Cerebral visual impairment: the role of ophthalmologists in its identification and intervention

Deficiência visual cerebral: como o oftalmologista pode identificar e intervir

Discapacidad visual cerebral: como el oftalmólogo puede identificar e intervenir

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ABSTRACT
Cerebral or cortical visual impairment is a consequence of impairment or injury in the retrogeniculate visual pathways, primary visual cortex, or associated pathways. Currently, it is one of the major causes of low visual acuity in infants and children in developed countries. Its incidence is high in individuals with multiple disabilities, particularly in those with cerebral palsy. Visual and behavioral findings exhibit specific features, varying in a broad spectrum according to the location and size of the lesion, which may compromise both primary visual and cognitive/perceptual visual functions. Cerebral visual impairment is often associated with ophthalmologic abnormalities and should be carefully investigated. This article aims to present common visual and behavioral characteristics of cerebral visual impairment to allow both the early identification of this condition, which is increasingly frequent in ophthalmologists’ practice, and an early intervention, thus enabling patients to overcome cerebral visual impairment or diminish its impact on the visual development of infants and children.
INTRODUCTION

The causes of visual impairment in childhood are markedly distinct in different regions of the world. This is probably related to social and economic development issues as well as the availability of primary healthcare and eye healthcare institutions. A significant prevalence of visual impairment owing to neurological causes is currently observed, particularly in high-income countries. Cerebral or cortical visual impairment (CVI)\textsuperscript{1,2,3} is particularly relevant because patients with CVI present other associated disabilities. This prevalence is explained by the advances in medicine, which allow high-risk infants to survive, and the identification of CVI based on knowledge regarding this condition and its wide variety of manifestations, in conjunction with families of children with multiple disabilities increasingly seeking early intervention and special education\textsuperscript{4}.

In this context, even in Brazil, it is becoming more and more common in general and pediatric ophthalmology clinics as well as in multidisciplinary healthcare institutes to find infants and children with neurologic lesions who, despite having normal ophthalmologic examinations, present altered or extremely poor visual behavior or even total absence of fixation.

Although CVI has been identified since the 1990s and investigated in several studies, understanding this condition and the implications of its assessment and intervention still poses great challenges, which are becoming more and more significant in the fields of low visual acuity and pediatric ophthalmology. Therefore, it is important to study this condition, considering that the ophthalmologist is the first medical professional to diagnose it.

Visual impairment is generally divided into ocular visual impairment and CVI\textsuperscript{5}. Ocular visual impairment is a consequence of diseases affecting the anterior segment of the eye, ocular attachments, or the anterior visual pathways (retina, optic nerve, optic chiasm, and optic tract). Regarding visual function impairment, it affects the quality of visual images in several aspects, including vision acuity, visual field, contrast sensitivity, color vision, light-dark adaptation, and glare.

CVI is characterized by damage to the visual system in the posterior visual or geniculostriate pathways, occipital cortex, and associated pathways\textsuperscript{5}. In addition to the potential impairment of primary visual functions, CVI may include impairment of visual functions, which several authors refer to as cognitive or perceptive visual dysfunction\textsuperscript{6,7,8}, as well as varied visual-motor, eye-hand coordination, and sensory integration processing issues\textsuperscript{7}. CVI is often associated with simultaneous damage to the anterior visual pathways, particularly the optic nerve or retina.

The first term used to describe the deficit caused by bilateral damage to the visual hemispheres in adults was “cortical blindness”\textsuperscript{9}. This term proved to be inappropriate for children because there is some residual vision and plasticity of the visual system in immature brains. Thus, “cortical visual impairment” has become the most widely used term since the 1990s\textsuperscript{8}. Currently, some authors consider the term “cerebral visual impairment” more appropriate because visual impairment, in addition to cortical alterations, includes subcortical alterations or even an absence of anatomical lesions, in the presence of seizures or metabolic changes\textsuperscript{10,11}.

Because of the underlying neurologic lesion, children and infants with CVI usually present other associated deficits. Therefore, according to Groenveld\textsuperscript{12}, CVI should not be analyzed as an isolated disability, but as a combination of deficits that compromise the access to different types of information and their integration, resulting in a complex of difficulties specific to each child.
ETIOLOGY OF CEREBRAL VISUAL IMPAIRMENT

Children and infants with CVI have a medical and developmental history that indicates the possibility of brain damage, often recognized via magnetic resonance imaging.

The most common impairment leading to CVI is cerebral hypoxia or anoxia, in both term and preterm infants, affecting different brain areas at different levels. In preterm infants, periventricular leukomalacia is a frequently observed condition that causes a wide and variable spectrum of impairments. The typical visual damage owing to a lesion in the upper optic radiations is bilateral inferior hemianopsia. Changes in visual-spatial abilities, visually guided motor actions, eye movements, and visual attention mechanisms are also caused by damage to the adjacent visual pathways of the dorsal stream. Regarding global development, periventricular leukomalacia may have consequences ranging from cerebral palsy—generally spastic diplegia, associated or not with intellectual disability—to only strabismus with learning disabilities.

In term infants, the areas most commonly affected by hypoxia–ischemia are the cortical, subcortical, and white-matter areas, leading to a clinical picture of multicystic encephalopathy, which results in cerebral palsy and intellectual disability, in addition to the visual impairments typical of CVI.

The second most frequent cause of CVI is changes in brain development from genetic, infectious, or idiopathic causes, resulting in structural abnormalities such as myelomeningocele, microcephaly, agenesia of the corpus callosum, and primary congenital hydrocephalus.

Other causes of CVI are brain hemorrhage, particularly intraventricular hemorrhage in premature infants, brain trauma, and hydrocephalus. The latter may lead to isolated or combined visual deficits.

CHARACTERISTICS OF CEREBRAL VISUAL IMPAIRMENT

The nature of the visual alterations in CVI is related to the location and size of the neurologic lesion; however, understanding these changes remains a great challenge. The very complexity of the visual system and its integration with other perception and motor systems, the difficulties inherent to the evaluation of infants and children with severe disabilities, particularly using standardized and quantitative behavioral tests, as well as the great variability observed in visual behavior, all make assessing CVI quite difficult.

IMPAIRMENT OF PRIMARY VISUAL FUNCTIONS AND OCULAR MOTILITY IN CEREBRAL VISUAL IMPAIRMENT

The impairment of primary visual functions such as visual acuity, visual field, contrast sensitivity, and color vision can occur with wide variability and in multiple combinations.

Visual acuity is generally reduced in CVI, but blindness is rare. Assessing resolution acuity with the gaze preference technique using Teller acuity cards is a procedure used to estimate visual acuity as well as visual evoked potentials. Although visual evoked potentials are used to determine visual acuity in preverbal children and in those with multiple disabilities, the result may not reflect the actual status of visual impairment in those with CVI. It can be overestimated owing to the cognitive visual dysfunctions resulting from changes beyond the primary visual cortex, either in the ventral or dorsal stream, that greatly impact vision. When recognition acuity assessment using symbols is possible, it provides more detailed information about the visual system’s discerning ability. A noteworthy phenomenon in CVI is crowding, in which visual acuity may be much better with symbols presented in isolation than when grouped in line, or even using specific crowding test tables.

Contrast sensitivity may be either normal or greatly decreased. It is observed in practice that high-contrast image contours, whether in symbols, figures, or even objects, facilitate visualization and perception, probably because they help decreasing crowding.

Regarding color vision, different authors report that it is usually normal. A strong preference for one color, usually red or yellow, is common.

Visual field changes related to the area where the lesion is located are frequent. Thus, unilateral damage to the occipital lobe or geniculostriate pathways can lead to contralateral hemianopsia. Lesions affecting the upper optic radiations bilaterally cause inferior visual field loss. While analyzing clinical data from 309 individuals with CVI, with most of them...
being children, Bosh et al. could assess the visual field of 249, of which 60% showed changes: 19% had hemianopsia, 25% had upper or inferior visual field impairment, and 56% had visual field constriction\(^1\). \(^1\)

Ocular motility and static disorders are frequent and include the following: strabismus; nystagmus; fixation instability; inaccuracy, restriction or absence of movement to a certain direction of gaze, both saccadic and/or pursuit movements; instability of eye and head movements; and conjugate eye deviation to a given direction of gaze.

Infants with extremely poor visual development commonly exhibit eye and head rotation to the same side, with pursuit movements that fail to cross the midline. However, even after having achieved good visual performance with the intervention process, this behavior may appear to be caused by fatigue or when concurrent and complex stimuli are received\(^1\).\(^1\)\(^1\)

Another atypical behavior in some children with CVI and cerebral palsy is repeatedly presenting intense extensor synergy posturing, in which the eyes gaze upwards. This behavior involves the neck, trunk, and lower limbs\(^1\).\(^1\)

**REFRACTION AND ACCOMMODATION**

Children with brain damage have a higher risk for significant refractive errors: myopia, hyperopia, and astigmatism. The presumed etiology is that at birth, the statistical distribution of refractive errors is similar to that in other children with normal vision; however, the process of emmetropization fails owing to low visual acuity, which leads to a high prevalence of refractive errors in children of school age\(^2\).\(^2\) It is often difficult for affected children to accept glasses, particularly those with cerebral palsy and tactile hypersensitivity. They also reject glasses when the vision does not significantly improve with the use of glasses. This is more frequent in near-sighted children whose near vision is adequate without correction\(^2\).\(^2\) Alternatively, it should be noted that accommodation may be impaired in children with brain damage\(^2\) and that in the case of hypermetropia, it leads to a constantly defocused retinal image.

**COGNITIVE VISUAL DYSFUNCTIONS**

To make comprehension easier, cognitive visual dysfunctions can be characterized as a consequence of lesions in the dorsal or ventral stream.

The system comprising the dorsal stream, located in the posterior occipital–parietal brain, is called the “how” system. It is responsible for visually guided actions (for example, reaching and grasping), visual–spatial demands, egocentric and allocentric localization, and control of saccadic eye movements\(^2\), as well as for attention mechanisms.

The attention mechanisms allow the visual system to select an object of interest that is part of a complex scene, putting the rest of the scene in the background. These mechanisms also allow us to switch the gaze and visual attention from one object to another at a different spatial location in the visual scene\(^2\)\(^2\).

Therefore, any damage to the dorsal system will primarily compromise the visually guided motor actions, for example, accurately finding and grasping an object. It affects the ability to process different concurrent visual information, which is necessary to understand complex scenes, compromising the capacity to find figures in non-uniform backgrounds, a specific object among others, a person in crowded places, or letters grouped in a word. These findings are part of the group of crowding difficulties, often expressed as irritability, fatigue, or lack of visual interest\(^1\),\(^2\),\(^2\)\(^2\),\(^2\)

The ventral stream, in the inferior posterior occipital–temporal brain, is responsible for visual perception and thus called the “what” system. In association with memory, semantics, and communication, it allows the identification or recognition of visual objects as well as the attribution of meanings from our internal representation of the visual world\(^2\).\(^2\) There is evidence that, for each category of images—i.e., for objects, faces, places, etc.—there are different specialized areas to decode them\(^2\)

Damage to the ventral stream can hinder visual recognition, which can manifest as difficulty in recognizing one category of stimuli by itself. This may be concrete objects, places, figures and symbols, faces (prosopagnosia), or facial expressions. It may also not be selective, compromising the whole perceptual function of visual recognition, totally or with different levels of difficulty.

Although parallel and independent, these two subsystems are complementary and allow the person to distinguish between the visual scene and the action performed on it\(^2\)\(^2\).

Children with CVI may also present other impairments, such as memory and spatial orientation
and sensory integration. When brain function is normal, sensory information from several senses and motor actions are integrated, resulting in a holistic experience on and with objects. If sensory integration is impaired, the perception of different sensory channels is not simultaneous but either isolated or sequential, which leads to an inability to simultaneously perform two actions, such as maintaining gaze fixation on an object and grasping it.

**BEHAVIORAL CHARACTERISTICS IN CVI**

Some visual behaviors typical of CVI are symptoms of visual dysfunction, and they interfere with the use of vision to varying degrees, depending on the severity of the condition and on the manner they manifest themselves, which is unique for each child.

Variability in visual functioning is one of the most frequently observed characteristics. Several environmental factors, general health status, and the occurrence of seizures are the leading causes of fluctuation in the use of visual function. Vision varies from one day to the next or even from one moment to another, which hinders the use of quantitative and standardized visual tests.

Attraction to lights (light gazing) and non-purposeful gaze are extremely common. This is an innate and compulsive behavior, for which there may be a specific neuropathologic substrate. Flickering hands in the direction of a light source is a derivation of the attraction to lights. Paradoxically, despite being attracted to lights, these children are often intensely photophobic.

In infants with an extremely low level of visual function, preference for a single color, generally red or yellow, as well as for objects with movement or the illusion of movement (such as acetate wigs) can be observed. Images on the TV or tablets and ceiling fans are attractive, but this does not mean the child is “seeing” them.

Familiar objects are preferred to new ones because repetition facilitates visual processing and recognition. This implies an extremely careful choice of assessment and intervention materials, which should have the qualities of the objects that draw the child’s attention and should be repeatedly presented and integrated to their daily life.

Another relevant characteristic is a greater delay in responding to a visual stimulus, i.e., increased visual latency. This is owing to difficulties in visual processing and organization of the motor response. This important aspect of behavior should be considered in assessment and intervention.

Several children with CVI show a preference for a region of the visual field, i.e., they can only maintain fixation on a certain region. Several factors might be involved, such as disorders of eye movement, with or without normal head position, visual field deficits, and left visual hemifield neglect, in which there is no awareness of anything on the left side, often including the individual’s own body.

Difficulty with simultaneous or cluttered visual information owing to the intrinsic qualities of either the visual stimulus itself or its surroundings is another typical visual behavior observed in CVI. Some children only respond visually to an object when it is illuminated with a light beam and the environment is darkened, so that only the object can be seen and the surroundings disappear from the visual sphere. They may need to approach the object, even when their visual acuity is not reduced, to decrease the amount of information in their visual field.

Visually locating an object, followed by turning the gaze or the head to the opposite side when performing the motor action of reaching for it, is another relevant behavioral characteristic.

Inadequate control of the head, neck, and trunk plays an extremely important role in the use of vision; therefore, positioning and balance issues influence the visual performance of children with CVI. Therefore, it is not always possible to obtain visual responses if the children have not been positioned according to their needs. Help from a physical or occupational therapist is often necessary to obtain better positioning and hence the best visual responses.

Changes in spatial perception, depth perception (both binocular and monocular), and distance judgment can hinder mobility even in functionally ambulant children. Vestibular changes as well as changes in the proprioceptive feedback from the neck and eye muscles and central balance pathways are often combined with dorsal stream damage. These changes play an important role in both eye statics and dynamics as well as in the conjugate movements of the head.

Another important characteristic, which influences the use of vision and the general behavior of children with CVI, is hypersensitivity: tactile, auditory, and related to other sensory channels. Hypersensi-
tivity manifestations are highly variable and lead to irritability or to task or contact avoidance\textsuperscript{28}.

**ASSESSMENT AND IDENTIFICATION OF CVI**

The task of assessing a child or infant with CVI is a major challenge owing to the complex patterns of possible findings in CVI, which can range from deep impairments to subtle deficits in specific visual functions. The neurologic lesion is usually non-focal and is comorbid with multiple neuropsychological inabilities\textsuperscript{32}.

This clinical picture of multiple disabilities, in addition to hampering the possibilities of response to standardized tests, hinders the determination of the cause of a particular impairment\textsuperscript{32} as a deficit in visual perception tasks may result from an ocular lesion or cognitive problems (among others). There may also be a combination of deficits in basic visual functions (such as visual field, visual acuity, and contrast sensitivity) and cognitive visual abilities\textsuperscript{28}.

Because of its particularities, every assessment of an individual with CVI must include, whenever possible, the evaluation of functional vision, in addition to ophthalmologic and standardized visual function examinations. The content of the functional vision evaluation should consider the specificities of CVI\textsuperscript{33}.

The evaluation and diagnostic process involves and is determined by other medical information, such as neurological and neuroimaging data as well as information from family and early-intervention and special-education staff\textsuperscript{34}.

The collaborative, transdisciplinary model is the most recommended model to respond to the questions related to the complexity of CVI\textsuperscript{7,34}. The transdisciplinary model, unlike the multidisciplinary one, enables all members of a team to collaborate in sharing their knowledge and skills, to go beyond each field’s specific goals. It allows professionals to cumulatively develop more efficient and comprehensive strategies that can promote global development. In this context, family is an integral part of this process\textsuperscript{4,34}.

Family provides important information about the child’s history and current state. Through comprehensive and systematic information collection by caregivers, it is possible to obtain knowledge regarding the child’s visual functioning in daily life and in different environments. The lead of this information collection on visual abilities is the behavioral characteristics of CVI, which provide clues regarding dysfunctions related to cognitive perception, and in association with team evaluations, help to understand the infant’s or child’s visual functioning\textsuperscript{17,27,35}.

A few authors have presented frameworks that can be used as guides to assess functional vision in CVI, provided they are contextualized according to the global development of the children or infants and combined with their medical information and with that obtained from their families\textsuperscript{7,17,34,36,37}.

**INTERVENTION IN CVI**

In ocular visual impairment, in which the region of the brain that receives and analyzes the visual information is intact, the intervention is focused on enriching visual information. However, in CVI, this approach is not effective because owing to the changes in the processing and interpretation of visual images, the greater the amount of information, the more difficult it is for the child to process the visual impulse and respond visually. Another dimension is added to the intervention program\textsuperscript{8,17}.

The premise on which the intervention program is based is that CVI can be mitigated or even overcome in some cases\textsuperscript{17}. Improvement occurs owing to brain plasticity and the possibility of neuronal growth and new specializations and brain synapses in infants and children\textsuperscript{38}. However, for this to happen, it is necessary to enable children to see and to motivate them to see through repeated structured and well-oriented seeing experiences.

The possibility of recovery depends on several factors, such as the type of intervention received, the etiology, and even the location of the brain injury. Improvement usually occurs with brain maturation and the experiences gained through sensory integration. It may occur over a period of many years; however, the best prognosis is when the intervention is administered up to the age of three years\textsuperscript{12}.

The intervention process should not be seen as a rehabilitation process but as an approach that should be a part of the individual’s whole routine as the strategies should be integrated in their daily life, functional learning activities, self-care, leisure activities, etc. Interventions depend on CVI characteristics, age, level of development, and level of school or preschool functioning\textsuperscript{33}.
Essentially, the child or infant should be shown what she or he is able to see; interventions should be planned to meet and not to surpass the level of visual functioning observed upon assessment\textsuperscript{17,33}.

Children with brain damage can rapidly fluctuate between different “behavioral states,” such as focused/absent, quiet/crying, or alert/drowsy, which has repercussions on their adaptability and learning ability\textsuperscript{35}, and consequently, in their visual functioning. Their behavior is generally influenced by both internal and external conditions. Internal conditions relate to their general state of health, fatigue, stress, unfamiliarity, hunger, or convulsions. External conditions are noise, temperature, excess information, and hyperstimulation. It is necessary to observe the favorable conditions for the infant or child to be alert, such as time control. The type, intensity, and duration of sensory information, both visual and from other channels, should be controlled. This should include the environment, stimuli, and also the professional’s behavior\textsuperscript{28}. For example, it is often necessary to give an oral command prior to an activity and refrain from speaking while the child does it and to always pay attention to the signs they show about their status\textsuperscript{28,22}.

As progress is perceived, a new assessment should be made, while always maintaining a systematic approach based on the levels of the CVI characteristics\textsuperscript{17}.

Another factor to be considered is the limits imposed by impairments from ocular causes, which may coexist with CVI.

**FINAL CONSIDERATIONS**

There are difficulties in identifying the wide and complex variety of visual dysfunctions in CVI, from those related to primary visual functions to cognitive or perceptual visual dysfunctions. The absence of robust assessment instruments and the presence of comorbidities, particularly in those with diffuse brain damage and those with severe disabilities, often hinder the assessment of visual changes.

A collaborative effort, including different fields of knowledge, is advocated because this model allows integrating clinical tests with evaluations of functional vision and visual behavior in different situations, enabling some degree of understanding regarding patient’s visual perception, use of vision in movements, and interaction with the environment.

In addition to the collaboration among several professionals (such as pediatricians, neuropsychiatrists, neuropsychologists, ophthalmologists, occupational therapists, physical therapists, speech-language therapists, and special-education and regular-education teachers), family participation is crucial, for not only providing information about visual behavior in conditions different from those seen at the clinic but also regarding active participation in the intervention process, which should be integrated into the child’s daily life to be effective.

**REFERENCES**


