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Skew Deviation: Report of a Case Treated with Prismatic Spectacles

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ABSTRACT

Introduction: This is a case report of a skew deviation (a vertical or oblique misalignment of the eyes caused by damage to the prenuclear vestibular input to the ocular motor nuclei) in a patient who presented with symptoms of diplopia and vertigo.

Case Report: A 77-year-old Caucasian male presented to the Illinois Eye Institute with long-standing binocular diplopia in both the horizontal and vertical meridians. After both objective and subjective measurements of the patient's strabismic angle, as well as an objective assessment of his extraocular muscles, a diagnosis of skew deviation secondary to a small transient ischemic event was determined. Treatment for this patient was provided in the form of spectacles with both ground-in and Fresnel membrane prism.

Discussion: A patient presenting with a skew ocular deviation caused by stroke, multiple sclerosis, trauma, tumor, abscess, hemorrhage, or neurosurgical procedure presents many challenges. Alternative treatment options for skew deviation may include occlusion, the use of Botulinum toxin, surgery, and prism. The use of ground-in and press on prisms relieved many of the symptoms presented by this patient.

Keywords: Prism, skew deviation, vertical diplopia, vertical strabismus

Correspondence regarding this article can be emailed to msallen@ico.edu or sent to Dr. Megan S. Allen, Illinois College of Optometry, 3241 S. Michigan Ave., Chicago, IL 60616. All statements are the author's personal opinion and may not reflect the opinions of the College of Optometrists in Vision Development, Optometry & Vision Development or any institution or organization to which the author may be affiliated. *OVD is indexed in the Directory of Open Access Journals. Online access is available at http://www.covd.org.*

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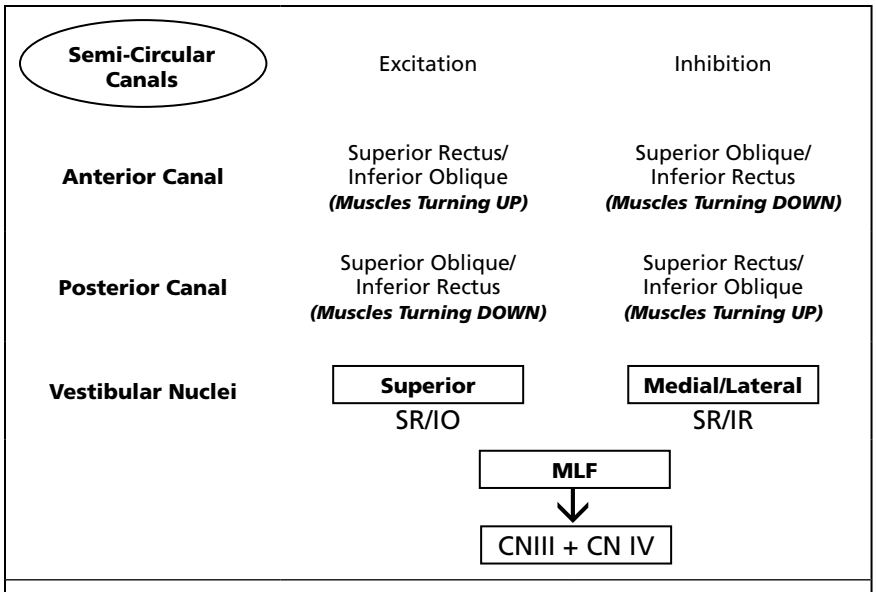
Introduction

Skew deviation has been defined as a vertical or oblique misalignment of the eyes caused by damage to the prenuclear vestibular input to the ocular motor nuclei.[1] This damage is typically seen in the area of the brainstem; however, some reports have also localized damage to the cerebellum. Damage is characteristically caused by stroke, multiple sclerosis, trauma, and tumor; as well as abscess, hemorrhage, or neurosurgical procedure. The majority of cases are seen with brainstem stroke association.[2] These cases often present with associated brainstem symptoms such as vertigo, hearing loss, ataxia, lack of coordination, and nystagmus. In skew deviation, the otolithic pathway is injured and this causes the patient to perceive the world as tilted.[1] Skew deviation can present as a comitant, noncomitant, or alternating deviation. A comitant deviation is caused by unilateral damage to the otolithic pathway in which both anterior and posterior semicircular canals of the inner ear are damaged equally. A noncomitant deviation occurs when the hyper deviation is greater in one lateral gaze than the other. This is caused by asymmetric injury to the unilateral otolithic pathways of the anterior and posterior semicircular canals. An alternating skew deviation occurs when the hypertropic eye reverses in each lateral gaze and is caused by bilateral damage to the otolithic pathways.[1,3]

The primary function of the vestibulo-ocular system is to maintain stable fixation during head movement. The system starts within the labyrinthine receptors of the semi-circular canals, located within the inner ear. The semi-circular canals sense angular acceleration and send excitatory and inhibitory innervations to the extra-ocular muscles (Figure 1). The superior extraocular muscles act as intortors of the eye, while the inferior extraocular muscles function to extort the eye. Lesions of the semicircular canal pathways can cause an imbalance of the vestibular tone, thus resulting in the cyclotorsion and vertical deviation seen in a skew deviation.[1,3-5]



The vestibulo-ocular system starts with the semi-circular canals located within the inner ear. They send signals to the extra-ocular muscles thru the vestibular nuclei and cranial nerves III and IV to create both intorsion and extortion. These muscles also act to elevate and depress the eye. They work together and act in opposite forces to create visual balance while the head is tilted. This is called the ocular tilt reaction, and it is affected in patients with skew deviation.

Figure 1

Clinical characteristics of skew deviation consist of a vertical strabismus that presents acutely with or without a horizontal component. The vertical deviation presents as a hypotropia that is most commonly found on the same side as the brainstem or cerebellar lesion. Additionally, and clinically important, the hypotropic eye extorts, or rotates away from the midline, and the hypertropic eye intorts, or rotates toward the midline. This phenomenon differentiates the skew deviation from a superior oblique palsy.[1,4,6] A further clinical sign seen in skew deviation is a head tilt.[4,6] The tilt is usually away from the hypertropic eye; however, not all cases follow this rule.[5] The head tilt is in response to the subjective visual vertical. In other words, the patient's head is tilting because they perceive that to be upright, versus a head tilt in response to vertical diplopia. This also differentiates a skew deviation from a superior oblique palsy.[1,7]

Case Report

A 77-year-old Caucasian male presented to the Primary Care Service of the Illinois Eye Institute with complaints of diplopia. The diplopia was binocular, oblique, occurred with and without glasses, and was worse in the distance. It started about 20 years prior and was relatively stable according to the patient. He stated that he had prior relief from spectacle prism, but often seemed to "outgrow" each pair of glasses. This patient's ocular history was remarkable for mild cataracts, the left eye having progressed more than the right. His last eye examination and prescription had been given only five months prior at a different eye clinic.

The patient reported that his medical history was remarkable for a genetic heart defect, for which he had had multiple pacemakers implanted. He denied any small vessel disease, such as hypertension or diabetes. He did suffer from vertigo, which was being managed by meclizine. In addition, he suffered from hearing loss in the left ear, for which he used a hearing aid. Both the vertigo and hearing loss have been present for the past 30 years.

Upon presentation to the primary care clinic, distance visual acuities were taken through his last prescribed bifocal spectacles (Bifocal #1 in Table 1). Acuity of 20/25- was found in the right eye, and 20/40- was found in the left. At 40 cm, acuities of 20/40+ in the right and 20/40- in the left were obtained. Pupils were equal and reactive to light to a grade of 3+ in both the right and left eye, and no afferent pupillary defect was present. Extraocular muscles, as assessed by the primary care optometrist, were documented as showing full range of motion; however, diplopia was noted in all fields of gaze. An esotropic and hypertropic posture was seen, but no magnitude was given. The patient was assessed as having long-standing diplopia from an unknown muscle paresis and was referred to the Pediatric/Binocular Vision Service for further assessment, as well as possible prism prescription.

The patient returned for a strabismus evaluation in the Pediatric/Binocular Vision Service with similar complaints as stated at the primary care encounter. A right head tilt was noted throughout the entire examination. The patient denied any symptoms of oscillopsia, as well as any systemic symptoms of giant cell arteritis or myasthenia gravis. In addition, it was also discovered that this patient had been seen by a neurologist three years prior at which time an MRI was performed. The patient was told he had evidence of a

past small stroke, which could account for the symptoms of vertigo and hearing loss.

Upon examination, similar visual acuity measurements to those from the primary care encounter were obtained. A distance cover test was performed unaided and showed a constant left esotropia of 25$^{\Delta}$ with a left hypotropia of 7$^{\Delta}$. This was measured using a 20/60 distance target. Cover test done at 40 cm resulted in a reduction of the left esotropia to 16$^{\Delta}$ with the left hypotropia being reduced to 4$^{\Delta}$. This was performed using a 20/50 near acuity target (Table 2). The extraocular muscles were assessed as being restricted in the left gaze, as well as the superior gaze to a mild degree of -1 under-action. This was seen in both ductions and versions.

Table 1.

	Spectacles/Lensometry		Ground Prism
Bifocal #1	OD Pl OS +1.00 -1.50 x 160	Add +2.75 Add +2.75	7Δ BO,7ΔBD 7Δ BO,7ΔBU
Bifocal #2	OD -0.25-0.25 x 150 OS -0.25-1.00 x 040	Add +2.75 Add +2.75	4ΔBO, 4ΔBD 4ΔBO, 4ΔBU
Distance #3	OD Pl OS +0.50 sph		2ΔBO, 2ΔBD 2ΔBO
Distance #4	OD +0.50 sph OS +1.00 -0.75 x 090		4ΔBO, 4ΔBD 4ΔBO, 4ΔBU
Near #5	OD +2.00-0.25 x 150 OS +2.00 sph		

Table 2.

Refraction:		**Deviation:**	
OD +0.25-0.50x 150 +2.50		Dist: 25Δ CLET, 7Δ LHypo	
OS +0.75-0.75x 040 +2.50		Near: 16Δ CLET, 4Δ LHypo	
Trialed	**Ground**	**+ Fresnel = Total**	**VA**
1. Distance Only **OD** +0.50 sph **OS** +1.00-0.75 x 090	4Δ BO, 4ΔBD 4Δ BO, 4Δ BU	8Δ BO (16Δ BO, 8Δ Vertical) 2Δ BΔ (12Δ BO, 2Δ Vertical) 12Δ BO	20/25- 20/60-
2. Near Only **OD** +2.00-0.25 x 150 **OS** +2.00 sph	No Ground	2ΔBU (12Δ BO, 2Δ Vertical) 12Δ BO	20/40+ (OU)
Final Rx	**Ground**	**+ Fresnel = Total**	**VA**
1.**OD** +0.25-0.50x 150 **OS** +0.75-0.75x 040	4Δ BO, 4ΔBD 4Δ BO, 4Δ BU	8Δ BO (16Δ BO, 8Δ Vertical)	20/25- 20/50
2.**OD** +2.75-0.50x 150 **OS** +3.25-0.75x 040	4Δ BO, 4ΔBD 4Δ BO, 4Δ BU	4Δ BO (12Δ BO, 2Δ Vertical)	20/30 (OU)

Objective measurement of comitancy was performed using a cover test in all diagnostic action fields. It revealed the left esotropia to be comitant within 5$^{\Delta}$ in all fields of gaze. However, the hypertropia appeared to increase in left gaze. Subjective testing was performed using the Maddox rod method with the results not matching the objective findings. This difference was most likely due to confusion during the testing and the patient's difficulty in maintaining an upright head posture. The Parks three-step test was performed and revealed under-action of the left superior rectus. The double Maddox Rod test was performed to subjectively assess the torsional component of the deviation. The patient reported excyclotorsion of the hypotropic eye with no subjective torsion being reported in the hypertropic eye.

Cranial nerve III palsy was ruled out due to the adequate function of the other ocular muscles controlled by both the superior and inferior divisions, as well as by the fact that there was no pupil involvement in this patient. With a vertical diplopia, it is also important to rule out a cranial nerve IV palsy. This could be ruled out due to the clinical sign of extortion in the hypotropic eye versus the hypertropic eye, as well as by the fact that the superior oblique was not isolated on the Parks three step.

A diagnosis of noncomitant skew deviation was made based on the findings surrounding the vertical misalignment of the eyes. These include the fact that the hypotropic eye showed extorsion, the patient demonstrated a head tilt posture, and the patient had a history of a small midbrain infarction that resulted in other brainstem symptoms including vertigo.[8]

Treatment for the skew deviation was achieved by using a method of combining both ground-in prism with temporary Fresnel prism in multiple pairs of glasses. Lensometry was performed on the various prescription glasses that were brought in by the patient. In order to have an accurate trial of prism treatment, the pair of glasses with the closest refraction to that found during the current examination was used along with temporary Fresnel prism to completely neutralize the remaining deviation (Table 2). The amount of Fresnel prism was determined by an in-office trial using a red lens and a transilluminator. Loose prism

was held over the patient's spectacles while viewing the transilluminator thru a monocular red lens. A small amount of prism was chosen at first and increased in small increments until the patient subjectively achieved fusion with the red lens. This allowed the patient to use his vergence skills and ultimately allowed for a smaller amount of prism to neutralize the deviation. This amount of prism was then used in the form of a Fresnel lens and applied over the spectacles. Two pairs of spectacles, one for distance viewing and the other for near, were sent home for a practical trial. The patient returned one week later and reported complete resolution of the diplopia while using the lenses, but he felt like his vision was slightly worse. In order to improve this complaint, a more accurate prescription was written in which the most current refraction was used along with the combination of ground prism and Fresnel prism that had been tried. The distance pair of spectacles contained a total of 4ΔBO each, as well as 4Δ each of vertical prism. An additional amount, 8ΔBO, of horizontal Fresnel prism was added to the non-dominant eye. This gave a total of 16ΔBO horizontal and 8Δ vertical that was neutralized by the prism. For a distance deviation measured at 25Δ esotropia and 7Δ L hypotropia, this resulted in 8Δ of horizontal divergence that the patient was able to overcome. Visual acuity was 20/25 in the dominant right eye, and 20/50 in the non-dominant left eye. The patient was very satisfied with this result. A similar technique was used for the near spectacles. (Table 2) In previous spectacles, the maximum amount of horizontal deviation that was neutralized was 14ΔBO. (Table 1) This could have been tolerable; however that same pair had an additional 7Δ of vertical prism that created diplopia and the patient found them extremely heavy. The spectacles given at this follow up achieved the best visual acuity, were able to neutralize the diplopia, and were the lightest in weight.

The patient was asked to continue follow-up care with the ophthalmologist, but to return if he began to notice a change in his vision or became diplopic while using the glasses.

Discussion

The diagnosis of skew deviation was slightly confounded by the fact that the patient exhibited an esotropic posture that increased in the distance. This phenomenon resembles a cranial nerve VI palsy, which would affect abduction and the function of the lateral rectus muscle. In order to rule out a concomitant CN VI palsy, versions and ductions were performed and showed an equal mild restriction in abduction. In skew deviation, the vestibular input to the extraocular muscles is damaged, thus affecting the torsional aspect of the eye. The superior rectus and superior oblique muscles, the intortors, were therefore weaker due to vestibular damage. It was postulated that the esotropic posture came from the loss of the tertiary abduction function of the superior oblique muscle.

The inferior rectus, superior rectus and both oblique extraocular muscles exhibit three movement actions of the eye. The primary function of the superior oblique muscle is to intort the eye. With the loss of this function, the eye then extorts. The secondary function is to depress the eye upon adduction. This function remained intact for this patient. The tertiary function of the superior oblique muscle is to abduct the eye. With the weakening of this function, the eye had difficulty abducting and thus an esotropic posture was observed.

Prognosis

The prognosis for recovery from skew deviation has been stated as being favorable.[9] A retrospective study completed by Borruat in 1998 found that 70% of patients recovered spontaneously, meaning they had relief of diplopia, after a median time of 7.5 months.[9] The reason why some patients recover completely and others do not is not completely understood. In some instances, the prognosis for recovery may depend on the severity of the brainstem damage; however this is not always the case. For the 30% of patients who remain symptomatic, treatment can be found through the many options discussed below.

Treatment Options

Occlusion: Skew deviation can be treated in various ways. The most simple and temporary way to relieve diplopia is to simply have the patient occlude, or patch the non-dominant eye. This can be done with a variety of methods. A less noticeable and cosmetically appealing method, would be to use the Bangerter Occlusion Foils.[10] These are available in different visual acuity levels and are used to create a blurred image that makes the diplopia less perceptible. The advantage of the foil is that it appears fairly clear on the spectacles to the average observer. The disadvantage is that the patient must tolerate the vision decreasing to an acuity of 20/100 or worse. To

alleviate this problem, the occlusion foil is usually placed over the non-dominant eye. This option is ideal for acute conditions, or as a temporary solution before more permanent treatment.

Botulinum toxin: Botulinum toxin (Botox) injection is another option for the treatment and management of skew deviation. Botulinum toxin type A has been used in the United States for treatment of blepharospasm, strabismus, and hemifacial spasm since approval in 1989.[11] Botulinum toxin acts by selectively blocking the release of acetylcholine from the cholinergic synapses within the muscle. This action ultimately blocks the nerve impulses of the muscle.[12] This option is less invasive in comparison to surgery, but also poses some disadvantages. These include the temporary nature of the muscle denervation, ptosis, injection site reaction, as well as the possibility of not relieving the diplopia, or making it worse. The duration for which the toxin causes paralysis is dose dependant, as well as dependant on the individual.[12] Typical doses last around 6 weeks, so multiple procedures need to be performed for any duration of relief greater than 6 weeks. Limited studies have been performed looking at the benefits of botulinum toxin injection for the relief of a vertical strabismus;[11] however it can still be considered as a temporary treatment option. According to one article by Ripley[12], successful outcomes were reported after 1-2 injections, but the results are best in cases of heterophoria with little near-distance angle disparity.

Surgery: The most aggressive method of treatment for skew deviation would be the surgical option. This option should be deferred for at least 8 months or longer due to the possibility of recovery and the variable nature of this condition. Different surgical options have been studied and trialed including horizontal or diagonal transposition of the vertical rectus muscles, oblique muscle surgery, vertical rectus muscle resections, or a combination of all three.[13,14] Adjustable sutures are also often used to allow the surgeon to adjust the muscles with the aid of the patients' subjective responses. Vertical rectus recession or resection has been stated to be the most common procedure, as well as the most successful.[14] Oblique muscle surgery is often associated with poorer outcomes.[14] Individual surgeons may approach this condition differently, based on experience. According to Nemoto, surgery should be performed on only one muscle at a time in order to prevent overcorrecting.[13] Further studies are still examining the stability and effectiveness of these procedures for the treatment of skew deviation over longer periods of time.

Prism: Prescribing prism for the treatment of skew deviation is a simple and beneficial way to relieve the patient's diplopia. It is a flexible approach that allows the prescription to be changed if the deviation proves to be unstable. This non-invasive simple approach can be done using two different forms of prism, or a combination of both. Disadvantages to ground-in prism include an increase in weight and thickness, increase in cost, and a limit to the amount of prism diopters allowed.[15] When ground-in prism is used clinically, the guideline of no more than 10 prism diopters per lens is applied. The amount of prism is usually split between the two lenses, unless the amount is less than two prism diopters.[15] The advantage to ground-in prism is that it does not alter the clarity or contrast sensitivity through the lenses.

Fresnel prism is a temporary plastic film that overlays the spectacle lenses. It is composed of tiny prismatic elements that are aligned with their bases in the same direction.[15] This allows the prism to be available in much larger increments without increasing the thickness or weight of the spectacles. It is also a temporary option, so it can be used to trial prism before ground prism is used. This option works well for temporary or continuously changing deviations such as those seen with myasthenia gravis or certain cases of Graves' ophthalmopathy. Fresnel prism also has the advantage of being altered and cut, within the office setting, to specific fields of gaze. For example, it can be applied only to the bifocal region if the diplopia only occurs at near. It is available in various increments of prism from 1-40 prism diopters. Fresnel prism holds the disadvantage in that it causes an increase in light scattering from the prismatic grooves, and thus causes a decrease in visual acuity and contrast sensitivity. Increasing prismatic power decreases the visual acuity (Table 3).[16]

Treatment for this Case:

With this patient, all treatment options were considered. Due to the fact that he had changed spectacle prescriptions frequently, it was determined that the patient demonstrated a fairly unstable deviation. Because of this, a surgical option was not chosen at this time. A Botox injection was discussed; however, he decided to attempt a new prescription of prismatic spectacles before any further invasive procedure was considered.

Table 3.

Fresnel Prism	Decrease in Acuity
5Δ	1 line decrease in VA
15Δ	3 line decrease in VA
20Δ	4 line decrease in VA
30Δ	5 line decrease in VA

In order to maximize the advantages of both options of prism treatment, ground prism and temporary Fresnel prism were used in combination. This allowed the spectacles to maximize the amount of ground prism in each lens without making the glasses too heavy or uncomfortable. It also allowed for a smaller amount of Fresnel to be used, thus decreasing the amount of visual distortion. The Fresnel prism was also found to be advantageous because the amount could be changed as the patient's condition changed. Both a distance only pair and reading only pair of spectacles were used, due to the difference in deviation at the two distances. In the final prescription, the Fresnel prism was placed over the non-dominant eye in order to ease the amount of distortion noticed binocularly.

Conclusion

Skew deviation is a vertical misalignment of the eyes most commonly seen after brainstem or cerebellar injury. It causes a triad of symptoms consisting of vertical diplopia, head tilt, and a torsional rotation of the eyes in which the hypotropic eye extorts away from the midline. In addition, patients with skew deviation may also present suffering from other brainstem related symptoms such as vertigo, hearing loss, or impairment of pain and temperature sensation. While many cases of skew deviation resolve spontaneously, many patients remain symptomatic. It is important to maintain close follow-up to monitor for changes in the deviation and to make possible changes in the treatment method. Treatment options include occlusion, prismatic spectacles, botulinum toxin injection, and muscle surgery. This case explains the successful relief of diplopia through the combined use of ground prism with temporary Fresnel prism. This is a simple and practical alternative for the optometrist to provide symptomatic relief from skew deviation.

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