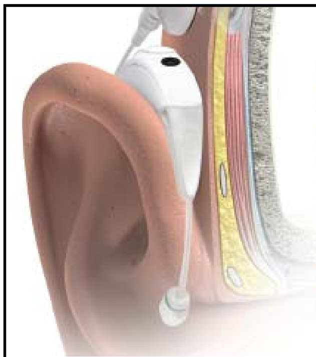

GRAND ROUNDS IN COCHLEAR IMPLANTS

Hosted by Advanced Bionics

Presented by Jennifer Wolf, Au.D., CCC-A
June 19, 2015

Advanced Management of Complex Cases: Enlarged Vestibular Aqueduct

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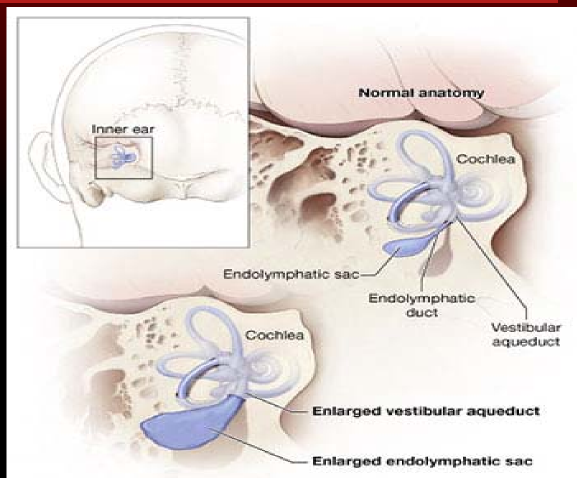
Advanced Management of Complex Cases: Enlarged Vestibular Aqueduct

Jennifer Wolf, AuD, CCC-A

Agenda

- Definition of EVA
- Audiologic configuration and progression of hearing loss
- Theories describing cause of hearing loss
- Audiologic (Re)habilitation
- Cochlear implantation considerations
- Case study

Anatomy



http://www.nidcd.nih.gov/health/hearing/pages/eva.aspx

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Describing EVA

- 1791 • Mondini
- 1969 • Valvasori
- 1978 • Valvasori and Clemis

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Defining EVA

Valvasori and Clemis (1978)

"a diameter greater than 1.5 mm halfway between the common crus and medial aspect of the operculum on the posterior wall of the temporal bone"

Defining EVA

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Imaging options

- Diagnosis can be made by a radiologist with either Computed Tomography (CT) scan or Magnetic Resonance Imaging (MRI) scan
  - CT Scan
    - Vestibular aqueduct is identified
  - MRI
    - Endolymphatic duct as well as endolymphatic sac can be identified

EVA is the ***most common*** inner ear anomaly identified on imaging for children with hearing loss

Development

Complete in Utero?

- Evidence that there is no change in VA size postnatally when comparing ***average*** measurements across children and adults

- Arrest in development causing the aqueduct to not elongate and form into a "j" shape

Continued Growth?

- Some data suggests that the VA continues to grow postnatally in nonlinear fashion until a child is 3-4 years of age

Statistics, for now…

- Prevalence of EVA is estimated to range from 1- 14% of in populations with SNHL
- Bilateral to unilateral ratio 2:1
- Female to male ratio 3:2

Associated Congenital Disorders

- Pendred syndrome
  - Autosomal recessive
  - Mutations on gene SLC26A4 resulting in hypothyroidism and goiter
  - Combination of thyroid dysfunction and EVA
- CHARGE syndrome
- Branchio-oto-renal syndrome

POLL QUESTION 1

POLL QUESTION 2

POLL QUESTION 3

Clinical Presentation

Hearing Concerns
- Failure on hearing screening(s)
- Reduced auditory responsiveness in daily activities, in some cases following minor head trauma
- Reported difficulty hearing
- Speech and language delay/ concerns

Vestibular Symptoms
- delayed ambulation in childhood
- episodes of vertigo with variance in length
- disequilibrium

Audiologic Test Battery

- Tympanometry
- Acoustic reflexes
- Pure tone thresholds
- Word recognition ability
- Otoacoustic emissions
- VEMP

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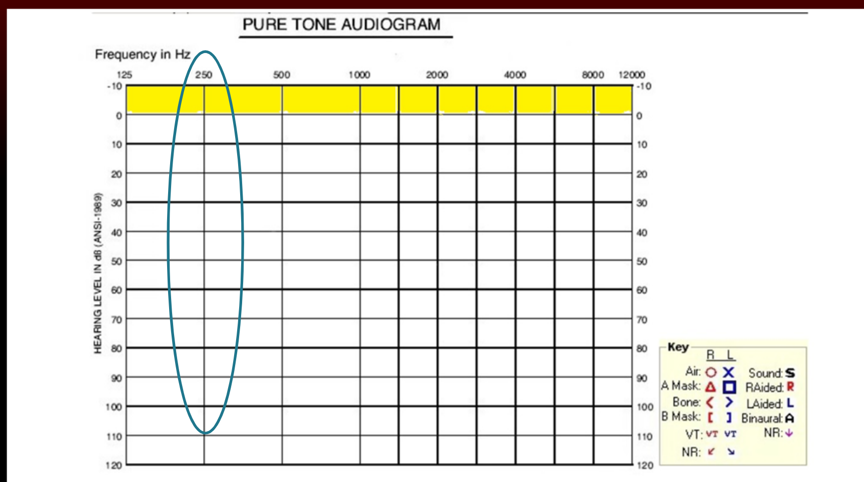
Pure Tone Thresholds



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Type and Configuration of HL

- **Type of Hearing Loss**
  - Conductive, Mixed AND Sensorineural hearing loss have ***ALL*** been reported in the literature
  - Conductive hearing loss or mixed components are most likely to occur in the in low frequencies (250 Hz and 500 Hz)
- **Configuration of Hearing Loss**
  - Most commonly reported are are down sloping, flat, and reverse cookie bite

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Degree of HL

- Degree of hearing loss ranges from mild to profound
- Hearing loss is reported to potentially fluctuate, rapidly change or gradually change over time with no specific incident.
- Hearing loss can also range from deafness in childhood to stable hearing loss into adult life

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Word Recognition Ability

- Often word recognition will decline with progression of hearing loss
- Word recognition may be poorer than expected when compared to conductive or mixed components of middle ear origin

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Additional Measurements

- Tympanometry
  - Expected to be within normal limits
- Acoustic Reflexes
  - Ipsilateral reflexes (tonal or BBN) can be present with conductive and/or mixed components
- Otoacoustic emissions

Vestibular Testing

- Vestibular Evoked Myogenic Potential (VEMP)
  - Present responses despite air-bone gaps
  - **Potential for HIGH** Ocular VEMP amplitude and **LOW** Cervical VEMP threshold

Differential Diagnosis

Test	Result	Rule Out
Tympanometry	Normal	ETD, fluid
Ipsi Reflexes	Likely present *	most ossicular concerns
OAEs	Present/Absent	likely absent in cases of true middle ear dysfunction
VEMP	Present; can be lower in threshold than normal, and greater in amplitude	middle ear dysfunction

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Interpretation of Results

Mixed

Cochlear conductive

Air-bone gap

sensorineural

Pseuodoconductive

How do we adequately describe the presence of conductive or mixed components when results are not consistent with middle ear pathology?

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Interpretation of Results

- Describe conductive components and air bone gaps identified on pure tone testing
- Indicate that despite conductive components, results are **not** consistent with middle ear pathology.

Precipitating Factors

- Head trauma, barotrauma, high fever, noise exposure, URI can all increase risk for sudden changes in hearing
- Minor head trauma does NOT always cause a further decline in hearing
  - ***ONE THIRD*** of patients reported a sudden decline in hearing following minor head trauma

Are there predictive values?

- In the majority of studies, vestibular aqueduct size or endolymphatic sac size is **not** correlated with degree of hearing loss
- Madden et al, 2003 reported that the mean VA at the operculum was significantly larger in patients with a progressive hearing loss versus those with a stable or fluctuating loss

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Clinical Implications

MONITOR, MONITOR, MONITOR!!!

- Use all diagnostic tools possible in order to help a physician make the correct diagnosis and avoid unnecessary surgery (such as middle ear exploration)
- Counsel patients and families on potential causes for further hearing loss, and activities they may wish to avoid

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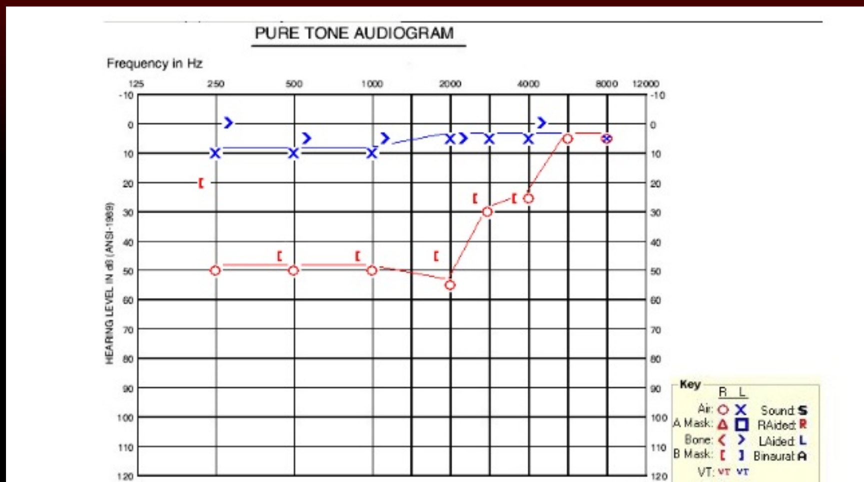
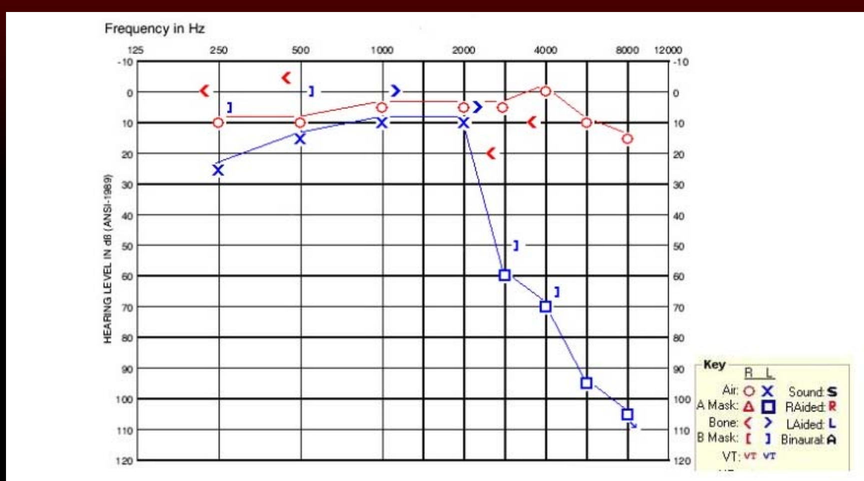
A


PURE TONE AUDIOGRAM
Frequency in Hz
HEARING LEVEL IN dB (ANSI-1989)
Key
Air: A Mask: Bone: B Mask: VT:
Sound: S RAided: R LAided: L Binaural: A

B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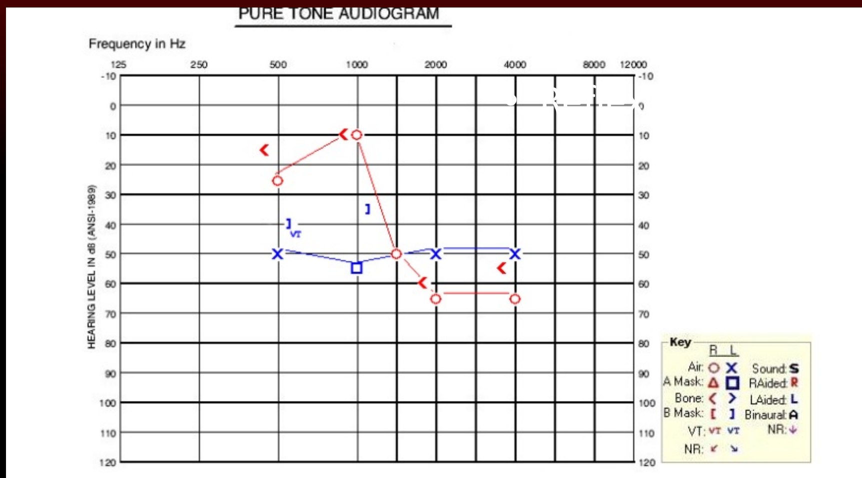

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D

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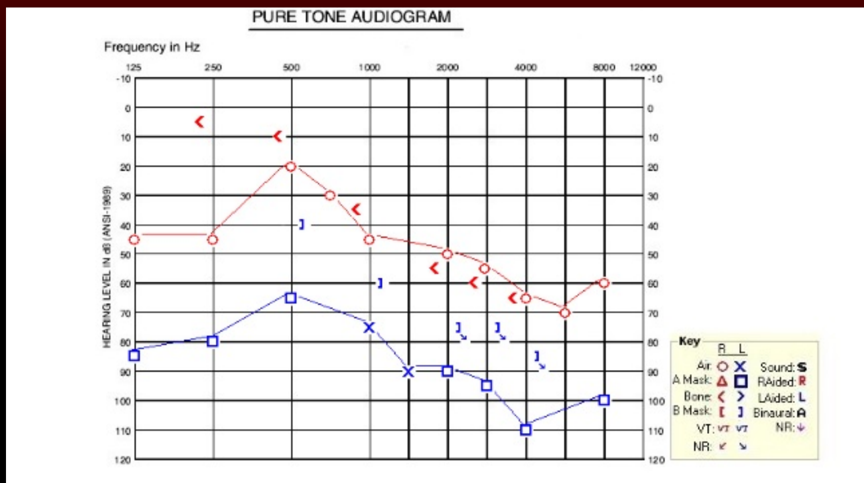
D, 1 year later



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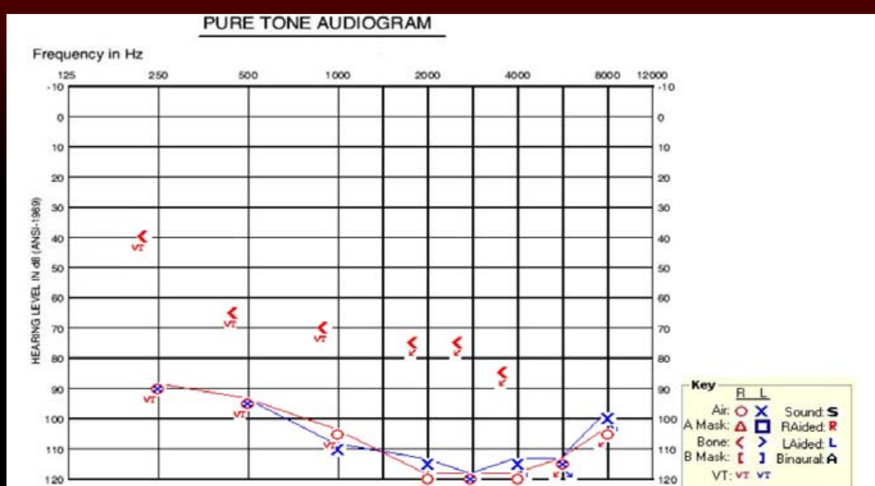
E



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POLL QUESTION 4

POLL QUESTION 5

Cause of HL?

EVA is a radiologic marker for HL, NOT the cause

EVA is the cause of HL

Theories of Hearing Loss

Pressure wave theory I

- Conductive/ mixed losses can be explained by back pressure of perilymph and endolymph causing decreased stapes mobility
- Potential reason for perilymph "gushers"

Pressure wave theory II

Greater pressure shifts from intracranial space cross through VA and damage the inner ear, specifically hair cells.

Theories of Hearing Loss

Electrolyte Imbalance Theory

- Proposed that endolymphatic sac itself disturbs homeostasis
- Large volumes of endolymph could overwhelm the the ion pump mechanism of the stria vascularis.

Hyperosmolar Fluid Reflux Theory

- Endolymphatic duct fluid which contains hyperosmolar fluid can reflux easily through the larger aqueduct and cause damage to the inner ear.

Theories of Hearing Loss

Middle Ear Involvement

- Ossicular Deformities
- Stapes Fixation
  - Middle ear exploration due to mixed hearing loss revealed fixated stapes footplate and perilymphatic “gusher” in the presence of EVA

Theories of Hearing Loss

Third Window Lesion

- Defined as any abnormal entrance to the inner ear
- Sound energy is shunted out of the cochlea, resulting in poorer air conduction and improved bone conduction

Treatment for Decline in Hearing?

- Endolymphatic sac shunt, occlusion or obliteration surgery
- Corticosteroid treatment

Audiologic (Re)habilitation Options

- Preferential Seating
- FM system (sound field or personal)
- Hearing Aids
- Cochlear Implant

OR

- Combination of the above, depending on degree of hearing loss and word recognition ability

Cochlear Implant Considerations

- Mixed components should not rule out CI candidacy
- Surgical
  - Electrode array (dependent on other anatomical considerations, progression of HL)
  - Reported “gushers”
  - Postoperative recovery

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Cochlear Implant Programming

- Potential for fluctuations in impedances, thereby impacting voltage compliance limitations
- Fluctuation in preferred loudness measurements
- May want to control each ear independently (volume, sensitivity) in case of fluctuations

Cochlear Implant Performance

Children

- In many cases, children will have acquired some speech and language before implanted, therefore may have limited time without auditory input and potential for improved performance

Adult

- Many adults hold on tightly to even a small amount of hearing and may be fearful to be implanted
- Duration of hearing loss and previous use of amplification can impact post-operative performance

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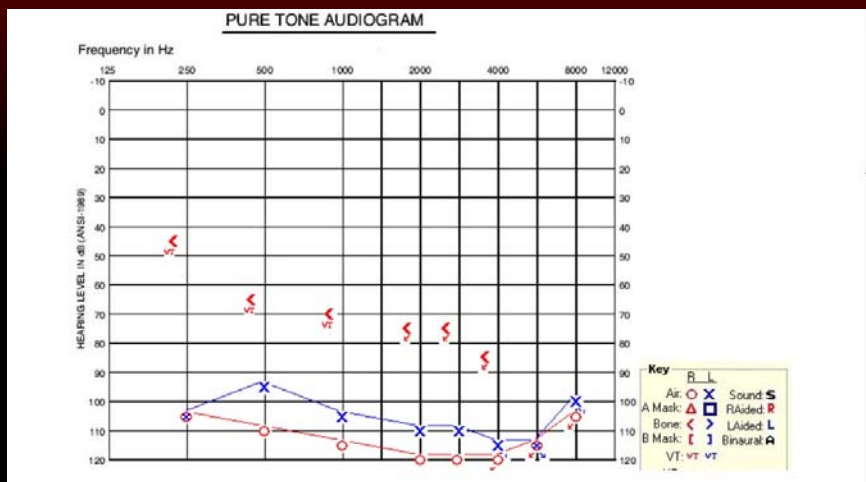
Case Study

Cochlear Implantation with bilateral EVA

Case

- 40 y.o male with reported bilateral progressive hearing loss first identified when he was 4
- Noted poorer hearing always in the LEFT ear from childhood
- Consistent use of amplification on the RIGHT and inconsistent use on the LEFT; access to speech on the RIGHT with environmental and VT on LEFT
- First identified with EVA at the age of 38

Cochlear Implant Evaluation



Cochlear Implant Evaluation

	Right	Left	Binaural
AzBio			0%
CNC Whole word Phoneme	 2% 7%	 0% 3%	 0% 6%
HINT	1%	1%	1%
Four-Choice Spondee (closed set)	60%*	90%*	90%*

*Chance is 25% correct

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Implantation and Activation

- RIGHT Advanced Bionics HiRes90k Advantage/ HiFocus1j electrode
  - No resistance on insertion; no evidence of a "gusher" per physician report
- Activation
  - Naida Q70 sound processor, HiResOptimaS processing strategy

Post-Op Performance

	Pre-Op	3 month	1 year
AzBio		95%	98% 74% (10 dB SNR)
CNC Whole word Phoneme	 2% 7%	 84% 91%	 88% 95%
HINT	1%	100% 95% with 10 dB SNR	100% DNT
Four-Choice Spondee	60%	100%	
BKB-SIN			5.5 dB SNR

Summary

- Hearing loss can present as conductive, mixed, or sensorineural in nature
- Hearing loss can remain stable, fluctuate or rapidly change
- When EVA is suspected, use all clinical tools to help decipher a conductive or mixed component of middle ear origin from inner ear origin
- Review potential precipitating hearing loss factors when with patients and families
- Monitor hearing, hearing aid performance and/ or cochlear implant programming ***closely*** for potential fluctuations

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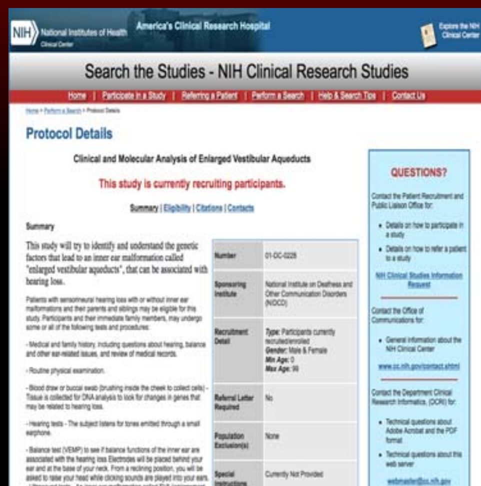
NIH Research

http://clinicalstudies.info.nih.gov/detail/A_2001-DC-0228.html



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