REVIEW ARTICLE

Brain visual impairment in childhood: mini review

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Abstract

Cerebral visual impairment (CVI) is one of the leading causes of severe visual impairment in childhood. This article was written to highlight any new knowledge related to cerebral visual impairment in childhood.

The international literature was searched to describe the type of visual, oculomotor and / or visuo-perceptual disturbances and to discuss the prognosis. CVI children show a wide range of visual disturbances. These could be either visual, oculomotor, perceptual or a combination of all. The severity of CVI depends on the time, location and extend of the brain damage.

The visual function seems to improve in CVI children, especially in the cortically damaged, mainly due to brain plasticity. The increased survival rate of very premature infants during the last decades has increased the incidence of CVI in childhood. Better understanding of the pathophysiological mechanisms of CVI, early diagnosis and early intervention could lead to a better quality of life of these children. Hippokratia 2010; 14 (4): 249-251

Key words: cerebral visual impairment, hypoxic ischemic injury, periventricular leukomalacia, review

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CVI is one of the leading causes of severe visual impairment in childhood – almost 2 per 1000 live births and 19 per 1000 live births for infants born at 20-27 weeks gestation[1,2]. Any visual dysfunction due to brain damage in a patient with an otherwise normal ocular examination is characterized as CVI[3]. In CVI patients the brain damage could be either subcortical, cortical or both. Subcortical damages are more common in preterms[3,4].

The causative factor of CVI in children could appear either pre-, peri- or postnatal. Although a perinatal hypoxic ischemic injury seems to be the most common cause of CVI in children, meningitis/encephalitis, hydrocephalus, congenital central nervous system malformation, trauma, brain tumours, cerebrovascular accidents, metabolic disorders, are some other possible causes[5].

Asphyxia leads to hypoxia and hypercarbia, loss of autoregulation of cerebral blood flow and decreased perfusion in watershed areas, between the anterior and middle cerebral artery, and between the middle and posterior cerebral artery. Decreased perfusion in these areas, in full term infants, leads to cystic infarctions in the frontal and parieto-occipital regions or to damage of the thalami and basal ganglia[5]. In preterm infants, hypoxia selectively injures mainly the periventricular deep white matter. In this area resides the germinal matrix, a temporary structure with a rich supply of fragile capillaries[6]. This structure produces neurons and glial cells. The fragile capillaries are prone to hemorrhage from hypoxia and ischemia, due to fluctuations in blood flow and blood pressure perinatally. The germinal hemorrhage could lead to damage of the white matter around the ventricles (periventricular leukomalacia - PVL), to intraventricular hemorrhage, to porencephalic cysts and to cortical neuronal disorganization, leading to visual, motor, cognitive and attentional problems[7,8]. Since the optic radiations and the corticospinal motor tracts travel within the periventricular white matter, PVL is strongly associated with CVI[9].

Visual disturbances

The type and the severity of visual disturbances vary a lot in CVI children. The sensory visual function, the oculomotor, the visuomotor and cognitive visual function could be either mildly or severely affected, leading to misinterpretation of the visual world.

A routine eye examination is not enough to identify the depth of the visual dysfunction in CVI children. Efforts to structure devices to evaluate the visual function globally in CVI children have been done. A system for quantifying functional vision into levels, was devised by Hoyt[6]. Level 1, perception of light; level 2, occasional fixation on large objects, faces, or movements in the environment; level 3, variable visual function but some moments of good visual fixation as indicated by the ability to see small objects or reliably fixate on the face; level 4, reliable fixation on a small target or with visual acuity measured in the range of 20/200 to 20/400 level 5, with one or both eyes open, vision between 20/50 and 20/200; level 6, normal sensory visual examination. By using this device in two groups of CVI children, with cortical and PVL accordingly, he found that in both groups the initial visual acuity was very poor (level 1 and level 2 ranging 22–50%), children with cortical damage showed visual improvement more often (78%), and that the incidence of strabismus, nystagmus and optic atrophy

was significantly higher in PVL CVI children. Kozeis et al[10], studying school aged cerebral palsied children found that the microsaccades, being used for reading, were also markedly affected in PVL children. Children with CVI could be either photophobic or photosensitive; they prefer coloured and highly contrasted figures.

Visual field defects could be or not present in CVI children, depending on the brain damaged area. In PVL patients have been found bilateral inferior visual field defects, possibly due to damage of the optic radiations (temporal lobe)[11].

Damage of visual associated cortical areas of the brain could lead to **cognitive visual dysfunction**. Visual information from the primary visual cortex is processed to these areas, following mainly two streams, the dorsal and the ventral stream. The dorsal stream connects the occipital area with the parietal lobe, responsible: to select accurately certain objects in a complex visual scene, to pay attention to those objects and to execute accurate gross and fine movements with his arms and legs in the space, visually guided. The ventral stream connects the occipital with the temporal lobe, where recognition of form, orientation in the space and visual memory is achieved[12]. Dutton et al[13] found that the most of the cerebral palsied children had visual acuity better than 20/30, but more than half of them had bilateral inferior visual field defects. All of them manifested inaccurate climbing, inaccurate reaching, knocking over objects, impaired simultaneous perception, inability to locate an object in a crowded visual field and difficulty seeing moving targets (dorsal stream dysfunction). 50% of patients manifested difficulties with orientation in unfamiliar places, recognizing faces, shapes or objects (ventral stream dysfunction). Although children with CVI usually function better in familiar environments, their visual attention is variable. Environmental distraction, fatigue, illness are some of the causes that could affect their attentiveness. Kozeis et al[14], studying the visual perceptual abilities related to reading process in CP school age children, found them markedly affected.

Other disorders

Additional ophthalmic deficits, such as: strabismus, nystagmus, optic nerve pathology, neurologic deficits like seizures, cerebral palsy, microcephaly or a combination of both, are very common[6,15].

Children with cortical damage commonly suffer horizontal tonic gaze deviation, while tonic downgaze is more common in children with subcortical damage. In homonymous hemianopia children appear with a conjugate gaze deviation towards the hemianopic field. Strabismus is very common (approximately 80%) in CVI children; exotropia is more common in cortical damage and esotropia is more common in subcortical damage[6]. Nystagmus is also common (approximately 40%) either in cortical or subcortical lesions[15,16].

Sometimes, CVI children appear with pale optic disc. When the insult affects the immature visual system a pseudoglaucomatous cupping of the optic nerve appears. When an insult affects the mature visual system optic atrophy appears[17-20].

Prognosis

Due to improved neonatal care and survival of very low birth weight preterm infants, the incidence of CVI patients has been increased. CVI is also one of the leading causes of severe visual impairment in childhood, though visual function of the CVI children usually improves, more commonly in cortically damaged children. It is believed that the visual improvement is mainly due to brain plasticity, followed by improvement of the cognitive status[21]. Baring in mind all the above, it is obvious that better understanding of the pathophysiological mechanisms of CVI in children, early diagnosis and early intervention will improve the rehabilitation status and the quality of life of these children.

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