

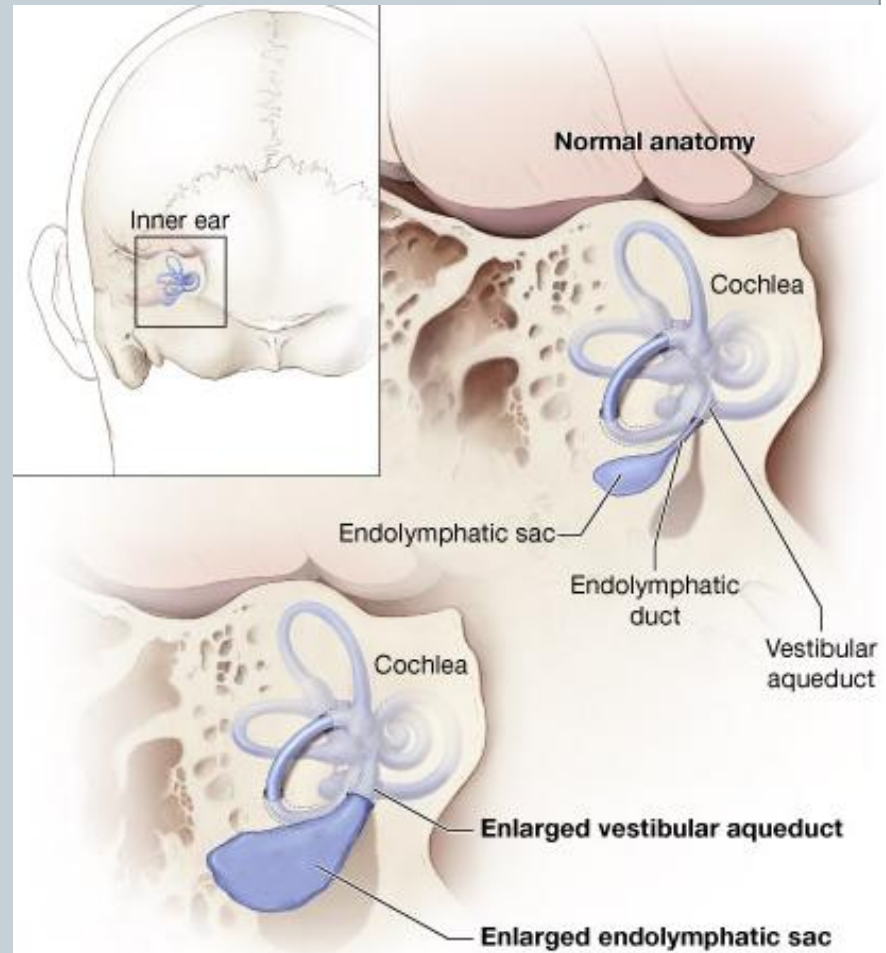
# Large vestibular aqueduct syndrome



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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 byproducts
  - > 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

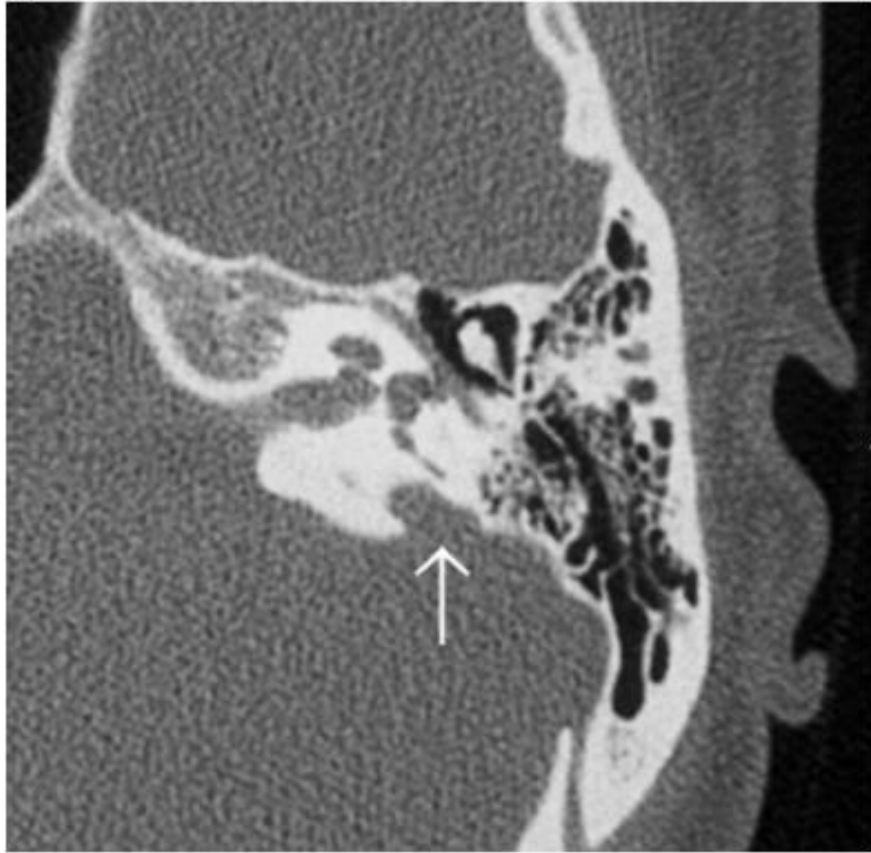
*Wilson et al.*

- 0.9 mm at the midpoint or 1.9 mm at the operculum

*Cincinnati group*

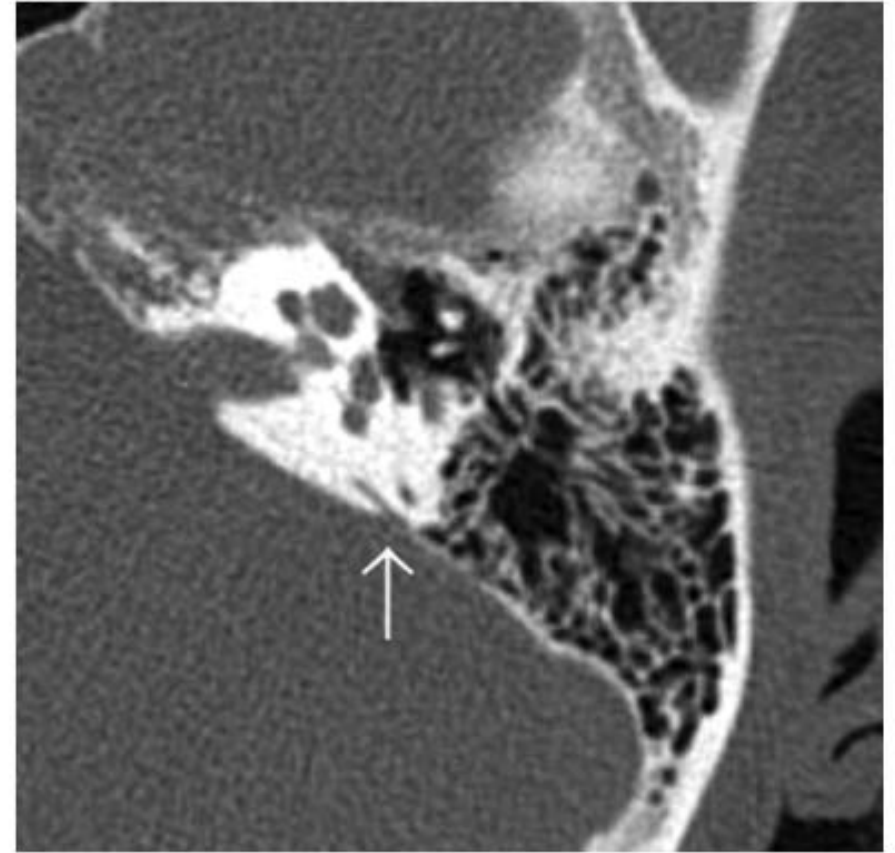
Large vestibular aqueduct

A



B

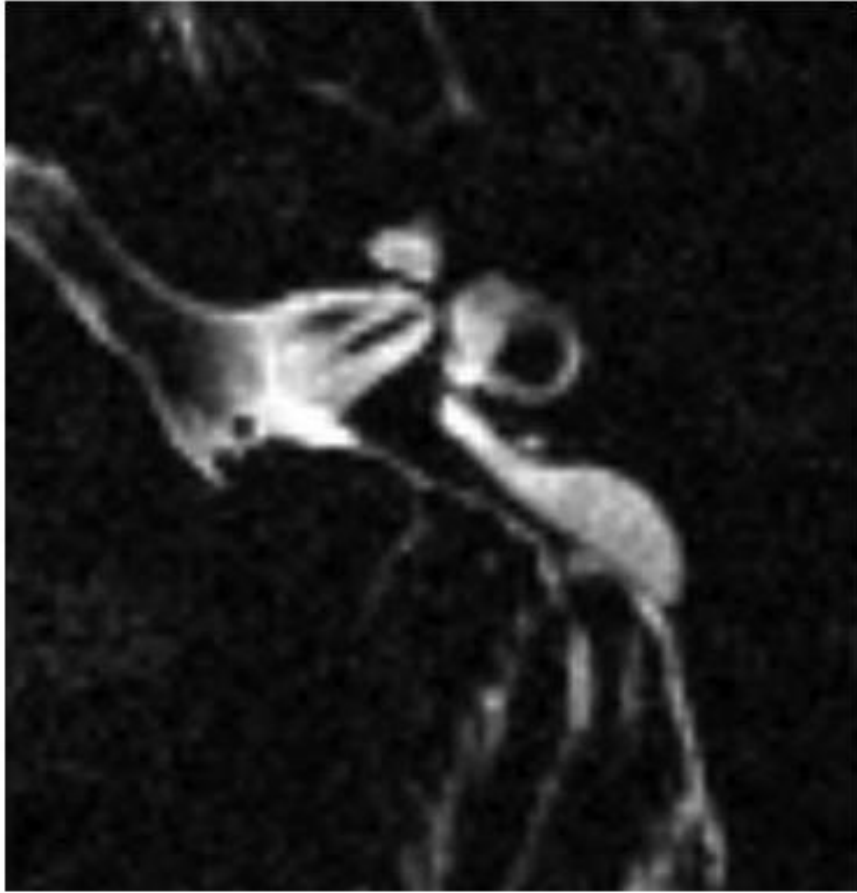
Normal



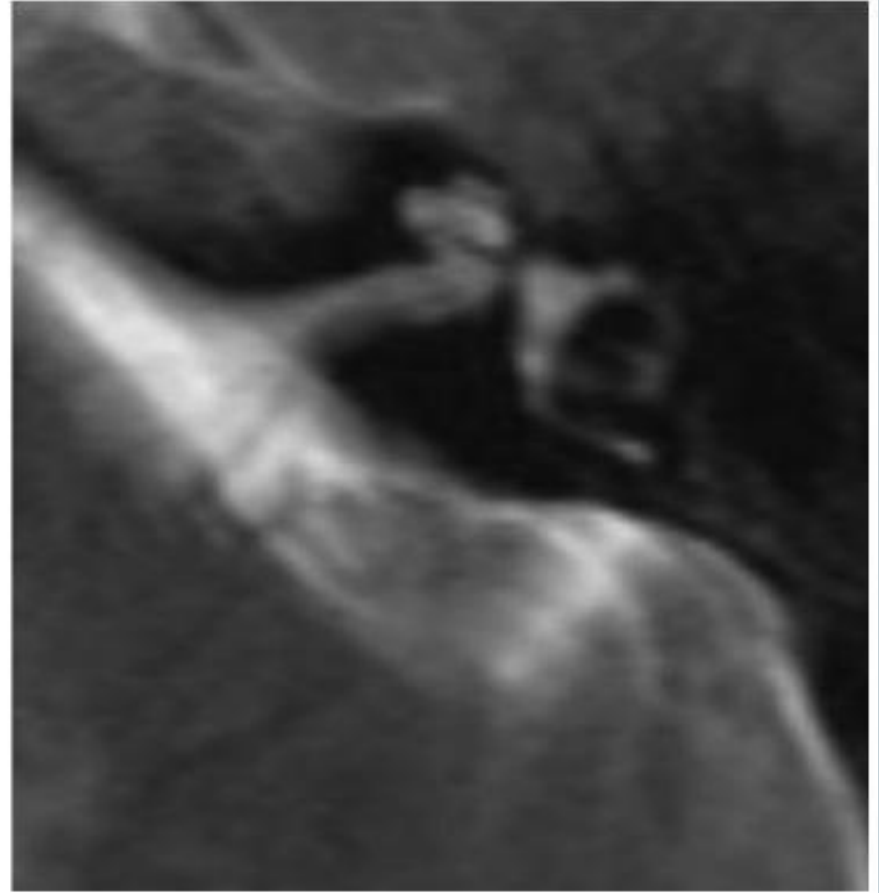
Large endolymphatic duct and sac

Normal

A



B



The large vestibular aqueduct case report and review of the literature  
*Acta Oto-Laryngologica*, 2006; 126: 788795

# 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

*Antonelli P. Am J Otol 1998;19:306–312*

- 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

*Naganawa S. AJNR 2000;21:1664–1669*

# Vestibular Symptoms



- Dysequilibrium
- Episodic 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

# Treatment



- **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.