

Visual Function and Age Related Macular Degeneration

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To call a person with severe vision loss "legally blind" is as preposterous as calling a person who is severely ill "legally dead". The old, simplistic dichotomy between those who are 'legally sighted' and those who are 'legally blind' is not valid, since there is a spectrum of vision loss from individuals who have lost a little to those who have lost almost all.

Importance of History Taking to Assess Functional Impact of Vision Changes

A thorough functional history is the key to assessing the impact of vision changes on the life of a patient. The functional history reveals how the patient is using remaining vision, and also gives clues to adaptations already being employed by the patient. It demonstrates the physician's concern and opens the door for the patient to relate fears and concerns which are often not communicated in routine eye care. Even for ophthalmologic practices not doing comprehensive rehabilitation, a functional history will guide appropriate referral.

Those with "normal" vision use it in widely varying ways. Likewise for those with decreased vision, the loss can have widely varying functional implications. These are related to the type of loss, the extent of the loss, the type of activities that were previously engaged in, as well as the emotional reaction to the loss.

The type of activities that the individual has previously made part of his or her life is very significant to the impact of vision loss. A mild reduction in central acuity affects an illiterate warehouse laborer who enjoys participating in wrestling and socializing at the bar very differently than a jeweler who stops at the library every third day to replenish his reading material and plays left field for his baseball team.

Depending on the individual's lifestyle, the impact of the same degree of vision loss can vary from having no effect to causing a major life disruption.

It is valuable to identify the nature of the support network available to the individual. Do they live with a healthy spouse or have the responsibility of caring for one with Alzheimers? Are children or friends nearby to offer assistance? Do they live in their own home or a care facility where meals and domestic care are provided. As well, other coexisting physical and sensory impairments can be significant in the rehabilitation plan and need to be identified.

Functional History Technique

Vague questions like: "How are you doing?" may or may not illicit useful information; a one word answer like "fine" is a frequent response that doesn't reveal much. More specific questions are generally needed and they can yield meaningful insights; they also demonstrate a sensitivity that will help to open communication lines further.

Examples would include

- General questions - to determine problem areas and their significance to the patient.
 - eg. "How has your life changed since the onset vision loss?"
 - eg. "What are your plans for the future?"

- Specific questions - directed at specific tasks that may be problematic. As appropriate for the individual, questions can be directed along such lines as:
 - eg. "Do you read the newspaper and how?"
 - "Do you do your own mending?"
 - (to assess ability to do detailed near tasks)
 - eg. "How do you get to the grocery store and manage in the aisles?"
 - (to assess orientation and mobility skills)
 - eg. "Can you follow the action on a television program, the ballet or read the overhead menu at a fast food restaurant?"
 - (to assess detailed distance vision abilities).

Phantom Vision

One unique aspect of the low vision history should be an inquiry into whether the Charles Bonnet phenomenon (phantom vision) has been experienced. In this phenomenon individuals with decreased vision experience photopsias or formed hallucinations (similar to phantom leg pain noted by amputees). This unusual visual experience has been reported in greater than 50% of patients seen for low vision evaluation.¹ The sensation is not usually reported voluntarily as patients are often concerned that doing so would be construed as evidence that they "are losing their marbles". Most patients are greatly appreciative to learn that their "seeing faces or animals" is a common experience and not a manifestation of some other serious problem such as a psychiatric condition.

Sensitive Issues

Many individuals feel significant discouragement and distress as a result of their visual impairment. Knowledge of this can aid in the use of reassurance therapy (see chapter 10) or guide referral for counseling or support groups.

In taking the history this information can often be gained by tactfully using a third party questioning technique such as this: "Many of my patients have experienced - feelings of discouragement - unusual visual experiences - etc. Have you?" State that other people have the problem in question and then ask them if they do. Knowing that they are not alone gives the patient an easier answer if they are troubled. This allows them to give an honest answer without feeling self conscious. It also allows the doctor to ask the question without looking suspicious or implying doubt in their coping abilities.

REFERENCE

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Visual Function Testing in Macular Disease

Assessment of the many aspects of visual function is a key component to management of ARMD. While visual acuity is an important measure of function at the center of the macula, by itself it does not provide the total picture of macular function. Contrast Sensitivity and Central Visual Field are also critical factors in determining macular function. Understanding these perimeters helps guide the therapy of the clinician and the evaluation process also provides an opportunity to educate patients about the visual resources they still have, emphasizing what is left rather than what is gone.

CONTRAST SENSITIVITY MEASUREMENT IN ARMD

Contrast Sensitivity describes the ability to detect objects of low contrast. Like visual acuity it can be a sensitive but non-specific indicator for a variety of problems in the visual system. Contrast Sensitivity is defined as the reciprocal of the lowest contrast that can just be detected (compare: visual acuity as the reciprocal of smallest symbol size that can just be recognized).

Poor low contrast vision can be related to significant difficulties in activities of daily living. Typical low contrast tasks include driving in rain or fog, walking down steps, pouring milk into a white cup, etc. Magnification is a useful remedy if small details of high contrast cannot be seen (e.g. reading); it does not necessarily help in the detection of objects of poor contrast. Reduced contrast sensitivity is often seen in patients with more generalized retinal involvement, such as macular edema and geographic atrophy. Such patients often are extremely responsive to an improvement in illumination, even more than to increased magnification.

The Peli-Robson chart is particularly useful, because it gives large letters/symbols that can be seen by most low vision patients. Contrast sensitivity charts should contain rows of at least 3 letters/symbols at each contrast where most patients can read the top row but not read all rows. It is critical that appropriate lighting is always maintained as the results of the contrast sensitivity test can be much poorer than the patients ability if the lighting is not at or above the appropriate level (commonly about 80 to 100 candela per meter squared). A different approach is the use of a low contrast letter chart with variable letter sizes of a fixed low contrast. This innovative approach is used in the SKILL chart (Smith-Kettlewell Institute Low contrast, Low Luminance chart), a handheld chart with a regular letter chart (black on white) on one side and a low contrast, low luminance (black on dark gray) chart on the other side. When both sides of the chart are viewed under the same office conditions (same illumination, same viewing distance) a normal eye will read two lines less on the low contrast side; a larger difference indicates a loss of contrast sensitivity.

VISUAL FIELD MEASUREMENT IN ARMD

Macular perimetry is one of the most important, but often neglected, aspects of vision assessment in macular disease. Macular scotomas have a major impact on the performance of activities of daily living and have been reported to exist in about 91% of patients referred to a low vision rehabilitation service.¹ For example, the presence or absence of a central scotoma or paracentral scotoma is a much more powerful predictor of reading speed problems than is visual acuity.² Knowledge of the central visual field has great clinical value for training the patient in the effective use of residual vision and facilitates rehabilitation in activities of daily living. Since most eye diseases often progress slowly over a considerable period of time, patients have the opportunity to adapt slowly by developing the habit of making more scanning eye movements. It is not unusual for patients to be aware that they are 'clumsy', but to be unaware that this is because they have a visual field loss. If patients have good central acuity, the practitioner may forget to do a visual field. Family members may also have a difficult time understanding the condition. Everyone can imagine what it is like to be in a fog: one can see in all directions, but only for a limited distance. Hence the common question: how far can you see? They cannot understand that the patient can read the clock across the room, but have great difficulty navigating a page of text at a close distance. It often is an enormous relief for patients to learn that they have a 'real' disease and are not just clumsy or malingering.

Macular Perimetry Testing

Perimetry in the central visual field (central 20 degrees or roughly the area between the temporal vascular arcades) presents unique challenges not encountered in the more well known peripheral visual field analysis. Patients with diseases of the macula often have disrupted fixation behavior.^{3,4,5} Screen and projection perimetry rely on steady and accurate fixation to accurately plot the visual field; when fixation is unsteady or located in different retinal areas these tests have no method of monitoring or compensating for retinal movement. Using the physiological blind spot as a fixation monitoring technique still allows for a 5 degree unsteadiness in fixation without detection. These strategies for testing the central visual field, at best, indicate where scotomas are found relative to fixation but they provide no information about the absolute retinal location or quality of fixation.

Much has been learned about the central visual field by Scanning Laser Ophthalmoscope (SLO) macular perimetry which allows the investigator to determine the retinal location of visual stimuli directly on the retinal image in real time. Fixation can be monitored and even in patients with very unsteady fixation, a macular scotoma can be identified in relationship to the anatomy of the macula. Accurate SLO central visual field analysis has demonstrated that 91% of low vision patients have significant central or paracentral dense scotomas in their central visual field.¹

SLO macular perimetry has shown that the size, location and density of scotomata cannot consistently be inferred from the ophthalmoscopic appearance of the macula. Macular areas with altered appearance, such as areas of retinal pigment epithelial atrophy or chorioretinal scarring, often have diminished or extinguished function but that the loss is not uniform. Grossly disturbed function has also been demonstrated in areas of normal appearing retina. Scotomata may be round or have complex ameboid shapes. The threshold sensitivity of the PRL has wide variation. Those with elevated threshold sensitivity at their PRL can be predicted to require higher intensity of lighting to achieve optimal performance. The PRL may be bordered on many sides by scotomata. SLO studies have shown that the 18% of low vision patient's eyes will have scotomas on 3 or 4 sides of their PRL; they are likely to have many more functional difficulties with activities of daily living than other patients with identical acuities but less encumbered PRLs.^{2,7,8}

Macular perimetry should be performed with flashing lights and not pattern instruments like the Amsler Grid. Amsler grid testing and other pattern perimetry instruments, although easy to use, are very insensitive to detecting the existence of macular scotomas and very inaccurate at determining the extent of the macular scotoma due to perceptual completion. While the scanning laser ophthalmoscope is the preferred instrument for macular perimetry, its cost prohibits widespread use. The tangent screen can be an effective instrument for macular perimetry if a skilled perimetrist uses the appropriate testing protocol. Macular perimetry is best performed with a hybrid technique because of the special characteristics found in patients with macular disease. These patient characteristics include poorer fixation stability ability and the possibility of eccentric fixation. The hybrid technique selects a single stimulus intensity for all test locations, like kinetic perimetry, but presents the target at specific retinal locations (visual field locations) as in static perimetry. This hybrid technique obtains isopter visual field information at a minimum amount of time but at the expense of obtaining full threshold values. Static techniques (like the Humphrey Visual Field Analyzer 10-2 protocol) are also insensitive to detecting small macular scotomas and inaccurate at determining the extent and depth of the macular scotoma. Finally, the kinetic protocols on the Humphrey Visual Field Analyzer or Goldmann Perimeter can be used if the perimetrist is careful to understand the problems associated with kinetic protocols (e.g., reaction time influencing the real border and eye movements tracking the movement of the stimulus).

Careful clinical observations often give hints about central visual field topography that are confirmed by the SLO. Much information can be gained from careful observation of the patient's performance on a variety of visual tasks. The manner in which a letter chart is read can give useful information. If it is noted that one side of the chart is consistently missed, it is likely that a scotoma is present on that side of fixation. Rapid wide searching motions of the eye or head while reading the chart imply a large central scotoma with an eccentric viewing location. Difficulty with both small and very large letters with best performance in the middle letter size range may indicate an island of vision surrounded by a ring scotoma. A marked improvement in reading rate with good illumination may indicate that fixation is located in the vicinity of a relative scotoma that

encroaches on the useful field when inadequate luminance is provided.

PREFERRED RETINAL ABILITY IN ARMD

The visual system of a patient with a central scotoma naturally and consistently chooses (unconsciously) a preferred eccentric retinal area to perform the visual tasks that the nonfunctioning fovea used to perform. The patient must use this eccentric retinal area (called the preferred retinal locus or PRL) for visual tasks because the foveal area can no longer perform visual tasks like fixation, reading, or tracking. For patients with a central scotoma, visual tasks are performed by aiming the eye such that the image of the visual target is placed within the PRL. The PRL in essence becomes a pseudofovea and assumes many of the tasks of the nonfunctioning fovea, i.e. focused attention, object recognition, detail discrimination, oculocentric reference point, etc. Thus, the ability of the preferred retinal locus to perform fixation stability, pursuit movement, and saccadic movements will determine the performance abilities in many activities of daily living. In addition, ARMD patients may use more than one preferred retinal locus depending on the stimulus conditions or the visual task. The relative location of the one or more preferred retinal loci to macular scotomas will also indicate the degree of difficulty in adapting to the existence of the macular scotoma and thus the ability of the preferred retinal locus. Those patients with macular scotomas that encircle the preferred retinal locus, ring scotomas, have been reported to experience great difficulty in activities of daily living while often still having fairly good acuity test results. Preferred retinal locus ability testing is easily done with a scanning laser ophthalmoscope but, again, the cost of the instrument prohibits its widespread use. Video eye tracking devices are a good cost alternative and may yet still retain adequate accuracy for the clinical testing of the preferred retinal locus ability. Regardless, the ability of the preferred retinal locus to perform fixation stability, pursuit movement, and saccadic movement along with the locations of the preferred retinal loci relative to the existing macular scotomas should be assessed.²

The binocular aspect of using the preferred retinal locus in activities of daily living also needs to be assessed. The binocular correspondence of the preferred retinal loci in each eye can be assessed with a "Worth 4 Dot" test. In the simple example of a patient with a central scotoma in one eye and a normal functioning fovea in the other eye, the binocular preferred retinal locus test would determine whether a stimulus is seen with both eyes (binocular correspondence) or only in the eye with the functioning fovea. The single eye perception, the use of a dominant preferred retinal locus, has been reported in about 2 out of 3 (67%) of low vision patients.⁶

A variety of methods have been proposed to teach ARMD low vision patients eccentric viewing techniques to compensate for central scotomas. SLO testing can identify where the naturally chosen PRL is located. Such information can abbreviate the training time. Words can be projected onto the retina and patterns of letter distortion/disappearance noted by both the patient and trainer. During SLO macular perimetry testing, the patient can be taught what a scotoma is and the relationship of the PRL to the scotoma can be demonstrated. Knowledge of the central field abnormalities can help in predicting what difficulties may be encountered in Activities of Daily Living (ADL). Knowing, for example, a scotoma to the left of the PRL makes the saccadic eye movement to the left hand margin of the page difficult after completing a line when reading. Patients with large central scotomata may experience difficulties with hand-eye coordination for such activities as writing, slicing foods, threading a needle, pouring liquids, filling insulin syringes, putting toothpaste on the toothbrush, etc. Patients with tight ring scotomas will have a limited response to magnification. The threshold sensitivity of the PRL is an indicator of the need for extra illumination for the patient. This knowledge helps to differentiate those that have a poor level of motivation for rehabilitation from those that have a greater visual deficit as an obstacle.

If a small island of reasonable vision is sandwiched between scotomas, letter chart acuity may be fairly good. But if the area is too small to accommodate words, reading acuity may be much worse and moderate magnification is not effective. In this case significant additional magnification may be needed to allow the patient to use a more eccentric area where a larger contiguous area is available. SLO studies have shown that some patients use different retinal loci for different purposes (small vs. large objects, dim vs. bright objects).

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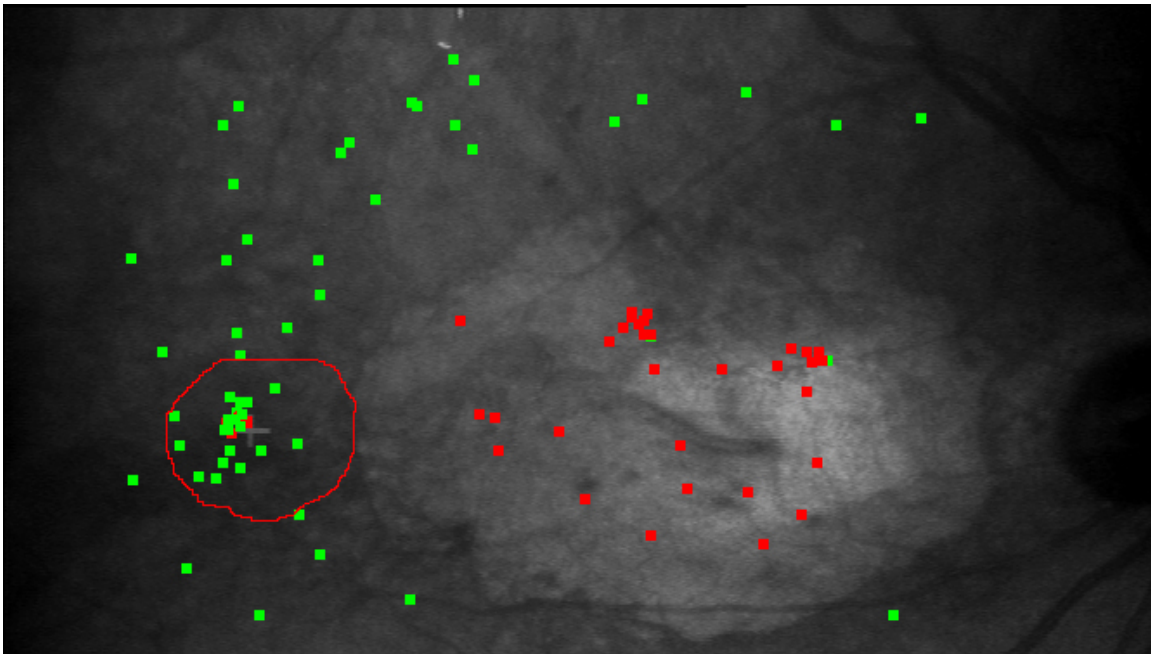


Figure #1

Scotoma Following Visible Anatomical Disruptions

(Note borders of dense scotoma correspond to areas of visible anatomical irregularity.)

Red dots indicate bright 50,000 Troland stimuli not seen by patient (dense scotoma)

Green dots indicate bright 50,000 Troland Stimuli seen by patient (functioning retina)

Red Circle indicates area of Preferred Retinal Locus (PRL)

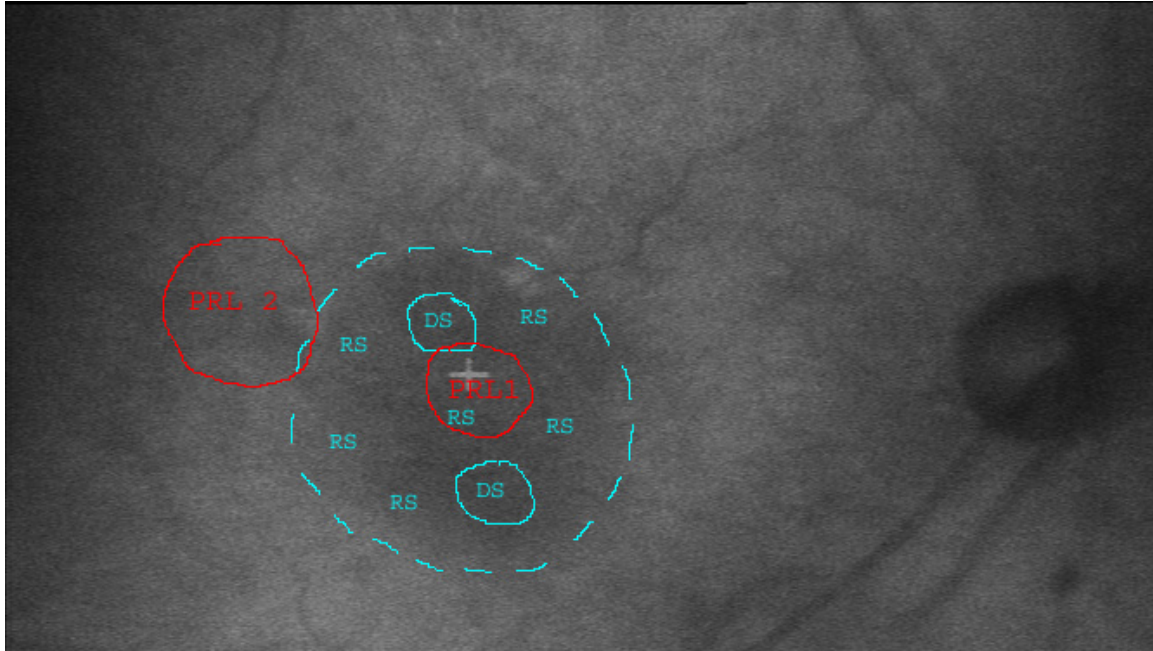


Figure #2

Scotomas Not Following Visible Anatomical Disruptions

DS indicates dense scotoma (50,000 Troland stimuli)

RS indicates relative scotoma (200 Troland, room light level stimuli)

Red Circles indicate areas of Preferred Retinal Loci (PRL)

PRL 1 was used for bright light stimuli and PRL2 was used for dim light stimuli

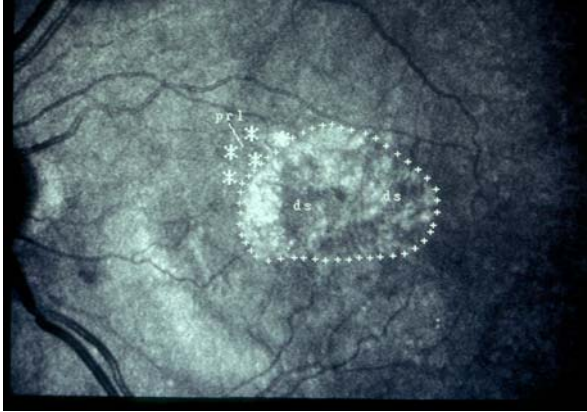


FIGURE #3

(2 slides, picture of SLO macular perimetry and how the patient might view a face)

Figure Caption: This SLO macular perimetry study in a patient with age related macular degeneration shows that the fovea is non functioning, lying within an area of dense scotoma(ds). The patient's visual system has chosen a Preferred Retinal Locus (PRL) that is anatomically superior and nasal to the non functioning fovea and central scarring. (PRL indicated by *s) While using the PRL for fixating the lady's nose, perceptual completion may render the image complete or some metamorphopsia may be noted as indicated to the right of fixation.