

"It was normal at first. Now it's gotten worse. I don't pronounce my words right. Some words I can't even say, and my voice is even different."

(64-year-old man with a mixed spastic-hypokinetic dysarthria associated with an unspecified neurodegenerative disease)

CHAPTER OUTLINE

I. Etiologies

- A. Degenerative diseases
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- C. Vascular disorders
- D. Trauma
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II. Speech pathology

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IV. Summary

Imposing functional and anatomic divisions on the nervous system helps us understand the brain's operations and establish a framework for localizing and categorizing nervous system diseases. Unfortunately, however, there is no rule of nature that obligates neurologic disease to restrict itself to the divisions we impose upon it. As a result, the effects of neurologic disease can be "mixed" or distributed across two or more divisions of the nervous system.

The frequent refusal of neurologic disease to be focal and compartmentalized has implications for motor speech disorders (MSDs). Chapters 4 through 9 focused on "pure" dysarthrias that reflect damage to only one of the divisions of the motor speech system. For practical clinical purposes at least, many people do have only a single type of dysarthria. However, it is often the case that the damage that causes dysarthria is not confined to a single component of the motor system. Thus many people with dysarthria have a *mixed dysarthria*, or combination of two or more of the types that have already been discussed.

Mixed dysarthrias are common. They are encountered as the primary speech disorder in a large medical practice at a considerably higher rate than any single dysarthria type. Based on data for primary communication disorder diagnoses within the Mayo Clinic Speech Pathology practice, it accounts for 29.1% of all dysarthrias and 26.9% of all MSDs (see Figure 1-3).

Does the fact that many dysarthrias are mixed minimize the value of categorizing them into types? No. In fact, because dysarthria type reflects underlying neuropathology, recognizing its mixed forms is also valuable to neurologic localization and diagnosis. For example, a patient with a diagnosis of Parkinson's disease (PD) who has a mixed hypokinetic-ataxic dysarthria may not have PD, or may have more than PD, because PD should not be associated with ataxic dysarthria. Thus the recognition of each component of a mixed dysarthria may help rule out certain neurologic diagnoses or make other diagnoses more likely.

Mixed dysarthrias represent a heterogeneous group of speech disorders and neurologic diseases. Virtually any combination of two or more of the single dysarthria types is possible, and in any particular mix any one of the components may predominate. In spite of its heterogeneity, and the fact that sorting out the various components of mixed dysarthrias can be quite difficult, many mixed dysarthrias are perceptually distinguishable. Also, like pure forms, they may be the first or among the first signs of neurologic disease.

In this chapter, common etiologies of mixed dysarthrias are reviewed, with an emphasis on diseases that are frequently encountered in neurology and medical speech pathology practices. The most common types of mixed dysarthrias and their relation to specific neurologic diseases are also addressed. Finally, the mixed dysarthrias that are encountered in several specific neurologic diseases are discussed because they have been studied sufficiently to permit clinical descriptions of their most salient characteristics. Their description helps establish that they are lawfully derived from diseases that affect more than one component of the brain's motor system.

ETIOLOGIES

Mixed dysarthrias can be caused by many conditions within each of the broad categories of neurologic disease. More than any other dysarthria type, they can result from combined neurologic events (e.g., multiple strokes) or the cooccurrence of two or more neurologic diseases (e.g., stroke plus PD). Also, they occur commonly in a number of degenerative diseases that affect more than one portion of the nervous system.

This section addresses conditions that can cause mixed dysarthrias more frequently than any single dysarthria type. The definition and description of conditions whose speech manifestations have been studied in some detail are emphasized. The specific speech characteristics associated with several of these disorders are addressed later in the section on speech pathology.

Degenerative Diseases

Because a number of degenerative diseases affect more than one portion of the motor system, they are commonly associated with mixed dysarthrias. Some of these diseases primarily affect motor functions. Others are more diffuse in their effects, also producing autonomic, sensory, and cognitive impairments.

Motor Neuron Disease—Amyotrophic Lateral Sclerosis

Motor neuron diseases (MNDs) are disorders characterized by progressive loss of upper motor neurons (UMNs) or lower motor neurons (LMNs), or both.

Spinal muscle atrophies are MNDs that affect LMNs only. Progressive limb wasting and weakness, with or without cranial nerve weakness, characterize them. They can be inherited or occur sporadically and may be congenital or develop in childhood or adulthood.¹⁶⁸ When dysarthria is present, it is flaccid, not mixed, so it is not discussed here further.

Progressive bulbar palsy (PBP) is a syndrome dominated by LMN weakness of cranial nerve muscles. Dysarthria and dysphagia are its predominant signs. UMN signs in the bulbar muscles may or may not be present.¹³¹ When it is confined to the LMNs, PBP is associated with flaccid, not mixed dysarthria. In a sense, PBP can be thought of as amyotrophic lateral sclerosis (ALS) without limb involvement.

Primary lateral sclerosis (PLS), or *progressive pseudobulbar palsy* when bulbar muscles are predominantly affected, is an MND that affects UMNs only. They are characterized by corticospinal or corticobulbar tract signs, or both, but without LMN involvement. They can be difficult to distinguish from ALS. They may be associated with spastic dysarthria (see Chapter 5).

ALS is the most common MND. It is characterized clinically by UMN and LMN signs in the limbs or bulbar muscles, or both, and neuropathologically by loss of motor neurons in the precentral and postcentral cortex, the corticospinal tracts, motor nuclei of cranial nerves, and anterior horns of the spinal cord. Atrophic PNS motor fibers and evidence of denervation are also present.¹⁶⁷ Because it is a mixed UMN and LMN disease that often affects the bulbar muscles, *ALS has a natural and common association with mixed spastic-flaccid dysarthria.*

The incidence of ALS is approximately 1 to 5 per 100,000 population. More men than women are affected. It usually occurs sporadically, but approximately 5% of cases are familial.^{131,168} Exact time of onset is often difficult to establish because more than half of the anterior horn cells must be lost before weakness is apparent and patients may adapt well to weakness.¹¹⁰ Approximately 80% of affected individuals develop symptoms between 40 and 70 years of age, and the peak rate of occurrence is between 60 and 70 years.^{84,110} The course of the disease is usually 1 to 5 years, but up to 25% of affected people live beyond 12 years.²¹ Death is usually related to respiratory failure.

Although its first signs and symptoms are usually in the limbs, *approximately 25% of patients have their initial problems in the bulbar muscles, most often first represented by dysarthria, and sometimes by dysphagia.*^{157,158} Patients with bulbar deficits as the first symptoms tend to have a more rapid course because dysphagia and airway problems represent major threats to life.

The diagnosis of ALS is made on the basis of its clinical profile and electrophysiologic confirmation. General clinical features include fatigue, cramping, fasciculations, weakness, and muscle atrophy, as well as hyperactive deep tendon reflexes with spasticity. Weakness is often focal initially. Electromyogram (EMG) findings of denervation (fibrillations) and reinnervation (large polyphasic motor unit action potentials) from two or more extremities (with the bulbar muscles counted as an extremity) are considered diagnostic of the disease.¹³¹ Eye movements and autonomic and cognitive functions are usually spared, but some patients have cognitive deficits or signs of parkinsonism.¹⁶⁸ Cognitive deficits tend to be greater and more frequently evident in people with bulbar onset of symptoms.^{98,144}

Multiple Sclerosis

Multiple sclerosis (MS) is the commonest acquired demyelinating central nervous system (CNS) disease and the commonest serious CNS disorder in young and middle-aged adults, affecting approximately 0.1% to 0.2% of the U.S. population. It affects women more often than men and usually begins between 20 and 40 years of age. Its cause is unknown, but it is believed to be an autoimmune disease triggered by environmental and genetic interactions.¹¹⁷

The disease affects scattered and diverse areas of the nervous system, with a predilection for white matter and periventricular areas, the brainstem, spinal cord, and optic nerves. MS plaques are characterized by demyelination (destruction of myelin sheaths with preservation of axons) and death of oligodendrocytes (cells that produce myelin) within the lesion.¹³⁰ Some lesions may be acute, with active myelin breakdown, whereas others reflect chronic, inactive demyelinated glial scars. In acute plaques, edema occurs in the area of affected nerve fibers. Resolution of edema may explain some of the recovery from deficits after an exacerbation.

*Initial presentation in the bulbar muscles aids neurologic differential diagnosis, because it is unusual in conditions that can mimic ALS, such as multifocal motor neuropathy, motor neuropathy, spinomuscular atrophy, or hyperthyroidism.¹⁵⁷

Diagnosis can be difficult, and approximately 10% of MS patients are misdiagnosed,⁵⁸ with misdiagnosis sometimes including hysteria.^{60,136} Current recommended diagnostic criteria¹⁰⁰ emphasize objective demonstration of disseminated lesions in both time and space, but clinical observations and other objective tests are also important. For example, a diagnosis of MS can be made if there is evidence of two or more attacks and objective evidence of two or more lesions. Various combinations of clinical findings and objective evidence of lesions can also permit the diagnosis, although with varying degrees of confidence. Among objective tests, magnetic resonance imaging (MRI) is emphasized because of its sensitivity to white matter lesions, but cerebrospinal fluid examination and visual evoked potentials are also helpful.

The course of MS is unpredictable. Some people have a benign course, with one or only a few attacks, and complete or nearly complete remission. Others have a relapsing-remitting course, with episodes of deterioration followed by near-complete recovery, a pattern that may persist for years. Still others have a remitting-progressive course with a slow accumulation of deficits. Finally, some have a progressing course, with the insidious onset and slow progression of disease without remission.¹¹⁷

MS can produce any sign or symptom of CNS disease. Problems with gait are common, as are visual and other sensory difficulties. Cerebellar dysfunction is often but not invariably present. Cranial nerve abnormalities may occur and can include trigeminal neuralgia, Bell's palsy, and facial myokymia. Psychiatric problems are not unusual and most often reflect affective disorders, which may be a direct consequence of the demyelinating process or a reaction to the disability caused by the disease.¹²² Cognitive deficits occur in as many as 25% of people with progressive MS.¹³⁹ Aphasia and apraxia of speech are rare but have been reported,^{57,92,119,124} as have difficulties with higher-level language functions that are not specifically aphasic in character.⁹¹

Dysphagia is relatively uncommon in patients who are ambulatory, but it does occur in others.⁹⁹ Dysarthria may occur in 50% of people with MS¹³³; it is uncommon at the onset of the disease but can be the presenting symptom (see Chapter 6 for a discussion of paroxysmal ataxic dysarthria). When present, *dysarthria may reflect nearly any single type or combination of single types. A spastic-ataxic dysarthria may be the most common mixed dysarthria associated with MS, but it should not be considered the dysarthria of MS.*

Friedreich's Ataxia

Friedreich's ataxia (FA) is an inherited degenerative disease that is predominantly spinocerebellar, but it may also be associated with spasticity, LMN weakness, and extrapyramidal movement disorders. It was discussed in Chapter 6, but it clearly can be associated with mixed dysarthria, most often *ataxic* and *spastic*.

Progressive Supranuclear Palsy

Progressive supranuclear palsy (PSP) is a multisystem neurodegenerative disease that is often mistaken for PD. Its incidence is approximately 1 to 6.5 per 100,000 population. It affects more men than women and usually begins after age 50; incidence increases with age. Average survival from symptom onset to death is approximately 6 to 7 years. It usually occurs sporadically rather than within families. Etiology is unknown.^{20,152}

Neuropathologic characteristics include cell loss in numerous areas of the brain, including structures and pathways of the motor system, such as the globus pallidus, substantia nigra, thalamus, subthalamic nucleus, midbrain, a number of brainstem nuclei, and the cerebellum. The cranial nerves and the cerebral cortex, with the exception of the frontal lobes, are usually spared.^{88,152}

Clinically, PSP is characterized by supranuclear ophthalmoparesis (paralysis of vertical gaze, especially downgaze), postural instability, and signs of parkinsonism (e.g., rigidity, bradykinesia). In spite of parkinsonian signs, tremor is usually not prominent, and responsiveness to antiparkinsonism drugs is usually poor or absent. *Dysarthria and dysphagia are common and often early and prominent signs.* Personality and cognitive changes associated with frontal lobe dysfunction (e.g., apathy, irritability, difficulty in planning and sequencing) can be quite evident.^{48,94}

The clinical signs and pathology of PSP are indicative of multisystem degeneration. Several dysarthria types are possible. Mixed dysarthria occurs frequently, most often in the form of various combinations of *hypokinetic*, *spastic*, and *ataxic* types.

Multiple System Atrophy

Multiple system atrophy (MSA) is a sporadic neurodegenerative condition characterized by varying combinations of parkinsonism, ataxia, spasticity, and autonomic dysfunction. Similar to PSP, it is sometimes mistaken for PD, but response to levodopa is suboptimal. Incidence is approximately 3 per 100,000 population. Onset is usually after 50 years

of age, and average survival from symptom onset until death is approximately 9 years.^{20,38}

Neuropathologic localization varies somewhat across MSA subtypes (discussed later), but the range of involvement includes neuronal loss and gliosis in the basal ganglia, substantia nigra, cerebellum, inferior olives, middle cerebellar peduncles, pontine nuclei, corticospinal tracts, and intermediolateral and anterior horn cells. Cerebral atrophy, especially in the frontal lobes, has also been documented.³⁸ These loci implicate the basal ganglia and cerebellar control circuits, as well as UMN pathways. As a result, *hypokinetic*, *hyperkinetic*, *ataxic*, or *spastic* dysarthria may be encountered in MSA.

MSA recently has become the preferred designation for three previously separated conditions, including *striatonigral degeneration*, *olivopontocerebellar atrophy (OPCA)*, and *Shy-Drager syndrome*. Two MSA subtypes are now identified, *MSA-P when parkinsonian features predominate* and *MSA-C when cerebellar features predominate*.^{*} Because the literature contains references to both sets of terminology, the labels of Shy-Drager syndrome, olivopontocerebellar atrophy, and striatonigral degeneration are retained when discussing studies that have used those designations.

In general, MSA-P (and striatonigral degeneration) reflects predominant, although not exclusive, nerve cell loss and gliosis in the basal ganglia and substantia nigra. As a result, parkinsonian features tend to dominate clinical signs and symptoms. When dysarthria is present, the hypokinetic type would most often be expected, either as the only dysarthria type or in combination with spastic or ataxic dysarthria, or both. Similarly, MSA-C and OPCA reflect predominant, although not exclusive, cerebellar involvement. When dysarthria is present, ataxia would most often be expected, either as the only dysarthria type or in combination with spastic or hypokinetic types, or both. In Shy-Drager syndrome, there are usually prominent autonomic nervous system deficits (dysautonomia), such as orthostatic hypotension,[†] incontinence, reduced respiration, and impotence. These problems stem from loss of preganglionic sympathetic neurons in the intermediolateral horns. Because the substantia nigra, striatum, cerebellum, and corticospinal tracts are also affected, various combinations of parkinsonism, ataxia, and spasticity, along with their associated dysarthrias (and, sometimes, laryngeal stridor), may predominate.

^{*}Based on recommendations of a Consensus Committee of the American Autonomic Society and the American Academy of Neurology.¹⁴⁶

[†]Orthostatic hypotension is characterized by a decrease in blood pressure upon standing.

Corticobasal Degeneration

Corticobasal degeneration (CBD) is an uncommon neurodegenerative disease of unknown etiology that is characterized by asymmetric cortical and extrapyramidal signs. A striking feature is the asymmetry with which CBD presents, even though its progressive course eventually includes the cortex (frontal and parietal lobes most prominently) and basal ganglia bilaterally. Onset is usually between 50 and 70 years of age, usually with a 5- to 15-year progression to death.^{13,17,38}

The most consistent clinical features of CBD are asymmetric limb rigidity and apraxia. Asymmetric dystonic limb posturing, myoclonus, tremor, and cortical sensory loss are also common. Other fairly distinctive signs include *alien limb phenomena* and *mirror movements*.^{*} Frontal release signs, ataxia, postural instability, nonaphasic cognitive deficits, aphasia, apraxia of speech, and dysarthria can also occur.¹⁶ The dysarthria is usually mixed, with *spastic* and *hypokinetic* types being most common, but *hyperkinetic* and *ataxic* components are possible.

Toxic-Metabolic Conditions

When toxic and metabolic diseases alter neurologic functions, their effects tend to be diffuse. When they affect the motor system, they commonly affect more than one of its components. When motor speech is affected, the result is often a mixed dysarthria. Some toxic-metabolic conditions that may be associated with mixed dysarthrias are described as follows.

Wilson's Disease

Wilson's disease (WD) is a rare autosomal recessive genetic metabolic disorder associated with inadequate processing of dietary copper. It is also known as *hepatolenticular degeneration* to indicate liver involvement and the consistent postmortem findings of degeneration in the lenticular nuclei of the basal ganglia. The metabolic inadequacy in WD leads to a buildup of copper in the liver, brain, and cornea of the eye, with the appearance of neuromotor signs by late adolescence or early adulthood. WD can be fatal if it goes undiagnosed.

WD may present with a hepatic, neurologic, or mood disturbance.⁴⁹ The pathognomonic sign of the disease is a golden brown ring (*Kayser-Fleischer*

rings) around the cornea of the eyes, reflecting copper deposits. Its classic neurologic manifestations are motor in nature and most frequently include a wing-beating tremor when the arms are outstretched; truncal rigidity; slowness of movement; incoordination; dystonia; dysarthria; drooling; and facial masking or a grinning, vacuous smile.^{2,49} The basal ganglia are usually the most severely affected structures. If diagnosed before permanent damage occurs, a low copper diet, substances such as zinc to reduce copper absorption, and agents such as penicillamine to promote urinary excretion of copper can control copper balance and reverse many of the neurologic manifestations. Unfortunately, dysarthria is one of the neurologic signs that tend to be resistant to these treatments.¹²¹ Liver transplant is pursued with fulminant liver failure.⁴⁹

*Dysarthria is considered a cardinal feature of WD, and it may be its initial sign.*¹¹⁸ The most common types of dysarthria associated with WD are *hypokinetic*, *spastic*, and *ataxic*.

Hepatocerebral Degeneration

Hepatocerebral degeneration can occur in people who have survived episodes of hepatic coma or in people with chronic liver disease. Common clinical manifestations include limb tremor, chorea or choreoathetosis in the face and limbs, unsteady gait, ataxia, and dysarthria. Corticospinal signs and mental deterioration may also be present. Pathologically, abnormalities are noted in the cerebral cortex, the lenticular nuclei, thalamus, and a number of brainstem nuclei. The lesions are similar to those encountered in WD.²

The dysarthrias associated with this condition have not been studied. The presence of *hypokinetic*, *hyperkinetic*, *spastic*, or *ataxic* forms seems possible.

Hypoxic Encephalopathy

Hypoxic encephalopathy is a diffuse neurologic condition resulting from a lack of oxygen to the brain because of failure of the heart and circulation or failure of the lungs and respiration. These failures most often involve myocardial infarction or cardiac arrest, carbon monoxide poisoning, suffocation (e.g., drowning, strangulation), diseases that paralyze respiratory muscles (e.g., Guillain-Barré syndrome), or diffuse CNS damage (e.g., traumatic brain injury [TBI]).

In general, when consciousness is lost and oxygen deprivation exceeds several minutes, permanent neurologic damage occurs. If consciousness and responsiveness are regained, several clinical abnormalities may emerge. Among the most common are memory

^{*}The alien limb phenomenon is characterized by involuntary extremity movements such as elevation of the arm or grasping of objects, with the limb often described by patients as having a mind of its own. Mirror movements are characterized by inappropriate, involuntary movements of a limb that crudely mirror those of the contralateral limb as it performs volitional activity; they are usually associated with parietal lobe damage.¹⁷

disturbances, personality changes and disruptive behavioral problems, poor insight, visuospatial problems, spasticity, ataxia, dystonia, parkinsonism, tremor, action myoclonus, and pseudobulbar palsy. *Delayed postanoxic encephalopathy*, characterized by mental status changes and parkinsonism, occurs in some patients, most often days to weeks after carbon monoxide poisoning.²⁷

The dysarthrias associated with hypoxic encephalopathy have not been described. The involvement of cortical, extrapyramidal, and cerebellar structures predicts the possible emergence of a number of dysarthria types that could include, at the least, *hypokinetic*, *hyperkinetic*, and *ataxic* forms, either singly or in combination.

Central Pontine Myelinolysis

Central pontine myelinolysis (CPM) is a serious metabolic condition characterized by destruction of myelin in the base of the pons. It also can affect the thalamus, subthalamus, amygdala, striatum, internal capsule, lateral geniculate bodies, white matter of the cerebellum, and deep layers of the cerebral cortex and adjacent white matter.

CPM is often associated with alcoholism and other conditions associated with malnutrition. It can occur in people with chronic liver or kidney disease or after organ transplant. It is believed that the basis pontis and other affected structures are especially susceptible to some acute metabolic fault, such as rapid correction or overcorrection of a profound electrolytic disturbance, such as hyponatremia.^{2,97}

A number of neurologic signs are possible in CPM. Quadriplegia, spasticity, pseudobulbar palsy, and dysarthria or anarthria are common.

The dysarthrias of CPM have not been studied. Clinical experience suggests that *spastic*, *ataxic*, and *hyperkinetic* forms, at the least, can occur.

Vascular Disorders

Multiple strokes that affect various components of the motor system have a natural association with mixed dysarthrias. They can produce any combination of dysarthria types. Single brainstem strokes can also result in mixed dysarthrias because of the close proximity of pyramidal and extrapyramidal fibers, the cerebellar control circuit, and cranial nerve nuclei in that area of the brain. As a result, various combinations of *spastic*, *ataxic*, and *flaccid* dysarthria are not uncommon in brainstem stroke and can even occur in hemispheric stroke. *Hyperkinetic* dysarthria (e.g., secondary to palatal myoclonus) can also occur.

Trauma

The diffuse or multifocal lesions associated with traumatic and closed head injuries (CHIs) can produce virtually any combination of dysarthrias. Trauma from neurosurgery, especially if it involves posterior fossa structures, can also result in various mixed dysarthrias.

Tumor

Tumors, especially in the brainstem, can cause mixed dysarthrias because they can invade or produce mass effects on multiple components of the nervous systems. Brainstem tumors can be associated with various combinations of *spastic*, *ataxic*, and *flaccid* dysarthria.

Infectious and Autoimmune Diseases

The diffuse or multifocal effects of infectious and autoimmune diseases such as meningitis, encephalitis, and acquired immunodeficiency syndrome (AIDS) can be associated with various mixed dysarthrias. Two examples of such conditions, *progressive multifocal leukoencephalopathy** and *systemic lupus erythematosus*, are addressed here.

Progressive multifocal leukoencephalopathy (PML) is a rare, usually viral demyelinating CNS disease. It tends to occur in people with autoimmune disorders (e.g., AIDS, lymphoma, chronic lymphocytic leukemia) or in people receiving immunosuppressive therapy. The predominantly white matter lesions in PML are most prominent in cerebral subcortical areas and the posterior fossa. Clinical features include personality changes; motor deficits; ataxia; visual and other sensory deficits; and speech, language, and cognitive problems.^{8,91}

The dysarthrias of PML have not been studied in detail. Lethlean and Murdoch,⁹¹ describing the language deficits of one woman with PML, noted the presence of severe dysarthria; type was not identified, but clinical features suggest it was mixed, possibly with spastic and ataxic components. Dysarthria has been among the initial neurologic manifestations of PML in people undergoing chemotherapy (5-fluorouracil and levamisole) for colon cancer.^{62,78} The presence of multiple and scattered hemispheric and brainstem lesions makes it likely that the dysarthrias were mixed. Because patients improved when chemotherapy was discontinued or modified, toxic effects of chemotherapy were the suspected cause.

*Leukoencephalopathy is also discussed briefly in Chapter 5.

Mixed hypokinetic, spastic, and ataxic dysarthria, plus apraxia of speech, have been reported in a patient without PML or any other structural lesions who was receiving the immunosuppressive agent FK-506 following liver transplantation.¹⁸

It appears that recognition of a developing dysarthria may be an early indication of neurotoxicity in this type of chemotherapy.

Systemic lupus erythematosus (SLE) is an autoimmune disease that can affect any organ, including the nervous system. Its effects on the nervous system can be temporary or permanent, and it can affect multiple, diffuse areas of the nervous system. Mechanisms of damage can be multiple and complex but are usually vascular. Various speech, language, and cognitive-communication disorders can result from SLE,¹⁷⁵ including dysarthria, which, on the basis of clinical experience, can be mixed. Specific speech characteristics and dysarthria types have not been studied carefully, however.

■ SPEECH PATHOLOGY

Sorting out the individual components of mixed dysarthrias can be difficult. Clinical uncertainty about all or some of the components of a mixed dysarthria probably occurs much more frequently than for the diagnosis of any single dysarthria type. It is not unusual, for example, to identify with confidence one of the components but to be uncertain if a second or third or fourth component is also present. Diagnostic impressions such as “the patient has an unambiguous mixed spastic-ataxic dysarthria, possibly with an

accompanying flaccid component,” or “ataxic dysarthria versus mixed spastic-ataxic dysarthria” are not unusual in clinical practice. This uncertainty probably reflects combinations of the natural overlap among manifestations of diseases affecting several portions of the motor system, the shortcomings of perceptual methods, and the “true” equivocal presence of certain neurologic signs in some cases. The need to draw equivocal or qualified conclusions can be unsettling to a clinician’s desire for certainty and precision, but there is little choice when uncertainty reflects clinical reality. It may be reassuring, or equally as unsettling, to know that clinical neurologic examinations frequently reach similar tenuous interpretations of signs and symptoms.

Table 10-1 summarizes the types of dysarthria that can be encountered in a number of neurologic diseases that can produce mixed dysarthrias. It may be useful for setting a range of expectations for types of dysarthria that may be present when a neurologic diagnosis is relatively unambiguous and for identifying mixed dysarthrias that may be incompatible with particular neurologic diagnoses. Chapter 15, which addresses differential diagnosis, summarizes distinctive features of each of the single dysarthria types in a manner that helps identify each component that makes up a mixed dysarthria (in particular, see Tables 15-3 and 15-4).

table

10-1

Types of dysarthria that may be present in neurologic diseases that can produce mixed dysarthrias. Dysarthria is not inevitably present in all people with these diseases, and the listed diseases are not exhaustive.

| Disease | Dysarthria | | | | | |
|--|------------|---------|--------|-------------|--------------|------|
| | Flaccid | Spastic | Ataxic | Hypokinetic | Hyperkinetic | UUMN |
| Degenerative | | | | | | |
| ALS* | ++ | ++ | ? | - | - | - |
| MS | + | +/++ | +/++ | + | + | + |
| Friedreich's ataxia | ?/+ | + | ++ | - | - | - |
| PSP | - | ++ | + | ++ | - | - |
| Multiple system atrophy | +/? | +/++ | ++ | ++ | +/? | - |
| Corticobasal degeneration* | - | +/++ | + | +/++ | ? | ? |
| Toxic-metabolic | | | | | | |
| Wilson's disease | - | +/++ | +/++ | ++ | ?/+ | - |
| Hepatocerebral degeneration [†] | - | + | + | + | + | - |
| Hypoxic encephalopathy [†] | - | ? | +/++ | +/++ | +/++ | - |
| CPM [†] | - | +/++ | +/++ | ? | +/++ | - |
| Vascular [†] | + | +/++ | +/++ | + | + | +/++ |
| Tumor [†] | + | + | + | +/? | +/? | + |
| Infectious* [†] | + | + | + | + | + | + |
| Traumatic* | + | +/++ | +/++ | +/++ | + | + |

ALS, Amyotrophic lateral sclerosis; CPM, central pontine myelinolysis; MS, multiple sclerosis; PSP, progressive supranuclear palsy; UMN, unilateral upper motor neuron; ++, often present when dysarthria is present—may be quite typical for a particular disease; +, sometimes present, but not necessarily “typical” for a particular disease; ?, uncommon or of uncertain presence; -, not present.

*Apraxia of speech may also be present.

[†]Dysarthria has not been explicitly studied in the particular disorder.

| box 10-1 | Etiologies for 406 quasirandomly selected cases with a primary speech pathology diagnosis of mixed dysarthria at the Mayo Clinic from 1969-1990 and 1999-2001. Percentage of cases under each broad etiologic heading is given in parentheses. |
|---|--|
| <p>Degenerative (66%)</p> <p>ALS (includes diagnoses of MND and progressive bulbar palsy) (43%) Nonspecific CNS degenerative disease (8%) PSP (4%) PD or parkinsonism (2%) Olivopontocerebellar degeneration (2%) Multiple systems atrophy (2%) Other (Wilson's disease, asymmetric cortical degeneration, Shy-Drager syndrome, cerebellar degeneration, striatonigral degeneration, hepatocerebral degeneration, Creutzfeldt-Jakob disease, PD vs. Shy-Drager disease, PD vs. PSP) (6%)</p> <p>Vascular (11%)</p> <p>Multiple strokes (7%) Single stroke (4%) Vascular malformation (<1%)</p> <p>Traumatic (5%)</p> <p>CHI (3%) Surgical (<1%)</p> | <p>Multiple Causes (5%)</p> <p>Various combinations of stroke, encephalopathy, cerebellar degeneration, Shy-Drager syndrome, postthalamotomy or other neurosurgery, PD, parkinsonism, drug toxicity, CHI, primary lateral sclerosis</p> <p>Demyelinating (4%)</p> <p>Multiple sclerosis (3%) Other (1%)</p> <p>Tumor (4%)</p> <p>Mass (mostly posterior fossa) (4%) Paraneoplastic (<1%)</p> <p>Undetermined (3%)</p> <p>Toxic/Metabolic (1%)</p> <p>Hypothyroidism, central pontine myelinolysis, gangliosidosis, neuroleptic toxicity, hypoxic encephalopathy, hepatic or metabolic encephalopathy, undetermined metabolic disease</p> <p>Inflammatory (1%)</p> <p>Postviral encephalopathy, progressive encephalopathy, spongiiform encephalopathy</p> |

ALS, Amyotrophic lateral sclerosis; CHI, closed head injury; CNS, central nervous system; MND, motor neuron disease; PD, Parkinson's disease; PSP, progressive supranuclear palsy.

In the remainder of this section, common etiologies of mixed dysarthrias and the most common mixed dysarthrias encountered in clinical practice are discussed. The dysarthrias encountered in specific neurologic diseases, the speech characteristics of which have been studied in some detail, are also summarized.

Distribution of Etiology, Types, and Severity in Clinical Practice

Etiologies

Box 10-1 and Figure 10-1 summarize the etiologies for 406 quasirandomly selected cases seen at the Mayo Clinic with a primary speech pathology diagnosis of mixed dysarthria. The cautions expressed in Chapter 4 about generalizing these data to the general population or all speech pathology practices apply here as well.

The data establish that mixed dysarthrias can be caused by a wide variety of neurologic conditions. Approximately two thirds of the cases were accounted for by degenerative diseases, and nearly

80% were accounted for by degenerative and vascular diseases.

By far, ALS was the most frequent degenerative neurologic disease associated with mixed dysarthria (43% of all cases).^{*} Eight percent of cases clearly had degenerative CNS disease, but the diagnosis was otherwise nonspecific. The remaining degenerative cases were spread across many of the neurodegenerative diseases discussed earlier in this chapter.

Multiple strokes accounted for more than half of the vascular cases (11% of all cases). The sites of the strokes were widely distributed within the CNS and included both hemispheres, the brainstem, and cerebellum. Single strokes causing mixed dysarthrias were nearly always in the brainstem.

CHIs accounted for most of the traumatic etiologies (4% of all cases). Lesions, when identifiable, were widely distributed in the brain, most

^{*}Although this figure may approximate that encountered in large tertiary medical care centers, it is almost certainly an overestimate of the percentage of cases seen in speech pathology practices in rehabilitation or primary care settings.

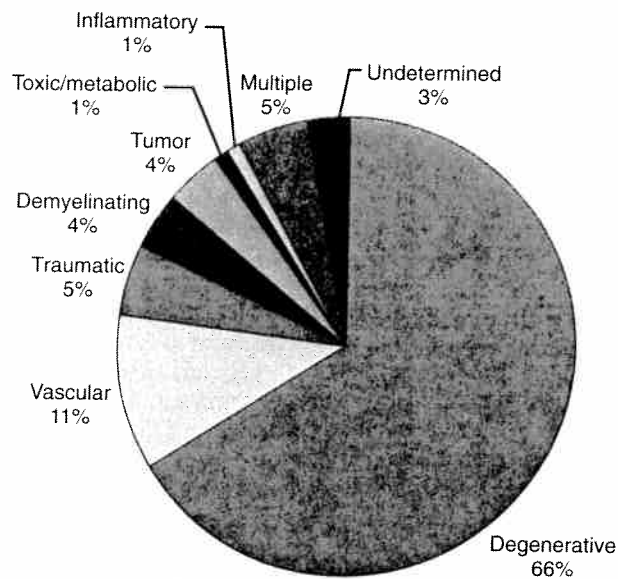


FIGURE 10-1 Distribution of etiologies for 406 quasirandomly selected cases with a primary speech pathology diagnosis of mixed dysarthria at the Mayo Clinic from 1969-1990 and 1999-2001 (see Box 10-1 for details).

often in subcortical areas, the brainstem, or cerebellum. Tumors accounted for 4% of the cases; a majority of the tumors were located in the posterior fossa.

Demyelinating diseases accounted for 4% of the cases. The majority of those cases had MS.

A combination of diseases was present in approximately 5% of the cases. These included various combinations of vascular, inflammatory, degenerative, traumatic, and toxic/metabolic conditions. Toxic/metabolic and inflammatory etiologies, by themselves, were responsible for only a small percentage of cases. Finally, approximately 3% of cases had undetermined neurologic diagnoses. The indeterminate nature of these disorders ranged from unexplained signs and symptoms (e.g., dysarthria, dystonia, blepharospasm) to identifiable lesions of undetermined etiology (e.g., undetermined posterior fossa lesion).

Types of Mixed Dysarthrias

The combination of dysarthria types was examined for the first 300 cases encountered within the sample summarized in Box 10-1. A combination of two dysarthrias represented 84% of the cases. Fourteen percent had three dysarthria types, and 2% had a combination of four types. The dominance of two dysarthria types in mixed dysarthria reflects either the "reality" of localization of neurologic disease in the sample or the limitations on auditory perceptual abilities to detect more than two dysarthria types in

any one person; a combination of these explanation is likely.*

Table 10-2 summarizes the frequency of occurrence of each single dysarthria type within the 300 patients. Because ALS occurred so frequently, the distribution for the entire sample and that portion of the sample minus ALS cases are also given. Spastic dysarthria was the most common type encountered being present in 91% of the entire sample and 85% of the sample without ALS. Across both samples ataxic dysarthria was the next most frequently encountered. Flaccid dysarthria was present in a majority of the entire sample but only one quarter of the sample without ALS. Hypokinetic dysarthria was present somewhat less frequently, although present in 35% of the sample without ALS. Hyperkinetic dysarthria was the least frequently encountered but nonetheless was present in far more than a few patients in both samples.

Table 10-3 summarizes the most common types of mixed dysarthria for the sample of 300 patients. The most common neurologic diagnosis for each mixed type is also given. Mixed flaccid-spastic dysarthria was the most frequent mixed dysarthria, accounting for 42% of the entire sample. The vast

*The difficulty that can be encountered in sorting out types in mixed dysarthrias is highlighted by the fact that one component of the mixed dysarthria was considered questionably or equivocally present in approximately 6% of the 300 cases. This uncertainty was associated with flaccid, spastic, ataxic, and hypokinetic types.

table

10-2

Distribution of individual dysarthria types encountered in a sample of 300 people with a primary speech pathology diagnosis of mixed dysarthria. Percentages are given for the entire sample and the portion of the sample without a diagnosis of ALS.

| Type | Entire Sample | Sample without ALS |
|--------------|---------------|--------------------|
| Flaccid | 54% | 25% |
| Spastic | 91% | 85% |
| Ataxic | 43% | 66% |
| Hypokinetic | 21% | 35% |
| Hyperkinetic | 13% | 21% |

ALS, Amyotrophic lateral sclerosis.

table

10-3

The most common types of mixed dysarthria and the most frequent neurologic diagnoses in a sample of 300 people with a primary speech pathology diagnosis of mixed dysarthria.

| Type (% of Entire Sample) | Neurologic Diagnosis (% of Category) |
|--------------------------------------|--|
| Flaccid-Spastic (42%) | ALS (88%) |
| | Vascular (5%) |
| | Tumor (2%) |
| | Other (5%) |
| | — |
| Ataxic-Spastic (23%) | Vascular (17%) |
| | Demyelinating (13%) |
| | CNS degenerative disease (12%) |
| | Inflammatory (9%) |
| | Cerebellar degeneration (7%) |
| | Spinocerebellar degeneration (6%) |
| | Tumor (6%) |
| | Trauma (6%) |
| | Other (24%) |
| | — |
| Hypokinetic-Spastic (7%) | Degenerative CNS disease (30%) |
| | PSP (20%) |
| | Vascular (20%) |
| | Multiple (15%) |
| | Other (15%) |
| Ataxic-Flaccid-Spastic (6%) | ALS (59%)* |
| | Vascular (18%) |
| | Other (23%) |
| Hyperkinetic-Hypokinetic (3%) | Parkinson's disease (67%) [†] |
| | Other (33%) |
| Other Types (19%) | — |

ALS, Amyotrophic lateral sclerosis; CNS, central nervous system; PSP, progressive supranuclear palsy.

*The ataxic component was equivocal in about half of these cases.

[†]Often associated with on-off medication effects.

majority (88%) of these cases had ALS. This suggests that *gradual onset and progression of a mixed flaccid-spastic dysarthria should generate a high index of suspicion about ALS*. The association of ALS with mixed flaccid-spastic dysarthria represents the strongest association of any mixed dysarthria with a specific neurologic disease in the sample.

Mixed ataxic-spastic dysarthria accounted for 23% of the mixed dysarthrias. Neurologic diagnoses were quite variable, with a majority of cases associated with vascular, demyelinating (usually MS), degenerative, and inflammatory etiologies.

Hypokinetic-spastic dysarthria accounted for 7% of the cases. Again, neurologic diagnoses were quite variable, although approximately 50% were associated with PSP or undefined degenerative CNS diseases.

Mixed ataxic-flaccid-spastic dysarthria accounted for 6% of the mixed dysarthrias. Of interest, 59% of these cases had a neurologic diagnosis of ALS. This supports the clinical impression that *ataxic-like speech features may be perceived in individuals with ALS, particularly when their dysarthria is mild*. This is discussed further when the specific speech characteristics of ALS are addressed.

Mixed hyperkinetic-hypokinetic dysarthria accounted for 3% of the cases. Approximately two thirds of these cases had a diagnosis of PD. The mixed dysarthrias probably reflected on-off medication effects.

Many other mixed dysarthrias were encountered. In fact, a total of 29 different combinations of single dysarthria types were documented. Other than the mixed types just discussed, however, none of the other mixed types occurred frequently.

Severity and Other Characteristics

This retrospective review did not permit a precise delineation of dysarthria severity. However, intelligibility was specifically commented on in 68% of the first 300 cases encountered within the sample summarized in Box 10-1. In those cases, 76% had *reduced intelligibility*. The degree to which this figure accurately estimates intelligibility impairments in mixed dysarthrias is unclear. It is likely that many patients for whom an observation of intelligibility was not made had normal intelligibility. However, the sample probably contains a larger number of mildly impaired patients than is encountered in a typical rehabilitation setting.

Because of its association with damage to more than one portion of the nervous system, it is reasonable to expect that some people with mixed dysarthrias will have cognitive disturbances. For the patients whose cognitive abilities were explicitly judged or formally assessed in the sample summa-

rized in Box 10-1 (83% of the sample). 26% exhibited some impairment of cognitive ability. The proportion of patients with cognitive impairments was much higher among those whose etiology was not ALS.

Finally, among the first 300 patients summarized in Box 10-1, dysarthria was the initial symptom in 20% and among the initial symptoms of neurologic disease in 25%. Perhaps more important, for 12% of that sample, dysarthria (sometimes with accompanying dysphagia) was the only complaint and neurologic finding at the time the patient presented for neurology and speech pathology diagnosis.

Motor Neuron Disease—Amyotrophic Lateral Sclerosis

Dysarthria may be the first manifestation of ALS, and it usually develops at some point during the disease's course. When dysarthria and dysphagia are the initial symptoms of ALS, they tend to remain the most functionally limiting symptoms as the disease progresses.¹⁷³ It has been estimated that approximately half of people with ALS who are receiving hospice care have reduced intelligibility, and only approximately 25% are intelligible just before death.¹³⁵ People requiring augmentative communicative devices need them within an average of 3 years following diagnosis and use them for an average of 2 years.¹³⁸ Once speech is affected, its decline is inevitable but not necessarily steady.³³ It is important to recognize that dysarthria in ALS may not be perceived as mixed at all points during the disease. It may present as either flaccid or spastic dysarthria; when mixed, either type may predominate.

Oral mechanism abnormalities are typically bilateral and generally consistent with those encountered in people with flaccid or spastic dysarthria of any etiology. Thus if spasticity is present, a jaw jerk, sucking reflex, hyperactive gag reflex, slow orofacial movements, and pseudobulbar affect may be evident. If LMNs are affected, the gag reflex may be reduced, the cough weak, and the face lacking in tone. Lingual fasciculations and atrophy can be prominent and early signs. Fasciculations may also be apparent in the chin and perioral area. Dysphagia may be present on UMN or LMN bases. It is not unusual for ALS patients with flaccid-spastic dysarthria to have an *audible reflexive dry swallow*. Some patients complain of shortness of breath, especially when lying down, and pulmonary function studies may demonstrate reduced vital capacity.

Nonspeech oral mechanism abnormalities are clearly relevant to speech findings. Clinical measures of strength and speed of tongue and lip movements, and other indices of respiratory and oromotor structure and function during nonspeech activities (e.g.,

lingual atrophy, dysphagia, velar movement, vital capacity) correlate strongly with measures of speech function, including intelligibility.^{25,169}

Darley, Aronson, and Brown (DAB)^{28,29} studied 30 people with ALS and found a combination of the deficits that were present in their groups with flaccid dysarthria alone and spastic dysarthria alone. The primary speech dimensions and clusters of deviant speech dimensions for these ALS patients are summarized in Table 10-4. It is apparent that some features are clearly associated with spastic or flaccid dysarthria and that others can be attributed to either type. The six clusters of deviant dimensions that were identified match with clusters found in spastic

Table 10-4 The clusters and most deviant speech dimensions, ranked from most to least severe, associated with the mixed flaccid-spastic dysarthria of ALS, as well as the degree to which the flaccid versus spastic component probably contributes to each feature.

| <u>Dimension/Cluster</u> | <u>Component</u> |
|--------------------------------------|-------------------|
| Dimension | |
| Imprecise consonants | Either or both |
| Hypernasality | Flaccid > spastic |
| Harshness | Spastic > flaccid |
| Slow rate | Spastic |
| Monopitch | Either or both |
| Short phrases | Either or both |
| Distorted vowels | Spastic |
| Low pitch | Spastic |
| Monoloudness | Spastic > flaccid |
| Excess and equal stress | Spastic |
| Prolonged intervals* | Combined |
| Reduced stress | Spastic |
| Prolonged phonemes* | Combined |
| Strained-strangled quality | Spastic |
| Breathiness | Flaccid > spastic |
| Audible inspiration | Flaccid |
| Inappropriate silences* | Combined |
| Nasal emission | Flaccid |
| Clusters | |
| Prosodic excess | Spastic |
| Prosodic insufficiency | Spastic |
| Articulatory-resonatory incompetence | Spastic |
| Phonatory stenosis | Spastic |
| Phonatory incompetence | Flaccid |
| Resonatory incompetence | Flaccid |

Data from Darley FL, Aronson AE, Brown JR: Clusters of deviant speech dimensions in the dysarthrias, *J Speech Hear Res* 12:462, 1969a and Darley FL, Aronson AE, Brown JR: Differential diagnostic patterns of dysarthria, *J Speech Hear Res* 12:246, 1969b.

*Not a prominent dimension in either flaccid or spastic dysarthria. May represent the combined effects of both dysarthria types.

and flaccid dysarthria and provide further support to the types of dysarthria that are prominent in the disorder.

In addition, three features were present that were not found in flaccid or spastic dysarthria alone: *prolonged intervals*, *prolonged phonemes*, and *inappropriate silences*. These mainly prosodic features may reflect a summation of flaccid and spastic influences on speech. The combined effects of UMN and LMN deficits on speech are also reflected in the finding that distorted vowels, slow rate, short phrases, and imprecise consonants were rated as more severe in the ALS group than in any other group studied by DAB.

Phonatory abnormalities are frequently present but are quite variable across speakers. Perceptual attributes of dysphonia that receive frequent mention include harshness, breathiness, tremor, strained-strangled quality, audible inhalation, and abnormally high or low pitch.²⁵ Even highly intelligible speakers have a relatively high frequency of voicing contrast errors, suggesting vulnerability of the laryngeal subsystem early in the disease course.¹²⁸ The specific phonatory (and other) acoustic attributes may not be uniform across ALS speakers, however,¹⁴³ possibly reflecting varying degrees to which spasticity and weakness are present. It also appears that gender may be related to certain patterns of phonetic contrast errors^{71,128}; for example, errors related to laryngeal functions are more frequent in men than in women.⁷¹

The frequent presence of tremor noted by Carrow et al.²⁵ is curious, because tremor is not a finding in flaccid or spastic dysarthria associated with other diseases. However, Aronson⁴ observed the presence of a *rapid tremor* or "*flutter*" in some people with ALS, a feature detectable during vowel prolongation but usually not during connected speech; perceptually it seems to fall in the 7 to 10 Hz range. Demodulation and spectral analysis of the vowel prolongations of ALS patients with perceived flutter has documented frequency and amplitude modulations ranging from 0 to 25 Hz, with most patients having amplitude or frequency peaks in the 6 to 12 Hz range.⁵ The physiologic cause of the vocal flutter associated with ALS is uncertain, but it is generally thought to reflect an LMN deficit rather than a "central" tremor.*

The vocal harshness perceived in mixed flaccid-spastic dysarthria often has a "*wet*" or "*gurgly*"

character. This is presumably due to turbulence during speech from saliva that has accumulated in the pyriform sinuses and on the vocal folds because of reduced frequency of swallowing or inadequate clearing of secretions.

Occasionally, when the dysarthria is mild, a patient may exhibit irregular articulatory breakdowns during contextual speech, leading to a perception of *ataxic* or *ataxic-like dysarthria*. The reasons for this are unclear, but they may be similar to those offered in Chapter 9 for the ataxic-like characteristics that may be perceived in unilateral upper motor neuron dysarthria.

Several studies shed light on the articulatory abnormalities that contribute most to reduced intelligibility in ALS.^{72,75,76,80} Among patients with varying degrees of intelligibility impairment, the most disturbed features tended to be related to velopharyngeal function (nasal-oral distinctions), lingual functions for articulatory manner contrasts (stop versus affricate), syllable shape, voicing contrasts, regulation of tongue height for vowels, and production of syllable final consonants. Data suggest that features affecting intelligibility tend to be consistent within speakers over time but may differ among speakers.⁸⁰ The findings demonstrate that all speech functions are not affected uniformly and, specifically, that some lingual functions are affected less than others are. For example, front versus back vowel, long versus short vowel, and general place of articulation distinctions seem relatively resistant to intelligibility problems.* Finally, it is noteworthy that perceptual ratings of speech alternate motion rate (AMR) articulatory precision and rhythmic consistency correlate strongly with ratings of sentence intelligibility.¹³²

The physiologic and acoustic characteristics of speech in ALS have received some attention. Findings have confirmed or modified perceptual hypotheses and extended our understanding of the disorder. The primary findings of these studies are summarized in Table 10-5.

Kinematic studies have identified increased nasal airflow, difficulty maintaining velar elevation for sequences requiring velopharyngeal closure, slow single and repetitive articulatory movements, reduced velocity of articulator movement, limited range of movement, and reduced maximum strength of voluntary jaw, lip, and tongue movement.^{31,32,34,36,61,85} Kinematic findings of slow rate

*The only other dysarthria type in which flutter is perceived is hypokinetic. Whether the flutter heard in the mixed dysarthria associated with ALS and the hypokinetic dysarthria associated with parkinsonism share the same acoustic characteristics and pathophysiology is uncertain.

*Everyday listeners, commenting on their attempts to understand the speech of speakers with ALS, identify articulatory imprecision, slow rate, monopitch, distorted vowels, and difficulty distinguishing word boundaries as contributors to reduced intelligibility.⁷⁹

Table 10-5 Summary of acoustic and physiologic findings in studies of ALS*

| <u>Speech Component</u> | <u>Acoustic or Physiologic Observation</u> |
|------------------------------------|--|
| Respiratory | Reduced vital capacity Chest wall muscle weakness |
| Laryngeal | Abnormal f_0 (too high or low) Abnormal jitter, shimmer, harmonic/noise ratio Decreased maximum phonatory frequency range Decreased maximum vowel prolongation |
| Velopharyngeal | Difficulty maintaining velar elevation Increased nasal airflow |
| Articulation, Rate, Prosody | Slow single & repetitive articulatory movements Reduced velocity & range of articulatory movements Reduced maximum strength of tongue, lip, & jaw movements Excessive jaw movement (probably compensatory) Lengthened segment and sentence duration Increased stop-gap duration Blurring of voiced-voiceless VOT distinctions (articulatory-laryngeal) Reduced spectral distinctiveness among lingual fricatives Increased vowel duration within syllables Reduced/shallow/flattened F2 slope within words Reduced vowel space Exaggerated formant trajectories at vowel onset within syllables Frequency or amplitude fluctuations, or both, during vowel prolongation related to perceived vocal flutter |

ALS, Amyotrophic lateral sclerosis; f_0 , fundamental frequency; F2, second formant; VOT, voice onset time.

*Note that many of these findings are based on only a few speakers and that not all speakers with ALS (or mixed spastic-flaccid dysarthria) exhibit all features. Note also that many of these characteristics are probably not unique to ALS or mixed spastic-flaccid dysarthria; some may be found in other motor speech disorders or other neurologic or nonneurologic conditions.

have been confirmed by acoustic studies that document abnormally slow segment and sentence duration.^{153,159,162,163} In general, physiologic findings suggest that *tongue functions are more severely affected than those of the lips and jaw.*⁷⁴ Taken together, the movement abnormalities identified in physiologic studies are consistent with the slow rate, imprecise articulation, vowel distortions, hypernasality, and nasal emission commonly perceived in speakers with ALS.

People with ALS may complain of shortness of breath when supine, and they can have reduced vital capacity in the upright position. Studies of respiration have documented chest wall muscle weakness, especially in inspiratory muscles. Low lung volumes can reduce utterance length and loudness, and respiratory weakness can lead to reduced loudness and stress contrasts, short phrases, and reduced power for coughing.¹²⁵ Respiratory decline can be expected in all people with ALS, and, unfortunately, severe compromise of bulbar functions is associated with severe compromise in respiratory status, an association that exacerbates dysarthria and dysphagia.¹⁷⁰

The relationship between weakness and speech disability in ALS is neither simple nor direct, at least partly because of the ability of some muscle groups

to compensate for weakness in others. For example, kinematic studies have documented excessive jaw displacements during speech-related lip and tongue movements⁶¹ and exacerbation of speech deficits when the jaw is fixed, suggesting that under normal speaking conditions the jaw is able to compensate for weakness in other articulators, because its strength is relatively preserved.³⁴ The complex relationship may also be related to the fact that patients with ALS can have significantly reduced muscle force before obvious effects on speech, probably because only approximately 10% of maximum muscle contraction forces are recruited during speech.⁸⁵

Acoustic studies have documented a number of abnormalities, although with substantial variability across patients.⁷⁴ Among the common findings are: abnormal fundamental frequency (f_0) (too high or low); abnormal jitter, shimmer, and harmonic/noise ratio; reduced maximum phonatory frequency range; longer stop-gap durations*; longer vowel duration in syllables; decreased maximum vowel duration;

*Stop-gap duration is the time from cessation of acoustic energy in a preceding vowel to the onset of acoustic energy from the articulatory burst for a subsequent initial stop-plosive.

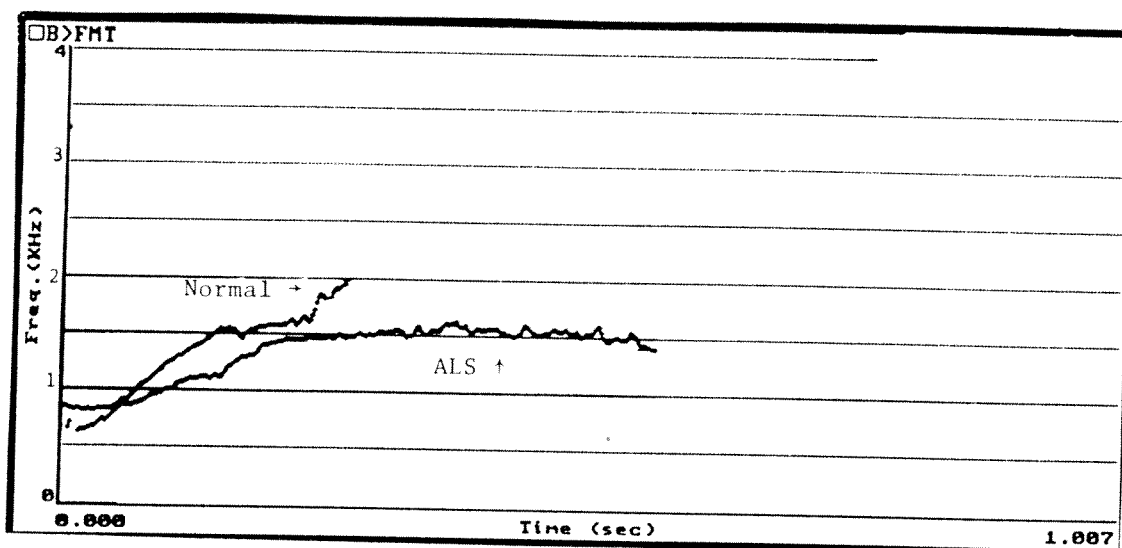


FIGURE 10-2 Second formant (F2) tracings for the vowel /æ/ in the word “wax” for a normal male speaker and a man with mixed spastic-flaccid dysarthria associated with ALS (analysis based on method described by Kent et al.⁷⁷). Relative to the normal speaker, the F2 slope for the dysarthric speaker is only approximately half as steep, covers a smaller frequency range, and takes approximately twice the time to complete. This long and flattened F2 trajectory is an acoustic correlate of slow speaking rate and slowed and restricted range of articulatory movements that can underlie mixed spastic-flaccid dysarthria.

abnormal rate and periodicity of vocal fold diadochokinesis (i.e., rapid repetitions of /hA/); slow and short phrase duration; reduced spectral distinctiveness between lingual fricatives; and longer segment and utterance durations.* Some studies document a blurring of the voice onset time (VOT) distinctions between initial voiced and voiceless stops,¹³⁷ but others do not,²⁶ and not all studies find consistent abnormalities in jitter, shimmer, and harmonic/noise ratio.^{72,75} This variability in findings probably reflects differences in severity across various speech subsystems and perhaps the degree to which weakness versus spasticity is predominant among the speakers who have been studied. Gender differences have also been noted for some measures.⁷² In general, however, these acoustic findings are indicative of slow lingual or laryngeal movements, aperiodicity, or instability of movements and weakness of movements during speech.

The acoustic characteristics of the vocal flutter that is present in some patients have been examined acoustically using fast Fourier transformation (FFT) after the signal from vowel prolongations with perceived flutter was demodulated into frequency and amplitude components.⁵ Results demonstrated mul-

tle frequency and amplitude modulations, with more prominent modulations in ALS than control subjects. The prominent frequencies spanned the range from 0 to 25 Hz, but most had peaks in the 6 to 12 Hz range. These findings provide support for the perception of flutter in some ALS patients, but they do not clarify the basis for the phenomenon. Aronson et al.⁵ speculated that the flutter is probably not central, because tremor is not typically heard in spastic dysarthria alone, the only obvious CNS dysarthria present in ALS. They noted that people with peripheral neuropathy could have tremor in the 8 to 12 Hz range. In addition, the flutter could be a sign of loss of motor units, resulting in an intermittent absence of motor unit firing that, when it affects intrinsic laryngeal muscles, might be perceived as a tremor or flutter.

Studies of the slope of the second formant (F2) in intelligibility test words have revealed reliable and useful findings. For people with ALS, the *F2 slope* seems to be a sensitive index of lingual function, and perhaps speech proficiency in general, because it probably reflects the rate at which lingual movements occur and, by inference, the rate at which motor units can be recruited. F2 slope declines along with intelligibility in subjects followed longitudinally and in groups of men and women with a range of intelligibility impairments^{71,75,77,112,162} (see Figure 10-2). Weismer et al.¹⁶⁴ found shallower slopes of formant transitions, exaggerations of formant trajectories at the onset of vocalic nuclei, and greater inter-

*References 26,71,72,75,126,127,129,153,154,160,162.

⁷They also observed tremulous movements of the true folds and supraglottic muscles on fiberoptic examination of ALS patients with vocal flutter.

speaker variability in ALS speakers than in control subjects. Patients who were less than 70% intelligible had more aberrant trajectory characteristics than those with better intelligibility. Poorly intelligible speakers tended to have flat trajectories or shallow slopes. It thus appears that measures of formant transitions, particularly F2, may be a useful index for monitoring the course of ALS and for making predictions about intelligibility impairments. It should be noted, however, that the F2 slope-intelligibility relationship may not be linearly correlated across the full range of intelligibility,^{77,112} so measures other than F2 may be required to predict the full range of intelligibility scores in people with ALS.*

Vowel space⁷ appears to be another useful acoustic correlate of impairment. In comparison to normal speakers, vowel space is reduced in some speakers with ALS at habitual, slow, and fast rates, and it is moderately correlated with speech intelligibility.^{159,162,163}

Is the typically slow speaking rate of speakers with ALS a primary problem or does it reflect a compensatory response to maintain intelligibility? Although both explanations could be true within or across speakers, it has been shown acoustically and perceptually that although ALS speakers can increase their rate when asked, they nonetheless remain slower than normal. Importantly, vowel space is compressed at habitual and faster rates, and perceptual measures of intelligibility and severity does not show any change between habitual and faster rates. This latter finding suggests that the habitually slow speech rate in speakers with ALS is not a product of compensation.¹⁶³

To summarize the primary speech findings commonly associated with ALS, the dysarthria associated with the disease may be *flaccid, spastic*, or, most often, *mixed flaccid-spastic*. The overall pattern of the mixed form, beyond mild degrees of impairment, is one of *labored, slowly produced speech with short phrases and intervals between words and phrases, grossly defective articulation, hypernasality, strained-strangled and groaning voice quality, and monopitch and monoloudness*. Although all levels of speech production are often affected in ALS, the degree of impairment across levels is not uniform, at least relative to their impact on intelligibility.

*Yorkston et al.¹⁷³ present data that suggest that reductions in speaking and AMR rates in people with ALS may be precursors of reduced intelligibility.

⁷The vowel space is the area of the quadrilateral formed by plotting F1 against F2 for the point vowels [i], [u], [a], and [æ].⁷³ Reduced vowel space implies reduced acoustic and perceptual distinctiveness among the plotted vowels and reduced distinctiveness among the articulatory movements that generate them.

Multiple Sclerosis

Dysarthria is the most common communication disorder associated with MS, occurring in 40% to 50% of people with the disease.^{12,53,107} Severity varies but is generally related to the overall severity of neurologic deficit, including physical and cognitive deficits, and to the number of neurologic systems involved. On average, communication deficits are mild, but a small percentage of affected people have moderate to severe difficulties, with some requiring augmentative or alternative means of communication.^{12,30,171} The type of dysarthria is also variable, consistent with the variable presentations of MS in general. *Ataxic and spastic dysarthria, often combined, are probably most common.*

Nonspeech findings in MS that have implications for speech production include the occasional presence of reduced vital capacity and inadequate ventilation.^{30,63} Respiratory complications are frequent in the terminal stages of MS and may also occur during disease relapses. Such impairments can include generalized or diaphragmatic respiratory muscle weakness, disordered regulation of automatic and voluntary breathing, and bulbar weakness leading to aspiration and infection. Some patients have obstructive sleep apnea, and some require mechanical respiratory support.⁶³ Facial paralysis similar to Bell's palsy occurs in approximately 10% of people with MS, and facial myokymia and trigeminal neuralgia may also be present.¹³⁹ Although tremor in speech system muscles is not generally present in MS, tremor elsewhere in the body occurs frequently (especially in the upper extremity), and its severity correlates with dysarthria severity.³ In general, it appears that lingual functions are more severely affected than lip functions, and abnormalities in lingual strength, endurance, and rate of repetitive movements have been demonstrated, even in nondysarthric MS speakers.^{52,115}

Table 10-6 summarizes the deviant speech characteristics and some of the related dysfunctions found in Darley, Aronson, and Goldstein's³⁰ study of 168 people with MS. The presence of impaired loudness and pitch control and sudden articulatory breakdowns are suggestive of ataxic dysarthria, but the dysarthria was spastic in some patients. The presence of spastic dysarthria in MS is also suggested by the findings of Farmakides and Boone,⁵⁷ who reported hypernasality, reduced pitch variability, and slow rate in some of their 82 people with MS.

It should be noted that *scanning speech*, as it may occur in some speakers with ataxic dysarthria,* is not

*See Chapter 6 for a discussion of scanning speech. The explanation provided for scanning speech by Hartelius et al.⁵⁴ was based on data obtained from speakers with MS.

Table 10-6 Speech deviations and related functions in a sample of 168 people with multiple sclerosis

| <u>Deviation</u> | <u>% of Sample</u> |
|----------------------------------|--------------------|
| Speech | |
| Impaired loudness control | 77 |
| Harshness | 72 |
| Defective articulation | 46 |
| Impaired emphasis | 39 |
| Impaired pitch control | 37 |
| Hypernasality | 24 |
| Inappropriate pitch level | 24 |
| Breathiness | 22 |
| Sudden articulatory breakdowns | 9 |
| Related Functions | |
| Decreased vital capacity | 35 |
| Nasal escape (on oral manometer) | 2 |
| Inadequate ventilation | 2 |

Based on Darley FL, Aronson AE, Goldstein NP: Dysarthria in multiple sclerosis. *J Speech Hear Res* 15:229, 1972.

a pathognomonic feature of dysarthria in MS. For example, only a small percentage of patients studied by Darley, Aronson, and Goldstein³⁰ had increased stress on unstressed syllables, the feature that is most relevant to a perception of scanning speech. This should temper descriptions of scanning speech as *the* speech of MS.

Instrumental measures of dysarthria in MS are quite limited, but they do support the ability of acoustic and kinematic measures to identify abnormalities at the phonatory and articulatory levels. For example, it has been established that measures of long-term phonatory instability during vowel prolongations can distinguish MS speakers with perceptible dysphonia from matched control speakers,⁵¹ and that temporal measures of speech AMRs and sequential motion rates (SMRs) are sensitive to distinctions between MS speakers and normal speakers and speakers with PD.¹⁵⁵ Electropalatography has also detected temporal abnormalities (articulatory "overshooting") in tongue movements during speech in one speaker with MS.¹¹³

It seems reasonable to conclude that *ataxic* and *spastic dysarthria* and *mixed ataxic-spastic dysarthria* are among the most frequent dysarthria types encountered in MS. Perhaps more than in any other of the degenerative diseases discussed here, however, the dysarthrias of MS are unpredictable. It is prudent to consider virtually any dysarthria type or combination of types as possible in people with the disease.

Friedreich's Ataxia

The few studies of speech in FA establish that, despite the disease's label, its associated dysarthria is not always ataxic and that the dysarthria can be mixed. This is a logical consequence of the disease's capacity to affect more than cerebellar structures.

The prominent deviant speech dimensions that have been noted in perceptual studies of FA include abnormal respiratory synchrony, harshness, breathiness, strained-strangled voice quality, audible inspiration, monopitch, pitch breaks, fluctuating pitch, inappropriate pitch level, monoloudness, excess loudness variation, hypernasality, imprecise consonants, distorted vowels, irregular articulatory breakdowns, prolonged phonemes, abnormal rate, excess and equal stress, inappropriate silences, prolonged intervals, slow rate, and slow AMRs.^{43,70} Acoustic analysis has quantified abnormal f_0 and intensity variability in vowel prolongation, abnormal variability in AMRs, slow speaking rate, and longer word durations and slower AMRs.^{1,42,87} Taken together, these observations suggest that more than a single dysarthria type can be present in FA. For example, the statistical analysis by Joannette and Dudley⁷⁰ of 22 patients with FA identified a cluster of features suggestive of underlying ataxia with predominant effects on articulation, as well as phonatory stenosis, which could reflect a spastic component (although the authors did not explicitly conclude that the phonatory abnormalities reflected spasticity). The presence of breathiness and audible inspiration raises the possibility of a flaccid component as well. These data plus clinical experience suggest that ataxic dysarthria is not *the* dysarthria of FA.

To summarize, the results of a few perceptual and acoustic studies, combined with the known sites of nervous system degeneration in people with FA, suggest that *ataxic dysarthria* may be the most frequently encountered dysarthria in FA, but that other types can also be present, particularly *spastic dysarthria*. Thus mixed dysarthria can be present in FA, and it seems that the most common mix is an *ataxic-spastic dysarthria*.

Progressive Supranuclear Palsy

Dysarthria is probably the least well described clinical sign of PSP, in spite of it being a frequent, early (often within 2 years), and prominent manifestation of the disease.^{88,95,96,108,111,123,140} It is more frequently among the initial manifestations of disease in PSP than PD and more prevalent overall in PSP than PD⁶⁹; dysarthria is present in 70% to 100% of unselected patients with PSP in several reports. Given the

predilection of PSP to produce parkinsonian, pseudobulbar, and sometimes ataxic features, it is reasonable to predict several types of dysarthria in PSP.

Oral mechanism examination can reveal various confirmatory signs encountered in people with hypokinetic, spastic, and ataxic dysarthria. Although orofacial manifestations of parkinsonism (e.g., facial masking) are most frequent, evidence of pseudobulbar palsy (e.g., pseudobulbar affect, hyperactive jaw jerk) are common. In addition, in contrast to the flexed neck posture often seen in PD, people with PSP may exhibit neck extension, with the head pointed upward.⁸⁸ Dysphagia is common,* with some studies suggesting it is more common than dysarthria,⁹⁵ and others suggesting it is less common.⁸³ Latency from disease onset to complaints of dysphagia is strongly correlated to total survival time.¹¹¹

The dysarthrias of PSP have been delineated in several group studies.^{82,83,96,108,140} Hypokinetic, spastic, and ataxic types (generally in that order of frequency) are consistently identified, sometimes singly, but more often in various combinations. A combination of all three types is common in some reports. The severity of the hypokinetic component is related to the degree of neuronal loss in the substantia nigra.⁸² In general, the combination of spastic, hypokinetic, and ataxic components coincides with the loci of neuropathologic changes found in PSP, and their recognition is considered important to clinical diagnosis.⁸³

Because PSP is often misdiagnosed as PD,⁶⁸ some attention to speech findings that seem to distinguish PSP from PD is warranted. Retrospective data suggest that characteristics found more frequently in PSP than PD include monopitch, hoarseness, nasal emission, excess and equal stress, hypernasality, imprecise articulation, and slow rate. Characteristics found more frequently in PD than PSP include vocal flutter, reduced loudness, reduced stress, tremor, breathiness, and rapid rate (Lu, Duffy, and Maraganore, 1992). These differences are logically related to the relative exclusivity of features of hypokinetic dysarthria in PD and the frequent added presence of features of other dysarthria types in PSP, particularly spastic dysarthria. These distinctions suggest that *the presence of a dysarthria type other*

than hypokinetic in people with a neurologic diagnosis of PD should raise questions about the accuracy of the PD diagnosis; PSP would be an alternative diagnosis.

Speech difficulties that extend beyond those that can be explained by dysarthria can be present. *Palilalia* is frequently noted,* and “stuttering” dysfluencies and *echolalia* are mentioned in some reports (Testa et al., 2001)^{83,86,88,108}; recall, however, that palilalia and certain dysfluencies have a strong association with hypokinetic dysarthria (see Chapter 7). Some patients produce *involuntary vocalizations* such as groaning or humming sounds,¹⁵² and some patients have language and cognitive deficits commonly associated with frontal lobe pathology.^{38,123} A single atypical case with apraxia of speech has been reported.¹⁶

To summarize, perceptual observations establish that dysarthria, often mixed dysarthria, is common and tends to appear early in PSP. *Hypokinetic, spastic, and ataxic* dysarthria are most commonly present, most often in varying combinations. Recognition of a mixed dysarthria in people with suspected PSP versus PD may be particularly helpful to differential diagnosis. Early in the course of neurologic disease, the presence of hypokinetic dysarthria or, especially, a mixed dysarthria with hypokinetic, spastic, or ataxic components, may be more strongly associated with PSP than with other degenerative neurologic diseases, particularly PD.

Multiple System Atrophy

The dysarthrias associated with MSA and its subtypes—MSA-P and MSA-C—have been sufficiently described to develop a picture of their prevalence, severity, and salient features. This can be supplemented by descriptions of the dysarthrias associated with striatonigral degeneration, OPCA, and Shy-Drager syndrome, conditions now encompassed by the MSA designation.

Dysarthria is common in MSA,[†] present in 100% of unselected patients in some series.⁸¹ It tends to emerge earlier in the disease course than it does in PD, within the first 2 years in approximately half of affected people. On average, dysarthria seems to be more severe than in PD.^{38,81,111,165}

Dysarthria type is usually correlated with other motor signs of MSA and is therefore, often mixed.

*Kluin et al.⁸³ reported that 73% of their patients with PSP had dysphagia. Litvan, Sastry, and Sonies⁹⁵ reported that 96% of their PSP patients had abnormal swallowing studies, although only 19% aspirated. A majority of their patients had delayed initiation of the swallow reflex, impaired tongue mobility, premature dripping into the pharynx, and pooling in the valleculae.

*Palilalia has been reported as an early-appearing clinical feature that helps distinguish patients with PSP from those with MSA.¹⁴⁵

†In spite of involvement of multiple systems, apraxia of speech and aphasia are rarely, if ever, encountered in MSA.³⁸

Hypokinetic, ataxic, and spastic types (generally in that order of frequency of occurrence) are most often noted. All three types were present in a majority of 46 unselected patients reported by Kluin et al.⁸¹ Recognizing dysarthria types other than hypokinetic can help distinguish MSA from PD.

Before it was absorbed under the heading of MSA, the dysarthrias associated with striatonigral degeneration (MSA-P) were not well described. Hypokinetic dysarthria is the most common expected type, but hyperkinetic and perhaps spastic dysarthria are possible based on the common loci of pathology.

The dysarthrias associated with OPCA have been well described in only a few patients.^{43,55} The reported deviant speech features are suggestive of mixed ataxic-spastic dysarthria, perhaps with a flaccid component based on observations in some patients of audible inspiration and vocal flutter.* Because OPCA (MSA-C) may also be associated with parkinsonian features, a hypokinetic dysarthria is also possible. Thus *ataxic, spastic, hypokinetic*, and, less frequently, *flaccid dysarthria*, singly or in combination, are the common expected dysarthria types. Additional but less consistently present deficits that can affect speech and communication include palatal myoclonus and dementia.³⁵

The dysarthrias associated with Shy-Drager syndrome are most adequately described in Linebaugh's⁹³ study of 80 people with the disorder. Forty-four percent had dysarthria. Among those with dysarthria, 43% had ataxic dysarthria, 31% had hypokinetic dysarthria, and 26% had various combinations of mixed dysarthrias. Three forms of mixed dysarthria were present, including *hypokinetic-ataxic, ataxic-spastic*, and *spastic-ataxic-hypokinetic*. These mixes are consistent with the involvement of direct and indirect motor systems and the basal ganglia and cerebellar control circuits that occurs in the disease.

An association of laryngeal stridor with Shy-Drager syndrome has long been recognized, and as many as one third of people with MSA may have stridor.¹⁹ Recognizing stridor within various combinations of spastic, ataxic, and hypokinetic dysarthria is diagnostically valuable because that combination of signs is probably uncommon in degenerative diseases other than MSA. Excessive snoring and sleep apnea are commonly associated with inhalatory stridor, but stridor can sometimes be heard just before speech is initiated or at phrase boundaries during ongoing speech; when more serious, it can be

*Hartman and O'Neill⁵⁵ described a man with OPCA whose speech characteristics suggested a mixed flaccid-spastic dysarthria plus stuttering-like dysfluencies, which may or may not have reflected a reemergence of developmental stuttering.

evident during quiet awake breathing. When severe upper airway obstruction occurs, continuous positive airway pressure or tracheostomy may be recommended. The cause of inhalatory stridor is traditionally thought to reflect abductor (posterior cricoarytenoid) laryngeal weakness secondary to involvement of the nucleus ambiguus,⁶ hence its frequent recognition as a sign of flaccid dysarthria.* However, recent evidence suggests that laryngeal dystonia may at least sometimes be the cause of stridor, at least as it occurs in MSA.^{47,65}

Corticobasal Degeneration

Communication deficits are common in CBD, and dysarthria is among the most frequent communication problems. For example, among 60 papers that have described speech and language characteristics in a total of 457 people with CBD, dysarthria was reported as present in 42%.⁹⁰ Dysarthria and other communication disorders can be early and prominent manifestations of CBD, and the prevalence of dysarthria increases with disease progression.^{9,39,90,166} Severity of dysarthria is related to overall disease severity¹²⁰ but is not necessarily correlated with disease duration.³⁹

Dysarthria type varies, but more often than not it is mixed. *Hypokinetic and spastic types are most common*, but ataxic dysarthria has also been reported, most often in combination with hypokinetic or spastic types, or both.^{39,90,120} *Apraxia of speech* may also be present, either as the sole MSD or in combination with dysarthria. Nonverbal oral apraxia is frequently reported, and echolalia and palilalia have been noted.^{39,90}

It is important to note that aphasia occurs frequently in CBD, in more than half of patients in some reports, and that aphasia type is most often described as nonfluent or anomic. Aphasia can be the first manifestation of the disease; when this is the case, it is frequently called *primary progressive aphasia*.^{41,64,90,106,109}

Patients with CBD may exhibit a behavior that has been called *yes-no reversals*, in which they spontaneously complain that they say or gesture "yes" when they mean "no," and vice versa, when responding to questions during social discourse; the behavior is often confirmed during examination.⁴⁰ This can occur in the absence of obvious aphasia, but it does occur more frequently in patients with predominant

*Loss of myelinated nerve fibers in the laryngeal branch of the recurrent laryngeal nerve has been documented in patients with MSA.⁵⁶

⁷It is known that dystonia can be present in cervical and limb muscles in people with MSA.¹⁵

left hemisphere involvement. It is correlated with frontal lobe functions related to mental flexibility, inhibitory control, and motor programming. Yes-no reversals in the absence of significant aphasia can be a useful differential diagnostic sign, because among people with degenerative neurologic diseases, they most often occur in CBD and PSP.⁴⁰

To summarize, mixed or isolated dysarthria types are common in CBD, with hypokinetic, spastic, and ataxic types being the most common. When the common asymmetry of the disease involves the left hemisphere, communication deficits are frequently mixed beyond dysarthrias, with frequent occurrence of aphasia and apraxia of speech. Communication can also be affected by additional problems that probably reflect frontal lobe dysfunction (e.g., yes/no reversals, reduced mental flexibility, impaired motor programming). This constellation of deficits can make the communication difficulties encountered in CBD more complex than in many other degenerative neurologic diseases.

Wilson's Disease

Wilson's disease (WD) often affects the bulbar muscles. Dysarthria is one of the most frequent and sometimes the only manifestation of the disorder.^{141,156} Oral mechanism findings in people with WD can be similar to those encountered in people with hypokinetic, ataxic, or spastic dysarthria. Dystonia also may be present and is considered responsible for the inappropriate and fixed vacuous or "pseudo smile" exhibited by some patients.¹⁴¹ Dysphagia and drooling are not unusual.

Berry et al.¹⁰ studied 20 patients with WD who had various combinations of ataxia, rigidity, and spasticity. The most prominent deviant speech characteristics and clusters of speech characteristics (based on factor analysis) derived from the study are summarized in Table 10-7. It was concluded that the dysarthria of WD can be mixed, containing various combinations of *hypokinetic*, *ataxic*, and *spastic*, but that each single type can occur alone in some people with the disease.

Monitoring the speech of patients undergoing penicillamine and low copper diet management of their WD has established a correlation between improvement in deviant speech characteristics and general neurologic improvement.¹¹ This suggests that careful monitoring of speech during medical treatment of WD can serve as an index of the effectiveness of treatment for the disease.

Traumatic Brain Injury

Communication deficits are common in TBI. They can include nonaphasic cognitive-communication

table

10-7

Prominent deviant characteristics and clusters of speech characteristics associated with Wilson's disease

| Features | Hypokinetic | Ataxic | Spastic |
|--|-------------|--------|---------|
| Characteristics | | | |
| Reduced stress | X | | X |
| Slow rate | | X | X |
| Excess and equal stress | | X | X |
| Low pitch | X | | X |
| Irregular articulatory breakdowns | | X | |
| Hypernasality | | | X |
| Inappropriate silences | X | | |
| Prolonged phonemes | | X | |
| Prolonged intervals | | X | |
| Strained voice | | | X |
| Short phrases | | | X |
| Clusters (Based on Factor Analysis) | | | |
| Prosodic insufficiency | X | | X |
| Phonatory stenosis | | | X |
| Prosodic excess | | X | |
| Articulatory-resonatory incompetence | X | | X |

Based on Berry WR et al: Dysarthria in Wilson's disease, *J Speech Hear Res* 17:169, 1974a. Only those characteristics that are *not* common to all three dysarthria types are listed (see the original study for a listing of all deviant characteristics).

disorders, aphasia, and MSDs. Our focus here is on the dysarthrias, which occur in approximately one-third of the TBI population overall,¹³⁴ with approximately 60% having dysarthria early after onset and approximately 10% chronically.¹⁷⁴ Yorkston et al.¹⁷ point out that the dysarthria varies significantly in severity and persistence, may or may not be accompanied by language and other cognitive disorders and has a more positive outcome in people younger than the age of 20. They also note that although a major portion of recovery tends to occur within the first several months, significant changes in speech can occur over many months or years.

Virtually any type of dysarthria can result from TBI, and mixed dysarthria is probably more common than any isolated dysarthria type. This is a logical consequence of the diffuse or multifocal injuries that are so often associated with the condition, and it is congruent with motor deficits that occur elsewhere in the body, including weakness, spasticity, ataxia, bradykinesia, rigidity, tremor, and dystonia. The locus of CNS lesions is diverse, but it appears that a majority of lesions in people with dysarthria from TBI are subcortical and that the dysarthria is less severe when lesions are predominantly cortical.⁸⁹ I

table 10-8 Summary of acoustic and physiologic findings in studies of dysarthric speakers with TBI*

| <u>Speech Component</u> | <u>Acoustic or Physiologic Observation</u> |
|------------------------------------|---|
| Respiratory | Reduced vital capacity & forced expiratory volumes Difficulty coordinating rib cage & abdominal movements |
| Laryngeal | Increased f_0 Abnormal vocal fold closing time & phonatory flow rate Decreased rate of adduction/abduction Increased or decreased subglottal pressure or laryngeal airway resistance Increased jitter, shimmer, amplitude perturbation, voice turbulence, & noise/harmonic ratio Continuous voicing during connected speech |
| Velopharyngeal | Increased nasal airflow & nasalance |
| Articulation, Rate, Prosody | Decreased strength, endurance, speed, & control of tongue & lip movements Increased articulatory effort during lip movements Syllable lengthening Reduced syllable & overall speech rate Slow AMR rates, with lengthened syllables & intersyllabic gaps Temporal & energy irregularities during AMRs Abnormal variability in VOT Multiple or missing stop bursts Spirantization |

AMR, Alternate motion rate; f_0 , fundamental frequency; TBI, traumatic brain injury; VOT, voice onset time.

*Note that many of these findings are based on only a few speakers and that not all speakers with TBI exhibit these features. Note also that most, if not all, of these characteristics are not unique to dysarthria in TBI; many may be found in other motor speech disorders or other neurologic or nonneurologic conditions.

is noteworthy that injury is not always confined to the CNS; approximately one third of people with severe TBI can have cranial nerve deficits,⁶⁹ sometimes with associated flaccid dysarthria. As with many dysarthria types, all levels of the speech system can be affected but not necessarily to the same degree. Sometimes impairment is evident at only a single level.

Many reports of dysarthria associated with TBI in both children and adults* fail to describe a dysarthria type, but those that do most often document flaccid, spastic, ataxic, hypokinetic, and hyperkinetic types.^{22,23,161,172} The reported mixed dysarthrias include spastic-ataxic, flaccid-spastic, flaccid-ataxic, spastic-hypokinetic, hypokinetic-ataxic, ataxic-hyperkinetic (palatal-laryngeal myoclonus), and spastic-hyperkinetic (dystonia and palatal-laryngeal myoclonus). More than two components are sometimes present. Also, because of the complexity of the motor impairments that can occur with TBI, it is not

unusual for a dysarthria type to be described as "undetermined."

A number of physiologic and acoustic studies have confirmed, refined, or modified perceptual findings and inferences about underlying deficits. The findings of these studies are quite variable across speakers within and among studies, at least partly secondary to differences in dysarthria type and subsystem impairments. Most studies have focused on adults, but similar abnormalities have also been demonstrated in children.¹⁵⁰ These findings are summarized in Table 10-8.

At the respiratory level, lower vital capacity, lower forced respiratory volumes, and problems coordinating rib cage and abdominal movements during speech have been documented.^{116,149} Such deficits may be related to a tendency to breathe at ungrammatical locations in some dysarthric speakers.⁵⁰

Phonatory dysfunctions are common in TBI^{101,147} but not homogeneous. Among patients with various perceived phonatory abnormalities, electrolaryngographic and aerodynamic assessments have demonstrated increased f_0 , abnormalities in vocal fold closing time and phonatory flow rate, decreased adduction/abduction rate, and increased or decreased subglottal pressure or laryngeal airway resistance.^{104,147-149} Acoustic abnormalities indicative of phonatory problems most often include abnormal values for jitter and shimmer, increased amplitude

*Cahill, Murdoch, and Theodoros (2003) studied 24 children between 5 and 18 years of age with TBI. Sixteen were dysarthric. Various dysarthria types were evident, and severity ranged from mild to severe. The authors concluded that the profile of speech deficits in children with TBI mirrors that of their adult counterparts. Thirty percent of the children with TBI reported by Stierwalt et al.¹⁴⁵ were dysarthric, similar to the incidence reported for adults.

perturbation, voice turbulence, and noise-to-harmonics ratio.^{66,101} Several of these physiologic and acoustic abnormalities are suggestive of laryngeal hyperfunction (e.g., strained voice quality) and are consistent with spasticity, but others suggest laryngeal hypofunction (e.g., breathiness) and, possibly, weakness.* Theodoros and Murdoch¹⁴⁸ noted that some intersubject differences on their instrumental measures could reflect compensatory adjustments for laryngeal spasticity or strategies to compensate for problems elsewhere in the system. These appropriate cautions about data interpretation highlight the importance of recognizing that abnormalities heard in dysarthric speakers, as well as abnormalities detected aerodynamically and kinematically, can reflect underlying pathophysiology, as well as, possibly, compensatory responses to the pathophysiology.

Problems at the velopharyngeal level have been documented with aerodynamic measures of nasal airflow, and, in general, they correlate with the perception of hypernasality.^{102,103,149,151} For clinicians who rely heavily on perceptual ratings, it is important to note that a perception of hypernasality sometimes can be an artifact of slow speech rate; that is, when hypernasality is perceived in the absence of instrumental findings of increased nasalance, rate of speech tends to be slow.¹⁰³

A number of instrumental studies document abnormalities at the articulatory level. Results vary among affected speakers, but a variety of kinematic and acoustic measures have identified decreased strength, endurance, speed or control of tongue or lip

movements, increased effort to achieve normal articulatory (lip) pressures, and syllable lengthening and reduced syllable and overall speech rates.^{24,44,45,47,67,114,142,149} In general, tongue movements appear more severely affected than lip or jaw movements.⁶⁷ Findings generally suggest that dysarthria in TBI is often associated with reduced speed, force, and endurance of movements, and these abnormalities generally correlate with the perception of slow rate and increased sound, syllable, and word durations. However, physiologic data do not always correlate with all perceptual ratings. Although Stierwalt et al.¹⁴² found that measures of tongue strength and endurance were significantly correlated with perceptual judgments of articulatory imprecision and overall speech defectiveness, other studies have failed to find a strong relationship between perceptual and kinematic measures.^{46,105} Goozée, Murdoch, and Theodoros⁴⁶ suggested that their dysarthric speakers might have been compensating in different ways for their physiologic impairments, thus weakening the ability of physiologic and perceived articulatory abnormalities to predict each other.

Speech AMRs, analyzed acoustically, can distinguish TBI speakers with dysarthria from normal control speakers.¹⁴ Wang et al.¹⁶¹ found that TBI speakers had slowed syllable AMR rates that were due to lengthened syllables and, to a lesser extent, lengthened intersyllable gaps. AMR syllable rates were correlated with conversational speech rates, overall dysarthria severity, intelligibility, and prosody. There were also irregularities in temporal and energy parameters within repetition sequences and abnormal variability in VOT. Qualitative analyses revealed evidence of explosive speech quality, breathiness, phonatory instability, multiple or missing stop bursts, continuous voicing, and spirantization. The detection of a large number of motor abnormalities on the basis of AMR data alone may be particularly valuable, because AMRs are probably less susceptible than many other speech tasks to the contaminating influences of the often-present and significant cognitive and linguistic deficits in the TBI population.

*It appears that laryngeal abnormalities are less pronounced or occur less frequently in children than adults with TBI. Cahill et al.²³ studied 16 speakers with TBI who were younger than 16 years old at the time of injury. They had normal or only minimally impaired laryngeal function compared to that reported for adults after TBI. The authors noted that the reasons for this are uncertain but could include different dynamics of TBI in children versus adults, better potential for recovery in children, or better ability in children to compensate for impairments because the pediatric larynx is still developing.

Cases

Case 10-1

A 68-year-old man presented stating: "I don't know what's the matter with me. If you have a cure, I'd be delighted." During the previous 4 years he had developed impotence, occasional stumbling and falling, dysphagia with aspiration of liquids, and occasional laryngeal stridor. He had recently developed urinary urgency and clumsiness in his hand.

Neurologic examination revealed axial rigidity, poor station, orthostatic hypotension, reduced upward gaze, and dysarthria. EMG revealed a mild, predominantly motor peripheral neuropathy. Autonomic reflex testing identified a generalized autonomic neuropathy. Laryngeal examination revealed left vocal fold paresis.

During speech examination, he noted a 1-year history of a "higher and weaker" voice and a sense that his speech was "clumsy." He reported choking on liquids and having occasional "laryngospasms" during sleep. He was no longer able to play the trumpet or flute because of respiratory fatigue; he stated, "I get out of breath for no good reason." Finally, he complained that his lips were "tight and being stretched across my mouth."

Examination revealed a slight left lower facial droop, equivocal bilateral reduction in tongue strength, a weak cough and glottal coup, and inhalatory laryngeal stridor at phrase boundaries during speech and when inhaling rapidly. His speech was characterized by accelerated rate (1), monopitch and monoloudness (3), imprecise articulation (1), and reduced loudness (1). Pitch was mildly elevated. Vowel prolongation was strained-harsh (1) and unsteady (1,2). Vocal flutter was sometimes evident.

Speech AMRs were irregular (1,2) and occasionally accelerated and "blurred."

The clinician concluded: "mixed dysarthria in which a hypokinetic component is most prominent. His mildly irregular AMRs and vocal unsteadiness suggest an ataxic component. The subtle strained component to his voice could represent a mild spastic component, although there are no other features of spasticity. His laryngeal stridor suggests posterior cricoarytenoid weakness, and his vocal flutter may reflect weakness of laryngeal adductors."

Pulmonary function tests were abnormal but nonspecific. MRI of the head showed moderate cerebellar atrophy and periventricular atrophy.

The neurologist concluded that the patient had MSA that most closely corresponded to Shy-Drager syndrome. Several drugs whose action would stimulate dopamine receptors were recommended. The patient declined speech therapy. He was told that therapy might help maintain intelligibility or could help develop augmentative means of communication if it became necessary.

Commentary. (1) Mixed dysarthria occurs commonly in degenerative neurologic disease. (2) A number of dysarthria types may be perceptually evident in mixed dysarthria. This patient had unequivocal hypokinetic and flaccid dysarthria, probable ataxic dysarthria, and possible spastic dysarthria. All of these types were compatible with the diagnosis of MSA or Shy-Drager syndrome. (3) Many people with obvious dysarthria decline speech therapy when intelligibility and speech efficiency are relatively well maintained.

Case 10-2

A 35-year-old woman with a 10-year history of chronic progressive MS presented for consideration of thalamotomy to control a severe bilateral upper limb tremor. Neurologic examination revealed hyperreflexia; pathologic reflexes; bilateral weakness; spasticity; impaired coordination; nystagmus and optic neuritis; and severe resting, postural, and movement tremor of the upper and lower extremities. Neuropsychological assessment demonstrated severe impairment of new learning and memory and a generalized loss of intellectual abilities.

During speech evaluation, the patient noted a 1-year history of progressive speech difficulty. She had reduced facial and lingual strength. Speech was characterized by slow rate (3), irregular articulatory breakdowns (2), breathy-hoarse voice quality (2), and hypernasality with nasal emission (2). Speech intelligibility was significantly reduced.

The clinician concluded that the patient had a "mixed spastic-ataxic dysarthria of moderate severity."

Unfortunately, the presence of abnormal somatosensory evoked potentials precluded adequate localization within the thalamus for lesion placement to abolish her tremor. Surgery was not recommended. She was not motivated to pursue speech therapy.

Commentary. (1) Mixed dysarthria is not uncommon in people with MS who are dysarthric. Mixed spastic-ataxic dysarthria may be the most common mixed dysarthria encountered in MS. (2) Cognitive deficits may be present in MS, and they can compound difficulties with communication. (3) In spite of reduced intelligibility, not all patients are motivated or interested in speech therapy.

Case 10-3

A 49-year-old woman was referred by her internist because of a 2-month history of speech difficulty that her family interpreted as a response to psychologic stress. During speech evaluation, she admitted to considerable family stress but felt she was handling it well. Her difficulty began with a cold. She described its initial character as "nasal." She had also developed swallowing difficulty characterized by food sticking in her throat after a swallow had been initiated and the need to swallow several more times to get it down. She had recently begun to choke on liquids. She admitted that food occasionally squirreled in her cheeks and that sometimes she needed to use a finger to remove it. She had begun to gag when brushing her teeth or swallowing saliva and reported "crying a lot" even when she did not feel sad. She admitted to some "twitching" around her eyes and left upper lip.

Oral mechanism examination revealed bilateral lower face and tongue weakness and reduced lateral tongue AMRs. Nasal emission was evident during pressure consonant production. Her gag reflex was hyperactive, but her cough and glottal coup were weak. A sucking reflex was present.

Contextual speech was characterized by groaning and strained voice quality (1), reduced loudness (-2), hypernasality (2), imprecise and weak pressure consonants (2,3), reduced rate (2), and short phrases and monopitch and monoloudness (2,3). Speech AMRs were slow but regular (2,3). Vowel prolongation was mildly strained and breathy.

The clinician concluded that the patient had a "mixed flaccid-spastic dysarthria of moderate severity." She was referred for neurologic evaluation. She declined a recommendation for speech therapy because her primary concern at the time was diagnosis.

Neurologic examination showed evidence of hyperreflexia and pathologic reflexes in all limbs and weak-

ness in her face. EMG examination failed to provide evidence for LMN disease in the limbs. A computed tomography (CT) scan of the head was normal.

Her speech continued to worsen. Two months later she was writing to communicate much of the time. She had moderate bilateral lower facial weakness, equivocal jaw weakness, markedly reduced tongue strength, and possible lingual atrophy. The gag reflex was hyperactive, and cough and glottal coup were markedly weak. She had an audible reflexive swallow and inhalatory stridor. Connected speech was characterized by strained-hoarseness (2), reduced loudness (2), hypernasality (2), and imprecise articulation (3). Rate was slow (2,3), and phrases were short, with monopitch and monoloudness (3). Stridor was present at phrase boundaries. Vowel prolongation was strained-harsh-wet. Speech AMRs were slow (3). She had pseudobulbar crying.

Speech therapy was recommended. Speech intelligibility improved for approximately 1 month but then deteriorated. EMG 1 month later demonstrated abnormalities in all limbs, consistent with ALS. The patient communicated fairly efficiently by writing until her death from respiratory and cardiac arrests approximately 6 months later.

Commentary. (1) Dysarthria can be the initial manifestation of neurologic disease and fairly frequently is the presenting sign of ALS. It can progress for some time before diagnosis is confirmed. (2) Initial signs of neurologic disease are sometimes misinterpreted as responses to psychologic stress. When the symptom is speech difficulty, careful examination can help distinguish a motor speech disorder from a psychogenic speech disturbance. (3) Mixed spastic-flaccid dysarthria is the "prototypic" mixed dysarthria of ALS. Its effects on intelligibility can be dramatic and often lead to a need for augmentative/alternative forms of communication. (4) Rate of decline can be quite rapid in some people with ALS.

Case 10-4

A 77-year-old woman developed difficulty with speech, swallowing, and right leg and left arm weakness. She was subsequently hospitalized for an apparent exacerbation of longstanding myasthenia gravis. Her prior symptoms of myasthenia gravis were predominantly ophthalmic, and the disease had been well controlled with Mestinon. Steroids and an increase in Mestinon dose did not help. Her lack of response to these treatments raised the possibility that myasthenia gravis might not be the only cause of her new difficulties.

During speech evaluation, the patient reported a 3-month history of speech and swallowing problems. She was frequently choking, with occasional nasal regurgitation. She also complained of increased ease of crying, even when she did not feel sad. She did not complain of dramatic worsening of her speech with extended talking.

Examination revealed mild jaw and lower facial weakness. The tongue was weak bilaterally, but fasciculations and atrophy were not evident. Palatal movement during vowel prolongation was minimal. A gag reflex could not be elicited. A sucking reflex was present. She had a prominent audible reflexive swallow. Her speech was characterized by slow rate (3), reduced phrase length, strained-harsh voice quality (3), hypernasality (3) with audible nasal emission on pressure sounds, and monopitch and monoloudness (3). Speech AMRs were slow (3), slower than expected for her degree of weakness. Vowel prolongation was strained-hoarse and occasionally characterized by flutter.

The clinician concluded that the patient had: "mixed spastic-flaccid dysarthria. I believe the spastic compo-

nent predominates and that respiratory weakness reflects the most significant flaccid component. The spastic component and her pseudobulbar affect are suggestive of UMN involvement and cannot be explained on the basis of weakness secondary to myasthenia gravis. On the basis of this examination, it is not possible to determine if the LMN component of her dysarthria is secondary to neuromuscular junction disease or some other disturbance in LMN function. However, there is no significant deterioration of her speech with stress testing."

Based on the speech evaluation, EMG studies were conducted. They failed to show evidence of ALS. A CT scan of the head showed moderate diffuse cerebral and cerebellar atrophy and a small lacunar infarct in the left basal ganglia. The neurologist concluded that the patient's difficulties were probably due to a combination of her myasthenia gravis and pseudobulbar palsy of undetermined origin. However, multiple small infarctions were suspected as the cause of her pseudobulbar palsy.

Commentary. (1) By definition, mixed spastic-flaccid dysarthria identifies the presence of upper and lower motor neuron dysfunction. In this case, the speech diagnosis helped establish that myasthenia gravis could not be the sole explanation for the patient's difficulties. (2) Mixed dysarthrias can result from the cooccurrence of two or more diseases. In this case, the patient had a confirmed diagnosis of myasthenia gravis and, possibly, vascular disease leading to multiple CNS strokes.

Case 10-5

A 55-year-old woman presented with a 9-month history of cervical pain and hoarseness following a motor vehicle accident. Laryngeal examination was normal. She was referred to speech pathology for evaluation of her hoarseness.

During speech evaluation, the patient noted that her dysphonia developed immediately after her motor vehicle accident and that vocal fold polyps were identified and removed by laser 4 months later. Her voice gradually returned to normal over the next few months, but hoarseness then returned, with an occasional "slurry" quality to her speech. She denied swallowing difficulty or problems with emotional expressiveness.

Examination revealed equivocal lingual weakness but bilateral lingual fasciculations. There was significant nasal emission during production of pressure-filled sentences, although the palate was symmetric and mobile. Speech was characterized by hypernasality (1), imprecise articulation (0,1), and hoarse-rough voice quality (1,2) with occasional diplophonia. Vowel prolongation was breathy-hoarse-rough-strained (1). Speech AMRs were normal, except for equivocal slowing on "tuh." There was a subtle vocal "flutter" during vowel prolongation.

The clinician concluded: "I believe the patient has a flaccid dysarthria that includes the cranial nerve X and cranial nerve XII. A component of her dysphonia may

Case 10-5—cont'd

indeed be due to excessive musculoskeletal tension in the laryngeal area, perhaps due to efforts to compensate for laryngeal trauma or weakness. However, findings are very suspicious for cranial nerve X and XII weakness. Neurologic examination is strongly recommended."

On neurologic examination, in addition to her speech and cranial nerve findings, phrenic nerve weakness was suspected, because she complained of shortness of breath when lying supine. On EMG, the phrenic nerve was normal, but mild neurogenic changes in the tongue bilaterally, of indeterminate duration and origin, were noted.

Eight months later the patient returned for follow-up assessment. She had had increased episodes of choking, and it had become "more difficult to form words and letters" when speaking. She complained that her swallow was often audible and that she swallowed more slowly, and that "when I cry my mouth wants to start laughing." Examination revealed bilateral chin fasciculations, lower facial weakness, lingual weakness, fasciculations and atrophy, nasal escape during pressure sound production, and a weak cough and glottal coup. A sucking reflex and subtle "on the verge of crying" facial expression were present. Speech was characterized by slow rate (1,2), excess and equal stress (1,2), hypernasality with nasal

emission (1), vocal "flutter"; strained-harsh voice quality (1), and reduced pitch (2). Vowel prolongation was characterized by flutter and a rough, strained voice quality. Speech AMRs were slow (1). Speech intelligibility was normal.

The clinician concluded: "mixed flaccid-spastic dysarthria, with clear worsening of speech difficulty and the emergence of a spastic component since she was last seen. Strongly suspect mixed bilateral upper and lower motor neuron dysfunction." The patient denied a need for speech therapy, and the clinician concurred. She was advised to seek reevaluation if her speech problems worsened.

Subsequent neurologic evaluation identified the presence of diffuse hyperreflexia and pathologic reflexes and weakness in her upper and lower extremities. EMG showed widespread denervation in three extremities, as well as the tongue, consistent with ALS.

Commentary. (1) Dysphonia may be the first sign of neurologic disease. It can occur simultaneously with or be mistaken for vocal abuse or musculoskeletal tension-related dysphonias. (2) Dysarthria associated with ALS does not always present initially as a mixed dysarthria. (3) When dysarthria is present in ALS, it is usually mixed flaccid-spastic in character eventually.

Case 10-6

A 51-year-old woman presented with a 13-year history of PD with marked fluctuations in her neurologic signs and symptoms during her parkinsonian medication cycle. Neurologic examination revealed dysarthria, right arm dystonia and rigidity, bradykinesia, and left arm and leg tremor.

The patient was seen for speech evaluation 1.5 hours after her last Sinemet dose. Severe limb, torso, and head dyskinesias were present. The oral mechanism was normal in size, strength, and symmetry. Dyskinetic movements of her jaw, face, and tongue were apparent but not prominent during speech. Her speech was characterized by accelerated rate (2,3), reduced loudness (1,2), imprecise articulation (1,2), monopitch and monoloudness (1,2), variable rate (1,2), and occasional inappropriate silences (1). Vowel prolongation was unsteady and intermittently mildly strained. Speech AMRs were irregular (1). Speech intelligibility was mildly reduced.

The clinician concluded that the patient had a "moderately severe mixed hypokinetic-hyperkinetic

dysarthria, with the hypokinetic component predominating." It was recognized that her speech probably fluctuated with Sinemet effects, and the patient was quite certain that it was more difficult to talk when her medication wore off. Speech therapy was undertaken, and the patient was quite successful in slowing her speech rate, with subsequent improvement in intelligibility and quality. With some adjustments in medication dosage and timing, there were fewer fluctuations in her speech and other neurologic signs.

Commentary. (1) A mixed hypokinetic-hyperkinetic dysarthria can occur in PD, reflecting the direct effects of the disease on speech and its interaction with medication effects. (2) Fluctuations in the severity and nature of dysarthria in people with PD can occur, sometimes dramatically, as a result of "on and off" effects associated with fluctuating medication effects. (3) Careful monitoring of speech can be a useful way to monitor medication effects in certain neurologic diseases.

Case 10-7

A 61-year-old woman presented with a 6-year history of progressive coordination difficulty and 18-month history of dysarthria. Clinical neurologic examination confirmed the presence of gait ataxia, upper limb incoordination, slowed and ataxic eye movements, and dysarthria.

During speech evaluation the patient described speaking as a "real effort." She felt she had to speak more slowly to be understood but admitted that she was unable to talk more rapidly. She had no chewing or swallowing complaints and denied drooling or difficulty with emotional control. Oral mechanism examination was normal, except that her cough and glottal coup were poorly coordinated. Speech was characterized by slow rate (2); irregular articulatory breakdowns (2); excess and equal stress (2); abnormal alterations in pitch, loudness, and duration of words and syllables (3); and strained voice quality (1). Vowel prolongation was hoarse and unsteady. Speech AMRs were slow and irregular (2).

The clinician concluded that the patient had a "mixed dysarthria, predominantly ataxic, but with a mild spastic

component." Intelligibility was minimally compromised, and the patient denied a need or desire for speech therapy. She was advised to pursue reassessment if her speech difficulty worsened.

Head CT scan demonstrated cerebellar and pontine atrophy. The neurologist concluded that the patient had OPCA. The patient's mother probably had a similar disease.

Commentary. (1) OPCA (MSA-C) is often associated with a mixed dysarthria, in this case a mixed ataxic-spastic dysarthria with the ataxic component predominating. This mix logically reflects the sites of prominent degeneration in MSA-C, and in this case it served as a confirmatory sign for the neurologic diagnosis. (2) Mixed ataxic-spastic dysarthria is not diagnostic of any particular neurologic disease. As in most cases, the speech diagnosis can contribute to localization and provide support for neurologic diagnosis.

Case 10-8

A 64-year-old woman with von Hippel-Lindau syndrome (defined in Chapter 6) was referred by a geneticist for speech assessment and recommendations. Her speech difficulty began following neurosurgery for removal of multiple cerebellar hemangioblastomas 2 years previously. She had a vocal fold paralysis as a complication of her neurosurgery.

She described her speech as sounding "drunk." She denied difficulty with chewing, swallowing, or saliva control. Examination revealed subtle myoclonic twitches in the right chin and tongue. The tongue was normal in strength and range of motion, and there was no atrophy or fasciculations. Palatal myoclonus was evident at rest and during phonation. Myoclonic movements in the external neck were also apparent. There were no pathologic oral reflexes. Her speech was characterized by: reduced rate (-1.2); brief voice interruptions or near-interruptions on a periodic basis, at a rate of approximately 2 to 4 Hz, consistent with myoclonus; infrequent subtle hypernasality and hyponasality; irregular articulatory breakdowns (1); and inhalatory stridor (2). Vowel prolongation was characterized by myoclonic variability at 2.5 to 3 Hz (measured acoustically). Speech AMRs were mildly irregular. Intelligibility was normal in the quiet one-to-one setting.

The clinician concluded: "mixed ataxic-hyperkinetic dysarthria. The hyperkinetic component is represented by a palatal laryngeal myoclonus. This latter problem is, in all likelihood, what is most bothersome to the patient.

She also has some inhalatory stridor, about which she does not complain, which could reflect the laryngeal myoclonus and/or a residual of her vocal fold paralysis."

The nature of the patient's speech difficulty was reviewed in detail with her, with particular attention paid to having her understand her palatal-laryngeal myoclonus. She was counseled that the myoclonus was not subject to behavioral management. Although Botox injection might have helped to manage palatal-laryngeal myoclonus in isolation, it was not recommended in her case because of the other components of her dysarthria, which were felt to put her at greater than average risk for significant dysphagia. A number of suggestions were made regarding strategies to maximize comprehensibility of speech. Formal therapy was not recommended because she was otherwise compensating well for her dysarthria.

Commentary. (1) Mixed dysarthria sometimes has more than a single cause. In this case, the dysarthria probably reflected the effects of the underlying disease as well as complications arising from the neurosurgery that was done to treat it. (2) Some speech abnormalities can have more than a single cause. The patient's stridor may have been a product of vocal fold weakness, laryngeal myoclonus, or a combination of the two. (3) Patient education is an important component of management, as much to promote understanding of why certain things cannot or should not be done as to promote understanding of what can be done.

Case 10-9

A 45-year-old man presented to his family physician complaining of a several-month history of speech difficulty. A general medical examination was normal, and it was thought that the patient's symptoms reflected anxiety. Two weeks later, he called to report that his speech was getting worse. He was referred for neurologic assessment, which was judged normal, including speech. However, because of his complaint, a speech pathology consultation was requested. Testing for myasthenia gravis was also ordered; the results were negative.

The patient was seen 2 weeks later for speech evaluation. He reported an approximately 5-month history of difficulty articulating words normally. He felt the problem had worsened. Only within the past several weeks had his wife agreed that there was some "thick-ness" in his speech.

An oral mechanism examination was normal. His speech was characterized by nonspecific hoarseness with occasional pitch breaks, equivocal hypernasality, and occasional lingual articulatory imprecision, especially for lingual affricates. Speech AMRs were equivocally slow but regular. Vowel prolongation was rough-hoarse with some vocal flutter. There was a trace of nasal airflow on a mirror held at the nares during repetition of sentences with pressure consonant sounds. During 4.5 minutes of continuous reading, there was no dramatic deterioration of voice or speech.

The clinician concluded that the patient had a subtle dysarthria of undetermined type, although with features suggestive of weakness and possible spasticity. Because the patient felt that his dysarthria was often at its worst later in the day, he was asked to call the clinician at home in the evening if he felt that his speech problem was more apparent.

The patient called the clinician several days later in the evening. His speech characteristics were similar to those noted during formal evaluation but worse and strongly suggestive of mixed spastic-flaccid dysarthria. When that observation was communicated to the referring neurologist, additional tests were ordered. Unfortunately, EMG revealed fasciculations and fibrillations in the left upper extremity, left tongue, and bilateral thoracic paraspinal musculature. An MRI was normal. A tentative diagnosis of ALS was made. Subsequent evaluation failed to identify other possible causes for his speech difficulty, and a definite diagnosis of ALS was eventually made. A session of speech therapy established that he would benefit from use of an amplifier in his work as a teacher, primarily to minimize fatigue. Arrangements were made to follow him on an as-needed basis to help manage his communication difficulties.

Commentary. (1) Changes in speech may herald neurologic disease. (2) Subtle changes in speech, in the absence of other symptoms, are fairly frequently misidentified as a reflection of stress or anxiety. (3) Speech changes can be subtle enough to defy a confident, specific speech diagnosis by an experienced clinician, but they may nonetheless be sufficient to warrant a diagnosis of dysarthria and neurologic disease. (4) Accurate recognition of dysarthria type can contribute significantly to a neurologist's decisions about the specifics of a neurologic workup. (5) Early identification of speech deficits in degenerative neurologic disease can establish strategies to maintain intelligible, efficient verbal communication, as well as anticipate and prepare for future communication needs.

SUMMARY

1. Mixed dysarthrias reflect various combinations of individual dysarthria types. They occur more frequently than single dysarthria types, highlighting the fact that dysarthria often reflects damage to more than one component of the speech motor system.
2. Mixed dysarthrias can be caused by many conditions that damage more than one portion of the nervous system, but degenerative diseases are probably their most frequent cause. Single strokes and neoplasms leading to mixed dysarthrias tend to occur in the posterior fossa. Mixed dysarthrias resulting from toxic-meta-

bolic conditions, infection, multiple strokes, and trauma may be the product of diffuse or multifocal damage in many portions of the nervous system.

3. Because a number of diseases are associated with damage to specific parts of the nervous system, the types of mixed dysarthrias encountered in them are somewhat predictable. This is best exemplified by the mixed spastic-flaccid dysarthria that is classically associated with ALS. It should be noted, however, that although most mixed dysarthrias help identify the locus of their causative underlying lesions, they do not, by themselves, usually indicate their specific etiology.

4. Spastic dysarthria is probably the most frequently occurring type of dysarthria encountered within mixed dysarthrias. Flaccid and ataxic dysarthrias also occur frequently. Hypokinetic, hyperkinetic, and unilateral UMN dysarthrias are also encountered in mixed dysarthrias but less frequently than the other dysarthria types.
5. Intelligibility is often affected in mixed dysarthrias. Patients with mixed dysarthrias, excluding those with ALS, frequently also have associated cognitive deficits.
6. Even though mixed dysarthrias reflect damage to more than one component of the motor system, they are fairly frequently the presenting complaints or among the earliest manifestations of neurologic disease. Thus accurate recognition of the components of mixed dysarthrias can aid the localization and diagnosis of neurologic disease and may contribute to the medical and behavioral management of affected individuals.

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