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Neurodegeneration with Brain Iron Accumulation

National Institute of Neurological Disorders and Stroke (NINDS)

Source

National Institute of Neurological Disorders and Stroke (NINDS). [Neurodegeneration with Brain Iron Accumulation Information Page.](#)

Neurodegeneration with brain iron accumulation (NBIA) is a rare, inherited, neurological movement disorder characterized by an abnormal accumulation of iron in the brain and progressive degeneration of the nervous system. Symptoms, which vary greatly among patients and usually develop during childhood, may include dystonia (slow writhing, distorting muscle contractions of the limbs, face, or trunk), dysarthria (slurred or slow speech) choreoathetosis (involuntary, purposeless jerky muscle movements), muscle rigidity (uncontrolled tightness of the muscles), spasticity (sudden, involuntary muscle spasms), and/or ataxia (inability to coordinate movements), confusion, disorientation, seizures, stupor, and dementia. Visual changes are also common, most often due to atrophy of the optic nerve (optic atrophy) or degeneration of the retinal layer in the back of the eye (retinal degeneration). Cognitive decline occurs in some forms of NBIA; the majority of individuals with NBIA do not have cognitive impairment. Several genes have been found that cause NBIA.