Article • Evaluation of Diplopia Complicated by Advanced Visual Field Loss: A Clinical Application of Luster Testing

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ABSTRACT

Background: Visual field constriction and binocularity may interact in such a way that slight ocular misalignments produce diplopia due to a loss of peripheral fusional lock. Limited research has characterized diplopia in visual field constriction, and existing case reports do not evaluate patients' fusion. In this paper, we discuss the prismatic management of a patient with significant visual field loss secondary to retinitis pigmentosa. We present a framework to allow simple and efficient clinical evaluation of fusion in this population.

Case Report: A 74-year-old male presented with an ocular history of advanced retinitis pigmentosa and a visual complaint of diplopia that manifested as difficulty focusing with changes in gaze. His habitual bifocals contained a total of 4$^{\Delta}$ base in. A red acetate lens over the right eye and a penlight were used to evaluate binocularity on four separate office visits. Luster was used to assess fusion and enabled a continuous suppression check throughout testing. Multiple evaluations were conducted during each office visit, as the patient's responses were variable. Fresnel prisms were trialed between office visits to ensure that prismatic correction was adequate. Ultimately, a total of 1.5$^{\Delta}$ base in, split between the two eyes, was needed to achieve luster. The patient noted significant improvement in symptoms with the updated prismatic correction and achieved luster with his new bifocals.

Conclusions: Luster testing can be an effective way to assess binocularity in patients with significant visual field constriction and is an easily accessible evaluation technique for most clinicians. Some patients with visual field constriction can be adequately corrected with prism, although due to the progressive nature of ocular disease, the amount of prism needed is expected to change over time. Further research on the prevalence and underlying mechanism of diplopia in patients with visual field constriction is needed.

Keywords: diplopia, peripheral fusional lock, prism, retinitis pigmentosa, visual field constriction

Introduction

Retinitis pigmentosa (RP) is the most prevalent hereditary retinal dystrophy and a relatively common cause of peripheral visual field loss.[1] Usually diagnosed in a patient's third decade of life, RP presents with a characteristic pattern of slow, progressive visual field loss that culminates as a central island of residual vision.[1,2] It is widely understood that the peripheral visual field plays an important role in fusional lock, which allows patients to maintain single binocular vision despite small amounts of retinal image slip.[3] However, the interaction between visual field constriction and binocularity is not well characterized. Literature on the management of diplopia in visual field constriction is limited to case series that do not assess fusion.[4,5] This case expands on previous work, discussing fusional outcomes for the prismatic management of a patient with advanced RP-associated visual field loss. We present a clinical framework for the efficient, effective assessment and management of diplopia in the setting of severe visual field loss.

Case Report

A 74-year-old male with advanced retinitis pigmentosa presented with a complaint of difficulty focusing when making head or eye movements to look in different positions of gaze. Further questioning revealed that this was due to variable diplopia when changing fixation. The diplopia was most often horizontal and most frequently noticed when shifting from distance to near, although it was also present when shifting from near to distance. The patient had been using 2$^{\Delta}$ base in prism OU in his habitual bifocal, prescribed a year and a half prior. At his most recent examination 2 months prior, prismatic prescription was increased to 4$^{\Delta}$ base in OU, which worsened symptoms.

Best-corrected Snellen distance visual acuities were 20/80 (6/24) OD and 20/50 (6/15) OS. Near acuities were 0.2/1M (Snellen equivalent 20/100) OU, 0.18/1.25M (Snellen equivalent 20/139) OD, and 0.18/0.8M (Snellen equivalent 20/89) OS. Eye movements were smooth, accurate, full, and extensive. Mars contrast sensitivity was severely reduced, at 0.96 logCS OD, OS, OU. Visual fields, as assessed using Goldmann kinetic perimetry with a III4e stimulus, were profoundly constricted, with central islands in each eye subtending 7° horizontal by 5° vertical OD and 13° horizontal by 6° vertical OS (Figure 1). There were no residual peripheral islands. While the binocular visual field, assessed as the sum of each eye's monocular visual field, subtended just less than 20°, the overlapping visual field area extended 7° horizontal by 5° vertical based on its widest diameters (Figure 1).

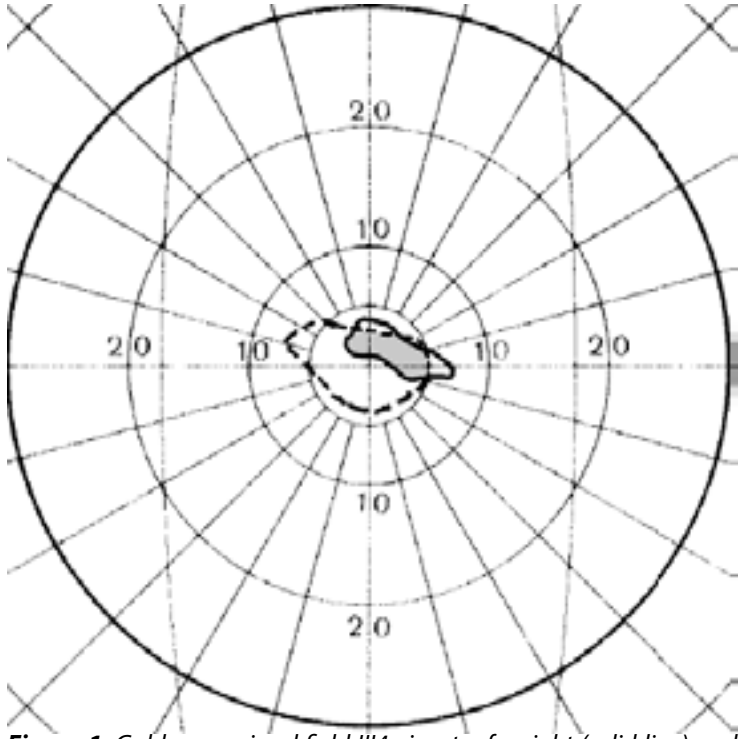

Figure 1. *Goldmann visual field III4e isopter for right (solid line) and left (dashed line) eyes superimposed over one another. Shaded grey area indicates overlapping visual field, or the visual field area where corresponding retinal points exist between the two eyes. This area has an irregular perimeter and extends 7° horizontal by 5° vertical at its widest diameters.*

The binocular vision evaluation was performed during four separate visits to ensure repeatability, and important findings from this (e.g., cover test, luster findings, prism measurement) are documented in Table 1. At each visit, testing was done with the patient wearing his habitual bifocals and holding a red acetate lens over his right eye while looking at a penlight held approximately 50 cm away. In this way, flat fusion in the form of luster and suppression could be evaluated simultaneously and continuously. At each visit, luster was evaluated first to prevent disruption of fusion due to prolonged occlusion during clinical examination. Rotary prism was used in this evaluation, and prism

Table 1. Luster and Cover Test Results at Near

Visit	Current prism	Ocular alignment (Cover test)	Sensory response (Luster test)	Rotary prism measurement to obtain luster
Initial examination (without correction)	None	Variable phoria ranging from a flick eso to flick exo	Vertical diplopia	
Initial examination	4$^{\Delta}$ base in ground in	Small right hyperphoria, moderate exotropia	Vertical diplopia	3$^{\Delta}$ base down OS
2 week follow-up	4$^{\Delta}$ base in ground in	Moderate exotropia	Uncrossed diplopia	2.5$^{\Delta}$ base out
4 week follow-up	4$^{\Delta}$ base in ground in 2$^{\Delta}$ base out Fresnel	Moderate exotropia	Uncrossed diplopia	0.5$^{\Delta}$ base out
3 month follow-up	1.5$^{\Delta}$ base in ground in	Variable phoria ranging from ortho to small exo	Luster	No additional prism

All measures were taken with correction unless otherwise specified

Table 2. Prism Bar Ranges at Near

	Break	Recovery
Without correction	2	0
	2	-2
	6	0
	2	-1
With correction	2	-2
	2	0
	2	0
	1	0

was incrementally added to eliminate diplopia and to allow fusion (Table 1). The cover test was also evaluated at each visit. Additionally, free-space vergence ranges and vergence facility were evaluated at the final visit when the patient had received his appropriate prismatic correction (Table 2). Throughout testing, a penlight was used as a fixation target, ensuring that the patient could see it despite reduced vision, while also allowing continuous luster and suppression testing.

During luster testing, subjective responses were initially variable. Therefore, testing was repeated multiple times within the same visit. Subjective responses provided our primary endpoint (i.e., luster, suppression, or diplopia), but at times the description of diplopia was unusual. Often, the patient described diplopia not as seeing two distinct lights, but as a single light of one colour that was "bleeding off" in the other colour, indicating that each eye's image was approximately aligned but not entirely fused. Often, if the patient blinked a few times, allowing himself to re-fixate, his responses would change significantly and become more consistent following the blink. Prior to prism evaluation, we ensured that the patient was aggressively treating any dry eye, and throughout luster testing, we monitored for monocular diplopia by inquiring whether he saw multiple lights of the same colour (e.g., two red lights would indicate monocular diplopia in the right eye). Consequently, any improvement in diplopia with blinking was unlikely to be related to concurrent ocular surface disease.

Prism trials were decided based only on results that could be repeated within the same visit. We noted that fine adjustments were needed to allow fusion, and large changes resulted in a suppression response.

During initial testing, the patient reported vertical diplopia, which was inconsistent with his chief complaint of horizontal diplopia. Vertical prism was used, and the patient responded most positively to 3^{Δ} base-down prism in the left eye. The patient thought that he might appreciate a slight improvement in focusing with this amount of prism, so a Fresnel prism was adhered to his left lens to trial for two weeks.

At his two-week follow-up (visit 2), the patient had removed the Fresnel prism, noting less diplopia without it. Luster testing at this visit revealed uncrossed diplopia, and fusion was achieved with 2.5^{Δ} base-out prism. A 2^{Δ} base-out Fresnel prism was applied to the patient's right eye, as 2.5^{Δ} base-out Fresnel prism was unavailable. With this in place, the patient noted an initial improvement in his ability to focus and appreciated improved comfort. The patient was encouraged to trial this new prism at home over the next two weeks to determine whether it would promote fusion.

At the four-week follow-up (visit 3), the patient noted that the improved focusing ability provided by the Fresnel prism persisted throughout the two weeks, and he was content with the results. He did, however, note that the Fresnel degraded image quality and induced some blur. Consequently, he was interested in learning whether the prismatic correction could be incorporated in a different way. At this visit, luster testing revealed a small amount of uncrossed diplopia, which was resolved with 0.5^{Δ} base out over the Fresnel prism, amounting to 2.5^{Δ} base out over his habitual spectacles. As measures were repeatable over two visits and the patient noted a marked improvement in symptoms with the Fresnel prism, 2.5^{Δ} base-out prism was incorporated into the patient's existing prismatic correction to be ground into a new pair of bifocals. This amounted to a total of 1.5^{Δ} base in (i.e., 4^{Δ} base in from the habitual bifocals combined with 2.5^{Δ} base out from prism evaluation), ordered as 0.75^{Δ} base in per eye (i.e., the prism was split equally between the two eyes). After the glasses were dispensed, the patient was encouraged to return for follow-up in 8 weeks to ensure proper adaptation to the new spectacles. He was reminded that should symptoms recur or worsen, he could return earlier.

Eight weeks after the patient received his glasses, he returned for follow-up (visit 4), noting resolution of both the focusing difficulty and the diplopia with his new glasses. He was content with the clarity of his vision through these bifocals. The patient achieved luster wearing his new bifocals without needing any additional prism. He reported clear, comfortable, single vision with his glasses and denied any residual difficulty when changing focus.

At this visit, near cover test responses (Table 1) were variable, but repeat measures indicated an

exophoria. All near vergence ranges (Table 2), which were tested with a penlight and red acetate lens over the right eye, were constricted, both with and without appropriate prismatic correction. Base-out ranges appeared to improve with prism, while base-in ranges seemed to worsen. However, ranges were so narrow that normal fixation changes and testing error may have accounted for the differences.

Standard vergence prism flippers (12$^{\Delta}$ base out/4$^{\Delta}$ base in) were also attempted at near using a penlight as a target with a red acetate lens over the right eye in order to evaluate vergence facility while the patient was wearing his habitual correction. The patient demonstrated more difficulty fusing base-out prism than base-in prism initially, and later in the test, he reported that the image was shifting left or right with each flip. This response indicates either suppression or that the image had been shifted onto non-seeing retina.

Discussion

The relationship between visual field loss and the ability to maintain clear and comfortable single binocular vision is not fully understood. Little research has been done to elucidate interactions between the two, although the hemi-field slide phenomenon has been thoroughly documented. In this entity, patients with bi-temporal hemianopia experience splitting or doubling of images with small amounts of ocular misalignment due to a lack of overlapping corresponding retinal points.[6-8] More recently, a case series has demonstrated that this effect can occur in patients with bilateral heteronymous altitudinal defects (i.e., loss of the superior hemi-field in one eye and the inferior hemi-field in the other), presumably by the same mechanism.[7] However, limited work has established the impact of non-hemispheric visual field loss on diplopia.

To our knowledge, only two recent case series discuss the management of diplopia for patients with visual field constriction.[4,5] One series reviews seven cases in which patients experienced visual field constriction and large-angle strabismus.[4] Many of these patients had gaze palsies, and all but one case underwent strabismus surgery. Only two patients included in this case series had visual field constriction: one due to glaucoma and one due to retinitis pigmentosa with a likely decompensated fourth nerve palsy. All others had neurological visual field loss. Both patients with visual field constriction noted alleviation of diplopia, although not complete resolution, following strabismus surgery.[4] However, no evidence of post-operative fusion was provided, so it remains unclear whether patients truly regained fusion or whether alignment was adjusted such that the non-foveated image less frequently fell on seeing retina in one eye. The second case series[5] discussed three patients with advanced glaucomatous visual field loss who had variable small-to-moderate sized tropias with symptoms of diplopia. All patients ultimately failed prismatic correction and preferred to ignore the double image, as the benefit of a larger binocular visual field outweighed that of single vision through monocular occlusion. The authors noted that the variable diplopia in advanced glaucomatous field loss seems to share characteristics with hemi-field slide, and perhaps these glaucoma patients experienced diplopia due to insufficient binocular overlap in the residual visual field.[5]

Previous case report findings[4,5] seem consistent with our understanding of normal fusion, in which the periphery enables robust maintenance of fusion through fusional lock.[3] Patients who lack adequate peripheral vision, therefore, depend solely on their central vision, which is limited in its capacity to correct for retinal image disparity continuously in order to promote binocular single vision.[3] Consequently, patients with peripheral visual field loss may be more likely to experience diplopia due to fatiguing of their over-taxed central fusional system. However, this has not been thoroughly investigated. One study examining the impact of chiasmal lesions on stereopsis theorized that a 5 to 6° central visual field is needed for patients to achieve stereopsis.[9] However, this analysis was conducted with a sub-group of the study sample, consisting of only six control-group RP patients, thus weakening results. As such, the amount of peripheral vision that is necessary to allow normal continuous fusion must be further evaluated.

Similar to the glaucoma patients studied by Khanna and Holmes,[5] our patient reported variable diplopia, and measurements fluctuated throughout testing. Our patient described his diplopia as a "focusing difficulty," reflecting the variability and resulting challenge that may be experienced in attempting to describe symptoms. This variability may not only make it increasingly difficult for patients to recognize symptoms, but also for clinicians to elicit them. Patients with significant visual field constriction have an underlying ocular disease, and some managing providers may not routinely assess binocularity and diplopia. While the prevalence of diplopia in patients

with visual field constriction has not been investigated, patient and provider factors likely contribute to underdiagnosis.

Methods employed to evaluate diplopia in this case differ from the usual techniques. For patients with vision loss, additional care must be taken to ensure that the target being used can be seen easily by each eye, as reduced contrast sensitivity and visual acuity may further disrupt fusion. Some techniques that are commonly used to assess binocularity (e.g., associated phoria, stereoacuity) could not be evaluated in this patient due to his reduced near vision. Even a cover test can produce spurious results for patients with vision loss. This patient had a near acuity in the worse-seeing eye of 20/139. A letter of this size would have a minimum angle of resolution of 6.95°. For this patient, an appropriately sized cover test target would subtend his entire visual field, making it impossible to ensure steady central fixation during testing. Consequently, cover test responses did not match sensory responses and became invalid. For this elderly patient, the usual accommodative target was replaced by a penlight. This enabled us to ensure target visibility and provided a continuous suppression check, using a red acetate lens over one eye. Through this method, as long as the patient perceived luster or two differently coloured lights, we knew that the percept was visible in each eye and our results were valid. Luster can also be evaluated in multiple positions of gaze to assess comitancy, although this was deferred in this case.

While luster evaluation was useful in this case, it did not provide complete information about binocularity and may not be appropriate in isolation for patients with stable central fixation, significant glare sensitivity, or who are able to accommodate (e.g., children). However, in cases where binocularity cannot otherwise be assessed due to reduced vision, it is an efficient tool that can allow most clinicians to diagnose and treat diplopia confidently.

During testing, a suppression response often indicated that the target had been shifted onto non-seeing retina rather than true sensory suppression. This is best demonstrated through the patient's responses to prism. This patient's horizontal visual field diameter subtended 7° (12.28$^{\Delta}$) in the right eye (Figure 1). Assuming the patient's visual field was centered over fixation and that fixation remained stable throughout testing, introducing prism of greater than 6.14$^{\Delta}$ base in or out over the right eye would shift the right eye's image onto non-seeing retina. If the same logic is applied to the left eye, which had a horizontal visual field diameter subtending 13° (23$^{\Delta}$), prism greater than 11.5$^{\Delta}$ base in or out would induce a suppression response (Figure 1). In either case, the suppression response would not indicate true sensory suppression; rather, it would indicate that the image had been shifted onto non-seeing retina. Introducing prism over the eye with a larger visual field enables larger prism powers to be assessed before field constriction precludes further evaluation. Fusional prism used in this case was less than the maximum possible based on the patient's visual field, and the patient's consistent report of luster indicates that fusion was in fact achieved. However, when evaluating vergence facility, a suppression response was inevitable because of the large amounts of prism that were used during testing. Valid responses might have been attainable if a smaller amount of prism was used (e.g., 2$^{\Delta}$ base out/1$^{\Delta}$ base in). In this case, small amounts of prism were needed to achieve fusion. However, for patients requiring larger amounts of prism, clinicians may need to consider asymmetrically splitting prism between the two eyes to ensure that the shifted image continues to fall on seeing retina in each eye.

RP-associated visual field loss is progressive;[1] therefore, the amount of prism needed to promote fusion is expected to change over time. If peripheral vision drives fusion, further loss of the periphery may result in decreased stability or a larger-angle tropia, which would require higher amounts of prism. The prism prescribed at this visit likely will no longer be appropriate in future evaluations. This prediction is supported by the patient's history with prismatic correction, as he had comfortably worn base-in prism for a year and a half prior to this evaluation.

One limitation of this case report is that a binocular Goldmann visual field was not performed, and we inferred it to be equivalent to the visual fields of the right and left eyes superimposed over one another. As the patient has a tropia, this simplification introduces inaccuracy. A tropia would cause one eye's visual field to be shifted relative to central fixation only when tested binocularly. For example, in the case of a left exotropia, the left eye's visual field would be shifted left, resulting in an overall larger binocular visual field, but a smaller area of binocular overlap. Since corresponding points within the area of binocular overlap would have different images, visual confusion would be induced. Literature has documented the effectiveness of using peripheral visual confusion induced through prism to produce visual field expansion for patients with peripheral visual field loss.[10,11] However, central

visual confusion is poorly tolerated, making prismatic management to eliminate diplopia an important goal.

Conclusions

Diplopia in patients with visual field loss is common and likely underdiagnosed; further research is needed to enable a full understanding of its prevalence and mechanism. For patients with RP and other conditions causing visual field constriction, luster testing is a simple strategy that can efficiently provide information about binocularity and is accessible to most eye care providers. Prismatic correction can successfully enable some patients with peripheral field loss to fuse, although prism will need to be adjusted over time.

Acknowledgements

We would like to thank Drs. Kenneth Ciuffreda and M. H. Esther Han for their valuable feedback on this project.

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Gobeille M, Patel S, Meltzer B, Fischer M. Evaluation of diplopia complicated by advanced visual field loss: A clinical application of luster testing. Optom Vis Perf 2020;8(3):122-7.