NANOS Patient Brochure

Progressive Supranucler Palsy

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Progressive Supranuclear Palsy (PSP) is a rare progressive neurodegenerative disorder that affects certain parts of the brain, resulting in difficulty with balance, walking, swallowing, and vision. PSP patients often report difficulties reading, double vision, dry gritty eyes and light sensitivity. PSP often mimics the symptoms of Parkinson's disease until later in the course when other features emerge. A small percentage of patients with PSP improve with typical Parkinson's disease medication early in the disease. Unfortunately, there is no cure, however, treatment of symptoms can improve the patient's quality of life.

Anatomy:

The brain's ability to control body and eye movements is based on a complex network of nerves. Functions such as reading, smiling, swallowing, and walking involve various parts of the brain, including the cortex (outer layers), which communicates with the basal ganglia (motor regulators) to start, stop, and modulate these movements; several other brain areas, including the midbrain, substantia nigra, globus pallidus, subthalmic nucleus, and the dentate nucleus, play an important role in generating normal eye movements. The signals for body and eye movement are then sent to the nuclei of the brainstem, and then to the muscles that carry out the movements. PSP causes a disruption in these pathways including those above the nuclei ("supranuclear") causing abnormal movements, which may be absent, slow, or over active.

Physiology:

PSP is caused by a build up of a normal protein called tau; the reason for this abnormal build up is currently unknown. The build up occurs in particular places within the brain, interfering with normal nerve cell function and causing the symptoms of PSP.

Symptoms:

Patients with PSP often develop symptoms in their 60's; frequent early symptoms include imbalance and unexplained falls. Within the several years, most patients experience a change in their voice, slowness of movements, difficulties with vision, and mild memory alterations. The voice of PSP patients can softer, slower, and lose the normal higher and lower infections of speech. Movements of the arms and legs take longer time to start; the face can appear frozen with limited expression, and decreased blink rate. Patients may develop difficulty reading, limitation of eye movements, double vision, light sensitivity, and dry eye symptoms such as tearing, burning or a gritty feel. Patients often develop difficulty swallowing, and may choke on liquids more easily than solids. Patients may cry or laugh more easily, become forgetful, exhibit mild personality changes, or lose interest in things once pleasurable.

Signs:

Signs that others may notice or are observed by the doctor include: postural instability or imbalance with a tendency to fall, stiffness of the neck and/or trunk, change in voice (dysarthria), slowed movements (bradykinesia), limited range of facial expression (masked facies), tendency toward sustained brow crease or forehead wrinkle (procerus sign), decreased blink rate, lid retraction/stare, involuntary sustained eyelid closure with slowed eyelid opening, “darting” eye movements when viewing a target (square wave jerks), slowed an incomplete voluntarily eye movements up or down (improved eye movement extent may occur with moving the patient’s head while viewing a stationary target), slowing of normal rapid eye movements, particularly vertically, misalignment of the eyes, difficulty following moving stripes on a rotating drum (abnormal optokinetic nystagmus response), and memory/cognition difficulty.

Diagnosis:

There are no blood tests to confirm the diagnosis of PSP. Therefore, the diagnosis of PSP is made on the clinical evaluation by a neurologist or neuro-ophthalmologist. The diagnosis of PSP may be supported by findings on a MRI of the brain; however, the MRI changes are not specific to PSP. PSP is only confirmed on autopsy studies of the brain. The microscopic evaluation reveals abnormal tau protein in certain areas of the brain as well as characteristic changes to the neurons e.g. neurofibrillary tangles and tufted astrocytes.

Prognosis:

PSP is a progressive disease extending over years; many patients will eventually experience the typical symptoms within 3 to 4 years. Walking and balance difficulty prompt many PSP patients to utilize a wheelchair to come degree often within 5-6 years. Most patients will require some assistance with daily activities such as feeding, dressing, and bathing. The pace of progression and extent of symptoms is variable between patients. Most patients have difficulty with walking or falls early in the course, and many patients develop dysarthria, bradykinesia, cognitive and vision problems within the first 1-2 years. Patients may develop swallowing problems within a few years, which increases the risk of aspiration (food down the “wrong pipe”) pneumonia. Pneumonia is the leading cause of death in PSP patients. The average life expectancy of patients with PSP is approximately 7 years with a relatively wide range of variability.

Treatment:

While there is no cure for PSP, many of the symptoms can be helped with simple treatments. Approximately 30% of patients with PSP respond favorably to traditional Parkinson’s disease

medications, at least initially. Mild to moderate dry eyes can be treated with artificial tears and ophthalmic ointments. Severe dry eyes may benefit from moisture chamber goggles or other dry eye therapies. Light sensitivity may respond to tinted lenses such as the FL-41 hue. Physical therapy and assistive devices such as a cane or walker can decrease the risk of falls, helping prevent injuries and broken bones. Swallowing exercises can decrease the chances for choking and aspiration pneumonia. Researchers are actively trying to figure out why PSP occurs, who is at risk, and possible medications for diseases that are caused by the tau protein.

Follow up:

Patients diagnosed with PSP should see their neurologist or neuro-ophthalmologist on a regular basis to monitor for progression of new symptoms that could be treated; these clinic visits should include discussions of advanced directives concerning treatment options for possible future symptom difficulties as appropriate.

Frequently Asked Questions

Is PSP hereditary?

Most patients with PSP do not have a family history of this disease, and PSP has only rarely occurred as a strongly genetic disorder in families. A gene variant on chromosome 17 known as the H1 haplotype is more common in patients with PSP; however, this alone will not result in the clinical disease PSP.

How did I get PSP?

Researchers do not currently understand the primary triggers for PSP. There are no definite known risk factors, behaviors, or exposures that cause PSP. Some studies suggest that rural living may be associated with a higher PSP risk for unknown reasons. Specific toxins found in certain fruits native to the island of Guadeloupe are reported to cause symptoms similar to PSP.

Why did it take so long to diagnosis my PSP?

Many patients with PSP are diagnosed with Parkinson's disease (PD) for several years before the presence of PSP is suspected. Both PSP and PD share early symptoms and examination findings may only reveal very subtle or no differences initially. Whereas Parkinson's disease is common, PSP is very rare, and most primary care doctors have never seen a case. It is estimated that only 5 to 6 people out of 100,000 have PSP. It is often not until patients are evaluated by a neurologist or neuro-ophthalmologist that the diagnosis of PSP is considered; progression of symptoms over time is an important feature to help confirm the clinical diagnosis of PSP. Eye doctors may not consider this diagnosis when patients report that difficulty reading despite normal "20/20" visual acuity.

Will I develop Alzheimer's disease?

PSP patients do not develop Alzheimer's disease, however, they can experience a different forms of cognition difficulty. PSP patients have slowed thinking without the dramatic loss of memory typical of Alzheimer disease. PSP patients can lose interest in activities they once found pleasurable. They may become more emotional either laughing or crying more easily. They can also become more irritable.

Support groups

Cure PSP: http://www.psp.org/

PSP Association: http://www.pspassociation.org.uk/

NINDS: http://www.ninds.nih.gov/disorders/psp/detail_psp.htm