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CONDUCTIVE/MIXED HEARING LOSS:
OSTOSCLEROSIS AND OTHER CAUSES

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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.
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.
MY OBJECTIVE WITH CHL/MHL...
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
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

Tympano-ossicular system
Tympanic membrane mechanics

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

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)
Levers of the tympano-ossicular system

- **Catenary lever** – gain realized by TM resting on manubrium while rigidly supported at annulus and acted upon by its 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 +
Hydraulic lever =

34 dB gain (actual 20-25 dB)
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)

Ossicular chain
Malleoincudal joint

Diarthrodial joint: hyaline cartilage with synovial fluid
Hinge-like motion at low frequency
Twisting motion at higher frequencies (“beveled gears”)

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)
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)

Stapes annular ligament

Syndesmotic (fibrous) joint
Stiffest posteroinferior
Non-linear mechanical properties
Preloading stiffens the annular ligament
Otosclerosis

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)
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

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
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

**Treatment Options**

- Observation
- Hearing Amplification
- Rx
- Surgery
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

Kessel, 1878
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

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
SUPERIOR SEMICIRCULAR CANAL DEHISCENCE

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

Chien et al, Otol Neurotol, 2007
Increased air conduction threshold in SCDS

Elevation of air conduction thresholds
Shunting of fluid volume

Lower input impedance of inner ear + umbo hypermobility

Decreased bone-condition thresholds

Decrease in bone-conducted thresholds
Increased response by cochlea to compressional wave

Suprathreshold fluid motions

Chien et al, Otol Neurotol, 2007

Mikulec et al, Otol Neurotol, 2004
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

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)
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

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

<table>
<thead>
<tr>
<th>Approach</th>
<th>Patients, n (%)</th>
<th>Preoperative</th>
<th>Postoperative</th>
<th>P Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>MFCA</td>
<td>6 (43)</td>
<td>15.8</td>
<td>10</td>
<td>.175</td>
</tr>
<tr>
<td>TMA</td>
<td>23 (59)</td>
<td>17.2</td>
<td>15.7</td>
<td>.144</td>
</tr>
</tbody>
</table>

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

### Table 4. Air-Bone Gap by Frequency.¹

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

Abbreviations: MFCA, middle fossa craniotomy approach; TMA, transmastoid approach.

¹Values are presented as mean dB.
ENLARGED VESTIBULAR AQUEDUCT

Photo courtesy of Dr. Larry Lustig
Largest ABG usually at 250, 500 Hz

(Zhou G et al, Laryngoscope, 2008)
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

Head trauma and EVAS

  - 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
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...

X-LINKED STAPES GUSHER SYNDROME
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


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 complications during CI surgery
  - CSF leak
  - Insertion into IAC (Fluoroscopy)
Ossicular abnormalities
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
Work-up

- Objective evaluation
  - Microscopy
  - Tympanogram (Ad)
  - ARTs typically absent
  - CT scan
Incus interposition
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
Surgical exploration

Linder and Fisch, 2007

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

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%
Congenital Footplate Fixation vs. Juvenile Otosclerosis

- **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; Bülent Sntar, MD

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

Laryngoscope 2015
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!