Chapter 13: Control of Movement

CONTROL OF MOVEMENT

In more advanced forms of animal life, reflexive motion is based on the transmission of impulses from a receptor through an afferent neuron and ganglion cell to motor neurons and muscles. This arrangement is found in the reflex arc of higher animals, including humans, in whom the spinal cord has further developed into a central regulating mechanism. Superimposed on these reflex circuits, the brain is concerned with the initiation and control of movement and the integration of complex motions.

Control of Movement in Humans

The motor system in humans controls a complex neuromuscular network. Commands must be sent to many muscles, and multiple ipsilateral and contralateral joints must be stabilized. The motor system includes cortical and subcortical areas of gray matter; the corticobulbar, corticospinal, corticopontine, rubrospinal, reticulospinal, vestibulospinal, and tectospinal descending tracts; gray matter of the spinal cord; efferent nerves; and the cerebellum and basal ganglia (Figs 13–1 and 13–2). Feedback from sensory systems and cerebellar afferents further influences the motor system.

FIGURE 13–1
Schematic illustration of some pathways controlling motor functions. Arrows denote descending pathways.
FIGURE 13–2


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Movement is organized in increasingly complex and hierarchical levels.

**Reflexes** are controlled at the spinal or higher levels.

**Stereotypic repetitious movements**, such as walking or swimming, are governed by neural networks that include the spinal cord, brain stem, and cerebellum. Walking movements can be elicited in experimental animals after transection of the upper brain stem, probably as a result of the presence of **central pattern generators**, or local circuits of neurons that can trigger simple repetitive motor activities, in the lower brain stem or spinal cord.

**Specific, goal-directed movements** are initiated at the level of the cerebral cortex.

**MAJOR MOTOR SYSTEMS**
Corticospinal and Corticobulbar Tracts

A. Origin and Composition

The fibers of the corticospinal and corticobulbar tracts arise from the sensorimotor cortex around the central sulcus (see Fig 13–1); about 55% originate in the frontal lobe (areas 4 and 6), and about 35% arise from areas 3, 1, and 2 in the postcentral gyrus of the parietal lobe (see Fig 10–11). About 10% of the fibers originate in other frontal or parietal areas. The axons arising from the large pyramidal cells in layer V (Betz's cells) of area 4 contribute only about 5% of the fibers of the corticospinal tract and its pyramidal portion.

The portion of the pyramidal tract that arises from the frontal lobe is concerned with motor function; the portion from the parietal lobe deals more with modulation of the ascending systems. The tracts have endings or collaterals that synapse in the thalamus (ventral nuclei), the brain stem (pontine nuclei, reticular formation, and nuclei of cranial nerves), and the spinal cord (anterior horn motor neurons and interneurons; Fig 13–3). A direct pathway to spinal cord motor neurons exists only for the musculature of the distal extremity, such as the fingers that require rapid and precise control.

FIGURE 13–3
Diagram of the corticospinal tract, including descending fibers that provide sensory modulation to thalamus, dorsal column nuclei, and dorsal horn.
B. Pathways

The corticobulbar (corticonuclear) fibers originate in the region of the sensorimotor cortex, where the face is represented (see Figs 10–13 and 10–14). They pass through the posterior limb of the internal capsule and the...
middle portion of the crus cerebri to their targets, the somatic and brachial efferent nuclei in the brain stem. The **corticospinal tract** originates in the remainder of the sensorimotor cortex and other cortical areas. It follows a similar trajectory through the brain stem and then passes through the pyramids of the medulla (hence, the name pyramidal tract), decussates, and descends in the lateral column of the spinal cord (see Figs 5–13, 13–1, and 13–3). About 10% of the pyramidal tract does not cross in the pyramidal decussation but descends in the anterior column of the spinal cord; these fibers decussate at lower cord levels, close to their destination. In addition, up to 3% of the descending fibers in the lateral corticospinal tract are uncrossed. These ipsilateral descending projections control musculature of the trunk and proximal limbs and thus participate in the maintenance of an upright stance and in gross positioning of the limbs.

The pyramidal tract has a somatotopic organization throughout its course. (The origin, termination, and function of this tract have been described more fully in Chapter 5.)

**The Extrapyramidal Motor System**

The extrapyramidal system is a set of subcortical circuits and pathways, phylogenetically older than the corticospinal system, which includes the corpus striatum (caudate nucleus, putamen, and globus pallidus) together with the subthalamic nucleus, substantia nigra, red nucleus, and brain stem reticular formation (Figs 13–2A, 13–4, and 13–5). Some authorities include descending spinal cord tracts other than the corticospinal tracts (such as the vestibulospinal, rubrospinal, tectospinal, and reticulospinal tracts) in the extrapyramidal motor system. Cortical and subcortical components of the motor system are richly interconnected, either directly and reciprocally, or by way of fiber loops that involve the extrapyramidal system, and the majority traverse the basal ganglia.

**FIGURE 13–4**
Magnetic resonance image of a coronal section through the head at the level of the lentiform nucleus.
Magnetic resonance image of an axial section through the head at the level of the lentiform nucleus.

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Basal Ganglia

Pathways and nuclei: The anatomy of the gray masses in the forebrain that make up the basal ganglia has been described in Chapter 10 (Fig 13–2). The striatum (caudate and putamen) is the major site of input to the basal ganglia (see Fig 13–2B). The striatum receives afferents via the corticostriate projections from a large portion of the cerebral cortex, especially the sensorimotor cortex (areas 4, 1, 2, and 3), the more anterior premotor cortex (area 6), and the frontal eye fields (area 8) in the frontal and parietal lobes. These corticostriate projections are excitatory. The striatum also receives inputs from the intralaminar thalamic nuclei, substantia nigra, amygdala, hippocampus and midbrain raphe nuclei. Many inhibitory (gamma-aminobutyric acid [GABA]-ergic) and a smaller number of excitatory interneurons (the latter in some cases using acetylcholine as a transmitter) are present within the striatum.

The caudate and putamen send inhibitory (GABA-ergic) axons to the inner part of globus pallidus (GPi), which is the major outflow nucleus of the basal ganglia. These projections provide a strong inhibitory input to the globus pallidus (see Fig 13–2C).

The globus pallidus, GPi (internal part) is one of the two major output nuclei of the basal ganglia. GPi sends inhibitory axons (GABA-ergic) to the ventral nuclei (ventral anterior, VA; and ventral lateral, VL) of the thalamus.
thalamus (which also receives input from the cerebellum, the subthalamic nucleus, and substantia nigra). Axons from the globus pallidus project to the thalamus by passing through or around the internal capsule. They then travel in small bundles (the **ansa lenticularis** and the **lenticular fasciculus**, also known as the $H_2$ field of Forel) before entering the thalamus (see Fig 13–2C). The VA and VL thalamic nuclei complete the feedback circuit by sending axons back to the cerebral cortex (see Fig 13–2D). The circuit thus traverses, in order,

Cortex → striatum → globus pallidus (internal, GPi) → thalamus → cortex

Another important feedback loop involves the second major outflow nucleus of the basal ganglia, the **substantia nigra**, which is reciprocally connected with the putamen and caudate nucleus. Dopaminergic neurons in the **pars compacta** of the substantia nigra project to the striatum (the **nigrostriatal projection**), where they form inhibitory synapses on striatal neurons that have D2 dopamine receptors, and excitatory synapses on neurons that have D1 dopamine receptors (see Fig 13–2B). Reciprocal projections travel from the striatum to the substantia nigra (**striatonigral projection**) and are also inhibitory (see Fig 13–2C). This loop travels along the pathway

Cortex → striatum → substantia nigra → striatum

Neurons in the substantia nigra and G Pi also send inhibitory projections to the thalamus (VA and VL), which, in turn, sends projections to the sensorimotor cortex. Substantia nigra (pars compacta) also sends modulatory projections (**mesolimbic** and **mesocortical projections**) to the limbic system and cortex. This pathway involves the following circuit:

\[\text{Cortex} \rightarrow \text{striatum} \rightarrow \text{substantia nigra} \rightarrow \text{thalamus} \rightarrow \text{cortex}\]

The **pars reticulata** of the substantia nigra (SNr) receives input from the striatum, and sends axons outside the basal ganglia to modulate head and eye movements.

The **subthalamic nucleus** (also called the **nucleus of Luys**) also receives inhibitory inputs from the globus pallidus and from the cortex; efferents from the subthalamic nucleus return to the globus pallidus (see Fig 13–2C). Thus, the subthalamic nucleus participates in the feedback loop:

Cortex → globus pallidus → subthalamic nuclei → globus pallidus → cortex

Another loop involves the cerebellum. Portions of the thalamus project by way of the central tegmental tract to the inferior olivary nucleus; this nucleus, in turn, sends fibers to the contralateral cerebellar cortex. From the cerebellum, the loop to the thalamus is closed via the dentate and contralateral red nuclei.

Although there are no direct projections from the caudate nucleus, putamen, or globus pallidus to the spinal cord, the subthalamic region, including the prerubral field and the red nucleus, is an important relay and modifying station. Projections from the globus pallidus to the red nucleus converge with inputs from the motor cortex and the deep cerebellar nuclei. Efferent fibers from the red nucleus descend in the spinal cord as the rubrospinal tract, which modulates the tone of flexor muscles (see the following section).
The organizational theme for the basal ganglia involves complex loops of neurons (including many inhibitory neurons) feeding back to the sensorimotor cortex. These neuronal loops play an important role in motor control. Electrical engineers are well acquainted with abnormal oscillations, or "ringing," that can occur when inhibitory feedback circuits are damaged. Disorders of the basal ganglia are often characterized by abnormal movements that can be repetitive or rhythmic.

The motor control circuits passing through the basal ganglia that are involved in movement disorders such as Parkinson's disease have been conceptualized as operating in a manner summarized in Figure 13–6A. According to this model, excitatory synaptic output from the precentral and postcentral motor and sensory cortex is directed to the putamen. The putamen also receives projections from the pars compacta of the substantia nigra (SNc). Output from the putamen is directed toward the internal portion of the globus pallidus (GPi) and the pars reticulata of the substantia nigra (SNr) via two pathways (the direct and indirect pathways). Monosynaptic inhibitory projections from the putamen project via the direct pathway to GPi/SNr and tend to enhance motor activity. A series of polysynaptic connections extends from the putamen, within the indirect pathway, through portions of the external part of the globus pallidus (GPe) and the subthalamic nucleus (STN), with the net outcome of suppression of motor activity. In addition, mutual inhibitory connections exist between GPe and GPi/SNr. Outputs from GPi/SNr project toward the ventrolateral nuclear group of the thalamus (VL), and the VL in turn projects back to the cortex. Importantly, most of the intrinsic connections within the basal ganglia, and the GPi/SNr projections, are inhibitory (GABA-ergic), except for the projection between STN and GPi/SNr. Changes in activity in this circuitry as a result of cell death in SNc (Fig 13–6B), which disturb the balance between enhancement and suppression of motor activity, are discussed later and have significant implications for Parkinson's disease.

**FIGURE 13–6**

A: Conceptual model of activity in the basal ganglia and associated thalamocortical regions under normal circumstances. Dark arrows indicate inhibitory connections, and open arrows indicate excitatory connections. B: Changes in activity in Parkinson's disease. As a result of degeneration of the pars compacta of the substantia nigra, differential changes occur in the two striatopallidal projections (as indicated by altered thickness of the arrows), including increased output from GPi to the thalamus.

D, direct pathway; I, indirect pathway; GPe, external segment of globus pallidus; GPi, internal segment of globus pallidus; SNc, substantia nigra (pars compacta); SNr, substantia nigra (pars reticulata); STN, subthalamic nucleus; VL, ventrolateral thalamus. (Reproduced, with permission, from Wichmann T, Vitek JL, DeLong MR: Parkinson's disease and the basal ganglia: Lessons from the laboratory and from neurosurgery. *Neuroscientist* 1995;1:236–244.)
Subcortical Descending Systems

Additional pathways—important for certain types of movement—include the rubrospinal, vestibulospinal, tectospinal, and reticulospinal systems (see Fig 13–1 and Chapters 5 and 8).

A. Pathways

Subcortical descending systems originate in the red nucleus and tectum of the midbrain, in the reticular formation, and in the vestibular nuclei of the brain stem.

The rubrospinal tract arises in the red nucleus. The red nucleus receives input from the contralateral deep cerebellar nuclei (via the superior cerebellar peduncle) and the motor cortex bilaterally. Axons descend from the red nucleus in the crossed rubrospinal tract within the lateral column and then synapse on interneurons in the spinal cord.

The sensorimotor cortex projects to several nuclei in the reticular formation of the brain stem, which then sends fibers to the spinal cord in the form of the reticulospinal tract in the lateral column. Descending axons in this tract terminate on interneurons in the spinal cord and on gamma motor neurons.
The vestibulospinal tract arises in the vestibular nuclei, located in the floor of the fourth ventricle. The four vestibular nuclei receive afferents from the vestibular nerve and cerebellum. The vestibulospinal tract arises primarily from the lateral vestibular nucleus and medial vestibular nucleus. This tract contains both crossed and uncrossed fibers that project to anterior horn neurons in the spinal cord. (These mostly are interneurons that project to alpha and gamma motor neurons; extensor muscle motor neurons may be supplied directly.) Activity in the vestibulospinal tract resets the gain on the gamma loop so as to facilitate the activity of motor neurons that innervate muscles that oppose the force of gravity. Thus, the vestibulospinal tract plays an important role in maintaining an erect posture.

The tectospinal tract arises from cells in the superior colliculus and crosses in the midbrain at the level of the red nuclei. Descending tectospinal fibers travel within the medial longitudinal fasciculus in the medulla. Other tectospinal fibers descend in the anterior funiculus of the spinal cord and terminate at cervical levels, where they form synapses with interneurons that project to motor neurons. The tectospinal tract carries impulses that control reflex movements of the upper trunk, neck, and eyes in response to visual stimuli.

B. Function

The corticospinal and rubrospinal systems appear to cooperate to control hand and finger movement. The rubrospinal tract appears to play a role in control of flexor muscle tone.

The reticulospinal, vestibulospinal, and tectospinal systems play a limited role in movements of the extremities; their main influence is on the musculature of the trunk. Pure unilateral lesions of the corticospinal tract (ie, lesions that spare the other descending pathways) may result in relatively minor weakness, although precise movements of distal musculature (eg, movements of the individual fingers) are usually impaired. In these cases, descending control of motor neurons innervating proximal parts of the limbs and the trunk is mediated by the reticulospinal, vestibulospinal, and tectospinal pathways and by uncrossed axons in the anterior and lateral corticospinal tract.

Decerebrate rigidity occurs when the posterior part of the brain stem and spinal cord are isolated from the rest of the brain by injury at the superior border of the pons. In decerebrate rigidity, the extensor muscles in all of the limbs and those of the trunk and neck have increased tone. When the brain stem is transected, inhibitory influences from the cortex and basal ganglia can no longer reach the spinal cord, and facilitatory influences, which descend in the vestibulospinal and reticulospinal tracts, dominate. This results in increased activity of alpha motor neurons innervating extensor muscles, which is due to increased gamma motor neuron discharge for these muscles (see Fig 5–20).

Cerebellum

A. Pathways

The cerebellum is interconnected with several regions of the central nervous system (Fig 13–7; see also...
from the opposite cerebral cortex and cerebellar efferent systems to the contralateral red nucleus, the reticular formation, and the ventral nuclei of the contralateral thalamus (which connects to the cerebral cortex). These regions were discussed in Chapter 7.

**FIGURE 13–7**
Schematic illustration of some cerebellar afferents and outflow pathways.

**B. Function**

The cerebellum has two major functions: coordination of voluntary motor activity (fine, skilled movements and gross, propulsive movements, such as walking and swimming) and control of equilibrium and muscle tone. Experimental work suggests that the cerebellum is essential in motor learning (the acquisition or learning of stereotyped movements) and memory mechanisms (the retention of such learned movements).

**MOTOR DISTURBANCES**

Motor disturbances include weakness (paresis), paralysis, abnormal movements, and abnormal reflexes. They can result from lesions of the motor pathways in the nervous system or from lesions of the muscles.
Fasciculations are spontaneous, grossly visible contractions (twitches) of entire motor units (Table 13–1).

**TABLE 13–1**

**Signs of Lesions of the Human Motor System.**

<table>
<thead>
<tr>
<th>Location of Lesion</th>
<th>Voluntary Strength</th>
<th>Atrophy</th>
<th>Muscle Stretch Reflexes</th>
<th>Tone</th>
<th>Abnormal Movements</th>
</tr>
</thead>
<tbody>
<tr>
<td>Muscle (myopathy)</td>
<td>Weak (paretic)</td>
<td>Can be severe</td>
<td>Hypoactive</td>
<td>Hypotonic</td>
<td>None</td>
</tr>
<tr>
<td>Motor end-plate</td>
<td>Weak</td>
<td>Slight</td>
<td>Hypoactive</td>
<td>Hypotonic</td>
<td>None</td>
</tr>
<tr>
<td>Lower motor neuron (includes peripheral nerve, neuropathy)</td>
<td>Weak (paretic or paralyzed)</td>
<td>May be present</td>
<td>Hypoactive or absent</td>
<td>Hypotonic (flaccid)</td>
<td>Fasciculations*</td>
</tr>
<tr>
<td>Upper motor neuron</td>
<td>Weak or paralyzed</td>
<td>Mild (atrophy of disuse)</td>
<td>Hyperactive (spastic). After a massive upper-motor-neuron lesion (as in stroke), reflexes may be absent at first, with hypotonia and spinal shock</td>
<td>Hypertonic (claspknife) or spastic</td>
<td>Withdrawal spasms, abnormal reflexes (eg, Babinski’s extensor plantar response)</td>
</tr>
<tr>
<td>Cerebellar systems</td>
<td>Normal</td>
<td>None</td>
<td>Hypotonic (pendulous)</td>
<td>Hypotonic</td>
<td>Ataxia, dysmetria, dysdiadochokinesia, gait</td>
</tr>
<tr>
<td>Basal ganglia</td>
<td>Normal</td>
<td>None</td>
<td>Normal</td>
<td>Rigid (cogwheel)</td>
<td>Dyskinesias (eg, chorea, athetosis, dystonia, tremors, hemiballismus)</td>
</tr>
</tbody>
</table>

*Fasciculations are spontaneous, grossly visible contractions (twitches) of entire motor units.*
Muscles

The actions of each muscle are documented in Appendix B. A muscle may be unable to react normally to stimuli conveyed to it by the lower motor neuron, which results in weakness, paralysis, or tetanic contraction. Muscle tone may be decreased (hypotonia), and deep tendon reflexes may be reduced (hyporeflexia) or abolished (areflexia) as a result of muscle weakness. The cause of these disturbances may lie in the muscle itself or at the myoneural junction. Myasthenia gravis is a disorder of the myoneural junction, characterized by decreased efficacy of acetylcholine receptors, that results in weakness and fatigue. Myotonia congenita and the progressive muscular dystrophies are examples of muscle disorders characterized by muscle dysfunction in the presence of apparently normal neural tissue.

Lower Motor Neurons

Clinicians tend to differentiate between lower motor neurons and upper motor neurons and between lower- and upper-motor-neuron lesions. The clinical state of the patient often makes this differentiation straightforward, and this distinction can be useful in localizing a lesion.

A. Description

Lower motor neurons in the anterior gray column of the spinal cord or brain stem have axons that pass by way of the cranial or peripheral nerves to the motor end-plates of the muscles (see Fig 5–22). The lower motor neuron is called the "final common pathway" for two reasons. It is under the influence of the corticospinal, rubrospinal, olivospinal, vestibulospinal, reticulospinal, and tectospinal tracts as well as the segmental or intersegmental reflex neurons, and it is the ultimate pathway through which neural impulses reach the muscle.

B. Lesions

Lesions of the lower motor neurons can be located in the cells of the anterior gray column of the spinal cord or brain stem or in their axons, which constitute the ventral roots of the spinal or cranial nerves. Signs of lower-motor-neuron lesions include weakness, flaccid paralysis of the involved muscles, decreased muscle tone, muscle atrophy with fasciculations and degeneration of muscle fibers over time, and histologic-reaction degeneration (10–14 days after injury). Reflexes of the involved muscle are diminished or absent, and no abnormal reflexes are obtainable.

Lesions of lower motor neurons are seen in poliomyelitis (a viral disorder that results in death of motor neurons) and motor neuron disease (including forms called amyotrophic lateral sclerosis and spinal muscular atrophy, in which motor neurons degenerate). Mass lesions such as tumors involving the spinal cord can also damage lower motor neurons.

Upper Motor Neurons

A. Description
The upper motor neuron is a complex of descending systems conveying impulses from the motor areas of the cerebrum and subcortical brain stem to the anterior horn cells of the spinal cord. It is essential for the initiation of voluntary muscular activity. The term itself is used mainly to describe neurons with bodies rostral to those of lower motor neurons in the spinal cord or brain stem, and their descending axons (see Fig 5–22). One major component, the corticospinal tract, arises in the motor cortex, passes through the internal capsule and brain stem, and projects within the spinal cord to the lower motor neurons of the cord. Another component, the corticobulbar tract, projects to the brain stem nuclei of the cranial nerves that innervate striated muscles. Upper motor neurons control voluntary activation (but not necessarily reflex activation) of lower motor neurons.

B. Lesions

Lesions in the descending motor systems can be located in the cerebral cortex, internal capsule, cerebral peduncles, brain stem, or spinal cord (see Table 13–1). Signs of upper-motor-neuron lesions in the spinal cord include paralysis or paresis (weakness) of the involved muscles, increased muscle tone (hypertonia) and spasticity, hyperactive deep reflexes, no or little muscle atrophy (atrophy of disuse), diminished or absent superficial abdominal reflexes, and abnormal reflexes (eg, Babinski’s response).

Damage to the cerebral cortex incurred in utero, during birth, or in early postnatal life may result in cerebral palsy. This is a heterogeneous group of disorders that often include a form of spastic paralysis; however, the disease may be characterized by other signs, such as rigidity, tremor, ataxia, or athetosis. The disorder may be accompanied by defects such as speech disorders, apraxia, and cognitive impairment in some (but by no means all) patients.

C. Patterns of Paralysis and Weakness

Hemiplegia is a spastic or flaccid paralysis of one side of the body and extremities; it is delimited by the median line of the body. Monoplegia is paralysis of one extremity only, and diplegia is paralysis of any two corresponding extremities, usually both lower extremities (but can be both upper). Paraplegia is a symmetric paralysis of both lower extremities. Quadriplegia, or tetraplegia, is paralysis of all four extremities. Hemiplegia alternans (crossed paralysis) is paralysis of one or more cranial nerves and contralateral paralysis of the arm and leg. The term paresis refers to weakness, rather than total paralysis, and is used with the same prefixes.

Basal Ganglia

Defects in function of the basal ganglia (sometimes termed extrapyramidal lesions) are characterized by changes in muscle tone, poverty of voluntary movement (akinesia) or abnormally slow movements (bradykinesia), or involuntary, abnormal movement (dyskinesia). A variety of abnormal movements can occur: tremors (resting tremor at rest and postural tremor when the body is held in a particular posture), athetosis (characterized by slow, writhing movements of the extremities and neck musculature), and chorea.
(quick, repeated, involuntary movements of the distal extremity muscles, face, and tongue, often associated with lesions of the corpus striatum).

A discussion of some particularly notable diseases of the basal ganglia follows.

**A. Huntington's Disease**

This autosomal-dominant disorder is characterized by debilitating abnormal movements (most often chorea; rigidity in early-onset cases) and cognitive and psychiatric dysfunction. Depression is common. The disorder progresses relentlessly to incapacitation and death. Onset usually occurs between the ages of 35 and 45 years, although a childhood form is sometimes present.

Huntington's disease is due to mutation of a gene located on chromosome 4. The function of the protein encoded by this gene (Huntingtin) is not known. In most cases, the mutation includes a trinucleotide (CAG) repeat, that is, an expanded region of the gene in which the sequence CAG is abnormally repeated.

The pathology of Huntington's disease includes striking loss of neurons in the caudate and putamen, which can be observed both microscopically and macroscopically (loss of bulk of the caudate nucleus where it indents the lateral wall of the lateral ventricle). Loss of GABA-ergic (inhibitory) neurons in the striatum results in chorea (Fig 13–8). The cerebral cortex also becomes atrophic. The steps leading from expression of Huntington's gene to the degeneration of inhibitory neurons in the striatum and clinical expression are not understood.

**FIGURE 13–8**
Schematic illustration of the processes underlying Parkinsonism. GABA, gamma-aminobutyric acid.
B. Hemiballismus

In this unusual movement disorder, one extremity or the arm and leg on one side engage in large, flailing movement. Hemiballismus usually results from damage to the contralateral subthalamic nucleus, commonly as a result of infarction. For reasons that are poorly understood, hemiballismus often resolves spontaneously after several weeks.

C. Parkinson's Disease

This disorder, with onset usually between the ages of 50 and 65 years, is characterized by a triad of symptoms: tremor, rigidity, and akinesia. There are often accompanying abnormalities of equilibrium, posture, and autonomic function. Characteristic signs include slow, monotonous speech; diminutive writing (micrographia); and loss of facial expression (masked face).

This progressive disorder is associated with loss of pigmented (dopaminergic) neurons in the substantia nigra (Figs 13–8 and 13–9). The cause of this degenerative disorder is unknown. Parkinsonian symptoms were seen in some survivors of the epidemic of encephalitis lethargica (von Economo's encephalitis) that occurred from 1919 to 1929 (postencephalitic parkinsonism). Some toxic agents (carbon monoxide, manganese) can damage the basal ganglia, and a rapidly developing Parkinson-like disease has been linked to the use of certain "designer drugs," for example, MPTP (1-methyl-4-phenyl-1,2,5,6-tetrahydropyridine), a synthetic narcotic related to meperidine. Moreover, use of some neuroleptics (eg, phenothiazines) can produce a drug-
induced parkinsonian syndrome. Most causes of Parkinson's disease, however, are idiopathic, and mechanisms leading to degeneration of neurons in the substantia nigra are not well understood.

**FIGURE 13–9**
Midbrain of a 45-year-old woman with Parkinson's disease, showing depigmentation of the substantia nigra.

![Image of midbrain showing depigmentation of substantia nigra](image-url)


**Cerebellum**

Disorders caused by cerebellar lesions are characterized by reduced muscle tone and a loss of coordination of smooth movements (see Table 13–1). Lesions in each of the three subdivisions of the cerebellum exhibit characteristic signs.

**CASE 17**

A 63-year-old, right-handed secretary/typist consulted her family physician when her right hand and fingers "did not want to cooperate." She also explained that her employers had become dissatisfied with her because her work habits and movements had become slow and her handwriting had become illegible over the preceding months. Her intellectual abilities were unimpaired.

Neurologic examination showed slowness of speech and mild loss of facial expression on both sides. The patient had difficulty initiating movements. Once seated, she did not move about much. Her posture was stooped and she walked with a small-stepped gait, with decreased arm swing. There was no muscular atrophy and no weakness. Muscle tone was increased in the arms, and "cogwheel rigidity" was present. There was a fine tremor in the fingers of the right hand (frequency of three to four times per second). The rest data were within normal limits.
What is the most likely diagnosis? Where is the lesion?

CASE 18

A 49-year-old woman with known severe hypertension complained of a severe headache. She then suddenly lost strength in the left leg and arm; she fell down and, when brought to the emergency room, seemed only partially conscious.

Neurologic examination showed an obtunded woman who had difficulty in speaking. There was no sensation on the left side of the face or body. Left-central facial weakness was present. When aroused, the patient complained that she could not see on the left side of both visual fields. Complete paralysis of the left upper and lower extremities was present. Deep tendon reflexes were absent in the left upper extremity and increased in the lower extremity. There was a left extensor plantar response. Vital signs and the complete blood count were within normal limits; blood pressure was 190/100.

What is the preliminary diagnosis? Would a lumbar puncture be indicated? Would imaging be useful?

Cases are discussed further in Chapter 25.

A. Vestibulocerebellum (Archicerebellum)

Loss of equilibrium, often with nystagmus, is typical.

B. Spinocerebellum (Paleocerebellum)

Truncal ataxia and "drunken" gait are characteristic.

C. Neocerebellum

Ataxia of extremities and asynergy (loss of coordination) are prominent. Decomposition of movement occurs; voluntary muscular movements become a series of jerky, discrete motions rather than one smooth motion. Dysmetria (past-pointing phenomenon) is also seen, in which people are unable to estimate the distance involved in muscular acts, so that their attempts to touch an object will overshoot the target. Dyssdiadochokinesia (the inability to perform rapidly alternating movements), intention tremor, and rebound phenomenon (loss of interaction between agonist and antagonist smooth muscles) are also typical. If there is a unilateral lesion of the cerebellum, these abnormalities present on the same side as the lesion.

BOX 13–1 Essentials for the Clinical Neuroanatomist

After reading and digesting this chapter, you should know and understand:

The corticospinal tract and its decussation (Fig 13–3)
Dopaminergic dysfunction in Parkinson's disease

Subcortical descending systems

Motor disorders: sites of pathology and clinical presentation (Table 13–1)

REFERENCES


