PERIPHERAL VISUAL FIELD LOSS & VISUAL NEGLECT

DIAGNOSIS & TREATMENT

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Abstract
Diagnostic and therapeutic approaches are reviewed for the management of peripheral visual field loss in those with acquired brain injury. Differentiation of field loss from visual neglect (aka visual unilateral spatial inattention) is a key component in diagnosis and treatment. Since the emphasis is on compensation, visual field loss requires less vision rehabilitation, although modest visual field recovery is a relatively common finding. Visual neglect requires considerably more rehabilitation, but it is more likely than visual field loss to be remediable.

Key Words
brain injury, hemianopia, perimetry, unilateral spatial inattention, visual field, visual neglect

INTRODUCTION
Visual field diagnosis and treatment must be a component of any brain injury rehabilitation program, as visual field loss is common following brain injury. The primary visual pathway, which is the neural substrate for visual field loss, extends from the retina to the occipital cortex. Thus, the pathway is long and frequently damaged in cases of significant brain injury1. The ganglion cells, emerging from each retina and forming the optic nerves, hemidecussate at the chiasm, so that images from the right side of space (imaged on the left half of the retina with the vertical divide at the fovea) end up neurally represented in the left cortical hemisphere, and images from the left side of space end up neurally represented in the right cortical hemisphere. The retinotopic mapping, or organization, of the neurons in this pathway is maintained well enough that the clinician can localize the lesion causing the visual field deficit reasonably well by the characteristics of the field deficit itself (Figure 1).

In the clinical setting, however, the lines are frequently less cleanly drawn; the ischemia and/or neural shearing that occurs with many hypoxic and traumatic brain injuries can cause visual field defects that do not match the traditional pathway analysis presented in Figure 1. Examples would be scattered islands of visual field loss or concentric visual field loss found in severe hypoxic or ischemic brain injury, and altitudinal (superior or inferior) visual field loss found with anterior ischemic optic neuropathy (AION). Visual field loss due to AION may occur without primary brain injury if there is significant blood loss due to trauma or surgical intervention, without prompt replacement.

Visual Field Testing and Diagnosis
Visual Field Defects
The standards in visual field testing are Goldmann kinetic testing and automated threshold-based perimetry. However, in the brain injury population, where attention, fatigue, postural difficulties, and limited response capability are significant constraints, patients are frequently unable to perform either a full Goldmann or automated perimetric evaluation. Hence, other techniques may be necessary to obtain this important information. The tangent screen is useful for some patients. It is particularly good for demonstrating small scattered scotomas, but it requires extended divided attention. I have found that an older instrument, which is no longer manufactured, the Harrington Flocks, is extremely useful in testing the brain injury population. It is a tachistoscopic central visual field test, which allows the patient as much time as needed to respond either by counting or pointing to the test dot stimuli that were presented. The Amsler grid may also be used, not only for delineating macular defects, but more importantly in this population as a miniature tangent screen for determining the boundary of a hemianopic defect. Knowing the degree, if any, of macular sparing is critical in determining what sort of compensations should be applied, particularly for reading. Lastly, the differential diagnosis of a visual field defect from visual neglect (aka visual unilateral spatial inattention) is a critical part of the visual field diagnosis.

Visual Neglect
The distinction between a visual field defect and visual neglect (aka unilateral visual spatial inattention) is an important clinical consideration. Visual neglect, like visual field defect, comes in varying
densities. A dense visual neglect will appear as a hemianopia. However, in visual neglect, the primary visual pathway (i.e., the substrate for sight) is intact. It is the ability to attend to, and therefore perceive and respond to, the stimulus that has been damaged. Neglect may involve separately, or concurrently, the patient’s own body (i.e., personal neglect), space within arms reach (i.e., peripersonal visual neglect), or space beyond arms reach (i.e., distant visual neglect). Object perception may be involved such that only half of objects are perceived, or the objects may be intact, and there may be inattention to areas of space—usually a hemifield. There may also be motor involvement on the same side of the body, which may appear as a hemiparesis. Thus, it is important, in diagnosing and treating visual neglect, to understand that neglect is not a unitary disorder, but rather a syndrome that involves one or more of multiple neural substrates with multiple manifestations (reviewed by Bartolomeo and Chokron).

Proposed Neural Substrates of Neglect

There have been a number of cerebral structures that have been associated with visual neglect. Portions of the dorsolateral frontal lobes, the inferior parietal lobule in the posterior parietal cortex, the temporo-parietal junction, the caudate portion of the superior temporal gyrus, the cingulate gyrus of the limbic system, as well as subcortical areas such as the caudate nucleus and putamen in the basal ganglia, and the pulvinar in the thalamus, are all neural substrates that have been implicated in visual neglect. Each of these is a substrate, or part of a network for visual attention. Additionally, damage to the white matter pathways connecting parietal lobe to frontal lobe—a portion of the superior occipitofrontal fasciculus—can cause visual neglect. Each of these neural substrates, damage to the posterior parietal lobe, superior temporal lobe, or the superior occipitofrontal fasciculus appears to be the most devastating. As demonstrated by split brain research, the left hemisphere allocates attention only to the right visual hemifield, while the right hemisphere allocates attention to both right and left visual fields (Figure 2). This finding in split-brain patients implies that right hemisphere’s allocation of attention to the right visual field is probably mediated through subcortical substrates. Thus, when the left hemisphere is damaged, there is an imbalance in attention between the left and right visual fields, with the right being less attended. However, both hemifields still have attention allocated to them by the right hemisphere. But when the right hemisphere is damaged, there is no cortical substrate for attending to the left hemifield. This results in a dense vi-


Figure 2. Allocation of spatial attention by the cortical hemispheres. The right hemisphere allocates spatial attention to both sides of space, while the left hemisphere allocates attention only to the right side of space. While damage to the left parietal lobe may disturb the balance of attention across visual space, damage to the right parietal lobe can leave the left side of space entirely unattended. With permission from Moore, J.C. and Warren, M. Effect of visual impairment on postural and motor control following adult brain injury. Continuing education workbook by visABILITIES Rehab Services, Inc. (www.visabilities.com).
visual neglect that mimics homonymous hemianopia on visual field testing.

Differential Diagnosis of Neglect

Differential diagnosis of visual neglect begins with a careful history from the caregivers and therapists. Often the occupational therapist will be aware and report that the patient seems to ignore the lower left quadrant or other area of space, or a family member will be able to describe neglect-like signs in the patient’s activities. In addition, there may be an accompanying motor neglect, which may appear as a hemiparesis, except that sometimes the limbs in question may move without volition on the part of the patient.

Double simultaneous stimulus confrontation testing to demonstrate the phenomenon of extinction is a method that can sometimes be used to demonstrate visual neglect. In visual neglect, there is competition between the inputs from the two hemifields. With a moderate visual neglect, one may find full counting fingers confrontation fields if stimuli are presented in only one visual field at a time. However, when simultaneously flashing fingers in opposite fields, the perception of the fingers in the less attended field is extinguished, so that the patient responds as if only the fingers in the well-attended field were presented. This generally indicates visual neglect. However, patients may have visual neglect without clinically noticeable extinction (reviewed by Vuilleumier and Rafal). In my experience, patients with very dense visual neglect will respond to unilateral confrontation as if they are hemianopic, limiting the usefulness of this test, and those with mild neglect may demonstrate visual neglect on line bisection or cancellation tasks, but not double simultaneous confrontations. Nonetheless, double simultaneous stimulus confrontation testing is a useful tool that is quickly and easily administered.

Cancellation (Figure 3a) and drawing tasks (Figure 3b) may also be used to demonstrate visual neglect. Cancellation tasks may also be used to demonstrate the phenomenon of extinction. When asked to cross out all of the stimuli, the patient with visual neglect may fail to cancel targets on the left half of the figure or page. However, if asked to erase the targets, the patient will frequently be able to complete more of the task, as the competing stimuli are being removed as they proceed.

Another task that allows one to investigate the presence of visual neglect is the line bisection task. This task demonstrates the spatial distortions caused by the visual neglect or visual field deficit. For example, if one presents a line 5 cm or longer at a distance of 40 cm from the patient’s eyes, and then asks them to place a mark that bisects the line, the patient with visual neglect will fail to perceive the neglect, or unattended end of the line, and so will bisect toward the intact field. In contrast, the patient with a visual field deficit will most often bisect in the opposite direction (i.e., toward the field defect). The patient with both visual neglect and hemianopia, will therefore tend to bisect lines accurately (reviewed by Kerkhoff et al). However, at times, rather than bisecting correctly, patients with both hemianopia and visual neglect have been found to make even greater bisection errors away from the affected field relative to those with neglect alone.

A quick screener for visual neglect which can be performed by most patients is the line bisection-crossout task that I developed for use in my office (Figure 4). This task must be interpreted in light of the visual field results (Figure 5). Line bisection and crossout tasks appear to reflect different aspects of neglect, and like double simultaneous confrontations, a line bisection test without a crossout task can be relatively insensitive. Occasionally, one will see a hemianopic pattern on the visual field test, with a centered line bisection, and will not be able to elicit functional evidence of visual neglect. More often, one may see patterns of line bisection where the “bisection” is shifted to the right or the left, but one is unable to elicit other evidence of visual neglect. Although the literature has interpreted an off-center line bisection as evidence of visual neglect, it just demonstrates a distortion in spatial perception. Indeed, in the normal population, there is a slight bias leftward from center. So one should not measure with a ruler...
when judging the bisection, as the bias should be evident by gross visual inspection alone. If a line bisection crossout task and visual field pattern are found that imply visual neglect, then one must test and observe carefully for functional visual neglect. More sensitive tests such as the Star Cancellation subtest from the Behavioral Inattention Test can be used to confirm the diagnosis, if necessary.
Another test which has demonstrated good sensitivity by combining the concept of line bisection with a crossout-like task is the Character Line Bisection Test developed by Lee et al. Since it has distractors built into the line, this appears to be a sensitive test, if the patient is able to complete it. A line of different characters is presented to the patient. There is a specific target character, which is never presented in the exact middle of the line. The patient’s task is to circle the target character that is closest to the center. Twelve repetitions are performed with different character sets.

A drawing task may further elicit visual neglect. If functional visual neglect cannot be observed or elicited, then one should treat the spatial perceptual distortion observed in the line bisection crossout task if possible.

**Visual Field Loss Treatment Therapy**

Treatment of visual field loss begins with education of the patient and, if applicable, the patient’s family or caregivers regarding the physical and behavioral aspects of the loss. Next, compensatory techniques are taught. Scanning is the mainstay of visual field loss treatment. When scanning into the blind field, the patient with hemianopia loses information from the normally-sighted side. Therefore, they must be able to scan, gather information, and move back to the other side quickly. Perceptual speed and span must be maximized. As demonstrated on a line bisection task, the hemianopic patient has visual-spatial distortions in perception. These are generally remediable with yoked prism procedures and other vision therapy techniques involving space perception.

Scanning strategies must be implemented and generalized to mobility and other daily activities. With consistent scanning into the blind field, some amount of visual restitution has been observed in patients with visual field loss. These gains in visual field tend to be modest, but may be significant in terms of quality of life.

Computerized instruments and programs have been developed for restitution of the visual field. However, thus far, there is no compelling evidence that they work any better than scanning training in the environment. Margolis and Suter discuss therapeutic techniques for peripheral visual field loss and visual neglect in more detail elsewhere.

**Compensatory Prism Systems**

There are two main types of prism systems to assist in compensation for peripheral visual field loss. The Peli “field expansion” prism system is a simultaneous perception system placed above and/or below primary gaze (Figure 6). The prisms are approximately 40 prism diopters in magnitude and directed with the base into the visual field deficit, thus effectively shifting the images of objects from the blind periphery into the sighted field. This allows the patient to monitor with their superior or inferior peripheral vision for objects of interest in the blind field, so that they can scan over to them when necessary. The disadvantage of this system is that if it is applied inferiorly to help the patient avoid obstacles during mobility, it can also cause confusion and obstruction when the patient looks downward for curbs or steps. The Peli system also requires considerable training and practice. In hemianopia, or visual neglect, this system may work best to apply only the superior prism for approximately two weeks to allow the patient to learn the system. The inferior prism can then be added. The advantage of this system is that it may be useful for patients with visual neglect to remind them that the unattended field exists.

The second prism system, the Rekindle Visual Awareness System, is a peripheral prism button fused into the lens in the blind field; it is unseen unless the patient scans into it (Figure 7). The prism is ground with the patient’s refractive prescription, and it is usually 18.5 prism diopters with the base directed into the visual field deficit. The advantages of this system are a) that it does not interfere with...
the patient’s vision until they scan into the blind field, and b) that the system requires scanning. Scanning into the blind field may be crucial to visual field restitution. Because this system requires scanning and furthermore serves as a reminder to the patient to do so, it is my first choice in a prism system for the patient with hemianopia. However, the disadvantage of this system is that it cannot be used in visual neglect, as the patient will not scan into the lens, as they will be unaware of it.

Visual Neglect Treatment

Much of the treatment for visual neglect is similar to the treatment for visual field loss. However, the patient with visual neglect requires considerably more vision rehabilitation therapy than the patient with visual field loss. They initially find it difficult to remember and act on the fact that they are not “seeing” a portion of their visual space, so constant cuing is required. Unlike the patient having visual field loss without visual neglect, the patient with visual neglect does not automatically implement compensatory strategies. Usually, over time, with consistent cuing to the unattended field, they can develop a cognitive realization that they are missing visual information on one side. Visual neglect treatment also requires more therapy to reestablish appropriate form perception and space perception. For example, one adjunct therapy that seems promising to help patients reorganize their space world is to adapt patients to high amounts of prism and furthermore serve as a reminder to the unattended field, they can develop a quad cane, without further physical therapy. The effects of visual neglect are pervasive. Rehabilitation dollars and efforts are both wasted when the visual aspect of unilateral spatial inattention is not diagnosed and effectively addressed as one of the primary rehabilitative issues following brain injury.

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Sources


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