

Hearing Considerations and Fanconi Anemia

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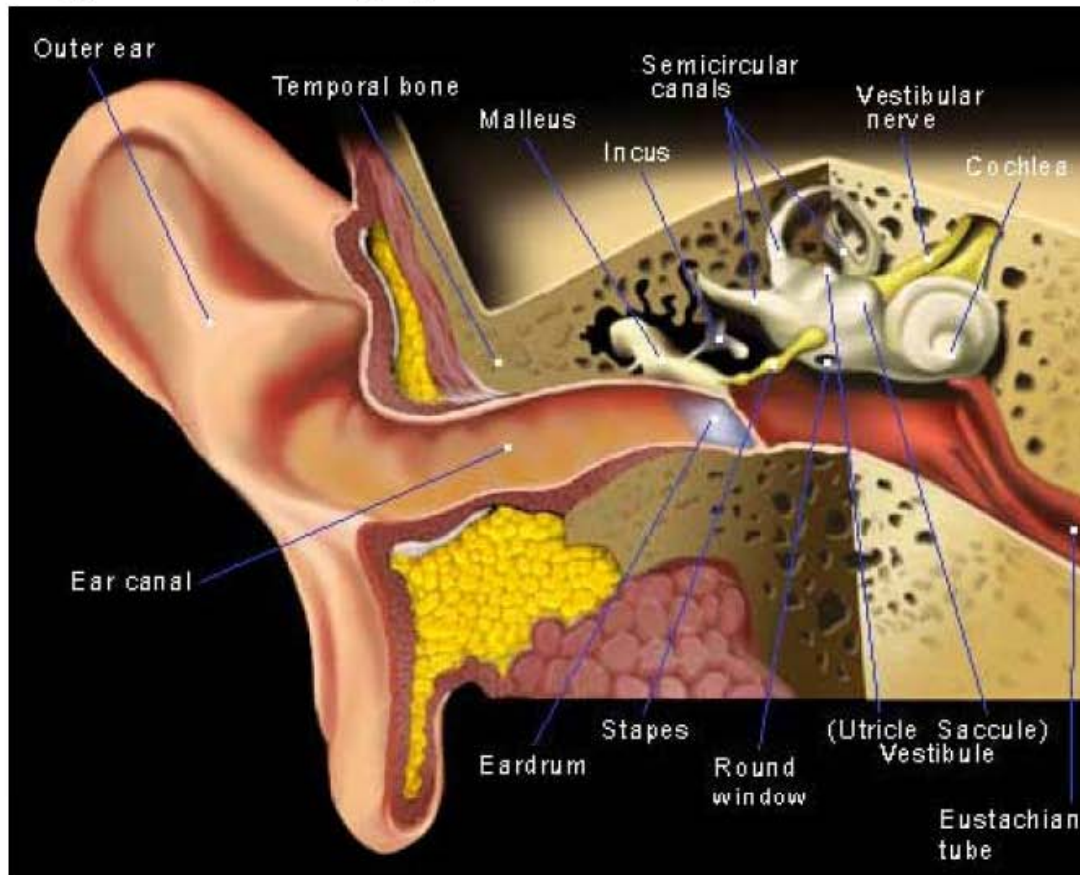


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Basic Ear Anatomy

Figure 1: Anatomy of Ear



From MedicineNet.com

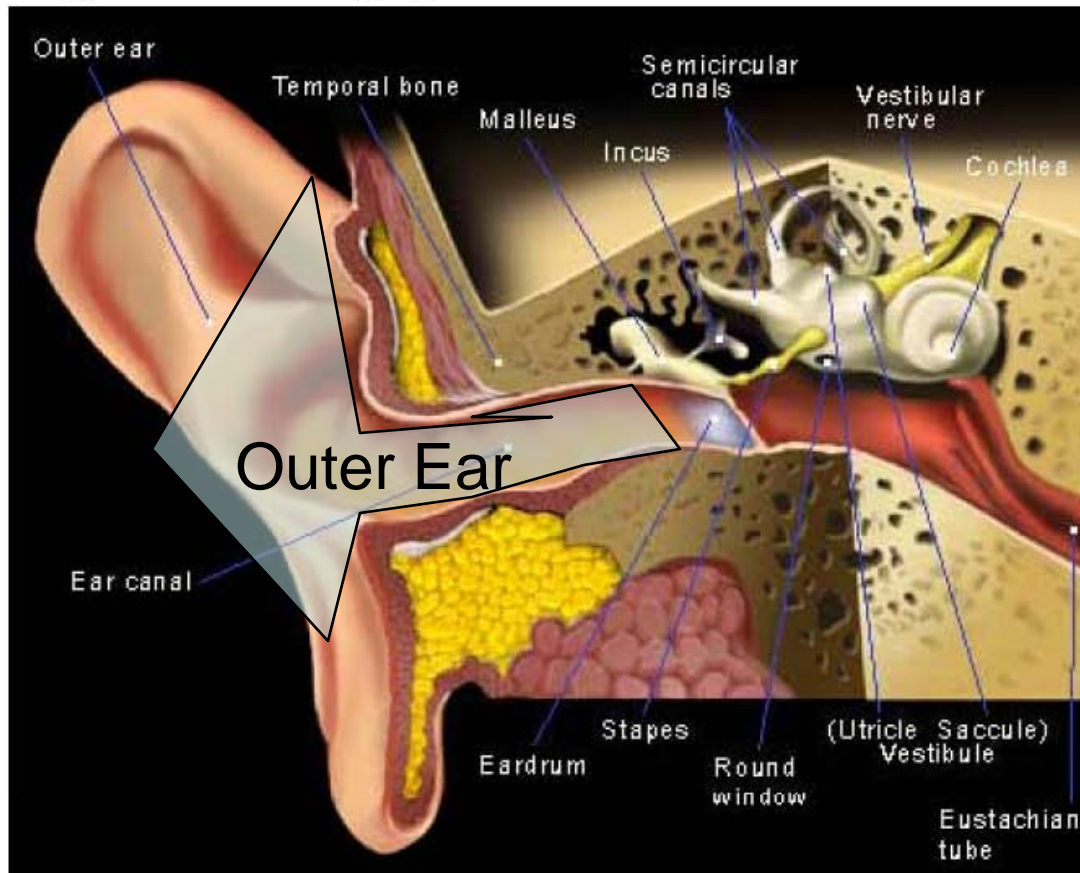


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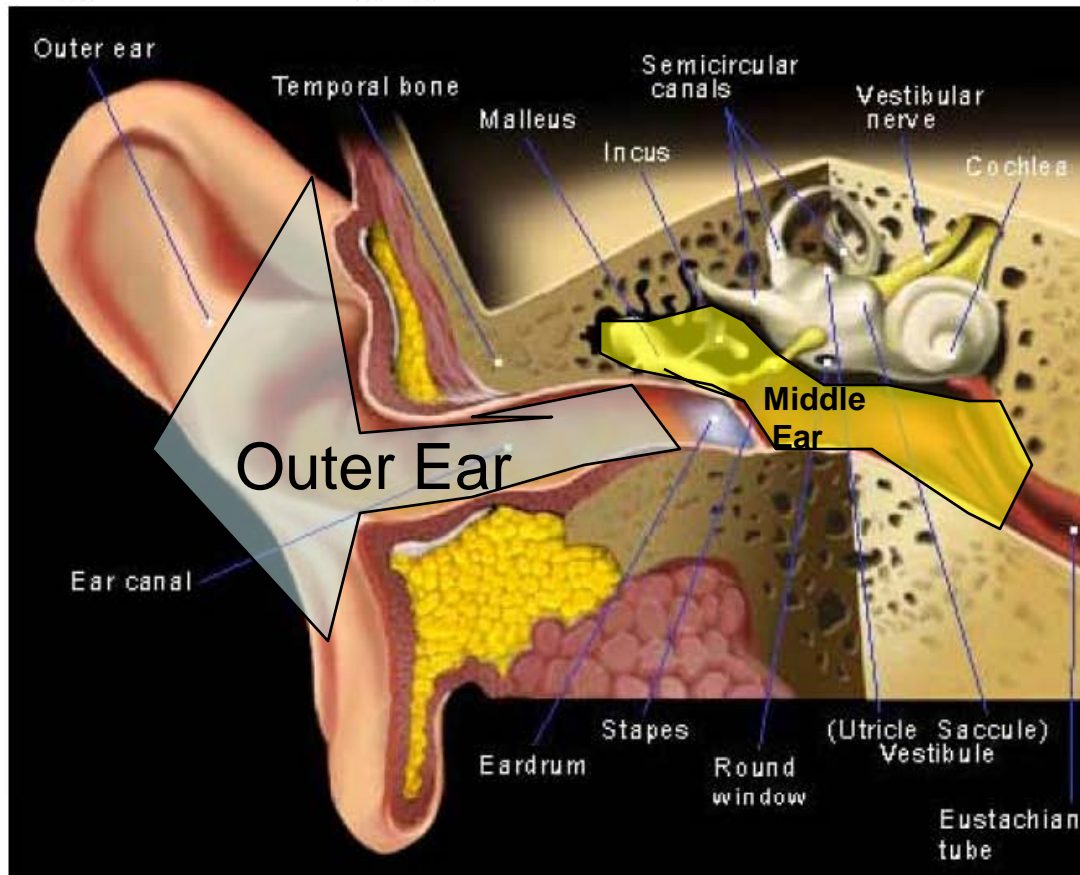


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Basic Ear Anatomy

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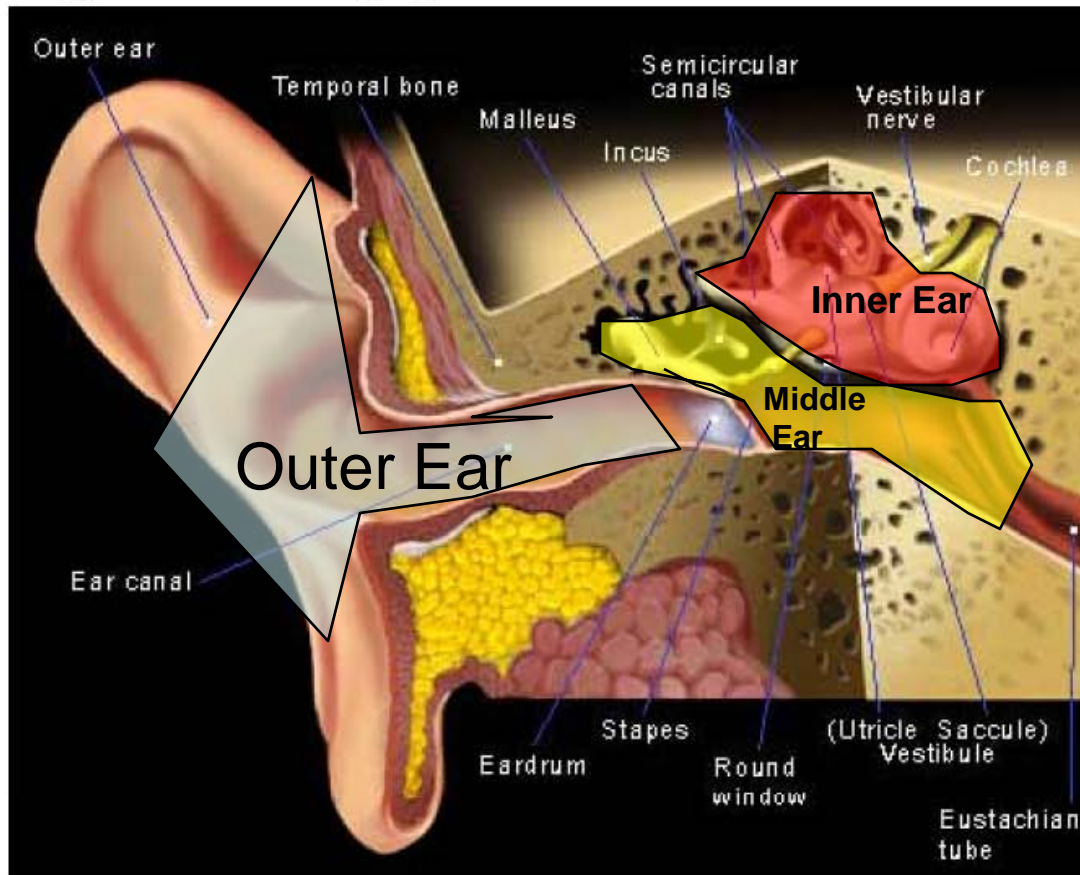


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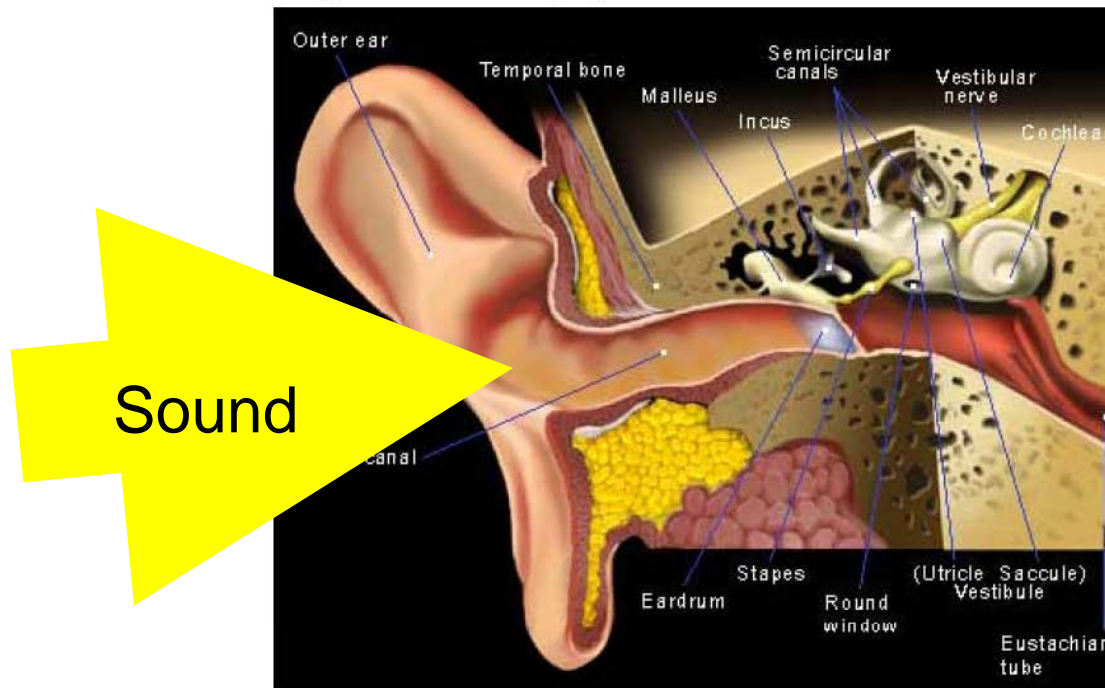
Special Sense of Hearing

- Sound is produced
- It is collected by the external ear, travels through the ear canals
- Vibrates the ear drums
- Is amplified by the tiny hearing bones in the middle ear (Malleus, Incus, Stapes)
- Is transmitted to the inner ear
- Is sent to the brain



Sound Transmission

Figure 1: Anatomy of Ear

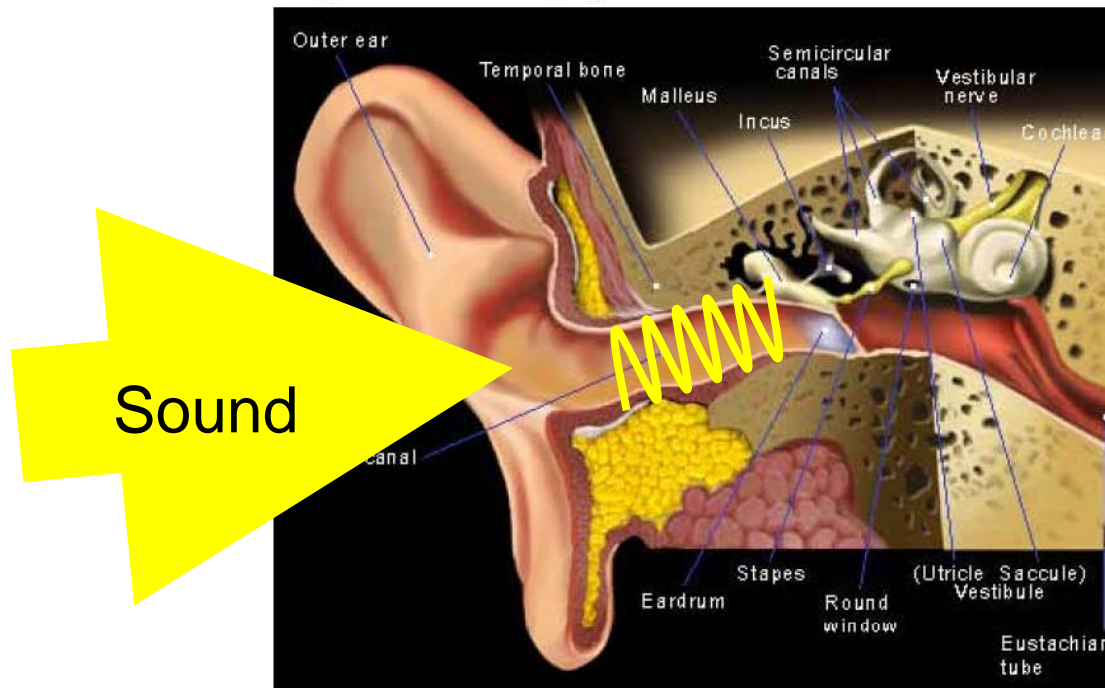


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Sound Transmission

Figure 1: Anatomy of Ear

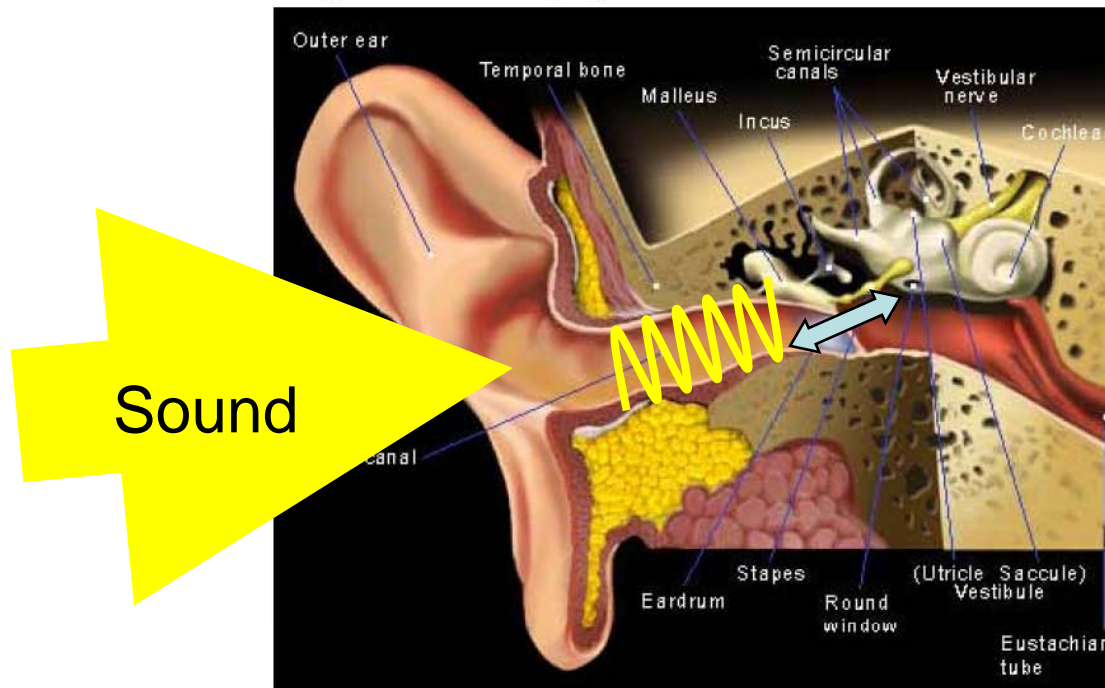


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Parts of the EAR

Figure 1: Anatomy of Ear

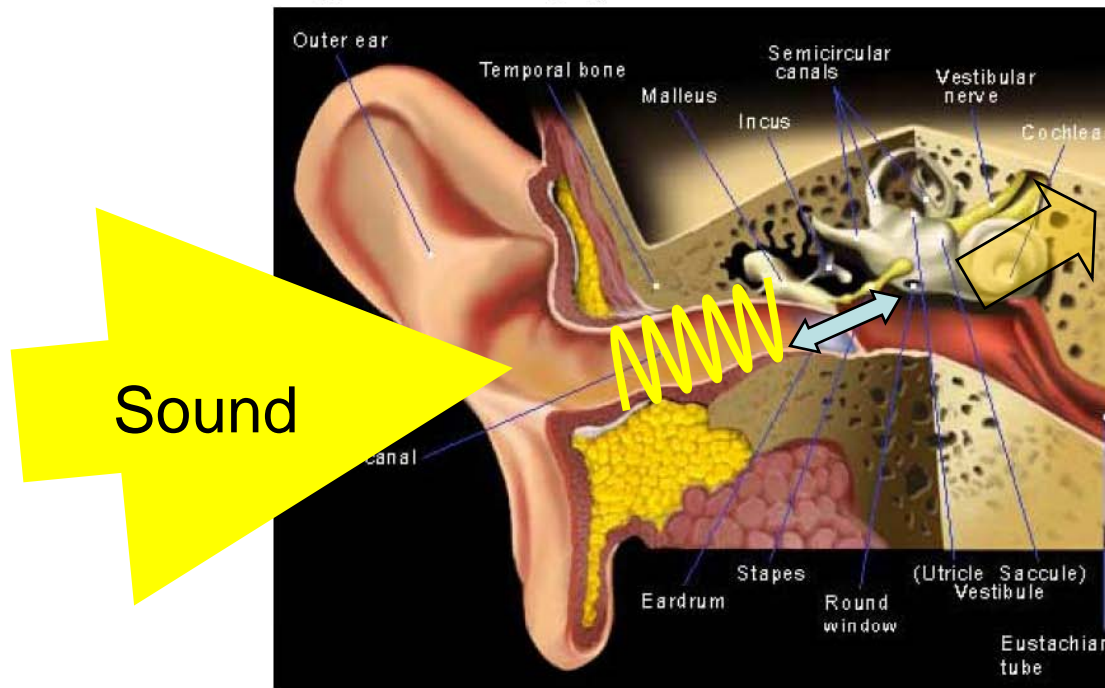


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Sound Transmission

Figure 1: Anatomy of Ear



From MedicineNet.com

30 fold amplification
Through the hearing
Bones before transduction
To the Brain



Normal Hearing

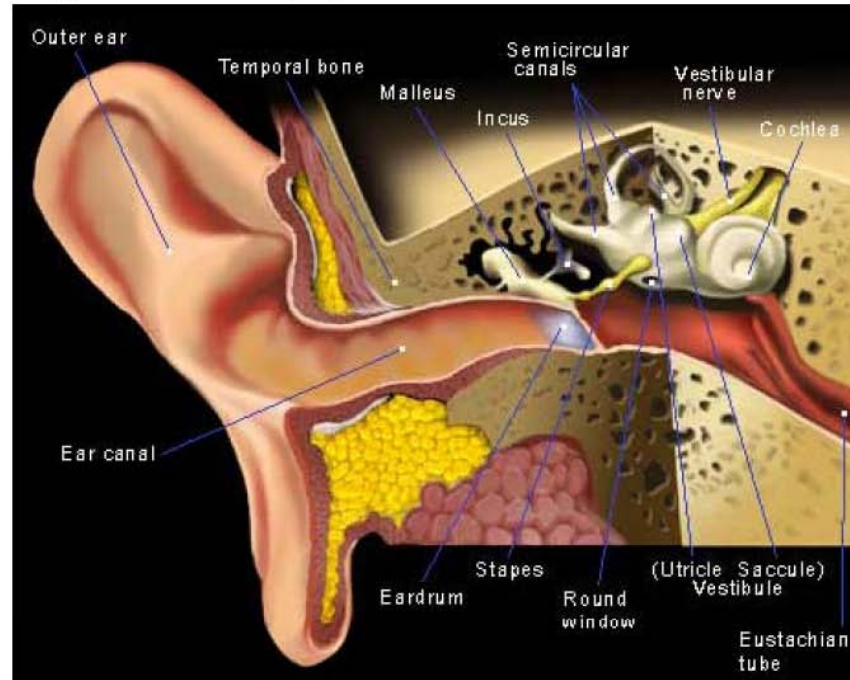
- The Conduction Path Needs to be entirely intact the whole way
- Any breach in integrity can cause diminished hearing



Problems in Sound Transmission

- 1) Deformed Ear
- 2) Deformed canal
- 3) Ear Wax (cerumen)
- 4) Ear drum hole
- 5) Ear bone Abnormality
- 6) Middle ear fluid
- 7) Inner ear damage
(congenital/ rock music)
- 8) Nerve Damage
- 9) Brain Damage (very rare)

Figure 1: Anatomy of Ear



From MedicineNet.com



The Audiogram

- Objective test of hearing evaluation that can be administered at all ages
- Behavioral audiometry is used in the very young.
- Otoacoustic emissions, ABR(BAER) can evaluate hearing as well

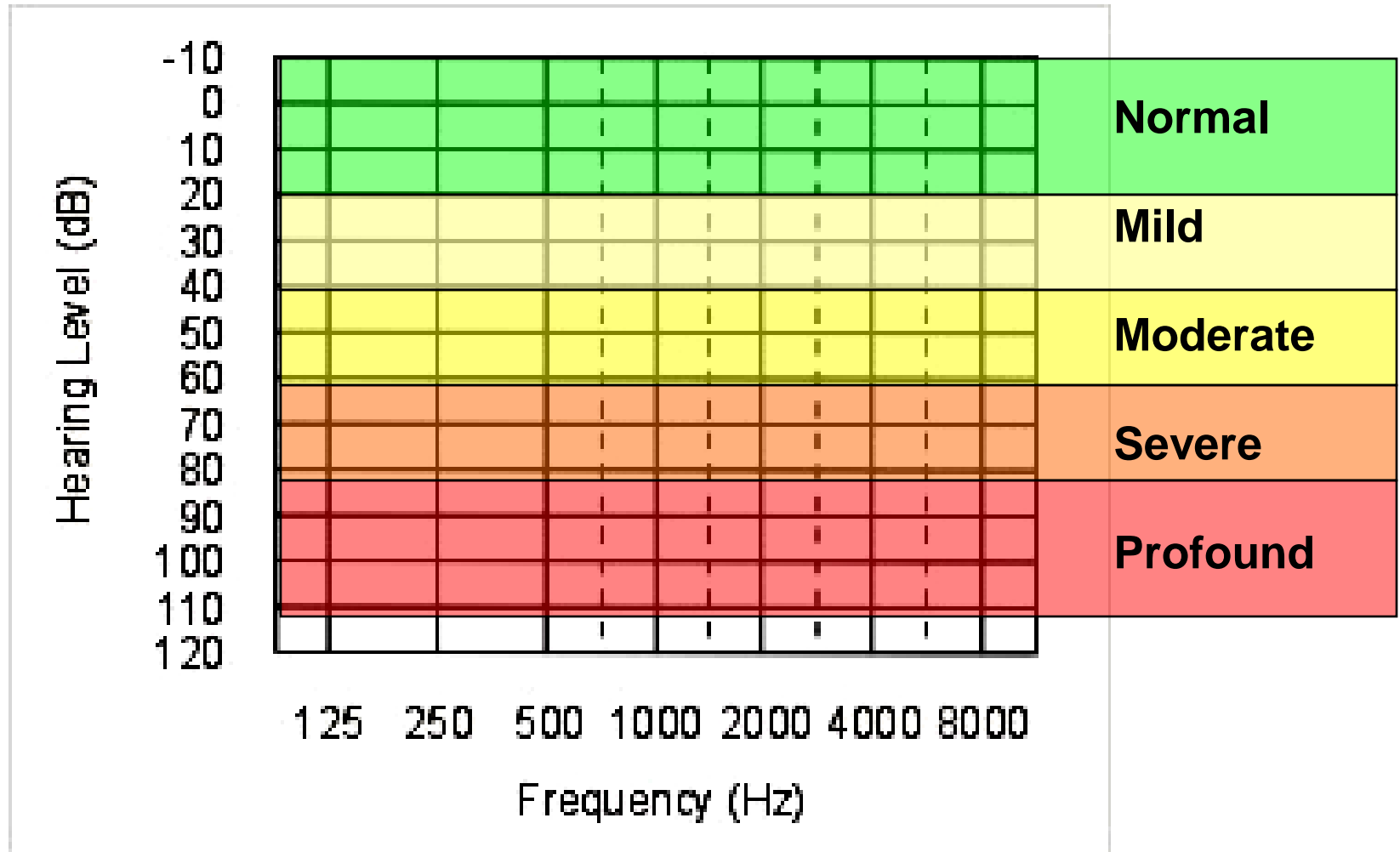


Audiogram- Continued

- Can reliably detect clinically significant hearing changes that require intervention (surgery, hearing aids, etc.)
- Hearing important for speech development so early diagnosis of abnormal hearing very important.



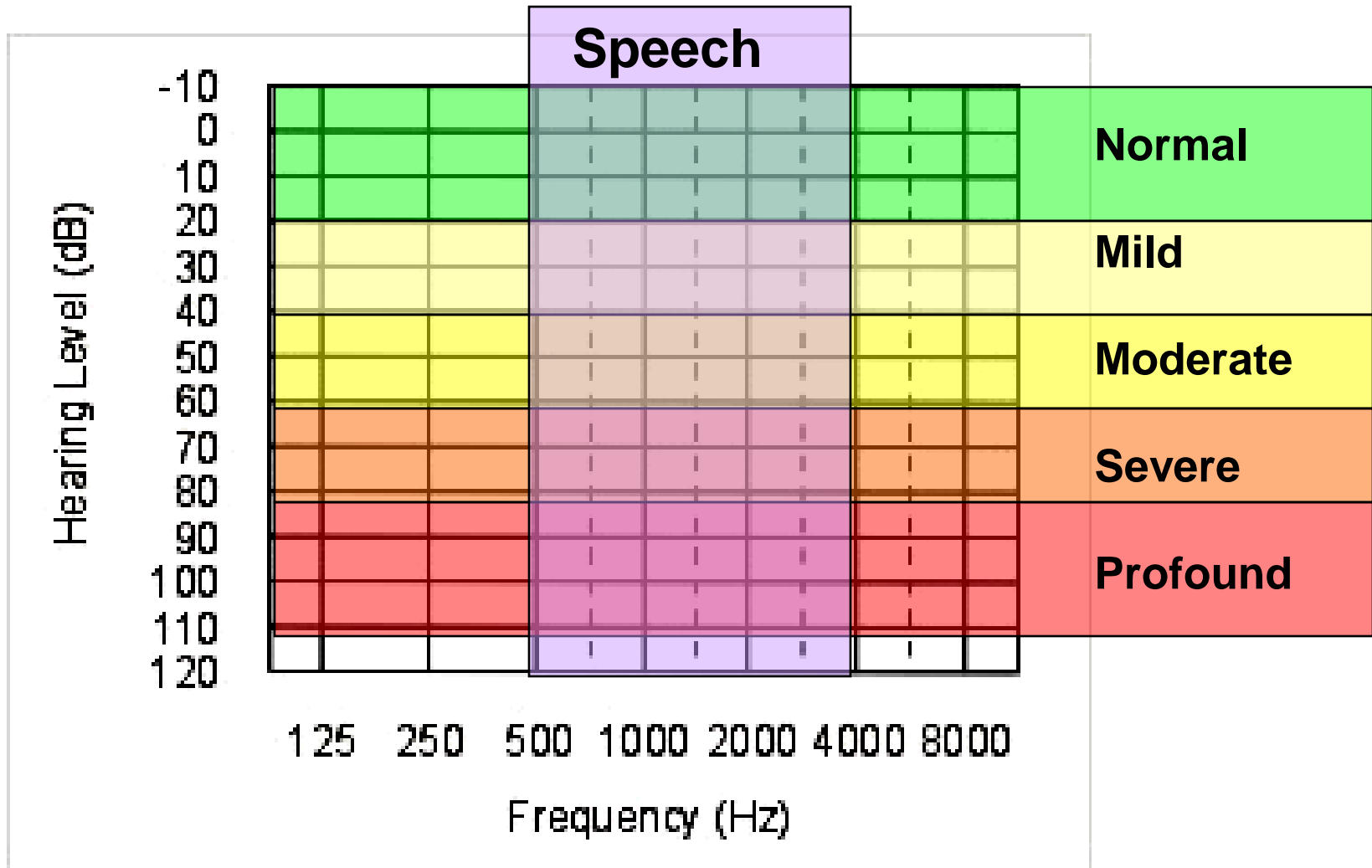
Audiogram



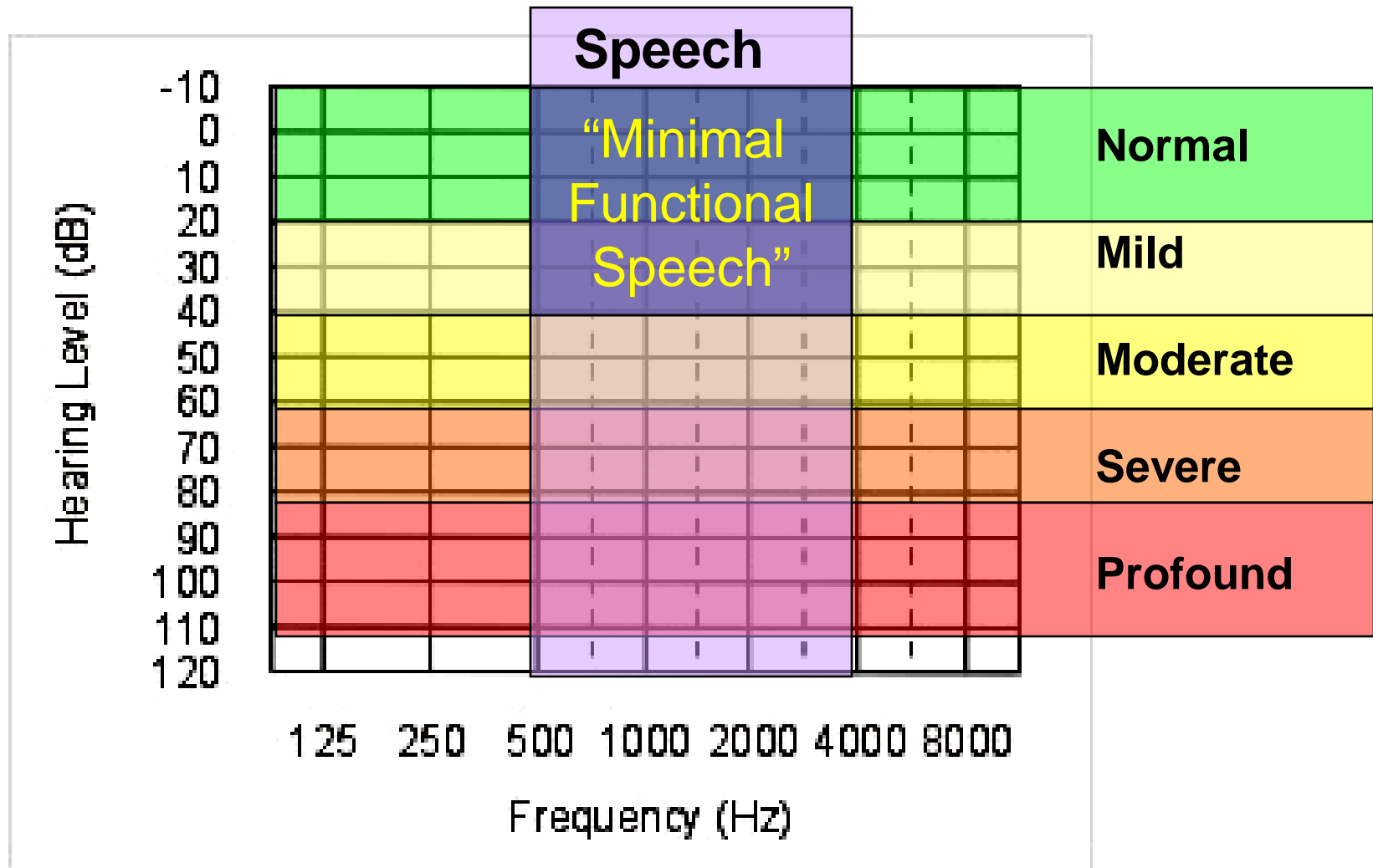
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Audiogram



Audiogram



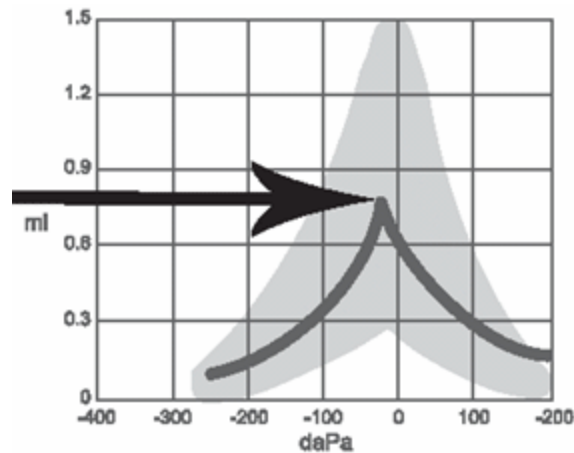
Speech Audiometry

- Read a standard word list and score by Percentage correct.

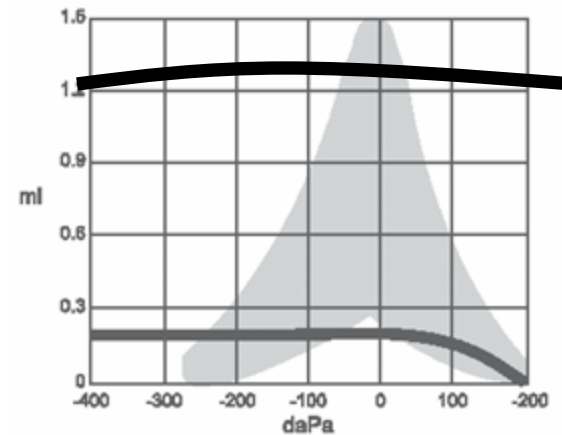


Tympanometry

- Normal



- Type B



Hole

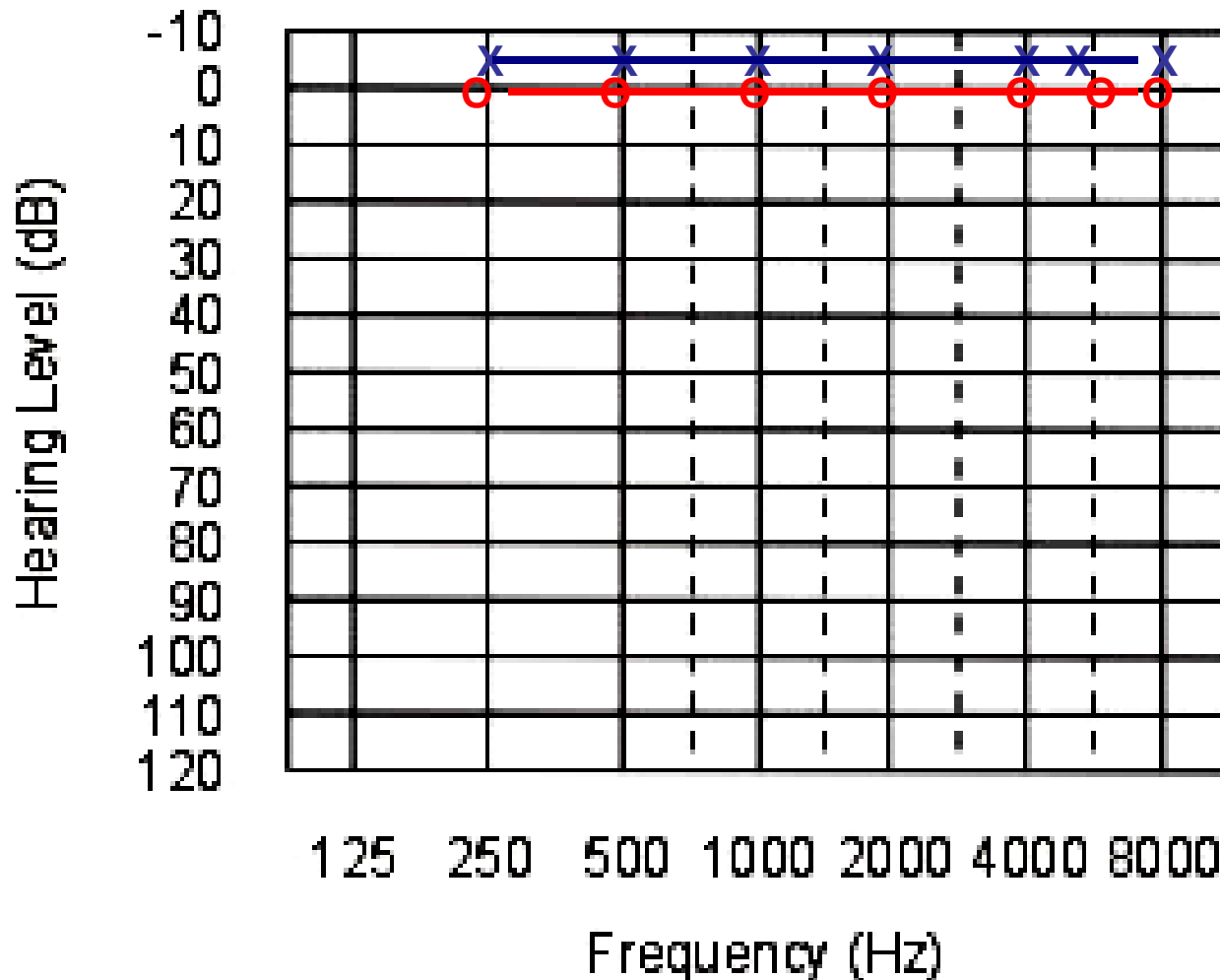
Fluid



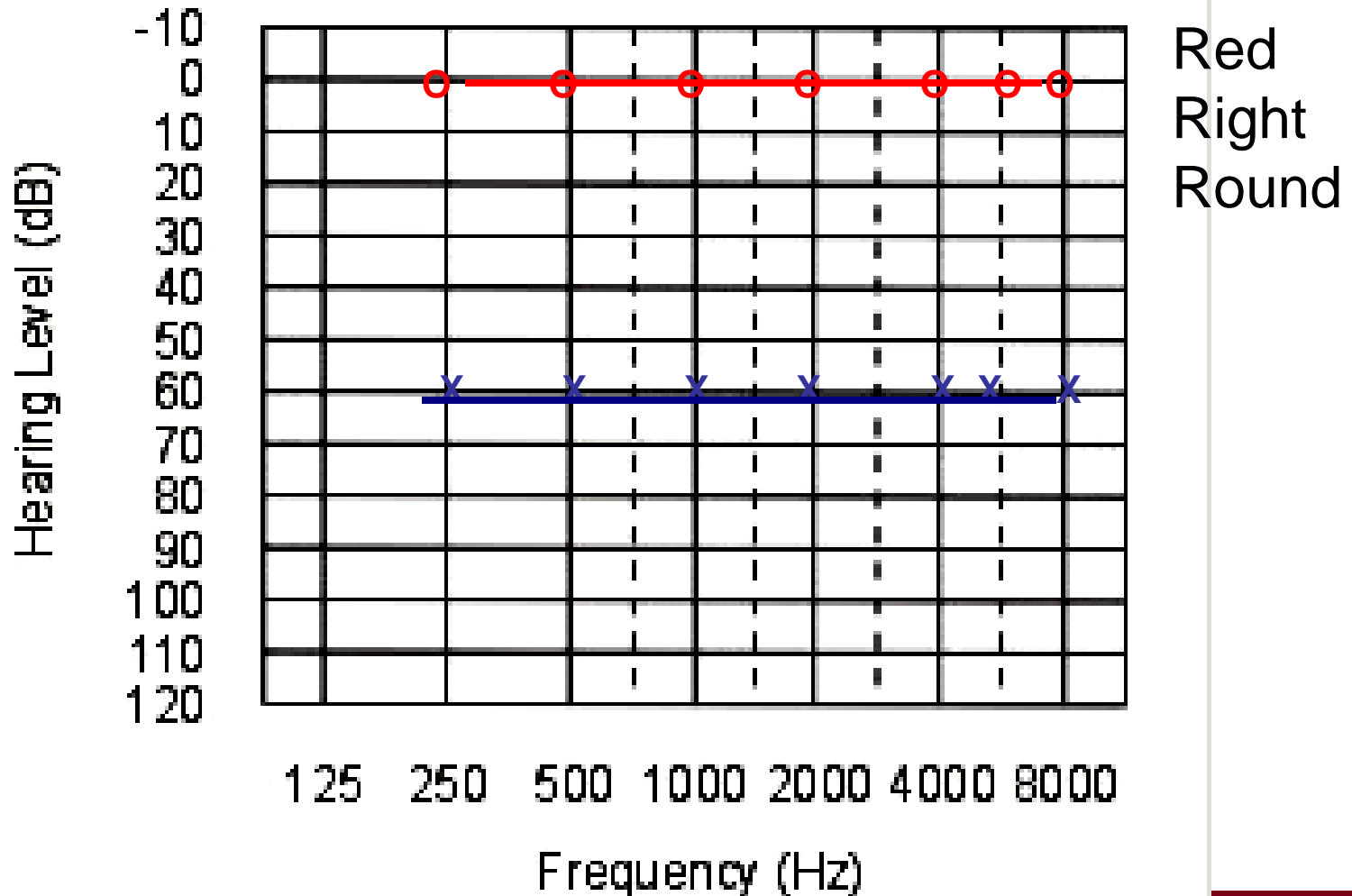
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Typical Normal Audiogram



Left severe hearing loss



Ear Abnormalities in Fanconi Anemia

- Approximately 15% with “ear” abnormality (Giampietro, 1993)
- Hearing loss in 17% of patients (Santos 2002)
 - 100% of the time conductive hearing loss
 - Fluid
 - Ear canal abnormalities
 - Ear bone abnormalities



Ear Abnormalities in Fanconi Anemia

- 53% hearing loss (Kim 2006)
- Mostly conductive and mild but some with Sensorineural hearing loss.



Treatment options

- Preferential classroom placement
- Assisted listening devices in the classroom
- Surgery for the conductive loss and >mild.
- Surgery for ear canal narrowing or absence
- Surgery for ear cosmetic abnormalities

- *Otolaryngologist, Neuro otologist, Plastic Surgery*



Hearing Monitoring

- In the classroom situation by school screening
- Formal audiometry early to determine normalcy of hearing
- Audiometry if any change noticed by parents at baseline.



Popular Demand- Oral cancer screening

- University of Minnesota oral cancer screening program
- Follow Fanconi patients for yearly exams for the past 3 years.
- No early cancers discovered in young patients < 18 years
- No precancerous lesions detected (approximately 30-40 patients).
- 2 Cancers in adults



Screening Methods

- Physical exam (MD or Dentist)
- Visilite
- Toluidine blue
- Visilite



Physical Exam Cancer Screening

- Gold standard
- Inspection and palpation of oral cavity and neck
- Possible use of Flexible laryngoscopy in older patients or symptomatic patients



Vizilite Cancer screening

- Combination of light and telenium chloride, assisted exam.
- Primarily through dental clinics
- Sensitive for any oral abnormality (cold sore, bite, cancer, etc.)
- Not shown superior to oral exam



Vizilite



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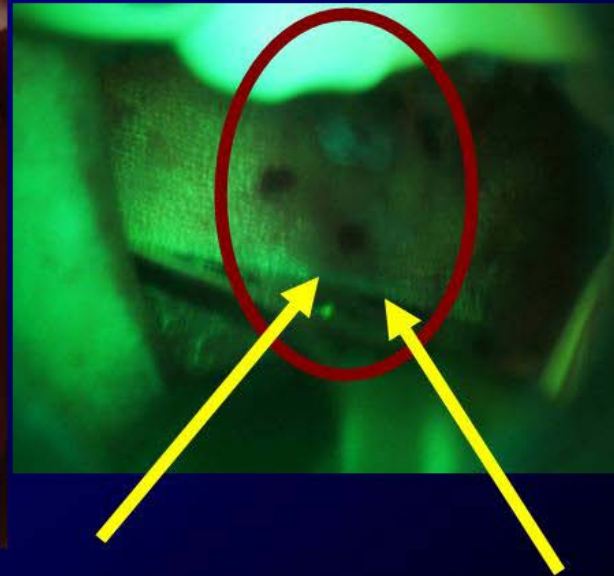
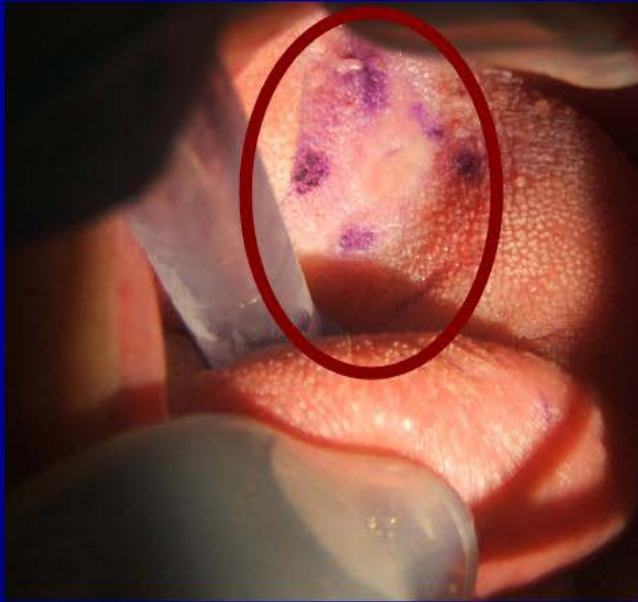
Velscope

- Normal tissues autofluoresce
- Abnormal tissues do not
- Therefore autofluorescent **NEGATIVE** areas might be suspicious for dysplasia or cancer
- Not shown to be better than physical exam



Velscope- surgical margins

Velscope- Pre/post



Questions On Hearing Loss or Oral Cancer Screening?



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