Session IV: The Gauntlet; Tell me What to Say in Common Systemic Disorders
1:40PM  Cerebral/Cortical Visual Impairment  Linda M Lawrence MD

Introduction:
Pediatric ophthalmologist not only care for children but support their families and caregivers. Therefore, communication about cerebral/cortical visual impairment, has to involve the family, and the team working with the child. The challenge is how the busy ophthalmologist can accomplish this. You have to have a strategy. I have worked in the field of CVI for over 30 years, so learned from many families, teachers/therapists, and other team members the importance of family centered interventions. The family has a child 80% of their awake time, and therefore needs to be empowered to integrate all “therapies” into daily routines in their family, to enhance the developmental outcomes of the child.

Brain based visual impairment (cortical, cerebral/cortical, cerebral, neurological visual impairment, all referred to as CVI) is the most common diagnosis for newly diagnosed visual impairment in the developed world, and also fast becoming so in the developing world. The reason is better pediatric and neonatal care. We are saving lives with better neonatal care and saving eyes/vision from other causes like congenital glaucoma, cataract, and retinopathy of prematurity. CVI is an invisible disability, and difficult for others to understand.

The Pediatric or Pediatric oriented ophthalmologist will be challenged to care for these infants and children. Not only with a diagnosis with optical, medical, and surgical treatment, but compassionate support for the family. Recommendations for basic intervention strategies start at the time of diagnosis by the ophthalmologist. Proper referral and guidance to early interventionists including teachers and therapists should be as soon as possible, not “let’s wait and see”. A baby or child with low vision can be significantly delayed in development and learning, this is a developmental urgency!

The ophthalmologist needs to understand, they are not the main interventionist. CVI is not a disease we cure, but we can lessen the effect of the disability with proper interventions, by those trained in these interventions. In the USA, referral at time of diagnosis of a visual impairment is made to an early intervention program, funded by IDEA Part C. The services in many states are free for the assessment of any child with a suspected disability, then depending on the State, intervention services may be covered partially or completely by Federal or state funds. Each ophthalmologist should understand their State’s Part C program referral system. The early intervention program then consults the Teacher of the Visually impaired (TVI or TSBVI) . These are highly trained professional special educators .There are also non for-profit organizations for preschool for children with VI as well as state and private schools for the blind. Once a child is in the public-school system they may qualify for Part B services under IDEA. These are free services to the child in public school, private schools may vary from state. The early intervention specialist and team makes the decision about the proper referral for vision services, then support the family on an ongoing basis. The school system typically makes the recommendation for the involvement from the school for the blind, or TVI.

The ophthalmologist must understand the local and state structure and referral system developmental and educational assessment and intervention. We cannot give “orders” to the educational system. We can make recommendations for the team and family.
When you start seeing babies and children with CVI, you need to have a strategy for assessment, intervention, and (re)habilitation.

1. The ophthalmologist’s role is to perform medical and vision history, assess the visual system both anatomically, and functionally, and make appropriate diagnoses.
2. The different between visual acuity and visual function (use of vision for function of the child) must be understood. These are NOT the same.
3. The ophthalmologist must understand visual developmental milestones when accessing babies and small children, and early intervention programs referral for those at risk including what is need for the IFSP (Individual family planning program)
4. The ophthalmologist must understand the requirements of the school system for assessment and educational interventions for older children including the IEP (individual educational plan) requirements in their state.
5. The ophthalmologist should have appropriate referral and resources available so that each family and child can have the best avenues for intervention.
6. The ophthalmologist should understand that the principle interventions in CVI involve simplification: decrease clutter and complexity, decrease crowding, increase contrast, allow time to process.
References you may not know about:

This chapter on Pediatric Low Vision outlines the referral system and resources for children with low vision

Typical Childhood development and developmental milestones

1. Ventura, LO, et al., Visual Impairment evaluation in 119 Children with congenital Zika syndrome, J AAPOS 2018;22:218-222) Infants with CZS present with severe visual impairment. A protocol for assessment of the ocular findings, visual acuity, and visual developmental milestones tested against age-matched controls is suggested

2. Kanlovkids website: example of visual development for a typical developing baby 7 months for parents and teachers https://drive.google.com/file/d/0B34fDbqTRfi9OUdLOG1JMkVKa3M/view

Strabismus surgery in CVI:


Review of strabismus surgical outcomes in CZS, all whom had CVI, addresses quality of life issues


Setting up CVI Clinic


How to set up an effective CVI clinic in a busy ophthalmology practice

Cerebral and Ocular Visual Impairment


Advances in understanding of CVI


Excellent summary of current state of the art


Entire issue based on current understandings of CVI from USA and Europe with multiple authors
Etiologies

Refraction in CVI
   Downloadable book for parents and teachers that explains why we use glasses in babies. Also in Spanish, and soon in Tamil

Other references:
AAPOS website: https://aapos.org/education/educational-resources/pediatric-low-vision-education
CVI (Cerebral Visual Impairment) Resources: For Families
(thanks Terry Schwartz)

1. CVI Scotland: [https://cviscotland.org](https://cviscotland.org). This is a very detailed website, started by a parent of an older child with CVI. A good start (when you have a spare 50 minutes or so) is Dr. Gordon Dutton’s video, which can be found under resources/films and videos ([https://cviscotland.org/mem_portal.php?article=78](https://cviscotland.org/mem_portal.php?article=78)). Parents of older children with CVI, as well as affected teenagers and adults themselves may benefit from the plethora of information available on this website. [www.emeraldeducationsystems.com/news/3310](http://www.emeraldeducationsystems.com/news/3310)
   Parent handbook: [https://cviscotland.org/downloads/CVIBookINT.pdf](https://cviscotland.org/downloads/CVIBookINT.pdf)

2. Parent Support Groups:
   - Start Seeing CVI [https://startseeingcvi.com](https://startseeingcvi.com)
   - CVI momifesto [https://cvimomifesto.com](https://cvimomifesto.com)


4. The Perkins School for the Blind: [www.perkinselearning.org](http://www.perkinselearning.org) (offers on-line courses, lectures and informational sessions)


6. The American Printing House for the Blind: [www.aph.org](http://www.aph.org) (products for students with CVI)

7. The American Conference on Pediatric Cortical Visual Impairment: [www.childrensomaha.org/continuinged](http://www.childrensomaha.org/continuinged) (yearly interdisciplinary conference for professionals and parents, usually held in late June)

8. For parents of children with hemispherectomy for seizure: [www.brainrecoveryproject.org](http://www.brainrecoveryproject.org)

9. Facebook groups:
   - CVI Neuroplasticity Research Group, CVI Phase III Community, CVI Connection

10. Strategytosee.com: Strategy To See’s 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.

11. CVI connect: iPad app for CVI [https://cviconnect.co/](https://cviconnect.co/) CVI Connect is a community of professionals and technology that offer hope and confidence in the form of personalized education for children with cortical visual impairment

12. KanLovKids: website with webinars on children with visual impairment and visual development. [kanlovkids.kssdb.org](http://kanlovkids.kssdb.org)

Resources for medical/educational team

Screening and History questionnaires
(Thanks Terry Schwartz)

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.


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.gestalrevision.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).

Additional Text references:
3. Hyvarinen L, Jacob N. WHAT and HOW Does this Child See? Helsinki, Finland Vistest 2011