VISUAL IMPAIRMENTS IN CHILDREN WITH CEREBRAL PALSY

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 Primljeno: 15.9.2011.
 Pregledni rad

 Prihvaćeno: 16.1.2012.
 UDK: 376.1-056.262

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Abstract: Cerebral palsy (CP) is the neurological developmental disorder mainly affecting motor abilities. Considering the high rate of associated impairments even the definition of CP is revised and changed.

Visual impairment is one of the most common associated impairment. Unfortunately, it is often unrecognized and considered to be a normal consequence of motor problems.

Sense of sight is most important for early child development, motivation, learning through imitation. It is, therefore, indispensable to know about visual problems of children with CP and possibilities of facilitating visual functioning in different situations. Early comprehensive, multidisciplinary assessment must be done whenever the child is at risk for developing neurological dysfunction.

This review gives informations about incidence and kind of visual problems in children with cerebral palsy.

Key words: cerebral palsy, visual impairment, cerebral visual impairment

INTRODUCTION

Cerebral palsy (CP) is the most common developmental disorder associated with lifelong motor impairment and disability. It is thought to affect two to four individuals per 1000 of the general population (Aisen et al., 2011; Jan, 2006). Originally reported by Little in 1861, CP has been the subject of books and papers by some of the most eminent medical minds of the past hundred years.

It has always been a challenge to define CP due to heterogeneity of disorders. Mac Keith et al. (1959) defined CP as "a persisting but not unchanging disorder of movement and posture, appearing in the early years of life and due to a non-progressive disorder of the brain, the result of interference during its development." Bax (1964) reported and annotated a definition of CP, which has been suggested by an international working group: "CP is a disorder of movement and posture due to a defect or lesion of the immature brain."

For practical purposes, it is usual to exclude from cerebral palsy those disorders of posture and movement which are (1) of short duration, (2) due to progressive disease, or (3) due solely to mental deficiency (Bax, 1964).

Those earlier definitions of CP were focused on motor aspects and specific brain damage. CP is, however, associated with other impairments. Therefore, on An International Workshop on Definition and Classification of Cerebral Palsy held in Bethesda, Maryland, July 11-13 2004, experts revised and in 2006 accepted the definition of CP as "a group of permanent disorders of the development of movement and posture, causing activity limitation, that are attributed to nonprogressive disturbances that occurred in the developing fetal or infant brain. The motor disorders of cerebral palsy are often accompanied by disturbances of sensation, perception, cognition, communication, and behaviour, by epilepsy, and by secondary musculoskeletal problems" (Rosenbaum et al. 2006).

Among the various associated handicaps in CP the most common is mental retardation, followed by speech impairment, seizure disorder, visual impairment, and hearing impairment (Majumdar et al., 2006).

The goal of this review is to present various studies investigating visual impairment in children with CP in order to report the incidence and kind of visual problems in children with CP.

OCULAR DISORDERS IN CHILDREN WITH CEREBRAL PALSY

The association of ocular problems with CP is known since Little's early papers. Over the last half of the century researchers from around the world perceived that visual impairment is common in children with CP (Altman et al., 1966; Arnoldi et al., 2006; Black, 1980; Breakey, 1955; Elmenshawy et al., 2010; Guzzeta et al., 2001; Hou et al., 2010; Schenk-Rootlieb et al., 1992; Venkateswaran & Shevell, 2008). The rates vary considerably from the study to study, depending on the selection criteria for the cohort the sophistication of the clinical tests used, and the terms used to describe both the CP and the visual deficits (Ghasia et al, 2008). Review of visual problems rates in children with CP from different studies is shown in Table 1.

Although rates of overall visual impairment vary from the study to study, all authors agree that ocular motility and refractive error are the most common visual problems in children with CP (Table 2) (Altman et al., 1966; Arnoldi et al., 2006; Black, 1980; Breakey, 1955; Elmenshawy et al. 2010). Among ocular motility problems, children mostly develop squint (esotropia more

common than exotropia) and nystagmus (Table 2) (Altman et al., 2006; Black, 1980; Breakey, 1955; Elmenshawy et al. 2010). The incidence of strabismus in general population reported in literature is from 1,28% to 6% (Matsuo & Matsuo, 2005; Stidwill, 1997; Yekta et al. 2010). As in children with CP, in general population more children develop esotropia (3.0%) than exotropia (1.2%) (Chew et al., 1994). Prevalence of nystagmus in general population of children is between 0,23 and 1% (Sarvananthan et al., 2009). In children with CP reported prevalences are different, but much higher than in general population (Altman et al., 1966; Black, 1980; Elmenshawy et al., 2010).

Children can develop amblyopia which, in many cases is not treated, considering it is a common problem in children with CP and cannot be treated (Graham, 1969; Erkkilä et al. 1996). Nevertheless, amblyopia therapy is necessary in any case of developing amblyopia in an attempt to preserve binocular vision as a prerequisite for normal visual field and stereopsis.

Despite many data about problems in ocular motility, there are only few reports about problems in smooth visual pursuit and generation of saccades, which are also common in children with CP (da Cunha Matta et al., 2008; Jackson et al., 2006; Mayberry & Gilligan, 1985). Those visual functions are important for fluent reading, and they have to be considered especially in case of school children with CP.

Incidence of refractive errors in general population is between 4.55% to 50%, and more authors report higher rates of hyperopia than myopia (Fotouhi et al., 2011, Logan & Gilmartin, 2004; Schimiti et al., 2001). Refraction errors are frequent

Table 1. Overall visual problem in children with cerebral palsy in percentages

Research	No. of participants with CP	Visual impairment (%)		
Breakey (1955)	100	56		
Altman et al. (1966)	64	92		
Black (1980)	117	77,8		
Schenk-Rootlieb et al. (1992)	164	71		
Arnoldi et al. (2006)	131	46		
Katoch et al. (2007)	200	68		
Venkateswaran & Shevell (2008)	92	80		
Hou et al. (2010)	354	38,98		
Elmenshawy et al. (2010)	46	76		

in children with CP (Table 2). In our practice, we noticed that more than half of children with CP and refractive errors requiring correction do not have prescription for glasses. There is a common opinion that children with CP "have bigger problems than refractive errors". Sometimes it is difficult to mount frames for the spectacles with no head control and parents and teachers just give up. However, knowing that vision is positively influencing the head control enabling, on the other hand, gathering of visual information, the best possible correction of refractive error should be provided in order to ease learning, facilitate communication and motor abilities.

Reduced accommodation is also common disorder in children with CP, although it often remains undetected (Duckman, 1979; Leat, 1996; Kozies et al., 2007). More than half of children with CP fail to accommodate normally, requiring plus correction for reading (McClelland, 2006; Kozies et al., 2007). Children with more severe motor impairments are at a greater risk of having accommodative deficits (Leat, 1996; McClelland, 2006). CP is a disorder of motor function, and, in cases with more severe motor impairment, the likelihood that the ocular muscles are involved is increased, and hence accommodative function is reduced. However, McClelland (2006) explains that the accommodation in the normal visual system is a complex response to a combination of visual, mechanical, and psychological stimuli, and implying a solely motor origin for the accommodative impairment demonstrated in CP is likely to be an oversimplification. Children with accommodative dysfunction are at a disadvantage to their visually normal peers in the school environment (Maples, 2003). Many visual tasks such as writing school work and looking at the blackboard, and other, may be affected by poor accommodation. Reduced accommodation conditions can often be successfully

treated with a vision therapy program and/or the use of plus lenses or bifocals (Rouse, 1987; Al-Bagdady, 2009). In school practice, enlargement of a print can also contribute to child's convenience reading.

CEREBRAL VISUAL IMPAIRMENT

Investigating visual problems in children with cerebral palsy ophthalmologists noticed visual problems that cannot be attributed to ocular problems. Some children with CP had no refractive errors or ocular motility problems but had poor perceptual performances, such as decreased visual acuity, intermittent fixation and lack of visual attention (Menken et al., 1987; Porro et al., 1998; Schenk-Rootlieb et al., 1992; Stiers et al., 2002). It is described as cerebral visual impairment (CVI).

Although the term CVI is often mentioned, the precise definition is lacking. Commonly, it is defined as a "disturbance of vision caused by a defective function of the retrochiasmatic part of the visual system in the absence of any important ocular disease" (Van Nieuwenhuizen, 1987).

CVI can manifest itself through a variety of clinical signs and symptoms, which seem to reflect the possible involvement of various parts of the retrochiasmatic visual pathways (Signorini et al., 2005). Acuity is commonly reduced in CVI but can be normal. Colour vision and contrast sensitivity are remarkably normal in many cases (in those in whom it can be tested) but are probably reduced in those with more profound damage in whom testing is impossible. The visual fields can be affected in a number of different ways. In children with CVI ability to see multiple targets at the same time can be impaired. That results in impaired attention. In addition, a disability to estimate the depth, particularly for walking down the stairs, can lead to injury. Disorders of eye move-

Table 2. Kinds of ocular disorders in children with cerebral palsy in percentages

		Research							
Visual problem (%)		Altman et al. (1966)		Black (1980)		Katoch et al. (2007)		Elmenshawy et al. (2010)	
Refraction	Myopia	90	15,5	49,6	-	33,5	13,5	67,2	41,4
	Hyperopia		33		-		20		20
	Astigmatism		27,5		-		-		10
Ocular	Esotropia	66	30	50,5	-	39	-	32,8	-
motility	Exotropia		14		-		-		-
	Nystagmus		5		16		5,5		11,14

ment control are common in children with CVI, who can exhibit one or more of the following eye motility problems: strabismus, nystagmus, unstable fixation, inaccurate fast eye movements (dysmetric saccades), deficient smooth pursuit movements and paroxysmal deviations, in which the eyes intermittently deviate upwards. Inability to recognize people, even close family members when they are seen out of context, can also occur. Brain damage causing cerebral visual impairment may lead to incomplete accommodation, persistent hypermetropia and consequent blurring of the retinal image (Dutton & Jackobson, 2001).

According to different authors (Table 3), incidence of CVI in children with CP is between 16% to 80% (Elmenshawy et al., 2010; Ghasia et al., 2008; Schenk-Rootlieb., 1992).

Table 3. Prevalence of cerebral visual impairment in children with cerebral palsy

Research	No. of participants with CP	Cerebral visual impairment (%)
Schenk-Rootlieb et al. (1992)	74	84
Katoch et al. (2007)	200	28
Ghasia et al. (2008)	50	16
Elmenshawy et al. (2010)	46	51,4

In children with cerebral palsy who can communicate and function socially, the above described visual problems can occur in any combination. In more profoundly affected children, it is very likely that they have cognitive visual problems in an equal measure, but they cannot be identified because they are masked by communication and motor problems (Dutton & Jackobson, 2001). In children with CVI ability to see can vary from one time to another, depending on child's overall physical comfortableness and on environmental and situational factors.

INCIDENCE OF VISUAL IMPAIRMENT ACROSS DIFFERENT TYPES AND SEVERITIES OF CEREBRAL PALSY

The occurrence and severity of visual problems in children with CP vary across different types of CP.

Until recently severity of CP was described as being 'mild', 'moderate', or 'severe. Nowadays,

severity is described through five levels according to The Gross Motor Function Classification System (GMFCS). GMFCS is a standardized method to classify gross motor function in children with cerebral palsy (CP) aged from 1 to 12 years (Palisano et al., 1997; Rosenbaum et al., 2008; Russell et al., 1989). This 5-level system was designed to reflect differences in gross motor function that are meaningful in the daily lives of children with CP and their families, with an emphasis on sitting and walking (Palisano et al., 1997; Rosenbaum et al., 2008; Russell et al., 1989). A classification is made by determining which level best corresponds to a child's present motor abilities. Generally older children on level I walk without limitations, on level II walk with limitations, on level III walks using a hand-held mobility device. Level IV means selfmobility with limitations; they may use powered mobility and children at level V are transported in a manual wheelchair (Palisano et al., 1997).

Authors have found that children on higher level of GMFCS have more associated impairments (da Costa et al., 2004; Ghasia et al., 2008). It is not surprising as it is known that CP levels with more severe deficits are mainly caused by higher degree of brain involvement (Mejaški – Bošnjak et al., 2005; Mejaški – Bošnjak, 2007; Rešić et al., 2008). Therefore, visual pathways are more likely to be injured with the consequence of visual impairment.

Different studies also report various rates of visual impairments in different types of CP. The children with diplegic and spastic CP seem to be more often hyperopic and esotropic, but retain the highest prevalence of fusion and stereopsis. In contrast, children with quadriplegic and mixed (dyskinetic, athetoid, hypotonic, and ataxic) CP tended to have high myopia, CVI, dyskinetic strabismus, and severe gaze dysfunction (Black, 1982; Buckley & Seaber, 1981; Erkkilä et al., 1996; Fantl & Perlstein, 1961; Ghasia et al., 2008; Jacobson et al., 2002; Kozeis et al., 2007; Pennefather & Tin, 2000).

(RE)HABILITATIONAL AND EDUCATIONAL CONSIDERATION

Elmenshawy et al. (2010) states that visual handicap plays a significant role in the overall disability of CP children. Usually children with CP are

not examined with care due to difficulties in assessment due to their mental and physical disability or with the idea that nothing much can be done to help them cope with their already poor condition. In Black's (1980) research, we can notice that many of the children with CP do not even have the best possible correction for refractive errors. Flanagan et al. (2003) also stated that children with CVI are mostly not known to services which would promote maximal visual and developmental potential at an early stage.

The recognition, assessment and (re)habilitation of children with cerebral dysfunctions is the most important in early age, during the critical period of visual function's development. Many researchers confirm significant improvement of visual functions when stimulations started in time (Alimović & Mejaški - Bošnjak, 2011; Fazzi et al., 2005). Stimulative environment and performing visual stimulations in early age can encourage development of visual functions and facilitate the use of vision in everyday situations. That would all lead to higher motivation and better performance of child in all activities.

The sense of sight has remarkable influence on overall child development (Alimović & Mejaški – Bošnjak, 2011). Visual objects are stimulating child to move towards them. Children learn through visual imitation. Therefore, it is necessary to have a good visual perception to facilitate overall child's development, especially in children with CP.

Visual impairment in older children with CP has an impact on quality of life. Reading abilities

are connected with visual functions such as saccades, smooth visual pursuit, ability to see two or more objects at the same time, visual acuity, etc. According to previously mentioned studies, problems in those visual functions are common in children with CP, so reading problems can occur in children with CP due to visual impairment. It is common that reading problems are misinterpreted as a consequence of mental retardation and are not treated properly. Those problems can often be easily avoided through text adaptations and optical and non-optical devices.

Children with poor contrast sensitivity or CVI can have problems in face recognition. This can cause problems in communication and socio-emotional development. By making some adaptations in the environment and increasing contrast sensitivity of familiar faces, those problems can easily be solved and enable normal socio-emotional relations.

CONCLUSION

Visual impairment is one of the most common associated problems in CP. Due to importance of visual perception for development and functioning, it is mandatory to do early visual assessments in any child under suspicion of neurological dysfunctions. Assessment of vision must encounter the assessment of visual functions and functional vision. It has to be done multidisciplinary where ophthalmologist, neuroophtalmologist, vision therapist, special education and rehabilitation teacher and occupational therapist are included.

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OŠTEĆENJA VIDA KOD DJECE S CEREBRALNOM PARALIZOM

Sažetak: Cerebralna paraliza (CP) je neurološki razvojni poremećaj koji uglavnom zahvaća motoričke sposobnosti. Obzirom da se uz dijagnozu cerebralne paralize veže velika stopa udruženih poremećaja, čak je i definicija revidirana i promijenjena.

Oštećenje vida je jedno od najčešćih udruženih poremećaja. Nažalost ono je često neprepoznato i smatra se normalnom posljedicom motoričkih problema.

Osjet vida je najvažniji za rani djetetov razvoj, motivaciju, učenje imitacijom. Stoga je neophodno poznavati vizualne probleme djece s CP i mogućnosti pospješivanja vizualnog funkcioniranja u različitim situacijama. Rana sveobuhvatna, multidicsiplinarna procjena je obavezna za svako dijete za koje postoji rizik razvoja neurološke disfunkcije.

Ovaj pregledni članak daje informacije o prevalencijama i vrstama oštećenja vida u djece s cerebralnom paralizom.

Ključne riječi: cerebralna paraliza, oštećenja vida, cerebralno oštećenje vida