

Article ▶ Corectopia: Facilitating a Preferred Retinal Site/Preferred Retinal Locus: A Theory on Enhancing the Potential for Reorganization of Visual Processing in Those with Macular Degeneration/Absolute Central Scotoma

Lawrence A. Routt, Sr., OD, Kosciusko, Mississippi

ABSTRACT

Background: Having a preferred retinal locus or loci refers to the utilization of one or more eccentric retinal fixation points, most often occurring with loss of foveal function. This article proposes the theory that surgical pupil displacement could facilitate development of an optimally functioning preferred retinal locus in those with macular degeneration/absolute central scotoma while enhancing the potential for reorganization of visual processing.

Case Report: A seventy-five year old aphakic male presented complaining of blurred vision with his habitual aspheric spectacles. He reported having had extracapsular cataract extraction for congenital cataracts OD, then OS, at seventeen years of age. He had mild corectopia OD, a severely updrawn peaked pupil OS, and best visual acuity (BVA) limited to 20/40-1 OD and 20/40 OS at distance. Scanning laser ophthalmoscopy indicated the use of a preferred retinal locus OU. With chronic dilation of the patient's updrawn pupil OS, the visual axis was enabled, the dimensions of the visual field OS were significantly increased, and BVA improved to near normal OS.

Conclusion: A patient with macular degeneration/absolute central scotoma who has a centered pupil and normal peripheral retina in the absence of other significant ocular or neurologic pathology could, in theory, benefit from surgically induced corectopia. This would enable direct light stimulation of a preferred retinal site, an area of viable retina determined to be the most appropriate for establishing a single, stable, preferred retinal locus when there has been loss of foveal function. This could enhance the potential for reorganization of visual processing.

Keywords: absolute central scotoma, alternative visual axis, age-related macular degeneration, corectopia, preferred retinal site, preferred retinal locus or loci, reorganization of visual processing, Stargardt disease

Introduction

Normal human vision is an on-axis optical system. It comprises four main coaxial elements, including the anterior and posterior corneal and lenticular surfaces and the pupil that enables stimulation of the retina, the light-sensitive tissue layer. A well-centered pupil acts as an optical aperture that facilitates the following: an optic axis that contains the center of curvatures of the optical surfaces of the eye and a visual axis for maximum stimulation of the fovea,¹ best visual acuity, stimulation of both central and peripheral retina for an optimal contiguous visual field (about 160 degrees horizontally and 120 degrees vertically), and optimal image quality centrally and peripherally.²

Corectopia, a displaced pupil (not in the center of the iris),^{2,3} can reduce visual acuity, visual field, light sensitivity, spatial resolution, and contrast sensitivity. A displaced pupil can also result in color distortions, unpredictable changes in refractive error, wavelength-specific distortions of visual direction, and wavelength-specific errors in the judgement of apparent depth and size.²

Because the potential complications of corectopia are so numerous and undesirable, there has been relatively little research concerning the off-axis human optical system. There

has been even less interest in pupil displacement except as a complication of ocular surgery due to vitreous or iris entrapment, intraocular lens placement, etc.⁴ Nevertheless, it is possible that surgical pupil displacement could benefit those with macular degeneration/absolute central scotoma.

Two hereditary diseases are responsible for most macular degeneration in children and adults. Stargardt disease begins in childhood and causes slowly progressive visual loss for 1 in 10,000 children, while age-related macular degeneration (ARMD) is the leading cause of blindness in the elderly, affecting nearly 15% of people over age 75.^{5,6}

In the majority of those with Stargardt disease, vision will decline to 20/200 or worse, and patients may become visually disabled by their 20s. Since high-resolution central vision is necessary for reading, driving, using computers, monitoring facial expressions, and other tasks, function is severely limited. Although there is presently no medical/surgical treatment for Stargardt disease,⁷ functional care, including low vision lenses and devices, can be beneficial.⁶⁻⁸

Early ARMD may show no symptoms or loss of vision. For intermediate and late ARMD, researchers in the Age-Related Eye Disease Studies (AREDS and AREDS2) at the National Eye Institute found that daily intake of certain high-dose

vitamins and minerals can slow progression.⁶ Although there is no cure for advanced neovascular ARMD, substantial progress has been made in the development of anti-VEGF (vascular endothelial growth factor) agents that can slow progression and may provide improvement. These anti-VEGF agents have essentially superseded earlier treatments such as photodynamic therapy using verteporfin, laser photocoagulation, and injection of steroids such as triamcinolone. Despite highly effective treatment with anti-VEGF agents, some patients still have significant vision loss, and others will have lost vision prior to receiving effective treatment. For those with dry ARMD, functional care that includes low vision lenses and devices is the only available treatment.⁶⁻⁸ It is possible that surgically induced corectopia could enhance the potential benefits of such rehabilitative care.

Case Report

A seventy-five year old aphakic, white male presented complaining of blurred vision with his habitual aspheric spectacles that were in poor condition. The subject reported having to hold reading material closer than desired. He had

a history of extracapsular cataract extraction OD at seventeen years of age and ECCE OS several months later (dates of surgery unknown).

Unaided visual acuity was 20/4667 OD, OS. The habitual prescription was:

Habitual spectacle Rx:

OD = +10.00-0.50x006 Add: +3.75

DVA = 20/50; NVA = 20/50

OS = +10.00-0.25x171 Add: +3.75

DVA = 20/50; NVA = 20/50

The Maddox Rod Test indicated 3^Δ BD and 1^ΔBO OS. There was no diplopia, and the patient had reliable alignment. Stereopsis was absent according to stereo tests for House Fly, Circles, and Animals. According to the Miles test, the patient preferred the left eye. There was bilateral vitreous to cornea with mild corectopia OD and severely updrawn, peaked pupil OS, apparently occurring at the time of extracapsular cataract extraction (Figures 1-3). Although there are congenital, non-surgical causes of corectopia, such as idiopathic tractional corectopia, Axenfeld-Rieger syndrome, and ectopia lentis et pupillae,⁹⁻¹¹ they are not pertinent to this case report.

There was significant opacification of the capsular vitreous face OU, but no other ocular pathology was noted. Blood pressure was 156/92 mmHg. The patient, who denied a history of hypertension, was referred to his primary care physician. Refraction was postponed. The patient was referred

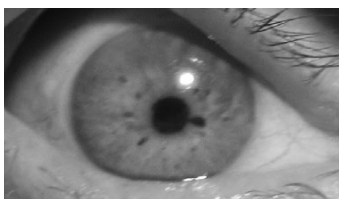


Figure 1. Pupil appearance pre-dilation OD

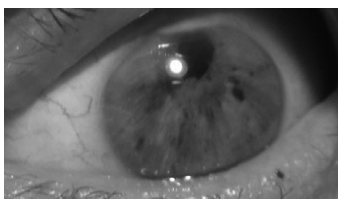


Figure 2. Pupil appearance pre-dilation OS



Figure 3. Pupil appearance pre-dilation OU



Figure 4. Pupil appearance post-dilation OD

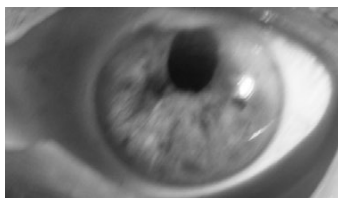


Figure 5. Pupil appearance post-dilation OS



Figure 6. Pupil appearance post-dilation OU

Table 1. Pupil Dimensions Pre- and Post-Dilation OU

PRE-DILATED PUPIL DIMENSIONS OD	POST-DILATED (atropine 1% bid) PUPIL DIMENSIONS OD
Vertical = 3.0mm Horizontal = 3.0mm	Vertical = 4.5mm Horizontal = 4.0mm
PRE-DILATED UPDRAWN PUPIL DIMENSIONS OS	POST-DILATED (atropine 1% bid) UPDRAWN PUPIL DIMENSIONS OS
Vertical = 3.0mm (pupil extends 1mm below the LUL margin, primary gaze) Horizontal = 2.0mm	Vertical = 4.5mm (pupil extends 2mm below the LUL margin, primary gaze) Horizontal = 3.5mm

to an ophthalmologist, who performed YAG laser to the capsular vitreous face opacification OS and OD, with one month separation between procedures. Six months later, the patient's systemic hypertension was well controlled.

Unaided VAs = 20/1167 OD, 20/500 OS. The patient was refracted and new aspheric spectacles were dispensed:

OD= +10.00-0.50x005 Add: +3.50;
DVA 20/40-1; NVA= 20/40

OS= +10.00-0.25x170 Add: +3.50;
DVA 20/40; NVA= 20/40

Three months later, the patient reported much improved vision with routine dilation OU. Thus, further testing was done to compare visual acuities, visual fields, and pupil dimensions pre- and post-dilation with atropine 1% OU bid (Figures 4-6). Scanning laser ophthalmoscopy was used to test for eccentric fixation pre- and post-dilation with atropine 1% OU bid. Unaided VAs remained unchanged despite dilation OU.

The subject's corectopia OD included mild displacement of the pupil 1.5 mm inferior nasal (Figure 1). A pupil is normally situated about 0.5 mm inferior nasal from center of iris. 12 Dilation caused no significant change in visual field OD (Figures 7, 8). According to scanning laser ophthalmoscopy, a preferred retinal locus was established 1.2 degrees temporal, 0.7 degrees superior and 1.4 degrees total distance from the fovea OD. With dilation of the right eye, see Table 1, fixation moved to the fovea. However, the right eye remained amblyopic at 20/40-1 BVA at far (see discussion).

The severely updrawn pupil OS (Figure 2) and the resulting 4.5 mm vertical disparity from the undilated pupil OD to the undilated pupil OS inhibited the visual axes OU. Instead, the patient established a preferred retinal locus in each eye with less than normal BVA, as indicated above. Scanning laser ophthalmoscopy indicated a preferred retinal locus located 1.2 degrees temporal, 0.49 degrees inferior and 1.3 degrees total distance from the fovea of the undilated OS (see Discussion). With dilation of the left eye (atropine 1% bid; Table 1), there was an increase in visible pupil area below the left upper eyelid margin in primary gaze.

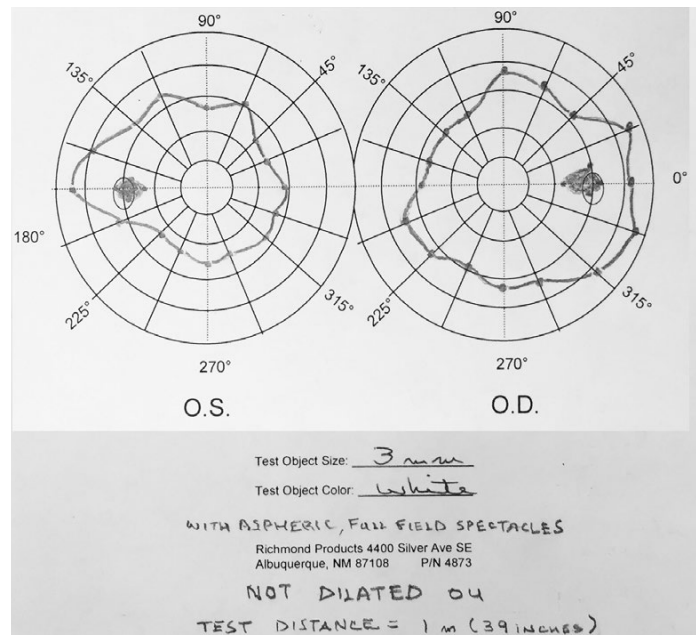


Figure 7. Tangent screen pre-dilation

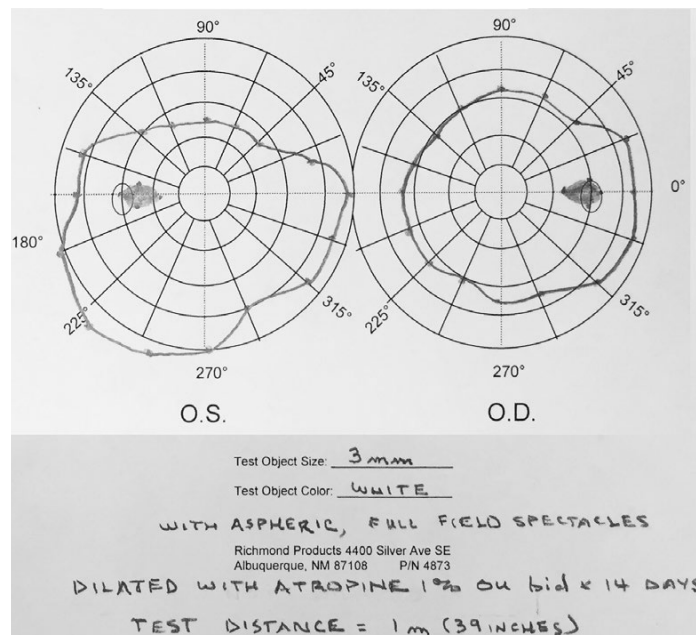


Figure 8. Tangent screen post-dilation

Since the patient was unable to perform reliably on automated visual fields, tangent screen visual fields were performed (Figures 7 and 8). As shown in Figure 8, there was a significant increase in the overall vertical and horizontal dimensions of the visual field OS with dilation.

Dilation of the severely updrawn pupil OS enabled a visual axis with fixation at the fovea, as indicated by scanning laser ophthalmoscopy. The BVA OS improved from 20/40 distance and near (undilated) to 20/20-1 at distance and 20/25 at near (dilated with atropine 1% OS bid x 2 weeks).

The patient's BVA OS was maintained with chronic dilation. The spectacle Rx OS was unchanged, and best visual acuity was 20/20-2 in the distance and 20/25 at near (dilated with atropine 1% OS bid x 14 months).

Discussion

Because of the subject's 4.5 mm vertical disparity from the center of the inferiorly displaced pupil OD to the center of the severely updrawn pupil OS when undilated, there was significant vertical disparity from the optic axis OD to the optic axis OS, and from the visual axis OD to the visual axis OS. Some adaptation of the undilated patient involved establishment of a preferred retinal locus located 0.49 degrees inferior to the fovea of the left eye that minimized his loss in the superior visual field OS. The patient had reliable alignment, and there was no diplopia. Prism was of no benefit.

The patient (undilated) unconsciously established a preferred retinal locus OU just as a patient with macular degeneration/absolute central scotoma might institute a preferred retinal locus. In both cases, eccentric viewing is an off-axis adaptation to a damaged on-axis optical system that typically involves defective visual fields and reduced visual acuities.¹⁻³

Dilation with atropine 1% OD bid x 2 weeks (Table 1) enabled fixation at the fovea OD. However, the right eye remained amblyopic at 20/40-1 BVA at distance. Because congenital cataract extraction OD, then OS (dates of surgery unknown) was not performed until the patient was seventeen years old, he may have developed a preference for the possibly better seeing left eye as a young child. This could explain the amblyopia OD in the absence of retinal pathology OU. Dilation with atropine 1% OS bid x 2 weeks (Table 2) reduced vertical disparity of the axes from OD to OS and enabled fixation at the fovea of the left eye, with improvement in BVA at far from 20/40 to 20/20-1 OS. The BVA of the left eye was maintained at 20/20-2 with chronic dilation (atropine 1% OS bid x 14 months).

Since visual acuities decline as fixation distance from the cone-dense fovea increases,^{13,14} progression in size of an absolute central scotoma can be debilitating. As a result, researchers have developed vision training programs that enhance the use of one or more preferred retinal loci.^{15,16} Unfortunately, much of this training and/or unconscious adaptation is limited by a centered optical aperture (pupil), which determines the area of the retina stimulated by light and places a macular scotoma in the center of the patient's visual field. A centered pupil also limits the area of healthy, functional peripheral retina that needs to be stimulated for development of a preferred retinal locus to a ring of tissue disrupted by the central lesion. This enables visual cues from viable retina surrounding the central lesion to direct the scotoma to the center of the patient's visual field in all positions of gaze. Such disruption of the central visual field may require the patient intermittently to move peripheral retinal fixation from one preferred retinal locus to another across the scotoma as in pursuits, saccadic movements,¹⁷ and other task-dependent efforts.

Whitaker et al. showed that multiple preferred retinal loci are more likely to occur as the central scotoma increases

Table 2. Tangent Screen Results OS

(15° Tangent Screen, Test object size = 3mm, Test object color = white Test distance = 1 meter, with aspheric spectacles, chin rest, lights dimmed)		
	PRE-Dilation	POST-Dilation
Total distance from superior to inferior limits of visual field	27°	38°
Distance from fovea to superior limit of visual field	14°	13°
Distance from fovea to inferior limit of visual field	13°	25°
Total distance from temporal to nasal limits of visual field	37°	46°
Distance from fovea to nasal limit of visual field	14°	24°
Distance from fovea to temporal limit of visual field	23°	22°

in size.¹⁸ This adaptation is contrary to the development of a single, optimal preferred retinal locus, which better simulates a normal fovea in a healthy on-axis human optical system. Furthermore, it is possible that multiple preferred retinal loci inhibit the potential for reorganization of visual processing.

Frolo et al. underscore the importance of facilitating a single, optimal preferred retinal locus in those with central visual field scotoma.¹⁹ These researchers used functional magnetic resonance imaging to show that direct stimulation of the patient's preferred retinal locus with object pictures led to significantly increased activation in striate and extrastriate cortices, as well as higher visual areas (fusiform gyrus, inferotemporal gyrus), in comparison to stimulating a matching peripheral area in the opposite hemifield. The difference was not seen in controls. Furthermore, patients with stable eccentric fixation showed a greater difference between preferred retinal locus and opposite hemifield preferred retinal locus in fusiform and inferotemporal gyrus than patients with unstable eccentric fixation.¹⁹ It is possible that multiple preferred retinal loci that may be necessary to move retinal fixation across a central scotoma are less stable than a single preferred retinal locus.

In contrast to the results just described, not all studies show clear evidence for cortical reorganization as a response to central scotoma.²⁰⁻²³ Yet, functional magnetic resonance imaging, microperimetry, and scanning laser ophthalmoscopy have assisted researchers in providing support for cortical reorganization in humans with central scotoma.^{6,19,24-29} This offers hope to those with macular degeneration/absolute central scotoma who experience loss of vision despite present medical/surgical care.⁸ While low vision lenses, devices, and functional treatment can be effective⁶⁻⁸ surgical pupil displacement superiorly could enhance the potential benefits of such rehabilitative care.^{2,5,15,16,19,24-29}

There are eight objectives/possible benefits of surgically induced corectopia for those with macular degeneration/absolute central scotoma: 1) to simulate an on-axis optical system² in which there is light stimulation of a preferred retinal site, with establishment of an optimally functioning preferred retinal locus; 2) to facilitate direct light stimulation of a single, preferred retinal locus such that its receptors have an opportunity to re-orient towards the center of the displaced pupil for optimal visual sensitivity and resolution;³⁰ 3) to enable the use of an alternative visual axis, the line

from the fixation target through the center of the surgically displaced pupil to a single, stable preferred retinal locus; 4) to stimulate as much healthy, viable superior retina as possible to provide the maximum available, contiguous inferior nasal, inferior central, and inferior temporal visual field needed for locomotion, uninterrupted lateral side-to-side gaze, reading, saccades, tracking, and pursuits;³¹⁻³³ 5) to move the scotoma effectively to the superior limit of the patient's visual field. This could help eliminate lateral disruption of the visual field that may be associated with the development of multiple, unstable preferred retinal loci;^{19,34} 6) to eliminate the need for unusual head turn³⁵ or tilt and unstable visual fixation,⁵ including low-frequency jerk nystagmus¹⁸ that possibly enables utilization of multiple preferred retinal loci; 7) to eliminate conscious or unconscious reinforcement of established multiple preferred retinal loci.⁵ A normal human on-axis optical system has only a single retinal fixation point, the fovea, and a single visual axis per eye. When there is loss of cortical input from macular degeneration, reinforcement of multiple preferred retinal loci may be confusing to visual processing;¹⁹ and 8) to enhance the effectiveness of vision training to establish and to reinforce a single, stable preferred retinal locus in the superior retina (inferior visual field) above the central lesion (below the central scotoma).^{15,16,19,36,37} If any or all of the possible objectives/benefits of surgically induced corectopia are realized, the potential for reorganization of visual processing in those with macular degeneration/absolute central scotoma could be enhanced.^{5,19}

For all of the reasons discussed, it is preferable for a patient with macular degeneration/absolute central scotoma to develop a single, stable preferred retinal locus as close as possible to the superior border of the central lesion but without any lateral obstruction. Although studies show that such a preferred retinal locus can be associated with activation of the visual cortex,^{5,13,14,17-19,30,39,40} there has been no known attempt at surgically induced corectopia to stimulate a preferred retinal site in order to establish an optimal preferred retinal locus, to enable an alternative visual axis, to improve the effectiveness of vision training, or to enhance the potential for reorganization of visual processing.^{19,24-30}

Despite present medical, surgical, and functional care, including low vision lenses and devices, some patients suffer loss of vision to less than 20/200 BVA in the better-seeing eye and are unable to function at an acceptable level as the result of macular degeneration/absolute central scotoma.⁵⁻⁸ For such a patient who has a centered pupil and normal peripheral retina in the absence of other significant ocular or neurological pathology, surgically induced corectopia is worthy of consideration.^{5,13,14,17-19,24-30,39,40}

The pupil should be updrawn superiorly so that the inferior margin of the surgically displaced pupil is at or slightly above the superior margin of the pre-surgical pupil with about 500-750 lux, lumen/m² indoor office illumination. The superior portion of the surgically displaced pupil may

be covered by the upper eyelid in primary gaze, but this is not expected to cause any adverse effects. Although visual acuities decline faster vertically than horizontally as distance from the fovea to the retinal fixation point increases,^{14,38} pupil displacement superiorly is preferable because it would most closely simulate an on-axis optical system and provide maximum available uninterrupted inferior visual field.^{1,2}

A second consideration for surgical pupil displacement is that the horizontal pupil diameter, with about 500-750 lux, lumen/m² indoor office illumination, should remain as nearly the same as possible to the pre-surgical dimension in order to simulate most closely the patient's pre-macular degeneration, on-axis optical system.^{1,2} The post-surgical pupillary distance should remain as close as possible to the pre-surgical measurement to assist in maintaining optimal binocularity when possible. Finally, the post-surgical updrawn pupil should remain responsive to topical pharmaceutical dilation such that light stimulation of the superior retina can be manipulated to extend or to limit the visual field, as preferred subjectively and objectively.

In research on monkeys with bilateral laser-induced foveal lesions, a preferred retinal locus developed within days, but it took several weeks to train the eye movements to place targets on the preferred retinal locus.⁴¹ In humans with less than 20/200 BVA in the better-seeing eye as a result of macular degeneration/absolute central scotoma, the timing for the development of a preferred retinal locus is uncertain.^{5,17,18,24,33,34,39} This has implications for vision training following surgical pupil displacement.

If acceptable to the patient, the unoperated eye should be patched/occluded for all waking hours beginning as soon as possible after superior displacement of the pupil in the operated eye.^{5,17,18,24,33,34,39} Patching/occlusion of the unoperated eye is intended to inhibit interference with the potential for reorganization of visual processing involving the operated eye.⁵

Because optimal stimulation of a preferred retinal site could aid in establishing a single, stable preferred retinal locus, refraction of the operated eye should be performed, when appropriate, following superior displacement of the pupil.^{2,30-33} If significant vision correction is prescribed for the operated eye, the spectacles (unoperated eye patched/occluded) should be recommended for all waking hours regardless of whether or not there is any immediate improvement in vision. If the patient has accommodative insufficiency/presbyopia, a lined bifocal is preferable, as the near power is constant throughout the seg. This could inhibit the establishment of multiple preferred retinal loci that may be associated with a progression of lens powers for near.¹⁷

Post-operative vision training should be continued, as needed, to aid in establishing a single, optimally functioning preferred retinal locus with utilization of an alternative visual axis.^{19,20} If this occurs as expected, the unoperated eye should remain occluded for all waking hours, if comfortably tolerated,

to inhibit interference with the potential for reorganization of visual processing in the operated eye. Even if the latter fails to occur, any or all of the other objectives/benefits of surgical pupil displacement superiorly may be significant for those with less than 20/200 BVA in the better-seeing eye as a result of macular degeneration/absolute central scotoma.^{5,19,31-35,37,39}

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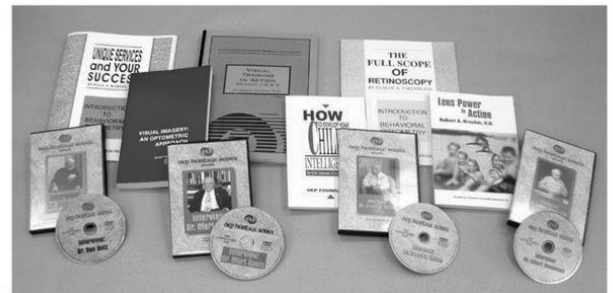
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