THE CEREBELLUM
anatomy, functioning and disorders

PREFACE

The cerebellum is a derivative of the hindbrain which is developed in connection with the receptors of statics. **The cerebellum,** the second-largest part of the brain, occupies the inferior and posterior aspects of the cranial cavity. It accounts for about a tenth of the brain mass yet contains nearly half of the neurons in the brain. The cerebellum is situated under the frontal lobes of the cerebral hemispheres, dorsal to the pons and medulla oblongata, and is lodged in the posterior cranial fossa. The large lateral parts, the hemispheres (hemispheria cerebelli) and a narrow, middle part lying between them, the vermis, are distinguished in it.

FUNCTION

The main function of the cerebellum is to evaluate how well movements initiated by motor areas in the cerebrum are actually being carried out. When movements initiated by the cerebral motor areas are not being carried out correctly, the cerebellum detects the discrepancies. It then sends feedback signals to motor areas of the cerebral cortex, via its connections to the red nucleus and thalamus. The feedback signals help correct errors, smooth movements, and coordinate complex sequences of skeletal muscle contractions. Besides coordinating skilled movements, the cerebellum is the main brain region that regulates posture and balance.

It is also considered one of the highest centers of the vegetative (sympathetic) nervous system.

The presence of reciprocal connections between the cerebellum and association areas of the cerebral cortex suggest that the cerebellum may also have nonmotor functions such as cognition (acquisition of knowledge) and language processing.

ANATOMY OF THE CEREBELLUM

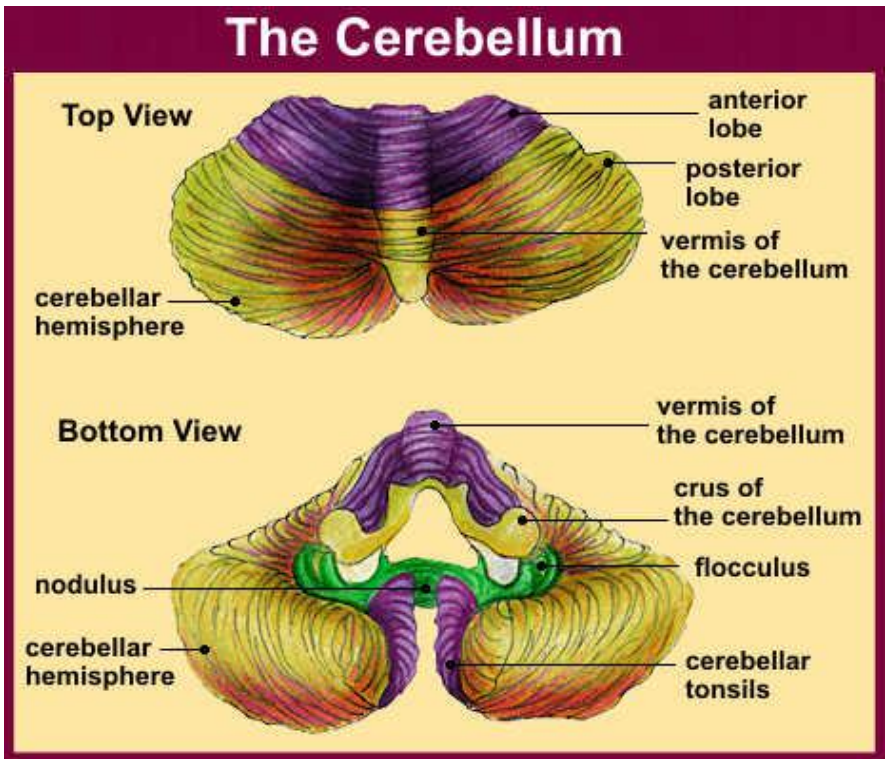
In superior or inferior views, the shape of the cerebellum is somewhat like a butterfly. The central constricted area is the vermis (“worm”), and the lateral "wings" or lobes are the cerebellar hemispheres. Each hemisphere consists of 3 lobes separated by deep and distinct fissures. The anterior lobe and posterior lobe govern subconscious aspects of the skeletal muscle movements. The flocculonodular lobe on the inferior surface contributes to equilibrium and balance.

The surface of the cerebellum is covered with a layer of grey matter constituting its cortex (three layers) and forms narrow cerebellar folia of the cerebellum (folia cerebelli) separated one from another by fissures (fissurae cerebelli). The deepest, horizontal fissure (fissura horizontalis cerebelli) passes to the posterior border of the cerebellum and separates the upper surface of hemispheres (facies superior) from the lower surface (farcies inferior). The horizontal and other large fissures divide the entire surface of the cerebellum into lobules (lobuli cerebelli). Among these it is necessary to point out the most isolated, small lobule, the flocculus, lying on the inferior surface of each hemisphere near the middle cerebellar peduncle, and the nodulus, a part of the vermis connected with the flocculus. The flocculus is joined to the nodulus by means of a fine strand, the peduncle of the flocculus (pedunculus flocculi), which is continuous medially with a thin crenate lamina, the inferior medullary velum (velum medullare inferius)/

Internal Structure of the Cerebellum

Paired nuclei of grey matter are embedded in the white matter of both halves of the cerebellum (Fig. 2). On either side of the midline, where the fastigium projects into the cerebellum, is the extreme medial nucleus fastigii.

Lateral to it lie small islets of the nucleus globosus, and still further lateral, the emboliformis nucleus. Finally in the centre of the hemisphere is the dentate nucleus (nucleus dentatus) which has the appearance of a grey curved lamina resembling the olivary nucleus and has a mouth open medially (the hilus of the nucleus dentatus). The resemblance of the cerebellar dentate nucleus to the crenate nucleus of the olive is not a chance phenomenon; both nuclei are connected with the conduction tracts, the fibrae olivocerebellares, and each folium of one nucleus is similar to the folium of the other nucleus. Thus, both nuclei contribute together to the accomplishment of balance.



Cerebellar peduncles.

1. The *inferior cerebellar peduncles* (pedunculi cerebellares inferiors, running to the medulla oblongata) convey to the cerebellum the tractus *spinocerebellaris posterior* (Flechsig's), the fibrae arcuatae externae from the nuclei of the posterior funiculi of the medulla oblongata, and the fibrae *olivocerebellares* from the olive. All these fibres end in the cortex of the vermis and hemispheres.

The posterior spinocerebellar (Flechsig's) tract. The cell body of the first neuron is in the spinal ganglion, the axon separates into a peripheral branch running in the muscular nerve to the receptor located in some part of the motor apparatus, and a central branch, which as a component of the posterior root, penetrates the posterior columns of the spinal cord and surrounds the thoracic nucleus of the posterior horns with its terminal rami and collaterals. The thoracic nucleus contains the cells of the second neuron which axons form the posterior spinocerebellar tract. As it is shown by its name, the thoracic nucleus is pronounced best in the thoracic segment between the level of the last cervical and the second lumbar vertebra. The posterior cerebellar tract, on reaching as a component of the lateral funiculus, the medulla oblongata on its own side passes to the inferior cerebellar peduncles to the cortex of the vermis. On its way to the spiral cord and medulla oblongata it does not cross over to the other side and is therefore called the direct cerebellar tract. On entering the cerebellum, however, most of its fibres cross in the vermis.

In addition, fibres pass into these peduncles from the nuclei of the vestibular nerve and end in the nucleus fastigii. These fibres convey to the cerebellum impulses from the vestibular apparatus and the proprioceptive field; as a result it becomes a nucleus of proprioceptive sensation concerned with automatic correction of the motor activity of the other parts of the brain. Descending tracts also pass in the opposite direction in the inferior peduncles from the nucleus fastigii to the lateral vestibular nucleus and from this nucleus to the anterior horns of the spinal cord (*the vestibulo-spinal tract)*. The cerebellum produces the affects the spinal cord functioning via this tract.

The cerebellum takes part in controlling the spinal cord motor neurons. This is accomplished through the cerebellorubrospinal tract (tractus cerebellorubrospinalis). The cell body of the first link of this tract lies in the cerebellar cortex (Purkinje cells). The axons of the cells terminate in the cerebellar dentate nucleus and, possibly, in other nuclei of the cerebellum. The second link arises here. The axons of the second neurons pass through the superior cerebellar peduncles to the midbrain and terminate in the red nucleus, in which the cells of the third link are located. As components of the rubrospinal

(Monakow's) tract (*tractus rubrospinalis*) the axons of these cells, after changing over in the anterior horns of the spinal cord (the fourth link), reach the skeletal muscles.

2. The *middle cerebellar peduncles* (pedunculi cerebellares medii) (running to the pons) contain fibres passing from the pontine nuclei to the cerebellar cortex. The *pontocerebellar tracts*, the conducting pathways to the cerebellar cortex, arise in the pontine nuclei and are a continuation of the corticopontine fibres which terminate in the nuclei of the pons after decussation. These pathways link the cerebral cortex with the cerebellar cortex, that explains the fact that the more developed is the cortex of the brain and in men the pons and the cerebellar hemispheres are more developed. The cerebral cortex, which is in charge of all body processes, also governs the cerebellum, the most important proprioceptive centre concerned with body movement. This is achieved due to the presence of a special tract descending from the cortex of the brain to that of the cerebellum, the corticopontocerebellar tract (*tractus corticopontocerebellaris*).

The first link of this tract consists of neurons whose cell bodies are located in the cerebral cortex while the axons descend to the nuclei of the pons, nuclei (proprii) pontis. These neurons form separate bundles termed, according to the different lobes of the brain, the frontopontine (*tractus frontopontinus*), occipitopontine (*tractus occipitopontinus*), and parietopontine (*tractus arietopontinus*) tracts. Thepontine nuclei give rise to the second neuron whose axons form the pontocerebellar tract (*tractus pontocerebellaris*) passing to the opposite side of the pons and reaching the cerebellar cortex (neocerebellum) as components of the middle cerebellar peduncles (Fig. 2). A connection is thus established between the cortex of the brain and the cerebellar hemispheres. (Each cerebral hemisphere is connected with the contralateral cerebellar hemisphere).

3. The *superior peduncles* (pedunculi cerebellares superiores) (running to the tectal lamina) consist of nerve fibres stretching in both directions: (1) to the cerebellum, the anterior spinocerebellar, or Gowers' tract (*tractus spinocerebellaris anterior*), and (2) from the cerebellar dentate nucleus to the tectum, the cerebellotegmental tract (*tractus cerebellotegmentalis*) which after decussation terminates in the red nucleus and thalamus. The cerebellum receives impulses from the spinal cord through the first tract and sends impulses to the extrapyramidal system along the second tract and thus it itself has an influence on the spinal cord through this system.

The anterior (Gowers') spinocerebellar tract (tractus spinocerebellaris anterior [Gowers']) has the first neuron in common with the posterior tract. The cells of the second neurons are in the posterior horn. Their axons form the anterior spinocerebellar tract and stretch in the anterior parts of the lateral white column on their side and on the opposite side to which they cross through the white commissure. The tract ascends through the medulla oblongata and the pons - to the superior medullary velum where it again crosses to the other side. Then the fibres enter the cerebellum through its superior peduncles and terminate in the cortex of the vermis. Thus, the whole tract forms two decussations owing to which proprioceptive sensibility is conveyed to the same side from which it had gone.

Therefore, the three pairs of cerebellar peduncles are responsible for its versatile connections. The cerebellum receives impulses from the spinal cord and medulla oblongata through the inferior peduncles, from the cortex of the cerebral hemispheres through the middle peduncles; the superior peduncles contain the main efferent tract along which the cerebellar impulses are conveyed to the cells of the anterior horns of the spinal cord. The connection of the cerebral hemispheres with the cerebellar hemispheres, i.e. with its new part (neocerebellum) is crossed, while the vermis, i.e. the old part of the cerebellum (palaeocerebellum), is connected with the spinal cord in a straight manner, i.e. homolaterally.

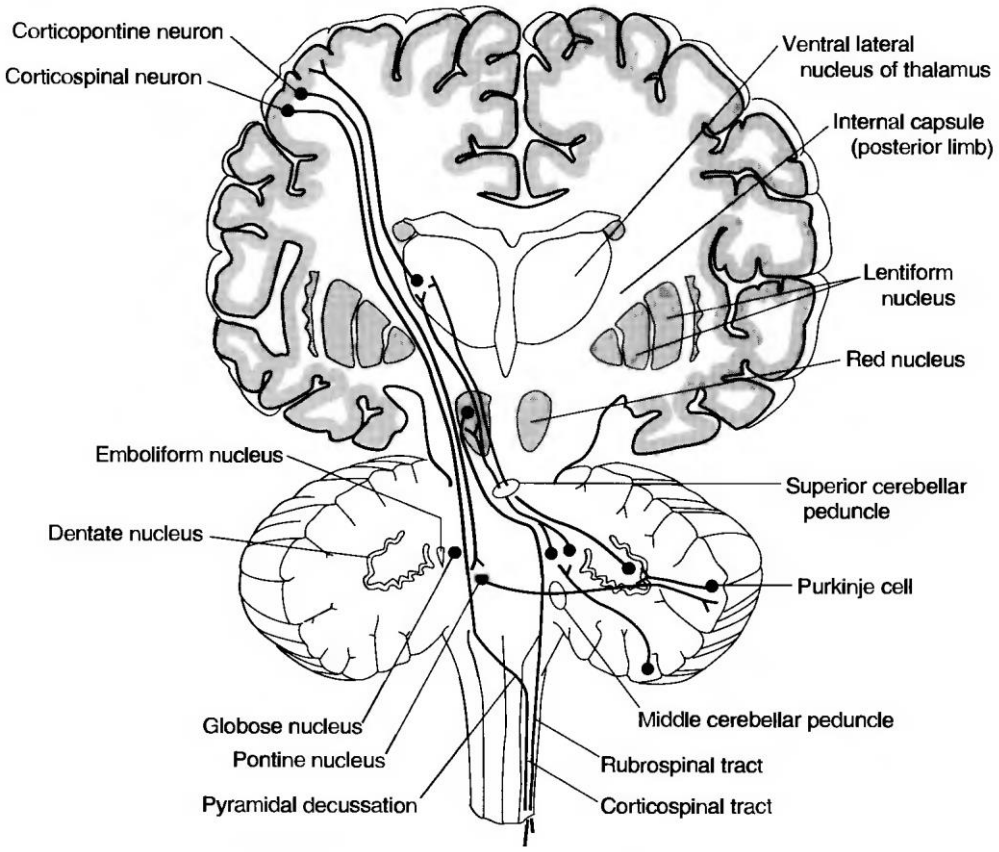


Figure 2. Principal cerebella connections.

CLINICAL EXAMINATION OF PATIENTS WITH CEREBELLAR DISFUNCTION.

There are several simple tests to demonstrate ataxia or incoordination.

Coordination. The patient is asked to touch the tip of his nose with the tip of his index finger slowly with his eyes open (*the finger nose test*). This is repeated with the eyes shut to differentiate loss of deep sensibility from **dynamic ataxia**. Incoordination, not of sensory origin (cerebellar), is present with both the eyes open and closed. Weakness may interfere with the testing procedure. With eyes closed the index fingers touch tip to tip in front of the thorax. To test the lower extremities the patient is asked to place the heel of one foot on the opposite knee and slide it down the shin bone (*the heel knee test*). The given test is performed with the eyes open and closed, the same type of incoordination is done the finger nose test. *Other tests for ataxia* consist in having the patient pour water from one test tube or glass to another. Ordinary movements, such as sitting up or buttoning clothes may furnish evidence of incoordination.

The Romberg test recognizes **static ataxia,** it if performed by having the patient stand with heels and toes together with open eyes and then with closed eyes. The tendency unstable position with the eyes close provides a positive Romberg probe.

Other tests: the patient is asked to touch his index finger to the examiner's finger which gradually recedes from the patient's finger. The same test is carried out with the feet, the patient attempts to touch examiner's finger with his big toe. A steadiness in movement without tremor is normal. The cerebellar patient has a tendency to move his finger in short jerky **decomposed movements** and not a smoothly coordinated total sweep **dysmetria**, or overshooting of the mark, is an accompaniment of cerebellar lesions. To compensate for this the patient quickens his speed and shorts out his finger at the target to get there before the unsteadiness appears. An intentional **tremor** which is brought on by action and ceases at rest is also characteristic.

Decomposition of movements is also tested by watching the patient arise from his bed or from a chair. Instead of one highly integrated movement with each individual muscle group functioning almost simultaneously, each movement is performed awkwardly in turn.

The rebound phenomenon of Holmes is elicited by having the patient flex his forearm forcibly against the examiner's resistance which is suddenly released. Normally, the tendency of the arm to fly into flexion is checked by a rapid innervation of the antagonistic extensors. But pathology of the cerebellar disturbance so alters the muscle tonus, that the antagonistic muscle cannot be innervated quickly enough and violent rebound develops. In testing the upper extremity, attention should be given to protect the patient's face.

The normal individual is able to perform successive movements rapidly, as alternate pronation and supination of the forearm, or extension and flexion of the fingers. In cerebellar lesions the patient's successive movements are increasingly clumsy and irregular in time, which is termed **adiodokokinesia.**

This is due to faulty reciprocal innervation of antagonistic and protagonistic muscles and also to a faulty synergism of the muscles which fix the neighboring joint, for example, fixation of the wrist in opening and shutting the fingers ***(in pronation and suppination testing).*** Wertham has brought out a rhythmic finger tapping test, for cerebellar incoordination. In cerebellar disease a good rhythm cannot be maintained.

Cerebellar signs. As noted under posture and gait, the position of the head is often important for the localization of tumors about the tentorium. In observing the gait, the examiner detects signs of unsteadiness and tendencies to fall or stagger backward or to either side. If there is doubt as to the side to which side the patient is not stable, the chair test is done. This involves having the patient circle around a chair first clockwise and then counterclockwise ***(in grasping / ungrasping)***. In circling the chair in one direction he closes, in the other he sways from it when there is a tendency to lateral pulsion.

The hypotonicity or atonicity can be tested by forcible swinging and flopping of the upper extremities. Weakness and slowness in muscular movements, as well as pendular patellar reflexes, may be associated with cerebellar damage. Also very important signs of cerebellar disorders include **nystagmus, scanning speech and dysarthria** should be carefully described, as these are often present in cerebellar diseases.

Nystagmus or spontaneous oscillatory movements are seldom seen in the position of fixation. Nystagmus usually appears when the eyes conjugately deviated to either lateral field, upward or downward. There are two components, quick and slow. The nystagmus is designated after the quick component, which is usually away from the center of fixation, i.e., outward (the cerebellar damage has the fast component maximal toward the side of the cerebellar lesion). Nystagmus is described as follows: direction of quick component, direction of gaze which produces it, speed, amplitude, and rotatory or linear direction of movement. Normally, fatigue produced a few nystagmoid mevements in extreme lateral deviation, rarely on vertical movement.

Scanning speech and dysarthria. Speech is usually tested while the history is taken and may early give a clue to patient.s disease. In cerebellar disturbances speech is slow, monotonous and interrupted by breath-taking pauses not at punctuation points. Speech may be slurred with marked tremor of lips, tongue and face made worse by talking.

LOCALIZATION OF CEREBELLAR DISORDERS

Disorders of the cerebellum and its inflow or outflow pathways produce deficits in the rate, range, and force of movement. Anatomically, the cerebellum has three subdivisions. The archicerebellum (vestibulocerebellum) comprises the flocculonodular lobe, helps maintain equilibrium and coordinate eyehead-neck movements, and is closely interconnected with the vestibular nuclei. The midline vermis (paleocerebellum) helps coordinate movement of the trunk and legs. Vermis lesions result in abnormalities of stance and gait. The lateral hemispheres, which make up the neocerebellum, control ballistic and finely coordinated limb movements, predominantly of the arms. Signs of cerebellar disease are listed in table.

Sign	Description
Ataxia	Reeling, wide-based gait
Dysdiadochokinesia (adiadochokinesis)	Inability to perform rapid alternating movements
Decomposition of movement	Inability to properly sequence fine, coordinated acts
Dysarthria	Inability to articulate words properly, with slurring and inappropriate phrasing
Dysmetria	Inability to control range of movement
Hypotonia	Decreased muscle tone
Nystagmus	Involuntary rapid oscillation of the eyeballs in a horizontal, vertical, or rotary direction with the fast component maximal toward the side of the cerebellar lesion
Scanning speech	Slow enunciation, a tendency to hesitate at the beginning of a word or syllabl
Tremor	Rhythmic, alternating, oscillatory movement of a limb as it approaches a target (intention tremor) or of proximal musculature when fixed posture or weight bearing is attempted (sustention tremor)

STRUCTURAL LESIONS OF THE CEREBELLUM

Infarcts, hemorrhages, or tumors can produce neurological deficits and, as they enlarge, may cause hydrocephalus or increased intracranial pressure with papilledema. The midline cerebellum is the most common site of primary brain tumors (medulloblastoma, cystic astrocytoma) in childhood. Cerebellar deficits may be caused by demyelinating plaques of multiple sclerosis (which may occur anywhere in the cerebellar white matter), the Chiari malformation (extension of cerebellar tissue into the cervical canal), and basilar invagination with platybasia (flattening of the skull base).

Alcoholism with nutritional deprivation can cause degeneration of the vermis and anterior cerebellum with profound gait ataxia. Acquired cerebellar syndromes may also be caused by hypothyroidism, various toxins (carbon monoxide, heavy metals, phenytoin), hyperpyrexia, and repeated head trauma. Rarely, reversible pancerebellar dysfunction may follow viral infections in children. A rare, profound cerebellar degeneration may accompany certain malignancies in adults.

Same hereditary disease may accompanied by cerebellar symptoms: Friedreich.s ataxia, abetalipoproteinemia, Refsum.s disease, multiple system atrophy, ataxia-teleangiectasia, mitochondrial multisystem disorders, etc.

EXTRAPYRAMIDAL SYSTEM

The *extrapyramidal system* consists of the following gray structures: caudate nucleus, putamen, pallidum, subthalamic nucleus, substantia nigra, and red nucleus.

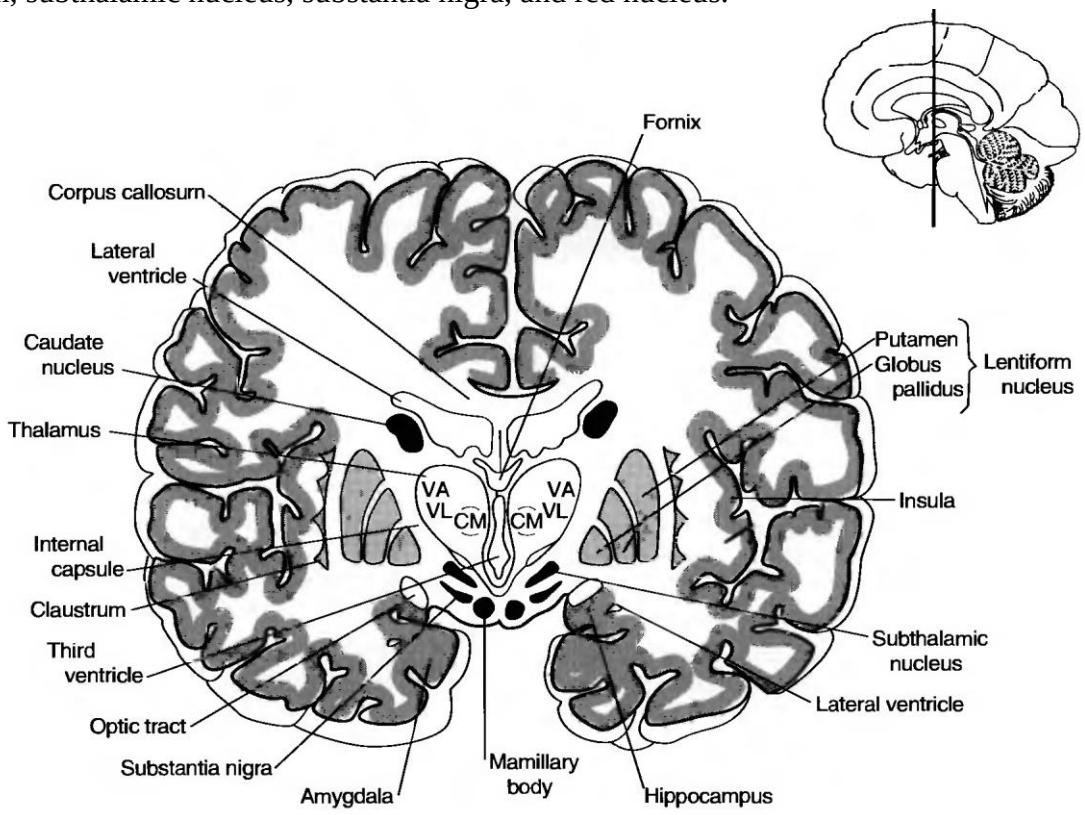


Figure 21-1. A coronal section through the midthalamus at the level of the mamillary bodies. The basal ganglia are all prominent at this level and include the striatum and the lentiform nucleus. The subthalamic nucleus and substantia nigra are important components of the striatal motor system.

THE BASAL GANGLIA OF THE HEMISPHERES

In addition to the grey cortex on the surface of the hemisphere, masses of grey matter are present in the depth of its tissue. These are called **basal, central, or subcortical nuclei,** in brief the "subcortex". As distinct from the cortex which has the structure of screen centres, the subcortical nuclei possess the structure of nuclear centres. Three conglomerates of subcortical nuclei are distinguished: **corpus striatum, claustrum, and the amygdaloid nucleus** (Fig. 1).

1. Corpus striatum consists of two parts, the caudate and lentiform nuclei, which are incompletely separated one from the other.

A. The caudate nucleus (nucleus caudatus) lies above and medial to the lentiform nucleus and is separated from it by a layer of white matter called the internal capsule (capsula interna). The thickened anterior part of the nucleus, its *head (caput nuclei caudati)* forms the lateral wall of the anterior horn of the lateral ventricle, while its attenuated parts, the *body* and *tail (corpus and cauda nuclei caudati),* stretch to the back on the floor of the central part of the lateral ventricle; the tail curves onto the superior wall of the inferior horn. Medially the caudate nucleus adjoins the thalamus from which it is separated by a band of white matter called the stria semicircularis (stria terminalis).

Anteriorly and inferiorly the head of the nucleus approaches the anterior perforated substance where it is united with the lentiform nucleus (with the part called the putamen). In addition to this wide union of these two nuclei on the ventral side, there are fine bands of grey matter joining them dorsally. These bands, alternating with the white bundles of the internal capsule, are responsible for the name corpus striatum.

B. The lentiform nucleus (nucleus lentiformis) lies lateral to the caudate nucleus and the thalamus and is separated from them by the internal capsule. On a horizontal section through the hemisphere the medial surface of the lentiform nucleus facing the internal capsule has the shape of an angle whose apex is directed centrally, the anterior side is parallel to the caudate nucleus, and the posterior side is

parallel to the thalamus. The lateral surface of the nucleus is slightly convex and is directed at the lateral side of the hemisphere in the region of the insula. As it is pointed out above, anteriorly and ventrally the lentiform nucleus is fused with the head of the caudate nucleus. On a frontal section the lentiform nucleus is wedge-shaped, the apex of the wedge is directed medially, and the base laterally. Two parallel white layers called the medullary laminae (laminae medullares) divide the lentiform nucleus into three segments, one lateral grey segment called the *putamen* and two medial lighter coloured segments united under the term *globus pallidus*.

The globus pallidus has a distinctive macroscopic appearance and also differs from the other parts of the corpus striatum histologically. It is phylogenetically older (palaeostriatum) than the putamen of caudate nucleus (neostriatum). In view of all these features, the globus pallidus is now distinguished as a specific morphological structure, *the pallidum*, while the term *striatum* is used as a designation only for the putamen and the caudate nucleus. As a consequence, the term "lentiform nucleus" loses its initial meaning and should be used only in a purely topographical meaning; the caudate and lentilorni nuclei are now called the striapallidal system instead of corpus striatum. This system is the principal part of the extrapyramidal system and, besides, it is the higher centre of control of vegetative functions concerned with thermoregulation and carbohydrate metabolism and predominates over similar vegetative centres in the hypothalamus.

2. The claustrum is a thin sheet of grey matter in the region of the insula between it and the putamen. It is separated from the putamen by a thin layer of white matter, the external capsule (capsula externa) and from the cortex of the insula by a similar layer called the capsula extrema.

3. The amygdaloid nucleus *(corpus amygdaloideum)*, or the epistriatum lies under the putamen in the anterior end of the temporal lobe. It does not reach the temporal pole, but lies in front of the apex of the inferior horn of the lateral ventricle. Morphologically, the amygdaloid nucleus is a posteroventral continuation of the claustrum. It is evidently a subcortical olfactory centre where a bundle of fibres passing from the olfactory lobe and the anterior perforated substance terminates.

The afferent nigrostriatal fibers have been described to be dopaminergic and to reduce the inhibitory unction of the strialum, On the other hand, the strionigral tract is *gamma-aminobulyric acid- or GABA-ergic* and has an inhibitory effect on the dopaminergic nigrostriatal neurons. This is a closed-loop feedback circuit. The GABA-ergic neurons of the strionigral fibers probably inhibit the descending, most likely dopaminergic, nigral neurons that control muscle tone via gamma neurons (Hassler).

SIGNS CAUSED BY LESIONS IN EXTRAPYRAMIDAL GRISEA

The main signs of extrapyramidal lesions are disorders of muscle tone *(dystonia)* and involuntary movement disorders *(hyperkinesia, hypokinesia, akinesia)* absent during sleep. Two clinical syndromes can be differentiated. One is characterized by a combination or hyperkinesia and hypotonia and is caused by a disease of the neostriatum. The other presents as a combination of hypokinesia and hypertonia or rigidity and stems from a disease of the substantia nigra.

Symptoms And Signs of The Hypokinesia-Hypertonia Syndrome (Parkinsonism)

In 50 to 80% of patients, the disease begins with a resting 4- to 8-Hz *pill-rolling tremor* of one hand. The tremor is maximal at rest, diminishes during movement, and is absent during sleep; it is enhanced by emotional tension or fatigue. Usually, the hands, arms, and legs are most affected, in that order. Jaw, tongue, forehead, and eyelids may also be affected, but the voice escapes the tremor.

Rigidity progresses, and *movement becomes slow* (bradykinesia), decreased (*hypokinesia*), and difficult to initiate (*akinesia*). Rigidity and hypokinesia may contribute to muscular aches and sensations of fatigue. In contrast to spastic elevation of muscle tone, rigor can be felt in extensors as a sticky, waxy resistance to all passive movements. The muscles cannot be relaxed. In passive movements one can feel that the tone of the antagonist muscles decreases in steps and not in an even, continuous fashion *(cog-wheel phenomenon).* The lifted head of a lying person, when suddenly released, does not fall down as usual but sinks gradually back onto the pillow *(head-dropping test).*

In contrast to their behavior in a spastic condition, the proprioceptive reflexes are not increased, and no pathologic reflexes can be observed. Paresis is absent. If it too difficult to elicit reflexes, it is

not possible to intensify the patellar reflex by *Jendrassik's maneuver*. (The patient hooks his hands together by the flexed fingers and tries to pull them apart as hard as he can while the patellar reflexes are checked). The result is an increase in the tonic stretching reflex, that is, an *activated rigidity.*

The face becomes masklike, with mouth open and diminished blinking, which may be confused with depression. *The posture becomes stooped*. *Patients find it difficult to start walking; the gait becomes shuffling with short steps, and the arms are held flexed to the waist and do not swing with the stride.* Steps may inadvertently quicken, and the patient may break into a run to keep from falling (*festination*). The tendency to fall forward (*propulsion*) or backward (*retropulsion*) when the center of gravity is displaced results from loss of postural reflexes.

Speech becomes hypophonic, with a characteristic *monotonous*, stuttering *dysarthria.* Hypokinesia and impaired control of distal musculature results in *micrographia* and increasing difficulty with activities of daily living. *Dementia* affects about 50% of patients, and depression is common (Fig. 2).

During ***examination***, passive movement of the limbs is met with a *plastic, unvarying lead-pipe rigidity;* superimposed tremor bursts may have a ratchet-like cogwheel quality. *The sensory examination is usually normal. Signs of autonomic nervous system dysfunction* (eg, seborrhea, constipation, urinary hesitancy, orthostatic hypotension) may be found. *Muscle strength is usually normal,* although useful power may be diminished and the ability to perform rapid successive movements is impaired. *Reflexes remain normal* but may be difficult to elicit in the presence of marked tremor or rigidity.

THE HYPERKINESIA-HYPOTONIA SYNDROME

Aetiology And Pathophysiology of The Hyperkinesia-Hypotonia Syndrome.

This syndrome develops if the neostriatum is damaged. Occasionally, such lesions are accompanied by others in the globus pallidus, thalamus, or cerebral cortex; in such cases the hyperkinesia is possibly caused by a loss of inhibitory neurons of the neostriatum that descend to pallidum and substantia nigra. In other words, a loss of a neuronal system of higher order has occurred, producing excessive excitation of the neurons of the next lower system. The resulting hyperkinesias are of different kinds: *athetosis, chorea, spasmodic torticollis, torsion dystonia,*
ballism, and other conditions

Athetosis

This kinetic disorder is usually caused by perinatal damage to the striate bodies. This damage takes the form of a circulatory loss of small neurons, resulting in irregular glial scars simulating veins in marble; hence the name *status marmoratus*. Involuntary movements are slow and wormlike, with a tendency to overextend the peripheral portions of the extremities. In addition, there are irregular, spasmodic increases in muscle tensions between agonists and antagonists. As a result, postures and movements are rather bizarre. Voluntary movements are severely distorted by the spontaneous appearance of hyperkinetic movements that may include face and tongue and thus cause grimacing with abnormal tongue movements. There may be spasmodic outbursts of laughing or crying. The athetosis may be combined with a contralateral paresis; it may also be bilateral and is then called double athetosis, which usually occurs in association with spastic paraplegia (Little's disease, Vogt's syndrome). Mentation may be preserved.

Chorea

The choreal syndrome is characterized by short, fast, involuntary jerks occurring in single muscles at random and producing various patterns of movements, some resembling voluntary movements. At first the peripheral portions of the extremities are involved; the proximal portions follow. Involuntary jerks of the facial muscles produce grimacing. In addition to the hyperkinesia, a decrease in the tone of the musculature is characteristic of chorea. The choreal restlessness seen in children as *chorea minor or Sydenham's chorea or St. Vitus's dance* is an acute, usually self-limiting disorder that appears to be linked closely to rheumatic fever and has been called *chorea infectiosa*. The neuropathologic findings vary greatly. Some patients show no alterations at all. Treatment: sedation, phenothiazines. The condition may become recurrent – during pregnancy, intercurrent infection. The same acute chorea may occur during early pregnancy and is referred to as *chorea gravidarum.*

Most important is *Huntington's chorea*, a dominant, hereditary, degenerative disease usually commencing in middle age. It may begin as early as adolescence with involuntary movements, which may be mistaken for parkinsonian hyperkinesia, or with mental changes, usually diagnosed as schizophrenia. The movements are generally not as jerky as those in chorea minor. They are more complex and sometimes slow like those seen in athetosis. They may be twisting, torque-like, and similar to those in torsion dystonia. The proximal extremities, the trunk, and the facial musculature are particularly involved, causing lively grimacing with forceful protrusion and retraction of the tongue. Speaking and swallowing are difficult. The early hypertonia changes later into rigor. Pathologic findings consist of an atrophy of the striate bodies associated with loss of the small neurons. Cortical neurons may also degenerate, and the disease may terminate in dementia.
These are the most important types of dystonia syndromes. In both diseases there are usually alterations within the putamen and the centromedian nucleus of the thalamus and in other extrapyramidal nuclei (pallidum, substantia nigra, and others).

Spasmodic torticollis, a tonic disorder, consists of spasmodic contractions of muscles in the neck region that result in slow, involuntary turning and bending movements of the head. The sternocleidomastoid and trapezius muscles are particularly often involved, in addition to other muscles of the neck. The causes vary. Occasionally, the spasmodic torticollis represents an abortive form of torsion dystonia or an early sign of another extrapyramidal disease, such as Huntington's chorea or Wilson's disease. Tonic spasms of the facial musculature have been considered psychogenic, as facial tics have been. After the epidemic of lethargic encephalitis in 1920, however, these hyperkinesias were very common, and postmortem studies revealed alterations, particularly in the striatum. Tic-like spasmodic contractions of the diaphragm are the cause of hiccups.

Torsion dystonia is characterized by rather extensive turning and twisting movements of trunk and proximal extremities. They can be so severe that the patient can neither stand nor walk without support. The disease may be idiopathic or symptomatic; in the latter case the cause may be birth injury, kernicterus, former encephalitis, early Huntington's chorea. Hallervorden-Spatz disease, or a hepatocerebral degeneration (Wilson's diease, Westphal-Strumpell disease).

Tics

Brief, rapid, simple or complex involuntary movements that are stereotypical and repetitive; but not rhythmic. Simple tics (eg, blinking) often begin as nervous mannerisms in childhood or later and disappear spontaneously. Complex tics often resemble fragments of normal behavior. Grilles de la Tourette's Syndrome - inherited multiple tic disorder that begins in childhood.

Gilles de la Tourette syndrome characterised by the movement disorder may begin with simple tics that progress to multiple complex tics, including respiratory and vocal ones. Vocal tics may begin as grunting or barking noises and evolve into compulsive utterances. Coprolalia (involuntary scatologic utterances) occurs in 50% of patients. Severe tics and coprolalia may be physically and socially disabling. Tics tend to be more complex than myoclonus, but less flowing than choreic movements, from which they must be differentiated. Motor and vocal tics, copropraxia (making obscene gestures), coprolalia (obscene utterances) and obsessive behaviour. Onset is in childhood, males are more often affected and the condition may be inherited. However results of a systematic genome screen were negative. A population study showed that 3% of all children and that up to 25% of children requiring special education may have mild to moderate Tourette's syndrome.

Ballistic syndrome

This syndrome usually occurs as hemiballism. Hemiballismus is violent, continuous proximal limb flinging movements confined to one side of the body, usually affecting the arm more than the leg. It is caused by a lesion, usually an infarct, in the region of the contralateral subthalamic nucleus of Luys. Differential diagnosis includes acute hemichorea, usually due to tumor or infarct of the caudate nucleus, and focal seizures. Although disabling, hemiballismus is usually self-limited, lasting 6 to 8 wk.

Myoclonic movements (brief, lightning-like contraction of a muscle or group of muscles) are usually indicative of damage in the area of the triangle of Guillain-Mollaret. Myoclonus may occur normally

as a person falls asleep (nocturnal myoclonus). Common hiccup (singultus) is a form of myoclonus affecting the diaphragmatic muscles. Etiology of abnormal myoclonus includes metabolic derangements (eg, uremia), various degenerative diseases (eg, Alzheimer's disease, progressive myoclonic epilepsy), and slow virus infections (eg, Creutzfeldt-Jakob disease, subacute sclerosing panencephalitis). Myoclonus developing after severe closed head trauma or hypoxic-ischemic brain injury may increase with intended movements; thus it is termed action myoclonus. Palatal myoclonus (a continuous, rhythmic contraction of the posterior pharyngeal muscles) is a form of tremor resulting from a lesion in the dentato-olivocerebellar circuit.