

Chorea and Athetosis

Chorea is a movement disorder that causes sudden, jerky movements. They are usually purposeless, arrhythmic, and mostly affect the distal extremities. Athetosis is a similar disorder that looks like slow, snake-like writhing movements. Lesions in the basal ganglia motor loops are responsible for most choreiform and athetotic pathology. Choreoathetosis is classically associated with Huntington disease and rheumatic fever. The diagnosis is clinical but genetic analysis may be helpful. Treatment is aimed at the underlying cause but symptomatic cases may benefit from dopamine blocking or depleting agents such as tetrabenazine.



PLAY PICMONIC

Characteristics

Sudden Jerky Movements

Choreographed Dancers

Chorea is a movement disorder that causes sudden jerky, random, and purposeless movements. It comes from the Greek word for "dance".

Athetosis

Athlete-tossing

Athetosis means “without position or place”. The most common underlying cause of athetosis is Huntington's disease. In children, athetosis is commonly associated with dyskinetic cerebral palsy caused by kernicterus, hypoxic-ischemic injury at term birth or prematurity with associated brain injury.

Snake-like, Writhing Movements

Snake-dancer

Atetosis most commonly involves the distal extremities: fingers (hand or feet). It can occur because of damage to the corpus striatum (caudate and putamen) in the basal ganglia.

Basal Ganglia Lesions

Broken Bass-guitar

Lesions in basal ganglia motor loops are responsible for loss of coordination of movements. These may include structures such as the caudate nucleus, putamen, and globus pallidus.

Associations

Huntington's Disease

Hunters

Huntington's disease is the most common cause of chorea in adults. Activation of the nigrostriatal dopamine pathway is believed to induce chorea.

Acute Rheumatic Fever

Roman Fever-beaver

Sydenham's chorea is a milder form of chorea and is listed as one of the five major manifestations of rheumatic fever (Jones criteria). Sydenham's chorea is believed to occur as a result of an autoimmune reaction towards basal ganglia circuitry.

Management

Treat Underlying Disorder

Treating Disorders Under the Table

The treatment of chorea depends on the underlying disease. If Huntington is the cause, tetrabenazine is effective. Patients with Sydenham chorea can be treated with antibiotics to handle the infection. If chorea is occurring as a side effect of a medication, stopping the agent will help to reduce the symptoms.

Tetrabenazine

Tetris on Benz

Treatment of chorea depends on the etiology. A common medication is tetrabenazine, which works by inhibiting vesicular monoamine transporter (VMAT) thereby reducing dopamine exocytosis from presynaptic neurons.