

Developing an Evidence Based Referral Protocol for Early Diagnosis of Vestibular Schwannomas

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- The views expressed in this presentation are those of the author and do not reflect the policy of the Department of the Army, Department of Defense, or US Government.



Introduction

- Vestibular Schwannoma (VS)
 - Slow growing benign tumor
 - Develops on Vestibulocochlear Nerve (CN VIII)
 - Overproduction of Schwann Cells
 - Wrap around nerve fibers
 - Help support and insulate for increased conduction
 - Common Symptoms (Cummings et al., 2005)
 - Hearing loss, Tinnitus, Vestibular, Aural Fullness, Otalgia
 - CN VII (Facial) or CN V (Trigeminal)
 - Labyrinthine artery
 - Incidence of 1 in 100,000 (NIDCD, 2004)

Introduction

- Tests that can potentially indicate the presence of a VS
 - Audiologic (Pure tone thresholds, speech discrimination testing, acoustic reflex thresholds, acoustic reflex adaptation (decay))
 - Vestibular (ENG/VNG, rotary chair testing, VEMP)
 - Evoked Potentials (ABR, ECoChG)
 - Radiologic (MRI, CT Scan)
- Most definitive test for diagnosing VSs
 - Contrast-enhanced MRI
 - Contrast (typically gadolinium) - provides a greater contrast between normal and abnormal tissue (Cummings et al., 2005)

Literature Review

- Previous Referral Criteria
 - Obholzer et al. (2004)
 - Unilateral hearing loss: ≥ 15 dB HL at two adjacent frequencies
 - Bilateral hearing loss: ≥ 20 dB HL at two adjacent frequencies
 - Sheppard et al. (1996)
 - ≥ 15 dB HL asymmetry
 - Unilateral tinnitus in the presence of normal hearing

Data Collection

- All data were collected retrospectively
 - Age/Gender
 - Previous Hx of Hazardous Noise Exposure
 - Hx of hearing loss (newly identified, stable, progressive, none)
 - Who referred them for a MRI (Audiology, ENT, Neurology, etc.)
 - Patient's presenting symptoms
 - Audiologic test results
 - Pure tone thresholds (excluded if air-bone gaps)
 - Speech discrimination scores
 - Tympanometry* (excluded if abnormal)
 - Acoustic reflex thresholds
 - Acoustic reflex adaptation (decay)
- Only demographic exclusion criteria: No patients < 18 years of age

Participants

- Original chart review – 628 patients
- Final participant groups ($N = 328$)
 - Positive MRI: 19 patients (age 31-73, $M = 53.37$)
 - Males ($n = 10$)
 - Females ($n = 9$)
 - Negative MRI: 309 patients (age 20-85, $M = 48.28$)
 - Males ($n = 221$)
 - Females ($n = 88$)

Referral Breakdown

- Audiology
 - Hearing Loss – 114
 - Tinnitus Only – 4
 - Dizziness/Vertigo Only – 0
- ENT
 - Hearing Loss – 160
 - Tinnitus Only – 16
 - Dizziness/Vertigo Only – 12
- Neurology
 - Hearing Loss – 111
 - Tinnitus Only – 0
 - Dizziness/Vertigo Only – 5
- PCM
 - Hearing Loss – 110
 - Tinnitus Only – 7
 - Dizziness/Vertigo Only – 4

Results/Conclusions

- Evaluation
 - Case Hx
 - Diagnostic Testing
 - Pure tones (air and bone)
 - Word recognition measures (in quiet and/or noise and to include measure of PIPB Rollover)
 - Immittance evaluation
 - Tympanometry
 - Acoustic Reflex Thresholds (specifically contralateral acoustic reflexes)
 - Acoustic reflex decay (adaptation)
 - Vestibular Testing (as needed based on presenting symptoms)

Results/Conclusions

- Diagnosis

- Presenting symptoms of unilateral/asymmetric hearing loss and unilateral/asymmetric tinnitus **AND** one or more of the following test outcomes
- Pure tone threshold asymmetry of 10 dB or more at three adjacent frequencies
- Asymmetric word recognition scores (not explained by hearing loss)
- Positive (measured) rollover
- Elevated or absent contralateral acoustic reflexes (not explained by degree of hearing loss, e.g., in the presence of a conductive component to the hearing loss).

Results/Conclusions

- If the patient has either unilateral/asymmetric hearing loss (asymmetry of 10 dB or more at three adjacent frequencies) **or** unilateral/asymmetric tinnitus with no other significant test results or symptoms the audiologist can choose to:
 - Based on clinical judgment of the specific case, refer the patient for an MRI, or
 - Refer for medical evaluation for evaluation of other non-retrocochlear (medical) etiology.

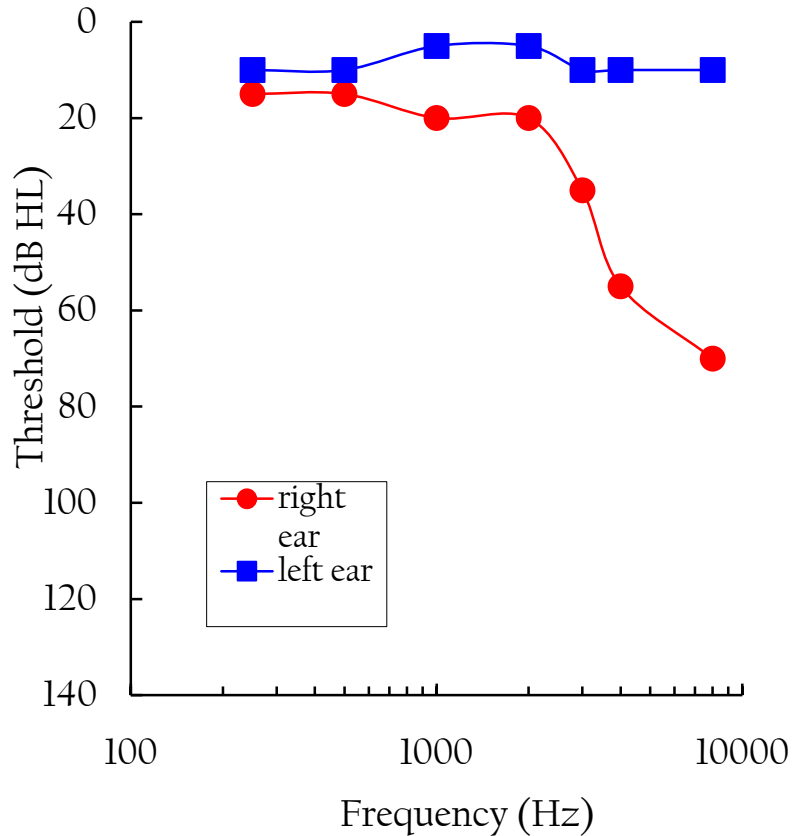
Case 1

- 32 y.o. M; Blackhawk Mechanic
- Hx of PE tubes as a child
- Seen in FEB 2008 for hearing screening
 - Identified as normal with no subsequent referral
 - Moderate loss noted at 6 kHz in the right ear only
- Seen by PCM in FEB 2008 for
 - Earache AD
 - Pressure/fullness with inability to pop AU
 - Otoscopy: Normal AD; Erythema noted AS
 - Tx for OM
- Seen by PCM in NOV 2008
 - Sudden onset (2 days prior) of bifrontal headache
 - Sudden hearing loss AD (1 week prior), although the pt. reported suspected hearing loss AD for 1 year w/o evaluation
 - Earache AD
 - Sore Throat – 2 days
 - Congestion
 - Evaluation
 - Hemorrhagic discharge noted AU
 - Right Otoscopy – decreased mobility with signs of fluid
 - Left otoscopy – unremarkable
 - Tx for OM and referred to Audiology for chronic hearing loss AD

Case 1

- Hearing Screening in DEC 2008
 - Moderate to severe responses AD above 2 kHz
- Initially seen in DEC 2008 in ENT
 - Subjective right sided hearing loss (HL)
 - HL often accompanied by
 - Crackling sounds
 - Sharp pains lasting seconds with rapid, spontaneous resolution
 - No balance complaints
 - Intermittent right ear tinnitus
 - Refer to Audiology

Case 1

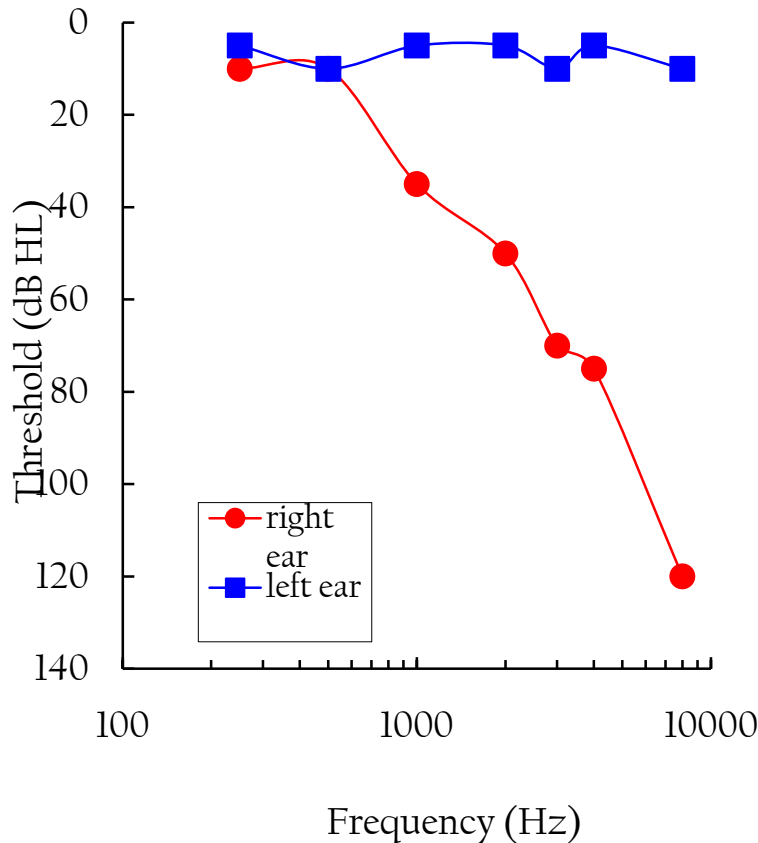


- OAEs
 - Present/robust AS
 - Largely absent above 1 kHz AD
- WR
 - AS: 100% @ 40 dBHL
 - AD: 98% @ 45 dBHL
- Normal tympanograms AU
- ARTs (I) – present AS (.5, 1, and 2 kHz) and AD (.5 and 1 kHz)

Case 1

- ENT Ordered MRI of the IACs with gadolinium enhancement
- Impression: 1.8 cm anterior-posterior x 2.5 cm transverse x 1.8 cm craniocaudal right cerebellar pontine angle mass. The lesion expands into the internal auditory canal. Some erosion of the petrous apex of the right temporal bone was also noted

Case 1



- WR
 - AS: 100% @ 50 dBHL
 - AD: 64% @ 50 dBHL; 96% @ 90 dBHL
- Normal tympanograms AU
- ARTs
 - Probe AS - Normal
 - Probe AD
 - Ipsi – Normal (.5, 1 kHz); Absent (2 kHz)
 - Contra – Elevated (.5 kHz); Absent (1, 2 kHz)
- Negative Acoustic Reflex Decay

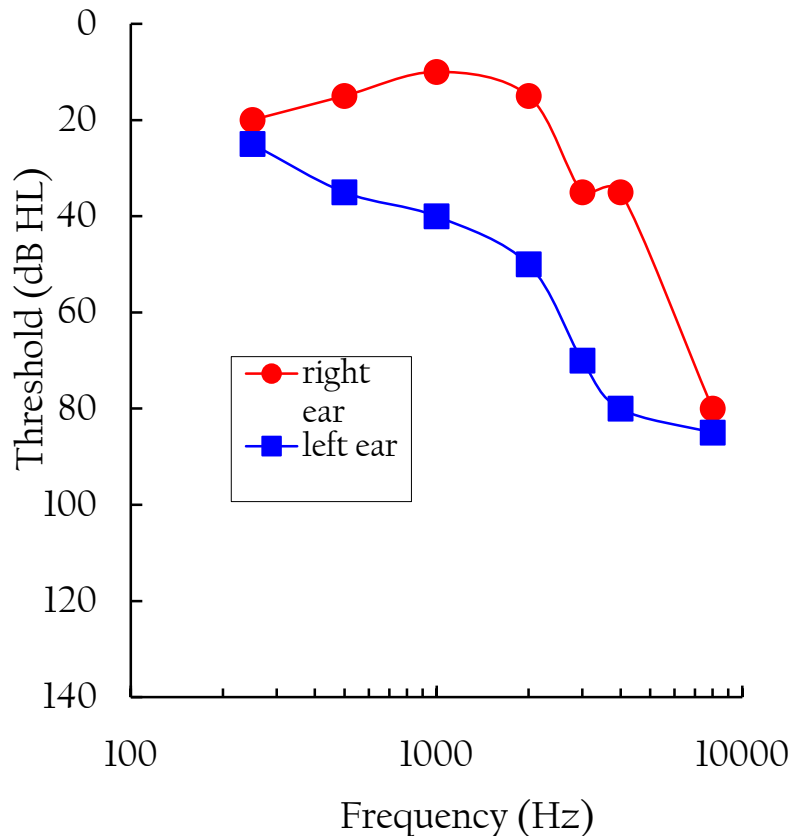
Case 1

- VS was removed in AUG 2009
 - Post-occipital approach with total severance of auditory/vestibular nerve
- Complication of CSF seroma
- Migraines – successfully tx with Floricet and caffeine pills
- Pt. initially had facial nerve paresis which is almost resolved by DEC 2009
- Also c/o balance issues and blurry vision which has also improved
- Pt. discussed BAHA vs. CROS aid
- ENT noted only a small portion of bone to work with for BAHA considering his craniotomy

Case 2

- 60 y.o. M initially seen in Audiology in June 1987
 - Presented with
 - Dysequilibrium problems
 - Hearing loss AS>AD
 - Distortion AS
 - Intermittent pressure in his head
 - Tinnitus AS

Case 2



- WR
 - AD: 96% @ 55 dBHL; 92% @ 90 dBHL
 - AS: 50% @ 65 dBHL; 44% @ 90 dBHL
- Normal tympanograms AU
- ARTs
 - Probe AD - Normal
 - Probe AS
 - Ipsi – Normal/Elevated (.5, 1, 2 kHz)
 - Contra – Normal (.5 kHz); Elevated/Absent (1, 2 kHz)
- Negative Acoustic Reflex Decay

Case 2

- ABR – June 1987

- AD

- Absolute wave latencies were delayed for Waves III and V.
 - Morphology was considered fair

- AS

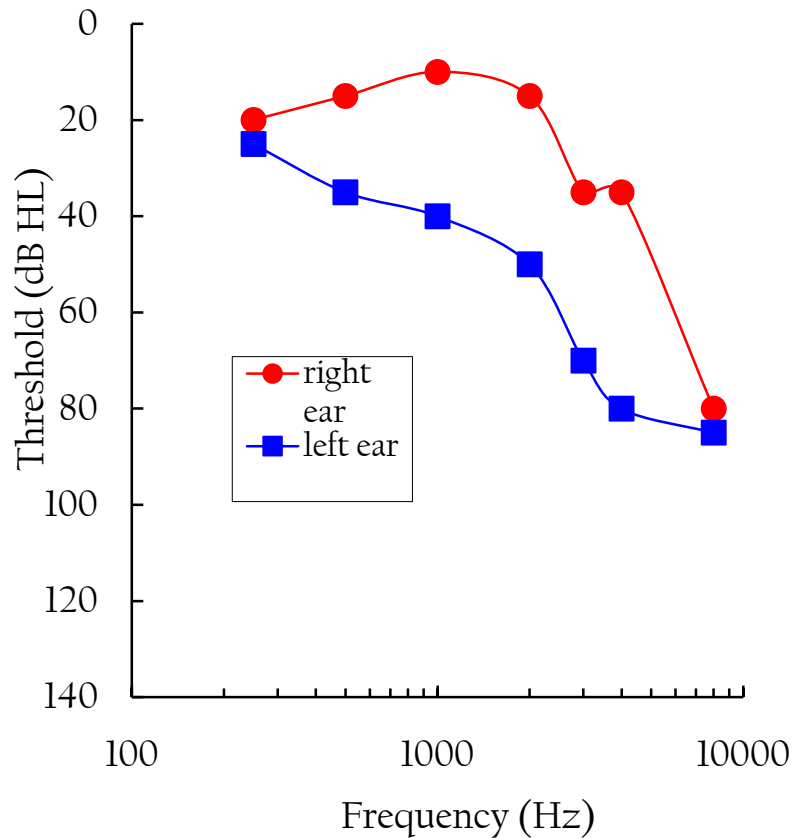
- Absolute latencies were delayed for Waves III and V.
 - Comparison between ears indicated significant delay for Wave V in the left compared to the right
 - Morphology was considered poor

Case 2

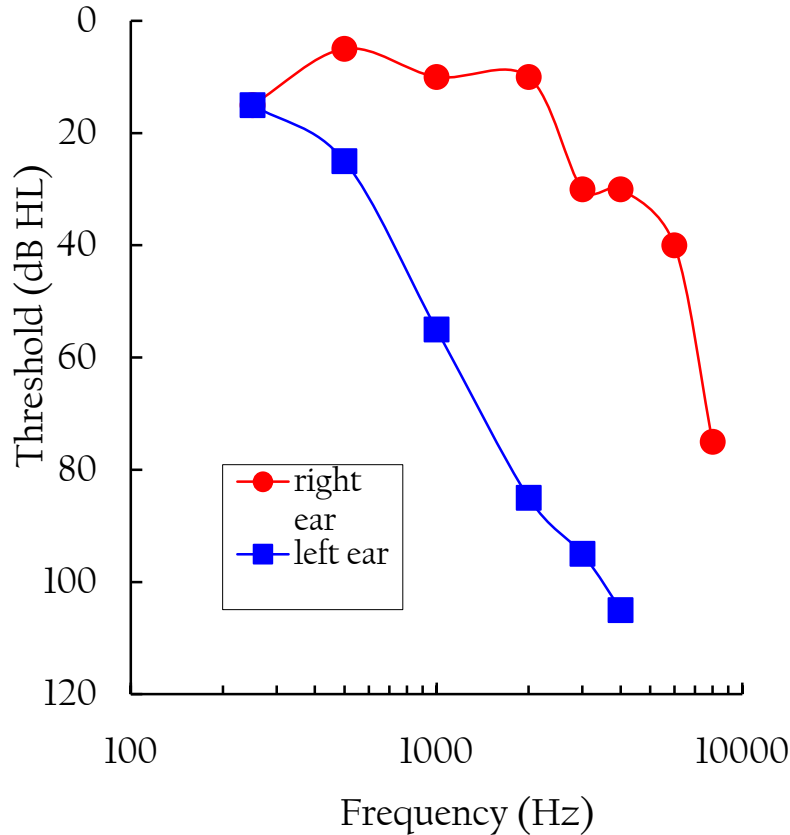
- Vestibular Evaluation in July 1987
 - Ocular Motor Evaluation
 - Significant for saccadic smooth pursuit at low frequencies
 - No spontaneous nystagmus
 - Dix Hallpike right – right beating, non-latent nystagmus with no symptoms. Not classic for BPPV
 - Dix Hallpike left – Normal
 - Direction fixed, right beating nystagmus of 7-9 deg/sec seen in head/body right positions
 - Bithermal calorics – 44% left weakness

Case 2

- Stable SNHL since JUN 1987
- WR
 - AD: 100% @ 50 dBHL
 - AS: 60% @ 60 dBHL



Case 2



- Significant decreased thresholds noted AS
- WR
 - AD: 96% @ 50 dBHL; 96% @ 90 dBHL
 - AS: 28% @ 55 dBHL; 8% @ 90 dBHL
- Normal tympanograms AU
- ARTs
 - Probe AD – Normal
 - Probe AS – Essentially Absent
- Negative Acoustic Reflex Decay – Probe Right Only

Case 2

- Referred for MRI by ENT
 - Noted a 1 x 1.5 cm acoustic neuroma on the left side

Case 2

- ENG evaluation in NOV 1990
 - Ocular Motor tests were normal
 - No spontaneous
 - No positional nystagmus
 - Negative Dix Hallpikes
 - Rotary Chair – abnormal phase leads and decreased gain

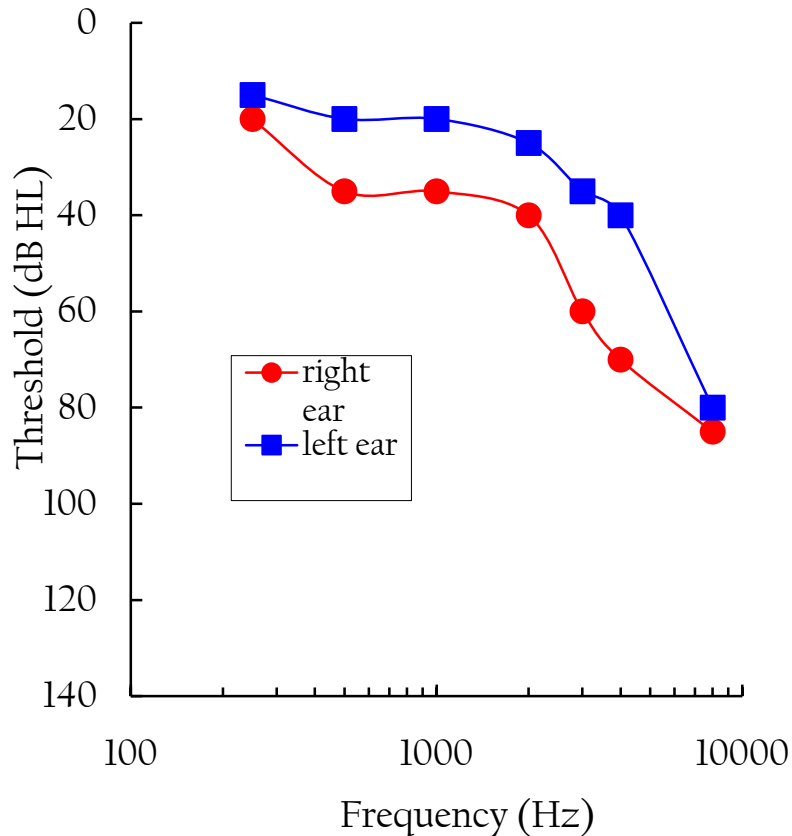
Case 2

- Removal of VS in NOV 1990.
- Audio was unchanged AD with an anacoustic AS.
- Pt. reports no significant hearing difficulties

Case 3

- 73 y.o. M
- Presented to Audiology in DEC 2008
- Previous Noise Exposure
- Fm hx: Father and brother have both had different brain tumors
- Presentation:
 - Hearing loss
 - Difficulty understanding speech
 - No earache
 - No tinnitus
 - No balance difficulties

Case 3



- WR
 - AS: 100% @ 90 dBHL
 - AD: 84% @ 75 dBHL; 48% @ 90 dBHL
- Normal tympanograms AU
- ARTs
 - Probe AS – Normal, except Absent at 2 kHz in contra condition
 - Probe AD – Absent

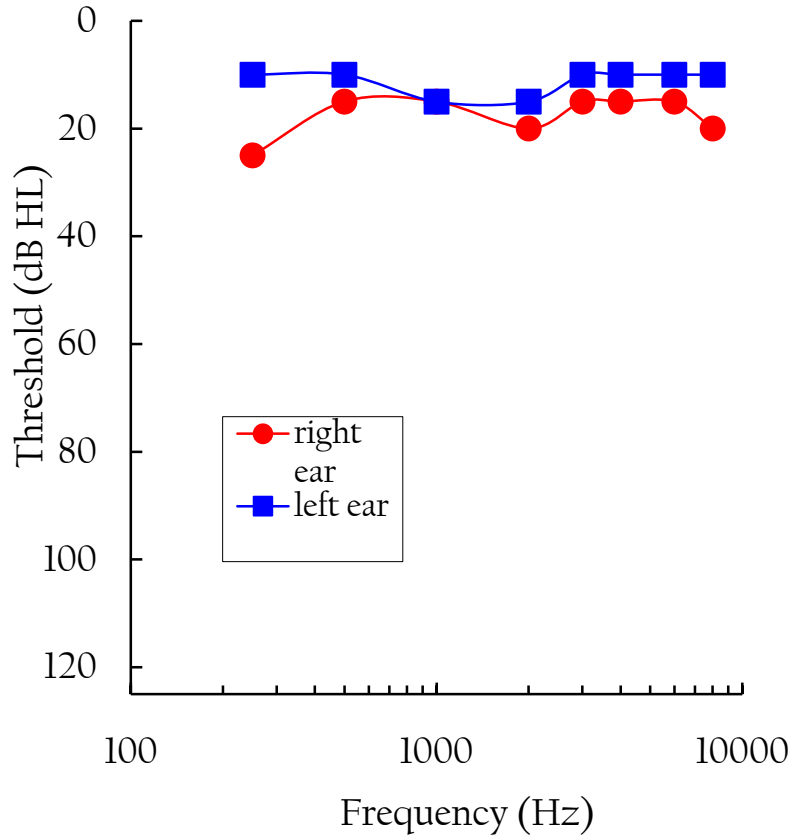
Case 3

- Negative MRI of the IACs
- Pt. was fit with hearing aids in both ears

Case 4

- 42 y.o. F
- Pt. initially presented to Audiology in OCT 2005 for a second opinion on a Meniere's Dx
- Presentation
 - No subjective hearing loss
 - Intermittent tinnitus
 - Vertigo, noted since Spring 2005
 - No ear pain

Case 4



- WR
 - AS: 92% @ 80 dBHL
 - AD: 100% @ 80 dBHL
- Normal tympanograms AU
- ARTs
 - Contralateral right and left reflexes were WNL
- Negative Acoustic Reflex Decay in both ears

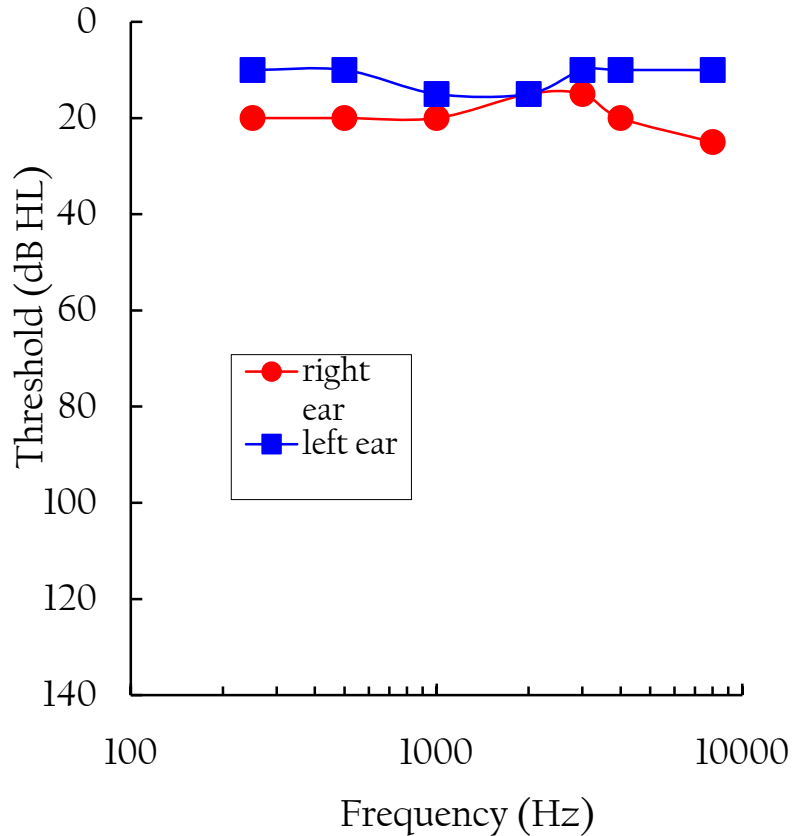
Case 4

- Next OTO-HNS note is from 5/2006
- Presentation
 - Vertigo following a sinus infection 4 weeks prior
 - Vertigo lasted 2.5 weeks. It began to resolve but not entirely
 - Today, she could say her symptoms occur looking to the right
 - Intermittent tinnitus (non-localized)
- Conclusion
 - Audiogram
 - Consult for VRT
 - If no improvement consider MRI

Case 4

- Seen in Audiology in May 2006
 - Presentation
 - Subjective hearing loss AD
 - Muffled sound AD
 - Pressured sensation AD
 - Tinnitus AD only

Case 4



- Stable hearing
- Excellent high level word recognition in both ears
- Normal tympanograms and ARTs

Case 4

- Seen again in ENT in AUG 2006
 - Positive Romberg to the right
- Suspected BPPV; however ordered MRI to rule out intracranial pathology considering
 - Unilateral tinnitus
 - Abnormal Romberg

Case 4

- MRI of the IACs
 - 3 x 4 mm lesion noted in the right IAC involving the right CN XIII

Case 4

- Seen in DEC 2007 in Audiology for monitoring
 - Severe Tinnitus AD
 - Difficulty understanding speech especially in background noise
 - Stable Normal hearing noted AU; Excellent WR with no evidence of rollover; Normal tympanograms and ARTs in both ears

Case 4

- FEB 2008 (Audio and Vestibular Testing)
 - Presentation
 - Constantly imbalanced
 - Positional vertigo
 - Vertigo to temperature changes
 - Uses CC on television
 - Right severe tinnitus
 - Has begun balance training

Case 4

- DHI 68/100
- Audiogram
 - Stable, normal thresholds
 - Excellent word recognition w/ no evidence of rollover
 - Normal tympanograms
 - Normal acoustic reflexes
 - Negative acoustic reflex decay in both ears
- ABR – Normal
- Normal OAEs
- No spontaneous
- Positional – not clinically significant
- Negative Dix Hallpike
- Left beating head shake nystagmus
- Calorics – 76% right unilateral weakness
- Normal VEMPs
- Slightly abnormal posturography. Normal sensory analysis

Case 4

- Removal of VS in APR 2008 via a middle fossa approach
 - Persistent tinnitus
 - Mild imbalance that is improving
 - Intermittent echo sound AD with occasional tinny quality
 - Some right ear pain behind the right pinna

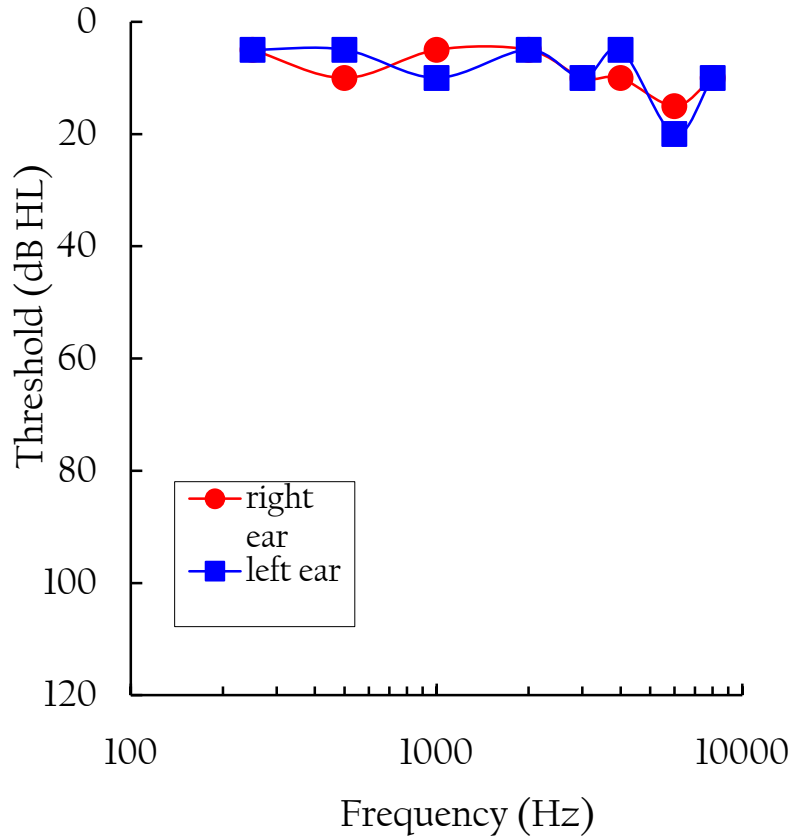
Case 4

- MRIs in 2009 have indicated a 7mm lesion in the right IAC
 - Residual lesion vs. granulation tissue
- Newest Audio in SEPT 2009 was essentially normal
- ENT in SEPT 2009
 - Right eye is “slow”
 - Pain in the medial side of R eyebrow
 - Mildly improved tinnitus
 - Dry eyes
 - Difficulty with taste on the right side of her tongue
 - Mild preauricular numbness and pain AD
 - Twitch in right eye tx with botox that is more tolerable

Case 5

- 37 y.o. M
- Presented to Audiology Clinic in JAN 2010
- C/o
 - Intermittent ear pain AS
 - Episodic, sharp pain
 - Mostly AS, but rarely noted AD
 - Pain for 10-15 seconds
 - Randomly several times a week
 - No hearing loss
 - No tinnitus
 - No dizziness
 - Ears frequently feel like there is cotton in them

Case 5



- WR
 - AS and AD: 100% @ 50 and 90 dBHL
- Normal tympanograms AU
- ARTs - WNL
- Negative Acoustic Reflex Decay in both ears

Case 5

- Initially had CT scan which was normal in DEC 2009
- Sent for MRI of the IACs for chronic left ear pain in JAN 2010
- Impression: 1.5 mm right intracanicular VS

References

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Thank You!!

