

What is Primary Lateral Sclerosis?

Primary lateral Sclerosis is a disorder of the upper motor neurons. The degeneration of these upper motor neurons in the brain and spinal cord, which control voluntary movements, cause disabling spasticity and weakness. As the muscles are not directly affected, there is no wasting or fasciculation's (rippling effect under the skin), with this condition. There are also lower motor neurons in the body, but these are unaffected by primary lateral sclerosis.

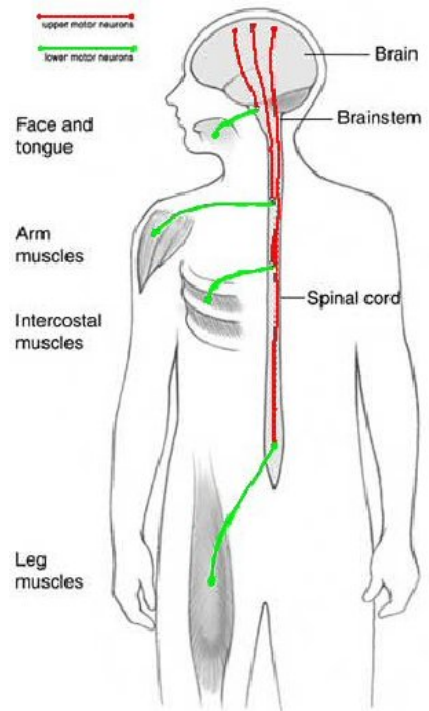
The red lines indicate upper motor neurons, which are affected by primary lateral sclerosis.

Green lines are the lower motor neurons, where no changes occur in primary lateral sclerosis.

How is PLS diagnosed?

Usually tests need to be carried out to exclude other conditions as the likely cause of symptoms. Other conditions may include multiple sclerosis, brain/spinal tumours, spinal injury or a virus.

Amyotrophic lateral sclerosis, which is the commonest form of motor neurone disease, and affects both upper and lower motor neurons, would also have to be excluded.



What are the tests for PLS?

- Magnetic resonance imaging (MRI) scan: used to detect any abnormality of the brain and spinal cord.
- Nerve conduction test: uses small electrical shocks to check the speed of nerve conduction to the muscles; in MND this would appear normal.
- Electromyogram (EMG): measures the activity of muscles to detect any dysfunction.
- Blood tests to check on general health.

PLS Symptoms

- Lower limb weakness, a decreased sense of balance, and stiffness in the limbs.
- Clumsy hands, slurred speech.
- Emotional lability (inappropriate or uncontrolled laughing or crying)
- Speech and swallowing difficulties.
- Bladder urgency in some cases.

Who gets Primary Lateral Sclerosis?

Generally it affects people over 50 years old, whether male or female. It is not passed on to other members in a family.

PLS Prognosis

Life span could essentially be normal, although it may be life limiting, depending on whether the progression remains as primary lateral sclerosis, or develops into amyotrophic lateral sclerosis.

How might I be affected by PLS?

You may have a greater tendency to fall, as balance is affected. There may also be some pain in the neck, back and legs.

Tasks such as washing, dressing, writing, and cooking may be difficult, because of decreased mobility.

Speech problems may occur due to the muscles in the throat and neck becoming sluggish. This may result in impaired control of the tongue, lip and palate, causing a loss of clear speech.

Swallowing difficulties may result in problems with eating and drinking. There may also be loss of emotional control, with inappropriate laughing or crying.

Management of Primary Lateral Sclerosis

Management of the disease is mostly by symptom control. This includes medication to reduce cramped and tightened muscles and physical therapy/exercise, to maintain flexibility and range of motion. Assistive devices and communication aids may also be appropriate.

Exercise

Exercise such as walking, or water aerobics, may help to strengthen muscles to compensate for those that are weakening. Stretching exercises may also lessen muscle cramps, and maintain motion. Always consult a doctor before undertaking any exercise.

Your doctor may prescribe medication to relieve cramps and stiffness.

Speech

Speech problems may become such, that speech is impaired over time. There are many aids that could be useful to continue communication, such as a light writer or computer adaptations.

Feeding

You may become dependent on others for your daily intake of nutrition and fluids. If feeding becomes difficult and tiring, due to the swallowing muscles becoming sluggish, it may be appropriate to consider PEG feeding. This would help you to get better nourishment, and a feeling of well being.

In all cases, assessment by a professional is advised, in order to benefit from the appropriate services.

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References: Primary Lateral Sclerosis by Carmel Armon, MD, MHS.(2003)