**CVI status post rare neonatal thalamic stroke**

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**Abstract**

This case report discusses the multidisciplinary approach in the diagnosis/management of CVI in a child with a history of cerebral palsy and global developmental delay after a rare symptomatic intracranial hemorrhage as a full-term neonate.

**Key Words:** Cortical/cerebral visual impairment, intracranial hemorrhage, cerebral palsy

**Case History**

A 2.5-year-old African American female presents for a functional vision evaluation at the New England Eye Low Vision Clinic at Perkins School for the Blind (Watertown, MA). Significant ocular history includes variable exotropia and cortical/cerebral visual impairment (CVI). Significant medical history includes rare thalamic neonatal stroke at 3 days of age, cerebral palsy (CP), epilepsy, global developmental delay, diabetes insipidus, and GERD. She is unable to sit unassisted, does not use speech, and there is minimal reaching and grasping per mother. She was born full term with no complications during pregnancy or birth. She takes Cyproheptadine, Desmopressin, Omeprazole, Miralax, Topiramate, Diazepam, and a multivitamin. Her mother reports that she appears to primarily respond by auditory stimulation and would like to know more about how she uses her vision. Her ocular health is managed in collaboration with the ophthalmology department at the Boston Children’s Hospital (BCH) and she has an appointment for a dilated exam there in October. Previous dilated ocular health exams by BCH have been unremarkable and they have previously deferred glasses. Her last cycloplegic refraction at BCH nine months ago revealed +2.50-2.00x180 OD and +3.00-2.50x180 OS.

**Pertinent Findings**

Although BCH’s ophthalmology department had previously measured a 20/150 acuity without correction OU via visual evoked potential one year ago, a Teller acuity is unable to be obtained, (despite the setting of a dim room and direct lighting on the Teller cards) due to lack of interest and/or attention. Intermittent fix and follow is possible with a quiet, unshaken, yellow mylar pom-pom. She does not reach for the pom-pom and there is a large latency in response. Alignment via Krimsky reveals a variable, ~35 prism diopter, intermittent and alternating exotropia, largely dependent on fixation and attention. Full lateral gaze is present with minimal down gaze. Up gaze is not evident throughout the exam. Roving nystagmus is also intermittently present. Dry retinoscopy shows less hyperopia than her previous cycloplegic refraction: OD +1.50-2.00x180 and OS +2.25-2.00x180.

Contrast sensitivity with Double Happy Low Contrast cards measures well below average at 0.15 log CS (71% Weber Contrast). Again, the cards were isolated in space by peripheral dim lighting with only the Double Happy cards illuminated. Latency was again a factor. Pupils are equal, round, reactive to light, without the presence of an afferent pupillary defect. Visual fields are tested via modified confrontation with both eyes open in a dim room. The central stimulus is the examiner while the peripheral stimuli are 1.5” diffusely illuminated LED “wands” that are alternately lit to estimate the extent of the patient’s visual field awareness via fixation changes. This is completed both on the mother’s lap and while laying supine on a floor mat to confirm that her response is not dependent on her posture. While her right field appears to be grossly present, the left field appears to be significantly reduced.

Most interestingly, when presented with familiar books and toys, the patient is much more responsive, reaching to touch the bright colored pages and pegboard pieces. This visual preference of familiar objects over novel objects, as well as distinct color preferences, is common in patients with CVI. Her acuity and visual function can easily be underestimated with novel acuity cards or fixation objects.

**Diagnosis and Discussion**
Symptomatic intracranial hemorrhage incidence in an otherwise healthy, full-term baby with an uncomplicated birth history is estimated to occur in 2.7/10,000 live births\(^1\). The affected neonate most often presents with seizures (~70%), apnea, respiratory distress, cyanosis, and/or poor feeding\(^1,3\). Major risk factors include congenital heart disease, fetal distress, and hemostatic abnormalities\(^2\), none of which were present in our patient at the time of her stroke. Mortality rates are reported at 12-25%\(^1,2\). Approximately 10% develop CP, with 50% patients demonstrating unresolved neurological deficits at one year of age, with greater risk in grade III-IV hemorrhage\(^2\). The most common neonatal intraventricular hemorrhage is subdural\(^4\), though our patient demonstrated an even rarer thalamic stroke, diagnosed via cranial ultrasound. Parallels can be found in premature infants who are at high risk for intraventricular hemorrhage, which puts the preemie at high risk for CP, global developmental delay, and CVI.

CVI is the leading cause of visual impairment in children in developed nations\(^5\). There are three major factors that describe CVI – the traditional ocular health exam does not accurately explain how they see, the child has history of a major neurological event, and they have ten major characteristics\(^6\) in common. These characteristics include, but are not limited to: unusual attention to color, attention to movement, latency, difficulty complexity, visual field differences, visual novelty problems, absence of reflex blink response, limited distance viewing, light gazing, and absence of visually directed reach. Theses characteristics are present in our patient further supporting the diagnosis of CVI. Due to the lack of a specific protocol to diagnose CVI, multiple questionnaires for caregivers have been developed that touch on the above characteristics to assist in the diagnosis.

Treatment, Management

The patient requires multidisciplinary care and communication among providers due to the complexity of her ocular and medical health conditions. Her internal ocular health is currently monitored at BCH’s ophthalmology department, while her functional vision is cared for by the low vision team at Perkin’s School for the Blind. She is also closely monitored via her neurologist and pediatrician. First and foremost, it is most important that the patient’s underlying neurological disease/dysfunction is appropriately managed via medication or therapy. Our patient’s epilepsy is controlled via oral medication, and she is seen by physical, occupational, speech/language therapists to assist with her CP and developmental delay. She is registered with the Massachusetts Commission for the Blind in order to receive services as needed.

Many children with CVI improve in visual function over time, making it imperative for the visual system to be stimulated frequently and in an appropriate manner based on CVI characteristics. It is vital to the patients’ visual development to have the ongoing services of a teacher of students with vision impairment (TVI) who are specially trained in such cases.

The trial of spectacle aware may be warranted at her age in order to optimize her visual function. Our notes and recommendations will therefore be distributed to her care team members, and to the family so that the decision to pursue spectacle wear can be made after her next cycloplegic refraction. Many children with CVI are tactilely sensitive; therefore spectacle wear may not be possible in all cases.

Conclusion

In patients of CVI, visual stimulation should include large, high contrast well lit targets, tactile or auditory cues to initiate fixation/attention, be presented in a simplified environment with extra time for response latency, and over stimulation should be avoided. All of the aforementioned suggestions should be tailored to each child’s visual abilities. Co-management with a multi-disciplinary team is imperative to ensure the best prognosis for these patients.

References


