

1.1 Introduction:

What is Progressive Supranuclear Palsy?

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 suggests where you can go for further information and support if you or someone you know has PSP. The information here is presented as a guide; it is important to discuss all symptoms and management strategies with your neurologist and other health professionals.

PSP is a rare neurological condition affecting the parts of the brain that control 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 tend to worsen over time. However the severity and rate of symptom progression varies widely.

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- '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 a part of the body

At the moment there are no treatments that are known to cure 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.

PSP occurs in only about five or six people per 100 000 (this is similar to the numbers of people with 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.



*For PSP information and support phone
Parkinson's Australia on 1800 644 189*





PSP was first described and named by doctors in 1964. Before then people with the condition were thought to have Parkinson's disease. PSP is sometimes called 'Steele-Richardson-Olszewski syndrome' after the doctors who first described the condition. Other terms sometimes used for PSP are 'Richardson's disease', 'PSP-parkinsonism' or (PSP-P) and '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, though research is currently underway. Researchers have found no evidence of any links between PSP and environmental factors such as exposure to toxins or chemicals. Research is ongoing in Australia and overseas. Contact Parkinson's Australia or your neurologist for further information about participating in PSP studies.

Who gets PSP?

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

Is PSP inherited or passed on through families?

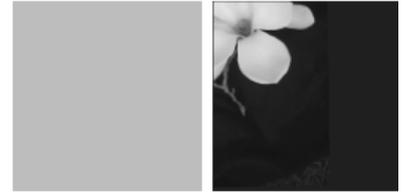
PSP does not run in families. However, a common genetic type has been found to be more common in people with PSP than the rest of the population. In other words, there may be some genetic susceptibility that makes some people more at risk than others, but PSP rarely affects more than one person in a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



Is PSP fatal?

PSP can not be cured, so everyone with PSP experiences deterioration prior to death. As the condition progresses, people with PSP are at risk of developing serious complications, such as pneumonia or choking caused by swallowing difficulties or head injury caused by falls. These complications can be fatal.

The rate of progression with PSP varies significantly from person to person. The **average** life expectancy after diagnosis is approximately seven years, which means that half the people with PSP will live longer, some up to 15 years, and half will live shorter, even as short as 3 years.

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. These get worse over time and can start to have a significant impact after five to six years.

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.
- Facial stiffness.
- Problems with eyesight, which may be described as tunnel vision, blurring, double-vision, dislike of bright lights and/or difficulty focussing.
- Problems with thinking.
- Changes in personality, such as loss of interest in activities that were previously enjoyed, irritability, depression and/or heightened emotional responses.
- Slurring of speech.
- Clumsiness.
- Mild shaking of hands.
- Small handwriting.



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, down or to the side 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 PSP Australia's series of Help Sheets on 'Managing Symptoms' or contact Parkinson's Australia.

How is PSP diagnosed?

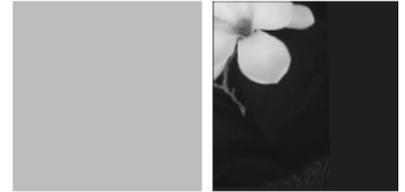
PSP is difficult to diagnose. There are no blood tests or brain scans that can diagnose it, although tests and scans are commonly used to rule-out other conditions. It is common for people with early symptoms of PSP to be misdiagnosed with Parkinson's due to similar symptoms, such as slowness of movement.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

People may experience a range of feelings after being diagnosed with PSP. Most people experience a feeling of grief, which may include fear, sadness, denial, anger or concern for the future. Some people may experience a feeling of relief at having a name for the difficulties



they have been experiencing after searching so long for answers. It is important to have a safe space in which to work through these feelings of grief and be aware that those around you will be experiencing the same range of emotions. Speaking

to a counsellor can help you to come to terms with your feelings and make meaning of the situation. Getting information and support can help you to adjust to the diagnosis and begin to make changes that will help with the daily management of the condition. Remember, you are not alone and help is available. The best place to start is by contacting Parkinson's Australia.

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 information, counselling, advice and respite to help them take a break from caring.

Help Sheet 1.1

Introduction
What is PSP?

Page 5

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 PSP Australia's Help Sheets 1.2: *Living with PSP* and 3.5: *Where Can I Go for Help*.



For further information about living with PSP, including the full range of PSP Australia Help Sheets, visit: www.psp-australia.org.au

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