



Disability Awareness Begins With You: Hallervorden-Spatz Disease

(Neurodegeneration with Brain Iron Accumulation)

What is Neurodegeneration with Brain Iron Accumulation?

Neurodegeneration with brain iron accumulation (NBIA) is a rare, inherited, neurological movement disorder characterized by progressive degeneration of the nervous system. Symptoms, which vary greatly among patients and usually develop during childhood, may include slow writhing, distorting muscle contractions of the limbs, face, or trunk, choreoathetosis (involuntary, purposeless jerky muscle movements), muscle rigidity (uncontrolled tightness of the muscles), spasticity (sudden, involuntary muscle spasms), ataxia (inability to coordinate movements), confusion, disorientation, seizures, stupor, and dementia. Other less common symptoms may include painful muscle spasms, dysphasia (difficulty speaking), mental retardation, facial grimacing, dysarthria (poorly articulated speech), and visual impairment.

Is there any treatment?

There is no cure for NBIA, nor is there a standard course of treatment. Treatment is symptomatic and supportive, and may include physical or occupational therapy, exercise physiology, and/or speech pathology.

What is the prognosis?

Most patients experience periods of rapid deterioration lasting 1–2 months, with relatively stable periods in between. The rate of progression correlates with the age at onset, meaning that children with early symptoms tend to fare more poorly. For those with early onset, dystonia and spasticity can eventually limit the ability to walk, usually leading to use of a wheelchair by the midteens. Life expectancy is variable, although premature death does occur in NBIA. Premature death usually occurs secondary to dystonia and impaired swallowing, which can lead to poor nutrition or aspiration pneumonia. With improved medical care,

however, a greater number of affected individuals reach adulthood. For those with atypical, late-onset NBIA, many are diagnosed as adults and live well into adulthood.

What research is being done?

The NINDS supports research on neurodegenerative movement disorders such as NBIA. The goals of this research are to increase understanding of these disorders, and to find ways to prevent, treat, and cure them.

Organizations:

NBIA Disorders Association

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