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# Movement disorders

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# Neuroanatomical consideration

**Many movement disorders are attributed to diseases in the basal ganglia which consists of group of ganglia & nuclei situated deeply in the brain & brain stem, the term basal ganglia refers to:**

***A- Caudate nucleus***

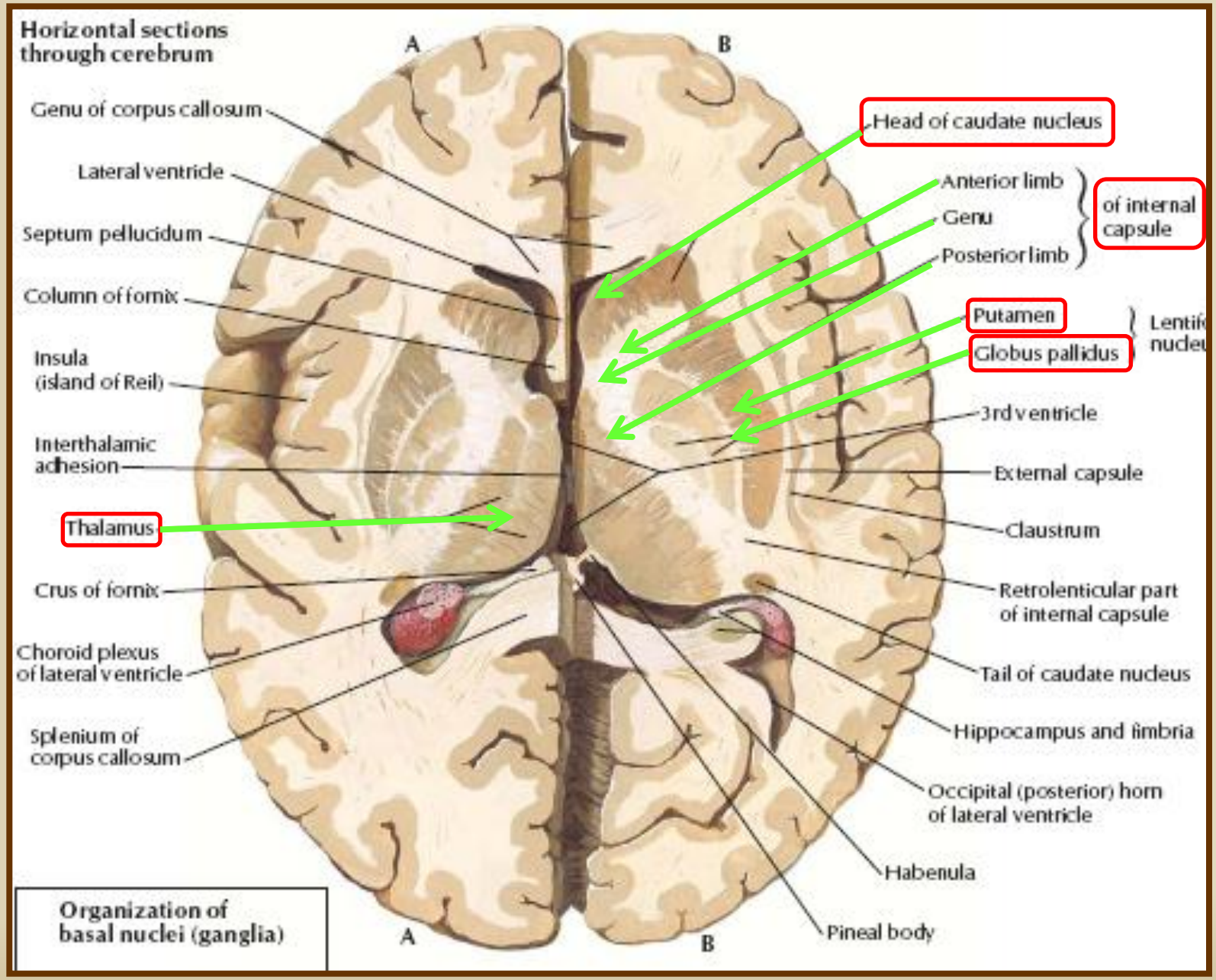
***B- Putamen***

***C - Globus pallidus***

***D- Substantia nigra***

***E - Subthalamic nucleus***

# The basal ganglia



# Neurophysiological considerations

**The basal ganglia receives afferents from:** ***the cerebral cortex, thalamus, brain stem & amygdala.***

**Sends efferents to:** ***the cerebral cortex, thalamus, brain stem & reticular formation.***

**These different connections consume many different neurotransmitters:** ***dopamine, glutamate, acetyl choline, GABA, serotonin, enkephalin, neurotensin & substance P.***

**Through these different connections the basal ganglia exert its role in controlling:**

***Voluntary motor activity → planning & programming of voluntary movements & control of subconscious automatic movements.***

***Muscle tone → Inhibition of muscle tone.***

***Cognitive function → Through connections with frontal, prefrontal & limbic structures.***

# Classification of movement disorders

**(A) Akinetic rigid syndrome (Parkinsonism).**

**(B) Dyskinesias:**

- ✓ **Chorea**
- ✓ **Dystonia**
- ✓ **Athetosis**
- ✓ **Hemiballismus**
- ✓ **Tics**
- ✓ **Myoclonus**

# Parkinsonism

**There's depletion up to 80% in striatal dopamine, also deficiency occurs to: *Tyrosine hydroxylate, melanin, Serotonin , GABA.***

**Neuropathological changes:**

- **Depletion of dopamine**
- **Decrease in the fresh volume of Substantia nigra with loss of nigral neurons**

# Akinetic rigid syndromes (Parkinsonism)

**i- Idiopathic Parkinson's disease**

**ii- Arteriosclerotic Parkinsonism**

**iii- Postencphalitic Parkinsonism**

**iv- Symptomatic Parkinsonism:**

| | |
|---|---|
| *Drugs: metoclopromide, methyldopa & phenothiazines.* | *Toxins & poisons; Co, manganese, MPTP, TIQ.* |
| *CVA* | *Cerebral anoxia* |
| *Repetitive head trauma* | *Brain tumors* |

**V-Degenerative causes (Parkinsonism plus syndromes):**

| | |
|---|---|
| *Progressive supranuclear palsy* | *Wilson's disease* |
| *Multiple system atrophy* | *Athetoid cerebral palsy* |
| *Corticobasal degeneration* | *Calcification of the BG* |

# Parkinson's disease (shaking palsy)

■**Etiology:**

**-Natural aging process in autoimmune predisposed patients.**

**-Exposure to endogenous toxins:**

- ***MPTP which is converted by MAO B to the toxic MPT which destroy nigral neurons.***
- ***TIQ***

**-May be genetically determined**

**-Defective anti- oxidant system**

■**Neuropathological & Neurophysiological changes:** ***discussed before.***

# Parkinson's disease, Clinical features:

## (1) Rigidity:

***Present in all cases with onset and presented as:***

- ***Clumsiness of movement.***
- ***Stiffness.***
- ***Difficulty in turning over the bed.***
- ***Difficulty in changing position.***
- ***Difficulty & fatigue in walking.***

***It is generalized in all muscle groups, more in axial, flexors & proximal muscles giving the body flexion attitude and stooping forward (gorilla like attitude).***

***May be equal all through the movement "lead pipe" or interrupted by the tremors "cog wheel".***

# Parkinson's disease, Clinical features:

**(2)-Tremors:**

*-Present in at least 80% of patients.*

*-Static, regular, rhythmic with rate of 4-6/sec.*

*-Pill rolling, counting money or drum tapping.*

*-Increased by emotions, fatigue & rest, decrease on movement & disappear on sleep.*

*-Marked in wrist joint & fingers, less in ankle & head.*

**(3)-Disorders of voluntary movements:**

*-Slowness of movements "bradykinesia" up to akinesia.*

*-Reduced amplitude "hypokinesia" (hand writing).*

*-Reduced or lost arm swinging*

*- Power, reflexes & planter responses, all are normal*

# Parkinson's disease, Clinical features:

**(4) Speech:** ***Soft monotonous voice.***

**(5) Gait:** ***Short steppage, festinant, shuffling.***

**(6) Facial features:** ***Mask face, reduced blinking.***

**(7)- Other features; "not in all patients"**

***-Akathesia.***

***-Propulsion–retropulsion.***

***-Autonomic features***

***-Mild eye movement disorders.***

***-Cognitive functions:*** ***Normal in early stages, later on, depression psychotic episodes may occur***

***NB;*** ***rigidity & tremors usually are confined to one limb months or ys before generalization to the other limbs.***

# Diagnosis & differential diagnosis of Parkinsonism

■ **Parkinson's disease:**

- ***Age of patient 55-70ys.***
- ***C/P as described above.***
- ***Asymmetrical presentation with gradual onset slowly progressive course.***
- ***Good response to levodopa.***
- ***Absence of symptomatic causes.***

■ **Idiopathic Parkinson's disease must be differentiated from:**

**i) Drug induced Parkinsonism:**

- ***History of taking neuroleptics in psychotic patient.***
- ***Taking antiemetics or vestibular sedatives.***
- ***Of acute or subacute onset.***
- ***Prominent extra pyramidal rigidity with tongue protrusion***

**ii) Other symptomatic cases:** ***from the history.***

# Diagnosis & differential diagnosis of Parkinsonism

**iii) Post encephalitic Parkinsonism:**

- ✓ *History of encephalitis "fever, convulsions, neurological deficits" months or years before.*
- ✓ *Any age.*
- ✓ *Prominent occulogyric crisis (spasm of up gaze).*
- ✓ *Rigidity more common.*
- ✓ *Pyramidal signs.*

**iv) Arteriosclerotic Parkinsonism:**

- ✓ *Older age.*
- ✓ *Presence of vascular risk factors: dyslipidaemia, hypertension, DM, cardiac disorders.*
- ✓ *Pyramidal signs.*
- ✓ *The complete C/P comes after frequent vascular insults.*
- ✓ *Signs & symptoms of pseudobulbar palsy.*

# Diagnosis & differential diagnosis of Parkinsonism

**v) Wilson's disease (Hepatolenticular degeneration):**

**-Age under 50 years. – Heredfamilial (AR).**

**-Diffiency of ceruloplasmin (enzyme binding copper) → deposition of copper in lentiform nucleus, liver, cornea, BG, Clinically →**

- **Behavioral & psychotic disorders.**
- **Hepatic disorders.**
- **Presence of Kayser Fleisher ring in the cornea (by slit lamp).**
- **Positive lab. Investigations:**
  - **Low ceruloplasmin in serum.**
  - **Liver biopsy → increased copper concentration.**

**Treatment:**

***D.penicillamine (copper chelating drug) 250 mg 4 times daily.***

***Oral zinc reduces copper absorption in early stages.***

***Liver transplantation in severe liver damage.***

# Treatment of Parkinson's disease:

## A -Medical treatment:

**i) Anticholinergic drugs e.g. parkinol, cogentine**

***Used as initial treatment in mild cases or with levodopa.***

***Helpful in reducing tremors & rigidity, have little effect on bradykinesia***

***Dose:*** *Cogentine 2 mg tablet, 1 tab t.d.s*

*Parkinol 5mg tablet, 1 tab t.d.s*

***Side effects:***

*-Blurring of vision - Dryness of mouth -Confusion*

*- Constipation & retention of urine especially in elderly*

***Contraindication:*** *Cardiac, hepatic patients, glaucoma.*

# Treatment of Parkinson's disease:

**ii) Levodopa;**

*Converts to dopamine by dopa decarboxylase.*

*Corner stone in treatment of Parkinson's disease*

*Given in combination with dopa decarboxylase inhibitor e.g. carbidopa to prevent peripheral effects of dopamine (ratio 10-1)*

*It s available as sinemet 275 mg tab*

***Dose:*** *must be introduced in small doses with gradual increase over a period of weeks or months up to 2gm in 4-6 divided doses*

***Side effects:***

-Nausea, vomiting -Postural hypotension

-Cardiac arrhythmias

-Psychiatric disturbances as confusion, agitation hallucination

-Movement disorders as end of dose akinesia or dyskinesia, on off effect

# Treatment of Parkinson's disease:

**iii) Monoamine oxidase B inhibitor**

***Deprenyl (jumex) prolongs levodopa action, dose; 5mg tab t.d.s up to 30 mg /day.***

**iv) Dopamine agonists (as bromocriptine)**

***Dose 2.5mg tab with gradual increase up to 20-40 mg/day.***

**(B) Surgical treatment: Deep brain stimulation.**

**(C) Recent concepts in treatment of Parkinsonism:**

**Controlled release of levodopa – carbidopa.**

**New dopamine agonist as pergolid.**

**Protein restriction.**

**COMT inhibitors.**

**Free radical scavengers & antioxidants.**

# Chorea

**Involuntary, static, sudden, jerky, dysrythmic, pseudopurposful movements due to lesion mainly in the caudate nucleus**

**Causes:**

**i) Genetically determined:**

***-Huntington chorea***

***-Benign hereditary chorea***

***-Neuroacanthocytosis.***

# Chorea, Causes:

## ii) Symptomatic chorea:

*1-Rheumatic chorea (Sydenham's chorea)*

*2 -Chorea gravidarum*

*3-Thyrotoxicosis*

*4 -SLE*

*5-Postencephalitic*

*6 -Hyperglycemia*

*7-Hypernatremia*

*8 –Hypoparathyroidism*

*9-Polycythemia*

# Chorea, Causes:

## iii) Drug induced chorea:

*1-Neuroleptics*

*2-Levodopa*

*3-Dopamine agonists*

*4-Anticholinergics*

*5-Contraceptive pills*

*6-Phenytoin*

*7-Antihistaminic*

*8-Cimitidine (H2 antagonist)*

## iv) Hemichorea & Hemiballismus:

*Stroke, tumor, trauma & multiple sclerosis.*

# Rheumatic chorea:

One of the major criteria of rheumatic fever & it is related to streptococcus group A infection but other streptococcal infection, scarlet fever & diphtheria can cause chorea

Age of onset 7-12ys, females more than males (2/3 of patients)

**Clinical picture:**

Irritability, behavioral & emotional disturbances.

Chorea may develop suddenly or gradually

Chorea may develop generalized or unilateral (20%).

In rheumatic patients, about 1/3 of patients have evidence of carditis

# Rheumatic chorea:

**Movements:**

*In the tongue, cannot keep it protruded .*

*In the face, frequent facial grimacing with sudden jerky facial muscle movement.*

*In the arm, sudden jerky movements with drop of objects, jerkiness of shoulder.*

*Movements are usually more proximal.*

**Hypotonia:**

*The affected limbs are hypotonic, on outstretching the arms there is flexion of the wrist, extension of metacarpophalangeal & interphalangeal joints giving the hand boat shaped appearance.*

# Rheumatic chorea:

**Treatment:**

***Complete rest in bed.***

***Mild sedation with diazepam, 2-4mg/t.d.s.***

***Dopamine antagonists:***

***-Haloperidol 3-10 mg t.d.s/day***

***-Chlorpromazine10 mg t.d.s up to 75 mg***

***-Na valporate may be used in resistant cases.***

***Course of long acting penicillin is needed up to the age of 20 years in rheumatic patients.***

# Differential diagnosis of choreic movement

**1-Rheumatic chorea: discussed above.**

**2-Huntington chorea:**

***-Heredofamilial, AD.***

***-Older age of onset 20 -55 years***

***-Clinically:***

- ***Generalized chorea***
- ***Early, evident psychiatric disturbances.***
- ***Prominent gait abnormalities***
- ***Dementia***
- ***Elements of Parkinsonism, dystonia & myoclonus***

# Differential diagnosis of choreic movement

**3-Neuroacanthocytosis;**

*-Age of onset 8-62 years*

*- Heredofamilial*

*-Acanthocytosis; spiky appearance of RBC (blood film).*

*-Cognitive dysfunction*

*-Axonal neuropathy*

*-Akinetic rigid syndrome & seizures may be present.*

**4- Chorea gravidarum;**

*-Common in primigravida, those with history of recurrent abortion, venous thrombosis*

*-May be associated with lupus anticoagulant*

*-Older age than that of rheumatic chorea*

# Dystonia

***Static, intermittent or repetitive, slow, sustained muscle spasm including dystonic spasm of neck muscles causing torticollis, retrocollis, anticollis, spasm of back muscles causing lordotic or scoliotic postures, planter flexion & inversion of foot, dystonic writers cramps.***

***Causes of dystonic syndromes:***

***Dystonia usually arise due to lesion in the putamen.***

***1-idiopathic dystonia, genetically determined.***

***2-symptomatic:***

| | |
|---|---|
| *Drug induced (neuroleptics, metoclopromide)* | *Trauma* |
| *Postencephalitic* | *Stroke* |
| *Toxic (CO, cyanide, methanol)* | *Post anoxic* |
| *Tumors* | *Sickle cell disease* |
| *Other genetically determined & metabolic disease (DRD, WD)* | |

# Clinical picture of idiopathic torsion dystonia

***A-Focal dystonia:***

***In adult life.***

***Axial dystonia in the trunk.***

***Spasmodic torticollis, retrocollis, antecollis .***

***Blepharospasm: contraction of orbicularis occuli.***

***Oromandibular dystonia: spasm of the jaw, mouth, tongue, pharynx & larynx causing lip protrusion, jaw closure or opening, facial grimacing, dysphagia, dysphonia.***

***Dystonic writers cramps affecting small muscles of the hand.***

***B-Generalized dystonia:***

***. Before the age of 20 years.***

***. Dystonic features appear in the arms ,trunk, neck& legs.***

***C-Segmental dystonia: wider range of age of onset, adjacent parts of the body e.g. arms & neck.***

# Treatment lines of dystonia.

***A) In generalized dystonia:***

***- Line 1→ all patients should receive 2-3 months trial of levodopa (250 mg tds)to exclude dopa responsive dystonia →on failure***

***-Line 2→ Anticholinergic e.g benzhexol up to 120 mg/day→ on failure***

***-Line 3→benzodiazepines in large doses***

***→ Tetrabenazines, phenothiazines → on failure***

***-Line 4 → unilateral or bilateral thalamotomy***

***B)In focal dystonia:***

***1- Benzhexol, benzodiazepines***

***2-Local injection of botulinum toxin type (A) in the affected muscle, will produce temporary paresis of the dystonic muscle for about 3 months, to be repeated, this treatment has changed dramatically the management of focal dystonia.***

# Hemiballismus

***Severe, static, involuntary, wide flinging movements of large amplitude due to lesions in the sub thalamus, usually cause skin abrasions, macerations, exhaustions even bone fracture.***

***Most frequently in elderly***

***Vascular causes e.g. infarction or hemorrhage (commonest causes).***

***Many patient may resolves into hemichorea after Hemiballismus.***

***Good response is usually achieved with tetrabenazines***

# Athetosis

***Distal movements, in frequency lies between chorea (more rapid) & dystonia (more slow), it is static, involuntary, snake like movement, eg; oriental dancing movement.***

***Usually accompanied clinically with chorea, choreo-athetosis.***

***Treatment with anticholinergic & dopamine antagonists.***

# Tics

***Involuntary, repetitive, stereotyped motor acts that resemble normal pattern of muscle contractions.***

***Common in childhood, common in hands, neck & eye muscles.***

***Motor tics: simple (blinking, hand nodding, blepharospasm), complex (copropraxia, echopraxia, jumping, hitting).***

***Vocal tics: simple (blowing, sneezing, suckling, nasal clearing), complex (palilalia, coprolalia, echolalia)***

***Gilles de la Tourette syndrome: complex tics, multiple motor & vocal ones, with behavioral & learning problems.***

***Haloperidol can control the tics.***


Thank You