

Pediatric Cortical Visual Impairment 101

An Introduction For The American Conference On Pediatric Cortical Visual Impairment

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Introduction

If you are reading this document, you have elected to attend The American Conference on Pediatric Cortical Visual Impairment. This conference has been held annually since 2012 in Omaha, Nebraska, USA. The purpose of this document is to provide the introductory information you will need to be familiar with in order to fully engage the conference. In the early years of this meeting, half of the time was spent covering basic topics for the good of those beginning their study of pediatric cortical visual impairment (PCVI). As the conference is developing and maturing, it became obvious that more time is needed at the conference to explore more advanced topics; such as scientific research, parent issues, advocacy, various approaches offered by vision, therapy and educational professionals with expertise in caring for children with cortical visual impairment (CVI), and so on. Though this information may be briefly covered in the meeting, there will be no intentional effort to do so. A deliberate study of this material prior to the American Conference on PCVI will give you the foundation needed for your maximum educational benefit.

Relevance

Cortical visual impairment has become the leading cause of visual impairment in children in developed countries (1,2,3). This has occurred because of medical advances that have led to better visual outcomes in retinopathy of prematurity, congenital glaucoma and congenital cataract; and at the same time, improved survival of children with central nervous system diseases. The number of children afflicted with CVI is increasing, elevating it to a public health concern of high significance. The National Institute of Health website sites a CVI prevalence of 10.5% of all children with developmental disabilities. Despite the concerted and productive efforts of researchers and clinicians in the field of PCVI, the knowledge

base surrounding PCVI is scant. Professionals and parents whom care for children with CVI frequently share feelings of inadequacy in confronting the questions of, “What does this child see? How does this child see?” and, “How can I improve this child’s ability to see?” The American Conference on Pediatric Cortical Visual Impairment was conceived to address these pressing problems.

Mission Statement of The American Conference on Pediatric CVI

- *To enhance interdisciplinary understanding of pediatric cortical visual impairment between the disciplines of vision education, occupational therapy, optometry and ophthalmology, ultimately leading to improved vision care for children with cortical visual impairment.*

Basic Science

The sense of sight is a complex series of processes consisting of sensation followed by integration of sensory information into higher cortical centers. Sensation (figure 1 and the color red in figure 2) is commonly measured with visual acuity, visual field studies, contrast sensitivity, stereopsis and color vision. These processes occur in the occipital lobe cortex of the brain. Integrated visual functions occur when this sensory information is transmitted via white matter pathways to cortices in the temporal, parietal and frontal lobes of the brain. Integrated or higher order visual functions include motion perception, visual memory, target acquisition with slow and fast eye movements, separation of figure from back-ground based on figure novelty and back-ground complexity and other integrated visual functions. Cortical visual impairment refers to dysfunction of any of these visual functions due to disease posterior to the optic tract/lateral geniculate body (figure 1).

It is now understood that the higher order visual functions occur through 2 principal pathways. The *dorsal stream* (the color green in figure 2) reaches the parietal lobe and pre-frontal cortex and is responsible for spatial orientation and visual target acquisition. The *ventral stream* (the color blue in figure 2) reaches the temporal lobe cortex and is responsible for form perception and visual memory.

This is a gross oversimplification, however, it will suffice as introductory information.

The Sensory System

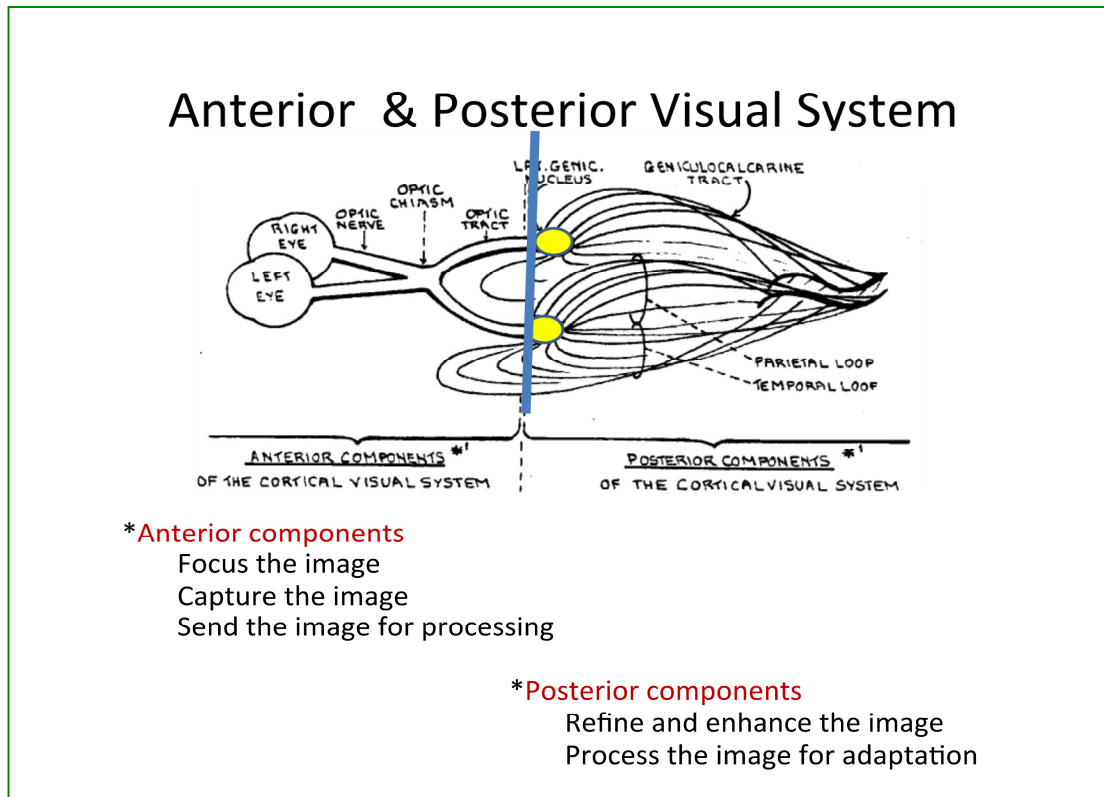


Figure 1 (With permission by Linda Baker-Nobles, OT/R)

Integrated or Higher Order Visual Processing

Visual input enters the eyeball, travels back to the occipital lobe, and then forward to the prefrontal cortex to complete processing

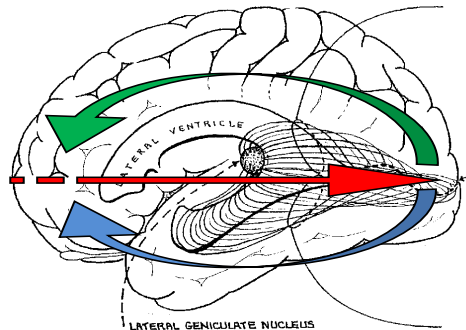


Figure 2 (With permission by Linda Baker-Nobles, OT/R)

Causes of Pediatric Cortical Visual Impairment

Disease states that cause dysfunction of these brain based visual pathways are the causes of PCVI. PCVI is not a disease. It is the end result of multiple brain diseases that affect children prior to birth, at birth, or after birth.

Table 1: Medical Causes of PCVI

(Courtesy of Alan Lantzy, MD)

- Asphyxia
- Periventricular Leukomalacia (PVL)
- Cerebral Vascular Accident (CVA)
- Trauma, including "Shaken Baby Syndrome"
- Hypoglycemia
- Congenital Brain Abnormalities
- Intrauterine Infections
- Acquired Infections
- Intra-ventricular Hemorrhage (IVH)
- Post-Hemorrhagic Hydrocephalus
- Kernicterus
- Chromosomal Abnormalities
- Infantile Spasm
- Mitochondrial Diseases

The CVI Range as Defined By Dr. Christine Roman

The CVI Range as defined by Dr. Roman consists of 10 characteristics of visual behavior that are quantifiable using techniques as described in her textbook, Cortical Visual Impairment (4). The Range does not measure visual sensation. The characteristics are visual processing functions that occur in cortices of the temporal, parietal and probably the frontal lobe. These characteristics are:

- Color Preference
- Need For Movement
- Visual Latency
- Visual Field Preference
- Difficulties With Visual Complexity
- Light-Gazing and Non-Purposeful Gaze
- Difficulty with Distance Viewing
- Atypical Visual Reflexes
- Difficulty With Visual Novelty
- Absence Of Visually Guided Reach

Each characteristic is scored on a 0.00-1.00 point scale. For example: Difficulties with Visual Complexity would be scored as follows.

0.00: Responds only in strictly controlled environments. Generally no regard for human face.

0.25: Visually fixates when environment is controlled.

0.50: Student tolerates low levels of familiar background noise. Regards familiar faces when voice does not compete.

0,75: Competing auditory stimuli tolerated during periods of viewing.

1.00: Only the most complex visual environments affect visual response. Views books or other two-dimensional materials.

If the child fixates only when the environment is controlled, they score a 0.25 for this characteristic. All 10 characteristics are scored and added together giving a CVI Range Score of 0.00 – 10.00

How a child scores on the range will place him/her in Phase I, Phase II or Phase III.

These phases may be summarized as:

Phase I (Range Score of 0.00 to 3.00)

Goal: Building visual behavior.

Phase II (Range Score of 3.25 to 7.00)

Goal: Integrating Vision and Function

Phase III (Range Score of 7.25 to 10.00*)

Goal: Resolution of CVI characteristics.

*Note: A score of 10.00 is currently theoretical as it has yet to be achieved.

Dr. Roman's system employs *interventions* specific to each *phase* that are designed to allow children to improve their phase level and hence, their visual engagement with the environment. Though there are other systems around the globe, Dr. Roman's is widely regarded as the most highly developed system for quantitating pediatric CVI. It is recommended that you familiarize yourself with Dr. Roman's textbook as the concepts in it are widely discussed at the American Conference on Pediatric CVI that you are going to attend.

Sandra Newcomb, PhD has published data addressing the reliability of Dr. Roman's Range (5). 104 children were rated by 12 professionals of various disciplines concerned with pediatric CVI. The internal consistency, test-retest and inter-rater reliability coefficients all exceed 0.83. Though design flaws in this study exist, it is concluded by Dr. Newcomb that the data supports the use of Dr. Roman's CVI Range in the management of children with CVI.

The Definition of Pediatric CVI As Determined by American Conference on Pediatric Cortical Visual Impairment

After deliberation by the speakers and moderators of the 2012 and 2013 meetings of the American Conference, the following definition of pediatric CVI was derived:

**“Congenital or acquired brain-based visual impairment
With onset in childhood, unexplained by an ocular disorder,
And associated with unique visual and behavioral characteristics.”**

This definition is interdisciplinary and was crafted by professionals from visual education, ophthalmology, optometry and occupational therapy. It is to serve as the beginning of a common language between disciplines. Note that such vision loss is “brain-based”. Though ocular disease can co-exist in a child with CVI, it does not explain the visual loss that the examiner is witnessing. “..onset in childhood” is understood to be prior to the age of puberty. “...unique and behavioral characteristics” refer to the *characteristics* of The CVI Range. Note that there is no mention of visual acuity, visual field, color vision, contrast sensitivity or stereopsis in this definition.

An Interdisciplinary Audience

By design, the conference you are about to attend is highly interdisciplinary. You will be able and encouraged to discuss openly aspects of PCVI with professionals that come with training and experience that is different from yours. This creates a rich environment for learning from each other, which serves the mission of the conference. The more you are willing to reach out to professionals of other fields and share information, the more you are likely to benefit from this conference.

A brief summary of represented professional fields follows.

Teachers Of The Visually Impaired—are specially trained and educated to teach students with visual impairments how to use their vision or adapt their environment to best learn. They teach Braille and other specialized areas of direct

instruction, make classroom accommodations, and connect students with assistive technology to enhance their educational experience.

Orientation and Mobility Specialists—are specifically trained and educated to teach students and adults with visual impairment and blindness how to safely and effectively navigate their environment, so that they can independently engage the world beyond their immediate surroundings.

Optometrists—are vision professionals who are concerned with how an individual gathers visual information from their environment. They are concerned with fine visual motor abilities, which include fixation, visual tracking, visual scanning, focusing, and binocular (eye teaming) function. Also of concern is how vision integrates with other sensory systems and how the brain perceives, interprets and processes sensory input. Optometrists assess these functions and maximize visual function with corrective lenses, prisms and visual therapies. They can make the diagnosis of CVI.

Occupational/Physical and Speech Therapists—are specially trained and educated professionals who provide therapies to improve outcomes in fine motor, gross motor, activities of daily living, and receptive/expressive language. They are concerned with how a child with CVI integrates his/her visual experience with the world around him/her.

Ophthalmologists—are medical doctors who specialize in the medical and surgical treatment of disorders of the eye and visual system. They can make the diagnosis of CVI. Ophthalmologists conduct eye surgery for ocular conditions such as misalignment, cataract and glaucoma.

Parents—are the relentless advocates for their children, and the children of others with CVI. They raise public awareness of CVI, expand their knowledge of CVI, and raise funds to support the Pediatric Cortical Visual Impairment Society.

The Pediatric CVI Society

The Pediatric Cortical Visual Impairment Society was created to more effectively further the aims of the Conference. Specifically, to further advocacy for children with CVI, heighten public awareness, promote research and other activities that would lead to improvement in vision care for children with CVI. A Board of 13 Directors and 5 Committees is established. The committees are Parent Affairs, Scientific Affairs, Board Affairs, Program and Meeting, and Advancement/Budget/Finance. Formal mission statement, by-laws, board and membership structure, 501c(3) status, website are all now in place. You are invited to attend the annual business meeting. You don't need to be member to attend. You may join the PCVI Society for \$50 at the annual meeting. Membership is open to all attendees of the Conference, as well as those parents and other parties interested in furthering the goals of the Society whom are unable to attend the yearly Conference.

THE PEDIATRIC CORTICAL VISUAL IMPAIRMENT SOCIETY

MISSION STATEMENT

The mission of the Pediatric Cortical Visual Impairment Society ("the Society") is to advocate for improvement in the quality of life of children with vision loss due to brain disorder, disease or injury. The mission is restricted to matters concerning the sense of vision.

Goals

The mission of the Society will be fulfilled through the pursuit of the following goals:

1. Advance interdisciplinary education and research.
2. Enhance dissemination of information and its communication among and between professionals, parents, government and educational institutions, and other groups whom impact children with cortical visual impairment.
3. Advocate for the appropriate allocation of resources to allow for improved vision services for children with cortical visual impairment.
4. Improve public and professional awareness of cortical visual impairment in children.
5. Advocate for governmental policy that improves quality and quantity of vision services available to children with cortical visual impairment.

6. Engage in fund raising activities that will allow for advancement of the Mission and the Goals of the Society.

Welcome

You have decided to spend 2 days of your year in Omaha strengthening your knowledge of pediatric CVI. On behalf of the planning committee, I would like to say that we are honored to have you at this Conference. Welcome! Get to all of the presentations that you can. Talk to people outside your discipline and your geographic home. You will find many who share your enthusiasm for learning how to better help these children whom are waiting for us to find ways to help them visually connect with our world.

Bibliography

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Revised 3/30/17