

Nonketotic hemichorea-hemiballism

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Chorea and Ballism

- Chorea : Involuntary hyperkinetic movement disorder consisting of sudden, **irregular, flowing**, purposeless movements that are **distally prominent**
- Ballism : **Proximal**, high amplitude, high velocity movements

Chorea: Classification

Primary (genetic): Huntington's disease, Neuroacanthocytosis, McLeod syndrome, Benign hereditary chorea, Wilson's disease, Dentatorubralpallidoluysian atrophy (DRPLA)

Secondary: Vascular chorea, Autoimmune chorea (Sydenham's chorea), Metabolic chorea (hyperglycemia), Drug-induced chorea, Infectious chorea

Nonketotic hemichorea-hemiballism (diabetic striatopathy)

Hemichorea-hemiballism is a rare hyperkinetic movement disorder characterized by typically continuous unilateral involuntary and nonrhythmic movements of one or both limbs. Though basal ganglia infarction accounts for most cases, uncontrolled and prolonged hyperglycemia now represents the second-most common etiology, designated by the clinical acronyms NKHH (nonketotic hemichorea-hemiballism) or HIHH (hyperglycemia-induced hemichorea-hemiballism).

The imaging features are relatively characteristic, typified by **increased T1 signal intensity** on MRI within the basal ganglia contralateral to the symptomatic extremity. There is a predilection for involvement of the putamen. **Rarely, bilateral involvement may be observed.**

The mainstay of diabetic striatopathy treatment is **control of hyperglycemia** with proper hydration to correct the underlying metabolic imbalance. Although chorea could be successfully treated with glucose control only in one-fourth of the patients, the majority needed additional anti-chorea medications for symptom control. There are four main categories of anti-chorea medications, namely antipsychotics, GABA-receptor agonists, selective serotonin reuptake inhibitors and dopamine-depleting agents.

References:

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