CASE REPORT

Optical Management Using Monovision and Yoked Prism for Acquired Strabismus and Nystagmus Secondary to a Neurodegenerative Disease

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ABSTRACT

Acquired involuntary eye movement disorders, including noncomitant strabismus, nystagmus, and saccadic dyskinesia, are common ocular manifestations of many neurodegenerative diseases. These patients may experience visual symptoms, such as blurred vision, diplopia, and oscillopsia, which can significantly impact their use of vision. The goal of the management for these patients is to reduce the visual symptoms using any combination of available management strategies. This case report discusses the effective optical management using the combination of spectacle monovision correction and yoked prism to improve visual symptoms in a patient with olivopontocerebellar atrophy.

Keywords: Diplopia, monovision, neurodegenerative disease, optical management, oscillopsia, yoked prism

INTRODUCTION

Olivopontocerebellar atrophy (OPCA) is a central neurodegenerative disease that fundamentally affects the cerebellum and pontine, inferior olivary, arcuate, and pontobulbar nuclei.^{1,2} Like many other neurodegenerative conditions, this condition often impacts the visual systems, including fixation, eye alignment, and movement.^{3,4} As a result, ocular manifestations, such as noncomitant (incomitant) strabismus and involuntary abnormal eye movements, may occur. Due to the acquired nature of the disease, these patients commonly become symptomatic with blurred vision, diplopia, and/or oscillopsia, which significantly impact their use of vision and quality of life. For these patients, it is important to manage their visual symptoms in addition to managing the underlying neurodegenerative conditions causing such abnormal ocular manifestations. The available options include spectacle correction, contact lens wear, vision therapy, prism compensation, medication management, botulinum toxin injection, and surgery. Unfortunately, effective management strategies may vary and be

limited based on the type and nature of ocular manifestations. Another challenge is that most published cases and studies of management strategies are focused on congenital conditions rather than acquired conditions. Despite these challenges, the best attempt should be made using any combination of the available options, since patients with severe visual symptoms appreciate any improvement. In many cases, optical management is the most desirable option, as it is relatively less invasive, safe, and has the advantage of easy modification or withdrawal if it is not effective. This report demonstrates a successful case of improving visual symptoms using a combination of medication, spectacle monovision prescription, and yoked prism in a patient experiencing blurred vision, diplopia, and oscillopsia secondary to OPCA.

CASE

A 66-year-old Caucasian male with the known diagnosis of olivopontocerebellar atrophy (OPCA) was referred to the Veterans Affairs polytrauma optometry

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clinic by a neuro-ophthalmologist for a vision assessment. He was diagnosed with OPCA at age 34 and had been monitored closely since. Consequently, he was experiencing various symptoms, including ataxia, gait difficulties, and speech difficulties. His visual symptoms included light sensitivity, blurry vision, constant horizontal diplopia, and oscillopsia, with or without his current glasses. According to the patient, up-gaze was his preferred gaze as the oscillating images became relatively more stable, although it forced him to look over his glasses. It was reported that these visual symptoms had been stable for 3 years with good compliance of prescribed baclofen.

The patient was unaware of any family history of OPCA, other neurodegenerative diseases, or any ocular conditions. The previous ocular diagnoses of the patient included large-amplitude, low-frequency nystagmus, multi-planar gaze-evoked nystagmus (GEN), approximately 20 prism dioptre alternating esotropia, and left hypertropia. These ocular manifestations were determined to be consistent with the patient's diagnosis of OPCA. Systemically, he was diagnosed and monitored for hypertension, type 2 diabetes, hyperlipidaemia, and post-traumatic stress disorder. His prescribed medication included albuterol inhaler, as needed, baclofen 10 mg, omeprazole 20 mg, oxybutynin chloride 5 mg, ranitidine 86 mg, simvastatin 20 mg, tamsulosin 0.4 mg, and vancomycin 50 mg. He was alert and oriented to time and place at the time of the examination.

The habitual glasses prescription was -0.50 dioptres (D), 6 prism dioptres (Δ) base-out (BO), 1.5Δ base-up (BU) right eye (OD), and -0.25 D, 13Δ BO, 1Δ base-down (BD) left eye (OS), with $+3.00$ D ADD both eyes (OU). He demonstrated a habitual slight head turn to the left and significant chin depression, which placed his gaze over the glasses most of the time. His visual acuity was measured in several different ways, although some results could not be attained due to the interference of severe oscillopsia. The aided distance visual acuity in primary gaze was $20/100^{+2}$ OD, unable to attain (UTA) OS, and OU. His aided near visual acuity in primary gaze was $20/60$ OD, OS, and OU. In addition, we attempted to measure the distance visual acuity in different gaze positions (Table 1). These measurements were obtained unaided. It was done because we considered the benefit of eliminating the induced prism effect caused by his current prism prescription was more valuable than the minimal visual acuity improvement the habitual glasses provided. The results were UTA OD, OS, and OU in primary gaze; $20/80^{+2}$ OD, $20/70^{+2}$ OS, and UTA OU in down-gaze. During the down-gaze measurement, the patient had to lift his chin up quite extensively to obtain the adequate resolution of the distance target. It was measured

$20/80^{+2}$ OD, $20/80^{+2}$ OS, UTA OU in up-gaze. The patient reported worsening of diplopia in this gaze despite it being his preference due to the least amount of oscillopsia. This is consistent with our experience with other diplopic patients who preferred to have the images widely separated than in close proximity, yet not fusible. The retinoscopy value was -0.50 – 0.50×180 OD, -0.50 – 0.50×010 OS. The final prescription determined by trial frame refraction was -0.50 D OD, -0.25 D OS, and $+3.50$ D ADD OU. The best-corrected visual acuity obtained with this prescription was $20/70^{-2}$ OD, $20/70^{-2}$ OS at distance and $20/50$ OU at near in up-gaze.

The patient could initiate saccades when he was asked to; however, the end position could not be maintained due to the interfering pendular nystagmus. The nystagmus was binocular, conjugate, and had a large amplitude and moderate frequency. It appeared to dampen slightly in both up- and down-gaze. In both right and left gaze, GEN was also present. Interestingly, occasional vertical nystagmus was also observed in the horizontal gaze positions. No latent component was observed.

Due to the constant abnormal eye movements, it was difficult to assess the ocular alignment by cover test or Hirschberg test. However, it was possible to appreciate the presence of constant alternating esotropia of approximately 20 – 25Δ that appeared larger at near. Additionally, a V-pattern was observed during the extraocular muscles (EOMs) testing. This test also suggested no apparent restriction of EOMs, although it was difficult to assess. With 20Δ BO prism in place, the patient reported constant alternation between fusion and uncrossed diplopia with both light and letter targets when viewing at distance. An additional horizontal prism caused crossed diplopia, and vertical prism caused vertical diplopia. Therefore, we were unable to obtain more than momentary fusion in any position of gaze.

Given the patient's unstable fusion ability, a spectacle monovision prescription was trial framed. The left eye was corrected for distance (-0.25 D) and the right eye was corrected for near ($+3.00$ D). With this prescription, the patient was able to ignore (or suspend) the right eye's blurry image when viewing distance. It was slightly more challenging to ignore the left eye's blurry image at near; however, the patient reported that it was not too bothersome. Visual acuity measured with this prescription was not compromised compared with his best-corrected visual acuity.

In order to shift the target of interest into the gaze that results in the greatest reduction of nystagmus to further improve his vision, oscillopsia, and head posture, BD yoked prisms* were trialled in addition to the monovision prescription. The patient did not notice any difference from his usual vision when both

*Yoked prism: Unlike compensating prism, yoked prisms are used in both eyes in the same direction and amount for the purpose of moving both eyes.

3Δ OU and 5Δ OU BD yoked prisms were demonstrated. Through several trials, the patient noticed the images were not moving as much with 7Δ OU and 10Δ OU BD yoked prisms. Although residual oscillopsia was present, he reported most stable vision with 10Δ BD. The patient did not prefer 15Δ BD yoked prism, as it caused increased oscillopsia after an initial dampening. Objectively, the nystagmus was still evident with 10Δ BD yoked prism; however, the frequency appeared to be dampened compared with when no prism was in place. The intensity of his abnormal head position of chin depression was reduced significantly, and the patient was no longer looking over his glasses. For the comparison purpose, 10Δ BD yoked prism was demonstrated with the patient's distance prescription alone; however, he preferred it with the monovision prescription. With the monovision prescription and 10Δ BD yoked prism in place, the visual acuities were 20/70⁻¹ at distance and 20/50 at near when measured binocularly. No further testing was done as the patient had a neuro-ophthalmology evaluation and dilated fundus examination 1 week prior to this visit.

The monovision prescription with yoked prism was dispensed for full-time wear. Anti-reflective coating and photochromic tint were added for his light sensitivity symptoms. The patient was informed about the adaptation period, and it was recommended that he occlude the left eye, which is corrected for distance, for a few minutes if he continuously had difficulty adapting at near. A Bernell clip-on occluder was dispensed for this purpose. He was instructed to return to clinic in 2 months for a follow-up visit, or sooner as needed.

TABLE 1 Aided and unaided visual acuities (VA) measured in different gazes at distance (20 ft).

	Gaze	OD	OS	OU
Unaided	Primary	UTA	UTA	UTA
	Up-gaze	20/80 ⁺²	20/80 ⁺²	UTA
	Down-gaze	20/80 ⁺²	20/70 ⁺²	UTA
Aided	Primary	20/100 ⁺²	UTA	UTA

UTA = unable to attain.

At the 2-month follow-up, the patient reported excellent compliance and adaptation with the new spectacle prescription. He was no longer bothered by diplopia when viewing at distance, and he was able to focus on the appropriate targets without a conscious effort. Although the temporary occlusion of the left eye was helping, the patient still found reading slightly challenging. However, he reported having minimal difficulty with any "spotting" tasks at near. The patient also appreciated the reduction of oscillopsia and improved abnormal head posture. He was also extremely satisfied with the improvement of visual clarity at both distance and near. Anti-reflective coating and photochromic tint were working effectively per the patient.

There was no change in his general health and the medication list compared with the initial visit. The pupils were round and equally reactive to light and the confrontational field was grossly full OD, OS. His corrected distance visual acuity was 20/70⁻² OS when measured monocularly and 20/80 when measured binocularly in the monovision condition. There was no improvement with pinhole measurement. At near, corrected acuity was 20/50⁻ when measured binocularly. Compared with the initial visit, the patient's head posture was significantly improved and he was looking through the glasses with no difficulties. The pattern of abnormal eye movements and V-pattern alternating esotropia were observed to be consistent from the initial visit. The patient was counselled on his good adaptation to the monovision and yoked prism prescription. He was recommended to return to clinic in 6 months for dilated fundus examination or sooner as needed. The optical management detail and its outcome are summarized in Table 2.

DISCUSSION

Abnormal ocular manifestations, including ocular misalignment and involuntary eye movements, commonly present in various neurodegenerative diseases. As many are acquired conditions, the patients often suffer from symptoms such as reduced vision,

TABLE 2 Summary of optical management and the outcome.

Habitual spectacle prescription	Final spectacle prescription
OD: -0.50 D 6Δ BO; 1.5Δ BU OS: -0.25 D 13Δ BO, 1Δ BD	OD: +3.00 D 10Δ BD (distance correction) OS: -0.25 D 10Δ BD (near correction) Additional features: photochromic lenses, anti-reflective coating
Initial symptoms	Outcome
- Blurry vision at all distances	- Better visual clarity at both distance and near
- Constant horizontal diplopia	- No distress from diplopia (due to suspension)
- Constant oscillopsia	- Reduced oscillopsia
- Abnormal head posture (chin depression)	- Improved abnormal head posture
- Light sensitivity	- Less light sensitivity

diplopia, and/or oscillopsia, which can greatly impact their quality of life. For these patients, managing the underlying condition should be the primary management goal, and it is the most effective way of controlling the ocular manifestations.^{5,6} However, this may be challenging for many cases, especially when the condition is progressive in nature. For these unfortunate cases in particular, it is important to help the patients reestablish or improve their daily living functions, including vision. The eye care professionals have the opportunity to help with these patients' visual function using any combination of the available management strategies.

Management of Diplopia

Patients with acquired strabismus are more likely to experience diplopia compared with patients who had infantile strabismus. Some patients with longstanding acquired strabismus may establish an adaptation strategy, such as suspension where a diplopic image is ignored.^{7,8} However, in our patient, it may have been more difficult to learn how to ignore the diplopic image due to the constant oscillopsia and movement of the image across the retina. In fact, an oscillation technique known as "macular massage", similar to the movement experienced in nystagmus, is used in orthoptic techniques to break suppression in strabismus.⁹ This same reason presumably hinders his ability to establish stable sensory fusion, even when the ocular alignment is achieved with neutralizing prism. When managing diplopia, the desired goal usually is to restore binocularity and attain fusion. Such may be achieved by strabismus surgery, prism, vision therapy, and/or botulinum toxin injection to EOMs.^{7,8,10} However, each of these options depend on the magnitude and state of comitancy of the deviation, as well as the patient's fusion ability.^{7,8,10} When it is not possible to reestablish functional binocularity with the listed methods, the management goal should be adjusted to obtain maximum patient comfort and visual efficiency by eliminating the diplopic image by making the patients monocular.⁸ Occlusion of an eye is a valid option; however, it may compromise peripheral vision and be cosmetically unacceptable to many patients.^{8,10} Monovision correction is another management option for presbyopic patients that can serve the purpose of eliminating the competition from diplopic images.^{7,8,10} When each eye is corrected for different distances by spectacles or contact lens, the patients can focus on an appropriate image of the two and learn to ignore the other, depending on the specific working distance. Many patients eventually develop this adaptation strategy called suspension and become asymptomatic of diplopia.⁷ It also allows one to retain the peripheral

vision, monocular acuity, and cosmesis, which is a superior option to occlusion.¹⁰ Furthermore, this allows the patient the full lens for viewing at near or distance respectively; a significant advantage for many patients who need a wide field of view. A limitation of this management option is that the best-corrected acuity must be similar in each eye that the degree of visual function is also comparable at each distance. There have been a number of successful case reports, and a small study ($n=20$) that showed the benefit of monovision correction in patients with diplopia.^{7,10}

Management of Oscillopsia

Oscillopsia caused by acquired involuntary eye movements, including nystagmus and saccadic dyskinesia, is another disturbing symptom that the patient with neurodegenerative conditions may experience. The type of abnormal eye movements and resultant oscillopsia may vary depending on the affected location of the lesion.¹¹ When the retinal image slip is greater than 5 degrees per second, it may also cause a reduction in visual acuity, as the target of interest is not fixed on fovea for a sufficient amount of time.^{12–15}

There are various management options suggested for controlling the involuntary eye movements, nystagmus in particular. Regardless of the method, the primary management goal for oscillopsia is to minimise its amplitude and frequency without affecting the normal eye movement function.^{12,13} In this sense, some management strategies that affect the normal range of eye movement, such as botulinum toxin injection, may not be the best option.^{12,14} Surgical techniques, such as four-muscle tenotomy, are also available to improve the control of nystagmus. However, they are known to have a limited benefit in acquired nystagmus compared with the congenital case.^{6,11,16} In addition, there has been suggestions of auditory or visual biofeedback training to help controlling nystagmus.^{17–19} The idea is to help the patients to realise when they have nystagmus and get them to make a conscious effort to control it.^{17,19} However, the benefit of this strategy is questionable for the acquired conditions, as these patients are often already aware of nystagmus due to oscillopsia. Contact lens wear is another known treatment of nystagmus with two possible supporting mechanisms.²⁰ First is that the sensory feedback from the edge of contact lens against the lids causing the dampening of nystagmus.²⁰ The second mechanism is that the better quality of image achieved by contact lens improves foveation, eliminates an induced prism effect, and further decreases oscillation.¹⁹ Unfortunately, most of the reported cases and studies are patients with congenital nystagmus and the benefit for acquired nystagmus is unclear. By far, the

most common management intervention for acquired nystagmus and saccadic dyskinesia is the use of medications, such as baclofen, gabapentin, memantine, and clonazepam.⁵ The effective medical management varies, depending on the type of involuntary eye movement. However, the pharmacologic agent is determined by trial-and-error in many cases and definite effectiveness of each drug is uncertain.^{5,13} Thus, more controlled studies are warranted to confirm their effectiveness.^{5,13}

Besides the use of contact lens, yoked prism is another viable optical management option.^{6,19,21} The yoked prism is prescribed so that the images are shifted towards the patient's null point. For this case, BD yoked prism was prescribed to shift the images towards his up-gaze where the nystagmus was dampened and reduced the intensity of his oscillopsia. Decreasing the frequency of nystagmus also allows better foveation and further improves the clarity of vision.^{6,19} In addition, the abnormal head posture can be improved by eliminating the patients' need to establish their null point themselves.¹⁹ There has been several successful case reports using this management strategy; however, due to the inherent subjectivity of the patient's perception, the magnitude must be determined by trial-and-error.

CONCLUSION

This case report demonstrates successful management of a case of concurrent diplopia and oscillopsia secondary to a neurodegenerative disease using optical management strategies, in addition to the previously dispensed medication. Such cases can be particularly challenging due to the complex underlying cause and symptoms that are often inconsistent. Although it is generally not possible to "cure" these visual signs and symptoms, judicious use of the optical management options described in this paper, applicable for many conditions, should be considered to maximise the patients' visual comfort and efficiency.

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