Hearing Loss Case

A Self-Directed Learning Module

Department of Otolaryngology – Head & Neck Surgery
Schulich School of Medicine & Dentistry, Western University

Click to Begin

Case Presentation

A 32-year-old female teacher presents to your family practice with a five year history of slowly progressive bilateral hearing loss.

You are the family physician, click through the module to diagnose and treat this patient.

Obtain a history

Back to Case Presentation

Patient History

"My hearing has been getting worse for the past five years. I often have trouble hearing people's voices and have to ask them to repeat themselves. The hearing loss is always there, along with some ringing in my ears. My hearing has been getting progressively worse, and I noticed more of a drop when I was pregnant. I have not had any dizziness or ear pressure and I haven't tried anything for the hearing loss so far.

Otherwise, I don't have any other medical problems. I do not take any medications, or have any allergies.

My mother had quite poor hearing, and so did her father. However, I don't think they were given a diagnosis."

What is on your differential diagnosis so far?

Back to Case Presentation

What diagnoses are coming to mind?

Think VINDICATE! (Click on heading to test yourself)

Vascular

Endocrine/Metabolic

Infectious

Trauma/Toxins

Neoplastic

Autoimmune/Allergic

Degenerative

Congenital/Genetic

Iatrogenic/Idiopathic

With your DDx in mind, proceed to focused physical exam.

Back to Case Presentation

What diagnoses are coming to mind?

Think VINDICATE! (Click on heading to test

Vascular - Unlikely X

Why not?
Bilateral and progressive hearing loss. No pulsatile characteristic.

Endocrine/Metabolic

Infectious

Trauma/Toxins

Neoplastic

Autoimmune/Allergic

Degenerative

Congenital/Genetic

Iatrogenic/Idiopathic

With your DDx in mind, proceed to focused physical exam.

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What diagnoses are coming to mind?

Think VINDICATE! (Click on heading to test yourself)

Vascular

Endocrine/Metabolic

Trauma/Toxins

Neoplastic

Autoimmune/Allergic

Degenerative

Congenital/Genetic

Iatrogenic/Idiopathic

Infectious - Unlikely

Why not?
Five year history, general good health

DDx
Luetic (otosyphilis) hearing loss

With your DDx in mind, proceed to focused physical exam.

Back to Case Presentation

What diagnoses are coming to mind?

Think VINDICATE! (Click on heading to test yourself)

Vascular

Endocrine/Metabolic

Infectious

Trauma/Toxins

Autoimmune/Allergic

Congenital/Genetic

Iatrogenic

X

Neoplastic

Why?
Progressive history over years.

DDx
Temporal bone neoplasm
Glomus jugulare tumor
Cholesteatoma (*NOT cancerous; keratinizing squamous epithelium in the temporal bone)

With your DDx in mind, proceed to focused physical exam.

Back to Case Presentation

What diagnoses are coming to mind?

Think VINDICATE! (Click on heading to test yourself)

Vascular

Endocrine/Metabolic

Infectious

Trauma/Toxins

Neoplastic

Autoimmune/Allergic

Congenital/Genetic

Iatrogenic/Idiopathic

Degenerative - Unlikely X

Why not?
Young age and no fluctuating course

DDx
Presbycusis
Multiple Sclerosis

With your DDx in mind, proceed to focused physical exam.

Back to Case Presentation

What diagnoses are coming to mind?

Think VINDICATE! (Click on heading to test yourself)

Vascular

Endocrine/Metabolic

Infectious

Trauma/Toxins

X Neoplastic

Autoimmune/Allergic

egenerative

Congenital/Genetic

Iatrogenic/Idiopathic - Unlikely

Why not?
No previous otologic surgery. No fluctuating course or associated vertigo.

DDx
Meniere's disease

With your DDx in mind, proceed to focused physical exam.

Back to Case Presentation

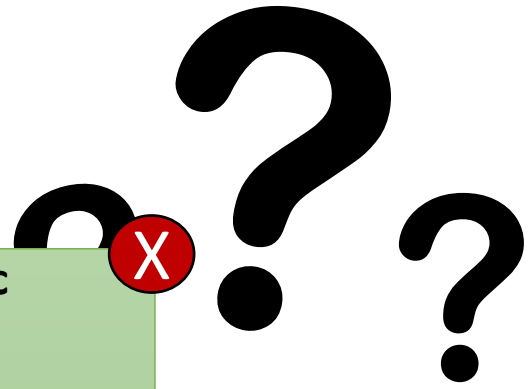
What diagnoses are coming to mind?

Think VINDICATE! (Click on heading to test yourself)

Vascular

Endocrine/Metabolic

Infectious



Trauma/Toxins

Neoplastic

Congenital/Genetic

Why?
Congenital less likely given prior normal hearing, but genetic diagnoses are possible.

DDx
Paget's disease
Osteogenesis imperfecta
Otosclerosis

A

Degenerative

Iatrogenic/Idiopathic

With your DDx in mind, proceed to focused physical exam.

Back to Case Presentation

What diagnoses are coming to mind?

Think VINDICATE! (Click on heading to test yourself)

Vascular

Endocrine/Metabolic

Infectious

Trauma/Toxins

Neoplastic

X

Autoimmune/Allergic

Why?
Middle-aged female.

DDx
Autoimmune inner ear disease

Degenerative

tal/Genetic

Iatrogenic/Idiopathic

With your DDx in mind, proceed to focused physical exam.

Back to Case Presentation

What diagnoses are coming to mind?

Think VINDICATE! (Click on heading to test yourself)

Vascular

Endocrine/Metabolic

Infectious

Trauma/Toxins - Unlikely X

Why not?
Unlikely based on history, but important to rule out on history.

DDx
Ototoxic medication
Noise-induced hearing loss
Head trauma
Lead and mercury toxicity
Barotrauma
Cerumen impaction

Neoplastic

Degenerative

al/Genetic

Iatrogenic/Idiopathic

With your DDx in mind, proceed to focused physical exam.

Back to Case Presentation

What diagnoses are coming to mind?

Think VINDICATE! (Click on heading to test yourself)

Endocrine/Metabolic - Unlikely X

Why not?
Unlikely based on history and no suggestive associated symptoms.

DDx
Hypothyroidism

Vascular

Infectious

Neoplastic

Autoimmune/Allergic

Degenerative

Congenital/Genetic

Iatrogenic/Idiopathic

With your DDx in mind, proceed to focused physical exam.

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Physical Exam *(Click on the physical examinations.)*

General inspection & Vitals

Cranial Nerve Exams

Head & Neck Exam

Cerebellar Tests

Proceed to investigations

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Physical Exam *(Click on the physical examinations.)*

General inspection & Vitals

Cranial Nerve Exams

Head & Neck Exam

Cerebellar Tests

Patient looks well and doesn't appear to be in any distress.

Vitals:

HR: 80 bpm
RR: 12 per minute
BP: 122/80 mmHg
Temperature: 37℃

Proceed to investigations

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Physical Exam *(Click on the physical examinations.)*

General inspection & Vitals

Cranial Nerve Exams

Head & Neck Exam

Cerebellar Tests

Cranial Nerve Exams:

CN I – VII, IX - XII : Within normal limits

CN VIII:
Weber test: No lateralization
Rinne test: Bone conduction > Air conduction

Review: Cranial Nerve Exams

Review: Weber & Rinne tests

Proceed to investigations

Review: Cranial Nerve Exams

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Cranial Nerve	Name	Function	Test
I	Olfactory Nerve	• Smell	• Test for sense of smell with coffee, alcohol swab, citrus, etc.
II	Optic Nerve	• Vision	• Visual acuity – Snellen's eye chart • Visual fields – Confrontation testing • Pupillary reflexes – Direct and consensual response • Fundoscopy
III	Oculomotor Nerve	• Motor innervation to most* extra-ocular muscles	• H test & convergence • Pupillary reflex
IV	Trochlear Nerve	• Motor innervation to superior oblique muscle*	• H test : Look for ability to look "down and out"
V	Trigeminal Nerve	• Sensory innervation to the face • Motor innervation to muscles of mastication	• Corneal reflex • Test sensory supply to the face – cotton swab • Test strength of muscles of mastication
VI	Abducens Nerve	• Motor innervation to lateral rectus muscle*	• H test: Look for ability to abduct eye
VII	Facial Nerve	• Motor innervation to muscles of facial expression • Taste – anterior 2/3 tongue	• Ask patient to do different facial expressions • Corneal reflex
VIII	Vestibulocochlear Nerve	• Hearing & balance	• Weber & Rinne tests
IX	Glossopharyngeal Nerve	• Sensory innervation to the palate • Taste – posterior 1/3 tongue	• Gag reflex • Say "Ahhh" – look for deviation of the uvula
X	Vagus Nerve	• Motor supply to the pharynx • PSNS supply to abdominal viscera	• Gag reflex
XI	Spinal Accessory Nerve	• Motor innervation to trapezius and sternocleidomastoid	• Shoulder shrug against resistance, head turn against resistance
XII	Hypoglossal Nerve	• Motor supply to muscles of the tongue	• Stick tongue out and move tongue from side-to-side

Review: Weber & Rinne tests

Back to Physical Exam

Back to Case Presentation

Review: Weber and Rinne tests

1. Weber test

- Strike a 512 Hz tuning fork and place on top of the patient's head
- A patient with normal hearing should hear the sound equally on both sides* (I.e. the sound shouldn't lateralize to one ear)
  - *Note: A Rinne test is needed to confirm normal hearing, as a patient with bilateral conductive hearing loss would also have no lateralization of sound.

2. Rinne test

- Strike a 512 Hz tuning fork and place it on the mastoid bone behind the patient's ear (Testing bone conduction (BC))
- When the patient signals that they can no longer hear the sound, move the tuning fork next to the patients external auditory canal (Testing air conduction (AC))
- A patient with normal hearing should hear the sound better through air conduction (AC > BC)
- Rinne test might not be negative if the conductive hearing loss is very mild

Results from the Weber and Rinne test can be used to determine the type of hearing loss:

Test	**Normal**	**Conductive Hearing Loss**	**Sensorineural Hearing Loss**
Weber	Sound heard in midline	Sound heard in affected ear	Sound heard in good ear
Rinne	AC > BC	BC > AC	AC > BC

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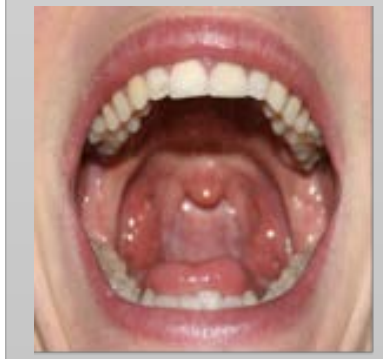
Focused Head & Neck Exam

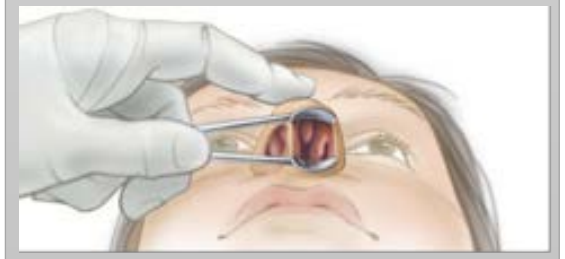
Inspection:
- No scars, asymmetry, enlarged thyroid or parotids, skin lesions
- No spontaneous nystagmus

Palpation:
- No palpable lymph nodes, salivary glands palpable, thyroid not palpable

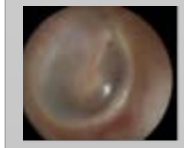
Oral Cavity Exam:



Nasal speculum exam:
- Turbinates, nasal mucosa, and nasal septum normal



Otoscopy:
- External auditory canals have minimal cerumen
- No foreign bodies, discharge or mass



Right Tympanic Membrane

Left Tympanic Membrane

Review: Otoscopy

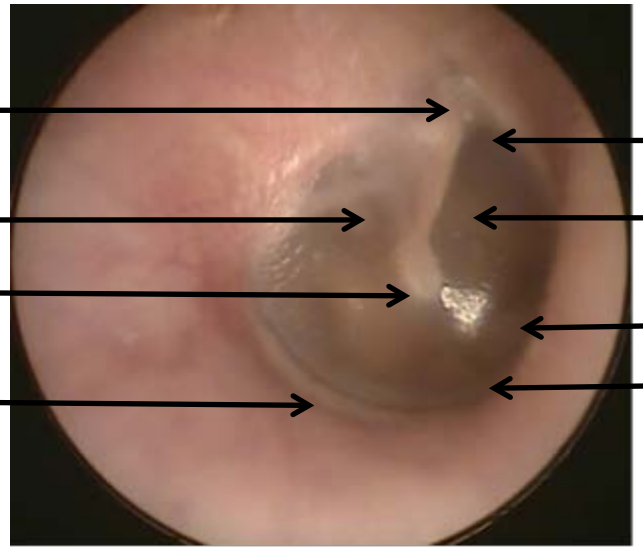
Back to Physical Exam

Review: Otoscopy

Normal Otoscopic Exam Findings:

- Tympanic membrane (TM) should be intact and appear pearly grey or whitish/pinkish grey

Normal TM – Right Side



Examples of Abnormal TMs

Bulging, red TM - Acute Otitis Media

Cholesteatoma

Otitis media with effusion

Myringosclerosis

Hemotympanum

Retracted TM

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Physical Exam *(Click on the physical examinations.)*

General inspection & Vitals

Cranial Nerve Exams

Head & Neck Exam

Cerebellar Tests

Cerebellar Tests

- Finger-nose, heel-shin, and rapid alternating movements tests are normal
- Gait (including tandem gait) is normal
- Rhomberg is normal

Proceed to investigations

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Investigations *(Click on the buttons to see investigation results.)*

Cochlear/Acoustic Testing

Vestibular Testing

Temporal Bone CT Scan

Internal Auditory Canal MRI

Continue to Diagnosis

Cochlear/Acoustic Testing

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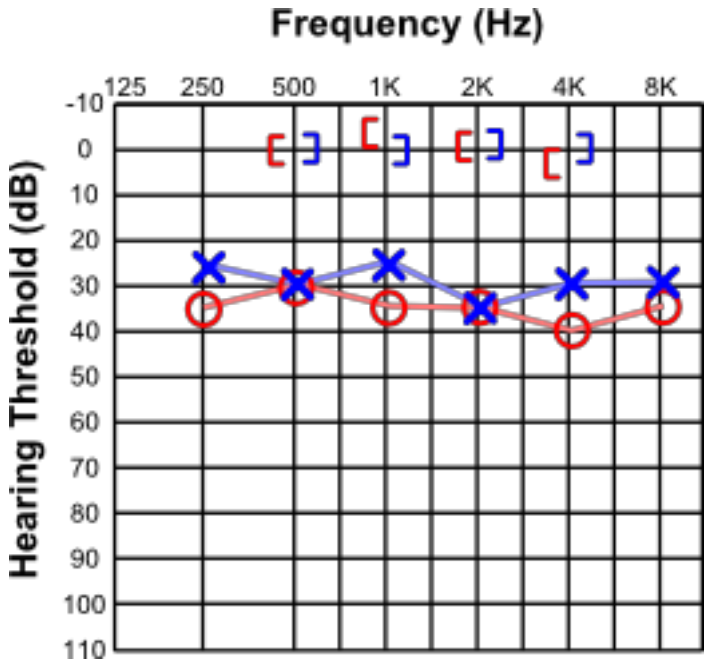
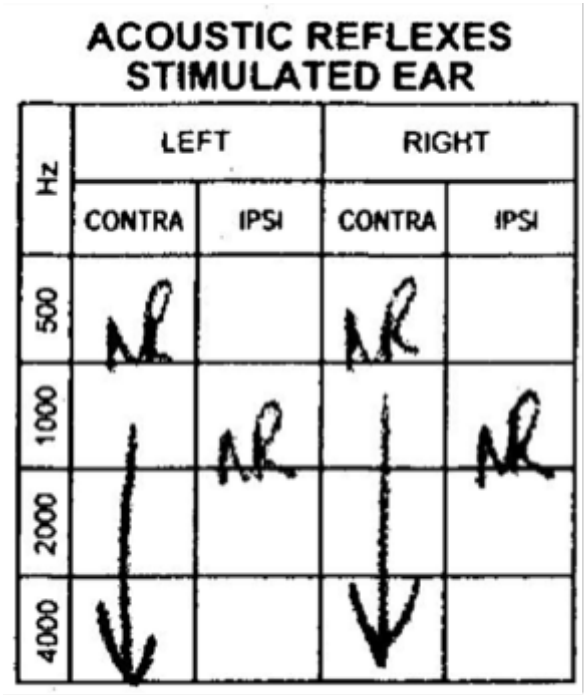
Audiogram



Test: Within normal limits?

Unsure? **Review:** Audiology Testing

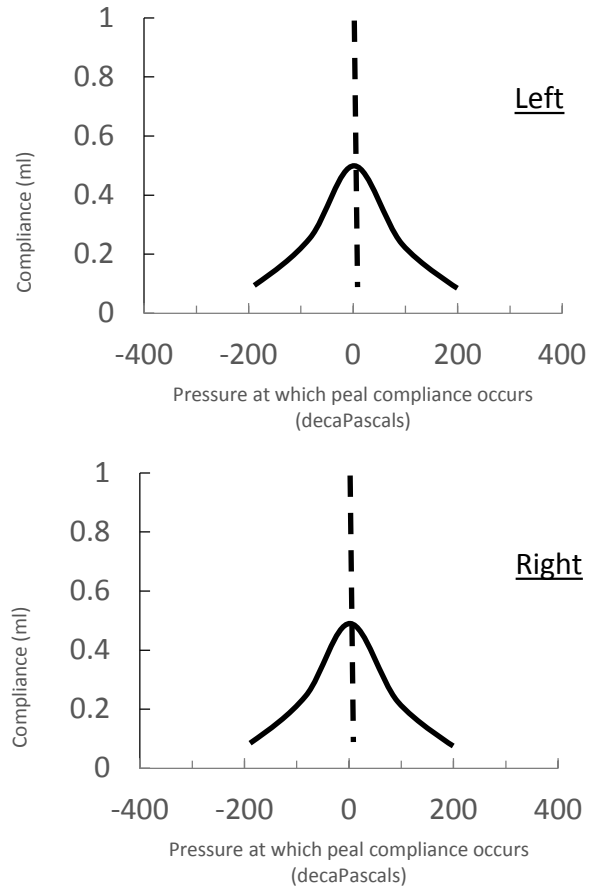
Acoustic Reflex



Test: Within normal limits?

Unsure? **Review:** Acoustic Reflex

Tympanometry



Test: Within normal limits?

Unsure? **Review:** Tympanometry

Cochlear/Acoustic Testing

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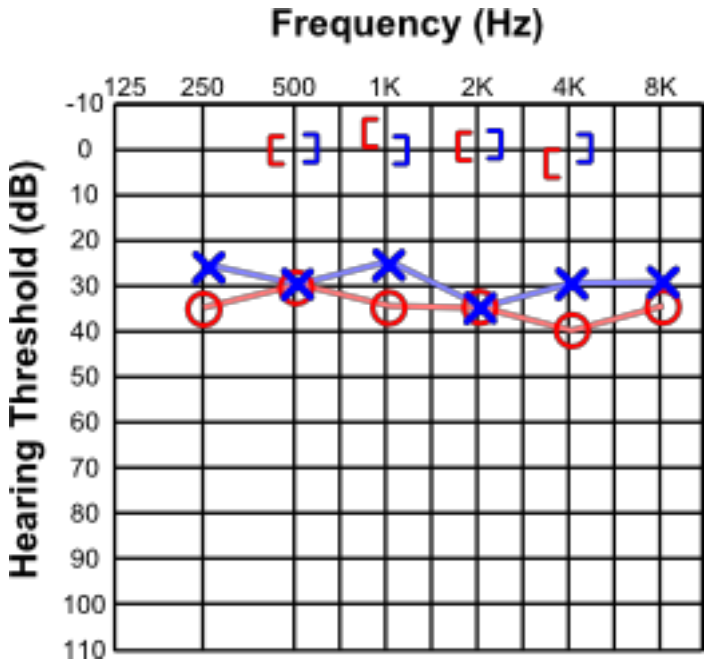
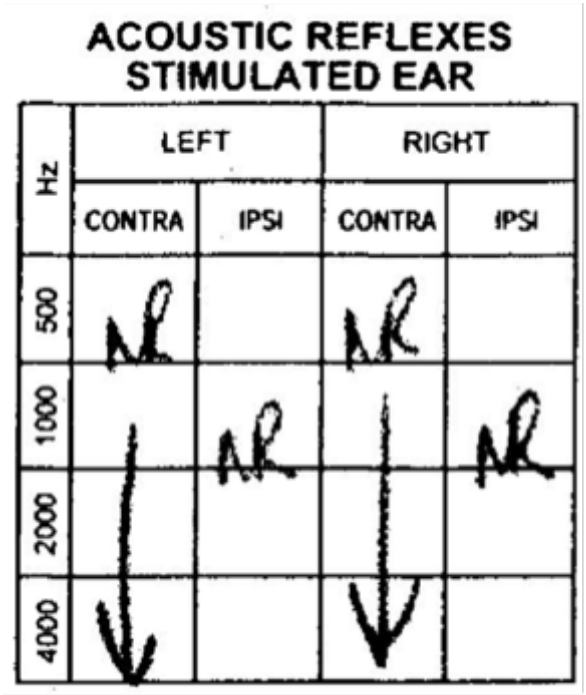
Audiogram



No, the audiogram shows bilateral conductive hearing loss.

Unsure? **Review:** Audiology Testing

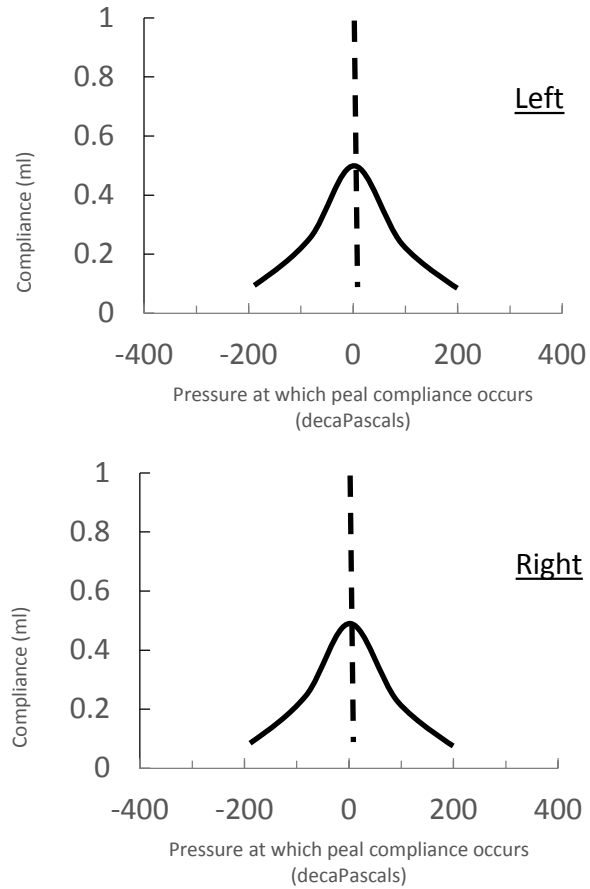
Acoustic Reflex



Test: Within normal limits?

Unsure? **Review:** Acoustic Reflex

Tympanometry



Test: Within normal limits?

Unsure? **Review:** Tympanometry

Cochlear/Acoustic Testing

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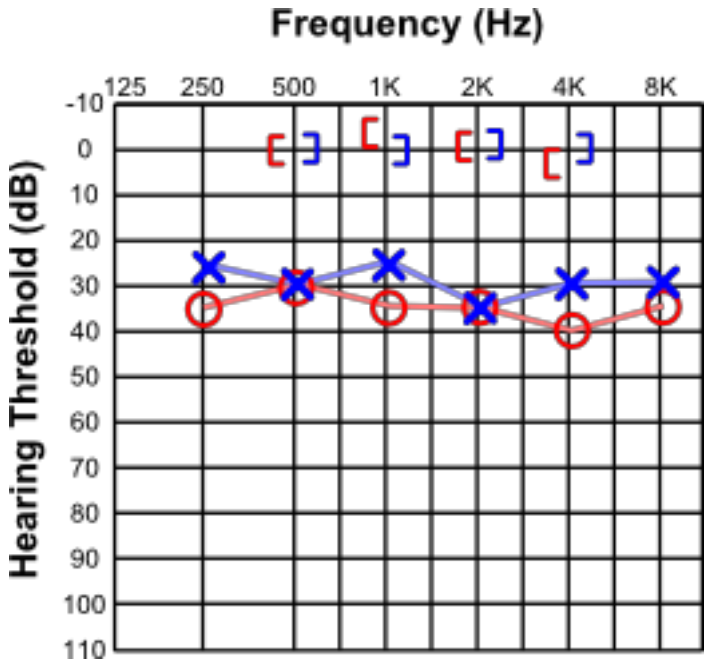
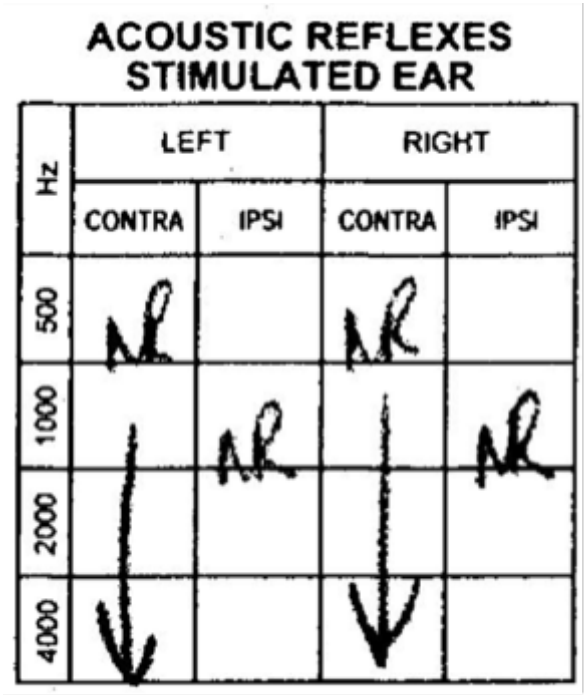
Audiogram



Test: Within normal limits?

Unsure? **Review:** Audiology Testing

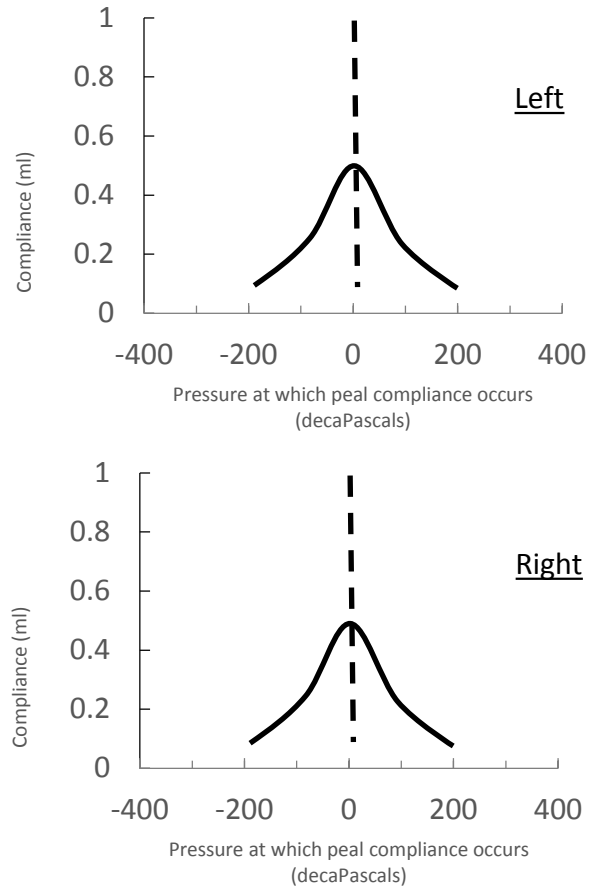
Acoustic Reflex



No, acoustic reflexes are absent bilaterally.

Unsure? **Review:** Acoustic Reflex

Tympanometry



Test: Within normal limits?

Unsure? **Review:** Tympanometry

Cochlear/Acoustic Testing

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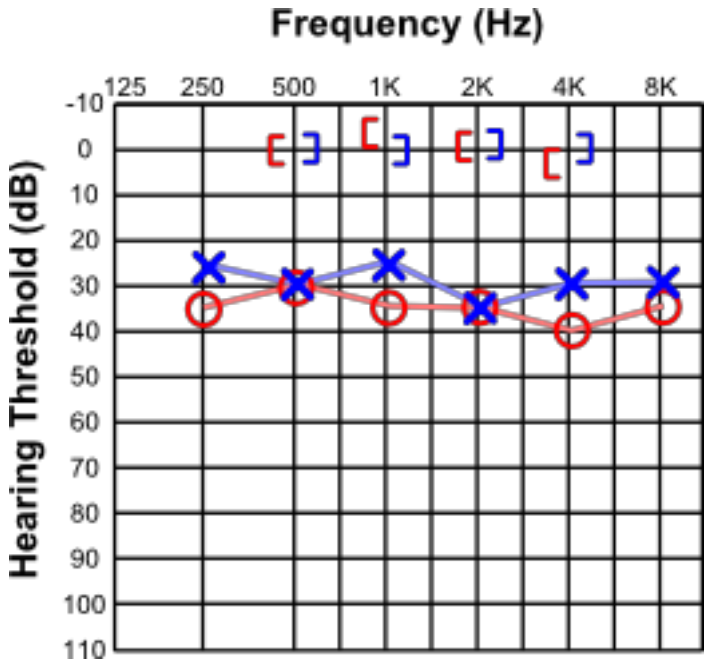
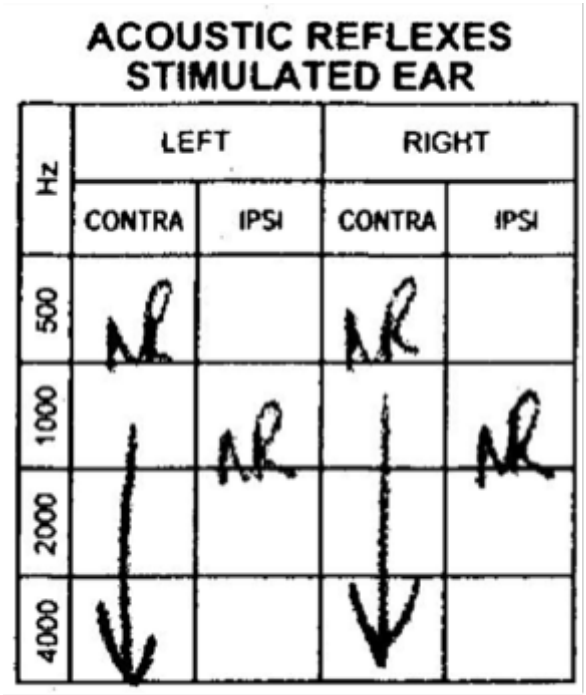
Audiogram



Test: Within normal limits?

Unsure? **Review:** Audiology Testing

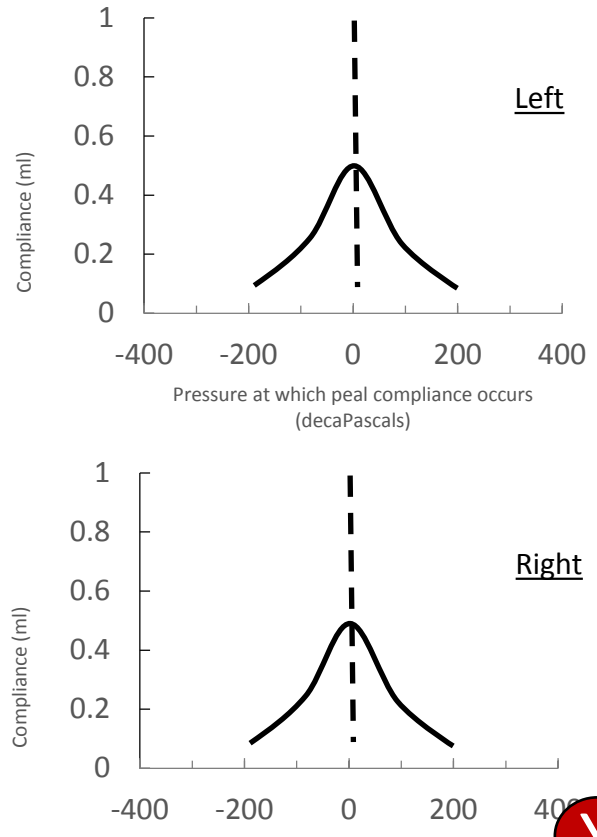
Acoustic Reflex



Test: Within normal limits?

Unsure? **Review:** Acoustic Reflex

Tympanometry



Yes, tympanometry is within normal limits.

Unsure? **Review:** Tympanometry

Review: Interpreting an Audiogram

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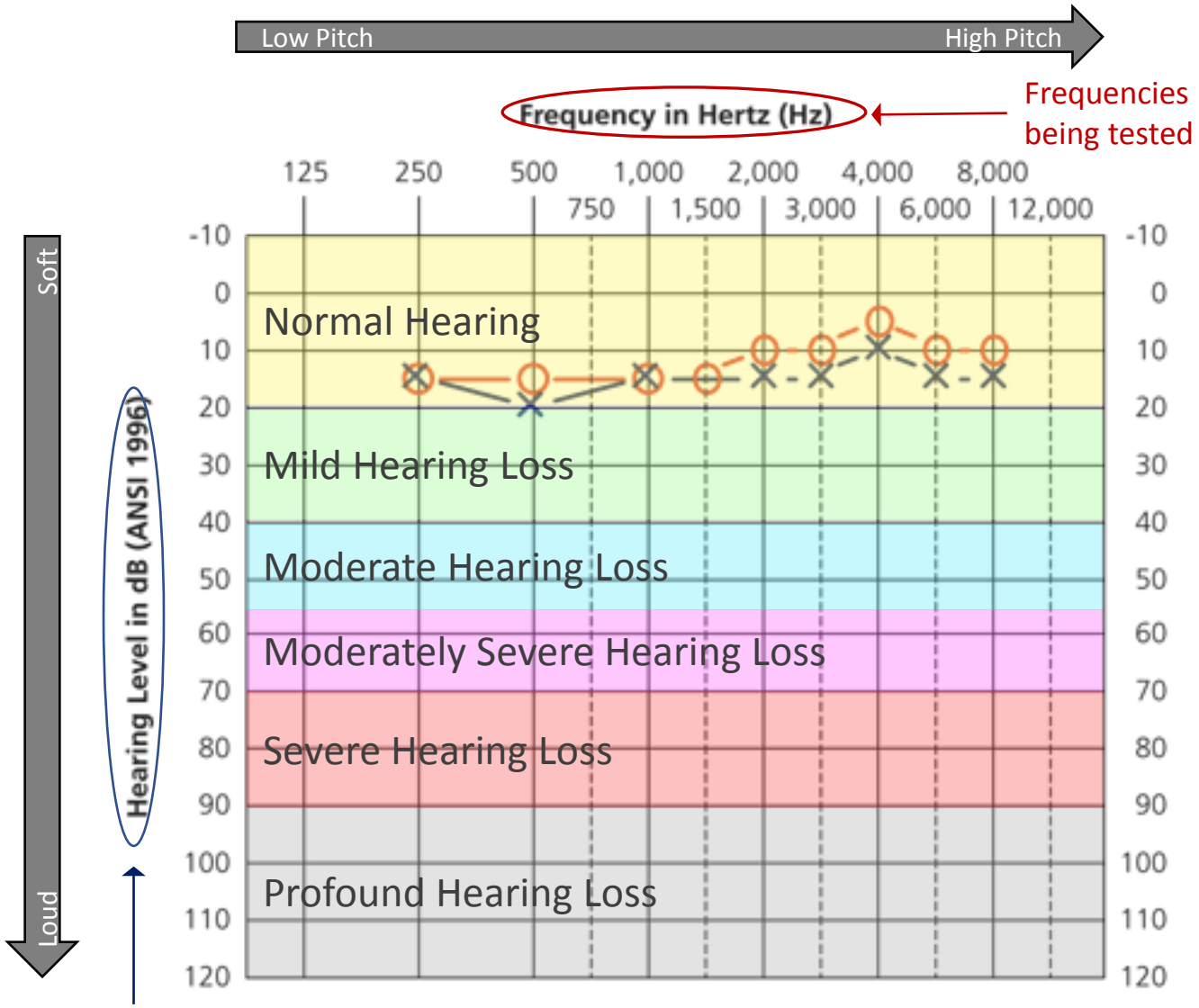

Air conduction testing:

- Sound delivered through headphones or loudspeakers, tests outer, middle, and inner ear.
  - Left ear = X
  - Right ear = O
- Different symbols are used when "masking" is used. Masking refers to noise presented to the non-test ear to prevent it from hearing sound presented to the test ear.
  - Left ear = ⍰
  - Right ear = △

Bone conduction testing:

- Bone vibrator placed behind the ear to deliver sound vibrations to the cochlea, bypassing the outer and middle ear.
  - Left ear = >
  - Right ear = <
- Masking symbols
  - Left ear =]
  - Right ear = [

Click to practice reading audiograms

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Review: Interpreting an Audiogram

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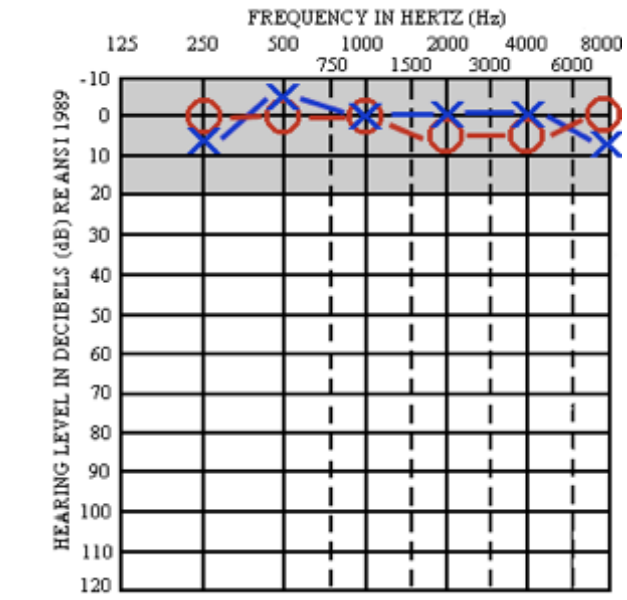
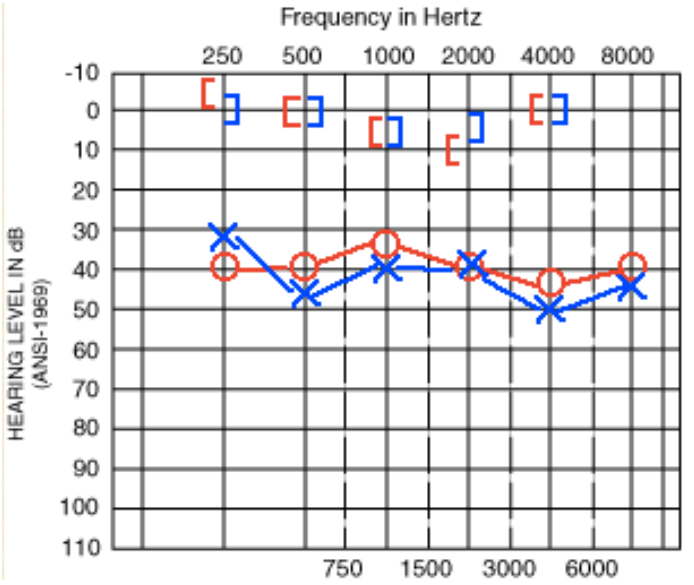


Unmasked air conduction audiogram.

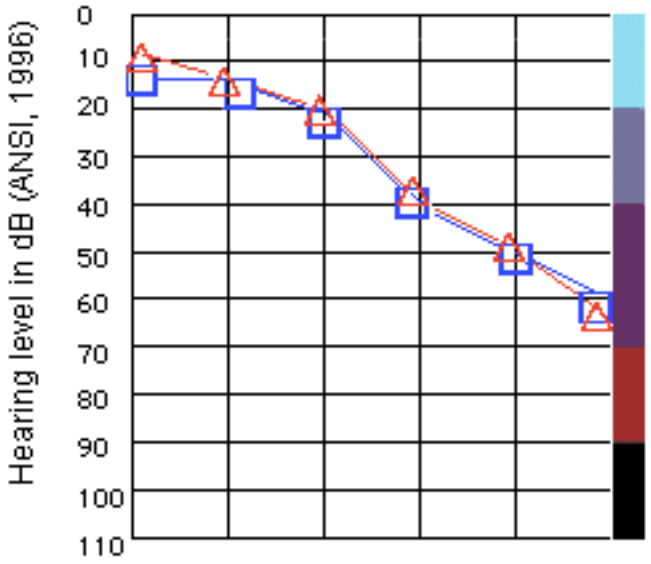
Interpretation:
Normal hearing



Air conduction testing compared to masked bone conduction testing.

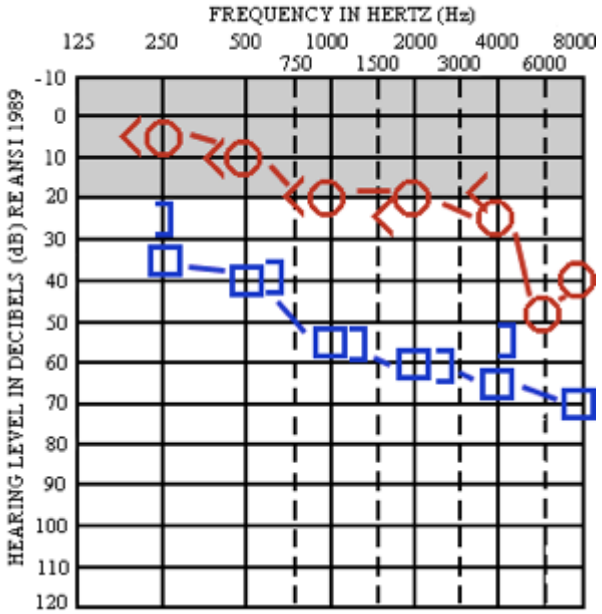
- Bone conduction: within normal range
- Air conduction: mild – moderate hearing loss

Interpretation:
Conductive hearing loss.
(I.e. Middle ear pathology)



Masked air conduction audiogram.

Interpretation: Moderately severe hearing loss at high frequencies.
(I.e. Presbycusis)



Right ear: Unmasked air and bone conduction testing
Left ear: Masked air and bone conduction testing

Interpretation:
Asymmetrical sensorineural hearing loss.
(I.e. Acoustic neuroma)

- Right ear: mild sensorineural hearing loss at higher frequencies
- Left ear: Mild to moderately severe hearing loss as move up frequencies

Review: Acoustic Reflex Testing

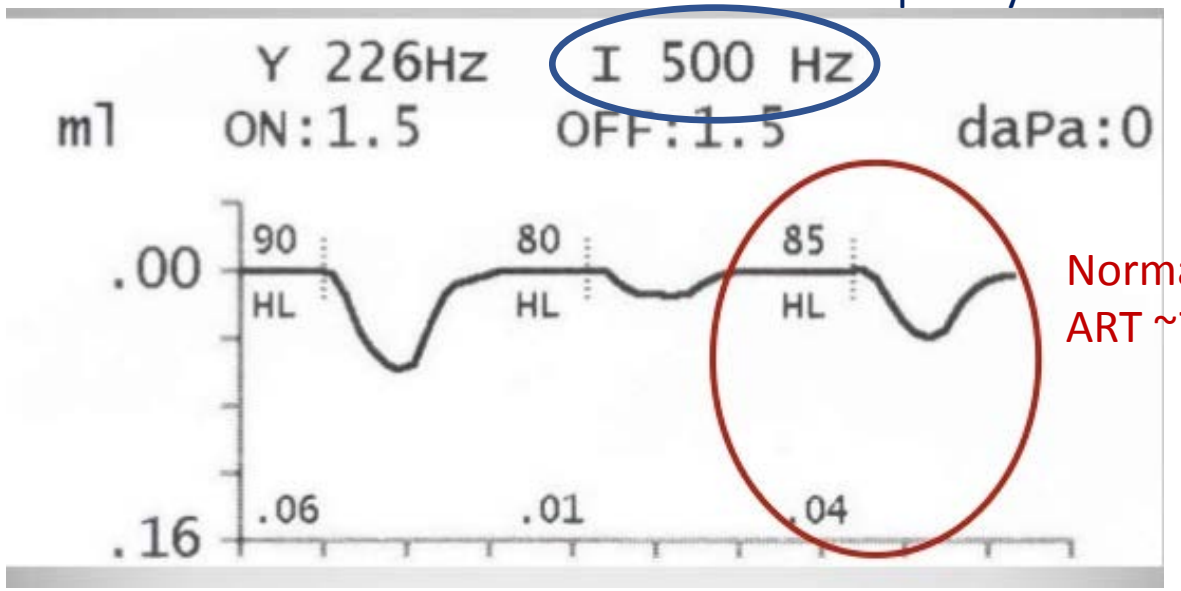
Back to Case Presentation

The acoustic reflex is the reflexive contraction of the stapedius muscle, and subsequent stiffening of the tympanic membrane (TM), in response to high-intensity sound or vocalization. *Anatomy reminder: stapedius is innervated by CN VII.

In acoustic (stapedial) reflex testing, acoustic signals at varying frequencies (usually 500, 1000, or 2000 Hz) are introduced into one ear and the acoustic impedance is measured in the both ears.

Acoustic Reflex Threshold (ART): Sound pressure level (SPL), in dB, from which a sound stimulus with a given frequency will elicit the acoustic reflex.

Frequency tested



Normal hearing:
ART ~70-100 dB SPL

Reflexes may be absent or harder to illicit in patients with:

- Conductive hearing loss
  - I.e. fixation of the ossicles
- Severe sensory hearing loss
- CN8 injury on side receiving sound
- CN7 injury on side being measured

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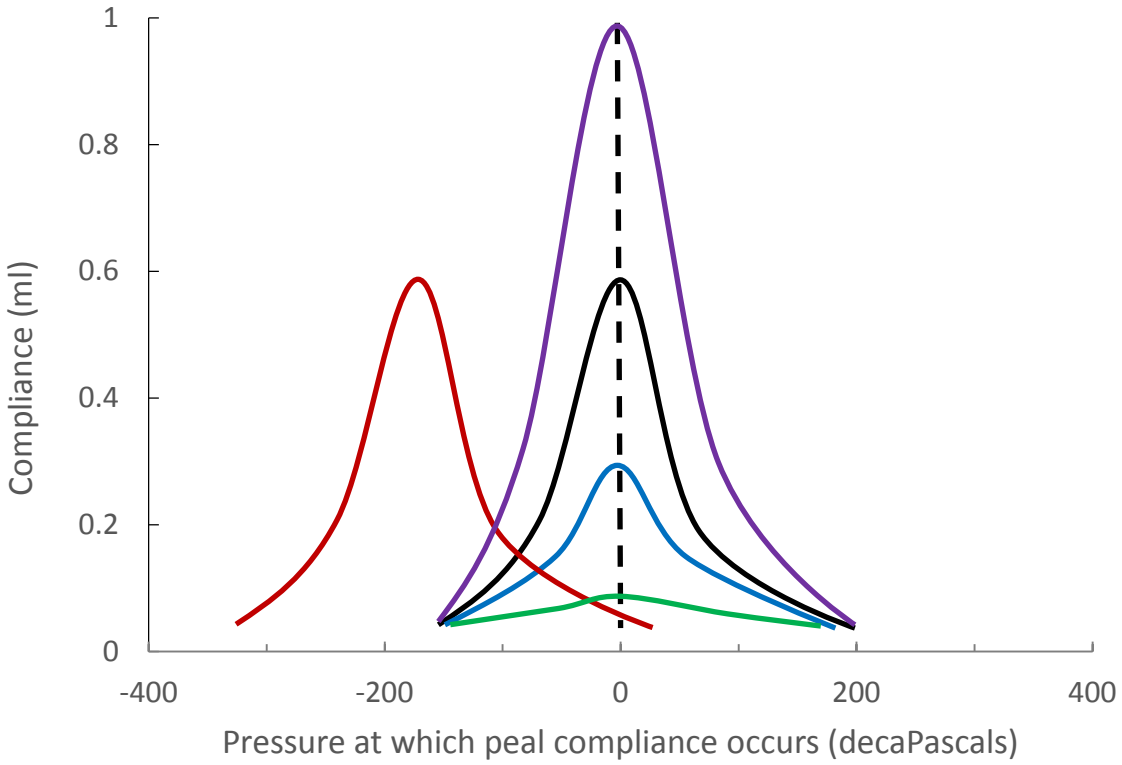
Review: Tympanometry

Tympanometry is an indirect test of middle ear function by the transmission/reflection of sound energy. A tympanogram plots compliance changes of the tympanic membrane (TM) versus air pressure in the external auditory canal.



High peak = hypercompliant TM
- I.e. Ossicular discontinuity, monomeric TM (thin TM from healed TM perforation)

Normal tympanogram

Shallow peak = stiff TM
- I.e. Otosclerosis, tympanosclerosis

No peak = non-mobile TM
- I.e. Effusion, perforation

Peak shifted to a more negative pressure = retracted TM
- I.e. Eustachian tube dysfunction, TM atelectasis

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Investigations *(Click on the buttons to see investigation results.)*

Cochlear/Acoustic Testing

Vestibular Testing

Unnecessary given no associated vestibular symptoms.

Temporal Bone CT Scan

Internal Auditory Canal MRI

Continue to Diagnosis

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Investigations *(Click on the buttons to see investigation results.)*

Cochlear/Acoustic Testing

Vestibular Testing

Temporal Bone CT Scan

Internal Auditory Canal MRI

Unnecessary given conductive loss with no associated vestibular or neurologic symptoms.

Continue to Diagnosis

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Investigations *(Click on the buttons to see investigation results.)*

Cochlear/Acoustic Testing

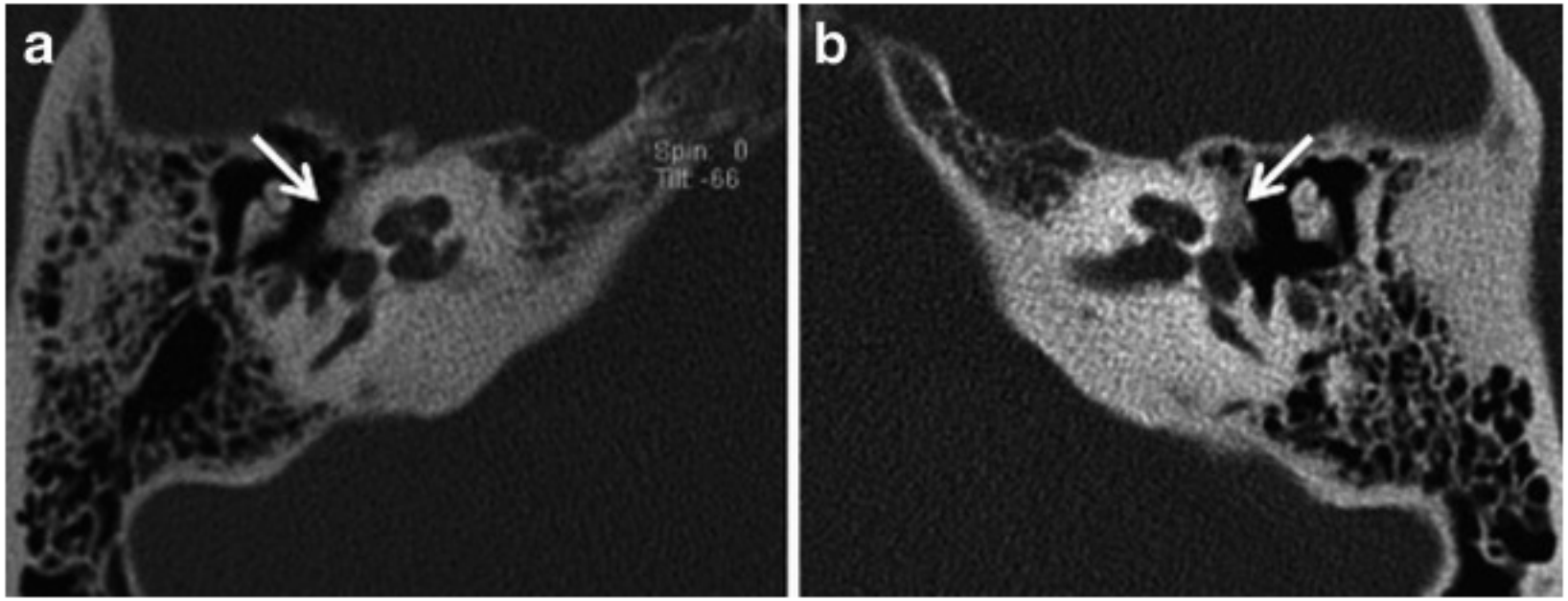
Vestibular Testing

Temporal Bone CT Scan

Internal Auditory Canal MRI

Right temporal bone – Axial plane

Left temporal bone – Axial plane



Interpretation: *Hypodense demineralized plaques (arrows)*

Review:
Reading a temporal bone CT scan

Continue to Diagnosis

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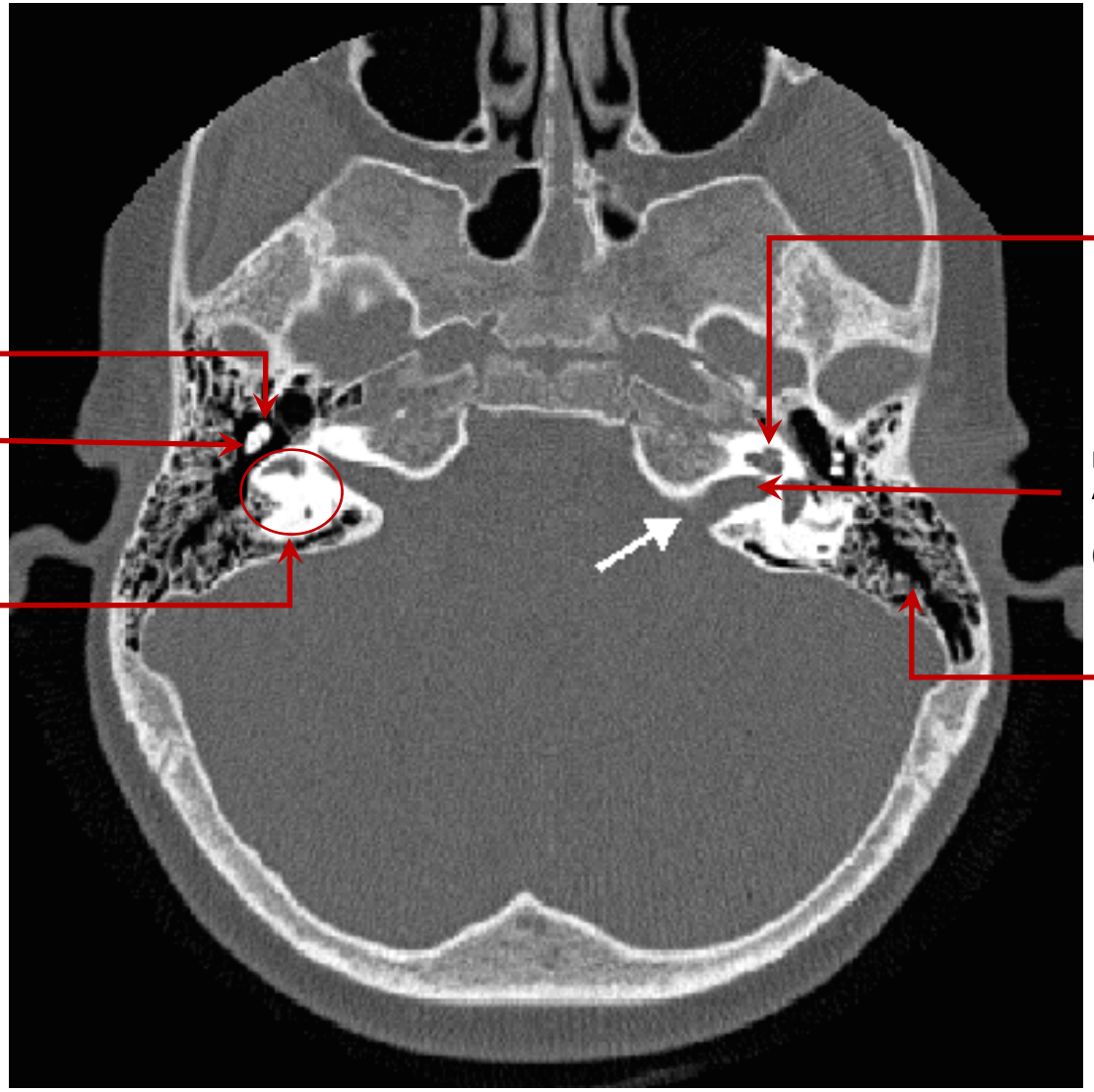
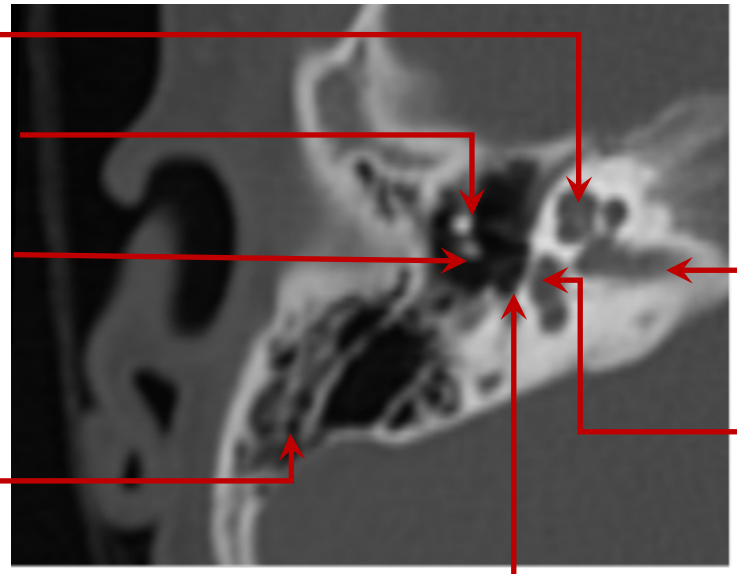
Review: Reading a Temporal Bone CT Scan

Normal temporal bone CT scan – Coronal plane



Magnified right temporal bone – Coronal plane



Back

Summary of findings

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Chief Complaints:
- 5 year history of progressive bilateral hearing loss
  - Exacerbated during pregnancy
- Episodic tinnitus

Physical Examination:
- Vitals and general inspection: normal
- Cranial nerve exams:
  - CN I – VII, IX – XII were normal
  - CN VIII
    - Weber test: No lateralization
    - Rinne test: BC > AC
- Head & neck exam: Normal
  - Otoscopy: Right ear Left ear

  - Positive Schwartze sign on left
- Cerebellar tests: Normal
- Systems review: Normal

Investigations:
- Acoustic/cochlear testing:
  - Bilateral conductive hearing loss, absent acoustic reflexes
- Temporal bone CT scan:
  - Bilateral hypodense demineralised plaques noted at fissula ante fenestramx

Based on your findings, choose the most likely diagnosis:

a. Presbycusis

b. Labyrinthitis

c. Otosclerosis

d. Paget's Disease

e. Ménière's Disease

f. Autoimmune inner ear disease

Back to Case Presentation

Diagnosis

Correct! The most likely diagnosis is **otosclerosis**. Otosclerosis is characterized by abnormal resorption and deposition of bone in the bony labyrinth and ossicles. Often patients become symptomatic over time due to stapes fixation and associated conductive hearing loss. Sensorineural hearing loss can also occur late in the disease progression. This is called cochlear otosclerosis and can be seen with demineralization of cochlea on CT (Double ring sign).

Otosclerosis is an autosomal dominant condition with incomplete penetrance. Symptoms begin to manifest by 20-40 years old. Pregnancy can be associated with acceleration of otosclerosis, as seen in our patient.

To note, definitive diagnosis of otosclerosis can only be made at the time of surgery or during a histological study of the temporal bone.

Now that you have made the correct diagnosis, choose the best treatment for this patient.

Treatments

Back to Case Presentation

Diagnosis

Incorrect. Presbycusis, also known as age-related hearing loss, is a progressive and irreversible sensorineural hearing loss, usually occurring after age 50.

While presbycusis can present in younger patients, our patient's occupation and lack of noise trauma in her history, makes age-related hearing loss highly unlikely. Further, our patient's audiogram showed a conductive hearing loss.

Please choose a different diagnosis.

Back to diagnosis

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Diagnosis

Incorrect. Our patient's five year history of slowly progressive bilateral conductive hearing loss and absence of vestibular symptoms is not consistent with a diagnosis of labyrinthitis.

Labyrinthitis is an infection within the inner ear that usually presents with sudden vertigo and sensorineural hearing loss.

Please choose a different diagnosis.

Back to diagnosis

Back to Case Presentation

Diagnosis

Incorrect. Paget's Disease is a rare autosomal dominant condition that causes excessive breakdown and abnormal remodeling of bone.

Paget's Disease involving the temporal bone could present similar to this case. However, Paget's typically causes sensorineural hearing loss due to compression of CN VIII within the internal auditory canal. Further, there were no signs of Paget's Disease on the CT scan.

Please choose a different diagnosis.

Back to diagnosis

Back to Case Presentation

Diagnosis

Incorrect. While our patient did report experiencing tinnitus, her audiogram showed a conductive hearing loss. Further, there were no associated vestibular symptoms.

Diagnosis of Ménière's Disease can be subdivided as definite versus probable. Below are the diagnostic criteria:

Definite Ménière's:

- Two or more spontaneous episodes of vertigo 20 minutes and 12 hours
- Low- to medium- frequency sensorineural hearing loss.
- Fluctuating aural symptoms (hearing, tinnitus and/or fullness) in the affected ears.

Probable Ménière's

- Episodic vestibular symptoms (vertigo or dizziness) 20 minutes to 24 hours
- Fluctuating aural symptoms (hearing, tinnitus or fullness)

Please choose a different diagnosis.

Back to diagnosis

Back to Case Presentation

Diagnosis

Incorrect. Autoimmune inner ear disease can manifest with progressive bilateral hearing loss and tinnitus. However, since it is caused by autoantibodies which attack the inner ear, it produces a sensorineural hearing loss, which is typically accompanied by vestibular symptoms.

Please choose a different diagnosis.

Back to diagnosis

Back to Case Presentation

Treatment for otosclerosis fits into 4 main categories.

Click on the links to learn about the treatments.

Observation

Medical management: Fluorides and Bisphosphonates

Amplification: Hearing aid

Surgery: Stapedotomy

Continue to Quiz

Back to Case Presentation

Observation

This option has the least risks and expense. It is preferred for patients with mild conductive hearing loss (i.e. this case) with minimal impact on quality of life.

If this option is chosen, the patient should be aware that the hearing loss will continue to progress slowly, and that yearly audiograms should be obtained.

Continue to Quiz

Back to treatments

Back to Case Presentation

Medical management: Fluorides and Bisphosphonates

Sodium fluorides and bisphosphonates have been proposed as options for medical therapy. These therapies theoretically prevent/slow the progression of otosclerosis through suppression of bone remodeling.

To date, medical treatment for otosclerosis remain controversial and has not been widely adopted.

Continue to Quiz

Back to treatments

Back to Case Presentation

Amplification: Hearing aid

Since the majority of patients with otosclerosis have normal cochlear function, they are good candidates for hearing aids.

Before a patient decides to have surgery, practitioners can encourage a trial hearing aids. This may help defer surgery or alternatively allow patients to appreciate the possible improvement following surgical intervention. Hearing amplification does not halt the disease process.

It would be useful in this case to for the patient to try a hearing aid to see if that would be a good solution for her.

Continue to Quiz

Back to treatments

Back to Case Presentation

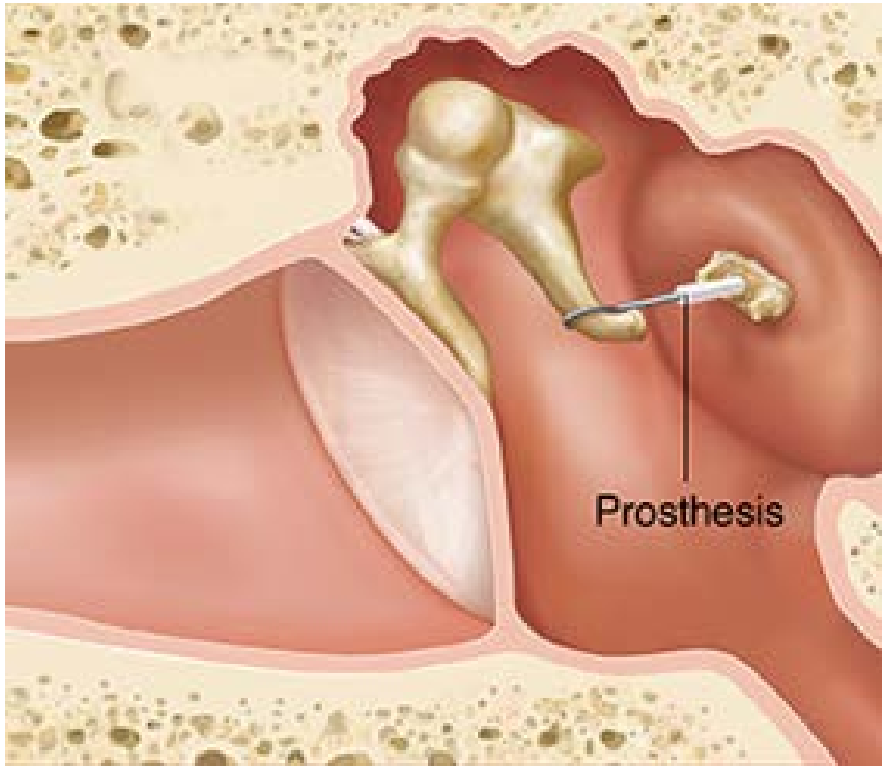
Surgery: Stapedotomy

Most patients with conductive hearing loss from otosclerosis are good surgical candidates.

Preferably, the patient would have air-bone gap of >20dB and a good speech discrimination.

Surgery for otosclerosis has evolved from total extraction of the footplate (stapedectomy) to creating a small hole in the stapes footplate (stapedotomy). Prosthesis connecting the the incus to the inner ear allows sound vibration to be transmitted and corrects the conductive hearing loss.

Stapes Prosthesis



Continue to Quiz

Back to treatments

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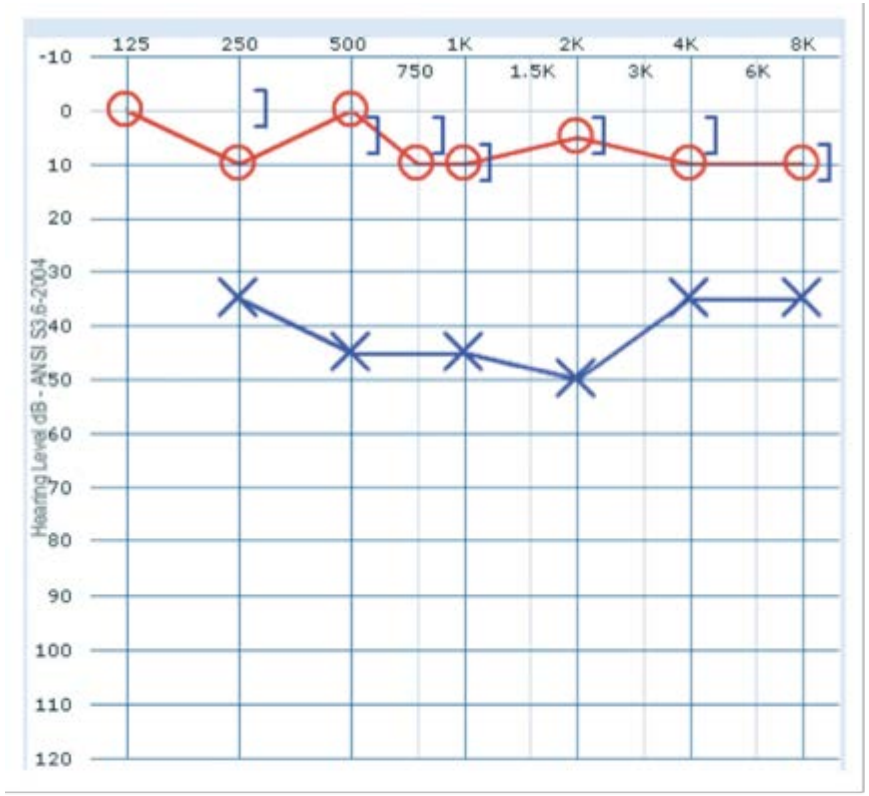
Quiz – Q1

Interpret the audiogram below:



a. Normal hearing

b. Unilateral sensorineural hearing loss

c. Asymmetrical conductive hearing loss

d. Bilateral sensorineural hearing loss

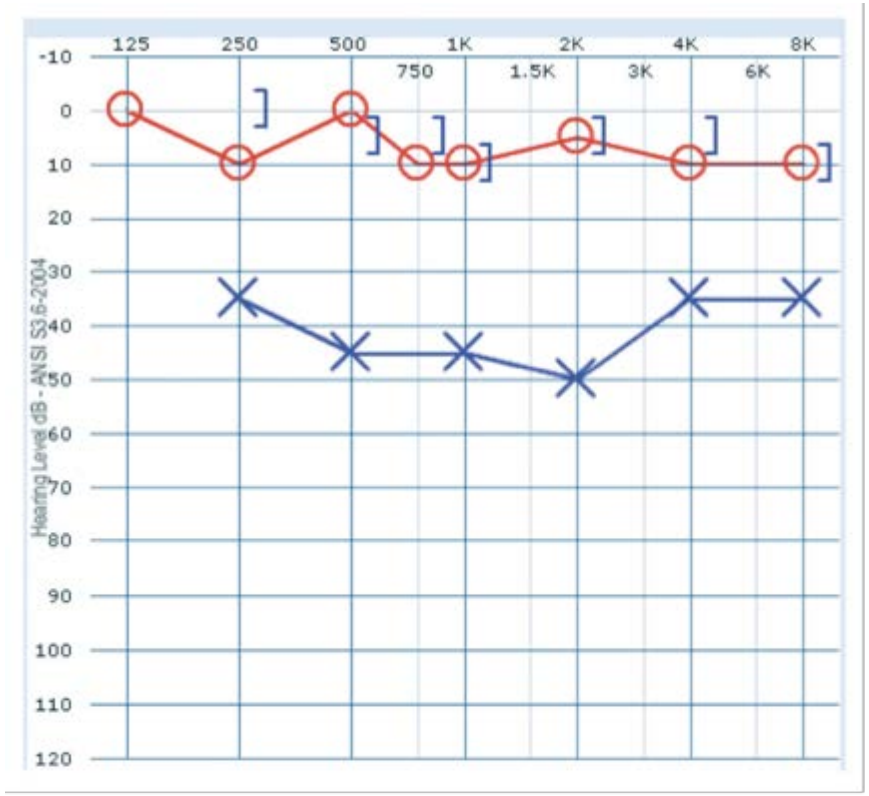
e. Bilateral conductive hearing loss

Back to Case Presentation

Try again

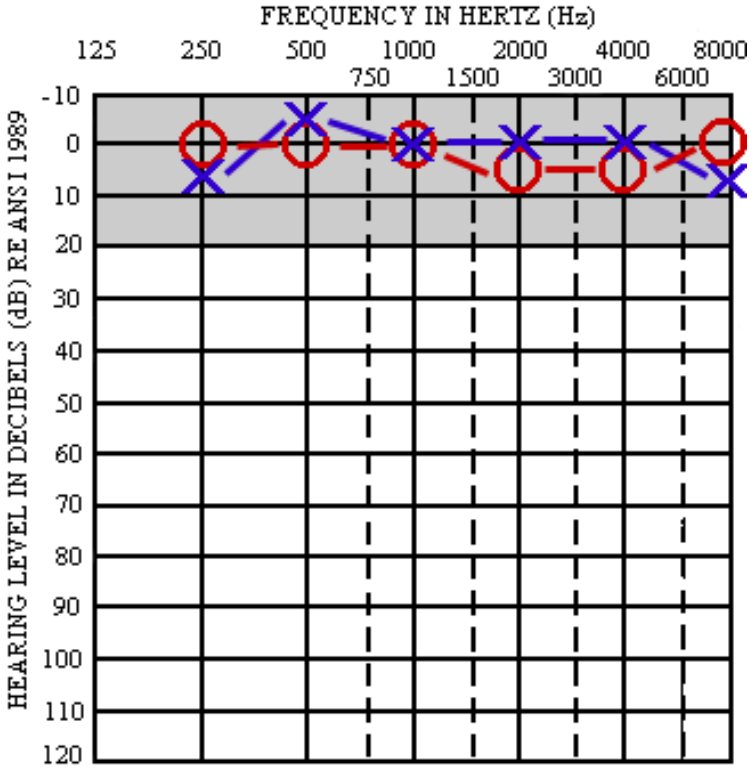
Quiz – Q1

Interpret the audiogram below:



a. Normal hearing

Incorrect. Normal audiogram:



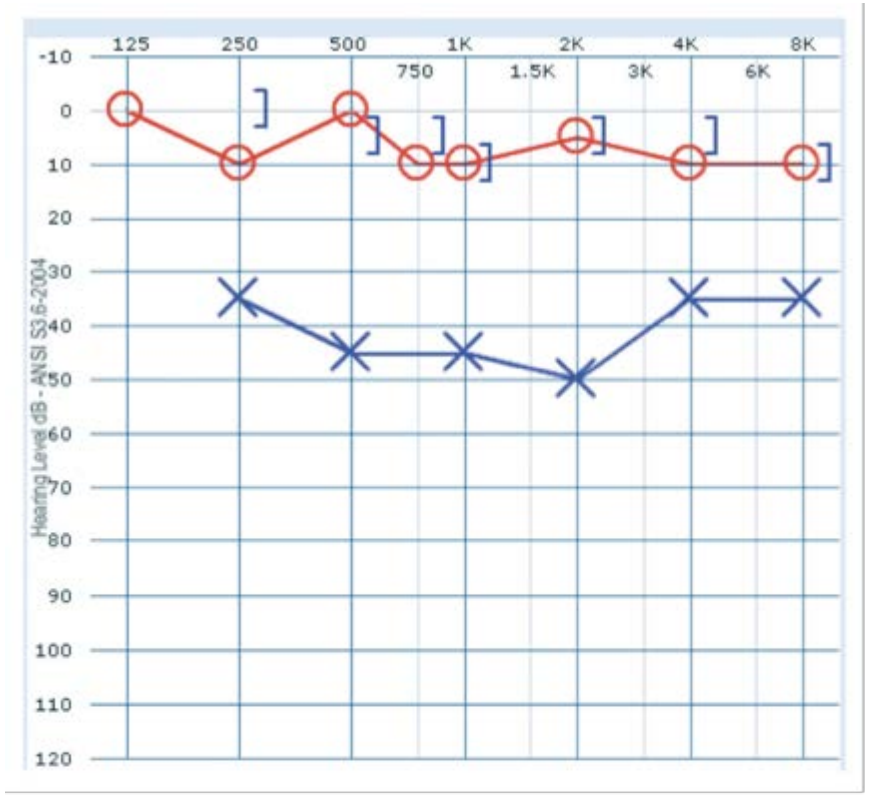
Review: Interpreting Audiograms

Back to Case Presentation

Try again

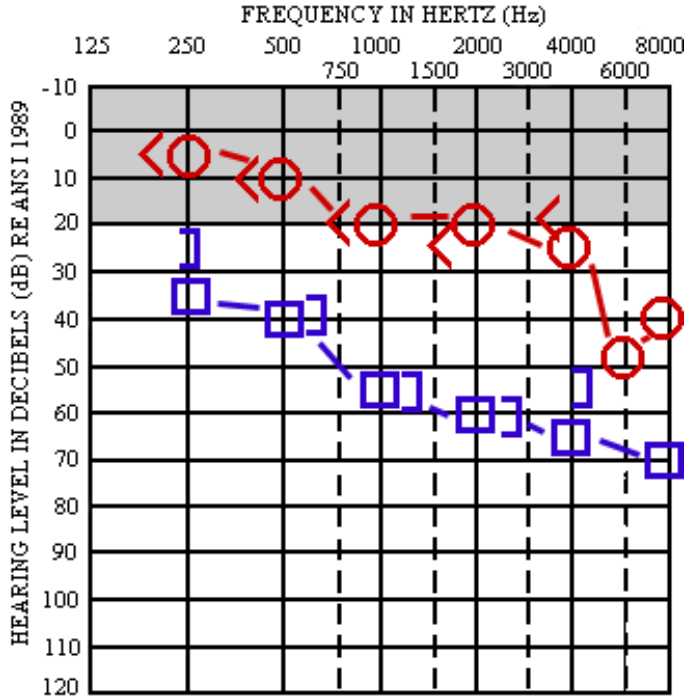
Quiz – Q1

Interpret the audiogram below:



b. Asymmetrical sensorineural hearing loss

Incorrect. Asymmetrical sensorineural hearing loss audiogram:
(*Note air conduction = bone conduction)

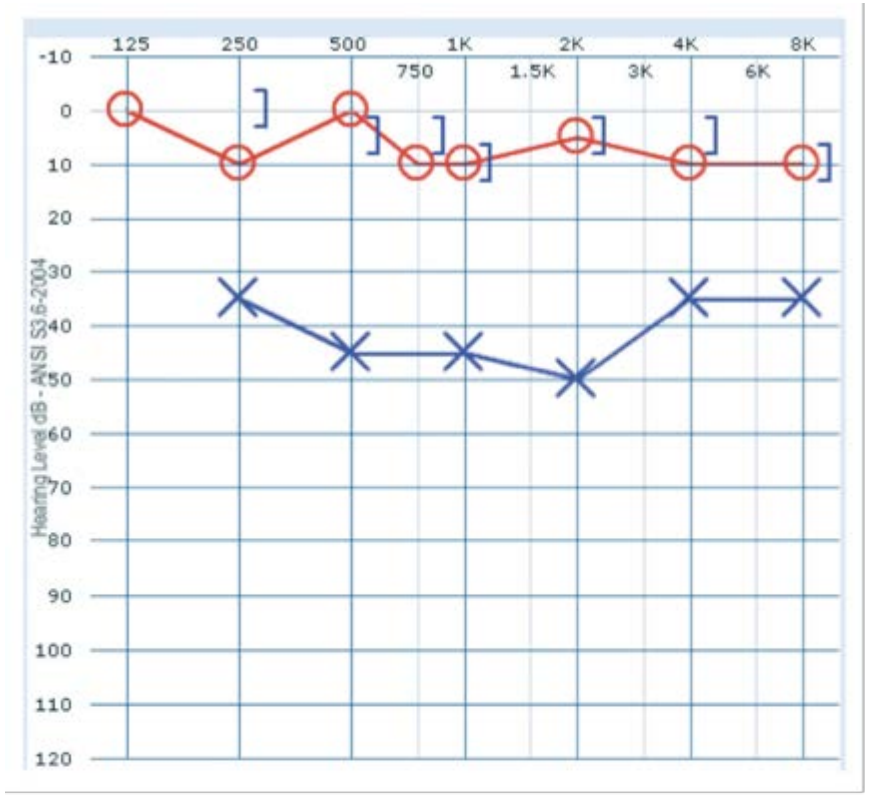


Review: Interpreting Audiograms

Back to Case Presentation

Quiz – Q1

Interpret the audiogram below:



a. Normal hearing

b. Unilateral sensorineural hearing loss

c. Unilateral conductive hearing loss

d. Bilateral sensorineural hearing loss

e. Bilateral conductive hearing loss

Correct! This patient has normal hearing in their right ear and conductive hearing loss in their left ear.

Review: Interpreting Audiograms

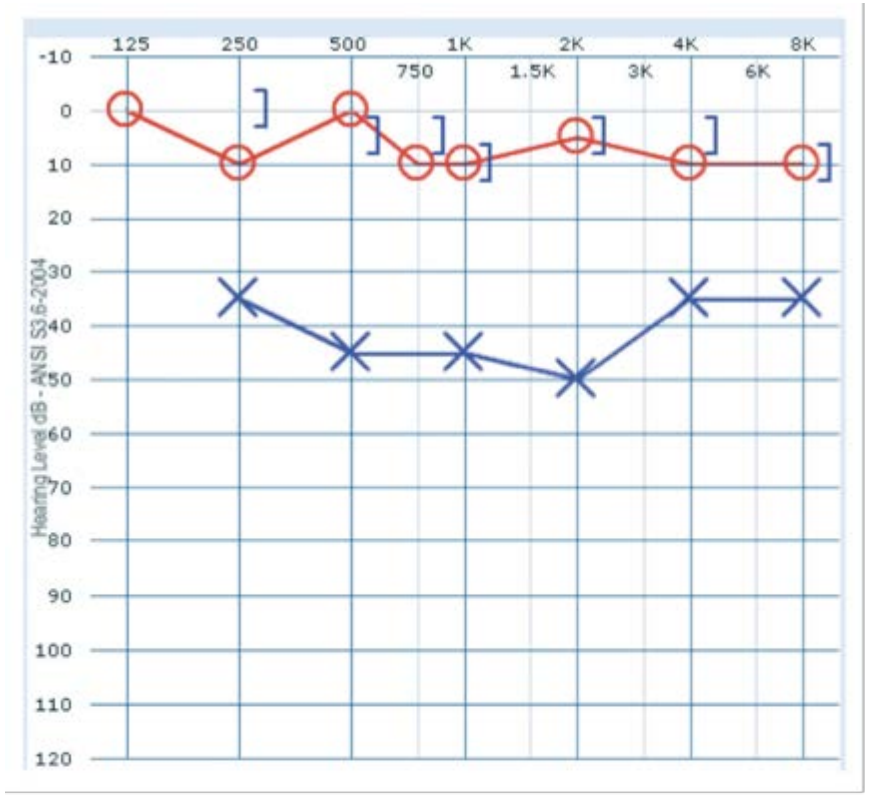
Proceed to Q2

Back to Case Presentation

Try again

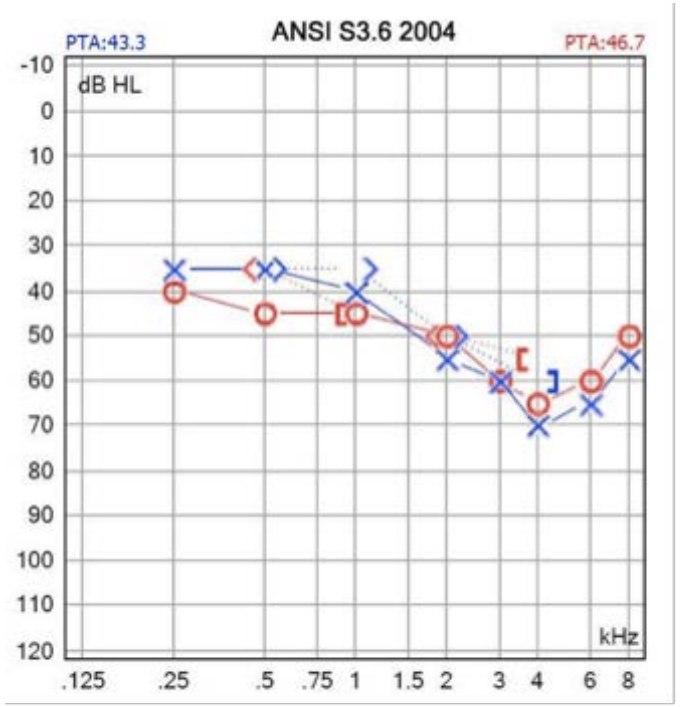
Quiz – Q1

Interpret the audiogram below:



d. Bilateral sensorineural hearing loss

Incorrect. Bilateral sensorineural hearing loss audiogram:
(*Note air conduction = bone conduction)



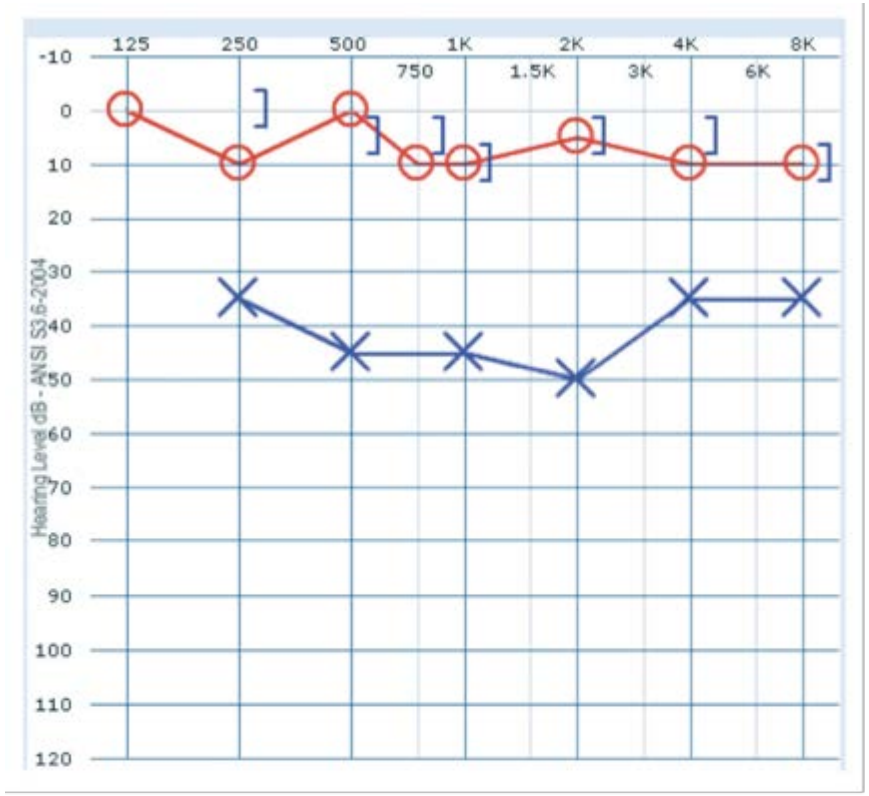
Review: Interpreting Audiograms

Back to Case Presentation

Try again

Quiz – Q1

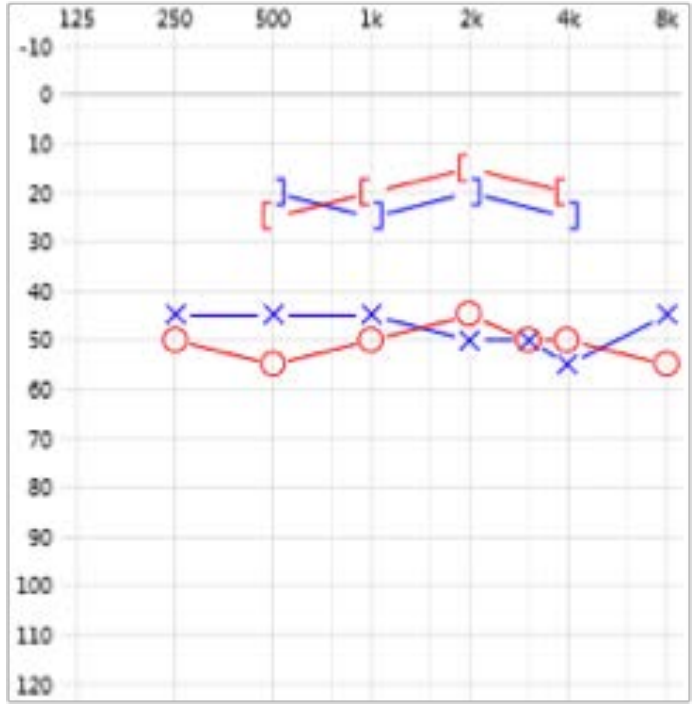
Interpret the audiogram below:



Review: Interpreting Audiograms

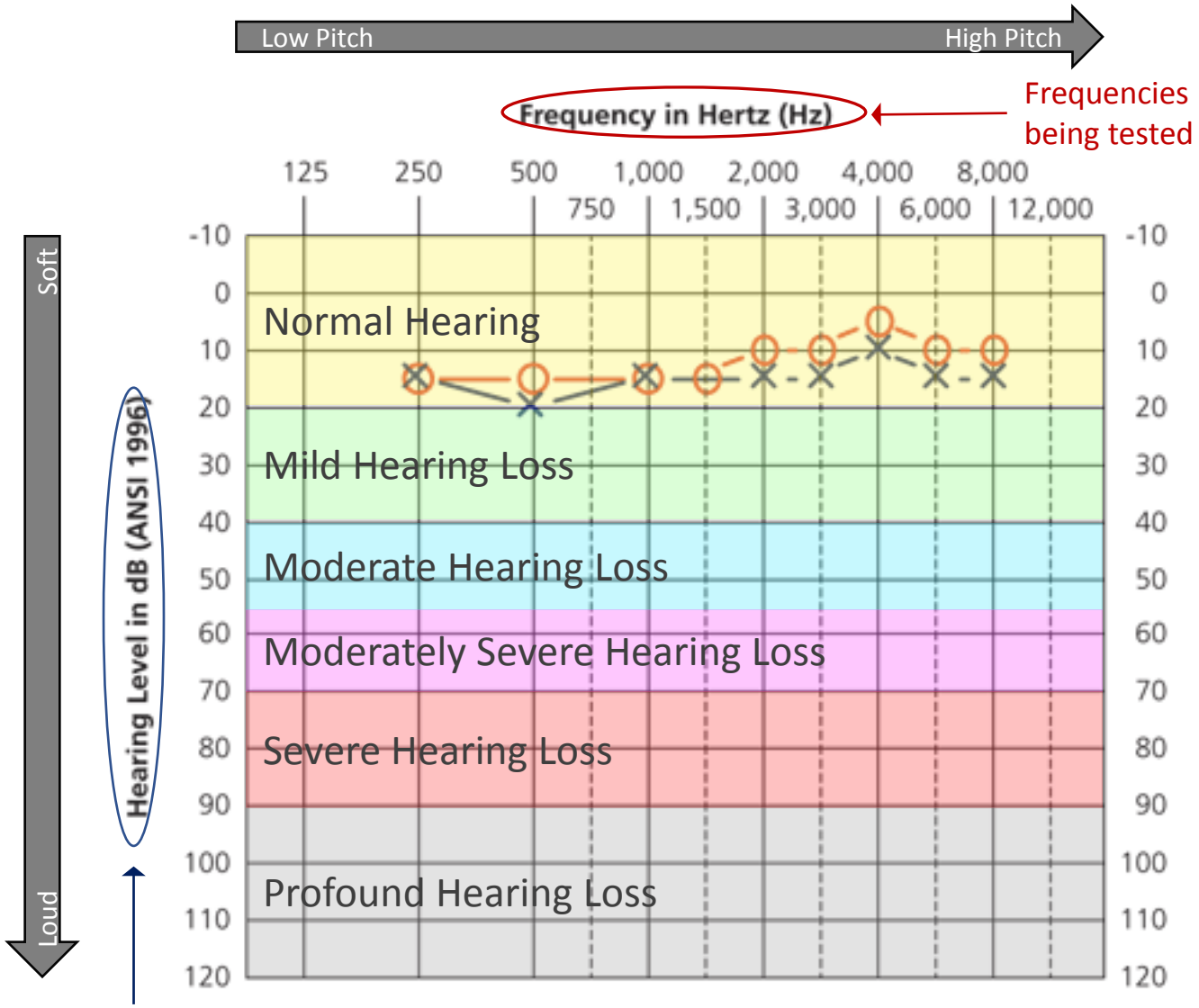
e. Bilateral conductive hearing loss

Incorrect. Bilateral conductive hearing loss audiogram:
(*Note air conduction does NOT equal bone conduction in both ears)



Review: Interpreting an Audiogram

Back to Case Presentation



How loud the sound needs to be, in order to be heard at that frequency

Air conduction testing:

- Sound delivered through headphones or loudspeakers, tests outer, middle, and inner ear.
  - Left ear = X
  - Right ear = O
- Different symbols are used when the "masking" is used. Masking refers to noise presented to the non-test ear to prevent it from hearing sound presented to the test ear.
  - Left ear = ⍰
  - Right ear = △

Bone conduction testing:

- Bone vibrator placed behind the ear to deliver sound vibrations to the cochlea, bypassing the outer and middle ear.
  - Left ear = >
  - Right ear = <
- Masking symbols
  - Left ear =]
  - Right ear = [

Click to practice reading audiograms

Back to Q1

Review: Interpreting an Audiogram

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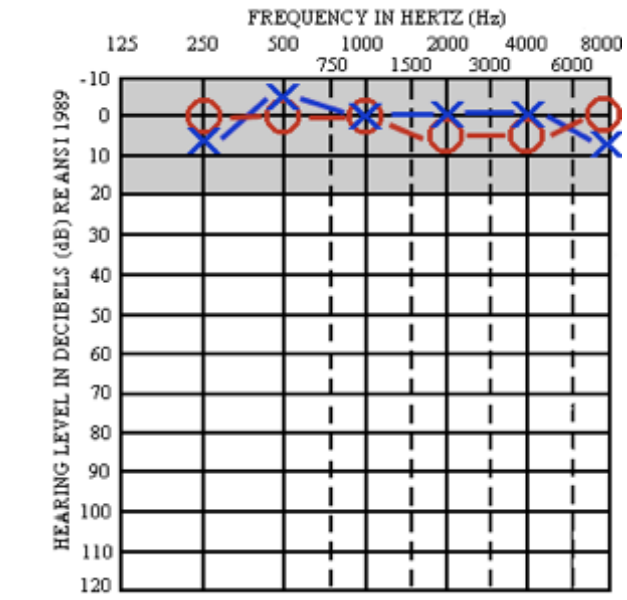
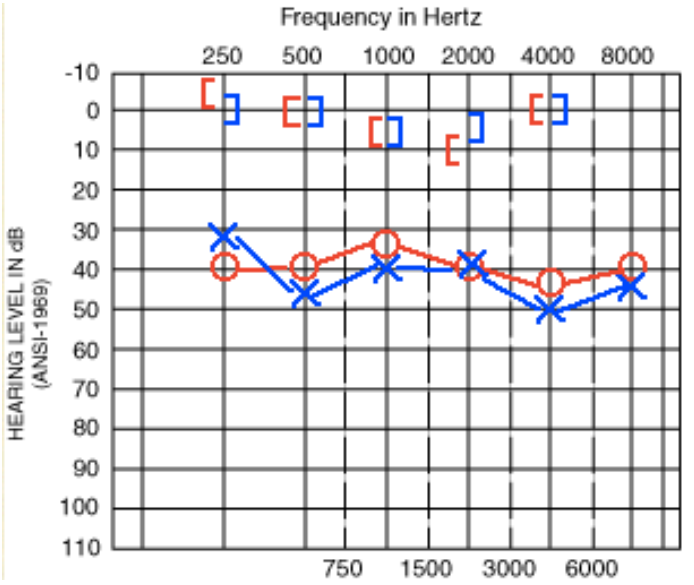
Back



Unmasked air conduction audiogram.

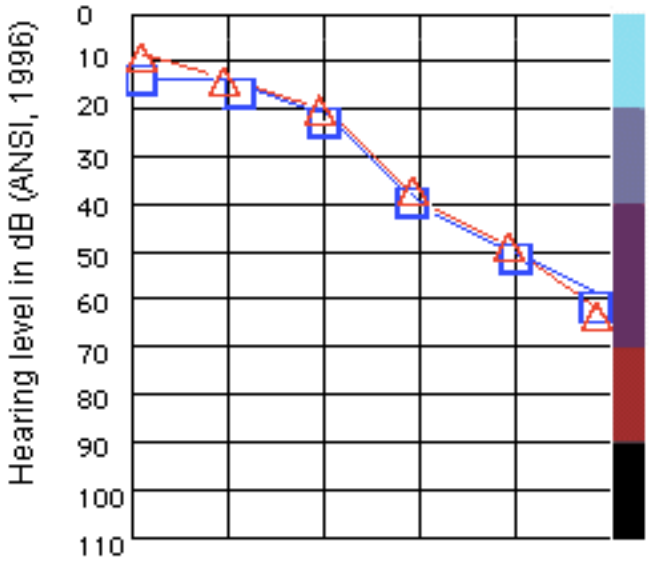
Interpretation:
Normal hearing



Air conduction testing compared to masked bone conduction testing.

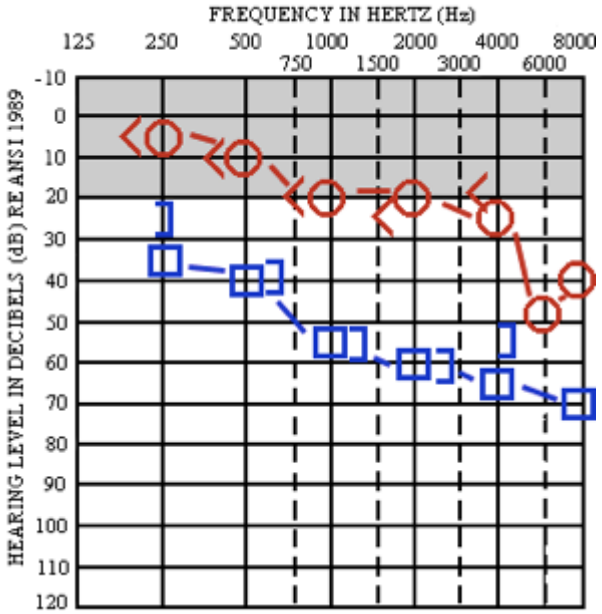
- Bone conduction: within normal range
- Air conduction: mild – moderate hearing loss

Interpretation:
Conductive hearing loss.
(I.e. Middle ear pathology)



Masked air conduction audiogram.

Interpretation: Moderately severe hearing loss at high frequencies.
(I.e. Presbycusis)



Right ear: Unmasked air and bone conduction testing
Left ear: Masked air and bone conduction testing

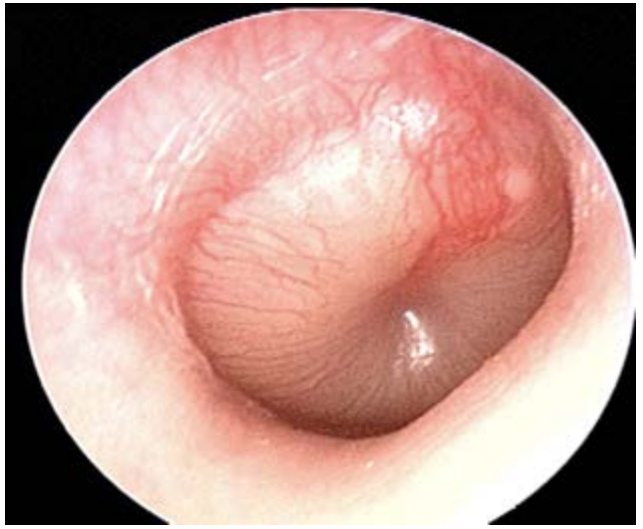
Interpretation:
Asymmetrical sensorineural hearing loss.
(I.e. Acoustic neuroma)

- Right ear: mild sensorineural hearing loss at higher frequencies
- Left ear: Mild to moderately severe hearing loss as move up frequencies

Back to Case Presentation

Quiz – Q2

Which of the following is a sign of severe otosclerosis on otoscopy?

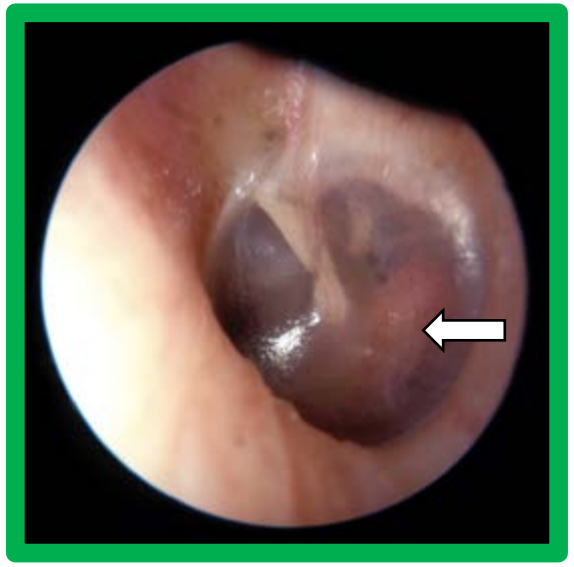


Back to Case Presentation

Quiz – Q2

Which of the following is a sign of severe otosclerosis on otoscopy?

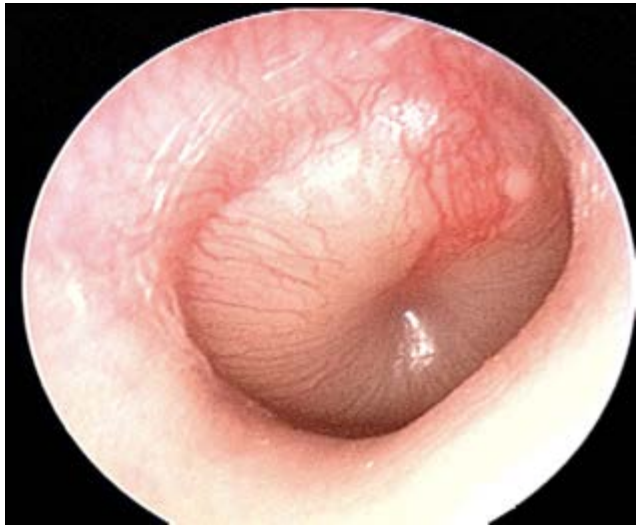
Correct!



Schwartze Sign

Myringosclerosis

Middle ear effusion



Acute Otitis Media

Schwartze sign: Increased vascularity on the promontory that is seen through the tympanic membrane. This indicates active otosclerosis. Only seen in ~10% of cases.

Proceed to Q3

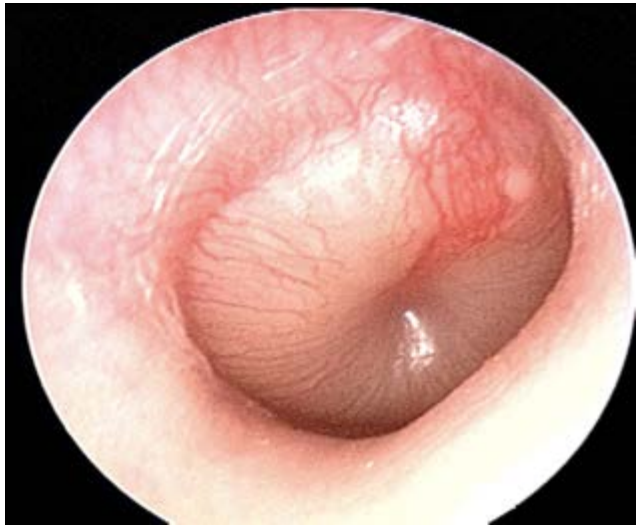
Back to Case Presentation

Quiz – Q2

Which of the following is a sign of severe otosclerosis on otoscopy?

Incorrect – Please choose again

Myringosclerosis



Try again

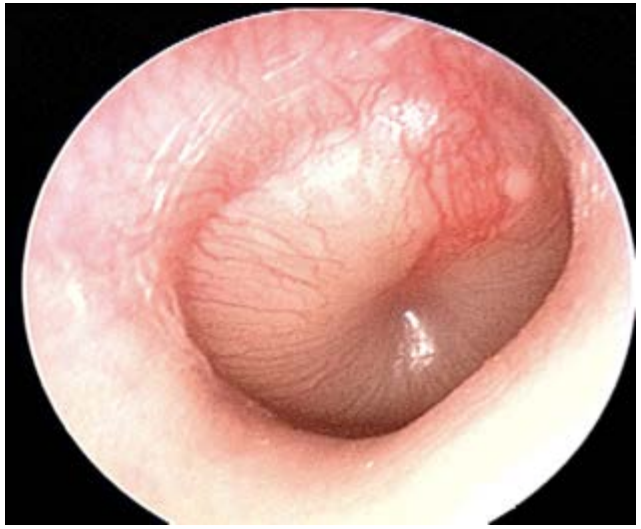
Back to Case Presentation

Quiz – Q2

Which of the following is a sign of severe otosclerosis on otoscopy?

Incorrect – Please choose again

Middle ear effusion



Try again

Back to Case Presentation

Quiz – Q2

Which of the following is a sign of severe otosclerosis on otoscopy?

Incorrect – Please choose again

Acute Otitis Media

Try again

Back to Case Presentation

Quiz – Q3

Otosclerosis can only be confirmed at the time of surgery or through histological analysis. What else is on the differential diagnosis for progressive conductive hearing loss?

a. Osteogenesis Imperfecta
b. Presbycusis
c. Tympanosclerosis
d. Middle ear effusion
e. Meniere's Disease
f. a, c, & d
g. b & e

Back to Case Presentation

Quiz – Q3

Otosclerosis can only be confirmed at the time of surgery or through histological analysis. What else is on the differential diagnosis for progressive conductive hearing loss?

a. Osteogenesis Imperfecta
b. Presbycusis
c. Tympanosclerosis
d. Middle ear effusion
e. Meniere's Disease
f. a, c, & d
g. b & e

Incorrect. Osteogenesis Imperfecta is on the DDx for otosclerosis, however it is not the only correct answer. Please choose again.

Try again

Back to Case Presentation

Quiz – Q3

Otosclerosis can only be confirmed at the time of surgery or through histological analysis. What else is on the differential diagnosis for progressive conductive hearing loss?

a. Osteogenesis Imperfecta
b. Presbycusis
c. Tympanosclerosis
d. Middle ear effusion
e. Meniere's Disease
f. a, c, & d
g. b & e

Incorrect. Presbycusis is characterized by a sensorineural hearing loss. Please choose again.

Try again

Back to Case Presentation

Quiz – Q3

Otosclerosis can only be confirmed at the time of surgery or through histological analysis. What else is on the differential diagnosis for progressive conductive hearing loss?

a. Osteogenesis Imperfecta
b. Presbycusis
c. Tympanosclerosis
d. Middle ear effusion
e. Meniere's Disease
f. a, c, & d
g. b & e

Incorrect. Tympanosclerosis is on the DDx for otosclerosis, but it is not the only correct answer.

When diagnosing progressive conductive hearing loss, tympanosclerosis can typically be ruled in or out through findings on otoscopy (see right).

Please choose again.

Try again

Back to Case Presentation

Quiz – Q3

Otosclerosis can only be confirmed at the time of surgery or through histological analysis. What else is on the differential diagnosis for progressive conductive hearing loss?

a. Osteogenesis Imperfecta
b. Presbycusis
c. Tympanosclerosis
d. Middle ear effusion
e. Meniere's Disease
f. a, c, & d
g. b & e

Incorrect. Middle ear effusion is on the DDx for otosclerosis, however it is not the only correct answer.

When diagnosing progressive conductive hearing loss, middle ear effusion can typically be diagnosed through otoscopy. On inspection of the tympanic membrane (TM), fluid or air-fluid levels behind the TM (see right).

Please choose again.

Try again

Back to Case Presentation

Quiz – Q3

Otosclerosis can only be confirmed at the time of surgery or through histological analysis. What else is on the differential diagnosis for progressive conductive hearing loss?

a. Osteogenesis Imperfecta
b. Presbycusis
c. Tympanosclerosis
d. Middle ear effusion
e. Meniere's Disease
f. a, c, & d
g. b & e

Incorrect. Meniere's Disease is characterized by episodic vertigo, sensorineural hearing loss, and tinnitus or aural fullness in the affected ear. Please choose again.

Try again

Back to Case Presentation

Quiz – Q3

Otosclerosis can only be confirmed at the time of surgery or through histological analysis. What else is on the differential diagnosis for progressive conductive hearing loss?

a. Osteogenesis Imperfecta
b. Presbycusis
c. Tympanosclerosis
d. Middle ear effusion
e. Meniere's Disease
f. a, c, & d
g. b & e

Correct! Osteogenesis imperfecta, tympanosclerosis, and middle ear effusion are on the DDx for otosclerosis.

Pathologies that impede sound transmission through the middle ear are on the DDx for otosclerosis. Other conditions on the DDx include:

- Chronic otitis media, with or without cholesteatoma
- Trauma
- Neoplasms of the middle ear or external auditory canal

Proceed to Q4

Back to Case Presentation

Quiz – Q3

Otosclerosis can only be confirmed at the time of surgery or through histological analysis. What else is on the differential diagnosis for progressive conductive hearing loss?

a. Osteogenesis Imperfecta
b. Presbycusis
c. Tympanosclerosis
d. Middle ear effusion
e. Meniere's Disease
f. a, c, & d
g. b & e

Incorrect. Presbycusis and Meniere's Disease are associated with sensorineural hearing loss. Please choose again.

Try again

Back to Case Presentation

Quiz – Q4

What do you expect to find on the Weber and Rinne tests in someone with left-sided otosclerosis (air bone gap >25dB)?

a. Weber: Lateralizes to left ear; Rinne both ears: AC > BC
b. Weber: No lateralization; Rinne both ears: AC > BC
c. Weber: No lateralization; Rinne both ears: BC > AC
d. Weber: Lateralizes to left ear; Rinne left ear: BC > AC

Back to Case Presentation

Quiz – Q4

What do you expect to find on the Weber and Rinne tests in someone with left-sided otosclerosis (air bone gap >25dB)?

a. Weber: Lateralizes to left ear; Rinne both ears: AC > BC
b. Weber: No lateralization; Rinne both ears: AC > BC
c. Weber: No lateralization; Rinne both ears: BC > AC
d. Weber: Lateralizes to left ear; Rinne left ear: BC > AC

Incorrect. This pattern is characteristic of sensorineural hearing loss in the right ear. With the presenting air-bone gap, one would expect the Rinne test to be negative

Review: Interpreting Rinne and Weber tests.

Try again

Back to Case Presentation

Quiz – Q4

What do you expect to find on the Weber and Rinne tests in someone with left-sided otosclerosis (air bone gap >25dB)?

a. Weber: Lateralizes to left ear; Rinne both ears: AC > BC
b. Weber: No lateralization; Rinne both ears: AC > BC
c. Weber: No lateralization; Rinne both ears: BC > AC
d. Weber: Lateralizes to left ear; Rinne left ear: BC > AC

Incorrect. This pattern is characteristic of normal hearing.

Review: Interpreting Rinne and Weber tests.

Try again

Back to Case Presentation

Quiz – Q4

What do you expect to find on the Weber and Rinne tests in someone with left-sided otosclerosis (air bone gap >25dB)?

a. Weber: Lateralizes to left ear; Rinne both ears: AC > BC
b. Weber: No lateralization; Rinne both ears: AC > BC
c. Weber: No lateralization; Rinne both ears: BC > AC
d. Weber: Lateralizes to left ear; Rinne left ear: BC > AC

Incorrect. This pattern is characteristic of conductive hearing loss in the both ears.

Review: Interpreting Rinne and Weber tests.

Try again

Back to Case Presentation

Quiz – Q4

What do you expect to find on the Weber and Rinne tests in someone with left-sided otosclerosis (air bone gap >25dB)?

a. Weber: Lateralizes to left ear; Rinne both ears: AC > BC
b. Weber: No lateralization; Rinne both ears: AC > BC
c. Weber: No lateralization; Rinne both ears: BC > AC
d. Weber: Lateralizes to left ear; Rinne left ear: BC > AC

Correct! This pattern is characteristic of left-sided conductive hearing loss.

Review: Interpreting Rinne and Weber tests.

Continue

Back to Case Presentation

Review: Weber and Rinne tests

1. Weber test

- Strike a 512 Hz tuning fork and place on top of the patient's head
- A patient with normal hearing should hear the sound equally on both sides* (I.e. the sound shouldn't lateralize to one ear)
    - *Note: A Rinne test is needed to confirm normal hearing, as a patient with bilateral conductive hearing loss would also have no lateralization of sound.

2. Rinne test

- Strike a 512 Hz tuning fork and place it on the mastoid bone behind the patient's ear (Testing bone conduction (BC))
- When the patient signals that they can no longer hear the sound, move the tuning fork next to the patients external auditory canal (Testing air conduction (AC))
- A patient with normal hearing should hear the sound better through air conduction (AC > BC)
- Rinne test might not be negative if the conductive hearing loss is very mild

Results from the Weber and Rinne test can be used to determine the type of hearing loss:

Test	**Normal**	**Conductive Hearing Loss**	**Sensorineural Hearing Loss**
Weber	Sound heard in midline	Sound heard in affected ear	Sound heard in good ear
Rinne	AC > BC	BC > AC	AC > BC

Back to Q4

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Review: Weber and Rinne tests

1. Weber test

- Strike a 512 Hz tuning fork and place on top of the patient's head
- A patient with normal hearing should hear the sound equally on both sides* (I.e. the sound shouldn't lateralize to one ear)
  - *Note: A Rinne test is needed to confirm normal hearing, as a patient with bilateral conductive hearing loss would also have no lateralization of sound.

2. Rinne test

- Strike a 512 Hz tuning fork and place it on the mastoid bone behind the patient's ear (Testing bone conduction (BC))
- When the patient signals that they can no longer hear the sound, move the tuning fork next to the patients external auditory canal (Testing air conduction (AC))
- A patient with normal hearing should hear the sound better through air conduction (AC > BC)
- Rinne test might not be negative if the conductive hearing loss is very mild

Results from the Weber and Rinne test can be used to determine the type of hearing loss:

Test	Normal	Conductive Hearing Loss	Sensorineural Hearing Loss
Weber	Sound heard in midline	Sound heard in affected ear	Sound heard in good ear
Rinne	AC > BC	BC > AC	AC > BC

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Congratulations! You have finished the hearing loss module.

Key points to remember:

- Otosclerosis is an **autosomal dominant** condition with incomplete penetrance.

- Typically presents in the **2nd to 4th decades** of life
  - Unilateral or bilateral progressive conductive hearing loss
  - May experience tinnitus
  - Rare to experience vertigo
  - Hearing loss may be exacerbated by pregnancy

- Diagnosis
  - History
  - Audiometry: **conductive hearing loss**
  - Otoscopy: typically normal
    - Active cases: Hyperaemia of cochlear promontory (Schwartze sign) – 10% of patients
  - **Temporal bone CT scan**: Hypodense demineralized plaques at fissula ante fenestram
    - *Note: CT scan is not necessary for diagnosis of otosclerosis
  - Note: definitive diagnosis can only be made at the time of surgery or through histological analysis

- **Three treatment options:**
  - Observation
  - Hearing aid
  - Surgery

Schwartze Sign

Continue or return to the **Review** sections.

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Module Review Sections

Physical Exam:

- Cranial Nerve Exams
- Weber & Rinne tests
- Otoscopy

Investigations:

- Audiograms
- Acoustic Reflex
- Tympanometry

Imaging:

- Temporal bone CT scan

Continue

Review: Cranial Nerve Exams

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Cranial Nerve	Function	Test
CNI – Olfactory Nerve	• Smell	• Test for sense of smell with coffee, alcohol swab, citrus, etc.
CN II – Optic Nerve	• Vision	• Visual acuity – Snellen's eye chart • Visual fields – Confrontation testing • Pupillary reflexes – Direct and consensual response • Fundoscopy
CN III – Oculomotor Nerve	• Motor innervation to most* extra-ocular muscles	• H test & convergence • Pupillary reflex
CN IV – Trochlear Nerve	• Motor innervation to superior oblique muscle*	• H test : Look for ability to look "down and out"
CN V – Trigeminal Nerve	• Sensory innervation to the face • Motor innervation to muscles of mastication	• Corneal reflex • Test sensory supply to the face – cotton swab • Test strength of muscles of mastication
CN VI – Abducens Nerve	• Motor innervation to lateral rectus muscle*	• H test: Look for ability to abduct eye
CN VII - Facial Nerve	• Motor innervation to muscles of facial expression • Taste – anterior 2/3 tongue	• Ask patient to do different facial expressions • Corneal reflex
CN VIII – Vestibulocochlear Nerve	• Hearing & balance	• Weber & Rinne tests
CN IX – Glossopharyngeal Nerve	• Sensory innervation to the palate • Taste – posterior 1/3 tongue	• Gag reflex • Say "Ahhh" – look for deviation of the uvula
CN X – Vagus Nerve	• Motor supply to the pharynx • PSNS supply to abdominal viscera	• Gag reflex
CN XI – Spinal Accessory Nerve	• Motor innervation to trapezius and sternocleidomastoid	• Shoulder shrug against resistance, head turn against resistance
CN XII – Hypoglossal Nerve	• Motor supply to muscles of the tongue	• Stick tongue out and move tongue from side-to-side

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Review: Weber and Rinne tests

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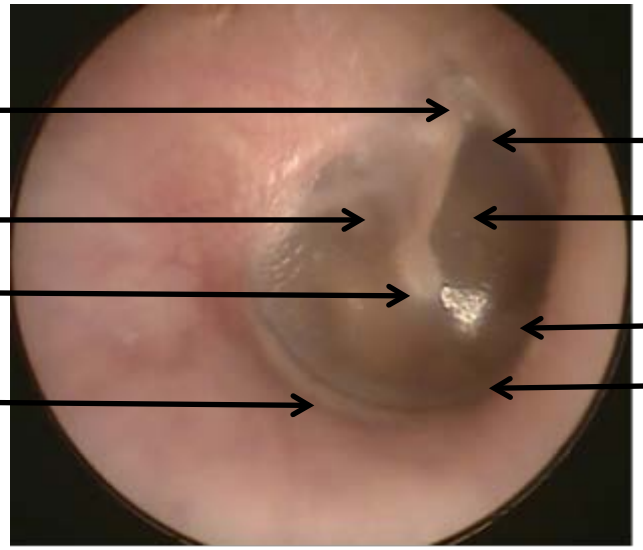
Review: Otoscopy

Normal Otoscopic Exam Findings:

- Tympanic membrane (TM) should be intact and appear pearly grey or whitish/pinkish grey

Normal TM – Right Side



Examples of Abnormal TMs

Bulging, red TM - Acute Otitis Media

Cholesteatoma

Otitis media with effusion

Myringosclerosis

Hemotympanum

Retracted TM

Review: Interpreting an Audiogram

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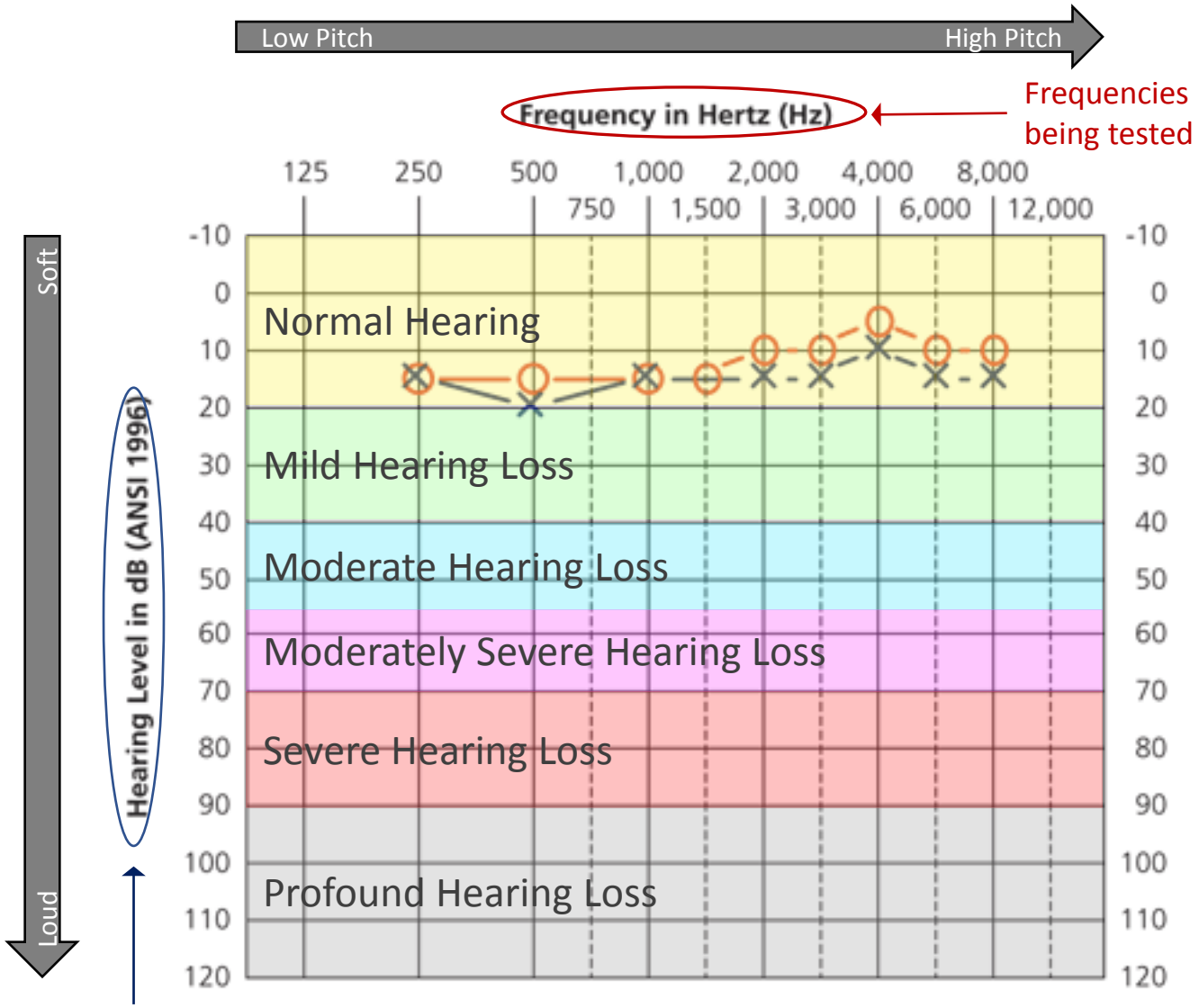

How loud the sound needs to be, in order to be heard at that frequency

Air conduction testing:

- Sound delivered through headphones or loudspeakers, tests outer, middle, and inner ear.
  - Left ear = X
  - Right ear = O
- Different symbols are used when the "masking" is used. Masking refers to noise presented to the non-test ear to prevent it from hearing sound presented to the test ear.
  - Left ear = ⍰
  - Right ear = △

Bone conduction testing:

- Bone vibrator placed behind the ear to deliver sound vibrations to the cochlea, bypassing the outer and middle ear.
  - Left ear = >
  - Right ear = <
- Masking symbols
  - Left ear =]
  - Right ear = [

Click to practice reading audiograms

Review: Interpreting an Audiogram

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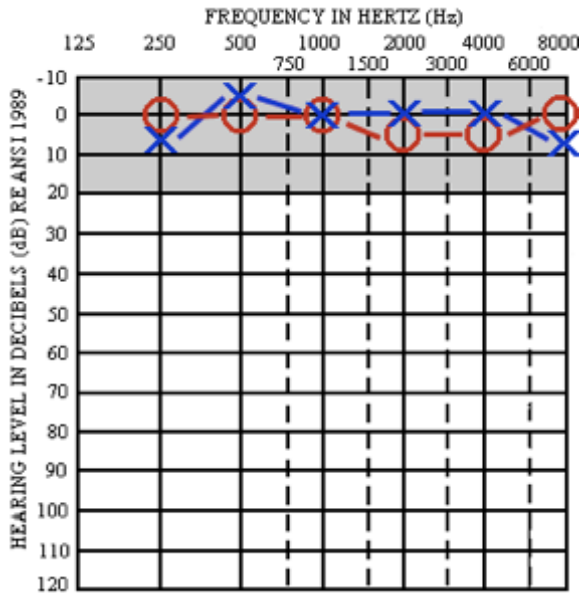
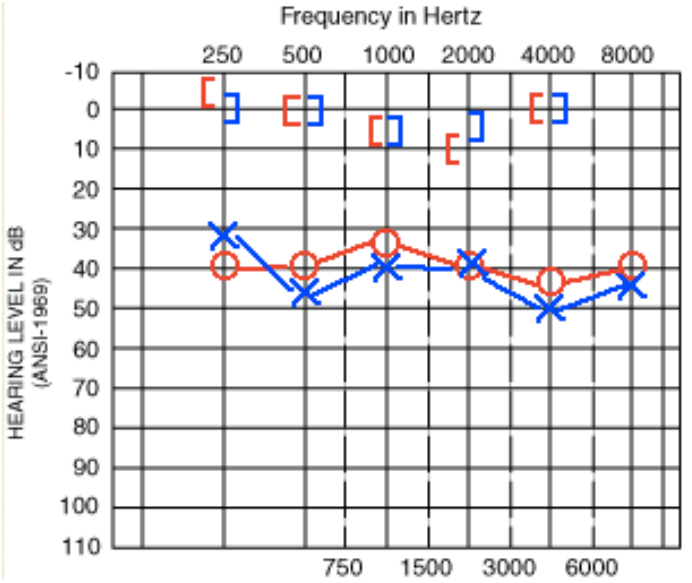


Unmasked air conduction audiogram.

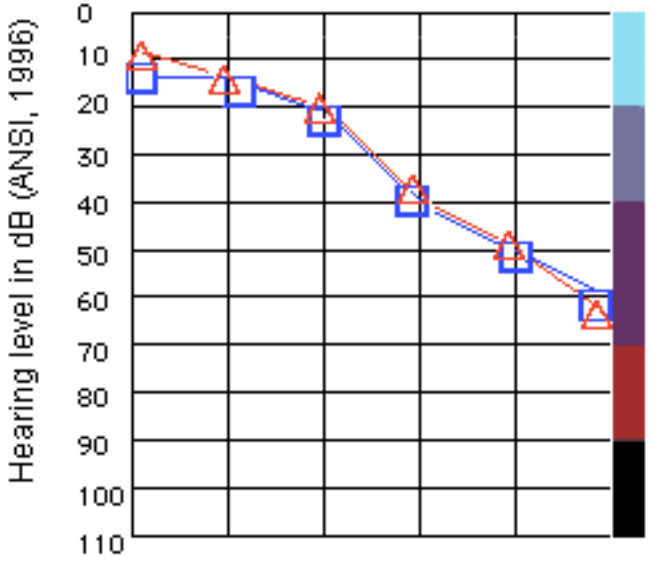
Interpretation:
Normal hearing



Air conduction testing compared to masked bone conduction testing.

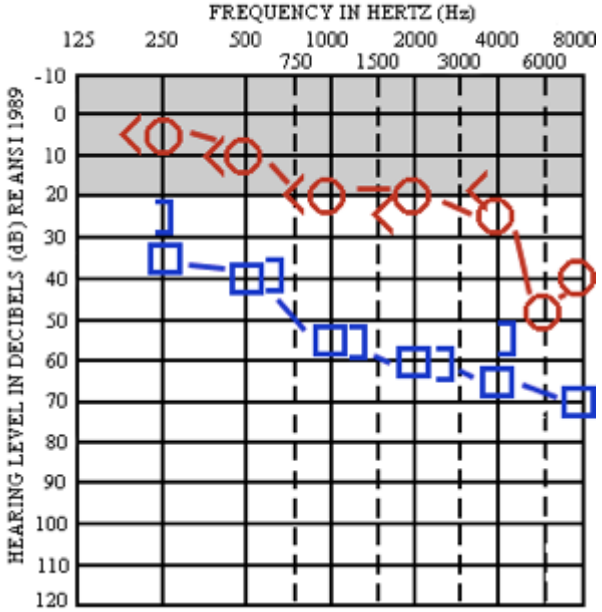
- Bone conduction: within normal range
- Air conduction: mild – moderate hearing loss

Interpretation:
Conductive hearing loss.
(I.e. Middle ear pathology)



Masked air conduction audiogram.

Interpretation: Moderately severe hearing loss at high frequencies.
(I.e. Presbycusis)



Right ear: Unmasked air and bone conduction testing
Left ear: Masked air and bone conduction testing

Interpretation:
Asymmetrical sensorineural hearing loss.
(I.e. Acoustic neuroma)

- Right ear: mild sensorineural hearing loss at higher frequencies
- Left ear: Mild to moderately severe hearing loss as move up frequencies

Review: Acoustic Reflex Testing

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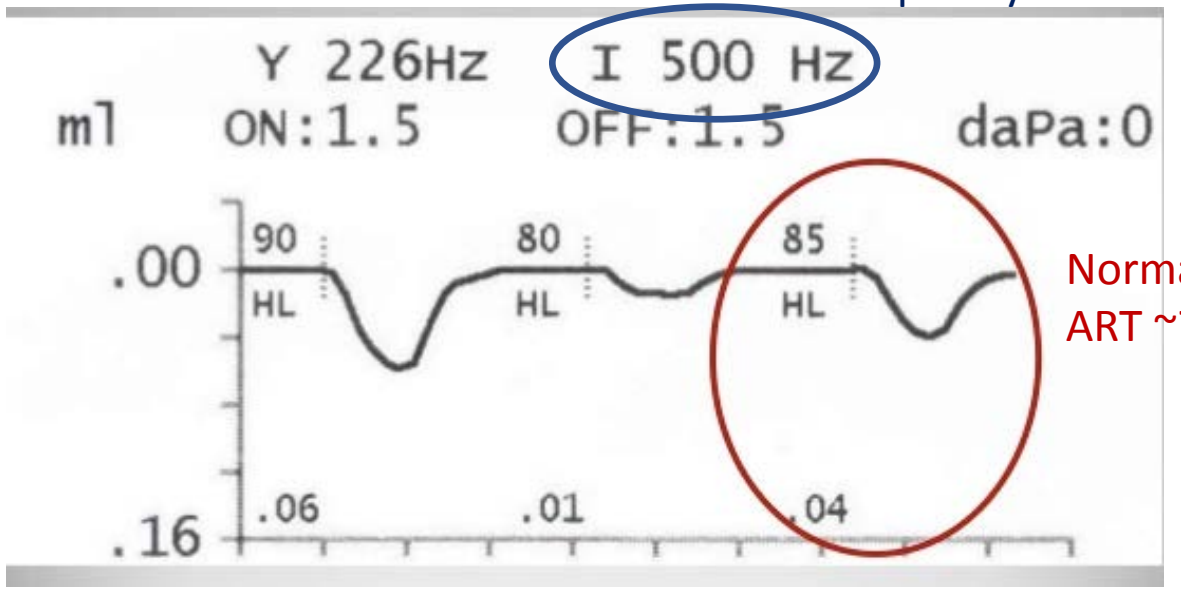
The acoustic reflex is the reflexive contraction of the stapedius muscle, and subsequent stiffening of the tympanic membrane (TM), in response to high-intensity sound or vocalization. Anatomy reminder: Stapedius is innervated by CN VII.

In acoustic (stapedial) reflex testing, acoustic signals at varying frequencies (usually 500, 1000, or 2000 Hz) are introduced into one ear and the acoustic impedance is measured in the both ears.

Acoustic Reflex Threshold (ART): Sound pressure level (SPL), in dB, from which a sound stimulus with a given frequency will elicit the acoustic reflex.

Frequency tested



Normal hearing:
ART ~70-100 dB SPL

Reflexes may be absent or harder to illicit in patients with:

- Conductive hearing loss
- Severe sensory hearing loss
- CN8 injury on side receiving sound
- CN7 injury on side being measured

Reflexes may also be absent if there is fixation of the ossicles.

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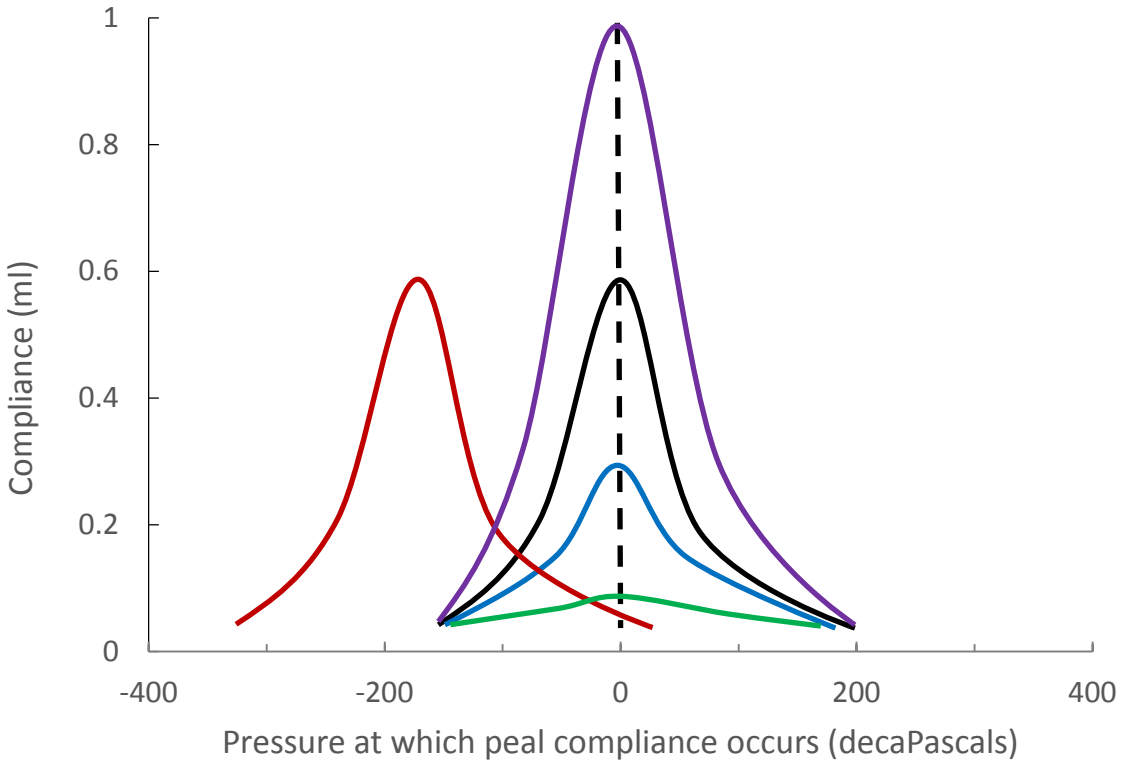
Review: Tympanometry

Tympanometry is an indirect test of middle ear function by the transmission/reflection of sound energy. A tympanogram plots compliance changes of the tympanic membrane (TM) versus air pressure in the external auditory canal.



High peak = hypercompliant TM
- I.e. Ossicular discontinuity, monomeric TM (thin TM from healed TM perforation)

Normal tympanogram

Shallow peak = stiff TM
- I.e. Otosclerosis, tympanosclerosis

No peak = non-mobile TM
- I.e. Effusion, perforation

Peak shifted to a more negative pressure = retracted TM
- I.e. Eustachian tube dysfunction, TM atelectasis

Review: Reading a Temporal Bone CT Scan

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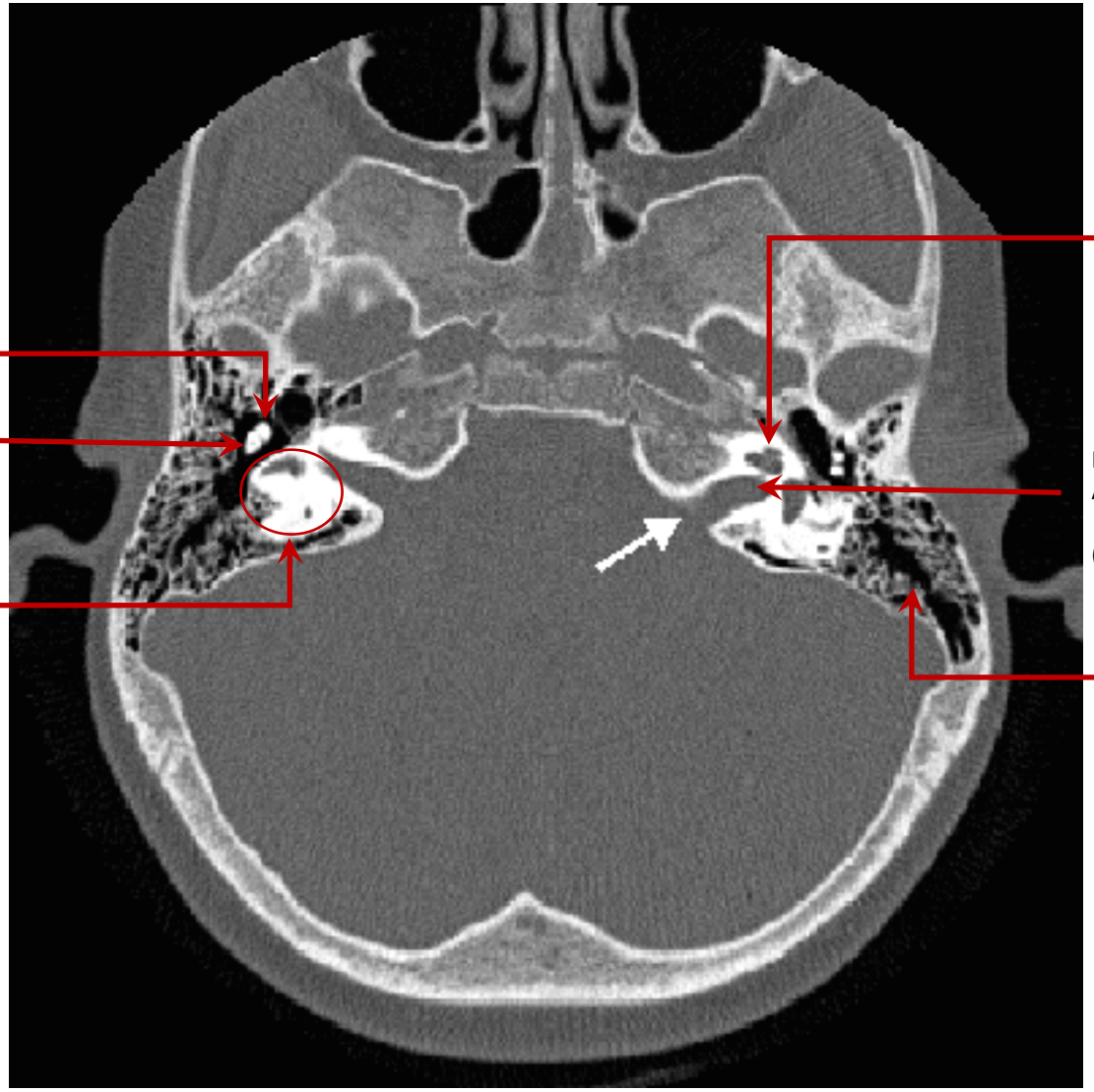
Normal temporal bone CT scan – Coronal plane



Tip: Look for the "ice cream cone"

Malleus

Incus

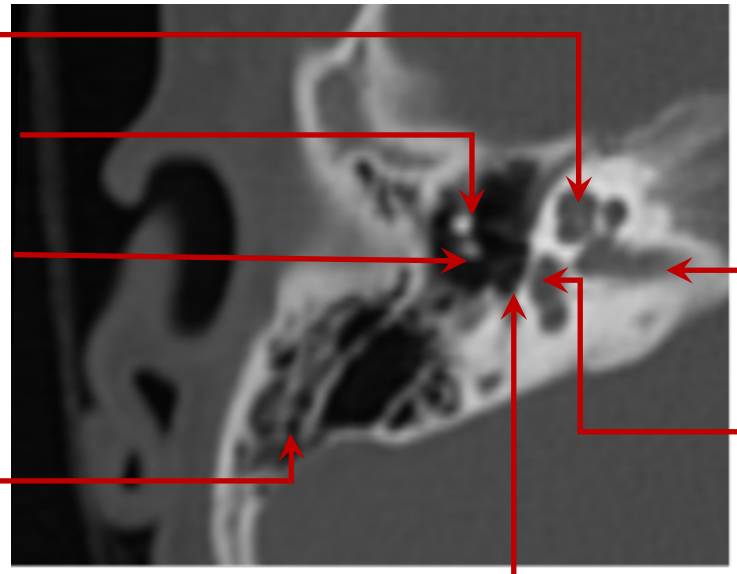
Semicircular Canal

Cochlea

Internal Auditory Canal (IAC)

Mastoid Air Cells

Magnified right temporal bone – Coronal plane



Malleus

Incus

IAC

Footplate of stapes at the oval window

Stapes

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Module Authors

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You may now exit, return to **Review** sections or retake the Quiz