

Progressive Supranuclear Palsy (PSP) is a rare progressive neurodegenerative disease that causes vision difficulties and loss of balance while walking. It is due to an abnormal accumulation of a protein in the nerve cells, called tau, which causes parts of the brain to deteriorate over time. As a result, people with PSP may experience increased difficulty with their thinking, speech, swallowing and movement.

About progressive supranuclear palsy

PSP, also known as the Steele-Richardson-Olszewski syndrome, is caused by an overproduction of a protein in the brain called tau which causes nerve cells in specific areas of the brain to become increasingly damaged over time. It is the specific involvement of these areas in the brain that cause the characteristic symptoms of PSP.

The outer layer of the brain (the cortex) becomes involved and causes difficulties with thinking, understanding and speaking (language). The involvement of the deeper areas of the brain, including the substantia, the basal ganglia, the subthalamus and the brainstem will cause the difficulties in balance, walking, eye movements, and the slowness and stiffness in movements.

What are the symptoms?

Symptoms of PSP are most commonly seen in people in their early 60's, but may begin in some people who are in their 40's. PSP can be confused with Parkinson's disease and Corticobasal Degeneration because some symptoms, the stiffness and slow movements, can resemble some of the common characteristics of these other neurodegenerative diseases. Unlike Parkinson's disease, tremor is not seen.

Early symptoms of this disease may be related to a person's increased difficulty with walking and balance, often resulting in frequent falls. It is common for a person in the early stages of PSP to develop other motor-related symptoms like slowed or awkward movements while walking.

Symptoms that help to differentiate PSP from other neurodegenerative diseases, like Parkinson's, are often related to a person's vision and eye movements. People with PSP often experience blurred vision and an inability to control eye movements. Some cannot look downward or cannot open their eyelids. This increased difficulty with eye movements can make it appear that a person is disinterested in conversation because of the limited eye contact.

While not a common early symptom of PSP, changes in thinking and cognition may be experienced as the disease progresses. People with the cognitive symptoms of PSP typically experience difficulties with carrying out a plan, problem solving, adapting to change, making financial decisions, memory and slurred speech.

On occasion, people with PSP may experience changes in their behaviour and emotions that are similar to people with behavioural variant frontotemporal dementia (bvFTD). The behavioural symptom that is commonly shared among people with bvFTD and PSP, is their inability to control their behaviour in social situations and may result in embarrassing social situations. People with the emotional symptoms of PSP may appear to be indifferent towards other people, depressed, or have an inability to control their emotions (prolonged crying).

How is progressive supranuclear palsy diagnosed?

It can be challenging to diagnose PSP early in the progression of the disease as there are a number of symptoms that are shared with similar neurodegenerative diseases, including Parkinson's disease, Alzheimer's disease, frontotemporal dementia and Creutzfeldt-Jakob disease.

People who develop movement difficulties prior to vision challenges are most often misdiagnosed with Parkinson's disease. PSP is often not considered until people experience challenges with their vision. It is also common for changes in a person's mood to be initially attributed to depression, or for changes in behaviour to be related to symptoms of another dementia like bvFTD and not PSP.

There is no single test to diagnose PSP. Physicians assess types of symptoms to help rule out other conditions that could be causing the symptoms, such as Parkinson's disease or strokes. Brain imaging (MRI) may be used to rule out other conditions, and helps to detect any abnormal changes, like shrinkage, in the parts of the brain that are associated with PSP. A thorough assessment may also include neuropsychological testing to evaluate cognitive functioning, physical and neurological exam to assess a person's movement and any changes in the person's vision.

What are the causes or the risk factors?

At present, there is no known cause of PSP. The disease appears to be sporadic, meaning that in most cases there is no family history of the disease, and that common risk factors have yet to be identified. However, similar to other types of dementia, PSP is characterized by an excess amount of the tau protein in the parts of the brain that are most affected by PSP. It is still unknown what causes this surplus of the tau protein, and why it results in the deterioration of brain cells.

Is there treatment?

Currently, there is no known cure for PSP. Medications that are used to manage symptoms of Parkinson's disease may be of some benefit to people with similar symptoms.

Therapeutic approaches are also supporting people living with PSP to manage symptoms of their disease. Physical therapy is often focused on fall prevention and exercises to improve balance. People with PSP are also frequently encouraged to use weighted walking aids to prevent them from falling backwards, such as a walker. Speech therapy may also be effective in addressing eating and swallowing challenges that are associated with the progression of PSP.

Support is available:

Visit the Alzheimer Society's website at www.alzheimer.ca or contact your local Alzheimer Society.

Additional resources:

- Alzheimer's Society UK:
http://www.alzheimers.org.uk/site/scripts/download_info.php?fileID=1767
- Foundation for PSP, CBD and Related Brain Diseases:
http://www.psp.org/file_download/bdbad4d5-6215-4624-9aec-6f909f459fb5
- The Progressive Supranuclear Palsy Association:
<http://www.pspassociation.org.uk/>

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Sources:

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Foundation for PSP, CBD and Related Brain Diseases, *A Guide for People Living with PSP, CBD, and Other Atypical Parkinsonian Disorders*

http://www.psp.org/file_download/bdbad4d5-6215-4624-9aec-6f909f459fb5

Mayo Clinic, *Diseases and Conditions: Progressive Supranuclear Palsy*

<http://www.mayoclinic.org/diseases-conditions/progressive-supranuclear-palsy/basics/definition/con-20029502>

National Institute of Neurological Disorders and Stroke, *Progressive Supranuclear Palsy Fact Sheet*

http://www.ninds.nih.gov/disorders/psp/detail_psp.htm#261703281

UCSF Memory and Aging Center, *Progressive Supranuclear Palsy*

<http://memory.ucsf.edu/education/diseases/psp>

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