Hearing Sensitivity & Connective Tissue Disorders

Jeffrey Marler, PhD
The OSU Eye and Ear Institute
Cognitive Auditory Research Lab
10-22-11 Cutis Laxa Clinic

• Risk factors of mild hearing loss (HL)
• Basics of ear structure and function
  – Middle ear
  – Inner ear
• How an audiologist measures integrity of function (is it working?)
  – Middle ear – tympanometry (ME pressure)
  – Inner ear – otoacoustic emissions (outer hair cells)
• These are important to the audiologist, and indirectly to you...but what you want to know is “IS MY HEARING NORMAL???”
• The audiogram
Demographics

- There are ~1.7 million children in the US aged 6-16 with MHL (unilateral or bilateral).
- If you measure children with “slight” hearing loss (PTA > 16 dB HL), there are ~1.1 million children in the US aged 6-16 with slight HL.
- There are ~2.8 million children in academic settings not identified as possibly at-risk.
Risk Factors for Mild Hearing Loss (MHL)

• 37% of children fail at least one grade
• 8% not performing at grade level
• Comprehensive Test of Basic Skills
  – In 3rd grade, children show lower scores for reading, language mechanics, word analysis, spelling, and science than normally hearing (NH) peers
• Ross et. al. (2005)
  – Children 6-16 years of age with MHL twice as likely to score 2 SD’s below the norm on standardized math and reading tests when compared to NH peers
  – Children with MHL at higher risk for speech and/or language difficulties than NH children

Bess, Dodd-Murphy, & Parker, 1998
<table>
<thead>
<tr>
<th>Gross division</th>
<th>Outer ear</th>
<th>Middle ear</th>
<th>Inner ear</th>
<th>Central auditory nervous system</th>
</tr>
</thead>
<tbody>
<tr>
<td>Anatomy</td>
<td>pinna</td>
<td>malleus</td>
<td>vestibule</td>
<td>facial n.</td>
</tr>
<tr>
<td></td>
<td>concha</td>
<td>incus</td>
<td>vestibular n.</td>
<td>cochlear n.</td>
</tr>
<tr>
<td></td>
<td>external auditory canal</td>
<td></td>
<td>semicircular canals</td>
<td>internal auditory canal</td>
</tr>
<tr>
<td></td>
<td>external auditory meatus</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Mode of operation</td>
<td>Air vibration</td>
<td>Mechanical vibration</td>
<td>Mechanical, Hydrodynamic, Electrochemical</td>
<td>Electrochemical</td>
</tr>
<tr>
<td>Function</td>
<td>Protection, Amplification, Localization</td>
<td>Impedance matching, Selective oval window stimulation, Pressure equalization</td>
<td>Filtering distribution, Transduction</td>
<td>Information processing</td>
</tr>
</tbody>
</table>
ME – EI Interactions

FEM model of the middle ear.

1. Tympanic membrane (pars tensa)
2. Tympanic membrane (pars flaccida)
3. Malleus; 4. Incus; 5. Stapes
6. anterior malleal ligament
7. Posterior incudal ligament
8. **Tensor tympani tendon**
9. **Stapedial tendon**
10. Annular ligament

*Sone (1998) reported elastin in the round and oval windows*
ELN in ME structures

- Tympanic membrane
  Ruah et al. (1992); Yan et al. (1988)

- Stapedius tendon
  Franz et al. (1993)

- Eustachian tube
  Matsune et al. (1992); Sando et al. (1994)
<table>
<thead>
<tr>
<th>Gross division</th>
<th>Outer ear</th>
<th>Inner ear</th>
<th>Central auditory nervous system</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Anatomy</strong></td>
<td>pinna</td>
<td>malleus</td>
<td>vestibule</td>
</tr>
<tr>
<td></td>
<td>concha</td>
<td>incus</td>
<td>vestibular n.</td>
</tr>
<tr>
<td></td>
<td>external auditory canal</td>
<td>semicircular canals</td>
<td>facial n.</td>
</tr>
<tr>
<td></td>
<td>external auditory meatus</td>
<td>cochlea</td>
<td>cochlear n.</td>
</tr>
<tr>
<td></td>
<td>ear drum</td>
<td>stapes</td>
<td>round window</td>
</tr>
<tr>
<td></td>
<td></td>
<td>eustachian tube</td>
<td>internal auditory canal</td>
</tr>
<tr>
<td><strong>Mode of operation</strong></td>
<td>Air vibration</td>
<td>Mechanical vibration</td>
<td>Mechanical, Hydrodynamic, Electrochemical</td>
</tr>
<tr>
<td>Function</td>
<td>Protection, Amplification, Localization</td>
<td>Impedance matching, Selective oval window stimulation, Pressure equalization</td>
<td>Filtering distribution, Transduction</td>
</tr>
</tbody>
</table>
1) Main Structures of the Inner Ear

- Cochlea
- Basilar membrane
- Hair cells
**ELN in Cochlea**

- Round Window
  - Sone (1998)
- Basilar membrane
  - Mikuni et al., (1994)
- Hair cell stereocillia
  - Osborn & Comis (1990)
- Not in hair cell stereocillia
  - Katori et al., (1996)
- Not in adult mouse cochlea

McLean et al., 2009
10-22-11 Cutis Laxa Clinic

• Risk factors of mild hearing loss (HL)
• Basics of ear structure and function
  – Middle ear
  – Inner ear
• How an audiologist measures integrity of function (is it working?)
  – Middle ear – tympanometry (ME pressure)
  – Inner ear – otoacoustic emissions (outer hair cells)
• These are important to the audiologist, and indirectly to you...but what you want to know is “IS MY HEARING NORMAL???”
• The audiogram
Measures of Middle-Ear Function

226-Hz Probe Tympanometry (typical)

Multifrequency tympanometry (typical)

Hz range 750 - 1200

daPa = +/- 100
mmho = 0.3 to 1.2
Point of reference for MFT & WB

- The frequency sweeps between 250 and 2000Hz in 50-Hz steps
- The resonant frequency is the frequency at which $\Delta B$ is equal to 0.
- Pathologies that increase the stiffness (mass controlled) increase the resonant frequency (high mean Hz)
- Pathologies that reduce the stiffness (stiffness controlled) decrease the resonant frequency (low mean Hz)
Point of reference for MFT & WB

Wideband ambient-pressure acoustic transfer functions (ATFs)

- very sensitive to presence of subclinical otitis media, particularly in regions above 1000 Hz

- Keefe & Simmons (2003) reported that both ambient and tympanometric WB ATFs were more accurate predictors or conductive hearing loss (0.90 and 0.95 are under the ROC curve respectively) in older children and adults

- WB ATFs would allow frequency-specific diagnostic information not available with standard 226-Hz probe tympanometry.

- WB ATFs ARE CURRENTLY NOT APPROVED BY THE FDA AND ARE ONLY AVAILABLE FOR RESEARCH

Normal middle-ear resonance (800 to 1200 Hz)
Otoacoustic Emissions

• Healthy cochlea more sensitive to low- to mid-level sounds (nonlinear/compressive)
• Outer hair cells (OHCs) support discriminating many different frequencies at once
• OHCs contribute to extreme sensitivity to very low-level sounds
• OHCs facilitate hearing in noisy environments
Risk factors of mild hearing loss (HL)
Basics of ear structure and function
  - Middle ear
  - Inner ear
How an audiologist measures integrity of function (is it working?)
  - Middle ear – tympanometry (ME pressure)
  - Inner ear – otoacoustic emissions (outer hair cells)
These are important to the audiologist, and indirectly to you...but what you want to know is: “IS MY HEARING NORMAL???”
The audiogram
**What is an audiogram?**

An audiogram is a graph on which the audiologist plots the results of your child's responses to different pitches (frequencies) and the loudness (decibel) level at which your child could hear that pitch.
• Any responses above the red line (≤15 dB HL) are within normal limits
## 2) Basics of Hearing – Testing

<table>
<thead>
<tr>
<th>Sound</th>
<th>Intensity (dB HL)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ticking of a Watch</td>
<td>20</td>
</tr>
<tr>
<td>Whisper</td>
<td>30</td>
</tr>
<tr>
<td>Normal Speech</td>
<td>50-60</td>
</tr>
<tr>
<td>Car Traffic</td>
<td>70</td>
</tr>
<tr>
<td>Alarm Clock - Blender</td>
<td>80</td>
</tr>
<tr>
<td>Lawn Mower - Blow Dryer</td>
<td>95</td>
</tr>
<tr>
<td>Chain Saw</td>
<td>110</td>
</tr>
<tr>
<td>Jackhammer</td>
<td>120</td>
</tr>
<tr>
<td>Jet Engine</td>
<td>130</td>
</tr>
</tbody>
</table>
## CLASSIFICATIONS OF HEARING LOSS

<table>
<thead>
<tr>
<th>Type</th>
<th>Progression</th>
<th>Frequency</th>
<th>Severity</th>
<th>Onset</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sensorineural</td>
<td>Progressive</td>
<td>Low ( ≤ 500 Hz)</td>
<td>Mild</td>
<td>Congenital</td>
</tr>
<tr>
<td>Conductive</td>
<td>Nonprogressive</td>
<td>Middle ( 501-2000 Hz)</td>
<td>Moderate</td>
<td>Childhood</td>
</tr>
<tr>
<td>Mixed</td>
<td>Fluctuating</td>
<td>High ( &gt; 2000 Hz)</td>
<td>Moderately severe</td>
<td>Adulthood</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Severe</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Profound</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>(21- 40 dB HL)</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>(41- 60 dB HL)</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>(61- 80 dB HL)</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>(81-100 dB HL)</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>( &gt; 100 dB HL)</td>
<td></td>
</tr>
</tbody>
</table>
Hearing evaluations of individuals with connective tissue disorders

• Disruption of the elastic fiber system negatively impacts middle-ear function in ADCL, and WS
• There appears to be a higher incidence of hearing loss in ARCL than would be expected in the general population
• Multifrequency tympanometry should be included as part of any audiometric battery when evaluating individuals with connective tissue disorders