Primary Lateral Sclerosis

Primary lateral sclerosis is a rare neuromuscular disease. It causes progressive muscle weakness in the voluntary muscles. Onset of the disorder generally occurs after age 50. PLS belongs to a group of disorders known as motor neuron diseases. It occurs when specific nerve cells in the cerebral cortex that control voluntary movement gradually degenerate. This causes the muscles under their control to weaken.

SYMPTOMS

Symptoms may include:

- Difficulty with balance.
- Weakness and stiffness in the legs.
- Clumsiness.
- Sudden, involuntary muscle spasms (*spasticity*) in the hands, feet, or legs. This produces slowness and stiffness of movement.
- Dragging of the feet. This leads to an inability to walk.
- Facial involvement. This can cause poorly articulated speech (*dysarthria*).
- The disorder usually begins in the legs. But it may also begin in the tongue or the hands.

DIAGNOSIS

Scientists believe PLS is not hereditary. It progresses gradually over a number of years, or even decades. It leads to stiffness and weakening of the affected muscles. In PLS, there is no evidence of degeneration of spinal motor neurons or muscle wasting (*amyotrophy*). Those do occur in a similar disorder called amyotrophic lateral sclerosis or ALS (Lou Gehrig's disease). For the first few years (3-5) after symptoms begin, there is a possibility that the true diagnosis is ALS. ALS can have some of the same symptoms as PLS. For this reason, most neurologists follow the affected individual's clinical course for a few years before making a diagnosis of PLS.

TREATMENT

Treatment for individuals with PLS is symptomatic. Baclofen and tizanidine may reduce spasticity. Quinine or phenytoin may decrease cramps. Physical therapy may help prevent joint immobility. Speech therapy may be useful for patients with dysarthria.

There is no cure for PLS. The disorder is not fatal.

RESEARCH BEING DONE

The NINDS conducts a broad range of research on neuromuscular diseases. It is aimed at:

- Finding better treatments.
- Preventing and curing these disorders.

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

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