Progressive Supranuclear Palsy: Disease Profile and Rehabilitation Strategies

Progressive supranuclear palsy (PSP) is the most common parkinsonian disorder after Parkinson disease (PD).\textsuperscript{1–3} Postural instability with frequent falls and difficulty moving the eyes in the vertical direction are the main symptoms of PSP.\textsuperscript{4,5} However, because the initial clinical features often resemble PD,\textsuperscript{6–8} many patients are referred for rehabilitation services with the wrong diagnosis. There is no cure or effective medication to manage PSP, and the progression of the symptoms is much faster than in PD.\textsuperscript{1,2} It is important that physical therapists be aware of the particularities of this disease to ensure that patients are referred to movement disorder specialists for the correct diagnosis. Despite the demand for rehabilitation in this population, there is no evidence in the literature to support its effectiveness. Studies with controlled methods are necessary to provide guidance to physical therapists in the management of this disease. The purposes of this update are to highlight the characteristics of this disease and to evaluate rehabilitation strategies.

**History and Nomenclature of PSP**

Progressive supranuclear palsy was first described as a distinct clinical entity in 1964.\textsuperscript{9} The syndrome was identified by Steele et al at a meeting of the American Neurological Association where they reported that a group of 9 patients with a progressive brain disorder did not conform to any classifications of diseases already known. Common symptoms included ophthalmoplegia, pseudobulbar palsy, dysarthria, dystonic rigidity of the neck and upper trunk, and dementia. Neuropathological alterations involved neuronal loss and neurofibrillary tangles in the basal ganglia, brain stem, and cerebellum. The disease was named \textit{progressive supranuclear palsy}, referring to the progressive degeneration of the brain structures localized superior to the oculomotor nuclei, causing palsy and eventual paralysis of ocular movements. Another term also used in the literature, but not as often, is \textit{Steele-Richardson-Olszewski syndrome} (SROS).\textsuperscript{10}


**Key Words:** Fall prevention, Gaze control, Progressive supranuclear palsy, Rehabilitation, Visual attention.

\textit{Cris Zampieri, Richard P Di Fabio}
Prevalence and Incidence
The prevalence of PSP has been reported in a number of studies along with PD or other parkinsonian syndromes. However, only a few epidemiological studies have specifically addressed the prevalence of PSP alone or its incidence. In 1988, Golbe et al assessed the crude prevalence of PSP in the general population in New Jersey and reported a rare occurrence of 1.39 cases per 100,000 people. A study conducted in Olmsted County, Minn, between the years of 1976 and 1990 showed an average annual incidence rate (new cases per 100,000 person-years) of 5.3. All cases were reported between the ages of 50 to 99 years; there were no cases before 50 years of age. More recent estimates from the United Kingdom have revealed that the disease is more common than previously considered, with a crude prevalence of 6.5 cases per 100,000 people. The studies used detailed methods for case identification to ensure a reliable prevalence estimate and showed that the true incidence of PSP may be masked by misdiagnosed cases. Moreover, Nath et al found that the majority of the patients are initially referred to non-neurologists who are not familiar with the disease, which makes an accurate diagnosis less probable.

The mean age of onset of the disease is between 60 and 65 years. Although there have been reports of a predominance of men with the disease, recent publications state that both sexes are equally affected. The average survival time is 7 years; however, there have been reports of neuropathologically confirmed cases of individuals with the disease who survived up to 11 years or 16 years. Patients usually die from complications of the disease, and the most common cause of death is pneumonia.

Etiology
The etiology of PSP is unknown. Pathologically, the disease is characterized by neurodegeneration, gliosis, and abnormal accumulation of tau protein in the basal ganglia, brain stem, prefrontal cortex, and cerebellum. In people who are healthy, the tau protein occurs normally and its function is to stabilize the cytoskeleton of neurons. In PSP, this protein becomes resistant to proteolysis and is partially crystallized, forming abnormal deposits of tangled fibers, which are called neurofibrillary tangles. Other diseases also present aggregates of tau protein and are called tauopathies. These diseases include corticobasal degeneration, Pick disease, frontotemporal dementia with parkinsonism associated with chromosome 17 abnormalities (FTDP–17), and Alzheimer disease. The degree to which these pathologies share the same pathophysiological mechanisms with PSP is not known. To date, researchers have found that tau filaments differ among these pathologies in terms of morphology and tau isoform content.

Regardless of the primary cause of the disease, research has shown that the occurrence of neurofibrillary tangles is related to 2 cellular events: mitochondrial dysfunction and oxidative stress. Albers and Augood postulated on how these 2 events contribute to generate a cycle of destruction in neurons. The mitochondria are the key intracellular structures controlling the production of free radicals; therefore, when their function is impaired, the levels of intracellular free radicals increase, causing additional damage to mitochondrial proteins, lipids, and DNA, which leads to further mitochondrial dysfunction. The interaction between the oxidative damage and the energy depletion inside the neuronal cell leads to a depolymerization of microtubules and hyperphosphorylation of the tau protein, which gives origin to the neurofibrillary tangles, resulting in cellular death. Although the specific cause of mitochondrial dysfunction and oxidative stress is not clear, there is evidence of a contribution of both environmental and genetic factors.

This update highlights the characteristics of progressive supranuclear palsy and evaluates rehabilitation strategies for the disease.

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The neurodegenerative effects of these exotic plants has been shown by experimental studies and animal models. The plants are of the Annonaceae family, in particular *Annona muricata*, and contain substances (ie, quinolines, acetogenins, and rotenoids) that have been found to be neurotoxic. When cultures of mesencephalic dopaminergic neurons prepared from the midbrain of rat embryos were exposed to quinolines contained in the root of *A. muricata*, a degeneration of 50% of the neurons was observed after a period of 24 hours. According to the authors, the neurotoxicity of quinolines comes from an inhibition of the mitochondrial function, leading to neuronal death by adenosine triphosphate depletion. Acetogenins also have been shown to cause neuronal degeneration in cultures of dopaminergic cells by the same mitochondrial inhibitory process. Chronic administration of quinolines to squirrel monkeys for up to 104 days produced motor symptoms similar to parkinsonism.

In humans, the particular devastating effects of one of the substances, the rotenoids, has been described in a group of 3 young drug addicts who self-administered this substance under the impression it was heroin and developed parkinsonian-like symptoms. A postmortem examination of the cases showed depletion of dopaminergic neurons and extensive gliosis in the substantia nigra.

Evidence supporting a genetic cause of PSP also has been found. Several authors have described the occurrence of familial postmortem confirmed cases. Alterations of the tau gene associated with PSP have been described in a number of publications. More recently, investigators have found that mitochondrial genetic alterations also may play a role in the pathogenesis of PSP.

**Clinical Features and Diagnosis**

Typically, the clinical picture of PSP is characterized by early postural instability with recurrent falls, vertical gaze palsy, pseudobulbar palsy with speech and swallowing problems, bradykinesia, axial rigidity, and subcortical dementia. The gait is clumsy, slow, and unsteady, resembling a “drunken sailor.” With the progression of the disease, walking is no longer independent, and after 5 years, on average, patients are unable to stand unassisted, requiring the use of a wheelchair.

Slowness of vertical saccades is one of main diagnostic criteria for PSP. However, it typically develops 3 years after onset of other supporting symptoms, which makes the diagnosis uncertain in early stages of the disease. The first manifestation of slowing of vertical saccades may be difficulty reading or seeing food on a plate. Other eye-movement impairments commonly observed are apraxia of lid opening or closing (difficulty or slowness with voluntarily opening or closing the eyes), blepharospasm (involuntary closure of the eye caused by spasms of the orbicularis oculi), and decreased blinking frequency. The combination of oculomotor abnormalities and facial dystonia, with overactivity of the frontalis, gives the patient a characteristic “staring face.”

The diagnosis of PSP is exclusively clinical; laboratory tests and imaging exams cannot detect the disease, but help rule out other pathologies. In 1995, the National Institute of Neurological Disorders and Stroke (NINDS) and the Society for Progressive Supranuclear Palsy (SPSP) established the most currently accepted criteria for the diagnosis of PSP. In 2003, these criteria were redefined, and patients can be classified as having possible, probable, or definite PSP. Vertical gaze palsy and postural instability with falls are key symptoms in this classification. The Table shows the clinical inclusion and exclusion diagnostic criteria by the modified NINDS-SPSP consensus. As shown in the Table, a definite diagnosis can only be confirmed with postmortem examination, where the clinical presentation has to match specific neuropathologic findings.

Despite the existence of diagnostic criteria, the diagnosis of PSP remains challenging, especially in the early stages of the disease. Misdiagnosis of PSP as PD is common. On a survey done with 437 patients recruited through the SPSP, Santacruz et al found that one third of the cases had been previously diagnosed as PD. Recent epidemiological studies also have revealed a number of mis-
diagnosed cases and suggest that not only the similarities of the symptoms to PD may be confounding, but also some clinicians may be inexperienced or may not be aware of this disease to provide an accurate diagnosis. In the United States, PSP is correctly diagnosed only 75% of the time, as shown by a retrospective analysis of 180 neuropathologically confirmed cases from the SPSP brain bank.19

For the practicing clinician, several clinical features should raise the suspicion of PSP and help differentiate PSP from PD:

- Failure to respond to antiparkinsonian medications such as levodopa
- Vertical gaze palsy
- Recurrent falls in a backward direction
- Bulbar signs (difficulty with speech, swallowing)

However, physical therapists should be aware that there are heterogeneous manifestations of symptoms in people with PSP. Although it is not common, there have been reports of atypical cases with postmortem confirmation as definite PSP. The atypical features include tremor,46,47 absence of eye gaze palsy,48,49 pure akinesia without rigidity,50 and asymmetric features.49

The detection of PSP with laboratory or imaging exams is not a reality in clinical practice; however, some progress has been made in this area in recent years. Sagittal magnetic resonance imaging (MRI) comparisons among PSP, PD, and other parkinsonian syndromes have revealed some particular alterations that may help differentiate among these diseases. The most common patterns of abnormalities are found to be atrophy of the midbrain area, also described as the “hummingbird” sign,51 increase of the third ventricle area,52,53 and atrophy of the superior cerebellar peduncle.54,55 However, a general limitation of MRI exams is that such alterations are detectable only in advanced stages of the disease, and they do not address the problem of early misdiagnosis.

Physical therapists play an important role in helping patients receive the correct diagnosis for the disease. It is important that PSP be diagnosed correctly as early as possible to allow patients and family members to prepare accordingly for the quickly progressive course of the symptoms. Considering that many patients referred for rehabilitation services may be potentially misdiagnosed, it is important that “suspicious cases” be referred back to a movement disorder specialist for a second opinion. The dominant clinical problems of a suspicious case are severe postural instability with frequent falls and vertical

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### Table.
National Institute of Neurological Disorders and Stroke–Society for Progressive Supranuclear Palsy Diagnostic Criteria for Progressive Supranuclear Palsy

<table>
<thead>
<tr>
<th>Inclusion Criteria</th>
<th>Possible</th>
<th>Probable</th>
<th>Definite</th>
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<tr>
<td>Gradually progressive disorder</td>
<td>Gradually progressive disorder</td>
<td>Gradually progressive disorder</td>
<td>Neuropathologic findings at autopsy confirming possible or probable diagnosis</td>
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<tr>
<td>Age of onset 40 y or later</td>
<td>Age of onset 40 y or later</td>
<td>Age of onset 40 y or later</td>
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<tr>
<td>Vertical supranuclear palsy</td>
<td>Vertical supranuclear palsy</td>
<td>Vertical supranuclear palsy</td>
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<tr>
<td>or Slowing of vertical saccades with postural instability and falls in the first year of the disease</td>
<td>Postural instability and falls in the first year of the disease</td>
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<table>
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<th>Exclusion Criteria</th>
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<tr>
<td>Recent history of encephalitis</td>
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<td>Alien limb syndrome</td>
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<tr>
<td>Cortical sensory deficits</td>
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<tr>
<td>Focal frontal or temporoparietal atrophy</td>
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<tr>
<td>Hallucinations or delusions unrelated to dopaminergic therapy</td>
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<tr>
<td>Cortical dementia of Alzheimer type</td>
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<tr>
<td>Prominent early cerebellar symptoms or unexpected dysautonomia</td>
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<tr>
<td>Evidence of other diseases that could explain the clinical features</td>
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<table>
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<tr>
<th>Neuropathologic Criteria</th>
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<tr>
<td>High density of neurofibrillary tangles and neuropil threads in at least 3 of the following areas: pallidum, subthalamic nucleus, substantia nigra, or pons</td>
</tr>
<tr>
<td>Low to high density of neurofibrillary tangles and neuropil threads in at least 3 of the following areas: striatum, oculomotor complex, medulla, or dentate nucleus</td>
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</table>

gaze palsy. The falls usually happen unexpectedly and very often are in a backward direction.6,7 Vertical gaze palsy may not be observable by the physical therapist until it is well developed, in which case the patient would simply not be able to look down. However, there are some signs that may indicate the development of a possible eye-movement palsy: (1) changes in the ability to see well, commonly presented as double vision, (2) difficulty reading (the patient would notice that the eyes cannot move down to the next line and the same line is read over and over again), and (3) difficulty guiding utensils to the mouth while eating.56

In addition to the NINDS-SPSP consensual criteria, physical therapists should be aware of “red flags” that suggest a diagnosis other than PSP1 such as: onset of symptoms earlier than age 40 years, aphasia, duration of more than 20 years, cortical dementia, cortical sensory or visual deficits, hallucinations or delusions not due to medications, fluctuating state of cognition and arousal, severe orthostatic hypotension, unilateral contractures, a response to levodopa, and levodopa-induced dyskinesias.

Rating Scales and Prognosis

Two rating scales have been proposed to assess the level of impairment of patients: the Unified Parkinson’s Disease Rating Scale (UPDRS) and the Progressive Supranuclear Palsy Rating Scale (PSPRS). The UPDRS is the most commonly used rating scale, and although it was designed for patients with PD, the motor section of the scale has been shown to yield valid and reliable data for patients with PSP as well.57 The rating scale for each item varies between 0 (no disability) and 4 (high level of disability), summing up to a maximum score of 56 points.

The PSPRS is a newer scale, designed by Lawrence I Golbe, MD, specifically to assess level of disability in people with PSP.58 It evaluates aspects of the disease in the domains of health history, mentation, bulbar function, eye and lid movement, limb movement, and trunk movement. The total maximum score is 100, reflecting the highest level of impairment (Appendix).

The assessment of functional levels and staging of the symptoms in PSP may be particularly helpful to predict the prognosis of the disease. Santacruz et al45 found that the early presence of falls, bradykinesia, and inability to move the eyes downward are negative factors in the survival time of patients. Nath et al10 confirmed the relationship between survival time and the onset of early falls, and they also reported bulbar problems and diplopia as negative predictors. Another recent study13 has shown that the impairment of gait is a key factor in the prognosis of PSP, as compared with other motor impairments such as speech difficulty and swallowing problems. This study involved a longitudinal assessment of clinical and videotape databases of 50 cases with probable diagnosis. The authors classified gait impairment at 3 different levels: loss of independent walking, inability to stand unassisted, or requiring a wheelchair. Their main finding was that 48% of the patients reached 1 of the 3 levels of impairment within 4 years of onset of the disease. Based on these results, they suggested that gait impairment be assessed as a means of verifying the effectiveness of new interventions.

Surgical Treatment and Medications

To date, there is no effective medication or surgical treatment to cure or delay the progression of the symptoms in PSP. Although palliative interventions may be used to alleviate major symptoms, no drug has been found to efficiently treat the origin of the problem. Palliative interventions include botulinum injections for blepharospasm or neck rigidity, glasses with prisms for visual disturbances, antidepressants and support therapy for depression, percutaneous endoscopic gastrostomy for swallowing problems, and speech therapy for dysarthria.1,5

Although some authors59 have reported benefits of dopaminergic drugs in some cases, the great majority of researchers argue that, on a larger scale, patients are unresponsive to levodopa or any other neurotransmitter-specific therapies.60–62 As stated earlier in this update, responsiveness to parkinsonian medication has been used as a “red flag” against the diagnosis of PSP.1 The reason behind the ineffectiveness of parkinsonian drugs for people with PSP is probably related to the widespread degenerative nature of the disease.4,5 While PD affects the substantia nigra primarily, PSP affects many other nuclei.1,9,19

Rehabilitation

Patients with PSP usually seek or are referred for rehabilitation for balance and gait problems with frequent falls.63 There are no reports in the literature of how often people with PSP are referred for rehabilitation. Nevertheless, the referrals and the demand for physical therapy certainly tend to increase with an increase in the awareness of the disease. Unfortunately, evidence-based approaches to rehabilitation in PSP are lacking, and the only research available consists of case reports involving 1 or 2 patients.63–65 Below is a summary of these studies and a discussion on the research that still needs to be conducted in this field.

Izzo et al65 were the first authors to address the rehabilitation of a patient with a neurological presentation indicative of PSP. Functionally, the patient was described as having moderate involvement in motor function. Cognitively, the authors reported mild dementia, slow processing of thought, memory function below normal,
and mild impairment of judgment skills. The rehabilitation program included limb-coordination activities, tilt-board balancing, ambulation activities incorporating trunk flexion and rotation, and strategies to compensate for impaired visual scanning. No further information was provided regarding the exercises or the frequency and duration of the treatment sessions. At the end of the exercise program, improvements were observed in the patient’s standing balance and ability to scan the environment. Fine coordination remained the same, and gait characteristics showed little improvement, although the patient reported feeling safer during ambulation.

A similar report by Sosner et al.64 described the rehabilitation of 2 patients. Like the previous case study, the diagnosis was based on clinical findings, and exams excluded other pathologies. Both patients showed moderate involvement in motor function. Cognitively, the first patient had mild memory impairment and slowness of thought. The second patient had no impairment in memory and thought processing, but impaired abstract thinking. Each patient followed an individualized rehabilitation program that involved strength training with progressive resistive exercises and isokinetic exercises, coordination exercises, gait training, transfer training to and from a bed and chair, and stretching of the neck muscles. In addition, the second patient was taught to compensate for downward gaze impairments by using head movements. As in the previous case study,65 no exercise description or information on the frequency and duration of treatment sessions was provided. The only information provided regarding the outcomes of the study was that patients were able to achieve safe ambulation. No observations were made related to changes in balance, coordination, strength, or transferring abilities after the exercise program.

The case studies described above are important because they represent the experiences of clinical practitioners in the management of PSP. Although the results cannot be generalized across all patients with PSP, because these are case reports, the authors’ observations raise questions that can guide future research concerning the effectiveness of balance and gait training programs to manage PSP. Still, many limitations can be found in these reports: (1) no rating scales were used to initially classify the patients, which makes it difficult to compare them with other patients in the clinical setting or in other studies, (2) the exercise program was not thoroughly described, which limits the replication of their approach, and (3) the assessment of many outcomes was not quantified, which can be biased by the observer, and may not allow small changes to be identified.

In 2002, Suteerawattananon et al.63 reported a case report using a body-weight–support training program for a patient with PSP. This case report was conducted under more controlled conditions compared with previous reports of Sosner et al.64 and Izzo et al.65 The patient had mild to moderate motor involvement, with a score of 24 out of 52 on the motor section of the UPDRS. Cognitive function was mildly affected, with a score of 27 on the Mini-Mental State Exam.66 Several measurements were obtained before and after rehabilitation. Mobility was assessed with the Timed “Up & Go” Test,35 a timed 360-degree test,63 and a timed 5-step test.63 Balance measures included the Functional Reach Test,67 a test of balance on a foam pad,68 the Berg Balance Scale,69 and a postural stability test done on a force plate.63 Temporal and spatial characteristics of the patient’s gait were assessed while he walked on a 3-m instrumented walkway.63 In addition, fall incidence before, during, and after treatment was monitored through a questionnaire answered by the caregiver. The treatment program consisted of body-weight–support treadmill training for 1½ hours, 3 days a week, for 8 weeks. Different directions of walking were practiced (forward, backward, and sideways [both left and right]) with 15% body weight support. In addition, postural reaction to perturbation was practiced in the same 4 directions, with the harness system for safety and with 0% body weight support. The results of the case study showed improvement on all measures except the Timed “Up & Go” Test. According to the authors, the lack of improvement on the Timed “Up & Go” Test can be justified by the fact that the patient was not trained in sequencing of motor tasks or sit-to-stand activities.63

Based on this case study, body-weight–support treadmill training was beneficial for 1 patient with PSP who had mild to moderate functional impairment. However, it remains unknown whether this approach would be effective when tested on a larger scale. Clinical trials with a larger number of subjects are necessary to confirm efficacy. As can be seen, the literature is very limited in the field of rehabilitation for PSP. Despite case reports showing benefits following rehabilitation, basic questions remain unanswered:

1. What is the most effective and comprehensive rehabilitation approach to treat patients with PSP, given that there are many approaches to the problem of impaired balance and gait?

2. Because patients may seek treatment at different stages of the disease, what would be realistic goals for patients with mild, moderate, and severe impairments?

3. Because the disease progresses so quickly, is it realistic to hope for an improvement in function or would it be more realistic to expect that patients maintain their function?
4. Would intervention for balance and mobility be the same for such problems in someone with PD?

Rehabilitation strategies for PSP would most likely differ from those for PD because vertical gaze palsy is unique to PSP and has the potential to create additional balance and mobility problems. A recent study by Ondo et al. showed that people with PSP had markedly worse postural control compared with people with PD matched for age and disease duration. A patient diagnosed with PSP will likely have some level of vertical oculomotor palsy, which contributes to poor postural control. Common complaints related to gaze palsy are difficulty scanning the environment, seeing curbs and obstacles during locomotion, judging distances, and going up or down the stairs.

Vision plays a critical role in the control of locomotion because it provides input for anticipatory reactions of the body in response to variations in and constraints of the environment. Studies of subjects who were healthy have shown that anticipatory saccades occur normally in situations that involve changing direction of walking or prior to obstacle avoidance. Di Fabio et al. have shown that down saccades are not generated as often during obstacle stepover in community-dwelling elderly people at a high risk for falls and fall-related injuries compared with elderly people at low risk for falls and fall-related injuries.

In the case of PSP, the input provided by vision is limited because patients have difficulty executing down saccades. As a consequence, they may lack appropriate anticipatory reactions to the environment, and they may be more susceptible to falls and accidents while walking. Although clinical practice shows that gaze limitations play a role in the balance and gait deficits in PSP, no studies have been conducted to investigate this relationship. Thus, it would be appropriate to ask whether rehabilitation for PSP should involve “eye-movement training” in addition to balance and gait training.

As described earlier in this update, Izzo et al. and Sosner et al. have incorporated strategies in their rehabilitation program to overcome gaze limitation problems by teaching patients to scan the environment where they walk or to move the head while maintaining the eyes fixated on the floor. Unfortunately, the success of such strategies and the extent to which they contributed to the improvement in the patients’ balance and gait were not discussed by the authors.

Recent work suggests that people with PSP have difficulty inhibiting visual reflexes that may interfere with gaze control or compound vertical gaze palsy. It is theoretically plausible that eye-movement exercise may improve the ability to suppress fixation and allow some degree of gaze shift to occur. Preliminary evidence indicates that a rehabilitation program emphasizing eye-movement exercise might increase eye range of motion and improve visual attention in some people with PSP. More research, however, is necessary to investigate the benefits of gaze-oriented interventions for PSP.

Summary

Progressive supranuclear palsy is a parkinsonian syndrome commonly misdiagnosed as PD. The progression of the disease is much faster, and the impairment of gait and balance is more dramatic, than in PD. We believe that the demand for rehabilitation in this population will increase; however, there is no evidence in the literature to guide clinical practice. More research is necessary to answer basic questions regarding the effectiveness of rehabilitation for patients with PSP.

References


## Appendix.
### Progressive Supranuclear Palsy (PSP) Rating Scale and Staging System

Medical Advisory Board of the Society for Progressive Supranuclear Palsy (SPSP)—Lawrence D Golbe, MD, Chair

For more copies or for information on PSP, contact the SPSP at 1-800-457-4777/www.psp.org

### History (from patient or other informant)

<table>
<thead>
<tr>
<th>Item</th>
<th>Description</th>
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<tbody>
<tr>
<td>1. Withdrawal (relative to baseline personality)</td>
<td>0 = None. 1 = Follows conversation in a group, may respond spontaneously but rarely if ever initiates exchanges. 2 = Rarely or never follows conversation in a group.</td>
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<td>2. Aggressiveness</td>
<td>0 = No increase in aggressiveness. 1 = Increased, but not interfering with family interactions. 2 = Interfering with family interactions.</td>
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<td>3. Dysphagia for solids</td>
<td>0 = Normal; no difficulty with full range of food textures. 1 = Tough foods must be cut up into small pieces. 2 = Requires soft solid diet. 3 = Requires pureed or liquid diet. 4 = Tube feeding required for some or all feeding.</td>
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<tr>
<td>4. Using knife and fork, buttoning clothes, washing hands and face (rate worst)</td>
<td>0 = Normal. 1 = Somewhat slow but no help required. 2 = Extremely slow; or occasional help needed. 3 = Considerable help needed but can do some things alone. 4 = Requires total assistance.</td>
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<tr>
<td>5. Falls (average frequency if patient attempted to walk unaided)</td>
<td>0 = None in the past year. 1 = &lt;1 per month; gait may otherwise be normal. 2 = 1–4 per month. 3 = 5–30 per month. 4 = &gt;30 per month.</td>
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<td>6. Urinary incontinence</td>
<td>0 = None of a few drops less than daily. 1 = A few drops staining clothes daily. 2 = Large amounts, but only when asleep; no pad required during day. 3 = Occasional large amounts in daytime; pad required. 4 = Consistent requiring diaper or catheter awake and asleep.</td>
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<tr>
<td>7. Sleep difficulty</td>
<td>0 = Neither 1° nor 2° insomnia (ie, falls asleep easily and stays asleep). 1 = Either 1° or 2° insomnia; averages ≥5 hours sleep nightly. 2 = Both 1° and 2° insomnia; averages ≥5 hours sleep nightly. 3 = Either 1° or 2° insomnia; averages &lt;5 hours sleep nightly. 4 = Both 1° and 2° insomnia; averages ≤5 hours sleep nightly.</td>
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### Mental Exam

Items 8–11, use this scale:

<table>
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<th>Item</th>
<th>Description</th>
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<tbody>
<tr>
<td>8. Disorientation</td>
<td>0 = Clearly absent 1 = Equivocal or minimal 2 = Clearly present, but not interfering with activities of daily living (ADL) 3 = Interfering mildly with ADL 4 = Interfering markedly with ADL</td>
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<tr>
<td>9. Bradyphrenia</td>
<td>0 = None 1 = 16%–50% of normal amplitude 2 = Slow or hypometric; 86%–100% of normal amplitude 3 = 51%–85% of normal amplitude 4 = Moderate lid-opening inhibition or blepharospasm causing partial visual disability.</td>
</tr>
<tr>
<td>10. Emotional incontinence</td>
<td>0 = None 1 = Absent. 2 = Slight or detectable only on activation. 3 = Only partial range of motion possible. 4 = Little or no passive motion possible.</td>
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<td>11. Grasping/imitative/utilizing behavior</td>
<td>0 = None. 1 = Minimal; all or nearly all words easily comprehensible (to examiner, not family). 2 = Definite, moderate; most words comprehensible. 3 = Severe; may be fluent, but most words incomprehensible. 4 = Mute; or a few poorly comprehensible words.</td>
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### Bulbar Exam

Items 12–15, use this scale. Rate by inspection of saccades on command from the primary position of gaze to a stationary target.

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<th>Item</th>
<th>Description</th>
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<tr>
<td>12. Dysarthria (ignoring palilalia)</td>
<td>0 = None. 1 = Minimal; all or nearly all words easily comprehensible (to examiner, not family). 2 = Definite, moderate; most words comprehensible. 3 = Severe; may be fluent, but most words incomprehensible. 4 = Mute; or a few poorly comprehensible words.</td>
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<tr>
<td>13. Dysphagia (for 30–50 cc of water from a cup, if safe)</td>
<td>0 = None. 1 = Fluid pools in mouth or pharynx. Or swallows slowly, but no choking/coughing. 2 = Occasionally coughs to clear fluid; no frank aspiration. 3 = Frequently coughs to clear fluid; may aspirate slightly; may expectorate frequently rather than swallow secretions. 4 = Requires artificial measures (oral suctioning, tracheostomy, or feeding gastrostomy) to avoid aspiration.</td>
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### Supranuclear Ocular Motor Exam

Items 14–16, use this scale. Rate by inspection of saccades on command from the primary position of gaze to a stationary target.

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<th>Description</th>
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<tr>
<td>14. Voluntary upward saccades</td>
<td>0 = None. 1 = Absent. 2 = Slight or detectable only on activation. 3 = Only partial range of motion possible. 4 = Little or no passive motion possible.</td>
</tr>
<tr>
<td>15. Voluntary downward saccades</td>
<td>0 = None. 1 = Absent. 2 = Slight or detectable only on activation. 3 = Only partial range of motion possible. 4 = Little or no passive motion possible.</td>
</tr>
<tr>
<td>16. Voluntary felt and right saccades</td>
<td>0 = None. 1 = Absent. 2 = Slight or detectable only on activation. 3 = Only partial range of motion possible. 4 = Little or no passive motion possible.</td>
</tr>
<tr>
<td>17. Eyelid dysfunction</td>
<td>0 = None. 1 = Blink rate decreased (&lt;15/min) but no other abnormalities. 2 = Mild inhibition of opening or closing or mild blepharospasm. 3 = Moderate lid-opening inhibition or blepharospasm causing partial visual disability. 4 = Functional blindness or near-blindness because of involuntary eyelid closure.</td>
</tr>
</tbody>
</table>

### Limb Exam

Items 18–20, use this scale. Rate by inspection of saccades on command from the primary position of gaze to a stationary target.

<table>
<thead>
<tr>
<th>Item</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>18. Limb rigidity (rate the worst of the four)</td>
<td>0 = Absent. 1 = Slight or detectable only on activation. 2 = Definitively abnormal. But full range of motion possible. 3 = Only partial range of motion possible. 4 = Little or no passive motion possible.</td>
</tr>
<tr>
<td>19. Limb dystonia (rate the worst of the four; ignore neck and face)</td>
<td>0 = Absent. 1 = Subtle or present only when activated by other movement. 2 = Obvious but not continuous. 3 = Continuous but not disabling. 4 = Continuous and disabling.</td>
</tr>
<tr>
<td>20. Finger tapping (if asymmetric, rate worst side)</td>
<td>0 = Normal (&gt;14 taps/5 s with maximal amplitude). 1 = Impaired (6–14 taps/5 s with moderate loss of amplitude). 2 = Barely able to perform (0–5 taps/5 s or severe loss of amplitude).</td>
</tr>
</tbody>
</table>
Appendix. Continued.

Progressive Supranuclear Palsy (PSP) Rating Scale and Staging System

21. Toe tapping (if asymmetric, rate worst side)
   0 = Normal (>1.4 taps/5 s with maximal amplitude).
   1 = Impaired (6–1.4 taps/5 s with moderate loss of amplitude).
   2 = Barely able to perform (0–5 taps/5 s or severe loss of amplitude).

22. Apraxia of hand movement
   0 = Absent.
   1 = Present, not impairing most functions.
   2 = Impairing most functions.

23. Tremor in any part
   0 = Absent.
   1 = Present, not impairing most functions.
   2 = Impairing most functions.

Gait/Midline Exam

24. Neck rigidity or dystonia
   0 = Absent.
   1 = Slight or detectable only on activation.
   2 = Definitely abnormal. But full range of motion possible.
   3 = Only partial range of motion possible.
   4 = Little or no passive motion possible.

25. Arising from chair
   0 = Normal
   1 = Slow but arises on first attempt.
   2 = Requires more than one attempt, but arises without using hands.
   3 = Requires use of both hands.
   4 = Unable to arise without assistance.

26. Gait
   0 = Normal.
   1 = Slightly wide-based or irregular or slight pulsion on turns.
   2 = Must walk slowly or occasionally use walls or helper to avoid falling, especially on turns.
   3 = Must use assistance all or almost all the time.
   4 = Unable to walk, even with walker; may be able to transfer.

27. Postural stability (on backward pull)
   0 = Normal (shifts neither foot or no foot).
   1 = Must shift each foot at least once but recovers unaided.
   2 = Shifts feet and must be caught by examiner.
   3 = Unable to shift feet; must be caught, but does not require assistance to stand still.
   4 = Tends to fall without a pull; requires assistance to stand still.

28. Sitting down (may touch seat or back but not arms of chair)
   0 = Normal.
   1 = Slightly stiff or awkward.
   2 = Easily positions self in chair, but descent into chair is uncontrolled.
   3 = Has difficulty finding chair behind him/her and descent is uncontrolled.
   4 = Unable to test because of severe postural instability.

Section Totals

History 0 –24
Mentation 0 –16
Bulbar 0 – 8
Ocular 0 –16
Limb 0 –16
Gait 0 –20
TOTAL 0–100

*PSP Staging System  1 2 3 4 5
(ignore any inability to stand up from the seated position)
1 = Gait and stability are normal or equivocal.
2 = Gait is abnormal but stable, requiring only 1–2 steps back on pull test.
3 = Would fall or retropulse on pull test; may require cane or intermittent assistance
4 = Can walk only with walker or continuous assistance
5 = No useful gait, but may be able to remain standing unassisted or transfer between chair and bed

*The PSP Staging System component of this assessment tool is currently under modification.