Basal Ganglia

■ INTRODUCTION

Basal ganglia are the scattered **masses of gray matter** submerged in subcortical substance of cerebral hemisphere (Fig. 151.1). Basal ganglia form the part of **extrapyramidal system,** which is concerned with motor activities.

■ COMPONENTS OF BASAL GANGLIA

Basal ganglia include three primary components:

1. Corpus striatum
2. Substantia nigra
3. Subthalamic nucleus of Luys.

■ CORPUS STRIATUM

Corpus striatum is a mass of gray matter situated at the base of cerebral hemispheres in close relation to thalamus (Fig. 151.2). Corpus striatum is incompletely divided into two parts by internal capsule:

i. Caudate nucleus
ii. Lenticular nucleus.

i. *Caudate Nucleus*

Caudate nucleus is an elongated arched gray mass, lying medial to internal capsule. Throughout its length, the caudate nucleus is related to lateral ventricle. Caudate nucleus has a head portion and a tail portion.

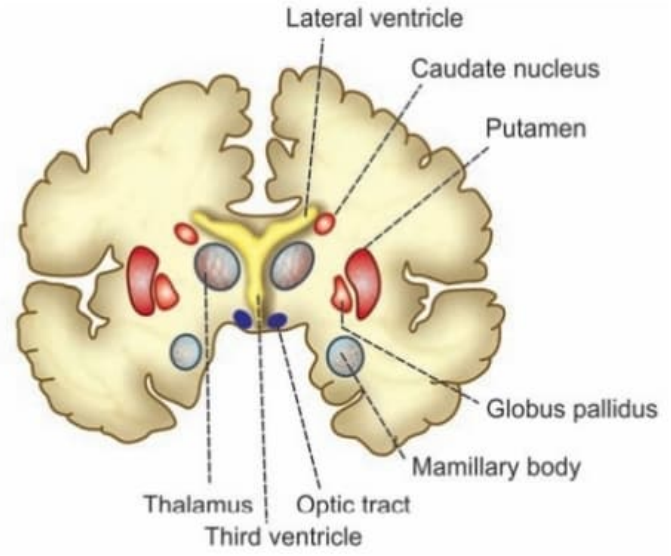

FIGURE 151.1: Basal ganglia

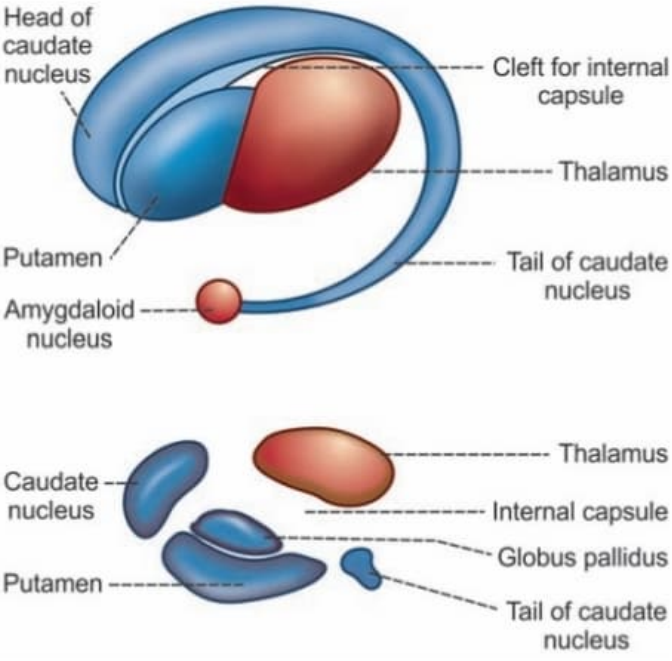

FIGURE 151.2: Corpus striatum

Head is bulged into lateral ventricle and situated rostral to thalamus. The tail is long and arched. It extends along the dorsolateral surface of thalamus and ends in amygdaloid nucleus.

ii. *Lenticular Nucleus*

Lenticular nucleus is a wedge-shaped gray mass, situated lateral to internal capsule. A vertical plate of white matter called **external medullary lamina,** divides lenticular nucleus into two portions:

a. Outer putamen
b. Inner globus pallidus.

Putamen and caudate nucleus are the phylogenetically newer parts of corpus striatum and these two parts are together called **neostriatum** or **striatum.** Globus pallidus is phylogenetically older part of corpus striatum. And, it is called **pallidum** or **paleostriatum.** Globus pallidus has two parts, an outer part and an inner part.

■ SUBSTANTIA NIGRA

Substantia nigra is situated below red nucleus. It is made up of large pigmented and small non-pigmented cells. The pigment contains high quantity of iron.

■ SUBTHALAMIC NUCLEUS OF LUYS

Subthalamic nucleus is situated lateral to red nucleus and dorsal to substantia nigra.

■ CONNECTIONS OF BASAL GANGLIA

Afferent and efferent connections of corpus striatum (Figs. 151.3 and 151.4), substantia nigra and subthalamic nucleus of Luys are given in Table 151.1.

In addition to afferent and efferent connections, different components of corpus striatum of the same side are interconnected by intrinsic fibers.

1. Putamen to globus pallidus
2. Caudate nucleus to globus pallidus
3. Caudate nucleus to putamen.

Different components of corpus striatum in each side are connected to those of the opposite side by commissural fibers.

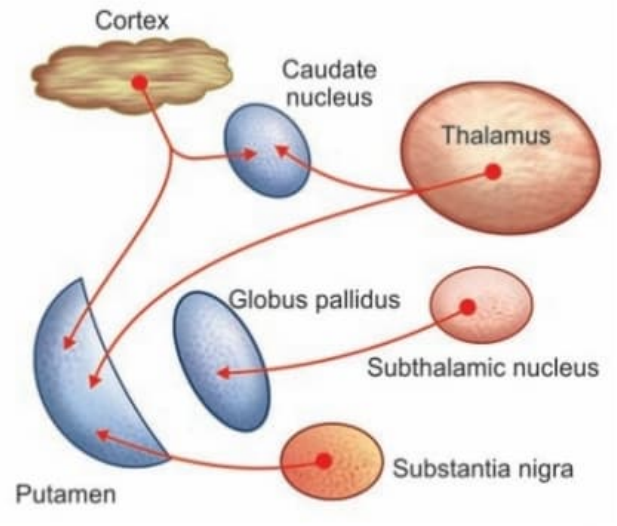

FIGURE 151.3: Afferent connections of corpus striatum

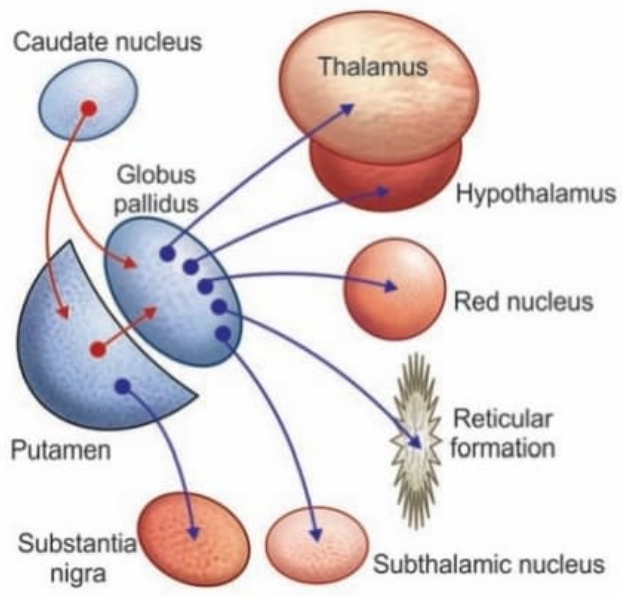

FIGURE 151.4: Efferent and intrinsic connections of corpus striatum

■ FUNCTIONS OF BASAL GANGLIA

Basal ganglia form the part of **extrapyramidal system,** which is concerned with integration and regulation motor activities. Various functions of basal ganglia are:

■ 1. CONTROL OF MUSCLE TONE

Basal ganglia control the muscle tone. In fact, gamma motor neurons of spinal cord are responsible for development of tone in the muscles (Chapter 157). Basal ganglia **decrease** the **muscle tone** by inhibiting gamma motor neurons through descending inhibitory reticular system in brainstem. During the lesion of basal ganglia, muscle tone increases leading to rigidity.

■ 2. CONTROL OF MOTOR ACTIVITY

i. *Regulation of Voluntary Movements*

Movements during voluntary motor activity are initiated by cerebral cortex. However, these movements are controlled by basal ganglia, which are in close association with cerebral cortex. During lesions of basal ganglia, the control mechanism is lost and so the movements become inaccurate and awkward.

Basal ganglia control the motor activities because of the nervous (neuronal) circuits between basal ganglia and other parts of the brain involved in motor activity. Neuronal circuits arise from three areas of the cerebral cortex:

a. Premotor area
b. Primary motor area
c. Supplementary motor area (Chapter 152).

All these nerve fibers from cerebral cortex reach the caudate nucleus. From here, the fibers go to putamen. Some of the fibers from cerebral cortex go directly to putamen also. Putamen sends fibers to globus pallidus. Fibers from here run towards the thalamus, subthalamic nucleus of Luys and substantia nigra. Subthalamic nucleus and substantia nigra are in turn, projected into thalamus. Now, the fibers from thalamus are projected back into primary motor area and other two motor areas, i.e. premotor area and supplementary motor area.

TABLE 151.1: Connections of basal ganglia

Component	Afferent connections from	Efferent connections to
Corpus striatum	1. Thalamic nuclei to caudate nucleus and putamen 2. Cerebral cortex to caudate nucleus and putamen 3. Substantia nigra to putamen 4. Subthalamic nucleus to globus pallidus	1. Thalamic nuclei 2. Subthalamic nucleus 3. Red nucleus 4. Substantia nigra 5. Hypothalamus 6. Reticular formation (Most of the fibers leave from globus pallidus)
Substantia nigra	1. Putamen 2. Frontal lobe of cerebral cortex 3. Superior colliculus 4. Mamillary body of hypothalamus 5. Medial and lateral lemnisci 6. Red nucleus	Putamen
Subthalamic nucleus of Luys	Globus pallidus	1. Globus pallidus 2. Red nucleus

ii. *Regulation of Conscious Movements*

Fibers between cerebral cortex and caudate nucleus are concerned with regulation of conscious movements. This function of basal ganglia is also known as the **cognitive control** of activity. For example, when a stray dog barks at a man, immediately the person, understands the situation, turns away and starts running.

iii. *Regulation of Subconscious Movements*

Cortical fibers reaching putamen are directly concerned with regulation of some subconscious movements, which take place during trained motor activities, i.e. skilled activities such as writing the learnt alphabet, paper cutting, nail hammering, etc.

■ 3. CONTROL OF REFLEX MUSCULAR ACTIVITY

Some reflex muscular activities, particularly **visual** and **labyrinthine reflexes** are important in maintaining the posture. Basal ganglia are responsible for the coordination and integration of impulses for these reflex activities.

During lesion of basal ganglia, the postural movements, especially the visual and labyrinthine reflexes become abnormal. These abnormal movements are associated with rigidity. Rigidity is because of the loss of inhibitory influence from the cerebral cortex on spinal cord via basal ganglia.

■ 4. CONTROL OF AUTOMATIC ASSOCIATED MOVEMENTS

Automatic associated movements are the movements in the body, which take place along with some motor activities. Examples are the swing of the arms while walking, appropriate facial expressions while talking or doing any work. Basal ganglia are responsible for the automatic associated movements.

Lesion in basal ganglia causes absence of these automatic associated movements, resulting in **poverty of movements.** Face without appropriate expressions while doing any work is called **mask-like face.** Body without associated movements is called **statue-like body.**

■ 5. ROLE IN AROUSAL MECHANISM

Globus pallidus and red nucleus are involved in arousal mechanism because of their connections with reticular formation. Extensive lesion in globus pallidus causes drowsiness, leading to sleep.

■ 6. ROLE OF NEUROTRANSMITTERS IN THE FUNCTIONS OF BASAL GANGLIA

Functions of basal ganglia on motor activities are executed by some neurotransmitters released by nerve endings within basal ganglia. Following neurotransmitters are released in basal ganglia (Table 151.2):

1. Dopamine released by dopaminergic fibers from substantia nigra to corpus striatum (putamen and caudate nucleus: dopaminergic nigrostriatal fibers): deficiency of dopamine leads to parkinsonism
2. Gamma-aminobutyric acid (GABA) secreted by intrinsic fibers of corpus striatum and substantia nigra
3. Acetylcholine released by fibers from cerebral cortex to caudate nucleus and putamen
4. Substance P released by fibers from globus pallidus reaching substantia nigra
5. Enkephalins released by fibers from globus pallidus reaching substantia nigra
6. Noradrenaline secreted by fibers between basal ganglia and reticular formation
7. Glutamic acid secreted by fibers from subthalamic nucleus to globus pallidus and substantia nigra.

Among these neurotransmitters, dopamine and GABA are inhibitory neurotransmitters. So, the fibers

TABLE 151.2: Neurotransmitters involved in the functions of basal ganglia

Neurotransmitter	Released by	Action
1. Dopamine	Fibers from substantia nigra to corpus striatum	Inhibition
2. Gamma aminobutyric acid	Intrinsic fibers of corpus striatum and substantia nigra	Inhibition
3. Acetylcholine	Fibers from cerebral cortex to caudate nucleus and putamen	Excitation
4. Substance P	Fibers from globus pallidus reaching substantia nigra	Excitation
5. Enkephalins	Fibers from globus pallidus reaching substantia nigra	Excitation
6. Noradrenaline	Fibers between basal ganglia and reticular formation	Excitation
7. Glutamic acid	Fibers from subthalamic nucleus to globus pallidus and substantia nigra	Excitation

releasing dopamine and GABA are inhibitory fibers. All other neurotransmitters have excitatory function.

APPLIED PHYSIOLOGY – DISORDERS OF BASAL GANGLIA

1. PARKINSON DISEASE

Parkinson disease is a slowly progressive degenerative disease of nervous system associated with destruction of brain cells, which produce dopamine. It is named after the discoverer **James Parkinson.** It is also called **parkinsonism** or **paralysis agitans.** Great boxer Mohammed Ali is affected by parkinsonism because of repeated blows he might have received on head resulting in damage of brain cells producing dopamine.

Causes of Parkinson Disease

Parkinson disease occurs due to lack of **dopamine** caused by damage of basal ganglia. It is mostly due to the destruction of **substantia nigra** and the **nigrostriatal pathway,** which has dopaminergic fibers. Damage of basal ganglia usually occurs because of the following causes:

i. Viral infection of brain like encephalitis
ii. Cerebral arteriosclerosis
iii. Injury to basal ganglia
iv. Destruction or removal of dopamine in basal ganglia. It occurs mostly due to long-term treatment with antihypertensive drugs like reserpine. Parkinsonism due to the drugs is known as **drug-induced parkinsonism.**
v. Unknown causes: Parkinsonism can occur because of the destruction of basal ganglia due to some unknown causes. This type of parkinsonism is called **idiopathic parkinsonism.**

Signs and Symptoms of Parkinson Disease

Parkinson disease develops very slowly and the early signs and symptoms may be unnoticed for months or even for years. Often the symptoms start with a mild noticeable tremor in just one hand. When the tremor becomes remarkable the disease causes slowing or freezing of movements followed by rigidity.

Following are the common signs and symptoms of Parkinson disease:

i. *Tremor*

Refer Chapter 147 for details of tremor. In Parkinson disease, the tremor occurs during rest. But it disappears while doing any work. So, it is called **static tremor** or **resting tremor.** It is also called **drum-beating tremor,** as the movements are similar to beating a drum. Thumb moves rhythmically over the index and middle fingers. These movements are called **pill-rolling movements.**

ii. *Slowness of movements*

Over the time, movements start slowing down **(brady-kinesia)** and it takes a long time even to perform a simple task. Gradually the patient becomes unable to initiate the voluntary activity **(akinesia)** or the voluntary movements are reduced **(hypokinesia).** It is because of hypertonicity of the muscles.

iii. *Poverty of movements*

Poverty of movements is the loss of all automatic associated movements. Because of absence of the automatic associate movements, the body becomes **statue-like.** The face becomes **mask-like,** due to absence of appropriate expressions like blinking and smiling.

iv. *Rigidity*

Stiffness of muscles occurs in limbs resulting in rigidity of limbs. The muscular stiffness occurs because of increased muscle tone which is due to the removal of inhibitory influence on gamma motor neurons. It affects both flexor and extensor muscles equally. So, the limbs become more **rigid like pillars.** The condition is called **lead-pipe rigidity.** In later stages the rigidity extends to neck and trunk.

v. *Gait*

Gait refers to manner of walking. The patient looses the normal gait. Gait in Parkinson disease is called **festinant gait.** The patient walks quickly in short steps by bending forward as if he is going to catch up the center of gravity.

vi. *Speech problems*

Many patients develop speech problems. They may speak very softly or sometimes rapidly. The words are repeated many times. Finally, the speech becomes slurred and they hesitate to speak.

vii. *Emotional changes*

The persons affected by Parkinson disease are often upset emotionally.

viii. *Dementia*

In later stages, some patients develop dementia (Chapter 162).

Treatment for Parkinson Disease

As Parkinson disease is due to lack of dopamine caused by damage of dopaminergic fibers, it is treated by **dopamine injection.**

Dopamine does not cross the blood-brain barrier. So, another substance called **levodopa** (L-dopa) which crosses the blood-brain barrier is injected. L-dopa moves into the brain and there it is converted into dopamine. Since, L-dopa can be converted into dopamine in liver, some side effects occur due to excess dopamine content in liver and blood. So, along with L-dopa, another substance called **carbidopa** is administered. Carbidopa prevents the conversion of L-dopa into dopamine and carbidopa cannot pass through blood-brain barrier. Thus, L-dopa moves into the brain tissues and is converted into dopamine.

Some of the symptoms of Parkinson disease such as tremor are abolished by **surgical destruction** of basal ganglia or thalamic nuclei.

■ 2. WILSON DISEASE

Wilson disease is an inherited disorder characterized by excess of copper in the body tissues. It is also known as **progressive hepatolenticular degeneration.** This disease develops due to damage of the lenticular nucleus particularly, putamen.

In Wilson disease, copper is deposited in the liver, brain, kidneys and eyes. **Copper deposits** cause damage of tissues. And the affected organs stop functioning.

In addition to symptoms of Parkinson disease, **liver failure** and damage to the central nervous system are the most predominant effects of this disorder. Wilson disease is fatal if not treated early.

■ 3. CHOREA

Chorea is an abnormal involuntary movement. Chorea means **rapid jerky movements.** It mostly involves the limbs. Chorea is due to the lesion in caudate nucleus and putamen.

■ 4. ATHETOSIS

Athetosis is another type of abnormal involuntary movement, which refers to slow **rhythmic and twisting movements.** It is because of the lesion in caudate nucleus and putamen.

■ 5. CHOREOATHETOSIS

Choreoathetosis is the condition characterized by aimless involuntary muscular movements. It is due to combined effects of chorea and athetosis.

■ 6. HUNTINGTON CHOREA

Huntington disease is an inherited progressive neural disorder due to the degeneration of neurons secreting GABA in corpus striatum and substantia nigra. This disease starts mostly in middle age. It is characterized by chorea, hypotonia and dementia. In severe cases bilateral wasting of muscles occurs. It is otherwise called **Huntington disease, chronic progressive chorea, degenerative chorea** or **hereditary chorea.**

■ 7. HEMIBALLISMUS

Hemiballismus is a disorder characterized by violent involuntary abnormal movements on one side of the body involving mostly the arm. While walking, the arm swings widely. These movements are called the **flinging movements.** These movements are due to the **release phenomenon** because of the absence of inhibitory influence on movements. Hemiballismus occurs due to degeneration of subthalamic nucleus of Luys.

■ 8. KERNICTERUS

Kernicterus is a form of brain damage in infants caused by **severe jaundice.** Basal ganglia are the mainly affected parts of brain. Refer Chapter 21 for details.