Statement on Cortical Visual Impairment

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In August 2008, the American Printing House for the Blind (APH) brought together an advisory group to provide guidance and clarity on a range of issues related to cortical visual impairment (CVI) as those issues relate to the development of products. The nationally drawn CVI Advisory Group, with members suggested by APH ex officio trustees and consultants, represents a continuing effort by APH to serve the growing group of students with CVI who are registered for Federal Quota funds. Previous activities conducted by APH on behalf of these students have included improving the Federal Quota registration process by better defining eligibility criteria; hosting a CVI Synergy meeting in Louisville, Kentucky; identifying existing APH products that serve the unique needs of this population; developing and manufacturing new products for students with CVI; creating a CVI web site; and providing information on APH products that are related to students with CVI through workshops of the APH National Instructional Partnership.

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Definition of CVI: A working definition for educational services

CVI is defined as impaired vision that is due to bilateral dysfunction of the optic radiations or visual cortex or both. It can coexist with ocular and ocular motor disorders and can be the result of perinatal brain dysfunction or be caused by trauma. Approximately 30%-40% of children with visual impairments have CVI (see Figure 1).

One concern of professionals in the field of education of students with visual impairments is to establish a standard definition of CVI. Accordingly, the purpose of this article is to clarify the differences between children who qualify for services from vision educators and those who have visual processing difficulties that are not considered visual impairment. Our perspective is that
all children who have CVI should be classified as visually impaired and receive the necessary services, regardless of the severity of the degree of CVI or additional disabilities. A child with CVI is distinguished from a child with learning disabilities or developmental disabilities by the following criteria: (1) an eye examination that cannot fully explain the child's use of vision; (2) a history or presence of neurological problems, even when the child's brain-imaging studies may appear normal (Dutton, 2008); and (3) the presence of the behavioral or visual responses that are collectively associated with CVI.

In most North American jurisdictions, low vision is defined as a reduction in visual acuity no better than 20/70 (6/21) but better than 20/200 (6/60) in the better eye with the best correction. Legal blindness is defined as visual acuity no better than 20/200 (6/60) in the better eye with the best correction. Legal blindness is also defined as a central visual field that is no greater than 20 degrees. Using this framework, CVI should be defined, albeit arbitrarily, by a reduction in visual acuity, in the visual fields, or in a child's ability to see compared to other children of the same age. Unfortunately, traditional methods of precisely determining acuity or visual field function in children with CVI are problematic.

Because children with CVI frequently have additional disabilities, it is often difficult to measure visual acuity. When it is possible to do so, standard visual acuity testing should be performed. Electrophysiological measures, such as visual evoked potential acuities, may also be used. When warranted, visual acuities should be measured using forced-choice preferential looking acuities or estimating visual function through the identification of sized objects at specific distances.

Dutton (2008) and others have proposed a theoretical construct for classifying higher-level visual processing. This framework considers the effects of damage to the dorsal and ventral streams of the brain to explain visual dysfunction. Although there are several explanatory models to identify and describe CVI (Colenbrander, 2009; Hyvarinen, 2005; Morse, 1990), it is becoming increasingly clear that the effects of CVI fall into a spectrum from mild to severe.

Recently, the term cortical visual impairment has been used interchangeably with the term cerebral visual impairment. For some (Hyvarinen, 2005), the term cortical visual impairment is limiting because cortical visual impairment does not describe the full extent of visual dysfunction caused by damage in areas other than the visual cortex. We believe that children with perceptual and cognitive dysfunction may have cerebral processing problems that do not represent visual impairment; cortical visual impairment, on the other hand, is a cortical disorder
that conforms to a unique set of visual and behavioral characteristics. That is, cortical visual impairment should be considered a subset of the more broadly defined term, cerebral visual impairment (see Figure 1).

In addition to a reduction in acuity or in the ability to see, children who are diagnosed with CVI also demonstrate the phenotypic characteristics of CVI, as defined in the literature (Jan, Groenveld, & Sykanda, 1990). Children with CVI have unique visual features, including light gazing (Jan et al., 1990); photophobia (Jan, Groenveld, & Anderson, 1993); poor visual attention (Jan, Groenveld, Sykanda, & Hoyt, 1987); color preference (Groenveld, 1993; Roman-Lantzy, 2007); restricted visual fields (Jan et al., 1993); difficulties discriminating or interpreting complex visual patterns, arrays, and scenes (Dutton, McKillop, & Saidkasimova, 2006; Roman-Lantzy, 2007); difficulties finding an object at a distance (Dutton et al., 2006; Roman-Lantzy, 2007); better recognition of familiar objects than novel ones (Jan et al., 1993; Roman-Lantzy, 2007); attention to moving objects (Jan et al., 1990; Roman-Lantzy, 2007); looking away when reaching (Groenveld, 1993; Dutton et al., 1996; Roman-Lantzy, 2007); visual latency (Roman-Lantzy, 2007); atypical visual reflexive responses (Roman-Lantzy, 2007); and variability in contrast (Good, 2001). A combination of these specific visual and behavioral features is needed to confirm a diagnosis of CVI. A trained clinician or educator should be able to identify these features systematically through interviews, observations, and direct assessments of the child (Roman-Lantzy, 2007). In summary, the definition of CVI must have several key components. First, the child must have reduced visual acuity or difficulty seeing compared to other children of the same age. Second, the child must have some brain injury, a suspected brain injury, or a malformation that causes dysfunction of the optic radiations or visual cortex or both. Finally, children must demonstrate the collection of visual behaviors that have been identified in the literature as characteristic of children with CVI. Beyond these criteria, other diagnostic features should also be considered. For example, comorbid conditions include cognitive impairment, cerebral palsy, hearing loss, and seizures (Huo, Burden, Hoyt, & Good, 1999; Matsuba & Jan, 2006). These conditions should be considered because they play a role in intervention and rehabilitation. Note, however, that the absence of some or all comorbid conditions does not exclude the diagnosis of CVI.

As with ocular visual impairments, the severity of CVI should be considered across a spectrum (Roman-Lantzy, 2007). The visual acuities, characteristic features, and comorbid conditions can vary within the population. In addition, some variation should be expected within the same individual, since fluctuations in environment, health, and rehabilitation occur. Fortunately, nearly all children with CVI will demonstrate improved vision, and many of the clinical characteristics
will be resolved; however, most children will continue to meet the definition of CVI and thus continue to qualify for vision support services (Baker-Nobles & Rutherford, 1995; Groenendaal & Hof-van Duin, 1992; Huo et al., 1999; Matsuba & Jan, 2006; Roman-Lantzy, 2007).

Clarifying the definition of CVI is important. Use of a clear and consistent definition will not only help APH, but will guide researchers, clinicians, and families in the development of appropriate educational and interventional strategies.

Screening of children with CVI

As we noted previously, children with CVI are a heterogeneous population with a spectrum of visual abilities. One child with CVI may have limited responses and skills, such as responding only to light sources or a bright red toy, whereas another child with CVI may demonstrate visual difficulty only in complex environments or with novel or visually cluttered materials. Milder forms of CVI are often not detected because visual difficulty is attributed to such factors as communication-language or motor delays. Therefore, vision educators must identify all children who are at risk for CVI on the basis of key etiologies and conditions that are known to result in damage to the visual pathways and visual processing areas of the brain. Specifically, CVI is most prevalent in the following etiologies or conditions: asphyxia and perinatal hypoxic-ischemic encephalopathy, cerebral vascular accident, periventricular leukomalacia, infection, structural abnormalities, chromosomal abnormalities, metabolic conditions, and traumatic brain injury. Therefore, all children who present with one or more of these conditions should be screened for CVI. Ideally, medical personnel in neonatal intensive care units or pediatric intensive care units should implement a screening procedure to detect infants or children who demonstrate the behaviors associated with CVI and then refer them to appropriate intervention programs that include a teacher of students with visual impairments. Personnel in early intervention or educational programs should refer a child to a teacher of students with visual impairments if the child has one or more of these conditions even if the infant, child, or adolescent does not yet have a diagnosis of CVI. Screening can be accomplished by asking key questions of parents and caregivers (Roman-Lantzy, 2007). If a child has a medical history that is consistent with the diagnosis of CVI and demonstrates visual behaviors that are consistent with the diagnosis of CVI, he or she should then be referred for more in-depth testing.

Medical assessment of children with CVI

Vision educators can help guide families as they navigate through the entire evaluation process. A pediatric ophthalmologist or pediatric optometrist who has experience in evaluating children
with CVI should evaluate all at-risk children. Ophthalmologists or optometrists play a key role in ruling out anterior pathway diseases, which can coexist with CVI and require thorough ophthalmologic management. Additional evaluations, such as imaging, electrophysiological studies, and laboratory investigations, are often performed. These evaluations can be used to confirm abnormalities to the posterior pathway, particularly when the history is less suggestive. It should be noted that specific evaluations are dependent upon the clinical presentation and are ordered at the discretion of the physician. The child's pediatrician, neurologist, or ophthalmologist must determine the course of investigation. The following key areas are included in a comprehensive ophthalmologic examination:

- Review of the child's birth history, maternal history, and overall medical history
- Review of all existing neurological records
- The child's overall physical appearance
- Evaluation of eye health
- Evaluation for refractive error
- Assessment of ocular pathology
- Visual acuity measures (nontraditional methods may be used when certain color resolutions or preferences are present)
- Visual fields
- Eye movements
- Motility or binocular vision
- Color vision
- Contrast sensitivity

With medical information from the ophthalmologist or optometrist, the team will be able to provide intervention services and address potential coexisting ocular conditions. In addition to medical assessments, the intervention team needs to understand the child's functional use of vision in living and learning environments. Therefore, a functional vision assessment is a priority.

Functional vision assessment of children with CVI

Program planning and adjustments in the environment begin with a systematic assessment of a child's CVI status on the basis of a functional vision assessment of the child's unique visual and behavioral characteristics (Roman-Lantzy, 2007). The teacher of students with visual impairments is responsible for conducting a thorough assessment that synthesizes recent
medical information, data from interviews, and findings from direct assessments. If the child has been referred for vision services and has not had an ophthalmologic examination within the past year, referral to a pediatric ophthalmologist is recommended.

The teacher of students with visual impairments should lead the functional vision assessment and collaborate with the primary team members (such as the parents, teachers, occupational therapist, physical therapist, speech therapist, orientation and mobility specialist, and deaf-blind specialist). While other personnel may have training and experience in CVI, it is ultimately the responsibility of the teacher of students with visual impairments to lead, complete, and interpret all the findings of the assessment. We suggest that traditional tools and methods of a functional vision assessment do not adequately assess key visual characteristics that are indicative of CVI. For children with CVI, a functional vision assessment that is conducted by the teacher of students with visual impairments should be grounded in the characteristics of CVI and should include the following:

- A review of the child's medical and visual history (from physicians’ reports)
- Face-to-face interviews with the parent, teacher, and child (if applicable)
- Direct observations in the classroom, community, and natural environments
- Direct assessment of visual behaviors related to the characteristics of CVI: color preference, need for movement, difficulty with visual complexity, difficulty with visual novelty, visual field preferences, difficulty with distance viewing, absence of visually guided reach, light gazing and nonpurposeful gaze, atypical visual reflexes, and visual latency (Dutton, McKillop, & Saidkasimova, 2006; Jan, Groenveld, & Anderson, 1993; Roman-Lantzy, 2007)
- An environmental assessment to evaluate the impact of the environment on the child’s visual performance and to determine possible environmental modifications to learning materials and for orientation and mobility purposes.

Functional vision assessment for a child with CVI and ocular impairment

Although CVI may coexist with ocular forms of visual impairment, components of each child's functional vision assessment (ocular or CVI) should consider the child's unique medical and ophthalmologic profiles. Children with CVI and ocular impairment (including strabismus, visual field loss, near sightedness, or far sightedness) require an evaluation for CVI on the basis of the
characteristics of CVI and a thorough assessment of the educational impact of the ocular condition (near and distance acuity, visual fields, binocularity, motility, color, light, and contrast).

Additional assessment considerations

Because children with CVI often have additional disabilities, the teacher of students with visual impairments and the educational team should also consider the following when conducting a CVI assessment:

- Use of familiar and unfamiliar items
- Consideration of the child’s communication level and formal communication strategies (such as sign language, an object or picture system, and partner-assisted scanning)
- Expectations for responding (verbal, pointing, eye gaze, vocalization, sign, gesture, and changes in biobehavioral states)
- Incorporation of the wait time for responding
- Use of natural routines for observations
- Proper physical positioning
- Level of fatigue

In summary, the role and responsibility of the teacher of students with visual impairments is to lead the educational team by sharing essential information from the functional vision assessment to assist in the development of the Individualized Family Service Plan (IFSP) or Individualized Education Program (IEP). Teachers of students with visual impairments need to adopt a functional vision assessment that assesses the visual and behavioral characteristics that are indicative of CVI and to integrate these findings into the child’s learning media assessment and Expanded Core Curriculum (Holbrook & Koenig, 1989). Collectively, these data provide the information that is needed for program planning, intervention, and service delivery.

Service delivery

Children with CVI should receive services from teachers of students with visual impairments who have expertise and training specific to CVI. Although other professionals or the children’s parents may have knowledge and skills in CVI, these specialist teachers must take the lead in
assessment, intervention, and service delivery (direct, coaching, consultative, or collaborative). They must work with other professionals (including orientation and mobility specialists, occupational therapists, physical therapists, speech-language therapists, and deaf-blind specialists) so that the team understands how CVI affects development in the areas of cognition, fine motor and gross motor skills, communication, social, self-care, and mobility. Teachers of students with visual impairments must also keep in mind that CVI may exist in individuals at all levels of functioning from those with severe disabilities to those with gifted abilities.

The initial role of the teacher of students with visual impairments is to provide information to the team for determining eligibility. Once eligibility has been established, the specialist teacher’s assessment information assists the team in educational planning. For a child with CVI, a significant amount of time must be allotted to the initial planning phase of the child's intervention program, followed by continuous consultation or collaboration with the team.

The teacher of students with visual impairments works with team members to describe and plan adaptations and specific instructional strategies. The IFSP or IEP contains distinct outcomes, goals, objectives, and components of specially designed instruction that are derived from the assessed needs and are based on the visual and behavioral characteristics associated with CVI. The CVI-specific adaptations allow access to information across living and learning settings. Program planning also includes collaboration with a certified orientation and mobility specialist regarding orientation in space and safe, efficient travel. O&M instruction will be most effective when it is designed according to CVI principles.

Effective implementation of interventions in meaningful and natural environments requires consistent instructional leadership by the teacher of students with visual impairments. Leadership includes one-to-one instruction, modeling and feedback to classroom personnel, the development of curricula and materials, data collection and analysis, coteaching with other professionals, and peer support. The leadership of the teacher of students with visual impairments also includes training parents and family members in methods of implementing appropriate CVI adaptations in the home. These leadership practices are essential whether the child participates in a functional or an academic program.

Once the assessment and program design are complete, the teacher of students with visual impairments may resume a more consultative or collaborative role. This role will include ongoing
assessment and adjustment of the program. Additional information may include formal assessment, observation, video reports, photo journals, and anecdotal reports.

Training needs related to CVI in the field of visual impairment

Preservice training

It is essential that teacher preparation programs for teachers of students with visual impairments develop program competencies that address the identification and assessment of CVI and intervention. Program competencies include the causes of CVI, the characteristics of children with CVI, conducting appropriate functional vision assessments of children with CVI, interventions for children with CVI, monitoring the progress of children with CVI, and making environmental modifications. Another consideration is the development of a subspecialty or endorsement within an existing program for training teachers of students with visual impairments. Given the incidence of CVI, programs could consider developing a specific course that provides knowledge and teaches skills to address the previously mentioned competencies, neuroanatomy, and neurological causes of CVI. Practicum experiences that provide direct experience with children who have CVI are essential.

To reflect the addition of CVI content in training programs for teachers of students with visual impairments, national examinations (like Praxis II) will need to include critical questions about CVI and program competencies, such as the causes of CVI, the characteristics of children with CVI, the appropriate functional vision assessment of children with CVI, appropriate interventions for children with CVI, monitoring the progress of children with CVI, and environmental modifications.

In-service training

Given the complex needs of children with CVI, vision professionals should increase training to other professionals, such as occupational therapists, physical therapists, deaf-blind specialists, speech language therapists, and physicians. In fact, there is a significant lack of in-service training for teachers of students with visual impairments and professionals in related fields. Ongoing training can be accomplished through online instruction, video conferencing, single- or multiple-day face-to-face training sessions, and peer support opportunities.
In-service training and outreach to medical professionals is important to facilitate the early identification and referral of infants and children with CVI. Collaborative training activities with professionals in neonatal and pediatric intensive care units and developmental follow-up clinics will create continuity in service delivery.

Conclusion

Over the past two decades, several theoretical and remedial perspectives have emerged that are related to (1) classification models of CVI and visual processing disorders (Colenbrander, 2009; Dutton, 2008; Hyvarinen, 2005; Morse, 1990; Good, Jan, & Leader, 1990) and (2) assessment and intervention strategies that are based on the behavioral characteristics of CVI (Baker-Nobles & Rutherford, 1995; Roman-Lantzy, 2007). However, the increasing incidence of CVI warrants further inquiry to develop and validate effective practices that apply to CVI. Finally, children whose visual impairments are due to damage to or malformation of the brain represent a large and growing cohort who require specialized services from teachers of students with visual impairments. It is critical that CVI be identified as distinct from visual processing disorders that are associated with other learning and developmental disabilities. Children who have CVI require educational vision supports that are equal to those of any child who is considered to be blind or to have low vision.

References


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Figure 1

Figure 1. The top circle represents visual impairment. The area above the top of the large circle-line represents ocular and optic nerve conditions (60%-70% of the cases of visual impairment). The area of intersection of the two circles represents CVI (30%-40% of the cases of visual impairment). The largest circle represents the processing of visual information.