Adaptation to Macular Scotomas in Persons With Low Vision

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Persons with scotomas in their central 20° of vision often do not notice these blind spots within their visual field and have visual performance difficulties far exceeding what would be expected from standard vision tests. Before persons with macular scotomas can be assisted to optimally use their remaining vision for a better quality of life, more must be known about how the visual system adapts to a macular scotoma. Important issues include spatial and temporal characteristics of perceptual completion and metamorphopsia, development of preferred retinal loci for fixation and visual search, and dynamics of the preferred retinal locus development in terms of the changes in the eye movement system. With a full understanding of the visual system's adaptation to macular scotomas, new low vision devices and training techniques can be proposed to promote independence in activities of daily living for the person with low vision.

It is not uncommon to hear comments such as the following from persons in a low vision rehabilitation clinical service:

- “When I try to catch fly balls while playing baseball, the ball disappears and then reappears but not always in time for me to catch it.”
- “When I look at a door or a window frame, it appears bent.”
- “When I look at a word or part of a sentence on a page, the letters or words to the right of where I am looking are so blurry that I cannot recognize them.”
- “I cannot seem to walk straight or walk without things hitting me; things (chairs, fences, and so forth) seem to leap out and hit me.”
- “Outdoors in the evening or when I go indoors, more of the objects (signs, faces, and so forth) become bent or blurry.”
- “My vision has not become any worse (and my physician agrees), but I cannot read as well as I did before, even with my low vision device.”

These persons are probably trying to perform activities of daily living (ADL) with one or more macular scotomas (sometimes called blind spots) unknown to them or their health specialists. Why are some persons with low vision aware that they have these blind spots, whereas others with low vision perform ADL completely unaware that their problems in visual performance are due to macular scotomas?

Characteristics of Macular Scotomas

Macular scotomas are retinal areas with reduced light sensitivity (compared with sensitivity results of persons with normal vision) in the central 20° of the visual field. Macular scotomas are specified by the retinal location in that central scotomas are retinal areas with reduced light sensitivity involving the fovea, whereas paracentral scotomas are retinal areas with reduced light sensitivity within the central 20° of the visual field but do not involve the fovea. Macular scotomas are further defined by the light intensity used to map out their extent. For example, dense scotomas (sometimes labeled absolute scotomas) are retinal areas that are insensitive to very bright objects, whereas relative scotomas are retinal areas that are insensitive to a light level relatively less than the very bright object. Most clinical services and studies of the central visual field have used standard clinical perimetry equipment (Amsler grid, tangent screen, and Humphrey Visual Field Analyzer1). Recent results have indicated that macular perimetry with the scanning laser ophthalmoscope (SLO) is more sensitive in detecting small localized scotomas than standard clinical perimetry equipment (Schu-

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chard, 1993; Schuchard & Hu, 1994). Therefore, these small (maybe 5° and less) macular scotomas more than likely have gone undetected by health specialists in low vision rehabilitation services and investigators in studies of the central visual field. Figure 1 shows an example of these types of macular scotomas in a patient with traumatic brain injury.

Macular scotomas are associated with most of the common low vision medical conditions such as age-related macular degeneration, diabetic retinopathy, and glaucoma, as well as less common medical conditions such as postchiasmal central nervous system disorders. In a typical low vision rehabilitation service, 83% of patient eyes were found by SLO macular perimetry to have dense macular scotomas present in the central visual field (Fletcher, Schuchard, Livingstone, Crane, & Hu, 1994). This prevalence of dense macular scotomas is nearly equal to the 80% of patient eyes found by SLO macular perimetry to have dense macular scotomas in a low vision research study (Schuchard, 1993). Fewer than half of these research subjects with low vision were documented, either by their referring clinician or low vision specialist, as having macular scotomas. Central scotomas, with a wide range of acuities, were found in 40% of the research subjects, whereas 50% of the patients with macular scotomas had central scotomas. Paracentral macular scotomas with no central scotoma (and thus often with fairly good acuity) were additionally found in 40% of all the research subjects, whereas 50% of patients with macular scotomas had paracentral macular scotomas. In the low vision rehabilitation study (Fletcher et al., 1994), only 16% of the patients with macular scotomas had scotomas of less than 5° in size. Most of the persons with macular scotomas (84%) referred to the low vision rehabilitation service had scotomas greater than 5° in size that substantially affected their ADL. We may never know the true prevalence of macular scotomas in the general population; many persons may have scotomas less than 5° in size and never know about them. Regardless, the prevalence of persons with low vision who have macular scotomas (especially paracentral scotomas because central scotomas are easier to document) is considerably greater than suspected by most low vision specialists.

Perception With a Macular Scotoma

One would think that persons with low vision would see black holes where their macular scotomas fall in the central visual field if the visual system was not able to adapt to macular scotomas. Instead, recent research has shown that the visual system has a great capacity to adapt to macular scotomas (Ramachandran, 1992; Schuchard, 1993, 1995). Thus, most persons with macular scotomas do not perceive a black spot in the visual field where the scotoma is located. Persons with normal vision do not perceive a black spot when viewing the visual world with one eye closed. Figure 2 demonstrates both the phenomena of having small objects jumping into and out of view and of larger objects always being in view without gaps, which shows that even persons with normal vision do not perceive a black spot in the visual field where the scotoma is located.

In another example, scotomas from laser treatments of retinal diseases are sometimes described by patients as “large black spots” or “boulders,” but over a period of months, these spots fade and can no longer be localized without macular perimetry testing. Even when persons with low vision are continuously aware of their scotomas, the visual world does not fall into a “black hole”; rather, these persons often see a blurry, hazy, dark or light area and a smaller blind spot size than the actual scotoma size (Schuchard, 1993).

The mapping of object space into perceptual space by observers with a scotoma can include warping or
central scotoma that had been produced by placing the object falls in a scotoma. Even though perceptual completion has been documented in observers with scotomas (Gerrits, De Haan, & Vendrik, 1966; Ramachandran & Gregory, 1993; Schuchard, 1993; Schuchard & Timmerman, 1969; Piantanida, 1985; Ramachandran & Gregory, 1991; Spillmann & Kurtenback, 1992). Perceptual completion can be regarded as a type of visual illusion (as a perception that does not represent the character of an object or a misconception of a sensory impression) (Gassel & Williams, 1963). For example, persons with normal vision will perceive edges or borders where no contract exists (see Figure 3).

Persons with low vision can have different perceptions depending on the expected visual target. Figure 4 shows examples of letters placed briefly over macular scotomas with an SLO. If told before the presentation that they will be seeing geometric objects (lines, circles, and so forth), persons with low vision will report that they see two parallel lines (for the H) and a cone or upside-down V (for the A). However, if told beforehand that they will be presented with letters, these same persons will almost always report that they see a complete H (with no breaks or irregularities) and then an A. Thus, the expected visual target plays an important part in the visual perception of persons with macular scotomas, as it does for persons with normal vision who can be tricked into different visual perceptions by visual illusions.

Preferred Retinal Loci With Central Scotomas

In an eye with a central scotoma, the development of one or more eccentric preferred retinal loci (PRL) naturally and reliably occurs (Culham, Fitzke, Timberlake, & Marshall, 1993; Cummings, Whittaker, Watson, & Budd, 1986; Fletcher et al., 1993; Guez, Francois, Rigaudeau, & O'Regan, 1993; Schuchard, 1993; Schuchard & Fletcher, 1994; Schuchard & Raasch, 1992; Timberlake et al., 1986; Timberlake, Peli, Essock, & Augliere, 1987; White & Bedell, 1990; Whittaker, Budd, & Cummings, 1988; Whittaker & Cummings, 1986). The term PRL is normally reserved for those persons whose visual system has chosen an eccentric retinal area to act as a pseudofovea for visual tasks that were performed with the fovea. The mechanisms in across the scotoma, although the thick rods were perceived to have a somewhat smaller thickness and less precise edges than when seen away from the scotoma. Remarkably, this report is probably the most complete documentation of perceptual completion development in a person adapting to a macular scotoma. Other reports show perceptual completion with cortical scotomas, but even in this literature, there is disagreement as to whether perceptual completion or uncontrolled experimental factors like poor fixation stability are actually producing the perception of complete objects (Sergent, 1988). As early as 1941, the filling in of physiological blind spots and of scotomas was thought to represent some intrinsic organizing function of the cortex (Lashley, 1941). Recent studies have indicated that filling in occurs extraretinally by interpolation of the visual information from the borders of the scotoma (Gerrits & Timmerman, 1969; Piantanida, 1985; Ramachandran & Gregory, 1991; Spillmann & Kurtenback, 1992). Perceptual completion can be regarded as a type of visual illusion (as a perception that does not represent the character of an object or a misconception of a sensory impression) (Gassel & Williams, 1963). For example, persons with normal vision will perceive edges or borders where no contract exists (see Figure 3).

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that decide the specific retinal location or the time course for PRL development have not been discovered. Regardless, the retinal location of PRL relative to the macular scotoma, and thus the position of gaze relative to the blind spot, is often critical for success of visual performance in persons with macular scotomas. Even the relative location of paracentral macular scotomas to the fovea can have drastic consequences for ADL. For example, persons with ring scotomas can have very poor reading rates (Plas, Fletcher, & Schuchard, 1995). In a typical low vision rehabilitation service, 51% of eyes were found to have the PRL to the left or right of a macular scotoma. These persons typically cannot see ahead or scan back to the beginning of a line while reading.

The only accurate means for determining the location of the PRL relative to macular scotomas, the physiological blind spot, or any other retinal landmark is to use a device like the SLO. For example, although one might think that, because the acuity is relatively poorer, the PRL is relatively farther away from the nonfunctioning fovea, the eccentric retinal location of the PRL cannot be determined by visual acuity alone (Schuchard & Raasch, 1992).

The PRL ability, besides the PRL retinal characteristics, has profound effects on ADL. In an eye with a functioning fovea (for example, paracentral scotoma), fixation is the act of directing the eye toward the visual object of regard, causing the image of the object to be within the fovea. Eccentric fixation is the act of directing the eye toward the visual object of regard, causing the image of the object to be within the fovea. Eccentric viewing refers to fixation performance in which the person has the sensation of looking directly at the visual object during fixation with the eccentric PRL. Eccentric viewing, on the other hand, refers to fixation performance in which the person has the sensation of looking above, below, or to either side of the object during fixation with the fovea or PRL. A shift from the fovea to an eccentric PRL in the perception of looking directly at an object is possible in persons with bilateral central scotomas (Schuchard & Raasch, 1992; White & Bedell, 1990). In a similar manner, the fovea or PRL controls large eye movements. How well the PRL develops should predict the ability for the person with a central scotoma to successfully perform ADL involving eye movements (such as fixation and visual search). When persons with low vision are asked to fixate (look directly at) small objects, they may use different retinal locations and therefore exhibit multiple PRLs. These persons with low vision may not even be consciously aware that they are looking at small objects such as words with different retinal locations. These different retinal locations can have different acuity capabilities as well as contrast and other visual function capabilities. When asked to keep their fixation on a small object (fixation stability), persons with low vision will often use a retinal area that is much larger than that used by a normally sighted person. The SLO has shown that persons with low vision can have PRL as small as that of persons with normal vision (1°–2°) and as large as 9°.

Persons with low vision must use their PRL when searching for objects during scanning or other eye movements. When asked to follow or track a moving object, the person with low vision may have difficulty if the object is moving into the macular scotoma. Additionally, the macular scotoma may inhibit their visual search capabilities.
Figure 4. Illustration of two letters, H and A, projected onto the retina so that the images of the cross bars of both letters (black pattern) are falling into the dense macular scotomas (gray pattern). Persons with low vision can report two different perceptions—two parallel lines or an H versus an upside-down V or an A, depending on what type of object the person with low vision is expecting to see.

scotomas have been found to place the center of a large object at their PRL, even when verbal instructions were given to place the center of the large object at their non-functioning fovea (Schuchard & Rassch, 1992; White & Bedell, 1990).

Up to this point, the discussion of PRLs has ignored the fact that persons with low vision exist in a binocular visual world, whereas most of the concepts of the PRL have been developed monocularly. In a study investigating binocular correspondence, 80% of the 67 subjects saw objects with only one eye (monocular perception) while performing a simple free viewing binocular visual task (Schuchard, Tekwani, & Hu, 1995). However, this study does not conclude that the subjects are seeing the world with only one eye. The task was to fixate small visual targets at the PRL location. Subjects reported seeing the world with both eyes, although not always appreciating that, when looking at a small object (such as a number, letter, or word), the small object was only seen by one eye. Previous studies of PRL abilities in eye movements (Schuchard & Fletcher, 1994; Whittaker & Cummings, 1986; Whittaker & Cummings, 1990; Whittaker, Cummings, & Swieson, 1991) have used similar tasks. Typically, the subject is asked to fixate, saccade to, or follow a small object. The eye used by the visual system in binocular tasks may be the dominant eye, whereas the fellow eye is actually following the lead of the dominant eye in eye movement and other visual tasks. Therefore, the poor results in the aforementioned PRL studies may be due to forcing the fellow eye to perform eye movement tasks without the dominant eye. The study by Schuchard et al. (1995) implied that the visual system does not (cannot?) typically retain binocular correspondence because only 20% of the subjects saw the object with both eyes (one subject retained binocular perception with anomalous retinal correspondence). This lack of binocular correspondence has dramatic implications for persons with low vision in ADL, possibly including the loss of visual function such as depth perception from disparity and rivalry, diplopia, or suppression instead of fusion when viewing binocular objects.

ADL and Macular Scotomas

Clearly the visual system has a great capacity for plasticity in adapting to macular scotomas. Persons can adapt to central blind spots by developing PRLs to perform foveal tasks (such as fixation and reading) on eccentric retinal areas. This adaptation may occur without the conscious knowledge of the person with low vision and may not be detected by the low vision specialist. Thus, the person with low vision may experience strange or unusual visual sensations during ADL. What are the possible visual circumstances that could lead to the comments by persons with low vision stated at the beginning of the article?
"When I Try to Catch Fly Balls While Playing Baseball, the Ball Disappears and Then Reappears but Not Always in Time for Me to Catch It."

Persons with small paracentral scotomas often have good acuity with small blind spots just above the fixation location in the visual field. Balls or other small objects flying in the air that are in the visual field corresponding to the macular scotoma cannot be seen until the image falls on healthy retina. Because the person does not see a black hole, there is no perception of a hole in his or her vision. Thus objects appear to dance into and then out of view, without any understanding of why this perception is occurring.

“When I Look at a Door or a Window Frame, It Appears Bent.”

A person with similar visual field conditions as described for the balls can look at large objects and “see” the whole object. However, if the person cannot fully adapt to the macular scotoma, or the macular scotoma is too large for the perceptual completion mechanisms to successfully fill in the visual information, the perception of the large object can be distorted. This perception is especially noticeable when the object has straight edges that the person with low vision knows should be uniform straight lines.

“When I Look at a Word or Part of a Sentence on a Page, the Letters or Words to the Right of Where I Am Looking Are So Blurry That I Cannot Recognize Them.”

Persons with a central scotoma to the right of the PRL or with a paracentral scotoma immediately to the right of the fovea may not be aware of the blind spot caused by the scotoma. The perceptual completion process naturally fills in the type of visual information that is expected (letters or words), but the visual system will not know the specific information that is at the spatial location of the scotoma. Thus the perception is often of blurry objects with no gap that are generally correct but without specific visual information. For example, the person perceives a blurry unrecognizable letter or word because the specific letters or words inside the scotoma cannot be “seen” by the visual system.

“I Cannot Seem to Walk Straight or Walk Without Things Hitting Me; Things (Chairs, Fences, and So Forth) Seem to Leap Out and Hit Me.”

Persons with a central scotoma to the right or left of the PRL or persons with a paracentral scotoma to the right or left of the fovea may have large scotomas without an appreciation of the true extent of the blind spot. Thus, the perceptual completion process will fill in the edge of the blind spot that gives the perception that the path is clear, whereas in reality the chair that is just inside the blind spot is not seen until the last second, when the image of the chair falls on healthy retina outside of the scotoma.

“Outdoors in the Evening or When I Go Indoors, More of the Objects (Signs, Faces, and So Forth) Become Bent or Blurry.”

Because relative scotomas are often larger than dense scotomas, the blind spot will suddenly become bigger when the person with a macular scotoma moves from the outdoors or any other well-lit environment to a dimly lit environment. Thus the light level of the environment can change the visual perception and exacerbate difficulties in ADL for the person with low vision.

“My Vision Has Not Become Any Worse (and My Physician Agrees), but I Cannot Read as Well as I Did Before, Even With My Low Vision Device.”

Unfortunately, macular scotomas can increase in size with the progression of many macular diseases, but this increase in the blind spots may not change the results of other vision tests. For example, a ring scotoma could substantially increase in size without affecting the center of the ring (the fovea), and thus visual acuity and contrast sensitivity would remain relatively unchanged. This change in size of the macular scotoma may not be detected by the low vision specialist, and thus the patient is left wondering what, besides a vision change, is causing the increased difficulties in ADL.

Conclusion

Persons with macular scotomas have visual performance difficulties far exceeding what would be expected from standard vision tests. Before professionals can assist these persons to optimally use their remaining vision for a better quality of life, we must know how the visual system adapts to a macular scotoma. Questions worth investigating are: (a) What are the limitations of the visual system’s plasticity in adapting to macular scotomas? (b) Why do only some persons develop PRL with relatively good ability? (c) Is there one PRL for all visual tasks or ADL? (d) Does the ability of the PRL in visual tasks predict rehabilitation potential? Because potentially four of five persons with low vision are performing ADL with macular scotomas, only when the answers to these types of questions are found and made available to low vision specialists, including occupational therapists, will these persons have thorough low vision rehabilitation care.
References


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