

What is Progressive Supranuclear Palsy (PSP)?

This Help Sheet gives a simple introduction to the condition known as Progressive Supranuclear Palsy (PSP for short). It explains its causes and symptoms, and gives information about who the condition affects and how it is diagnosed. It also tells you where you can go for further information if you or someone you know has PSP.

PSP is a rare neurological condition affecting the part of the brain that controls walking, eye movements, balance, speech and swallowing. PSP has a wide range of symptoms, but few people experience them all and symptoms vary from person to person. It is a progressive condition, meaning that the symptoms continue to get worse over time, however the severity and rate of progression varies widely between individuals. Symptoms can fluctuate from day to day or even from hour to hour.

There is no known cure for PSP and we do not know of any way of preventing it from occurring. Treatments, therapies and strategies are available to help people with PSP and their families and carers to make the most of living with the condition. The information here is a guide; it is important to discuss all symptoms and management strategies with your neurologist and other health professionals.

‘P’	Progressive	means the symptoms get worse over time
‘S’	Supranuclear	refers to the part of the brain that is affected
‘P’	Palsy	means a weakness or paralysis in part of the body.

PSP occurs in only about five or six people per 100 000 (this is about the same as motor neurone disease). It is a difficult condition for doctors to diagnose, so actual numbers of people with PSP may be higher. Approximately 180 Australians will develop the first signs of PSP each year and currently around 1300 Australians are living with PSP. Only 22 per cent of people with PSP are given the correct diagnosis in the first 2 years, with many receiving an initial diagnosis of Parkinson’s disease due to the similarity of symptoms.

PSP was first described and named by doctors in 1964. Before then people with the condition were thought to have Parkinson’s disease. In the past PSP was sometimes called ‘Steele-Richardson-Olszewski Syndrome’. In its different manifestations it may be called ‘Richardson’s disease’ after the doctors who first described the condition, PSP-parkinsonism (PSP-P) or pure akinesia with gait freezing.

What causes PSP?

PSP is associated with an over-production of a protein called ‘tau’ in certain areas of the brain. Clumps of tau (called ‘tangles’) build up and these are believed to cause damage to nerve cells. We do not know what causes the over-production or the build up of tau, however research is currently underway. Researchers have found no evidence of any links between PSP and environmental factors such as exposure to toxins or chemicals. There is ongoing research going on in Australia, and study

participants are always needed. Parkinson's Australia or your treating neurologist may know about these studies.

Who gets PSP?

People with PSP usually start to have symptoms between the ages of 60 and 70. PSP can affect people from all nationalities and lifestyles and many of those affected have always lived a healthy lifestyle.

Is PSP inherited or passed on through families?

PSP does not run in families. However a common genetic type has been found to be more common in people with PSP than the rest of the population. In other words, there may be some genetic susceptibility that makes some people more at risk than others, but PSP rarely affects more than one person in a family.

Can it be treated?

Treatments and therapies are available to help manage symptoms of PSP, however there is no cure and we do not know of any way of slowing its progression.

It is important that people with PSP see a neurologist, preferably one with expertise in movement disorders such as PSP and Parkinson's. Depending on symptoms, it is also important to seek the advice of other health professionals with expertise in the condition, such as a physiotherapist, occupational therapist, speech pathologist (speech therapist) or counsellor. Parkinson's Australia can provide information about Neurologists and other health professionals in your area.

For further information about treatments, therapies and strategies to manage the symptoms of PSP see our series of Help Sheets on managing symptoms.

Is PSP fatal?

PSP cannot be cured, and so everyone with it experiences deterioration prior to death. Symptoms become progressively worse over time and management strategies may fail to be effective. People with PSP commonly contract serious respiratory problems in the later stages of the condition due to being immobile, and these problems often contribute to death. The **average** life expectancy after diagnosis is approximately seven years which means that half of people with PSP will live longer (even up to 15 years) and half will live shorter (even as short as 3 years). There is significant variation from person to person and, with good symptom control, some people live significantly longer.

Research into PSP is currently underway, giving us hope for the future, particularly in relation to more effective management of symptoms.

What are the Symptoms of PSP?

PSP affects different people in different ways. It has a wide range of symptoms, few people experience all symptoms and the severity and progression of symptoms varies

widely between individuals. Symptoms can also fluctuate from day to day or even from hour to hour.

The most significant problems in the early stages of PSP are usually related to walking. Symptoms get worse over time and can start to have a very significant impact after five to six years. Problems with vision tend to occur on average three to five years after the walking problems.

Early symptoms of PSP may include:

- Problems with walking (or 'gait'). These may include stiffness, awkwardness or problems with balance and unexplained falls, particularly backwards.
- A feeling of dizziness.
- Slow movements.
- Problems with eyesight, which may be described as tunnel vision, blurring, double-vision, dislike of bright lights and/or difficulty focussing, dry eyes or discomfort.
- Problems with thinking.
- Changes in personality, such as loss of interest in activities that were previously enjoyed, irritability and/or depression.
- Slurring of speech.
- Clumsiness.
- Mild shaking of hands.
- Small handwriting.
- Facial stiffness
- Depression.

Later symptoms of PSP may include:

- Worsening of movement problems making walking very difficult or impossible and some people become unable to move voluntarily.
- Recurrent falls that can lead to bruises and fractures.
- Eye problems that can include involuntary closure of eyelids, difficulty looking up or down and loss of ability to focus.
- Difficulties with swallowing that can lead to weight loss and bouts of choking
- Communication difficulties
- Increased difficulty with thinking

There are a range of treatments and therapies that can help to manage symptoms and achieve the best possible level of independence and quality of life. For more information, see our series of Help sheets on 'Managing Symptoms' or contact Parkinson's Australia.

How is it Diagnosed?

PSP is difficult to diagnose. There are no blood tests or brain scans that can diagnose it, although tests and scans are commonly used to rule-out other conditions. It is common for people with early symptoms of PSP to be misdiagnosed with Parkinson's

due to similar symptoms, such as slowness of movement. Two of the key signs indicating the need to review a diagnosis of Parkinson's and consider PSP are a limited response to Parkinson's medication and changes in the movement of the eyes.

Some people with PSP are also misdiagnosed with dementia due to problems they may experience with their thinking.

Because PSP is rare, many doctors are not even aware of the condition and do not know what symptoms to look for, although this situation is improving. It is important to see a neurologist, who is a doctor who specialises in brain conditions. People with PSP should visit their neurologist regularly for ongoing treatment and advice. If possible, see a neurologist with expertise in movement disorders, as they are more likely to be up to date with the latest advances in treatment and management.

Coming to terms with a diagnosis of PSP

Most people will experience a range of feelings after being diagnosed with PSP. Family members and the person diagnosed may feel relief at having a name for the difficulties they have been experiencing, combined with anger, grief, denial or concern. Getting information and support can help you to adjust to the diagnosis and begin to make changes that will help with the management of the condition on a day to day basis. Remember, you are not alone and help is available.

Where can I go for further information and support?

PSP is a complex and challenging condition. It is important to get help and to communicate your needs. Parkinson's Australia provides information and support for people with PSP and their families and carers. We also provide a free telephone interpreter service for people who speak a language other than English.

There are a range of services available to assist people with daily living and to help members of the community to make the most of living with an illness or disability, including:

- Information and emotional support – either over the phone or in person
- Therapies and resources to maintain independence
- Financial support and assistance
- Support groups where you can share your experiences and learn from others
- Help with personal care, such as bathing, dressing and eating
- Home delivered meals and other food services
- Home help for assistance with housework, laundry and shopping.
- Help for carers, including counselling, advice and respite to help you take a break from caring

For further information about services that can help, contact Parkinson's Australia or your local council. The **Commonwealth Respite and Carelink Centre on 1800 052 222** can also provide information about services in your local area.

Please also refer to our 'Living with PSP' Help Sheet and 'Where to Go for Help'.