PROGRESSIVE SUPRANUCLEAR PALSY:

AN OVERVIEW & IMPORTANCE OF REHABILITATION

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Abstract

Much attention has been devoted to neuro-degenerative disorders of late, particularly Parkinson's disease and progressive supranuclear palsy (PSP). This article reviews the symptomatology and neurology of PSP, which is also known as Dudley Moore's disease. The utility of optical and non-optical intervention, for purposes of visual rehabilitation, is explored.

The major visual sequelae of progressive supranuclear palsy are as follows: restriction of vertical gaze (both saccades and pursuits), saccadic intrusions in fixation, decreased blink rate, and nonparalytic eyelid apraxia. The progressive nature of this degeneration eventually extinguishes all eye movements, yielding complete external ophthalmoplegia.

Key Words

supranuclear gaze palsy, vertical gaze restriction, convergence paralysis, saccadic intrusions, yoked prism, prismatic redirection

Fortunately, there are few progressive diseases suffered by humans which rob us of our self, but in the infrequent instance where one is diagnosed with such a condition—Alzheimer's disease, Parkinson's disease, Lou Gehrig's disease—the consequences can be devastating. Progressive supranuclear palsy (PSP) is a progressive, degenerative disease of the mesencephalon region of the brain,[1] that can leave the person as a masked shell of his former self. PSP presents a clinically similar picture to other neurological degenerative diseases, such as Parkinson's disease. PSP, however, is relatively rarer than its widely publicized counterpart; there is a prevalence of 500,000 Americans suffering from Parkinson's disease, while only 4% of this same number are affected by PSP.[2] The disease almost equally affects men and women, in their sixth or seventh decade.[3] As with Parkinson's disease, the etiology of this progressive degeneration is unknown; and diagnosis is confirmed only on autopsy.[4]

Due to the pattern of midbrain and extrapyramidal (non-motor tract) involvement, patients with PSP usually experience difficulties with balance in the early stages. While walking, the individual with PSP may have stiff gait and upright posture. Companions note personality changes; the individual may seem more irritable or more apathetic than normal. Emotional tone seems to increase, and patients respond with greater emotion than before. Early signs include vision changes such as blurry vision and eyelid abnormalities. The patient may blink less frequently, and may also experience blepharospasm. Companions comment that the patient seems to "stare through them," and has difficulty maintaining eye contact in conversation. Speech difficulties emerge, in which the patient may exhibit delays, and the voice may take on a hoarse or raspy quality.[2] The phenomenon of pseudobulbar dysarthria, where speech tends to rise in volume near the end of a spoken sentence, presents later in the course of the disease.[5] Loss of involuntary muscle control soon affects the swallowing mechanism, and death often occurs as a consequence of aspiration pneumonia.[6] Various degrees of dementia may emerge during the course of the disease.

REVIEW OF THE DISEASE

Symptoms

At the onset of PSP, patients experience difficulties with walking, balance, and report visual disturbances.[7] These early symptoms generalize to universal difficulties of movement and vision; cognitive changes and emotional disturbances do not affect all PSP patients.[8] The symptomatology of 50 patients diagnosed with PSP in New Jersey was delineated by the time course. Golbe et al.,[9] found that the earliest presenting symptom (after tentative diagnosis) was the necessity for gait assistance at a time course of 3.1 years post diagnosis; visual symptom presentation followed closely at 3.9 years. For perspective, death occurred in this sample at 9.7 years post diagnosis. A review of the most common symptoms experienced by PSP patients may be found in Table 1.

The premier visual symptom for these patients is blurred vision. Diplopia is rarely reported in the early stages, and when it is, it is likely caused by dry eye.[4] Patients frequently note ocular irritation and photophobia. For perspective, Friedman et al., found half of their PSP patient sample to have complaints of blur, whereas eye irritation and diplopia were less frequent complaints, showing nearly

Table 1.
Most frequently presenting symptoms, at onset of progressive supranuclear palsy.[7]

Body System	Effects
BALANCE	Fall backward
GAIT	Unsteady; Drift to one side
MENTAL CAPACITY	Forgetfulness
SPEECH	Slurred
VISION	Blur; Diplopia

equal prevalence in one-third of their sample.[4]

Ocular and Visual Signs

Visual system signs in PSP can be divided into three groups: I) Deficiencies of conjugate gaze and fixation; II) Deficiency in vergence; and III) Eyelid abnormalities. In general, difficulty with voluntary eye movements pervades this disease; involuntary eye movements (those which can be elicited by "doll's eyes" reflex) remain intact until the very late stages of the disease.[3,10]

Saccades are usually affected before pursuits.[3,11] Saccades primarily show slowing (hypometria) and restriction, as well as increased latency as the disease progresses.[12] Restriction tends to occur first in downgaze saccades, then in upgaze. Smooth pursuits are the next to be affected. They are often seen to break down into a series of small saccades. And unlike saccades, increased latency occurs initially. Thus, compared to normal elderly adults, patients with PSP show difficulty in pursuit *initiation*. Normal elderly subjects show slight increased latency of pursuit initiation compared to young adults, but it has not been found to be statistically significant.[13] Pursuit gain may be reduced with the ensuing brainstem and cerebellar involvement; when this occurs, eye-head tracking may become superior to the visual tracking ability alone.[14] Gradually, all voluntary saccade and pursuit eye movements are extinguished, yielding complete external ophthalmoplegia.[4] One case report noted an OKN response at the end stage of PSP, but it was coupled with a rotary head turn; Bisdorff et al., postulated this finding occurred due to a "disinhibited vestibulocollic reflex" in these patients. The patient had no voluntary eye movements, and only very slow, voluntary head movements.[15] This illustrates the pervading rigidity these patients experience late in the course of PSP. Fixational ability, or zero-velocity pursuit, is also affected in PSP; the fixation becomes unstable with multiple refixational eye movements, termed "saccadic intrusions" or "square wave jerks."[4]

After the patient encounters difficulties with vertical eye movements, horizontal eye movements become affected. Horizontal conjugate gaze becomes restricted, and then vergence ability is affected, yielding a convergence paralysis.[3] A bilateral internuclear ophthalmoplegia (INO), caused by degeneration of the Medial Longitudinal Fasciculus between cranial nerves III and VI, was noted in 25 to 30% of one PSP patient sample.[4] A bilateral INO (generated by an anterior midbrain or supranuclear infarct) would give a clinical finding of deficient vergence response.[12] Table 2 gives a summary of how the visual system is affected.

Eyelid involvement contributes significantly to a person's complaint of irritation or dry eye with this disease. The blink rate of PSP patients was found to be substantially decreased (three blinks per minute) compared to normals (nearly 16 blinks per minute).[4] The likelihood of resulting ocular surface disease is high. In addition to decreased blinking, blepharospasm and apraxia of eyelid opening or closing are involved.[1] The origin of the eyelid apraxia difficulty is in the supranuclear system. The patient shows transient inability to initiate lid opening (or closing).[4] Eyelid retraction is a fourth lid finding in this disease; caused by disruption in the levator—superior rectus synkinesis, the lids elevate disproportionately when the superior division of cranial nerve III is stimulated.[4]

Table 2.
Progression of supranuclear visual system involvement, PSP.

1. SACCADES
   Hypometria (slowing)
   Downgaze affected first
2. PURSUITS, Voluntary
   Restriction of gaze
   a). Downward
   b). Upward
   c). Horizontal
3. FIXATION STABILITY
   Square wave jerks
   (saccadic intrusions)
4. VERGENCE
   Convergence paralysis
5. COMPLETE EXTERNAL OPHTHALMOPLEGIA

CASE PRESENTATION

History

DC, a 69-year-old Caucasian male, presented to the Head Trauma Vision Rehabilitation Unit at the University Optometric Center, State University of New York, State College of Optometry clinic with complaints of vision that "blurs, then clears" as well as horizontal and vertical diplopia while reading and watching TV. He sustained a concussion two months prior to his visit, and reported frequent problems with loss of balance and vertigo. At this visit, he used a cane for support. During the history, the patient seemed to "stare through" the person to whom he was speaking; his speech was slow, forced, and raspy. Mr. C's previous ocular and visual history was notable for cataract surgery in both eyes, as well as a constant "sandy" feeling in the eyes. Family medical history revealed his father's diagnosis of Parkinson's disease. Synomed was the only medication reportedly used by the patient. He had received a diagnosis of PSP by his neurologist.

Findings

Visual acuity with his current plano progressive addition bifocal prescription was 20/50 OD, 20/25 OS. Best subjective visual acuity was improved, as follows:

OD: - 0.50 sph 20/30
OS: - 0.50 sph 20/25
Add: +2.50 sph OD, J2 at 15"; and
+2.50 sph OS, J2 at 15"

Distance cover tested elicited no horizontal or vertical deviation, but on near cover test, the patient manifested an intermittent 14 p.d. exotropia. Fixational testing showed more than 10 saccadic intrusions OD, OS, and OU in a 10-second time frame. Delayed latency and hypometria were noted on saccadic testing, and pursuit testing revealed marked superior and inferior gaze restriction, monocularly and binocularly. Phorometic testing yielded the following:

Distance Phoria: 3 Eso / 1 Right Hyper
Distance BI: X / 6 / 5
Distance BO: 10 / 14 / 0
Near Phoria: 19 Exo / No Hyper

Near BI: X / 24/ 16
Near BO: needed 8 p.d. BI to attain single vision
Fused Cross Cyl: +2.50

The ocular health examination was significant for 2+ conjunctival injection and moderate tear layer insufficiency. Much debris was noted on the anterior corneal surface. DC had to be reminded several times to blink during the evaluation. Ocular tensions, visual field testing, and posterior segment evaluation were unremarkable.

Assessment

Mr. C's complaint of blurred vision can be attributed to several of the examination findings, most notably the dry eye and significant number of saccadic intrusions during fixation. The diplopia complaint can be ascribed to the intermittent exotropia, and quite possibly the tear layer insufficiency. Mr. C had been diagnosed in 1998 with Parkinson's disease, yet that diagnosis was changed to progressive supranuclear palsy in 1999 as the picture of his symptomatology changed.

Plan

To address the intermittent exotropia, a trial of 10 p.d. base-in prism (with the appropriate add power) was conducted. Mr. C became emotional over the change, and he appreciated the prescription for comfort and clarity. Considering DC's overall inability with downgaze, the use of a bifocal was discouraged and single vision reading glasses with base-in prism were prescribed. Mr. C was reminded to "think blink," and was instructed on the use of artificial tears QID, with the addition of Celluvisc at night.

DISCUSSION

Etiology of PSP

Presently the cause of PSP is unknown. Suggested causative agents include free radical damage, viral etiology, random genetic mutation, or an unknown chemical. Progression of PSP in families may exist, in autosomal dominant form.[16] Brain degeneration in PSP occurs secondary to an abnormality in microtubule binding (protein tau).[1,10,17] The effects of such abnormal binding are neurofibrillary tangles (also common to Alzheimer's disease), gliosis, and atrophy.[17]

In the early stages, the PSP patient may not show gaze palsy. The dentate nucleus of the cerebellum (involved in coordination of movement and balance) is one of the first structures to be affected, whereas the superior colliculus, which is invoved with voluntary, conjugate gaze is initially spared.[18] The main structures eventually affected are the dentate nucleus, subthalamic nuclei, substantia nigra (yielding difficulty with initiation of movement), and the superior colliculus.[1,3,10,18] The frontal lobe may also become involved, as atrophy of the corpus callosum has been evidenced in PSP patients on CT or MRI.[1]

Table 3. Control of voluntary eye movements, via cortex.[2, 20, 21]

AREA	FUNCTION	LOCATION
Frontal Eye Field (FEF)	Trigger voluntary saccade Trigger voluntary pursuit Disengages fixation	Precentral Gyrus
Supplemental Eye Field (SEF)	Control timing of a saccade sequence Saccade control during head or body movement Eye-hand coordination	Medial wall, Frontal Lobe
Prefrontal Cortex (PFC)	Short-term spatial memory of saccades Inhibits reflexive saccades	Midfrontal Gyrus
Cingulate Eye Field	Voluntary saccade moderation, secondary to motivation effects	Anterior Cingulate Cortex

Visual System Neuropathology

The supranuclear control system is key to stabilizing images on the retina. Four component neural control networks comprise the supranuclear system: saccades, pursuits, vergence, and the vestibulo-ocular reflex.[12] Eye movement difficulties in progressive supranuclear palsy involve voluntary saccades, first affecting the vertical meridian. Impulses for voluntary saccades first originate in the frontal eye field or superior colliculus[12, 19] and are temporized, if necessary, by input from the supplemental eye field (body movement/eye-hand coordination)[20, 21] or prefrontal cortex (memory-guided saccades) (Table 3). The later substrates for saccadic eye movements are the Paramedian Pontine Reticular Formation (horizontal saccades) and the rostral interstitial nucleus of the Medial Longitudinal Fasciculus (vertical saccades). Neural degeneration at any point in this impulse, from initiation to execution, may induce saccadic latency and/or hypometria. Pathologically short latency to saccadic onset is likely due to frontal involvement, whereas lengthier latencies are more likely due to several paths involving the superior colliculus.[22] A saccadic sign, which is relatively unique to PSP, involves diagonal eye jumps: on diagonal trajectory, the eyes of the PSP patient tend to drift toward the horizontal midline, thereby indicating a reduction of vertical range of saccades.[23] Unwanted saccadic intrusions, another abnormal characteristic of PSP patients, point to *disinhibition* of a saccadic reflex—due to degenerative lesions in the prefrontal cortex and/or the substantia nigra.[24]

The voluntary pursuit pathway involves the parieto-occipito-temporal junction, as well as the frontal eye field.[12,19] Signs of pursuit degeneration are seen on the slow phase of nystagmus (for involuntary, or foveal, pursuit testing) as well as overt pursuit testing (pursuits break down to a series of saccades). As a reminder, the most significant deficit relating to pursuits in PSP is vertical gaze restriction.

Vergence anomalies in PSP may be supranuclear (comitant deviation in all positions of gaze), internuclear (yielding a "convergence paralysis," or bilateral adduction deficit), or infranuclear, yielding noncomitant strabismus. It is important to remember that in PSP all levels of oculomotor control may be affected by degeneration. As mentioned previously, the vestibulo-ocular reflex remains intact until the late stages of PSP.

Eyelid apraxia and other lid anomalies are induced by degeneration in blink control centers, particularly the frontal eye field, supplemental eye field, and basal ganglia.[25] In normal subjects, a voluntary blink yielded greater activity on functional MRI testing in the regions of the

supplemental eye field and posterior parietal cortex, whereas voluntary pursuits showed greater activity in the frontal eye fields and visual cortex.[25]

Other Sequelae of PSP

PSP sufferers show particular degradation of speech pattern. Speech has been described as "raspy," and "gutteral," yet the most distinguishing characteristic of speech is *pseudobulbar dysarthria*.[5, 26] The term "pseudobulbar" refers to the similarity of this speech product abnormality to one which would originate in the medulla's motor centers for speech. The dysarthria in PSP shows characteristic raising of the speech volume near the end of a sentence. Initiation of the swallowing reflex is impaired in the later stages of PSP.

Cognitive effects of the disease are variable. The nature of the dementia has been classified as both cortical and subcortical.[27] The frontal lobe and temporal lobe seem particularly affected, yielding difficulties with sustained attention, reading, "visual dyslexia," dysgraphia, and increased reaction time.[26,28,29,30]

Differential Diagnosis

PSP shares a host of symptoms with other degenerative disease processes, but none bears a more striking resemblance to PSP than does Parkinson's disease. Like PSP, Parkinson's disease patients tend to be elderly, but may be affected at younger ages than those who suffer from PSP. Both diseases share gait abnormalities, bradykinesia, and trunk / neck rigidity.[6, 31] Ocular similarities include slow saccades, decreased blinking, and lid apraxia. In contrast to PSP, patients with Parkinson's disease show greater difficulties looking up. They also exhibit greater saccadic latency than that of PSP patients.[6,10] The movement patterns of these two degenerative diseases vary as well; Parkinson's disease shows the characteristic shuffle and forward learning during mobility, while PSP patients are upright. They may show posture instability and fall backward as a result. The key features to keep in mind when trying to differentiate the relatively more rare PSP from Parkinson's are the following: I) Supranuclear gaze palsy; and II) History of balance problems and falls.[32] Table 4 illustrates similarities and differences between Parkinson's disease and progressive supranuclear palsy. PSP shares some visual and cognitive signs with other degenerative diseases, particularly Alzheimer's disease (large saccadic intrusions), corticobasal degeneration (greater latency of saccades versus saccade slowing in PSP), Creutzfeld-Jakob disease (slow saccades, vertical and horizontal), and Huntington's Chorea (slow saccades, with difficulty suppressing reflex saccades).[1,19]

Table 4. Similarities and differences between Parkinson's disease and progressive supranuclear palsy (PSP).

FEATURE	Similarities	PD Characteristic	PSP Characteristic
Age at Onset	Mid-age to elderly	Mean age 54 yrs	60 years and up
Progression	Gradual	Insidious	Rapid deterioration
Visual	Saccade hypometria Decreased blink rate	Increased saccadic latency	Vertical gaze difficulty Saccadic intrusions
Gait / Movement	Delayed initiation of movement Cogwheeling	Lean forward Shuffling steps	Fall back, or to one side
Speech	Affected in later stages	Garbled, muted	Pseudobulbar dysarthria
Affect	Flat, stare	Smoothed, mask-like	"Astonished," with Frontalis contraction
Therapy	L-Dopa (premier)	Reduction of symptoms	Only reduces initiation of movement difficulties and cogwheeling

Treatment

Drug therapies have shown little promise in slowing the degeneration in PSP. The most common drug treatment implemented with this patient group is L-Dopa, which is prescribed with success for Parkinson's disease. Up to half of the PSP patients on L-Dopa therapy may have exacerbation of supranuclear gaze palsy and other symptoms, and as such, it is not used aggressively within this population.[31] When L-Dopa is implemented, it tends to help alleviate any parkinsonian symptoms (bradykinesia, cogwheeling movements, balance difficulties). Physostigmine, alternately, has shown promise in reducing disinhibition of reflex saccades and improvement on neuropsychological performance—yet it has no effect on the pyramidal motor aspects of the disease.[33] Potential areas of therapeutic benefit may be discovered in research with trophic factors and free radical scavengers.[31] As such, there is no current therapeutic regimen for PSP that ameliorates all symptoms concurrently.

Visual Rehabilitation

The supranuclear gaze palsy these patients experience undermines their ability to function efficiently in daily tasks. Unexplained difficulties when eating meals or frustration with reading may be experienced in early stages of the disease. Bifocals are contraindicated because of the impaired downward gaze ability of these patients. The first intervention that needs to be considered is transitioning the patient, from the bifocal, to a single vision lens for the appropriate working distance(s). To remediate vergence difficulty, vision therapy may be considered, yet it becomes less effective for controlling gaze difficulties as the disease progresses. Compensating treatment modalities, such as base-in prism for reading, become more useful for the convergence difficulty.[7]

Difficulties with downgaze tasks may be addressed simply by bringing materials into the field of primary gaze with reading stands or music stands. The recommendation of a closed-circuit television (CCTV) may be warranted, especially if image enlargement is helpful in reducing the frequency of blur and/or diplopia.[34] Alternatively, a prism prescription may be useful for moving inferior visual space to primary gaze position. Two prism modifications, worn over current spectacles, may be implemented for the sake of maintaining a patient's habitual reading posture (or for any other activity that may require looking inferiorly). Yoked base-down prism can be worn on a temporary basis for inferior-gaze tasks; it may also be incorporated into spectacles—up to 15 prism diopters—with a light tint to reduce glare from direct lighting sources above the patient's working area (Figure 1).[3,5] Yoked prisms are indicated when they diminish saccadic intrusions, by establishing a null point which dampens the frequency or amplitude of the disturbance.[35] The other effective means for re-

directing inferior views to a straight-ahead position is accomplished by the BedSpec[a] mounted prism lenses (Figure 2).[5] The BedSpec prisms are composed of two prisms with the base parallel to the frontal surface of each eye, with a mirror visor on the planar side of the prisms. The inferior field of view is internally reflected in the prism, with the resultant upright image directed through the base to the visual axis. Patients can also opt to watch TV with the BedSpec while supine, aiming their gaze toward the ceiling in this case (primary gaze position). The advantage to having an accessory piece for spectacle wear is that it may be used as needed (for mobility or cosmetic concerns).

A patient's complaint of irritation must be addressed, with reminders to "think blink," as well as to supplement their tear layer with artificial tears and ointments. Pending the severity of ocular surface disease, punctal plugs or other permanent alternatives should be considered to prolong the integrity of the cornea.

CONCLUSION

Patients presenting with a known history of PSP, or even where a neuro-degenerative disease is suspected, experience profound visual symptoms which affect the quality of vision. The most common visually-related symptoms PSP individuals experience are blur, diplopia, and dry eye. The findings most frequently found with progressive supranuclear palsy include: restriction of inferior and superior gaze, slow saccades, saccadic intrusions, and ocular surface disease secondary to decreased blinking. Presenting visual symptoms are commonly accompanied by a history of frequent falls or difficulty with balance.

The limitations of medical intervention to date for PSP dictate the palliative management of the patient's multisystem disturbances. Visual rehabilitation is an important piece of the management for patients with PSP, as there are few alternatives that can help with visual function in the patient's vocational and avocational interests. Our rehabilitative tools—base in prism, yoked prism, the BedSpec prism modification—provide a unique offense for the severe limitations encountered by patients with neuro-degenerative disease, particularly those patients afflicted with PSP.

Figure 1. Base-down yoked prism.

Figure 2. The BedSpec.

Acknowledgment

I wish to thank Dr. Irwin Suchoff for his guidance in the production of this article as well as for his mentorship in the clinical realm of head trauma rehabilitation.

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Date accepted for publication:
June 1, 2000