

2019 AAPOS 45th Annual Meeting

Workshop Handout

Cortical/Cerebral Visual Impairment 2019: What You Need To Know To Diagnose and Treat

Lehman

Definition of CVI

- Bilateral visual impairment due to brain damage of the posterior visual pathway
- Eye structure is typically normal or the pathology found (optic atrophy) does not explain visual impairment
- Children with CVI display characteristic behaviors

Causes

Structural: brain malformations, tumors

Vascular: periventricular leukomalacia (PVL) secondary to prematurity, hypoxic/ischemic event, perinatal stroke

Infectious: meningitis, encephalitis

Inflammatory: vasculitis (inflammation of blood vessels)

Trauma: TBI, nonaccidental trauma

Metabolic: neonatal hypoglycemia, mitochondrial disease, lysosomal disorders

Neurologic disease: seizure, hydrocephalus

Cortical vs. Cerebral Visual Impairment

Various interpretations:

Outside the United States, the term cerebral is more commonly used

Some use terms interchangeably

Some use cerebral as a broader term which includes vision issues due to brain damage of more superficial parts of the brain (cortical) as well as deeper structures in the brain

Some use term cerebral to mean higher level perceptual and processing issues

Characteristics in infants or children with significant neurologic impairment

Variable, poor or atypical response to visual stimuli

Poor or inefficient use of visually guided (eg.: eccentric viewing or frequent looking away)

Latency (delay in visual response)

Difficulty with distance viewing

Difficulty with novelty

Preference for familiarity

Light gazing (preference for looking at light)

Color preference

Difficulty with complexity

Preference for certain visual fields

Better visual performance with movement

Reference:

Roman-Lantzy, C. (2007). Cortical visual impairment: An approach to assessment and intervention. New York, NY: AFB

Characteristics in older children with higher function (Cerebral?)

- Slow and inefficient visual performance
- Deficiency of visual perception and integration
- Contrast sensitivity impairment
- Difficulty with complexity
- Short visual attention
- Poor visual memory
- Difficulty with object recognition/face recognition

Damage to connections of visual centers

Dorsal stream function – getting there

Connections between occipital area to parietal areas

Responsible for:

Finding objects in space

Figure/background

Extremity movement

Examples of deficits:

Difficulty with steps or changes in surfaces

Inaccurate reach

Difficulty with complexity

Ventral stream function – who or what is there

Connect between occipital area and temporal lobe

Responsible for:

Form recognition

Visual memory

Examples of deficits:

Forget location of objects

Difficulty with recognition of faces, shapes, objects

Reference: Lueck AH, Dutton GN, editors. Vision and the Brain: Understanding Cerebral Visual Impairment in Children. New York, NY: AFB Press; 2015

Visual acuity

Variable

Acuity may be normal

Deficit may be higher level

Processing or interpretation of visual information

May be called cognitive visual dysfunction

Visual Field

Various types may be seen:

Hemianopsia

Inferior field loss

CVI Phases (Roman-Lantzy)

Phase is a period in the resolution of CVI that indicates degree or severity of CVI

Phase I: Building visual behavior

Phase II: Integrating vision with function

Phase III: Resolution of remaining CVI characteristics

Phases are useful in planning for appropriate interventions and environmental accommodations

Reference:

Roman-Lantzy, C. (2007). *Cortical visual impairment: An approach to assessment and intervention*. New York, NY: AFB

Recovery of visual function in CVI

Most children have some degree of improvement

Recovery can occur over months to years

Degree of recovery cannot be predicted from imaging studies

References:

Matsuba CA, Jan JE. Dev Med Child Neurol. Long-term outcome of children with cortical visual impairment; 2006 Jun;48(6):508-12.

Watson T¹, Orel-Bixler D, Haegerstrom-Portnoy G; Optom Vis Sci.; Longitudinal quantitative assessment of vision function in children with cortical visual impairment 2007 Jun;84(6):471-80.

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Comorbid conditions

Prematurity, cerebral palsy, ADHD, autism, cognitive impairment

Effect of comorbid conditions:

Difficulty in developmental/educational testing

Behavioral mannerisms of autism confused with characteristics

CVI Workup/Treatment with Pediatric Ophthalmologist

- Ophthalmologic evaluation with pediatric ophthalmologist
- Perform history and physical
- Treat ophthalmologic problems
- Provide diagnosis of cortical visual impairment
- Provide medical necessity necessary to obtain vision services
- Refer for vision services
- Provide educational information for family
- Make specific recommendations for child based on characteristics
- Provide information concerning child medical condition to family and team

Language to use for documentation:

Declare visual impairment:

- List cortical visual impairment and visual impairment in diagnosis list
- Cortical visual impairment interferes with child's ability to access educational materials.

Declare need for vision services:

- (Patient's first name) requires direct ongoing evaluation and follow up of a teacher of the visually impaired experienced with cortical visual impairment.

CVI Treatment-Don't Forget

- Correct refractive error
- Treat accommodative insufficiency
- Treat amblyopia if needed

CVI Workup/Treatment with Teacher of Visually Impaired

- Functional evaluation performed by teacher of the visually impaired experienced in cortical visual impairment
 - History
 - Functional evaluation
 - Recommendations based on characteristics
 - Periodic reassessments

Multidisciplinary team approach

- parents/family/guardians
- primary care physician
- pediatric ophthalmologist
- pediatric neurologist
- teacher of visually impaired
- occupational therapist
- physical therapist
- speech therapist
- teacher of the hearing impaired
- augmentative and alternate communication specialist
- orientation and mobility specialist
- feeding specialist

Parental Involvement

Parents are experts about their children. It is important for parents to share their observations about their child's visual function with members of the child's care team.

There should be trust for parental observations about their child's vision.

Parent's concerns and observations should be validated.

Parental expectations should be discussed.

Parental advocacy should be supported and encouraged.

Prevention

Improved prenatal care

Safety recommendations in order to reduce TBI (helmet use, seat belt)

Refinement of techniques to reduce neuronal damage (head and body cooling to reduce damage in hypoxic-ischemic encephalopathy)

CVI Diagnosis and Assessment of Visual Function: Evaluation tools 2019 - Schwartz

1. Focused medical history:

Looking for disease associated with damage to vision processing parts of the brain
In children with good visual acuity, abnormal prenatal/perinatal medical hx is the most important risk factor for CVI, in deciding which kids should be referred for assessment.

Premature infants: PVL (White matter disease of Immaturity)

Term infants: Hypoxic Ischemic Encephalopathy

Cerebral vascular accident (stroke)

Meningitis/encephalitis

Hydrocephalus

Head trauma

Seizures

In utero drug exposure

9% no known cause

Diagnosing Cerebral Visual Impairment in Children with Good Visual Acuity. M Gendern et al. Strabismus 20(2), 2012

2. Screening and History questionnaires

Preverbal Visual Assessment Questionnaire (PreViAs): Useful to detect abnormal visual function from 0 - 24 months of age. 4 visual domains: visual attention, visual communication, visual-motor coordination, and visual processing

García-Ormaechea, Inés, et al. "Validation of the preverbal visual assessment (PreViAs) questionnaire." Early human development 90.10 (2014): 635-638.

Visual Skills Inventory: Inventory (administered by parents & caregivers) which may help provide guidance about the quality of visual function. This study identified questions with the highest validity for predicting vision.

McCulloch, Daphne L., et al. "A visual skills inventory for children with neurological impairments." Developmental Medicine & Child Neurology 49.10 (2007): 757-763.

Visual Function Questionnaire: A screening tool for CVI: Parent questionnaire to screen for CVI. Therefore good predictive value for identifying children with CVI.

Ortibus, Els, et al. "Screening for cerebral visual impairment: value of a CVI questionnaire." *Neuropediatrics* 42.04 (2011): 138-147.

Inventory of Visual Function: Questionnaire: Assessment of visual function in children with CVI. Cerebral Visual Impairment in Children: Visuoceptive and Visuocognitive Disorders 2015 Edition. Josef Zihl, Gordon N. Dutton

3. Online aids for screening and intervention:

- Teach CVI: <https://www.teachcvi.net/>

TEACHCVI is a partnership that aims to create collaborative tools for teachers and health care professionals. It is meant to build a bridge between teachers/educators and health care professionals so they can work together to benefit the target group: children with Cerebral Visual Impairment (CVI).

SCREENING TOOLS

Screening tools developed as a first step to decide when to refer children with a suspicion of CVI for further assessment.

3 screening lists:

List 1. Focused on children with a motor disability who are non-ambulatory

List 2. Focused on children with developmental age between 2 – 6 years

List 3. Focused on children with developmental age between 6 – 12 years

- Gestalt ReVision Leuven: <http://www.gestaltrevision.be>

- The Leuven Perceptual Organization Screening Test (L-POST)

The L-POST is designed to offer clinicians, neuropsychologists and researchers a tool to assess potential deficits in mid-level vision in different patient populations. The screening test contains 15 subtests which test a range of mid-level processes, including figure-ground segmentation, local and global processing, shape perception and the ability to use a range of grouping cues including common fate, co-linearity, proximity and closure.

Torfs, K et al. (2013). The Leuven Perceptual Organization Screening Test (L-POST), an online test to assess mid-level visual perception. *Behavior research methods*, 1-16 (* joint first authors).

Vancleef, K., et al. (2015). Reliability and validity of the Leuven Perceptual Organization Screening Test (L-POST). *Journal of Neuropsychology*, 9(2), 271-298.

Educational Resource Material for Physicians

Lueck AH, Dutton GN, editors. Vision and the Brain: Understanding Cerebral Visual Impairment in Children. New York, NY: AFB Press; 2015

Dutton GN, Bax M, editors. Clinics in developmental medicine no. 186: Visual Impairment in children due to damage to the brain. London: Mac Keith Press; 2010

Roman-Lantzy C. Cortical Visual Impairment: An Approach to Assessment and Intervention (second edition) New York, NY: AFB Press; 2018

CVI Resources: Can be located on AAPOS Website Low Vision Committee Section

<https://aapos.org/education/educational-resources/pediatric-low-vision-education>

Cerebral Visual Impairment (CVI) Scotland:

CVI Scotland is devoted to helping people understand cerebral visual impairments, and together working towards beginning to master this complex spectrum of conditions.

CVI Information Day Presentation:

A fifty minute video of Gordon Dutton's presentation at a CVI Information Day that offers a comprehensive introduction to vision and cerebral visual impairments.

Sharing & Developing Our Understanding of CVI:

Guide for Parents.

Pediatric Cortical Visual Impairment Society:

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.

The Perkins School for the Blind:

Offers on-line courses, lectures and informational sessions.

The American Foundation for the Blind:

Textbooks associated with education of children with CVI.

The American Printing House for the Blind:

Products for students with CVI.

The American Conference on Pediatric Cortical Visual Impairment:

Interdisciplinary conference for professionals and parents, usually held in late June.

Strategy To See's:

The mission is to provide strategies, suggestions and techniques to parents, caretakers, teachers and other action heroes, who hope to encourage more consistent and efficient use of vision in children with Cerebral/Cortical Visual Impairment.

KanLovKids:

Website with webinars on children with visual impairment and visual development.

CVI Connect:

iPad app for CVI and a community of professionals and technology that offer hope and confidence in the form of personalized education for children with CVI.

Facebook groups:

- [CVI Neuroplasticity Research Group](#)
- [CVI Phase III Community](#)
- [CVI Connection](#)

Parent Support Groups:

- [Start Seeing CVI](#)
- [CVI Momifesto](#)

School Design & Vision Impairment:

The development of a new school for children with multiple disabilities and visual and hearing dual sensory impairment presents particular challenges in design, equipment and staffing. The environment of the school should ideally allow children to be as independent as possible. The design of the school needs to take into account the sensory and central processing functions of the children so that the impediments caused by these limitations are minimised. Such an approach is likely to lead to enhanced learning with fewer resources and should therefore prove both beneficial for the children and cost efficient. This paper provides a preliminary set of ideas from the perspective of an ophthalmologist who looks after these children.

The Importance of Vision:

Why is the vision so important? What special role does it play in learning and guiding development from the moment of birth? What is the value of good visual functioning? How does vision affect and impact the development and activities of daily living of children and adolescents?

Vision After Hemispherectomy, TPO Disconnection, and Occipital Lobectomy:

An Introductory Guide

Vision Evaluations after Epilepsy Surgery

Zika Care Connect:

The Zika Care Connect website contains information to help pregnant women and families find the appropriate healthcare professional and resources that can address their specific needs.