

Primary lateral sclerosis (PLS) is a rare, neuromuscular disorder that affects the central motor neurons and is characterized by painless but progressive weakness and stiffness of the muscles of the legs. Such weakness may progress to affect the arms and the muscles at the base of the brain (bulbar muscles). Less frequently, the muscles of the face are affected. In most cases, the disorder affects adults during midlife. The exact cause of primary lateral sclerosis is unknown. The exact cause of primary lateral sclerosis is not known. Most cases seem to occur randomly, for no apparent reason (sporadically). Primary lateral sclerosis is one of a group of disorders known as motor neuron diseases. Motor neuron diseases are characterized by malfunction of the nerve cells (motor neurons) within the brain and spinal cord that carry instructions from the brain to the muscles. Primary lateral sclerosis is a rare disorder that affects males and females in equal numbers. In most cases, the disorder occurs during the fifth decade. However, according to the medical literature a familial form may exist that affects children. The exact prevalence of primary lateral sclerosis and motor neuron diseases is unknown. Treatment of primary lateral sclerosis involves the use of drugs to help control specific symptoms. Baclofen and tizanidine may be prescribed for spasticity, quinine for cramps, and diazepam, a drug that relaxes muscles, for muscular contractions. Additional treatments may include physical therapy to prevent stiffness of joints, and speech therapy may be needed to aid affected individuals whose ability to speak has been impaired by muscle weakness. Other treatment is symptomatic and supportive.