Progressive supranuclear palsy

WHAT IS IT?
Progressive supranuclear palsy (PSP) involves the progressive death of neurons in the brain stem. It results in problems with balance, movement, vision, speech and swallowing. The rate of progression varies considerably among individuals. PSP is often misdiagnosed because its symptoms are similar to those of Parkinson’s and Alzheimer’s diseases. Problems with vision are the hallmark of PSP and differentiate it from similar conditions.

CAUSES
PSP is caused by a death of neurons in the brain stem and basal ganglia. The basal ganglia control balance, movement, vision, speech and swallowing. The precise causes are unknown but progression, especially neurofibrillary tangles deposited in the brain, are similar to the processes of Alzheimer’s disease, although those occur in a different part of the brain.

SYMPTOMS
Symptoms vary considerably from person to person but they include:
- Loss of balance – usually the first symptom. Patients may have unexplained falls that are often reported initially as being caused by dizziness;
- Stiffness of gait;
- Changes in personality such as a loss of interest in everyday activities or irritability;
- Memory loss;
- Blurred vision and difficulty controlling eye movement. Eye problems are the definitive diagnosis of PSP. Patients have difficulty focusing downwards and controlling the eyelids, leading to involuntary blinking and difficulty opening the eyes;
- Difficulty maintaining eye contact;
- Slurred speech;
- Difficulty swallowing.

Diagnosis
There is no blood test or other test that can provide a definitive diagnosis and although an MRI scan can illustrate neurological degeneration in the brain, diagnosis is based on symptoms. Diagnosis can prove difficult as symptoms are similar to those of other conditions. For example, changes in mood and memory problems may lead to an initial diagnosis of Alzheimer’s disease and motor problems lead to confusion with Parkinson’s disease.
The diagnostic pointers to PSP are early gait instability and difficulty moving the eyes.

TREATMENT
At present there is no cure for PSP although certain symptoms can be managed. Motor difficulties may respond to medications such as levodopa, on its own or in combination with anticholinergics.
- Antidepressants such as fluoxetine and amitriptyline can help with mood disturbances.
- Patients may also use walking aids because of their tendency to fall backwards. Bifocals or special glasses called prisms are sometimes prescribed for PSP patients to remedy the difficulty of looking down.
- Gastrostomy is used where swallowing difficulties interfere with nutrition.
- The speech, vision, and swallowing difficulties do not usually respond to any drug treatment.

REFERENCES