

CHAPTER 22

Overview of the Motor System

Examination of motor functions includes the determination of muscle power, evaluation of muscle tone and bulk, and observation for abnormal movements. Examination of coordination and gait is closely related to the motor examination. Coordination is often viewed as a cerebellar function, but integrity of the entire motor system is essential for normal coordination and control of fine motor movements. Examination of cerebellar function is discussed in [Chapter 43](#). Station (standing) and gait (walking) are complex and involve much more than motor function; they are usually assessed separately from the motor examination ([Chapter 44](#)).

Both the peripheral and central nervous systems participate in motor activity, and various functional components have to be evaluated individually. Our motor systems move our bodies in space, move parts of the body in relation to one another, and maintain postures and attitudes in opposition to gravity and other external forces. All movements, except those mediated by the autonomic nervous system, are effected by contractions of striated muscles through the control of the nervous system.

LEVELS OF MOTOR ACTIVITY

The intricate organization of the motor system and its evolutionary development from the simple responses of unicellular organisms to the patterns of behavior of animals and man account for the complexity of motor function. From anatomic and functional standpoints, there are certain phylogenetic motor levels, or stages of development, which increase in complexity with evolution. In lower vertebrates, motor activities are effected through subcortical centers, but with the greater development of the cerebral cortex in higher mammals, some of these functions are significantly altered. The more primitive centers retain some of

their original functions, although modified by cortical control. They are not replaced but are incorporated into an elaborate motor system, subordinate to the cortex. The phylogenetically old and new systems work together, and the efficiency of each depends upon collaboration with the other.

The evolutionary development of motor function from simple to complex movements is duplicated to a certain extent in the maturation of motor skills in man. At the time of birth, simple spinal and brainstem reflexes are already present. A newborn held supported will take rudimentary steps (walking reflex). More complex postural and righting reflexes appear during the first few weeks of life. With maturation of the cortex and commissural pathways, acts requiring associated sensory functions (grasping and groping) are possible, followed by volitional control of movement. Finally, the ability to perform skilled acts with a high degree of precision emerges.

Complex mechanisms underlie even the simplest discrete voluntary movement. All of the levels of motor integration contribute to the precision of movement. Initiation of contraction of the agonist (prime mover) must be accompanied by graded relaxation or contraction of the antagonists and synergists. Smooth, accurate movement requires the ability for the movement to be stopped at any point, reversed, and started again at a different degree of contraction or in a different direction. Stereotyped and patterned movements, integrated at lower levels, may be part of the act. Postures must be assumed that can be modified or shifted easily and instantly for adjustment to the next movement. Throughout all of this, the volitional elements and purposeful aspects of the act are of paramount importance.

Knowing the structure and function of the different levels of motor control, the relationships between the motor systems, and the changes in motor activity that occur in disease helps in understanding disorders of the motor system. Many neuroscientists over the years have envisioned various hierarchical schemes with different levels of complexity of motor activity. In this text, we consider the following levels: the motor unit (lower motor neuron, final common pathway) and the segmental (spinal cord), brainstem, cerebellar, extrapyramidal, and pyramidal levels.

The lowest echelon of motor activity is the motor unit, which consists of an alpha motor neuron in the spinal cord or brainstem, its axon, and all of the muscle fibers it innervates. The segmental or spinal cord level mediates simple segmental reflexes, such as the withdrawal reflex, and includes the activity of many motor units and elements of both excitation and inhibition involving

agonists, synergists, and antagonists. Various descending suprasegmental motor systems modulate the activity that occurs at the segmental level ([Figure 22.1](#)). The pyramidal (corticospinal) system arises from the primary motor cortex in the precentral gyrus. The corticospinal system is the primary, overarching suprasegmental motor control mechanism. The function of the corticospinal system is modulated and adjusted by the activity of the extrapyramidal and cerebellar systems. The extrapyramidal system arises primarily in the basal ganglia. Centers in the brainstem that give rise to the vestibulospinal, rubrospinal, and related pathways are of importance in postural mechanisms and standing and righting reflexes. The psychomotor, or cortical associative, level has to do with memory, initiative, and conscious and unconscious control of motor activity that arises primarily from the motor association cortex anterior to the motor strip.

These levels are not individual motor systems and do not normally act individually or separately. Anatomists continue to have difficulty in even defining the constituents of some of these levels (e.g., the corticospinal or pyramidal vs. the basal ganglion or extrapyramidal). These levels are components of the motor system as a whole; each is part of the complex motor apparatus. Each contributes its share to control of the lower motor neuron on which, as the final common pathway, all motor control systems converge. Disease at each of these levels causes characteristic signs and symptoms ([Table 22.1](#)). Some disorders of the motor system may involve more than one level. In addition, all purposeful movements are guided by a constant stream of afferent impulses that impinge on various levels of the motor system. Sensory and motor functions are interdependent in the performance of volitional movement, and it is not possible to consider the motor system apart from the sensory system. Impairment of sensation may affect all aspects of motion—volitional, reflex, postural, tonic, and phasic.

A brief sketch of the organization of the motor system may help to lay a foundation before discussing each level in detail ([Figures 22.1](#) and [22.2](#)). The premotor and supplementary cortices control the planning and preliminary preparation for movements, which the primary motor cortex in the precentral gyrus then executes. The primary motor cortex also receives input from the basal ganglia and the cerebellum ([Figure 22.2](#)). The corticospinal (pyramidal) and corticobulbar tracts arise from the precentral gyrus, descend through the corona radiata, and enter the posterior limb of the internal capsule. The internal capsules merge in their descent with the cerebral peduncles, which form the base of the

midbrain. Corticobulbar fibers terminate in the lower brainstem on cranial nerve nuclei and other structures. Corticospinal fibers aggregate into compact bundles, the pyramids, in the medulla. At the level of the caudal medulla, 90% of the pyramidal fibers decussate to the opposite side and descend throughout the spinal cord as the lateral corticospinal tract. About 10% of the corticospinal fibers descend ipsilaterally in the anterior corticospinal tract and decussate at the level of the local spinal synapse. Pyramidal fibers preferentially innervate certain lower motor neuron groups.

Descending motor system fibers send collaterals to other structures to help control and coordinate movement. These structures in turn project back to the cortex to form feedback loops that ensure coordinated interactions between the suprasegmental motor systems ([Figure 22.2](#)). The thalamus, especially the ventral lateral (VL) and the ventral anterior (VA) nuclei, serves as the relay station for projections from the other centers back to the cortex. The VL projects predominantly to the primary motor cortex and the VA to the premotor regions.

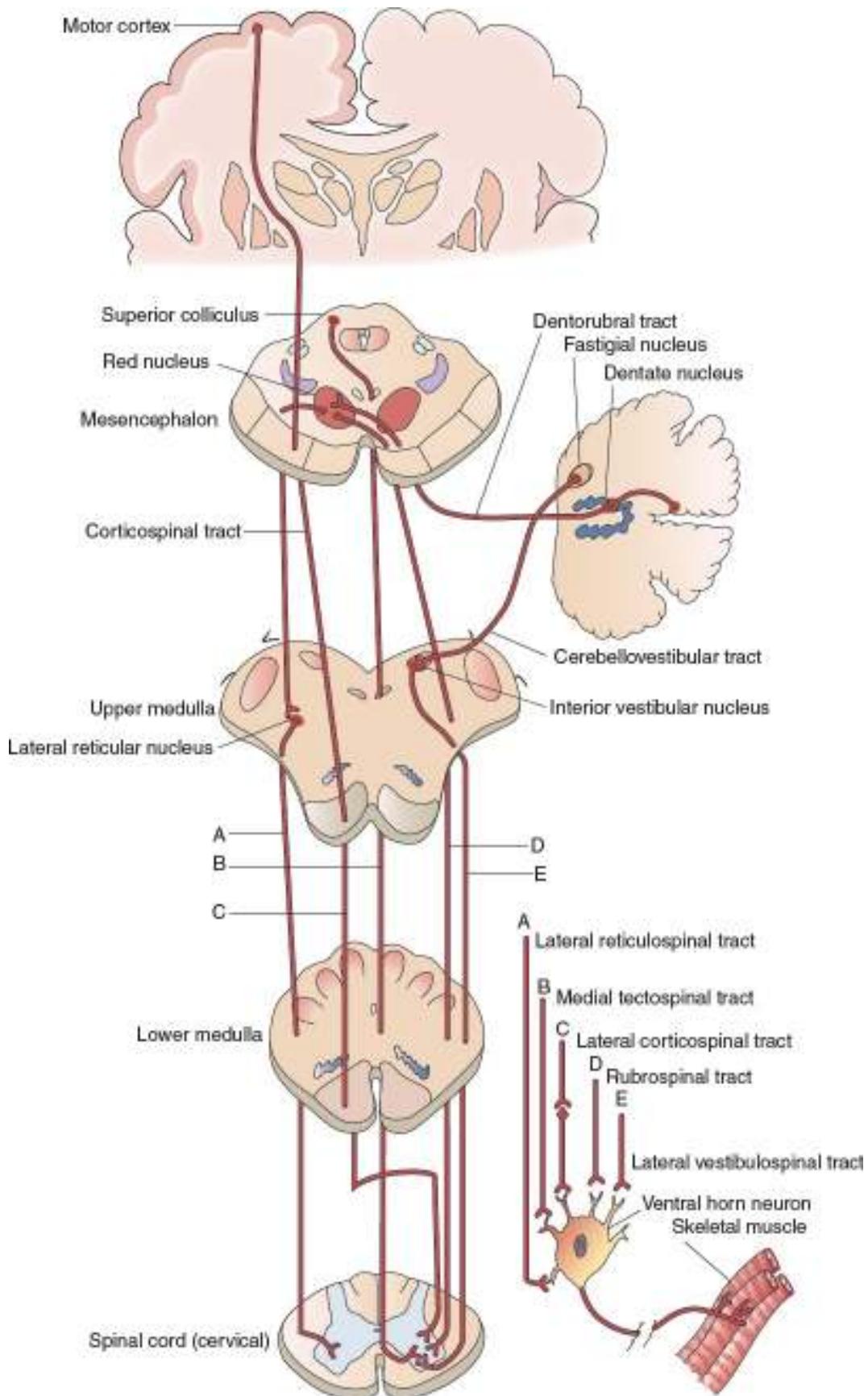


FIGURE 22.1 The most important descending pathways that act upon the anterior horn cell of the spinal cord (final common pathway).

TABLE 22.1

Changes in Motor Function

Level	Weakness	Tone	Vibration and Contour	Fasciculations	Atonia	Deep Tendon Reflexes	Abnormal Movements	Pathologic Associated Movements
Motor Unit Lesions								
a. Lower motor neuron	Focal or segmental; bilateral	Hypotonic	Absent vibration contour	Common	Absent	Usually decreased	None except fasciculations	Absent
b. Nerve root; plexus; peripheral nerve	Focal or segmental	Hypotonic	Absent vibration contour	Occasional	Absent	Decreased or absent	None except rare fasciculations	Absent
c. Neuromuscular junction	Diffuse or proximal; bilateral	Usually normal	Usually no change	Serotonia present because of anticholin- esterase drugs	Absent	Usually normal	None	Absent
d. Muscle	Diffuse, proximal, or distal	Normal or hypotonic	Normal; atrophy, hypertrophy, or pseudohypertrophy	None	Absent	Normal unless weakness is very severe	None	Absent
Corticospinal tract lesion	Monic; hemi-, pono-, quadriparetic, often incapacitating distribution	Specific	Normal	None	Absent	Increased unless paroxysms acute	None	Present
Extrapyramidal lesion	None or mild	Rigid	Normal	None	Absent	Normal	Present	Absent
Corticofugal lesion	None; atonia may simulate weakness	Hypotonic	Normal	None	Absent	Paradoxical normal	None except intention tremor	Absent
Neuropathic lesion	Bilat. breakdown; no true loss of power; may simulate any type	Normal or variable; often fatigably increased	Normal	None	Absent; but incoordination may simulate ataxia	Normal; may have poor relaxation and ataxic; char. jerkiness	May be present	Absent

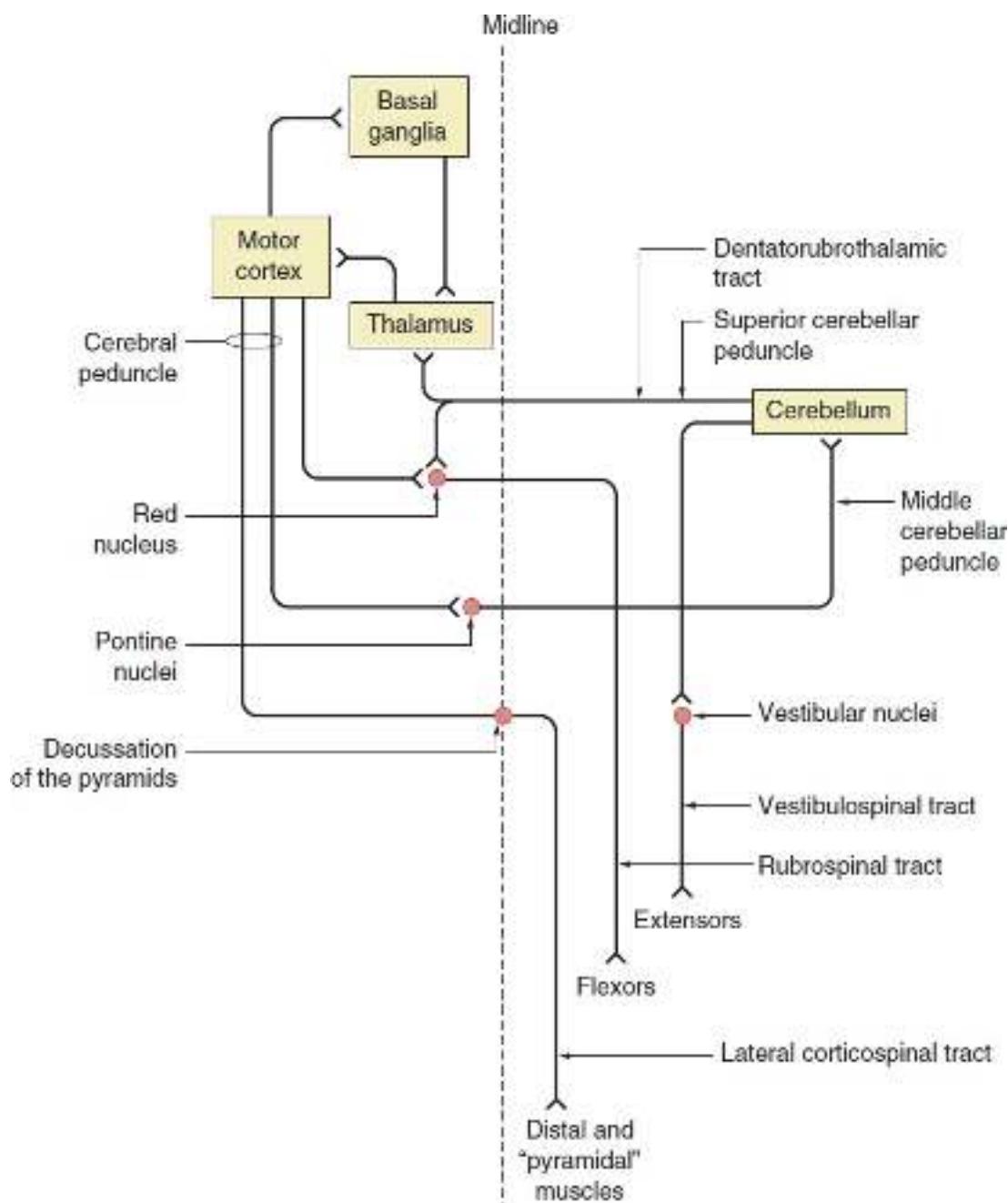


FIGURE 22.2 Major connections of the motor system. Note feedback loops between cortex and cerebellum and cortex and basal ganglia. (Modified from Campbell WW, Pridgeon RP. *Practical Primer of Clinical Neurology*. Philadelphia: Lippincott Williams & Wilkins, 2002, with permission.)

As fibers from the motor cortex run downward through the internal capsule, they send collaterals to the basal ganglia. Fibers from the basal ganglia project to VA and VL, which in turn project to the cortex, creating a feedback loop. The substantia nigra also projects to the striatum and influences its activity. The

motor cortex and cerebellum are also part of a circuit. The pontine nuclei lie scattered among the descending motor and crossing pontocerebellar fibers in the basis pontis. Corticopontine fibers synapse on pontine nuclei, which then give rise to pontocerebellar fibers that project across the midline to the contralateral cerebellar hemisphere through the middle cerebellar peduncle. The cerebellum in turn projects to the contralateral VL via the superior cerebellar peduncle, which decussates in the midbrain. The VL nucleus in turn projects to the motor cortex to complete the circuit. The cerebellum also receives unconscious proprioception from muscle spindles and Golgi tendon organs via the spinocerebellar and cuneocerebellar tracts. The cerebellum also projects to the ipsilateral vestibular nuclei, which give rise to the vestibulospinal tracts. The lateral vestibulospinal tract descends from the lateral vestibular nucleus to the spinal cord, where it facilitates ipsilateral extensor muscle tone of the trunk and extremities. As they descend, corticospinal fibers send collaterals to the ipsilateral red nucleus. The rubrospinal tract arises from the red nucleus and then immediately decussates and descends to facilitate flexor muscle tone, primarily in the upper extremities. The tectospinal tract arises from the superior colliculus, crosses in the dorsal tegmental decussation, and descends to influence muscles of the neck and upper back. It functions to move the head in response to external stimuli and to maintain head position in relation to the body position. The uncrossed pontine (medial) reticulospinal tract arises from the oral and caudal pontine reticular nuclei and facilitates extensor muscles, especially of the trunk and proximal extremities. The medullary (lateral) reticulospinal tract arises from the gigantocellular reticular nucleus and is primarily uncrossed but with a small crossed component. It travels just anterior to the rubrospinal tract. It inhibits antigravity muscles and is involved with autonomic functions.

The cerebellar hemispheres influence muscles on the ipsilateral side of the body. The cerebellum projects to the contralateral red nucleus, and the rubrospinal tract then crosses back. Because of the “double decussation,” the rubrospinal tract controls muscles ipsilateral to the cerebellar hemisphere where the impulse originated. The vestibulospinal system remains ipsilateral throughout. The cerebral motor cortex on one side and the cerebellar hemisphere on the opposite side act in concert to control the arm and leg on a particular side of the body. Their actions are coordinated by projections from the cerebrum to the pontine nuclei, which send fibers to the contralateral cerebellum, which in turn projects back to the thalamus and cerebrum on the original side via the decussation of the dentatothalamic tract. Consider the right cerebellar

hemisphere, it receives input from the left cerebral cortex via the middle cerebellar peduncle and projects back to the left thalamus and motor cortex via the superior cerebellar peduncle. So, both the left cerebral hemisphere and the right cerebellar hemisphere control movements on the right side of the body.

OVERVIEW OF CLINICAL MANIFESTATIONS OF DISEASE OF THE MOTOR SYSTEM

The most common manifestation of motor system disease is weakness. Other abnormalities include alterations in muscle tone, changes in muscle size and shape, abnormal involuntary movements, and defective coordination. Subsequent chapters deal with abnormal motor functions in more detail.

Motor Strength and Power

Weakness is a common abnormality and can follow many patterns. Terminology may become problematic. For instance, weakness may be generalized or localized, symmetric or asymmetric, proximal or distal, or upper motor neuron or lower motor neuron. The term focal is often used to imply asymmetry; a patient with a hemiparesis is said to have a focal examination. The term generalized is often used to imply symmetry, even though the weakness may not truly be generalized. A disease may cause weakness in a particular distribution that is bilaterally symmetric (e.g., the scapuloperoneal syndromes), but these are not generally regarded as focal even though the involvement is very localized. A patient with bilateral carpal tunnel syndrome or bilateral peroneal nerve palsies would most properly be described as having a multifocal pattern of weakness, even though the weakness is bilateral and symmetric. The term nonfocal is often used to describe a patient's neurologic examination, particularly by nonneurologists. The implication is usually that the examination is normal or at least that there is no asymmetry. It is a poor and not very helpful term. A patient with Guillain-Barré syndrome causing generalized weakness and impending respiratory failure would have a nonfocal examination, yet be critically ill.

Focal weakness may follow the distribution of some structure in the peripheral nervous system, such as a peripheral nerve or spinal root. It may affect one side of the body in a hemidistribution. A hemidistribution may affect the arm, leg, and face equally on one side of the body, or one or more areas may

be more involved than others. Muscle groups preferentially innervated by the corticospinal tract are often selectively impaired. When weakness is nonfocal, it may be generalized, predominantly proximal, or predominantly distal. These various patterns have differential diagnostic and localizing significance. Identification of the process causing weakness is further aided by accompanying signs, such as reflex alterations and sensory loss ([Table 21.1](#)).

Generalized Weakness

Weakness may involve both sides of the body, more or less symmetrically. With truly generalized weakness, bulbar motor functions are also impaired. When the bulbar functions are intact and there is weakness of both arms and both legs, the patient is said to have quadriplegia—if only the legs, then paraparesis. When weakness affects all four extremities, the likely causes include myelopathy, peripheral neuropathy, a neuromuscular junction disorder, or a myopathy.

When myelopathy is the culprit and the deficit is incomplete, more severe involvement of those muscles preferentially innervated by the corticospinal tract can frequently be discerned. Reflexes are usually increased (though in the acute stages, they may be decreased or absent); there is usually some alteration of sensation, sometimes a discrete spinal sensory “level”; superficial reflexes disappear; and there may be bowel and bladder dysfunction. Generalized peripheral nerve disease tends to predominantly involve distal muscles. There is no preferential involvement of corticospinal innervated muscles, reflexes are usually decreased, sensory loss is frequently present, and bowel and bladder functions are not disturbed. With a neuromuscular junction disorder, the extremity weakness is likely to be worse proximally, sensation is spared, reflexes are normal, and there is usually involvement of bulbar muscles. With myopathy, weakness is usually more severe proximally, reflexes are normal, sensation is normal, and, with a few exceptions, bulbar function is spared except for occasional dysphagia. These are generalizations. Some neuropathies may cause proximal weakness, and some myopathies may affect distal muscles; not all patients with a neuromuscular transmission disorder have bulbar involvement.

Localized Weakness

When the arm and leg on one side of the body are weak, the patient is said to have a hemiparesis. This may range in severity from very mild, manifest only as

pronator drift and impairment of fine motor control, to total paralysis. Monoparesis is weakness of only one extremity, such as the leg contralateral to an anterior cerebral artery stroke. Reflexes—typically increased unless the process is acute—and accompanying sensory loss help identify such focal weakness as central in origin.

A mononeuropathy, such as a radial nerve palsy, or a spinal root lesion, such as from a herniated disc, causes weakness limited to the distribution of the involved nerve or root. A plexopathy may cause weakness of the entire limb or weakness only in the distribution of certain plexus components. With such lower motor neuron pathology, reflexes are typically decreased, and there is often accompanying sensory loss. Localization of focal weakness due to root, plexus, and peripheral nerve pathology requires intimate familiarity with peripheral neuroanatomy.

Motor neuron disease is a special case. Amyotrophic lateral sclerosis (ALS) characteristically involves both the upper and lower motor neurons. It produces a clinical picture of weakness and wasting due to involvement of the lower motor neurons in the anterior horn of the spinal cord, combined with weakness and hyperreflexia due to involvement of the upper motor neurons in the cerebral cortex that give rise to the corticospinal tract. There is upper motor neuron weakness (cerebral cortex pathology) superimposed on lower motor neuron weakness (spinal cord pathology). ALS usually begins with focal weakness, often involving one hand or one foot.

A multineuro, multiroot distribution of weakness, normal or increased reflexes, and a lack of sensory loss are usually the earliest suggestions as to the nature of the problem. In ALS, the weakness early tends to be asymmetric and associated with hyperreflexia. An extremity that is both atrophic and hyperreflexic is characteristic. As the condition progresses, the clinical picture evolves into one of generalized weakness.

It is very rare for a myopathy or neuromuscular junction disorder to cause focal weakness, although these disorders may have a predilection for certain muscles, such as the extraocular muscles in myasthenia gravis or the thigh muscles in quadriceps myopathy.

Other Motor System Abnormalities

Muscle tone may be increased (hypertonia) or decreased (hypotonia). Hypertonia comes in two common variants: rigidity and spasticity. When the increased tone

occurs to more or less the same degree throughout the range of passive motion of a limb, and is independent of the speed of the movement, it is referred to as rigidity. When the hypertonia is most marked near the middle of the range of motion and is more apparent with fast than with slow passive movement, it is referred to as spasticity. One of the key characteristics of spasticity is that the hypertonus is velocity dependent, most evident with rapid movements. In lead pipe (plastic) rigidity, there is smooth resistance throughout the range independent of the rate of movement. Gegenhalten (paratonia) is an increase in tone in a limb more or less proportional to the examiner's attempt to move it. In cogwheel rigidity, there is ratchety, jerky, tremulous variation in the hypertonia, due primarily to superimposed tremor. Spastic hypertonia is typically associated with increased deep tendon reflexes, loss of superficial reflexes, and Babinski signs. Cogwheel rigidity occurs in Parkinson's disease and related conditions. Gegenhalten is usually associated with other abnormal neurologic signs depending on the etiology. The term dystonia refers to transient or sustained hypertonic conditions that do not fit into the other categories. Hypotonia occurs in two primary settings in the adult: myopathies and cerebellar disease. Infantile hypotonia (floppy baby) is a common clinical problem. The differential diagnosis of infantile hypotonia is extensive, and the workup of a floppy baby is a frequent exercise in pediatric neurology.

Muscle Volume and Contour

Muscle mass or volume may be decreased (atrophy) or increased (hypertrophy). Neurogenic atrophy results from a lesion involving the anterior horn cells, nerve root, or peripheral nerve innervating a muscle; it may be severe. Muscle diseases usually cause only mild to moderate atrophy of the involved muscles. Disuse atrophy occurs after immobilization, as when a limb is in a cast, and is usually mild to moderate in severity and recovers quickly with resumption of use.

True muscle hypertrophy results from an increase in the size of the muscle. It is most often physiologic hypertrophy from heavy use, but it can occur in certain neuromuscular disorders. Pseudohypertrophy refers to apparent muscle enlargement due to replacement of diseased muscle by fat and fibrous tissue. Enlarged calf muscles in patients with Duchenne muscular dystrophy are a classic example of muscle pseudohypertrophy.

Abnormal Movements

Abnormal involuntary movements occur in a host of neurologic conditions. They come in many forms, ranging from tremor to chorea to muscle fasciculations to myoclonic jerks. The only common characteristic is that the movements are spontaneous and not under volitional control. Involuntary movements may be rhythmic or random, fleeting or sustained, and predictable or unpredictable. They may occur in isolation or be accompanied by other neurologic signs. Common types include tremor, chorea, athetosis, hemiballismus, dystonia, tics, and dyskinesias.

Coordination

Coordination and control of fine motor movements are delicate functions that require smooth interactions between the different components of the motor system as well as normal sensory function. The cerebellum is a critical component, and disease of the cerebellum frequently causes impaired coordination in the absence of weakness or other motor abnormalities. But poor coordination may also be a manifestation of corticospinal tract or extrapyramidal disorders.

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CHAPTER 23

The Motor Unit Level

The motor unit consists of an alpha motor neuron (lower motor neuron), its axon, and all its subject muscle fibers; it is the final common pathway for all motor activity, both voluntary and involuntary ([Figure 23.1](#)). Clinical disorders may affect any portion of the motor unit (cell body, nerve root, plexus, peripheral nerve, neuromuscular junction [NMJ], or muscle), and diseases at different sites have different clinical features. Sensory dysfunction—such as pain and paresthesias—may accompany lesions involving the nerve root, plexus, and peripheral nerve portions of the motor unit. Nevertheless, the motor unit remains a useful conceptual framework for understanding disorders involving the peripheral neuromuscular apparatus.

THE MOTOR UNIT

Alpha motor neurons reside in the anterior horn of the spinal cord and the brainstem motor nuclei. The axon traverses the anterior root and the peripheral nerve en route to the muscle. The peripheral nerve enters the muscle at the motor point and divides into intramuscular branches. These arborize within a muscle fascicle and terminate as fine twigs, which end as axon boutons. Terminal boutons abut the motor end plates of individual muscle fibers across a synaptic cleft, forming NMJs. Each muscle fiber has only a single end plate.

Terminal axonal twigs ramify in the muscle and innervate widely dispersed muscle fibers. A motor unit may have anywhere from a handful of muscle fibers to more than a thousand. The innervation ratio refers to the number of muscle fibers in a motor unit. A low innervation ratio means few muscle fibers are innervated by a single axon and is characteristic of muscles under precise and finely graded voluntary control, such as extraocular or laryngeal muscles. A muscle performing a gross motor movement may have several hundred muscle

fibers per motor unit. The gastrocnemius has about 2,000. The innervation ratios for the brachial biceps, tibialis anterior, and deltoid are 209, 329, and 239 fibers, respectively. The anatomical scatter of fibers belonging to the same motor unit may vary from muscle to muscle. Electrophysiologic motor unit counting techniques estimate that an intrinsic hand muscle has about 100 motor units. Beyond the age of 60 years, there is a decline in the number of functioning motor units.

Motor units vary by size within a muscle. Smaller motoneurons have smaller motor unit territories. To produce a smoothly graded muscle contraction, motor units are recruited, more or less, in order of increasing size. Small motoneurons are first recruited, and increasing force of contraction calls forth activity from increasingly larger motoneurons: the size principle.

Motor units are classified as type 1 or type 2. Muscles were broadly divided into red muscle and white muscle, dark meat and light meat, long before the basis for the difference was understood. The red or dark color is now known to result from the presence of the instruments for oxidative metabolism: myoglobin, mitochondria, and a vascular network for delivery of oxygen to the metabolizing muscle cells. Histochemical stains help separate the different types of muscle fibers and often allow the appreciation of structural detail and abnormalities not seen with routine hematoxylin and eosin stains. The myosin ATPase (adenosine triphosphatase) histochemical stain identifies two distinct populations of muscle fibers, referred to as type 1 and type 2, which correspond to type 1 and 2 motor units. Further adjustment of pH allows separation of type 2 muscle fibers into types 2A and 2B ([Box 23.1](#)). An average muscle contains about 40% type 1 fibers and 60% type 2 fibers. However, this ratio varies with the anatomical location and function of the muscle, and similar muscles may vary among individuals. All fibers in a particular motor unit are of the same type, and there is good correlation between the mechanical properties and other attributes of a motor unit and the histochemical reactions of its muscle fibers. The fiber type mix of a muscle is determined by its innervation and ultimately by its function. In needle electromyography, the summated electrical activity of all the muscle fibers of a motor unit—a motor unit action potential—is recorded by a needle electrode inserted into the muscle.

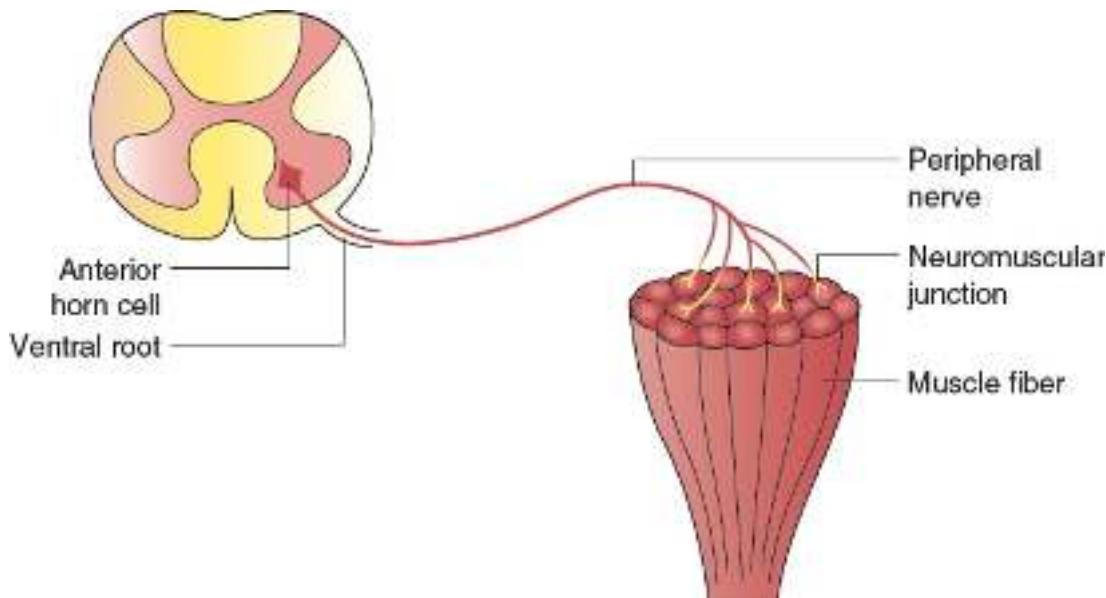


FIGURE 23.1 The motor unit. An alpha motor neuron in the anterior horn gives rise to an axon, which branches and arborizes in the periphery to innervate scattered muscle fibers within a muscle fascicle.

BOX 23.1

Type 1 and Type 2 Muscle Fibers

The myosin ATPase (adenosine triphosphatase) stain identifies the two distinct populations of type 1 and type 2 muscle fibers; the differences are summarized in [Table 23.1](#). Cross sections reveal a random admixture of the two fiber types creating a checkerboard pattern. The type 2 fibers stain darkly and the type 1 fibers lightly at pH 9.4; the staining characteristics reverse at pH 4.3. Preincubation at pH 4.6 identifies two kinds of type 2 fibers: type 2A and type 2B. The 2B fibers are the classic fast twitch, fatigue-sensitive glycolytic fibers, whereas 2A fibers have characteristics intermediate between type 1 and type 2B, with some oxidative capability, slower twitch, and more fatigue resistance than the 2B fibers. Pathologic processes may cause characteristic abnormalities of fiber type distribution or proportion or produce changes primarily in one particular fiber type.

Type 1 muscle fibers are rich in oxidative enzymes and mitochondria but sparse in glycogen; they are designed for sustained, long-duration contraction under aerobic conditions. Red, or dark, meat is high in type 1 fibers. Type 2

fibers are rich in glycogen and glycolytic enzymes but sparse in oxidative enzymes, mitochondria, and lipid. They are designed for brief, intense bursts of activity under anaerobic conditions. White meat is high in type 2 fibers. The mnemonic “one, slow, red ox” helps recall the essentials: type 1 fibers, slow muscle, red meat, oxidative metabolism.

TABLE 23.1	Attributes and Characteristics of Type 1 and Type 2 Muscle Fibers	
Attribute	Type 1	Type 2
ATPase stain at pH 4.6	Dark	Light
ATPase stain at pH 9.4	Light	Dark
Oxidative enzymes	High	Low
Lipid	High	Low
Mitochondria	High	Low
Glycogen	Low	High
Glycolytic enzymes	Low	High
Function	Sustained contraction	Brief contraction
Twitch speed	Slow	Fast
Metabolism	Aerobic	Anaerobic
Fatigue	Resistant	Sensitive

ATPase, adenosine triphosphatase.

In another functional and physiologic scheme, motor units are classified into three different types: fast twitch, fatigue sensitive, or fast fatigable (FF); slow twitch, fatigue resistant (S); and intermediate or fast fatigue resistant (FR). Type FF units are fast twitch and fatigue sensitive, type 2B histochemically, rich in glycogen but poor in oxidative enzymes, and designed for brief, phasic activity. Type S units are slow twitch, fatigue resistant, type 1 histochemically, low in

glycolytic but high in oxidative enzymes, and designed for sustained, tonic activity. Type FR is intermediate, fast twitch but more fatigue resistant than type FF, type 2A histochemically, high in glycolytic, and intermediate in oxidative enzyme activity.

Myotomes

A myotome consists of all the muscles innervated by a specific nerve root. Most skeletal muscles receive innervation from two or more roots, and there is inherent variability in the myotomal patterns among individuals. Early anatomists reported myotome innervation from detailed dissections, and some errors have been perpetuated through the years. Many different innervation charts are available, and most vary in some details. Some misinformation remains, such as the inclusion of C6-C7 innervation to the thenar muscles, which does not fit clinical reality. The issue has been compounded by observations during intraoperative recordings that indicate contributions from unexpected sources to leg muscles and anomalous innervation so frequent as to be the rule rather than the exception. Liveson and Ma present charts derived from seven different sources, separating “new myotomes” derived from electromyographic data.

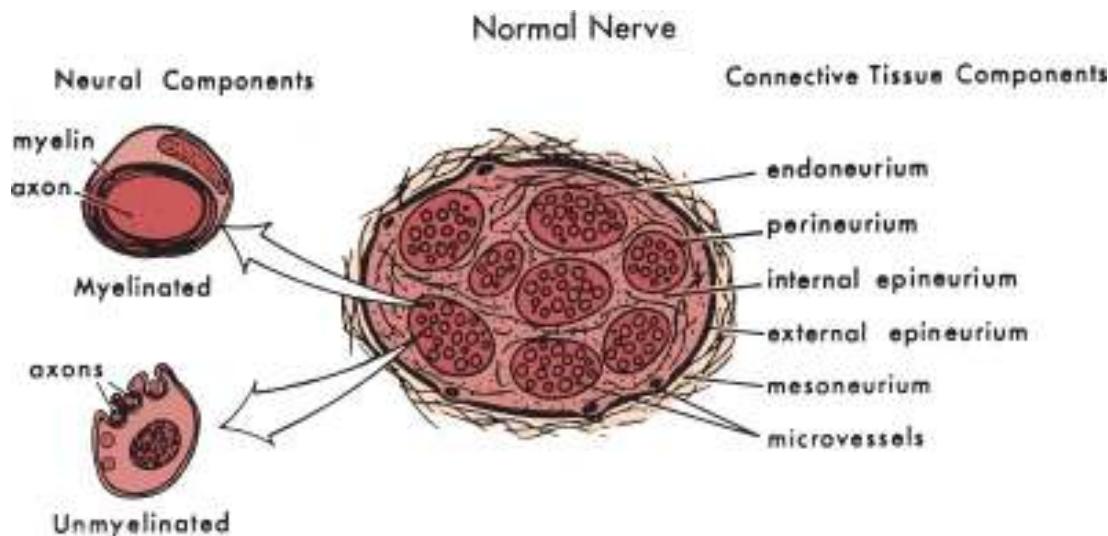


FIGURE 23.2 The normal peripheral nerve is composed of connective tissue and neural tissue components. The nerve fibers may be myelinated or unmyelinated. (Modified with permission of Georg Thieme Verlag KG from Mackinnon SE, Dellon AL. *Surgery of the Peripheral Nerve*. 1st ed. New York: Thieme, 1988; permission conveyed through Copyright Clearance Center, Inc.)

Microanatomy of the Peripheral Nerve

Peripheral nerves are composed of myriad axons, ensheathed by myelin of varying thickness and supported by Schwann cells, all imbedded in a matrix of connective tissue. Nerves are divided into discrete internal fascicular compartments by perineurium. The blood-nerve barrier is a physiologic partition, created by the perineurium and the endothelium of intrafascicular capillaries. It regulates the nerve microenvironment and acts as a diffusion barrier. The extreme terminal ends of nerve fibers are not protected by perineurium and have no effective blood-nerve barrier, a detail of probable importance in the pathogenesis of some peripheral neuropathies. Within each fascicle, endoneurium separates individual axons and their Schwann cells ([Figure 23.2](#)). Fascicles are bound together into nerve trunks by the epineurium, loose areolar connective tissue that also contains blood vessels, lymphatics, and the nervi nervorum. The epineurium also serves an important cushioning role. The interfascicular (internal) epineurium lies between fascicles; the epifascicular (external) epineurium circumferentially envelops the entire nerve.

Fascicles bifurcate, join with adjacent fascicles, redivide, and recombine to create a complex internal fascicular network ([Figure 23.3](#)). Yoon et al. have imaged the individual fascicles beautifully with 7T MRI ([Figure 23.4](#)). Nerves can be classified into monofascicular, oligofascicular, and polyfascicular types. A polyfascicular pattern is common in regions subject to mechanical stress and where there is heavy fiber exchange, such as the brachial plexus. Plexiform fascicular exchange is most prominent proximally, and a constant fascicular pattern is present for only a short distance in proximal regions of a nerve. Fascicles innervating a particular muscle or sensory zone become more discrete and constant in position as they approach the target organ. This complex intraneuronal topography has important clinical, surgical, and electrophysiologic implications.

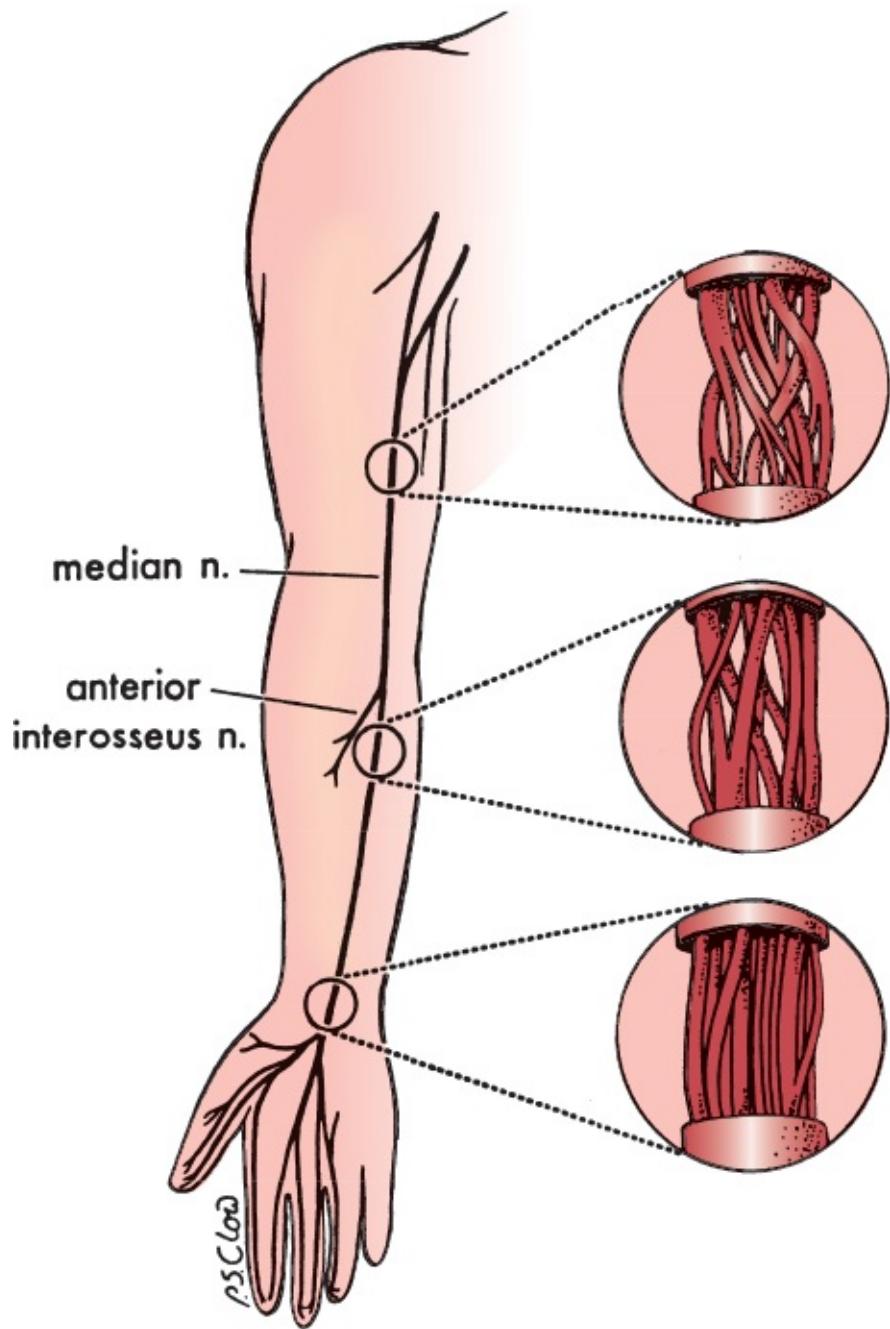


FIGURE 23.3 An illustration of the internal topography of the median nerve at different levels. The complexity of the internal fascicular anatomy is apparent. The degree of plexus formation between fascicles decreases in the distal portion of the nerve as the bundles approach their target muscles. (Modified with permission of Georg Thieme Verlag KG from Mackinnon SE, Dellon AL. *Surgery of the Peripheral Nerve*. 1st ed. New York: Thieme, 1988; permission conveyed through Copyright Clearance Center, Inc.)

Axons are divided into three major size groups: large myelinated, small myelinated, and unmyelinated. Large myelinated axons have diameters in the 6-

to 12- μm range, small myelinated axons are 2 to 6 μm , and unmyelinated axons are 0.2 to 2.0 μm . Small myelinated fibers are about three times more numerous than large myelinated axons. The myelin sheath adds additional thickness. Conduction is most efficient when the ratio of the axon diameter to total fiber diameter is 0.5 to 0.7.

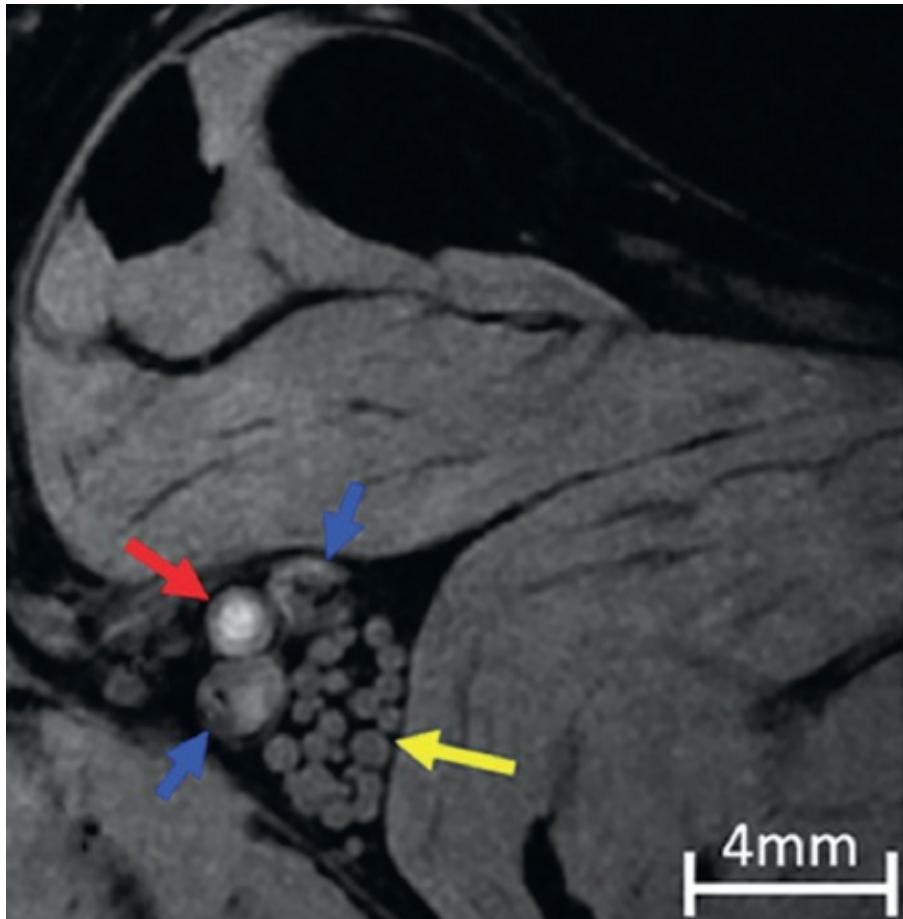


FIGURE 23.4 Individual fascicles of the tibial nerve are clearly identified at high resolution on 7T MRI, marked by the yellow arrow. Posterior tibial artery is marked by the red arrow and posterior tibial veins are marked by the blue arrows. (Modified from Yoon D, Biswal S, Rutt B, et al. Feasibility of 7T MRI for imaging fascicular structures of peripheral nerves. *Muscle Nerve*. 2018;57(3):494-498. Copyright © 2017 Wiley Periodicals, Inc. Reprinted by permission of John Wiley & Sons, Inc.)

Ultrastructurally, axons contain cytoskeletal elements, neurofilaments, and neurotubules, which are synthesized in the cell body and move slowly down the axon at a rate of 3 mm/d. Neurotubules consist of polymerized dimers of tubulin protein forming longitudinally oriented hollow tubes about 20 nm in diameter and 1 mm long, linked by crossbridges to the neurofilaments. Neurofilaments are

smaller organelles that maintain axonal structure. Neurotubules are responsible for fast antegrade and retrograde axonal transport (see Physiology of the Peripheral Nerve below).

In myelinated axons, a single Schwann cell wraps a single internodal segment in concentric layers of myelin ([Figure 23.5](#)). Schmidt-Lantermann incisures are bits of Schwann cell cytoplasm sequestered between layers of myelin. The external plasma membrane of the Schwann cell is continuous with the outermost layer of myelin; the inner membrane of the Schwann cell is immediately adjacent to the outer surface of the axolemma. The external lamina is a condensation of extracellular matrix surrounding the entire external surface of the Schwann cell. The nodes of Ranvier are gaps in the myelin coverage between the territories of adjacent Schwann cells. Internodal length varies with fiber size; it is about 1 mm for large-diameter fibers. For unmyelinated axons, a single Schwann cell, sometimes referred to as a Remak cell, sends out processes to support several adjacent axons, lending to each primarily a cytoplasmic coat and only a minimal investment of myelin. A complex of several unmyelinated axons and their supporting Remak cell is encased by an external lamina.

Longitudinal section Cross section

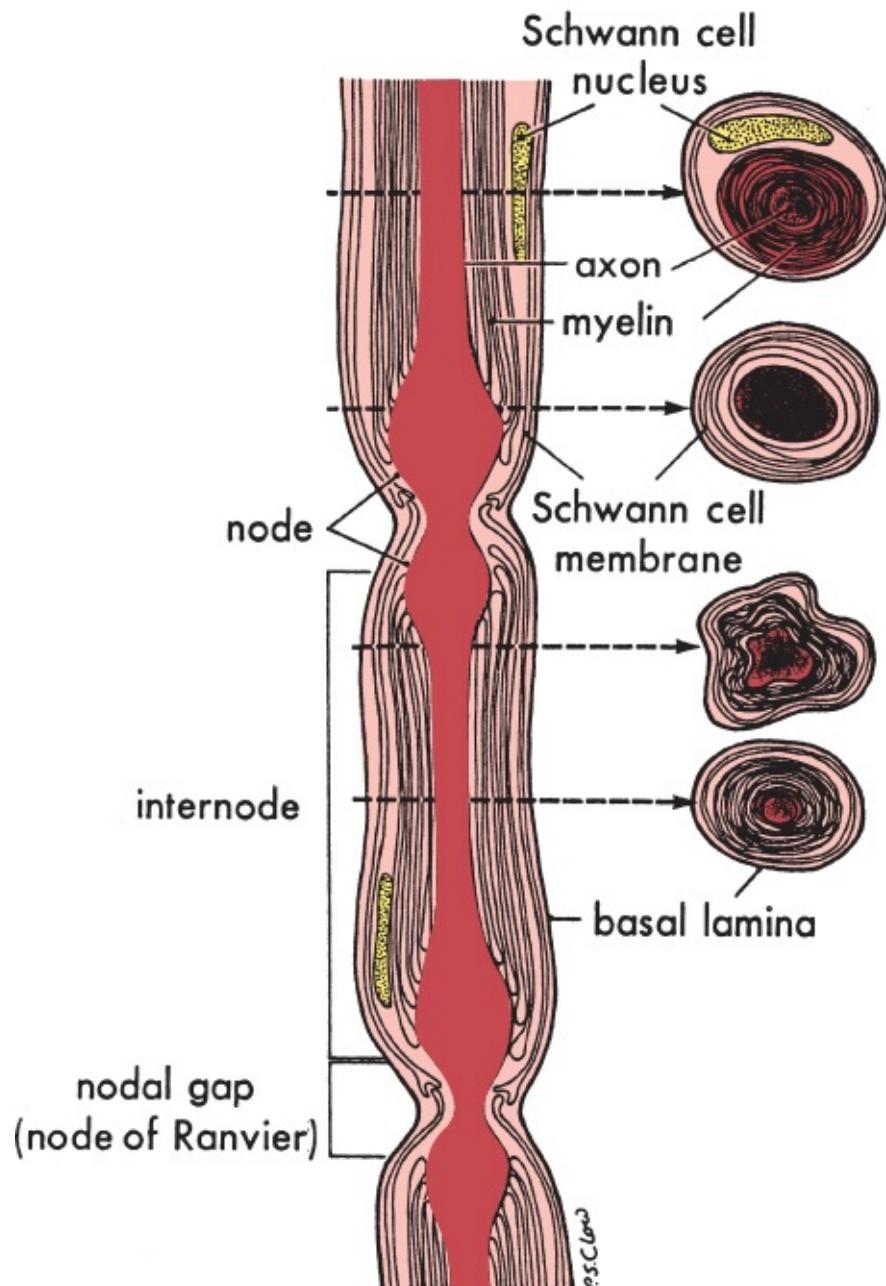


FIGURE 23.5 Illustration of a single myelinated nerve fiber demonstrates the relationships between the axon, the Schwann cell, and the myelin sheath at different points. (Modified with permission of Georg Thieme Verlag KG from Mackinnon SE, Dellen AL. *Surgery of the Peripheral Nerve*. 1st ed. New York: Thieme, 1988; permission conveyed through Copyright Clearance Center, Inc.)

Peripheral nerves receive blood supply from penetrating segmental arteries usually derived from adjacent vessels. Penetrating arterioles then form an

extensive longitudinal anastomotic network that runs within the nerve. Watershed zones of precarious perfusion within the nerve may explain some of the clinical manifestations seen in ischemic neuropathies, especially in vasculitis.

Physiology of the Peripheral Nerve

Peripheral nerve fibers are classified according to two schemes: the ABC and the I/II/III/IV systems ([Chapter 31](#)), both ranging from largest (A, I) to smallest (C, IV). The conduction velocity (CV) of a fiber depends on its diameter and degree of myelination; it ranges from less than 1 m/s for small, unmyelinated fibers to greater than 100 m/s for large, myelinated fibers. The compound nerve action potential—recorded *in vitro* from a mixed peripheral nerve—separates fibers into groups based on their CV. Aa and Ag fibers are efferent fibers from alpha and gamma motoneurons, respectively. Ab and Ad fibers are primarily cutaneous afferents. Group B fibers are preganglionic autonomic. Group C fibers are postganglionic autonomic, visceral afferents, and pain and temperature fibers. The Roman numeral system applies only to afferent fibers. The Ia fibers arise from nuclear bag muscle spindle fibers and joint receptors, the Ib fibers from Golgi tendon organs, and the II fibers from nuclear chain muscle spindle fibers. Class III fibers are cutaneous axons that correspond more or less to Ad fibers, and type IV fibers correspond to C fibers. Some neuropathies have a predilection for certain types and sizes of fibers. Large-fiber neuropathies affect strength, reflexes, and proprioception with relative sparing of pain and temperature sensation, whereas small-fiber neuropathies primarily affect pain, temperature, and autonomic function. Differential involvement of large versus small sensory fibers can sometimes be discerned clinically. The immunologic and biochemical differences between fibers, which might explain differential involvement, are just beginning to be understood. For example, the L2 membrane protein is expressed only on motor axon Schwann cells, and the nerves to the extraocular muscles are especially rich in ganglioside GQ1b, which may relate to their involvement in Miller-Fisher syndrome.

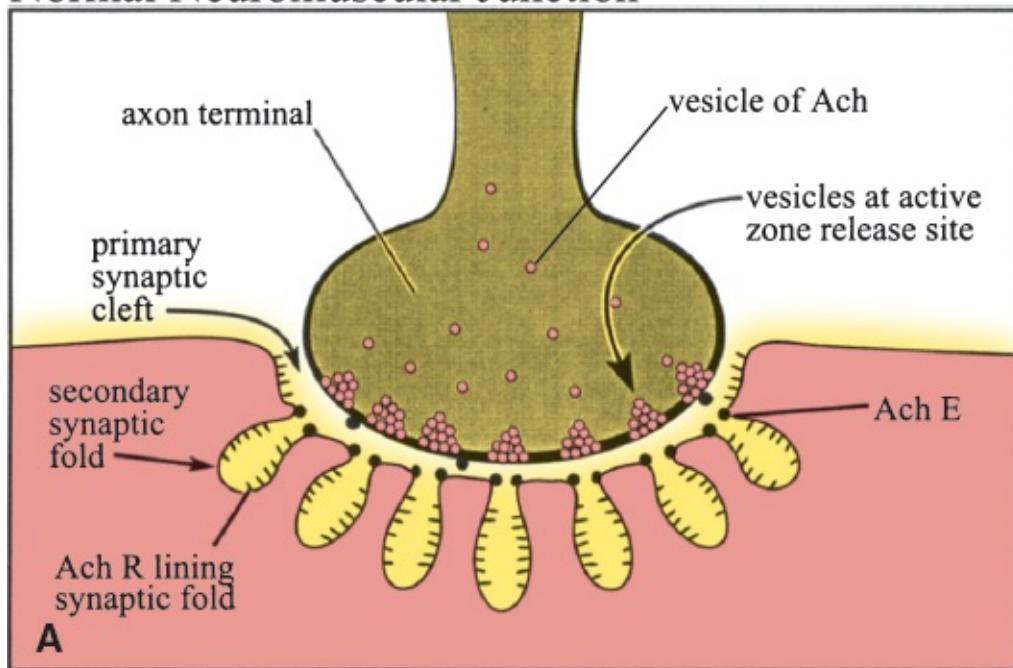
The axoplasm is in constant flux, containing elements that flow to and fro along its length between the cell body and the periphery. Antegrade axoplasmic flow moves from the cell body distally; retrograde flow moves centripetally. Antegrade flow has multiple components. Slow axonal transport, 1 to 3 mm/d, conveys cytoskeletal proteins to the periphery for maintenance and renewal of

axoplasm, along with neurotransmitters, enzymes, and other components. Fast axonal transport, 400 mm/d, largely transports membrane-bound vesicles that are propelled by kinesin, a microtubule-associated ATPase. Abnormalities of axonal transport are likely important in the mechanism of dying back or length-dependent neuropathies. Several substances produce neuropathy by disrupting the cytoskeletal elements: vinca alkaloids, taxoids, and hexacarbons, for example. Retrograde flow moves materials from the periphery back to the cell body; it is the mechanism through which some neurotrophic viruses reach the central nervous system.

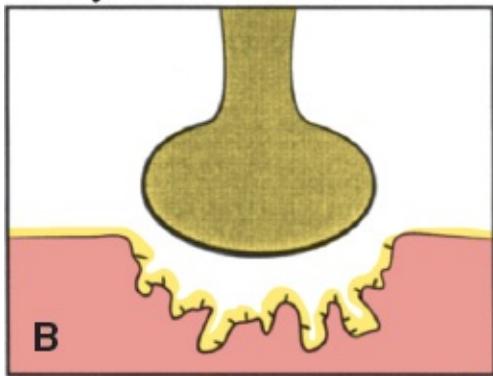
Anatomy and Physiology of the Neuromuscular Junction

In the nervous system, presynaptic electrical events are converted to chemical events at the synapse and are converted again into electrical events postsynaptically. The NMJ is a specialized synapse through which electrical events in the peripheral nerve are transduced into chemical events that then induce depolarization of the postsynaptic muscle membrane, which in turn induces muscle contraction. Disturbed neuromuscular transmission (NMT) results in several different clinical disorders, which are characterized primarily by weakness and fatigability.

Normal Neuromuscular Junction



Neuromuscular Junction
in Myasthenia Gravis



Neuromuscular Junction
in Lambert-Eaton Syndrome

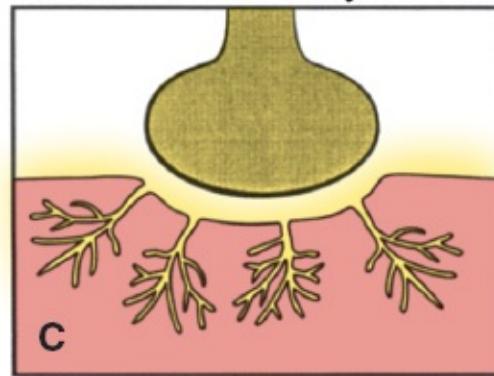


FIGURE 23.6 A. A normal neuromuscular junction (NMJ). B. An NMJ in myasthenia gravis, degraded by immunologic attack, simplified, and depopulated of acetylcholine receptor. C. An NMJ in Lambert-Eaton's syndrome, highly complex and convoluted with increased surface area. (From Campbell WW. *Essentials of Electrodiagnostic Medicine*. Philadelphia: Lippincott Williams & Wilkins, 1999. Modified with permission of Dr. William W. Campbell.)

An intramuscular nerve branch ends by forming a bulbous swelling—the terminal bouton. The primary synaptic cleft separates the terminal bouton from the postsynaptic muscle membrane, which is in turn divided into a number of secondary synaptic clefts, or junctional folds. The postsynaptic muscle

membrane is blanketed by a dense array of nicotinic acetylcholine receptor (AChR) molecules. Freeze-fracture techniques show the AChR as large particles concentrated on the tips of junctional folds, extending about halfway down into the secondary synaptic clefts. The AChR is a complex structure, consisting of two alpha subunits and beta, gamma, and delta subunits, plus an ion channel. The main immunogenic region of the AChR is the site that is attacked by autoantibodies in the majority of cases of myasthenia gravis. In addition, there are acetylcholine esterase molecules on both presynaptic and postsynaptic membranes (Figure 23.6).

The terminal bouton is a beehive of metabolic activity. It is packed with cytoskeletal proteins, mitochondria, and numerous chemicals. Most importantly, it contains vesicles, which are membrane-bound collections of acetylcholine (ACh). In the cytoplasm of the terminal bouton, ACh is packaged into vesicles, which then migrate to and collect at primary release sites, or active zones. The active zones of the presynaptic membrane tend to line up opposite the secondary synaptic clefts of the postsynaptic membrane. The active zones are the sites of both exocytosis of ACh vesicles and ingress of calcium.

Events of Normal Neuromuscular Transmission (events outside boxes occur in the synaptic cleft)

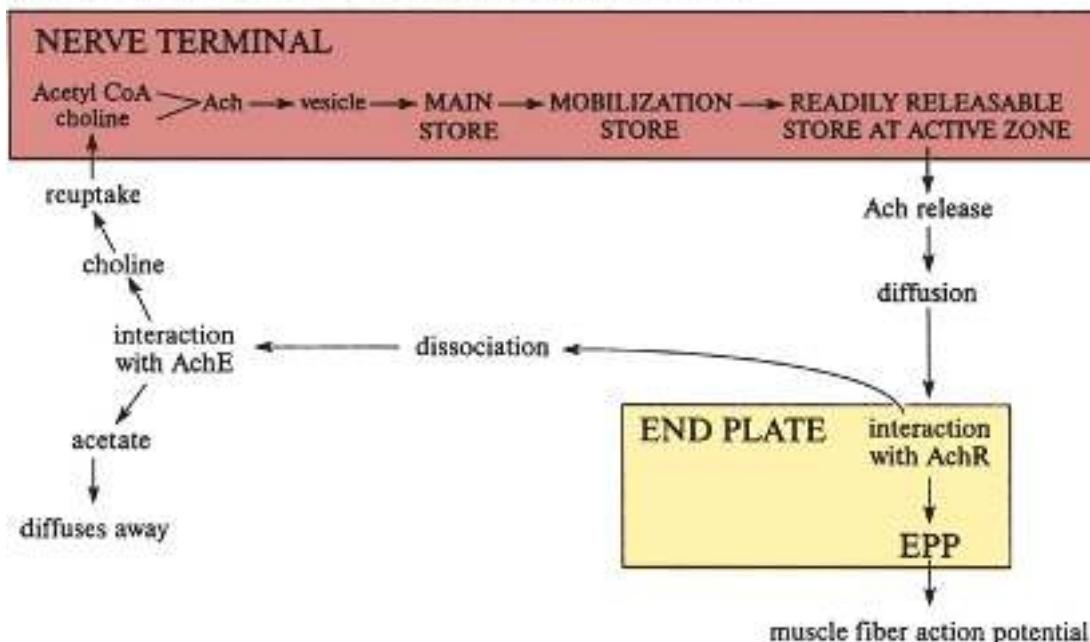


FIGURE 23.7 Schematic of the events of normal neuromuscular transmission. Ach, acetylcholine; AchE, acetylcholinesterase; AchR, acetylcholine receptor; EPP, end plate potential. (From Campbell WW. *Essentials of Electrodiagnostic Medicine*. Philadelphia: Lippincott Williams & Wilkins, 1999. Modified with permission of Dr.

The presynaptic membrane contains voltage-gated calcium channels. In response to nerve depolarization, these channels permit the influx of calcium into the presynaptic terminal, which greatly facilitates the release of neurotransmitter with the next nerve impulse. Magnesium has the opposite effect and inhibits the release of transmitter. After a nerve impulse, calcium diffuses out of the nerve terminal and is largely gone within 100 to 200 ms. Repetitive nerve impulses also increase the mobilization of ACh vesicles toward the active zones. As a result, sustained voluntary muscle contraction has a transient facilitatory effect on transmitter release. Repetitive nerve stimulation (RNS) is a clinical neurophysiologic technique used to investigate NMT disorders. The timing of the calcium fluxes is extremely important in determining the response to different rates of RNS.

Vesicles of ACh are released sporadically and irregularly while the membrane is at rest and in flurries after nerve depolarization. Each vesicle contains about 5,000 to 10,000 molecules of ACh. Upon activation, the vesicles fuse with the presynaptic membrane and pour their ACh contents into the primary synaptic cleft. The molecules of ACh diffuse rapidly across the primary synaptic cleft and into the secondary clefts. Anywhere two molecules of ACh encounter an AChR, a chemical interaction takes place. This causes opening of sodium channels in the postsynaptic membrane, producing a brief nonpropagated localized depolarization. The depolarization produced by the contents of one vesicle is referred to as a miniature end-plate potential (MEPP). The summation of many MEPPs produces a localized, nonpropagated depolarization in the region of the end plate, referred to as an end-plate potential (EPP). The EPPs in turn summate, and if above threshold spawn a propagated, all-or-none muscle fiber action potential. The summated electrical activity of hundreds to thousands of muscle fiber action potentials produces the motor unit action potential, which can be recorded by needle electromyography.

The events of normal NMT are summarized in [Figure 23.7](#). Defects in NMT may develop at a number of points in the process.

Anatomy and Physiology of Muscle

A muscle is composed of hundreds to thousands of individual muscle fibers ([Figure 23.8](#)). Each fiber is a multinucleated syncytium, roughly cylindrical in

shape and encased in a connective tissue covering of endomysium, which extends over a long distance within a muscle fascicle. Fibers are polygonal in cross section; the diameter may vary depending on a number of factors but is relatively constant within a given muscle. A muscle fascicle is a group of fibers lying together within a sheath of perimysium. Intramuscular nerve twigs, capillaries, and muscle spindles also occupy the perimysium.

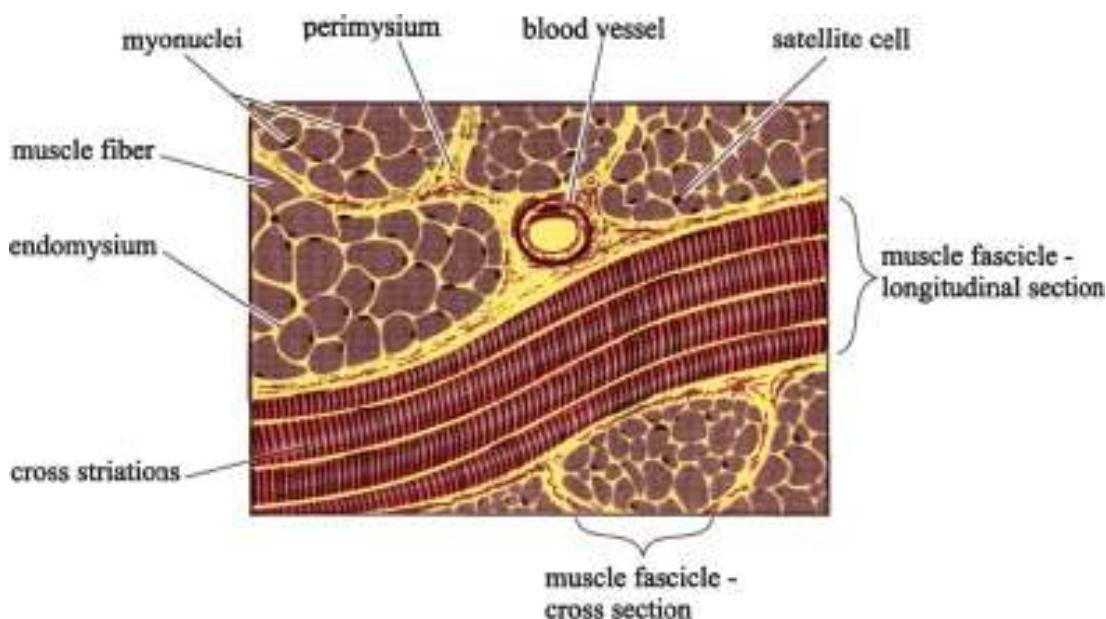


FIGURE 23.8 Cross section of muscle showing several fascicles, with muscle fibers, connective tissue septa, and blood vessels. Note the endomysium surrounding individual muscle fibers, the perimysium surrounding and separating fascicles, and the epimysium surrounding the entire muscle. Myonuclei and satellite cells lie peripherally and cannot be distinguished histologically. Longitudinal fibers demonstrate cross striations. (From Campbell WW. *Essentials of Electrodiagnostic Medicine*. Philadelphia: Lippincott Williams & Wilkins, 1999. Modified with permission of Dr. William W. Campbell.)

Epimysium separates groups of fascicles and also provides a covering for the entire muscle. The surface epimysium, which encases the muscle proper, is continuous with the fascia, which covers the muscle, and in turn with the tendons, which anchor it at the origin and insertion. The nuclei supporting a fiber lie peripherally just under the sarcolemmal membrane. Just external to the sarcolemma is the dense basement membrane. Satellite cells lie between the basement membrane and the sarcolemma. These dormant, omnipotent stem cells, whose nuclei resemble the sarcolemmal nuclei, can serve as the source of regeneration of muscle fibers following injury. Cellular organelles, glycogen granules, and lipids lie interspersed between the myofibrils and near the

sarcolemmal nuclei.

Each muscle fiber is composed of thousands of myofibrils, which are in turn made up of myriad myofilaments, the contractile elements ([Figure 23.8](#)). The myofibril is composed of repeating identical segments called sarcomeres. A sarcomere is anchored at each end by a condensation of protein referred to as a Z disk. From each Z disk arise thin filaments, made up of a long double helix of two chains of actin, which project toward the center of the sarcomere. From a condensation in the center of the sarcomere—the M line—thick filaments of myosin project outward toward the Z lines. Where the myosin and actin filaments overlap, the sarcomere appears denser and transmits less light—the anisotropic or A band. At the sarcomere's ends, where thin actin filaments exist alone, the appearance is lighter—the isotropic or I band. In the paramedian zone, where myosin filaments exist alone, the appearance is intermediate—the H zone. There are twice as many actin filaments as myosin filaments. During muscle contraction, the filaments slide past each other as side arms on the myosin molecule ratchet the actin molecule and draw it past. At maximal shortening, the Z disks are drawn together and the I bands are obliterated as the overall length of the sarcomere decreases ([Figure 23.9](#)).

Myosin is composed of two fragments: heavy meromyosin, which has ATPase activity, and light meromyosin, which does not. Actin is composed of three fragments: actin, troponin, and tropomyosin. Lying in the groove between the two chains in the actin are long filaments of tropomyosin molecules. Troponin molecules are small globular units located at intervals along the tropomyosin molecules. Troponin is made up of three subunits: troponin T, troponin I, and troponin C. Troponin T binds the troponin components to tropomyosin, troponin I inhibits the interaction of myosin with actin, and troponin C contains the binding sites for the Ca^{2+} that helps to initiate contraction. Troponin can reversibly bind with calcium. A troponin-tropomyosin complex inhibits the interaction of myosin and actin while the muscle is at rest. The binding of calcium to troponin disinhibits the interaction and allows reactions to occur between the cross bridges on the myosin molecule and active sites on the actin molecule. Other important skeletal muscle proteins are actinin, titin, and desmin. Actinin binds actin to the Z lines. Titin connects the Z lines to the M lines and provides a scaffold for the sarcomere. Desmin adds structure to the Z lines.

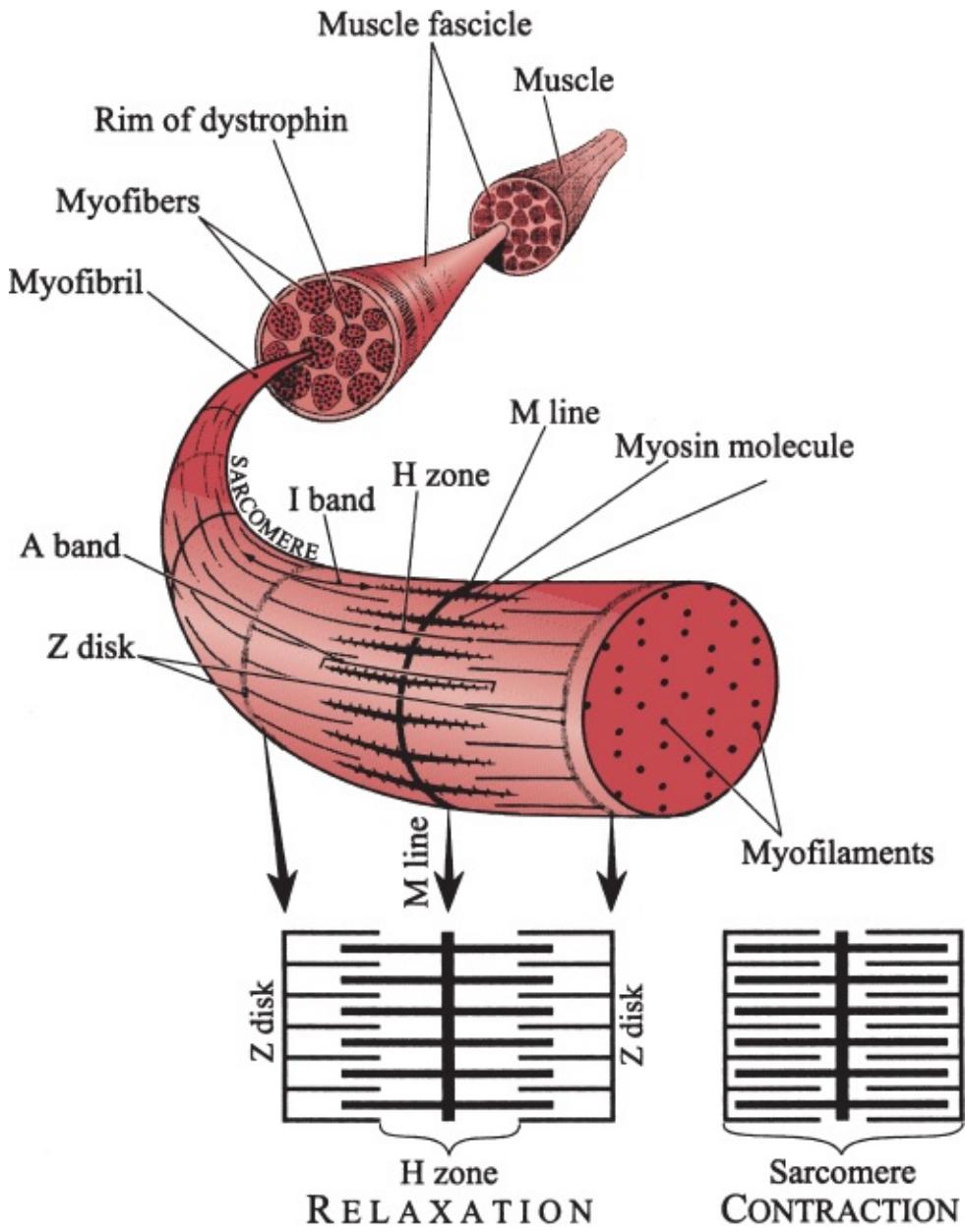


FIGURE 23.9 Myofibrils are composed of repeating sarcomeres. The sarcomere extends from Z line to Z line and consists of the I band (actin filaments only), the A band (actin and myosin filaments overlapping), the H zone (myosin filaments only), and the M line (a central condensation of the myosin filaments). The myosin molecules have cross bridges that interact with the actin molecules. When the muscle shortens, the overlapping of the myosin and actin molecules increases as the filaments slide, drawing the Z lines together and obliterating the I band. Dystrophin lies beneath the sarcolemma and helps reinforce it against stretching and buckling. (From Campbell WW. *Essentials of Electrodiagnostic Medicine*. Philadelphia: Lippincott Williams & Wilkins, 1999. Modified with permission of Dr. William W. Campbell.)

At the junction of the A and I bands, the transverse (T) tubular systems arise

as invaginations of the plasmalemma and ramify as an intricate network within the sarcomere. The T tubules allow communication between the muscle interior and the extracellular space and are the conduits along which the action potential is transmitted to the depths of the sarcomere. The sarcoplasmic reticulum (SR) is a closed internal labyrinth of vesicles that surrounds the myofibrils. The SR ends as focal dilatations, the terminal cisterns, which contain calcium. A pair of terminal cisterns abuts a T tubule to form a triad. The action potential conducted into the fiber along the T tubule causes calcium release from the terminal cisterns, which in turn activates myosin ATPase and initiates sliding of the filaments. This sequence is referred to as excitation contraction coupling. Following contraction, calcium ions are sequestered back into the terminal cisterns of the SR.

In addition to the contractile elements, skeletal muscle contains important cytoskeletal proteins, which help provide it structure. Elastic elements are vital to allow for contraction and relaxation. One of the key cytoskeletal proteins is dystrophin, a large molecule that forms a reinforcing meshwork just beneath the sarcolemma, and links the sarcomere to the sarcolemma and the extracellular matrix. Dystrophin is not directly connected to the membrane, but anchored to it at each end by a glycoprotein complex (dystrophin-associated glycoprotein), which spans the membrane and binds externally to laminin in the extracellular matrix. Dystrophin appears to lend mechanical support to the sarcolemma to help stabilize and brace it against the forces of muscle contraction. Genetic derangements of these cytoskeletal proteins underlie many of the muscular dystrophies.

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CHAPTER 24

The Spinal Cord Level

Above the motor unit, the next level of motor system integration is the spinal cord. The spinal cord begins at the cervicomedullary junction and ends at the conus medullaris. It is slightly flattened in an anteroposterior direction. Fissures and sulci mark the external surface of the spinal cord; most are of little clinical importance. A deep anterior median fissure and a posterior median sulcus partially divide it into two symmetrical halves. The anterior and posterior roots form the spinal nerves, which are segmentally arranged in 31 pairs. There are 8 pairs of cervical nerves, 12 thoracic, 5 lumbar, 5 sacral, and 1 coccygeal ([Figure 24.1](#)). Situated on each dorsal root is a dorsal root ganglion (DRG).

In newborns, the spinal cord may extend as far caudally as L3. During maturation, the vertebral column elongates more than the spinal cord, and the adult spinal cord is about 25 cm shorter than the vertebral column. The variability of the lower level has some correlation with the length of the trunk, especially in females. Radiographically, the conus is usually seen at the L1-L2 interspace in adults; if the level of the tip of the conus is below the mid-L2 vertebral body, the conus is considered low-lying. Because the spinal cord normally ends at the level of the L1-L2 interspace, lumbar punctures are done well below this level.

The length discrepancy between the spine and the spinal cord creates a difference between the segments of the spinal cord and the vertebral level that progressively increases from rostral to caudal. In the upper cervical area, the numerical cord level is about one segment greater than the corresponding vertebral spinous process (e.g., the C5 spinous process lies at the C6 segment of the spinal cord). In the lower cervical and thoracic areas, there is a difference of about two segments; in the lumbar region, there is a difference of almost three segments ([Figure 24.1](#)). Because of the offset, the spinal nerves below the cervical region course downward before exiting through the intervertebral

foramina. Nerve roots exit through the foramen above the vertebra of like number from C1 to C7. The C8 root exits below C7 and sets the pattern of root exit below like-numbered vertebra followed down the rest of the vertebral column. The lumbar and sacral roots descend almost vertically to reach their points of exit. These long trailing roots from the lower cord segments make up the cauda equina.

The spinal cord is greater in width and diameter in the cervical and lumbosacral regions, forming the cervical and lumbar enlargements, the site of nuclear centers that supply the extremities. The cervical enlargement extends from the C3-T2 spinal cord segments; it innervates muscles of the upper limb ([Figure 24.2](#)). The lumbar enlargement extends from the L1-S3 spinal cord segments; it innervates muscles of the lower limb. The segments of the cervical enlargement match fairly well with the corresponding vertebral levels. The lumbar enlargement extends over vertebral levels T9-T12. Below T12, the spinal cord tapers to form the conus medullaris.

Each segment of the spinal cord gives rise to a mixed spinal nerve that contains motor, sensory, and autonomic fibers ([Figure 24.3](#)). Motor axons arising from the anterior horn cells of the spinal cord travel in the converging filaments of the anterior spinal root. On each posterior root, within the intervertebral foramen and just proximal to the junction with the anterior root, lies a DRG. The DRG is made up of unipolar neurons, and the posterior roots are made up of the central processes of these neurons. Acetylcholine is the only neurotransmitter in the anterior roots; the posterior roots contain several, including substance P, glutamate, calcitonin gene-related peptide, vasoactive intestinal polypeptide, cholecystokinin, somatostatin, and dynorphin. The anterior roots convey motor and autonomic fibers into the peripheral nerve; they join the posterior root to form the mixed spinal nerve. In the thoracolumbar region, white and gray rami communicantes connect the spinal nerve to the paravertebral sympathetic chain ([Figure 24.3](#)).

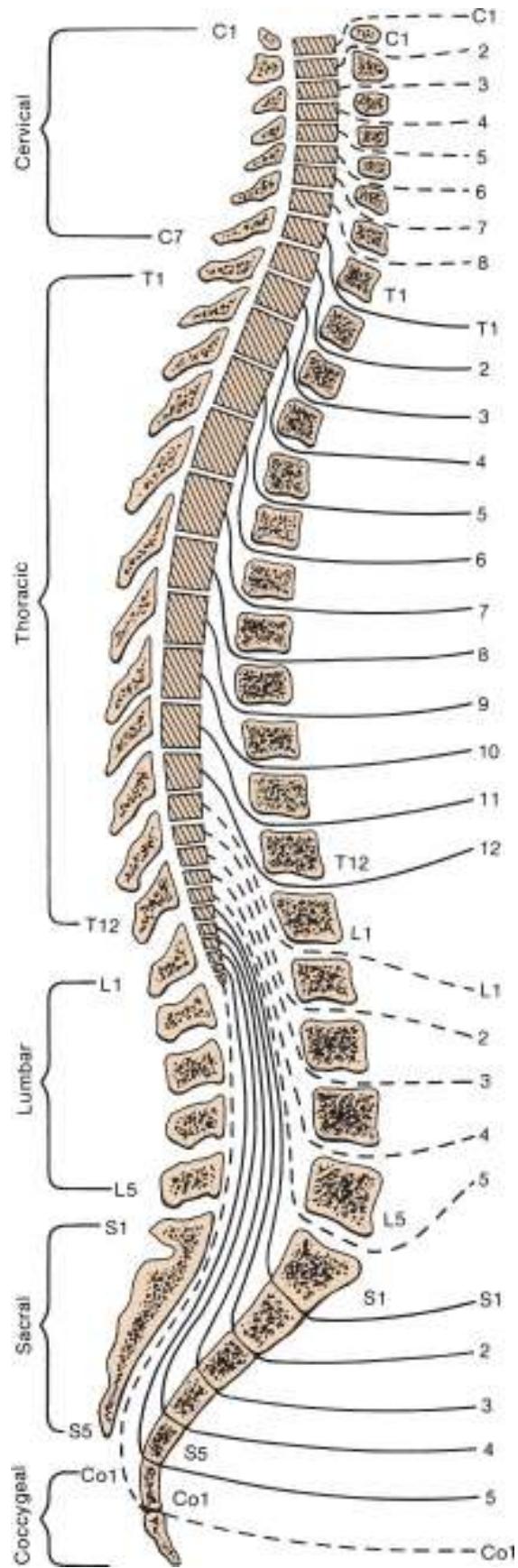


FIGURE 24.1 The relationship of the spinal cord segments and spinal nerves to the vertebral bodies and spinous processes.

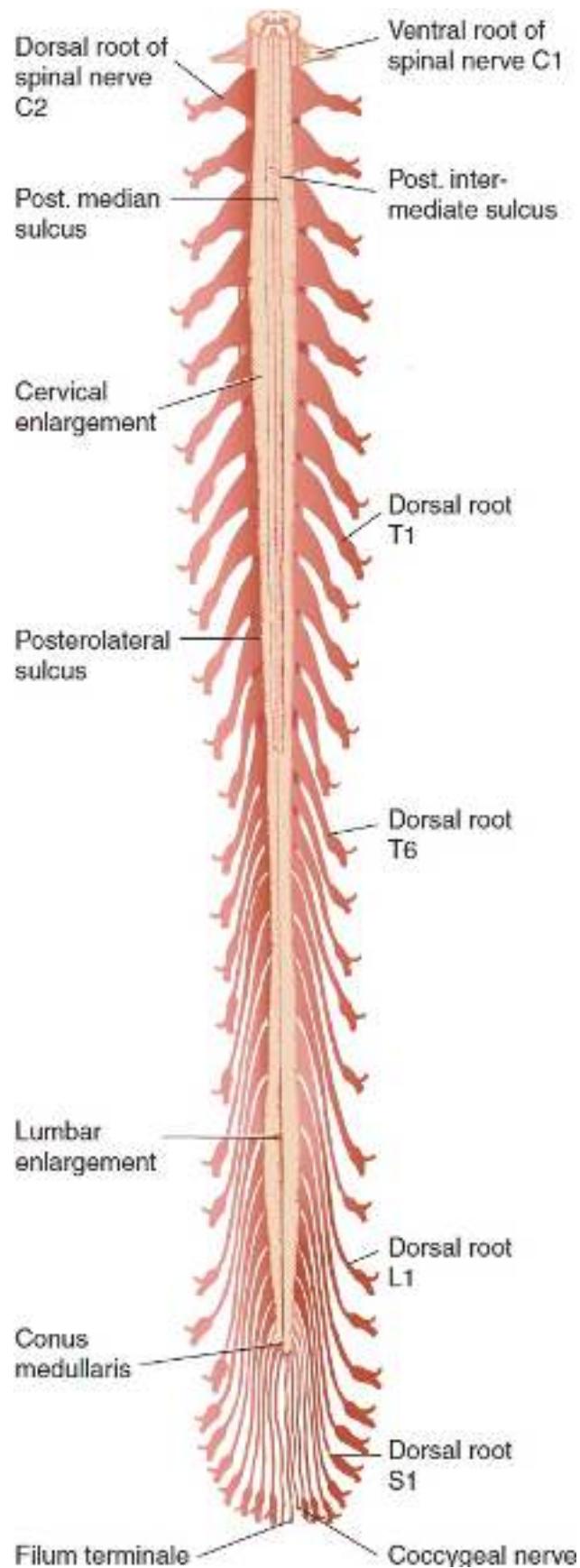


FIGURE 24.2 Posterior view of the spinal cord showing attached dorsal root filaments and spinal ganglia. Letters and numbers indicate corresponding spinal nerves. (Modified from Carpenter MB, Sutin J. *Human Neuroanatomy*. 8th ed. Baltimore: Williams & Wilkins, 1983, with permission.)

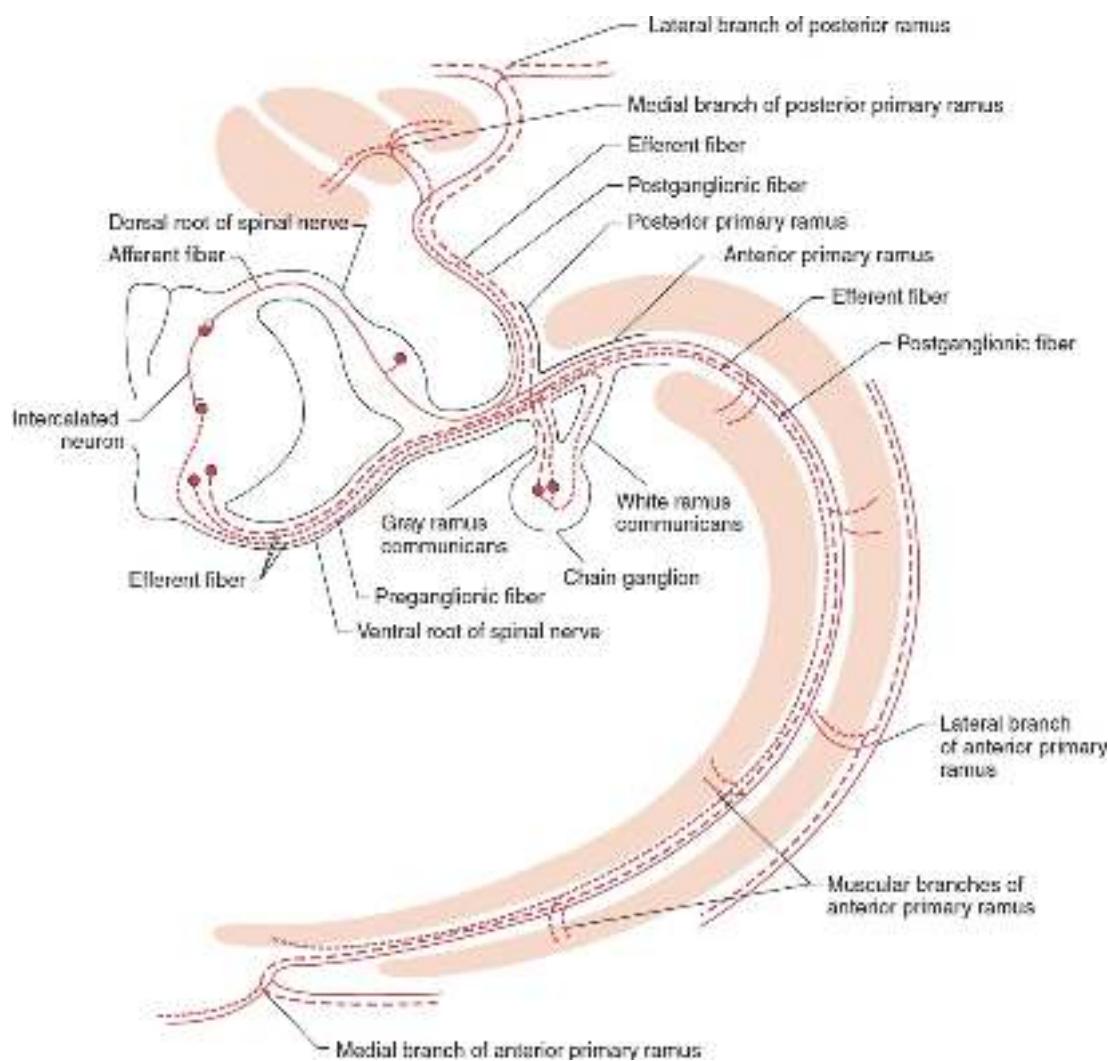


FIGURE 24.3 Segmental spinal nerve showing the course of motor, sensory, and preganglionic and postganglionic sympathetic fibers.

The Bell-Magendie law (for Sir Charles Bell and Francois Magendie) states that the anterior roots are motor and the posterior roots sensory; this discovery was one of the seminal developments in early neurobiology. However, it now appears there may be some afferent nerve fibers in the anterior roots, and up to 3% of the fibers in the posterior root may be efferent. The roots pass through the dura separately and then unite in the intervertebral foramina just distal to the DRG to form the mixed spinal nerve. After the mixed spinal nerve exits the

intervertebral foramen, it divides into anterior and posterior primary rami. The smaller posterior primary rami supply the skin of the back and the paraspinal muscles. The anterior primary rami are continuations of the mixed spinal nerves and supply motor and sensory innervation to all other structures of the body. The anterior primary rami from the cervical and lumbar enlargements form the brachial and lumbosacral plexuses that innervate the limbs. The anterior primary rami of the thoracic segments of the spinal cord continue as the intercostal nerves. The anterior primary ramus of the mixed spinal nerve is sometimes referred to as a root, especially by surgeons and especially regarding the brachial plexus. When anatomy sources say that the C5 and C6 roots join to form the upper trunk, they are actually referring to the anterior primary rami of the spinal nerve. Similarly, when the dorsal scapular nerve is said to arise from the C5 root.

The delicate pia-arachnoid closely invests the spinal cord. The tough, fibrous membrane of the dura mater forms a firm, tubular sheath around the exiting nerve roots. The spinal cord is separated from the walls of the vertebral canal by the epidural space, which contains areolar tissue and a plexus of veins. The subdural space is a potential space containing a small amount of fluid. The subarachnoid space is a well-defined cavity containing cerebrospinal fluid that extends to about the level of the second sacral vertebra, forming the lumbar thecal sac. Systemic malignancies frequently metastasize to the capacious spinal epidural space. Spinal hematomas may accumulate in the epidural, subdural, or subarachnoid space. Subarachnoid hematomas can extend along the entire length of the subarachnoid space. Epidural and subdural spinal hematoma present with intense, knifelike pain at the location of the hemorrhage (“coup de poignard”) followed by progressive paralysis below the affected level. Spinal epidural abscess classically causes a triad of fever, back pain, and a neurologic deficit.

A section of spinal cord where the rootlets of a pair of spinal nerves attach is referred to as a segment, although there is no internal demarcation to separate one segment from another. Each spinal cord segment can function as an independent entity for purposes of some very basic functions, such as the segmental muscle stretch reflex. Each segment controls the resting muscle tone of the muscles it innervates. The motor units supplying the myotomal muscles innervated by a segment carry out voluntary activity. The motor function of a spinal cord segment is modulated and influenced by suprasegmental impulses from several descending motor tracts.

The spinal cord parenchyma consists of an H- or butterfly-shaped core of gray matter that contains nerve cells, surrounded by white matter made up of

longitudinally arranged ascending and descending nerve fibers, mainly myelinated. The relative proportion of white to gray matter varies depending on the cord level. In the center of the gray matter, running throughout the entire length of the cord and for a short distance into the filum terminale, is a minute central canal consisting of a single layer of ependymal cells. The two halves of the spinal cord are joined by a commissure made up of a core of gray matter surrounded by anterior and posterior white commissures. Internally, the white matter of the spinal cord is divided into posterior, lateral, and anterior funiculi. The posterior funiculus extends from the posterior median sulcus to the attachment of the posterior rootlets at the posterolateral sulcus. Rostral to the upper thoracic segments, a posterior intermediate sulcus separates the medial fasciculus gracilis from the lateral fasciculus cuneatus. The lateral funiculus lies between the attachments of the posterior and anterior spinal rootlets. The anterior funiculus extends from the anterior rootlets to the anterior median fissure.

The spinal cord gray matter consists of anterior and posterior horns with a lateral concavity. In the thoracic and upper lumbar regions, an intermediolateral column of autonomic neurons forms a small projecting lateral horn between the anterior and posterior horns. The sympathetic axons project through the anterior horn and anterior root and then through the gray rami communicantes to enter the sympathetic chain ganglia. The gray matter contains neurons, nerve fibers, supporting neuroglia, and blood vessels. Neurons are not distributed uniformly but are collected into functional groups that consist of columns of cells extending over many segments ([Figure 24.4](#)). The most elementary division is into posterior horns that contain sensory neurons and anterior horns that contain motor neurons. The posterior horn is relatively narrow and capped by a thin crescent of tissue, the substantia gelatinosa (of Rolando). The tip of the posterior horn is separated from the surface by a thin white matter tract, the dorsolateral tract (of Lissauer).

Within the anterior horn, there are alpha motor (skeletomotor) neurons, gamma motor (fusimotor) neurons, beta motor neurons, and interneurons. The alpha motoneurons innervate common, extrafusal striated skeletal muscle; the gamma motoneurons innervate intrafusal, muscle spindle fibers (L. *fusus* “spindle”). Beta motor neurons innervate both intrafusal and extrafusal fibers. All of these fibers are classified as general somatic efferent. At any given level, there is a somatotopic arrangement of motor neurons. A medial cell group that extends throughout the length of the spinal cord innervates the trunk and

proximal muscles. A lateral cell group found only in the cervical and lumbar enlargements innervates limb muscles. The expansion of the anterior horns in the cervical and lumbar enlargements reflects the presence of this lateral cell column that supplies limb muscles. In both the cervical and lumbar enlargements, neurons innervating proximal muscles are located more rostrally and those innervating distal muscles more caudally; cells supplying extensors are more ventral than cells innervating flexors. Some motor neurons are aggregated into well-defined nuclear groups. The phrenic nucleus is a central collection of cells from C3 to C7 that innervates the diaphragm. Onuf's nucleus is a ventrolateral cell group at S1 and S2 that supplies the striated muscles of the perineum. For unknown reasons, the cells of Onuf's nucleus are relatively spared in motor neuron disease but disproportionately involved in multiple system atrophy.

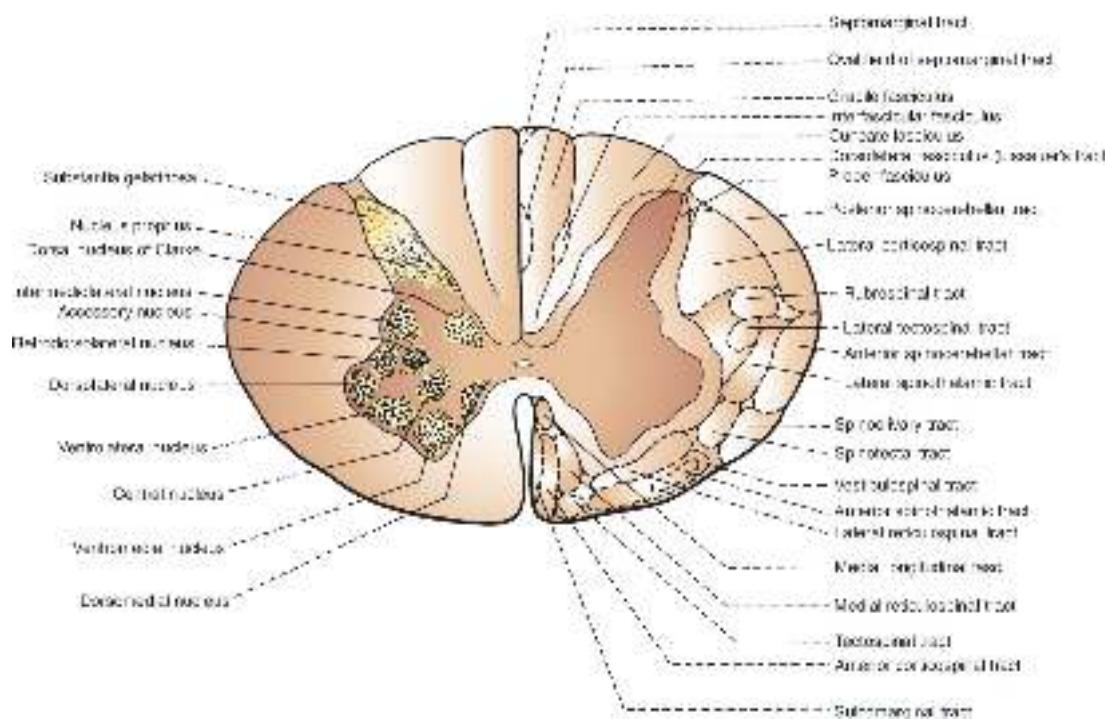


FIGURE 24.4 Cross section of the spinal cord showing the arrangement of cellular groups in the gray matter and fiber pathways in the white matter.

The collections of neurons in the gray matter of the spinal cord are not as well defined as nuclei in other parts of the nervous system. In experimental animals, Rexed identified 10 regions, or laminae, of the spinal cord gray matter. Rexed laminae are more commonly used in the study of spinal cord neurons than the named nuclei (Figure 24.5, Box 24.1). There is evidence supporting the laminar scheme in humans as well. Laminae I to IV make up most of the posterior horn

and receive cutaneous primary afferents. Lamina IX contains the motor neurons that supply striated muscles. Small motor neurons tend to innervate type S (slow twitch, fatigue resistant) motor units, and larger neurons type FF (fast twitch, fatigue sensitive) and FR (intermediate, fast twitch but more fatigue resistant than FF) motor units.

The white matter of the spinal cord consists of ascending and descending long fiber tracts and short intersegmental and intrasegmental tracts ([Figure 24.4](#)). The ascending pathways carry sensory impulses of various types from the extremities, trunk, or neck to higher centers. The major ascending tracts are the posterior columns, the spinothalamic/anterolateral system, and the spinocerebellar tracts.

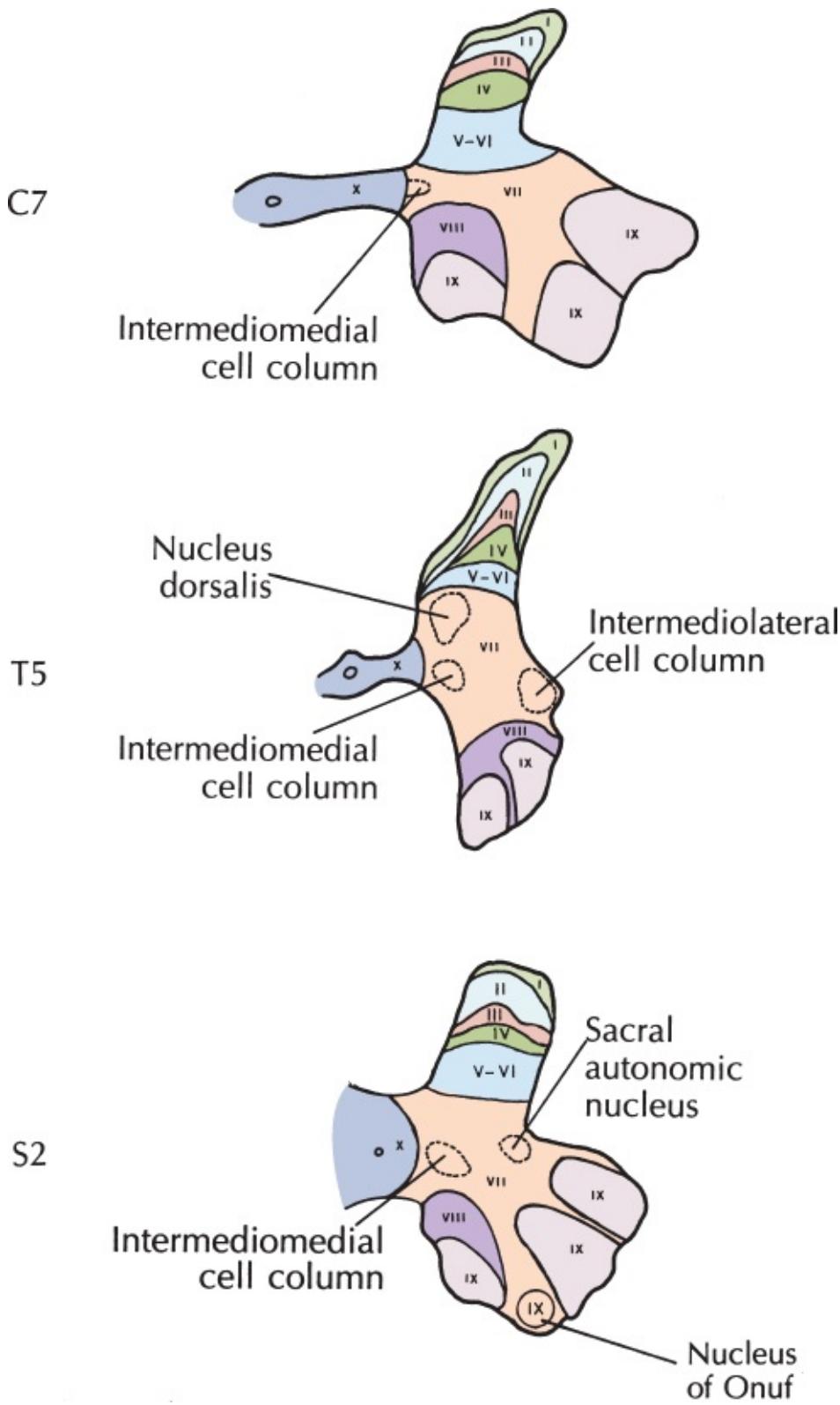


FIGURE 24.5 Positions of the Rexed cytoarchitectonic laminae at three levels of the spinal cord gray matter. (Modified from Kiernan JA. *Barr's The Human Nervous System: An Anatomical Viewpoint*. 9th ed. Philadelphia: Wolters Kluwer Health/Lippincott Williams & Wilkins, 2009, with permission.)

BOX 24.1

The Rexed Laminae

Lamina I consists of scattered large neurons in the superficial region of the posterior horn that receive afferents from Lissauer's tract; these neurons are primarily involved with pain impulses. Lamina II, with overlap into lamina III, is approximately coterminous with the substantia gelatinosa. These regions contain interneurons that receive pain and temperature afferents from Lissauer's tract but make no contribution to the long ascending sensory pathways. Lamina IV contains polymodal, or wide dynamic range, sensory neurons that are activated by many different stimuli. Cells in lamina IV give rise to the contralateral spinothalamic tract. Laminae V and VI are indistinguishable in humans. Cells in laminae IV and V to VI make up the nucleus proprius. Laminae V to VI receive many descending fibers, particularly those of the corticospinal tract. Lamina VII is the most extensive cytoarchitectonic layer. Between C8 and L3, the medial portion of lamina VII contains the nucleus dorsalis (of Clarke), which is the origin of the posterior spinocerebellar tract. Laterally in lamina VII is the intermediolateral gray column that contains preganglionic sympathetic neurons between T1 and L2 and preganglionic parasympathetic neurons between S2 and S4. Lamina VIII contains interneurons involved in motor control, including Renshaw cells that receive collaterals from nearby lamina IX motor neurons. Lamina IX contains the motor neurons. There is a medial nuclear group at all levels that innervates axial and proximal muscles and a lateral nuclear group in the cervical and lumbar enlargements that innervates distal extremity muscles. Lamina X surrounds the central canal and contains small neurons that receive dorsal root afferents involved with pain, temperature, and visceral sensation.

The descending pathways carry impulses from higher centers; these terminate in spinal cord nuclei on which they have regulatory and inhibitory functions. The major descending pathway is the pyramidal tract, which includes the lateral corticospinal (crossed pyramidal) and ventral corticospinal (uncrossed pyramidal) tracts. The lateral corticospinal tract is a massive bundle taking up

most of the lateral funiculus of the cord. It contains descending pyramidal tract fibers from the giant pyramidal Betz cells in the motor cortex, but these comprise only 3% of the bundle; fibers making up the bulk of the tract come from other cortical areas. Lateral corticospinal axons drop off to innervate segmental motor neurons all along the cord, so the tract becomes progressively smaller as it descends.

In humans, corticospinal tract fibers originating in areas 4 and 6 synapse not only with interneurons but directly with the large, multipolar, spinal motor neurons in lamina IX. The direct synapse with anterior horn motor neurons is one of the defining characteristics of the corticospinal system. The direct projections from the precentral gyrus to spinal motor neurons are concerned with discrete, fractionated limb movements and fine motor control, and the distribution of precentral corticospinal fibers is primarily to motor neurons that supply distal extremity muscles. Other descending tracts that influence segmental spinal cord motor activity include the rubrospinal, vestibulospinal, medial and lateral reticulospinal, olivospinal, and the tectospinal tract. Fractionated, fine movements of distal extremity muscles are controlled primarily by the corticospinal and rubrospinal tracts; control of proximal and postural muscles is primarily by extrapyramidal pathways, particularly the reticulospinal and vestibulospinal tracts. There are other less well-defined tracts, as well as intersegmental, intrasegmental, and association pathways. There is some intermingling of fibers within the various tracts, and individual pathways are not as distinctly delineated as diagrams would indicate.

BLOOD SUPPLY

There is some individual variation in the blood supply of the spinal cord ([Figure 24.6](#)). The anterior spinal artery is formed by the union of the arterial branches that pass caudally from each vertebral artery and unite in the midline near the foramen magnum. It descends the entire length of the spinal cord, taking a somewhat undulating course, and lies in or near the anterior median fissure. Below the fourth or fifth cervical segment, the anterior spinal artery is fed or reinforced by unpaired anterior medullary arteries that arise from the lateral spinal arteries. These latter vessels enter the vertebral canal through the intervertebral foramina, and in the cervical region are branches of the ascending cervical artery; in the thorax, of the intercostals; and in the abdomen, of the

lumbar, iliolumbar, and lateral sacral arteries. They pierce the dural sheaths of the spinal roots and split into anterior and posterior radicular branches. The radicular arteries are asymmetric and sometimes absent. The largest medullary artery, the great anterior radicular artery of Adamkiewicz, arises between T9 and L2, usually on the left side, and supplies the lumbar enlargement. The blood supply to any given level of the spinal cord is proportional to its cross-sectional area of gray matter, and the caliber of the anterior spinal artery is largest at the level of the lumbar and cervical enlargements.

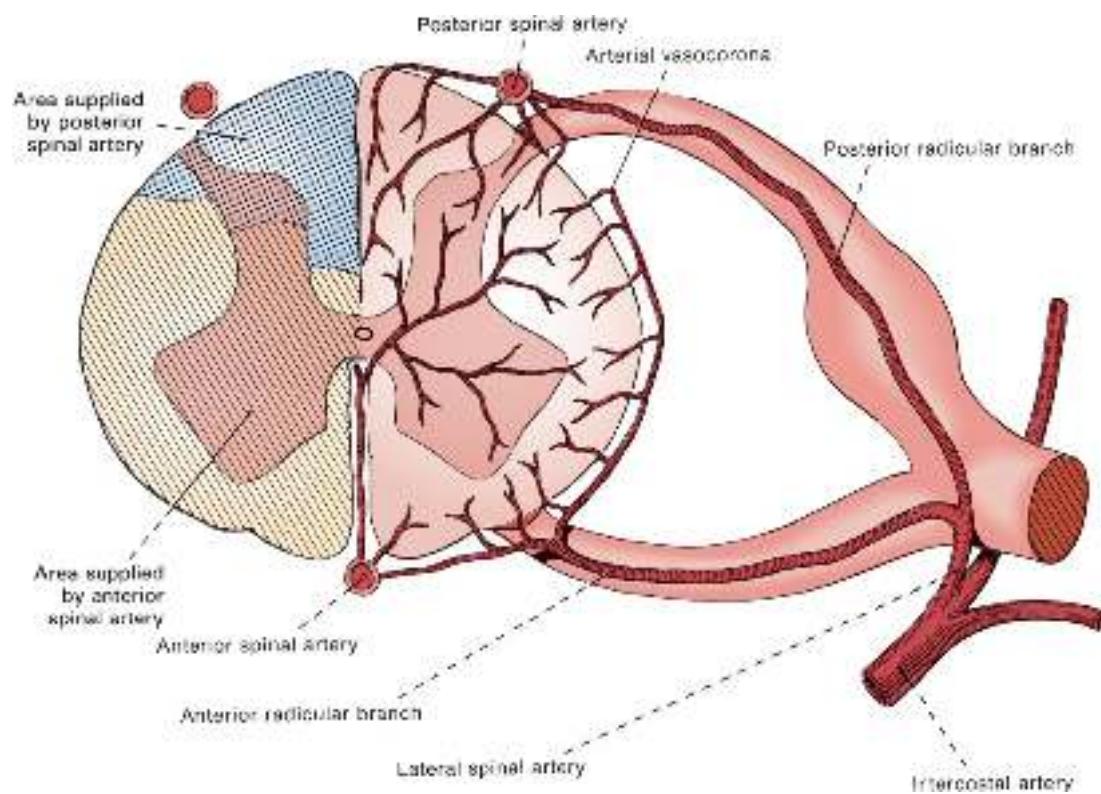


FIGURE 24.6 Arterial supply of the spinal cord.

The posterior spinal arteries are really plexiform channels rather than distinct single vessels that lie near the posterolateral sulci and the entrance of the rootlets of the posterior spinal nerves. They also arise from the vertebral arteries, and posterior medullary arteries join them at irregular intervals. Central arteries, branches of the anterior spinal, are given off alternately to the right and left halves of the spinal cord at different levels to supply the anterior and central portions of the cord. Branches of the anterior and posterior spinal arteries form a peripheral anastomosis, the arterial vasocorona, which supplies the periphery of the cord, including the lateral and ventral funiculi. This anastomosis is least

efficient in the region of the lateral columns. Within the substance of the cord, the posterior spinal arteries supply the posterior horns and most of the dorsal funiculi; the anterior spinal artery supplies most of the remainder of the cord. Certain boundary zones between ascending and descending sources of the blood supply are sites of least adequate circulation in the spinal cord. The cervical and lumbosacral spinal cord are more richly vascularized than the thoracic cord. The upper thoracic segments near T4 have been traditionally thought particularly vulnerable to ischemia. More recent evidence finds the lower thoracic or upper lumbar cord more vulnerable. Patients with myelopathy after cardiac arrest or severe hypotension were found to have predominant involvement of the lumbosacral level with relative sparing of the thoracic region; none of the patients had damage isolated to the thoracic region. In a series of 44 cases of spinal cord infarction, the mean level of deficit in cases of global ischemia was at T9.

The venous drainage of the spinal cord courses from the capillary plexuses to peripheral venous plexuses that correspond somewhat to the arterial supply. The major portion of the venous drainage takes place through the intervertebral foramina into veins in the thoracic, abdominal, and pelvic cavities, but the valveless spinovertebral venous plexus (Batson's plexus) also continues upward into the intracranial cavity and venous sinuses and may be a means of transport of tumor cells. Abnormalities of the cerebrospinal venous system have been implicated in the pathogenesis of multiple sclerosis (MS).

PHYSIOLOGY AND PATHOPHYSIOLOGY

Spinal reflexes are responses to stimuli that are mediated at the spinal cord level. A spinal reflex may be monosynaptic, with an afferent and efferent limb mediated by only two neurons joined by a single synapse, or polysynaptic when one or more interneurons are involved. A spinal reflex may be segmental (intrasegmental), mediated at only one level, or intersegmental, with several levels participating. Long-loop reflexes involve circuits that reach as far as the cerebral cortex, allowing modulation of spinal reflexes by suprasegmental mechanisms.

The motor unit consists of an alpha motor neuron and all its subject muscle fibers ([Chapter 23](#)). A parallel system of innervation arises from gamma motor neurons that innervate muscle spindles. Muscle spindles, or neuromuscular

spindles, are small structures (1 to 3 mm long in small muscles, 7 to 10 mm long in large muscles), composed of specialized muscle fibers. Spindles lie widely interspersed in the muscle, affixed with connective tissue parallel to the larger, extrafusal muscle fibers. Muscles that require finely graded contractile control, such as small hand muscles, have a greater density of spindles. The extrafusal fibers provide the force for muscle contraction; the muscle spindles provide modulation and control of that force. They also help regulate the underlying tone in the muscle. Muscle contraction always occurs against some setting of background tone. When the underlying, background level of muscle tone is either too high or too low, voluntary activity cannot occur with normal effectiveness. Thus, the gamma efferent system is a critical component of motor control. The descending extrapyramidal motor pathways, such as the reticulospinal and vestibulospinal tracts, have an important influence on the gamma efferent system. Pathways from the motor cortex and cerebellum tend to influence both the alpha and gamma motor neurons simultaneously, an effect referred to as alpha-gamma coactivation.

Muscle spindles consist of a small group of intrafusal muscle fibers surrounded by a connective tissue capsule. Tonic firing of the gamma motor neuron produces slight contraction of the intrafusal fibers, leaving them under tension. The spindles send information regarding the level of tension to the spinal cord. There are two primary types of intrafusal fibers, nuclear chain and nuclear bag, in a ratio of 3 to 4:1. In nuclear bag fibers, a collection of myonuclei creates a bulge in the fiber's center; in nuclear chain fibers, the myonuclei are linear. There are two types of nuclear bag fibers: bag 1 and bag 2. The bag 2 fibers are smaller than the bag1 fibers, intermediate in size between the bag 1 fibers and the nuclear chain fibers. Muscle spindles supply the nervous system with information about the length of a muscle and, if the length is changing, about the rate of change. The bag 1 fibers convey dynamic information about changes in length; the bag 2 fibers relay data about static muscle length. Nuclear chain fibers respond only to static muscle length.

The efferent nerve supply to the muscle spindles arises from gamma or beta motor neurons. There are two types of efferent nerve ending configurations: plate endings occur primarily on nuclear bag fibers; trail endings are common on both bag and chain fibers. Afferent nerve fibers from the muscle spindles are group Ia (primary spindle afferents) and group II (secondary spindle afferents). The group Ia afferents arise from primary annulospiral endings that form spirals around the equatorial region of nuclear bag fibers; the group II afferents arise

from both annulospiral and flower spray endings, primarily on nuclear chain fibers. Primary spindle afferents are large, heavily myelinated, rapidly conducting fibers; they are the fastest conducting fibers in the peripheral nervous system. Centrally, they make monosynaptic contact with alpha motor neurons innervating agonist and synergist muscles. They also send collaterals to the gamma motor neurons innervating the same muscle, as well as inhibitory collaterals to alpha motor neurons innervating antagonist muscles (reciprocal inhibition). Secondary spindle afferents make monosynaptic contact only with agonist muscle motor neurons. Spindle afferents are classified as general somatic afferent type fibers.

Renshaw described the effect of the discharge of motoneurons on neighboring motoneurons. Alpha motor neurons give off collateral fibers that synapse on nearby inhibitory interneurons, which then modulate the discharge of the alpha motor neuron (recurrent inhibitory Renshaw loop, lateral inhibition). Renshaw cells are present in laminae VII and VIII, immediately medial to the motor neurons in lamina IX. The synapse of the alpha motor neuron on the Renshaw cell is cholinergic. Both glycine and gamma-amino butyric acid (GABA)—inhibitory amino acids—appear to play a role in the recurrent inhibition mechanism of Renshaw cells on motor neurons. Autoantibodies to glutamic acid decarboxylase—a key enzyme in GABA synthesis—cause stiff-person syndrome, a condition of generalized stiffness and increased tone because of impaired inhibitory mechanisms.

Another important component in this system is the Golgi tendon organ (GTO). The GTO, from its position in the tendon, further helps to regulate muscle tone. In contrast to the parallel arrangement of the muscle spindles, GTOs are connected in series with the muscle. The GTO is a mechanism for force feedback to the contracting muscle. It may also serve as a protective mechanism against overstretch of the muscle tendon, either from active contraction of the muscle or from passive stretch. When tension in the tendon increases beyond a certain level, afferent traffic travels centrally via group Ib fibers from the GTO to inhibit contraction of the agonist and to cause contraction of the antagonist. The inhibition of the agonist (autogenic inhibition) is mediated by glycinergic interneurons. For force generated by active muscle contraction, autogenic inhibition helps to unload the tension on the tendon by causing the muscle to relax. The effects of the GTO/Ib fiber system are opposite (inhibition of the agonist) those of the muscle spindle/Ia fiber system (facilitation of the agonist). Further modulation of the motor system at the local segmental level is

provided by afferents coming from the skin and joints that help to convey additional information regarding the position of the limb in space.

The simplest example of spinal cord segmental modulation of motor unit activity is the monosynaptic stretch reflex. If a muscle is suddenly stretched, as by percussion of its tendon with a reflex hammer, the passive stretch of the tendon stretches the muscle belly, which in turn leads to passive stretch of the muscle spindles. This lengthening of the intrafusal fibers triggers a volley of impulses in the primary spindle afferents. These synapse with alpha motor neurons innervating the muscle. The alpha motor neurons fire, producing a contraction of the muscle, which then, because of the parallel configuration of intrafusal and extrafusal fibers, unloads or takes the stretch off the muscle spindles. The muscle then returns to a state of relaxation. The sequence of percussion, contraction, and then relaxation is a muscle stretch or myotatic reflex (myo + Gr. teinein “to stretch”). Because the muscle stretch is produced by percussion of the tendon, the terms tendon reflex and deep tendon reflex are also commonly used. The contraction of the agonist may be accompanied by relaxation of the antagonist mediated by inhibitory interneurons (inverse myotatic reflex). If the resting tone of the muscle spindles is increased, and the tension of the intrafusal fibers at a higher than normal level, then the additional passive stretch due to percussion of the tendon produces a markedly exaggerated response. This is hyperreflexia, which is seen with upper motor neuron lesions.

In addition to the monosynaptic reflex arc, there are complex polysynaptic spinal reflexes that involve excitation or inhibition of agonists, synergists, and antagonist muscles and even contralateral muscles. The withdrawal reflex consists of a movement to escape from a cutaneous stimulus, usually noxious (e.g., flexion of the lower extremity in response to a painful stimulus on the sole of the foot). Afferent cutaneous nociceptive fibers synapse on both excitatory and inhibitory interneurons, which cause flexion of the hip and thigh, and dorsiflexion of the foot, with appropriate inhibition of their antagonists. The reflex activity is intersegmental, spread over several cord segments. More complex yet is the crossed extensor (Phillipson) reflex, in which withdrawal of the ipsilateral limb is accompanied by extension of the contralateral limb to provide body support whereas the stimulated limb escapes the provoking trigger. The crossed extensor reflex is not merely polysynaptic and intersegmental; the contralateral side of the spinal cord also participates. The crossed flexor reflex is when the contralateral leg flexes rather than extends.

CLINICAL MANIFESTATIONS OF DISINHIBITED SPINAL CORD SEGMENTS

The activity of the motor neurons in the spinal cord is regulated and modulated by the descending motor pathways. When the influence of the descending motor pathways is removed, as in a spinal cord injury, the result is a disinhibition of the segmental motor neuron pools below the level of the lesion, resulting in a higher level of gamma efferent resting traffic. This increases the gain on the muscle spindles, leaving them under an increased level of resting tone, which leads to spasticity and hyperreflexia.

Segmental spinal cord reflexes can be responsible for fairly elaborate motor phenomena. A dog making vigorous kicking movements with one leg in response to being scratched is exhibiting a complex, polysynaptic, intersegmental spinal reflex. The descending motor pathways—particularly the pyramidal tract—in general inhibit segmental reflexes and act to suppress excess activity. Because descending motor pathways normally suppress segmental activity, the eloquence of spinal reflexes is seen to best advantage when suprasegmental control is defective. In the immature nervous system of the neonate, suprasegmental pathways are not fully developed; a variety of spinal reflexes occur normally, such as stepping and placing reactions, crossed extensor reflex, and the tonic neck reflex. In the normal neonate, the crossed extensor and placing/stepping reactions disappear by 1 to 2 months of age, the tonic neck reflex by 3 months, and the extensor plantar response by 12 months. In patients with severe myelopathy that has interrupted the suprasegmental pathways rostrally, spinal reflex activity may be prominent, including withdrawal and crossed extensor reflexes. The Babinski's sign and related extensor plantar responses are fragments of the withdrawal reflex that occur with damage to the descending motor pathways. A more fully developed variation is the triple flexion response, in which extension of the great toe is accompanied by dorsiflexion of the foot and flexion of the knee and hip, essentially a withdrawal reflex. Brain-dead patients can display impressive spinal reflex movements, including the dramatic "Lazarus sign" (bilateral arm flexion, shoulder adduction, raising of the arms, and crossing of the hands). These are all ostensibly due to local spinal cord reflexes, which have become autonomous and are under no suprasegmental control.

SPINAL CORD SYNDROMES AND DISORDERS

Common or classic spinal cord syndromes include transverse myelopathy, Brown-Séquard syndrome, central cord syndrome, syringomyelic syndrome, anterior cord syndrome, posterior column syndrome, posterolateral column syndrome, anterior horn syndrome, and anterior horn-corticospinal tract syndrome.

Complete transverse myelopathy causes total loss of function below the level of the lesion; with incomplete myelopathy, there is some preservation of function (see [Chapter 53](#)). Common etiologies include trauma, cord compression, and myelitis, due to MS, neuromyelitis optica, a parainfectious event, or an isolated clinical syndrome. Patients present with acute transverse myelopathy if compression involves the spinal cord proper. If compression involves the cauda equina, the patient presents with a cauda equina syndrome rather than a transverse myelopathy (see [Chapter 47](#)). Longitudinally extensive transverse myelitis refers to florid and widespread inflammation of the spinal cord causing T2 hyperintensity on spinal magnetic resonance imaging that is seen to extend over three or more vertebral segments. It is classically associated with neuromyelitis optica, but there are many other causes, including other inflammatory etiologies, infection, malignancy, and metabolic disturbances.

Brown-Séquard described the clinical picture that follows functional hemisection of the spinal cord. It is actually more often seen with extramedullary tumor compression than with trauma. Patients with Brown-Séquard syndrome have corticospinal tract and posterior column dysfunction ipsilateral to the lesion and spinothalamic tract-mediated pain and temperature loss contralateral to the lesion. There may be evidence of root dysfunction at the level of the lesion. Brown-Séquard-plus syndrome is associated with additional neurologic findings involving the eyes, bowel, or bladder.

Central cord syndrome is one of the regularly recurrent variants seen with incomplete cervical spinal cord injury; it involves necrosis with softening of the central aspect of the spinal cord, with relative sparing of the periphery. Patients have segmental weakness at the involved level because of anterior horn gray matter necrosis, with only minor long tract findings, that is, they are not paraplegic or quadriplegic. The segmental weakness typically involves the hands and distal upper extremities. Hand weakness may also occur with a lesion several segments higher.

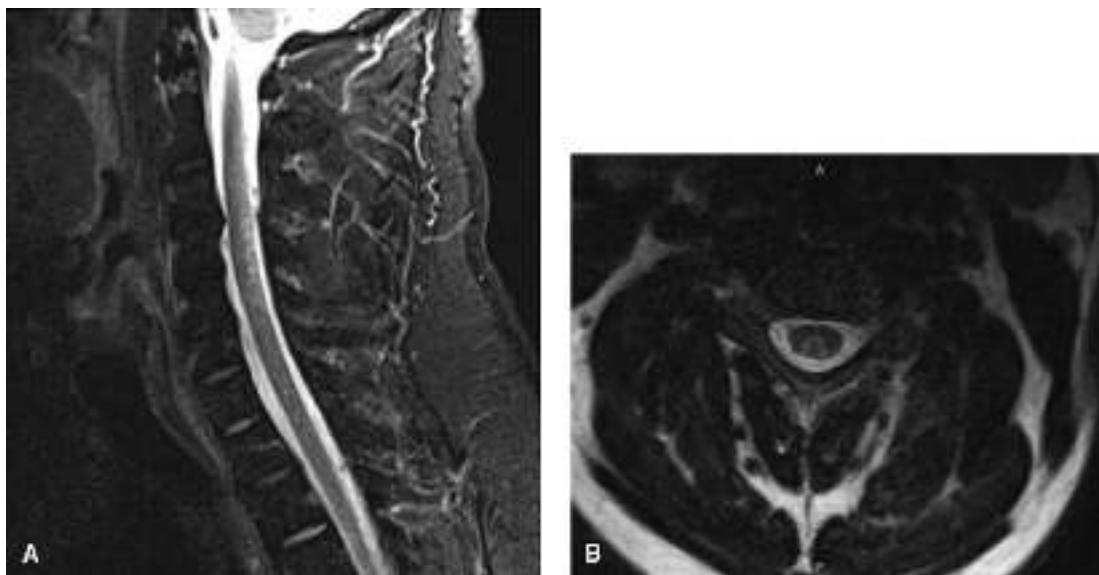


FIGURE 24.7 Nitrous oxide abuse myelopathy. Sagittal short T1 inversion recovery MRI showing hyperintensity in the posterior cervical and thoracic spinal cord (**A**); axial T2 hyperintensity (**B**). (Reprinted from Probasco JC, Felling RJ, Carson JT, et al. Teaching NeuroImages: myelopathy due to B_{12} deficiency in long-term colchicine treatment and nitrous oxide misuse. *Neurology*. 2011;77[9]:e51, with permission.)

Anterior cord syndrome is due to ischemia in the distribution of the anterior spinal artery (anterior spinal artery syndrome). There is dysfunction of the entire spinal cord, except for the posterior columns. Patients are typically paraplegic or quadriplegic with loss of pain and temperature sensation below the level of the lesion but with retained sensation to light touch, position, and vibration. In the posterolateral column syndrome (subacute combined degeneration), most often because of vitamin B_{12} deficiency, there is demyelination and gliosis of the posterior and lateral columns. Clinically, affected patients have weakness, spasticity, and prominent loss of vibratory and position sense with relative preservation of pain and temperature. A similar syndrome may occur in nitrous oxide exposure, HIV myelopathy, copper deficiency, or zinc toxicity (Figure 24.7). Syringomyelia, often associated with Chiari malformations, produces suspended, dissociated sensory loss (see Chapter 36).

In the posterior column syndrome, dysfunction is limited to the posterior columns. The primary cause is syphilitic myelopathy, now rarely seen (tabes dorsalis; tabes, L. consumption). Isolated degeneration of the posterior column, without demonstrable etiology, occurs as a clinically benign rare condition, without progression to other systems. The etiology is most likely a sporadic degenerative disease of the cord. The anterior horn syndrome is characterized by

loss of anterior horn cells and occurs in such conditions as spinal muscular atrophy (hereditary or acquired) and poliomyelitis. The anterior horn-corticospinal tract syndrome causes a combination of spasticity and anterior horn cell dysfunction and occurs in amyotrophic lateral sclerosis.

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CHAPTER 25

The Corticospinal (Pyramidal) Level

In common parlance, the corticospinal level of motor integration is also referred to as the pyramidal level, cortical level, or upper motor neuron level. From a strict anatomical perspective, these terms are not synonymous. Objections can be raised to the terminology because neither corticospinal, pyramidal, nor upper motor neuron precisely and unambiguously describes the voluntary, direct descending motor pathway, but a better term has not come into use. The pyramidal tract is only one of the descending motor systems that converge on the anterior horn cell, so there are other “upper motor neurons.” The corticobulbar tract supplies brainstem structures in the same way the corticospinal tract (CST) innervates the spinal cord but does not pass through the medullary pyramids and is not therefore “pyramidal.” Neurons that are not part of the pyramidal system project from the cortex to the spinal cord. The terms pyramidal and extrapyramidal have become blurred anatomically; some anatomists suggest that they be abandoned. Clinicians, however, continue to find them useful because the clinical manifestations of lesions of the direct (pyramidal) motor pathways differ from those of the indirect (extrapyramidal) system.

ANATOMY AND PHYSIOLOGY

For clinical purposes, the CST is the principal efferent system through which purposive movements are initiated and performed. The CST is by no means the sole cortical mechanism for movement; it acts primarily to integrate highly skilled, fine, discrete movements of the distal extremities. It is responsible for the contraction of agonist muscles as well as the inhibition, or graded relaxation, of antagonist muscles necessary to perform skilled acts. By its integration and

control, individual muscle contractions are coalesced into complex motor acts. The corticospinal level does not function independently. Normally, and in the presence of disease, it is closely integrated with other levels of motor activity, as well as with a constant stream of incoming sensory impulses. The CST, along with other cortical and brainstem pathways, constantly supplies lower centers with impulses that have a generally inhibitory effect. Disease involving the pyramidal pathways results in a release of this inhibiting effect, resulting in hyperactive and autonomous function of the affected spinal cord segmental levels. This results in excessive activity of the lower centers that are normally suppressed by cortical control mechanisms.

Area 4 (area gigantopyramidalis) of the precentral gyrus is the primary motor cortex (M-I); it is the region having the lowest threshold for stimulation to cause contraction of muscles of the opposite side of the body. The cortex of M-I is agranular and heterotypical; its most characteristic feature is the presence of giant pyramidal neurons (Betz cells) in lamina V. The localization of function within the precentral gyrus is depicted by the motor homunculus ([Figure 6.5](#)). The corticospinal system is phylogenetically relatively new. It is fully developed only in mammals and reaches its highest development in apes and man. The phylogenetic acquisition of speech and complex hand function resulted in expansion of cortical areas representing the tongue, mouth, lips, thumb, and fingers, displacing the cortical representation for the lower extremities and sacral regions upward and onto the medial surface of the hemisphere. Areas for the tongue, face, and digits are exceptionally large and out of proportion to those of the proximal musculature. The extension of the precentral gyrus onto the medial aspect of the frontal lobe forms the anterior portion of the paracentral lobule. Neurons controlling the lower extremities and perineal musculature are in the paracentral lobule, which plays an important role in bowel and bladder sphincter control.

There are reciprocal connections between the primary motor cortex and the primary somatosensory cortex in the postcentral gyrus. M-I receives association fibers from the premotor and supplementary motor areas and from the insula. These connections are involved in the preparation and planning for voluntary movements that are then executed by the primary motor cortex. There are also connections between the primary motor cortices in the two hemispheres. The posterior division of the ventral lateral nucleus of the thalamus receives input from the cerebellum and projects to area 4.

The term pyramidal tract arose because these fibers make up most of the

medullary pyramids. It was once thought that the pyramidal tract consisted mainly of the axons of the Betz cells in the primary motor cortex. However, of the approximately 1 million fibers in the CST at the level of the pyramids, only 20% to 30% arise from M-I and only 3% arise from the Betz cells. These fibers are large, heavily myelinated and conduct rapidly. In addition to the contribution from M-I, the CST contains fibers from the premotor cortex (area 6), the supplementary motor area and adjacent regions, and the postcentral gyrus (areas 3, 1, and 2). The majority of the CST arises in approximately equal thirds from area 4, area 6, and the postcentral gyrus, with an additional contribution from the adjoining parietal cortex (area 5) and other portions of the brain, including the temporal and occipital lobes, cingulate gyrus, and certain subcortical centers (Figures 25.1 and 25.2).

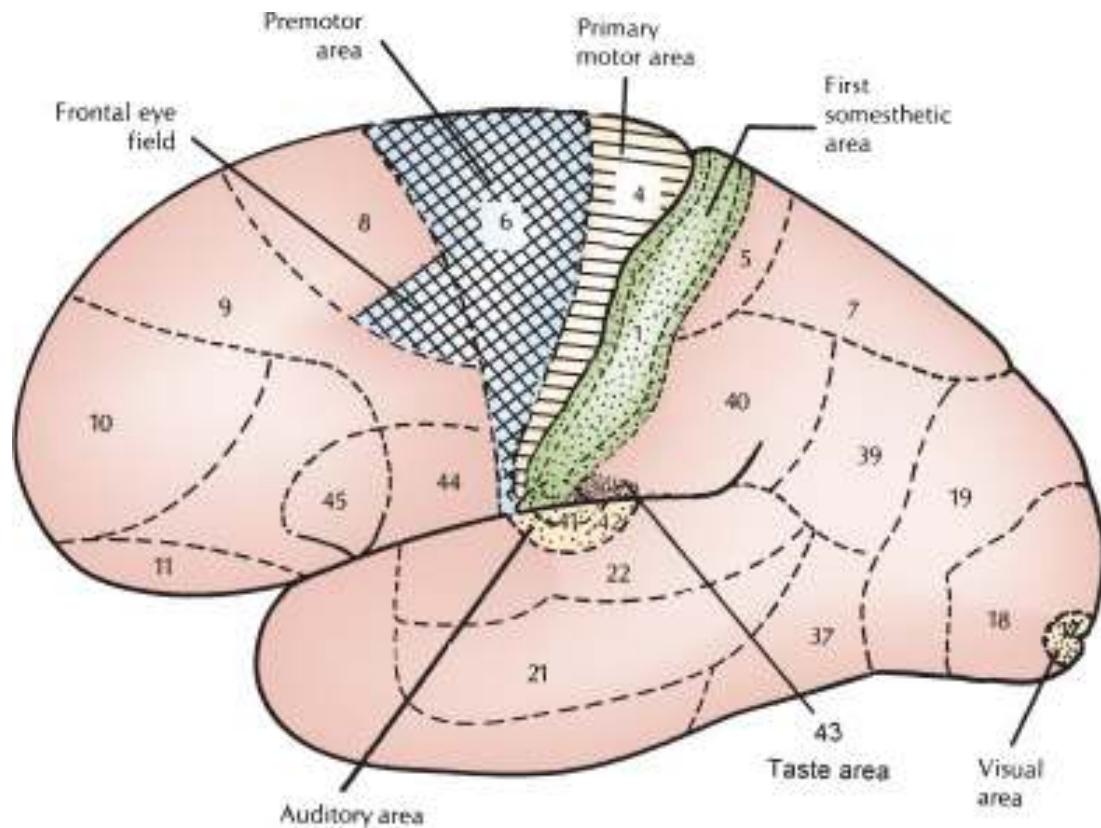


FIGURE 25.1 A lateral view of some of the clinically relevant cortical areas from the Brodmann cytoarchitectonic map. (Modified from Kiernan JA. *Barr's The Human Nervous System: An Anatomical Viewpoint*. 7th ed. Philadelphia: Lippincott-Raven, 1998, with permission.)

The premotor region (area 6), located just rostral to area 4, is closely related to the motor cortex, both anatomically and functionally. The premotor cortex is

similar histologically to the motor cortex but lacks the giant pyramidal cells. Some fibers from area 6 pass to area 4 and then downward with the CST; others descend with the pyramidal fibers. There is probably less complete crossing of the fibers from the premotor cortex than of those from the motor cortex. In addition, the premotor region communicates with the basal ganglia and other portions of the extrapyramidal system, including the subthalamic nucleus, red nucleus, superior colliculus, vestibular nuclei, inferior olive, and brainstem reticular formation ([Figure 25.3](#)).

The CST is important in controlling discrete, isolated motor responses, especially fine voluntary movement of individual digits. The CST provides speed and agility of distal extremity movements. The premotor cortex and its pathways are concerned with larger coordinated responses, with more stereotyped movements that are partly automatic and involve the trunk and the proximal limbs, and with postural mechanisms. It is the principal cortical component of the extrapyramidal system. Stimulation of area 6 causes contraversive movements of the head and trunk. The premotor region is involved in movements guided by visual, auditory, and somatosensory stimuli. The supplementary motor area (M-II) is part of area 6 that lies on the medial aspect of the frontal lobe just anterior to the paracentral lobule (area 6ab). It communicates with the primary motor cortex as well as with the supplementary motor cortex in the opposite hemisphere. M-II seems to be involved particularly in planning and integrating bilateral body movements. M-II is also somatotopically organized, but the homunculus is cruder and less detailed than the one in M-I. The cingulate motor area in the anterior half of the cingulate gyrus projects to the primary motor cortex and also contributes descending fibers to the corticobulbar and CSTs. There is also a secondary motor area in the depths of the central sulcus where the precentral and postcentral gyri merge.

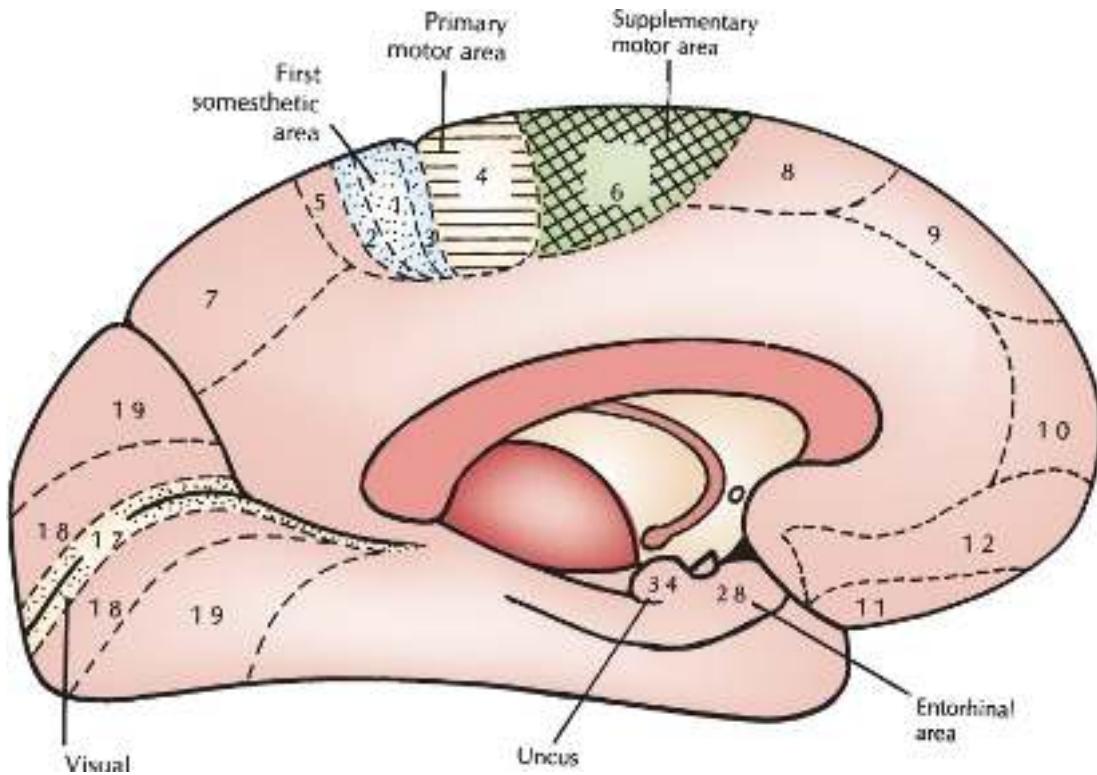


FIGURE 25.2 A medial view of some of the clinically relevant cortical areas from the Brodmann cytoarchitectonic map. (Modified from Kiernan JA. *Barr's The Human Nervous System: An Anatomical Viewpoint*. 7th ed. Philadelphia: Lippincott-Raven, 1998, with permission.)

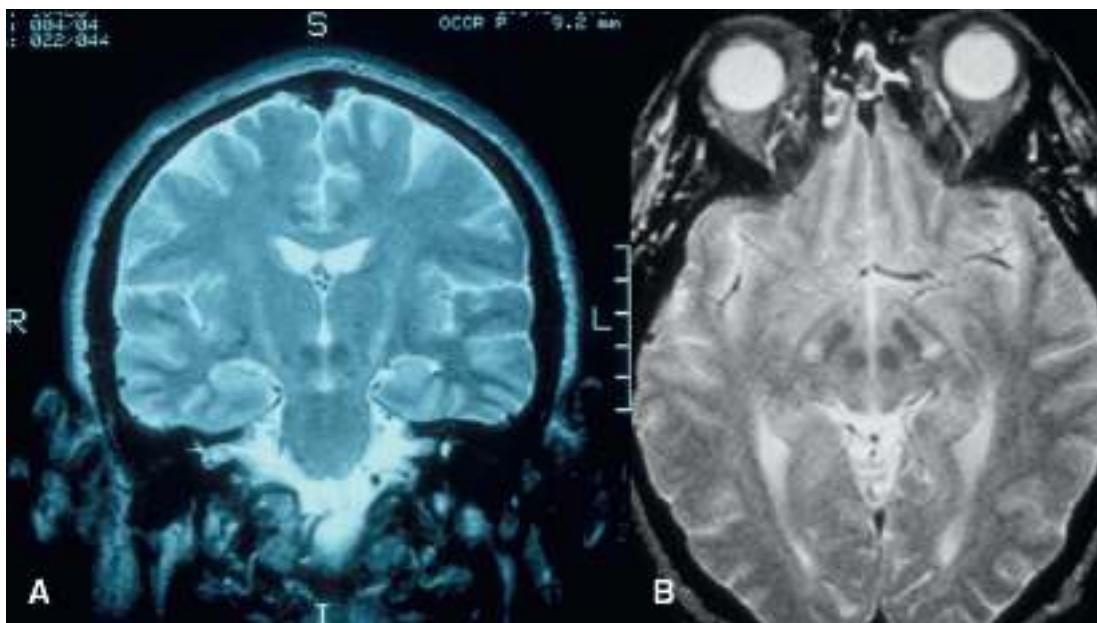


FIGURE 25.3 Degeneration of the corticospinal tracts in primary lateral sclerosis. In (A), there are streaks of high signal on the coronal view. In (B), the transverse view demonstrates ovoid areas of high signal intensity in the posterior portions of the internal capsules.

Axons from the motor neurons of the precentral gyrus descend through the corona radiata and the posterior limb of the internal capsule where corticobulbar fibers are anterior, followed posteriorly by those to the upper extremity, trunk, and lower extremity. About 90% of the CST fibers are small myelinated axons with a diameter of 1 to 4 μm , and most of the remaining fibers are 5 to 10 μm in diameter. The small numbers of fibers that arise from Betz cells are very large, with diameters of 10 to 22 μm . The posterior limb of the internal capsule is that portion of the capsule between the lenticular nucleus and the thalamus. The retrolenticular part lies posterior to the lenticular nucleus. In the rostral part of the internal capsule, the corticospinal fibers lie in the anterior portion of the posterior limb. As the capsules descend, the corticospinal fibers move posteriorly and come to occupy a position in the posterior third to posterior quarter of the posterior limb. In normal individuals, the CST can often be visualized as a subtle hyperintensity in the posterior portion of the posterior limb of the internal capsule on T2-weighted axial magnetic resonance images. These areas of hyperintensity are found near the junction of the posterior limb and the retrolenticular portion of the capsule. The signal change is thought to mark the very large heavily myelinated, rapidly conducting fibers that constitute the Betz cell component of the pyramidal tract. Degeneration of the CST, as occurs in amyotrophic lateral sclerosis or primary lateral sclerosis, accentuates this signal change and may be useful diagnostically ([Figure 25.4](#)).

At the midbrain level, pyramidal fibers traverse the middle three-fifths of the cerebral peduncle, with corticobulbar fibers most medial ([Figure 25.5](#)). The majority of corticobulbar fibers decussate before synapsing with the specific cranial nerve nuclei, but most of the cortical innervation of brainstem centers is both crossed and uncrossed. The descending fibers then pass through the basilar portion of the pons as disjointed fascicles and enter the medulla. In the caudal medulla, the CSTs gather into the discrete twin columns of the medullary pyramids that make up the base of the medulla ([Figure 11.11](#)). Approximately 85% to 90% of the fibers cross in the decussation of the pyramids, with those destined for the upper extremity decussating more rostrally than those for the lower extremity ([Figure 11.12](#)). There is considerable variation in the proportion of crossed and uncrossed CST fibers in man. Instances of ipsilateral hemiplegia because of an uncrossed pyramidal tract have been reported. Terakama et al. used functional magnetic resonance imaging, motor-evoked potentials, and somatosensory-evoked potentials to investigate a patient with an ipsilateral

hemiplegia following a cerebral hemorrhage. They were able to demonstrate that the affected limbs were controlled by the ipsilateral cerebral cortex. In some instances, an uncrossed CST is associated with congenital anomalies of various sorts.

The fibers that decussate descend in the lateral funiculus of the spinal cord in the lateral CST, lying anterolateral to the posterior gray horn, medial to the posterior spinocerebellar tract, and posterior to the plane of the denticulate ligaments, to supply the muscles of the opposite side of the body. In the lumbosacral cord, where there is no posterior spinocerebellar tract, the lateral CST abuts the surface of the cord. The lateral CSTs may also contain other corticofugal fibers as well as some ascending ones. About 50% of the fibers of the lateral CST terminate in the cervical region, 20% in the thoracic area, and 30% in the lumbosacral portion of the cord. The tract ends at about the S4 level. The smaller anterior CST usually contains about 10% to 15% of the corticospinal fibers; it descends uncrossed in the ipsilateral anterior funiculus and usually does not extend below the midthoracic region. These fibers cross in the anterior white commissure at the segmental spinal cord level before terminating; they primarily supply axial muscles.

The axons of the corticobulbar and CSTs terminate on motor nuclei of the cranial nerves and the anterior horn cells of the spinal cord. Those traveling to the cord terminate primarily in laminae IV to VI, lamina VII, lamina VIII, and lamina IX on the side opposite the hemisphere of origin. Fibers from areas 4 and 6 end more ventrally, mainly in laminae VII and VIII, and are concentrated in the cervical and lumbosacral enlargements. The majority of CST fibers synapse on an interneuron, but about 10% end directly on alpha motor neurons in lamina IX. Those fibers that project directly from the cortex to anterior horn cells mediate discrete, skilled, fine motor movements of the distal extremities. Impulses then travel from the brainstem motor nuclei and anterior horn cells to the neuromuscular junctions of striated muscles (final common pathway). A single corticospinal fiber innervates more than one neuron in the spinal cord, and some probably innervate many. The pyramidal tract affects the activity of both alpha and gamma motor neurons. Alpha gamma coactivation serves to maintain a consistent level of stretch on the intrafusal muscle fibers during contraction and relaxation of the extrafusal fibers.

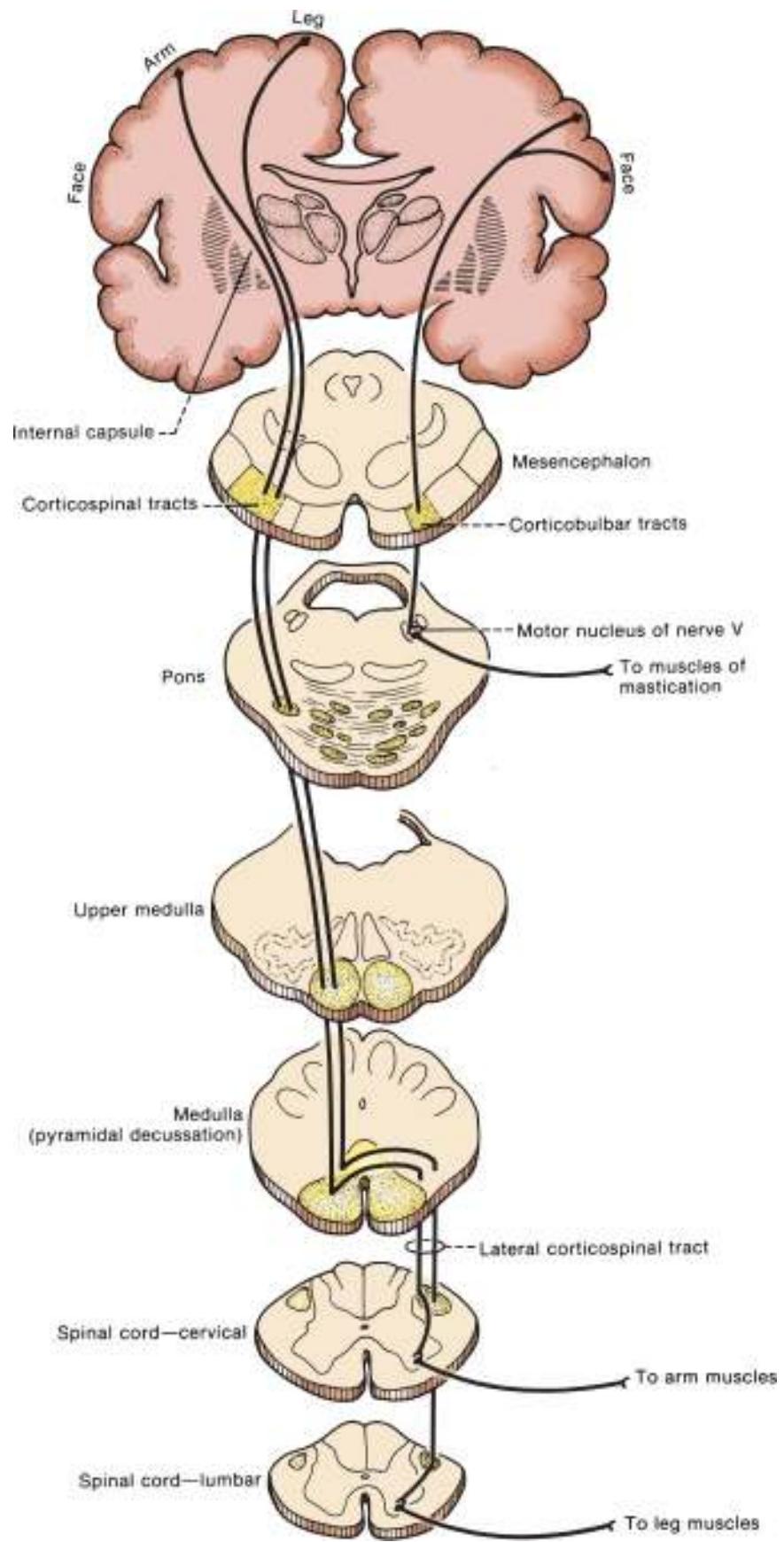


FIGURE 25.4 The corticobulbar and corticospinal pathways.



FIGURE 25.5 This patient suffered a major intracranial injury at age 3 and was left with refractory seizures and a spastic right hemiparesis with hemiatrophy.

The CST preferentially innervates certain muscles, and this “pyramidal distribution” is important clinically. Lateral vestibulospinal tract effects are largely the opposite of CST effects. Glutamate or aspartate may be the excitatory neurotransmitters in some corticospinal neurons. The fibers from the parietal

lobe that descend in the pyramidal tract terminate on sensory neurons in the dorsal horn and play a role in modulating sensory impulses in the long, ascending sensory pathways.

Neuronal discharge of the primary motor cortex, as from electrical stimulation or seizure activity, causes muscular contractions on the opposite side of the body. The response is one of groups of muscles rather than simple contraction of isolated muscles, but individual muscles as well as movements may be finely represented in the motor cortex. Stimulation of area 4 can cause discrete movements of the digits and the muscles supplied by the cranial nerves. With some overlap, areas controlling movements of the thumb, index finger, hallux, and face have the widest distribution and the lowest threshold. Stimulation of area 6 also causes a contralateral motor response, but a stronger stimulus is necessary than when area 4 is stimulated. The resulting movements are more complex and consist of slow, synergistic, postural, or patterned contractions of a generalized type that involve large muscle groups.

The CST lesions induced by ablation experiments in animals cause various deficits, depending on the animal and the area ablated. From a phylogenetic standpoint, voluntary motor control is extrapyramidal in submammalian species, mixed pyramidal/extrapyramidal in nonhuman mammals, and essentially pyramidal in humans. The effects of medullary pyramid transection (pyramidotomy) illustrate the differences in the function of the CST in different species. In the chimpanzee, it causes paralysis of the whole limb. In the rhesus monkey, the same lesion causes paralysis of distal extremity muscles with permanent loss of independent hand and finger movements. In the cat, it causes minimal deficits. Because of differences between the effects of lesions in man and those in experimental animals, such experiments have been of only limited usefulness in understanding human CST pathophysiology.

Questions remain about the relationship of the CST to spasticity. Primarily because of animal experimentation, it has been thought that lesions restricted to the pyramidal tract may cause weakness but not spasticity or hyperreflexia. The increase in tone that follows CST lesions may be related more closely to dysfunction of the extrapyramidal rather than the pyramidal system or to interruption of corticofugal fibers other than pyramidal in the corticospinal pathway. Although experimental pyramidotomies in animals may not lead to spasticity, in humans the evidence is that spasticity does eventually develop in the majority of cases. This is true even after very restricted lesions involving the medullary pyramids, although after a longer interval than is typical of other

pyramidal lesions. It is likely that early flaccidity followed by the development of spasticity occurs with infarction of the CST at any level. More slowly developing lesions are likely to manifest spasticity at the time of initial presentation. Spasticity probably results from imbalance of the facilitatory and inhibitory centers in the midbrain and brainstem reticular formations, as well as altered balance between the alpha and gamma motor systems in the spinal cord. The reduction in the threshold and exaggeration of the stretch reflexes, an essential aspect of spasticity, may be mediated by the rubrospinal and vestibulospinal rather than the CSTs.

Hemiplegia in man is often produced by combined lesions of the motor and premotor components of the upper motor neuron. Various pathologic responses—such as the Babinski, Chaddock, and Hoffmann signs—appear. The affected extremities may at first be flaccid with depressed reflexes, but spasticity and reflex exaggeration typically develop within a few days. The pathologic reflexes may remain permanently.

CLINICAL MANIFESTATIONS OF DISEASE OF THE CORTICOSPINAL LEVEL

The corticospinal pathways may be involved in diverse disease processes, including vascular disease, neoplasm, degeneration, trauma, and others. The essential manifestations of a CST lesion consist of loss of skilled voluntary movements, or impairment of integration of movements, along with an overactivity of lower segmental centers because of disinhibition. The loss of voluntary movement is accompanied by increased tone in the involved muscles. The paresis from a lesion involving the pyramidal tract tends to involve entire extremities or certain muscle groups. Pyramidal lesions disrupt movements; any muscle that participates in the movement will be weakened, regardless of its specific lower motor neuron innervation. In contrast, lower motor neuron lesions involve muscles innervated by a specific structure, such as a nerve root or peripheral nerve. For instance, a CST lesion causes weakness of external rotation of the shoulder. This movement is carried out by both the infraspinatus and teres minor muscles. The teres minor is innervated by the axillary nerve and the infraspinatus by the suprascapular nerve; both are weakened despite their different segmental innervations.

Pyramidal tract lesions do not cause the sort of severe, early focal muscle

atrophy seen in lower motor neuron lesions, but there may be some mild, late atrophy of the involved part because of disuse (see [Chapter 29](#)). With lesions that are congenital or occur early in life, the involved limbs may fail to grow normally, resulting in hemiatrophy to varying degrees in adulthood ([Figure 25.5](#)). Such hemiatrophy may be difficult to detect; comparing the size of the thumbnails is a traditional technique for detecting subtle hemiatrophy. When atrophy does occur with CST disorders, it usually affects the small muscles of the hand. There are no fasciculations.

Deep tendon (muscle stretch) reflexes—rather than being lost as is the usual case with neurogenic atrophy because of a lower motor neuron lesion—are increased, and clonus may be elicited (see [Chapter 38](#)). The superficial reflexes are diminished or absent (see [Chapter 39](#)). Various pathologic reflexes, such as the Babinski sign, often termed pyramidal reflexes or upper motor neuron signs, often appear (see [Chapter 40](#)). Normal associated movements may be lost, and abnormal associated movements may appear (see [Chapter 42](#)). Trophic changes are uncommon, but occasionally, there is edema, desquamation, pigmentary changes, or glossy skin.

A pyramidal tract lesion causes weakness in a characteristic distribution. Facial weakness is limited to the lower face, although occasionally eyelid closure may be slightly weak. Voluntary facial movements are affected more than emotional ones, and movement in response to emotional stimuli may be normal (dissociated facial palsy; [Chapter 16](#)). There is slight, if any, detectable involvement of the muscles innervated by the spinal accessory nerve. There may be slight weakness of the affected side of the tongue, but the throat and jaw muscles function normally. Deglutition, articulation, movements of the trunk, and other functions with bilateral supranuclear innervation are little affected. Voluntary, skilled, and learned actions are most impaired, and there is loss of the ability to carry out fine, independent, fractionated movements, especially with the distal portions of the extremities, with precision and delicacy. Gross movements and those that are habitual or have little voluntary control are relatively spared.

The extremity weakness reflects the preferential CST innervation of certain muscle groups and has a characteristic pattern: the pyramidal or corticospinal distribution. When weakness is mild, it may be detectable only in the corticospinal distribution. Distal muscles, especially hand muscles, receive more pyramidal innervation than proximal muscles and are particularly affected. In the upper extremity, weakness preferentially involves the wrist, finger and elbow

extensors, supinators, and external rotators and abductors of the shoulder; there is relative sparing of the flexor, pronator, and internal rotation muscles. In the lower extremity, weakness is most marked in the foot and toe dorsiflexors, knee flexors, and flexors and internal rotators of the hip, with relative sparing of the extensors, external rotators, and plantar flexors. When weakness is severe, the stronger, non-CST-innervated muscles overcome the weak muscles, producing the characteristic posture of a spastic hemiplegia. The arm is held in adduction, with internal rotation at the shoulder, flexion and pronation at the elbow, and flexion of the wrist and fingers. Additional flexion may still be carried out, but there is marked weakness of extension. There is loss of isolated movements of the wrist and fingers; movements at the elbow and shoulder are less affected. In the lower extremity, there is weakness of flexion at the hip and knee; the hip is extended, adducted, and often externally rotated, and the knee is extended. There is weakness of the dorsiflexors and everters of the foot, often with a pes equinovarus deformity causing plantar flexion and inversion of the foot and toes.

The spasticity, or increase in tone, is most marked in the flexor and pronator muscles of the upper limb and the extensors of the lower, more apparent with an attempt to extend or supinate the muscles of the upper extremity or flex those of the lower. Passive motion may be carried out with little difficulty if done through a small range of movement, but resistance increases if an attempt is made to move the extremities through a greater range. Slow, passive movement may be carried out with relative ease, but on rapid movement, there is a “blocking” or “catching,” often with a waxing followed by a sudden waning of tone at the extremes of the range of motion (clasp-knife phenomenon). Abnormalities of muscle tone are discussed further in [Chapter 28](#).

The paralysis that follows a vascular lesion of the internal capsule provides a common example of the effects of a CST lesion. Examination soon after the event typically reveals flaccid paralysis and areflexia on the opposite side of the body (“cerebral shock”) but is soon followed by spasticity and hyperreflexia. When the corticospinal pathways are affected by a spinal cord lesion of sudden onset, especially if bilateral, there may also be a period of flaccidity and areflexia accompanying the paralysis below the level of the lesion. This is the period of “spinal shock,” which sooner or later gives way, in most instances, to the corticospinal syndrome. During the neural shock phase, the plantar responses may be mute and the superficial reflexes absent. The pyramidal, or upper motor neuron, syndrome gradually emerges over hours to weeks with spasticity, hyperactive tendon reflexes, extensor plantar responses, and continued absence

of superficial reflexes. With spinal cord lesions, there is also impairment of bowel, bladder, and sexual function.

The motor deficit with CST lesions is only occasionally complete. This may be the result of the same, largely unknown, factors and mechanisms responsible for the recovery of function that follows many such lesions. Some muscles may have bilateral innervation, or there may be incomplete decussation in the medulla. The CST receives fibers from regions of the cortex other than the motor strip, and many of the motor centers in the cortex occupy a large area with overlap of the foci of localization. The primary motor cortex is only one portion of the motor system; other cortical and subcortical centers, such as the supplementary and secondary motor cortices, may assume function in the face of disease of the corticospinal system. Sensory factors influence the type and degree of paralysis and the degree of motor recovery; the prognosis for return of function is less optimistic if there is significant sensory loss. In patients with infantile hemiplegia, hemispherectomy done for seizure control may not increase the motor deficit. Removal of normally developed cortex in an adult (e.g., surgical extirpation of a neoplasm) causes a spastic hemiplegia. Yet similar removal in patients who have had a spastic hemiparesis since birth or early childhood causes a transient flaccid deficit that later becomes spastic, and the residual weakness after surgery is no more than before, sometimes less. In such patients, it is likely that either the other hemisphere or some subcortical or cortical structures have previously assumed a portion of the function of the diseased cortex.

With cerebral neoplasms or other pathologic processes affecting the motor cortex, there may be a corticospinal type of paresis, together with recurrent jacksonian convulsions of the involved extremities. A lesion of the pyramidal pathway after it has left the cortex, however, produces only paresis, and convulsions do not occur.

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CHAPTER 26

The Extrapyramidal Level

The extrapyramidal system is more of a functional concept—derived primarily from the study of patients with neurologic disease—than an anatomic or physiologic entity. Patients with disease of the extrapyramidal system have disorders that involve the motor system, but the clinical phenomenology is distinctly different from the weakness, spasticity, and hyperreflexia that mark the pyramidal syndrome. The term extrapyramidal was first used by Wilson in describing hepatolenticular degeneration (Wilson's disease). Wilson's patients had a type of motor disturbance with different clinical characteristics than seen with pyramidal system disease, which was associated with lesions in the basal ganglia (BG). Because the principal component of the extrapyramidal motor system is the BG, the term extrapyramidal came to be used to refer to the BG and their connections. Extrapyramidal disorders cause a category of neurologic illness now more often referred to as movement disorders. Such conditions may produce excessive movement (e.g., Huntington's chorea), a poverty of movement (e.g., Parkinson's disease), or a disturbance of posture, tone, righting reflexes, or other manifestations.

The extrapyramidal system is phylogenetically old. Much of its function arises through modulation of the pyramidal system rather than by direct projections to the spinal cord. The extrapyramidal system can be looked at as a neural network that influences motor control. It is not directly concerned with the production of voluntary movement but is closely integrated with other levels of the motor system to modulate and regulate the motor activity that is carried out by way of the pyramidal system.

ANATOMY AND PHYSIOLOGY

Anatomists disagree as to what should be properly included in the

extrapyramidal system and even whether it should be recognized as an entity. They speak of “nonpyramidal corticofugal systems” but are loath to discuss the “extrapyramidal system.” Many decry the continued use of the expression, and the term does not even appear in many major neuroanatomy textbooks. There are other important nonpyramidal motor systems that are not related to the BG. Other nonpyramidal, nonbasal ganglia pathways include the rubrospinal, vestibulospinal, olivospinal, and reticulospinal tracts. The pyramidal system is the final effector because the extrapyramidal system serves to modulate the activity of the pyramidal system and does not itself project to the spinal cord. Despite these limitations, the concept of an extrapyramidal level continues to be clinically useful. Although the cerebellum is a nonpyramidal motor system, it is not considered part of the extrapyramidal system.

As mentioned in [Chapter 2](#), BG nomenclature is not used consistently. The BG that contribute most extensively to the extrapyramidal system and are most important for clinical purposes include the caudate, putamen, globus pallidus (GP), substantia nigra (SN), and subthalamic nucleus (STN). In addition to the motor circuits controlling voluntary movement, the BG have limbic connections involved with the emotional aspects of movement, as well as connections with the oculomotor system. They also play a role in cognition.

The caudate and putamen are frequently referred to as the striatum, in contrast to the GP, or pallidum. Sometimes, all three are included as parts of the striatum, with the caudate- and putamen-labeled neostriatum and the GP-labeled paleo-(less often archi-) striatum. The terms striatum and corpus striatum are sometimes used synonymously; at other times, a distinction is drawn between the striatum as caudate and putamen (or caudatoputamen) and the corpus striatum as caudate, putamen, and GP. For purposes of this discussion, striatum refers to the neostriatum, the caudate and putamen. Some authorities include other gray matter masses that lie at the base of the brain as part of the BG, including the substantia innominata, nucleus accumbens, claustrum, amygdala, anterior perforated substance, and olfactory tubercle. Most of these are not functionally related to the BG motor system and are usually not considered parts of the extrapyramidal system for clinical purposes. There is a dorsal striatum, or dorsal division of the striatum, and a smaller ventral striatum, or ventral division. The dorsal striatum is the caudate and putamen; the ventral striatum consists of the nucleus accumbens and the anterior portion of the anterior perforated substance and olfactory tubercle. The connections of the ventral striatum are predominantly with the limbic system. Similarly, there is a dorsal pallidum (the

GP proper) and a ventral pallidum (the posterior portion of the anterior perforated substance). For clinical purposes, neither the ventral striatum nor the ventral pallidum plays a significant role in voluntary motor function.

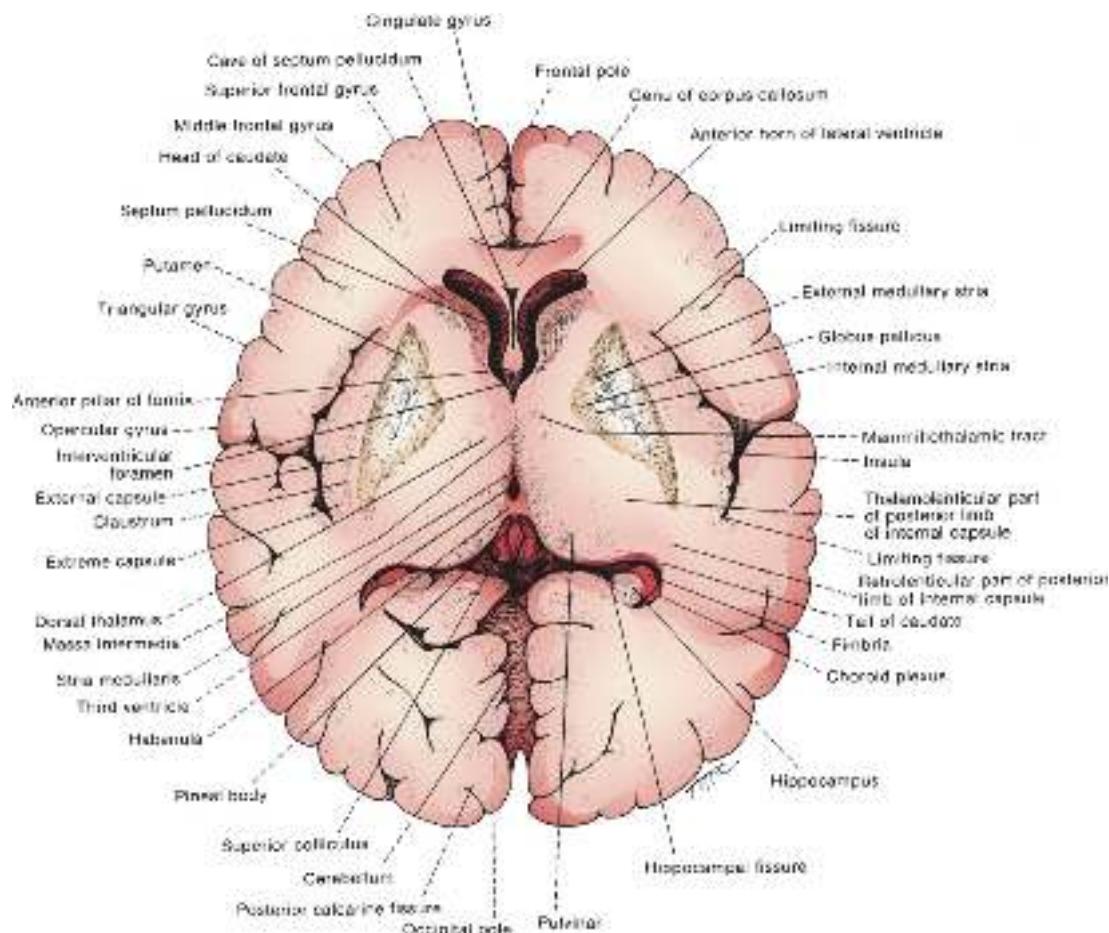


FIGURE 26.1 Drawing of the superior surface of an unstained horizontal section of the adult human brain, through the internal capsule, basal ganglia (BG), and thalamus.

The caudate nucleus lies deep in the substance of the cerebral hemisphere between the lateral ventricle and the insula (Figure 26.1). The head of the caudate nucleus is a pear-shaped mass of gray matter that bulges into the lateral aspect of the frontal horn of the lateral ventricle; its tail runs backward in the floor of the ventricle and then downward and forward in the roof of the temporal horn. The tail of the caudate abuts but remains separate from the amygdala. The sulcus terminalis is a groove that separates the caudate nucleus from the thalamus; it contains the stria terminalis and the thalamostriate vein. The lenticular, or lentiform, nucleus is composed of the putamen and GP, with the

putamen forming the most lateral part of the lentiform complex and the GP the medial part. The claustrum is an island of gray matter in the subcortical white matter of the insula. The putamen is separated from the insula by, in order, the external capsule, claustrum, and extreme capsule.

The internal capsule thrusts itself squarely through the region of the BG, vastly complicating its anatomy and connections. Important components of the BG and extrapyramidal system lie separated on opposite sides of the capsule. The putamen and GP are lateral to it; the caudate, thalamus, STN, and SN lie medial. All efferent and many afferent connections of the BG are made with the diencephalon and midbrain, which are separated from the BG by the internal capsule and crus cerebri. The caudate and putamen are cytologically and functionally nearly identical, but the anterior limb of the internal capsule separates one from the other. Fibers of the internal capsule and crus cerebri also separate the SN from the GP, which are functionally closely related.

The caudate nucleus and putamen are semicontinuous, separated by the streaming fibers of the anterior limb of the internal capsule ([Figure 26.1](#)). The alternating gray and white strands led to the name striatum (striped). Bundles of finely myelinated, small-diameter fibers (“Wilson’s pencils”) crossing the striatum toward the GP also contribute to the marbled appearance. Inferiorly, just above the anterior perforated substance, the head of the caudate fuses with the inferior part of the putamen and is continuous medially with the nucleus accumbens. Histologically, the caudate and putamen are identical; they have a common embryologic origin. They contain a few large and many small neurons, with the small cells predominating by 20:1. Dendrites may be spiny or aspiny. The most common cell type in the striatum is small and spiny and contains gamma-amino butyric acid (GABA), plus enkephalin (ENK), dynorphin, or substance P (SP). The small, spiny neurons are the primary source of striatal efferents. The small, aspiny neurons are cholinergic.

The microstructure of the striatum consists of a matrix of cells that stain histochemically for acetylcholine (ACh), with patches or islands of cells that contain other neurotransmitters. The islands are referred to as striosomes, and the striatum is a mosaic arrangement of islands or patches of striosomes lying in the matrix of cholinergic cells. The striosomes primarily contain SP and ENK. The ENKergic neurons have D2 dopamine receptors; SP neurons have D1 receptors. In the caudate, striosomes have a higher concentration of dopamine compared to the matrix. The striosome-matrix pattern is not as evident in the putamen, which consists mostly of matrix, or in the ventral striatum, which

consists mostly of striosomes. The cholinergic neurons of the matrix are facilitatory to the projection neurons and are inhibited by dopamine.

The GP is medial to the putamen, just lateral to the third ventricle. An external medullary lamina separates the GP from the putamen. Internally, the GP is divided by an internal medullary lamina into a lateral, or external, zone (the globus pallidus externa, GPe); and a medial, or internal, zone (the globus pallidus interna, GPi). The GP contains only about 5% as many cells as the striatum, and all are large neurons. Neurons throughout the GP use primarily GABA as a neurotransmitter, less often ACh. The associated neuropeptide is SP in the GPi and ENK in the GPe.

The SN is a gray mass that lies in the cerebral peduncle between the crus cerebri and the tegmentum of the midbrain at the level of the superior colliculi. The SN is composed of two parts: the deep pars or zona compacta (SNc), which contains the large, melanin-containing, dopaminergic neurons that give the structure its name; and the more superficial zona reticulata (SNr), which contains large, multipolar, nonpigmented, GABAergic neurons similar to those in the GP. The SNr is closely related functionally to—essentially a midbrain extension of—the GPi; they are separated only by fibers of the internal capsule and crus cerebri, and both are involved in BG efferent functions. The SNr receives afferents through the striatonigral comb system or comb bundle, which crosses through the crus cerebri obliquely. The STN (corpus Luysii) is a small, lens-shaped gray mass situated in the ventral thalamic region just dorsal and medial to the cerebral peduncle ([Figure 26.2](#)).

Other important structures involved in the extrapyramidal motor control system include the thalamus, red nucleus (RN), the brainstem reticular formation (RF), the inferior olfactory nucleus in the medulla, the zona incerta (ZI), the vestibular nuclei, the pedunculopontine nucleus (PPN), and the gray matter of the quadrigeminal plate. Several thalamic nuclei are involved, and these are sometimes referred to as the motor thalamus, including the nucleus ventralis lateralis (VL), pars oralis (VLo); ventralis lateralis, pars caudalis (VLC); ventralis posterior lateralis, pars oralis (VPLo); and portions of ventralis anterior (VA). The RN is located in the tegmentum of the midbrain at the level of the superior colliculi. It has magnocellular (large-celled) and parvocellular (small-celled) portions. The caudal magnocellular part gives rise to the rubrospinal tract and the parvocellular part to the central tegmental tract. The lateral and medial nuclear groups of the RF are situated in the tegmentum of the midbrain, and other constituents of the RF that either inhibit or facilitate motor responses are

placed caudally in the brainstem. The PPN is a cholinergic nucleus that lies caudal to the SN in the brainstem tegmentum, partially buried in the superior cerebellar peduncle. It receives afferents primarily from GPi/SNr and sends cholinergic projections to the dopaminergic neurons in the SNC. It may be involved in locomotion. Patients with Parkinson's disease have significant loss of PPN neurons, and dysfunction of the PPN may be important in the pathophysiology of the locomotor and postural disturbances of parkinsonism.

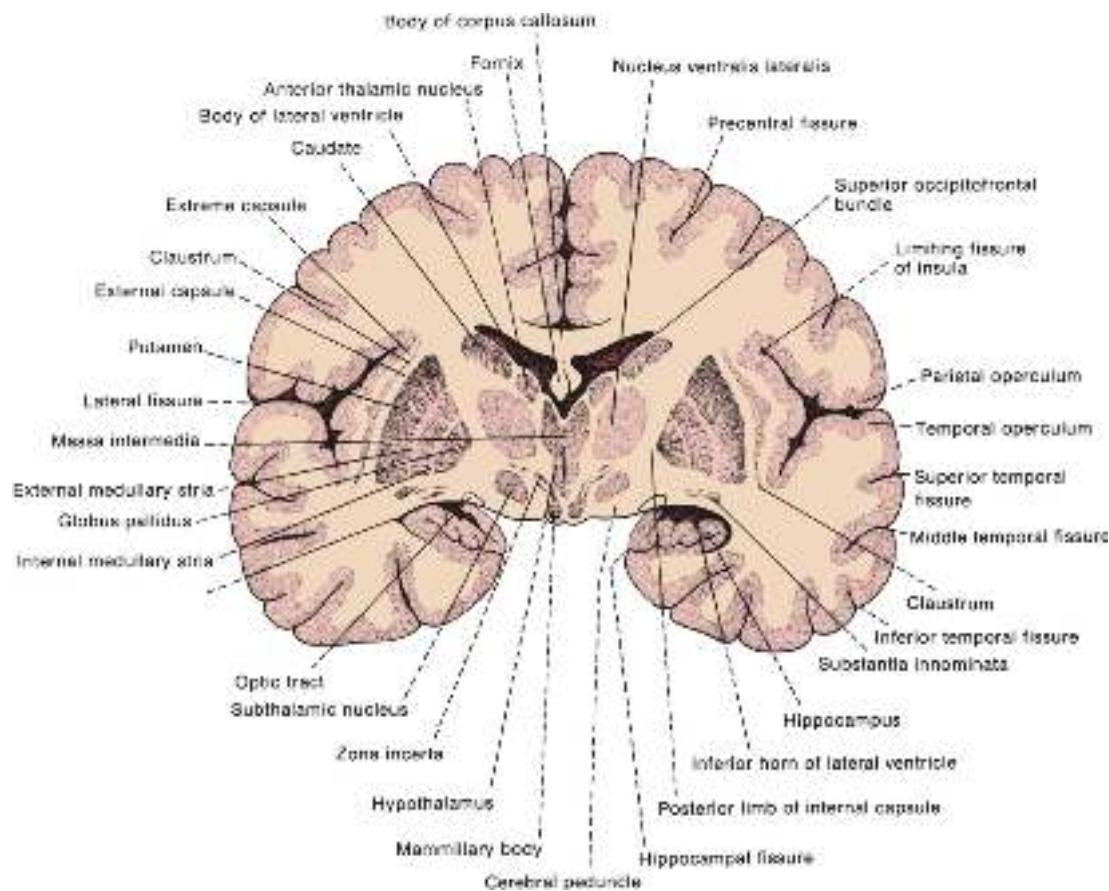


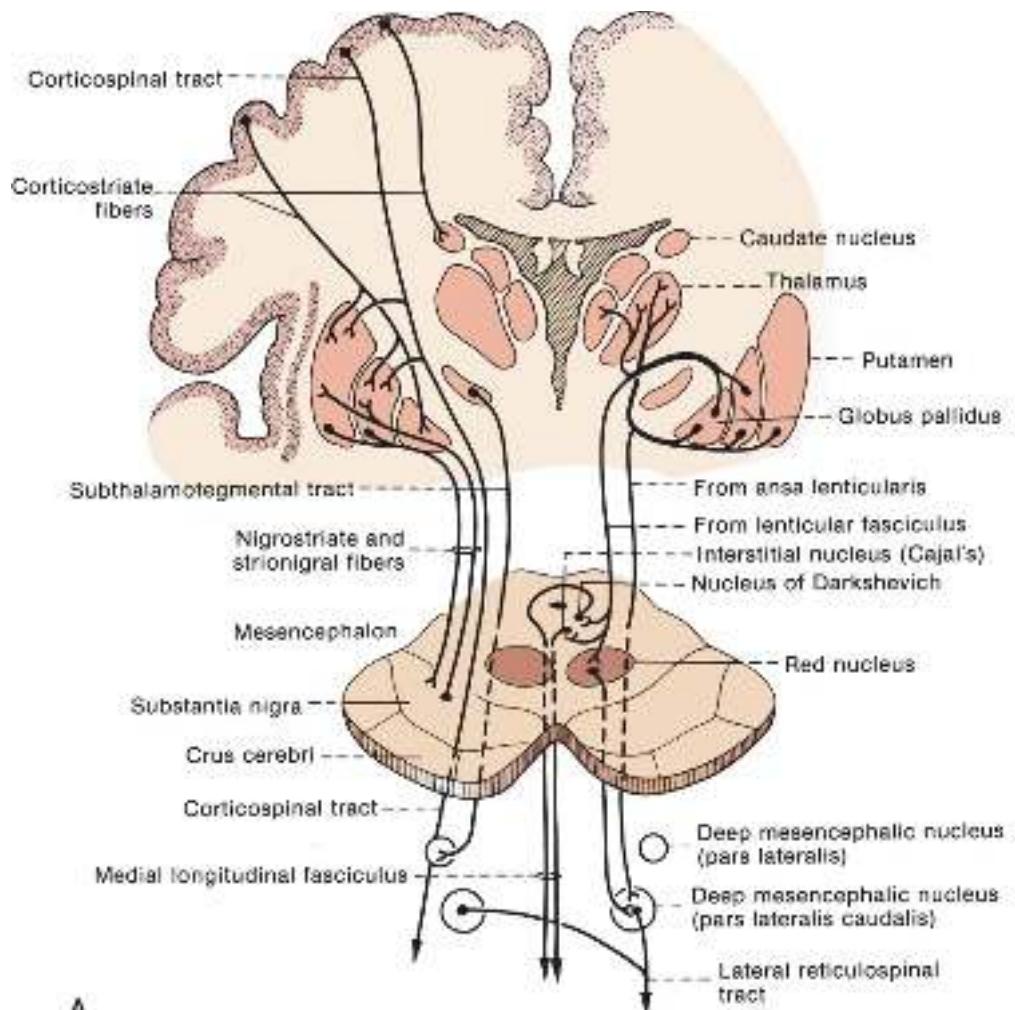
FIGURE 26.2 Drawing of the posterior surface of an unstained coronal section of the adult human brain, through the posterior limb of the internal capsule, BG, and mammillary bodies.

The BG have rich connections with one another and with brainstem structures, as well as with the cerebral cortex and with lower centers (Figure 26.3). The area of the cortex involved is the motor cortex, which includes the precentral motor regions including the supplementary and premotor areas. In essence, the cerebral cortex projects to the striatum, which in turn projects to the GP and SNr; efferents go to the thalamus, which then projects back to cerebral

cortex, primarily to the motor areas. [Table 26.1](#) summarizes some of the major connections.

Striatal Afferents

The striatum receives topographically organized glutaminergic axons that originate from small pyramidal cells in layers V and VI of the entire ipsilateral neocortex. The striatum also receives afferents from the thalamus. These connections provide the striatum with sensory and cognitive inputs. The head of the caudate receives projections from the frontal lobe, the body from the parietal and occipital lobes, and the tail from the temporal lobe. The massive input from the frontal lobe to the head is the reason it is so much larger than the remainder of the nucleus. These connections form the anatomical substrate for the role of the caudate in cognition. The caudate also receives fibers from the dorsomedial (DM) and VA nuclei of the thalamus (thalamostriate fibers) and from the putamen.



A

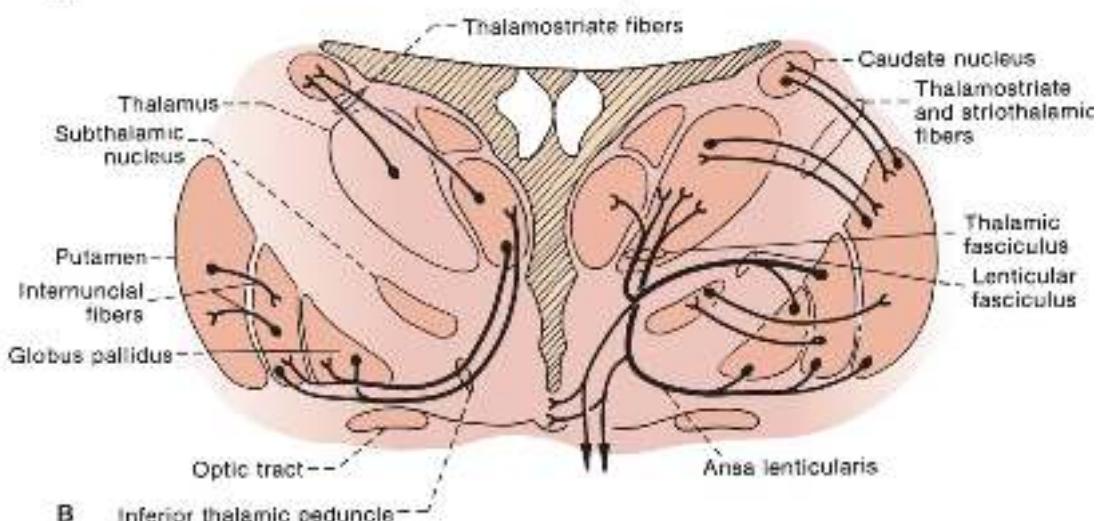


FIGURE 26.3 A. Principal connections of the BG. The thalamocortical portions of the loops (cortex-BG-thalamus-cortex) are omitted. B. Detail giving connections between the BG and thalamus.

TABLE 26.1**Major Basal Ganglia Pathways**

Tract	Origin	Termination	Neurotransmitter
Corticostriate	Cerebral cortex	Striatum*	Glutamate
Striatopallidal	Striatum	GP	GABA
Pallidothalamic [†]	GP	Thalamus	GABA
Pallidosubthalamic [‡]	GPe	Subthalamic nucleus	GABA
Striatonigral	Striatum	SN	GABA
Nigrostriatal	SNC	Striatum	Dopamine
Nigrotectal	SNr	Superior colliculus	GABA
Nigrothalamic	SNr	Thalamus	GABA

*The dorsal striatum, the caudate, and putamen.

[†]Via the fasciculus and ansa lenticularis.

[‡]Via the subthalamic fasciculus.

GABA, gamma-amino butyric acid; GP, globus pallidus; SN, substantia nigra; SNC, pars compacta; SNr, pars reticulata.

The connections of the putamen are more focused; it receives fibers from cortical areas 4 and 6 and from the parietal lobe, the perirolandic motor centers, which are essentially the same areas that give rise to the corticospinal tract. It has a large connection with the caudate. It also receives fibers from the SN by the comb bundle.

Striatal Efferents

The caudate sends fibers to the thalamus (striothalamic fibers) and to the putamen and GP. The primary efferent fibers from the striatum project to the GP and SN. The striatopallidal fibers from the caudate run directly through the anterior limb of internal capsule, and those from the putamen project medially

through the external medullary lamina into the GP.

Pallidal Afferents

The principal afferents to the GP are from the caudate and putamen. The STN also sends fibers to the GP in the subthalamic fasciculus; some pass through the internal capsule into the medial segment of the GP; some cross in the supraoptic commissure (of Gudden). There are also fibers from the SNC to the GP. The GP also receives impulses from DM and VA, through thalamostriate fibers in the inferior thalamic peduncle, and from area 6 and possibly area 4 through corticospinal collaterals.

Pallidal Efferents

The pallidal efferents are the principal outflow of the BG. There are four major bundles: (a) the fasciculus lenticularis; (b) the ansa lenticularis; (c) pallidotegmental fibers, which arise from GPi; and (d) pallidosubthalamic fibers, which arise from GPe. Pallidofugal fibers are often discussed in terms of their relationships to the “fields of Forel.” August Forel performed some of the early anatomical studies of the BG and subthalamic region using degeneration techniques. (Although his name is inextricably linked to the neuroanatomy of the subthalamic region, his principal reputation arises from seminal studies of the social behavior of ants.)

Forel identified fields or regions in the subthalamic and BG area and labeled them “H” for “Haubenregionen” because of their resemblance to a hat plume. There are Forel fields H, H₁, and H₂; these refer to different fiber bundles that course in the region of the RN, STN, and ZI. The prerubral or tegmental field (Forel field H) lies just rostral to the RN and primarily contains dentatothalamic fibers from the contralateral superior cerebellar peduncle and rubrothalamic fibers from the ipsilateral RN ascending toward the thalamus through the subthalamic region. The dentatothalamic and rubrothalamic fibers form a capsule around the RN; the portion of this capsule just rostral to the RN is the prerubral field. Pallidofugal fibers stream into the prerubral field and then turn upward. As these fibers ascend toward the thalamus, the prerubral field divides into dorsal and ventral lamina; the dorsal division consists of the lenticular fasciculus (Forel field H₂), and the ventral division is the thalamic fasciculus

(Forel field H₁).

Both the ansa and fasciculus lenticularis have the same origin, GPi, and the same destination, the thalamus; the difference is that the fasciculus penetrates through, and the ansa curves around, the internal capsule. Both pass through field H, and both join with the thalamic fasciculus. The fasciculus lenticularis emerges from the dorsal surface of GPi and then pierces the internal capsule to lie just above the STN and below the ZI. The ansa lenticularis emerges from the ventral surface of GPi, runs ventromedially to loop around the posterior limb of the internal capsule, and then enters the prerubral field. After traversing the internal capsule, the fasciculus lenticularis joins the ansa lenticularis at the medial border of the ZI; both then enter the thalamic fasciculus just above the ZI.

The thalamic fasciculus (Forel field H₁) is a complex bundle that lies just dorsal to the ZI. It carries pallidofugal fibers as well as rubrothalamic and dentatothalamic fibers. The thalamic fasciculus enters the rostral ventral tier of thalamic nuclei, primarily VL and VA. Some pallidofugal fibers separate from the thalamic fasciculus and enter the centromedian thalamic nucleus. The dentatothalamic fibers in the thalamic fasciculus are destined primarily for VL, but some enter the intralaminar nuclei. The VL is involved in integration and coordination of BG and cerebellar function. The VL in turn projects to area 4 of the motor cortex. The point where the thalamic fasciculus converges on VL is of singular strategic importance in the function of the motor system.

Subthalamic Nucleus

The STN is reciprocally connected with the GP via the subthalamic fasciculus, a bundle that runs directly through the internal capsule. The connection to the STN is the only pallidal efferent to arise from GPe; all others come from GPi. The STN sends fibers back to GPe, as well as to GPi, through the subthalamic fasciculus.

Substantia Nigra

The SN extends from the pons to the subthalamic region and makes up the primary dopaminergic cell population of the midbrain; cholinergic cells are also present. The SNC of one side is continuous across the midline with the SNC of

the opposite side. Cells of the SNc contain neuromelanin, a by-product of dopamine synthesis. The striatonigral afferents from the striatum use GABA and either SP or ENK as transmitters. The SNr receives strionigral fibers from the striatum, GP, and STN. The primary efferents from the SN are to the striatum, midbrain tectum, and thalamus (Table 26.1). The SNr is functionally related to the GPi; its efferents are GABAergic. Dopaminergic nigrostriatal fibers from SNc project to the striatum. The nigrothalamic tract runs to VA and DM. The nigrotectal tract connects the SN with the ipsilateral superior colliculus and may be involved in the control of eye movements. There are also connections between the SN and the PPN and RF.

BASAL GANGLIA PHYSIOLOGY

The connections of the motor system are complex (Figure 22.2). There are two major loops: the BG and the cerebellar. The essential connections in the BG loop are cortex → striatum → globus pallidus → thalamus → cortex. The projections of the thalamus, cortex, and STN are excitatory; the outputs of the striatum and pallidum are primarily inhibitory. The projections from the cortex to the striatum and from the thalamus to the cortex are both excitatory (glutaminergic). The pathway from the striatum to the thalamus may be either excitatory or inhibitory depending on the route. Current models of BG function include a direct and an indirect loop or pathway for the connection between the striatum and thalamus (Box 26.1). In brief, the direct loop is excitatory and the indirect loop is inhibitory. The indirect loop brings in GPe and STN, which are not involved in the direct pathway (Figure 26.4). The caudate, putamen, and STN make up the BG input nuclei; the GPi and SNr are the output nuclei. The input and output nuclei are connected by the direct and indirect loops. In essence, the output nuclei, GPi and SNr, tonically inhibit the motor thalamus; the input nuclei either facilitate cortical motor activity by disinhibiting the thalamus or inhibit motor activity by increasing thalamic inhibition. The direct pathway serves to facilitate cortical excitation and carry out voluntary movement. The indirect pathway serves to inhibit cortical excitation and prevent unwanted movement. Disease of the direct pathway produces hypokinesia, for example, parkinsonism; disease of the indirect pathway produces hyperkinesias, for example, chorea or hemiballismus.

BOX 26.1

Direct and Indirect Pathways

As an overview, the direct pathway is mediated by D1 dopamine receptors and results in a facilitation of movement; the indirect pathway is mediated by D2 receptors and results in an inhibition of movement. In the direct pathway, the motor cortex sends glutaminergic-activating signals to the caudate and putamen and excites D1 receptors. Striopallidal fibers to the globus pallidus interna (GPi) and superficial zona reticulata (SNr) are inhibitory (GABAergic); fibers from the GPi to the thalamus are also inhibitory (GABAergic). At rest, GPi/SNr exert an inhibitory influence on the thalamus, which decreases the excitatory influence of the thalamus on the cortex. Activation of the direct pathway inhibits the activity of GPi/SNr. The direct loop projections from the striatum to GPi inhibit the inhibitory pallidothalamic pathway and result in net cortical excitation and facilitation. The direct loop excites the thalamic projections to the cortex by inhibiting the pallidothalamic pathway.

In the indirect pathway, the motor cortex sends glutaminergic-activating signals to the caudate and putamen and excites D2 receptors. Striopallidal fibers then project to the globus pallidus externa (GPe), causing inhibition (GABAergic). The resting activity of GPe decreases. The GPe projects inhibitory GABAergic fibers to the subthalamic nucleus (STN) by the subthalamic fasciculus. The STN then projects to the GPi, but its fibers are excitatory (glutaminergic). Indirect pathway activity reduces the inhibition of the STN. The STN then facilitates the inhibitory projection of the GPi to the thalamus, resulting in a net decrease in activity in the thalamocortical pathways. The indirect loop inhibits the thalamic projections to the cortex by increasing the inhibition mediated by the pallidothalamic projection. Activity in the indirect loop prevents activation of motor cortical areas that might interfere with the voluntary movement executed by the direct pathway.

Activity in the direct loop results in a net increase in cortical excitation; activity in the indirect loop results in a net decrease. When cortical excitation is at a normal level, voluntary movements are normal. When there is a pathologic net decrease in cortical activation because of disease involving the direct pathway, voluntary movements are inhibited; this causes hypokinetic movement disorders, such as Parkinson's disease. When there is

a pathologic net increase in cortical activation because of disease involving the indirect pathway, movements are augmented; this causes hyperkinetic movement disorders, such as chorea. The final common result in Parkinson's disease is an increase in the firing rate of the GPi, resulting in a suppression of activity in the thalamocortical circuits (the GPi rate theory).

The direct and indirect pathway model and GPi rate theory fail to explain some features of movement disorders, for example, the hyperkinetic component, tremor, in a disease primarily producing hypokinesia. It fails to explain some of the effects of deep brain stimulation in Parkinson's disease or the efficacy of anticholinergic agents. Various authors have proposed refinements, modifications, and alternatives, including the bursting neurons theory, synchronization theory, prokinetic/antikinetic oscillators theory, and the systems oscillators theory. Changes in GPi firing patterns rather than simply the overall firing rate as emphasized in traditional wiring diagrams of the basal ganglia may explain many disease features not well accounted for by the GPi rate theory.

The SNC projects dopaminergic fibers to the striatum, causing excitation or inhibition depending on the receptor. There are five subtypes of dopamine receptor, D1 through D5. The D1 and D2 receptors are the primary ones involved in regulating movement. The dopamine effect on the D1 family of receptors is excitatory; the effect on D2 receptors is inhibitory. The direct loop is routed through the D1 receptors and the indirect loop through the D2 receptors. Dopamine excitation of D1 receptors increases the inhibitory effect of the striatum on the GPi/SNr through the direct pathway, which results in a decrease of the inhibitory effect of GPi/SNr on the thalamus and a net increase in thalamocortical excitation. The net effect of dopamine on the D1 receptor is to facilitate the direct loop and increase thalamocortical excitation. Dopamine inhibition of D2 receptors decreases the inhibitory effect of the striatum on the GPe through the indirect pathway, which results in a decrease of the inhibitory effect of GPe on STN. The disinhibition of STN causes an increase in its ability to excite GPi/SNr, increasing the inhibitory output of GPi/SNr and causing a net decrease in thalamocortical excitation. The net result is that the nigrostriatal system facilitates activity in the direct loop, which increases thalamocortical excitation and inhibits activity in the inhibitory indirect loop, which also increases thalamocortical excitation. When there is dopamine deficiency, cortical

activation is decreased both because of decreased facilitation through the excitatory direct loop and lack of inhibition of the inhibitory indirect loop.

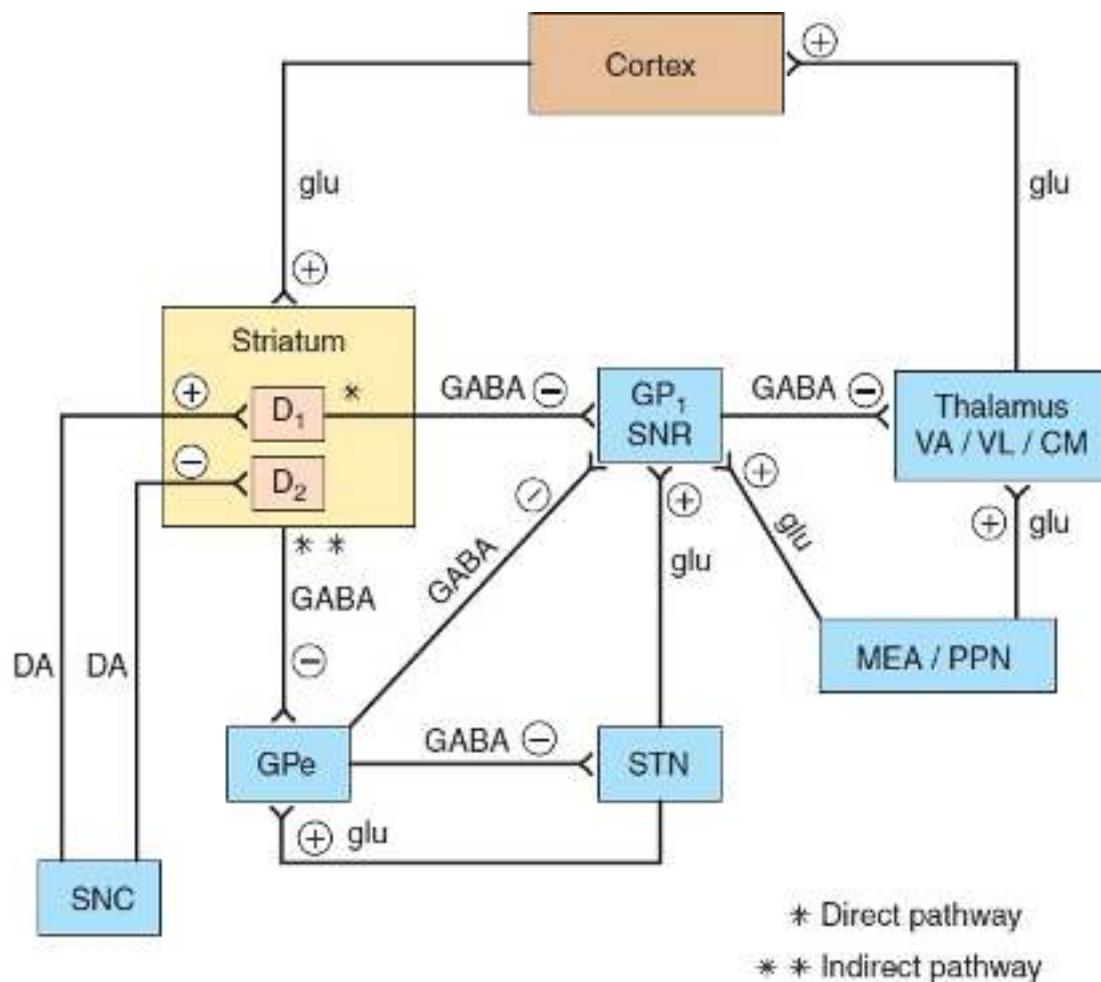


FIGURE 26.4 The direct and indirect BG loops. The indirect loop includes the GPe and subthalamic nucleus. The direct loop results in excitation of the thalamus and cortex, the indirect loop in inhibition. +, excitatory; -, inhibitory; CM, centromedian thalamic nucleus; D₁, D₁ dopamine receptor; D₂, D₂ dopamine receptor; DA, dopamine; GABA, gamma-amino butyric acid; glu, glutamic acid; GPe, globus pallidus externa; GPi, globus pallidus interna; MEA, midbrain extrapyramidal area; PPN, pedunculopontine nucleus; SNC, substantia nigra, compacta; SNr, substantia nigra, reticulata; STN, subthalamic nucleus; VA, ventral anterior thalamic nucleus; VL, ventral-lateral thalamic nucleus.

The inhibitory effect of the BG output neurons affects not only the motor thalamus but the midbrain extrapyramidal areas as well. The effects have been likened to a brake. Increased braking through increased activity from GPi/SNr inhibits motor pattern generators in the cerebral cortex and brainstem; decreased GPi/SNr activity decreases the braking and results in a net facilitation of cortical

and brainstem motor activity. The STN increases the braking, whereas the striatum decreases it. The striatal input to GPi/SNr is organized to provide a specific, focused inhibition (unbraking) in order to selectively facilitate desired movements, whereas the input from the STN causes a more global excitation of GPi/SNr (braking), perhaps to inhibit potentially competing movements.

BASAL GANGLIA PATHOPHYSIOLOGY

Hypokinetic movement disorders, such as parkinsonism, are thought to result from an increase of the normal inhibitory effects of the BG output neurons. Hyperkinetic movement disorders—such as chorea, hemiballismus, and dystonia—presumably result from a reduction in the normal inhibition.

The most common hypokinetic movement disorder is Parkinson's disease. Pathologically, there is loss of the pigmented cells in the SNC, as well as loss of other pigmented cells in the central nervous system, such as the locus coeruleus. The SNC cells are the origin of the nigrostriatal dopaminergic pathway. Loss of dopamine input to the striatum decreases thalamocortical activation by effects mediated by both D1 and D2 receptors. There is decreased activity in the direct loop, mediated by the D1 receptor, causing loss of striatal inhibition of GPi/SNr and increased inhibition of the motor thalamus, resulting in decreased cortical activation. There is also decreased inhibition of the indirect loop, mediated by the D2 receptor. The STN is released from the inhibitory control of GPe, which causes increased activity of the STN; this in turn increases the inhibitory effects of GPi/SNr. Both of these effects decrease the thalamic drive to the motor cortex, causing hypokinesia and bradykinesia. There is a net increase in activity through the indirect over the direct pathway, resulting in a net hyperactivity of GPi/SNr and subsequent inhibition or braking of the thalamocortical circuits.

In hyperkinetic movement disorders, the inhibition of the motor thalamus by the GPi/SNr is impaired. Hemiballismus results from a lesion of the contralateral STN, usually infarction. The damage to the STN removes its normal facilitation of the inhibitory effects of GPi/SNr. The loss of facilitation of GPi/SNr output (less braking) disinhibits the motor thalamus and the cortex, resulting in hyperkinetic movements of the involved extremities. In Huntington's disease, there is loss of ENKergic spiny neurons in the striatum, which project primarily to GPe. Loss of these neurons removes inhibition from GPe, the effect of which is to profoundly inhibit STN, incapacitating it. As with hemiballismus, without

STN input, GPi/SNr inhibition of the motor thalamus decreases, releasing the brake, disinhibiting VL, and causing increased thalamocortical activity and hyperkinesis. Experimentally, chorea can be produced by lesioning STN, disinhibition of GPe, or the administration of dopaminergic agents.

OTHER BASAL GANGLIA FUNCTIONS

In addition to its functions in the regulation of voluntary movement, the BG also have connections involved in cognition, emotion, and oculomotor control. The BG have links to parts of the brain involved in behavior, memory, attention, and reward processes. In the motor loop, cortical projections are to the putamen; in the other loops, cortical projections are to the caudate. In the cognitive loop, projections from the frontal lobe to the caudate travel via the GP to the VA and DM thalamic nuclei, which then send fibers back to the frontal lobe. The cognitive connections of the BG are important in learning new motor tasks. The limbic loop originates in the orbitofrontal and anterior cingulate cortex and travels via the ventral striatum, particularly the nucleus accumbens, to the ventral pallidum, and then to DM, which projects back to the cortex. The limbic loop may be involved in the motor expressions of emotion. The oculomotor loop originates in the cortical eye movement control areas, including the frontal eye fields and posterior parietal cortex, and projects to the caudate and then to both the SNr and the superior colliculus. Impulses from SNr are routed via VA and DM back to the cortex. The oculomotor loop is involved in the control of saccadic eye movements.

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CHAPTER 27

Motor Strength and Power

Motor strength and power indicate the capacity of muscles to exert force and expend energy. Decreased strength is weakness, or paresis; absence of muscle contraction is paralysis, or plegia. Weakness may cause loss of the speed, rapidity, or agility of movement and a decrease in the range, or amplitude, of movement before there is loss of power to formal strength testing. Other manifestations of impaired motor function include fatigability, variation in strength on repeated tests, diminished range and rate of movement, loss of coordination, irregularity and clumsiness of motion, tremulousness, loss of associated movements, and lack of ability to carry out skilled acts.

Although judgment of the force exerted in either initiating or resisting movement is the major criterion in the evaluation of strength, observation and palpation of either the contraction of the muscle belly or its movement of its tendon may be helpful adjuncts. The contraction of an extremely weak muscle may sometimes be felt when it cannot be seen. In nonorganic weakness, contraction of the apparently weak muscle may be felt when the patient is asked to carry out movements with synergistic muscles (cocontraction sign), or the antagonists may be felt to contract when the patient is asked to contract the weak muscle. Weakness may be masked when attempts to contract individual weak muscles are accompanied by activation of other muscles to compensate for the loss of power. In these substitution, or “trick,” movements, the patient exploits a strong muscle with similar function to compensate for the loss of action of a weak muscle. Careful observation for alterations in normal movement patterns and substitution movements may indicate loss of function. Endurance is the ability to perform the same act repeatedly. Loss of endurance, or abnormal fatigability, may occur in myasthenia gravis. Conversely, a patient with Lambert-Eaton myasthenic syndrome may grow transiently stronger with successive contractions.

For clinical purposes, it is usually possible to evaluate muscle strength adequately without resorting to special equipment. Although the subjective impression of the examiner is usually adequate, it is at best semiquantitative and varies with the experience and ability of the examiner. There is a subjective element, with significant interexaminer and intraexaminer variability. When more quantitative determinations are necessary, various dynamometers, myometers, and ergometers are available.

The strength examination assesses primarily voluntary, or active, muscle contraction rather than reflex contraction. Strength may be classified as kinetic (the force exerted in changing position) and static (the force exerted in resisting movement from a fixed position). Strength may be tested in two ways. The patient may place a joint in a certain position, and then the examiner tries to move it. Alternately, the patient may try to move a joint or contract a muscle against the fixed resistance of the examiner. In most disease processes, both are equally affected, and the two methods can be used interchangeably. Some patients may comprehend and cooperate better with the first method, but having the patient initiate movement may better detect mild weakness. There is disagreement about how the examiner should apply force. Some authorities recommend a slow application of resistance in which the patient and examiner match effort; others contend that a rapid movement by the examiner will better detect mild weakness. With very weak muscles, strength may have to be judged without resistance or only against the resistance offered by gravity.

Many factors may complicate the strength examination and make assessment more difficult. The experience gained from examining many patients helps in buttressing an examiner's impression of loss of strength, especially when the impairment is mild. Fatigue, systemic illness, failure to understand or cooperate with strength testing, and many other factors may result in a false or distorted impression of weakness. In extrapyramidal disease, rigidity may interfere with apparent muscle power, and bradykinesia delays the onset of muscle contraction and causes retardation of movement. Hyperkinesias of various types and ataxia may make motor activity difficult. Loss or impairment of motion may also occur with pain, swelling, spasm, fractures, dislocations, adhesions or ankylosis of joints, contractures of either agonists or antagonists, loss of position sense, hysteria, malingering, and catatonia.

There is wide individual variation in the speed of voluntary movement. It may be increased in hyperthyroidism and mania and decreased in hypothyroidism, depression, parkinsonism, fatigue, and various myopathies. Slowness of

movement (bradykinesia) may be the first manifestation of extrapyramidal disease. Abnormalities in the smoothness and regularity of movement may be due to ataxia, tremor, or chorea, but incoordination may also be caused by weakness. Motor impersistence is the inability to sustain voluntary motor acts that have been initiated on verbal command. The patient is unable to sustain an activity, such as keeping the eyes closed or the hand raised. It may be a form of apraxia and has been said to occur most often with left hemisphere lesions.

Weakness of a muscle must be distinguished from loss of range of motion for other reasons and from contracture of antagonists. Passive movements to assess range of motion are sometimes necessary to help distinguish whether limitation of movement is due to weakness, pain, muscle spasm, or fibrous or bony changes. Limitation of movement due to severe weakness may ultimately result in contracture and deformity. With contracture, a muscle cannot be stretched to its normal limits without considerable resistance and the production of pain. Patients with spasticity are at particular risk for contractures, especially if the muscles are not passively stretched at regular intervals. Contractures are particularly common in the calf muscles, drawing the foot downward. There is lack of full range of motion on attempted passive dorsiflexion ("tight heel cords"). Contractures may ultimately result in periarthritic changes, joint ankylosis, and fixed deformities.

In evaluating contractures and deformities, it is important to differentiate between those of neurogenic origin and those due to orthopedic disease, congenital abnormalities, habitual postures, occupational factors, or other factors that cause mechanical difficulty with movement. An equinus or equinovarus deformity of the foot may result from any of the following: footdrop due to peroneal (fibular) nerve palsy; spasm of the calf muscles in foot dystonia; spasticity due to a corticospinal tract (CST) lesion; a developmental anomaly, such as a congenital clubfoot; or trauma or arthritis of the ankle joints. The limitation of movement in the shoulder that often complicates hemiplegia must be differentiated from shoulder joint or rotator cuff disease. Flexion contracture of the wrist joint associated with a long-standing wrist drop due to a radial nerve palsy, and the claw hand due to ulnar neuropathy, must be differentiated from the deformities seen in such conditions as arthritis, Volkmann's contracture, and Dupuytren's contracture.

The motor examination may have to be modified, often with only a rough estimation of function, in various disease states, in confused or stuporous patients, and in infants and young children. Detection of weakness in the patient

with altered consciousness or coma requires special techniques. In coma, assessment of motor function depends on spontaneous movements, the position of an extremity, or withdrawal of an extremity in response to painful stimulation, noting particularly any asymmetry of spontaneous or reflex movements on the two sides. The only manifestations of hemiplegia in the comatose patient may be the absence of contraction of the facial muscles on one side following pressure on the supraorbital ridge, the flail dropping of the wrist and forearm when released while the flexed elbow is resting on the bed, and extension and external rotation of the thigh and leg when released after having been placed in flexion with the heel resting on the bed. These are discussed in [Chapter 51](#). In infants and young children, the motor examination may be largely limited to observing spontaneous activity and noting the general posture and the positions of the extremities when the patient is prone, supine, seated, and upright. Resistance to passive movement, reflex motor responses, and palpation may provide indirect evidence of muscle strength.

Strength may be assessed in absolute terms (e.g., the examiner comparing the patient's power to a belief of what normal should be), or it may be assessed in comparison with the patient's other muscles. The comparison is most often to a homologous muscle on the other side, as in comparing the two biceps muscles. But proximal strength should be commensurate with distal strength in the same patient. A patient with polymyositis may have weakness of the deltoids on both sides, so one deltoid cannot be judged against the other. But the deltoids may be obviously weaker than the wrist extensors, so there is a proximal to distal gradient of increasing strength that is clearly abnormal. The muscles on the dominant side are usually slightly stronger.

STRENGTH SCALES

Quantitative measurements and permanent records help in diagnosis and in evaluating disease progression or recovery. In manual muscle testing, the strength of individual muscles is tested and graded quantitatively using some scale. Strength is most commonly graded using the five-level MRC (Medical Research Council) scale, which was developed in Britain in World War II to evaluate patients with peripheral nerve injuries ([Table 27.1](#)). The MRC scale has been widely applied to the evaluation of strength in general. However, the scale is heavily weighted toward the evaluation of very weak muscles ([Video 27.1](#)).

In a severe peripheral nerve injury, improvement from grade 0 (no contraction) to grade 1 (a flicker) is highly significant, as it signals the beginning of reinnervation. A patient with a nerve injury who eventually recovers to grade 4 has had an excellent outcome. In contrast, a patient with polymyositis who is diffusely grade 4 has severe disease and is doing poorly. So, the most commonly used strength grading scale has significant limitations when dealing with many patients.



Video 27.1 The MRC scale. (Courtesy Nandedkar Productions, LLC, EMG on DVD Series: Volume XIII.)

The levels of the MRC scale are precisely defined, but not linear. It is a common error to believe the MRC grades are evenly spaced and that grade 5 is normal, grade 4 is minimal or mild weakness, grade 3 is moderate weakness, grade 2 is severe, and so forth. In fact, anything less than grade 5 denotes significant weakness. Grade 4 is moderate weakness, and anything less is severe weakness. A patient who is diffusely 4/5 does not have mild or equivocal weakness but major and serious involvement. One must not confuse poor effort with weakness. A muscle is graded by the maximal power demonstrated, even if it is only briefly.

**TABLE
27.1**

The Medical Research Council Scale of Muscle Strength

- | | |
|---|-----------------------------------|
| 0 | No contraction |
| 1 | A flicker or trace of contraction |

- 2 Active movement with gravity eliminated
 - 3 Active movement against gravity
 - 4– Active movement against gravity and slight resistance
 - 4 Active movement against gravity and moderate resistance
 - 4+ Active movement against gravity and strong resistance
 - 5 Normal power
-

The muscle strength scale used by physical therapists grades muscles on a 6-point scale from 0 (no motion) through trace, poor, fair, and good to normal. The scale used at the Mayo Clinic grades muscle strength on a 5-point scale that is more linear. Normal is designated as 0, mild weakness is –1, and total paralysis is –4.

The different levels of the MRC scale are so precisely defined that there is good interexaminer consistency once the fine points of proper positioning and other details are mastered. In clinical practice, the MRC scale is often expanded to include subgrades (e.g., 5–, 4+). The MRC eventually saw the need to include grades 4– and 4+. Mendell and Florence developed a formal modified MRC scale. In general use, the subgrades are not so precisely defined, and there is much less inter- and even intraexaminer consistency. Approximate guidelines are all that is possible. Before attempting to describe these guidelines, the important issue of examiner-patient mismatch should be recognized.

There is obviously considerable individual variation in muscle power, dependent in part upon size, gender, body build, age, and activity level. This variability affects examiners as well as patients. Patients come large and small, young and old, male and female, and physically strong and relatively weak. So do physicians. A large, young, powerful, male physician examining a small, old, sick, female patient has an unfair advantage. He may tend to think she is weak when she is in fact normal for her age, sex, and circumstances. Conversely, a small, relatively weak female physician examining a large, powerful man may miss significant weakness because of strength mismatch.

As a general principle, reliable strength testing should attempt to break a given muscle. Muscles are most powerful when maximally shortened. Another consideration is the lever effect. Attempting to overpower a muscle, such as a

deltoid, using a very short lever (examiner's hand at the mid-upper arm) is much less likely to meet with success than using a long lever (patient's elbow extended and examiner pressing down on the wrist). A small, weak examiner can overcome the deltoid of the most powerful man by keeping the patient's elbow extended and using both hands to pull down the wrist. Technique matters.

By varying the length of lever and the shortening of the muscle permitted, the examiner may give or take mechanical advantage as necessary to compensate for strength mismatch. Many patients of different ages, sizes, and strength levels must be examined in this fashion in order to develop an appreciation of the expected strength of a muscle for a given set of circumstances.

So, for an examiner of average size and strength examining a patient of average size and presumed normal strength, the following is a useful guideline for assessing power in the major muscle groups. If it requires the whole hand and a firm push to break the muscle, the power is grade 4+. If the muscle can be broken using three fingers, it is grade 4. If it can be broken using one finger, it is grade 4-. Some clinicians use the grades of 3- and 2- to describe muscles that can move against gravity or with gravity eliminated, but not through a full range, and the grade of 5- to indicate borderline or equivocal weakness.

Some muscles are “special cases.” The small hand muscles are best examined by matching them against the examiner’s like muscle (e.g., abductor pollicis brevis [APB] to abductor pollicis brevis). This method is beautifully described and illustrated in *Segmental Neurology* by John K. Wolf. The gastrocnemius muscles are normally so powerful it is virtually useless to examine them using hand and arm strength, unless they are very weak. Having the patient walk on tiptoe, hop, support the entire body weight on one tiptoe, or do one-legged toe raises are usually better methods.

**TABLE
27.2**

Features of Upper Motor Neuron versus Lower Motor Neuron Weakness

Feature	Upper Motor Neuron	Lower Motor Neuron
Weakness distribution	Corticospinal distribution; hemiparesis, quadriparesis, paraparesis,	Generalized, predominantly proximal, predominantly distal or focal. No preferential involvement of corticospinal innervated muscles

		monoparesis, faciobrachial
Sensory loss	Central pattern distribution	None, stocking glove or peripheral nerve or root distribution
Deep tendon reflexes	Increased unless very acute	Normal or decreased
Superficial reflexes	Decreased	Normal
Pathologic reflexes	Yes	No
Sphincter function	Sometimes impaired	Normal (except for cauda equina lesion)
Muscle tone	Increased	Normal or decreased
Pain	No	Sometimes
Other CNS signs	Possibly	No

CNS, central nervous system.

PATTERNS OF WEAKNESS

There are common patterns of weakness. Recognition of a pattern may help greatly in lesion localization and differential diagnosis. Identification of the process causing weakness is further aided by accompanying signs, such as reflex alterations and sensory loss. [Table 27.2](#) reviews the features of upper motor neuron versus lower motor neuron weakness. [Table 27.3](#) summarizes some common patterns of weakness and their localization.

Weakness may be focal or generalized. When focal, it may follow the distribution of some structure in the peripheral nervous system, such as a peripheral nerve or spinal root. It may affect one side of the body in a “hemi”

distribution. A hemi distribution may affect the arm, leg, and face equally on one side of the body, or one or more areas may be more involved than others. The CST preferentially innervates certain muscle groups, and these are often selectively impaired (see [Chapter 25](#)). When weakness is nonfocal, it may be generalized, predominantly proximal, or predominantly distal.

Generalized Weakness

The term *generalized weakness* implies that the weakness involves both sides of the body, more or less symmetrically. When a patient has truly generalized weakness, bulbar motor functions—such as facial movements, speech, chewing, and swallowing—are involved as well. Weakness of both arms and both legs with normal bulbar function is quadriplegia or tetraparesis. Weakness of both legs is paraparesis. When weakness affects all four extremities, the likely causes include spinal cord disease, peripheral neuropathy, a neuromuscular junction disorder, or a myopathy.

**TABLE
27.3**

Common Patterns of Weakness with Lesions at Different Locations in the Neuraxis

Location of Lesion	Distribution of Weakness	Sensory Loss	DTRs*	Possible Accompanying Signs
Middle cerebral artery	Contralateral arm and face > leg ^b	Y	Incr	Aphasia, apraxia, visual field deficit, gaze palsy
Anterior cerebral artery	Contralateral leg > arm and face ^b	Y	Incr	Cortical sensory loss in contralateral leg, frontal lobe signs; sometimes incontinence
Internal capsule	Contralateral face = arm = leg ^b	N	Incr	None ("pure motor stroke")
Brainstem	Ipsilateral cranial nerve and contralateral body ^b	Y	Incr	Variable, depending on level
Cervical cord (transverse)	Both arms and both legs ^b	Y	Incr	Bowel, bladder, or sexual dysfunction common
Thoracic cord (transverse)	Both legs ^b	Y	Incr	Bowel, bladder, or sexual dysfunction common
Cauda equina	Both legs, asymmetric, multiple root pattern	Y	Decr	Occasional bowel, bladder, or sexual dysfunction; sometimes pain
Anterior horn cell	Focal early, generalized late	N	Incr	Atrophy, fasciculations, bulbar weakness
Single nerve root	Muscles of the affected myotome	Y	Decr	Pain
Plexus	Plexus pattern, complete or partial	Usually	Decr	Pain is common, especially with brachial "plexitis"
Mononeuropathy	Muscles of the affected nerve	Usually	Decr	Variable atrophy, variable pain
Polyneuropathy	Distal > proximal	Usually	Decr	Variable pain, atrophy late
Neuromuscular junction	Bulbar, proximal extremities	N	Normal	Ptosis, ophthalmoparesis, fatigable weakness, fluctuating weakness
Muscle	Proximal > distal	N	Normal	Pain uncommon, many potential patterns (limb girdle, facioscapulohumeral, etc.), pseudohypertrophy, myotonia

*With corticospinal lesions, DTRs acutely may be normal or decreased (neural shock).

^aExtremity weakness in a corticospinal tract distribution.

Decr, decreased; DTR, deep tendon reflex; Incr, increased; N, no; Y, yes.

When spinal cord disease is the culprit and the deficit is incomplete, more severe involvement of those muscles preferentially innervated by the CST can frequently be discerned. Reflexes are usually increased (though in the acute stages they may be decreased or absent); there is usually some alteration of sensation, sometimes a discrete spinal "level"; superficial reflexes disappear; and there is often bowel and bladder dysfunction. Generalized peripheral nerve disease tends to predominantly involve distal muscles, although there are exceptions. There is no preferential involvement of CST innervated muscles; reflexes are usually decreased; sensory loss is frequently present; and bowel and bladder functions are not disturbed. With a neuromuscular junction disorder, the weakness is likely to be worse proximally; sensation is spared; reflexes are normal; and there is usually involvement of bulbar muscles, especially with ptosis and ophthalmoplegia. When the problem is a primary muscle disorder, weakness is usually more severe proximally; reflexes are normal; sensation is normal; and with only a few exceptions, bulbar function is spared except for occasional dysphagia.

Amyotrophic lateral sclerosis (ALS) causes a characteristic pattern of

weakness. The weakness and wasting due to lower motor neuron involvement are accompanied by weakness and hyperreflexia due to upper motor neuron involvement.

Focal Weakness

Weakness of the arm and leg on one side of the body is hemiparesis. Monoparesis is weakness limited to one extremity. Diplegia is weakness of like parts on the two sides of the body; the term spastic diplegia refers to weakness of both legs that occurs in cerebral palsy; and facial diplegia is weakness of both sides of the face. Spastic weakness of one arm and the opposite leg is referred to as cruciate or crossed paralysis, or hemiplegia alternans.

Certain patterns of muscle weakness point to a peripheral nerve, plexus, or root lesion. With a peripheral nerve lesion, all muscles below the level of the lesion are at risk. It is increasingly recognized, however, that not all muscles distal to a peripheral nerve lesion are necessarily equally affected. When multiple muscles of an extremity are weak, localization depends on recognizing the common innervating structure. In cervical radiculopathy, the muscles involved are innervated by different peripheral nerves and different brachial plexus components, but all by the same root. For instance, lesions of the middle trunk of the brachial plexus are exceedingly rare, so weakness of the triceps (radial nerve) and the pronator teres (PT) (median nerve) usually means a lesion of the C7 root.

A focal neuropathy, such as a radial nerve palsy, or a spinal root lesion, such as from a herniated disc, causes weakness limited to the distribution of the involved nerve or root. A complete plexopathy, such as a traumatic brachial plexopathy, may cause weakness of the entire limb. Partial lesions may cause weakness only in the distribution of certain plexus components. With such lower motor neuron pathology, reflexes are typically decreased, and there is often accompanying sensory loss. Localization of focal weakness due to root, plexus, and peripheral nerve pathology requires intimate familiarity with peripheral neuroanatomy. Anterior horn cell disease often begins with focal weakness that may simulate mononeuropathy, but it evolves into a more widespread pattern as the disease progresses, culminating in generalized weakness. Except for extraocular muscle involvement in myasthenia gravis, it is rare for a myopathy or neuromuscular junction disorder to cause focal weakness.

NONORGANIC (FUNCTIONAL) WEAKNESS

Nonorganic weakness due to psychiatric disorders—such as hysteria, conversion disorder, malingering, or depression—is common. The first step in evaluating weakness is often deciding whether it is organic or nonorganic. This distinction is not always easy. Patients with nonorganic weakness are commonly thought to have neurologic disease, but it seems nearly as often patients with real weakness are thought to be hysterical or malingering. Organic and functional deficits may coexist—the problem of functional or psychogenic overlay. Some patients embellish by nature; others may exaggerate a real deficit for fear the physician may not take them seriously. A patient may have real weakness in one area and functional weakness in another area. Some patients with organic weakness may allow resistance to collapse when the examiner begins to defeat a muscle so that real weakness may appear to be related to poor effort. When a movement causes pain, resistance may be less than full and the effort may be erratic.

Coaching is often helpful. The examiner exhorts the patient not to give up, to keep pushing or pulling no matter what. Simple encouragement to keep trying even if the patient is losing may suffice to improve effort. Some patients, in spite of all, will simply not give full effort. Their efforts are erratic and variable. Some things are often useful in distinguishing organic from psychogenic weakness. Patients with bona fide organic muscle weakness will yield smoothly as the examiner defeats the weak muscle. The patient gives uniform resistance throughout the movement. If the examiner decreases his resistance, the patient will begin to win the battle. If the examiner drops the resistance level, the patient with nonorganic weakness will not continue to push or pull. Instead, the patient will also stop resisting so that no matter how little force the examiner applies, there is an absence of follow-through and the patient never overcomes the examiner. When there is nonorganic weakness, resistance is erratic and often collapses abruptly. The muscular contractions are poorly sustained and may give way suddenly, rather than gradually, as the patient resists the force exerted by the examiner. Some patients will give up entirely and allow the muscle or limb to flop; others will provide variable resistance throughout the range of motion with alternating moments of effort and no effort. At the peaks of contraction, strength is normal; in the valleys, there is little or no resistance. This pattern of variable strength is referred to as “collapsing,” “ratchety,” “give way,” “breakaway,” or “catch and give.” It is characteristic of nonorganic weakness. With nonorganic weakness, there may be an increase or a decrease in strength with repeated

testing. Contraction of the apparently weak muscle may be felt when the patient is asked to carry out movements with synergistic muscles, or the antagonists may be felt to contract when the patient is asked to contract the agonist (e.g., the triceps muscle twitches when the patient is told to flex the elbow). Functional testing may fail to confirm weakness suspected during strength testing. For example, there may be apparent foot dorsiflexion weakness, yet the patient is able to stand on the heel without difficulty.

The patient with nonorganic weakness may make little effort to contract the muscles necessary to execute a particular movement. He may be calm and indifferent while demonstrating the lack of strength, showing little sign of alarm at the presence of complete paralysis, and smile cheerfully during the examination. If the examiner raises and drops an extremity, a limb with psychogenic paralysis may drop slowly to avoid injury, whereas an extremity with real weakness would drop rapidly, especially if the paralysis is flaccid. In psychogenic paralysis of the arm, the latissimus dorsi may appear paretic when tested by having the patient adduct the arm but contract normally on coughing. In simulated hemiplegia, the patient may be unable to adduct either the affected arm or leg against resistance, yet if asked to keep both arms against the body or both legs close together, the adductors contract strongly on both sides because it is difficult to adduct one extremity without adducting its apparently paralyzed fellow. In testing paralysis of the finger muscles, the patient may be asked to pronate the forearms and interlock the fingers so that the left fingers are on the right and vice versa ([Figure 36.6](#)); the examiner then points to the individual fingers and tells the patient to move them. It is difficult for one to determine immediately whether the indicated finger is on the right side or on the left; if the patient attempts to respond promptly, he or she makes many mistakes. Similar tests may be carried out by asking the patient to perform individual movements with the hands behind the back.

The patient with functional weakness cannot control associated movements, which forms the basis of the Hoover and abductor signs. The Hoover (automatic walking) sign is useful for evaluating suspected nonorganic leg weakness. When a normal supine patient flexes the hip to lift one leg, there is an involuntary downward movement of the other leg. The extension countermovement of the opposite leg is a normal associated movement (see [Chapter 42](#)). An extension movement of one leg normally accompanies flexion of the other leg, as in walking. In organic leg weakness, the downward pressure of the contralateral heel occurs when the patient tries to raise the weak leg, and the examiner can

feel the extension pressure by placing a hand beneath the heel that remains on the bed. In nonorganic leg weakness, there is no downward pressure of the contralateral heel as the patient tries to raise the weak leg, but an extension movement of the “paralyzed” leg may be felt as the good leg is raised. Hoover’s sign is the absence of the expected extension movement of the good leg on attempting to raise the bad leg and normal involuntary extension of the bad leg on raising the good leg.

In the quest for ways to distinguish real from hysterical weakness at the turn of the 20th century, Hoover thought this maneuver was more useful than the Babinski sign ([Chapter 40](#)). The abductor sign is similar and seeks to elicit synergistic movement of the nonparetic leg when the patient is asked to abduct the paretic leg. In nonorganic paresis, the paretic leg demonstrates synergistic abduction when the sound leg is tested, and the sound leg does not exert normal abduction power and can be moved into a hyperadducted position when the paretic limb is tested. It may be useful when relatively preserved hip extensor strength limits the utility of Hoover’s test. In the Spinal Injuries Center (SIC) test, patients unable to raise their knees spontaneously have a positive test, indicating nonorganic weakness, when their knees remain up after being lifted by the examiner. See LaFrance for video of the Hoover, abduction, and SIC tests. Dynamometers have been used in various ways in an attempt to prove weakness is nonorganic, and investigators have developed computerized, quantitative methods for detecting functional weakness based on these principles.

Patients with nonorganic hemiparesis are more likely to have weakness of the sternomastoid, usually ipsilateral, but this muscle is rarely involved in bona fide hemiparesis because of its bilateral innervation ([Chapter 19](#)). With the arm-dropping test, transient voluntary contraction in functional weakness may cause a brief hang-up before the arm collapses. On the Babinski trunk-thigh test, a leg with nonorganic weakness fails to flex as expected. Looking for the presence or absence of other associated movements may sometimes help in the differentiation between organic and psychogenic weakness ([Chapter 42](#)). The patient with functional weakness may have a dramatic display of effort, grimacing and grunting when trying to contract the weak muscle. A movement such as a squat may be executed in a slow and laborious manner that actually requires greater than normal strength. The onset of functional weakness is often sudden.

In psychogenic weakness, muscle tone may be normal, decreased, or variable, but it is often increased with pseudorrigidity or pseudospasticity. Rigidity, if

present, resembles voluntary resistance. The part may be held firmly in a bizarre position. Abnormalities in tone usually vary from time to time, especially under the influence of suggestion.

Normal tone, normal deep tendon reflexes (DTRs), normal facial movements in the face of hemiparesis, an absence of trophic changes, normal sphincter function despite paraparesis or quadriplegia, aphonia, and a monoplegic dragging gait all may suggest that weakness is nonorganic. It is very unusual for functional weakness to present in a peripheral nerve, plexus, or root distribution or to display a proximal/distal gradient. Evidence does not support the notion that functional weakness is more common on the left.

DSM-V emphasizes the importance of “positive signs” of nonorganicity to support a diagnosis of conversion disorder. Many such positive signs have been reported, but only a minority have been validated. The positive signs that have been carefully studied generally have low sensitivity but high specificity. In a carefully done pilot study, Daum et al. concluded that four positive signs were highly reliable in the diagnosis of functional weakness: Hoover’s sign, giveaway weakness, cocontraction, and drift without pronation.

EXAMINATION OF MOTOR STRENGTH AND POWER

Evaluation of the strength of various muscle groups and movements can become complex, depending upon the degree of detail necessary. Isolated contraction of a single muscle is rarely possible because muscles with similar functions participate in almost every movement. Normal contraction of synergists and fixating muscles and relaxation of antagonists are also necessary. Still, the predominant action of a single muscle can usually be determined and tested. Some functions are carried out by many muscles acting synergistically (e.g., flexion and extension of the trunk), and the muscles must be tested as a group by assessing the movement rather than individual muscles. It is helpful to be fluent with the primary actions of the major muscles and their peripheral nerve, plexus, and root innervations. The strength of each major pertinent muscle group should be determined individually and its strength grade recorded.

Reliable strength testing requires proper patient positioning and avoidance of unwanted movements. Testing may be done in various positions, depending on the muscle to be tested and its power. Testing in the seated position suffices

under most circumstances. It is important to fix the proximal portion of a limb when the movements of the distal portion are being tested. For instance, when testing forearm pronation strength, the patient must not be allowed to internally rotate the shoulder to compensate for lack of pronation power. When evaluating very weak muscles, gravity must be eliminated to detect residual power. A very weak biceps muscle (MRC grade 2/5), even when it cannot succeed against gravity, may be able to flex the elbow if the arm is raised to shoulder height so that the forearm can be moved horizontally. The wrist and finger drop of radial nerve palsy creates such a mechanical disadvantage for contraction that the patient may appear to have weakness of grip and finger abduction, but these functions are intact when the wrist and fingers are passively extended.

EXAMINATION OF SPECIFIC MOVEMENTS AND MUSCLES

The motor examination of the muscles supplied by those cranial nerves that have motor functions is discussed separately. For the trunk and extremities, the strength and power of the individual muscles and of movements are assessed as appropriate for the clinical circumstances. Many reference sources are available to assist in learning muscle examination techniques. There is some difference regarding the exact innervation of individual muscles among different reference sources, and occasionally, there is variable or anomalous innervation. [Tables 27.4 to 27.7](#) give the most generally accepted spinal cord segment and peripheral nerve innervation of the more important muscles. [Tables 27.8 and 27.9](#) give the innervation by root.

The action of a muscle about a joint may vary depending on the part of the muscle activated and the position of the joint. Parts of some large muscles, such as the gluteus maximus, may have secondary actions that are different from other parts. For instance, the upper part of the gluteus maximus abducts the hip, whereas the lower part adducts it. Still, each muscle has a primary action in which all parts participate, and in this instance, it is hip extension. The angle of the joint about which a muscle acts may influence its leverage and angle of pull. In extreme cases, a muscle may move to the opposite side of the axis of rotation and have an action in one joint position that is different from its action in another joint position (inversion of action). For instance, the hip adductors act secondarily as hip flexors when the hip is extended but as extensors when the hip

is flexed; the piriformis externally rotates the extended hip but internally rotates the flexed hip.

**TABLE
27.4**

Innervation of Muscles Responsible for Movements of the Head and Neck

Parentheses signify minor contribution

Muscle	Segmental Innervation	Peripheral Nerve
Sternocleidomastoid	Cranial XI; C(1)2-C3	Spinal accessory nerve
Trapezius	Cranial XI; C(2)3-C4	Spinal accessory nerve
Scalenus anterior	C4-C7	
Scalenus medius	C4-C8	
Scalenus posterior	C6-C8	
Longus capitis	C1-C4	
Longus colli	C2-C6	
Rectus capitis anterior	C1-C2	Suboccipital nerve
Rectus capitis lateralis	C1	Suboccipital nerve
Rectus capitis posterior	C1	Suboccipital nerve
Obliquus capitis inferior	C1	Suboccipital nerve
Obliquus capitis superior	C1	Suboccipital nerve
Splenius capitis	C2-C4 (C1-C6)	
Splenius cervicis	C2-C4 (C1-C6)	

Semispinalis capitis	C1-C4
Semispinalis cervicis	C3-C6
Spinalis cervicis	C5-C8
Sacrospinalis	C1-C8
Iliocostalis cervicis	C1-C8
Longissimus capitis	C1-C8
Longissimus cervicis	C1-C8
Intertransversarii	C1-C8
Rotatores	C1-C8
Multifidi	C1-C8

Examination of Movements and Muscles of the Neck

The principal neck movements are flexion, extension (retraction), rotation (turning), and lateral bending (tilting, abduction). Many different muscle groups contribute to the various neck movements. Except for the sternocleidomastoid (SCM) and trapezius, it is not possible to examine them individually, and the assessment is made of movement (e.g., neck flexion) rather than particular muscles. The spinal accessory nerve, along with the second, third, and fourth cervical segments, supplies the SCM and trapezius muscles. The SCM is a flexor and rotator of the head and neck; the trapezius retracts the neck and draws it to one side. Other muscles that contribute to neck flexion include the platysma, suprathyroid, infrathyroid, scalenes, and prevertebral group of muscles (longus colli and capitis, rectus capitis).

Many muscles contribute to neck extension, including the trapezius and the paravertebral muscles. Many of these muscles when contracting unilaterally rotate the spine. The paravertebral musculature is a massive, complex amalgam of individual muscle groups that primarily serve to extend and rotate the neck.

and trunk. Four principal muscle subgroups combine to form the paravertebral muscles: the splenii, erector spinae, transversospinalis, and interspinal-intertransverse. All of these muscles lie deep and medial in the groove formed between the transverse and spinous processes of the vertebral bodies. They are further named for the vertebral segment in which they lie (e.g., transversospinalis cervicis). The splenius capitis is a powerful ipsilateral rotator of the head; the splenii contracting bilaterally extend the neck. Muscles commonly injected with botulinum toxin to treat cervical dystonia include the SCM, trapezius, splenius capitis, scalenes, longissimus, and levator scapula. The paraspinal muscle complex generally receives its innervation from multiple levels. These muscles arise from a common embryologic precursor muscle mass, and their innervation shows extensive longitudinal overlap ([Table 27.4](#)).

Neck flexors are tested by having the patient try to place the chin on the chest as the examiner applies extension force to the forehead ([Figure 27.1](#)). Extensors are tested by having the patient extend against the examiner's resistance applied to the occiput ([Figure 27.2](#)). Neck rotation is accomplished by the contralateral SCM and ipsilateral splenius capitis and trapezius; examination of the SCM and trapezius muscles is discussed in [Chapter 19](#). Neck flexor strength may be tested with the patient sitting or supine and neck extension sitting or prone. The neck flexion test consists of measuring the time the supine patient can keep the head raised with the chin on the chest; most patients can keep their head in this position for at least 1 minute. This test is sometimes useful in the evaluation of myopathies and neuromuscular junction disorders, the principal conditions that cause neck flexor or extensor weakness. Abnormalities of neck position may occur in conditions in which there is no muscle weakness, and these can sometimes be confused with the effects of a weak muscle. The patient with meningismus may have the head retracted, whereas the patient with Parkinson disease may have it flexed. Cervical dystonia can cause an almost infinite number of abnormal head positions, including torticollis (head turned or tilted), anterocollis (head flexed), and retropcollis (head extended). Examination of the neck muscles must be done carefully in any patient at risk for cervical spine disease.

TABLE 27.5 Innervation of Muscles Responsible for Movements of the Shoulder Girdle and Upper Extremity

(Parentheses signify minor contribution)

Muscle	Segmental Innervation	Peripheral Nerve	Muscle	Segmental Innervation	Peripheral Nerve
Trapezius	Cranial XI; C12/3-C4	Spinal accessory nerve	Extensor indicis proprius	C7-C8	Radial nerve
Levator scapulae	C3-C4	Nerves to levator scapulae	Extensor digiti minimi	C7-C8	Radial nerve
	C5	Dorsal scapular nerve	Extensor pollicis longus	C7-C8	Radial nerve
Rhomboideus major	C4-C5	Dorsal scapular nerve	Extensor pollicis brevis	C7-C8	Radial nerve
Rhomboideus minor	C4-C5	Dorsal scapular nerve	Abductor pollicis longus	C7-C8	Radial nerve
Serratus anterior	C5-C7	Long thoracic nerve	Pronator teres	C6-C7	Median nerve
Deltoid	C5-C6	Axillary nerve	Flexor carpi radialis	C6-C7	Median nerve
Teres minor	C5-C6	Axillary nerve	Pronator quadratus	C7-C8	Median nerve
Supraspinatus	C4/5-C6	Suprascapular nerve	Palmaris longus	C7-C8	Median nerve
Infraspinatus	C4/5-C6	Suprascapular nerve	Flexor digitorum sublimis	C7-T1	Median nerve
Latissimus dorsi	C6-C8	Thoracodorsal nerve	Flexor digitorum profundus (radial half)	C8-T1	Median nerve
Pectoralis major	C5-T1	Lateral and medial anterior thoracic nerves	Lumbricales 1 and 2	C8-T1	Median nerve
Pectoralis minor	C7-T1	Medial anterior thoracic nerve	Flexor pollicis longus	C7-T1	Median nerve
Subscapularis	C5-C7	Subscapular nerves	Flexor pollicis brevis (FPB) (lateral head)	C8-T1	Median nerve
Teres major	C5-C7	Lower subscapular nerve	Abductor pollicis brevis	C8-T1	Median nerve
Subclavius	C5-C6	Nerve to subclavius	Opponens pollicis	C8-T1	Median nerve
Coracobrachialis	C6-C7	Musculocutaneous nerve	Flexor carpi ulnaris	C7-T1	Ulnar nerve
Biceps brachii	C5-C6	Musculocutaneous nerve	Flexor digitorum profundus (ulnar half)	C8-T1	Ulnar nerve
Brachialis	C5-C6	Musculocutaneous nerve	Interossei	C8-T1	Ulnar nerve
Brachioradialis	C5-C6	Radial nerve	Lumbricales 3 and 4	C8-T1	Ulnar nerve
Triceps brachii	C6-C8	Radial nerve	FPB (medial head)	C8-T1	Ulnar nerve
Anconeus	C7-C8	Radial nerve	Flexor digiti minimi brevis	C8-T1	Ulnar nerve
Supinator	C6-C7	Radial nerve	Abductor digiti minimi (ADM)	C8-T1	Ulnar nerve
Extensor carpi radialis longus	C5/6-C7	Radial nerve	Opponens digiti minimi	C8-T1	Ulnar nerve
Extensor carpi radialis brevis	C7-C8	Radial nerve	Palmaris brevis	C8-T1	Ulnar nerve
Extensor carpi ulnaris	C7-C8	Radial nerve	Adductor pollicis	C8-T1	Ulnar nerve
Extensor digitorum communis	C7-C8	Radial nerve			

TABLE 27.6 Innervation of Muscles Responsible for Movements of the Thorax and Abdomen

(Parentheses signify minor contribution)

Muscle	Segmental Innervation	Peripheral Nerve
Diaphragm	C3-C5	Phrenic nerve
Intercostal muscles (internal and external)	T1-T12	Intercostal nerves
Levatores costarum	C8-T11	Intercostal nerves
Transversus thoracis	T2-T7	Intercostal nerves
Serratus posterior superior	T1-T4	Intercostal nerves
Serratus posterior inferior	T9-T12	Intercostal nerves
Rectus abdominis	T5-T12	Intercostal nerves
Pyramidalis	T11-T12	Intercostal nerves
Transversus abdominis	T7-L1	Intercostal, ilioinguinal, and iliohypogastric nerves
Obliquus internus abdominis	T7-L1	Intercostal, ilioinguinal, and iliohypogastric nerves
Obliquus externus abdominis	T7-L1	Intercostal, ilioinguinal, and iliohypogastric nerves

Examination of Movements and Muscles of the Upper Extremities

The responsible muscles and their innervation are given in [Table 27.5](#).

The Shoulder

Movements of the shoulder take place at the sternoclavicular, acromioclavicular, and glenohumeral joints. Because the scapula is firmly connected to the clavicle at the acromioclavicular joint, the two bones tend to move as a unit with the motion taking place primarily at the sternoclavicular joint. Movements of the scapula are elevation, depression, retraction (movement away from the chest wall), protraction (movement toward the chest wall), and rotation. The ventral surface of the scapula is a concavity known as the subscapular fossa that is filled mostly with the subscapularis muscle. The serratus anterior lies between the subscapularis and the chest wall and inserts into a thin rim of the scapula along the vertebral border and slightly expanded triangular areas at the superior and inferior angles ([Figure 27.3](#)). The serratus runs obliquely from its origination from the upper eight ribs along the lateral chest wall to its attachment to the scapula. The trapezius is a diamond-shaped muscle that attaches widely to the shoulder girdle. The superior fibers insert along the posterior border of the clavicle and scapular spine, the middle and lower fibers along the scapular spine. The upper and middle fibers insert laterally along the scapular spine and the lower fibers more medially. The rhomboids (major and minor) arise from the spinous process of the upper thoracic vertebrae and insert along the medial border of the scapula. The levator scapulae originates from the upper cervical vertebra and drops diagonally to insert along the upper medial border of the scapula.

The upper fibers of the trapezius, assisted by the levator scapulae, elevate the scapula and the point of the shoulder and rotate the scapula upward. The middle fibers rotate the scapula upward and assist the rhomboids in retraction. The lower fibers rotate and depress the scapula and draw it toward the midline. The rhomboids act primarily to retract the scapula, bracing the shoulder backward. The levator scapulae acts with the trapezius to elevate the scapula. The serratus anterior, assisted by the pectoralis minor, protracts the scapula, pulling it anteriorly. It is critical in all functions that involve reaching or pushing forward. The expanded insertion at the inferior angle helps to pull the inferior scapular angle forward around the chest wall. It also, along with the trapezius, rotates the scapula and raises the point of the shoulder to abduct the arm above horizontal. It helps to fix the scapula while other muscles abduct or flex the arm.

Elevation of the scapula, as in shrugging the shoulder, is carried out by the upper trapezius and levator scapulae muscles, assisted by the SCM. The levator scapulae is innervated by direct branches from C3 and C4 with a contribution from C5 via the dorsal scapular nerve. The levator scapulae draws the scapula

upward and rotates it so that the inferior angle approaches the spinal column. Depression of the scapula is carried out primarily by the lower trapezius, pectoralis minor, and subclavius muscles.

TABLE 27.7 Innervation of Muscles Responsible for Movements of the Lower Extremities

Parentheses signify minor contribution					
Muscle	Segmental Innervation	Peripheral Nerve	Muscle	Segmental Innervation	Peripheral Nerve
Psoas major	L1 2-L3 4	Nerve to psoas major	Tibialis posterior	L5-S1	Tibial nerve
Psoas minor	L1-L2	Nerve to psoas minor	Flexor digitorum longus	L5-S1	Tibial nerve
Iliacus	L2-L3 4	Femoral nerve	Flexor hallucis longus	L5-S1	Tibial nerve
Quadriceps femoris	L2-L4	Femoral nerve	Biceps femoris (short head)	L5-S2	Common peroneal nerve
Sartorius	L2-L3	Femoral nerve	Tibialis anterior	L4-L5	Deep peroneal nerve
Pectenius	L2-L3	Femoral nerve	Peroneus tertius	L5-S1	Deep peroneal nerve
Gluteus maximus	L5-S2	Inferior gluteal nerve	Extensor digitorum longus	L5-S1	Deep peroneal nerve
Gluteus medius	L4-S1	Superior gluteal nerve	Extensor hallucis longus	L5	Deep peroneal nerve
Gluteus minimus	L4-S1	Superior gluteal nerve	Extensor digitorum brevis	L5-S1	Deep peroneal nerve
Tensor fasciae latae	L4-S1	Superior gluteal nerve	Extensor hallucis brevis	L5-S1	Deep peroneal nerve
Piriformis	L5 S1-S2	Nerve to piriformis	Peroneus longus	L5-S1	Superficial peroneal nerve
Adductor longus	L2-L4	Obturator nerve	Peroneus brevis	L5-S1	Superficial peroneal nerve
Adductor brevis	L2-L4	Obturator nerve	Flexor digitorum brevis	S1-S2	Medial plantar nerve
Adductor magnus	L2-L4	Obturator nerve	Flexor hallucis brevis	S1-S2	Medial plantar nerve
Adductor magnus	L4-L5	Sciatic nerve	Abductor hallucis	S1-S2	Medial plantar nerve
Gracilis	L2-L4	Obturator nerve	Lumbricales (medial 1 or 2)	S1-S3	Medial plantar nerve
Obturator externus	L2-L4	Obturator nerve	Quadratus plantae	S1-S2	Lateral plantar nerve
Obturator internus	L5-S1	Nerve to obturator internus	Adductor hallucis	S2-S3	Lateral plantar nerve
Gemellus superior	L5-S1	Nerve to obturator internus	ADM pedis	S1-S3	Lateral plantar nerve
Gemellus inferior	L5-S1	Nerve to quadratus femoris	Flexor digiti minimi brevis	S2-S3	Lateral plantar nerve
Quadratus femoris	L5-S1	Nerve to quadratus femoris	Lumbricales (lateral 2 or 3)	S1-S3	Lateral plantar nerve
Biceps femoris (long head)	L5-S1	Tibial nerve	Interossei	S2-S3	Lateral plantar nerve
Semimembranosus	L5-S1	Tibial nerve			
Semitendinosus	L5-S2	Tibial nerve			
Popliteus	L5-S1	Tibial nerve			
Gastrocnemius	S1-S2	Tibial nerve			
Soleus	S1-S2	Tibial nerve			
Plantaris	S1-S2	Tibial nerve			

Retraction of the scapula is carried out primarily by the rhomboids and the middle trapezius. The rhomboids also draw the scapulae together, as in standing at attention. The rhomboids are innervated by a twig directly from the C5 nerve

root, and not via the brachial plexus. Examination of the rhomboids is important in the differentiation of C5 radiculopathy from upper trunk brachial plexopathy. In protraction of the scapula, the scapula moves forward as in throwing a punch. This movement is carried out primarily by the serratus anterior (long thoracic nerve, C5-C7). The serratus keeps the vertebral border of the scapula applied to the thorax and pulls the scapula forward and laterally. Rotation of the scapula is accomplished by the trapezius, serratus anterior, pectorals, rhomboids, and latissimus dorsi. Normal scapular rotation is essential to efficient shoulder abduction.

**TABLE
27.8**

**Major Upper-Extremity Muscles Innervated by
Different Roots**

Parentheses signify minor contribution

Root Muscles Supplied

C4	Levator scapulae, rhomboids
C5	Levator scapulae, rhomboids, supraspinatus, infraspinatus, teres major and minor, deltoid, biceps, brachialis, BR, serratus anterior, pectoralis
C6	Supraspinatus, infraspinatus, teres major and minor, deltoid, biceps, brachialis, BR, supinator, serratus anterior, pectoralis, FCR, pronator teres, latissimus dorsi, ECRL (triceps)
C7	Serratus anterior, pectoralis, teres major, latissimus dorsi, triceps, anconeus, pronator teres, FCR, ECRL, EDC, ECU, supinator (EIP, FCU, FDS, FPL, extensor pollicis longus/brevis)
C8	Latissimus dorsi, pectoralis, triceps, anconeus, EDC, ECU, EIP, extensor pollicis longus/brevis, FCU, FDS, FDP, FPL, PQ, APB, APL, OP, AP, ADM, lumbricals, interossei
T1	T1 Pectoralis, FCU, FDS, FDP, FPL, APB, OP, AP, ADM, lumbricals, interossei

ADM, abductor digiti minimi; AP, adductor pollicis; APB, abductor pollicis brevis; APL, abductor pollicis longus; BR, brachioradialis; ECRL, extensor carpi radialis longus; ECU, extensor carpi

ulnaris; EDC, extensor digitorum communis; EIP, extensor indicis proprius; FCR, flexor carpi radialis; FCU, flexor carpi ulnaris; FDP, flexor digitorum profundus; FDS, flexor digitorum superficialis; FPL, flexor pollicis longus; OP, opponens pollicis; PQ, pronator quadratus.

Scapulohumeral Rhythm

There are two motions involved in abducting the arm: scapulothoracic and glenohumeral. The scapulothoracic motion is the movement of the scapula in relation to the chest wall; the glenohumeral is the movement at the shoulder joint. These two motions normally occur in harmony to effect smooth arm movements. When the scapula does not move normally, the arm cannot raise normally. As abduction begins, the scapular muscles—especially serratus anterior—fix the scapula, so the pull of the deltoid is on the humerus and not the scapula. As the deltoid abducts the shoulder toward 90 degrees, the serratus anterior and trapezius rotate the scapula. The serratus pulls forward and laterally on the inferior angle, holding the scapula close to the chest wall, while the upper fibers of the trapezius pull up on the lateral end of the clavicle and the lower fibers pull down on the medial part of the scapular spine (Figure 27.4). The smooth interaction of scapula and shoulder joint movements is referred to as the scapulohumeral rhythm. For every 2 degrees of motion at the glenohumeral joint, there is 1 degree of scapular rotation. After the deltoid abducts the arm to the horizontal, further action by the upper trapezius elevates and rotates the scapula further to allow the arm to be raised overhead.

**TABLE
27.9**

**Major Lower-Extremity Muscles Innervated by
Different Roots**

Parentheses signify minor contribution

Root Muscles Supplied

- | | |
|----|---|
| L2 | Iliopsoas, sartorius, quadriceps (adductors, gracilis) |
| L3 | Iliopsoas, sartorius, adductors, gracilis, quadriceps |
| L4 | Gracilis, gluteus medius, TFL, quadriceps, adductor magnus, TA (iliopsoas, adductor longus) |
| L5 | Gluteus maximus, internal hamstring, biceps femoris, gluteus |

medius, TFL, peronei, TA, EHL, EDL, EDB, TP, FDL, FHL
(adductor magnus)

- S1 Internal hamstring, biceps femoris, gluteus maximus, gastrocnemius, soleus, FDL, FHL, ADMP, AH, EDB, lumbricals (gluteus medius, TFL, peronei, EDL, TP)
- S2 Gluteus maximus, gastrocnemius, soleus, AH, ADMP, interossei, lumbricals (internal hamstring, short head of biceps femoris)
- S3 Interossei, lumbricals, ADMP

ADMP, abductor digiti minimi pedis; AH, abductor hallucis; EDB, extensor digitorum brevis; EDL, extensor digitorum longus; EHL, extensor hallucis longus; FDL, flexor digitorum longus; FHL, flexor hallucis longus; TA, tibialis anterior; TFL, tensor fascia lata; TP, tibialis posterior.



FIGURE 27.1 Examination of flexion of the neck. The patient attempts to flex his neck against resistance; the sternocleidomastoid, platysma, and other flexor muscles can be seen and palpated.

The Scapular Muscles

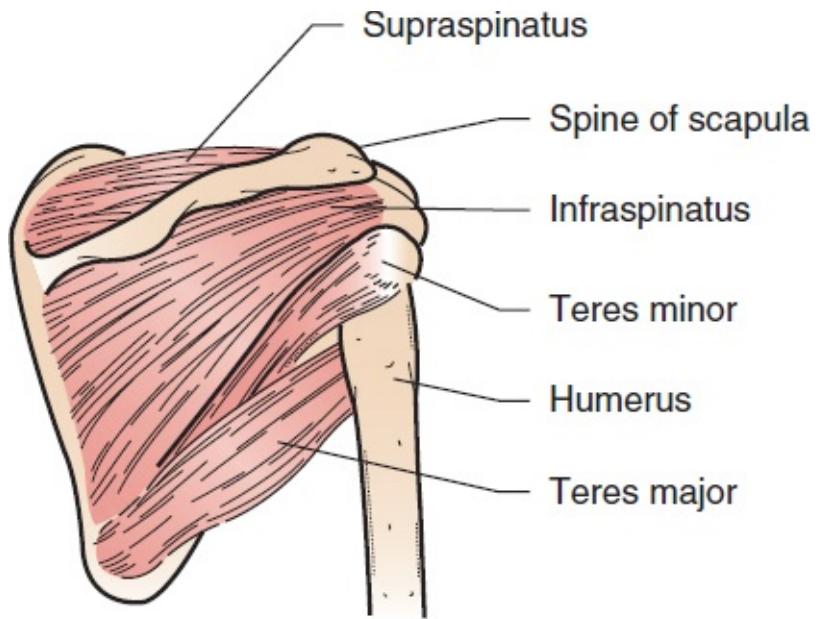
The rhomboids can be tested by having the patient, with hand on hip, retract the shoulder against the examiner's attempt to push the elbow forward ([Figure 27.5](#)). If the patient braces the shoulders backward as if standing at attention, the bulge of the rhomboids can be seen and palpated along the medial border of the

scapula. Another test of rhomboid function is to have the patient place the back of the hand against the small of the back and to push backward with the palm against the examiner's resistance. The rhomboid major contracts vigorously as a downward rotator of the scapula. Lifting the hand off the small of the back is also used to test the subscapularis. The levator scapulae is tested by observing elevation of the scapula; it is rarely possible to detect weakness of the levator scapulae on clinical examination.

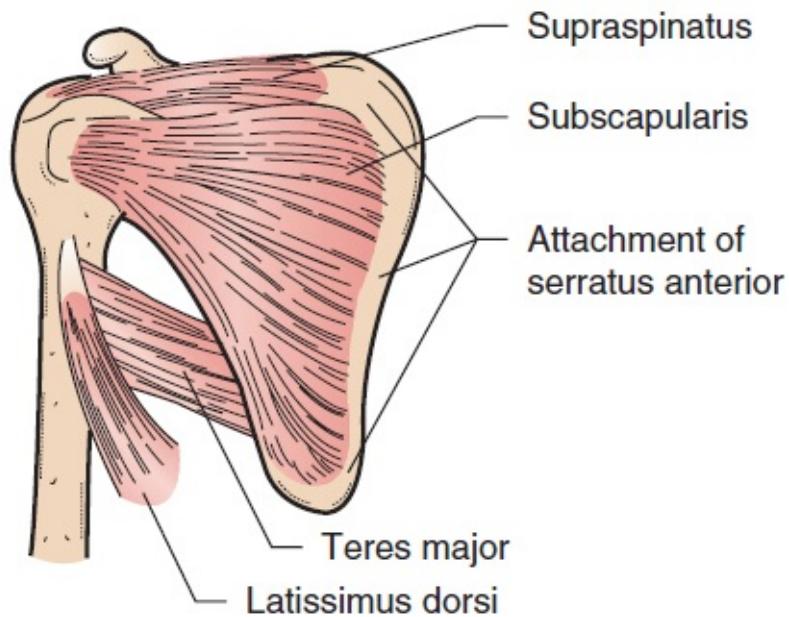


FIGURE 27.2 Examination of extension of the neck. The patient attempts to extend his neck against resistance; contraction of the trapezius and other extensor muscles can be seen and felt, and strength of movement can be judged.

The different parts of the trapezius must be tested separately. One test of the upper fibers is to have the patient shrug the shoulders against resistance ([Figure 19.4](#)). A better test is resisting the patient's attempt to touch the occiput to the acromion. The middle fibers may be tested by having the patient retract the scapula against resistance ([Figure 27.6](#)) or having the patient hold the arm horizontally abducted, palm up, and attempting to push the elbow forward.



Posterior View



Anterior View

FIGURE 27.3 The muscles of the scapula.

With unilateral trapezius paralysis, the patient cannot retract the shoulder or abduct the arm above horizontal. Because of the weight of the arm, the upper portion of the scapula falls laterally, the inferior angle moves medially, and the vertebral border is flared. The sagging of the shoulder causes a drooping of the

entire arm, and the fingertips on the involved side are at a lower level than on the normal side. With trapezius atrophy, the superior angle of the scapula may bulge beneath the skin. There is little loss of shoulder-shrug ability because the levator scapulae and rhomboids are able to elevate the scapula, but the normal slant from the base of the neck to the shoulder becomes squared off because of loss of trapezius bulk. The patient may be able to elevate the arms forward with little or no difficulty because the serratus anterior is primarily responsible for scapular fixation and rotation in that plane.

The serratus anterior can be tested by having the patient make movements that involve forward reaching or pushing and observing for evidence of scapular winging (see next section). When there is significant weakness, abnormalities may appear when the patient simply tries to raise the arm overhead. More subtle degrees of weakness may be brought out by having the patient push forward against resistance. The classical test is to have the patient push against a wall, comparing how well the scapulae remain against the chest wall on the two sides ([Figure 27.7](#)).

Winging of the Scapula

Normally, the medial border of the scapula remains close to the chest wall when the arms are raised. However, with weakness of either the serratus anterior or the trapezius, the vertebral border or the entire scapula protrudes posteriorly, away from the thoracic wall. This causes the deformity known as “winging” ([Figure 27.8](#)). The trapezius is a rotator and retractor of the scapula and functions primarily during abduction of the arm to the side in the coronal plane of the body. When the trapezius is weak, scapular winging is more apparent on attempted abduction of the arm than on forward elevation. Trapezius winging may be made more conspicuous by having the patient bend forward at the waist so the upper body is parallel to the ground and then raise the arms to the sides, as if beginning a swan dive. This requires strong action by the trapezius to retract the scapula and accentuates the posterior displacement of the shoulder girdle.

The serratus anterior is primarily a protractor of the scapula and functions during forward arm elevation. When the serratus is weak, the inferior angle is shifted medially and the entire vertebral border rides up from the chest wall. Serratus anterior weakness causes winging that is more obvious when trying to elevate the arm in front, in the sagittal plane of the body; it is less obvious when the arms are abducted to the sides. This difference aids in differentiating serratus

anterior winging (as from a long thoracic nerve palsy) from the flaring of the scapula that occurs with trapezius weakness (as from a spinal accessory nerve palsy). Serratus winging may be accentuated by having the patient protract the scapula against resistance ([Figure 27.7](#)). Another method to bring out mild serratus winging is to have the patient slowly lower the outstretched arms. This downward movement may exacerbate the winging, and at a certain point as the arms descend the scapula will suddenly snap backward. Scapular winging is also discussed in [Chapter 19](#). For a video of scapular winging, see [Video Link 27.1](#).

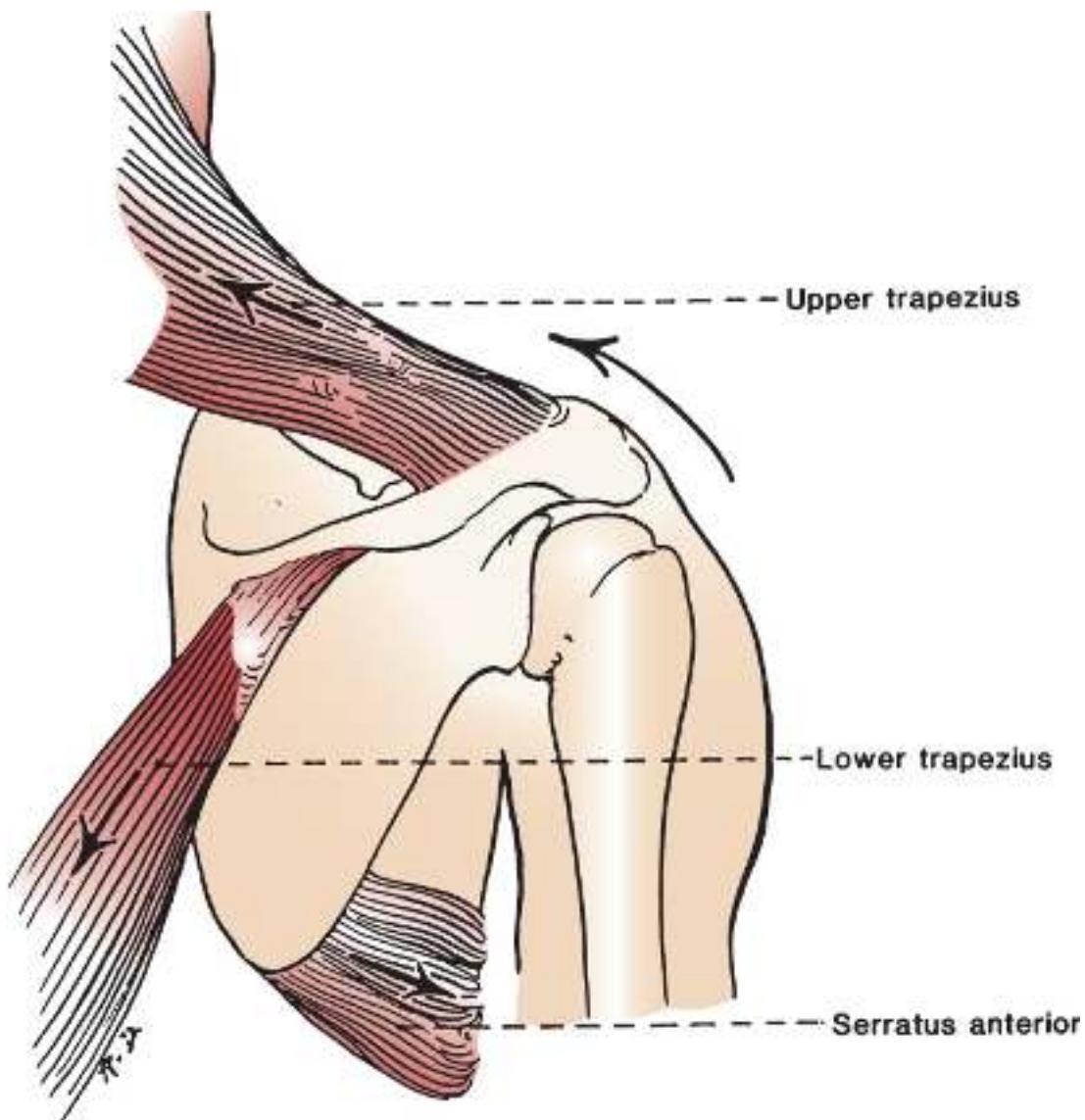


FIGURE 27.4 Upward rotators of the scapula. (Modified from Weibers DO, Dale AJD, Kokmen E, et al., eds. *Mayo Clinic Examinations in Neurology*. 7th ed. St. Louis: Mosby, 1998. Copyright © 1998 Elsevier. With permission.)

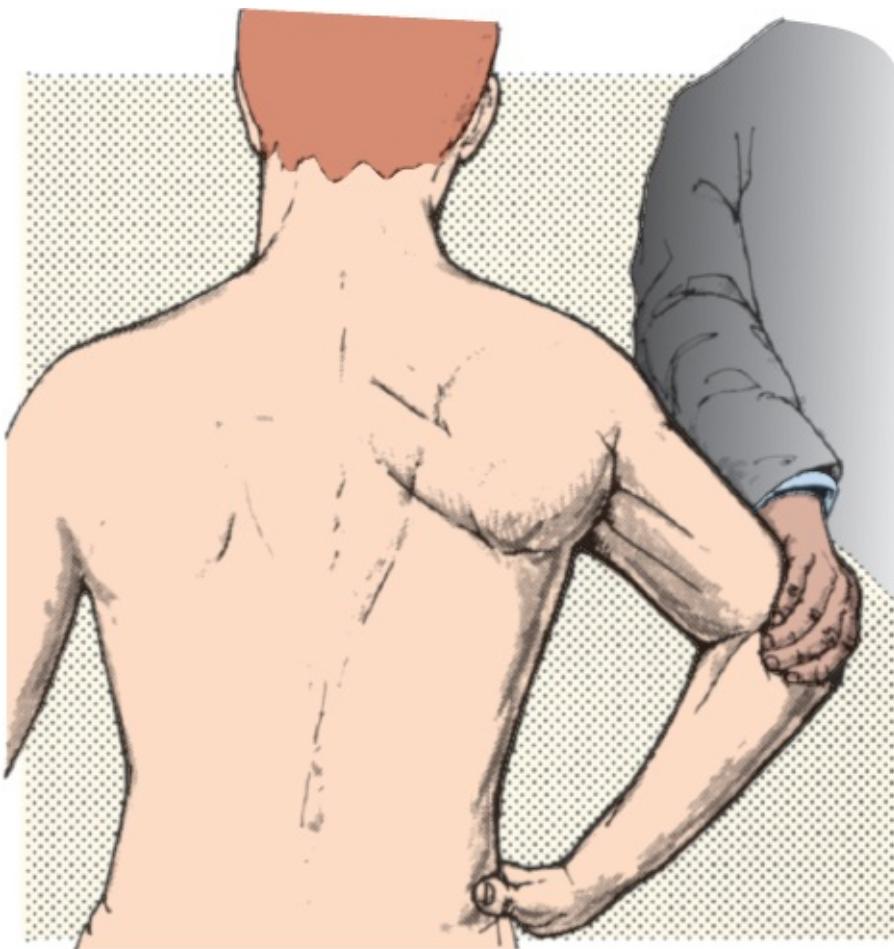


FIGURE 27.5 Examination of the rhomboids. With hand on hip, the patient retracts the shoulder against the examiner's effort to push the elbow forward; the contracting muscles can be seen and palpated.

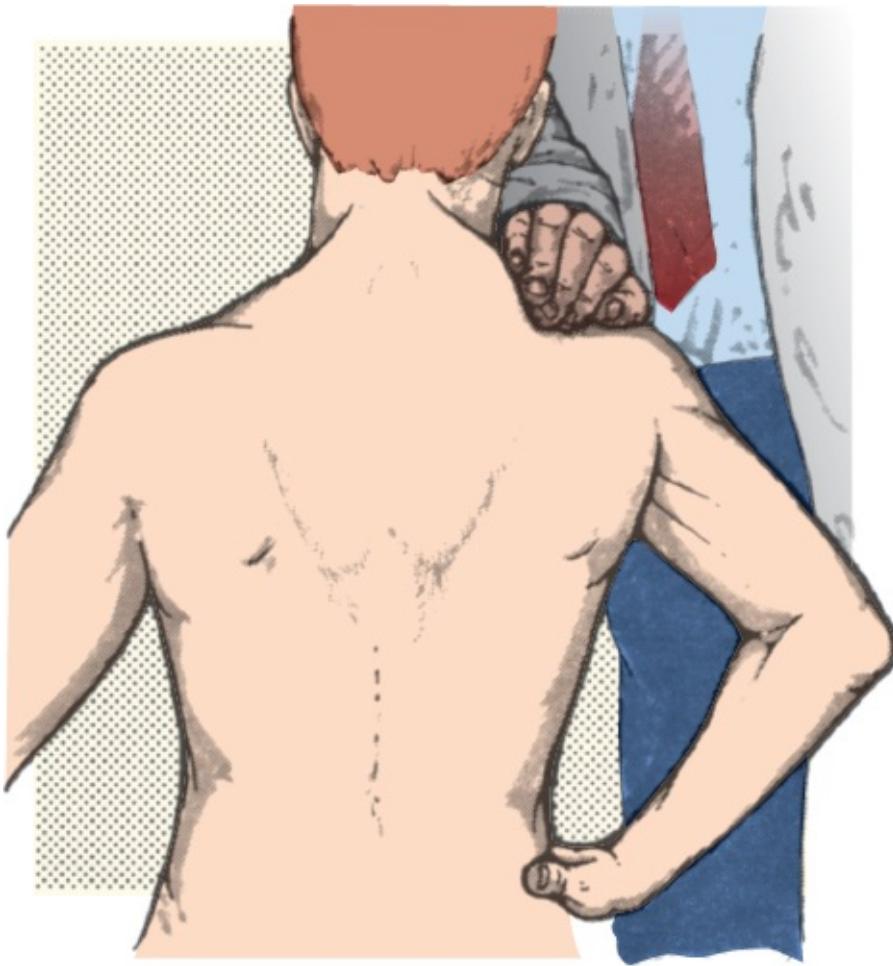


FIGURE 27.6 Examination of the trapezius. On retraction of the shoulder against resistance, the middle fibers of the muscle can be seen and palpated.

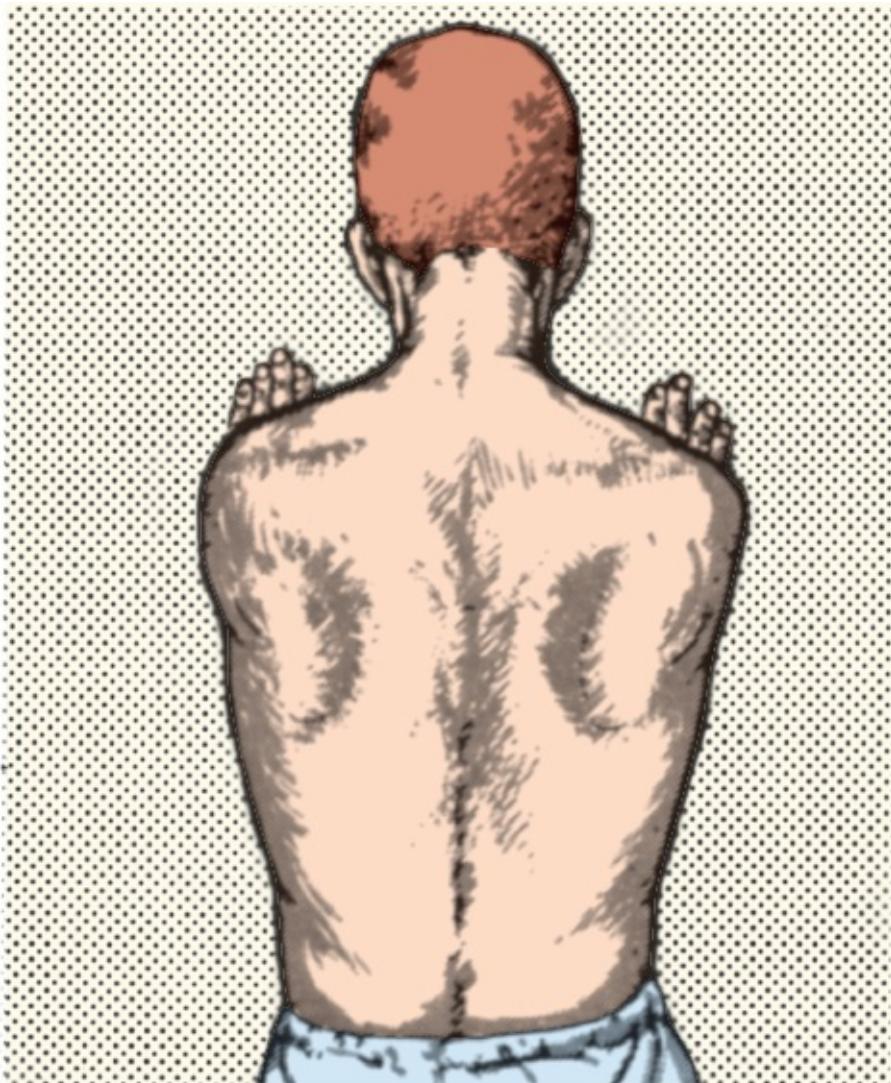


FIGURE 27.7 Examination of the serratus anterior. The patient pushes against a wall with his arms extended horizontally in front of him; normally, the medial border of the scapula remains close to the thoracic wall.

In the muscular dystrophies, particularly facioscapulohumeral (FSH) dystrophy, there is often weakness of all the shoulder girdle muscles, with prominent scapular winging, typically bilateral ([Figures 27.8](#) and [29.6](#)). Another manifestation of shoulder girdle weakness, seen most often in patients with myopathy, is a change in the position of the arms. Normally, as the arms hang at the sides, the thumb faces to the front. With shoulder girdle weakness, the scapula tends to slip laterally so that even at rest the shoulders tend to turn slightly anteriorly. This causes the entire arm to rotate internally, making the back of the hand rather than the thumb face anteriorly. Another effect, especially if there is also pectoral muscle atrophy, may be to produce a crease running

diagonally from the anterior axillary fold toward the neck ([Video Link 27.2](#)).

The Glenohumeral Joint

The principal movements at the glenohumeral joint are abduction, adduction, external and internal rotation, flexion, extension, and elevation of the arm. These movements are best appreciated as taking place in the plane of the body of the scapula rather than in the body as a whole.



FIGURE 27.8 Scapular winging in a patient with facioscapulohumeral muscular dystrophy. **A.** With the arms at rest. **B.** With the arms raised.

The deltoid is the most prominent muscle in the shoulder region. It is supplied by C5 and C6 through the axillary nerve, a branch of the posterior cord of the brachial plexus. The deltoid has three portions: anterior, middle, and posterior. The middle deltoid and supraspinatus muscles, aided by the subscapularis and the upper part of the infraspinatus, abduct the shoulder. With deltoid contraction, the arm is abducted (raised laterally) to the horizontal plane. Further abduction, or elevation above the horizontal plane, is carried out by the associated action of the trapezius and the serratus anterior, which rotate the scapula and tilt the angle of the glenoid fossa upward. In the first 15 degrees, the abduction motion by the deltoid is aided by the supraspinatus, and the synergistic actions of the subscapularis, infraspinatus, and teres minor prevent the humeral head from translating upward. The posterior fibers of the deltoid also assist in extension

and external rotation of the arm and the anterior fibers in flexion and internal rotation; but electromyography shows the deltoid is not very active in these movements.

The major function of the deltoid is tested by noting the ability of the patient either to abduct the arm through the range up to 90 degrees against resistance ([Figure 27.9](#)) or to hold the arm in abduction to the horizontal level, either laterally or forward (the elbow may be either flexed or extended) and to resist the examiner's attempt to push it down. Testing both sides simultaneously helps the patient maintain balance and also helps in the comparison of strength on the two sides. With MRC grade 3/5 weakness, the patient can abduct the arm against gravity but not against significant resistance. With more severe weakness, the patient may lean in the opposite direction and raise the tip of the shoulder to aid in the attempt (trick movement). When active elevation to the horizontal plane is impossible, the passively abducted arm may be held up against gravity. With grade 2/5 weakness, the patient may be able to abduct the arm when lying flat but not when erect. In complete paralysis, no contraction of the muscle is possible. When weakness of the deltoid is due to a lesion of the anterior horn cells, brachial plexus, or axillary nerve, atrophy appears promptly and may be severe. This leaves the bulge of the acromion visible through the atrophic muscle belly, which simulates the appearance of shoulder dislocation. Because of myotomal overlap, isolated cervical root lesions do not cause the same degree of atrophy. In ALS or cervical spondylosis, fasciculations are often noted.

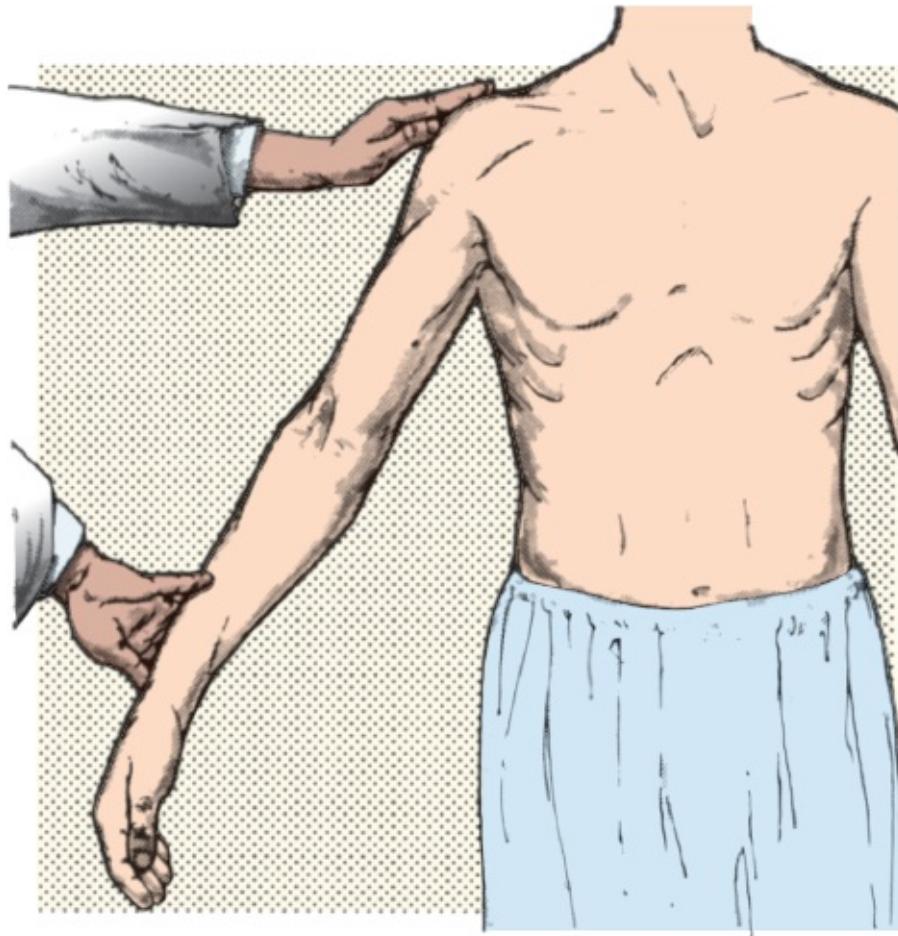


FIGURE 27.9 Examination of the deltoid. The patient attempts to abduct his arm against resistance; the contracting deltoid can be seen and palpated.

The supraspinatus helps abduct the shoulder through the first 15 degrees. The muscle belly lies in the supraspinous fossa of the scapula; its contraction can be palpated and sometimes seen when the arm is abducted less than 15 degrees against resistance (Figure 27.10). The supraspinatus is innervated primarily by C5 and C6 and by the suprascapular nerve, which arises directly from the upper trunk of the brachial plexus. Its tendon crosses over the shoulder joint to attach to the greater tuberosity of the humerus, forming one component of the rotator cuff (Chapter 47).

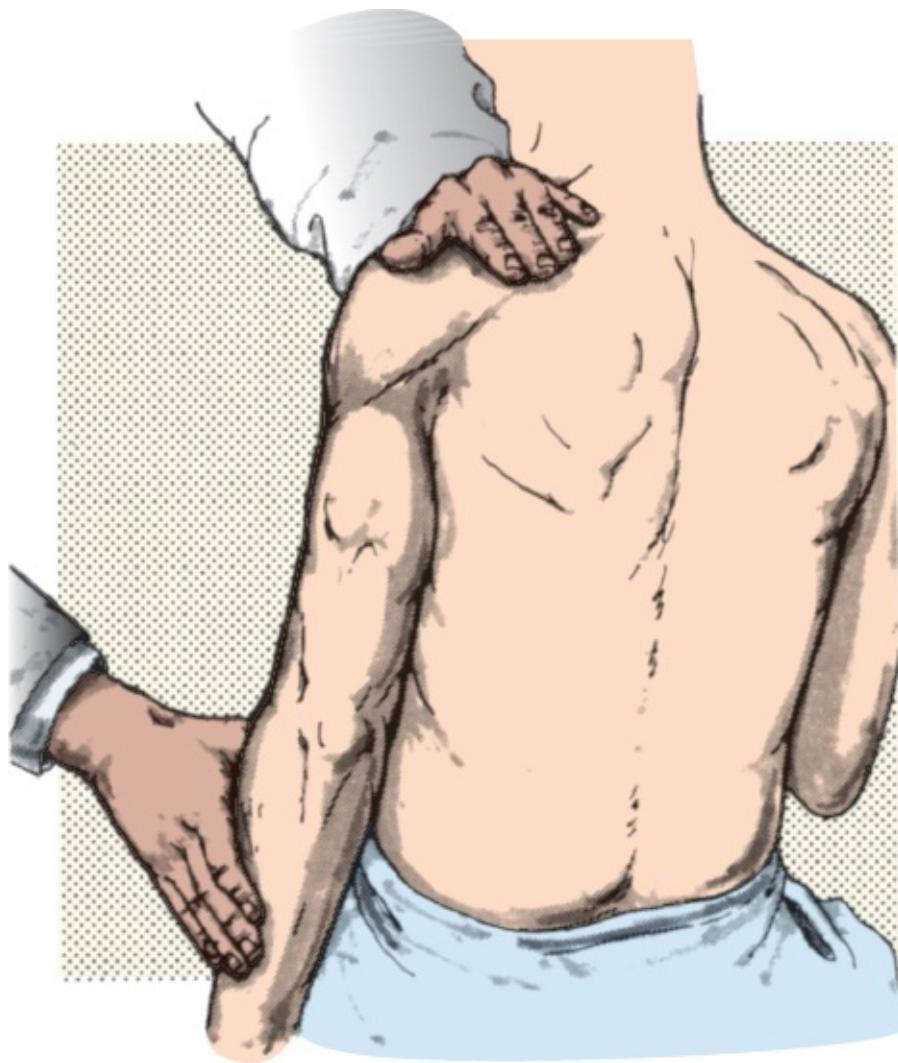


FIGURE 27.10 Examination of the supraspinatus. Contraction of the muscle fibers can be felt during early stages of abduction of the arm.

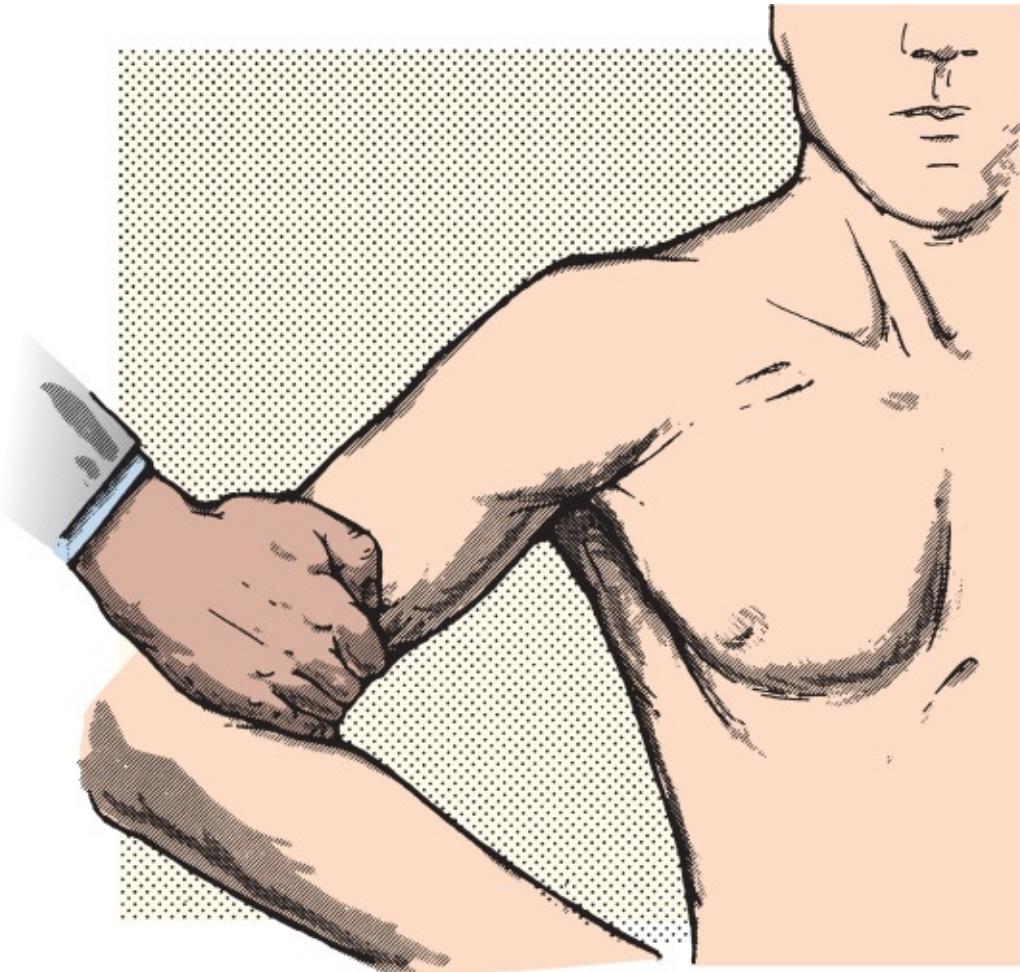


FIGURE 27.11 Examination of the pectoralis major. Contraction of the muscle can be seen and felt during attempts to adduct the arm against resistance.

The primary adductors of the shoulder are the pectoralis major and latissimus dorsi. Their adduction actions are difficult to separate. The pectoralis major receives innervation from all levels of the brachial plexus, C5-T1, through the medial and lateral pectoral nerves. It is the principal shoulder adductor and is also a flexor and internal rotator. When the arm is fixed, the muscle draws the chest upward, as in climbing. On attempts to adduct the horizontally abducted arm against resistance, the contraction of the sternocostal and clavicular portions of the muscle can be seen and felt (Figure 27.11). The muscle can also be tested and palpated by having the patient move the horizontally abducted arm forward, try to press the hands together with the arms in front, or try to internally rotate the forearms with the elbows at the side and flexed, in a position as if holding a book.

The latissimus dorsi is supplied by C6-C8 through the thoracodorsal nerve off

the posterior cord of the brachial plexus. It adducts, extends, and medially rotates the shoulder and may be tested in various ways. The muscle acts when the patient tries to adduct the raised arm against resistance ([Figure 27.12](#)). Along with the teres major, the latissimus forms the posterior axillary fold. The fold becomes prominent when the arm is adducted against resistance and the muscle belly can be easily seen and palpated. The muscle belly can also be felt when the patient coughs or pushes the arm downward and backward. When the humerus is fixed, the latissimus draws the pelvis and the lower part of the trunk forward and upward. When the arm is hanging by the side, it depresses, retracts, and rotates the scapula.

External rotation of the shoulder is carried out principally by the infraspinatus with a minor contribution by the teres minor (axillary nerve, C5-C6). The infraspinatus is innervated by the suprascapular nerve, C5 and C6, which also innervates the supraspinatus. To test these muscles, the patient attempts to externally rotate the shoulder by turning the forearm laterally and backward against resistance while the elbow is flexed at an angle of 90 degrees and held at the side ([Figure 27.13](#)).

Internal rotation at the shoulder chiefly results from contraction of the subscapularis and teres major muscles. These muscles are tested by having the patient move the forearm medially against resistance with the elbow flexed and at the side—the opposite motion from external rotation. Internal rotation can also be tested by having the patient lift the back of the hand off the small of the back against resistance, as is done when testing the rhomboids (see “The Scapular Muscles” above).

Flexion of the shoulder (forward elevation of the arm in the sagittal plane of the body) is carried out by the anterior fibers of the deltoid and the pectoralis major, subscapularis, coracobrachialis, and biceps muscles. The muscles, especially the deltoid and pectoralis, may be palpated when raising the arm forward against resistance. Extension of the shoulder (backward elevation of the arm) is carried out by the posterior fibers of the deltoid together with the latissimus dorsi, triceps, subscapularis, and teres major muscles. This action can be tested by having the patient attempt to extend the arm against resistance. The individual muscles concerned are better examined by the tests discussed in the preceding paragraphs.

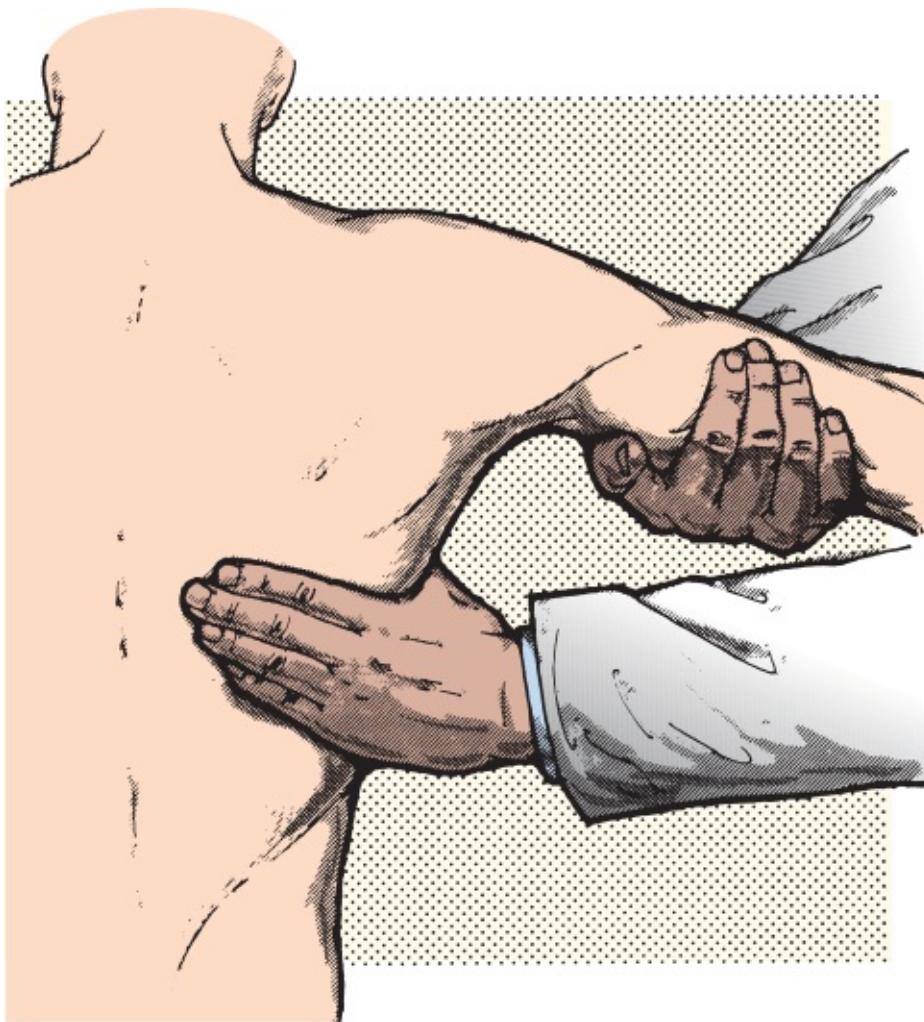


FIGURE 27.12 Examination of the latissimus dorsi. On adduction of the horizontally and laterally abducted arm against resistance, the contracting muscle fibers can be seen and palpated. An effective way to compare the latissimus contraction strength on the two sides is to firmly grasp the muscle bellies from behind and have the patient cough.



FIGURE 27.13 Examination of the external rotators of the arm. On external rotation of the arm while the elbow is flexed and kept close to the body, the contracting infraspinatus muscle can be seen and palpated.

The Rotator Cuff

The subscapularis, supraspinatus, infraspinatus, and teres minor form the rotator cuff. The infraspinatus and teres minor rotate the humerus externally and the subscapularis internally. The subscapularis, infraspinatus, and teres minor help keep the humeral head from sliding upward when the deltoid contracts during the initial stages of abduction. The supraspinatus, more than the deltoid, keeps the humeral head from migrating downward when the arm is hanging down. Rotator cuff tears are a common clinical problem. Rotator cuff pathology often enters the differential diagnosis in patients with arm pain and weakness. A torn

rotator cuff, especially impaired arm abduction due to a ruptured supraspinatus muscle or tendon (the most common component of the rotator cuff to suffer a tear), can be confused with a neurologic process ([Chapter 47](#)).

The Elbow

The principal movements at the elbow are flexion and extension of the forearm at the elbow joint and pronation and supination at the radioulnar joint.

Many muscles contribute to elbow flexion; the primary ones are the biceps brachii, brachialis, and brachioradialis. Which muscle is the prime mover depends on the position of the forearm. The biceps muscle is innervated by C5-C6 through the musculocutaneous nerve, a branch of the lateral cord of the brachial plexus. It is an elbow flexor and also a strong supinator of the forearm. Its supination power is greatest when the forearm is flexed and pronated. Its flexion power is greatest when the forearm is supinated. The brachialis has the same innervation; it flexes the elbow regardless of forearm position. The musculocutaneous nerve passes through the coracobrachialis and may be compressed there, causing weakness of the biceps and brachialis muscles. The brachioradialis is innervated by C5-C6 through the radial nerve. It acts as an elbow flexor when the forearm is held midway between pronation and supination (thumb up). The brachioradialis acts as a supinator when the forearm is extended and pronated but as a pronator when the forearm is flexed and supinated.

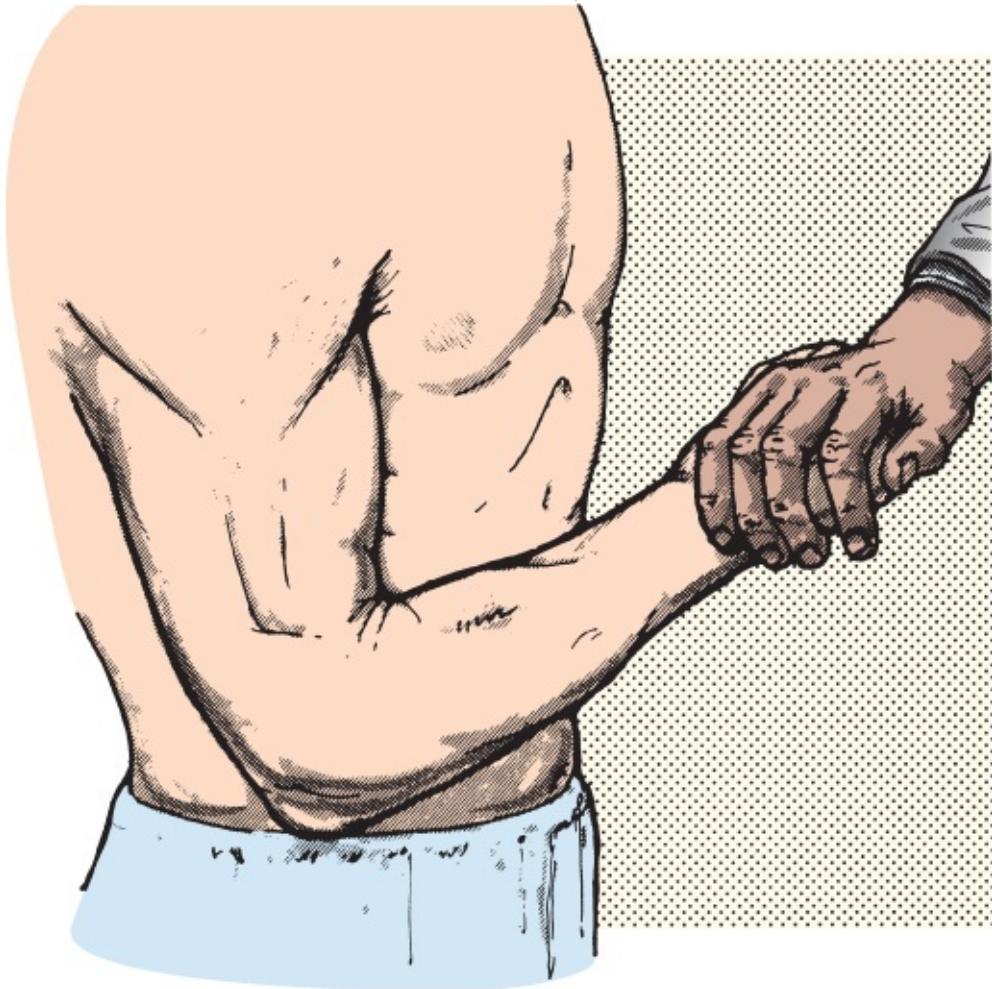


FIGURE 27.14 Examination of the biceps brachii. On attempts to flex the forearm against resistance, the contracting biceps muscle can be seen and palpated.

Biceps and brachialis functions are tested by having the patient attempt to flex the elbow against resistance. The biceps contraction can be seen and felt, but the brachialis is buried (Figure 27.14). The brachioradialis is tested by attempts to flex the semipronated forearm (Figure 27.15). Even when the biceps and brachialis are completely paralyzed, the brachioradialis is still capable of flexing the elbow to some degree. When the biceps muscle is weak, the patient may employ trick movements by putting the forearm into midpronation and bringing in the brachioradialis or pulling the elbow backwards. The latter resembles the movement bartenders make when drawing a draft beer and has been called the “bartender’s sign.”

The triceps brachii is the principal elbow extensor. It is innervated by C6, C7, and C8 through the radial nerve, a branch of the posterior cord of the brachial

plexus. The branches to the triceps come off before the nerve enters the spiral groove. The anconeus aids the triceps in extension. To test these muscles, place the elbow in a position midway between flexion and extension and have the patient attempt to either extend the elbow or to hold position against the examiner's resistance ([Figure 27.16](#)). The triceps muscle is less powerful when the elbow is fully flexed, and slight weakness may be more easily detected with testing in this position. With mild triceps weakness, the examiner may be able to pin the triceps in extreme flexion on the involved but not the normal side.

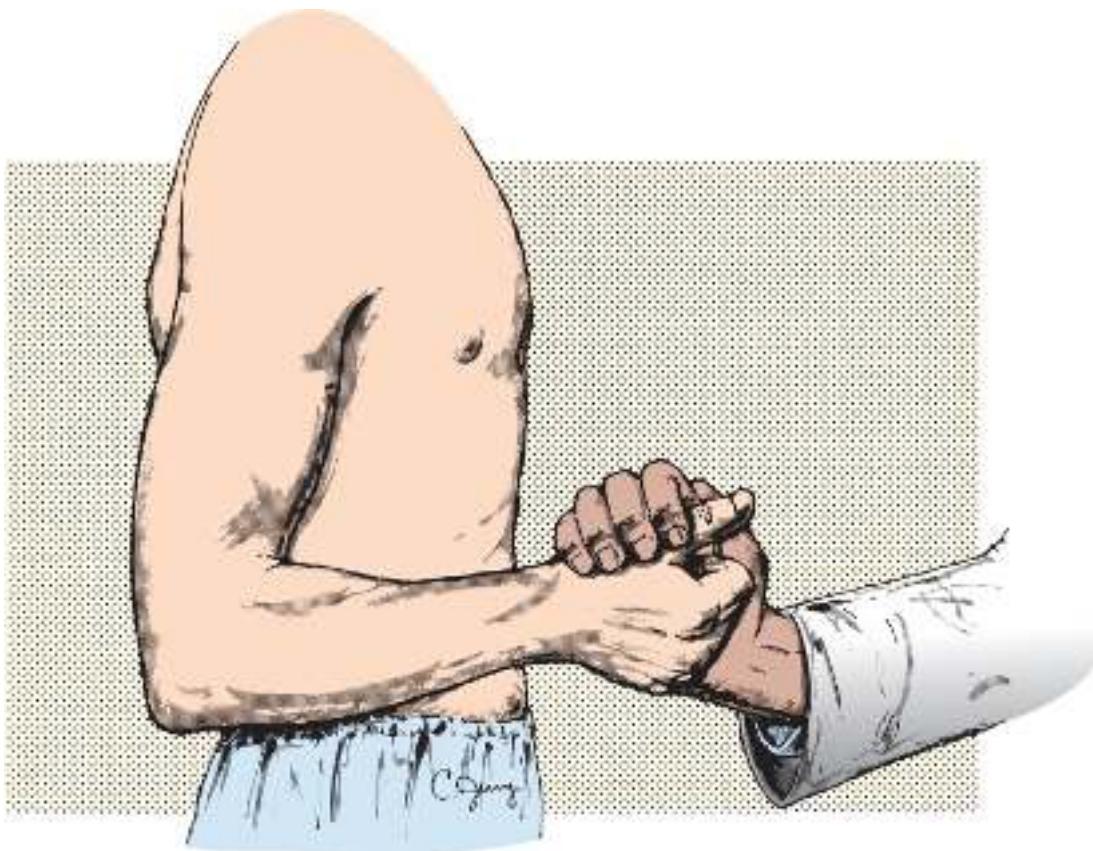


FIGURE 27.15 Examination of the brachioradialis. On flexion of the semipronated forearm (thumb up) against resistance, the contracting muscle can be seen and palpated.

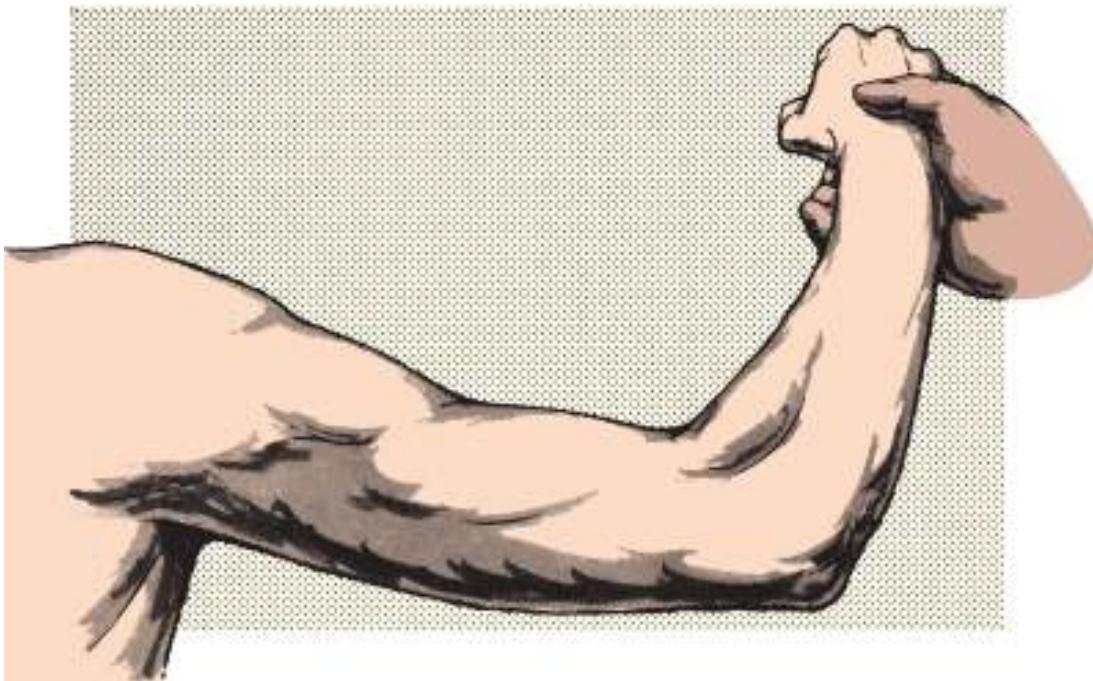


FIGURE 27.16 Extension of the forearm. On attempts to extend the partially flexed forearm against resistance, contraction of the triceps can be seen and palpated.

Supination of the forearm is done primarily by the supinator muscle, assisted by stronger muscles, especially the biceps, for movements requiring power.

The supinator has superficial and deep layers; the proximal edge of the superficial head is the arcade of Frohse. The posterior interosseous nerve passes beneath the arcade and may be compressed there. The supinator is innervated by C6 and C7, either by the posterior interosseous nerve or the radial in different individuals. The biceps muscle is the most powerful forearm supinator; its action is strongest when the forearm is flexed and pronated. The supinator is less powerful, but it acts through all degrees of flexion and supination. Supination is tested by having the patient supinate against the examiner's resistance. With the forearm in extension, the brachioradialis also participates; with the forearm in flexion, the biceps also participates ([Figure 27.17](#)).

Pronation is brought about primarily by the pronator quadratus (PQ), which is assisted by the much stronger PT for movements requiring power. Other muscles may play a minor role. The PT originates from the common flexor tendon (CFT) that arises from the medial epicondyle. Like the other muscles that arise from the CFT (flexor carpi radialis [FCR], palmaris longus, flexor carpi ulnaris [FCU], and flexor digitorum superficialis [FDS]), the PT is also an elbow flexor. To test the PT and PQ, the patient attempts to pronate against resistance ([Figure 27.18](#)).

The PT has a humeral and an ulnar head. The median nerve usually enters the forearm between the two heads and supplies the muscle (C6 and C7). The PQ is innervated by the anterior interosseous branch of the median nerve (C7-C8). The anterior interosseous nerve comes off the median nerve between the two heads of the PT. The anterior interosseous nerve, less often the main trunk of the median nerve, may be compressed between the two heads of the PT (pronator syndrome). To isolate the action of the PQ, pronation should be tested with the elbow extended, when the PT is maximally lengthened and exerts its weakest pull. Flexion of the elbow would signal that the patient is trying to bring the PT into play.

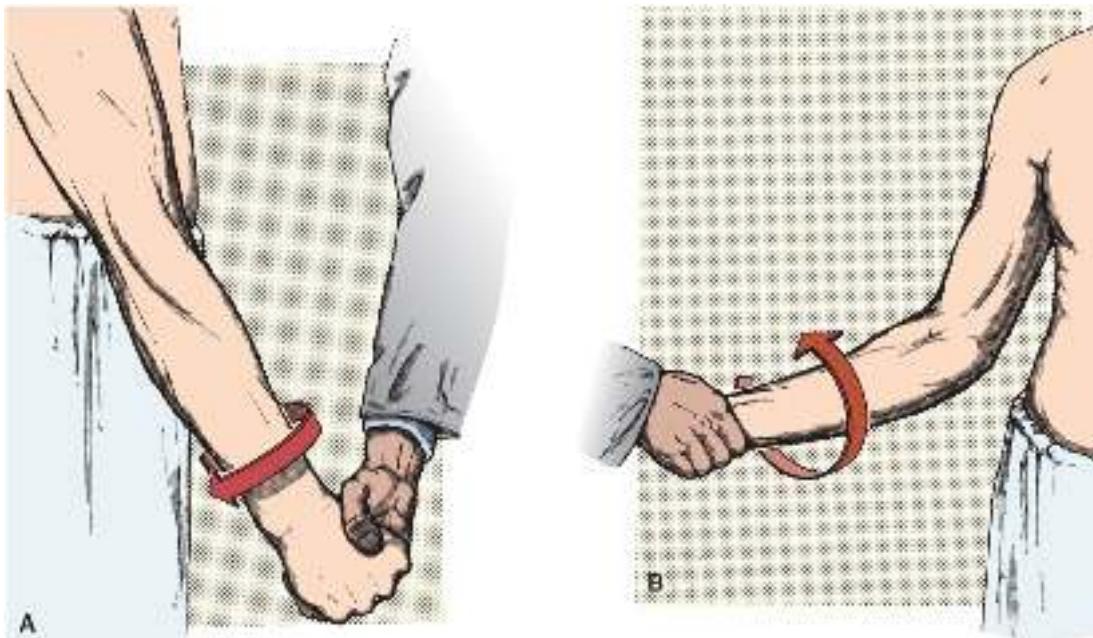


FIGURE 27.17 Supination of the forearm. **A.** On attempts to supinate the extended forearm against resistance, the contracting brachioradialis can be seen and palpated. **B.** On attempts to supinate the flexed forearm against resistance, the contracting biceps can be seen and palpated.

The Wrist

The principal movements at the wrist are flexion and extension; adduction (ulnar flexion) and abduction (radial flexion) are minor movements.

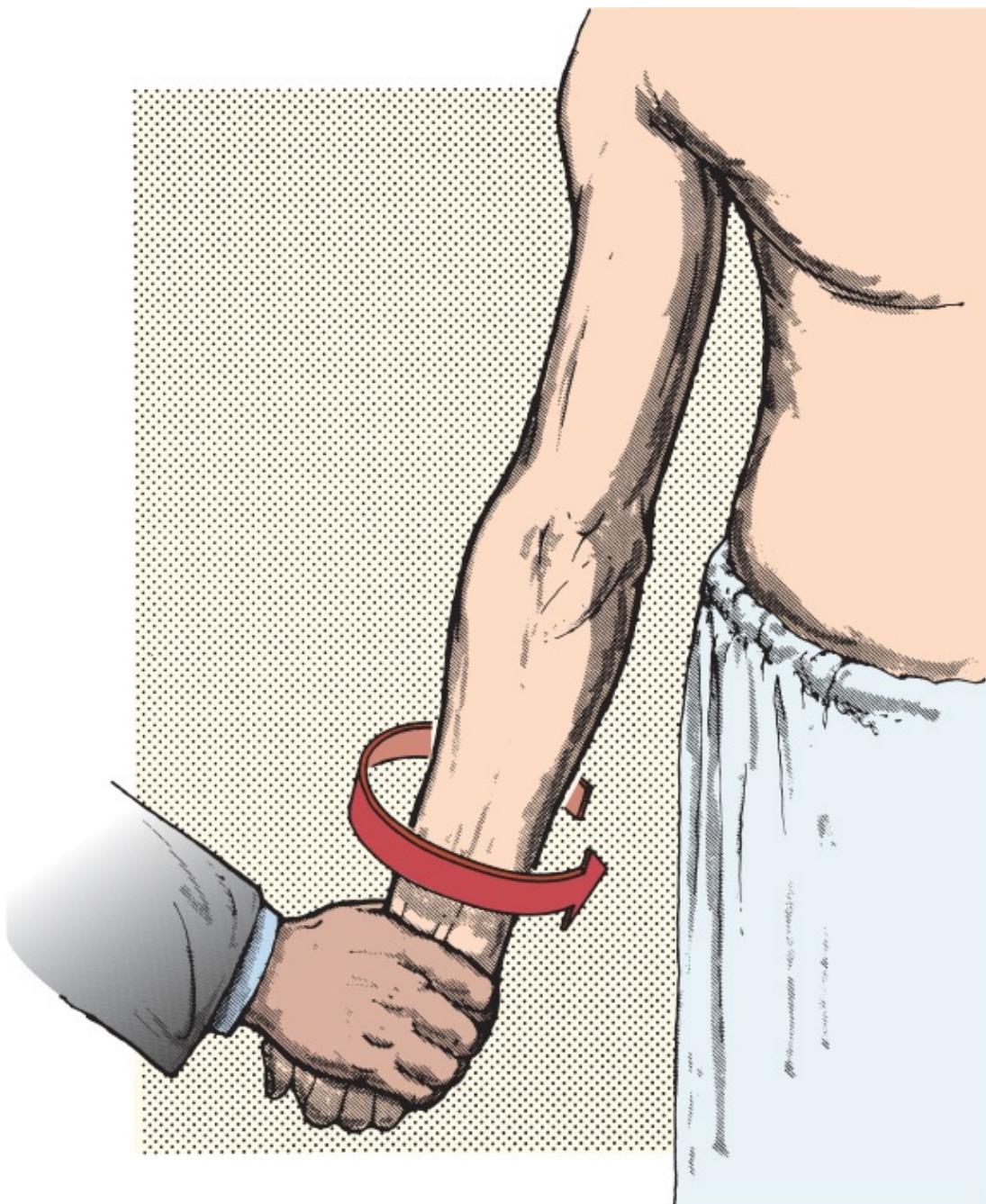


FIGURE 27.18 Pronation of the forearm. On pronation of the forearm against resistance, contraction of the pronator teres can be seen and palpated.

Flexion of the wrist is carried out principally by the FCR and FCU muscles. The FCR originates from the CFT and is innervated by the lateral head of the median nerve (C6-C7). When making a fist, the wrist usually extends slightly as the fingers flex; the FCR counteracts the wrist extension motion. The FCR also assists in elbow flexion and in forearm pronation. It inserts on the second

metacarpal and is a weak abductor of the wrist. The FCU is supplied by the ulnar nerve (C7-T1). It has two heads of origin: from the CFT on the medial epicondyle of the humerus and from the olecranon process of the ulna. The two heads are joined by an aponeurosis, the humeroulnar arcade (Osborne's band). After passing through the retroepicondylar groove, the ulnar nerve passes beneath the aponeurosis and may be compressed by it. The FCU attaches to the pisiform bone on the medial aspect of the wrist. In addition to flexing the wrist, it also works with the extensor carpi ulnaris (ECU) to adduct (ulnar deviate) the hand. Other muscles that aid in wrist flexion include the palmaris longus, flexor digitorum profundus (FDP), FDS, flexor pollicis longus (FPL), and abductor pollicis longus (APL).

Wrist flexion is tested by having the patient resist the examiner's attempts to extend the wrist ([Figure 27.19](#)). Both the FCR and FCU are superficial; their contraction can be seen and felt. On resisted wrist flexion, the FCR and palmaris longus tendons stand out on the volar wrist surface; the median nerve lies between the two tendons. The FCR can be tested individually by having the patient flex the wrist toward the radial side against resistance directed toward the thumb. Function of the FCU can be tested by having the patient flex the wrist toward the ulnar side while the examiner presses on the hypothenar region. The FCU also acts as a synergist for the abductor digiti minimi (ADM), stabilizing the pisiform bone; its contraction can be seen and felt during resisted small finger abduction.

Extension (dorsiflexion) of the wrist is executed primarily by the extensor carpi radialis longus (ECRL), extensor carpi radialis brevis (ECRB), and ECU. The extensors of the digits play a minor supportive role. The ECRL (radial nerve, C6 and C7) is the most powerful wrist extensor. The ECRB and ECU are innervated by the posterior interosseous branch of the radial nerve (C7-C8). In a posterior interosseous neuropathy, the wrist deviates radially on extension because of the unbalanced pull of the ECRL.

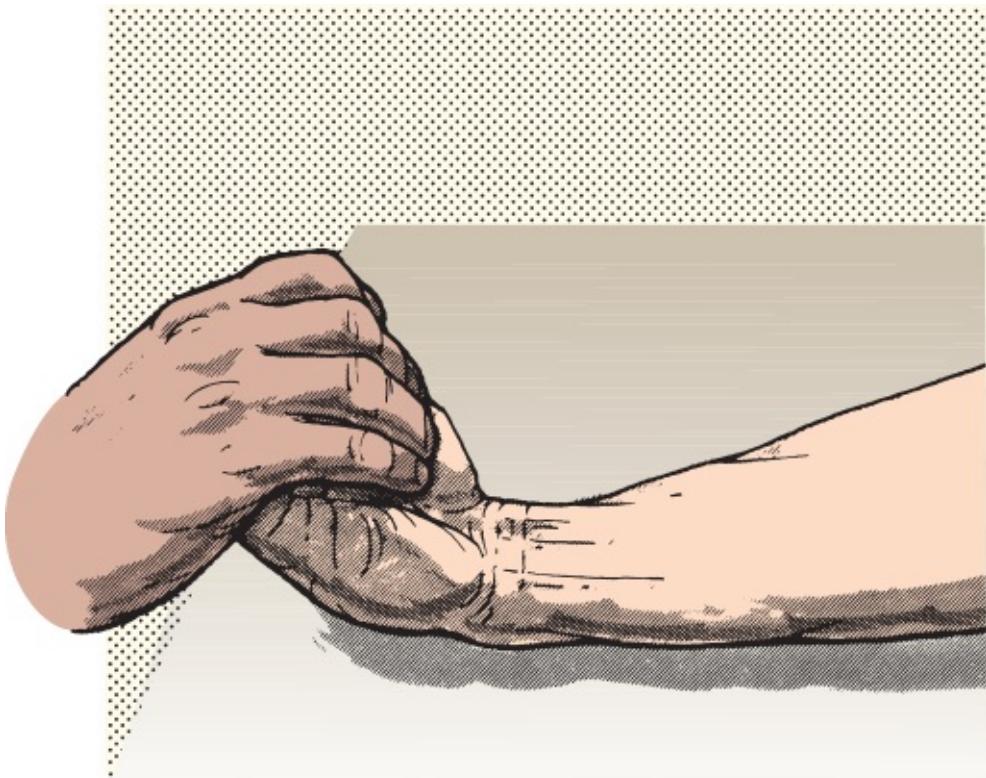


FIGURE 27.19 Flexion at the wrist. On flexion of the hand at the wrist against resistance, the tendon of the flexor carpi radialis can be seen and palpated on the radial side of the wrist and that of the flexor carpi ulnaris on the ulnar side; the tendon of the palmaris longus can also be seen and palpated.

To test the wrist extensors, the forearm is held in pronation with the wrist partially extended. The patient then resists the examiner's attempts to pull the wrist into flexion ([Figure 27.20](#)). With mild weakness, the examiner may be able to hold the wrist in extreme flexion using one or two fingers against the patient's efforts to extend it on the involved but not the normal side. Moderate weakness of the extensors results in involuntary flexion at the wrist when the patient attempts to make a fist; marked weakness causes a wrist drop, the major finding in a radial nerve palsy.

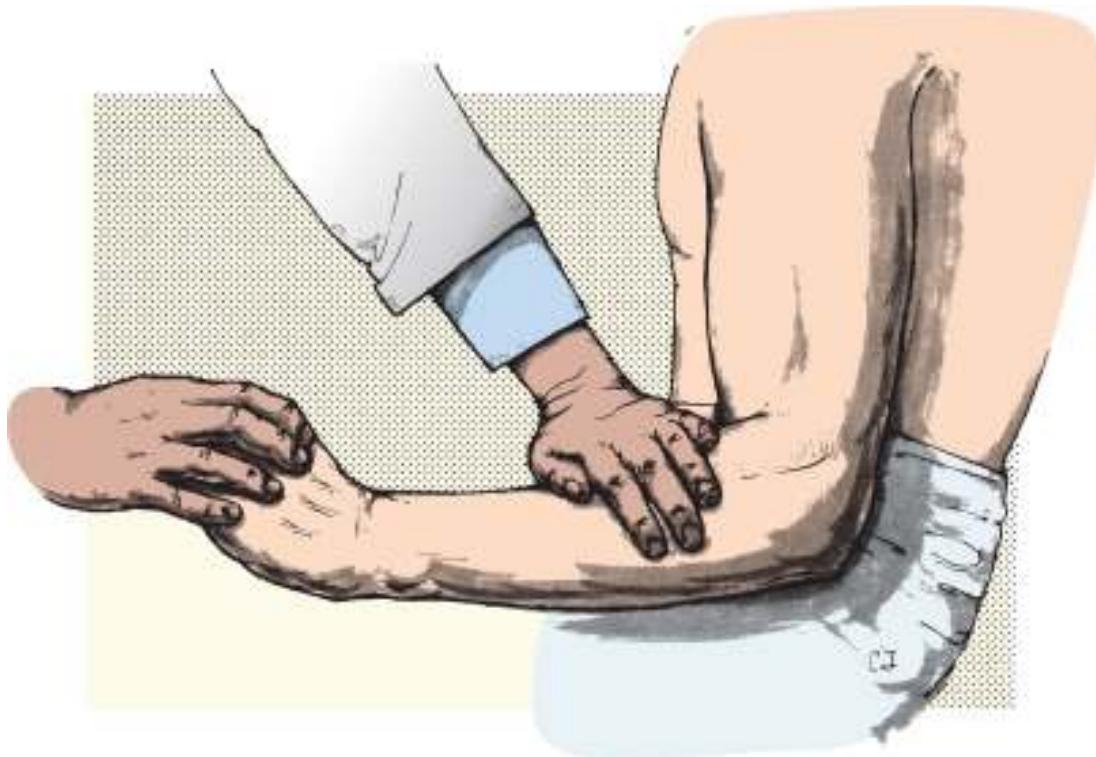


FIGURE 27.20 Extension at the wrist. On attempts to extend the hand at the wrist against resistance, the bellies of the extensors carpi radialis longus, carpi ulnaris, and digitorum communis can be seen and palpated.

Adduction, or ulnar deviation or flexion, of the wrist is carried out principally by the FCU and ECU; abduction, or radial deviation or flexion, is carried out by the FCR, ECRL, and ECRB. Other muscles may make minor contributions. These movements, too, may be tested by carrying them out against resistance.

The Hands and Fingers

Examination of the hand and finger muscles is difficult. Innervation is complex, and the numerous possible substitution movements can lead to misinterpretation. Possible movements include flexion, extension, adduction, abduction, and opposition. The muscles that power the hand can be divided into extrinsics and intrinsics. The extrinsic muscles originate in the forearm and insert on hand structures; the intrinsics originate and insert within the hand.

Flexion of the Fingers

The primary finger flexors are the FDS and the FDP. The FDS and FDP are both

innervated by C8 and T1. The FDS is supplied by the main trunk of the median nerve. Its tendons pass through the carpal tunnel and then diverge to insert on the palmar surfaces of the middle phalanges. The FDS primarily flexes the proximal interphalangeal (PIP) joints of the four fingers; a continuation of its action flexes the metacarpophalangeal (MCP) joints and ultimately the wrist. There are separate muscle slips for each finger, so the PIP joints can be flexed independently. The FDP has two parts: (a) The lateral or radial head is innervated by the anterior interosseous branch of the median nerve and (b) the medial or ulnar head by the ulnar nerve. The four tendons of the FDP pass through the carpal tunnel and then pierce the tendons of the FDS and insert on bases of the distal phalanges. The main action of the FDP is flexion of the distal interphalangeal (DIP) joints of the fingers; continuing this action flexes the remaining phalanges and finally the wrist. The muscle slip and tendon that flex the index finger are usually distinct; the remaining slips of the FDP are often partially conjoined, making it difficult to flex the other DIP joints independently.

The fingers are flexed at the MCP joints by the interossei and the lumbricals. On the dorsal surface of the proximal phalanx of each finger, there is a dorsal extensor expansion, a fibrous enlargement of the tendon of the extensor digitorum. The finger extensor tendons blend into the expansion. The dorsal interossei lie between the metacarpal bones, from which they originate, and insert on the proximal phalanges. They also insert separately into the extensor expansions and are therefore functionally connected to the finger extensor tendons. Finger adduction and abduction are judged in relation to an imaginary line down the center of the middle finger. From the insertion on the proximal phalanx, the dorsal interossei flex the MCP joint. From their insertion on the extensor expansion, they extend the PIP joint and also abduct the fingers. The smaller palmar interossei arise from the palmar surfaces of the metacarpal bones, rather than between them, and insert on the side of the extensor expansion so as to adduct the finger; they also flex the MCP joint and extend the PIP joint. The interossei are innervated by C8-T1 through the deep palmar branch of the ulnar nerve.

The lumbricals arise from the tendons of the FDP and insert into the extensor expansions on the dorsal surfaces of the phalanges. The two lumbricals on the ulnar side of the hand are innervated by the deep palmar branch of the ulnar nerve, the two on the radial side of the hand by the median nerve, all C8-T1. The lumbricals are weak flexors of the MCP joints. Their more important function is to extend the PIP joints.

The flexor digiti minimi brevis flexes and slightly abducts the proximal phalanx of the little finger. Two other muscles acting on the little finger are the ADM, which abducts the little finger, flexes its proximal phalanx, and extends the middle phalanx, and the opponens digiti minimi, which flexes, adducts, and slightly rotates the fifth metacarpal. All three muscles are supplied by C8-T1 through the ulnar nerve proximal to the origin of the deep palmar branch. The palmaris brevis has the same innervation; it wrinkles the skin over the hypothenar eminence and deepens the hollow of the hand. The palmaris brevis sign is wrinkling of the skin over the hypothenar eminence with small finger abduction in the face of weakness of the ulnar hand intrinsics; it proves the lesion involves the deep palmar branch.

Function of the FDP is tested by having the patient flex the distal phalanges of the individual fingers against resistance while the middle phalanges are fixed ([Figure 27.21](#)). The FDS is tested by having the patient flex the fingers at the PIP joints while the proximal phalanges are fixed ([Figure 27.22](#)). The patient should try to relax the distal phalanges to eliminate any action of the FDP on the PIP joint. The interossei and lumbricals flex the MCP joints and extend the interphalangeal (IP) joints. Weakness of these intrinsic hand muscles causes loss of MCP joint flexion and loss of PIP joint extension, together with loss of adduction and abduction of the fingers. The hand assumes a position of rest in which the MCP joints are held in extension and the PIP and DIP joints are flexed (claw hand). Ulnar neuropathy is the most common cause of claw hand (ulnar griffe). Ulnar clawing primarily affects the ring and small fingers because both lumbrical and interosseous functions are lost.

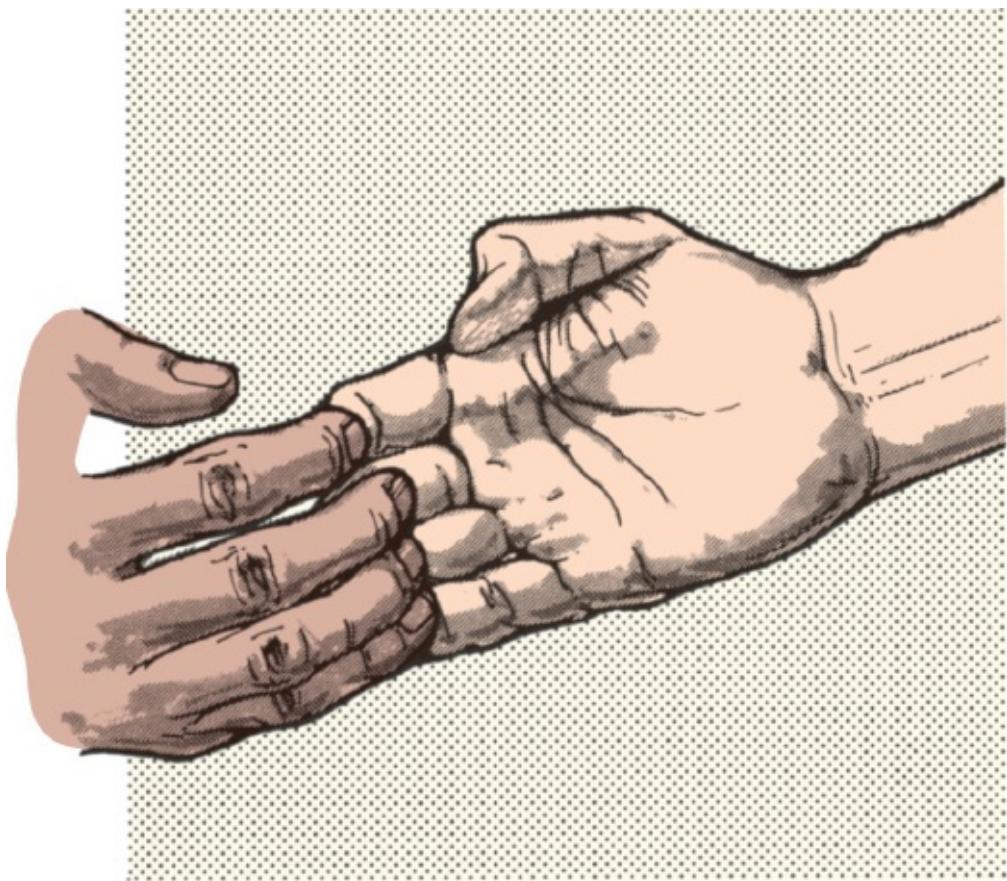


FIGURE 27.21 Examination of the flexor digitorum profundus. The patient resists attempts to extend the distal phalanges while the middle phalanges are fixed.

Making a fist requires flexion of the fingers at all joints. The strength of the grip depends on the degree of flexion at the MCP and IP joints, on the position of the thumb and its ability to flex and to brace the fingers, and on the synergistic actions of the wrist extensors in fixation of the wrist. A firm fist can be made only with the wrist in extension. Grip is commonly used in the assessment of upper-extremity strength. An examiner cannot easily extricate his fingers from the clenched hand of a person with normal grip strength. For quantitative testing, a dynamometer may be used. In fact, although commonly done, grip power is not very useful in assessing upper-extremity motor function in neurologic patients for the following reasons. The finger and wrist flexors are not corticospinal innervated and are not likely to be weak with a mild CST lesion. Grip strength is so unaffected by CST pathology that many patients with a severe, spastic hemiparesis have a tightly fisted hand—enough that palmar hygiene may become a problem. In addition, grip is a complex movement with many different muscles involved, so it is insensitive to peripheral pathology as

well. A good rule is that one can use grip as a strength test only if prepared to name all the muscles involved along with their peripheral nerve, brachial plexus, and root innervations. An examiner sophisticated enough to know this information will be testing individual hand muscles, not grip strength.

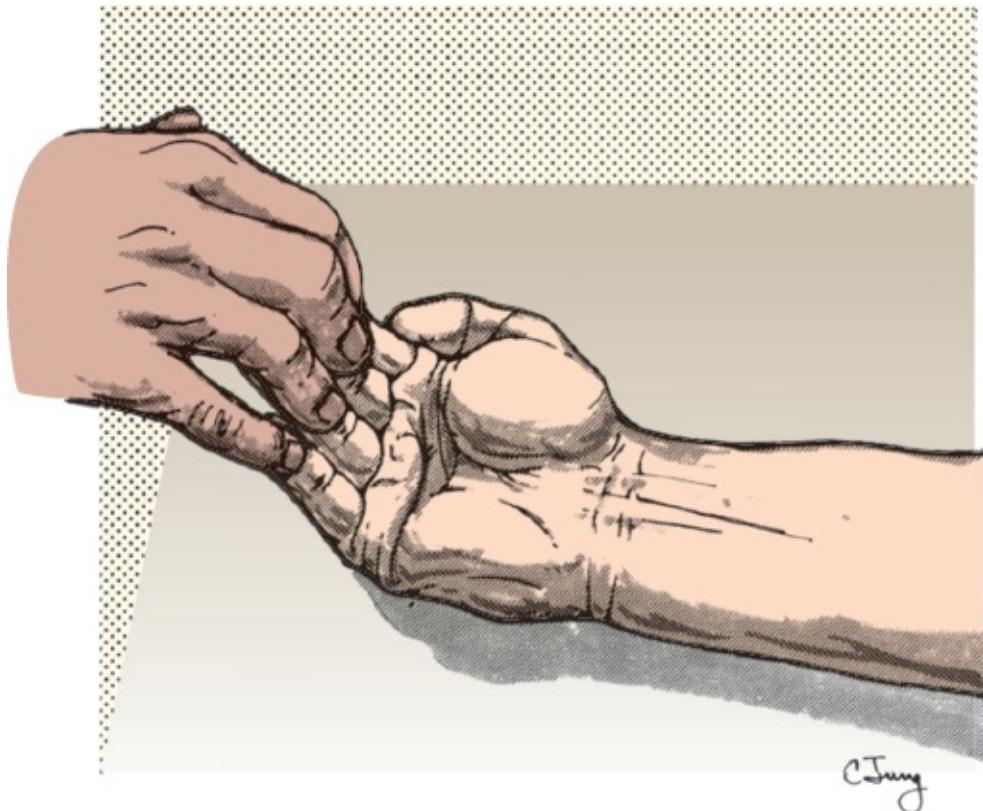


FIGURE 27.22 Examination of the flexor digitorum superficialis. The patient resists attempts to straighten the fingers at the first interphalangeal (IP) joint.

Extension of the Fingers

The long extensors of the fingers include the extensor digitorum communis (EDC), extensor indicis proprius (EIP, aka extensor indicis), and the extensor digiti minimi (EDM). All the finger extensors are innervated by C7-C8 through the posterior interosseous branch of the radial nerve. The tendons insert on the dorsal extensor expansions of the first phalanges of the fingers. The primary action of the EDC is extension of the MCP joints. However, the EDC can exert some force to extend each joint it crosses, including the wrist and—through the extensor expansion complexes—the PIP and DIP joints. Its action on the MCP joints causes some spreading of the fingers, and patients with weakness of the

interossei may use this as a trick movement to abduct the fingers. The EIP extends the index finger and adducts it slightly. The EDM extends the little finger. The interossei and lumbricals also extend the PIP and DIP joints of the fingers.

To test the action of the EDC, EIP, and EDM, the patient resists attempts to push the fingers down at the MCP joints with the forearm pronated and the wrist stabilized ([Figure 27.23](#)). A useful technique is for the examiner to try to overcome the patient's finger extensors with his own. Because they have separate muscles, the index and little fingers can extend independently, but it is very difficult to extend either the middle or ring finger without moving the other. The extensor function of the lumbricals and interossei is tested by having the patient try to extend the PIP and DIP joints against resistance while the MCP joints are hyperextended and fixed ([Figure 27.24](#)).

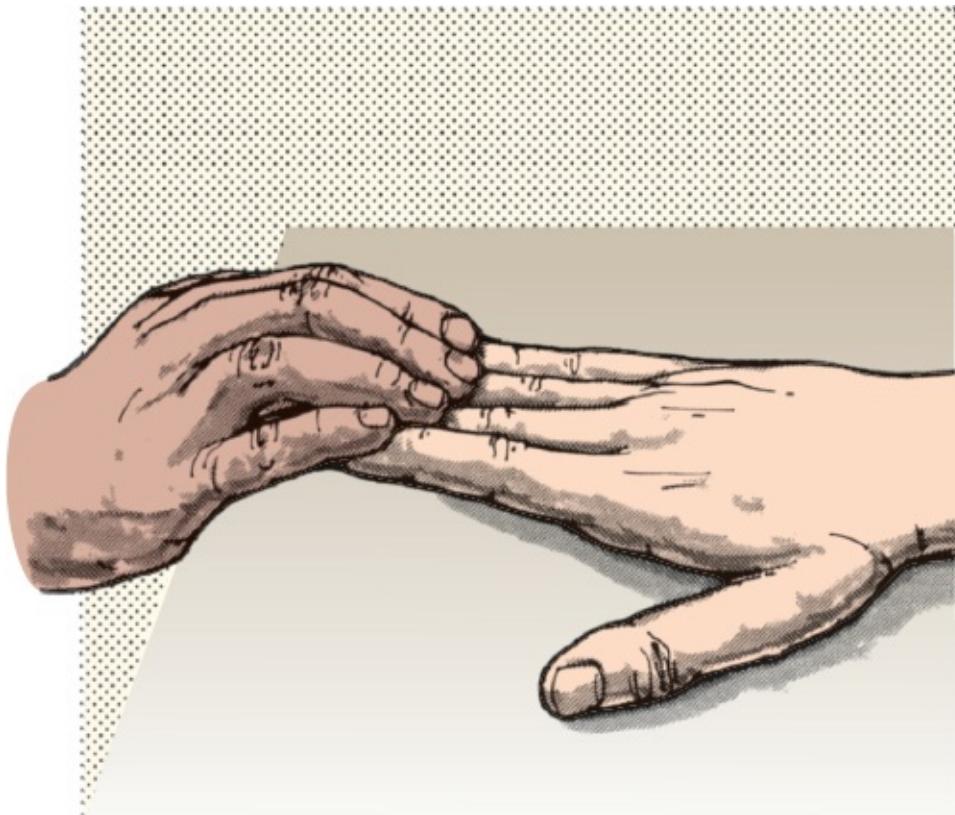


FIGURE 27.23 Examination of the extensor digitorum communis. With hand outstretched and IP joints held in extension, the patient resists the examiner's attempt to flex the fingers at the metacarpophalangeal (MCP) joints.

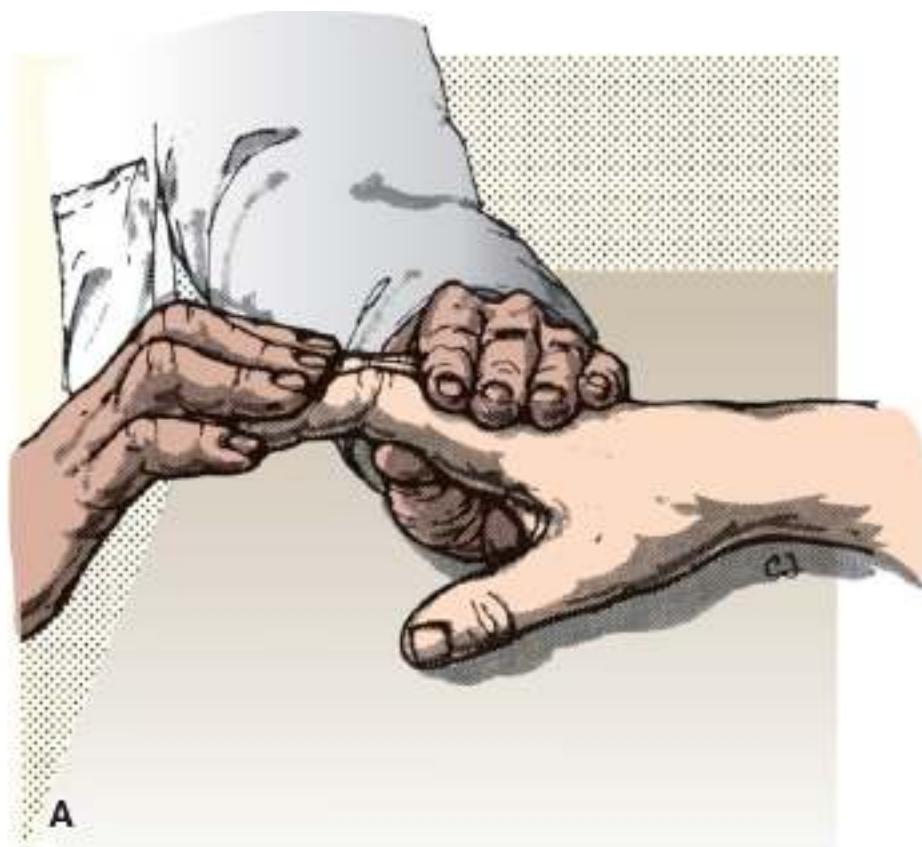
The Thumb and Its Muscles

The thumb is a complex bit of machinery; small wonder it conveyed such an evolutionary advantage. It is capable of movement in many directions. The difference in some of the motions is subtle (e.g., flexion vs. adduction), but the muscle involved and the clinical significance may be marked. Two sets of muscles control thumb motion: those in the forearm (extrinsic thumb muscles) and those that make up the thenar eminence (intrinsic thumb muscles).

The mobility of the opposable thumb requires more elaborate muscle control compared to the other digits. Because the classical anatomical terms describing directions of movement are not easily applied to the thumb, additional directions are designated: palmar, dorsal, ulnar, and radial.

The IP and MCP joints can flex and extend. The carpometacarpal joint can move in many directions. In palmar abduction, the thumb moves upward at right angles to the plane of the palm; in radial abduction, the thumb moves away in the plane of the palm. Ulnar and palmar adductions are movements that touch the first and second metacarpals together. Opposition (anteposition) is the motion of circumduction of the thumb with extended MCP and IP joints; this turns the thumb into semipronation and touches the palmar surface of the tip of the thumb to the palmar surface of the tip of the small finger.

The forearm muscles involved in controlling the thumb are the APL, extensor pollicis longus (EPL), extensor pollicis brevis (EPB), and FPL. The APL abducts the thumb and extends it to a slight degree. The EPL extends the terminal phalanx; the EPB extends the proximal phalanx. These three muscles are supplied by the C7-C8 cervical segments through the posterior interosseous branch of the radial nerve. The APL is the only muscle in the flexor pronator compartment of the forearm supplied by the radial nerve. The FPL (anterior interosseous branch of the median nerve, C8-T1) flexes the distal phalanx of the thumb. To test the FPL, the patient flexes the distal phalanx of the thumb while the proximal phalanx is flexed and immobilized ([Figure 27.25](#)). The EPL is tested by having the patient extend the thumb at the IP joint while the proximal phalanx is immobilized ([Figure 27.26](#)). The EPB is tested by having the patient extend the thumb at the MCP joint while the metacarpal bone is immobilized ([Figure 27.27](#)). Hyperextending the thumb, especially at both joints, causes the tendons of the EPL, EPB, and APL to stand out, forming the “anatomic snuff box.”



A



B

FIGURE 27.24 A and B. Extension of the middle and distal phalanges. The patient attempts to extend the fingers against resistance while the MCP joints are fixed.

The thenar eminence is the mass of muscles on the palmar surface overlying the first metacarpal. The muscles that make up the thenar eminence are the APB, opponens pollicis (OP), and flexor pollicis brevis (FPB). The APL and APB muscles produce palmar abduction. The OP pronates the thumb, turning the volar thumb surface down, to touch the tip of the thumb to the small finger. The FPB flexes the MCP joint of the thumb. In testing the FPB, the patient is asked to flex the MCP joint of the thumb while keeping the IP joint extended. The FPB has a superficial head supplied by the median nerve and a deep head by the deep palmar branch of the ulnar nerve, all C8-T1. The rest of the thenar muscles are supplied by C8-T1 through the median nerve.

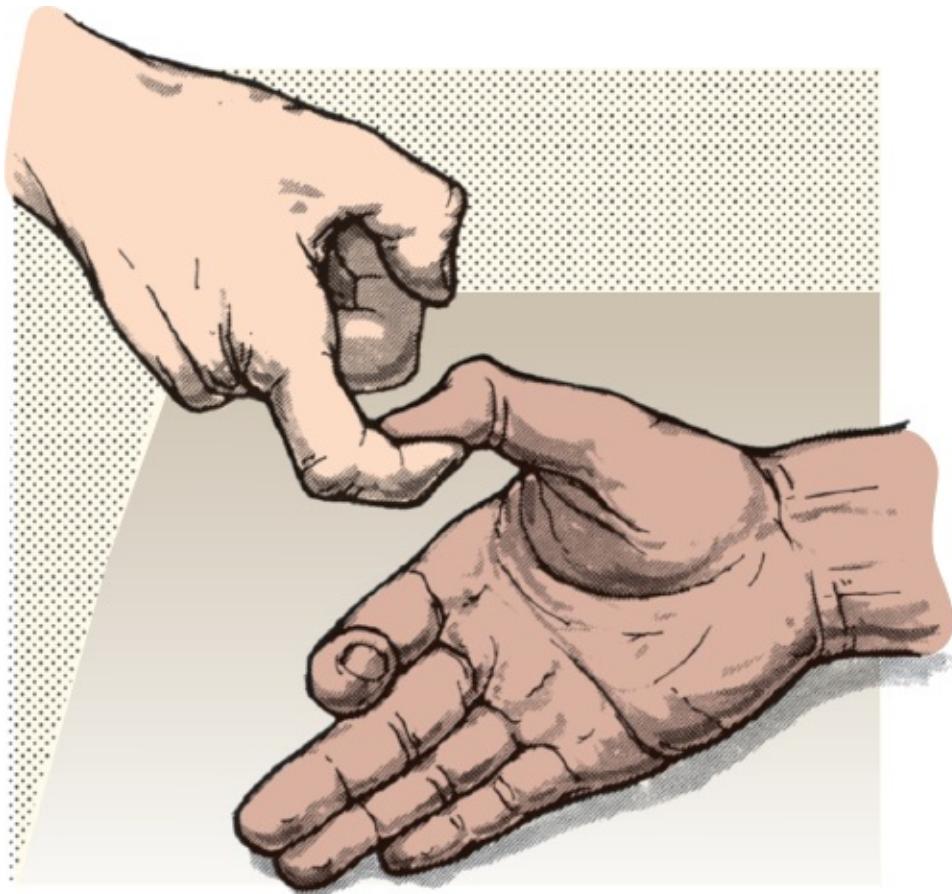


FIGURE 27.25 Examination of the flexor pollicis longus. The patient resists attempts to extend the distal phalanx of the thumb while the proximal phalanx is fixed.

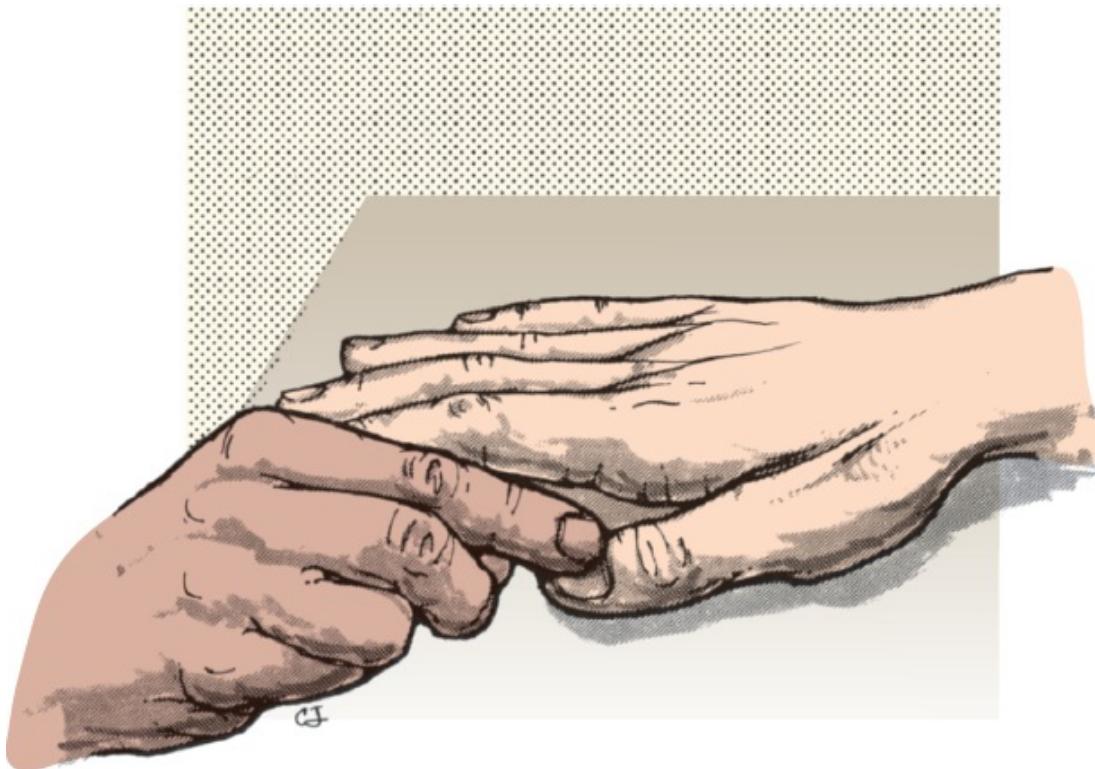


FIGURE 27.26 Examination of the extensor pollicis longus. The patient attempts to resist passive flexion of the thumb at the IP joint; the tendon can be seen and palpated.

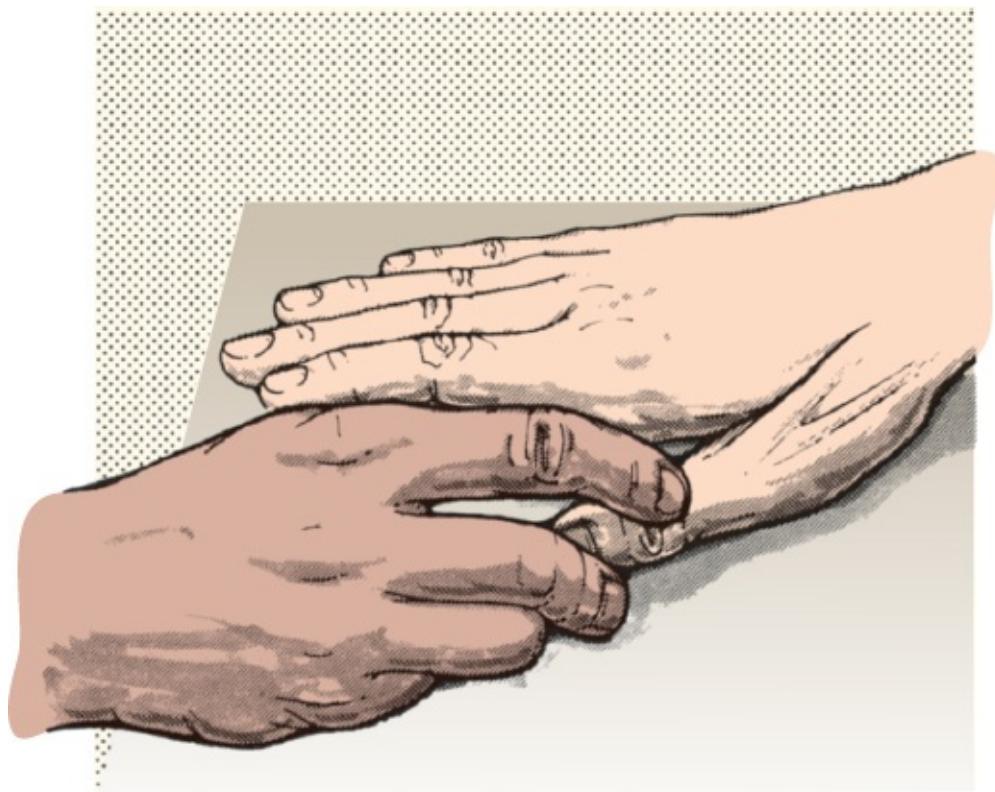


FIGURE 27.27 Examination of the extensor pollicis brevis. The patient attempts to resist passive flexion of the thumb at the MCP joint; the tendon can be seen and palpated.



FIGURE 27.28 Radial abduction of the thumb. The patient attempts to abduct the thumb in the same plane as that of the palm; the tendon of the abductor pollicis longus can be seen and palpated.

Abduction of the thumb is carried out in two planes: in the same plane as the palm (radial abduction) and at right angles to the plane of the palm (palmar abduction). To test radial abduction, the thumb is moved outward if the hand is horizontal, and upward if the hand is vertical, against resistance. This movement is executed by the APL and EPB (Figure 27.28). The APB is a thin sheet of muscle lying just medial to the first metacarpal that performs palmar abduction. To test palmar abduction, the thumb is moved upward at right angles to the palm, inside the radial margin of the hand, against resistance. It is very easy for both

patient and physician to confuse abduction with extension. One trick is to place a pencil or similar object between the thumb and the palm, or radial to the thumb, perpendicular to the palm. The patient then raises the thumb to a point vertically above its original position, keeping it parallel to the pencil with the thumbnail at right angles to the palm ([Figure 27.29](#)). In paralysis of abduction, the thumb is adducted and rotated, thumbnail parallel rather than perpendicular to the fingernails, falling into the plane of the palm (simian or ape hand, [Figure 46.6](#)).

The APB is most often tested by having the patient place the thumb in full palmar abduction and hold it there as the examiner tries to force the thumb down toward the palm. The examiner may do this in several ways. A commonly used technique is for the examiner to hold the back of the patient's hand in his palm, touch the radial side of the patient's thumb with the palmar aspect of his own (usually right hand against right hand), and then make a compound motion with the long and short flexors and opponens to pull the patient's thumb down. The patient, using only his thumb abductors, is outnumbered and outgunned and will usually lose. A better technique is to use APB against APB. The examiner holds the patient's hand as above. The examiner and patient both hold their thumbs in full abduction, radial aspect to radial aspect, IP joint to IP joint; then, the examiner, using only his thumb abductors, tries to press the patient's thumb down. This is normally a match. If the patient's APB yields, the degree of weakness may be semiquantitated if the examiner then uses progressively weaker muscles against the patient's APB. This is sometimes useful for follow-up. Suppose the patient's APB could be overcome by the examiner's APB, first dorsal interosseous (first DI), and ADM on the first visit, but after treatment, the patient's APB can stand up to the examiner's ADM. This means that there has been improvement. If the patient's APB could stand up to the examiner's ADM initially but fails later, then there has been worsening.

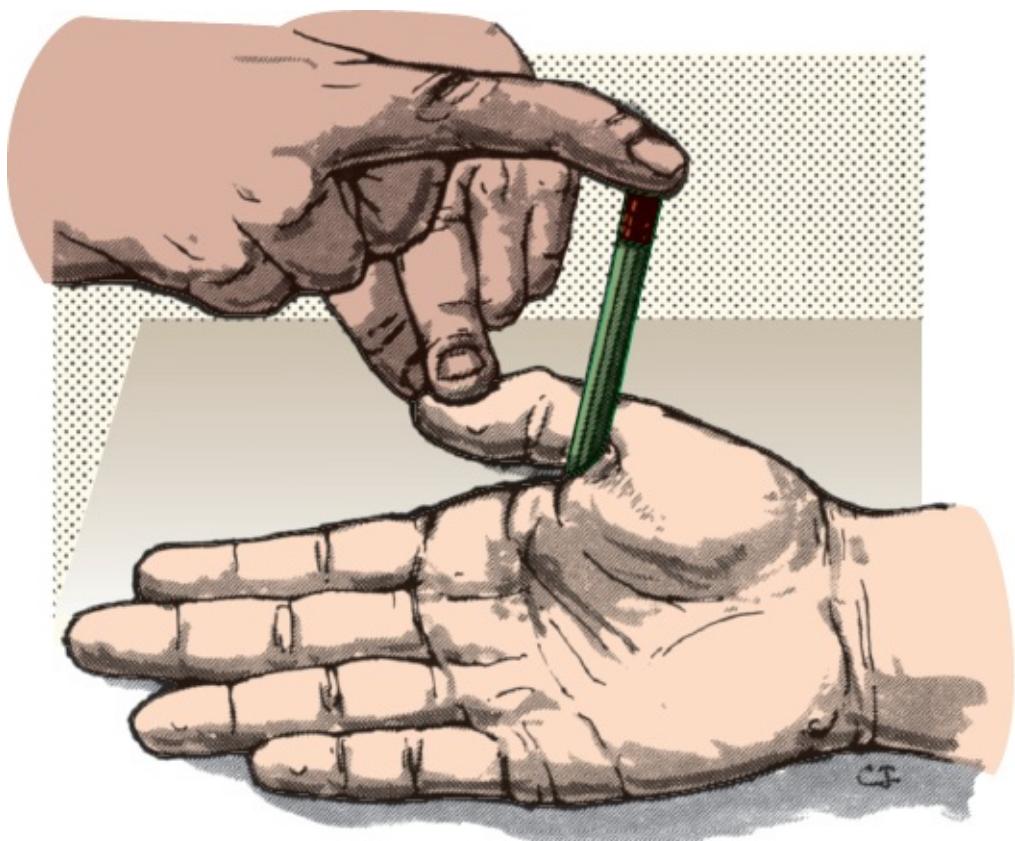


FIGURE 27.29 Palmar abduction of the thumb. The patient attempts, against resistance, to bring the thumb to a point vertically above its original position.

Side-to-side “confrontational testing” is a similar like-muscle-against-like-muscle method for testing hand intrinsic strength. The patient pushes his own abducted small fingers or index fingers together. If one side is weak, the finger on the strong side will force the finger on the weak side into adduction. See Dr. John K. Wolf’s *Segmental Neurology* for further elaboration on these elegant muscle-to-muscle techniques.

Opposition of the thumb is tested by having the patient touch the little finger with the thumb (Figure 27.30). With the thumbnail on a plane approximately parallel to the palm, the palmar surface of the tip of the thumb should contact the palmar surface of the tip of the little finger. When the OP is weak, the patient may be able to oppose the thumb to the index or middle finger, but not the little finger. In testing opposition of the little finger by the opponens digiti minimi (ulnar nerve, C8-T1), the patient moves the extended little finger in front of the other fingers and toward the thumb (Figure 27.31). Opposition of the thumb and little finger may be tested in one maneuver. When both are opposed, their extended tips meet and form an arch over the cupped palm (Figure 27.32). The

strength of the combined movement may be gauged by the patient's ability to hold onto a piece of paper held between finger and thumb as the examiner tries to pull it free, or the examiner may attempt to pull his finger between the touching tips of the thumb and little finger. The flexors of the thumb and little finger and the short abductor of the thumb probably enter into these movements.



FIGURE 27.30 Examination of the opponens pollicis. The patient attempts, against resistance, to touch the tip of the little finger with the thumb.

The adductor pollicis is the final muscle innervated by the deep palmar branch of the ulnar nerve (C8-T1). It adducts the thumb and flexes the first metacarpal. Adduction of the thumb is also carried out in two planes: in the plane of the palm (ulnar adduction) and in a plane at right angles to the palm (palmar adduction). Ulnar adduction is touching the ulnar aspect of the thumb to the radial aspect of the second metacarpal, the thumb in the same plane as the palm and the thumbnail as nearly as possible parallel with the other fingernails, as if to put the hand into salute position. In palmar adduction, the ulnar aspect of the thumb touches the palmar aspect of the second metacarpal so that the thumb and index finger lie perpendicular to each other, with the thumbnail at right angles to the other fingernails (Figure 27.33). A commonly used test of adduction power in either of these positions is to have the patient try to hold a

piece of paper tightly between thumb and hand as the examiner tries to extract it ([Figure 27.34](#)). When thumb adduction is weak, the patient may make a substitution movement, flexing the IP joint with the FPL and trying to secure the paper with the tip of the thumb (Froment's sign), a common finding in ulnar neuropathy.

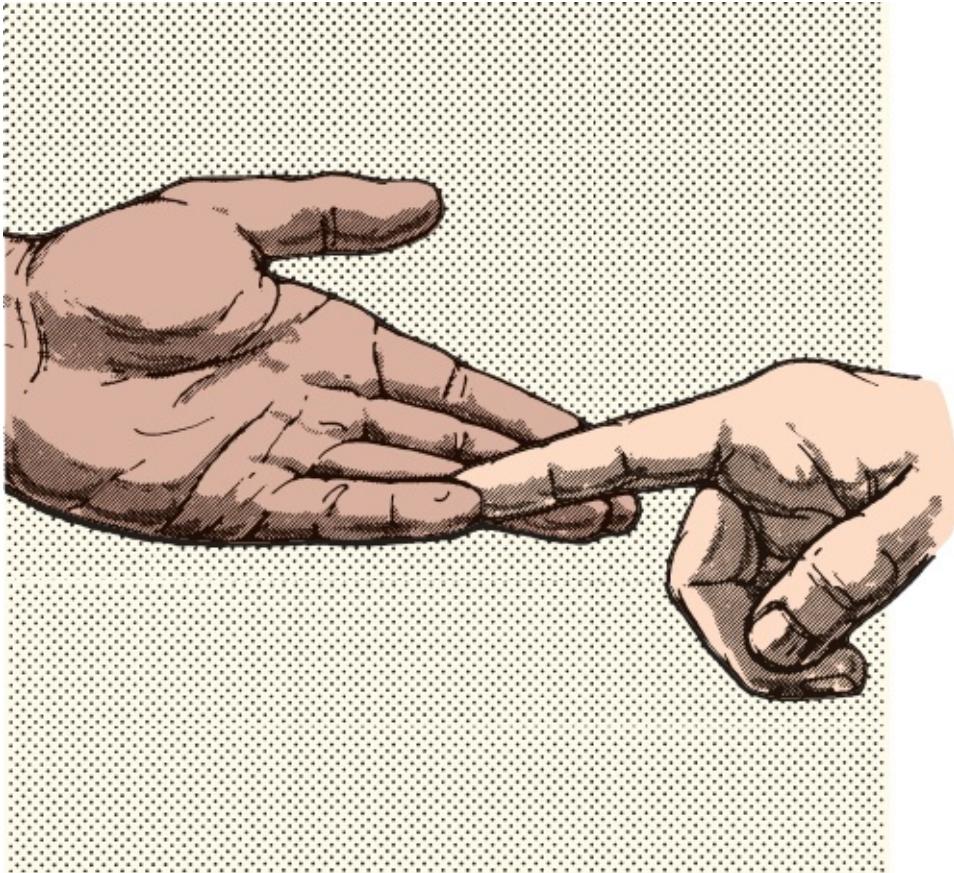


FIGURE 27.31 Examination of the opponens digiti minimi. The patient attempts to move the extended little finger in front of the other fingers and toward the thumb.

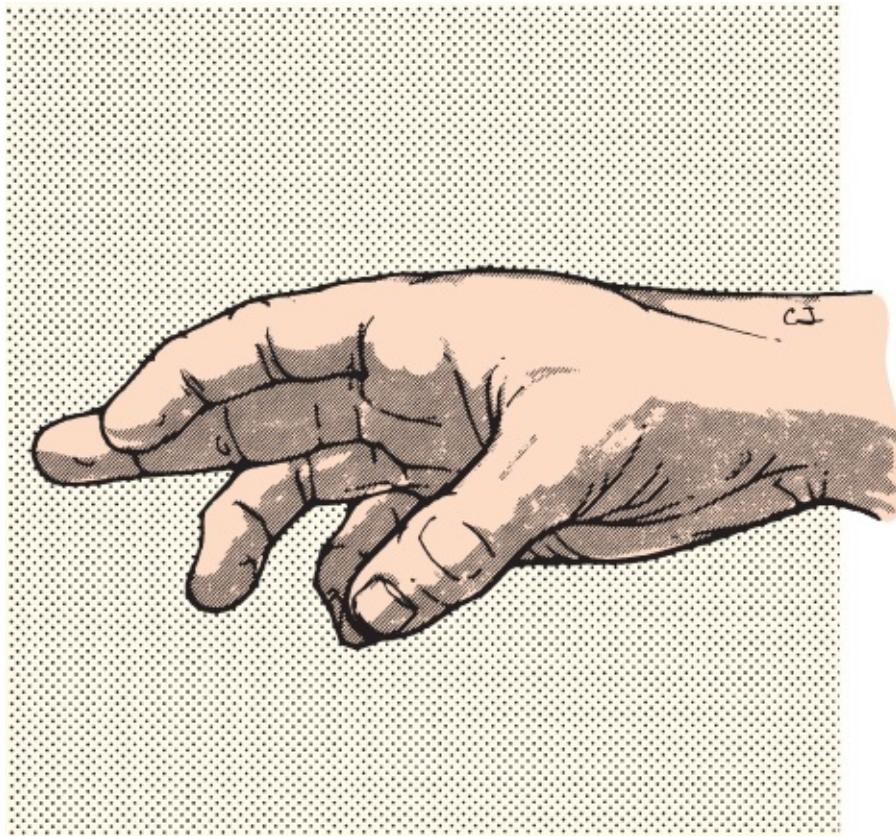


FIGURE 27.32 Opposition of the thumb and little finger.

Adduction of the fingers is the movement that brings the fingers tightly together; abduction spreads the fingers apart. Adduction is a function of the volar interossei, whereas abduction is a function of the dorsal interossei. Abduction of the little finger is done by the ADM. Adduction may be tested in several ways. With the fingers abducted and extended, the patient may try to adduct the fingers against resistance (Figure 27.35). The patient may try to clutch a piece of paper between two fingers and resist the examiner's attempts to withdraw it. The examiner may interdigitate his fingers between the patient's and have the patient squeeze as tightly as possible. Another test is to have the patient make a "finger cone," by holding the hand palm up, touching the index and ring fingers together above the middle finger, and then laying the small finger atop the ring finger. This movement requires good adduction power. If the patient then opposes the thumb to the small finger, all major muscle groups of the hand have been tested in one quick maneuver. Yet another adduction test is to have the patient, palm down, lay the middle finger across the index finger as far as possible, trying to touch the ulnar aspect of the middle finger to the radial aspect of the index finger.

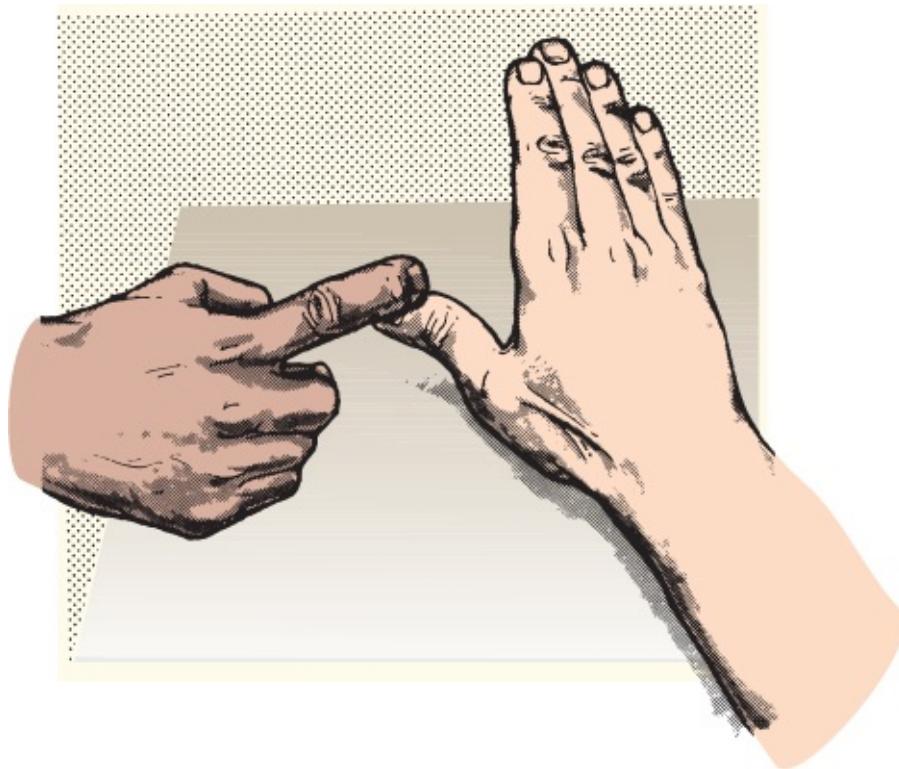


FIGURE 27.33 Palmar adduction of the thumb. The patient, against resistance, attempts to approximate the thumb to the palmar aspect of the index finger; the thumbnail is kept at a right angle to the nails of the other fingers.

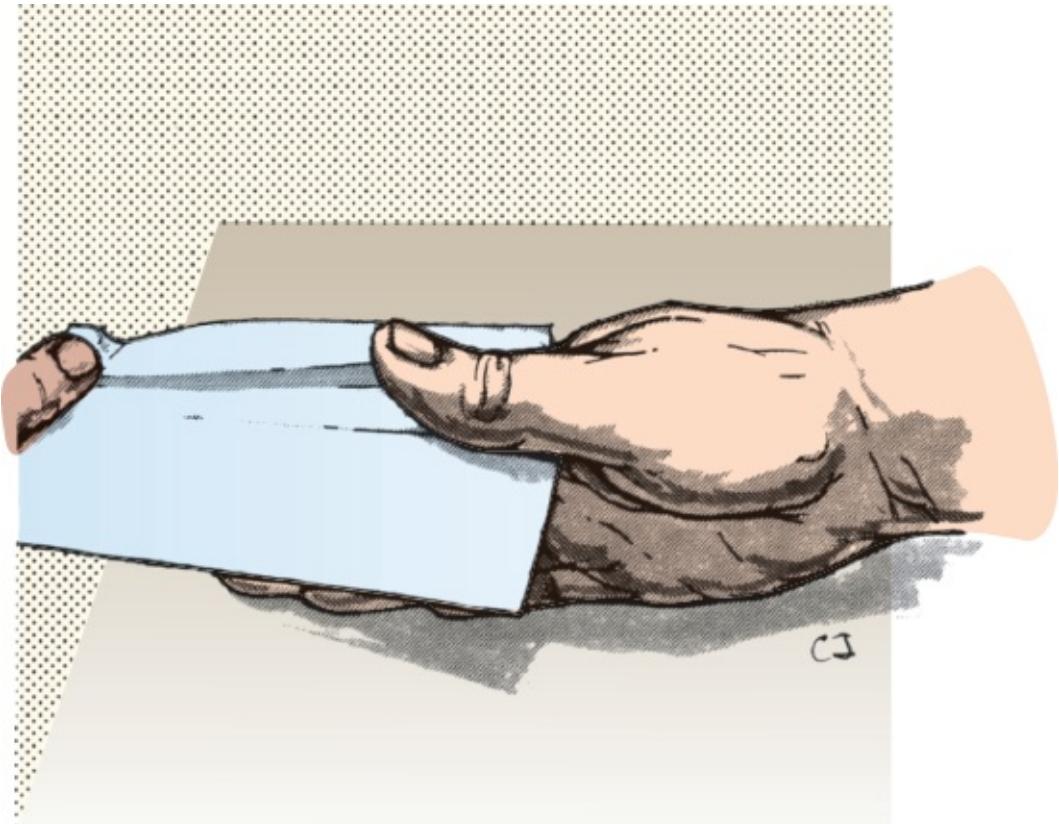


FIGURE 27.34 Ulnar adduction of the thumb. The patient attempts to grasp a piece of paper between the thumb and the radial border of the index finger while the thumbnail is parallel to the nails of the other fingers. In ulnar neuropathy, the IP joint flexes to compensate for weakness of the adductor pollicis (Froment's sign, [Chapter 46](#)).



FIGURE 27.35 Adduction of the fingers. The patient attempts to adduct the fingers against resistance.

The usual test of abduction is to have the patient keep the fingers fully extended and spread apart and resist the examiner's attempt to bring them together ([Figure 27.36](#)). In most circumstances, the examination concentrates on the first DI and the ADM. A commonly used technique is for the examiner to use a claw-like grip with his palm against the back of the patient's fingers, patient and examiner both palm down, examiner's small finger hooked around the patient's small finger, and thumb around the patient's index finger; then, the examiner pulls the thumb and small finger together to overpower the patient's finger abductors. This is a mismatch in that the muscles the examiner is using are more powerful than the muscles the patient is using, so the examiner always overcomes the finger abductors and must develop a feel for how much resistance

is normal. A better technique is for the patient and examiner to match muscles. The patient and examiner both have their hands palms down, fingers extended and abducted. With the radial aspect of his right index finger against the radial aspect of the patient's right index finger, the fingertip of each about at the level of the PIP joint of the other, the examiner tries to overcome the patient's first DI with his own first DI, like muscle against like muscle (as described for the APB above). Similarly for the ADM, but patient and examiner are both palms up. If the patient and examiner are both right handed or both left handed, this method has the advantage of pitting dominant hand against dominant hand and nondominant against nondominant. A more efficient if less ideal technique for testing finger abduction of the patient's right hand is as follows. The patient holds his right hand palm down, fingers fully abducted. The examiner holds his right hand palm down, his left palm up, right index finger against the patient's right index finger, and left small finger against the patient's right small finger. Using these like muscles, the examiner then tries to force the patient's fingers together.



FIGURE 27.36 Examination of the abduction of the fingers. The patient resists the

examiner's attempt to bring the fingers together.

Examination of Movements and Muscles of the Thorax, Abdomen, and Trunk

The actions of the large muscles of the trunk, chest, and abdomen are often combined, and it is difficult to evaluate them individually (Table 27.6). Except for the respiratory muscles, most of these muscles have scant neurologic significance.

The Muscles of the Thorax

The major thoracic muscles consist of the internal and external intercostals and diaphragm. Muscles attached to the sternum, clavicles, and scapulae act as accessory muscles of respiration. The intercostal muscles are innervated by the intercostal nerves, which are the anterior divisions of the 12 thoracic spinal nerves. The diaphragm is innervated by C3-C5 through the phrenic nerves, which arise directly from the nerve roots. The diaphragm is the principal muscle of respiration. During quiet inspiration, intercostal contraction expands the anteroposterior and transverse diameters of the thorax, and the vertical diameter is increased by the descent of the diaphragm. In deep inspiration, additional muscles are brought into action, including the scaleni, SCMs, and other muscles that act on the shoulders, clavicles, and scapulae. The diaphragm also contracts during various expulsive acts such as coughing, sneezing, laughing, vomiting, hiccuping, urination, defecation, and parturition.

Weakness of the intercostal muscles causes adduction of the costal margins and abdominal respiration, with alternate bulging and retraction of the epigastrium as increased diaphragmatic contraction compensates for the intercostal weakness (abdominal breathing). The intercostal spaces may retract during inspiration, and the ribs do not rise and separate. When bilateral paralysis of the diaphragm is present, the excursion of the costal margins is increased and the epigastrium does not bulge during inspiration. The moving shadow caused by retraction of the lower intercostal spaces during inspiration (Litten's sign) is absent. The expulsive acts, primarily coughing, are carried out with difficulty. Quick, forceful diaphragmatic contractions are impaired; one manifestation of this may be the inability to sniff. Unilateral diaphragmatic weakness is difficult

to detect, but the excursion of the costal margin on the affected side during quiet inspiration may be slightly increased, and Litten's sign is absent. Fluoroscopy, ultrasonography, and phrenic nerve conduction studies, not to mention pulmonary function tests, are far superior to physical examination in detecting diaphragmatic impairment. Diaphragm function should be particularly assessed in patients with spinal cord lesions that involve the C3-C5 segments. If there is diaphragmatic or intercostal weakness, the accessory muscles that act in deep inspiration are brought into play, and breathing recruits the scaleni, SCMs, serrati, and pectorals.



FIGURE 27.37 Examination of the abdominal muscles. The recumbent patient attempts to raise his head against resistance.

The Muscles of the Abdomen

The abdominal muscles are the rectus abdominis, pyramidalis, transversus abdominis, and obliqui. Weakness of these muscle groups does not often occur in neurologic patients. The abdominal muscles are innervated by the anterior divisions of the thoracic spinal nerves. The rectus abdominis flexes the vertebral column, compresses the abdominal viscera in such acts as defecation and

parturition, and aids in forced expiration.

When performing a sit-up, the abdominal muscles contract strongly during the initial phase of the movement, when raising the head and shoulders. After the shoulders have been raised about 8 in, the hip flexors contract strongly and bring the trunk to an upright position. The abdominal muscles may be tested by having the patient raise the head against resistance ([Figure 27.37](#)), cough, or do a sit-up. If the abdominal muscles are weak but the hip flexors normal, hyperextension of the spine will occur during an attempted sit-up. If the abdominal muscles contract equally in all four quadrants, the umbilicus will not move. If the lower abdominal muscles are paralyzed, as in a T10 myelopathy, the upper abdominal muscles will pull the umbilicus cephalad when the patient raises the head or attempts a sit-up (Beevor's sign, see [Video Link 27.3](#)). More rarely, abnormal movement of the umbilicus may occur with weakness of the upper abdominals or if the weakness is unilateral.

The Muscles of the Pelvis

The pelvic muscles, including the urinary bladder and the perineal and external genital muscles, are not accessible for the usual clinical testing. A crude assessment can be obtained by reflex activation, for example, cremasteric, bulbocavernosus, and anal wink. These are discussed in [Chapter 39](#). The bladder and some of the functions of the genitalia are discussed in [Chapter 45](#).

The Muscles of the Spine

The muscles that extend and rotate the spine were introduced above in the discussion of neck muscles. Most of these muscle groups extend down the entire spine, and for general clinical purposes, they are considered as a group: the erector spinae, paravertebral, or paraspinal muscles. Paraspinal examination is an important part of needle electromyography. Abnormalities in the paraspinal muscles help differentiate nerve root disease from plexus and peripheral nerve disease. The paraspinals are also often abnormal electromyographically in myopathies. The movements of the spine are flexion, extension, rotation, and lateral bending. The muscles that produce these movements are examined en masse by examining the movements rather than individual muscles. All the paraspinal muscles are innervated by the posterior primary rami of the spinal nerves.

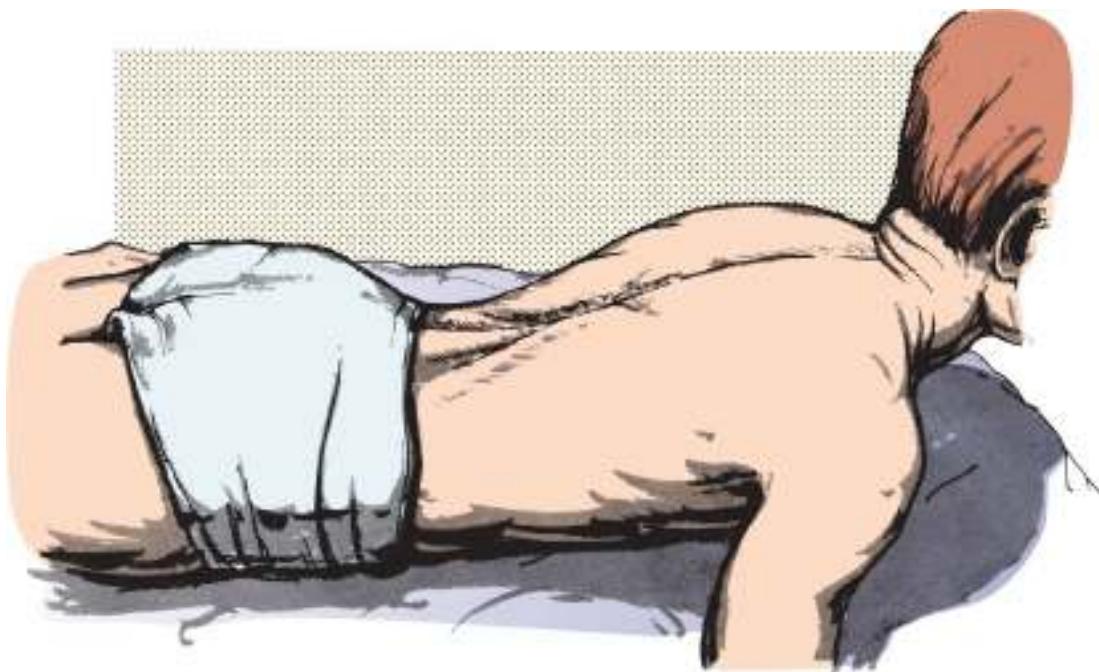


FIGURE 27.38 Examination of the extensors of the spine. The patient, lying prone, attempts to raise the head and upper part of the trunk.

The extensors of the spine are tested by having the prone patient raise the head and shoulders without the assistance of the hands (Figure 27.38); in a continuation of this, the young and supple may be able to raise the legs and balance on the belly. The ability to perform these maneuvers indicates normality, but further assessment may be done by pressing downward against the patient's extension motion. The most common cause of paraspinal weakness is primary muscle disease, particularly muscular dystrophies, especially FSH dystrophy. Patients with weak spine extensors often adopt a lordotic, sometimes hyperlordotic, posture. This is because the paraspinals normally function like guy-wires on a tower antenna, helping to balance the spine above the pelvis. With forward bending, contraction of the paraspinals prevents tipping over. Patients with paraspinal weakness cannot rely on their spine extensors to keep them from falling forward, so they lean backward to compensate and maintain balance. The result is the lordotic posture. In FSH, the hyperlordosis may reach bizarre proportions, with the patient leaning so far backward the spine approaches horizontal. The lordosis disappears when the patient lies down. When patients with some dystrophies, particularly Duchenne's, become wheelchair bound, the paraspinal weakness may lead to scoliosis, often severe enough to require surgical intervention.

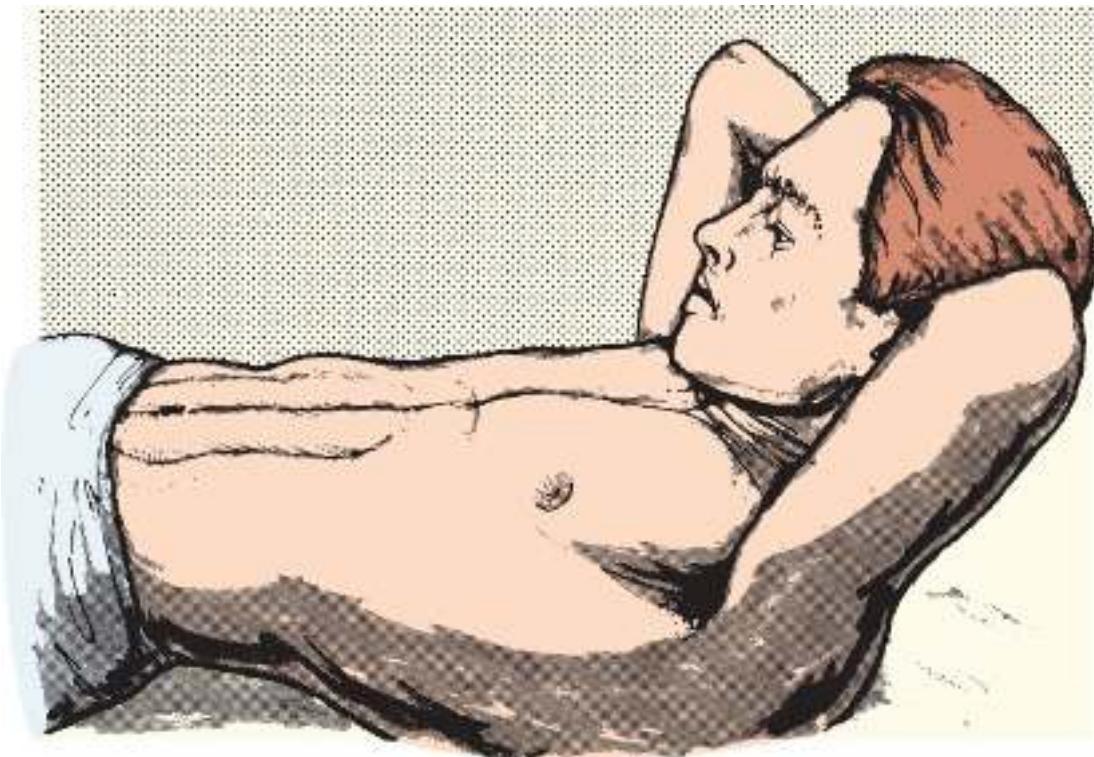


FIGURE 27.39 Examination of the abdominal muscles and flexor muscles of the spine. The patient attempts to rise from a recumbent to a sitting position without the use of the hands.

The flexors of the spine are tested by having the patient rise from recumbent to seated and then to a standing position without using the hands (Figure 27.39). The flexors of the abdomen and hips are also involved in this movement. The flexors and extensors of the spine may both be tested by having the patient try to touch toes with fingertips and stand back up.

Examination of the Movements and Muscles of the Lower Extremities

The movements of the lower extremities are less complex than those of the upper extremities, and there are fewer substitution movements. Table 27.7 lists the pertinent muscles and their innervation. Some muscles of the lower extremity have complicated actions that seem discordant (e.g., the hamstrings flex the knee but extend the hip). This is largely due to the rotation of the lower extremity into a new functional position that occurred during the evolution to land vertebrates. The ventral surface of the thigh came to lie posterior so that the knee flexes

backward even though the hip flexes forward.

The Hip Joint

The movements that take place at the hip are flexion, extension, abduction, adduction, and internal and external rotation. The principal hip flexor is the iliopsoas. Important accessory hip flexors are the rectus femoris, sartorius, and tensor fascia lata. The iliopsoas has two parts, the psoas and the iliacus, which have the same function. The psoas portion is innervated by branches from the lumbosacral plexus (L1-L4). The psoas arises from both the transverse processes and the bodies of the lumbar vertebra. The intervertebral foramina of L1-L4 lie between these two points of origin so that the roots that form the lumbar plexus exit into the substance of the muscle and the plexus lies within it. This anatomy accounts for the severe damage to the lumbosacral plexus that commonly occurs with hemorrhage into the psoas muscle. The iliacus portion arises in the iliac fossa and is innervated by the femoral nerve (L2-L4).

The two iliopsoas muscles acting together from each side help to maintain an erect posture by balancing the spine and pelvis over the femurs, preventing a backward tilt. When the legs are fixed, they flex the trunk and pelvis forward, as in doing a sit-up. Hip flexor strength is tested by having the patient flex the hip against resistance ([Figure 27.40](#)). This may be done in the sitting or supine position. If sitting, the patient should not be permitted to lean backward. When testing in the sitting position, normal hip flexors cannot be overcome by an examiner using hand and arm strength from an arm's length away. If the examiner stands close and uses his body weight, the hip flexors can usually be defeated. When testing supine, a patient with mild hip flexor weakness may still be able to raise the leg with the knee extended; with more severe weakness, the hip can only be flexed with the knee flexed. Another hip flexor test is to have the supine patient attempt to maintain both lower extremities flexed at the hip and extended at the knee, the legs at about a 45-degree angle off the bed, feet apart. This is a difficult callisthenic maneuver that not all patients can perform. If the hip flexors are mildly weak unilaterally, as in a CST lesion, the involved lower extremity will drift downward more rapidly than its fellow (leg drift). Another version of this test can be used to look for subtle hamstring weakness (see “The Knee Joint”).

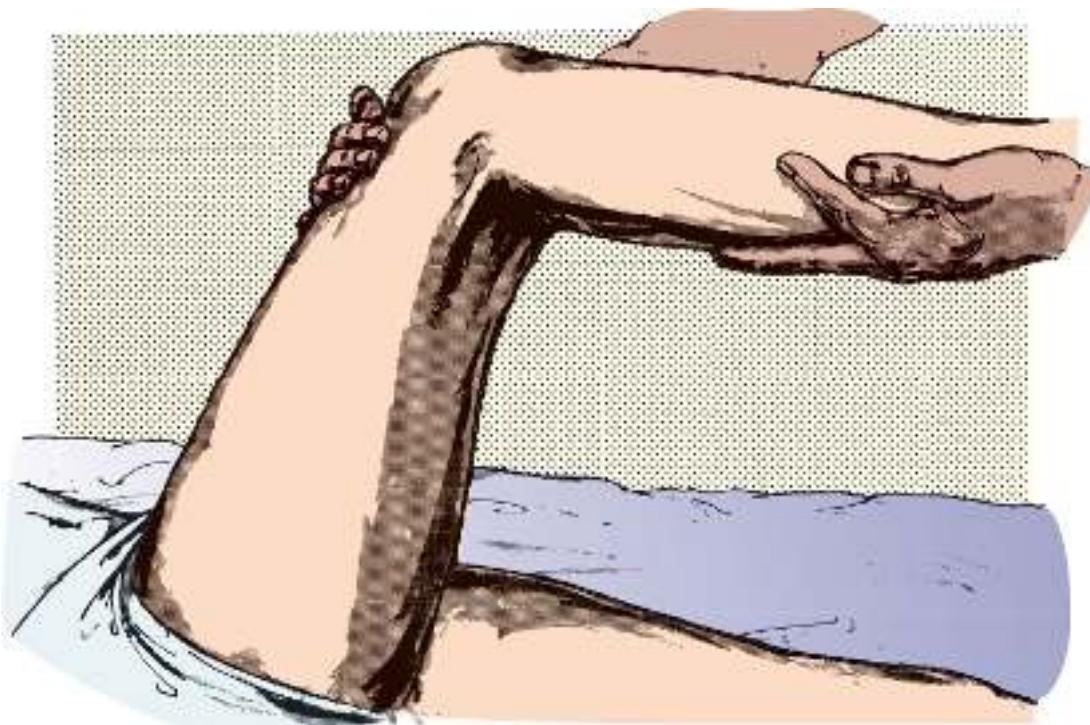


FIGURE 27.40 Examination of the flexors of the thigh. The patient attempts to flex the thigh against resistance; the knee is flexed and the leg rests on the examiner's arm.

The major hip extensor is the gluteus maximus (inferior gluteal nerve, L5-S2). The gluteus maximus is the most powerful extensor and external rotator of the thigh. Accessory hip extensors are the glutei medius and minimus, hamstrings, and hip adductors. The gluteus maximus is important in climbing steps, jumping, and rising from a chair. Hip extensor function is best tested with the patient prone, raising the flexed knee up from the table against downward pressure from the examiner (Figure 27.41). Having the knee flexed minimizes any contribution from the hamstrings. The gluteus maximus can also be tested with the patient lying on the side and extending the hip, or seated and trying to press the raised knee back down as the examiner holds it up, or by testing the ability to stand upright from a stooped position. With hip girdle weakness, particularly in the muscular dystrophies, there is marked weakness of the hip extensors, and the patient arises from a stooped position by using his hands to “climb up the legs” (Gowers’ sign or maneuver, see [Video Link 27.4](#)).

The primary abductors of the hip are the gluteus medius, gluteus minimus, and tensor fasciae lata (TFL). These muscles are supplied by L4-S1 through the superior gluteal nerve. They also function as internal rotators of the hip. The TFL also tightens the fascia lata and flexes the hip. The hip abductors may be

tested either supine or sitting by having the patient attempt to hold the lower extremities outward with ankles spread far apart as the examiner tries to force the ankles together ([Figure 27.42](#)). Weakness of hip abduction is often present in myopathies affecting the hip girdle musculature.

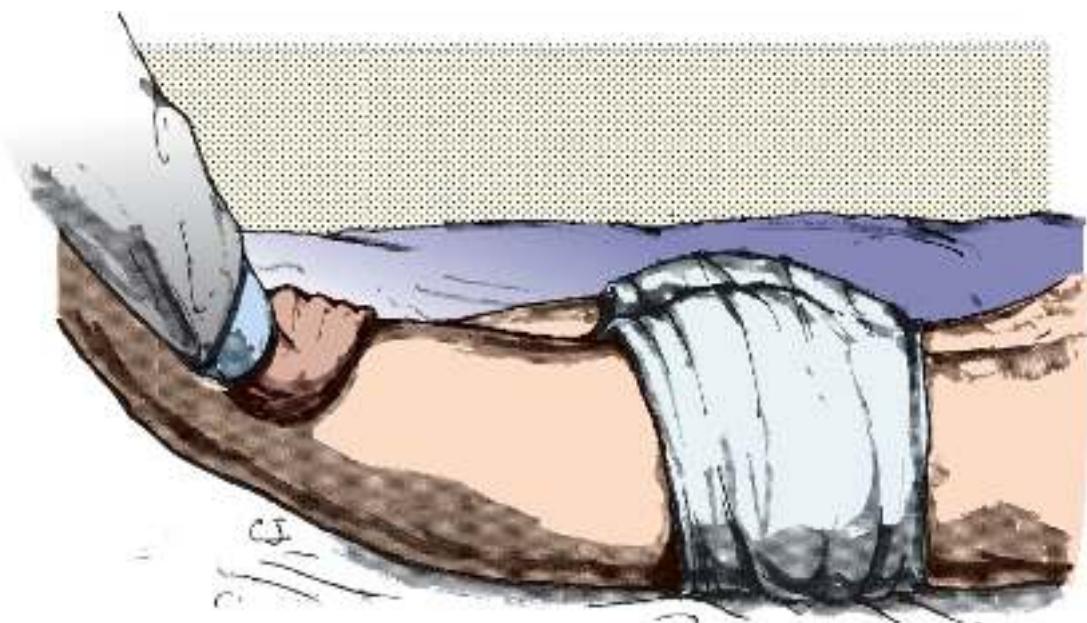


FIGURE 27.41 Examination of the extensors of the thigh at the hip. The patient, lying prone with the leg flexed at the knee, attempts to extend the thigh against resistance; contraction of the gluteus maximus and other extensors can be seen and palpated.

The hip abductors are very important in walking. With each step, the abductors of the stance leg must generate enough force to balance all the weight of the rest of the body in order to keep the pelvis level. Without contraction of the hip abductors, the hip would slide laterally toward the stance leg as the pelvis tilted and the stance leg adducted. When the hip abductors are weak, there is an exaggerated pelvic swing during the stance phase as the pelvis on the side of the swing leg drops downward (Trendelenburg's sign). When bilateral, the result is a gait pattern referred to as a pelvic waddle, which resembles the exaggerated hip swing of a fashion model. The waddling gait is particularly common in myopathies that weaken the pelvic girdle musculature. A unilateral Trendelenburg sign can occur with processes that weaken the hip abductors unilaterally, such as lumbosacral radiculopathy ([Chapters 44 and 47](#)).



FIGURE 27.42 Abduction of the thigh at the hip. The recumbent patient attempts to move the extended leg outward against resistance; contraction of the gluteus medius and tensor fasciae latae can be palpated.



FIGURE 27.43 Examination of adduction of the thigh at the hip. The recumbent patient attempts to adduct the extended leg against resistance; contraction of the adductor muscles can be seen and palpated.

Adduction of the hip is principally a function of the three adductors: longus, brevis, and magnus. The adductor magnus is the longest and strongest hip adductor. Other muscles may play a minor role. The three adductors are supplied by L2-L4 through the obturator nerve. The adductor magnus also receives a twig from the sciatic nerve carrying innervation from L4 to L5. The adductors can be tested with the patient supine, sitting, or lying on one side. The patient attempts to bring the legs together as the examiner tries to keep them apart ([Figure 27.43](#)). As with the abductors, the adductors are so powerful it is helpful to keep the patient's knees extended to give the examiner the advantage of a longer lever. When testing with the patient in the decubitus position, the examiner raises the uppermost leg with the patient trying to keep the legs together; this should raise the entire body, with the lowermost leg remaining apposed and following the upward movement. When the uppermost adductors are weak, the leg will passively abduct and the torso will not move upward; when the lowermost adductors are weak, the lower leg will not follow the upward motion, remaining behind on the bed.

Internal, or medial, rotation of the hip is carried out principally by the hip abductor muscles (glutei medius and minimus, and TFL), with some contribution from the adductors. To test internal rotation, the patient lies supine with the hip and knee flexed or prone with the knee flexed. He then attempts to move the foot laterally against resistance, thus rotating the hip medially ([Figure 27.44](#)). Internal rotation can also be tested with the patient supine and the leg extended, rotating the foot medially as if to touch the big toe to the bed. Rotating the foot medially with the knee extended produces the same hip motion as carrying the foot laterally with the knee flexed. With a unilateral CST lesion (e.g., acute stroke), the internal rotators are weak. When the patient lies supine, the involved leg lies externally rotated compared to its fellow. This asymmetry of leg position may be a clue to the presence of a hemiparesis in an obtunded patient.

External, or lateral, rotation of the thigh at the hip is carried out primarily by the gluteus maximus. The piriformis externally rotates the extended thigh but abducts the flexed thigh. External rotation is tested by maneuvers similar to those for testing internal rotation, but the patient rotates the hip externally by attempting to carry the foot medially against resistance with the knee flexed. If these muscles are paralyzed, the entire leg is turned inward.

An array of short muscles around the hip (obturators, gemelli, quadratus femoris, piriformis, and pectineus) probably play a more important role as postural muscles maintaining hip stability than as prime movers. The sartorius

(femoral nerve, L2-L3), the longest muscle in the body, has a complex set of actions. It is an abductor, flexor, and lateral rotator of the hip and a flexor and medial rotator of the knee. The sartorius would be active when trying to look at the bottom of one's foot.



FIGURE 27.44 Examination of internal rotation of the thigh. The patient, lying prone with the leg flexed at the knee, attempts to carry the foot laterally against resistance, thus rotating the thigh medially.

The Knee Joint

The major movements that take place at the knee joint are flexion and extension. Flexion of the knee is carried out primarily by the hamstring muscles (biceps femoris, semimembranosus, and semitendinosus). Other muscles may play a contributory role. The hamstrings also act as powerful hip extensors. The biceps femoris (external or lateral hamstring) has two heads, long and short. The belly of the long head overlies the short head except just above the popliteal fossa. Both heads are innervated by the sciatic nerve (L5, S1-S2), but the long head is supplied by the tibial division and the short head by the peroneal division. The innervation of the short head by the peroneal division is important in distinguishing lesions involving the peroneal (fibular) nerve at the knee from

those involving the peroneal division of the sciatic, which may be otherwise inseparable. Involvement of the short head is not detectable by physical examination but can be detected electromyographically. The common peroneal nerve at the knee lies just medial to the biceps femoris tendon. The semimembranosus and semitendinosus muscles (internal or medial hamstrings) are supplied by branches of the sciatic nerve (L5, S1-S2).

The knee flexors may be tested with the patient prone ([Figure 27.45](#)), supine, or sitting. With the knee in partial flexion, the patient resists the examiner's attempts to straighten the knee. The knee flexors are powerful and cannot normally be overcome. Another test is to have the prone patient attempt to maintain both knees flexed at about 45 degrees from horizontal with the feet slightly apart. When the knee flexors are weak on one side, as in a CST lesion, the involved leg will sink, gradually or rapidly (leg drift, leg sign of Barré). Examination of knee flexion with the patient prone makes it easier to see and palpate the muscle contractions and lessens the likelihood of misinterpretation because of simultaneous action of the hip flexors. The sartorius may be examined by having the patient attempt to flex the knee against resistance with the hip flexed and rotated laterally ([Figure 27.46](#)).

The quadriceps femoris (femoral nerve, L2-L4) is the primary knee extensor. It is composed of four large muscles, rectus femoris, vastus lateralis, vastus medialis, and vastus intermedius, which are united into a common tendon inserted into the upper border of the patella. The quadriceps is very powerful. It is capable of generating as much as 1,000 lb of force—three times more than the hamstrings. The rectus femoris originates from the ilium and runs straight down the middle of the thigh. The other three muscles originate from the shaft of the femur and only cross the knee joint. Because the rectus femoris also crosses the hip, it serves as a hip flexor as well as a knee extensor. The vastus medialis is sometimes divided into two parts: vastus medialis longus and vastus medialis oblique. Although the oblique head is often examined electromyographically, it in fact is the only portion of the quadriceps that is incapable of extending the knee.



FIGURE 27.45 Examination of flexion at the knee. The prone patient attempts to maintain flexion of the leg while the examiner attempts to extend it; the tendon of the biceps femoris can be palpated laterally and the tendons of the semimembranosus and semitendinosus medially.



FIGURE 27.46 Examination of the sartorius. With the thigh flexed and rotated laterally and the knee moderately flexed, the patient attempts further flexion of the knee against resistance.

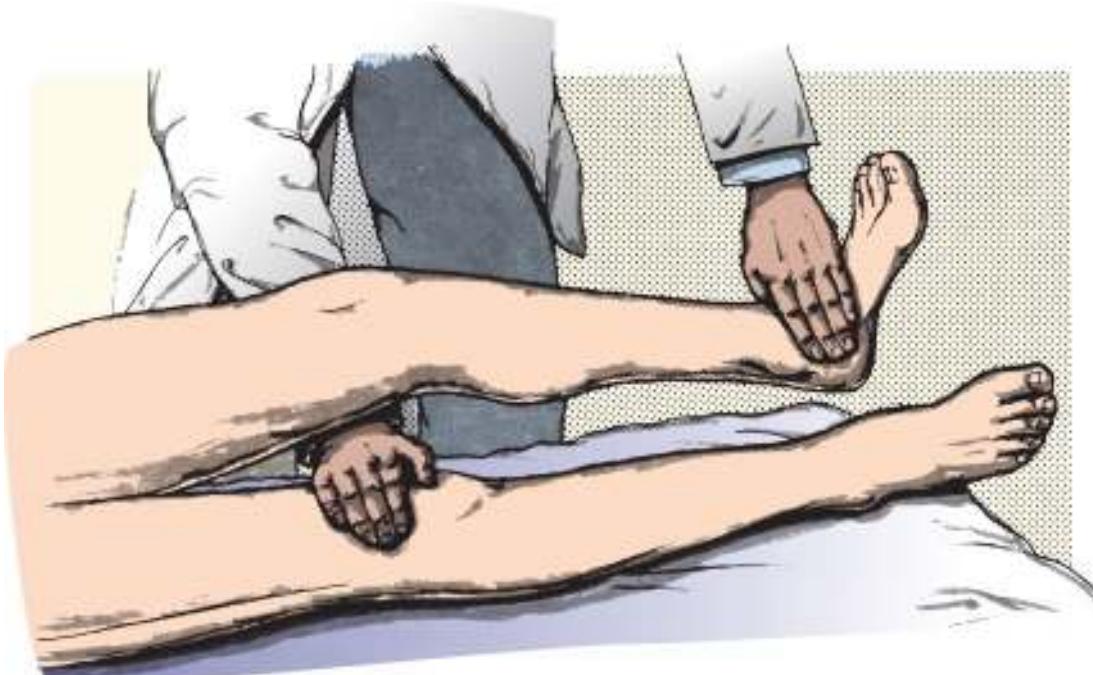


FIGURE 27.47 Examination of extension of the leg at the knee. The supine patient attempts to extend the leg at the knee against resistance; contraction of the

quadriceps femoris can be seen and palpated.

The quadriceps may be tested when the patient, sitting or supine, attempts to extend the knee against the examiner's resistance ([Figure 27.47](#)). The quadriceps is so powerful it is nearly impossible to overcome in the normal adolescent or adult except by taking extreme mechanical advantage. A sometimes useful technique for testing knee extension is the "barkeeper's hold," a hold usually applied to the elbow to control unruly patrons. To examine the right quadriceps, the examiner, standing on the outer aspect of the knee and reaching around from the inner aspect, places his left elbow, forearm pronated, beneath the patient's flexed knee, puts his right hand as far down on the shin as possible, and then grips his right forearm with his left hand, locking the patient's leg in a vice from front and back. The examiner then pulls upward with the elbow while pushing downward with the hand as the patient tries to extend his knee ([Figure 27.48](#)).

With severe quadriceps weakness, the sitting patient may lean backward when trying to extend the knee, attempting to muster some knee extension force by allowing the rectus femoris to contract across the hip. The patient will have marked difficulty in rising from a kneeling position and in climbing stairs; he can walk backward but has difficulty walking forward.

The Ankle Joint

Movements about the ankle joint are plantar flexion, dorsiflexion, eversion, and inversion. Plantar flexion (flexion) of the foot is carried out principally by the gastrocnemius and soleus muscles (gastrosOLEUS, triceps surae). Other muscles cross posterior to the axis of rotation of the ankle but because of mechanical factors are not very effective plantar flexors. The gastrocnemius also assists in flexing the knee. The gastrosOLEUS raises the heel, as in walking, and inverts the foot. The calf muscles are innervated by the tibial nerve (S1-S2).

The function of these muscles is tested manually by having the patient plantar flex the ankle as the examiner offers resistance by pressure against the sole of the foot ([Figure 27.49](#)). The plantar flexors of the ankle are among the most powerful muscles in the body. They cannot normally be defeated by hand and arm strength alone, even when the examiner takes maximal mechanical advantage. A helpful technique is to use the forearm as a lever by grasping the patient's heel with the hand and pushing against the ball of the foot with the volar forearm. Normal plantar flexors will hold fast even against this power

move. A better test of plantar flexor strength is to have the patient stand on tiptoe. Normally, a patient can easily support the entire body weight on one tiptoe, hop on one foot, and even do multiple toe raises on one foot. The number and ease of toe raise repetitions on the two sides may be compared to detect subtle weakness, as in S1 radiculopathy.



FIGURE 27.48 The “barkeeper’s hold,” a powerful move against the quadriceps. (From Wolf JK. *Segmental Neurology*. Baltimore: University Park Press, 1981. Reprinted with permission from Dr. John K. Wolf.)



FIGURE 27.49 Examination of plantar flexion of the foot. The patient attempts to plantar flex the foot at the ankle joint against resistance; contraction of the gastrocnemius and associated muscles can be seen and palpated.

Dorsiflexion (extension) of the ankle is carried out primarily by the tibialis anterior muscle, assisted by the extensor digitorum longus (EDL) and extensor hallucis longus (EHL). The tibialis anterior is supplied by the deep peroneal nerve (L4-L5). It also functions as an invertor, more so when the ankle is dorsiflexed. When the foot is planted on the ground, the tibialis anterior tilts the lower leg forward, as in walking. The foot dorsiflexors are tested by having the patient pull the foot up against the examiner's resistance ([Figure 27.50](#)). The dorsiflexors are powerful and cannot normally be overcome, even with maximal effort from the examiner. Subtle weakness can sometimes be detected by placing the patient at maximal mechanical disadvantage with the foot plantar flexed and trying to hold the foot in that position as the patient tries to dorsiflex it. This technique is most useful with unilateral weakness when the two sides can be compared. Dorsiflexion may also be tested by having the patient stand on the heels, raising the toes as high as possible. The toes on the weak side cannot be lifted as far. The tibialis anterior is the major muscle innervated by the L5 myotome; L5 radiculopathy and peroneal neuropathy are the most common causes of weakness.

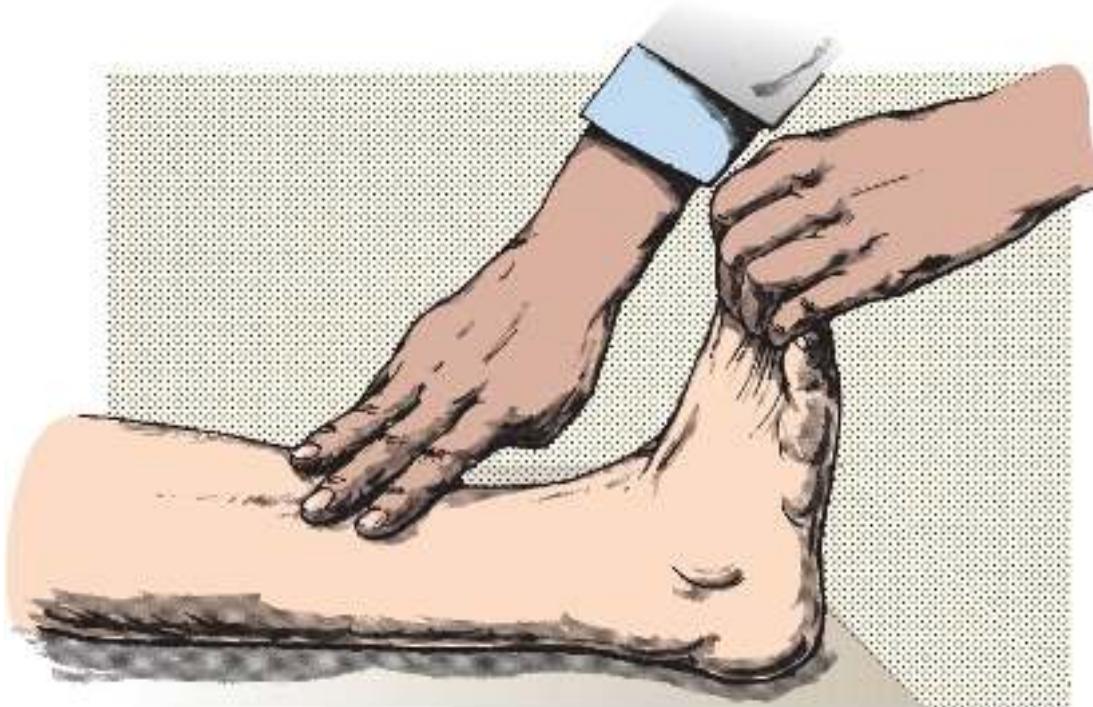


FIGURE 27.50 Examination of dorsiflexion (extension) of the foot. The patient attempts to dorsiflex the foot against resistance; contraction of the tibialis anterior can be seen and palpated.

With severe weakness of dorsiflexion, there is a footdrop. The patient walks with a “steppage gait,” raising the affected leg higher off the ground by exaggerated flexion at the hip and knee, to permit the toes to clear during the stride phase (see [Video Link 27.5](#)). In normal walking, heel strike occurs first. With a footdrop, there may be an audible double slap as the toes contact the floor first, followed by the heel. There may be inability to raise the forefoot off the ground while trying to stand on the heel. The term footdrop is sometimes applied to any degree of dorsiflexion weakness, even when too mild to cause a steppage gait. The term steppage gait is also used to refer to the high-stepping, drum-major gait of a patient with sensory ataxia as he slams the feet to the ground to reinforce proprioception ([Chapter 44](#)).

Inversion at the ankle is elevation of the inner border of the foot to turn the sole medially. Several muscles can perform this action with different degrees of efficiency, determined in part by whether the ankle is dorsi- or plantar flexed. The tibialis posterior (tibial nerve, L5-S1), the strongest invertor, is also a plantar flexor and is strongest as an invertor when the ankle is plantar flexed. The tibialis anterior functions as an invertor when the ankle is dorsiflexed. Inversion is tested by having the patient attempt to invert the ankle against resistance

(Figure 27.51). Weakness of ankle inversion is a key clinical sign indicating that a footdrop is due to L5 radiculopathy and not peroneal neuropathy at the knee.

Eversion, or lateral deviation, is elevation of the outer border of the foot to turn the sole laterally. This movement is carried out by the peroneus longus, brevis, and tertius and the EDL. The peronei are supplied by spinal segments L4-L5 and S1, the longus and brevis through the superficial peroneal nerve, and the tertius through the deep peroneal nerve. To test these muscles, the patient attempts to evert the ankle against resistance applied to the lateral border of the foot (Figure 27.52).

The evertors and invertors of the ankle are very important in ankle stability. When these muscles are weak, the patient is susceptible to ankle sprains; an early symptom of ankle instability is difficulty walking over rough or uneven terrain.



FIGURE 27.51 Examination of inversion of the foot. The patient attempts to raise the inner border of the foot against resistance; the tendon of the tibialis posterior can be seen and palpated just behind the medial malleolus.

Muscles of the Foot and Toes

The function of individual foot and toe muscles is not as clearly defined as in the hand, and muscle testing cannot be carried out with as much detail. The principal movements are extension (dorsiflexion) and flexion (plantar flexion) of the toes. With plantar flexion, there is cupping of the sole. Abduction and adduction of the toes are minimal.



FIGURE 27.52 Examination of eversion of the foot. The patient attempts to raise the outer border of the foot against resistance; the tendons of the peronei longus and brevis can be seen and palpated just above and behind the lateral malleolus.

The toe extensors are the extensor digitorum longus (EDL) and extensor digitorum brevis (EDB) and the extensor hallucis longus (EHL) and extensor hallucis brevis (EHB). These muscles are all supplied by the deep peroneal nerve (EHL, L5; EDL and EDB, L5-S1).

The long toe extensors extend the metatarsophalangeal (MTP) and IP joints

and dorsiflex the ankle joint. The EDL is also an evertor. The EDB aids the EDL in extending the four medial toes. Dorsiflexion of the toes against resistance may be used as a test for the function of these muscles. The tendons of the long extensors and the belly of the EDB can be palpated during this maneuver ([Figure 27.53](#)). The EDB normally forms a prominent bulge on the dorsolateral aspect of the foot. Its most medial and largest belly is the EHB. The EDB is the muscle used for recording the compound muscle action potential when performing peroneal nerve conduction studies. The EDB atrophies easily in neurogenic processes, and its bulge may disappear in patients with radiculopathy or peripheral neuropathy. Preservation of EDB bulk in the face of foot dorsiflexion weakness suggests a myopathic process (e.g., distal myopathy, scapuloperoneal dystrophy, FSH dystrophy). Weakness of the EHL is a key clinical sign of L5 radiculopathy; it is sometimes the only weak muscle. In testing for subtle weakness between the two sides, a useful technique is for the examiner to test the muscle using a relatively weak hand muscle, such as the first DI or the ADM. Either of these muscles may be able to overcome the EHL on the weak side but not the normal side. When the EHL is severely weak in the absence of severe weakness of the other foot and toe extensors, the patient may have a “toe drop” rather than a “footdrop.”



FIGURE 27.53 Examination of dorsiflexion (extension) of the toes. On attempts to dorsiflex the toes against resistance, the tendons of the extensors digitorum and hallucis longus and the belly of the extensor digitorum brevis can be seen and palpated.



FIGURE 27.54 Examination of flexion of the toes. The patient attempts to flex the toes against resistance.

Flexion of the toes is carried out by the flexors digitorum and hallucis longus, flexors digitorum and hallucis brevis, and some of the intrinsic muscles of the sole of the foot. These muscles are tested by having the patient flex the toes against resistance ([Figure 27.54](#)). The long toe flexors are calf muscles, innervated by the tibial nerve (L5, S1-S2). These muscles flex the phalanges of all five toes, acting chiefly at the distal IP joints. They also plantar flex the ankle joint and invert the foot. The short toe flexors (medial plantar nerve, S1-S2) act at the proximal IP and MTP joints. Testing of the intrinsic muscles of the sole of the foot is difficult and not clinically useful. Abduction and adduction are

extremely weak movements, and the short toe flexors are more powerful than the other intrinsic muscles. These muscles may be tested together by asking the patient to cup the sole of the foot ([Figure 27.55](#)). Most of the intrinsic foot muscles are more important in maintaining the longitudinal arches of the foot than in moving the toes.

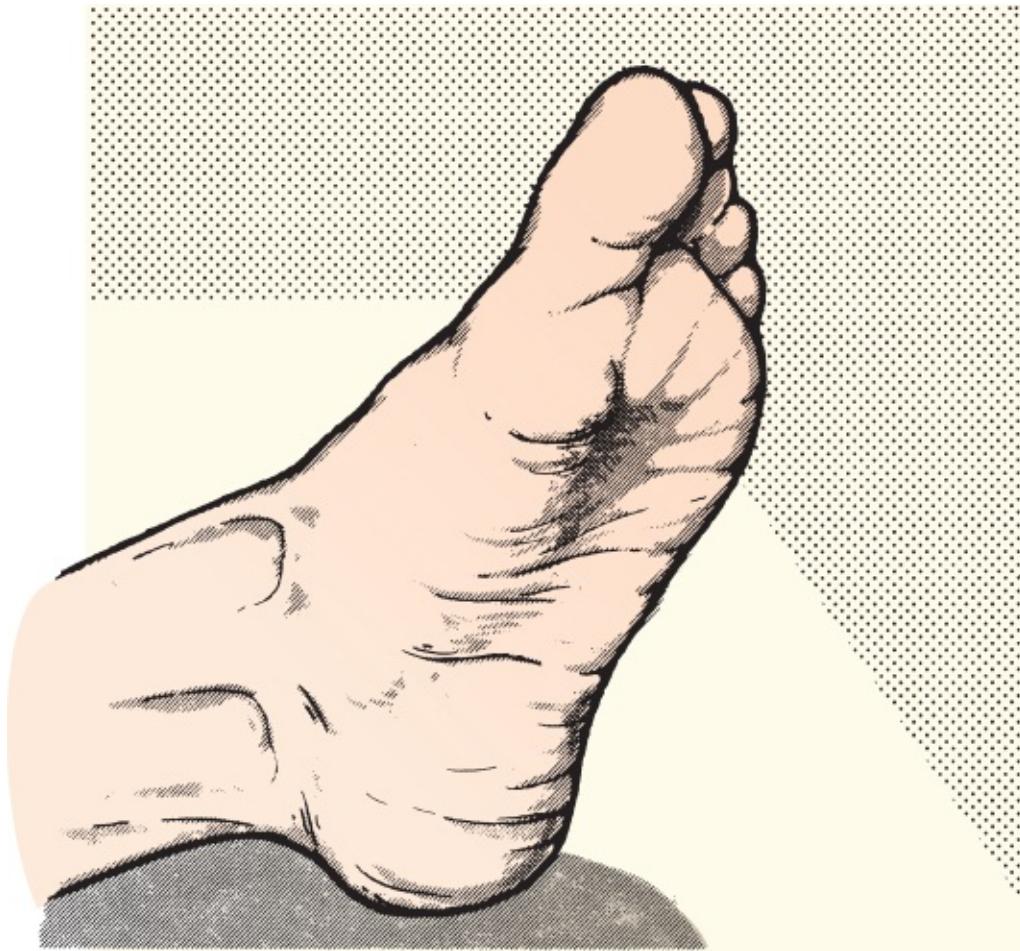


FIGURE 27.55 Cupping of the sole of the foot.

EXAMINATION FOR SUBTLE HEMIPARESIS

The motor examination is not concluded just with the formal strength assessment. Patients with mild CST lesions may have normal strength to routine testing, but the neurologic deficit may be brought out using ancillary maneuvers. The most important of these is the examination for pronator drift (Barré's sign). With the patient's upper extremities outstretched to the front, palms up and with the eyes closed, observe the position of each extremity ([Figure 27.56](#)). The

patient should hold this position for at least 20 to 30 seconds. In normals, the palms will remain flat, the elbows straight, and the limbs horizontal. Any deviation from this position will be similar on the two sides. One exception to the usual symmetry is that the dominant hand occasionally may pronate slightly more than the nondominant, perhaps because the nondominant extremities tend to be more flexible than the dominant extremities, making it more difficult to stretch the dominant hand to a horizontal position. Slight pronation, without downward drift, of the dominant arm (pseudodrift) due to decreased flexibility is not necessarily abnormal and must be interpreted in clinical context. However, greater pronation of the nondominant arm is sometimes an indication of subtle hemiparesis. Except for pseudodrift and for coincidental orthopedic or musculoskeletal problems, there should be no difference between the positions of the two limbs.

The patient with a mild CST deficit may demonstrate “pronator drift” to varying degrees ([Video 27.2](#)). With mild drift, there is slight pronation of the hand and slight flexion of the elbow on the abnormal side. With more severe drift, there is more prominent pronation and obvious flexion of the elbow, and there may be downward drift of the entire arm ([Figure 27.57](#)). Because of the innervation pattern of the CST, the minimally weak CST innervated muscles are overcome by the non-CST muscles. With a mild CST lesion, the minimally weak muscles in the upper extremity are the extensors, supinators, and abductors. These are overcome by the uninvolved and therefore stronger muscles: the pronators, biceps, and internal rotators of the shoulder. As these overcome the slightly weakened CST innervated muscles, the hand pronates, the elbow flexes, and the arm drifts downward. The tendency to pronation and flexion in mild hemiparesis has also been attributed to subtle hypertonicity in the pronator and flexor muscle groups. Imagine what would occur if this motion continued to the extreme: the hand would become hyperpronated, the elbow fully flexed, and the shoulder internally rotated, that is, the position of spastic hemiparesis ([Figure 27.58](#)). The abnormal upper limb positions in minimal pronator drift and in severe spastic hemiparesis are due to the same underlying phenomenon: strong non-CST muscles overcome variably weak CST muscles involved by the disease process. Another sign occasionally useful is the *digitus quinti* sign (see [Video Link 27.6](#)). With the hands outstretched in drift position, the small finger on the hemiparetic side may be abducted more than on the normal side.



Video 27.2 Discussion of the examination for pronator drift and its underlying pathophysiology.



FIGURE 27.56 Technique for testing for pronator drift. In the presence of a corticospinal tract (CST) lesion, the selectively weakened muscles are the shoulder abductors and external rotators, the supinators, and the elbow extensors. These muscles are overcome by their antagonists to cause pronation, elbow flexion, and downward drift. This is an illustration of mild pronator drift of the right upper extremity. Patients with mild CST lesions may demonstrate a pronator drift or have an abnormal arm or finger roll test in the absence of clinically detectable weakness to formal strength testing.

Hachinski recently described the upgoing thumb sign as a subtle clinical

finding in minor stroke. The thumb is extended (radially abducted) but with the palms facing it is upgoing. Souques described the “interosseous phenomenon” in 1907, in which elevation of a paretic arm caused abduction of all the fingers. The interosseous phenomenon, or finger sign, is usually classified as an associated movement ([Chapter 42](#)). Several subtle upper motor neuron signs that involve finger abduction are probably all related to the interosseous phenomenon originally described by Souques, including the digitus quinti sign; the upgoing thumb sign; myelopathy hand, a hand posture seen in patients with cervical spondylotic myelopathy; and the finger escape sign, widely cited in Orthopedic and Neurosurgery textbooks as a sign of cervical spondylotic myelopathy.

The examination for pronator drift is a very important part of the neurologic examination. If only one motor test could be done on a patient, the best single test to use would probably be examining for drift. While waiting for drift to occur, because it is not instantaneous, the examiner may simply wait or hasten the development of drift by tapping on the palms or having the patient turn the head back and forth or both. The examination for drift is often combined with the Romberg test because both require the patient to have the eyes closed.



FIGURE 27.57 Moderate drift with further development of the posture.



FIGURE 27.58 Further development of pronator drift, with the evolution of severe drift to show how marked weakness of the corticospinal innervated muscles produces the posture of spastic hemiparesis. The pathophysiologic basis for pronator drift and for the upper-extremity posture of fully developed spastic hemiparesis and for the upper-extremity posture of decorticate rigidity is the same; it is only a matter of degree. A mild CST lesion results in mild pronator drift; a severe lesion results in spastic hemiparesis.



FIGURE 27.59 Updrift due to a parietal lobe lesion with loss of position sense.

Abnormal drift can occasionally occur with lesions elsewhere in the nervous system. Cerebellar disease may cause drift to some degree, but the movement is outward and usually slightly upward. In parietal lobe lesions, there may be “updrift,” with the involved arm rising overhead without the patient’s awareness, ostensibly because of loss of position sense (Figure 27.59). Additional pronation phenomena have been described by Babinski and Wilson. In the former, the palmar aspects of the hands are held in approximation with the thumbs up and are then jarred or shaken; the paretic hand falls into a position of pronation. In the latter, there is pronation of the forearm along with internal rotation at the shoulder when the arms are held overhead, palms facing; as a result, the affected palm turns outward. Pronation may also occur on the paretic side when the arms are actively abducted with the forearm supinated or when the arms are passively abducted with the forearm supinated and then suddenly released. Downward drift without pronation has been reported as a sign of nonorganic weakness.

Similar procedures can be used to detect lower-extremity weakness. Examination for leg drift is possible (see “The Hip Joint”) but is not nearly as

useful as testing for arm drift and therefore seldom done. Checking for a decreased rate of foot tapping on the involved side is simpler and more useful. In the leg or knee-dropping test, the patient lies supine with the hips and knees flexed, the knees forming an angle of about 45 degrees, and heels resting on the table. When a CST lesion is present, the affected heel will gradually slide downward so that the knee slowly extends, and the hip goes into extension, external rotation, and abduction.

Other useful maneuvers include examination of forearm roll, finger roll, thumb roll, and rapid alternating movements ([Video 27.3](#)). Abnormal forearm rolling is a sensitive indicator of neurologic pathology. To test it, the patient is instructed to make fists, to hold the forearms horizontally so that the fists and distal forearms overlap with the palms pointed more or less toward the umbilicus, and then to rotate the fists around each other, first in one direction and then the other ([Figure 27.60](#)).



Video 27.3 Other subtle signs of hemiparesis, including forearm and finger rolling.

Normal patients will have about an equal excursion of both forearms so that the fists and forearms roll about each other symmetrically. With a unilateral corticospinal lesion, the involved side does not move as much as the normal side, so the patient will appear to plant, fix, or “post” one forearm and to rotate the opposite forearm around it ([Video 27.4](#)). Finger roll is an even more sensitive version of the same test. The patient is asked to rotate the extended forefingers around each other, moving just the fingers. Again, the finger on the abnormal side will move less than its fellow. In the thumb rolling test, the patient rotates each thumb around its fellow. Forearm rolling has a sensitivity of 17% to

87%, specificity of 97% to 98%, positive LR of 15.6, and negative LR of 0.6 in the detection of contralateral hemispheric disease. Index finger rolling has a sensitivity of 33% to 42%, a specificity of 92% to 98%, positive LR of 6.0, and negative LR of 0.7 in the detection of contralateral hemispheric disease. In a series of patients with mild hemiparesis, thumb rolling was more sensitive (88%) than pronator drift (47%), forearm rolling (65%), or index finger rolling (65%). Patients with bradykinesia or rigidity from extrapyramidal disease may also show decreased excursion of the affected limbs.



Video 27.4 Abnormal forearm rolling in a patient with a left hemiparesis.



FIGURE 27.60 Testing for a CST lesion using arm roll. The involved extremity tends

to have a lesser excursion as the forearms roll about each other so that the normal extremity tends to rotate around the abnormal extremity, which tends to remain relatively fixed ("posted"). Patients with mild CST lesions may have an abnormal arm roll test in the absence of clinically detectable weakness to formal strength testing.

Normal fine motor control requires functional integrity of both the CST and the cerebellum. Testing for rapid alternating movements is part of the cerebellar examination, but the primary function of the CST is to provide discrete, fractionated movements to the distal extremities. Either CST or cerebellar disease may interfere with fine motor control of distal muscles. Normal fine motor control also requires intact proprioceptive pathways. Traditionally, different tests have been done to look for CST signs than for cerebellar signs, but both involve rapid alternating movements. This test is also referred to as assessment of alternate motion rate, but in fact more than the rate of motion provides useful information. Fine motor control can be tested in numerous ways, most advantageously by comparing the dexterity and precision of the two hands while performing rapid, repetitive movements, making allowance of course for hand dominance. The patient may be asked to repetitively, and as quickly as possible, touch the tip of the index finger to the tip of the thumb, as in making the OK sign. Any finger can be used, but the index and small fingers are favorites. The movements will be slower and less agile on the abnormal side. This test is often done by having the patient touch the IP joint rather than the tip of the thumb. Rough quantitation can be done by counting the number of touches in a set period of time. Other tests requiring a high level of coordination include quickly touching the tip of each finger in turn to the thumb, flicking the fingers as if flicking off water, doing one-handed clapping, and making quick, small finger movements as if playing a piano. Patients with extrapyramidal disorders, especially parkinsonism, may show more of a decrease in the amplitude than the rate of motion, especially if the task is continued for more than a few seconds. Fine motor control of the foot can be assessed by having the patient do rapid, repetitive foot taps on the floor if standing and against the examiner's palm if supine.

Abnormalities of associated movements may also occur relatively early in a CST lesion. These may include the absence of a normal associated movement, such as a decreased arm swing while walking, or the presence of an abnormal associated movement, such as Wartenberg's thumb adduction sign, the Babinski trunk-thigh sign, or the anterior tibial sign ([Chapter 42](#)).

Video Links

Video Link 27.1. Scapular winging. <http://www.youtube.com/watch?v=dfTe0nPclDE>

Video Link 27.2. Pectoral crease in shoulder girdle weakness.
http://neurosigns.org/wiki/Pectoral_creature

Video Link 27.3. Beevor's sign. http://neurosigns.org/wiki/Beevor%27s_sign

Video Link 27.4. Gowers' sign. http://neurosigns.org/wiki/Gower%27s_sign

Video Link 27.5. Steppage gait. http://neurosigns.org/wiki/Steppage_Gait

Video Link 27.6. Digitus quinti sign. http://neurosigns.org/wiki/Digitus_quinti_sign

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CHAPTER 28

Muscle Tone

Muscle tone has been defined as the tension in the relaxed muscle or the resistance to passive movement when voluntary contraction is absent. Because of resting tone, normal muscles have slight resistance to passive movement even in the relaxed state. The inherent attributes of muscle tissue—such as viscosity, elasticity, and extensibility—contribute to resting tone. Even apparently relaxed muscle fibers have a constant slight fixed tension by which they hold their resting position, resist changes in length, prevent undue mobility at joints, and are in position to contract when necessary. Resting muscle tone is greatest in the antigravity muscles that maintain the body in an erect position.

The resting level of tone in a muscle is dependent on activity in the spinal cord segment that innervates it, primarily the gamma motor neuron. Efferent impulses from the gamma motor neuron set the level of contraction of the intrafusal fibers of the muscle spindles. Spindle afferents in turn convey impulses to the spinal cord segment to complete the gamma loop. Descending influences from higher motor centers regulate and modulate the activity at the local spinal cord segment. All of these factors interact to determine the level of resting tone. When a muscle with normal segmental innervation is passively stretched, reflex shortening may occur; this is the stretch reflex.

The background level of muscle tone maintains normal resting limb positions and attitudes. Active muscle contraction takes place on the background of the resting level of muscle tone, and normal background tone is important for proper coordination of movement. Activity mediated by the reticular formation, the otolith organs, the vestibular apparatus, and other higher centers is important in maintaining the steady contraction of the antigravity muscles that is necessary to the standing position, as well as to other postural and righting reflexes.

Tone may be affected by disease at different levels of the nervous system. Interruption of the local spinal reflex arc abolishes resting muscle tone. Most

types of hypertonicity can be abolished by interrupting either the gamma efferent impulses to the intrafusal fibers or the afferent impulses from the muscle spindles. Denervated muscle is flaccid and behaves as noncontractile tissue. Loss of impulses from the supraspinal pathways that normally inhibit lower reflex centers usually causes an increase in tone. Loss of the normal balance between higher facilitatory and inhibitory centers may either decrease or increase tone.

EXAMINATION OF TONE

Tone is difficult to assess. The determination of tone is subjective and prone to interexaminer variability. There are no methods that can measure tone quantitatively. The determination is based solely on the clinical judgment of the examiner; accurate assessment of tone requires clinical experience. It is difficult to separate slightly increased tone from poor relaxation in a tense or apprehensive patient. Tone is especially difficult to evaluate in infants, where there may be wide variations in apparent tone on different examinations, in either health or disease.

The examination of tone requires a relaxed and cooperative patient. Small talk may help the patient relax. Simple observation may reveal an abnormality of posture or resting position that indicates an underlying change in tone. Muscle palpation is sometimes useful, but well-muscled individuals may have firm muscles despite normal resting tone, whereas in other individuals, the muscles may feel flabby despite an underlying hypertonicity. Muscles may have a firm consistency to palpation because of edema, inflammation, spasm due to pain, or pseudohypertrophy.

The most important part of the examination of tone is determination of the resistance of relaxed muscles to passive manipulation as well as the extensibility, flexibility, and range of motion. Abnormalities of tone are more easily detected in extremity than in trunk muscles. The limb is moved passively, first slowly and through a complete range of motion and then at varying speeds. The examiner may shake the forearm to and fro and note the excursions of the patient's hand, brace a limb and then suddenly remove the support, or note the range of movement of a part in response to a slight blow. Bilateral examination of homologous parts helps compare for differences in tone on the two sides of the body.

Tone should be assessed by both slow and rapid motion and through partial

and full range of motion, documenting the distribution, type, and severity of any abnormality. Certain specific maneuvers may be helpful in evaluation of abnormal tone.

The Babinski Tonus Test

The arms are abducted at the shoulders, and the forearms are passively flexed at the elbows. With hypotonicity, there is increased flexibility and mobility, and the elbows can be bent to an angle more acute than normal. With hypertonicity, there is reduced flexibility, and passive flexion cannot be carried out beyond an obtuse angle.

The Head-Dropping Test

The patient lies supine without a pillow, completely relaxed, eyes closed, and attention diverted. The examiner places one hand under the patient's occiput and with the other hand briskly raises the head and then allows it to drop. Normally, the head drops rapidly into the examiner's protecting hand, but in patients with extrapyramidal rigidity, there is delayed, slow, gentle dropping of the head because of rigidity affecting the flexor muscles of the neck. When meningismus is present, there is resistance to and pain on flexion of the neck.

Pendulousness of the Legs

The patient sits on the edge of a table, relaxed with legs hanging freely. The examiner either extends both legs to the same horizontal level and then releases them (Wartenberg pendulum test, see [Video Link 28.1](#)) or gives both legs a brisk, equal backward push. If the patient is completely relaxed and cooperative, there will normally be a swinging of the legs that progressively diminishes in range and usually disappears after six or seven oscillations. In extrapyramidal rigidity, there is a decrease in swing time but usually no qualitative change in the response. In spasticity, there may be little or no decrease in swing time, but the movements are jerky and irregular, the forward movement may be greater and more brisk than the backward, and the movement may assume a zigzag pattern. In hypotonia, the response is increased in range and prolonged beyond the normal. In all of these maneuvers, a unilateral abnormality will be more apparent.

The Shoulder-Shaking Test

The examiner places her hands on the patient's shoulders and shakes them briskly back and forth, observing the reciprocal motion of the arms. With extrapyramidal disease, there will be a decreased range of arm swing on the affected side. With hypotonia, especially that associated with cerebellar disease, the excursions of the arm swing will be greater than normal.

The Arm-Dropping Test

The patient's arms are briskly raised to shoulder level and then dropped. In spasticity, there is a delay in the downward movement of the affected arm, causing it to hang up briefly on the affected side (Bechterew's or Bekhterew's sign); with hypotonicity, the dropping is more abrupt than normal. A similar maneuver may be carried out by lifting and then dropping the extended legs of the recumbent patient.

Hand Position

Hypotonicity, especially that associated with cerebellar disease or Sydenham's chorea, may cause the hands to assume a characteristic posture. With the arms and hands outstretched, there is flexion at the wrists and hyperextension of the fingers ("spooning") accompanied by moderate overpronation. With the arms raised overhead, the overpronation is exaggerated with the palms turned outward. This overpronation phenomenon differs from the pronator drift sign, in which the overpronation is due to weakness of corticospinal innervated muscles or increased tone in the pronator muscles.

MYOTATIC IRRITABILITY, MYOEDEMA, AND TENDERNESS

In addition to the inspection, palpation, and resistance to passive motion used in the assessment of tone, it is sometimes useful to observe the reaction to direct percussion of the muscle belly. The idiomuscular contraction is the brief and feeble contraction of a muscle belly after it is tapped with a percussion hammer, causing a slight depression even when the deep tendon reflex (DTR) is absent.

Myotactic irritability has been defined as both the response to direct percussion as well as the ability of a muscle to contract in response to sudden stretch.

The response to direct muscle percussion in normal muscle is very slight and, in most muscles, is seen or felt with difficulty. The reaction may be more pronounced in wasting diseases, such as cachexia and emaciation, and in some diseases of the lower motor neuron. Hyperexcitability to such stimulation occurs in tetanus, tetany, and certain electrolyte disturbances. Occasionally, after a muscle is percussed with a reflex hammer, a wave of contraction radiates along the muscle away from the point of percussion. A small ridge or temporary swelling may persist for several seconds at the point of stimulation. This stationary muscle mounding is known as myoedema. There is no accompanying electrical muscle activity. The idiomuscular contraction causes a slight depression, myoedema a rounding up. The mechanism of myoedema is poorly understood, but it is probably a normal physiologic phenomenon. Its presence alone does not indicate a neuromuscular disorder, but the response may be exaggerated in some circumstances, most notably hypothyroid myopathy and cachexia. Myoedema is electrically silent on electromyography. Hypothyroidism may also cause an electrically active muscle mounding and spreading contraction, manifest by a burst of normal motor unit action potentials upon percussion (for video, see Loomis et al.). Myotonia is a persisting contraction following mechanical stimulation of muscle that is quite different from myoedema (see below). In rippling muscle disease, there are wave-like muscle contractions evoked by muscle stretch that move laterally along the muscle over 5 to 20 seconds. The phenomenon is especially prominent in large proximal muscles (see [Video Link 28.2](#)).

During muscle palpation, muscle tenderness may sometimes be elicited. Muscle tenderness on squeezing the muscle belly, or even with very slight pressure, may cause exquisite pain. Widespread muscle tenderness to palpation may occur with inflammatory myopathy, especially polymyositis and dermatomyositis, in some neuropathies, and in acute poliomyelitis. Focal muscle tenderness occurs with trauma or overexertion of muscles.

ABNORMALITIES OF TONE

Pathologic conditions may cause an increase or decrease in tone. In addition, there are different varieties of hypotonicity and hypertonicity. Hypotonicity may

develop from disease of the motor unit, the proprioceptive pathways, cerebellar lesions and in the choreas. The muscle may be flaccid, flabby, and soft to palpation. The involved joints offer decreased resistance to passive movement. The excursion of the joint may be increased with an absence of the normal “checking” action on extreme passive motion. If the involved extremity is lifted and allowed to drop, it falls abruptly. A slight blow causes it to sway through an excessive excursion. The DTRs are usually decreased or absent when hypotonia is due to a lesion involving the motor unit or proprioceptive pathways.

Hypotonia

When hypotonia is due to disease of the motor unit, there is invariably some degree of accompanying weakness. The hypotonia that results from central processes (e.g., cerebellar disease) does not cause weakness; muscle power is preserved even though hypotonia is demonstrable on examination. Infantile hypotonia (floppy baby syndrome) is a common clinical condition in which there is a generalized decrease in muscle tone, typically affecting a neonate. There are numerous causes, both central and peripheral.

Tone may also be decreased when disease affects the muscle spindle afferent system. Tabes dorsalis affects proprioceptive fibers in the posterior root and may cause muscle hypotonia with joint hyperextensibility. Hypotonia may occur with some lesions of the parietal lobe, probably because of disturbances of sensation. Hypotonicity may occur with various types of cerebellar disease but is never as severe as that which occurs with diseases of the lower motor neuron. Cerebellar hypotonia is not associated with weakness and the reflexes are not lost, although they may be pendular; there are no pathologic reflexes. Muscle tone is, of course, decreased in deep sleep, coma, and other states of impaired consciousness.

Sudden attacks of impaired muscle tone in an awake patient occur in akinetic epilepsy and in cataplexy. With atonic (akinetic) seizures, the attacks of sudden loss of muscle tone occur spontaneously, and the patient may fall to the ground (drop attack or drop seizure). Less severe attacks may cause only a head drop. In cataplexy, there are attacks of decreased tone after strong emotion, such as laughter or anger. With severe attacks, the patient falls to the ground but without loss of consciousness. With incomplete attacks, there may be slackening of facial muscles, jaw drop, head drop, slumping of the shoulders, or knee buckling without a fall. Cataplexy is usually a component of narcolepsy. Sleep paralysis is

a state common in narcolepsy, in which a patient has diffusely decreased tone and is unable to move immediately after awakening from sleep. The hemiparesis that is present acutely following hemispheric stroke may be associated with hypotonia (cerebral or neural “shock”), which gradually evolves into hypertonia with the passage of time. Some conditions may cause abnormal joint laxity, which may be confused with muscle hypotonia (e.g., Ehlers-Danlos syndrome).

Hypertonia

Hypertonia occurs under many circumstances. It is a routine feature of lesions that involve the corticospinal tract after the acute stage. It can occur with diffuse cerebral disorders, with disease involving the extrapyramidal system, with disease of spinal cord interneurons (e.g., stiff-person syndrome), and even with muscle disorders in continuous muscle fiber activity syndromes. The most common causes of hypertonia are spasticity and rigidity. For a discussion and demonstration of spasticity versus rigidity from the 2015 Stanford Medicine 25 Skills Symposium, see [Video Link 28.3](#).

Extrapyramidal Rigidity

Extrapyramidal rigidity is a diffuse increase in muscle tone to passive movement that occurs primarily with lesions that involve the basal ganglia. There is a fairly constant level of increased tone that affects both agonist and antagonist and is equally present throughout the range of motion at a given joint. Both flexor and extensor muscles are involved, with resistance to passive movement in all directions. The increased tone is equally present from the beginning to the end of the movement and does not vary with the speed of the movement. This type of rigidity is referred to as “lead-pipe.” The involved muscles may be firm and tense to palpation. After being placed in a new position, the part may remain there, causing the limbs to assume awkward postures. Both neural-mediated excitation of shortening muscles (the shortening reaction) and inhibition of stretched muscles contribute to the rigidity; which mechanism predominates is associated with the direction of movement. An increase in spinal interneuron excitability mediated through specific descending motor pathways may underlie parkinsonian rigidity.

In cogwheel rigidity, there is a jerky quality to the hypertonicity. As the part is manipulated, it seems to give way in a series of small steps as if the limb were

attached to a heavy cogwheel or ratchet. The jerky quality of the resistance may be due to tremor superimposed on lead-pipe rigidity. Cogwheel rigidity is most commonly encountered in Parkinson's disease and other parkinsonian syndromes. It appears first in proximal muscles and then spreads distally. Any muscle may be affected, but there is predominant involvement of neck and trunk muscles and the flexor muscles of the extremities. The rigidity of extrapyramidal disease may be brought out by the head-dropping, shoulder-shaking, and similar tests. The rigidity on one side may be exaggerated by active movements of the contralateral limbs (see [Chapter 30](#)).

In extrapyramidal disease, there is usually associated hypokinesia and bradykinesia, but no real paralysis. With repeated active movements, there is a gradual decrease in speed and amplitude. This may be brought out by having the patient rapidly open and close the eyes or mouth, open and close the hand, or oppose finger and thumb. Patients may also show slowness of starting and limitation of the amplitude of movement, loss of pendulousness of the arms and legs, inability to carry out rapid repeated movements or to maintain two simultaneous voluntary movements, and impairment of associated movements, such as swinging of the arms when walking.

Paratonia is an alteration in tone to passive motion that is often a manifestation of diffuse frontal lobe disease. It has been divided into inhibitory paratonia and facilitatory paratonia. Gegenhalten (inhibitory paratonia, paratonic rigidity) is a form of rigidity in which the resistance to passive movement seems proportional to the vigor with which the movement is attempted. The resistance of the patient increases in proportion to the examiner's efforts to move the part; the harder the examiner pushes, the harder the patient seems to push back. It seems as though the patient is actively fighting, but the response is involuntary. It is said that the severity of gegenhalten can be judged by the loudness of the examiner's exhortations to relax.

In the limb placement test, the examiner passively lifts the patient's arm, instructs the patient to relax, releases the arm, and notes whether or not it remains elevated. The arm remaining aloft, in the absence of parkinsonism or spasticity, indicates paratonia. In facilitatory paratonia (mitgehen), the patient cooperates too much. The patient actively assists the examiner's passive movements, and the limb may continue to move even after the examiner has released it. In the modified Kral procedure, the examiner instructs the seated patient to relax and then passively flexes and extends the elbow several times through a full range of motion, releasing the arm with the patient's hand at the

level of the thigh. Further movement is scored on a scale of 0 (no movement) to 4 (elbow flexes fully or cycles of flexion and extension continue).

Using a Delphi procedure, experts agreed on the following definition of paratonia: paratonia is a form of hypertonia with an involuntary variable resistance during passive movement; the nature of paratonia may vary from active assistance to active resistance; the degree of resistance depends on the speed of movement (e.g., slow → low resistance, fast → high resistance); the degree of paratonia is proportional to the amount of force applied; and the resistance to passive movement is in any direction and there is no clasp-knife phenomenon. The Paratonia Assessment Instrument is an assessment tool for paratonia. For a training video on paratonia from the Geriatric Medicine Research Unit at Dalhousie University, see [Video Link 28.4](#).

Spasticity

Spasticity is due to lesions involving the corticospinal pathways. The hypertonicity to passive movements differs from that of rigidity because it is not uniform throughout the range of movement, and it varies with the speed of movement. In addition, rigidity tends to affect all muscles to about the same degree, whereas the hypertonia of spasticity varies greatly from muscle to muscle. In spasticity, if the passive movement is made slowly, there may be little resistance. But if the movement is made quickly, there will be a sudden increase in tone partway through the arc, causing a catch or a block as though the muscle had impacted a stop. The relationship of the hypertonus to the speed of movement is a key feature distinguishing spasticity from rigidity. In the upper extremity, it is useful to look for spasticity involving the pronator muscles. With the patient's elbow flexed to about 90 degrees and the forearm fully pronated, the examiner slowly supinates the patient's hand. Unless spasticity is severe, there will be little or no resistance to this slow movement. If, after several slow repetitions, the examiner supinates the patient's hand very quickly, there will be sudden resistance at about the midrange of movement, referred to as a "pronator catch." The catch will then relax, and the supination movement can be completed. When hypertonus is severe, this maneuver may elicit pronator clonus.

A similar slow then rapid motion technique can be used to detect lower-extremity spasticity. With hands behind the knee, the examiner slowly flexes and extends the knee of the supine and relaxed patient. With adequate relaxation, the

foot remains on the bed. After several slow repetitions, from the position of full extension, the examiner abruptly and forcefully pulls the knee upward. When tone is normal, the foot will scoot back, remaining in contact with the bed. When there is spasticity, the foot flies upward in a kicking motion (spastic kick). In the heel- or foot-dropping test, the examiner holds the patient's leg flexed at the knee and hip, one hand behind the knee and the other supporting the foot. The foot is suddenly released. Normally, its descent is smooth, but when there is spasticity in the quadriceps muscle, the foot may hang up and drop in a succession of choppy movements.

Spastic muscles may or may not feel firm and tense to palpation. The range of movement of spastic extremities, and the degree of hypertonicity, often varies between examinations. No devices for quantitating spasticity exist, and clinical evaluation remains the most useful tool. The Ashworth scale is commonly used to quantitate spasticity on a scale from 1 (no increase in muscle tone) to 5 (affected part rigid in flexion or extension). Its validity and reliability have been questioned. In the presence of spasticity, the DTRs are exaggerated, and pathologic reflexes such as the Babinski and Chaddock signs can often be elicited. Clonus is often present. There may be abnormal associated movements.

Upper motor neuron weakness is often accompanied by sustained contraction of specific groups of muscles. With hemiparesis or hemiplegia, spasticity is most marked in the flexor and pronator muscles of the upper and the extensor muscles of the lower extremity; this causes a posture of flexion of the arm and extension of the leg, the characteristic distribution in cerebral hemiplegia (Figure 25.6). The arm is adducted, flexed at the elbow, and the wrist and fingers are flexed; there may be forced grasping. The lower extremity is extended at the hip, knee, and ankle, with inversion and plantar flexion of the foot; there may be marked spasm of the hip adductors. There is more passive resistance to extension than to flexion in the upper extremities and to flexion than to extension in the lower extremities. With bilateral lesions, the increased tone of the hip adductors causes a scissors gait, in which one leg is pulled toward the other as each step is taken (see [Chapter 44](#)). Although spasticity in the lower extremities usually affects the extensors most severely, in some patients with severe myelopathy or extensive cerebral lesions, there is marked hypertonicity in the flexor muscles, drawing the legs into a position referred to as paraplegia in flexion.

Catatonic Rigidity

The abnormal muscle tone in catatonia is in many respects similar to extrapyramidal rigidity and may be physiologically related. There is a waxy or lead-pipe type of resistance to passive movement that may be accompanied by posturing, bizarre mannerisms, and evidence of psychosis. It may be possible to mold the extremities into any position, in which they remain indefinitely. Catatonia may be induced by neuroleptics and may progress to neuroleptic malignant syndrome.

Decerebrate and Decorticate Rigidity

Decerebrate rigidity is characterized by marked rigidity and sustained contraction of the extensor muscles of all four extremities; in decorticate rigidity, there is flexion of the elbows and wrists with extension of the legs and feet. These are discussed further in [Chapter 41](#).

Similar generalized rigidity with neck extension can occur with severe meningismus (opisthotonus), as well as in the tonic phase of a generalized seizure. Cerebellar or posterior fossa fits are probably attacks of decerebrate rigidity due to brainstem dysfunction related to mass effect in the posterior fossa.

Voluntary Rigidity

Various muscle groups may be consciously tensed or braced to protect against injury or in response to pain. It is often difficult to differentiate between tension that is truly volitional and that which is unconscious or involuntary, especially when related to excitement, alarm, pain, or fatigue. Tense, apprehensive individuals may show increased muscular tension at all times and may have exaggerated tendon reflexes. The reflex exaggeration is one of range of response, and the latent period is not shortened. Conversely, the reflexes may be suppressed because the semivoluntary contraction prevents normal movement.

Involuntary Rigidity

Rigidity that is involuntary, reflex, or nonorganic may resemble voluntary rigidity. Rigidity of psychogenic origin may be bizarre and may simulate any type of hypertonicity. Hysterical rigidity may simulate decerebration or catatonia. It may be extreme, with neck retraction and opisthotonus, the body resting with only the head and heels upon the bed (arc de cercle, [Chapter 52](#)).

Reflex Rigidity

Muscles may develop reflex rigidity, or spasm, in response to afferent impulses, particularly pain. Muscle spasm is a state of sustained involuntary contraction accompanied by muscle shortening. The abnormal contraction is visible and palpable. Common examples of reflex muscle spasm are the board-like abdomen of acute abdominal disorders, rigidity of the neck and back in meningitis, and the localized spasm in the extremities following trauma. Reflex rigidity may follow other sensory stimuli, such as cold. Muscle contracture may follow prolonged spasm. In some metabolic myopathies (e.g., McArdle's disease), painful muscle cramps and spasms are brought on by exercise; the muscle cramp is a physiologic form of contracture due to abnormal metabolism and is not accompanied by electrical activity.

Myotonia

Myotonia is a disorder of the muscle membrane that can occur in many different conditions. Tone is usually normal when the muscles are relaxed, but contraction produces a temporary involuntary tonic perseveration of muscle contraction with slow relaxation ([Video 28.1](#)). Sudden movements may cause marked spasm and inability to relax. In grip myotonia, the patient has difficulty letting go of an object after gripping it strongly. The myotonia usually decreases with repetition of the movement. In rare instances, the myotonia increases with repetitive movement (paradoxical myotonia). Percussion myotonia is elicited by tapping on the muscle. Percussion over the thenar eminence produces a prolonged tonic abduction and opposition movement lasting several seconds, over which the patient has no control. Tapping over the extensor digitorum communis to the middle finger causes the finger to snap into extension, after which it slowly falls over a much longer period of time than normal. Percussion myotonia can also be elicited over other muscles. Oblique elimination with a penlight may help to make the slowly disappearing depression or dimple more visible. Percussion of a tongue blade placed transversely on edge across the tongue may produce a segmental myotonic contraction that constricts the tongue circumferentially (napkin-ring sign, [Chapter 20](#)).



Video 28.1 Video demonstrating grip and percussion myotonia in a patient with myotonic dystrophy, followed by eyelid, grip, and percussion myotonia with paradoxical myotonia in two patients with paramyotonia congenita. Paradoxical myotonia worsens with successive contractions. (Courtesy Dr. Richard Barohn.)

Other Types of Rigidity

Muscular rigidity may also occur in epilepsy, tetany, and tetanus. In epilepsy, there may be generalized rigidity during the tonic phase of the fit. Occasionally, there are tonic seizures with no clonic phase (tonic fits). In tetany, there is generalized irritability of the peripheral and central nervous systems, with tonic muscle spasms leading to localized or generalized hypertonicity, hypersensitivity to stimuli, cramps, and muscle twitching (see [Chapter 52](#)).

In tetanus, there is usually generalized rigidity with increased muscle tone in the entire body. In most instances, it begins in the face and jaw muscles and then spreads to affect the abdominal muscles, extremities, and spinal muscles, causing abdominal rigidity, extensor rigidity, and opisthotonus. In cephalic tetanus, disease manifestations occur primarily in head and neck muscles (for video, see You et al.). Both agonist and antagonist muscles are simultaneously hypertonic. Spasm of the muscles of mastication causes trismus (lockjaw). Retraction of the angles of the mouth causes the facial dystonia referred to as risus sardonicus. Paroxysms of muscle spasm progressively increase in intensity and propagate to other muscles. Spasms may occur spontaneously, after voluntary contraction or after mechanical, tactile, auditory, visual, or other stimuli. Between spasms, there is usually some persisting muscular rigidity. The reflexes are grossly exaggerated, and a light tap on a tendon may throw the limb into violent spasms. The clinical manifestations of tetanus are due to the action of the exotoxin of

Clostridium tetani on the inhibitory internuncial neurons of the brainstem and spinal cord. In the stiff-person (stiff-man) syndrome, there are painful tonic muscular spasms and progressive rigidity of the muscles of the trunk, neck, abdomen, back, and proximal parts of the extremities. Other disorders causing increased muscle tone are discussed in [Chapter 30](#).

Video Links

Video Link 28.1. Wartenberg pendulum test. <https://www.youtube.com/watch?v=yYtGjvCcA7o>

Video Link 28.2. Rippling muscle disease. <https://www.youtube.com/watch?v=bdiwylu3Oro>

Video Link 28.3. Discussion and demonstration of spasticity versus rigidity from the 2015 Stanford Medicine 25 Skills Symposium.
<https://www.youtube.com/watch?v=gLZoYLxdXCQ&index=5&list=PL5o6KWShAMajcL3piv2wi>

Video Link 28.4. Training video on paratonia. <https://www.youtube.com/watch?v=Z-NjgIPbuEU&t=89s>

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CHAPTER 29

Muscle Volume and Contour

A search for evidence of muscle atrophy or hypertrophy is an important part of the motor examination. There is normally an appreciable individual variation in muscular development, but noteworthy changes in the size or shape of individual muscles or muscle groups, especially when focal or asymmetric, may be significant.

Muscle atrophy (amyotrophy) causes a decrease in muscle volume or bulk and is usually accompanied by changes in shape or contour. Neurologic conditions likely to cause muscle atrophy are primarily those that affect the following components of the motor unit: the anterior horn cell, the nerve root(s), the peripheral nerve, or the muscle. Neuromuscular junction disorders do not cause muscle atrophy. Atrophy may also result from such things as disuse or inactivity, immobilization, tendonotomy, muscle ischemia, malnutrition, endocrine disorders, and normal aging.

Muscle hypertrophy is an increase in the bulk, or volume, of muscle tissues. It may result from excessive use of the muscles (physiologic hypertrophy) or occur on a pathologic basis. Hypertrophied muscle is not necessarily stronger than normal. Persistent abnormal muscle contraction may cause hypertrophy. Patients with myotonia congenita have a diffuse muscularity without significant increase in strength. Patients with dystonia may develop hypertrophy of the abnormally active muscle. In cervical dystonia (spasmodic torticollis), it is common to see hypertrophy of one sternomastoid muscle. Muscular dystrophies, especially Duchenne's dystrophy, often cause pseudohypertrophy of muscle, with enlargement because of infiltration of the muscle with fat and connective tissue without an actual increase in muscle fiber size or number.

EXAMINATION OF MUSCLE VOLUME AND

CONTOUR

There is a great deal of individual variation in muscular development, in part constitutional and in part because of training, activity, and occupation. Certain individuals have small or poorly developed muscles, whereas others show outstanding muscular development. The sedentary, the elderly, and those with chronic disease may have small muscles without evidence of wasting or atrophy. Athletes may develop physiologically hypertrophic muscles. In normal individuals, the dominant side may exhibit an increase in the size of the muscles, even of the hand and foot. The appraisal of bulk and contour should be correlated with the other parts of the motor examination, especially with the evaluation of strength and tone.

Muscle volume and contour may be appraised by inspection, palpation, and measurement. Inspection generally compares symmetric parts on the two sides of the body, noting any flattening, hollowing, or bulging of the muscle masses. The muscles of the face, shoulder, and pelvic girdles, and distal parts of the extremities—especially the palmar surfaces of the hands, the thenar and hypothenar eminences, and the interosseous muscles—should be examined specifically. A useful technique for comparing extremities is to look down the long axis. Hold the patient's arms outstretched and close together, comparing “down the barrel” from fingertips to shoulders for any asymmetry.

Palpation assesses muscle bulk, contour, and consistency. Normal muscles are semielastic and regain their shape at once when compressed. When hypertrophy is present, the muscles are firm and hard; in pseudohypertrophy, they appear enlarged but may feel doughy or rubbery on palpation. The feel of pseudohypertrophy has been likened to that of a plastic, gelatinous toy such as an imitation snake. Atrophic muscles are often soft and pulpy in consistency. When degenerated muscles have undergone fibrotic changes, they may feel hard and firm. Those infiltrated or replaced by fat may feel pliant and flabby.

Measurements may be very useful in assessing atrophy or hypertrophy. A pronounced difference in muscle size may be recognized at a glance, especially when confined to one side of the body, one extremity, or one segment of a limb. Slight differences are more difficult to detect, and measurements with a tape measure or calipers may be necessary. Measurements should be made from fixed points or landmarks and the sites—such as the distance above or below the olecranon, anterior superior iliac spine, or patella—recorded. The extremities should be in the same position and in comparable states of relaxation. It may

also be valuable to measure the length of the limbs.

Atrophy or hypertrophy may be limited to an individual muscle, to muscles supplied by a specific structure (e.g., a nerve or root), to those muscles supplied by certain spinal cord segments, or to one-half of the body; or it may be multifocal or generalized. In atrophy related to arthritis and disuse, there may be a pronounced decrease in volume with little change in strength. In myopathies, on the other hand, there is often little atrophy in spite of a striking loss of power. Examination of the skin and subcutaneous tissues may also be relevant, especially in such conditions as dermatomyositis.

ABNORMALITIES OF VOLUME AND CONTOUR

Muscular Atrophy

Many processes may cause muscular atrophy. Neurogenic atrophy follows disease of the anterior horn cell, root, or peripheral nerve. Atrophy because of other neurologic processes, such as the hemiatrophy associated with congenital hemiplegia, is not typically considered neurogenic atrophy even though it is related to nervous system disease. The term neurogenic atrophy as commonly used implies disease affecting some part of the lower motor neuron. Myogenic atrophy is that due to muscle disease, such as muscular dystrophy. As a generalization, when weakness and wasting are comparable, the process is more likely to be neurogenic; when the weakness is disproportionately greater than the wasting, the process is more likely to be myopathic. When a muscle appears wasted but is not weak, the cause is likely to be nonneurologic, such as disuse.

Neurogenic Atrophy

The anterior horn cell and its processes exert a trophic effect on skeletal muscle. The nature of the trophic effect remains poorly understood, but it is not as simple as the effect of nerve impulses. Electrical stimulation of the peripheral nerve, sometimes done after peripheral nerve injury or Bell's palsy, does not help prevent or reverse neurogenic atrophy. Nerves may be involved in regulating the trophic actions of insulin-like growth factor, calcitonin gene-related peptide, and other neurotrophic factors that have an influence on skeletal muscle. When a lesion completely disrupts the lower motor neuron or its peripheral processes,

the affected muscle lies inert and flaccid and no longer contracts voluntarily or reflexively. Muscle fibers decrease in size, causing wasting or atrophy of the entire muscle mass. Without timely reinnervation, the muscle may become fibrotic, with an increase in connective tissue and fatty infiltration.

The more abrupt or extreme the interruption of nerve supply, the more rapid the wasting. The atrophy may either precede or follow other signs, such as weakness. In rapidly progressing diseases, weakness precedes atrophy, but in slowly progressive diseases, the atrophy may precede appreciation of weakness. If the pathologic process is confined to the anterior horn cells or the spinal cord, the neurogenic atrophy is segmental in distribution. Some conditions cause rapid destruction of the anterior horn cells and atrophy in the distribution of the affected spinal cord segments that develops within a short period of time (e.g., poliomyelitis).

In more slowly progressive disorders of the motor neuron (e.g., amyotrophic lateral sclerosis [ALS]), there is a gradual but widespread degeneration of the brainstem motor nuclei and anterior horn cells, causing progressive muscular atrophy that may appear before paralysis is evident ([Figure 29.1](#)). The distribution of the atrophy is important. To make a diagnosis of motor neuron disease, it is necessary to demonstrate widespread denervation in a multiple nerve, multiple root distribution. Eventually, the disease becomes widespread, but it often begins segmentally in one limb. Rarely, it may remain confined to one limb (monomelic motor neuron disease, Hirayama disease, O'Sullivan-McLeod syndrome). Particular groups of muscles are often affected. In classical ALS and in progressive spinal muscular atrophy (SMA) of the Aran-Duchenne type, atrophy is usually first seen in the distal musculature—the thenar, hypotenar, and interosseous muscles of the hand and the small muscles of the foot—and then spreads up the limbs to the proximal parts. In some patients, ALS seems to have a tendency to preferentially involve the muscles of the lateral half of the hand, median-innervated thenar, and ulnar-innervated first dorsal interosseous muscles while sparing the hypotenar muscles (split hand syndrome). Although not common, this pattern seems to be relatively specific for anterior horn cell disorders. In progressive bulbar palsy, the atrophy is first noted in the muscles supplied by the 12th, 10th, and 7th cranial nerves. In hereditary motor neuron syndromes, the involvement is often proximal. In SMA type 1 (infantile progressive SMA, Werdnig-Hoffmann disease), the atrophy first involves the trunk, pelvic, and shoulder muscles and then spreads toward the periphery. The proximal distribution and slow progression in SMA type 3

(juvenile proximal SMA, Kugelberg-Welander disease) may simulate muscular dystrophy. Segmental atrophy may also follow focal spinal cord lesions involving the anterior horn cells (e.g., syringomyelia). The rapidity of the progress depends upon the type of pathologic change.



FIGURE 29.1 A patient with amyotrophic lateral sclerosis showing advanced atrophy of the muscles of the shoulder girdle and upper arms (**A**) and intrinsic hand muscles (**B**).

Involvement of nerve roots, plexus elements, or peripheral nerves leads to atrophy of the muscles supplied by the diseased or injured component (Figure 29.2). With severe lesions involving a peripheral nerve or nerve plexus, atrophy may develop within a short period of time. Within 1 month after denervation, there may be a 30% loss of weight in the affected muscle and a 50% loss within 2 months; thereafter, the atrophy progresses more slowly and replacement by connective tissue and infiltration by fat follows. Lesions involving single nerve roots usually do not cause much atrophy, because most muscles are innervated from more than one level. Marked wasting in a disease that appears consistent

with radiculopathy suggests multiple root involvement. In generalized peripheral neuropathy, weakness and wasting are usually greatest in the distal portions of the extremities. The amount of atrophy depends on the severity and chronicity of the neuropathy. The hereditary peripheral neuropathy, Charcot-Marie-Tooth disease (peroneal muscular atrophy), typically causes marked atrophy in a characteristic distribution involving the lower legs (inverted champagne bottle deformity; [Figure 29.3](#)).

The complete syndrome of peripheral nerve dysfunction, with paralysis, atrophy, sensory impairment, areflexia, and trophic changes in the skin and other tissues, is the result of interruption of motor, sensory, and autonomic fibers. Interruption of sensory nerve fibers alone does not lead to muscular atrophy except as related to disuse, but loss of pain sensation may predispose one to painless injuries, including ulcerations following minor trauma and burns. The autonomic system is involved in trophic function by regulating the nutrition and metabolism of muscle and other tissues. Because of interruption of autonomic pathways, diseases of the lower motor neuron may be associated with trophic changes in the skin and subcutaneous tissues: edema, cyanosis or pallor, coldness, sweating, changes in the hair and nails, alterations in the texture of the skin, osteoporosis, and even ulcerations and decubiti. Autonomic fibers may be a factor in muscle atrophy because of “trophic dysfunction” and loss of vasomotor control.

Upper motor neuron lesions in adults are usually not followed by atrophy of the paralyzed muscles except for some generalized loss of muscle volume and secondary wasting because of disuse, which is seldom severe. With lesions dating from birth or early childhood, there may be a failure of growth of the contralateral body ([Figure 25.5](#)). Such congenital hemiatrophy may involve one side of the face or the face and corresponding half of the body; it is characterized by underdevelopment not only of the muscles but also of the skin, hair, subcutaneous tissues, connective tissue, cartilage, and bone. Congenital hemihypertrophy is rarer than the corresponding hemiatrophy, and there are usually other anomalies. There may be underdevelopment of one-half of the body because of either lack of development or atrophy of the opposite cerebral hemisphere (cerebral hemiatrophy). Severe cerebral insults in early life may lead to hemiplegia, hemiatrophy, partial or hemiseizures, and the development of delayed hemidystonia (“4-hemi” syndrome). Experimentally, lesions of the motor cortex and the descending corticospinal pathways may be followed by muscular atrophy, and on occasion, severe wasting appears with cerebral

hemiplegias. Usually, there are associated trophic and sensory changes, and the wasting may in part be secondary to involvement of the postcentral gyrus or parietal lobe, lesions of which are known to be followed by contralateral atrophy. The loss of muscle bulk associated with lesions of the parietal lobe may appear promptly; the degree of atrophy depends upon the size and character of the lesion and the extent of the hypotonia and sensory change. The distribution is determined by the localization of the process within the parietal lobe. It is most severe if the motor cortex or pathways are involved along with the sensory areas of the brain. Hemiatrophy may also complicate hemiparkinsonism. Rarely, hemiatrophy is idiopathic.



FIGURE 29.2 Atrophy of the left abductor pollicis brevis in a plumber with carpal tunnel syndrome.

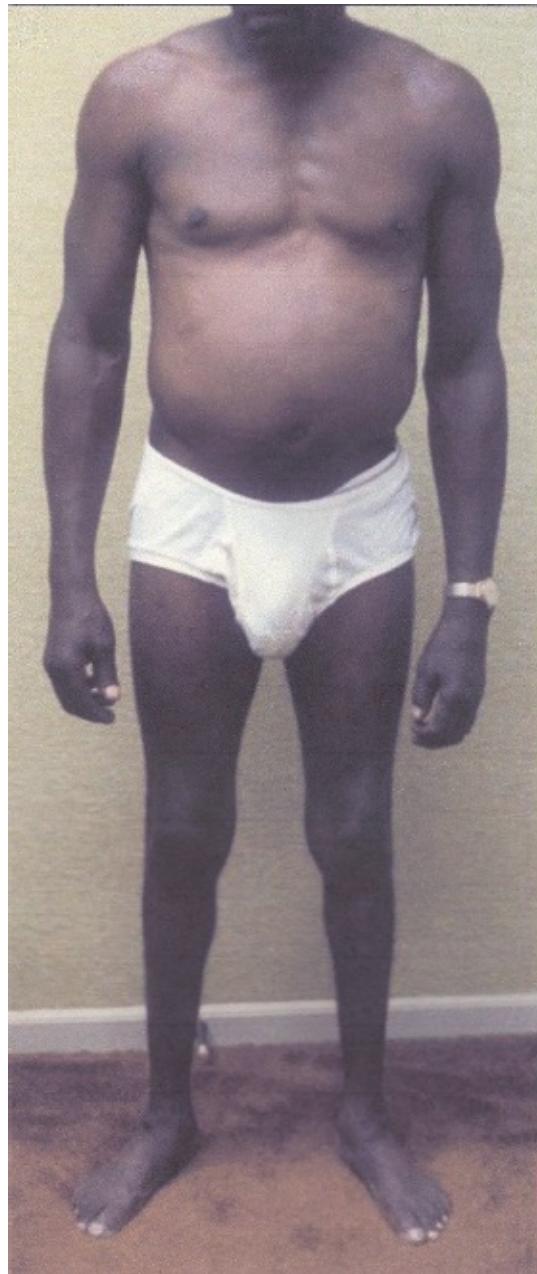


FIGURE 29.3 A patient with type 1 Charcot-Marie-Tooth disease. Despite a muscular upper body, there is marked atrophy of the lower extremities below the knees.

Other Varieties of Muscular Atrophy

Myogenic, or myopathic, atrophy occurs as a result of primary muscle disease. In some conditions, there may be prominent wasting without much weakness. In most of these, the primary pathologic change is type 2 fiber atrophy. Wasting with little weakness occurs in disuse, aging, cachexia, and some endocrine

myopathies. Weakness out of proportion to wasting occurs in inflammatory myopathy, myasthenia gravis, and periodic paralysis.

Muscle wasting is common in muscular dystrophy, and the distribution of the wasting parallels the weakness. In dystrophinopathies, the weakness and atrophy primarily involve the pelvic and shoulder girdle muscles ([Figure 29.4](#)). Weakness of the hip and spine extensors causes difficulty in assuming the erect position, and the patient “climbs up his thighs” (Gowers’ sign or maneuver) in order to stand ([Video Link 29.1](#)). As the disease progresses, there is increasing wasting of all muscles of the shoulders, upper arms, pelvis, and thighs. In the face of all of the atrophy, certain muscles—particularly the calf muscles—are paradoxically enlarged because of pseudohypertrophy (see below). The limb-girdle syndromes also primarily involve the pelvic and shoulder girdles. In facioscapulohumeral (FSH) (Landouzy-Dejerine) dystrophy, the atrophy predominates in the muscles of the face, shoulder girdles (especially the trapezius and periscapular muscles), and upper arms, especially the biceps ([Figure 29.5](#)). Involvement is often asymmetric, and occasionally, there is pseudohypertrophy of the deltoid and other shoulder muscles.

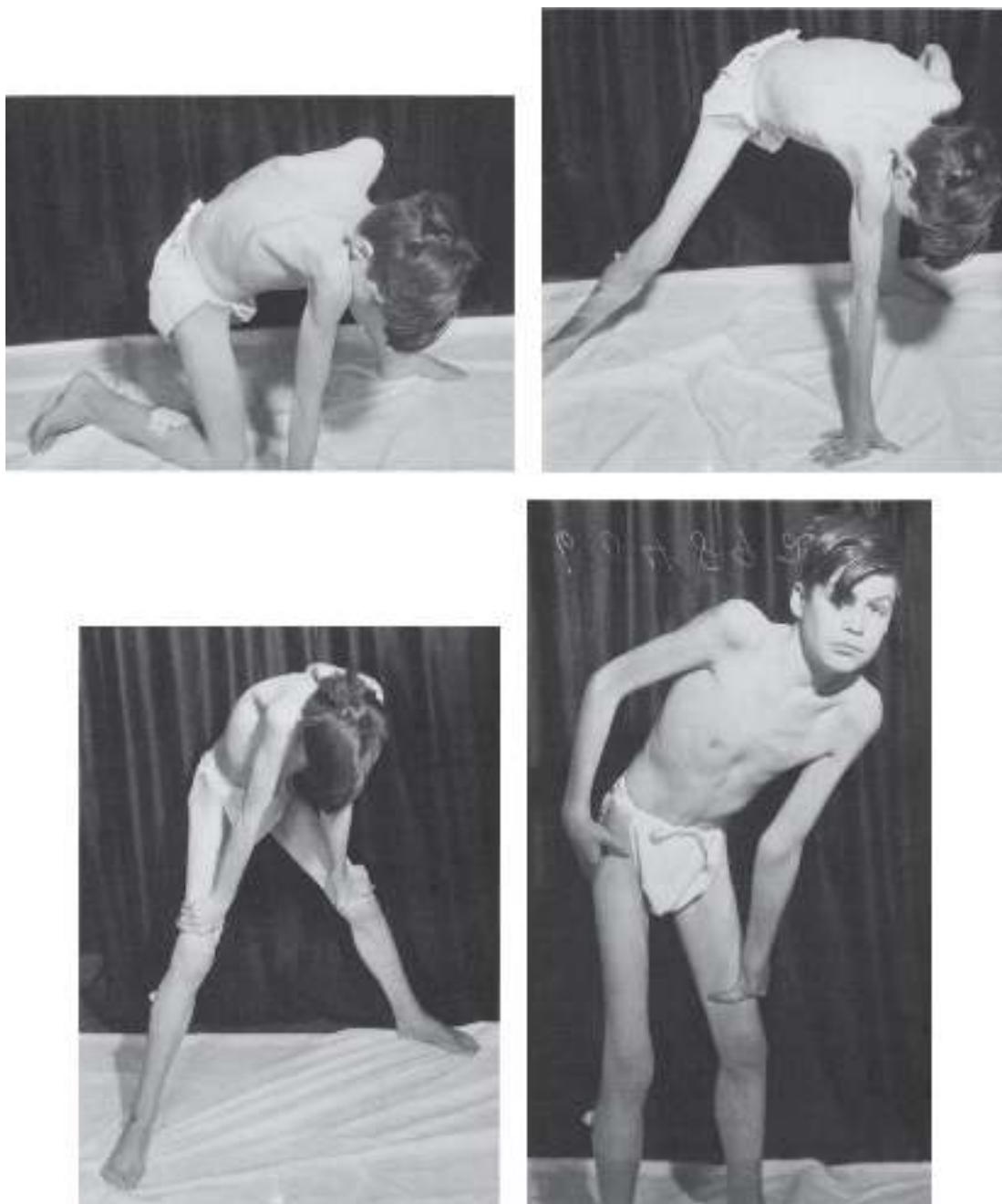


FIGURE 29.4 A boy with Duchenne's muscular dystrophy, showing wasting of the musculature in the shoulders and thighs; weakness and atrophy of the glutei cause difficulty in assuming the erect position, and the patient "climbs up on his thighs" (Gowers' maneuver) in order to stand erect.

Distal myopathies, affecting the muscles of the hands and feet, are occasionally seen. Wasting involving the distal extremities, with relative sparing of the hands and feet, is likely to be myopathic. In contrast, denervation atrophy involves the entire distal extremity, including the hand or foot. Some myopathies

cause striking weakness and atrophy involving certain muscles or muscle groups. In myotonic dystrophy, there is prominent atrophy of the sternocleidomastoid muscles. Scapuloperoneal syndromes involve the periscapular and peroneal muscles. Selective involvement of the quadriceps occurs in inclusion body myositis and type 2B limb-girdle muscular dystrophy (dysferlin deficiency). Some myopathies have a curious tendency to affect certain muscles while sparing nearby muscle groups. FSH characteristically causes wasting of the biceps and triceps with sparing of the deltoid and forearm musculature creating the “Popeye” arm appearance ([Figure 29.6](#)). The diamond on quadriceps sign refers to asymmetric diamond-shaped bulges seen in the anterolateral thighs of patients with dysferlinopathy (LGMD 2B and Miyoshi myopathy) when standing with the knees slightly bent. Proximal and distal atrophy causes the curious island of sparing to stand out. Other unusual muscle shapes have been described as a manifestation of dysferlinopathy, such as selective biceps atrophy causing a “bowl-shaped biceps.”



FIGURE 29.5 A patient with facioscapulohumeral muscular dystrophy showing atrophy of the muscles of the shoulders and upper arms and pronounced scapular winging.

Disuse atrophy follows prolonged immobilization of a part of the body. It may be rapid in onset and can sometimes simulate neurogenic atrophy. Disuse atrophy may occur in an extremity after immobilization, such as casting, one that cannot be moved normally because of joint disease, such as arthritis, paresis following a cerebral lesion, or after prolonged bed rest. The quadriceps femoris is particularly susceptible to disuse atrophy because of bed rest or because of pain in the knee or hip. The degree of muscle wasting is greater than the degree

of weakness, which may be minimal or absent. Muscle biopsy shows atrophy of type 2 fibers, with the earliest changes in the type 2B fibers. Disuse atrophy can occasionally occur in extremities that are not used because of nonorganic paralysis.



FIGURE 29.6 The Popeye arm appearance in FSH dystrophy.

Arthrogenic atrophy may appear in association with joint disease. It is more severe and develops more rapidly in acute arthritis. Both rheumatoid arthritis and osteoarthritis may cause periarticular atrophy, with loss of muscle bulk around involved joints. Periarticular muscle atrophy may be particularly prominent in

patients with HIV-associated arthritis. Atrophy of this type may in part be the result of inactivity or disuse, but other factors are likely involved.

Muscle atrophy may accompany malnutrition, weight loss, cachexia, and other wasting diseases. The loss of muscle mass is typically greater than the degree of accompanying weakness. A normal blood supply is essential to the nutrition and oxygenation of muscles, and ischemia may lead to muscle atrophy as well as to alterations in the skin and other trophic changes. In Volkmann ischemic contracture, atrophy accompanies the muscle shortening.

Endocrine dysfunction of various types may lead to atrophy and other changes in muscle. In thyrotoxic myopathy, atrophy is particularly prone to involve the shoulder girdle and may lead to scapular winging. Coarse fasciculations are often seen in the affected areas. With primary hyperparathyroidism, weakness may be associated with atrophy, hyperreflexia, and fasciculations simulating ALS (Vical's syndrome). Myopathy because of excess corticosteroids, exogenous or endogenous, may be associated with muscle wasting. Muscular weakness and atrophy are also frequent findings in hypopituitarism because of loss of thyroid and adrenal cortical hormones. Muscle wasting also occurs with diabetes. Distal weakness and atrophy are common in diabetic distal axonopathy. Diabetic amyotrophy (or radiculoplexus neuropathy) is a common syndrome of bilateral but asymmetric weakness and atrophy that involves the pelvic and thigh muscles because of involvement of the lumbosacral plexus and nerve roots. It is usually associated with severe pain. Patients with diabetes may also develop either localized lipoatrophy or areas of focal muscular atrophy because of repeated injections of insulin in the same area. The loss of subcutaneous tissue may simulate muscle atrophy. In adiposis dolorosa (Dercum's disease), the muscles may be replaced with fat.

Congenital hypoplasia or absence of a muscle may be mistaken for atrophy. Almost any muscle may be congenitally absent, but some are particularly prone, including the depressor anguli oris, palmaris longus, trapezius, peroneus tertius, and anterior abdominal muscles (prune-belly syndrome). In the Holt-Oram syndrome, there are absent or hypoplastic thenar muscles. Poland syndrome is a rare anomaly characterized by unilateral absence of the pectoral muscles and ipsilateral hand abnormalities; it may be associated with a variety of other congenital anomalies. Other syndromes of congenital muscle abnormality include Duane retraction syndrome, Möbius' syndrome, and congenital ptosis.

Muscular Hypertrophy and Pseudohypertrophy

Enlarged muscles are encountered less frequently than atrophy. In true muscle hypertrophy, the muscle is enlarged; in pseudohypertrophy, the muscle appears enlarged because it is replaced by fat and fibrous tissue. Extremely muscular individuals may show pronounced development of certain groups of muscles because of functional or physiologic hypertrophy, often found in athletes and heavy manual laborers. Microscopic examination shows an increase in the diameter of muscle fibers, primarily the type 2 fibers, without any increase in the number of fibers. Except for physiologic hypertrophy because of exercise, pseudohypertrophy is encountered more commonly than true hypertrophy.

Pseudohypertrophy is common in some forms of muscular dystrophy. Muscle biopsy reveals severe myopathy, with fatty and connective tissue infiltrations. Pseudohypertrophy is common in Duchenne's and Becker's dystrophy; an alternate term for Duchenne's dystrophy is pseudohypertrophic muscular dystrophy. Certain muscles, particularly the calf muscles and the infraspinatus, are often strikingly enlarged because of pseudohypertrophy ([Figure 29.7](#)). Comparing the circumference of the calf to the knee is most informative. In the early stages of the disease, the enlarged muscles may feel firm and hard and remain strong, and there may actually be an element of true hypertrophy. With progression, they develop a soft doughy or rubbery feeling.

Muscle hypertrophy is common in myotonia congenita, especially the dominant form (Thomsen's disease), because of the excessive contraction. These patients may have the impressive muscularity of a bodybuilder; although they may appear strong and muscular, strength is normal, or there is even slight weakness. Hypertrophia musculorum vera is a hereditary syndrome causing enlargement of the muscles, usually those of the limbs, but any area may be affected. Mutations in the myostatin gene may cause muscle hypertrophy.



FIGURE 29.7 Pseudohypertrophy of the calves in Duchenne's muscular dystrophy. (Reprinted from Schaaf CP, Zschocke J, Potocki L. *Basiswissen Humangenetik*. Heidelberg: Springer Medizin; 2008. Figure 31.11. Reproduced with permission of Springer in the format Book via Copyright Clearance Center.)

Muscle enlargement, either true hypertrophy or pseudohypertrophy, occurs as an occasional feature in other neuromuscular disorders, including Kugelberg-Welander disease, central core disease, centronuclear myopathy, autosomal recessive limb-girdle muscular dystrophy, acid maltase deficiency, polymyositis, FSH muscular dystrophy, inclusion body myositis, hyperkalemic periodic paralysis, paramyotonia congenita, proximal myotonic myopathy, Isaac's syndrome, focal myositis, and in manifesting dystrophinopathy carriers. Chronic partial denervation of muscle occasionally leads to focal muscle hypertrophy, presumably because of compensatory physiologic hypertrophy of unaffected fibers or parts of the muscle. Muscle hypertrophy has been reported as a manifestation of radiculopathy and rarely in other neurogenic processes. Use or abuse of androgenic steroids or beta-2 adrenergic agonists may lead to muscle hypertrophy.

Muscle enlargement may occur in hypothyroidism. The enlarged muscles have reduced strength, fatigability, and slowness of contraction and relaxation.

The Kocher-Debré-Semelaigne (infant Hercules) syndrome is diffuse muscular hypertrophy because of hypothyroidism, particularly early in life. Hoffman syndrome refers to a hypertrophic myopathy because of hypothyroidism in adults. Early acromegaly may cause generalized muscular hypertrophy with increased strength, but in later stages, there is weakness and amyotrophy. Edema and inflammation of muscles may simulate hypertrophy. Muscle enlargement may also occur due to interstitial infiltrates, as occurs in cysticercosis, trichinosis, sarcoidosis, and amyloidosis. Focal muscle enlargement may occur with benign or malignant neoplasms. The masseters may become enlarged because of bruxism or as a familial condition.

Loss of body fat may lend the appearance of muscle enlargement. In the lipodystrophies, which may be familial and acquired, there is loss of adipose tissue that may be focal or generalized, often associated with metabolic complications, such as diabetes mellitus and hypertriglyceridemia. Kobberring-Dunnigan syndrome is a familial partial lipodystrophy that may result in an appearance of excessive muscularity, particularly in females.

Video Links

Video Link 29.1. Gowers' sign. http://neurosigns.org/wiki/Gower%27s_sign

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CHAPTER 30

Abnormalities of Movement

Movement disorders may involve any portion of the body. They usually result from disease involving various parts of the motor system, and the etiologies are many. The character of the movement depends on both the site of the lesion and the underlying pathology. Lesions in different sites sometimes cause identical movements, but different etiologic processes involving the same part of the motor system may cause different movement abnormalities.

Movement disorders disrupt motor function not by causing weakness but by producing either abnormal, involuntary, unwanted movements (hyperkinetic movement disorders) or by curtailing the amount of normal free-flowing, fluid movement (hypokinetic movement disorders). The hypokinetic movement disorders are usually accompanied by abnormal states of increased muscle tone. Pathology in movement disorders primarily involves the basal ganglia: caudate, putamen, globus pallidus, substantia nigra, or subthalamic nucleus. The rich connections between the subcomponents of the basal ganglia and between the basal ganglia and other motor systems, as well as the numerous neurotransmitters involved, make the clinical manifestations of basal ganglia disease complex and varied. Depending on the precise location of the abnormality, the particular cell type involved, and the neurotransmitter affected, the clinical picture may range from abnormally decreased movement (the akinesia/bradykinesia of Parkinson's disease [PD]) to abnormally increased movement (chorea, hemiballismus, dystonia).

HYPOKINETIC MOVEMENT DISORDERS

The archetype of hypokinetic movement disorders is PD. Other disease processes may produce a similar clinical picture, characterized by decreased movement and rigidity; these have been grouped together as the akinetic-rigid

syndromes. About 80% of the instances of akinetic-rigid syndrome are due to PD ([Table 30.1](#)). The terms Parkinson syndrome and Parkinson plus are sometimes used to designate such other disorders, and the features that resemble PD are referred to as parkinsonism, or parkinsonian. Parkinsonism is a clinical diagnosis appropriate in the presence of resting tremor, bradykinesia, rigidity, and impaired postural reflexes. PD is but one cause of parkinsonism, and it must be differentiated from other conditions that may have some of its typical features as a component of another disorder.

Parkinson's Disease

PD is due to a degeneration of neurons in the dopaminergic nigrostriatal pathway. It is the second most common movement disorder behind essential tremor (ET), affecting about 1% of the population over the age of 50. The prevalence increases exponentially between the ages of 65 and 90 and reaches 3% of the population over 65. Cardinal manifestations include bradykinesia, rigidity, tremor, an expressionless face, and postural instability. Asymmetry is characteristic. The disease often begins asymmetrically; the signs may be so lateralized as to warrant the designation of hemi-PD, and some asymmetry usually persists even when the disease is well established. The major manifestations vary from case to case. In some, tremor is the outstanding symptom and in others the rigidity, the bradykinesia, or the loss of associated movements. Akinetic, tremor, and postural instability subtypes have been recognized. Asymmetric rest tremor is the most common presentation of PD, occurring in about 75% of patients.

PD causes marked hypertonia, or rigidity, which principally affects the axial muscles and the proximal and flexor groups of the extremities, causing an increased tone to passive movement. The rigidity has a rhythmic quality referred to as cogwheel rigidity (Negro's sign), presumably because of the superimposition of the tremor. Cogwheeling may be brought out as the examiner passively moves an elbow or wrist by having the patient grit the teeth, look at the ceiling, use the opposite hand to make a fist, trace circles in the air, or imitate throwing a ball. The rigidity is present evenly throughout the range of movement, without the ebb at the extremes of the range that occurs in spasticity.

**TABLE
30.1**

**The Differential Diagnosis of Parkinson's Disease
(PD)**

PD

Parkinsonian syndromes

Progressive supranuclear palsy

Multisystem atrophy (MSA)

 MSA-parkinsonian (striatonigral degeneration)

 MSA-cerebellar (olivopontocerebellar degeneration, sporadic form)

 MSA-autonomic (Shy-Drager syndrome)

Diffuse Lewy body disease

Corticobasal degeneration

Drug-induced parkinsonism

Dopa-responsive dystonia

Other non-Parkinson's akinetic-rigid syndromes

Huntington's disease (rigid or juvenile form)

Wilson's disease

Essential tremor

Depression

Arthritis, polymyalgia, and fibromyalgia

In PD, there is a paucity of movement and a slowing of movements. Strictly speaking, akinesia means an absence of movement; bradykinesia, a slowness of movement; and hypokinesia, a decreased amount or amplitude of movement, but the term bradykinesia is often used to encompass all three. Bradykinesia is not due simply to the rigidity and may have an independent pathophysiologic basis. There is loss of associated and automatic movements, with masking of the face, infrequent smiling and blinking, and loss of swinging of the arms in walking ([Figure 30.1](#)). Normal fidgeting and adventitial movements are decreased or absent. Because of the rigidity and bradykinesia, strength may seem to be decreased, but (although an alternate name for PD is paralysis agitans and Parkinson referred to the disorder as "shaking palsy") there is no true loss of power such as is seen in corticospinal lesions. The rigidity and bradykinesia involve movements and not muscles or muscle groups and do make locomotion and motor activity slow and difficult. Under acute emotional stress, the extremities can often be used rapidly and effectively, as when an otherwise immobile patient escapes from a fire (kinesia paradoxica).

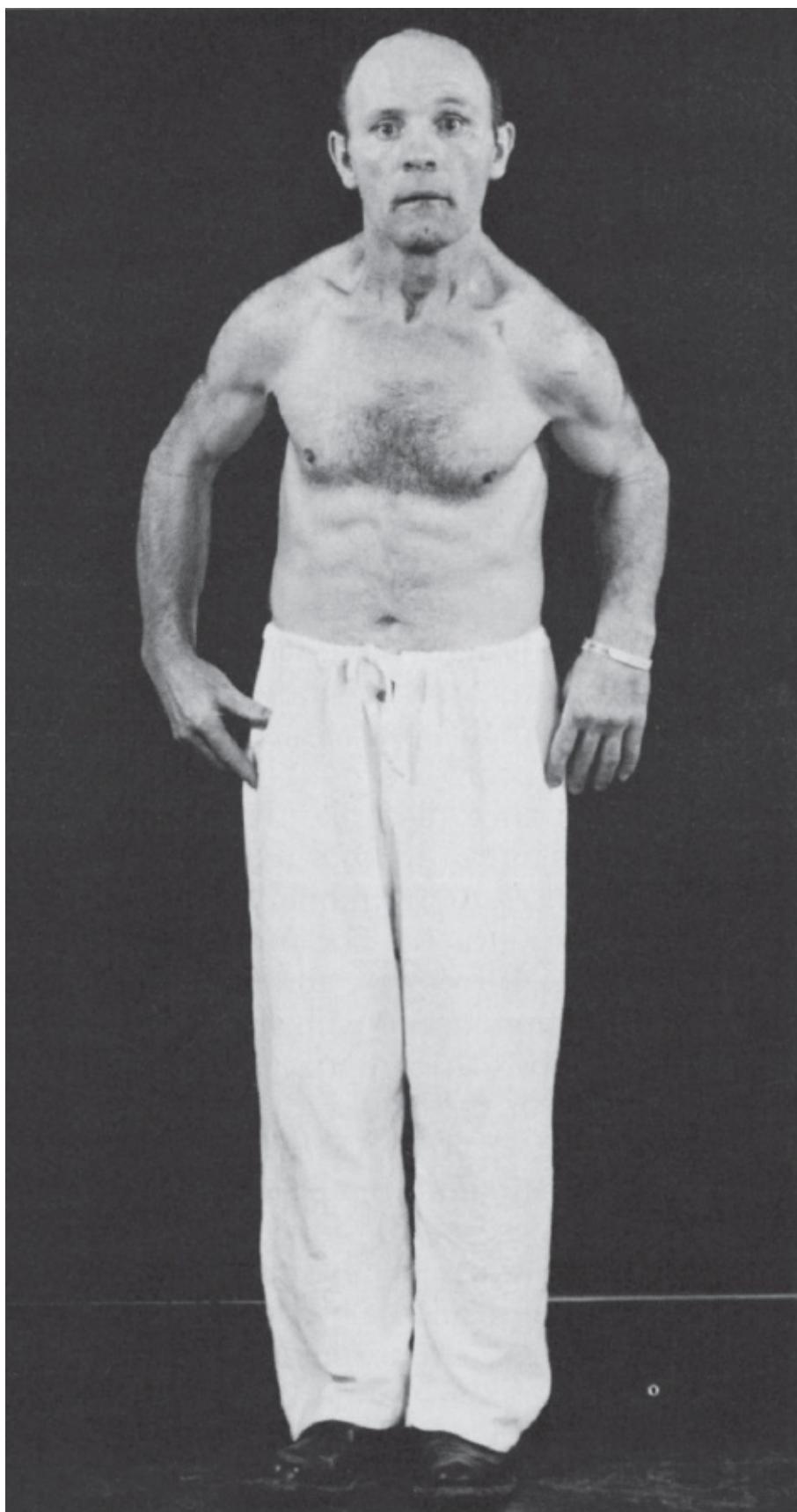


FIGURE 30.1 A patient with Parkinson's disease, showing rigidity, masked facies, and typical posture.

The tremor of PD is a coarse “pill-rolling” movement, so named because of its resemblance to the motion pharmacists of a bygone era used to make pills ([Video 30.1](#)). The tremor is fairly rhythmic and gross, from 2 to 6 Hz, and may involve the hands, feet, jaw, tongue, lips, and pharynx, but not the head. It is typically a resting tremor that lessens during voluntary movement and disappears in sleep. Parkinson said the tremor was present “whilst the limb is at rest and unemployed.” The tremor fluctuates, increasing in amplitude but not rate when the patient becomes excited. The tremor often is more apparent when the patient is walking. Some patients may also have a low-amplitude 7- to 8-Hz tremor during voluntary movement that is suppressed by relaxation.



Video 30.1 Examples of parkinsonian and essential tremor. The parkinsonian tremor is prominent at rest and dampens with the arm outstretched. Essential tremor is an action tremor, usually not evident at rest but appearing with the hands outstretched; it often involves the head and the voice. (Courtesy Dr. Stephen G. Reich.)

Patients with PD have poor balance, a tendency to fall, and difficulty walking. Gait and balance are not prominently affected in most patients with early PD, and significant postural instability early in the course suggests an alternate diagnosis, such as progressive supranuclear palsy (PSP) (see below). The gait abnormality is stereotypical: slow and shuffling with a reduced stride length, sometimes markedly so; a stooped flexed posture of the body and extremities (simian posture); a tendency to turn “en bloc;” and reduced arm swing ([Video 30.2](#)). Head drop and camptocormia may occur. Lower-half (lower-body,

arteriosclerotic) parkinsonism causes gait difficulty out of proportion to other manifestations ([Chapter 44](#)). Impaired postural reflexes lead to a tendency to fall forward (propulsion), which the patient tries to avoid by walking with increasing speed but with very short steps, the festinating gait. Falls are common. If a patient, standing upright, is gently pushed either backward or forward, she cannot maintain balance and will fall in the direction pushed. When a patient, seated in a chair, is suddenly tilted backward, there is absence of the normal reflex leg extension to counteract the loss of balance (Souques' leg sign).



Video 30.2 The first segment of the video demonstrates typical features of a parkinsonian gait with a stooped, flexed posture; short steps; en bloc turning; tremor; reduced arm swing; and impaired postural reflexes. The second segment shows two patients with a festinating gait and the third shows a patient with severe, frequent freezing of gait. (Courtesy Dr. Stephen G. Reich.)

Facial immobility and lack of expressiveness are common features of PD (hypomimia, masked face; [Video 30.3](#)). A decreased rate of blinking (5 to 10 per minute rather than the normal 12 to 20), accompanied by slight eyelid retraction (Stellwag's sign, also seen in thyroid eye disease), causes patients to have a staring expression (reptilian stare). Hanes describes the whistle-smile sign as efficient way to bring out the masked facies in PD. When the normal individual is asked to whistle, he does so and then smiles, probably as a response to the apparent absurdity of the request. The patient suffering from parkinsonism does not smile after whistling because of bradykinesia affecting the facial muscles.



Video 30.3 Evolution of Parkinson's disease over 12 years. (Courtesy Dr. Stephen G. Reich.)

Abnormalities of phonation and articulation are common (bradyphonia). The voice is typically soft, breathy, monotonous, and tremulous. Lack of movement of the lips and tongue causes articulatory imprecision. Temporarily overcoming vocal bradykinesia, words pour out in a short rush of rapid speech. Patients with PD tend to be soft, fast, mumbly talkers. They are often unable to speak loudly or shout. There is also slowness of chewing and swallowing, and the decreased swallowing is largely responsible for the drooling that sometimes occurs.

The freezing phenomenon is common in PD. In the midst of a motor act, the patient will suddenly freeze in place, unable to move for a short time because of simultaneous activation of agonists and antagonists ([Video 30.2](#)). Freezing may occur when first starting to walk (start hesitation), when turning (turn hesitation), when approaching an obstacle, and even when talking or eating. Associated involvement of midbrain structures may cause changes in ocular movements, including fixation instability, hypometric saccades, convergence insufficiency, and impaired upgaze. Oculogyric crisis, forced involuntary eye deviation, usually upward, is a feature of postencephalitic PD and can occur in drug-induced parkinsonism, but it does not happen in idiopathic PD.

In PD, there is no atrophy, fasciculations, reflex changes, or pathologic reflexes of the type seen in corticospinal tract disorders (see [Table 22.1](#)). Reflex changes may occur if there is associated corticospinal tract involvement, but this does not occur in idiopathic PD. Even when the extrapyramidal signs are asymmetric, the reflexes remain normal and equal. Fragments of dystonia sometimes occur. A “striatal toe” (dystonic toe) is an apparent extensor plantar response, without fanning of the toes that occurs in isolation, without other signs

suggesting corticospinal tract dysfunction, in patients with extrapyramidal disorders such as PD ([Figure 30.2](#)). The extended toe may occur as part of a foot dystonia that includes ankle inversion, arching of the sole, and flexion of the other toes (striatal foot). The parkinsonian hand is slightly extended at the wrist and flexed at the metacarpophalangeal joints with the fingers extended and adducted. There is often exaggeration of the orbicularis oculi and orbicularis oris reflexes. Myerson's sign (glabellar tap reflex) is blinking of the eyes on tapping over the glabella. In PD, the patient is unable to inhibit the response and will continue to blink over and over; normals do not continue to blink with repetitive tapping (see [Chapter 16](#)). Patients with PD may exhibit a variety of nonmotor manifestations, such as anosmia, hyperhidrosis, greasy seborrhea, constipation, somnolence, REM sleep behavior disorder, difficulty turning over in bed, blepharospasm, apraxia of eyelid opening, depression, and dementia. Nonmotor manifestations may occur before motor symptoms develop.



FIGURE 30.2 A “striatal toe” (dystonic toe) resembles the great toe extension of Babinski’s plantar sign but is tonic and sustained, not accompanied by fanning of the toes, and occurs without plantar stimulation; it occurs in extrapyramidal disease, in this case Wilson’s disease.

In the early stages of PD, typical signs are often subtle, and patients may present complaining only of stiffness, impaired handwriting (especially micrographia), or difficulty getting about. Stiffness and myalgic pains may suggest a diagnosis of arthritis, polymyalgia, or fibromyalgia. The facial masking and bradykinesia often lead to a misdiagnosis of depression. Early PD may also be mistaken for the effects of aging. Rest tremor, a flexed posture, and mild cogwheel rigidity provide important clues to the possibility of early

parkinsonism in such patients. The clinical diagnosis of PD at the time of initial presentation is occasionally wrong. The pattern of disease progression and the response to medication are important additional factors in determining whether the patient suffers from PD or one of the other akinetic-rigid syndromes.

Advancing disease is characterized by increasing gait difficulty, worsening of tremor and bradykinesia, motor fluctuations related to levodopa therapy, behavioral changes, dysphagia, speech difficulty, intractable drooling, and sleep impairment ([Video 30.3](#)). Some degree of cognitive blunting may occur in 20% to 40% of patients. Severe dementia is rare; when present, especially early in the course, the possibility of dementia with Lewy bodies should be considered. Depression occurs in approximately one-third of patients, and psychosis and hallucinations occur in about one-fourth of patients. Early, prominent, and nonvisual hallucinations raise the possibility of dementia with Lewy bodies. The absence of tremor in the early stages may suggest a possible Parkinson-plus syndrome. Advanced age at onset, severe depression, dementia, and an akinetic-rigid presentation are risk factors for rapid disease progression. The tremor may begin to abate in the very late stages.

PD usually affects older patients, the mean age at onset is around 60 years, but some cases began relatively early in life. The nosology describing these patients is inconsistent. Juvenile Parkinson's disease (JPD) has its onset before 20 and young-onset Parkinson's disease (YOPD) between 20 and 50, but the terms are sometimes used interchangeably. JPD was first described by Ramsay Hunt and is one of the several "Ramsay Hunt syndromes" ([Box 16.2](#)). YOPD is probably the same entity as idiopathic PD, with a younger age of onset. In a series of 953 individuals with YOPD, 17% were found to have a genetic etiology. In contrast, JPD is a heterogeneous group of disorders. Many patients with JPD have a genetic disorder due to a parkin gene mutation. Patients with parkin mutations may have atypical clinical features such as dystonia at onset and marked diurnal symptom fluctuation. YOPD tends to have more gradual progression of parkinsonian signs and symptoms and earlier treatment-related complications. Susceptibility to developing late-onset PD has been associated with polymorphisms or mutations in several genes.

The diagnosis of PD is predominantly clinical, and differential diagnosis essentially is between other conditions causing tremor, of which ET is the commonest, and other akinetic-rigid syndromes. Clinical features that favor PD include prominent rest tremor, asymmetric signs, preservation of balance and postural reflexes in the early stages of the disease, and a good response to

levodopa replacement therapy. The other degenerative disorders with parkinsonian features typically produce other neurologic signs, such as gaze limitation, cerebellar signs, pyramidal signs, severe dementia, apraxia and other parietal lobe signs, or dysautonomia, although these other manifestations may not be apparent early in the course. In the past, a diagnosis of PD required two of three parkinsonian features (tremor, rigidity, bradykinesia). Using these criteria resulted in a 24% error rate based on pathologic studies. Using revised criteria (the UK Brain Bank criteria) of rest tremor, asymmetry, and a good response to levodopa improved accuracy and resulted in pathologic confirmation of the diagnosis in 99% of cases determined by clinicopathologic correlation studies. The most common condition confused with PD is PSP (see below).

Certain drugs can induce a reversible condition that mimics PD. The most common agents that cause drug-induced parkinsonism are antipsychotics, especially the high-potency piperazine compounds such as haloperidol. The atypical neuroleptics are as potent in their antipsychotic effects as traditional compounds but less likely to induce parkinsonism. Drug-induced parkinsonism can mimic PD closely, even to the point of causing asymmetric signs. Although dopamine receptor-blocking agents, especially those that block the D2 receptor, are the most common offenders, other agents can induce parkinsonism. Some patients are much more prone to develop extrapyramidal side effects than others, but most individuals will eventually develop parkinsonism if treated with high doses. Metoclopramide is a dopamine blocker most often used for gastrointestinal disease; it may have extrapyramidal side effects.

In the presence of typical clinical signs and symptoms and the typical age of onset, extensive workup is not required. Imaging studies are usually normal. A thorough medication history, complete neurologic examination to look for nonextrapyramidal abnormalities, and screening for orthostatic hypotension are useful. Certain features suggest an alternative diagnosis, such as prominent dementia and hallucinations in the patient suffering from dementia with Lewy bodies, prominent dysautonomia in the patient with multisystem atrophy (MSA), dysarthria, and early age of onset in Wilson's disease. Single-photon emission computed tomography (SPECT) imaging of the dopamine transporter system can confirm or exclude loss of nigrostriatal dopaminergic neurons and help distinguish PD from some of the conditions that may mimic it, such as ET. These scans cannot distinguish between PD and conditions such as MSA and PSP.

Pathologically, the disease is characterized by depigmentation and cystic degeneration of the pars compacta of the substantia nigra with cell loss and the

presence of intracytoplasmic Lewy bodies in surviving neurons. A major chemical constituent of the Lewy body is alpha-synuclein, a synaptic protein. Abnormal aggregation of alpha-synuclein has been advocated in the pathogenesis of PD, dementia with Lewy bodies, and multisystem atrophy, and these conditions have been grouped together as synucleinopathies. In PD, there is depletion of dopamine; replacement by its precursor, levodopa, has been well recognized as effective therapy since the landmark studies of the 1960s. The etiology of PD remains unknown, but the basis is likely multifactorial, possibly involving hereditary factors, environmental influences that may selectively affect dopaminergic nigral cells, and free radical toxicity.

The pathophysiology of parkinsonism is complex. Dopamine deficiency ultimately results in an increased output from the internal segment of the globus pallidus and subthalamic nuclei, which results in excessive inhibition of the thalamus and suppression of the cortical motor system (see [Chapter 26](#)). Pharmacologic treatment modalities include anticholinergic drugs, dopamine-releasing agents, dopamine agonists that directly stimulate the dopamine receptor in the striatum, catechol-O-methyl transferase inhibitors, and levodopa. Dopamine and acetylcholine are in balance in the striatum, and anticholinergics increase the effect of dopamine by influencing this balance. These agents were the earliest treatment available for PD. The discovery of L-dopa as a treatment for PD was a major medical advance. The story of that discovery is told in British neurologist Oliver Sacks' book, *Awakenings*, and its motion picture adaptation. Over 90% of patients with PD respond very well to the initiation of levodopa replacement. The absence of a good initial response suggests the possibility of another diagnosis, although some of the mimickers of PD, especially MSA, may also have a good initial response.

Some of the other conditions important in the differential diagnosis of PD include multisystem atrophy, PSP, corticobasal degeneration (CBD), and diffuse Lewy body disease.

Multisystem Atrophy

MSA produces degeneration involving the basal ganglia, cerebellum, anterior horn cells, cerebral cortex, and brainstem in varying combinations and usually includes parkinsonian features. Patients with MSA may have elements of cerebellar ataxia, dementia, amyotrophy, parkinsonism, corticospinal tract dysfunction, dysautonomia, and urinary dysfunction.

There are three subtypes: MSA-P (parkinsonian), MSA-C (cerebellar), and MSA-A (autonomic). In MSA-P (formerly striatonigral degeneration), the primary manifestation is parkinsonism; it accounts for about 80% of the cases of MSA. About 10% of patients presenting with parkinsonism will evolve into MSA-P. In MSA-C (formerly the sporadic form of olivopontocerebellar atrophy), the primary manifestations are cerebellar. In MSA-A (formerly Shy-Drager syndrome), the primary manifestation is dysautonomia, particularly postural hypotension. Suspicion of MSA should arise in the patient with atypical parkinsonism in conjunction with cerebellar signs and/or early and prominent autonomic dysfunction, usually orthostatic hypotension. There is some overlap of pathologic findings in the different forms of MSA. Recently discovered glial cytoplasmic inclusions containing alpha-synuclein confirm these three disorders are different forms of the same disease. The dysautonomia is due to degeneration of the neurons in the intermediolateral gray column of the thoracic and lumbar spinal cord. Impaired peripheral vasomotor control in MSA may cause cold, dusky, purplish discoloration of the digits (cold hands sign; [Figure 30.3](#)).

Parkinsonism is the most frequent motor manifestation of MSA. MSA-P has been associated with relative symmetry of findings, urinary incontinence, frequent falls, an absence of tremor, and the frequent presence of pyramidal or cerebellar signs. MSA-A consists of a combination of parkinsonian signs and symptoms coupled with severe dysautonomia, most often manifest as orthostatic hypotension. The primary manifestations of MSA-C are ataxia and brainstem dysfunction. Approximately one-quarter of patients with MSA-C will develop parkinsonian features within 5 years; such evolution carries a poor prognosis for survival.



FIGURE 30.3 Dusky, violaceous fingers typical of multisystem atrophy. (Reprinted from Reich SG. The cold hands sign in MSA. *Neurology* 2003;60[4]:719, with permission.)

Progressive Supranuclear Palsy

In PSP (Steele-Richardson-Olszewski syndrome, Richardson's syndrome), degenerative changes in the rostral brainstem and thalamus result in impairment first of downgaze, then of upgaze, and eventually in global gaze paresis. Three subtypes are currently recognized: classic Richardson's syndrome (PSP-RS), PSP parkinsonism (PSP-P), and pure akinesia with gait freezing. In PSP-P, the clinical picture resembles PD early in the course. In PSP-RS, the gaze abnormalities are accompanied by parkinsonian signs, frontal lobe-type dementia, postural instability, pseudobulbar palsy, and a pronounced tendency to extensor axial rigidity, especially involving the neck muscles, and sometimes causing frank retrocollis. Gait difficulty is prominent, and a tendency to fall is an early and conspicuous feature. Some have likened the hypererect gait with abducted arms seen in PSP to that of a gunslinger. The falls are often backward because of the increased extensor tone and postural instability. There is particular difficulty walking downstairs because of the combination of retrocollis and impaired downgaze. Tremor is not usually pronounced, and the disease does not respond significantly to levodopa. The gaze abnormalities at first involve voluntary, vertical, saccadic gaze, sparing reflex eye movements; the name of the

disease refers to this supranuclear ocular motility disturbance. The disordered motility may progress to complete ophthalmoplegia terminally.

Facial dystonia may contort the face, particularly the forehead muscles, into a characteristic expression of “perpetual surprise” or astonishment with raised eyebrows, lid retraction, and reduced blinking (procerus sign; [Figure 16.4](#)). The facial expression is markedly different from the hypomimia and masking of PD. The characteristic ocular motility disturbance may not be present early in the course and, in rare instances, never appears. The “applause sign” is an inability to stop clapping after being asked to clap three times. It was initially touted as a way to distinguish PSP from PD and frontotemporal dementia. Later studies found the applause sign in cortical dementia and suggested that it is a nonspecific sign of frontal lobe dysfunction. On magnetic resonance imaging (MRI), a characteristic atrophy of the midbrain with relative preservation of the pons (the “hummingbird sign” on midsagittal images) may be seen. MRI measurement of the midbrain/pons ratio and combined measurements, such as midbrain area/pons area and especially the magnetic resonance parkinsonism index, are very useful in distinguishing PSP from other parkinsonian syndromes.

Corticobasal Syndrome

In CBD (corticobasal degeneration), abnormalities involve both the basal ganglia and the cerebral cortex. Pathologically, there is asymmetric frontoparietal neuronal loss and gliosis, with swollen, achromatic neurons, tau-immunoreactive neuronal and glial inclusions, and nigral degeneration. Clinically, the corticobasal syndrome (CBS) is characteristically very asymmetric initially, with rigidity, bradykinesia, and occasionally tremor, accompanied by evidence of higher cortical dysfunction such as apraxia, agnosia, cortical sensory loss, focal myoclonus, or pyramidal signs. It has been increasingly recognized that not all patients with CBS have CBD pathologically. Many have other pathology, most commonly PSP or Alzheimer’s disease. Conversely, some patients with a clinical picture of PSP, frontotemporal dementia, or Alzheimer’s disease may have CBD pathologically.

CBS typically begins with clumsiness, stiffness, or jerking of an arm. The alien limb phenomenon is common. There may be dystonic posturing as well as spontaneous and reflex myoclonus of the involved limbs. The involved limbs become stiff, jerky, and eventually useless. The combination of unilateral parkinsonism unresponsive to levodopa, accompanied by ideomotor apraxia of

the involved extremities, is very suggestive. MRI frequently shows asymmetric cortical atrophy. Cognitive impairment may emerge as the disease progresses and becomes generalized. The disorder progresses slowly and does not respond to levodopa. Tau protein is a microtubule-associated protein, which when aggregated causes neurofibrillary tangles. Other conditions are associated with abnormalities of tau protein, especially PSP and frontotemporal dementia, collectively referred to as tauopathies.

Diffuse Lewy Body Disease

In diffuse Lewy body disease, the usual clinical picture is progressive dementia with added parkinsonian features in an elderly patient. It is the second most common degenerative dementia after Alzheimer's disease ([Chapter 8](#)). Parkinsonism may occur early or late and varies in severity. The parkinsonian features are typically more symmetric and milder than in PD. Tremor occurs but is less common and less severe than in PD. Another characteristic feature is psychotic behavior with visual hallucinations, delirium, and paranoia. Other common clinical features include cognitive fluctuations; dysautonomia; sleep disorders, especially REM sleep behavior disorder; and neuroleptic sensitivity.

Wilson's Disease

Wilson's disease (hepatolenticular degeneration) is a rare, autosomal recessive disorder due to abnormal copper deposition in the brain, especially the basal ganglia, liver, eye, and other tissues because of a genetic defect in an ATPase involved in copper transport (ATP7B), usually accompanied by a defect in the copper transport protein ceruloplasmin. The genetic defect leads to impaired copper excretion and systemic copper accumulation. A low ceruloplasmin level is found in the serum in 95% of patients. Ceruloplasmin is involved in the transfer of copper to copper-containing enzymes such as cytochrome oxidase, and the dysfunction of these enzymes may underlie the clinical manifestations.

The usual age of onset is between the ages of 10 and 20, and major manifestations include tremor, rigidity, dystonia, abnormal involuntary movements of various types, dysarthria, dementia, parkinsonian features, spasticity, cerebellar signs, and psychiatric abnormalities (anxiety, depression, psychosis). The tremor may be present at rest and increased by voluntary movement. Most characteristic is a "wing-beating" tremor of the proximal upper

extremities, consisting of a slow, high-amplitude, up-and-down movement of the elbow when the arm is held with the shoulder abducted and the elbow flexed (see [Video Link 30.1](#)). Pathologically, there is symmetric degeneration of the lenticular nuclei, with widespread neuronal loss and proliferation of Alzheimer-type II astrocytes.

Kayser-Fleischer rings are crescents of green-brown discoloration of the cornea because of copper deposits in Descemet's membrane ([Figure 30.4](#)); these are essentially always present in patients with neurologic involvement but may not be visible without a slit lamp. Rarely, the disease may present without the rings. Slit lamp examination may also detect sunflower cataracts because of copper deposition in the lens.

Risus sardonicus refers to an unusual forced “smile” because of facial dystonia and may be seen in Wilson's disease and other disorders ([Figure 30.5](#)). Other manifestations of the disease include cirrhosis, atypical hepatitis, hemolytic anemia, and renal disease. Patients may present with liver disease alone, brain disease alone, or evidence of both. In a series of 282 patients seen over three decades, the mean age at diagnosis was 16, and the predominant features were neurologic (in 69%); the predominant neurologic manifestations were parkinsonism (62%) and dystonia (35%).



FIGURE 30.4 Copper deposits in Descemet's membrane producing the brownish Kayser-Fleischer ring in Wilson's disease. (Reprinted from Chern KC, Saidel MA. *Ophthalmology Review Manual*. 2nd ed. Philadelphia: Wolters Kluwer Health/Lippincott Williams & Wilkins, 2012, with permission.)



FIGURE 30.5 Risus sardonicus in a patient with Wilson's disease.

Pantothenate Kinase–Associated Neurodegeneration

Pantothenate kinase–associated neurodegeneration (PKAN, neurodegeneration with brain iron accumulation type 1, Hallervorden-Spatz syndrome) is a rare autosomal recessive disorder associated with macroscopic rust-brown discoloration of the globus pallidus and substantia nigra because of iron deposition. The clinical phenotype is variable. The disease usually begins in the

first to the fourth decade of life, with rigidity, involuntary movements, ataxia, and dystonia, followed by pyramidal signs and progressive dementia. MRI findings are characteristic. T2-weighted sequences show bilaterally symmetric low signal intensity in the globus pallidus, because of iron deposition, surrounding a focus of high signal intensity, because of gliosis. This “eye of the tiger” image pattern is virtually diagnostic for PKAN.

Dentatorubropallidolysian Atrophy

Dentatorubropallidolysian atrophy is a heredofamilial degeneration, because of CAG repeats, in which the pathologic changes involve primarily the dentate, globus pallidus externa, red nucleus, and subthalamic nucleus. Clinical manifestations include choreoathetosis, dystonia, dementia, myoclonus, and ataxia; the disease is often included as one of the hereditary ataxia syndromes. Most reported cases have been from Japan. A cluster of cases occurred in the United States around the Haw River in North Carolina (Haw River syndrome).

HYPERKINETIC MOVEMENT DISORDERS

Hyperkinesia refers to increased movement. Hyperkinesias are abnormal involuntary movements that occur in a host of neurologic conditions. Hyperkinesias come in many forms, ranging from tremor to chorea to muscle fasciculations to myoclonic jerks. Any level of the motor system, from the motor cortex to the muscle itself, may be involved in their production. The only common characteristic is that the movements are spontaneous and, for the most part, not under volitional control. They may be rhythmic or random, fleeting or sustained, and predictable or unpredictable and may occur in isolation or accompanied by other neurologic signs. [Table 30.2](#) summarizes some of these features.

In the examination of abnormal movements, the following should be noted: (a) the part of the body involved or the exact location of the movements; (b) the extent of the movement, or its distribution as it regards to part of a muscle, an entire muscle, movement involving joints, or more complex or composite patterns consisting of a sequence of different movements; (c) the pattern, rhythmicity, uniformity, multiformity, and regularity of recurrence—there may be a regular or rhythmic recurrence of activity involving the same muscle or

groups, or there may be an irregular pattern of constantly changing motion of different parts; (d) the course, speed, and frequency of each particular movement; (e) the amplitude and force of the movement; (f) the relationship to posture, rest, voluntary activity or exertion, involuntary activity, various stimuli, fatigue, and time of day; (g) the response to heat and cold; (h) the relationship to emotional tension and excitement; (i) the degree that movements are suppressible by attention or the use of sensory tricks; and (j) the presence or absence of the movements during sleep. In general, involuntary movements are increased by stress and anxiety and decrease or disappear with sleep. Truly involuntary movements must be separated from complex or bizarre voluntary movements, such as mannerisms or compulsions.

TABLE 30.2 Abnormal Involuntary Movements as a Spectrum of Movements

Regular/Predictable	Intermediate	Fleeting/Unpredictable
Tremor	Most dystonias	Fasciculations
Hemiballism	Myokymia	Myoclonus
Palatal myoclonus	Athetosis	Chorea
	Tic	Dyskinesias
	Stereotypy	
	Myorhythmia	

It may be possible to name movements that fit a well-defined clinical pattern, but it is often better to describe the abnormality. Palpation may sometimes be useful, especially if the movements are very fine and limited to individual muscles. Videos are often very useful in the diagnosis and management of movement disorders.

TREMOR

A tremor is a series of involuntary, relatively rhythmic, purposeless, oscillatory

movements. The excursion may be small or large and may involve one or more parts of the body. A simple tremor involves only a single muscle group; a compound tremor involves several muscle groups and may have several elements in combination, resulting in a series of complex movements (e.g., alternating flexion and extension together with alternate pronation and supination). Not only the agonist and antagonist, but muscles of fixation and synergists may play a part in the movements. A tremor may be present at rest or with activity. Some tremors are accentuated by having the patient hold the fingers extended and separated with the arms outstretched. Slow movements, writing, and drawing circles or spirals may bring tremor out.

Tremors may be classified in various ways: by location, rate, amplitude, rhythmicity, relationship to rest and movement, etiology, and underlying pathology. Other important factors may include the relationship to fatigue, emotion, self-consciousness, heat, cold, and the use of medications, alcohol, or street drugs. Tremor may be unilateral or bilateral and most commonly involves distal parts of the extremities—the fingers or hands—but may also affect the arms, feet, legs, tongue, eyelids, jaw, and head and may occasionally seem to involve the entire body. The rate may be slow, medium, or fast. Oscillations of 3 to 5 Hz are considered slow and 10 to 20 Hz rapid. Amplitude may be fine, coarse, or medium. Tremor may be constant or intermittent, rhythmic or relatively nonrhythmic, although a certain amount of rhythmicity is implied in the term tremor. Irregular “tremor” may be due to myoclonus.

The relationship to rest or activity is the basis for classification into two primary tremor types: rest and action. Resting (static) tremors are present mainly during relaxation (e.g., with the hands in the lap) and attenuate when the part is used. Rest tremor is seen primarily in PD and other parkinsonian syndromes.

Action tremors appear when performing some activity. Action tremors are divided into subtypes: postural, kinetic, task-specific, and isometric. Only when they are very severe are action tremors present at rest. Postural tremors become evident when the limbs are maintained in an antigravity position (e.g., arms outstretched). Common types of postural tremor are enhanced physiologic tremor and ET. Kinetic tremor appears when making a voluntary movement and may occur at the beginning, during, or at the end of the movement. The most common example is an intention (terminal) tremor. Intention tremor is a form of action tremor seen primarily in cerebellar disease (see [Chapter 43](#)). The tremor appears when precision is required to touch a target, as in the finger-nose-finger or toe-to-finger test. It progressively worsens during the movement.

Approaching the target causes the limb to shake, usually side-to-side perpendicular to the line of travel, and the amplitude of the oscillation increases toward the end of the movement. Some tremors fall into more than one potential classification. Most tremors are accentuated by emotional excitement, and many normal individuals develop tremor with anxiety, apprehension, and fatigue. A shivering type of tremor (rigors) may be brought on by cold, but identical movements can be psychogenic.

Physiologic tremor is present in normal individuals. The frequency varies from 8 to 12 Hz, averaging about 10 Hz in the young adult, somewhat slower in children and older persons. The frequency for an individual is the same at different sites in the body. The visible tremor brought out in normal persons by anxiety, fright, fatigue (rock climber's tremor, Elvis leg), and other conditions with increased adrenergic activity is accentuated or enhanced physiologic tremor. A typical example of enhanced physiologic tremor is that seen in hyperthyroidism. The tremor involves principally the fingers and hands and may be fine and difficult to see. It may be brought out by placing a limb in a position of postural tension, by performing voluntary movements at the slowest possible rate, or holding the index fingertips as close together as possible without touching. The tremor may be better appreciated by placing a sheet of paper on the outstretched fingers; shaking of the paper may be obvious even though tremor is not grossly visible. Physiologic tremor may be present both at rest and on activity, but it is accentuated by activity as well as by anxiety and emotional stress. Similar tremor occurs due to the effects of alcohol, nicotine, caffeine, amphetamines, ephedrine, and other stimulants ([Table 30.3](#)). A fine tremor of the closed eyelids is seen in hyperthyroidism (Rosenbach's sign).

TABLE 30.3

Some Drugs That Cause Tremor

- Sympathomimetics (epinephrine, pseudoephedrine, isoproterenol, metaproterenol, albuterol, terbutaline, ritodrine)
- Aminoglycoside antibiotics (amikacin, kanamycin, tobramycin)
- Methylxanthines (aminophylline, theophylline)
- Amphetamines
- Anticholinergics
- Antihistamines
- Bupropion

Carisoprodol, orphenadrine (centrally acting muscle relaxants)
Antipsychotics
Cyclosporine
Benzodiazepines (diazepam, oxazepam)
Selective serotonin reuptake inhibitors
Other antidepressants (mirtazapine, amoxapine, trazodone, clomipramine)
Lithium
Thyroid supplements
Antiarrhythmics (mexiletine, amiodarone, quinidine)
Opioid antagonists (naloxone)
Phenytoin
Tramadol
Valproic acid
Vasopressin
Yohimbine

Tremor of medium amplitude and rate is often evident in anxiety. Such tremor may occur in the absence of neurologic disease. The tremor is usually postural, most evident with the hands outstretched, made worse by movement, and may interfere with motor activity. ET is often of medium amplitude and rate but may be coarse when severe. The intention tremor of multiple sclerosis (MS) and cerebellar disease is usually of medium amplitude and may vary in degree from mild to severe; it may be coarse and irregular, especially when associated with ataxia. Coarse tremors occur in a variety of disease states and are usually slow. Parkinsonian tremor is one of the most characteristic. Coarse tremor also occurs in Wilson's disease and other extrapyramidal syndromes. The tremor of general paresis and alcoholism may also be coarse, especially if the movements are diffuse, as in delirium tremens. Psychogenic tremor and the tremor associated with midbrain and cerebellar disease may also be coarse and slow.

Parkinsonian Tremor

Resting, static, or nonintention tremor occurs most frequently in diseases of the

basal ganglia and extrapyramidal pathways ([Video 30.1](#)). The most characteristic tremor of this type is seen in PD and the various parkinsonian syndromes (see PD above). The tremor is slow, coarse, and compound in type. The rate may vary from 2 to 6 Hz, averaging 4 to 5 Hz. The movement in the hand characteristically consists of alternate contractions of agonist and antagonist, involving the flexors, extensors, abductors, and adductors of the fingers and thumb, together with motion of the wrist and arm, including flexion, extension, pronation, and supination. As a result, there is a repetitive movement of the thumb on the first two fingers, together with the motion of the wrist, producing the classical pill-rolling. The tremor is relatively rhythmic, present at rest, and may be temporarily suppressed by movement. It may disappear temporarily, whereas the limb is engaged in a voluntary effort. Because of the uniformly alternating movements at regular intervals, it is sometimes called an alternating tremor. The tremor may be unilateral at onset; it may even begin in a single digit but, in most cases, eventually becomes bilateral. It disappears during sleep and is aggravated by emotional stimulation, fatigue, and anxiety. Tremor resembling that of parkinsonism may also occur in other extrapyramidal syndromes.

Essential Tremor

ET is the most common of all movement disorders ([Video 30.1](#)). It is higher in frequency and lower in amplitude than the tremor of PD. The etiology and pathophysiology remain obscure, but recent evidence suggests a cerebellar abnormality.

ET is often familial. The prevalence of ET increases with age, may first appear anywhere between the second and sixth decades of life, and tends to be slowly progressive. ET is a postural and action tremor that tends to affect the hands, head, and voice. It is made worse by anxiety. The movement of the head may be in an anteroposterior (affirmative, yes-yes) or a lateral (negative, no-no) direction. Senile tremor is ET occurring during senescence with a negative family history.

A common problem is differentiating the tremor of early PD from ET. The tremor of PD is most prominent at rest, whereas that of ET occurs with a sustained posture, such as with the hands outstretched, or on action. Parkinsonian tremor may persist with hands outstretched but usually damps, at least momentarily, when making a deliberate movement, whereas ET usually

worsens with any attempt at a precise action. The ET patient may have great difficulty sipping water from a cup, but the PD patient may do so without spilling a drop. The head and voice are often involved with ET, only rarely with PD, although the tremor in PD may involve the lips and jaw. Alcohol and beta-blockers often improve ET but have no effect on parkinsonian tremor. Some patients thought to have ET go on to develop PD.

Other Forms of Tremor

Cerebellar tremor is a low-frequency tremor that occurs primarily as the finger approaches a target (intention tremor); it may have a postural component (see Chapter 43). Rubral (Holmes, cerebellar outflow, midbrain) tremor refers to a severe, large-amplitude, relatively slow (2 to 5 Hz) tremor, involving both proximal and distal muscles, present at rest but made worse with action. The clinical picture resembles a combination of parkinsonian and cerebellar tremors. It may be unilateral and is usually due to stroke or trauma. Formerly thought to result from an abnormality of the red nucleus, it is now believed to be due to a lesion involving cerebellar efferent and nigrostriatal fibers coursing through the midbrain.

Orthostatic tremor (shaky legs syndrome) is a variant of ET that involves the legs. It is an isometric tremor most apparent when standing, and it abates when walking. Neuropathic tremor is a coarse postural and action tremor seen in patients with peripheral neuropathy. Dystonic tremor is typically a localized, irregular postural tremor that resembles ET but is more irregular and asymmetric. It usually but not invariably occurs with evidence of dystonia. Head tremor in a patient with cervical dystonia and hand tremor in a patient with writer's cramp are examples. Task-specific tremors appear during performance of a particular activity (e.g., primary writing tremor). Nonorganic or psychogenic tremor is typically complex and does not fit well into the classification scheme. The patient may have action as well as resting tremor, with rapidly changing clinical features and disability out of proportion to the tremor. Typical features include abrupt onset with maximal disability immediately, onset in one limb with rapid generalization, spontaneous resolution and recurrence, easy distractibility, entrainment, and refractoriness to conventional antitremor treatment.

CHOREA

Chorea (Gr. “dance”) is characterized by involuntary, irregular, purposeless, random, and nonrhythmic hyperkinesias ([Video 30.4](#)). The movements are spontaneous, abrupt, brief, rapid, jerky, and unsustained. Individual movements are discrete, but they are variable in type and location, causing an irregular pattern of chaotic, multifocal, constantly changing movements that seem to flow from one body part to another. The movements may at times appear purposeful to a casual observer, but they are actually random and aimless. They are present at rest but are increased by activity, tension, emotional stress, and self-consciousness. The patient may be able to temporarily and partially suppress the movements, and they disappear in sleep.



Video 30.4 Chorea in a patient with Huntington's disease.

The distribution of the choreic movements is variable. They may involve one extremity, one-half of the body (hemichorea), or be generalized. They occur most characteristically in the distal parts of the upper extremities but may also involve the proximal parts, lower extremities, trunk, face, tongue, lips, and pharynx. There may be repeated twitching and grimacing movements of the face that change constantly in character and location. Involvement of the vocal tract may cause abnormal vocalizations, difficulty in maintaining phonation, or aphonia.

The abnormal movements interrupt the harmonious coordination of prime movers, synergists, and antagonists. They interfere with and distort voluntary movements, and the latter may be short, jerky, and unsustained. Difficulty with rapid, repetitive movements and problems performing a sequence of hand movements indicate the disturbed motor function. Constant unwanted movements of the hands may interfere with activities of daily living. When

asked to hold the hands outstretched, there may be constant random movements of individual fingers (piano-playing movements). If the patient holds the examiner's finger in her fist, there are constant twitches of individual fingers (milkmaid grip). Chorea movements may often be brought out by having the patient carry out two simultaneous acts. She may touch finger to nose or protrude the tongue in isolation, but when attempting to do both at the same time, the jerky movements become noticeable. The patient may also have difficulty chewing and swallowing.

The patient may try to incorporate a spontaneous, involuntary movement into a semipurposeful movement in order to mask the chorea (parakinesia). If a choreic movement suddenly makes a hand fly upward, the patient may continue the movement and reach up and scratch her nose. When chorea is generalized, the patient is in a constant state of motion with continual adventitious movements randomly scattered.

In addition to the abnormal movements, there is hypotonia of the skeletal muscles, with decreased resistance to passive movement. The outstretched hands are held with hyperextension of the fingers with flexion and dorsal arching of the wrist (spooning). The fingers are separated and the thumb is abducted and droops downward. When the arms are raised overhead, the hands may turn into a position of hyperpronation. Motor impersistence—the inability to sustain a contraction—frequently accompanies chorea. The patient is frequently unable to hold the tongue out for any length of time; when asked to do so, the tongue shoots out and then jerks back quickly (snake, darting, flycatcher, or trombone tongue). The blink rate is increased.

There is no paralysis, but the hypotonia and constantly repeated hyperkinesias may interfere with voluntary movement enough to cause significant impairment of motor function. The hypotonia may result in pendular deep tendon reflexes. Many disorders may cause chorea, among them Huntington's disease (HD) and Sydenham's chorea.

Huntington's Disease

HD (Huntington's chorea) is a progressive, fatal, AD, neurodegenerative condition with variable penetrance, because of an unstable CAG trinucleotide repeat expansion on chromosome 4. The gene produces a protein called huntingtin. The mutant protein results in an expanded polyglutamine tract and a buildup of deleterious β -amyloid-like aggregates, which primarily affect the

caudate and putamen. The onset is usually between the ages of 35 and 50, but the disease may begin in childhood or senescence. Onset is often earlier when the disease is inherited from the father. Age of onset correlates inversely with the length of trinucleotide repeats. Repeats tend to lengthen in succeeding generations, causing earlier onset (anticipation), particularly with paternal transmission. DNA diagnostic testing can confirm the diagnosis, even in presymptomatic individuals.

Huntington described three cardinal features: dementia, emotional disturbance, and the familial nature of the disorder. The condition is inexorably progressive and ultimately fatal. The typical course is from 15 to 20 years. Some symptomatic treatments are available, but nothing which will arrest the progression. The choreic movements may cause the patient to complain of clumsiness or tremor. The movements are similar to those of Sydenham's chorea but are often somewhat slower, less jerky, and more bizarre, widespread, and violent. They may be seemingly purposeful, and the same pattern may be repeated over and over. Frequently, the larger muscle groups and the proximal extremities are affected; there may be repeated shrugging of the shoulder or flail-like movements of the arm and twisting and lashing movements that lie between those of chorea and athetosis. Facial grimacing may be marked. Movements of the fingers and hands are often accentuated as the patient walks. Pronounced chorea of the arms and legs when walking may lead to a bizarre, prancing gait (see [Video Link 30.2](#)). In the later stages of the disease, the chorea may progress to athetosis or dystonia. Rigidity may become a conspicuous feature in the later stages of choreic disease. In juvenile HD, with onset prior to age 20, rigidity is often more prominent than chorea (Westphal, rigid, or pseudoparkinsonian variant).

HD is accompanied by progressive intellectual deterioration. Cognitive impairment usually begins at about the same time as the abnormal movements but may precede it and progresses in tandem. Most patients also develop psychiatric abnormalities, particularly personality changes and mood disorders. The degree of dementia is out of proportion to the cortical pathology and may reflect impairment related to the role of the caudate in cognition. Patients are typically reduced to a vegetative state about 10 to 15 years after onset.

Neuropathologically, there is atrophy most prominent in the caudate, putamen, and cerebral cortex. The neuron loss initially affects the enkephalinergic (ENKergic) spiny neurons of the striatum. Loss of ENKergic striatal neurons lessens the inhibitory influence of the striatum on the external

segment of the globus pallidus, allowing it to increase its inhibition of the subthalamic nucleus. This results in a decrease in the facilitatory influence of the subthalamic nucleus on the internal segment of the globus pallidus, decreasing the inhibition of the motor thalamus (releasing the brake), increasing thalamocortical activity, and resulting in hyperkinesia (see [Chapter 26](#)). Imaging studies may show atrophy of the caudate, producing a square-shaped lateral ventricle ([Figure 30.6](#)). The cortical atrophy primarily affects the frontal and temporal regions. HD-like 1 and 2 are hereditary disorders clinically and pathologically very similar to HD.

Other Forms of Chorea

Sydenham's chorea (chorea minor, rheumatic chorea, St. Vitus' dance) occurs in childhood and adolescence in relationship to streptococcal infection. Like rheumatic fever, it has become a rarity in developed countries. After recovery, a patient may retain minimal choreic movements or have minor tic-like movements that are difficult to differentiate from chorea. Some of the other conditions associated with chorea include chorea gravidarum (occurs during pregnancy), systemic lupus erythematosus, antiphospholipid syndrome, neurosyphilis (usually with concomitant HIV infection), hyperthyroidism, polycythemia vera, nonketotic hyperglycemia, adult polyglucosan body disease, Behçet's disease, and neuroacanthocytosis. In children, chorea may occur after cardiac surgery (postpump chorea). Sydenham's chorea may recur, at times, as chorea gravidarum. There is also a form of nonprogressive chorea that is inherited as a recessive trait (benign hereditary chorea).

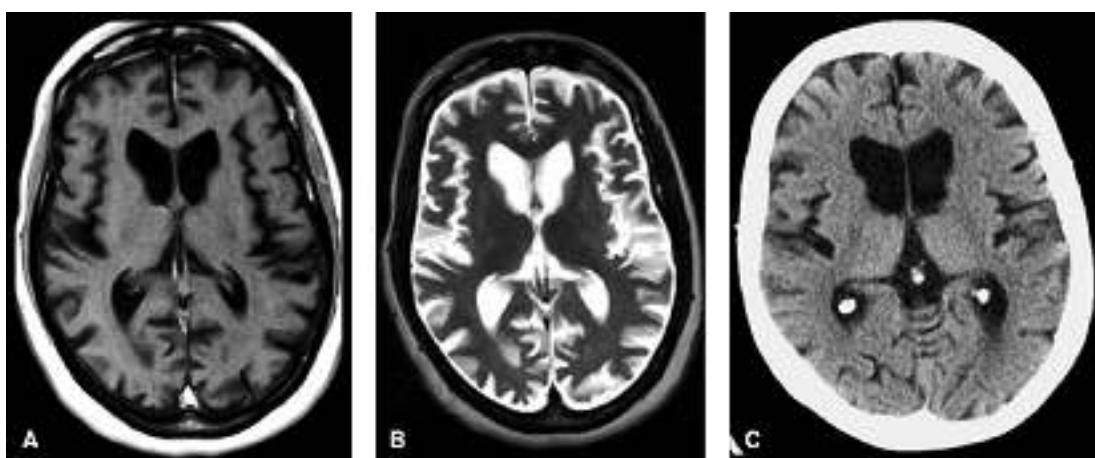


FIGURE 30.6 T1-weighted MRI (A), T2-weighted MRI (B) and axial CT (C) images show atrophy of the caudate and frontal horn enlargement. (From Sethi KD. Magnetic

resonance imaging in Huntington's disease. *Mov Disord* 1991;6[2]:186. Copyright © 1991 Movement Disorder Society. Reprinted by permission of John Wiley & Sons, Inc.)

Structural lesions of the basal ganglia, such as infarct, neoplasm, or trauma, may occasionally cause chorea. It may occur with inborn errors of metabolism, such as Lesch-Nyhan syndrome, Niemann-Pick disease, and gangliosidosis. Chorea is nearly universal in the neuroacanthocytosis syndromes, in which various neurologic abnormalities are associated with the presence of acanthocytes on peripheral smear. Some patients have a genetic abnormality involving the protein chorein. Others may have decreased or absent (Bassen-Kornzweig disease) beta-lipoproteins. Neuroferritinopathy is an AD disorder in which various extrapyramidal features, including chorea, develop in the third to fifth decade. It is often mistaken for HD. Low serum ferritin is common but not invariable.

Chorea may be a transient side effect of many medications, such as psychotropic agents, phenytoin, antihistamines, levodopa, methylphenidate, lithium, oral contraceptives, estrogen, tricyclic antidepressants, isoniazid, and others. Drugs of abuse may cause chorea, including cocaine ("crack dancing") and amphetamines. Cocaine abuse is also associated with other movement disorders, including dystonia, exacerbation of Tourette's syndrome, multifocal tics, opsoclonus-myoclonus, and stereotyped behavior. Chorea may be a persisting feature of past or present exposure to psychoactive drugs as part of the syndrome of tardive dyskinesia. It may be a remote effect of carcinoma or be part of multisystem familial degenerative disorders. Hemichorea may follow structural lesions of the contralateral basal ganglia (see Hemiballism).

ATHETOSIS

In athetosis (Hammond's disease), the hyperkinesias are slower, more sustained, and larger in amplitude than those in chorea. They are involuntary, irregular, coarse, somewhat rhythmic, and writhing or squirming in character. They may involve the extremities, face, neck, and trunk. In the extremities, they affect mainly the distal portions, the fingers, hands, and toes. The movements are characterized by any combination of flexion, extension, abduction, pronation, and supination, often alternating and in varying degrees ([Figure 30.7](#)). They flow randomly from one body part to another, and the direction of movement changes

randomly. The affected limbs are in constant motion (athetosis means “without fixed position”). Hyperextension of the fingers and wrist and pronation of the forearm may alternate with full flexion of the fingers and wrist and supination of the forearm. Facial grimacing, slower and more sustained than in chorea, often accompanies the movements of the extremities, and there may be synkinesias affecting other parts of the body. The hyperkinesias may not be constant or continuous. The movements can often be brought out or intensified by voluntary activity of another body part (overflow phenomenon). They disappear in sleep. Voluntary movements are impaired and coordinated action may be difficult or impossible. Athetosis is usually unilateral; bilateral involvement is called double athetosis.

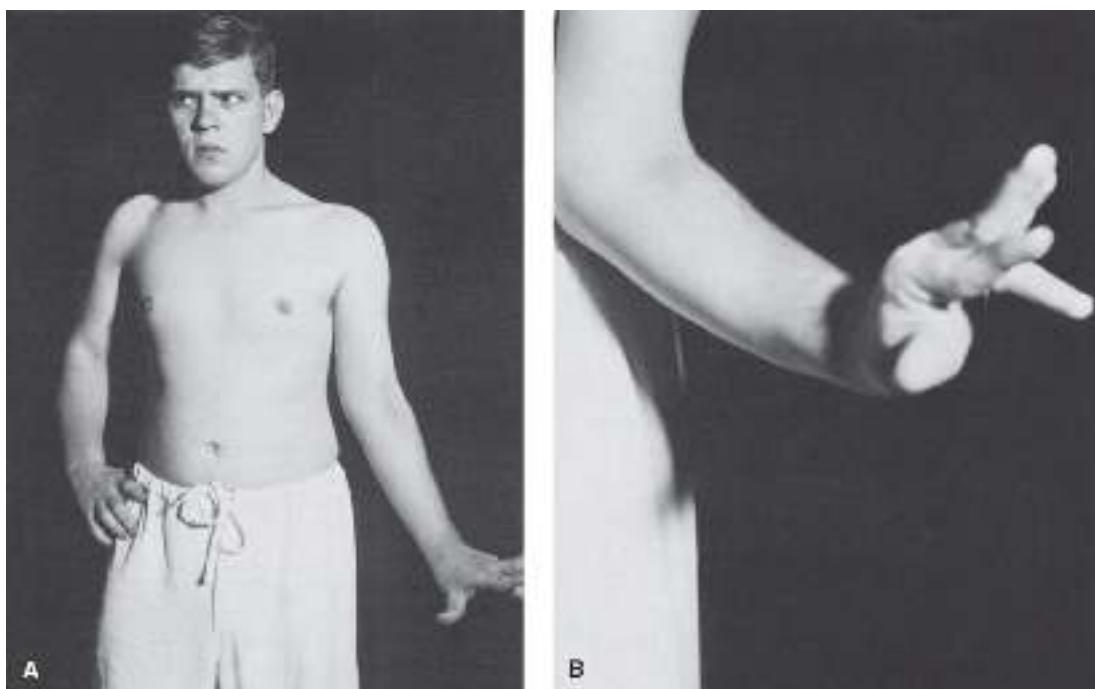


FIGURE 30.7 A and B: A patient with congenital unilateral athetosis.

Athetosis is usually congenital, the result of perinatal injury to the basal ganglia, and may be present in association with other neurologic deficits (athetotic cerebral palsy). It may be either unilateral or bilateral. The predominant pathologic changes are in the caudate and putamen, although there may also be cortical involvement. Double athetosis may be associated with status marmoratus of the basal ganglia, which is usually due to anoxic birth injury. Acquired athetosis may follow disease or trauma in later life. Many of its causes overlap with those of chorea, and in fact, many patients have features of

athetosis plus chorea. Choroathetosis refers to movements that lie between chorea and athetosis in rate and rhythmicity and may represent a transitional form. Slow athetoid movements begin to blend with dystonia. Pseudoathetosis (sensory athetosis) is a term used to describe similar undulating and writhing movements of the extremities because of loss of position sense as a result of a parietal lobe lesion or peripheral deafferentation because of such conditions as tabes dorsalis, posterolateral sclerosis, and peripheral nerve disease ([Figure 30.8](#)). The movements are more marked when the eyes are closed and are usually unassociated with an increase in muscle tone.



FIGURE 30.8 Pseudoathetosis of the hand in a patient with a parietal lobe lesion.

DYSTONIA

Dystonia refers to spontaneous, involuntary, sustained muscle contractions that force the affected parts of the body into abnormal movements or postures, sometimes with cocontraction of agonists and antagonists. Dystonia often affects the extremities, neck, trunk, eyelids, face, or vocal cords. It may be either constant or intermittent and generalized, segmental, focal, multifocal, or in a hemidistribution. Dystonic movements are patterned, tending to recur in the same location, in contrast to the random and fleeting nature of chorea. The speed of dystonia varies widely, from slow, sustained, and cramp-like (athetotic

dystonia) to quick and flicking (myoclonic dystonia). When the duration is very brief (less than 1 second), the movement may be referred to as a dystonic spasm, when more sustained (several seconds) as a dystonic movement, and when prolonged (minutes to hours) as a dystonic posture. Occasionally, dystonia is associated with rapid rhythmic tremulous movements (dystonic tremor). Action dystonia occurs when carrying out a voluntary movement. As in athetosis, overflow may occur, with the dystonia brought out by use of another part of the body.

Generalized dystonia causes involuntary movements similar in many respects to athetosis but involving larger portions of the body, often producing distorted postures of the limbs and trunk. The movements are slow, bizarre, and sometimes grotesque, with an undulating, writhing, twisting, turning character, and a tendency for the contraction to be sustained at the peak of the movement (torsion dystonia, torsion spasm). Generalized dystonia may start distally, usually in the foot, with plantarflexion and inversion, and then spread to the opposite side, the upper extremities, the trunk, face, and tongue. In severe cases, there are writhing movements of the shoulder muscles, hip girdles, and trunk. There is peculiar, axial twisting of the spine, causing marked torsion of the entire vertebral column with lordosis, scoliosis, and tilting of the shoulders and pelvis. Dysarthria, facial grimacing, and torticollis may also be present. The muscles are often in a constant state of hypertonicity, and the muscular contractions may cause severe pain. The movements are involuntary but are increased by voluntary activity and emotion. Eventually, postures become fixed by contractures, and deformities develop. The term dystonia is sometimes used to describe the postures or positions assumed by the patient, as well as for the hyperkinesia itself. Dystonia may be intermittent or paroxysmal, lasting minutes.

Dystonia musculorum deformans (DMD, idiopathic torsion dystonia) is a rare progressive disease that usually begins in childhood. DMD-1 (DYT1 dystonia) is AD, and DMD-2 (DYT2 dystonia) is recessive; numerous other genetic forms of dystonia exist. Dystonia also occurs in Wilson's disease, acquired hepatocerebral degeneration, PKAN, kernicterus, in HD as it progresses, in PD, and occasionally with structural lesions affecting the basal ganglia. Dystonia can occur as a drug side effect, particularly as a dose-related manifestation of treatment with levodopa and other dopaminergic drugs. Other offending agents include cimetidine, anticonvulsants, calcium channel blockers, and anxiolytics. Tardive dystonia is related to treatment with phenothiazines and other psychotropic drugs. Dopa-responsive dystonia (Segawa's disease and many

other names) is a distinctive and common type of generalized dystonia that presents in childhood or adolescence and is characterized by marked diurnal variation in severity and exquisite responsiveness to small doses of levodopa. It is a consideration in the differential diagnosis of diplegic cerebral palsy, sporadic spastic paraparesis, hereditary spastic paraparesis, and JPD. Misdiagnosis is frequent. Hemidystonia is along the spectrum of hemichorea and hemiballismus but because of a lesion of the contralateral striatum.

The focal dystonias are disorders causing involuntary contractions in a limited distribution. A relatively common form of focal dystonia is cervical dystonia (spasmodic torticollis; [Chapter 19](#)), which affects the neck, and sometimes the shoulder, muscles, producing either a sustained or jerky turning of the head to one side, often with some element of head tilt. “Torti” implies a twisting or turning movement; less common variants of cervical dystonia include retrocollis (extension movement) and anterocollis (flexion movement). In the beginning, the twisting and turning may be intermittent or present only in paroxysms (spasmodic), but later in the course of the syndrome, there is persistent contraction of the involved muscles with resulting deviation of the head. Many if not most patients with cervical dystonia learn they can straighten their head by placing a hand or finger somewhere on the face or performing some other maneuver to provide sensory stimulation or light counterpressure (*geste antagoniste*, sensory trick, counterpressure sign; [Figure 30.9](#)). Notoriously refractory to medical therapy, cervical dystonia is now often treated by the injection of infinitesimally small amounts of botulinum toxin to weaken the abnormally contracting muscles.

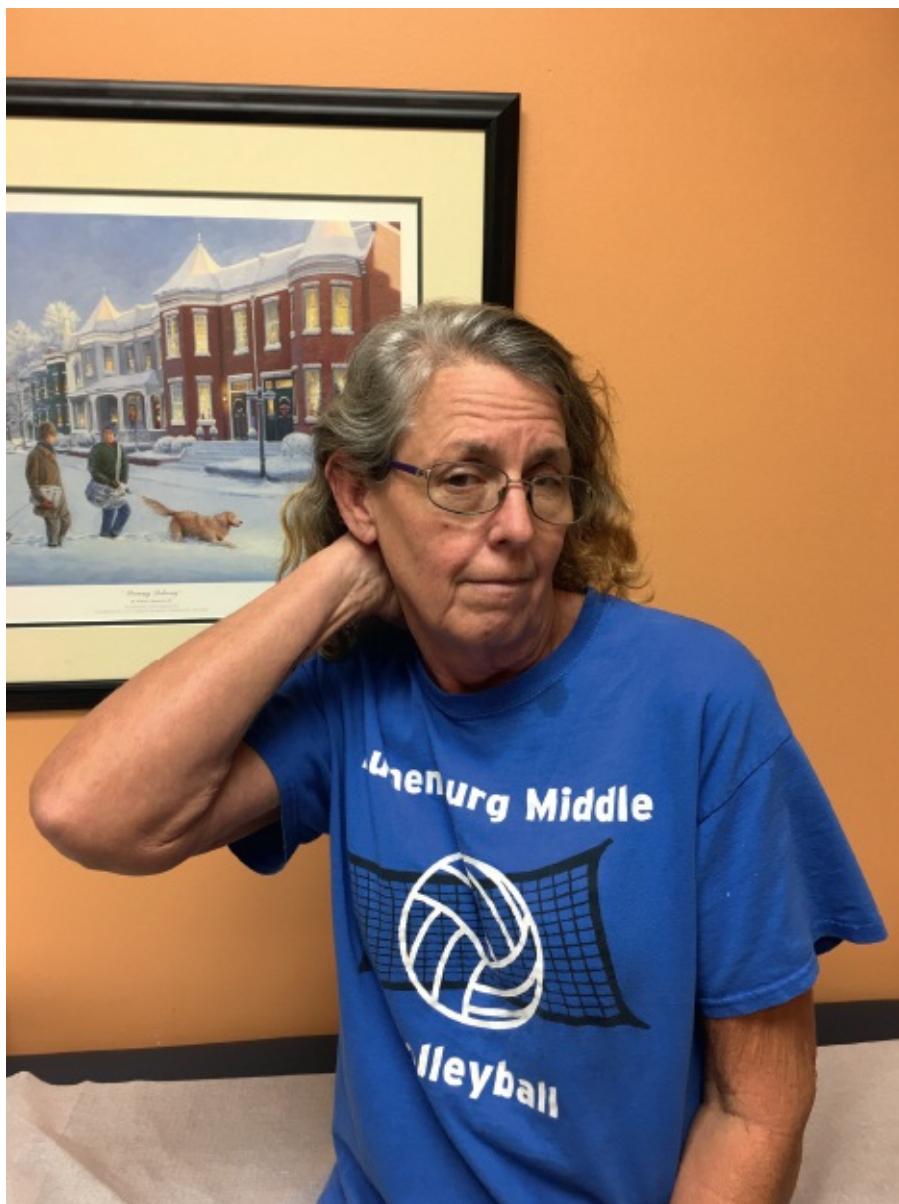


FIGURE 30.9 Sensory trick in cervical dystonia. The patient found that keeping her right hand behind the neck helped keep her head straight. She kept the right elbow flexed so constantly she developed a right ulnar neuropathy at the elbow.

Writer's cramp (graphospasm) is a focal dystonia of the hand or forearm muscles brought on by use of the part, most frequently by writing. There are a number of other focal, occupational, or task-specific dystonias related to specific activities. Musicians may develop hand or embouchure dystonia. The *yips* in golfers may be a task-specific dystonia. Blepharospasm (upper facial dystonia) causes involuntary closure of both eyes. The spasms may be brief or sustained. Patients with sustained spasms become functionally blind during the paroxysms.

Involuntary closure of one eye is usually due to early hemifacial spasm. Oromandibular dystonia involves the mouth, lips, and jaw. The combination of blepharospasm and oromandibular dystonia constitutes Meige's (Brueghel's) syndrome ([Video 30.5](#)). Auctioneer's jaw is a task-specific oromandibular dystonia. Spasmodic dysphonia is dystonia of the vocal cords (see [Chapter 9](#)). Belly dancer's dystonia refers to involuntary movements affecting the abdominal musculature. Focal dystonias of peripheral origin have been described in relation to nerve, plexus, or nerve root lesions. A segmental dystonia is more extensive than a focal dystonia and involves contiguous body regions (e.g., cervical dystonia accompanied by oromandibular dystonia).



Video 30.5 Examples of blepharospasm and Meige's syndrome (blepharospasm with oromandibular dystonia). (Courtesy Dr. Stephen G. Reich.)

HEMIBALLISMUS

Hemiballismus (hemiballism) refers to a dramatic neurologic syndrome of wild, flinging, incessant movements that occur on one side of the body ([Video 30.6](#)). It is classically due to infarction or hemorrhage in the region of the contralateral subthalamic nucleus; the lesion results in disinhibition of the motor thalamus and the cortex, resulting in contralateral hyperkinetic movements. Case series with modern neuroimaging have shown that only a minority of cases have lesions in the subthalamic nucleus. Nonketotic hyperglycemia is also a common etiology. The ballistic movements of hemiballismus resemble those of chorea but are more pronounced. The clinical distinction between severe hemichorea and hemiballismus becomes arbitrary. Like chorea, hemibalistic movements are involuntary and purposeless movements, but they are much more rapid and

forceful and involve the proximal portions of the extremities. When fully developed, there are continuous, violent, swinging, flinging, rolling, throwing, and flailing movements of the involved extremities. The movements are ceaseless during the waking state and disappear only with deep sleep. They are usually unilateral and involve one entire half of the body; their intensity may cause movement of the entire body. Rarely, they are bilateral (biballismus or paraballismus) or involve a single extremity (monoballismus). The movements may spare the face and trunk. Hemiballismus is difficult to treat, incredibly disabling, and sometimes fatal because of exhaustion and inanition.



Video 30.6 Hemiballismus. The movements were unremitting and medically intractable but resolved after pallidotomy. (From Suarez JI, Metman LV, Reich SG, et al. Pallidotomy for hemiballismus: efficacy and characteristics of neuronal activity. *Ann Neurol* 1997;42:807–811.) (Courtesy Dr. Stephen G. Reich.)

DYSKINESIAS

All hyperkinetic movements are technically dyskinesias, but the term is often used to encompass complex involuntary movements that do not neatly fit into another category. Dyskinesia is used most often to refer to abnormal involuntary movements related to drugs. Dyskinesias are a common dose-related complication of the treatment of PD with levodopa and dopamine agonists. In some disorders, the dyskinesias occur paroxysmally. Paroxysmal dyskinesias strike suddenly and unexpectedly when the patient is engaged in otherwise normal motor behavior. The dyskinesias may be precipitated by movement (paroxysmal kinesigenic dyskinesia) or by other factors, such as stress, heat, or

fatigue (paroxysmal nonkinesigenic dyskinesia). Paroxysmal hypnogenic dyskinesias occur during sleep.

OROFACIAL DYSKINESIAS

Orofacial dyskinesias are involuntary movements of the mouth, face, jaw, or tongue that may consist of grimacing, pursing of the mouth and lips, “fish-gaping” movements, and writhing movements of the tongue. These often develop as tardive dyskinesias (TD) after the use of phenothiazines and other psychotropic drugs.

TD are involuntary movements that usually develop in patients who have received phenothiazines or related compounds, usually as treatment for major psychosis, for prolonged periods. Dyskinesias may also occur shortly after initiation of psychotropic therapy and may be associated with the use of other drugs. The movements typically involve primarily the mouth, tongue, and jaw with incessant chewing, smacking, licking, and tongue-thrusting movements that are difficult to eradicate. Some patients are unaware they have these movements. It seems likely that long-term dopamine receptor blockade leads to denervation hypersensitivity of the receptor. The movements not infrequently first appear when the dose of dopamine-blocking agent is reduced and can often be controlled, at least temporarily, by reinstating or increasing the dose of the drug. TD are more prone to develop in older patients, especially women. Unfortunately, the term TD is often used for all involuntary orofacial movements, which may develop with no drug exposure, especially in older or edentulous patients. Other abnormal movements may arise as a tardive phenomenon, including tremor, dystonia, akathisia, tics, and chorea. Rabbit syndrome refers to a rhythmic perioral tremor, generally associated with the use of psychotropics. The movements are more rapid and regular and do not involve the tongue, helping distinguish them from TD.

MYOCLONUS

The term myoclonus has been used for several differing motor phenomena. In general, myoclonus may be defined as single or repetitive, abrupt, brief, rapid, lightning-like, jerky, arrhythmic, asynergic, involuntary contractions, involving portions of muscles, entire muscles, or groups of muscles. The movements are

quicker than chorea. Myoclonus is seen principally in the muscles of the extremities and trunk, but the involvement is often multifocal, diffuse, or widespread. It may involve the facial muscles, jaws, tongue, pharynx, and larynx. There may be successive or simultaneous involvement of many muscles. Myoclonus may appear symmetrically on both sides of the body; such synchrony may be an attribute unique to myoclonus. The sudden, shock-like contractions usually appear in paroxysms at irregular intervals, during either the resting or active state, and may be activated by emotional, mental, tactile, visual, and auditory stimuli. Myoclonic movements, like fasciculations and myokymia, occasionally are too weak to cause joint movement. More often, they affect entire muscles or muscle groups, producing clonic movements of the extremities. They may be so violent as to cause an entire limb to be suddenly flung out and may even throw the patient to the ground. An excessive startle response causing a massive whole-body myoclonic jerk occurs in some conditions, especially Creutzfeldt-Jakob disease (see “Hyperekplexia”). Myoclonus may also be subtle, a quick flick of a finger or foot.

Myoclonus has been classified in numerous ways, including the following: positive versus negative, epileptic versus nonepileptic, stimulus sensitive (reflex) versus spontaneous, rhythmic versus arrhythmic, anatomically (peripheral, spinal, segmental, brainstem, or cortical), and by etiology (physiologic, essential, epileptic, and symptomatic). Asterixis may be viewed as negative myoclonus, the transient, unwanted, abnormal relaxation of a muscle group. As typically used, the term myoclonus refers to positive myoclonus: abnormal jerks. Cortical reflex myoclonus is focal myoclonus triggered by stimulation or movement of the affected part.

Myoclonic movements may occur in a variety of conditions, and their significance varies. Physiologic myoclonus occurs in normals. Sleep starts (hypnic jerks) are myoclonic jerks that appear during the process of falling asleep but disappear during sleep. Hiccups are another form of physiologic myoclonus. In essential myoclonus, there are no accompanying abnormalities; it may be sporadic or familial. Paramyoclonus multiplex is a disorder of adult life causing paroxysmal contractions of the limb and trunk muscles. The etiology is not known, and the existence of the entity has been questioned.

Myoclonus is frequently encountered in epilepsy. Many epileptic patients have occasional random myoclonic jerks of the axial or proximal limb musculature, which may appear or increase in frequency immediately prior to a seizure. Beginning around puberty, patients with juvenile myoclonic epilepsy

(Janz syndrome) have generalized tonic-clonic seizures that are associated with frequent myoclonic jerks predominantly affecting the arms, especially on awakening. Massive myoclonic spasms of infancy are characterized by frequent, sudden, violent jerking attacks with flexion of the neck and trunk and adduction or abduction and extension of the arms and legs (infantile spasms, West syndrome). The body may bend forward (salaam or jackknife attacks).

The progressive myoclonic epilepsies are a group of disorders that include Unverricht-Lundborg syndrome and Lafora body disease. Progressive myoclonic epilepsy can also occur without Lafora bodies and in type 3 Gaucher's disease, sialidosis, gangliosidoses, ceroid lipofuscinoses, and myoclonic epilepsy with ragged red fibers. In dyssynergia cerebellaris myoclonica, one of the several Ramsay Hunt syndromes, myoclonus is accompanied by progressive cerebellar degeneration and seizures.

Myoclonus occurs without prominent seizures in a number of other conditions, including metabolic disorders (especially uremic and anoxic encephalopathy), subacute sclerosing panencephalitis, PKAN, Creutzfeldt-Jakob disease, Alzheimer's disease, Wilson's disease, HD, CBD, viral encephalitis, general paresis, Hashimoto's encephalopathy, and the lipidoses. Opsoclonus refers to random, chaotic, lightning-fast eye movements (see [Chapter 14](#)). Opsoclonus accompanied by myoclonus (opsoclonus-myoclonus syndrome, dancing eyes-dancing feet, Kinsbourne's syndrome) may occur as a postinfectious encephalopathy or as a paraneoplastic syndrome, especially because of occult neuroblastoma. Action myoclonus occurs with use of the involved limb. A syndrome of action or intention myoclonus may develop as a sequel to cerebral anoxia (Lance-Adams syndrome).

Myoclonus may sometimes be benign and without serious significance and even have a psychogenic basis. Movements difficult to differentiate from myoclonus may be nonorganic. Benign nocturnal myoclonus may occur in healthy persons. Paroxysmal kinesigenic myoclonus has also been described.

Myoclonus is typically arrhythmic and diffuse, but the term has also been applied to rhythmic and localized motor phenomena. Palatal myoclonus is characterized by involuntary, rhythmic movements of the soft palate and pharynx, sometimes of the larynx, eye muscles, and diaphragm, and occasionally of other muscles. The movements are generally not influenced by drugs or sleep. The palate may bounce up and down or twitch rhythmically to one side ([Video 30.7](#)). The posterior pharyngeal wall moves laterally, and the larynx moves in an upward and downward direction. Movements involving the diaphragm or larynx

may cause a grunting respiratory noise. Opening and closing of the eustachian tube sometimes causes a clicking sound accompanying the movements, audible to the patient and sometimes the examiner.



Video 30.7 Palatal myoclonus (microtremor). (Courtesy Dr. Jason Hawley.)

Palatal myoclonus occurs in essential and symptomatic forms. Symptomatic palatal myoclonus occurs with lesions involving the connections between the inferior olfactory, dentate, and red nuclei. The Guillain-Mollaret (myoclonic) triangle is a loop: inferior olive → inferior cerebellar peduncle → dentate nucleus → superior cerebellar peduncle → red nucleus → central tegmental tract → inferior olive. A lesion anywhere in this loop, most often brainstem infarction, may cause palatal myoclonus and its variants. Lesions of the central tegmental tract may cause hypertrophy (pseudohypertrophy) of the olive. There is gliosis of the amiculum of the olive, increasing the size of the olive grossly, which may be visualized by MRI.

Palatal myoclonus is also referred to as palatal microtremor. Tremors are due to alternating agonist-antagonist contractions, rhythmic myoclonus to contraction-relaxation cycles of an agonist. In addition, tremors usually disappear in sleep and these palatal movements do not. Whether palatal myoclonus is best characterized as rhythmic myoclonus or a tremor remains unclear.

ASTERIXIS

Seen primarily in metabolic encephalopathy, particularly hepatic encephalopathy, asterixis is an inability to sustain normal muscle tone ([Video](#))

30.8). With the arms outstretched and wrists extended, “like stopping traffic,” the lapse in postural tone may cause the hands to suddenly flop downward, then quickly recover, causing a slow and irregular flapping motion that led to the term “liver flap.” When severe, the entire arm may drop. Other body parts may exhibit the phenomenon (e.g., inability to keep the foot dorsiflexed [foot flap]). Unilateral asterixis may occur with focal brain lesions, particularly involving the contralateral thalamus. In metabolic encephalopathy, there may be a high-frequency postural tremor that begins after a latent period of 2 to 30 seconds and attenuates on movement (mini-asterixis, metabolic tremor). In unresponsive patients, asterixis at the hip joints can sometimes be brought out by passively flexing and abducting to hips so that the thighs form a “V.” In this position, the knees may flap up and down.



Video 30.8 Asterixis. The first segment shows asterixis of the hands, the second segment of the feet. (Courtesy Dr. Robert Laureno.)

MYORHYTHMIA

The term myorhythmia refers to slow (typically 2 to 3 Hz), rhythmic alternating movements that resemble tremor. The primary distinguishing characteristics are the slow rate and the often widespread involvement. It may be intermittent or continuous, synchronous, or asynchronous when involving multiple body parts and is absent during sleep. The movements may involve one or several limbs, the head, the eyes, or various combinations. Myorhythmia has been described in cerebrovascular disease, anti-NMDA receptor encephalitis, Hashimoto’s encephalopathy, and other disorders. Oculomasticatory myorhythmia refers to pendular vergence movements of the eyes synchronous with contractions of the

masticatory muscles. It is a distinctive movement disorder that appears to be specific for central nervous system (CNS) Whipple's disease (see [Video Link 30.3](#)).

TICS

The hyperkinesias discussed to this point have been involuntary movements. In another type of abnormal movement, the patient has some degree of awareness of the movement but must make a movement in response to the urge of some compelling inner force. The patient experiences tension and restlessness, which are temporarily relieved by making a particular movement. Such movements have been called "unvoluntary." Examples include tics, akathisia, stereotypies, compulsions, and restless legs.

Tics (habit spasms) are quick, irregular but repetitive movements that are more often seen in children than adults. A tiqueur is a person who is subject to one or multiple tics. A tic may be defined as a coordinated, repetitive, seemingly purposeful act involving a group of muscles in their normal synergistic relationships. Tics are stereotyped, recurrent movements that may seem purposeful but are relatively involuntary, consisting of brief contractions of whole muscles or groups of muscles, always accompanied by motion of the affected part. Patients are able to suppress the movements temporarily with concentration, but they quickly return when attention is diverted to some other task. Voluntary suppression causes a sense of intolerable mounting tension and an urge to move that is temporarily relieved by indulgence in a tic. Tics are exaggerated by emotional strain and tension; they cease during sleep.

Tics may involve any portion of the body. Common examples of simple motor tics include repetitive blinking, facial contortions, or shoulder shrugging. More complicated tics can occur, and tics can also involve the vocal tract (phonic or vocal tic), producing throat-clearing as well as bizarre vocalizations, such as barking and grunting or sounds resembling a hiccup.

Patients affected with Gilles de la Tourette syndrome (maladie des tics) have multifocal tics, compulsive behavior, imitative gestures, stereotyped movements, grunts and groans, and evidence of regressive behavior. There are explosive vocalizations, and the patient may utter profanity and obscenities over which they have no control (coprolalia). The condition has its onset in childhood and occurs most frequently in boys, usually in the preadolescent period. Tics are very

common and usually benign; patients with Tourette's syndrome have exaggerated, complex tics, which together with the other features of the disease can be very disabling (see [Video Link 30.4](#)). The large repertoire of tics and the combination of motor and vocal tics distinguish Tourette's syndrome from ordinary tics. The disease is hereditary, probably AD with variable expressivity, and related to some dysfunction of dopamine receptors. It is likely that the noted historical literary figure, Dr. Samuel Johnson, the famously eccentric author of the first English language dictionary, suffered from Tourette's syndrome.

AKATHISIA

Patients suffering from akathisia experience an inner restlessness and urge to move that causes them to remain in almost constant motion. It occurs most often as a result of treatment with dopamine-blocking agents. Patients with PD may experience akathisia but are not able to move in response to it.

STEREOTYPY

A stereotypy is a repetitive, purposeless but often seemingly purposeful, involuntary, patterned motor activity. Common foot shaking and other mannerisms are examples of simple stereotypies. More complex stereotypies may involve ritualistic behavior, such as the compulsions of obsessive-compulsive disorder. Stereotypies most commonly occur in psychiatric disorders: anxiety, obsessive-compulsive disorder, schizophrenia, autism, and mental retardation. They may also be a part of neurologic disorders, such as tardive dyskinesia and Tourette's syndrome. The hand-wringing in Rett's syndrome is a stereotypy. "Punding" refers to complex, purposeless, stereotyped behavior seen in cocaine and amphetamine abuse and in patients with PD disease treated with dopaminergic agents. Stereotypies may resemble motor tics but do not share the suppressibility, variability, or mounting-tension compulsion to make the movement. Mannerisms are somewhat more complicated and stereotyped and are usually carried out in a more leisurely manner. They may appear only under emotional stress or when the patient is engaged in some particular activity.

HYPEREKPLEXIA

Hyperekplexia (startle disease, pathologic startle) refers to disorders characterized by an excessive startle response in the absence of other evidence of neurologic disease, sometimes accompanied by echolalia, automatic behavior, or automatic obedience (see [Video Link 30.5](#)). It may be sporadic or hereditary. Colorful names have been used for variants of the condition described in different geographic regions (jumping Frenchmen of Maine, latah, myriachit). An exaggerated startle response may also occur in Creutzfeldt-Jakob disease, Tay-Sachs disease, stiff-person syndrome, and lipidoses.

SLEEP-RELATED DYSKINESIAS

Except for palatal myoclonus, involuntary movements generally do not occur during sleep. There are some disorders, however, that occur primarily during sleep. Restless legs syndrome (RLS, Ekbom's syndrome, fidgety feet, jimmy legs) is a common disorder causing unpleasant and difficult-to-describe sensations in the legs that are temporarily relieved by movement. The symptoms commonly occur at night as the patient is drifting off to sleep. Many affected individuals get up and walk around to obtain respite. The patient support newsletter is NightWalkers. In many patients, RLS is accompanied by spontaneous movements of the legs during sleep (periodic movements in sleep, nocturnal myoclonus), best documented by polysomnography. The disorder is likely due to a central disturbance of dopamine metabolism. The restless red legs syndrome is RLS associated with telangiectasias of the legs. Patients with spinal stenosis may have leg pain in recumbency with similarities to RLS (vespers curse; [Chapter 47](#)). Restless arms and restless abdomen may also occur.

FASCICULATIONS

Fasciculations are fine, rapid, flickering or vermicular twitching movements due to contraction of a bundle, or fasciculus, of muscle fibers. They are usually not extensive enough to cause movement of joints, except occasionally the digits. They vary in size and intensity, from so faint and small as to only slightly ripple the surface of the overlying skin to coarse and impossible to overlook. They are random, irregular, fleeting, and inconstant. At times, they are abundant; at other

times, they require a careful search. Fasciculations often seem to strike where the examiner is not looking and are usually seen from the corner of the eye.

Fasciculations are brought out by fatigue and cold. When assessing fasciculations, the patient should be warm, comfortable, and completely relaxed. It is written that fasciculations may be brought out by mechanical stimulation of the muscle (e.g., light tapping), and neurologists frequently engage in a ritual of tapping a muscle with a reflex hammer and peering intently for a resultant fasciculation. Whether this really occurs is debatable. Good light is necessary in order to visualize fasciculations; oblique lighting is best. They may be more difficult to see in women than men because of the overlying subcutaneous fat. When not visible, they can occasionally be palpated or heard with a stethoscope. Many patients are unaware of fasciculations; others may see or feel them, or both. Needle electromyography can detect their presence even when they cannot be seen. High-resolution sonography may reveal fasciculations not visible from the surface. Fasciculations continue in sleep. They are exaggerated by the administration of cholinergic drugs (e.g., pyridostigmine). Hypercaffeinism is a common cause of fasciculations in normal individuals. At one time, the terms fibrillation and fasciculation were used synonymously. But fibrillation potentials are the contractions of single muscle fibers too small to be visible through the skin; they can only be detected by needle electromyography. Fasciculations are contractions of a large group of fibers, all or part of a single motor unit. Fasciculations are much more gross than fibrillations and can be seen through intact skin.

Fasciculations are a characteristic feature of motor neuron disease ([Video 30.9](#)). They serve as a very useful marker for the disease, and the diagnosis should remain circumspect when fasciculations are not present. Their exact mechanism remains debatable. Fasciculations were once thought to represent the dying gasps of sick motor neurons, but current evidence indicates they more likely arise much more distally in the neuron, perhaps in immature, unstable peripheral sprouts. In amyotrophic lateral sclerosis (ALS), abundant fasciculations may be an indication that disease progression will be rapid. Fasciculations of small hand muscles in chronic anterior horn cell disease, particularly spinal muscular atrophy, may cause small amplitude, subtle finger twitches called minipolymyoclonus, which are of course not real myoclonus.



Video 30.9 Fasciculations in a patient with end-stage amyotrophic lateral sclerosis.

Although fasciculations are most characteristic of motor neuronopathies, they can occur in any chronic denervating process, including radiculopathy and peripheral neuropathy. Fasciculations can also occur when anterior horn cells are involved in intrinsic spinal cord disease, such as syringomyelia or tumor. Except for thyrotoxicosis, myopathies generally do not cause fasciculations. In chronic denervating disease resulting in an enlarged motor unit territory, slight muscle contraction may activate a larger than normal number of muscle fibers causing a visible twitch referred to as a contraction fasciculation. These do not have the same significance as spontaneous fasciculations. In Kennedy's disease (bulbospinal muscular atrophy), contraction fasciculations occur in the chin with slight pursing of the lips. Contraction fasciculations may be seen occasionally in normal individuals, especially in the small hand muscles.

Fasciculations unaccompanied by atrophy or weakness do not necessarily indicate the presence of a serious disease process. About 70% of the population, especially health care workers, have occasional benign fasciculations. Some patients, most often older men, have prominent fasciculations without other abnormality. These most often occur in the calves, and the patients are quite aware of the movements, whereas most patients with ALS seem surprisingly oblivious to their fasciculations. The clinical examination is otherwise normal, and needle electromyography is normal except for the fasciculations. There is no infallible way to distinguish benign from malignant fasciculations from the fasciculations alone; judgment is made by the company they keep. A nonprogressive course over time is more reassuring than a single normal electrodiagnostic evaluation. Of 121 patients with benign fasciculations followed

up to 32 years, none developed ALS. In another report, 6.7% of ALS patients had fasciculations as an isolated, initial manifestation of the disease. Cramp-fasciculation syndrome is a syndrome of cramps and fasciculations because of hyperexcitability of the peripheral nerve.

MYOKYMA

Myokymia (Gr. *kyma*, “wave”) refers to involuntary, spontaneous, localized, transient or persistent quivering movements that affect a few muscle bundles within a single muscle but usually are not extensive enough to cause movement at a joint. The movements are somewhat coarser, slower, and undulating (“wormlike”), usually more prolonged, and involve a wider local area than fasciculations. They usually are not affected by motion or position, and they persist during sleep. On needle electromyography, clinical myokymia is accompanied by electrical discharges, either myokymic discharges or, less often, neuromyotonic discharges. Myokymic discharges, the electrical phenomenon, may or may not be accompanied by clinical myokymia, the visible, vermicular undulations on the skin surface.

Myokymia often occurs in normal individuals, causing persistent, focal twitching of a muscle, most commonly the orbicularis oculi. Myokymia usually occurs in isolation, without evidence of an accompanying neurologic disease; it is exacerbated by fatigue, anxiety, and caffeine. Myokymia in normal individuals and benign fasciculations may represent similar alterations in muscle physiology. Myokymia occurs in a variety of disease states; it is thought to arise because of biochemical perturbations in the nerve microenvironment because of demyelination, a toxin (such as rattlesnake venom or gold salts), edema, a decrease in ionized Ca^{++} concentration, or other factors (see [Video Link 30.6](#)). The generator lies somewhere along the motor axon.

Myokymia may be generalized or focal/segmental. Focal myokymia is much more common than generalized myokymia. The superior oblique muscle may develop episodic twitching producing a low-amplitude monocular intorsional movement (microtremor) that may cause annoying monocular oscillopsia and diplopia. It may be a microvascular compression syndrome with contact between the trochlear nerve and a vascular structure that may be seen by high-resolution thin slice magnetic resonance images. Myokymia sometimes occurs in the facial muscles in patients with MS or other lesions of the brainstem or cranial nerves,

such as pontine glioma, syrinx, or Guillain-Barré syndrome. Facial myokymia is usually transient but may persist for long periods when due to channelopathy or a structural lesion, such as pontine glioma or syringobulbia. Other abnormal facial movements, including synkinesias because of aberrant facial nerve regeneration and hemifacial spasm, are discussed in [Chapter 16](#). Focal limb myokymia is particularly characteristic of radiation damage to a nerve or plexus. Myokymia is only rarely associated with nerve compression syndromes. Myokymia may be seen, along with fasciculations, in motor neuronopathies. The pacemaker site varies with the condition. The response pattern of myokymic discharges to sleep, anesthesia, nerve blocks, and curare suggests a distal origin in many instances.

Generalized myokymia (Isaacs' syndrome, syndrome of continuous muscle fiber activity, neuromyotonia) causes generalized muscle stiffness and persistent contraction because of underlying continuous muscle fiber activity. Needle electromyography discloses spontaneous repetitive firing of motor unit potentials, creating myokymic and neuromyotonic discharges. Morvan's syndrome (Morvan's fibrillary chorea) is a dubious entity also associated with clinical myokymia. Generalized myokymia also occurs in episodic ataxia with myokymia.

STIFF-PERSON SYNDROME

Stiff-person (stiff-man, Moersch-Woltman) syndrome is due to hyperexcitability of anterior horn cells related to interference with gamma-aminobutyric acid–mediated spinal cord inhibitory mechanisms. There is progressive, often painful rigidity, punctuated by intense muscle spasms, particularly affecting the axial and paraspinal muscles. The axial rigidity causes hyperlordosis and prominent paraspinal muscle contractions. Superimposed on the stiffness are spasms provoked by movement or external stimuli. Most patients have antibodies to glutamic acid decarboxylase. In stiff-limb syndrome, symptoms are limited to one extremity. Jerking stiff-person syndrome is characterized by muscles stiffness and spasms affecting the legs. Progressive encephalomyelitis with rigidity and myoclonus causes a clinical picture similar to classic stiff-person syndrome but is more rapidly progressive.

SPASMS

Spasms are involuntary contractions of a muscle or group of muscles. The tonic contraction may cause either alteration of position or limitation of movement. They may occur in almost any muscle. A painful, tonic, spasmotic muscular contraction is often spoken of as a cramp. Spasms that limit movement may be defensive or protective. Prolonged spasm may cause reflex rigidity or be followed by muscle contracture. Spasms are often of reflex origin because of peripheral irritation affecting either muscles or nerves. Pain is a common cause of defensive spasm and reflex rigidity. Muscle spasm may also be voluntary or occur in response to fear or excitement. Carpopedal spasm is a common manifestation of tetany and hyperventilation ([Chapter 52](#)). Muscle spasms may also result from central processes. Prolonged and severe muscle spasms occur in tetany and tetanus. Characteristic painful tonic spasms occur in MS and stiff-person syndrome.

OTHER HYPERKINESIAS

In the painful legs and moving toes syndrome, there are continuous, involuntary movements of the toes associated with pain in the legs (see [Video Link 30.7](#)). The condition is sometimes a manifestation of peripheral neuropathy, but the responsible lesion in many is not clear. Variants are painful arms and moving fingers and painless legs and moving toes. Jumpy stump is involuntary movements of an amputated limb.

FUNCTIONAL MOVEMENT DISORDERS

Functional (psychogenic, nonorganic) movement disorders can simulate virtually any type of movement disorder. Functional disorders do not correspond to any of the organic types of abnormal involuntary movement; they are bizarre, change in type from time to time, and are influenced by emotional state and suggestion. Onset is often sudden. If a movement disorder is bizarre and defies classification, the possibility that it may be psychogenic should be borne in mind. Peculiar motor behaviors occur frequently in major psychiatric illnesses such as schizophrenia. However, being bizarre and difficult to characterize does not necessarily mean a movement disorder is psychogenic.

Psychogenic tremor is typically complex and does not fit well into the tremor classification scheme. The patient may have rapidly changing clinical features