

Hyperkinetic Dysarthrias

"The flow of speech is often jerky, generated in fits and starts. As they proceed, patients are seemingly on guard against anticipated speech breakdowns, making compensation from time to time as they feel the imminence of glottic closure, respiratory arrest, or articulatory hindrance."

(Description of effects of chorea on speech—Darley, Aronson, and Brown [DAB]³⁶)

CHAPTER OUTLINE

- I. Anatomy and basic functions of the basal ganglia control circuit
- II. Clinical characteristics of basal ganglia control circuit disorders associated with hyperkinetic dysarthrias
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 - B. Myoclonus
 - C. Tics
 - D. Chorea
 - E. Ballismus
 - F. Athetosis
 - G. Dystonia
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- III. Etiologies
 - A. Toxic-metabolic conditions
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 - A. Distribution of etiologies, lesions, and severity in clinical practice
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 - F. Spasmodic torticollis (cervical dystonia)
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- K. Spasmodic dysphonia

VI. Cases**VII. Summary**

Hyperkinetic dysarthrias are a perceptually distinguishable group of motor speech disorders (MSDs) that are most often associated with diseases of the basal ganglia control circuit. They may be manifest in any or all of the respiratory, phonatory, resonatory, and articulatory levels of speech, and they often have prominent effects on prosody. Unlike most central nervous system (CNS)-based dysarthrias, they can result from abnormalities of movement at only one level of speech production, sometimes only a few muscles at that level. Their deviant speech characteristics are the product of abnormal, rhythmic or irregular and unpredictable, rapid or slow involuntary movements.

The designation of this dysarthria type as a plural disorder, the *hyperkinetic dysarthrias*, is justified by the existence of different kinds of involuntary movements that can cause them. Thus the singular term, hyperkinetic dysarthria, serves to identify a type of MSD that reflects the effects of involuntary movements on speech. Its subtypes designate the specific kind of involuntary movement. As with any classification scheme, there is overlap among subtypes, and clinical distinctions are sometimes difficult to make. Nonetheless, recognizing subtypes is useful for various reasons. This chapter's organization uses the notion of subtypes as a vehicle for discussing the shared features, as well as the remarkable variability

among speech problems caused by different involuntary movement disorders.

Hyperkinetic dysarthrias are encountered in a large medical practice at a higher frequency than other major single dysarthria types. Based on data for primary communication disorder diagnoses in the Mayo Clinic Speech Pathology practice, they account for 21.6% of all dysarthrias and 19.9% of all MSDs (see Figure 1-3). However, neurogenic spasmodic dysphonia and organic voice tremor accounted for approximately 75% of the hyperkinetic cases in the database. Thus if those two disorders are excluded, hyperkinetic dysarthrias are encountered somewhat less frequently than the other major single dysarthria types.

Hyperkinetic dysarthrias are perceptually distinguishable from other types of dysarthria, and observing the visible abnormal orofacial, head, and respiratory movements that underlie them often facilitates their diagnosis. The bizarreness of these involuntary movements and resultant speech abnormalities frequently raises suspicions about psychogenic etiology, so proper recognition of these dysarthrias can be essential for accurate medical diagnosis. Their diagnosis implies pathology in the basal ganglia or related portions of the extrapyramidal system or sometimes the cerebellar control circuit. The diversity of lesion loci associated with them (and movement disorders in general) reflects the diversity of abnormal movements that may occur in CNS disease and our limited understanding of their anatomy and pathophysiology.

The clinical features of hyperkinetic dysarthrias illustrate the devastating effects that involuntary movements and variations in muscle tone can have on voluntary movement. Hyperkinetic speech often gives the impression that normal speech is being executed but then is interfered with by regular or unpredictable involuntary movements that distort, slow, or interrupt it.

■ ANATOMY AND BASIC FUNCTIONS OF THE BASAL GANGLIA CONTROL CIRCUIT

The anatomy and functions of the basal ganglia control circuit and other portions of the CNS that may be implicated in this dysarthria type were discussed in Chapter 2 and reviewed in Chapter 7. The anatomy and functions of the circuit are the same as those discussed for hypokinetic dysarthria. They are reviewed briefly here, with specific focus on the possible anatomic and pathophysiologic bases of hyperkinetic dysarthrias.

The ventrolateral nucleus of the thalamus has a primarily excitatory effect on the cortex. The nuclei of the basal ganglia have complex interconnections

whose output is channeled to the cortex through the ventrolateral nucleus. The aggregate impulses from the basal ganglia have an inhibitory effect on the thalamus. As a result, they tend to inhibit cortical neuronal firing as well. Many hyperkinesias seem to result from a failure of these pathways to properly inhibit cortical motor discharges. This may happen in a number of ways. For example, the subthalamic nucleus normally exerts an inhibitory effect on the thalamus via its regulation of the globus pallidus. Destruction of the subthalamic nucleus causes reduced inhibitory output from the basal ganglia, with resultant increased thalamic and, subsequently, cortical firing. Consequently, uninhibited abnormal movement commands are "released" through the motor cortex to the corticospinal or corticobulbar pathways. Other movement disorders may have similar explanations. For example, a loss of neurons in the striatum, which normally modulates the globus pallidus, can result in abnormal involuntary movements.

Hyperkinesias can also result from a disruption of the normal equilibrium between excitatory and inhibitory neurotransmitters. For example, a relative increase in dopaminergic activity or a relative decrease in cholinergic activity within the circuit may result in hyperkinesia. Finally, the basal ganglia control circuit's role in movement disorders is demonstrated by the outcome of neurosurgical lesions or stimulators placed in the globus pallidus or ventrolateral nucleus of the thalamus. Such lesions can abolish tremor, rigidity, and involuntary limb movements by interrupting the loop through which the abnormal movements are generated.

Portions of the cerebellar control circuit can be similarly implicated in movement disorders. For example, lesions in cerebellar structures such as the dentate nucleus, or in brainstem structures such as the inferior olive or red nucleus, can alter the circuit's discharge patterns to thalamocortical pathways. The resultant input to the cortex can ultimately lead to abnormal motor cortex discharges through the corticospinal and corticobulbar pathways, with subsequent abnormal, involuntary patterns of movement.

■ CLINICAL CHARACTERISTICS OF BASAL GANGLIA CONTROL CIRCUIT DISORDERS ASSOCIATED WITH HYPERKINETIC DYSARTHRIAS

Some involuntary movements are normal. Startle reactions to loud noises, fear-induced tremor of the hands, shivering in response to cold, and jerking of body parts when falling asleep are all normal involuntary responses to certain intrinsic conditions or external stimuli. *Abnormal involuntary movements*

are those that occur in conditions where motor steadiness is expected. They can occur at rest, during static postures, or during voluntary movement. They are usually abolished by sleep and exacerbated by anxiety and heightened emotions. In some cases only specific movements trigger them, and sometimes adopting specific postures can inhibit them. The term *hyperkinesia* refers to these abnormal or excessive involuntary movements. The prefix "hyper" does not necessarily reflect excessive speed of voluntary movement; it indicates the presence of "extra" or involuntary movements that can range in rate from slow to fast. In fact, voluntary movements are generally slowed in body parts affected by hyperkinesias.

The precise location and underlying pathophysiology of many movement disorders are poorly understood. As a result, classifications are descriptive, often based on the speed of the involuntary movements (i.e., quick or slow hyperkinesias). Such divisions are often inadequate, because quick and slow involuntary movements occur on a continuum and often reflect a mixture of slow and quick components. However, some descriptive terms are useful, because they convey something about the

predominant character of the abnormal movement. In general, it is important to recognize that some hyperkinesias are rapid, unsustained, and unpatterned, whereas others are slower to develop, may be sustained for seconds (or longer), or may be prolonged to a degree that distorts posture in a constant or waxing and waning manner. Combinations of these characteristics are often apparent.

The varieties of movement disorders that are most relevant to understanding hyperkinetic dysarthrias are discussed as follows. Their basic characteristics are summarized in Table 8-1. Additional concepts that describe some associated nonspeech motor behaviors are also addressed.

Dyskinesias

Dyskinesia is a general term used to refer to abnormal, involuntary movements, regardless of etiology. *Orofacial dyskinesias* are involuntary orofacial movements that can occur without hyperkinesias elsewhere in the body. Most hereditary and acquired diseases that cause orofacial dyskinesias are associated with basal ganglia pathology.

Table 8-1 Categories of abnormal movement and their predominant rate and rhythm characteristics and presumed anatomic substrates. All but the movements under "Other" may be associated with hyperkinetic dysarthria.

<u>Designation</u>	<u>Speed</u>	<u>Rhythmicity</u>	<u>Anatomic Substrate</u>
Dyskinesia	Fast or slow	Irregular or rhythmic	Basal ganglia control circuit
Myoclonus	Fast or slow	Irregular or rhythmic	Cortex to spinal cord
Palatopharyngo-laryngeal	Slow	Regular	Brainstem (Guillain-Mollaret triangle)
Action	Fast	Irregular	Basal ganglia or cerebellar control circuit
Tics	Fast	Irregular but patterned	Basal ganglia control circuit
Chorea	Fast	Irregular	Basal ganglia control circuit
Ballism	Fast	Irregular	Area of subthalamic nucleus
Athetosis	Slow	Irregular	Basal ganglia control circuit
Dystonia	Slow	Irregular or strained	Basal ganglia control circuit
Spasmodic dysphonia	Slow	Irregular or sustained	?
Spasmodic torticollis	Slow	Irregular or sustained	? Basal ganglia control circuit
Blepharospasm	Slow	Irregular	? Midbrain, cerebellum, facial nucleus
Spasm	Slow or fast	Irregular	? Basal ganglia control circuit
Hemifacial spasm	Fast	Irregular	Facial nucleus cerebellopontine angle, facial canal
Essential tremor	Slow or fast	Rhythmic	? Striatum
Organic voice	Slow	Rhythmic	Cerebellar control circuit
Spasmodic dysphonia	Slow	Rhythmic	?
Other*			
Fasciculations	Fast	Irregular	LMN
Synkinesis	Fast or slow	Irregular	LMN
Facial myokymia	Intermediate	Rhythmic	LMN

CNS, Central nervous system; LMN, lower motor neuron.

*These abnormal movements may be visibly apparent in the speech muscles, but they are not considered hyperkinesias because they do not, by themselves, interfere with voluntary movement. Fasciculations, synkinesis, and facial myokymia may be associated with flaccid, not hyperkinetic, dysarthrias and are signs of LMN, not CNS pathology.

Orofacial dyskinesias are a common side effect of prolonged use of antipsychotic drugs, a condition known as *tardive dyskinesia (TD)*. The most common manifestation of TD is an *oro-buccal-lingual dyskinesia* that can be characterized by involuntary *stereotyped and repetitive lip smacking; pursing; puffing and retraction; tongue protrusion; or opening, closing, or lateral jaw movements*. TD can also affect respiratory function, with subsequent effects on speech.^{26,41,99} The emergence of hyperkinetic dysarthria caused by dyskinesias can represent TD, and its early recognition may help prevent a permanent TD if drug withdrawal or dosage modifications are possible.

Akathisia is a condition characterized by an inner sense of motor restlessness, which can be manifest by overt motor restlessness (e.g., weight shifting, pacing) to relieve the sensation. It can occur in parkinsonism and Parkinson's disease (PD) and sometimes in response to dopamine antagonist drugs (e.g., neuroleptic or antiemetic agents). It occurs in approximately 25% of patients with tardive dyskinesia.^{19,76}

Myoclonus

Myoclonus is characterized by involuntary single or repetitive brief, lightninglike jerks of a body part; if repetitive, jerks can be rhythmic or nonrhythmic. It cannot be inhibited willfully. Myoclonic movements can be confined to a single muscle or can be multifocal. They may occur spontaneously or be induced by visual, tactile, or auditory stimuli, or, sometimes, by voluntary movements. When brought on by movement, the condition is known as *action myoclonus (AM)*.

Myoclonus can be associated with lesions anywhere from the cortex to the spinal cord. It can occur in epilepsy (*myoclonic epilepsy*), where it is considered a component of a seizure. A common form of acquired myoclonus is *postanoxic myoclonus*, which can occur in the aftermath of cardiorespiratory arrest.^{99a}

Hiccups (singultus) are a form of complex myoclonus produced by a brief spasm of the diaphragm with subsequent adduction of the vocal folds. They commonly result from irritation of the peripheral sensory nerves in the stomach, esophagus, diaphragm, or mediastinum and may be associated with some toxic-metabolic conditions, such as uremia. Hiccupping may be a sign of medullary involvement in the region of the tractus solitarius, which has important respiratory control functions.⁵⁸

Palatal or palatopharyngolaryngeal myoclonus (PM) (also referred to as *palatal tremor*) is a unique, complex form of myoclonus associated with lesions

in the area of the brainstem known as the *Guillain-Mollaret triangle*. It can be associated with specific speech characteristics and is discussed later in a section on speech pathology.

Tics

Tics are rapid, stereotyped, coordinated, or patterned movements that are under partial voluntary control. They tend to be associated with an irresistible urge to perform them and often can be temporarily voluntarily suppressed. Simple tics are difficult to distinguish from dystonia or myoclonus. Complex tics, however, are coordinated and sometimes include jumping, noises, coprolalia, lip smacking, and touching. The prototypical tic condition is *Gilles de la Tourette's syndrome (TS)*, which is discussed later in a section under speech pathology.

Chorea

Chorea is characterized by involuntary rapid, non-stereotypic, random, purposeless movements of a body part. It may be present at rest and during sustained postures and voluntary movement. Choreiform movements can be subtle or can grossly displace body parts. They are sometimes modified by the patient to make them appear intentional in order to mask them and avoid embarrassment. Chorea can be degenerative (e.g., Huntington's chorea) or inflammatory or infectious in origin (e.g., Sydenham's chorea, encephalitis). It can occur in response to drugs, during pregnancy (*chorea gravidarum*), in association with some metabolic abnormalities, sometimes from neoplasm, and occasionally from vascular lesions of the subthalamic nucleus, striatum, or thalamus. Etiology can be undetermined. Rarely, the condition is benign and familial.²⁷

Ballismus

Ballismus involves gross, abrupt contractions of axial and proximal muscles of the extremities that can produce wild flailing movements; when unilateral, the condition is called *hemiballismus*. Lesions of the subthalamic nucleus are often responsible, and stroke is the most common cause.²⁷

Athetosis

Athetosis is characterized by slow, writhing, purposeless movements that tend to flow into one another. It is often considered a major category of cerebral palsy; when acquired, it may be caused by various conditions. Athetotic movements, especially when acquired, are often considered a combination

of chorea and dystonia; when chorea is predominant, the term *choreoathetosis* is sometimes used to describe them.⁶

Dystonia

Dystonia is a relatively slow hyperkinesia characterized by involuntary abnormal postures resulting from excessive cocontraction of antagonistic muscles. The primary abnormal movements tend to be slow and sustained, but there may be superimposed quick movements. The abnormal posture may involve torsion of a body part. Dystonias probably reflect a combination of dopaminergic and cholinergic overactivity in the basal ganglia.⁸⁹

Dystonia may involve only one segment of the body or contiguous regions (segmental). When only orofacial muscles are affected, the condition is often called *focal mouth dystonia* or *orofacial dystonia* or *dyskinesia*. Many occupational cramp syndromes, such as writers' cramp, are probably forms of dystonia. Dystonia can also be generalized and, when not associated with other neurologic deficits, is known as *primary generalized dystonia*.

Torticollis (cervical dystonia) is a segmental dystonia characterized by tonic or clonic spasms of the neck muscles, especially the sternocleidomastoid and trapezius. This causes deviation of the head to the right or left or, less frequently, backward (*retrocollis*) or forward (*antecollis*). Torticollis is generally considered a basal ganglia disease, and it is most often idiopathic. Cervical spine abnormalities and focal lesions in the putamen, caudate, thalamus, and globus pallidus, or their connecting pathways, have been associated with the condition.^{17,90}

Blepharospasm is characterized by a forceful, spasmodic, relatively sustained closure of the eyes. It can occur alone or with other dystonic disorders, particularly those involving orofacial muscles. Its biochemical and neuroanatomic mechanisms are poorly understood, but bilateral lid closure and blinking can be caused by stimulation of the mid-brain and cerebellum. It is usually taken as a sign of extrapyramidal disease; it has also been associated with disturbances in the thalamus, putamen, and lower pontine tegmentum.¹⁷

Spasm

Spasm is a general descriptive term that designates various muscular contractions. Tonic spasms are prolonged or continuous. Clonic spasms are repetitive, rapid in onset, and brief in duration.

Spasms are usually involuntary, even when they result from fear, anxiety, and conversion disorders. They often result in movement, but sometimes they limit motion (e.g., when attempting to avoid back

pain that may arise from movement). The term *spasm* is sometimes used to describe the abnormal postures seen in dystonia.

Hemifacial spasm is characterized by paroxysms of rapid, irregular clonic twitching of half of the face. The causative lesion affects the facial nerve in the cerebellopontine angle or facial canal and is often thought to result from a pulsating blood vessel (see Chapter 4). This interesting phenomenon illustrates that not all movement disorders result from primary lesions of the CNS control circuits or extrapyramidal system.

Tremor

Tremor is the most common involuntary movement. It involves the rhythmic (periodic) movement of a body part. It may be characterized as resting, postural, action, or terminal. *Resting tremor* occurs when the body part is in repose, *postural tremor* when the body part is maintained against gravity, *action tremor* during movement, and *terminal tremor* as the body part nears a target. Some clinically observable tremors are *physiologic*, meaning they are exaggerations of the normal tremor that exists in muscle, becoming of sufficient amplitude to be visible under conditions of extreme fatigue or emotion. Physiologic tremor is in the 10- to 12-Hz range until the fifth decade, after which it progressively decreases with age.⁶⁶ Toxic tremors can be induced by endogenous toxic states, such as thyrotoxicosis and uremia, or by medications, toxins, or during withdrawal from drugs or alcohol.

Essential (familial) tremor occurs with sustained posture and action and commonly affects the upper limbs, head, or voice. It tends to be reduced by alcohol.

Cerebellar tremor was discussed in Chapter 6. It occurs during sustained postures and action and terminally, and it is primarily due to involvement of the dentatorubrothalamic pathway; lesions of the superior cerebellar peduncle can cause severe tremor.⁶ *Wing-beating tremor* (frequently present in Wilson's disease) is a severe proximal postural tremor and is considered a special type of cerebellar tremor. It has a wing-beating appearance when the arms are held in an outstretched or abducted position.¹⁷

ETIOLOGIES

Hyperkinetic dysarthrias can be caused by any process that damages the basal ganglia control circuit or portions of the cerebellar control circuit or indirect activation pathways that can lead to hyperkinesias. Known causes include degenerative, vascular, traumatic, inflammatory, toxic, and metabolic

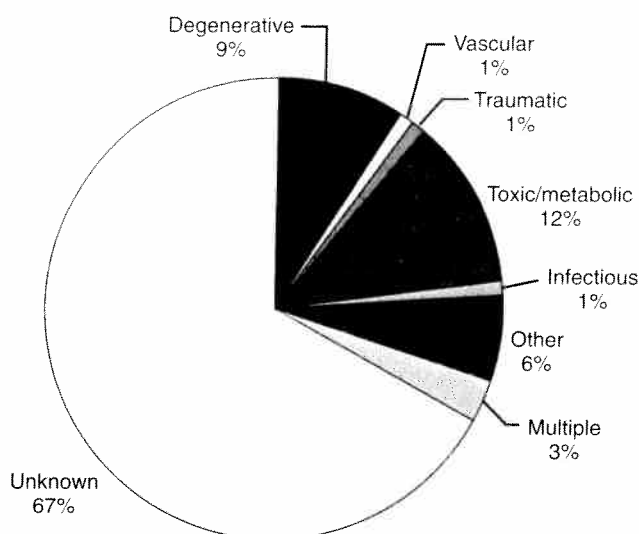


FIGURE 8-1 Distribution of etiologies for 141 quasirandomly selected cases with a primary speech pathology diagnosis of hypokinetic dysarthria at the Mayo Clinic from 1969-1990 and 1999-2001 (see Box 8-1 for details).

diseases. These broad etiologic categories are associated with hyperkinetic dysarthrias with varying frequency, but the exact distribution of etiologies is unknown. Idiopathic, toxic-metabolic, and degenerative causes are probably the most frequent etiologies, however (Figure 8-1 and Box 8-1).

Some of the common neurologic conditions associated with hyperkinetic dysarthrias with noticeably greater frequency than other dysarthria types are discussed as follows; much of the specific information provided is based on known causes of hyperkinesias in general, with an assumption that dysarthria can be caused by them as well. Conditions associated with hyperkinetic dysarthrias but that are more frequently associated with other dysarthria types (e.g., PD) are discussed in chapters dealing with those dysarthrias.

Toxic-Metabolic Conditions

Drugs that affect the balance of neurotransmitters in the basal ganglia are a common cause of acute or delayed-onset involuntary movements. *Neuroleptic* (meaning that which takes on the neuron) or *antipsychotic drugs*, whose actions block (antagonize) dopamine receptors, are the most frequent culprits. In some populations, drug-induced movement disorders are quite common. For example, the overall prevalence of tardive dyskinesia (TD) in schizophrenic patients on long-term treatment is 24%,⁹³ and the elderly who take antipsychotic drugs are at considerably higher risk for TD than younger people.⁶¹

TD can be associated with all classes of dopamine-blocking agents, including phenothiazines (e.g., chlorpromazine [Thorazine], thioridazine [Mellaril]) and butyrophenones (e.g., haloperidol [Haldol]).* Dopamine-blocking drugs used to control gastrointestinal disorders (e.g., metoclopramide [Reglan], prochlorperazine [Compazine]) can also cause TD.⁹⁴ Although the first step in treating TD is drug withdrawal, the dyskinesia often worsens in the first weeks after withdrawal and sometimes does not emerge until drug use is stopped. Drug withdrawal may be associated with remission of the dyskinesias, perhaps in 60% of patients, but it may take 3 to 5 years.⁷⁵ Acute dystonic reactions can also be triggered by dopamine receptor blocking agents.⁹⁴ Other drugs that can cause dyskinesias include levodopa, amphetamines, cocaine, tricyclic antidepressants, and phenytoin.⁹³

Chorea and dystonia can be caused by antiparkinsonian drugs and usually occur at the time of peak levodopa effect.⁷ These dyskinesias are frequently evident in limb and orofacial muscles, and sometimes they alter respiration.⁸⁷ L-Dopa-induced dyskinesias seem to reflect effects of excessive dopaminergic stimulation of certain striatal neurons, with subsequent thalamic disinhibition and excessive positive feedback to precentral motor areas, resulting in excessive, involuntary (hyperkinetic) movements.⁸¹

*A newer generation of "atypical" antipsychotic drugs (e.g., clozapine, risperidone, olanzapine, amisulpride) seem to carry less risk for TD than conventional neuroleptics, even in the elderly.^{40,71,101}

box
8-1

Etiologies for 141 quasirandomly selected cases with a primary speech pathology diagnosis of hyperkinetic dysarthria at the Mayo Clinic from 1969-1990 and 1999-2001. Percentage of cases under each heading is given in parentheses. Specific etiologies under each heading are ordered from most to least frequent. Cases with isolated essential voice tremor or isolated neurogenic spasmodic dysphonia are not included, because etiology is nearly always idiopathic in those disorders; if they were included in these data, the percentage of unknown etiologies would be substantially higher.

Unknown (67%)

Orofacial dyskinesia, dystonia, or tremor (21%)
(other descriptors include oromandibular dystonia, lingual-mandibular dystonia, focal dystonia, orallaryngeal dystonia, lingual-pharyngeal dystonia, palatal-pharyngeal-lingual-facial dystonia, oral dyskinesia, buccolingual dyskinesia, extrapyramidal facial movement disorder)
Essential tremor (11%)
Segmental dystonia or tremor (7%)
Spasmodic torticollis or retrocollis or antecollis (6%)
Face or neck or axial dyskinesia (4%)
Chorea (2%)
Meige's syndrome (2%)
Dystonia, not otherwise specified (2%)
Other (12%)
Movement disorder; dyskinesia; extrapyramidal syndrome; acquired basal ganglia disorder; myoclonus; indeterminate brainstem lesion; isolated hyperkinetic dysarthria; torticollis and facial dystonia; respiratory-laryngeal dystonia; abdominal myoclonus; action dystonia; focal seizure disorder; generalized dystonia

Toxic or Metabolic (12%)

Tardive dyskinesia (7%)
Drug-induced dyskinesia (2%)
Other (3%)
Dialysis encephalopathy; hepatic encephalopathy; hypoparathyroidism

Degenerative (9%)

Huntington's chorea (3%)
Other (6%)
Unspecified degenerative CNS disease; dystonia musculorum deformans; amyotrophic lateral sclerosis; multiple sclerosis; Parkinson's disease

Multiple (3%)

Multiple sclerosis + drug-induced parkinsonism; familial ataxia + alcoholism; senile chorea + stroke; tardive dyskinesia + stroke + anxiety

Infectious (1%)

Sydenham's chorea

Trauma (1%)

After cerebellar tumor removal (ataxia and palatal myoclonus)

Vascular (1%)

Brainstem stroke

Other (6%)

Tourette's syndrome; myoclonic epilepsy; focal seizure disorder; familial tremor-dystonia syndrome; seizure-related generalized polymyoclonus; focal myoclonus; paraneoplastic encephalopathy; no neurologic diagnosis

CNS, Central nervous system.

Chorea, including choreiform facial movements, can be associated with oral contraceptive use, alcohol withdrawal, and certain metabolic conditions including hyperthyroidism, anoxic or hepatic encephalopathy, hypernatremia, hypoglycemia, choreoacanthocytosis, and hypoparathyroidism.^{3,27,60}

Action or postural tremor can be associated with valproic acid, lithium, and theophylline derivatives and may occur during alcohol or other drug withdrawal states. Tremor (and dystonia) may also occur in Wilson's disease.⁷⁷

A number of toxins can cause myoclonus (e.g., mercury, lead, strychnine, marijuana), as well as a number of drugs (e.g., psychiatric medications, antiinfectious agents, narcotics, anticonvulsants, anesthetics, cardiac medications, and antihistamines).²⁸

Degenerative Diseases

Huntington's disease is an inherited autosomal dominant degenerative CNS disorder. Because it has complete penetrance, half of the offspring of individuals with the gene are affected. It usually begins insidiously by the fourth or fifth decade, with progression to death within 10 to 20 years.²⁷ Cellularly, there is severe loss of neurons in the caudate nucleus and putamen and diffuse neuronal loss in the cortex. Functionally, positron emission tomography (PET) has shown impaired activity of the striatum and its frontal projection areas.^{14,98} The disease's most characteristic clinical feature is chorea, which can be generalized, but it is sometimes initially manifest only in the face or hands. Dementia, depression, personality changes, and attention deficits are also

characteristic, and dysarthria and dysphagia are common.

Primary generalized dystonia (also called *idiopathic torsion dystonia* or *dystonia musculorum deformans*) usually results from autosomal dominant inheritance, with marked variation in clinical expression. It is often associated with gait abnormalities and postural deformities in the neck, trunk, and extremities. Usually beginning in childhood as a focal dystonia, it eventually spreads over months or years to affect other body parts.^{88,94}

Involuntary movements may also occur in degenerative diseases that primarily affect cognitive abilities. For example, orofacial dyskinesia in the elderly tends to be associated with dementia.³³ Dyskinesia, especially orofacial dyskinesia, has been reported in 17% of individuals with a diagnosis of Alzheimer's disease.⁸⁰

Infectious Processes

Sydenham's chorea is associated with streptococcal infection or rheumatic fever; it occurs in 26% of patients with rheumatic fever. It affects mostly the young and usually resolves in a relatively short time, but it is sometimes persistent.²⁵ Single photon emission computed tomography (SPECT) has documented hyperperfusion of the basal ganglia in people with recent onset of symptoms.¹³

Other infectious causes of chorea include diphtheria, rubella, systemic lupus erythematosus, and acquired immune deficiency syndrome (AIDS).^{85,89}

Vascular Disorders

Although stroke is the usual cause of hemichorea and hemiballismus, vascular lesions are not a common cause of hyperkinesias.⁵² Nonetheless, stroke or other vascular disturbances in the basal ganglia control circuit, and sometimes the cerebellar control circuit, can lead to movement disorders and hyperkinetic dysarthria. For example, dystonia can result from putaminal stroke^{49,89}; chorea, dystonia, athetosis, or action tremor can result from lateral-posterior thalamic stroke⁶⁵; and blepharospasm, *Meige's syndrome* (see later discussion), and palatal myoclonus have been reported in brainstem stroke or hypoxic encephalopathy.^{37,60a}

Neoplasm

Tumors of the basal ganglia and thalamus have been associated with chorea and dystonia.^{89,90}

Other

Movement disorders, particularly dystonias, are often considered *primary* or *unassociated with a*

known cause or other neurologic abnormalities. A genetic basis for some primary dystonias has been established and is suspected for others. Primary dystonia can be generalized or focal.⁹⁴ Meige's syndrome is a primary focal cranial dystonia characterized by a combination of blepharospasm and oromandibular dystonia. Many spasmodic dystonias are considered primary focal dystonias.

Although uncommon, some hyperkinetic movement disorders are paroxysmal or evident only during brief (minutes to hours) recurring episodes.* Examples of these disorders include *paroxysmal kinesigenic choreoathetosis*, which is precipitated by sudden movement; *paroxysmal exercise-induced dystonia*, which is induced by prolonged exercise; and *paroxysmal (nonkinesigenic) dystonic choreoathetosis*, which can be triggered by various factors, such as alcohol, coffee, tea, fatigue, stress, and anxiety. Many of these disorders are thought to represent "channelopathies," or dysfunction of ion channels involved in neurotransmission. Cases are frequently idiopathic and sporadic, but a family history with autosomal dominant inheritance is common. Occasionally, paroxysmal dyskinesias are symptomatic of other conditions (e.g., multiple sclerosis, progressive supranuclear palsy [PSP], perinatal hypoxic encephalopathy, endocrine disorders, diabetes mellitus, vascular lesions, traumatic brain injury). In at least some of these disorders, speech can be affected during episodes.¹⁵

TS is characterized by motor and vocal tics. The disorder has a significant genetic component, and its signs are always apparent before adulthood. Its vocal and speech characteristics are discussed in a section under speech pathology.

Abnormalities of the dental arch in edentulous elderly people have been associated with involuntary chewing movements. Disruption of dental proprioception has been suggested as a general explanatory mechanism.⁶⁰

Facial dyskinesias may be observed in schizophrenic individuals, and they can occur before the introduction of antipsychotic drugs.⁶⁰

Chorea gravidarum is a rare, benign choreiform disorder that occurs during pregnancy, most frequently in women with chronic rheumatic heart disease.⁴⁴

■ SPEECH PATHOLOGY

Distribution of Etiologies, Lesions, and Severity in Clinical Practice

Box 8-1 and Figure 8-1 summarize the etiologies for 141 quasirandomly selected cases seen at the Mayo

*Episodic ataxias are discussed in Chapter 6.

Clinic with a primary speech pathology diagnosis of hyperkinetic dysarthria. Cases with organic voice tremor and neurogenic spasmodic dysphonia were not included in the review, because their etiology is nearly always unknown. The cautions expressed in Chapter 4 about generalizing these data to the general population of patients with hyperkinetic dysarthrias or to all speech pathology practices also apply here.

The data establish that hyperkinetic dysarthrias can result from several medical conditions and that distribution of the etiologies is quite different from that for most other dysarthria types. Sixty-seven percent of the cases were of undetermined etiology, with toxic or metabolic causes accounting for an additional 12% of the cases. These percentages illustrate the elusive nature of the neuroanatomic bases of movement disorders, and they suggest that their causes often lie in neurochemical abnormalities rather than structural lesions.

The nature and muscular locus of involuntary movements of unknown etiology illustrate the heterogeneity that exists in this group of speech disorders. The abnormal movements were given numerous descriptive labels, including dyskinesia, dystonia, torticollis, retrocollis, antecollis, chorea, tremor, myoclonus, AM, and action dystonia. In some cases the neurologic diagnosis was limited to general labels such as "movement disorder," "extrapyramidal syndrome," and "acquired basal ganglia disorder."

The largest single diagnosis for the 141 cases was orofacial dyskinesia of unknown etiology (21%). This means that the patients' movement disorders were confined to the face, jaw, tongue, pharynx, or larynx. This highlights the predilection of many movement disorders for the orofacial muscles, the likelihood that many generalized movement disorders may be manifest first in the orofacial area, and the importance of recognizing the meaning of abnormal orofacial movements and associated dysarthria as signs of neurologic disease. The prevalence of movement disorders that are limited to the head and neck muscles in people with dysarthria is further illustrated by the frequency of cases with spasmodic torticollis, retrocollis, or antecollis (6%), face or neck or axial dyskinesia (4%), and tardive dyskinesia (7%). Thus approximately 85% of the group had a dysarthria in which the underlying movement disorder was not evident in the limbs. This percentage would be even higher if organic voice tremor and neurogenic spasmodic dysphonias were included in the sample.

Most cases with toxic or metabolic etiology were drug related, most often involving neuroleptic or anticonvulsive medications. Although most were delayed in onset (tardive), some occurred soon after

medication was started. In contrast to the distribution of etiologies for other dysarthria types, drugs were the most frequent known cause of hyperkinetic dysarthrias in this sample.

Huntington's chorea was the most frequent degenerative disease (3% of the entire sample). The remaining etiologic categories (infectious, trauma, vascular) were not frequently represented but did contain conditions with established and prominent associations with hyperkinesias (e.g., TS, myoclonic epilepsy, Sydenham's chorea).

To what extent was dysarthria the only manifestation of an involuntary movement disorder? A substantial minority of the sample had involuntary movements that were confined to the orofacial muscles but present at rest or during nonspeech movements. Several cases had a dysarthria in which the hyperkinesia was triggered by speech and not present during orofacial movements other than speech (this percentage would be considerably higher if cases with organic voice tremor and neurogenic spasmodic dysphonia were included). Thus *movement disorders may become clinically evident only during speech*. Unfortunately, when this is the case, the problem is often diagnosed as psychogenic, and affected people may have a painfully long history of repeated psychiatric assessments and treatment, without any connection emerging between psychopathology and the speech disorder and without benefit from psychotherapy, behavioral interventions, or psychotropic medications. Such cases illustrate a lack of understanding about the possible connection between speech disorders and neurologic disease, especially when speech is the presenting and only obvious physical problem.

What speech structures were involved in these cases, and how often was only a single structure involved? Table 8-2 summarizes the percentage of

table 8-2 Percentage of patients with involvement of the jaw, face, tongue, palate, larynx, and respiratory muscles—singly and in combination—for 86 cases with hyperkinetic dysarthrias (excluding organic voice tremor and spasmodic dysphonia)

Structure Involved	% in Combination	
	with Other Structures	% Isolated
Jaw	52	3
Face	67	1
Tongue	56	3
Palate	13	0
Larynx	44	1
Respiratory	7	1

cases (among the first 86 cases in the group of 141 reviewed in Box 8-1) in which a clear indication was given about involvement of the jaw, face, tongue, palate, larynx, and respiratory muscles, and the percentage of cases in which only one of those structures was affected. It is clear that more than one speech structure is involved in most cases and that face, tongue, and jaw involvement are most frequently recognized (note again that organic voice tremor and spasmodic dysphonia were not included in the sample, so laryngeal involvement is underrepresented). Combined jaw, face, and tongue hyperkinesia was the most frequently recognized combination of involved structures. It is also apparent that only a single speech structure may be involved in hyperkinetic dysarthria. Nine cases (10%) had only one structure affected; the palate was the only structure not affected in isolation. In a few cases, involuntary movements in the singly involved structure were induced only by speech; for example, two cases with jaw dystonia had abnormal jaw movements only during speech. Thus these data suggest that involuntary movements underlying dysarthria usually involve more than a single speech structure, but they sometimes affect only a single speech structure and sometimes occur only during speech.

Precise anatomic localization of lesions for the patients in this sample, as predicted by the high frequency of undetermined etiologies, was sparse. A majority of those who had computed tomography (CT) or magnetic resonance imaging (MRI) had no identifiable pathology, and detected abnormalities were often nonfocal or not necessarily related to the movement disorder. For example, several cases had evidence of general cerebral or cerebellar atrophy, or both; bilateral white matter changes; or ventricular dilatation. A few had evidence of cortical lesions, but there was no common site among them. Some did have evidence of basal ganglia pathology, and a few had evidence of cerebellar pathology or a thalamic lesion. Even in cases with identifiable lesions in the basal ganglia, cerebellum, and thalamus, it was sometimes concluded that they were not directly responsible for the movement disorder (e.g., one patient's movement disorder was most likely due to tardive dyskinesia). Thus although CT and MRI sometimes reveal abnormalities, the connection between the identified lesion and the movement disorder is not always clear.

This retrospective review did not permit a clear delineation of dysarthria severity. However, in those patients for whom a judgment of intelligibility was explicitly stated (60% of the sample), 49% had reduced intelligibility. The degree to which this figure accurately estimates the frequency of intelligibility impairments in the population with hyperki-

netic dysarthrias is unclear. It is likely that many patients for whom an observation of intelligibility was not made had normal intelligibility, but the sample probably contains a larger number of mildly impaired patients than is encountered in the typical rehabilitation setting. In addition, reduced intelligibility is only one measure of deficit and may not accurately represent degree of handicap or disability. For example, many hyperkinetic dysarthrias significantly reduce efficiency (speed) of communication without affecting intelligibility, and the visible, bizarre involuntary movements that are responsible for many aspects of the dysarthria can have devastating social and emotional consequences. The relatively low frequency of intelligibility impairments in this dysarthria is fortunate, but to conclude that the disorder often does not have a significant impact on verbal and nonverbal communication, and its emotional and social consequences, probably grossly underestimates its impact on affected individuals.

Finally, movement disorders and hyperkinetic dysarthria can occur in conditions that also affect cognitive functions (e.g., Huntington's disease). Of the patients in the sample whose cognitive abilities were explicitly commented on or formally assessed (80% of the sample), 20% had some impairment of cognitive ability.

Patient Perceptions and Complaints

Patient complaints often depend on the type of movement disorder and the level of the speech system it affects. Those with nonrhythmic hyperkinesias (e.g., chorea, dystonia) affecting the jaw, face, tongue, and larynx tend to describe their speech as slurred, slow, halting, or "hard to get out." Somewhat surprisingly, those with hyperkinesia at several levels of the speech system may not be aware of the abnormal movements, even when they are visibly apparent to the examiner. They may, however, recognize their inability to maintain a steady jaw, face, or tongue posture when requested by the examiner. Failure to spontaneously complain about orofacial hyperkinesias is more frequent in patients whose hyperkinesia is apparent only during speech, chewing, or swallowing (i.e., they complain of difficulty with speech but not the underlying abnormal movement). Chewing and swallowing complaints are common in chorea and dystonia.

Patients whose hyperkinesia is limited to a single or a few structures may complain of abnormal movements, both at rest and during speech. Complaints about abnormal movements at rest may predominate in patients whose hyperkinesias are mild or can be suppressed temporarily during speech. These complaints include feelings of tightness in affected structures, an inability to move a structure, an inability to

control or inhibit abnormal movements, or a sense that the structure simply "doesn't work right." Some patients report being able to suppress the abnormal movements for a time but find that they return with a vengeance when their efforts cease.

Patients with prominent laryngeal hyperkinesias (usually associated with tremor or dystonia) often complain that their voice is shaky, tight, closes off, or does not want to come out. Because of increased resistance to airflow with laryngeal spasm during speech, they may complain of shortness of breath or physical exhaustion during speech and associate it with respiratory difficulty. When the problem is isolated to the larynx, however, the patient usually does not note similar fatigue during strenuous nonspeech physical activities. Patients with respiratory hyperkinesias that are triggered only by speaking may be unaware of the locus of the problem, even when they are acutely aware of their abnormal speech.

Some patients learn that the severity of orofacial dystonic posturing or movements can be reduced or eliminated by certain tactile or proprioceptive *sensory tricks*. For example, a patient with torticollis may learn that bringing the hand to the chin or the back of the head allows them to posture the head more normally; a patient with involuntary jaw opening may learn that lightly touching the hand to the jaw may prevent the movement. With increasing severity or duration of the disorder, however, the facilitory effect of these stimuli diminishes. The term "sensory tricks" is descriptive, and the real mechanism for their effect is unknown.⁴³ Nonetheless,

patient reports of previously successful sensory tricks, or observation of them during examination, are useful diagnostically, because they are rarely developed in other dysarthria types or in psychogenic speech disorders characterized by abnormal movements.

The following sections review the primary oral mechanism and speech characteristics associated with each of a number of movement disorders known to underlie the hyperkinetic dysarthrias. Related acoustic and physiologic findings are summarized when appropriate.

Chorea

Nonspeech Oral Mechanism

The jaw, face, tongue, and palate are usually normal in size, strength, and symmetry. The gag reflex is usually normal, and pathologic oral reflexes are usually absent. Drooling is occasionally observed, and chewing and swallowing difficulties are not uncommon. The most striking abnormality is motor unsteadiness and, often, easily observed choreiform movements. At rest or during attempts to maintain steady orofacial postures, *quick, unpredictable, involuntary movements* may occur. They can range from subtle exaggerations of facial expression to movements that are so pervasive and prominent that affected structures seem never to be at rest (Figure 8-2). Difficulty recognizing these movements as hyperkinesias occur when (1) movements are subtle



FIGURE 8-2 Quick, involuntary head, jaw, face, lip, and eye movements in a woman with generalized chorea. The depicted movements in panels **A** and **B** were brief and separated from each other by less than 1 second.

and infrequent, because they may be difficult to distinguish from normal unsteadiness, and (2) patients display motor impersistence or cognitive impairments that raise doubts about their ability to sustain adequate effort on the task.

Speech

Conversation, reading, and speech alternate motion rates (AMRs) are useful for eliciting the unpredictable breakdowns of articulation and the abnormalities of rate and prosody that may predominate. Vowel prolongation is indispensable, because it is an opportunity to observe fluctuations in the steady state of the vowel induced by choreiform movements. The open vowel "ah" is particularly useful, because adventitious movements of the jaw, face, tongue, and palate can be easily observed and heard during the sound.

Careful visual observation of the patient during speech is important. It provides confirmatory evidence of abnormal movements as the source of the speech deficit and permits the identification of at least some of the structures involved in the movement disorder.

Table 8-3 summarizes the neuromuscular deficits presumed by DAB^{34,36} to underlie the dysarthria of chorea. Nearly all aspects of movement may be disturbed. Involuntary movements may alter the direction and rhythm of movement, and they generally slow rate. Force and range of individual and repetitive movements may vary from reduced to normal to excessive depending on the presence or absence of choreic movements at the moment and the relationship of their direction to that of the intended voluntary speech gesture. Muscle tone may be excessive and, when it is, it tends to be biased. The relation-

ship of these characteristics to specific deviant speech characteristics is discussed in the following section, as are findings from relevant acoustic and physiologic studies.

Clusters of Deviant Dimensions and Prominent Deviant Speech Characteristics

DAB³⁵ identified several clusters of deviant speech dimensions in their patients with chorea, but a complete review of each is unnecessary to appreciate the major features of the disorder. Only those clusters that are most prominent and distinctive of the hyperkinetic dysarthria of chorea are addressed. Here the focus primarily is on the distinct clusters and most deviant or unique speech characteristics, as well as their relationship to movement abnormalities at each level of the speech system. These characteristics are useful to understanding the disorder's underlying neuromuscular deficits, the components of the speech system that tend to be most prominently involved, and the features that help to distinguish the hyperkinetic dysarthria of chorea from other dysarthria types. The most deviant speech characteristics encountered in the dysarthria of chorea are summarized in Table 8-4.

Respiration. Choreiform movements that affect respiration during speech can be reflected in *sudden, forced, involuntary inspiration or expiration*. Although not pervasive, and not necessarily severe, this feature was not encountered in any other dysarthria type by DAB.³⁶

Phonation. Patients may exhibit *harsh voice quality, excess loudness variations, and a strained-strangled voice quality*. These features correlated

table 8-3 Neuromuscular deficits found in hyperkinetic dysarthria associated with dystonia and chorea

Direction	Rhythm		Rate		Range		Force	Tone
	Individual Movements	Repetitive Movements	Individual Movements	Repetitive Movements	Individual Movements	Repetitive Movements	Individual Movements	Muscle Tone
Dystonia	Inaccurate due to slow involuntary movements	Irregular	Slow	Slow	Reduced to normal	Reduced to normal	Normal	Excessive (biased)
Chorea	Inaccurate due to quick > slow involuntary movements	Irregular	Slow	Slow	Reduced to excessive	Reduced to excessive	Reduced to excessive	Often excessive (biased)

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

table
8-4

The most deviant speech dimensions encountered in the hyperkinetic dysarthria of chorea by Darley, Aronson, and Brown,³⁴ listed in order from most to least severe. Also listed is the component of the speech system associated with each characteristic. The component "prosodic" is listed when several components of the speech system may contribute to the dimension. Characteristics listed under "Other" include speech features not among the most deviant but that may occur and are not typical of most other dysarthria types.

Dimension	Speech Component
Imprecise consonants	Articulatory
Prolonged intervals*	Prosodic
Variable rate*	Prosodic
Monopitch	Phonatory-prosodic
Harsh voice quality	Phonatory
Inappropriate silences*	Prosodic
Distorted vowels	Articulatory-prosodic
Excess loudness variations*	Respiratory-phonatory-prosodic
Prolonged phonemes*	Prosodic
Monoloudness	Phonatory-prosodic
Short phrases	Prosodic
Irregular articulatory breakdowns	Articulatory
Excess and equal stress	Prosodic
Hypernasality	Resonatory
Reduced stress	Prosodic
Strained-strangled quality	Phonatory
Other	
Sudden forced inspiration or expiration*	Respiratory-prosodic
Voice stoppages*	Phonatory-prosodic
Transient breathiness*	Phonatory

*Tend to be distinctive or more severely impaired than in any other single dysarthria type.

with one another in DAB's subjects to form the cluster of *phonatory stenosis*, presumably resulting from relatively brief, random hyperadduction of the vocal folds (excess loudness variations could also result from choreic respiratory movements). Phonatory stenosis may be sufficient to cause *voice stoppages* in some patients. Acoustic analyses of patients with Huntington's disease have documented abnormal fundamental frequency (f_0) variability (often with abrupt changes), abnormally variable voice onset time (VOT), voice arrests, and reduced maximum vowel duration.^{56,86,104} These perceptual and acoustic findings mostly reflect instability of laryngeal movements during speech.

Although infrequent, some patients may exhibit *transient breathiness*. This may occur as a result of brief, involuntary vocal fold abduction, poor timing between expiration and phonation, or possibly in response to, or in compensation for, the physically exhausting effects of phonatory stenosis.

Resonance. Chorea may lead to *hypernasality* in some patients, although it is rarely pronounced. Hypernasality was correlated with imprecise consonants and short phrases in DAB's subjects, forming the cluster of *resonatory incompetence*. This suggests that air wastage through the velopharyngeal port may at least partially explain the occurrence of imprecise consonants and short phrases in some speakers.

Articulation. *Imprecise articulation* is the most prominent but not the most distinguishing feature of the dysarthria of chorea. It tends to occur simultaneously with *distorted vowels* and *hypernasality*; in DAB's patients these features formed the cluster of *articulatory-resonatory incompetence*. *Irregular articulatory breakdowns* also occur frequently. All of these features are presumably the result of various combinations of choreiform movements of the jaw, face, tongue, and palate. Acoustic analyses of people with Huntington's disease have supported this conclusion by documenting disproportionate lengthening of short vowels, slowed* and markedly variable speech AMRs or sentence duration, and abnormal first formant variability (suggestive of abnormal jaw movements) and second formant variability (suggestive of abnormal tongue position and shape) during steady state vowels.^{1,56,104} Many of these features have a significant impact on prosody.

Prosody. *Prosodic disturbances are prominent*. They can reflect the primary effects of chorea on speech, as well as the individual's response to the unpredictable movements. As DAB stated, "The flow of speech is often jerky, generated in fits and starts. As they proceed, patients are seemingly on guard against anticipated speech breakdowns, making compensation from time to time as they feel the imminence of glottic closure, respiratory arrest, or articulatory hindrance."³⁶

The most prominent cluster of speech characteristics in DAB's study was *prosodic excess*, composed of *prolonged intervals*, *inappropriate silences*, *prolonged phonemes*, and *excess and equal stress*. Patients also exhibited *prosodic insufficiency*, characterized by *monopitch*, *monoloudness*, *reduced stress*, and *short phrases*. Many patients also had

*Reduced syllable repetition rates have been detected before the emergence of other signs of disease in patients with Huntington's disease.³⁰

variable rate, possibly reflecting their efforts to complete phrases quickly before the next involuntary movement. The cooccurrence of the seemingly mutually exclusive clusters of prosodic excess and prosodic insufficiency attests to the moment-to-moment variability that occurs in this form of dysarthria, as well as the unpredictability of the effects of relatively quick and variable involuntary movements on speech. They probably also reflect the combined effects of the primary motor disturbance and compensatory or cautious responses to it.

What features of the dysarthria of chorea help identify and distinguish it from other MSDs? Most apparent is the *transient and unpredictable nature of the deviant speech characteristics*, the most obvious of which are *hypernasality, strained-harshness, transient breathiness, articulatory distortions and irregular articulatory breakdowns, loudness variations, and sudden forced inspiration or expiration*. These features, often in combination with the speaker's attempt to avoid or compensate for them, lead to *prolonged intervals and phonemes, variable rate, inappropriate silences, voice stoppages, and excessive or insufficient stress patterns*. The primary and distinguishing speech and speech-related findings in this form of hyperkinetic dysarthria are summarized in Table 8-5.

Dystonia*

Nonspeech Oral Mechanism

As in chorea, the oral mechanism is often normal in size, strength, and symmetry. Reflexes may be normal. Drooling may occur, and chewing and swallowing complaints are common. Patients frequently complain that food gets stuck in the throat or that chewing is difficult because of involuntary jaw or tongue movements.⁵³ The striking features of the nonspeech oral mechanism examination are most evident at rest or during attempts to maintain steady facial postures. Dystonic movements are slower than those of chorea, and they have a *waxing and waning* character. Blepharospasm and facial grimacing may be present, as may intermittent, relatively sustained spasms that lead to mouth opening and closing, lip pursing or retraction, and protrusion or rotary movements of the tongue (Figure 8-3). Affected neck muscles may cause elevation of the larynx; torsion of the neck may be marked in patients with torticollis. Recognition of dystonia is most difficult when

*This section addresses the general effects of dystonia on speech. Spasmodic dysphonia—because it is encountered fairly frequently in speech pathology practices, and because it often occurs as an isolated problem—is addressed separately in a subsequent section.

table
8-5

Primary distinguishing speech and speech-related findings in the hyperkinetic dysarthria of chorea

Perceptual Phonation-respiration	Sudden forced inspiration-expiration, voice stoppages, transient breathiness, strained-harsh voice quality, excess loudness variations
Resonance Articulation	Hypernasality (intermittent) Distortions & irregular breakdowns, slow & irregular AMRs
Prosody	Prolonged intervals & phonemes, variable rate, inappropriate silences, excessive-inefficient-variable patterns of stress
Physical	Quick, unpatterned involuntary head or neck, jaw, face, tongue, palate, pharyngeal, laryngeal, thoracic-abdominal movements at rest, during sustained postures & movement Dysphagia
Patient Complaints	Effortful speech, involuntary orofacial movements Chewing & swallowing problems

AMRs. Alternate motion rates.

movements are subtle or when cognitive or other motor deficits make valid observations difficult.

Patients may use sensory tricks to inhibit dystonic movements, and it is important to ask if they are aware of such tricks when they do not use them spontaneously. These often involve pressure or light touch to the jaw, cheek, or back of the neck; some patients will hold a pipe in the mouth, because it inhibits jaw, lip, or tongue dystonias.

In some cases the nonspeech oral mechanism examination can be entirely normal, with dystonic movements being triggered only by speech. There is a tendency for such patients to have focal dystonic movements that may involve only the jaw, tongue, pharynx, larynx, or respiratory muscles.

Speech

Conversational speech or reading, speech AMRs, and vowel prolongation are useful when assessing the dysarthria of dystonia. Careful visual observation of the patient during speaking is similarly important.

Table 8-3 summarizes the neuromuscular deficits presumed by DAB^{34,35} to underlie the dysarthria of



FIGURE 8-3 Woman with oromandibular-lingual dystonia attempting to sit in a relaxed posture with, **A**, relatively slow involuntary jaw opening and tongue lateralization and protrusion and, **B**, involuntary lip pursing.

dystonia. Nearly all aspects of movement may be disturbed. Dystonic movements may alter direction and rhythm of movement, and rate is generally slow. Range of individual and repetitive movements may be normal but can be reduced by excessive and biased muscle tone. The relationship of these characteristics to specific deviant speech characteristics is discussed as follows, as are findings from relevant acoustic and physiologic studies.

Clusters of Deviant Dimensions and Prominent Deviant Speech Characteristics

DAB³⁵ found several clusters of deviant speech dimensions in their patients with dystonia. Only those that are most prominent and distinctive are addressed here; the focus is on the most deviant or distinctive speech characteristics and their relationship to movement abnormalities at each level of the speech system.* The most deviant speech characteristics encountered in the dysarthria of dystonia are summarized in Table 8-6.

Respiration. The dystonic speakers in DAB's study did not exhibit speech characteristics that

*Golper et al.⁵¹ found a pattern of speech deficits similar to DAB's dystonic patients in a group of 10 patients with focal cranial dystonia (Meige's syndrome).

clearly reflected respiratory dystonia. However, some had *excess loudness variations*, and a small number had mild *alternating loudness*. These features could reflect abnormal respiratory movements or respiratory movements made in an effort to overcome phonatory stenosis.

Phonation. Several phonatory deviations may be present, including *harshness*, *strained-strangled voice quality*, *excess loudness variations*, and *voice stoppages*. Combined with *short phrases*, these characteristics combined in DAB's dystonic speakers to form the cluster of *phonatory stenosis*, a cluster also found in speakers with chorea. All of these characteristics can be related to dystonic hyperadduction of the vocal folds during phonation.

Some patients exhibit *audible inspiration*, probably secondary to involuntary vocal fold adduction during inhalation. With the exception of abductor vocal fold weakness, this feature is rarely encountered in other dysarthria types.

Although not prominent in frequency of occurrence or severity, *voice tremor* may be present. In fact, voice tremor was more evident in speakers with dystonic speech than in any other dysarthric group studied by DAB.

Resonance. Although dystonic movements can affect velopharyngeal function during speech, hyper-

The most deviant speech dimensions encountered in the hyperkinetic dysarthria of dystonia by Darley, Aronson, and Brown,³⁴ listed in order from most to least severe. Also listed is the component of the speech system associated with each characteristic. The component "prosodic" is listed when several components of the speech system may contribute to the dimension. Characteristics listed under "Other" include speech features not among the most deviant but that may occur and are not typical of most other dysarthria types.

<u>Dimension</u>	<u>Speech Component</u>
Imprecise consonants	Articulatory
Distorted vowels*	Articulatory-prosodic
Harsh voice quality*	Phonatory
Irregular articulatory breakdowns*	Articulatory
Strained-strangled quality*	Phonatory
Monopitch	Phonatory-prosodic
Monoloudness	Phonatory-prosodic
Inappropriate silences*	Prosodic
Short phrases	Prosodic
Prolonged intervals	Prosodic
Prolonged phonemes	Prosodic
Excess loudness variations*	Respiratory-phonatory-prosodic
Reduced stress	Prosodic
Voice stoppages*	Phonatory-prosodic
Slow rate	Articulatory-prosodic
Other	
Audible inspiration*	Phonatory-respiratory
Voice tremor*	Phonatory
Alternating loudness*	Respiratory-phonatory-prosodic

*Tend to be distinctive or more severely impaired than in any other single dysarthria type.

nasality is not a pervasive characteristic and is usually rated as mild when present.³⁴

Articulation. *Imprecise consonants, distorted vowels, and irregular articulatory breakdowns* may be prominent when dystonia affects articulators. These features formed the cluster *articulatory inaccuracy* in DAB's dystonic speakers. They logically reflect the effects on articulation of adventitious involuntary jaw, face, lip, or tongue movements (Figures 8-4 and 8-5). This conclusion receives support from acoustic analyses that have identified excessive second formant fluctuations during

steady state vowels in patients with tardive dyskinesia.⁵¹ This acoustic variability reflects vocal tract instability induced by orofacial dyskinesia (Figure 8-6).

Prosody. Prosodic disturbances are prominent and are similar to those encountered in chorea. *Monopitch, monoloudness, short phrases, and reduced stress* may be present, and in DAB's speakers they combined to form the cluster of *prosodic insufficiency*. Also commonly heard are *prolonged intervals, prolonged phonemes, and slow rate*, features that combined to form the cluster of *prosodic excess*. *Inappropriate silences* and *excess and equal stress* may also be detected, and they contribute to the general perception of exaggerated stress patterns. All of these features may reflect the effect of slowness of movement or interruptions in the flow of normal speech movements.

Similar to chorea, the cooccurrence of clusters of prosodic excess and prosodic insufficiency may reflect the variable nature of dystonia with its underlying cooccurring slowness and reduced range of movement, and the speaker's compensatory or cautious response to the primary disorder.

What features of the dysarthria of dystonia help identify and distinguish it from other motor speech disorders? Most apparent is the variable nature of the deviant speech characteristics, the most prevalent of which are *imprecision and irregular breakdowns of articulation, inappropriate variability of loudness and rate, strained harshness, transient breathiness, and audible inspiration*. These features, often in combination with the speaker's attempt to avoid or compensate for dystonic movements, may lead to *slow rate, prolonged intervals and phonemes, inappropriate silences*, and prosodic features that lead to both *excessive and insufficient stress patterns*. The primary and distinguishing speech and speech-related findings in this form of dysarthria are summarized in Table 8-7.

Athetosis

Although athetosis is a major subcategory of cerebral palsy, the term "athetosis" is rarely used to describe acquired movement disorders (possibly because many neurologists consider athetosis to be synonymous with dystonia). As a result, the literature on the dysarthria of athetosis is based exclusively on studies of children or adults with cerebral palsy. The results of such studies suggest that the speech characteristics of athetosis are probably captured within the descriptions of dysarthria associated with dystonia and, perhaps, chorea. Because of this apparent overlap, and because the focus of this book is on acquired MSDs, the literature on the speech of



FIGURE 8-4 Woman with predominantly lingual dyskinesia during normally rapid production of alternate motion rates for /pA/: involuntary tongue protrusion prevents bilabial closure.



FIGURE 8-5 Man with jaw opening dystonia during production of the /r/ in "grand." Excessive jaw opening and lingual retraction occurred only during speech and were associated almost exclusively with production of open vowels or velar consonants (see Case 8-2 for complete description).

individuals with athetotic cerebral palsy is not discussed here.*

*The interested reader is referred to reviews or comprehensive descriptive studies by DAB³⁶; Hardy⁵⁴; Kent and Netsell⁶²; Nielson and O'Dwyer^{81a}; Platt, Andrews, and Howie⁸²; Platt et al.⁸³; and Putnam.⁸⁴

Spasmodic Torticollis (Cervical Dystonia)

Spasmodic torticollis (ST) affects the cervical neck muscles and not the cranial nerve–innervated speech muscles. Unless accompanied by dystonia that directly affects the speech muscles, any ST-related speech abnormalities are presumably secondary to the effects of neck postural deviations

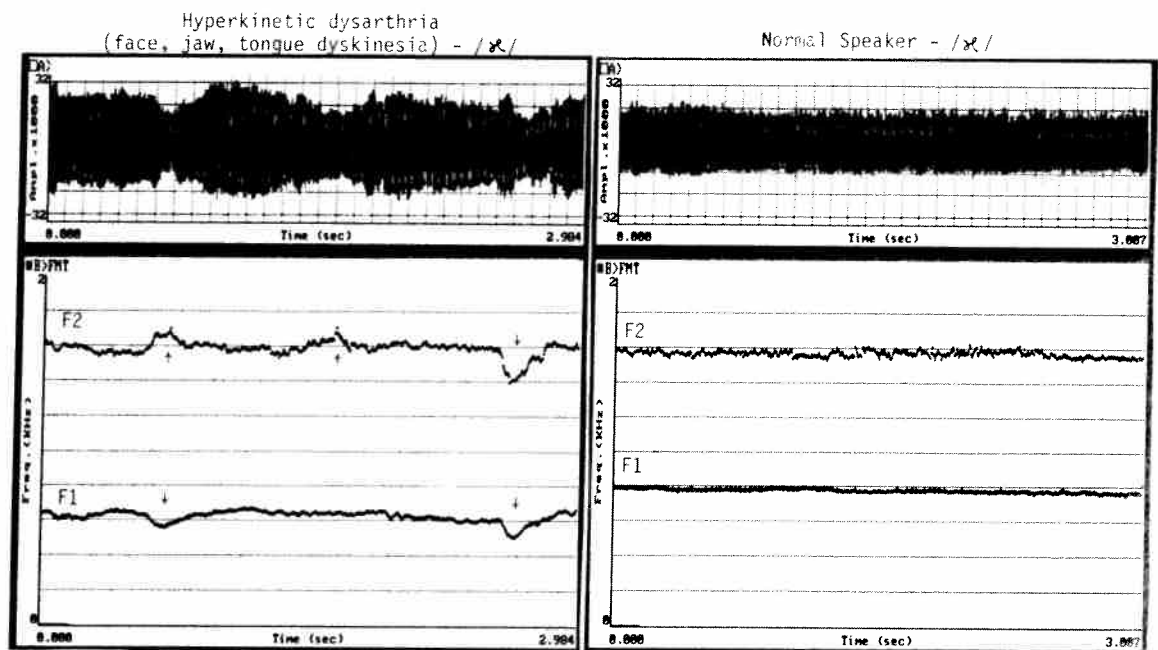


FIGURE 8-6 Raw acoustic waveform and first (*F1*) and second (*F2*) formant tracings for a 3-second prolongation of the vowel /æ/ by a man with face, jaw, and tongue dyskinesia (*left side*) and a normal male speaker (*right side*). Maintenance of steady vocal tract posture is reflected in all tracings for the normal speaker. In contrast, the hyperkinetic speaker's abnormal movements are reflected in the raw waveform and in the relatively sustained (≈ 230 to 280 ms) fluctuations in *F1* and *F2* (see *arrows*). The fluctuations are sometimes apparent in *F1* and *F2* simultaneously, but not always, and there is variability in their amplitude and direction. The fluctuations reflect auditorily perceptible abnormal movements of the tongue, lips, or jaw.

table 8-7 Primary distinguishing speech and speech-related findings in the hyperkinetic dysarthria of dystonia

Perceptual	
Phonation-respiration	Strained-harsh voice quality, voice stoppages, audible inspiration, excess loudness variations, alternating loudness, voice tremor
Resonance	Hypermasality
Articulation	Distorted vowels, irregular articulatory breakdowns, slow irregular AMRs
Prosody	Inappropriate silences, excess loudness variations, excessive-inefficient-variable patterns of stress
Physical	Relatively slow, waxing & waning head-neck, jaw, face, tongue, palate, pharyngeal, laryngeal, thoracic-abdominal movements Present at rest, during sustained postures & movement, but sometimes only during speech Improvement with "sensory tricks" Dysphagia
Patient Complaints	Effortful speech, involuntary orofacial movements "Tricks" that improve speech temporarily Chewing & swallowing problems (e.g., food "sticks" in throat)

AMRs, Alternate motion rates.

on primary speech muscle activity, or to alterations in the shape of the subglottic, glottic, or supraglottic vocal tract induced by abnormal neck postures (Figure 8-7). Given the severe distortions of neck posture that may occur in ST, it is surprising

that speech is not affected more frequently and dramatically.

The most detailed study of speech in people with ST is that by LaPointe, Case, and Duane.⁷⁰ In comparison to age-matched controls, their group



FIGURE 8-7 A, Front and B, back views of a woman with spasmodic torticollis attempting to sit in a normal resting posture. The neck rotation and head turning are sustained and involuntary.

of 70 people with ST exhibited reduced reading rate; reduced speech AMRs and sequential motion rates (SMRs); reduced maximum duration of /s/, /z/, and vowel prolongation; reduced phonation reaction time; and, in women, reduced habitual pitch, highest pitch, and pitch range. Intelligibility was also reduced, although the authors stated the overall impression "was that it was functional and intelligible, even if subtly different along some parameters." Increased vocal jitter and shimmer and decreased harmonic-to-noise ratio during vowel prolongation has also been reported.¹⁰³ In general, these studies suggest that the speech of some speakers with ST may be perceived as *slowly initiated, reduced in maximum duration of utterances, reduced in pitch and pitch variability, dysphonic, and reduced in rate.* These deficits, when present, are usually mild, and intelligibility is usually maintained.

The exact manner in which ST affects speech movements or vocal tract configurations is unclear. The speech characteristics of the disorder deserve further study, however, as much to establish how speakers adapt so well to abnormal head or neck postures as to understand the physiologic bases of the relatively mild speech abnormalities that may occur. The primary speech and speech-related findings associated with ST are summarized in Table 8-8.

Palatopharyngolaryngeal Myoclonus

PM is a rare disorder characterized by relatively abrupt rhythmic or semirhythmic unilateral or bilat-

table

8-8

Primary distinguishing speech and speech-related findings in the hyperkinetic dysarthria of spasmodic torticollis

Perceptual	
Phonation-respiration	Reduced pitch & pitch variability Dysphonia
Articulation-prosody	Reduced rate, delayed speech initiation, slow AMRs
Physical	Relatively sustained deviation of head to right or left, forward or back Sensory tricks reduce abnormal posturing
Patient Complaints	Speech often reported as normal Complaints related to neck movement & pain Occasional dysphagia Aware of sensory tricks that reduce spasm temporarily

AMRs, Alternate motion rates.

eral movements of the soft palate, pharyngeal walls, and laryngeal muscles.* The lesion causing it has

*Because PM is rhythmic and not lightninglike, neurologists have argued that the preferred term for this disorder should be "palatal tremor."¹⁸

been localized to an area of the brainstem and cerebellum known as the Guillain-Mollaret triangle, encompassing the loop among the dentate nucleus, red nucleus, and inferior olive (the dentatorubro-olivary tracts). PM is sometimes regarded as the prototypic movement disorder that depends on a central pacemaker that generates myoclonic jerks time-locked in different muscles. The inferior olive is thought to be the pacemaker, and hypertrophic degeneration of it is a common autopsy finding in people with the condition.³⁸ The MRI image in Figure 8-8 illustrates hypertrophy of the inferior olives that can be associated with PM.

PM is usually caused by a brainstem or cerebellar vascular event, but neoplasm, multiple sclerosis, encephalitis, and other degenerative diseases affecting the same general areas have been reported as causes.¹⁷ When caused by an acute lesion, there may be a delay of several months to years before the PM emerges.⁷⁸

PM can also be idiopathic. Deuschl et al.³⁸ found 27% of 287 cases with PM to have unknown etiologies. They referred to this condition as “essential rhythmic palatal myoclonus,” similar to other benign extrapyramidal conditions of undetermined origin, such as essential tremor (which includes essential



FIGURE 8-8 Transverse magnetic resonance image at the level of the medulla and cerebellum illustrating bilateral hypertrophy of the inferior olives (arrows) in a 45-year-old man with palatopharyngolaryngeal myoclonus (and ataxic dysarthria). See Case 8-7 for an illustrative case.

or organic voice tremor). Patients with this idiopathic condition had a much higher frequency of “earclicks” (explained later) and a slower rate of myoclonus (<120/min) than patients with symptomatic PM (i.e., PM with established etiology). They were generally younger than 40 years of age at onset and frequently also had myoclonus of the chin or perioral area.

Nonspeech Oral Mechanism

PM is present at rest, during sustained postures and movement, and during sleep. In some cases the eyeballs, diaphragm, tongue, lips, and jaw are also involved. The most common finding in PM is *abrupt, rhythmic, beatinglike elevation of the soft palate at a rate of 60 to 240 per minute*. Pharyngeal contractions also may be apparent and, because of activity of the tensor veli palatini, may produce opening and closing of the eustachian tube with an associated clicking sound that sometimes can be heard by others. These “earclicks” are a frequent complaint when PM is idiopathic but uncommon when PM is symptomatic.³⁸

Myoclonic movements of the larynx can sometimes be seen on the external surface of the neck, and patients may complain of a clicking sensation in the larynx or a sensation of laryngeal spasm. It is important to distinguish myoclonic movements in the external neck from carotid pulses, which are usually slower and do not visibly displace the laryngeal cartilages. Myoclonic movements of the lips and even the nares are sometimes present. Apparent lingual myoclonus may be seen, but lingual jerks may be secondary to laryngeal myoclonus.

Speech

The effects of PM on speech, even when it affects the jaw, lips, tongue, palate, pharynx, and larynx, are not apparent under most circumstances and may not be detectable at all during conversational speech, because they are so brief and relatively low in amplitude. If apparent during connected speech, it is perceived as a slow voice tremor and, less frequently, as intermittent hypernasality.³⁶ However, the effects of PM can usually be heard during vowel prolongation as *momentary rhythmic arrests or tremorlike variations* at a rate that matches the rate of palatal myoclonus. Myoclonic variations can be distinguished from those of essential voice tremor by their slower frequency and the relatively abrupt character of each cycle (voice tremor has a slower, more sinusoidal, waxing and waning character). Rarely, a clicking noise is audible at rest, reflecting eustachian tube opening. There may be occasional prolonged

silent intervals or inappropriate silences during speech if myoclonic vocal fold adduction occurs before inhalation at phrase boundaries is completed.⁹ If the diaphragm is involved, there may be momentary interruptions in phonation due to interrupted airflow.

The dysarthria of PM is probably rare as an isolated speech disturbance. Except when idiopathic, PM is usually accompanied by other signs of posterior fossa damage. As a result, PM probably most often occurs as one part of a speech disturbance that may include spastic, ataxic, flaccid, or unilateral upper motor neuron (UMN) dysarthria. Table 8-9 summarizes the primary speech and speech-related findings associated with PM.

Action Myoclonus

The effect of AM on speech has received little attention, but dysarthria can result from it.¹² The character of AM is quite different from that of PM, and its effects can have a greater functional impact on speech than PM.

Dysarthria is apparently common in patients with AM in nonspeech muscles. Fahn, Davis, and Rolland⁴² noted that at least 40% of their 59 cases with AM had dysarthria; it is unclear, however, whether the dysarthria was due to the AM or other cooccurring neuromuscular deficits.

AM is distinguished from other myoclonic conditions because it is induced by volitional muscle activity and is less generalized and rhythmic than other forms. In their classic description of four cases, Lance and Adams⁶⁹ noted that "the essential clinical picture was that of an arrhythmic fine or coarse jerking of a muscle or group of muscles in disorderly fashion, excited mainly by muscular activity when a conscious attempt at precision was required, worsened by emotional arousal, suppressed by barbiturates, and superimposed on a mild cerebellar ataxia." The jerks were typically less than 200 ms in duration and occurred singly or in series. Each patient had slow and "slightly slurred" speech.

AM is often associated with cerebellar, basal ganglia, or pyramidal system involvement. Lance and Adams⁶⁹ suggested that it might be a product of unrestrained synchronous or repetitive firing of thalamocortical neurons in the ventrolateral thalamus, the main relay nucleus from the cerebellum to the cortex. These abnormal discharges are then relayed from the cortex to the corticospinal and corticobulbar tracts, where they result in AM. Abnormal thalamocortical activity may be explained by inadequate control from the pontine reticular formation or by abnormal synchronous impulses through dento-thalamic pathways.

table 8-9 Primary distinguishing speech and speech-related findings in the hyperkinetic dysarthria of palatopharyngolaryngeal myoclonus

Perceptual	
Phonation-respiration	Often no apparent abnormality Momentary voice arrests during contextual speech when severe Voice arrests or myoclonic beats at 60-240 Hz during vowel prolongation
Resonance	Usually normal, but occasional intermittent hypernasality
Articulation-prosody	Usually normal, but brief silent intervals if myoclonus interrupts inhalation or initiation of exhalation, phonation, or articulation
Physical	Myoclonic movements of palate, pharynx & larynx, & sometimes lips, nares, tongue, & respiratory muscles Laryngeal or pharyngeal myoclonus sometimes observable beneath neck surface
Patient Complaints	Earclicks Patient often unaware of myoclonic movements & usually does not complain of speech difficulty

The most common etiology of AM is anoxic encephalopathy (e.g., due to cardiorespiratory arrest), in which the principal findings have been degeneration of cells and fibers in the globus pallidus, hippocampus, deep folia in the cerebellum, and the deep layers of the cerebral cortex, especially the parietal and occipital lobes.⁶⁹ Multiple other causes also have been documented, including myoclonic epilepsy, toxic-metabolic disturbances (e.g., lithium exposure, immunosuppressive drugs in transplant patients), infectious processes (e.g., encephalitis), paraneoplastic cerebellar degeneration, stroke, and multiple sclerosis.^{29,68,100}

Nonspeech Oral Mechanism and Speech

By definition, AM is not present at rest, and nonspeech oral mechanism examination may be entirely

normal unless deficits in addition to the AM are present.

Aronson, O'Neill, and Kelly¹² have presented the only specific description of the dysarthria of AM. The disorder appears to have its primary perceptual effects on articulation (labial) and phonation. Their four cases were described as having stable orofacial muscles at rest but quick, gross, or fine jerky movements during attempts to speak. "*Repetitive fluctuation of phonation*" and *adductor voice arrests*, which were synchronous with myoclonic spasms of the lips, were characteristic. Slow speech rate was apparent in each case, and the myoclonic movements worsened with increased speech rate. Slow rate could be compensatory, although the authors noted that, consistent with the general physiology of myoclonus, it could also reflect brief periods of inability to contract muscles following myoclonic jerks. These observations suggest that patients suspected of having the disorder should be asked to speak at slow, average, and rapid rates. Noticeable deterioration of voice quality or articulatory adequacy with increased rate or the emergence of myoclonic facial movements with increased rate can help confirm the diagnosis, because other dysarthrias are generally not triggered by increases in rate; intelligibility may improve at slowed rates, but the underlying disordered movements are generally not altered. Speech AMRs may be particularly useful for making such observations. Table 8-10 summarizes the primary speech and speech-related findings associated with the dysarthria of AM.

Tics—Tourette's Syndrome

Tics may occur as an isolated, nonspecific disorder, but TS is the prototypic tic disorder. It is defined operationally in the *Diagnostic and Statistical Manual of Mental Disorders*³⁹ as including: (1) multiple motor and one or more vocal tics, (2) tic-free periods not exceeding 3 months, (3) marked distress or impairment in daily functioning, (4) onset before 18 years of age, and (5) not due to physiologic effects of a substance (e.g., stimulants) or other medical condition (e.g., Huntington's disease, postviral encephalitis). It is generally felt that most cases are genetically determined and suspected that pathophysiology involves striatal dopamine receptor supersensitivity. Although traditionally viewed as severe and disabling, TS is now recognized as a clinically heterogeneous disorder with motor and behavioral features that vary along a severity continuum.⁶⁷

TS, which affects mostly males ($\approx 3:1$ ratio), frequently cooccurs with obsessive-compulsive disorder or attention deficit hyperactivity disorder.⁶⁷ Stuttering, dyslexia, conduct disorder, panic attacks,

table 8-10 Primary distinguishing speech and speech-related findings in the hyperkinetic dysarthria of action myoclonus

Perceptual	
Phonation-respiration	Occasional adductor voice arrests
Articulation-prosody	Slow rate, decreased precision with increased rate Marked deterioration of AMR regularity with increased rate
Physical	Normal at rest unless other neuromuscular deficits present Quick, gross, or fine jerky movements of orofacial muscles during speech—especially lips—worsening with increased rate
Patient Complaints	Awareness of imprecise speech & inability or reluctance to speak at normal or rapid rates

AMR, Alternate motion rate.

multiple phobias, depression, and mania reportedly are more common than in control subjects.³¹

Tics are brief involuntary movements or sounds that occur over normal background motor activity. They can be brief and isolated (e.g., eye blink, head twitch, facial grimace) or can consist of coordinated and seemingly purposeful movements (e.g., touching, jumping, obscene gestures). Some patients experience "sensory tics," which are somatic sensations, such as pressure, tickling, and temperature changes, that may lead to movements intended to relieve the sensation, such as tightening or stretching of muscles.⁶⁷ Tics are often bizarre appearing and frequently misinterpreted as signs of psychiatric disease.

In addition to motor tics and behavioral disorders, TS is characterized by *vocal tics* that can be isolated or embedded within voluntary verbal utterances. Vocal tics are unique, because they represent *the only dysarthria in which specific sounds or spoken words represent the disorder*.

Simple vocal tics include noises and sounds that are made repetitively and sometimes can be suppressed temporarily. The most common of these are throat clearing and *grunting*, but *yelling-screaming*, *sniffing*, *barking*, *snorting*, *coughing*, *spitting*, *squeaking*, and *humming* can also occur.³² These sounds are usually executed rapidly, and some of them may reflect a response to a sensation in the

larynx or throat.⁶⁷ More complex vocal tics may include *echolalia* (repetition of other's utterances), *palilalia* (discussed in Chapter 7), and *coprolalia*.

Coprolalia (copro = feces; lalia = lips), or involuntary, compulsive, repetitive, almost ritualistic swearing, is one of the most dramatic, although not universally present, features of TS (the words "fuck," "shit," and "piss" are the most common scatological utterances, according to Comings³²). The words are often said softly or incompletely and are sometimes accompanied by throat clearing or other noises, possibly reflecting an attempt to suppress or mask the coprolalia. They may emerge independent of any volitional verbal expression or at the start of or within volitional utterances. They sometimes may appear socially acceptable or even humorous, but the social and psychologic consequences for the patient are often tragic.

The primary speech and speech-related characteristics of TS are summarized in Table 8-11.

Organic (Essential) Voice Tremor

Organic or essential (idiopathic) voice tremor is often simply viewed as a voice disorder and not a neurologic disorder or a dysarthria. However, it occurs in approximately 20% of patients with essential tremor elsewhere,⁶⁰ and clearly can be classified as a hyperkinetic dysarthria of tremor.*

Essential tremor is the most common movement disorder, and a family history of tremor is present in 17% to 96% of affected people.⁵⁹ It can begin at any age, often before 50 years, and incidence increases with age. It occurs most frequently in the hands but, on average, is present in the voice in approximately 14% of affected people.⁵⁹ Although generally benign, it usually slowly progresses in severity (tremor amplitude). A focal presentation of the disorder (e.g., voice tremor) sometimes spreads to include other body parts, and it is sometimes a precursor to, or associated with, other movement disorders such as focal dystonias, dystonia musculorum deformans, and ST.^{45,66}

The localization of essential tremor is unknown, but it is probably related to a CNS oscillatory abnormality. The red nucleus, cerebellum, and inferior olivary and ventrolateral thalamic nuclei are possible sites on the basis of their inherent rhythmic physiology, the results of PET studies in people with the

*Essential tremor can affect the tongue, sometimes in isolation, but essential lingual tremor is rare in comparison to essential voice tremor. Patients with lingual tremor are usually unaware of it. It occurs at a rate of 4 to 8 Hz, is generally apparent on protrusion but not at rest, and is often alcohol responsive.¹⁶ Its effects on speech are unclear.

table

8-11

Primary distinguishing speech and speech-related findings in the hyperkinetic dysarthria of Tourette's syndrome

Perceptual

Phonation-respiration Coughing, grunting, throat clearing, screaming, moaning, etc.

Resonance

Articulation-prosody Sniffing
Humming, whistling, lip smacking, echolalia, palilalia, coprolalia

Physical

Multiple motor tics (e.g., eyeblinks, head twitch, facial grimacing, jumping, touching, obscene gestures)

Patient Complaints

Awareness of vocal and motor tics, compulsion to perform them, & inability to inhibit them for sustained periods

Behavioral & psychiatric disorders may be present (e.g., obsessive-compulsive, phobias, hyperactivity & attention deficit disorder, learning disability)

disorder, or known sites of surgical or vascular lesions that may abolish or cause it.⁵⁹

The onset of essential voice tremor is usually gradual. When mild, patients may not be aware of its presence. Those who are aware often note that it worsens with fatigue and psychologic stress and improves with alcohol intake. The voice tremor can be an isolated problem, but more often is accompanied by head or extremity tremor. When isolated, it is sometimes misdiagnosed as a psychogenic disorder.

Nonspeech Oral Mechanism

Lingual tremor may be apparent at rest or on protrusion in patients with organic voice tremor. When present during phonation, it may represent genuine

*Patients with cerebellar disease sometimes have voice tremor. The tremor frequency is in the range of 3 Hz, similar in frequency to other forms of cerebellar postural tremor.² This is slower than that of essential voice tremor and usually occurs with an ataxic dysarthria. Although its frequency is in the general range of palatal-laryngeal myoclonus, it can occur without evidence of myoclonic movements at rest.

lingual tremor or be secondary to vertical oscillations of the larynx. Tremulous movements of the jaw and lips are often apparent at rest, during sustained postures, and during vowel prolongation. Palatal and pharyngeal tremor are often obvious during sustained "ah," synchronous with the perceived voice tremor; this can be evident even when resonance is perceived as normal. Fiberscopic observation of the larynx may reveal rhythmic vertical laryngeal movements and adductor and abductor oscillation of the vocal folds, synchronous with the perceived voice tremor. Vertical oscillations of the larynx also can often be seen on the external neck during vowel prolongation. Tomoda et al.⁹⁶ recorded electromyographic evidence of tremor in the cricothyroid muscle and expiratory muscles (rectus abdominis) in three patients with organic voice tremor, synchronous with voice tremor. They suggested that voice tremor may be an action tremor of voluntary expiratory muscles that affects phonatory function. Although respiratory tremor should be considered a possible source of voice tremor, it is probably not a primary factor in most cases of organic voice tremor.

Speech

Aronson⁹ describes three effects of essential tremor on voice: (1) a "typical" organic voice tremor when the adductor and abductor vocal fold tremor components are relatively equal, (2) an adductor spasmodic dysphonia when the adductor component is predominant, and (3) an abductor spasmodic dysphonia when the abductor component is predominant. Only the typical voice tremor is addressed here (spasmodic dysphonias associated with tremor are discussed in the next section).

Voice tremor may not be apparent during contextual speech, especially when mild, which may be why some patients are unaware of it. Its rhythmic fluctuations are most easily perceived during vowel prolongation. To rule out a respiratory contribution to the voice tremor, it is often useful to have the patient prolong /s/ and /z/. If the /s/ is steady and the /z/ or vowel contains tremor, a prominent respiratory contribution to the voice tremor is unlikely.

Organic voice tremor most often occurs at a frequency of 4- to 7-Hz, most often in the 5- to 6-Hz range, with a tendency for tremor frequency to be slower with increasing age.^{10,18} The tremor has a *sinusoidal, quavering, or rhythmic waxing and waning character* during vowel prolongation, presumably due to rhythmic alterations in pitch, loudness, or both (Figure 8-9). When severe, there may be abrupt, staccato voice arrests that, in most cases, are rhythmic. However, the tremor may lose its rhythmic character when arrests are present, pos-

sibly because of the speaker's efforts to avoid or otherwise compensate for them. In such cases, having the patient prolong a vowel at a higher pitch may abort the arrests and allow the tremor to be heard more easily. Additional acoustic attributes associated with essential voice tremor include increased jitter, reduced harmonic or noise ratio, and reduced dynamic range at the natural frequency of phonation.⁵⁰

Patients with marked to severe organic voice tremor may have *reduced speech rate* secondary to phonatory interruptions; speech rate may also be reduced secondary to jaw, lip, and tongue tremor. When voice and oromandibular tremor occur simultaneously and are marked, the effects on speech can be pronounced, and the disorder can become more complex than the smooth modulations of a sinusoidal tremor. Kent et al.⁶³ reported such a case with severely reduced intelligibility. Acoustic analyses documented variable patterns of phonation, with dysphonic intervals, harmonic doubling, and noise. Single word rates and AMRs were slow and variable, and the jaw tremor interfered with stability of articulation. Of interest, articulatory movements sometimes seemed timed to the 3- to 5-Hz tremor cycle; Kent et al.⁶⁴ note that one way an individual with tremor can contend with it "is to coordinate voluntary movements with the tremor, which then acts as an internal pacemaker."

The primary speech and speech-related characteristics associated with organic voice tremor are summarized in Table 8-12.

Spasmodic Dysphonia

Spasmodic dysphonia (SD) designates a group of voice disorders that are characterized by strained or breathy voice qualities resulting from adductor or abductor laryngospasm. Concepts of the disorder have an interesting history. For a number of years SD was thought to be a manifestation of psychopathology, usually stemming from psychologic trauma, stress, or anxiety. Neurologic etiologies were rarely considered. Over the past 2 decades the etiologic pendulum has swung to a point where many investigators and clinicians assume that the disorder is always neurogenic, with only lesion loci and the specific neurophysiologic nature of the disorder in question. Along with this trend has come a shift from use of the term "spastic" to the term "spasmodic" to characterize the dysphonia. This latter trend is useful, because, although the term "spastic" describes the strained character of the adductor form of the disorder, it does not appear that spasticity (in the physiologic sense) is responsible for most cases of SD. The term "spasmodic" retains descriptive power and at the same time suggests that spasm (or

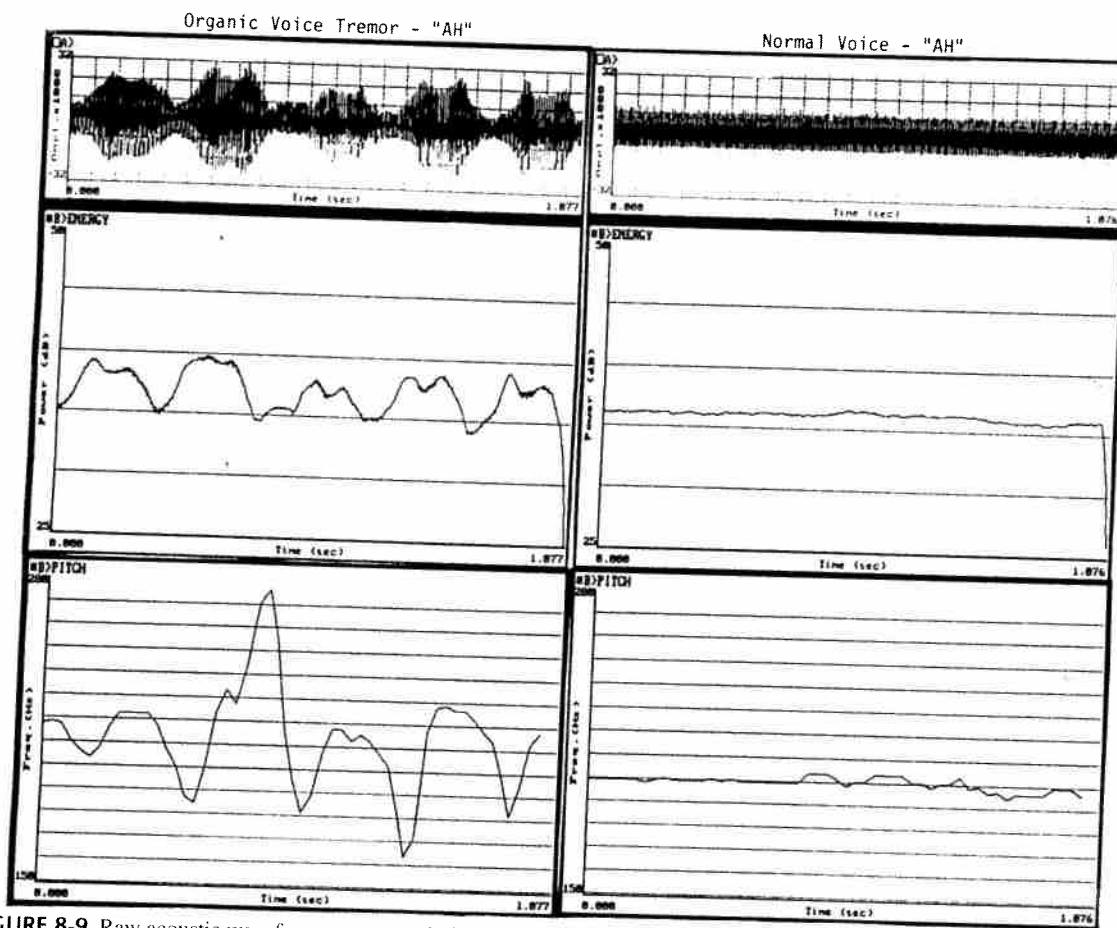


FIGURE 8-9 Raw acoustic waveform, energy, and pitch contours for an approximately 1-second prolongation of /a/ by a female speaker (see Case 8-4 for full clinical description) with an approximately 5-Hz organic voice tremor (left side) and a normal female speaker (right side). The fairly regular tremor is apparent in all tracings and stands in marked contrast to the steady maintenance of the same parameters by the normal speaker.

dystonia) is the basis for at least some or many forms of the disorder.

Aronson⁹ has argued that any diagnosis of SD should be modified to specify likely etiology. There are three broad etiologic possibilities: *neurogenic*, *psychogenic*, and *idiopathic*. This recognizes that SD can have at least two etiologies—neurogenic and psychogenic—and that etiology sometimes cannot be specified with any degree of confidence (hence is idiopathic). Importantly, psychogenic and neurogenic SD may not be distinguished on the basis of auditory perceptual characteristics; the distinction is often made on the basis of history and other examination findings. Evidence that SD can have a psychogenic etiology is discussed in Chapter 14. Here are addressed only neurogenic varieties of SD, which justifiably can be considered a subtype of hyperkinetic dysarthria.

It is also important to establish if SD is *adductor*, *abductor*, or *mixed* in form. Adductor SD, by far the most common variety, is characterized by adductor laryngospasms that give the voice a *strained, tight* character. Abductor SD is characterized by abductor laryngospasms that give it an *intermittent breathy* or *aphonic* character. Some patients have a mixed form, with both intermittent strained and breathy qualities. There is no evidence that these symptomatic types vary in distribution as a function of neurogenic versus psychogenic etiology.

Ignoring possible etiologic differences, SD has an average age of onset of approximately 45 to 50 years, but it can develop anywhere from the third to eighth decades. Male-to-female ratio ranges from 1:1 to 1:4. It may develop suddenly but usually begins insidiously, often taking a year or longer to develop into its full-blown state. Remissions are rare when

table 8-12 Primary distinguishing speech and speech-related findings in the hyperkinetic dysarthria of organic voice tremor

Perceptual	
Phonation-respiration	Quavering, rhythmic, waxing & waning tremor, most evident on vowel prolongation, at a rate of =4 to 7 Hz Voice arrests may occur in severe forms but may disappear if pitch is raised
Articulation	Usually normal, but rate may be slowed
Prosody	Normal pitch & loudness variability may be restricted or altered by tremor
Physical	Rhythmic, vertical laryngeal movements & adductor & abductor oscillations of the vocal folds synchronous with voice tremor. Tremor of jaw, lips, tongue, & palate or pharynx may be present, especially during phonation. Lingual & jaw tremor may be secondary to laryngeal tremor.
Patient Complaints	Shaky or jerky voice Worse with fatigue or anxiety Improves with alcohol Frequent family history of tremor

the cause is neurologic. It is not unusual for the disorder to begin during a flulike illness or during a period of acute or chronic psychological stress; this may be the case even when SD is clearly neurogenic in origin.

In an extensive review of evidence for the neurologic underpinnings of SD, Cannito²¹ concluded that SD probably results from an impairment of the volitional motor system, rather than the limbic or lower brainstem centers for vocal control. He based this conclusion on evidence from several studies of (1) a relatively high incidence of extrapyramidal and pyramidal motor signs in patients with SD, (2) neuroimaging evidence for such lesions,^{*46} (3) the tendency for the disorder's manifestations to be greater for complex than simple verbal activities, and (4) a strong association between SD voice and speech characteristics and those found in spastic

dysarthria and the dysarthrias of dystonia and chorea. He tied these data to models of hyperkinesias that implicate cortical premotor-striatal-pallidal-thalamic control circuits that are normally involved in the control of complex, voluntary sequential movements.

Numerous factors influence SD, and wide intraindividual fluctuations are common. Emotional stress, anxiety, depression, and physical exertion often make symptoms worse. These influences are superficially suggestive of psychogenic etiology, but it is important to note that many of them also affect severity in people with unambiguous neurogenic movement disorders, and that they are common complaints in many patients with other dysarthria types. In contrast to most other dysarthria types, however, voice in SD may be normal during singing or laughter, and under conditions of surprise or quickly emitted "automatic" utterances. Most often, the movement disorder underlying neurogenic SD is *action induced*, the triggering action being volitional speech.

Tremor and dystonia have a strong causal association with neurogenic SD. This suggests that SD is related to extrapyramidal system dysfunction in the broad sense, more specifically to dysfunction in the basal ganglia or cerebellar control circuit. SD may be an isolated, focal manifestation of tremor or dystonia, or may coexist with more widespread manifestations of them.

That dystonia or tremor can be focal to the larynx receives support from other focal manifestations of tremor and dystonia (e.g., blepharospasm, oromandibular dystonia or tremor, torticollis) and the fact that the basal ganglia have a somatotopic organization. Jankovic⁶⁰ notes that the somatotopic organization of the basal ganglia plays an important role in the muscular distribution of hyperkinesias and the preferential involvement of head and orofacial structures in them.

SD of essential voice tremor presumably occurs (1) when the adductor component of the tremor predominates and causes adductor laryngospasms and squeezing of the glottis (adductor SD of essential voice tremor), (2) when the abductor component of the tremor predominates and causes abductor laryngospasm and widening of the glottis (abductor SD of

*The interested reader is referred to the work of Finitzo and Freeman⁴⁶ and an exchange of letters between Aronson and Lagerlund¹¹ and Finitzo et al.⁴⁷ for a comprehensive and critical overview of approaches to establishing the neurologic locus of SD and the methodological and interpretive challenges and controversies faced by such efforts.

essential voice tremor), or (3) when the tremor amplitude is relatively balanced in adductor and abductor muscles but sufficient to cause both abductor and adductor laryngospasm (mixed adductor and abductor SD of essential voice tremor). Thus there appears to be a continuum along which a diagnosis of essential voice tremor can merge into a diagnosis of SD. The continuum seems to include severity as well as the balance of muscle forces involved in adductor and abductor laryngeal activity. In addition to perceptual evidence, a link between SD and essential tremor comes from evidence of nonlaryngeal tremor in patients with SD. For example, approximately one third of patients with SD have evidence of essential tremor elsewhere⁶⁰; Aronson and Hartman¹¹ found a similar tremor frequency between patients with essential voice tremor and patients with SD and voice tremor, and their SD patients had a high incidence of tremor elsewhere in the body.

Nonspeech Oral Mechanism

Affected patients often have a normal oral mechanism examination. If associated with generalized or other orofacial tremor or dystonia, Meige's syndrome, or ST, manifestations of those problems will be apparent. Soft neurologic signs such as facial or palatal asymmetry, mild weakness, or pathologic oral reflexes can be present, but not usually, and probably not as a reflection of any causal relationship. *Because SD is an action-induced disorder, evidence for it might be found only during voluntary speech.*

Speech (Adductor Spasmodic Dysphonia)

The primary perceptual feature of adductor SD is a *strained, jerky, grunting, squeezed, groaning, and effortful* voice quality. When mild, there may be no more than a mild strained quality. These qualities can be intermittent or relatively continuous. *Silent articulatory movements or sound repetitions*, presumably as a result of unanticipated laryngospasms, may be present. Additional dysfluent characteristics, such as tense pauses, dysrhythmic phonation, part and whole word repetitions, and revisions have been documented and contribute to clinical impressions of overall severity.²⁴

When tremor underlies adductor SD, there may be a *staccato quality* or an *obvious tremor or rhythmic character* to the laryngospasms. This is heard most easily during vowel prolongation; when voice

arrests are prominent, having the patient phonate at a higher pitch may attenuate the arrests and permit perception of the tremor. There may be associated head, jaw, lip, tongue, palatal, pharynx, and thoracic tremor during vowel prolongation.

When dystonia underlies the laryngospasm, the spasmodic voice may be continuous, or arrests may be unpredictable. Speech rate may be slow secondary to laryngospasm, or perhaps, in some cases, because of involvement of supralaryngeal muscles.^{20,22} Intermittent or fairly constant *hypernasality* is perceptually evident in some patients. Jerky and dysrhythmic movements of the thorax and abdomen may be apparent during speech, synchronous with strained voice and voice arrests; these are probably a secondary effect of uncontrolled glottic closure.

When adductor SD is severe, there may be *facial grimacing, associated neck contractions, and movements of the shoulder girdle and upper arms*. The overall picture may be one of *extreme physical effort* during speech. This may be one of the primary complaints of people with the disorder, sometimes exceeding their dissatisfaction with the voice itself.

Patients with adductor SD can have abnormally increased subglottal air pressure, abnormal variability of phonatory airflow, and increased laryngeal resistance during speech.^{5,48,57} Electromyogram (EMG) examinations have revealed delays in speech initiation and overactivity of laryngeal muscles.⁷² Videofiberoptic laryngoscopy during speech may reveal rhythmic, arrhythmic, or relatively sustained adductor spasms of the true vocal folds and arytenoids, but as severity increases, spasm of the false folds and even the inferior pharyngeal constrictor muscles may be observed. In some cases the entire larynx may move upward, implicating spasmodic activity in the extrinsic laryngeal muscles as well. High-amplitude muscle burst activity in the 6- to 7-Hz range, synchronous with fluctuations in the speech waveform, have been observed in the thyroarytenoid and levator palatini muscles.^{46,97} Lundy et al.,⁷⁴ using fiberoptic videonasolaryngoscopy, found abnormal soft palate posturing during speech in 84% of 83 patients with laryngeal movement disorders who did not have perceptually abnormal oral or nasal resonance; speech diagnoses included adductor, abductor, and mixed SD and voice tremor. The findings suggest that the abnormal soft palate posturing reflects compensatory behavior or an additional area of primary involvement. Similar explanations may apply to a finding of abnormal kinematic patterns of lip movements during speech in two speakers with adductor SD (and another with abductor SD).⁹⁵

Table 8-13 Primary distinguishing speech and speech-related findings in the hyperkinetic dysarthria of spasmodic dysphonia

Perceptual	
Phonation-respiration	<p><i>Adductor:</i> Continuous or intermittent strained, jerky, squeezed, effortful quality, with voice arrests when severe. If tremor based, voice tremor may be apparent, especially during vowel prolongation at higher pitches.</p> <p><i>Abductor:</i> Brief, breathy or aphonic segments, most obvious at beginning of utterances or in voiceless consonant environments</p>
Resonance	<p><i>Adductor:</i> Usually normal but occasionally hypernasal</p> <p><i>Abductor:</i> Usually normal, but occasionally hypernasal</p>
Articulation-prosody	<p><i>Adductor:</i> Inappropriate silences, silent articulatory movements, & sound repetitions, especially when voice arrests are prominent</p> <p>Contextual speech and AMRs may be slow secondary to laryngospasms</p> <p><i>Abductor:</i> Short due to air wastage through glottis during abductor spasms</p>
Physical	<p>Nonlaryngeal muscles are usually normal, unless tremor or dystonia present elsewhere (e.g., head or limb tremor, orofacial dyskinesia, torticollis)</p> <p>Rhythmic or arrhythmic spasms of true folds & arytenoids, & sometimes false folds & pharyngeal constrictors, usually only during speech</p> <p>Jerky & arrhythmic thoracic or abdominal movements, usually secondary to adductor laryngospasm</p> <p>Facial grimacing & neck or shoulder movements secondary to severe adductor laryngospasms</p>
Patient Complaints	<p><i>Adductor:</i> Tight, strained voice</p> <p><i>Abductor:</i> Intermittent, weak, breathy, aphonic voice</p> <p><i>Adductor & abductor:</i> Increased physical effort & fatigue associated with speaking</p> <p>Occupational, social, & emotional impact may be significant. Voice may improve with alcohol when tremor based.</p>

Acoustic analyses have documented phonatory breaks, aperiodicity, breakdown in formant structure, elevated standard deviation of f_0 , abnormal frequency shifts and intensity fluctuations, widely spaced vertical striations at irregular intervals (reflecting reduced f_0 and aperiodicity of phonation), increased jitter and shimmer, interruptions in articulation, separation of sounds in syllables, delayed onset of phonation in vowels, and a tendency toward reduced f_0 and loudness (e.g., Adams et al.⁴; Cimino-Knight and Sapienza, 2001^{29a}; Hertegard, Granqvist, and Lindestad³⁵; Ludlow and Connor⁷³; Sapienza, Walton, and Murry⁹¹; Sapienza et al.⁹²; Wolfe and Bacon¹⁰²; Zwirner, Murry, and Woodson¹⁰⁵).^{*} A number of these acoustic measures have served as indices of change in response to treatment with botulinum toxin. These acoustic and physiologic attributes are generally consistent with and further refine perceptual descriptions of the disorder.

^{*}Despite evidence of phonatory instability or unsteadiness in the disorder, the number of phonatory breaks, frequency shifts, and aperiodic segments seems consistent across repeated trials and over time (Cimino-Knight and Sapienza, 2001).^{29a} However, the relative predominance of acoustic abnormalities may vary as a function of speech task (e.g., reading versus vowel prolongation).⁹¹

Speech (Abductor Spasmodic Dysphonia)

In abductor SD, the voice is interrupted by brief, inappropriate breathy or aphonic segments that are most easily triggered by voiceless consonants in the beginning of an utterance or syllable. During these segments airflow increases, speech rate may be slowed.⁷⁹ As in adductor SD, dysfluencies and hypernasality are sometimes apparent.

Acoustic analyses have revealed increased aspiration time for initial stops, a loss of energy in the higher formants, intensity fluctuations, prolonged VOT for voiceless consonants, elevated average f_0 , and increased sentence articulation times.^{23,102} Direct observation of the larynx may show abduction of the vocal folds during phonation, resulting in a wide glottal chink; these coincide with breathy releases. If tremor is present, abductor movements may be rhythmic. EMG has identified increased activity in the thyroarytenoid and posterior cricoarytenoid muscles in speakers with this form of SD.⁹⁷ There can be a breakdown of formant structure or superimposed noise on vertical striations in spectrograms of affected speakers.¹⁰²

The primary speech and speech-related findings associated with SD are summarized in Table 8-13.

Cases

Case 8-1

A 73-year-old man presented with a 5-month history of "hesitation" in speech, which had initially worsened for a few months and then plateaued. Neurologic examination was normal with the exception of abnormal orofacial movements. His CT scan and electroencephalogram were normal. Routine laboratory studies and screening for heavy metal poisoning were within normal limits. He had never taken neuroleptic medications.

During speech examination, he complained of halting and slurred speech as well as involuntary mouth movements. Examination revealed tremor of the lips at rest and on lip rounding and semirhythmic movement of the tongue at rest. A regular voice tremor was present during vowel prolongation. Relatively rapid chewing and smacking and rounding movements of the lips interrupted contextual speech. They were noticeably reduced if the patient spoke while biting on a tongue depressor or with some pressure at the angle of the mouth.

The clinician concluded that the patient had a "hyperkinetic dysarthria associated with oro-

facial dyskinesia with an accompanying tremor component."

The patient was advised to speak while holding a pipestem in his mouth and biting down, and he was given some practice at doing it. The neurologist recommended a trial of Inderal for the movement disorder. Several weeks later, the patient wrote to indicate that the Inderal had significantly reduced, but not eliminated, his facial grimacing. He also noted that "a pipe held between my teeth is definitely effective and socially acceptable."

Commentary. (1) Orofacial dyskinesias or focal mouth dystonias often develop without a clear etiologic explanation. They can be present in the absence of any other neurologic symptoms and in the absence of abnormalities on neuroimaging studies. (2) Orofacial dyskinesias can affect speech. (3) Sensory tricks, such as biting down or exerting some pressure on the cheek, can be effective (temporarily) in relieving abnormal movements and improving speech.

Case 8-2

A 53-year-old man presented with an 18-month history of speech difficulty and a right upper and lower limb movement disorder. The course was one of gradual onset and progression. A question had been raised about manganese intoxication, possibly secondary to exposure when welding or from materials used in refinishing a boat.

Neurologic evaluation revealed the presence of torsion dystonia of the right foot during walking and right upper extremity cogwheel rigidity. A jaw-opening dystonia during speech was also apparent. He was referred for speech evaluation.

The patient was aware of abnormal jaw movements during speech but was unaware of difficulty at other times, including during chewing and swallowing. His speech tended to worsen when he was anxious, excited, or consuming alcohol. Speech was better in the morning, following relaxation exercises, and when writing or drawing while speaking; he felt this latter activity distracted his attention from speech.

Oral mechanism examination at rest and during non-speech sustained postures was normal. During speech he had intermittent marked jaw opening and tongue retraction. Superficially, these movements were random, but

careful analysis established that they were strongly associated with the occurrence of open vowels and velar consonants (i.e., sounds requiring jaw opening or back-of-tongue elevation). Speech improved somewhat during whispering and noticeably when he clenched his jaw during speaking. It improved moderately when he wrote while talking. His jaw opening was often sufficient to arrest speech, with continuation possible only after the dystonic interval passed.

MRI and SPECT scans were normal. Additional laboratory and radiologic studies were negative. An EMG revealed normal blink and facial nerve conduction and a normal masseter-inhibiting reflex. There was no evidence of abnormal activity in the lateral pterygoid muscles and digastric muscles at rest or during chewing and drinking, but tonic spasms of 500 to 3000 ms were present in those muscles during speech.

It was concluded that the patient had a progressive extrapyramidal disorder of unclear etiology. It was recommended that he avoid welding, painting, and other heavy metal exposure. Sinemet was prescribed. There was some improvement in the patient's limb symptoms but no change in speech. Subsequent examination of the paint and several metals to which he had been exposed

Case 8-2—cont'd

failed to provide convincing evidence that his disorder was due to heavy metal intoxication.

Commentary. (1) Dystonia affecting speech can be specific to small groups of muscles (jaw and possibly tongue in this case) and, in fact, may be present only during speech. In some cases focal, speech-induced dystonias can be relatively pho-neme specific; in this case they were triggered by open vowels and back of tongue

elevation. (2) Focal speech-induced dystonias sometimes improve with altered postures and distraction and are generally better under conditions of relaxation. The worsening of dystonia or speech difficulty under conditions of anxiety does not establish anxiety as a cause of the speech problem. (3) Perhaps more frequently than any other dysarthria type, the cause of hyperkinetic dysarthria may be indeterminate.

Case 8-3

A 49-year-old woman presented with a 1-year history of ataxia and movement difficulties, including speech. Her problems had begun suddenly with a severe headache and "drunken" speech. Within days she noticed some twitching of the right facial muscles and shaking and twitching in her hands. Medical workup shortly after onset suggested a diagnosis of myoclonic epilepsy of cortical origin.

During speech examination she complained of slurred speech. She noted a feeling of "tightness" in her face and neck intermittently when speaking. Oral mechanism examination was normal at rest. Myoclonic or tremorlike movements of the tongue were present during protrusion and lateral movements. There was no obvious palatal or pharyngeal myoclonus. Jaw and perioral myoclonus was more apparent during sustained phonation than when her mouth was open without phonation. Traces of nasal emission were apparent during pressure sound production. Some dystonic-like perioral movements were also apparent during speech. Her speech was characterized by reduced rate (3), and imprecise articulation (3), with difficulty achieving bilabial closure during connected speech, apparently secondary to dystonic lip contractions. Voice quality was strained-hoarse (2,3) with monotony and monoloudness (3). Prosody was characterized by excess and equal syllabic stress. Prolonged "ah" was unsteady (2). Speech AMRs were regular when produced

at a rate of one per second but markedly irregular when she attempted to maximize rate. Intelligibility was reduced.

The clinician concluded that the patient had a "hyperkinetic dysarthria of AM. In addition, there appear to be some dystonic perioral movements during speech that make it difficult for her to achieve bilabial closure. Speech clearly worsens during attempts to increase speech rate, and she has consciously reduced her rate because of this." Some suspicion was raised about accompanying ataxic and spastic components to her dysarthria, although it was felt that her scanning prosody and strained voice could well be secondary to efforts to compensate for her hyperkinetic dysarthria.

Neurologic evaluation indicated the presence of ataxia in the limbs, hyperreflexia, and action-induced myoclonus of the trunk, extremities, and face.

A complete workup confirmed a diagnosis of myoclonus epilepsy of cortical origin. Etiology was unclear, but an undiagnosed viral illness was felt to be the most likely cause.

Commentary. (1) Some movement disorders may be speech specific. (2) AM can cause dysarthria, one whose manifestations are noticeably exacerbated by increased speaking rate. In some cases, the myoclonus is associated with dystonic-like movements.

Case 8-4

A 35-year-old woman presented with a 2-year history of gradual mental deterioration, handwriting difficulty, reduced ability to concentrate, and reduced personal hygiene. She complained of speech difficulty, stating, "Nobody can understand me." There was a family history of Huntington's disease, most convincingly present in the patient's father, who died at age 45. Neurologic evaluation identified difficulty with balance and the presence of involuntary movements, generalized motor impersistence, mild cogwheel rigidity, and probable dementia. Neuropsychological assessment confirmed the presence of significant cognitive or memory limitations.

MRI showed an abnormality in the right putamen that could represent the iron deposition sometimes seen in Huntington's disease. Mild generalized atrophy was also present.

During speech examination, rapid, unsustained, choreic-like movements of the lower face, jaw, and tongue were present at rest. Involuntary tongue clicking was noted. She had difficulty maintaining a protruded tongue, open mouth, and lip retraction, as much because of motor impersistence as involuntary movements. Speech was characterized by accelerated rate (1,2), imprecise articulation with irregular articulatory breakdowns (1,2), dysprosody (2), and variable rate (1,2). Choreiform movements tended to delay the initiation of speech or delay continuation of speech at phrase boundaries. Vowel prolongation was characterized by a low amplitude tremor. Speech AMRs were irregular (1,2).

Pitch and loudness variability was reduced, but pitch and loudness occasionally varied inappropriately.

The clinician concluded, "hyperkinetic dysarthria associated with dyskinetic or choreiform movements of the lower face, jaw, and tongue. Her tendency toward accelerated rate and monopitch and monoloudness raise the possibility of an accompanying hypokinetic component, although it is possible that those characteristics are secondary to efforts to race through speech before the next occurrence of orofacial involuntary movements." The clinician also noted that the patient seemed impaired cognitively and often responded impulsively. She was seen for one session of speech therapy, during which she demonstrated an ability to slow her rate and improve articulatory precision. She was unable to do this without constant reminders, however. The family was counseled about the best strategy to use when they were unable to understand the patient; this focused primarily on cueing her to reduce speech rate.

Commentary. (1) Hyperkinetic dysarthria and orofacial choreiform movements may be among the presenting signs of Huntington's disease. (2) Cognitive deficits and personality changes often accompany the dysarthria in Huntington's disease. (3) People with chorea affecting speech sometimes accelerate rate in order to complete a statement before the next involuntary movement. This may give the appearance of an accompanying hypokinetic component to their dysarthria; the distinction between hypokinetic dysarthria and such compensatory efforts can be difficult to make in such cases.

Case 8-5

A 70-year-old woman presented with a 1-year history of voice difficulty. She denied chewing or swallowing difficulty. She had never smoked and did not abuse alcohol. An ear, nose, and throat (ENT) examination was normal. She was referred for speech evaluation.

During speech assessment, she reported the gradual emergence of voice difficulty that she described as "a quiver." This worsened under conditions of stress and fatigue. She denied other speech difficulties or problems with chewing or swallowing. She felt self-conscious about her voice, and it occasionally made her reluctant to speak. She reported that her father had "parkinsonism" and that her 71-year-old brother had some "shaking in his hands."

Oral mechanism examination was normal in size, strength, and symmetry. There was a subtle low-amplitude tremor of her lips at rest, and tremor of her jaw, tongue, palate, and pharynx were quite apparent during vowel prolongation. Articulation and resonance were normal. During conversation, a voice tremor with occasional voice interruptions was apparent. The tremor was particularly apparent during vowel prolongation. There was no evidence of respiratory tremor during prolonged voiceless fricatives or prolonged audible exhalations.

The clinician concluded, "Organic voice tremor with tremor frequency in the 5- to 8-Hz range. No other speech-language abnormalities detected. There are no

Case 8-5—cont'd

other deviant speech characteristics to suggest the presence of hypokinetic dysarthria, which might reflect early Parkinson's disease." This impression was discussed with the patient, who was relieved to have a diagnosis. She expressed concern, however, that her voice difficulty might reflect Parkinson's disease. She was referred for neurologic assessment to rule out PD. Neurologic examination was normal, with the exception of the voice tremor. There was no evidence of parkinsonism. Propranolol was prescribed in an effort to reduce the voice tremor but was ineffective.

Commentary. (1) Voice tremor can be an isolated manifestation of dysarthria. (2) Laryngeal tremor may not be apparent (or may be missed) during laryngeal

examination, and correct diagnosis is often made solely on the basis of perception of voice tremor. (3) Organic voice tremor can occur in the absence of other neurologic signs. (4) In addition to voice tremor's effect on communication ability, it often raises concerns in the patient about more serious neurologic disease. In this case the patient could be reassured that her condition was probably benign. The speech pathologist's impression was confirmed during neurologic evaluation. (5) In some cases the most effective management of a speech problem is correct diagnosis. This patient expressed relief about her diagnosis and a relative lack of concern about the minor difficulties her voice problem was causing her in some social situations.

Case 8-6

A 73-year-old woman presented with a 10-year history of voice difficulty that was present upon awakening one day, without obvious explanation. The problem worsened but had been stable for 3 to 4 years. She had had several periods of speech therapy, without benefit. Neurologic evaluation identified the presence of a head tremor, postural upper extremity tremor, and "spastic speech." A cause for these abnormal movements was not identified during a complete neurologic workup.

During speech examination, the patient associated onset of her voice problem with a period of considerable psychologic stress (her adopted son was having difficulty with drugs and was in the process of attempting to locate his biologic parents). She also noted that her voice worsened when she was anxious or spoke in a group. She noted mild improvement in her voice when she had a glass of wine.

Her voice was characterized by a tremor that consistently interrupted her voice and mildly slowed speech rate. Prolonged "ah" contained consistent, somewhat irregular, and strained voice interruptions. At higher pitches, voice interruptions disappeared and a tremor became apparent. Tremor was not apparent during prolongation of voiceless fricatives.

The clinician concluded, "Adductor spasmodic dysphonia of essential voice tremor, moderate to marked in severity."

Botox injection (discussed in Chapter 17) was recommended. Her voice improved significantly after several weeks of a weak-breathy dysphonia and mild swallowing difficulty. She noted a marked reduction in physical effort to speak and was pleased with her voice quality. Voice quality was indeed markedly improved,

although evidence of mild voice tremor persisted, but without voice interruptions.

Commentary. (1) Adductor spasmodic dysphonia can develop in association with organic voice tremor. Voice tremor may be accompanied by tremor elsewhere in the body, particularly in the jaw, face, tongue, palate, and pharynx. (2) The onset of spasmodic dysphonia is often associated with psychologic stress, even when examination reveals an organic basis for the problem. The relationship between psychologic stress and neurogenic spasmodic dysphonia is unclear, but the presence of psychologic stress at the time of onset does not rule out the possibility of neurogenic etiology for persistent voice difficulty. (3) Proper diagnosis of adductor spasmodic dysphonia can lead to fairly effective treatment of the disorder. A 68-year-old man presented with complaints of gradually progressive dizziness, visual difficulties, and slurred speech and mild swallowing difficulty. He had had a "mild" stroke and subsequent left carotid endarterectomy 8 years previously, but his speech and visual difficulties did not emerge until 2 years later. Neurologic examination was normal except for abnormal speech. Concern was raised about motor neuron disease. The patient was referred for EMG, MRI, and ENT and speech consultations.

During speech evaluation the patient reported having some mild difficulty with speech following his stroke, with subsequent improvement, but then worsening in recent years, characterized by voice difficulty and occasional problems with pronunciation. He did not complain of swallowing difficulties during meals but felt he was having problems controlling saliva.

Case 8-7

Oral mechanism examination was normal with the exception of some quick myoclonic-like movements of the tongue and 2- to 4-Hz myoclonic movements of the palate at rest and during phonation. Hoarse-rough voice quality (2,3), sporadic voice breaks, and inconsistent, imprecise articulation (1) of lingual fricatives and affricates characterized speech. Speech AMRs and SMRs were normal in rate and rhythm. Vowel prolongation was variable but consistent with the rate of the palatal myoclonus.

The clinician concluded: "The patient has a palatolaryngeal and perhaps lingual myoclonus suggestive of dysfunction in the Guillain-Mollaret triangle (brainstem or cerebellum). I think his myoclonus can explain some of the variability in his voice and some of his inconsistent articulatory imprecision. I do not think it explains very well his rough-hoarse voice quality."

EMG and ENT evaluations were normal. MRI showed old lacunar strokes in the thalami and right caudate nucleus but no lesion in the brainstem or cerebellum. His speech difficulties may have resulted from

an undetectable brainstem stroke, but a degenerative neurologic disorder could not be ruled out. Clonazepam was prescribed in the hope that it would help the myoclonus.

The patient returned for neurologic reassessment year later with worsening of symptoms. He had been unable to tolerate the side effects of Clonazepam and had discontinued it. Examination revealed palatal myoclonus and obvious but mild gait unsteadiness. An MRI now showed clear evidence of hypertrophic olivary degeneration (see Figure 8-8 for illustration of hypertrophic olivary degeneration). He was not seen for speech reassessment. It was concluded that he had a neurodegenerative disorder that, at the present time, could not more clearly be defined.

Commentary. (1) Palatal-laryngeal myoclonus is well-localized disorder. Its presence in this case predicted the MRI abnormality that eventually emerged. (Although uncommon, PM can be the result of degenerative neurologic disease. (3) Changes in speech and oral mechanism examination can be among the first signs of neurologic disease.

SUMMARY

1. Hyperkinetic dysarthrias are usually associated with dysfunction of the basal ganglia control circuit, but can also be related to involvement of the cerebellar control circuit or other portions of the extrapyramidal system. They probably occur somewhat less frequently in speech pathology practices than other dysarthria types, but if organic voice tremor and neurogenic spasmodic dysphonias are included in such comparisons, they may be more prevalent than all other single dysarthria types. Their characteristics can be manifest in the respiratory, phonatory, resonatory, and articulatory levels of speech, and prosody is often prominently affected. The deviant speech characteristics of hyperkinetic dysarthrias reflect the effects on speech of abnormal rhythmic or irregular and unpredictable, rapid or slow involuntary movements.
2. Hyperkinetic dysarthrias are heterogeneous, both in terms of the various abnormal movements that can lead to it and the particular speech muscles affected by the involuntary movements. The movement disorders underlying them are often categorized by the degree to which they vary in speed and rhythmicity. The most

common abnormal movements associated with hyperkinetic dysarthrias include chorea, dystonia, athetosis, ST, myoclonus, tics, and tremor.

3. The cause of hyperkinetic dysarthrias is of unknown. This is particularly true when the movement disorder is limited to the speech cervical muscles. Toxic and metabolic conditions are frequent known causes, with anticholinergic or neuroleptic medications representing the most frequent toxic cause. Orofacial dyskinesias and dysarthria are often the first or only manifestation of drug toxicity and tardive dyskinesia. Hyperkinetic dysarthrias are not uncommonly associated with degenerative neurologic conditions. Infection, neoplasm, trauma, and stroke are possible but infrequent causes.
4. The jaw, face, and tongue are frequently affected by hyperkinesias, usually in combination, sometimes only a single speech structure involved. Organic voice tremor and spasmodic dysphonias frequently have speech abnormalities that are perceptually limited to phonatory functions. Sometimes the involuntary movements are action induced and occur only during speech. In such cases the dysarthria is sometimes misdiagnosed as psychogenic in origin.

5. Patient complaints and specific deviant speech characteristics are quite variable, and they depend on the type of involuntary movement and the specific levels of the speech system affected. Distinctions can generally be made among dysarthrias that are due to chorea, dystonia, athetosis, ST, PM, AM, tics, organic voice tremor, and spasmodic dysphonias. These distinctions are what justify consideration of hyperkinetic dysarthria as a plural disorder, with subtypes based on the nature of the underlying involuntary movement.
6. In general, acoustic and physiologic studies have provided support for the auditory-perceptual characteristics of hyperkinetic dysarthrias, have specified more precisely the disorder's acoustic and physiologic characteristics, and have established approaches to documenting and quantifying relevant parameters of the disorder.
7. Hyperkinetic dysarthria can be the only, the first, or among the first and most prominent manifestations of neurologic disease. Its recognition can aid neurologic localization and diagnosis and may contribute to the medical and behavioral management of the individual's disease and speech disorder.

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