

## THE USE OF VERTICAL PRISMS IN PROGRESSIVE SUPRANUCLEAR PALSY (STEELE RICHARDSON OLSZEWSKI DISEASE)

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### Abstract

*Progressive supranuclear palsy is a degenerative disease, ultimately resulting in total external ophthalmoplegia. Problems of down gaze palsy may be the most distressing. Two cases are presented where 15 dioptre prisms were used base down to help these symptoms.*

**Key words:** *progressive supranuclear palsy, prism treatment.*

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Progressive supranuclear palsy (Steele Richardson Olszewski disease) is a degenerative condition of the central nervous system resulting in defective and ultimately absent saccadic and smooth pursuit eye movements. It affects individuals in the sixth and seventh decade<sup>1</sup> and although early reports suggested that males are more affected than females, Pfaffenbach *et al.*<sup>2</sup> in a study of 44 cases found an almost equal sex distribution.

The earliest ocular sign is usually defective downward voluntary gaze, followed by upward and horizontal gaze defects, and paralysis of convergence. Smooth pursuit movements also become affected, resulting in a striking pattern of apparent complete external ophthalmoplegia. Optokinetic responses are usually absent. Doll's head movements can usually be demonstrated until the very late stages of the disease, however, an associated feature of the disease, neck stiffness, may make this difficult to assess.

Pfaffenbach's study found Bell's phenomenon to be absent in all 13 cases tested, and, although

conjugate movements were markedly affected, only two cases out of 44 had a manifest squint.<sup>2</sup>

Other associated features are pseudobulbar palsy, dysarthria and, later, dysphagia and extensor posturing of the head and neck, with rubbery rigidity of the posterior cervical muscles. Progressive supranuclear palsy can be distinguished from Parkinsonism by the pattern of ophthalmoplegia, lack of tremor and the presence of pyramidal tract signs.<sup>1</sup> The signs are due to a progressive degeneration and gliosis of the subthalamic nuclei, substantia nigra, superior colliculi and dentate nucleus of the cerebellum, resulting within approximately five years (in the majority of cases) in dementia and death.<sup>1</sup>

Although there is no known treatment to affect the ultimate outcome of this disease, two patients seen at Concord Hospital have been helped by the use of base down prisms to overcome (partly) one of the main visual problems—being unable to look down. The presenting symptoms are often related to defective downward gaze because, not only is it

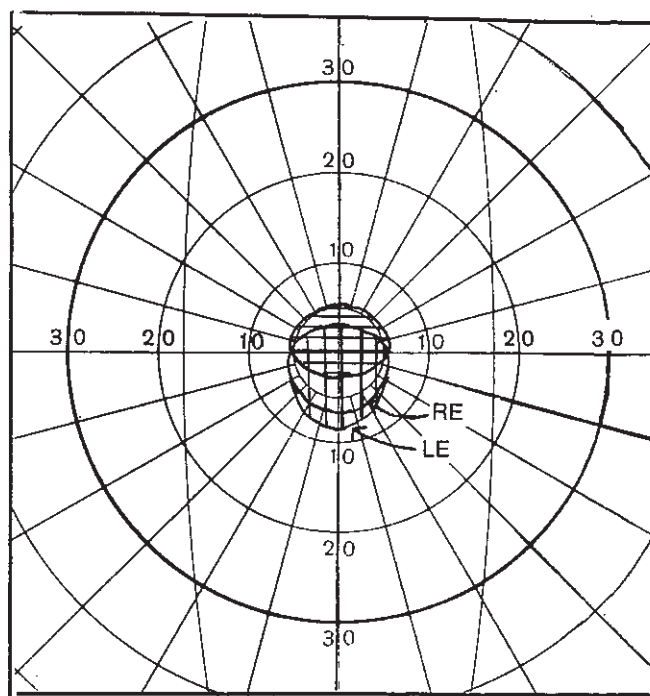


Figure 1: Field of eye movement.  
Without prisms: — With prisms: - - -

one of the first symptoms, it is also the most disabling when all eye movements are lost. Base down prisms (in front of each eye) have the effect of moving the whole visual field up, so that downward gaze is less required and visual performance related to objects in the patients inferior visual field is improved.

The first of the two men given this form of help, on being asked to sit down for his initial examination, missed the chair and fell over. He was found to have completely absent down gaze (saccadic and pursuit). There were no upward saccades, but some upward pursuit movements. Horizontal movements were also defective (saccades more so than pursuit) and he had no convergence. Although his neck was definitely stiff, doll's head vertical and horizontal movements could be demonstrated.

He was subsequently diagnosed as being at a moderately late stage of the disease, with consequent moderate dementia and dysphagia, so it was difficult to communicate with him. His

wife, however, reported that she had to guide him virtually all the time when he was walking.

15 prism dioptre base down press on prisms were attached to his glasses, and, although he showed initial disorientation, he adjusted to them remarkably quickly. On a subsequent visit his wife commented that it had taken about half a day for him to become used to them, and she felt that they were definitely helping him.

The second man, although showing almost complete vertical gaze palsy (saccadic and pursuit), severely limited horizontal movements and moderately affected convergence, was mentally more alert and could communicate well. His main visual complaint was of being unable to read, and he also had trouble eating because of his deficient downgaze. Although he was wearing bifocals, it was considered that it was worth trying vertical prisms, again 15 prism dioptre base down either eye, were applied as press on prisms. Subjectively he reported a definite improvement, and became adjusted to

them in less than a day. Although the prisms had the effect of bringing the bifocal segment into the main field of vision and the patient reported some distortion of distance objects, he was prepared to accept this for the improvement in other visual tasks. He has had a pair of light weight reading glasses ordered with this prismatic correction incorporated.

A field of fixation was plotted for each eye using the Goldmann perimeter with the patient being asked to follow the target and report when he felt that he was no longer able to look directly at it. This was done with each eye separately as his defective convergence meant that, at the close range, the target was horizontally double.

Figure 1 shows the severe limitation of eye movements in all directions, but with the prisms allowing more direct fixation of targets on depression. It can be seen that the displacement of the visual field is only small, suggesting that prisms of greater power could perhaps be used. It is interesting to note that the amount of movement is a little better in the left eye.

This form of therapy has only been reported briefly in the literature. In Pffafenbach's study, three patients were given "special 90 degree glasses" which were reported as being "more bother than benefit". However, these were obviously much more powerful than the approximately 7 degree displacement in this study. Walsh and Hoyt<sup>3</sup> reported that "prisms with base down added to their reading glasses gave much relief" to two elderly males with the disease, but give no further details of the power used.

Thus, although offering only marginal help, vertical prisms do appear to be one positive way of assisting these people with down gaze palsies.

#### References

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