Examples of Specific Interventions Based on CVI Characteristics

Characteristic	**Interventions for child with limited function (lower phase)**	**Interventions for child with better function (higher phase)**
Latency	Provide extra time for localization and following of visual stimuli.	Gradually work toward decreasing time needed for localization and following. Provide extra time for completion for educational tasks.
Preference for specific color	Exploit color preference in choice of visual objects of regard.	Use objects of preferred color for ADL. Have teacher, instructor, or therapist wear preferred color in order to maintain interest and localization for following (e.g.: learning to drive motorized chair).
Absent or clumsy visually guided behavior	Use high contrast, lighted, shiny and bright visual objects to grab visual attention. Use sound, vibration and tactile cues to draw visual attention. Be sensitive to overstimulation. It may be necessary to hold	Encourage cause and effect activities in order to motivate child to explore new activities by making something happen. Use bold lines and highlighting to aid child in staying within lines.

	objects further away or use a single sensory stimulation (a lighted toy without sound).	Ensure that child's body is fully supported when performing visual activities.
Difficulty with complexity	Present high contrast objects on black background. Limit distractions. Keep environment simple and uncluttered.	Isolate math problems by presenting one at a time. Limit complexity initially and increase as tolerated based on performance. Keep environment simple and uncluttered. Teach organized scanning of Where's Waldo and Hidden Picture activities.
Improved visual response with movement	Use movement of visual objects to grab visual attention.	Use movement of visual objects to grab visual attention.
Abnormal visual field	Exploit field where child sees better by presenting visual objects in that field. Place stickers or pictures on four corners of tray or desk and teach child to scan the entire	Teach scanning where child turns head to bring objects into field where child sees. Teach child to scan environments before entering them in order to make a mental

	visual field by looking for and identifying all four stickers. Place objects so child will have increased chance of finding it.	picture identifying obstacles so they can be avoided. Elevate visual materials if child has inferior field deficit.
Preference for looking at lights and non-purposeful gaze	Use lighted toys to attract visual attention.	Position child in environment so that there are no distracting lights (next to window, under a ceiling light).
Difficulty with distance viewing	Present visual materials within 1-3 feet.	Gradually increase the distance at which objects are presented. Use other cues to help child locate objects at a distance.
Difficulty with visual novelty	Use familiar objects to grab visual attention. Avoid introducing more than one new object at a time.	Use salient feature of known favored object to introduce new object (color, shape). Describe new objects before introducing them visually to child.
Atypical visual behaviors	Allow child to look away intermittently when attempting to follow visual objects. Do not punish this behavior.	Encourage child to look at objects and maintain gaze when reaching for them.

Cortical/Cerebral Visual Impairment 2023 Update: What You Need To Know

Overview of Clinical Approach

Richard H. Legge, MD
Assistant Professor of Ophthalmology and Visual Science
University of Nebraska Medical Center
Truhlsen Eye Institute

UNMC

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No Financial Disclosures

UNMC

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Relevance

Cortical Visual Impairment (CVI) is the leading cause of visual impairment amongst children in developed countries.

--Chang 2021, McConnell, Nielson, Hattan, Matsuba

The incidence of CVI is increasing.

--Kheptal

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When you first encounter a CVI patient on your clinic schedule, you may think …

YIKES!

4

Our goal today is to convert "yikes!" to …

YIPPEE!

5

So how do we get from

YIKES! → YIPPEE!?

Answer: Have A Plan

6

Effective clinical evaluation of CVI children requires a different approach then what we were classically trained to do.

In short, we need a new plan.
Some parts of the plan we already know.
Some parts of the plan will be new.

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A plan for evaluating Children with CVI.

1. Does this child have CVI?
   The diagnosis needs to be made, or discarded, or suspected.
2. How does this child see?
   There is both visual function, and functional vision to consider.
3. What ocular co-morbidities does this child manifest, and should they be treated?
4. Can I determine the severity of this child's CVI?
   Useful in monitoring for improvement over time.

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A plan for evaluating Children with CVI.

5. Assess the level of CVI assessment, accommodation and intervention that is occurring through the school system, via therapists (OT/PT/Vision Therapy), and at home.
6. Determine and supply for the family the appropriate referrals and documentation to allow for the provision of vision service.
7. Provide additional information resources for the family.
8. Be the child's eye doctor. Support the child's eye care for the long-run.
   This will produce the YIPPEE experience the parents, child and now the doctor, are looking for.

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1. Does this child have CVI? The diagnosis needs to be made, or discarded, or suspected.

MAKE the Diagnosis

Diagnostic Criteria for CVI *(Legge, 2017)*

1. A neurologic diagnosis or condition must be present.
   --Chang 2020, Roman 2007
2. Vision dysfunction must be detected by some method.
   --Chang, 2020
3. Vision dysfunction must exceed what is anticipated based on the ocular pathology present.
   --Chang 2020, Roman 2007
4. >6 months of age (Not published)
   Need to rule out delayed visual maturation/attention
   –C.Hoyt, 1983

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1. Does this child have CVI? The diagnosis needs to be made, or discarded, or suspected.

DISCARD the Diagnosis

Discard the diagnosis if:

-a neurologic diagnosis/condition is NOT present

OR

-vision dysfunction is NOT detected.

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1. Does this child have CVI? The diagnosis needs to be made, or discarded, or suspected.

SUSPECT the Diagnosis (at risk for CVI)

SUSPECT the diagnosis if:

Vision dysfunction is not clearly detected.

OR

It's not clear that the vision dysfunction exceeds that due to a detected ocular pathology

OR

The child is less than 6 months of age, corrected for any prematurity

If a CVI Suspect, follow the child as it will become clearer over time.

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What about using VEP to diagnose Cortical Visual Impairment?

Flash and pattern VEP testing is not useful, unless you are concerned that the child is blind.

Do at >1 year of age to reduce false positives.

This test is not perfect.

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2. How does this child see? There is both visual function, and functional vision to consider.

Visual Function
(we do this)

- -visual acuity
- -stereopsis
- -color vision
- -contrast sensitivity
- -tests of preferential looking
- -peripheral vision

31% of CVI children have at least 1 of these indexes measurable
--Legge, 2016

Functional Vision
(we don't do this)
(More coming soon …Chang,Shah, Lehman)

-novelty, distance viewing, color preference, visual latency, field preference, light gazing and non-purposeful movement, decreased visual-motor skills, decreased visual attention to salient features, lack of eye contact and others

Functional Vision Assessment
-Lueck & Dutton, Kran, others

CVI Range
--Roman 2018

Others
-Pehere 2019, Ravenscroft, Others

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3. What ocular co-morbidities does this child manifest, and should they be treated?

This is our wheel-house.

Refractive error
- treat to prevent amblyopia/PEDIG guidelines *--Beck*
- treat if materially impacting visual function

Accommodative weakness. 12% *--Pehere 2018*
- Consider treating hyperopia <+4.00
  - I commonly treat +1.50 to +3.50
- lined bifocals (dynamic retinoscopy is hard sometimes)

Strabismus *--West* More coming soon … Dr. Melinda Chang

ROP – no change in treatment criteria

Cataract – treat if you think the cataract is contributing to the decreased visual function or functional vision that is detected. Consider post-op risks

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4. Can I determine the severity of this child's CVI? Useful in determining improvement over time.

Ask the parent how the patient is interacting with the environment.
- Phase I – not
- Phase II – is
- Phase III – is finding meaning in the environment

--Adapted from Roman 2007

Obtain the child's Functional Vision Assessment or CVI Range if done by a TVI or OT.

Metrics of CVI severity are not well validated.
This will likely be the next big thing in CVI.
Find something that works for you and use it.

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5. Assess the level of CVI assessment, accommodation and intervention that is occurring through the school system, via therapists (OT/PT/Vision Therapy), and at home.

This can be done by asking the parents:
- Does the child have a vision teacher?
- What has the vision teacher done for the child?
- What kind of technology is being employed by the TVI?

Determine if you think this level of vision care is appropriate.

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5. Assess the level of CVI assessment, accommodation and intervention that is occurring through the school system, via therapists (OT/PT/Vision Therapy), and at home.

Determine if you think this level of vision care is appropriate.

It is important you do this for the following reasons:

1. To know whether they are getting any services at all. If not, print your note and send your recommendation for services home with the family, to be delivered to the school system.

2. If the school system is not the provider of vision services, seek out OTs in your area who have interest and training in CVI. As a physician, you can prescribe OT for vision assessment, accommodations and interventions.

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6. Determine and supply for the family the appropriate referrals and documentation to allow for the provision of vision services.

My dot-phrase:

"Please provide teacher of the visually impaired (TVI) to conduct evaluations, accommodations and interventions to the best of the ability of the TVI."

I put this in the "Plan" section of my clinic note, print it up for the patient and have them hand carry it to their school and therapists.

Much more on this later from Dr. Sharon Lehman.

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7. Provide additional information resources for the family.

https://aapos.org/syndicated/pediatric-low-vision

Many more coming soon …Dr. Sharon Lehman

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8. Be the child's eye doctor

You patient's family wants an eye doctor who understands CVI.

This is now you!

Familiarity matters.

The child becomes comfortable with you over time, allowing a better exam without anesthesia.

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8. Be the child's eye doctor

You will build better understanding of the child's vision over time, through the history the family and TVI gives, along with your examination.

It doesn't all come with one visit.

The needs of the child will change over time, and knowing the child over time will allow you to better meet and anticipate them.

(i.e. after high school planning)

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8. Be the child's eye doctor

It's FUN!

It's JOYFUL!

Just like with your patients without CVI, they grow, mature and accomplish.

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Now you have a plan to get you from YIKES! → YIPPEE!?

YOU CAN DO THIS!

1. Does this child have CVI?
   The diagnosis needs to be made, or discarded, or suspected.
2. How does this child see?
   There is both visual function, and functional vision to consider.
3. What ocular co-morbidities does this child manifest, and should they be treated?
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   Useful in determining improvement over time.
5. Assess the level of CVI assessment, accommodation and intervention that is occurring through the school system, via therapists (OT/PT/Vision Therapy), and at home.
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8. Be the child's eye doctor. Support the child's eye care for the long-run.
   This will produce the YIPPEE experience the parents, child and now the doctor, are looking for.

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'Low Functioning' CVI: Evaluation and Management

Melinda Chang, MD
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AAPOS Workshop: CVI 2023 Update
March 31, 2023

0

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- Saban Research Institute
- Blind Children's Center
- Knights Templar Eye Foundation
- Children's Eye Foundation of AAPOS
- Research to Prevent Blindness
- Ingerman Foundation

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Case Presentation

- 9 month old male referred for 'poor tracking'
  - Also noted to have 'crossed' and 'jumping' eyes
- Born at 35 weeks, urgent C-section, maternal history of intrauterine multi-substance abuse and no prenatal care
- APGAR scores 3 and 8
- NICU for 3 months
  - Extensive intracranial hemorrhage with infarction
  - Seizures
  - Anemia, hyperbilirubinemia, respiratory distress

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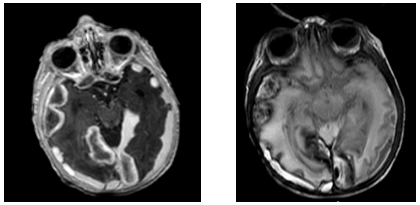
6 days old



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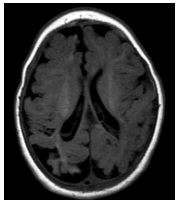
- Current medical problems
  - Encephalomalacia
  - Global developmental delay
  - Seizures
- Medications
  - Levetiracetam
  - Phenobarbital
  - Albuterol
- Family history: unknown
- Social history: foster care



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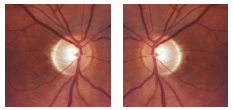
Ophthalmology examination

- Visual acuity
  - Poor fixation OD, OS, OU
  - Could not elicit fixation preference
- Ductions: full
- Krimsky: ET 45 PD
- Nystagmus: conjugate horizontal jerk, no null point
- CRx: +1.00 OU



Representative fundus photos
https://moranocore.utah.edu/basic-ophthalmology-review/optic-atrophy/

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Diagnosis of CVI

Diagnostic Criteria for CVI *(Legge, 2017)*

1. A neurologic diagnosis or condition must be present.
2. Vision dysfunction must be detected by some method.
3. Vision dysfunction must exceed what is anticipated based on the ocular pathology present.
4. >6 months of age
   - Rule out delayed visual maturation/attention

6

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Visual assessment

- Visual function

Grade	Visual Behavior
1	Light perception only
2	Occasional fixation on large objects, faces, or movement
3	Occasional fixation on small objects or reliable fixation on large objects (4-inch lighted toy at 1 foot) or faces, or optotype acuity worse than 20/400
4	Reliable fixation on small objects (2-inch toy at 1 foot), or optotype acuity between 20/400 to 20/200
5	Reliable fixation and pursuit of small objects (2-inch toy at 1 foot), or optotype acuity between 20/200 to 20/50
6	Reliable fixation and pursuit of smallest objects (1-inch toy at least 2 feet away), or optotype acuity between 20/50 to 20/20

Chang MY, Borchert MS. J AAPOS. 2021 Dec;25(6):334.e1-334.e5.

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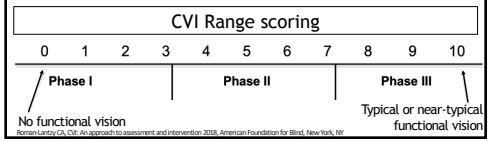
Visual assessment

- Functional vision
  - Not using vision to interact with environment
  - CVI Range: Phase I



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Ocular co-morbidities

- Optic atrophy
- Sensory nystagmus
- Sensory esotropia
- Refractive error: +1.00 OU

9

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Ocular co-morbidities

- Optic atrophy
- Sensory nystagmus
- Sensory esotropia: hold off on surgery until vision improves
- Refractive error: +1.00 OU: no spectacle Rx

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Follow-up

- Visual acuity improved over 1 year
  - Still poor fix OD
  - OS: reliable fixation on small objects
  - OU: grade 4/6 on visual behavior scale
  - Fixation preference OS (strabismic amblyopia OD)

Grade	Visual Behavior
1	Light perception only
2	Occasional fixation on large objects, faces, or movement
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5	Reliable fixation and pursuit of small objects (2-inch toy at 1 foot), or optotype acuity between 20/200 to 20/50
6	Reliable fixation and pursuit of smallest objects (1-inch toy at least 2 feet away), or optotype acuity between 20/50 to 20/20

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Vision Improvement in CVI

- Spontaneous improvement rate: 46-83%
- Factors associated with lack of improvement:
  - Older age at presentation
  - Hypoxic-ischemic encephalopathy, p=0.043
  - ≥3 anti-epileptic medications, p=0.053
  - Cerebral palsy, p=0.029
  - Abnormal fundus examination, p=0.023
- Factors positively associated with improvement:
  - Any therapy (PT/OT/early intervention/glasses), p=0.043

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Jimenez-Gomez A, Fisher KS, Zhang KX, Liu C, Sun Q, Shah VS. Front Hum Neurosci. 2022 Aug 16;16:772353.

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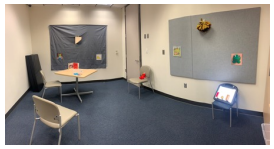
Functional vision assessment

- CVI Range-CR
  - 3 components
    - Interview
    - Observation
    - Direct Assessment
  - Trained examiner
    - TVI or OT
    - Neuropsychologist
  - Scoring: phase 1-3



Chang M, Roman-Lantzy C, O'Neil SH, Borchert MS, *et al BMJ Open Ophthalmology* 2022;7:e001144.

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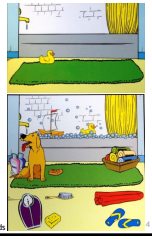
Functional vision assessment

- CVI Range-CR
  - 3 components
    - Interview
    - Observation
    - Direct Assessment
  - Trained examiner
    - TVI or OT
    - Neuropsychologist
  - Scoring: phase 1-3



APH Complexity Cards

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CVI Range scoring

- 10 characteristics

Lower-order visual preferences	Higher-order visual processing	Motor responses
Color preference	Complexity	Latency
Visual field preference	Novelty	Visually guided reach
Preferred viewing distance	Movement	Visual reflexes
Light-gazing		

- 2 methods of scoring
  - Across-CVI Characteristics (score 1)
  - Within-CVI Characteristics (score 2)

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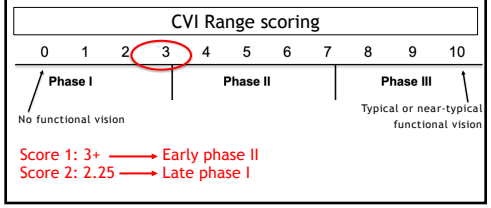
Our patient's CVI Range score



Score 1: 3+ → Early phase II
Score 2: 2.25 → Late phase I

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Additional follow-up

- Fixation preference OS (strabismic amblyopia OD)
  - Started patching OS up to 4 hr/day with good compliance
- 6 months later
  - No fixation preference!
  - Still grade 4/6 on visual behavior scale (binocular)
  - ET stable at 45 PD near and distance, comitant

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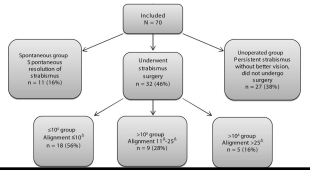
Strabismus in patients with cortical visual impairment: outcomes of surgery and observations of spontaneous resolution

Nicholas R. Binder, MD,[a,b,c] Jacqueline Kruglyakova, BS,[d] and Mark S. Borchert, MD[a,b]

JAAPOS 2016;20:121-125



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Children's Hospital Los Angeles
USC University of Southern California

Ophthalmologic characteristics and outcomes of children with cortical visual impairment and cerebral palsy

Michael R. West, BS,[a] Mark S. Borchert, MD,[a,b] and Melinda Y. Chang, MD[a,b]

(J AAPOS 2021;25:223.e1-6)

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Children's Hospital Los Angeles
USC University of Southern California

- Retrospective study of outcomes in children with CVI with and without cerebral palsy (CP)
- Strabismus
  - 'Good' outcome: horizontal deviation <10 PD
  - Surgical candidates
    - Stable deviation
    - Good control of underlying neurologic condition
    - Visual acuity at least 3 on 6-level scale

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Children's Hospital Los Angeles
USC University of Southern California

Demographics and etiologies of CVI

Table 1. Demographics of children with cortical visual impairment with comorbid cerebral palsy (CVI + CP) and without cerebral palsy (CVI − CP)

	CVI + CP (n = 151)	CVI − CP (n = 153)	*P* value
Age at first visit, years, median (range)	3.0 (0.2-18)	1.5 (0.13-18)	0.0003[a]
Follow-up time, years, median (range)	2.6 (0-12)	1.6 (0-11)	0.09
Male (%)	76 (50.3)	78 (51.0)	0.99

[a]$P < 0.05$.

Table 2. Etiologies of cortical visual impairment in children with comorbid cerebral palsy (CVI + CP) and without cerebral palsy (CVI − CP)

Etiology	CVI + CP, no. (%) (n = 151)	CVI-CP, no. (%) (n = 153)	*P* value
Hypoxic-ischemic encephalopathy	62 (41.1)	40 (26.1)	0.006[a]
Seizures	58 (38.4)	70 (45.8)	0.26
Structural brain malformation[a]	33 (21.2)	31 (20.3)	0.73
Trauma	15 (9.9)	7 (4.6)	0.07
Hydrocephalus	17 (11.3)	10 (6.5)	0.15
Infectious (meningitis, encephalitis)	6 (4.0)	5 (2.6)	0.74
Prematurity with periventricular leukomalacia	25 (16.6)	2 (1.3)	0.000003[a]
Genetic disorder	10 (6.6)	24 (11.1)	0.012[a]
In utero drug exposure	1 (0.7)	2 (1.3)	0.57
Posterior reversible encephalopathy syndrome (PRES)	0 (0)	1 (0.7)	1.00
Multiple etiologies listed above	62 (41.1)	47 (30.7)	0.06

[a]Structural brain malformations include cortical heterotopia, polymicrogyria, schizencephaly, hydranencephaly, lissencephaly, microcephaly, megalencephaly, encephalocele, and holoprosencephaly.
[a]$P < 0.05$.

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USC University of Southern California

Baseline characteristics

Table 3. Ophthalmic characteristics and diagnoses at presentation in children with cortical visual impairment with comorbid cerebral palsy (CVI + CP) and without cerebral palsy (CVI − CP)

	CVI + CP (n = 151)	CVI-CP (n = 153)	*P* value
Visual acuity on six-level scale, median (range)[a]	5 (1 to 6)	4 (1 to 6)	0.06
Significant refractive error[a] (%)	81 (53.6)	63 (41.2)	0.03
Myopia	44 (54.3)	38 (60.3)	0.47
Hyperopia	37 (45.7)	24 (38.1)	0.36
Astigmatism	66 (81.5)	54 (85.7)	0.50
Anisometropia	11 (7.3)	11 (7.2)	0.97
Amblyopia (%)	9 (6.0)	5 (3.3)	0.26
Strabismus (%)	125 (82.8)	111 (72.5)	0.03
Esotropia	40 (32.0)	24 (21.6)	0.10
Exotropia	79 (63.2)	82 (73.9)	
Hypertropia	0 (0)	2 (1.8)	
Combined horizontal and vertical deviation	6 (4.8)	3 (2.7)	
Nystagmus (%)	35 (23.2)	29 (19.0)	0.37
Optic atrophy (%)	70 (46.4)	50 (32.7)	0.01
Congenital optic nerve anomaly[a] (%)	4 (2.6)	4 (2.6)	0.98

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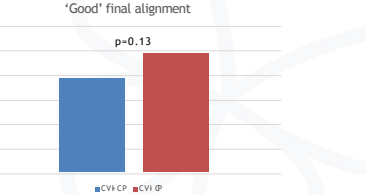
Children's Hospital Los Angeles
USC University of Southern California

Strabismus outcomes



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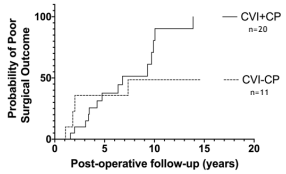
Strabismus surgery outcomes



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Our patient

- Underwent strabismus surgery
  - Right medial rectus recession
  - Right lateral rectus resection
  - Target angle 45 PD
- Orthotropic at POW1!
- 1 year follow-up: consecutive X(T) 20 PD

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Summary

- Diagnose CVI when vision loss is worse than expected based on degree of ocular pathology
  - 'High functioning' CVI may require assessment of visual processing or perception
- Assess both visual function and functional vision
- Majority of patients improve over time
- Strabismus may improve as vision improves
- Strabismus surgery is an option
  - Patients with co-morbid cerebral palsy may have worse outcomes

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'High Functioning' CVI: Evaluation and Management

AAPOS Workshop CVI 2023 Update
March 31st 2023

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Pediatric Neuro-Ophthalmology
Assistant Professor of Ophthalmology and Neurology
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No financial disclosures

2

Case Presentation

CC : "trouble seeing in school"

- 14 y/o girl referred due to difficult time seeing in school
- Certain classes that she has hard time seeing "uncomfortable, overwhelming, crisscrossed vision"
- Certain classes (such as gym) are much more difficult than others
- No other significant PMHx, SHx, or FHx;
  - mild developmental delays early in life, but a normal MRI
- Meds/ Allergies: None

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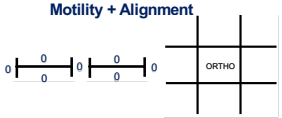
Ocular examination

	OD	OS
VA	20/20	20/20
Pupils	Reactive; no rAPD	Reactive No APD
IOP	13	16
Color	11/11	11/11
VF	Full	Full

Motility + Alignment



- Normal Anterior segment and Dilated Fundus exam

What is the diagnosis ???

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Diagnostic Criteria for CVI *(Legge, 2017)*

1. ~~X~~ A neurologic diagnosis or condition must be present. ***
2. ~~X~~ Vision dysfunction must be detected by some method. ***
3. ~~X~~ Vision dysfunction must exceed what is anticipated based on the ocular pathology present. ***
4. ~~X~~ >6 months of age
   – Rule out delayed visual maturation/attention

Can you have NORMAL vision and neuroimaging and have CVI ???

YES

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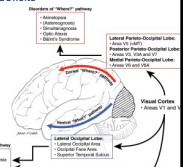
High Vision Functioning Deficits (HVFD) CVI : ***Exception to the rule !

- High Vision Processing/ Cognitive/Vision Perception deficits

Dorsal and Ventral stream pathways
Combination of dysfunction

Spectrum of Agnosias

- Most HF-CVI have neuroimaging findings BUT neuroimaging may be NORMAL!
- HARD to Identify : Low Fxn >> High Fxning CVI!
  → Use R.A.R.E. to spot High Fxning CVI



Haque S, Vaphiades MS, Lueck CJ. The Visual Agnosias and Related Disorders. J Neuroophthalmol. 2018 Sep;38(3):379-392.

6

What to I do as a Pediatric Ophthalmologist or Provider ??

- Recognize : Listen, not hear our patients!
- Ask the Correct Questions!
- Refer the patient !
- Examination for the CVI Dx/Management

7

Back to our patient : Recognition: Listen, not hear our patients!

"uncomfortable, overwhelming, crisscrossed vision "

- She has difficulty walking in the hallways
  - → Can't find a friend in the crowd
  - → Gym class and riding school bus - hard to see
  - - Chaotic, object face recognition and feels lost
- Certain classes are much more difficult than others
  - → Sitting in front – misses a lot of information
  - → While loves drawing, Art Class most difficult
  - - Inability to perceive more than one object at time



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Higher Visual Function Deficits in Children With Cerebral Visual Impairment and Good Visual Acuity

Ask the Correct Questions!

Developed TOP- 11 Screening questions for distinguishing High Functioning CVI vs Normal developing children

1. Does your child have difficulty seeing something that is pointed out at a distance ?
2. Does your child find uneven ground difficult to work on?
3. Does your child bump into things when walking and having a conversation?
4. Does your child have difficulty walking downstairs?
5. After being distracted does your child find it difficulty to get back to get to what they were doing?
6. Does your child have difficulty finding a close friend or relative who is standing in a group?
7. Does your child find copying words or drawings time-consuming and difficult ?
8. Does your child trip at the edges of pavements going down?
9. Does your child find inside floor boundaries difficult to cross?
10. Does your child have difficulty seeing scenery from a moving vehicle?
11. Does your child look down when crossing floor boundaries?

9

Refer the patient !

Multi-Disciplinary Vision Evaluation

Multi-Disciplinary Vision Clinic

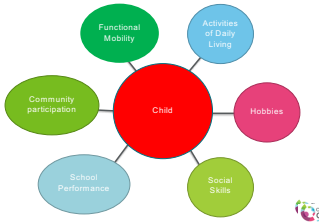
- Ophthalmologist/ Optometrist
- Ophthalmic Technician
- Occupational Therapist
- Teacher of the Visually Impaired (TVI) and/or Education Coordinator
- Clinic Coordinator

Ophthalmology Evaluation → Referral → Occupational Therapy Evaluation

10

Refer the patient !

Occupational Therapists Evaluate and Treat



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Examination for the Dx/Management

Formal Exam/Assessments:

- Beery- Buktenica Visual Motor Integration Development
  - Age 2-100 year-olds
  - 30 item Full form for ages >7 years; 21-item Short form age 2 to 7 years
  - Visual-Motor Integration: The degree to which visual perception and finger- hand movements are well coordinated

Quantify and define the high-order vision deficits

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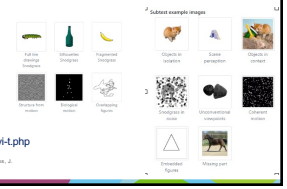
Examination for the Dx/Management

Formal Exam/Assessments : Younger Kids

- Children's Visual Impairment Test for 3-6 year-olds (CVIT 3-6)
  - Computer based visual perception
    - 4 Domains, 14 subtests
      - Object recognition
      - Degraded object recognition
      - Motion perception
      - Global-local processing



Resource:
https://psytests.be/clinicians/test-centrum/cvi-t.php

Vancleef, K., Petré, Y., Janssens, E., Bäumer, S., Oribus, E., & Wagemans, J. (2017). Screening test for cerebral impairment in young children.

13

Back to our patient :

Examination for the Dx/Management

DX: High-Functioning CVI

	Visual Motor Integration	Visual Perception	Motor Coordination
Raw Score	16	20	19
Standard Score	72	83	76
Scaled Score	3	6	5
Percentile	3	13	5
Age Equivalent	5.11	7.4	6.8

She has difficult walking in the hallways
→ Can't find a friend in the crowd
→ Gym class and riding school bus - hard to see
- Chaotic, object face recognition and feels lost

→ Akinetopsia
Prosopagnosia

Certain classes are much more difficult than others
→ Sitting in front – misses a lot of information
→ While loves drawing, Art Class most difficult
- Inability to perceive more than one object at time

→ Simultanagnosia

14

Back to our patient :

Examination for the Dx/Management

Management: Adaptations Catered to the patient

- Environmental Adaptations
  - Position or locate child away from door or part of the room where there are a lot of people moving , or lighted objects / windows with distracting visual stimuli
  - Child given extra time, with NO auditory distractors, to visually process the learned activity (schoolwork tasks that are visually complex)
  - Learned activities earlier in the day due visual latency that may develop as child gets more tired later in the day.
- Adaptations to Reduce Visual complexity during learned activity
  - Consider reducing the complexity of the task or environment
  - Clear clutter away from workspace and /or line of vision to help visually attend

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Revisiting the Diagnostic Criteria for CVI

Diagnostic Criteria for CVI *(Legge, 2017)*

1. A neurologic diagnosis or condition must be present. → Agnosia ✓ ***
2. Vision dysfunction must be detected by some method. → Beery VMI ✓ ***
3. Vision dysfunction must exceed what is anticipated based on the ocular pathology present. ✓ ***
4. >6 months of age
   – Rule out delayed visual maturation/attention

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Take home points to your Clinic

- High functioning CVI patient are HARD to diagnosis! →For everyone
- Pediatric ophthalmologist and Primary vision providers:
  - Recognition: Listen, not hear our patients
  - Ask the Right questions
  - Referral to CVI multidisciplinary team
  - Examination for CVI Diagnosis and Management
- Management: Recommendations and catered therapy for child with High-Order Vision dysfunction and Perceptual deficits

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Cortical/Cerebral Visual Impairment 2023 Update

AAPOS Workshop
March 31, 2023

Sharon S. Lehman, MD

Jefferson
NEMOURS CHILDREN'S HEALTH

1

Disclosure

- I have no financial disclosures.

2

Role of Pediatric Ophthalmologist

- Prompt diagnosis.
- Treatment of underlying medical conditions.
- Treat ophthalmic problems.
- Document medical necessity for vision services
- Prompt referral for services for vision.
- Make specific recommendations.
- Provide education and opportunity for family questions.
- Communicate with team.

3

Role of Pediatric Ophthalmologist

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- Communicate with team.

4

Language to use for documentation:

- List **cortical visual impairment** as diagnosis
- Cortical visual impairment interferes with child's ability to access educational materials.
- Evaluation for vision services medically necessary.
- (Patient's first name) requires direct ongoing evaluation and follow up of a teacher of the visually impaired experienced with cortical visual impairment.

5

Role of Pediatric Ophthalmologist

- Prompt diagnosis.
- Treatment of underlying medical conditions.
- Treat ophthalmic problems.
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6

Refer for Vision services

- Distributed by each individual state
- Each state has its own definition of eligibility based on acuity, visual field or the diagnosis of a chronic condition causing disability
- Educational resources through the state or non-profit organizations support families by providing equipment and educational planning by teacher visually impaired

7

Refer for Vision Services

- Before age of 3:
  - Early intervention program
- After age of 3:
  - School system
- Know the process for obtaining an evaluation for services in your state

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Teacher of Visually Impaired

- assess a child's visual function
- make recommendations for classroom that address the specific characteristics of CVI demonstrated by the child
- work to provide accommodations and equipment
- TVI's prepare:
  - functional vision assessments (FVA)
  - learning media assessments (LMA)
  - assistive technology assessments.

https://familyconnect.org/education/your-childs-educational-team-and-placement/

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Role of Pediatric Ophthalmologist

- Prompt diagnosis.
- Treatment of underlying medical conditions.
- Treat ophthalmic problems.
- Document medical necessity for vision services
- Prompt referral for services for vision.
- Make specific recommendations for use until services start.
- Provide education and opportunity for family questions.
- Communicate with team.

10

Characteristics of Cortical visual impairment

- Color preference and attraction
- Attraction to movement
- Visual latency
- Visual field preference
- Difficulty with visual complexity
- Need for light
- Difficulty with distance viewing
- Atypical visual reflex responses
- Difficulty with visual novelty
- Absence of visually guided reach

Reference: Roman-Lantzy, Chrisitine(2018).Cortical Visual Impairment (2nd edition) AFB.

11

Interventions/Recommendations based on characteristics - Example

Characteristic	Interventions for child with limited function (lower phase)	Interventions for child with better function (higher phase)
Latency	Provide extra time for localization and following of visual stimuli.	Gradually work toward decreasing time needed for localization and following. Provide extra time for completion for educational tasks.

12

Interventions/Recommendations based on characteristics - Example

Characteristic	Interventions for child with limited function (lower phase)	Interventions for child with better function (higher phase)
Preference for specific color	Exploit color preference in choice of visual objects of regard.	Use objects of preferred color for ADL. Have teacher, instructor, or therapist wear preferred color in order to maintain interest and localization for following (e.g.: learning to drive motorized chair).

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Role of Pediatric Ophthalmologist

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- Treat ophthalmic problems.
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- Prompt referral for services for vision.
- Make specific recommendations.
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- Communicate with team.

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Provide education and dialogue for family

- Discuss diagnosis with family
- Provide educational materials to family
  - AAPOS website
  - List of internet resources
- Provide opportunity for family to ask questions

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CVI Resources for families:

- American Printing House for the Blind (APH): http://www.aph.org/cvi/index.html
- American Foundation for the Blind: http://www.afb.org/default.aspx
- American Association of Pediatric Ophthalmology and Strabismus (AAPOS): http://www.aapos.org/
- Family Connect: http://www.familyconnect.org/eyeconditions.aspx?EyeConditionID=6
- Little Bear Sees: http://www.littlebearsees.org/
- National Association of Parents of Children with Visual Impairments (NAPVI): http://www.lighthouseguild.org/programs-services/education/napvi/
- Perkins School for the Blind: http://www.perkins.org/
- Texas School for the Blind and Visually Impaired: http://www.tsbvi.edu/
- Pediatric Cortical Visual Impairment Society: http://www.pediatriccvisociety.org/
- CVI Scotland: https://cviscotland.org/
- Brain recovery Project: http://www.brainrecoveryproject.org/

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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

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Communicate with team

- Use EMR
- Give copy of recommendations and educational materials to family who can share with patient's team
- If family has already accessed vision services, review recommendations of teacher of visually impaired

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Transition of care

- "Risk and vulnerability encompass many dimensions of the transition from adolescence to adulthood, and the transition from pediatric, parent-supervised health care to more independent, patient-centered adult health care is no exception."

Pediatrics Nov 2018, 142 (5) e20182587; DOI: 10.1542/peds.2018-2587;http://pediatrics.aappublications.org/content/142/5/e20182587

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Strategies for improving transition to adult services

- Educate about diagnosis and interventions for CVI
- Develop inventory of needs
- Increase awareness of needs
- Partner with appropriate agencies, educational institutions, occupational agencies

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Strategies for navigating transition

- Start transition planning early
- Experiment with age-appropriate topics
- Involve the patient and move towards independence
- Everyone wants to be heard
- In whatever way the individual can, let he/she answer the questions posed by physician before you jump in
- Encourage transition from pediatric parent supervised to adult patient centered care at early age

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22

Role of Pediatric Ophthalmologist

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