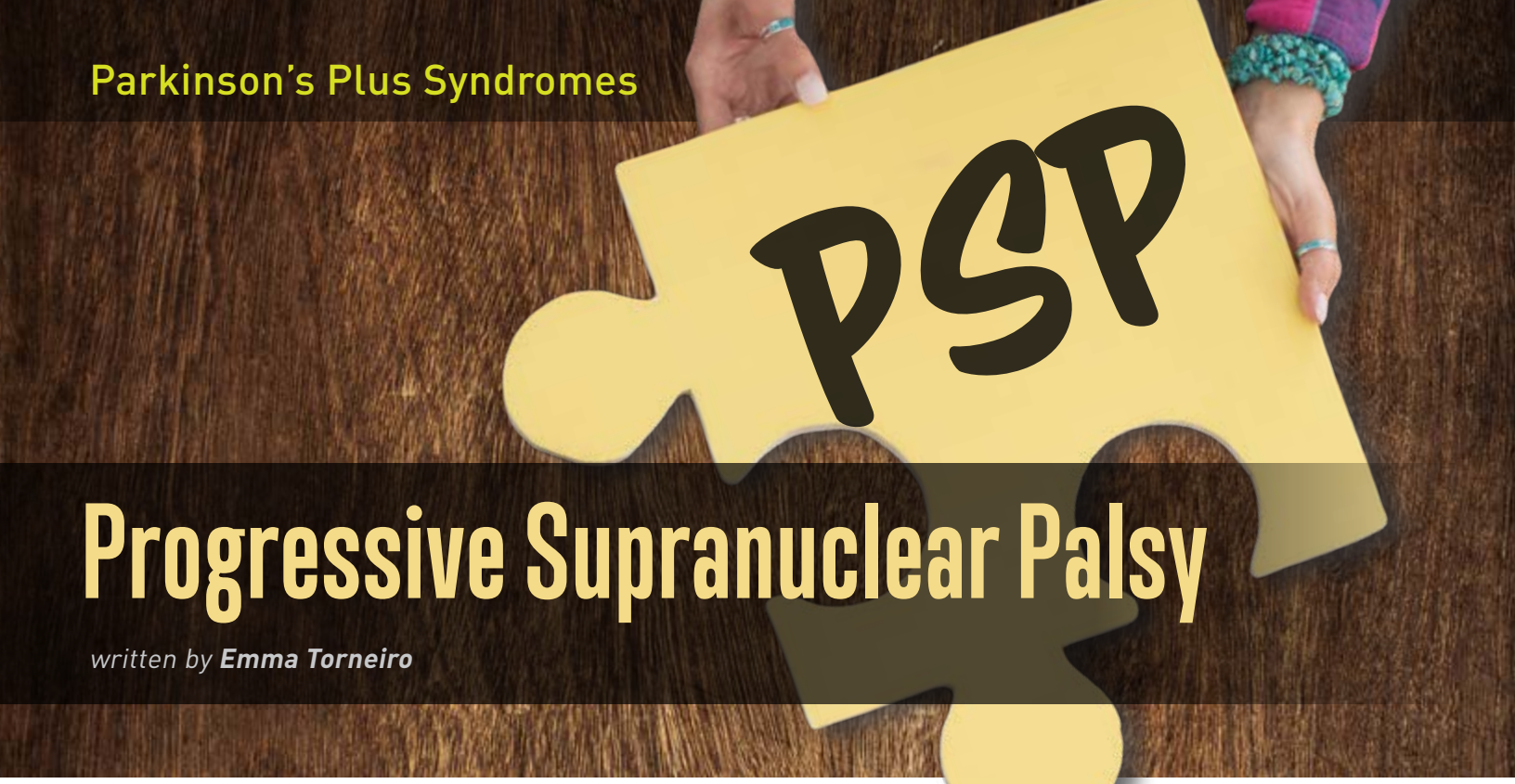

Progressive Supranuclear Palsy

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Progressive Supranuclear Palsy (PSP) is a rare neurodegenerative tauopathy condition resulting from abnormal buildup of tau protein in the brain causing cell death[1].

What do these medical terms really mean? In PSP, the cell degeneration and symptoms are caused by the malfunctioning of clumped tau protein (tauopathy) in the brain. The term progressive refers to the development over time, supranuclear refers to the vision issues and the areas of the brain impacted, and palsy refers to weakness or paralysis of muscles/body[2]. The motor and cognitive symptoms associated with PSP are connected to the deterioration of cells within the substania nigra responsible for movement, thinking, and coordination. The vision symptoms of PSP results from impairments to the facial muscles responsible for controlling eye movements[2].

The condition was initially titled Steele-Richardson-Olszewski syndrome in 1963 after the Canadian physicians who defined it[3]. The cause of malfunctioning tau protein and cell death in PSP is currently unknown, however research has furthered our understanding of the processes within the brain. The tau protein, which appears in all our brains, is primarily responsible for holding structures called microtubules together and transporting messages of movement between our brain and skeletal system. The microtubules are responsible for cell growth, movement, and cellular transport. In PSP, the tau protein clumps into knots called neurofibrillary tangles and the microtubules separate, leading to the dysfunction of the protein[3]. This means that clumps of tau protein are floating within the brain, leading to the brain being unable to send accurate messages for movement to the body and being unable to promote cell growth. The initial clump of tau protein creates a blueprint for future tau proteins and continues the chronic cycle of clumping and degeneration[3].

PSP is categorized into two main types: Richardson's Syndrome and PSP-Parkinsonism (PSP-P)[3]. The Richardson's Syndrome form primarily involves balance and gait problems. The form can also involve issues with speech, cognition, and vision. The secondary form, PSP-P, resembles Parkinson disease (PD) with more impactful tremor and followed by the Richardson type symptoms. The condition differs from typical Parkinson's through frequent, unexplained falls and difficulties with vision occurring 2-3 years after diagnosis[3]. The second difference between PD and PSP is the number of systems in the body being impacted. For example, PD only involves the dopamine systems within the brain, but PSP involves deficiencies in multiple systems with increased damage to brain cells[3].

The diagnostic process for PSP involves an examination of symptoms and may include brain imaging scans (e.g., MRI for shrinkage or PET for changes in brain). For many, the diagnosis process begins by investigating the loss of balance while walking and increased falls[3]. Due to the scans being unable to differentiate between conditions and the Parkinsonian symptoms, it is common to initially receive a Parkinson diagnosis. The process of diagnosing generally does not include examinations of familial medical history, as PSP is rarely genetic or hereditary.

The Facts

- The average age of onset is 45-70
- Life expectancy is 7-10 years following symptom onset
- Prevalence as the most common form of Parkinson's Plus Syndromes is 5-7 per 100,000

Motor Symptoms

- Parkinsonian symptoms
- Difficulties with Gait
  - Loss of balance and walking challenges
  - Freezing
  - Tendency to fall backwards
- Axial Rigidity (*straight posture or tilting head backwards*)
- Unilateral, action tremor known as PSP tremor[3]
- Dystonia

Vision Symptoms

- Blepharospasm (*involuntary closing of eyelids or increase in blinking*)
- Eye Apraxia (*difficulty coordinating vertical eye movements*)
- Photophobia (*sensitivity to bright lights*)
- Inability to focus eyes resulting from palsy in eye muscles
- Reduction in blinking, leading to dry eyes and loss of clarity from surface damage
- Loss of corneal sensitivity, leading to progressive damage of eye and increased difficulties blinking
- Difficulty shifting vision downwards, creating difficulty in daily living tasks (*e.g., eating, reading, driving*)
- Difficulty maintaining eye contact during interactions or conversations
- Tunnel Vision
- Double Vision
- Square Wave Jerks (*rapid, involuntary vertical eye movements*)[3]
- Supranuclear or Vertical Gaze Palsy

Speech Symptoms

- Dysarthria (*difficulty with speech*)
- Slurred speech
- Dysphagia (*difficulty swallowing*) with solids and liquids from incoordination and weakened muscles
- Hypophonia (*soft speech*)
- Aphasia
- Spastic speech (*inability to articulate or irregular speech patterns*)
- Ataxic speech (*speech has qualities of "drunken speech"*)[3]

Cognitive Symptoms

- Dementia
- Personality Changes (*e.g., lack of interest in activities previously enjoyed or increased irritability*)
- Slowness in mental processing, which creates challenging engaging in conversation due to delayed responses
- Inability to control emotions and expression of emotions (*laughing or crying at inappropriate times*)
- Difficulties with Impulse Control
- Difficulties with Executive Functioning (*organizing information, understanding instructions, and decision-making processes*)

Treatment Options

There is no specific medication to treat PSP or slow the progression of symptoms, therefore treatments target daily management of symptoms. The Parkinson medication, Levodopa Carbidopa, prescribed in a higher dose may initially improve the motor functions of PSP-P with benefits being limited to 2-3 years after diagnosis[3,4]. The secondary Parkinson medication that could provide limited relief for gait challenges is Amantadine[3]. The healthcare team can also prescribe additional medications (*e.g., depression, anxiety, dystonia*) and may provide recommendations for the vision symptoms of PSP (*e.g., Yoked Vertical or Horizontal Prism lenses or artificial tears*[5]).

The main treatment options for PSP focus on using therapies for management of symptoms and improvements to quality of life. The recommended therapies include physiotherapy for mobility and balance, occupational therapy for assessment of mobility aids, and speech therapy for challenges with speech, swallowing, and communication. The use of a dietitian is beneficial for help with proper nutrition and finding easy to swallow diets. Finally, psychologist or social workers can provide support for the non-motor symptoms associated with the diagnosis.

Resources

1 *Progressive Supranuclear Palsy. (n.d.). National Organization of Rare Disorders (NORD). https://rarediseases.org/rare-diseases/progressive-supranuclear-palsy/*

2 *Progressive Supranuclear Palsy. (2020, October 23). Cleveland Clinic. Retrieved from https://my.clevelandclinic.org/health/articles/6096-progressive-supranuclear-palsy*

3 *Golbe, L.I. (2021, July). PSP: Some Answers. CurePSP. https://www.psp.org/wp-content/uploads/2021/08/2021-PSP-SOME-ANSWERS-BROCH_web.pdf*

4 *Progressive Supranuclear Palsy. (n.d.). Mayo Clinic. https://www.mayoclinic.org/diseases-conditions/progressive-supranuclear-palsy/symptoms-causes/syc-20355659*

5 *Haines, S. (2021, May 21). Neuro-ophthalmology [Conference Presentation]. 2021 CurePSP Virtual Family Spring Conference, San Francisco, CA, United States. https://youtu.be/V4yXp_ZdwcI*