Title: Strategies for objective visual acuity assessment in pediatric patients with cortical visual impairment (CVI)

Abstract: Objective evaluation of visual acuity can be challenging in patients with cortical visual impairment. Presented are two unique cases of CVI and methods for estimating visual acuity in order to make prescribing and functional recommendations.

Patient 1
I. Case History
- Patient demographic: 13-year-old, African-American female
- Chief complaint
  - Primary: Repeat dilated fundus exam and cycloplegic retinoscopy, poor cooperation at last visit
  - Secondary: Patient often breaks glasses, spectacles fit poorly
- Ocular and medical history: history of high hyperopia and mild astigmatism OU; paternal family history of retinitis pigmentosa; Dravet’s syndrome (severe myoclonic epilepsy of infancy): most recent seizure 2 months ago; history of encephalopathy; developmental delays; feeding disorder
- Medications: Canabidiol oil, L-carnitine, Miralax® and Vitamin B6 supplements
- Other information: non-verbal and non-ambulatory; no current therapies; does not go to school

II. Pertinent findings - Clinical exam
- Visual acuity: Unable to fix and follow with or without trial-frame correction, no behavioral response to lights being turned off, negative menace response
- Alignment: Aligned by Hirschberg with and without correction, grossly full versions by observation
- Field of view: Unable to be assessed
- Cycloplegic retinoscopy: OD: +8.00 -2.00 x 180, OS: +8.00 -2.00 x 180
- Ocular health: Clear media and normal anterior segment health OD, OS; normal optic nerves without pallor or hypoplasia OD, OS; unable to obtain satisfactory views of the peripheral retina
- Physical: 60 pounds, less than four feet tall, confined to stroller
- Others: Affect and orientation: Consistent rocking back and forth, chewing on hands

Visual Evoked Potential for visual acuity and field evaluations - Scheduled for October 3, 2017
- Results pending (Figure Available)

III. Differential diagnoses
- Primary: Cortical visual impairment
- Others: Optic nerve disorders, amblyopia, retinitis pigmentosa (RP)

IV. Treatment and management
- Patient 1 exhibited no visual interest in any introduced visual stimuli or room lighting alterations with or without correction while in the exam room. Because of this, a VEP was ordered to assess visual functions and determine if her mother should continue to struggle with fulltime spectacle wear.

Patient 2
I. Case History
- Patient demographic: 6-year-old, African-American male
- Chief complaint: Neurologist referral for suspect cortical visual impairment
- Ocular and medical history: Intraocular lens implants OU status-post steroid-induced cataracts; mild, bilateral optic nerve hypoplasia; born at 30 weeks gestation; agenesis corpus callosum; tracheotomy; moderate hearing loss; ear tubes; feeding disorder
- Medications: Gabapentin, Nasonex®, Miralax®
- Other information: Currently in speech therapy, physical and occupational therapies; non-ambulatory

II. Pertinent findings - Clinical
- Visual acuity: Fix and follow OD, OS; no resistance to occlusion, reaches for small toys in dim and bright lighting; unable to test with forced-choice preferential looking; negative menace response
- Extraocular muscle movements are full in both eyes with mild, end-gaze nystagmus
- Field of view: Non-seeing to seeing, grossly full in all quadrants OD, OS
- Habital correction: OD: Plano, OS: Plano, Add: +2.00
- Ocular health: Normal anterior segment ocular health OD, OS (clear media)
- Physical: Non-verbal, wheelchair-bound

III. Differential diagnoses
- Primary: Cortical visual impairment
- Others: Optic nerve head hypoplasia, deprivation amblyopia

IV. Treatment and management
- Diagnosis of CVI in this patient is primarily informational. His resource teachers and occupational, speech and physical therapists will be informed of his diagnosis and adjust their therapies accordingly. Through habilitation, acuity may enable improved motor skills, social interaction and visual behavior.

V. Diagnosis and discussion
Elaborate on the condition: Cortical visual impairment (CVI) is a neurological disorder caused by a lesion beyond the optic chiasm that results in bilateral vision loss. In pediatric patients, CVI manifests as impaired visual function with normal ophthalmologic evaluation and normal pupil function. Visual field defects are common in CVI.

Expound on unique features: In many patients with CVI, there is a concurrent motor compromise. Head and eye movements may be inaccurate or spastic, making use of preferential looking tasks inconclusive as inattention or inability to direct gaze may prevent a patient from following a stimulus. The use of Visual Evoked Potential (VEP) testing for quantifying visual function in patients can provide general information about responses to visual stimuli within the occipital lobe. It is particularly useful with the CVI population because of their propensity to stare at luminescent sources. The patient can be situated such that the screen subtends a wide angle on the subject’s field. This allows coverage over any areas of visual field loss, or any motor deficits causing head or eye turning.

VI. Conclusions
Traditional exam techniques may not accurately assess visual function in patients with CVI. Therefore, for these patients, as well as those with motor impairments, it may be useful to recommend a VEP evaluation to aid in the assessment of visual discrimination. The results from such referrals could be an effective tool for determining the extent of CVI and the potential benefit of spectacle wear when visual acuity cannot be formally determined. The following list of objective methods are alternative options for the determination of visual acuity/function in patients with suspected CVI:
- Forced choice preferential looking: Teller Acuity Cards, Patti Paddles®
- Ability to fixate and/or follow an item of visual interest, particularly lights
- General observation of behaviors to light projection, extinguishing room lighting, various visual stimuli

References