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Physical Therapy Management of a Patient with Cortical Visual Impairment to Improve Functional Mobility: A Case Report

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Abstract

Background: Cortical visual impairment (CVI) is the leading cause of visual impairment in developed countries. CVI is caused by any process that damages the central visual pathways in the brain, including stroke, seizure, hypoxia, hydrocephalus and other neurologic disorders. Individuals with CVI oftentimes have a normal eye exam, making early detection difficult. **Purpose:** The purpose of this case report is to present the multi-dimensional treatment approach to a patient with cortical visual impairment and abnormal gross motor development, as well as the challenges associated with evaluating progress. **Case Description:** The patient was a 5-year-old female with a medical diagnosis of paraventricular leukocytosis, resulting in cerebral palsy. During the case report period, the patient scored at level 4 on the Cortical Visual Impairment scale, meaning that she consistently responded to visual stimuli. The patient required hand-hold assist for ambulation, and maintained a crouched posture in standing. **Interventions:** Both visual tracking and ambulation training were utilized to improve the patient's functional mobility and independence. **Discussion:** During the case reporting period, the patient demonstrated improvements in visual tracking, as well as postural stability in static stance and during ambulation. Multiple outcome measures were utilized, including the GMFM-88, PDMS-2, and a Functional Visual Evaluation. Due to the numerous confounding factors that play into a pediatric patient's progress, it is difficult to produce a cause and effect relationship between outcome and intervention.

Background

Cortical visual impairment (CVI), also referred to as cerebral visual impairment, is defined as a visual impairment due to brain damage. It is the leading cause of visual impairment in developed countries, and is on the rise in developing nations as well.¹ CVI is caused by any process that damages the visual pathways of the brain, including stroke, seizure, hypoxia, infection, hydrocephalus, and other neurologic disorders.^{1,2}

Patients with CVI often have a normal eye examination, making it difficult to detect early in life.² Another difficulty when diagnosing CVI is the variable presentation, and the large spectrum of visual disabilities that fall under the umbrella term.³ While there is a vast range of behavioral characteristics associated with CVI, the most common characteristics, as listed by Roman-Lantzy et al., are as follows: a distinct color preference, an attraction to movement, visual latency, visual field preferences, difficulty with environmental complexity, light-gazing, absent or atypical visual reflex responses, difficulty with visual novelty and absence of visually guided reach.¹

Early recognition of CVI is oftentimes critical due to neuroplasticity of the juvenile brain. Collignon et al. established that there is cross-modal plasticity when vision is lost early in life. There is a sensitive period for auditory spatial processing in the occipital cortex when the brain is devoid of visual stimulation.⁴ Additionally, lack of visual projections to the cerebellar vermis and cortex can lead to delays in control of balance, leading to postural instability that can sustain into adulthood.⁵ Therefore, it is imperative to rehabilitate the visual system in order to preserve visual function in the occipital cortex.

There is evidence that CVI has the capability to improve over time. Huo et al. compiled data of 170 patients with CVI seen in pediatric ophthalmology over a 15-year period and found that 60.4% of the patients demonstrated some improvement in vision, 37.5% demonstrated no change in vision, and only 2% demonstrated a decline in vision.⁶ Huo et al. also found that patients diagnosed with CVI that were older than 3 years of age had less improvement in vision than patients who were diagnosed at an earlier age.⁶ Watson et al. described a critical period for development of acute vision between birth to 10 months, followed by a period of slower development from 10 months to 10 years.⁷ Additionally, research supports that in the first 2 years of life, neuro-connectivity increases rapidly, and connections are double that of an adult. After 2 years, considerable synaptic pruning occurs and connections that are not used are lost. This synaptic pruning pertains to vision as well.^{7,8} Early studies performed by Hubel and Wiesel on young cats demonstrated that visual deprivation during a critical period of visual development leads to permanent damage to vision.⁸ While there is no consensus on a time-frame surrounding cortical plasticity, there is evidence that the greatest opportunity for improvement in vision occurs early in life.^{1,6,7,8} There is a limited amount of time to expose children to visual stimulation to develop their visual function in the most optimal way.

The American Foundation for the Blind reports that approximately 30%-40% of children with a visual impairment have CVI.¹ Additionally, the prevalence of CVI in children with developmental disabilities is 10.5%, making it imperative to screen for visual impairments in pediatric physical therapy.⁹

Developmental disabilities are often accompanied by CVI, and pediatric physical therapists will need to address the visual component while managing a patient with CVI. Visual stimulation exercises help encourage visual processing and discrimination, visual perception, visual input, concentration, focus, visual awareness, hand-eye coordination, and encourage social development.¹⁰

There is a scarcity of literature supporting the physical therapy management of CVI, with much of the research focusing on the cognitive visual dysfunction of the injury.¹¹ The purpose of this case report is to highlight the incorporation of visual stimulation exercises, not typically thought of as physical therapy interventions, into the rehabilitation of a patient with cerebral palsy (CP) and CVI. This paper will discuss the challenges associated with documenting progress in a complex pediatric patient, and lay out the multidimensional treatment approach used to progress towards functional independence in a young patient with cortical visual impairment.

Patient History

The patient was a 5-year-old female who was first seen at Genesis Pediatric Therapy in 2013 at 13 months of age. She presented with the diagnosis of developmental delay, and paraventricular leukomalacia after contracting meningitis at one week of age. The patient also had a medical history including myocarditis and an atrial-septal defect. The patient was born at 30-1/2 weeks gestation. At the time of her initial evaluation in 2013, the patient held her eyes in a medial position bilaterally and was wearing an eye patch to correct for medial gaze palsy. The parents were primarily concerned that the patient was not sitting, crawling or walking independently. Prior to her evaluation at Genesis Pediatric Therapy, she was receiving therapy through another pediatric therapy service in the area.

Initial Examination and Evaluation

During the initial evaluation in 2013, the patient had heightened muscle tone, especially in her lower extremities. She was unable to roll between prone and supine positions, she demonstrated no initiation to transition to sitting, and had no interest in initiating movement towards toys. The patient required support at the thorax to maintain sitting balance, but was able to maintain her head at midline. She sat with a kyphotic posture, and had increased lumbar lordosis and bilateral knee hyperextension in supported standing. The patient demonstrated no form of functional mobility at the initial evaluation. She had bilateral supportive reflexes, impairing her standing balance. A Functional Vision Evaluation (FVE) was also performed. The patient was placed in a dark room, and colored lights were used for visual contrast. Upon initial evaluation, the patient was only able to perceive a red colored light when it was presented directly in front of her, centrally. She did not demonstrate visual tracking in any direction, regardless of color presented. The patient did not blink to touch and did not have a threat blink reflex. The patient did have a pupillary constriction reflex.

Re-Evaluation

During gait analysis in July 2017, the major concerns were as follows: the patient demonstrated scissoring gait, hyperextended lower extremities, and minimal heel contact throughout the gait cycle. She required 2 hand-hold assist for ambulation. The patient's functional vision was re-tested in December 2015 after receiving physical therapy at Genesis Pediatric for both functional vision and gross motor development, and again in August 2017 during this case reporting period. The following is a compilation of the patient's visual processing behavior collected by Maria Teresa Ferrer, PT,DPT,C/TBI, C/NDT and classified by Roman-Lantzy.¹

Color:

The patient did not rely on the color red, as she did upon initial evaluation. She was able to view objects with four different colors and complex patterns.

Movement:

2015: The patient did not require movement, but sound cues helped gain the patient's interest.

2017: The patient was able to follow moving objects as long as the object is within 100 cm and no higher than 85 cm from the ground.

Visual Fields:

2015: The patient was able to perceive objects placed at midline at a distance of up to 90 cm

2017: The patient had improved peripheral field vision and was able to perceive objects that were placed in midline at a distance of up to 100 cm.

Complexity:

2015: The patient did best with simple images and colors. Visual attention increased with multiple colors used in a dark environment

2017: The patient was able to attend to complex images and colors. Visual attention increased with multiple colors used in a dark environment.

Distance Viewing:

2015: Did not extend past 90 cm

2017: Did not extend past 85 cm

Visual Reflexive Responses:

Consistent response to reflexes during both testing periods

Visual Novelty:

2015: Visual attention to new objects was an issue, especially in unfamiliar environments

2017: Visual attention to new objects or people improved, preference to follow musical voices

Visual Motor:

The patient's ability to look and reach at the same time was impacted by her balance in both testing periods. There were no changes noted in this category. When support was withdrawn, the patient was apprehensive. With support, she was able to look and reach in quick sequence.

Based on the patient's results during the visual assessment, the patient scored at Phase 2 in December 2015, meaning that she was able to briefly fixate on an object in a controlled environment, but did not demonstrate appropriate attention. She progressed to Phase 4 during testing in August 2017, meaning that she functioned with more consistent visual responses to surroundings.

Interventions

The patient was seen three times a week for 40-minute physical therapy sessions, occupational therapy and speech therapy from 13 months of age to present. Her current physical therapy sessions focused primarily on functional mobility, as well as visual attention to the environment. At the start of this case study, in July 2017, the patient's preferential mode of ambulation was crawling. The patient utilized a reverse walker at home and at school, and she was beginning to pull to stand and cruise on furniture.

The human balance system is comprised of the vestibular, somatosensory and visual systems. When any of the three components of this system are altered, balance can be compromised.¹² Spasticity can also alter how a person reacts to perturbations to balance.¹³ Since the patient in this case report had both compromised vision and irregular muscle tone, both factors contributed to the patient's decreased stability during functional activities. Additionally, the patient would startle frequently, leading to a further increase in tone, resulting in a loss of balance. When standing and ambulating, the patient would maintain cervical and trunk flexion causing decreased visual attention to the environment and loss of balance forward.

Visual Stimulation Training: The patient was placed in a dark room, with visual targets used to promote the use of visual pathways. This improved postural orientation by encouraging proper head alignment. During this activity, the patient would initially perform static stance, either against the wall or with moderate support at the lower extremities to maintain balance. The patient's vestibular system was engaged by moving the lights both horizontally and vertically, so the patient was forced to rotate her head to track the lights. The lights were first placed close to the patient, approximately 6 inches away, so the patient would fixate on the visual stimuli. The lights were advanced further until the patient was no longer able to consistently fixate on, and track the light. Since the patient responded strongly to auditory stimuli, the person holding the lights was instructed not to speak so that the patient would use only vision to find the stimulus. This activity was meant to stimulate the patient's visual pathways and promote postural awareness, since the patient had to bring her body to vertical to find the light. While the patient was in supported stance, the patient was instructed to reach out and touch the visual stimulus to promote visual-motor integration and spatial awareness. To turn visual tracking into a functional task, the patient was positioned at one side of the dark room and was instructed to walk

forward toward the visual stimuli. This activity promoted forward gaze and increased trunk and cervical extension, since the patient was to fixate on visual stimulus during ambulation.

Ambulation Training: Since walking independently was a primary therapeutic goal for the patient, much of her therapy consisted of ambulation training. The patient became apprehensive with decreased support from the therapist and would reach for additional tactile input, causing a loss of balance. To combat the patient's need for support, she was given a single elastic cord support that she could utilize for proprioceptive input, which would stretch when she applied weight. This forced the patient to learn ankle, hip, and step strategies to maintain balance. Visual targets were used to promote trunk alignment and forward gaze. The patient often struggled to fixate on visual targets in a busy environment, so verbal cueing was utilized as well. To progress towards independent functional mobility, the patient was instructed on walking with one upper extremity supported on a wall for balance training. By using a wall for support as opposed to hand hold or cord assist, the patient was only able to utilize her lower extremities for ambulating rather than shifting some weight to her upper extremities. This activity allowed the patient to begin gaining confidence standing and walking with full weight acceptance on the lower extremities.

As noted earlier, if not cued verbally or visually to maintain trunk extension, the patient would flex at her trunk, resulting in hip and cervical flexion as well. To combat scissoring, multiple interventions were attempted. A bolster was placed between the patient's lower extremities, at the level of the knee to promote an increase in base of support and hip abduction. The patient was also verbally cued to her widen base of support to increase her body position awareness. Finally, the patient would have her lower extremities manually guided by the physical therapist during swing phase into proper position. To decrease trunk flexion during gait, Neurodevelopmental treatment techniques and Votja therapy were utilized to promote upright posture.^{14,15} Visual targets, as discussed earlier, were also used to promote vertical orientation. As the patient demonstrated increased independence with ambulating without an assistive device, her base of support became wider and she displayed decreased knee hyperextension during static stance and walking.

Outcome Measures

Throughout her time at Genesis Pediatric Therapy, the Peabody Developmental Motor Scale-Second Edition (PDMS-2) was administered. The PDMS-2 has a high test-retest reliability (intraclass correlation coefficient of .88-1.00), a sensitivity to change coefficient between 1.6 and 2.1, and a responsiveness coefficient between 1.7 and 2.3. These metrics demonstrate that the PDMS-2 is an appropriate tool to evaluate children with cerebral palsy.¹⁶ At 30 months of age, both the patient's age-equivalent stationary gross motor score, and locomotion gross motor score were that of a 9-month-old. The PDMS-2 was re-assessed during the 9-week case study period when the patient was 5 years old. The patient had an age-equivalent stationary score of 13 months, and a locomotion age-equivalent score of 11 months. During both testing periods, the patient was only able to complete 2 of the 3 subsections due to her visual impairment. The patient was not able to react quickly with her upper extremities to moving objects, which is the preliminary measure in the Object Manipulation subsection of the PDMS-2. The patient reached her ceiling level in the PDMS-2 when the tasks called for independent stance or ambulation.

An additional measure that was utilized during the 9-week case report period was the Gross Motor Function Measure-88 (GMFM-88) which has been shown to reliably detect changes in gross motor function over time. Since the GMFM-88 can be utilized for patients of any functional status (as defined by GMFCS levels I-V), the minimal clinically important difference varies for each GMFCS level. The MCID is greater for GMFCS levels I and II compared to GMFCS levels IV and V. The interrater and intrarater reliability of the GMFM-88 are >.97 and >.94, respectively, showing that the GMFM-88 is a reliable measure of gross motor function in children with developmental delays.¹⁷ The GMFM-88 is composed of 5 dimensions: lying/rolling, sitting, crawling/kneeling, standing, and walking/running/jumping. A higher score indicates greater proficiency. The patient scored 100% in the lying and rolling category. The patient scored 90% in the sitting dimension, with the only items not

achieved being transitioning from the floor to sitting on a bench. The patient was content maintaining a seated position and did not attempt the task. In the kneeling and crawling dimension, the patient scored 57% due to her inability to maintain tall kneeling or half kneeling without support. In the standing dimension, the patient scored 21%. The patient was able to complete each task in the standing subsection that involved at least one upper extremity support, but was limited when the tasks required hands-free actions. The patient scored 15% in the walking, running and jumping subsection, and was limited in the same manner she was in the standing category. The GMFM-88 was not utilized previously, but contained additional items that the PDMS-2 lacked to gain a comprehensive sense of the patient's ability level.

While a formal, computerized gait analysis was not performed, the patient's gait was utilized as an outcome measure based on the amount of support the patient required for walking, and the quality of motion.¹⁸ The patient's gait was documented, including location of support, type of support, and gait deviations. At the start of the 9-week case study, the patient required moderate assist at the hips for weight shifting and moderate assist at the lower extremities to correct foot placement. By the end of the case study duration, the patient was able to ambulate with one upper extremity supported on the wall, with contact guard assist. Additionally, by the end of the case study the patient demonstrated decreased knee hyperextension during gait and demonstrated independent weight shifts to advance her lower extremities approximately 75% of the time. The patient also displayed increased awareness of visual surroundings and maintained head elevation approximately 50% of the time during gait. It is important to note that the patient's performance during gait training fluctuated over the 9-week period based on participation, fatigue, and time of day.

To address visual changes relating to function, a Functional Visual Evaluation and the patient's level of Cortical Visual Impairment, as described by Roman-Lantzy, were re-assessed at multiple points in the patient's rehabilitation.^{1,19} The results of the Functional Visual Evaluation were presented previously in this paper. The patient's vision showed improvements, as demonstrated in the FVE and CVI level. The CVI range is scored from Phase 0 to Phase 10, with Phase 0 being no visual response and Phase 10 being resolution of visual processing issues.¹ Upon initial evaluation the patient's vision was rated as Phase 1, meaning that she had minimal to no visual response to stimulus. At the midpoint of the 9-week period of this case study, the patient was rated in Phase 4 of CVI, meaning that she functioned with more consistent visual responses to surroundings.

Considerations When Performing Pediatric Standardized Tests

The PDMS-2 and the Gross Motor Function Measure (GMFM) have been shown to be effective outcome measures to detect change in gross motor function over time.¹⁷ While both of these measures have been deemed appropriate measures for patients with brain injuries, neither of these standardized tests takes into account visual impairment associated with brain injury.¹⁷ Due to the patient's vision, the patient was limited by both her ability to reproduce a demonstrated task and her fear of movement, which are characteristic of cortical visual impairment.¹

Discussion

The purpose of this case report was to describe the multi-dimensional intervention approach to a patient with cortical visual impairment and abnormal gross motor development, as well as the challenges associated with evaluating progress.

Early intervention was likely important in preserving the patient's motor and visual function. While visual impairments have the potential to improve over time, implementing visual stimulation during the critical period of neural plasticity will increase a child's potential for functional sight and control of posture.^{1,20,21,22} Visual stimulation exercises are not typically thought of as physical therapy interventions, but pediatric patients who are limited in their visual abilities will be limited in their motor learning as well. Westcott et al. acknowledged that vision is used as the primary information source when first learning a task in a novel environment, emphasizing the important role that vision plays in motor learning.²³

A study done by El-Maksoud et al. demonstrated that visual-based training programs can improve motor function in patients with cortical visual impairment and cerebral palsy, compared to routine physical therapy. Individuals who received physical therapy interventions that incorporated a visual component showed statistically significant improvements in GMFCS and PDMS-2 scores over a 3-month period of intensive therapy.²⁴

Visual stimulation exercises can be performed by families to improve visual awareness, independent of physical therapy. The role of physical therapy in the management of cortical visual impairments is incorporating visual training with balance and mobility training. The patient described in the case study had decreased awareness of her environment. This was a result of both her impaired vision, and her inability to maintain vertical orientation during functional tasks. It was critical to address each of these underlying factors.

Eck et al. noted that factors such as increased height, changes in body mass index, and hormone levels can influence gross motor function.²⁵ As the patient grew and her body changed throughout her time in physical therapy, there were fluctuations in gross motor function. These body changes could have affected the patient's functional mobility, leading to a perceived discrepancy in progress. Finally, adherence to home exercise programs is critical for maintaining and progressing visual and motor function. Motor learning requires repetitive, task specific training, as noted by Richards et al. Additionally, repetition is critical to provoke and maintain functional changes.²⁶ Home programming is essential to promote and sustain the greatest change in behavior with regards to visuospatial awareness and motor learning.²⁷

As discussed previously, there are numerous confounding factors that play into a pediatric patient's progress, making it difficult to produce a cause and effect relationship between intervention and outcome. The patient's progress, as measured by the PDMS-2 and GMFM, was confounded by her cortical visual impairment. Both the patient's quality of movement and vision have improved over the past 2 years. At this time there are no gross motor standardized tests that are specific to individuals with visual impairments. Due to the increasing prevalence of cortical visual impairment, developing a standardized measure that combines visual and motor function would be beneficial to advance patient care.

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