The Complicated Diagnosis of Cortical Vision Impairment in Children with Multiple Disabilities

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ABSTRACT

Cortical vision impairment is caused by a variety of neurological insults affecting the posterior visual pathways and/or visual cortex. Children with cortical vision impairment often have additional disabilities and ophthalmic findings, which can complicate the diagnosis of CVI. Children with multiple disabilities and a suspected vision impairment require a thorough investigation to determine the source of their reduced visual responses. Orthoptists may be involved in this process. The importance of distinguishing between reduced visual responses due to cortical vision impairment as opposed to attention difficulties or cognitive delay is discussed in this paper.

KEY WORDS: attention, arousal, cognitive delay, intellectual disability, intervention

INTRODUCTION

Cortical vision impairment (CVI) is a complex condition in which the vision impairment, rather than being of an ocular cause, is primarily due to damage to cortical areas that process vision. Many children with CVI also have additional impairments such as an intellectual disability or cerebral palsy[1]. However there is little emphasis in the literature on the diagnosis of cortical vision impairment in children who have multiple disabilities. Given the increasing incidence of CVI in children over time[2,3], it appears that there is a need for specific information about the diagnosis and prognoses of CVI in children with multiple disabilities.

The neuroanatomical structures related to the processing of vision are implicated in CVI. Significant processing of visual input occurs within the retina and brain before images reach the primary visual cortex. Initially, information from the retina is relayed to the lateral geniculate nucleas (LGN) of the thalamus via the optic nerves and optic tracts. This pathway is referred to as the anterior visual pathway. In addition to the anterior visual pathway it is hypothesized that there are two visual pathways present in the human brain; the geniculostriate pathway and the extragenciculate pathway. These have implications for the diagnosis of CVI. It is well established that the geniculostriate pathway is responsible for the conscious visual analysis of the environment[4]. Involved in this pathway are the LGN, optic radiations, primary visual cortex and visual association cortex (within the occipito-parietal regions)[5]. There is increasing evidence for an extrageniculate visual pathway, which is thought to be responsible for visual attending to a stimulus away from the point of fixation[6]. The extrageniculate pathway extends from the superior colliculus of the midbrain through the pulvinar nucleas of the thalamus, to the visual association cortex[2]. Damage to varying areas of this pathway appears to produce differing types of deficits in being able to shift attention covertly to a stimulus[6]. The extrageniculate pathway may also be responsible for pupillary and blink responses to light, optokinetic nystagmus, pattern discrimination and colour sense[7].

CVI occurs when there is damage to the geniculostriate pathway, extrageniculate pathway or visual cortex[2] and should be suspected in any child with decreased vision that cannot be explained by ocular findings[8]. Different types of neuroimaging techniques have been used with various levels of success to identify damage to the visual pathways and visual cortex in children suspected with having CVI[9]. Identification of regions of dysfunction within the visual pathways assists in confirming a diagnosis of CVI.

In pure CVI, pupillary functions are normal and there is an absence of nystagmus as these are controlled by the anterior portion of the visual pathway[2]. Any insult to the brain may cause CVI, with the main causes being hypoxia-ischemia, head injuries (over half due to 'shaken baby'), shunt failure in hydrocephalus, developmental brain defects, infections of the central nervous system and infantile spasms[10,11]. CVI can be present in children as an isolated finding[12]. However, given that many of the causes of CVI result in diffuse brain damage or are associated with syndromes, it is not surprising that many children who have a vision impairment due to cortical damage, also have ocular findings, additional disabilities and medical conditions[11,13]. The presence of additional problems in children with CVI complicates the process of diagnosing this condition according to the traditional diagnostic criteria.

Implications of additional findings

It is evident that children who have CVI may have ocular findings including nystagmus and abnormal pupillary responses. These may be due to ocular conditions related to the cause of CVI, conditions unrelated to this cause or additional damage to the anterior visual pathways. As many as 65.3% of patients with CVI have been found to have at least one ophthalmological deficit[13]. Optic atrophy is one of the main ophthalmic findings in children with CVI. Groenvald, et al.,[10] found that the prevalence of optic atrophy in children with CVI was 26%. Optic atrophy most commonly occurs due to transynaptic degeneration of the optic nerve following damage to

the posterior visual pathways, but may occur at the time of insult as part of the widespread brain damage[14]. The presence of nystagmus in up to 11.2% of children with CVI[13] is further evidence of involvement of the anterior visual pathway in CVI. The nystagmus may occur subsequent to optic atrophy or be due to additional damage to the anterior pathways. This ocular finding is in conflict with the definition of 'pure CVI'. It is of interest though that nystagmus can be absent in the presence of ocular or anterior pathway defects when there is extensive damage to posterior visual pathways[2]. In addition to the above ocular findings, children with CVI have also been found to have strabismus, ocular motor apraxia, gaze palsy, significant refractive error and retinal conditions[13]. In light of the vast array of ophthalmic findings in children with CVI, it is apparent that a thorough ophthalmic investigation, functional vision assessment and sound clinical judgement is necessary to determine whether or not the level of vision impairment can be related to ocular causes, or is in fact attributed to cortical damage. The orthoptist may have a role in performing functional vision assessments and explaining their findings to parents, teachers and other allied health or medical professionals.

The high incidence of additional impairments and medical conditions in children who have CVI may further complicate the diagnosis of CVI. The most common of these are cerebral palsy, epilepsy, hydrocephalus, deafness and intellectual disability[1]. The characteristics of CVI as described by Crossman[4] include the following: poor visual communication skills, fluctuating vision and visual inattention or lack of curiosity. Additional behaviours include that spontaneous visual activity is of short duration, objects may be mouthed instead of explored visually and body position may influence use of vision. Good, et al.,[15] stated that the clinical examination is usually sufficient to establish the diagnosis of CVI. Hence the presence of characteristics, such as the above, in a child with evidence of brain damage but an absence of ocular findings may result in a diagnosis of CVI being made. This is supported by Utley, et al., (1998) who stated that CVI is frequently best diagnosed by considering characteristic behaviours. However, much care must be taken to establish that the visual behaviours are attributed to CVI rather than being directly attributed to a related condition. Difficulties in determining the primary cause of some visual behaviours have been identified[15]. An example of such difficulties includes children who have ocular motor disturbances, as in cerebral palsy. Their inability to shift their gaze towards objects may be mistaken for visual inattention, which is a characteristic behaviour of CVI. A further example of such difficulties is children who have subclinical seizures due to epilepsy. The disturbance of a seizure during certain activities may appear as the child having a short visual attention span[15].

Due to the high incidence of additional disabilities in children with CVI, a thorough medical investigation is necessary to determine whether or not a child has CVI or whether their behaviours are characteristic of their additional disabilities. There is also a need to actively deter professionals and teachers from labeling a child with the often confusing diagnosis of CVI on the basis of visual behaviours, until appropriate investigations have taken place. Orthoptists working with children who have CVI and multiple disabilities would benefit from knowledge of the implications that various impairments such as cerebral palsy and epilepsy have on the use of vision. This knowledge would assist them to make accurate judgements about the cause of a specific visual behaviour in a child with multiple disabilities.

Implications of attention problems and cognitive delay

In addition to difficulties with diagnosis arising from additional conditions, one concern raised in this article is that a child may meet the criteria for diagnosis of CVI and demonstrate the characteristic behaviours, but may experience difficulties in other areas of development at a similar level to their vision impairment. For example, a child may be equally as inattentive and lacking in curiosity to auditory stimuli as to visual stimuli. This raises the concern that CVI may not be an appropriate diagnosis for children who have general cognitive delay or general attention difficulties in several areas including vision. The definition of CVI by Whiting, et al.,[17], encompasses the issue addressed above. Their definition includes that "the diagnosis of CVI should be suspected when there is a greater delay in the visual development than in other areas" (p. 738). In addition Luna, et al.,[18] have commented that it is difficult to determine whether or not deficits in CVI are related to specific anatomical lesions or to more global deficits including attention problems. They stated that techniques for separating attention deficits from visual deficits would be advantageous.

As yet there does not appear to be a method of separating attention deficits from visual deficits in children with multiple disabilities. It is apparent that determining a child's comparative level of development across a variety of areas would be a detailed task and may require the involvement of several professionals in the area of childhood development.

It is likely that the cause of a child's visual behaviour will influence the likelihood of visual improvement over time. Differentiating between visual behaviours specifically due to CVI as opposed to visual behaviours due to cognitive delay or attention deficits raises the issue of whether there are differences in the expected visual prognoses for children in these two groups.

Prognostic outcomes

In general, little is known about specific prognostic outcomes in CVI[15]. Studies have shown that some degree of visual improvement in CVI is common but not to normal levels of vision, with many children remaining vision impaired[2]. Of interest are studies that investigate the visual outcome in children with CVI and multiple disabilities. On the whole it has been found that a poorer visual outcome is more likely in children with extensive neurological damage and a lower level of intellectual attainment in addition to

CVI[2,18,19,20]. More specifically a poorer visual outcome for children with CVI has been found in those with lower IQs[19] and those with neonatal seizures[20]. In children with hypoxic insults but no seizures, visual outcome has been related to general neurological outcome[20]. Additionally it has been well documented that treatment of underlying disorders, such as reduction in seizures in children with CVI and treatment of hydrocephalus is usually accompanied by an improvement in vision[2,21]. The above conclusions clearly identify a relationship between visual outcomes and overall developmental outcomes, with improvements in vision occurring as other areas of development also improve. This supports the author's premise that many children diagnosed with CVI and multiple disabilities may be experiencing visual difficulties which are more related to their general cognitive delay or attention problems rather than being specifically due to CVI. According to the additional definer of CVI put forward by Whiting et al.,[17] children in the above categories may not meet the proposed criteria for diagnosis of CVI.

The importance of an appropriate diagnoses

Children with CVI and multiple disabilities are often involved in programs where the aim is to maximise the use of functional vision[15] and to improve residual vision. Commonly suggested strategies for improving residual vision in children with CVI and for encouraging the use of vision in these children include providing a visually stimulating environment, using strong colours and patterns, using movement and performing tasks in a consistent and standardised fashion[4,15]. However, the notion presented in relevant professional literature, that the level of vision improvement in children with CVI and multiple disabilities is dependant on overall development, indicates that improvements in vision, for these children, may be closely related to attentional or cognitive development. Geniale[22] discussed the importance of arousal level for improvements in overall abilities for children with cerebral palsy. She stated that a prerequisite for optimising visual interest and visual motor control is to obtain an appropriate level of arousal and postural activity. It is apparent that a child's diagnoses may influence whether or not the intervention provided has a specific vision bases or is more related to general development.

Identifying the cause of visual difficulties in a child with multiple disabilities is an important consideration for the appropriate design and implementation of programs to encourage the use of vision. The diagnosis of CVI for some children with multiple disabilities may lead to confusion for parents and teachers, with the misleading belief that the vision impairment exists as a separate entity to the presence of diffuse neurological damage. As a result of this belief children with the diagnoses of CVI may receive a decreased amount of intervention in the area of general attention and arousal, in relation to specific vision intervention. Providing only minimal intervention in the area of attention and arousal would be unfortunate for children whose reduced visual responses are more related to their cognitive delay.

Conclusion

The diagnosis of cortical vision impairment in a child with multiple disabilities is complicated. One of the roles of professionals involved in making such a diagnosis is to carefully evaluate the child's visual behaviours to determine whether these can be attributed to a primary diagnosis of cortical vision impairment or are more related to their additional physical, cognitive or ophthalmic findings. The orthoptist may have the role of performing a functional vision assessment and determining whether the child's visual behaviours can be explained by their ophthalmic findings.

A primary diagnosis of cortical vision impairment as explanation for reduced visual responses should be made with caution, even in the presence of neuroimaging which indicates damage to the visual pathways. Children with extensive neurological damage causing severe intellectual disability may demonstrate reduced responses to stimuli from a variety of mediums. Reduced responses to visual stimuli as well as auditory and tactile stimuli may be indicative of reduced cognitive function rather than a specific vision impairment.

As yet there is no simple means of identifying whether or not poor visual response in a child with multiple disabilities is due to CVI or deficits of attention. Therefore the author recommends that a child with multiple disabilities and suspected cortical vision impairment should be carefully assessed by a multidsciplinary team who are experienced in this field. The overall aim of the team assessment would be to identify whether or not the child's primary impairment is their vision impairment or cognitive delay. The outcome of such assessments may have direct implications for the type of intervention that a child with multiple disabilities and a vision impairment receives.

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