



## EVALUATION OF VARIOUS CAUSES OF CEREBRAL VISUAL IMPAIRMENT IN CHILDREN AT TERTIARY CARE CENTRE

### Ophthalmology

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### ABSTRACT

**Purpose:** The purpose of this study is to evaluate the etiology, associated ophthalmological and neurological findings of cerebral visual impairment (CVI) in children at a tertiary care center.

**Methods:** A retrospective study was conducted on 71 patients visiting between February 2020 to August 2021 at a tertiary care hospital, J.L.N. medical college and hospital, Ajmer, Rajasthan. Diagnosis of CVI was based on ocular examination found poor visual function that could not be accounted by age, ocular findings and neuroimaging. **Results:** The records of 44 boys and 27 girls were analysed. Majority of them were <3yrs (40.8%). The most common etiology was perinatal hypoxia (29%), followed by prematurity (22.5%). Moreover, 9(12.7%) showed mixed aetiology, 8(11.3%) had hydrocephalus and 7(9.9%) had hypoglycemia. Amongst the ocular associations, refractive error was the most common, seen in 32 (45%) children, while strabismus in 31 (44%), nystagmus in 22(31%). Thirty two (45%) children had associated developmental delay, while 38(53%) had seizures. The most common findings were ischemic encephalopathy (31%), periventricular leukomalacia (23%), cortical and subcortical atrophy and structural malformations (24%).

**Conclusion:** CVI is of increasing importance as a cause of vision impairment in children. The major causes for CVI tend to be perinatal hypoxia followed by prematurity. Establishing the etiology for these children at early age may aid in early rehabilitation of these children.

### KEYWORDS

Cerebral visual impairment, developmental delay, ischemic encephalopathy, nystagmus, refractive error

### INTRODUCTION

Cerebral visual impairment (CVI) is vision loss as a result of damage to the retrogeniculate pathway (optic radiations, occipital cortex, and visual association areas) of the visual system in the absence of any ocular pathology. With the improvement of perinatal care and increased survival of the preterm babies, it emerged as one of the leading cause of visual impairment.<sup>1</sup>

CVI has multiple causes, but the most common cause is perinatal hypoxia. Other causes of pediatric CVI are meningitis, encephalitis, congenital central nervous system malformation, hydrocephalus, hypoglycaemia, trauma, cerebrovascular accidents, and rarely metabolic disorders.<sup>2,3,4,5,6</sup>

Other causes of CVI include shunt failure (which can lead to ischemia and occipital lobe infarction), antenatal use of cocaine and amphetamines by the mother, twin pregnancy, and epilepsy.<sup>7,8,9,10</sup>

CVI may have associated ophthalmological abnormalities including refractive errors, nystagmus and strabismus are common associations. Other associations are structural anomalies such as cataract, coloboma, optic atrophy, and retinal dystrophy have also been found.<sup>9,11</sup>

CVI is also associated with neurological deficits including cerebral palsy, mental retardation, and hemiparesis.<sup>3</sup>

### METHODS

We retrospectively studied all the children attending ophthalmology outpatient department of J L N Medical college, Ajmer between february 2020 and August 2021.

Our study included children aged <16 years diagnosed with CVI.

The diagnosis of CVI was based on an ophthalmic examination showed poor visual function bilaterally that could not be accounted for by age, structural ocular findings, optic atrophy, or high refractive error.

We also made a note of relevant birth history. This included stormy perinatal course, use of supplemental oxygen and its duration or ventilator support, anemia requiring transfusion, repeated episodes of hypoglycemia, which could be an indicator of sepsis, seizures, meningitis, or encephalitis.

Visual responses in infants and young children (<3years) who could

not identify letters or pictures had been qualitatively assessed by their quality of fixation and following.

In older children, the quantitative records (Snellen) were considered. Pupillary examination performed with a moderately bright torch light with dimmed room, was looked and recorded. Anterior segment examination findings of associated anomalies, if any, such as a developmental cataract or other media opacities and distant direct ophthalmoscopy were recorded. Data on dilated fundoscopy especially optic atrophy were also collected.

The findings of retinoscopy done preferably using homatropine 2%, were also noted.

The findings of any associated neurological abnormalities were also noted.

Further, findings of CT and MRI brain reports and VEP (if needed) in the records were studied and recorded.

### STATISTICAL ANALYSIS

The data obtained were subjected to statistical analysis in a prepared master with subdivided data. Statistical analysis was done by using Statistical Package of Social Science software (SPSS Version 20).

### RESULTS

Records of 71 children, diagnosed with CVI were reviewed. Of these, there were 44(61.9%) boys and 27(38%) girls. Majority of them 29(40.8%) were upto 3 years of age.

**Table 1: Age distribution of CVI study subjects (n=71)**

| Age group    | No. | %    |
|--------------|-----|------|
| Upto 3 years | 29  | 40.8 |
| 4-6 years    | 24  | 33.8 |
| 7-15 years   | 18  | 25.4 |

Mean age was 4.78±3.18 years

Visual function was poor in most children. 59 (83.1%) of children were suffering with poor vision, deviations of eyes in 14(19.7%), Thirty nine children were born full term delivery, 32 were premature delivery.

The ocular associations in our patients are summarized in Tables 2 and 3. Ocular findings were present in almost all the patients, with significant refractive error (>-2/+2 Ds) being the most common (45%)

followed by strabismus (43%). Of the 31 patients with strabismus, majority (21) had exotropia. Nystagmus was seen in 22(31%) patients. Fundus examination revealed that 34 of the 71 children (48%) had normal fundi, 26 had disc pallor (37%) and 8 had optic atrophy.

**Table 2: Type of refractive error in CVI study subjects (n=71)**

| Refractive error  | No. | %    |
|-------------------|-----|------|
| Absent            | 39  | 54.9 |
| Hypermetropia     | 16  | 22.5 |
| Myopia            | 8   | 11.3 |
| Mixed astigmatism | 8   | 11.3 |

**Table 3: Ocular manifestations in CVI study subjects (n=71)**

| Ocular manifestations                 | No. | %    |
|---------------------------------------|-----|------|
| <b>Posterior segment</b>              |     |      |
| Normal Disc                           | 34  | 47.9 |
| Disc oedema                           | 2   | 2.8  |
| Disc pallor                           | 26  | 36.6 |
| Optic atrophy                         | 8   | 11.3 |
| Multiple intraretinal haemorrhage +nt | 1   | 1.4  |
| <b>Strabismus</b>                     |     |      |
| Absent                                | 40  | 56.3 |
| Esotropia                             | 21  | 29.6 |
| Exotropia                             | 10  | 14.1 |
| <b>Nystagmus</b>                      |     |      |
| Absent                                | 49  | 69.0 |
| Present                               | 22  | 31.0 |

The neurological associations in our patients are summarized in Tables 4. Fifty four of the 71 patients had multiple neurological deficits. The common neurological findings were developmental delay (45%), seizures (53%) and Cerebral palsy (28%). Amongst the less common associations were third nerve palsy (1.4%) and hemiparesis (1.4%).

**Table 4: Associated neurological abnormalities in CVI study subjects (n=71)**

| Associated neurological abnormalities     | No. | %    |
|-------------------------------------------|-----|------|
| Developmental delay                       | 32  | 45   |
| Cerebral palsy                            | 20  | 28.2 |
| Third nerve palsy, Right side Hemiparesis | 1   | 1.4  |
| No abnormality                            | 18  | 25.3 |

When examined for the aetiological factors of CVI, it was found that, 21(29.6%) of children had perinatal hypoxia (most common), followed by prematurity was observed in 16(22.5%) children. Moreover, 9(12.7%) showed mixed aetiology, 8(11.3%) had hydrocephalus, 7(9.9%) had hypoglycemia summarized in Table 5.

**Table 5: Etiology of CVI in study subjects (n=71)**

| Aetiology of CVI                     | No. | %    |
|--------------------------------------|-----|------|
| Perinatal hypoxia                    | 21  | 29.6 |
| Prematurity                          | 16  | 22.5 |
| Mixed aetiology                      | 9   | 12.7 |
| Hydrocephalus                        | 8   | 11.3 |
| Hypoglycaemia                        | 7   | 9.9  |
| Epilepsy                             | 4   | 5.6  |
| Meningoencephalitis                  | 4   | 5.6  |
| CVA Secondary to septic endocarditis | 1   | 1.4  |
| Traumatic brain injury               | 1   | 1.4  |

All the patients undergone CT or a MRI which were done in different radiology centers which shows damage to both gray and white matter with cerebral, cortical, subcortical atrophy, and paucity of white matter. The most common findings were ischemic encephalopathy (30%), periventricular leukomalacia (23%), cortical and subcortical atrophy, and structural malformations (24%) such as agenesis or thinning of the corpus callosum.

18 patients had VEP records with them showing prolonged P100 and decreased amplitude.

## DISCUSSION

A total of 71 children were included in this study, out of which 44(61.9%) were male children and 27(38.0%) were female children who met our inclusion criteria. In our study, we observed more number of male children than female children. Khetpal et al<sup>8</sup> included 98

patients ranged from 0.2 to 19 years, with an average age of 3.1 years. There were 56 males and 42 females. Pehera et al.<sup>12</sup> included 428 severely visually impaired children aged 3 years with average age at presentation being 14.02 months. There were 264 (62%) male and 164 (38%) female children. Males were commonly affected in both studies, which is in accordance with our study.

Age wise distribution was also analyzed and observed that up to 3 years 29(40.8%) children were possessing the clinical condition, as the age proceeded, between 4 to 6 years, the number of children affected were found to be decreased. In a recent study done by Geeta V K P et al<sup>13</sup>, Hyderabad, has stated that, it was found that CVI was the most common cause of profound visual impairment in <3 years of age (33%). our study is in accordance with it. In another study performed by Huo R et al<sup>14</sup>, reviewed the patients records and from 170 CVI cases, they observed that, the average age of patient was 3 years, which is in accordance with our study.

Children with CVI can have a myriad of ocular manifestations. The results from our study highlighted that the major causes of visual impairment were refractive error (45%) and strabismus (esotropia > exotropia) (44%). While this was true for the majority, the ones more refractory to treatment, like disc pallor (36%) and nystagmus (30%), also contributed to the spectrum. This was in conjunction with findings from similar studies by Khetpal and Pehera et al.<sup>8,12</sup> in which, refractive errors followed by strabismus, and optic atrophy were found to be the most common ocular associations.

The associated neurological deficits were reported in our study, was seizures (54%) and developmental delay(45%) was the most common neurological association followed by cerebral palsy (28%), and hemiparesis (1.4%). This was comparable to the finding from Huo, Good, Hoyt, Khetpal et al.<sup>14,8</sup> though grossly analogous. Seizures followed by cerebral palsy were found to be the major neurological association. However, the statistical results of a similar study in the Indian subcontinent, by Pawar et al.<sup>15</sup>, varied in the order of frequency found developmental delay and seizures to be the most common neurological associations.

When examined for the aetiological factors of CVI in our study, it was found that, 21(29.6%) of children had perinatal hypoxia as most common etiology, later prematurity was observed in 16(22.5%) of CVI children. Moreover, 9(12.7%) showed mixed aetiology, 8(11.3%) had hydrocephalus and 7(9.9%) had hypoglycemia. This was comparable to the results of a few similar studies. Huo et al. reported that the four most common causes of CVI were perinatal hypoxia (22%), cerebral vascular accident (14%), meningitis (12%), and acquired hypoxia (10%).<sup>6</sup> While Khetpal et al.<sup>8</sup> reported perinatal hypoxia (35%), prematurity (29%), hydrocephalus (19%), structural central nervous system abnormalities (11%) as the common aetiologies. Many children (69%) had multiple aetiologies. Pehera et al.<sup>12</sup> found perinatal hypoxia to be the most common aetiological association.

The most common findings of neuroimaging in our study were ischemic encephalopathy, followed by periventricular leukomalacia and cortical and subcortical atrophy and structural malformations, which is similar to study done by Khetpal and Donahue.<sup>8</sup>

The main limitation of our study is that prognosis of vision of children with CVI were not assessed. Long-term follow-up of these children could give different results of visual prognosis. This study is a retrospective study performed in a single hospital with limited number of CVI patients. However, compared with other study parameters, we have successfully evaluated all the selected parameters, but could not accomplish appropriate statistical analysis to rule out other comorbidities contributing this disease.

## CONCLUSION

CVI is of increasing importance as a cause of vision impairment in children. The major risk factors for CVI tend to be perinatal hypoxia, prematurity followed by mixed etiology, structural malformations and hypoglycaemia. Establishing the etiology at early age in them may aid in early rehabilitation of these children.

In this present study, the selected parameters remained as strength of this study, we could assess that, age if children, gender, CP, HIE and perinatal hypoxia were significantly associated with the disease condition. Most of the studies have selected these parameters,

however, we correlated the logical explanation behind selecting these clinical parameters and even we have route out the mechanism involving in the etiopathogenesis. Further, it provides a scope to designing future studies and implementing neuroprotective strategies for treatment of this high-risk premature population.

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