# AIOC 2026 – Instruction Course

# Introduction to Cerebral Visual Impairment (CVI), Visual Characteristics of CVI Children and Red Flags in Diagnosis

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## 1. Introduction to CVI

**Cerebral Visual Impairment (CVI)** is a **brain-based visual disorder** caused by damage or dysfunction of the retrochiasmal visual pathways, **without sufficient ocular pathology** to explain the visual deficit.

- Now one of the **leading causes of childhood visual impairment**
- Results from impaired **visual processing in the brain**, not the eyes

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## 2. Common Causes

- Hypoxic ischemic encephalopathy (HIE)
- Prematurity with periventricular leukomalacia (PVL)
- Hydrocephalus
- CNS infections
- Seizure disorders
- Congenital brain malformations
- Traumatic brain injury

**Frequently Associated With:**

- Cerebral palsy
- Epilepsy
- Global developmental delay

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## 3. Pathophysiology (What Gets Affected?)

Damage may involve:

- Optic radiations
- Occipital cortex
- **Dorsal stream ("Where" pathway)** – spatial processing
- **Ventral stream ("What" pathway)** – object recognition

👉 **Dorsal stream dysfunction is common in children.**

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# 4. Visual Characteristics

### General Features

- Inconsistent visual responses
- Delayed visual latency
- Preference for moving objects
- Light attraction
- Visual fatigue
- Better performance in familiar settings
- Difficulty in cluttered environments
- Visual acuity may be near normal but functional vision impaired

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### Dorsal Stream–Related Signs

- Difficulty locating objects in clutter
- Problems with navigation (stairs, crowded spaces)
- Inaccurate reaching (optic ataxia)
- Reduced spatial awareness
- Simultanagnosia

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### Ventral Stream–Related Signs

- Difficulty recognizing faces
- Impaired object identification
- Problems with shapes and symbols

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# 5. Red Flags for CVI

Suspect CVI when:

- Visual behavior is worse than ocular findings
- Anterior segment & fundus exam are normal/minimally abnormal
- Vision fluctuates
- Child struggles in visually complex environments
- Strabismus without clear ocular cause
- Persistent poor fixation despite correction
- Presence of CP, seizures, or developmental delay

⚠️ **A normal eye exam does NOT exclude CVI.**

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# 6. Clinical Approach

Evaluation should include:

- Detailed perinatal & developmental history
- Functional vision assessment
- Cycloplegic refraction
- Comprehensive ocular exam
- Behavioral visual fields
- MRI brain (when indicated)

👉 **CVI is primarily a clinical diagnosis supported by neuroimaging.**

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# Key Take-Home Messages

- CVI is a **brain-based visual disorder**
- Ocular exam may be normal
- **Dorsal stream dysfunction is common**
- Early identification + multidisciplinary intervention improves outcomes

**TOPIC**: The tools used for visual acuity, field and functional assessment in CVI ---Soveeta Rath

Introduction:

Cerebral visual impairment was defined as visual impairment due to retrogeniculate visual pathway pathology, as determined by clinical assessment and neuroradiological evidence. It has become the leading cause of visual impairment in developing countries, with the increase in survival of premature infants and infants with perinatal insult and metabolic disorders. The assessment of these children is crucial yet challenging as they are also associated with neurological deficits that affect their cognitive and motor abilities.

## FEATURES AND ASESSMENT :

The major ophthalmic manifestations of these children include **poor visual behaviour** where children do not fix on light, do not recognise their mother and are unable to grab an object.

Nystagmus, strabismus, and leukocoria are also noted in these children like children with ocular visual impairment(OVI).

However, the basic assessment in these varies and needs a clutter-free room and specific tools, including a Lea paddle, Lea mailbox, etc. The vision assessment has been described by Huo et al in 6 levels, wherein level 1 starts with only light perception and ends with level 6 wherein the child has a normal sensory visual examination.

Cycloplegic refraction is a must for these children who have at least attained neck control and can support glasses, which will improve the clarity of their near work. Cyclopentolate is avoided in these children with neurological issues. It has been seen that CVI children with a history of prematurity have myopia, whereas others are hypermetropic.

The majority of these children have accommodation insufficiency, which emphasises the role of dynamic retinoscopy in them and benefits from bifocals.

Studies have also demonstrated a reduction of colour and contrast sensitivity in patients with developmental delays. Hiding Heidi contrast cards is an important tool in assessing contrast in these children

Assessment of the visual field using a pupillary light reflex or a Lea wand is essential, as many prefer looking up, suggesting an inferior field defect.

Apart from these dysmetric saccades, deficient smooth pursuit movements are seen in a substantial group of children.

Strabismus has a characteristic shifting trait in children with CVI. They may have exotropia, esotropia, and dissociated vertical deviation.

The angle and the type of strabismus vary in different episodes during the examination in some of these children, whereas in others, it is a constant deviation. Though esotropia has been noted as the most common type of strabismus in these children, exotropia in children with neurological associations is on the rise. Repeated orthoptic evaluation with consistent measurements is necessary before planning surgical correction in children with strabismus.

On fundus examination children with early periventricular leukomalacia (PVL) (<28 weeks)- have small discs, and optic nerve hypoplasia, whereas those born after 28 weeks have normal discs with large cups called as pseudo glaucomatous discs or PVL discs also.

Children with PVL affecting the cortex have a horizontal conjugate deviation, whereas those with subcortical lesions prefer a tonic downgaze.

**Functional vision assessment** is essential to know the practical implications to initiate rehabilitation therapy. The first period of a functional assessment is simply to watch the use of vision, and depending on the child's comfort, begin simple interactions to observe the impact of different factors. **Visual attention, visual robustness, tracking and scanning, shifting gaze, colour and contrast sensitivity, visual fields, light sensitivity, how the eyes work together, and visual stamina are the key pointers while evaluation.**

Various questionnaires are designed for functional assessment by Dutton and Roman Lantzy. These give us a quantification apart from the quality of functional vision, which further helps the role of therapy. Visual functioning in CVI is measured across the Range (0-10), with three major phases describing level of impact of the CVI Characteristics on the individual's ability to use *his/her* vision functionally:

- 0-3, Phase I: Most severe impact on visual functioning
- 3-7, Phase II: Moderate impact on visual functioning
- 7-10, Phase III: Closest to typical visual functioning

Thus, it is imperative to detect and treat these children with early intervention. Since it not only affects the functional vision, it also retards the overall psychomotor, social and emotional development of the child. A comprehensive assessment of these special children should be included in our training programs.

# Higher Vision function deficits and MRI findings in CVI children

Cerebral visual impairment (CVI) is brain-based vision impairment caused by damage to retrogeniculate pathways in the absence of anterior afferent visual pathway or ocular structure damage. CVI is characterized by abnormal visually guided behaviors indicating higher vision functions deficits that can occur even in the presence of good visual acuity and so, might remain undiagnosed.

The higher vision functions can be explained by cerebral network consisting of dorsal stream and ventral stream. Dorsal stream or 'where' stream connecting occipital, parietal lobe, middle and superior temporal gyrus is responsible for spatial localization, movement and spatial relations while ventral stream or 'what' stream connecting occipital and inferotemporal gyrus is responsible for recognition of faces, objects, colors and shapes. However, there is overlap between the two streams in implementing most of the visual functions.

Characteristic abnormal visually guided behaviors typically seen in CVI children:

1) Visual latency
2) Light gazing
3) Visual field preference
4) Color preference
5) Preference for motion clues or tactile clues
6) Difficulty in visual complexity
7) Difficulty in visual novelty
8) Absence of visually guided reach

CVI is caused by brain insult during perinatal period or of late onset. The common causes are perinatal hypoxia, neonatal hypoglycemia, epilepsy, traumatic brain injury, hemorrhagic lesions, cerebral dysgenesis leading to defective neuronal migration.

MRI is most sensitive modality in assessing the extent and nature of injury and thus prognosticating the outcome and planning rehabilitation therapy. Pattern of injury depends on brain maturity at the time of insult, severity, and time of imaging.

The commonest risk factor for CVI is hypoxic brain injury that leads to infarction in the watershed areas. In preterm infants, ventriculopetal penetrating arteries supply periventricular region therefore watershed areas are located in periventricular white matter leading to periventricular leukomalacia (PVL)in preterm infants. While in term infants, ventriculofugal arteries extend from lateral ventricles and watershed zone moves peripherally in parasagittal location leading to involvement of subcortical white matter and parasagittal cortex in term infants. Profound hypoxic injury in the form of acute severe asphyxia in term infants leads to basal ganglia and thalamus injury and altered signal intensities in these areas.

Neonatal hypoglycemia particularly shows most commonly white matter abnormalities with parietooccioital distribution of lesions.

Common MRI findings:

1) PVL: The commonest MRI finding is PVL which is seen as white matter hyperintensity adjacent to lateral ventricle or scattered punctate white matter abnormalities.
2) Irregular ventriculomegaly with dilatation trigones and occipital horns of lateral ventricles.
3) Thinning of corpus callosum

4) Cystic lesions or infarcts in parietal, occipital, or temporal lobes
5) Diffuse cerebral white matter loss
6) Altered signal intensities in thalamus, basal ganglia
7) Cerebral dysgenesis like lissencephaly, polymicrogyria

The lesions in PV white matter that is traversed by optic radiations and the lesions involving occipital cortex often found to have severe visual impairment. However, amazing plasticity of neonatal brain in few cases with appropriate vision stimulation therapy might give surprising outcomes. The early intervention programs can favorably help the visual development of children with CVI, if adequate vision stimulation provided during critical period of early neonatal development.

Dr Sujata Guha /

Dr Aditi Yadgire

# Strabismus In CVI children


## Dr Suma ganesh


The rate of coexistent strabismus has been reported to be as high as 73% in patients with CVI. Prevalence of strabismus in CP varies from 19% to 50% . Esotropia is the most common type of strabismus (30-50%). Exotropia Constant is associated in more severely affected patients (10%). Dyskinetic Strabismus- Exotropia changes to esotropia on momentary basis. Specific traits of Strabismus in CVI- *Angle variability* and Shifting pattern. Vertical deviations associated are hypertropia 3% and DVD 11.6%.

Children with cerebral palsy and visual dysfunction caused by periventricular leukomalacia may have exo or esotropia, whereas those who have escaped cerebral palsy often present with esotropia . Mixed deviations like Upshoot in adduction , "V" pattern (inferior oblique overaction) ,An isolated elevation deficit and monocular adduction, elevation, and depression deficit associated with partial ptosis may be seen .

**Management criteria**:

Observe children with poor visual behaviour and with unstable deviation or shifting pattern Operate on them who have constant angle of >15 PD once they are able to follow faces or large toys irrespective of age (>4 years). Repeated measurements- MUST- At least 4 stable and constant measurement before planning surgery. 16% showed spontaneous resolution.

Strabismus and motility evaluation is difficult owing to the behavioural aspects, yet has to performed in multiple visits to know about stability of deviation which helps decide optimal timing of surgical intervention. Esotropia being a common association in neurologically impaired child warrants early surgery. However the surgical results following bilateral medial rectus recession have shown overcorrection as reported by multiple authors. Undercorrection of normal dosage by 15-20% is always done to avoid consecutive exotropia over long term follow up.42,43 There is sparse literature regarding surgical correction of exotropia in these children.

| AUTHOR | MEAN FOLLOW UP DOSAGE for BMR | | REMARKS |
|---|---|---|---|
| Pickering et a<br>JPOS 1994 | 0.54 mm less(10.8%) | 24 months | Increase in effect of the same amount of surgery averaged **5.28 prism diopters** |

| | | | |
|---|---|---|---|
| Wilner JH et al<br>JAAPOS 2006 | 0.84 mm less (16.8%) | 17 months | Overcorrections on long term follow up |
| Wilner JH et al<br>JAAPOS 2012 | 0.75 mm less (15%) | 5 years | **Effect:2.26 degrees/mm** |
| van Rijn LJ et al<br>Strabismus 2009 | Normal dose | 2years | **Effect per mm of surgery in bimedial**<br>**recession larger** than generally experienced |
| Zehavi-Dorin T et al<br>Strabismus 2016 | 0.66 mm less | 4.6 years | Consecutive exotropia with time increased |

# Optical, Accommodation Anomalies and Low Vision Management in Children with CVI

## 1. Introduction

Cerebral Visual Impairment (CVI) is a neurological visual disorder caused by damage to the visual pathways or visual processing centers in the brain, rather than abnormalities of the eyes themselves. It is one of the leading causes of visual impairment in children.

## Common Causes

- Hypoxic ischemic encephalopathy (HIE)
- Prematurity
- Periventricular leukomalacia (PVL)
- Traumatic brain injury
- Seizure disorders
- Central nervous system infections

## 2. Visual Characteristics in CVI

Children with CVI often present with:

- Variable visual attention
- Light gazing behavior
- Difficulty handling visual complexity
- Crowding phenomenon
- Reduced contrast sensitivity
- Visual field defects (commonly inferior field loss)
- Better near vision compared to distance vision
- Delayed visual latency
- Fluctuating visual responses
- Squinting of eyes

Vision in CVI is frequently inconsistent and influenced by environmental and neurological factors.

## 3. Optical Management in CVI

## A. Refractive Error Correction

- Cycloplegic refraction is essential.
- Even mild refractive errors should be corrected.
- Full correction is generally preferred unless contraindicated.

Common refractive findings include:

- Hyperopia
- Astigmatism
- Anisometropia

Early correction supports visual development and improves functional vision.

## B. Special Optical Considerations

- Bifocals in cases of reduced accommodation
- Near add lenses
- Prisms (in selected cases with strabismus, diplopia and field enhancement)
- Tinted lenses in selected cases with photophobia

## 4. Accommodation Anomalies in CVI

Accommodation deficits are common due to neurological involvement.

## Types of Accommodation Anomalies

- Accommodative insufficiency
- Accommodative lag
- Poor accommodative facility
- Inaccurate accommodative response

## Assessment Methods

- Dynamic retinoscopy
- MEM retinoscopy
- Near point of accommodation
- Functional near task observation

## Management Strategies

- Executive-style bifocals (preferred in children)
- Appropriate near add
- Magnification support
- Reduced working distance guidance
- Regular monitoring and adjustment

Addressing accommodation significantly improves near task performance, reading ability, and classroom participation.

## 5. Low Vision Management in CVI

Low vision management in CVI is functional and individualized. It focuses on improving usable vision rather than only visual acuity.

## A. Environmental Modifications

- Reduce visual clutter
- Use plain, simple backgrounds
- Increase contrast (black/white, black/yellow combinations)
- Provide controlled and directed lighting
- Present objects singly
- Position materials in the child's preferred visual field

## B. Optical Low Vision Devices

- Hand magnifiers
- Stand magnifiers
- Dome magnifiers
- Electronic magnifiers (CCTV)
- Tablets with magnification applications

## C. Non-Optical Strategies

- Enlarged print
- Bold fonts
- Increased spacing between letters and lines
- Simplified visual presentation
- Tactile and auditory reinforcement
- Repetition to support visual learning

## 6. Multidisciplinary Approach

Effective management of children with CVI requires collaboration between:

- Pediatric ophthalmologist
- Optometrist
- Orthoptist
- Occupational therapist
- Special educator
- Neurologist
- Parents and caregivers

Caregiver education is critical for consistent home-based reinforcement.

## 7. Clinical Pearls

- Visual acuity alone does not reflect functional vision.
- Near vision is often better than distance vision.
- Contrast enhancement is often more beneficial than magnification alone.
- Accommodation deficits are common and frequently underdiagnosed.
- Vision may improve in a quiet, uncluttered environment.
- Early intervention improves visual outcomes.

## 8. Practical Management Flow

1. Detailed medical and developmental history
2. Cycloplegic refraction
3. Functional vision assessment
4. Accommodation evaluation
5. Prescribe full correction ± bifocal
6. Implement environmental modifications
7. Trial appropriate low vision devices
8. Caregiver counselling
9. Regular follow-up (every 3–6 months)

## 9. Take-Home Message

Cerebral Visual Impairment is a brain-based visual disorder requiring:

- Accurate and early optical correction
- Identification and management of accommodation anomalies
- Functional low vision rehabilitation
- Environmental modifications
- A multidisciplinary approach

Management should focus on enhancing functional vision, supporting developmental milestones, and improving quality of life.

# Early Intervention & Rehabilitative Support Therapy for Maximizing Visual Development in CVI

By Amrita Sharma

Special Educator

## Why Early Intervention is Important

- Rapid brain development during early years.
- Early therapy strengthens visual attention and tracking.
- Delayed intervention limits functional visual growth.
- Parents learn effective home-based strategies.
- Improves school readiness and overall independence.

## Scoring System – CVI Phase Assessment (Roman-Lantzy)

Children are assessed and categorized into three phases based on their visual functioning score.

### Phase I (Score 0–3): Focus – Building Visual Response

- Color Preference Activity: Identify which color the child visually responds to best.
- Use colored objects in preferred colors (yellow, red) with high contrast backgrounds.
- Use visually simplified backgrounds such as black mat or cloth.
- Pause → Move → Pause method to encourage visual attention.
- Light Gazing Activity: Encourage visual attention to light sources using light-up toys, shiny sheets, or torch patterns.
- Movement-Based Activity: Present moving shiny objects slowly (shiny ribbon, lighted toy, pom-poms).
- Sensory Play (Tactile + Vision): Build visual attention through simple sensory experiences.

Success Story Case 1: Nirvan – Improvement observed after consistent early intervention.

### Phase II (Score 4–7): Focus – Integrating Vision with Function

- Look and Reach Activity: Place colorful object within reach after the child looks at it.
- Mirror Play for self-recognition.
- Hand-Under-Hand and Hand-Over-Hand support techniques.
- Identify favorite color and reach for it.
- Face-to-Face interactions: Singing songs, facial expressions, showing pictures of familiar people.
- Visual Tracking Activity: Rolling brightly colored ball, follow-the-light activity.
- Object Recognition: Match real object to picture and match colors.

Success Story Case 2: Yuvansh – Functional improvement observed with structured activities.

### Phase III (Score 7–10): Focus – Functional Daily Routine Activities

- Using vision for learning activities such as dressing, eating, and classroom work.
- Identify colors and clothing items.
- Visual Guided Reach Activity.
- Visual Scanning Activities using worksheets and object search tasks.
- Room-Based Activity: Find the object placed in different locations.
- Visual Motor Integration Activities: Drawing, coloring, building blocks, sorting colored beads, stacking blocks.
- Identification of big and small objects.
- Stimulation activities follow a process depending on the child's ability.

Success Story Case 3: Adib – Noticeable progress in functional vision.

Success Story Case 4: Raghav – Improved engagement and visual participation.

Success Story Case 5: Sanskar – Enhanced visual response and daily activity performance.

Thank You!