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Practice Report

Low Vision Rehabilitation of Retinitis Pigmentosa

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Retinitis pigmentosa is a rod-cone dystrophy, commonly genetic in nature. Approximately 60–80% of those with retinitis pigmentosa inherit it by an autosomal recessive transmission (Brilliant, 1999). There have been some reported cases with no known family history.

The symptoms of retinitis pigmentosa are decreased acuity, photophobia, night blindness, and a progressive constriction of the visual field. Visual acuity can vary from 20/20 to no light perception. Typically, acuity is severely affected only in the later stages, when macular changes and glare from posterior subcapsular cataracts may cause moderate to severe vision loss. A person with retinitis pigmentosa has difficulty making the transition between different levels of lighting because of decreased rod-cone function. The visual field loss usually begins in the midperiphery and works in and out, creating a ring scotoma. Owing to the slow, progressive nature of the field loss, many individuals are unaware of it initially.

These visual symptoms create a challenge to visual function. Decreased acuity can make reading and writing difficult, glare can cause a person with good acuity to be severely impaired, and mobility can be limited. Since the constricted visual field hampers the individual from gathering visual information from his or her surroundings, he or she may be hesitant while traveling, often reverting to tactile methods, which are inefficient. These issues can be addressed through comprehensive low vision rehabilitation. A precise refraction, magnification for near tasks, and an evaluation of glare can maximize acuity. Visual field enhancement, in conjunction with orientation and mobility (O&M) training, can help a person travel more safely and confidently. The following case report illustrates many aspects of comprehensive low vision rehabilitation.

Case report

James, a 54-year-old white man, who had been diagnosed with retinitis pigmentosa 10 years earlier, was referred to the low vision center for a low vision evaluation. He reported no family history of retinitis pigmentosa. He said that his vision had deteriorated in the past few years to the point where it was starting to affect his life, he was no longer driving, and was having difficulty with mobility. He felt unsafe in unfamiliar settings, was constantly bumping into things, and his near tasks had become more challenging. James was light sensitive on bright days, had poor adaptation from light to dark, and had poor night vision.

James's major concern was his employment at an automotive plant, which required desk work and walking the floor in a supervisory capacity; people who came to his desk from behind or the side startled him. James had four years until retirement and wanted to continue to work until then. Most people at work were unaware of his impairment, but he thought that he now needed help to continue his job. His prior ocular history, besides the diagnosis of retinitis pigmentosa, was remarkable only for a spectacle correction for myopic astigmatism and presbyopia. His medical history was unremarkable. He denied taking any medications and had no known medical allergies. He had no prior low vision rehabilitation.

Upon evaluation, his distance visual acuity with his current spectacle correction was $20/63^{-}$ OD and $20/63^{-}$ OS with direct fixation and moderate illumination OU on a modified ETDRS chart. James often lost his place and bounced between lines while reading off the chart. His near acuity with his current bifocal add was $1.25M^{+}$ OU on a Lighthouse Near Acuity Card. His distance acuity through trial-frame refraction was improved to 20/50 OD, 20/50 OS, which James thought was a significant improvement. James's near acuity with a +3.00D add improved to .6M single letters, and James was able to read 1M continuous text without difficulty. Goldmann visual fields, using a III4e stimulus, showed a severe constriction of 10 degrees in the right eye and 7 degrees in the left.

In a tint evaluation, James was shown a variety of Corning CPF lenses, gray lenses of 88% and 60% absorption, amber lenses, and kalichrome lenses outdoors on a bright, sunny day and indoors with moderate illumination. James preferred the Corning CPF527 lenses with side shields for outdoors and clear lenses for overcast days and indoors. The concept of field enhancement with prisms was discussed with him, and displacement was shown to him with a handheld prism. The method of using Fresnel prisms temporal and nasal on each eye with a clear central zone for scanning was explained. James was willing to try anything that might help.

A convex mirror to be used as a field-enhancement spotting tool, which James could use at his desk to help locate people who approached from the sides and rear, was demonstrated to him, and a wide-angle mobility light was recommended for nighttime travel. O&M training was discussed and recommended. James was hesitant at first, but consented when it was explained that the success rate for using prisms was much greater when O&M training was provided.

After the initial visit, the following devices were ordered: (1) clear Rx: OD –3.00 –1.75 X 012, OS –

1.00 –2.00 X 175, add +3.00; (2) outdoor Rx: OD –3.00 –1.75 X 012, OS –1.00 – 2.00 X 175, CPF527 tint with side shields; (3) four 20-diopter Fresnel prisms; (4) a convex mirror; and (5) a mobility light. In addition, an initial O&M assessment was done. Strategies to improve mobility may include the introduction of a long cane or dog guide, changes in walking speed and visual scanning corresponding to the environment, and optical devices (Geruschat & Turano, 2002). The low vision practitioner and O&M instructor determined that introducing the long cane technique, along with field enhancement with Fresnel prisms, was the best approach for James.

When James returned to the center to receive the devices and to have the prisms placed, he had a positive initial reaction to the updated spectacles. The prisms were initially placed on the clear Rx and would later be placed on the sunglasses, if successful. James was placed 1 meter from a tangent screen and asked to point his nose directly at the center. He was then instructed to look at a target 20 degrees to the left without moving his head. With one eye occluded, a piece of adhesive tape was moved from the area of deficit to the field of vision. James was asked to indicate as soon as he saw the leading edge of the tape. This procedure was repeated for the other eye. James then looked at the target 20 degrees to the right without moving his head, and the placement of the adhesive tape was repeated. Once all four edges were determined, binocularity was assessed. The edge of the tape was modified until both eyes reached the edge of the prism at the same time to eliminate diplopia. Once the edges were determined, the prisms were placed on the rear of the lenses with the base to the deficit and were trimmed to maximize cosmesis.

Next, James was instructed in the use of the prisms. Training started with simple tasks and progressed to more complex tasks, as described by Weiss and Brown (1995). Initially, James was asked to look straight ahead to avoid looking through the prism. While he looked straight ahead, a pen was place to the right outside his field. He was asked to look right with just his eyes to locate the pen and then touch the tip with his finger. He then tried to touch the tip, but, because of the image displacement, he missed the pen. Next, James was told to turn his head to the right and find the pen through the clear center zone to see the actual image. Then, he was asked to locate a moving target while sitting; he was to spot a person walking through a doorway to his left with the prisms. This concept was reinforced by taking him into the hallway and asking him to try to spot objects through the prism and then to locate the objects through the center of the lenses by turning his head while moving. The use of prisms was described as an early warning system, and was compared to the use of a rearview mirror to check a blind spot while driving. Once James understood the proper use of the prisms, he was told to use them only in a familiar setting and during his O&M lessons until his next appointment, when they would be reevaluated.

During this visit, James also received the convex mirror, which he was to anchor to his desk at work and position so he would be able just to glance into it and locate someone approaching him from the side and rear. He also received the mobility light and was instructed in its use. One mobility session was to be scheduled at night to practice with the device.

At his next follow-up visit, James reported that the prisms were “strange initially” but that he thought they were helping. He did not feel the prisms were in the way. The O&M instructor’s assessment concurred with his. James was asked to walk down the hallway and to locate a moving object to demonstrate how he was using the prisms. He initially located an object in the prisms and then looked at the actual image with the center of the lenses. James was instructed to use the prisms in more complex settings and return in one month.

When James returned in one month, he said that the prisms were successful. He was able to use them in the grocery store and while walking in his neighborhood and continued to use them during O&M training, when their proper use was emphasized. He did not use them at work because he thought that his depth perception was altered because of the motion of the machinery; it seemed as if objects were coming toward him when they were actually not nearby. James was more comfortable wearing his old safety glasses without the prisms at work. Also, convex mirrors that were located in hallways and at various intersections in the plant aided his mobility. After James received the desk convex mirror, he realized that it was a great aid to him and made an effort to use it. Prisms were also placed on his sunglasses at this time.

At this point, James had completed five O&M lessons, during which the long cane technique was integrated with the prisms. With this technique, James used a sweeping motion of the cane in front of him that allowed him to keep his eyes up and warned him of changes in contour, curbs, potholes, or other objects. The prisms also allowed James to keep his focus forward while gathering information from the periphery. He was taken on routes that he would normally take to increase his familiarity with traffic patterns while using the techniques, including one night lesson, when the mobility light was incorporated, which increased his sense of safety and confidence while traveling.

At his six-month visit, James said that he was still using the prisms. The prisms occasionally came off during cleaning, but he was able to reapply them without difficulty. The position of the prisms was checked and found to be proper. James was told that the prisms could yellow and harden with age and might need to be replaced at regular intervals. He was to return in one year, so that his vision and use of the prisms could be evaluated.

Discussion

Retinitis pigmentosa offers the low vision practitioner a great challenge. A person with a constricted field (with or without reduced acuity) acts much differently from one with macular degeneration. A good refraction needs to be done—a step that is often neglected when the medical aspect of the disease is emphasized. With persons with reduced visual acuity, the use of magnification can be applied conservatively. Glare control and illumination play a larger role. Mobility, in most cases, is the biggest problem. Persons with retinitis pigmentosa travel slower than do those with typical vision and are five times more likely to have a mobility incident under reduced illumination (Geruschat, Turano, & Stahl, 1998). Despite low success rates, field enhancement should be attempted.

The first step is to determine a precise refractive correction. A trial-frame refraction with a variable illuminated chart maximizes acuity (Weiss, 1991). The trial frame allows for a wider field of view and gives the person the ability to scan. By varying illumination, the person can be made comfortable with the least amount of glare. Abnormalities in binocularity should be addressed to limit asthenopia, since the person tries to maintain fusion with significantly reduced fields (Faye, 1984). In persons with better acuity, reading difficulties may be addressed simply with an appropriate add.

People with retinitis pigmentosa are affected by glare in two ways: (1) general photophobia because of light scatter, since the retinal pigmented epithelium can no longer absorb light, and (2) poor adaptation to different lighting levels because of the lack of photoreceptor function (Brilliant, 1999). Therefore, a tint evaluation should be done with various tints. Using wraparound-style frames, side shields, and sun visors helps to cut down additional glare from the sides and above. When adaptation to various lighting levels is an issue, photochromatic lenses or a gradient tint may be appropriate. It is not unusual to prescribe three or four different pairs of spectacles because of the various lighting situations that individuals may encounter. Impaired scotopic vision makes mobility at night and in dim illumination difficult. A wide-beam mobility light, which can illuminate an area up to 10 feet wide in front of the individual, is a simple solution.

Magnification needs to be used conservatively for persons with decreased acuity and constricted fields. Too much magnification may enlarge the image outside the field and hence make reading more difficult. For those with a moderate impairment, simple reading glasses or a high-add bifocal may be sufficient. For those with a more severe impairment, stand magnifiers, hand magnifiers, and closed-circuit televisions (CCTVs) can be effective. CCTVs allow a person to use more magnification from a greater viewing distance, giving a larger field of view. The contrast and brightness can be controlled, allowing the person to personalize the image. Some prefer reverse polarity, and others like various color combinations. Distance magnification is even more restricted (Weiss, 1991). Higher powers are not successful. A 2.2X or 2.5X can be tolerated in some cases. The minimum amount of magnification to achieve the goal will be the most successful for distance and near.

Constricted fields usually give persons with retinitis pigmentosa the most difficulty. Although field-enhancement techniques have had limited success, they should be attempted. Field enhancement can used for sighting or mobility. Sighting assumes that the person is stationary while trying to observe an area larger than his or her field. A large minus Fresnel lens can be placed on a door or window to gather information (Cohen, 1992). Minus lenses held at arm’s length allow the person to survey an area. Strategically placed convex mirrors, on desks or in a work space, allow the person to locate objects behind him or her or in a blind spot more efficiently (Weiss, 1991). Mirrors can also be placed in hallways or rooms to provide information about doors, people, or other obstacles (Brilliant, 1999).

Field-enhancement systems for mobility work by increasing the effect of the person’s habitual scanning skills. They should be taught in conjunction with an O&M instructor. Various systems have been described, including reverse telescopes, amorphic lenses, and the use of various prismatic systems. Since the success rate of these systems is not high, a person should be motivated and need to travel independently, and his or her acuity should be only moderately impaired, better than 20/100 (Jose, 1983). Goldmann visual fields should be plotted; a person whose visual field is less than 20 degrees but greater than 5 degrees is the best candidate. Close attention should be paid to peripheral islands that the person may not even be aware of. The more recent the field loss, the more easily the person will adapt. In addition, a person needs to have sufficient cognitive ability and be physically able to use the system. Thus, the system is contraindicated for those who are wheelchair bound, use walkers, or have a hemiparesis (Perlin & Dziadul, 1991).

Reverse telescopes can be used in handheld or bioptic form (Cohen, 1992; Drasdo, 1976; Mehr & Quillman, 1979). The overall field of view is constricted into the individual’s field. People can have a good initial reaction to this system in the examination room but a poor reaction while moving in the real world. Their visual efficiency has been shown not to improve significantly with the use of reverse telescopes (Krefman, 1981), because spatial location becomes more difficult (Weiss, 1991) and their visual acuity suffers because of the minification.

The use of amorphic lenses has also been attempted (Cohen, 1992; Szlyk et al., 1998; Weiss, 1991). An amorphic lens is a cylindrical reverse telescope with minification only in the horizontal meridian. It reduces the decrease in acuity but induces a large amount of visual distortion and can cause people to become dizzy and disoriented. The use of an amorphic lens in bioptic form, along with a comprehensive training program, has been somewhat successful in areas of peripheral detection, recognition, scanning, tracking, visual memory, and mobility (Laderman, Szlyk, Kelsch, & Seiple, 2000).

The use of various prism systems is another alternative method for field enhancement. One type is a fixed prism system (Gottlieb, Freeman, & Williams, 1992). Another approach is the use of Fresnel prisms, which are less costly and easily removed if they are not tolerated. Although more success has been reported in rehabilitation of hemianopias (Cohen, 1992; Perez & Jose, 2003), attempts have been made with overall constrictions. Several techniques have been described. Weiss (1990) attempted to butt the prisms together with the base out, bisecting the pupil. Jose (1983) arbitrarily selected 30-diopter prisms and placed them base out around the constriction just outside the field in a more peripheral location (Jose & Smith, 1976).

The technique used in the case presented here was described by Perlin and Dziadul (1991). Fresnel prisms were placed base out around the constriction, but a central zone was left for scanning. It was determined that an ocular rotation of 20 degrees in both directions was comfortable, leaving a central area of 40 degrees for scanning. This area could be modified, depending on the individual's reaction during training. The optimum initial prism power was determined to be 20 diopters, since 30 diopters is often rejected (Perlin & Hoppe, 1992), and anything less than 20 diopters would not give enough displacement to be effective. As with the bioptic amorphic lens, the best success was with extensive training in conjunction with an O&M instructor.

Good communication between the low vision provider and the O&M instructor is essential. The O&M instructor can evaluate the individual's use or lack of use of prisms in the real world and may observe modifications that need to be made in the placement of the prisms. The prisms may need to be moved in or out, depending on the individual's reaction, and more or less image displacement may be needed to ease the location of objects.

Even though James had a good reaction to the therapy, there are still many negatives to this method. In contrast to conventional prisms, there is a significant loss of clarity and brightness and an increase in distortion and glare with the Fresnel prisms (Cheng & Woo, 2001). Furthermore, prisms can be difficult to clean, are often displaced, harden and yellow over time, and need to be replaced. The concept of image displacement can be confusing, and image jump can be disorienting.

Conclusion

In the case presented here, James benefited from comprehensive vision rehabilitation and continued to work. He discussed his condition with his supervisors and asked to have his floor duties reduced and to concentrate more on desk work. This request was granted. The simple update in eyeglasses clarified his distance vision and allowed him to continue with the demands of his near work without difficulty. James's quality of vision was increased through the use of tints for glare control. The addition of a mobility light allowed James to travel at night. Although the use of prisms was not a total success in the workplace, James did benefit from the prisms and the O&M training. He became a much more efficient scanner with and without the prisms and much more confident in general.

Even though there is a high rate of rejection with the various field-enhancement systems, these systems should be attempted. The process of training with field-enhancement techniques, even if they are rejected, makes the individual more aware of his or her field and a better scanner—and thus a safer and more confident traveler. More research needs to be done in this area to develop a more effective system that allows a person to continue to use scanning methods while enhancing the visual field.

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Errata:

In the article, "Low Vision Driving with Bioptics: An Overview," by Chuck Huss and Anne Corn, which appeared in the October 2004 issue of the Journal of Visual Impairment & Blindness (JVIB) (Volume 98, Number 10), the definition of low vision should have appeared as "less than 20/40 best corrected vision in the better eye." In addition, the section on low vision driver programs should have stated, "When a prospective driver demonstrates competence behind the wheel, a referral is made by the driver education instructor who determined his or her competence to the motor vehicle authority…." JVIB regrets these errors.

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