Understanding Cortical Visual Impairment in Children

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This article presents a review of the literature and a case study on a child with cortical visual impairment. The literature review covers the diagnosis, etiology, prevalence, prognosis, and a comparison of the differences between children with cortical visual impairment and those with ocular impairment. The case study presents occupational therapy intervention strategies specific to cortical visual impairment and results of treatment.

Etiology of Cortical Visual Impairment

It is speculated (Groenveld, 1990) that much of what is learned in the early developmental years is reinforced visually. Vision is the sense that enables humans to perceive simultaneously an object in its complete form and the object's relationship to other objects in the environment (Groenveld, 1990). Understanding visual impairment in children can be complicated. Traditionally, visual impairment has been thought of as an ocular disorder (Groenveld, Jan, & Leader, 1990). However, visual impairment can be a result of an ocular disorder or a cortical disorder. An ocular impairment refers to eye conditions or to lesions of the anterior visual pathway (from the retina to the lateral geniculate body). Ocular impairments negatively affect visual acuity. Cortical visual impairment refers to lesions in the posterior visual pathway (from the lateral geniculate body to the visual cortex) and represents difficulty in processing and interpreting visual information in the visual cortex (Jan & Groenveld, 1993).

CVI is often referred to as cortical blindness, which is defined clinically as a bilateral loss of vision, with normal pupillary response and an eye examination that indicates no other abnormalities (Whiting, Jan, Wong, Ferrell, & McCormick, 1985). According to Good et al. (1994), the term cortical blindness implies no sensory visual responsiveness. Because most children with a lesion in the visual pathways or visual cortex have some residual vision, the term CVI is a more appropriate diagnosis.

Because CVI is a new area of study, the visual impairments of children with CVI have often been ignored by...
physicians, educators, and other professionals (Jan & Wong, 1991) and many standard textbooks on ophthalmology and neurology do not present information on CVI. According to Groenveld et al. (1990), CVI has been incorrectly perceived to be "nothing more than a fancy term for brain damage, although the majority of brain-damaged children are not visually impaired." The diagnosis of CVI has become more common in the past 5 years and can now be considered one of the major causes of visual impairment in children in the developed countries (Good et al., 1994). According to Jan and Wong (1991), approximately 50% of all children with congenital ocular abnormalities present with additional disabilities, whereas all children with CVI present with associated neurological problems.

There are several causes of CVI, but the most probable cause in children is perinatal hypoxia-ischemia (Flodmark, Jan, & Wong, 1990). The posterior visual pathways and the visual cortex seem to be most vulnerable to damage during the perinatal phase (Good et al., 1994). Although the infant brain becomes more resistant to hypoxic-ischemic episodes as it develops, CVI can still result and has been reported in children after cardiac arrest and open heart surgery. Other causes of CVI include trauma, epilepsy, cerebral angiography, infections, drugs or poisons, and metabolic diseases (Good et al., 1994).

The diagnosis of CVI can be made through several medical tests that also can offer information in planning intervention strategies. However, these tests are expensive and difficult to access, and some are invasive, so it may be better to understand and interpret the visual behaviors of the child and then decide whether these tests can offer essential information (Jan & Groenveld, 1993). Electroencephalography (EEG) provides general information about geniculocalcarine dysfunction and occipital responses to photic stimulation. Visual evoked potential (VEP) assesses the function of the visual pathways, although results vary with this test. Imaging techniques such as computed tomography (CT), magnetic resonance imaging (MRI), and positron emission tomography (PET) are also helpful in identifying where damage has occurred in the brain. Visual evoked potential mapping (VEPM) is a recent technique used for studying CVI; it examines larger areas of the brain in a dynamic manner rather than just measuring the electrical activity over the visual cortex (Good et al., 1994).

Comparison of Visual Responses in the Child With CVI to Those of the Child With Ocular Impairment

The eyes of the child with CVI generally look normal, whereas the eyes of the child with an ocular impairment often look abnormal. Sensory nystagmus is associated with bilateral ocular lesions. These eye oscillations can be slowly drifting, aimless movements or jerky movements. Sensory nystagmus is a symptom of ocular impairment and indicates instability of fixation. In comparison, sensory nystagmus is absent in pure CVI, except when it is accompanied by an ocular impairment. Children with CVI sometimes demonstrate a motor nystagmus, which is an unsteady, tremulous fixation of gaze that is due to impaired brain control and not an ocular impairment (Jan & Groenveld, 1993).

Children with ocular disorders and some vision may have a normal attention span and maintain fixation, whereas children with CVI usually demonstrate a very short attention span (Jan & Groenveld, 1993). They often make a characteristic head turn when looking at or reaching for an object. They do not appear to look at the object with central vision, but instead seem to use peripheral vision (Good et al., 1994; Jan & Groenveld, 1993). Children with ocular disorders that include peripheral field loss are able to use their heads to scan the environment; children with CVI that includes visual field loss are not able to do this. They generally turn their heads and look away from the side of the field loss (Jan & Groenveld, 1993).

Although it is common for both children with ocular disorders and children with CVI to lean close to an object or to bring an object very close to the eyes, they do so for different reasons. Ocular impairments often cause the child to need additional magnification, which is achieved through bringing the object closer to the eyes. In CVI, bringing the object close to the eyes is done to reduce the "crowding effect" (Jan & Groenveld, 1993, p. 103). Groenveld et al. (1990) stated that CVI causes difficulty in seeing objects or pictures placed close together, but the pictures or objects are more readily identifiable when they are spaced farther apart. They claimed that by bringing the object close to the visual field, the child fills the visual field with one object and therefore reduces the amount of nonessential visual information from the background. This allows the child to better process the simpler visual information (Groenveld et al., 1990).

Children with CVI often exhibit compulsive light gazing (staring at a source of light), whereas children with only ocular impairments do not (Jan, Groenveld, & Sykanda, 1990). Jan and Groenveld (1993) also noted that eye pressing, which is prevalent in children with ocular disorders that have retinal involvement, is never seen in CVI.

Color perception in children with CVI is generally intact because color perception has bilateral hemisphere representation and requires fewer functioning neurons than does form perception. Children with CVI are particularly attracted to bright colors such as red and yellow. Sensitivity to light is found in certain ocular conditions and is also found in about one third of children with CVI (Jan & Groenveld, 1993).

The prognosis for improvement in children with CVI depends on cause, age of onset, and severity and type of
damage. Most children with CVI show some improvement in visual responses (Good et al., 1994). To optimize visual responses in these children, modifying the visual environment is critical. Traditional methods of maximizing residual vision in children with ocular impairments rely on enriching and enhancing the visual environment. These methods of intervention may not help children with CVI because as the amount of visual input is increased, visual processing becomes more difficult (Groenveld et al., 1990). Techniques that use simplification of the visual environment, high contrast color, common objects, the integration of touch and vision, and computer technology are stressed for therapists working with children with CVI. It is important to individualize these programs and monitor visual responses during intervention because visual responses are variable in children with CVI (Good et al., 1994).

Case Study

History

J. is a 14-month-old girl, the larger of fraternal twins born near term as a result of an uneventful pregnancy, labor, and delivery. The placentas of the twins were fused on delivery. Seizures and apnea developed in J. 2 weeks after delivery. The seizures lasted for 3 days and required hospitalization. Another brief episode of seizures occurred at the age of 8 weeks. No seizures have occurred since that time. Left-sided hemiparesis and cortical visual impairment were subsequently diagnosed. The history of the fused placentas and J. being the larger twin suggested an intrauterine blood transfusion resulting in a perinatal coagulopathy as the cause of a neonatal infarction at the time of the seizures. Because most of the damage occurred in the visual cortex with minimal damage in the periventricular regions, the pediatric ophthalmologist believed that J.'s condition had a good prognosis for some visual recovery.

Evaluation and Implications for Occupational Therapy

J.'s parents were referred to the infant intervention program by the developmental pediatrician when J. was 5 months old. The infant team consisted of the team coordinator, who is a nurse; an infant teacher; an occupational therapist; a speech pathologist; and a social worker. A developmental evaluation performed by the team revealed that J. was functioning at a developmental level of 2 to 3 months in all areas. The functional visual evaluation performed by the occupational therapist and nurse revealed inattention and lack of fixation on real objects and faces in normal lighting, absence of reaching attempts, inability to visually track or follow a lighted toy in a darkened room, and short visual attention to colored gels on the lightbox (a large box with fluorescent lighting covered with a white opaque piece of plastic). The pattern of J.'s visual responses consisted of a momentary peripheral glance and then looking away, with a return peripheral glance after this pause. Left esotropia and a fine rotary motor nystagmus were also observed. Her parents reported a lack of visual alertness to all environmental stimuli in the home.

Functional long-term team goals were established and included (a) improve visual awareness to light and promote central fixation, (b) improve gaze shifting and visual following of a moving object, and (c) promote reaching. These goals were set to encourage visual awareness and self-directed exploration of real objects in the natural environment.

Equipment used included a lightbox with colored gels and forms, flashlights, toys with light and sound, and a color monitor computer screen with programs that promoted cause and effect with a jelly switch. The color monitor computer provided simple high-contrast visual experiences and immediate visual reinforcement for developing cause and effect abilities. All of the equipment with the exception of the computer was also used by the parents in the home environment.

Simplification of the visual environment was the primary modification made in J.'s home and play environment. Some strategies used included use of a solid colored blanket on the floor when toys were presented, presentation of one object or toy at a time against a solid colored background, and reduction of the amount of visual stimuli in her crib and bedroom. Reaching for a visual object was encouraged through the provision of a tactile cue to the hand with the object and by assisting J. to touch the light, toy, or lightbox when she was visually engaged.

J. was seen weekly by the occupational therapist or nurse on the infant intervention team. Her visual needs were also explained to family members and other professionals working with her. Initially, there were no visual expectations during her motor therapy sessions because it was too difficult for her to visually engage when she was working on specific motor goals. When children with CVI have impaired motor skills, they appear to use a larger share of energy for posture and cannot maintain visual attention, so it is recommended that intervention plans for the different problems not be carried out simultaneously (Groenveld et al., 1990).

Occupational Therapy Intervention and Results

Results, interventions, and short-term goals are presented on a biweekly basis.

7 months. Results for J. were as follows:

1. Demonstrated lower peripheral field predominance with several short periods of central fixation (3–5 sec) on solid colored gels on lightbox. No attention to gels with complex patterns noted.
2. Followed a lighted toy in a darkened room horizontally from peripheral field to midline.
3. Tactile cues to the hand precipitated momentary looking at a lighted toy.
4. Performed short peripheral glances followed by gaze aversion predominated when looking at real objects.
5. Began to lift head in prone position to look at the lightbox.

Goals and interventions for next 2 months were as follows:
1. Elicit visual fixation and reach: (a) provided tactile cue to hand while moving lighted toy into visual field; (b) continued to use lighted toys in a darkened room to provide optimum contrast and reduce confusing, irrelevant visual input; (c) recommended simple toys and mobiles that were permanently attached over the infant seat and crib to promote reach, exploration, and development of cause and effect.
2. Improve head control through use of one lighted toy or lightbox as a visual stimulus when playing in the prone position.

9 months. Results for J were as follows:
1. Showed improved ability to handle increased visual stimuli.
2. Visual pauses consisted of blank stares as well as looking away.
3. Shifted gaze between two different colored gels on lightbox.
4. Demonstrated preference of one lighted toy over another.
5. Showed central fixation (20–25 sec) on complex gels on lightbox.
6. Improved spontaneous reaching for lighted toy with fewer tactile cues.

Goals and interventions for next 2 months were as follows:
1. Improve gaze shifting: (a) introduced two different geometric patterns of different color on lightbox, (b) presented pairs of different lighted toys.
2. Develop understanding of cause and effect: (a) used lighted switch toys that activated music, (b) introduced simple cause and effect programs on computer with jelly switch.

11 months. Results for J were as follows:
1. Consistently reached for lighted toy with both hands and no tactile cue.
2. Showed central fixation of 40 sec on large simple picture of face on lightbox.
3. Vocalized pleasure at happy face on lightbox and turned away from sad face.
4. Assumed hands and knees position to look at lightbox.
5. Paid brief visual attention to mother’s face.
7. Consistently activated jelly switch on computer.

At 18 months, J spontaneously crept to familiar toys in her environment for self-directed play. She was also observed to perform her first incidental visual learning. After she observed the occupational therapist turning a handle on an unfamiliar jack-in-the-box several times, she reached for the handle to attempt the action. Her mother reported that she was becoming increasingly visually aware of familiar objects in her environment.

Discussion
The major treatment goal with infants and children with CVI is to maximize the use of residual vision so that these children have a better opportunity for learning from the
environment. Occupational therapy is frequently a primary intervention service that is provided when a child has multiple disabilities, specifically to improve the child’s ability to play and learn. Many with multiple disabilities present with CVI and it is critical that occupational therapists address the visual needs of these children to maximize their potential for developing functional skills. Today there is a variety of technology, such as computers and communication systems, available to assist this process. However, most of this technology requires the child to have some visual functioning to access it for learning and communication. As noted in the literature review, standard visually enriched environments usually associated with working with children with visual impairments may not work for children with CVI, because as the visual input is increased it becomes harder for these children to visually process the information (Groenveld et al., 1990). This rich visual environment may result in children with CVI not being able to use their residual vision (Good et al., 1994). If these children do not learn to use their vision, then they may not be able to use that technology.

Our infant intervention program designed to promote residual vision in children with CVI has been in existence for 6 years. During that time, we have observed some or all of the visual behaviors that were noted in the literature. One of the behaviors that is mentioned in the literature that has been of particular interest to us is the frequent gaze aversion immediately before reach. It is not known why children avert their gaze. Some have suggested that it may be due to the use of peripheral fields (Good et al., 1994; Jan & Wong, 1991). We believe that this gaze aversion may be present because the child needs the pause time to assimilate and process the visual information before being able to return to looking at the stimulus. In a review of literature on visual gaze behaviors, Morse (1991) cited several studies that reported that subjects looked at their partner approximately 50% of the time in routine communicative exchange and looked more frequently and for longer periods when they were listening rather than speaking. It was hypothesized that gaze aversion controlled for possible distracting information and this gaze pattern may have helped the subjects organize the visual information (Rutter, cited in Morse, 1991). A similar phenomenon may be true of children with CVI—that breaks in fixation allow the child time to assimilate and organize. We often see a pattern of looking, looking away, and looking again. It seems that the more complex the visual information becomes, the more frequently these gaze aversions occur. As the child improves in visual abilities, the length of time that fixation occurs often increases while the frequency of the pauses decreases. Certainly, more information needs to be gathered on these visual responses.

The importance of simplifying and modifying the visual environment seems critical when working with children with CVI. In the case study presented, J. initially had almost no functional visual responses. Her parents reported that she was virtually “blind” and totally visually unresponsive to her surroundings. With modifications to her visual environment using visual simplification, high-contrast colored lighting on the lightbox, elimination of visual distractors in her immediate environment to reduce the “crowding effect” of too many visual stimuli, tactile integration of visual input, and use of common objects, J. demonstrated definite improvements in her visual responses. At this time she is learning to use and enjoy a computer, which may be a primary tool for her learning in the future. As occupational therapists in the field of pediatrics, we have an important role in understanding and addressing the visual needs of children who present with multiple disabilities and cortical visual impairment. If we address these visual problems when planning our interventions, we may achieve greater successes with children who present with CVI.

References