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Progressive Supranuclear Palsy

Progressive supranuclear palsy (PSP) belongs to the category of FTD disorders that primarily affect movement. Some symptoms of both PSP and corticobasal syndrome — another FTD disorder associated with a decline in motor function — resemble those often seen in people with Parkinson's disease. In fact, these features are sometimes referred to as "atypical Parkinsonism."

The earliest motor symptoms are stiffness in the axial muscles, the neck and trunk, along with poor balance and more frequent falls. The earliest visual signs are a decrease in upward vertical movement of the eyes (vertical saccades) and a progressive inability to move the eyes, including opening or closing the eyes. PSP can also affect coordination, and movement of the mouth, tongue, and throat. In addition to motor symptoms, people with PSP may exhibit changes in behavior and language skills common to bvFTD and PPA, particularly as the disease progresses.

Know the Signs…Know the Symptoms

Supranuclear gaze palsies

The person affected may experience an inability to move or aim the eyes vertically (particularly downward) or horizontally (left and right). They may experience rapid involuntary eye movements, or difficulty blinking or excessive blinking. Supranuclear gaze palsies may be experienced as blurring. The person affected may experience difficulty reading, make poor eye contact during conversations, have difficulty going down stairs, or experience impaired vision while driving.

Postural instability

Difficulty maintaining balance, which can lead to frequent, unexplained falls.

English

Gait instability
An unsteady, awkward gait.

Akinesia/bradykinesia
Absence of movement (akinesia) or abnormally slow (bradykinesia) movement.

Rigidity
Stiffness, resistance to movement.

Dysphagia
Difficulty swallowing, including gagging or choking. This can lead to aspiration pneumonia.

Dysarthria
Slurred or slowed speech due to difficulty moving the muscles controlling the lips, tongue and jaw.

Behavioral and emotional symptoms that may occur in PSP
A progressive deterioration in the diagnosed person's ability to control or adjust their behavior appropriately in different social contexts is the hallmark of the behavior changes, and results in the embarrassing, inappropriate social situations that can be one of the most disturbing facets of FTD and related disorders.

In addition to the depression, apathy and inability to control emotions noted above, PSP patients may manifest emotional blunting or indifference toward others and a lack of insight into changes in their own behavior.

Cognitive symptoms
PSP patients may suffer increasing impairment in "executive functions," such as distractibility, mental rigidity and inflexibility, impairments in planning and problem solving, and poor financial judgment. PSP patients may also have memory problems. They also develop progressive language

disturbance.

Diagnosis

Based on overlapping symptoms, PSP is frequently misdiagnosed as Parkinson's or another movement disorder. Some symptoms – especially the potential behavioral and cognitive ones – may appear later in the progression of the disease. If you have concern that you or a loved one may have been misdiagnosed – or about any of the signs and symptoms listed above – it is important to consult a doctor.

Treatment, Management and What to Expect

There is currently no effective treatment for PSP. In some patients, the slowness, stiffness, and balance problems of PSP may respond to anti-parkinsonian agents such as levodopa, or levodopa combined with anticholinergic agents, but the effect is usually temporary. Decreased transmission of GABA in the mid-brain is considered to contribute to the symptoms of PSP and so drugs that work on the GABA neurotransmitter may be helpful. Zolpidem, in limited case studies, has improved speech and facial expressions, as well as akinesia rigidity and dysarthria. However, this drug is short acting and prolonged use is not recommended. In general, speech, vision, and swallowing difficulties usually do not respond to any currently available drug treatment.

Another group of drugs that has been of some modest success in PSP are antidepressant medications. The most common antidepressants used for PSP have been Prozac, Elavil and Tofranil. The anti-PSP benefit of these drugs seems to be unrelated to their ability to relieve depression.

Management of PSP symptoms can take many forms. Patients frequently use weighted walking aids to counteract their tendency to fall backward. Bifocals or special glasses called prisms are sometimes prescribed for PSP patients to remedy the difficulty of looking down. Although formal physical therapy is of no proven benefit in PSP, exercises can be done to keep the joints limber. Prevention of injuries related to recurrent falls is a major focus of physical therapy.

PSP predisposes those affected to serious complications such as pneumonia followed from difficulty in swallowing (dysphagia). The most common complications are choking and pneumonia, head injury, and fractures caused by falls. A surgical procedure involving a gastrostomy tube may be done when there are significant and severe swallowing disturbances. This surgery involves the placement of a tube through the skin of the abdomen into the stomach for feeding purposes.

The most common cause of death is pneumonia. With good attention to medical and nutritional needs, however, most PSP patients live well into their 70s and beyond.

Pathology

Like all FTD disorders, PSP is associated with degeneration of the brain's frontal and temporal lobes. In addition, brain regions that help to control and coordinate movement and balance, especially areas in the brainstem controlling ("supra") groups of brain cells ("nuclei") related to eye

movements (hence, “supranuclear” palsy, or weakness), are also affected — as well as the basal ganglia, substantia nigra, and subthalamic nucleus.

In contrast to most other FTD disorders, the protein that accumulates in the brains of all people with PSP is tau — most commonly a type known as 4R tau. For this reason, pathologists sometimes refer to PSP as a “pure tauopathy.” In addition to being folded incorrectly, the 4R tau in the protein deposits is *hyperphosphorylated*, meaning it contains an unusually large number of chemical modifications called phosphate groups.

More Information

- Managing FTD (https://aftd2.wpengine.com/living-with-ftd/managing-ftd/)
- Coordinating Care (https://aftd2.wpengine.com/living-with-ftd/coordinating-care/)
- Support for People with FTD (https://aftd2.wpengine.com/living-with-ftd/support-for-people-with-ftd/)
- Support for Care Partners (https://aftd2.wpengine.com/living-with-ftd/support-for-care-partners/)
- CurePSP (http://www.curepsp.org/)

References

Lamb, R., Rohrer, J.D., Lees, A.J., Morris, HR. Progressive supranuclear palsy and corticobasal degeneration: Pathophysiology and treatment options. *Current Treatment Options Neurology*. 2016, September 2016.

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Have Questions?

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