Original Article

Cerebral visual impairment and effect of phase-wise stimulation strategies—An interim analysis

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Context: Cerebral visual impairment (CVI) is an overarching term, defined as a brain-based visual impairment with onset in childhood, unexplained by an ocular disorder and associated with unique visual and behavioral characteristics. Good vision and awareness of visual function in a child are highly essential as neuroplasticity is maximum in the first three years of life and response to intervention is utmost in this period. Awareness is lacking regarding CVI, and the diagnosis is largely missed. This can be easily addressed if a structured approach is employed. **Purpose:** This study aims to evaluate the etiology and radiological correlation with the severity of CVI and outcome after structured intervention in children with CVI. **Settings and Design:** Prospective—interventional study. **Methods and Material:** Children attending the Child Development Centre (CDC) of a tertiary care hospital in North Karnataka and diagnosed with CVI in the age group of six months to 12 years and meeting the sampling criteria were screened and enrolled consecutively after obtaining parental consent/assent. Statistical analysis used is nonparametric test with SPSS software. **Results:** Age showed a significant association with the phase of CVI. Perinatal insult was associated significantly with the severity of CVI. Magnetic resonance imaging (MRI) findings did not hamper the recovery of CVI. **Conclusions:** Enrolment in early intervention programs tailored according to child's specific needs should be encouraged, with stress on ophthalmic screening of preterm and high-risk babies with perinatal hypoxia and history of convulsions, as early as six months.

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Cerebral visual impairment (CVI) is an overarching term, defined as a brain-based visual impairment with onset in childhood, unexplained by an ocular disorder and associated with unique visual and behavioral characteristics.[1-4] Children with CVI often have multiple associated conditions, either neurological or developmental. The degree of brain damage and visual impairment depend on the timing, location, intensity, and specificity of the insult. Perinatal injury is a common cause, which hampers the neurological development (motor and cognitive) and disturbs the five constructs of vision which comprises light, color vision, contrast, movement, and stereopsis. CVI-related impairments can be mild to severe; basically, it leads to functional limitations that impact the child's learning, movements, and overall quality of life.

Cerebral visual impairment (CVI) is a major cause of childhood blindness in developed countries and is gradually increasing in developing countries. In developing countries like India, increasing rate of survival of premature babies with perinatal brain injury is leading to rise in cases of CVI in children.[5] Moreover, CVI affects not only vision but also the attainment of developmental milestones.

In developed and developing countries, the prevalence of CVI in children under 16 years of age is between 10 and 22 subjects per 10,000 births and 10 per 10,000 births, respectively.[6] In India, prevalence of CVI in children with cerebral palsy is around 28%.[7] The prevalence of visual impairment in children with multiple disabilities is found to be 10.5%.[8] A recent study performed in Scotland found that at least one in every 30 children of mainstream schools had symptoms related to CVI.[9]

Awareness of visual disability and functional impairment in the child is essential as neuroplasticity is maximum in the first three years of life and response to intervention is utmost in this period. In spite of this, there is minimal knowledge regarding CVI and the diagnosis is largely missed. It is to be borne in mind that once diagnosed, phase-wise management and structured habilitation approach can be employed with favorable outcomes.

However, CVI is still in early stages of both research and clinical practice due to lack of clarity in terms of terminology, assessment methods, and diagnosis. Structured evaluation and management of cerebral visual impairment (CVI) in children has been under rated. Impact on overall development of the child needs to be studied. There is a lacuna in studies on childhood visual disorders caused by brain injury due to

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Table 1: Comparison of age with different phases

Age group	CVI phase I	CVI phase II	CVI phase III	Total
<1 Years	10	0	0	10
1-5 Years	12	6	8	26
>5 Years	1	0	3	4
Total	23	6	11	40

Pearson Chi-square value is significant at 0.008

Table 2: Comparison of phases of CVI with MRI findings

CVI phases/ MRI findings	Normal (%)	Hypoxic–ischemic encephalopathy/ Periventricular leukomalacia (HIE/PVL) (%)	Others (%)
Phase I	7 (30)	13 (57)	3 (13)
Phase II	2 (33)	2 (33)	2 (33)
Phase III	2 (18)	5 (46)	4 (36)

Pearson Chi-square value was not significant

Table 3: Comparison of phases of CVI with type of perinatal insult

CVI phases/ perinatal insult	Normal (%)	Seizure (%)	Jaundice (%)	Others (%)
PHASE I	6 (26)	12 (52)	3 (13)	2 (9)
PHASE II	5 (83)	0 (0)	1 (17)	0 (0)
PHASE III	4 (36)	1 (9)	4 (36)	2 (18)

Pearson Chi Square value is significant at 0.022

Table 4: Follow-up data

Total Number	18
Improvement in CVI phase	5
Within same CVI phase—RATING SCORE improvement	9
Improvement in visual acuity	4

challenges encountered in testing young children. There are very few studies dealing with systematic treatment approaches in these children, especially in Indian scenario. Hence, this study aims to plug the gap.

Objectives

Primary objective: To study the outcome after structured intervention in children with cerebral visual impairment (CVI).

Secondary objective: To evaluate the etiology and radiological correlation with the severity of CVI.

Methods

Children attending the Child Development Centre (CDC) of a tertiary care hospital in North Karnataka and diagnosed with CVI in the age group of six months to 12 years and meeting the sampling criteria were screened and enrolled consecutively after obtaining parental consent/assent. Children in whom visual deficit was attributed to anterior segment anomalies like corneal opacity, congenital glaucoma, congenital or developmental cataract, and posterior segment pathologies like sequelae of retinopathy of prematurity or retinal dystrophies were excluded from the study.

The diagnosis of CVI was made when the ophthalmological findings did not explain the visual performance of the child, and there was associated neurological involvement. CVI assessment was made by pediatric ophthalmologist running the CVI clinic using the ten characteristics of CVI, which is structured for all pediatric age groups with suitable age-wise modifications.[10] MRI brain was evaluated to know the extent of the injury. Perinatal history regarding the occurrence of neonatal seizures, hypoglycemia, jaundice, oxygen therapy, or any other significant event was documented.

TAC was used to assess and quantify visual acuity in all children. Static and dynamic retinoscopy and ocular examination was performed as per the standard protocol.[11,12] Refractive correction was prescribed to children with significant errors and compliance was noted as minimum six waking hours of spectacle use, as confirmed by the parent. CVI was classified on the basis of ten characteristics as phase I, II, or III at their initial evaluation.[10] Severity was based on the rating, range and phase of CVI. Appropriate treatment/educational strategies were briefed to the parents according to the phase of CVI. A CVI Kit encompassing all the interventional activities was indigenously developed to ensure better compliance. It included an inventory list. Duration of activities advised ranged from one to three hours in divided sessions according to the phase of CVI.[10] Parents were asked to mention the number of hours of activity performed in the list. Smartphone-based messenger apps were utilized for monitoring and guiding. Follow-up was performed every three months.

The protocol was reviewed and approved by the Institutional Internal Review Board. In all children, consent for evaluation and treatment was obtained from the responsible parent/guardian. Data were collected and analyzed with SPSS software.

Results

During the study period, a total of 40 children referred by CDC with a diagnosis of CVI were enrolled. Twenty-seven (67.5%) were born at term and rest were preterm (gestational age <36 weeks at birth). There was a history of difficulty in breathing/delayed birth cry in 14 (35%). The mean birthweight was 2.43 kg.

In our study, the majority of children were in the age group of 1-5 years [Table 1]. Most children were grouped in CVI phase 1 (58%). In the age group less than one year, all children were diagnosed with CVI phase I. 16 (40%) children had strabismus. Among them, 11 had exotropia (68.75%) and five had esotropia (31.25%). We found significant refractive error in 18 subjects (45%) children.

Based on MRI findings, most common cause of CVI in our group was hypoxic ischemic encephalopathy (HIE) seen in 20 subjects (50%). In a significant number of children (27.5%) with CVI, the MRI findings were normal [Table 2].

Among the 23 subjects grouped in Phase I CVI, 17 had perinatal insult. A history of neonatal seizures was present in 13 subjects (32.5%). Fifteen children (37.5%) had an

uneventful perinatal period. Neonatal jaundice was seen in eight subjects (20%) [Table 3].

Other causes included neonatal hypoglycemia, meningitis, and hydrocephalus.

Discussion

CVI is a disability, which is on the rise not only in developed countries but also in the developing countries. It is a matter of rising concern as it is a disability which is hidden, hence requires extra effort to diagnose. Studies reveal that recognition and training at an early stage is helping these children overcome their difficulties. A step toward implementing the interventions is to identify the present stage of development and determine the level of CVI and to classify CVI into phases based on severity. Depending on the CVI characteristics, scoring system of Christine Roman-Lantzy is followed.[10] Interventions can be planned in phased manner, and quantitative evaluation of progress is recorded. The plasticity of the visual system and results of previous studies showed that training can enhance visual function which led to the concept of developing training programs matching the nature and degree of visual dysfunction.[13]

The purpose of management is to identify the problems faced by each child and to device practical solutions to these problems. Many children adopt unique strategies to overcome their difficulties, which help them with everyday living. The full range of visual limitations is identified and characterized, and full range of capabilities and strengths is utilized to utmost advantage.[14]

This study has focused on evaluating these children under phases of CVI and providing them with an intervention plan accordingly. Creating visual awareness and training different areas of visual perception is the core plan. This is an ongoing prospective study and an interim analysis has been attempted. In this study, 65% children were in the age group of 1-5 years.

46% of these children were in CVI I, 23% in CVI II and 31% in CVI III, thus ruling out the age effect. This is the age when the developing brain is maximally receptive to intervention. Any strategy planned at this stage would seemingly produce better results. The reason for increased proportion of children in phase I could be due to delayed presentation to the hospital and late diagnosis which meant they were not under any habilitation plan. HIE was the major MRI finding in all the phases, which was similar to other studies.[15] Among 14 children, three had periventricular leukomalacia (PVL) in phase I and two in phase III. 27.5% had normal MRI suggesting rehabilitation strategies need not be based on MRI findings.

According to a study performed in Canada, asphyxia, acute or prolonged, results in severe form of CVI.[16] Hypoxic insult from seizures results in severe damage to the visual processing pathways, which comprises 80% of the brain. This study showed similar results as there was a statistically significant association between the type of insult and the severity of CVI. In CVI phase I, convulsions were the most common perinatal insult (52%), whereas in phase III, jaundice was the most common insult. Irrespective of age and prematurity, children with convulsions in the immediate postnatal period developed severe form of CVI (phase I). History of jaundice or oxygen therapy did not have any influence on the severity of CVI.

Due to the pandemic, follow-up was hindered [Table 4]. 18 children have turned up for the three month follow-up and all have shown significant improvement either in phase or in their rating scores within the phase. Five children improved to the next phase, and in another nine children who maintained in the same phase, CVI rating score showed improvement. Four kids remained in the same phase but visual acuity improved as assessed by Teller Acuity Cards.

Hoyt *et al.* (2003) stated that in children with severe disabilities and MRI changes, visual recovery was incomplete and was not meaningful. They concluded that the lesions in MRI did not hamper the recovery and children with epilepsy and periventricular leukomalacia had poor prognosis.[17,18] Most studies stress that severe asphyxia leads to severe form of CVI, which is the most challenging group to evaluate and habilitate.[16] This study found otherwise. Improvement was seen in severe phases of CVI in spite of severe causes of asphyxia like PVL and HIE on MRI. Likewise, few children with normal MRI did not show expected improvement. Again proving that MRI findings need not correlate with the severity of CVI as well as outcome.

The improvement was observed in both term and preterm children. In contrast to Good *et al.* study (2001), improvement in functional vision was observed.[17] Evaluating these children based on CVI characteristics and following adaptive strategies accordingly helps to achieve greater time bound functional vision improvement. This is in confirmation with the study which says there is considerable neuroplasticity in this challenging group, with neural connections developing all the time.[14] Purposeful planned stimulation gives this whole network a jumpstart. Hence, it is important to know the level of child's development and frame individualized strategies, which are practical and provide environmental support at home and school likewise. Keeping in mind the socio-economic strata of majority of these parents, our study aimed at substitution of locally available affordable quality tools, developed and provided in the form of a kit. The kit incorporated feasible activities and therapies easily practiced at home. Moreover, training and monitoring using the smartphone apps served a dual purpose of compliance and guidance. This was found to be a user-friendly approach by the parents too.

Conclusion

Enrolment in early intervention programs tailored according to child's specific needs should be encouraged, with stress on ophthalmic screening of preterm and high-risk babies with perinatal hypoxia and history of convulsions as early as six months. This interim analysis has fortified the belief that structured interventions is beneficial for children with CVI and positively reinforces researchers to conduct the study on a large scale.

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Conflicts of interest

There are no conflicts of interest.

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