# Occupational Therapy: Cortical Visual Impairment

807155

College of Western Idaho

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Megan Nichols

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# Cortical Visual Impairment

## Condition Description

Cortical visual impairment (CVI) is a lifelong neurological disorder caused by injury to the brain's visual pathways and visual processing centers resulting in abnormal or unique visual responses to people, objects, and the environment (Baker-Nobles & Rutherford, 1995). CVI primarily affects infants and children but can persist throughout development into adulthood. CVI is exceedingly common and is known to be the leading cause of visual impairment in children in the United States and developed countries (Wilkinson & Wolf, 2021).

It is important to note that the name cortical visual impairment can be interpreted as misleading as it can potentially allude the reader to assume it is an eye impairment like visual blindness. In fact, this disorder was once labeled as cortical visual blindness, but that term has since been abandoned for inaccuracy. The actuality is that many children with CVI often have completely healthy eyes and attain eye exams that are typical or nearly typical (Dutton & Lueck, 2015). CVI differs from ocular, eye-based forms of visual impairment in that the interference in visual function exists not in the structures of the eye or the optic nerve, but in the visual processing centers and visual pathways of the brain (Ospina, 2009).

In typical vision, the eyes take a picture of an object, and that message is sent to the brain by way of the optic nerves. The brain recognizes the image and integrates it with other sensory messages (Scott, 2022). The brain then responds to the sensory input by sending a motor response to the appropriate part of the body. In individuals with CVI, the eye takes a normal picture of the object and sends the message to the brain. The message is not properly processed or integrated because of the bilateral damage to the posterior visual pathways to the occipital cortex (Scott, 2022). This damage represents difficulty in processing and interpreting visual

information. This results in characteristics of abnormalities of the visual field, including blind spots, swirling masses of light, and the world around them may appear as a visual kaleidoscope of meaningless color and patterns. Although these children live in the visual world, they are unable to interpret what is seen, and their brains cannot make sense of the visual information. Children with CVI demonstrate multiple developmental delays as all behavior and development are visually driven throughout life. In addition, children with CVI can have problems with visual concentration, depth perception, eye-hand coordination, recognizing objects, focusing on near objects, fast eye movement, and visual field loss (ElMaksound et al., 2016).

According to the National Institute of Health, visual function in CVI is measured across the CVI Range (0-10) (Chang et al., 2022). This range contains three major phases (I-III) describing the level of impact of the CVI characteristics on the individual's ability to use their vision functionally. Phase I (0-3) is the most severe impact on visual functioning. Phase II (3-7) is considered to have a moderate impact on visual functioning, and Phase III (7-10) is the closest to typical visual functioning" (Change et al., 2022). Although CVI is irreversible, therapy can be crucial and important to help individuals diminish CVI. By receiving early intervention and effective therapy, individuals with CVI may improve their functional vision. There is significant importance in identifying a child's specific visual function in the CVI range with regard to each condition characteristic. The range is provided to guide future intervention approaches needed to support progress in visual functioning unique to each phase (Chang et al., 2022). To elaborate, in Phase I building visual behavior is the focus. The therapist will work with the client to consistently start looking at a small range of specific objects. Compared to Phase III, the refinement of CVI characteristics is the main goal. The therapist will work on fine-tuning and

identifying salient visual details and targets that are situated in complex arrays. With proper interventions, CVI can evolve over time, and an individual can show a progression of improvement from Phase I and move systematically into Phases II and III.

**Etiology**

The etiology behind CVI is complex as this damage to the brain may affect not only the primary visual cortex but also the associative areas, such as the optic radiations, optic nerves, and even visual attention pathways (Ospina, 2009).

According to research, perinatal hypoxic-ischemic encephalopathy (HIE) is by far the most common cause of pediatric CVI (Ospina, 2009). To simplify, HIE is a type of newborn brain damage caused by both limited blood flow and oxygen deprivation. Too little oxygen, or hypoxia, disrupts the autoregulation of the major blood supply to the brain, creating too little blood flow or ischemia. The extent of damage from hypoxia-ischemia depends on both the severity and duration of the episode (Chhablani, et al., 2014). If the period of oxygen deprivation is short, the body may recover without long-term effects. If the deprivation of oxygen to the brain is severe, cellular death is the result, and the effects are lifelong. (Chhablani, et al., 2014). According to statistics, in 2018, “HIE was the cause of CVI in 151 of 423 (35.7%) children diagnosed with CVI” (Pehere et al., 2018).

CVI can also be caused by premature birth. Babies who are born prematurely, before thirty-seven weeks, have not had time to fully develop. This has potential consequences for the visual system because these infants are at increased risk as blood and, therefore, oxygen has not reached all parts of the brain (Pehere, 2018). This occurrence can cause the onset of periventricular leukomalacia or PVL. PVL occurs when the white matter adjacent to the lateral ventricles is deprived of oxygen, and the nerves in this area die, becoming soft, and scar tissue

develops (Pehere, 2018). The reason why PVL occurs as a result of premature birth is because a premature baby's lungs aren't yet fully developed. The undeveloped lungs deliver less oxygen and blood flow to the baby's brain, resulting in damage to brain cells.

CVI can also be caused by hydrocephalus. Hydrocephalus is when the cerebrospinal fluid does not circulate properly around the brain, and collects in the head, causing an abnormal build of CSF, ultimately putting pressure on the brain (Pehere, 2018). This build-up of fluid and intense pressure in the ventricles can cause damage to the brain.

The etiology behind CVI can be caused by a variety of additional factors, such as seizures, trauma, TBI, developmental brain defects, chromosome disorders, infections associated with the CNS, and more (Chhablani, 2014). On the occasion, there have been instances where the cause of CVI in a particular patient has been unknown. The etiology behind CVI is very significant. It will help practitioners and professionals better understand a client's personal and unique diagnosis of CVI and better guide future rehabilitation and interventions.

**Prevalence**

As noted earlier, CVI is known to be the leading cause of visual impairment in children ages zero to three, with an estimated 150,000 children diagnosed in the U.S. alone. (*What is it like to have CVI,* 2022*).*

CVI primarily affects infants and children but can persist throughout development into adulthood. The prevalence of children diagnosed with CVI has continued to rise over the past few decades because of advancements in the medical field and technological discoveries. With access to improved neonatal care and medical sciences, the survival rates of premature infants are increasing. With increased survival rates comes consequence as more children born prematurely are likely to be diagnosed with developmental delays, disabilities, and disorders like

CVI. The National Institutes of Health website cites that "approximately 30-40% of children with visual impairments have CVI, and there is a CVI prevalence of 10.5% of all children with developmental disabilities" (*Cerebral Visual Impairment (CVI)* 2020).

It is worth mentioning that although CVI cases are becoming more substantially prominent in the US, many cases go undiagnosed or misdiagnosed on a regular basis (Maitreya, et al., 2018). This is a challenging disorder to diagnose for medical doctors because it is a neurological disorder. Often times, brain imaging studies, like an MRI, may or may not indicate the neurological issue of CVI, resulting in misdiagnoses. Medical doctors must solely rely on previous medical records, birth records, and observations of unique visual and behavioral characteristics in order to diagnose a patient with CVI. Adversities with accurately diagnosing a patient with CVI make the research on the prevalence of this condition limited.

**Prevention**

When understanding the etiology behind CVI, it is recognized that there are no factual preventative measures to prevent the occurrence of this condition. Although this is the case, looking at the bigger picture, it is possible to identify factors that could possibly prevent prematurity and hypoxic ischemia since they are the leading cause of CVI. Unfortunately, with the incidence of premature infants on the rise and over 11% of all births in the US, we are miles from this goal (Good, 2009). Still, it is worth reflecting on the healthcare system and approaches that could reduce conditions that damage infants. Premature birth can affect any socioeconomic group, but lower socioeconomic status confers significant risk. This is due to poor access to prenatal care. Ultimately, health care policies that increases access to medical care could have a significant impact on the incidence of all diseases associated with prematurity, not simply CVI.

The cooling protocol is also an innovative treatment for hypoxia, one of the leading causes of CVI (Good, 2009). This treatment involves the reduction of the body temperature in newborns who meet specific criteria associated with hypoxia. According to a research article titled, Cortical Visual Impairment: New Directions, "the infant's body temperature is reduced for 72 hours and then gradually rewarmed to best reduce the effects of reperfusion injury associated with restoring the blood flow after severe hypoxia" (Good, 2009). This approach is based on the concept that inflammation plays an important role in the spread of CNS injury after an initial insult to the brain, and that inflammation can be reduced by lowering tissue temperatures (Good, 2009). Although there is no cure for CVI and limited preventative measures, individuals with CVI have the potential to improve their functional vision by receiving therapy. Children and infants with CVI require early intervention, occupational therapy, vision rehabilitation, educational support, and other special services to help them develop and learn (*Cerebral Visual Impairment (CVI)* 2020).

## Short-Term Goals and Occupational Therapy Interventions

For this part of the assignment, I have chosen to base my short-term goals and interventions for a CVI client that shows characteristics on the CVI Range of a Phase III. I have chosen a Phase III CVI client for the fact that a prominent majority of Phase I and Phase II clients require the use of Augmentative and Alternative Communication (AAC) for daily activities, communication, and interventions. Since the use of AAC's are something I am not yet accustomed to currently, I contemplated that with the preexisting knowledge I have gained thus far as a student, I could create and execute a better intervention with a client that is in Phase III of the CVI Range.

### Short-Term Goal A

To help with navigation of daily environments, visual tracking, distance viewing, and figure foreground discrimination, the client will display the ability to correctly seek and find 7 out of 10 pre-determined sequences while walking down a hallway and scanning a busy visual field 3 out of 4 trials for 5 consecutive sessions.

**Intervention A: Where are the Animals? Zoo Escape**

For intervention A, regarding the short-term goal, I will implement an activity that will work towards improving the client's skills in visual tracking, distance viewing, and figure foreground discrimination. These listed characteristics are described as challenging for CVI clients in Phase III. For the short term-goal, I will administer a "Where are the Animals? Zoo Escape" activity. For this activity, the client would walk around the outpatient clinic hallway and have to find a list of zoo animals in order. Materials needed for this activity include cut-out pictures representing ten zoo animals, string lights/fairy lights, a cardboard box decorated to portray a mini zoo, and an adaptive reference sheet of the printed-out animals. This activity encourages the occupation of play, particularly pretend play, while still working on functional skills needed for everyday use. This intervention will help the client to be able to look at a visual field with overlapping visual features, visually cluttered backgrounds, and a complex array of objects to target the predetermined sequences.

1. To prepare for this intervention, cut out pictures representing ten zoo animals. Next, glue or tape the string lights on the back of the animals in order to provide a slight illuminance factor to the cutouts. For better structure and identification, create a two-dimensional object by gluing the animal cut out and lights to the back of cardboard painted in the child's preferred color (blue, yellow, red, etc). Creating this type of structure will help the child target and

locate the animals without getting overstimulated from the complex visual field. Then, craft a cardboard box to portray a zoo cage to encourage pretend play.

2. To finish preparing for this intervention, create an adaptive reference sheet of the animals. To do this create a booklet containing enlarged photos on the animals presented in this activity that the child can refer to. Make sure to include salient features listed on the side so the therapist can further assist in helping the child identify and differentiate the animal pictures. The final step is to place the luminated animals around the hallway for the child to find. It is important to remember that children with CVI Phase III continue to have difficulties with lower visual field function, so adequately placing the animals at eye level or above will make this intervention more successful (Roman-Lantzy, 2018).
3. To start out this activity I would explain to the client that all of the animals in the zoo escaped. I would go on to explain that it is our job to find the "hiding animals" throughout the clinic hallway and return them to the zoo. It is important that the child is familiar with the environment and location of this activity for both safety reasons and to help regulate emotional behaviors.
4. Hand the reference book to the client and describe the salient features of each animal. For example, the therapist will communicate salient features of an elephant such as, big floppy ears, really long nose, very large animal, sharp white tusks, gray, etc. This helps the individual with CVI continue to develop the ability to discriminate greater degrees of details associated with image, symbol, and distance viewing (Roman-Lantzy, 2018).
5. Have the client walk throughout the hallway visually scanning for animals and collecting the animals when found. According to research, distance viewing for an individual with Phase III CVI exists beyond ten to fifteen feet (Roman-Lantzy, 2018). Help the client place the

animals back into the zoo when completed. Provide assistance and verbal cues for moving through the environment as needed. Also, be aware of visual latency and provide enough time for the child to be able to generalize or locate the picture of the animal.

This activity works on the main skills of improving visual complexity, distance viewing, and visual scanning. To support the improvement of these skills, this intervention provides adaptive techniques by providing luminance, color preference, and contrast factors. CVI clients in Phase III often continue to have difficulty with lower visual field function and may require the addition of color and backlighting to perceive details in complex presentations (Roman-Lantzy, 2018).

According to an evidence-based research article titled Effects of Visual Rehabilitation on a Child with Severe Visual Impairment, manipulating the luminance, color, and contrast components of training materials (toys, objects, etc.) during play activities resulted in both motivation from the client and improvement of functional vision (Tsai et al., 2013). This article goes on to describe that "because play is a functional task for children, it can facilitate and modulate children's motivation and attention during interventions, further enhancing the visual learning effect" (Tsai et al., 2013). It is expected that the use of CVI-adapted toys or objects during play will allow the client to be more likely to translate their new learning abilities to their daily activities. The hope for this intervention is that with repetitive task practice, a client with CVI will gain new connections and improve brain plasticity for future functional performance. Improving visual complexity, distance viewing, and visual scanning with the use of targeted luminance, contrast, and colored objects will help a client navigate more efficiently through everyday life. This intervention will be helpful for future experiences because children with CVI

often have a difficult time locating the restroom sign, exit sign, get lost easily, and navigating through their environments.

**Short-Term Goal B**

Client will actively engage in visually guided reach of both preferred and nonpreferred items when given one verbal cue with 85% accuracy for 3/4 trials for 5 months.

**Intervention B: Salient Features Go Fish**

For this intervention, my goal is to improve on the client's visually guided reach, generalization of everyday objects, and the use of salient features to identify targeted objects. This game is a fun twist on “Go Fish” that is designed especially for those that are visually impaired or those that have CVI. I give credit to the website titled Strategy to See for this activity idea for those individuals that have Phase III CVI (Sheline, 2020).

As expressed earlier in this paper, salient features describe the prominent features of an object, picture, or person, and help a client distinguish one target from another. Silent features are used with CVI clients often as it helps them to better understand what they are visually perceiving. Materials needed for this activity are homemade cards cut out from thick stock paper. I would recommend using black stock paper as this will create a better contrast to support a child's vision needs with CVI. Also needed for this intervention are cut-out pictures of an assortment of everyday items that a child uses. This could be their preferred items, grooming products, eating utensils, food, clothing, etc. The last detail needed for this intervention is the matching 3-D object correlating to the cut-out pictures.

1. Glue the cut-out pictures of items and place them on the homemade black cardstock cards. Create enough cards to have about at least 20 matching cards for the deck.

2. Locate 3-D objects that will correlate to the pictures posted on the cards. Place down a black tablecloth on the tabletop to again create contrast between the different 3-D objects and the 2-D cards.
3. Next, both the therapist and the child will collect 4 cards each. If it is easier for the client, place all of the cards face up in front of the client to create contrast from the black tablecloth. Give them time to process all of the pictures in front of them. It is important to remember that those with CVI have visual latency that might require additional time for their brains to process what they are seeing.
4. The therapist will then choose a card out of their hand and start describing the salient feature descriptions. To describe a spoon in this game, the therapist could ask the client, "do you have a card that is oval, long, skinny, and has a curved part at one end?". The client will then attempt to locate and visually reach for the appropriate card that describes a spoon. This may require hand-under-hand support, or verbal cues to locate and reach for the correct object.
5. The 3-D objects correlating to the cards come in handy to further review the salient features associated with a particular object. This better helps the client recognize cards displaying this object (Sheline, 2020). For example, use a 3D ball to review the salient features associated with the object to better help the client recognize the card displaying this object.
6. If the client is showing success in this activity, have them describe the salient features of their cards to the therapist, in order to make a match.
7. The winner is the one who gets to a certain number of matches.

It is my belief that visually guided motor responses will be improved in this intervention. Although we are implementing a goal for increased visually guided reach, we are using the occupation of play as a medium for this intervention. Implementing a fun game similar to "Go

Fish" into the occupational therapy session will motivate the child to engage in the activity, all while unknowingly working on function skills to improve visual processing.

Visually guided reach is a critical skill for everyday functional activities, such as reaching for a spoon, a cup, toys, or for communicating. Adaptations based on a client's CVI Phase should be used to promote increased visually guided reach (Roman-Lantzy, 2018). An example of an adaptation I used in this activity included reducing the visual complexity of the array of objects and backgrounds in front of the client. I used this by creating a black contrast on both the cards and the table. This allows the client to focus on the cards/object and not get overwhelmed or fixated due to a visually complex background. Techniques that use a simplification of the visual environment, high contrast color, common objects, integration of touch and vision, are stressed for therapists working with children with CVI (Sheline, 2020).

According to the article titled, Understanding Cortical Visual Impairment in Children, using a solid color to create contrast in order to promote visually guided reach for play was used in this study (Baker-Nobles, et al., 1995). It explains that reaching for an object was encouraged with the provision of tactile cues when a child was visually engaged (Barker-Nobles, et al., 1995). This proves that simplifying the visual complexity of an environment will improve the probability of visual guided movement towards an object.

It has been identified throughout this paper that visually guided reach is a challenge for individuals with CVI. This is simply because neurologically they have a difficult time processing visual information, so attempting to make motor movements seems impossible. Although this is the case, it has been proven that individuals with CVI are able to accurately complete the task of reaching for an object while looking. According to the research article titled Cerebral Visual Impairment and Clinical Assessments, children with CVI tend to use their peripheral vision to

look at an object and appear not to use their central vision (Ortibus et al., 2019). It quotes, “children with CVI often lean forward to bring their eyes close to an object to reduce the “crowding” effect or to improve resolution” (Ortibus et al., 2019). The article also explains the importance of good appraisal of the visual stimulus for children with CVI. It states that the use of good lighting, optimal contrast, and a clutter free environment can increase the overall outcomes and motivation of visually guided reaching as well as being able to understand the defining elements that distinguish one object from the other (Ortibus at el., 2019).

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