

Progressive Supranuclear Palsy (PSP)

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July 2012

Progressive Supranuclear Palsy (PSP) is a rare neurological disease which affects parts of the brain involved in balance and walking, eye movements and speech. It affects around 7 people in 100,000 over the age of 40, does not usually run in families and can affect both men and women. It was previously called Steel-Richardson-Olszewski syndrome after the doctors who first described the condition in 1964. It is often included in a group of conditions which can have similar symptoms, termed “atypical parkinsonism” which includes Multiple System Atrophy and Corticobasal Degeneration Syndrome.

Symptoms can vary from person to person, but most people experience progressive difficulty with walking and balance that leads to falls, slurred speech, difficulty with swallowing, and eye movement problems that cause difficulty with reading. Many people have changes in their thinking with slowed thoughts and difficulty processing information though they remain aware of their environment and those around them. Some people have changes in their personality, in particular with less interest in activities.

The cause for PSP is not known. It is associated with the build up of “tau” protein in the brain. Although tau is a usual part of normal brain cells, in PSP the tau proteins collect into clumps called “tangles”. How these damage the brain cells is not known, but areas of the brain affected can shrink over time. Although these changes can be seen on MRI scans of the brain late in disease, scans performed earlier cannot demonstrate this change and are often reported as “normal” even when clinical symptoms are present.

There are two main forms of PSP, “Richardson’s syndrome” presents with early falls and eye movement problems. Some people with PSP can have symptoms similar to Parkinson’s disease in the beginning with slowed movements of their limbs, stiffness or tremor. Unlike Parkinson’s disease these symptoms don’t improve very much with medications. This form of the disease is known as “PSP-parkinsonism” and people with this presentation are often diagnosed initially with Parkinson’s disease. There is currently no diagnostic test which is able to separate PSP from Parkinson’s disease so accurate diagnosis relies on clinician experience. A third form, Primary Akinesia with Gait Freezing is very rare and progresses very slowly, producing problems with walking and falls.

Symptoms of PSP worsen over time, but the rate of progression and severity varies between people. People affected by PSP are at risk of serious complications such as pneumonia and injuries from falls, which can be fatal. The average life expectancy after diagnosis is seven years, though some people will live much longer than this.

There is no treatment that cures PSP or slows the progression of the disease. However there are some treatments that can help the symptoms and improve quality of life. A management team that involves the primary care giver, general practitioner, neurologists and an experienced allied health care team is important to optimise independence and maintain function whenever possible. A neurologist can assist in ensuring the diagnosis is correct and if medications can help with any of the symptoms. For some people dopamine replacement medication used in Parkinson’s disease (Sinemet, Madopar and Kinson) may assist with limb stiffness and slow movements. Amantadine may help freezing of gait though it can produce side effects including constipation and confusion and needs to be monitored carefully. Some

people have problems with involuntary closure of the eyelids that may respond to botulinum toxin injections. Unfortunately surgical approaches (deep brain stimulation surgery) used in Parkinson's disease are rarely useful in PSP. A speech pathologist can assist with swallowing problems and communication aids if the voice becomes soft or difficult to understand. Falls are a common problem and physiotherapy input is important to help maintain safe mobility as long as possible. Massage and heat packs can help with muscle stiffness. Eye movement problems affect quality of life as they interfere with the capacity to read. As they are a progressive problem obtaining new glasses rarely helps, and using other visual aids such as large print, magnifying sheets, devices to hold books higher into the line of sight and using talking books or computer screens with adjustable text size are often more successful.

Further information about PSP can be obtained from medical websites and support groups.

PSP- Australia has many useful documents to help understand the condition and cope with symptoms, and contact information for people who have experience caring for people with PSP

www.psp-australia.org.au

Other useful sites include:

USA PSP support group www.curepsp.org

Movement Disorder Society www.movementdisorders.org
www.wemove.org

Support for carers can also be found at: www.carersaustralia.com.au