



Progressive Supranuclear Palsy

Progressive supranuclear palsy (PSP) is a condition that causes changes in movement, language and behavior. In its typical form, PSP causes difficulties with balance that lead to frequent falls. Eye movement problems are also a characteristic symptom of PSP, although these are often discovered when a doctor examines a person with PSP. Slowed movement, stiffness and difficulty walking are common. People with PSP may also have changes in speaking, thinking or behavior.

What Causes PSP?

The cause of PSP is unknown. Scientists know that in PSP there is a large build up of a protein called *tau*. Some researchers group PSP along with [CBS](#) and [FTD](#) under a single term called *Pick-complex disorders* or *primary tauopathies*. It is likely that whether one presents with PSP, CBS or FTD depends in part on the location in the brain of these microscopic changes, although individual differences may play an important role as well. Tau occurs normally in the brain, but

we do not yet understand what causes it to build up in large amounts. As more and more proteins clump in the nerve cells, the cells lose their ability to function and eventually die. This causes the affected parts of the brain to shrink.

There have been some cases of a genetic form of PSP that follows an autosomal dominant inheritance pattern with reduced penetrance.

How is Age Related to PSP?

Most people with PSP start having symptoms after age 60, although some people have shown signs earlier or later.

What Happens in PSP?

A clinical evaluation by a neurologist is important in the diagnosis of PSP, as it is often misdiagnosed and difficult to diagnose early. This involves an interview with the patient and a partner (such as a spouse, relative or close friend) to provide examples of behavior and daily functional activities, a physical exam to assess mobility and vision, and a neuropsychological evaluation for evaluation of cognition.

The clinical features used to diagnose PSP are:

1. Progressive difficulty with walking (gait) and balance resulting in frequent falls
2. Progressive loss of voluntary control of eye movements (gives the disorder its name)
3. Progressive changes in behavior or cognition

The most common first sign of PSP is trouble with balance. Because of their balance problems, people with PSP may have trouble walking and fall. Frequently, people with PSP feel stiffness in the neck and back, and their movements may be slowed. A person with PSP will begin to experience eye problems, such as difficulty opening and closing their eyes, blinking, seeing clearly or moving their eyes side to side or up and down. The decreased blinking, along with constantly raised eyebrows, gives the face a fixed staring expression, characteristic of the disease. The gaze difficulties can lead to problems such as difficulty making eye contact, difficulty reading (because of inability to scan lines on a page), and difficulty eating (because of inability to look down at their food).

Dysarthria (slow or slurred speech) is a very common symptom in PSP. People with PSP often find it difficult to converse with others because of their delayed responses and difficulties with speech pronunciation. Eventually, swallowing food, and particularly liquids, can be poorly

coordinated, leading to the leakage of food into the windpipe (dysphagia). This can result in pneumonia, the most common cause of death in PSP. Some warning signs caregivers should look for are drooling, food collecting in the mouth, increased effort in swallowing, chest congestion, trouble talking and weight loss.

PSP patients also experience cognitive and behavioral changes consistent with a decline in frontal lobe functioning, such as slow information processing and retrieval, concrete thinking, impaired reasoning, difficulty planning and shifting between tasks. Behaviorally, patients often exhibit apathy, including decreased motivation and withdrawal, impulsivity and perseveration, an inability to switch tasks or change topic. Depression is also common.

In contrast with PSP, people with [Parkinson's disease](#) (PD) don't experience severe balance dysfunction until later in the course of their disease. They also experience tremors that are uncharacteristic of PSP. In PSP, the posture is stiff and upright with a tendency to fall backwards, as opposed to the stooped posture seen in PD.

While balance and walking problems are usually early features of the disease, the illness can present with cognitive and behavioral changes. Visual and oculomotor problems tend appear early as well.

Later in the disease, people with PSP may feel increasing weakness in their limbs. Their balance may get worse, and they may fall more often while walking. Eye problems, such as keeping eye contact with another person, will get worse. PSP can also affect a person's ability to speak, and their voice may become soft and weak. Mood and behavior changes may occur. People with PSP may become irritable, forgetful, or depressed, and they may become less interested in things. They may also become more impulsive in their decision-making.

PSP is a disease that changes with time. A person with PSP can live many years with the disease. Research suggests that a person with PSP may live an average of seven years with the disease, although this can vary from person to person.

Are There Medicines to Treat PSP?

Though there are no effective treatments for PSP yet, there are medications that help manage the symptoms. Movement symptoms may be treated with medications used for [Parkinson's disease](#), such as levodopa, although the effect of these medications in PSP is debated.

At UCSF, we are working on developing new treatments for this disorder. If you, or someone you know, is interested in participating in a study of a new potential treatment for PSP, please read about our [clinical trials for people with PSP](#).

What Other Things Help?

Speech therapy may be helpful for people with PSP who have problems with speaking and expression. Physical therapy and stretching exercises may help strengthen stiff muscles and improve some movement difficulties. A professional should evaluate fall risks at the home of the person with PSP and make changes as necessary, such as installing rails, removing carpets or adding a shower chair. Using a walking aid with a heavy front to prevent falling backwards, eating more solid foods and less thin liquids, and physical therapy or exercise programs to improve mobility may be helpful. If swallowing problems become severe, insertion of a feeding tube directly into the stomach can significantly decrease the risk of pneumonia.

Research has shown that physical exercise helps to enhance brain health and improves mood and general fitness. A balanced diet, enough sleep, and limited alcohol intake are other important ways to promote good brain health. Other illnesses that affect the brain, such as diabetes, high blood pressure and high cholesterol, should also be treated if present.

Resources

- [NINDS Progressive Supranuclear Palsy Information Page](#)
- [Diagnostic Criteria for Progressive Supranuclear Palsy](#)
- [CurePSP](#)
- [PSP Association](#)
- [Brain Support Network](#)
- [National Organization for Rare Disorders](#)
- [Family Caregiver Alliance](#)
- [National Institutes of Health](#)

Participate in Research

- [Progressive Supranuclear Palsy Trial of ABBV-8E12](#)
- [Progressive Supranuclear Palsy Trial of Oral Salsalate](#)
- [4 Repeat Tauopathy Neuroimaging Initiative – Cycle 2 \(4RTNI-2\)](#)
- [Advancing Research and Treatment for Frontotemporal Lobar Degeneration \(ARTFL\)](#)
- [Creation of Stem Cells from Patients with FTD](#)
- [Eye Movements in Dementia](#)