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CONDUCTIVE/MIXED HEARING LOSS: OTOSCLEROSIS AND OTHER CAUSES

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22 April, 2020

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Disclosures

- Financial: Daniel Zeitler is an Otologist/Neurotologist at Virginia Mason Medical Center in Seattle, WA. He received an honorarium for presenting this course.
- Non-financial: Daniel Zeitler has no relevant non-financial relationships to disclose.

2

Learning Outcomes

After this course, participants will be able to:

- Discuss etiologies for conductive hearing loss with an intact tympanic membrane and without middle ear disease.
- Describe the differences and similarities between conditions.
- Explain an overview of audiological and otologic work-up.

3

4

MY OBJECTIVE WITH CHL/MHL…

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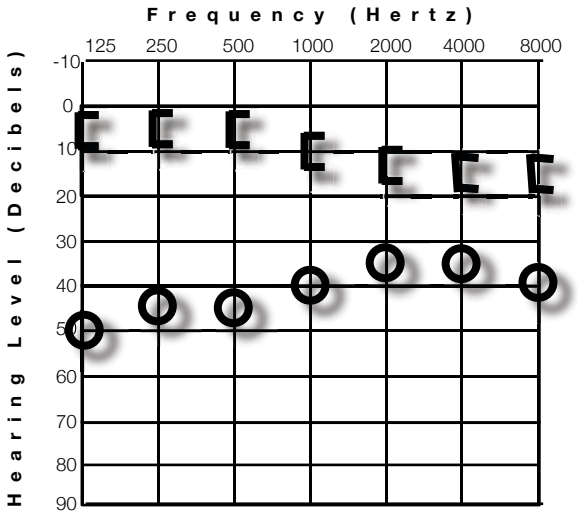

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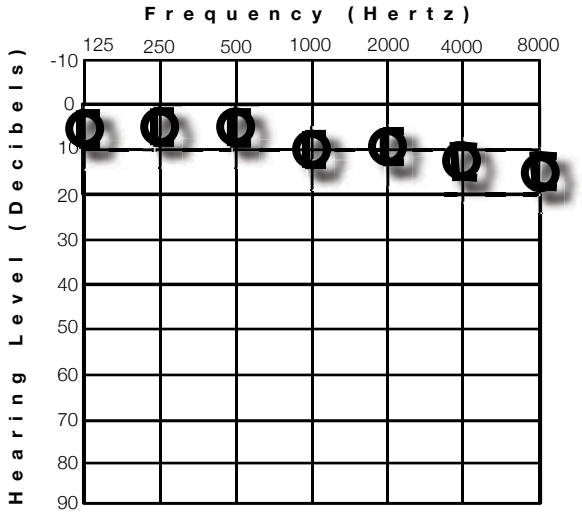

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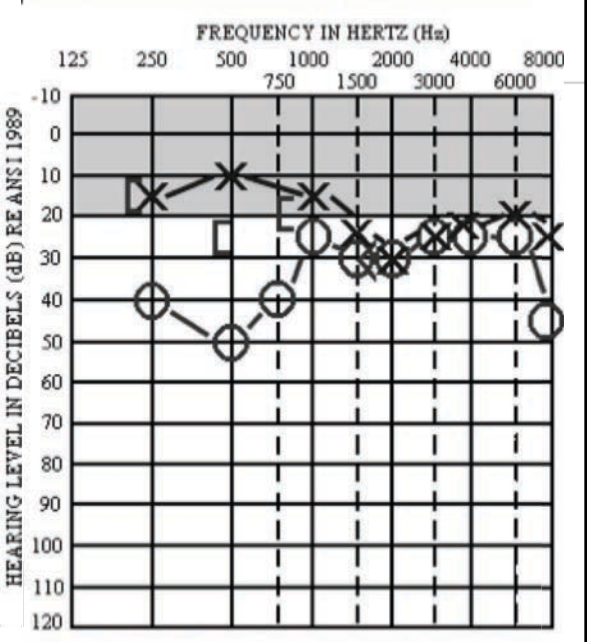
Probably Otosclerosis



8

Middle ear mechanics

9

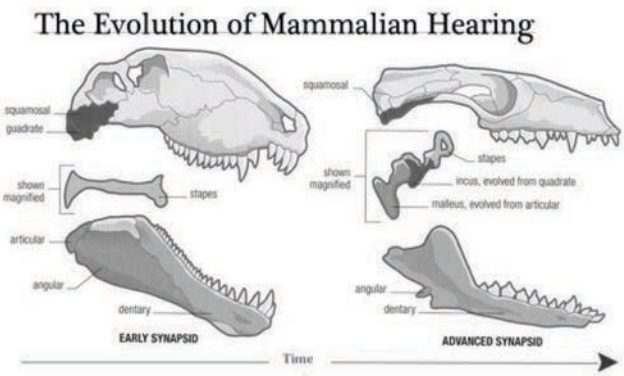
Function of the middle ear

- Transform acoustic waves into mechanical vibration that stimulates inner ear fluid
- For distant ancestors, the medium was water
- Evolutionary adaptation to air
- Required increased efficiency

10

Evolution of mammalian middle ear

- Exaptation: repurposing structures during evolution
- Reptiles: columella connecting the TM to the inner ear
- Stapes formation earlier and distinct
- 2 reptilian jaw bones not seen in mammals incorporated into ME
  - Articular bone (lower jaw) = malleus
  - Quadrate bone (upper jaw) = incus



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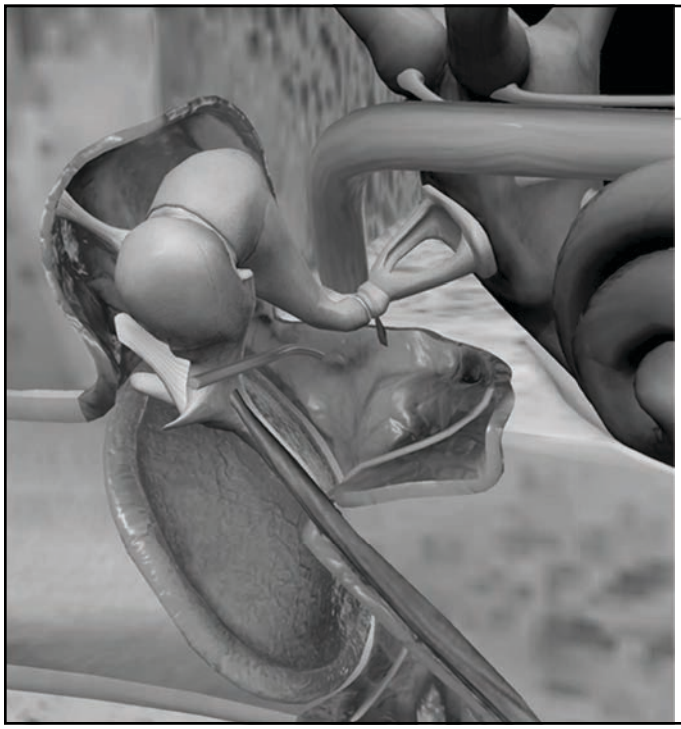
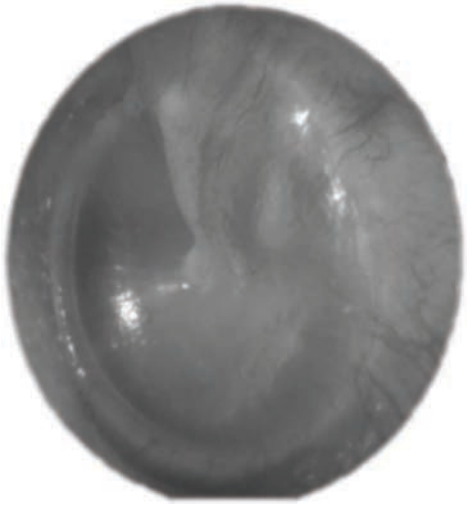


Tympano-ossicular system

12



Tympanic membrane mechanics

- 9.5 x 8.5 mm
- Circumferential and radial collagen
- Circumferential “softness” = low frequency
- Radial “stiffness” = high frequency

Q2, Q3

13

Biomechanics of tympanic membrane

- Low frequencies (< 1 kHz)
  - Entire TM in phase as unit
  - Malleus moves as lever
- High frequencies (> 3 kHz)
  - Complex, maximal displacement around umbo
  - Malleus decoupled, moves in elliptical fashion
  - Stresses importance of mobile + flexible connection between TM and malleus (manubrial fold)

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Levers of the tympano-ossicular system

- **Catenary lever** – gain realized by TM resting on manubrium while rigidly supported at annulus and acted upon by it's own weight
- **Ossicular lever** – gain provided by length of manubrium of malleus / long process of incus
- **Hydraulic lever** – size differential between the TM and oval window (20x)

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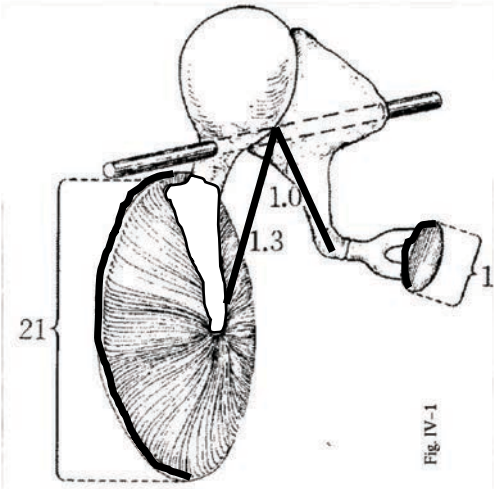
The tympano-ossicular system

Catenary lever +
Ossicular lever +
Hydrolic lever =

34 dB gain
(actual 20-25 dB)



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Why theoretical gain ≠ actual gain

1. Different portions of TM vibrate at different frequencies
2. Slippage of ossicles around lever axis
3. Forces needed to overcome stiffness and mass of TM itself
4. Reduction of middle ear space aeration which can impair impedance matching

(Rosowski et al., Am J Otol, 1996)

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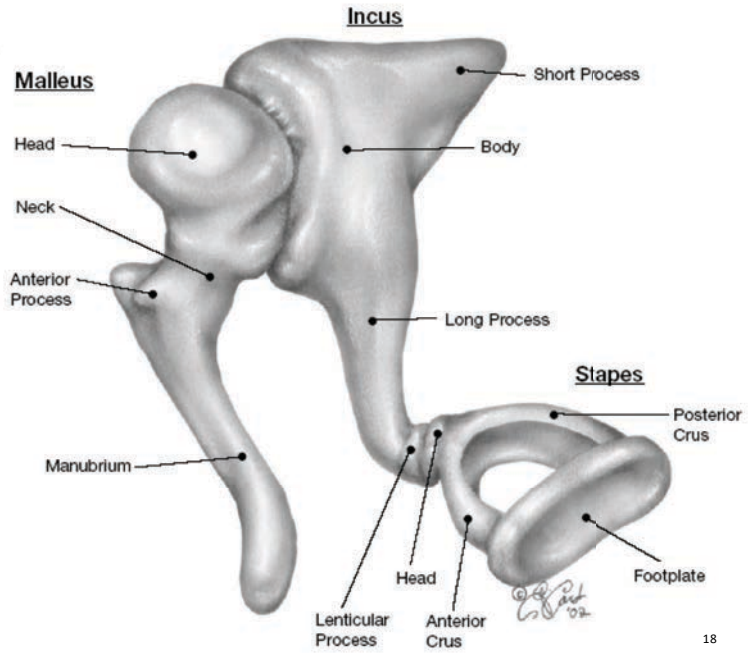
Ossicular chain



Q4

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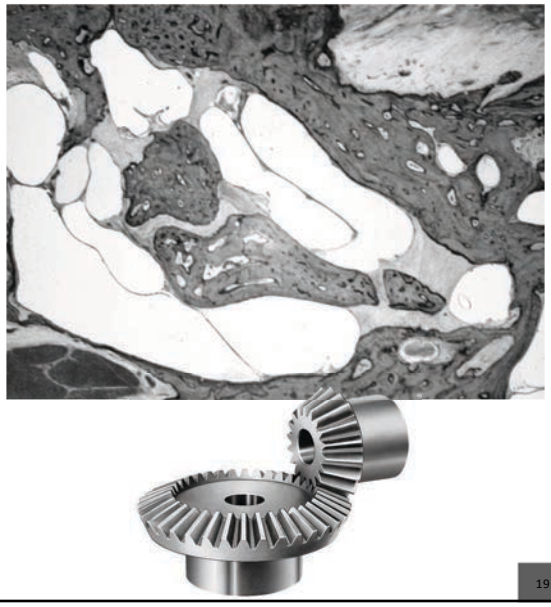
Malleoincudal joint

Diarthrodial joint: hyaline cartilage with synovial fluid

Hinge-like motion at low frequency

Twisting motion at higher frequencies ("beveled gears")



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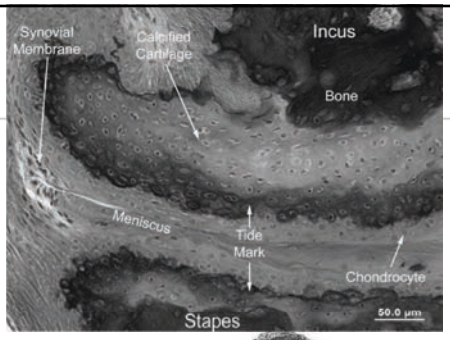
Incudostapedial joint

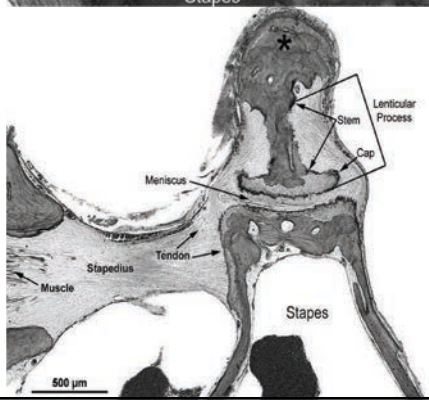
Diarthrodial joint
- Hyaline cartilage
- Synovial fluid

Articular disk
- Fibrous meniscus

Joint capsule
- Encompasses entire lenticular process
- Prevents "lift-off"

(Karmody et al. Otol Neurotol, 2009)





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Biomechanics of stapes motion

- Two components: piston and rocking
- Piston motion dominates
- Rocking motion has higher threshold, lower sensitivity for cochlear activation
  - Thus, not as important

(Heiland KE. Am J Otol,1999 ; Sim, JH. MEMRO, 2012)

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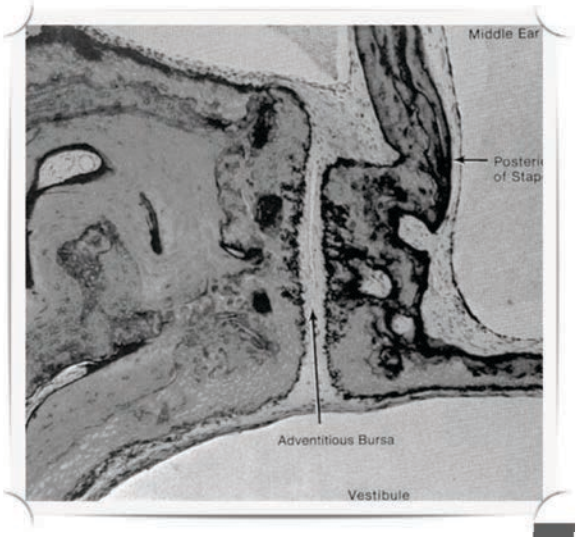
Stapes annular ligament

Syndesmotic (fibrous) joint

Stiffest posteroinferior

Non-linear mechanical properties

Preloading stiffens the annular ligament



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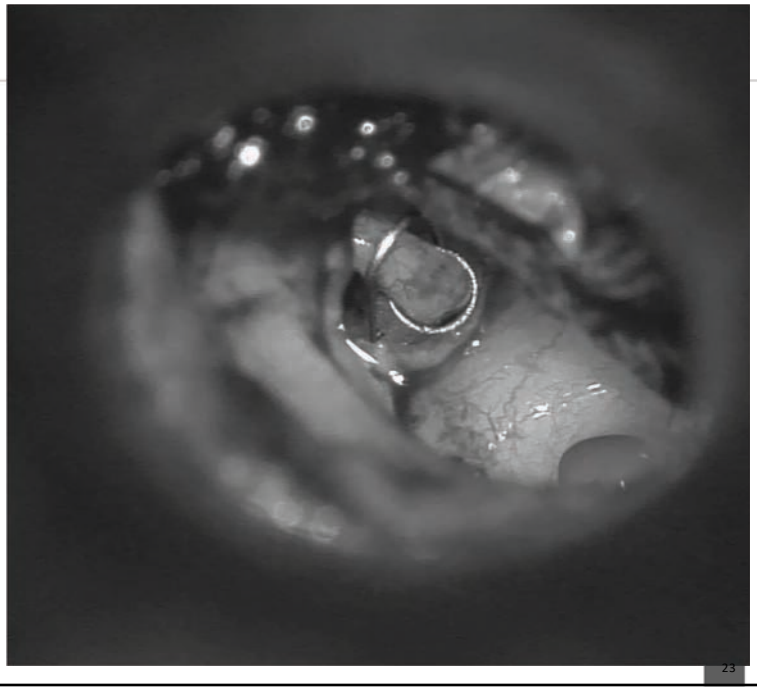
Otosclerosis



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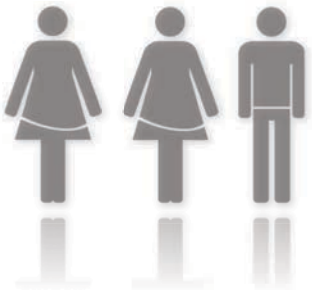
Epidemiology

- Prevalence
  - Clinical otosclerosis 0.5-1%
  - Histological evidence of disease > 10%
  - Bilateral in 60-90%
- Gender predilection
  - 2:1 Female to Male
- Age
  - 20-45 years
- Race
  - Most common among Caucasians (10x AA)



Q5, Q6

24

Endocrine etiology

- Clinical disease 2x more common in women
- Bilateral disease 30% more common in women
- Clinical onset and disease progression more common during pregnancy

- Estrogen receptors found in otosclerotic plaques

25

Diagnosis: Audiometry

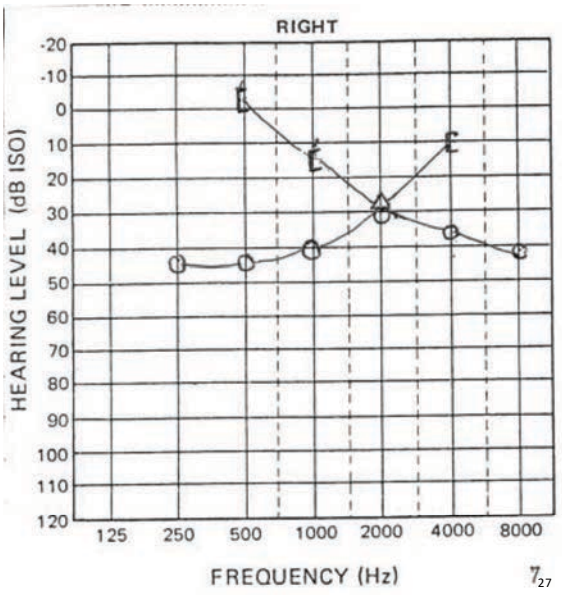
- Best characterizes severity of disease
- Early disease – low frequency CHL
- Late disease – flat CHL through all frequencies
- Labyrinthine or cochlear disease – MHL or SNHL
  - Can often cause drop in WRS

Q7, Q8

26

Diagnosis: Audiometry

- **Carhart's Notch**
  - Depression in BC threshold at 2000 Hz (15 dB)
  - 5 db @ 500, 10 @ 1000, 5 @ 4000 Hz
  - Artifact – primary resonant frequency of ossicular chain for BC around 1700 Hz
  - Disappears after surgery



Q1, Q9

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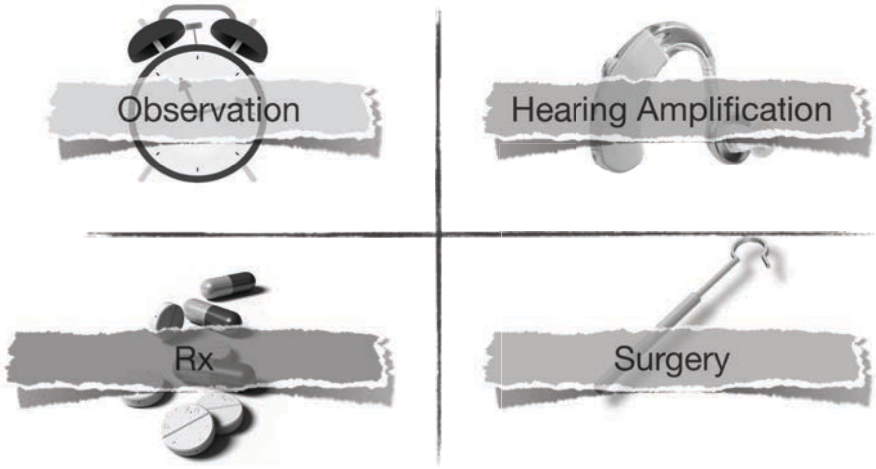
Treatment Options



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CONTINUED

Treatment: Fluoride

- Mechanism: decrease osteoclasts, increase osteoblasts, reduced bone remodeling, inhibits cytotoxic enzymes
- Indications: new or rapidly progressive disease, inner ear manifestations (vertigo, SNHL)
- Histology shows no evidence of effectiveness
- CT studies have confirmed resolution
- Equipoise in literature

Contraindications
Renal disease
Rheumatoid arthritis
Pregnancy

Side-Effects
GI upset
Skeletal Fluorosis

Q10

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CONTINUED

Kessel, 1878

30

CONTINUED

Treatment: stapedectomy

- Minimum ABG 15-20 dB >= 2 consecutive frequencies, ”flip” fork
- Relative contraindications:
  - Unfit for surgery
  - Active OE or OM
  - Perforated TM
  - Only/better hearing ear
  - MD (saccular enlargement)
  - Inner ear malformations (X-linked gusher syndrome)
  - Far advanced otosclerosis

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Surgical outcomes

- ABG closure <= 10 dB in 90-95%
- Partial success 10-20 dB in 3-5%
- 1% unsuccessful
- <1% chance of SNHL

- Delayed hearing loss infrequent over 20 years
  - 1.6% CHL
  - 1.2% SNHL

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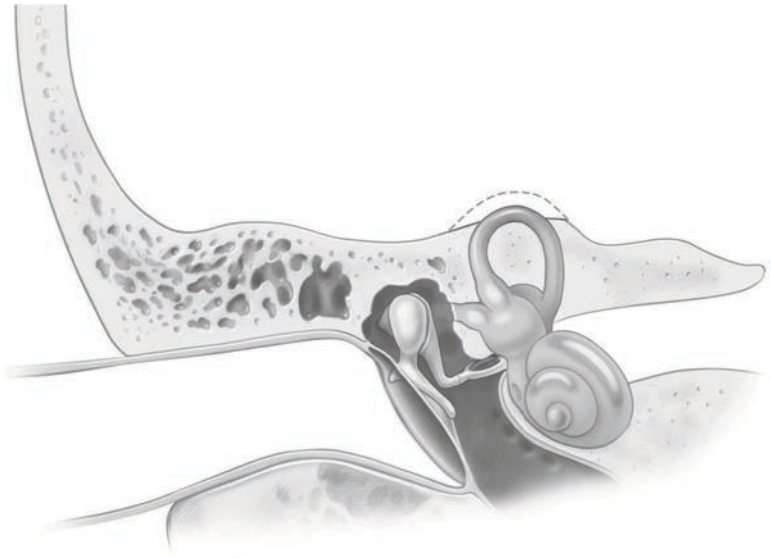
SUPERIOR SEMICIRCULAR CANAL DEHISCENCE



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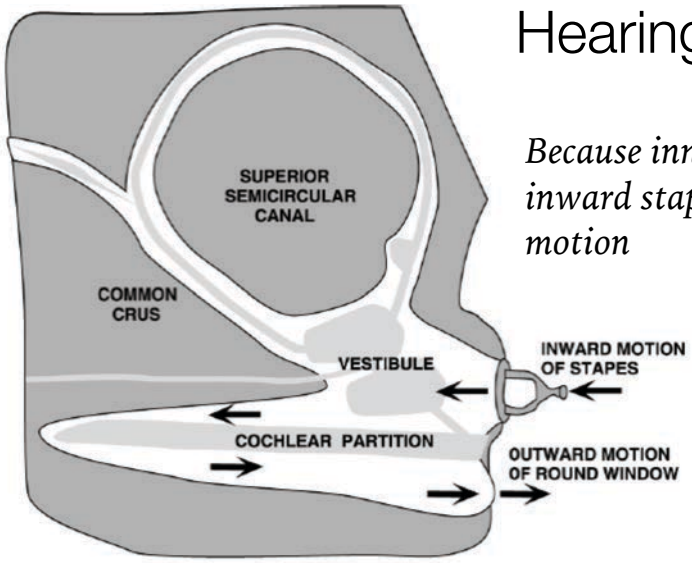
CONTINUED

Hearing in a normal ear



Because inner ear is non-compressible, inward stapes motion = outward stapes motion

Chien et al, Otol Neurotol, 2007

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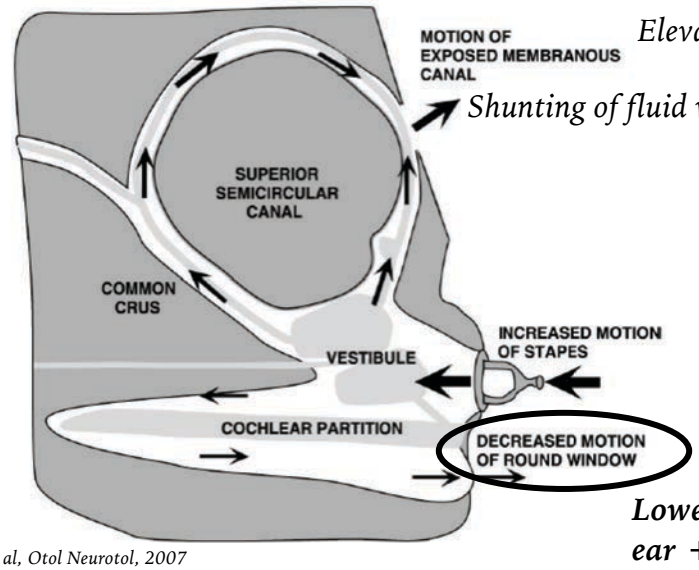
Increased air conduction threshold in SCDS



Elevation of air conduction thresholds

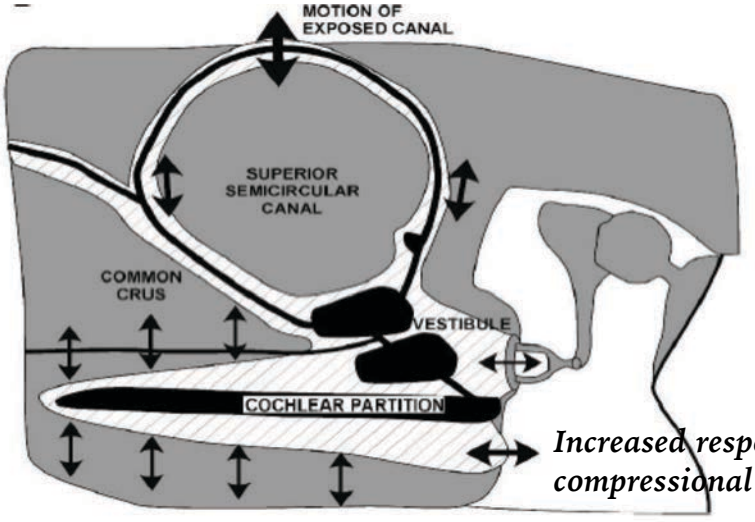
Shunting of fluid volume

Lower input impedance of inner ear + umbo hypermobility

Chien et al, Otol Neurotol, 2007

35

Decreased bone-condition thresholds



Decrease in bone-conducted thresholds

Increased response by cochlea to compressional wave

Suprathreshold fluid motions

Mikulec et al, Otol Neurotol, 2004

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SSCD can mimic otosclerosis

- SSCD can present with audiometric pattern with ABG
- Vertigo, sound/pressure sensitivity can be absent
- Aural symptoms including aural fullness, autophony, pulsatile tinnitus can be absent

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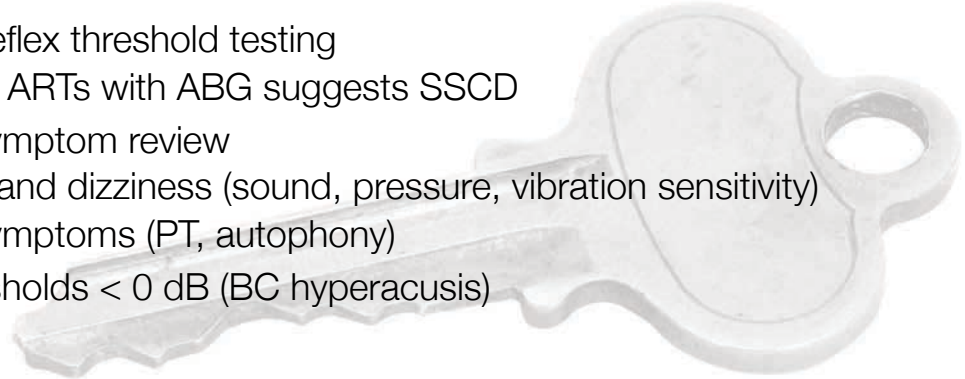
Keys to diagnosing SSCD



- Acoustic reflex threshold testing
  - Present ARTs with ABG suggests SSCD
- Detailed symptom review
  - Vertigo and dizziness (sound, pressure, vibration sensitivity)
  - Aural symptoms (PT, autophony)
- Bone thresholds < 0 dB (BC hyperacusis)
- VEMP
- CT imaging (thin cuts, dedicated views)

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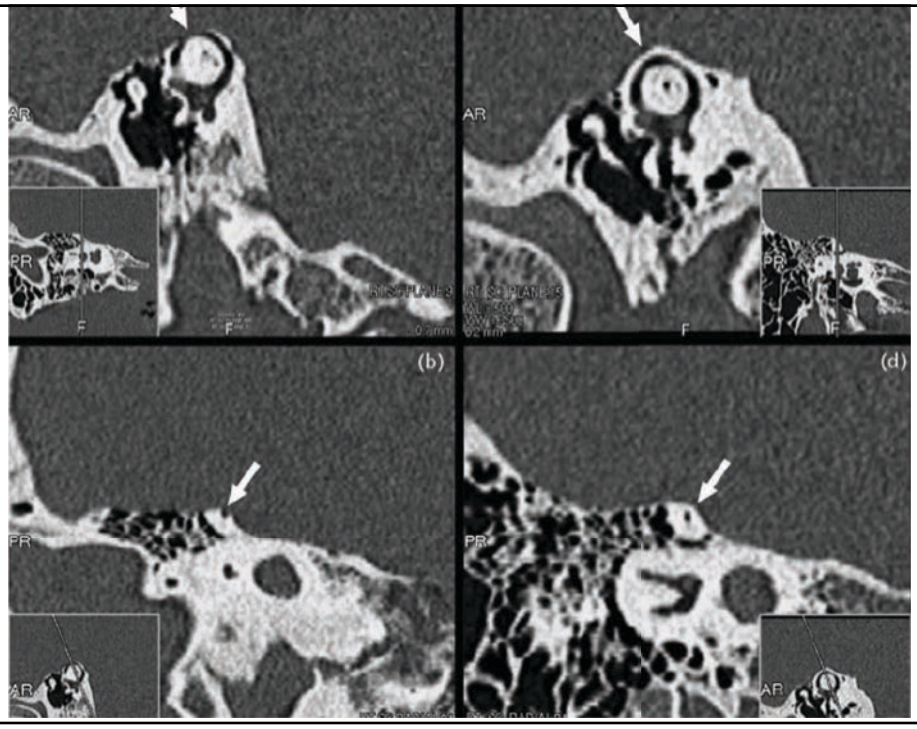
AR
PR
(b)
(d)

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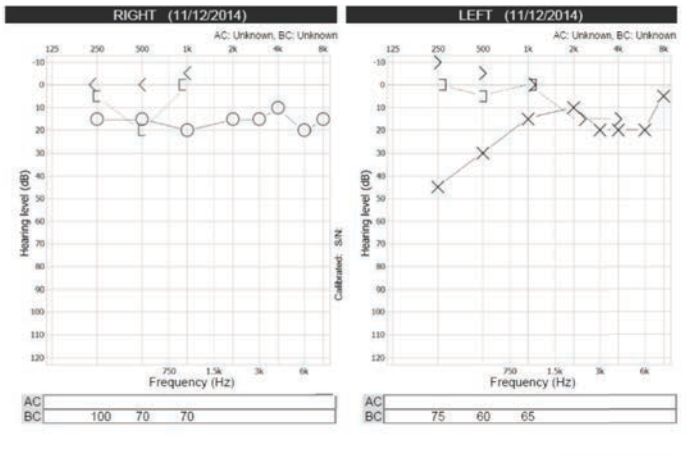
RIGHT (11/12/2014)
AC: Unknown, BC: Unknown
Hearing level (dB)
Frequency (Hz)
AC
BC
100 70 70
LEFT (11/12/2014)
AC: Unknown, BC: Unknown
Hearing level (dB)
Frequency (Hz)
AC
BC
75 60 65
PTA (dB HL) / AI (%)
AC BC AI
Right 16 98
Left 18 6 86
Reliability
Speech SDT SRT WRS / SRS 1 WRS / SRS 2 MCL UCL
dB HL [m] dB HL [m] % dB HL [m] S/N % dB HL [m] S/N dB HL dB HL
Right 20 96 60 [55]
Left 20 100 60 [55]
Legend
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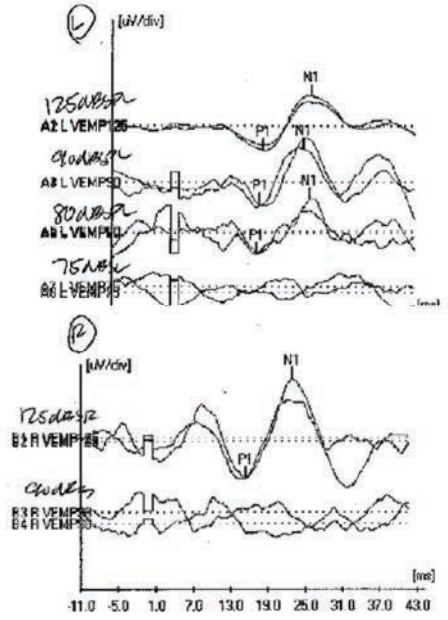
AC
BC
SF
MCL
UCL
NR
PTA
AC: 500, 1k, 2k
BC: 500, 1k, 2k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40

Hearing outcomes following SSCD repair

- Ward (2012): 43 ears, ABG 16 dB — 8 dB; 53% increased PTA (8 dB — 19 dB)
- Limb (2006): 19 primary ears, no change AC or BC, partial closure of ABG in 5/19
- Goddard (2013): 24 ears, no difference in AC thresholds
- Yuen (2006): 10 ears, 1 complete ABG closure, 6 partial, 3 increased

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Table 3. ABG Scores of Patients with Preoperative ABG >10 dB.

		Mean ABG, dB		
Approach	Patients, n (%)	Preoperative	Postoperative	*P* Value
MFCA	6 (43)	15.8	10	.175
TMA	23 (59)	17.2	15.7	.144

Abbreviations: ABG, air-bone gap; MFCA, middle fossa craniotomy approach; TMA, transmastoid approach.

Table 4. Air-Bone Gap by Frequency.[a]

Frequency	MFCA	TMA
500 Hz		
Preoperative	13.6	19.6
Postoperative	7.9	15.1
P value	.079	.131
1000 Hz		
Preoperative	13.6	14.7
Postoperative	8.6	15.8
P value	.121	.636
2000 Hz		
Preoperative	4.29	4.23
Postoperative	2.86	3.97
P value	.414	.841
4000 Hz		
Preoperative	7.5	11.2
Postoperative	11.42	11.5
P value	.151	.861

Abbreviations: MFCA, middle fossa craniotomy approach; TMA, transmastoid approach.
[a]Values are presented as mean dB.

(Schwartz et al, Otolaryngol Head Neck Surg, 2019)

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Photo courtesy of Dr. Larry Lustig

ENLARGED VESTIBULAR AQUEDUCT

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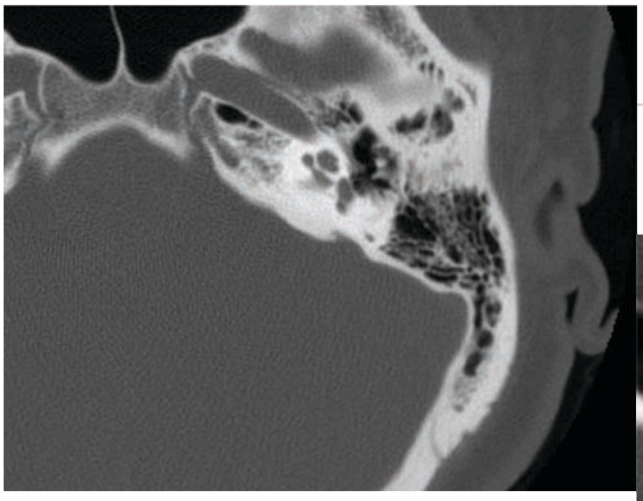

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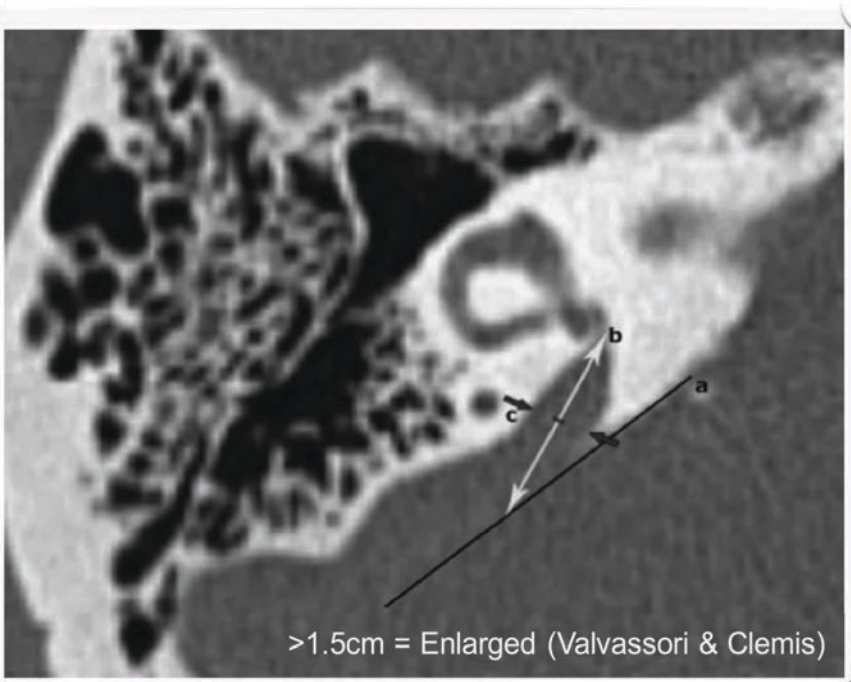

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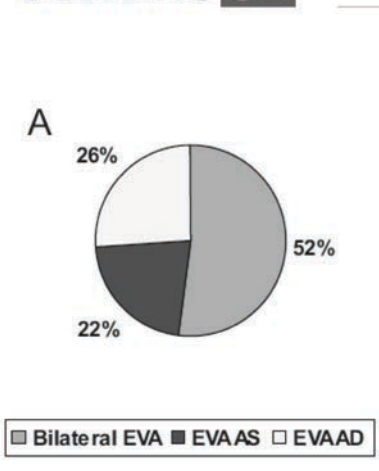

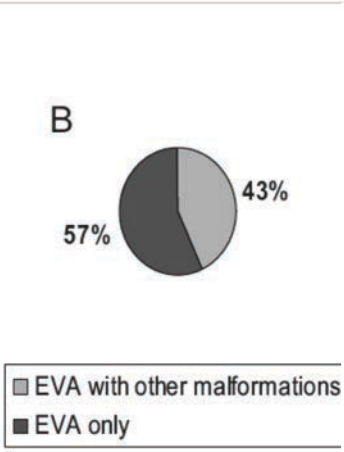

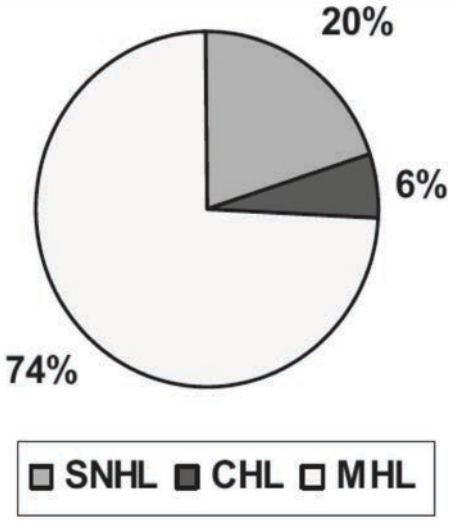

(Zhou G et al, Laryngoscope, 2008)

Largest ABG usually at 250, 500 Hz

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CONTINUED

Etiology of hearing loss in EVA

- Physiology poorly understood

1. Decreased stapes mobility due to increased perilymphatic pressure at the footplate
2. Direct communication between CSF and inner ear increases bone conduction (may explain link between head trauma and HL)
3. Third window phenomenon

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Head trauma and EVAS

- Noordman BJ et al, *Otol Neurotol*, 2014
  - 31 studies, 179 patients, 351 EVAs
  - 34% SHL after head/noise/barotrauma
  - Pre-trauma fluctuating hearing correlates with SHL after trauma (OR 8.6)
  - No risk for SHL after trauma with progressive HL, vestibular symptoms, or Pendred

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AMERICAN JOURNAL OF OTOLARYNGOLOGY–HEAD AND NECK MEDICINE AND SURGERY 34 (2013) 619–625

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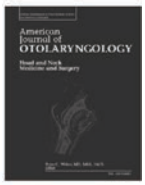

Original contributions

Enlarged vestibular aqueduct syndrome mimicking otosclerosis in adults

*Stephanie S. Wieczorek, BS, Martin E. Anderson Jr., MD, Dave A. Harris, PhD, Anthony A. Mikulec, MD, FACS**

Otolaryngology–Head and Neck Surgery, Saint Louis University School of Medicine, Saint Louis, MO, USA

3 cases previously underwent stapedectomy…

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X-LINKED STAPES GUSHER SYNDROME

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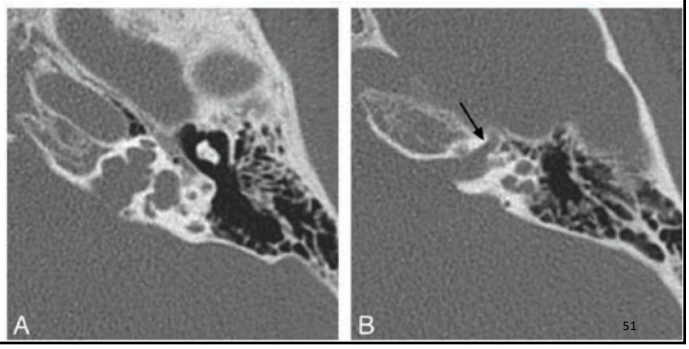
X-linked stapes gusher syndrome

- Pou3F4 mutation - DFN3
- X-chromosome
- Communication between subarachnoid and perilymphatic space
- Pressure under the stapes
- Classic CT findings
- MHL most common



Huang et al, Am J Neuroradiol, 2012

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Stapedectomy and X-linked stapes gusher syndrome

- Poor outcomes after stapedectomy
- Profound hearing loss, failure to close ABG
- Consider HA vs CI
- Be aware of compilations during CI surgery
  - CSF leak
  - Insertion into IAC (Fluoroscopy)

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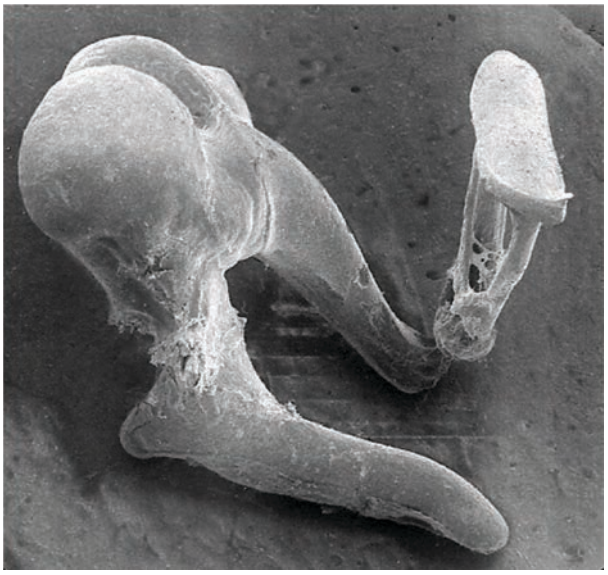

Ossicular abnormalities

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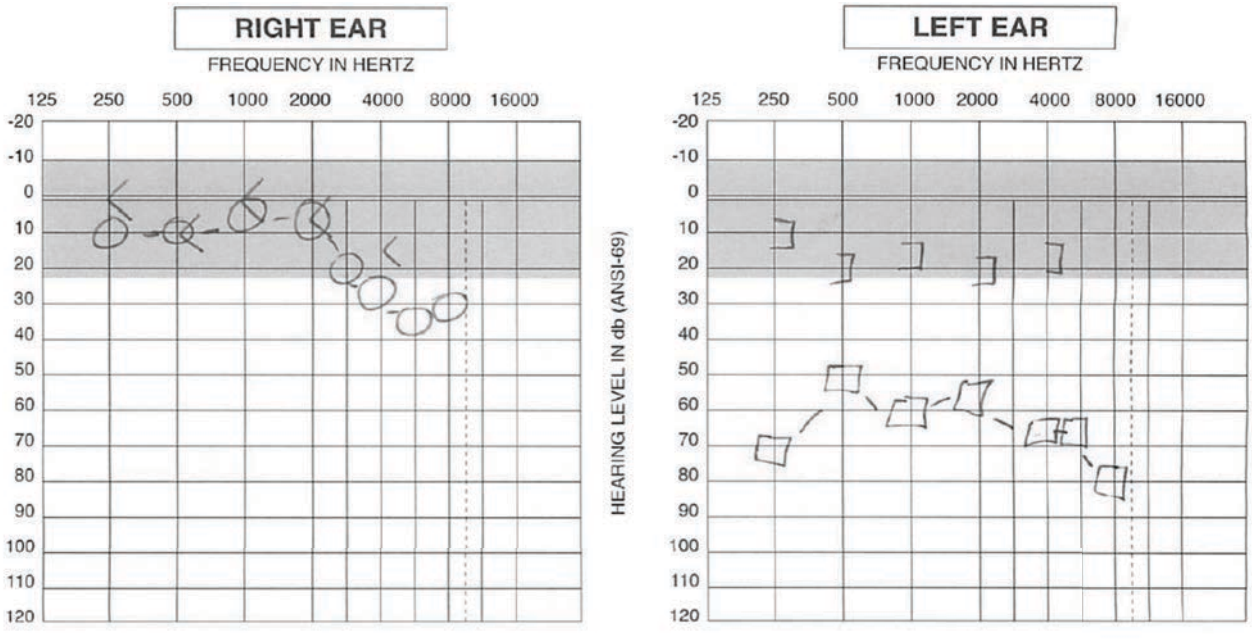

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Ossicular malfunction

- Intact TM + ossicular disjunction = maximum CHL
- ABG < 50 dB - incomplete ossicular disjunction
- DifFERENtial diagnosis
  - Incus necrosis (most common) - watershed
  - Stapes crura fracture/dislocation
  - Congenital malformation
  - Lateral chain fixation
  - Traumatic injury

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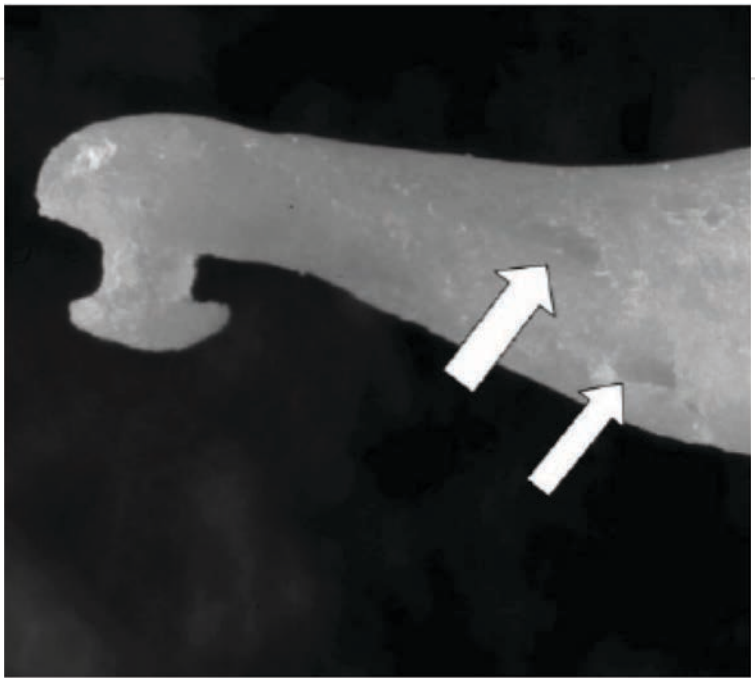

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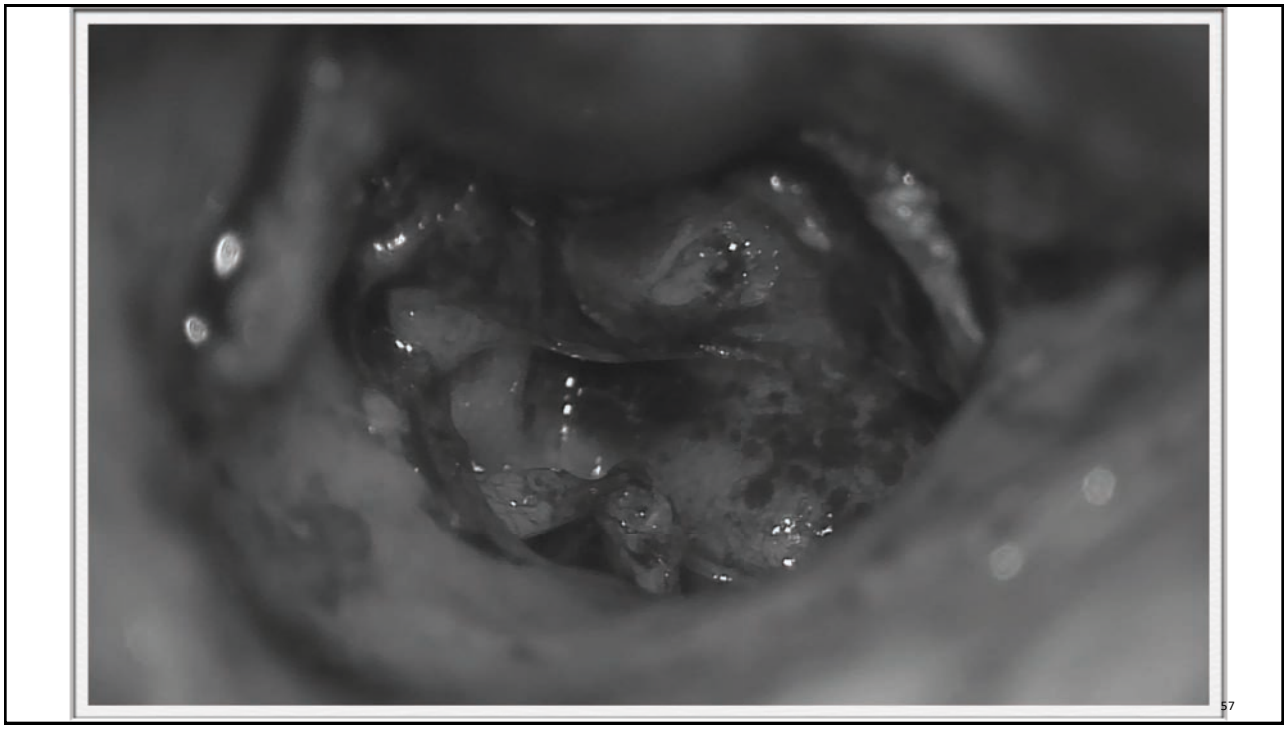

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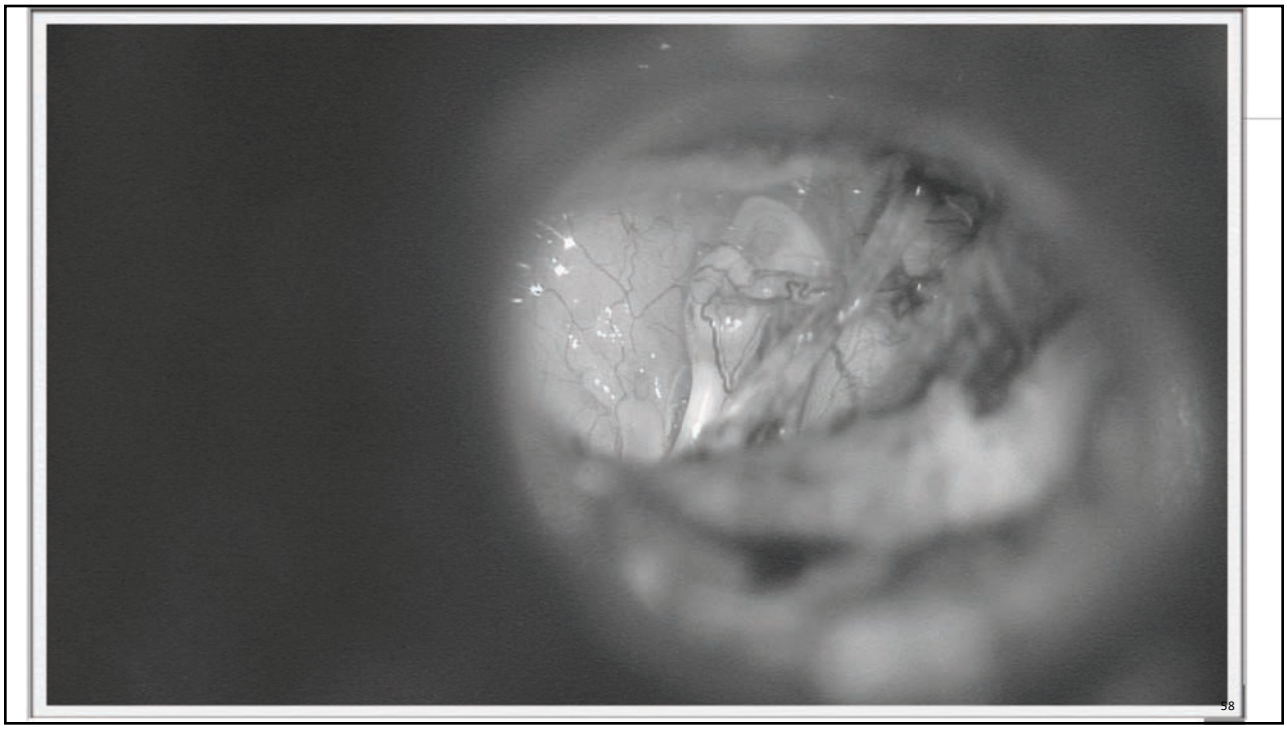

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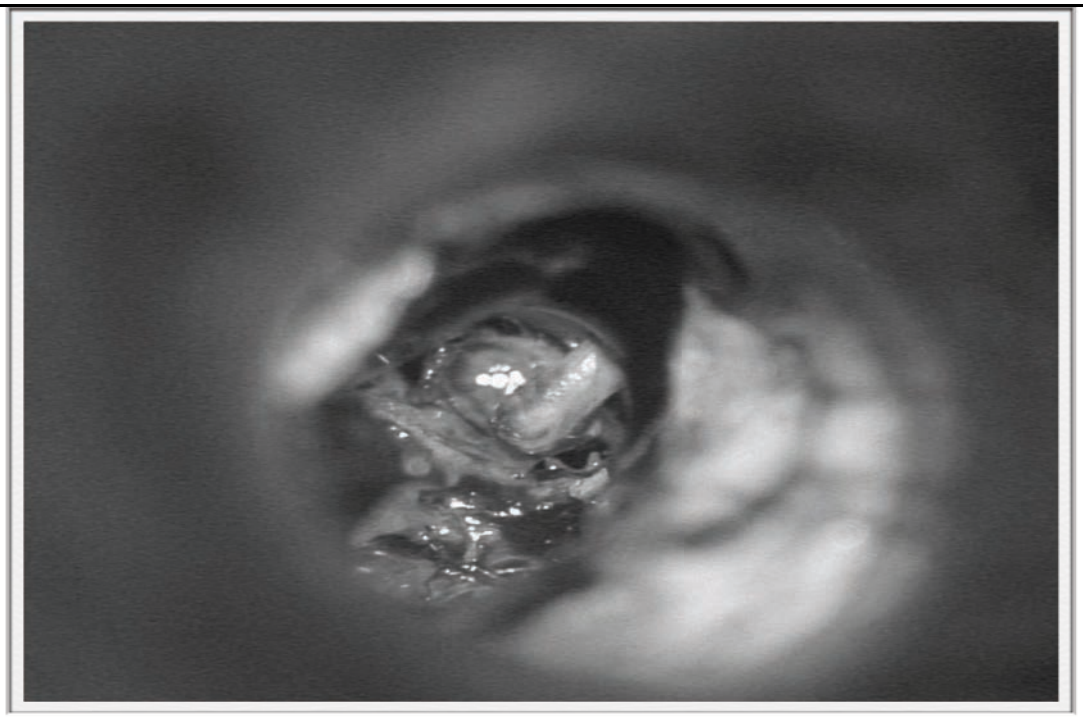

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Work-up

- Objective evaluation
  - Microscopy
  - Tympanogram (Ad)
  - ARTs typically absent
  - CT scan

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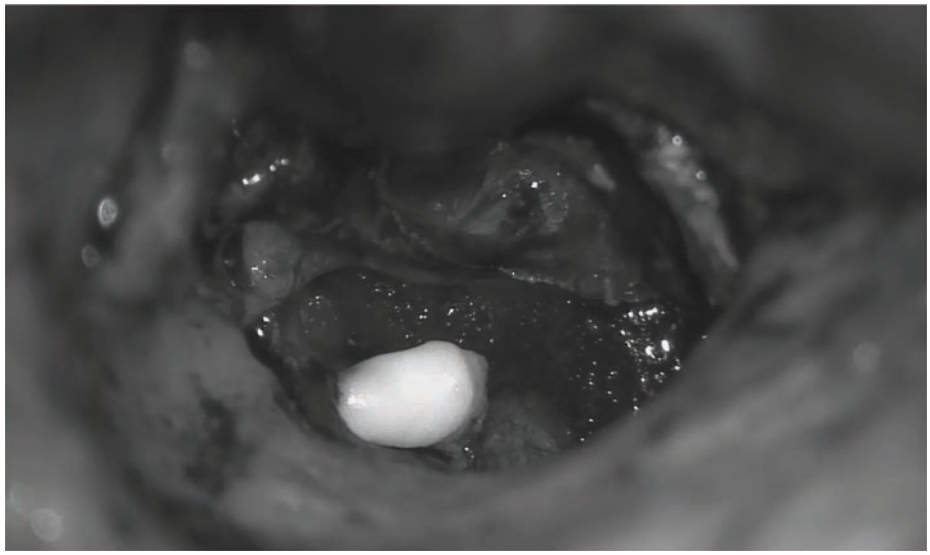

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OSSICULAR RECONSTRUCTION

DANIEL M. ZEITLER, MD

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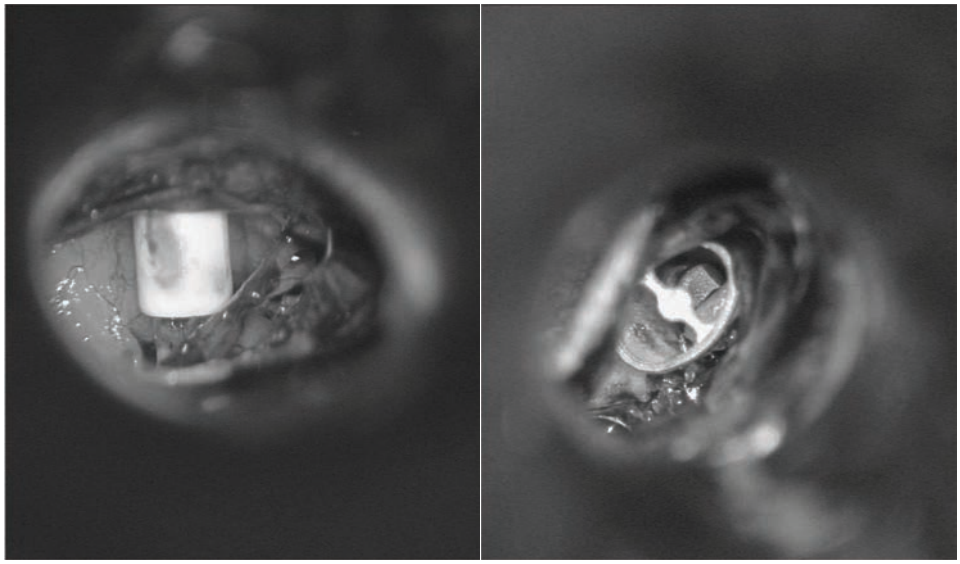

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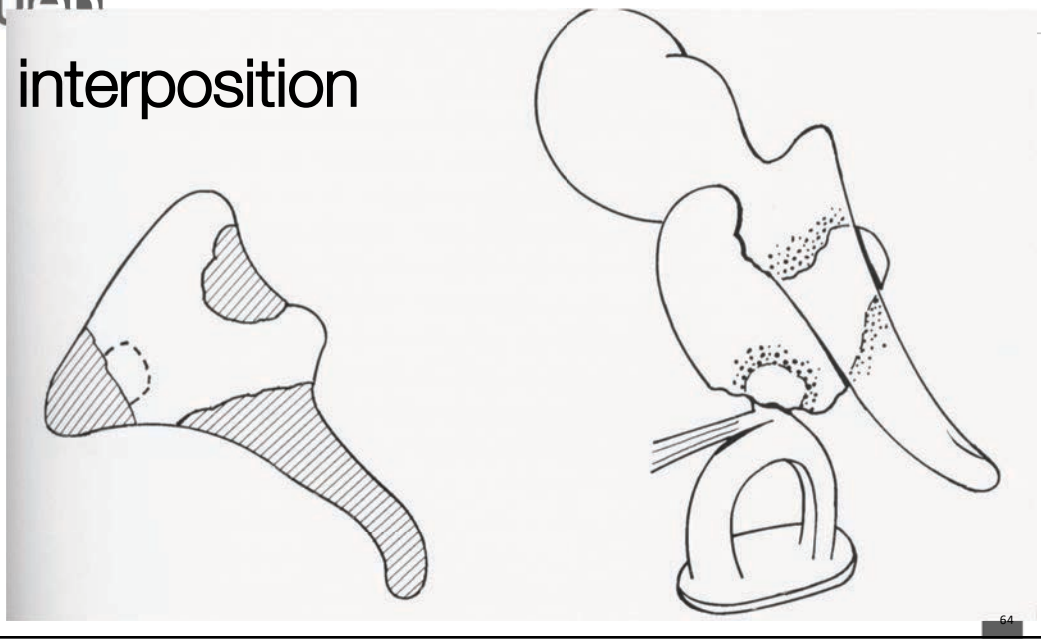
Incus interposition



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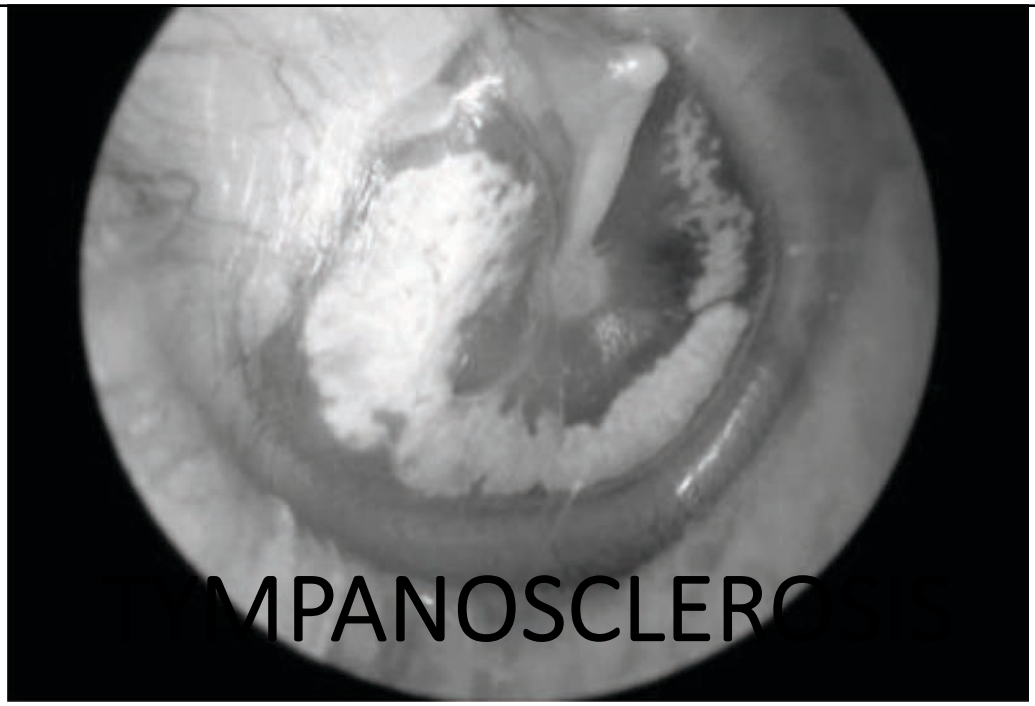

TYMPANOSCLERO

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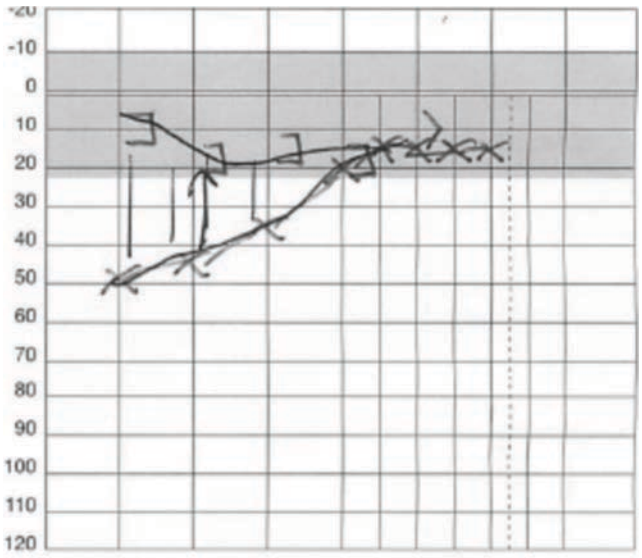

Tympanosclerosis

- Hyaline degeneration + calcification in lamina propria
- Etiology unknown (infectious, inflammatory)
- Plaques can lead to fixation
- Limited vs. extensive disease
- Disease severity ≠ severity of hearing loss

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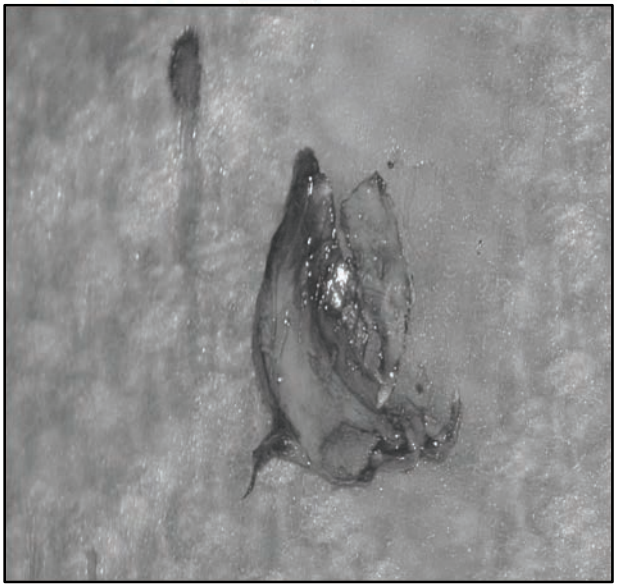

Surgical exploration

Linder and Fisch, 2007

1. Anterior ligament of malleus
2. MI joint
3. Stapes and pyramidal process
4. Round window niche

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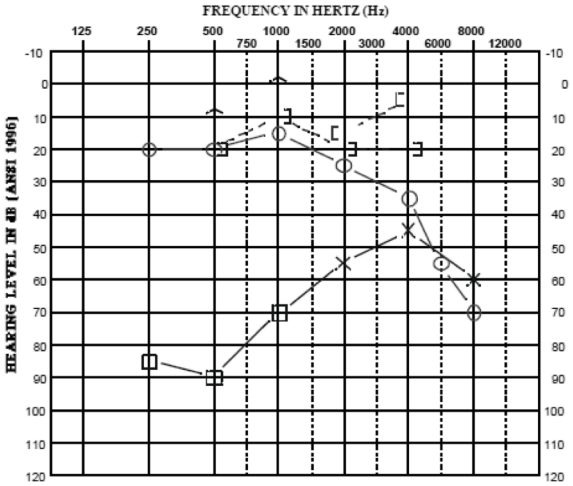
Pediatric considerations



Middle ear malformations
- 1 in 11,000 to 15,000
- Isolated stapes ankylosis 30.6%
- Stapes ankylosis w/ other anomalies 38.2%
- Ossicular malformation with mobile stapes 21.5%
- Severe aplasia/dysplasia of OW or RW 9.7%

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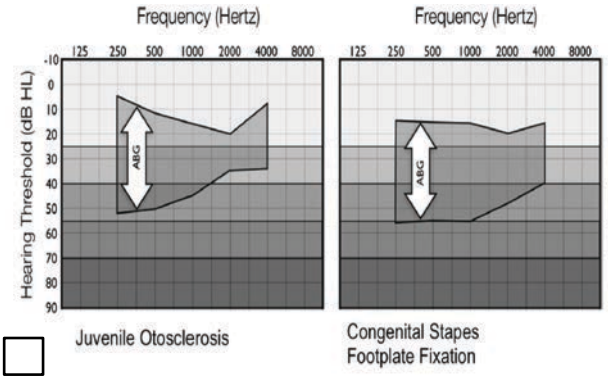
Congenital Footplate Fixation vs. Juvenile Otoscerlosis

- Age:
  - Younger (12 yrs vs 16 yrs)*
- Audiogram
  - Worse (PTA 52 dB vs 42 dB)*
- Other ossicular anomalies
  - Greater (37% vs 0%)
- Outcomes
  - Worse (PTA 32.8 dB vs 22.4 dB)*



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Systematic Review

A Meta-analysis of Surgical Success Rates in Congenital Stapes Fixation and Juvenile Otosclerosis

Burak Asik, MD; Murat Binar, MD; Muhittin Serdar, MD; Bulent Satar, MD

- Overall success rate of ABG closure <10dB was 70%
  - Congenital footplate fixation: 80.2%
  - Juvenile otoscerlosis: 54%

Laryngoscope 2015

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Conclusions

- Otosclerosis is most common cause of CHL, and can be diagnosed without additional testing
- Many etiologies for CHL/MHL other than otosclerosis
- DDx: audiogram, acoustic reflexes, H&P, imaging, VEMP
- Predict etiology based on audiometric pattern
- Knowing diagnosis before surgery improves safety + outcomes
- Ossicular reconstruction techniques vary
- Know when not to operate!

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