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Special Commentary: Cerebral/Cortical Visual Impairment Working Definition

A Report from the National Institutes of Health CVI Workshop

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Cerebral/cortical visual impairment (CVI), a brain-based condition, has emerged as a leading cause of pediatric visual impairment in the United States and other industrialized nations. The National Eye Institute (NEI) recognized CVI as a priority area for research as part of their 2021 NEI Vision for the Future Strategic Plan and partnered with the Eunice Kennedy Shriver National Institute of Child Health and Human Development and the National Institute of Neurologic Disorders and Stroke within the National Institutes of Health (NIH) to sponsor a CVI Workshop in November 2023. A panel consisting of a group of clinicians with expertise in diagnosing CVI convened to draft a working definition for this condition. Five key elements were identified: (1) CVI encompasses a spectrum of visual impairments caused by an underlying brain abnormality that affects the development of visual processing pathways and is characterized by deficits in visual function and functional vision; (2) the visual dysfunction in CVI is greater than expected by any comorbid ocular conditions alone; (3) the visual dysfunction in CVI may manifest as lower-order or higher-order afferent visual deficits, or both, leading to characteristic behaviors in affected individuals; (4) although CVI may be comorbid with other neurodevelopmental disorders, CVI is not primarily a disorder of language, learning, or social communication; and (5) the underlying neurologic insult of the developing brain may go unrecognized or undiagnosed until later in life. Future work is needed to achieve consensus on nomenclature, diagnostic criteria, and strategies for early identification and intervention. The NIH is developing a CVI registry to collect relevant demographic and clinical data prospectively and longitudinally to help inform future research questions and to provide insight into considerations for future clinical trials in the field of CVI. *Ophthalmology 2024;131:1359-1365 © 2024 by the American Academy of Ophthalmology*

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In the past several decades, cerebral/cortical visual impairment (CVI), a brain-based condition, has emerged as a leading cause of pediatric visual impairment in the United States and other industrialized nations.[1–4] It is a diagnostic entity distinct from cortical blindness. Historically, Whiting et al[5] noted that "acquired cortical blindness in adults may be quite different from congenital or early-onset cortical visual loss in children." Cortical blindness in adults is characterized by complete (or near complete) loss of vision resulting from bilateral lesions of the occipital cortex, most frequently in the setting of posterior circulation strokes.[6] In contrast, individuals with CVI (for further details, see Note regarding nomenclature at end of commentary) may have lesions that extend beyond the occipital cortex and may have visual deficits beyond reduced visual acuity.[7–10] These patients usually are not completely blind, and instead have some level of vision and functional visual abilities that may improve over time.[5,11,12] The difference in trajectory is presumed to be related to underlying neuroplasticity in the developing visual system.[13]

In the first descriptions of CVI, children demonstrated (often profound) decreased visual acuity associated with damage to the occipital cortex, optic radiations, or both.[5,14] Many clinicians now have adopted a broader definition inclusive of CVI in individuals with higher-order visual processing deficits in the setting of normal to near-normal visual acuity.[15–17] In 2018, Sakki et al[18] proposed a definition of CVI based on a systematic review of available literature. Using a thematic analysis approach, the authors defined CVI as "a verifiable visual dysfunction which cannot be attributed to disorders of the anterior visual pathways or any potentially co-occurring ocular impairment."[18] Although representing an important step forward in helping to improve communication regarding this condition, this definition is limited. It primarily focuses on distinguishing CVI from ocular-based causes of visual

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impairment (which may coexist in some patients), and as a result, CVI is rendered more as a diagnosis of exclusion without further characterization of the types of visual deficits experienced by these individuals. Moreover, although visual dysfunction in CVI indeed should be determined objectively, additional details from appropriate assessments are needed, and it is important to disentangle the contributions of other neurodevelopmental comorbidities. Finally, the definition does not explicitly identify a neurologic insult or maldevelopment of the visual pathways in the brain as the underlying pathophysiologic feature of CVI.

The National Eye Institute (NEI), one of the National Institutes of Health (NIH), has recognized CVI as a priority area for research as part of their 2021 NEI Vision for the Future Strategic Plan.[19] Further, the NEI acknowledged that the interdisciplinary nature of this condition requires coordinating efforts across specialties. As such, the NEI partnered with the Eunice Kennedy Shriver National Institute of Child Health and Human Development and the National Institute of Neurological Disorders and Stroke to sponsor an NIH CVI Workshop (Roadmap to Consensus and Building Awareness, https://www.nei.nih.gov/events/cvi-workshop) in Bethesda, Maryland, on November 16-17, 2023. As part of this workshop, a group of clinicians with expertise in diagnosing CVI convened to initiate a discussion on how to reconcile conflicting views and to propose a definition for this condition. With these goals in mind, the panel began drafting a working definition of CVI and settled on 5 key elements listed below. Although all panelists believed that more discussion was necessary to solidify the definition, we were in overall agreement that these elements encompass most patients with CVI typically encountered in clinical practices.

Cerebral/Cortical Visual Impairment Working Definition

(1) CVI encompasses a spectrum of visual impairments caused by an underlying brain abnormality that affects the development of visual processing pathways and is characterized by deficits in visual function and functional vision. (2) The visual dysfunction in CVI is greater than expected by any comorbid ocular conditions alone. (3) The visual dysfunction in CVI may manifest as lower-order or higher-order afferent visual deficits, or both, leading to characteristic behaviors in affected individuals. (4) Although CVI may be comorbid with other neurodevelopmental disorders, CVI is not primarily a disorder of language, learning, or social communication. (5) The underlying neurologic insult of the developing brain may go unrecognized or undiagnosed until later in life.

Rationale for 5 Elements of the Cerebral/Cortical Visual Impairment Working Definition

1. CVI encompasses a spectrum of visual impairments caused by an underlying brain abnormality that affects the development of visual processing pathways and is characterized by deficits in visual function and functional vision.

CVI is a heterogeneous disorder with respect to cause and clinical presentation. It impacts multiple aspects of visual function.[7,9,14,20] Furthermore, all individuals with CVI are likely to have some degree of functional vision impairment, although this may be difficult to assess because of reduced visual acuity, cognitive abilities, or overall level of functioning. As defined by Colenbrander,[21] visual function refers to how the eye functions (measured by clinical tests such as visual acuity), whereas functional vision refers to how the person functions while performing activities that require vision. Observations in a standard clinical setting may not accurately reflect difficulties with functional vision at home, school, or other situations.[15,20] It is also critical to understand that in this context, functional vision is unrelated to the terms *functional vision loss* or *functional vision disorder*, both of which are used to indicate nonorganic visual disturbances that are, in some cases, suspected to be psychogenic in origin.[22]

All patients with CVI have an underlying neurologic condition impacting the development and functioning of visual pathways in the brain that accounts for their visual deficits. Neurologic disorders associated with CVI include, but are not limited to, hypoxic−ischemic encephalopathy, prematurity with periventricular leukomalacia, trauma, hydrocephalus, seizures (especially with epileptic encephalopathy), as well as genetic and metabolic conditions. In many patients with CVI, a structural abnormality implicating the optic radiations, occipital cortex, visual association areas, or a combination thereof is detectable on neuroimaging.[23,24−27] It is important to note that structural abnormalities may not always be readily evident in association with certain causes (e.g., seizure activity with epileptic encephalopathy or a genetic or metabolic disorder) with standard brain neuroimaging methods.[28] However, underlying structural and functional changes may be detected using specialized neuroimaging techniques, such as diffusion-based imaging,[29,30] functional neuroimaging,[31] and resting-state functional connectivity.[10]

2. The visual dysfunction in CVI is greater than expected by any comorbid ocular conditions alone.

Ocular comorbidities are common also in patients with CVI, especially optic nerve abnormalities such as optic nerve atrophy, optic nerve hypoplasia, and the characteristic increased cupping seen in periventricular leukomalacia.[5,9,32−36] Retinal disorders such as retinopathy of prematurity[32−34] also have been described in association with CVI. Because these ocular conditions are expected to impact visual function, CVI should be diagnosed only when visual dysfunction cannot be explained by the degree of ocular pathologic features alone.[18] Observed visual deficits may differ in nature and severity from those expected based on the appearance of the retina and optic nerve.

Oculomotor comorbidities can include strabismus, nystagmus, oculomotor apraxia, absent vestibular−ocular reflex, delayed or dysmetric saccades, impaired pursuits, and fixation instability.[34,37−40] Patients with CVI also may experience difficulties with binocular function and accommodative insufficiency.[41] These comorbid oculomotor conditions may cause secondary deficits of vision and thus

should be addressed, given that they may impact an individual's functioning in other ways (such as reading) and overall development and quality of life. Although assessing oculomotor function was deemed important, the panel also reached agreement that these disturbances alone do not constitute a diagnosis of CVI.

Other ophthalmologic conditions that are considered separate from CVI include amblyopia and delayed visual maturation (DVM),[42,43] which is in the differential diagnosis of an infant with visual impairment. Although the underlying pathophysiologic features in amblyopia relate to maldevelopment of the visual cortex, amblyopia is not considered to be CVI because the inciting factor is an ocular-based or binocular-based disorder (such as uncorrected refractive error, strabismus, or both) that causes abnormal visual input to the brain.[44] In DVM, the pathophysiologic features remain unknown, and infants with DVM initially may demonstrate visual behaviors similar to those associated with CVI.[45] Crucially, however, in DVM, visual behavior generally normalizes by 6 months,[42,43,46,47] whereas visual dysfunction persists in children with CVI.

3. The visual dysfunction in CVI may manifest as lower-order or higher-order afferent visual deficits, or both, leading to characteristic behaviors in affected individuals.

The initial presentation of young children with CVI is likely to be focused primarily on lower-order afferent visual deficits (i.e., abnormalities that are attributed to dysfunction of the visual pathway from the eye to the primary visual cortex). These include decreased visual acuity, reduced contrast sensitivity, and visual field defects.[5,9,48,49] When lower-order visual deficits are profound, higher-order visual dysfunction may be difficult or impossible to detect and confirm. However, abnormalities of visual processing may be suspected on the basis of some of the visual behaviors that are common in young children with CVI. Behavioral characteristics of CVI include light gazing, eccentric gaze preference, preference for brightly colored and moving objects, difficulties with detecting objects that are closely spaced or on complex backgrounds, and fluctuating levels of vision, with worse visual behavior associated with fatigue, illness, or unfamiliar environments.[14,50–53]

In patients with CVI whose visual acuity is measured at normal or near normal levels (frequently in older patients), higher-order visual processing deficits manifest as the main visual impairment.[17] Therefore, normal performance on the standard ophthalmic clinical examination does not exclude the presence of CVI. Higher-order visual deficits are attributed to dysfunction of visual processing pathways, including visual association areas and extending to the dorsal and ventral stream pathways.[13,30,54] Examples of higher-order visual function deficits described in individuals with CVI include difficulties with complex motion perception, face and object recognition, visual search, visuospatial orientation, and simultanagnosia.[7,31,55–59]

Directed history, visual assessment, and eye examination remain the mainstay for making the diagnosis of CVI.[60,14,61] Semistructured history-taking tools (questionnaires and inventories) should be used to elucidate higher-order visual processing deficits, especially in patients with good visual acuity.[17,62–65] A variety of ancillary techniques also may be useful to screen and measure the visual deficits in CVI. The appropriateness of each assessment varies depending on an individual's visual, motor, and cognitive abilities (detailed discussion is beyond the scope of this article). The ancillary techniques may include perimetry,[48] preferential looking tests,[66–68] standardized neuropsychological and functional vision assessments,[60,28,69–71] visual evoked potentials,[38,58,67,72,73] and eye tracking.[40,68,74–76]

4. Although CVI may be comorbid with other neurodevelopmental disorders, CVI is not primarily a disorder of language, learning, or social communication.

Several co-occurring neurodevelopmental conditions are common in the setting of CVI, and their assessment and characterization are crucial to understand better their impact on visual function and processing.[21,77] Cerebral palsy is a frequent comorbidity with CVI.[34] Other neurodevelopmental disorders, such as autism and dyslexia, may co-occur with CVI, but also may have overlapping manifestations, leading to diagnostic difficulties. Individuals on the autism spectrum have challenges with social communication and social interaction, including difficulties with social–emotional reciprocity, nonverbal communication behaviors, and social relationships, along with restricted and repetitive patterns of interests, behaviors, activities, or a combination thereof.[78] Dyslexia is a learning disability characterized by difficulties with word recognition, spelling, and decoding.[79,80] Although vision is an important sensory method for social interaction and reading text, abnormal visual function or processing alone cannot explain the difficulties experienced by individuals with autism or dyslexia. When present in the case of CVI, the severity of these conditions is likely related to the cause, timing, and nature of the underlying neurophysiological differences. A diagnostic dilemma remains in that CVI may be misdiagnosed or underdiagnosed in children who are suspected to have other neurodevelopmental disorders. Uncovering the overlap and boundaries of these comorbidities remains challenging in the case of CVI. At the same time, consideration should be given to diagnostic prioritization and the impact of multiple diagnoses for the same individual, with the understanding that a multidisciplinary approach in such cases will be essential.

Assessment tools and batteries appropriate for individuals with CVI (which may need to be adapted for specific comorbid conditions such as cerebral palsy) are necessary to disentangle core and secondary features across ages and ranges of functioning. Modifications of standard assessments for autism in children with ocular causes of visual impairment have been published,[81] and similar studies are needed in individuals with CVI.

5. The underlying neurologic insult of the developing brain may go unrecognized or undiagnosed until later in life.

In some cases, the CVI diagnosis is not made early on because children may be unable to recognize or express their functional vision deficits until later in development. Screening for CVI should be considered in individuals with a high suspicion of early neurologic injury impacting visual pathways and processing areas, such as those with periventricular leukomalacia. The term *CVI suspect* was proposed at the workshop to refer to individuals for whom a

high degree of clinical suspicion is present based on medical history and initial presentation, but further objective testing and treatment of ocular comorbidities are needed before confirming the diagnosis of CVI. The currently held view among experts is that an early diagnosis of CVI is imperative so that appropriate accommodative strategies are in place during development.[61,82] Indeed, a concerted effort is underway to diagnose CVI early, as soon as alternate diagnoses (such as delayed visual maturation) can be ruled out. An important component of early diagnosis is screening by pediatricians. A recent American Academy of Pediatrics Clinical Report on the diagnosis and care of children with CVI provides guidance to help pediatricians and other clinicians identify and refer children with CVI as early as possible.[83]

The first decade of life is the most critical time for the maturation of visual function and functional vision or perception.[84,85] Despite these well-established critical periods of vision development,[85] evidence is emerging that visual processing pathways have a sensitive period or window of time with active structural and functional brain alterations extending past the first decade.[13,86,87] Whether these are dynamic physiologic changes or continued developmental processes remains an ongoing debate. It is important to realize that the timing of brain injury significantly impacts the functional, structural, and behavioral consequences of visual impairment.[88]

It is crucial that appropriate support is available for individuals with CVI throughout their lifespan. At this time, management of CVI primarily is focused on environmental adaptations, accommodations, and habilitation with treatment of any associated ocular comorbidities.[9,82,89] Future research is needed to identify evidence-based medical treatments specific to CVI. Management strategies should adjust to the differing needs of individuals as they age; however, evidence exists that most children with developmental disabilities, including visual impairment, do not receive adequate transition planning to adult care.[90,91] Recommendations include initiating early planning for transition and establishing care with an adult provider before stopping care from a pediatric provider.[90,91] Individuals with CVI also require individualized support for educational, vocational, recreational, and social needs across their lifespans.[90,91] Thus, both pediatric and adult practitioners should be aware of the impact of this condition and should provide long-term follow-up and support at each stage of life.

Conclusions

The NIH CVI Working Group proposes that CVI is a neurodevelopmental disorder characterized by deficits of visual function and functional vision that are caused by neurologic damage to visual pathways and processing areas in the brain. Anterior visual pathway disorders, such as optic atrophy or retinopathy of prematurity, may be comorbid with CVI, but do not explain the level of visual impairment. Other neurodevelopmental disorders, such as autism and dyslexia, are distinct from CVI, but also may be comorbid. Diagnostic assessments to support the requirement that visual deficits exceed those expected by any comorbid conditions may include quantitative tests of visual acuity, electrophysiologic testing (such as visual evoked potentials), semistructured history-taking questionnaires, neuropsychological testing, standardized functional vision assessments, and investigational techniques such as eye tracking. Neuroimaging and genetic testing may be indicated to identify and characterize underlying neurologic conditions, but abnormalities on these tests are not required to make a diagnosis of CVI. Although early diagnosis remains the goal, CVI may be recognized and diagnosed in both childhood and adulthood.

We acknowledge that this working definition of CVI must be followed by standardized diagnostic criteria for CVI and considerations for identifying CVI suspects, as well as standardized nomenclature of CVI (e.g., cortical vs. cerebral) to promote appropriate clinical care (especially early diagnosis) as well as research.[92] Areas of future research include establishing the neurophysiological basis and timing of neuroplasticity in the developing brain of individuals with CVI, standardizing methods of visual assessment, clarifying the impact of underlying cause on visual manifestations, identifying prognostic markers, understanding how the functional deficits of individuals with CVI change throughout their lifespan, and evaluating the efficacy of proposed interventions and rehabilitation strategies. The NIH is developing a CVI registry to collect relevant demographic and clinical data prospectively and longitudinally to help inform these research questions and to provide insight into considerations for future clinical trials in the field of CVI.

Note regarding nomenclature: Although Whiting et al used the term *cortical visual impairment* in their original report, more recently, *cerebral visual impairment* has become more widespread in the literature, acknowledging that subcortical structures, including white matter pathways, and visual association areas may be involved in CVI.[18,60] This report is intended to establish a working definition of CVI that encompasses both individuals who have received a diagnosis of cortical visual impairment, consistent with the original description, as well as others with a diagnosis of cerebral visual impairment who may have deficits that involve other areas of the visual pathway.[23,29]

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Abbreviations and Acronyms:
CVI = cerebral/cortical visual impairment; **DVM** = delayed visual maturation; **NEI** = National Eye Institute; **NIH** = National Institutes of Health.

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