Pediatric Cortical Visual Impairment

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Next PCVIS Conference:
June 27-28, 2014 Oaha, NE

OD Speakers include:
Dr. Joseph Maino,
Dr. Curt Baxstrom,
Dr. Dominick Maino

Case #1
Hx: 2 year 4 mo old, ischemic changes in the cortex with both white and deep grey matter diffuse abnormalities, CP spastic quad, DD, seizures since birth (infantile spasms)

Case #2
Hx: 2 y 5 mo female, picks up toys more, increased facial expressions, still using g-tube. No change in mobility, feeding improving. Eye Health unremarkable
XT onset after head trauma, all milestones delayed shaken baby syndrome, retinal signs resolved, seizures, Prevacid, Topamax

Case #3
11 yr 6 m F. vision problems noted at 8mos of age, optic nerve hypoplasia, nystagmus. VEP all results delayed. Peak poorly formed but consistent with optic nerve hypoplasia, nystagmus intermittent, gtube, seizures, poor handeye. Mobility rolls over

Pediatric Cortical Visual Impairment

An Introduction

1. Define pediatric cortical visual impairment (PCVI)

Definition confusing, misunderstood and imprecise.
Pediatric Cerebral Visual Impairment
Pediatric Cortical Visual Impairment
Delayed Visual Development
History of CVI

- Brain injury 19th century with Phineas P. Gage

World War I, wounded veterans with brain injury
Display perceived motion in the “blind, non-seeing” visual field.
Ability to sense motion, lights, and colors
Conscious or subconscious.

Statokinetic dissociation (in children)

- Greater reduction in sensitivity to stationary visual stimuli relative to similar targets in motion
- Riddoch phenomenon (adults)
  - Ability to sense movement even though blind
  - “See” moving objects...but not stationary ones
- blindsight
  - Ability to ‘sense’ objects in the way

Statokinetic dissociation (in children)

Movement in the peripheral visual field may elicit a smile in the blind child with quadraplegia and profound intellectual disability.
Children who are fed with a spoon may intermittently open their mouths to receive food when the spoon is moved in an arc from the peripheral visual fields, but not when it approaches the mouth from straight ahead.

Statokinetic dissociation (in children)

- For those children who understand language stating what is being seen as the child reacts to it may enhance both visual and language development.
- Such children may rock to and fro. Whether this generates an image is difficult to know.
- Rarely, children with cerebral blindness who are mobile move slowly around obstacles. This phenomenon has been called travel vision.

1980’s adults with bilateral occipital cortex insult (cortical blindness)

- Term applied to children.
- Cortical visual impairment used in the 1980’s onward
- Definition of CVI includes injury lateral geniculate nucleus/visual cortex
Reduced visual acuity identifying feature.
Many children damage to white matter surrounding the ventricles (perventricular leukomalacia PVL)
Cerebral Visual Impairment now used (especially in Europe)

The ventral stream (also known as the "what pathway") travels to the temporal lobe and is involved with object identification. The dorsal stream (or, "where pathway") terminates in the parietal lobe and process spatial locations.

• Delayed Visual Maturation (DVM)
  • DVM type I: Visually impaired infants: improved visual abilities by the age of 6 months, often without treatment.
  • DVM type II: attention problems, associated with neurological/learning abnormalities. Improvement takes longer
  • DVM III: children have nystagmus, albinism. Vision improves later, can improve to low-normal levels.
  • DVM IV: associated with retinal, optic nerve, macular anomalies

Pediatric Cortical Visual Impairment Society
Next PCVIS Conference: June 27-28, 2014 Oahu, NE
Congenital or acquired brain-based visual impairment with onset in childhood, unexplained by an ocular disorder and associated with unique visual and behavioral characteristics.

Founding Board: Lindsay Hillier, Alan Lantzy, Richard "Skip" Legge, Dominick Maino, Linda Nobles, Christine Roman, Jacy VerMaas-Lee
Pediatric Cortical Visual Impairment

**Vision Function**
- Clarity of vision
- Oculomotor ability
- Accommodation
- Binocularity

**Eye health**
- Biomicroscopy, Tonometry
- Dilated Fundus Evaluation
- Special diagnostic tools
  - EOG (electrooculogram)
  - ERG (electroretinogram)
  - VER/VEP (visually evoked response/visual evoked potential)

**Functional Vision**
- Functionally induced disability *that overlays* pathologically induced disability
- Uncorrected refractive error: Amblyopia
- Constant Strabismus: Amblyopia
- Oculomotor dysfunction, Binocular vision dysfunction, Accommodative dysfunction:
  - Attention

**Functional vision**
- Vision information processing (VIP)/Visual perceptual skills
- laterality/directionality
- visual motor integration
- non-motor perceptual skills
- auditory perceptual/processing

**History**
- All the usual questions AND
- General/Motor/Visual/Auditory Development
- Daily Living Skills
- Skills needed for Learning

**Vision Function**
- Clarity of vision
- What is visual acuity?
- What is contrast sensitivity?
- What is refractive error?
Vision Function: Clarity of vision

What is visual acuity?
The ability to see a certain size object at a certain distance

What is contrast sensitivity?
Contrast sensitivity measures the ability to see details at low contrast levels. Visual information at low contrast levels is particularly important:

1. in communication, since the faint shadows on our faces carry the visual information related to facial expressions.
2. in orientation and mobility, where we need to see such critical low-contrast forms as the curb, faint shadows, and stairs when walking down. In traffic, the demanding situations are at low contrast levels, for example, seeing in dusk, rain, fog, snow fall, and at night.
3. In every day tasks, where there are numerous visual tasks at low contrast, like cutting an onion on a light colored surface, pouring coffee into a dark mug, checking the quality of ironing, etc.

4. In near vision tasks like reading and writing, if the information is at low contrast, as in poor quality copies or in a fancy, barely readable invitation, etc.

Pediatric Cortical Visual Impairment

Refractive Error

Myopia (Nearsightedness)
Hyperopia (Farsightedness)
Astigmatism

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Refractive Error: Assessment Objective

Dry Retinoscopy
Mohindra Dynamic Retinoscopy
Cycloplegic Retinoscopy

Objective: Auto-refraction

Mohindra Dynamic Retinoscopy
Dark room
50 cm
Neutralize main meridians
Algebraically add -1.25 to sph

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Refractive Error: Assessment

Subjective
Which is better 1 or 2?

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Refractive Error: Assessment

Subjective
Which is better 1 or 2?
Oculomotor ability
basic extra-ocular muscle assessment
EOMs
Pursuits
Saccades
Convergence
Divergence

Accommodation (focusing)
MEM Nott
Book Bell

Accommodation (focusing)
MEM
Room illumination should be dim but with target illuminated
Briefly insert lens into line of sight
Measurements should be made within 1 second per lens used to minimize the dazzle of light and the effect of lens on accommodation system
The lens that creates neutrality is the value

Accommodation (focusing)
Monocular Estimate Method (MEM): you neutralize the reflex while the patient accommodates to a target at near (usually at 40cm)
With motion: Lag of accommodation --- Add PLUS
Against motion: Lead of accommodation - Add MINUS
Use patient’s correction for distance or near
TRUE measurement of lag/lead if measured with BVA
Place the target at their working distance
Adults: usually 40 cm  Children: use Harmon’s distance

Accommodation (focusing)
Nott Method: clinician moves toward and away from the patient until neutrality is seen (Dioptric difference between neutral and your beginning distance is the lead/lag)
Against motion: move closer to the patient
With motion: move further away from patient
Accommodation (focusing)

Book Retinoscopy

Technique developed at the Gesell Institute by Gerry Getman, OD working with Arnold Gesell, MD.

Accommodation (focusing)

1. Free and Easy reading level, reflex varied from neutral to with motion with bright, sharp edges and had a pinkish color.
2. Instructional reading level (maintaining the reading task with comprehension in spite of being stressed) the reflex was a varying fast against motion while the color was bright, sharp, and very pink.

Accommodation (focusing)

Bell Retinoscopy

A small shiny bell dangling from a string is used as a fixation target (now use a silver ball on the top of a stick). The ball is moved closer to and farther from the patient along this midline. The retinoscope is positioned slightly above this line at a fixed distance of 50 cm. (20 inches) from the patient. Watch what happens to the reflex as you move the ball.

Accommodation (focusing)

Bell Retinoscopy

The distance between the retinoscope and the target when the change in motion occurs is a physical measure of the lag of accommodation. Typically we expect to see a change from “with” to “against” on the way in at 35 - 42 cm. (14 - 17 inches) and a change from “against” to with at 37.5 - 45 cm. (15 -18 inches).

http://www.oepf.org/VTAids/Retinoscopy.pdf
Color Vision

Binocularity (?)

- Fusion
- Stereopsis
- Depth Perception (3D vision)

Binocularity

Eye Health

Functional Vision Anomalies in PCV

- Amblyopia, Strabismus, Oculomotility Disorder, Accommodative Disorders, Binocular Vision Disorders
  - Lea H, Jacob N. What and how does this child see? Vistest Ltd. Helsinki, Finland, 2011.
  - Cerebral Palsy Review (see Taub MB, Reddell AS. Cerebral Palsy. In Taub M, Bartuccio M, Maino D. (Eds) Visual Diagnosis and Care of the Patient with Special Needs; Lippincott Williams & Wilkins. New York, NY; 2012:21-30.)

Treatment of Functional Vision Anomalies in PCV

- Treatment begins with the basics
  - Vision function
  - Refractive error & quality of life
  - Spectacles therapeutic
  - Eye health
Treatment with spectacles
multi-focal prescription/bifocal prism
occlusion
task specific glasses
high “+” adds (magnification)
Low Vision Aids

“The medicine in my glasses has run out!”

Vision Therapy/Vision Rehabilitation/
Vision Stimulation

Use Principles of Neuroplasticity
Use it or lose it
Use it and improve it
Specificity
Repetition matters
Intensity matters

Medications and Alternative Therapies

Medications: Prescribed many more medications
Higher affinity for adverse effects due to systemic/environmental factors
Seldom complain of symptoms related to their disability, systemic anomalies, or medication side effects

RJ Donati RJ. Medications and the Lack of Oculo-Visual Complaints from those with Mental Illness and Dual Diagnosis. Optometry 2009;80:249-254
Medications and Alternative Therapies

Alternative and complementary medical therapies


Traditional allopathic approaches

Medications and Alternative Therapies

Mental illnesses in children
Pediatric Bipolar disorder/ depression


Case Reviews

Case #1

Hx: 2 year 4 mo old, ischemic changes in the cortex with both white and deep grey matter diffuse abnormalities, CP spastic quad, DD, seizures since birth (infantile spasms), Placental umbilical cord problems

Dx: CVI, Delayed visual maturation, exotropia., lower heart rate, meconium aspiration, profound hearing loss bilateral cochlear implants, encephalopathy

Case Reviews

Case #1

Medications: Multiple medications
Participates in vision therapy, developmental tx, speech/OT/PT, PT pool, VA 20/300 PL Teller Cards, 38 cm test dist. OU
Horizontal tracking fine, vertical much more difficult
Binocularity inadequate most of the time, IAXT 30-35PD

Acknowledgements:

Dr. Tracy Matchinski: The Chicago Lighthouse for People who are Blind or Visually Impaired

Dr. Mary Flynn-Roberts: Illinois Eye Institute/Illinois College of Optometry Electrodiagnostic Service

Movies, etc.

Hyvarinen L, Jacob N. What and How does this Child See? Vistest, Ltd. Helsinki, Finland. 2011
Case Reviews

Case #1
VF using toys/OKN drum. Responded well in all visual fields. Contrast sensitivity at 10% level, moderately reduced for his age.
Refraction hyperopia/astigmatism. Tolerates glasses well. No change from last prescription.

Case #1
OD +2.50-2.00X005 OS +2.50-2.50X177
Old Rx Mohindra Ret +3.75-2.50X180 OD +3.50-2.50X180
Near VA good, accommodation/interested in near objects appears to function well.
Health of eyes: normal size, shape, clarity, structure, pupils. DFE previously done.

Case #2
Hx: 2 y 5 mo female, picks up toys more, increased facial expressions, still using g-tube.
No change in mobility, feeding improving. Eye health unremarkable
XT onset after head trauma, all milestones delayed shaken baby syndrome, retinal signs resolved, seizures, Prevacid, Topamax, phenobarbital, ROS unremarkable except for what is noted above. Strong tracking all quadrants, + convergence, +OKN, pupil acc response, Teller 20/200 50cm, Cardif 20/253 at 20 cm, IET, IXT, nystagmus, cyclo +.50-4.00X170 OD +.50-4.00X010 OS
Dx CVI, strabismus, nystagmus OT/PT/speech/developmental tx

Case #3
11 yr 6 m F. vision problems noted at 8mos of age, optic nerve hypoplasia, nystagmus
VEP all results delayed. Peak poorly formed but consistent with optic nerve hypoplasia, nystagmus intermittent, gtube, seizures, poor handeye, Mobility rolls over

Recommendations
High degree of vision function. Continue to work with developmental therapist. Visual search, scan, tracking vertically and hand-eye coordination therapy.
Case #3
OD +75-3.00X170 OS +1.00-4.00X010 cyclo OKN/Teller UTT, can separate head from eye movement, IAXT 10 with 5 R hyper, VF UTT, contrast sensitivity UTT, ref +.50-3.25X180 OD, +.75-3.75X015 OS, pupils OD 2mm OS 3mm RRL, ocular allergies Pataday Rx’d Light stimulus therapy

Case #4
2 y/o HM, genetic mutation L1CAM that lead to hydrocephalus and developmental delays, had VP shunt, in early intervention program, no self feeding, hearing ok, Lissencephaly, ROS unremarkable, born c-section because of large head, APGAR 9 and 9, no meds

Case Reviews
Teller 20/180, Cardif 20/80, +tracking, +OKN, +eyehand, FROM, Ta 26, 26 lids held, +2.25 OD/OS IRET 10PD, PERRL – apd Dx: CVI, IAET, Hordeolum, hyperopia, eye health unremarkable


...Criteria were set to extract a fairly homogeneous group of 21 children with CVI due to perinatal HIE or postnatal anoxia who had extensive gray and white matter injury and multiple neurological deficits; 20 of 21 (95%) had symptomatic epilepsy as well. Subjects entered the study with responses ranging from just a pupillary light reflex to rudimentary perception of outline. Each subject underwent an at-home treatment program. Twenty of 21 children (95%) manifested significant improvement after 4 to 13 months on the program. Results indicate that even in this challenging group, there may be considerable neuroplasticity in visual systems leading to reintegration and visual recovery.

Cortical Visual Impairment Pediatric Visual Diagnosis Fact Sheet http://www.aap.org/cvi/articles/bbf_1.html

Cortical Visual Impairment http://www.aapos.org/terms/conditions/40

Blind Babies Foundation http://blindbabies.org/learn/diagnoses-and-strategies/


...We found that cortical or cerebral vision impairment (CVI) was the most common condition causing vision impairment, accounting for 27.6% of cases. This was followed by albinism (10.6%), retinopathy of prematurity (ROP; 7.0%), optic atrophy (6.2%), and optic nerve hypoplasia (5.3%). Vision impairment was associated with ametropia. ...The mean spherical equivalent refractive error of the children (n = 813) was -0.78 +/- 6.00 D with 0.94 +/- 1.24 D of astigmatism and 0.92 +/- 2.15 D of anisometropia. ...The relative frequency of ocular conditions causing vision impairment in children has changed since the 1970s. Children with vision impairment often have an associated ametropia suggesting that the emmetropization system is also impaired.
Social Media

Pinterest
http://pinterest.com/pediastaff/visual-impairment/

Facebook
Present Blindness American
https://www.facebook.com/preventblindness?fref=ts
Thinking Outside the Lightbox
https://www.facebook.com/Thinkingoutsidethelightbox?ref=ts&fref=ts

Resources:

Taub M, Bartuccio M, Maino D. (Eds) Visual Diagnosis and Care of the Patient with Special Needs; Lippincott Williams & Wilkins. New York, NY. 2012


Hyvarinen L, Jacob N. What and How does this Child See? Vistest, Ltd. Helsinki, Finland. 2011


Resources

Internet
http://drleahyvarinen.com/
http://Mainosmemos.com
http://www.slideshare.net/DMAINO/
https://www.facebook.com/Thinkingoutsidethelightbox?ref=ts&fref=ts

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Treatment of Functional Vision Anomalies in PCV

**How To Modify your Home for Visual Stimulation**

Environment- directly impacts visual development and brain cells

- **Lighting**: to increase stimulation of brain cells
  - Open drapes- position child’s back to windows/doors
  - Use In-direct lighting – floor or desks lamps are best and reduce glare (direct light may damage retinal tissues);
  - compact fluorescent bulbs -16 or 22 Watt with warm color

**Suggestions from members**

**Increase contrast**:
- Use electrical colored tape, stickers, decals to add to objects (bottles, cups) walls, cribs
- Use plain colored sheets, poster board to hang on walls/corners to then attach objects, fabrics to make play spaces or rooms around the home more stimulating
- Use patterned fabrics, carpet squares, cellophane, clear plastic- to add to walls, windows, play spaces
  - Make a “stained glass” window or mobile- use cellophanes, CD’s, Mylar wrapping papers
  - Use carpet squares on floor to mark areas; paint/tape on floor moldings or door jams

**Suggested Materials and Activities to try**:
- **Mobiles**: suspend colorful Mylar, CD’s, strings- provides movement and shiny objects
- Screen savers- computer backgrounds are very stimulating and can become a cause and effect activity
- Household items- use soup cans, quacker oats containers, spoons, metal bowls, colorful cups
- Adhesives- wall decals, stickers; add to lower places on walls

**Be aware** of what you wear or what other sounds are in the environment; competing stimuli make it harder to visually attend and focus

Barbara Halton-Bailey, Education Coordinator Virginia Dept. f/t Blind

**Show, Tell & Reach**:

- Develops understanding of objects and immediate world through hands on experience
- Helps understand daily routines
- Develops better visual and/or motor responses
- Builds sound localization
- Increases active involvement
- Lays the groundwork for crawling and walking
**Treatment of Functional Vision Anomalies in PCV**

Show, Tell & Reach- How to do-

- Slow down the pace during activities
- Routinely take 5 minutes or so; tell what object is and what is happening, allow extra time for baby to “study” with hands, ears, eyes and body
- Provide assistance with reaching
- Babies may need to hold and “get to know” it by touching it before understanding and reaching for it away from the body
- Gradually put familiar toys a few inches away (after initially touching) and make a sound for baby to reach for the object

Barbara Halton-Bailey, Education Coordinator Virginia Dept. of Blind

**Treatment of Functional Vision Anomalies in PCV**

**Defined Spaces or Play spaces**

Defined Spaces or Play spaces-

- Provides incentive for movement, exploration, and independent interaction
- A life-long organizational strategy to enhance efficiency of movement, independence and self-esteem-the use of defined spaces expands and grows with the child
- Use walls and furniture as reference points in each room of the house
- First place toys touching body as baby plays on tummy, back, side or seated on the play space
- Move objects further away and make sounds with the object for baby to reach for

Barbara Halton-Bailey, TVI, NBCT
Education Coordinator, DBVI

**Treatment of Functional Vision Anomalies in PCV**

I love ... the use of shiny emergency blankets. They are like large sheets of reflective Mylar material that kids love to wrap themselves in and look at the reflection of the light off of the wrinkles created in the sheets. ...reflective Christmas gift bags, water bottles filled with glitter, snap and light up neon bracelets or necklaces, pompoms, shiny reflective beaded necklaces, feather boas and the list goes on and on. Sometimes just using neon coloured duct tape over a baby bottle or favorite toy works wonders.

Jody Whelan, Specialist, Early Intervention Early Childhood Vision Consultant Northeast Blind Low Vision Early Intervention Program