

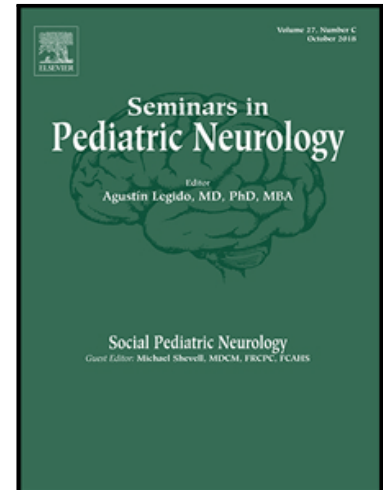
Accepted Manuscript

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PII: S1071-9091(19)30041-5
DOI: <https://doi.org/10.1016/j.spen.2019.05.005>
Reference: YSPEN 760

To appear in: *Seminars in Pediatric Neurology*



Please cite this article as: Barry S. Kran OD, FAAO , Linda Lawrence MD , D. Luisa Mayer PhD , Gena Heidary MD, PhD , Cerebral/Cortical Visual Impairment: A Need to Reassess Current Definitions of Visual Impairment and Blindness, *Seminars in Pediatric Neurology* (2019), doi: <https://doi.org/10.1016/j.spen.2019.05.005>

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Cerebral/Cortical Visual Impairment: A Need to Reassess Current Definitions of Visual Impairment and Blindness

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ABSTRACT

Cerebral/cortical visual impairment (CVI) is characterized by higher order visual dysfunction caused by injury to the retrogeniculate visual pathways and brain structures which sub-serve visual processing. CVI has become the leading cause of significant vision loss in children in developed countries, but continues to be an underrecognized cause of visual disability with respect to services aimed at maximizing visual development. Current criteria which are used to define visual disability rely on measures of visual acuity and visual field. Many children who require specialized vision services do not qualify, because these standard definitions of vision impairment do not account for CVI. In order to appropriately identify patients with CVI and offer the resources which may positively impact functional use of vision, the definition of visual impairment and blindness needs to be modified. This commentary calls for a change in the definition of visual impairment and blindness to acknowledge those persons with brain-based vision impairment.

“The definition of a word is fixed by usage, Blindness has been so widely used by people who had no exact knowledge of what they meant by it that it cannot be restricted to a sharply defined impairment to vision.”¹

What one “sees” begins with visual processing in the eyes that continues along the anterior visual pathway, lateral geniculate nucleus, retrogeniculate visual pathways, primary visual cortex, and vision association areas. An individual’s view of the world is further influenced by other sensory inputs, environment, experience and attention. Given that all these resources are utilized to create visual impressions, it is no longer sufficient to define vision impairment and blindness solely based on one’s ability to see detail (acuity) or on a diameter of a visual field. The purpose of this commentary is to examine current definitions of visual impairment and highlight the need to incorporate a more encompassing rubric which takes into account the diagnosis of cerebral/cortical visual impairment (CVI).

Current definitions of blindness and visual impairment are used to classify the level of visual dysfunction on the basis of visual acuity and visual field. Other vision function findings are also important but are not part of most national or international descriptions of vision impairment and blindness. This categorization is used to determine inclusion or exclusion for various important vision services (e.g., school-based educational plans, vocational training, mobility or aid with activities related to daily living); this is also used to determine eligibility for tasks such as driving and disability benefits. It is important to note that visual disability can be considered relative

to a particular task and profession. Regarding driving, in states such as Massachusetts, there exists a stratified approach to levels of licensure based upon acuity and field. For example, so long as best corrected distance acuity is not worse than 20/40, there is no special restriction on licensure.²

The problem clinicians are confronted with is that current categorizations of visual impairment for entitlement to services based upon visual acuity and visual field-based do not account for visual impairment associated with children who have CVI. That is, many children with CVI would not qualify as legally blind under the current definitions, but their level of visual dysfunction is poorer than what would be predicted on the basis of visual acuity or visual field. Some children may even have normal visual acuity, but have substantive issues with visual processing.^{3,4} Since CVI is the leading cause of pediatric vision impairment in the developed world and many developing countries, and may be associated with deficits in motor function (cerebral palsy or CP), cognition and/or sensory processing, the identification of children with CVI is critical in order to provide timely and appropriate assessment and intervention to lessen the impact of the disability.^{5,6,7}

“Cortical blindness” is the only ICD code to describe CVI.⁸ Cortical blindness was first used by Gordon Holmes in 1918 to describe visual impairment of soldiers who were blind secondary to injuries to the occipital cortex.⁹ In 2003, Hoyt noted that cortical blindness is “bilateral loss of vision with normal pupillary responses and an eye examination, which shows no abnormalities” but that this does not reflect the

constellation of patients we see with brain-based vision impairment. Further, Dr. Hoyt writes that “there is an obvious need for the establishment of an international classification of neurologic visual disorders.”¹⁰

Since 2015, the American Association of Pediatric Ophthalmology and Strabismus (AAPOS) has noted on its website that one can have both a brain-based vision impairment and an impairment in the anterior visual pathway. They, like many in North America, continue to use the term “cortical” rather than “cerebral” which is the predominant term elsewhere to describe this brain-based vision impairment.^{11,12} Per AAPOS, CVI “is a decreased visual response due to a neurological problem affecting the visual part of the brain. Typically, a child with CVI has a normal eye exam or has an eye condition that cannot account for the abnormal visual behavior. It is one of the most frequent causes of visual impairment in children from developed countries.”¹¹

A UK-based team led by Sakki et al. in 2017 published a systematic literature review of original articles focused on CVI in an effort to develop an internationally accepted definition of CVI.¹² From their work, the authors propose the following definition: “Childhood cerebral impairment is a verifiable visual dysfunction which cannot be attributed to disorders of the anterior visual pathways or any potentially co-occurring ocular impairment.”

Gordon Dutton and Amanda Hall-Lueck described CVI as fully and as functionally as the condition can be understood at this time. “Vision impairment due to damage or

disorder of the visual pathways and visual centers in the brain, including the pathways serving visual perception, cognition, and visual guidance of movement.”¹³

The World Health Organization (WHO) defines visual impairment and blindness as follows: “Blindness” is defined as a presenting visual acuity of worse than 3/60 (20/400) or a corresponding visual field loss to less than 10° in the better eye. “Severe visual impairment” is defined as a presenting visual acuity of worse than 6/60 (20/200) and equal to or better than 3/60 (20/400). “Moderate visual impairment” is defined as a presenting visual acuity in the range from worse than 6/18 (20/63) to 6/60 (20/200) WHO considers the term “visual impairment” to include moderate and severe visual impairment as well as blindness.”¹⁴

For each level of vision impairment, Colenbrander has provided examples of changes in how a person functions and participates in society.¹⁵ The International Congress of Ophthalmology (ICO) 2002 document takes the position that that vision impairment and blindness “should be based on function rather than on visual acuities alone.”¹⁵

Vision function tests measure the threshold of a specific visual ability such as visual acuity, contrast sensitivity, or extent of the visual field. Functional vision is how the person uses their visual abilities to interpret visual information and react to that visual information for example, by attending to that information or guiding movements of the body based on that information.¹⁶ Can the child’s use of vision for a specific task be

both meaningful and sustainable?¹⁷ There are assessment tools/inventories which are available to evaluate the level of functional vision. These are most commonly administered in educational or community-based settings, rather than during a routine eye examination.^{18,19} Although a typical eye examination does not formally focus on functional vision, the clinician may still get insight into the impairment of functional vision through modifications, which are needed to perform the ophthalmological evaluation successfully. For example, a child with CVI may not perform optimally on visual acuity testing if there are environmental distractions such as a fully illuminated room or auditory distractions from a conversation between the eye doctor and parent. The examiner may notice that a modification of the environment by changing illumination to focus only on the testing materials (such as Teller acuity cards) in a quiet room will improve the visual acuity measurement. The examiner observes that as the environment is simplified, visual acuity, the patient's ability to sustain visual attention, and the latency to respond all improve. This example shows that altering the environment may change both vision function (threshold) and use of vision (improved attention to the task, reduction in time to reach threshold and in latency of response). In this way, although functional vision is not being formally assessed, the eye care provider may gain insight into the impact of CVI on how the patient is utilizing his or her visual potential.

There are examples where functional vision and vision function are relatively independent of each other. We assess patients with relatively good visual acuity, contrast sensitivity, and visual field who nevertheless function visually at a much lower

level than the vision function data would indicate. These patients may not be able to recognize faces or familiar objects and have difficulty with visually guided movement; some may not accurately process motion of things around them, nor manage moving through space, especially in environments that are novel or complex.

In our eye care practices, these individuals are diagnosed with CVI but it is difficult to translate our findings and history into effective advocacy since they do not meet the traditional definition of visual impairment/blindness. Unless a child is categorized as visually impaired or blind, the US educational system does not have the legal obligation to provide services that may include services from a teacher of students with vision impairment and/or an orientation and mobility specialist, to this growing number of students.²⁰ Denying or withholding vision services will have significant impact for the child, family and to our society as a whole.

The range and types of visual impairment (and other impairments) in individuals with CVI are diverse. There may or may not be motor dysfunction (such as cerebral palsy, but also including speech), some level of intellectual disability or developmental delay, and other sensory impairments (i.e., auditory) or visual processing issues. Prototypical examples of low, moderate, and high functioning individuals with CVI are described by Dutton and Lueck as well as on the website, cviscotland.org.²¹ Individuals with good vision function but poor functional vision who are passing their grades academically with significant effort may not qualify for vision services. Poor functional vision findings might include inaccurate visually guided reach or movement in space

which can be worse in visually complex or novel settings. This can result in an inability to travel safely such as when navigating a street crossing or finding the way to a new location in a school building or neighborhood. In children with some combination of these findings, when testable, various visual perceptual and visual motor tests may be well below average. While some self-taught compensatory skills may occur “organically”, it is more likely that children with these difficulties would habilitate more efficiently and quickly with services from individuals trained to provide habilitation services. In many instances, the education team should include vision educators (teacher of visual impairments, orientation and mobility instructor) as well as occupational therapists, physical therapists, speech and language therapists and assistive technology specialists whose collaboration with the student, teachers and parents will yield positive progress towards maximizing the child’s potential. The impact on the child by not providing these services includes potential delays in overall development, limited progress in developing visual skills, poor self-esteem, lack of self-confidence, and can give rise to behavioral problems.²⁰

The eventual cost to society later in life will far outweigh the cost of appropriate services provided earlier in the lives of the persons with CVI. For example, consider a child with ocular blindness who was doing well in school and lost her sight late in elementary school. If the student was not offered Braille despite having adequate tactile skills until just before aging out of the educational system, this child would be functionally illiterate and unable to progress in college or live as an independent adult. Another example would be if a student was not offered instruction in appropriate technological accommodations despite having the skills to use them. This student would

be technologically illiterate and like the Braille case described above, might not reach their educational potential, live independently, or contribute to society. Providing services when deficits are noted early in life can yield a significant return on investment. Once the child has plateaued with appropriate services, the need for or level of intensity of services (social, medical, other) for the remainder of the person's life would be either dramatically reduced or not necessary.

Several studies looking at preschool school and early childhood education utilizing rigorous research designs, reveal that those who received additional services had statistically better educational outcomes than those who did not.^{23, 24} Further, one study showed that the treatment group obtained significantly higher earnings than the control group. They determined that each dollar invested for services early in life yielded approximately \$13.00 in return.²³

Since 1879, when the US Congress passed the Act to Promote the Education of the Blind, the American Printing House for the Blind (APH), through funds allocated by congress, has provided materials to qualified students. How students qualified has changed over the years. The latest change occurred in 2010 when APH expanded eligibility to from those who Meet the Definition of Blindness (MDB) to those who either MBD or Function at the Definition of Blindness (FDB). This was done to more accurately reflect the true group of individuals for whom services are needed but were unable to respond to traditional methods of collecting acuity. These two categories are defined as follows:

- Meets the *Definition of Blindness*: Central visual acuity of 20/200 or less in the better eye with correction or a visual field diameter no greater than 20 degrees.
Or
- *Function at the Definition of Blindness*: Visual performance reduced by brain injury or dysfunction when visual function meets the definition of blindness as determined by an eye care specialist or neurologist.^{25,26}

For the last census available (January 2, 2017), 69.18% of the registrants to APH met the definition of legal blindness and 30.19% qualified as functioning at the definition of legal blindness. From a simplistic view, this implies that 30% fewer children would have been identified and served by vision educators if this alternative qualifier did not exist.²⁵

The US Department of Education, Office of Special Education and Rehabilitative Services Memo dated May 22, 2017 (OSEP 17-05), clarifies issues around state determination of eligibility for vision services relative to being in compliance with the Individuals with Disabilities Education Act (IDEA).²⁷ They note that “...*any* impairment in vision, regardless of significance or severity, must be included in a State’s definition, provided that such impairment, even with correction, adversely affects a child’s educational performance.” However, some states had been using a two-step eligibility process. The first step is to determine if the child had one of the conditions they determined could “...affect a child’s vision functioning” such as a specific constriction of visual field or acuity level. The second step is to then determine if that condition

“...adversely affects the child’s educational performance.” This memo makes it clear that the first step of this process is “inconsistent with IDEA” as one could be eliminated from consideration for services if they had a condition that was not on the state list.²⁷ This memo has been helpful for our children with CVI, but has not been sufficient to ensure needed services.

According to Goodrich, the US Department of Veteran Affairs realized that they were not meeting the needs of veterans of the conflicts in Afghanistan and Iraq with respect to visual sequela secondary to traumatic brain injuries.²⁸ Although these veterans did not qualify for vision rehabilitation services because they did not meet legal blindness criteria, they nevertheless had significant loss in functional use of vision. In 2008, the VA amended eligibility requirements by utilizing the concept of “excess disability”. “This term refers to vision loss that has a substantial impact on the individual’s functional independence that is out of proportion to the degree of impairment as measured by visual acuity or visual field (Department of Veteran Affairs, 2008).”

There are additional avenues towards identifying children with CVI. As Lena Jacobsen states in a commentary in 2014, “The time has come to re-evaluate the diagnostic system in which some functional diagnoses due to brain damage are currently recognized and given an ICD code, whereas others, such as cerebral visual and cerebral hearing impairment, are not included. Maybe a new approach would be better: a diagnostic code for cerebral dysfunction with sub-codes for each of the specific

conditions that are impaired.”²⁹ This would include codes for motor dysfunction (CP), CVI, and central auditory processing dysfunction. While these codes could be helpful and would acknowledge the predominant source of impaired function, the connection to needed services is not part of the solution. Significant changes to educational law would be required linking these conditions to the need for assessment and intervention.

Since 2001, the International Classification of Functioning, Disability and Health (ICF) is a companion set of codes to the World Health Organization’s International Classification of Diseases.³⁰ The ICF provides a standardization of assessment and description of functioning of an individual. These data are then used to assign health services, intervention and the management of other services and interventions. Such data sets could also be used for outcome assessments. The full ICF code book is difficult to utilize so various core code sets (a specific set of codes from the complete set that have been culled for a specific disability) have been developed over the years. Thus, a second alternative to acknowledging the disability caused by CVI is to allow for identification through the utilization of a vision core set under the international core set rubric. However, at the present time, a validated core set for children with vision impairment/blindness is not available.³¹

There is an understandable need to balance quantitative and qualitative data when considering a diagnosis and, in fact, a hallmark of major clinical treatment trials over the last 20 years has been to include both as primary and secondary outcome measures. The time has come to translate this to clinical practice and to the delivery of subsequent

necessary services. The fact that all visual disability codes were removed from a previous draft proposal for ICD 11 highlights that the committee is not ready to move in this direction at this time. Once an ICF vision disability core set is validated, it should be embraced by not only by the medical community, but by those setting policy regarding services such as in the areas of rehabilitation and education. Unfortunately for patients with CVI, eye care providers cannot wait until such a system is in place. It is incumbent upon us to identify and treat all children who are functioning with some level of vision impairment/blindness in spite of vision function data suggesting otherwise. Thus, the approach taken by AFB (now APH) and by the US Department of Veteran Affairs to broaden how individuals could qualify for services is a model that should be employed both nationally and internationally. Doing so will have the impact of including not only children with brain damage affecting the visual system, but older children and adults with visual symptoms secondary to traumatic brain injury. To effect change on a national or state level, it will be necessary for a cohort of advocacy groups (medical, education, service delivery providers, parent groups, appropriate non-profits, etc.) to lobby for changes to legislation to both allow for appropriate diagnosis of these children as well as to then assure provision of habilitative services as early as possible. For example, on a state level the appropriate coalition of advocacy groups could work with their state commissions for the blind to adjust the definition of blindness to include language similar to that used by AFB (now APH) in the “functions at the definition of blindness category.”

In conclusion, CVI is increasingly being recognized as the leading cause of visual impairment in children of developed countries. In order to appropriately identify patients with CVI and offer the resources which may positively impact visual development and visual function, the definition of visual impairment and blindness needs to be modified. The time has come to change the definition of visual impairment and blindness to acknowledge those persons with brain-based vision impairment who function at the level of visual impairment or blindness independent of visual acuity and visual field.

Disclosure of interests:

D. Luisa Mayer, PhD is a Designated Advisor for the Teller Acuity Cards® with the University of Washington. The other authors have no commercial, proprietary, or financial interest in any products or companies described in this article.

No other authors have any commercial, proprietary, or financial interest in any products or companies described in this article.

REFERENCES

1. Jackson E, Snell AC, Gradle HS. REPORT OF THE COMMITTEE ON DEFINITION OF BLINDNESS. JAMA. 103:1445-1446; 1934.
2. Medical standards for passenger (Class D) and motorcycle (Class M) driver's licenses <https://www.mass.gov/info-details/medical-standards-for-passenger-class-d-and-motorcycle-class-m-drivers-licenses> accessed August 22, 2018.
3. Hyvarinen L. Considerations in evaluation and treatment of the child with low vision. Am J Occup Ther. 49:891-897;1995.
4. Saidkasimova S, Bennett DM, Butler S, Dutton GN. Cognitive visual impairment with good visual acuity in children with posterior periventricular white matter injury: a series of 7 cases. J AAPOS;11:426-30; 2007. Epub 2007 Jul 16
5. Flanagan NM, Jackson AJ, Hill AE. Visual impairment in childhood: insights from a community-based survey. Child Care Health Dev. 29:493-9; 2003.
6. Rahi JS, Cable N. British Childhood Visual Impairment Study G. Severe visual impairment and blindness in children in the UK. Lancet 362:1359-65; 2003.
7. Kong L, Fry M, Al-Samarraie M, Gilbert C, Steinkuller PG. An update on progress and the changing epidemiology of causes of childhood blindness worldwide. J AAPOS. Dec;16(6):501-7; 2012. D oi: 10.1016/j.jaapos.2012.09.004
8. Centers for Medicare and Medicaid Services. CMS.gov <https://www.cms.gov/Medicare/Coding/ICD10/2019-ICD-10-CM.html> accessed August 23, 2018.

9. Holmes G. Disturbances of Vision by Cerebral Lesions. *Br J Ophthalmol* 2:353-84; 1918.
10. Hoyt CS. Visual Function in the brain damaged child. *Eye* 369–384; 2003.
11. AAPOS patient information on Cortical Visual Impairment available at: <http://www.aapos.org/terms/conditions/40> accessed July 26, 2018.
12. Sakki HEA, Dale NJ, Sargent J, Perez-Roche T, Bowman R1. Is there consensus in defining childhood cerebral visual impairment? A systematic review of terminology and definitions. *Br J Ophthalmol*. 102:424-432; 2018. doi: 10.1136/bjophthalmol-2017-310694. Epub 2017 Nov 16.
13. Dutton DN, Lueck AH. Impairment of Vision due to Damage to the Brain. In Dutton GN, Lueck AH, editors. *Impairment of vision due to damage to the brain in Vision and the Brain 2015* AFB Press p. 4.
14. Universal eye health: A global action plan 2014-2019. Available at: http://www.who.int/blindness/AP2014_19_English.pdf?ua=1 Accessed June, 10 2018.
15. Report prepared for the International Council of Ophthalmology at the 29th International Congress of Ophthalmology Sydney, Australia, April 2002 p.32.
16. Colenbrander A. Towards the development of a classification of vision-related functioning- a potential framework in Dutton GN, Bax M, editors *Clinics in Developmental Medicine* No. 186 *Visual impairment in children due to damage to the brain*. Mac Keith Press 2010. p.284.
17. Colenbrander A. Towards the development of a classification of vision-related functioning- a potential framework. In Dutton GN, Bax M editors. *Clinics in*

- Developmental Medicine No. 186 Visual impairment in children due to damage to the brain. Mac Keith Press 2010 p 286.
18. Teach CVI available at: <https://www.teachcvi.net/screening-tools>, accessed June 10, 2018
19. Roman-Lanzky C. Cortical Visual Impairment: An approach to assessment and intervention, 2nd ed. AFB Press 2018.
20. IDEA Sec 300.8 Child with a disability Available at: <https://sites.ed.gov/idea/reg/b/a/300.8> accessed July 17, 2018.
21. Lueck AH, Gordon GN Assessment of Children with CVI: Introduction and Overview appendix in Dutton GN, Lueck AH, editors Vision and the Brain AFB Press 2015 p 207-260.
22. AAPOS, Importance of Vision available at: [https://aapos.org/client_data/files/2018/1662_importance of vision.pdf](https://aapos.org/client_data/files/2018/1662_importance_of_vision.pdf). accessed 9 July 2018
23. Belfield CR, Nores M, Barnett S, Schweinhart L. The High/Scope Perry Preschool Program cost-benefit analysis using data from the age 40 followup. J Hum Resour 41:162-90; 2006.
24. Campbell FA, Ramey CT, Pungello EP, Sparling JJ, Miller-Johnson S. Early Childhood Education: Young Adult Outcomes from the Abecedarian Project. Appl Dev Sci. 6:42-57; 2002.
25. Personal communication with Janie Blome, Director of Field Services APH July 19, 2018

26. What is Federal Quota available at: <https://www.aph.org/federal-quota/what-is-federal-quota/> accessed June 8, 2018
27. OSEP memo 17-05 available at: <https://sites.ed.gov/idea/files/letter-on-visual-impairment-5-22-17.pdf>. Accessed July 14, 2018
28. Goodrich GL. The evolution of the definition of legal blindness. In Lueck AH and Dutton GN editors. Vision and the Brain 2015 AFB Press p. 7-8.
29. Jacobsen L. Cerebral dysfunction in children: should this be the central tenet for a new system of classification? *Develop Med & child Neurol* 56:101-102; 2014. doi: 10.1111/dmcn:12328
30. Bickenbach J, Cieza A, Rauch A, Stucki, G Editors. ICF Core Sets: Manual for Clinical Practice. Hogrefe Publishing 2002.
31. Yazar F, Cavlak., Basakci Calik B. Applying the International Classification of Functioning, Disability, and Health in children with low vision: difference between raters. *Turk J Med Sci* 46:1694-1699; 2016. doi10.3906/sag-1506-152