Abnormal Audiograms in Ear Pathology

Presented by Lori Klingenberg, Au.D. CCC-A
Ear, Nose, Throat and Plastic Surgery Associates
Winter Park, FL
ENTorlando.com

Disclaimer

None to report

Objectives of this session

The attendee will
- Improve interpretation of audiograms
- Distinguish ear pathologies with their accompanying audiograms
- Narrow the differential diagnosis of hearing loss
- Recognize the third window phenomenon
- Identify audiometric findings that suggest non-organic hearing loss
Audiogram review

• Pure tone testing
  – Thresholds are obtained by air and bone conduction. Threshold is determined by at least 2 responses on ascending presentation of tones.
  – Air conduction ONLY tells us the DEGREE of hearing loss
  – Bone conduction tells us what TYPE of hearing loss is present (sensorineural, mixed, conductive)
  – Describe the audiogram by configuration from least amount of hearing loss to the most (ex: mild to severe)
  – Pure tone average is 500 Hz, 1000 Hz, and 2000 Hz added and divided by 3.

Degree of hearing loss

Degree of hearing loss:
• 0-15 dB WNL
• 16-25 dB Slight
• 26-40 dB Mild
• 41-55 dB Moderate
• 56-70 dB Moderately-severe
• 71-90 dB Severe
• 90+ dB Profound

![Diagram of audiogram configuration](image)
Speech Testing

Speech Recognition Threshold
- The lowest level at which at least 50% of spondees are identified correctly.
- Purpose = determine the lowest level a person can hear speech.
- Crosscheck accuracy of pure tone audiometry
- SRT should be within 7 dB of PTA.

Word Recognition (WRs)
- Percentage of single words or words in sentences patient hears correctly.
- Purpose = determining how well the person understands speech in each ear. Used diagnostically and for hearing aid purposes.
- Lists are presented at suprathreshold

Question 1: Masking...Why do we need it?

A. It keeps the worse cochlea from responding with bone conduction.
B. It keeps the worse ear from crossover response in air conduction.
C. It prevents crossover response in air conduction and bone conduction.
D. It focuses the patient’s attention away from distracting external sounds.

Masking...Why do we need it?
• Prevention of cross hearing in both air conduction and bone conduction.
• Air conduction = If there is a large difference between ears, it is possible the intensity of sound in the test ear could cross over and be heard in the non-test ear.
• Bone conduction = Does not have any inter-aural attenuation (0 dB HL)...so without masking the better cochlea is responding.
Tactile responses

- Important to understand as this may alter treatment method and possibly cause unnecessary surgery
- Patient feels the vibration of bone conduction vs hearing it.
- Most prevalent at high intensity/low frequency bone conduction (ex: 500 Hz)
- Why is this an issue?
  - Could cause elevated (better) bone conduction threshold and false referral for surgery to close the “air/bone gap.”
  - Also ask mature patient if they “felt or heard the sound?”

Variability in audiograms

- Testing outcome can vary based upon:
  - Proper calibration of equipment (done annually)
  - Test environment - proper sound booth, minimal extraneous noise
  - Examiner experience
  - Patient (performance on test)
    - False negative or false positive responses

Audiometric abbreviations:

- CNT - Could not test
- DNT - Did not test
- HA - Hearing aid
- HAE - Hearing aid evaluation
- NR - No response
- SNHL - Sensorineural hearing loss
- WNL - Within normal limits
- AU - Both sides (ears)
- AS - Left
- AD - Right
- VT - Vibrotactile response
- RTC - Return to clinic
- BC - Bone conduction
- AC - Air conduction
- PTA - Pure-tone average
- UCL - Uncomfortable loudness level
- MCL - Most comfortable loudness level
- HFA - High frequency average
- HL - Hearing level
- SPL - Sound pressure level
- SRT - Speech reception threshold
- SAT - Speech awareness threshold (Martin and Clark, 2015)
Type of hearing loss

<table>
<thead>
<tr>
<th>AC Loss?</th>
<th>BC Loss?</th>
<th>Significant Air/bone gap?</th>
</tr>
</thead>
<tbody>
<tr>
<td>• CHL=</td>
<td>YES</td>
<td>YES</td>
</tr>
<tr>
<td>Conductive</td>
<td></td>
<td></td>
</tr>
<tr>
<td>• SNHL=</td>
<td>YES</td>
<td>NO</td>
</tr>
<tr>
<td>Sensorineural</td>
<td></td>
<td></td>
</tr>
<tr>
<td>• Mixed=</td>
<td>YES</td>
<td>YES</td>
</tr>
</tbody>
</table>

Example Audiograms

Ear Pathology and Audiograms
Let’s start with more questions........

Question 2: Based on this audiogram and knowing the patient had normal tympanometry, what possible disorder could cause this hearing loss, left ear?

A. Meniere’s disease
B. Otitis media with effusion
C. TM perforation
D. Otosclerosis

Question 3: Based on this audiogram and knowing the patient had normal tympanometry, **WHY** do you think the disorder causing this hearing loss, left ear is....?

A. Meniere’s – because this is low-frequency hearing loss.
B. OME – because this is conductive hearing loss.
C. TM perf – because the SRT is worse on the left
D. Otosclerosis – because this is conductive hearing loss with a normal tympanogram
Question 4: What type of hearing loss is present, left ear?

A. Conductive  
B. Sensorineural  
C. Mixed  
D. Non-organic

Question 5: What would be a probable diagnosis?

A. Meniere's disease  
B. Large vestibular aqueduct syndrome  
C. Cochlear otosclerosis  
D. Sudden idiopathic SNHL

Idiopathic SNHL, AS

What to look for:
- Decrease in pure tone thresholds of more than 30 dB at 3 consecutive frequencies  
- Asymmetrical SNHL with poor word recognition on affected ear  
- Normal middle ear function  
- Pt may or may not present with vertigo  
- Often present with tinnitus and aural fullness  
- Hearing loss recovery greater for LF thresholds (vs high-frequency)
Idiopathic SNHL

• In many cases, the cause of sudden SNHL cannot be identified. Known causes of sudden SNHL to rule out:
  Causes?
  – Viral or vascular are most suspected cause (Hall 2014)
  – Perilymph fistula (hearing loss with vestibular symptoms)
  – Autoimmune disorders
  – Meniere’s disease (Vestibular symptoms present)
  – Tumors
  – Closed head trauma
  – Rupture of basilar membrane
  – Neurological disorders

Noise induced hearing loss “NIHL”

• 1 of 2 most common causes of acquired SNHL (the other is presbycusis)
  – Caused by impulsive or long term noise exposure
  – Permissible noise levels=85 dB SPL/8 hours
• Most preventable cause of acquired hearing loss
• Landmark “noise notch” generally around 3-6 kHz
• Recovery at 8 kHz

NIHL

• Temporary threshold shift
  – Decrease in hearing persisting 16-48 hours after exposure (Hall, 2014)
  – Accompanied by tinnitus and distortion of speech
• Permanent threshold shift
  – Repeated exposure causes irreversible loss-characteristic notch around 3-4 kHz. Pattern can widen with continued exposure (Kramer, 2014)
• Differential Diagnosis?
  – Sudden idiopathic SNHL (case hx and audiometric pattern will differ)
  – Presbycusis (WILL not have recovery at 8 kHz as in NIHL)
• Recommendations?
  – Proper ear protection
  – Antioxidant therapy?
**Question 6:** What disorder(s) would you suspect with this audiogram?

A. Presbycusis, AS and Vestibular schwannoma, AD
B. Meniere’s disease, AD and presbycusis AS
C. Bilateral Meniere’s disease
D. Bilateral vestibular schwannomas

---

**Vestibular Schwannoma**

- According to Hall (2014), autopsy results 8 per 1000 persons.
- More common in age 30-60 years, females
- Early stages, high frequency hearing loss as outer fibers of auditory are affected due to compression
- Tinnitus secondary to tumor pressure
- Poor word recognition on affected ear (asymmetrical)
7.5 mm left vestibular schwannoma

- Smaller tumor than previous slide at 1.4 cm
- HF asymmetry present, but word recognition remains normal on affected ear.
- Size and location of tumor causes variance in symptoms.

Audiometric findings for vestibular schwannoma-red flags

- Asymmetrical, HF SNHL (progressive with tumor growth)
- Unilateral tinnitus
- Pt may not present with vertigo, slow tumor growth results in imbalance due to central compensation
- Facial “tingling” or weakness
- Asymmetrical word recognition with rollover present
- Abnormal acoustic reflex day and acoustic reflexes
- OAEs may be normal if hearing loss is truly neural
- Normal middle ear function
- ABR testing=increased absolute and interpeak (I-III) wave latencies. Wave V interaural difference greater than 0.3 msec.
- Vestibular testing may show unilateral weakness on affected side

Differential diagnosis of Vestibular Schwannoma’s

- Neurofibromatosis (NF2)
  - Often bilateral symptoms-also mistaken for Meniere’s disease
- Meningioma involving auditory nerve (Hall, 2014)
- Vascular loop syndrome (Hall 2014)
- Multiple sclerosis (Martin and Clark, 2015)
- Hx of noise induced hearing loss (asymmetrical due to recreational or work exposure)
**Question 7:** Why is this audiogram consistent with possible superior canal dehiscence?

A. Mixed hearing loss at 4000 Hz  
B. Enhanced thresholds for LF bone conduction  
C. High frequency conductive hearing loss  
D. Decreased bone conduction at 2000 Hz.

---

**3rd Window Phenomenon?**

- Most common and most researched disorder is Superior Canal Dehiscence syndrome (SCDS)  
- Abnormal thinning of the bone that covers the superior semicircular canal  
- Creates unnatural “third window”  
- Most often unilateral but can be bilateral (Hall, 2014)  
- Other disorders that can cause “third windows” include:  
  - Large Vestibular Aqueduct syndrome  
  - Carotid-cochlear dehiscence  
  - X-linked deafness with stapes gusher  
  - Paget’s disease (Merchant & Rosowski, 2008)

---

**SCDS**

- Window on the scala vestibule side of cochlea due to thinning of bone on superior semicircular canal  
- Results in worsening of air conduction thresholds and improvement of bone conduction thresholds.  
- Why?  
- Air conduction sound dissipated from cochlear partition. Decreased sound pressure=reduced hearing sensitivity (Merchant & Rosowski, 2008)  
- Bone conduction: Enhanced bone conduction due to vibration of fluids in cases of SCDS. (Hall, 2014)
Symptoms of SCDS

- Hearing loss
- Vertigo intensified with loud sounds or pressure changes
- Imbalance
- Tulio phenomenon-Nystagmus with loud sounds (Hall, 2014)
- Hennebert sign-Nystagmus with pressure changes (Hall, 2014)
- Internal sounds magnified such as footsteps, heartbeat, own voice (due to elevated bone conduction)
  - Pt in clinic-bothered by eyelids fluttering

Audiometric characteristics of “third window” SCDS

- Normal middle ear function with CHL
- Suprathreshold bone conduction (better than 0 dB HL in low frequencies)
- Acoustic reflexes present despite air/bone gap
- VEMPS present and often enhanced with CHL-particular to SCDS
- OAEs can be present as “CHL” is not secondary to middle ear pathology

But wait...how to differentiate SCDS from Otosclerosis?

Cannot always differentiate based on the audiogram alone
Question 8: So how do we differentiate SCDS from Otosclerosis?
A. SCDS patients will have normal bone conduction.
B. Otosclerosis patients will also have vestibular complaints.
C. SCDS patients will have poor compliance on tympanograms.
D. Otosclerosis patients will have acoustic reflexes absent in affected ear.

Differential diagnosis of Otosclerosis vs SCDS

**Otosclerosis**
- CHL with normal ME and TM
- Reduced compliance on tympanometry
- Normal but not enhanced bone conduction thresholds
- Acoustic reflexes absent in affected ear
- Usually not associated with complaints of vertigo
- VEMPS absent due to true CHL

**SCDS**
- CHL with normal ME and TM
- Tympanogram compliance and pressure normal
- Enhanced bone conduction thresholds
- Acoustic reflexes present despite air/bone gap and CHL
- Auditory and vestibular complaints present
- VEMPS present and enhanced

Question 9 ...In differentiating otosclerosis from superior canal dehiscence, what test would help indicate the latter?
A. Vestibular evoked myogenic potentials (VEMPS) would be absent in SCD and present in otosclerosis
B. Acoustic reflexes would be present in SCD despite air/bone gap
C. Acoustic reflexes would be absent in SCD due to air/bone gap
D. The low frequency CHL in otosclerosis would be greater than in SCD
SCDS without enhanced bone conduction

- LF CHL, AS
- Bone conduction not enhanced
- Differential diagnosis of SCDS, otosclerosis, or other middle ear disorder
- Include tympanometry, acoustic reflexes and VEMPS

Additional symptoms of Otosclerosis

- Osteospongeosis in early stage of the disease-otosclerosis as bone hardens.
- 20-30% of patients will develop cochlear otosclerosis (Hall 2014)
- 80% bilateral (Hall 2014)
- Caucasian, 2:1 female, 30-50 yrs.
- Normal TM with Schwartz sign
- Slowly progressing loss-not sudden
- Paracusis willisi
- CHL greater in low frequencies
- Carhart’s notch at 2 kHz

Normal TM with Schwartz sign
Bilateral otosclerosis with Carhart notch at 2kHz

In summary....SCD vs Otosclerosis

- Always assess bone conduction less than 0 dB HL
- Bone conduction testing on normal hearing patients with report of dizziness or vertigo
- Order acoustic reflexes on patient with LF CHL, air bone gap, and normal tympanograms.
- VEMPS will be present/enhanced for SCDS and absent for otosclerosis (due to true CHL)
- Differential diagnosis will help prevent unnecessary exploratory middle ear surgery and/or stapedectomy
- CT scan for diagnosis

Question 10: Based on the configuration of this audiogram, what disorder would you consider?

A. Bilateral otosclerosis
B. Vestibular Schwannoma, AS
C. Superior canal dehiscence, AD
D. Bilateral Meniere’s disease
Meniere’s Disease

- Audiometric configuration:
  - SNHL, rising configuration at initial stages of disease.
  - Fluctuating hearing loss, tinnitus, vertigo, and pressure in affected ear.
  - Fluctuations in hearing and vertigo major parts of criteria for diagnosis.
  - Most common in 40-60 age range and women (Hall 2014).
  - 12% have bilateral involvement within 8 years (Hall 2014)

Nonorganic Hearing Loss

- Exaggerated or false hearing loss that is not supported by objective audiometric testing.

  - Risk factors:
    - Accident or injury involving a lawsuit
    - “Sudden” hearing loss following an accident or incident
    - Adolescent girls with unexplained hearing loss
    - Exaggerating hearing loss with history of noise exposure
    - Disability claims
    - Hx of abuse (especially in children)
    - Patients under psychiatric care
    - Children seeking attention

Differential Diagnosis of Meniere’s Disease

- Cogan’s syndrome-Will also have ocular symptoms
- Autoimmune disease-Fluctuating hearing loss but may not have vertigo (Kramer, 2014)
- Vestibular neuritis: Vertigo but without aural fullness
- Syphilis
- NF2: Bilateral hearing loss, poor word recognition
- Cochlear Meniere’s disease-No vestibular symptoms
- Vestibular Meniere’s disease
  (Martin and Clark, 2015)
Other testing in diagnosis of Meniere’s disease

- Repeat audiometric testing to document fluctuations in pure tones and word recognition scores.
- Vestibular work up including electrocochleography
  - Enhanced SP/AP ratio
  - Vestibular weakness on affected side depending on stage of disease
  - Bilateral vestibular weakness?
- Normal middle ear function despite “pressure” in the ear.
- Acoustic reflexes and OAEs will coincide with audiogram.
- Repeat audiometric testing to document fluctuations in pure tones and word recognition scores.
- Vestibular work up including electrocochleography
  - Enhanced SP/AP ratio
  - Vestibular weakness on affected side depending on stage of disease
  - Bilateral vestibular weakness?
- Normal middle ear function despite “pressure” in the ear.
- Acoustic reflexes and OAEs will coincide with audiogram.

Key Factors on audiometric testing

- Patient is able to carry on conversation despite “hearing loss” on audiogram
- Difficult to establish thresholds inconsistent responses
- PTA and SRT differ by more than 10 dB HL
- Word recognition scores not consistent with audiogram
- Bone conduction scores worse than air conduction
- Mixed hearing loss without evidence of middle ear pathology
- Stenger and Lombard testing positive (Hall 2014)

Objective testing for Nonorganic hearing loss

- Otoacoustic emissions will be normal and not in agreement with degree of “hearing loss”
- Acoustic reflex testing at normal levels and not in agreement with pure tone loss.
- Auditory brainstem response testing
  - Threshold estimation with ABR Wave V for click and tone burst stimuli.
- Repeat audiometric testing and re-instruction.
- Referral for counseling as necessary.
Last but not least…. Auditory Neuropathy Spectrum Disorder
Lack of transmission of sound from the inner ear to the brain. Disorder discovered in the 1990s when OAEs became available.

• **Where is the issue?** Difficult to pinpoint however the following are likely:
  – Inner hair cells
  – Connection or synapse between IHC and auditory nerve
  – Auditory nerve

• **Causes?**
  – Infants with birth complications (low birth weight, jaundice, oxygen deprivation)
  – Genetics

Symptoms and test findings of ANSD

• Various thresholds on audiogram, according to Hall (2014), low-frequency hearing loss is the most common.
• Poor functional speech understanding in quiet and noise (worse than expected based on pure tones)
• Acoustic reflexes absent
• Present OAEs with absent ABR
• ABR findings: Cochlear microphonic and “mirrored” waveforms with polarity reversal.

Avoiding errors in audiograms

**Undermasking:**
  – Can cause false conductive hearing loss for bone conduction
  – Poor word recognition with “CHL”
  – Present thresholds in single-sided deafness (undermasking)
  – Abnormally good WRs for single sided SNHL

**Poorly inserted inserts**
  – False CHL in low frequencies with normal tympanograms

**Collapsing canals**
  – CHL in high frequencies with normal tympanograms
Avoiding Audiogram errors

- Unmasked bone conduction with air/bone gap causing false CHL
- Patient reliability
- Audiologist experience
- Equipment issues
- Confirm audiometric results are consistent with case history and tuning forks.

Martin and Clark, 2015

In Conclusion.....

- Audiogram alone can determine type, degree of hearing loss, and insight into certain disorders.
- Use additional testing for differential diagnosis:
  - Tympanograms
  - Acoustic reflex and decay testing
  - Otoacoustic emissions
  - Auditory brainstem response
  - Electrocochleography
  - VEMPS
  - Bone conduction testing below 0 dB HL

Let’s try some questions again......
**Question 11:** Based on the configuration of this audiogram, what disorder would you consider?

A. Bilateral otosclerosis  
B. Vestibular Schwannoma, AS  
C. Bilateral Meniere’s disease  
D. Superior canal dehiscence, AD

**Question 12:** What type of hearing loss is present, AS, and what would be a probable diagnosis?

A. Mixed, Large vestibular aqueduct syndrome  
B. Mixed, Cochlear otosclerosis  
C. SNHL, Sudden idiopathic SNHL  
D. SNHL, Meniere’s disease

**Question 13:** Based on this audiogram and assuming the patient had normal tympanometry, what possible disorder could cause this hearing loss, left ear?

A. Meniere’s disease  
B. Otosclerosis  
C. Otitis media with effusion  
D. TM perforation
**Question 14:** What disorder(s) would you suspect with this audiogram?

A. Bilateral vestibular schwannomas  
B. Meniere’s disease, AD and presbycusis AS  
C. Bilateral Meniere’s disease  
D. Presbycusis, AS and Vestibular schwannoma, AD

**Question 15:** Why is this audiogram consistent with possible superior canal dehiscence?

A. Mixed hearing loss at 4000 Hz  
B. Enhanced thresholds for LF bone conduction  
C. High frequency conductive hearing loss  
D. Decreased bone conduction at 2000 Hz.

**Question 16** and final...In differentiating otosclerosis from superior canal dehiscence, what statement would apply?

A. Acoustic reflexes would be present in SCD despite air/bone gap  
B. Vestibular evoked myogenic potentials (VEMPS) would be absent in SCD and present in otosclerosis  
C. Acoustic reflexes would be absent in SCD due to air/bone gap  
D. The low frequency CHL in otosclerosis would be greater than in SCD
THANK YOU!
Questions? Comments?
Lori Klingenberg, Au.D CCC-A
lklingenberg@ENTorlando.com

References