

# Article • Cerebral Visual Impairment and the Association with Cerebral Palsy

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## ABSTRACT

**Background:** Approximately 60-70% of children with cerebral palsy also display cerebral visual impairment, both of which often share common origins that can involve the retrogeniculate visual pathways. The purpose of this case report is to explore the association of cerebral palsy and cerebral visual impairment, as well as the ocular manifestations typically present in children with cerebral palsy.

**Case Report:** A three-year-old full-term Caucasian male with spastic quadriplegia presented for a clinical functional low vision evaluation in order to determine how to optimize his visual learning environment. Despite normal MRI scans, normal EEG, and a healthy anterior visual pathway, he presented with severely reduced vision, slightly reduced contrast sensitivity, variable strabismus, oculomotor impairment, a bilateral congruous inferior field defect, and visual perceptual dysfunction. Completion of the Teach CVI/Ortibus CVI Survey Level 1 by the patient's parents and nurse revealed 4/6 screening items suspicious of CVI and 10/19 answers suspicious of CVI. The diagnosis of CVI was most consistent with the patient's level of visual function and inability to sustain the use of vision for a task, as well as the response to the CVI survey. The family was given extensive education on how to optimize the patient's visual function and was recommended

to undergo a learning media assessment from a teacher of the visually impaired.

**Conclusions:** The clinical diagnosis of cerebral palsy should include not only the characteristics of the motor aspects of the disorder but also the neuro-ophthalmological and perceptual visual aspects that have been shown to be present in a majority of cases. Early intervention and rehabilitation of visual impairments, including CVI, through a multidisciplinary approach can contribute to the improvement of the motor impairments in cerebral palsy and lead to a better overall prognosis.

**Keywords:** cerebral palsy, cerebral visual impairment, functional low vision, neuro-ophthalmology, visual perceptual dysfunction

## Introduction

Cerebral visual impairment (CVI) is a deficit of visual function caused by damage to the retrogeniculate visual pathways, which include the optic radiations, occipital cortex, and associative visual areas, in the absence of any major ocular disease.<sup>1</sup> The well-accepted two-stream model of higher visual processing systems, the dorsal and ventral streams, aids in describing the visual-cognitive disorders that are prevalent in CVI.<sup>2</sup> The dorsal stream is the occipito-parietal pathway that involves visual-spatial abilities, and the ventral stream is the occipito-temporal pathway that involves visual object recognition.<sup>3</sup>

Cerebral palsy (CP) is a group of disorders of development of movement and posture that are caused by early non-progressive lesions of the central nervous system.<sup>4,5</sup> The presentation of CP can include motor, sensation, cognitive, communicative, perceptual, and behavioral disorders, as well as seizures.<sup>4</sup> The most common form of CP is spastic diplegia that exhibits bilateral hypertonic involvement of the lower extremities.<sup>4</sup> CP and CVI share common origins that can involve the retrogeniculate visual pathways, in which periventricular leukomalacia is

often the causative factor.<sup>6</sup> Literature shows that 60-70% of children with CP have CVI.<sup>6,7</sup>

CVI is the leading cause of childhood visual impairment in developed countries, and its incidence continues to increase.<sup>8</sup> This reflects the improvements in diagnosing CVI and the advances in neonatal intensive care, leading to an increased survival rate of preterm children.<sup>1</sup> Although consensus on its definition is still highly debated, CVI has been defined as a verifiable visual dysfunction that cannot be attributed to disorders of the anterior visual pathway or to any potentially co-occurring ocular impairment.<sup>9</sup> A functional approach in assessing and diagnosing CVI has led to an improvement in identifying those with it, as well as to establishing individualized treatment plans. Early diagnosis, intervention, and rehabilitation have been shown to impact children with CVI positively in numerous studies.<sup>6,10</sup>

The etiology of CVI is most often multifactorial and can include hypoxic, ischemic, and inflammatory events that disrupt white-matter development.<sup>11</sup> The most frequent cause of CVI is perinatal hypoxic ischemic damage, with prematurity and low birth weight being prominent risk factors for CVI.<sup>3,11</sup> Approximately 60% of children with perinatal hypoxic ischemic damage have CVI.<sup>1</sup> Other causes of CVI include cerebral malformations, hydrocephalus, CNS infections, epilepsy, head trauma, metabolic and neurodegenerative diseases, and exposure to drugs.<sup>1</sup>

The purpose of this case report is to explore the association between CP and CVI, as well as the ocular manifestations typically present in children with CP. This report describes the case of a full-term, three-year-old patient with spastic quadriplegia and CVI despite normal MRI scans and EEG.

### Case Report

A three-year-old Caucasian male presented as a new patient accompanied by his parents and nurse for a clinical functional low vision evaluation. The primary goal for the patient was to establish how to optimize his visual learning environment. His parents were concerned about a possible inferior field defect. He had previously been seen by an audiologist, who suspected that there might have been a sensory processing disorder at play, although this is not typically diagnosed until children are older. The patient had an MRI once in infancy and again at two years of age, as well as an EEG, all of which appeared normal. He had an evaluation from a teacher of the visually impaired (TVI); however, his parents reported

low success with the current services. He was not prescribed any optical correction.

The patient was born full term with a normal birth weight of seven and a half pounds. His health history included pyloric stenosis, cerebral palsy with spastic quadriplegia (hypertonia of trunk and neck and hypotonia of limbs), and global developmental delay. He had a chromosomal micro array done, showing homozygosity of chromosome three. The patient underwent surgery for pyloroplasty at approximately three and a half weeks of age. It was noted that following the procedure, the patient had prolonged paralysis from succinylcholine, remained intubated, and had an apneic event requiring bagging with adjustment of the endotracheal tube. His lowest O2 saturation was 70%, leading to an unclear etiology of the patient's neurological problems.

The patient received early intervention services, including therapy from a speech and language therapist, a physical therapist, an occupational therapist, and a teacher of the visually impaired. He used a communication board, and his primary learning media were auditory. His medications included omeprazole, fluticasone, a probiotic, CBD/THC oils, gripe water, milk of magnesia, and albuterol and acetaminophen as needed. He also had a G-tube. His allergies included suxamethonium, latex, garlic, and adhesive remover.

The patient had been seen previously by two other eye care professionals. Below is a summary of the exam reports.

#### ***Ongoing eye care from 12 months to present age (02/17/16 to 9/27/17)***

- CVI secondary to lack of findings for level of vision function in the anterior visual pathway consistent with light perception
- External & internal ocular health normal OU
- Reduced nystagmus
- Aligned via Hirschberg
- (+) foveal reflex OU
- Pink and healthy optic nerves OU

#### ***Exam at 31 months (9/18/17)***

- (+) OKN
- Teller acuity cards attempted
- Eyes grossly aligned and positioned mostly in upgaze
- Full motilities
- External & internal ocular health normal OU
- Cycloplegic refraction: ~ +2.00 sph OU

The results of our exam findings on 02/20/2018 are depicted in Table 1.

Following our exam, the patient was diagnosed with CVI, hyperopia, intermittent alternating exotropia, bilateral congruous inferior visual field defects, developmental delay, and CP. The diagnosis of CVI was most consistent with his level of acuity and inability to sustain the use of vision for a task and was supported by the parents' responses on the CVI survey. Approximately thirty minutes of the exam was spent on extensive parent education, in which it was recommended to register the patient with the blindness agency in their state and the continued services of a TVI, who should administer a learning media assessment. The family was provided with multiple resources relating to CVI and children with vision impairment and was scheduled to return to the clinic for a functional low vision assessment in one to two years, and they were instructed to maintain eye care in the community. Due to the small amount of hyperopia found, glasses were not indicated at the correct time.

## Discussion

Our patient presented with CP with spastic quadriplegia and CVI. The literature reports that 60 to 70% of children with CP also manifest CVI.<sup>6,7</sup> Damage to the retrogeniculate visual pathways is common in both of these disorders, since hypoxic-ischemic encephalopathy and periventricular leukomalacia involve all levels of the visual pathway. Neuro-ophthalmologic and perceptual visual disorders are now considered core symptoms of CP in addition to the motor aspects. The severity of visual impairment shows a relationship to the severity of the visual pathway involvement and has been shown to manifest differently in different types of CP (hemiplegia, diplegia, tetraplegia).<sup>6</sup> Multiple studies on the neuro-ophthalmological disorders in cerebral palsy have shown that visuo-cognitive dysfunction, reduced visual acuity, reduced visual fields, and impaired oculomotor abilities are seen in various degrees across the spectrum of CP and CVI.<sup>1,6</sup>

Fazzi et al.<sup>6</sup> found that those with the most extensive visual impairment had tetraplegic CP, which included severely reduced visual acuity (98%), impaired/absent oculomotor abilities (98%), abnormal eye movements, fundus abnormalities, and other neurobehavioral impairments. Those with spastic diplegia, the most common form of childhood CP,<sup>4</sup> showed a high percentage of visuo-cognitive dysfunction, hyperopia, strabismus (90%), reduced

**Table 1. Exam Results from 2/20/18**

Entrance Testing/Refraction	Results
Near grating acuity sc (Teller acuity cards @ 55cm)	OU: 1.30 cycles per degree (~20/470) - Quiet, dim room with cards illuminated - Head tilt to the right with right gaze preference - Latency in response
Contrast sensitivity sc (Double Happy II @ 40cm)	OU: 1.35 log CS, 4.5% Weber contrast
Ocular alignment sc	- Grossly aligned when visually attentive - Variable intermittent alternating exotropia at near
Pupils	ER, minimally responsive to light, (-) RAPD
EOMs	- Limited left and downgaze, compensates with head movement - Full superior and right gazes - Mylar toys used for fixation (initially just visual, then noise was added to engage)
Dry retinoscopy	OD: +1.50 sph OS: +1.00 sph
VF (Modified confrontation with both eyes open in a dim room; central stimulus: examiner; peripheral stimulus: 1.5" diffusely illuminated LED (Kong wand))	Bilateral congruous constriction inferiorly, latency in response to left field
<b>Slit Lamp Exam (Burton Lamp)</b>	
Lids/lashes	Clean OU
Conjunctiva/sclera	White & quiet OU
Cornea	Clear OU
Angle	1:1 T OU
Anterior chamber	D&Q, angles open via penlight OU
Iris	Flat & blue OU
Lens	Clear OU
Tonometry (via iCare @ 1:46pm)	OD: 14 mmHg OS: 16 mmHg
<b>Dilated Fundus Exam: Deferred due to ongoing eye care</b>	
<b>Additional Diagnostic Tests</b>	
Teach CVI/Ortibus CVI Survey Level 1 <sup>11</sup> (Completed by parents and nurse)	4/6 screening items suspicious of CVI 10/19 answers suspicious of CVI
Attention	3/6
Ventral	3/5
Dorsal	
- Complexity	3/4
- Visual guidance	0/2
- Movement	1/2

stereopsis, altered contrast sensitivity, and moderately reduced acuity (82%). Individuals with hemiplegia had strabismus (71%), refractive errors (88%), and altered visual fields (64%).<sup>6</sup> While subjects diagnosed with spastic quadriplegia were not included in this study, many of our patient's findings (including hyperopic refractive error, severely reduced visual acuity, slightly reduced contrast sensitivity, intermittent alternating

exotropia, oculomotor impairment, constricted visual field, and visual perceptual dysfunction) are consistent with the characteristics of the neuro-ophthalmologic manifestations of CP and CVI. It has been shown that damage to the optic radiations or calcarine cortex can result in visual field deficits - with which our patient had been diagnosed and was one of his parent's concerns.<sup>1</sup>

Fazzi et al.<sup>2</sup> also demonstrated that conventional brain MRI did not document major alteration or damage to parietal and temporal white matter or other areas involved in the visual pathway in children with periventricular leukomalacia, spastic diplegia, and normal intelligence; all of their subjects demonstrated deficits in visual object recognition, visual imagery, and visual-spatial skills, among others. They proposed that the lack of findings on MRI might be due to the limitations of its analysis and do not correlate with their findings of impairment of the higher visual processing systems of ventral and dorsal stream processing.<sup>2</sup> Using more advanced forms of neuro-imaging, including diffusion-MRI and f-MRI techniques, has shown promise in finding underlying neurophysiological changes in brain structure and function linked to visual dysfunctions.<sup>12</sup> One study with diffusion MRI showed that white matter arborization was reduced in individuals with CVI and PVL compared to age-matched controls, as well as those with CVI who were born full-term.<sup>12</sup>

Although both of our patient's MRI scans were concluded to be normal, the lack of findings correlating to his neuro-ophthalmologic impairments may be due to limitations in technology and does not affect the diagnosis of CVI. Screening surveys are tools to help identify risk factors for when CVI is suspected in order to detect deficits in functional vision of the dorsal or ventral pathways even when acuity and fields are at near-normal levels; however, they are not diagnostic.<sup>13</sup> Visuo-cognitive dysfunction can only be accurately assessed at ages four to five years.<sup>6</sup> Assuming that our patient's visual function and acuity improved, visual perceptual testing such as the DTVP would allow us to assess the affected visual pathways in order to create a rehabilitation plan. Fazzi et al.<sup>1</sup> reported that 88.9% of the children with CVI whom they were able to test using the DTVP presented with visuo-cognitive disorders. In a systematic review, it was reported that approximately 50% of children with CP have visual perceptual impairment.<sup>5</sup>

Many studies have shown that prematurity has an influence on visuo-perceptual deficits, with

a significantly greater compromise in visuomotor abilities than nonmotor visuo-perceptual abilities when compared to preterm children.<sup>4</sup> In a study, preterm children's scores on the DTVP test were below normal, while the group of term children had normal scores, with a significant difference between the two.<sup>4</sup> Another finding showed that preterm children had a higher presence of oculomotor impairments such as strabismus (93%) than the term children (22%).<sup>4</sup> This study also supports previous evidence that the severity of periventricular leukomalacia correlates with visuo-perceptual deficits, as well as the Griffiths performance score that assesses cognitive abilities. Although this study did not find any major differences in severity or location of periventricular leukomalacia in either preterm or term groups, the authors suggest that using higher-resolution MRI studies to examine the visual pathways in detail and determining whether there are specific anatomical defects related to the visual impairments revealed by the neuropsychological tests could highlight the reasons for these differences and would be a great next step.<sup>4</sup>

## Conclusion

This case illustrates the high comorbidity of CP and CVI. Eye care providers working with patients at risk for CVI should be careful to classify suspect findings as consistent with retinal/anterior visual pathways or posterior pathways. The clinical diagnosis of CP should include not only the motor aspects of the disorder but also the neuro-ophthalmological aspects that have been shown to be present in a majority of cases. A child may present with typical behaviors of CVI despite normal brain imaging, and it is important to identify and to diagnose these patients so that their needs may be met. With advances in structural and functional neuro-imaging, discoveries of abnormalities of white matter connectivity and cortical activation patterns may help to determine visual development potential and may aid in diagnosis.

Intervention through rehabilitation of visual impairment can contribute to the improvement of the motor impairment in CP and lead to a better overall prognosis. Any treatable abnormalities related to CVI, including refractive errors and strabismus, should be carefully managed so as to prevent or treat amblyopia and facilitate a positive visual prognosis. Optometrists and ophthalmologists play a large role in identifying the neuro-ophthalmological

manifestations of CP by conducting a careful history, including reviewing reports from other health care providers and also using screening surveys to help detect functional visual impairment. It is crucial to perform a modified functional visual exam that is adapted to the patient and to discern both visual function and functional vision. Care providers of these patients need to advocate for patients to obtain low vision services, including classrooms and resources for visually impaired students, specialists trained to improve orientation and mobility skills, and the creation of individualized education plans. Early diagnosis, intervention, and rehabilitation through a multidisciplinary approach can positively impact children with CP and CVI.

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