What is progressive supranuclear palsy?

PSP is a brain disease in the category of “neurodegenerative” diseases. Alzheimer’s, Parkinson’s and Lou Gehrig diseases are in the same category – where brain cells cumulatively break down for unclear reasons. PSP affects cells that control walking, balance, mobility, vision, speech, swallowing and behavior. Five to six people per 100,000 have PSP, a number similar to that of Lou Gehrig disease, but only about a third of these have received the correct diagnosis. Symptoms begin, on average, when an individual is in the early 60’s, but may start as early as in the 40’s. It is slightly more common in men than women, but PSP has no known geographical, occupational or racial preference.

What are the symptoms of PSP?

The most common symptoms of PSP are:

- Loss of balance
- Changes in personality such as a loss of interest in ordinary pleasurable activities or increased irritability
- Difficulty organizing thoughts, following a plan and maintaining attention.
- Weakness of eye movements, especially in the downward direction. Later, there is often difficulty controlling eyelids, producing a wide-eyed stare or difficulty opening the eyes.
- Slurred speech
- Difficulty swallowing—the reduced ability of throat muscles to seal off the windpipe during swallowing often results in aspiration pneumonia—the most common cause of death in PSP patients.
- Constipation and difficulty controlling the bladder

What eventually happens to someone with PSP?

The course of PSP, at least until better treatment or a cure is found, is of slowly worsening symptoms until the person is quite disabled and requires help with walking and most daily activities. At that point, they are vulnerable to such complications as pneumonia, injuries from falls, bladder infections, bedsores and blood clots. The
average person has the initial symptoms at age 63 and survives an average of about seven years. This compares with an average of 19 years that the general population lives after age 63.

What causes PSP?

PSP is caused by a gradual deterioration and death of brain cells in many small parts of the brain, especially in the brainstem, the part that controls such actions as eye movement, speech and swallowing. We do not yet know the cause of that deterioration, but most scientists think that it’s a combination of genetic and environmental factors.

Is PSP genetic or hereditary?

PSP very rarely runs in families. Less than one in 100 persons with PSP knows of even one other family member with PSP. However, some research has found that close family members of people with PSP are more likely than other people to have various other types of neurological symptoms. A specific gene variant is more common in PSP patients than in the rest of the population. About 95% of people with PSP and 65% of the rest of the population have this gene variant. A handful of other gene variants are over-represented in PSP, although to a lesser degree.

How is PSP similar to and different from Parkinson’s disease?

Both PSP and Parkinson’s disease cause stiffness, slowness, and clumsiness, a combination called parkinsonism. This is why PSP may be difficult to distinguish from Parkinson’s disease in the early stages. In PSP, there is also severe damage to certain brain cells which remain intact in Parkinson’s. Medications that may be effective for Parkinson’s disease are unfortunately of much less benefit in PSP.

Does PSP lead to dementia like that in Alzheimer’s disease?

Most people with PSP do eventually develop some degree of mental impairment. Some are mislabeled as having Alzheimer’s disease although PSP lacks the profound memory impairment of AD. This is similar to the situation in Parkinson’s disease. In PSP, dementia is characterized by slowed and disorganized thought and difficulty in putting together several different ideas into a new idea or plan. People with PSP also seem to lose interest in their surroundings, creating the impression of loss of thinking ability.

Is there treatment for PSP?

PSP currently has no effective treatment or medication, although some drugs may have a temporary, modest benefit.

What research is being done?

Research is proceeding in the areas of genetics, epidemiology, treatment trials, and molecular studies:

- CurePSP’s genetics program has searched the entire genome for genes related to PSP and corticobasal degeneration (CBD). The results, which showed at least four genes not previously suspected of a relation to these diseases, will be
• CurePSP is working with researchers at the University of Louisville to study environmental, occupational, and genetic risk factors associated with PSP.
• CurePSP and other funding agencies are presently sponsoring exciting and promising research into many aspects of PSP, including ways to prevent the brain cells from dying and treatment of the symptoms with drugs, magnetic stimulation, and deep-brain electrical stimulation.
• Two drug companies are presently running large, formal trials of new medications designed to slow progression of PSP.
• CurePSP holds an annual international research symposium attended by scientists, researchers, and physicians from around the world to share their findings and discuss new research.