

# Movement disorders in clinical practice 2

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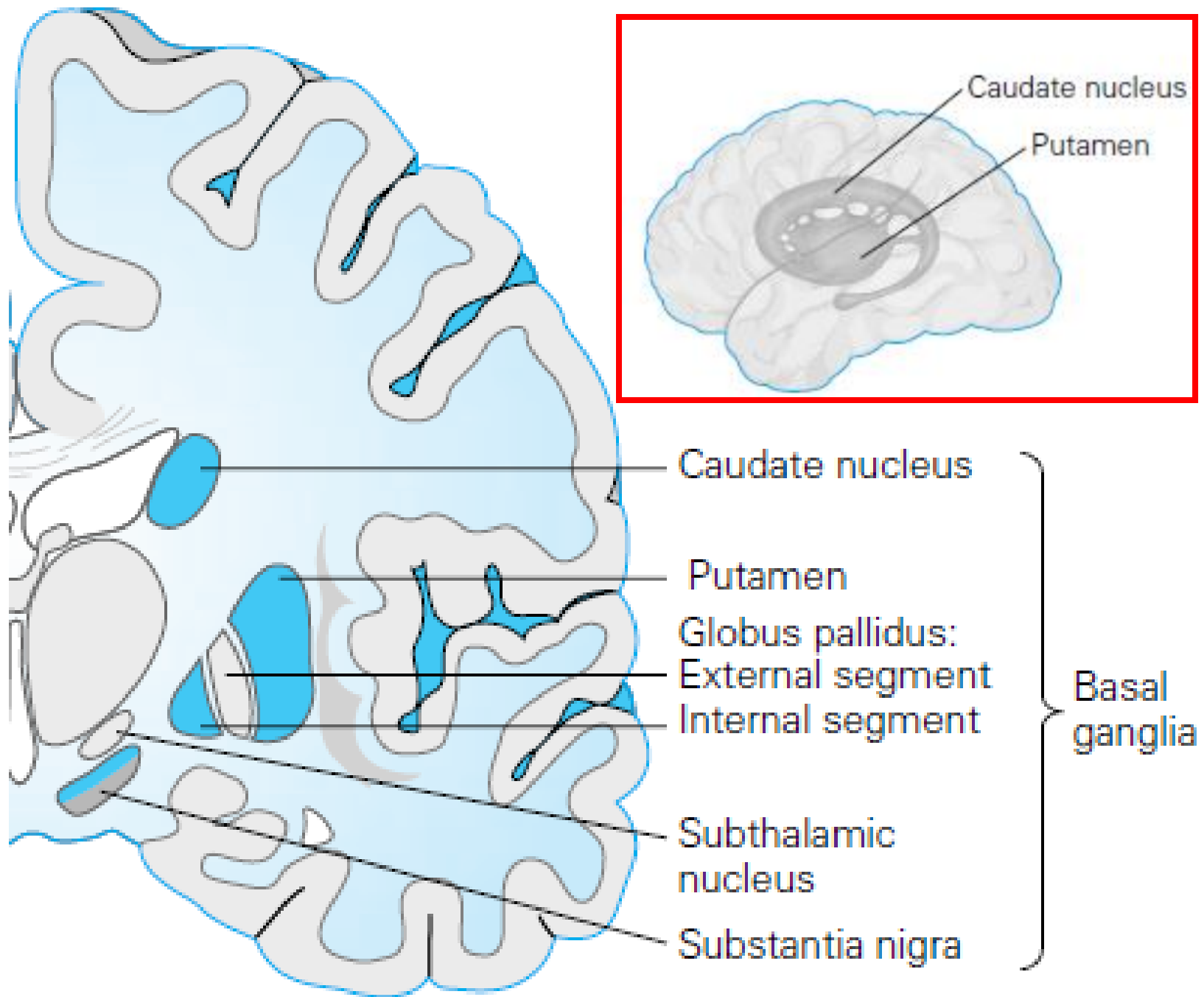
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- ❑ Chorea
- ❑ Ballismus
- ❑ Athetosis
- ❑ Tics
- ❑ Parkinsonian tremor
- ❑ Myoclonus



Basal ganglia

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graph TD; BG[Basal ganglia] --> CS[Corpus striatum]; BG --> AM[Amygdala]; CS --> NS[Neostriatum (Striatum)]; CS --> PS[Paleostriatum]; NS --> CA[Caudate]; NS --> PU[Putamen]; PU --> LN[Lentiform nucleus]; PS --> GP[Globus pallidus];
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Corpus striatum

Amygdala

Neostriatum (Striatum)

Paleostriatum

Caudate

Putamen

Globus pallidus

Lentiform nucleus

# Chorea

- ❑ Greek word meaning “dance”
- ❑ Involuntary arrhythmic movements of a forcible, rapid, discrete and jerky type
  - Chorea may be limited to one side of the body (*hemichorea*)



# Causes of chorea

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## **Inherited disorders**

- Huntington disease
- Benign hereditary chorea
- Neuroacanthocytosis
- Dentatorubropallidoluisian atrophy
- Wilson disease

## **Rheumatic chorea**

- Sydenham, chorea gravidarum

## **Drug-induced chorea**

- Neuroleptics (phenothiazines, haloperidol, and others)
- Oral contraceptives
- Phenytoin (occasionally other anticonvulsants)
- Excess dosages of L-dopa and dopamine agonist medications
- Cocaine

# Causes of chorea

## **Chorea symptomatic of systemic disease**

Lupus erythematosus with antiphospholipid antibodies

Thyrotoxicosis

Polycythemia vera

Hyperosmolar, nonketotic hyperglycemia

AIDS

Paraneoplastic, often with other movements

## **Hemichorea, rarely associated with**

Stroke

Tumor

Vascular malformation



# Clinical tests in chorea

MR brain imaging

Full blood count  
Red cell mass

Blood film

Serum caeruloplasmin  
Urinary copper level  
Liver biopsy

Pregnancy test

- Stroke or other focal basal ganglia pathology
- T2\* imaging abnormal in iron accumulation disorders
- FLAIR imaging abnormal in prion disease
- Deep brain T2 hyperintensities in Wilson's disease
- Haematocrit raised in polycythamia
- Sensitive test for diagnosis of polycythaemia rubra vera
- Multiple films necessary to exclude neuroacanthocytosis
- Reduced in Wilson's disease
- Raised in Wilson's disease
- Sometimes required to diagnose Wilson's disease
- Chorea gravidarum in first trimester

HIV test

Erythrocyte sedimentation rate and antinuclear antibody

Anti-dsDNA

Anticardiolipin antibodies and lupus anticoagulant

Thyroid function tests

Anti-streptolysin O titre

Antibasal ganglia antibodies

Muscle biopsy

Cerebrospinal fluid analysis

- Chorea may be presenting feature of AIDS
- Sensitive tests to reveal systemic lupus erythematosus
- Relatively specific to systemic lupus erythematosus
- Antiphospholipid syndrome is a cause of chorea
- Thyrotoxicosis
- Recent streptococcal infection suggests Sydenham's chorea/PANDAS
- Associated with post-infective chorea, chorea gravidarum and oral contraceptive-induced chorea
- Ragged red fibres or respiratory chain abnormalities in mitochondrial disease
- May reveal inflammatory/neoplastic causes

# Pharmacological management of chorea

- ❑ Treatment of underlying cause(s), behavioral and psychiatric features
  - more important than symptomatic management
- ❑ For functionally disabling chorea
  - Sulpiride, **olanzapine** and **risperidone** at low doses
  - Tetrabenazine is a good option, but may cause sedation and depression

# Ballismus

- ❑ An uncontrollable, poorly patterned flinging movement of an entire limb
- ❑ Ballistic movements are usually unilateral, involve proximal limb muscles and are of wide range and flinging in nature, the condition is called *hemiballismus*





# Athetosis

- ❑ Greek word meaning “unfixed” or “changeable”
- ❑ Characterized by an inability to sustain the fingers and toes, tongue, or any other part of the body in one position
  - The maintained posture is interrupted by relatively *slow, sinuous*, purposeless movements that have a tendency to *flow into one another*







- The movements appear to be slower than those of chorea
  - In some cases, it is impossible to distinguish between them, hence the term *choreoathetosis*
  - Chorea + slower writhing movement that can be part of dystonia, hence the term *choreodystonic*

# Tics

- ❑ Repeated, individually recognizable, intermittent movements or movement fragments that are almost always *briefly suppressible* and are usually associated with awareness of an urge to perform the movement

# Tics

- ❑ Motor tics
  - sudden, brief intermittent *movements*
- ❑ Vocal or phonic tics
  - sudden, brief intermittent *utterances*



## Motor

## Vocal

Simple

Eye blinking,  
eyebrow/forehead/  
shoulder raising,  
winking, nose  
wrinkling

Grunting, sniffing,  
coughing,  
humming, whistling

Complex

Head shaking,  
touching self or  
others, kicking,  
jumping, hopping,  
copropraxia,  
echopraxia

Saying words or  
parts of words,  
coprolalia,  
echolalia, palilalia

# Gilles de la Tourette syndrome

- Begins in children and teenagers with:
  - Multiple *motor tics*, with a gradually evolving repertoire of movements
  - *Phonic tics*, commonly sniffs and grunts, rarely repetitive speech (echolalia) or swearing (coprolalia)

# Gilles de la Tourette syndrome

- Comorbidities
  - Obsessive–compulsive disorder (OCD), such as repeated checking or complex rituals
  - Attention-deficit hyperactive disorder (ADHD)

- Two classes of drugs are used in treatment
  - Alpha 2-adrenergic agonists clonidine and guanfacine (first choice because of better side-effect profile in spite of lesser potency)
  - Neuroleptics: *haloperidol*, pimozide, sulpiride, and tiapride

- ❑ OCD (which is often more disabling) may improve with SSRIs (like *fluoxetine*) or behavioural therapy
- ❑ ADHD may improve with methylphenidate



# Tremor

Mainly at rest (often suppressed by movement)

**Parkinson's disease**

Mainly on sustained posture (e.g. outstretched hands)

Physiological tremor  
Anxiety  
Thyrotoxicosis  
Essential tremor  
Drugs (including salbutamol, lithium, valproate, etc.)

Mainly on movement (but also with sustained posture)

**Cerebellar disease**

# Parkinsonian Tremor



## Parkinsonian Tremor

## Essential Tremor

High amplitude, slower frequency

Low amplitude, faster frequency

Mostly seen at rest

Mostly seen during action

Bradykinesia (B), rigidity (R) & postural instability (P)

B, R & P not commonly seen

Family history (<10%)

Family history (>50%)

Re-emergent / postural tremor observable after mean latency of 5 sec

Postural tremor immediately observable

## Parkinsonian Tremor

## Essential Tremor

Starts unilaterally & progresses to the other side; usually remains asymmetrical

Starts bilaterally; symmetrical

No effect from alcohol

Alcohol often improves tremor

Voice and head almost never affected

Head and voice affected

Worsens with emotional stress

Worsens with emotional stress







# Myoclonus

- ❑ Sudden, *shock-like jerks* of a group of muscles which are irregular in rhythm and amplitude
- ❑ Can be due to dysfunction in the cerebral cortex, basal ganglia, brainstem or spinal cord, nerve roots and plexuses

# Causes of myoclonus

- ❑ Myoclonus as part of general medicine:
  - Hepatic encephalopathy ('liver flap')
  - Renal failure
  - Carbon dioxide retention



- ❑ Myoclonus as part of degenerations of the cerebral cortex:
  - Alzheimer's disease
  - Lewy body dementia
  - Creutzfeldt–Jakob disease

## □ Myoclonus as part of epilepsy:

- Juvenile myoclonic epilepsy (jerks in the morning: 'messy breakfast syndrome')
- Severe infantile epilepsies

- ❑ Myoclonus due to basal ganglia disease:
  - Jerking on attempted movement ('action myoclonus') after anoxia due to cardiorespiratory arrest or carbon monoxide poisoning

- ❑ Myoclonus due to brainstem disease:
  - Exaggerated jerks in response to sudden noise ('startle myoclonus') in rare metabolic and degenerative disorders

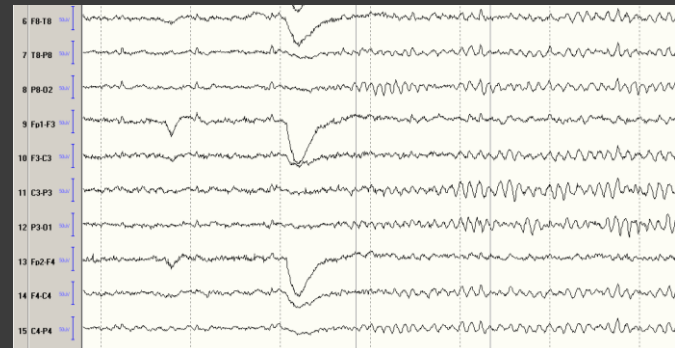
# Cortical

- ❑ Distal
- ❑ Small amplitude
- ❑ Focal
- ❑ Touch sensitive

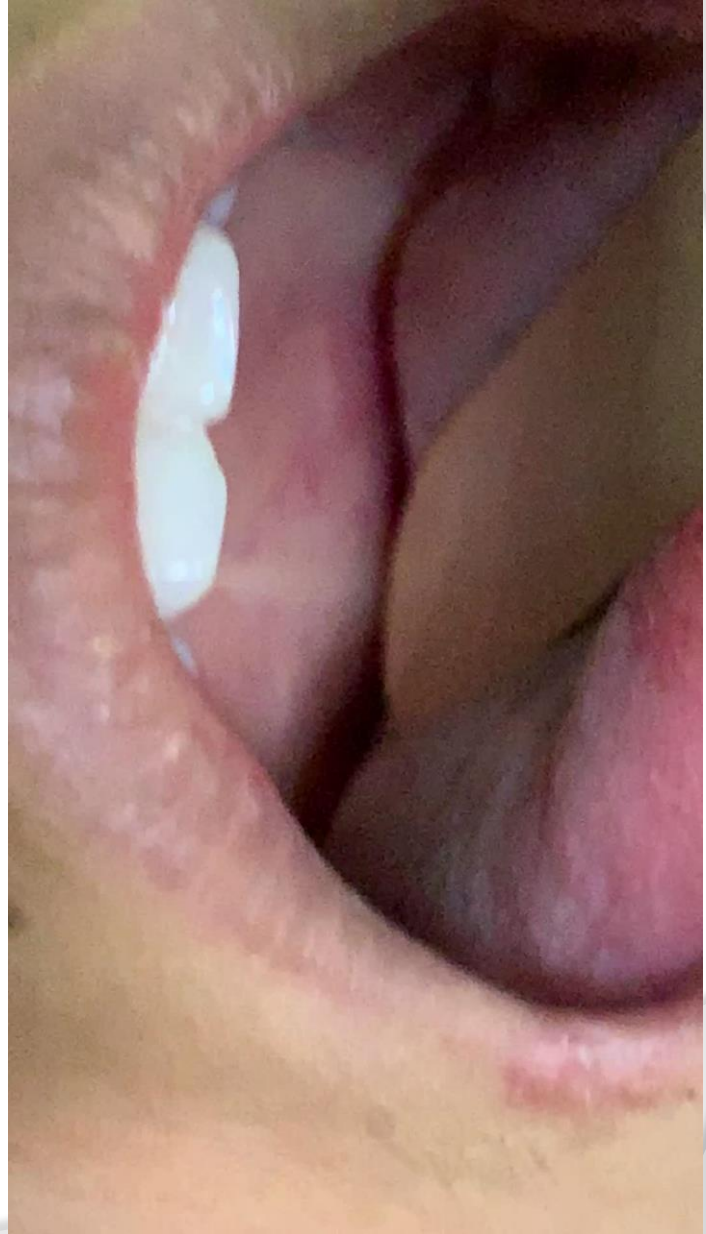


# Subcortical

- ❑ Proximal
- ❑ Large amplitude
- ❑ Generalized
- ❑ Sound sensitive







# Palatal myoclonus

- ❑ A rare movement disorder consisting of brief and rhythmic movements of the soft palate
- ❑ Described in 1886 by Spencer



# Palatal myoclonus or tremor

- ❑ Less 'shock-like'
- ❑ More regular and continuous

# Palatal tremor

- Two different forms of palatal tremor
  - Essential palatal tremor (EPT)
  - Symptomatic palatal tremor (SPT)

# Palatal tremor

- Essential palatal tremor (EPT)
  - Palatal tremor and ear clicks are the sole clinical manifestations

# Palatal tremor

- ❑ Symptomatic palatal tremor (SPT)
  - Usually no ear clicks
  - Tremor may involve facial, ocular, and extremity muscles
  - Olivary pseudohypertrophy m/b seen

# Effect on swallowing

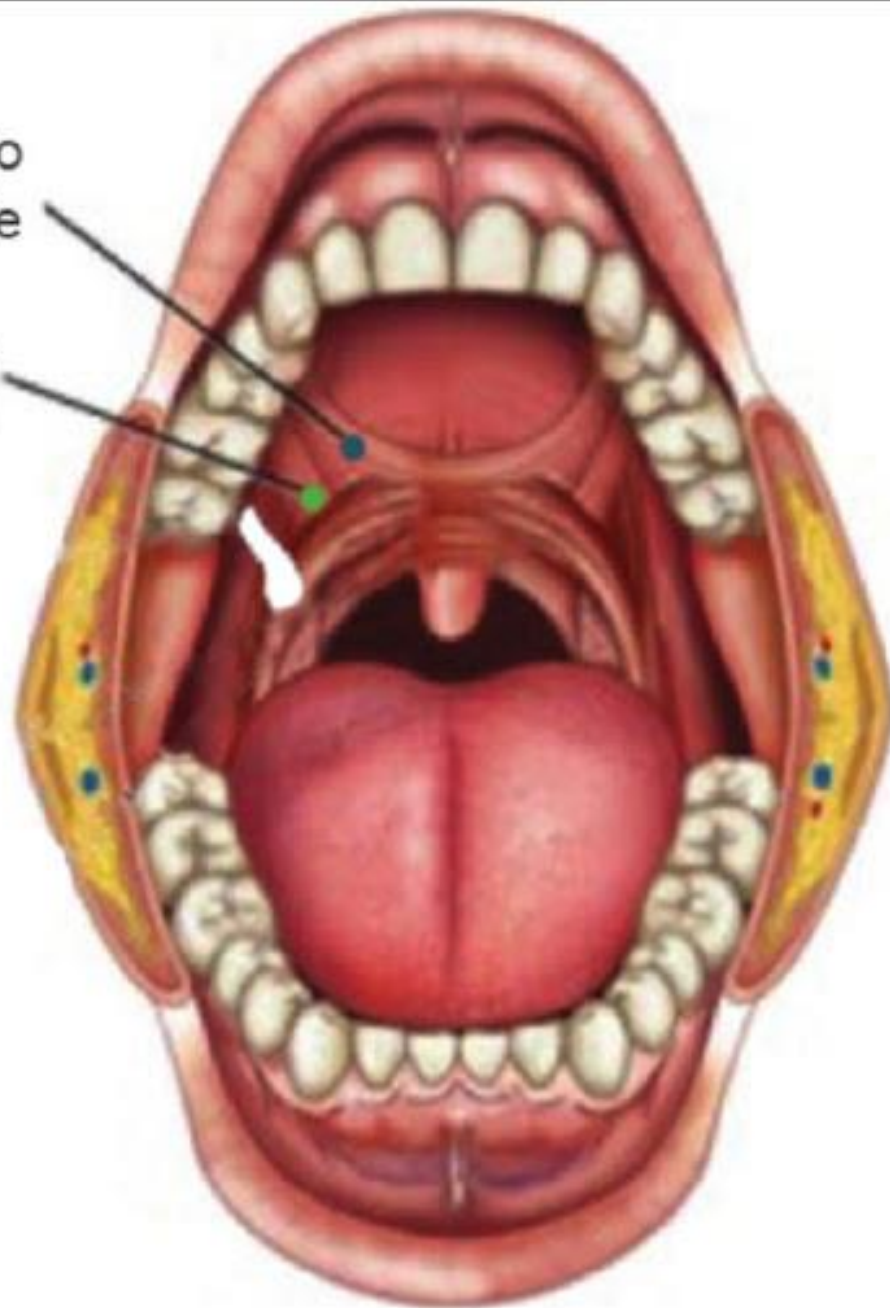
- ❑ Surprisingly little effect on swallowing

# Treatment

- ❑ Sodium valproate
- ❑ Clonazepam
- ❑ Levetiracetam/piracetam
- ❑ Carbamazepine
- ❑ Trihexyphenidyl
- ❑ Botulinum toxin

Injection into  
TVP muscle

Injection into  
LVP muscle



Thank you!