Pediatric Cerebral Visual Impairment

Notes from the American Conference on Pediatric Cerebral Visual Impairment

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The Children's Hospital and Medical Center of Omaha, NE recently sponsored a symposium with the purpose being to bring together professionals from several fields of study. These professional were to share information, learn from each other, discuss controversial topics, and develop a document suitable for publication detailing principles that we could all agree upon. A second document may also be developed that discusses the controversies in this area and the foundations for these controversies. These documents or transactions will serve as reference guides to all involved professionals, with derivative publications for the lay public to follow.

The professional team invited to keynote this conference included: Mark Borchert, M.D.; Associate Professor of Clinical Ophthalmology and Neurolog, University of Southern California, The Vision Center, Children's Hospital Los Angele, Los Angeles, CA; Christine Roman Lantzy, Ph.D.; Director, Pediatric View Program, The Western Pennsylvania Hospital. CVI Consultant, The American Printing House for the Blind Educational Consultant, Pittsburgh, PA; Jacy VerMaas-Lee, M.A., OTR/L; Assistant Professor of Occupational Therapy, Creighton University, Omaha, NE and Dominick M. Maino, OD, MEd, FAAO, FCOVD-A; Professor of Pediatrics/Binocular Vision Illinois Eye Institute/Illinois College of Optometry, Chicago, Il. The program was developed and moderated by neuro-ophthalmologist, Richard H. Legge, M.D.; Adjunct Assistant Professor, Department of Ophthalmology, University of Nebraska Medical Center, Omaha, NE. The audience included optometrists, ophthalmologists, other MDs, occupational therapists, physical therapists, speech and language therapists and a large number of teachers of the visually impaired.

Conference Topics

1. Defining Pediatric Cerebral Visual Impairment

The definition of brain related visual impairment had been and even today is often confusing, misunderstood and imprecise. It is now, however, frequently referred to as Pediatric Cerebral Visual Impairment (PCVI). Initially Pediatric Cerebral Visual Impairment had also been referred to as Pediatric Cortical Visual Impairment and mistaken for Delayed Visual Development. 1

Commentary in the Journal of Visual Impairment and Blindness 2 noted that in North America the phrase Cortical Visual Impairment was frequently used while elsewhere Cerebral Visual Impairment was considered the preferred terminology.

The story of the development of the concepts of visual impairment due to brain injury begins in the 19th century. Later during World War I, wounded veterans with brain injury displayed an ability to perceive motion in the “blind, non-seeing” visual field. This ability to sense motion, lights, and colors even though the individual has brain injury induced blindness may be conscious or subconscious. This is also referred to as statokinetic dissociation or the Riddoch phenomenon when discussing adults. 3 The
ability to sense such motion was called blindsight\textsuperscript{4} which also appeared to include the ability to ‘sense’ objects in one’s way so that these could be avoided when walking into a room or down a hall way.

Prior to the 1980’s adults with bilateral insult to the occipital cortex were referred to as having cortical blindness. At this time, this term was also applied to children. Cortical visual impairment was used in the late 1980’s onward with the definition of CVI being injury between the lateral geniculate nucleus and the visual cortex with reduced visual acuity being the identifying feature. When it was noted that many children had damage to the white matter surrounding the ventricles of the brain (periventricular leukomalacia PVL), the term cerebral visual impairment was coined and was used to describe the condition (especially in Europe).

Cerebral visual impairment is a more inclusive term that allows for not only significantly reduced visual acuity but also the frequently associated oculomotor anomalies, visual field loss, and vision information processing problems seen in children.\textsuperscript{3} Some researchers suggest that the phrase cognitive visual dysfunction (CVD) be used to identify the many visual perceptual anomalies associated with this condition.\textsuperscript{5}

Colanbrander classified the various areas associated with CVI, these included:

1. Ocular visual impairment: Anomalies of refractive state and optics and eye health.
2. Cerebral visual impairment: Abnormalities associated with pathway problems, cortical problems, and oculomotor dysfunction as well as vision information processing anomalies (dorsal and ventral streaming processing mechanisms).\textsuperscript{6}

Delayed Visual Maturation. (DVM) describes infants who appear to be visually impaired, but usually demonstrate improved visual abilities by the age of 6 months, often without treatment. At this point the children frequently then go on to mirror more normal infant visual development.\textsuperscript{7} Even though infants with DVM were first described in the 1920s, there is little consensus as to the etiology of this disorder. There are several types of DVM with type I being described earlier in this paragraph. DVM type II is characterized by problems with attention and fixation but is also usually associated with other neurological and/or learning abnormalities. Improvement in the infant’s vision takes longer and the end point visual acuity is typically not of the same quality as in DVM I. Many in this category have intellectual disability, seizures, and other developmental issues. In DVM III, the children frequently have congenital nystagmus and albinism. Their vision starts to improve later than infants with DVM type I and can improve to low-normal levels. When Delayed Visual Maturation is associated with retinal, optic nerve and macular anomalies, it is referred to as being Type IV.\textsuperscript{8}

Defining PCVI. Variability with defining various disorders is not all that uncommon. For instance Autism used to be a relatively rare anomaly. Once this definition was altered to reflect a spectrum of individuals with behaviors that have autistic like characteristics, the number of those on the Spectrum is now considered (by some) to have reached almost epidemic status.\textsuperscript{9,10,11} Interestingly the neurological/brain changes associated with this disorder can even mimic many of the behaviors seen in those with PCVI as well.\textsuperscript{12}

Should we be concerned about how PVCI is defined? Absolutely. There are instances where not only do the numbers of individuals with the diagnosis increase exponentially (like that which occurred for Autism), but can also decrease significantly. When the American Association on Intellectual and Developmental Disabilities changed the definition of mental retardation by decreasing the IQ cut off point from 80 to 70 and by adding adaptive behavior qualifications they instantly cured hundreds of thousands of those with mental retardation overnight.\textsuperscript{13} The AAIDD has not only changed the definition of mental retardation, but also the words used to describe the condition. Many years ago the classifications used such derogatory terms as idiot and moron; then mental retardation and now, the preferred terminology in this area is either developmental disability or intellectual disability.

What we call a thing is very important for to name it is to have power over it.

Post conference commentary: After the meeting the group of presenters met to review all that was discussed the previous day. It was decided that the term “pediatric cortical visual impairment” was the preferred terminology to use since this is very specific about the group of individuals being discussed (those with cortical visual impairment only and no other developmental, cognitive or developmental issues). I
noted that most practitioners, therapists, and teachers see children with additional issues such as motor challenges, vision information dysfunction, and other non-cortical anomalies. For these individuals I suggested that the term, pediatric “cerebral visual impairment” is much more appropriate.


There are numerous areas that require a significant number of assessment procedures to ascertain the level of ability of those with pediatric cerebral/cortical visual impairment. We need to assess vision function as well as functional vision.

An assessment of vision function can include determination of the clarity of vision (visual acuity, contrast sensitivity, refractive error), oculomotor ability (pursuits and saccades; convergence and divergence), accommodation (focusing), depth perception (3D vision) and eye health. It is also often appropriate to use special diagnostic tools such as the EOG (electrooculogram), ERG (electroretinogram) and the VER (visually evoked response; VEP, visual evoked potential) to determine the level of ability present. Those with a wide range of disability tend to show many anomalies in the various areas of vision function noted above.

An assessment of functional vision should then be conducted as well. Those with disability tend to have functionally induced disability that often overlays pathologically induced disability, so that the end result is often greater than one might expect from either anomaly individually. For instance a large amount of uncorrected refractive error (hyperopia, myopia, astigmatism) could cause amblyopia (a functional anomaly) that magnifies any vision loss due to cerebral/cortical impairment. The amblyopia also induces numerous vision information processing anomalies that impede a child’s daily living skills development and his or her ability to navigate the world about them. Children with Down Syndrome for example have very poor accommodative abilities that can interfere with all near-point activities from using a computer to reading a book. Those with Cerebral Palsy will display oculomotor, visual motor integration and accommodative problems along with high refractive errors as well.

Another area of concern is that of vision information processing (VIP) and the development of appropriate visual perceptual skills. Directionality, visual motor integration, non-motor perceptual skills, and auditory perceptual/processing skills all have a role to play in child development. Unfortunately those with disability tend to have both functional vision and vision function anomalies that interfere with the development of appropriate vision and auditory information processing ability.

Post conference commentary: Some members of the group thought that the use of electro-diagnostics (VEP, ERG, EOG) were not needed and only confirmed what you already knew. These tests added to the costs involved in managing patients and used valuable resources unwisely. I noted that the use of VEPs could establish a valuable objective baseline of vision function that could be utilized to confirm improvement once therapy was begun and could help guide the therapist in what approaches worked best.

3. Therapeutic Strategies For the Treatment of Pediatric Cerebral Visual Impairment

All treatment should begin by paying attention to the basics. These basics include the various areas of vision function and eye health discussed above. Any problems that need to be addressed to insure the best possible eye health should be instituted. If uncorrected refractive error is present, it should be diagnosed and a prescription for glasses given to the child. It has been noted that even correcting a relatively small amount of refractive error for those with traumatic brain injury can improve these individuals’ quality of life. Remember that spectacles can not only be corrective/compensative in nature but they can also be therapeutic as well.

Children with high amounts of hyperopia and those with accommodative dysfunction (including individuals with Down Syndrome, Cerebral Palsy and brain injury) often benefit from a multi-focal prescription where an added “+” power is given either in a multifocal prescription (bifocal) or as a secondary pair of spectacles to use for specific tasks. Individuals with significantly decreased vision at near can also benefit from high “+” adds and the magnification that results.

Once the refractive prescription is determined and corrected, and any therapeutic applications addressed appropriately for use with a spectacle prescription (bifocals, prism, sector occlusion, etc), then it is time to determine other therapeutic interventions required for any additional vision function anomalies present. It was suggested that Facebook can be a unique resource for therapeutic ideas as well as other internet resources.
(Thinking Outside the Box, Maino’s Memos, Pinterest). Therapeutic procedures to improve eye movement and hand-eye, accommodation, convergence/divergence, and other aspects of both vision function and functional vision, as well as visual stimulation activities were presented as well.

Although children with cortical/cerebral visual impairment have significant neurological impediments, the principles of neuroplasticity can be applied to the various therapeutic approaches utilized for this population. In most instances the therapy is not rehabilitative in nature, but rather habilitative. This difference is important to remember when caring for those with PCVI.

Post conference commentary: The large number of teachers in the audience often directed the discussion towards diagnosing and treating the educational concerns present for these children. Christine Roman Lantzy, Ph.D, frequently referred to her text, Cortical Visual Impairment: An Approach to Assessment and Intervention, as the best way to determine various levels of function and to treat PCVI.

4. How Do Environmental Factors, Medications and Non-Visual Handicaps Affect the Evaluation and Treatment of Pediatric Cerebral Visual Impairment?

Individuals with a handicap tend to be prescribed many more medications than those not demonstrating a disability. They also often have a slightly higher affinity for the development of adverse effects due to various environmental factors. A paper in Optometry discussed adults with not only a developmental disability but also a psychiatric illness that noted many of these individuals were taking 10 or more, high powered neurotropic and systemic medications. Interestingly, seldom did any of these individuals complain of symptoms related to their disability, systemic anomalies, or medication side effects. Certainly those who are significantly younger than the population described above may also find it difficult to communicate their needs, wants and symptoms as well.

Various medications, alternative and complementary medical therapies and even more traditional allopathic approaches to health care can result in adverse, unintended events. (See Table 1). Although you may think that a child is too young for many of these major drugs, you should realize that various psychiatric anomalies such as pediatric bipolar disorder is now one of the most frequently diagnosed mental illnesses in children. Pediatric depression is also being diagnosed often, let alone all the medications currently being used for behavioral issues such as attention deficit hyperactivity disorder.

One of the major environmental hazards those with disability encounter are people. Many do not know how to respond to an individual with a disability. They may make assumptions that are false and then act on those assumptions. This is true not only for lay individuals but also for teachers and health care professionals.

Post conference commentary: Little was discussed on this topic after the meeting. It was decided,
however, that the American Conference on Pediatric Cortical Visual Impairment would be held on a yearly basis over the next two years with support from the Children’s Hospital and Medical Center of Omaha, NE. A website, Brainblind.org (http://www.brainblind.org/) is being developed and should be online sometime during the summer of 2012 for additional information.

For those in attendance, the first ever American Conference on Pediatric Cerebral/Cortical Visual Impairment began a process to determine the optimum way to diagnose and treat those with PCVI. Many left with answers. More left with questions. The planning process continues for the next conference. What would you, as developmental optometrists, want out of such a conference? Contact Dr. Dominick Maino for additional information (dmaino@ico.edu).

References

Note: URLs are functional hyperlinks to internet addresses.