"Basal Nuclei / Basal Ganglia (Part 1/1) "

Th. I CN I IC I LN (P+GP) I EC I Clau. I Insula
(Medial -> Lateral)

》 Role

- Control of posture and voluntary movement.
- No direct input/output connections with the spinal cord.

》 Terminology: Collection of masses of gray matter within each cerebral hemisphere

> Structures in Basal Nuclei:
- Corpus striatum
- Amygdaloid nucleus
- Claustrum

> Functionally Related Structures (not included in basal nuclei):
- Subthalamic nuclei
- Substantia nigra
- Red nucleus

"Corpus Striatum"

》 Location and Structure

- Lateral to the thalamus.
- Divided by internal capsule into:
  - Caudate nucleus
  - Lentiform nucleus

》 Striatum Appearance

- Striated appearance due to gray matter strands connecting caudate nucleus to putamen.

"Caudate Nucleus"

》 Description

- Large C-shaped mass of gray matter.
- Lateral to thalamus, related to lateral ventricle.
- Divided into head, body, and tail.

》 Head

- Large and rounded.
- Forms lateral wall of anterior horn of lateral ventricle.
- Continuous with putamen inferiorly (neostriatum/striatum).
- Striated appearance due to gray matter strands through internal capsule.

》 Body

- Long and narrow.
- Continuous with head at interventricular foramen.
- Forms part of the floor of the body of the lateral ventricle.

》 Tail

- Long and slender.
- Continuous with body at posterior end of thalamus.
- Follows contour of lateral ventricle, continues in the roof of the inferior horn of the lateral ventricle.
- Terminates anteriorly in the amygdaloid nucleus.

"Lentiform Nucleus"

》 Structure and Location

- Wedge-shaped mass of gray matter.
- Broad convex base directed laterally; blade directed medially.
- Buried in the white matter of the cerebral hemisphere.

》 Medial Relation

- Internal capsule separates it from the caudate nucleus and thalamus.

》 Lateral Relation

- Separated from the claustrum by the external capsule (thin sheet of white matter).
- Claustrum separates it from the subcortical white matter of the insula.
- Divided by vertical white matter plate into:

》 Putamen:

- Larger, darker lateral portion.

》 Globus Pallidus:

- Inner, lighter portion with high concentration of myelinated nerve fibers.

》 Connections

- Putamen is continuous inferiorly at its anterior end with the head of the caudate nucleus.

"Amygdaloid Nucleus"

》 Location

- Situated in the temporal lobe near the uncus.
- Part of the limbic system

》 Function

- Influences body responses to environmental changes, e.g., fear alters heart rate, blood pressure, skin color, and respiration rate.

"Susbstantia Nigra and Subthalamic Nuclei"

》 Substantia Nigra

- Located in the midbrain.
- Functionally related to basal nuclei activities.
- Contains dopaminergic and inhibitory neurons.
- Many connections to the corpus striatum.

》 Subthalamic Nuclei

- Located in the diencephalon.
- Functionally related to basal nuclei activities.
- Contains glutaminergic and excitatory neurons.
- Many connections to the globus pallidus and substantia nigra.

"Claustrum"

》 Structure and Location

- Thin sheet of gray matter.
- Separated from the lentiform nucleus by the external capsule.
- Lateral to the claustrum is the subcortical white matter of the insula.

》 Function

- Function is currently unknown.

"Connections of the Corpus Striatum and Globus Pallidus"

》 Input and Output Sites

> Caudate Nucleus and Putamen:
- Main sites for receiving input to the basal nuclei.

> Globus Pallidus:
- Major site from which output leaves the basal nuclei.
- No direct input from or output to the spinal cord.

"Corpus Striatum Afferent Fibers"

》 Corticostriate Fibers

- Projections from all parts of the cerebral cortex to the caudate nucleus and putamen.
- Each cortex part projects to a specific part of the caudate-putamen complex.
- Largest input from the sensory motor cortex.
- Neurotransmitter: Glutamate.

》 Thalamostriate Fibers

- Projections from the intralaminar nuclei of the thalamus to the caudate nucleus and putamen.

》 Nigrostriatal Fibers

- Projections from neurons in the substantia nigra to the caudate nucleus and putamen.
- Neurotransmitter: Dopamine.
- Function: Inhibitory.

》 Brainstem Striatal Fibers

- Ascending fibers from the brainstem ending in the caudate nucleus and putamen.
- Neurotransmitter: Serotonin.
- Function: Inhibitory.

"Corpus Striatum Efferent Fibers"

》 Striatopallidal Fibers

- Projections from the caudate nucleus and putamen to the globus pallidus.
- Neurotransmitter: Gamma-aminobutyric acid (GABA).

》 Striatonigral Fibers

- Projections from the caudate nucleus and putamen to the substantia nigra.
- Neurotransmitters: GABA, acetylcholine, substance P.

"Globus Pallidus Afferent Fibers"

» Striatopallidal Fibers

- Pass from the caudate nucleus and putamen to the globus pallidus.
- Neurotransmitter: GABA.

"Globus Pallidus Efferent Fibers"

» Pallidofugal Fibers: Divided into following groups:

i) Ansa Lenticularis: Pass to the thalamic nuclei.

ii) Fasciculus Lenticularis: Pass to the subthalamus.

iii) Pallidotegmental Fibers: Terminate in the caudal tegmentum of the midbrain.

iv) Pallidosubthalamic Fibers: Pass to the subthalamic nuclei.

"Basal Nuclei Functions"

I) Connections and Information Flow

- Basal nuclei are interconnected with numerous regions of the nervous system via complex neural circuits.

》 Afferent Inputs to Corpus Striatum

- Cerebral cortex
- Thalamus
- Subthalamus
- Brainstem (including substantia nigra)

》 Integration

- Information is processed within the corpus striatum.
- Output is relayed back to the cerebral cortex, thalamus, subthalamus, and brainstem.

》 Sources of Initiation: Activity of basal ganglia is initiated by information received from:

- Premotor and supplemental motor areas
- Primary sensory cortex
- Thalamus
- Brainstem

》 Output Pathway (Feedback loop):

- Outflow from basal nuclei is routed through the globus pallidus.
- Influences motor areas of the cerebral cortex or brainstem motor centers.

2) Role in Movement Control

- Basal nuclei influence muscular movements indirectly by modulating the cerebral cortex.
- They do not directly control motor pathways descending to the brainstem and spinal cord.
- Effects:
  - Regulate voluntary movement.
  - Facilitate learning of motor skills.

3) Influence on Skilled Cortical Motor Activities:

- Writing (letters, words)
- Drawing (diagrams, shapes)
- Sports (passing a football)
- Vocalization (talking, singing)
- Eye movements (tracking objects)

4) Impact of Damage

> Primary Motor Cortex Damage:

- Loss of fine, discrete movements of hands and feet on the opposite side.
- Gross movements of opposite limbs remain possible.

> Corpus Striatum Damage:

- Results in paralysis of remaining movements on the opposite side.

5) Preparatory Role for Movement

- Basal nuclei assist in preparing for movement by:
- Controlling axial (trunk) and girdle movements.
- Positioning proximal limbs.

"Clinicals"

》 Disorders of the Basal Nuclei: Types of Disorders

> Hyperkinetic Disorders: Excessive and abnormal movements (e.g., chorea, athetosis, ballism).

> Hypokinetic Disorders: Lack or slowness of movement.

> Parkinson Disease: Includes both hyperkinetic and hypokinetic motor disturbances.

"Chorea"

》 Characteristics:

- Involuntary, quick, jerky, irregular movements.
- Nonrepetitive.
- Examples: Swift grimaces, sudden head or limb movements.

"Huntington Disease"

》 Inheritance: Autosomal dominant, onset in adult life, death 15–20 years after onset.

» Genetic Cause: Single gene defect on chromosome 4, protein huntingtin.

» Pathophysiology:

- CAG codon for glutamine repeated excessively.
- Affects men and women equally, often manifests after childbearing age.

» Signs and Symptoms:

1. Choreiform Movements:

- Initial involuntary extremity movements, facial grimacing.
- Later, more muscle involvement, leading to immobility, inability to speak/swallow.

2. Progressive Dementia:

- Loss of memory, intellectual capacity.

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》 Neuronal Degeneration:

- GABA-secreting, substance P-secreting, acetylcholine-secreting neurons of the striatonigral-inhibiting pathway degenerate.
- Overactive dopamine-secreting neurons of the substantia nigra.
- Nigrostriatal pathway inhibits caudate nucleus and putamen.
- Abnormal movements due to this inhibition

》 Imaging: Enlarged lateral ventricles on CT scans due to caudate nuclei degeneration.

》 Treatment: Medical treatment has been disappointing.

"Sydenham Chorea"

》 Characteristics: Rapid, irregular, involuntary movements of limbs, face, trunk.

》 Association: Childhood disease linked to rheumatic fever.

》 Pathophysiology:

- Antigens of streptococcal bacteria mimic proteins in striatal neuron membranes.
- Host antibodies attack both bacterial antigens and basal ganglia neurons.

》 Outcome: Transient choreiform movements with full recovery.

"Hemiballismus"

》 Characteristics:

- Involuntary movement confined to one side of the body.
- Involves proximal extremity musculature.
- Limb flies out of control in all directions.

》 Lesion:

- Usually a small stroke.
- Occurs in the opposite subthalamic nucleus or its connections.
- Smooth movements integrated in the subthalamic nucleus.

"Parkinson Disease"

》 General Information:

- Progressive disease of unknown cause.
- Onset between ages 45-55.
- Affects about 1 million people in the United States.
- Associated with neuronal degeneration in the substantia nigra, globus pallidus, putamen, and caudate nucleus.

》 Pathophysiology:

- Neuronal degeneration in substantia nigra → reduced dopamine release in corpus striatum.
- Results in hypersensitivity of dopamine receptors in postsynaptic neurons in the striatum.
- Cause of most symptoms: Increased inhibitory output from basal nuclei to thalamus and precentral motor cortex.

» Signs and Symptoms:

1) Tremor:

- Alternating contraction of agonists and antagonists.
- Slow tremor, most obvious at rest, disappears during sleep.
- Different from intention tremor in cerebellar disease.

2) Rigidity:

- Present equally in opposing muscle groups.
- Resistance to passive movement (plastic rigidity).
- If tremor present, muscle resistance felt as jerks (cogwheel rigidity).

3) Bradykinesia:

- Difficulty initiating (akinesia) and performing new movements.
- Slow movements, expressionless face, slurred and unmodulated voice.
- Loss of arm swinging during walking.

4) Postural Disturbances:

- Stooped posture, flexed arms.
- Short steps, difficulty stopping, may shuffle/run to maintain balance.

5) Muscle Power and Sensibility:
- No loss of muscle power or sensibility.
- Normal corticospinal tracts: normal superficial abdominal reflexes, no Babinski response, normal deep tendon reflexes.

》 Known Causes

> Postencephalitic Parkinsonism:
- Developed after viral encephalitis outbreak (1916-1917).
- Damage to basal nuclei.

> Iatrogenic Parkinsonism:
- Side effect of antipsychotic drugs (e.g., phenothiazines).

> Other Causes:
- Meperidine analogues (used by drug addicts).
- Carbon monoxide and manganese poisoning.
- Atherosclerotic parkinsonism in elderly hypertensive patients.

» Treatment

> Elevating Brain Dopamine Level:

- L-Dopa: Precursor to dopamine, crosses the blood-brain barrier, converted to dopamine in basal nuclei.

- Selegiline: Inhibits monoamine oxidase, slows degeneration of dopa-secreting neurons.

> Transplantation:

- Human Embryonic Dopamine-Producing Neurons:
  - Improvement in motor function.
  - Grafts can survive and form synaptic contacts.
  - Many grafted neurons do not survive, continuing degeneration counteracts improvement.

- Autotransplantation of Suprarenal Medullary Cells: Source of dopa-producing cells.

- Genetically Engineered Cells: Future potential source of dopa.

> Surgical Lesions:

• Pallidotomy: Effective in alleviating parkinsonian signs.
- Restricted to patients not responding to medical treatment.

"Drug-Induced Parkinsonism"

》 Prevalence: Increasing in clinical practice.

》 Causes:
- Drugs blocking striatal dopamine receptors (D2) for psychotic behavior (e.g., phenothiazines, butyrophenones).
- Drugs depleting striatal dopamine (e.g., tetrabenazines).

》 Reversibility: Disappears once the agent is withdrawn.

"Athetosis"

》 Characteristics:
- Slow, sinuous, writhing movements.
- Commonly involve distal segments of limbs.

» Pathophysiology:

- Degeneration of the globus pallidus.
- Breakdown of circuitry involving basal nuclei and cerebral cortex.