I Heard it Through the Grapevine: Evaluation and Management of Congenital and Acquired Hearing Loss

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I have no financial relationships to disclose

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Learning Objectives

- Review the anatomy and physiology of hearing and where structural anomalies, injuries, and illnesses can cause hearing loss
- Describe the appropriate screening, history, physical exam, and additional testing, if indicated, required for the appropriate evaluation of hearing loss
- Recognize the symptoms and exam findings that indicate a need for urgent management of new onset hearing loss to prevent permanent changes in hearing
- Identify treatments for hearing loss including current medical, surgical, and technological options

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Question 1

What is the lifetime prevalence of hearing loss in the United States?

A. 2%

B. 21%

C. 43%

D. 61%

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Prevalence of Hearing Loss

United States

- 0.1-0.2% of newborns
- 0.2% young children
- 3% ages 21 to 34
- 6% ages 35 to 44
- 11% ages 44 to 54
- 25% ages 55 to 64
- 43% ages 65 to 84

World

- Around 466 million people worldwide have disabling hearing loss; 34 million of these are children
- It is estimated that by 2050 over 900 million people will have disabling hearing loss
- Unaddressed hearing loss poses an annual global cost of US$ 750 billion

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B. 21%

C. 43%

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Question 2

If a patient complains of muffled hearing, where would the abnormality be located? (choose all that apply)

A. External ear

B. Middle ear

C. Inner ear and/or Auditory Nerve

D. Sensation of muffled hearing unrelated to ears

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Physiology of Hearing

https://youtu.be/PeTriGTENoc

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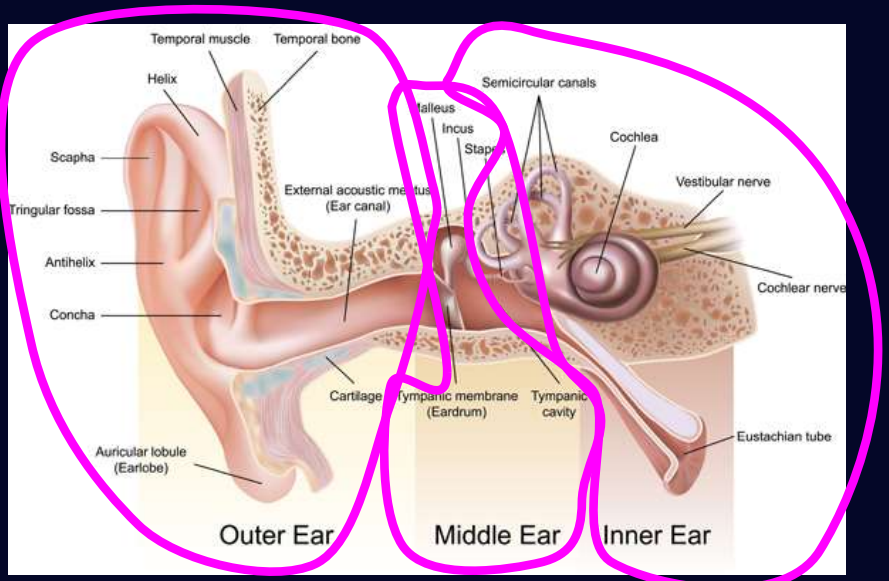
Anatomy



http://www.audiologyspecialists.com/anatomy-of-the-ear

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Other Causes of Muffled Sensation

- Temporomandibular joint dysfunction
- Migraine or other neurologic disorder
  - Concussion
  - Cognitive disorders

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Question 2

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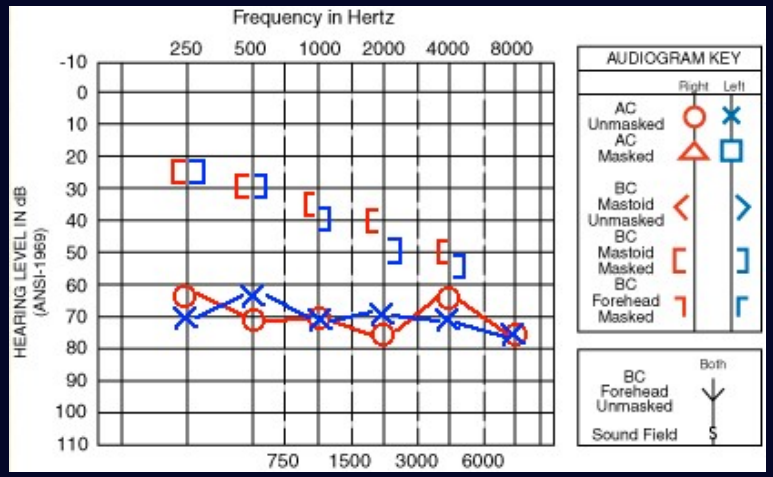
Question 3

How would you interpret this audiogram?

A. Normal hearing
B. Moderate sensory neural hearing loss
C. Severe conductive hearing loss
D. Severe mixed hearing loss



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How to Read an Audiogram

- Brackets are bone conduction, x marks and round marks are air conduction
- Normal above 20hz
- Right
  - Round marks, red ink, bracket opens to right
- Left
  - X marks, blue ink, bracket opens to left
- Word recognition score

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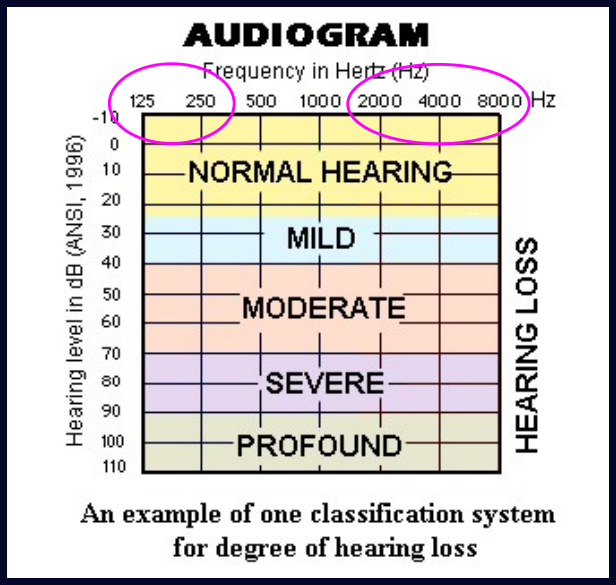
How to Read an Audiogram

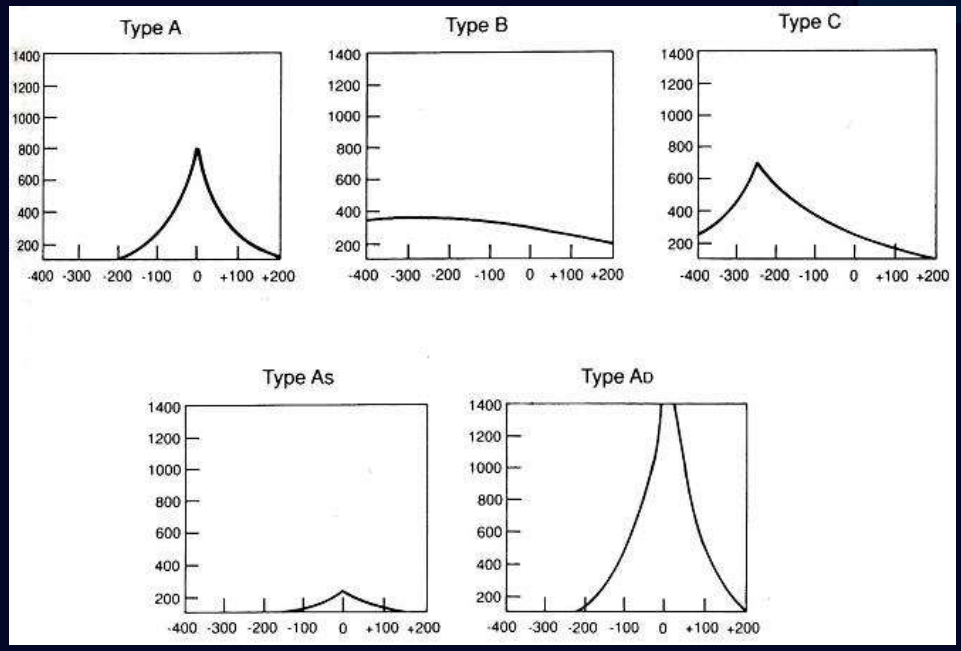
Low frequency → High frequency



An example of one classification system for degree of hearing loss

https://www.google.com/url?sa=i&rct=j&q=&esrc=s&source=images&cd=&cad=rja&uact=8&ved=0ahUKEwi-i6P68_OAhUG4yYKHQZ5ASAQjRwIBw&url=http%3A%2F%2Ffirstyears.org%2Fc3%2Fc3.htm&psig=AFQjCNGPSQ_qfEeeC8L4JBAoYUS4dkIlNQ&ust=1473428659252426

How to Read a Tympanogram



http://www.cybersight.org/data/1/rec_imgs/6208_fig.7.15.jpg

15

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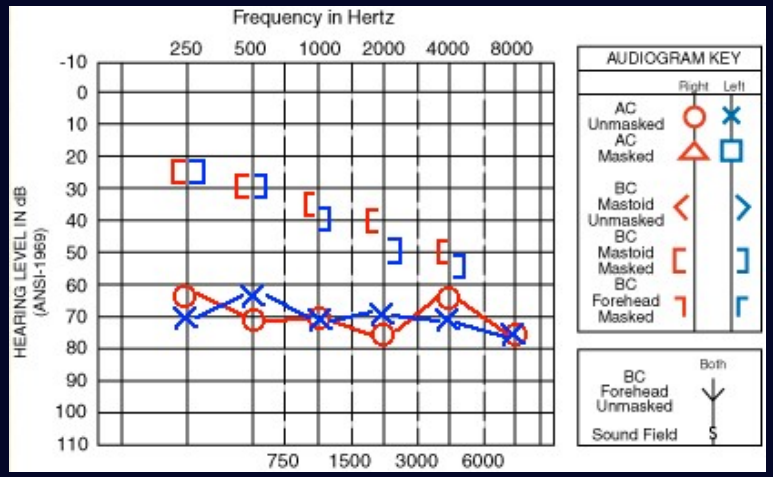

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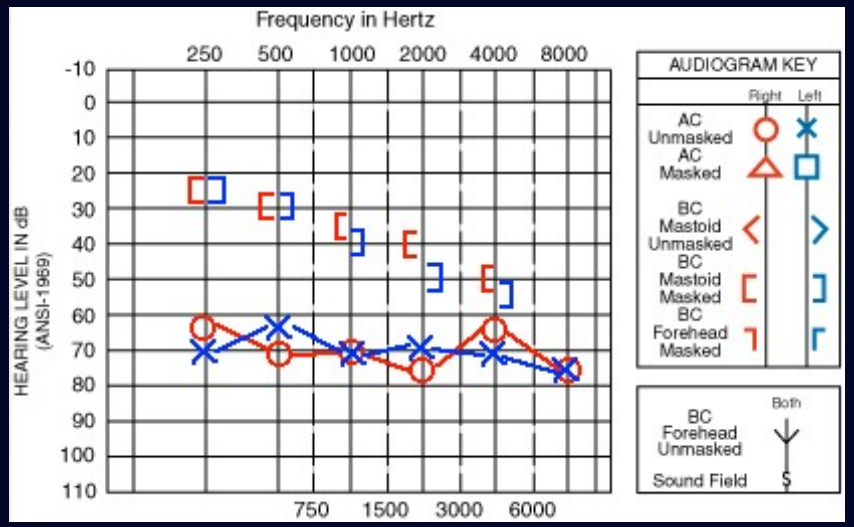
Question 4

Where could/should the abnormality in hearing be located in this audiogram?

(choose all that apply)

A. External auditory canal
B. Middle ear
C. Acoustic nerve
D. No abnormality on audiogram



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Types of Hearing Loss

- Sensory neural
  - Acoustic nerve dysfunction
- Conductive
  - Issue in the middle ear, tympanic membrane, or ear canal
- Mixed
  - Combination of sensory neural and conductive
- *Malingering

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Question 4

Where could/should the abnormality in hearing be located in this audiogram?

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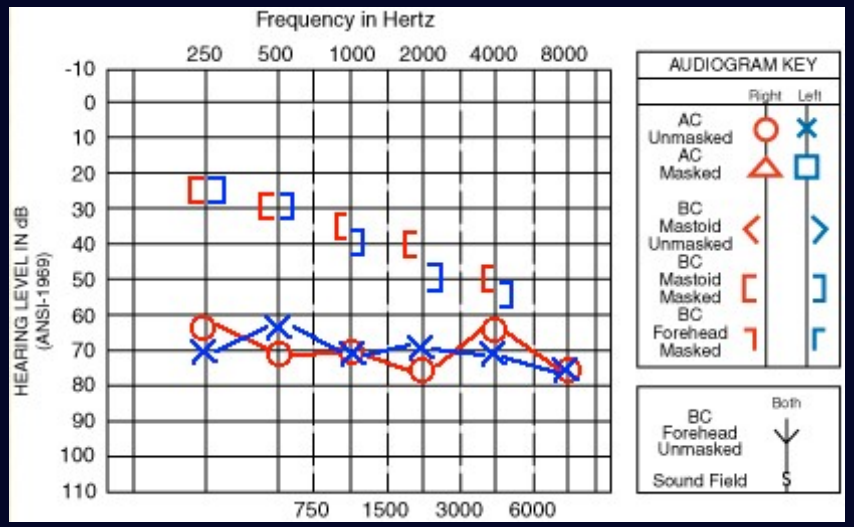

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Question 5

20 year old female complains of waking up in the morning 1 week ago with hearing loss in her right ear. What is the next appropriate step?

A. Obtain further history

B. Perform physical exam

C. Order audiogram

D. Prescribe decongestants

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History

- When did it start? Has it changed or worsened since then?
- Can you not hear people talking or can you not understand them?
- Associated symptoms
  - Pain
  - Drainage
  - Vertigo
  - Tinnitus
  - Headache, visual changes
  - Autophony
  - Tullio phenomenon
  - Jaw pain
- Any head or ear injury?
  - Including barotrauma
- History of ear infections?
- History of ear surgeries?
- Family history of hearing loss?
  - Any surgery for this hearing loss?
- Medication history
- Past Medical History
  - Diabetes
  - Smoking
  - Vascular disease
  - Autoimmune diseases

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Physical Exam

- Visual inspection
  - External ear
  - Post and pre auricular area
  - Ear canal
  - Tympanic membrane
- Palpation
  - External ear
  - Post and pre auricular area
  - TMJ
- Hearing evaluation
  - Audiogram if possible
  - Weber
    - Tuning fork in midline of head and see which side the sound lateralizes toward
  - Rinne
    - Air versus bone conduction

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Rinne and Weber

	Normal	Right CHL	Left CHL	Right SNHL	Left SNHL
Weber Equal	X				
Weber to Right		X			X
Weber to Left			X	X	
Rinne Right Air > Bone	X		X	X	X
Rinne Left Air > Bone	X	X		X	X
Rinne Right Bone > Air		X			
Rinne Left Bone > Air			X		

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Why Not Decongestants?

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Question 6

Exam of this patient's tympanic membranes are as pictured below, face is symmetric, Webber lateralizes to the left ear, Rinne shows air is greater than bone bilaterally. What is the treatment?

A. Nasal steroid spray
B. Oral steroid taper
C. Diuretic
D. Antibiotic

https://doomandbloom.net/wp-content/uploads/2012/02/ear_drum.png

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Eustachian Tube Dysfunction

- Inability of eustachian tube to equalize pressure in the middle ear
- Causes
  - Anatomic abnormalities
  - Rhinitis
  - Age related anatomical factors
  - Mass
  - Laryngeal pharyngeal reflux
  - Unknown

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Eustachian Tube Dysfunction

- Symptoms
  - Blocked ear feeling
  - Unable to “pop ears”
  - Discomfort
  - Some people have hearing change
- Exam
  - May be normal
  - Retracted tympanic membrane
  - Effusion
  - Motion with breathing*
- Audiogram
  - Flat or reduced tympanogram, otherwise normal

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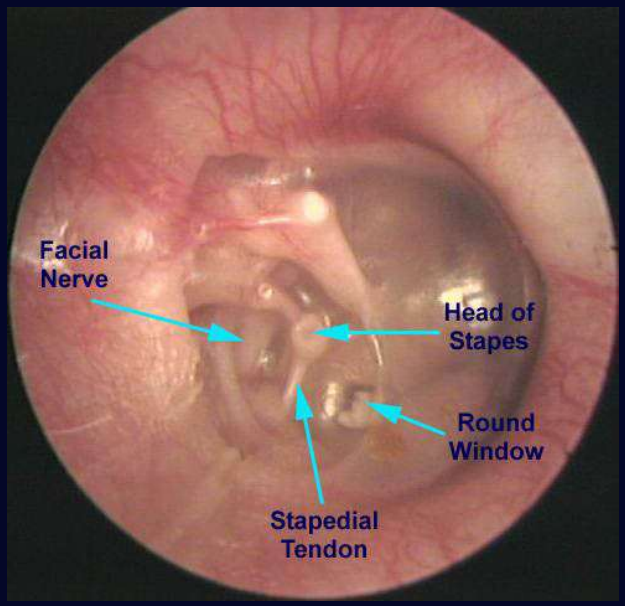
Eustachian Tube Dysfunction



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Eustachian Tube Dysfunction

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Eustachian Tube Dysfunction

- Treatment
  - Treat the underlying problem
    - Nasal steroid sprays
    - Proton pump inhibitors +/- H2 blocker for reflux
  - Myringotomy and ventilation tube placement

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*Patulous Eustachian Tube

- Eustachian tube is open when it shouldn't be
- Autophony
- May see tympanic membrane move with inspiration
- Associated with weight loss
- Very hard to treat
  - Nasal estrogen drops
  - PatulEND
  - Dampen the vibration of the TM

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Meniere's Disease

- Symptoms
  - Episodic sensory neural hearing loss in one ear
    - Accompanied by vertigo and tinnitus in that ear
  - May recover between episodes
  - Over time causes permanent sensory neural hearing loss

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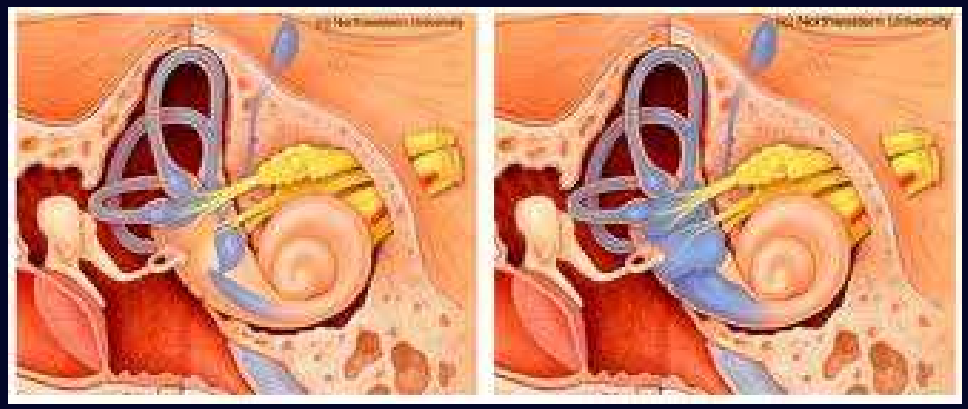
Meniere's Disease

- Probably caused by endolymph hydrops



Normal Ear

Ear with Meniere's Disease

https://dnbhelp.files.wordpress.com/2011/10/menieres-disease.jpg

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Meniere's Disease

- Exam will be normal
- Audiogram
  - Low frequency sensory neural hearing loss in one ear
- Treatment
  - Dietary changes
    - Low salt, decrease caffeine, processed foods, red wine, chocolate
  - Diuretics
  - +/- oral or intratympanic steroids for acute symptoms
  - Meclizine, phenergan, or alprazolam for acute symptoms

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Sudden Sensory Neural Hearing Loss

- Acute hearing loss
- Causes
  - Most sudden sensory neural hearing loss is idiopathic
  - Viral
  - Vascular
  - Autoimmune
  - Acoustic neuroma (schwannoma)

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Sudden Sensory Neural Hearing Loss

- Symptoms
  - Hearing loss
    - Usually unilateral
    - "I woke up and couldn't hear"
  - Tinnitus
  - If has vertigo should consider other diagnosis in differential
- Physical exam should be normal
- Audiogram
  - Sensory neural hearing loss in one ear

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Sudden Sensory Neural Hearing Loss

- Workup
  - MRI of Internal Auditory Canals **without and with gadolinium**
- Treatment
  - Steroid taper
    - At least 60mg of Prednisone for 3-5 days with taper
  - Intratympanic injection of dexamethasone
  - Antivirals?

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Exam of this patient's tympanic membranes are as pictured below, face is symmetric, Webber lateralizes to the left ear, Rinne shows air is greater than bone bilaterally. What is the treatment?

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https://doomandbloom.net/wp-content/uploads/2012/02/ear_drum.png

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Question 7

A 3-year-old girl presents with her parents with a complaint of ear infections. She has had 5 ear infections this year and 4 last year. The family feels she hears well, but her speech is delayed. Her tympanic membranes look as below.

https://i.ytimg.com/vi/jVWhOnY14uw/maxresdefault.jpg

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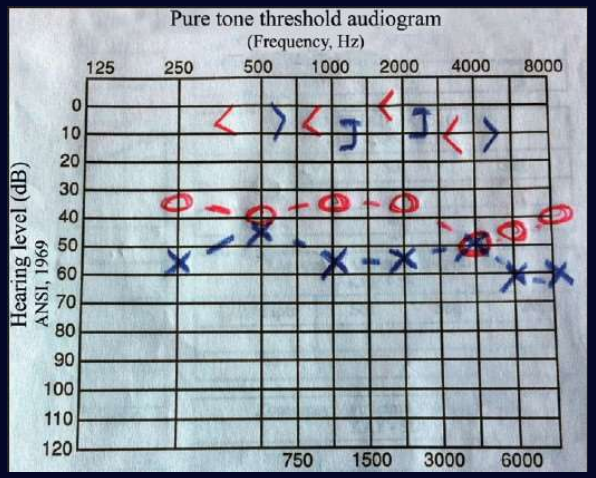
Question 7

Her audiogram as is follows



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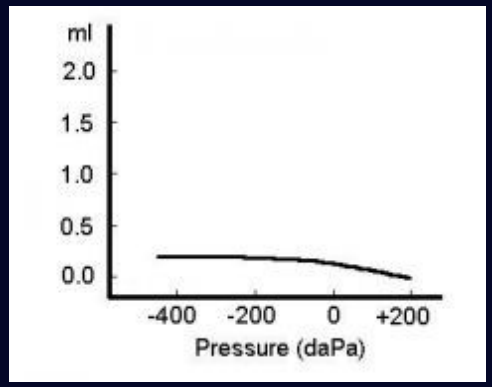

http://fce-study.netdna-ssl.com/images/upload-flashcards/back/7/2/47727781_m.jpg

40

Question 7

What is the most appropriate treatment for this patient?

A. Bilateral myringotomy and tube placement

B. Amoxicillin/clavulanic acid 90mg/kg/day divided BID for 7 days

C. Observation for 3 months

D. Hearing aids

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Indications for Myringotomy and Tube Placement in Children

- Otitis media with effusion (OME) in one or both ears lasting at least 3 months AND hearing difficulties
- Recurrent acute otitis media (AOM) with OME in at least one ear at the time of the visit
- OME of any duration or recurrent AOM in children at risk of speech, language, or learning problems

Rosenfeld, R.M. et al. (2013). Clinical practice guidelines: Tympanostomy tubes in children. *Otolaryngol Head Neck Surg.* 145(1), S1-S35.

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Acute Otitis Media Diagnosis

- Bulging of the tympanic membrane

OR

- Other signs of inflammation and OME
- Be cautious
  - If the tympanic membrane is erythematous and no other signs
  - Bubbles/air fluid level
- If you are not sure
  - Tympanogram or pneumatic otoscopy

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Acute Otitis Media Treatment

- Acetaminophen alternating with ibuprofen
- Antibiotics?

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AOM Antibiotic Recommendations

- No recent beta-lacatam therapy or no recurrent AOM
  - Amoxicillin 90mg/kg/day divided BID
- Recent beta-lactam therapy or recurrent OM
  - Amoxicillin/Clavulanate acid 90mg/kg/day divided BID

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AOM Antibiotic Recommendations

- Penicillin allergic without history of type 1 reaction, use one of the following:
  - Cefdinir 14 mg/kg per day QD or divided BID
  - Cefpodoxime 10 mg/kg per day divided BID
  - Cefuroxime 30 mg/kg per day divided BID
  - Ceftriaxone 50 mg/kg intramuscularly QD for one to three doses

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AOM Antibiotic Recommendations

- Penicillin allergic with history of type 1 reaction, use one of the following:
  - Azithromycin 10 mg/kg per day as a single dose on day one and 5mg/kg per day for days two through five
  - Clarithromycin 15 mg/kg per day divided BID
  - Erythromycin-sulfisoxazole 50 mg/kg per day of the erythromycin component divided TID or QID
  - Clindamycin 10 to 25 mg/kg per day orally divided TID for mild to moderate infections and 30 to 40 mg/kg per day orally divided TID for severe infections

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Why Not Hearing Aids?

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Question 8

A 9-month-old child presents with complaint of failed newborn hearing screens bilaterally. Patient has had repeat screening exams and has failed both ears. Parents feel that he hears, as he turns to look at them when they enter the room but are unsure how well he hears as he has no words. Auditory brainstem response shows severe to profound hearing loss bilaterally. What is the recommended treatment?

A. Hearing aids only

B. Repeat testing in 3 months

C. Cochlear implant evaluation

D. Observe

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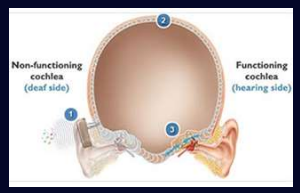
Hearing Aids

- Provide amplification for patients with hearing loss
- Can be both traditional and bone anchored hearing aid (BAHA)



http://www.hearingaidtechnologies.net/
http://www.cochlear.com/wps/wcm/connect/in/home/discover/baha-bone-conduction-implants/baha-connect-system
http://www.earassociates.com/services-bone-anchored-hearing-aids-san-jose-ca.html
https://en.wikipedia.org/wiki/Bone-anchored_hearing_aid

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Cochlear Implant Indications

- Desire for hearing and language
- Severe to profound sensory neural hearing loss with no benefit with hearing aids for at least 3 months

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Cochlear Implant Evaluation

- Evaluation of sensory neural hearing loss
  - MRI internal auditory canals with contrast
  - CT temporal bones without contrast
  - Renal ultrasound
  - Dilated eye exam
  - Electrocardiogram
- If syndromic appearing or family has concerns about future children can do Geneticist evaluation

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Cochlear Implant Evaluation

- Audiology evaluation with hearing aids in place
  - Evaluates for word recognition and hearing threshold
- Prior to implantation
  - Meningitis vaccinations
  - All other pediatric immunizations up to date

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Risks of Cochlear Implant

- Meningitis
- Loss of residual hearing
- Facial nerve paralysis
- Tinnitus
- Abortion of the procedure
- Implant failure
- Altered sense of taste
- Incomplete insertion
- No improvement in speech
- Cerebrospinal fluid leak
- Vertigo
- Skin flap necrosis
- Bleeding

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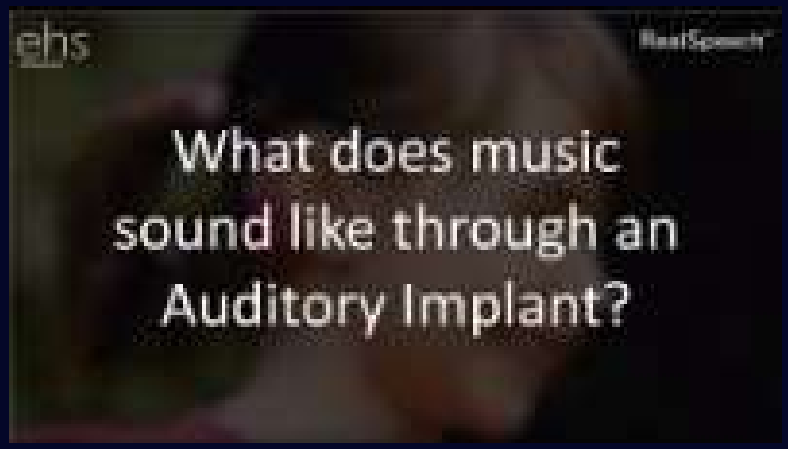
Cochlear Implant



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Question 9

42-year-old female dives into the quarry and has immediate pain and hearing loss when entering the water. She notes bloody drainage from her left ear and comes to the urgent care to be evaluated. Exam reveals her drum appears as below.

https://www.google.com/url?sa=i&rct=j&q=&esrc=s&source=images&cd=&cad=rja&uact=8&ved=0ahUKEwim9K-CgbnNAhUOID4KHS2jA90QjRwIBw&url=http%3A%2F%2Fme.hawkelibrary.com%2Fnew%2Fmain.php%3Fg2_itemId%3D1468&psig=AFQjCNFgACaImkSagoV1Ghi0M5YsIYmSdg&ust=1466594629600205

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Question 9

What is the appropriate treatment?

A. Irrigate ear for better evaluation

B. Cortisporin Otic 5 drops left ear TID for 10 days

C. Floxin Otic 5 drops left ear BID for 10 days

D. Urgent Otolaryngology referral for surgical intervention

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Tympanic Membrane Perforation

- Causes
  - Trauma
  - AOM
  - Residual after ventilation tube extrusion
  - Cholesteatoma
- Symptoms
  - Hearing loss
    - Conductive on audiogram
  - If traumatic, pain
  - If from AOM, pain will resolve when perforates

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Tympanic Membrane Perforation

- Treatment
  - Do NOT irrigate ear if you suspect a perforation
    - You will wash everything into the middle ear
  - Ear drops to prevent blood from drying in ear canal and prevent infection
    - Floxin otic (ofloxacin)
    - Ciprodex otic (ciprofloxacin + dexamethasone)
    - Cipro HC (ciprofloxacin + hydrocortisone)
    - Cipro (ciprofloxacin)
    - Ototvel (ciprofloxacin + fluocinolone acetonide)
    - Cortisporin otic (cin + polymyxin B + hydrocortisine)

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Tympanic Membrane Perforation

- Treatment
  - Observe for 3 months
  - If not healed
    - Continue observation
    - Tympanoplasty

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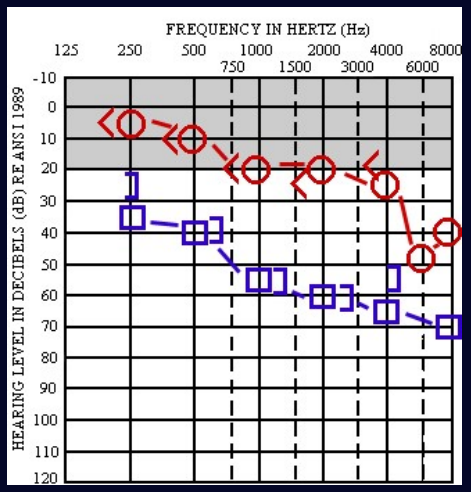
Question 10

68-year-old male complains of hearing loss for 6 years. Left ear is worse than the right ear, tinnitus is louder in left ear, no vertigo. Audiogram noted below. What treatment would you offer?

A. Hearing aids
B. Bone anchored hearing aid
C. Further workup needed before treatment
D. Myringotomy and tube placement in left ear



http://www.dizziness-and-balance.com/disorders/hearing/sensorineural.htm

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Asymmetric Sensory Neural Hearing Loss (SNHL)

- Possible causes
  - Noise exposure or event
  - Congenital
  - Acoustic neuroma (schwannoma)
  - Trauma
  - Meniere's Disease
  - Viral infections
  - Vascular events
  - Idiopathic

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Asymmetric SNHL

- Evaluation
  - MRI of internal auditory canals with gadolinium
  - If unable to have MRI can evaluate with auditory brainstem response testing
- Treatment
  - If sudden
    - Oral steroids
      - We use prednisone taper starting at 60mg for 4 days and tapering over 10-14 days
    - Intratympanic steroids
  - If not sudden or sudden hearing loss does not improve with steroids
    - Hearing aids
    - Utilize hearing protection to preserve remaining hearing and hearing in "good" ear

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Acoustic Neuroma

- Occurs in 10 per 1 million people in the US
- Symptoms
  - 95% of people with acoustic neuroma will have asymmetric SNHL
    - 25% of people with an acoustic neuroma will have sudden SNHL
    - 1%-5% of people with sudden SNHL will have an acoustic neuroma
  - Tinnitus in affected ear
  - Vertigo
  - Facial Paralysis if grows large enough

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Acoustic Neuroma

- Treatment
  - Gamma knife
    - Used in tumors 4cm or less
  - Open resection
    - Reserved for tumors too large for gamma knife

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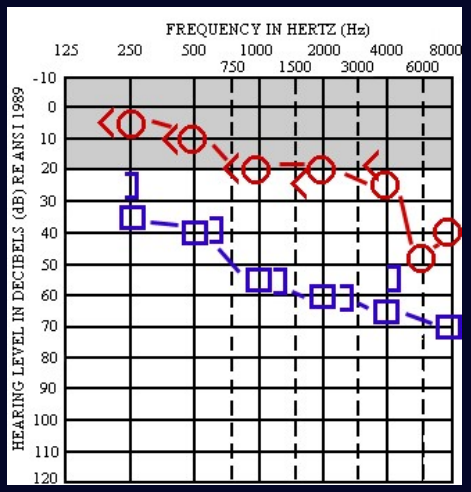

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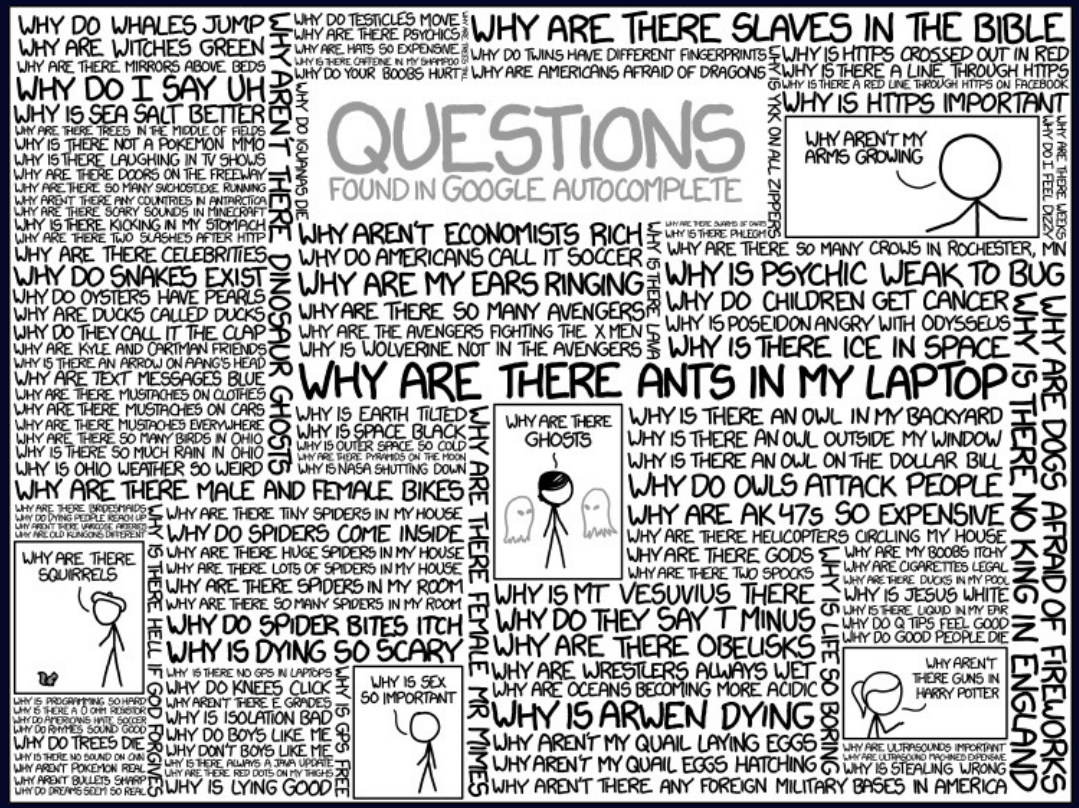
Questions?



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