Hereditary Spastic Paraplegia

Hereditary spastic paraplegia (HSP) is a rare group of inherited disorders. This disorder is also called familial spastic paraparesis (FSP). This disorder can be passed from a parent to child through genes. These disorders are characterized by progressive weakness and stiffness of the legs.

SYMPTOMS

The main problem is severe, progressive, lower extremity stiffness. Mild stiffness and problems with gait are typical, early in the disease process and progress to the point that assistive devices such as canes or wheelchairs are needed. Then the disorder can have other neurological problems. These include:

- Damage of the optic nerve (*optic neuropathy*).
- Diseases of the retina (*retinopathy*).
- Cataracts.
- Seizures.
- Dementia.
- Lack of muscle control (*ataxia*).
- A skin disorder that causes dry, rough, scaly skin (*icthyosis*).
- Mental retardation.
- Problem with the nerves that connect to the brain and spinal cord (*peripheral neuropathy*).
- Deafness.

DIAGNOSIS

The diagnosis is mostly made by:

- Neurological examination.
- Testing to exclude other disorders.

Specialized genetic testing and diagnosis are available at some medical centers.

TREATMENT

There are no specific treatments to prevent, slow or reverse HSP. Symptomatic treatments used for other forms of longstanding (*chronic*) paraplegia are sometimes helpful. Regular physical therapy is important for:

- Improving muscle strength.
- Preserving range of motion.

The likely outcome (*prognosis*) for individuals with HSP varies. Some cases are seriously disabling. Others are less disabling. They are compatible with a productive and full life. The majority of individuals with HSP have a normal life expectancy. It is not possible to predict how quickly or severe symptoms will progress. Once symptoms start, they slowly progress throughout life. In some childhood forms, there is progression that stabilizes once a child reaches adolescence. HSP rarely results in total loss of mobility of limb.

RESEARCH BEING DONE

The National Institute of Neurological Disorders and Stroke (NINDS) supports research on genetic disorders such as HSP. Genes that are responsible for several forms of HSP have already been identified. Many more will likely be identified in the future. Understanding how these genes cause HSP will lead to ways to prevent, treat and cure HSP.

Research is continually being done to learn more about this problem.

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