SECTION H Coordination and Gait

CHAPTER 43

Cerebellar Function

The cerebellum is tasked with bringing finesse to the motor system. Although not primarily involved in the mechanisms for production of muscle power, it is necessary for normal control and regulation of muscle contraction. The major function of the cerebellum, from a clinical point of view, is the coordination of movement. The cerebellum is the portion of the brain through which the cerebral motor cortex achieves the synthesis and coordination of individual muscle contractions required for normal voluntary movements. Without it, movements are gross, uncoordinated, clumsy, and tremulous, and precise movements become impossible. Lesions of the cerebellum do not cause weakness, but rather loss of coordination and inability to gauge and regulate, as Gordon Holmes said, the "rate, range, and force" of movement. Although motor strength and power are preserved, active movements are severely compromised.

In order to perform any movement—especially a complex act involving many muscle groups—contractions of the agonists, antagonists, synergists, and muscles of fixation must be adequately coordinated. To begin a movement, the agonists contract to execute the movement; the antagonists relax or modify their tone to facilitate it; the synergists reinforce the movement; and the fixating muscles prevent displacements and maintain the appropriate posture of the limb. To terminate the movement, the antagonists contract and the agonists relax. The individual muscles that enter into the act must be controlled and coordinated, as a conductor would direct an orchestra, precisely regulating the action of the individual parts. The cerebellum is the conductor. It is essential to the synergy of muscle contraction and is the center of coordination for voluntary movement. It does not provide power and does not play an instrument, but without it the symphony of normal movement degenerates into a cacophony of disorganized muscle contractions.

A major manifestation of cerebellar lesions is ataxia (Gr. a "without," taxis

“order”); a rough translation is “not orderly.” The essential feature in ataxia is that movements are not normally organized. Although the term is a general one, indicating chaotic and disorganized movement, it is used clinically primarily to refer to the motor control abnormalities—including incoordination, tremor, and impaired rapid alternating movements (RAMs)—that occur with cerebellar lesions. Ataxia is not specific for cerebellar disease, and lesions in other parts of the nervous system must be excluded before attributing ataxia to cerebellar disease. Impaired proprioception may cause sensory ataxia, and lesions involving pathways that originate in the frontal lobe may cause frontal lobe ataxia. Other common manifestations of cerebellar disease include nystagmus, impaired balance, and difficulty walking.

ANATOMY

The cerebellum is located in the posterior fossa, beneath the tentorium cerebelli. Below and anteriorly, it is separated from the dorsal pons by the fourth ventricle and from the medulla and the dura mater covering the atlanto-occipital membrane by the cisterna magna. Grossly, there are three parts: (a) the cerebellar hemispheres, two larger, lateral masses; (b) the vermis, a small, unpaired median portion that connects the hemispheres (Figures 43.1 and 43.2); and (c) the flocculonodular (FN) lobe, a small, midline structure that lies on the anterior part of the inferior surface. The FN lobe consists of the paired lateral flocculi and the midline nodulus (Figure 43.3). The vermis is separated from the hemispheres by the paramedian sulci. The cerebellar tonsils are small, rounded lobules on the inferior aspect of the cerebellar hemispheres, just above the foramen magnum.

Gross Anatomy

Anatomically, the cerebellum is divided into three lobes: anterior, posterior, and FN. Each has a vermis and hemisphere portion (Figure 43.3). The deep transverse primary fissure divides the cerebellum into anterior and posterior lobes. The posterolateral fissure separates the FN lobe from the posterior lobe. Anatomists further divide the cerebellum by fissures and sulci into 10 lobules that bear arcane names of no clinical relevance (Figure 43.4). In terms of afferent and efferent connections, the cerebellum can also be organized into three parallel, sagittal zones: vermian, paravermian, and lateral. As mentioned

previously, clinicians divide the cerebellum functionally into three parts: (a) the hemispheres, responsible for appendicular coordination; (b) the anterior, superior vermis (or simply the vermis), responsible for gait and other axial functions; and (c) the FN lobe, or vestibulocerebellum. The FN lobe is phylogenetically the oldest and is referred to as the archicerebellum. The FN lobe has extensive connections with the vestibular nuclei and is concerned primarily with eye movements and gross balance. It also receives visual afferents from the superior colliculi and the visual cortex. The primary concerns of the archicerebellum are eye movement control and gross orientation in space, such as up and down. The next area of the cerebellum to evolve was the paleocerebellum or spinocerebellum. In humans, the paleocerebellum consists of the anterior, superior vermis and adjacent paravermal cortex; it corresponds approximately to the anatomical anterior lobe. The paleocerebellum developed during a period of evolution when extremity control was not a concern; it is concerned primarily with posture, muscle tone, axial muscle control, and locomotion. There are extensive connections between the vermis and spinal cord pathways. The most phylogenetically recent part of the cerebellum is the neocerebellum, or the cerebellar hemispheres, which make up the bulk of the cerebellum. The neocerebellum corresponds approximately to the posterior lobe. The hemispheres are concerned with coordinating movement and providing fine motor control for precise movements of the extremities. The primary afferents to the hemispheres are from the pontine nuclei, which receive the corticopontine fibers from the cerebral cortex.

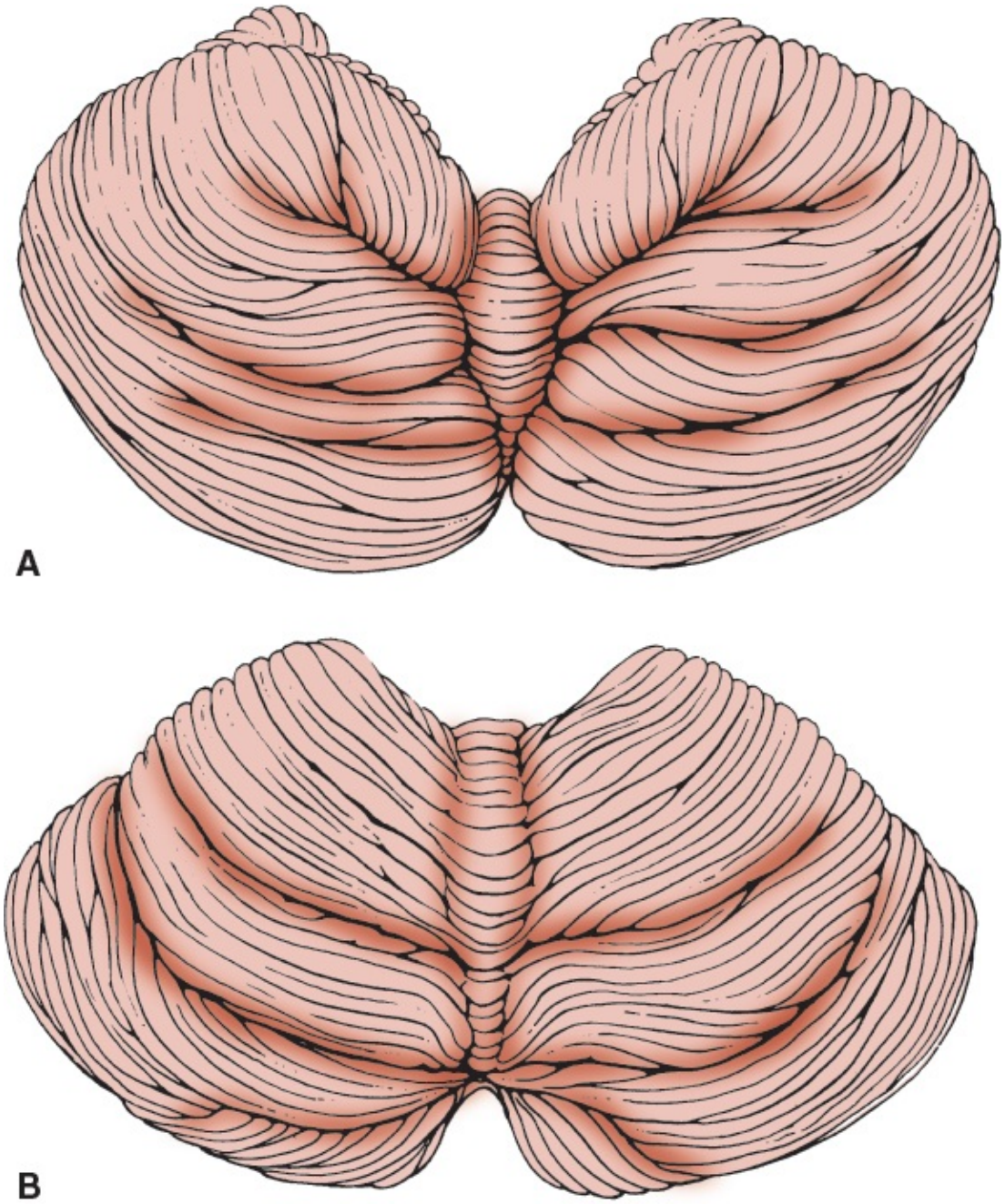

FIGURE 43.1 Ventral **(A)** and dorsal **(B)** views of the human cerebellum. See Figure 43.4 for names of lobes and lobules.

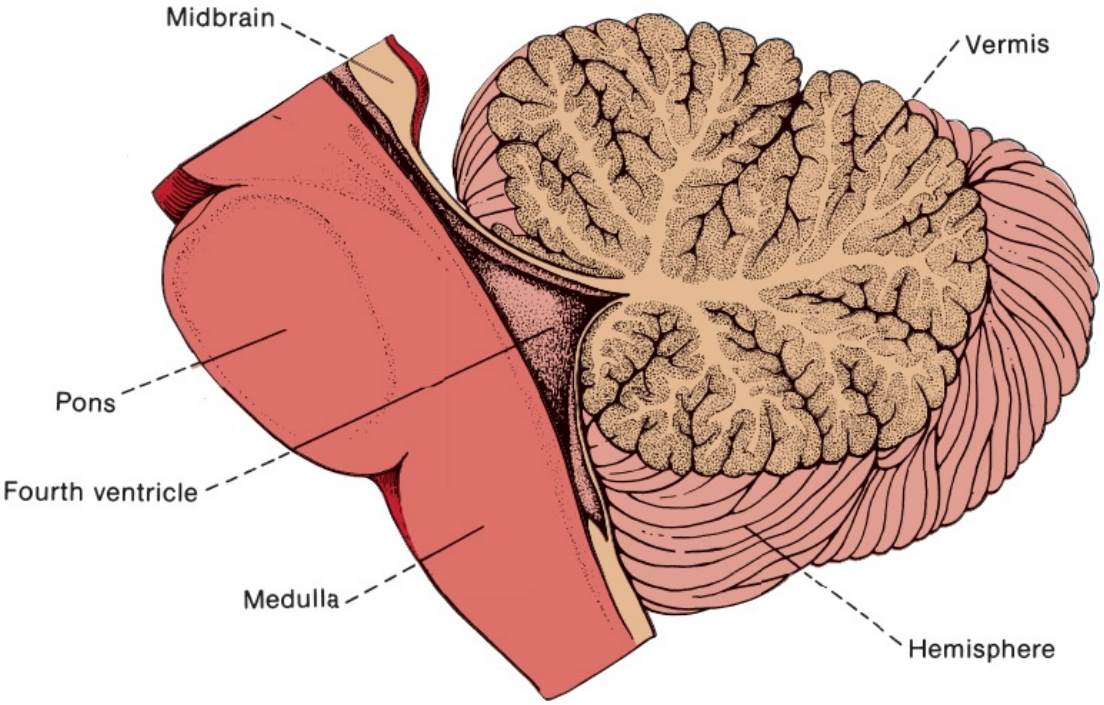

FIGURE 43.2 Median longitudinal section through the human cerebellum, pons, and medulla.

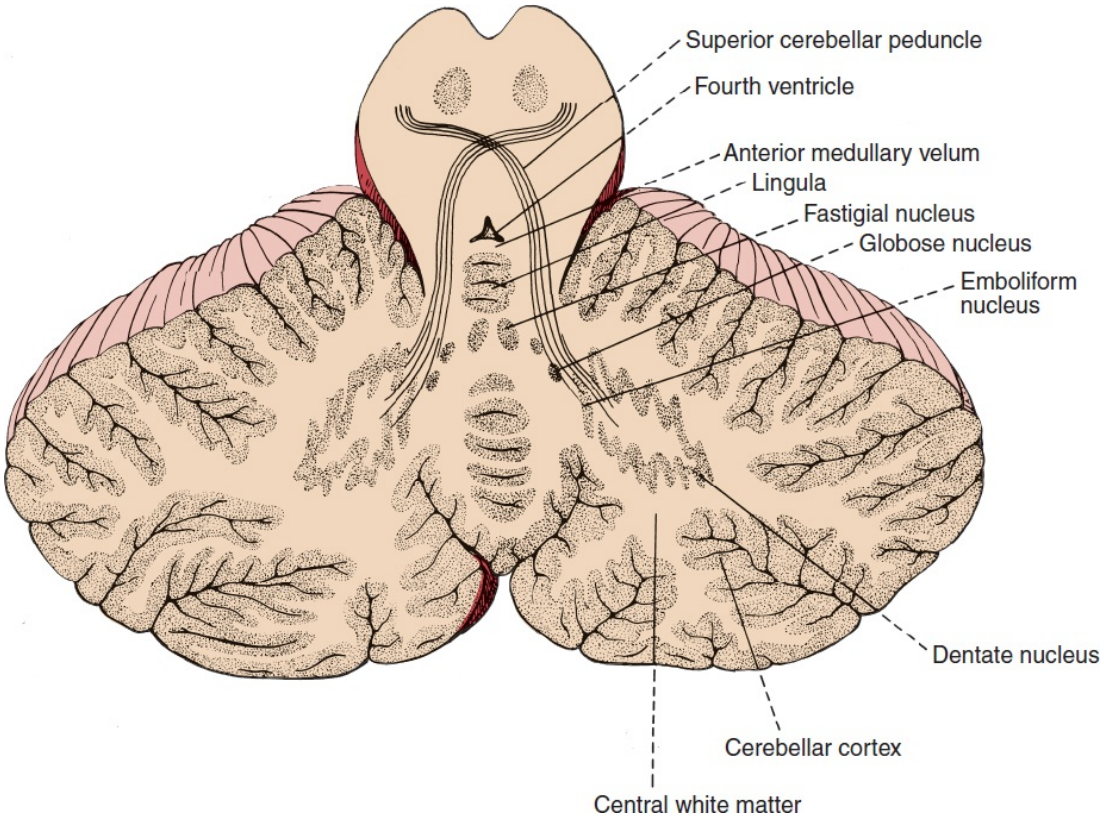

FIGURE 43.3 Horizontal section through the human cerebellum showing the arrangement of the cortical gray matter and locations of the nuclei within the white

matter.

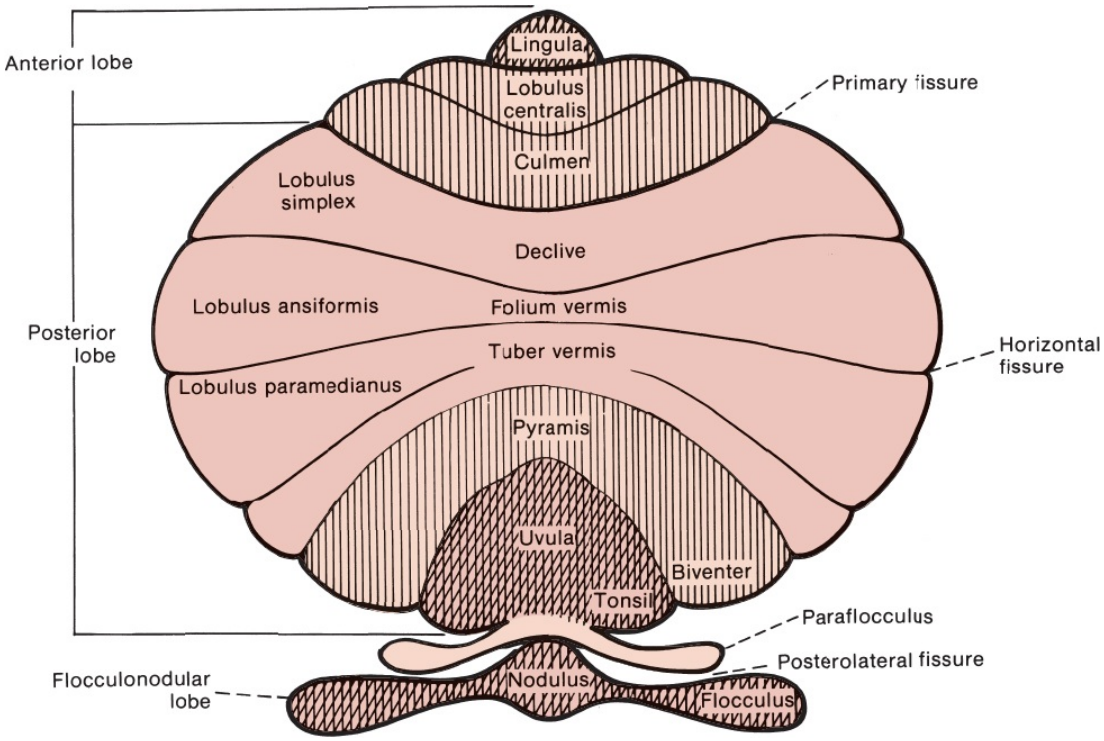

FIGURE 43.4 Diagram of the cerebellum showing the lobes and lobules.

Another way to view the cerebellum is in terms of its primary afferent connections: the vestibulocerebellum (input from vestibular nuclei to the FN lobe), the spinocerebellum (input from the spinocerebellar tracts to the anterior vermis), and the pontocerebellum (input from the pontine nuclei to the hemispheres).

The cerebellum is made up of a white matter core, covered with a thin layer of gray matter, the cerebellar cortex. Deep in the white matter are several gray masses, the cerebellar nuclei. The dentate nuclei, the largest of the cerebellar nuclei, are gray matter structures situated deep in the white matter of each hemisphere (Figure 43.3). In the hilus of each dentate nucleus lie the emboliform nuclei; medial to the emboliform are the globose nuclei. The globose and emboliform nuclei together are called the nucleus interpositus. In the white matter of the vermis, at the roof of the fourth ventricle, are the fastigial, or roof, nuclei. From medial to lateral, the deep nuclei are the fastigial, globose, emboliform, and dentate. The major cerebellar connections are to the vestibular system, the spinal cord, and the cerebral cortex (Figure 43.5). Microscopically, the cortex is made up of three layers: the outer, nuclear, or molecular layer; the

layer of Purkinje cells; and the inner, or granular, layer (Figure 43.6).

The FN lobe is a primitive part of the cerebellum primarily concerned with vestibular function. The connections of the FN lobe are primarily, if not entirely, vestibular. The FN lobe receives afferent impulses from the labyrinths and vestibular centers, spinal cord, and brainstem—including the reticular formation and olivary bodies—and projects to the vestibular nuclei, vestibulospinal tracts, and reticular formation. The cerebellum and vestibular centers function together to maintain equilibrium, the orientation of the body in space, and the regulation of muscle tone and posture. The clinical manifestations of disease of the FN lobe are difficult to separate from the invariably accompanying vestibular findings, primarily nystagmus. Isolated FN lobe dysfunction is usually caused by ependymomas and medulloblastomas in children.

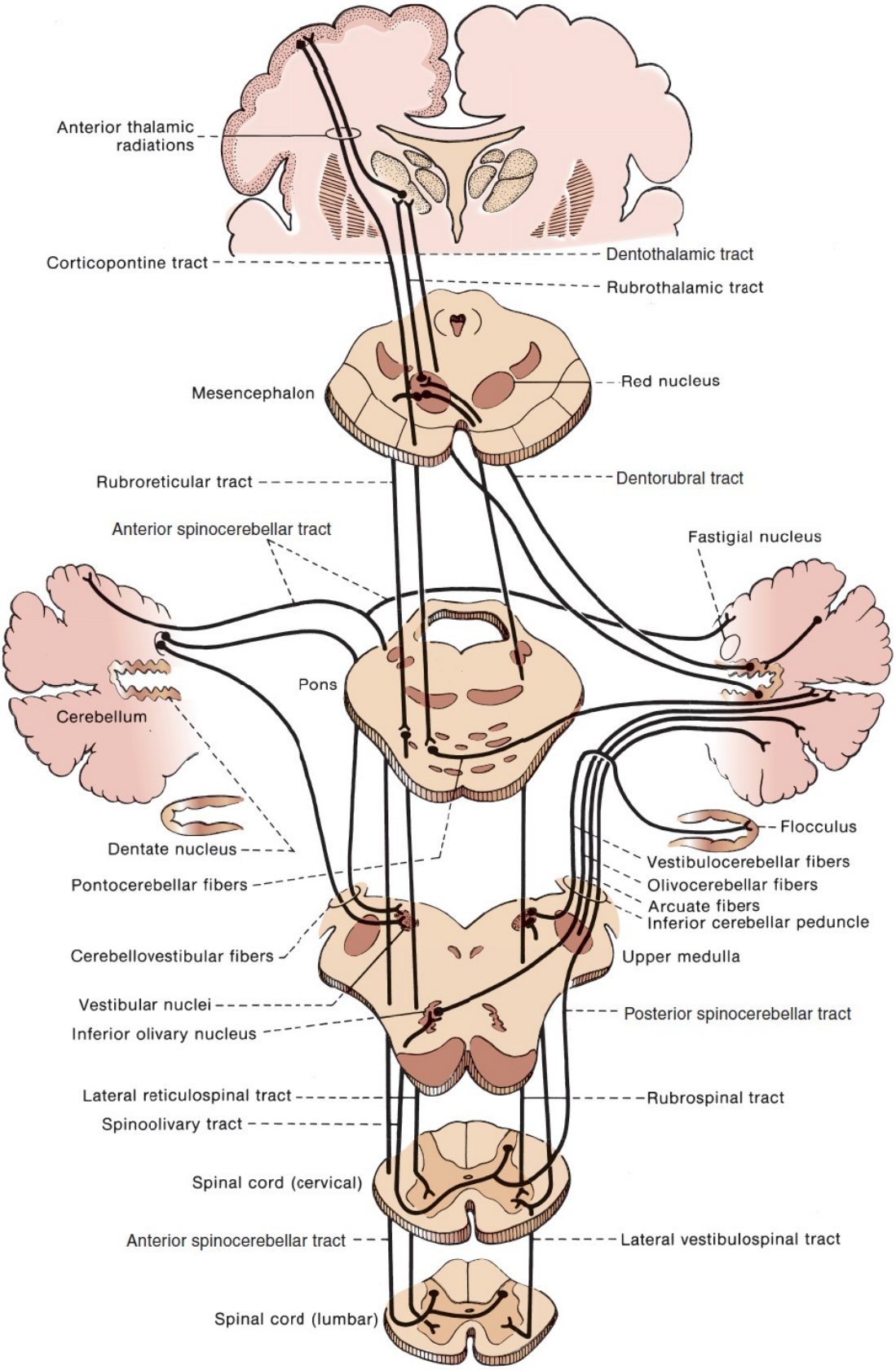

FIGURE 43.5 Principal afferent and efferent connections of the cerebellum.

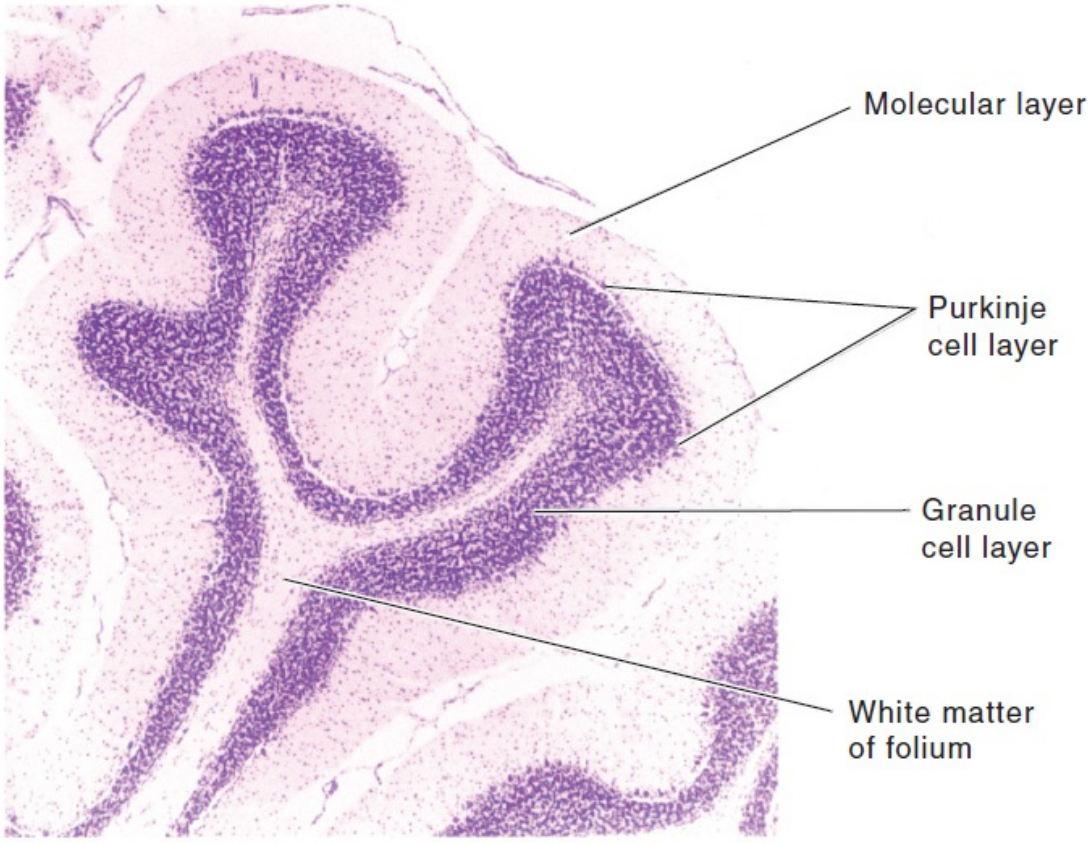

FIGURE 43.6 Transverse section of cerebellar folia showing the three layers of the cortex and the underlying white matter (*cresyl violet*). (Reprinted from Kiernan JA. *Barr's The Human Nervous System: An Anatomical Viewpoint*. 9th ed. Philadelphia: Wolters Kluwer Health/Lippincott Williams & Wilkins, 2009, with permission.)

The paleocerebellum communicates with the spinal cord, brainstem, and vestibular centers. The principal afferent connections of the anterior lobe come from the anterior spinocerebellar tract, although it receives trigeminocerebellar fibers, input from the vestibular nuclei, and some corticocerebellar fibers. The discharges are to the vestibular nuclei, brainstem, and spinal cord.

The neocerebellum (pontocerebellum) communicates with the cerebral cortex. It is enormously developed in mammals in association with growth of the cerebral hemispheres. In primates, the hemispheres overshadow the rest of the cerebellum. Its afferent connections are principally corticopontine, or corticopontocerebellar, although there are some spinocerebellar fibers; it discharges through the dentate nucleus to the red nucleus and thalamus, and thus to the cerebral cortex.

The cerebellum is connected to the brainstem by the three cerebellar peduncles. The inferior cerebellar peduncle (ICP) connects the cerebellum with

the spinal cord and the medulla. The ICP lies medial to the middle cerebellar peduncle (MCP, brachium pontis). The ICP has two parts: the restiform body and the juxtarestiform body. Ascending fibers in the restiform body include the posterior spinocerebellar and cuneocerebellar (from the accessory cuneate nucleus) tracts, the dorsal and ventral external arcuate fibers from the nuclei gracilis and cuneatus, and the olivocerebellar, trigeminocerebellar, and reticulocerebellar pathways. Lying just medial to ICP is the juxtarestiform body, made up of fibers traveling directly between the vestibular nuclei and the FN lobe. The restiform body is an afferent system; the juxtarestiform body contains both vestibulocerebellar and cerebellovestibular fibers. The juxtarestiform body is mainly efferent. Although it carries primary afferent vestibulocerebellar fibers from the vestibular nerve and secondary vestibulocerebellar fibers from the vestibular nuclei, its primary component is cerebellovestibular fibers from the vermis and the FN lobe (fastigiobulbar tract). Other cerebellovestibular fibers run from the fastigial nucleus to the vestibular nuclei in the uncinate fasciculus, which enters the brainstem adjacent to the ICP. The MCP connects the cerebellum with the pons, and through it run the pontocerebellar tracts; these are the final neurons of the corticopontocerebellar pathway that comes mainly from the frontal, temporal, and other areas of the cortex to communicate with the contralateral cerebellar hemisphere.

The superior cerebellar peduncle (SCP, brachium conjunctivum) contains the principal efferent fibers of the cerebellum and the dentatorubral and the dentatothalamic pathways. It also carries the afferent anterior spinocerebellar tract, as well as cerebellovestibular fibers in the uncinate fasciculus. The cerebellotegmental, cerebellotectal, and tectocerebellar tracts also travel in the SCP. The afferent fibers to the cerebellar cortex arrive primarily by tracts that enter through the middle and inferior peduncles, but the anterior spinocerebellar tract enters via the SCP.

Microscopic Anatomy

The molecular layer of the cerebellar cortex contains the dendritic arborizations of the Purkinje cells, radial fibers of the Bergmann glial cells, basket cells, stellate cells, climbing fibers, and parallel fibers (Figure 43.6). The flattened dendritic trees of the Purkinje cells spread out perpendicular to the long axis of the cerebellar folium. Climbing fibers are the terminal ramifications of fibers from the inferior olivary nucleus that ascend through the granular layer to

contact Purkinje dendrites in the molecular layer. Each climbing fiber forms an excitatory synapse with a single Purkinje cell. Climbing fibers also synapse on the neurons of the deep cerebellar nuclei. Parallel fibers are granule cell axons that extend upward into the molecular layer where they bifurcate to send branches in opposite directions along the axis of a folium to terminate on the Purkinje cell dendrites. The parallel fibers intersect the Purkinje cell dendrites like telephone wires over the cross pieces of a telephone pole.

The Purkinje cell layer contains the perikarya of the large Purkinje cells and the smaller Bergmann (epithelial) glial cells. The granule cell layer lies between the white matter and the Purkinje cell layer; it contains granule cells, Golgi cells, brush cells, and the cerebellar glomeruli (Figure 43.7). The granule cells send their axons to the molecular layer where they branch to form parallel fibers. Mossy fibers are the predominant afferent system to the cerebellum. They arise from the spinal cord, the trigeminal, reticular formation, vestibular and pontine nuclei of the brainstem. The mossy fibers terminate as mossy fiber rosettes that lie at the center of each cerebellar glomerulus. Like the climbing fibers, mossy fibers are excitatory. The mossy fibers primarily use glutamate; the climbing fibers release glutamate or aspartate. Cerebellar glomeruli are synaptic formations with mossy fiber rosettes at the center, surrounded by granule and Golgi cell dendrites (Figure 43.8).

The Purkinje cells are excited by the parallel and climbing fibers and send inhibitory GABA-ergic projections to the deep cerebellar and the vestibular nuclei. Input from the basket and stellate cells inhibit the Purkinje cells. The granule cells are excitatory, glutaminergic neurons. The parallel fibers that arise from the granule cells excite the Purkinje, basket, stellate, and Golgi cells. The granule cell is excited by mossy fiber input at the cerebellar glomerulus and inhibited by Golgi cells. The mossy fiber input excites Purkinje cells indirectly through the granule cell–parallel fiber system and produces simple spikes from the Purkinje cell. The climbing fibers are entwined around the Purkinje cell, like a vine around a tree trunk, and excite it directly, producing complex spikes. The cerebellar cortex also receives noradrenergic fibers from the locus coeruleus, dopaminergic fibers from the substantia nigra, and serotonergic fibers from the raphe nuclei. All the aminergic afferents are probably inhibitory. Afferents to the cerebellar cortex and deep nuclei generally produce an increase in excitability. The Purkinje cells impose inhibitory control over the cells of the deep nuclei. Mossy fiber input causes strong direct excitation of the deep nuclei; the additional input via the granule cell-parallel fiber system provides inhibitory

control and modulation of the direct excitatory pathway. The climbing fibers modulate the activity of the Purkinje cells by controlling the influence of the different systems that converge on it.

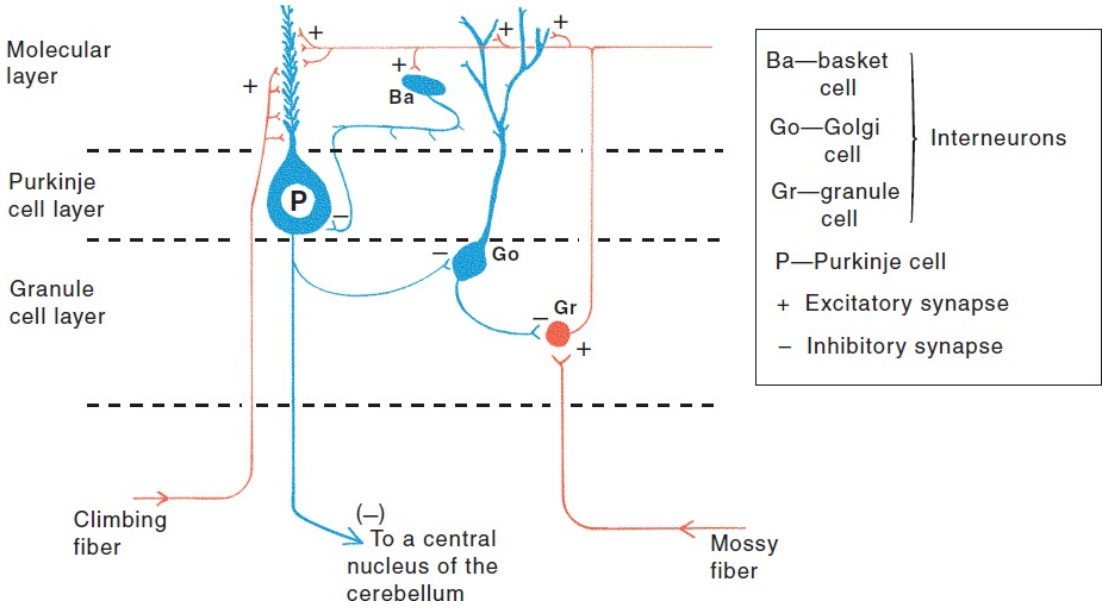

FIGURE 43.7 Neurons in the cerebellar cortex showing excitatory and inhibitory synapses. The diagram represents a longitudinally sectioned folium, with an edge-on view of the dendritic tree of the Purkinje cell. Glutaminergic (excitatory) neurons are *red*; GABA-ergic (inhibitory) neurons are *blue*. (Reprinted from Kiernan JA. *Barr's The Human Nervous System: An Anatomical Viewpoint*. 9th ed. Philadelphia: Wolters Kluwer Health/Lippincott Williams & Wilkins, 2009, with permission.)

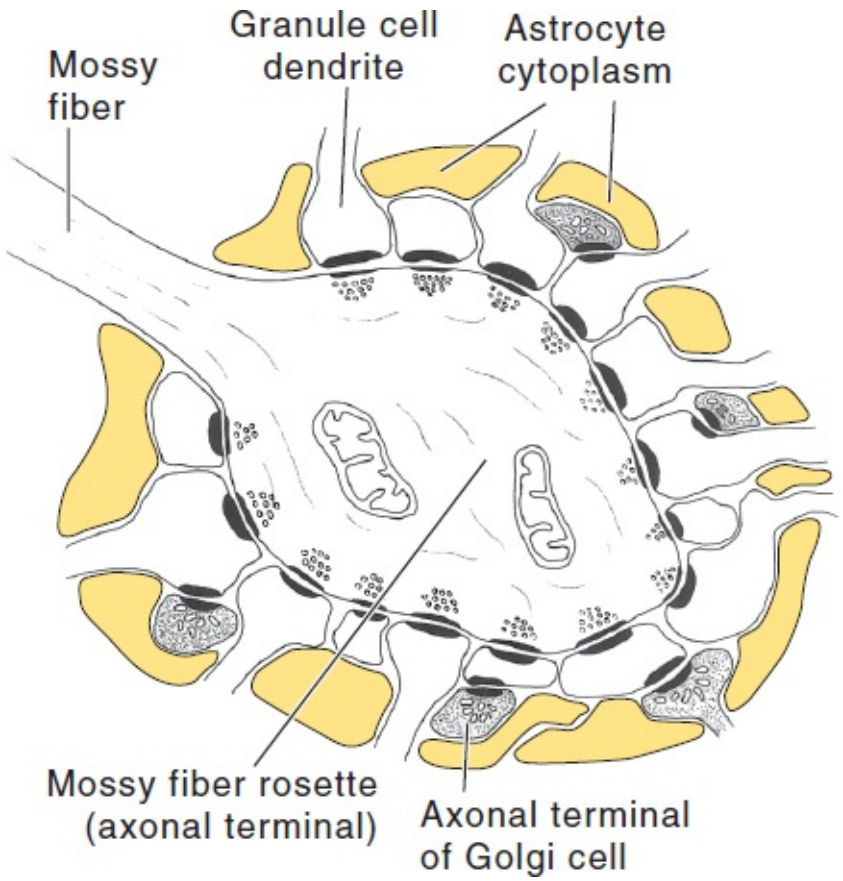

FIGURE 43.8 Ultrastructure of a synaptic glomerulus in the granule cell layer. The astrocytic processes (*yellow*) prevent diffusion of neurotransmitters to adjacent synapses. (Reprinted from Kiernan JA. *Barr's The Human Nervous System: An Anatomical Viewpoint*. 9th ed. Philadelphia: Wolters Kluwer Health/Lippincott Williams & Wilkins, 2009, with permission.)

The efferent fibers from the Purkinje cells in the cerebellar cortex are nearly all relayed to the deep nuclei, where the cerebellar outflow originates (Figure 43.5). The output from the deep cerebellar nuclei is excitatory and glutaminergic except for the projection to the inferior olive, which uses gamma-aminobutyric acid. The fastigial nucleus, the oldest of the cerebellar nuclei, receives afferent fibers from the paleocerebellum and also from the vestibular nuclei and the eighth cranial nerve. Its efferent impulses, many of them crossing in the roof, pass into the brainstem to the vestibular nuclei—especially the lateral vestibular nucleus—and to the reticular formation. Some of these go through the ICP; others travel in the SCP in the uncinate fasciculus. The fastigial nucleus also projects to the ventral lateral (VL) nucleus of the thalamus, which in turn projects to the trunk area of the motor strip. The interposed nuclei receive afferents primarily from the paravermal cortex and project to VL and to the

magnocellular part of the contralateral red nucleus, which gives rise to the rubrospinal tract. The dentate nuclei, the most important of the nuclear masses in terms of clinical function, receive afferents principally from the Purkinje cells of the neocerebellum. The dentate projects to the ipsilateral VL and intralaminar thalamic nuclei and to the contralateral red nucleus and inferior olivary nucleus.

The cerebellum is part of complex feedback loops that are involved in the coordination of motor activity (Figures 43.9 to 43.11). Large myelinated muscle spindle and Golgi tendon organ afferents travel to the cerebellum via the spinocerebellar tracts and enter primarily through the ICP. This information is processed in the hemispheres and influences the activity of Purkinje cells in the deep, midline (primarily dentate) nucleus. The Purkinje cells send axons via the SCP to the contralateral VL nucleus of the thalamus, which in turn projects to the motor cortex. Descending corticopontine fibers then synapse on pontine nuclei in the basis pontis, which in turn send pontocerebellar axons via the MCP to the cerebellar hemispheres. Other descending corticomotor fibers actually execute the task at hand. The cerebellum can thereby communicate the need for fine adjustment of movement to the cortex, and the cortex can take corrective action while simultaneously informing the cerebellum of the extent of the correction so further adjustments can be made (see Chapter 22). The motor thalamus serves to integrate cerebellar, basal ganglia, and cortical activity. Another loop consists of fibers that arise from the inferior olive and travel via the ICP to the dentate nucleus, with projections from the dentate going to the red nucleus, which then projects to the inferior olive (Guillain-Mollaret triangle, see Chapter 30). The direct vestibulocerebellar and returning cerebellovestibular fibers form yet another circuit. The cerebellum also receives input from the hypothalamus.

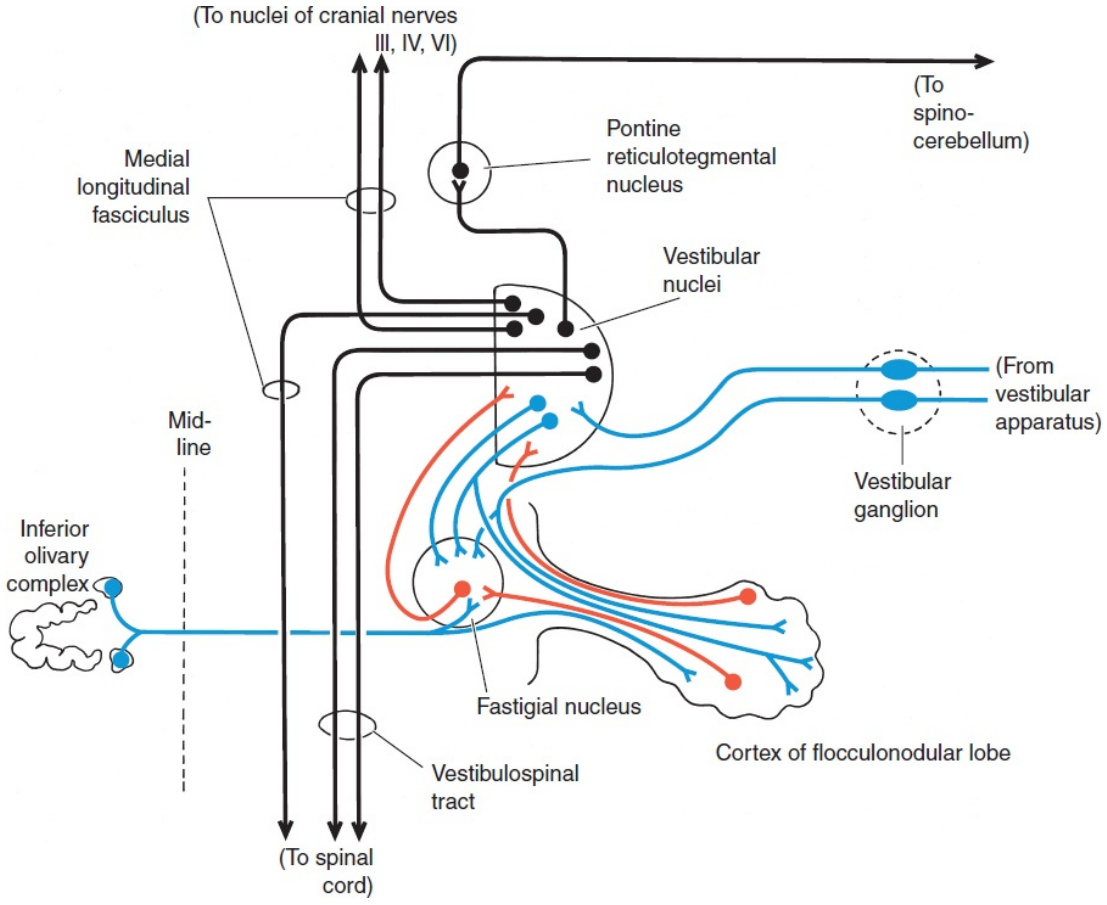

FIGURE 43.9 Connections of the vestibulocerebellum and vestibular nuclei. Afferents to the cerebellum are *blue*, cerebellar efferents are *red*, and other neurons are *black*. (Reprinted from Kiernan JA. *Barr's The Human Nervous System: An Anatomical Viewpoint*. 9th ed. Philadelphia: Wolters Kluwer Health/Lippincott Williams & Wilkins, 2009, with permission.)

CLINICAL MANIFESTATIONS OF CEREBELLAR DYSFUNCTION

Patients with cerebellar dysfunction suffer from various combinations of tremor, incoordination, difficulty walking, dysarthria, and nystagmus, depending on the parts of the cerebellum involved (see Table 22.1). Ataxia is the cardinal sign of cerebellar disease; it consists of varying degrees of dyssynergia, dysmetria, lack of agonist-antagonist coordination, and tremor. Ataxia may affect the limbs, the trunk, or the gait. Cerebellar disease may also cause hypotonia, asthenia or slowness of movement, and deviation or drift of the outstretched limbs. Disease involving the cerebellar connections in the brainstem causes abnormalities indistinguishable from disease of the cerebellum itself. When cerebellar ataxia

results from dysfunction of the cerebellar connections in the brainstem, there are usually other brainstem signs.

Dyssynergia

The essential disturbance in cerebellar disease is dyssynergia. Normally, there is harmonious, coordinated action between the various muscles involved in a movement so that they contract with the proper force, timing, and sequence of activation to carry out the movement smoothly and accurately. Cerebellar disease impairs the normal control mechanisms that organize and regulate the contractions of the different participating muscles and muscle groups to insure smooth, properly coordinated movement. There is a lack of speed and skill in performing movements that require the coordinated activity of several groups of muscles or of several movements. The cerebellum is instrumental in timing the activation of the different muscles involved in a movement. Lack of integration of the components of the act results in decomposition of movement—the act is broken down into its component parts and carried out in a jerky, erratic, awkward, disorganized manner. The cerebellum is particularly important in coordinating multijoint movements.

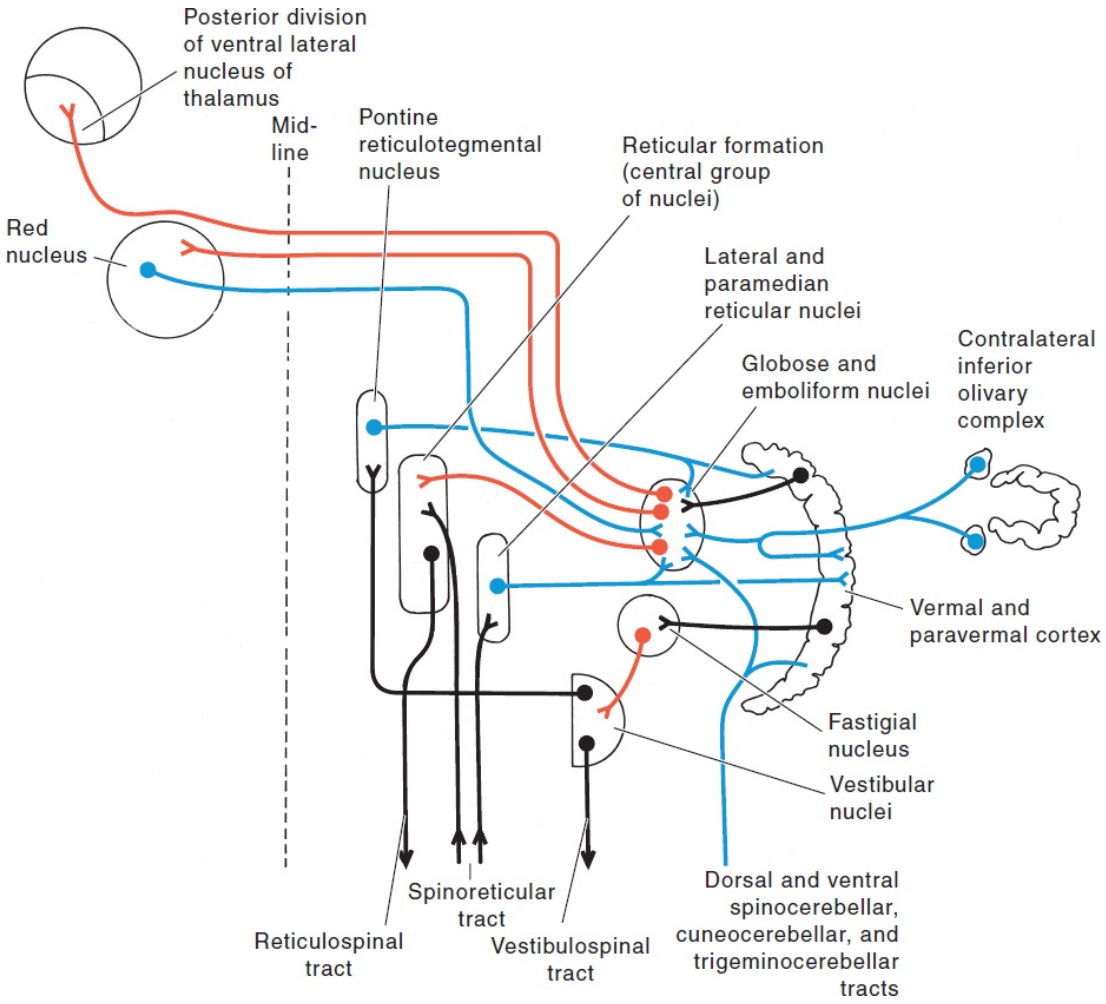

FIGURE 43.10 Connections of the spinocerebellum. Afferents to the cerebellum are *blue*, cerebellar efferents are *red*, and other neurons are *black*. (Reprinted from Kiernan JA. *Barr's The Human Nervous System: An Anatomical Viewpoint*. 9th ed. Philadelphia: Wolters Kluwer Health/Lippincott Williams & Wilkins, 2009, with permission.)

Dysmetria

Dysmetria refers to errors in judging distance and gauging the distance, speed, power, and direction of movement. Cerebellar dysfunction leads to loss of the normal collaboration between agonist and antagonist. When reaching for an object 50 cm away, the hand shoots out 55 cm, overshooting the target (hypermetria), or fails to reach the target (hypometria). Hypermetria is more common. The movement may be carried out too slowly or too rapidly with too much or too little force. The patient with dysmetria does not make a movement along a straight line between two points but erratically deviates from the

intended track. Electromyographic studies have shown that dysmetria is associated with abnormalities of the timing and force of antagonist contraction necessary to decelerate the movement. Hypermetria is associated with a more gradual buildup and prolongation of agonist activity with a delayed onset of antagonist activity or with a slower rate of rise of activity in the antagonists. Evidence suggests that different mechanisms may underlie dysmetria, depending on the anatomical location of the cerebellar lesion.

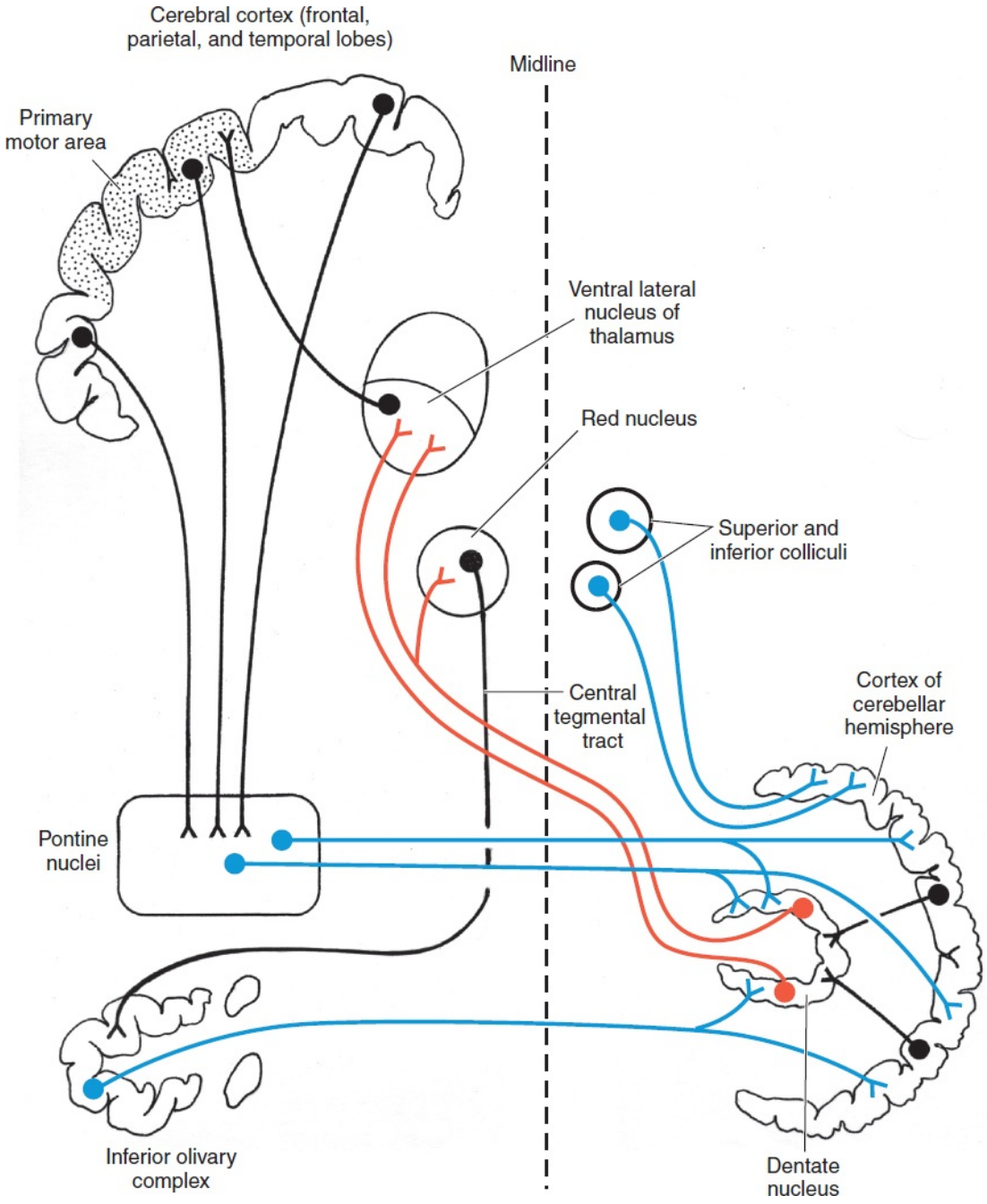

FIGURE 43.11 Connections of the pontocerebellum and vestibular nuclei. Afferents to the cerebellum are *blue*, cerebellar efferents are *red*, and other neurons are *black*. (Reprinted from Kiernan JA. *Barr's The Human Nervous System: An Anatomical Viewpoint*. 9th ed. Philadelphia: Wolters Kluwer Health/Lippincott Williams & Wilkins, 2009, with permission.)

Agonist-Antagonist Coordination

A disturbance in reciprocal innervation results in a loss of the ability to stop the contraction of the agonists and rapidly contract the antagonists to control and regulate movement. In patients with cerebellar deficits attempting to make rapid, voluntary movements, the first agonist burst is frequently prolonged, the acceleration time is longer than normal, and the acceleration time may exceed the deceleration time. The normal triphasic agonist-antagonist-agonist sequence of activity is disturbed by a too long or too short agonist burst, or an agonist burst that continues into the antagonist activity. Impairment of the ability to carry out successive movements and to stop one act and follow it immediately by its diametric opposite causes dysdiadochokinesia, loss of checking movements, and the rebound phenomenon. Dysdiadochokinesia (or adiadochokinesia) is a clumsy term (coined by Babinski) that means inability to make rapid alternating movements or RAMs. The patient with impaired RAMs has difficulty with such tests as patting the palm of one hand alternately with the palm and dorsum of the other hand, rapid tapping of the fingers, tapping out a complex rhythm, or tapping the foot in steady beat. Inability to rapidly reverse an action also causes impairment of the check response, producing the Holmes rebound phenomenon (see section on "Impaired Check and the Rebound Phenomenon").

Tremor

The most common type of cerebellar tremor is an intention (active, kinetic, or terminal) tremor that is not present at rest but becomes evident on purposeful movement. In the upper extremity, when the patient reaches to touch an object, there are irregular, to-and-fro, jerky movements perpendicular to the path of movement that increase in amplitude as the hand approaches the target. A postural tremor of the outstretched limbs may also occur, without the patient reaching for a target. Cerebellar tremor often involves the proximal muscles. When severe, cerebellar tremor may involve not only the extremities but also the head or even the entire body. Severe cerebellar tremor may at times take on an

almost myoclonic character. The tremors and other movements probably result from disease involving the cerebellar efferent pathways or their connections with the red nucleus and thalamus (dentatorubral and dentatothalamic pathways, or SCP) and are sometimes referred to as a cerebellar outflow tremor. A rubral tremor is present at rest but worsens with action and probably also results from a lesion involving the cerebellar outflow tracts (see Chapter 30).

Hypotonia

Hypotonia, or muscle flaccidity, with a decrease in resistance to passive movement, is often seen in cerebellar disease. Cerebellar dysfunction results in a decrease in the tonic output of the cerebellar nuclei, causing loss of cerebellar facilitation to the motor cortex. The muscles are flabby and assume unnatural attitudes; the parts of the body can be moved passively into positions of extreme flexion or extension. The stretch reflexes are normal or diminished in disease limited to the cerebellum. Occasionally, the tendon reflexes are "pendular." Tapping the patellar tendon with the foot hanging free results in a series of to-and-fro movements of the foot and leg before the limb finally comes to rest. Pendular reflexes are caused by muscle hypotonicity and the lack of normal checking of the reflex response. The superficial reflexes are unaffected by cerebellar disease. Cerebellar disease may also cause a characteristic position of the extended hand, probably because of hypotonia. The wrist is flexed and arched dorsally, with the fingers hyperextended, and a tendency toward overpronation. The hand is similar to that seen in Sydenham's chorea. A cerebellar lesion may cause a decrease in the normal pendular movement of the affected arm when walking. A decreased arm swing may also occur with extrapyramidal disorders and with mild hemiparesis. In the shoulder-shaking test, a cerebellar lesion causes an increase in the range and duration of swinging of the involved arm, although the movements may be irregular and nonrhythmic (see Chapter 28).

Dysarthria

Cerebellar disease often affects speech. Articulation may be slow, ataxic, slurred, drawling, jerky, or explosive in type because of dyssynergy of the muscles of phonation. A scanning type of dysarthria is particularly characteristic of cerebellar disease (see Chapter 9). The scanning speech of multiple sclerosis and

the staccato speech of Friedreich's ataxia (FA) are probably the result of cerebellar dysfunction. Dysarthria may be an isolated manifestation of paravermal cerebellar infarction.

Nystagmus

Nystagmus and other disturbances of ocular motility may occur with lesions of the cerebellum. Nystagmus often indicates involvement of vestibulocerebellar pathways. The ocular abnormalities often result from involvement of the connections of the cerebellum with other centers rather than actual cerebellar dysfunction. Cerebellar disease may cause gaze paretic nystagmus. The patient is unable to sustain eccentric gaze and requires repeated saccades to gaze laterally. With a lesion of one hemisphere, the eyes at rest may be deviated 10 to 30 degrees toward the unaffected side. When the patient attempts to gaze elsewhere, the eyes saccade toward the point of fixation with slow return movements to the resting point. The movements are more marked and of greater amplitude when the patient looks toward the affected side. When a tumor of the cerebellopontine angle is present, the nystagmus is coarse on looking toward the side of the lesion and fine and rapid on gaze to the opposite side (Bruns' nystagmus). Other ocular motility disturbances seen with cerebellar disease include skew deviation, ocular dysmetria, ocular flutter, opsoclonus, ocular tilt reaction, and saccadic intrusions. Rebound nystagmus is a type of nystagmus that may be unique to cerebellar disease; the fast component is in the direction of lateral gaze but transiently reverses direction when the eyes come back to primary position (see Chapter 14). For a detailed discussion of cerebellar eye signs, see *Cerebellar Eye Signs* from Dr. Robert B. Darroff, Neuro-ophthalmology Virtual Education Library (NOVEL), University of Utah, **Video Link 43.1.**

Other Abnormalities

Abnormalities of posture and gait with abnormal attitudes and spontaneous deviation of the head and parts of the body may be seen in cerebellar disease. In unilateral cerebellar disease, there may be deviation of the head and body toward the affected side, with past pointing of the extremities toward the affected side. When standing, there is an inclination to fall, and when walking a tendency to deviate, toward the side of the lesion. The outstretched extremities deviate laterally, toward the affected side. There may be a decrease or absence of the

normal pendular movement of the arm in walking. In midline, or vermis, lesions, the patient may not be able to stand erect and may fall either backward or forward. The gait is staggering, reeling, or lurching in character, without laterality. “Cerebellar fits” is an antiquated term, referring to episodes of decerebrate rigidity because of brainstem dysfunction due to mass effect from lesions in the cerebellum.

EXAMINATION OF COORDINATION AND CEREBELLAR FUNCTION

Clinical tests for cerebellar dysfunction are basically designed to detect dyssynergia, decomposition of movement, and dysmetria. The combination of incoordination, awkwardness, errors in the speed, range and force of movement, along with dysdiadochokinesia, and intention tremor is referred to as cerebellar ataxia. Simple observation can be as informative as a detailed clinical examination. Watching as the patient is standing, walking, dressing and undressing, buttoning and unbuttoning clothing, and tying shoelaces may reveal tremor, incoordination, clumsiness, and disturbed postural fixation. The patient may be asked to write, use simple tools, drink from a glass, and trace lines with a lightweight pen while no support is given at the elbow. The examination of infants and children may be limited to simple observation, noting the child’s ability to reach for and use toys and objects. Tests for coordination may be divided into those concerned with equilibratory and nonequilibratory functions.

Equilibratory Coordination

Equilibratory coordination refers to the maintenance of balance and the coordination of the body as a whole. The examination of station and gait assesses equilibratory coordination; these are discussed further in Chapter 44.

Nonequilibratory Coordination

Tests of nonequilibratory coordination assess the patient’s ability to carry out discrete, oftentimes relatively fine, intentional movements with the extremities. Although these are primarily tests of coordination, other neural systems must be intact for normal performance. The other components affecting fine motor

control are discussed in Chapter 27. It is important to consider handedness in judging coordination and to allow for the normal slight clumsiness of the nondominant side. Patients who are fatigued or sedated may have incoordination that is not normal for the individual. Fine motor skills may also be assessed functionally by asking the patient to do such things as thread a needle, pick up a pin, string beads, pour water, or draw circles.

The Finger-to-Nose (Finger-Nose-Finger) Test

There are several variations on the theme of having the patient touch the index finger to the nose, all of which will be included as the finger-to-nose (FTN) test. All may be carried out with the patient lying, seated, or standing. The patient extends the arm completely and then touches the tip of the index finger to the tip of the nose, slowly at first, then rapidly, with the eyes open and then closed. The examiner may place the outstretched extremity in various positions and have the test carried out in different planes and from various angles. The patient may be asked to touch the tip of his index finger to his nose, then touch the tip of the examiner's finger, and then back to the tip of his nose. It is helpful to demonstrate the requested movement, lest the patient make some odd interpretation of the verbal request; an occasional patient will attempt to put his index finger on the examiner's finger without removing it from his own nose. The examiner's finger may be moved about during the test, and the patient asked to try to touch the moving target as the finger is placed in different locations at different distances and to move both slowly and quickly. The examiner may pull his finger away and make the patient chase it (finger chase test). Fully extending the arm in this way can bring out mild intention tremor.

During these movements, note the smoothness and accuracy with which the act is executed and look for oscillations, jerkiness, and tremor. An intention tremor becomes more marked, coarse, and irregular as the finger approaches the target. There may be little tremor during the midrange of the movement, but near the end, the tremor erupts; when the finger contacts the target, the tremor stops. In cerebellar ataxia, the difficulty may vary from slight incoordination, with a blundering type of movement, to wild oscillations causing complete inability to execute the act. A patient with severe appendicular ataxia may not be able to touch hand to head, much less finger to nose. **Video 43.1** demonstrates a patient with appendicular ataxia.

Video 43.1 Cerebellar ataxia with titubation and severe appendicular ataxia causing intention tremor on finger to nose and heel to shin testing.

With dysmetria, the patient may stop before he reaches his nose, pause and then complete the act slowly and unsteadily, or overshoot the mark and bring the finger to the nose with too much speed and force. With dyssynergy, the act is not carried out smoothly and harmoniously; there may be irregular stops, accelerations, and deflections, or the movement may disintegrate into its component parts. Performing the FTN test against slight resistance may cause mild ataxia to become more obvious, or latent ataxia evident. The examiner may apply resistance by placing his fingers against the patient's forearm and exerting slight pressure as the patient moves his arm toward the nose, or by placing a long rubber band around the patient's wrist and pulling gently on it during the test. Another test is to have the patient draw a line, starting and then stopping at fixed points. He may have difficulty in starting at the correct point and may either stop short of the second point or overshoot the mark. This may also demonstrate tremor, with side-to-side oscillations along the intended tract. The patient with cerebellar disease may have macrographia, using large characters that become larger across the page, the opposite of the writing disturbance seen in Parkinson's disease.

In the finger-to-finger (fingertips in the midline) test, the patient abducts the arms widely to the horizontal and then brings in the tips of the index or middle fingers through a wide arc to touch them exactly in the midline. This is done slowly and rapidly, with the eyes first open and then closed. With unilateral cerebellar disease, the finger on the involved side may fail to reach the midline, and the finger on the normal side may cross the midline to reach it. Also, the arm on the affected side may sag or rise, causing the finger on that side to be below

or above the one on the normal side.

In hysteria or malingering, there may be bizarre responses of various types. The patient may act as if unable to touch the finger to the nose, or circle around it with widespread, wandering movements but eventually touch the very tip. Or the patient may repeatedly but precisely touch some other part of the face, implying there is no loss of sensation or coordination.

Similar tests may be used to evaluate the lower extremities. In the heel-to-shin (heel-knee-shin/toe) test, the patient is asked to place the heel of one foot on the opposite knee, tap it up and down on the knee several times, push the point of the heel (not the instep) along the shin in a straight line to the great toe, and then bring it back to the knee. The patient with cerebellar disease is likely to raise the foot too high, flex the knee too much, and place the heel down above the knee. The excursions along the shin are jerky and unsteady. With sensory ataxia, the patient may have difficulty locating the knee with the heel, groping around for it; there is difficulty keeping the heel on the shin, and it may slip off to either side while sliding down the shin. In the toe-to-finger test, the patient tries to touch his great toe, knee bent, to the examiner's finger. If there is dysmetria, the patient will undershoot or overshoot the mark; intention tremor and oscillations may also be evident. The patient may be asked to draw a circle or a figure eight with his foot, either in the air or on the floor; in ataxia, the movement will be unsteady and the figure irregular.

Rapid Alternating Movements

With dysdiadochokinesia, one act cannot be immediately followed by its diametric opposite; the contraction of one set of agonists and relaxation of the antagonists cannot be followed immediately by relaxation of the agonists and contraction of the antagonists. Patients with cerebellar ataxia may have great difficulty making these kinds of movements **(Video 43.1)**. A common test for dysdiadochokinesia is to have the patient alternately pronate and supinate his hands, as in patting alternately with the palm and dorsum of the hand on the thigh or on the palm or dorsum of the other hand, or imitating screwing in a light bulb or turning a doorknob. The movements are performed repetitively and as rapidly as possible. Any movement involving reciprocal innervation and alternate action of agonists and antagonists can be used, such as alternate opening and closing of the fists, quickly flexing and extending individual fingers, touching the tip of the index finger to the tip or extended interphalangeal joint of the thumb, or patting rapidly against a table top with hand or fingertips.

A good test is to have the patient touch the tip of his thumb with the tip of each finger rapidly and in sequence—starting with the index finger and proceeding to the little finger, repeating with the little finger and going to the index finger, and so forth. Another good test is to have the patient tap out a simple rhythm with each hand (e.g., 1-2-3/pause in steady beat), and then a more complex but familiar rhythm (e.g., Happy Birthday song). Testing RAMs in the lower extremity is much more limited. The patient may be asked to pat the foot steadily, against the floor if standing, and against the examiner's palm if recumbent or to repetitively touch the heel up and down to the knee if supine. RAMs of the tongue may be tested by having the patient move the tongue in and out or from side to side as rapidly as possible.

In all of these tests, note the rate, rhythm, accuracy, and smoothness of the movements. In patients with ataxia, the RAMs are carried out either slowly and hesitantly, with pauses during transition between the opposing motions, or unsteadily and irregularly, with loss of rhythm. There may be a rapid fatigability: The movements may be executed satisfactorily in the beginning, but after a few attempts, they become awkward and clumsy. The two extremities are usually compared, but patients with bilateral abnormalities are common, and the examiner must rely on experience or use another control. Demonstrating the movements to the patient provides an opportunity for the examiner to be the control. For some maneuvers, such as rapid, repetitive finger movements, the two extremities can be examined simultaneously and one side compared with the other. Simultaneous testing may also cause accentuation of the abnormality on the affected side.

Impaired Check and the Rebound Phenomenon

Checking movements involve contraction of the antagonists after a load is unexpectedly removed during strong contraction of the agonists. The agonists must immediately relax and the antagonists must contract to provide braking after the sudden release of resistance. Because cerebellar dysfunction causes impairment of the reciprocal relationship between agonist and antagonist, patients may have impairment of the checking response.

In the Holmes (Stewart-Holmes) rebound test, the patient holds the arm adducted at the shoulder and flexed at the elbow, with the forearm supinated and the fist firmly clenched. The elbow may rest on a table or be held unsupported close to the body. The examiner pulls on the wrist, and the patient strongly resists the examiner's attempt to extend the elbow. The examiner then suddenly

releases his grip on the wrist. Normally, with the sudden unloading, the contraction of the elbow flexors immediately ceases and is rapidly followed by contraction of the elbow extensors to arrest the sudden flexion movement and stop the patient from hitting himself. The normal patient is able to control the unexpected flexion movement of the elbow. In cerebellar disease, when the strongly flexed extremity is suddenly released, the patient cannot stop the flexor contraction and engage the extensors to stop the elbow movement. Because of loss of the checking response, the fist flies up to the shoulder or mouth, often with considerable force. The examiner's free arm should be placed between the patient's fist and face to block the blow. The prevalent description of this as the Holmes rebound phenomenon is not precisely correct. Stewart and Holmes used rebound to refer to the jerk back in the opposite direction, the recoil, on release of the restraint. The rebound phenomenon is present normally and exaggerated in spastic limbs. It is the absence of rebound (usually accompanied by impaired checking) in limbs affected by cerebellar disease that is abnormal. It is the loss of rebound that is abnormal and characteristic of cerebellar disease, not its presence.

The rebound test may be carried out in other ways. Elbow extension against resistance may be tested instead of flexion. With both arms outstretched in front of the patient, the examiner may press either down or up on them as the patient resists and then suddenly lets go. This allows comparison of the rebound phenomenon and loss of checking movements on the two sides. In the lower extremities, rebound can be tested by sudden release after the patient has been resisting either flexion or extension at the knee, hip, or ankle. Impaired checking and loss of rebound are not invariably present in cerebellar disease and may sometimes be present in normal limbs. Loss of rebound is more significant unilaterally than bilaterally. In the arm-stopping test, the patient holds both arms overhead or by his sides, the examiner holds his arms outstretched horizontally, and then the patient tries to quickly bring his arms up or down so that his fingertips are at the exact same level as the examiner's. With a unilateral hemispheric lesion, the good arm will stop on target, the affected arm often overshoots and then corrects in the opposite direction, oscillating around the target before eventually coming to rest.

Deviation and Past Pointing

Patients with cerebellar disease often have difficulty maintaining normal alignment of the limbs or body when performing a task such as holding the arms

outstretched or walking, especially with eyes closed. The patient may miss when trying to reach out to touch a target (past pointing), drift to one side when walking eyes closed, or have drift of the outstretched arm **(Video 43.2)**. Similar findings may occur with vestibular lesions.

Video 43.2 Vestibular and cerebellar past pointing and the stepping test.

To perform the traditional test for past pointing, the patient and examiner should be facing, either seated or standing, the outstretched upper extremity of each held horizontally with the index fingers in contact. The patient raises his arm to a vertical position, finger pointed directly upward, and then returns to horizontal to again touch the examiner's finger. The maneuver should be tried a few times with the eyes open and then executed with the eyes closed. The arms may be tested sequentially or simultaneously. The test is less commonly done with the patient raising the arm from below up to the horizontal. Normally, the patient will return to the starting position fairly accurately, without any drift or deviation. In labyrinthine disease or with a cerebellar hemispheric lesion, the arm will deviate to the involved side on the return track, more so with the eyes closed. This deviation is called past pointing. A simpler way to test for past pointing is to have the patient close his eyes while doing the finger-nose-finger test. With eyes open, the pointing is accurate, but with eyes closed, the patient points off to the side of the target. Repeating the test several times may produce greater deviation. With severe lesions, past pointing may occur even with eyes open. The pattern is different in vestibular as opposed to cerebellar past pointing. In vestibular disease, past pointing occurs with both upper extremities toward the involved side; in unilateral cerebellar disease, past pointing occurs toward the side of the lesion, or erratically to either side, but only in the ipsilateral arm. Past

pointing is discussed further in Chapter 17.

A cerebellar lesion may also cause drift of the outstretched upper extremities. Three types of drift may occur when the patient attempts to hold the arms outstretched with eyes closed: pyramidal drift, parietal drift, and cerebellar drift. In pronator drift (Barré's sign) due to a pyramidal lesion, the arm sinks downward and there is accompanying pronation of the forearm (see Chapter 27). In parietal drift, the arm usually rises and strays outward (updrift). With cerebellar drift, the arm drifts mainly outward, either at the same level, rising, or less often sinking. Testing is done with arms outstretched and eyes closed. With disease involving one cerebellar hemisphere, the arm drifts toward the side of the lesion. The deviation may be accentuated by tapping the patient's outstretched wrists. Tapping on the wrists may also create an up-and-down oscillation because of impaired checking, so that the arm swings up and down a few times and gradually drifts laterally and often upward.

Position holding can also be tested in the lower extremities. The patient, lying supine, raises the legs one at a time. When there is ataxia, the leg cannot be lifted steadily or in a straight line. There may be adduction, abduction, rotation, oscillations, or jerky movements from one position to another. When the limb is lowered, the patient may throw it down heavily, and it may not return to its original position beside its mate but may be deviated across it or away from it. When the seated patient extends the legs without support and attempts to hold them steady, a unilateral cerebellar lesion may cause oscillations and lateral deviation of the ipsilateral extremity. If the prone patient bends the knees and tries to maintain the shins vertically, there may be marked oscillations and lateral deviation of the leg on the side of the lesion.

Deviation and drift may also occur when the patient tries to walk with eyes closed. As in vestibulopathy, the patient drifts to the side of the lesion (see Chapter 17). Walking back and forth with eyes closed may reveal a "compass" or "star" gait due to deviation toward the involved side. When walking around a chair, the patient shows a tendency to fall toward the affected side.

CEREBELLAR SYNDROMES

Cerebellar disease may affect all or only a specific part of the cerebellum. There are two clearly defined cerebellar syndromes: a midline or vermis syndrome and a lateral or hemispheric syndrome. With the vermis, or midline, syndrome, the

outstanding symptoms are abnormalities of station and gait, with abnormalities ranging from slight widening of the base on walking in mild disease (gait ataxia) to total inability to sit or walk in severe disease. Disease of the cerebellar hemispheres produces appendicular ataxia, disturbance in coordination of the ipsilateral extremities, the arm more than the leg. The primary clinical manifestations of dysfunction of the FN lobe or its connections are disturbances of equilibrium; nystagmus, often positional; and other abnormalities of extraocular movement. There is no limb ataxia. Table 43.1 summarizes the clinical manifestations of disease of these parts of the cerebellum.

TABLE 43.1 Clinical Manifestations of Disorders of the Cerebellum (Related to the Different Zones of the Cerebellum)

Zone of Cerebellum	Clinical Manifestation	Possible Disorder
Flocculonodular lobe (archicerebellum)	Nystagmus; extraocular movement abnormalities	Medulloblastoma
Vermis (paleocerebellum)	Gait ataxia	Alcoholic degeneration
Hemisphere (neocerebellum)	Appendicular ataxia	Tumor; stroke
Pancerebellar	All of the above	Paraneoplastic

The manifestations of cerebellar disease differ markedly in severity, depending upon the acuteness or chronicity of the process. The ability of the nervous system to compensate for a cerebellar lesion can be remarkable. If the lesion is acute, the symptoms are profound; if it is slowly progressive, they are much less severe. There may be considerable recovery from an acute lesion. If a lesion develops insidiously, there may be extensive involvement of the hemispheres without much in the way of clinical findings. The neural plasticity and compensation are such that some patients with little remaining cerebellar tissue can eventually function quite well. The symptoms of cerebellar disease are similar regardless of the etiology of the disease process, and whether the lesion is congenital or acquired.

Midline Syndrome

The vermis is important in the control of axial structures, or those that are bilaterally innervated; vermian lesions primarily affect midline functions, such as walking and coordination of the head and trunk. A patient with mild vermian disease has gait ataxia. The base is widened, tandem gait is particularly difficult, and there may be decompensation on turning. The Romberg test is negative—the imbalance does not worsen significantly with eyes closed. With severe dysfunction of the vermis, there may be gross postural and locomotor disturbances of the entire body. There is no lateralization, and the tendency to fall may be either backward or forward. The gait is wide based and characterized by swaying and staggering; the patient may reel in a drunken manner to either side. Cerebellar ataxia is discussed further in Chapter 44.

With truncal ataxia, there is swaying and unsteadiness when standing, and the patient may be unable to maintain an upright position. There may be loss of the ability to remain erect when seated or to hold the neck and head steady and upright; when severe, the standing and sitting balance disturbance leads to constant, to-and-fro swaying, nodding, and weaving movements of the head and trunk when the patient is upright known as titubation. The head movements in titubation are primarily anteroposterior (yes-yes) at 3 to 4 Hz. Vermis dysfunction causes little or no abnormality of the extremities, especially the upper extremities, although all coordinated movements may be poorly performed. Muscle tone and reflexes are normal. Nystagmus may be present but is usually not marked. Ocular dysmetria, rebound nystagmus, and pursuit abnormalities may also occur. Lesions involving the vermis may cause upbeat nystagmus. Dysarthria is often present. There is sometimes an abnormal rotated or tilted head posture.

Common causes of a midline cerebellar syndrome are alcoholic cerebellar degeneration and medulloblastoma. Alcohol preferentially poisons the vermis, leading to a characteristic syndrome of gait ataxia with sparing of the limbs. Such patients may have no demonstrable lower-extremity ataxia while lying supine, yet be totally unable to walk. Unwary examiners may conclude such findings represent hysteria. Medulloblastomas occur most often in the cerebellar vermis.

Hemispheric Syndrome

With a lesion involving one cerebellar hemisphere, the manifestations are appendicular rather than axial. Cerebellar hemispheric deficits are unilateral and ipsilateral to the lesion, as the pathways are uncrossed (or, more correctly, double crossed). There is a disturbance of skilled movements of the extremities, with ataxia, dysmetria, dyssynergy, dysdiadochokinesia, and hypotonicity affecting the arm and hand more than the leg and foot. Distal movements are affected more than proximal and fine movements more than gross ones. Movements are performed irregularly, and there may be intention tremor or other hyperkinesias if the dentate nucleus or its efferent pathways are involved.

Posture and gait are not impaired as severely as in the vermis syndrome, but abnormalities do occur. There may be swaying and falling toward the side of the lesion. The patient may be able to stand one legged using the contralateral but not the ipsilateral foot. He may be unable to bend his body toward the involved side without falling. The abnormalities often resemble those of a unilateral vestibular lesion. On walking, there may be unsteadiness, with deviation or rotation toward the involved side. There may be drift and past pointing toward the involved side. Dysarthria may occur, although disturbances of articulation are not as severe as in vermis lesions. Nystagmus is a common finding, usually horizontal but sometimes rotatory. It is usually more prominent when looking toward the side of the lesion. Common causes of a cerebellar hemispheric syndrome include cerebellar astrocytoma, multiple sclerosis, and lateral medullary stroke.

Diffuse Cerebellar Dysfunction

Some conditions affect the cerebellum diffusely, causing midline and bilateral hemispheric abnormalities. Patients may have nystagmus, gait and truncal ataxia, and appendicular incoordination. Etiologies include the hereditary spinocerebellar ataxia (SCA) syndromes, drugs (especially phenytoin), toxins, and paraneoplastic cerebellar degeneration.

Sensory Ataxia

Incoordination may also result from a lack of proprioceptive input from the limbs. Sensory ataxia results from peripheral nerve disease affecting primarily sensory fibers; pathology involving the dorsal root ganglia, dorsal roots, or posterior columns of the spinal cord; interruption of the proprioceptive pathways

in the brainstem; or disease of the parietal lobe. Incoordination due to sensory ataxia can closely mimic that of cerebellar ataxia (Table 43.2). With cerebellar ataxia, it makes little difference whether the patient's eyes are open or closed. In sensory ataxia, performance is not normal with eyes open but worsens markedly with eyes closed. The different components of the abnormality may behave slightly differently when visual input is removed. Some of the tremor in sensory ataxia is due to visually guided voluntary corrections of deviations from the intended track. Because of loss of appreciation of limb position in space, with eyes closed, the patient may be unable to find his nose or the examiner's finger, but the tremor may actually abate because the patient cannot see that a deviation is occurring and does not attempt to correct it. He may be wildly off target but move in a straighter line. The distinction between cerebellar and sensory ataxia is also made by the associated findings (Table 43.2).

TABLE 43.2 Associated Findings Helpful in Distinguishing Sensory from Cerebellar Ataxia

Sensory Ataxia	Cerebellar Ataxia
Sensory loss, especially for joint position and vibration	Nystagmus, ocular dysmetria, and other eye movement abnormalities
Steppage gait	Reeling, ataxic gait
Decreased reflexes	Other signs of cerebellar disease (dyssynergia, dysmetria, dysdiadochokinesia, hypotonia, rebound, impaired check response)

Other Abnormalities

There are many potential causes for a lack of coordination of movement. All of the levels of the motor system are involved in performing smooth and accurate movement. Weakness of any origin may interfere with skill and precision. Abnormalities of tone of any type may interfere with coordination. Diseases of the extrapyramidal system may impair motor control because of rigidity, akinesia

or bradykinesia, lack of spontaneity, and loss of associated movements. A corticospinal tract lesion may cause jerkiness and clumsiness of movement, loss of motor control, and poor integration of skilled acts. Nonorganic illness may cause difficulty with coordination simulating true ataxia. Hyperkinetic movement disorders may cause irregularity in the timing and excursion of successive movements. Proprioceptive abnormalities may impair motor performance. To always attribute ataxia to cerebellar disease is an oversimplification because many conditions can cause incoordination and clumsiness. Often, the cause is multifactorial. A good general rule is to avoid drawing conclusions about the meaning of "cerebellar signs" in the face of any significant degree of weakness, spasticity, rigidity, or sensory loss. When the examination shows no other abnormalities, incoordination and awkwardness of movement are usually due to cerebellar disease.

Frontal lobe ataxia refers to disturbed coordination due to dysfunction of the contralateral frontal lobe; it may resemble the deficits due to abnormalities of the ipsilateral cerebellar hemisphere. Frontal lobe ataxia results from disease involving the frontopontocerebellar fibers en route to synapse in the pontine nuclei. Frontal lobe lesions may produce other abnormalities, such as hyperreflexia, increased tone, and pathologic reflexes, whereas purely cerebellar lesions typically cause hypotonia, diminished or pendular reflexes, and no pathologic reflex responses. Pressure on the brainstem by a cerebellar mass lesion may cause corticospinal tract findings that can confuse the picture. Bruns' ataxia refers to a gait disturbance seen primarily in frontal lobe lesions (see Chapter 44).

A variety of other functions have been attributed to the cerebellum, and there has been increasing awareness of its nonmotor functions. Roles for the cerebellum in learning, planning, emotion, and cognition have been proposed. It may play a role in sensory-motor integration, motor coordination, motor learning, and timing. A cerebellar cognitive affective syndrome has been described, characterized by disturbed executive function, visuospatial disorganization and impaired visuospatial memory, personality change, and linguistic difficulties such as dysprosodia, agrammatism, and mild anomia. The cerebellum has considerable influence in language processing. Children with cerebellar malformations have a high prevalence of nonmotor developmental and functional disabilities including cognitive, language, and social-behavioral deficits. The cerebellar mutism syndrome (posterior fossa syndrome) consists of diminished speech progressing to mutism, emotional lability, hypotonia, and

ataxia. It is common following resection of a midline posterior fossa tumor in children, particularly medulloblastoma. Dysarthria may occur as a sequela. Nonmotor dysfunction of the cerebellum has been implicated in conditions as diverse as autism, dyslexia, and schizophrenia.

CEREBELLAR DISORDERS

Conditions causing a relatively acute ataxia include metabolic disorders, infections, toxins, neoplasms, infarction, hemorrhage, and demyelinating disease. An idiopathic condition, acute cerebellar ataxia or "cerebellitis," is most common in children. Autoimmunity may account for some cases of idiopathic sporadic cerebellar ataxia. The metabolic disorders include Wernicke's encephalopathy, biotinidase deficiency, and hyperammonemia. Conditions causing episodic or recurrent ataxia include channelopathies, such as the episodic ataxia syndromes, basilar artery migraine, recurrent toxin exposure (alcohol is the archetypal cerebellar toxin), and metabolic disorders such as Hartnup's disease, Leigh's syndrome, and organic acidurias.

Chronic ataxia may be relatively fixed or progressive. Static forms include alcoholic cerebellar degeneration and malformations, such as the Dandy-Walker and Chiari malformations. The Chiari I malformation is relatively common. It is often asymptomatic and discovered incidentally on neuroimaging. When symptomatic, typical other manifestations include headache and neck pain (worsened by cough or Valsalva maneuver), evidence of lower brainstem dysfunction (e.g., dysarthria, dysphagia, downbeat nystagmus), myelopathy, and syringomyelia. Causes of chronic progressive ataxia include the hereditary SCAs and acquired disorders such as hypothyroidism, paraneoplastic cerebellar degeneration, and multiple sclerosis.

TABLE 43.3 The Common Forms of Spinocerebellar Ataxia (SCA)

Name	Genetics	Phenotype
SCA1	6p22–p23; CAG repeats; ataxin-1	Ataxia; ophthalmoparesis; pyramidal and extrapyramidal findings

SCA2	12q23–q24.1; CAG; ataxin-2	Ataxia; slow saccades; minimal pyramidal and extrapyramidal findings
SCA3 (Machado-Joseph disease)	14q24.3–q32; CAG repeats; ataxin-3	Ataxia; ophthalmoparesis; variable pyramidal, extrapyramidal, and amyotrophic signs
SCA6	19p13.2; CAG repeats; CACNA1A protein, P/Q-type calcium channel subunit	Ataxia; dysarthria; nystagmus; mild proprioceptive sensory loss
SCA7	3p14.1–p21.1; CAG repeats; ataxin-7 binding protein	Ophthalmoparesis; visual loss; ataxia; dysarthria; extensor plantar response; pigmentary retinal degeneration
SCA8	13q21 with CTG repeats; noncoding; 3′ untranslated region of transcribed RNA	Gait ataxia; dysarthria; nystagmus; leg spasticity; reduced vibratory sensation

Modified from Rosenberg RN. Ataxic disorders. In: Longo D, Fauci AS, Kasper DL, et al., eds. *Harrison's Principles of Internal Medicine*. 18th ed. New York: McGraw-Hill, 2011.

The inherited ataxias may be transmitted through autosomal dominant, autosomal recessive, or maternal (mitochondrial) modes of inheritance. A genomic classification has now largely superseded previous ones based on clinical expression alone. The clinical manifestations and neuropathologic findings of cerebellar disease dominate the clinical picture; there may also be characteristic changes in the basal ganglia, brainstem, spinal cord, optic nerves, retina, and peripheral nerves. The conditions clinically range from purely cerebellar syndromes to mixed cerebellar and brainstem disorders, cerebellar and basal ganglia syndromes, and spinal cord or peripheral nerve disease.

The most common form of hereditary ataxia is the autosomal recessive condition called FA. The most common molecular abnormality in FA is a

trinucleotide repeat expansion in the gene encoding *frataxin*. The condition is characterized by progressive gait and limb ataxia with associated limb muscle weakness, absent lower limb reflexes, extensor plantar responses, dysarthria, decreased vibratory and proprioception sense, scoliosis, pes cavus, hammer toes, and cardiac abnormalities. Onset is usually in the first or second decade, before the end of puberty. The triad of hypoactive knee and ankle jerks, signs of progressive cerebellar dysfunction, and preadolescent onset is commonly regarded as sufficient for diagnosis. Uncommon features and atypical forms have been recognized. As many as one-quarter of the patients, even homozygotes, have atypical features, including older age at presentation and intact tendon reflexes. Smaller trinucleotide repeat expansions correlate with later onset and longer times to loss of ambulation.

The autosomal dominant SCAs include SCA types 1 through 47 (at this writing). The most common disorders are SCA 1, 2, 3 (Machado-Joseph disease), 6, 7, and 8 (Table 43.3). Many of the conditions are nucleotide repeat disorders; others are channelopathies. Most of the CAG repeat disorders result in proteins, termed *ataxins*, that produce a toxic gain of function. Although the phenotype is variable for any given disease gene, a pattern of neuronal loss with gliosis is produced that is relatively unique for each ataxia.

Video Links

Video Link 43.1. Cerebellar eye signs. https://collections.lib.utah.edu/details?id=188448

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CHAPTER 44

Gait and Station

It is likely possible to learn more about neurologic status from watching a patient walk than from any other single procedure, and observation of gait should always be part of a neurologic examination. Abnormalities of gait are a common clinical problem with numerous causes, both neurologic and nonneurologic. A careful general evaluation is always necessary to exclude a nonneurologic cause.

Station is the way a patient stands and gait the way she walks. Standing and walking are active processes that depend upon a number of factors and reflex responses. The mechanisms are complex, especially in the human, whose biped gait and erect position over a narrow base require more efficient maintenance and control of equilibrium than is necessary in quadrupeds. Normal resting tone, especially in the antigravity muscles, is essential. The postural and righting reflexes described in Chapter 41 are particularly important. Standing may be considered a postural reflex that is dependent on reflexes mediated through the brainstem and influenced to a major degree by tonic neck and labyrinthine reflexes. If the mechanisms mediating static and postural reflexes are impaired, normal standing and walking will be impaired. In addition, proprioceptive sensation must be received, the skeletal system must be intact, the muscles must be functioning normally, and coordination must be adequate. Gait and station may be affected by abnormalities of proprioception, abnormalities of muscle power or tone, and abnormalities of vestibular function and by dysfunction of the basal ganglia, the cerebellum, or their connections.

Neurologic causes of an abnormal gait include conditions as varied as footdrop due to peroneal nerve palsy, myopathy, hydrocephalus, and cerebellar degeneration. The various gait abnormalities have different findings on physical examination in regard to the gait itself, such as a steppage pattern as opposed to a pelvic waddle. The differential diagnosis of the gait abnormality is also very dependent on the history and the other clinical signs present. Some of the more

common abnormal gait patterns are summarized in Table 44.1.

EXAMINATION OF STATION

Station is the patient's attitude, posture, or manner of standing. The healthy individual stands erect with her head up, chest out, and abdomen in. Abnormality of station may be an important indicator of neurologic disease. Station is tested by having the patient stand, feet closely together, noting any unsteadiness or swaying. More rigorous testing includes having the patient stand eyes open and eyes closed, on one foot at a time, on toes and heels, and tandem with one heel in front of the toes of the other foot. She may be given a gentle push to see whether she falls to one side, forward, or backward.

Patients with unsteadiness standing often attempt to compensate by placing the feet wide apart in order to stand on a broader and steadier base. In cerebellar disease, the patient usually stands on a broad base and there is swaying, to more or less an equal degree, with eyes open and closed. With a lesion of the vermis, the patient may sway backward, forward, or to either side. With a lesion of one hemisphere, she sways or falls toward the affected side. Unilateral vestibular disease also causes falling toward the affected side. In a unilateral, cerebellar hemispheric lesion, or in a unilateral vestibulopathy, the patient may tilt the head toward the involved side with the chin rotated toward the sound side, with the shoulder on the involved side somewhat higher than the other and slightly in front of it. If the patient with a cerebellar hemispheric lesion is given a light push—first toward one side and then toward the other—she will lose balance more easily when pushed toward the involved side. If asked to stand on one foot at a time, the patient with a cerebellar hemispheric lesion may be unable to maintain equilibrium standing on the ipsilateral foot but may stand without difficulty on the contralateral foot.

TABLE 44.1 Some of the More Common Neurologic Abnormalities of Gait

Gait Disorder	Gait Characteristics	Usual Associated Findings
Spastic	Stiff legged, scissoring (wooden soldier)	Hyperreflexia, extensor plantar

		responses
Cerebellar ataxia	Wide based, reeling, careening (drunken sailor)	Heel-to-shin ataxia, other cerebellar signs
Sensory ataxia	Wide based, steppage	Positive Romberg, impaired joint position sense
Hemiparetic	Involved leg spastic, circumduction, often with footdrop	Weakness, hyperreflexia, extensor plantar response
Parkinsonian	Small steps, flexed posture, shuffling, festination	Tremor, rigidity, bradykinesia
Marche à petits pas	Small steps, slow shuffling	Dementia, frontal lobe signs
Footdrop (unilateral or bilateral)	High steppage pattern to clear the toes from the floor, double tap with toe strike before heel strike	Foot dorsiflexion weakness
Myopathic	Exaggerated “sexy” hip motion, waddling, lumbar hyperlordosis	Hip girdle weakness

Other abnormalities may be apparent during station testing, particularly movement disorders. Patients with chorea seem unusually fidgety and often have small adventitial finger movements **(Video 30.4)**. Skeletal changes (such as kyphosis, scoliosis, or lordosis); abnormalities in the position of the head, shoulders, hips, or extremities; asymmetries; anomalies of development; and abnormalities of contour may be apparent. Patients who are weak or debilitated may need support to stand erect. If the patient is unable to stand alone, or unable to stand at all, document how much support and assistance is required (e.g., stands with a walker, chair bound). If the patient is chair bound or bed bound, describe the sitting or recumbent posture. The patient with a hemiparesis may

stand with the upper extremity flexed and pronated and the lower extremity extended. Patients with Parkinson's disease stand in a flexed posture, stooped over with head and shoulders bent forward and arms and knees flexed. Pelvic girdle weakness may cause pronounced lordosis, especially in muscular dystrophy (Figure 4.5). Depressed patients may appear stooped and dejected; in manic states, an erect, domineering, aggressive posture may be present. In schizophrenia, the patient may assume bizarre postures and hold them for long periods of time. Hyperkinesias, such as athetoid and choreic movements, may become evident during the evaluation of station.

The Romberg Sign

When proprioception is disturbed, the patient may be able to stand with eyes open but sways or falls with eyes closed (Romberg sign). The Romberg sign is often misunderstood and misinterpreted. The essential finding is a difference between standing balance with eyes open and closed. In order to test this function, the patient must have a stable stance eyes open and then demonstrate a decrease in balance with eyes closed when visual input is eliminated, and the patient must rely on proprioception to maintain balance (see **Video Link 44.1**). Romberg described this sign in patients with tabes dorsalis and thought it was pathognomonic. He said, "If he is ordered to close his eyes while in the erect posture, he at once commences to totter or swing from side to side; the insecurity of his gait also exhibits itself more in the dark." Romberg did not state that the feet should be placed together; that was a later addition. Nor did he comment on where the arms were to be positioned. It is common practice to have the patient hold the arms outstretched in front, but this is in order to check simultaneously for pronator drift or to perform finger-to-nose testing; it is not what Romberg described. Some authorities recommend the arms be held at the sides, others that the arms be crossed on the chest. Whether arm position makes any difference in test sensitivity is unknown. Turning the head side to side eliminates vestibular clues and increases the reliance on proprioception (Ropper's refined Romberg test).

The Romberg test can be difficult to interpret. There is some variability, even among expert examiners, in how the Romberg test is performed and interpreted. Many patients sway slightly with eyes closed, and minimal amounts of sway, especially in elderly patients, are seldom significant. Minor, normal swaying may stop if the patient is simply asked to stand perfectly still. Most clinicians

discount sway at the hips and insist on seeing sway at the ankles before calling the test positive, some require the patient take a corrective step to the side, and others that the patient nearly fall. Some require the patient be barefoot. The “sharpened” or tandem Romberg is done by having the patient stand in tandem position with eyes open and closed; the limits of normality for this variation are conjectural.

The Romberg sign is used primarily as a test of proprioceptive, not cerebellar, function. The pioneering 19th-century clinicians thought it was particularly useful in separating tabes dorsalis from cerebellar disease. In fact, patients with cerebellar disease, particularly disorders of the vestibulocerebellum or spinocerebellum, may have some increase in instability with eyes closed, but not usually to the degree seen with impaired proprioception. A patient with an acute unilateral vestibulopathy may fall toward the side of the lesion when standing with eyes closed. Patients with cerebellar disease, or those with severe weakness, may not have a stable base eyes open. It may help to have the patients widen stance to the point where they are stable eyes open, then close the eyes, and check for any difference. Only a marked worsening of balance with eyes closed qualifies as a positive Romberg sign. A patient who cannot maintain balance feet together and eyes open does not have a positive Romberg.

Some histrionic patients will sway with eyes closed in the absence of any organic neurologic impairment (false Romberg sign). The swaying is usually from the hips and may be exaggerated. If the patient takes a step, the eyes may remain closed, which never happens with a bona fide Romberg. The instability can often be eliminated by diverting the patient’s attention. Effective distracters are to ask the patient to detect numbers the examiner writes with her finger on the forehead, to wiggle the tongue, or to perform the finger-to-nose testing. Having the shoes off and watching the toe movements may be very informative. The toes of the patient with histrionic sway are often extended; the patient with organic imbalance flexes the toes strongly and tries to grip the floor.

PHYSIOLOGY OF GAIT

The brainstem and spinal cord in lower forms contain “central pattern generators,” which are groups of interneurons that coordinate the activity in pools of motor neurons to produce patterned movements. Although the existence of such cell groups in humans is unproven, locomotion likely depends on

activity in pattern generators. The pattern generators control the activity in lower motor neurons that execute the mechanics of walking. Higher centers in the subthalamus and midbrain, particularly the pedunculopontine nucleus, modulate the activity in the spinal cord pattern generators through the reticulospinal tracts.

The gait cycle refers to the events that transpire between the time that one heel strikes the ground and the time the same heel strikes the ground again (see **Video Link 44.2**). The gait cycle begins when the forward foot hits the ground (heel strike or initial contact). During the stance phase, the stance leg supports all or most of the weight. At the end of the stance phase, there is a push off (toe-off or preswing phase) after which the leg swings forward to contact the ground again. The stance period is divided into four phases: initial contact, loading, midstance, and terminal stance. The swing phase is also divided into four parts: preswing, initial swing, midswing, and terminal swing. The functional task during the stance phase is to bear weight; the functional task during swing phase is to advance the limb. Periods of single limb support (one foot on the ground) alternate with periods of double limb support (two feet on the ground).

Various parameters are used to measure and characterize gait, including gait velocity, stride time, step time, stride length, step width, and step length. Stride length is the distance covered by a gait cycle; step length is the distance covered by the swing phase of one leg. When walking, at least one foot is in contact with the ground at all times, and there are two periods of double limb support. When there is no longer a moment in time when both limbs are in contact with the ground, walking has become running. A typical adult walking comfortably on a level surface walks at a velocity of about 80 m/min, taking about 113 steps per minute, with a stride length of 1.41 m. About 60% of the gait cycle is spent in stance, 40% in swing, and 10% in double limb support. Periods of double limb increase quickly to compensate for instability.

The body’s center of mass is located just anterior to the S2 vertebral body. An efficient gait minimizes the displacement of the center of mass by rotating and tilting the pelvis and flexing and extending the various joints involved. Gait disturbances that increase the normal displacement of the center of mass are less efficient and require an increased expenditure of energy. Patients typically compensate by walking more slowly and employing compensatory maneuvers to regain lost efficiency. In addition to the increased energy requirement, abnormal gaits increase the risk of falling and the susceptibility to biomechanical injury.

EXAMINATION OF GAIT

The first step in analyzing gait is to check the width of the base, or the step width. The wider the base, the better the balance, and spreading the feet farther apart is the first compensatory effort in most gait disorders. Under normal circumstances, the medial malleoli pass within about 2 in of each other during the stride phase, a narrow and well-compensated gait. Any spread more than this may signal some problem with gait or balance.

The forefoot on each side should clear the ground to about the same degree; asymmetry of toe lift may be the earliest evidence of footdrop. A shortened stride length may be early evidence of bifrontal or extrapyramidal disease. Excessive movement of the hips may occur with any process causing proximal muscle weakness. Note the reciprocal arm swing; a decreased swing on one side is sometimes an early indicator of hemiparesis or hemiparkinsonism. Watch the hands for tremor or chorea.

Tandem walking stresses the gait and balance mechanisms even further. Elderly patients may have difficulty with tandem gait because of obesity or deconditioning. In relatively young patients with a low likelihood of neurologic disease, a quick and effective substitute for the Romberg is simply to have the patient close her eyes while walking tandem. This is a difficult maneuver and has high value as a screening test. Having the patient walk briskly and then stop abruptly on command, or make quick turns, first in one direction and then in the other, may bring out ataxia and incoordination not noticeable on straightaway walking. Rather than turning easily, the patient may have to turn in small, staccato steps. The patient may be asked to walk sideways, backward, and overstep, or cross one foot over the other. Having the patient walk on heels and toes may bring out weakness of dorsiflexion or plantar flexion. An excellent screening test is to have the patient hop on either foot. This simultaneously assesses lower-extremity strength, especially of the gastrosoleus, plus balance functions. Individuals who can hop adroitly on either foot are unlikely to have significant neurologic disease. Note if the patient is able to maintain balance with a sudden push or pull backward, forward, or to the side. This can be done by gently tapping the upper chest or pulling on the shoulders while standing behind the patient, reassuring the patient, “I will catch you if you begin to fall.” Note whether the patient has any obvious orthopedic limitations, such as a varus deformity of the knee, genu recurvatum, pelvic tilt, or any other abnormalities. Also note whether there is any difficulty rising from a chair or initiating gait.

Note posture, arm swing, extraneous movements, step height, and any side to side lurching.

ABNORMAL GAITS

A nosology has been suggested that classifies abnormal gait syndromes into low-, mid-, and high-level disorders. Low-level disorders are due to peripheral motor or sensory abnormalities; mid-level disorders include hemiplegic, paraplegic, cerebellar ataxic, parkinsonian, choreic, and dystonic disorders. Highest-level disorders include cautious gait, subcortical and frontal disequilibrium, isolated gait ignition failure, frontal gait disorder, and psychogenic gait disorder (PGD). Description of the clinical semiology of gait disorders continues to be the most common approach.

Cerebellar Ataxia

The gait of cerebellar disease is caused by involvement of the coordinating mechanisms in the cerebellum and its connecting systems **(Video 44.1)**. The only sign of mild ataxia may be the inability to walk tandem. Sudden stopping or turning may bring out a stagger. With more severe disease, there is a clumsy, staggering, unsteady, irregular, lurching, titubating, and wide-based gait, and the patient may sway to either side, back, or forward. Leg movements are erratic, and step length varies unpredictably. The patient may compensate by avoiding periods of single limb support, creating a shuffling gait. The patient is unable to walk tandem or follow a straight line on the floor. There may be tremors and oscillatory movements involving the entire body.

Video 44.1 Composite video demonstrating cerebellar ataxia, sensory ataxia in a patient with sensory neuropathy, spastic gait with pronounced scissoring in siblings with hereditary spastic paraplegia, gait apraxia in a patient with normal pressure hydrocephalus, steppage gait due to dense bilateral foot drops in a patient with Charcot-Marie-Tooth disease, and hemiparetic gait following stroke. (Cerebellar ataxia video courtesy John C. Pearson, PhD, and Thomas Mathews, MD, Neurological Teaching Videos, Wright State University Boonshoft School of Medicine.)

Ataxia of the lower extremities when tested separately usually accompanies cerebellar gait ataxia, except when disease is limited to the vermis (see below). With a lesion of the cerebellar vermis, the patient will exhibit a lurching, staggering gait, but without laterality, the ataxia will be as marked toward one side as the other. Cerebellar ataxia is present with eyes both open and closed; it may increase slightly with eyes closed, but not so markedly as in sensory ataxia. A gait resembling cerebellar ataxia is seen in acute alcohol intoxication. With a cerebellar hemispheric lesion, the patient will stagger and deviate toward the involved side. In disease localized to one cerebellar hemisphere or in unilateral vestibular disease, there is persistent swaying or deviation toward the abnormal side. As the patient attempts to walk a straight line or to walk tandem, she deviates toward the side of the lesion. Walking a few steps backward and forward with eyes closed may bring out “compass deviation” or a “star-shaped gait” (see Chapter 17). When attempting to walk a fixed circle around a chair, clockwise then counterclockwise, the patient will tend to fall toward the chair if it is on the side of the lesion, or to spiral out away from the chair if on the opposite side.

Either unilateral cerebellar or vestibular disease may cause turning toward the

side of the lesion on the Unterberger-Fukuda stepping test (Chapter 17). For all the tests that bring out deviation in one direction, other findings must be used to differentiate between vestibulopathy and a cerebellar hemispheric lesion. Unilateral ataxia may be demonstrated by having the patient attempt to jump on one foot, with the eyes either open or closed. The patient with bilateral vestibular disease may seek to minimize head movement during walking, holding the head stiff and rigid; having the patient turn the head back and forth during walking may bring out ataxia. Cerebellar gait ataxia is common in multiple sclerosis (MS), alcoholic cerebellar degeneration, cerebellar tumors, stroke, and cerebellar degenerations. With alcoholic cerebellar degeneration, pathology is restricted to the vermis. Nystagmus, dysarthria, and appendicular ataxia, even of the legs, are typically absent.

Sensory Ataxia

Sensory ataxia occurs when the nervous system is deprived of the sensory information, primarily proprioceptive, necessary to coordinate walking **(Video 44.1)**. Deafferentation may result from disease of the posterior columns (e.g., tabes dorsalis or subacute combined degeneration) or disease affecting the peripheral nerves (e.g., sensory peripheral neuropathy). The term “spinal ataxia” is sometimes used, but the pathology is not always in the spinal cord. The patient loses awareness of the position of the lower extremities in space, or even of the body as a whole, except as provided by the visual system. The patient is extremely dependent on visual input for coordination. When deprived of visual input, as with eyes closed or in the dark, the gait deteriorates markedly. The difference in walking ability with and without visual input is the key feature of sensory ataxia. If the condition is mild, locomotion may appear normal when the patient walks eyes open; more commonly, it is wide based and poorly coordinated.

The term “steppage gait” refers to a manner of walking in which the patient takes unusually high steps. Sensory ataxia is one of the causes of a steppage gait. The patient takes a high step, throws out her foot, and slams it down on the floor in order to increase the proprioceptive feedback. The heel may land before the toe, creating an audible “double tap.” An additional sound effect may be the tapping of a cane, creating a “slam, slam, tap” cadence. The sound effects may be so characteristic that the trained observer can make the diagnosis by listening to the footfalls. The patient with sensory ataxia watches her feet and keeps her

eyes on the floor while walking. With eyes closed, the feet seem to shoot out, the staggering and unsteadiness are increased, and the patient may be unable to walk. There is less reeling and lurching in sensory ataxia than with a comparable degree of cerebellar ataxia. The difficulty is even worse walking backward, because the patient cannot see where she is going. The patient with bilateral footdrops, however, also has a steppage gait and a double tapping sound (see "Steppage Gait" below). For a striking illustration of a tabetic gait in sequential photographs by Eadweard Muybridge in 1887, see Lanska and Goetz.

In all of these tests, sensory ataxia can be differentiated from predominantly cerebellar ataxia by accentuation of the difficulty with eyes closed; and unilateral cerebellar or vestibular disease from vermis involvement by laterality of unsteadiness. Ropper and Samuels point out the similarity of the wide based, stamping gait of sensory ataxia to film depictions of Frankenstein.

The Gait of Spastic Hemiparesis

The gait of spastic hemiparesis may be caused by a lesion interrupting the corticospinal pathways to one-half of the body, most commonly stroke. The patient stands with a hemiparetic posture, arm flexed, adducted, and internally rotated, and leg extended. There is plantar flexion of the foot and toes, either due to foot dorsiflexion weakness or to heel cord shortening, rendering the lower extremity on the involved side functionally slightly longer than on the normal side, referred to as an equinus deformity. (A horse's foot is digitigrade; horses stand on the tips of their toes. The human foot is normally plantigrade, with the whole plantar surface in contact with the ground. The human foot with a footdrop or shortened heel cord resembles a horse's, hence the term.) When walking, the patient holds the arm tightly to the side, rigid and flexed, extends it with difficulty, and does not swing it in a normal fashion **(Video 44.1)**. The leg is held stiffly in extension and the patient drags or shuffles the foot and scrapes the toes. With each step, the pelvis tilts upward on the involved side to aid in lifting the foot off the floor (hip hike), and during swing phase, the entire extremity sweeps around in a semicircle from the hip (circumduction). The stance phase is shortened because of weakness, and the swing phase shortened because of spasticity and slowing of movement. The sound produced by the scraping of the toes, as well as the wear of the shoe at the toe, may be quite characteristic. The patient is able to turn toward the paralyzed side more easily than toward the normal side. Loss of normal arm swing and slight circumduction

of the leg may be the only gait abnormalities in very mild hemiparesis.

Spastic Gait, Scissoring

This gait pattern occurs in patients who have severe spasticity of the legs. It occurs in patients who have congenital spastic diplegia (Little's disease, cerebral palsy) and related conditions and in chronic myelopathies due to conditions such as MS and cervical spondylosis. It is essentially a bilateral hemiplegic gait affecting the legs. There is characteristic tightness of the hip adductors causing adduction of the thighs, so that the knees may cross, one in front of the other, with each step (scissors gait). The patient walks on an abnormally narrow base, with a stiff shuffling gait, dragging both legs and scraping the toes **(Video 44.1)**. The steps are short and slow; the feet seem to stick to the floor. There may be a marked compensatory sway of the trunk away from the side of the advancing leg. Swaying and staggering may suggest an element of ataxia, but usually there is no true loss of coordination. The shuffling, scraping sound—together with worn areas at the toes of the shoes—are characteristic. The equinus position of the feet and heel cord shortening often cause the patient to walk on tiptoe.

The Spastic-Ataxic Gait

Some neurologic disorders cause involvement of both the corticospinal and the proprioceptive pathways (e.g., combined system disease due to vitamin B_{12} deficiency, or MS), resulting in a gait that has features of both spasticity and ataxia. The relative proportion of each abnormality depends on the particulars of the case. The ataxic component may be either cerebellar or sensory. In vitamin B_{12} deficiency, it is predominantly sensory; in MS, both components may be present.

The Parkinsonian Gait

The gait in most akinetic-rigid, parkinsonian syndromes is characterized by rigidity, bradykinesia, and loss of associated movements. The patient is stooped, with head and neck forward and knees flexed; the upper extremities are flexed at the shoulders, elbows, and wrists, but the fingers are usually extended (Figure

30.1). The gait is slow, stiff, and shuffling; the patient walks with small, mincing steps. Other features include involuntary acceleration (festination), decreased arm swing, en bloc turning, start hesitation, and freezing when encountering obstacles such as doorways **(Video 30.2)**.

The patient loses balance easily with a gentle shove or pull. Difficulty walking may be one of the earliest symptoms of the disease. The gait of Parkinson's disease is further described in Chapter 30. The same gait disorder can occur with any condition causing parkinsonism, such as drug side effects. Gait difficulty and a tendency to fall is particularly prominent in progressive supranuclear palsy (PSP).

Frontal Lobe Gait Disorders

A number of gait disorders have been ascribed to dysfunction of the frontal lobes. Lesions of the frontal lobe, or of the frontal lobe connections with the basal ganglia and cerebellum, may lead to a gait disorder characterized by a slightly flexed posture, short, shuffling steps, a widened base, and an inability to integrate and coordinate lower-extremity movements to accomplish normal ambulation. There is particular difficulty with starts and turns. Some of these are poorly understood and the relationship between them unclear. In some, the frontal lobe "dysfunction" has been attributed to normal aging. Many terms have been used, which refer to more or less the same phenomenon, including gait apraxia, frontal disequilibrium or ataxia, Bruns' apraxia/ataxia, magnetic gait, and lower half/body or vascular (arteriosclerotic) parkinsonism. Some of the gait disorders often included under this rubric include gait apraxia, the gait of normal pressure hydrocephalus (NPH), marche à petits pas, and the cautious (senile) gait.

Marche à Petits Pas

The marche à petits pas (walk of little steps) gait resembles that of parkinsonism but lacks the rigidity and bradykinesia. Locomotion is slow, and the patient walks with very short, mincing, shuffling, and somewhat irregular footsteps. The length of the step may be less than the length of the foot. There is often a loss of associated movements. This type of gait may be seen in normal elderly persons but also occurs in patients who have diffuse cerebral hemispheric dysfunction, particularly involving the frontal lobes. It may also occur as part of the syndrome

of NPH and in other types of hydrocephalus. The same gait disturbance is typical of multi-infarct dementia or lacunar state. In some patients with marche à petits pas, there are bizarre movements such as dancing or hopping. There may be generalized weakness of the lower extremities or of the entire body, with the patient fatiguing easily.

Gait Apraxia

Apraxia of gait is the loss of the ability to use the legs properly in walking, without demonstrable sensory impairment, weakness, incoordination, or other apparent explanation. The term has been criticized as not being loss of a learned skill. Gait apraxia is seen in patients with extensive cerebral lesions, especially of the frontal lobes. It is a common feature of NPH and may occur in frontal lobe neoplasms, Binswanger's disease, frontotemporal dementia, and other conditions that cause diffuse frontal lobe dysfunction. The patient cannot carry out purposeful movements with the legs and feet, such as making a circle or kicking an imaginary ball. In rising, standing, and walking, there is difficulty in initiating movement, and the automatic sequence of component movements is lost. The gait is slow and shuffling, with short steps. The patients may have the greatest difficulty initiating walking, making small, feeble, stepping movements with minimal forward progress. Eventually, the patient may be essentially unable to lift the feet from the floor, as if they were stuck or glued down, or may raise them in place without advancing them (magnetic gait, gait ignition failure, start hesitation). After a few hesitant shuffles, the stride length may increase (slipping clutch gait). When trying to turn, the patient may freeze (turn hesitation). The patient may be able to imitate normal walking movements when sitting or lying down, but eventually, even this ability is often lost. In addition, perseveration, hypokinesia, rigidity, and stiffness of the limb in response to contact (gegenhalten) are often seen. In the syndrome of isolated gait ignition failure, or freezing of gait, patients have difficulty starting to walk, but with continued stepping, the gait improves. They may again freeze when turning or encountering an obstacle.

Gait of Normal Pressure Hydrocephalus

Gait difficulty is typically the initial and most prominent symptom of NPH. The primary changes are slow walking, widened base, short steps, and shuffling, all

nonspecific features and natural compensations seen in patients with various gait disorders. It may range from mild, with only a cautious gait or difficulty with tandem walking, to severe, when unaided gait is impossible **(Video 44.1)**. It has been referred to as a gait apraxia. Features in common with other frontal lobe gait disorders include reduced velocity, stride length, and step height. NPH causes more widening of the base and outwardly rotated feet and is less responsive to external cues such as marching to a cadence or in step with the examiner. As with other disorders of frontal lobe function, patients may mimic stepping motions while supine or sitting.

Cautious (Senile) Gait

A cautious gait is seen in older patients who have no neurologic disease but are uncertain of their balance and postural reflexes. The gait takes on the characteristics seen when a healthy person walks on an icy surface: velocity slows, steps shorten, double support time increases, and the base widens. The foot-floor clearance is not decreased, and the patient does not shuffle. There is no difficulty with gait initiation, nor is there freezing.

There is an ostensible "multimodal" gait disorder in the elderly, attributed to aging of the vestibular system, impaired proprioceptive function caused by distal neuropathy in the elderly, and impaired vision. Baloh et al. found that age-related decreases in vestibular, visual, auditory, and somatosensation occurred in normal older people but were only weakly correlated with changes in gait and balance. White matter hyperintensities on magnetic resonance imaging were more highly correlated with changes in gait.

Steppage Gait

A problem arises with the use of the term "steppage," which means that the patient is lifting one or both legs high during her respective swing phases, as though she were walking up steps though the surface is level. Patients with footdrop may do this in order to help the foot clear the floor and avoid tripping. Patients with sensory ataxia, classically tabes dorsalis, may also lift the feet up high and then slap them down smartly to improve proprioceptive feedback. Because both of these gaits are "high stepping," both have been referred to as steppage gaits, but the causes and mechanisms are quite different.

A patient with footdrop has weakness of the dorsiflexors of the foot and toes.

When mild, this may be manifest only as a decrease in toe clearance during the swing phase. With more severe footdrop, the patient is in danger of tripping, and may drag the toe when she walks, characteristically wearing out the toe of her shoe. When footdrop is severe, the foot dangles uncontrollably during the swing phase. To compensate, she lifts the foot as high as possible, hiking the hip and flexing the hip and knee **(Video 44.1)**. The foot is thrown out and falls to the floor, toe first. The touching of the toe, followed by the heel creates a "double tap" that has a different sound than the heel first double tap of sensory ataxia. The patient is unable to stand on her heel, and when standing with her foot projecting over the edge of a step, the forefoot drops. The footdrops and steppage gait may be unilateral or bilateral. Common causes of unilateral footdrop and steppage gait include peroneal nerve palsy and L5 radiculopathy. Causes of bilateral footdrop and steppage gait include amyotrophic lateral sclerosis, Charcot-Marie-Tooth disease and other severe peripheral neuropathies, and certain forms of muscular dystrophy. In severe polyneuropathies, the steppage gait may have components of both sensory ataxia and footdrop.

The Myopathic (Waddling) Gait

Myopathic gaits occur when there is weakness of the hip girdle muscles, most often due to myopathy and most characteristically due to muscular dystrophy. If the hip flexors are weak, there may be a pronounced lordosis (Chapter 27). The hip abductor muscles are vital in stabilizing the pelvis, while walking. Trendelenburg's sign is an abnormal drop of the pelvis on the side of the swing leg because of hip abductor weakness (Chapter 27). When the weakness is bilateral, there is an exaggerated pelvic swing that results in a waddling gait. The patient walks with a broad base, with an exaggerated rotation of the pelvis, rolling or throwing the hips from side to side with every step to shift the weight of the body. In the extreme forms, this gait pattern has a bizarre appearance. The patient walks with a pronounced waddle, shoulders thrown back and pelvis thrust forward (see **Video Link 44.3**). This form of gait is particularly common in facioscapulohumeral muscular dystrophy. The myopathy patient has marked difficulty climbing stairs, often needing to pull herself up with the hand rail. Patients also have difficulty going from a lying to standing position without placing the hands on the knees and hips to push themselves up (Gower's sign, Figure 29.4). For a vintage motion picture showing a myopathic gait with calf pseudohypertrophy, toe walking, and Gower's sign in a boy with Duchenne's

dystrophy, see **Video Link 44.4**).

Hyperkinetic Gait

In conditions such as Sydenham's chorea, Huntington's disease, other forms of transient or persistent chorea, athetosis, and dystonia, the abnormal movements may become more marked while the patient is walking, and the manifestations of the disease more evident. Walking may accentuate not only the hyperkinesias but also the abnormalities of power and tone that accompany them. In Huntington's disease, the gait may be grotesque, dancing or prancing with abundant extraneous movement. It may look histrionic but is all too real **(Video Link 44.5)**. The distal movements in athetosis and the proximal movements in dystonia may be marked during walking, and in both, there are accompanying grimaces. Some movement disorders may present as a gait disturbance. Oppenheim termed the walking pattern in dystonia musculorum deformans "dromedary" because of the exaggerated lumbar lordosis and hip flexion.

Gaits Associated with Focal Weakness

In addition to the steppage gait that accompanies footdrop, weakness limited to other muscle groups may cause gait difficulties. With paralysis of the gastrocnemius and soleus muscles, the patient is unable to stand on the toes, and unable to push off to enter the swing phase with the affected leg. This may cause a shuffling gait that is devoid of spring. In weakness of the quadriceps muscle (e.g., femoral neuropathy), there is weakness of knee extension, and the patient can only accept weight on the affected extremity by bracing the knee. When walking, the knee is held stiffly, and there is a tendency to fall if the knee bends. The patient has less difficulty walking backward than forward. Lumbosacral radiculopathy may cause either footdrop or a unilateral Trendelenburg's gait, or both (see Chapters 27 and 47). In addition, the patient with acute radiculopathy may walk with a list or pelvic tilt, accompanied by flattening of the normal lumbar lordosis because of low back muscle spasm. The patient may walk with small steps; if the pain is severe, she may place only the toes on the floor, because dorsiflexion of the foot aggravates the pain. Patients commonly use a cane to avoid bearing weight on the involved leg.

Other Gait Disorders

Patients with unilateral thalamic lesions may have an inability to stand or sit out of proportion to weakness or sensory loss, with a tendency to fall backward or to the side contralateral to the lesion (thalamic astasia). A toppling gait refers to a tendency to totter and fall seen with brainstem and cerebellar lesions, perhaps due to a failure of righting reflexes and slow motor responses. Primary progressive freezing gait causes early and progressive gait freezing; it is not a distinct disorder but a syndrome with diverse causes.

NONNEUROLOGIC GAIT DISORDERS

Abnormalities of gait may occur for many other reasons and may be confused with neurologic disorders. An antalgic gait is one in which walking is disordered because of pain. Pain in a lower extremity, for whatever reason, causes a shortening of the stance phase on the involved limb as the patient seeks to avoid bearing weight. On more than one occasion, neurologic consultation has been requested in a patient who ultimately proved to have acute podagra or a hip fracture. An antalgic gait may also occur with peripheral neuropathy causing painful dysesthesias and allodynia of the feet. The patient walks as if on hot coals. Arthritis may cause difficulties with gait that are secondary to both pain and deformity. In pregnancy, ascites, and abdominal tumors, there may be a lordosis that resembles that seen in the muscular dystrophies. With dislocation of the hips, there may be waddling suggestive of a myopathic gait. A waddling gait is also typical of advanced pregnancy. Patients with severe orthostatic hypotension may complain of difficulty walking rather than dizziness. Marked stooping in ankylosing spondylitis may resemble parkinsonism. A gait abnormality due to generalized weakness may occur after a period of bed rest, or in wasting and debilitating diseases. It is characterized mainly by unsteadiness and the wish for support. The patient staggers and sways from side to side with a suggestion of ataxia. She moves slowly, and the knees may tremble. If the difficulty is marked, she may fall.

NONORGANIC (FUNCTIONAL) GAIT ABNORMALITIES

Derangements of station and gait on a nonorganic basis are common. Affected patients may be unable either to stand or walk, despite the absence of weakness or other objective neurologic abnormalities. Testing for strength, tone, and coordination is normal if carried out supine. Astasia-abasia is an old term meant largely to describe the gait in conversion disorders. PGD is preferable and includes gait changes related to depression, anxiety, and phobic states.

The gait may suggest the presence of a monoparesis, hemiparesis, or paraparesis, yet the limbs can be used in an emergency. In Keane's series of 60 patients, 23 mimicked paresis and most of the remainder had various ataxic or histrionic patterns. Several authors have noted that knee buckling is a common type of PGD. The histrionic type of PGD is nondescript and bizarre and may take any number of forms that do not conform to a specific organic disease pattern (see **Video Link 44.6**). The gait is irregular and variable, with a great deal of superfluous movement and often marked swaying from side to side. The patient may appear to be in great danger of falling but rarely does so, often demonstrating superb balance during the contortions. If she does fall, it is in a theatrical manner without injury. The bizarre movements often require better than normal coordination. The patient may balance on the stance leg for a prolonged period of time, while bringing up the swing leg with a great show of effort. The gait may show skating, hopping, dancing, or zigzag characteristics; the legs may be thrown out wildly, or there may be a tendency to kneel every few steps. Tremulousness of the extremities or tic-like or compulsive features may be present. Although the patient cannot walk forward, she may be able to walk backward or to one side or to run without difficulty. Better ability to propel a wheeled, swivel chair than to walk ("chair test") favors PGD. In most patients with PGD, the similarity to neurologic disease is slight. Hyperkinetic gait disorders are most likely to be confused with functional conditions.

The term astasia-abasia originated in an 1888 monograph by Blocq, and the condition is sometimes referred to as Blocq's syndrome. Blocq described patients who were able to jump, or walk on all fours, but unable to stand upright (astasia) or to walk (abasia). There is normal lower-extremity function when recumbent, yet an inability to walk. This same pattern can occur in lesions involving the cerebellar vermis, such as alcoholic cerebellar degeneration or medulloblastoma, and in frontal lobe disorders. Astasia-abasia is sometimes used to refer to any inability to either stand or walk normally but generally refers to a histrionic and dramatic gait disturbance with wild lurching and near falls.