# MOVEMENT DISORDERS

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# Topics of discussion

- What is movement disorder?
- Types of movement disorder
- Hypokinetic - parkinsonism
- Hyperkinetic
  - chorea
  - athetosis
  - hemiballismus
  - tremor
  - dyskinesias
  - dystonia
  - myoclonus
  - asterixis
  - tics
  - fasciculations

# Introduction

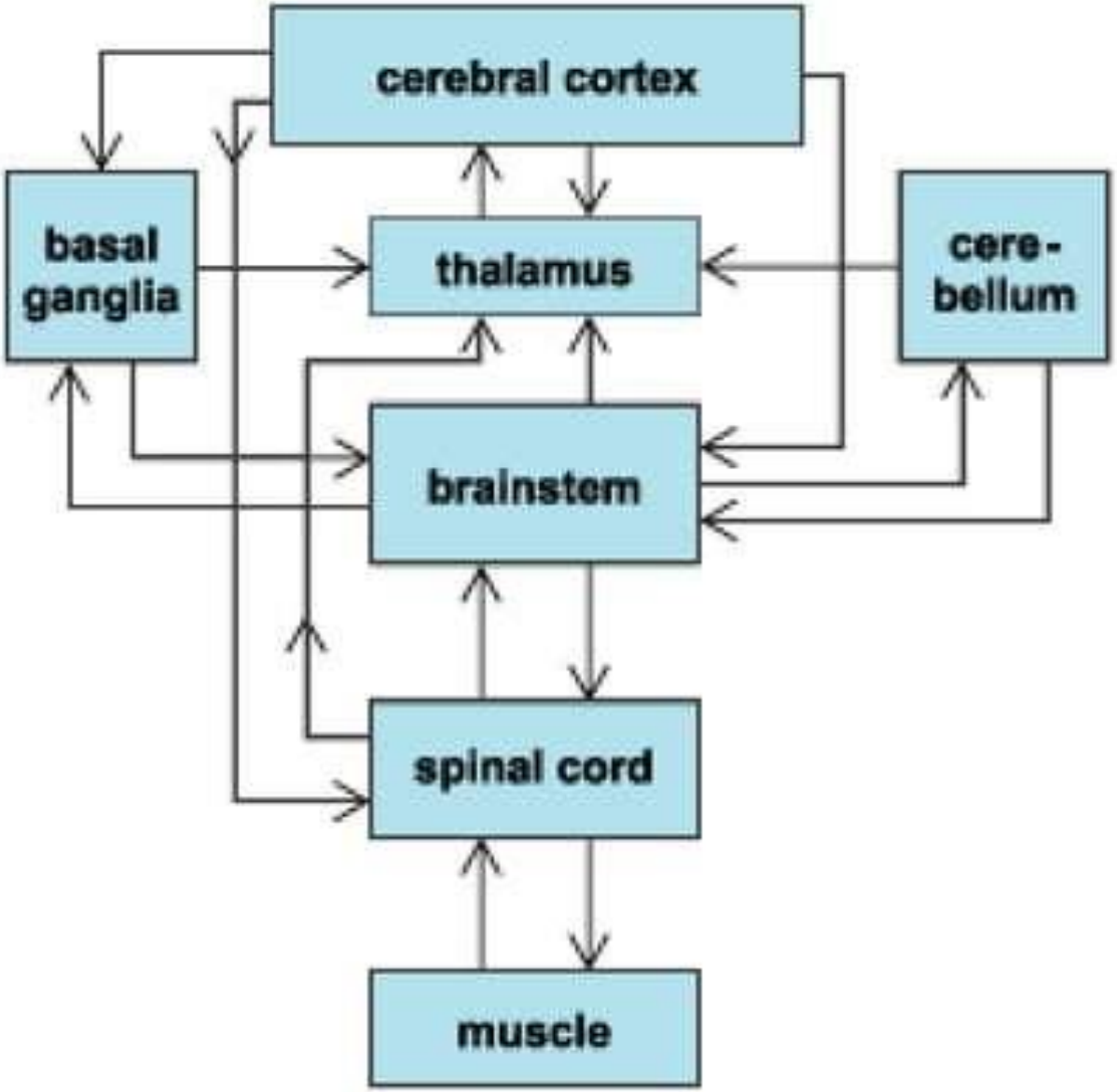
- **Movement disorder:** Term used for

1. physical sign of abnormal movement in absence of weakness
2. the syndrome that causes such motor abnormalities

- Movement **disorders disrupt motor function** by

1. Abnormal, involuntary, unwanted movements (hyperkinetic movement disorders)
2. **Curtailing [restricting]** the amount of normal free flowing, fluid movement (hypokinetic movement disorders)

- hypokinetic movement disorders are accompanied by **abnormal states of increased muscle tone**
- Pathology is in basal ganglia


cerebral cortex
basal ganglia
thalamus
cere-bellum
brainstem
spinal cord
muscle

# Basal Ganglia : Anatomy

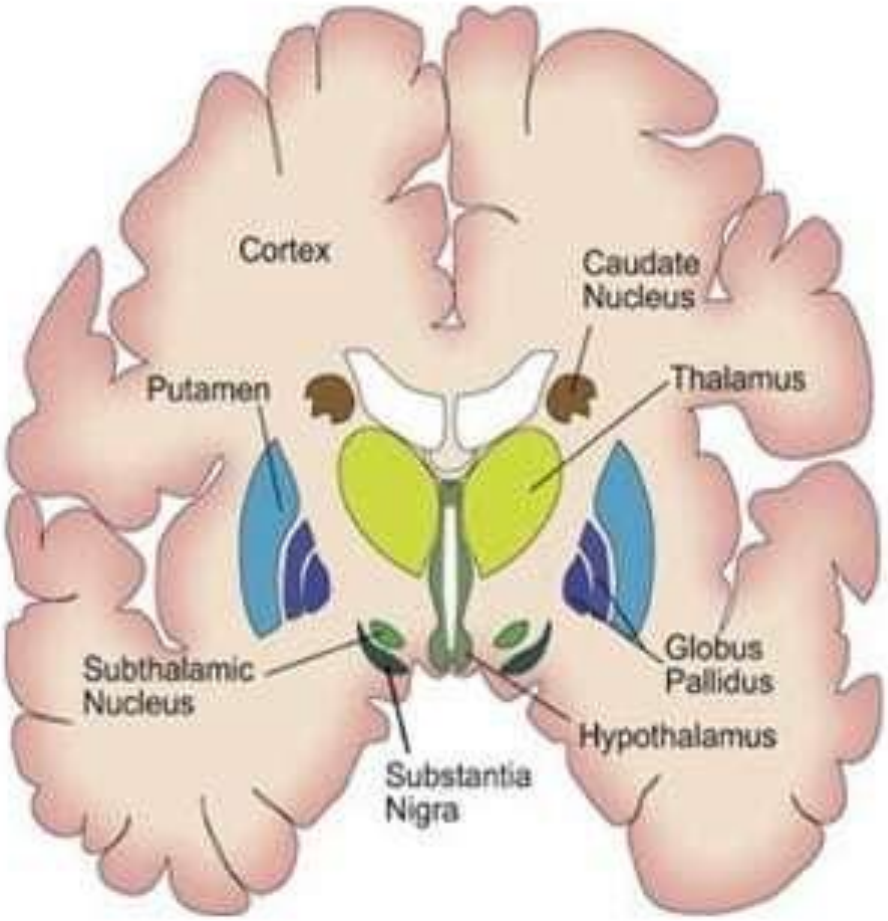
- **Caudate nucleus** : along lateral side of each lateral ventricle
- **Putamen**
- **Globus pallidus** (GPi & GPe)
- **Subthalamic nucleus** : small structure on border between brain stem & cerebrum, lateral & inferior to hypothalamus
- **Substantia nigra** (SNc & SNr): Histologically, 2 portions - SNc & SNr

# PHYSIOLOGY:

- BG **do not connect directly** to spinal neurons
  - exert their influence indirectly via **corticospinal, tectospinal and reticulospinal** pathways

- Lack of direct sensory input and motor output suggests that they act **as modulators** rather than executors of movement

- **Cortex → striatum → pallidum → thalamus → cortex**



# PHYSIOLOGY:

- **Striatum - major receiving area**: Input from all areas of cortex
- Output to GPe + SNr
- GPe + SNr are the main **output stations** of the basal ganglia → thalamus → cortex, superior coll, reticular formation
- **Subthalamic nucleus controls (brake)** this output through a feedback circuit with GP
- **Nigrostriatal pathway (dopaminergic) modulates striatal output**

# Clinico-pathologic correlation

| | |
|---|---|
| Parkinsonism | C/l substantia nigra |
| U/l hemiballismus | c/l subthalamic nucleus |
| Chronic chorea - 's | Caudate nucleus/putamen |
| Athetosis , dystonia | c/l putamen or thalamus |
| myoclonus | ? Cerebellar cortex/ thalamus |
| Rhythmic palatal/facial myoclonus | Central tegmental tract, inf olivary nuc, olivodentate fibres |

# Hypokinetic Movement Disorder

- Parkinson's disease (PD): Archetype of hypokinetic movement disorders

- Hypokinetic movement disorders are usually called Akinetic-rigid syndromes
- About 80% of akinetic-rigid syndrome are due to PD

- Disorders, whose features resemble PD are referred to as parkinsonism, or parkinsonian or Parkinson syndrome or parkinson plus

- Parkinsonism is a clinical diagnosis appropriate in the presence of

1. resting tremor,
2. bradykinesia,
3. rigidity, and
4. impaired postural reflexes

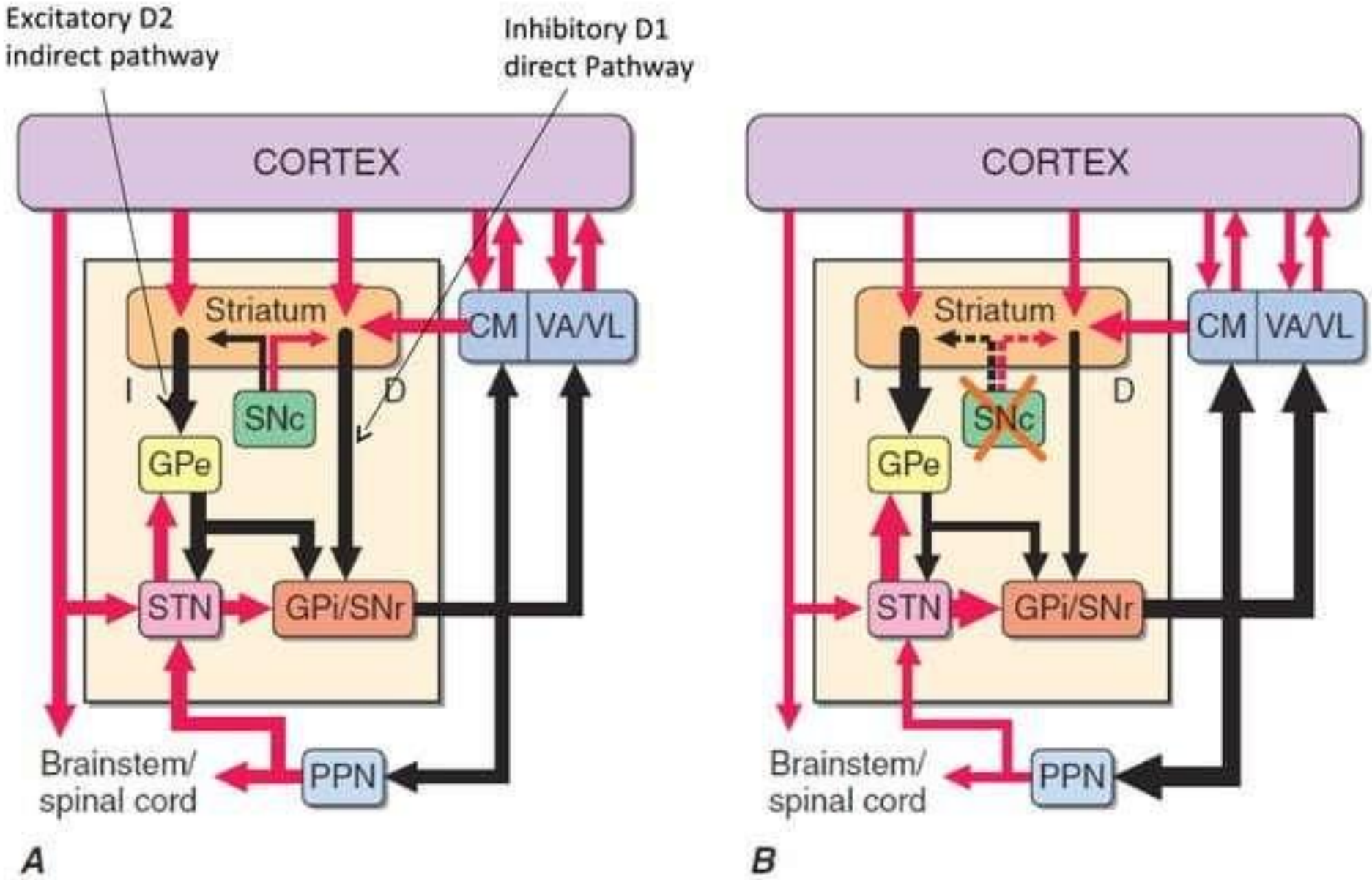
# Parkinson's disease

- Parkinson's disease is a progressive neuro-degenerative disorder that is associated with the loss of dopaminergic neurons in the substantia nigra pars compacta.

- The hallmarks of the disease are its triad of motor features
  - resting tremor
  - rigidity
  - akinesia/bradykinesia

- **Gait and postural disturbances** also characterize the disease.

# Cortico-striatal-thalamic circuits



# How movements produced in PD?

- Projections from SNpc to striatal targets are inhibitory in the dopamine **type 1 ($D_1$) receptor-mediated direct path** and **excitatory in the $D_2$ receptor-mediated indirect path.**

- The loss of dopamine results in hypokinetic symptoms secondary to **overactivity of the STN and Gpi via the indirect path, which increases inhibitory input to the thalamus** and then reduces the excitatory thalamocortical activity that ordinarily facilitates motor movements.

- Hyperkinetic movements, such as levodopa-induced dyskinesias occur **with overactivation of the direct pathway.**

## Direct and Indirect Pathways

- **Direct pathway**
  - **Facilitates movement**
- **Indirect pathway**
  - **Inhibits movement**

**Dopamine facilitates movement through the activation of the direct pathway and the inhibition of the indirect pathway**

# Parkinsonian Tremor


- Resting, static, or non-intention tremor
- slow, coarse, and compound in type

- Onset is **usually in one hand**; it may later involve the contralateral upper limb or ipsilateral lower limb.
- The rate vary from 2 to 6 Hz, **averaging 4 to 5 Hz**
-
- **Movement consists: alternate contractions of agonist and antagonist**, involving the flexors, extensors, abductors, and abductors of the fingers and thumb, together with motion of the wrist and arm
- It leads to pill-rolling
- relatively rhythmic
- present at rest
- may be temporarily suppressed by movement.

# Rigidity

- Rigidity is an **increased resistance to passive stretch**
- **Normally:** resistance is nearly equal in both agonist and antagonist muscles and generally uniform throughout the range of motion of the joint being tested
- It may be **sustained** (plastic or "lead pipe") or **intermittent** and ratchetty ("cogwheel")
- **Cogwheel rigidity** : parkinsonian rigidity complicated by parkinsonian tremor
- **asymmetric** in early PD
- commonly present at one or both **wrists** and in the **neck**
- **may manifest as a slightly flexed elbow on the more affected side in early disease**

# Bradykinesia

- bradykinesia is usually its **most disabling** component

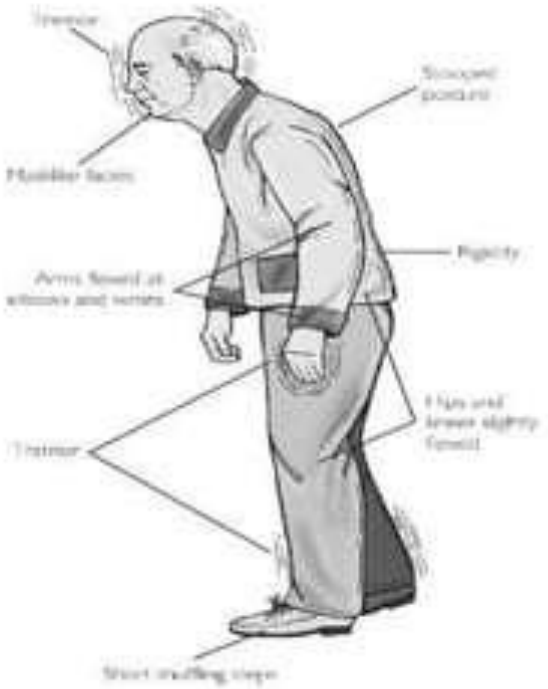
- **Bradykinesia**: slowness of voluntary movements and poverty of normal associated movements
- **Akinesia**: extension of bradykinesia implying nearly absent voluntary movement

- **Can be early sign in different body parts**
- In **eyes** it presents with **saccadic hypometria**
- In **face**
  - reduced frequency of blinking
  - diminished facial animation

# Postural Instability and Gait Disturbance

- Significant impairment is **rare** in early PD
- Usually occurs **about 5 years** after the onset of the disease

- **In early PD**, the posture may show
  - **slight flexion of the neck** or trunk
  - **slight lean** to one side
- **Abnormalities of gait** include
  - asymmetrically **reduced arm swing**,
  - overall **slowing of gait** and early fatigue,
  - **shortened stride length**
  - **intermittent shuffle**, or tripping over objects,
  - sometimes with **ankle dystonia**
  - **inability** to turn quickly



- As the **disease progresses**, gait initiation becomes a problem, the steps become shorter and more uncertain, and there is **festination**
- **Fear of falling** further contributes to a progressively hesitant gait

## ❑ Parkinsonian syndromes

- Progressive supranuclear palsy
- Multisystem atrophy
- Olivopontocerebellar degeneration (sporadic form)
- Striatonigral degeneration
- Shy-Drager syndrome
- Diffuse Lewy body disease
- Corticobasal degeneration
- Drug-induced parkinsonism
- Dopa responsive dystonia

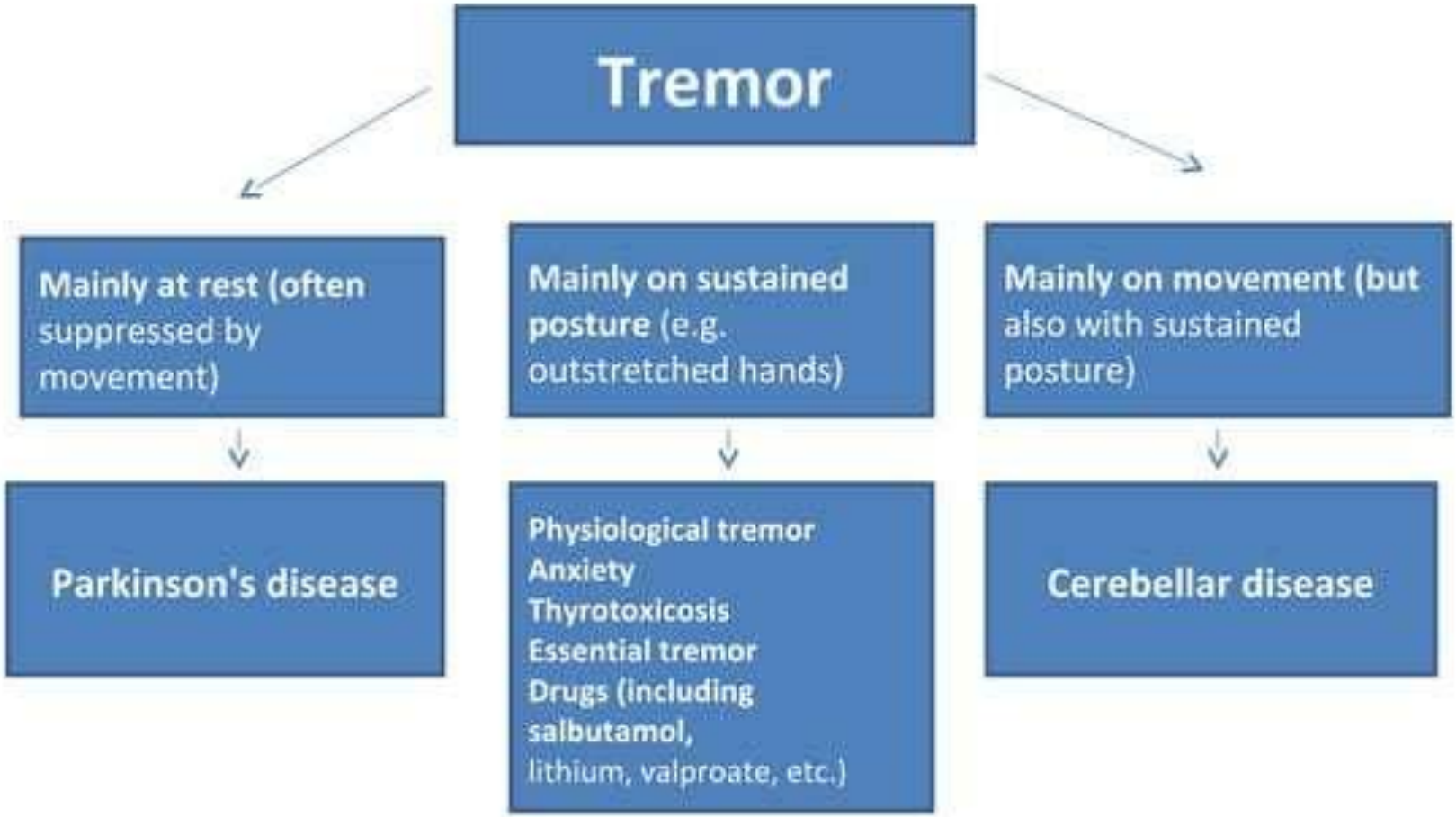
# Tremor

- **Tremor**: series of involuntary, relatively rhythmic, purposeless, oscillatory movements due to intermittent muscle contractions

- **Simple tremor** involves only a single muscle group
- **Compound tremor** involves several muscle groups
  - several elements in combination
  - resulting in a series of complex movements

- may be **unilateral or bilateral**

- most commonly involves **distal parts** of the extremities
  - fingers or hands
- may also affect the arms, feet, legs, tongue, eyelids, jaw, and head
- may occasionally involve the entire body


Tremor
Mainly at rest (often suppressed by movement)
Mainly on sustained posture (e.g. outstretched hands)
Mainly on movement (but also with sustained posture)
Parkinson's disease
Physiological tremor
Anxiety
Thyrotoxicosis
Essential tremor
Drugs (including salbutamol, lithium, valproate, etc.)
Cerebellar disease

# Tremor

- **Rate** may be slow, medium, or fast
- **Slow**: Oscillations of 3 to 5 Hz
- **Rapid**: Oscillations of 10 to 20 Hz

- **Amplitude** may be fine, coarse, or medium

- The **relationship to rest or activity** is the basis for classification into two primary tremor types:
  - resting
  - action

- **RESTING (static)** tremors are present mainly during relaxation (e.g., with the hands in the lap)
- **Attenuate when the part is used**
- **Rest tremor is seen primarily in PD** and other parkinsonian syndromes

# Tremor cont...

- **ACTION TREMORS** are divided into subtypes:
  - postural
  - kinetic
  - task-specific

- Postural:
  - ✓ **Postural tremors** become evident when the limbs are maintained in an antigravity position (e.g., arms outstretched)
  - ✓ Types of postural tremor:
    - enhanced physiologic tremor
    - essential tremor (ET)

- **Kinetic tremor:**

✓ Appears when making a voluntary movement

✓ May occur at the beginning, during or at the end of the movement

✓ Example : **intention (terminal) tremor**

✓ Intention tremor is a form of action tremor seen primarily in cerebellar disease.

- **Intention tremor**: appears when **precision** is required to touch a target, as in **finger-nose-finger** or **toe-to-finger test**

- Progressively worsens during the movement

- Approaching the target causes
  - the limb to shake,
  - side-to-side perpendicular to the line of travel,
  - amplitude of the oscillation increases toward the end

# Tremor cont...

- Tremors are **accentuated** by emotional excitement

- Many normal individuals develop tremor with anxiety, apprehension, and fatigue

- **Physiologic tremor:** is present in normal individuals
- frequency varies from 8 to 12 Hz
- Can occur normal persons by anxiety, fright, fatigue (rock climber's tremor)
- In conditions with **increased adrenergic activity**

# Essential Tremor

- Often **familial**
- ET may be a form of **enhanced physiological tremor**
- **Prevalence** of ET increases with age
- First appear anywhere between the second and sixth decades of life
- Tends to be **slowly progressive**

- ET is a **postural and action tremor**, that tends to affect the hands, head, and voice
- Made worse by anxiety
- **Senile tremor** is ET occurring during senescence with a negative family history

# Chorea

- **Chorea (Gr. "dance")** is characterized by involuntary, irregular, purposeless, random, non-rhythmic hyperkinesias
- movements are **spontaneous, abrupt, brief, rapid, jerky, and unsustained**
- movements are actually **random and aimless**
- They are present at rest but **increased** by activity, tension, emotional stress and self-consciousness
- patient may be able to temporarily and partially **suppress** the movements
- **disappear in sleep**

# Causes of chorea

**Drugs:**

- levodopa in Parkinson's patients
- oral contraceptive pill
- many psychiatric drugs

**Vascular disease of the basal ganglia:**

- atheroma
- systemic lupus erythematosus

**Degenerative diseases:**

- Huntington's disease

**Post-infectious:**

- Sydenham's chorea

**Other causes:**

- Thyrotoxicosis

# Distribution and characteristics

- **Distribution** is variable
- May **involve one extremity, one half of the body (hemichorea), or be generalized**
- Occur most characteristically in the **distal parts** of the upper extremities
- **May also involve** the proximal parts, lower extremities, trunk, face, tongue, lips, and pharynx
- May **be repeated twitching and grimacing** movements of the face that change constantly in character and location
- Involvement of the **vocal cord** may cause abnormal vocalizations, difficulty in maintaining phonation, or aphonia

# Chorea cont...

- They **interfere** with and **distort** voluntary movements, and the latter may be short, jerky, and unsustained
- When asked to hold the hands outstretched, there may be constant random movements of individual fingers **(piano-playing movements)**
- If the patient holds the examiner's finger in her fist, there are constant twitches of individual fingers **(milkmaid grip)**
- **Blink rate is increased**
- There is **hypotonia** of the skeletal muscles, with decreased resistance to passive movement

Milk maid grip of chorea

Piano playing movements of chorea

# Athetosis

- Athetosis means **"without fixed position"**
- Involuntary, irregular, coarse, somewhat rhythmic, and **writhing or squirming in character (twisting)**
- Hyperkinesias are **slower, more sustained, and larger in amplitude** than those in chorea
- May involve the **extremities, face, neck, and trunk**
- In the extremities they affect mainly the **distal portions, the fingers, hands, and toes**
- movements are characterized by any **combination** of flexion, extension, abduction, pronation, and supination, **often alternating and in varying degrees**

# Athetosis cont...

- They **flow randomly** from one body part to another, and the direction of movement changes randomly
- **Affected limbs are in constant motion** (**athetosis means "without fixed position"**)
- ***Hyperextension of the fingers and wrist and pronation of the forearm may alternate with full flexion of the fingers and wrist and supination of the forearm***
- **Disappear in sleep**
- Voluntary movements are impaired, and coordinated action may be difficult or impossible

# Causes

- May be **congenital** due to perinatal injury to the basal ganglia
- May be present in association with other neurological deficits (**athetotic cerebral palsy**)
- It may be either unilateral or bilateral
- The predominant pathologic changes are in the **caudate and putamen**, there may also be cortical involvement
- Many patients have features of athetosis plus chorea
- **Choreoathetosis** refers to movements that lie between chorea and athetosis in rate and rhythmicity, and may represent a transitional form

# Ballism

- Greek word *ballismos,* ***which means a jumping movement***

- Latin *ballista, which refers to an ancient* military machine, similar to a catapult, used for throwing large stones

# Hemiballismus

- Dramatic neurologic syndrome of **wild, flinging (forceful), incessant (uninterrupted or continuous)** movements that occur on one side of the body
- Due to infarction or haemorrhage in the region of the **contralateral subthalamic nucleus**
- Results in **disinhibition** of the motor thalamus and the cortex, resulting in contralateral hyperkinetic movements

# Distribution and characteristics

- Movements are involuntary and purposeless movements
- More rapid and forceful
- Involve the proximal portions of the extremities

- When fully developed, there are continuous, violent, swinging, flinging, rolling, throwing, flailing(thrashing) movements of the involved extremities

- They are usually unilateral, and involve one entire half of the body

- Rarely, they are bilateral (biballismus or paraballismus) or involve a single extremity (monoballismus)

- movements may spare the face and trunk
- Hemiballismus is difficult to treat, incredibly disabling, and sometimes fatal because of exhaustion and inanition

| Chorea | Ballismus | Athetosis |
| --- | --- | --- |
| Rapid | Rapid | Slow |
| Involuntary | Involuntary | Involuntary |
| Non-stereotypical | Non-stereotypical | Non-stereotypical |
| Semi-purposeful / non-purposeful | Non-purposeful | Non-purposeful |
| Dance-like | violent flinging movement | writing |
| More on distal | More on proximal | Has propensity affecting upper limb |

# Myoclonus

- ***Single or repetitive, abrupt, brief, rapid, lightning-like, jerky, arrhythmic, asynergic, involuntary contractions involving portions of muscles, entire muscles, or groups of muscles***

- **Movements are quicker than chorea**

- Seen principally in the **muscles of the extremities and trunk**, but the involvement is often multifocal, diffuse, or widespread

- May involve the facial muscles, jaws, tongue, pharynx, and larynx

- May **be successive or simultaneous involvement** of many muscles

- Myoclonus may appear **symmetrically** on both sides
- Such synchrony may be an attribute unique to myoclonus

# Classification

- **Myoclonus** has been classified in numerous ways, including the following:
  - **positive versus negative;**
  - **epileptic versus nonepileptic;**
  - **stimulus sensitive (reflex) versus spontaneous;**
  - **rhythmic versus arrhythmic;**
  - **anatomically (peripheral, spinal, segmental, brainstem, or cortical)**
  - **by etiology (physiologic, essential, epileptic, and symptomatic)**

- **Asterixis** may be viewed as negative myoclonus, the transient, unwanted, abnormal relaxation of a muscle group

- As typically used, the term myoclonus refers to **positive myoclonus**: abnormal jerks

- **Physiologic myoclonus** occurs normally
- **Hypnic jerks** are myoclonic jerks that appear during the process of falling asleep, but disappear during sleep
- **Hiccups** are another form of physiologic myoclonus
- **Essential myoclonus** there are no accompanying abnormalities
- may be sporadic or familial (hereditary essential myoclonus, paramyoclonus multiplex)

- **Myoclonic epilepsy**: occasional random myoclonic jerks of the axial or proximal limb musculature, which may appear or increase in frequency immediately prior to a seizure

- **Juvenile myoclonic epilepsy** (JME, Janz' syndrome) have generalized tonic-clonic seizures that are associated with frequent myoclonic jerks predominantly affecting the arms, especially on awakening

- The condition is familial, with both dominant and recessive forms, and is relatively benign

- **Myoclonus occurs without prominent seizures** in a number of other conditions, including
  - ✓ *Metabolic disorders (especially uremic and anoxic encephalopathy)*
  - ✓ *Subacute sclerosing panencephalitis*
  - ✓ *Hallervorden-Spatz syndrome*
  - ✓ *Creutzfeldt-Jakob disease*
  - ✓ *Alzheimer's disease*
  - ✓ *Wilson's disease*
  - ✓ *Huntington's disease*
  - ✓ *Corticobasal degeneration*
  - ✓ *Viral encephalitis*
  - ✓ *General paresis*
  - ✓ *Hashimoto's encephalopathy*
  - ✓ *Lipidoses*


Thank you