CLEAR-CP: A Transdisciplinary Approach to Diagnosis and Management of Cortical Visual Impairment (CVI) in Children with CP and other Motor Impairments

Focused Ophthalmology history and exam used to support a diagnosis of CVI and evaluation of functional vision

CVI: the challenge
   Diffuse brain pathology and systemic conditions that defy standardized measures
   Impaired global cognitive function, speech and motor function that challenge conventional examination
Visual impairment is often: Unrecognized, poorly characterized, difficult to quantify

Focused medical history:
   Goal is to identify diagnoses/disorders associated with damage to vision processing parts of the brain (Gendern et al 2012):
     Premature infants: PVL aka White matter disease of infancy (WMDI)
     Term infants: Hypoxic Ischemic Encephalopathy
     Cerebral vascular accident (stroke)
     Meningitis/encephalitis
     Hydrocephalus
     Head trauma
     Seizures
     In utero drug exposure

Focused visual history:
   Visual curiosity (present/absent)
   Classic Visual Behaviors associated with CVI
     Variable visual attention (unfamiliar/complex environment)
     Supplementing vision with touch
     Looking away when reaching
     Close viewing (despite corrected R.E.)
     Increased attention for moving object
     Attraction to colored objects
     Light gazing / Photophobia

Visual function questionnaires
   1. Visual skills inventory (McCulloch et al 2007): Visual impairment can be difficult to elicit and can be missed. This study demonstrates that a visual skills inventory (administered by parents & caregivers) is strongly associated with visual function and may provide guidance about the quality of visual function in such children.
   2. PreviAs questionnaire: The preverbal visual assessment (PreViAs) questionnaire (García-Ormaechea et al 2014): The PreViAs questionnaire is useful to detect abnormal visual maturation in infants from birth to 24 months of age. It improves the anamnesis process in infants at risk of visual dysfunctions.
      Assesses four visual domains: attention, communication, visual–motor coordination, processing
   3. Visual function questionnaire: (Ortibus et al 2011): Evaluates the utility of a questionnaire to screen for CVI. Parent questionnaire followed by diagnostic
evaluation for CVI of child. **Conclusion:** CVI questionnaire is a viable took that has the potential of being implemented as part of a routine screening procedure for CVI.

46 items exploring CVI characteristics

Visual attitude, Ventral/Dorsal stream, Complex problems, Other senses, Associated characteristics

4. Questionnaire developed by Dutton et al as a screening questions for older children.

**Focused examination:**

![Diagram of visual pathways]

**Visual pathways:**

**Anterior pathway** (eye, optic nerve, chiasm, optic tract):

- Visual acuity, Contrast Sensitivity, Accommodation
- Oculomotor functions: fixation (steadiness), smooth pursuit, saccades, convergent spasm, paroxysmal deviations, strabismus, nystagmus

**Posterior pathway** (optic radiations, primary (occipital) visual cortex): Most of our cognitive **functions** and perceptual processes are carried out by the cortex which is the largest part of the human brain. The **primary visual cortex** is that area of the brain that receives **visual** input from the retina and is just the beginning of complex visual processing.

- Visual acuity, contrast sensitivity, color perception, stereopsis, target localization, spatial perception, motion perception, visual field

**Retino-Tectal pathway:** Subcortical or “subconscious” visual system that allows us to move effortlessly through the visual world while talking or thinking about something else. It is this path that is involved in orienting the eye toward a stimulus that in the peripheral field of vision. It is the primary visual system up to 2 months, after which time the cortex takes over executive function with different streams processing specific aspects of visual information. It accounts for fixation and following behavior in infants with extensive cortical lesions.

**Visual processing** (and spectrum of visual deficits)

**Ventral (WHAT) stream**

Recognition of objects and shapes as well as problems of spatial orientation.
Vision Library of Faces/Objects/Routes
- Process physical attributes of an image
- Language processing
- Visual memory and visually identifying emotion

Damage to Ventral Stream:
- Distinguishing between objects with similar features (dog-cat)
- Recognizing faces
- Recognizing facial expressions (decoding emotions)
- Visually recalling a route
- Picturing what objects are in a drawer
- Visual amnesia (no access to stored visual objects)
- Visual alexia (reading)
- Visual anomia (naming)

Dorsal Stream (WHERE/HOW) Stream
- Simultaneous perception and detection of movement, framework for spatial awareness, orientation in space, and planning for motor functions (eye-hand coordination).
  - Analyze the size and direction of objects
  - Location of objects in visual space
  - Visual guidance of movement

Damage to Dorsal Stream cause difficulty with:
- Handling “lots of information”
- Visually mapping
- Communication between vision and movement
- Communication between sight and decision making
- Function more “like a baby” (Babies do one thing at a time, when doing one task, they are less aware of other stimuli)

Focused Exam:
1. Pupil response to light: 30 weeks gestation, retino-pupil pathway no cortex involved
2. Blink to light: 25 - 30 weeks gestation, requires intact optic nerve/pretectum not cortex
3. Blink to threat: 2 – 5 month, Not co-natal, a learned response, Higher order processing (intact 1° visual cortex) and visual attention (parietal/frontal/cingulate gyrus)
4. Visual fixation: reliable by 2 months (corrected gestational age)

Eye Movements to Assess Vision
- Eye movement development
  - Fixation and following movement – 2 months
  - Saccades – 4 months (hypometric before)
  - Smooth pursuit – 6 – 8 weeks
- Limitations: Limited eye movement? (Oculomotor apraxia), Poor head control? (Hypotonia)

Eliciting Visual Response: wait for it
- Indirect signs of vision
  - **Direct Va Signs:**
    - Object localization
    - Head or limb movement toward object
    - Eye movement toward object (pursuit/saccade)
  - **Indirect Va Signs:**
    - Posture alteration
Change in breathing
Avoidance
Smiling (or other change in expression)

**OKN (optokinetic nystagmus):** Present at birth. Subcortical response to large field motion which can be voluntarily suppressed.

**Visual acuity:** About 20/600 at birth with earliest 20/20 by 2 – 3 years (~age 5)

**Color perception:** Can’t distinguish different colored bits until > 3 m, preference for high contrast primary colors (early), probably adult-like by 2 – 3 years

**Peripheral visual field:** full (adult like) by 1 year of age. Plastic reorganization after damage to optic radiations allows rewiring of connections and restoration of full field vision (unless basal ganglia or thalamus involved). Visual field defects are common especially in the lower hemifield except for spastic diplegia in which we see homonymous hemianopia.

**Depth perception (stereopsis):** adult-like by 3 – 7 months

**Accommodation:** adult – like by 4 months

**Directed exam based on dorsal and ventral stream function**
- Facial expression (ventral stream)
- Facial recognition (ventral stream)
- Spatial orientation (dorsal stream) Mailbox game: Assesses visual perception of line orientation.
- Size perception/orientation (dorsal stream) with LEA rectangles game
  - By 3 years child can visually distinguish between a longer and a shorter rectangle

**Occupational Therapist components used to support a CVI diagnosis**
- A medical history that includes neurological problems associated with a diagnosis of CVI
- See focused History above for a list of the conditions
- Near normal eye exam that reveals an eye condition which cannot explain their lack of functional vision
- The presence of unique visual and behavioral characteristics
  - The 10 characteristics of the CVI Range
    - Color preference
    - Need for movement
    - Visual latency
    - Visual field preference
    - Difficulty with visual complexity
    - Need for light
    - Difficulty with distance viewing
    - Atypical visual reflexes
    - Difficulty with novelty
    - Absence of visual guided reach

**CLEAR CP clinic, a low vision rehabilitation clinic**
- History
  - CLEAR CP is Children’s Low Vision Education and Rehabilitation – Cerebral Palsy
Began in 2012 – Is Grant funding from the Ettlinger Trust Fund to establish a model clinic at the Perlman Center to evaluate children with cerebral palsy/physical disabilities and cortical visual impairment (CVI)

The CLEAR-CP Clinic – is comprehensive evaluations by an ophthalmologist from the Division of Ophthalmology and an occupational therapist from the Perlman Center.

Those involved and their roles
Medical Director of CLEAR CP clinic
Occupational therapist
Ophthalmologic Tech
Teacher of the visually impaired (TVI’s)
Customer Service representative

CLEAR CP Clinic Process (see flow chart)

A transdisciplinary collaboration model is vital because:

- Increases the likelihood of comprehensive care
- Improves the quality of patient education and support delivered to patient
- Ensures patient centered, coordination of care
- Strategies to increase the success of our transdisciplinary collaboration
  - Education and training for occupational therapists to administer and interpret the CVI Range
  - Communication between all occupational therapists within our facility who give the CVI range
  - Collaboration among departments to ensure consistency of documentation

Strategies for effective clinic follow-up and management

- Provide a comprehensive report to families with medical professional and therapist notes and recommendations
- Schedule a follow up with occupational therapist 4-6 weeks after the clinic visit to answer questions
- Provide further suggestions to integrate recommendation into home and community settings
- Provide contact information to child’s school teams and other therapist
- Recommend follow-up CVI range about 1 time yearly to progress the interventions as needed
CLEAR CP clinic
- CLEAR CP clinic is on the 4th Tuesday of each month
- Up to 4 patients are seen in the clinic each month
- Patient interaction/assessment half of the day (4-5 hours)
- Documentation and write up of detailed report

Referrals
- Cincinnati Children’s CP clinic
- Perlman Center Therapeutic Programs
- Other community or Children’s departments

Scheduling
- Referrals are triaged through ophthalmology and funneled to CLEAR CP clinic
- Questionnaires are sent to families once identified
- Child is scheduled once paperwork is returned

Day of clinic flow
- Check in, brief history, functional vision assessment – Ophthalmology Tech
- Provide child’s positioning needs for best visual attention - Occupational Therapist
- Brief eye exam – Medical director of CLEAR CP
- Parent identifies and communicated community needs – Teacher of visually impaired
- CVI Range administered – Occupational Therapist

Follow up
- Combined, detailed ophthalmology and occupational therapy report sent to family 1-2 weeks after visit
- Follow-up visit scheduled 4-6 weeks after visit with occupational therapist
- Review specific, client centered recommendations from CVI Range report
- Provide additional resources to the family
- Provide resource for community school outreach visit
Bibliography

**Questionnaires for Vision:**


**Assessing visual function:**


**Other:**


**Cortical Visual Impairment Assessment:**


**Transdisciplinary Care**