Original Article

Profiles of Cortical Visual Impairment (CVI) Patients Visiting Pediatric Outpatient Department

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Abstract

Introduction: Cortical visual impairment denotes vision loss from pathology posterior to the lateral geniculate nucleus. The pathology may involve the optic radiations, as well as the occipital cortex.

Objective: To find out the profiles of cortical visual impairment patients visiting the pediatric outpatient department.

Materials and Methods: The study is a hospital based retrospective study in which all consecutive patients diagnosed with cortical visual impairment were included. A total of 40 patients were collected. Detailed history taking and clinical examination was done. Visual acuity was taken by fixation and follows method. Among 40 patients, only two patients were advised to use glass and the rest did not have significant refractive error. Myopia ranged from (-2D to -5D) and five patients were myopic. Astigmatism ranged from (-0.5 to -2.5 x 108°) and 10 patients had astigmatism. Suspected patients were advised for Computed Tomography/ Magnetic Resolution Imaging (CT/MRI) of the brain.

Results: The male: female ratio was 3:2, the age group ranged from 4 months to 8 years old, antenatal checkup history was uneventful in 77.5% cases, history of birth asphyxia was present in 87.5% cases, postnatal checkup history was eventful in 67.5%, associated systemic illness was present in 60%, anterior segment examination was normal in 92.5%, posterior segment examination was normal in 72.5%, CT/MRI findings were abnormal in 57.5% and was not done in 30% of cases. Antenatal history was described as uneventful if there was absence of diabetes mellitus, hypertension, fever and intake of any medicine. Postnatal history was described as uneventful if there was absence of febrile convulsion, meningitis, encephalocele, encephalopathy, epilepsy or hydrocephalus.

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Conclusion: Birth asphyxia and postnatal infections are the major causes for cortical visual impairment. We can mitigate cortical visual impairment by limiting birth asphyxia and postnatal infections.

Key words: Cortical visual impairment, Birth asphyxia, Febrile convulsion.

Introduction

Cortical visual impairment (CVI) is a condition of bilateral visual loss from pathology posterior to the lateral geniculate nucleus. The pathology may involve the optic radiations as well as the occipital cortex. CVI is usually brain-based; always involves the neural pathways and/or the brain itself. Visual information is relayed through the eye as it should be; however, the brain cannot always make sense of the information it receives (Morse, 1999; Baker-Nobles & Rutherford, 1995; Jan & Groenveld, 1993; Flodmark et al, 1990). The most common causes of CVI are hypoxic/ischemic and lack of oxygen to the brain which usually occur during or shortly after birth from complications of prematurity or other gestational/delivery difficulties. The other causes of CVI include cardiac arrest/respiratory failure, increased intracranial pressure, head trauma, hydrocephaly, and/or shunt failure which leads to decrease of cortical oxygen and/or damage brain tissues. Congenital brain malformations secondary to genetic syndromes and/or other birth defects are also implicated in the incidence of CVI, as are CNS infections like meningitis, cytomegalovirus, encephalitis, and herpes simplex. Poisoning, certain drug exposures (e.g., Cisplatin), various sedating anticonvulsant drug therapies, carbon monoxide poisoning, intrauterine cocaine exposure, and accidental ingestion of other drugs or chemicals can also cause or exacerbate CVI. Finally, secondary complications such as seizures, metabolic diseases, hypoglycemia, and progressive genetic syndromes may cause or intensify cortical visual impairment. Brain malformations, head injury, CNS infections like CMV infection, meningitis, poisoning/drug exposure, birth trauma, cerebral palsy, seizures/epilepsy leading to lack of oxygen to brain and causes CVI (Good et al, 1994; Jan & Wong, 1991; Wong,1991; Flodmark et al, 1990; Groenveld et al, 1990).

Methodology

All records of patients diagnosed as CVI visiting pediatric OPD of Tilganga Institute of Ophthalmology from 2015 September to 2016 September were included in the study retrospectively. Ethical clearance was obtained from the Institutional Review Board of Tilganga Institute of Ophthalmology. History taking was done in detail. Antenatal history about fever or any other disease and history of per vaginal bleeding during pregnancy were taken. Detailed history during delivery like prolonged labor, meconium stain, birth asphyxia were taken. Postnatal history of high grade fever, seizures etc were taken in detail. Then visual acuity test and retinoscopy examination was done. Visual acuity was taken by fixation and follows method. Detailed eye examinations and fundus examinations were carried out. Then patients were advised for CT scan or MRI brain. Patients were also advised to consult a pediatrician if needed. Statistical analysis was done with Statistical Package for the Social Sciences version 20.

Results

The male: female ratio was 3:2 (Figure 1), antenatal checkup history was uneventful in 77.5% cases (Figure 2, Table 1), history of birth asphyxia was present in 87.5% cases (Figure 3), postnatal checkup history was eventful in 67.5% (Table 2, Table 3), associated systemic illness was present in 60% (Figure 4, Table

4), anterior segment examination was normal in 92.5%, posterior segment examination was normal in 72.5% (Table 5, Figure 5), Computed Tomography/ Magnetic Resolution Imaging findings were abnormal in 57.5% (Figure 6) and was not done in 30% of cases.

Table 1: Specification of antenatal history of mothers of patients who developed cortical visual impairment

Specification of events	Frequency	Percent
Diarrhoea	1	11.1
Diabetes, Hypertension	1	11.1
Fever (non-specific)	2	22.2
Fever (Typhoid)	1	11.1
Gestational Hypertension	2	22.2
Intake Of medicine	1	11.1
Urinary tract infection	1	11.1
Total	9	100.0

Table 2: Postnatal history of patients who developed cortical visual impairment

Postnatal history	Frequency	Percent	p -value
Eventful	27	67.5	0.027
Uneventful	13	32.5	
Total	40	100	

Table 3: Specification of eventful postnatal history of patients who developed cortical visual impairment

Events of postnatal history	Frequency	Percent
Cerebral palsy	3	11.1
Encephalocele	1	3.7
Encephalopathy	1	3.7
Epilepsy	2	7.4
Febrile convulsion	3	11.1
Hydrocephalus	1	3.7
Meningitis	1	3.7
Meningitis, hydrocephalus	1	3.7
Microcephaly, secondary craniosynostosis	1	3.7
Neonatal jaundice	1	3.7
Neonatal sepsis	1	3.7
Obstructive hydrocephalus, craniopharyngioma	1	3.7
Seizure disorders	10	37.0
Total	27	100.0

Table 4: Specification of systemic illness in patients with cortical visual impairment

Systemic illness	Frequency	Percent
Cerebral palsy	3	12.5
Congenital heart disease	1	4.2
Encephalocele	1	4.2
GDD with encephalopathy	1	4.2
GDD with febrile convulsion	2	8.3
GDD with microcephaly with secondary crainocynostosis	1	4.2
GDD with seizure disorders	2	8.3
Hydrocephalus	3	12.5
Meningitis, hydrocephalus	1	4.2
Seizure disorders	9	37.5
Total	24	100.0

Table 5: Specification of fundus examination in patients who had abnormal findings

Findings	Frequency	Percent
Pale disc	1	9.1
Pale disc, myopic fundus	1	9.1
Temporal pallor	9	81.8
Total	11	100.0

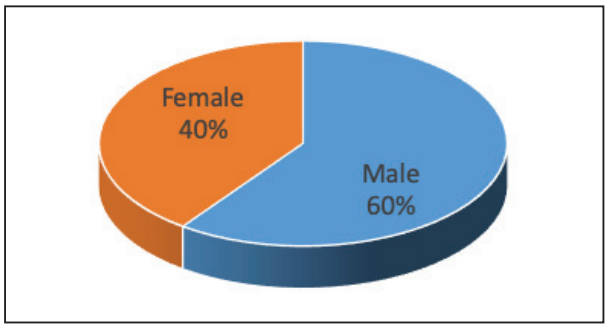

Figure 1: Gender distribution of patients with cortical visual impairment

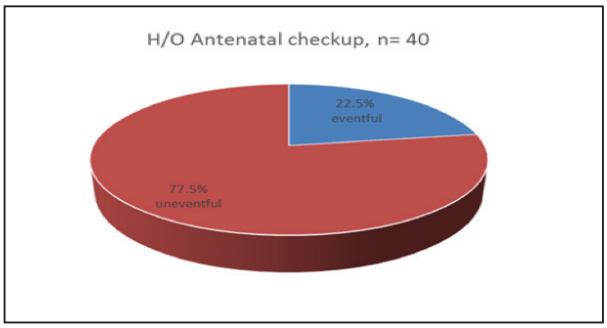

Figure 2: Antenatal checkup history (ANC) of mothers of patients who developed cortical visual impairment

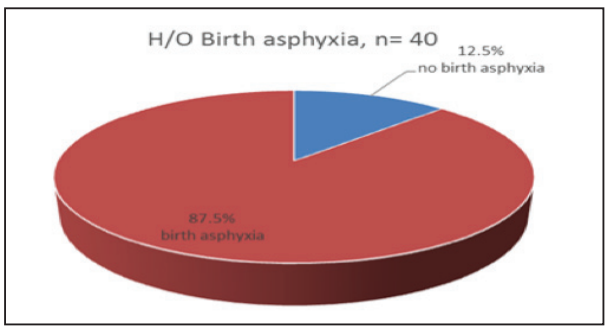

Figure 3: History of birth asphyxia during delivery

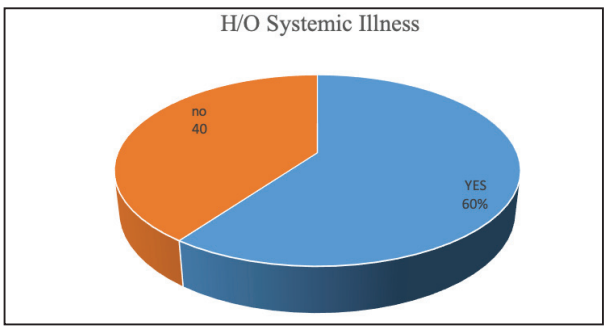

Figure 4: History of associated systemic illness in patients with cortical visual impairment

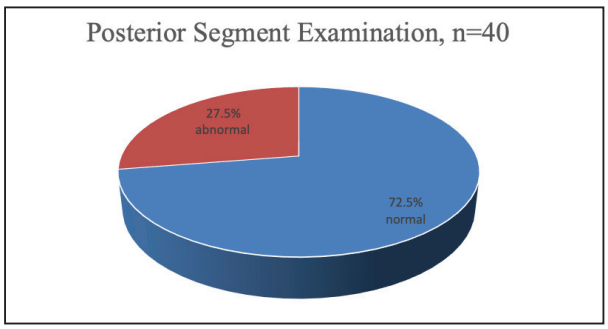

Figure 5: Posterior segment examination

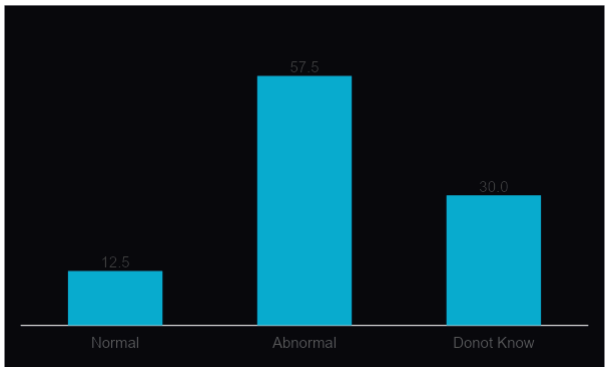

Figure 6: CT/MRI finding of patients with cortical visual impairment

Discussion

In our study, the male: female ratio was 60:40 (3:2), ANC history was uneventful in 77.5%, (p value 0.001), birth asphyxia was present in 87.5% (p value 0.001), PNH history was eventful in 67.5% (p value 0.027). In eventful PNH, 37% had seizure disorder, 11.1% had febrile convulsion, 11.1% had cerebral palsy, 7.4% had epilepsy, 3.7% had encephalocele, 3.7% had encephalopathy, 3.7% had hydrocephalus, 3.7% had meningitis, 3.7% had hydrocephalus with meningitis, 3.7% had microcephaly secondary craniosynostosis, 3.7% had neonatal jaundice, 3.7% had neonatal sepsis and 3.7% had obstructive hydrocephalus with craniopharyngioma.

Systemic illnesses were present in 60% of the patients. Among them 37.5% had seizure disorders, 21.5% had cerebral palsy, 8.3% had GDD with seizure disorders, 8.3% had hydrocephalus, 4.2% had meningitis with hydrocephalus, 4.2% had CHD, 4.2% had encephalocele, 4.2% had GDD with encephalopathy, 4.2% had GDD with microcephaly with secondary craniocynostosis and 4.2% had meningitis with hydrocephalus.

Anterior segment examination was normal in 92.5%. 57.5% had abnormal CT scan findings, among them HIE sequelae were common. Posterior segment examination was normal in 72.5% and abnormal in 27.5%. Among them, temporal pallor was seen in 81.8%, pale disc in 9.1% and pale disc with myopic fundus in 9.1%.

Children with CVI exhibit distinct visual behaviors which are often misinterpreted. The incidence of CVI has been growing causing greater need for identification and intervention from teaching and therapy service providers. Recognizing distinctively the children with CVI from children with other types of visual impairments in intervention designs and other educational planning is a paramount for constructing effective programs (Swift SH et al, 2008).

CVI is a condition where bilateral visual loss occurs by injury at visual areas of the brain without significant impairment on eye or anterior visual pathway. The most common causes of CVI are perinatal hypoxic ischemic encephalopathy (HIE) and postnatal anoxia giving rise to more extensive gray and white matter injury affecting optic radiations and visual cortex. Children with CVI often have other significant neurological disabilities and seizures as well.

In the evaluation of children with CVI conducted between January 1996 and March 2003 as for the study - Rehabilitation of cortical visual impairment in children, International Journal of Neuroscience, the results of an intensive visual stimulation program were retrospectively

examined. Standards were formulated in the manner that ensured the selection of a fairly homogeneous group of 21 children with CVI caused by perinatal HIE or postnatal anoxia having extensive gray and white matter injury along with multiple neurological deficits. 20 of 21 (95%) had symptomatic epilepsy as well. The study with responses ranging from just a pupillary light reflex to rudimentary perception of outline was carried out and an at-home treatment program was conducted for each subject. The remarkable improvement seen in 20 of 21 children (95%) after 4 to 13 months indicated the probability of considerable neuroplasticity in visual systems leading to reintegration and visual recovery even in the challenging group (Malkowicz DE et al, 2009).

The proportion of visual impairment in children with multiple disabilities is high; these children can have a combination of motor, cognitive, and visual dysfunction. 10.5% of children with developmental disabilities have been found to have suffered from visual impairment. In the United States, CVI is found to be one of the most important causes of bilateral blindness. It is also becoming an emerging significant cause of blindness in the developing world too, along with the increase in survival rate of children suffering from perinatal hypoxia. These developmentally handicapped and cortical visually impaired children may have a wide range of visual deficits. In children with CVI, number of perceptive visual functions such as face recognition, object recognition, motion processing, visual memory, orientation, visual spatial perception, and simultaneous perception may be affected. Studies, however, have not observed all of perceptive visual functions systematically. Vernier acuity has been found to be more affected than grating acuity. Some evidence indicates that dorsal stream / magnocellular pathway deficits may be more common in children with CVI. Empirical studies have suggested that the finding of periventricular leukomalacia (PVL) on magnetic resonance imaging (MRI) of the brain, as an important anatomic finding in children with CVI.

Recognizing CVI is the first and foremost step towards rehabilitation and prevention. These children need to be examined with comprehensive and broad examination techniques to assess vision and visual dysfunction. Conventional vernier acuity measurements on one hand are often not possible and on the other, it gives an erroneous picture in the CVI patient. It is important to examine via record grating acuity, which helps in follow-up. Often qualitative visual acuity testing methods supplement the quantitative visual acuity recording methods. Tests to detect the various types of perceptive dysfunctions need to be developed and tailored, taught, and made available to the ophthalmologists in the developing world.

In CVI patients, the prognosis for improvement of visual impairment is controversial. Earlier studies suggest that prognosis is poor, however, longitudinal studies show the contrary results suggesting improvement over time. Prevention involves early identification of the risk factors. Rehabilitation methods are to be tailored to each child. Each child with CVI possibly has a unique visual and motor deficit that demands an individualized approach (Swaminathan M, 2011).

A study on children with CVI and the literature review cover the diagnosis, etiology, prevalence, prognosis, and a comparison of the differences between children with CVI and those with ocular impairment. The case study presents occupational therapy intervention strategies specific to CVI and result of treatment (Baker-Nobles et al, 1995).

In the study, chronic cortical visual impairment in children: etiology, prognosis, and associated neurological deficits, 7200 outpatients were

examined over the past 15 years and were reviewed compiling data. In addition, the authors devised and applied a system for grading visual recovery in order to assess prognosis. CVI occurred in 2.4% of all patients examined. The four most common causes of CVI were perinatal hypoxia (22%), cerebral vascular accident (14%), meningitis (12%), and acquired hypoxia (10%). Most children with CVI had associated neurological abnormalities. The most common were seizures (53%), cerebral palsy (26%), hemiparesis (12%) and hypotonia (5%). Associated ophthalmological problems were esotropia (19%), exotropia (18%), optic nerve atrophy (16%), ocular motor apraxia (15%), nystagmus (11%) and retinal disease (3%). On average, CVI patients improved by two levels as measured by the authors' scale and they concluded that the majority of children with CVI showed at least some recovery. In this group of children, CVI is often accompanied by additional ophthalmological problems and is nearly always associated with other serious neurological abnormalities (Huo R et al, 2009).

In this study 87.5% had birth asphyxia, 37% had seizure disorders, 11.1% had cerebral palsy, 11.1% had febrile convulsion, 7.4% had epilepsy, 3.7% had encephalocele, encephalopathy, hydrocephalus, meningitis, meningitis with hydrocephalus, microcephaly with secondary craniocynostosis, neonatal sepsis, obstructive hydrocephalus with craniopharyngioma and neonatal jaundice.

In the study done by Giovanni C et al (2000), 10 patients with infantile spasms, hypsarrhythmic electroencephalograms and developmental delay (i.e. West syndrome) presented with severe visual inattention despite a normal ocular examination in follow-up (14 months to 6 years) and 5 patients (50%) had no improvement in their visual behavior. Despite some degree of improvement observed in the others, their visual function remained abnormal. All patients had moderate or severe mental retardation. CVI with infantile spasms is an important association, which the pediatric ophthalmologists should recognize (Castano G et al, 2000).

Conclusion

Birth asphyxia, perinatal and postnatal hypoxia are the common causes of birth asphyxia. We can mitigate cortical visual impairment by limiting birth asphyxia, perinatal and postnatal hypoxia and working together with a gynecologist and obstetrician to address the same.

Limitations of the study: Small sample size, retrospective study.

References

Baker-Nobles L, Rutherford A (1995). Understanding cortical visual impairment in children: American journal of occupational therapy; 49(9):899-903.

Castano G, Lyons CJ, Jan JE, Connolly M (2000). CVI in children with infantile spasm: journal AAPOS; 4(3):175-8.

Huo R, Burden SK, Hoyt CS, Good WV (1999). Chronic cortical visual impairment in children: aetiology, prognosis, and associated neurological deficits. Br J Ophthalmol;83(6):670-5. doi:10.1136/bjo.83.6.670

Malkowicz DE, Myers G, Leisman G (2006). Rehabilitation of cortical visual impairment in children: International Journal of Neuroscience; 116(9):1015-33.

Swaminathan M (2011). Cortical visual impairment in children: A new challenge for the future. Oman J Ophthalmol;4(1):1-2.

Swift SH, Davidson RC, Weems LJ (2008). Cortical visual impairment in children: Presentation Intervention, and Prognosis in Educational Settings. Teaching exceptional children plus;4(5):1-14.