Article • Management of Diplopia from Multiple Cranial Nerve Palsies: A Case Report

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ABSTRACT

Background: In a patient with an acquired noncomitant deviation, recent-onset diplopia is a very distressing problem. Initial management includes correction of significant refractive error, occlusion to prevent diplopia, prescription of Fresnel prism to allow for binocular fusion and vision therapy. This case report will show the benefit of treatment.

Case Report: A patient was referred by his optometrist for the evaluation and treatment of recent-onset diplopia. Exam findings were remarkable for right cranial nerve IV and right cranial nerve VI palsies that presented as binocular diplopia. The patient was followed over the course of 3 months, during which occlusion and Fresnel prism were used in the management and treatment of his diplopia.

Conclusion: This case illustrates how optometric management methods facilitated the recovery of a patient with cranial nerve IV and VI palsy.

Keywords: cranial nerve palsy, diplopia, Fresnel prism, occlusion, strabismus

Introduction

Cranial nerve IV innervates the superior oblique muscle, which depresses the eye in adduction and intorts the eye. It is the only nerve that exits the brainstem dorsally, with the longest course. Cranial nerve IV also decussates so that its nucleus innervates the contralateral superior oblique muscle. Common complaints of cranial nerve IV palsy are vertical diplopia, typically oblique in nature, and difficulty with downgaze that worsens with adduction. Common causes of acquired cranial nerve IV palsy may include head trauma, metabolic disease, microvascular infarction, tumors, multiple sclerosis, or infectious disease.[1]

Cranial nerve VI also has one of the longest intracranial courses. Cranial nerve VI innervates the ipsilateral lateral rectus muscle and ultimately travels to the contralateral medial rectus muscle. Cranial nerve VI palsies result in esodeviation and ipsilateral abduction deficits. The more frequent causes of cranial nerve VI palsies in adults are vasculopathic infarction, trauma, cerebral spinal fluid infection or inflammation, mass lesions, increased intracranial pressure, multiple sclerosis, stroke, or congenital reasons.[1]

The sixth and fourth cranial nerves are most commonly affected in cranial nerve palsies.[1] Acquired palsies involving one or more cranial nerves warrant further examination. A referral to the primary care provider and to a neurologist with a request for further laboratory testing and neuroimaging should be part of the patient's management. The lesion may affect an area involving more than one cranial nerve, such as the brainstem or the cavernous sinus. Further investigation is required to determine whether one or multiple lesions are causing the clinical signs.[1]

All optical and surgical treatments could be considered to alleviate the distressing problems of diplopia due to an acquired strabismus. Initial optometric management should include the correction of significant refractive error, occlusion, prism, and vision therapy with the appropriate co-management

to diagnose and treat the underlying cause. Strabismus surgery could also be an option, particularly in large-angle deviations that persist after the underlying problem has been addressed. However, because binocularity has a cortical basis, strabismus surgery may not reinstate normal binocular function.[2] Furthermore, appropriate compensatory lenses and vision therapy techniques are much less invasive means of treatment. These optometric treatment techniques provide the potential to restore proper visual function and sensorimotor fusion as the patient progresses.

Case History

A 48-year-old Asian male was referred to our office by an optometrist for the evaluation and treatment of recent-onset diplopia. The patient's chief complaint was double vision, diagonal in orientation, worse in right gaze, occurring 2 to 3 weeks prior, with coexisting symptoms of headaches, asthenopia, and instability of print when reading at near. Prior ocular history was unremarkable. He associated his visual symptoms with sudden-onset facial numbness on his right side after an acupressure appointment 3 months earlier. He wore a full-time patch over his right eye to resolve the diplopia.

The patient had undergone neurological evaluations with two normal MRIs 3 months previously (1.5 weeks prior to the eye exam) to rule out causes for the facial numbness after a sudden onset of diplopia. Other than bilateral photorefractive keratotomy for high myopia 6 years prior, ocular and medical health was unremarkable during recent medical and ophthalmological examinations, with no history of head injuries or trauma. He also denied a family history of ocular disease.

Case Report

Initial Exam #1

At the initial examination, unaided distance visual acuities were right eye 20/20 and left eye 20/25; near visual acuities were right eye 20/25 and left eye 20/20. Subjective refraction was right eye -0.25-0.25x030 (20/20) and left eye -0.25 DS (20/20). The patient also presented with a minimal compensatory left head tilt and a slight head turn favoring left gaze. Cover testing showed a constant right hypertropia of 5^{Δ} at distance and 2.5^{Δ} at near, with an esotropic component measuring 10^{Δ} at distance and 2^{Δ} at near. Extraocular muscle testing showed restriction of the right lateral rectus muscle in abduction. Near point of convergence was 5 cm break and 12 cm recovery. Park's 3 step testing with Maddox rod showed a right hyperdeviation greater in left gaze and right head tilt. Worth 4-dot testing showed constant uncrossed diplopia with a right hyperdeviation at 20 feet and 16 inches. He was unable to appreciate random dot or lateral disparity stereopsis on the Randot stereotest. His fine saccades on the King-Devick test for reading was reduced in speed and accuracy, totaling 109 seconds with 5 errors (age norms are <52 seconds and <2 errors). Visual midline shift testing showed a right, anterior visual midline shift.

Pupils were equal, round, and reactive to light with no afferent pupillary defect. Color vision was normal. Automated 24-2 screening visual fields were full and free of defects in each eye. Non-contact tonometry findings were normal, measuring right eye 11 mmHg and left eye 10 mmHg. Slit lamp biomicroscopy showed normal lids and lashes, white and quiet conjunctivas, clear corneas, deep and quiet anterior chambers, angles of grade 3 nasally and temporally, flat irises, and clear lenses in both eyes. Fundus examination showed clear media, cup-to-disc ratios of 0.35/0.35, distinct and healthy optic nerve rim tissue, clear maculae, normal retinal vessel caliber, and normal retinal health in both eyes.

Prism of 6^{Δ} base down and 10^{Δ} base out over the right eye allowed the patient to maintain

single vision. Using the Pythagorean theorem, calculations based on clinical measurements showed a resultant prism power and direction of 11.7$^{\Delta}$ at 211°.

$$P = \sqrt{(H^2+V^2)}$$
$$P = \sqrt{(10^2+6^2)}$$
$$P = 11.7$$

$$Tan\ \theta = V/H$$
$$Tan\ \theta = (-6/-10)$$
$$\theta = 31° (+180°) = 211° \text{ (in the 3rd quadrant)}$$

However, trial framing 9$^{\Delta}$ of Fresnel prism at 45° in the base-down and base-out position over the right eye (or 225°) produced the best results in visual comfort and eliminated diplopia.

Differential Diagnoses

- Myasthenia gravis, a neuromuscular disease, may present with variable deviation and lid involvement that worsens with fatigue.
- Duane retraction syndrome type I is usually congenital and presents as an abduction deficit, lid narrowing, and globe retraction in adduction.
- Brown syndrome is usually congenital and presents as a limitation of elevation in adduction.
- Orbital apex syndrome may be caused by lesions that involve multiple cranial nerves and may present with double vision, ptosis, proptosis, optic neuropathy, facial pain, and/or numbness.[3]
- Lyme disease is a tick-transmitted multisystem inflammatory disease that may include Bell's palsy and various ocular manifestations, such as neuroretinitis, multiple cranial nerve involvement, optic atrophy, and disc edema.[4,5] It is not the likely cause of this patient's visual issues since he denied travelling to any endemic areas for Lyme disease.

We diagnosed a cranial nerve IV palsy based on the right hyperdeviation that was greater in left gaze and with right head tilt, as measured on Park's 3-step testing by Maddox rod. The diagnosis of a cranial nerve VI palsy was based on the right esotropia and the abduction deficit.

Treatment

Nine prism diopters of Fresnel prism (equivalent to 6.4$^{\Delta}$ base down and 6.4$^{\Delta}$ base out) was applied over the right lens of the patient's current single-vision distance spectacles (OD +0.25-0.50x040 and OS plano-0.25x090) at 45°. A translucent patch was given to use as needed. Fit-over sunglasses were recommended for outdoor UV light protection. Smooth pursuit and visual tracking activities were prescribed for vision therapy at home to increase movement and range of motion of the right eye. A 4-6 week follow-up was scheduled to monitor. Maintaining follow-up appointments scheduled with his neurologist was also encouraged.

Exam #2: 1-month follow-up

The patient reported marked improvement of his symptoms at the one-month follow-up, having less frequency of diplopia, smaller image separation, and complete resolution of headaches. Without his prism glasses, he experienced brief periods of single vision in the morning that became constant, diagonal diplopia at all distances as the day progressed. The patient reported that he no longer needed to use the patch with the prism glasses and that the prescription now felt too strong.

Aided distance visual acuities were OD 20/25- and OS 20/20. Cover testing through the prism glasses was orthophoria at distance and near. Unaided cover testing showed intermittent right esotropia of 2$^{\Delta}$ and right hypertropia of 4$^{\Delta}$ at distance. Near cover testing showed a hyperphoria of 4$^{\Delta}$ and an esophoria of 4$^{\Delta}$. Extraocular muscle testing still showed

restriction of the right lateral rectus muscle in abduction but with improved range of motion. Near point of convergence was to the nose. Stereopsis showed 400 sec of lateral disparity, but there was no random dot stereopsis. The King-Devick test now showed a normal saccade speed of 36 seconds with no errors.

Without the prism glasses, Worth 4-dot testing showed constant, uncrossed diplopia at 20 feet and 16 inches. Testing with Park's 3-step subjective responses showed a right hyperdeviation that was slightly greater in left gaze and right head tilt.

Pythagorean theorem calculations based on clinical measurements showed a resultant prism power and direction of 4.5^{Δ} at 243°.

$$P = \sqrt{(H^2+V^2)}$$
$$P = \sqrt{(2^2+4^2)}$$
$$P = 6.3$$

$$Tan\ \theta = V/H$$
$$Tan\ \theta = (-4/-2)$$
$$\theta = 63° (+180°) = 243°$$

The patient had responded well to the initial Fresnel prism treatment but now needed an adjustment. During in-office evaluation, 6^{Δ} at 60° in the base-down and base-out direction over the right eye (or 240°) was the most comfortable and eliminated diplopia.

Diagnosis

Binocular diplopia
Right cranial nerve IV palsy
Right cranial nerve VI palsy
Visual midline shift syndrome

Treatment

The previous prism was replaced with 6^{Δ} with the base oriented at 60° (equivalent to 5.2^{Δ} base down and 3^{Δ} base out). The patient was also reminded to wear fit-over sunglasses for outdoor UV light protection and to perform daily smooth pursuits with visual tracking activities at home. A one-month follow-up appointment was scheduled. The patient also had an appointment scheduled with his neurologist within the week.

Exam #3: 2-month follow-up

The patient reported further improvement at the two-month follow-up, experiencing much less diplopia. On the day of the exam, he was not diplopic and felt that his prism glasses were too strong. The patient also reported that his laboratory tests came out negative for myasthenia gravis and autoimmune disorders during recent visits to his neurologist and primary care physician.

Aided distance visual acuities were right eye 20/25 and left eye 20/20. Cover testing through the Fresnel prism prescription was orthophoria at distance and near. Unaided cover testing showed orthophoria at distance and 3^{Δ} of right hyperphoria and 6^{Δ} esophoria at near. Extraocular muscle testing was full in all gazes; no restrictions were noted. The near point of convergence was an 8 cm break with a 30 cm recovery. The King-Devick test showed a normal saccade speed of 36 seconds with 1 error. Prism bar horizontal distance vergences measured BO x/8/1 and BI x/6/1. Horizontal vergences at near were BO x/8/0 and BI x/6/0. Prism bar vertical vergences at distance measured right BD 5/7/5 and BU 2/1/0. Vertical vergences at near were right BD x/9/7 and BU x/1/0.

Diplopia was reported on Worth 4-dot testing; a right hyperdeviation and an esodeviation were present at 20 feet and 16 inches. Testing with Park's 3-step showed a right hyperdeviation, slightly greater in right gaze and right head tilt. Visual midline shift testing showed a right, anterior visual midline shift.

In-office evaluation showed that a Fresnel prism of 2^{Δ} at 30° in the base-down and base-out direction (or 210°) improved comfort and reduced visual symptoms.

Table 1. Types of Occluders: Advantages and Disadvantages[2] Reprinted with permission from OEP Foundation.

Types	Features	Advantages	Disadvantages
Bandage	Opaque	Total occlusion, convenient, effective, different sizes	Allergies to adhesive, poor cosmesis
Tie-on	Pirate/elastic patch	Total occlusion	Loose, moveable, difficult to wear with specs
Clip-on	Clips on to plastic frames, sizes 32-50	Partial occlusion can be given	Easily removed
Translucent	Tape, nail polish, frosted lens, optical blur	Degrades form resolution of nonamblyopic eye, fusion, cosmesis acceptable	Children can peek around lens
Filter	Neutral density	Breaks suppression, allows some fusion	Effective VA improvement uncertain
Contact lens	Opaque soft lens	Convenient, effective, acceptable cosmesis	Difficult handling, requires cleaning, expensive

Treatment

The previous Fresnel prism over the right eye was removed and replaced with a reduced Fresnel prism of 2$^{\Delta}$ at 30° (equivalent to 1$^{\Delta}$ base down and 1.7$^{\Delta}$ base out). A one-month follow-up was scheduled.

Follow-up #4

The patient cancelled this follow-up appointment. With the marked improvement of his diplopia and visual symptoms measured clinically and reported subjectively, it is hopeful that his symptoms had become minimal or completely resolved.

No definitive diagnosis was made, but based on the positive VMSS[6] and the negative lab tests and MRI reports, the most likely etiology of the nerve palsy was trauma sustained during the acupressure session that preceded the onset of his diplopia.

Discussion

Optometrists can effectively treat patients with disruptive symptoms from acquired paretic extraocular muscles using various options.

Occlusion

The initial management approach is directed towards eliminating diplopia. Prescribing an occluder can easily address this problem. The best type of occlusion depends on the diagnosis and specific treatment goals for each patient. Pros and cons of different types of occlusion techniques should be considered, as shown in Table 1.3. Total occlusion of the nonpreferred eye will eliminate binocular diplopia. Patches and rigid or soft occlusion contact lenses can also be prescribed when necessary. However, patching should be considered as temporary treatment if possible, as it prevents binocular fusion. For patients wearing spectacle correction, central or sector occlusion using translucent tape often provides relief from diplopia and may be more cosmetically acceptable. Treatment options engaging both eyes during visual tasks can result in improvement of binocularity and recovery of stereopsis.[2]

It is typically recommended that for cases of recent-onset diplopia from mild to moderate paresis, prism should be considered as the initial treatment if fusion can be maintained.[8] Fresnel prism, as opposed to ground-in prism, is a flexible, economical option that can easily be changed as needed. A retrospective study of 64 patients by Tamhankar et al. indicated that even patients with larger, combined, and noncomitant deviations should be prescribed Fresnel prism for diplopia.[9] Overall, 72% of their patients were reported to be satisfied with prism use, with satisfaction being the highest for patients with vertical diplopia. During follow-up from 2 months to 5 years, 47 patients (73%) continued with prism even when strabismus surgery was offered as a treatment option (23% of patients opted for strabismus surgery). These follow-up results were a strong indicator of prism effectiveness in the initial management of diplopia with the consideration of comitancy, magnitude of deviation, presence of significant

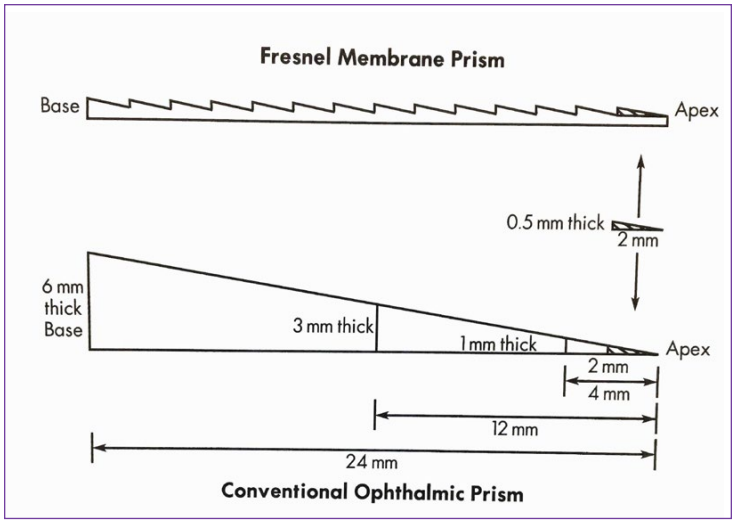

***Figure 1**. Fresnel membrane prism compared to conventional ophthalmic prism.[7] Reprinted with permission. Retrieved from www.fresnel-prism.com.*

motility restriction, head and neck posture, and degree of fusional ability, as well as patient motivation.[9] Proper education and goal setting during periodic follow-ups to assess appropriate prism prescription promoted satisfaction among prism users.

In another study by Gunton and Brown, approximately 80% of all adult patients with diplopia were satisfied with prism correction.[10] Patients with vertical diplopia and skew deviation/fourth nerve palsy had the highest satisfaction rates of 100 and 92%, respectively. Careful selection of patients for prismatic correction, management of patients' expectations, and continual monitoring of patient symptoms were critical to successful use of prisms.[10] Emphasis on how the condition has affected the patient's priorities and performance will determine the strategy for optometric management.[11]

Fresnel Prism

Fresnel prism can be modified, concurrent with changes in the magnitude of the deviation, during the recovery period. The initial power should be conservative, prescribing the minimum amount of prism that eliminates diplopia and reducing the power gradually if the paresis improves over time. Patients with non-comitant deviations resulting from a muscle paresis sometimes require prism only in the field of action of the affected muscle.[8] In many cases of paresis, different prisms can be prescribed for a specific task and distance to maintain fusion and promote recovery.[7] Separate single-vision lenses may be preferred over multifocal lenses for these cases.

Fresnel prisms are thin flexible plastic prisms that can be pressed onto spectacle lenses. They consist of a series of tiny prisms aligned with their bases in the same direction (Figure 1). Powers from 1 to 40 prism diopters are available and can be positioned in the horizontal, vertical, or even oblique directions. In cases of combined horizontal and vertical deviations, a Fresnel prism can be used to correct both horizontal and vertical components when applied in the oblique orientation to produce an effect equivalent to that of a single prism power.[8] There are various ways to determine oblique prism correction, but calculations using the Pythagorean theorem will be used in this discussion.

The pythagorean theorem applies the following equation for right triangles: $P = \sqrt{(H^2+V^2)}$. The angle θ for the direction of the base can be determined using the tangent, keeping track of the signs to determine the direction of the resulting prism in the proper quadrant: $\text{Tan}\,\theta = V/H$. In horizontal distances, the right direction is notated as positive and the left direction is notated as negative. A vertical distance upward is positive, and a vertical distance downward is negative (relative to the clinician's viewpoint, as shown in Figure 2). For the right eye, base-out prism is in the minus direction, and for both eyes, base-down prism is in the minus direction. The sign of the final calculation determines the resulting direction, unless the direction lies in quadrant 2 or 3, in which 180 must be added to obtain the correct answer.[8]

This prism calculation should be used as a guide, as trialing the resultant prism is a necessary step in determining the effect

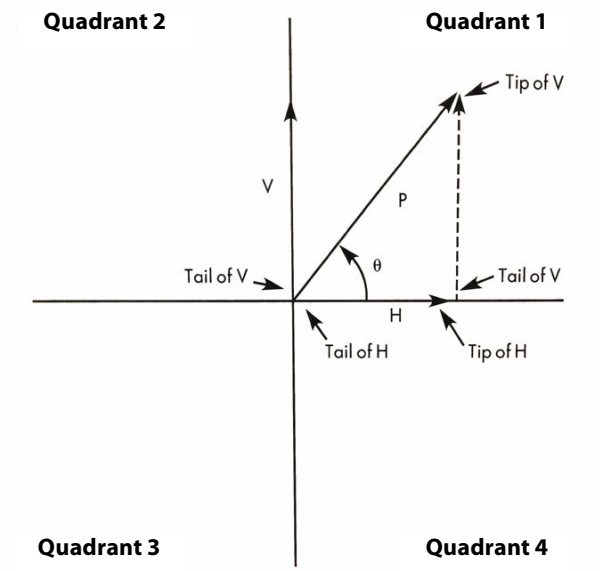

Figure 2. *Oblique prism with a power (P) from the horizontal (H) and vertical (V) directions.[7] Adapted from Clinical uses of prism: A spectrum of applications.*

on eye alignment and visual comfort with movement through free space. Based on this patient's subjective responses and clinical findings, such as cover testing and Worth 4-dot testing, loose prisms were trialed. The optimal prism based on overall considerations was then prescribed.

For comitant deviations, the prism power is generally split equally between the eyes to balance the thickness and weight. For paretic strabismus like the one in this particular case, the prism power was prescribed before the eye with limited movement. This allows prescription of less prism power than if it were split equally between the two eyes. When the prism is placed in front of the paretic eye, and the unaffected eye is fixating without prism correction, the smaller, primary angle of deviation can be neutralized.[12] If prism is prescribed in front of the unaffected eye, however, both eyes would be directed toward the action field of the weakened muscle under Hering's law.[12] Therefore, less prism correction is needed when prescribed for the paretic eye than if prism is placed in front of the unaffected eye.

The adjustment of prism power depends on the efficiency and stability of the patient's binocular vision system and whether symptoms have improved. Close monitoring with follow-up visits, typically within 4 to 6 weeks, is helpful in determining whether changes in prism power are appropriate.

Home vision therapy exercises can be given in conjunction with prism correction to prevent contracture of the ipsilateral muscle for patients with muscle paresis.[7] It is a beneficial management option because visual efficiency can be improved with training. Vision therapy can be considered when patients are unsuccessful with their prism prescriptions, when they are successful with correction but want to avoid dependency on prism glasses, or to enhance patient comfort in wearing prism correction. Prescribing smooth pursuits, visual tracking activities, ocular rotations, and stretches to further increase range of motion assists in preventing secondary ocular muscle contraction or adhesions, while promoting restoration of function in the paretic muscle.[7,11]

Conclusion

In cases of recent-onset diplopia due to multiple cranial nerve palsies, ruling out systemic and neurological causes is of primary importance to diagnose and treat the underlying cause. Numerous optometric treatment options are available to eliminate the diplopia and to enhance binocular fusion following a thorough history and optometric examination. Based on the patient's diagnostic profile, treatment considerations such as spectacle correction, occlusion, prism, and vision therapy are important treatment considerations. Monitoring the patient with education on the dynamic recovery process will improve patient satisfaction and the success of treatment.

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