





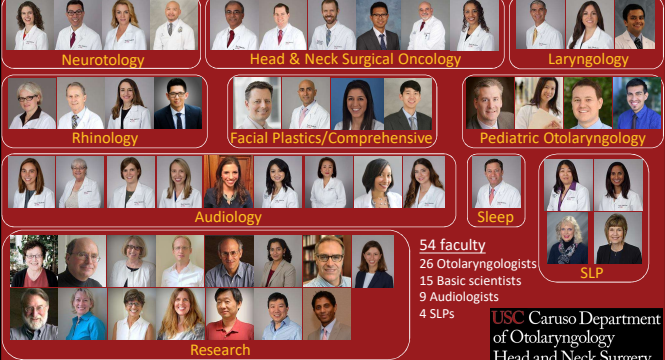
Sensorineural Hearing Loss: How, Why, Where?

John S. Oghalai, MD
 Leon J. Tiber and David S. Alpert Professor and Chair
 Caruso Department of Otolaryngology – Head and Neck Surgery
 Keck School of Medicine of USC
 University of Southern California
 4/10/2020

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Neurotology **Head & Neck Surgical Oncology** **Laryngology**
Rhinology **Facial Plastics/Comprehensive** **Pediatric Otolaryngology**
Audiology **Sleep** **SLP**
Research

54 faculty
 26 Otolaryngologists
 15 Basic scientists
 9 Audiologists
 4 SLPs

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Kubler-Ross stages of grief as they pertain to otolaryngology residents and COVID-19

- **Denial:** *This virus won't affect me. I'm young. Anyway, I'm pretty sure I had COVID a month ago.*
- **Anger:** *It isn't fair that you're canceling surgery and clinics. My patients need me and I need to complete a minimum number of cases to graduate.*
- **Bargaining:** *Okay, if we get preop COVID testing and I wear proper PPE, then can I operate?*
- **Depression:** *I don't know when this will end. This is really dragging on.*
- **Acceptance:** *This is happening and every resident is affected, but there are ways to proceed. I can do telemedicine; I can see some patients in person with proper PPE; I can do some surgeries; I can participate in the online classes.*

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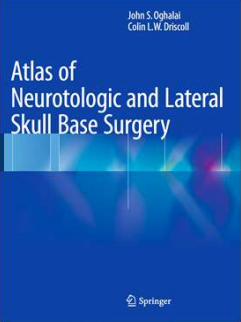
Learning objectives

- Improve your understanding of cochlear anatomy
- Learn the basic mechanisms of sensorineural hearing loss
- Be able to tell a patient what a hearing aid does, why it can help in some situations, and why it can't help much in other situations
- Basically, to understand the disease at a deeper level than the standard:
 - Patient notices hearing loss and makes an appointment to see you
 - You document the hearing loss with an audiogram
 - You tell them they have sensorineural hearing loss
 - You tell them to get hearing aids

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Disclosures

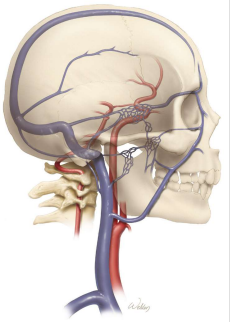


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Illustrations by Scott Weldon



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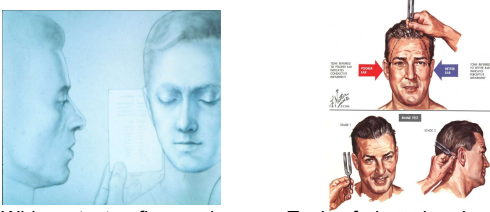
Diagnosing sensorineural hearing loss



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Office Evaluation of Hearing



Whisper test or finger rub:
Simple test of air conduction

Tuning forks or head scratch:
Conductive vs sensorineural

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What To Do When a Patient Presents with Hearing Loss

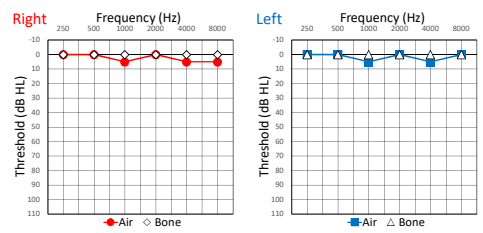
- Adult with progressive hearing loss
 - Refer to any otolaryngologist or audiologist
- Child with speech delay or possible hearing loss
 - Refer to a pediatric otolaryngologist or pediatric audiologist
- Sudden hearing loss is an emergency
 - Same day appointment with Otolaryngology
 - Oral or intratympanic steroids



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Normal Hearing

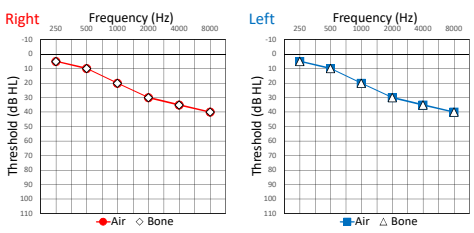


Word Recognition Score
R = 100%
L = 100%

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Mild Sensorineural Hearing Loss

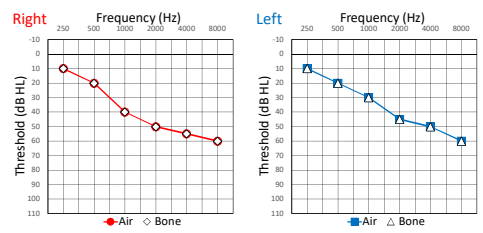


Word Recognition Score
R = 92%
L = 88%

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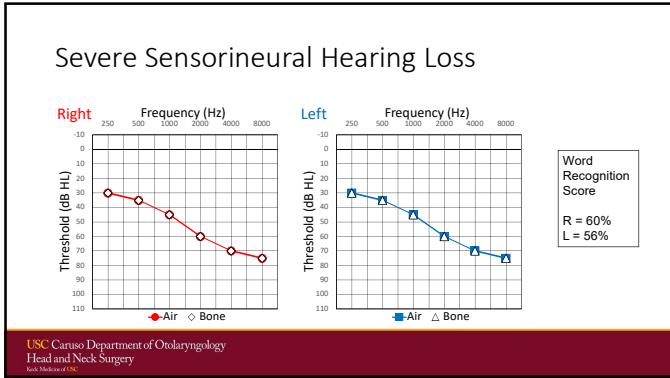
Moderate Sensorineural Hearing Loss



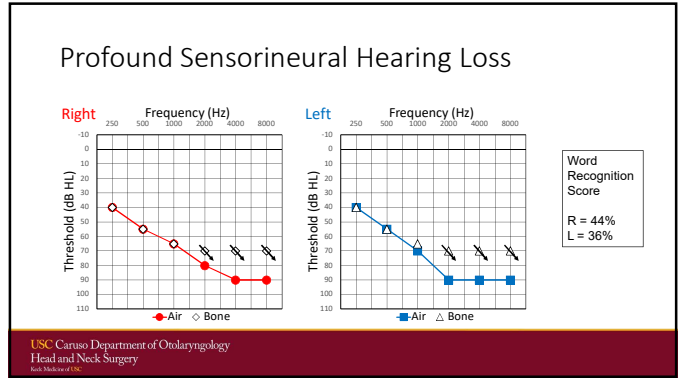
Word Recognition Score
R = 72%
L = 76%

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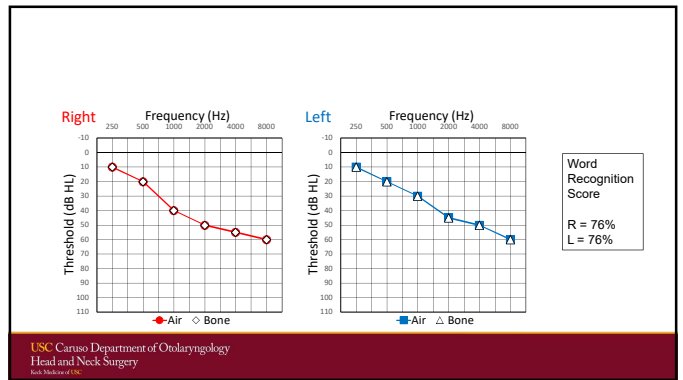
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Case Presentation

- A 72 yo male presents for evaluation of hearing loss. His new wife complains that he turns up the TV too loud. He says people have been telling him this for years, but he doesn't feel like anything has changed. No noise exposure.
- Family history negative.
- PMH/PSH negative.

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What is the next step? (select all that are correct)

1. Order a CT scan
2. Order an MRI scan
3. Order a TSH
4. Order an autoimmune panel
5. Order genetic testing
6. Tell him he has "nerve damage" and sell him digital hearing aids

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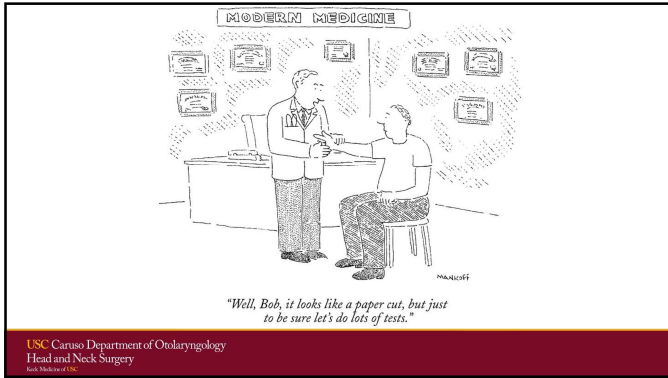
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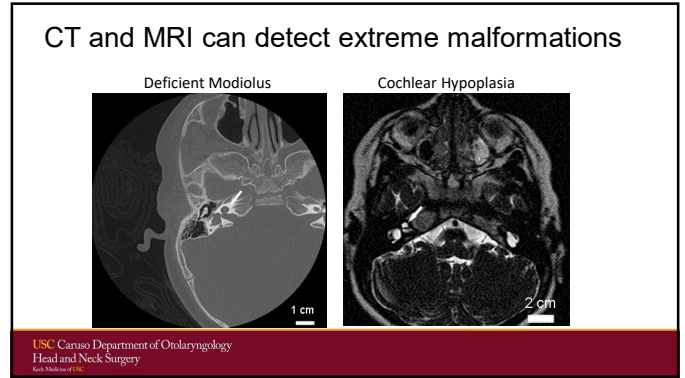
None of the answers are correct

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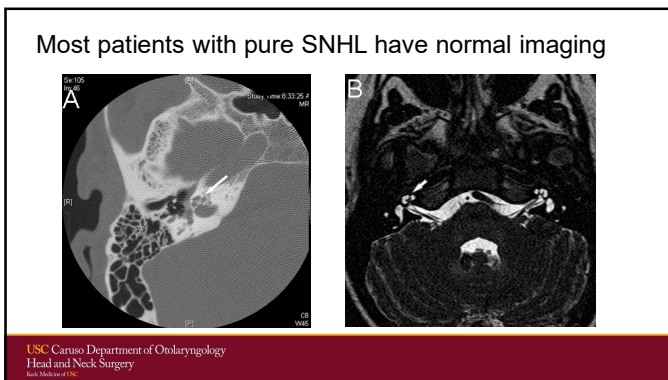
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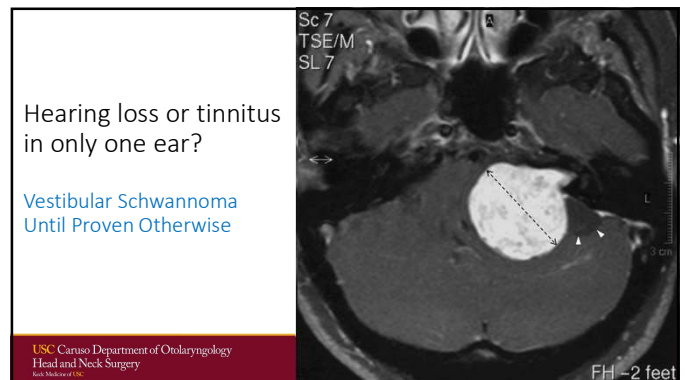
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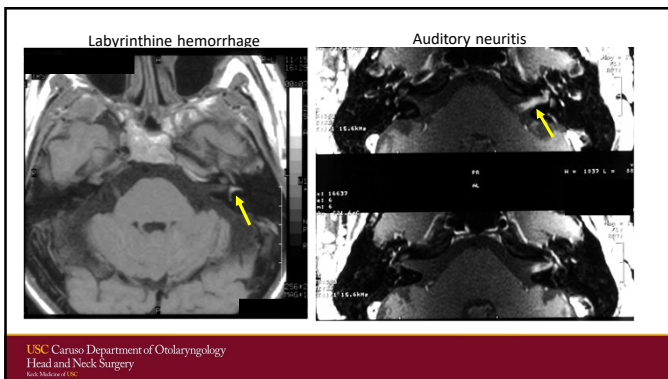
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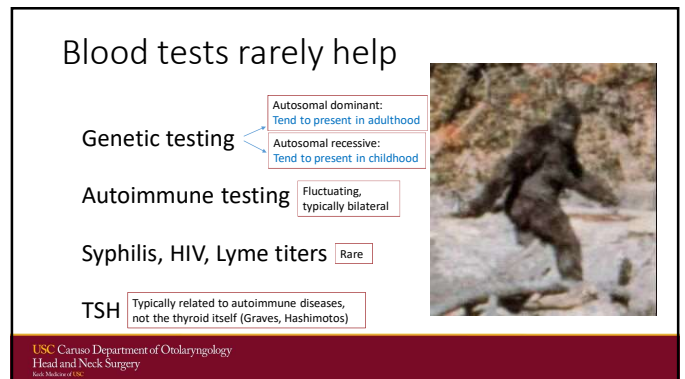
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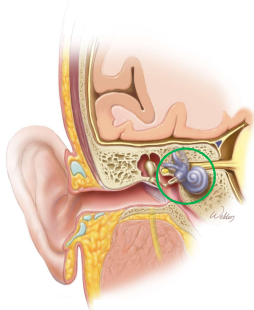
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How do we hear?



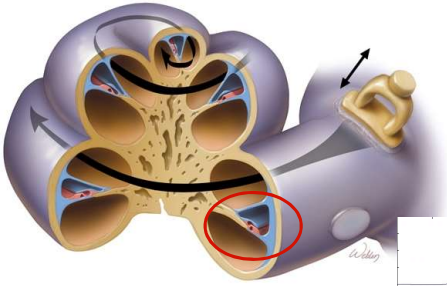
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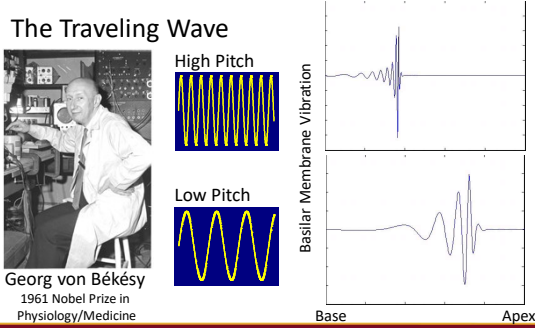
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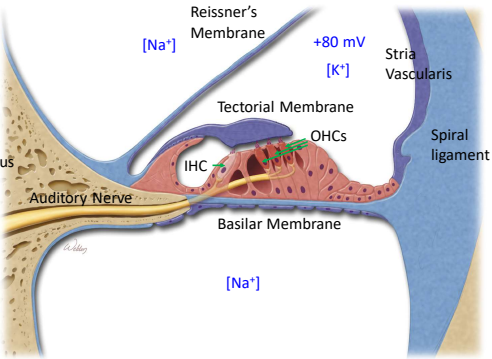
The Traveling Wave



Georg von Békésy
1961 Nobel Prize in Physiology/Medicine

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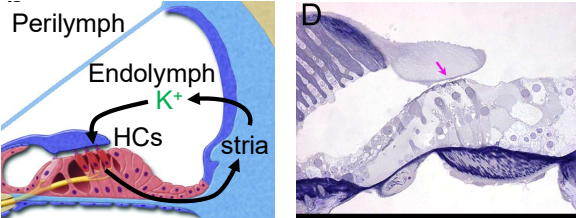


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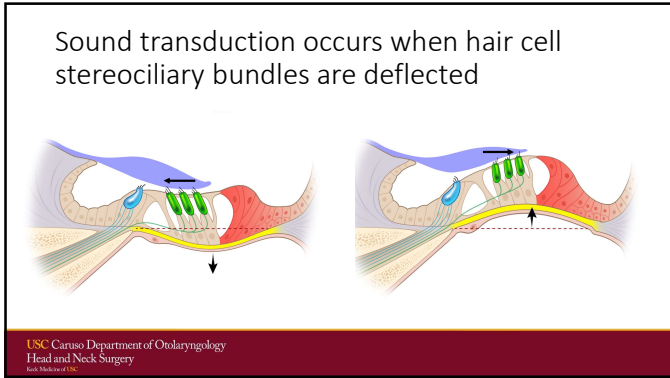
K⁺ recycling:

- Secretion by stria vascularis
- Uptake by hair cells through mechano-electrical transduction channels
- Transport through supporting cells back to the stria

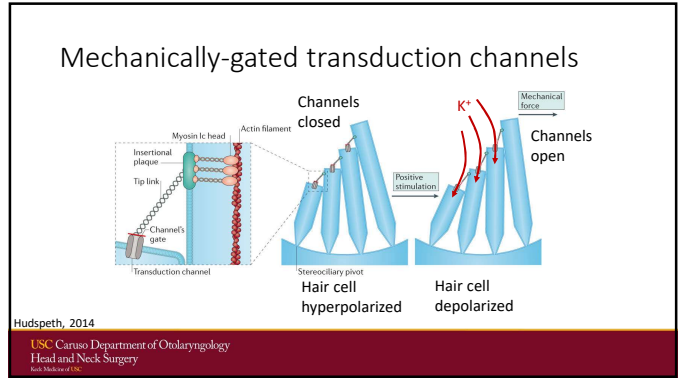


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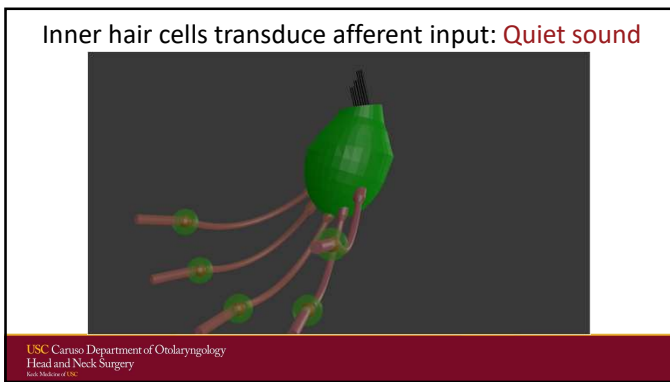
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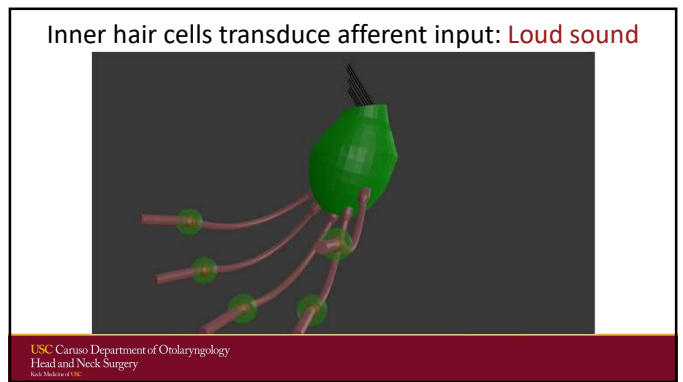
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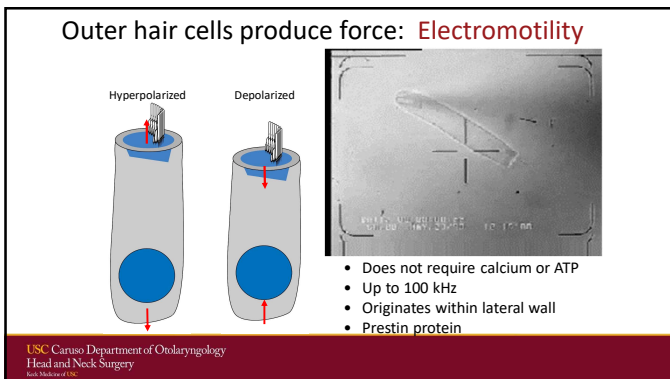
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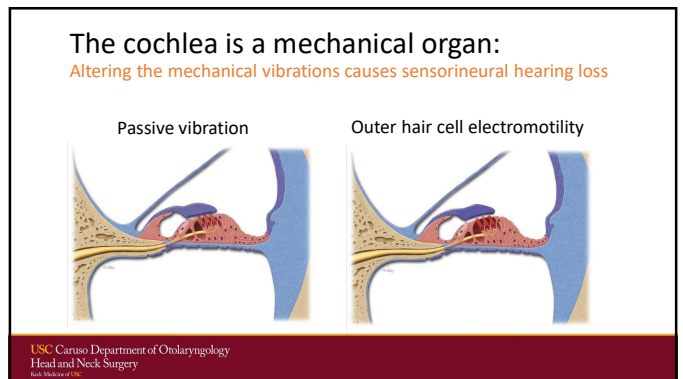
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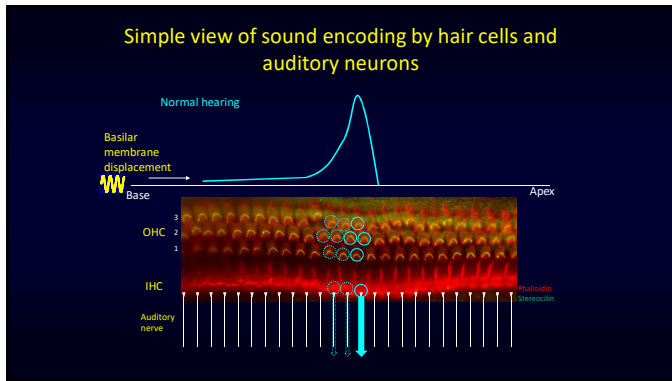
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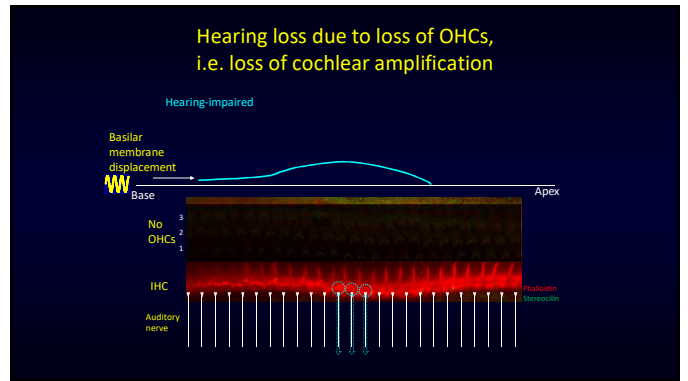
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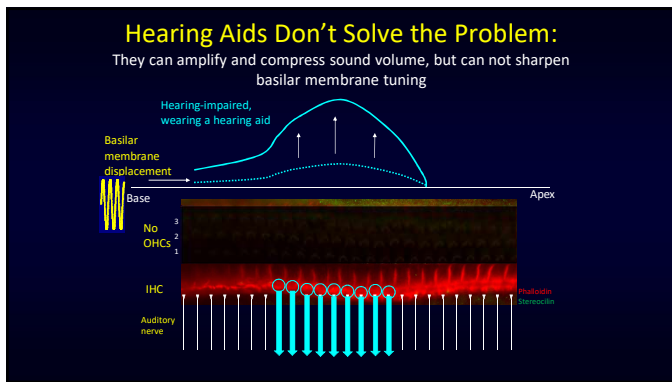
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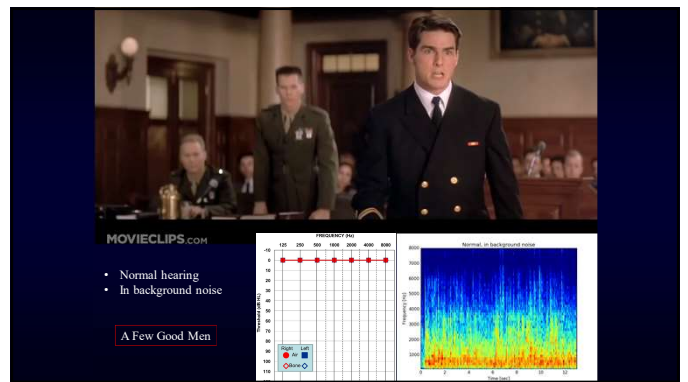
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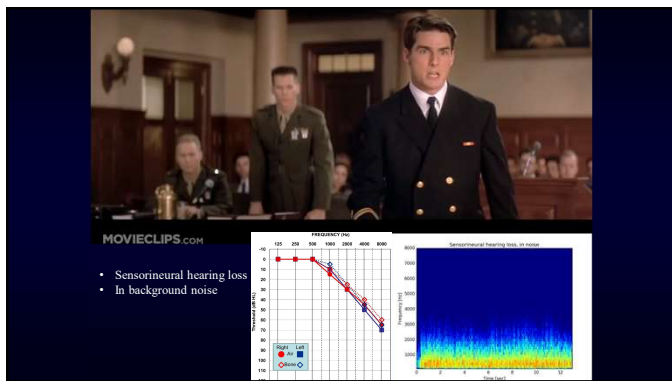
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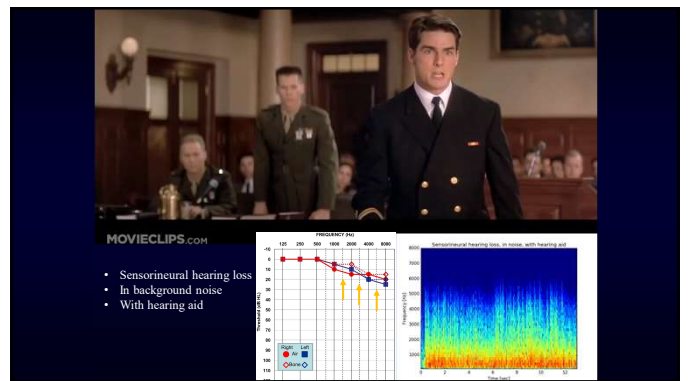
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Question:

Select all the correct statements

1. Hair cell mechano-electrical transduction means converting sound vibrations into voltage changes.
2. Inner hair cells transduce the traveling wave and convey the majority of the sound signal to the afferent auditory neurons.
3. Outer hair cells transduce the traveling wave and convey the majority of the sound signal to the afferent auditory neurons.
4. Inner hair cells amplify and sharpen the traveling wave using electromotility.
5. Outer hair cells amplify and sharpen the traveling wave using electromotility.
6. Hearing aids help by sharpening the tuning curve and improving frequency discrimination

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Question:

Select all the correct statements

- ★ 1. Hair cell mechano-electrical transduction means converting sound vibrations into voltage changes.
- ★ 2. Inner hair cells transduce the traveling wave and convey the majority of the sound signal to the afferent auditory neurons.
- 3. Outer hair cells transduce the traveling wave and convey the majority of the sound signal to the afferent auditory neurons.
- 4. Inner hair cells amplify and sharpen the traveling wave using electromotility.
- ★ 5. Outer hair cells amplify and sharpen the traveling wave using electromotility.
- 6. Hearing aids help by sharpening the tuning curve and improving frequency discrimination

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Pathophysiology of sensorineural hearing loss



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What causes sensorineural hearing loss?

- Aging
 - Noise exposure
- } Common
-
- Tumors
 - Stroke to the inner ear
 - Ototoxic drug exposure
 - Autoimmune disease
 - Meningitis
 - Otosclerosis
- } Rare

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We all develop sensorineural hearing loss

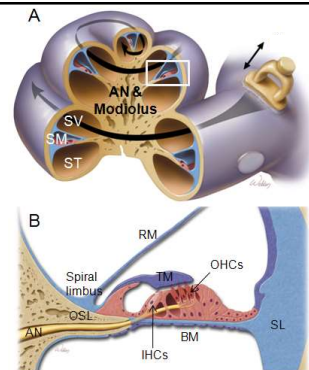
- Age > 65 years
 - 2/3 people have hearing loss that affects them
- United States
 - 30 million with hearing loss
 - 2 million deaf

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Common Causes of Hearing Loss:

- Alter the Soft Tissue Structures
- For example:
 - Hair cell loss
 - Auditory nerve loss
 - Tectorial membrane mutations
 - Gap junction mutations
 - Atherosclerosis within stria vascularis
- CT/MRI don't have the resolution to detect these problems



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Stereociliary trauma is found immediately after loud noise

Control
Blast-exposed

2 μm

Nicolas Grillet

USC Caruso Department of Otolaryngology
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Noise causes loss of hair cells in the base

7d after blast

a Apex a' Middle a'' Base

1 IHC
2 OHC
3 Phalloidin Prestin Myosin VIIa

Low frequencies Middle frequencies High frequencies

Jinkyung Kim

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Visualizing endolymph within scala media

SV SM TM
RM ST BM

SV SM ST

Kim et al, PNAS, 2018

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Time lapse OCT imaging in the mouse cochlea

Noise-damaged stereocilia causes K⁺ build up in endolymph No secretion of K⁺ from stria vascularis

control blast dead

Perilymph Endolymph HCs cells
Perilymph Endolymph HCs cells

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Noise also causes loss of synaptic connections with the auditory nerve (cochlear synaptopathy)

Normal Noise-exposed

Ido Badash
Juemei Wang

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Case Presentation: Same patient as before.
A 72 year-old male has mild-moderate SNHL and WRS 76% in each ear. What are the most likely source of his problem? (select all that are correct)

1. He is missing mostly inner hair cells, and so the afferent signal is reduced.
2. He is missing outer hair cells, and so cochlear amplification is reduced.
3. His stria stopped secreting K⁺, and so cochlear amplification is reduced.
4. He has auditory nerve loss, and so the afferent signal is reduced.

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Case Presentation: Same patient as before.
 A 72 year-old male has mild-moderate SNHL and WRS 76% in each ear. What are the most likely source of his problem? (select all that are correct)

- ★ 1. He is missing some inner hair cells, and so the afferent signal is reduced.
- ★ 2. He is missing outer hair cells, and so cochlear amplification is reduced.
- ★ 3. His stria is secreting less K⁺, and so cochlear amplification is reduced.
- ★ 4. He has auditory nerve loss, and so the afferent signal is reduced.

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Treating sensorineural hearing loss




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Impact of Hearing Loss

- Reduced social activity, isolation
- Depression
- Anxiety; Insecurity
- Children
 - Speech delays
 - Lower IQ
 - Lower income
- Adults
 - Untreated mild hearing loss: 2X risk of dementia
 - Untreated severe hearing loss: 5X risk of dementia

Healthy Hearing
 =
 Healthy Brain



Gurgel et al., 2014; Thomson, Auduong, Miller and Gurgel, 2017; Panza et al., 2018;

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Hearing Aids:

- Make sounds louder
- Do not make speech clearer
- Some models help with background noise
- Tinnitus masking



Cochlear Implants:

- Stimulate auditory nerve directly
- Make speech clearer
- Not ideal for music
- Tinnitus usually improved

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Hearing Aids


- Daily wear vs. extended wear
- 45 day trial (California state law)
- Technology improvements
 - Bluetooth connectivity
 - Find my hearing aid
 - Directional microphones
 - Background noise suppression

Big box model	vs.	Medical model
Cheaper		Wider range of technology
Convenient		Better programming
		Validation of fit
		Custom earmolds
		Physician involvement


In the canal (ITC)




In the ear (ITE)



Behind the ear (BTE)





Behind the ear (BTE) with a custom mold



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Deep insertion, long-term wear hearing aids





- Tinnitus management
- Might have a more natural sound
- Analog

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1998

2018

OTC devices:
Caveat Emptor

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Cochlear Implants

- Not natural hearing
- Expanding criteria because they work so well

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Outcomes after Cochlear Implantation

- Improved emotional health
- Reduced isolation
- Expanded options in education and work
- Improved cognition

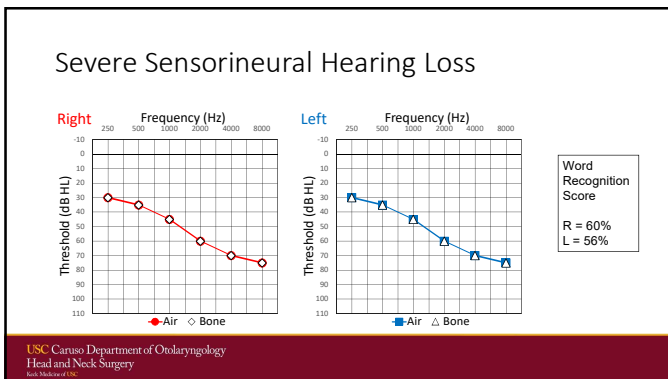
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Case presentation

- 74 year-old retired mechanical engineer
- Has noticed trouble understanding what people are saying
- Particularly bad in the presence of background noise
- Tried hearing aids about 3 years ago, but gave up
 - Uncomfortable
 - Feedback
 - Didn't help him in public
 - Didn't help him hear the TV better

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What is the next step? (select all that are correct)

1. Try to fix his current hearing aids
2. Sell him a more expensive set of hearing aids that has more bells and whistles
3. Ask him to try using a headphone when watching the TV
4. Order a cochlear implant evaluation
5. Tell him this is a normal part of aging

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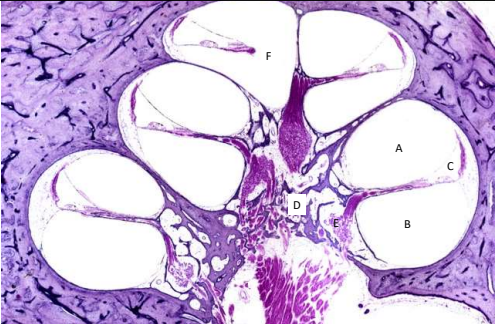
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- ? 4. Order a cochlear implant evaluation
- ★ 5. Tell him this is a normal part of aging

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Name the structures

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100 μm

Name the structures

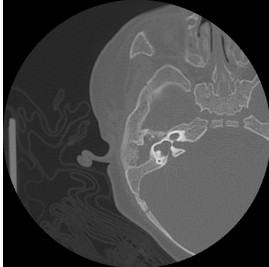


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Case Presentation:

What should you be concerned about when performing cochlear implantation?



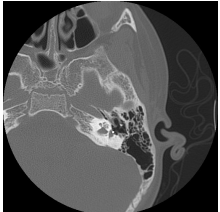
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Case Presentation:

9 year old boy had normal bone line 3 months after temporal bone fracture. TORP placement performed for incus/stapes fracture. Had 5-10 dB ABG 2 months after surgery. At routine 1 year f/u had profound SNHL.

What is the diagnosis?

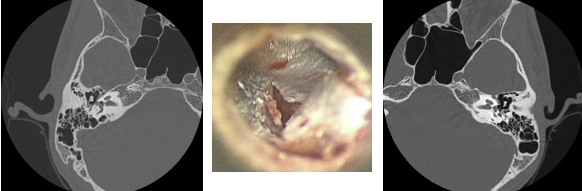


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Case Presentation:

16 year old male who had a stick jabbed in his right ear. Has dizziness, a 55 dB ABG, and a normal bone line.



What is the diagnosis?

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Case Presentation:

What should you advise this patient?

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Case Presentation

A 3 year old girl has bilateral progressive hearing loss. The left ear was implanted 1 year ago and the child is making excellent progress. What is the next step?

1. Use the cochlear implant in the left ear alone.
2. Use the cochlear implant in the left ear and a hearing aid in the right ear.
3. Put another implant in the right ear.
4. Save the right ear for stem cell therapy.

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Thank you and stay safe!

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