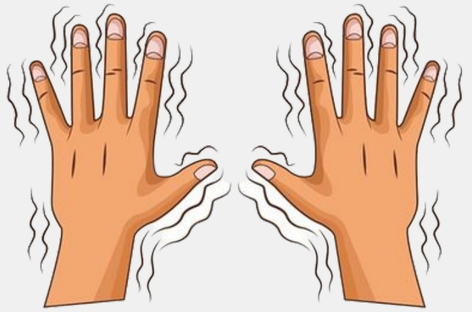
NEUOROLOGY
MOVEMENT DISORDERES

DONE BY: GROUP C3

INTRODUCTION

Ibrahem Alaqrbawy

Introduction:



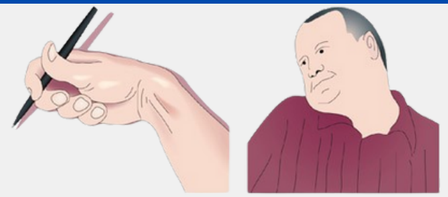
- Tremor involuntary oscillatory movement of a body part (arm, leg, head, jaw, lips, palate)

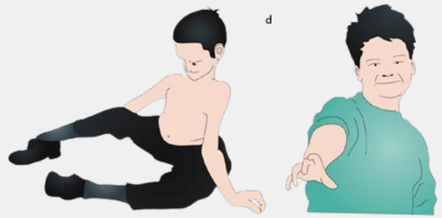


- Chorea is an irregular, twisting or jerky movement of a group of muscles.

- Ballism is a brief flinging movement of a limb, most often unilateral (hemiballismus).

Introduction：





- **Dystonia** is a sustained contraction of agonist and antagonist muscles producing twisting movements or abnormal postures.



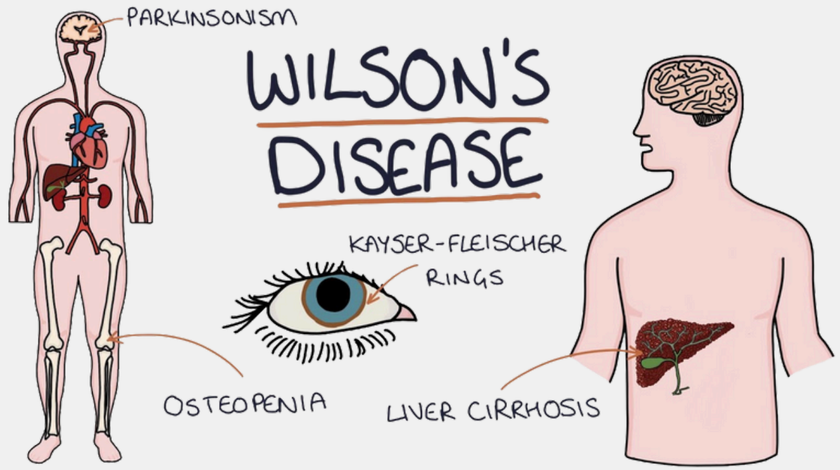
- **Myoclonus** is a sudden jerking movement of a muscle that is sufficient to move a joint.



- **Tics** are abnormal, brief muscle contractions that may involve the face, extremities,

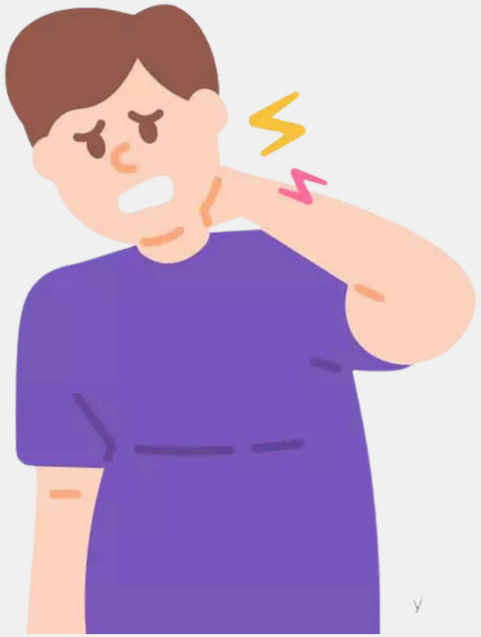
Introduction：

- **Wilson Disease** ：impairs copper transport, leading to excessive copper accumulation whchi result in various Manifestations (liver, nuro , psychic)

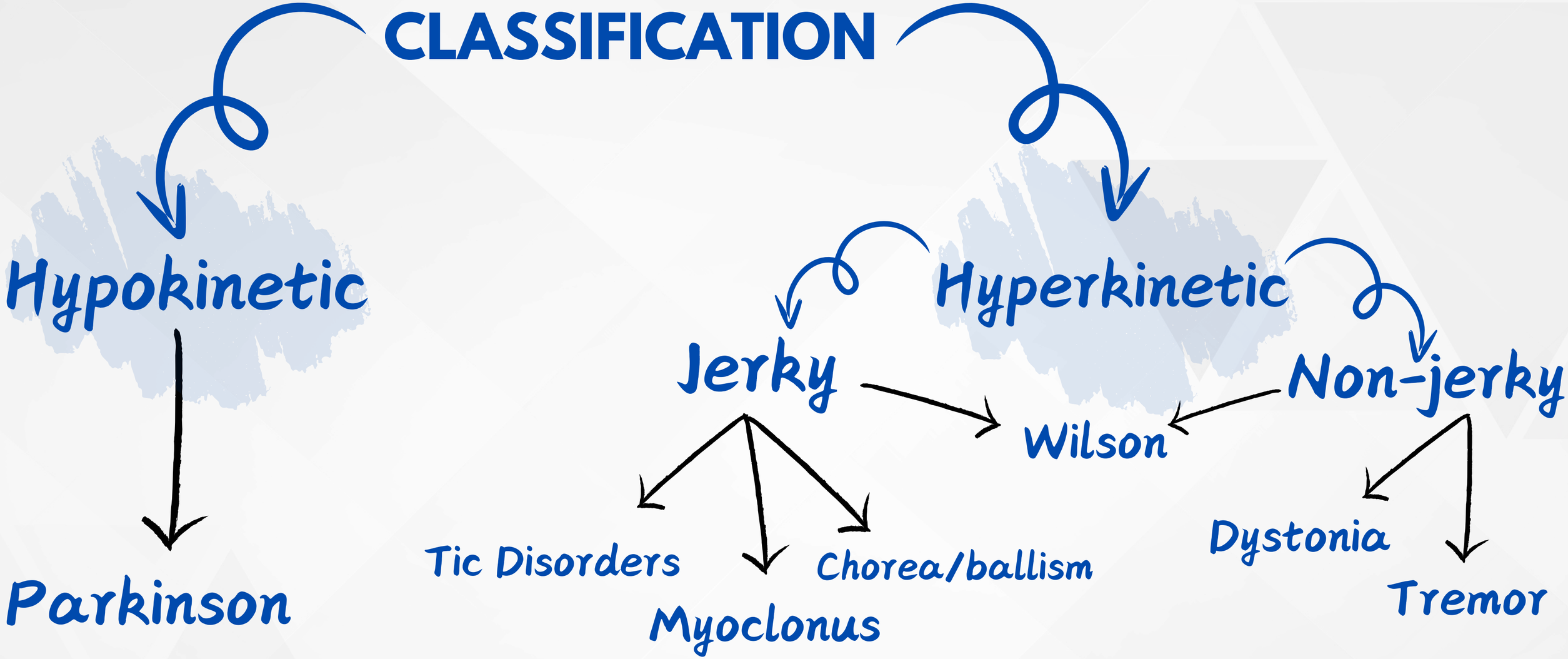


- **Stiff-person Syndrome**：

muscle rigidity and spasms. Symptoms typically start with axial and trunk



CLASSIFICATION



DRUG INDUCED MOVMENT DISORDERS

Ibrahem Alaqrbawy



Drug induced movment disorders



Medications which block dopamine receptors including antipsychotic medications (both traditional dopamine blockers and newer, atypical agents) and the promotility agent metoclopramide may produce a variety of movement disorders:

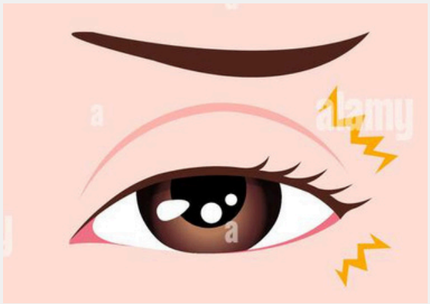
1- Acute dystonic reactions occur when patients are introduced to dopamine-blocking medications or given high doses of dopamine blockers to which they are not accustomed.

Intermittent or sustained contraction in any of the muscles in the face, limbs, or trunk may occur. Forced contraction of the extraocular muscles and tonic deviation of the eyes may occur.



Acute dystonic reactions are best treated with anticholinergic agents and benzodiazepines. The reaction is short-lived and does not produce any long-term consequences





2- Parkinsonism may occur as the result of long-term use of any neuroleptic agent.
The symptoms are similar to those seen in PD, but tremor is less common and patients tend to be less responsive to levodopa.

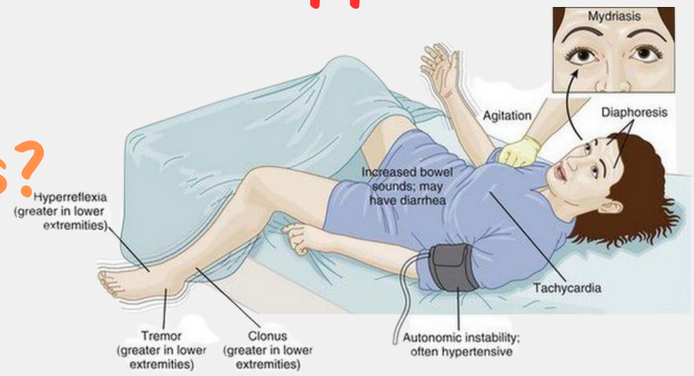
When does it happen ?
Difference about PD ?

3- Neuroleptic malignant syndrome : occurs when patients are exposed to high doses of dopamine-blocking medications or when levodopa or dopamine agonists are withdrawn abruptly.

When does it happen ?

The syndrome includes fever, autonomic instability, encephalopathy, and muscular rigidity.

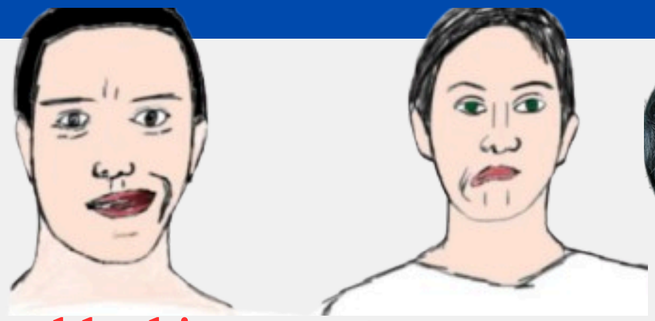
Symptoms?



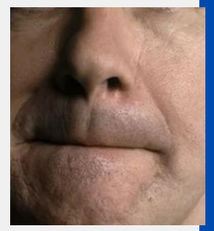
The offending agent must be stopped, but a combination of bromocriptine, dantrolene, and benzodiazepines is usually required to control the muscle rigidity.

Dealing with rigidity?

Tardive dyskinesia (TD)







4- Tardive dyskinesia (TD)

is a disorder that occurs after chronic exposure to dopamine-blocking agents.



Commonly observed movements include

chewing, lip smacking , grimacing and tongue thrusting.

The limbs, trunk, and even diaphragm may be affected.

Treatment

Treatment of TD is challenging:

Withdrawal of the offending agent often makes the movements worse.

The dopamine-depleting agent tetrabenazine may be helpful



So , what is the most common drug that induce movement disorders? Dopamine blocking agent

MYOCLONUS

Ibrahem Alaqrbawy



Myoclonus

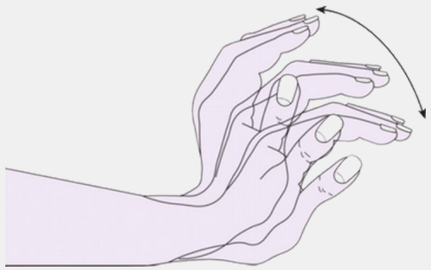


- Myoclonus is a sudden jerking movement of a muscle that is sufficient to move a joint.

- Asterixis is a negative form of myoclonus in which a patient is suddenly, but briefly, unable to hold the arms and hands up against gravity, resulting in an erratic and repetitive downward jerking movement.



- Both are signs of central nervous system dysfunction.
- The etiologic categories of myoclonus are physiologic, essential, epileptic, and symptomatic (Box 16-3).



Myoclonus

Physiological

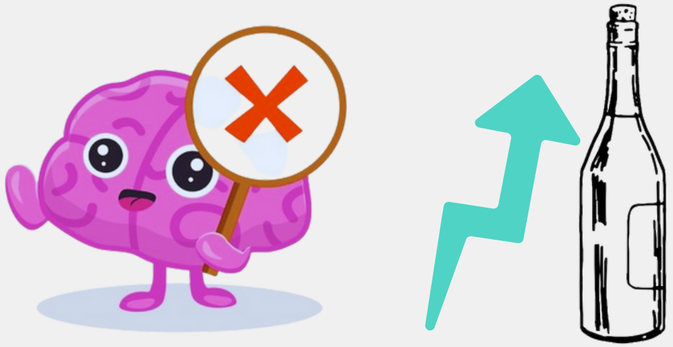
sleep starts/hypnic jerks/hiccups.



Essential

without other neurologic symptoms

responsive to alcohol



Epileptic

juvenile myoclonic epilepsy and others ?



Symptomatic myoclonus accompanies a wide variety of metabolic disturbances and neurodegenerative diseases.

TREATMENT

Clonazepam and valproate





Look at box 16-3 to know more about different causes of different types of myoclonus

KEY POINTS

- Myoclonus is a sudden, brief jerking movement of a muscle or group of muscles.
- Myoclonus has a wide variety of causes and may respond to medications such as clonazepam or valproate.

TREMOR

Bayan Al dabbas



Tremor

- Tremor is an involuntary oscillatory movement of a body part (arm, leg, head, jaw, lips, palate).
- It can be divided into :

1. **Resting** : occurring when the body part is at rest
2. **Postural** : occurring when maintaining a fixed posture
3. **Action** : occurring with movement

(Postural and action tremors usually accompany each other.)



- **Intention tremor** is applied when an action tremor worsens as the body part approaches its target.



- **Essential tremor** (ET) is the most common tremor and overall, the most common movement disorder. It can begin at any age and tends to get worse over time.

⋙ Common types and causes of tremors are summarized in the Box .

TABLE 16-1. Parkinsonian Syndromes

Parkinsonian Syndrome	Distinguishing Clinical Features
Progressive supranuclear palsy	Supranuclear ophthalmoplegia, with greatest limitation of downward gaze; axial rigidity; early falls due to rigidity, impaired postural reflexes, neck hyperextension, and inability to look down
Corticobasal ganglionic degeneration	Limb apraxia; cortical sensory impairment; alien-limb phenomenon; asymmetric rigidity; dementia
Diffuse Lewy body disease	Early dementia; prominent visual hallucinations; cognitive fluctuations; extreme sensitivity to extrapyramidal side effects of antidopaminergic neuroleptic drugs
Vascular parkinsonism	"Lower-half" parkinsonism in which rigidity in the legs is greater than in the arms, resulting in slow, shuffling gait
Multiple system atrophy	Early and prominent features of autonomic dysfunction (MSA-A); cerebellar dysfunction (MSA-C); parkinsonism refractory to levodopa (MSA-P); high-pitched, quivering dysarthria

- Because there is often a family history, the term "familial tremor" is sometimes applied.
- The tremor is a postural and action tremor, which involves the hands, head, and voice.

Treatement :

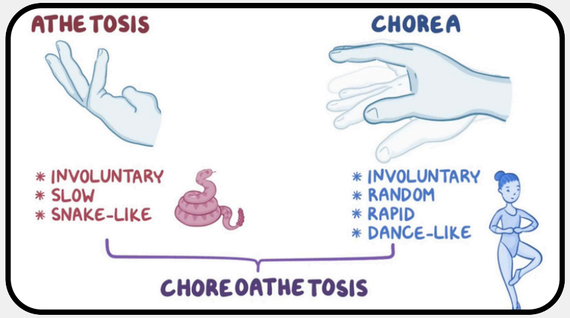
- It often improves with small quantities of alcohol, though this is not recommended as a treatment strategy.
- The most effective treatment options are propranolol and primidone.
- Deep brain stimulation of the ventral intermediate nucleus of the thalamus may help patients with disabling ET refractory to medications.

HUNTINGTON DISEASE AND OTHER CAUSES OF CHOREA

Reema Alshrouf



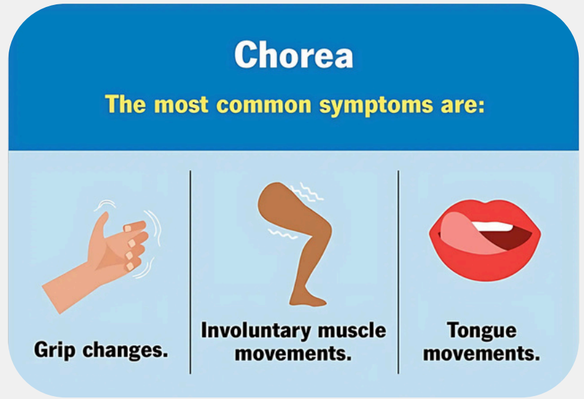
Chorea



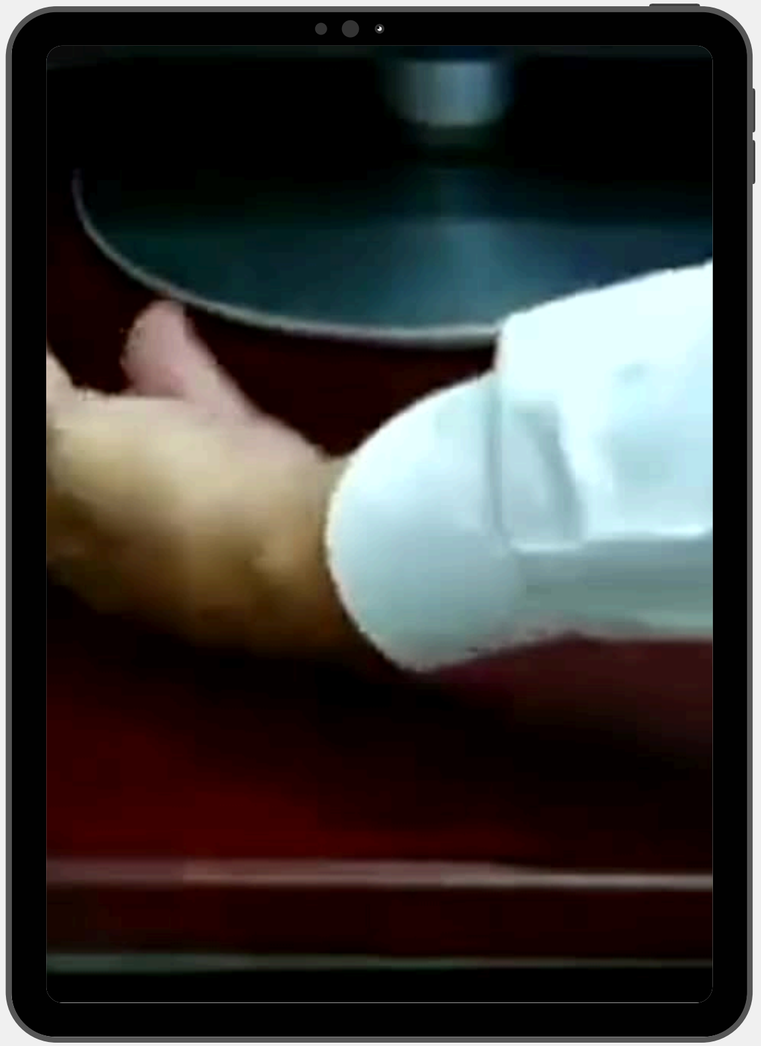
- Chorea is an irregular, twisting or jerky movement of a group of muscles.
- In most cases, chorea flows from one muscle group to an adjacent group in a random-appearing pattern.



- Chorea is usually accompanied by athetosis, a writhing movement of the limbs.
- Motor impersistence often occurs with chorea; two examples are the darting tongue and milkmaid handgrip, which are seen in patients with Huntington disease (HD).



athetosis



Chorea

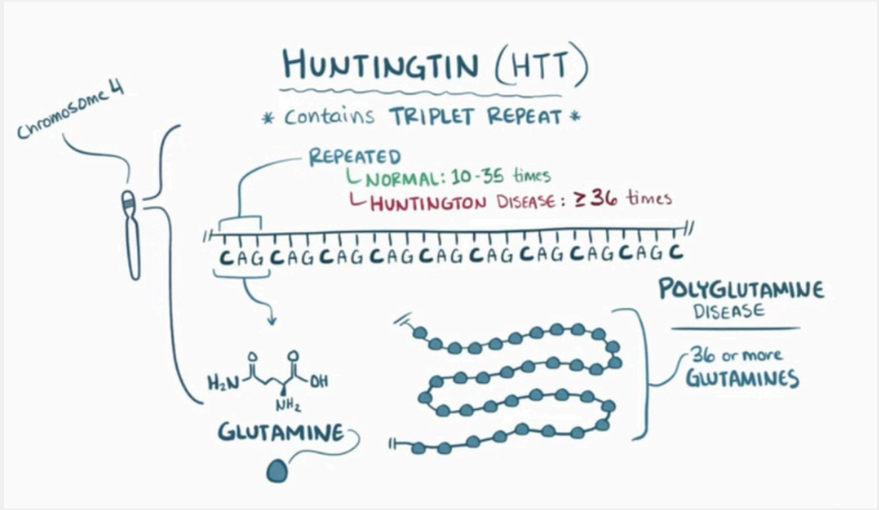


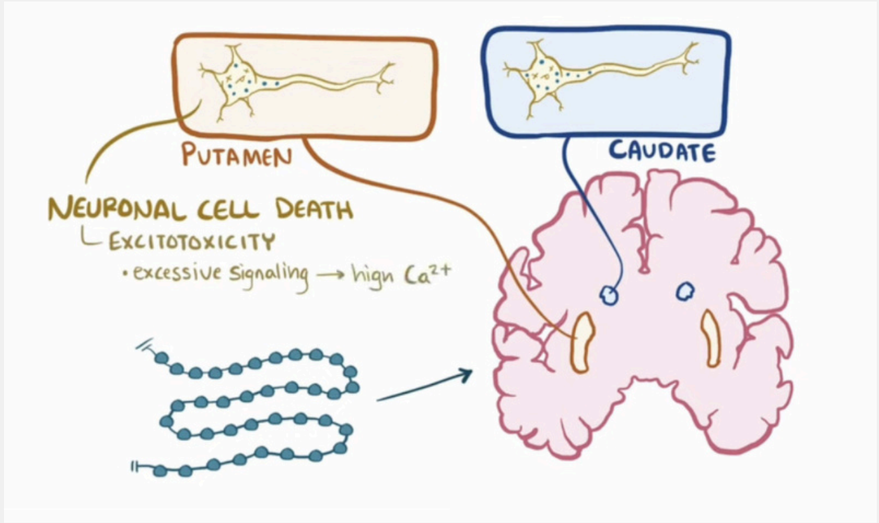
HUNTINGTON DISEASE

- **CLINICAL MANIFESTATIONS**

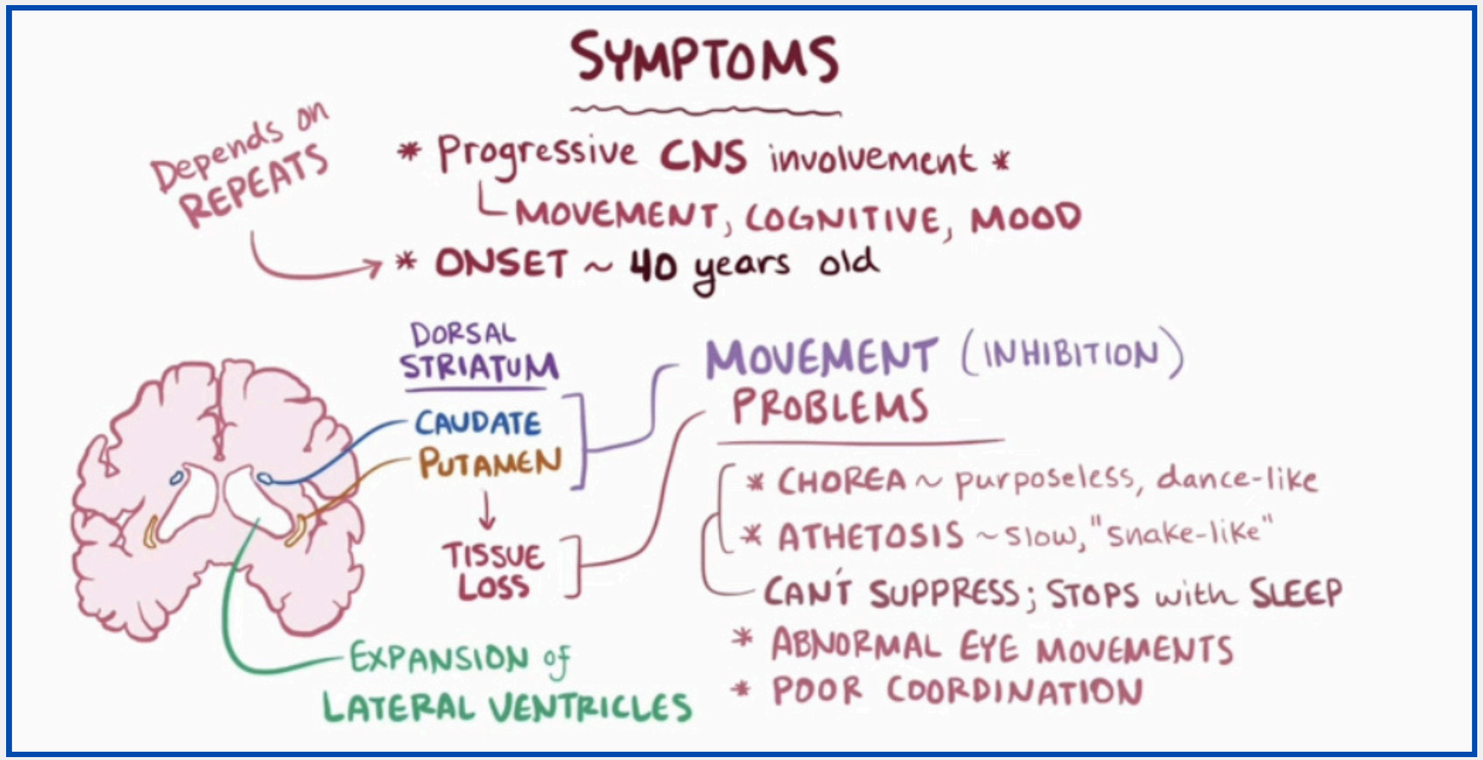
  - HD, an autosomal dominant neurodegenerative disorder is the most important and serious cause of chorea.

  - HD is characterized by chorea, cognitive impairment, dystonia, and psychiatric illness.



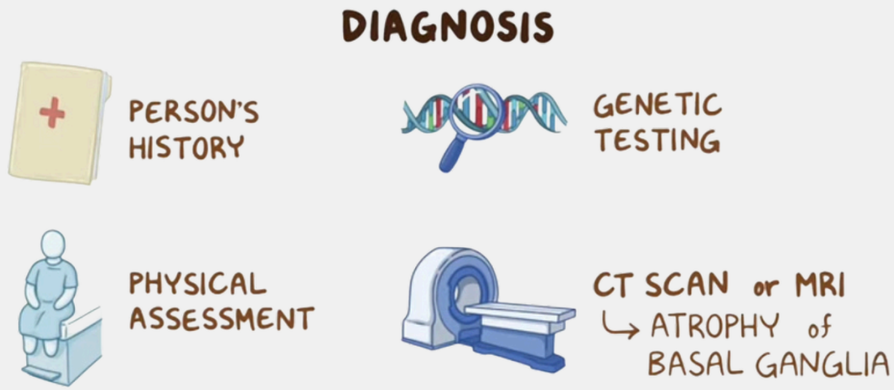


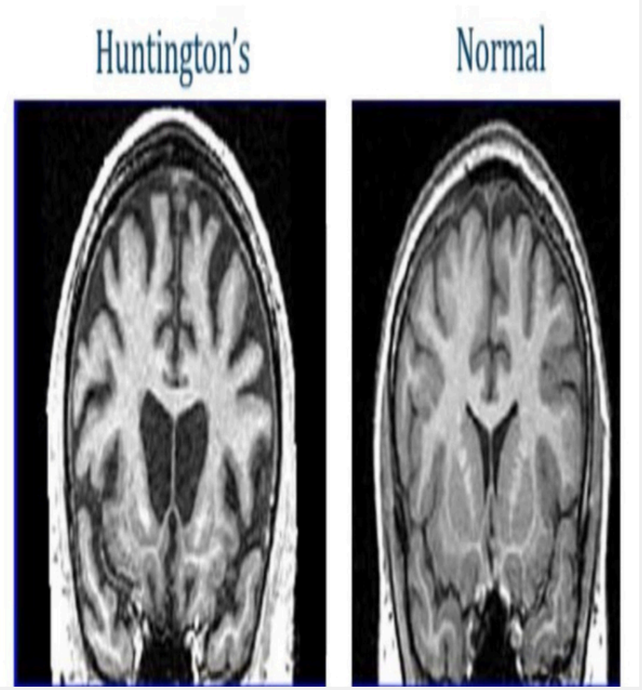
- Symptoms usually appear between the ages of 35 and 45 years and include the triad of chorea, behavioral changes or personality disorder (frequently obsessive-compulsive disorder), and dementia. The three may occur together at onset, or one may precede the others by years.



DIAGNOSTIC EVALUATION

- Diagnosis is by personal and family history, clinical signs, imaging, and genetic testing.
- Caudate atrophy, sometimes severe, is the characteristic finding on magnetic resonance imaging (MRI).
- Definitive diagnosis is made by finding an expansion of more than 40 cytosine–adenine–guanine trinucleotide repeats in the HTT gene on chromosome 4.





PATHOLOGY

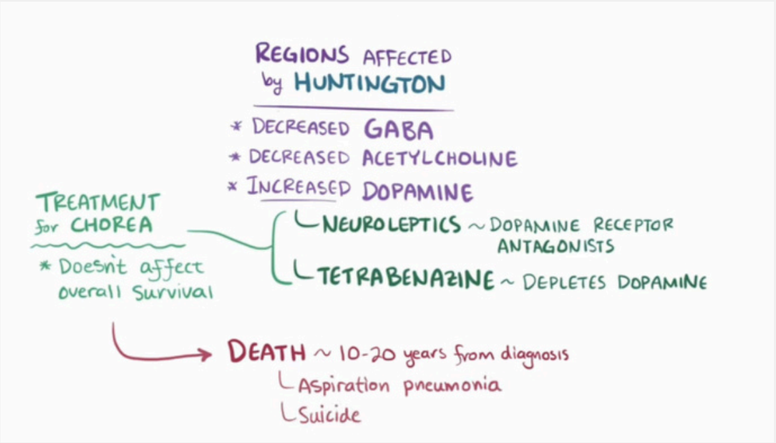
- Pathologic examination shows severe destruction of the caudate and putamen (striatal and nigral GABA-ergic neurons) and loss of neurons in the cerebral cortex (layer 3).

- The molecular mechanisms of HD are unclear but involve accumulation of abnormal intracellular proteins which triggers cell death.

TREATMENT

- Pharmacologic management of dementia and chorea often involves dopaminergic antagonists, including neuroleptic drugs, but it is far from adequate.

- Neuroleptics can ameliorate the chorea, but the other neuropsychiatric symptoms ultimately prove disabling. Unfortunately, HD is a progressive and ultimately fatal disorder; death occurs 10 to 20 years after onset.



Chorea has many causes

Hereditary

- Huntington disease (HD)
- HD-like syndromes
- Neuroacanthocytosis
- Dentatorubral pallidoluysian atrophy
- Wilson disease

Drugs

- Neuroleptics
- Antiparkinsonian medications

Toxins

- Alcohol
- Anoxia
- carbon monoxide

Metabolic

- Hyperthyroidism
- Nonketotic hyperglycemia
- Hepatocerebral degeneration

Vascular

- Caudate infarction or hemorrhage

Pregnancy

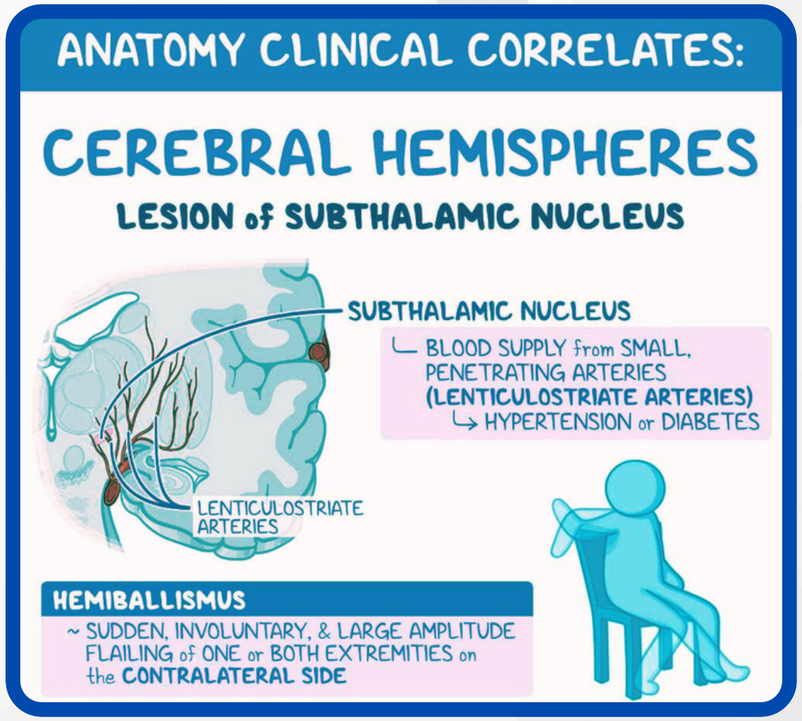
- Chorea gravidarum

Immunologic

- Systemic lupus erythematosus
- Antiphospholipid antibody syndrome
- Poststreptococcal (Sydenham chorea)

Ballism

Aseel Alneis







Ballism

- Ballism is a rare movement disorder characterized by violent, involuntary flinging movementof the limb, usually of one arm or leg (hemiballismus), but can affect both sides in rare cases.
- Etiology:



- Often due to damage or dysfunction of the subthalamic nucleus (part of the basal ganglia)
- Can be caused by lesion such as stroke or tumor, particularly affecting the contralateral subthalamic nucleus ,but also by infections, trauma, or metabolic disorders like hyperglycemia .
- The classic lesion responsible for hemiballismus is an ischemic stroke in the contralateral STN, though lesions in other components of the basal ganglia may be causative.
- Can be seen in neurodegenerative diseases, such as Wilson disease, Huntington's disease, and other basal ganglia pathologies.

Ballism :

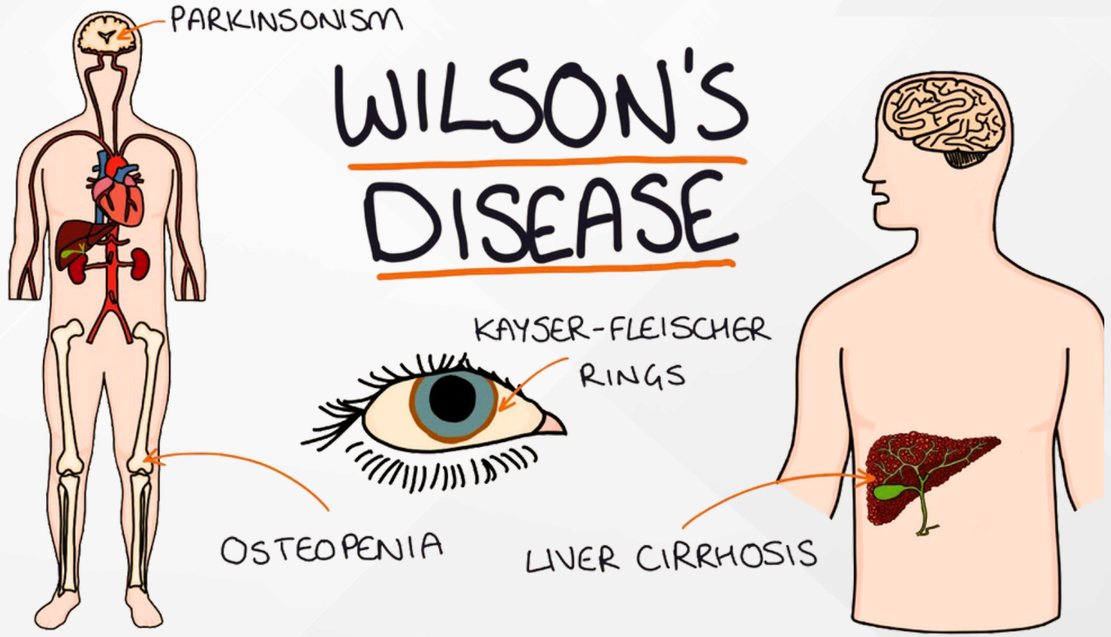
- Lesion in the subthalamic nucleus reduces its inhibitory output to the thalamus, leading to excessive motor activity.
- Movements are often severe enough to cause injury or interfere with daily activities.

Treatment:

- In many cases, ballism resolves on its own, but dopamine-blocking agents may be helpful if this spontaneous improvement does not occur
- Dopamine-blocking agents like antipsychotics (haloperidol) or dopamine-depleting drugs (tetrabenazine) to reduce hyperkinetic movements.
- Treat the underlying cause (e.g., stroke management, metabolic correction).

Wilson's disease

Aseel Alneis





Wilson's disease

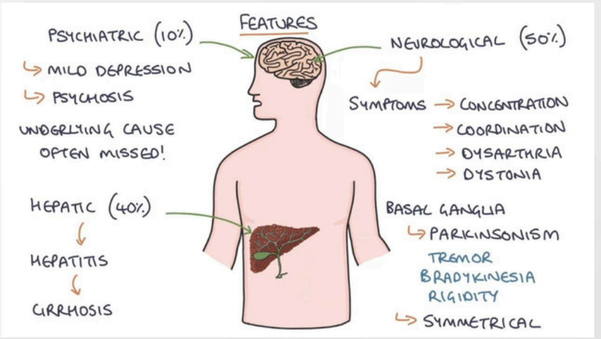
- Wilson's disease (hepatolenticular degeneration) is a rare but important autosomal recessive disorder of copper metabolism caused by a variety of mutations in the ATP7B gene on chromosome 13 leading to copper accumulation in various organs, particularly the liver, brain, kidneys, and corneas. Without proper excretion of copper, it builds up to toxic levels, causing damage to these organs.

- The possibility of Wilson's disease should be considered in any patient under the age of 40
- Fatal if left untreated but manageable with early diagnosis.



Wilson's disease

Normally, dietary copper is absorbed in the stomach and proximal small intestine and then taken up by the liver, where it is incorporated into apocaeruloplasmin to form caeruloplasmin and secreted into the bloodstream, with excess copper excreted via bile, all of which occurs in the presence of the ATP7B protein. However, in Wilson's disease, mutations in the ATP7B gene lead to impaired copper transport, resulting in decreased incorporation of copper into caeruloplasmin and reduced excretion of excess copper. This accumulation can cause copper toxicity in various organs, particularly the central nervous system (CNS), liver, kidneys, and eyes.



- Main organs affected:
- Liver: Initial site of copper buildup, leading to cirrhosis.
- Brain: Particularly affects the basal ganglia, which is crucial for movement control.

Also when free copper is released into the bloodstream, it can cause massive haemolysis , renal tubulopathy and osteoporosis.

Neurological Symptoms

There are multiple neurologic manifestations of Wilson Disease (WD), including:

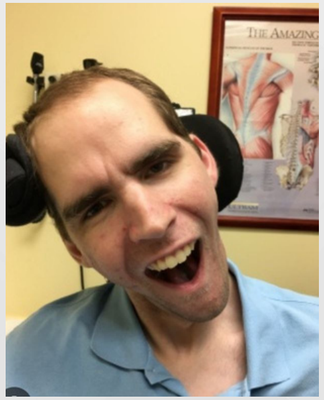
Tremor

Patients may exhibit a coarse tremor, often described as a "wing-beating tremor," resembling the flapping of a bird's wings.



Dystonia

Muscle contractions may lead to abnormal postures or movements; a common presentation is facial dystonia, producing a grimace known as risus sardonicus.



Chorea

Sudden, unpredictable, dance-like movements.

Parkinsonism

Tremors, bradykinesia (slow movements), and rigidity.

Neurological Symptoms

- **Dysarthria**: Slurred speech due to impaired muscle control.

- **Ataxia**: This involves a lack of voluntary coordination of muscle movements, resulting in unsteady gait and difficulties with fine motor skills

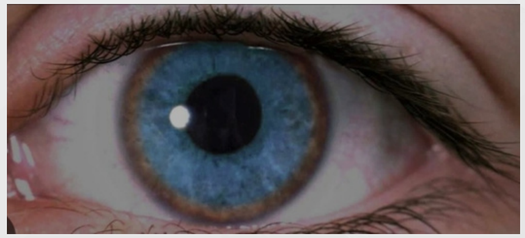
- **Cognitive Decline**: Memory loss, reduced attention span, and executive dysfunction.

- **Psychiatric Symptoms**:
  - Depression, anxiety, irritability.
  - Psychosis and personality changes may also occur.

Diagnostic Tools

The diagnosis of Wilson Disease is established through a combination of clinical evaluation and laboratory findings:

- Serum Copper: Increased serum copper levels are indicative of copper overload.
- Ceruloplasmin: Typically decreased in WD; this copper-carrying protein is often deficient.
- 24-hour Urinary Copper Excretion: Increased urinary copper levels (>100 mcg/day) help confirm excessive copper accumulation.
- Kayser-Fleischer rings: Copper deposits around the cornea (visible with slit-lamp exam).



- Liver biopsy: Confirms excessive hepatic copper deposition
- Imaging: MRI of the brain can show characteristic changes associated with copper accumulation

Treatment:



Treatment options for Wilson Disease include:

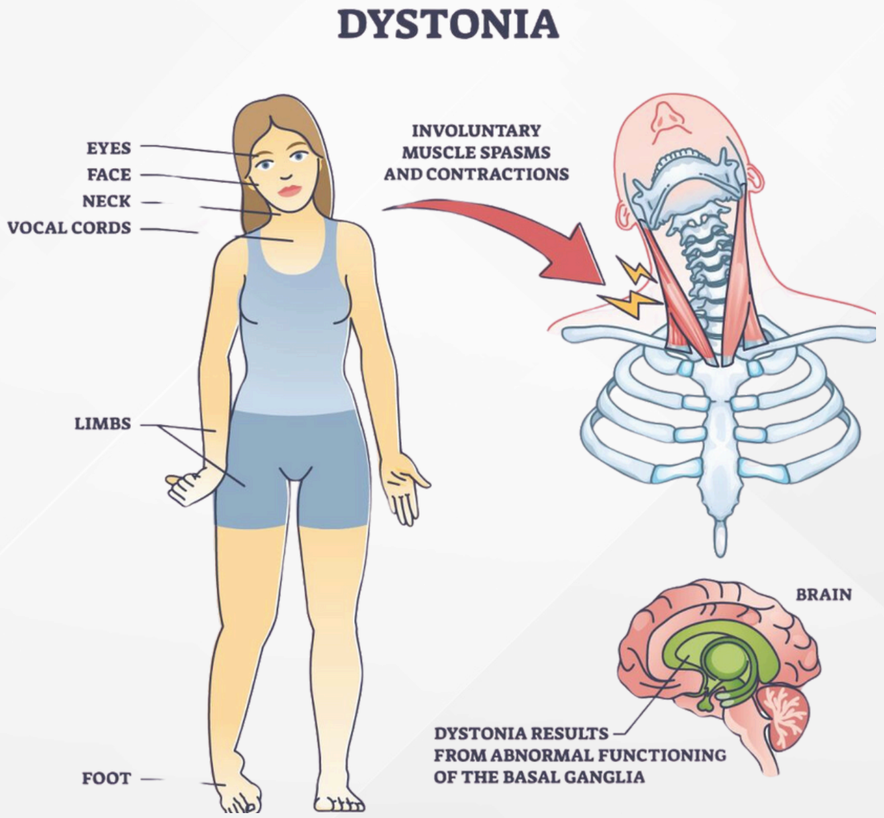
- **Copper-Chelating Agents:**

Penicillamine and Trientine are commonly used to bind copper and facilitate its excretion from the body.

- **Zinc Supplementation**: Zinc competes with copper for absorption in the intestines and reduces copper accumulation.
- **Dietary Management**: Patients are advised to restrict copper-rich foods such as liver, shellfish, nuts, and mushrooms.
- **Liver Transplantation**: In cases of severe liver damage or fulminant hepatic failure, liver transplantation may be necessary.
- Regular monitoring of copper levels and liver function tests is essential to evaluate treatment effectiveness and adjust therapy as needed.
- Earlier diagnosis and treatment result in better long-term neuropsychiatric and hepatic outcomes, emphasizing the importance of awareness and screening for Wilson Disease.

DYSTONIA

Albasal Jalal



DEFINITION :

- Is a sustained contraction of agonist and antagonist muscles , producing twisting movements or abnormal postures.
- This abnormal movements are worsend by movement , and relieved by sensory tricks such as touching or stroking the affected body part.

TYPES :

1. focal affecting one body part and it is more common in adults.
2. segmental affecting one region.
3. generalized affecting multiple body parts and it is more common in children.

FOCAL DYSTONIA EXAMPLES

1. Torticollis which is excessive contraction of the neck muscles resulting fixed head position and it's often painful.
2. spasmodic dysphonia involve the laryngeal muscles and may result in choppy or strangled speech (adductor spasmodic dysphonia) or a breathy voice quality (abductor spasmodic dysphonia).
3. Blepharospasm which is sustained contraction of the orbicularis oculi and forced closure of the eyelids.
4. Writer's cramp which is a focal dystonia of the hand and arm muscles that prevents a patient from using a writing implement properly.

Botulinum toxin injections are the best treatment option for the focal type

DYT–TOR1A DYSTONIA

- It's the most common example of generalized dystonia
- It's an autosomal dominant disorder caused by a mutation in the gene that encodes the protein torsinA
- It starts in childhood or adolescence and often begins in the foot and leg muscles.

TREATMENT

- Anticholinergic agents such as trihexyphenidyl, or baclofen, benzodiazepines.
- Deep brain stimulation of the globus pallidus interna.

Dopa– responsive dystonias

Are an important group of childhood-onset dystonias and are caused most often by A mutation in the gene encoding the enzyme GTP cyclohydrase 1.

- Parkinsonism may accompany the dystonia.

from it's name this group of conditions is responsive to dopamine
that's why we try levodopa in all children who develop dystonia.

#Dystonia may also occur as a manifestation of other central nervous system disorders including Wilson disease (WD), PD, HD, anoxic brain injury, stroke, multiple sclerosis, and medication-induced movement disorders.

TICS

Roaa Al-dreini



TICS

- Tics are abnormal, brief muscle contractions that may involve the face, extremities, or speech.
- They tend to vary in intensity and are irregular in frequency, sometimes occurring in runs of multiple tics and often suppressible for short periods of time.
- Patients with tics describe an internal sensation of an urge to move or perform the tic, with a sense of relief after the tic has occurred.
- Stress tends to exacerbate tics.
- Tics are most often idiopathic, but head trauma, encephalitis, and cerebrovascular disease may produce tics.



Tics can be divided into :

- **Simple Motor** — Include eye blinks, facial grimaces, and shoulder shrugs
- **Complex Motor** — Include spitting and finger cracking
- **Simple Vocal** — Include grunting, throat clearing, and coughing.
- **Complex Vocal** — More extensive vocal utterances of several words blurted out. Including foul language (coprolalia),

Tourette syndrome

- Gilles de la Tourette syndrome is a pediatric-onset disorder in which patients develop motor and vocal tics.
- It has a presumed genetic origin, but a single responsible gene mutation has not been identified.
- Boys tend to be affected more often than girls.
- Multiple motor and vocal tics are present; they may change over time and even go into periods of remission.
- There is a tendency for the tics to improve in adulthood.
- Obsessive-compulsive disorder, attention-deficit hyperactivity disorder, and depression often accompany Tourette syndrome.



PANDAS

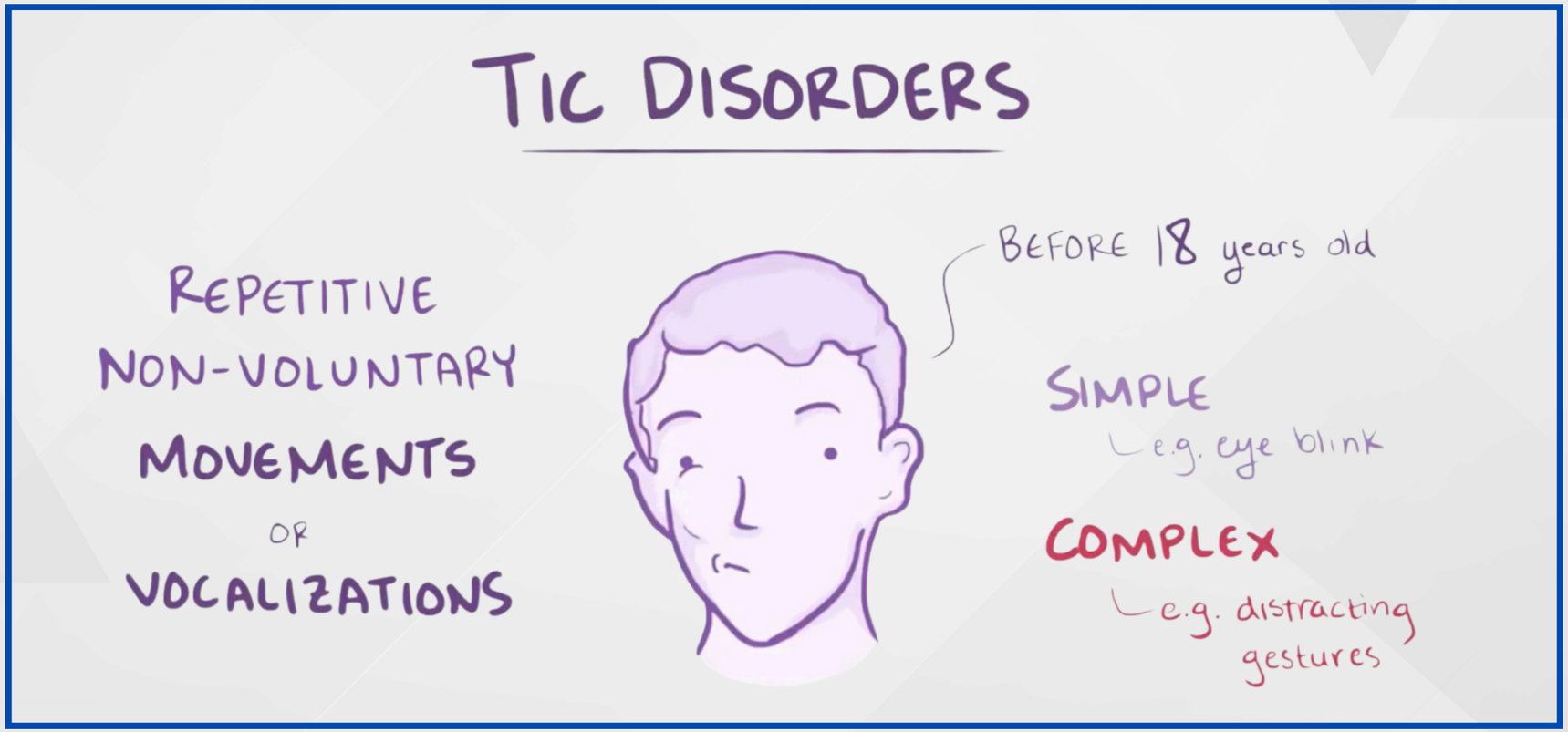
- Pediatric autoimmune neuropsychiatric disorders associated with streptococcal infections (PANDAS) are a combination of tics, obsessive-compulsive disorder, and anxiety following group A ß-hemolytic streptococcal infection.

Etiology of PANDAS

- Is controversial, but presumably the streptococcal infection triggers are autoimmune reaction against the basal ganglia.
- The symptoms are temporary and respond to treatment with antibiotics and immunomodulatory therapy.

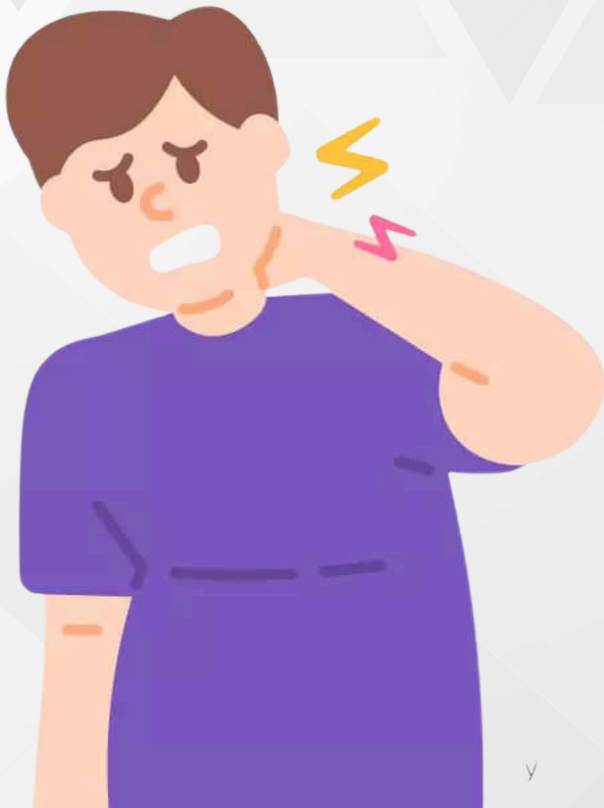
Treatement of Tics

- Dopamine antagonists (haloperidol, risperidone, and pimozide are used most commonly), guanfacine, clonazepam, and clonidine.
- In many cases, tics improve during youth and disappear by the teenage years or early adulthood.



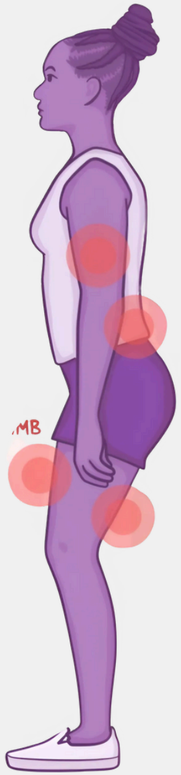
STIFF-PERSON SYNDROME

Sahar Hussein



Stiff–person syndrome

- is an autoimmune or paraneoplastic condition characterized by disabling muscle rigidity and spasms.
- Symptoms typically start with axial and trunk muscles and spread throughout the body over time.
- In addition to progressive muscle stiffness, patients have episodic worsening of their muscle stiffness often provoked by physical or psychological stress.



Stiff–person syndrome

- Other autoimmune diseases including diabetes and thyroiditis are often present.
  Finding antibodies to glutamic acid decarboxylase-65 (GAD-65) supports the diagnosis .

- Antispasmodic medications including : benzodiazepines and baclofen are the standard treatments for stiff-person syndrome.

- patients with refractory disease, immunosuppressive therapy including corticosteroids and intravenous immunoglobulin may be helpful.

PAROXYSMAL DYSKINESIAS

Sahar Hussein



PAROXYSMAL DYSKINESIAS

rare, episodic movement disorders, which typically take the form of chorea, athetosis, or dystonia. Consciousness is preserved during episodes, and there are no other neurologic symptoms .

Inparoxysmal kinesigenic dyskinesia, the dyskinetic movements are triggered by sudden movements and are brief, lasting for seconds or minutes.

Paroxysmal nonkinesigenic dyskinesias occur independent of movement and tend to last for minutes to hours. They are often triggered by alcohol, fatigue, and stress.

Carbamazepine is often a helpful treatment for paroxysmal dyskinesias.

Thank You!

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Reference:

Blueprints Neurology
5th Edition