Progressive Supranuclear Palsy

Progressive supranuclear palsy (PSP) is a rare brain disorder. It causes serious and permanent problems with control of gait and balance. The symptoms of PSP are caused by a gradual deterioration of brain cells in a few tiny but important places at the base of the brain. This region is called the brainstem.

SYMPTOMS

- The most obvious sign of the disease is an inability to aim the eyes properly. This happens because of lesions in the area of the brain that coordinates eye movements. Some patients describe this effect as a blurring.
- PSP patients often show changes mood and behavior. This includes:
  - Depression.
  - Apathy.
  - Progressive mild dementia.
- The pattern of signs and symptoms can vary greatly from person to person.

DIAGNOSIS

PSP is often misdiagnosed because some of its symptoms are very much like those of:

- Parkinson's disease.
- Alzheimer's disease.
- Rare neurodegenerative disorders, such as Creutzfeldt-Jakob disease.

The key to establishing the diagnosis of PSP is the identification of early gait instability and difficulty moving the eyes, the hallmark of the disease. Also important is ruling out other similar disorders, some of which are treatable. Although PSP gets progressively worse, no one dies from PSP itself.

TREATMENT

- There is currently no effective treatment for PSP. Scientists are searching for better ways to manage the disease.
- In some patients, the slowness, stiffness, and balance problems of PSP may respond to antiparkinsonian agents such as levodopa, or levodopa combined with anticholinergic agents. But the effect is usually temporary.
- The speech, vision, and swallowing difficulties usually do not respond to any drug treatment. Another group of drugs that has been of some modest success in PSP are antidepressant medications. The most commonly used of these drugs are Prozac, Elavil, and Tofranil. The anti-PSP benefit of these drugs seems not to be related to their ability to relieve depression.
- Non-drug treatment for PSP can take many forms. Patients often use weighted walking aids because of their tendency to fall backward.
- Bifocals or special glasses called prisms are sometimes prescribed for PSP patients. They can correct the difficulty of looking down.
- Formal physical therapy is of no proven benefit in PSP. But certain exercises can keep the joints limber.
- A surgical procedure may be needed when there are swallowing disturbances. It is called a gastrostomy. This surgery involves the placement of a tube through the skin of the abdomen into the stomach (*intestine*) for feeding purposes.

WHAT WILL HAPPEN

PSP gets progressively worse. But it is not itself directly life-threatening. It does, however, predispose patients to serious complications. This includes 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. But with good attention to medical and nutritional needs, most PSP patients live well into their 70s and beyond.

RESEARCH BEING DONE

Therapeutic trials with free radical scavengers are being planned for the future. These are agents that can get rid of potentially harmful free radicals. Research is ongoing on Parkinson's and Alzheimer's diseases. Better understanding of those common, related disorders will go a long way toward solving the problem of PSP. Studying PSP may help shed light on Parkinson's and Alzheimer's diseases, too.

Research is continually being done to learn more about this problem.

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