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### **Navigating Cerebral Visual Impairment in Children**

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#### **Introduction**

Imagine, you are a new clerk about to start your Pediatrics rotation. You are so excited to be joining the Developmental Pediatrics team this week in clinic. To prepare, you have reviewed the developmental milestones and are eager to see your first patient – Quinn. The astute clerk that you are, you came in extra early to read Quinn's file. He is a busy 2-year-old boy who has been followed by the Developmental Pediatrician since birth when he was diagnosed with cerebral palsy. His parents have booked an appointment today because his mom has noticed that Quinn fails to make eye contact with her when they interact. She states the Quinn's eyes are always looking around and he can't focus on objects if the room is busy. Mom does note that Quinn is very fond of his red fire truck, and other red items. When he plays with it, she notices his eyes light up and they will track the truck as it moves.

You are excited but nervous! The curious case of Quinn is not one you have ever encountered before! Nor are any of mom's concerns found in your developmental milestones table. What are you to do?

Welcome everyone, my name is Josie, and I am a third-year medical student at Queen's university in Kingston, Ontario. Today, I will be helping you develop an approach to assessing and managing children like Quinn, those with suspected cerebral visual impairment or CVI. This podcast was made in collaboration with Dr. Dawa Samdup, a Developmental Pediatrician from Queens University. The objectives of this podcast are to:

- 1) Define cerebral visual impairment or what I will be referring to as CVI
- 2) List the potential causes of CVI
- 3) Discuss the presentation and diagnosis of CVI in children
- 4) Review the potential impact of CVI on development and learning

#### **First, we will be taking about the definition of CVI**

Vision plays a crucial role in child development as it impacts all areas of growth and learning. From acquiring knowledge and understanding the environment to developing motor skills and social interactions, vision is fundamental in shaping a child's cognitive, emotional, and physical development. Now, let's delve into the anatomy and physiological aspects of vision. The journey begins with the eyes that take a "picture" of an object, where light travels through the cornea and passes through the lens to focus on the retina. The photoreceptors (rods and cones) detect the light and transmit electrical signals through the optic nerve, which synapses on the lateral geniculate nucleus (LGN) in the thalamus – the relay station of the brain. The visual information is then organized in the LGN before being sent to the retro-geniculate pathway, and

finally off to the primary visual cortex. The visual cortex relies on two streams, the occipital temporal area known as the ventral or “what” pathway that recognizes objects, faces, and processes information such as color, shape, texture, and size. The occipital parietal visual area known as the dorsal stream or “where” pathway tells you where objects are and what to do next with that object. For example, the ventral pathway sees an apple, and the dorsal pathway recognizes it’s on a shelf and you should reach for it.

The visual pathways are complex, so to help organize this journey, we can simplify this into the anterior pathway (the eye) and posterior visual pathways (the brain). Visual impairments that are a result of disruptions within the brain are referred to as cerebral visual impairment (CVI). As a result of these brain-based impairments, an individual living with CVI may have altered visual acuity, visual fields, and visual attention<sup>1-3</sup>. These individuals may also face challenges with visual-motor and visual-spatial coordination, but this will vary based on the cause of CVI and what area of the brain is affected. Notably, many children with CVI have no pathology occurring in the eye<sup>1-3</sup>, while others have refractive errors or misalignment that may mask their CVI.<sup>1,5,6</sup>

As you continue to read Quinn’s file, you note that he was seen by a Pediatric Ophthalmologist who found no abnormality with his eyes. Where do you think Quinn’s visual problems may lie? If you guessed within the brain, you are correct!

With an understanding that Quinn may have CVI, you – the well-prepared medical student- check UpToDate to learn more about CVI. But you are surprised because nothing comes up! Despite being the leading cause of childhood blindness and low vision<sup>4</sup>, there is little information on CVI, which may be attributed to the lack of a universally accepted definition<sup>2,5</sup>. In 2022, however, the *American Association of Pediatric Ophthalmology and Strabismus* described CVI as “*resulting from a decreased visual response due to a neurological problem affecting the visual part of the brain.*” They also went on to state that “*A child with CVI has a normal eye exam that cannot account for the abnormal visual behavior.*”

To recap, in an individual with CVI, their eyes are normal, or near normal, so the “picture” taken travels to the brain but is not properly processed or integrated by the brain due to abnormal brain function. This seamlessly brings us to our second objective: Exploring the various causes of CVI.

### **Causes of CVI**

CVI is caused by any process that damages the visual parts of the brain, such as prematurity, stroke, decreased blood supply, brain malformation, hydrocephalus (increased pressure in the brain), seizure, metabolic disease, infection, head trauma<sup>6</sup>. Of note, in children with hypoxic ischemic encephalopathy, approximately 60% have CVI<sup>7</sup>. CVI can also co-occur with many conditions such as cerebral palsy, Down syndrome, Rett syndrome and several genetic variants<sup>4</sup>. The prevalence of CVI has increased sharply, likely due to increased survival rates of children who sustain severe neurological conditions during the perinatal period<sup>1-3</sup>.

### **Presentation and Diagnosis of CVI**

And there we have it, Objective 3: Diagnosis of CVI. Despite the increasing prevalence of CVI<sup>8-10</sup>, the diagnostic process is complex, and relies on a multidisciplinary team of ophthalmologists, healthcare professionals, therapists, educators, and parents<sup>1,9,11</sup>. These specialists bring a diverse range of training and expertise to their understanding of CVI, enabling them to provide a tailored approach to a child’s specific needs. Nonetheless, CVI is

often underdiagnosed and misunderstood. According to new research conducted by the Perkins School for the Blind with analysis from McKinsey & Company, less than 20% of children with CVI have been diagnosed<sup>4</sup>. Several factors contribute to children with CVI going unrecognized and unsupported. First, familiarity with CVI among providers is limited. Next, availability and accessibility of support remains an obstacle for many families. Finally, other social risk factors such as poverty exacerbate these obstacles, leading to further gaps in diagnosis, support, and eventual achievement<sup>4</sup>.

Every child's visual journey with CVI is unique, and a diagnosis is often made by ruling out other causes. There are, however, common visual and behavioural characteristics that may help determine if a child has CVI<sup>6</sup>:

- **Latency:** A child may take a long time to look at an object to understand what they are seeing.
- **Visual motor:** A child will not reach for an object or looks away when reaching.
- **Complexity:** Occurs if there is difficulty seeing when a background is complex or cluttered and in a busy environment.
- **Visual field:** A child might not respond to items on one side of their visual field. Lower field visual loss is common in children with periventricular leukomalacia (PVL) i.e tripping and falling on stairs.
- **Movement:** A child may have difficulty seeing stationary objects, needs a viewer or the object needs to be moving. They might also need an object to move to know it's there or they might have trouble assessing distance and speed, such as a ball or car coming toward them.
- **Color:** Many kids with CVI prefer high contrast, one color that is saturated over another, like Quinn preferring red objects.
- **Light gazing:** Children with CVI may stare at light or light sources and find it difficult to redirect attention. Lights should be used to accentuate or highlight the item.
- **Novelty:** A child with CVI may prefer familiar items over novel items and may ignore something that is new.
- **Visual reflex:** Visual blink or threat response may be absent or atypical.
- **Distance viewing:** Some children may have difficulty with distance viewing and may hold objects close to their eye to reduce visual complexity.

Again, CVI is a spectrum disorder and not all children will have all these visual behaviours. Remember, children living with CVI have diverse functional vision, which may fluctuate due to physiological, psychological, and environmental factors<sup>1,5,6</sup>.

### **Impact of CVI on Developmental and Learning**

The manifestations of CVI in children is unique. Some may struggle with finding items in a cluttered scene, others may bump into objects or face difficulties copying from the classroom whiteboard to their workbooks, or even exhibit challenges in effectively controlling their eye position to stay focused on tasks<sup>10</sup>. For primary school-aged children, the failure to fully recognize and understand these difficulties may lead to misinterpretations such as a perceived lack of comprehension, clumsiness, inattention, or even social and communication issues, especially if the child has other co-occurring developmental concerns. Children with CVI may be misdiagnosed with attention deficit disorder, learning problems, autism spectrum disorder or Developmental co-ordination disorder.

Thinking back to our patient Quinn, it is easy to see how he may struggle with his fine motor development, such as coloring, drawing, dressing, or cutting with scissors while doing a

craft. Thinking about his transition to daycare or school, which are often crowded and loud environments, Quinn may struggle to focus his vision, and his inability to excel in this busy environment might be attributed to his cerebral palsy. Challenges with these tasks may also impact Quinn in other developmental domains such as his social skills (playing or sharing with others).

To help children with CVI excel in their home/school/community environment, the following approaches may be helpful<sup>6</sup>:

- Use large, high contrast lighting.
- Use touch or sound to attract child's attention.
- Present objects in a simple uncluttered manner.
- Present objects from different directions/angles.
- Allow extra time for responses to visual stimuli.
- Avoid over stimulation.
- Avoid visual tasks when child is hungry, tired, frustrated, etc.

Ultimately, children living with CVI are unique in their presentation and abilities. It is important for health care providers to work as part of a multidisciplinary team to care for children with CVI. Going back to our patient Quinn, you observe the Developmental Pediatrician interact with Quinn and his mother. Quinn can identify objects when they are presented one-by-one on a mat, but when a whole bucket of toys is dumped on the floor at once, Quinn can't seem to find his toy car. When you interacted with Quinn, he was very fascinated by your red name tag, until he got hungry and no longer was able to pay attention to you. The Developmental Pediatrician then discusses with mom the diagnosis of CVI and what it will mean for Quinn. A meeting is set up between an OT, PT, low vision specialist and daycare providers. Quinn will have a therapy plan in place to help him excel and participate.

So, with that, a few things to review:

When do you suspect CVI?

1. When the degree of vision impairment is unexplained by ocular findings
2. Significant pertinent past medical history: brain injury, seizures, prematurity etc.
3. Neuroradiological findings of brain injury +/-
4. Behavioral and visual responses associated with CVI

How do you manage CVI

1. Refer to a Pediatric ophthalmologist.
2. Refer for Blind low vision therapy (Ontario blind low vision Early Intervention Program, BLVEIP 0-6 years), vision loss rehabilitation.
3. Multidisciplinary team: OT, PT, SLP, physicians, orientation/ mobility experts, educators.

### Take Home Points

What a wonderful day you have had caring for Quinn and learning about CVI. To recap what was discussed in this podcast, lets highlight some main points of this podcast.

- 1) CVI is the leading cause visual impairment in children.
- 2) CVI refers to visual impairments resulting from brain damage rather than eye abnormalities.

- 3) The vision impairment caused by CVI may affect a child's gross and fine motor skills, cognitive, language and social interactions, interventions are focused on a child's unique abilities and school accommodations are often required.
- 4) Early identification and appropriate support services are key to optimize developmental potential.
- 5) A multidisciplinary team is required to support a child with CVI

Thanks for listening!

## **Reference**

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