We are called to make a healthy difference in people's lives.

"Can You Hear Me Now?": Inner Ear Emergencies

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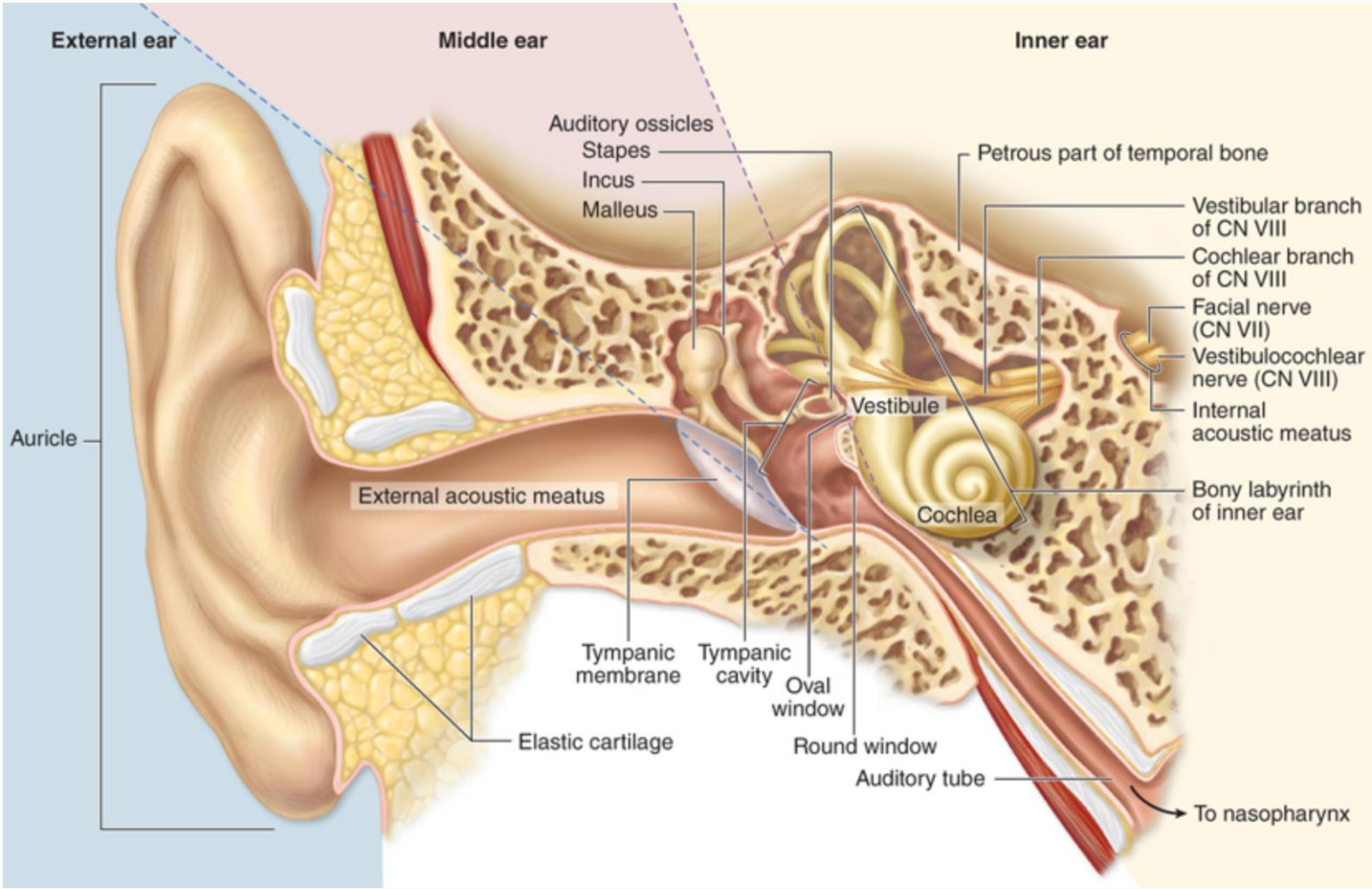
Essentia Health

The College of St. Scholastica

Objectives

- Following today's lecture, you should be able to:
  - Differentiate the basic types of acute inner ear pathologies and their respective treatments
  - Recognize key history and exam findings to help guide your way
  - Identify referral criteria and implement options

Divisions of the Ear



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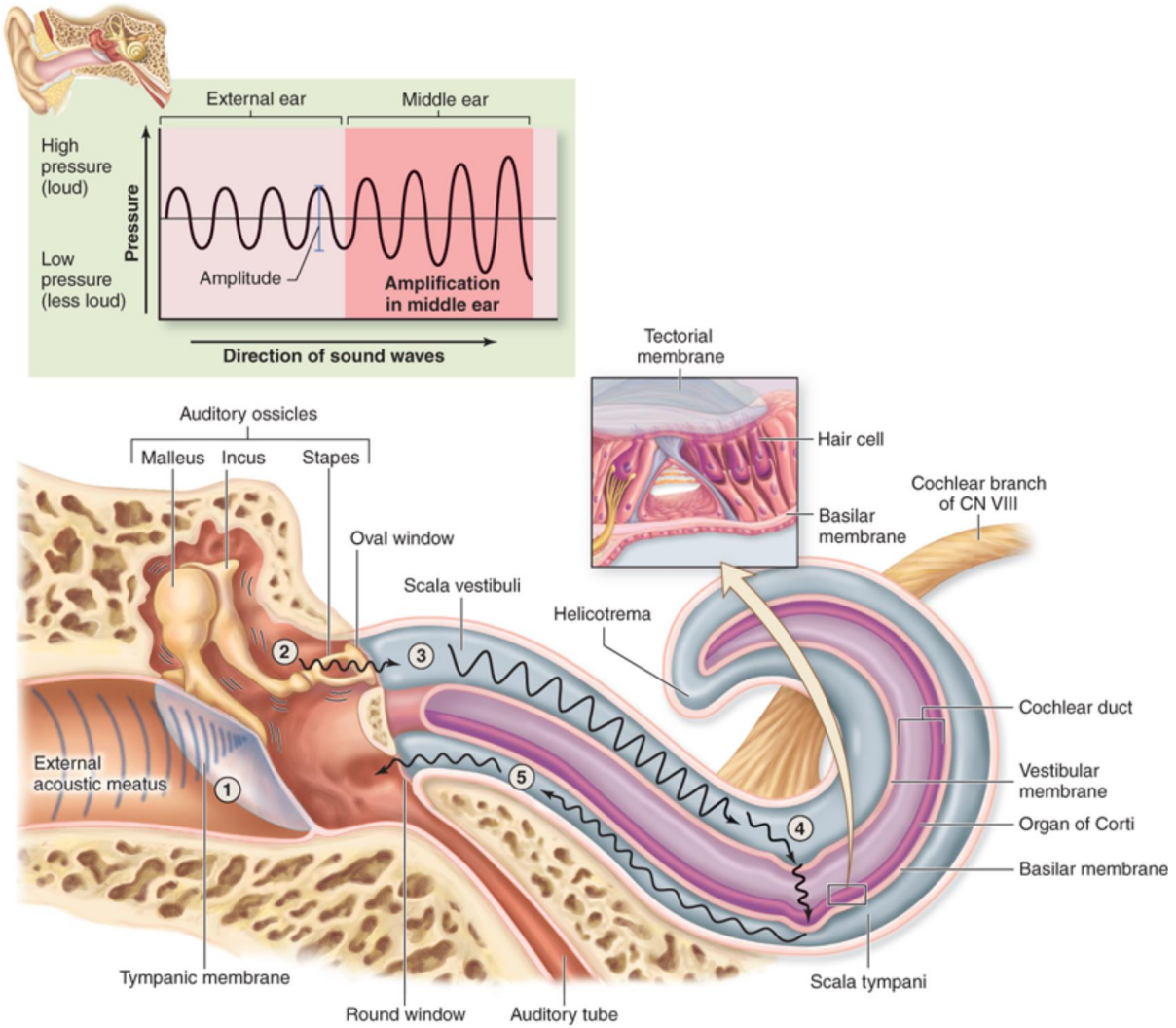
Path of Sound Wave Through the Ear



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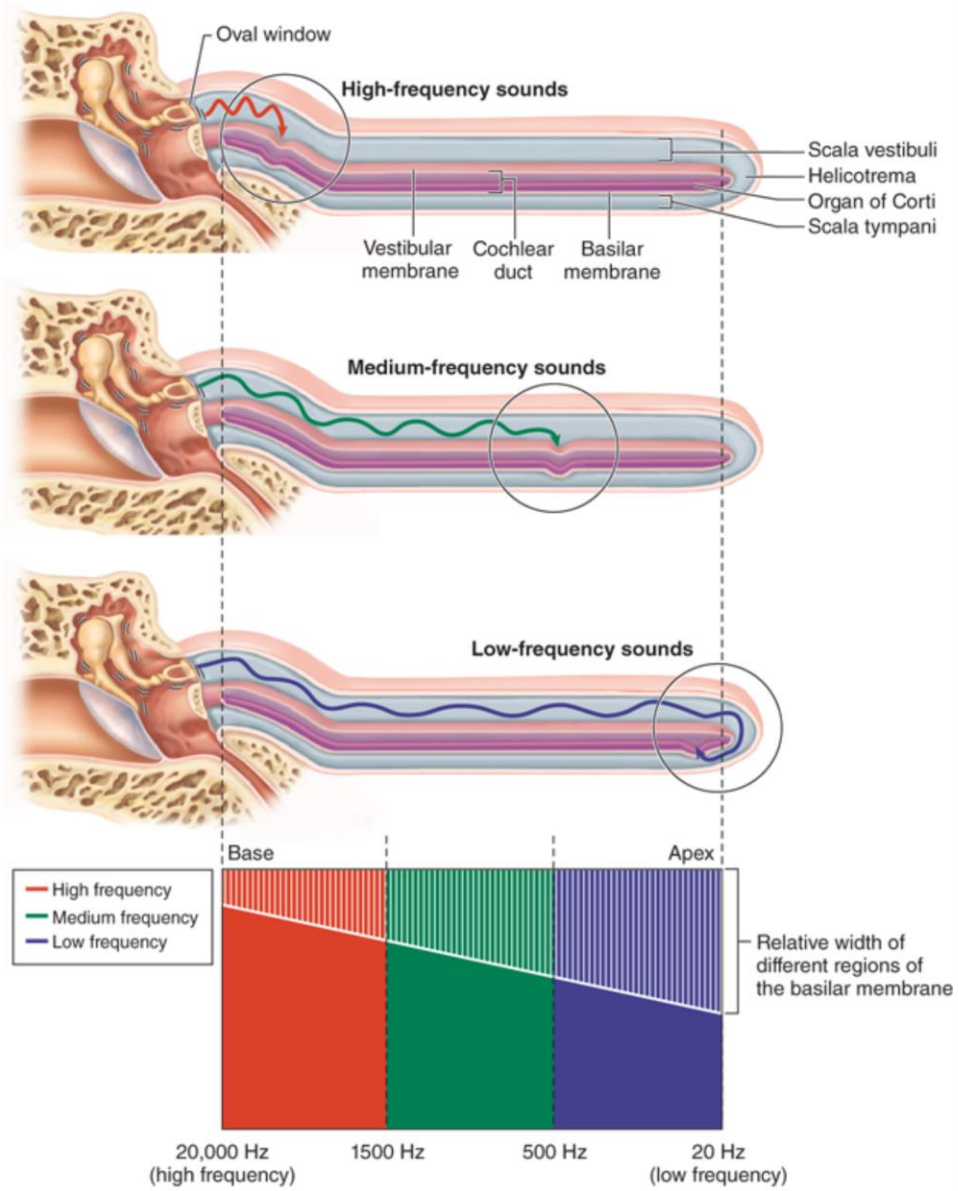
Tonotopic Map



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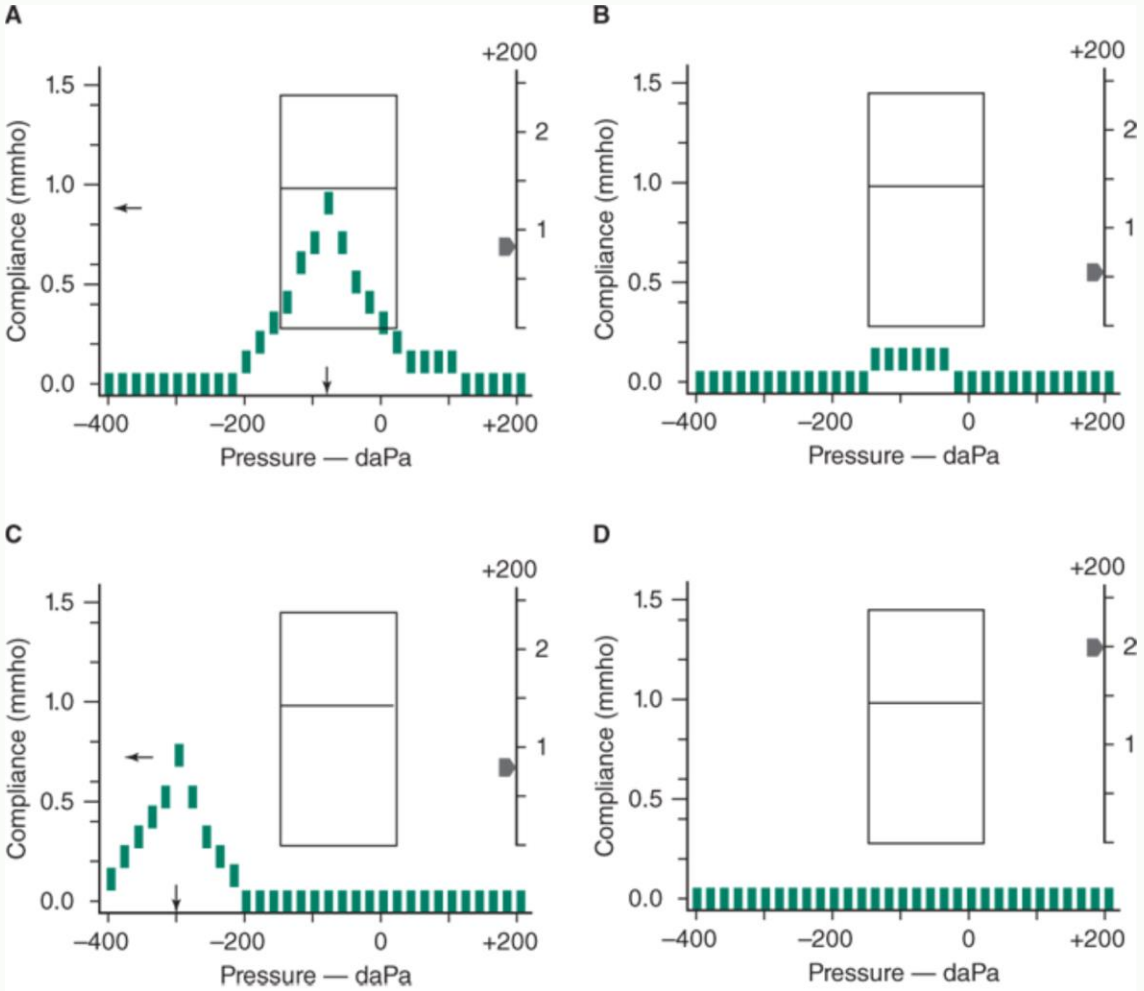
Tympanogram



Ear, Nose, & Throat, *CURR*: Yoon PJ, Scholes MA, Herrmann BW. Yoon P.J., & Scholes M.A., & Herrmann B.W. Yoon, Patricia J., et al.Ear, Nose, & Throat. In: Hay Jr. WW, Levin MJ, Abzug MJ, Bunik M. Hay Jr. W.W., & Levin M.J., & Abzug M.J., & Bunik M(Eds.),Eds. William W. Hay Jr., et al.eds. *Current Diagnosis & Treatment: Pediatrics, 25e*. McGraw Hill; 2020.
Accessed October 05, 2021. https://accessmedicine-mhmedical-com.akin.css.edu/content.aspx?bookid=2815&sectionid=244260113

Hearing Loss Classification

- Conductive Hearing Loss
  - Can occur due to dysfunction of the external auditory canal (EAC), the TM, or the ossicles
- Sensory-Neural Hearing Loss
  - Caused by dysfunction of the sensory (cochlea) or neural components of the auditory system
- Mixed Hearing Loss
  - Combination of the conductive and sensory-neural hearing loss

Tuning Fork Tests

- Helpful for differentiating between CHL and SNHL

Weber Test

Tuning fork is placed on the center of the forehead

- Normal Hearing
  - Sound is heard equally in both ears
- CHL
  - Sound lateralizes to the “bad” ear
- SNHL
  - Sound lateralizes to the “good” ear

Rinne Test

512 Hz tuning fork is held first on mastoid tip and then on shoulder

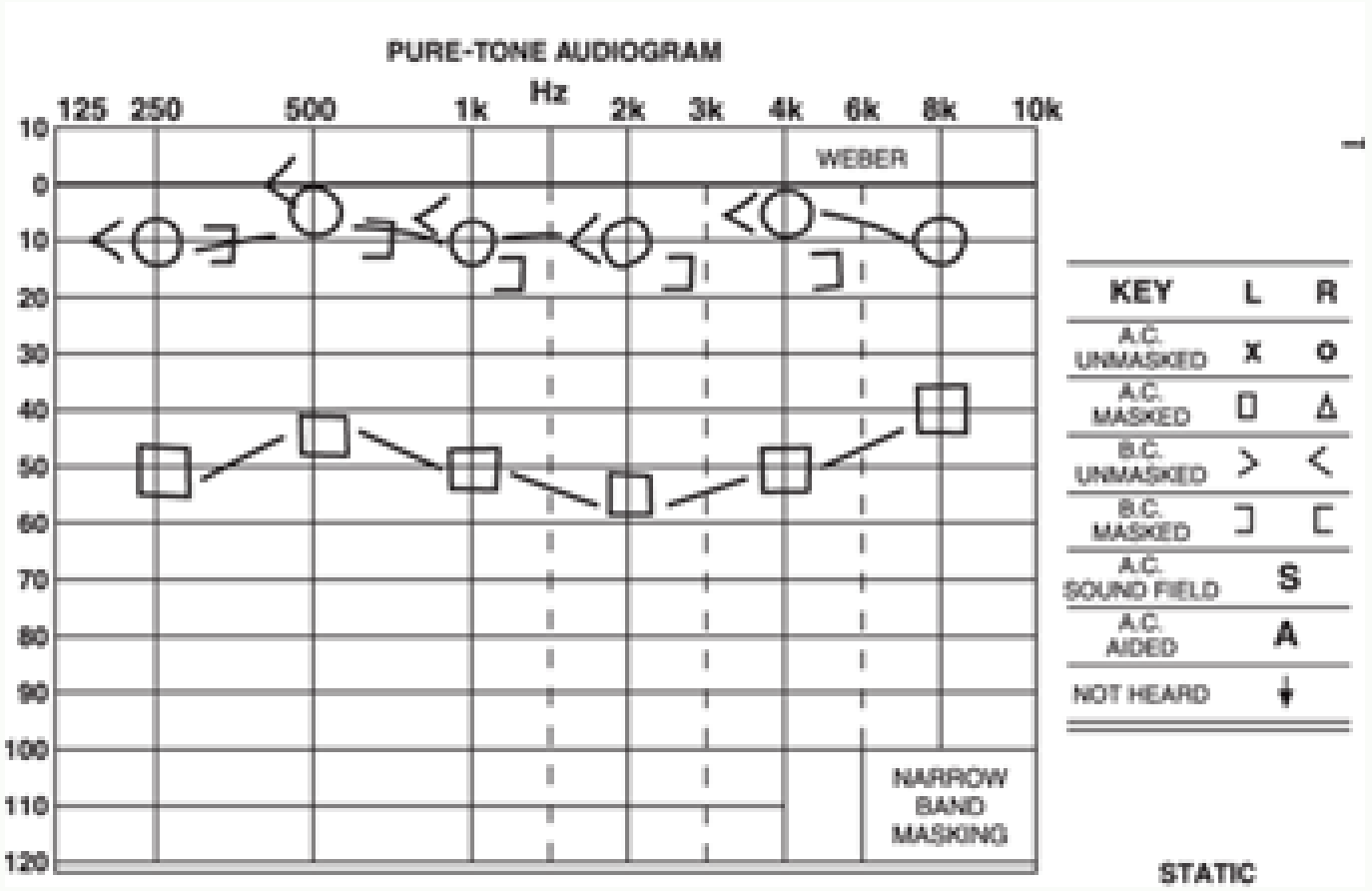
- Normal Hearing
  - Air conduction (AC) > Bone conduction (BC)
- CHL
  - AC = BC or BC > AC
- SNHL
  - AC > BC

Interpretation of Tuning Fork Tests

Results of Weber Test	Results of Rinne Test	Interpretation
Sound does not lateralize	Air conduction greater than bone conduction bilaterally	No hearing loss or equal sensorineural hearing loss bilaterally
Sound does not lateralize	Bone conduction greater than air conduction bilaterally	Equal conductive hearing loss bilaterally
Sound lateralizes to one side	Air conduction greater than bone conduction bilaterally	Sensorineural hearing loss on the side opposite of the lateralization
Sound lateralizes to one side	Bone conduction greater than air conduction on the side of lateralization	Conductive hearing loss on the side of lateralization
Sound lateralizes to one side	Bone conduction greater than air conduction bilaterally	Bilateral conductive hearing loss, greater on the side of lateralization

Middle Ear Effusions & Conductive Hearing Loss (CHL)

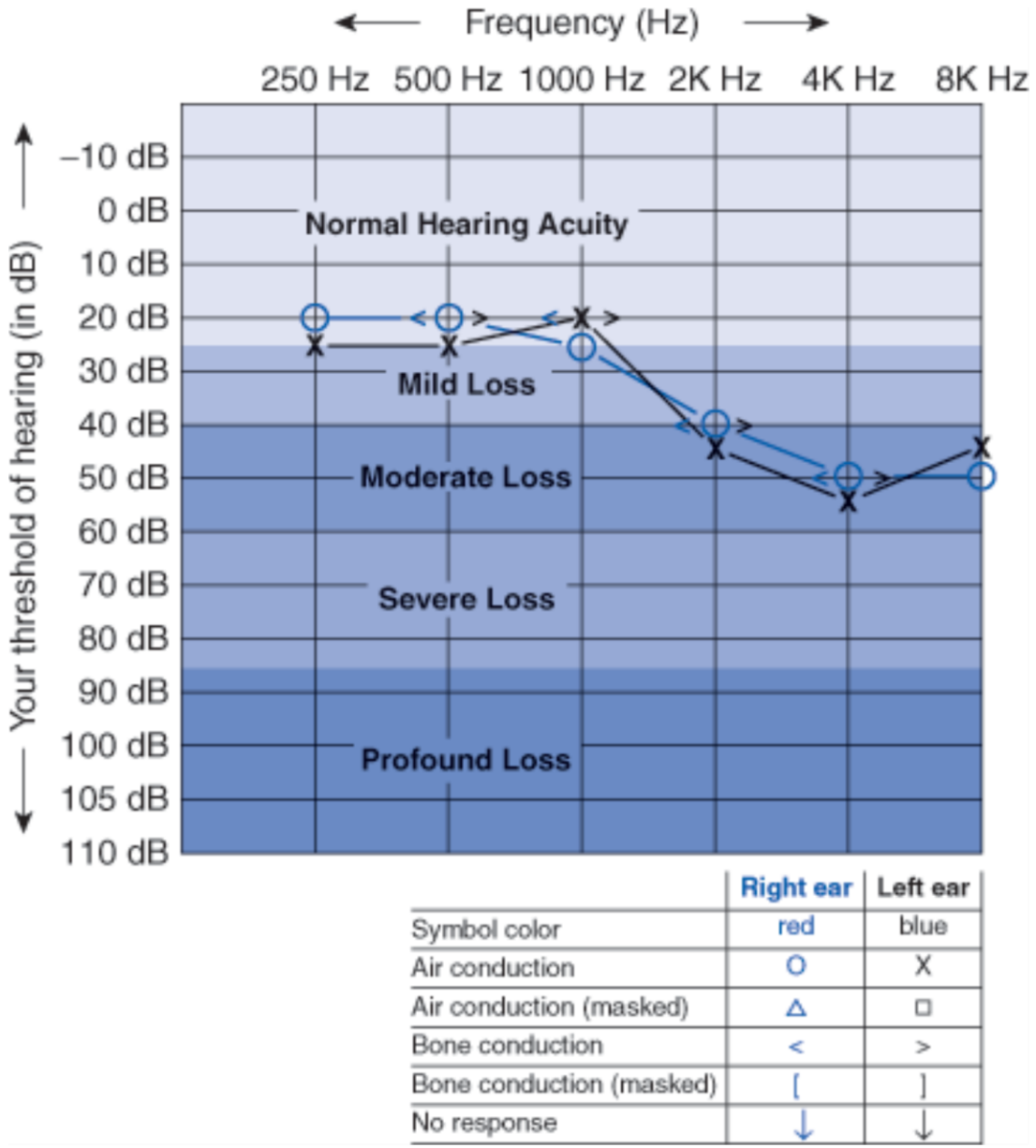
- TALK ABOUT THIS!!!!



Sensory-Neural Hearing Loss (SNHL)

- *Presbycusis*
- Ototoxic drugs
- Meniere's disease
- *Sudden Sensorineural Hearing Loss (SSNHL)*
- *Acoustic neuroma*
- Multiple sclerosis
- *Autoimmune disease*

Audiogram



Managing Hearing Impairment in Older Adults, *Current Diagnosis & Treatment: Geriatrics, 2e*

Williams BA, Chang A, Ahalt C, Chen H, Conant R, Landefeld C, Ritchie C, Yukawa M. *Current Diagnosis & Treatment: Geriatrics, 2e*; 2014

What noises CAN'T we hear?

If you have a **mild** hearing loss, you couldn't hear . . .

- Whispers
- A fan blowing
- A clock ticking

If you have a **moderate** hearing loss, you couldn't hear . . .

- Normal conversation
- A dishwasher
- A clothes dryer

What noises CAN'T we hear?

If you have a **severe** hearing loss, you couldn't hear . . .

- Almost all conversation
- Music
- A hand drill

If you have a **profound** hearing loss, you couldn't hear . . .

- A chainsaw
- A lawnmower
- A siren

Sudden Sensory-Neural Hearing Loss (SSNHL)

- Hearing loss that has a rapid onset, occurring within 72 hours
  - 30 dB or greater over at least three contiguous audiometric frequencies
  - OR 20 dB or greater at 3000 Hz
  - Typically involves one ear
- Epidemiology
  - 5-20 per 100,000
  - Affects males and females equally
  - peak incidence is between the 5^{th}-6^{th} decade of life
- Etiology and Pathophysiology

Kuhn, M., Heman-Ackah, S., Shaikh , J., & Roehm, P. (2011, May). Sudden Sensorineural Hearing Loss, *Trends in Hearing*, 15(3). doi: 10.1177/1084713811408349

Etiology and Pathophysiology

Table 1. Identifiable Causes of Sudden Sensorineural Hearing Loss

Autoimmune	Autoimmune inner ear disease	Neurologic	Migraine
	Behcet's disease		Multiple sclerosis
	Cogan's syndrome		Pontine ischemia
	Systemic lupus erythematosis		
Infectious	Bacterial meningitis	Otologic	Fluctuating hearing loss
	Cryptococcal meningitis		Meniere's disease
	HIV		Otosclerosis
	Lassa fever		Enlarged vestibular aqueduct
	Lyme Disease		
	Mumps	Toxic	Aminoglycosides
	Mycoplasma		Chemotherapeutic agents
	Syphilis		Non-steroidal anti-inflammatories
	Toxoplasmosis		Salicylates
Functional	Conversion disorder	Traumatic	Inner ear concussion
	Malingering		Iatrogenic trauma/surgery
Metabolic	Diabetes mellitus		Perilymphatic fistula
	Hypothyroidism		Temporal bone fracture
Neoplastic	Vestibular schwannoma	Vascular	Cardiovascular bypass
	CPA or petrous meningiomas		Cerebrovascular accident/stroke
	CPA or petrous apex metastases		Sickle cell disease
	CPA myeloma		

Kuhn, M., Heman-Ackah, S., Shaikh , J., & Roehm, P. (2011, May). Sudden Sensorineural Hearing Loss, *Trends in Hearing*, 15(3). doi: 10.1177/1084713811408349

Diagnosis

- Risk Factors
  - Recent viral infection
- Normal otoscopic and neuro examination
- Differential
  - “KITTENS”
    - Congenital
    - Idiopathic/Infectious/Inflammatory
    - Traumatic/Toxic
    - Endocrine
    - Neoplastic
    - Systemic (Autoimmune)

Diagnosis

- Diagnostic Tests/Interpretation
  - MRI brain/ & IACs with and without contrast
  - CT Temporal Bone
  - Labs
    - VDRL
    - TSH w/reflex (+Thyroid antibodies if TSH abnormal)
    - Glucose
    - Lyme titre

Treatment

- General Measures
  - Referral to ENT and Audiology ASAP
- Medications
  - *High-dose oral steroids*
- Surgery/Procedures
  - Intratympanic dexamethasone (ENT)

Ongoing Care

- Follow up
  - Serial audiograms
- Patient Education
  - Reassurance and importance of ENT follow-up
- Prognosis
  - ~2/3 will experience recovery, but often not complete
- Complications
  - Depression

Acoustic Neuroma

- Schwann cell-derived tumors
  - commonly originating from the vestibular portion of CN VIII
- Epidemiology
  - incidence of 10 per 1 million individuals each year
  - No gender bias
  - Age of presentation is between 40 and 60 yrs of age
- Risk Factors
  - Neurofibromatosis (type II)
  - Family history

Diagnosis

- History/Symptoms
  - Most common manifestation is unilateral SNHL and tinnitus
  - May complain of balance problems
- Exam Findings/Signs
  - Poorer word recognition on the involved side
  - Abnormal tandem gait
  - CN VII asymmetry in large tumors

Diagnosis

Differential

"KITTENS"

- Congenital
- Idiopathic/Infectious/Inflammatory
- Traumatic/Toxic
- Endocrine
- Neoplastic
- Systemic (Autoimmune)

Diagnostic Tests

- Audiogram
- MRI brain/ & ACs with and without contrast

Treatment

- General Measures
  - Observation
- Medications
  - High-dose oral steroids
- Surgery/Procedures
  - Surgical resection
  - Stereotactic radiosurgery

Ongoing Care

- Follow up
- Patient Education
- Prognosis
- Complications
  - Unusable ear
  - Chronic vertigo
  - Unilateral facial paralysis
- When to Refer

Autoimmune SNHL

- May be associated with a wide array of systemic autoimmune disorders (15-30%)
- In many cases, the autoimmune pattern of audio-vestibular dysfunction presents in the absence of recognized systemic autoimmune disease
- Epidemiology
  - Incidence
  - Prevalence

Goodall, AF & Siddiq, MA. (2015). Current understanding of the pathogenesis of autoimmune inner ear disease: a review. *Clin Otolaryngol,40*(5):412–9.

Diagnosis

- History/Symptoms
  - Loss is most often bilateral and progressive
  - The hearing level often fluctuates, with periods of deterioration alternating with partial or even complete remission
  - Vestibular dysfunction, particularly disequilibrium and postural instability, may accompany the auditory symptoms

Diagnosis

- Exam Findings/Signs
  - Normal ENT exam
  - Underlying autoimmune diseases
- Differential
  - Meniere's vs idiopathic SSNHL vs other
- Diagnostic Tests/Interpretation
  - Audiogram
  - laboratory tests to screen for autoimmune disease

Treatment

- General Measures
  - **Refer to audiology and ENT!**
- Medications
  - Corticosteroids
- Surgery/Procedures
  - PE Tube placement (Pope wick) and steroid ear drops
  - intratympanic injections

Ongoing Care

- Follow up
  - Serial audiograms and treat underlying autoimmune disease (if any)
- Patient Education
- Prognosis
- Complications
- When to Refer

Tinnitus

- A perception of abnormal sounds such as ringing, buzzing, or roaring noises, often accompany hearing loss
- Epidemiology
  - More common with advancing age
  - Men are 1.4 to 1.8 times more likely to be affected by tinnitus than women
  - Whites are 2.2 times more likely to be affected than blacks
  - In children, the prevalence of tinnitus ranges from 1% to 13%
  - Estimated 50 million people in US have chronic tinnitus
- Etiology and Pathophysiology
- Risk Factors

Hyperacusis

- Excessive sensitivity to sound
- Epidemiology
  - 40% of patients with tinnitus also have hyperacusis
  - 86% of patients with hyperacusis also have tinnitus
- Etiology and Pathophysiology
- Risk Factors

Diagnosis

- Exam Findings/Signs
  - Symptoms worse with stress, anxiety, fatigue, caffeine, ASA/Ibuprofen
  - Tinnitus worse in quiet environments, at night
  - Hyperacusis worse with loud/high-pitched/metallic sounds

Differential Diagnosis

Peripheral

- Hearing loss
- Stapedectomy
- Perilymph fistula
- Head injury/whiplash
- TMJ

Central

- Ototoxic medications
- Infectious
- Metabolic
- Neurologic
- Autoimmune
- Depression/Anxiety/PTSD
- Vascular

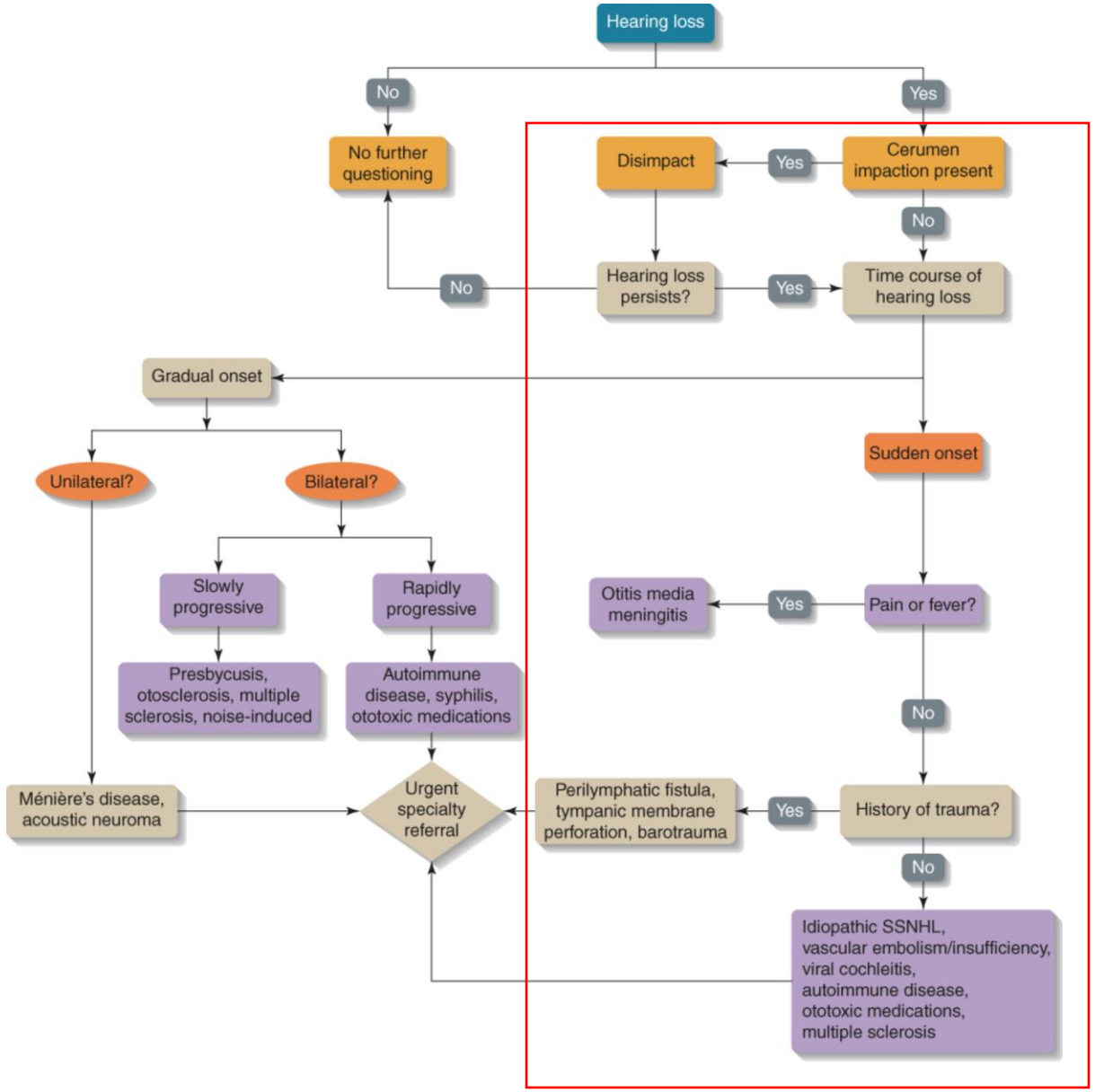
Treatment

- General Measures
  - Amplification
  - Distraction and relaxation
  - Biofeedback
  - Habituation/desensitization
  - Cognitive Behavioral Therapy (CBT)
- Medications
  - +/- Antidepressants

Ongoing Care

- Follow up
- Patient Education
- Prognosis
- Complications
  - Depression, anxiety
- When to Refer

Hearing Loss Diagnostic Algorithm

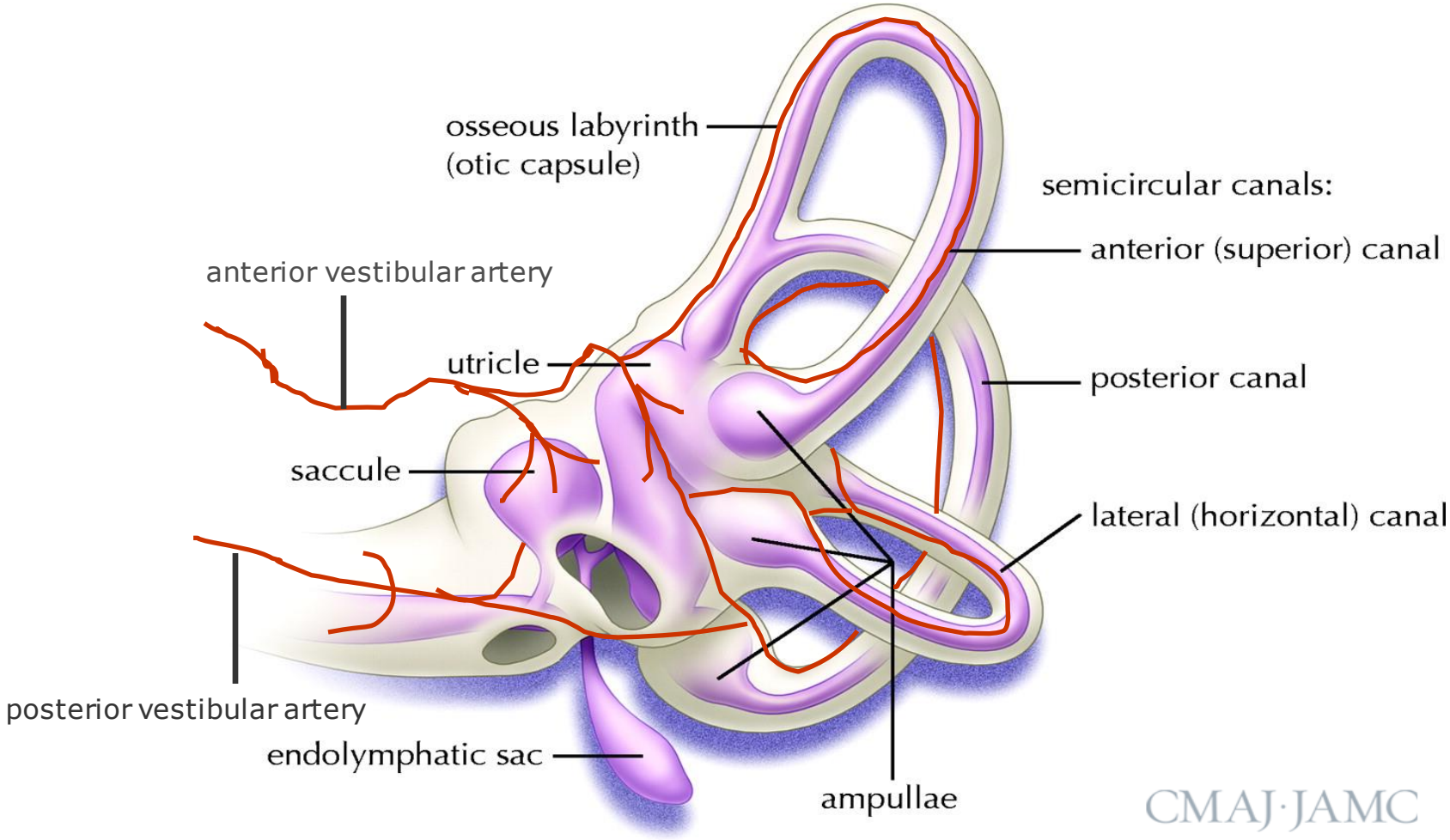


Source: Chapter 18. Hearing Loss, *The Patient History: An Evidence-Based Approach to Differential Diagnosis, 2e*

Citation: Henderson MC, Tierney LM, Jr., Smetana GW. *The Patient History: An Evidence-Based Approach to Differential Diagnosis, 2e*; 2012

Labyrinthine Anatomy

Osseous and membranous labyrinth, and vestibular arteries (off the anterior inferior cerebellar artery) of the left inner ear



Parnes, L S et al. CMAJ 2003;169:681-693

Vestibular Neuronitis and Labyrinthitis

- Epidemiology
  - Third most common cause of peripheral vestibular vertigo after BPPV and Meniere disease
  - No gender bias and typically affects middle-aged people
  - Less than half the patients have an antecedent or concurrent viral illness
- Etiology and Pathophysiology
- Risk Factors

Diagnosis

- Exam Findings/Signs
- Differential
- Diagnostic Tests/Interpretation
  - Audiogram
  - VNG
  - PT Evaluation

Vestibular Neuronitis and Labyrinthitis

- Medications
  - Corticosteroids
  - Benzodiazepines, anticholinergics very helpful first two weeks
    - Prolonged use, however, can delay/impede CNS compensation
- Will need intensive vestibular rehabilitation therapy

Ongoing Care

- Follow up
- Patient Education
- Prognosis
  - Recurrent attacks in the same or contralateral ear have been reported but are unusual
- Complications
- When to Refer

We are called to make a healthy difference in people's lives.

Questions?

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References

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