Large vestibular aqueduct syndrome

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Vestibular aqueduct

- a temporal bone structure that runs from the vestibule to the posterior cranial fossa
- contains the endolymphatic duct, which terminates at the endolymphatic sac
- Adult size 0.4-1.0 mm

Large vestibular aqueduct syndrome

- First described by Valvassori and Clemis in 1978
- 50 cases out of 3,700 tomograms, or 1.4%
  - congenital hearing loss and vestibular symptoms
  - enlarged vestibular aqueducts from 1.5–8.0 mm
- 50% of the individuals with LVA were children and teenagers
- female to male ratio was 3:2
- bilateral to unilateral 2:1
- 60% associated with other inner ear abnormalities
  - enlarged vestibule, enlarged semicircular canals, hypoplastic cochlea
- SLC26A4 (PDS) gene
- Pendrin, cellular transport of chloride, iodine, and bicarbonate anions
- Pendred’s syndrome
  - thyromegaly
  - Mondini malformations
  - enlargement of vestibular aqueduct
### Prevalence

- 0.64% of children with SNHL of unknown etiology
- 1% of patients with various otologic problems
- 2.25% of patients referred for inner ear tomography
- 4% of children in a cochlear implant program
- 7% of patients with SNHL of unknown etiology
- 12% of deaf children
Hypothesis

• Back pressure/damaging pressure wave theories
• Electrolyte imbalance theory
• Hyperosmolar fluid reflux theory
• Ossicular discontinuity theory
• Third-window lesion
Back pressure/damaging pressure wave theories

- back pressure of perilymphatic and endolymphatic fluid
  - decreased stapes mobility
  - conductive hearing loss
- pressure shifts generated from the intracranial space to cross through EVA and damage the inner ear
- acoustic reflexes remain intact, inconsistent with stapes fixation
- does not explain the high incidence of sensorineural hearing loss
Electrolyte imbalance theory

- enlarged and dysfunctional endolymphatic sac
  - electrolyte derangement or toxic biproducts
  - damage the inner ear
- Large volumes of endolymph introduced from the enlarged system might overwhelm the ion pump mechanism of the stria vascularis
Hyperosmolar fluid reflux theory

- endolymphatic sac fluid, contain hyperosmolar fluid
  -> reflux through enlarged endolymphatic sac and duct
  -> enter the inner ear
  -> damage to the inner ear structures
Ossicular discontinuity theory

- temporal bone study demonstrating a 38% incidence of ossicular deformities in enlarged vestibular aqueduct cases
- air bone gap
Third-window lesion

- Any abnormal opening into the inner ear excluding the normal oval window (first window) and round window (second window)
- Sound energy being shunted out of the cochlea
Diagnosis

- diameter > 1.5 mm at the midpoint  
  Valvassori and Clemis

- diameter > 2 mm at the midpoint  
  Arcand et al., Jackler and De La Cruz

- diameter was more than double the diameter of the posterior semicircular canal  
  Wilson et al.

- 0.9 mm at the midpoint or 1.9 mm at the operculum  
  Cincinnati group
Large vestibular aqueduct case report and review of the literature
Acta Oto-Laryngologica, 2006; 126: 788-795
Large endolymphatic duct and sac

Normal

The large vestibular aqueduct: case report and review of the literature
Acta Oto-Laryngologica, 2006; 126: 788-795
Hearing Loss

- All three types of hearing loss (sensorineural, mixed, and conductive) have been reported
- down-sloping
- progressive or fluctuating
- sensorineural hearing loss progresses at an average rate of 4 dB per year

*Govaerts et al. Int J Pediatr Otorhinolaryngol 1999*
Precipitating Factors for Hearing Loss

- head trauma
- barotrauma (scuba diving, airplane flights, Valsalva)
- upper respiratory tract infections
- high fevers
- noise trauma
- physical exercise
EVA Size Correlations with Hearing Loss

- Enlargement and morphology of the vestibular aqueduct correlated highly with the severity of hearing loss

- Midpoint diameter correlated with the frequency and severity of hearing loss fluctuations, but not with the progression of hearing
  Lai C. Laryngoscope 2004;114:832–838

- T2-weighted MR, volume of the vestibular aqueduct and sac, the diameter of the duct and sac, and the signal intensity of the endolymphatic sac

- No significant correlation with the degree of hearing loss
Vestibular Symptoms

- Dysequilibrium
- Epidosic vertigo attacks of variable length
- Motor delays
  - Delayed ambulation
  - Poor coordination
- Severe vestibular loss in the perinatal period or early during development, may display few vestibular symptoms later in life
  - Compensated through development
Corticosteroid therapy

- Grimmer: retrospective cohort of 12 patients
- hearing improvement in four of five patients with corticosteroids
- lack of hearing improvement in six of seven patients without steroids
Endolymphatic Sac Surgery

- Jackler, 1989
  - shunting of the endolymphatic sac to the subarachnoidal space
  - four of seven patients had a significant early decline in hearing thresholds

- Welling, in 1999
  - endolymphatic sac occlusion on 10 patients, with 9 patients having some degree of additional hearing loss after the procedure
Hyperbaric oxygen therapy

- Nakashima et al
- After 22 sessions, the patient’s sudden hearing loss exhibited thresholds comparable to those prior to the hearing loss
- Hearing remained stable during an 8-month follow-up period
• **Hearing aids or cochlear implantation**
  - Bent, 1999: 10 patients of EVA undergo CI, 7/8 gained good speech recognition
  - Harker et al: perform the implant as soon as possible after a profound hearing loss to avoid neural degeneration
Thanks for your attention.