Cortical/Cerebral Visual Impairment 2019

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:

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


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 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:

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:

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


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


   **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.

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


Inventory of Visual Function: Questionnaire: Assessment of visual function in children with CVI.


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).

SCRENNING 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).
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Educational Resource Material for Physicians


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.