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# Cerebral/Cortical Visual Impairment

“Focus” on Vision Impairment and Blindness  
Conference



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# What is the “visual brain”?

- Eyes capture visual information and transmit it to the striate cortex
- This information is processed
- The interpretation of this information informs
  - Our recognition of elements of the scene
  - How we move our eyes towards items of interest and attend to these items
  - Visually guided movements of our body
- The interpretation needs to be continuously updated and processed

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# Cerebral or cortical visual impairment

- Visual impairment that cannot be attributed to pathology within the eye or the anterior visual pathways
  - Retro-geniculate pathway injury which may include
    - Optic radiations
    - Occipital cortex
    - Higher order visual association centers
  - Ocular injury and cortical visual impairment may coexist
  - The diagnosis is a clinical one and requires a multidisciplinary approach
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# Epidemiology of CVI

- Leading cause of significant visual impairment in children in developed countries
  - Study of 3070 US children attending schools for the blind

Diagnosis	1999		2012	
	No	% of total	No	% of total
Cortical visual impairment	515	20	556	18
Optic nerve hypoplasia	166	7	445	15
ROP	320	13	442	14
Optic Atrophy	124	5	211	7
Albinism	140	5	126	4
Coloboma	26	1	86	3

- UK population based study of children with profound vision loss: 48% CVI

# Common Etiologies of CVI

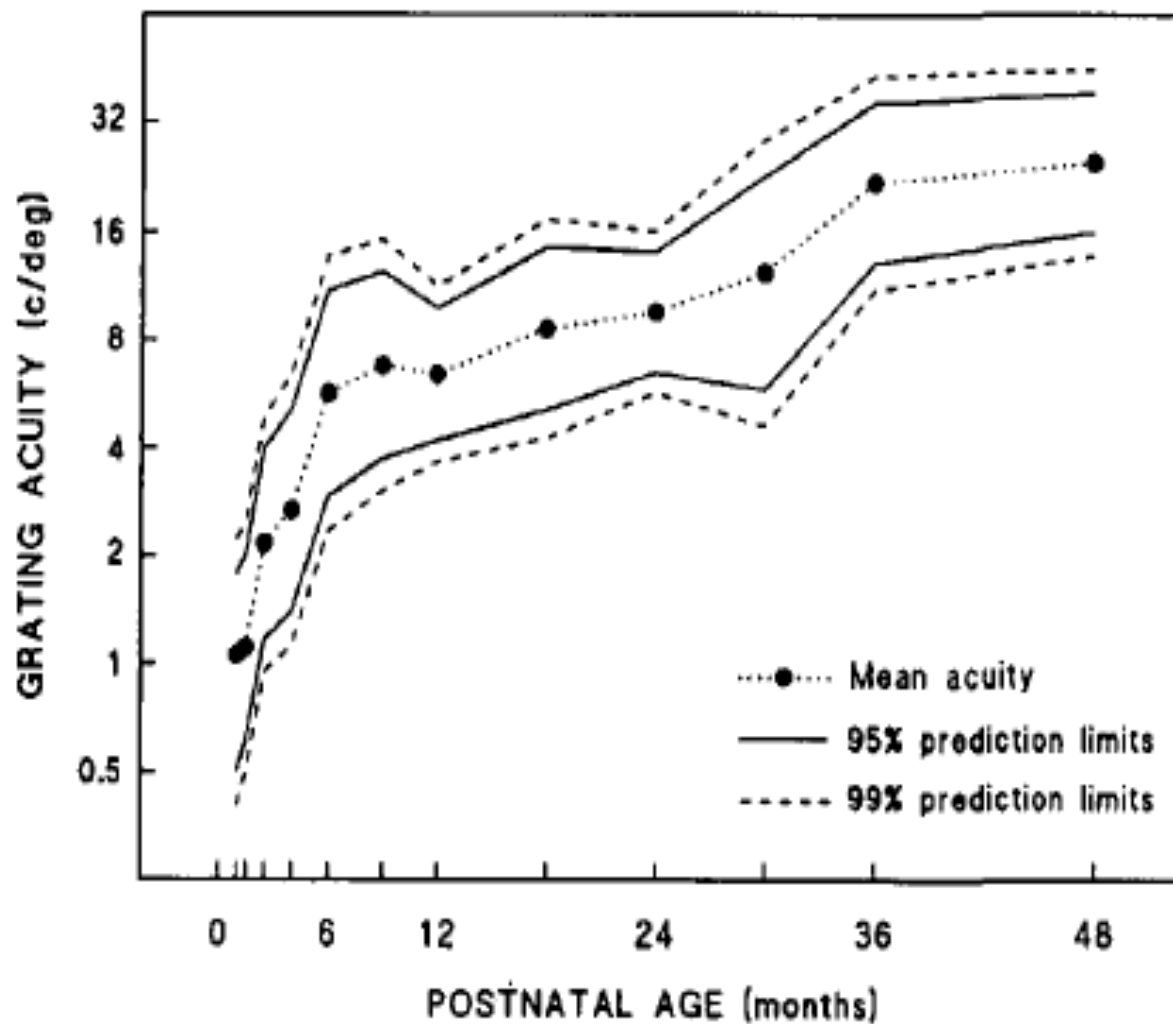
- Perinatal hypoxia/ischemia
    - Term infant there is injury to the cortical watershed areas: parieto-occipital infarction
  - Periventricular leukomalacia (hypoxic/ischemic)
    - Premature infant there is injury to the subcortical visual pathways: this affects the optic radiations
  - Head injury/ trauma
  - Hydrocephalus
  - Infection (e.g. encephalitis, meningitis)
  - CNS malformations
  - Metabolic disorders
  - Neurodegenerative disorders
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# Co morbidities in the Setting of CVI

- Cognitive deficits/delay
- Seizures
- Motor deficits/cerebral palsy
- Sensory deficits (hearing)

# Normal Grating Acuity



# Visual Development is Impaired in CVI

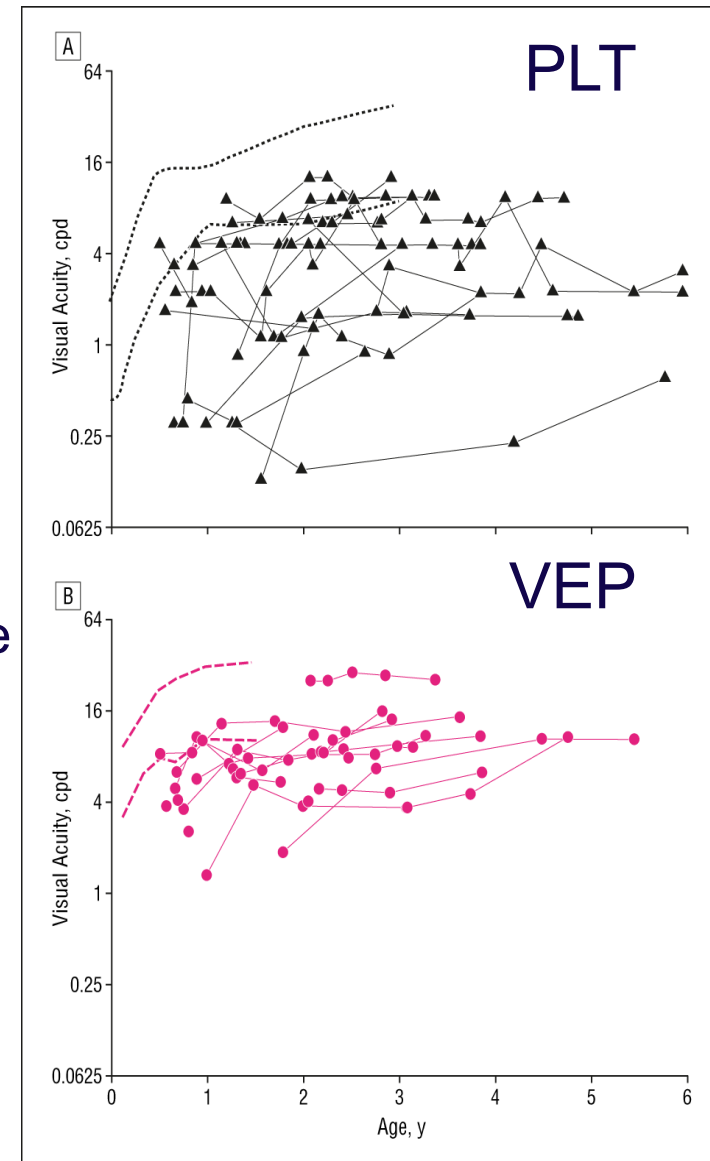
## ■ Study of vision in CVI

### □ 19 term babies

- Hypoxic-ischemic encephalopathy
- Seizures
- MRI-confirmed injury

### □ Results

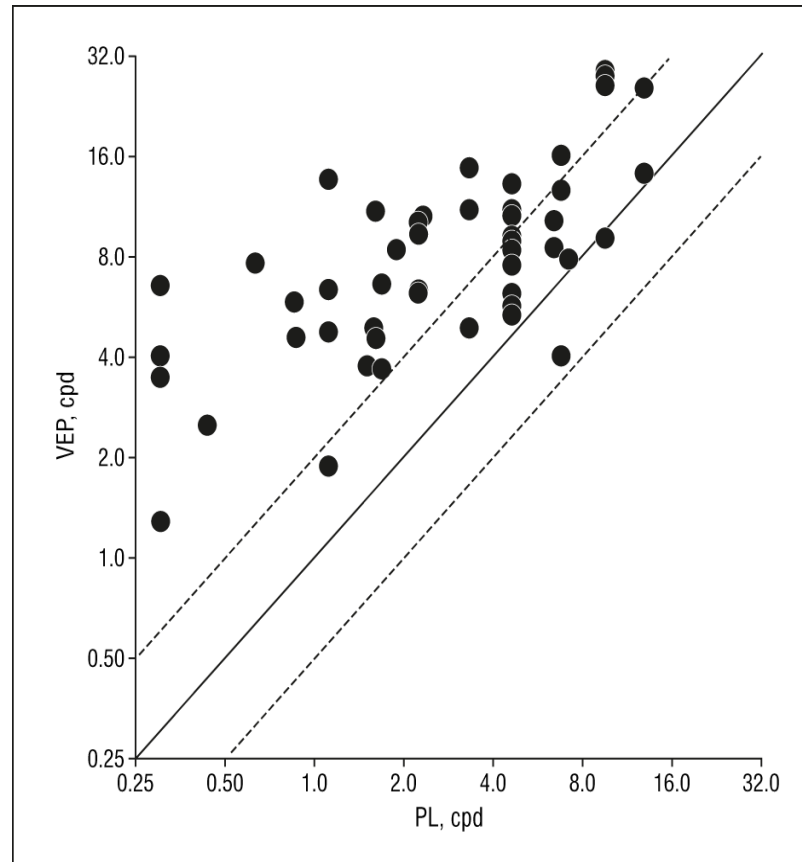
- PL improved 1 octave on average
- VEP improved 1 octave on average





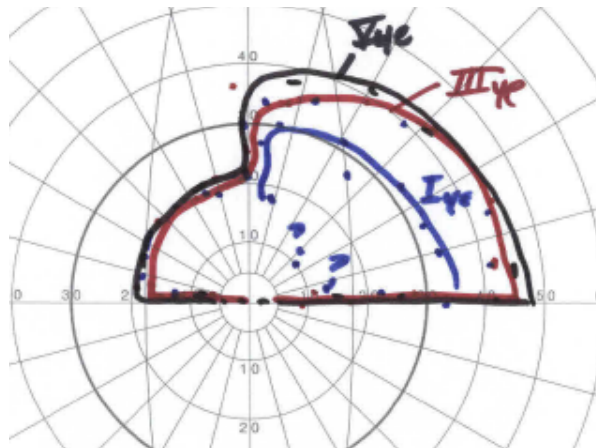
# Visual Development is Impaired in CVI

- VEP and PLT results are discrepant in CVI
  - Normally VEP acuity may measure better than PLT by ~1 octave
  - In most CVI patients, VEP acuity abnormally exceeded PLT

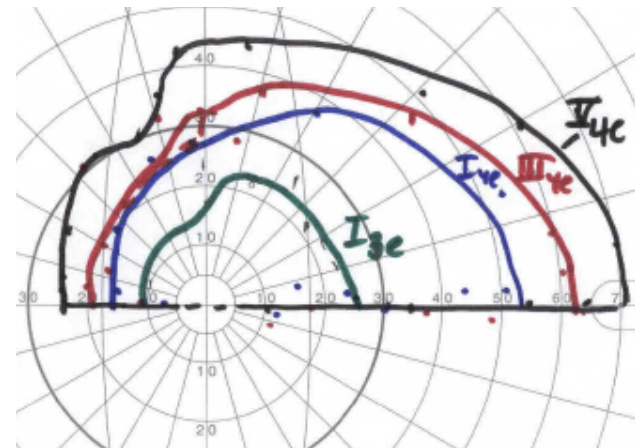


# Visual Fields may be Affected in CVI

- Homonymous visual field loss
- Bilateral inferior visual field loss in the setting of PVL



OS



OD

# Efferent Dysfunction is Common

- Strabismus (range 22%-73%)
  - 70 patients with CVI and strabismus
  - 16% had spontaneous resolution

Table 2. Strabismus type<sup>a</sup>

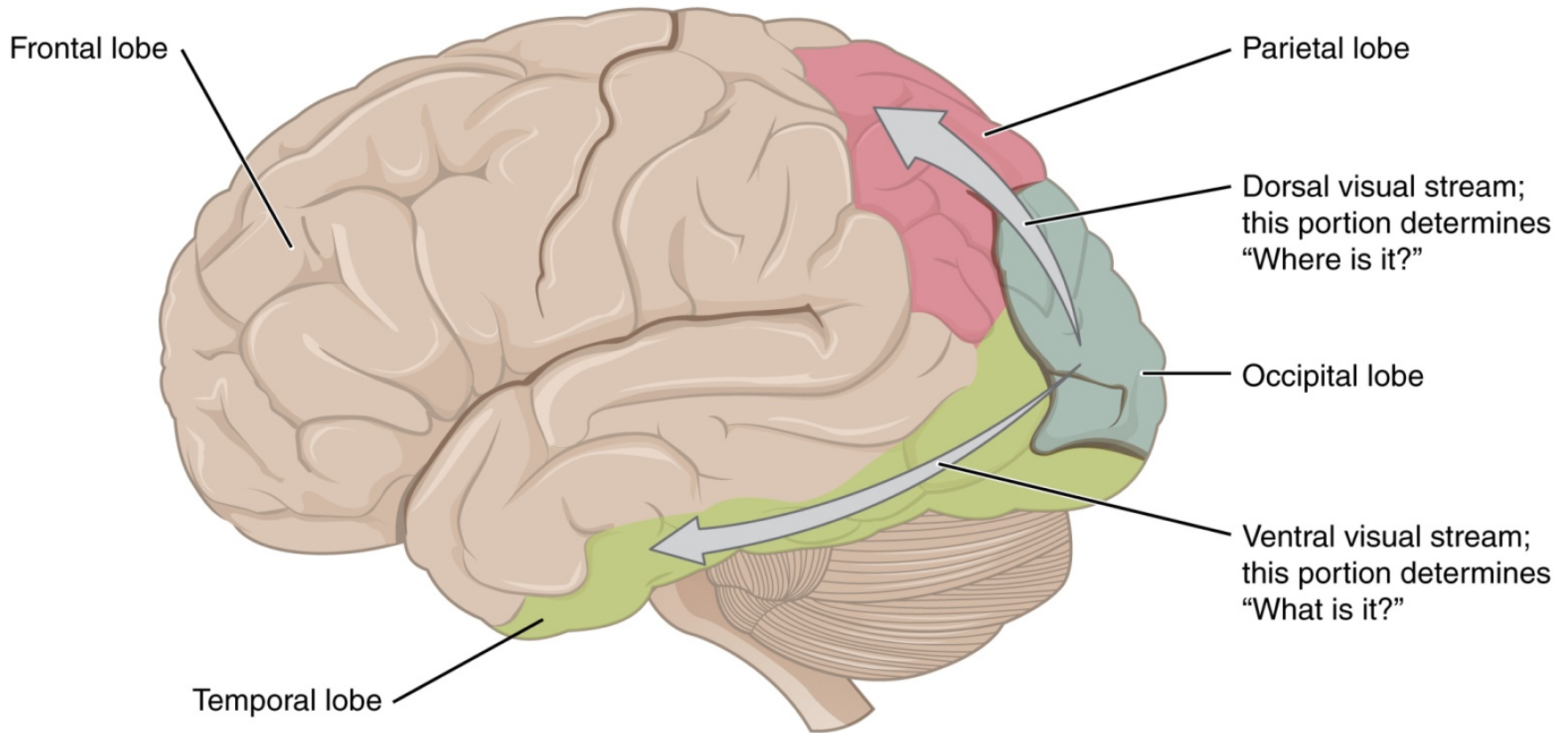
Outcome category	ET	E(T)	XT	X(T)	All ET	All XT
Spontaneous resolution	2 (18%)	1 (9%)	6 (55%)	2 (18%)	3 (27%)	8 (73%)
Unoperated	6 (22%)	2 (7%)	15 (56%)	4 (15%)	8 (30%)	19 (70%)
Operated						
Final ≤10 PD	6 (33%)	2 (11%)	7 (39%)	3 (17%)	8 (44%)	10 (56%)
Final >10 PD	6 (43%)	0 (0%)	6 (43%)	2 (14%)	6 (43%)	8 (57%)
All	20 (29%)	5 (7%)	34 (48%)	11 (16%)	25 (36%)	45 (64%)

ET, constant esotropia; E(T), intermittent esotropia; XT, exotropia; X(T), intermittent exotropia.

<sup>a</sup>Four patients had a vertical strabismus in addition to horizontal, but in these cases the primary deviation was horizontal. No statistically significant difference was found in the types of strabismus present between groups.

- Nystagmus
- Gaze palsy
- Difficulty with saccades

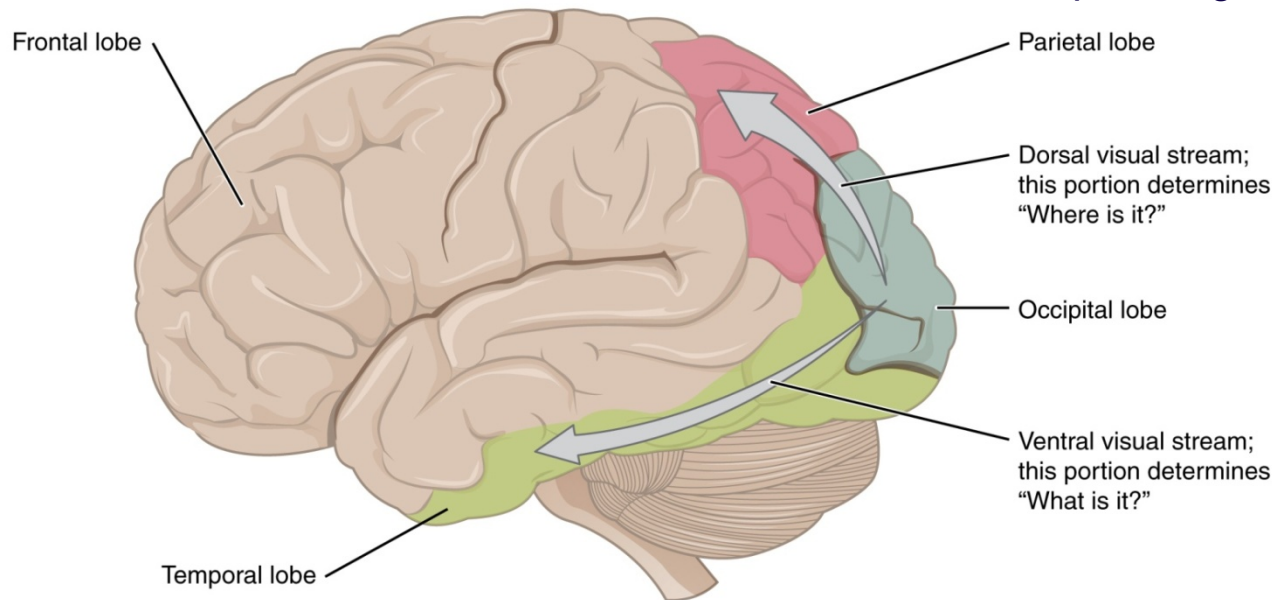
# Two Stream Hypothesis of Visual Processing



# Impact of CVI on Visual Processing

## Dorsal Stream Dysfunction

Difficulty with visual complexity  
Inability to sustain visual attention  
Difficulties with visual-motor planning



## Ventral Stream Dysfunction

Difficulty with recognition of objects or faces  
Difficulty with visual memory

# Behavioral Characteristics

- Light gazing
- Gaze avoidance
- Use of peripheral vision to locate an object
- Increased latency of visual response
- Difficulty with visual complexity
- Difficulty with visual novelty
- Proclivity for a color to see
- Use of movement to see
- Difficulty with distance vision
- Variability of visual attention
  - Exacerbated by concurrent auditory or tactile stimuli

# Elements of the CVI Assessment

- Ophthalmology visit
- Functional vision assessment
- Low vision assessment
- Involvement of a teacher for visual impairment
- Additional services
  - OT, PT, Orientation and Mobility
- Input from multidisciplinary team
  
- Tools for assessment of CVI
  - Christine Roman-Lantzy CVI Range
  - Gordon Dutton or Els Ortibus CVI inventories

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# The Anatomy of CVI

- What is the neuro-anatomic basis of CVI?



# Case 1

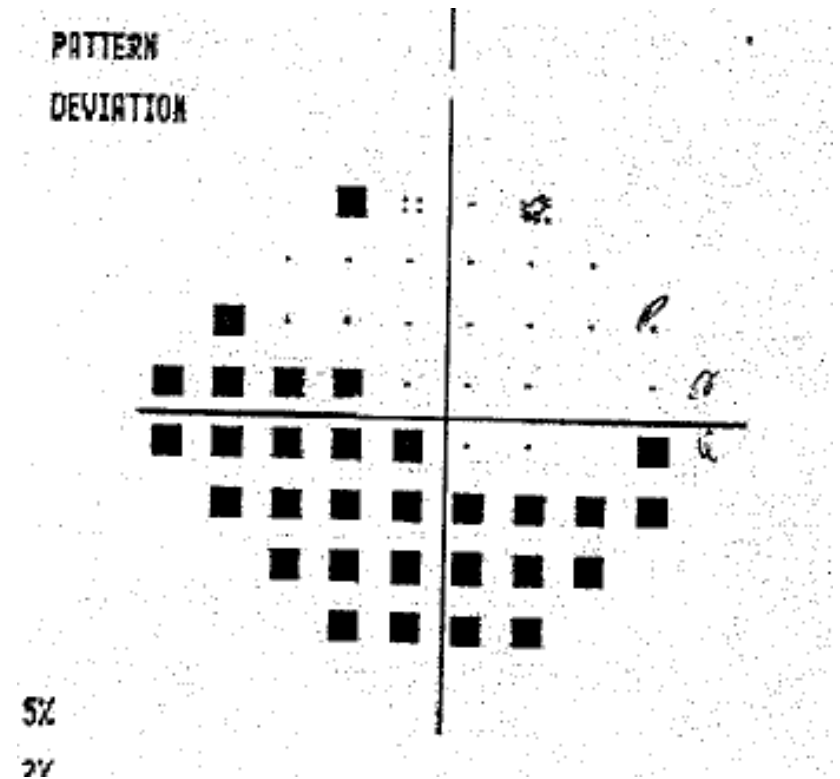
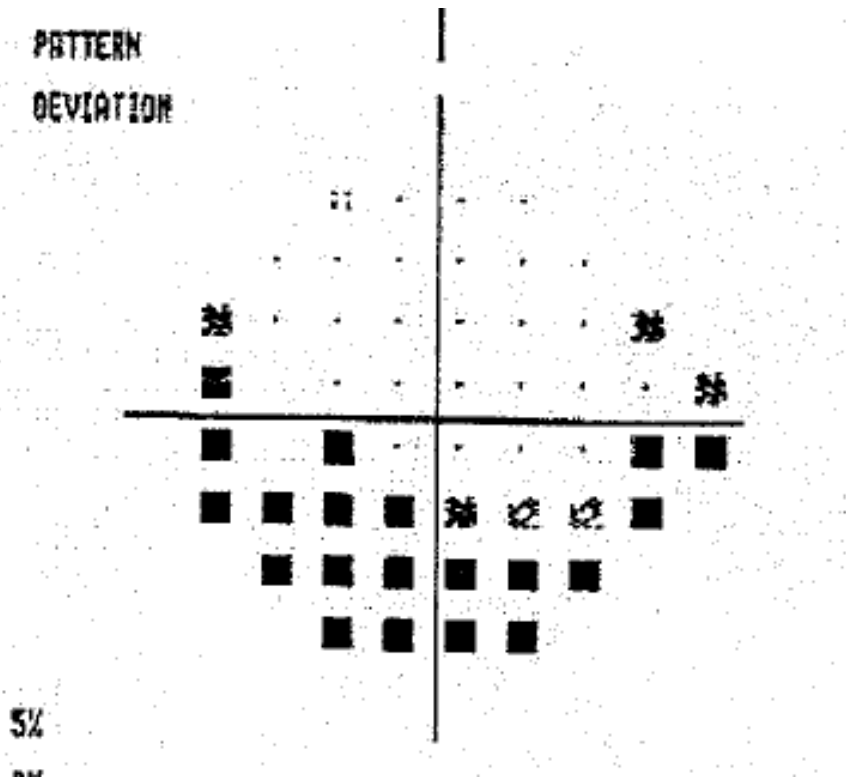
16 year old girl with difficulty in school characterized by tracking issues, visual inattentiveness, and hardship with complex images particularly interpreting those on a grid.

Birth History	<ul style="list-style-type: none"><li>• Born at 32 weeks gestation</li><li>• Developed grade III intraventricular hemorrhage and subsequent hydrocephalus</li><li>• Diagnosed with periventricular leukomalacia</li></ul>
Ocular History	<ul style="list-style-type: none"><li>• Strabismus surgery at 11 months for esotropia</li></ul>
Medical History	<ul style="list-style-type: none"><li>• Spastic diplegia</li></ul>

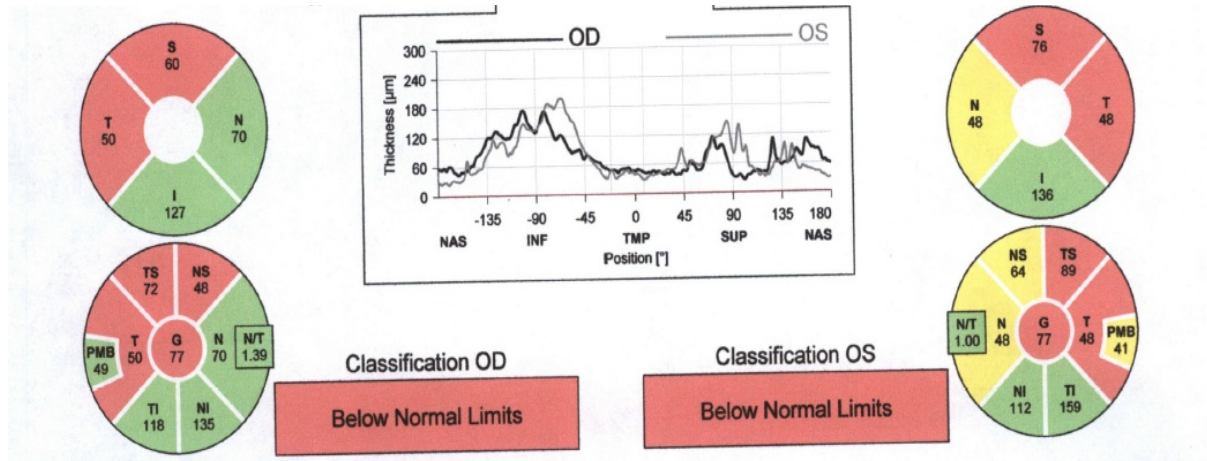
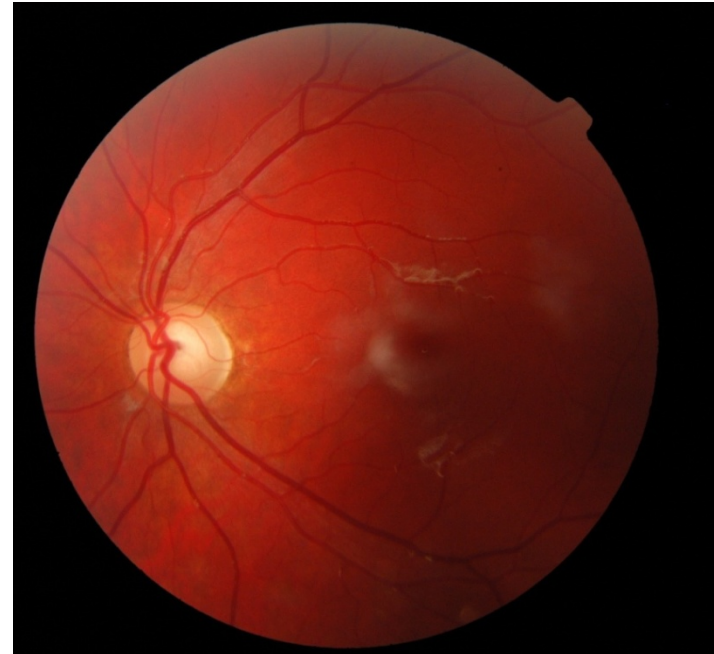
# Examination

	Right Eye	Left Eye
Visual acuity	20/50	20/40
Cycloplegic refraction	Mild hyperopic astigmatism	
Pupils	Normal with no relative afferent pupillary defect	
Contrast sensitivity	Reduced	Reduced
Anterior segment	Normal	Normal

# Humphrey Visual Fields 24-2



# Fundus Photos and OCT



# Case 2

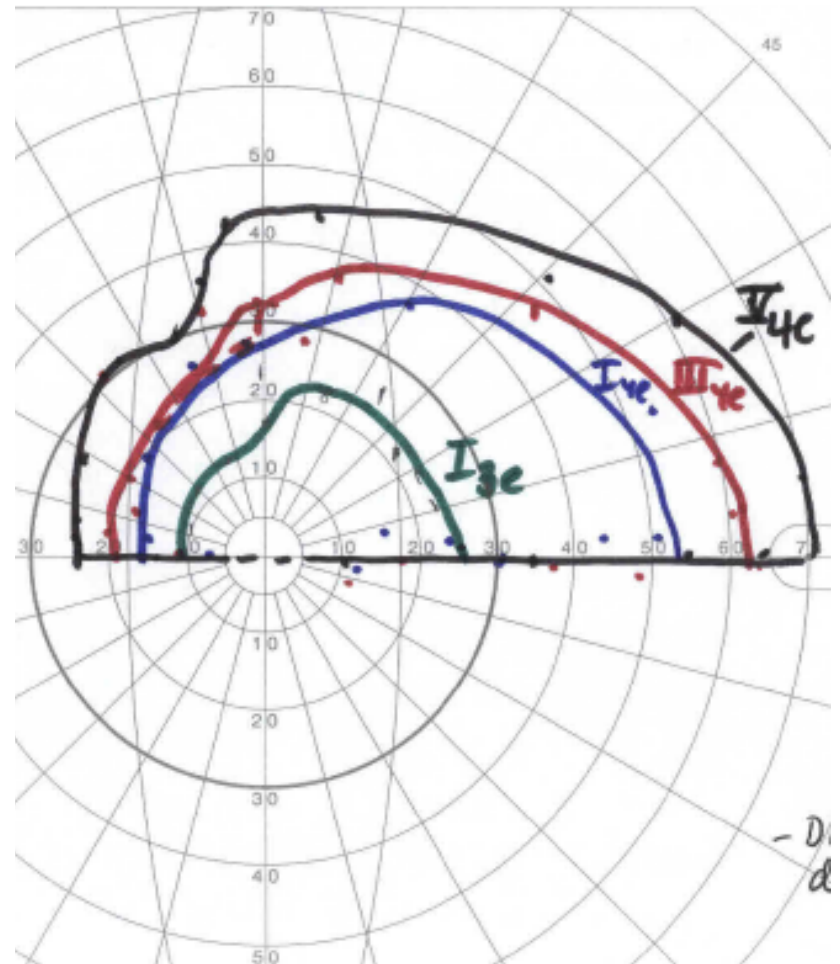
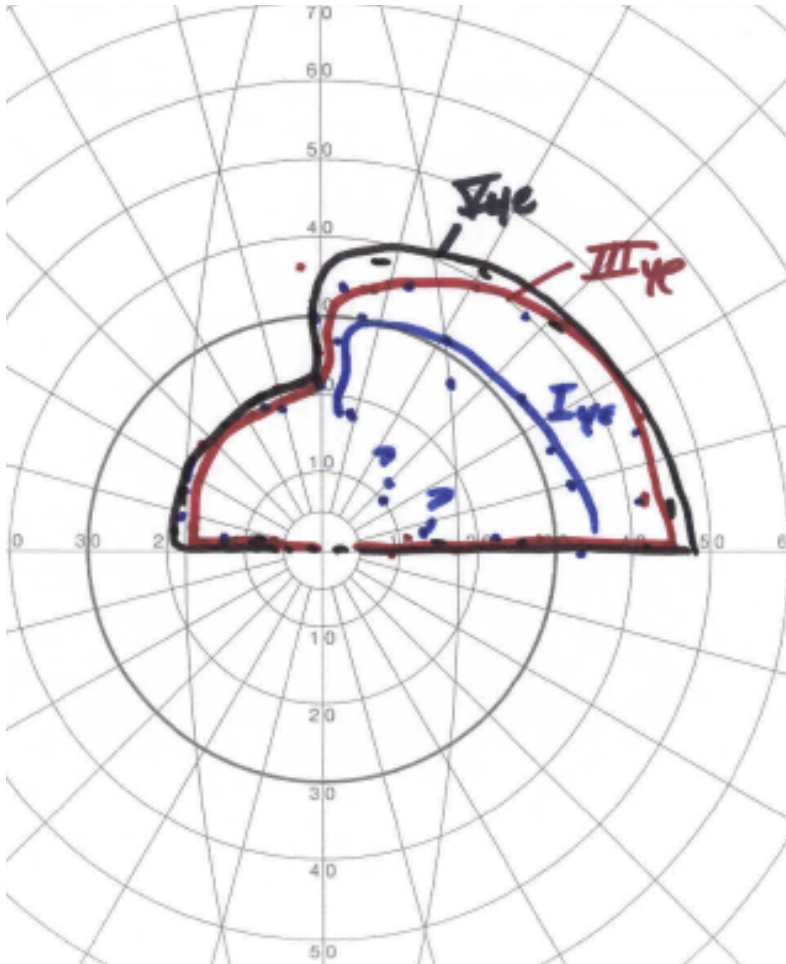
22 year old man with significant cognitive delay. Visual difficulty characterized by visual inattentiveness and inability to identify objects in a crowded scene.

Birth History	<ul style="list-style-type: none"><li>• Born at 28 weeks gestation</li><li>• Developed perinatal brain hemorrhage</li><li>• Diagnosed with periventricular leukomalacia</li></ul>
Ocular History	<ul style="list-style-type: none"><li>• Strabismus surgery before age 3 for esotropia</li></ul>
Medical History	<ul style="list-style-type: none"><li>• Marked developmental delay</li><li>• Cerebral palsy</li></ul>

# Examination

	Right Eye	Left Eye
Visual acuity	20/100	20/100
Cycloplegic Rx	+1.50+1.25 x 15 OD +1.50+1.25 x 175 OS	
Pupils	Normal with no relative afferent pupillary defect	
Contrast sensitivity	Reduced	Reduced
Anterior segment	Normal	Normal
Sensorimotor	Full motility Manifest fine amplitude nystagmus Consecutive exotropia	
Posterior segment	Optic atrophy bilaterally	

# Goldmann Visual Fields



# Hypothesis: CVI is a disease of abnormal neuronal connectivity

- Pilot study to map major white matter connections between visual cortex and higher order visual association centers using HARDI (high-angular-resolution diffusion imaging)
  - Patients 14-21 years of age
  - Corinna Bauer, PhD and Lotfi Merabet, OD, PhD

Superior longitudinal fasciculus (SLF)

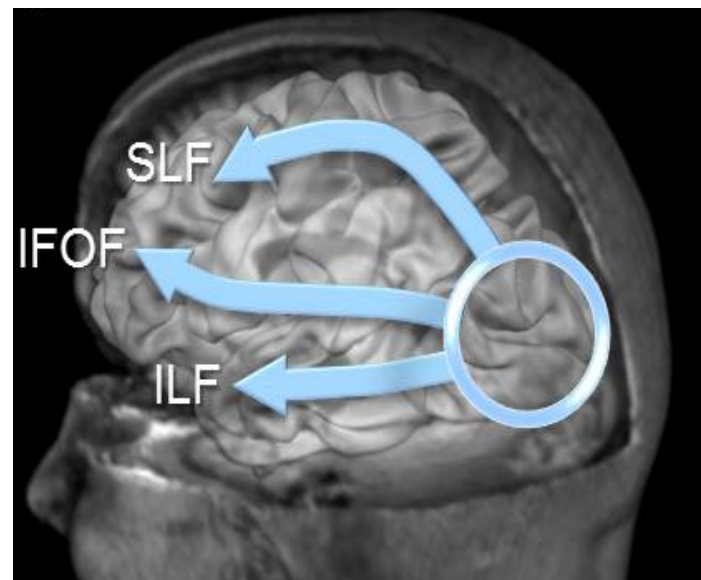
-occipital-parietal: visual complexity, visual attention

Inferior fronto-occipital fasciculus (IFOF)

-occipital-ventral frontal: visual attention, eye movement control

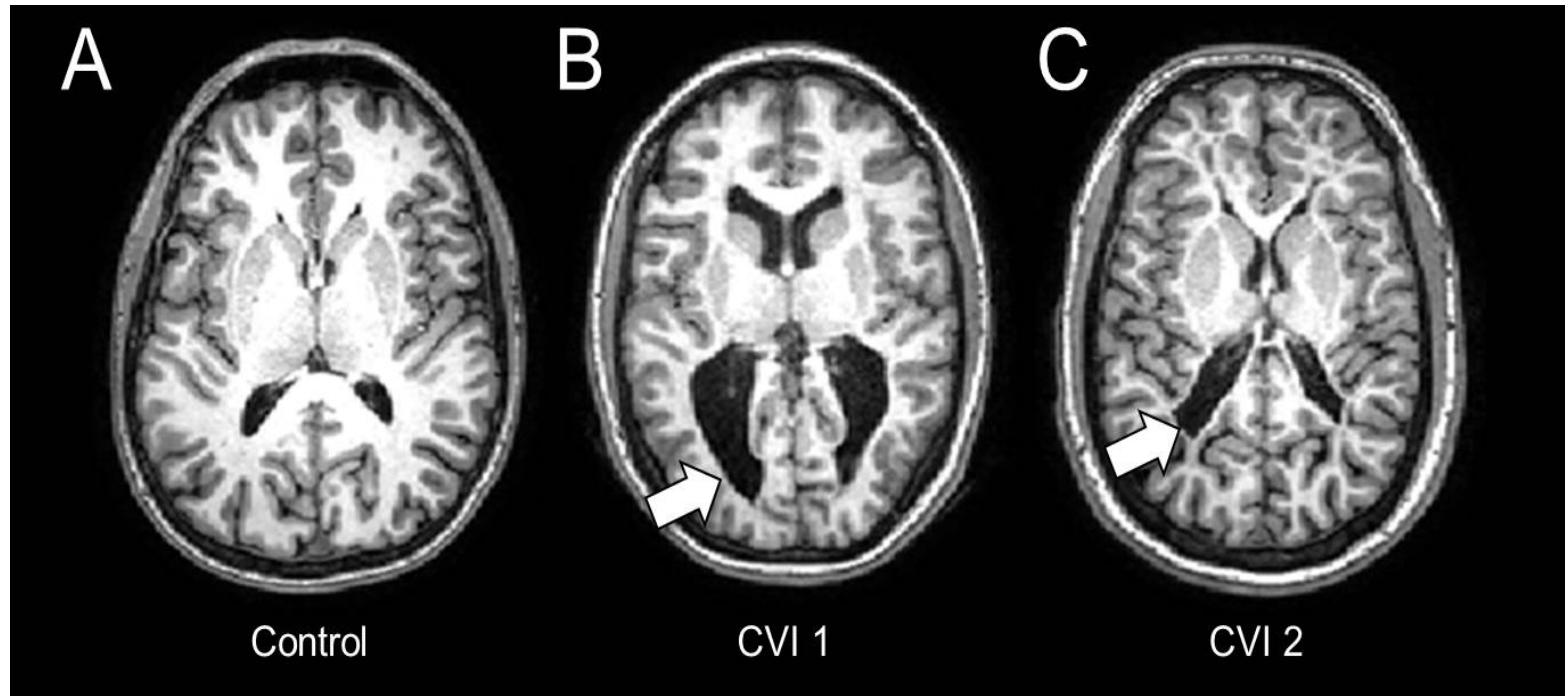
Inferior longitudinal fasciculus (ILF)

- occipital-temporal: object vision, color, faces





# Axial T1 weighted MRI Images

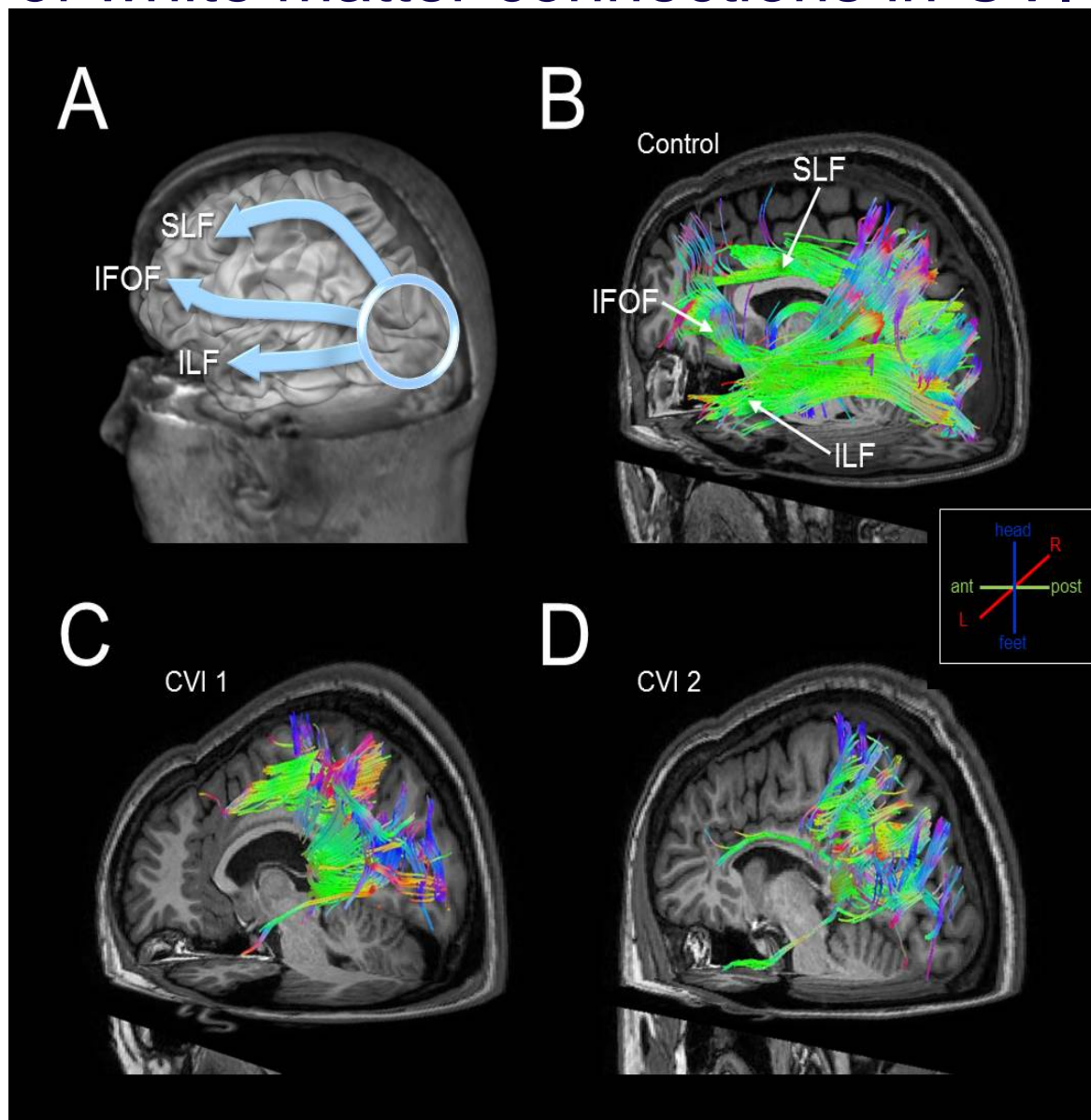


Control

Case 1  
Preterm 32 weeks  
PVL

Case 2  
Preterm 28 weeks  
PVL

# Paucity of white matter connections in CVI patients



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# The Anatomy of CVI

- CVI may be secondary to abnormal connectivity amongst vision association centers
- May we use this understanding to develop an individualized approach towards habilitation or an improved educational plan for our CVI patients?

# Ophthalmologist's Role in CVI

- Maximize vision function
  - Prescribe glasses when relevant
  - Manage strabismus when relevant
  - Specifically evaluate visual fields to determine whether there is visual field loss
- Recognize CVI and initiate appropriate referrals
  - Low vision assessment
  - Functional vision assessment
  - Ensure patient has a teacher for visual impairment
  - Register with State Services

## MA Commission for the Blind

using an ETDRS chart, they would not be classified as legally blind because they were able to read one letter on the 20/100 line. If acuity testing is problematic due to a vision condition such as cortical visual impairment, please answer the optional question below.

Standard acuity testing is impossible or unreliable and, in my medical opinion, the functional vision is at the level of legal blindness as defined above?

Yes

No

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# Summary

- CVI is the leading cause of childhood blindness in developed countries
  - The condition is likely underreported
  - There are visual and behavioral findings which characterize CVI
  - A multidisciplinary approach towards evaluation and treatment is essential
  - Recognition of CVI will facilitate our children receiving the appropriate services to maximize visual function
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