PRIMARY LATERAL SCLEROSIS (PLS)

FACT SHEET | AMYOTROPHIC LATERAL SCLEROSIS SOCIETY OF CANADA | SOCIÉTÉ CANADIENNE DE LA SCLÉROSE LATÉRALE AMYOTROPHIQUE

WHAT IS PLS?

Like ALS, primary lateral sclerosis (PLS) is a progressive degenerative disease of the motor neurons. It is characterized by progressive spasticity (involuntary muscle tension and spasms) and it affects the lower limbs, trunk, upper limbs, and bulbar muscles, usually in that order.

HOW DOES PLS DIFFER FROM ALS?

PLS affects the upper motor neurons alone, while ALS affects the upper and lower motor neurons. The nerve cell bodies of the upper motor neurons reside in the brain, where they control the activity of the lower motor neurons, which reside in the spinal cord. In PLS, there is no evidence of the degeneration of spinal motor neurons that occurs in ALS.

The progress of PLS is more gradual and less devastating than that of ALS. Unlike ALS, PLS does not result in muscle wasting, and although it is disabling, it is not fatal. Some ALS specialists believe that PLS is on the ALS continuum and may not be a separate disease, but a very slow-progressing type of ALS. ALS may begin with signs of only upper or lower motor neuron involvement, so a process that begins with upper motor neuron degeneration and that initially is considered to be PLS has the potential to be reclassified as ALS if sufficient signs of both upper and lower motor neuron involvement develop over time.

WHAT ARE THE SYMPTOMS OF PLS?

PLS usually begins with lower-extremity stiffness and pain due to spasticity. With progression, which is a gradual process, people with PLS may develop balance problems and lower back and neck pain. As the upper limbs become affected, activities of daily living may become difficult to perform. Speech impairment may occur as a result of damage to the muscles involved in forming words, and swallowing and breathing may be compromised in the later stages of the disease.

The slow rate of progression allows people with PLS and their families and caregivers more time to adapt to changes than with the more rapidly progressive symptoms of ALS, but the longer duration of the disease places upon them a greater burden of care.

WHAT IS THE LIFE EXPECTANCY OF A PERSON WITH PLS?

In general, PLS is not considered to shorten life expectancy, although people who are young at the onset of the disease may live a shorter life span.

WHAT CAUSES PLS?

The cause of PLS is unknown.

WHAT IS THE TREATMENT FOR PLS?

There is no treatment that will reverse or slow the progress of PLS. Symptoms may be treated with drugs to reduce muscle spasticity or cramps, physical therapy to reduce joint immobility, and speech therapy to help those whose facial muscles are affected.

HOW FREQUENTLY DOES PLS OCCUR?

The incidence of PLS is rare, but the exact number is uncertain. ALS affects two individuals per 100,000 each year, and tentative estimates put the annual PLS incidence rate at half a per cent of that number. The median duration of PLS is approximately 20 years, while the duration of ALS is two to five years, so PLS prevalence is high relative to incidence because people with the disease live longer. If tentative U.S. statistics are accurate, we can extrapolate that there are approximately 50 Canadians living with PLS.

IS PLS HEREDITARY?

PLS is not considered a genetic disorder since almost all cases occur sporadically. However, a very rare form of PLS with an early childhood or juvenile onset has been linked to a mutation in one gene. There is also a genetically mediated look-alike, progressive familial paraparesis (a partial paralysis of the legs) which is a separate condition with more limited symptoms and a more benign course.

DOES PLS AFFECT ONE SEX MORE THAN THE OTHER?

The numbers under study are too small to make a definitive link to gender.

WHAT IS THE AVERAGE AGE OF ONSET OF PLS?

The age of onset ranges from 35-66 years with a median age of 50.5.

HOW IS PLS DIAGNOSED?

PLS is diagnosed through a process of eliminating the possibility of other neurological disorders. Affected individuals typically have no family history of similar disorders. There should be signs of upper motor neuron dysfunction and no signs of involvement of other systems. Early on, PLS and ALS can look very similar because ALS sometimes begins as an upper motor neuron disease, and the average age of onset is similar. Lower extremity onset and slow progression of at least three to five years without lower motor neuron involvement would increase confidence in the diagnosis of PLS and decrease the likelihood of a later evolution into ALS. People who have been diagnosed with PLS are advised to see a neurologist regularly to be tested for the characteristic neuronal degeneration of ALS.

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