Primary lateral sclerosis

FACT SHEET | WWW. ALSMB.CA

The ALS Society of Manitoba
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Primary Lateral Sclerosis (PLS) is a rare progressive neurodegenerative disease which is similar to ALS. However, ALS and PLS are not the same thing. Unlike ALS, PLS only affects the upper motor neurons. ALS affects both the upper and lower motor neurons.

In PLS, the motor neurons in the spine do not degenerate. Therefore, people with PLS do not tend to experience muscle wasting. The most common symptoms include muscle stiffness, spasticity, and cramps.

PLS also has a much more gradual progression than ALS, and it does not shorten a person's life expectancy. It is not fatal.

PLS is very rare, even rarer than ALS. It is estimated that PLS affects 0.5% as many people compared to the number of people with ALS. The number of people with ALS is estimated at 2 in 100,000 per year.[1]

People with PLS live longer than people with ALS. This means the ALS and PLS prevalence rates are difficult to compare because of the difference in lifespan -- relatively more people with PLS will be alive than they would be if they had ALS. There are approximately 50 Canadians living with PLS at a given time.

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

IS PLS SEPARATE FROM ALS?

There is some debate as to whether PLS and ALS are separate disorders, or if they form two parts of a continuum. Some researchers believe PLS may simply be a milder, slower form of ALS. Future research will reveal the ways PLS and ALS are related.

Some people are initially diagnosed with PLS, but the condition progresses to ALS. Probable PLS is defined by the absence of significant active lower motor neuron degeneration 2–4 years from symptom onset. Definite PLS is defined by the absence of significant active lower motor neuron degeneration 4 or more years from symptom onset. After 4 years, if there is no sign of lower motor neuron involvement, a diagnosis of probable PLS becomes definite.

WHAT CAUSES PLS?

The causes of PLS are currently unknown. It is likely that both genetic and environmental factors contribute to PLS.

New research has linked genetic mutations to some forms of PLS. Primary lateral sclerosis appears to be mainly a sporadic disorder, however, with diagnosis based primarily on clinical features rather than genotype.

A rare, juvenile form of PLS results from a genetic mutation, and is considered hereditary.

WHAT ARE THE SYMPTOMS OF PLS?

PLS typically begins with stiffness, spasticity, and pain in the lower extremities. As the disease progresses gradually, people with PLS may experience balance problems and lower back and neck pain. The upper limbs usually become affected at the later stages of the disease, causing difficulty with the activities of daily living.

People with PLS may also experience bulbar symptoms, causing weakness in the tongue. Speech, swallowing, and breathing may be affected.

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.

HOW IS PLS DIAGNOSED?

PLS is diagnosed through a process of elimination of other possible neurological disorders. People affected by PLS typically do not have a family history of similar disorders. In PLS, there should be 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. If a person does not experience muscle wasting or lower motor neuron involvement within 3 to 4 years, it is likely that PLS is the correct diagnosis.

WHAT CARE DO PEOPLE LIVING WITH PLS NEED?

People who have been diagnosed with PLS are advised to see a neurologist regularly for symptom management and to be monitored for any characteristic neuronal degeneration of ALS. People living with PLS may also require the use of mobility and communication devices, as well as home safety equipment.

PLS RESEARCH AND EXPERIMENTAL TREATMENT

Individuals with PLS are typically excluded from ALS clinical trial eligibility criteria. In April of 2020, a group of prominent ALS clinician researchers from around the world published a manuscript highlighting a collaborative effort to prioritize better understanding, diagnosis and emphasis on PLS as its own entity, separate from ALS. This includes development of future measures that would allow PLS clinical trials to be pursued.

SUMMARY

- PLS and ALS are separate, but related disorders that are often thought to be on a continuum.
- It is difficult to differentiate PLS from ALS at the early stages of illness.
- PLS affects only the upper motor neurons, rather than both the upper and lower motor neurons as in ALS.
- People with PLS do not experience muscle wasting. The most common symptoms include stiffness, spasticity, and cramps.
- PLS typically does not shorten the lifespan, and it is not fatal.
- PLS remains a clinical diagnosis. If clinical signs and symptoms change over time, a person who was initially diagnosed with PLS may be rediagnosed with ALS.
- Researchers are interested in determining the mechanisms involved in PLS, investigating potential treatments, and improving diagnostic tools.

KNOW THAT WE ARE HERE TO HELP I For people and families living with ALS in Ontario, ALS Canada can assist in connecting you to support services, equipment, and ALS clinics. Whether you are a person living with ALS, a family member or a caregiver, we will strive to support you along this journey. If you live outside of Ontario, please contact your provincial ALS Society for information on support available in your region. Learn more at www.als.ca.

Special thanks to everyone who helped write and review this fact sheet.

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