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Original Research Article

Profile of cerebral visual impairment in children less than 7 years – A prospective observational study

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ABSTRACT

Aim: To study the incidence of cerebral visual impairment among children with history of perinatal neurological insult and congenital brain anomalies and to analyze the outcome following visual intervention programme.

Materials and Methods: A prospective interventional study, examining 100 children less than 7 years over a period of 1 year. All children with history of perinatal hypoxia, neonatal hypoglycemia, neonatal seizures, infantile spasm, epilepsy, congenital hydrocephalus, congenital brain anomalies, CNS infections, traumatic brain injury and post cardiac surgery, excluding children with ocular visual impairment.

Results: Of the 100 children studied, the incidence of CVI was 87%. The associated ocular problems were found to be low visual acuity seen in 85%, strabismus in 25%, refractive error in 39%, Nystagmus in 21% and oculomotor apraxia in 6%. The most common etiology was hypoxic ischemic encephalopathy, contributing to 30% of the cases and most common MRI findings were periventricular leukomalacia, seen in 42%. CVI Range was used for functional visual assessment. Visual intervention rather than visual stimulation was taught to the primary care giver and children were followed up every 3 months. 26% of children had improved in their functional vision after 6 months of Individualized intervention.

Conclusion: Early identification and individualized visual intervention, integrating them into many activities of the child across the whole day, form a crucial part of the visual outcome. Improvement in vision creates access to a world of cognitive and motor development.

1. Introduction

Although vision or sight, as a sense is primarily associated with eyes, in reality vision is the product of complex system of which eyes are a part.[1–5] The processing of visual information involves the receipt of visual stimuli from eyes, its interpretation in various brain centers and translation into visual images, which involves up to 40% of the brain. Traditionally educators of visually impaired assisted only those whose eye conditions were associated with ocular visual impairment. Now it has become necessary to offer services to those whose visual loss is due to brain damage. Thus, the definition of "cerebral visual impairment" is born.[6] With good perinatal and pediatric care, the number of children surviving the neurological insult has increased.[7] Thus, a system where there is early identification and individualized intervention is no longer a choice but a necessity.

CVI is commonly defined as a loss in visual function in the absence of damage to anterior afferent visual pathways

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or ocular structures.[8] CVI has multiple causes, but the most common is perinatal hypoxia. CVI can result from the insults at the level of the cortex, subcortex or a temporary dysfunction of higher cortical centers. The risk factors are hypoxia–ischemia, CNS infections, hydrocephalus, trauma, epilepsy hypoglycemia, maternal intake of drugs, metabolic disorders, congenital neurological malformation, cardiac arrest, respiratory arrest and complication of cardiac surgery.[9] For a child with CVI their behavior is their means of communication.[10] A child's behavior may not be what it seems. What we see on the surface is their behavior.[11] Visually impaired children have different blind mannerisms like rocking; thumb sucking, head banging, head flopping, eye pressing, light gazing and flicking their fingers.[12] What we don't see beneath the behaviour is their problem with visual and sensory complexity, difficulty in distance vision, latency and visual novelty.

2. Materials and Methods

It was a prospective interventional study with a sample size of 100 children less than 7 years of age. All children less than 7 years of age with history of cerebral palsy, hypoxic ischemic encephalopathy, congenital brain malformations, neonatal seizures, infantile spasms, epilepsy, CNS infections, congenital hydrocephalus, neonatal hypoglycemia and traumatic brain injury were included and those with retinopathy of prematurity, congenital cataract, corneal dystrophy and congenital glaucoma. Materials employed in the study include[13] preferential looking grating acuity cards, Snellens chart, Prism bars, hand held slit lamp biomicroscopy, streak retinoscope, Indirect Ophthalmoscope, Tools to assess functional vision (toys with single color, toys with two colours, toys with multiple colours, toys with sound).

All children are evaluated after their parents have given their informed consent.[14] The CSM (Central Steady Maintained Fixation to light) is used to measure visual acuity in children less than 3 years old.[15] Preferential looking test (Lea paddles) is used in 3- to 5-year-olds.[16] Allens Picture Card test and Snellens visual acuity chart are used above 5-year-olds. Anterior segment examination by slit lamp biomicroscopy. Cover uncover test and prism bar cover test to assess squint. Nystagmus and Oculomotor apraxia were looked for. Atropine refraction with 1 percent atropine eye ointment applied twice a day for 3 days. Dilated Fundus Examination by Indirect Ophthalmoscope.[17] All these children, even those with normal visual acuity, are tested for functional vision.[18] Functional visual assessment - showing the child first with a single- coloured toy (red, yellow, green), moving toy with reflective material, a familiar object of the child, Multicoloured toys and in the presence of visual and auditory complex surroundings (Figure 1).

The following 10 categories (Table 1) in functional vision are assessed based on the presence or resolving of CVI characteristics and added together to obtain a total score. The score of the CVI range is then used to calculate the phase of CVI.[19]

Range 1–3.5 – Phase 1
Range 3.5 –7.5 – Phase 2
Range 7.5 –10 – Phase 3
0 – Not resolved
0.25 – Resolving, often a factor affecting visual functioning
0.50 – Resolving, sometimes a factor affecting visual functioning
0.75 – Resolving, rarely a factor affecting visual functioning 1–Resolved, not a factor affecting visual functioning

1. After categorizing the child in the CVI Phase, the primary care giver of the child is taught CVI therapy, which varies according to phases. The 3 Ds of the intervention programme are
   (a) Development: promote stimulation and activities at the current levels and increase complexity over time.
   (b) Diversity of visual stimulation: Initiate with regard to preferred objects, advance to generalized objects rather than novel ones.
   (c) Duration: Increase the length of time of visual use over the day.
2. These children are followed up at 3 and 6 months and the functional Visual assessment is assessed by the same questionnaire and CVI range score is calculated and categorised into functional vision CVI phase.[20]
3. MRI brain findings in the study population was analyzed in the study.
4. VEP (Visual Evoked Potential) was done for children with normal brain MRI.

2.1. Analysis

The collected data was analysed with IBM SPSS statistics software, 23.0 version. Frequency analysis and percentage analysis were used for categorical variables and the mean and variance were used for continuous variables. Chi–square contingency calculator has been used to calculate chi–square statistics, p-value and statements of significance.

3. Results

3.1. Etiology of CVI

The most common etiology contributing to CVI were Hypoxic ischemic encephalopathy (30%), Traumatic brain Injury (11%), Epilepsy (11%), congenital brain anomalies

(9%), congenital hydrocephalus (5%), neonatal seizures (9%), Infantile spasm (6%), neonatal hypoglycemia (3%) and post cardiac surgery (2%). (Figure 1)

3.2. MRI brain

Periventricular Leukomalacia was seen in 42%.[21,22] Dilatation of the lateral ventricles in 5%, basal ganglio-thalamic lesion in 27%, normal in 10% and the remaining 27% consisted of corpus callosum agenesis, lissencephaly, holoprosencephaly, Dandy Walker and Arnold Chiari malformation (Figure 2).[23] VEP done for 10 children had prolonged P100 latency in 9 and no waveform was obtained in 1.

3.3. Prognosis

At the end of follow up at 6 months, 26 (26%) of children had improved to CVI Phase 3 from CVI Phase 1 and 2. 13% had improvement in follow up 1 and sustained it in follow up 2. 19% had no improvement at follow up 1 but has improved at follow up 2. 23 % had no improvement at follow up 1 and 2 (Figure 3). The chi –square statistic is 26.6819. The p–value is .000023.The result is significant at p <.05.

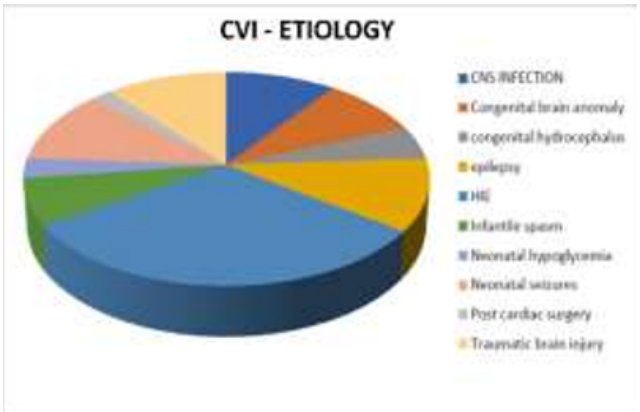

Fig. 1: Etiologies of CVI and their proportions

4. Discussion

The reported incidence of CVI has increased from 36 per 1,00,000 in late 1980s to 161 per 1,00,000 in 2003.[24,25] Prevalence of cerebral visual impairment in developed country varies from 10 to 22 cases /10,000 births in children less than 16 years of age, while reported around 10/10,000 birth in developing country.[26] Low visual acuity was seen in 85% and 45% of children had very low visual acuity such that their visual acuity could be measured by only CSM method of which 2% had no light perception, 33% had central steady unmaintained fixation and 10% had central steady maintained fixation to light. Strabismus was seen in 25% of children of which 18% had Exotropia and 7%

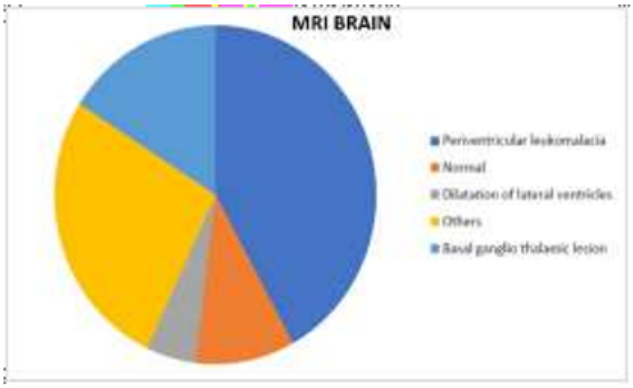

Fig. 2: MRI findings and their proportions

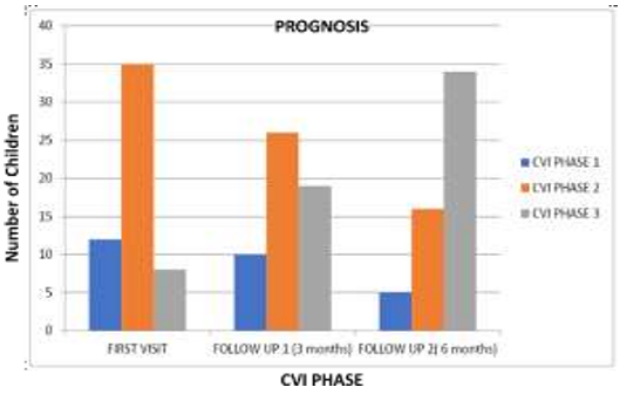

Fig. 3: Prognostic improvement on follow up visits

Fig. 4: Phasic introduction of different toys

Table 1: Parameters for assessment

S.No.	Character	Not Resolved		Resolving		Resolved
1	Color preference	0	0.25	0.50	0.75	1
2	Need for movement	0	0.25	0.50	0.75	1
3	Visual latency	0	0.25	0.50	0.75	1
4	Visual Fields preferences	0	0.25	0.50	0.75	1
5	Difficulties with visual complexity	0	0.25	0.50	0.75	1
6	Light gazing, non purposeful gazing	0	0.25	0.50	0.75	1
7	Difficulty with distance viewing	0	0.25	0.50	0.75	1
8	Atypical Visual distance viewing	0	0.25	0.50	0.75	1
9	Difficulty with visual novelty	0	0.25	0.50	0.75	1
10	Absence of Visually guided reach	0	0.25	0.50	0.75	1

had esotropia.[24] CVI is most commonly associated with exotropia whereas Periventricular Leukomalacia is most commonly associated with Esotropia.[24] Suma et al., have found that an undiagnosed Periventricular Leukomalacia may present with a strabismus and no other apparent neurological abnormality. Nystagmus was seen in 21% of children. Presence of nystagmus shows associated afferent visual pathway involvement or the loss of vision has started before 1 year of age.[26] In Eliza et al., study, contrary to popular belief, Periventricular Leukomalacia may present with latent or Manifest Nystagmus due to subcortical loss or white matter lesion affecting corticotectal pathway.[24] A study by Suma et.al observed nystagmus may not be present when both anterior and posterior visual pathway defects are significant. In our study Oculomotor apraxia was seen in 6% of children.

Refractive error was seen in 39% of children. The most common refractive error observed was Compound Hypermetropic Astigmatism which contributed to 15% followed by simple hypermetropia, observed in 8%, simple myopia in 7% and Simple Myopic Astigmatism seen in 9%. Significant refractive errors must be corrected to improve the visual prognosis of these children.

Cerebral visual impairment was associated with Temporal Disc pallor (30%), optic disc atrophy (14%), large optic disc cup (3%) and 1 child had optic nerve hypoplasia. This is in accordance with the previous studies, which show the associated optic disc abnormalities in CVI are 24%-43%.[24] Suma et. al., study suggests that the cause of optic atrophy is Tran synaptic retrograde axonal degeneration. In our study, functional visual assessment was done by Roman–Lantzy CVI range.[5] According to the Newcomb et al. study, the CVI range is a very reliable method for quantifying functional vision as well as evaluating prognosis. The CVI Range has also been used for individualized visual intervention.[17] Niranjan et. al., study found that individualized visual intervention rather than visual stimulation improves the functional component of vision. The most common etiology contributing to CVI is Hypoxic Ischemic Encephalopathy (HIE) which contributes to 30% of cases. The other etiologies are Traumatic brain injury (11%), Congenital Brain anomaly (9%), Congenital Hydrocephalus (5%), Neonatal Hypoglycemia (3%), Post Cardiac Surgery (2%),[20] Infantile spasm (6%), Neonatal seizures (9%), CNS infections (6%) and Epilepsy (11%). This is in accordance with the previous studies in which the most common etiology contributing to CVI were Hypoxic-ischemic encephalopathy (47.2%), unknown (15.5%), Neonatal Seizures (11.3%), Epilepsy (7.8%), Hydrocephalus (4.9%), Neonatal hypoglycemia (4.2%), Infantile spasms (3.5%), Traumatic Brain Injuries (2.1%), Congenital anomalies of brain (2.1%) and stroke (1.4%).

The incidence of cerebral visual impairment was 87%. Thus, the incidence of CVI in children with perinatal hypoxic insult, associated neurological and congenital brain anomalies was as high as 87% in our study. This is in accordance with the previous studies.[27] In a study of preterm and term children with perinatal hypoxic insult,[13] Giovanni et al. discovered that children who had severe hypoxic insult with MRI findings of multi- cystic encephalopathy had severely diminished visual acuity, while PVL alone had normal visual acuity but with a visual field defect, and even those children who were considered normal had some deficit during schooling.

At the end of the follow-up period, 26 (26%) of the children had progressed from CVI Phase 1 and 2 to CVI Phase 3. 32% of children had improvement in the CVI range score. Catteno has said that in children with damage to the visual brain, training that is carefully matched to the child's profile, and that the child enjoys, wishes to join in and is spontaneously driven to complete, will enhance intact functions and redeploy other functioning brain areas to adapt additional compensatory abilities through neuroplasticity.

5. Conclusion

Even though CVI is one of the most common causes of visual impairment, it may go unobserved and undetected. With improved perinatal and paediatric care, the number of children surviving this neurological insult has greatly increased. Early identification and timely intervention form

a crucial part of Visual Rehabilitation. An individualized approach in the form of CVI Range assessment and visual intervention based on it should be integrated into many activities of the child across the whole day. Improvement in vision in these children creates access to a world of cognitive and motor development that makes what seems impossible not only possible, but likely.

6. Conflict of Interest

The authors declare no relevant conflict of interest with respect to research, authorship and or publication of this article.

7. Source of Funding

None.

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