Cortical / Cerebral Vision Impairment (CVI): An overview of the attributes, range of clinical presentation, research findings and challenges to habilitate and educate this burgeoning population

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• Graduated from University of Western Ontario with degrees in physiology and psychology as well as in music
• BUSM – MA biomedical imaging, PhD in Anatomy and Neurobiology
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• Emeritus Professor of Vision Science, Department of Vision Science, Caledonian University, Glasgow
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  – Commitment to his patient and to their families
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  – Several textbooks
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Amanda Hall Lueck, PhD

• Direct Service Provider
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• Collaborator
• Mentor

6 📜 Overview of CVI

Barry S Kran, OD, FAAO

7 📜 Setting the Stage: Intro, History, Terminology and Definitions

8 📜 Who is the “C” in CVI?

9 📜 What is the “C” in CVI?

• Cortical Blindness:
  – The “term ‘cortical blindness’ is used to refer to the patient who has been rendered blind by bilateral damage to the occipital cortex.
  • WWI Gordon Homes
  • It is a term introduced as the result of studies of adult patients.
  • ICD 10 code: H47.619 (Cortical blindness, unspecified side of the brain)
    – Only code that is remotely close to describing what we see in children
  –

• Cortical Visual Impairment
  – “Bilateral loss of vision with normal pupillary responses and an eye examination, which shows no abnormalities. In adults, this is considered to be an infrequent event and usually the result of arterial circulatory disease.”
  – Older term which is commonly used in US; rarely elsewhere in the world

10 📜 CVI is the #1 cause of pediatric vision impairment in the developed world!

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  • Tremendous variation of presentation with many either misdiagnosed
CVI is the #1 cause of pediatric vision impairment in the developed world!

• Tremendous variation of presentation with many either misdiagnosed or not diagnosed at all.
  – Individual born prematurely with significant visual, motor, intellectual and other issues
    • May have very reduced acuity, contrast, & an inferior field defect
  – May be much higher functioning but (as examples)
    • Cannot read well without “excessive” enlargement or masking
    • Cannot cross a street or recognize familiar people by sight
  – It is present in a very significant number of individuals with CP and other co-morbidities

11 What is Delayed Visual Maturation?
• Delayed visual maturation is similar to cortical visual impairment in that an infant has a normal eye exam but does not demonstrate typical visual behavior. Unlike CVI, the visual response improves in a child with delayed visual maturation and resolves by one year of life.

• Revised 11/2015

• Important caveat: “Because improvement of vision is mandatory to make a diagnosis, the condition can only be suspected initially, with confirmation of the diagnosis made retrospectively following visual improvement.”

12 Brain Damage vs. Brain Injury

13 What is the “C” in CVI?
• **What is cortical / cerebral visual impairment?**
  
  – 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.

  • Revised 11/2015 AAPOS
  
  • AAPOS definition uses cerebral rather than cortical


  14 □ **What is the “C” in CVI?**

  • Structural definition
    
    – “Bilateral diminished visual acuity caused by damage shown on MRI to the occipital lobes or to the visual pathways” (2009)
    
    • Mezer E et al. Trends in the incidence and causes of severe visual impairment and blindness in children from Israel. JAAPoS 2015; 19:260-265

  • Functional definition
    
    – “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.”
    
    – Dutton GN, Lueck AH. Impairment of vision due to damage to the brain in *Vision and the Brain* edited by Lueck AH, Dutton GN. 2015 AFB Press Page 4

  15 □ **What may be impacted in children with CVI?**

  • Concept of vision function Vs. functional vision
    
    – Colenbrander A. Towards the development of a classification of vision-related functioning – a potential framework in Dutton & Bax 2010

  • Acuity and contrast
    
    – Reduction not explained by “ocular” conditions
Vision-related functioning – a potential framework in Dutton & Bax 2010

- Field
  - For children who are premature, look carefully for an inferior field defect
    - May be complete or partial or relative vs. absolute
  - Neglect

What may be impacted in children with CVI?

- Appearance of the optic nerve
  - Differential diagnosis with congenital glaucoma or familial large cups
  - “….the brain lesion has most likely occurred between the 24th and 35th gestational week, i.e., before the optic nerve is fully developed.”
  - Optic Nerve hypoplasia vs. Atrophy, role of OCT

- Higher order visual processing issues
  - Dorsal Stream
    - Posterior Parietal lobes
    - Where is it system, visual guidance
      - Ventral Stream
        - Inferior temporal lobes
        - What is it system – visual library

Patient history, examination, advocacy
(How do you prepare for, see and then appropriately advocate for your patient who may have CVI?)
What is helpful for assessment

1. Pre-Evaluation
   - Correctly appointed
   - Medical history
   - Birth history
   - Developmental history
   - “CVI” surveys

2. Reports from:
   - Pediatric Ophthalmology
   - Neurology
   - Genetics/metabolism
   - Readings from scans
   - Neuropsychology
   - Educational — including vision educators
   - Occupational therapy
   - Physical therapy
   - Speech and Language

What is the thought process of seeing a patient who may have CVI?

• What, in the reports reviewed prior to the visit, alerts for the possibility of CVI?
• What findings need to be obtained which have not been?
• How can this information be collected in a manner which can aid in ruling in or out CVI and can provide insight relative to habilitation and advocacy?

Patient History / Causes of CVI

• Maternal health
• Pregnancy — term or not, complications during pregnancy
• Anything which can cause inflammation in the developing fetus or peri-natally
  — Infection
  — Anoxia/hypoxia
  — Trauma
• Seizures
• Genetic – those that do impact biochemical pathways in the brain and some that do not but impact function
• Abnormal development without an identified cause
• Presence of CP or hypo/hypertonicity

22 History
• Expressed concerns of parents and others
  – How to engage vision – for ADLs, Communication, Learning, other
• Use of surveys / directed history questions
  – Surveys should not be the sole basis upon for determining the a diagnosis of CVI
  – Surveys are helpful to direct the examination or to highlight areas for further assessment in your office or elsewhere

23 Additional History: Surveys
• Surveys
  – Dutton
    – Ortibus
      • https://www.teachcvi.net/screening-tools Accessed 29 Aug 2017
  – Israeli

24 Directed History Questions
  – Q2. Does your child have difficulty walking down stairs?
  –
  – Q18. Does your child have difficulty seeing things which are moving quickly, such as small animals?
Q19. Does your child have difficulty seeing something which is pointed out in the distance?

Q24. Does your child have difficulty locating an item of clothing in a pile of clothes?

Q27. Does your child find copying words or drawings time consuming and difficult?

Paradigm shift: Adjusting the Exam

• It is not the patient that is uncooperative. Rather, the issue with cooperation is the doctor's inability to create an environment in which the patient can be successful.

• It is not only how one introduces or models and adapts a technique, but when one performs the technique during the exam that will ultimately determine one's success in acquiring that information.
  • Doing the right test at the right time and the right way!

Examples of out-of-the-box testing

• Acuity – procedure and patching
  – Detection vs. Resolution
• Contrast – preferential looking
• Modified dynamic retinoscopy
• Field assessment
• Puzzles

Examination Outcomes/Habilitation

• Use data and observations to implement a treatment plan
  – Spectacles, LV devices, e-magnification, referral for Neuro-psych, determination of initial print size, referral for learning media assessment
  – Someone transitioning from EI to preschool who needed a visually quiet room to engage vision and needed frequent visual breaks
  – Someone who is transitioning from lower school to middle school and can not cross a street or recognize familiar people
  – Someone who is non-verbal, has a sense of cause and effect and family and others is wondering about using vision as part of a communication system
Future Directions

Future Directions

• Need for inter-disciplinary collaboration for patient care and research
• Need for additional training to work with this population by eye care providers, vision educators, others
• Need for forums, like this meeting, to have sessions which would include grand rounds with a panel of professionals with experience in this area.

Resources for CVI

– Some CVI resource links
  • cviscotland.org
    – Parent site with accessible information. Vetted by Gordon N Dutton
  • http://biomed.science.ulster.ac.uk/vision/-Visual-skills-inventories_60_.html
    – Ideas for strategies for children of various ages and needs
  • http://www.ssc.education.ed.ac.uk/resources/vi&multi/cpvi/ssreport.pdf
    – Short document introducing CVI and important aspects
  • http://www.ssc.education.ed.ac.uk/resources/vi&multi/cpvi/ssreport.pdf
    – Very long document with a more in depth overview of both CP and CVI.
  • ech.aph.org/cvi/
    – American Printing House for the Blind site on CVI

Resources for CVI


– Kran BS, Mayer DL. Chapter 14 Vision impairment and brain damage in Taub, Bartuccio, Maino eds Visual diagnosis and care of the patient with special needs. Lippincott 2012


Please remember to complete your session evaluations online.
What do neuroimaging studies tell us about how the brain develops in individuals with CVI versus other types of visual impairment?

(22 minutes)
Corinna Bauer, PhD

1. Overview of neuroplasticity in relation to brain development and blindness
   a. Principles related to neuroplasticity
   b. Neuroplasticity in the context of ocular blindness

2. Common imaging techniques and their application to the study of neuroplasticity
   a. Structural MRI
   b. Diffusion MRI
   c. Functional MRI

3. Neuroimaging related to CVI
   a. Do standard clinical neuroimaging techniques inform us as to the type and severity of visual impairments observed in CVI? (This will tie into previous talks)
   b. Changes to structural white matter networks associated with functional vision impairments of the dorsal stream (e.g. visual attention and motion processing) as shown by diffusion imaging
   c. Changes to functional networks associated with functional vision impairments as shown by resting state fMRI
   d. Relationship between functional neuroimaging and deficits in motion perception in CVI as shown by task-based fMRI.
   e. Changes observed in the ventral stream – not observed consistently, but does relate to subject-reported visual deficits

4. Implications of brain changes associated with CVI – What does it mean and why is it important
   a. Prognostic value of neuroimaging for CVI
   b. How advanced neuroimaging can be used to monitor changes in vivo
   c. Future research directions
Difficult seeing one’s feet, ‘clumsiness’, and seeing only part of the scene at each glance, related to posterior parietal dysfunction: a common form of cerebral visual impairment.

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Nicola McDowell, MSpTchg

ABSTRACT
Dysfunction or injury of the posterior parietal lobes impairs 3D mapping of the visual scene and can...

(1) Make body movement guided by vision inaccurate (optic ataxia).

(2) Limit the number of items seen at a glance, due to simultanagnosic visual dysfunction.

INTRODUCTION
The way the brain processes the visual image is the subject of a vast literature. This mandates...
the need for an academically sound practical model that helps ensure that visual disorders due to brain injury are recognised, identified, understood, and optimally managed. In-depth studies of the visual outcome of focal brain injury have historically proven an effective basis for such an approach.¹

Without our being aware of it, the top of the brain near the back on both sides (the posterior parietal lobes) subconsciously maps and monitors the three dimensional characteristics of the surroundings in relation to the body, in terms of visual,² auditory,³,⁴ and tactile (5) constructs, allowing us both to map our surroundings and to locate ourselves within them. This ever-changing ostensibly non-conscious internal mental representation of our environment, is integrated with proprioceptive mapping of the location of all parts of one’s body, and data provided by the balance system, to provide the framework for visual guidance of movement, visual search and visual attention.²,⁶,⁷

The posterior parietal lobes on both sides are where the three arteries that supply each side of the brain terminate in a ‘watershed zone’, leading to an enhanced risk of damage in this area due to reduced blood flow,⁸ impaired oxygen delivery,⁹ or insufficient glucose supply.¹⁰ Head injury can also affect this site,¹¹,¹² as can many other disorders.

The outcome is disabling, but the diagnosis can be missed because neither long-term impairment, nor recent loss of these non-conscious visual functions can be recognised or described by those affected, culminating in ‘hidden disability’. Unilateral and bilateral posterior parietal brain injury adversely affecting vision is, in our experience, common in both children and adults, but its disabling features can easily be misinterpreted or even go unrecognised.

This paper presents key aspects of a series of salient case reports, that when assembled, help explain the nature of disorders of visual function due to bilateral posterior parietal brain injury. The aim is to provide a coherent set of insights into this common disabling condition.

**CASE REPORTS**

1. The woman who learned to walk freely through her invisible surroundings.

MC was 30 when she developed a severe lung infection and very low blood pressure, leading to coma for more than a month. On waking, she could not see because both her occipital lobes had lost their blood supply.¹³

Two years later she presented with little useful vision, but she asked why she had recovered her ability to see rainwater running down a window but not to see through it; why she could see her daughter’s pony tail moving from side-to-side when she walked away, but...
not see her daughter; and why she could see water spiralling down the plug hole but not see her daughter in the bath. As these features indicated intact motion perception with absent static perception (the Riddoch phenomenon), she was invited to walk in figure of eight fashion around a row of chairs. Much to her surprise she could do so. A rocking chair was recommended, and she still uses it to help her see round the room, as she can only see when the scene is moving. During the ensuing 7 years she has learned to move freely round obstacles. She can accurately catch rolling balls and she can pick up a drinking glass as if she were sighted when she taps the table, because the resulting ripples render the glass visible and accessible to reach for accurately.

Her CT scan at the time showed strokes affecting both occipital lobes and both temporal lobes, but her posterior parietal

![Figure 2: Case 2: Original diagram in Gordon Holmes paper (1918) (17) depicting the nature of the penetrating head injury and the visual field in Sergeant K. (With permission of the British Journal of Ophthalmology.)](image-url)
lobes were not affected. More recent fMRI scanning reveals evidence of bilateral residual functioning middle temporal lobe tissue – serving perception of movement – becoming active in response to moving imagery (Figure 1). As for the similar case DF, described by Milner and Goodale, she has learned to read by imagining she is moving a forefinger over each letter, despite not being able to see them consciously.

MC has little or no occipital lobe tissue. Her type of vision has been described as ‘Blindsight’, which is conceptually the antithesis of the pattern described in the introduction. This illustrates the nature and character of the visual functions served by the middle temporal lobes and posterior parietal lobes functioning in harmony, but in the absence of occipital lobe function. She can walk accurately through her surroundings, which become mapped in her mind when she starts to move, as the surroundings move in the opposite direction with respect to her body. It is likely that the combination of her functioning middle temporal and posterior parietal lobes allows her to do this in spite of the infarction and loss of both occipital lobes. Once she had gained insight and understanding of the origin and nature of her residual non-conscious vision, she progressively gained the confidence to use it to move independently without assistance. A film about MC’s vision entitled ‘The Blind Woman who Saw Rain’ can be viewed on YouTube (https://youtu.be/9ABQ-U6V0tY).

2. The soldier who could see but couldn’t see.

In July 1917, Sergeant K sustained a probable rifle bullet injury through the lower part of his posterior parietal lobes (Figure 2). He was ‘intelligent and well educated’. His visual acuities were 20/40 and J1 and he could see stereoscopically. He had lost his lower visual fields, and attended poorly to his right. He could only read single words very slowly and uncertainly, and could not follow a passage unless single sequential words were viewed through a slot in a piece of paper. Apart from impaired convergence and accommodation, his oculomotor functions were intact. He retained good visual memory and could correctly recognise objects, but could not reach out and grasp them. When walking he consistently collided with objects he could see. He was unable to direct his gaze at someone speaking with him face to face. When 4 to 6 coins were placed inside his sighted area, he could not see nor count them all, tending to re-count ones he had already identified.

This First World War account, graphically shows that injury of the lower portion of the posterior parietal lobes leads to loss of the lower visual field in both eyes, accompanied by inaccurate visual guidance of movement (optic ataxia) and inability to see more than two or three items at a time (simultanagnosia), resulting in profound impairment of visual search. In essence the functions lost were those retained by MC (case 1), while those retained were those she has lost. The clinical features described for this soldier are typical of those seen today, in varying severity in children and adults with bilateral posterior parietal dysfunction or injury.

3. The woman who couldn’t find things and felt clumsy.

One of the authors was contacted for advice by a 58 year-old family friend. She felt there was something the matter with her vision but wasn’t sure what. She was progressively losing her ability to find items in a kitchen drawer, to identify clothes in her wardrobe and to find a friend in a group. She felt she was becoming clumsy, and found it helpful if she ran her hand along a surface to reach for an item. These are typical presenting features of posterior parietal dysfunction. The rare diagnosis of posterior cortical atrophy was mentioned as a possible explanation, and strategies to help were suggested, and immediately taken up and
implemented to good effect. Two years, and three separate neurology consultations later, this diagnosis was finally made. Now, despite normal visual acuities for single letters and intact ability to detect peripheral movement, she can see and identify only one or two items at a time. At present her lack of visual function resulting from her focal posterior parietal pathology is frustrating but accepted.

Posterior cortical atrophy causes progressive simultagnostic visual dysfunction culminating in complete simultanagnosia. One item only can be seen at a time. Her visual guidance of movement is inaccurate. She is however, incompletely aware of the severity of her visual disability because the visual functions she has lost are non-conscious. Agnosia is not to know. Anosognosia is not to know that one does not know. Simultanagnosia and optic ataxia tend to be anosognostic (which is arguably a saving grace).

4. Heart infection in a child that led to bleeding into both posterior parietal lobes.

Jacob was three when he developed infective endocarditis. Bleeding from secondary infected aneurysms in both posterior parietal lobes was drained neurosurgically. He recovered well with antibiotic treatment. He was ten when first referred for visual assessment owing to disability reading long words, following text, writing words in line, and copying. The progressively diminishing text size and increased crowding of print given to 10 year olds were described as rendering the text illegible, and he was losing self-esteem and confidence. He often walked into people as if they were not there. His difficulty visually following moving objects (e.g. a car or an aeroplane) and his difficulty reading text were associated with impaired visual scanning. Difficulty climbing down stairs and stepping off kerbs were evident. He had no refractive error. His visual acuities were 20/15 in each eye. Stereoacuity was normal as was colour vision. His visual fields elicited using confrontation methods appeared full, but were difficult to plot because he could not simultaneously perceive both the central and peripheral targets. His eyes were aligned but he showed jerky pursuit eye movements. Voluntary saccades to visual targets were difficult to elicit but he had no difficulty making saccades to commands, like ‘look up’. He could identify events in the ‘Cookie Theft’ picture from the Boston Diagnostic Aphasia Examination (used to assess simultanagnosia), yet took a long time to identify a pencil on a cluttered desk. Brain imaging showed severe damage to both posterior parietal lobes (Figure 3). His behavioural features, symptoms, signs and imaging were consistent with the diagnosis of a variant of Balint syndrome.

An adaptive approach using his strengths and abilities was encouraged. He learned to scan the ground ahead. His environment was de-cluttered. Well-spaced text and masking of text above what he was reading (rather than below, which interferes with accessing the next line) was adopted. Auditory strategies, of listening and dictating to a scribe were
implemented. The senses of hearing, touch, and proprioception to locate and reach for objects were employed, while deliberate systematic scanning of the environment was achieved. All this led to greater independence and improved self-esteem, while his prior behavioural outbursts due to frustration became infrequent.

Adaptive and training strategies matched to the needs of those with bilateral posterior parietal pathology can prove effective.

5. The teenage girl with an arteriovenous malformation in her left occipitoparietal area that bled.

In 1996 Nicola sustained a spontaneous rupture of an arteriovenous malformation into her left occipital lobe. After 8 days of drug-induced coma the malformation was neurosurgically excised. After an early episode of spontaneously resolving left sided weakness, perhaps due to vascular spasm, she recovered well, with initial reports of ‘no lasting damage’, but outpatient visual field testing identified a right-sided homonymous hemianopia with central sparing. No other visual difficulties were identified. However, from that time Nicola experienced profound visual difficulties in cluttered or crowded conditions, but was unable to articulate what they were. She called them her ‘visual gremlins’. Seventeen years later, she learned at a lecture on cerebral visual impairment that her symptoms fitted with dorsal stream dysfunction. These included her difficulty finding objects in cluttered surroundings, inability to pick out and recognise familiar faces in a crowd, and feeling overwhelmed in crowded places. She struggled to read crowded text and could not locate where sounds or voices were coming from. She has also felt clumsy. All these difficulties persist.

Nicola cannot focus on a single object surrounded by other objects, because she feels compelled to look at another one, with the result that the first one ‘disappears’ and cannot easily be found again. This is consistent with competitive mutual inhibition of object recognition, which is exacerbated when the objects are moving, or competitive simultanagnostic visual dysfunction. This complex difficulty was dramatically demonstrated to others when she was asked to look briefly at a large audience and count how many people she saw. She explained that she could only focus completely on a single face in the crowd. The other people were seen as blurred images surrounding this one face. Her ‘need’ to look at another face, which she fulfilled, rendered the first face invisible and subsequently inaccessible. On looking away from the audience and back again she found it impossible to locate the person she saw the first time, and once again could only isolate another single clear face. She described her emotional reaction to this exercise. Anxiety had overwhelmed her and led her to frantically scan the audience, darting from one image to another with no discernable pattern to her scanning technique. This panicked approach did not give her time to focus on any individual, and she found it impossible to estimate how many people were in front of her. Prior to this experience she had not been able to describe her simultanagnostic vision because she had had no awareness of what her ‘visual gremlins’ were, but once she had had her vision explained, everything fell into place. Her ‘clumsiness’ was found to have the pattern of optic ataxia.

This long-term simultanagnosia and optic ataxia had ‘impacted hugely’ on her ability to cope in congested environments and social settings, whether familiar or unfamiliar. Her prior unawareness of the nature of her visual problems and why she found certain situations hard to cope with had progressively eroded her self-confidence and led her to withdraw from many social activities. However, having gained insight and understanding, and having become able to acknowledge the origin of her ‘gremlins’, Nicola is now developing coping strategies in crowded and cluttered places.
First she allows herself time to slowly scan a crowded location, and consciously suppress her otherwise overwhelming emotions. Second, to help her identify specific people in a large group, she now slowly scans to seek obvious features. She notes who is wearing a bright red top, for example, and focuses upon their exact location, being ready to relocate them on her next scan, with the aim of building up her best overall picture. Her new knowledge and understanding of her condition have considerably improved her quality of life because she can now use rational approaches to make best use of her vision.

Simultanagnostic visual dysfunction can cause overwhelming discomfort in crowded locations. Giving motivational support to those affected by simultanagnosia, and an understanding of their condition, along with ideas to help them to develop strategies to overcome their unique difficulties, can prove very effective in helping them learn to make best use of their vision. Nicola is one of the authors of this paper and has worked together with the mother of a child with similar visual difficulties to create simulations her own visual difficulties. Figure 4 shows two pictures that look the same to Nicola when she views the person who has been rendered clear in the ‘doctored’ image and the same person in the ‘undoctored’ image.

This pair of photographs, illustrates the qualitative nature of Nicola’s simultanagnosia as she experiences it, now that she has insight into her condition.

6. The boy who couldn’t, then could place a peg into a peg board.

One of the authors was shown and asked to explain a pair of videos at a meeting. In the first, a kneeling boy, known to have bilateral periventricular white matter pathology affecting his posterior parietal territory, was struggling to place a peg into a peg-board. In the second, taken a few minutes later, he easily performed the task. A key feature had been missed. In the second video, but not the first, the boy’s knee was touching the peg-board. This was later found to account for the differing performances.

Tactile input that supplements the degraded non-conscious posterior parietal representation of visual space serves to enhance egocentric localisation, and thereby helps diminish impaired visual guidance of movement.

7. The teenager who played the piano using his thumbs to locate the keyboard.

A 19 year-old man with multiple disabilities and visual impairment enjoyed creating his own ‘music’ at the piano. It was questioned why he pushed his thumbs against the front of the piano whilst doing so. It was found that he no longer needed to do this when his piano stool was elevated so that his legs touched the piano instead of this thumbs.
Again, this anecdote highlights that tactile supplementation of visual guidance adds data that enhances accuracy of self-localisation in surrounding 3D space.

8. The child who did not know where sound was coming from, who lost moving balls and who wouldn’t look at his mother when she was talking to him.

A 10 year old boy with superior posterior periventricular white matter pathology, lower visual field impairment and difficulty finding a toy in a toy box, or his mother in a group of mothers waiting at the school gate, was described as not being able to locate where someone was calling from, and rarely if ever looking at the face of those who talked to him. When asked about this he replied that he could not locate the origin of sounds, and that he was unable to listen when looking at a face, and needed to look away towards an uncluttered area to hear what was being said.

He was given a mobile phone and taught how to use it when out and about so that he could find his family members who now phone him rather than calling out. He was also taught how to glance, then turn and smile at his mother’s face once she has finished speaking. This has proved effective in helping to build up their relationship.

When playing football he chooses to play in goal every time. He was asked what a ball looks like when it is kicked. “It disappears for about a meter of course, then it comes back when it slows down” was his reply. On assessment he could not count fingers on a quickly moving hand, only identifying them when the hand moved slowly.

The posterior parietal and middle temporal lobes together dynamically map the surroundings for both sound and sight, and damage to this area can impinge on both functions. Children with posterior periventricular white matter pathology causing cerebral (cortical) visual impairment can have difficulty dual processing sight and sound. They can also have difficulty locating where sound is coming from. They can be taught simple strategies that help them to overcome these difficulties.

Inability to see fast movement (dyskinesia) is not uncommon and needs to be sought in children with this history. Slowing facial expressions, watching films without fast movement, and training ball skills with light slow balls initially are warranted, and can all prove effective. Playing in goal prevents the need to identify all the players, and approaching balls do not disappear, they simply appear to get bigger.

9. The baby who reached out for the first time

An eleven-month old boy had sustained bilateral posterior parietal damage owing to lack of blood supply to his brain during delivery leading to hypoxic ischemic encephalopathy. Despite apparently normal motor function, he had never reached out for anything, and sat smiling with his hands by his sides. It was hypothesised that the explanation could relate to impaired posterior parietal function leading to deficient visual mapping of his surroundings.

He was placed in a cot lined with a white sheet. A single toy was placed beyond reach. About twenty minutes later he had worked out how to move to reach for the toy. He picked it up and played with it. He had never done this before. Gradually his skills developed, as a sequel to a range of parenting strategies, and he now attends mainstream nursery school.

It was not possible to know why this child would not reach out. However, from the distribution of his brain injuries it was surmised that he could have had simultanagnistic visual dysfunction and optic ataxia. Simplifying his visual environment enabled him to locate and play with a toy for the first time, adding weight to this hypothesis.
10. The girl whose upward, but not downward, reach and grasp are accurate

An 18 year old girl who had been born at 24 weeks gestation approached one of the authors at a meeting where both were contributing. She asked why she was clumsy.

She was found for the first time to have no detectable visual function below thirty degrees from the horizontal. When asked to reach out for objects in her intact lower visual field she consistently did so inaccurately, with a wide gap between fingers and thumb as she did so. However, her reach in her upper visual field was accurate, with a normal in-flight gap between fingers and thumb as her hand reached out for the proffered objects.

In those with periventricular white matter pathology associated with premature birth, lower visual field impairment is common. Not only can the peripheral lower visual field be absent, but also the apparently intact paracentral lower visual field can have a reduced resolution. In our experience this can be associated with inaccurate reach and grasp in the lower visual field, but accurate reach and grasp in the intact upper visual field. This knowledge can be used to advantage.

11. The girl who lost her ability to perceive the whole scene, but could still see in minute detail

At age 9 Lea underwent cardiac surgery for a complex congenital cardiac anomaly diagnosed before birth. Low cardiac output during surgery and probable cerebral emboli led to multiple bilateral infarctions in the parietal and occipital lobes. Good recovery of vision followed a short period of cortical blindness and she returned to school a few weeks later. She had been a gifted pupil pre-operatively, but post-operatively she could no longer read and could not visually encompass a complete page of a textbook, the class or the playground, despite normal visual acuities. Lea found this stressful as she had been told her vision had recovered. All tasks at school and at home involving visual search had become impossible. She could perceive tiny detail in an object or on a textured carpet but could not find a specified object in a room, in a drawer or on a cluttered table. The apparently inexplicable paradox of her clear detailed central vision but lack of global vision, were a cause of anxiety for Lea and her family.

Lea was offered and underwent intensive training for five half-days a week. She was trained to detect, orient to and grasp visual stimuli presented peripheral to central fixation with the aim of enlarging her attentional visual field. She was trained to pay attention to the global scene, in favour of the local aspects of shape and detail. Hierarchical stimuli (such as a big circle made up of a circular line of small squares) were presented, and she was asked to identify and name each large shape while disregarding the small ones. Reading training included tactile exploration of raised letters and words while reading aloud. After a few weeks, her simultanagnostic visual dysfunction had regressed almost entirely. She regained her reading skills and visual detection in her peripheral visual field, and returned to full time schooling.

The dissociation between intact central detailed vision allowing a child to see and pick up a paper clip from a carpet, in the context of a major deficit in global vision, can be counter-intuitive for parents, teachers and clinicians. Bilateral parieto-occipital damage commonly is accompanied by acute cortical blindness initially. Central visual function tends to recover, but in our experience, a spectrum of visual attentional dysfunctions can persist. This includes the condition of peripheral global attentional dysfunction – yet with persisting capacity to detect single peripheral moving targets – ranging to full-blown Balint syndrome in which only single entities can be seen (simultanagnosia) and visual guidance of movement is profoundly impaired (optic ataxia). The intensive visual training Lea received was accompanied by good recovery. There is need
for clinical trials to standardize and evaluate the efficacy of this approach.

12. The 17 year old boy with multiple disabilities and visual impairment (MDVI) who looked around for the first time in a bright monochromatic tent.

Tom is 17. He has profound quadriplegic cerebral palsy and is registered blind due to CVI. He is sociable, and responds well to sound, but is startled by sudden fast movement. For most of the day he used to hold his head down, which was ascribed to poor head control. He rarely used his limited vision.

However, when surrounded by a fluorescent orange ‘tent’ for the first time at age 17, he lifted his head to look around and laughed with pleasure, something he had never done before. As a sequel to daily 15-20 minute sessions in the tent, he became motivated. He concentrated on single items, and he reached out and touched a slightly moving tambourine. He also began to track it in his more functional left visual field. Within a week he had started, for the first time, to hold his head up. Later he became able to do this outside the tent as well.

A year later he can still only focus on single items in the ‘tent’, providing evidence that he can only process one item at a time owing to profound simultanagnosia. Tom has now gained a better posture, holds his head up to view his surroundings and can now locate light and colour movement in a multisensory room. He has also developed more engaging behaviour in communal activities.

It is not possible to test for objective evidence of simultanagnosia in those with profound learning difficulties, but Tom’s markedly improved use of vision after a trial of removal of all pattern and clutter in a monochromatic tent, gives compelling evidence that this phenomenon is present. His newly found capacity to look around when not inside his tent, indicates that this ability is transferable to the world outside of it too.

EXPLANATION AND DISCUSSION

Taken together, the case histories described show that the combination of apparent clumsiness, impaired visual search and evident discomfort in crowds are highly suggestive of bilateral posterior parietal dysfunction, or injury. When we became attuned to this clinical picture, it was remarkable how common this disorder in its various guises turned out to be.

Without one knowing, the posterior parietal lobes serve the essential function of creating a moment-to-moment 3D mental non-conscious emulation of the surrounding moving visual scene that facilitates visual guidance of movement amongst the entities that make up that scene. These entities are given their conscious identities by interplay with the image libraries within the temporal lobes, while attention to and choice of these identified items, is mediated by the prefrontal area of the brain. To ensure accurate visual guidance of movement, the centre of this emulation is coincident with and relates to the midline of the body. Head and eye movement, supported by extraocular muscle and neck proprioception to stabilise the imagery with respect to the body, serve to extend this field of perception. The vertical and horizontal orientation of the scene is integrated with the balance (labyrinthine) system, while coincident timing to events in one’s surroundings is supported by the motion perception brain areas including the middle temporal lobes, the upper mid-brain and cerebellum.

In our clinical experience posterior parietal dysfunction degrades this non-conscious emulation, potentially resulting in inaccurate visual guidance of movement and/or incomplete visualisation of the scene, in any combination or degree, depending on the nature and extent of the functional deficit.

The posterior parietal lobes thus reconfigure visual information received via the dorsal stream from the occipital lobes and from the middle temporal lobes that process moving imagery. They create the non-
conscious virtual, dynamic three-dimensional pictorial and auditory\textsuperscript{3,4} mental emulation of the structure of our surroundings that enables us to move accurately without collision and injury. This constantly changing data-set is integrated and cross-referenced with:

- Synchronous input from the two eyes. (Asynchrony leads to the Pulfrich phenomenon, where the apparent vectors of moving targets are not coincident with their actual vectors.\textsuperscript{28})
- The automatic reflex protective visual system served by the upper mid brain and thalamus\textsuperscript{29}
- Conscious visual storage and analysis of what things are, served by the temporal lobes, connected to the occipital lobes via the ventral stream
- Predictive visual memory input allowing one to move amongst moving targets such as in a crowd
- The internal ‘plumb line’ (vestibular) input from the utriculus and sacculus of the inner ear that creates the percept of what is vertical and horizontal - even when the eyes are closed
- The dynamic movement input from the semicircular canals of the inner ear that detect and accord knowledge of the rate and direction of one’s change of momentum
- Accurate timing – to integrate with one’s relative movement through the environment – accorded by the cerebellum
- A dual proprioceptive system –
  - from the body’s muscles that inform of the position in space of body parts, and
  - from the extra-ocular and neck musculature, which relates the relative positions of the head and eyes in relation to the body, thereby stabilising image location with respect to the body, despite head and eye movement.\textsuperscript{6}

- Frontally mediated choice of attention and action, linked to the capacity to plan and predict outcome

Any of these functions can be disturbed and each needs to be considered in anyone who has suffered brain injury affecting their perception of where they are in visual space.

The clinical presentations of posterior parietal brain injury described in this selected series of clinical vignettes can be drawn together to provide a concept framework that gives a practical working model for recognising difficulties consistent with posterior parietal brain injury, in order to understand, conceive of and implement salient customised strategies to deal with the resulting issues for each affected adult or child, as each is unique.

Initial history taking from a close family member or friend is essential, because those with injury or dysfunction of the posterior parietal lobes can rarely describe their visual difficulties. The non-conscious nature of their visual dysfunctions means that although the resulting behavioral features are evident to those who know them well, those affected often find it very difficult to understand and to describe their own visual difficulties.

Subsequent careful observation of behaviors under the circumstances described in the history, serves to both corroborate and characterise the nature and degree of the specific disabilities identified. For each of the cases described, environmental adaptation and tailored compensatory strategies proved highly effective. While targetted rehabilitational training also proved effective in case.\textsuperscript{11} An additional fundamental approach that was particularly effective for case,\textsuperscript{5} was in depth explanation of what became her erstwhile anosognostic visual difficulties. Now that Nicola understands the origin and nature of her difficulties she has been motivated to work on her lifestyle and to devise multiple strategies of her own to enable her to deal with her difficulties constructively. This process has been life enhancing not only because...
it has been empowering, but it has allowed Nicola to share her new knowledge about her vision with friends and family, who are also empowered to make salient accommodations which have now become second nature. The whole family has benefitted.

The posterior parietal lobes perform myriad rapid computations to create the non-conscious moment-to-moment internal representation of our surrounding environment enabling us to parallel process vast amounts of information to create the ever-changing emulation through which we elect to move and interact. Damage to this area alone or in combination with adjacent brain areas, renders this process deficient, but in ways that are unique for every individual. To complement the systematic evaluation of visuo-attentional processes a specific and structured comprehensive assessment of neurovisual function using investigations matched to the child’s, age abilities and vision need to be performed in every case.

Successful habilitation and rehabilitation are contingent upon starting from scratch with each affected individual to build up a comprehensive profile of the resultant disordered and deficient mapping of surrounding sights and sounds, and the nature and degree of the resulting subjective disruption of knowledge or where one is in the environment (egocentric localisation) and the trajectories of moving surrounding elements of the scene, such as people in a crowd (allocentric localisation).

The impact that this profile has upon the individual concerned is characterised, and the individual’s strengths and abilities are ascertained. A customised matching profile of habilitational strategies is designed and implemented in such a way that it evolves to match the progressively changing needs of the affected individual, as quality of life improves.

It is never to late to start this process!

**Addenda**

Of the clinical vignettes described in this paper case 5 is autobiographical, cases 1, 4, 9 and 12 describe published material. The remainder are published material co-written by the first author.

Each of the authors contributed case material for this paper and were involved in the design of the overall paper. They have all edited the manuscript and approved the final version.

None of the authors is aware of any conflict of interest. Additional information about CVI is available at: [http://cviscotland.org](http://cviscotland.org).

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Maximizing Access to Learning and Educational Content in the Schools for Children with CVI

Amanda Lueck, Ph.D.

How Do We Transform What We Know into Effective Programs for Students in the Schools?

- COPE number (54869-NO)
  - Amanda Lueck, Ph.D.
    - AAO, 2017

Overview of Session

- Setting the Stage: Impact of CVI
- Collaborative Solutions
- Identification
- Assessment
- Universal Design for Learning

Overview of Session

- Differentiated Instruction for Optimal Access to Learning
- Entitlement to Services
- Service Needs to Move Forward
- How Can Optometry Help?

Setting the Stage: Impact of CVI

- CVI is leading cause of visual impairment in high-income countries and is on the rise in low-income countries
- Increase in survival rate of brain injured children born prematurely
- Improvement in treatments for eye disorders
  - 14 million surviving pre-term births worldwide in 2010

Three Subgroups of Children

- Children with profound visual impairment due to CVI
- Children with CVI who have functionally useful vision and cognitive challenges
- Children with CVI who have functionally useful vision and who work at or near the expected academic level for their age group

Quick Review of Potential Behavioral Consequences

Behavioral Consequences of CVI

- Acuity
- Contrast
- Color
- Visual Fields
- Control of Eye Movement (e.g., nystagmus, unstable fixation, inaccurate saccades, deficient smooth pursuit movements)
- Accommodation disorders
- Perception of movement (can be impaired or absent)

Quick Review of Potential Behavioral Consequences

- Visually-guided movement concerns
- Object recognition issues (objects, shapes, faces, facial expressions)
- Difficulty with route-finding
- Visual memory task difficulties
- Visual attention and perception issues (visual neglect, difficulty seeing things in visually cluttered targets, difficulty separating foreground and background)
- Auditory processing issues
- Social and self-esteem issues

Holistic Concerns

- CVI can affect many areas of development and learning
- When there are issues acquiring basic skills these can affect the acquisition of more complex skills required to complete pre-academic, functional academic, and
academic tasks
• Need to look at the holistic needs of children

• Collaborative Solutions
• Because the manifestations of CVI can be quite diverse:
  • Requires collaboration of medical and educational systems
  • Continuous transfer of information among professionals and families
  • Highlighting the importance of team approaches to assessment and intervention with multidisciplinary members on the team

• Identification:
  Identify Children Early
• understand their visual capabilities including visual processing
• how these impact their development

• Identification
• Systematic methods to identify children in the schools, including those with normal near-normal visual acuity but whose vision is compromised such that they cannot use vision to extract information from educational materials
• Differentiate children with CVI from those who have Autism Spectrum Disorder, ADHD, Learning Disability including Dyslexia, or Developmental Coordination Disorders (or children may have more than one condition)

• Identification Of Children
• Look for more subtle signs of the disorder in children who have CVI with additional disabilities including those with
  • Cerebral palsy (Ego, 2015)
  • The impairment of visual-perceptual skills must be considered a core disorder within the CP syndrome.
• A more systematic approach to neuropsychological testing within the CP population should be adopted. Not all children with CP have visual perceptual issues but this should be considered in this population.
• Profound visual deficits and/or profound additional additional disabilities

• Assessment
• Knowing what to
• look for
• Thorough understanding of visual and other consequences of cerebral visual impairment sets the stage for a comprehensive evaluation by members of the educational and medical team and the development of appropriate interventions
• **Uncovering Learning Issues**
• **Collaborative Approach to Assessment and Intervention**
  • Systematic, Multidisciplinary and Multidimensional Assessment
  • Evaluate possible consequences of both ocular and brain-based visual impairment
  • Consider possible attendant disorders
  • Look at vision within the context of the whole child
  • Pinpoint areas of strength as well as challenges

• **Multiple Approaches to Assessment**
  • Triangulation of Data: A Research Method that Can be Applied to the Assessment of Children who have CVI
    • Using multiple data sources in an investigation to produce understanding that captures different aspects of the same phenomenon.
    • There may be inconsistencies across data sources. Such inconsistencies are a source of strength as they provide an avenue to explore deeper meaning in the data.
    • For children with a complex condition such as CVI, using multiple methods of investigation is a reasonable course of action because at this time we don’t have one system that evaluates all possible factors associated with CVI.
    • Requires a variety of professionals who look at the condition from different but critical perspectives

• **Multiple Approaches to Assessment**
  • Example: viewing a specified object in a distant scene: is lack of recognition due to visual acuity, contrast sensitivity, visual field, visual complexity, visual attention, not understanding the directions, too many sensory distractions.
  • Example: different results for visual acuity in clinic vs. classroom: is it environment, familiarity of evaluator, attention issues, behavioral state, type of target.

• **Universal Design for Learning**
  • Inclusion of Children with Cerebral Visual Impairment
    • Many children with cerebral visual impairment are included regular education.
    • The goal is for children to participate as fully as possible with general education peers. (Cheetham, 2014)
    • Children with CVI require access to environments, materials, and methods to reap the full benefit of inclusive settings including full inclusion.
      1. Cheetham, N. (2014). To explore how children with cerebral visual impairment can effectively be included within a mainstream setting, University of Portsmouth, School of Education and
• **Universal Design for Learning**
  
  • **Universal Design**
  
  Design with sufficient flexibility to meet the needs of all users, both abled-bodies and those who have disabilities
  
  Everyone benefits from approaches that address the diverse needs of the whole population


• **Universal Design for Learning**
  
  • **Environments (some examples)**
  
  Visual clutter is minimized in classrooms, hallways, specialized areas such as the school cafeteria and auditoriums
  
  Auditory distractions are minimized
  
  Acoustics are maximized for understanding speech from more than one person at a time
  
  Children with CVI have access to safe and inclusive play yards to accommodate any inability to see fast moving objects and multiple elements at one time
  
  Travel routes throughout school are clear and easy to follow with destination landmarks clearly visible and distinct

• **Differentiated Instruction for Optimal Access to Learning**
  
  • **Methods and Materials as well as Environmental Supports**
  
  Learning options and instructions are implemented that meet individual learning styles and interests for all children in inclusive settings and special setting

• **What are some possible accommodation requirements in this area for children with CVI that must be considered?**

• **Differentiated Instruction for Optimal Access to Learning**
  
  • **Sample factors to consider in differentiated instruction**
  
  • Use of methods that help focus visual attention to promote visual inspection
• Access to perceptible visual input that takes into account requirements related to size, color, contrast, movement, complexity of form, spacing of material
• Access to perceptible auditory input that considers the pace of input, modulation of speech, reduction of competing auditory stimuli
• Provision of support to children when working in collaborative groups at different age levels
• Implementation of creative technology solutions that focus on the highly specific needs of all children including those with cerebral visual impairment

• Role of Teacher of Students with Visual Impairments (TSVI)
• From a new position paper from AERBVI
• Interpret medical reports related to visual impairment
• Conduct a Functional Vision Assessment (how a child uses his or her vision
• Complete a Learning Media Assessment
• Vision specific assessment within typical activities
• Recommendations to optimize visual skills in cooperation with other professional within child’s educational plan
• Translate assessment findings into educational instruction and adaptations in typical settings for a child and for the selection of appropriate literacy media, methods and materials


• Entitlement to Services
• Ensure that all children who have CVI receive appropriate educational services even when they do not fall within service guidelines based on acuity and field

• Eligibility Criteria
• Parameters for entitlement to services have not been universally established
• Some criteria involve discrete measures of visual acuity and/or visual field. These do not incorporate the current understanding of visual impairment due to damage to the visual brain.
• Other criteria involve defined levels of task performance for eligibility

• Entitlement to Services Related to Visual Impairment
• OSEPs memo of 5/22/17 re: Eligibility under Individuals with Disabilities Act
• Written in response to a query about convergence insufficiency:
• Any impairment of vision provided that such impairment, even with correction, adversely affects a child’s educational performance. The term includes both partial sight and blindness
• States must not narrow the definitions in the IDEA; This therefore includes any impairment of vision regardless of significance or severity
• Basing consideration for special education and related services on the presence of designated criteria or physical condition is inconsistent with the IDEA

• **Entitlement to Services Related to Visual Impairment**
  • OSEPs memo of 5/22/17 re: Eligibility under Individuals with Disabilities Act
  • No single measure or assessment may be used as the sole criterion for determining whether the child is a child with a disability and for determining an appropriate educational program for the child
  • In conducting the evaluation, the public agency must use a variety of assessment tools and strategies to gather relevant functional, developmental, and academic information about the child that may assist in determining whether the child is a child with a disability and the educational needs of the child. This can include information from a physician if determined appropriate

• **Entitlement to Services Related to Visual Impairment**
  • OSEPs memo of 5/22/17 re: Eligibility under Individuals with Disabilities Act
  • The group of qualified professionals and the parent must, upon information from a variety of sources, including aptitude and achievement tests, parent input, and teacher recommendations, as well as information about the child’s physical condition, social or cultural background, and adaptive behavior
    • Such evaluations should include a data-based media assessment, be based on a range of learning modalities (including auditory, take, and visual), and include a functional vision assessment

• **Entitlement to Services Related to Visual Impairment**
  • OSEPs memo of 5/22/17 re: Eligibility under Individuals with Disabilities Act
  • In a prior memo: An assessment of a child’s vision status generally would include the nature and extent of the child’s visual impairment and its effect on the child’s ability to learn to read, write, do mathematical calculations, and use computers and other assistive technology, as well as the child’s ability to be involved in and make progress in the general curriculum offered to nondisabled students
  • In addition, because the evaluation must assess a child’s future needs, a child’s current vision status should not necessarily determine whether it would be inappropriate for that child to receive special education and related services while in school
• **Service Needs to Move Forward**
  - Institute methods to identify children in all three subgroups as early as possible, particularly those children with seemingly subtle manifestations of cerebral visual impairment
  - A medical professional needs to make the diagnosis of CVI, but the way the behavioral manifestations for a particular child requires a multidisciplinary team

• **Potential Members of Multidisciplinary Teams**
  - ophthalmologists
  - optometrists
  - pediatric neurologists
  - neuro-radiologists
  - teachers of visually impaired
  - orientation mobility specialists
  - occupational therapists
  - pediatricians
  - physical therapists
  - psychologists/neuropsychologists
  - speech-language pathologists
  - caregivers
  - teachers/ specialty teachers

• **Service Needs to Move Forward**
  - More research on the effects of a variety of interventions addressing the multidimensional needs of all children with cerebral visual impairment and the development of evidence-based curricula for this population

• **Service Needs to Move Forward**
  - More valid and reliable assessment tools developed specifically for children with cerebral visual impairment that address the full spectrum of behavioral consequences of cerebral visual impairment
  - Establishment of school environments conducive to the education of children with cerebral visual impairment – universal design implications

• **Service Needs To Move Forward**
  - Dissemination of information to medical practitioners, a range of school staff, policy makers, and to families about cerebral visual impairment
  - Appropriate transition programs within adult services for young adults with cerebral visual impairment
• Intelligent use of existing resources to meet the needs of children with cerebral visual impairment
  - **Service Needs To Move Forward**
  • Increased pre-service and in-service training among various disciplines and professions within those disciplines that address
  • wide array of consequences of cerebral visual impairment
  • how this impacts learning and development
  • how to assess children with cerebral visual impairment
  • appropriate interventions for these children

• **How Can Optometry Help?**
  **History with Low Vision**
  • Optometry propelled low vision services in the schools forward in the 20th century in the US along with the groundbreaking word of the esteemed educator, Dr. Natalie Barraga
  • William Feinbloom and Arthur Kestenbaum led the way for optometry to take a major role in low vision in the mid 20th century, and low vision clinics opened and increased their reach in the late 20th century
  • Low Vision Clinics opened and increased their reach and low vision evaluations have become the norm for children who have visual impairments with functional vision in the schools

• **How can Optometry Help?**
  **Another Turning Point**
  • What is Optometry’s role in diagnosing, assessing and determining interventions for children who have cerebral visual impairment?
  • How can Optometric methods be applied and expanded to provide a deeper understanding of the complex manifestations of this condition?
  • How can Optometry coordinate services with schools and families?
  • How best to train Optometry personnel to address this population?

• Thank you!

• Please remember to complete your session evaluations online.