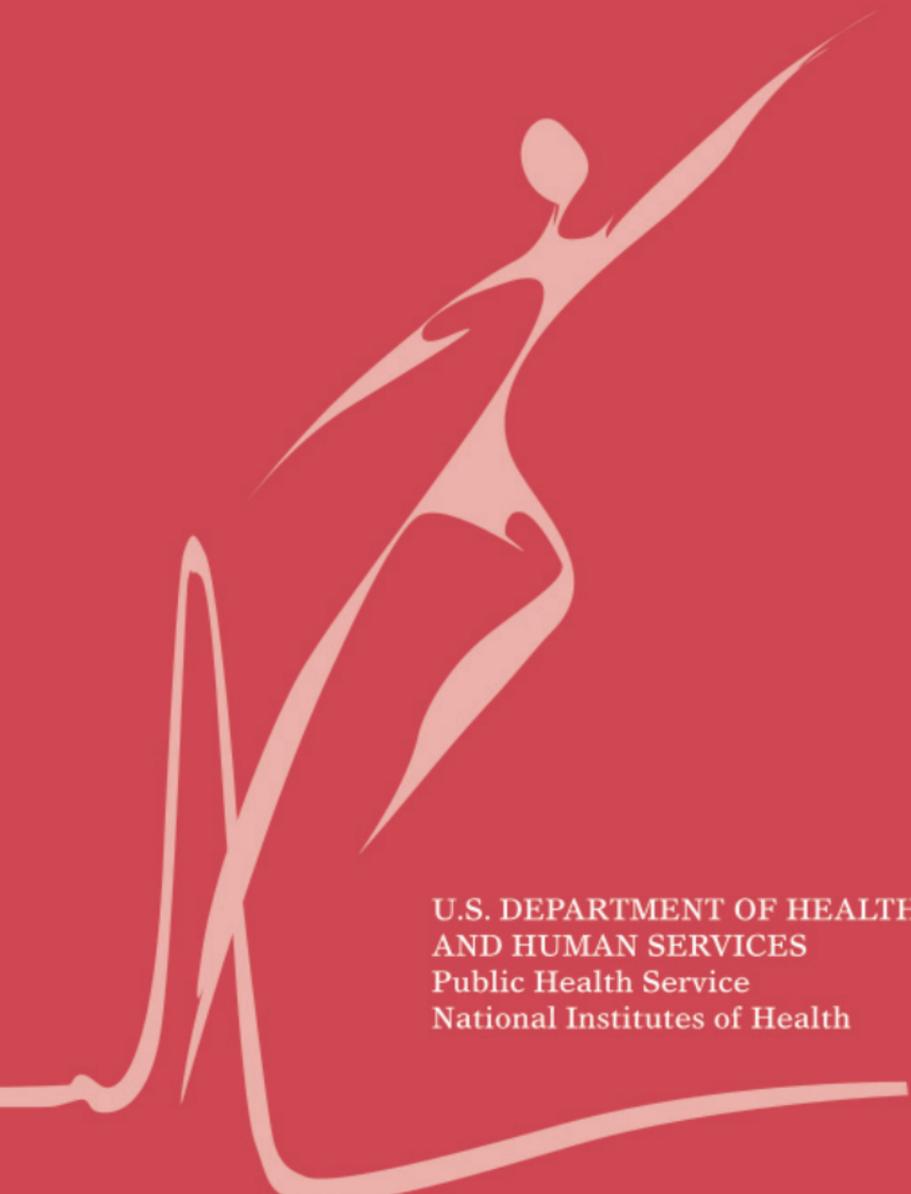


Progressive Supranuclear Palsy



U.S. DEPARTMENT OF HEALTH
AND HUMAN SERVICES
Public Health Service
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Progressive Supranuclear Palsy

What is progressive supranuclear palsy?

Progressive supranuclear palsy (PSP) is a rare brain disorder that causes serious and progressive problems with control of walking or movement by foot (gait) and balance, along with complex eye movement and thinking problems. One of the classic signs of the disease is an inability to aim the eyes properly, which occurs because of lesions in the area of the brain that coordinates eye movements. Some individuals describe this effect as a blurring. Affected individuals often show alterations of mood and behavior, including depression and apathy as well as progressive mild dementia.

The disorder's long name indicates that the disease begins slowly and continues to get worse (*progressive*), and causes weakness (*palsy*) by damaging certain parts of the brain above pea-sized structures called nuclei that control eye movements (*supranuclear*).

PSP was first described as a distinct disorder in 1964, when three scientists published a paper that distinguished the condition from Parkinson's disease. It is sometimes referred to as Steele-Richardson-Olszewski syndrome, reflecting the combined names of the scientists who defined the disorder. Although PSP gets progressively worse, no one dies from PSP itself.

Who gets PSP?

Approximately 20,000 Americans—or one in every 100,000 people over the age of 60—have PSP, making it much less common than Parkinson’s disease, which affects about 500,000 Americans. Affected individuals are usually middle-aged or elderly, and men are affected more often than women. PSP is often difficult to diagnose because its symptoms can be very much like those of other, more common movement disorders, and because some of the most characteristic symptoms may develop late or not at all.

What are the symptoms?

The most frequent first symptom of PSP is a loss of balance while walking. Individuals may have unexplained falls or a stiffness and awkwardness in gait. Sometimes the falls are described by the person experiencing them as attacks of dizziness.

Other common early symptoms are changes in personality such as a loss of interest in ordinary pleasurable activities or increased irritability, cantankerousness, and forgetfulness. Individuals may suddenly laugh or cry for no apparent reason, they may be apathetic, or they may have occasional angry outbursts, also for no apparent reason. The pattern of signs and symptoms the pattern of signs and symptoms can be quite different from person to person.

As the disease progresses, most people will begin to develop a blurring of vision and problems controlling eye movement. In fact, eye problems usually offer the first definitive

clue that PSP is the proper diagnosis. Individuals affected by PSP have trouble voluntarily shifting their gaze downward, and also can have trouble controlling their eyelids. This can lead to involuntary closing of the eyes, prolonged or infrequent blinking, or difficulty in opening the eyes.

Another common visual problem is an inability to maintain eye contact during a conversation. This can give the mistaken impression that the person is hostile or uninterested.

Speech usually becomes slurred and swallowing solid foods or liquids can be difficult.

In rare cases, the symptoms will be more similar to those of Parkinson's disease, and some individuals may even have tremors. This version is often referred to as "Parkinsonian PSP" or PSP-P.

What causes PSP?

The symptoms of PSP are caused by a gradual deterioration of brain cells in a few specific areas of the brain, mainly in the brainstem. One of these areas, the substantia nigra, is also affected in Parkinson's disease, and damage to this region of the brain accounts in part for the motor symptoms that PSP and Parkinson's have in common.

Scientists do not fully know what causes these brain cells to degenerate, but it is known that a hallmark of the disease is the accumulation of an abnormal protein called tau. There is no evidence that PSP is contagious, and genetic factors have not been implicated in most individuals. No ethnic or racial groups have been

affected more often than any others, and PSP is no more likely to occur in some geographic areas than in others.

There are, however, several theories about PSP's cause. One possibility is that an unconventional virus-like agent infects the body and takes years or decades to start producing visible effects. Another possibility is that random genetic mutations, of the kind that occur in all of us all the time, happen in particular cells or certain genes, in just the right combination to injure these cells. A third possibility is that exposure to some unknown chemical in the food, air, or water slowly damages certain vulnerable areas of the brain. This theory stems from a clue found on the Pacific island of Guam, where a common neurological disease occurring only there and on a few neighboring islands shares some of the characteristics of PSP, Alzheimer's disease, Parkinson's disease, and amyotrophic lateral sclerosis (Lou Gehrig's disease). Its cause is thought to be a dietary factor or toxic substance found only in that area.

Another possible cause of PSP is cellular damage caused by free radicals, reactive molecules produced continuously by all cells during normal metabolism. Although the body has built-in mechanisms for clearing free radicals from the system, scientists suspect that, under certain circumstances, free radicals can react with and damage other molecules. A great deal of research is directed at understanding the role of free radical damage in human diseases.

How is PSP diagnosed?

The key to diagnosing PSP is identifying early gait instability and difficulty moving the eyes, the hallmark of the disease, as well as ruling out other similar disorders, some of which are treatable.

Initial complaints in PSP are typically vague and an early diagnosis is always difficult. The primary complaints fall into these categories: 1) symptoms of disequilibrium, such as unsteady walking or abrupt and unexplained falls without loss of consciousness; 2) visual complaints, including blurred vision, difficulties in looking up or down, double vision, light sensitivity, burning eyes, or other eye trouble; 3) slurred speech; and 4) various mental complaints such as slowness of thought, impaired memory, personality changes, and changes in mood.

PSP is often misdiagnosed because some of its symptoms are very much like those of Parkinson's disease, Alzheimer's disease, and more rare neurodegenerative disorders, such as Creutzfeldt-Jakob disease. In fact, PSP is most often misdiagnosed as Parkinson's disease early in the course of the illness. Memory problems and personality changes may also lead a physician to mistake PSP for depression, or even attribute symptoms to some form of dementia.

How is PSP different from Parkinson's disease?

Both PSP and Parkinson's disease cause stiffness, movement difficulties, and clumsiness. However, people with PSP usually stand straight or occasionally even tilt their

head backward (and tend to fall backward), while those with Parkinson's disease usually bend forward. Problems with speech and swallowing are much more common and severe in PSP than in Parkinson's disease, and tend to show up earlier in the course of the disease. Eye movements are abnormal in PSP but close to normal in Parkinson's disease. Both diseases share other features: onset in late middle age, bradykinesia (slow movement), and rigidity of muscles. Tremor, very common in individuals with Parkinson's disease, is rare in PSP. Although individuals with Parkinson's disease markedly benefit from the drug levodopa, people with PSP respond poorly and only transiently to this drug.

What is the prognosis?

PSP gets progressively worse but is not itself directly life-threatening. It does, however, predispose individuals to serious complications such as pneumonia secondary to difficulty in swallowing (dysphagia). The most common complications are choking and pneumonia, head injury, and fractures caused by falls. The most common cause of death is pneumonia. With good attention to medical and nutritional needs, it is possible for most individuals with PSP to live a decade or more after the first symptoms of the disease.

Is there any treatment?

There is currently no effective treatment for PSP, although scientists are searching for better ways to manage the disease. In some individuals the slowness, stiffness, and balance problems of PSP may respond to

antiparkinsonian drugs such as levodopa, but the effect is usually temporary. Excessive eye closing can respond to botulinum injections.

Non-drug treatment for PSP can take many forms. Individuals frequently use weighted walking aids because of their tendency to fall backward. Bifocals or special glasses called prisms are sometimes prescribed for people with PSP to remedy the difficulty of looking down. Formal physical therapy is of no proven benefit in PSP, but certain exercises can be done to keep the joints limber.

A gastrostomy (or a jejunostomy) may be necessary when there are swallowing disturbances or the definite risk of severe choking. This minimally invasive surgical procedure involves the placement of a tube through the skin of the abdomen into the stomach (intestine) for feeding purposes. Deep brain stimulation and other surgical procedures used in individuals with Parkinson's disease have not been proven effective in PSP.

What research is being done?

Studies to improve the diagnosis of PSP have recently been conducted at the National Institute of Neurological Disorders and Stroke (NINDS), a component of the National Institutes of Health (NIH). Experiments to find the cause or causes of PSP are currently under way. Clinical trials are testing medications used for other conditions in PSP, but the results so far have not been very encouraging.

In addition, there is a great deal of ongoing research on Parkinson's and Alzheimer's diseases at the NIH and at university medical

centers and institutes throughout the country. Better understanding of those common related disorders will go a long way toward solving the problem of PSP, just as studying PSP may help shed light on Parkinson's and Alzheimer's diseases.

Where can I get more information?

For more information on neurological disorders or research programs funded by the National Institute of Neurological Disorders and Stroke, contact the Institute's Brain Resources and Information Network (BRAIN) at:

BRAIN

P.O. Box 5801
Bethesda, MD 20824
800-352-9424
www.ninds.nih.gov

Information also is available from the following organizations:

CUREPSP (Foundation for PSP/CBD and Related Brain Diseases)

30 E. Padonia Road, Suite 201
Timonium, MD 21093
410-785-7004
800-457-4777
www.curepsp.org

WE MOVE (Worldwide Education & Awareness for Movement Disorders)

204 West 84th Street
New York, NY 10024
212-875-8312
www.wemove.org



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