Progressive Supranuclear Palsy

What is progressive supranuclear palsy (PSP)?

Progressive supranuclear palsy (PSP) is a degenerative brain disease leading to difficulties with walking and balance, problems with eye movements, changes in behavior, difficulty with speech and swallowing, and dementia.

PSP occurs primarily in middle-aged adults and the elderly, with slightly more males being affected than females. Approximately 1.39–6.4 in every 100,000 individuals are estimated to have PSP, but because the disorder is difficult to diagnose, this is thought to be considerably underestimated. In part because it is relatively rare, PSP is frequently misdiagnosed as Parkinson’s disease (PD). However, its treatment response and clinical symptoms are different, making an accurate diagnosis important for patient management.

Most known forms of PSP are sporadic, but there have been some cases of a genetic relationship, following an autosomal dominant inheritance pattern with reduced penetrance.

PSP, corticobasal degeneration (CBD) and frontotemporal dementia (FTD) are not associated with amyloid plaques in the brain, such as those seen in Alzheimer’s disease, but are associated with the abnormal accumulation of a protein called tau. Some researchers have chosen to group PSP along with CBD and FTD under a single term called Pick-complex disorders or primary tauopathies. It is likely that whether one presents with PSP, CBD 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.

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.

Signs & symptoms of progressive supranuclear palsy (PSP)

The clinical features used to diagnose PSP are:

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

Classically, PSP has been portrayed as an illness that presents with imbalance and frequent falls and visual symptoms. In the last decade, there is an appreciation of the significant behavioral and cognitive changes that these patients experience. The motor symptoms observed in PSP include a combination of slowed movement and stiffness in the neck and trunk, in addition to the imbalance and falls. Visual symptoms are quite prominent and characteristic of PSP. The earliest of these are slowing of vertical saccades (the quick eye movements we use in redirecting our vision), causing difficulty with changing to a new visual target. Other difficulties with eye movement include difficulty opening and closing the eyes and decreased blinking. The decreased blinking, along with a constant raised-eyebrows facial expression, gives the face a fixed stare, characteristic of the disease. The gaze difficulties can lead to problems such as difficulty in making eye contact, difficulty in reading (because of inability to scan lines on a page), and difficulty with eating (because of inability to look down at their food).

Dysarthria (slow or slurred speech) is a very common symptom in PSP. Patients often find it difficult to carry conversations with others because of the delay of their responses and their difficulties with speech pronunciation. Eventually, difficulties with control of oral movements can progress to the point where 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 suggesting abnormal function in the frontal lobes. Cognitive changes consist of 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 perseverance, an inability to switch tasks or change topic. Depression is also common.

In contrast with PSP, PD patients 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.

Progression of progressive supranuclear palsy (PSP)

Balance and walking problems are usually early features of the disease but the illness can present
with cognitive and behavioral changes. Visual and oculomotor problems tend appear early as well. Late in the course of this disease, all these symptoms progress to the point where walking becomes very difficult, if not impossible, eye movement problems get to be more disabling, and cognitive impairment progresses to dementia.

Treatment of progressive supranuclear palsy (PSP)

Currently, there are no effective treatments for PSP. 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 [12].

There are medications, however, that may relieve some of the symptoms. Mostly, these are medications used for typical Parkinson’s disease (PD) [8]. People with PSP do not respond to these agents as well as a person with typical PD.

Lifestyle changes may help alleviate some of the problems associated with PSP. These include use of 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.

If swallowing problems become severe, insertion of a feeding tube directly into the stomach can significantly decrease the risk of pneumonia.

Resources for progressive supranuclear palsy (PSP)

Families and caregivers of PSP patients often have feelings of anger, frustration, depression, guilt and isolation and may be reluctant to share their feelings with others. It is important for caregivers to seek support for these difficulties.

- NINDS Progressive Supranuclear Palsy Information Page [13]
- CurePSP [14] is dedicated to assisting patients and their families, increasing public awareness, educating healthcare professionals and supporting research toward better diagnosis, effective treatments, and eventual cures of progressive supranuclear palsy, corticobasal degeneration [9] and related brain diseases.