Title: Management Considerations for a Child with Neurologic Injury with Yoked Prism combined with Vision Therapy

Abstract: This is a case report of a 9 year-old Hispanic female presenting with left homonymous hemianopia, acquired nystagmus secondary to ventriculoperitoneal shunt malfunction due to hydrocephalus.

I. Case History

- **Patient Demographics:** 9 year old Hispanic female
- **Chief complaint:** Patient was referred in by outside ophthalmologist for evaluation of left homonymous hemianopia and nystagmus, which occurred 3 weeks after surgery for ventriculoperitoneal (VP) shunt malfunction. Mother reports left pupil blew on 08/01/2013 and needed an emergency VP shunt revision surgery, which resulted in current symptoms.
- **Ocular History:**
  - Left homonymous hemianopia
  - Acquired Nystagmus
  - Exposure keratopathy of the left eye secondary to left facial nerve palsy
- **Medical History:**
  - Congenital Hydrocephalus in 2003
  - Ventriculo-peritoneal shunt at birth with 2 revision surgeries
  - Inguinal surgery
  - Cranioplasty Surgery x 2
  - Left facial nerve palsy
  - Stroke post VP shunt malfunction
- **Birth History:**
  - Length of pregnancy- 8-9 months, Delivery- Caesarian, Apgar score- 9/9, Birth Weight- 8 lbs and 4 oz
- **Allergies:**
  - Nut flavor, Cat Hair, Oak
- **Ocular medications:** Lacri-lube
- **No systemic medications**

II. Pertinent Findings

- **Pupils:** OD, OS unremarkable
- **EOMs:** OD, OS EOM is full, upgaze nystagmus
- **FU EOMs:** Distance horizontal pendular nystagmus, Near vertical downbeat nystagmus
- **Confrontation fields:** OD restricted Sup and Inf nasal, OS restricted Sup and Inf Temp
- **Color vision:** OD, OS pass
- **Distance VA cc:** OD 20/40+, OS 20/50+, OU 20/40+
- **Near VA cc:** OD 20/20, OS 20/20, OU 20/20
- **Refraction:** OD -1.75 sph, OS -1.25 sph
- **CT:** Distance (cc) ~14-18 ILET, Lhyper in all fields of gaze and Near cc: 10 ILET, Lhyper
- **Stereo cc:** Global 250”, Local 25”
- **Worth 4 Dot:**
  - Bright distance- 5 dots, intermediate 2 dots, near 2 dots
  - Dim distance- 2 dots, intermediate 2 dots, near 2 dots
  - Interpretation: LE suppression overall expect with distance view under bright conditions show uncrossed diplopia
- **Ocular health:** OD unremarkable, OS PEK inf
- **Visual Field:** Left homonymous hemianopia 24-2, reconfirmed with 30-2
General Observation: left head turn causing neck strain

III. Differential Diagnosis

- Left Homonymous Hemianopia
  - Stroke
  - Trauma
  - Space occupying lesion: gliomas, meningiomas, metastasis

- Acquired Nystagmus
  - Gaze evoked: Cerebellar or brainstem injury/disease, alcohol, intoxication, sedatives
  - See-saw: Lesion involving parasellar region and chiasm. If pendular with chiasmal region involvement and if jerking movement then midbrain involvement.
  - Convergence retraction: Papilledema, pineal region tumor or other dorsal midbrain abnormality.
  - Downbeat: Lesion at cervicomedullary junction (e.g. Arnold-Chiari malformation) or a manifestation of cerebellar degeneration.
  - Periodic alternating: Congenital or acquired most commonly associated with cervicomedullary junction and posterior fossa or MS, medications, rarely blindness.
  - Peripheral vestibular/Spasmus Nutans: dysfunction of vestibular end organ, eighth cranial nerve, or eighth cranial nerve nucleus or irritative lesions (Meniere disease). If vestibular nystagmus associated with interstitial keratitis called Cogan syndrome.
  - Other: Congenital nystagmus, Rebound nystagmus (cerebellar lesions), Bruns nystagmus (cerebellopontine angle), oculomasticatory myorhythmia (Whipple disease), oculopalatal myoclonus (prior brainstem stroke).
  - Superior oblique myokymia
  - Opisoclonus/saccadomainia
  - Internuclear Ophthalmoplegia

- Esotropia
  - Comitant or Concomitant Esotropia
    - Divergence insufficiency r/o elevated intracranial pressure, neurologic trauma, and pontine tumor
    - Congenital (infantile) esotropia
    - Accommodative esotropia
    - Sensory-deprivation esotropia
    - Intermittent esotropia secondary to decompensating esophoria
  - Incomitant Esotropia
    - CNS pathology: Acquired sixth nerve palsy, MG
    - Medial Rectus restriction: Thyroid eye disease, medial orbital wall facture
    - Lateral Rectus Weakness
    - Duane syndrome, Brown syndrome
  - Other
    - Mobius syndrome, congenital fibrosis syndrome

IV. Diagnosis and Discussion

- Left Homonymous Hemianopia: Stroke secondary to VP shunt malfunction
- Acquired nystagmus: Cerebellar or brainstem injury secondary to VP shunt malfunction
- Monocular esotropia: Suspected secondary to increased intracranial pressure from VP shunt malfunction causing neurological damage to cranial nerve six.

This case is very unique because it is not common to be functionally treating a young patient for a visual field defect, acquired nystagmus, and monocular esotropia secondary to VP shunt malfunction in treatment for hydrocephalus. Although CSF shunting remains the most common procedure used to treat hydrocephalus, it is fraught with complications and has a
high likelihood of revision. In these cases, once a patient has suffered from these complications, how do we help them function in everyday life.

V. Treatment and Management

- Left Homonymous Hemianopia: 10 PD base left Fresnel yoked prism at initial visit and reduction in prism thereafter. Although the patient was able to appreciate her extended visual field, the prism caused diplopia at intermediate and distance viewing and secondary right head turn resulting in neck strain. Patient also reported trouble reading the right end of lines and tends to look above glasses or closes one eye most likely due to apical scotomata caused by bilateral sector prism. Since patient did not have issues in scanning while reading without prism, no prisms were prescribed for the field loss.
  - Will discuss other types of prisms that could help patients scan and spot targets that are in their lost field (ex. Gottlieb prism, etc).1
- Acquired nystagmus: Head posture of chin down position reduced vertical nystagmus and null point of nystagmus at upgaze. Trial framed and prescribed 16 PD Yoked BD (after a couple of weeks of vision therapy), which reduced neck strain with near and intermediate work. Educated adaptation to BD yoked prism and peripheral awareness due to the effect of BD prism on gait may be due in part to a perception of increased height.2
- Vision Therapy goals are to improve scanning ability with yoked prism and function in everyday life. Other goals are to incorporate balance and peripheral awareness activities to reduce bumping into things. During vision therapy, it was also observed that she had visual processing issues and was tested as well. Vision therapy was structured to work on her visual memory, visual spatial relationships, laterality/directionality, and ocular motor dysfunction. Will discuss tests used and some examples of vision therapy activities patient is performing.
- Exposure keratopathy secondary to facial nerve palsy: continue with lubrication and patch of left eye

VI. Conclusion

- Consider unilateral or bilateral off setting of yoked prism or peripheral prism to avoid visual confusion or diplopia.
- Considerations of vertical yoked prisms for acquired nystagmus and education of adaptation to yoked prism.
- Co-management with other therapist to enhance rehabilitation for the patient.
- Always rule out other ocular pathological etiologies before treatment.

Bibliography
