PREFACE

The field of movement disorders is relatively broad, encompassing disorders of increased movement, such as tremors, dystonia, and tics, to disorders characterized by a paucity of movement, such as Parkinson's disease. Our understanding of the pathogenic mechanisms and our treatment options are expanding at a rapid pace. This expansion ranges from the medical and surgical advances in treating Parkinson's disease to the flood of genetic abnormalities that have now been found to cause various movement disorders. Although many patients are seen by the movement disorders specialist in neurology clinics around the country, most of these patients receive their follow-up care from a primary care physician or "general" neurologist who must be versed in the characteristics and treatment plans of this diverse group of disorders.

The major goal of *Parkinson's Disease and Movement Disorders: Diagnosis and Treatment Guidelines for the Practicing Physician* is to distill this immense amount of information and to educate the practitioner about the many facets of the movement disorders field. We believe that this book fills a large void, since most texts on movement disorders are more detailed and geared toward the specialist. We have asked the chapter authors to emphasize the clinical characteristics of each disorder, discuss the differential diagnosis and the diagnostic testing, and then outline the various treatment options, as if they were teaching during a preceptorship in their clinic. To this end, we have not designed the book to be an exhaustive review of each topic; rather, it takes a general approach to each subject. We have avoided referencing each statement; a short list of further recommended reading sources is given at the end of each chapter.

The purpose of this text is to help the practitioner distinguish which disorder is being encountered, give a basic understanding for test and treatment options that are required, and synthesize any recommendations made by a consulting specialist. As the movement disorders specialist becomes busier and insurance regulations limiting specialty referrals increase, the burden of caring for these patients by the primary care physician will continue to grow. Thus, we hope that this text will offer the reader full confidence in approaching patients with movement disorders.

The text is organized into five sections: basic diagnostic principles, Parkinson's disease, other parkinsonian disorders, hyperkinetic movement disorders, and other movement disorders. In Chapter 1 of Section A: Basic Diagnostic Principles, Dr. J. Eric Ahlskog provides an extensive overview on the neurologic examination. Since movement disorders can involve all parts of the neurologic system, a detailed neurologic examination is often imperative when these patients are evaluated. Changes in speech often occur in many of the disorders, and the speech characteristics may provide important diagnostic clues. In Chapter 2, Dr. Joseph Duffy describes the varieties of motor speech abnormalities that may be encountered and provides a systematic approach to their recognition.

Given the frequency of Parkinson's disease (PD), the tremendous advances made over the past two decades in understanding the disease and its treatment, and the debates on the "best" way to treat the patient, we have devoted 12 chapters to this entity in Section B: Parkinson's Disease. In Chapter 3, Dr. Howard Hurtig discusses the pathophysiology, neurochemistry, and neuropathology of PD. This is followed by Dr. Richard Dewey's

description of the clinical characteristics of PD in Chapter 4, outlining not only the typical features, such as rest tremor and slowness of movement, but also clinical signs suggesting an atypical form of parkinsonism.

What causes PD? The intriguing search for etiologic answers has generated volumes of studies and papers, sometimes with conflicting results. This information is distilled in two chapters. The epidemiologic studies of PD, which have generated multiple clues to the etiology, are reviewed by Dr. Demetrius Maraganore in Chapter 5. This chapter covers reported risk factors and the role of genetics (including the defects in the α-synuclein gene) in addition to basic incidence and prevalence data. On the other hand, basic bench research has produced multiple lines of evidence for a variety of possible causal factors. Etiologic hypotheses generated from knowledge of biochemical mechanisms are comprehensively reviewed by Dr. Peter LeWitt in Chapter 6. This chapter includes discussion of the roles of oxidative stress, mitochondrial dysfunction, and neurotoxins and addresses etiologic mechanisms that have received less publicity, such as possible autoimmune and infectious causes.

We have divided the discussion of the medical treatment of PD into three chapters. The issue of whether our current drug arsenal includes medications that will slow the progression of PD is hotly debated, and in Chapter 7, Dr. Ahlskog tackles the theories and practical issues for the practitioner. Chapter 8, also by Dr. Ahlskog, is devoted to the various treatment options available for the patient with newly diagnosed PD. This includes decision-making regarding the use of levodopa, dopamine agonists, and numerous other agents. The complicated issue of how to treat the patient with more advanced PD is covered by Dr. Ryan Uitti in Chapter 9.

Patients with PD also have nonmotor manifestations that can be as disabling as the tremor and bradykinesia. The sleep problems of PD, including insomnia and daytime drowsiness, are addressed by Dr. Cynthia Comella in Chapter 10. Neurogenic bladder and bowel problems and symptomatic orthostatic hypotension are common issues in the PD clinic; these autonomic problems and treatment strategies are covered by Dr. Bradley Hiner in Chapter 11. Dementia, psychosis, and depression can be overwhelming factors in the patient with more advanced PD, and Dr. Erwin Montgomery discusses these in Chapter 12.

Currently, the most visible topic concerning PD is surgical treatment, which has made newspaper and television headlines for the past several years. In Chapter 13, Dr. Kathleen Shannon discusses neurosurgical intervention, including pallidotomy, thalamotomy, deep brain stimulation, and cerebral transplantation, and reviews the prospects for the future. She provides guidelines on which patients may benefit and which of the different procedures may be appropriate for a given patient.

Therapy for patients with PD does not end with medications and surgery; Chapter 14, written by Drs. Padraig O'Suilleabhain and Susan Murphy, addresses adjunctive treatments. They include nutrition and dietary issues, which are especially important in advancing disease. They also address the role of physical therapy in management of parkinsonian motor problems.

All disorders characterized by slowness of movement are not PD, and in Section C we have devoted six chapters to discussing these other disorders. In Chapter 15, Drs. Eric Molho and Stewart Factor cover secondary causes of parkinsonism, such as vascular, toxic, and traumatic etiologies; they also provide a practical strategy for the workup of parkinsonism. Among the more common neurodegenerative disorders sometimes mistaken for PD is progressive supranuclear palsy; the key clinical signs and points that

separate this disorder from PD are covered by Dr. Mark Stacy in Chapter 16. When patients have cerebellar signs, prominent autonomic dysfunction, or resistance to dopaminergic therapy, one must consider the multiple system atrophies discussed by Dr. James Bower in Chapter 17. Inherited cerebellar disorders, including the autosomal dominant spinocerebellar ataxias, sometimes resemble sporadic multiple system atrophyand occasionally PD; these familial ataxic syndromes are reviewed by Dr. Bower in Chapter 18. Corticobasal degeneration may resemble PD early in the course. The clinical hallmarks that allow differentiation are covered by Dr. Brad Boeve in Chapter 19. The final chapter in this section, Chapter 20, by Dr. Richard Caselli, describes the various primary dementing disorders, including Alzheimer's disease, that often include components of parkinsonism.

Section D begins the discussion of disorders characterized by too much movement, or hyperkinetic movement disorders. All the chapters in this section address the characteristics of the individual disorders, diagnostic considerations, and treatment options. We begin by a discussion of the most commonly encountered movement disorder, tremor. Chapter 21, by Dr. Joseph Matsumoto, describes the different types of tremor and how to differentiate and treat them. Chapter 22, by Dr. Jean Hubble, goes into further detail about the most commonly seen tremor, essential tremor.

Dystonia is a more common disorder than is often appreciated, occurring in adulthood as torticollis, blepharospasm, writer's cramp, and other focal or segmental dystonias. These along with generalized dystonic conditions, including primary torsion dystonia developing in childhood, are reviewed by Dr. Daniel Tarsy in Chapter 23. Hemifacial spasm is sometimes mistaken for facial dystonia; this disorder, due to compression of the seventh cranial nerve, is discussed by Dr. Mark Lew in Chapter 24.

The dancing movements of chorea have origins ranging from inherited (Huntington's disease) to infectious (Sydenham's chorea) causes. Clinical characterization and treatment are covered by Dr. John Caviness in Chapter 25. Tardive dyskinesias are sometimes confused with primary choreiform syndromes. These iatrogenically induced conditions are discussed by Dr. Kapil Sethi in Chapter 26.

The lightning-like jerks of myoclonus occasionally cause diagnostic confusion; if repetitive, they may resemble tremor or the phasic movements seen in some dystonic conditions. In Chapter 27, Dr. Caviness discusses diagnostic criteria, categorization, and treatment of myoclonus.

Simple spasm of muscle may have a wide variety of causes, ranging from peripheral to central nervous system origins. In the most elementary sense, the concept of muscle spasm should include the sustained muscle contraction state of dystonia. Primary disorders characterized by muscle spasm, however, have their own distinguishing features, which separate them from primary dystonias. These disorders of muscle spasm, including the prototypical condition, stiff-man syndrome, are discussed by Dr. Michel Harper in Chapter 28.

The most common movement disorder of childhood is that of tics. This problem, however, is not confined to children and occasionally confronts internists with adult practices. The spectrum of motor and other tics, as well as the constellation of symptoms that make up Tourette's syndrome, is the topic of Chapter 29 by Drs. Kathleen Kujawa and Christopher Goetz.

A disorder that has gained much recognition in the past few years is restless legs syndrome, discussed by Drs. Virgilio Evidente and Charles Adler in Chapter 30. Dr.

Adler then covers the various uses of botulinum toxin (Chapter 31), an injectable agent that reduces movement and has application for treating multiple different movement disorders. This drug has revolutionized the treatment of dystonia and certain other hyperkinetic movement disorders.

We conclude this book with Section E, which covers other movement disorders, those that do not fit well into previous sections. Dr. Katrina Gwinn-Hardy, in Chapter 32, discusses the autosomal recessively inherited disorder Wilson's disease, which can present with hyperkinetic or bradykinetic features. This is critical to diagnose, since treatment is available; if unrecognized, it can result in irreversible neurologic damage and even death by hepatopathy.

Abnormal gait is a common component of many neurologic disorders and especially the conditions covered in this text. Recognition of the prototypical types of gaits is critical to diagnosis. Dr. Frank Rubino applies his years of clinical savvy in Chapter 33, which addresses the broad topic of gait disorders.

Commonly, patients attribute their condition to some prior trauma. How often does this occur? Although subject to much debate, the topic of trauma-induced movement disorders is covered in Chapter 34 by Dr. Sotirios Parashos. Possibly the most difficult problem for clinicians is that of a psychogenic movement disorder. In Chapter 35, Drs. David Glosser and Matthew Stern have written a very reader-friendly review of what the practitioner should consider when approaching these patients. We conclude this book with an appendix that lists many of the organizations and foundations devoted to the disorders discussed in the book.

We wish to thank all the authors for their hard work and excellent contributions. We thank the Mayo Clinic Section of Scientific Publications, specifically Roberta Schwartz, Marlené Boyd, Reneé Van Vleet, and John Prickman, and Humana Press, specifically Paul Dolgert, for their diligent effort in publishing this text. We both would especially like to thank our wives, Laura Adler and Faye Ahlskog, as well as our children, Ilyssa and Jennifer Adler and Michael, John, and Matthew Ahlskog, for their support, encouragement, and patience during the long hours it took to complete this book. We hope that our combined efforts have created a readable text for the primary care physician that has distilled the tremendous advances made in the movement disorders field leading up to the millenium.

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2 Motor Speech Disorders: Clues to Neurologic Diagnosis

Joseph R. Duffy, PhD

Contents

INTRODUCTION

Speech is the most complex of innately acquired human motor skills, an activity characterized in normal adults by the production of about 14 distinguishable sounds per second through the coordinated actions of about 100 muscles innervated by multiple cranial and spinal nerves. The ease with which we speak belies the complexity of the act, and that complexity may help explain why speech can be exquisitely sensitive to nervous system disease. In fact, changes in speech can be the only evidence of neurologic disease early in its evolution and sometimes the only significant impairment in a progressive or chronic neurologic condition. In such contexts, recognizing the meaning of specific speech signs and symptoms can provide important clues about the underlying pathophysiology and localization of neurologic disease.

Neurologic speech disorders are known as *motor speech disorders*. They can be divided into two major categories, the *dysarthrias* and *apraxia of speech*. There are several types of dysarthria, but each reflects some disturbance of neuromuscular execution or control that can be attributed to weakness, incoordination, a variety of muscle tone abnormalities, or a variety of involuntary movements. The second category, apraxia of speech, is attributable to abnormalities in the programming of movements for speech rather than to their neuromuscular execution. (For readers relatively unfamiliar with concepts of speech production and speech disorders, Table 2-1 defines the basic components of speech production and some of the disturbances within each component that can be caused by neurologic disease.)

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Table 2-1
Definitions of the Basic Components of Speech Production and the Primary Speech Disorders Caused by Neurologic Disease

Basic components of speech production	
Respiration	Exhalatory air provides the transmission medium for speech. The diaphragm and thoracic and abdominal muscles are included. Disturbances at this level generally lead to abnormalities in loudness and in the number of words that can be uttered within a single exhalatory cycle.
Phonation	The production of voice. Laryngeal muscle activity to adduct the vocal cords during exhalation is required. Abnormal voice production, regardless of cause, is called dysphonia, a common problem in dysarthria.
Resonance	The quality of voice determined by the pharyngeal and velopharyngeal muscles, which control the shape of the vocal tract above the larynx, most crucially the degree to which the voice is transmitted through the oral cavity rather than the nasal cavity and vice versa. Excessive nasal transmission leads to a perception of hypernasality, frequently a problem in several types of dysarthria.
Articulation	The production of specific, distinguishable sounds of speech that, when combined, give speech its meaning. This is primarily a function of lingual, facial, and jaw muscle activity, although laryngeal and velopharyngeal movements, which produce many of the distinguishing features among speech sounds (phonemes), also contribute.
Prosody	The variations in pitch, loudness, and duration across syllables that help convey stress, emphasis, and emotion. They are a reflection of the combined activities of respiration, phonation, resonance, and articulation.
Primary speech disorders associated with neurologic disease	
Aphasia	A disturbance of language that can affect speech, comprehension of speech, reading, and writing (not discussed herein).
Apraxia of speech	A disturbance in the programming (selection and sequencing of kinematic patterns) of movements for speech production.
Dysarthria	Neuromuscular disturbance of strength, speed, tone, steadiness, or accuracy of the movements that underlie the execution of speech.

Table 2-2
Primary Tasks and Observations
for the Clinical Assessment of Motor Speech Disorders

Task	*Observation*
Nonspeech	
Jaw, face, tongue, palate	Symmetry at rest
	Symmetry and range of movement during spontaneous, reflexive, and imitated movement
	Involuntary movement at rest and during movement (e.g., fasciculations, tremor, dyskinesias)
Jaw, face, tongue	Strength against resistance
	Ability to smack lips, blow, click tongue (praxis)
Face, tongue	Atrophy
Palate	Movement during "ah"
	Gag reflex
Larynx	Sharpness of cough and grunted "uh"
	Ability to cough volitionally (praxis)
Speech	
Alternating motion rates "puh-puh-puh..." (lips) "tuh-tuh-tuh..." (anterior tongue) "kuh-kuh-kuh..." (mid and back tongue)	Rate, rhythm, range of movement, precision
Sequential motion rates "puh-tuh-kuh, puh-tuh-kuh..."	Sequential accuracy and speed (praxis)
Vowel prolongation ("ah")	Pitch, quality, loudness, duration, and steadiness
Conversation and reading	Rate, phrase length, prosody, voice quality, resonance, and precision

In this chapter, the basic methods for clinical examination of motor speech disorders are described and the distinctions among the disorders highlighted. Each disorder is reviewed in relation to its perceptible deviant speech characteristics, presumed underlying neuropathophysiology, associated signs and symptoms, and localization within the central or peripheral nervous system. Clues to distinguishing among the motor speech disorders also are highlighted. Finally, broad management strategies are reviewed.

CLINICAL ASSESSMENT OF MOTOR SPEECH DISORDERS

The clinical assessment of motor speech disorders often can be accomplished efficiently and simply. It includes (1) an oral mechanism examination, (2) a few examiner-specified speech tasks, and (3) assessment of conversation or reading aloud. A brief description of each task, its purposes, and the basic questions addressed during the task follow. Specific observations derived from these tasks are summarized during subsequent discussion of specific motor speech disorders. The clinical assessment is summarized in Table 2-2.

Oral Mechanism Examination

The oral mechanism is examined to assess the strength, speed, symmetry, range of movement, and steadiness of the jaw, face, tongue, and palate. These structures are observed at rest, during static postures, and during nonspeech movements.

1. *Jaw*. At rest, does the jaw hang excessively open? When opened, does it deviate to one side? Can the patient resist attempts by the examiner to open or close the jaw? Are there any adventitious movements at rest or when attempting to maintain mouth opening?
2. *Lower face*. At rest, are the angles of the mouth symmetrical? Are fasciculations apparent in the perioral area? Do the angles of the mouth elevate equally during an emotional smile or voluntary lip retraction? Is lip rounding symmetrical? Is there any symmetrical or asymmetrical air leakage during attempts to puff up the cheeks?
3. *Tongue*. Is the tongue normal in size bilaterally? Is there any grooving of the tongue indicative of atrophy? Are there any fasciculations? Are there any quick, slow, or sustained adventitious movements of the tongue at rest or on protrusion? Can the tongue be protruded fully, or does it deviate to the right or left on protrusion? Can it be wiggled to the left and right with normal rate, rhythm, and range of movement?
4. *Palate*. Are the arches of the soft palate symmetrical at rest? Can a gag be elicited with equal ease on both sides? Are there any rhythmical or arrhythmical movements of the soft palate? Does the soft palate elevate symmetrically during prolongation of the vowel "ah"?
5. *Larynx*. The integrity of vocal cord adduction can be crudely judged by asking the patient to cough or produce a glottal coup (a gruntlike "uh"). The cough and coup should be sharp, a gross indication of adequate adduction.

Nonverbal Oral Movements

For assessment of the ability to organize and program some fairly reflexive oromotor movements in a volitional way, the patient should be asked to cough, blow, smack the lips, and click the tongue. Any groping, off-target movements or tendency to verbalize the command rather than perform the movement suggests a nonverbal oral apraxia, a problem that tends to co-occur with apraxia of speech and limb apraxia. These apractic difficulties indicate left cerebral hemisphere disease.

Speech Alternating Motion Rates

Following examiner demonstration, the patient is asked to take a breath and repeat "puh-puh-puh-puh-puh…" as fast and as steadily as possible for 3 to 5 seconds. This is followed by similar repetitions of "tuh-tuh-tuh-tuh-tuh…" and "kuh-kuh-kuh-kuh-kuh…." These alternating motion rate (AMR) tasks permit judgments of rate, rhythm, precision, and range of motion of rapid movements of the lips, jaw, and tongue. Normal adults can produce an even rhythm at a rate of about 5 or 6 syllables per second.

Speech Sequential Motion Rates

Following examiner demonstration, the patient is asked to take a breath and produce "puh-tuh-kuh, puh-tuh-kuh, puh-tuh-kuh…" repetitively for 3 to 5 seconds. This task assesses the ability to program a sequence of speech movements rapidly and successively. After brief practice at slow rates, normal speakers accomplish this task rapidly and accurately. The ability to accurately sequence these sounds is the important observation on this task.

Vowel Prolongation

Following examiner demonstration, the patient is asked to take a deep breath and say "ah" for as long and as steadily as possible. Many normal adults can do this for 15 or more seconds, but 5 seconds usually suffices. The purposes of this task are to isolate the respiratory and laryngeal mechanisms and to evaluate their ability to produce a sustained tone at normal pitch, quality, loudness, duration, and steadiness.

Conversational Speech or Reading Aloud

Many important judgments about motor speech ability can be made when patients respond during the medical history and casual conversation or when they read aloud a standard paragraph. Such speech samples permit judgments of speech rate, phrase length, voice quality, resonance, and precision of articulation. Connected speech also permits assessment of the prosodic features of speech (defined below). Prosodic abnormalities often provide valuable clues that distinguish among the various motor speech disorders.

THE DYSARTHRIAS

The dysarthrias reflect neuromuscular disturbances of strength, speed, tone, steadiness, or accuracy of the movements that underlie the execution of speech. By definition, they do not include disorders attributable to anatomical deformities, faulty learning, or psychopathology.

Although dysarthria is manifested as a disorder of movement, it is important to recognize that sensorimotor integration (with tactile, proprioceptive, and auditory feedback representing the crucial sensory components) is essential to speech motor control. From this standpoint, most or all of the dysarthrias localized to the central nervous system should be thought of as sensorimotor rather than simply motor disturbances.

The dysarthrias can reflect disturbances at any one or a combination of the major components of the speech mechanism, including respiration, phonation, resonance, articulation, and prosody (see Table 2-1 for basic definitions). There are several subtypes of dysarthria. Subtype distinctions reflect (1) different auditory perceptual characteristics of speech, (2) different locations of central or peripheral nervous system lesions, and (3) different underlying neuromuscular dysfunctions. The ability to identify the type of dysarthria can be very useful for determining the underlying pathophysiology and lesion localization within the nervous system.

Each of the major categories of dysarthria is described below relative to its lesion locus, pathophysiology, nonspeech oral mechanism findings, and distinguishing speech characteristics. Localization and distinctive neuromuscular deficits are summarized in Table 2-3. Nonspeech oral mechanism findings are summarized in Table 2-4, and distinguishing speech characteristics are summarized in Table 2-5.

Flaccid Dysarthria

Lesion Loci

Lesions associated with flaccid dysarthria lie in the cell bodies, axons, or neuromuscular junction of the lower motor neurons that supply the speech musculature. These most often include cranial nerve V (trigeminal), VII (facial), X (vagus), or XII (hypoglossal) or the cervical and thoracic spinal nerves innervating the diaphragm and other respiratory muscles.

Table 2-3
Types of Dysarthria, Their Associated Lesion Loci, and the Neuromuscular Deficits That Probably Explain Their Distinctive Speech Characteristics

Dysarthria type	*Lesion locus*	*Distinctive neurologic deficit*
Flaccid	Lower motor neurons (cranial and spinal nerves)	Weakness
Spastic	Upper motor neurons (bilateral)	Spasticity
Ataxic	Cerebellum (cerebellar control circuit)	Incoordination
Hypokinetic	Basal ganglia control circuit	Rigidity, reduced range of movement
Hyperkinetic	Basal ganglia control circuit	Involuntary movements
Unilateral upper motor neuron	Upper motor neuron (unilateral)	Weakness, (?) incoordination, (?) spasticity
Mixed	Two or more of the above	Two or more of the above

Pathophysiology

With the exception of compensatory speech movements, the abnormal speech characteristics associated with flaccid dysarthria are all attributable to underlying weakness.[1] Because neurologic disease can affect the cranial or spinal nerves unilaterally or bilaterally, singly or in combination, the specific speech deficits depend on which lower motor neurons are affected. These effects are reviewed in the next section and are summarized in Table 2-3.

Nonspeech Oral Mechanism Findings and Distinguishing Speech Characteristics

Trigeminal Nerve (V)

Unilateral trigeminal lesions generally do not result in significant speech deficits. Bilateral trigeminal weakness, however, can significantly reduce jaw movement, especially closure, and severely affect articulatory contacts among the tongue, lips, and teeth. This can result in significant imprecise articulation of many sounds.

Facial Nerve (VII)

Lesions can cause imprecise articulation of sounds requiring facial movement. Facial weakness can affect sounds that require movement or contact of the upper and

[1] In persons with flaccid dysarthria associated with myasthenia gravis, speech may be normal if utterances are brief. However, if they are required to read aloud for several minutes without rest, their speech may deteriorate dramatically and any of the characteristics of flaccid dysarthria mentioned here may emerge. Speech may then recover to normal after a brief rest.

Table 2-4
Nonspeech Oral Mechanism Findings Frequently Associated With Each of the Major Motor Speech Disorders

	Dysarthria						
Physical findings	*Flaccid*	*Spastic*	*Ataxic*	*Hypokinetic*[a]	*Hyperkinetic*[b]	*Unilat UMN*[c]	*Apraxia of speech*
Hypoactive gag	+	-	-	-	-	-	-
Hypotonia	+	-	+	-	-	-	-
Atrophy	++	-	-	-	-	-	-
Fasciculations	++	-	-	-	-	-	-
Nasal regurgitation	++	-	-	-	-	-	-
Unilateral palatal weakness	++	-	-	-	-	-	-
Weak cough or coup	++	+	-	+	-	-	-
Dysphagia	+	+	-	+	+	+	-
Drooling	+	+	-	+	+	+	-
Pathologic oral reflexes	-	++	-	-	-	-	-
Hyperactive gag	-	++	-	-	-	-	-
Pseudobulbar affect	-	++	-	-	-	-	-
Masked facies	-	-	-	++	-	-	-
Tremulous jaw, lips, tongue	-	-	-	++	+	-	-
Reduced range of movement	-	+	-	++	-	-	-
Head tremor	-	-	+	-	+	-	-
Adventitious orofacial movements	-	-	-	-	++	-	-
Sensory tricks	-	-	-	-	++	-	-
Torticollis	-	-	-	-	++	-	-
Palatal myoclonus	-	-	-	-	++	-	-
Unilateral tongue and lower face weakness	-	-	-	-	-	++	+
Nonverbal oral apraxia	-	-	-	-	-	+	++

+, may be present but not generally distinguishing; ++, distinguishing when present; -, not usually present.
[a]Parkinsonian.
[b]For example, dystonia, chorea, myoclonus, tremor.
[c]Unilateral upper motor neuron.

Table 2-5
Primary Distinguishing Speech Characteristics Associated With Major Motor Speech Disorders

	Dysarthria						
Speech characteristics	*Flaccid*	*Spastic*	*Ataxic*	*Hypokinetic*	*Hyperkinetic*	*Unilat. UMN*	*Apraxia of speech*
Phonatory or respiratory hoarseness	+	-	-	+	-	+	-
Breathiness (continuous)	++	-	-	+	-	-	-
Diplophonia	++	-	-	-	-	-	-
Stridor	++	-	-	-	+	-	-
Short phrases	++	+	-	-	+	-	-
Strained-harsh voice	-	++	-	-	+	-	-
Excess loudness variation	-	-	++	-	++	-	-
Monopitch	+	+	-	++	+	-	+
Monoloudness	+	+	-	++	-	-	+
Reduced loudness	+	-	-	++	-	-	-
Vocal flutter	++	-	-	++	-	-	-
Forced inhalation or exhalation	-	-	-	-	++	-	-
Voice stoppages or arrests	-	-	-	-	++	-	-
Transient breathiness	-	-	-	-	++	-	-
Voice tremor	-	-	-	-	++	-	-
Myoclonic vowel prolongation	-	-	-	-	++	-	-
Intermittent strained or breathy voice breaks	-	-	-	-	++	-	-
Resonatory							
Hypernasality	++	+	-	+	-	-	-
Audible nasal emission	++	-	-	-	-	-	-
Intermittent hypernasality	-	-	-	-	++	-	-

Articulatory or prosodic							
Imprecise articulation	+	+	+	+	+	+	+
Slow rate	-	++	+	-	+	-	+
Slow and regular AMRs	-	++	-	-	-	-	-
Excess and equal stress	-	+	++	-	-	+	+
Irregular articulatory breakdowns	-	-	++	-	+	+	+
Irregular AMRs	-	-	++	-	++	-	-
Distorted vowels	-	-	++	-	++	-	+
Prolonged phonemes	-	-	+	-	+	-	+
Reduced stress	-	-	-	++	-	-	-
Short rushes of speech	-	-	-	++	-	-	-
Variable rate	-	-	-	++	+	-	-
Rapid “blurred” AMRs	-	-	-	++	-	-	-
Repeated phonemes	-	-	-	++	-	-	-
Palilalia	-	-	-	++	-	-	-
Prolonged intervals	-	-	-	-	++	-	+
Poorly sequenced SMRs	-	-	-	-	-	-	++
Articulatory groping	-	-	-	-	-	-	++
Articulatory substitutions	-	-	-	-	-	-	++
Attempts at self-correction	-	-	-	-	-	-	++
Articulatory additions	-	-	-	-	-	-	++
Automatic > volitional speech	-	-	-	-	-	-	++
Inconsistent errors	-	-	+	-	+	-	++
Increased errors with increasing length	-	-	-	-	-	-	++
Any component							
Rapid deterioration and recovery with rest	++	-	-	-	-	-	-

+, may be present but not generally distinguishing; ++, distinguishing when present; -, not usually present.
AMRs, alternating motion rates; SMRs, sequential motion rates; Unilat. UMN, unilateral upper motor neuron.

lower lips (p, b, m, w) or contact between the lower lip and the upper teeth (f, v). Unilateral paresis or mild-moderate bilateral weakness generally results in distortion of these sounds, whereas bilateral paralysis can completely prevent their production.

Vagus Nerve (X)

Branches of the vagus nerve supply the laryngeal and velopharyngeal muscles for speech. Unilateral lesions of the recurrent laryngeal nerve can produce vocal cord paralysis in the paramedian position, with resultant hoarseness, breathiness, and, sometimes, diplophonia (two perceived pitches rather than one, because the two vocal cords are vibrating at different rates). Bilateral lesions of the recurrent laryngeal nerve do not significantly alter phonation, but the resulting bilateral paramedian positioning of the vocal cords may seriously compromise the airway and cause inhalatory stridor. Unilateral lesions of the superior laryngeal nerve do not produce significant dysphonia, but bilateral lesions can restrict control of vocal pitch. Unilateral lesions of the pharyngeal branch of the vagus nerve can cause mild to moderate hypernasality and nasal air flow during production of consonants requiring pressure build-up in the mouth. Bilateral lesions can result in marked to severe hypernasality, audible nasal emission, and significant weakness of sounds requiring intraoral pressure (i.e., all non-nasalized consonant sounds); nasal regurgitation of liquids may occur during swallowing.

Intramedullary or extramedullary lesions (i.e., above the pharyngeal branch of the vagus nerve) can interfere with both phonation and resonance. The resultant unilateral or bilateral vocal cord paralysis leaves the affected vocal cord in the abductor position, causing short phrases and severe breathiness as well as hypernasality and nasal emission due to velopharyngeal weakness. The cough and glottal coup may be weak with unilateral or bilateral vocal cord paralyses.

Hypoglossal Nerve (X)

Unilateral or bilateral lesions produce weakness, atrophy, and fasciculations of the tongue. This results in articulatory imprecision of all consonants requiring lingual movements (e.g., as in do, to, no, key, go, sing, them, see, zoo, shoe, measure, chew, jump). Unilateral hypoglossal lesions usually result in only mild imprecision, whereas bilateral lesions can severely affect articulatory precision, sometimes including vowels.

Spastic Dysarthria

Lesion Loci

Spastic dysarthria is the result of central nervous system lesions that affect the upper motor neuron pathways bilaterally. Lesions can occur anywhere along these pathways, from their origin in the right and left cerebral hemispheres to their corticobulbar or corticospinal destinations in the brain stem and spinal cord. When vascular in origin, single lesions that produce the disorder are usually in the brain stem. Offending lesions outside the brain stem are usually multifocal or diffusely located in motor pathways of the cerebral hemispheres.

Pathophysiology

Upper motor neuron lesions usually produce a combination of weakness and spasticity. Some degree of weakness is usually evident in the speech musculature of persons with spastic dysarthria, and weakness certainly contributes to some of its

deviant characteristics. However, it is the resistance to movement and the tendency toward vocal cord hyperadduction generated by spasticity that give spastic dysarthria its distinctive speech characteristics.

Nonspeech Oral Mechanism Findings

Slow rate of orofacial movements, a hyperactive gag reflex, and pathologic oral reflexes (including suck, snout, and palmomental reflexes) are frequently present. Persons with spastic dysarthria often exhibit pseudobulbar affect, or pathologic laughter and crying due to poor reflex control. Drooling and dysphagia are common.

Distinguishing Speech Characteristics

Spasticity usually affects all components of speech and generates multiple abnormalities in speech, but the diagnosis of spastic dysarthria is often based on a gestalt impression generated by just a few distinctive speech characteristics. Among the most distinctive is a strained-harsh voice quality, often accompanied by reduced variability of pitch (monopitch) and loudness (monoloudness). These features reflect an apparent bias of spasticity toward hyperadduction of the vocal cords during speech as well as a slowing of the rapid muscular adjustments necessary for normal pitch and loudness variability. Also prominent and distinctive is slow rate of speech, often with classically slow and regular speech AMRs.

Ataxic Dysarthria

Lesion Loci

Ataxic dysarthria is associated with abnormalities of the cerebellum or cerebellar control circuit functions that influence the coordination of volitional movement. Cerebellar speech functions are not well localized, but prominent and lasting dysarthria is most often associated with bilateral or generalized cerebellar disease.

Pathophysiology

Reduced muscle tone and incoordination are associated with ataxic dysarthria. They appear largely responsible for the slowness of movement and inaccuracy in the force, range, timing, and direction of movements that underlie the distinctive speech characteristics of the disorder.

Nonspeech Oral Mechanism Findings

Oral mechanism size, strength, symmetry, and reflexes may be entirely normal. During oromotor AMR tasks, the repetitive movements of the jaw, face, and tongue are performed slowly and irregularly.

Distinguishing Speech Characteristics

Ataxic dysarthria is primarily a disorder of articulation and prosody. Although many abnormal speech characteristics can be present, only a few give speech its distinctive character. Conversational speech is often characterized by irregular breakdowns in articulation (giving speech an intoxicated or drunken character), sometimes including distorted vowels and excess variation in loudness. Speech rate is often slow, and some patients place excess and equal stress on each syllable produced, giving prosody a scanning character. Vowel prolongation is sometimes unsteady. Speech AMRs are often slow and distinctively irregular.

Hypokinetic Dysarthria

Lesion Loci

Hypokinetic dysarthria is associated with disease of the basal ganglia control circuit, probably bilateral in most instances. It is most frequently encountered in persons with Parkinson's disease or related parkinsonian conditions (see subsequent chapters for more complete discussion).

Pathophysiology

Rigidity, reduced force and range of movement, and slow (and sometimes fast) repetitive movements probably account for many of the distinctive deviant speech characteristics associated with hypokinetic dysarthria. These effects may be evident in the respiratory, laryngeal, velopharyngeal, and articulatory components of speech production.

Nonspeech Oral Mechanism Findings

The face may be masked or expressionless at rest and lack animation during social interaction. Reflexive swallowing may be reduced in frequency, and drooling may be apparent. Tremor or tremulousness may be apparent in the jaw, face, or tongue at rest and may be quite evident when the tongue is protruded. Size, strength, and symmetry of the jaw, face, and tongue may be surprisingly normal, but reduced range of motion may be evident in those structures on AMR tasks. Laryngeal examination may reveal bowing of the vocal cords. Reduced excursion of the chest wall and abdomen may be evident during quiet breathing and speech.

Distinguishing Speech Characteristics

The distinctive speech characteristics are primarily phonatory, articulatory, and prosodic. They often combine to convey a gestalt impression that speech is "flat," attenuated and unemotional, sometimes rapid but slow to start, and lacking in vigor and animation to a degree that is not explainable by weakness.

The very frequent phonatory abnormalities may include a tight breathiness with hoarseness and sometimes a vocal "flutter" (rapid voice tremor or tremulousness) that is most apparent during vowel prolongation. Reduced loudness and reduced pitch and loudness variability (monopitch and monoloudness) are also common, resulting in attenuation of normal prosodic variability. Hypernasality may be present but usually is not prominent.

Imprecise articulation is often evident secondary to reduced range of articulatory movement. Prolonged intervals between phrases reflect problems with initiation of movements for speech. Dysfluency characterized by sound prolongations or rapid sound repetitions sometimes occurs and can be prominent. When these dysfluencies include rapid or accelerating word and phrase repetitions, they are known as palilalia, a disorder rarely encountered in other types of dysarthria. Finally, hypokinetic dysarthria is the only type of dysarthria in which the speech rate may be rapid or accelerated and speech AMRs rapid and "blurred," with associated visually apparent reduced range of articulatory movements of the jaw, face, and tongue. Rapid or accelerating rate combined with prolonged intervals may be perceived as short rushes of speech.

Hyperkinetic Dysarthria

Lesion Loci

Similar to many hyperkinetic movement disorders, most varieties of hyperkinetic dysarthria probably derive from abnormalities in the basal ganglia circuitry (exception: palatal tremor).

Pathophysiology

Hyperkinetic dysarthrias reflect the influence of quick or slow, rhythmic or arrhythmic involuntary movements that interrupt, distort, or slow intended speech movements. Such movements can affect respiration, phonation, resonance, and articulation, singly or in combination, and often have prominent effects on prosody. Although movements may be present constantly, they may be clinically detectable only during speech, an observation that sometimes leads clinicians to misdiagnose the problem as psychogenic.

Hyperkinetic dysarthrias can be divided into subtypes according to the specific involuntary movements that underlie them. Most of these hyperkinesias can occur elsewhere in the body and sometimes in the bulbar muscles but without affecting speech. The various hyperkinetic dysarthrias are too numerous to review here, but the nonspeech oral mechanism and distinguishing speech characteristics associated with those that occur most commonly are briefly summarized below. They help give a sense of the variety of forms taken by hyperkinetic dysarthria, most of which are clearly distinctive from other types of dysarthria.

Nonspeech Oral Mechanism Findings and Distinguishing Speech Characteristics

Hyperkinetic Dysarthria of Chorea

The adventitious movements of chorea are quick, unsustained, and unpredictable in course. They may occur in the jaw, face, tongue, palate, larynx, or respiratory system. They may be evident at rest, during sustained postures or volitional movement, and during speech. The movements may cause dysphagia, but the oral mechanism may be normal in size, strength, and symmetry and without pathologic oral reflexes. They may range from subtle exaggerations of facial expression to movements so pervasive and prominent that the face never seems to be at rest.

Abnormal speech characteristics depend on the structures affected. Choreiform respiratory movements may lead to sudden forced inspiration and expiration. Quick, involuntary adductor or abductor movements of the vocal cords can produce sudden voice arrests with an intermittent strained quality or transient breathiness. Similar movements in the velopharynx can produce mild intermittent hypernasality and nasal emission. Choreiform articulatory movements can lead to brief speech arrests, irregular articulatory breakdowns, and articulatory imprecision, sometimes including distorted vowels, usually with visually apparent quick adventitious movements such as lip compression, facial retraction, darting of the tongue, and head jerking. Irregular AMRs and unsteadiness of vowel prolongation may be striking because of the unpredictable movements superimposed on the steady vocal tract. Prosodic abnormalities are prominent, and the flow of speech can have a jerky, fits-and-starts character, as if the patient were trying to say as much as possible before the next involuntary movement occurs.

Hyperkinetic Dysarthria of Dystonia

Dystonic speech movements are relatively slow and sustained, but quick movements are sometimes superimposed. Similar to chorea, dystonia may affect any or all parts of the speech system. Slow adventitious movements of the lips, tongue, or jaw may be evident at rest, during sustained postures and voluntary movements, and during speech. Dystonia in nearby structures that have minimal influence on speech, such as blepharospasm or torticollis, may be present. Patients sometimes develop sensory tricks or postures that help inhibit dystonic movements (e.g., a hand placed under the chin may inhibit a jaw-opening dystonia). It is important to recognize that dystonia can be confined to muscles for speech and swallowing and triggered only by the act of speaking.

When dystonia affects the larynx, it is variably known as spasmodic dysphonia or laryngeal dystonia. It may occur in isolation or be associated with dystonia in other craniofacial or neck structures. Laryngeal dystonia occurs in two forms, the more common of which is adductor spasmodic dysphonia, characterized by intermittent, waxing and waning, or constant strained-harsh voice quality, sometimes sufficient to cause voice stoppages. Less common is abductor spasmodic dysphonia, characterized by intermittent breathy or aphonic segments of speech. Laryngeal dystonia leading to spasmodic dysphonia is the most common focal, speech-induced hyperkinetic dysarthria.

Resonance abnormalities characterized by hypernasality are uncommon as the only manifestation of dystonic dysarthria, but they may be part of the clinical picture in spasmodic dysphonia. Focal face, jaw, and tongue dystonias can devastate articulation and lead to imprecise consonant articulation, distorted vowels, and irregular articulatory breakdowns. Dystonia anywhere within the speech system can have prominent effects on prosody, manifested as monopitch, monoloudness, short phrases, prolonged intervals and phonemes, inappropriate silences, and slow rate, creating a rhythm that reflects both excessive and insufficient prosody.

Hyperkinetic Dysarthria of Tremor

This dysarthria results from tremor, usually essential tremor, that affects speech muscles. The larynx is the most commonly affected speech structure, and organic or essential voice tremor is present in a substantial minority of patients with a neurologic diagnosis of essential tremor elsewhere in the body. Again, it is important to recognize that voice tremor may be present even though tremor is absent elsewhere in the body and may be evident only during speech. Tremor affecting speech can also arise from respiratory muscles and the velopharynx, tongue, and jaw.

Tremor of the arytenoid cartilages of the larynx may be apparent during laryngeal examination, and vertical oscillations of the larynx can sometimes be seen in the external neck during vowel prolongation. Similarly, tremor may be seen in the tongue, lips, or palate during vowel prolongation. It may also be evident during rest or sustained postures, and jaw tremor may be evident at rest or during mouth opening. In general, laryngeal and jaw tremors have the most significant impact on speech production.

Essential voice tremor is most easily detected during vowel prolongation and has a rhythmic, sinusoidal, quavering, waxing and waning character, with fluctuations in the range of 4 to 7 Hz. If the tremor is severe, there may be abrupt staccato voice arrests, sometimes leading to the designation "spasmodic dysphonia of essential voice tremor." When tremor is marked, speech rate may be slow. Prosodic abnormalities may be present,

reflected in altered pitch and loudness. Tremor in other speech muscles, especially those of the jaw, may also lead to slow rate and, sometimes, imprecise articulation.

Hyperkinetic Dysarthria of Palatopharyngolaryngeal Myoclonus

Caused by lesions in the dentatorubroolivary tracts in the brain stem and cerebellum (Guillain-Mollaret triangle), this unusual disorder (so-called palatal tremor) probably results from abnormal activity of a central pacemaker that generates jerks that are time-locked in different muscles. It is characterized by visually apparent, relatively abrupt, semirhythmic, unilateral or bilateral movements of the soft palate, pharynx, and laryngeal muscles at a rate of about 2 to 4 Hz. They are present at rest as well as during speech. Infrequently, the myoclonus includes the tongue, face, or nares. Patients may be unaware of the movements but may complain of a clicking sound or sensation in the ear secondary to myoclonic opening and closing of the eustachian tube.

The effects of the myoclonus on conversational speech often are not detectable, nor do they generally lead the patient to complain of speech difficulty. However, the effects can be very evident during vowel prolongation. They are heard as momentary semirhythmic arrests or tremor-like variations in voice at a rate that matches that of the visually evident palatal myoclonus (2 to 4 Hz). Rarely, intermittent hypernasality is evident. Articulation and prosody are usually normal, but brief silent intervals may occur if myoclonus interrupts inhalation, initiation of exhalation, or phonation. Because of the localization of the offending lesion, it may be accompanied by other types of dysarthria associated with posterior fossa lesions (e.g., flaccid, spastic, ataxic).

Unilateral Upper Motor Neuron Dysarthria

Lesion Loci

The label for this type of dysarthria defines its localization, which is the same as that of spastic dysarthria, except that the lesion is unilateral, not bilateral. Common sites of the lesion are the internal capsule (e.g., as in lacunar stroke) and the brain stem.

Pathophysiology

The underlying bases for this dysarthria may vary but probably reflect unilateral upper motor neuron weakness in most cases and, in some cases, various degrees of spasticity or ataxia-like incoordination. The variability in underlying substrates probably reflects different effects of lesions at different points along the upper motor neuron pathways between the cerebral cortex and the brain stem and spinal cord.

Nonspeech Oral Mechanism Findings

Most patients have a unilateral "central" facial (i.e., lower face only) weakness that may be evident at rest and during emotional smiling or voluntary lip retraction. Most also have unilateral tongue weakness, most obviously evident as deviation of the tongue on protrusion to the side of the lesion. Clinically obvious jaw, palate, or laryngeal weakness is unusual. Drooling from the weak side of the face may be apparent, and dysphagia can occur, although less frequently and severely than in spastic dysarthria.

Distinguishing Speech Characteristics

A number of speech abnormalities may be detected, but imprecise articulation is most prominent. Irregular breakdowns in articulation are not infrequent. Harshness, reduced loudness, hypernasality, and slow rate can occur but are usually mild. Speech AMRs can be mildly

slow and imprecise and sometimes irregular. In general, the dysarthria is rarely worse than mild to moderate, and when it is, an additional or different speech diagnosis (usually spastic or ataxic dysarthria or apraxia of speech) should be considered. The occasional association of ataxia-like irregular articulatory breakdowns and strained voice quality can sometimes suggest mixed dysarthria (e.g., multiple system involvement) even though the single offending lesion is only in the upper motor neuron pathway. The other results of the neurologic examination, particularly evidence of unilateral limb, face, and tongue weakness and the relative mildness of the dysarthria, often clarify the probable location of the lesion.

Mixed Dysarthrias

Combinations of two or more types of dysarthria occur more commonly than any of the single types just discussed. This reflects the basic fact that many neurologic diseases affect more than one component of the motor system. This does not minimize the value of distinguishing among single types of dysarthria or recognizing combinations when they occur. Recognition of a mixed dysarthria may help confirm expectations for a given disease, call into question a particular diagnosis, or raise questions about the presence of an additional condition. For example, amyotrophic lateral sclerosis can be associated with flaccid or spastic dysarthria and is "classically" associated with mixed flaccid-spastic dysarthria because the disease affects upper and lower motor neurons bilaterally. A type of dysarthria other than flaccid or spastic in amyotrophic lateral sclerosis should raise questions about the diagnosis or the possible existence of an additional disease.

A number of the conditions discussed in this book serve as additional examples. That is, with the exception of medication-related hyperkinesias causing hyperkinetic dysarthria, Parkinson's disease should be associated only with hypokinetic dysarthria. Additional types (e.g., spastic, ataxic, flaccid) in someone with a diagnosis of Parkinson's disease can suggest an alternative disease diagnosis, such as multiple system atrophy or progressive supranuclear palsy.

APRAXIA OF SPEECH

Apraxia of speech represents a disturbance in the selection and sequencing of kinematic patterns necessary to carry out intended speech movements. Its localization, underlying nature, and clinical characteristics differentiate it from the dysarthrias. Although it can affect any component of speech production, it is primarily a disturbance of articulation and prosody.

Lesion Loci

Praxis is a function of the dominant hemisphere, and apraxia of speech is associated with left (dominant) hemisphere disease in the great majority of affected persons. Lesions are nearly always in the distribution of the middle cerebral artery and usually involve the posterior portions of the frontal lobe (Broca's area), the insula, the parietal lobe, or the basal ganglia. Because it represents a left hemisphere pathologic condition, apraxia of speech is very frequently accompanied by aphasia, a disturbance of language that can affect verbal expression. It may also be accompanied by a unilateral upper motor dysarthria.

Pathophysiology

The clinical characteristics of apraxia of speech are not explainable on the basis of weakness, incoordination, disturbances of muscle tone, or involuntary movements. Conceptually, it is a disorder of programming of the temporal and spatial components of movements within and among the many muscles that generate the sequence of speech sounds necessary for intelligible speech.

Nonspeech Oral Mechanism Findings

The size, strength, symmetry, and reflexes of oral mechanism muscles may be normal. Right facial and tongue weakness is often present because of involvement of the nearby corticobulbar pathway, but such weakness does not explain the defining characteristics of the disorder. Frequently, but not invariably, a nonverbal oral apraxia is evident, characterized by groping and off-target efforts on nonspeech oromotor tasks (e.g., cough, blow, click tongue), sometimes with inappropriate accompanying verbalization (e.g., asked to blow, the patient with apraxia may say "blow" while trying to execute the act) (see Table 2-4 for summary).

Distinguishing Speech Characteristics

Articulation is usually imprecise or distorted and often accompanied by perceived substitutions, omissions, or even additions of sounds, with a tendency for more complex sounds and sound sequences to be more frequently in error. False articulatory starts and restarts, repetitive attempts at self-correction, sound and syllable repetitions, and visible and audible trial-and-error groping for correct articulatory postures are often apparent and reflect an acute awareness of errors. Speech rate is usually slow, consonants and vowels may be prolonged, and speech initiation may be delayed. Prosody is further disturbed by a tendency to equalize stress across syllables and words, with restricted alteration of pitch, loudness, and duration within utterances, conveying an impression that speech is being programmed and executed one syllable at a time. Rarely, but sometimes dramatically, prosody is altered in a manner that conveys an impression of a pseudoforeign accent.

In apraxia, unlike in dysarthria, a number of linguistic factors can affect speech accuracy. That is, error rates tend to increase for nonsense or unfamiliar words and as word length or complexity increases. For example, speech AMRs may be normal in apraxia, but sequential motion rates ("puh-tuh-kuh") are very often poorly sequenced. Simple words like "mom," "kick," and "baby" may be articulated without error, whereas more complex utterances like "statistical analysis," "stethoscope," and "we saw several wild animals" may elicit many apractic characteristics. In some patients, automatic speech (e.g., counting, social amenities, singing a familiar tune) may be more normal than more volitional or novel utterances. The patient with severe apraxia may be able to produce only a limited repertoire of sounds or words. Muteness can occur, although after stroke apractic mutism rarely persists for more than a few weeks if there are no accompanying language or other cognitive disturbances (see Table 2-5 for summary).

MANAGEMENT OF MOTOR SPEECH DISORDERS

The effective pharmacologic and surgical management of diseases that cause motor speech disorders can result in improvements in speech. When such treatments are not optimally effective, a number of additional behavioral, prosthetic, and instrumental management

approaches may maximize the patient's speech intelligibility or ability to communicate effectively and efficiently. Some of these approaches are common to all motor speech disorders, whereas others are unique to specific motor speech disorders. A few examples are provided here to illustrate the variety of strategies that can have a modest to dramatic effect on speech and communication ability. Identification of the best options usually requires careful speech assessment by a speech-language pathologist, and implementation often requires collaboration among a neurologist, otorhinolaryngologist, plastic surgeon, and prosthodontist.

Pharmacologic Management

Injection of botulinum toxin (Botox) into the vocal cords for the treatment of spasmodic dysphonias or into the jaw, face, or neck muscles for orofacial dystonias and torticollis often results in substantial and sometimes dramatic improvement in speech. Injection of botulinum toxin is often done for the primary or sole purpose of improving speech in persons with hyperkinetic dysarthria of dystonia, especially one of the spasmodic dysphonias, and for many it is the only or most effective treatment for the disorder.

Surgical Management

Persons with flaccid dysarthria and significant hypernasality and nasal emission may benefit from a pharyngeal flap or sphincter pharyngoplasty procedure that provides surgical obturation of the weak velopharyngeal mechanism. Similarly, certain thyroplasty procedures or vocal cord collagen injection may significantly improve weak voice in persons with vocal cord bowing, weakness, or paralysis.

Prosthetic Management

A number of mechanical and electronic prosthetic devices can improve speech or assist communication. A palatal lift prosthesis may reduce hypernasality and nasal emission in persons with flaccid or spastic dysarthria. Voice amplifiers may be very useful for patients in whom reduced loudness is the primary speech deficit, such as in flaccid or hypokinetic (parkinsonian) dysarthria. Pacing boards, metronomes, and delayed auditory feedback devices may be effective in slowing speech rate and reducing dysfluencies in persons with hypokinetic dysarthria. A wide variety of devices for augmentative and alternative communication can dramatically improve communication in severely affected patients, even when the underlying motor speech disorder does not change or worsens. They include picture, letter, and word boards as well as sophisticated computerized devices with a variety of output options, including synthesized speech.

Behavioral Management

Behavioral management can include efforts to improve physiologic support for speech, develop strategies for compensatory speaking or augmentative and alternative communication, and modify the environment and interactions in a way that facilitates communication. These approaches are too numerous to summarize here, but a few examples can illustrate their breadth.

Programs of vigorous vocal exercise may improve vocal loudness and quality for patients with weak voices associated with flaccid or hypokinetic dysarthria. Similar exercise for a weak face or tongue may lead to improved strength for articulation. Postural

adjustments and modified breathing strategies may improve respiratory support for speech and result in increased loudness and phrase length per utterance.

Slowing of speech rate may be the most effective strategy for improving speech intelligibility in dysarthria, regardless of type, and a number of behavioral strategies, with and without prosthetic assistance, can help patients accomplish this. Similarly, emphasizing the articulation of each sound or syllable may help slow rate and improve articulatory precision. Patients with apraxia of speech often require intensive and extensive drill work to develop reliable articulatory accuracy.

Behavioral strategies often focus on enhancing communication rather than improving motor speech per se. For example, the speaker may establish the topic of conversation explicitly at the outset of an interaction because it narrows the possible vocabulary and aids predictions about what will come next. When not understood, the patient may learn to point to the first letter of each word on a letter board before saying it. Listeners may learn to confirm their understanding of each word or phrase before moving on with the conversation, so that breakdowns can be repaired immediately.

To summarize, motor speech disorders often can be managed effectively, sometimes beyond what is accomplished through medical treatment of the underlying disease, especially if effectiveness is defined in terms of improving or maintaining the ability to communicate. This can be the case even in patients with chronic or degenerative disease. Some treatments reduce the underlying impairment or otherwise "normalize" speech. Others enhance speech intelligibility or efficiency or improve communication through the use of prosthetic or alternative communication devices or adaptive speaker or listener strategies.

SELECTED READING

Darley FL, Aronson AE, Brown JR. Differential diagnostic patterns of dysarthria. *J Speech Hear Res* 1969; 12: 246-269.

Darley FL, Aronson AE, Brown JR. Clusters of deviant speech dimensions in the dysarthrias. *J Speech Hear Res* 1969; 12: 462-496.

Duffy JR. *Motor Speech Disorders: Substrates, Differential Diagnosis, and Management,* Mosby, St. Louis, 1995.

McNeil MR. *Clinical Management of Sensorimotor Speech Disorders,* Thieme, New York, 1997.

Yorkston KM. Treatment efficacy: dysarthria. *J Speech Hear Res* 1996; 39: S46-S57.

Yorkston KM, Beukelman DR, Bell KR. *Clinical Management of Dysarthric Speakers,* Little, Brown & Company, Boston, 1988.