CHAPTER 2

SIGNS OF CENTRAL NERVOUS SYSTEM DISORDERS

Disorders of the brain and the spinal cord – the two major components of the central nervous system (CNS) – typically cause readily recognizable combinations of paresis, sensory loss, visual deficits, and neuropsychologic disorders (Box 2.1). Such signs of CNS disorders differ from those of the peripheral nervous system (PNS) and both differ from the signs of psychogenic disorders. Neurologists formulate their preliminary diagnosis and often initiate treatment on the basis of the patient's history and the examination, but if results of investigations – such as laboratory testing or magnetic resonance imaging (MRI) – contradict the initial clinical impression, they will usually revise or at least reconsider it.

SIGNS OF CEREBRAL HEMISPHERE LESIONS

Hemiparesis, usually accompanied by changes in reflexes and muscle tone, is one of neurology's most prominent and reliable signs. Damage to the *corticospinal tract*, also called the *pyramidal tract* (Fig. 2.1), in the cerebrum or brainstem above (rostral to) the decussation of the pyramids, causes contralateral hemiparesis (Box 2.2) with weakness of the arm and leg – and, if the lesion is high enough, the lower face – opposite the side of the lesion. Damage to this tract within the spinal cord causes *ipsilateral* arm and leg or only leg paresis, but no face paresis.

The division of the motor system into upper and lower motor neurons is a basic construct of clinical neurology. During the corticospinal tract's entire path from the cerebral cortex to the motor cranial nerve nuclei and the anterior horn cells of the spinal cord, this tract consists of *upper motor neurons* (*UMNs*) (Fig. 2.2). The anterior horn cells, which are part of the PNS, begin the *lower motor neuron* (*LMN*). Cerebral lesions that damage the corticospinal tract cause *signs of UMN injury* (Figs. 2.2–2.5):

- Paresis with muscle spasticity
- Hyperactive deep tendon reflexes (DTRs)
- Babinski signs.

In contrast, PNS lesions, including motor neuron diseases (diseases of the anterior horn cells) and disorders of nerves (neuropathy), cause *signs of LMN injury*:

- Paresis with muscle flaccidity and atrophy
- Hypoactive DTRs
- No Babinski signs.

Another indication of a cerebral lesion is loss of certain sensory modalities over one half of the body, i.e., *hemisensory loss* (Fig. 2.6). A patient with a cerebral lesion characteristically loses contralateral position sensation, two-point discrimination, and the ability to identify objects by touch (stereognosis). Neurologists often describe loss of those modalities as "cortical" sensory loss.

Pain sensation, a "primary" sense, is initially received by the thalamus, from which it is relayed to the cortex, limbic system, and elsewhere. Because the thalamus is situated above the brainstem but below the cerebral cortex, most patients with cerebral lesions still perceive painful stimuli. For example, patients with cerebral infarctions may be unable to specify a painful area of their body, but they will still feel the pain's intensity and discomfort (see Chapter 14).

Visual loss of the same half-field in each eye, *homonymous hemianopia* (Fig. 2.7), is a characteristic sign of a contralateral cerebral lesion. Other equally characteristic visual losses are associated with lesions involving the eye, optic nerve, or optic tract (see Chapters 4 and 12). Because they are situated far from the visual pathway, lesions in the brainstem, cerebellum, or spinal cord do not cause visual field loss.

Another conspicuous sign of a cerebral hemisphere lesion is *focal (partial)* or *focal-onset seizures* (see Chapter 10). In fact, the majority of focal seizures that alter awareness or induce psychomotor phenomena originate in the temporal lobe.

Signs of Damage of the Dominant, Nondominant, or Both Cerebral Hemispheres

Although hemiparesis, hemisensory loss, homonymous hemianopia, and focal seizures may result from lesions of either cerebral hemisphere, several neuropsychologic deficits are referable to either the dominant or nondominant hemisphere. Neurologists usually ask a patient's handedness when taking a history, but if this information is unavailable, because approximately 85% of people are right-handed, they assume with reasonable confidence that the left hemisphere serves as the dominant hemisphere.

Lesions of the dominant hemisphere may cause language impairment, *aphasia*, a prominent and frequently occurring neuropsychologic deficit (see Chapter 8). Because the corticospinal tract sits adjacent to the language centers, right hemiparesis often accompanies aphasia (see Fig. 8.1).

Lesions of the nondominant parietal lobe tend to produce one or more striking neuropsychologic disturbances (see Chapter 8). For example, patients may neglect or ignore left-sided visual and tactile stimuli (*hemiinattention*). They may fail to use their left arm and leg because they neglect their limbs rather than because of

BOX 2.1 Signs of Common CNS Lesions

Cerebral hemisphere*
- Hemiparesis with hyperactive deep tendon reflexes, spasticity, and Babinski sign
- Hemisensory loss
- Homonymous hemianopia
- Focal (partial) seizures
- Aphasia, hemi-inattention, and dementia
- Pseudobulbar palsy

Basal ganglia*
- Movement disorders: parkinsonism, athetosis, chorea, and hemiballismus
- Postural instability
- Rigidity

Brainstem
- Cranial nerve palsy with contralateral hemiparesis
- Internuclear ophthalmoplegia (MLF# syndrome)
- Nystagmus
- Bulbar palsy

Cerebellum
- Tremor on intention^
- Impaired rapid alternating movements (dysdiadochokinesia)^
- Ataxic gait
- Scanning speech

Spinal cord
- Paraparesis or quadriparesis
- Spasticity
- Sensory loss up to a "level"
- Bladder, bowel, and sexual dysfunction

*Signs contralateral to lesions
#MLF, Medial longitudinal fasciculus
^Signs ipsilateral to lesions

BOX 2.2 Signs of Common Cerebral Lesions

Either hemisphere*
- Hemiparesis with hyperactive deep tendon reflexes and a Babinski sign
- Hemisensory loss
- Homonymous hemianopia
- Focal seizure

Dominant hemisphere
- Aphasia: fluent, nonfluent, conduction, or isolation
- Gerstmann syndrome: acalculia, agraphia, finger agnosia, and left–right confusion
- Alexia without agraphia

Nondominant hemisphere
- Hemi-inattention
- Anosognosia
- Constructional apraxia

Both hemispheres
- Dementia
- Pseudobulbar palsy

*Signs contralateral to lesions

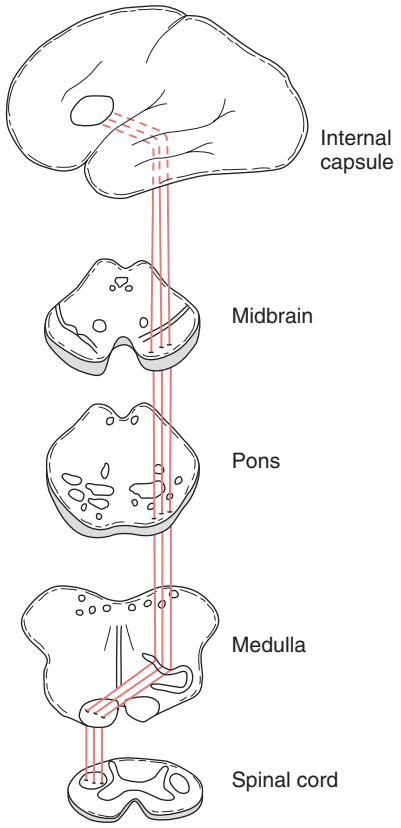

FIGURE 2.1 Each corticospinal tract originates in the cerebral cortex, passes through the internal capsule, and descends into the brainstem. The tracts cross in the pyramids, which are protuberances on the inferior portion of the medulla, to descend in the spinal cord mostly as the *lateral corticospinal tract*. The corticospinal tracts synapse with the *anterior horn cells* of the spinal cord, which give rise to peripheral nerves. Neurologists often call the corticospinal tract the *pyramidal* tract because it crosses in the pyramids. The *extrapyramidal* system, which modulates the corticospinal tract, originates in the basal ganglia and cerebellum, and remains within the brain.

paresis. When they have left hemiparesis, patients may not appreciate it (*anosognosia*). Many patients lose their ability to arrange matchsticks into certain patterns or copy simple forms (*constructional apraxia*, Fig. 2.8).

As opposed to signs resulting from unilateral cerebral hemisphere damage, bilateral cerebral hemisphere damage produces several important disturbances that psychiatrists are likely to encounter in their patients. One of them, *pseudobulbar palsy*, best known for producing emotional lability, results from bilateral *corticobulbar tract* damage (see Chapter 4). The corticobulbar tract, like its counterpart the corticospinal tract, originates in the motor cortex of the posterior portion of the frontal lobe. It innervates the brainstem motor nuclei, which in turn innervate the head and neck muscles. Traumatic brain injury (TBI), multiple cerebral infarctions (strokes), and frontotemporal dementia (see Chapter 7), are apt to strike the corticobulbar tract, as well as the surrounding frontal lobes, and thereby cause pseudobulbar palsy.

Damage to both cerebral hemispheres – from large or multiple discrete lesions, degenerative diseases, or metabolic abnormalities – also causes dementia (see Chapter 7). In addition, because CNS damage that causes dementia must be extensive and severe, it usually also produces at least subtle physical neurologic findings, such as hyperactive DTRs, Babinski signs, mild gait impairment, and frontal lobe release reflexes. However, many neurodegenerative illnesses that cause dementia, particularly

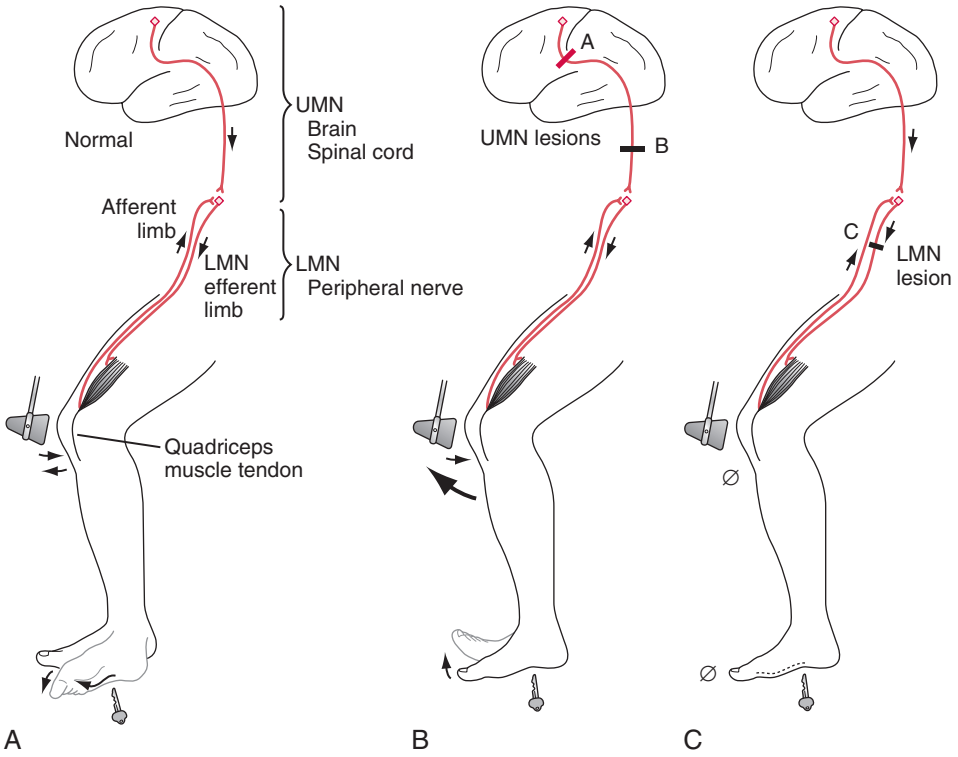

FIGURE 2.2 ■ A, Normally, when neurologists strike a patient's quadriceps tendon with a percussion hammer, the maneuver elicits a DTR. In addition, when they stroke the sole of the foot to elicit a plantar reflex, the big toe bends downward (flexes). B, When brain or spinal cord lesions injure the corticospinal tract, producing upper motor neuron (UMN) damage, DTRs react briskly and forcefully, i.e., DTRs are hyperactive. As another sign of UMN damage, the plantar reflex is extensor (a Babinski sign). C, In contrast, peripheral nerve injury causes lower motor neuron (LMN) damage, the DTR is hypoactive and the plantar reflex is absent.

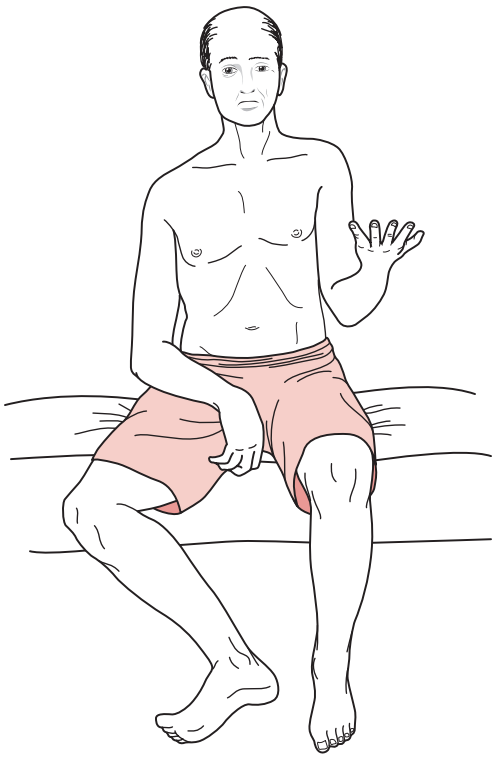

FIGURE 2.3 ■ This patient shows right hemiparesis with weakness of the right arm, leg, and lower face. The right-sided facial weakness causes the flat nasolabial fold; however, the forehead muscles remain normal (see Chapter 4 regarding this discrepancy). The right arm moves little, and the elbow, wrist, and fingers take on a flexed position; the right leg is externally rotated; and the hip and knee are flexed.

FIGURE 2.4 ■ When the patient stands up, his weakened arm retains its flexed posture. His right leg remains externally rotated, but he can walk by swinging it in a circular path. This maneuver is effective but results in *circumduction* or a *hemiparetic gait*.

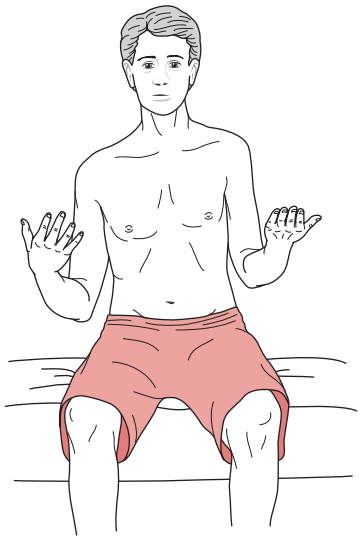

FIGURE 2.5 ■ Mild hemiparesis may not be obvious. To exaggerate it, the physician has asked this patient to extend both arms with his palms held upright, as though his outstretched hand were supporting a pizza box. His weakened arm sinks (drifts) and his palm turns inward (pronates). The imaginary pizza box would slide to his right. His arm drift and pronation represent a *forme fruste* of the posture seen with severe paresis (Fig. 2.3).

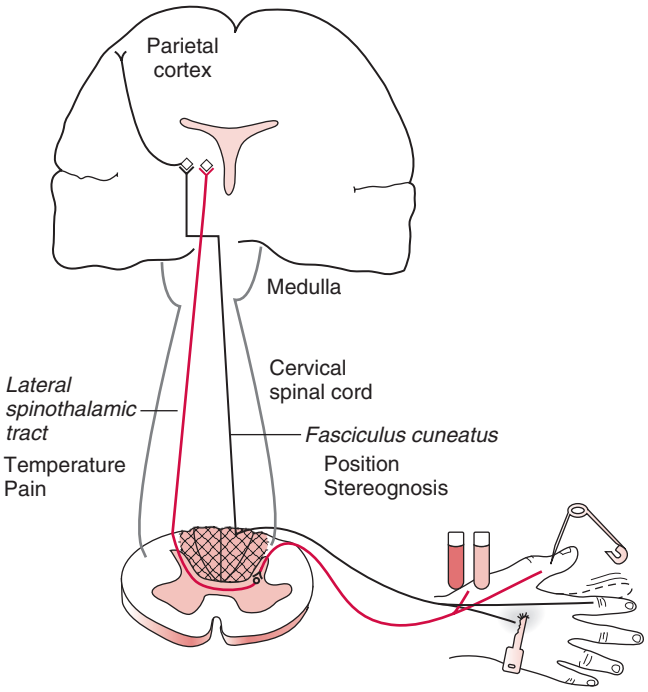

FIGURE 2.6 ■ Peripheral nerves carry pain and temperature sensations to the spinal cord. After a synapse, these sensations cross and ascend in the *contralateral lateral spinothalamic tract* (*pink*) to terminate in the thalamus. From there, tracts relay the sensations to the limbic system, reticular activating system, and other brainstem regions as well as the cerebral cortex. In parallel, the peripheral nerves also carry position and vibration sense and stereognosis to the *ipsilateral fasciculus cuneatus* and *fasciculus gracilis*, which together constitute the spinal cord's *posterior columns* (*cross-hatched*) (Fig. 2.15). Unlike pain and temperature sensation, these sensations ascend in the spinal cord via ipsilateral tracts (*black*). They cross in the decussation of the medial lemniscus, which is in the medulla, synapse in the thalamus, and terminate in the parietal cortex. (To avoid spreading blood-borne illnesses, examiners should use a disposable instrument when testing pain.)

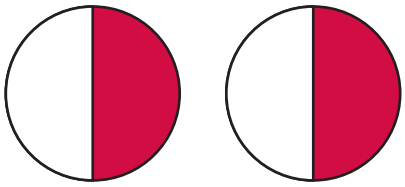

FIGURE 2.7 ■ In homonymous hemianopia, the same half of the visual field is lost in each eye. In this case, damage to the left cerebral hemisphere has caused a right homonymous hemianopia. This sketch portrays visual field loss, as is customary, from the patient's perspective; the colored area represents the defect (see Figs. 4.1 and 12.7).

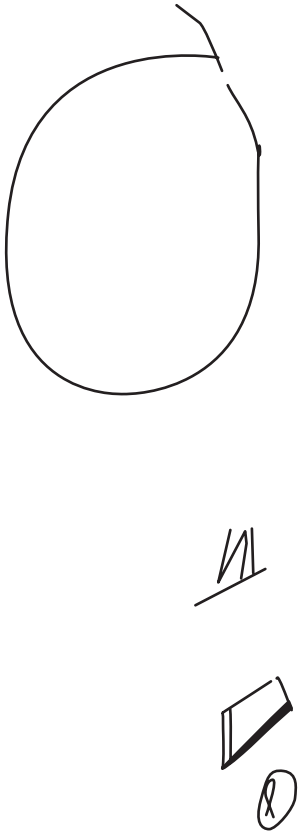

FIGURE 2.8 ■ A patient showing constructional apraxia from a right parietal lobe infarction was unable to complete a circle (*top figure*), draw a square on request (*second figure*), or even copy one (*third figure*). She spontaneously tried to draw a circle and began to retrace it (*bottom figure*). Her constructional apraxia consists of rotation of the forms, perseveration of certain lines, and the incompleteness of the second and lowest figures. In addition, the figures tend toward the right-hand side of the page, which indicates that she has neglect of the left-hand side of the page, i.e., left hemi-inattention (see Chapter 8).

Alzheimer disease, do not cause overt findings such as hemiparesis.

While certainly not peculiar to cerebral lesions and even typically absent in early Alzheimer disease, gait impairment is a crucial neurologic finding. Because walking requires intact and well-integrated strength, sensation, and coordination, testing the patient's gait is the single most reliable assessment of a patient's noncognitive neurologic function. Gait impairment constitutes the primary physical component of the subcortical dementias, such as vascular dementia, dementia with Lewy

TABLE 2.1 Gait Abnormalities Associated With Neurologic Disorders

Gait	Associated Illness	Figure
Apraxic	Normal pressure hydrocephalus	7.10
Astasia-abasia	Psychogenic disorders	3.4
Ataxic	Cerebellar damage	2.13
Festinating (*marche à petits pas*)	Parkinson disease	18.9
Hemiparetic/ hemiplegic	Strokes, congenital injury (cerebral palsy)	
• Circumduction		2.4
• Spastic hemiparesis		13.4
Diplegic	Congenital injury	13.3
Steppage	Tabes dorsalis (CNS syphilis)	2.20
Waddling	Duchenne dystrophy and other myopathies	6.4

bodies disease, and Parkinson disease dementia (see Chapter 7). Several distinct gait abnormalities are clues to specific neurologic disorders, such as normal pressure hydrocephalus (Table 2.1). As a general rule, slow gait speed, e.g., 0.7 m/sec or less, is associated with an increased risk of dementia, stroke, falls, disability, hospitalization, and death.

SIGNS OF BASAL GANGLIA LESIONS

The basal ganglia, located subcortically in the cerebrum, consist of the caudate and putamen (together constituting the *striatum*), globus pallidus, substantia nigra; and subthalamic nucleus (corpus of Luysii) (see Fig. 18.1). They give rise to the extrapyramidal motor system, which modulates the corticospinal (pyramidal) tract. It controls muscle tone, regulates motor activity, and generates postural reflexes. Its efferent fibers play on the cerebral cortex, thalamus, and other CNS structures. Because its efferent fibers are confined to the brain, the extrapyramidal tract does not act directly on the spinal cord or LMNs.

Signs of basal ganglia disorders include a group of fascinating, often dramatic, *involuntary movement disorders* (see Chapter 18):

- *Parkinsonism* is the combination of resting tremor, rigidity, bradykinesia (slowness of movement) or akinesia (absence of movement), and postural abnormalities. It usually results from Parkinson disease and related neurodegenerative illnesses, exposure to dopamine receptor-blocking antipsychotic medications, or toxins.
- *Athetosis* is the slow, continuous, writhing movement of the fingers, hands, face, and throat. Kernicterus or other perinatal basal ganglia injury usually causes it.
- *Chorea* is intermittent, randomly located, jerking of limbs and the trunk. The best-known example occurs in *Huntington* disease (previously called "Huntington chorea"), in which the caudate nuclei characteristically atrophy.
- *Hemiballismus* is the intermittent flinging of the arm and leg of one side of the body. It is classically associated with small infarctions of the contralateral subthalamic nucleus, but similar lesions in other basal ganglia may be responsible.

In general, when damage is restricted to the extrapyramidal system, patients have no paresis, DTR abnormalities, or Babinski signs – hallmarks of corticospinal (pyramidal) tract damage. More important, in many of these conditions, such as hemiballismus and athetosis, patients have no cognitive impairment or other neuropsychologic disorder. On the other hand, several conditions – such as Huntington disease, Wilson disease, and advanced Parkinson disease – affect the cerebrum as well as the basal ganglia. In them, dementia, depression, and psychosis frequently accompany involuntary movements (see Box 18.4).

With unilateral basal ganglia damage, signs develop in the contralateral limbs. For example, an infarction of the subthalamic nucleus causes contralateral hemiballismus, and degeneration of the substantia nigra causes contralateral parkinsonism ("hemiparkinsonism").

SIGNS OF BRAINSTEM LESIONS

The brainstem contains, among a multitude of structures, the cranial nerve nuclei, the corticospinal tracts, other "long tracts" that travel between the cerebral hemispheres and the limbs, and cerebellar afferent (inflow) and efferent (outflow) tracts. Combinations of cranial nerve and long tract signs, and the *absence* of signs of cerebral injury, such as visual field cuts and neuropsychologic deficits, indicate the presence and location of a brainstem lesion. For example, brainstem injuries cause *diplopia* (double vision) because of cranial nerve impairment, but visual acuity and visual fields remain normal because the visual pathway, which passes from the optic chiasm to the cerebral hemispheres, does not travel within the brainstem (see Fig. 4.1). Similarly, a right hemiparesis associated with a left third cranial nerve palsy localizes the lesion to the brainstem (particularly the left midbrain). Moreover, that pair of findings indicates that further examination will reveal neither aphasia nor dementia.

Several brainstem syndromes illustrate critical anatomic relationships, such as the location of the cranial nerve nuclei or the course of the corticospinal tract; however, none of them involves neuropsychologic abnormalities. Although each syndrome has an eponym, for practical purposes it is only necessary to identify the clinical findings and, if appropriate, attribute them to a lesion in one of the *three major divisions of the brainstem: midbrain*, *pons*, *or medulla* (Fig. 2.9). Whatever the localization, most brainstem lesions result from occlusion of a small branch of the basilar or vertebral arteries.

In the midbrain, where the oculomotor (third cranial) nerve fibers pass through the descending corticospinal

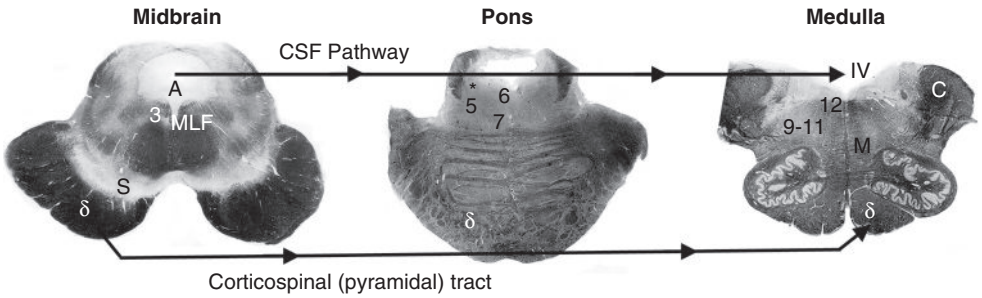

FIGURE 2.9 ■ Myelin stains of the three main divisions of the brainstem – midbrain, pons, and medulla – show several clinically important tracts, the cerebrospinal fluid (CSF) pathway, and motor nuclei of the cranial nerves. (*Midbrain*) The midbrain is identifiable by its distinctive silhouette and gently curved (pale, unstained in this preparation) substantia nigra (S). The aqueduct of Sylvius (A) is surrounded by the periaqueductal gray matter. Ventral to the aqueduct, near the midline, lie the oculomotor (3) and trochlear (not pictured) cranial nerve nuclei. The nearby MLF, which ascends from the pons, terminates in the oculomotor nuclei. The large, deeply stained cerebral peduncles, ventral to the substantia nigra, contain the corticospinal (pyramidal [Δ]) tract. Originating in the cerebral cortex, the corticospinal tract (Δ) descends ipsilaterally through the midbrain, pons, and medulla until it crosses in the medulla's pyramids to continue within the contralateral spinal cord. CSF flows downward from the lateral ventricles through the aqueduct of Sylvius into the fourth ventricle (IV), which overlies the lower pons and medulla. CSF exits from the fourth ventricle into the subarachnoid space. (Also see a functional drawing [Fig. 4.5], computer-generated rendition [Fig. 18.2], and sketch [Fig. 21.1].) (*Pons*) The pons (Latin, bridge) houses the trigeminal motor division (5), abducens (6), facial (7), and acoustic/vestibular (not shown) cranial nerve nuclei and, inferior and lateral to the fourth ventricle, the locus ceruleus (*). In addition to containing the descending corticospinal tract (Δ), the basilar portion of the pons (*basis pontis*) contains large crisscrossing cerebellar tracts. (Also see a functional drawing [Fig. 4.7] and an idealized sketch [Fig. 21.2].) (Medulla) The medulla (Latin, marrow), readily identifiable by the pair of unstained scallop-shaped inferior olivary nuclei, includes the cerebellar peduncles (C), which contain afferent and efferent cerebellar tracts; the corticospinal tract (Δ); and the floor of the fourth ventricle (IV). It also contains the decussation of the medial lemniscus (M), the nuclei for cranial nerves 9–11 grouped laterally and 12 situated medially, and the trigeminal sensory nucleus (not pictured) that descends from the pons to the cervical–medullary junction. (Also see a functional drawing [Fig. 2.10].)

tract, a single small infarction can damage both pathways. Patients with oculomotor nerve paralysis and contralateral hemiparesis typically have a lesion in their midbrain ipsilateral to the paretic eye (see Fig. 4.9). In an analogous situation, patients with an abducens (sixth cranial) nerve paralysis and contralateral hemiparesis have a lesion in the pons ipsilateral to the paretic eye (see Fig. 4.11).

Lateral medullary infarctions create a classic but complex picture, *the lateral medullary syndrome*. Patients have dysarthria because of paralysis of the ipsilateral palate from damage to cranial nerves IX through XI; ipsilateral facial numbness (*hypalgesia*) (Greek, decreased sensitivity to pain) because of damage to cranial nerve V, with contralateral anesthesia of the body (*alternating or crossed hypalgesia*) because of ascending spinothalamic tract damage; and ipsilateral ataxia because of inferior cerebellar peduncle dysfunction. They also have nystagmus and vertigo from damage to the vestibulocochlear nerve and ipsilateral Horner syndrome (ptosis, miosis, anhydrosis) due to interruption of sympathetic fibers. In other words, the most important elements of this syndrome consist of damage to three groups of nuclei (V, VIII, and IX–XI) and three white matter tracts (spinothalamic, sympathetic, and inferior cerebellar peduncle). Although the lateral medullary syndrome commonly occurs and provides an excellent example of clinical-pathologic correlation, physicians need not recall all of its pathology or clinical features; however, they should know that lower cranial nerve palsies accompanied by alternating hypalgesia, without cognitive impairment or limb paresis, result from a lesion in the lower brainstem (Fig. 2.10). They should also know that the lateral medullary syndrome causes bulbar palsy (see Chapter 4).

Nystagmus, repetitive jerk-like eye movements that are usually conjugate (i.e., affecting both eyes equally and simultaneously), is not peculiar to the lateral medullary syndrome, but rather may result from any type of injury to the brainstem's large vestibular nuclei. Nystagmus can be a manifestation of various disorders, including intoxication with alcohol, phenytoin (Dilantin), phencyclidine (PCP), or barbiturates; ischemia of the vertebrobasilar artery system; multiple sclerosis (MS); Wernicke–Korsakoff syndrome; or viral labyrinthitis. Among individuals who have ingested PCP, coarse vertical and horizontal (three directional or multidirectional) nystagmus characteristically accompanies an agitated delirium and markedly reduced sensitivity to pain and cold temperature. Unilateral nystagmus may be a component of *internuclear ophthalmoplegia*, which is usually a manifestation of MS or a small brainstem infarction (see Chapters 4 and 15).

SIGNS OF CEREBELLAR LESIONS

The cerebellum (*Latin*, diminutive of cerebrum) consists of two hemispheres and a central portion, the *vermis*. Each hemisphere controls coordination of the ipsilateral limbs, and the vermis controls coordination of "axial" or "midline structures": the head, neck, and trunk. Because the cerebellum controls coordination of the limbs on the same side of the body, it differs from the cerebrum wherein each hemisphere governs the contralateral body.

Another unique feature of the cerebellum is that when one hemisphere is damaged, the other will eventually assume the functions for both. In other words, although loss of one cerebellar hemisphere will temporarily cause

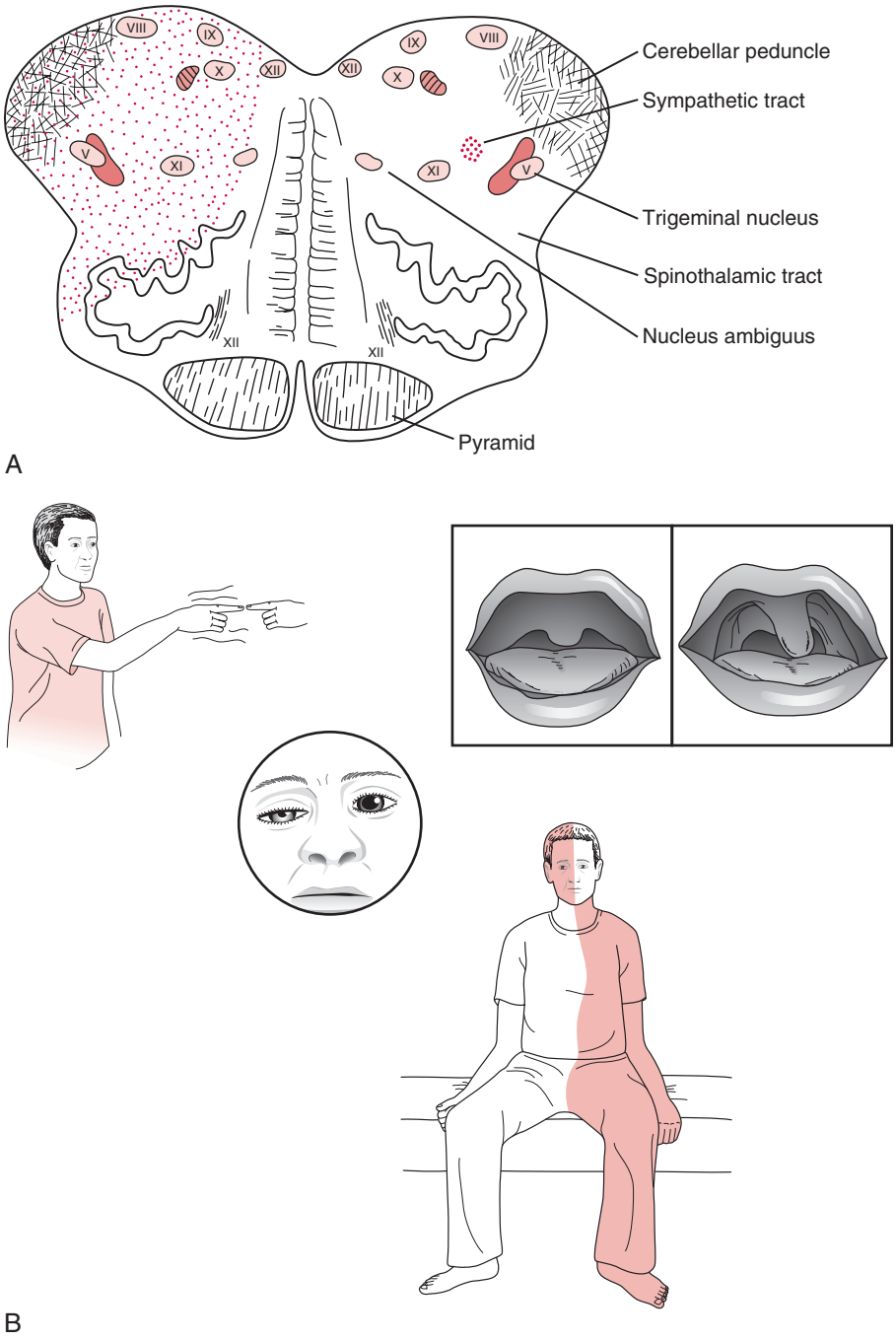

FIGURE 2.10 ■ A, An occlusion of the right posterior inferior cerebellar artery (PICA) or its parent artery, the right vertebral artery, has caused an infarction of the lateral portion of the right medulla (stippled). This infarction damages important structures: the inferior cerebellar peduncle, the spinal trigeminal nerve (V) sensory nucleus, the spinothalamic tract (which arose from the contralateral side of the body), the nucleus ambiguus (cranial nerves IX and X motor nuclei), and poorly delineated sympathetic fibers. However, this infarction spares medial structures: the corticospinal tract, medial longitudinal fasciculus (MLF), and hypoglossal nerve (XII) nucleus. B, Because he has sustained an infarction of his right lateral medulla, this patient has a right-sided Wallenberg syndrome. He has a right-sided Horner syndrome (ptosis and miosis) because of damage to the sympathetic fibers (also see Chapter 12). He has right-sided ataxia because of damage to the ipsilateral cerebellar tracts. He has an alternating or crossed hypalgesia: diminished pain sensation on the *right* side of his face, accompanied by loss of pain sensation on the *left* trunk and extremities (shaded). Finally, he has hoarseness and paresis of the right soft palate because of damage to the right nucleus ambiguus. Because of the right-sided palate weakness, the palate deviates upward toward his left on voluntary phonation (saying "ah") or in response to the gag reflex.

incapacitating ipsilateral incoordination, the patient's deficit lessens as the remaining hemisphere compensates almost entirely. For example, patients who lose one cerebellar hemisphere to a stroke or TBI typically regain their ability to walk, although they may never dance. Children who sustain such an injury are more resilient and often can learn to ride a bicycle and participate in athletic activities.

In addition to causing incoordination, cerebellar lesions cause subtle motor changes, such as muscle

hypotonia and pendular DTRs. However, cerebellar lesions do not cause paresis, hyperactive DTRs, or Babinski signs.

Although several technically sophisticated studies have shown that the cerebellum contributes to cognition and emotion, it does not play a discernible role in these functions in everyday endeavors. For example, lesions restricted to the cerebellum do not lead to dementia, language impairment, or other cognitive impairment. A good example is the normal intellect of children and young adults despite having undergone resection of a cerebellar hemisphere for removal of an astrocytoma (see Chapter 19).

On the other hand, several conditions damage the cerebrum as well as the cerebellum. For example, alcohol, phenytoin (Dilantin), lithium, and toluene may cause prominent ataxia and cognitive impairment.

For practical purposes, neurologists assess cerebellar function in tests of coordinated motor function. Thus, *intention tremor*, demonstrable in the finger-to-nose (Fig. 2.11) and heel-to-shin tests (Fig. 2.12), characterizes cerebellar dysfunction. This tremor is evident when the patient moves to a target but is absent when the patient rests. In a classic contrast, Parkinson disease causes a *resting tremor* that is present when the patient sits quietly and reduced or even abolished when the patient moves (see Chapter 18). Physicians should not confuse the neurologic term "intention tremor" with "intentional tremor," which would be a self-induced or psychogenic tremor.

Another sign of incoordination due to a cerebellar lesion is *dysdiadochokinesia*, impaired rapid alternating movements of the limbs. When asked to slap the palm and then the back of the hand rapidly and alternately on his or her own knee, for example, a patient with dysdiadochokinesia will do so with uneven force and irregular rhythm, and lose the alternating pattern.

Damage to either the entire cerebellum or the vermis alone causes incoordination of the trunk (*truncal ataxia*). This manifestation of cerebellar damage forces patients to place their feet widely apart when standing and leads to a lurching, unsteady, and wide-based pattern of walking (*gait ataxia*) (Table 2.1 and Fig. 2.13). A common example is the staggering and reeling of people intoxicated by alcohol. In addition, such cerebellar damage prevents people from walking heel-to-toe, i.e., performing "tandem gait." Another common example of ataxia occurs in individuals who have inherited genetic mutations that cause combinations of cerebellar and spinal cord degeneration. In several disorders, patients have abnormalities beyond the nervous system (Fig. 2.14). Extensive damage of the cerebellum causes *scanning speech*, a variety of dysarthria. Scanning speech, which reflects incoordination of speech production, is characterized by poor modulation, irregular cadence, and

FIGURE 2.11 ■ This young man has a multiple sclerosis plaque in the right cerebellar hemisphere. During the *finger-to-nose* test, his right index finger touches his nose and then the examiner's finger by following a coarse, irregular path. The oscillation in his arm's movement is an *intention tremor,* and the irregularity in the rhythm is *dysmetria.*

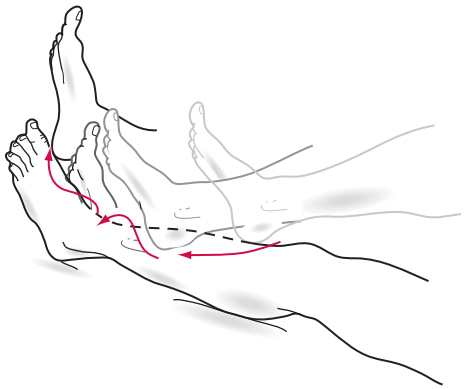

FIGURE 2.12 ■ In the *heel-to-shin test*, the patient with the right-sided cerebellar lesion in the previous sketch displays limb *ataxia* as his right heel wobbles when he pushes it along the crest of his left shin.

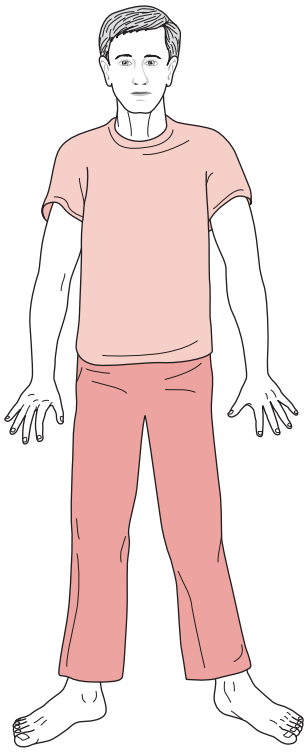

FIGURE 2.13 ■ Because this man has developed cerebellar degeneration from alcoholism, he has a typical *ataxic gait.* His stance is broad-based. His gait is unsteady, and he is uncoordinated.

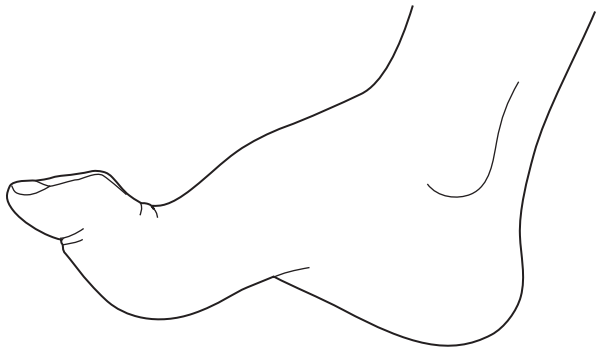

FIGURE 2.14 ■ The *pes cavus* foot deformity consists of a high arch, elevation of the dorsum, and retraction of the first metatarsal. When pes cavus occurs in families with childhood-onset ataxia and posterior column sensory deficits, it is a reliable sign of Friedreich ataxia, which is the most common hereditary ataxia in the United States and Europe.

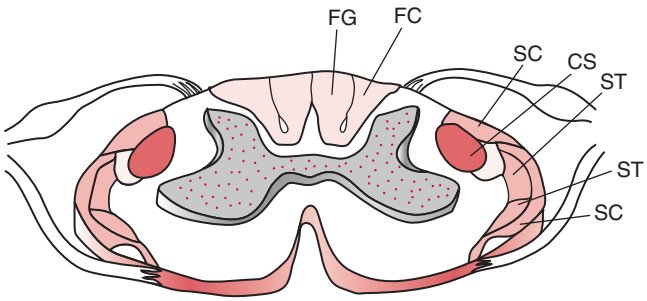

FIGURE 2.15 ■ In this sketch of the spinal cord, the centrally located gray matter is stippled. The surrounding white matter contains myelin-coated ascending and descending tracts. Clinically important ascending tracts are the spinocerebellar tracts (SC), the lateral spinothalamic tract (ST), and the posterior columns [fasciculus cuneatus (FC), from the upper limbs, and fasciculus gracilis (FG), from the lower limbs]. The most important descending tract is the lateral corticospinal (CS) tract.

inability to separate adjacent sounds. Physicians should be able to distinguish dysarthria – whether caused by cerebellar injury, bulbar or pseudobulbar palsy, or other neurologic disorder – from aphasia (see Chapter 8).

Before considering the illnesses that damage the cerebellum (see Section 2), physicians must appreciate that the cerebellum normally undergoes age-related changes that appear between ages 50 and 65 years in the form of mildly impaired functional ability and abnormal neurologic test results. For example, as people age beyond 50 years, they walk less rapidly and less sure-footedly. They begin to lose their ability to ride a bicycle and to stand on one foot while putting on socks. During a neurologic examination they routinely topple during tandem walking.

SIGNS OF SPINAL CORD LESIONS

The spinal cord's gray matter, which when viewed in the axial plane appears as a broad H-shaped structure in the center of the spinal cord, consists largely of neurons that transmit nerve impulses at one horizontal level. The spinal cord's white matter, composed of myelinated tracts that convey information in a vertical direction, surrounds the central gray matter (Fig. 2.15). This pattern – gray matter on the inside with white outside – is opposite that of the cerebrum. Interruption of the myelinated tracts causes most of the signs of spinal cord injury, which neurologists call "myelopathy."

The major *descending* pathway, entirely motor, is the *lateral corticospinal tract*.

The major *ascending* pathways, entirely sensory, include the following:

- *Posterior columns* (or *dorsal columns*), comprised of the *fasciculi cuneatus* and *gracilis*, carry position and vibration sensations to the thalamus.
- *Lateral spinothalamic tracts* carry temperature and pain sensations to the thalamus.
- *Anterior spinothalamic tracts* carry light touch sensation to the thalamus.
- *Spinocerebellar tracts* carry joint position and movement sensations to the cerebellum.

Spinal Cord Transection

If an injury severs the spinal cord, the *transection's* location – cervical, thoracic, or lumbosacral – determines the pattern of the ensuing motor and sensory deficits. Cervical spinal cord transection, for example, blocks all motor impulses from descending and sensory information from arising through the neck. This lesion causes paralysis of the arms and legs (*quadriparesis*) and, after 1–2 weeks, hyperactive DTRs, and Babinski signs. In addition, it prevents the perception of all limb, trunk, and bladder and bowel sensation. Similarly, a midthoracic spinal cord transection causes paralysis of the legs (*paraparesis*) with similar reflex changes, and sensory loss in the trunk and below (Fig. 2.16). In general, all spinal cord injuries disrupt bladder control and sexual function, which rely on delicate, intricate systems (see Chapter 16).

Another motor impairment attributable to spinal cord damage, whether from a specific lesion or a neurodegenerative illness, is pathologically increased muscle tone, which neurologists label *hypertonicity* or *spasticity*. It often creates more disability than the accompanying paresis. For example, because spasticity causes the legs to be straight, extended, and unyielding, patients tend to walk on their toes (see Fig. 13.3). Similarly, spasticity greatly limits the usefulness of patients' hands and fingers.

In a variation of the complete spinal cord lesion, when a penetrating injury severs only the lateral half of the spinal cord, neurologists refer to the injury as a spinal cord hemitransection. The lesion causes the classic *Brown–Séquard syndrome*, which consists of ipsilateral paralysis of limb(s) from corticospinal tract damage and loss of vibration and proprioception from dorsal column damage combined with loss of temperature and pain (hypalgesia) sensation in the opposite limb(s) from lateral spinothalamic tract damage (Fig. 2.17). In the vernacular of neurology, one leg is weak and the other is numb.

Even with devastating spinal cord injury, cerebral function is preserved. In a frequently occurring and tragic example, soldiers surviving a penetrating gunshot wound of the cervical spinal cord, although quadriplegic, retain intellectual, visual, and verbal facilities. Nevertheless, veterans and other individuals with spinal cord injuries

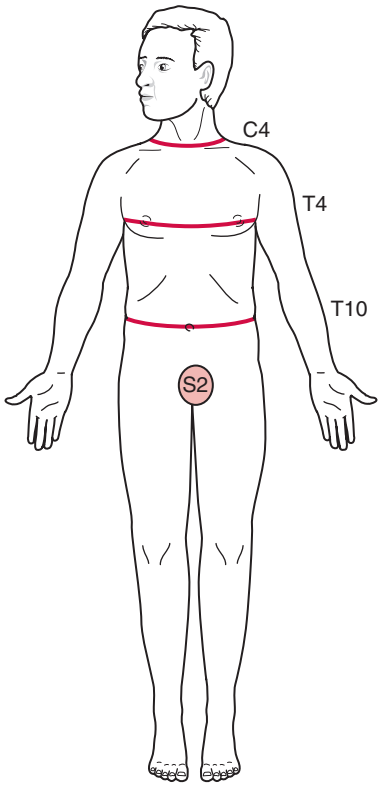

FIGURE 2.16 ■ In a patient with a spinal cord injury, the "level" of hypalgesia indicates the site of the damage. The clinical landmarks are C4, T4, and T10. C4 injuries cause hypalgesia below the neck; T4 injuries, hypalgesia below the nipples; T10 injuries, hypalgesia below the umbilicus.

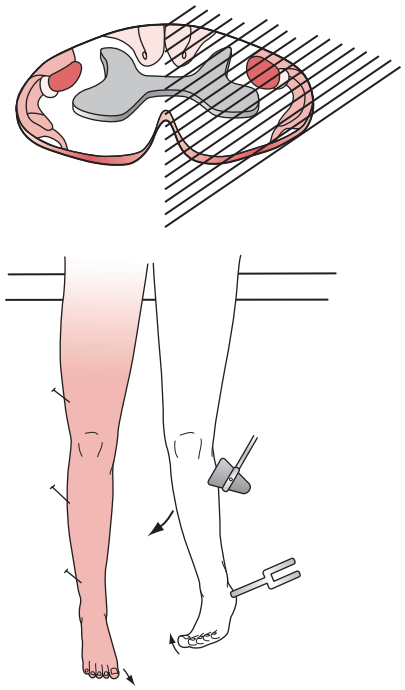

FIGURE 2.17 ■ In this case of hemitransection of the thoracic spinal cord from a knife wound, the patient shows the Brown–Séquard syndrome. Injury to the left lateral corticospinal tract results in the combination of left-sided leg paresis, hyperactive DTRs, and a Babinski sign; injury to the left posterior column results in impairment of left leg vibration and position sense. Most striking, injury to the left spinothalamic tract causes loss of temperature and pain sensation in the right leg. Loss of pain sensation contralateral to paresis is the signature of the Brown–Séquard syndrome.

often despair from isolation, lack of social support, and loss of their physical abilities.

Syringomyelia

A lesion that often affects only the cervical spinal cord consists of an elongated cavity, *syringomyelia* or simply a *syrinx* (Greek, *syrinx*, pipe or tube + *myelos* marrow). The syrinx occurs in the substance of the spinal cord, adjacent to its *central canal*, which is the thin tube running vertically within the gray matter. It usually develops, for unclear reasons, during adolescence. Traumatic intraspinal bleeding may cause a variety of syrinx, a *hematomyelia*. These conditions produce clinical findings that reflect their neuroanatomy (Fig. 2.18). In both cases, as the cavity expands, its pressure rips apart the lateral spinothalamic tract fibers as they cross from one to the other side of the spinal cord. It also presses on the anterior horn cells of the anterior gray matter. The expansion not only causes neck pain, but a striking loss in the arms and hands of pain and temperature sensation, muscle bulk, and DTRs. Because the sensory loss is restricted to patients' shoulders and arms, neurologists frequently describe it as *cape*- or *shawl-like*. Moreover, the sensory loss is characteristically restricted to loss of pain and temperature sensation because the posterior columns, merely displaced, remain functional.

NEUROLOGIC ILLNESSES

Several illnesses damage only specific ascending and descending spinal cord tracts (Fig. 2.19). The posterior columns – *fasciculus gracilis* and *fasciculus cuneatus* – seem particularly vulnerable. For example, tabes dorsalis (syphilis), combined system degeneration (vitamin B_{12} deficiency, see Chapter 5), and the spinocerebellar ataxias (SCAs) each damages the posterior columns alone or in combination with other tracts (also see Box 15.1). In these conditions, impairment of the posterior columns leads to a loss of position sense that prevents patients from being able to stand with their eyes closed (*Romberg's sign*). When they walk, this position sense loss produces an ataxic gait or possibly a *steppage gait* (Fig. 2.20).

In another example, the human T-lymphotropic virus type 1 (HTLV-1) infects the spinal cord's lateral columns. The infection, which is endemic in Caribbean islands, causes *HTLV-1 associated myelopathy* (or simply *HTLV-1 myelopathy*) in which patients develop spastic paraparesis that resembles MS. Perhaps more than in any other common myelopathy, the spasticity is disproportionately greater than the paresis.

Several toxic-metabolic disorders – some associated with substance abuse – damage the spinal cord. For example, nitrous oxide (N_2O), a gaseous anesthetic that may be inhaled as a drug of abuse by thrill-seeking

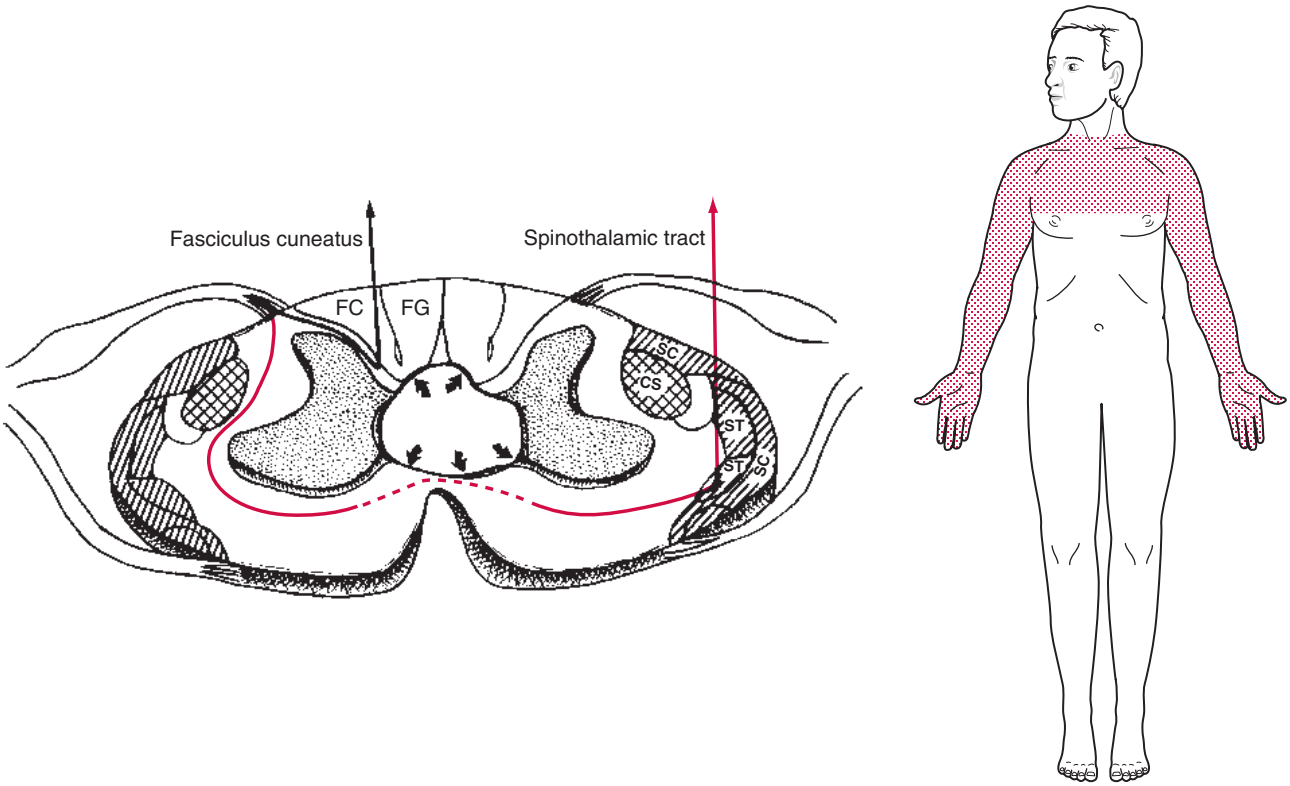

FIGURE 2.18 ■ *Left,* A syringomyelia (syrinx) is an elongated cavity in the spinal cord. Its expansion disrupts the lateral spinothalamic tract as it crosses, and compresses the anterior horn cells of the gray matter. It does not impair the function of the posterior columns and corticospinal tracts. *Right,* The classic finding is a shawl-like pattern of loss of pain and temperature sensation in the arms and upper chest (in this case, C4–T4) that is accompanied by weakness, atrophy, and areflexia in the arms.

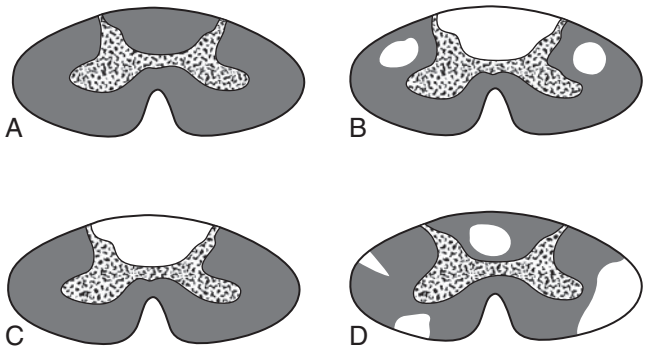

FIGURE 2.19 ■ A, A standard spinal cord histologic preparation stains normal myelin (white matter) black and leaves the central H-shaped column gray. B, In combined system degeneration (vitamin B_{12} deficiency), posterior column and corticospinal tract demyelination causes their lack of stain. C, In tabes dorsalis (tertiary syphilis), damage to the posterior column leaves them unstained. D, MS leads to asymmetric, irregular, demyelinated unstained plaques.

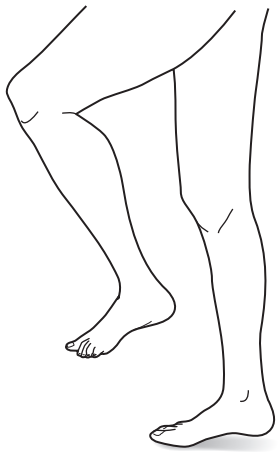

FIGURE 2.20 ■ The steppage gait consists of the patient's raising each knee excessively, as if perpetually climbing a staircase. This maneuver compensates for a loss of position sense by elevating the feet to ensure that they will clear the ground. Although the steppage gait is a classic sign of posterior column spinal cord damage from tabes dorsalis, peripheral neuropathies that impair position sense are a more frequent cause of this gait abnormality.

dentists, causes a pronounced myelopathy by inactivating vitamin B_{12} (see Chapter 5). Copper deficiency, often from excess consumption of zinc by food faddists or inadvertently ingested from excess denture cream, leads to myelopathy. Also, unless physicians closely monitor and replace vitamins and nutrients following gastric bypass surgery, patients remain at risk of developing myelopathy for up to several years after the surgery.

Most important, dementia accompanies myelopathy in several illnesses because of concomitant cerebral damage. Examples of this association include tabes dorsalis, vitamin B_{12} deficiency, AIDS, and, when disseminated throughout the cerebrum, MS.