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Primary Lateral Sclerosis

National Institute of Neurological Disorders and Stroke (NINDS)

Source

National Institute of Neurological Disorders and Stroke (NINDS). [Primary Lateral Sclerosis Information Page](#).

Primary lateral sclerosis (PLS) is a rare neuromuscular disease with slowly progressive weakness in voluntary muscle movement. PLS belongs to a group of disorders known as motor neuron diseases. PLS affects the upper motor neurons (also called corticospinal neurons) in the arms, legs, and face. It occurs when nerve cells in the motor regions of the cerebral cortex (the thin layer of cells covering the brain which is responsible for most higher level mental functions) gradually degenerate, causing movements to be slow and effortful. The disorder often affects the legs first, followed by the body, trunk, arms and hands, and, finally the bulbar muscles (muscles that control speech, swallowing, and chewing). Symptoms include weakness, muscle stiffness and spasticity, clumsiness, slowing of movement, and problems with balance and speech. PLS is more common in men than in women, with a varied gradual onset that generally occurs between ages 40 and 60. PLS progresses gradually over a number of years, or even decades. Scientists do not believe PLS has a simple hereditary cause. The diagnosis of PLS requires extensive testing to exclude other diseases. When symptoms begin, PLS may be mistaken for amyotrophic lateral sclerosis (ALS) or spastic paraplegia. Most neurologists follow an affected individual's clinical course for at least 3 to 4 years before making a diagnosis of PLS.