPTOMS IN INDIVIDUALS WITH DYSTONIA	OUTCOMES OF TREATMENT
4	Cervical (41)	VFSE	68.3% pre† 95.1% post†	• Swallow abnormalities most common during pharyngeal phase • Abnormal bolus preparation • Postswallow vallacular residue • Pharyngocele (pharyngeal outpouching) following surgery encountered • Aspiration more common post surgery • Duration of torticollis and subjective complaints correlated significantly with swallow dysfunction	Rhizotomy: • Aggravated pharyngeal swallow dysfunction • Gradual improvement 4–24 weeks postsurgery via self-report noted
5	Cervical (8)	VFSE	75%*	• Asymmetric bolus transport • Postswallow pharyngeal residue, particularly in the vallacular	N/A
12	Meige's syndrome (7 patients) (7 controls)	Mastication via surface EMG from: -masseter -orbicularis oris	N/A	• Excessive muscle activation • Frequent contraction • Loss of rhythmicity • Increased co-activation • Abnormal chewing to swallow phase	N/A
13	Cervical (12)	VFSE	92%*	• Pharyngeal swallow delay • Decreased tongue base retraction • Poor oral bolus preparation • Subclinical dysphagia—only 17% and 33% of patients complained of dysphagia pre- and postsurgery, respectively	Selective peripheral denervation surgery: • Worsening of pharyngeal swallow delay and oral bolus preparation in 25% of patients • 58% of patients demonstrated improved tongue base retraction

14	Neuro-epileptic drug-induced OMD (32)	CSE	15.6%†	• Decreased bolus preparation and bolus propulsion due to involuntary jaw opening and tongue thrust behavior • Painful chewing necessitating a pureed diet • Inability to intake food (liquids only through a straw) due to jaw-closure dystonia • Weight loss	• Tetrabenazine with antidystonic meds and/or botulinum toxin injections to lateral ptyergoids improved dysphagic symptoms
8	Cervical (43)	VFSE	51.2%†	• Postswallow vallecular residue • Pharyngeal swallow delay • Abnormal bolus preparation • UES dysfunction • Pharyngocele occasionally encountered • Subclinical dysphagia noted with only 34.9% of patients c/o swallowing problems	N/A
10	Focal: lingual (8)	CSE	N/A	• Choking and aspiration pneumonia • Drooling and poor oral control due to involuntary jaw opening and tongue protrusion • Feeding difficulties due to jaw closure requiring pureed diet and percutaneous gastrostomy tube in one case	• Botulinum toxin injections into the genioglossis muscle effective to control tongue protrusions and both masseter muscles in the patients who also had jaw closure • Bilateral globus pallidus pars interna deep brain stimulation surgery greatly relieved tongue protrusion dystonia

(*continued*)

TABLE 5.1 (*continued*)

REF.	DYSTONIA TYPE (N)	EVALUATION METHOD	DYSPHAGIA INCIDENCE	SWALLOWING SIGNS AND SYMPTOMS IN INDIVIDUALS WITH DYSTONIA	OUTCOMES OF TREATMENT
					and resulted in marked improvement of swallowing
10	Cervical (327)	Evaluated side effects of botulinum toxin injections in cervical dystonia	27%†	• Dysphagia was the most common side effect of botulinum toxin injections in cervical dystonia • Pain during swallowing was the third most common side effect (5.1%)	Botulinum toxin injections: • On average, dysphagia occurred 8.2 days after injection and lasted 14.9 days

CSE, clinical swallowing examination; VFSE, videofluoroscopic swallowing examination; OMD, oromandibular dystonia; UES, upper esophageal sphincter; †, swallowing dysfunction was not a prerequisite for inclusion into study; *, patients selected for study were referred to swallowing clinic due to suspected dysphagia.

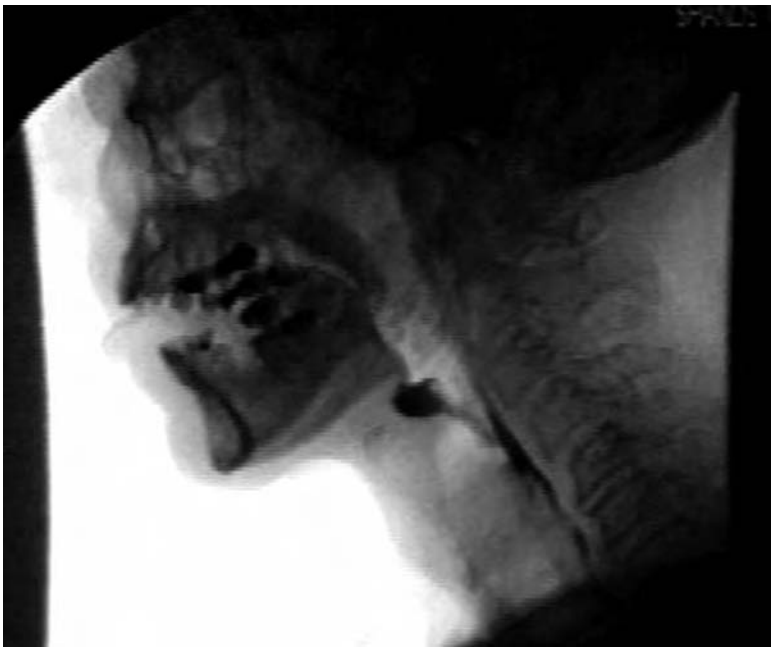

FIGURE 5.1 Lateral view of a patient during VFSE. Patient is swallowing food of a mechanical soft consistency with residue in the vallecular, aryepiglottic folds and into the upper esophageal sphincter (UES).

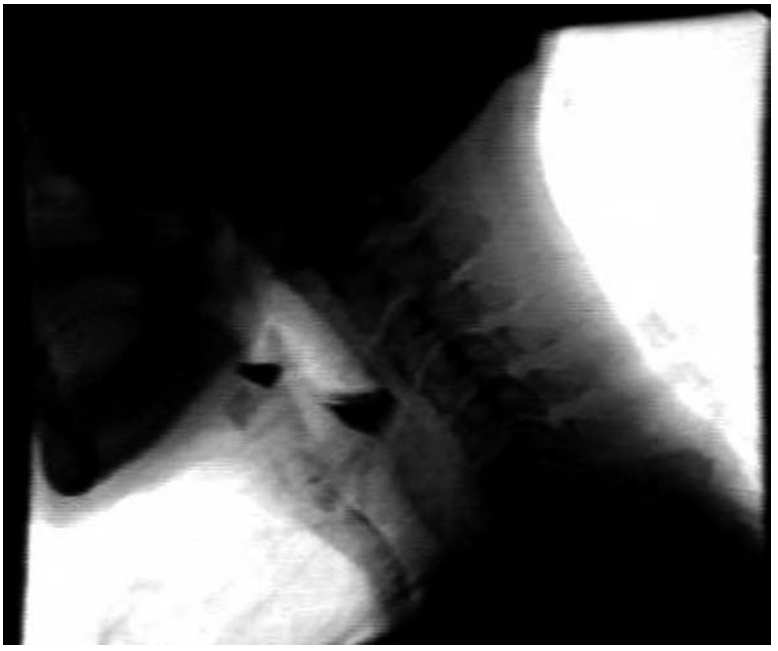

FIGURE 5.2 Lateral view of a patient during VFSE. The patient is swallowing thin liquid barium and demonstrates aspiration during the swallow, as well as postswallow residue in the vallecular and pyriform sinus.

The first-line treatment for cervical dystonia is local injection of botulinum toxin to the muscles causing the abnormal posture. Surgical treatments include bilateral C1–C3 rhizotomy, selective peripheral denervation, and, more recently, deep brain stimulation (DBS) of the globus pallidus pars interna (GPi). These treatments have been reported to have differing effects on swallowing function. While botulimum toxin injections may improve posture for safe swallowing in patients with cervical dystonia, the onset of dysphagia or an increase in its severity has also been reported (9,10). Slawek and colleagues (10) reported that dysphagia was the most common side effect of this treatment, with pain during swallowing (odynophagia) constituting the third most reported side effect. On average, dysphagia occurred 8 days after injection and persisted for approximately 15 days. Deterioration in swallow function status postinjection may be due to poor target selection, injection of unaffected muscles, medication diffusion, and other factors (16).

Horner et al. (4) found that dysphagia increased following rhizotomy in 41 patients with cervical dystonia. VFSE revealed dysphagia signs in 68% of patients prior to rhizotomy, which were considered mild in severity. Following surgery, 95% of patients demonstrated dysphagia signs that were more severe in nature. Noted swallowing signs included abnormal bolus preparation, vallecular residue, and pharyngocele (pharyngeal outpouching). Aspiration was noted more often postsurgery and swallow abnormalities were most common in the pharyngeal stage than in the oral stage. Disease duration and subjective complaints of dysphagia were found to correlate significantly with swallowing dysfunction. A subjective improvement in swallowing was reported by most patients 4–24 weeks after surgery.

Munchau and colleagues (3) studied 12 patients undergoing selective peripheral denervation surgery following failed botulinum toxin treatment. A blinded SLP and radiologist jointly evaluated two aspects of swallowing on a four-point ordinal scale for (1) bolus preparation/mastication and (2) initiation of swallow. The authors reported no significant changes in videofluoroscopic scores pre- and postsurgery. However, worsening of pharyngeal swallow delay and oral bolus preparation occurred in four participants following surgery. Interestingly, tongue base retraction was reported to improve in seven patients. The authors concluded that selective peripheral denervation surgery did not significantly compromise swallow function.

Kiss and colleagues (17) conducted a multicenter trial using bilateral DBS of the GPi in patients with cervical dystonia. Speech and swallowing were influenced

in a stimulation-dependent manner in some patients. More specifically, one case of dysarthria postoperatively that resolved with a change in stimulation parameters was reported. Another patient developed dysphagia a few weeks postsurgery that resolved with a decrease in stimulation amplitude. The authors also describe two patients who developed mild swallow impairments, while two others had improved swallowing. Thus, it appears that DBS treatment for dystonia may influence speech and swallow function in patients with cervical dystonia, though further systematic work is needed in this area.

Oromandibular Dystonia

OMD involves the masticatory, facial, and lingual musculature in various combinations and forms. OMD may, for example, lead to abnormal tongue movements (e.g., lingual protrusion dystonia) or repetitive and at times sustained jaw opening, closure, and deviation. OMD often has a negative influence on swallowing. For example, lingual protrusion dystonia may result in the anterior loss of food from the mouth. Jaw dystonia, depending on its presentation, may make it difficult to place food in the mouth (in the case of a jaw closure dystonia) or to normally manipulate food and liquid during the oral stage (in the case of jaw opening or deviating dystonia). Swallow deficits will depend on what specific structures are involved (i.e., the jaw, tongue, lower face, or a combination of these structures). Several investigators have reported on swallow characteristics in this group.

Schneider and colleagues (15) described eight patients with OMD with intermittent or continuous tongue protrusion as well as jaw closure dystonia in some. These patients experienced difficulties with swallowing, choking, speaking, tongue biting, and drooling. Several patients could only consume small amounts of a pureed diet, with weight loss necessitating a gastrostomy tube in one individual. Two individuals experienced life-threatening episodes of choking during a tongue protrusion episode, and a history of aspiration pneumonia was reported in another. The most successful treatments were botulinum toxin injections into the genioglossus muscle to control tongue protrusion and both masseter muscles in the patients who additionally had jaw closure dystonia. Two patients underwent bilateral GPi DBS that relieved tongue protrusion dystonia and resulted in marked improvement of swallowing function.

Papapetropolous and Singer (14) reported eating dysfunction in 16% of patients with OMD. Signs and symptoms included decreased bolus preparation and propulsion due to involuntary jaw opening and tongue thrusting, weight

loss, painful chewing (requiring a pureed diet), and an inability to get food into the mouth, necessitating a liquid only diet through a straw due to jaw closure dystonia. Tetrabenazine combined with other antidystonic medications and/or botulinum toxin injections was considered by the authors to be beneficial to those affected.

When OMD is accompanied by blepharospasm, the disorder is often referred to as Meige's syndrome. Mascia and colleagues (12) investigated mastication abilities in seven patients using surface electromyography (EMG) of the masseter and orbicularis oris muscles. In comparison to age-matched controls, patients with Meige's presented with excess duration of muscle activity, loss of rhythmicity, increased muscle coactivation, and disturbances in the chew to swallow transition phase.

Also using surface EMG, Bakke and colleagues (18) investigated chewing patterns across neurologic disease types and reported abnormal chewing patterns in four patients with dystonia. Specifically, two patients with primary focal dystonia of the jaw demonstrated involuntary "munching" (chewing-like movements with no food in the mouth), and two patients (one with general dystonia and the other with OMD) had abnormal mastication described as an abrupt, slow chewing rhythm that was due to repeated disturbances in either the opening or closing jaw phase. Per results of a self-report pain questionnaire, the only patients to report pain while chewing were those with a primary focal dystonia.

Medical treatment via muscle afferent block (injection of affected muscle with lidocaine and ethanol) in jaw deviation dystonia has been successful, with reports of abolished jaw deviations and significant improvements in speech and mastication (19).

Evaluation of Swallowing

Regarding swallowing, a SLP's scope of practice generally entails assessment and treatment of the oral and pharyngeal phases (or oropharyngeal function), while gastroenterologists, radiologists, and other physicians typically assess and manage esophageal disorders. An interdisciplinary team of healthcare professionals, including physicians, surgeons, occupational therapists, physical therapists, dieticians, and nurses, is most effective in the evaluation and management of dysphagia in patients with dystonia.

Evaluation of oropharyngeal swallowing typically begins with a clinical swallowing exam (CSE), which minimally includes the following components:

1. A history
2. An oral motor examination, often with sensory testing
3. A physical examination to assess items such as voice quality, strength of cough, and palpation of hyolaryngeal movement with swallowing
4. Observation of how foods and liquids are swallowed

For a comprehensive description on procedures of the swallowing examination in patients with movement disorders, the reader is directed to the book-length discussion by Rosenbek and Jones (20). The following provides special observations and procedural modifications that might be valuable in the evaluation of swallowing in people with dystonia.

Clinical Swallow Exam

The CSE is a powerful tool for swallowing assessment in the hands of a skilled dysphagia clinician. Commencing with patient history, information gleaned during the CSE helps to focus subsequent examinations and provide potentially crucial information for treatment planning. Items deserving particular attention during history taking include the following:

1. Relationship of dysphagia symptoms to medical and surgical treatments (e.g., botulinum toxin injections)
2. Present or past history of unintended weight loss
3. Benefit (or lack thereof) of sensory tricks during meals (e.g., light touch to chin or lips)
4. Rate of eating and drinking
5. Influence of control of the upper extremities, trunk, and head on eating
6. Preferences for liquids versus solids
7. Determination of whether dysphagia is exacerbated or elicited by eating, as some dystonic conditions may only occur with particular behaviors (i.e., task-specific dystonias)
8. Awareness that dysphagia may be present on instrumental exam, even when symptoms are not reported

While observing the patient eat and drink during the CSE, the following should be noted in the patient with dystonia:

1. Ability to handle utensils and glassware
2. Ability to get food into mouth (particularly when jaw closure dystonia is present)

3. Ability to keep food in the mouth (particularly when jaw opening or lingual dystonia is present)
4. Pace of eating
5. Posture
6. Use and benefit of sensory tricks
7. Coordination of swallowing and respiration (i.e., observation of the pattern of respiration with and without swallowing)

Instrumental assessment techniques may also be necessary, such as VFSE and/or endoscopic evaluation of swallowing. These approaches allow the skilled dysphagia clinician to assess the integrity of the oropharyngeal swallowing mechanism, determine the presence of penetration and aspiration, establish or at least develop hypotheses about the biomechanical abnormalities causing dysphagia, and complement other CSE findings to assist in decisions regarding oral intake, therapeutic intervention, and consultations with other healthcare providers.

VFSE

The VFSE is a radiologic procedure allowing visualization of a radiopaque substance during swallowing. Movements of anatomic structures, bolus transportation, and trials of specific strategies to improve swallow function are evaluated. During this procedure, particular attention should be paid to the following:

1. When possible, approximate the patient's normal eating posture during the examination and compare it with swallowing with improved posture.
2. Further explore the effect of sensory tricks on swallowing.
3. Careful attention should be focused on the movement of all structures involved in swallowing, even when those structures are not thought to be involved. Such careful attention is warranted in all cases, but perhaps especially so in patients with dystonia, as it is not uncommon for the distribution of affected musculature to be more widespread than suggested by a patient's medical diagnosis.
4. The penetration-aspiration scale (PAS) (21) should be used to quantitatively measure the depth of airway entry and if any attempts are made to eject material. This scale is provided in Table 5.2.

TABLE 5.2 The Penetration-Aspiration Scale (PAS)

PA SCORE	DESCRIPTION
1	Material does not enter the airway
2	Material enters the airway, remains above the vocal folds, and is ejected from the airway
3	Material enters the airway, remains above the vocal folds, and is not ejected from the airway
4	Material enters the airway, contacts the vocal folds, and is ejected from the airway
5	Material enters the airway, contacts the vocal folds, and is not ejected from the airway
6	Material enters the airway, passes below the vocal folds, and is ejected into the larynx or out of the airway
7	Material enters the airway, passes below the vocal folds, and is not ejected form the trachea despite effort
8	Material enters the airway, passes below the vocal folds, and no effort is made to eject

Source: Rosenbek JC, Robbins JA, Roecker EB, Coyle JL, Wood JL. A penetration-aspiration scale. Dysphagia 1996;11:94.

Endoscopic Swallowing Examination

Another increasingly used method for assessing swallow function is the endoscopic swallowing examination. During this procedure a small-diameter endoscope is passed through the nose into the pharynx, affording direct visualization of many of the structures of the larynx and pharynx involved in swallowing. There is little, if any, reported use of endoscopy to evaluate swallowing function in dystonia. In our own clinical practices, we generally find VFSE to be more useful in dysphagia assessment in patients with hyperkinetic movement disorders such as dystonia. At the very least, endoscopic swallowing evaluation presents with physical challenges for both the patient and the clinician due to the abnormal, twisting postures exhibited in this condition. However, in some patient populations, such as patients with diagnosed or suspected laryngeal dystonia, the endoscopic swallowing exam may have particular value. Reports of the use of the endoscopic swallowing examination in patients with dystonia will be a valuable contribution to the literature on swallow function in this population.

Treatment Considerations

When swallowing is unsafe, inadequate to maintain hydration and nutrition, or requires more effort than the patient can tolerate, a variety of behavioral

treatments should be considered (22). These behavioral treatments can be classified as rehabilitative or compensatory approaches. A summary of both rehabilitative and compensatory strategies is provided in Tables 5.3 and 5.4, respectively. These tables provide the clinician with an array of treatment options that need to be specifically tailored to the swallow signs and symptoms of individual patients. It is important to note that some rehabilitative maneuvers will be too difficult, inappropriate, or maladaptive for the patient with dystonia. For example, the Showa and Masako maneuvers require prolonged muscle holds with the tongue that may be impossible for a patient with lingual dystonia to perform. These are, however, included in Table 5.3 to provide a more complete list of potential treatment options.

TABLE 5.3 Rehabilitative Swallow Treatment Strategies

STRATEGY	DESCRIPTION	REPORTED EFFECT ON SWALLOW
Supraglottic swallow	Involves forceful laryngeal adduction followed by a throat clear/cough and a repeated swallow	Improve airway protection
Mendelsohn maneuver	Patient holds larynx in most anterior-superior position for 1–3 seconds, followed by completion of the swallow	Prolong duration and extent of UES opening
Shaker head raise	Patient lies supine and repeatedly raises and lowers head	Increase duration and extent of UES opening
Expiratory muscle strength training	Patient blows into a one-way spring loaded valve against a pre-set resistance	Improves submental muscle contractile properties, cough function and peripheral and central neural control mechanisms during swallowing (23)
Showa maneuver	Requires forceful elevation of tongue against the hard palate followed by a long hard swallow during which the patient squeezes all the muscles of the face and neck	Improves oral and pharyngeal movements during swallow (24)
Masako maneuver	Protruding and holding the tongue out while performing a dry swallow	Increase posterior pharyngeal wall movement and decrease residue (25)
Sensory therapies	Stimulation with cold, sour, and electrical current	May improve oral and pharyngeal stage function (26,27)

UES, upper esophageal sphincter.

TABLE 5.4 Compensatory Swallow Treatment Strategies

STRATEGY	EFFECT ON SWALLOW
Postural stabilization	Reduce movement or asymmetry
Liquid wash (alternate bite/sip)	Reduce postswallow residue
Double swallow	Reduce postswallow residue
Throat clear/cough postswallow	Clear airway postswallow
Chin tuck	Postural adjustment that will aid airway protection
Head turn	Postural adjustment that may aid bolus transit and reduce residue
Dietary changes	Avoid troublesome textures, switch to softer foods or in severe instances a pureed diet
Use of adaptive feeding utensils	Will aid in the hand to mouth movement of the preswallow period
Cut food into small pieces/small sips of liquid	Decrease effort required and allow more manageable bolus size
Decrease eating rate	Allow more time and decrease demands on swallow system
Time meals appropriately	To coincide with maximal medication effects and times of day when patient is most alert

Due to difficulties in performing many of the rehabilitative maneuvers listed in Table 5.3, as well as the pathophysiology of dystonia, rehabilitative treatments represent a challenge for clinicians working with this population. In addition, limited data are available to provide guidance. We have generally found that compensatory approaches are most valuable in patients with dystonia. For example, postural adjustments, when possible, may provide benefit. Sensory tricks, such as a light touch by the patient to the chin, may also be helpful in providing temporary relief from dystonia.

By the time patients see the SLP for swallowing evaluation, they may have made some adjustments in what and how they eat. It has been reported, for example, that patients with cervical dystonia cut food into smaller pieces, drink with their meals, and perform special head and neck maneuvers while swallowing (3,7).

Compensatory techniques (Table 5.4) need to be tailored to specific patients and their swallowing signs and symptoms. For example, in a patient with a jaw dystonia characterized by severe involuntary jaw closure dystonia,

getting food into the mouth may present a problem, with significant health consequences. Use of a straw may be beneficial in these instances for oral liquid intake.

Other patients with impaired mastication abilities from OMD may benefit from a pureed diet. If rehabilitative and compensatory strategies are ineffective and if safe, enjoyable, and adequate nutrition and hydration are impossible, enteral nutrition may become necessary.

Quality of Life

The impact of dysphagia on quality of life (QOL) in patients with dystonia has received little attention in the literature, though Papapetropoulos and Singer (14) report social embarrassment with eating in patients with OMD. In our clinical experience, patients with dystonia and dysphagia may report other related QOL influences, including slowness of eating; decreased enjoyment of eating and drinking; "messy" eating, and a reluctance to eat in public.

Nutrition

Dysphagia in the patient with dystonia can have a significant impact on nutritional status. It is recommended that the SLP work closely with a registered dietician to ensure the patient is obtaining adequate oral intake to meet nutritional requirements. Failure to do this can result in an array of macronutrient and micronutrient deficiencies and protein-calorie malnutrition (28). The importance of adequate nutrition care cannot be underestimated in this population. Early dietary intervention can help to maintain strength, functional status, immune function, and nutritional status (28). Weight loss, a common sequela in patients with dystonia and dysphagia (15), is one of the first signs of nutritional compromise. When possible, a nutritional screening should be conducted by a dietician and, if a patient is identified to be at risk for malnutrition, a full assessment implemented. Calorie, protein, and fluid requirements should be determined. If the dysphagic patient fails to meet these requirements via oral intake, a number of strategies may be employed. First, every effort to obtain nutritional requirements via oral intake should be attempted. This might include dietary changes such as calorie- and protein-rich foods and liquids. In the case of a patient with a severe jaw closing dystonia that restricts him or her to a full liquid diet through a straw or the patient with OMD with

masticatory deficits, implementation of calorie- and protein-rich drinks or shakes such as Boost© or Ensure© to increase caloric intake may be recommended. In some cases enteral feeding, either alone or in combination with an oral diet, may be needed to maintain adequate nutrition. In the most severe case, enteral feeding alone may be indicated, though this should be avoided whenever possible.

The Speech System

Functional Components of the Speech System

Although not often appreciated when functioning appropriately, normal speech is a complex sensorimotor process that involves all levels of the nervous system and a distributed network of structures, pathways, and musculature (29). In addition to being a sensorimotor process, speech production is dependent on a variety of cognitive, linguistic, and sensory mechanisms. Speech is commonly considered in terms of what have come to be known as the "functional components" of the speech mechanism. These components traditionally include the respiratory mechanism, larynx, pharynx, velopharynx, and orofacial structures of the jaw, face, and tongue (30). When nervous system support for any of these functional components is disrupted by neurologic disease, abnormal speech or, more precisely, dysarthria, is the result. Duffy (29) defines dysarthria as "a collective name for a group of neurologic speech disorders resulting from abnormalities in the strength, speed, range, steadiness, tone, or accuracy of movements required" for control of the functional components of the speech mechanism. Additionally, it is traditional and often helpful clinically to identify the speech processes of respiration, phonation, resonation, and articulation. To this list is often added prosody, which minimally includes perceptual features such rate, intonation, stress, and rhythm. Naturalness and intelligibility of speech are also being measured more commonly and with greater sophistication.

Dysarthria in Dystonia

When dystonia involves the musculature of the speech mechanism (including the respiratory mechanism), dysarthria can occur. The specific form of dysarthria occurring in dystonia is commonly described as a hyperkinetic

TABLE 5.5 Deviant Speech Auditory Perceptual-Speech Characteristics Reported in the Hyperkinetic Dysarthria of Dystonia Listed on Order of Severity (31)

Imprecise consonants
Distorted vowels*
Harsh voice quality*
Irregular articulatory breakdowns*
Strained-strangled quality*
Monopitch
Monoloudness
Inappropriate silences*
Short phrases
Prolonged intervals
Prolonged phonemes
Excess loudness variations*
Reduced stress
Voice stoppages*
Slow rate

*Distinctive feature of hyperkinetic dysarthria in dystonia or those more severely impaired than in other dysarthria.
Source: Ref. 31.

dysarthria. Darley et al. (31) performed the seminal investigation into the various dysarthria types and described the distinguishing auditory-perceptual features of hyperkinetic dysarthria in dystonia as listed in Table 5.5.

A discussion of how dystonia can affect each of the components of the speech mechanism follows.

Respiratory Mechanism

Although Darley et al. (31) did not directly discuss the respiratory mechanism in hyperkinetic dysarthria, the inclusion of *excess vocal loudness* as an auditory-perceptual feature and *alternating loudness* as a distinguishable feature from other dysarthria types suggests some abnormal respiratory function. Since this time, other investigators have documented respiratory involvement in dystonia.

LaBlance and Rutherford (32) compared respiratory dynamics in six patients with generalized dystonia with controls. They noted that patients with dystonia had faster respiratory rates, less rhythmic breathing patterns, decreased

lung volume, and apnea-like periods with decreases in arterial blood flow during quiet breathing and monologue. Speech intelligibility was seen to be strongly related to respiratory function. Specifically, speech was seen to be less intelligible when there were faster breathing rates, variability in respiratory cycle duration, variability in arterial blood oxygen saturation, and reduction in inspiratory volume during monologue. This study suggests the importance of respiratory function to speech intelligibility and highlights the respiratory mechanism as a therapeutic target for improving speech intelligibility in these patients.

Larynx

Laryngeal deficits observed in Darley et al.'s (31) group of dystonic speakers included *harshness, strained-strangled voice quality, excess loudness variations,* and *voice stoppages*. Combined, these speech dimensions formed what Darley et al. referred to as the cluster of "phonatory stenosis" thought to be related to hyperadduction of the vocal folds during phonation (29). In addition to these distinguishing perceptual features of the disease, the presence of *voice tremor* was also reported. Since this work, others have noted similar findings or added to knowledge regarding laryngeal disturbances in patients with dysarthria and dystonia.

Acoustic analysis of speech has revealed that persons with cervical dystonia have lower habitual fundamental frequency, lower ceiling fundamental frequency, restricted frequency range, shorter /s/ and /z/ durations, shorter maximum phonation duration, slower sequential movement rates, slower alternative movement rates, longer phonatory reaction time, slower reading rate, decreased intelligibility, increased jitter and shimmer values, and decreased harmonic-to-noise ratio (33).

Zraick and LaPointe (34) note that a distinguishing feature of hyperkinetic dysarthria is the variable, fluctuating, and unpredictable nature of speech. This is likely the result of abnormal patterns of vocal fold vibration (e.g., hypoadduction, hyperadduction, or fluctuating adduction).

Finally, spasmodic dysphonia (SD), which comprises 9% of all dystonia types (35), is a focal laryngeal dystonia with prominent effects on speech. SD can be classified as the adductor, abductor, or mixed form. Adductor SD is characterized by irregular hyperadduction of the vocal folds leading to strained, strangled, and effortful speech with frequent stops in phonation (36–38). Abductor SD is characterized by laryngospasms of the posterior cricoaryteniod muscles (adductor muscles), resulting in breathy, aphonic, or whispering voice

with abrupt terminations of voicing as the vocal folds are abducted during phonation. Some patients have a mixed SD that is characterized by both intermittent strained and breathy qualities (29). Adductor SD is the most common type of SD, accounting for a reported 82–89% of laryngeal dystonia (36,39).

Velopharynx

Disturbances of velopharyngeal function during speech are not commonly associated with hyperkinetic dysarthria. When encountered, intermittent hypernasality is the most common perceptual feature exhibited. Indeed, Duffy (29) reports intermittent hypernasality to be a distinguishing feature of hyperkinetic dysarthria from other dysarthria types. Resonance abnormalities may be expected most commonly when velopharyngeal dystonia is observed, though this can probably not be adequately determined with visual observation of the velum during nonspeech tasks, as dystonia is variable during different tasks and conditions, as is velopharyngeal function. As an example of the latter, velopharyngeal function in disordered populations may be different in its adequacy during speech and swallowing (31,40).

Orofacial Mechanism

Darley et al. (31,40) observed several prominent perceptual features related to disordered function of the orofacial mechanism, such as *imprecise consonants, distorted vowels,* and *irregular articulatory breakdown*. Together these aberrant speech features form the cluster termed *articulatory inaccuracy*. This subsystem is largely influenced by respiratory, laryngeal, and velopharyngeal function, and disturbances in these systems can be expected to affect articulatory proficiency and overall speech intelligibility (34).

Speech in OMD has also been described as having imprecise consonants (41,42). Individuals with OMD are likely to have disturbed orofacial performance due to the lack of control and involuntary movement of the structures of this mechanism such as the tongue, lips, and jaw. Other dystonias that affect such structures may be expected to impair articulatory function.

Prosody

Darley et al. (23) noted the following prosodic abnormalities in dystonic speakers: *monopitch, monoloudness, short phrases*, and *reduced stress*. These dimensions combined to form the cluster *prosodic insufficiency*. More recently, LaBlance and Rutherford (32) noted lower pitch and a restricted pitch range in

six patients with cervical dystonia, while abnormalities in stress have been reported in individuals with OMD (41,42).

Slowness of movement and interruptions in the flow of speech movements commonly observed in dystonia can lead to other aberrant prosodic features of such as prolonged intervals, prolonged phonemes, and slow rate, which Darley et al. referred to as "prosodic excess" (29). The prosodic subsystem is likely affected by reduction in range of movement, rigidity, and slowness of movement.

Examination of Speech

The traditional speech exam consists of four substantive parts:

1. History
2. Oral mechanism exam
3. Maximum performance testing
4. Evaluation of the speech mechanism during speech tasks—most importantly during connected speech

In some instances additional instrumental examination such as acoustic analysis, videostroboscopy, nasoendoscopy, electromyography, or kinematic analyses may be needed to supplement the traditional exam. For example, when examining a patient with laryngeal dystonia, videostroboscopy or endoscopic exam may complement the traditional speech exam by allowing direct visualization of the vocal folds during speech and phonation and at rest.

Table 5.6 provides an overview of the key components of the traditional motor speech evaluation. Though the use of nonspeech or speech-like activities has become a controversial topic (see Ref. 43 for a review), in our clinical experience such an exam is generally a valuable starting point. Duffy (29) and Yorkston et al. (44) are valuable resources for additional information. The following provides special observations and procedural modifications the SLP should consider when performing the motor speech evaluation in a patient with dystonia.

History

While taking the history, the clinician is able to observe connected speech while obtaining the necessary information. The clinician should pay attention to the use of any compensatory strategies such as postural shifts or sensory tricks.

TABLE 5.6 Key Components of a Traditional Motor Speech Exam

HISTORY OF THE SPEECH PROBLEM	EXAMINATION OF THE SPEECH MECHANISM WITH NONSPEECH ACTIVITIES	MAXIMUM PERFORMANCE TESTING OF THE SPEECH MECHANISM	EVALUATION OF THE SPEECH MECHANISM WITH SPEECH TASKS
• Onset and course Insidious or acute, fluctuations over time, effects of medication • Associated deficits Difficulty with swallowing, cognition, language, and/or changes in affect or emotions, physical function • Patient perception Patient describes change in speech and strategies to improve speech • Consequences of speech disorder Changes in ability to participate in vocational or social activities • Overall health care Other professionals involved, services provided, current medications, utilization of community resources	• Respiratory mechanism - Observe for posture, breathing at rest, and with physical exertion. - Elicitation of brisk sniff and rapid pant determines strength and coordination of the respiratory mechanism • Larynx -Laryngeal integrity assessed by eliciting volitional coughs and grunts. -Direct visualization of the larynx via flexible fiberoptic endsoscopy or rigid oral laryngoscopy may be necessary in some patients. • Velopharynx (VP) -Evaluate VP at rest for symmetry, involuntary movements, or structural abnormalities	• Respiratory mechanism -Assess range of loudness and maximum loudness during phonation. -Measure maximum phonation duration • Larynx -Assess vocal quality during 3 seconds of the optimal phonation. -Assess pitch range with pitch glide from lowest to highest pitch • Velopharynx (VP) -Assess resonance during assimilative nasality task ("Make me a Hong Kong cookie") and during production of a standard sentence ("Buy Bobby a poppy") with the nares open and occluded.	• Connected speech -Considered the most critical part of the evaluation. -Used to determine how components of an individual's speech mechanism work together. -Used to assess speech characteristics including rate, intonation, stress, rhythm, and naturalness. -Elicited during history or conversation (i.e., "Tell me about your family") • Repeating words/ sentences: 1. Snowball 2. Impossibility 3. Catastrophe 4. Please put the groceries in the refrigerator 5. The valuable watch was missing 6. The shipwreck washed up on the shore

	• Orofacial mechanism (face/lips, jaw, tongue) -Determines symmetry, strength, range of motion, and coordination -Observe for involuntary movements, structural abnormalities, and abnormal posturing	• Orofacial mechanism -Assess alternating motion rate by instructing patients to repeat "puh," "tuh," "kuh" as quickly, precisely, and regularly as they are able. -To isolate the tongue for "kuh," have patient put thumb between teeth, and bite down lightly	• Reading a standard passage -Use a passage with known number of words, frequency of sounds, and established rate norms, such as the Grandfather Passage

Duffy (29); Yorkston et al. (44); Kent et al. (45); Wertz et al. (46).

Source: Adapted from Jones HN, Donovan NJ, Rosenbek JC. Speech and swallowing disorders in patients with movement disorders. In: Fernandez HH, Rodriguez RL, Skidmore FM, Foote KD, Okun MS, eds. A Practical Approach to Movement Disorders: Diagnosis and Medical and Surgical Management. New York: Demos Medical Publishing, 2007:205–236.

The clinician should probe the following areas:

- If use of sensory tricks is noted, determine if the patient is aware of such strategies.
- If sensory tricks are not observed, ask the patient if they have ever tried such techniques, their effect, and when they might be used.
- How does speech change in different environments and during different tasks (e.g., reading, singing, conversation).
- Obtain specific information regarding medical treatments (e.g., botulinum toxin or surgical intervention) and their effect on speech.
- Estimate intelligibility across different environments based on patient and family report.

Oral Mechanism Exam

The oral mechanism of the dystonia patient is often normal in size, strength, and symmetry (29). The striking feature of the exam is at rest or while a patient attempts to maintain steady facial postures. Blepharospasm or facial grimacing may be present, as well as intermittent, relatively sustained dystonic posturing of the mouth, tongue, and jaw. In some patients, however, these involuntary movements are triggered by speech, leading to a normal oral mechanism exam.

Maximum Performance Testing

Early studies performed by Case et al. (47) and Zraick and colleagues (48) reported the following acoustic speech features as significantly differing in patients with cervical dystonia during maximum performance testing:

- Shorter maximum vowel prolongation durations
- Slower sequential movement rates (SMRs)
- Slower and irregular alternate motion rates (AMRs)
- Shorter /s/ durations
- Shorter /z/ durations

Auditory-Perceptual Assessment During Speech

It is important to assess speech in a variety of contexts. These include: conversation, reading, speech alternative motion rate, and vowel prolongations. Careful visual inspection of speech is particularly important in the patient with dystonia.

Treatment Considerations

A thorough examination can assist in the development of a sensible treatment plan. The primary goal in the management of these patients should be to maximize the effectiveness, efficiency, intelligibility, and naturalness of communication (29). Hyperkinetic dysarthria may be managed medically, surgically, and behaviorally; these treatment options will be outlined below.

Medical and Surgical Management of Hyperkinetic Dysarthria

Botulinum toxin is currently regarded as the most effective medical treatment for speech improvement in individuals with SD (35,36,39). Biltzer et al. (39) reported an average benefit of 90% of normal function lasting an average of 15 weeks in adductor SD and an average 67% benefit lasting 11 weeks in abductor SD patients in 900 patients with SD. Poorer outcomes with botulinum toxin are associated with older age and abductor SD (36). In addition to its use with focal laryngeal dystonia, botulinum toxin has been reported to be effective in treating tongue protrusion dystonia and jaw closure dystonia via injection into the genioglossus or masseter muscles, respectively (15).

DBS may also be used as a treatment for dystonia and has been reported to have differential effects on speech function. While some investigators have described improvements in speech function following DBS (15), others have reported dysarthria to be an adverse event following surgery (17,49). Indeed, Kupsch and colleagues (49) reported dysarthria to be the most common side effect post-DBS, occurring in 12% of patients, though the authors indicated this was typically resolved with adjustments of stimulation settings. The literature on the effects of DBS on speech function is only emerging and warrants further systematic study.

Behavioral Management

A number of behavioral treatment strategies can be employed by the SLP when working with the patient with dystonia. It is important that treatment goals be discussed with the patient and his or her family members, with the overall goal centered on maximizing intelligibility and naturalness of speech.

The application of a sensory trick maneuver ("geste antagoniste") may reduce agonistic and antagonistic muscle activity and lead to behavioral improvements in patients with dystonia (50). It is reported that 54–73% of

cervical dystonia patients use such tricks to surpass dystonic drive (51–53). The most common sensory trick is a slight touch to the chin or cheek, usually ipsilateral to the direction of head rotation (51,53).

Sensory stimulation to the affected body part(s) has been documented via EMG to decrease activity of the involved musculature. For example, Schramm et al. (50) studied 26 patients with cervical dystonia. The 19 patients who used a beneficial sensory trick demonstrated reduced EMG activity when using this maneuver. Sensory tricks in this group of patients included touching the chin, neck, or check with a finger or plastic stick. An individual's primary head position was found to have a strong influence on the benefit of the sensory trick. Interestingly, mere imagination of performing these maneuvers also led to a reduction in EMG activity (albeit decreased in amplitude), suggesting the involvement of a higher level of sensorimotor integration during the preparation phase of movement (50).

In OMD, effective sensory tricks include touching the lips, chin, or submental region, chewing, or biting (54). Baik and colleagues (55) described a case of focal lingual dystonia specifically induced by speaking. The patient had dysarthria due to uncontrolled tongue protrusions and used sensory tricks as a compensation, including chewing gum when she spoke on the telephone. Her condition eventually resolved after a 2-year treatment with trihexyphenidyl.

The use of a bite block, a custom-fitted prosthesis placed between the upper and lower teeth, has also been reported to be beneficial in patients with OMD to help inhibit jaw movements during speech (22). Schramm et al. (56) studied the effects of placing a wooden stick between the teeth and cheek or biting slightly on the stick while counting in seven patients with jaw-opening dystonia. The authors reported an improvement in orofacial function based on a blinded perceptual analysis, improved patient perception based on self-rating scales, and a reduction in EMG muscle activity of the temporalis, orbicularis oris, and digastric musclulature. Schramm et al. (56) suggested that the bite block provided both tactile stimulation and jaw occlusion and concluded that altered somatosensory feedback and voluntary antagonistic activation can modify symptoms of OMD. The benefit of such an approach can often be quickly explored by the SLP in the clinical setting by having the patient perform various speech tasks with and without a tongue depressor between the teeth.

Some investigators (29,32) have reported the utility of using biofeedback to modify lip dystonia or respiratory involvement in generalized dystonia that

lead to improved speech intelligibility. While more data supporting its benefit are needed, this could be a treatment approach for some patients.

Practical Tips

1. Be aware of the high incidence of subclinical dysphagia in the dystonia patient.
2. In our experience, compensatory techniques are often most beneficial in the treatment of swallowing and speech disorders in dystonia. Table 5.4 provides a list of compensations to consider for patients with dysphagia and dystonia.
3. In patients with dysphagia, consultation with a dietician may be helpful to optimize health and nutritional status.
4. Use of sensory tricks (chewing gum, postural adjustments, light touch) can be effective in reducing dystonic symptoms during speech and swallowing and should be explored if patients have not already done so on their own.
5. Dystonia is often more distributed than suggested by the medical diagnosis, and structures involved in swallowing and speech require careful assessment.
6. The influence of medical and surgical treatments on swallowing and speech is not completely understood. However, changes in swallowing and speech should be monitored, and positive, negative, or neutral outcomes may be encountered.
7. Determination of whether dysphagia is exacerbated or elicited by eating should be attempted.
8. Use of a bite block can be beneficial in improving speech intelligibility in patients with dystonia, and its benefit should be explored during the speech exam when the orofacial mechanism is involved.
9. Consider biofeedback approaches during treatment (e.g., EMG).
10. Many patients with dystonia will benefit from an interdisciplinary treatment approach involving professionals such as physicians, surgeons, SLPs, dieticians, physical therapists, and occupational therapists.

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6

The Role of Physical Therapy in the Management of Dystonia

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Barbara Bour

Dystonia is defined as a neurologic syndrome characterized by involuntary, patterned, sustained, or repetitive muscle co-contractions, which cause twisting movements or abnormal postures (1). Dystonia can be considered primary, secondary, or paroxysmal (2). Primary dystonia is considered idiopathic in nature with no evidence of a secondary cause for the symptoms. Secondary dystonia may be caused by abnormal birth or developmental history, exposure to certain drugs, neurologic illness, or other cause (3). Dystonia can be further subclassified as focal, segmental, multifocal, generalized, or hemidystonia depending on the region of the body or parts affected. Many focal dystonias, particularly those that begin early in life, will progresses to a more severe generalized form. Disease that begins in the third or fourth decade of life may involve the craniocervical muscles and may even remain focal or segmental (4).

Management of disability that results from dystonia is not straightforward and usually involves a multidisciplinary or interdisciplinary approach (Figure 6.1). This chapter will focus on the physical therapy management strategies to address disability and impairment of normal movement as a result of dystonia. The role of the physical therapist needs to be dynamic and must consider the needs of patients, especially if their condition is progressive. Despite the availability of tools to classify the type and severity of dystonia, each patient must be independently evaluated, as they will not often present with identical muscular imbalances or postural deviations (5). It is this diversity of presentations to the physical therapist that makes dystonia a challenging disorder to manage.

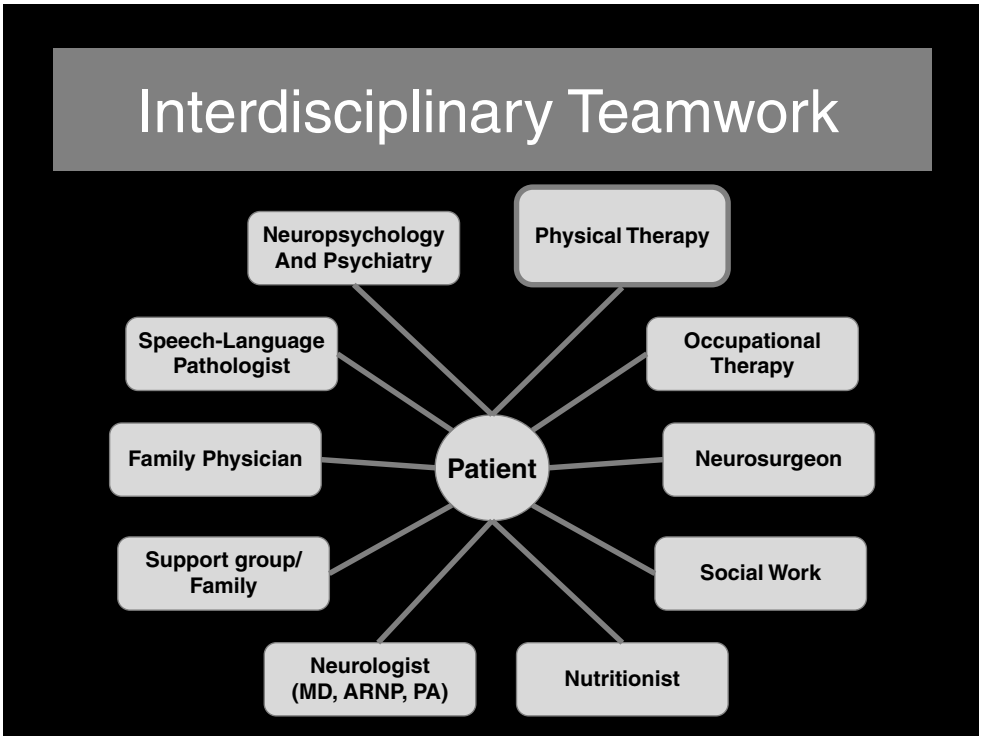

FIGURE 6.1 Effective management of dystonia requires interdisciplinary or multidisciplinary care. Physical therapy is one component of the network.

Role of the Physical Therapist

Physical therapy can offer opportunity for those living with dystonia. Dystonia presents in a variety of forms, and therapists should be aware that it can affect mobility, posture, and balance and therefore affect an individual's ability to perform everyday tasks.

Dystonia is a neurologic disorder. Physical therapy does not address the underlying dystonia mechanism but rather focuses on treating symptoms and secondary conditions that may accompany the dystonic phenomena. Through the management of these secondary conditions, therapists can create a foundation to maintain and improve function for select patients (Figure 6.2). The physical therapist may employ various treatment modalities and provide guidance to help individuals recognize and manage compensatory movements as well as habits that may have developed as a result of the dystonia. Making sufferers aware of activities that aggravate symptoms and teaching beneficial substitute methods may contribute to improved motor control and to quality

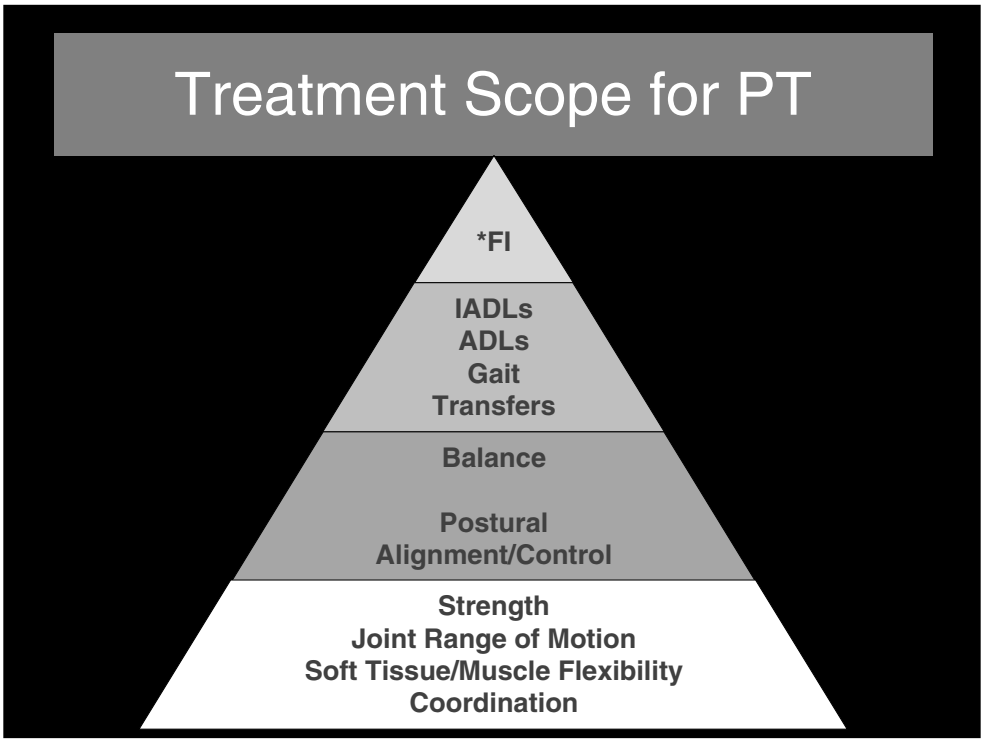

FIGURE 6.2 Developing a strong foundation can aid in improving or maintaining function in dystonia. FI, functional independence; ADLs, activities of daily living; IADLs, instrumental activities of daily living.

of life. Physical therapy may also enhance the benefits from other medical treatments, such as oral medications, botulinum toxin injections, and surgical interventions (6).

Physical therapy is a slow process that should be approached with the expectation that there will be a significant commitment in time, but with that commitment should come optimism. Results may not be immediately apparent, but a physical therapy program can influence many aspects of daily living (6).

Physical Therapy Examination

Prior to any treatment it is important for the therapist to conduct a thorough examination. This should consist of a detailed history, a review of systems, and finally specific tests and measures in order to objectify limitations and appropriately select interventions (Table 6.1). The examination process may also

TABLE 6.1 Physical Therapy Examination

EXAMINATION COMPONENT	EXAMINATION PROCEDURES	OBJECTIVE
PATIENT HISTORY	• Interview patient, family, caregiver • Review medical record • Consult with other members of the care team	• Learn patient's chief complaint and expectations • Identify needs related to restoring health, prevention, wellness, and fitness • Obtain demographics, social history, PMH/PSH, work/school/leisure status, growth and development history, living environment, social and health habits, past and current level of function, family history, medications • Results of other tests • Identify caregiver needs
SYSTEMS REVIEW	Brief or limited exam of: • Cardio-pulmonary system • Integumentary system • Musculoskeletal system • Neuromuscular system • Communication • Cognition • Language	• Gather overview of how each of these areas may be affecting the patient's ability to function • Assist in identifying problems that require referral to another provider
TESTS AND MEASURES	• Aerobic capacity/endurance • Anthropometric characteristics • Current of use of assistive/adaptive devices • Home, work, school and community environment barriers • Ergonomics and body mechanics • Gait, locomotion, and balance • Integumentary integrity • Motor function (motor control/learning—dexterity and coordination) • Muscle performance (strength, power, endurance) • Sensory integration—postural/equilibrium/righting reactions, evolution of motor skills (developmental inventories, infant and toddler motor assessments, reflex tests) • Use of orthotic, protective, or supportive devices • Pain assessment—provocation, discrimination, location, cessation. • Postural alignment	• Identify and define additional problems not uncovered during the history and systems review. • Confirm or reject hypotheses about the factors causing or contributing to the patient's diminished function • Support clinical judgments about appropriate interventions and expected outcomes

TABLE 6.1 *(continued)*

EXAMINATION COMPONENT	EXAMINATION PROCEDURES	OBJECTIVE
	• Joint range of motion and muscle length/flexibility • Sensory exam—deep reflexes, tone, sensation, vision, vestibular • ADL, IADL, and functional mobility performance • Job/School/Leisure and community integration or reintegration	

ADLs, activities of daily living; IADLs, instrumental activities of daily living; PMH, past medical history; PSH, past surgical history.
Source: Ref. 7.

identify potential issues that will require referral to another multidisciplinary/interdisciplinary provider. Additional specific considerations for evaluation are discussed later in the chapter, particularly for age (younger vs. older) and specific dystonia subtypes (7).

Management of Dystonia

A variety of tools can be utilized to classify and rate the severity of dystonia; these include the Unified Dystonia Rating Scale, Burke-Fahn-Marsden Scale, and the Global Dystonia Rating Scale (8). The selection of particular treatments is largely guided by a patient's age, the dystonia subtype (primary vs. secondary), and the anatomic distribution of the dystonia. The selection of the optimal treatment strategy by physical therapy is usually based on personal clinical experience, which is weighted against knowledge of efficacy and potential for adverse events (9). Most available treatments have not been subjected to rigorous double-blinded controlled trials. For the purposes of this chapter we will separate the management strategies into the broad categories of focal dystonia and generalized dystonia. We will discuss management ranging from onset of the disease to advanced stages. Through the use of this model we will provide an overview of the goals of physical therapy and introduce intervention strategies appropriate for each type. It is important to remember that the dystonia syndrome is really a complex disease process, and individuals will present with differing symptoms and may have differing responses to treatment. We will discuss the most efficacious interventions available and

introduce methods that have been investigated in the literature that will apply to this population of patients (9). Table 6.2 presents a summary of treatment interventions (6,10–27).

TABLE 6.2 Physical Therapy Treatment Modalities

MODALITY	TREATMENT GOAL	LIMITATIONS/PRECAUTIONS
CASTING/SPLINTING	1. Limb immobilization to remove sensory/motor inputs to the limb and to allow the cortical map to reset to normal topography (21) 2. Reduce or prevent contractures and joint deformity 3. Provide static support or sensory cue in the case of cervical dystonia	1. Immobilization can exacerbate or precipitate dystonia in a limb (18,20) 2. Skin integrity and pain as the dystonic activity may continue to push against or into the splint/cast 3. Muscle weakness/instability due to dependence on cervical brace
THERAPEUTIC EXERCISE	1. Aerobic conditioning to maintain cardiovascular fitness 2. Strengthening of postural muscles and antagonist muscles to improve alignment and decrease pain (11,15) 3. Active and passive stretching of agonist muscles to improve alignment and muscle length tension relationships 4. Maintain bone, joint, and soft tissue health	1. Excessive exercise in some instances may exacerbate limb dystonia (paroxysmal exercise induced dystonia) (27) 2. May be difficult to facilitate contraction of antagonistic muscles through range of motion during continuous agonist activity; should begin in a relaxed position and progress to functional positioning for therapeutic exercise.
MANUAL STRETCHING	1. Improve soft tissue extensibility/ flexibility 2. Reduce possible contractures of muscular, ligamentous and capsular structures from prolonged dystonic posturing (24)	1. Use caution to avoid overstretching any structure which may lead to static or dynamic joint instability 2. Stretching against dystonic muscle unable to relax
SOFT TISSUE AND JOINT MOBILIZATION	1. Promote muscle relaxation 2. Facilitate appropriate tissue extensibility to allow proper posture 3. Decrease pain	Patient may be unable to relax or "turn off" muscles during joint mobilization; techniques should be gentle and graded in nature (13)
POSTURAL TRAINING	1. Maintain/improve head- and trunk-centered coordinate systems to keep appropriate perception of head-trunk alignment 2. Increase voluntary control of head and trunk 3. Decrease pain through decreased maintenance of anomalous postures (23)	Patient compliance with cues and training at home

TABLE 6.2 (*continued*)

MODALITY	TREATMENT GOAL	LIMITATIONS/PRECAUTIONS
EMG BIOFEEDBACK (10)	1. Train patients to inhibit unwanted muscle contractions at rest and during movement 2. Facilitate improved positioning and posture 3. Decrease pain	1. Availability of EMG equipment 2. Difficult to transfer to independent program at home
TENS	1. Improve co-contraction of agonist and antagonist muscles. TENS may promote a reshaping of dysfunctional reciprocal excitatory and inhibitory muscle activity in agonists and antagonists (26) 2. Decrease pain (14)	1. Temporary treatment effect 2. Needs further investigation
NEUROMUSCULAR ELECTRICAL STIMULATION	Promote contraction of the antagonist muscle	Standard precautions for use of this modality
MUSCLE VIBRATION AND VESTIBULAR STIMULATION	Reduce activity in dystonic muscle groups to promote improved posture and decreased pain (19,22)	Needs further investigation
CONSTRAINT-INDUCED MOVEMENT THERAPY	Force use of the more affected extremity and minimize the effects of learned nonuse (25)	1. Widely used in poststroke rehabilitation with some evidence to support its use in focal hand dystonia (12) 2. May cause an increase in dystonic movement
ACTIVITY AND ENVIRONMENTAL MODIFICATION (16)	1. Avoid activity that exacerbates dystonia. 2. Reduce compensatory movements of surrounding anatomic structures 3. Use of mirrors for instant visual feedback on positioning 4. Adaptive or ergonomic modification to home, school or work environment 5. Ergonomic adaption to specific tools or instruments used regularly 6. Energy conservation 7. Fall prevention and safety	1. Requires consistent active involvement by patients, caregivers, and community 2. Finding the least restrictive environment for optimal function
GAIT AND LOCOMOTION TRAINING	1. Increase gait stability through adaptive stepping techniques and use of tactile, visual and verbal cueing strategies 2. Prescribe ambulatory assistive devices	Finding the least restrictive devices to maintain maximum independence and safety

(*continued*)

TABLE 6.2 (*continued*)

MODALITY	TREATMENT GOAL	LIMITATIONS/PRECAUTIONS
	3. Wheelchair positioning and mobility training 4. Task specific training for their environment	
ACTIVITY OF DAILY LIVING AND TRANSFER TRAINING	1. Maintain/Improve independence with bed mobility, transfers, dressing, grooming, bathing, eating, toileting 2. Promote ability for home management: shopping, chores, daregiving, yard	May require modification of the activity rather than training or use of assistive device
COMPLEMENTARY THERAPIES (FELDENKRAIS METHOD, ALEXANDER TECHNIQUE, RELAXATION TECHNIQUES, SOFT MARTIAL ARTS, YOGA, ETC.)	1. Address the whole person—body, mind, and spirit 2. Decrease physical and emotional stress that may exacerbate symptoms. 3. Increased awareness of movement during every day activity leading to higher functional ability (6,17)	Supported by anecdotal reports, but current research-based evidence cannot guide clinicians regarding the effectiveness of these techniques in this population (17)

Focal Dystonia

Primary focal dystonias occur much more frequently than generalized torsion dystonia (some estimates showing approximately 10 times more frequent) (28) This type of dystonia occurs frequently in adults. It usually affects the neck, face, or arm. The leg is rarely involved (3) Cervical dystonia or spasmodic torticollis is the most common form of focal dystonia and represents the area where a majority of investigation for physical therapy interventions has been focused to date.

Cervical Dystonia

Cervical dystonia (CD) usually begins with initial neck stiffness and restricted head mobility. Abnormal head postures may in some cases follow soon after and are sometimes asssociated with irregular head tremor. Neck and shoulder pain occur in approximately 75% of cases (3). This condition is often misdiagnosed as a musculoskeletal disorder. Other differential diagnoses include essential head tremor, tardive dystonia, anterocollis caused by cervical

myopathy, multiple system atrophy, secondary torticollis associated with neck injury, atlantoaxial dislocation, cervical disc disease, spinal cord neoplasm, or cervical soft tissue infection (3)

The cause of CD is often unknown. Although in most cases it is idiopathic, it can also be a result of genetic mutation or occur post head or neck trauma. There is usually persistent co-contraction of agonist and antagonist muscles, and this has been postulated to be a result of impaired reciprocal inhibition (13). Patients with CD have been shown to have somatosensory dysfunction and may also have concomitant vestibular abnormalities (13). The involvement of multiple systems makes a thorough examination the crucial first step in a therapy program (see Table 6.1). The physical therapist should pay particular attention to the following areas (13):

- Palpation of the cervical area and shoulder girdle to determine which muscles are overactive (electromyogram [EMG] analysis is most accurate to identify overactive agonists and underutilized antagonist muscle groups)
- Pain assessment
- Postural assessment
- Patient response to geste antagoniste (sensory cues)—assess responsiveness to cues as well as the timing and segment of the cues that are most effective.
- Cervical active and passive range of motion pre- and postsensory cues
- Muscle length and strength throughout the entire spine and shoulder region
- Preferred movement patterns of the cervical, thoracic, and lumbar spine as well as shoulder girdle during active motion and functional tasks
- Appropriate and commonly used outcome measures: Toronto Western Spasmodic Torticollis Rating Scale (TWSTRS) (29) and Cervical Dystonia Impact Profile (CDIP-58) (30)
- Balance deficits and postural control, including righting and equilibrium reactions, postural sway, and vestibulo-ocular (VOR) testing

Treatment of CD

Idiopathic CD is often associated with various degrees of disability. The issues encountered by patients may range from subjective discomfort in social situations with little to no major consequences in daily life activity to appreciable and substantial alterations to the home and work situation (31). Many treatments for CD are typically provided by physicians and can range from

pharmacologic to surgical intervention. Intuitively the role of physical therapy may be significant in this patient population, but few studies have examined traditional physical therapy approaches (see Table 6.2 for a review of available treatment modalities). The approach to treatment should be tailored to each individual patient and should be coordinated with interventions provided by the physician or other members of the multidisciplinary or interdisciplinary team.

In focal dystonia, botulinum toxin injections are the treatment of choice (9). It has been suggested that utilizing a rehabilitation program in association with botulinum toxin injections will increase the effectiveness of the toxin and in some cases lower doses may be administered (24). It has also been postulated that the timing of physical therapy after Botox treatment can impact the effectiveness of the physical therapy program (11). By initiatiating physical therapy within the first 2 weeks following Botox injection, you can in many cases take advantage of the weakened muscles that result in the CD. The antagonist muscles that require training to promote improved posture may, following treatment, be more compliant and able to contract in meaningful ways.

Deep brain stimulation (DBS) has demonstrated excellent improvement of dystonia, pain, and function in select cases with CD (32). No large studies have examined the use of physical therapy as an adjunct to DBS surgery (13). The role of the physical therapist can be dynamic in the management of CD. It is reasonable for physical therapy to be administered in combination with DBS to assist the patient with management of volitional muscle control and postural alignment and to improve the modulation of control accessed as a consequence of the surgery. Primary dystonias seem to have a better response to DBS, and that should be kept in mind when treating patients (32).

Goals of the Physical Therapy Program

- Preserve flexibility of the cervical spine and soft tissues
- Reduce the intensity of spasms
- Control head position/posture (first voluntarily, then automatically)
- Reduce or eliminate pain
- Allow the patient to maintain or regain a more preferable level of function

Successful intervention requires the application of an appropriate treatment plan and adequate patient education, especially on expectations. It is imperative

that patients learn which muscles need to be reinforced and which need to be relaxed. They will need to engage in the regular practice of activities that reinforce the use of the antagonist muscles and attempt to maintain the head in a straight and stable position at rest and during everyday activity (11).

The most effective treatment techniques include:

- Manual stretching of the cervical muscles
- Strengthening of antagonist muscle groups or exercises that reinforce appropriate posture
- Neuromuscular electrical stimulation to the primary corrective muscles/antagonists
- EMG/biofeedback to reduce spasm and muscle activity of agonists as well as learn to appropriately activate the antagonist muscles (10)
- Water as a medium to promote a relaxing environment for exercise
- Relaxation techniques
- Activity and environmental adaptations to remove stimuli that exacerbate the dystonia
- Use of sensory cues or tricks to improve head position—touching the chin, top of the head or back of the head, or contralateral side of the head
- Postural reeducation with emphasis on midline alignment; use of a mirror for visual feedback

Additional techniques reported in the literature may prove beneficial for managing CD, but their effectiveness is more questionable:

- Cervical collar or brace: use caution if used to provide static support for the neck and head, but specially designed braces may substitute for a sensory input/trick by touching certain portions of the head or neck. Caution should always be exercised when immobilizing, which can lead to contractures (9).
- Soft tissue and joint mobilization
- Transcutaneous Electrical Nerve Stimulation (TENS)
- Muscle vibration and vestibular stimulation

Instruction for Prescribing Home Exercises (11)

- The more often the corrective/antagonist muscles are worked, the greater the chance for obtaining normal muscular contraction.
- Introduce into the daily routine or activity the promotion of motor relearning.

- Exercises should be performed slowly while focusing on maintenance of appropriate posture.
- Allow adequate rest periods.
- Start from a resting position and then, once control improves, begin dynamic postures and functional movement. Integrate activities into function such as play, chores and work.
- Exercise in front of a mirror.
- Discontinue or modify any activity that aggravates spasms or postural deviation.

Recommendations for Daily Activity and Environmental Adaptations (11)

- Sit in comfortable chairs where the head can rest in midline against the back and the arms are supported. This technique can be applied in the car as well by using the headrest for support or sensory cue.
- Manage head position during sustained activity such as TV watching or reading.
- For sleeping, try to remove all pillows from the bed except a small, thin one. Otherwise lie prone with the head turned toward the corrected side.
- At home or work ensure visitors sit at the side that corrects your torticollis.
- Ergonomic assessment of your office/work space: arrange desk to facilitate correct posture while using the computer or phone.
- Use strategies for body mechanics to prevent or reduce exacerbation of symptoms.
- Discuss with family and colleagues to enhance their understanding and facilitate their interpersonal relationships.

Considerations for Management of CD or Torticollis in Pediatric Patients

Although spasmodic torticollis (also referred to as CD) is primarily seen later in life, it is not uncommon to see infants with asymmetry consistent with torticollis. This asymmetry may be congenital or induced by preferred positioning/posture of the infant. It may be present at birth or develop in the first months of life. There seems to exist a significant association of torticollis with plagiocephaly and craniofacial asymmetry. Conservative management strategies, including physical therapy, seem to be beneficial when

applied early (between 2 and 8 months of age) (33). Physical therapy should include (33):

- Education to parents/caregiver regarding positioning, handling, and carrying to prevent deformity
- Range of motion of the cervical spine and positioning for play with emphasis on "tummy time" and toy placement
- Specific stretching and range of motion exercises aimed at correcting asymmetry
- Avoidance of passive manipulations or manual stretching which provoke discomfort in the child
- Consideration of orthotic devices if deformation of the skull or neck is present and persists beyond 5–6 months

When torticollis occurs in a child older than one year of age, differential diagnosis may include medication-induced response, encephalitis, toxin exposure, Benign Paroxysmal Positional Vertigo (BPPV), visual impairment, brain tumor, and Sandifer syndrome.

In the case of Sandifer syndrome, the torticollis occurs intermittently and can change sides. The abnormal and changing postures are associated with gastroesophageal reflux disease (GERD) and failure to thrive or growth retardation in the older child. In Sandifer syndrome, the abnormal posturing with GERD is characterized as dystonic and usually occurs in the early phase of presentation. Typically, it is observed from infancy to early childhood (34). There is an unusual combination of GERD with or without hiatal hernia, torticollis, and dystonic body movement mainly involving the neck and upper extremities. The movements are thought to be an anatomic defense mechanism against repetitive acid reflux. It is considered that the abnormal movements during the reflux are the result of a mechanism to protect the air passages from reflux or to relieve the abdominal pain caused by acid reflux (35).

It is important to note that the clinical manifestation of Sandifer syndrome can be resolved with successful diagnosis and treatment (36). The intermittent occurrence of torticollis with alternating directions, normal sternocleidomastoid muscles, and normal cervical radiographic findings make Sandifer syndrome a probable diagnosis and necessitate upper gastrointestinal studies (37). With proper treatment, the change can be dramatic, with a rapid disappearance of symptoms.

Sandifer syndrome is commonly exhibited in the pediatric population with brain damage or metabolic disorders and is often not differentially

diagnosed (36). The child with neurologic deficits must be examined thoroughly to determine the etiology of the clinical manifestations so that the child will be treated appropriately.

Physical therapy intervention for CD requires time, patience, and commitment. The course of treatment may be prolonged, taking several months, and may require semiannual or more frequent follow-up with therapeutic intervention. It is important for the patient to be diligent regarding the home program and communicate with his or her physician or physical therapist regarding any changes (11,23,38).

Limb Dystonia

Limb dystonia is a much less common focal dystonia. It is often brought on by skilled or strenous activity and may respond poorly to rest. In rare cases focal limb dystonia may be associated with structural lesions of the basal ganglia or other disorders such as corticobasal ganglia degeneration and progressive supranuclear palsy. Limb dystonia in the foot can be a presenting sign for Parkinson's disease. In adults, focal task–specific dystonia is more common in the arm than the leg (3). Common manifestations of limb dystonia in the arm may be associated with musicians as well as with writer's cramp. There are specific treatment approaches to maximize the therapeutic benefit for these patients. See Chapter 7 for the details of treatment approaches for arm and hand dystonia.

Although primary upper extremity and hand dystonia has been well characterized, primary lower extremity and foot dystonia has not. Adult-onset primary dystonia in the lower extremity is rare, in contrast to childhood-onset dystonia, which typically begins in the foot (39). Adult-onset cases are often considered to have an identifiable cause such as trauma, medication, Parkinson's disease, or psychogenic factors but can be primary and minimally progressive in nature (40). The physical therapy management of primary lower limb dystonia has not been well documented primarily due to the rarity of the disorder. Patients may present with abnormal posturing of the foot in plantar flexion, inversion, and toe flexion or extension, which may worsen with activity. The approach of the physical therapist should be designed to:

- Preserve joint and soft tissue flexibility and range of motion
- Reduce spasm intensity and pain

- Promote appropriate postural alignment
- Facilitate the preferred level of function for each individual

The physical therapist should be sure to coordinate care with the rest of the interdisciplinary team when available. Treatment techniques identified in Table 6.2 should be considered for patients with lower limb dystonia and timed appropriately in collaboration with physician interventions. In focal dystonia, botulinum toxin injections are often the treatment of choice (9). The physical therapist should specifically consider:

- Orthotic devices to facilitate and maintain appropriate foot and akle position
- Strengthening exercises to activate antagonistic muscles (ankle eversion and dorsiflexion) as well as strengthen hip and trunk muscles to promote improved proximal stability
- Range of motion and flexibility exercises to preserve joint and soft tissue mobility
- Somatosensory stimuli and proprioceptive training to promote cortical remodeling (refer to the occupational therapy section on focal hand dystonia)
- Education for activity modification to reduce or remove provocative stimuli
- Assistive devices for ambulation to reduce physical stress on the affected lower extremities

Considerations for Management of Lower Limb Dystonia in Pediatric Patients

In childhood the initial presentation of the foot turning in may represent the early manifestation of primary generalized torsion dystonia. In order to facilitate the appropriate differential diagnosis, the therapist needs to be aware of alternative causes of lower limb dystonia.

One type of lower limb dystonia that can be misdiagnosed is dopa-responsive dystonia (DRD). It is a rare disorder presenting in early childhood with foot dystonia, gait abnormality, and hyperreflexia (3). A classic feature of DRD is worsening of symptoms at night. There may also be a lessening of the dystonic posturing at rest. It is important that DRD be considered in the case of a child with clinical symptoms, such as dystonic posturing of the foot, causing loss of balance and frequent falling, which gets worse at night and improves with rest. Since the clinical presentation could be identified as neurologic findings, DRD is often misdiagnosed as cerebral palsy. The hallmark

of DRD is a dramatic and sustained response to levodopa (3). There is a rapid response with marked changes in the clinical outcome and the quality of life of the child. The clue to the diagnosis of DRD is the diurnal fluctuation and lower limb onset. The physical therapist should be alert to the symptoms that are described by the parents/caregiver so they may be able to provide feedback to the referring physician.

If the single limb dystonia progresses to generalize to other body parts, a diagnosis may be made of primary generalized torsion dystonia which often shows no response to levodopa (3). At onset, a typical clinical presentation in a child is the foot turning in at initiation of gait. The parents may report that the dystonia seems to be related to the child's emotional state or level of rest. They may report a unilateral or bilateral presentation. The primary concern of the parents is the increase in falling related to the aberrant gait pattern. Depending on the age of the child, the clinical symptoms may mimic cerebral palsy and be diagnosed as such. It is imperative that the physical therapist perform a thorough examination and reassess the patient on a regular basis in order to identify any changes in the clinical presentation. The findings from the examination can assist in the differential diagnosis.

It should also be considered that dystonia can be caused by medications that block dopamine receptors, such a neuroleptics (41). Neuroleptics can affect the extrapyramidal system and are therefore sparsely used for the pediatric patient, but may be prescribed for the young adult. The physical therapist should be aware that the prescribed medication may cause changes in the patient's clinical presentation.

One additional type of limb dystonia that is rarely encountered is paroxysmal exercise-induced dystonia, which can be mislabeled as stress or psychogenic. This phenomenon has been described to typically affect the lower extremities, but cases have been reported that affect the arm and shoulder region. It is characterized by episodic dystonic attacks or dystonic attacks after prolonged exercise. Each episode can last 5–30 minutes, but in some cases this may progress toward more of a segmental or generalized dystonia (27,42,43). Physical therapy intervention has not been documented in this population mainly because of its paroxysmal nature. It is important to complete a thorough examination (Table 6.1), and one may consider having the patient videotape the dystonia if it cannot be reproduced in the clinic. The therapist should be sure to coordinate care with the rest of the interdisciplinary team and to consider the treatment strategies in Table 6.2.

Generalized Dystonia

Generalized dystonia refers to the involvement of multiple anatomic areas. It typically begins (especially DYT-1 genetic dystonia) as a focal dystonia in the foot and ankle (a foot turn) and progresses to involve the entire lower extremity with any combination of trunk, opposite lower extremity, upper extremities, neck and face within several years of the first manifestation. The first manifestation does not have to be in the foot. It is a chronic, "progressive" disorder, with the most severe cases typically presenting in childhood, although in many cases the progression levels off and remains stable for the remainder of the sufferer's life (3). Physical therapy management should begin early in the process and adapt as symptoms change. The interventions provided by the physical therapist should progress from corrective techniques and mild adaptations in the early stages to compensatory strategies and caregiver training in more advanced stages (Figure 6.3). Physical therapy interaction with the interdisciplinary or multidisciplinary team can enhance the comprehensive management of the patient.

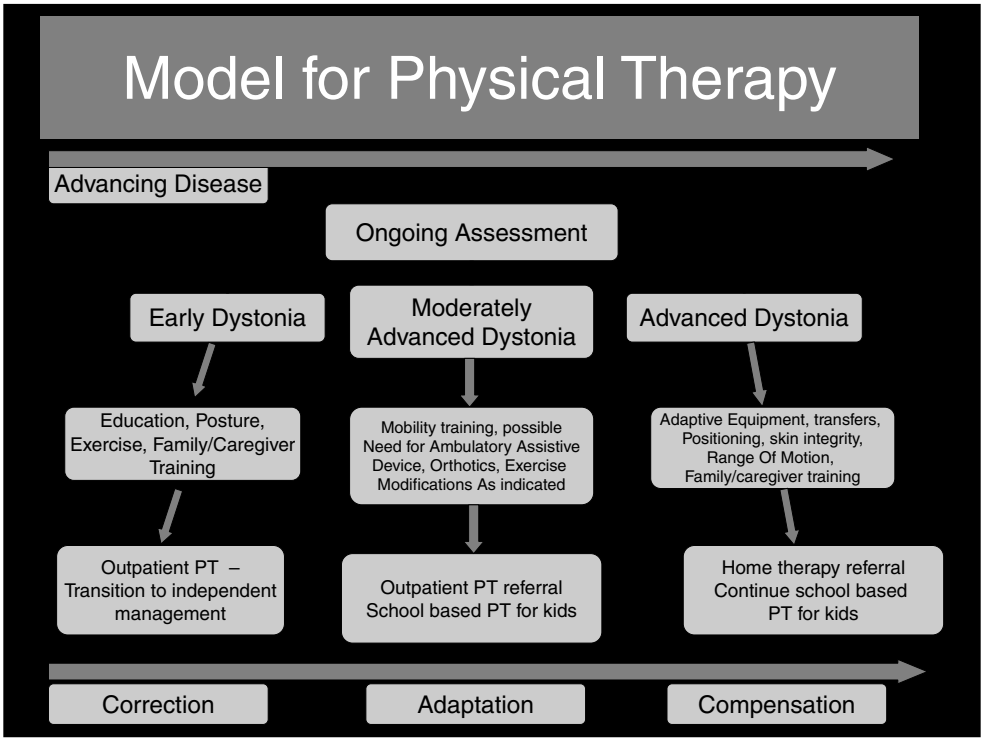

FIGURE 6.3 The strategy of the physical therapist changes with dystonia progression. Treatment planning should include long-term follow-up.

Physical Therapy Management

Physical therapy management should ideally begin with a comprehensive examination (Table 6.1) in order to fully appreciate functional abilities and limitations. The examination should also include standardized tests for symptom severity as well as functional performance. It may be beneficial to utilize all or part of the same standardized tests utilized by physicians (Global Dystonia Rating Scale, Unified Dystonia Rating Scale, Fahn Marsden Scale) (8). Standardized tests of functional mobility should also be employed. Although no tests have been validated specifically for this population, you may consider using the Adult and Pediatric Functional Independence Measure (FIM [44] and WeeFIM [45]). It has been shown that these two tests can be used for monitoring functional status (through adolescence) in children with spastic cerebral palsy, and it is reasonable to apply the measures to dystonia patients (46). As dystonia progresses, these may be two tests that can be used to track patients from child to adulthood. Once the examination is complete, the therapist will be able to shape the treatment plan to the patient's interests and needs. Although this is often a progressive condition that starts with mild impairment and disability and may progress to severe impairment and dependence in activities of daily living (ADLs) and functional mobility, a prepared physical therapist will be able to manage the variations that present over time. The therapist should manage the symptoms and promote maximum functional independence. The focus should be on:

- Preservation of joint and soft tissue flexibility and range of motion
- Reduction of spasm intensity and pain
- Promotion of appropriate posture
- Facilitation to maintain the preferred level of function for each individual

The key difference for management in the early stage compared to the advanced stage is the involvement of different body areas. Therapy usually begins with strategies to manage one or two anatomic areas, and over time the plan can be adapted to apply techniques to multiple anatomic areas. Strategies to maximize function will also vary from early to advanced stages as the need for increased adaptation and compensation arises. It is also important to note that since early onset is typically under the age of 25, special considerations need to be made for the younger patients, particularly those in the pediatric population.

The stages below are arbitrary and only meant as a guide for the physical therapist as they will not apply to all patients.

Early Disease Duration Stage

Dystonia symptoms in the early stage are often focal in nature, commonly beginning with a specific action, and are not present at rest. If symptoms begin in one leg, they may only be present when walking and may disappear when the individual runs or walks backwards. Patients will often present to the clinic ambulatory, with mild gait deviation. Although some movements may be challenged due to pain, fatigue, and decreased efficiency, patients are typically still involved in all of their life roles (work, school, leisure, family, etc.). Therapy should focus on providing education and training for patients, family, and caregivers. This strategy should include:

- Role of physical therapy now and if the disease progresses
- Postural education for maintaining midline alignment and normal joint position
- Exercise to optimize physical performance, including aerobic capacity, muscle strength, and edurance
- Flexibility and range of motion exercises to prevent deformity
- Gait training to maintain independence and reduce fall risk
- Use of orthotic devices or casting in select patients

Moderate Disease Duration Stage

Symptoms in moderately advanced dystonia may progress to include more than one region of the body. Some of these patients have hemidystonia, multifocal or segmental dystonia depending on the relationship of the regions affected.(3). Individuals at this stage may be experiencing more difficulty performing in their roles (family, school, work). Patients at this point may start to develop more significant movement dysfunction. Ambulation will be more challenging, and the patient will present with significant gait deviations and may be at risk for falling. There are no standardized balance and gait assessment tools validated for dystonia. Depending on the age and general moblity of the patient, you should consider 1) the Berg Balance Test (47), 2) the Dynamic Gait Index (48), and 3) the Timed Up and Go Test (49). Another test that has recently been validated for use in traumatic brain-injured patients is the High-Level Mobility Assessment Tool (HiMAT) (50).

The HiMAT may also be useful earlier in dystonia as it has less of a ceiling effect (51). In this stage the therapist needs to address:

- Consideration of assistive devices for ambulation
- Balance assessment and training
- Task-specific training strategies for mobility
- Contracture prevention through exercise and orthotics
- Ergonomic and adaptive changes to the home, school, and work environment
- Adaptive strategies to continue life role activities and leisure activities
- Continued patient, family, caregiver education
- Revisit early-stage recommendations

Advancing Disease Duration Stages

In the advanced stages of dystonia, it may become generalized. The symptoms can affect many body regions (3). When patients have reached this stage, they may encounter severe movement dysfunction. Often ambulation will be very labored, and patients may be at a high risk of falling. Patients will be dependent for transfers, bed mobility, and ADLs. There will be significant challenges to remain integrated into their life roles (family, work, school, leisure). Spasms and dystonic posturing may result in significant, unrelenting pain. In addition to the early and moderately advanced-stage management strategies, the therapist needs to also consider the following issues:

- Wheelchair seating and positioning for mobility, skin protection, and joint positioning (manual and power mobility options)
- Positioning at home, work, and school
- Contracture management/prevention—education to patient, family, caregiver regarding positioning, range of motion exercises, orthotics
- Adaptive equipment for ADLs (see Chapter 7)
- Adaptive equipment for transfers
- Continued patient, family, and caregiver education

In patients with progressive dystonia, physical therapy intervention will often be provided in conjunction with other medical and surgical therapies. Botox is often used to treat more localized areas. Physical therapy intervention should be initiated during the first 2 weeks following injection to obtain the maximum benefit of the dystonic muscle being weakened (which happens many days following the injection). In cases where symptoms are more generalized, it is

more common that aggressive approaches such as surgical procedures like deep brain stimulation will be employed. The physical therapist should be involved in the patient's care prior to surgery and continue to follow the patient for 3–6 months or more after surgery to assure maximal benefit (52). As the patient responds to programming of the stimulator, the therapist should assist the patient with the management of volitional muscle control, postural alignment, and general control.

The use of the intrathecal baclofen pump has been shown to be effective in secondary or spastic dystonias and in some generalized cases (53). As with deep brain stimulation, the physical therapist should be involved in the patient's care prior to implantation of the baclofen pump and continue to follow the patient for several months after the procedure. As the patient responds to the medication, the therapist should facilitate more independent functional mobility through the management of volitional muscle control, postural alignment, and decreased spasticity/hypertonia. Strategies for transfers and mobility may need to be modified. Some patients may not realize significant functional improvement from the baclofen, but it may reduce their pain and allow for improved positioning at home, in the wheelchair, or outside of the home when interacting in the community. This may relate to improved quality of life and ease of care (54).

Pediatric Considerations

The onset of dystonia in childhood presents unique problems (55). The patient is challenged with the clinical manifestation of the involuntary abnormal posturing, which may or may not be associated with pain. It is not uncommon for the child to be assessed by multiple physicians prior to a definitive diagnosis. Once the diagnosis is rendered, it is imperative that the patient and parents be educated regarding the signs and symptoms of dystonia. Emotional and psychological support should be available for the patient and family members. The members of the multidisciplinary or interdisciplinary team should recognize the emotional needs of the patient and family. The child must be taught to cope with the changes occurring in his or her body. The family must be supported and encouraged to assist the child to develop coping strategies at home, in school, and in the community. There will be continuous challenges for the child and the family throughout the progression of the disease.

As the disease duration lengthens, changes may occur in the child's level of independent function. The clinical manifestations may include involuntary

muscle activation at rest or during movement, co-contraction of antagonistic muscles triggered during task performance, and/or overflow to uninvolved muscles during active movement (55). In addition to the physiological changes, there will be changes in emotional, cognitive, and physical growth and development, which may be age related. All of these factors must be considered in physical therapy in reference to goal setting, plan of care development, and treatment implementation.

The overall goal of physical therapy is to minimize the impact of the disability on the child and to retain optimal functional independence. It is important to find a means to ensure that the child can fully participate in family interaction in the home, peer interaction in the school, and an active plan for interaction with the community. The intent is that the child will continue to participate in his regular routine in the family and community. When establishing goals, the child and family should be consulted. The child and family will be motivated to comply with the therapeutic process, especially if they are integrated in the planning process. The process of planning and intervention should be family centered. The physical therapy program must be meaningful to the participants for optimal outcome.

In the early stage of the disease process, the physical therapy program should address correction and management such as:

- Child and parent education regarding the impact of emotional and behavioral state on the presentation of abnormal postures. The pattern as well as the magnitude of involuntary muscle activity varies with arousal, emotional and behavioral state, tactile contact, or attempted task (55).
- The child should be trained in reduction or control of posturing using sensory or motor tricks. The internally driven tactile cueing can result in a cessation of the involuntary muscle activation. Often children will have developed their own strategies for dampening or terminating the posturing. It is helpful to understand the child's mechanism for managing the abnormal posturing.
- Self-management of safety and energy conservation. The child will be driven to continue participating in play. The child will most likely still be independent in ambulation and be motivated to interact with their peers. The child must be taught about personal safety and energy conservation during play. The muscular activation during play may be increased, thereby increasing the magnitude and frequency of the abnormal posturing. Pain may occur due to co-contraction of antagonist musculature during dystonic posturing. The

child must learn to monitor pain and be taught to manage the posturing by taking frequent rest periods. The dystonia may decrease or stop at rest.

In the advanced disease durations, the physical therapy program should address the following:

- Ongoing consultation regarding adaptive and assistive equipment and devices should continue as the disease duration lengthens. There must be child and family education and training regarding the positioning for alignment and the use of adaptive and assistive equipment. The child may develop a marked reduction in joint ROM due to chronic muscle shortening during posturing. The child may require adaptive seating and perhaps orthotics in an attempt to maintain biomechanical alignment and to reduce secondary impairment of scoliosis or hip dislocation. If recommending an orthotic or splint, consideration should be given to the intended purpose of the device and the composition of the material used in its design. There is a possibility that the posturing will be aggravated by the points of contact with the splint or orthotic and cause skin breakdown or result in pain. The goal of the recommended device should be thoroughly explained to the child and to the family so that compliance can be met and there will be a successful outcome. To ensure success, it is important that the use of the device or equipment be implemented into the child's daily routine and not require additional attention to time. Any prescribed piece of equipment or device should be monitored closely for skin integrity and assessed regularly for proper fit due to growth and development. The device or equipment must also be evaluated on a regular basis to determine if the goal or objective for use is being accomplished or needs modification for improved functional outcome.
- There should be a modification of the child's coping strategies for "quieting" the dystonic posturing so that the child can continue to participate in family, school, and community activities. The family may continue to require ongoing support to assist the child to cope with further disability.

Considerations for the Home, School, and Community

Initially, in the home, attention should be focused on safety during independent function and play. Efforts should be made to maintain the child's role in the family unit, including participation in sibling and parent play activity.

Because of the effect of dystonic posturing on changing the center of gravity with respect to the base of support, balance will be compromised. Fall-prevention strategies such as removing throw rugs should be implemented. The bedroom and bathroom should be accommodated with a safety rail. In the bedroom, the safety rail will assist with stability and thereby independence during dressing. In the bathroom, the safety rail will help to avoid loss of balance by providing stability during toileting, hygiene, and bathing. As the disease progresses, additional adaptive and assistive equipment will be necessary to provide stability and alignment during sitting activities such as family dinner, watching TV, or playing interactive video games. The parent and/or caregiver will need instruction in transfer training and lifting using proper body mechanics. Contracture management should be a part of the home program. Attention should be given to the prevention of secondary impairments due to the chronic dystonic posturing.

At school the child will face greater challenges. There will be increased stress involved with academic performance and peer interaction. Emotional and physical challenges will face the child in the classroom, on the playground, during physical education, in the cafeteria, and negotiating the school campus. Transportation on the school bus may also prove to be a challenge. The parents may choose to have the child evaluated for and classified as having special needs so that the child can be placed in the least restrictive classroom and be eligible for related services such as physical therapy, occupational therapy, and speech and language pathology. The school physical therapist should be instrumental in addressing the child's special needs as these will be relevant to his or her educational environment and should include:

- Adjustment and adaptation of classroom seating. The child's desk position in the classroom is critical for visual orientation and feedback and to ensure optimal cognitive processing. Midline orientation must be addressed in the seating arrangement for adequate postural alignment. The child's chair should promote an erect, symmetric posture with an upright pelvis and feet supported on the floor for a stable base of support at rest. The desk height must be determined based on the presentation of posturing in the upper extremities. The occupational therapist should be consulted regarding positioning of the upper body posture to effect the best possible outcome for tabletop and fine motor activities. Adaptive or assistive devices such as

upright pegs (to be held in the hands) for distal stabilization may be necessary to utilize optimal performance and participation. The overall goal for the classroom seating is to provide appropriate postural alignment for optimal academic performance.

- Consult with the teacher regarding classroom structural arrangement to allow for safety while navigating the classroom. The classroom should be set up to provide space between desks for safe classroom negotiation since coordination and balance will be impaired. In the early phases of the disease, the child may need the assistance of an aide or another classmate to transfer to the floor or ground as required by the classroom activity. The child may need help transferring into and out of the desk chair. There should be room allowed for each activity so that the child can function with the least necessary assistance required to maintain independence. The child should be assisted only when he requests it, or if safety is a concern of the teacher or therapist. The therapist should be available for consultation with the teacher on a regular basis to ensure safety in the classroom. As the disease progresses to include the trunk and/or multiple extremities, supportive, adaptive equipment should be considered for the child in the classroom. Consideration should be given to the acquisition of a wheelchair to allow functional independence for the child in the classroom. If indicated, the child should be provided with power mobility to preserve energy during classroom activities. The use of power mobility will also allow the child to keep up with his peers, which will assist in social emotional development and self esteem.
- Consult with the teacher and aide regarding safe play on the playground.
- The physical therapist should instruct the child in safe negotiation on the playground. The child should be introduced to play strategies, which will conserve energy. The physical therapist should instruct the child as to whether there is a need for intermittent rests or other strategies to "quiet" the dystonic posturing.
- Consult with the physical education teacher. The physical therapist should consult with the physical education teacher regarding recommendations for adaptations so that the child can continue to participate in the physical education class. The physical therapist may decide to use the class time as the treatment session to integrate the child for optimal participation in the group activity. As the dysfunctions increase in correlation to the disease

progression, assistive equipment may be indicated. The physical therapist should be integral in the recommendation and acquisition of equipment. Every effort should be made to adapt the physical education activity to meet the needs of the child so that he or she will be motivated to continue participation to his or her greatest capacity.

- Consultation and training in the cafeteria. During the initial stage of the disease, the child may need assistance with carrying the lunch tray and/or negotiating the crowded environment. Seating placement should be addressed for safety and ease in transfers. The school occupational therapist can assist with feeding strategies for independence.
- Consultation and training for safe toileting. The physical therapist should evaluate the child for safety in transfers in the bathroom. They should also evaluate the technique of toileting used by the child to ensure ease and efficiency in performance. There may be a need for toilet adaptation to provide optimal postural alignment for comfort during excretion. Relaxation during toileting is critical to reduce the dystonic posturing and make the toileting experience successful.
- The child's mode of mobility to negotiate the school campus should be assessed for conservation of energy, safety in function, and optimal independence. In the early stages, the child may only need supervision. Later, as the disease progresses, a manual or power wheelchair may be indicated to provide functional independence in mobility. If the child must be transported on the school bus, the physical therapist should consult with the child, parents, and bus driver to determine the safest strategy for getting on and off the bus. Safety in positioning on the seat during travel should also be assessed and considered. When indicated, the physical therapist can instruct the child in transfer techniques on and off the bus using a lift on an accessible bus. The child should then be able to maintain functional independence and fully participate in wheelchair mobility including bus transfers.

The family should be encouraged to continue participating in community activities and adapt to ensure full participation by the involved child. The family should be instructed in awareness of architectural barriers, which may require them to assist the child for optimal mobility and maneuvering. The family should also be made aware of the potential need for the child to conserve energy and take frequent rests. It is important that the parents be

aware of the child's cues for being tired or in pain. It is important to empower the parents to become the advocates for their child's continuing physical needs and to encourage them to seek out available modalities for their child's physical needs, including the use of adaptive or assistive equipment as the disease process advances.

The physical and emotional needs of the child will change with the progression of the disease. The physical therapist should be able to adapt the plan of care to the changing needs of the child through adolescence and then on to adulthood.

Summary

Dystonia is a complex disorder that may have an onset from early childhood to middle age. It may affect different body regions and have differential responses to treatments depending on type (primary vs. secondary) and body regions involved. Management of disability in dystonia is not always straightforward and should, if possible, utilize a multidisciplinary or interdisciplinary approach. The role of the physcial therapist should be dynamic and consider the needs of children and adults with a potentially progressive neurologic disorder. The physical therapy role may change based on the age of the patient and presentation of the dystonia. Initially, the physical therapist will be instrumental in educating the patient and family in correction and management strategies. As the dystonia progresses, the role should evolve to assess and recommend adaptive equipment and steer planning strategies based on functional presentation. The overall goal of physical therapy for the patient with dystonia is to minimize disability and maintain as much functional indepedence as possible. The physical therapist has the potential to enhance the quality of life for the patient with dystonia.

Pearls for Physical Therapists

1. The physical therapist needs to be dynamic and must consider the needs of patients, especially if their condition is progressive. The patient with a diagnosis of dystonia must be assessed on a regular basis for changes in functional status. The changes will occur as the disease progresses. It should be noted that patients often present with normal range of motion

and muscle tone in the early phases of the disease, and it is our role to preserve that as the dystonia progresses. In the pediatric case, normal growth and development will also influence changes in level of function. Recommendations will be needed for adaptive and assistive equipment.

2. Physical therapy for the patient with dystonia should be focused on treatment of the symptoms and secondary impairments rather than the dystonia itself. Successful intervention requires the application of an appropriate treatment plan in conjunction with adequate patient education, especially as to expectations.
3. Physical therapy is a slow process that should be approached with the expectation that there will be a significant commitment in time, but with that commitment should come optimism. Results may not be immediately apparent, but a physical therapy program can influence many aspects of daily living.
4. A child older than one year of age with alternating torticollis and persistent vomiting and dystonic posturing should be referred to the physician to rule out Sandifer syndrome.
5. Certain medications can induce dystonic posturing. The physical therapist should be aware of the timeline of presentation of the dystonia relative to the onset of the prescribed medication. This can be determined in the patient/parent interview at initial examination.
6. Dopa-responsive dystonia should be considered in any child who presents with paroxysmal or progressive hypertonia of unknown etiology. The child may initially present clinically as a toe walker. It should be considered because there is typically a dramatic response to levodopa in these children. These children are often misdiagnosed with cerebral palsy or another neurodegenerative disease.
7. Botox has been found to be effective in the treatment of focal dystonia to weaken the agonistic musculature. Initiating physical therapy immediately after Botox treatment can affect the effectiveness of the physical therapy program. Antagonistic strengthening and muscle reeducation can be the focus of the physical therapy program during this time.
8. When prescribing assistive devices or adaptive equipment, we should not only consider how it will immediately improve the patient's balance, mobility, or independence with a functional task, but also if it can be used to improve technique or eliminate positioning that would otherwise exacerbate the dystonic activity.

9. Dystonia can be minimized or temporarily relieved by patient-initiated sensory or motor tricks. The physical therapist can guide the patient to recognize this process in order to control the dystonic posturing.
10. The physical therapist must be aware that cognitive processing, visual feedback using a mirror, and proprioceptive feedback can be used in treatment for effective knowledge of performance and motor learning.

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7

The Role of the Occupational Therapist on the Interdisciplinary Team for the Evaluation and Treatment of Dystonia

Portia Gardner-Smith

The occupational therapist (OT) has a unique and compelling role in the evaluation and treatment of individuals with dystonia. Occupational therapy is an applied science and rehabilitation profession aimed at enabling individuals to reach their maximum potential in performance of daily living skills, in work and school productivity, and in leisure through the use of purposeful activity (1). Occupational therapy uses purposeful, therapeutic activities to prevent and mediate dysfunction and to promote adaptation in an individual. The ultimate goal of the treatment program is to restore the individual to his or her maximum level of performance in valued occupational roles through restorative or compensatory treatment approaches (2).

The diagnosis of dystonia is not a common one. The incidence is 29.5 per 100,000 for focal dystonias and 3.4 per 100,000 for generalized dystonia (3). Unfortunately, the patients who get an accurate dystonia diagnosis usually are not referred to occupational therapy. When they are referred, often the therapist has not had previous experience with dystonia. When my first patient with dystonia entered the clinic several years ago, I was baffled. I turned to other skilled occupational therapists in the area only to find that they had little to offer in the way of experience or advice. Likewise there is little clinical research available to guide the therapist when treating individuals with dystonia.

What we do know is that dystonia is a rare disease leading to dysfunctions that are not easily addressed with routine approaches. There is currently little written to guide therapists in their assessments and treatment regimens. Consequently, many OTs may feel challenged, afraid, or even confounded as

to where to start or what to do next when assessing and treating a patient with dystonia. The purpose of this chapter is to provide guidelines for the occupational therapist in the evaluation and treatment of the patient with dystonia.

As with most neurologic disorders, the occupational therapist is one part of an interdisciplinary team. A multidisciplinary approach to the treatment of dystonia is a well-supported construct with literature supporting its effectiveness in other neurologic disorders such as Parkinson's disease, Huntington's disease, and multiple sclerosis. In fact, a multidisciplinary approach is essential to promote optimal quality of life at each stage of the disease process (2,4–6).

This chapter will focus on two focal dystonias of the hand—writer's cramp and musician's dystonia—and on generalized dystonia such as DYT1 type or secondary dystonia. The evaluation and treatment principles outlined here can also be generalized to other types of dystonia. Occupational therapists possess a unique approach and skill set that can positively influence these conditions.

Dystonia Defined

Dystonia is a family of movement disorders characterized by sustained muscle co-contraction in the extreme end range of a movement, frequently with a rotational component, causing involuntary, often twisting movements. There is excessive co-activation of agonists and antagonists that interferes with the timing, the execution, and the performance of independent motion. Dystonic movements share a directional quality and are consistent and predictable, although they vary greatly per the individual. There is rarely a problem with tone or strength. Dystonia is characterized based on age of onset, area of the body affected, and primary versus secondary (5,7).

Focal Dystonia

Focal dystonia involves only one part of the body (Figure 7.1). The etiology remains unclear. Imaging studies suggest that the central sensory representation of the hand degrades, leading to a loss of motor control (8). Of the primary dystonias, focal dystonia has the highest incidence rate at 117 per million. Although focal dystonia can occur at any age, it usually occurs in adulthood;

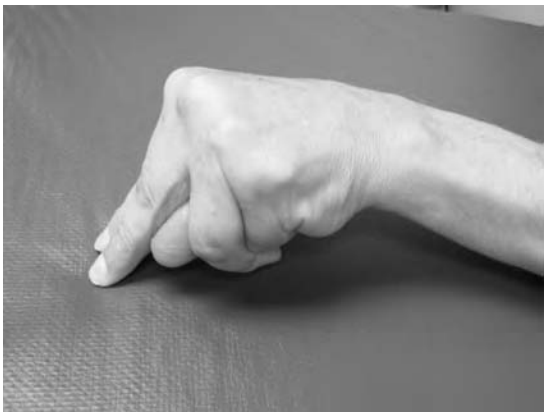

FIGURE 7.1 The positioning pattern of a patient with focal hand dystonia.

the peak incidence age is in the 40s, and 75% of those affected are men. Of the focal dystonias, spasmodic torticollis is the most common followed by blepharospasm and then writer's cramp (3).

Recent neurophysiologic findings can provide guidelines for the occupational therapist's assessment and intervention approach with these patients. Interestingly, abnormal sensory processing is implicated in this motor problem.

Normally discrete somatotopic areas assigned to individual fingers of the hand have begun to fade into one another. Loss of these clearly defined functional regions in the somatosensory cortex results in difficulty discriminating the digits from one another. This faulty somatosensory input is directly related to the abnormal motor output. The abnormalities in the cortex differ in patients with writer's cramp and with musician's dystonia. However, patients with different forms of focal hand dystonias share a variety of abnormalities in sensory processing, sensorimotor organization, and motor excitability. Experimental treatment approaches manipulating the sensory input of the hand have had promising results (8–14).

Writer's Cramp

Writer's cramp is a task-specific dystonia triggered by writing. Writer's cramp is unlike the other types of dystonias in that it is task-specific, usually not affecting the hand in any other activities. Tremor or myoclonus can be superimposed on the dystonic spasms or could be the main feature of the writer's cramp. There are a variety of clinical presentations of abnormal dystonic postures. Most commonly observed will be wrist flexion or extension,

pronation or supination of the forearm, flexion or extension of the fingers, forced flexion or extension of the thumb and index finger, or shoulder elevation (13,15–18).

Musician's Dystonia

Although it may occur in any instrumentalist, musician's dystonia is more common in musicians who play keyboards, strings, and woodwinds. Overuse appears to contribute to the disorder (13,19). Repetitive movement under high cognition or attention with strong desire for perfection has been linked to musicians who develop dystonia. The rapid high repetition is interpreted by the brain as a simultaneous contraction, and a learned sensorimotor dysfunction results as the homunculus seems to smear (20,21). Most musicians report that the symptoms increase with stress. Anxiety, tension, and other psychological factors appear to play a role in the development of the disorder. Jabusch (24) found a higher prevalence of anxiety and perfectionism in musicians with focal hand dystonia compared with other musicians.

Biomechanical disadvantages in the hand may also contribute to dystonia in this group. Small hands and decreased digit span measures can promote compensatory movements. Over time with excessive practice, these compensations can lead to functional changes in the cortical mapping (19,22,23).

OT Evaluation of the Patient with Focal Hand Dystonia

Patient Screening and Interview

The patient interview can provide the therapist with the essential clues necessary to distinguish the contributing factors leading to the focal dystonia. First and foremost, the therapist must evaluate how the dystonia is affecting the individual's quality of life. Evaluation of occupational performance involves discussion of the patient's life roles, daily routines, environments most encountered, and personal goals. This will provide the foundation for the rest of the exam.

Taking a thorough medical history is an essential step in the successful management of focal hand dystonia. During the initial interview, the therapist should screen for several specific indicators associated with focal hand

dystonia (FHD). While not the cause of focal hand dystonia, injuries resulting in loss of upper extremity joint motion have been identified as a risk factor for FHD (9). Therefore, a comprehensive fall history is prudent. Specific attention should be focused on falls onto an outstretched arm, upper extremity fractures, nerve compression syndromes (including thoracic outlet syndrome), head injuries, and other injuries of this nature. Note duration of the dystonia and the age of onset. What has been the progression of the disease? The pattern of progression can lend information to strategies formerly used that may be reconstituted in the treatment process.

Is there overflow, meaning does it appear with voluntary movements of other body parts? Is the dystonia present at rest? The presence of a rest dystonia is suggestive of a secondary dystonia. Investigate if the patient can identify a trigger. Often the patient may not be aware of this until the issue is explored by a skilled examiner. The patient's use of sensory or motor "tricks," such as a touch on the hand to relax it, are unique to focal dystonias (3,5,13). Ask questions regarding the effect of emotional status or fatigue on the dystonia symptoms.

Assess pain level, including questions regarding what activities make the pain worse and what interventions lessen the pain. Pain is not always a feature in the patient with focal hand dystonia; however, when pain is reported, it is generally secondary to compensatory movements that are being made.

Posture and Functional Mobility

Because restricted range of motion is a risk factor for focal dystonia, the therapist should screen for range-of-motion limitations. Restricted range of motion is a risk factor, even though it is not a cause of the dystonia (9,23). Evaluate any anatomic restriction, especially looking for decreases in finger abduction, forearm rotation, or shoulder rotation. Manual muscle testing of the digits is suggested as often the intrinsic muscles of the hand, or some of them, will be weak (9). When evaluating motor skills, look for impairment by errors in timing and/or force. Is there cramping when at rest?

Since the use of ergonomic principles and an alteration of postural approach have been linked to decreases in dystonic disruption of functional movement, note the postural pattern chosen during each of the tasks where dystonic movements are demonstrated (24). Do the dystonic movements change in severity or frequency if the posture is altered?

A posture evaluation in sitting and in standing is important (17). Look proximal to distal. Are the symptoms reduced when the proximal arm is stabilized? Does stabilization, either proximal or distal, help decrease the dystonia postures?

Observe the patient first sitting or standing statically for 30 seconds. Note any changes that may occur as he or she maintains that position, including involuntary movements or postural asymmetry.

Next, consider more dynamic postures. Take the opportunity to observe the patient when the patient is unaware of your observation. Note the upper extremity posture and movements during gait, while navigating among other people and objects, movement transitions such as coming from sit to stand, signing the intake forms, and other basic activities.

Note endurance and fatigue factors. Does the functional mobility decrease as the evaluation process advances?

Describe the Movement Pattern

When not performing the target tasks, many patients with focal dystonia will show normal results on the traditional neurologic exam. Generally they report that the involuntary co-contraction of the muscles of the hand occurs during specific tasks such as writing or playing a musical instrument (9,23).

Observe or trigger the abnormal movement. Differentiate between the types of movements that may occur. Dystonic contractions are associated more with prolonged bursts of electrical activity than with the short bursts of myoclonus. They tend to have a sustained, directional nature rather than the random flowing of chorea. They are distinguished from the involuntary, rhythmic "back-and-forth" movement characteristics of tremor. However, in some patients tremor-like muscle spasms, referred to as dystonic tremors, may be present when attempts are made to resist abnormal involuntary movement (3).

In writer's cramp, observe the patient writing and describe this movement in detail. Often the OT is asked by the physician to assist in determining sites for Botox injection. Having a true understanding of the musculature involved and the movement patterns displayed will be essential when offering suggestions of this type.

With musician's dystonia, observe the patient playing the instrument. In each affected musician, the dystonic hand behaves in a pattern unique to that musician (18). However, between individuals the pattern of dystonia varies.

Document the observation. Listen for unevenness in the playing. The patient may have more difficulty with certain types of drills or passages, so test a sample. The most affected hand is typically the one that does the most work or is positioned in the most awkward position (19).

Observe Coordination

Scores on standardized coordination assessments are less important than the opportunity to observe both extremities during the performance of these novel tasks. Look at errors in rate, rhythm, range, direction, and force of movement. Examples of tests that can be used are: rapid alternating supination/pronation, Minnesota Rate of Manipulation, the Nine Hole Peg Test, and the Crawford Small Parts Dexterity Test (2,25–27). The rapid alternating supination/pronation test is a simple yet effective tool in observing the speed and timing during reversal of movement. The Minnesota Rate of Manipulation allows the evaluator to measure one-handed performance, two-handed integration, and functional reach.

For more fine motor assessment, the Nine Hole Peg Test is a quick and simple measure of one-handed fine coordination. In patients with early stages of dystonia, the Crawford Small Parts Dexterity Test may be useful to obtain objective data on refined dexterous skills.

Activities of Daily Living

Often patients with focal hand dystonias report no decline in independence in ADLs. This may be due to the dystonia affecting a nondominant hand, the compensatory techniques already used by the patient, or the lack of dystonic posturing with tasks other than the target task.

Nevertheless, each area of self-care should be addressed since each task carries an intrinsic assessment of mobility. Self-care tasks include feeding, hygiene, grooming, dressing, toileting, and bathing. Does the patient with writer's cramp have the ability to manage a toothbrush or a fork without difficulty? Are the dystonic postures that are demonstrated during self-care position-dependent? Do the postures change if the shoulder is elevated or if the task is performed in standing versus sitting?

Communication skills are evaluated. Communication includes writing, computer and mouse use, and use of a telephone and cell phone (Figure 7.2).

FIGURE 7.2 An example of dystonia hand posture when attempting to pick up a pen.

Interaction with the community is assessed. This includes discussion of the effects of the abnormal movement on the patient's ability to drive. If driving safety is a concern, the patient should be referred for a comprehensive driving evaluation, often offered at an adult inpatient rehab facility. Community interaction includes the patient's involvement in recreational activities. Does embarrassment over the movement disorder hinder social interactions? Table 7.1 summarizes the activities of daily living tasks that should be considered.

The Jebsen Test of Hand Function may be used. This test is an objective measure of standardized tasks. It has seven subtests, which include writing, turning cards, picking up small objects, stacking checkers, simulated eating, moving lightweight objects, and moving weighted cans. Norms are provided for patient comparison (28). Not only is this test important for the standardization of the task completion and the speed of completion, it is also an excellent time to make observations. The appearance of mirrored movements in the relaxed dominant hand while writing with the nondominant hand is a valuable clue. When these mirrored movements show a reproduced dystonia posture in the dominant hand, similar to that shown when the hand is writing, the prognosis is more guarded (15).

Sensory Testing

Basic sensation is usually normal in dystonic patients, but general sensory testing using the Semmes Weinstein Monofilament Sensory Test is recommended (29,30). This test will identify the occasional patient who also

TABLE 7.1 Assessment of ADLs

ACTIVITIES OF DAILY LIVING	
FEEDING	HYGIENE AND GROOMING
Use of utensils	Shaving
Cutting with knife	Application of makeup
Drinking from cup	Hair care
TOILETING	DRESSING
On/off toilet	Pull-over and button-up shirts
Management of clothes	Buttons and zippers
Clean-up	Pants
	Socks and shoes
BATHING	COMMUNICATION
In/out of tub or shower	Writing
Underarms	Keyboard and mouse use
Feet	Telephone and cell phone use
Drying off	
INSTRUMENTAL ACTIVITIES OF DAILY LIVING	
HOME MANAGEMENT	COMMUNITY LIVING
Shopping	Transportation
Meal prep	Recreation
Cleaning and laundry	
Child care	

demonstrates abnormalities associated with other disorders, such as carpal tunnel syndrome. The majority of patients with focal hand dystonia show limitations in higher sensory processing such as localizing touch, graphesthesia, stereognosis, and kinesthesia. Specific testing of these perceptual domains is strongly recommended (10). Byl et al. outline a suggested assessment tool for testing stereognosis (31).

Occupational Performance

Occupation is the hallmark of occupational therapy. The occupational therapist's primary goal is to translate the clinical evaluation results into functional terms (32). Occupational roles are investigated. Priorities are established by identifying the value the patient places on return to self-care tasks, work or school performance, and leisure or recreational activities. This will also include exploration of the psychological adjustment to the impairment. OTs must not underestimate the difference between impairment

and disability. An impairment is a documentable physical limitation. Disability is the functional limitation imposed by the impairment. Therefore, the level of disability will be different for individuals even when the impairment is the same. For example, disability is very high for the patient with a dystonic hand who earns a living performing with an orchestra compared to a patient who can compensate for an inability to write by adapting to a laptop.

Use of standardized assessments may shed light on the need to further collaborate with the psychologist or the physician regarding adjustment issues. Standardized assessments such as the Quality of Life Scale can explore perceived impact of problems on quality of life and personal feelings about each problem (33). Another is the Beck Depression Inventory, which is self-administered and looks at manifestations of depressive symptoms (34).

OT Management of the Patient with Focal Hand Dystonia

Lie-Nemeth suggests a multifaceted approach with patients who have dystonia (19). Throughout this section on management, you will notice that rarely is one treatment technique used independently. Also, as part of the interdisciplinary team, the occupational therapist must collaborate with other team members during the development of the treatment plan and throughout the treatment process (13). For example, the OT will work closely with the physician if a Botox injection to the hand is considered.

Prognostic predictors have been identified in clinical trials. Older adults show better overall improvement than younger ones. The presence of dystonic tremor or the presence of overflow is a predictor of poor results. Djebbari et al. found that patients with a pronation/flexion pattern of dystonia showed the best and the most sustained improvement after Botox treatment than other groups. They also reported the pattern of thumb extension showed good functional improvements in patients with writer's cramp. Other studies reported poor response to Botox if there was tremor or inability to perform individual finger movements (15).

With musicians, psychological factors and hand biomechanics have been implicated in the development of dystonia. The presence of these potential red flag factors should be considered in the development of the treatment plan.

There is no definitive treatment approach for focal dystonia. However, several investigators have outlined specific treatment approaches that have proven successful, and these techniques will be discussed below.

Sensory Integration

The basis of sensory integration approaches is the assumption that carefully organized sensory input can result in the desired motor output. This approach emphasizes the use of only normal, tension-free movements with avoidance of abnormal movement strategies. Intense practice and cognitively demanding discrimination activities are required for task relearning. This approach is time-intensive for the patient (19).

Nancy Byl (8) has built upon this approach in the development of an evidence-based sensory discrimination model of therapeutic intervention. Her treatment approach is based upon findings from basic research with primates and applied research in humans. One of the primary findings in FHD is a change in the orderly representation of the somatosensory cortex. Previously discrete areas representing each digit have become faded, smearing into one another. Byl's approach attempts to restore the somatosensory representation of the hand in the sensory cortex through systematic modification of the sensory input into the dystonic hand.

Byl Treatment Protocol

First, the patient must stop performing the task.

Second, sensory discrimination retraining is performed. The patient is seen one-on-one in the clinic for 1–1.5 hours each session, one to two sessions per week. With vision occluded, a variety of stimuli are presented and repeated until the patient can accurately identify the stimulus. If the patient is having difficulty accurately identifying the stimulus, the stimulus is enlarged or vision is allowed until accurate identification can be made. Accuracy is essential.

To modify the sensory input, different positions are recommended. These may include supine, prone, or simply a position of the upper extremity that would not normally be associated with the task such as elevating the arm for writing. The three task classifications addressed are localization, graphesthesia, and stereognosis. All sensory discriminatory activities are initiated with the eyes closed and include identification of:

Various textures and temperatures
Objects or designs drawn on the hand and fingers
Use of one finger to read Braille letters, or playing games with Braille cards

Shapes, letters, or numbers drawn on the skin
Raised letters or numbers
Touching specific points on the skin

Other discriminatory tasks involved matching, such as:

Objects presented in a bowl of rice or beans
Dominoes
Shapes to openings for those shapes
Coins, beads, and buttons

In the third step, techniques are used to teach stress-free hand use. These include:

1. Mirror imagery: A mirror is placed between the affected and nonaffected hand. Simple sensory and simple motor activities are performed one-handed. The patient focuses on the mirror image of the performance of the unaffected hand. The patient is asked to make the affected hand look like the unaffected image.
2. Mental rehearsal: The patient is asked to imagine normal sensory processing, performing with normal motor control, and effectively completing the task. Therefore, the patient is to mentally rehearse the target task without performing any abnormal or involuntary movements. The goal is to reinforce normal perception and to increase confidence in normal movement.

Step four is the development of a customized home exercise program. The key elements of the home exercise program include:

Exercises to address postural alignment that are specific to the individual
Neural tension exercises
Relaxation techniques
Aerobic fitness through use of a stationary bike, walking, or running program
Practice of ergonomic principles for all regular hand use
Strengthening of the hand intrinsics (lumbricals and interossei)

To reinforce the importance of the custom home exercise program and the sensory retraining at home, the patient is asked to engage in these activities at least 1–2 hours each day. The patient is also asked to perform mental rehearsal at least 30 minutes per day. The patient is required to keep a log of these activities. The use of the log has been shown to increase compliance.

Splinting

There are various approaches to the treatment of dystonia that use splinting. Splinting can be used to promote neuroplastic changes or simply as a compensatory technique (7,13,29,20,35).

Writing Splint

This is a compensatory treatment technique. A writing splint may be fabricated to encourage retraining the task of writing by using the proximal muscles for writing rather than the fingers and thumb (Figure 7.3) (17). Provision of a writing splint implies a time commitment from the patient for intensive practice in writing drills. The patient is asked to relearn writing movements by relaxing the offending fingers and thumb and using alternative, larger muscle groups. Time spent in intensive practice is required for the patient to be successful with this approach (15).

Immobilization by Splinting

Priori et al. (20) advocate hand immobilization as a way to promote inactivity-dependent plasticity changes. This approach is a simple, effective, safe, and inexpensive treatment for focal, occupational upper limb dystonias. Prolonged limb immobilization in nondystonic subjects causes the motor cortical representation of the immobilized limb to shrink. Other researchers have found that immobilization of fingers combined with rehab will improve focal dystonia. Limb immobilization prompts inactivity-dependent plastic changes to reverse the functional abnormalities present at the cortical level in focal dystonia. Immobilization helps normalize the abnormally enlarged cortical representation of dystonic muscles. Priori et al. postulated that by removing all motor and sensory input to the limb, the cortical map will reset to the previous

FIGURE 7.3 Example of a custom-designed writing splint.

normal topography. The idea is that immobilization will cause a "fading" of inappropriate movement combinations typical of dystonia, so that retraining can begin on a relatively normal system.

The affected hand is immobilized in a custom thermoplastic splint, in the position of function, for 4–6 weeks (Figure 7.4). The immobilization of the wrist, thumb, and fingers is continued for 24 hours, 7 days a week. The splint is removed one time per week for 10 minutes for hygiene purposes. Four weeks later, the patient is allowed to begin controlled use of the hand in daily activities, but only as long as normal voluntary movement is demonstrated. This time allowed for controlled use of the hand is progressed over time at the following rate:

> 10 minutes per day during the first week
> Next 3 weeks, 30 minutes per session, three times per day
> Second month, one hour per session three times per day (musicians are allowed to begin practice of regular beginner's drills)
> After 3 months, patients can practice as often and as long as they wish but are instructed to stop as soon as they feel muscle fatigue or discomfort (20)

Initially after splint removal, it is common for patients to report clumsiness, unusual sensations, and the feeling they do not have normal control of the hand. Some report clumsiness to the level that they cannot complete simple individual finger movements. There is also weakness of involved muscles and occasionally edema and joint pain. However, with Priori et al.'s subjects, no contractures resulted, and within one week the joint pain and edema had disappeared, weakness was less, and dystonic postures had decreased. No

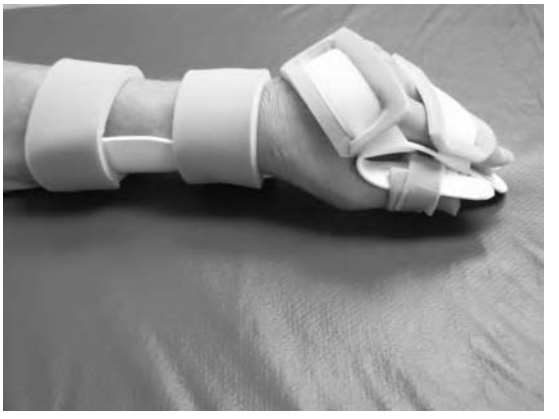

FIGURE 7.4 An immobilization splint.

patients had worsening of their dystonia with the immobilization, and there were no persistent adverse effects (20).

Priori et al. noted that patient's motor performance increased and was sustained longer than his 24-week follow-up, reflecting that motor learning had occurred (20).

Splinting for Sensory Trick

Jankovic has cited use of a splint to substitute for a sensory trick. This technique suggests trial and error by the therapist based on information from the extensive initial assessment (7). For example, a splint on the hand may be used to provide specific sensory input. The input is applied to the area that the patient has identified as being responsive to touch and, in the past, used successfully as a sensory trick to decrease the dystonic postures.

Sensory Motor Retuning

Sensory motor retuning was formerly referred to as constraint-induced movement therapy with the focal hand dystonia patients (36). In this approach, Candia (37) suggests an alternate method to promote normalization of the cortical map. Rather than immobilizing the dystonic fingers, the fingers that are used most to compensate for the dystonia are immobilized. The immobilizing splint is applied to the finger or fingers in a characteristic resting posture that they would use on their instrument. This is followed by intense repetitive exercises (sensory-motor retuning) of the dystonic fingers. This research was done with musicians (37,38). The sensory motor retuning treatment regimen is outlined in Table 7.2.

At the end of the supervised treatment course of 8 days, the patient is given the splint and asked to practice the exercises while wearing the splint for one hour daily for one year. The patient is instructed to also engage in unsplinted musical practice for 10% of what had been their customary time prior to the onset of dystonia. This is increased by 10% each month if motor control does not deteriorate. This method of treatment was found most effective for pianists and guitarists (37,38).

The basic principles are:

The most dystonic finger is not splinted

The main compensatory finger (or fingers) are splinted similar to the normal resting angle on the instrument

TABLE 7. 2 Sensory Motor Retuning (SMR)

SETS	DURATION	ACTIVITY
5	10 min exercise/2 min rest	• Apply splint • Sequential movement of free digits into extension in ascending then descending order • Example: If small finger is immobilized, movement sequence of the fingers is index, middle, ring, middle, index • Pace is set by a metronome; begin at a medium tempo, then alternate tempo throughout exercises
1	10 min	• Rest
4	10 min exercise/2 min rest	• Repeat the sequential movement of free digits into extension in ascending, then descending order
1	40 min	• Splint removal • Rest
2	15–30 sec play/2 min rest	• Play instrument without splint
IF TIME PLAYING INSTRUMENT IS DYSTONIA-FREE, PROCEED TO THE NEXT SEQUENCE.		
1	0–15 min play	• Play instrument without splint • Complexity and duration decided by OT
1	5 min	• Rest
5	5 min	• Apply splint • Sequential movement exercises continued

Exercises are performed with all the unsplinted fingers

The speed of movement of the dystonic finger moving with the other free fingers is increased and then decreased with progressively more exacting requirements

Daily practice with familiar music is valuable for motivation and to transfer the improved "exercise" movement to the target task movement

Practice is intensive but not to the point of excessive fatigue

A home exercise program, with daily drills, is important for maintenance and continued improvement (37,38)

A comparison of the Byl, Priori, and the Candia treatment approaches can be found in Table 7.3.

Development of Sensory Tricks

A sensory trick is a tactile or proprioceptive input to a particular area that results in a decrease in the muscle contraction severity or can even stop the

TABLE 7.3 Comparison of Treatment Approaches for Focal Hand Dystonia

	MECHANISM	TIME FRAME	SPECIAL CONSIDERATIONS
BYL	Sensory discrimination retraining	1–1.5 hours, 1–2 times per week plus home exercise program 3–6 months	Time intensive Cognitively and attentively strenuous
PRIORI	Immbolization	4–6 weeks, followed by slow, graduated return to use	No use of affected hand Possible skin or hygiene challenges
CANDIA	Sensory-motor returning	8 days in clinic Home exercise program for 1 year	Strict protocol-driven Compliance with time-limited practice drills

contraction (13,19). This is reported in up to 64% of patients with cervical dystonia and has been reported in focal hand dystonias as well. Many patients present with sensory tricks that help them to control their dystonic posturing. However, when not already in use, the OT may assist in the development of these tricks.

Common sensory tricks include touching the chin or lips to reduce dystonia of the mouth during feeding; touching the hand with the unaffected hand to help with writer's cramp; and pressing on the hips to assist in postural control and reduce trunkal dystonia. With musician's dystonia, a sensory trick that is often effective is a touch near the abnormal moving digit. Splinting to substitute for a sensory trick has been discussed in the previous section on splinting.

Stress Management

Stress-management techniques should be incorporated into any chosen treatment regimen. This is supported in Lie-Nemeth's work regarding use of a multifactorial approach (19). Stress management should be addressed by two methods: physiologically based relaxation and behavior modification techniques. The goal is twofold—to promote muscle relaxation and to build elements of stress-free time into one's day.

Physiologically, relaxation techniques reduce muscle contraction. These relaxation practices involve deep breathing techniques, visualization, and progressive muscle relaxation. Meditation is another technique that patients find helpful in stress reduction (17).

FIGURE 7.5 Normal hand positions on a guitar.

For musicians, training to modify their playing style is warranted (13). The emphasis is on modification to a less stressful technique. A lighter touch with the fingers or a less forceful grip on the guitar neck are examples of this type of approach (39). The goal is to reduce tension and practice good ergonomics when playing (Figure 7.5). Musicians should also avoid rapid increases in practice time or intensity.

The patient must be instructed about the importance of building in respite time as a stress-management technique. The patient must be made aware of the importance of making this time-out a priority. Control of stress, tension, and anxiety are essential elements in the overall treatment of the patient with dystonia (19). Emphasis on leisure activities can be also be helpful in stress reduction. Active leisure activities such as walking, water exercises, and stationary bike riding are recommended. Healthy habits such as general body conditioning, strength training, flexibility, reinforcement of good posture, and practice of general ergonomic principles also contribute to this management of stress (19,40).

Anecdotally, patients have reported that aerobic exercise has helped them to manage their dystonic symptoms in the early years after onset. One gentleman reported that for 2 years, jogging 2–3 miles helped the stiffness in his hand and kept him functional and able to manage the symptoms before the disease progressed.

Adjunct Modalities

Functional electrical stimulation can enhance and facilitate functional movement, particularly after Botox injections. For example, after Botox injections

are used to quiet the wrist flexors, FES can be applied to the wrist extensors to facilitate functional control over wrist positioning (41).

Biofeedback can be used to identify and relax overly active muscles and to increase the power and activation of the passive ones. Biofeedback may also be used in conjunction with the physiologic stress-reducing activities described in the earlier section on stress management (17).

ADL Training

ADL dependence is rarely a problem with the patient with focal hand dystonia. However, if there are tasks that are affected, OT can address compensatory techniques so ADLs can be performed safely, in a more satisfying manner, and continue to be done independently. Often this involves identifying a change in position where the posturing is less severe. An example is the elevation or slanting of a table or writing surface to assist with writing.

Because fatigue is often a complaint of the patient with dystonia, ADL training will encompass instruction in energy-conservation techniques. Basic principles include rest at intervals; pacing of large projects; looking at daily routines to evaluate for efficiency and safety; and suggestions of less energy-demanding postures for the task performance.

Adaptive and assistive devices may be recommended to address specific ADL challenges. For example, an upright vacuum may be easier to handle than a canister. Use of an electric or a one-handed can opener may facilitate function with less frustration.

Summary of Focal Dystonia

Focal hand dystonia is one of the most disabling limb dystonias. However, with the use of techniques to reduce the dystonic posturing, coupled with intensive retraining of movement, partnered with traditional fitness concepts to facilitate musculoskeletal health, the patient can improve sensory processing and motor control of the hand.

Generalized Dystonia

Full body dystonia is a poorly understood disease of the basal ganglia and has both genetic and nongenetic varieties. These deep brain centers, among other

things, modulate the speed and control of voluntary movement and prevent unwanted movements. The person with generalized dystonia may try to initiate a simple hand movement only to find that the movement has triggered a cascade of unwanted movement from the shoulder, trunk, and leg. The contractions may be so strong that the individual cannot continue to move at all. In addition, many patients complain of significant pain and distress resulting from the prolonged contractions.

Age of onset is strongly related to dystonia type. Generalized dystonia usually occurs in childhood, first with lower limb involvement, that later generalizes to the entire body within 5 years (in contrast to focal dystonias, which are most common in middle age, and remain localized to a specific body area). Childhood-onset generalized dystonia is the most severe form (3,17).

Secondary dystonias occur from secondary causes such as exposure to toxins/drugs, other diseases such as Parkinson's disease and multiple sclerosis, congenital disorders such as cerebral palsy, and from actual injury to the brain. In these cases the dystonia is typically generalized and affects the entire body.

OT Management of the Patient with Generalized Dystonia

The role of the OT is a dynamic one when considering the needs of individuals with this chronic, progressive condition. Their needs will change as the disease progresses and the OT's clinical knowledge, skills, and judgment will be a valuable addition to the multidisciplinary team (42). In the early stages of generalized dystonia, the OT will promote adaptations that enable patients to continue to function much in the same manner as they did prior to disease onset. As the disease progresses into the middle stages, more compensatory techniques will be required. In later stages, intervention emphasis shifts again and often focuses on education and training for the caregiver and modifications to the environment.

The patient with generalized dystonia may present to the OT clinic at any stage of the disease process. Many of these individuals may have experienced mild symptoms for some time before an investigation of these symptoms was initiated. Even after seeking medical attention, their symptoms may not have been accurately interpreted and they may not have received an accurate diagnosis. The patient may have been rated by their physician using the Burke Fahn Marsden Scale or the Unified Dystonia

Rating Scale (43). Both scales may be obtained from the WE MOVE website (3). The therapist may also use these scales to score the individual's status at the time of the initial evaluation.

Each stage of the disease brings its own challenges and, therefore, its own treatment emphasis and approach. Physical, emotional, social, and financial factors present a significant treatment challenge for patients diagnosed with generalized dystonia. Providing appropriate intervention to meet the comprehensive needs of the patient at each stage is best met by the multidisciplinary team (42).

Based on the framework developed by the National Parkinson's Foundation Allied Team Training, assessment and intervention in progressive neurologic disorders may be more easily understood if there is a breakdown of the disease process into stages (4). One method to functionally stage those with generalized dystonia is through the use of the Global Dystonia Rating Scale (GDS) (43). This valid and reliable tool is simple and easy to use and provides a global picture of the patient's current functional status (the scale can be found at wemove.org). Of course, presenting symptoms and rate of disease progression will vary from person to person. However, the scale forms a foundation from which to discuss evaluation and treatment strategies.

For the purpose of this chapter, we will separate the stages of the disease process and treatment strategies by broad categories of mild, moderate, and advanced, based on the score of the GDS. Primary goals in physical management, occupational performance intervention, and emotional support will be listed for each stage. Although this model will provide guidelines for the occupational therapist, we caution that given the complexity of the impairment in this population and the multifactorial contributors to disability, the choice of treatment approaches must ultimately be made on an individual basis.

Early Stage

Evaluation—GDS Score: 0–50

Individuals in the early stage, as represented by the 0–50 GDS score, will often present to the clinic walking independently, but with some detectable gait changes. They usually report still being involved in all of their occupational roles, such as self-care, school or work, and sports or other leisure activities. They often report decreased speed with activities and report frustration due

TABLE 7.4 Core Evaluation Elements for Patients with Dystonia

Thorough history and interview
Posture and functional mobility
Movement pattern analysis
Coordination
ADLs and I-ADLs
Sensory testing
Occupational performance

to decreased efficiency with movement. Fatigue and some level of pain are common complaints.

Symptoms at rest are a hallmark sign of generalized dystonia and can appear in these early stages. Early in the disease process, the patient may display awkward posturing of one extremity, most likely a foot or a hand. External factors and task specificity are uncommon in generalized dystonia. It is rare for the patient with generalized dystonia to benefit from use of a sensory trick.

In the OT clinic, all patients' needs, no matter the stage or the age, can be approached by looking at the patient's occupational therapy profile:

Patterns of daily living
Interests and needs
Problems or concerns over daily life tasks
Priorities

Evaluation of the patient with a generalized dystonia diagnosis should begin with the suggested categories identified in Table 7.4.

Management

Physical Management

A detailed outline of the findings from the assessment is crucial for this group. The problem list is then matched with an intervention and a home exercise program. Occupational therapy goals are as follows.

Promote Motor Relearning: Motor relearning programs, as described by Carr and Shepherd (44), require analysis of the task and the individual's performance. Difficult or poorly performed components of the task are practiced, followed by practice of the full task. Then the training is transferred into varied situations. Morris (6) suggests the following strategies to promote motor learning:

Break up long movement sequences into separate steps (not unlike the strategy proposed by Carr and Shepherd)
Perform one task at a time
Mentally rehearse before doing
Attend throughout the task
Use cues or tricks to trigger the movement
Practice in varied settings to generalize the task for motor learning

Age-appropriate games that stimulate a necessary skill set for a functional task can be used. For example, use of the Wii will require motor interaction to perform as well as motivation to succeed. Other forms of physical activity, such as yoga or tai chi, may motivate the patient motorically as well as reinforce group activity.

Restore Function Following Botox Injections: Functional electrical stimulation may be used to facilitate movement of one muscle group after the opposing muscle group has been injected. For example, if the bicep is injected, FES can be initiated 7–10 days after the injection to facilitate the use of elbow extension in functional tasks and allow the therapist to promote reeducation.

Facilitate Normal Development in Children: Children with cerebral palsy and upper extremity dystonia are often unable to reach for objects, manipulate toys, feed themselves, or use assistive communication devices. Therefore, even small improvements in these reaching tasks have large benefits for the child. Sanger et al. found that Botox injections improved the speed of outward reaching in children with arm dystonia (45).

An added consideration theorized by Sanger et al. is that the relationship between hypertonia and development of motor skills is unknown and that reduction in hypertonia during early development could potentially lead to long-lasting benefit (45).

Maintain and Increase Aerobic Fitness: Aerobic exercise has been proven to be of benefit in other neuromuscular disorders (6). Aerobic exercise can be implemented for muscle reeducation and functional movement and should be defined by the patient's goals (e.g., walking or biking) and adapted to their postural limitations.

Occupational Performance Intervention

Occupational skills enhancement is the primary goal. Many of these patients are elementary to middle school age. For these patients, referral to a pediatric

TABLE 7.5 Ergonomic Checklist for Desk Activities

CHAIR
Chair height allows the feet to rest flat on the floor, or, if not, a footrest is used
The chair provides lumbar support, or if a lumbar roll is added, it does not sacrifice seat depth
The seat depth provides coverage for most of the thigh yet does not allow the feet to dangle
UPPER EXTREMITY POSTURE
The upper arm is able to stay in a relaxed position, close to the body
The elbow is positioned at a 90-degree angle
Both wrists are maintained in a neutral position
HEAD AND NECK POSTURE
Computer monitor height facilitates a neutral cervical position
The distance between the individual and the computer monitor does not necessitate a head-forward position

OT clinic is recommended. The pediatric clinic will provide age-appropriate intervention tools and an encompassing network of services and support for the family. Treatment should focus on the following areas.

Promote the Use of Ergonomic Principles in Daily Life Tasks: Use of ergonomic principles is strongly recommended to facilitate proper movement patterns and to help decrease the fatigue often reported by this group (Table 7.5). Neutral positioning is a goal for work at a computer station at a school, work, or home computer. These neutral postures are also important in the school setting at a desk as well as the study area at home. Supporting good postural alignment and promoting core support is instrumental in controlling fatigue and pain.

Maintain Involvement in All Occupational Performance Roles: Facilitate the patient's active engagement in their usual roles by identifying barriers and proposing solutions.

Emotional Support

Teach Stress-Management Techniques: Since stress has been shown to exacerbate the symptoms, guide the patient and family to adapt routines and habits to decrease the stress level of the patient during task performance (3,35). Old routines may no longer be appropriate. Assist the family with development of new ones.

Patients of any age can benefit from stress-reduction techniques such as guided imagery and visualization. Biofeedback can also be used for stress reduction. For young children, the therapist can instruct parents in calming

strategies involving tactile input, holding and rocking, and use of gentle vestibular input.

Facilitate Social Interaction: Group interaction is extremely important at this stage to promote social interaction and to encourage physical activity. A therapy play group may be sought if the child is starting to avoid play with others due to embarrassment. Adapted aquatics or summer camps for children with disabilities may be options to consider.

Middle Stage

Evaluation—GDS Score: 55–100

By this stage the occupational roles of the patient are being threatened by the impairments resulting from the disease process. The patient is generally showing motor performance fluctuations, which are exacerbated by fatigue and stress. There is a pain level change, generally with higher pain levels reported and more frequency of occurrence.

Again reference to the general OT evaluation in Table 7.4 is the basis for the initiation of the evaluation. At this stage, addition of a pain scale is needed. A simple and consistent way to measure pain level is with the FACES Pain Scale, which is easy enough for children to understand, or for adults, a Visual Analog Scale (46,47).

ADL assessment will be more in-depth at this stage (Table 7.1). Instrumental ADLs (I-ADLs) should be considered and include home-management skills such as shopping, meal preparation, cleaning, laundry, and child care. Interaction with the community is also assessed. This includes driving or use of public transportation or school buses for children, shopping, and recreational activities. For adults, the FIM is a detailed assessment which may be used to identify the ADL independence level (48). For children, the Wee FIM may be used (49). The PEDI is another assessment for children, which addresses ADL functional levels (50).

Postural assessment is covered in the referenced OT assessment but will be a more time-consuming part of the evaluation at this stage. Patients usually exhibit moderate deviations in posture which are predictable, intermittent, and increase with intention. A detailed assessment of postural changes will be required to adequately address treatment strategies that can promote functional performance (51).

Communication changes may present new challenges for the patient and present new opportunities for OT collaboration with the speech pathologist.

The OT role is to assist in evaluation for specific communication devices, to adapt the device for the patient, and to train in the most efficient use. Writing and use of a computer keyboard and mouse are also assessed.

Assessments of the environment become important at this stage. The school-based OT may be an excellent resource for the student in a classroom situation. Ergonomic principles can be used as the foundation for the evaluation. In a school setting, attention to seat assignment in the classroom is an important element of the assessment, since vision will be influenced by postural deviations. Taking these same assessments and management techniques into the home for study habits or into the workplace for an adult patient is extremely important to control fatigue and frustration and promote success in the occupational roles.

This stage is also characterized by an increase in levels of stress, anxiety, and depression. The concerns of the caregiver and other family members are important to assess. The level of physical assistance required has increased. There is also a higher level of supervision required with tasks in which the patient has retained their independence. There will be changes in quality of life for both the patient and the family members. Assessment scales such as the Quality of Life Scale or the Satisfaction with Life Scale may be used (33,52).

Management

Physical Management

At this stage, more medical interventions, such as Botox and DBS, are being offered to the patient. Therefore, these treatment changes necessitate more involvement by the OT. Objectives of treatment at this stage include the following.

Improve Posture and Range of Motion: Postural enhancement can be provided by weight-bearing activities, closed chain exercises, weight shifting, and crossing the midline activities. Gait belts may be useful to enhance safety while allowing a form of independent movement.

Although a baclofen pump is usually used for treatment of spasticity, Motta et al. reported that it can also be used effectively with dystonia in children with CP. Usually it is indicated in patients who have moderate to severe dystonia that prevents patient management or function (53). In this case, the goal of the OT is to promote improvement with posture and ease the management of the patient by the care giver.

After Botox injections or DBS, the role of the occupational therapist may be to address specific joint limitations. Serial splinting can be used in these cases.

Promote Relaxation: Instruction in diaphragmatic breathing can be used to decrease fatigue and to promote relaxation. Other techniques to enhance relaxation are biofeedback, guided imagery, visualization, and music and art therapy.

Occupational Performance Intervention

Increase ADL Independence: Performance of basic ADLs will be an issue for this population. Each area of self-care should be addressed as outlined in Table 7.1. Each of these areas carries an intrinsic assessment of mobility limitations. Before abandoning an activity because of the dystonia, modify the task, by applying the principles of task analysis. Suggest an early morning routine and consistent adherence to the steps. Assistive devices may help the patient to maintain independence in some areas of ADLs. For example, suggesting changes to clothing choices will ease the challenges. Some suggestions include elastic waists versus zippers, and a choice of Velcro versus buttons.

Management of instrumental I-ADLs will need to be addressed. Safety is a concern and must be addressed in moderate to advanced stages of generalized dystonia, as the patient may not be mobile enough to manage without renovation or an individualized home plan.

Facilitate Adaptation to the Environment: Adaptation to the physical environment at home, at work, and in school is a major goal at this stage of the disease. The key to maintenance of the patient's participation in their occupational roles is environmental changes to enhance function. Use ergonomic principles as the foundation for positioning. For the child, address school-based physical demands such as getting to class, carrying books, and use of a locker. Social activities and participation in physical activities will require adaptations, based on the therapist's judgment.

Emotional Support

Monitor the Patient's Function and the Caregiver's Response: Another role of the OT is as a monitor for the team. As we are assessing and treating function and discussing the role of the caregiver in ADLs, the OT may be the best person on the team to monitor changes in these areas. Making appropriate recommendations for referrals to other team members is essential.

Educate the Caregiver: The primary caregiver requires special attention at this stage. Their responsibilities have increased due to the heightened physical

and emotional needs of the patient. The caregiver is balancing many roles at this stage, such as parent, physical handler, emotional stabilizer, financial provider, as well as child advocate in the medical system. Collaboration with the psychologist and/or the social worker is imperative to address the stability of the family structure.

Advanced Stage

Evaluation—GDS: Score 105–140

At this stage, the patient's level of dependency for routine activities has significantly increased. The frequency of pain complaints will also increase.

A detailed report of a normal day for the patient and his or her family is helpful to address use of routines and adaptations. A full interview with the caretaker is essential in assessing the challenges with management of the patient's ADLs. Strong emphasis should be placed on the preservation of health of the caregiver, and the essential principles of safe patient handling and movement should be enforced. A valuable resource for the occupational therapist is the Assessment Form and Algorithms illustration (Figure 7.6) found at www.visn8.med.va.gov/patientsafetycenter (54).

Assessment at this stage includes a home evaluation. Going into the home is the best practice. Due to geographical location or funding restrictions, this is not always feasible. If it is not possible to go into the patient's home, providing the caregiver with a checklist and a list of specific questions will help the therapist gain the information needed to make recommendations for accommodations, for safety, and for equipment.

Management

To promote the patient's confidence and to provide success in small steps of the tasks, use simple goals. From there incorporate challenges to inspire confidence, motivation, and interest.

Physical Management

Prescribe Appropriate Wheelchairs: Ambulation is less functional at this stage; therefore, use of a wheelchair is often recommended. Wheelchair consideration may start in this stage or, if started earlier, will certainly change at this stage. The goal for wheelchair use is for postural alignment, support for ease in UE function, and for optimal independence in mobility. This

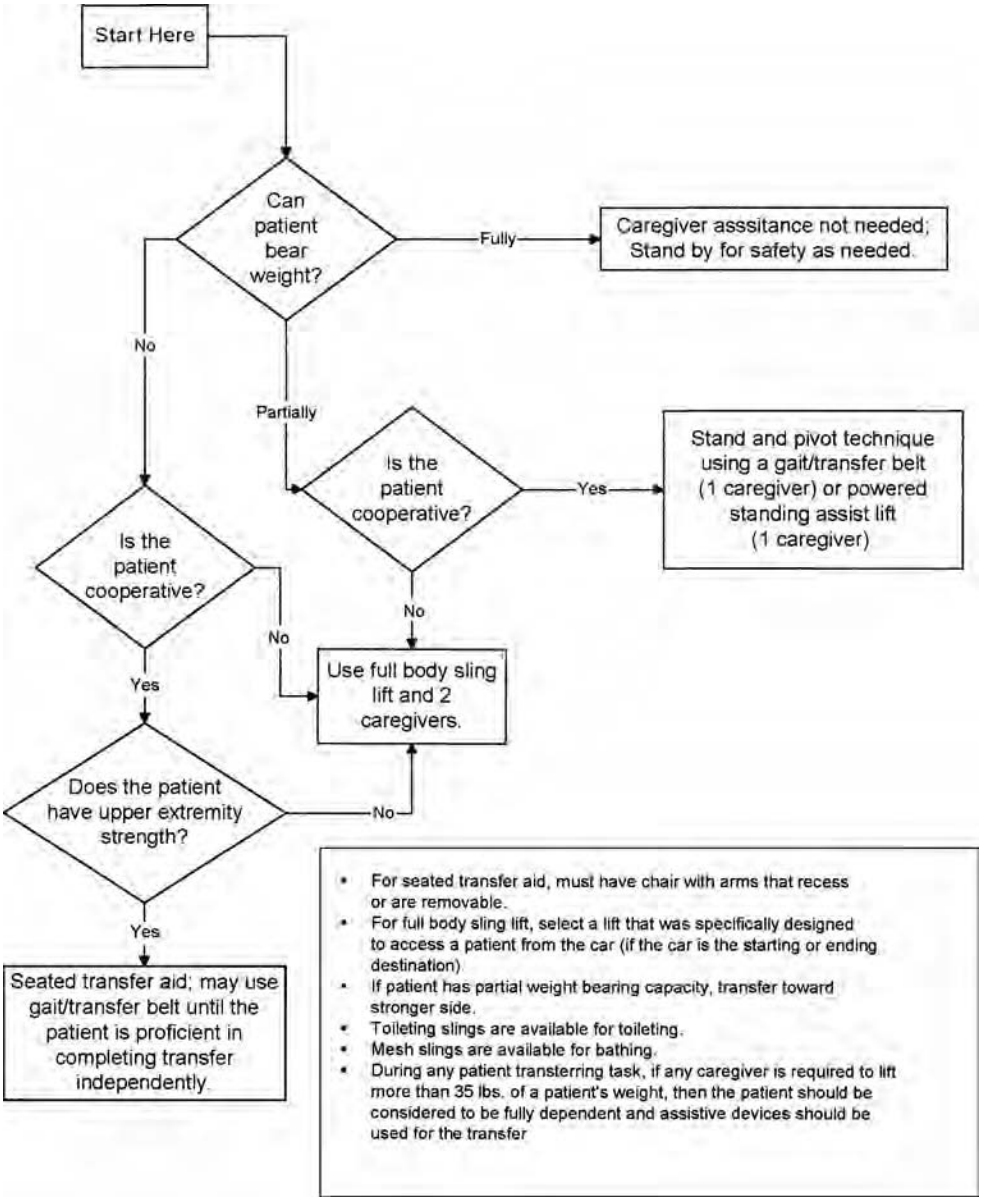

FIGURE 7.6 Transfer to and from bed to chair, chair to toilet, chair to chair, or car to chair. (Reprinted with permission from Audrey Nelson, Patient Safety Center.)

independence is important for the patient's emotional satisfaction and motivation to remain an active participant in their family, school, and community. A wheelchair assessment should identify the postural support needs of the patient. Seating systems can be customized to fit the needs of each individual. A seat cushion to distribute pressure is recommended. Collaboration

with the social worker is imperative for assisting with identification of funding sources.

Instruct the Caregiver in Safe Patient-Handling Techniques and Use of Proper Equipment: The patient will require moderate to maximum assistance in some or all transfers for ADL performance. Equipment will be needed to address the safety not only of the patient, but also of the caregiver. Care of the patient in the home will inherently involve risk to the family members. Transfers and other hands-on skills will challenge the family. Discussions with the caregivers regarding the risks involved should be reinforced routinely. Recommendations for equipment use are integral to the safety of the patient and to the caregiver. These recommendations include:

Friction-reducing devices may be of assistance for bed mobility challenges.
Rails on the bed may enhance bed mobility skills and increase the safety of independent movement.
Adaptation of the height of the bed for ease of assist during transfers or use of a twin bed if two caregivers are needed.
Raised toilet seats or toilet seats with grab bars may be considered.
Transfer tub seats, bathmats, grab bars, or mechanical bath chairs are options for increasing levels of safety during bathing for both the patient and the caregiver.
After a bath, use of a terry cloth bath robe for drying will be faster than use of a towel.
Mechanical lift devices such as floor models or ceiling-mounted models can be used to safely complete any transfers and can be a way to promote comfort and enhance dignity for the patient. An excellent reference for the therapist is www.visn8.med.va.gov/patientsafetycenter.

With all of the factors mentioned above, the occupational therapist has a major role in the provision of education to the caregiver. Instruction in specific handling techniques is imperative and safety elements should be reinforced regularly.

Promote Appropriate Positioning: General strategies for enhancing positioning include distal fixation, proximal stabilization, and/or constraining degrees of freedom.

Although use of such treatment approaches like DBS or a baclofen pump may not restore movement for independent use, often the caregiver will identify changes in the patient's posture and report that management of that patient for ADLs may be improved. Therefore, after these medical interven-

tions, the occupational therapist will need to adapt the treatment approach to incorporate these changes.

Specifically, positioning for feeding is extremely important to promote success, to increase the chances of stress-free mealtimes, and to enhance the safety of the process. Positioning to enhance proper posture with a chin tuck position and upright trunk to enhance digestion may be suggested. Another strategy is constraining the degrees of freedom by stabilization of the upper arm at 90 degrees, leaving the elbow flexion and extension as the primary movement pattern required for the feeding task. At this stage, feeding issues will require collaboration with the speech pathologist.

Occupational Performance Intervention

Facilitate the Patient's Involvement in All Occupational Roles: Facilitation of a level of active participation in roles at school, work, as a family member, and in the community will be challenging. However, the reward to the patient and their family members is enhancement of their quality of life.

Emotional Support

Train the Caregiver and the Patient in OT Strategies to Promote Relaxation: Use of the physiologic techniques and the behavior modification techniques previously covered in this chapter are valuable at this stage of the dystonia for both the patient and the caregiver.

Recommend Layers of Support: Collaborate with the psychologist and the social worker to suggest local organizations for support for the patient and the caregivers. Due to the rarity of the diagnosis, local support groups are often not specific to the dystonia diagnoses. However, based on the age of the patient, consideration of a stroke or a Parkinson's support group may be helpful. Caregiver websites may offer support to caregivers of children. Another excellent source of information and support for caregivers who are parents of young children with dystonia is the outpatient pediatric OT/PT clinic.

Summary

Dystonia is a disease of varied and sometimes unknown causes affecting different anatomic distributions and resulting in clinical manifestations that are unique to each individual. Therapeutic options must be tailored to meet the needs of the individual and are always chosen based on the age of the patient

and the part of the body affected. As part of a multidisciplinary team, the occupational therapist has a role to design a treatment plan that not only addresses the symptoms, but also helps the patient return to their life roles as successfully and joyfully as possible.

Ten Practical Tips

1. Get to know the members of your multidisciplinary team. One discipline alone cannot effectively manage the dystonia patient.
2. Assess how the dystonia affects the individual's quality of life.
3. Principles of postural support and ergonomic positioning should be established early in the treatment process.
4. Encourage relaxation strategies. The value can be significant.
5. Facilitate involvement in adapted aerobic fitness activities. Additional benefits in functional movement, stress relief, and social interaction will follow.
6. Explore the range of splinting options available to address FHD.
7. Educate and support the caregiver in cases of middle and advanced-stage generalized dystonia.
8. Avoid scheduling an OT treatment session immediately after a DBS programming session.
9. Dystonia is a rare diagnosis. Establish an OT network and encourage discussion of research articles and effectiveness of treatment techniques.
10. Remember, the OT's role is not only to treat the symptoms, but to assist the individual in maintaining involvement in their life roles as successfully as possible.

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8

The Role of the Psychologist in Dystonia

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> I hated looking in a mirror or at photographs because I was upset at the person looking back at me. I did not like that person. Just as the intenseness of my green eyes gazed back at me, so did the posture that I saw as grotesque and horrible. There were moments when I felt caged by the disease. . . . My mental anguish persisted, often unbeknownst to family, friends and colleagues.
>
> Beka Serdans, RN, MS, NP
> Founder of Care4Dystonia, Inc.

Dystonia can be emotionally devastating, as eloquently expressed by the above quote. To date, the unique psychological challenges faced by patients with dystonia have not been extensively addressed, and our current understanding of the psychological aspects of dystonia are drawn from two major sources: 1) the subjective experiences of individuals who are diagnosed with dystonia (and their family members) and 2) knowledge about issues and effective interventions with other chronic and/or progressive disorders. This chapter will provide an overview of current knowledge and practice and will highlight potential contributions of clinical psychologists to dystonia, including the role of psychologists in multidisciplinary treatment teams. While insights from related patient populations will be discussed, we will focus on issues unique to dystonia that may require special attention and services provided by psychologists.

The Psychology of Adult Dystonia

Psychological Factors in Dystonia

Like many chronic diseases, dystonia is associated with a unique constellation of physical, emotional, and psychological issues. These include frustrations experienced in the initial diagnosis/misdiagnosis of symptoms, psychological adjustment to the diagnosis, social stigma associated with physical changes, pain, impact on family and work, and a host of other issues somewhat unique to individuals with dystonia. As depicted in Figure 8.1, patient, disease and environmental factors, alone or in combination, can potentiate psychological distress in the dystonia patient.

Diagnosis and Misdiagnosis

Dystonia was not originally thought to have an organic basis (1), and misattribution of organic dystonia as psychogenic still occurs. One study reported that 25–52% of patients with idiopathic (primary) dystonia were initially

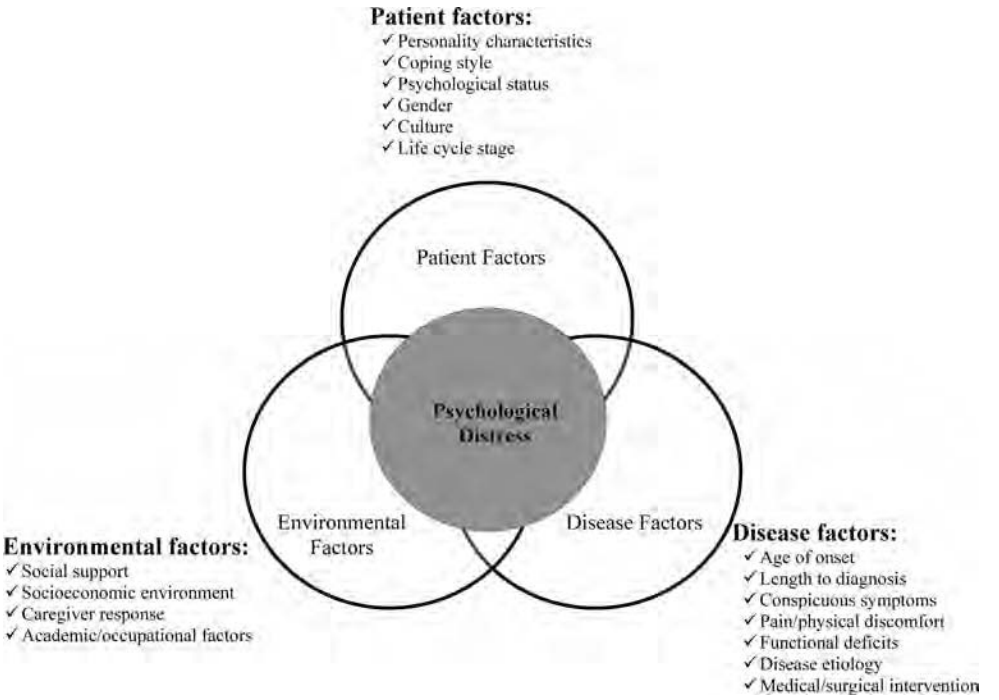

FIGURE 8.1 Psychological distress results from a complex interplay between patient-, environmental-, and disease-related factors, each of which can differ across patients. By considering these factors during psychological assessment and intervention, clinicians can individually tailor treatment to address each patient's unique needs.

diagnosed with a psychogenic movement disorder (2). Misdiagnosis and prolonged time to correct diagnosis may cause significant psychological distress for both patients and caregivers, and patients may continue to resent or distrust healthcare professionals even after being accurately diagnosed.

Social Stigma

A unique feature of dystonia is the conspicuous and often embarrassing nature of its primary symptoms, including body contortions and muscle spasms. Symptoms affecting the head, neck, and voice are particularly prominent in social interactions (1). Patients often feel self-conscious, unattractive, or apologetic (3); others may perceive them as *less* accountable, likable, trustworthy, attractive, and self-confident, and *more* "odd and different," reserved and piteous than individuals without dystonia (4). This raises a critical need for public education about dystonia as well as support for patients themselves.

Due to concerns about social stigma and embarrassment, individuals with dystonia tend to avoid social situations and perceive themselves as being avoided by others (3). Disfigurement may compromise social interactions, and negative feedback from others may perpetuate avoidant behaviors. Similarly, clumsiness may prompt patients to discontinue previously enjoyed activities. Avoidance eliminates the experience of negative social interactions, which further reinforces the withdrawal and isolation from others that can contribute to poor health outcomes (5). This phenomenon occurs in other physically conspicuous disorders such as essential tremor and can compromise quality of life (6). Also, body image concerns lead to negative self-cognitions (e.g., self-depreciation) that are linked to mood disturbances in dystonia (7). One study identified *body image concerns* as the main determinant of depression among spasmodic torticollis patients (8). Psychologists can aid in breaking this harmful cycle to promote more enjoyable social experiences, as discussed later.

Pain and Physical Discomfort

Aside from physical changes resulting from dystonia, pain is the most prominent symptom that contributes to psychological sequelae in this population, particularly in cervical dystonia (9). Pain is directly linked to depression and anxiety in a variety of diseases (10); fortunately, it responds well to psychological intervention, particularly behavioral therapy (11). Painful muscle spasms are also associated with sleep disturbance, fatigue, and mood dysregulation, all of which can worsen the symptoms of dystonia (12).

Loss of Independence

Dystonia may compromise one's ability to function independently, and, depending on life stage, this can result in different functional consequences for the adult "breadwinner" compared with a young child or a retired adult. Loss of independence and increased reliance on family members can create feelings of helplessness and worthlessness, and some disabled patients perceive themselves as "burdens." Furthermore, job loss can lead to economic distress, which is associated with psychological morbidity in chronic illness (13). Conversely, adequate financial resources to care for the individual with dystonia can buffer against distress associated with caring for a family member with this chronic disease.

Caregiver Response

Psychological distress can be thought of as "contagious" within the family. Caregivers of adult patients often experience emotional reactions to role changes, financial stress, and observing a loved one suffer from chronic physical discomfort and/or mood disturbance. Risk factors for caregiver distress include the nature and severity of the disease, uncertainty about prognosis and disease course, availability of financial and social resources, and caregiver characteristics such as age, gender, and relationship to patient (13).

Psychological Comorbidities

Depression is the most frequent psychological comorbidity in dystonia, although prevalence varies across dystonia subtypes. One study identified depression in 47% of cervical dystonia and 37% of blepharospasm patients (14). Others documented depressive symptoms in 72% and major depressive disorder in 25% of patients with spasmodic torticollis (15,16). In a study with mixed types of dystonia, clinically significant depression was identified in 37% of the sample, with no significant differences in rates or severity of depression between dystonia and other movement disorders such as Parkinson's disease (17). Importantly, depression was a primary predictor of quality of life in a large, community-based sample of dystonia (18).

Depression is linked to basal ganglia circuitry (19) and frequently antedates onset of physical dystonia symptoms (20). Therefore, it may result from the neuropathological effects of dystonia or a common genetic mechanism. One study reported increased risk for recurrent major depression among symptomatic and asymptomatic carriers of the DYT1 mutation (21), which

suggests that depression may be part of this genetic phenotype. Others have reported major depression in 28% of first-degree relatives of patients with cervical dystonia (15).

General and specific anxiety disorders are also common in dystonia, which may be attributed to worry about physical symptoms, dysregulation of neuro-affective processing, or both. Regarding general anxiety, reported rates vary from 25% in a mixed sample (22) to 53% in cervical dystonia (15). Social phobia was diagnosed in 56% of a sample of torticollis patients, (23) compared with 17.9% of a mixed sample (22). Interestingly, social anxiety is associated with younger age and depression but not with dystonia severity or duration (24). Panic disorder is also relatively prevalent among dystonia patients and was diagnosed in 29.5% of one sample of cervical dystonia patients (16).

Obsessive-compulsive disorder (OCD) is consistently linked to dystonia and has overlapping neurobiologic underpinnings (25). Antagonistic gestures (i.e., "sensory tricks") adopted by patients to alleviate dystonic symptoms can resemble compulsive behaviors; more traditional manifestations of OCD, including contamination fears and compulsive washing, are also seen (26). OCD symptoms often precede onset of spasmodic torticollis, which suggests that OCD, like depression, may be an early manifestation of basal ganglia circuitry instability. Notably, OCD is not strongly associated with DYT1 mutations (27).

Psychological Adjustment and Individual Differences

Adjustment to chronic disease involves changes in multiple life domains. The adjustment process is dynamic and continues throughout the disease course, particularly for progressive forms of dystonia that result in continuing loss of functional abilities and that may require increasingly aggressive treatments. Notably, successful psychological adjustment is an active process that involves many factors, including positive affect, effective coping strategies, mastery of adaptive skills, functional status, quality of life, and perceived meaning and purpose of life in the context of dystonia (28).

Adjustment in chronic illness is heterogeneous across individuals (29–31). Framed another way, not everyone reacts the same, and there seem to be a variety of factors that "moderate" how an individual uniquely adjusts to chronic disease. These include individual-specific factors like gender, personality characteristics (e.g., optimism, resiliency), how one typically deals with stressors, and previous history of psychological difficulties (i.e., depression, anxiety). Stage of life (i.e., the child vs. the young adult vs. the older adult) becomes important, as

well as social support from family members and friends, financial resources, and the suddenness and seriousness of the disorder (32).

Psychological Interventions

Psychologists can contribute to the clinical care of patients with dystonia in several ways. First, formal psychological assessment aids in the differential diagnosis of organic versus psychogenic movement disorder (PMD) by evaluating psychogenic risk factors, including mood disorder, precipitating events, social factors (e.g., relationship problems, abuse, secondary gain), and associated personality traits and coping styles. Periodic evaluation throughout the disease course can also contribute to the development of individually tailored treatment programs and medical decision making. Second, psychologists offer various behavioral treatments (i.e., therapies) that can help the patient with dystonia and their families learn to more effectively cope with their particular set of circumstances. This is important for a variety of reasons, one of which is that psychological stress can itself exacerbate dystonic symptoms (12). Some classic intervention methods used with adults are described below.

Foundations of Effective Therapy

At the core of any effective psychotherapeutic interaction is development of a psychologically safe, supportive environment for the patient. This includes rapport building through empathy, reflective listening, and validation. A strong therapeutic alliance is particularly important in the treatment of dystonia patients, who may have initially undergone an overwhelming array of challenges (e.g., misdiagnosis) before seeking psychological support. Individualized psychoeducation is also important in that it reduces faulty beliefs or expectations and helps the patient feel positively engaged in managing his or her own health. Psychoeducation also promotes a more realistic assessment of risks and benefits associated with treatments (e.g., deep brain stimulation surgery) and life planning (e.g., reproductive decision-making amongst patients with genetic forms of dystonia).

Supportive Psychotherapy

One goal of supportive psychotherapy is to help the patient increase "self-efficacy" to face life challenges resulting from dystonia. This is done by

helping the patient modify negative self-appraisals as well as mobilize personal and social resources. The therapist helps the patient formulate more compatible life goals and work toward *meaning making*. Meaning making involves making sense of a negative life event and identifying ways in which this negative event enriches one's life experience (32).

Supportive psychotherapy serves to build a repertoire of coping skills that can be strategically employed, such as avoidance strategies and approach strategies. While avoidance strategies (e.g., suppression, disengagement) may be helpful in managing acute crises, approach strategies are more effective long-term (32). Among the approach strategies, problem-focused coping (e.g., problem solving) is more common when facing controllable stressors, while emotion-focused coping (e.g., seeking social support) is more common when facing uncontrollable stressors (33).

Cognitive Behavioral Therapy

Cognitive behavioral therapy (CBT) focuses on replacing negative, automatic thoughts and managing stress and pain (34). Stress may result from catastrophic thoughts and abnormal illness beliefs that are perpetuated by an overemphasis on or misinterpretation of somatic symptoms. Examples of faulty beliefs in dystonia include anticipation of disease progression and assumptions about other people's perceptions (34). Using CBT, the therapist helps the patient identify negative thoughts, recognize contributing factors, and develop alternative cognitions. Contributing patient behaviors (e.g., excessive rest, avoidance of social situations) may also be examined and replaced with more adaptive alternatives (e.g., increased structured social activities).

Autogenic Relaxation

Relaxation techniques are highly effective in reducing stress and mitigating pain (11). Here the patient learns to use diaphragmatic breathing, progressive muscle relaxation, and mindfulness meditation. Visual imagery is also useful and can be tailored to a patient's interests (e.g., the relaxing beach scene). Primary benefits of learning these various relaxation techniques include reductions in abnormally heightened levels of physiologic arousal and improvements in focus and concentration. Moreover, relaxation techniques require the patient not only to master the procedure but also to recognize when and how to integrate it into daily life. This self-generated, or

"autogenic," aspect helps patients regain a sense of control and self-reliance. Increased self-awareness is also particularly beneficial for patients whose pain or dystonic symptoms are triggered or exacerbated by stress. In such cases, relaxation training should be augmented by activity pacing, which helps patients tailor their personal levels of activity and relaxation to minimize pain and symptoms.

Group Therapy

Support groups are often helpful in facilitating adjustment throughout the disease process (35). Support groups normalize the illness experience and provide a safe forum for the expression of emotions and concerns, thereby lessening psychological burden. Support groups for adult patients with other neurologic disorders, such as stroke, dementia, and multiple sclerosis, and their families are common in many medical centers and communities and could serve as models for dystonia patients and their families.

Family Therapy

Psychologists can also address the needs of caregivers and other loved ones. Interventions can range from extensive individual, couples, and/or family psychotherapy to more focused and short-term provision of psychoeducation and resources during a patient's hospital stay. Even just acknowledging the role of the caregiver, normalizing and validating their experience, and providing practical tips for self-care can have a profound impact.

Special Issues in Children and Adolescents

Age of onset for dystonia is bimodally distributed, with one peak at around age 9 and a second around age 45 (36). Early-onset dystonia is typically associated with more severe symptoms than its adult-onset counterpart, is more likely to progress from focal to generalized symptoms, and often has genetic etiology (37,38). Psychological factors and therapeutic targets in children and adolescents therefore warrant special attention.

Psychological Factors in Pediatric Populations

Children with dystonia face particular challenges that are complicated by 1) the often severe, progressive, and hereditary nature of early-onset

dystonia; 2) complex, reciprocal interactions between psychosocial, academic, and neurobiologic development; and 3) the pivotal role of family systems. To our knowledge there are no investigations into the specific psychological sequelae of childhood or adolescent dystonia. In the following sections we discuss some of the major challenges facing children with progressive and chronic illnesses such as dystonia. Obviously, every child and his or her family are different. Importantly, parents, extended family, and the community at large can work together to modulate the influence of these factors on an individual child.

Attachment and Psychosocial Development

Chronic and prolonged adverse medical events magnify a child's sense of helplessness and compromise feelings of safety, predictability, and trust. Children may become overly dependent on caregivers or generally distrustful of parents and authority figures, resulting in blunted development of emotional insight and self-regulation (39). Poor emotional self-regulation increases children's risk of clinically significant aggression, anxiety, depression, dissociative disorders, and substance abuse (39,40). In the case of complex chronic illnesses such as dystonia, children and adolescents may also feel concerned or guilty about the effects of their illness on parents and siblings.

School: Social and Academic Considerations

Physically limiting and socially stigmatizing symptoms of dystonia are often prominent in children, who must face hurtful teasing from peers. Physical symptoms can also interfere with the child's performance on relatively simple, mundane tasks during school. For example, hand contortions and spasms make it difficult to write and to keep up with in-class note taking, assignments, and timed exams. Secondary factors such as pain, fatigue, and social or emotional distress may indirectly compromise school performance by impeding a student's ability to focus, concentrate, and process information.

Potentially Traumatic Events

Medical visits, surgical procedures, and hospitalizations can be particularly traumatic for young patients, and a small number of proposed models attempt to address the specific psychological effects of medical illness in children and adolescents. For example, the National Child Traumatic Stress Network (2003)

defines pediatric medical traumatic stress (PMTS) as "a set of psychological and physiological responses of children and their families to pain, injury, serious illness, medical procedures, and invasive or frightening treatment experiences." The extent to which an event is "traumatic" depends on the child's subjective perception, and this is moderated by preexisting factors (e.g., preexisting psychopathology or family systems dysfunction), parental factors (e.g., parental response during the event; parental psychopathology), and/or characteristics of the event (e.g., perception of life threat; length of hospitalization) (41).

Models of PMTS are primarily based on evidence from acute injuries such as head trauma, illnesses such as cancer, and surgical procedures such as organ transplantation. Such models provide helpful conceptualization but may not fully account for psychological factors in children with chronic, degenerative disorders, let alone the unique experience of pediatric dystonia. The psychology of early-onset dystonia is likely best described as complex and multifactorial, including a tremendous amount of both inter- and intraindividual variability. What is clear is that the diagnosis of dystonia sets up a series of stressful events and experiences for the patient and his or her family and that these stressors need to be carefully and differentially addressed in light of age-related differences.

Caregivers and Siblings

Parents and siblings of chronically ill children or adolescents experience significantly elevated levels of psychological distress comparable to or exceeding those of the sick family member (41–43). Chronic stress also increases risk of compromised immune function and cardiovascular disease, and may even accelerate aging at a cellular level (44).

Illness in a child can cause siblings to develop negative responses. These may include guilt, helplessness, anger about attention paid to the ill child, and fears about their own health and security. Common caregiver concerns in childhood illness include changes in parental roles, feelings of helplessness, anxiety about unknown medical and psychological prognosis, and fear of potential loss or separation from their child (45). Stress and lifestyle changes also compromise self-care. For example, caregiver sleep quality was found to mediate the relationship between child health and caregiver depression (46).

The often hereditary nature of dystonia also contributes to caregiver and sibling response. Parents may blame themselves or their spouse for genetically

predisposing their child to this devastating illness. Anxiety regarding sibling health should also be expected, including heightened vigilance about subtle physical symptoms. Even genetic testing cannot provide clear resolution due to the low and variable penetrance of mutations such as those implicated in DYT1, or Oppenheim's dystonia (47). Moreover, even asymptomatic DYT1 mutation carriers may demonstrate nonmotor symptoms, including early-onset recurrent depression (21).

The mental health of caregivers and siblings also directly influences pediatric patients. A child's response and recovery is closely related to caregiver response and family functioning (39). Again, there is no dystonia-specific research on this subject, but it is safe to assume that these factors play an important role in the gestalt of the dystonia experience.

Psychological Intervention for Pediatric Populations

A recent meta-analysis found that psychological intervention in children with chronic illnesses is generally effective (48), and many of the same principles of psychotherapeutic intervention in adults can be applied to pediatric populations. However, children's conception of illness varies with age and with emotional and cognitive maturation. Intervention must therefore be tailored to the unique and age-specific needs of children, adolescents, and families of young dystonia patients. Psychotherapy should be flexible and should take into consideration developmental and social context, in addition to the role of ongoing exposure to related stressors (49).

Attachment and Psychosocial Development

To address concerns about emotional and psychosocial development, it is particularly important to establish a positive, supportive, and safe therapeutic environment. Depending upon the age of the child, this can be fostered via games, drawing, and activities that encourage silly/goofy, uninhibited behavior. Integrating predictable routines within therapy may also enhance perceived safety, particularly for children whose lives are marked by unpredictability. Encouraging patients to make choices during therapy (e.g., which game to play) is a simple way to help the patient feel safe expressing opinions free of judgment or criticism. This also helps shift perceived lack of self-control and self-competence.

Emotional awareness and regulation is another important component of therapy in this population. This includes 1) increasing emotional vocabulary, 2) learning to identify and differentiate between emotions, including awareness of a patient's own internal experience and interpretation of others' emotions, and 3) developing strategies for appropriately expressing and effectively coping with negative emotions. Emotional expression may be quite difficult for some patients, and play-based therapy can be useful in these situations (e.g., use of puppets, imaginary characters, story telling). Building skills to cope with teasing by others or social rejection by peers can also be a needed component of treatment.

Caregivers can enhance the effectiveness of in-session exercises. For example, parents can be engaged in treatment by helping children complete their therapy homework and/or asking their child about what he or she did in therapy. Caregivers should also be coached on how to provide appropriate praise and support at home, including encouraging appropriate emotional expression.

School: Social and Academic Challenges

Psychologists, teachers, and parents should be aware of potential social stressors for the child with dystonia, particularly at school. Pediatric patients are often reluctant to talk about teasing, and it is important to find a balance between encouraging communication and patiently waiting until the child feels comfortable enough to talk. The methods discussed above apply to this topic, and play techniques, such as talking about imaginary characters, making up stories, or playing with dolls or puppets, may be particularly useful.

Psychologists are also valuable informants for developing individually tailored academic accommodations. For dystonia, this may include recommendations such as computer training in anticipation of potential declines in spoken and/or written language and flexible time provisions for exams and in-class assignments. Psychoeducation for classmates, teachers, and other members of the school community is also appropriate.

Potentially Traumatic Events

To the extent that dystonia can be conceptualized within the context of medical traumatic stress, models of PTMS highlight areas for potential intervention. For example, variability in perceived traumatic value (41) suggests that psychotherapy before, during, or immediately following a potentially traumatic event may mitigate a patient's subjective experience of the event as

"traumatic." Guided imagery and other relaxation strategies can be individually tailored to younger patients for this purpose.

Caregivers and Siblings

In addition to the methods discussed earlier, caregivers and siblings of pediatric patients are particularly vulnerable to psychological distress, and psychotherapy is strongly encouraged. Also, the National Child Traumatic Stress Network has a variety of resources and handouts for parents and professionals, all of which are available for free at www.nctsnet.org. In siblings, parents and clinicians should carefully monitor for signs of psychological distress, and many of the same issues and treatment approaches recommended for patients similarly apply to healthy siblings. Within the pediatric cancer community, there have been numerous examples of group-based interventions for siblings, including treatment camps, and these seem to be quite effective in improving participants' anxiety and perceived levels of social competence and social acceptance (50,51). Similar group treatments may benefit siblings of children with dystonia, but to our knowledge this has not yet been tried.

Neuropsychological Aspects of Dystonia

Dystonia is a primary motor disorder and is not associated with significant intellectual impairment or marked cognitive decline (52). However, mild attentional-executive deficits have been noted (15). Deficits likely result from basal ganglia dysfunction and frontal-striatal involvement and may predate dystonia onset (53). In fact, nonmanifesting carriers of the DYT1 mutation demonstrate subtle abnormalities, including impaired motor sequence learning (54) and different patterns of brain activation (e.g., less prefrontal involvement and compensatory cerebellum activation) during motor learning (55). Other controversial findings suggest that patients with dystonia often exhibit preonset ADHD (56). However, attentional problems are commonly found in patients with disease-related pain and/or fatigue.

Cervical dystonia (CD) has also been associated with selective impairments in spatial processing, including mental manipulation of personal space (57), allocentric or egocentric navigation, and body-centered spatial perception (58). These results are likely related to persistent effects of abnormal head posturing of patients with CD.

Neurocognitive Deficits in Children and Adolescents

There is very little research on the neurocognitive features of dystonia in children or adolescents, with the exception of a few case studies and inclusion of pediatric patients in studies of DBS. For example, a recent report (59) described a 7-year-old boy with dystonia whose drawings of healthy family members consistently featured dystonic postures, highlighting the role of perception in dystonia and potential utility of neuropsychological evaluation. Subclinical sensory deficits were recently demonstrated in both symptomatic and asymptomatic individuals with the DYT1 mutation (60). Although participants were all adults, this study suggests that the mutation itself may affect sensory processing. Future research must address the neurocognitive consequences of DYT1 mutation in younger individuals. It is also likely that there are interactions between dystonia, neurodevelopment, and academic achievement, though these also have not yet been characterized.

Cognitive Side Effects of Medication

Medications used to treat dystonia have potential cognitive side effects. For example, anticholinergic medications (e.g., trihexyphenidyl) have been associated with deficits in visual sustained attention, verbal memory, and processing speed that were not better explained by depression or anxiety (15,61).

Potential Roles for Neuropsychologists

Periodic neuropsychological evaluations may aid in monitoring medication-related cognitive side effects, developing individually tailored medical and psychological treatment plans, and identifying compensatory strategies for occupational and functional performance. For pediatric patients, neuropsychological evaluation is also helpful in developing individualized school accommodations. Note that it is important to consider the likely need for repeat assessment when choosing which tests to administer (e.g., measures with multiple/alternate forms can minimize risk of practice effects). Children and adolescents are most likely to require repeat assessment as they progress through school, which raises additional issues. Age ranges accommodated by normative samples differ across tests and should factor into

test selection. For estimating intellectual functioning, the Wechsler Abbreviated Scale of Intelligence (WASI) has been validated across the widest age range (6–89 years) but does not include the same extensive selection of subtests as the other Wechsler Tests of Intelligence (i.e., WPPSI, WISC, WAIS). Therefore, it is important to consider the individual needs of each patient to determine the ideal approach, which may include a combination of measures (e.g., WASI plus select subtests from the age-specific test version). Additionally, due to the often rapidly progressive nature of early-onset dystonia, pediatric patients often face progressive physical limitations that affect performance on neuropsychological tests. It is therefore necessary to adapt test selection and administration accordingly, such as choosing multiple-choice versions of tests. For severely affected patients, unstandardized administration (e.g., improvising multiple-choice response options on tests without validated multiple-choice administration) may be the only option.

Deep Brain Stimulation: The Role of Clinical Psychology and Neuropsychology

DBS presents a host of unique opportunities—and challenges—for mental health professionals. Psychological issues have been explored in the context of DBS for Parkinson's disease (62,63), many of which are relevant to DBS treatment of dystonia as well. However, it is also likely that there are clinical, etiologic, and/or epidemiologic features unique to dystonia that should be considered. In the following section we will highlight potential roles for mental health professionals both before and after DBS surgery. Note that there may also be opportunities for mental health professionals to assist intraoperatively, including administration of relaxation exercises and/or guided visual imagery immediately before, during, or following DBS-related procedures (e.g., lead implantation, programming).

Preoperative Screening and Preparation

DBS is clearly an invasive procedure and requires postoperative medical compliance. Moreover, this procedure is still relatively new, particularly for dystonia. Therefore, preoperative psychological evaluation may provide valuable information regarding patient and caregiver preparedness. Screening for DBS should evaluate the following domains:

Knowledge and expectations regarding:

- General concept of DBS, including broad understanding of medical procedure
- Medical risks and potential complications, both during and after implantation
- Variability of potential outcome
- Requirements for pre-, peri-, and postoperative compliance

Factors related to compliance or complication risk:

- Premorbid behavioral and/or emotional problems
- Insufficient motivation
- Inadequate social support
- Poor or maladaptive coping strategies
- Psychosocial stressors, including family, social, and/or financial distress
- Spiritual and/or cultural beliefs that may be incompatible with surgical and/or postoperative demands

If preoperative psychological evaluation raises concerns about patient or caregiver preparedness, prophylactic interventions are often beneficial. Examples include DBS-specific psychoeducation, training in relaxation and visual imagery, and psychotherapy focused on development of adaptive coping strategies.

Preoperative Neurocognitive Evaluation

At a handful of programs, including ours at the University of Florida, neuropsychological evaluation is already integrated into multidisciplinary screening for DBS (62). Cognitive and behavioral measures provide additional information about potential contraindications, including progressive cognitive impairment. Characterization of preoperative functioning also facilitates postsurgical monitoring of surgical complications and/or neurocognitive disease changes. Assessment should therefore include neuropsychological measures across all major cognitive domains, including global cognitive functioning, verbal and visual memory, expressive and receptive language, visuo-perception and visuo-construction, and executive functions such as set-shifting and planning.

Postoperative Evaluation and Intervention

From the standpoint of general, postoperative factors, psychological evaluation and intervention should focus on medical compliance, patient

expectations regarding effects of surgery, and psychosocial barriers to successful ongoing adjustment. On a positive note, research suggests that motor improvements subsequent to DBS are generally accompanied by improvements in quality of life and mood, both in the general DBS population (64) and in studies restricted to patients with dystonia (65,66). However, suicide rates are debatably high immediately following DBS in dystonia; rates vary across studies (e.g., 6/140 in one study [67]; 2/16 in another [68]) and are independent of surgical outcome, and reports of suicidal ideation or attempt are even higher (for a review, see Appleby et al., 2007[64]). Voon and colleagues recently conducted a multicenter study of several thousand patients with STN DBS for Parkinson's disease and found the suicide rate to be lower than the general parkinsonian population (69). The high rate, particularly in a single dystonia study, may be reflective of the failure to utilize a multidisciplinary team and to prepare patients for the process of DBS. More studies of suicide and dystonia DBS will need to be published. In our own anecdoctal experience of over 30 patients treated by a multidisciplinary team approach, we have yet to have a suicide attempt. Potential explanations range from psychosocial to neuropathologic contributions, but the precise etiology and predictive factors are not yet known. Clearly this topic warrants further attention, and clinicians should be vigilant about monitoring suicidal ideation, particularly in patients with preoperative mood dysfunction.

Postoperative Neurocognitive Evaluation

For dystonia, post-DBS changes in cognition are even less consistent and less well characterized than affective and psychiatric sequelae. Recent studies with Parkinson's disease patients who undergo DBS in the subthalamic nucleus (STN) suggest that some (20–50%) (70–72) patients experience subtle declines in several cognitive domains after surgery, including verbal fluency, processing speed and memory for lists of unrelated words, with the former being the most robust and replicable finding (62,63). Similar declines occur less frequently in Parkinson's patients with stimulation targeting the globus pallidus (GPi) (73). This has important implications for dystonia patients, for whom the GPi is the most common DBS target. Stimulation parameters are typically more intense for dystonia than for Parkinson's disease (74), but the relationship between stimulation intensity and neurocognition has yet to be determined.

A few recent studies have investigated neurocognitive changes after GPi DBS in dystonia (75–77). Combined, these studies suggest that GPi DBS for dystonia does not generally have a deleterious effect on neurocognition and in fact may improve performance. However, there is also significant variability in postoperative cognitive outcome, and one challenge for neuropsychologists will be to determine predictors of positive and negative sequelae. To that end, regular follow-up neuropsychological evaluations are essential for monitoring individual response to surgery. Other surgical targets and effects on cognition in dystonia have been less studied.

DBS: Special Considerations for Pediatric Patients

There is almost no empirical evidence regarding the unique aspects of pediatric DBS, particularly with respect to psychosocial, emotional, and neurocognitive factors. Yet psychological assessment and intervention before, during, and after DBS surgery is equally critical to the care of younger patients.

Pre-DBS

Research shows that presurgical anxiety is not only unpleasant, but may also increase a child's risk for postoperative pain, mood, and sleep problems (78,79). Psychologists can contribute to presurgical assessment by evaluating the child's knowledge, compliance, and contraindications; presurgical psychotherapeutic intervention can reduce anticipatory anxiety and enhance patient and family preparedness. Examples include individualized and age-appropriate education regarding surgical procedures and expectations and evaluation and treatment of surgery-related phobias (e.g., fear of needles). Presurgical relaxation training is also helpful and can be combined with the educational components of intervention. For example, therapists can work with patients and caregivers to develop a game plan for the day of surgery, including planned relaxation strategies. Discussing and practicing aspects of this game plan during therapy can enhance a patient's familiarity and comfort during surgery and help ease presurgical anxiety. All of these strategies are also applicable to caregivers and siblings, who may experience similar levels of anxiety and fear about their loved one's upcoming procedure.

Psychologists can also assess and make recommendations about social and academic considerations related to the timeframe of surgical intervention. Although medical factors clearly take precedence, there may be leeway in the

timing of DBS procedures that would allow for presurgical intervention to address psychosocial and educational factors. Similarly, psychologists can provide resources and suggestions for enhancing social and academic adjustment, including helping patients develop ways to respond to questions from friends and teachers about their surgery.

Hospitalization and Surgery

Inpatient psychotherapy can be very effective, particularly when the psychologist already has a therapeutic relationship with the child and family. Ideally, this stage flows seamlessly from outpatient treatment, and techniques taught and practiced in preparation for surgery can be administered by the same clinician. In some cases psychologists can also stay with the child during surgery, whether to administer relaxation strategies or simply to serve as a trusted and familiar face dedicated to the child's comfort and mood during the procedure.

Psychologists also serve as resources and support for family members during this time. Again, psychoeducation, empathic and reflective listening, and encouragement are the most important features of inpatient therapy. Caregivers may need reassurance and would likely benefit from a concrete list of tips/reminders. The National Child Traumatic Stress Network also has free downloadable tip sheets for parents (http://www.nctsnet.org/nccts/nav.do?pid=typ_mt_ptlkt). Tips include being patient, supportive, and understanding; openly discussing feelings; and practicing self-care. For young children, being left alone can be particularly scary, so caregivers are encouraged to do so as little as possible and, when unavoidable, to explain to the child why you are leaving and when you will be back. Adolescents have the same needs for support and encouragement but may be less open in the expression of these needs to their parents. However, the presence of parents in their hospital room and throughout the presurgical procedures provides comfort and reassurance to the adolescent facing DBS.

After Surgery

The National Child Traumatic Stress Network also has free tip sheets pertaining to adjustment after hospitalization due to injury or illness. Many of these tips are similar to those for hospitalization, including encouraging expression of feelings, being patient and supportive, and practicing caregiver self-care. Returning to normal routines is also helpful in that it engenders a sense of

stability, predictability, and competency. Similarly, children and adolescents should be encouraged to socialize with friends and to resume appropriate levels of independence. To that end, psychologists can provide ongoing psychoeducation for caregivers, including age-appropriate reactions to medical stress. For example, children may become clingy, fearful, or irritable and may have changes in eating or sleeping patterns. These are normal reactions but should subside within a few days or weeks. Behavioral changes that may warrant additional attention include avoidance of school and/or social activities, development of new fears, and fighting with friends and family.

Adjustment after DBS surgery may be further complicated by other factors. These include negative responses from classmates and friends, requirements for medical compliance and safety, and pain or complications associated with the DBS equipment, all of which may additionally interfere with a patient's academic motivation or focus. Also, the use of DBS in children and adolescents is less well validated than in adults. Appropriate outcome expectations are therefore less clear, and patients may encounter unforeseen adverse effects. Of primary concern is that DBS leads implanted before the patient has completed physical development may have to be adjusted or replaced as the child continues to grow. Therefore, unlike a child who undergoes curative surgery, children and adolescents who undergo DBS for dystonia have a more uncertain immediate and long-term outcome, which can exacerbate psychological adjustment.

Conclusions

The constellation of psychosocial and emotional risk factors discussed in this chapter may seem to paint a bleak picture, particularly in severe cases of dystonia. However, the nature and heterogeneity of these factors suggests that psychological assessment and intervention have tremendous potential as integrated parts of multidisciplinary dystonia treatment. DBS provides an excellent example of the multiple roles for mental health professionals. Psychological and neurocognitive assessment of DBS candidates provides valuable information regarding outcome-related risk factors, including comorbid psychopathology and cognitive deficits. Psychotherapy can specifically target many problem areas detected during assessment, and continual intervention before and after surgery can serve as a powerful, consistent source of support. Therapeutic approaches may include psychoeducation (e.g., developing more realistic expectations for DBS outcome),

supportive and cognitive-behavioral therapy (e.g., enhancing utility of adaptive coping strategies by replacing negative cognitions), behavior modification, and skills-based training (e.g., identifying and addressing factors that interfere with medical adherence).

Training in relaxation strategies may be particularly helpful for DBS, especially for children, adolescents, and those with significant presurgical anxiety. Psychologists can even provide support, reassurance, and relaxation guidance during surgery. On a final note, the mental health of caregivers, siblings, and other family members is also of paramount importance, particularly with pediatric patients and with severe and functionally limiting cases.

On a positive note, dystonia is not typically associated with extensive cognitive deficits or decline, and DBS does not significantly compromise most aspects of cognitive functioning. However, patients are vulnerable to executive dysfunction, consistent with frontal-striatal involvement. Moreover, the disorder is very heterogeneous, and cognitive sequelae of DBS may be variable.
Based on the above review, two clear themes emerge:

1. The dystonia experience is not merely one of motor dysfunction, but includes a range of factors that may place patients, caregivers, and other relatives at increased risk for psychological and/or functional deficits
2. Literature on the psychological and neurocognitive aspects of dystonia is still in its infancy. It is therefore vital that mental health professionals provide psychological and neurocognitive clinical services as part of multidisciplinary dystonia care and that we play a greater role in a more comprehensive characterization of this complex, multifactorial disorder.

Ten Practical Tips

1. Dystonia occurs in the context of psychosocial and family systems factors that should be taken into consideration in treatment planning. Psychologists are therefore valuable members of multidisciplinary dystonia treatment teams, and it is critical to develop and maintain effective collaborations with physicians, surgeons, and medical support staff.
2. Psychological assessment can contribute to treatment planning by providing information about the following:
   a. Psychological risk factors (e.g., coping style, preexisting psychopathology, personality characteristics)

   b. Potential barriers for medical compliance
   c. Environmental and psychosocial factors that may affect psychological adjustment (e.g., financial stress, social support, caregiver response, occupational factors)
3. Comprehensive neuropsychological assessment provides critical supplemental information for comprehensive psychological evaluation and should include tests of all major cognitive and psychological domains, including:
   a. General intellectual functioning
   b. Attention, concentration, and working memory
   c. Language
   d. Memory
   e. Visuo-perception
   f. Executive function
   g. Frontal-motor function
   h. Personality and/or mood

4. There are numerous ways in which psychological intervention can enhance patient adjustment, treatment compliance, and quality of life. Psychotherapeutic approaches that may be helpful include:
   a. Psychoeducation
   b. Relaxation and stress-reduction training
   c. Cognitive-behavioral therapy
   d. Group therapy

5. Patients who undergo DBS for dystonia may experience presurgical anxiety and/or postsurgical adjustment issues. Individually tailored autogenic relaxation strategies are particularly helpful in such cases.
6. Children and adolescents with dystonia face unique psychological and neurocognitive issues. In these cases, intervention should take into consideration age-related factors such as emotional and cognitive development, social and academic demands, and family systems. Specific areas that may warrant attention include:
   a. Medically related phobias (e.g., needles)
   b. Heightened anxiety before, during, and after medical and surgical procedures
   c. Stunted development of emotional awareness and self-regulation
   d. Concerns about health and emotional well-being of siblings and parents

   e. Overdependence on or reactive withdrawal from parents/caregivers
   f. Peer response, including teasing and withdrawal as the child's symptoms progress
7. Psychoeducational assessment in children and adolescents with dystonia can provide valuable input for development of individualized education plans (IEP) to adequately accommodate physical, cognitive, and psychological challenges for academic achievement. Test selection should take into account the likely need for follow-up assessments and progressively worse physical limitations. School accommodations that may be useful for these children include:
   a. Additional time for class assignments and exams
   b. Alternative response methods, including use of computers and other assistive technologies
   c. Psychoeducation for teachers and classmates
8. In cases with known or suspected genetic contributions, psychologists should be aware that patient and caregiver stress may be exacerbated by concerns about health of other family members and/or guilt about genetic lineage.
9. The psychological demands and impact of dystonia frequently affect caregivers, who may therefore also benefit from psychotherapy and tips for self-care.
10. Young siblings of children and adolescents with dystonia may experience psychological and academic challenges as well, including feelings of guilt, helplessness, anger about attention paid to the ill child, and fears about their own health and security. Psychotherapy can be effective in such cases.

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9

Psychiatric Considerations in the Dystonia Patient

Herbert E. Ward

Screening for disturbances of mood and perception comprise an integral part of the assessment of a patient with dystonia. Disturbances in mood may be widely variable and range from adjustment disorders to major depression and even mania. Disturbances in perception also occur in dystonia and range from subtle delusional states to frank hallucinations. Additionally there must be an assessment of anxiety symptoms that may yield treatable conditions such as obsessions, compulsions, excessive worry, or panic attacks. It is important to have a comprehensive psychiatric evaluation in all dystonia patients. Quality of life and overall treatment success may hinge on recognition of and appreciation for psychiatric illness in this challenging patient group. In this chapter we will review all of the potential psychiatric manifestations that may occur in dystonia and the potential role for the psychiatrist on the multidisciplinary/interdisciplinary care team.

Depression

A major depressive episode is characterized by depressed mood or anhedonia and a constellation of symptoms that often include disturbances in weight, appetite, sleep, energy, and concentration. Cognitive distortions are common and may include an inappropriate sense of guilt or suicidal ideations. Table 9.1 shows the Diagnostic and Statistical Manual of Mental Disorders, 4th ed. (DSM-IV) criteria for a major depressive episode. Untreated, depression can be a lethal illness. The one-year prevalence rate for major depression in the general population ranges from 2.7 to 10.3%. Lifetime prevalence rates range from 7.8 to 17.1% (1,2). Depression in dystonia is common and occurs at a frequency similar to that seen in Parkinson's disease and essential tremor (3).

Antidepressant medications have become the foundation for treatment of major depression, and the clinician faced with depression in a dystonia patient now has a broad range of therapeutic options. All antidepressants have an acute effect of increasing the availability of neurotransmitters in the central nervous system (CNS) synapse (serotonin, norepinephrine, dopamine). The earliest pharmacologic agents utilized on a regular basis were the tricyclic antidepressants and the monoamine oxidase inhibitors. These agents remain efficacious but have fallen out of first-line favor because of their side effect profiles (confusion, memory loss, urinary retention, blurred vision, orthostatic hypotension, etc.). With the introduction of the selective serotonin reuptake inhibitors (SSRIs) in 1989, treatment of depression has become safer and better tolerated. We now can selectively inhibit the reuptake of both serotonin and norepinephrine (SNRIs) as well as dopamine and norepinephrine (DNRI) with minimal postsynaptic effects (antimuscurinic, antihistaminergic, and α_1-receptor blocking). This is an important feature of these medications as the postsynaptic effects often mediate side effects. Additionally, we have one α_2-receptor antagonist, mirtazapine, that works by blocking the negative feedback within noradrenergic and serotoninergic systems to increase these neurotransmitters without affecting reuptake. We do use the old tricyclic antidepressants (which may be more efficacious but have higher rates of side effects) in some cases, but much less often now with the newer agents available (4,5). Table 9.2 outlines the antidepressant medications by subclass.

TABLE 9.1 DSM-IV Diagnostic Criteria for Major Depressive Episode

A. Five (or more) of the following symptoms have been present during the same 2-week period and represent a change from previous functioning. At least one of the symptoms is either depressed mood or loss of interest or pleasure:

   (1) Depressed mood most of the day, nearly every day
   (2) Markedly diminished interest or pleasure in activities
   (3) Significant change in weight or appetite
   (4) Insomnia or hypersomnia
   (5) Psychomotor agitation or retardation
   (6) Fatigue
   (7) Feelings of worthlessness or excessive guilt
   (8) Diminished concentration or indecisiveness
   (9) Recurrent thoughts of death or suicidal ideation

B. Symptoms cause clinically significant distress or impairment in social, occupational, or other important areas of functioning

C. Symptoms are not due to the direct effects of a substance or general medical condition

Source: Ref. 6.

TABLE 9.2 Antidepressant Medications

GENERIC NAME	TRADE NAME	USUAL STARTING DOSE (mg)	USUAL DAILY DOSE (mg)	FORMULATIONS
SELECTIVE SEROTONIN REUPTAKE INHIBITORS				
Citalopram	Celexa	20	20–40	10, 20, 40, L
Escitalopram	Lexapro	10	10–20	5, 10, 20, L
Fluoxetine	Prozac	20	20–40	10, 20, 40, L
Fluvoxamine*	Luvox	50	50–300	25, 50, 100
	Luvox CR	100		100, 150
Paroxetine	Paxil	20	20–60	10, 20, 30, 40, L
Paroxetine CR	Paxil CR	25	25–62.5	12.5, 25, 37.5
Sertraline	Zoloft	50	50–200	25, 50, 100
SEROTONIN-NOREPINEPHRINE REUPTAKE INHIBITORS				
Duloxetine	Cymbalta	30	60–90	20, 30, 60
Venlafaxine	Effexor	37.5	75–225	37.5, 75, 50, 75, 100
VenlafaxineXR	Effexor XR	37.5	75–225	37.5, 75, 150
Desvenlafaxine XR	Pristiq	50	50–200	50, 100
SEROTONIN MODULATORS				
Trazodone	Desyrel	50	75–300	50, 100, 150, 300
NOREPINEPHRINE-SEROTONIN MODULATORS				
Mirtazapine	Remeron	15	15–30	7.5, 15, 30, 45, soltab
DOPAMINE-NOREPINEPHRINE REUPTAKE INHIBITORS				
Bupropion	Wellbutrin	150	300	75, 100
Bupropion SR	Wellbutrin SR	150	300	100, 150
Bupropion XL	Wellbutrin XL	150	300	150, 300
HETEROCYCLIC ANTIDEPRESSANTS				
Amitriptyline	Elavil	25–50	100–300	10, 20, 50, 75, 100, 150
Amoxapine	Asendin	50	100–400	25, 50, 100, 150
Clomipramine	Anafranil	25	100–250	25, 50, 75
Desipramine	Norpramin	25–50	100–300	10, 25, 50, 75, 100, 150
Doxepin	Sinequan	25–50	100–300	10, 25, 50, 75, 100, 150, L
Imipramine	Tofranil	25–50	100–300	10, 25, 50, 75, 100, 125, 150
Maprotiline	Ludiomil	50	100–225	25, 50, 75
Nortriptyline	Pamelor, Aventyl	25	50–150	10, 25, 50, 75, L
Protriptyline	Vivactil	10	15–60	5, 10
Trimipramine	Surmontil	25–50	100–300	25, 50, 100
MONOAMINE OXIDASE INHIBITORS				
Selegeline transdermal	EMSAM	6	6–9	Transdermal doses: 6 mg/24 hours 9 mg/24 hours 12 mg/24 hours
Isocarboxazid	Marplan	10	20–60	10
Phenelzine	Nardil	15	15–90	15
Tranylcypromine	Parnate	10	30–60	10

L, liquid; CR, controlled release; SR, sustained release; XL or XR, extended release; soltab, orally disintegrating tablet.

*FDA approved only for obsessive-compulsive disorder and social anxiety disorder.

Source: Ref. 7.

Depression is the most common psychiatric disorder associated with dystonia (8). Heiman and colleagues found that the risk for recurrent major depression was increased in patients carrying the DYT1 gene mutation (9). Symptoms of depression often overlapped with motor symptoms and could be overlooked during clinic visits. The Beck Depression Inventory (BDI), which is a 21-item self-report questionnaire that takes only a few minutes for patients to fill out, can be a useful screening tool in the clinic. Psychometric properties of the BDI are well established. Scores range from 0 to 63. Cutoffs of 0–9 for nondepressed, 10–18 for mild to moderate depression, 19–29 for moderate-to severe, and >29 for severe depression are generally accepted by clinicians and researchers (10). Miller and colleagues found BDI scores > 10 in 37.5% of 83 dystonia patients seen within a movement disorders center practice. Additionally, they failed to identify a relationship between depression and severity of motor symptoms (3). Another study using DSM-III interviews identified a depressive syndrome in 40% of 28 dystonia patients (11). A study of 40 patients with cervical dystonia failed to reveal an association with one specific psychiatric disorder. Thirty-eight percent met diagnostic criteria for major depression, and 40% had anxiety disorders. No causal relationship was found between the movement disorder and psychiatric illness (12). The data for depression and anxiety in dystonia patients are compelling and argue in favor of a psychiatrist as an important member of the multidisciplinary team.

Social Anxiety Disorder

Individuals with social anxiety disorder (SAD) or social phobia experience distressing anxiety while under the scrutiny of others or when in potentially embarrassing situations. SAD may occur in dystonia. This disorder can generalize to any situation that involves appraisal by others. In its mildest form this type of anxiety is often described as shyness. However, for the individual with SAD the term shyness does not communicate the pain they endure when exposed to novel social interactions. In addition to the subjective experience of anxiety, the individual with SAD can experience intense autonomic arousal with muscle tension, and this may result in robotic movements, sweating that soaks clothing, and a tremor that renders handwriting nearly impossible. The Leibowitz Social Anxiety Scale can be helpful in quantifying symptom severity (13). Treatment usually involves a form of exposure

TABLE 9.3 DSM-IV Diagnostic Criteria for Social Anxiety Disorder or Social Phobia

A. A marked and persistent fear of situations in which the person is exposed to unfamiliar people or to possible scrutiny by others
B. Exposure to the feared social situation provokes anxiety
C. The person recognizes that the fear is excessive or unreasonable
D. The feared situation is avoided or endured with intense anxiety and distress
E. The anxiety significantly interferes with the person's routine
F. Duration at least 6 months
G. Fear or avoidance is not due to a substance or general medical condition
H. If a general medical condition is present, the fear is unrelated to it

Source: Ref. 6.

therapy with cognitive restructuring (14) as well as medication (15). More specifically, the SSRIs and SNRIs have become first-line agents for SAD (16) (Table 9.2). Additionally, the benzodiazepines have proven effective in reducing symptoms (17). Table 9.3 details the criteria for social anxiety disorder or social phobia.

Gundel and colleagues reported that of 116 consecutive torticollis patients, 41.3% had social anxiety disorder as a primary psychiatric diagnosis. This finding was not related to body image, severity of dystonia, pain, or other psychiatric conditions (18). Compared to the general population, this represented a 10-fold higher prevalence of social anxiety disorder among patients with spasmotic torticollis. In addition to primary social anxiety disorder, social anxiety can occur secondary to a physical disability (19). Dystonic symptoms are often exacerbated by stress, which can lead to a cycle that feeds back on itself in social interactions. Treatment for social anxiety thought to be secondary to dystonia should not be withheld just because it is not considered a primary diagnosis (20,21).

Generalized Anxiety Disorder

Generalized anxiety disorder (GAD) is characterized by excessive worry that cannot be controlled. Patients often seek relief from the somatic complaints that accompany the disorder. Muscle tension and soreness are common. Patients complain of fatigue, poor concentration, disturbance in their sleep,

and a waxing and waning sense of restlessness. Clinicians often approach these target symptoms as they appear without appreciating the cluster representing the disorder. The syndrome often responds to a single medication. More specifically, the SSRIs and SNRIs have proven efficacy in GAD (Table 9.2). There is a high comorbidity of GAD and depression. The advantage of these agents is that they will address both conditions. However, in patients who do not respond to an antidepressant, the benzodiazepines have proven effective. The disadvantage of benzodiazepines is that this syndrome is chronic in nature. We have one nonbenzodiazepine anxiolytic, buspirone, classified as a 5HT1A partial agonist, that has a similar onset as the antidepressants, has no risk of chemical dependence, but has no appreciable antidepressant properties. This can be tried in dystonia. Some patients will, however, require a more aggressive regimen including a benzodiazepine (4,5). Table 9.4 details the criteria for GAD from DSM-IV. Table 9.5 lists anxiolytic medications.

Lauterbach and colleagues have explored the relationship between movement disorders and anxiety disorders. In 28 subjects with either primary dystonia or Parkinson's disease, they discovered that GAD occurred in 25% of dystonia subjects in the cohort and that more commonly they occurred after the onset of dystonia (22).

TABLE 9.4 DSM-IV Diagnostic Criteria for Generalized Anxiety Disorder

A. Excessive anxiety and worry for at least 6 months about a number of events or activities

B. The person finds it difficult to control the worry

C. The anxiety and worry are associated with three (or more) of the following six symptoms:

1. Restlessness or feeling keyed up or on edge
2. Being easily fatigued
3. Difficulty concentrating
4. Irritability
5. Muscle tension
6. Sleep disturbance

D. Not secondary to another disorder

E. The anxiety, worry, or physical symptoms cause significant distress or impairment

F. Not due to the effects of a substance or medical condition

Source: Ref. 6.

TABLE 9.5 Anxiolytic Medications

GENERIC NAME	TRADE NAME	DOSE EQUIVALENCE (mg)	TYPICAL STARTING DOSE (mg)	TYPICAL DOSAGE RANGE (mg)
BENZODIAZEPINES				
Alprazolam	Xanax	0.5	0.25–0.5 tid	0.75–4.0
Alprazolam extended-release	Xanax XR	NA	0.5–1.0	3–6
Chlordiazepoxide	Librium	10	5–25 tid or qid	15–100
Clonazepam	Klonopin	0.25	0.25 bid	1–4
Clorazepate	Tranxene	7.5	3.75–7.5 tid	15–60
Diazepam	Valium	5	2–10 bid/qid	4–40
Lorazepam	Ativan	1	0.5–1 tid	0.75–3.0
Oxazepam	Serax	15	10–30 tid	30–120
NONBENZODIAZEPINE ANXIOLYTICS				
Buspirone	Buspar	NA	5–10 tid	30–60

Bid, twice-daily dosing; tid, three times per day dosing; qid, four times per day dosing; CR, extended release; NA, not applicable.
Source: Ref. 7.

Panic Disorder

Panic attacks are characterized by the sudden onset of intense fear and alarming physical sensations. Patients often describe a sense of impending doom or feel that death is imminent. Physical symptoms may mimic medical catastrophes and may include chest pain, shortness of breath, sweating, dizziness, or a choking sensation. Patients are often evaluated in emergency departments or undergo exhaustive workups and numerous specialty evaluations. As a result of the panic attack, phobic avoidance of places or situations can develop and severely limit activities and quality of life. Early and definitive treatment can aid in avoiding an erosion of one's quality of life as well as unnecessary medical procedures. Panic attacks can be rapidly blocked with benzodiazepines prescribed on a regular dosing schedule (Table 9.5). There is usually no place for PRN dosing of these medications in panic disorder mainly because there is no prodrome to signal need for the medication. With the exception of bupropion, all of the antidepressants are effective in treating panic. The SSRIs are the first line (Table 9.2). However, when

using antidepressants, starting doses must be a fraction of the starting dose for depression, and titration must be slow to avoid activation and worsening of panic. Buspirone is not effective for panic disorder. Clinicians will often use a combination of a benzodiazepine and an antidepressant. This combination allows for rapid control of panic while titrating the antidepressant up to a therapeutic dose (and waiting for its onset in 2–3 weeks or longer). Once therapeutic antidepressant levels have been reached and the panic attacks have been blocked, the benzodiazepine can be slowly tapered and discontinued. The use of an antidepressant for panic disorder has the advantage of treating comorbid depression if it is identified (4,5). Table 9.6 details the diagnostic criteria from the DSM-IV for a panic attack, and Table 9.7 outlines the criteria for panic disorder. It should be noted whether the disorder is *with* or *without* agoraphobia.

Obsessive-Compulsive Disorder

Patients with obsessive-compulsive disorder (OCD) are usually plagued with intrusive and persistent thoughts that produce intense anxiety. They are

TABLE 9.6 DSM-IV Diagnostic Criteria for Panic Attacks

A discrete period of intense fear or discomfort, in which four (or more) of the following symptoms developed abruptly and reached a peak within 10 minutes:

1. Palpitations, pounding heart, or accelerated heart rate
2. Sweating
3. Trembling or shaking
4. Sensations of shortness of breath or smothering
5. Feeling of choking
6. Chest pain or discomfort
7. Nausea or abdominal distress
8. Feeling dizzy, unsteady, light-headed, or faint
9. Derealization (feeling of unreality) or depersonalization (being detached from oneself)
10. Fear of losing control or going crazy
11. Fear of dying
12. Paresthesias (numbness or tingling sensations)
13. Chills or hot flushes

Source: Ref. 6.

TABLE 9.7 DSM-IV Diagnostic Criteria for Panic Disorder

A. Both (1) and (2):

1. Recurrent unexpected panic attacks
2. At least one of the attacks has been followed by 1 month (or more) of one (or more) of the following
   a. Persistent concern about having additional attacks
   b. Worry about the implications of the attack or its consequences
   c. A significant change in behavior related to the attacks

B. The panic attacks are not due to the effects of a substance or medical condition

C. The panic attacks are not secondary to another psychiatric disorder

Source: Ref. 6.

compelled to engage in behaviors to try to neutralize the anxiety. For example, the obsession may have a contamination theme and require excessive hand washing to lower the anxiety. This disorder can be debilitating. Obsessional thinking and the overwhelming need to perform rituals can completely consume the OCD patient's day, leaving no time for work or relationships. A combination of cognitive behavior therapy and medication is usually the best approach to treatment. The SSRIs are the mainstay of pharmacotherapy. One tricyclic agent, clomipramine, is indicated for OCD (4,5) (Table 9.2) Table 9.8 summarizes the DSM-IV criteria for OCD.

Mania

A manic episode is usually best characterized as an energized state in which thought production is increased, mood is elevated, and the need for sleep is decreased. Grandiosity and poor judgment often result in negative financial and behavioral consequences. Mood may be euphoric, irritable, or frankly hostile. The disturbance in thought can quickly escalate to a manic psychosis and can require emergency management to ensure safety of the patient and others. The antipsychotic medications are usually utilized to get manic excitement under control, and this may require parenteral dosing initially with transition to oral medications when stable. Mood stabilizers such as valproate, lamotragine, and lithium can be used as maintenance medications (4,5). Mania may occur in dystonia and must be carefully monitored for particularly following deep brain stimulation (23). Table 9.9 summarizes the DSM-IV criteria for manic episodes, and Table 9.10 summarizes the pharmacotherapy that may be used for mania.

TABLE 9.8 DSM-IV Diagnostic Criteria for Obsessive-Compulsive Disorder

A. Either obsessions or compulsions

Obsessions are defined by the following:

1. Recurrent and persistent thoughts, impulses, or images that are experienced as intrusive and inappropriate and that cause marked anxiety or distress
2. The thoughts, impulses, or images are not simply excessive worries about real-life problems
3. The person attempts to ignore or suppress such thoughts, impulses, or images or to neutralize them with some other thought or action
4. The person recognizes that the obsessional thoughts, impulses, or images are a product of his or her own mind

Compulsions are defined by the following:

1. Repetitive behaviors (e.g., hand washing, ordering, checking) or mental acts (e.g., praying, counting, repeating words silently) that the person feels driven to perform in response to an obsession or according to rules that must be applied rigidly
2. The behavior or mental acts are aimed at preventing or reducing distress or preventing some dreaded event or situation. These behaviors or mental acts either are not connected in a realistic way with what they are designed to neutralize or prevent or are clearly excessive

B. At some point during the course of the disorder, the person has recognized that the obsessions or compulsions are excessive or unreasonable

C. The obsessions or compulsions cause marked distress, are time-consuming, or significantly interfere with the person's normal routine

Source: Ref. 6.

TABLE 9.9 DSM-IV Diagnostic Criteria for a Manic Episode

A. A distinct period of abnormally and persistently elevated, expansive, or irritable mood, lasting at least 1 week

B. At least three of the following symptoms:

1. Inflated self-esteem or grandiosity
2. Decreased need for sleep
3. More talkative than usual or pressured speech
4. Flight of ideas or subjective experience that thoughts are racing
5. Distractibility
6. Increase in goal directed activity or psychomotor agitation
7. Excessive involvement in pleasurable activities that have a high potential for painful consequences

C. Symptoms are severe enough to cause marked impairment in functioning or there are psychotic features

D. Symptoms are not due to the effects of a substance or medical condition

Source: Ref. 6.

TABLE 9.10 Atypical Antipsychotic Dosing in the Treatment of Acute Mania

GENERIC NAME	TRADE NAME	STARTING ORAL DOSE (mg/day)	TITRATION SCHEDULE	TARGET DOSE (mg/day)
Aripiprazole	Abilify	15–30	15 mg/week	30
Olanzapine	Zyprexa	5–10	5 mg/day	10–20
Risperidone	Risperdal	1–3	1 mg/day	2–6
Quetiapine	Seroquel	100	100 mg/day	300–600
Ziprasidone	Geodon	80	40 mg/day	120–160

Source: Ref. 7.

Psychosis

There are multiple etiologies for psychosis, which, although it rarely occurs, can accompany dystonia. After determining that the patient is delusional or hallucinating, the next step is to determine etiology. Structural, metabolic, toxic, and infectious etiologies need to be considered in an effort to be sure that all identifiable and reversible causes have been addressed. While the search for a reversible cause is underway, symptomatic treatment is usually indicated. The newer antipsychotics, termed atypical antipsychotics, have come into favor because of better side effect profiles than older agents such as haloperidol and chlorpromazine. In patients with movement disorders, drugs that spare the basal ganglia dopamine receptors are preferred. More specifically, agents with low D2 antagonism have the lowest chance of worsening involuntary movements. Quetiapine has become popular for this indication. Dosing should be low initially and titrated up slowly with close attention to side effects (4,5). Table 9.11 lists the atypical antipsychotics.

Adjustment Disorder

An adjustment disorder develops as a maladaptive response to a stressor and is commonly encountered in the clinical practice of movement disorders and particularly dystonia. The magnitude of the stressor in the eyes of the clinician may seem out of proportion to the catastrophic impact the stressor has had on the patient. Stress-induced psychiatric symptoms can be variable, but are usually linked temporally with the stressor. Symptoms may involve depressed mood, anxiety, or a disturbance in the patient's behavior. Psychotherapy is the foundation of treatment for adjustment disorders.

TABLE 9.11 Atypical Antipsychotic Medications

GENERIC NAME	TRADE NAME	USUAL ADULT DAILY DOSE (mg)	PREPARATIONS	AVAILABLE ORAL DOSES (mg)
Aripiprazole	Abilify	15–30	T, L, DT, IM	5, 10, 15, 20, 30
Clozapine	Clozaril	200–500	T, DT	12.5, 25, 50, 100, 200
Olanzapine	Zyprexa	10–20	T, DT, IM	2.5, 5,7, 5, 10, 15, 20
Paliperidone	Invega	3–12	T	3, 6, 9
Quetiapine	Seroquel	200–600	T	25, 100, 200, 300
Quetiapine extended-release	Seroquel XR	200–800	T	200, 300, 400
Risperidone	Risperdal	2–6	T, L, DT, D	0, 25, 0.5, 1, 2, 3, 4
Ziprasidone	Geodon	80–160	C, IM	20, 40, 60, 80

T, tablet; C, capsule; L, liquid; DT, orally disintegrating tablet; IM, intramuscular injection; D, depot.
Source: Ref. 7.

TABLE 9.12 DSM-IV Diagnostic Criteria For Adjustment Disorders

A. The development of emotional or behavioral symptoms in response to an identifiable stressor occurring within 3 months of the onset of the stressor

B. Symptoms cause either of the following:

1. Marked distress that is in excess of what would be expected from exposure to the stressor
2. Significant impairment in social or occupational functioning

C. Symptoms do not persist more than 6 months after the stressor ends

Acute: < 6 months

Chronic: > 6 months

Source: Ref. 6.

When the stressor is a medical condition such as dystonia, optimal management of the movement disorder is the first step. The therapist may employ interpersonal, cognitive-behavioral, group, or family therapies. Duration of therapy is usually brief but may require more long-term interventions (24). Table 9.12 details the DSM-IV diagnostic criteria for adjustment disorder.

Conclusions

Just as disturbances in brain function cause abnormal movements, alterations in brain function can result in disabling and sometimes lethal psychiatric illness.

A candid survey for problems with mood, anxiety, and perception can identify comorbid psychiatric illness in the dystonia patient, preserve quality of life, and optimize dystonia treatment. By using DSM criteria for comorbid psychiatric illness, clinicians can standardize communications and be confident with indications for treatment. Subclassification of the antidepressants allows for methodically choosing agents based on side effects and targeted neurotransmitter systems. The benzodiazepines are effective anxiolytics, but we now have a broad list of antidepressants with proven efficacy in the anxiety disorders. The newer atypical antipsychotics can be used to bring manic excitement under control or treat psychosis in the dystonia patient while minimizing the effect on the primary movement disorder. Movement disorder centers are recognizing the need for psychiatric expertise on the treatment team. Our patients have taught us the utility of integrating psychosocial and biologic treatments. Inclusion of psychiatric expertise on the movement disorders treatment team is rapidly becoming the standard of care for this challenging group of patients.

Ten Pearls for Clinical Practice in Utilizing Psychiatry as Part of Dystonia Care

1. Screening for disturbances in mood, anxiety, and perception is an integral part of the assessment of a patient with dystonia.
2. Depression in dystonia is common and occurs at a similar frequency as seen in Parkinson's disease and essential tremor.
3. The Beck Depression Inventory is an excellent self-report screening tool for a busy clinical practice.
4. Antidepressant treatment options now include selective effects on serotonin, norepinephrine, and dopamine reuptake without postsynaptic side effects typical of older agents.
5. Rapid relief of anxiety can be achieved with the use of benzodiazepines, but the antidepressants offer the advantage of treating comorbid depression without the risk of chemical dependence.
6. Treatment of social anxiety disorder should not be withheld just because it is thought to be secondary to dystonia.
7. Generalized anxiety disorder often presents with multiple somatic complaints that yield to a single psychotropic medication.
8. Early diagnosis and treatment of panic attacks can avoid exhaustive medical workups and decline in function from agoraphobia.

9. While searching for a reversible cause of psychosis, the atypical antipsychotics can bring symptoms under control, preserve patient safety, and usually avoid exacerbation of involuntary movements seen with the older agents.
10. Adjustment disorders are common in medical illness and warrant brief psychotherapy to preserve quality of life.

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10

Programming Deep Brain Stimulators in Dystonia

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Deep Brain Stimulators in the Dystonia Patient

In 2003 the U.S. Food and Drug Administration (FDA) approved the Medtronic deep brain stimulation (DBS) system for the treatment of dystonia under its "Humanitarian Device Exemption" program. DBS therapy for dystonia is indicated for stimulation of the internal globus pallidus (GPi) for the management of chronic, intractable (drug refractory) primary dystonia, including generalized and/or segmental dystonia, but has also been applied to hemidystonia, focal dystonia, and most recently cervical dystonia (torticollis). The therapy is usually applied in patients 8 years of age or older, but exceptions are made on a case-by-case basis (1–6). DBS has been applied by many centers worldwide as an off-label therapy for secondary dystonic syndromes.

Educating the dystonia patient prior to DBS implantation is an important first step as expectations for the patient, family, and physician must be realistic. DBS is not a cure for dystonia and will theoretically not be disease modifying, although potential neuroplasticity is being studied in many research laboratories. It is difficult to predict the extent of potential DBS response for an individual dystonic patient. When approaching the idea of surgical intervention, the patient should be informed that benefit can range from 0% to as much as 80–90% improvement and depends mainly on the type of dystonia, the presence of contractures, and the presence of other comorbidities. Current available information and experience has indicated that DBS works best in people with primary generalized dystonia, segmental dystonia, and focal dystonia of the neck region (cervical dystonia or spasmodic torticollis). Studies have demonstrated a 40–90% improvement in primary dystonia when treated with DBS (4). Limb improvement is much more likely than axial improvement in typical cases, with the possible

exception of tardive drug–induced dystonia. Non–DYT-1 primary dystonia patients can also benefit from DBS with 50–75% benefit in dystonia, especially if they have a primary generalized dystonia presenting like a DYT-1 genetic case. Late-onset dystonia, secondary dystonia, and focal dystonia patients may have a less predictable response. There have been several recent observations of STN DBS for various forms of dystonia, and STN as a rescue strategy, in addition to bilateral GPi leads, when outcome is suboptimal or there is stimulation induced Parkinsonism. These observations have been made by the author who has also discussed similar findings with Dr. Starr, Dr. Ostrem, and Dr. Baltuch who have published and presented their experiences. There have also been several articles published alluding to the potential benefit of the STN target in dystonia, but more studies are needed (9–12). Patients with severe disability resulting from secondary dystonia (such as encephalitis, tremors, or tardive dystonia) may also benefit from DBS. Patients can experience reappearance or progression of dystonic symptoms following periods of prolonged benefit following stimulation (5–8). Many sufferers also can, following GPi DBS, reduce or discontinue antidystonia medications, such as anticholinergics, muscle relaxants, and benzodiazepines (8). Unfortunately a subset of DBS dystonia patients may be "nonresponders" and not derive benefit from DBS.

For GPi DBS, the target location for the lead is typically the posteroventral region, immediately superior to the dorsal border of the optic tract. There is greater variability in the anterior-posterior coordinates in relation to the mid-commissural point. The lateral coordinate for the lead tip may range from approximately 19 to 23 mm from midline depending on identification by microelectrode recording (3,6). Leads that are slightly suboptimally placed may result in side effects such as "pulling," usually due to the field expanding into the internal capsule, which is located medial and posterior to GPi. Even if the stimulation spreads and results in internal capsule pulling sensations, simple adjustments can be made to reshape the field and to maintain benefit for dystonia as well as to eliminate side effects. This same type of programming adjustment can be performed to improve outcome with other side effects, such as seeing lights/spots (phosphenes), abnormal sensations, or occulomotor difficulties. It should be kept in mind that improvements in dystonia can result in increased dystonic pulling, which can be misinterpreted as "capsular side effects." This issue may crop up 2–3 days following a programming parameter adjustment. The potential for increased dystonic pulling sensations should be discussed with the patient, and if the pulling is mild and tolerable it may be wise, if possible, to wait to see if it will resolve over time. If the pulling is still present after a week or two, then it is likely

related to capsular responses, and reprogramming should be performed. When settings lead to progressive improvements in dystonic posturing, it may be beneficial to wait for maximal improvement prior to programming. If you adjust the setting too soon in dystonia, missed opportunities for improvement at lower current densities may occur, as dystonia patients typically experience delayed benefits following programming. These benefits may not manifest until weeks to months following reprogramming, making it difficult to know when to make adjustments. In this chapter we will review the basics of DBS for dystonia.

Using DBS in Children

Children 7–8 years of age and older can be considered candidates for DBS. The procedure has a relative contraindication in children under this age as growth of the skull may result in migration of the DBS lead (4). Age should be treated as a relative contraindication in young children, but DBS may be medically necessary below the age of 7–8 years because of rapidly progressive symptoms or because of the formation of orthopedic contractions.

Addressing Fixed Joint Contractures

Patients who are surgical candidates preferably should be considered prior to the onset of fixed orthopedic deformities, as these may limit functional improvement even when dystonia symptoms may be ameliorated (3). Some dystonia patients may have fixed joint contractures as a result of shortening of muscle and changes in connective tissue—unlike muscle tension, these contractures are usually not resolved by the stimulation. In these complicated cases, physical measures such as stretching and physiotherapy, compresses, and in very rare cases plaster casts may aid recovery. If such measures do not provide enough relief, correcting the contractures by orthopedic surgery may be considered, although to date the track record for this type of surgery following DBS has been anecdotally poor (4).

Implanting the DBS System

The surgical procedure may vary slightly between neurosurgeons, and the details relevant to your specific institutional procedures should be reviewed individually with the patient. The surgery is comprised of two stages. The first stage involves the placement of the leads into the brain and usually

utilizes a procedure called microelectrode recording to physiologically refine lead placement into the sensorimotor region of pallidum. The first stage of the surgery is usually performed in the awake patient. In the second stage of the procedure (which may be performed on a different day), the neurostimulator and extension wires are placed in the chest wall, similar to a pacemaker (4). The second stage of the procedure is usually performed under anesthesia and in many centers is delayed until several weeks following stage one.

Components of the DBS System

DBS is similar in design and function to a cardiac pacemaker. Impulses from the system's "neurostimulator," which is located "subclavicularly" in the chest wall, are sent to the globus pallidus (GPi), subthalamic nucleus (STN), or the thalamus in an effort to neuromodulate abnormal firing rates and patterns. These target structures have been implicated in the regulation of movements as well as in systems modulating mood and cognition (4).

The three main components of the DBS system include the neurostimulator, the DBS lead, and the extension cable/connector cable (Figure 10.1):

- **Neurostimulator**: This pacemaker-like device contains a small battery that powers the system. The neurostimulator has also been referred to as an implantable pulse generator (IPG). The dimensions of the currently approved device are approximately 7.5 cm (3 inches) wide and 1.3 cm (0.5 inches) thick, or slightly smaller depending on the specific model

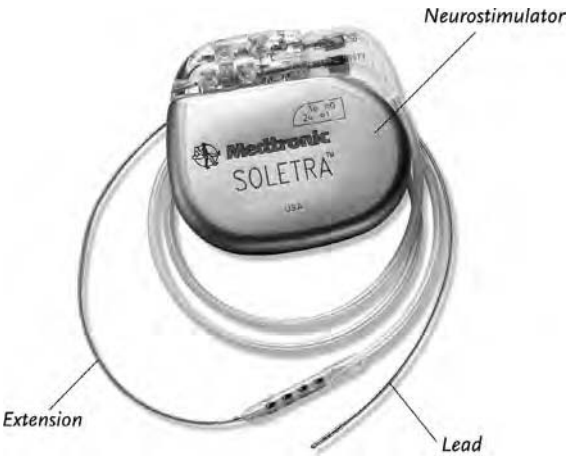

FIGURE 10.1 Components of the DBS system. Copyright © 2008, Medtronic, Inc. Medtronic is providing permission to use its copyrighted images of devices and therapies but does not control whether that use conforms with FDA-approved or -cleared indications. To view the approved or cleared indications associated with this image, please go to the relevant condition on www.medtronic.com and click on "Important Safety Information."

selected by the surgeon. In addition to the battery, the neurostimulator contains a computer chip that may be programmed to send electrical impulses that have the potential to neuromodulate dystonia symptoms. Medtronic offers two neurostimulators: Soletra, a single-chamber neurostimulator that powers one lead, and the Kinetra, a dual-chamber neurostimulator that can power two leads. Currently, Medtronic has the only FDA-approved device for dystonia. Kinetras are "thicker" than Soletras, and this must be kept in mind when implanting children or adults with lower than ideal body weight (children may require, for example, two Soletras).

- **DBS Electrode:** Medtronic offers two electrode models (3387 and 3389). Both models consist of quadripolar or four contact electrodes. The lead model 3387 has an intercontact distance of 1.5 mm between each active contact. The lead model 3389 has an intercontact distance of 0.5 mm between each lead contact. Current is delivered through one or more of the four cylindric electrode contacts, which are 1.27 mm in diameter and 1.5 mm in length. Each contact electrode surface is 5.99 mm^2. Both model leads are insulated and must be carefully placed within a brain region (13).
- **Extension**: The insulated extension wire is placed under the scalp but outside the skull, connecting the DBS lead to the neurostimulator. The extension cable runs behind the ear, down the neck, and into the chest below the collarbone, where it attaches to the neurostimulator. The extension cable can in certain circumstances be run into the abdominal region if the neurostimulator is preferred not to be left infraclavicularly.

Most dystonia patients will require bilateral GPi or STN DBS leads unless they are experiencing hemidystonic or focal limb dystonia. The leads can be connected to a Kinetra neurostimulator or to two Soletra neurostimulators as may be required in individual patients.

Medtronic Activa Soletra Neurostimulator

Soletra neurostimulators are single chambered and allow only one lead to be attached, activated, and programmed. (Figures 10.2 and 3) The Soletra does not have the capability to allow a patient to adjust settings with their Access Review remote (unlike the Kinetra), but patients do maintain the ability to turn the stimulator off and on (Figure 10.4).

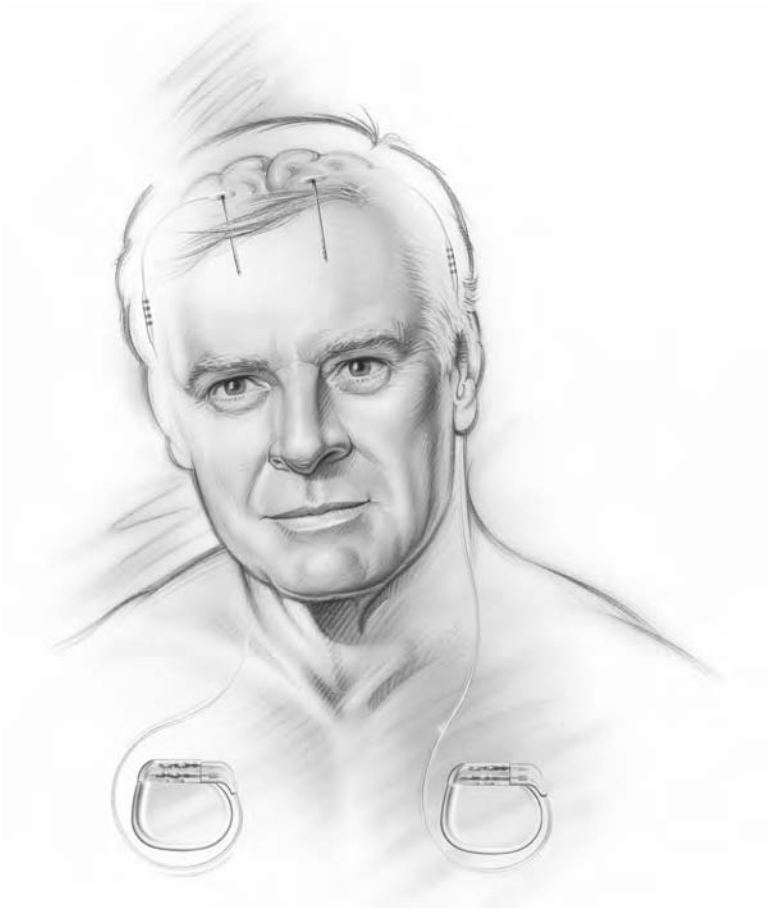

FIGURE 10.2 Medtronic Soletra neurostimulator. Copyright © 2008, Medtronic, Inc. Medtronic is providing permission to use its copyrighted images of devices and therapies but does not control whether that use conforms with FDA-approved or -cleared indications. To view the approved or cleared indications associated with this image, please go to the relevant condition on www.medtronic.com and click on "Important Safety Information."

FIGURE 10.3 Medtronic Soletra neurostimulator with extension cables and lead. Copyright © 2008, Medtronic, Inc. Medtronic is providing permission to use its copyrighted images of devices and therapies but does not control whether that use conforms with FDA-approved or -cleared indications. To view the approved or cleared indications associated with this image, please go to the relevant condition on www.medtronic.com and click on "Important Safety Information."

FIGURE 10.4 Access review. A patient controller for the Soletra neurostimulator. Copyright © 2008, Medtronic, Inc. Medtronic is providing permission to use its copyrighted images of devices and therapies but does not control whether that use conforms with FDA-approved or -cleared indications. To view the approved or cleared indications associated with this image, please go to the relevant condition on www.medtronic.com and click on "Important Safety Information."

Medtronic Activa Kinetra Neurostimulators

Kinetra neurostimulators are dual chambered and allow two leads to be activated, programmed, and monitored by a single device (Figures 10.5 and 6). The Kinetra also has the capability to allow a patient to adjust some settings at home utilizing their Access remote that can be set within a specified range by the DBS programmer (Figure 10.7). The DBS programmer can independently change the voltage and pulse width for each lead, but with Kinetra devices the frequency must remain the same for both connected leads.

Review of Postoperative Lead Locations

Lead location is an important factor in outcome. No amount of expert programming can compensate for a misplaced lead. A 1-month postoperative CT scan can be obtained and fused with a preoperative MRI in order to obtain

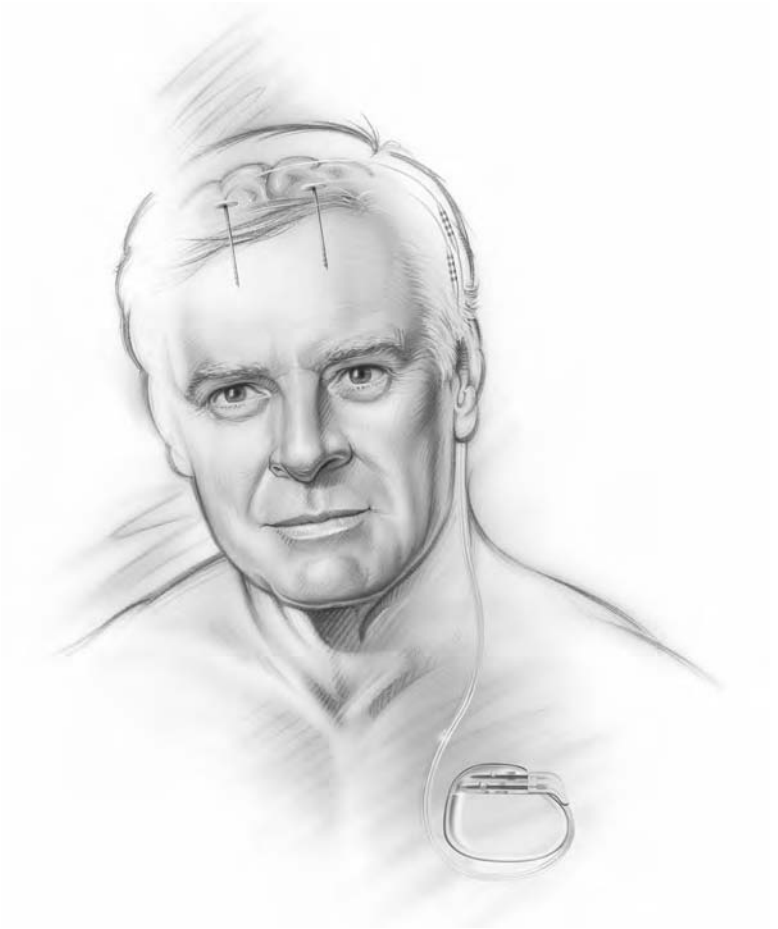

FIGURE 10.5 Medtronic Kinetra neurostimulator. Copyright © 2008, Medtronic, Inc. Medtronic is providing permission to use its copyrighted images of devices and therapies but does not control whether that use conforms with FDA-approved or -cleared indications. To view the approved or cleared indications associated with this image, please go to the relevant condition on www.medtronic.com and click on "Important Safety Information."

accurate lead measurements (or alternatively a simple postoperative MRI). For patients new to a center with already implanted DBS devices, a MRI or MRI/CT fusion should be used to evaluate lead measurement(s). MRI Guidelines for Medtronic Deep Brain Stimulation Systems can be found on the Medtronic website (http://www.medtronic.com/physician/activa/downloadablefiles/M925038A_a_001.pdf) and should be reviewed prior to sending implanted DBS patients for scanning. A protocol should be established with the institution's DBS programming clinic and radiology department to ensure safe imaging of DBS patients. Prior to MRI, neurostimulators should be turned to zero volts and placed in the off condition.

FIGURE 10.6 Medtronic Kinetra neurostimulator with extension cables and lead. Copyright © 2008, Medtronic, Inc. Medtronic is providing permission to use its copyrighted images of devices and therapies but does not control whether that use conforms with FDA-approved or -cleared indications. To view the approved or cleared indications associated with this image, please go to the relevant condition on www.medtronic.com and click on "Important Safety Information."

FIGURE 10.7 Access review. A patient controller for the Kinetra neurostimulator. Copyright © 2008, Medtronic, Inc. Medtronic is providing permission to use its copyrighted images of devices and therapies but does not control whether that use conforms with FDA-approved or -cleared indications. To view the approved or cleared indications associated with this image, please go to the relevant condition on www.medtronic.com and click on "Important Safety Information."

Programming of the Deep Brain Stimulator

The DBS system should be programmed and individually tailored to a patient's needs (Figure 10.8). Programming is accomplished by use of a portable programming device that the physician or practitioner can utilize to set appropriate parameters for the neurostimulator by adjusting electrode polarity, voltage, pulse width, and frequency.

- **Electrode Polarity:** Each electrode contact can be programmed as bipolar (cathode lead negative polarity; anode lead positive polarity) or as monopolar stimulation (cathode lead negative polarity, case set as anode). Monopolar stimulation provides a more spherical current field, whereas bipolar stimulation provides a more elliptical field (the maximal effect with either is centered around the cathode). One should try monopolar stimulation first, which usually requires lower stimulation intensity (voltage) to achieve the same clinical benefit when compared to bipolar stimulation.

FIGURE 10.8 The programmer for the DBS device. Copyright © 2008, Medtronic, Inc. Medtronic is providing permission to use its copyrighted images of devices and therapies but does not control whether that use conforms with FDA-approved or -cleared indications. To view the approved or cleared indications associated with this image, please go to the relevant condition on www.medtronic.com and click on "Important Safety Information."

Often a single cathode (monopolar setting) is chosen for DBS, although it may be necessary in some cases to activate two adjacent contacts (double monopolar) to achieve more diffusion of current. When monopolar settings lead to side effects, bipolar stimulation can be utilized to reduce diffusion into adjacent structures and to avoid/minimize side effects such as pulling or dysarthria (13).

- **Amplitude:** Medtronic neurostimulators allow one to vary voltage in 0.1 V steps between 0 and 10.5 V. The current versions of the device are not constant current. In the Soletra neurostimulator energy consumption is linear up until it reaches 3.6 V and then rises abruptly above this level, because a voltage doubler (3.6) or tripler (7.3) circuit becomes activated (13). When programming above 3.6 V, the benefit a patient receives must be significant enough to warrant the added battery drain. With regard to the Kinetra neurostimulator, a different electrical circuitry may allow an increased voltage (up to 10.5 V) with a linear increase in current consumption (maintained throughout the whole range of amplitudes).
- **Pulse Width:** Pulse width may be varied in steps usually ranging between 60 and 450 μsec.
- **Frequency:** Frequency can be set between 2 and 185 Hz for the Soletra neurostimulator and between 2 and 250 Hz for the Kinetra. Although most patients have been traditionally programmed using high frequency (>100 Hz), lower frequency settings (50–60 Hz) have been effective in some patients (particularly those with DYT-1 dystonia) (Tagliati and Alterman patient observations).

The patient can be issued a handheld programmer that they or the caregiver may use in order to turn the neurostimulator on or off (Access Review for the Soletra IPG) or to adjust settings within limits set by the doctor (The Access for the Kinetra IPG) (4). Patients can alternatively be issued a magnet, which they may hold over their device for a few seconds to cycle it from on to off or from off to on. When using a magnet, an AM/ FM radio transmitter can be effective to check and see if the device is on. A change in static on the AM/FM radio can be perceived audibly if the neurostimulator is in the on position when the patient holds the radio transmitter over the chest device or neck connector wire (13).

Unlike disorders such as Parkinson's disease and essential tremor, where significant improvement can be seen during initial programming of the DBS device, the benefits in dystonia may take time to fully manifest. Often continuous

stimulation over several weeks to months is required before changes in the dystonia become clinically apparent. Monthly adjustments may be required, and the programmer must be careful not to make changes too frequently to avoid missing a good programming setting (3,5,8).

Positive benefits of DBS have been observed in dystonia over months to years even when keeping the setting constant. Patients who have reported positive changes in their dystonia symptoms with a single setting may continue to have improvement over time, and the programming team should be careful in making major changes when significant improvements appear. Many patients require monthly programming for the first year. Some require bimonthly adjustments, although caution should be exercised as this may prove too frequent. Bimonthly adjustments may be needed for severe dystonia (the disadvantage of bimonthly adjustments is the potential of missing a beneficial setting as a result of a delayed response) (3,8). Additionally and as mentioned previously, some individuals will complain of pulling and will request changes in the DBS setting. Caution should be exercised as pulling could be a manifestation of initial improvement in dystonia. A subset of patients during initial and follow-up DBS programming will have subjective relaxation when first exposed to a setting that will be beneficial for long-term dystonia relief.

To help determine which lead to initially activate, the DBS programmer can obtain thresholds for adverse events on each DBS contact by slowly increasing the voltage and recording levels that elicit transient and permanent side effects (this should be performed on each of the four contacts) (8). Assessment of efficacy is accomplished by activating each lead using high frequency (130–185 Hz) and low pulse width (60–90 μsec) in monopolar stimulation. The best results for chronic programming in dystonia are usually seen with the deepest available contacts. Initial programming should utilize the optimal contact, which can be selected during thresholds for side effect and benefit, and also may be assisted by reviewing lead location using MRI or MRI/CT fusion. Benefit may not be manifest at initial programming in dystonia patients, although some will report subjective relaxations in symptoms. Each center may have their own protocol for setting the DBS system at initial programming, and there is no right or wrong approach as long as it is done in a systematic fashion. At subsequent visits the programmer should assess the patient's response to DBS and decide on an individual basis whether further adjustments are necessary, but usually several adjustments are required. If efficacy is not apparent with monopolar

stimulation, then an additional contact can be added (double monopolar). Bipolar configuration may be required if side effects are encountered such as capsular pulling or higher voltages or pulse widths lead to side effects (8).

Settings that are efficacious for one patient may not be adequate for another patient. There should not be a dogmatic approach to programming for dystonia. Each center must set a protocol that is systematic, organized, and comprehensive. Pulse widths in dystonia may vary from 60 to 450 μsec, and frequency may vary from 60 to 185 Hz. Some patients, especially patients with DYT-1 dystonia, seem to respond to lower frequency stimulation, which has the benefit of saving battery life. Over the years centers have migrated to the use of lower pulse widths of 120–210 μsec rather than 450 μsec, although higher pulse widths may be needed on a case-by-case basis. A trial of low frequency (60 Hz) for 3 months, especially in DYT-1 patients, may also be an effective way to save battery life. Higher amplitude, pulse width, and frequency may lead to excessive drain of the neurostimulator battery and can lead to the frequent replacement of the neurostimulator, sometimes in intervals less than a year. Voltage settings above 3.6 V (on the Soletra battery) will cause depletion to occur twice as fast. If patients present with acute worsening of dystonia, then sudden battery failure should be anticipated. To prevent sudden battery failure from occurring, providers should confirm battery status at each visit. Once a patient is on stable settings, the provider can call Medtronic to request an estimated length of battery life. It is optimal to replace the neurostimulator prior to failure; some patients may have a worsening of dystonia as the neurostimulator nears end of battery life. A small subset of patients may continue to have benefits even with battery failure.

Troubleshooting of the Deep Brain Stimulator

When patients experience no stimulation response during thresholds, loss of benefit, or intermittent/continuous side effect of stimulation, the DBS programmer should consider possible device failure (but not overreact as there may be no response initially in dystonia). Evaluating the device can be achieved by reviewing the device's control counter, battery check, and general/therapeutic impedance check:

- **Control Counter**: The magnet activation counter should routinely be set to zero at the end of each patient visit to allow for detection of unusually

high activation cycles as well as compliance review. One of the most common causes of sudden decrease in symptom suppression is an accidental turning off of one or both stimulators. Some patients can be off for several hours to days before return of symptoms is noticed by the patient or by a significant other. In addition, unusually high numbers of on-off cycles, in a range of >25 events in the neurostimulator log, can suggest deactivation/activation of the DBS system. Possible sources of electromagnetic interference are household devices used in close proximity to the neurostimulator such as electric shavers, electric toothbrushes, microwaves, mixers, electric drills, other power tools, or loudspeakers. Antitheft devices in stores can also cause deactivation/activation of the neurostimulator. In addition, magnets in brooches or name badges can deactivate the neurostimulator when placed over the device. Patients can be given a remote to check whether their neurostimulator is on or off. The Access remote for Kinetra neurostimulators and the Access Review remote for Soletra neurostimulators can also allow the patient to turn the device on or off. Kinetra neurostimulators can have the "read" switch disabled to decrease electromagnetic interference.

- **Therapeutic and General Impedances:** These should be obtained in Soletra neurostimulators using the standard setting of 1.0 V, 210 μsec, and 30 Hz. The Kinetra neurostimulator will require each electrode to be tested individually by selecting each electrode for general impedance. In addition, the voltage for impedance for Kinetra should be tested at 3.5 V for the most accurate results. Typically the measured impedances lie in a range of 500–1500 Ohms. An impedance >2000 Ohms for the Soletra and >4000 Ohms for the Kinetra indicates a connection problem, a broken cable, or a lead fracture. Impedance readings of <50 with current drains in the 200s indicate a possible short circuit (13).
- **Battery Check:** Neurostimulator battery strength should be evaluated at each visit. Neurostimulators should be replaced prior to battery depletion to avoid loss of dystonic efficacy. Estimation of battery life can be obtained from Medtronic technical services once chronic stimulation settings are established. Battery life varies depending on patient programming settings, with higher settings having a higher drain on the neurostimulator. For the dystonia patient, neurostimulator life typically varies from 1 to 3 years. The Soletra neurostimulator voltage at the time of implantation can range from 2.69 to 2.74. Over the span of the Soletra life the voltage will remain

constant. As the Soletra neurostimulator nears the end of its battery life, the battery voltage will begin to slowly decline for several months; then a more rapid depletion of the neurostimulator battery will occur. Kinetra neurostimulator battery voltage will decrease over time and will display an indication of battery capacity used overtime. As the Kinetra neurostimulator reading nears 2.44 V, plans should be made to replace the neurostimulator.

- **Palpation of System Components:** A noninvasive approach to detect intermittent stimulation is palpation of the implanted components with stimulation in the "on" condition in order to identify the location of a loose connection. If the patient reports tingling pain or dysaesthesias at a location near the implanted system, damage to the insulation and an exposed conductor should be suspected (e.g., a short) (13).
- **X-Ray**: If a hardware problem is suspected, then plain film x-rays should be obtained. Shunt series x-rays without abdomen (unless the patient's neurostimulator is implanted in the abdomen) can show possible hardware problems. A broken lead, dislodgement of the lead from the extension connector, or gross damages to the insulation may be visible on these scans and help to narrow down the location of the system problem.

Potential Risk of the Surgery and Side Effects from Stimulation

Common side effects reported by DBS patients include dysarthria, tingling sensations, dizziness, jolting sensations, shocking sensations, and numbness. Sudden loss of electrical signal from battery depletions or accidental turning off of stimulator(s) can also exacerbate a dystonia symptom. Many of the side effects can be managed by a simple noninvasive adjustment of the stimulation parameters. It may, however, require several follow-up visits to empirically discover the right stimulation settings in order to achieve the best result while minimizing side effects (Table 10.1).

Potential risks of DBS surgery include intracranial hemorrhage, infection, stroke, seizure, failure of the device to provide benefit, lead migration, lead fracture, need for lead adjustment, hydrocephalus, worsening of preexisting cognitive or mood/anxiety, suicide, visual loss, pulmonary

embolus, air embolus, cognitive dysfunction, and death. Additional deficits that may appear following surgery may include weakness, numbness, and changes in vision, speech, cognition, mood (potential suicidality), and gait. Complications can also result from breakdown of the skin surrounding the stimulator or dysfunctions in the hardware (Table 10.2). Details of the surgical procedure are provided in the chapter on medical and surgical therapies for dystonia.

Common DBS-induced side effects in dystonia patients include dysarthria, dysphagia, and capsular pulling. Capsular pulling and pulling

TABLE 10.1 Potential Side Effects of Deep Brain Stimulation

Tingling sensation (paresthesia)
Temporary worsening of the patient's disease symptoms
Speech problems such as whispering or strained voice (dysarthria or dysphonia)
Forming words (dysphasia, verbal fluency) or soft speech (hypophonia)
Vision problems (double vision, spots)
Dizziness or lightheadedness (disequilibrium)
Facial and limb muscle weakness or partial paralysis (paresis)
Abnormal, involuntary movements (chorea, dystonia, dyskinesia, ballism)
Movement problems or reduced coordination
Jolting or shocking sensation
Numbness (hypoesthesia)

TABLE 10.2 Deep Brain Stimulation Risks

Paralysis, coma, and/or death
Bleeding inside the brain (stroke)
Leakage of fluid surrounding the brain
Seizures
Infection
Allergic response to implanted materials
Temporary or permanent neurological complications
Confusion or attention problems
Pain at the surgery sites
Headache

TABLE 10.3 Common Deep Brain Stimulation–Induced Side Effects in Dystonia Patients

Dysarthria
Dysphagia
Capsular pulling
Numbness of the face, arm, hand, or leg
Stiffness or weakness of the limb
Double vision
Closure of the eyelids
Change in mood
Thinking problems
Facial weakness
Dizziness/lightheadedness
Imbalance, walking problems
Suicide/mood/cognitive issues

related to positive changes in dystonia are often similar and difficult to differentiate (Table 10.3).

Precautions for Patients Following Implantation

Since the entire DBS system is implanted under the skin, patients can swim, shower, and be exposed to water. Precautions must be taken in the vicinity of strong electromagnetic fields. In particular, this may also limit the capacity to undergo examination by MRI. As mentioned above, the MRI Guidelines for Medtronic Deep Brain Stimulation Systems should be carefully reviewed prior to sending implanted DBS patients to MRI. Deep-heat treatments (diathermy or therapeutic ultrasound), which are occasionally used in physical therapy or by chiropractors for treating muscular spasms or pain, must also be avoided in patients with DBS as they can lead to heating of the hardware and in turn heating of the brain. Using kitchen equipment (microwave) and mobile phones does not seem to interfere with DBS devices (4). Dental drills should not be placed over the neurostimulator. Precautions for specific surgeries should be discussed with Medtronic. In general, bipolar cautery should be used during surgery and grounding pads should not be placed over the neurostimulator.

Benefits of the DBS System

Improvement of symptoms following DBS implantation usually does not manifest until weeks to months following activation, and improvements in some cases may be seen even past one year. Often, the amount of medication consumed can be reduced following DBS (8). In rare cases patients will no longer require any medication. By reducing the frequency of muscle spasms and correcting abnormal postures, DBS may facilitate reintegration into society and a better chance at a more normal life. Additionally, many patients post-DBS will be able to take a more active role in social activities without increased self-esteem and decreased embarrassment. Additionally, people with dystonia are less likely to feel depressed or anxious following DBS (4).

Role of the DBS Programmer in Coordinating Care

Providing care and programming for the dystonia DBS patient can be time-consuming, and staff should be prepared for initial and follow-up appointments to run potentially past a single hour. Utilizing the services of a nurse practitioner or physician assistant to perform programming and follow-up can free the neurologist or neurosurgeon, who can then move into a supervisory role. Nurse practitioners and physician assistants provide quality care that is comprehensive and cost-effective (14,15). The DBS programmer coordinates and manages the care of these patients.

The role of the programmer is also covered in Chapter 3.

The nurse practitioner or physician assistant can assist in the coordination of referrals to members of the multidisciplinary/interdisciplinary team evaluating potential DBS surgical candidates. This team could include the neurologist, neurosurgeon, neurophysiologist, psychiatrist, and radiologist and in some cases representatives from the physical therapy, occupational therapy, speech therapy, and social work departments. The programmer may be the first to identify and stratify surgical- versus medication- versus stimulation-induced adverse effects and formulate the treatment plan. DBS programmers also play a critical role in the coordination of follow-up imaging studies for lead localization and other potential DBS complications.

Some institutions utilize registered nurses (RNs) to provide programming of the DBS device. Registered nurses can be trained on programming but cannot diagnosis or adjust patient medications. The neurologist should adjust

patient medications as needed and assess the need for referral to interdisciplinary team members.

Once the dystonia patient has undergone implantation of the DBS device, follow-up visits should be provided to the patient at intervals designated as standard of care for that particular institution. We use once a month for the first 6 months, with reassessment at 6 months and extending follow-up intervals to tailor patient needs following this point. At each visit the DBS programmer should check impedances and battery status. If impedances are abnormal, this may indicate a lead fracture (with high impedances) or a lead short (low impedances), and patients should be sent for a shunt series plain film x-ray without abdominal view, unless there is an abdominal neurostimulator. At each visit the DBS programmer should also reset the counters, as this will help monitor for any unintentional turning off of the neurostimulator, which can happen when patients pass through magnetic fields, such as store security panels or other high-field magnetic sources; patients should be made aware of this possibility. If numerous activations are apparent on the device counter since the time of the last visit, the programmer should inquire as to whether the device has been turned off and on intentionally for any reason. If there have not been deliberate deactivations of the device, a search for exposure to a magnetic field should be undertaken (this may include many diverse sources such as freezers as well as security systems in retail stores). Magnetic name badges or magnetic broaches can turn devices to the on or off condition. In addition, worsening of dystonia may be observed in a delayed manner once stimulation is turned off, with battery failure, or with hardware breakage (8). If these symptoms present, accidental on/offs should be considered. When programming children it is important if they have two chest neurostimulators that they sit back in a chair during changes and that the devices are kept 6 inches apart. If kept too close during programming they may reset to factory default settings found on the Invision Programmer when it first interrogates a DBS device (0- 3+ 0.0 volts, 210 μsec pulse width, 60 Hz frequency).

Physical and Occupational Therapy

Once the patient begins to manifest benefit as a result of the DBS device, the programmer may pursue in select cases physical therapy and/or occupational therapy. Therapeutic passive and active range of motion exercises may be aimed at strengthening muscles and improving coordination, especially in patients who can tolerate full rehabilitation programs. The aim of the therapy

should be to attempt to relearn more normal movement and to increase the general psychological and physical capacities in order to reintegrate into society. Hydrotherapy can also be beneficial.

Eleven Clinical Pearls for the DBS Programmer

1. Obtain lead location to ensure optimal placement.
2. Obtain thresholds for side effect profile.
3. Educate the patient pre and post implantation for expectations of benefit and side effects.
4. Ensure neurostimulators are at least six inches apart when programming, especially in children.
5. If loss of a benefit is manifested then evaluate for accidental "off," battery life depletion, impedances for technical failure, or for lead migration (especially in children).
6. Plan for battery replacement before battery failure.
7. Clear neurostimulator counter each visit to allow for monitoring of unintentional deactivation/activation of system.
8. Encourage the patient to work through pulling for one to two weeks unless extreme.
9. Refer appropriate patients to physical therapy, occupational therapy, and speech therapy.
10. Educate the patients on system precautions such as avoiding MRI/diathermy.
11. Establish monthly follow-up visits for the first year.

Websites

American Association of Neuromuscular & Electrodiagnostic Medicine: http://www.aanem.org/education/patientinfo/dystonia.cfm

American Speech-Language-Hearing Association: http://www.asha.org/public

The Bachmann-Strauss Dystonia & Parkinson Foundation, Inc.: http://www.dystonia-parkinsons.org/

The Canadian Movement Disorder Group, Dystonia: http://www.cmdg.org/Movement_/dystonia/dystonia.htm

Care4Dystonia, Inc.: http://www.care4dystonia.org/

Dystonia Association of Kentucky Going the DYSTance for all dystonia-affected individuals: http://www.dystoniaassociation.org/aboutdeepbrain stimulation.htm

Dystonia Ireland: http://www.dystonia.ie/menu.asp?Menu=23

Dystonia Medical Research Foundation: http://www.dystonia-founda-tion.org/

Dystonia Spasmodic Torticollis: http://www.spasmodictorticollis.org/

Dystonia-Support Group-Alabama: http://www.dystonia-alabama.org/

The Dystonia Society: http://www.dystonia.org.uk/about-dystonia-page 43048.html

The European Dystonia Portal: http://www.dystonia-europe.org/europe/ indexB.htm

Medtronic Activa Deep Brain Stimulation: http://dystonia.activadbs.com/

The Movement Disorder Society: http://www.movementdisorders.org/

National Institute of Health, Fact Sheet on Dystonia: http://www.nih.gov/ about/researchresultsforthepublic/Dystonia.pdf

National Institute of Neurological Disorders and Stroke: http://www.ninds.nih.gov/disorders/dystonias/detail_dystonias.htm

National Spasmodic Dysphonia Association: http://www.dysphonia.org/

National Spasmodic Torticollis Association: http://www.torticollis.org/

PBS: Twisted: http://www.pbs.org/independentlens/twisted/dystonia.html

Tyler's Hope for a Dystonia Cure: http://www.tylershope.org/

University of Florida, Movement Disorders Center: http://mdc.mbi.ufl.edu/

WE MOVE (Worldwide Education & Awareness for Movement Disorders): http://www.wemove.org/

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