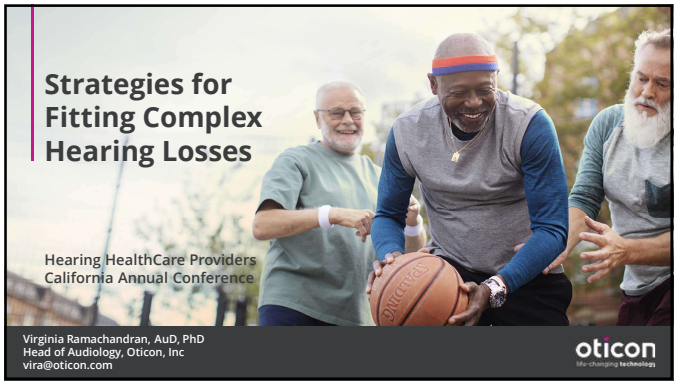
Strategies for Fitting Complex Hearing Losses
Hearing HealthCare Providers California Annual Conference
Virginia Ramachandran, AuD, PhD
Head of Audiology, Oticon, Inc
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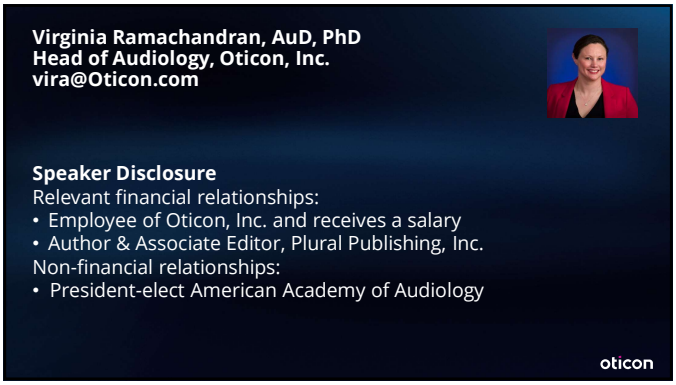
Virginia Ramachandran, AuD, PhD
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Speaker Disclosure
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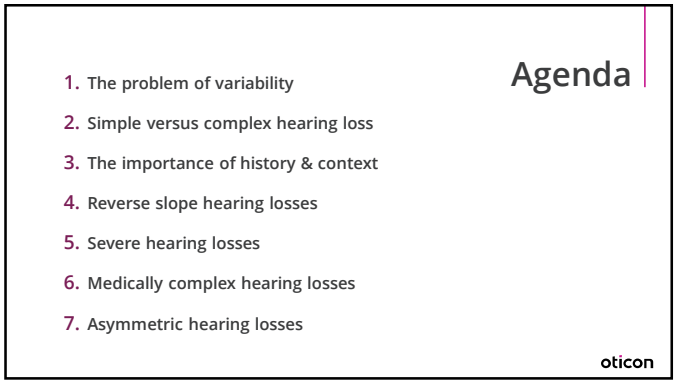
Agenda
1. The problem of variability
2. Simple versus complex hearing loss
3. The importance of history & context
4. Reverse slope hearing losses
5. Severe hearing losses
6. Medically complex hearing losses
7. Asymmetric hearing losses
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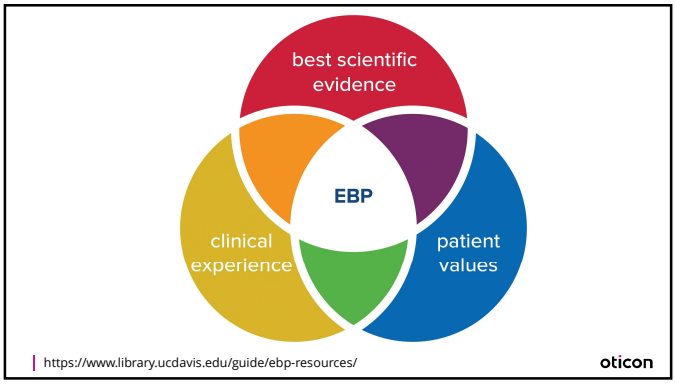
best scientific evidence
EBP
clinical experience
patient values
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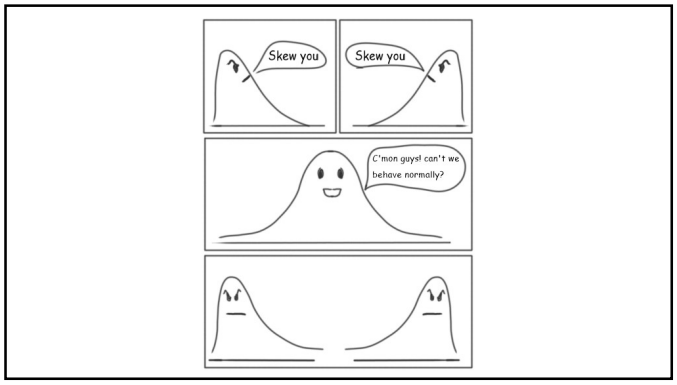

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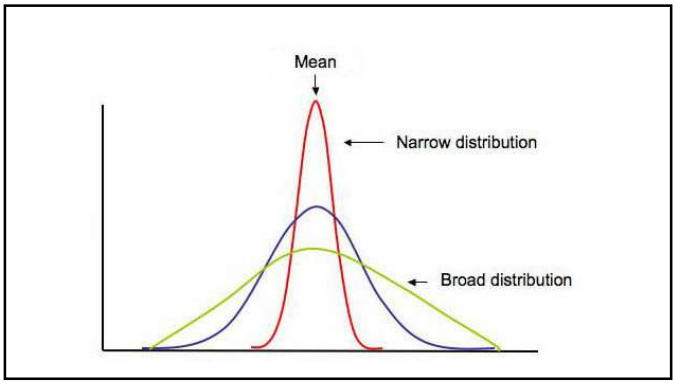

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Variability

- High variability in a "normal" population
  - Some populations more variable than others
- High variability due to diversity in a population
  - Diversity necessarily introduces variability
  - Diversity of population characteristics
  - Diversity of etiology

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WOMEN'S HEALTH RESEARCH ▾ SEX & GENDER ▾ IN THE SPOTLIGHT ▾ SCIENCE POLICY ▾ CAREER

Related to sex-specific reporting, but distinct from inclusion, NIH announced in 2014 a new policy requiring that preclinical research consider sex as an important biological variable (Clayton and Collins 2014) in both vertebrate animal and human studies. Under the new policy, in effect as of January 2016, applicants would be asked to "explain how relevant biological variables, such as sex, are factored into research designs and analyses." Strong justification from the scientific literature, preliminary data, or other relevant consideration is required from researchers planning to study only one sex. The absence of evidence about differences between males and females in previously published research could not serve as a justification for as single sex study. (See Consideration of Sex as a Biological Variable in NIH-funded Research, companion reference to NIH Guide Notice NOT-OD-15-102 (PDF - 74.6KB)).

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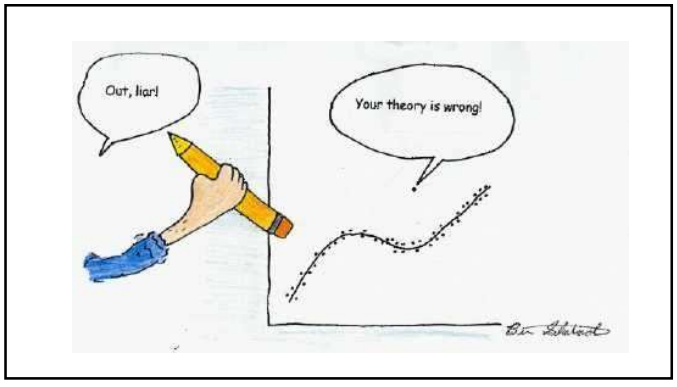

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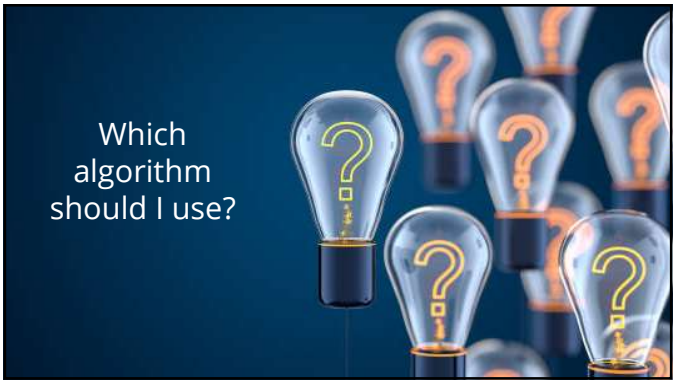

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What are some assumptions we make?

- All speech is valuable
- Restore audibility as much as possible
- Prescriptive approach
- Correct for threshold loss
- Measurable hearing is useable hearing
- Make the full range of inputs fit
- The more bandwidth the better
- Both ears contribute equally

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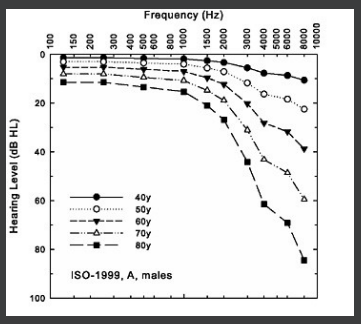
Normal Hearing Loss



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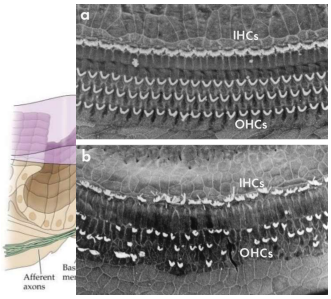
Sensorineural hearing loss

Something causes reduction of sound intensity due to damage to the cochlear mechanisms

Damage to the cochlear mechanisms also causes distortion because the original sound cannot be coded faithfully

In a **SIMPLE** SNHL, the damage is limited primarily to the outer hair cell (OHC) function, resulting in reduced sensitivity to sound and impaired fine-tuning of the signal



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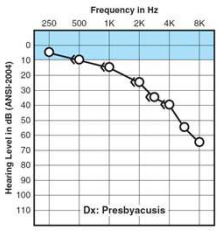
Audiometric results – simple SNHL

- Reduced air- and bone-conduction thresholds. No meaningful/significant difference between bone-conduction and air-conduction responses
- In simple SNHL, speech perception (WRS) generally reduced only in proportion to the audiometric thresholds*
- *Unless person older (around 80 years of age)



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Etiologies – simple SNHL

- Age
- And maybe…
- Some levels of noise exposure
- Some levels of ototoxicity
- Some levels of oxygen deprivation
- Some genetic disorders

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Complex sensorineural hearing loss

Something causes reduction of sound intensity due to damage to the cochlear mechanisms, auditory nerve, or central auditory nervous system (CANS)

Results in reduced sensitivity to sound. May results in severely impaired fine-tuning of the signal, AND some aspects of the signal that cannot be represented to the brain at all

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Audiometric results – COMPLEX SNHL

- Reduced air- and bone-conduction thresholds. May or may not be differences between bone- and air-conduction responses (aka an air-bone gap)
- Any bone-conduction thresholds greater than about 40 dB or so begins to be complex because IHCs necessarily involved
- In complex SNHL, speech perception (WRS) may be reduced disproportionally to the audiometric thresholds

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Etiologies – COMPLEX SNHL

- Endless…
- Severe noise exposure (acoustic trauma)
- Physical trauma (temporal bone trauma, cotton swab accidents)
- Severe ototoxicity (most often chemotherapy)
- Oxygen deprivation
- Infectious diseases (measles, mumps, maternal rubella, maternal CMV, meningitis, "viral")
- Acoustic neuroma/vestibular schwannoma/meningioma
- Autoimmune diseases
- Physical anomalies affecting structure (semi-circular canal dehiscence (SCD), enlarged vestibular aqueduct (EVA))
- Physical anomalies affecting function (maternal CMV, Usher's)
- Cochlear otosclerosis
- Unknown etiology

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The importance of history & context

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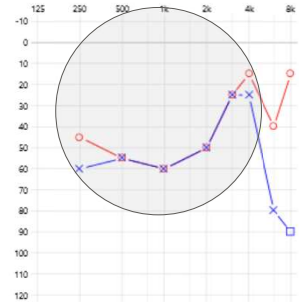

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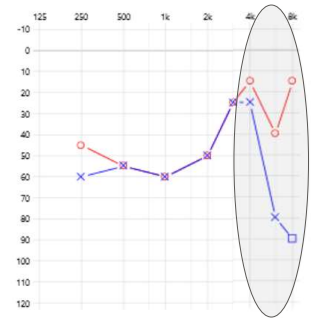

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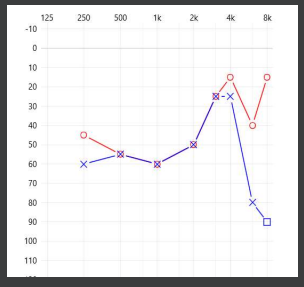

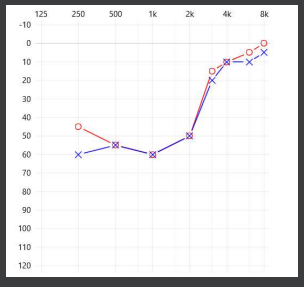

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Newborn infant hearing screening

AUDITORY BRAINSTEM RESPONSE

OTOACOUSTIC EMISSIONS

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When should I switch from a pediatric to an adult algorithm?

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Reverse slope hearing loss

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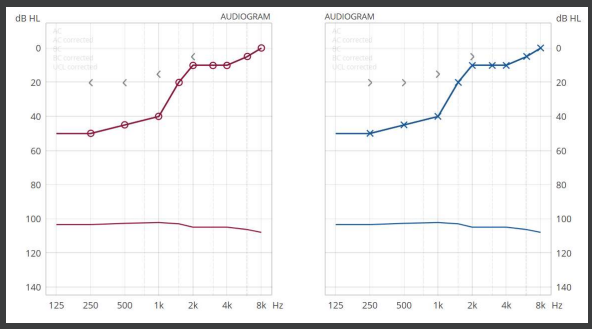

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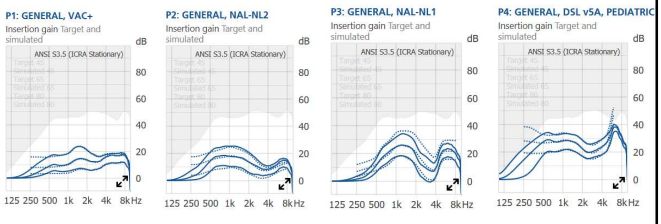

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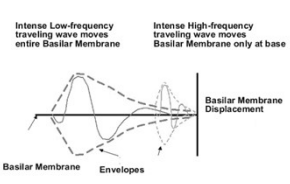
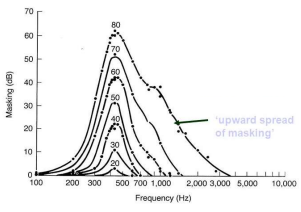
Upward spread of masking





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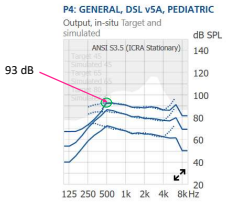
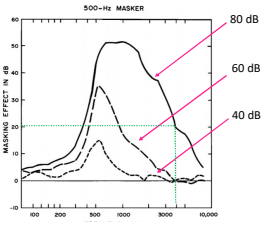
Prescriptive settings





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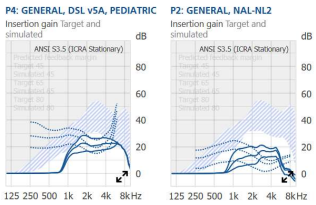
Alternatives



- Open dome
- Mid- and high-frequency emphasis
- Mid-frequency emphasis

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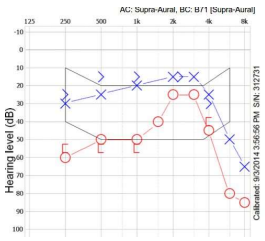
Meniere's Disease

AKA Endolymphatic Hydrops
- Name representative of disorder as due to buildup of endolymphatic fluid in membranous labyrinth of inner ear

Characteristics:
- Asymmetric sensorineural hearing loss
- Low-frequency (reverse slope) hearing loss
- Fluctuating hearing loss
- Poorer than expected speech understanding
- Low-pitch roaring tinnitus
- Dizziness/vertigo (nausea/vomiting)
- Feeling of fullness in the ear



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Severe hearing loss

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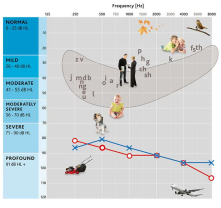
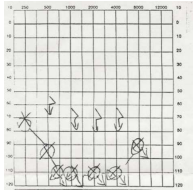
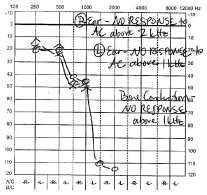
Audiometric results – Severe SNHL







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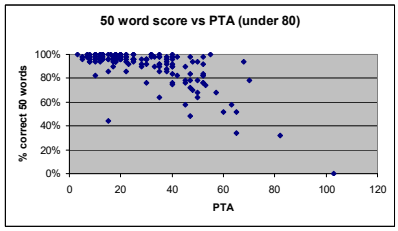
Speech Understanding



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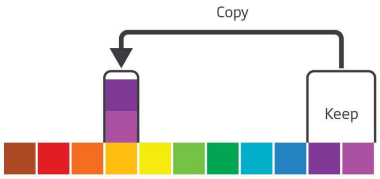
Frequency-lowering



The Copy and Keep strategy gives patients access to usable high frequency cues even when frequency lowering is utilized.

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Medically complex hearing losses – the importance of diagnostics

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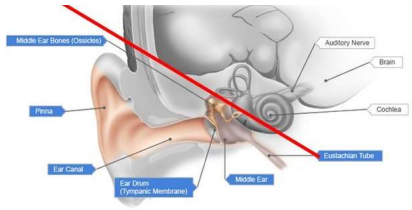
Conductive hearing loss

Something causes reduction of sound intensity as it travels to the cochlea



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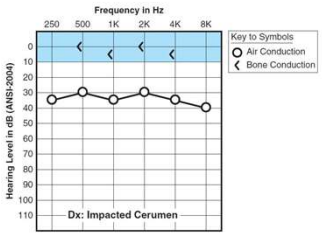
Audiometric results

- Differences between bone-conduction and air-conduction responses
- Speech perception (WRS) generally not reduced
- Maximal conductive hearing loss maybe up to 60 dB – generally far less



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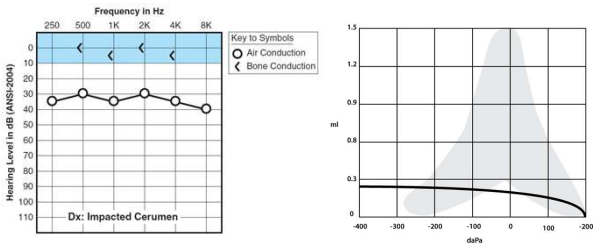

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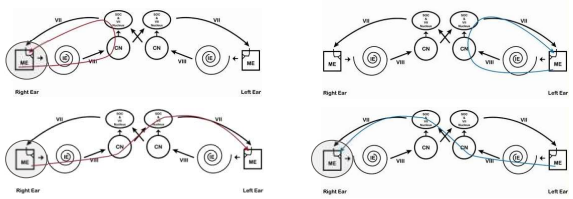
Acoustic Stapedial Reflex Thresholds with Conductive Hearing Loss



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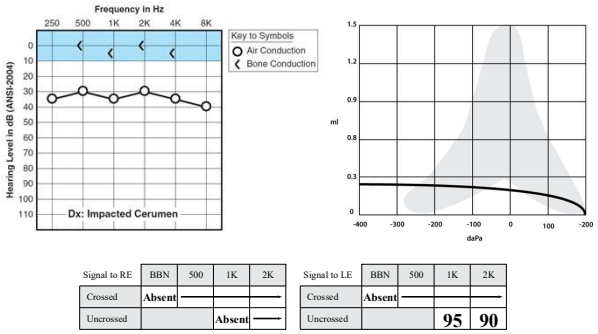

Signal to RE	BBN	500	1K	2K
Crossed	Absent			
Uncrossed			Absent	

Signal to LE	BBN	500	1K	2K
Crossed	Absent			
Uncrossed			95	90

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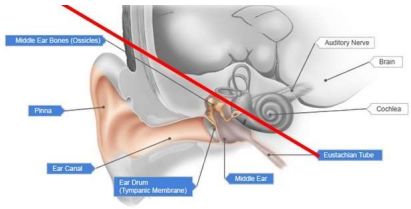
Conductive hearing loss

Something causes reduction of sound intensity as it travels to the cochlea



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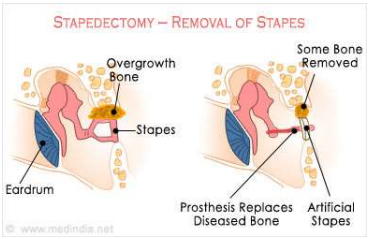
Otosclerosis



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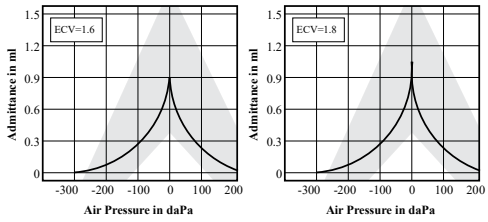
Tympanograms with Otosclerosis



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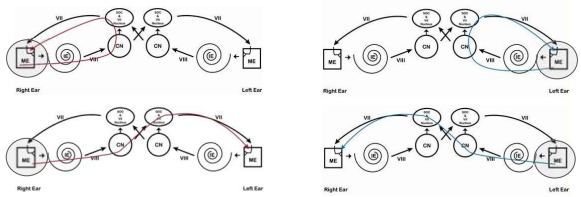
Acoustic Stapedial Reflex Thresholds with Otosclerosis



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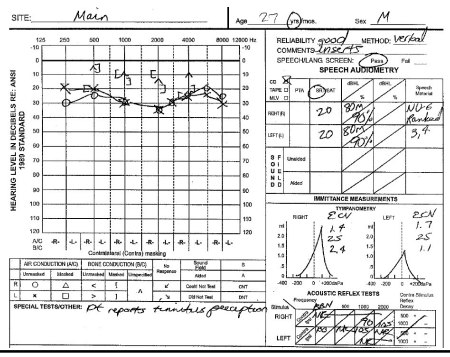

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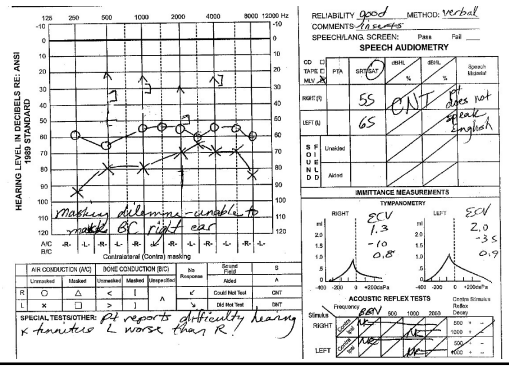

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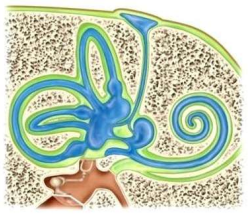
Third-Window Syndromes

- Defects in the integrity of the bony structure of the **inner ear**



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Symptoms of Third-Window Syndromes

- Vertigo
- Sound- or pressure-induced vertigo/nystagmus (Tullio and Hennebert signs)
- Can sometimes hear internal body sounds (heart beat, eye movement)
- Low-frequency air-bone gap (better than actual bone-conduction thresholds)
- Hearing loss
- Sensitivity to loud sound – hyperacusis-like symptoms
- Own voice too loud

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Causes of Third-Window Syndrome

- Enlarged vestibular aqueduct (EVA)
- Dehiscence of the scala vestibuli side of the cochlea
- Bone dyscrasias (Paget's disease, osteogenesis imperfecta)
- X-linked stapes gusher
- Perilabyrinthine fistula
- Superior Semicircular Canal Dehiscence (SSCD)

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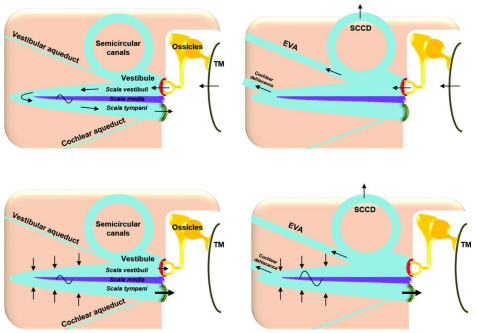

M.-L. Ho, G. Moonis, C.F. Halpin and H.D. Curtin. Spectrum of Third Window Abnormalities: Semicircular Canal Dehiscence and Be... American Journal of Neuroradiology January 2017, 38 (1) 2-9.

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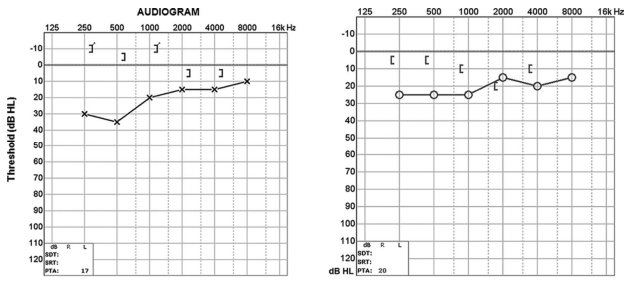

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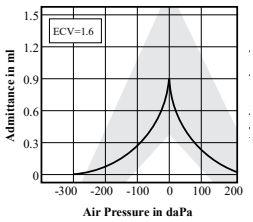
Tympanometry with Third-Window Syndrome



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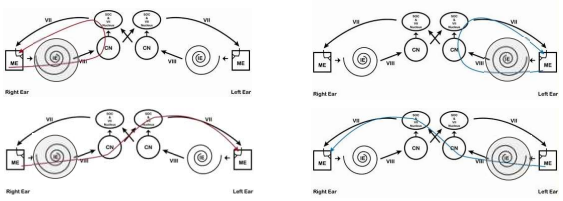
Acoustic Stapedial Reflex with Third-Window Syndrome



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Acoustic Stapedial Reflex with Third-Window Syndrome

- Reflexes **present** up to moderately-severe sensorineural hearing loss
- Reflexes **absent** above moderately-severe sensorineural hearing loss

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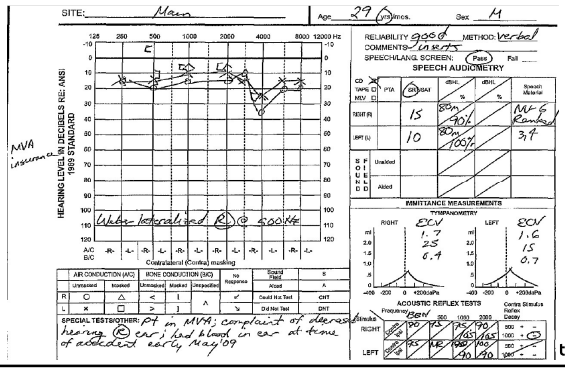

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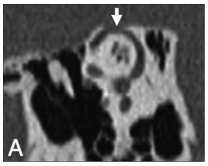

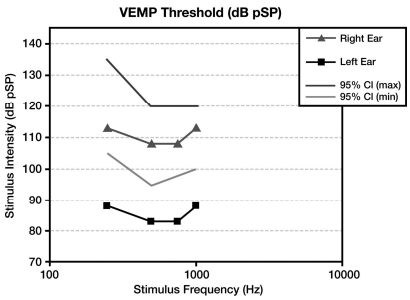

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Clinical Problem Fitting Hearing Aids

- Genie will believe that air-bone gaps indicate a conductive hearing loss and will prescribe extra gain accordingly
- These people typically have loudness tolerance problems
- The algorithm compounds the problem

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Suggestions

- To attempt to identify:
  - Ask to see any tympanograms or acoustic reflexes
  - Ask about any history of ear surgery (particularly unsuccessful ear surgeries)
  - Look for reduced word recognition scores – particularly if they are done at high levels
- To address problems:
  - Measure frequency-specific LDLs and use REM to ensure not exceeding these levels
  - Unocclude ear canal as much as possible and reduce low-frequency gain

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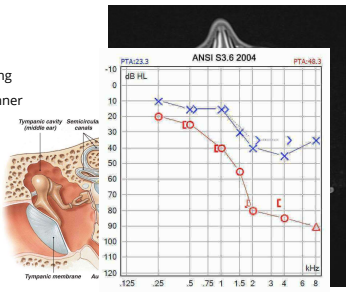
Vestibular schwannoma

- AKA acoustic neuroma
- noncancerous and usually slow-growing tumor that develops on the main (vestibular) nerve leading from your inner ear to your brain
- Characteristics:
  - Depends on location and direction of growt
  - Asymmetric high-frequency sensorineural hearing loss
  - Speech understanding poorer than expecte (rollover at high intensities)
  - Abnormal acoustic reflex thresholds; poten normal otoacoustic emissions
  - Unilateral tinnitus
  - Facial nerve problems (weakness, numbnes
  - Dizziness/imbalance



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Asymmetric Hearing Loss

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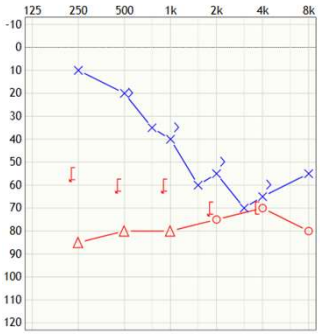

	Right	Left
SRT	90	40
WRS	0%	72%

AIDED RESULTS		
	Quiet	+10 S/N
Left Only	76%	56%
Binaural	92%	88%

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Suggestions

- Test binaurally to identify improvements or binaural interference
- Serial monaural fittings
- Fit better ear first
- Adjust gain in poorer ear to balance

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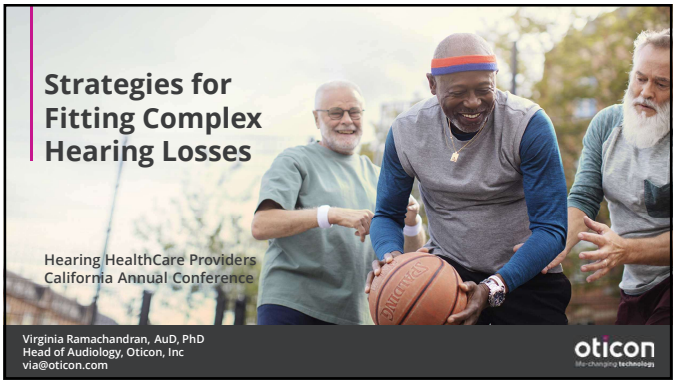

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