ABSTRACT

Background: Visual field loss is a common result of cerebrovascular accidents (CVAs), with the location of the vascular incident correlating with the location and extent of visual field loss. Many CVA patients also experience hemi-spatial visual neglect, lacking attention to one side of space. Those suffering from these conditions may find simple tasks such as reading, self-grooming, and navigating common areas to range from difficult to nearly impossible. Although CVAs are more common in the elderly population, the following presents a case of an infant suffering a CVA shortly after birth. This provides a unique case of a patient facing a diverse set of challenges while in a highly plastic stage of neural development.

Case: An 11-month-old female born prematurely with a history of CVA presented for a visual function consult. Her father expressed concern for useful vision in her left eye and was seeking further evaluation and therapeutic options. Examination revealed inattention to the left visual field with no fixation preference toward either eye. Ocular motility was limited in upgaze, but the patient had full motility in both left and right gazes. There was a positive OKN response to 1.6 cycles/deg (~20/400 Snellen equivalent) in each eye.

Discussion: An infant suffering field loss faces different challenges than an adult. Techniques to evaluate and treat the patient may be adapted to maximize the patient’s visual performance.

Keywords: cerebrovascular accident, field loss, neglect, prism, vision therapy

Introduction

Visual field loss and visual neglect are common findings in patients who experience a cerebrovascular accident (CVA).\(^1\)\(^2\) Visual field loss results from disruption of the visual signal being passed along the visual pathway. The extent of vision loss depends on the location of the CVA along the pathway. The majority of CVAs affect vision along the optic radiations between the lateral geniculate nucleus and the occipital cortex. A disruption of Meyer’s loop, the portion of the optic radiations traversing the temporal lobe, will cause an incongruent asymmetric superior quadrantanopia of the contralateral visual field, and a disruption of the neural fibers traversing the parietal lobe will cause an incongruent inferior quadrantanopia of the contralateral visual field. Likewise, a stroke occurring posteriorly (closer to the occipital cortex) will typically cause a more complete homonymous hemianopsia in the visual field contralateral to the side of the side of the lesion.\(^3\)

Such field loss is often detected through Goldmann perimetry, static perimetry, or visually evoked potentials. In Goldmann perimetry, a target of fixed size (standard 0.43°) and supra-threshold brightness is moved along a neutral background from the patient’s non-seeing to
the patient’s seeing field. Variation in patient fixation is controlled for by using the examiner’s eye as the patient’s fixation target. Goldmann perimetry is considered a test of the patient’s full visual field.\textsuperscript{4} The most common test using static perimetry is the Zeiss Humphrey Visual Field Analyzer (Zeiss, Carlsbad, California). The standard test used for detecting neuro-ophthalmic visual field defects is the 30-2, which analyzes test points in a 30-degree radius from the point of fixation. The patient is presented with a series of both supra- and infra-threshold stimuli at the various test points. Reliability is determined by the frequency of times a patient correctly responds to a supra-threshold stimulus and correctly does not respond to either no stimulus or an infra-threshold stimulus. Fixation is monitored by presenting stimuli in the physiologic blind spot and by measuring the stability of the corneal reflex through gaze tracking.\textsuperscript{5} Visual evoked potentials evaluate the integrity of the visual pathway from the retina to the cortex. The patient fixates a central target, and a stimulus (often a checkered grid) is presented in the area of the visual field that is being tested. The amplitude and latency of the electrical response is then measured. The advantage of this test over perimetry is that it requires no subjective response from the patient, but it is technically more difficult to perform, and poor fixation must be accounted for.\textsuperscript{6,7}

When hemianopic field loss is detected, further testing must be performed to determine whether it is true field loss or the result of hemi-spatial neglect. While field loss is caused by disruption to the visual pathway, hemi-spatial neglect occurs with damage to the parietal lobe. In this case, the patient is inattentive to all visual stimuli to one side of their midline. If neglect is not present, patients with visual field loss will be able to direct their eyes towards their non-seeing area, but a patient with neglect is unaware that this area of inattention exists.\textsuperscript{8}

The most common tests for neglect include line bisection and line/star/letter cancellation tests. For line bisection, the patient is presented with three horizontal lines perpendicular to their midline. They are asked to draw a line at the exact midpoint of each of the lines. A patient with neglect will draw the line significantly towards their area of usable vision. For cancellation tests, the patient is presented with an array of multiple lines, stars of various sizes, or random letters. The patient is asked to draw a line through each of the printed lines, circle all of the stars of a certain size, or circle one or two specific letters. A patient with neglect will miss all of the targets presented in their field of inattention.\textsuperscript{9}

Treatment for hemianopic field loss and hemi-spatial neglect is targeted at making the patient aware of their non-seeing area and maximizing their usable vision. The two most common forms of treatment include prismatic spectacle correction and vision therapy. Prism may be applied either to expand the visual field or to raise awareness of the visual field loss. Dr. Eli Peli has proposed a method for visual field expansion using sectoral application of Fresnel prism. Base out prism is applied to the superior and inferior portion of the lens over the eye ipsilateral to the visual field defect. This method induces diplopia in this eye but allows the patient to see areas of the defective visual field through the prism. It may be compared functionally to the rear view mirror of a car. Base out prism may also be placed over the entire temporal portion of the lens ipsilateral to the visual field defect, but this method would be ineffective for those suffering neglect, as the patient must be aware of the defect and voluntarily direct their eyes towards the area of prism to take advantage of its effect on field expansion.\textsuperscript{10} Finally, yoked prism may be prescribed. Since the prism is yoked, there is no actual expansion of the field, but rather the entire visual field is shifted. In the case of hemianopia without neglect, the prism is
applied with the base towards the defect to shift the non-seeing area into the patient’s field of view. This shift is intended to make the patient aware of the missing field. If the patient is experiencing neglect, one may consider applying prism with the base away from the area of neglect to encourage the patient to become attentive to the area of their neglect. Vision therapy seeks to promote patient awareness of the visual field defect and to encourage frequent scanning into the area of non-seeing. The end goal is to make these scanning techniques automatic as the patient navigates their environment. Initial therapy often requires auditory cues and external encouragement. It may include repetitively having patients direct their eyes toward a target in their non-seeing field. Similarly, a therapist will have the patient repetitively turn their head towards the area of the defect. Therapy begins in simple environments and builds complexity as the patient improves and is able to process more information. Tasks such as walking are then added in order to make the scanning and head turns more automatic. Weeks to months of therapy are often required, but successful treatment allows the patient to return to many activities of daily living and to resume normal function.

**Case Report**

**History**

An 11-month-old female patient presented for a complete eye examination. A twin, she was born prematurely at 26 weeks’ gestation weighing 1 lb 13 oz (822 g) at birth. The patient was on oxygen for 3 months after birth and was found to have stage 3 retinopathy of prematurity in each eye. An intracranial cyst was removed at age 4 months, and the patient suffered a post-surgical CVA. An abnormal eye posture was noted post-surgically, and the patient was put on phenobarbital for a suspected seizure condition.

**Examination Findings**

The patient presented wearing a habitual spectacle prescription of OD: -0.25-2.50x148, OS: +0.50-1.75x147. A visual evoked potential (VEP) was attempted to evaluate the patient’s visual system, but the patient was uncooperative. To further assess visual function, a series of flashing targets in a dark room was presented to the patient, beginning with a 5-inch-diameter ball and decreasing in size to a 1-inch mechanical toy. The patient was inattentive to any target presented in the left visual field but was full to fix and follow in each eye when targets were presented from the right side. Optokinetic nystagmus (OKN) elicited by a 1.6 cycle per degree OKN drum was symmetric in the right eye but demonstrated temporal-to-nasal asymmetry in the left eye.

Hirschberg and Bruckner testing was within normal limits, and a cover test using a penlight target revealed 8 esophoria at near. The patient showed no fixation preference with alternate occlusion. Both eyes favored the right-sided posture, and there was a slight aversion to superior gaze. Mohindra retinoscopy over the patient’s habitual spectacle prescription revealed +2.50 DS in the right eye and +1.50 DS in the left eye. Pupils were equal, round, and reactive to light. External features and the anterior segment were unremarkable. Evaluation of the posterior segment was deferred to a future visit with the pediatric ophthalmologist. The patient was scheduled for a one-month follow-up to reattempt VEP and confirm testing. This visit will occur after the time of this writing.

**Discussion**

The occurrence of a CVA in an infant produces unique challenges in diagnosis and treatment of the infant’s visual function. As an infant, the patient is unable to perform formal visual field and acuity testing; thus, the clinician must rely heavily on objective findings and the patient’s interaction with her environment. The evaluation is intended to assess the current
state of the infant’s visual system and detect any hindrance towards normal development. A main concern with visual development is equal cortical input from each eye. All testing suggested that the patient was, in fact, using both eyes. The patient showed no occlusion preference, and no tropia was detected on cover test, Bruckner, or Hirschberg. Retinoscopy also confirmed that the near refractive state of each eye was relatively equal when using correction. The visual evoked potential will be repeated at the patient’s one-month follow-up to further confirm equal use of each eye.

Although both eyes were able to fix and follow, the patient was inattentive to any stimulus presented in her left visual field. She was able to track a target to the left, but this task required significant reinforcement. Although hemi-spatial neglect cannot be fully confirmed, certain aspects of the exam were suggestive that neglect was present. The temporal-to-nasal asymmetry in OKN testing was consistent with damage to the right parietal lobe, which is the most common cause of left-sided neglect. Furthermore, the postural shift of her eyes towards the right may be indicative of abnormal egocentric localization, otherwise called a midline shift. Her inattention to the left side may have caused her to perceive her midline to be located to her right; however, her left hemiparesis may suggest that there was a motor component to the abnormal eye posture.

Homonymous hemianopia has been shown to have a negative effect on normal visual development. A recent study of 45 children with the average age of 2.1 years examined the effects of homonymous hemianopia on visual acuity and ocular alignment. The authors noted mild to severe visual impairment in 50% of the subjects and strabismus in 62% of the subjects. Additionally, 53% of the subjects developed an abnormal head posture.

Given the strong connection between visuomotor development and sensory development, it is pertinent to target visuomotor skills when treating the patient’s visual field deficits. Her father was instructed to present objects such as food and toys always from her left side. This will increase her awareness of her left field and prompt her to initiate ocular movements and reaching towards the left side. He was also instructed to turn her head to the right while she maintained fixation on a target. This method takes advantage of the vestibulo-ocular reflex (VOR) and causes her eyes to initiate movement towards left gaze. The technique known as “spinning” was also recommended to promote eye movements. In this procedure, the father will hold the child in his arms and spin in circles while she is looking outward. This method also takes advantage of the VOR. After spinning for a period of time, the eyes will respond to the deceleration effect that is produced once the spinning is stopped. The after effect produces a jerk nystagmus which may be utilized to train the brain to initiate movements toward the direction of field loss.

As the child develops, her visual system must be monitored closely to maximize her visual development. When she matures, therapy may become more complex to reinforce proper development of visual perception and visuomotor integration. Furthermore, therapy may permit development of binocular function and stereopsis, allowing proper alignment and few debilitating visual symptoms.

Conclusion

In conclusion, the occurrence of CVA in an infant presents a unique case in a highly neuroplastic individual. Unlike with the more common older CVA patient, this patient’s visual system had not fully developed prior to the event. While the CVA may hinder proper visual development, the patient may adapt well to the situation. A proactive approach likely involving prism and vision therapy is indicated to maximize the infant’s visual function and visuomotor integration throughout the developmental process.
References


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