Original Article

Cerebral visual impairment in children: Causes and associated ophthalmological problems

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Purpose: The aim of this study is to identify common causes, associated ophthalmological abnormalities, and systemic comorbidities in children in Andhra Pradesh, India, with cerebral visual impairment (CVI). **Methods:** A retrospective review of case records of all children aged <16 years with diagnosis of CVI seen between January 2016 and December 2016 was carried out. Data were collected for their age, gender, cause of CVI, refraction, accommodation, anterior and posterior segment examination findings, and systemic problems. **Results:** A total of 124 patients were identified and studied (80 boys and 44 girls, mean age 5.23 years, 44.8% aged <2 years). The most common causes of CVI were hypoxic–ischemic encephalopathy (HIE) (34.4%), undetermined etiology (32.8%), neonatal seizures, and infantile spasms (16% each). The most common presenting complaints were poor vision (76%) and squint (11.2%). Profound visual impairment was seen in 88.8%, and 11.2% had high functioning CVI. Fifty-eight (46.4%) patients had significant refractive errors, 40 (32.25%) had strabismus, 4 (3.2%) had visually significant cataract, and 40 (32%) had optic atrophy. Motor delay was observed in 39.5%, speech delay was evident in 22.4%, and cognitive delay in 16%. **Conclusion:** HIE is the most common cause (one-third) of CVI in our population, and the majority of them presented at age <2 years (44.8%) with profound visual impairment (88.8%). A significant number of them have treatable ophthalmic conditions such as refractive errors (46.4%), accommodative insufficiency (12.1%), and cataract (3.2%), and more than one-third of them also have delay in other areas of development.

Key words: Accommodative insufficiency, cerebral visual impairment in children, developmental delay, refractive errors

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Cerebral or cortical visual impairment (CVI) is an overarching term covering a wide range of visual and perceptual visual impairments resulting from dysfunction, anomaly or injury to the retrogeniculate visual pathways (optic radiations, occipital cortex, and visual association areas), and oculomotor control, which is more profound if the thalamus is affected.[1] CVI has recently become the most common cause of visual impairment in children in developed countries.[2-11] This is probably due to improved management of treatable causes of childhood blindness including cataract, glaucoma, and retinopathy of prematurity (ROP)[12] combined with improved intensive care of infants and children with cerebral pathology.[13] Improved diagnosis and increased reporting of this condition may also have led to this reported increase.[14]

In developing countries like India, increasing numbers of premature infants and babies with perinatal brain injury are surviving, leading to an increasing prevalence of CVI in children. Refractive errors and anomalies of accommodation are common in those affected.[15,16] As a part of the syndrome, structural problems such as cataract, coloboma, optic atrophy, and retinal dystrophy may coexist.[17,18] ROP may be seen in association with periventricular white matter injury. Optic nerve hypoplasia and optic atrophy are associated with a wide range of brain disorders, many of which impair visual function.[19] Disorders of eye movement control, including strabismus, nystagmus, unstable fixation, inaccurate fast eye movements (dysmetric saccades), deficient smooth pursuit movements, and paroxysmal deviations, in which the eyes intermittently deviate upward (most commonly) are common in children with CVI. The problems with visually guided eye movements can be compensated for in part by head movements (in children who have head control).[1,20]

Since it is often difficult to examine such children, some of these issues may be missed. Even if they are detected, the child may not receive appropriate management if those responsible for looking after the child believe that the strategies suggested would prove ineffective. Above all, in countries with limited

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resources, many of these children do not receive an eye examination.

The purpose of this study was to understand common causes of CVI in our population and common ophthalmic problems that they have.

Methods

We performed a retrospective review of case records of children aged < 16 years with a diagnosis of CVI, seen between January 2016 and December 2016 at The David Brown Children's Eye Care Center, L V Prasad Eye Institute, Kode Venkatadri Chowdary Campus, Vijayawada, Andhra Pradesh, India. The diagnosis of CVI was made when the ophthalmological findings did not explain the visual performance of the child, and there was associated neurological pathology. The following details were noted for each case: age at presentation, presenting complaint, visual acuity, cycloplegic refraction, ocular alignment and motility, and anterior and posterior segment findings. Significant refractive error was identified as per the preferred practice pattern published by the American Academy of Ophthalmology, with one modification for the hypermetropia, where we considered + 3.0 diopter sphere as a significant error, considering the fact that in children with developmental delay, low hypermetropia also becomes visually significant.[21] Cases with no significant ophthalmic pathology were labeled as isolated CVI and those with significant associated ophthalmic pathology (e.g., myopia >6D, hypermetropia >5D, and media opacities such as visually significant cataract and optic atrophy) were classified as mixed ocular visual impairment (OVI) with CVI.

Results

During the study period, a total of 124 children with a diagnosis of CVI were seen. Eighty were boys and 44 were girls. The median age at presentation was 3 years (mean 5.24, standard deviation [SD] 4.61). Fifty-six (45.16%) were aged < 2 years, and among them 36 (29.03%) were infants.

The presenting complaint in the majority (95 children, 76.61%) was poor vision. Fourteen (11.3%) presented with ocular misalignment (strabismus). White reflexes from eyes (leukocoria) were noted by parents in three cases (2.42%). Some of the uncommon presenting complaints were frequent fall or tripping over things on the floor while walking (6), difficulty in walking up and down the stairs (3), slow visual responses (1), and inability to reach out to objects accurately (1).

The most common cause of CVI in our group was hypoxic–ischemic encephalopathy (HIE) seen in 50 cases (40.32%). In a significant number of children (40 cases, 32.26%) with CVI, the etiology remained unknown. Sixteen children (12.9%) had epilepsy. A history of neonatal seizures was present in 24 cases (19.36%). Other less common causes included neonatal hypoglycemia (4 cases, 3.23%), meningitis (3 cases, 2.42%), hydrocephalus (3 cases, 2.42%), and stroke (4 cases, 3.23%). Apart from these, other neurological diagnoses were cerebral palsy in 59 (47.6%) children, metabolic storage disorders in 5 (4%), and brain malformations in 2 (1.6%) cases.

Fifty-four children (43.55%) were referred by pediatricians. Eighty-four (67.74%) were born at term and forty (32.23%) were born prematurely (gestational age <34 weeks at birth). There was a history of difficulty breathing/delayed birth cry in 71 (57.26%). The mean birth weight was 2.53 kg (SD 0.76).

Sixty-one children (49.2%) had strabismus. Thirty-nine had exotropia (mean deviation 36.2 prism diopters) and 22 had esotropia (mean deviation 29.25 prism diopters). Nystagmus was found in 9 children (7.3%).

We found significant refractive error in 62 (50%) children. Compound myopic astigmatism was the most common refractive error seen in 44 children (35.48%). Five (4.03%) had high myopia (>−6.0 DS) and 6 (4.89%) had high hypermetropia (>+5.0 DS) (4 following cataract surgery). Accommodative insufficiency was present in 15 (12.1%) children, as assessed by dynamic retinoscopy. None of these children were wearing glasses at the time of presentation.

The most common anterior segment anomaly was cataract that was present bilaterally in 5 (4.03%) children (10 eyes), and morphological subtypes included total cataract (7 eyes), anterior subcapsular (2 eyes), and visually insignificant sutural cataract (1 eye). One child had bilateral lower eyelid colobomata and one child had oculocutaneous albinism. The most common posterior segment anomaly found was optic atrophy, seen in 40 cases (32.26%). Thus, we had a total of 53 (42.7%) cases of isolated CVI and 71 cases (57.3%) of mixed CVI and OVI (accommodative insufficiency 15, high myopia 5, high hypermetropia 6, visually significant cataract 5, optic atrophy 40, and oculocutaneous albinism 1).

We could get reliable visual fields by automated perimetry (Humphrey Field Analyzer) in 4 children. In two of them, there was right homonymous hemianopia and two had inferior arcuate scotoma in both the eyes. In the rest of the children, depending on the level of cooperation by the child, we used confrontation testing, the leg raise test, Puppet faces, and Lea wand test. We also enquired about any history suggestive of lower visual field impairment, such as tripping over things on the floor and difficulty in walking down the stairs. By all these methods, lower visual field impairment was found in additional 10 children. Hence, in total, 12 (9.68%) children had lower visual field impairment and 2 (1.61%) had right homonymous hemianopia.

Seventy-one children (57.26%) had delay in one or more areas of development (motor delay in 49, speech delay in 28, cognitive delay in 20, and global delay in 29 children).

Discussion

The most common cause of CVI in this series was HIE. Majority of the children were aged <2 years at presentation and had delay in multiple areas of development. About half had treatable ophthalmic conditions such as significant refractive error, accommodative insufficiency, and strabismus. Four had cataract which required surgery. However, none of these problems had been known to the family. This highlights the need for regular and comprehensive eye examination for children with brain damage.

Our findings are in accordance with previous studies which have reported the prevalence of refractive errors

in such children to be in the range of 10%–60%.[2-12] Accommodative insufficiency in children is frequently underdiagnosed due to lack of awareness. In this series, 12% of children were affected, and hence, it should be actively looked for in every child with CVI. These are easily treatable conditions, which should be picked up earlier in life to avoid amblyopia. But often, such children do not receive a comprehensive eye examination; since they are unable to cooperate well, it is time-consuming and sometimes the resources and workforce may be limited. In addition, it may be perceived that the correction of refractive error or performance of cataract surgery may not benefit the child, due to his neurological condition. This is arguably incorrect, and in fact, it adds an avoidable disability on top of a preexisting disability. It was our experience that on account of these issues, almost all of our patients had not previously received a complete eye examination and consequently did not get appropriate help for their visual dysfunction at an earlier stage. Hence, there is an acute need to create awareness about CVI in the ophthalmic community as well as in society in general. Since the data show that in 57.3% cases, there was combined OVI and CVI, it is important for ophthalmologists to be aware of this combination too. When treatment of the cause of OVI does not improve vision to the desired level, the additional component of CVI should be suspected, especially when the child is known to have a neurological ailment or developmental delay, and necessary steps need to be taken to optimize the visual experiences of the child.

In the fact that the majority of these children were aged < 2 years at presentation and had delays in multiple areas of development along with visual disorders, there is a need for pediatric ophthalmology units to equip themselves with the necessary skills and facilities to manage such children. Such a learning experience is usually not available during most postgraduate and fellowship training programs. This may render clinicians underconfident while dealing with such children. In view of the increasing prevalence of children with neurological issues, there is a need to include this element as an integral part of all ophthalmology training programs.

Since most of the skills impacted as part of delayed development are also vision dependent, it is important that the experts handling such issues understand how the child sees, and how any visual deficiency compounds the disability, so that they can take appropriate action and render material accessible for learning. Hence, it would be ideal if pediatric ophthalmology services also offer services for motor, speech, and cognitive delay under one roof, without requiring children to move from one place to another. We have found that this approach extends the formal scope of ophthalmic service delivery but benefits such children a lot.

Conclusion

In summary, this study emphasizes the importance of comprehensive eye examination of every child with neurological problems and the need for a team approach to address challenges in other areas of development that are vision dependent. We suggest that any neonate with a stormy perinatal course of events should have an assessment of vision and an eye examination at 3 months of age and any child with a cerebral neurological ailment should undergo eye examination soon after the diagnosis when the systemic condition of the child permits.

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Conflicts of interest

There are no conflicts of interest.

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