

VISUAL FIELD DEFECTS & UNILATERAL SPATIAL INATTENTION: Diagnosis and Treatment

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Abstract

Patients with acquired visual field defects and unilateral spatial inattention are often not exposed to optical systems or visual rehabilitation therapies that can facilitate their recovery. Practitioners may not be aware of the different categories of optical systems available or may not understand that altered visual spatial references or spatial inattention may be the reason for reduced orientation and visual awareness. This article describes the different types of visual defects most commonly acquired through head injury using both anatomical and functional criteria. The use of different prism field compensatory systems and strategies, as well as the practical advantages and disadvantages of each system are reviewed from a clinical perspective. Treatment recommendations and examples of therapeutic techniques are described.

Key Words

behavioral therapies, confrontation testing, hemianopia, quadrantanopia, perimetry, prism, scotoma, unilateral spatial inattention, visual rehabilitation, visual field defects, visual neglect, yoked prism

INTRODUCTION

Visual field defects are common sequelae of acquired brain injury (ABI). They vary from small isolated blind spots, or scotomata, to loss of vision in an entire hemifield, or from an entire eye. The visual consequences of visual field loss are not simply a matter of loss of visual information being gathered from the affected areas in the visual field. The loss may include changes in the entire perceived structure of three dimensional space,¹ as well as shifts in the center of gravity,² which affect balance. Small scotomatas can be extremely disruptive if they are near the area of central fixation, causing disruption of reading and other activities of daily living (ADLs) that require fine visual discrimination, or orderly movement of the eye on a page. Ingrained patterns of eye movements with which one scans the world must often be entirely restructured following visual field loss, in order to effectively gather information from spaces that fall within the blind areas of visual field.^{3,4} Visual perceptual speed and span become critical; more fixations are required to cover the relevant space. Ishiai, et al,³ have shown that patients with hemianopia have different fixation patterns from persons without, in that they fixate longest on targets in the blind field, whereas persons without field defects fixate longest on central targets.

Management of visual field defects covers a broad spectrum of education, vision rehabilitation therapy, and optical aids. Unfortunately, most patients with vi-

sual field defects are not referred for visual rehabilitation. Because of the lack of understanding of the rehabilitation issues involved, many patients with significant visual field loss are told that they will simply adapt to the loss. These patients may end up severely curtailing their activities because they do not feel safe in environmental situations which are not tightly controlled, or they may end up on the road driving, without the benefit of either visual rehabilitation services or a driving evaluation. Every member of the rehabilitation team should be aware that vision rehabilitation is available for these patients, and that it can significantly impact the quality of the overall rehabilitation outcome. These patients should be referred to a visual rehabilitation specialist, such as an optometrist practicing neuro-optometric rehabilitation, for treatment (Appendix).

Neuroanatomy of Visual Field Defects

Visual field defects result from disruption of the primary retino-geniculate-occipital pathway. The neuroanatomy of visual field defects is quite orderly, allowing one to ascertain the general location of the lesion from the pattern of the defect (Figure 1). Unilateral visual field loss results from damage to the retina or optic nerve prior to the optic chiasm. At the optic chiasm, which rests above the sella turcica where the pituitary gland is seated, the ganglion cells from each eye hemidecussate; the axons which originate

rately define how close to fixation the edge of the of field defect extends. The Amsler grid is a small graph paper with a fixation dot in the center, which can be used to detect visual distortions or scotomata within the macular area. See Figure 2. For this testing, the patient fixates the central dot of the Amsler grid at a distance of 40 cm, while the examiner moves a small target such as a pen tip from the unsighted field toward the sighted field. The patient is instructed to report as soon as the pen tip comes into view. This information is critical in determining the sort of interventions that may be required for reading, writing, and various search tasks.

Confrontation Testing

Confrontation testing is a method for determining the most peripheral extent of the intact visual field. The patient is to indicate when he first perceives a target being moved from an area peripheral to the expected visual field extent into the expected range. One must remember that the sensitivity of confrontation testing is poor, ranging in one study from 38% detection of abnormal visual field quadrants to 90% for detection of hemianopias.⁵ Gianutsos and Suchoff⁶ described how confrontation testing is made more sensitive by using a two-person method. One person sits directly in front of the patient to check straight ahead fixation, which is a point on that person's face. The second person stands behind the patient and introduces the stimulus (a finger or small target) randomly from the periphery. The advantage of this method is that there is reduced cuing for the target stimulus. This test should be performed monocularly when possible.

A further use of this testing involves the extinction phenomenon. Here, the patient is aware of single stimulation in both the left and right fields, but is not aware of one field when both are stimulated simultaneously. This phase is particularly useful in determining the existence of Unilateral Spatial Inattention (see below).

Functional testing does not restrict fixation but determines how efficiently different visual field quadrants are attended to. Large interactive electronic boards such as the Dynavision,^a Wayne Saccadic Fixator,^b as well as targets presented on computer screens, as with Gianutsos' visual field software may be used.⁷

Following ABI, various homonymous visual field defects may occur with or without unilateral spatial inattention (USI), which is also known as "neglect". The differential diagnosis or the co-existence of these entities must be made. It is still the case that most general vision care practitioners, when finding a deficit on a perimetric test, will assume that it is due exclusively to a post chiasmal defect. In this type of defect, the primary retina-LGN-occipital pathway is disrupted and there is a loss of visual input. USI is a higher cerebral disorder where the substrates for "sight" are intact, but the ability to attend to and therefore perceive the visual information is disrupted. Further, USI can be found in the presence or in the absence of a post chiasmal defect.⁸ Differential diagnosis of pure USI vs. pure post chiasmal visual field defect, or whether the two entities are co existing is described by Suchoff and Ciuffreda.⁸ USI is a much more dangerous condition than the post chiasmal defects; in USI there is a general unawareness of the loss of information, and an associated lack of compensation.

As with a basic visual field defect, USI presents in different depths or densities. For instance, a person with severe USI may not demonstrate any evidence of sight in the neglected hemifield on automated perimetry or confrontation fields. On the other hand, the USI may be so mild that the patient only demonstrates deficits in visual perception on specialized testing such as line bisection, complex saccadic exercises, or tasks that require simultaneous processing or divided attention.⁸

Patients with defects that originate posterior to the chiasm or retinal based scotomas are usually aware of the field defect; those with USI will most often remain unaware of the defect.

FUNCTIONAL IMPLICATIONS OF VISUAL FIELD DEFECTS

Monocular Defects

Monocular field defects can range from something as small as a macular hole, to loss of the vision of the entire eye in patients with TBI. When a small central area of vision is lost in the non dominant eye, one experiences a loss of fine stereopsis, but does not generally require aggressive rehabilitation intervention. However, if the loss of central visual acuity occurs in the dominant eye, patients

sometimes express that while they are able to see, they have more difficulty comprehending or "thinking" while using the formerly nondominant eye for detailed acuity tasks. The sighting dominant eye is the eye that is used as a primary reference for spatial orientation. While sighting dominance changes with eccentric gaze,⁹ most of our fixations are central, and thus there is a preferred eye for most tasks that require visual-motor responses.¹⁰ The information from the sighting dominant eye is used for planning motor actions. A sudden disruption of this input (through pathology or injury) creates a constant daylong challenge. Persons with one eye enucleated have been shown to undergo a significant shift of their egocenter,¹¹ in addition to the consequent loss of stereopsis. Both of these changes can make simple, formerly automatic ADLs time consuming and difficult.¹² Most patients who become monocular will eventually learn to compensate, but would benefit from rehabilitation techniques that assist them in re-establishing a consistent monocular space world with a stable egocentric reference within it.

Homonymous Defects

Homonymous visual field deficits include sector defects, quadrantanopia, and hemianopia. Homonymous hemianopia (HH) is the most common visual field defect following brain injury.¹³ Patients with this condition may experience frequent running into, or tripping over objects on the blind side. Efficient adaptive scanning patterns are not generally acquired without some direction and practice.¹⁴ Walking through chaotic environments, such as busy public places, poses a safety risk for which the patient must internalize habitual scanning patterns. The family pet may pose a hazard if it walks into the path of the patient from the unseen side. A right HH, without several degrees of macular sparing, causes severe disruption of reading, as the patient lacks the visual information required for preprogramming the eye movement to the next word. Left HH also slows reading and can cause the patient to lose his place as they move from line to line

Patients with HH frequently experience egocentric visual midline shifts—i.e. a shift where the perceived straight ahead is shifted away from physical straight-ahead, causing a mismatch be-

tween their perception and the physical reality.^{15,16} These egocentric visual midline shifts cause veering—typically into the blind field—during mobility and driving. They also disrupt simple visual motor tasks such as eating or picking up a glass, demanding constant small corrective adjustments throughout the day. This can be a fatiguing experience and can leave patients feeling unsettled and unsure of themselves without knowing why.

Superior and Inferior Defects

Superior visual field loss seldom requires rehabilitation, other than education that there are unseen objects in that field. Garage doors, overhead kitchen cabinets that protrude, and low hanging lights are perhaps the most common obstacles here. It is uncommon for these patients to have much difficulty with orientation or safety issues. On the other hand, patients with homonymous inferior visual field loss may be severely impaired in mobility, particularly in an environment where there are children, animals or other objects in the affected field. If the margin of the field deficit is close to the fixation point, they may also have difficulty with losing their place during reading.

Concentric Defects

Patients with concentric visual field loss, may have severely restricted visual fields on visual field testing, which we have found to be caused by selective magnocellular damage during diffuse cerebral hypoxia, edema, or shearing. However, these individuals may not always act as if they have severe visual field loss during mobility.

TREATMENT OF VISUAL FIELD DEFECTS

Monocular Defects

Patients with monocular loss or degradation of central vision in their dominant eye may have difficulty restructuring their egocentric visual midline, and visual space. If they are still having difficulties with ADLs after several weeks they may benefit from therapies to help them restructure their visual space and visual-motor responses. In a situation where both maculas have reduced acuity (low vision) the use of telescope lenses and magnifiers become appropriate. Therapies can include instruction in the use of these devices and eccentric viewing.

HH and USI General Considerations

Considerably more patient education is required in treating USI than in treating HH; the patient with HH, once aware of the defect, will most frequently apply compensations to one degree or another. On the other hand, the patient with USI remains unaware of the defect and consequently perceives her incomplete spatial world as being whole.

It is important to inform the HH patient that the purpose of the optical devices and vision rehabilitation therapy is to help them gain a more complete awareness of their surroundings in the most efficient manner. A clear differentiation between the prognoses for performance improvements versus actual visual field recovery should be made at the very outset before intervention is initiated. This is important in order to create realistic expectations and to avoid a sense of failure over time. Partial recovery of visual field defect, both through spontaneous recovery, and also resulting from vision rehabilitation therapy following spontaneous recovery, are common findings.¹⁷⁻¹⁹ While it is not possible initially to predict which patients will show recovery, the prognosis for improvement is better for those patients who have a gradient of sensitivity at the edge of their visual field loss,^{20,21} as opposed to a sharp edged field cut. Most recovery of visual field is modest, averaging five to seven degrees in hemianopia, and found at the border of the sighted field.²¹⁻²³

The purpose of using optical prism systems in glasses for HH compensation is to shorten the time required to access information from the affected field, and to decrease the time that attention is being diverted from the functional side. When prisms are used to compensate for field loss, the base is always placed in the direction of the field loss. Thus, images of objects in the periphery of the lost field are moved closer to the straight-ahead, or functional side.⁸ The use of prism requires therapy to retrain eye movements and to adjust to the altered perception of space.

Yoked prism

The optical effect of wearing yoked prisms (prisms in front of both eyes with the base in the same direction) in glasses is that there is a shift of the visual image from the hemianopic field into the func-

tional field. The amount of image shift (field enhancement) measured in degrees, is equal to about half the prism diopters employed. The advantage of full field yoked prism for HH compensation is the instant access to the field enhancement with no special eye movements required. The disadvantage is the relatively low amount of prism that can be realistically used to keep spatial distortion at an acceptable level. It would require a relatively high amount of prism to achieve a significant increase in peripheral field using full field yoked prisms. Nevertheless, full field yoked prism can be an effective “early warning system” so that patient can become visually aware of oncoming objects or people in the affected field.⁸

Single Prism

The Peli system^c uses relatively high power prism wedges mounted on the lens of the patient’s glasses, corresponding to the side of the affected visual field above and below the line of sight.²⁴ Since the wedges are placed on just one lens, we consider it as a single prism device. Each wedge has its base placed into the affected field. This produces the simultaneous perception of a central image created from the functional field and a peripheral image shifted over (by the narrow prism wedges) from the hemianopic side. Essentially, it expands the limits of the affected field. The advantage of this system is that the use of higher powered prisms creates wider field awareness while the design allows simultaneous access to both right and left visual field. We have found that the system requires significant patient adaptation.

The Visual Field Awareness SystemTM (VFAS)^{d,25} uses a combination of eye movements into the hemianopic field side and relatively high amount of prism to increase field awareness. A round prism is inserted in a lens and placed just temporal to the pupil or at the temporal limbus on the affected side. The user is taught to look into the prism to gain quick access to information on the hemianopic side. Therapy often uses the analogy of glancing into a side mirror when driving. The advantage of this system is relatively larger field awareness with minimal optical distortion. This system also reinforces scanning into the blind hemifield, which is critical to maintaining adequate spatial constructs in hemianopia. The disadvan-

tage is confusion with the resulting double vision when the patient is viewing through the prism. Szlyk et al²⁶ have found improvement in multiple visual skill categories with use of such peripheral prism systems, as well as continued usage of peripheral prism systems for two or more years by most patients who were dispensed with VFAS-like systems.

All these system options can be demonstrated to the patient using temporary, flexible press-on prisms (i.e., Fresnel prism lenses). The press-on prisms have inferior optics but are inexpensive and versatile.

The application of any of these prism lenses requires some vision rehabilitation therapy in order for the patient to achieve maximum benefit from the lenses. For yoked prism and Peli prism systems, therapy generally incorporates activities that teach the patient to reorient their spatial judgments to the changes caused by the prism. Typically this takes the form of visual-motor matching activities. With peripheral prism systems, the patient is also taught how to fixate quickly into the prism to gain awareness of objects in the hemianopic field, ignoring the double vision that this creates. If something of interest is viewed in the prism, the patient must then turn her head toward the target to view it outside of the prism.

The authors have found the peripheral prism (e.g. VFAS) type of prism system to be the most popular system selected by patients given the choices described above. Ultimately the patient should achieve the ability to move forward while performing automatic fixations into the temporal prism. This is akin to adopting a radar mode of scanning where wide field awareness is gained without necessarily recognition. The repeated fixations into the temporal prism allow rapid access to object awareness in the hemianopic side with minimal loss of attention to the functional field.

Our experience indicates that patients with both USI and HH or those with just marked USI do not typically appreciate the changes as described above. Patients with marked USI are not aware of the need to compensate for the affected side so make little attempt to look into peripheral prisms on the side of the hemianopia.⁸ The visual misperceptions associated with USI result in difficulty integrating the simultaneous peripheral and central visual

information of the Peli type prism. Full field yoked prism glasses do manifest functional improvements in some patients by creating a shift in the image space to better match the shifted perceived egocentric visual midline in USI.

The egocentric visual midline shift, or perception of "straight ahead", tends to be in the opposite direction for patients with USI, vs. those with HH.^{1,15} That is, the perceived visual midline is shifted toward the defect for patients with "pure" hemianopia, and away from the defect for patients with "pure" USI. Interestingly, Ferber and Karnath show evidence that patients with both HH and USI have no shift in their perception of "straight ahead."²⁷ Patients will frequently tend to veer in the direction of their new perceived midline. In both HH and USI, restructuring perceived visual space to create a better visual and visual-motor match between the perceived and actual location of objects in space is critical. This may require the use of lenses, prisms and/or rehabilitation therapy.

Padula¹⁶ describes a quick screening for this egocentric visual midline shift. The patient fixates a vertically held pen, while the examiner passes it across in front of the patient's eyes. The patient is to report when the pen appears centered in front of their nose. The evaluator stands facing the patient, but off center, and introduces the pen first from the functional and then from the affected field side. Patients with visual midline shift describe the pen as appearing centered when in fact it is to the right or left of center depending on the direction of the visual midline shift. An alternative method of comparing the patients "internal" spatial coordinates with external space is the Spatial Localization Board.²⁸ With this instrument, the patient views targets on a board held under his chin, and makes a fast pointing movement with a pen from underneath the board (so that the hand is unseen) to mark the perceived position of the target. This method has the advantage of resulting in a hard copy recording of the spatial distortion. However, the disadvantage is that it requires a motor response, which may in itself be impaired, confounding the test.

Behavioral Therapies

Some patients achieve improved efficiency in compensating for their HH using head turning and scanning techniques that

require minimum demonstration by the rehabilitation professional. A small habitual head turn into the affected field may allow faster access for eye movements into that field, but head turning as a method of scanning is inefficient and counterproductive.¹³ Compensatory oculomotor scanning training is a mainstay of treatment for both HH and USI.^{4,13,22,23} The training has proven to be useful in both improvement of function^{25,26} and sometimes true expansion of residual visual field.¹⁸ We have found that in general, the patient with HH without USI will typically take considerably less training, and will be able to scan into the blind field, on instruction. This training begins in predictable situations, but must be generalized to non-predictable situations.

For visual demands where the margins are static and predictable such as paragraph reading, brightly colored strips held against the text margin on the affected side serve to encourage more complete sentence scanning. Scanning playing card layouts, dotting specific letters in a paragraph and calling out the first and last letter of each line are examples of therapies that can be used to improve page-scanning accuracy. A method for providing feedback on scanning accuracy while reading, is to have the patient trail under the word with a pen as they read. The pen trail provides feedback that the therapist can share with the patient as to what was actually read versus what was missed. Processing speed and span of recognition should be maximized along with the scanning training. Therapy for these perceptual responses employs interactive computer programs along with tachistoscopic presentations.

For non predictive or dynamic situations such as walking or navigating a wheelchair, compensatory techniques cannot be as simple. Classically the patient is taught to turn his head or scan towards the affected side. Unfortunately the patient has a poor concept of how far to turn his head or how far to scan to ensure a complete awareness of the visual surround. Consequently visual information on the affected side is typically missed. Furthermore, there are frequently head-centric components to USI.²⁹ This implies that teaching the patient with USI to head turn, rather than teaching scanning into the affected field with the eyes, can

further embed the USI, rather than remediating it. However, many patients with USI have difficulty when told to move their eyes toward the affected visual field.

Margolis³⁰ has developed and successfully used for many years an alternative approach (Margolis Eye Throwing Technique) which relies on non visual criteria to foster more complete environmental scans. The patient is taught in a step like fashion how to repeatedly and rapidly begin a scan from the most extreme lateral position on the affected side. The patient is taught to “throw” his eyes as far as physically possible into the direction of the affected field. This is initially done with eyelids closed, and then, once the proprioceptive concept has been learned, with them open. Once the patient has learned to begin the eye scan from the extreme temporal position on the affected side, he makes a complete visual sweep towards the functional side. The success of this technique is postulated to be due to the following points: the patient creates his own appropriate and repeatable starting point for a complete scan; the motor act of “throwing” the eyes into the affected field serves as an arousal mechanism to increase attention towards that side;³¹ visual attention tends to follow the visual axis as it moves towards the affected side,³² and visual scanning towards the direction of the functional field is usually intact.³³ At this point the patient can effectively engage in scanning activities in the therapy room, which must then be generalized to real world situations. A programmable pager set to vibrate every few seconds can be worn next to the skin on the patients affected side. This serves as a trigger to remind the patient to carry out the activity in everyday situations. Over time the pager should become unnecessary.

In addition to these scanning techniques there are a variety of therapies designed to rehabilitate concepts of body image, directionality, visual spatial midline and visual spatial relations. One example of a direct feedback method is to require the patient to walk towards a full length mirror while lining up a vertical strip of tape on their midline with a vertical line placed centrally on the mirror. The conscious adjustment required by the patient to achieve the match heightens awareness of their habitual mismatch.

Yoked prisms may also be used in a therapeutic manner to compensate for the mismatch between the physical world and the patient’s inner spatial construct. The prisms are worn during programmed movement activities to create adaptive learning experiences.

Altitudinal and Concentric Visual Field Loss

Patients with superior visual field loss generally adapt well and treatment requires only education regarding awareness of obstacles in superior space. When there is either a concentric or an inferior visual field loss, the primary concern is creating awareness of objects in the inferior visual field so that patients avoid tripping over objects on the floor immediately in front of themselves. Base down yoked prisms have been used with considerable success for this purpose. Also, for concentric visual field loss, to increase overall perspective for orientation and awareness, a reverse bioptic spotting telescope (miniature telescope mounted above the eye) may be inserted in the patient’s glasses. The reverse telescope projects a miniaturized view of a wide area of visual field into the patient’s functional vision. The patient is taught how to repeatedly fixate into the telescope to gain access to peripheral visual information.

Blindsight

Blindsight refers to the phenomenon whereby individuals are able to respond to visual stimuli within the area of their visual field defect. Usually the response takes the form of the detection, localization, or form perception of the object when it is presented within the visual field defect. Notwithstanding these responses, the patients report that they are unable to see any stimulus within the field defect area. Although they may be able to state whether a target in the HH field is present or absent, localize the object when told to point to it, and perhaps even identify the form of the object, they do not consciously perceive it, and will deny that they can see it. There is still controversy whether blindsight results from residual connections in a damaged visual cortex, or from intact extra-striate pathways.³⁴ Attempts to elicit blindsight responses usually take the form of a forced choice paradigm, or making the patient point toward the “unseen” target. In cases of inferior visual field defect or complete HH, the vision re-

habilitation practitioner may attempt to elicit and train blindsight responses³⁵ which can then be used to avoid obstacles, even though they are not consciously seen.

Driving Rehabilitation

Both sensitive evaluation techniques along with experienced clinicians are required to determine whether an individual has the simultaneous processing skills necessary to drive safely while compensating for a field defect. Any trace of USI can be reason to deny driving privileges. Prior to returning to driving, patients with significant HH visual field deficits should have an adaptive driving evaluation (Appendix). We have found that many patients with HH field defects without USI have been driving safely for years—some with and others without prism systems.

Summary

Visual field deficits can cause severe loss of function, spatial disorientation, and significant personal safety concerns following brain injury. Patients with visual field deficits should be referred to a vision rehabilitation specialist for diagnosis and rehabilitation. Pure HH must be carefully differentiated from pure USI, as the appropriate education and therapeutic plan are dependent on this differential diagnosis. Rehabilitation for visual field loss includes use of lenses, prisms, and low vision devices, as well as vision rehabilitation therapy and compensatory strategies. These rehabilitative treatments are primarily aimed at reestablishing a stable visual spatial construct which matches the physical world. Patients with significant visual field deficits who are given the opportunity to take advantage of the options most appropriate for them have the best prognosis for achieving personal independence and safety goals.

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Sources

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Markam, Ontario, Canada

- b. Wayne Engineering
8242 N. Christina Ave, Skoke, Illinois 60076
- c. Chadwick Optical, Inc.
PO Box 485, White River Junction, VT 05001
- d. Gottlieb Vision Group
5462 Memorial Dr., #101, Stone Mountain,
GA 30085

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Appendix Resources for Accessing Post-brain-injury Vision Rehabilitation Professionals

The following websites provide geographic listings of potential visual rehabilitation practitioners.

Neuro-Optometric Rehabilitation Association, International	www.nora.cc
College of Optometrists in Vision Development	www.covd.org
Optometric Extension Program	www.oep.org

Adaptive driving evaluations and training can be obtained from members of
The Association for Driver Rehabilitation Specialists www.driver-ed.org

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