

Progressive Supranuclear Palsy

Progressive Supranuclear Palsy (PSP or Steele-Richardson-Olsewski syndrome) is a rapidly degenerative neurological disorder. The main symptoms of PSP typically include vertical gaze palsy (difficulty looking up or down), postural imbalance and falls, dysphagia (difficulty eating or drinking) and/or dysarthria (speech disorder).

Prevalence:

- > The prevalence of PSP is 6.5 per 100,000 (similar to Motor Neuron Disease).
- > Average age of onset is 60 to 65 years.
- > Average life expectancy is six to seven years from diagnosis.
- > **Bowel and bladder difficulties** - constipation, incontinence.
- > **Sleep disturbances** - insomnia, REM sleep behaviour disorder, excessive daytime sleepiness, restless legs syndrome.

Cause:

- > PSP belongs to the family of tauopathies. Abnormalities in the protein tau lead to damage in both cortical and subcortical areas of the brain.

Diagnosis:

- > PSP is difficult to diagnose.
- > Patients may remain undiagnosed for approximately half of the natural history of their disease.
- > Patients with PSP present with parkinsonian symptoms of slowness of movement, difficulty initiating movement, and rigidity with or without resting tremor.
- > Patients with PSP differ from Parkinson's Disease with rapid progression of their disorder, absent/poor/waning response to dopaminergic medications, early instability or falls, early dysphagia and/or dysarthria.

Symptoms may include:

- > **Mobility difficulties** - falls (often backwards), balance, postural instability, slow movements, rigidity, dystonia.
- > **Vision difficulties** - vertical gaze palsy (may not be present in early stages), photophobia, diplopia (blurred vision), involuntary eyelid spasms, dry eyes.
- > **Swallowing difficulties** - dysphagia, impulsive feeding, excess saliva/drooling, needing PEG Feeding (late stages).
- > **Communication difficulties** - speech problems, occasionally stuttering.
- > **Cognition/mood difficulties** - apathy, depression, anxiety, disinhibition, irritability, emotional lability, executive dysfunction, slowness of thought.

Possible cures for PSP are being researched. Currently, there are a number of symptomatic therapies available which can help to improve quality of life.

Medications:

- > Appropriate medications for symptomatic management continue to evolve and it may be beneficial to consult with a Neurologist and/or Geriatrician.
- > High levodopa in some circumstances has limited effect.
- > Atropine eye drops or atrovent nasal spray administered under the tongue may assist in drying excess secretions/assist with drooling, medications to assist with constipation.
- > Medications may assist with constipation.

Non-pharmacological Treatments:

- > Constipation: daily movement, sufficient fluid and fibre intake.

Management of symptoms:

Movement Disorder Clinics and Parkinson's Clinics

- > Some regions provide specialist services to help manage PSP.

Psychology/counselling:

- > Patients and families may benefit from a mental health care plan through their GP to access services from a mental health practitioner.
- > Counselling is also available to anyone affected by PSP through Parkinson's SA **1800 644 189**.

Eye Specialist/Ophthalmologist

- > Can assist if eye and vision symptoms are

particularly severe.

Independent Living Centre of South Australia

- > Provides information and advice about resources, equipment or aids that can assist **(08) 8266 5260**.

SA Continence Resource Centre

- > Provides information, education and advice regarding continence issues, pads and appliances **1300 885 886**.

Continence Nurse Specialist

- > Continence nurses may be available through hospital/community services and can provide information and education.

Accessing Allied Health Care

- > Patients with PSP may have rapid and progressive changes to their mobility and swallowing placing them at high risk of falls/aspiration pneumonia.
- > They may benefit from ongoing monitoring of their Allied Health needs and for those with functional goals, short periods of intensive rehabilitation.
- > Allied Health may be accessed through Day Rehabilitation through public and private hospitals, Day Therapy through residential care facilities, private clinicians, community services provided through a range of aged care organisations, Domiciliary Care (over 65 years) and Disability SA (under 65 years), and local community health centres in regional areas.

Physiotherapy

- > Can provide patient/carer education, management of falls risk, exercise, carer assistive techniques, equipment.

Occupational Therapy

- > Can provide patient/carer education, management of falls risk, home modification, equipment, strategies for vision difficulties.

Dietetics

- > Can provide patient/carer education, management of nutrition, prevention of weight loss, input regarding need for PEG placement.

Speech Pathology

- > Can provide patient/carer education, management of dysphagia, input regarding need for PEG placement, communication strategies, and equipment for communication.

Swallowing Disorders Clinic RGH

- > Outpatient access to objective assessment of swallowing including Modified Barium Swallow and Fiberoptic Endoscopic Evaluation of Swallowing. GP Referral Liaison Services **(08) 8275 1837**.

Parkinson's SA

- > Parkinson's SA provides support and information to anyone affected by PSP **1800 644 189**.

Technical Aid to the Disabled (SA)

- > Can offer one-off solutions to help people with disabilities.
- > Their technical volunteers design, make or modify equipment not readily available to suit individual needs.

Useful References/Associations:

- > Williams DR, Litvan I. Parkinsonian syndromes. Continuum (Minneapolis, Minn). 2013 Oct;19 (5 Movement Disorders):1189-212.
- > PSP Australia website: www.psp-australia.org.au/
- > CurePSP USA website: www.psp.org/
- > PSP Association UK website: www.pspassociation.org.uk/

For more information

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