AMERICAN ACADEMY OF OPHTHALMOLOGY®

Special Commentary: Cerebral/Cortical Visual Impairment Working Definition

A Report from the National Institutes of Health CVI Workshop

Melinda Y. Chang, MD,[1,2,] Lotfi B. Merabet, OD, PhD,[3,4,*] for the CVI Working Group*

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

https://doi.org/10.1016/j.ophtha.2024.09.017
ISSN 0161-6420/24

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]

Acknowledgments

The workshop organizers thank all speakers for their contributions, including individuals with CVI and their families, to the overall success of the workshop.

Footnotes and Disclosures

Originally received: August 1, 2024.
Final revision: September 6, 2024.
Accepted: September 12, 2024.
Manuscript no. OPHTHA-D-24-01592.

[1] Children's Hospital Los Angeles, Division of Ophthalmology, Department of Surgery, Los Angeles, California.

[2] Department of Ophthalmology, University of Southern California, Los Angeles, California.

[3] Massachusetts Eye & Ear, Boston, Massachusetts.

[4] Department of Ophthalmology, Harvard Medical School, Boston, Massachusetts.

*Both authors contributed equally to this work and are co-chairs of the CVI Working Group.

Disclosure(s):

All authors have completed and submitted the ICMJE disclosures form.

The author(s) have made the following disclosure(s):

M.Y.C.: Financial support — National Eye Institute (grant no.: K23EY033790), Research to Prevent Blindness, Inc., Knights Templar Eye Foundation

This work was supported by the National Eye Institute, the National Institute of Neurological Disorders and Stroke, and the Eunice Kennedy Shriver National Institute of Child Health and Human Development within the National Institutes of Health. This article represents the perspective of the National Institutes of Health CVI Workshop expert panel and does not necessarily represent the views of the National Institutes of Health or the United States Department of Health and Human Services.

CVI Working Group Members: Mark Borchert (Children's Hospital Los Angeles, University of Southern California), Arvind Chandna (Smith Kettlewell Eye Research Institute, Alder Hey Children's Hospital), Gena Heidary (Boston Children's Hospital, Harvard Medical School), Barry Kran (New England College of Optometry, Perkins School for the Blind), Linda Lawrence (Private Practice), Richard H. Legge (Truhlsen Eye Institute, University of Nebraska Medical Center), Sharon Lehman (Wills Eye Hospital, Nemours Children's Hospital, Sydney Kimmel Medical College), Melissa Rice (Cincinnati Children's Hospital Medical Center, University of Cincinnati), Veeral Shah (Cincinnati Children's Hospital Medical Center, University of Cincinnati), Steve Stasheff (Children's National Hospital, George Washington University), and the following staff of the National Institutes of Health: Daofen Chen (National Institute of Neurologic Disorders and Stroke), Theresa Cruz (National Institute of Child Health and Human Development), Kerry Goetz, (National Eye Institute), Shefa Gordon (National Eye Institute), Kristi Hardy (National Institute of Neurologic Disorders and Stroke), Alicia Kerr (National Eye Institute), Erika Nelson (National Eye Institute), Ralph M. Nitkin (National Institute of Child Health and Human Development), Melissa A. Parisi (National Institute of Child Health and Human Development), Amber Reed (National Eye Institute), Santa J. Tumminia (National Eye Institute), Cheri Wiggs (National Eye Institute).

HUMAN SUBJECTS: Human subjects were not included in this study. This is a special commentary that is not considered human research and the tenets of the Declaration of Helsinki do not apply. There was no requirement for informed consent because there were no human subjects.

No animal subjects were included in this study.

Author Contributions:

Conception and design: Chang, Merabet

Analysis and interpretation: Chang, Merabet

Data collection: Chang, Merabet

Obtained funding: N/A; Study was performed as part of the authors' regular employment duties. No additional funding was provided.

Overall responsibility: Chang, Merabet

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

Keywords:
Cerebral visual impairment, Cortical visual impairment, Working definition.

Correspondence:
Melinda Y. Chang, MD, 4650 Sunset Boulevard, Mailstop No. 88, Los Angeles, CA 90027. E-mail: Melinda.y.wu@gmail.com.

References

1. Williams C, Pease A, Warnes P, et al. Cerebral visual impairment-related vision problems in primary school children: a cross-sectional survey. *Dev Med Child Neurol*. 2021;63(6):683–689.
2. Solebo AL, Rahi J. Epidemiology, aetiology and management of visual impairment in children. *Arch Dis Child*. 2014;99(4):375–379.
3. Kong L, Fry M, Al-Samarraie M, et al. An update on progress and the changing epidemiology of causes of childhood blindness worldwide. *J AAPOS*. 2012;16(6):501–507.
4. Chong C, McGhee CNJ, Dai SH. Causes of childhood low vision and blindness in New Zealand. *Clin Exp Ophthalmol*. 2019;47(2):165–170.
5. Whiting S, Jan JE, Wong PK, et al. Permanent cortical visual impairment in children. *Dev Med Child Neurol*. 1985;27(6):730–739.
6. Aldrich MS, Alessi AG, Beck RW, Gilman S. Cortical blindness: etiology, diagnosis, and prognosis. *Ann Neurol*. 1987;21(2):149–158.
7. Dutton G, Ballantyne J, Boyd G, et al. Cortical visual dysfunction in children: a clinical study. *Eye (Lond)*. 1996;10(Pt 3):302–309.
8. Good WV, Jan JE, DeSa L, et al. Cortical visual impairment in children. *Surv Ophthalmol*. 1994;38(4):351–364.
9. Chang MY, Borchert MS. Advances in the evaluation and management of cortical/cerebral visual impairment in children. *Surv Ophthalmol*. 2020;65(6):708–724.
10. Martin MB, Santos-Lozano A, Martin-Hernandez J, et al. Cerebral versus ocular visual impairment: the impact on developmental neuroplasticity. *Front Psychol*. 2016;7:1958.
11. Handa S, Saffari SE, Borchert M. Factors associated with lack of vision improvement in children with cortical visual impairment. *J Neuroophthalmol*. 2018;38(4):429–433.
12. Matsuba CA, Jan JE. Long-term outcome of children with cortical visual impairment. *Dev Med Child Neurol*. 2006;48(6):508–512.
13. Bennett CR, Bauer CM, Bailin ES, Merabet LB. Neuroplasticity in cerebral visual impairment (CVI): assessing functional vision and the neurophysiological correlates of dorsal stream dysfunction. *Neurosci Biobehav Rev*. 2019;108:171–181.
14. Jan JE, Groenveld M, Sykanda AM, Hoyt CS. Behavioural characteristics of children with permanent cortical visual impairment. *Dev Med Child Neurol*. 1987;29(5):571–576.

15. Kran BS, Lawrence L, Mayer DL, Heidary G. Cerebral/cortical visual impairment: a need to reassess current definitions of visual impairment and blindness. *Semin Pediatr Neurol*. 2019;31:25–29.
16. Good WV, Jan JE, Burden SK, et al. Recent advances in cortical visual impairment. *Dev Med Child Neurol*. 2001;43(1):56–60.
17. Chandna A, Ghahghaei S, Foster S, Kumar R. Higher visual function deficits in children with cerebral visual impairment and good visual acuity. *Front Hum Neurosci*. 2021;15:711873.
18. Sakki H, Dale NJ, Sargent J, et al. Is there consensus in defining childhood cerebral visual impairment? A systematic review of terminology and definitions. *Br J Ophthalmol*. 2018;102(4):424–432.
19. National Eye Institute. *NEI Strategic Plan: Vision for the Future, 2021-2025*. National Eye Institute; 2021.
20. Bennett CR, Bex PJ, Bauer CM, Merabet LB. The assessment of visual function and functional vision. *Semin Pediatr Neurol*. 2019;31:30–40.
21. Colenbrander A. Visual functions and functional vision. *International Congress Series*. 2005;1282:482–486.
22. Raviskanthan S, Wendt S, Ugoh PM, et al. Functional vision disorders in adults: a paradigm and nomenclature shift for ophthalmology. *Surv Ophthalmol*. 2022;67(1):8–18.
23. Sakki H, Dale NJ, Mankad K, et al. Exploratory investigation of brain MRI lesions according to whole sample and visual function subtyping in children with cerebral visual impairment. *Front Hum Neurosci*. 2021;15:765371.
24. Cioni G, Fazzi B, Ipata AE, et al. Correlation between cerebral visual impairment and magnetic resonance imaging in children with neonatal encephalopathy. *Dev Med Child Neurol*. 1996;38(2):120–132.
25. Eken P, de Vries LS, van Nieuwenhuizen O, et al. Early predictors of cerebral visual impairment in infants with cystic leukomalacia. *Neuropediatrics*. 1996;27(1):16–25.
26. Bauer CM, Papadelis C. Alterations in the structural and functional connectivity of the visuomotor network of children with periventricular leukomalacia. *Semin Pediatr Neurol*. 2019;31:48–56.
27. Jimenez-Gomez A, Fisher KS, Zhang KX, et al. Longitudinal neurological analysis of moderate and severe pediatric cerebral visual impairment. *Front Hum Neurosci*. 2022;16:772353.
28. Ortibus E, Lagae L, Casteels I, et al. Assessment of cerebral visual impairment with the L94 visual perceptual battery: clinical value and correlation with MRI findings. *Dev Med Child Neurol*. 2009;51(3):209–217.
29. Bauer CM, Merabet LB. Aberrant white matter development in cerebral visual impairment: a proposed mechanism for visual dysfunction following early brain injury. *J Integr Neurosci*. 2024;23(1):1.
30. Ortibus E, Verhoeven J, Sunaert S, et al. Integrity of the inferior longitudinal fasciculus and impaired object recognition in children: a diffusion tensor imaging study. *Dev Med Child Neurol*. 2012;54(1):38–43.
31. Pamir Z, Bauer CM, Bailin ES, et al. Neural correlates associated with impaired global motion perception in cerebral visual impairment (CVI). *Neuroimage Clin*. 2021;32:102821.
32. Huo R, Burden SK, Hoyt CS, Good WV. Chronic cortical visual impairment in children: aetiology, prognosis, and associated neurological deficits. *Br J Ophthalmol*. 1999;83(6):670–675.
33. Khetpal V, Donahue SP. Cortical visual impairment: etiology, associated findings, and prognosis in a tertiary care setting. *J AAPOS*. 2007;11(3):235–239.
34. West MR, Borchert MS, Chang MY. Ophthalmologic characteristics and outcomes of children with cortical visual impairment and cerebral palsy. *J AAPOS*. 2021;25(4):223e1–223e6.
35. Jacobson L, Hellstrom A, Flodmark O. Large cups in normal-sized optic discs: a variant of optic nerve hypoplasia in children with periventricular leukomalacia. *Arch Ophthalmol*. 1997;115(10):1263–1269.
36. Brodsky MC. Semiology of periventricular leucomalacia and its optic disc morphology. *Br J Ophthalmol*. 2003;87(11):1309–1310.
37. Mansukhani SA, Ho ML, Brodsky MC. Abnormal vestibular-ocular reflexes in children with cortical visual impairment. *J Neuroophthalmol*. 2021;41(4):531–536.
38. Kelly JP, Phillips JO, Saneto RP, et al. Cerebral visual impairment characterized by abnormal visual orienting behavior with preserved visual cortical activation. *Invest Ophthalmol Vis Sci*. 2021;62(6):15.
39. Salati R, Borgatti R, Giammari G, Jacobson L. Oculomotor dysfunction in cerebral visual impairment following perinatal hypoxia. *Dev Med Child Neurol*. 2002;44(8):542–550.
40. Mooney SWJ, Alam NM, Prusky GT. Tracking-based interactive assessment of saccades, pursuits, visual field, and contrast sensitivity in children with brain injury. *Front Hum Neurosci*. 2021;15:737409.
41. Rice ML, Sandoval MA, Castleberry KM, Schwartz TL. Physician prescribing and referral patterns in children with cerebral visual impairment. *Optom Vis Sci*. 2021;98(9):1078–1084.
42. Russell-Eggitt I, Harris CM, Kriss A. Delayed visual maturation: an update. *Dev Med Child Neurol*. 1998;40(2):130–136.
43. Tresidder J, Fielder AR, Nicholson J. Delayed visual maturation: ophthalmic and neurodevelopmental aspects. *Dev Med Child Neurol*. 1990;32(10):872–881.
44. Campos E. Amblyopia. *Surv Ophthalmol*. 1995;40(1):23–39.
45. Hoyt CS. Constenbader lecture. Delayed visual maturation: the apparently blind infant. *J AAPOS*. 2004;8(3):215–219.
46. Lambert SR, Kriss A, Taylor D. Delayed visual maturation. A longitudinal clinical and electrophysiological assessment. *Ophthalmology*. 1989;96(4):524–528; discussion 9.
47. Lloyd A, Hadfield M. Average recovery time from delayed visual maturation. *Int Ophthalmol*. 2021;41(6):2241–2247.
48. Merabet LB, Devaney KJ, Bauer CM, et al. Characterizing visual field deficits in cerebral/cortical visual impairment (CVI) using combined diffusion based imaging and functional retinotopic mapping: a case study. *Front Syst Neurosci*. 2016;10:13.
49. Good WV, Hou C, Norcia AM. Spatial contrast sensitivity vision loss in children with cortical visual impairment. *Invest Ophthalmol Vis Sci*. 2012;53(12):7730–7734.
50. Jan JE, Groenveld M, Sykanda AM. Light-gazing by visually impaired children. *Dev Med Child Neurol*. 1990;32(9):755–759.
51. Porro G, Dekker EM, Van Nieuwenhuizen O, et al. Visual behaviours of neurologically impaired children with cerebral visual impairment: an ethological study. *Br J Ophthalmol*. 1998;82(11):1231–1235.
52. Boyle NJ, Jones DH, Hamilton R, et al. Blindsight in children: does it exist and can it be used to help the child? Observations on a case series. *Dev Med Child Neurol*. 2005;47(10):699–702.
53. Cohen-Maitre SA, Haerich P. Visual attention to movement and color in children with cortical visual impairment. *J Vis Impair Blind*. 2005;99(7):2006.

54. Dutton GN. 'Dorsal stream dysfunction' and 'dorsal stream dysfunction plus': a potential classification for perceptual visual impairment in the context of cerebral visual impairment? *Dev Med Child Neurol*. 2009;51(3):170−172.
55. Bauer CM, Manley CE, Ravenscroft J, et al. Deficits in face recognition and consequent quality-of-life factors in individuals with cerebral visual impairment. *Vision (Basel)*. 2023;7(1):9.
56. Manley CE, Walter K, Micheletti S, et al. Object identification in cerebral visual impairment characterized by gaze behavior and image saliency analysis. *Brain Dev*. 2023;45(8):432−444.
57. Weinstein JM, Gilmore RO, Shaikh SM, et al. Defective motion processing in children with cerebral visual impairment due to periventricular white matter damage. *Dev Med Child Neurol*. 2012;54(7):e1−e8.
58. Chandna A, Nichiporuk N, Nicholas S, et al. Motion processing deficits in children with cerebral visual impairment and good visual acuity. *Invest Ophthalmol Vis Sci*. 2021;62(14):12.
59. Barton JJ. Higher cortical visual deficits. *Continuum (Minneap Minn)*. 2014;20(4 Neuro-ophthalmology):922−941.
60. Boonstra FN, Bosch DGM, Geldof CJA, et al. The multidisciplinary guidelines for diagnosis and referral in cerebral visual impairment. *Front Hum Neurosci*. 2022;16:727565.
61. Pilling RF, Allen L, Bowman R, et al. Clinical assessment, investigation, diagnosis and initial management of cerebral visual impairment: a consensus practice guide. *Eye (Lond)*. 2023;37(10):1958−1965.
62. Macintyre-Beon C, Young D, Calvert J, et al. Reliability of a question inventory for structured history taking in children with cerebral visual impairment. *Eye (Lond)*. 2012;26(10):1393.
63. Ben Itzhak N, Vancleef K, Franki I, et al. Visuoperceptual profiles of children using the Flemish cerebral visual impairment questionnaire. *Dev Med Child Neurol*. 2020;62(8):969−976.
64. Pehere NK, Dutton GN. Perceptual visual dysfunction in children—an Indian perspective. *Indian J Ophthalmol*. 2021;69(8):2004−2011.
65. McCulloch DL, Mackie RT, Dutton GN, et al. A visual skills inventory for children with neurological impairments. *Dev Med Child Neurol*. 2007;49(10):757−763.
66. Birch EE, Bane MC. Forced-choice preferential looking acuity of children with cortical visual impairment. *Dev Med Child Neurol*. 1991;33(8):722−729.
67. Raja S, Emadi BS, Gaier ED, et al. Evaluation of the relationship between preferential looking testing and visual evoked potentials as a biomarker of cerebral visual impairment. *Front Hum Neurosci*. 2021;15:769259.
68. Chang MY, Borchert MS. Comparison of eye tracking and Teller acuity cards for visual acuity assessment in pediatric cortical/cerebral visual impairment (CVI). *Am J Ophthalmol*. 2024;260:115−121.
69. Chang MY, Roman-Lantzy C, O'Neil SH, et al. Validity and reliability of CVI Range assessment for Clinical Research (CVI Range-CR): a longitudinal cohort study. *BMJ Open Ophthalmol*. 2022;7:e001144.
70. Vancleef K, Janssens E, Petre Y, et al. Assessment tool for visual perception deficits in cerebral visual impairment: reliability and validity. *Dev Med Child Neurol*. 2020;62(1):118−124.
71. McDowell N, Butler P. Validation of the Austin assessment: a screening tool for cerebral visual impairment related visual issues. *PLoS One*. 2023;18(11):e0293904.
72. Good WV. Development of a quantitative method to measure vision in children with chronic cortical visual impairment. *Trans Am Ophthalmol Soc*. 2001;99:253−269.
73. Hamilton R, Bach M, Heinrich SP, et al. VEP estimation of visual acuity: a systematic review. *Doc Ophthalmol*. 2021;142(1):25−74.
74. Chang MY, Borchert MS. Validity and reliability of eye tracking for visual acuity assessment in children with cortical visual impairment. *J AAPOS*. 2021;25(6):334e1−334e5.
75. Manley CE, Bennett CR, Merabet LB. Assessing higher-order visual processing in cerebral visual impairment using naturalistic virtual-reality-based visual search tasks. *Children (Basel)*. 2022;9(8):1114.
76. Kooiker MJG, Verbunt HJM, van der Steen J, Pel JJM. Combining visual sensory functions and visuospatial orienting functions in children with visual pathology: a longitudinal study. *Brain Dev*. 2019;41(2):135−149.
77. Chokron S, Kovarski K, Dutton GN. Cortical visual impairments and learning disabilities. *Front Hum Neurosci*. 2021;15:713316.
78. American Psychiatric Association. *Diagnostic and Statistical Manual of Mental Disorders*. Fifth Edition. American Psychiatric Publishing, Inc; 2022. Text Revision (DSM-5-TR).
79. Lyon GR, Shaywitz SE, Shaywitz BA. A definition of dyslexia. *Ann Dyslexia*. 2003;53:1−14.
80. Peng P, Zhang Z, Wang W, et al. A meta-analytic review of cognition and reading difficulties: individual differences, moderation, and language mediation mechanisms. *Psychol Bull*. 2022;148(3-4):227−272.
81. Williams ME, Fink C, Zamora I, Borchert M. Autism assessment in children with optic nerve hypoplasia and other vision impairments. *Dev Med Child Neurol*. 2014;56(1):66−72.
82. Philip SS, Dutton GN. Identifying and characterising cerebral visual impairment in children: a review. *Clin Exp Optom*. 2014;97(3):196−208.
83. Lehman SS, Yin L, Chang MY, et al. Diagnosis and care of children with cortical visual impairment: clinical report. Pediatrics. Forthcoming.
84. Braddick O, Atkinson J, Wattam-Bell J. Normal and anomalous development of visual motion processing: motion coherence and 'dorsal-stream vulnerability.'. *Neuropsychologia*. 2003;41(13):1769−1784.
85. Braddick O, Atkinson J. Development of human visual function. *Vision Res*. 2011;51(13):1588−1609.
86. Connell N, Merabet LB. Uncovering the connectivity of the brain in relation to novel vision rehabilitation strategies. *Neurology*. 2014;83(6):484−485.
87. Voss P. Sensitive and critical periods in visual sensory deprivation. *Front Psychol*. 2013;4:664.
88. Merabet LB, Mayer DL, Bauer CM, et al. Disentangling how the brain is "wired" in cortical (cerebral) visual impairment. *Semin Pediatr Neurol*. 2017;24(2):83−91.
89. Fazzi E, Micheletti S, Calza S, et al. Early visual training and environmental adaptation for infants with visual impairment. *Dev Med Child Neurol*. 2021;63(10):1180−1193.
90. Robertson AO, Tadic V, Rahi JS. Transition from paediatric to adult ophthalmology services: what matters most to young people with visual impairment. *Eye (Lond)*. 2018;32(2):406−414.
91. White PH, Cooley WC, Transitions Clinical Report Authoring Group, et al. Supporting the health care transition from adolescence to adulthood in the medical home. *Pediatrics*. 2018;142(5):e20182587.
92. Lueck A, Harpster K, Hartmann E, et al. Toward removing barriers in the evaluation, diagnosis, and care of individuals with cerebral visual impairments. *Policy Insights Behav Brain Sci*. 2023;10(2):296−307.