

# Cerebral Visual Impairment in Children

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## Abstract

Cerebral visual impairment (CVI) is vision loss as a result of damage to the retrogeniculate pathway of the visual system in the absence of any ocular pathology and with the improvement of perinatal care and increased survival of the preterm babies, it accounts for the leading cause of visual impairment today. Perinatal hypoxic ischemic encephalopathy has been the most common cause of CVI. Morphology of lesion due to hypoxic ischemic injuries to the retrogeniculate visual pathways varies with the timing of the event and the mechanism. Injury during the first trimester results in congenital malformations. Liquefaction necrosis and tissue resorption without any gliotic changes are the major pathological changes. However, damage during the late second to third trimester causes periventricular leukomalacia from ischemic damage followed by gliosis of the subcortical areas. A careful assessment of functional vision as well as level of visual acuity is the first and foremost step in managing these children. Rehabilitation strategies form the backbone of complete management of children with CVI. Every child needs different and special services based on their functional visual and neurological deficit. A multidisciplinary approach is always the mainstay of treatment. Nevertheless, parental education and their cooperation optimises the management outcomes in children with cerebral visual impairment. Prompt diagnosis and timely intervention in terms of neurological, ophthalmological and rehabilitation services always has a rewarding result for children with CVI and their parents.

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## Introduction

Cerebral visual impairment (CVI) is vision loss as a result of damage to the retrogeniculate pathway of the visual system in the absence of any ocular pathology and with the improvement of perinatal care and increased survival of the preterm babies, it accounts for the leading cause of visual impairment today. The term *cerebral visual impairment* is preferred to *blindness*, because the plasticity of the central nervous system and the presence of extra-geniculostriate visual pathways precludes total loss of sight, even when there is complete destruction of the striate cortex.<sup>1</sup>

Over the past decades, the term cerebral is preferred over cortical owing to the subcortical involvement of optic radiations in premature infants. However, many of these children with selective white matter lesions on neuroimaging also have global brain injury with significant cognitive visual disturbance. Cortical visual malfunction per se results from pathology affecting the visual pathways and the pathways serving the higher visual functions. The sequelae of perinatal injury is also related to the timing, degree, duration and mechanism of damage to the developing child's brain. The prognosis and further intervention directly correlates with this.

Nevertheless, management of children with CVI begins with prompt and coordinated effort of ophthalmologist, neurologist and rehabilitation services. CVI thus demands a special emphasis in today's ophthalmology practice.

## Definition

CVI is commonly defined as a loss in visual function in the absence of damage to the anterior afferent visual pathways or ocular structures. CVI includes<sup>2</sup>

CORTICAL	SUBCORTICAL	DELAYED VISUAL MATURATION
Geniculostriate lesions	Focal white matter lesions- Periventricular Leucomalacia (PVL)	Temporary dysfunction of higher cortical centers

## Epidemiology

The reported incidence of CVI in childhood has been increasing from a 36 per 100, 000 in the late 1980s to 161 in 100, 000 in 2003.<sup>3,4</sup> However, PVL has been noted to have a large incidence ranging from 32% to as high as 87%, detected on neuroimaging.<sup>5,6</sup>

## Pathogenesis

Perinatal hypoxic ischemic encephalopathy has been the most common cause of CVI. Morphology of lesion due to hypoxic ischemic injuries to the retrogeniculate visual pathways varies with the timing of the event and the mechanism. Injury during the first trimester results in congenital malformations. Liquefaction necrosis and tissue resorption without any gliotic changes are the major pathological changes. However damage during the late second to third trimester causes periventricular leukomalacia from ischemic damage followed by gliosis of the subcortical areas. Both cortical and subcortical encephalomalacia is the sequelae of term hypoxic injury affecting the parasagittal watershed zones.<sup>7</sup> Lesser degree of term hypoxic ischemic events affects parasagittal and the parietooccipital cortex. But, severe insult in a term newborn can involve thalami and basal ganglia.<sup>8</sup> In a preterm baby, subcortical areas have metabolically active germinal matrix, which produces neurons that migrate to populate the cerebrum. Fragile capillaries in this

layer is prone to hypoxic insult secondary to fluctuations in blood flow and results in haemorrhage. This damages the periventricular areas (watershed zone) causing loss of tissue and permanent scarring.<sup>9</sup>

In a term baby the watershed areas are between anterior and middle cerebral artery territories and between middle and posterior cerebral artery (similar to that of adult). Hypoxia and hypercarbia results in loss of autoregulation and impaired perfusion to frontal and parietooccipital areas. Subsequently, cortical thinning and dilatation of lateral ventricles secondary to loss of gray matter occurs.<sup>5</sup>

Multiple other mechanisms that also have been proposed include oxidative and inflammatory damage that induces death of immature oligodendrocytes. Complex cognitive and attentional deficits are also the result of injury to the *subplate* neurons that form connections between the thalamus and the visual cortex (whose peak timing of development coincides with PVL and IVH).<sup>10</sup>

### Etiology

The leading cause of CVI in children is hypoxic ischemic injury in the perinatal period.<sup>1,2,11-13</sup> Other causes include traumatic brain injury, congenital central nervous system (CNS) malformation and infections, hydrocephalus, cerebrovascular accidents and metabolic disorders.<sup>4,9,14</sup>

In Infants	Acquired Causes (Late Childhood)
<ul style="list-style-type: none"> <li>• Hypoxia – Ischemia</li> <li>• Traumatic Brain Injury</li> <li>• Infections of the CNS</li> <li>• Neonatal Hypoglycemia</li> <li>• Seizure Disorder</li> <li>• Metabolic conditions</li> <li>• Maternal Intake of Drugs</li> </ul>	<ul style="list-style-type: none"> <li>• Respiratory arrest</li> <li>• Head injury</li> <li>• Complications of cardiac surgery</li> <li>• Cardiac arrest</li> <li>• Focal infection</li> <li>• Cerebrovascular accidents</li> <li>• Status epilepticus</li> <li>• Encephalitis</li> </ul>

### Perinatal Hypoxia- Ischemia:

Hypoxic brain damage in full term infants results in loss of auto regulation that affects the watershed areas affecting anterior parasagittal involving the frontal cortex or the posterior parasagittal cortex in parietooccipital region.<sup>4,15</sup>

### Postnatal Hypoxia-Ischemia:

Perinatal strokes are mostly ischemic in origin and result in hemiplegic cerebral palsy. Associated hypoxic ischemic events are usually present may result in CVI, due to decrease in blood supply to the posterior visual pathway.<sup>16</sup>

### Head trauma:

CVI has been one of the major cause for permanent visual loss in children with non accidental head trauma (shaken baby syndrome). Posterior cerebral ischemia and subdural hematoma identified in diffusion weighted MRI characterize this condition. Retinal hemorrhages, retinal detachment, retinal folds, macular hole, or epiretinal membrane are other causes of visual morbidity.<sup>17</sup>

### Congenital CNS Malformations:

Occipital or parietal encephaloceles, Chiari malformations, Dandy–Walker complex, hydranencephaly, porencephalic cysts and other neuronal migrational abnormalities may be associated with CVI.<sup>2</sup>

### Congenital CNS Infections:

Neonatal herpes simplex is the most common cause in cases with severe CVI.<sup>18</sup> Eighty percent of post meningitic CVI are caused by type 2 herpes simplex virus. Bacterial meningitis rarely cause 5% of the cases.<sup>19</sup> Post meningitic CVI are mainly can be explained due to venous sinus thrombosis, thrombophlebitis, hydrocephalus, or hypoxic-ischemic insult in the watershed areas.

### Hydrocephalus:

In patients with hydrocephalus both anterior and posterior visual damage is usually seen either primarily or following shunt malfunction. CVI due to posterior visual pathway damage in these children is possibly due to vascular dysfunction mediated by intracranial hypertension or compression of the posterior cerebral arteries.<sup>20</sup>

### Metabolic disorders:

CVI may be one of the clinical features of various neurodegenerative conditions, including MELAS ornithine transcarbamylase deficiency, Fabry's disease, Leigh's disease, and X-linked adrenoleukodystrophy. Metabolic and neurodegenerative causes of cortical visual loss usually present later in childhood.<sup>21</sup>

### Subcortical Visual Loss: Periventricular Leucomalacia (PVL)

Prematurity is the most common cause of neurologic morbidity and visual impairment in children with CVI.<sup>4,9,11</sup> Injury to the preterm developing brain affects the subcortical white matter, resulting in PVL. The watershed zone is particularly close to the trigone of lateral ventricles, posteriorly rather than anteriorly adjacent to frontal horns. The optic radiations and corticospinal tracts are affected resulting in visual impairment and cerebral palsy as well as intellectual impairment.<sup>9</sup> Obstetric risk factors for periventricular leukomalacia among preterm infants include intrauterine infection, premature rupture of membranes, maternal chorio- amnionitis, hemorrhage during the first trimester.<sup>2</sup>

### Clinical Features

The qualifying remarks of CVI include<sup>2,4,9,22</sup>

1. CVI represents a spectrum of disability
2. Diagnosis of CVI should be suspected in all children with unexplained bilaterally decreased vision, even when there are no overt neurological problems
3. Pupillary reaction to light may not be completely normal
4. May display intermittent, unsustained bursts of nystagmus
5. Coexistent optic atrophy may be present

Moreover, CVI is usually associated with other systemic and neurologic disorders that include Cerebral palsy, seizures, microcephaly, hydrocephalus, sensorineural hearing loss, myelomeningocele, and progressive CNS degeneration.<sup>23</sup> Huo et al reported 19% of patients had ophthalmic deficits (strabismus, nystagmus, optic nerve pathology), 30% had neurologic deficits (seizures, cerebral palsy, microcephaly, hemiparesis) and 46% had both neurologic and ophthalmic deficits.<sup>13</sup> Hence in view of close link between the neurological and visual functions, it is very important to evaluate vision in terms of "4 A"- Acuity, Assimilation, Attention, Apraxia.

#### Visual Function:

Visual impairment in CVI can be as severe as no light perception to normal visual acuity. However cognitive visual function may be impaired in most of these children leading to misinterpretation in relation to what objects and where they are. Assessment of functional visual acuity in these children has been assessed by Hoyt et al in 6 different levels.<sup>3</sup>

- level 1: perception of light
- level 2: occasional fixation on large objects, faces, or movements in the environment
- level 3: variable visual function but some moments of good visual fixation as indicated by the ability to see small objects or reliably fixate on the face
- level 4: reliable fixation on a small target or with visual acuity measured in the range of 20/200 to 20/400
- level 5: with one or both eyes open, vision between 20/50 and 20/200
- level 6: normal sensory visual examination

Other notable features of visual assessment includes:

These children see better in familiar environment. They choose to view objects at closer range. Identification of objects is mainly done by touch than vision. They also have crowding phenomenon preferring individual objects over a group of toys. Their performance decreases in a cluttered environment.<sup>2,3</sup> Many of these children have a wide fluctuation in the visual function based on lighting conditions, tiredness, medications etc. Some of them prefer light-gazing whereas other parents complain of photophobia (less severe as opposed to retinal lesions). The cause of both remains unexplained, however possible explanation for photophobia is explained by damage to thalamic or cortical structures.<sup>8</sup> Again some of these children see better in static environment unlike others who prefer to see moving objects.

**Visual Fields:** Children with CVI usually have bilateral inferior field defects. Many of them present with homonymous hemianopia in association with hemiplegia. Interruption of axons of optic radiations explains the restriction of visual fields in these children. It can partly be explained by the problems of simultaneous attention in them.<sup>7,9</sup>

#### Ocular Findings

**Refraction:** Myopia is common in premature children who also have retinopathy of prematurity (ROP). In children with PVL without ROP hypermetropia in combination

with astigmatism has been frequently reported. However in children with cerebral palsy, hypermetropia is the common refractive error. Accommodation dysfunction is also common in most of these children.<sup>9,24</sup> In our experience in children with PVL simple hyperopic astigmatism was the commonest refractive error. Myopia ranged upto - 9.25 whereas hyperopia ranged upto 4.5D.<sup>26</sup>

**Strabismus and Ocular Motility:** It is prudent to consider CVI as a cause in children with isolated congenital exotropia.<sup>7</sup> These children usually have congenital exotropia as compared to esodeviation. On the contrary children with PVL present with esotropia commonly.<sup>9</sup> This needs to be differentiated from infantile esotropia based on other concurrent signs of retinopathy of prematurity, optic nerve changes or neuroimaging findings. Another specific trait of strabismus in these children is the shifting pattern or the dyskinetic strabismus, where esotropia changes to exotropia on momentary basis.<sup>9</sup> Repeated evaluation and a constant deviation is thus crucial before planning any surgical intervention. It is always important to remember that children with undiagnosed PVL may present to the ophthalmologist with strabismus and no other apparent neurological abnormality.<sup>25</sup> In our experience with PVL the commonest presenting ocular complaint was strabismus (59.3%). Exotropia was seen in 12 (37.5%) patients whereas 10 (31.2%) children had esotropia.<sup>26</sup>

**Nystagmus:** Although a rhythmic nystagmus is very rare, roving eye movements with intermittent or unsustained beats of nystagmus is not uncommon in cortical visual loss. Presence of nystagmus in these children implies coexistent anterior visual pathways involvement, or the onset of visual loss has been before first year of life.<sup>2</sup> However, children with PVL do display latent or manifest nystagmus either due to damage of the optic radiations or the white matter lesions disturb the neural generator of latent nystagmus located in the cortico-tectal pathways. Children with mixed mechanism visual loss where both anterior and posterior pathways are affected may not have severe nystagmus if posterior pathway damage is significantly more and vice versa.<sup>27</sup>

In children with PVL defective smooth pursuit movements and inability to perform visually guided saccadic movements is usually documented. Ocular motor apraxia and absence of fixation has been noted in some children with extensive PVL and cerebral palsy.<sup>9</sup> Though the cause for ocular motility is not definite, these functions are subserved by the dorsal stream pathway, which could be disrupted in white matter lesions of PVL.<sup>28,29</sup> In a study by Nielsen et al, refractive errors and strabismus were significantly correlated with low IQ.<sup>30</sup> Apart from decreased visual acuity and restricted fields, horizontal gaze deviation is an important diagnostic sign of CVI.<sup>31</sup> Here both eyes are deviated to one side and head turned to the same side This is explained by the asymmetric injury to the cortical command centres.<sup>7</sup> Children with cortical visual loss mostly had horizontal tonic gaze deviation whereas tonic downgaze was preferred by children with subcortical visual loss.<sup>7,9</sup>

**Optic Discs:** Brodsky et al reported normal discs in 56%, optic atrophy in 24%, optic nerve hypoplasia without

atrophy in 8%, and combined hypoplasia with atrophy in 12% in children with cortical visual loss.<sup>7</sup> Posterior visual pathways are increasingly susceptible to hypoxic damage. PVL is characterized by optic nerve hypoplasia or pseudo glaucomatous optic discs. These discs represents a classical optic nerve configuration with an abnormally large optic cup and a thin neuroretinal rim in a normal-sized optic disc.<sup>9,32</sup> Retrograde transsynaptic degeneration of retinogeniculate axons and bilateral damage to optic radiations in these children stands as the probable explanation for such appearance. Brodsky et al also explains the pattern of optic disc appearance in relation to the timing of injury in PVL. Early PVL before 28 weeks is usually associated with small optic discs whereas normal-sized optic disks with large cupping with a reduced neuro-retinal rim area was in PVL after 28 weeks.

### Dorsal and Ventral stream dysfunction:

In the developing brain 2 major pathways have been well delineated which control the cognitive visual function in these children. These include the dorsal and ventral stream.<sup>33</sup> To elaborate, the dorsal stream connects the occipital area with the posterior parietal cortex, which allows the mind to encompass the whole visual scene and to elect to pay attention to chosen components representing '*WHERE*'. The ventral stream connects the occipital and temporal lobe territories and subserves recognition of geometric and biological form, route finding, and visual memory i.e. *WHAT pathway*.

Ventral field deals with conscious analysis and understanding of the visual world. Recognition of shapes, objects, facial expressions, people is the major clinical function of this path. Accurate movement of the body through visual space but at a subconscious level is brought about by the dorsal stream. Other functions like visual attention and visual guidance of movements of head and body are controlled by the dorsal stream.<sup>13,16</sup>

In a study by Dutton et al, dorsal stream dysfunction was noted in all children with spastic diplegia or hemiplegia, but less than 50% had ventral stream dysfunction. Difficulty in climbing stairs, identification by touch; inaccurate reaching and knocking over objects; impaired simultaneous perception, inability to locate an object in a crowded visual field, such as toys in a toy box or a parent in a crowd were the major difficulty faced by majority with dorsal stream abnormality. Ventral stream impairment was seen only in children with severe CVI, faced problems with route finding in unfamiliar places; forgetting where objects were located; and difficulty recognizing faces, shapes or objects.<sup>34</sup>

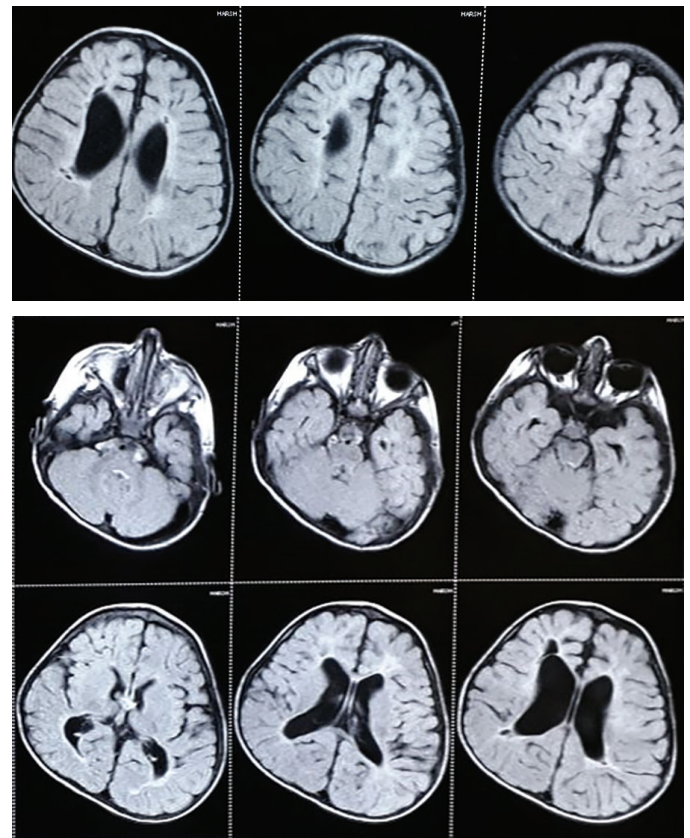
### Diagnostic and Prognostic considerations

Diagnosis of this entity is of utmost importance in a child with normal ocular examination. However suspicion begins in the immediate neonatal period. A perinatal ultrasound in children with low APGAR scores is always recommended. Although a positive ultrasound favours PVL in most of these immature infants, subtle changes may be missed.<sup>35</sup> Neuroimaging has been the modality in young infants who present for the first time, magnetic resonance imaging (MRI) with contrast is always preferred over computed

tomography (CT). Normal CT does not exclude presence of mild CVI.

Grossly, the classical findings noted in children with CVI in MRI include- Diffuse cerebral atrophy, bioccipital lobe infarctions, periventricular leukomalacia, cerebral dysgenesis, and parieto-occipital and parasagittal "watershed" infarction.<sup>4</sup> More specifically, MRI changes in PVL are reduction in periventricular white matter, prominently in the posterior regions with compensatory ventricular dilatation and irregular outlines.<sup>9,36</sup> In T2 weighted sequences, abnormally increased signal intensity of the periventricular white matter is another notable imaging feature. In extremely severe cases or children with associated mental retardation, volume loss of corpus callosum and shrinkage of basal ganglia, thalami, cerebellum have also been found.<sup>9</sup>

Fazzi et al in his study, established a correlation between severity of MRI changes and visual prognosis. Greater is the severity of periventricular leukomalacia in the peritrigonal white matter and calcarine atrophy, lower was the grading of visual acuity in children.<sup>37</sup> Functional MRI analyses the changes seen with visual stimulus and helps understanding of the pathophysiology. The three dimensional view of optic radiations, white matter tracts and localization of damage along these tracts can be detected by Diffusion tensor imaging scans (DTI). It not only analyses structural changes, but can also correlate with visual field changes also.<sup>38</sup>



**Figure 1:** Shows Mild white matter paucity in bilateral parieto occipital lobe. Bilateral frontoparietal periventricular white matter and periventricular white matter suggestive of PVL. Ex vacuo dilatation of ventricles.

Visual evoked potentials have been used to compare visual function with preferential looking tests.<sup>39</sup> Flash VEP evaluates a gross visual function, whereas sweep VEP predicts some correlation with neurodevelopmental function.<sup>40</sup>

### Management

A careful assessment of functional vision as well as level of visual acuity is the first and foremost step in managing these children. Dynamic retinoscopy is necessary before glasses are prescribed. Amblyopia management with patching therapy remains the crux of management though compliance has been a major concern.<sup>41</sup> A close monitoring of occlusion therapy is warranted in these children to prevent further psychosocial harm and developmental delay.

Strabismus and motility evaluation is difficult owing to the behavioural aspects, yet has to be 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.<sup>42,43</sup> There is sparse literature regarding surgical correction of exotropia in these children.

Rehabilitation strategies forms the backbone of complete management of children with CVI. Every child needs different and special services based on their functional visual and neurological deficit. Exercises that focuses on use of clutter free environment, development of hand eye coordination, enhancement of mobility by visual guidance are needed by the majority.

### Conclusion

A multidisciplinary approach is always the mainstay of treatment. Nevertheless, parental education and their cooperation optimises the management outcomes in children with cerebral visual impairment. Prompt diagnosis and timely intervention in terms of neurological, ophthalmological and rehabilitation services always has a rewarding result for children with CVI and their parents.

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