

# Cerebral visual impairment is a major cause of profound visual impairment in children aged less than 3 years: A study from tertiary eye care center in South India

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**Purpose:** The purpose of this study was to evaluate causes for profound visual impairment in children  $\leq 3$  years of age at a tertiary eye care center in Andhra Pradesh, India. **Methods:** A retrospective study was conducted for all the children ( $\leq 3$  years) who attended the pediatric ophthalmology service between January 2012 and February 2017. **Results:** A total of 428 severely visually impaired children aged  $\leq 3$  years were seen during the study period: 264 (62%) of them were boys and 164 (38%) were girls. The average age at presentation was 14.02 months. The causes of visual impairment were cerebral visual impairment (CVI) 142 (33%), a combination of CVI and ocular visual impairment (OVI) 48 (11%), and OVI only 236 (56%), which included congenital cataract 56 (13.1%), retinopathy of prematurity 52 (12.6%), optic atrophy 17 (4.5%), congenital nystagmus (4.4%), congenital globe anomalies 21 (5.2%), and high refractive errors - 10 (2.8%). Delays in different areas of development were seen in 103 out of 142 children with CVI (72.5%), which included motor delay 53 (51.5%), cognitive delay 15 (14.6%), speech delay in 3 (2.9%), and delay in multiple areas of development (like combination of motor, cognitive, and speech delay) in 32 (31.1%). **Conclusion:** In children under 3 years of age, CVI is a major cause of profound visual impairment in our area and the majority of them manifest delay in several areas of development.

**Key words:** Age less than 3 years, cerebral visual impairment, profound visual impairment, south India

Children with blindness account for 3.1% (1.4 million) of the global blind population. Although this number is relatively small, the larger number of future blind years resulting from childhood blindness, as compared to adult blindness, makes it a critical form of blindness needing attention. Approximately, 5,00,000 children become blind every year and 70 million blind person-years are added each year due to childhood blindness.<sup>[1]</sup> After cataract, this ranks second in cumulative blind person-years contributing to worldwide figures.<sup>[2]</sup> Childhood blindness has a significant impact on the development of the child encompassing psychomotor, educational, social, and emotional aspects of development. Blind children in developing countries also have a higher death rate than their sighted counterparts, and 60% are thought to be dying within the first year of becoming blind.<sup>[1]</sup> Hence, the World Health Organization has included childhood blindness as one of the five priority areas under Vision 2020, the global initiative to eliminate avoidable blindness by the year 2020.<sup>[3]</sup>

As a first step to tackle the problem, it is necessary to have reliable epidemiological data regarding childhood blindness

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because the strategies for control of childhood blindness are significantly different from those for adults. The prevalence and causes of childhood blindness vary from country to country, reflecting the socio-economic development and differences in the health care delivery systems. In addition, a multidisciplinary team approach is required, needing specific training, expertise, and equipment at different levels.

For effective planning to deal with childhood blindness in an area, it is important to understand the common causes of the same in that area. This helps in deciding on priorities and allocating resources. Vijayawada is a tier 3 city located in the South Indian state of Andhra Pradesh. Being a tertiary referral center, we get children from a surrounding population of 5.5 million people. We have a dedicated Children's Eye Care Center and an Early Intervention Unit, which provide rehabilitation services to young children with irreversible visual impairment.

Studies from India report the following conditions as common causes of childhood blindness: congenital globe anomalies (microphthalmos/anophthalmos), cataract/lens related, corneal opacity (including vitamin A deficiency),

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retinal dystrophies, optic atrophy, glaucoma, amblyopia owing to high refractive error, and abnormality of higher visual pathways.<sup>[4-15]</sup>

Data from the developed countries suggest that cerebral/cortical visual impairment (CVI) and optic neuropathy have recently become the most common causes of visual impairment in children.<sup>[16-25]</sup> This is probably owing to better management of avoidable causes of childhood blindness such as cataract, glaucoma, and retinopathy of prematurity (ROP) in children<sup>[26]</sup> combined with improving survival of children who survive severe neurological damage during the perinatal period.<sup>[27]</sup> Improved diagnosis and reporting of this condition also may have led to this increase.<sup>[28]</sup>

The first 3 years of life are of great importance to the overall development of the child. For optimal development, good vision is necessary because most of the tasks learned in this period are vision dependent. From that perspective, we wanted to find out the common causes of severe visual impairment/blindness in this age group in our set-up.

## Methods

This was a single center, hospital-based study conducted at a tertiary level referral center in south India. We performed a retrospective review of all children aged less than 3 years at the time of first examination at our service seen by a single pediatric ophthalmologist, between February 2012 and February 2017 (identified from electronic medical records). Any child who could not look at the largest grating card on the Teller Acuity Card test (0.23 cycles/cm at 38 cm) was considered as severely visual impaired. The following data were collected for each child: age, findings on ocular examination, and the principal cause of visual impairment. In instances of multiple causes of visual impairment, the pediatric ophthalmologist ascertained the primary and contributory causes of visual impairment. We divided affected children into 3 groups depending on their causes of visual impairment- children with ocular causes of visual impairment (OVI), cerebral causes of visual impairment (CVI)- 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 as evidenced by reports from pediatric neurologists/neuroimaging/history of birth asphyxia, epilepsy, stroke, hydrocephalus, etc) and combinations of both (CVI+OVI- where significant ophthalmic pathology was present but did not explain visual performance of the child and there was associated neurological pathology). Systemic comorbidities such as motor, speech, hearing, and cognitive impairment were recorded, assessed, and diagnosed by relevant professionals in our team including referring pediatric neurologists/pediatricians and the in-house physiotherapist, speech therapist, and special educator.

## Results

A total of 428 severely visual impaired young children aged less than 3 years were seen during the study period: 264 (62%) of them were boys and 164 (38%) were girls. The age range was between 4 months to 3 years (median 13 months).

CVI was a leading cause of severe visual impairment followed by congenital cataract and retinopathy of prematurity. Details are given in Table 1.

One hundred and thirteen of 142 children (79.6%) with CVI were referred to us by treating physicians (87 by Pediatric Neurologists/Pediatricians and 26 by Ophthalmologists) and 29 (20.4%) were brought by parents on their own to us. The causes of CVI were hypoxic-ischemic encephalopathy 67 (47.2%), unknown 22 (15.5%), neonatal seizures 16 (11.3%), epilepsy 11 (7.8%), hydrocephalus 7 (4.9%), neonatal hypoglycemia 6 (4.2%), infantile spasms 5 (3.5%), traumatic brain injuries 3 (2.1%), congenital anomalies of brain 3 (2.1%), and stroke in 2 (1.4%). Findings of neuroimaging (MRI brain, CT brain, or neurosonogram) were available in 112 (78.9%) children, which included periventricular leucomalacia, infarcts involving occipital or parieto-occipital areas, enlargement of lateral ventricles, cystic encephalomalacia, thinning of basal ganglia, thinning of corpus callosum, cortical atrophy, subdural hemorrhage, intraventricular hemorrhage, and the congenital anomalies such as schizencephaly, lissencephaly, and porencephaly. Neuroimaging was normal in 10 of the children with clinical features of CVI, 5 of them had a history of neonatal seizures, 2 had epilepsy, and in 3, the cause of CVI was unknown.

Seventy-one out of 142 children (50%) with CVI had significant refractive error on cycloplegic refraction with 1% cyclopentolate and 0.5% tropicamide. Different types of refractive errors seen were compound hypermetropic astigmatism in 23 (32.4%), simple myopic astigmatism in 18 (25.4%), compound myopic astigmatism in 16 (22.5%), simple hypermetropia in 10 (14.1%), simple myopia in 2 (2.8%), and mixed astigmatism in 2 (2.8%). Ocular alignment could be assessed reliably in 112 children (78.9%) and 54 (48.2%) of them had a squint (36 exotropia and 18 esotropia). One hundred and seventeen of them (82.4%) had optic nerve head pallor with normal retinal findings, whereas the cause was hypoxic-ischemic encephalopathy in 100 (85.5%) children identified by neuroimaging. Other causes of optic nerve head pallor included hydrocephalus (7, 6%), stroke (2, 1.7%), traumatic brain injury (2, 1.7%), and unexplained (6, 5.1%).

We found that the prevalence of CVI is similar among children belonging to rural 34 (42.3%) and urban 56 (50.5%) areas ( $P = 0.10$ ).

Delays in different areas of development were seen in 103 out of 142 children with CVI (72.5%), which included motor

**Table 1: Causes of profound visual impairment**

Cause of Profound Visual Impairment	Number (percentage)
Cerebral Visual Impairment (CVI)	142 (33%)
Cerebral Visual Impairment (CVI) + Ocular Visual Impairment (OVI)	48 (11%)
Ocular Visual Impairment (OVI)	236 (56%)
Congenital cataract	56 (13.11%)
Retinopathy of prematurity (ROP)	52 (12.64%)
Optic atrophy	17 (4.48%)
Microphthalmos	21 (5.15%)
High refractive errors (+/- > 6.0 Dsph)	10 (2.81%)
Others, (Leber's congenital amaurosis, retinitis pigmentosa, aniridia, uvealcoloboma, and infantile nystagmus syndrome)	48 (11.21%)

delay 53 (51.5%), cognitive delay 15 (14.6%), speech delay in 3 (2.9%), and delay in multiple areas of development (such as combination of motor, cognitive, and speech delay) in 32 (31.1%).

Interventions included ophthalmic interventions such as prescription of glasses for significant refractive error or hypo-accommodation and strabismus surgery for constant uniocular deviations. Rehabilitational measures were directed by the functional vision assessment, with emphasis on incorporating the child's vision into daily routine activities. A team comprising a pediatric ophthalmologist, optometrist, vision rehabilitation specialist, physiotherapist, speech therapist, and special educator, took care of these children under one roof in a transdisciplinary manner. Out of 142 children with CVI, 47 had a follow-up of more than 6 months. Among them, 21 showed significant improvement in visual functioning, 10 showed mild improvement, and 16 showed no change.

## Discussion

We found that in our area, CVI is a leading cause of visual impairment among young children aged less than 3 years, exceeding congenital cataract and retinopathy of prematurity. The prevalence of CVI among children from rural and urban backgrounds was no different, and close to three-fourth of them showed a delay in other areas of development.

Refractive error was identified in half of the children with CVI; this proportion is very similar to that for cerebral palsy, showing that every affected child must be refracted and managed appropriately.<sup>[29]</sup>

The emergence of CVI as one of the most common causes of profound visual impairment could be owing to better survival of several premature newborns and of those with "stormy" perinatal courses of events. However, because of increased availability of tertiary level pediatric eye care facilities; cataract, ROP, and glaucoma are becoming less common as causes of childhood blindness.

Previous studies from India show that 4.6 to 24.2%<sup>[7-15]</sup> children were blind due to optic atrophy, but the cause of optic atrophy is not mentioned in these papers. It is possible that in many of these cases, the optic atrophy may have been due to retrograde trans-synaptic neuronal degeneration following an injury to the brain, which is a well-established entity.<sup>[30]</sup> Thus, many of the reported cases of childhood optic atrophy may have had undiagnosed CVI. In our series, the cause of optic atrophy was hypoxic-ischemic encephalopathy in 100 children. This emphasizes the fact that optic atrophy is a sign that requires further investigations to find its cause and that optic atrophy cannot be considered a diagnosis in its own right. Major studies on childhood blindness from India have not hitherto included CVI as a diagnosis; hence, we have no data on its prevalence. In cases with optic atrophy, there is often a dilemma as to whether the cause of visual impairment is OVI due to optic atrophy or CVI. In such cases, we look for any cause in the eye, which can lead to optic atrophy (e.g., retinal dystrophy). If we find any ocular cause for optic atrophy, then we consider it as OVI. In the absence of such ocular cause and if there is a central nervous system (CNS) lesion, which can cause optic atrophy due to retrograde trans-synaptic degeneration,

then we considered the cause of visual impairment in such cases as CVI. In cases such as traumatic brain injury with lesions affecting the occipital cortex, where there was co-existing traumatic optic neuropathy, then we considered the cause of visual impairment to comprise both OVI and CVI.

The association of profound visual impairment with delay in several areas of development is explained by the fact that most of these children have suffered from damage to multiple areas of the brain as detailed in the results section. Being aware of such delays and guiding them to respective specialists is very important. Managing such complex issues of severe visual impairment with developmental delay requires a team approach with specialists in each of these areas working together.

We receive referrals of significant numbers of children from rural areas. It was surprising that the CVI was almost equally common among children belonging to rural and urban areas. The challenges involved in managing such children from disadvantaged places are greater than in urban areas thus the factors in urban areas leading to equal levels of CVI in the two communities warrant investigation.

These findings emphasize the need to equip ophthalmologists with the necessary skills to diagnose and manage CVI in young children and also to develop partnerships with specialists from other specialties involved in the care of the child including neurologists, physiotherapists, occupational therapists, speech therapists, and special educators. It is an encouraging development in our country that the Government of India has already established 92 District Early Intervention centers as a part of the Rashtriya Bala Swasthya Karyakram initiative and 366 more have been approved for different parts of the country, where such therapists are available under one roof at no cost to the families.<sup>[31]</sup> Awareness about this facility among ophthalmologists is important for appropriate referral of children with CVI to these DEICs for the various therapies and habilitational approaches that they need.

One major limitation of the study is that this is a single center study from a tertiary eye care center, and we run a separate clinic for children with CVI. This may attract more children with CVI to us, and thus, skew our data toward an apparently greater prevalence of CVI. Notwithstanding, we believe our findings reflect the impression of several pediatric ophthalmologists in our country that CVI is emerging as a major cause of visual impairment in children. To understand the nature, current incidence and prevalence of CVI in India, we need a larger epidemiological study involving major eye institutes across the country.

## Conclusion

In summary, CVI is a leading cause of severe visual impairment in children aged less than 3 years in our unit, and the majority of them also have delay in other areas of development. This calls for an emerging need to equip eye care facilities with the necessary systems to cater to the needs of such children.

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## Conflicts of interest

There are no conflicts of interest.

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