Anatomy & Physiology of the Auditory System

- Gonzalo Otazu, PhD
- Biomedical Sciences

Session objectives

- Students will be able to describe the anatomy of the outer-, middle-, and inner ear including the structures located therein.
- Students will be able to describe the basic principles of mechanotransduction found in the inner ear.
- Students will be able to describe the anatomy auditory pathway beginning from the inner ear to the primary auditory cortex.
- Students will be able to describe the basic physiology of the nuclei of the central auditory as well as that of the primary auditory cortex.
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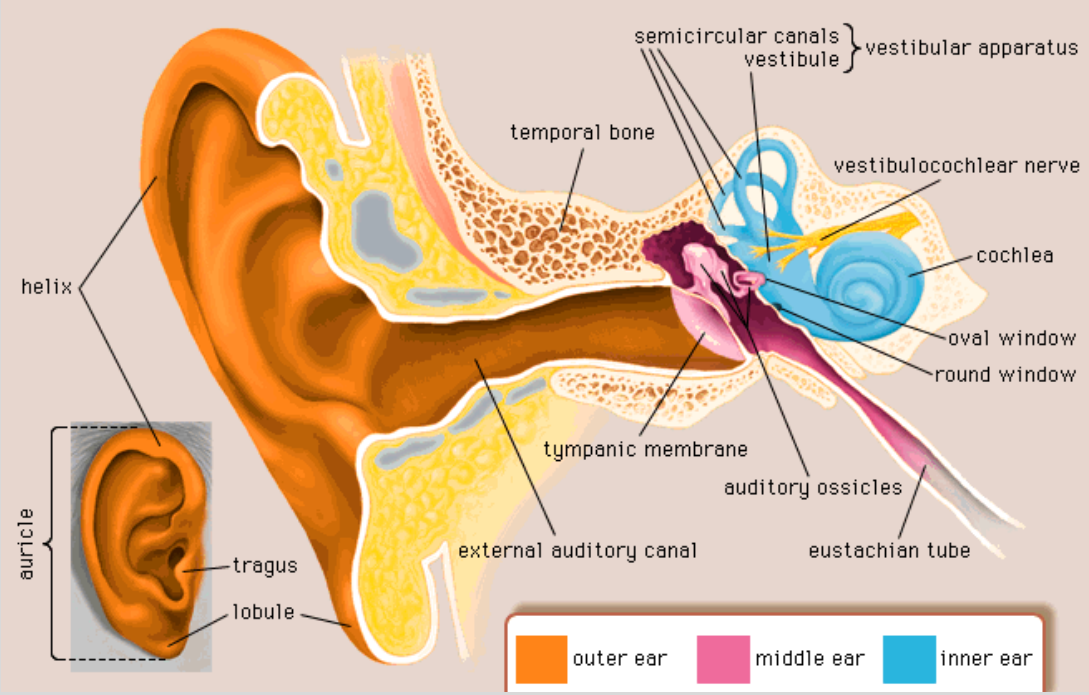
Auditory anatomy



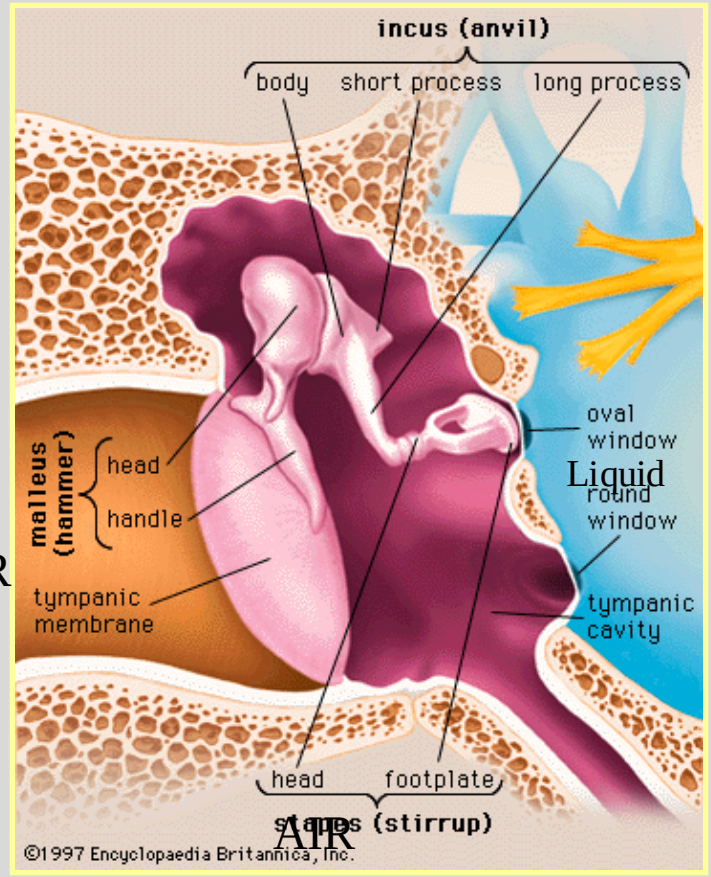
- Outer ear serves to efficiently capture the sound pressure waves and channel into into the middle ear (air transmission).
- In the middle ear, air vibrations of the tympanic membrane and converted into vibrations of the auditory ossicles (solid transmission).
- In the inner ear, the solid vibrations are converted to waves propagating in liquid and converted into nerve impulses.
- Sound pressure of barely audible sound is 20µPa (micro-Pascals) which is 10^8 times weaker than atmospheric pressure.

Middle Ear: Tympanic Membrane & Ossicles

Bones of the middle ear:

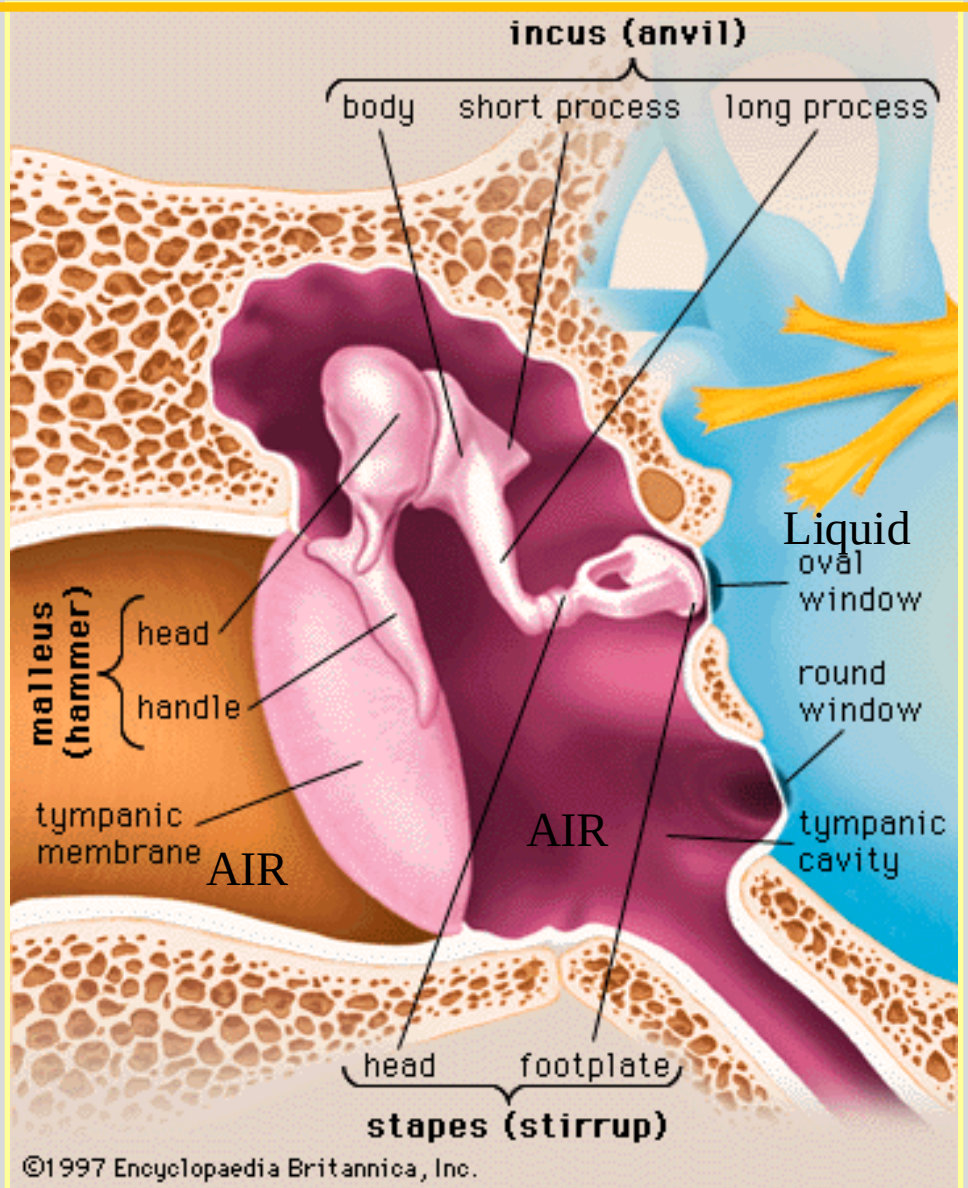
Malleus – hammer
Incus – anvil
Stapes – stirrup

•Transmit the air vibrations hitting the tympanic membrane to the oval window.
•Smallest bones of the human body.
•Malleus couples to the tympanic membrane.
•Incus transmits the vibration to the stapes.
•The stapes is coupled to the oval window.
•Although the transmission of the vibration in the middle ear is through the ossicles (solid), the tympanic cavity is filled with air and is connected to the nasopharynx through the eustachian tube.



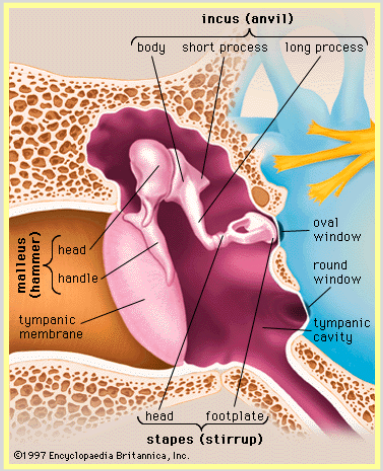
Middle Ear: convert air waves into liquid waves

- Air waves do no penetrate well into liquids.
- Liquids are less compressible than air and higher pressures are required for similar displacements.
- Smaller area of oval window compared to tympanic membrane increases the pressure in the cochlea.

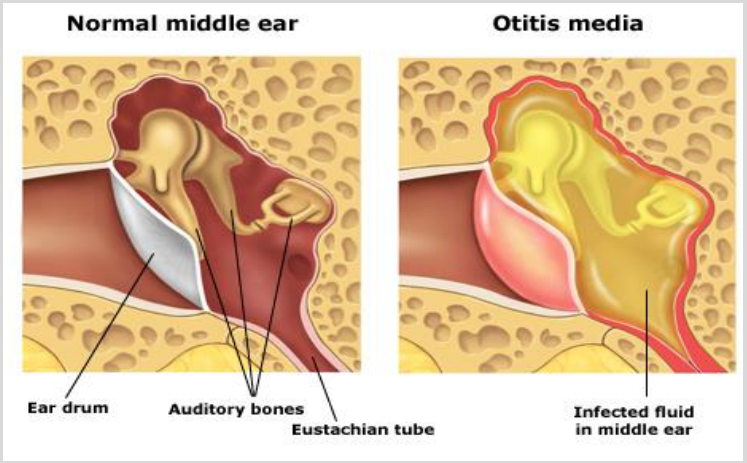


Middle Ear: Conductive hearing losses

- Conditions that affect middle ear conduction
  - Damage to the tympanic membrane
  - Fluid in the tympanic cavity
  - Damage to the ossicles

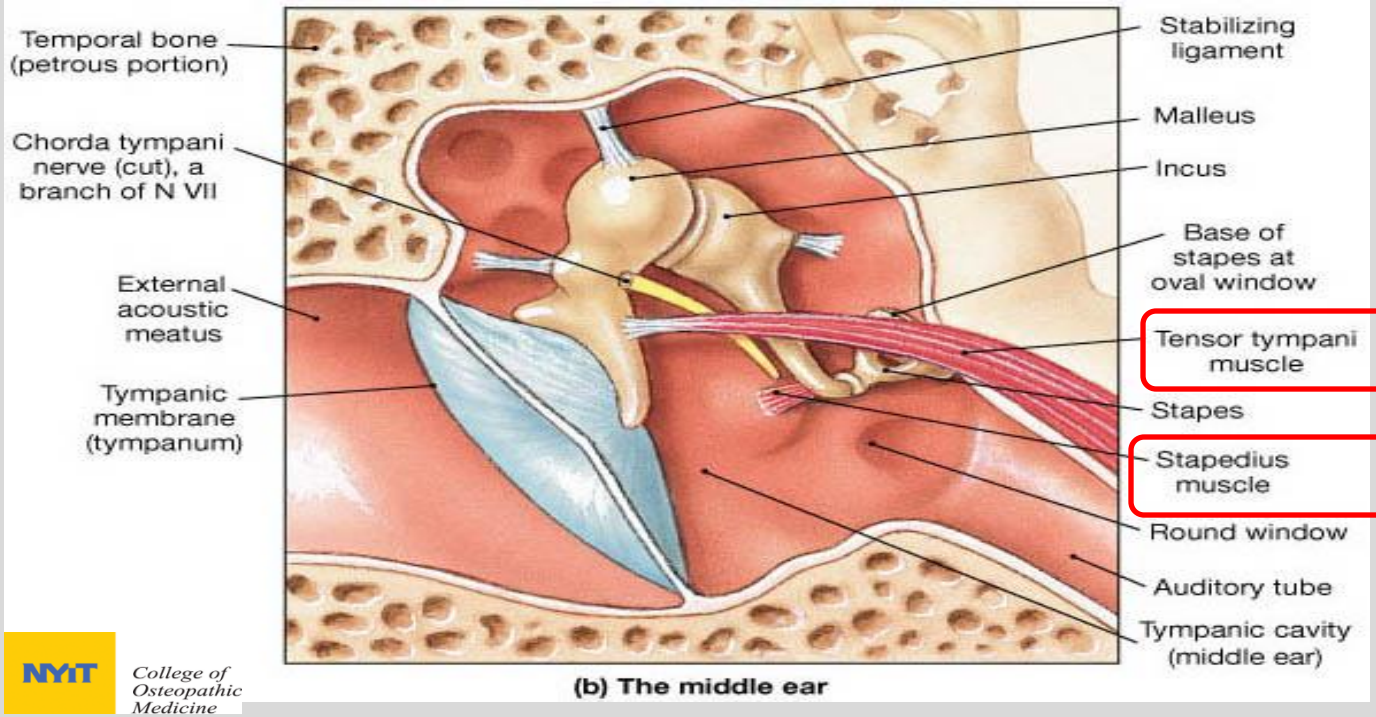


- Perforation of the tympanic membrane equalizes the pressure on both sides, so there is no pressure difference and the tympanic membrane will not vibrate.
- The sound wave applies directly to the oval window without amplification by the ossicles, so there a significant attenuation of the input to the cochlea.



In otitis media, the tympanic cavity fills with fluid and attenuates the vibrations of the ossicles and the tympanic membrane

Muscles of the middle ear



- Contractions of the muscles of the middle ear attenuates the propagation of sound through the middle ear and protect the delicate hair cells in the inner ear.
- Tensor timpani muscle decouples the malleus from the tympanic membrane and is innervated by the trigeminal nerve.
- Stapedius muscle decouples the stapes from the oval window.
- Both muscles are part of the acoustic reflex to protect the inner ear from loud sounds.
- Sudden loud sounds can still damage the hair cells.

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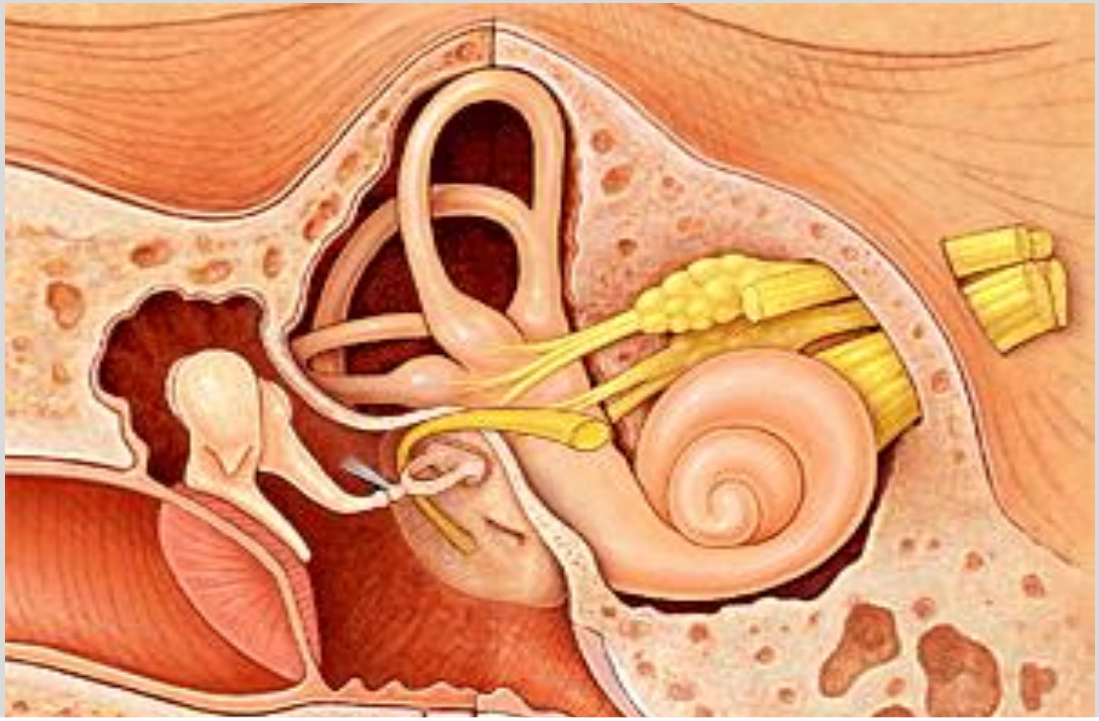
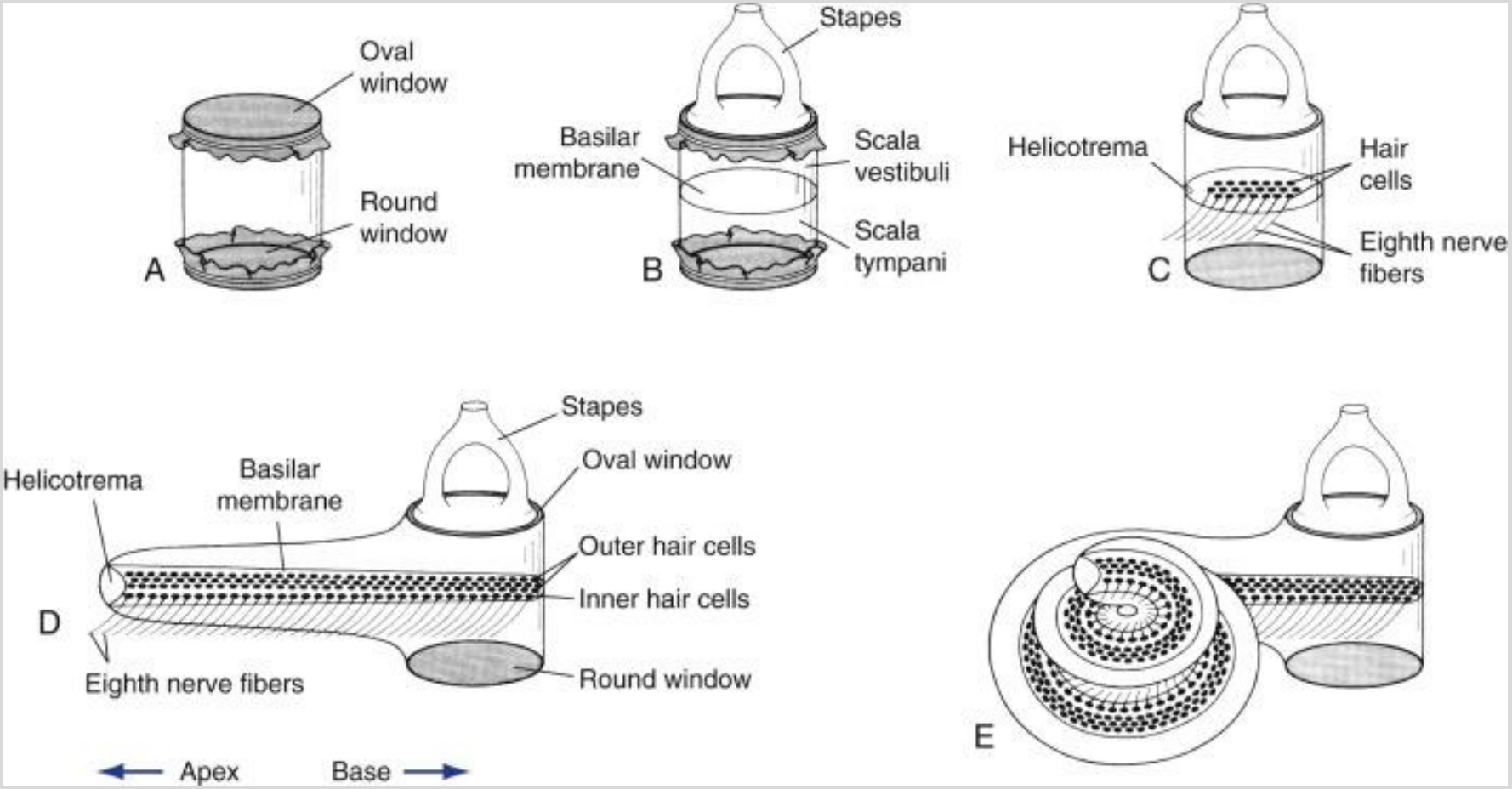
Inner ear - cochlea

- Located in the petrous portion of temporal bone.
- 2.5X coils
- Decreasing diameters w/ each coil.
- About the size of a chickpea.



The cochlea decomposes vibration into its frequency components

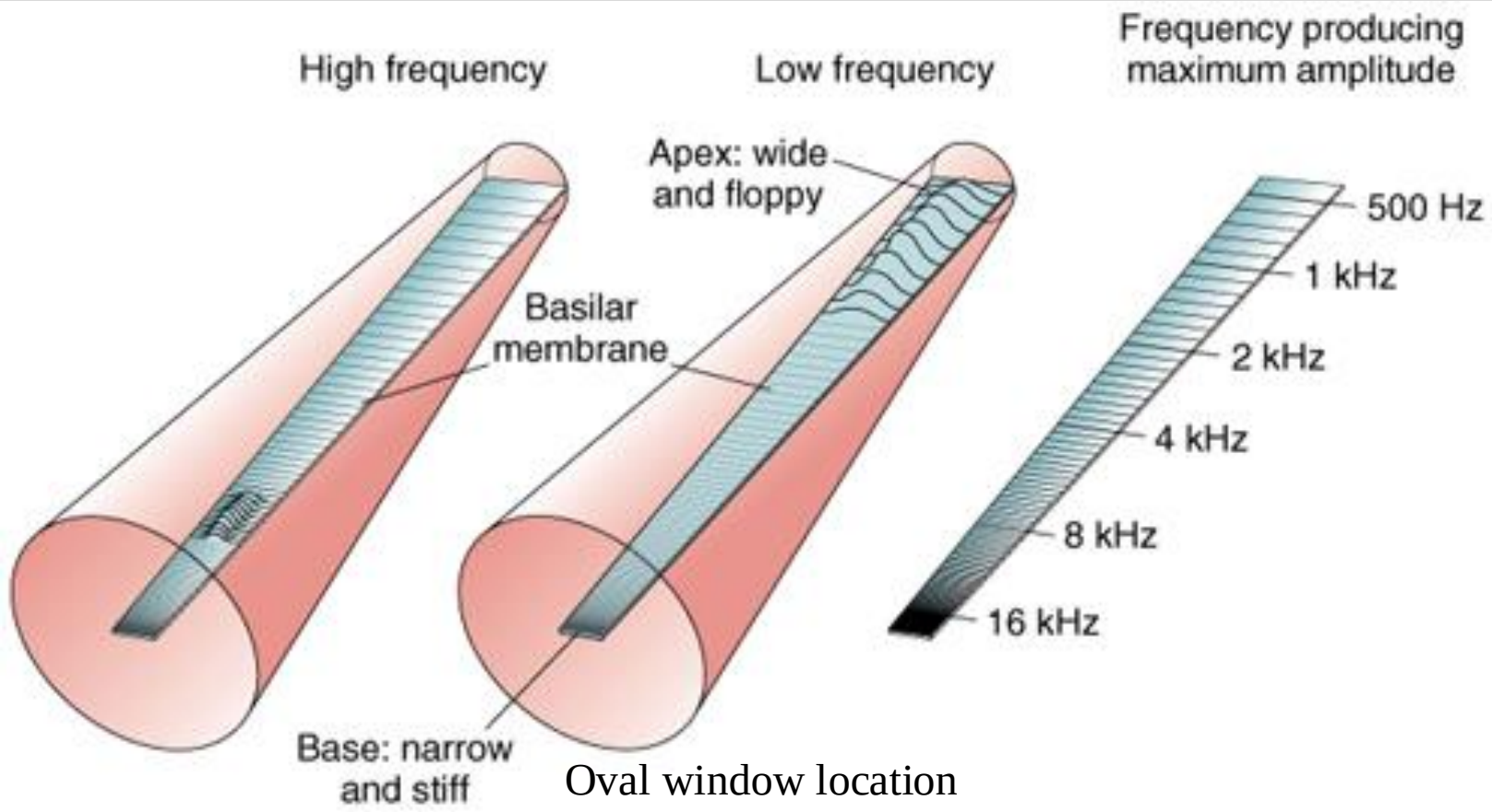


Physiological differences between the apex vs. base of cochlea

-basilar membrane is relatively wide, thin & floppy at the apex of the cochlea but narrow, thicker & more taut toward the base.

Base responds to high frequencies and apex responds to lower frequencies

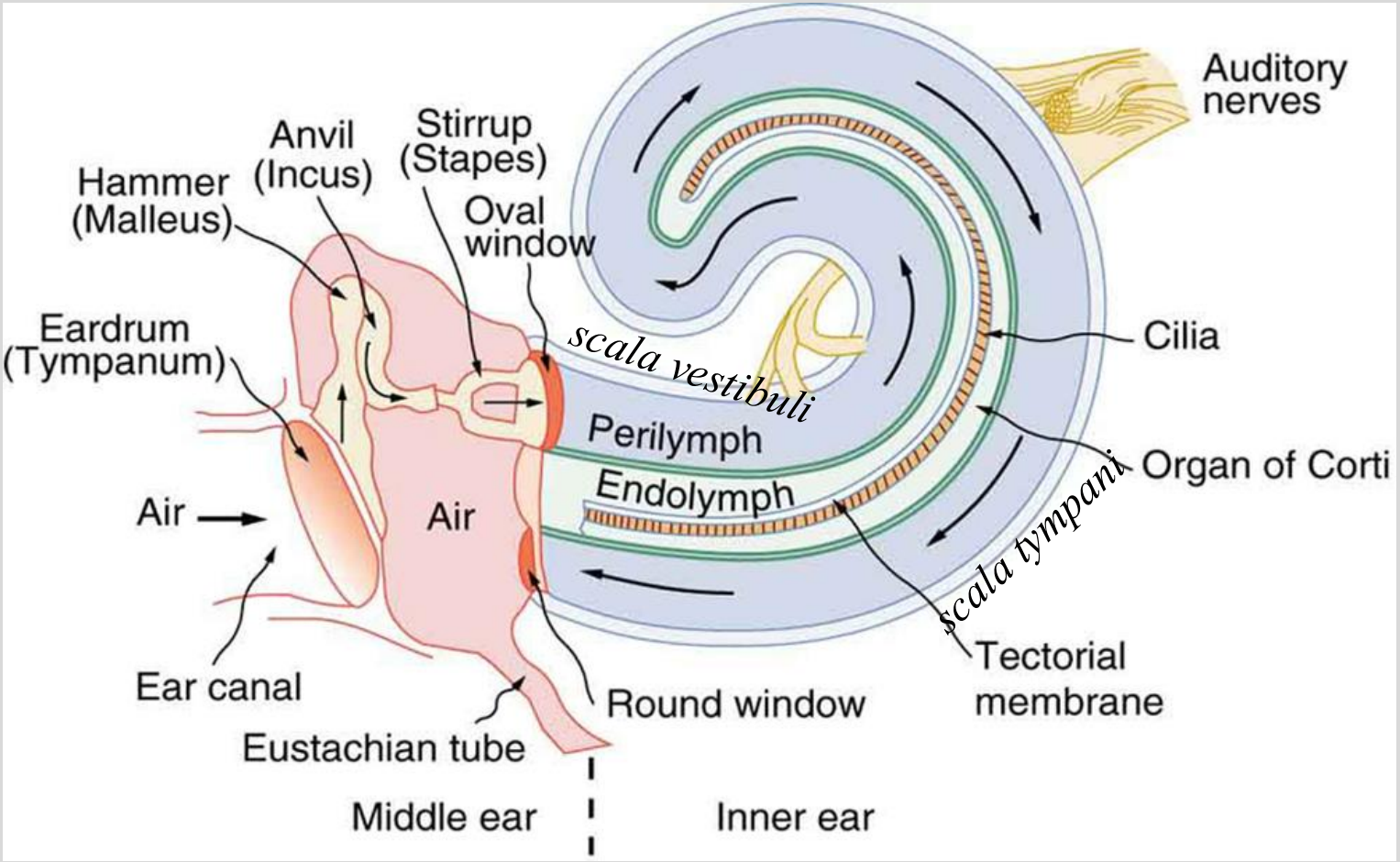
The cochlea decomposes vibration into its frequency components



Oval window location

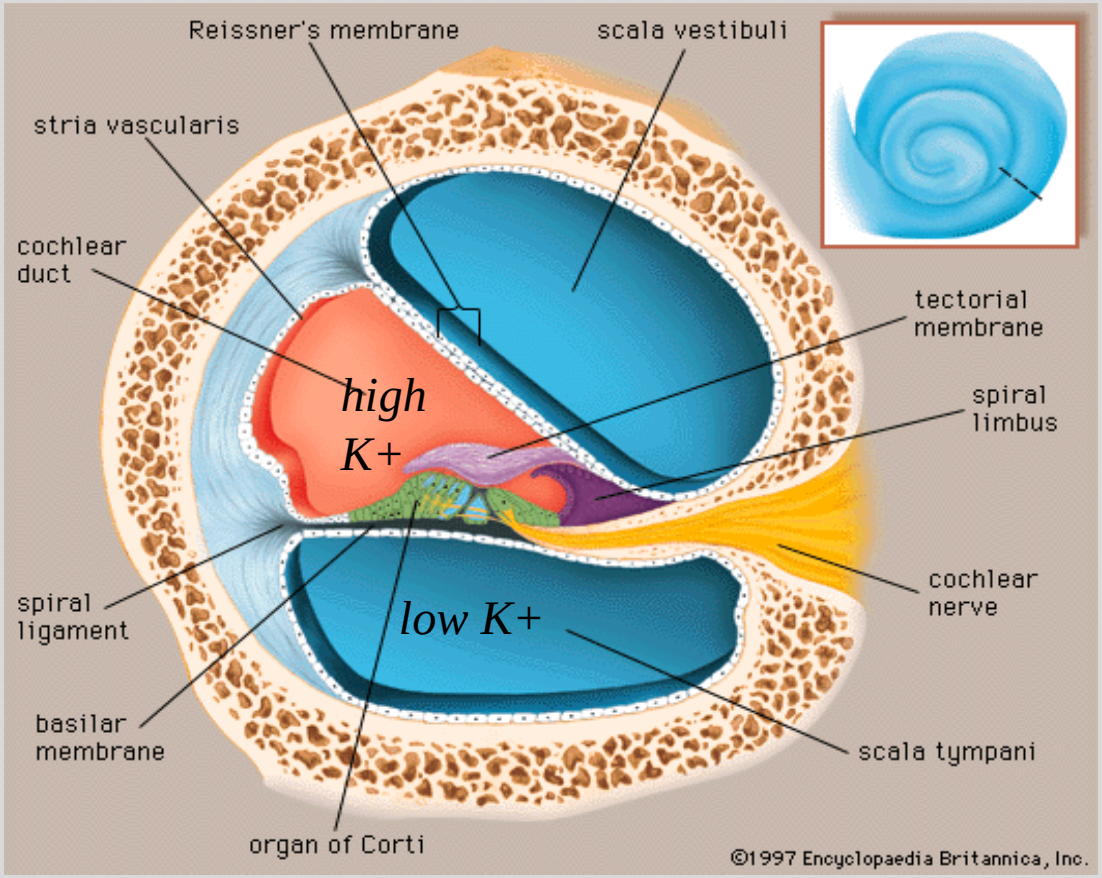
- Differences in stiffness determines the location of the oscillation of the basilar membrane, converting a frequency into a place code.
- Notice that the diameter of the cochlea is reversed of that of the basilar membrane: at the oval window the cochlea is broadest and the basilar membrane is narrowest

Anatomy of the cochlea



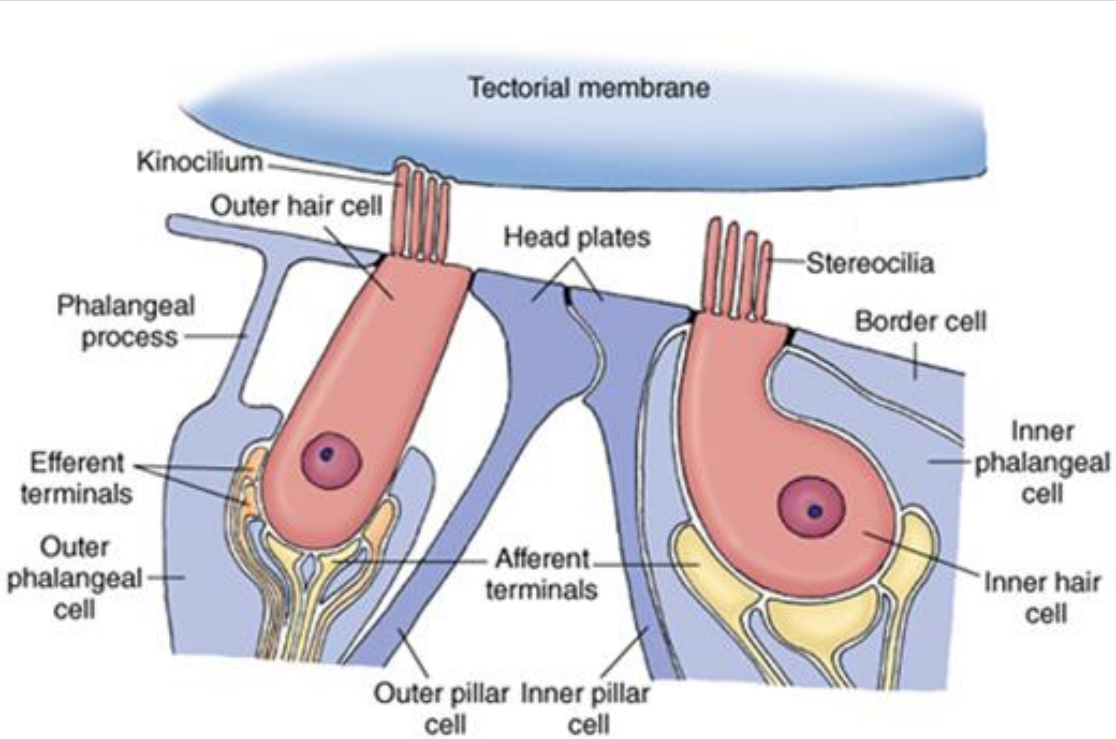
- *Waves travel from the oval window to the round window. Oval window->scala vestibuli->helicotrema->scala tympani..*

Anatomy of the cochlea



- Difference in ionic concentration of endolymph in the cochlear duct surrounding the hair cells (high potassium) vs perilymph in the scala vestibuli and scala tympani (low potassium concentration).

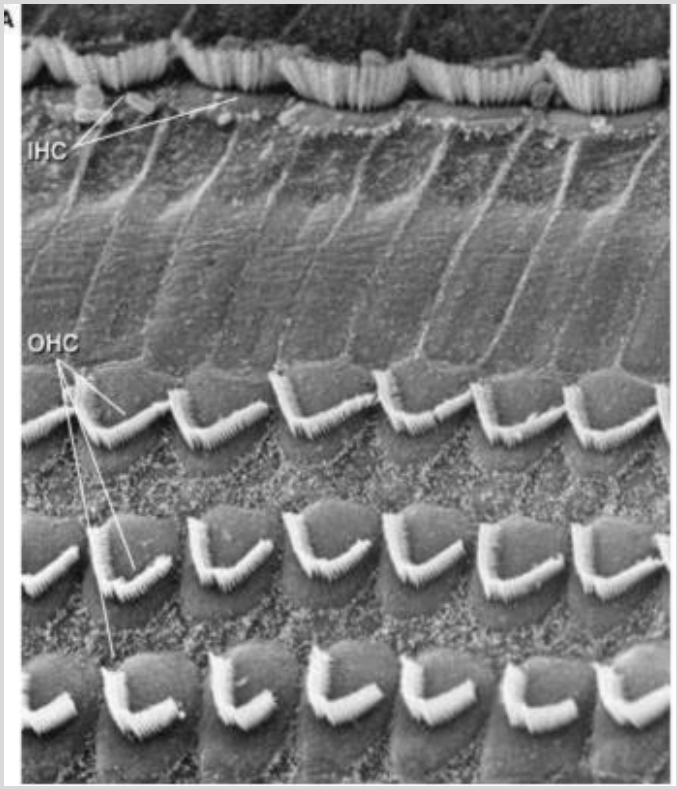
Organ of Corti

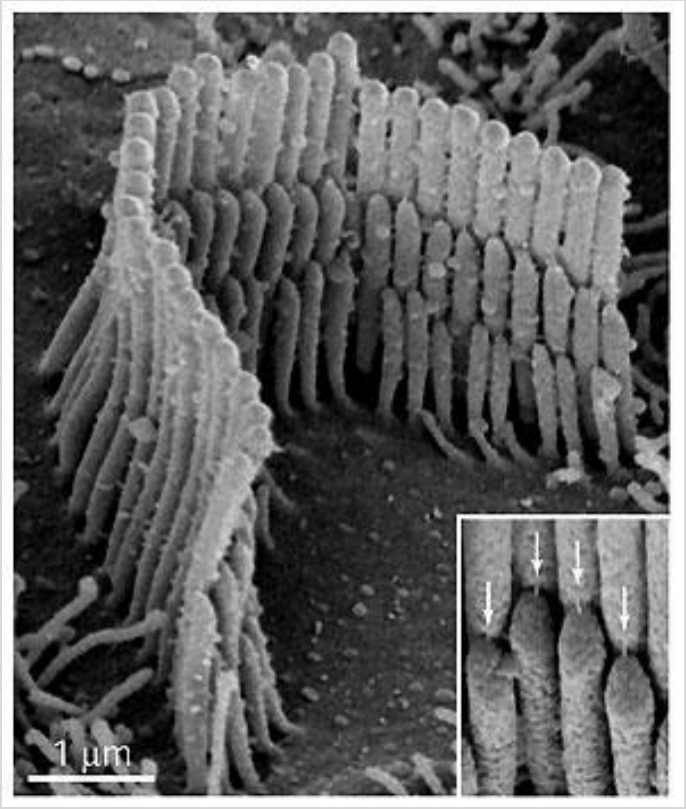


- Two types of hair cells: inner hair cells and outer hair cells.
- The inner hair cells are the actual sensory receptors, and 95% of the fibers of the auditory nerve that project to the brain arise from this subpopulation.
- Stereocillia are sound mechanoelectric transducers found on the tops of hair cells.

3000 inner hair cells!!

Inner & Outer hair cells



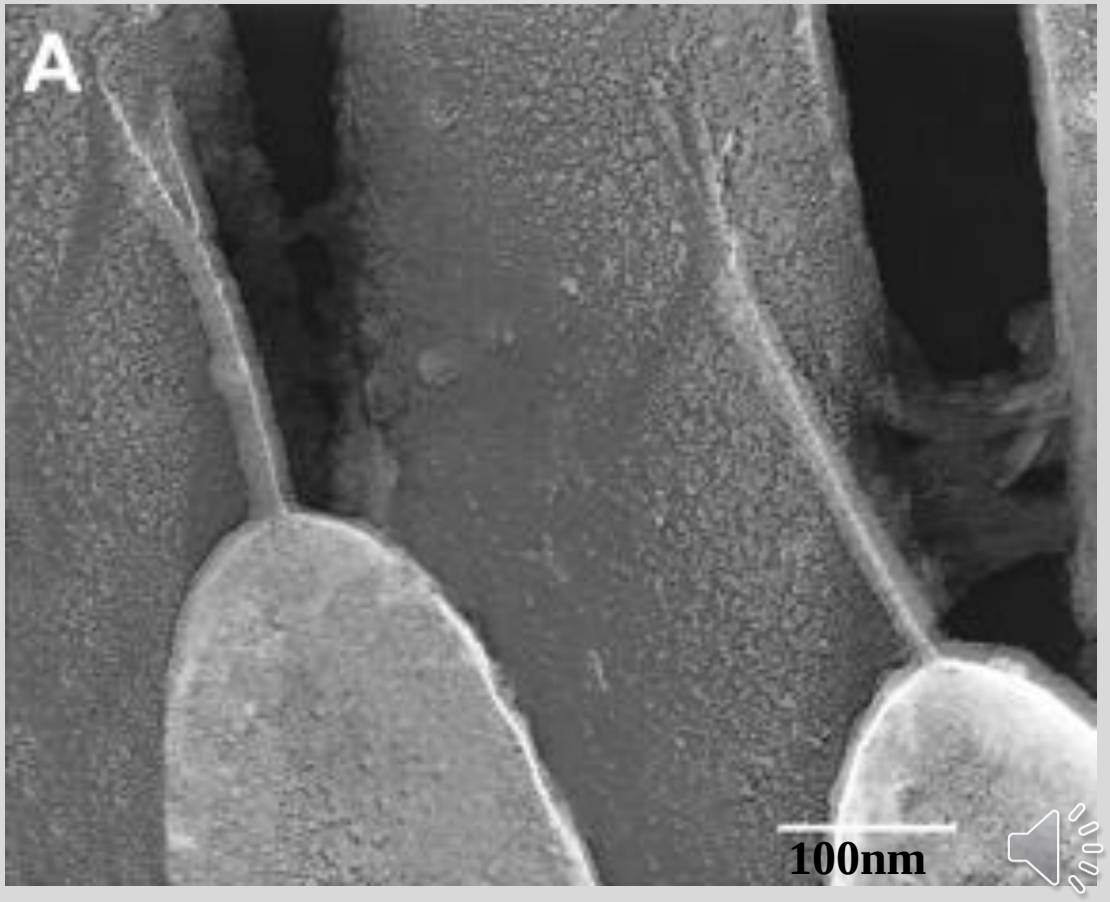


- The cochlear hair cells in humans consist of one row of **inner hair cells** and three rows of **outer hair cells**.
- Very small number of inner hair cells: 3500 inner hair cells.

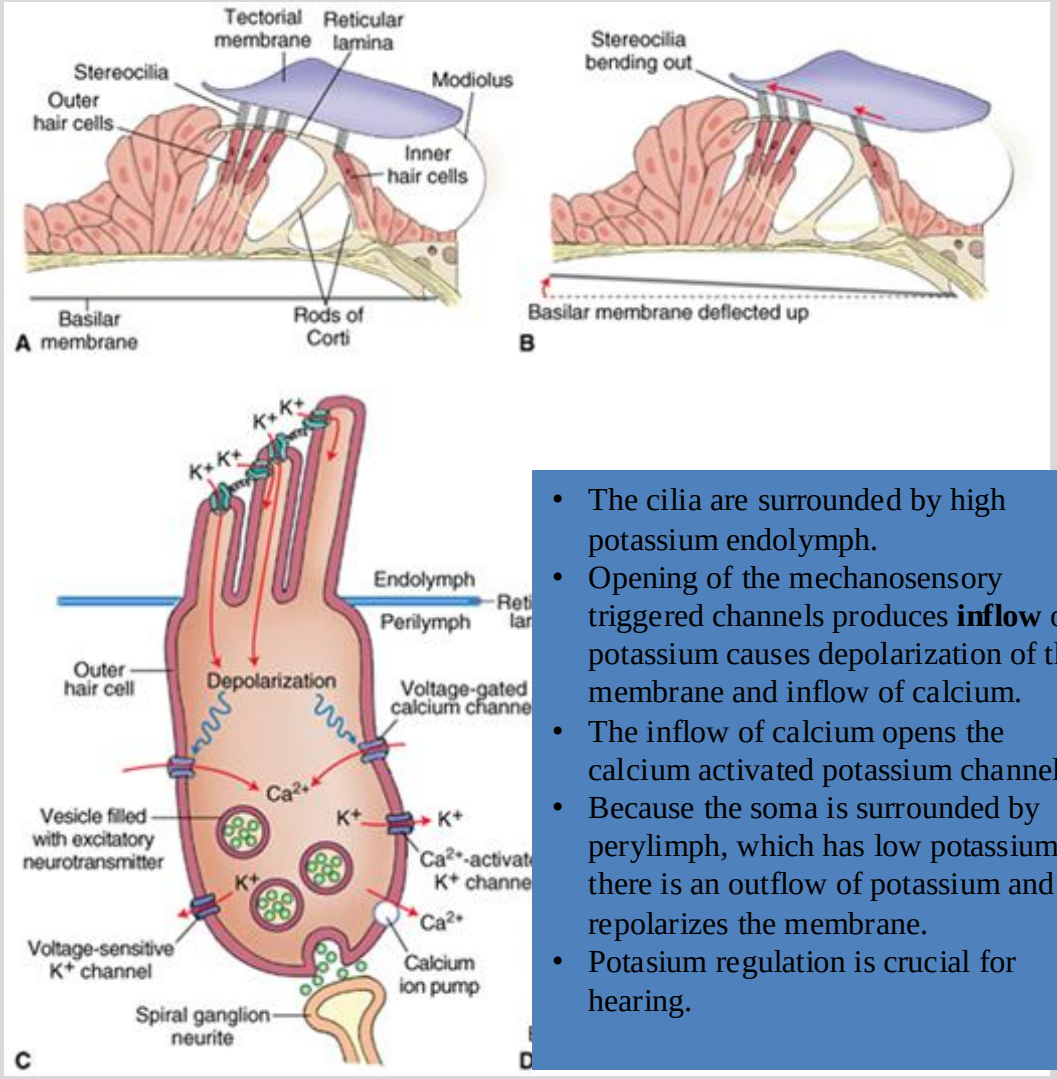
Tip links connect stereocillia.

Tip links functionally coupled to mechano – electric transduction channel (MET)

Stretch of steroecillia causes opening of MET channel.



Hair cells detect deflections of the basilar membrane



- The cilia are surrounded by high potassium endolymph.
- Opening of the mechanosensory triggered channels produces **inflow** of potassium causes depolarization of the membrane and inflow of calcium.
- The inflow of calcium opens the calcium activated potassium channels.
- Because the soma is surrounded by perylimph, which has low potassium, there is an outflow of potassium and repolarizes the membrane.
- Potasium regulation is crucial for hearing.

- The tips of stereocilia of outer hair cells are embedded in the tectorial membrane.
- Movement of the basilar membrane relative to the tectorial membrane results in displacement of the cilia that is necessary for generating afferent signals.
- Inner hair cells are not attached to the tectorial membrane and movement of their stereocilia is induced by movement of the endolymph.
- Hair cells synapse onto Spiral ganglia neurons
- 1 Spiral ganglia neuron connected to 1 inner hair cell.
- Hair cells release glutamate but do not fire action potentials
- Axons of Spiral ganglia neurons form CNVIII
- Efferent axons also innervate hair cells

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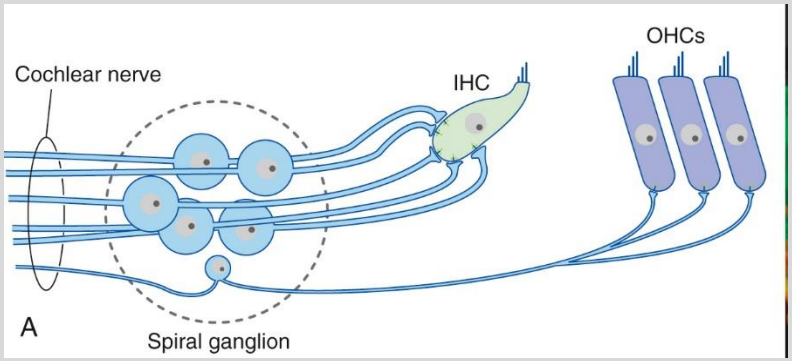
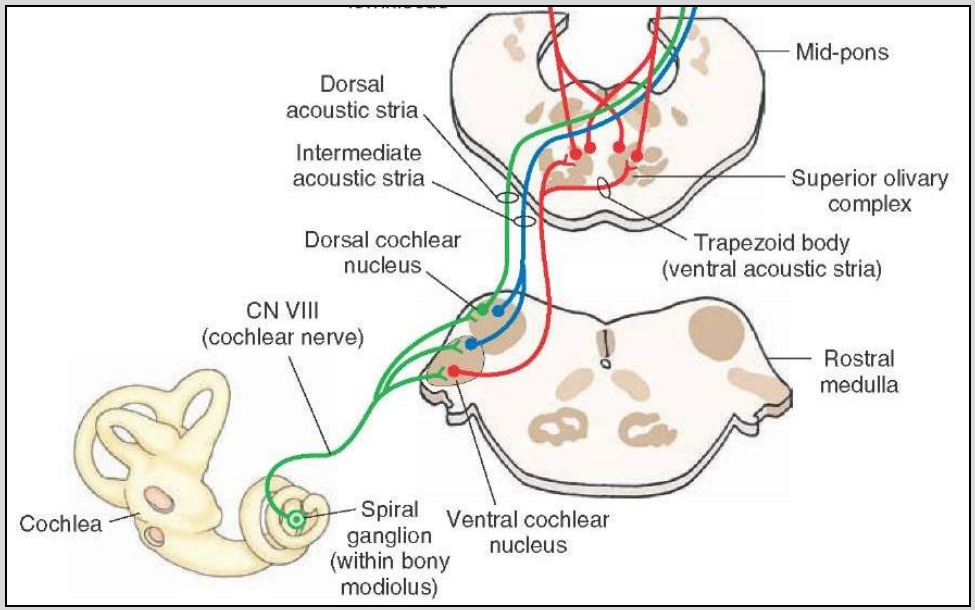
Ascending auditory pathway



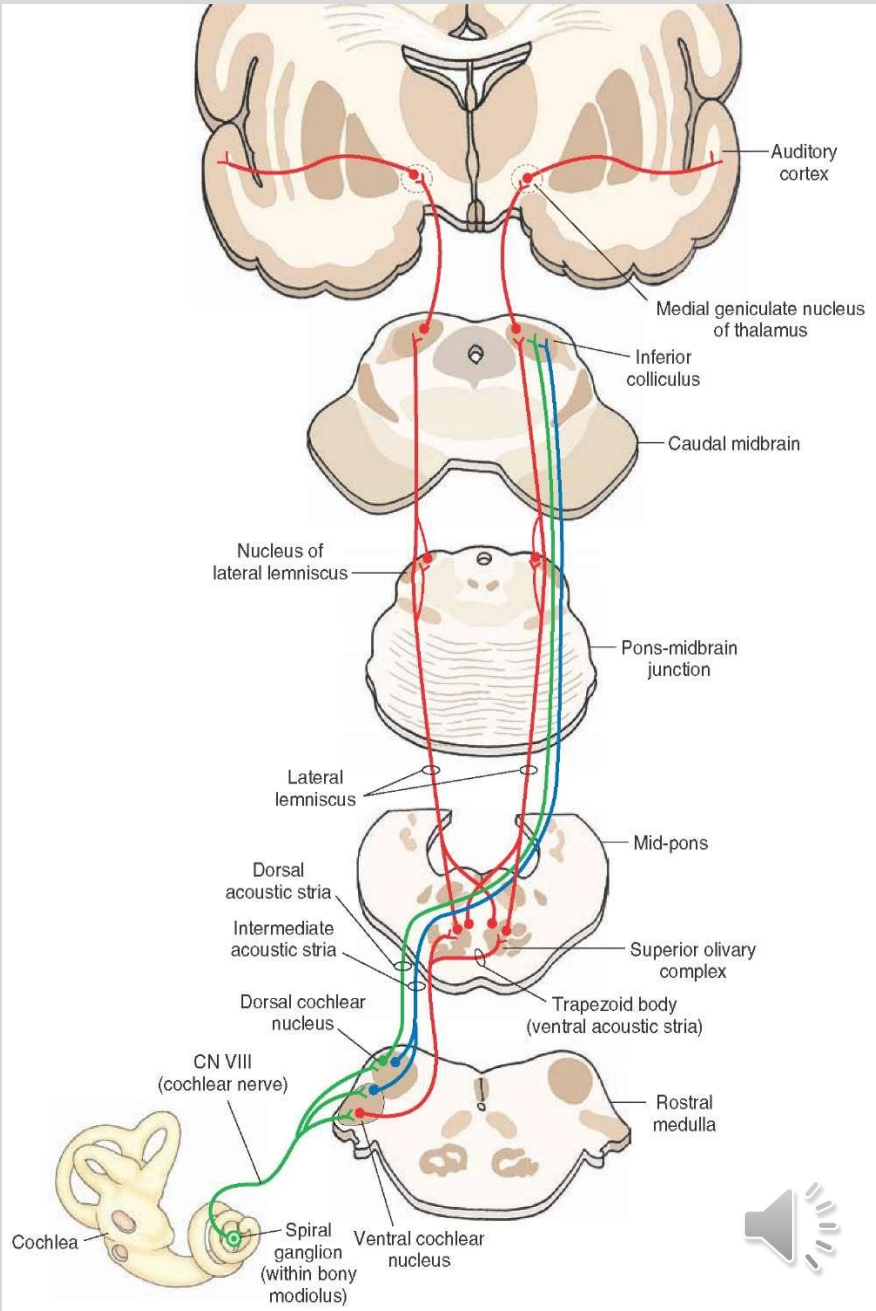
Nolte. The Human Brain



- Neurons in the spiral ganglion receive input from the inner hair cells and send their axons into the ipsilateral cochlear nucleus in the rostral medulla.
- Axons of the cochlear nucleus neurons ascend to the midpons where the fibers decusate and continue ascending.
- Some fibers make synapses in the superior olivary complex in the midpons. Some other fibers decusate and make synapse on the contrallateral superior olivary complex. There information from both ears can be compared and sound localization computation can be performed using intensity cues and time differences cues.

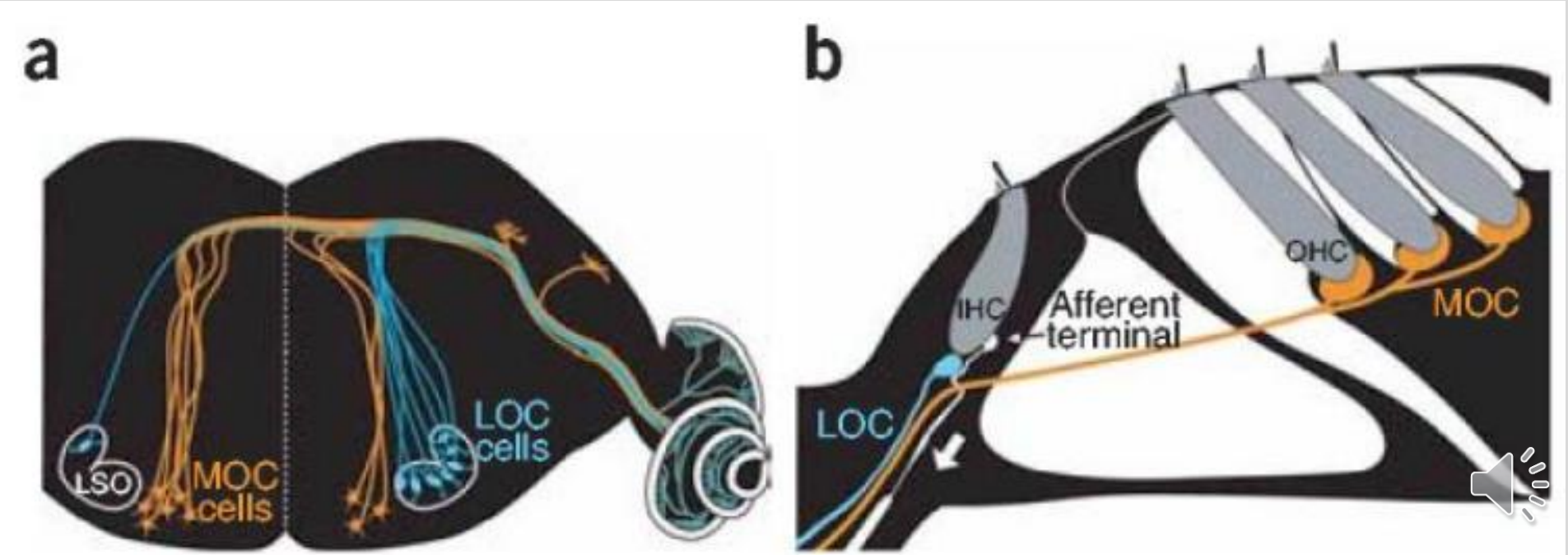
Auditory pathway

- Fibers ascend through the lateral lemniscus.
- After the decussation in the midpons, the fibers carrying auditory information make synapses in the inferior colliculus.
- The inferior colliculus project to the medial geniculate body of the thalamus bilaterally(auditory thalamus)
- Thalamic neurons project to the auditory cortex in the temporal lobe.



Olivocochlear descending pathway - two major subsystems

The medial (MOC) component is made up of myelinated axons of neurons of the medial superior olive projecting to the cochlear outer hair cells.
The lateral (LOC) component is made up of unmyelinated axons of neurons of the lateral superior olive projecting to cochlear nerve fibers, near their afferent synapses with cochlear inner hair cells.



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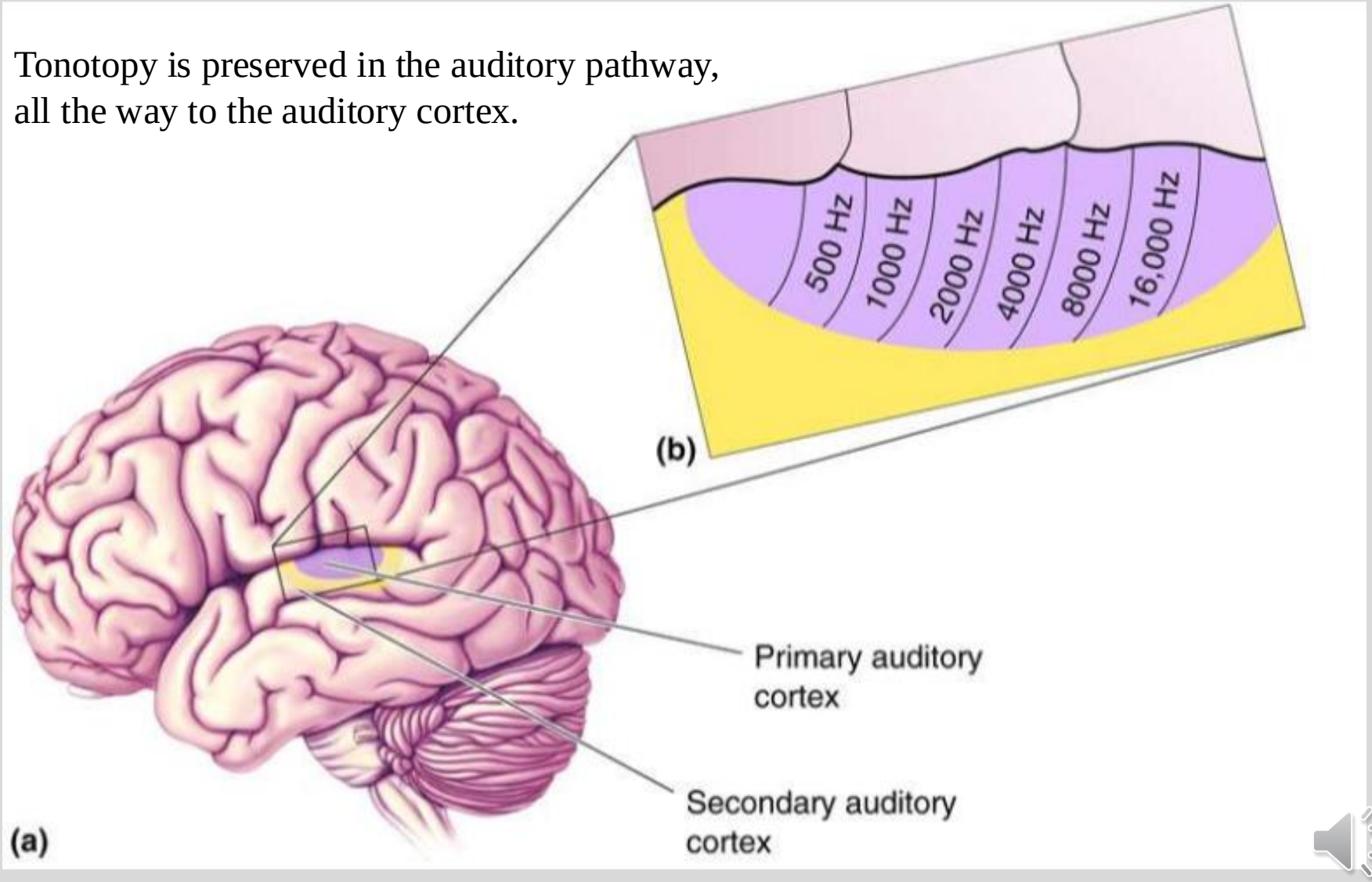
Tonotopy is preserved in Auditory Cortex

Tonotopy is preserved in the auditory pathway, all the way to the auditory cortex.



Response profile of a human auditory cortical neuron to tone presentations during neurosurgery.

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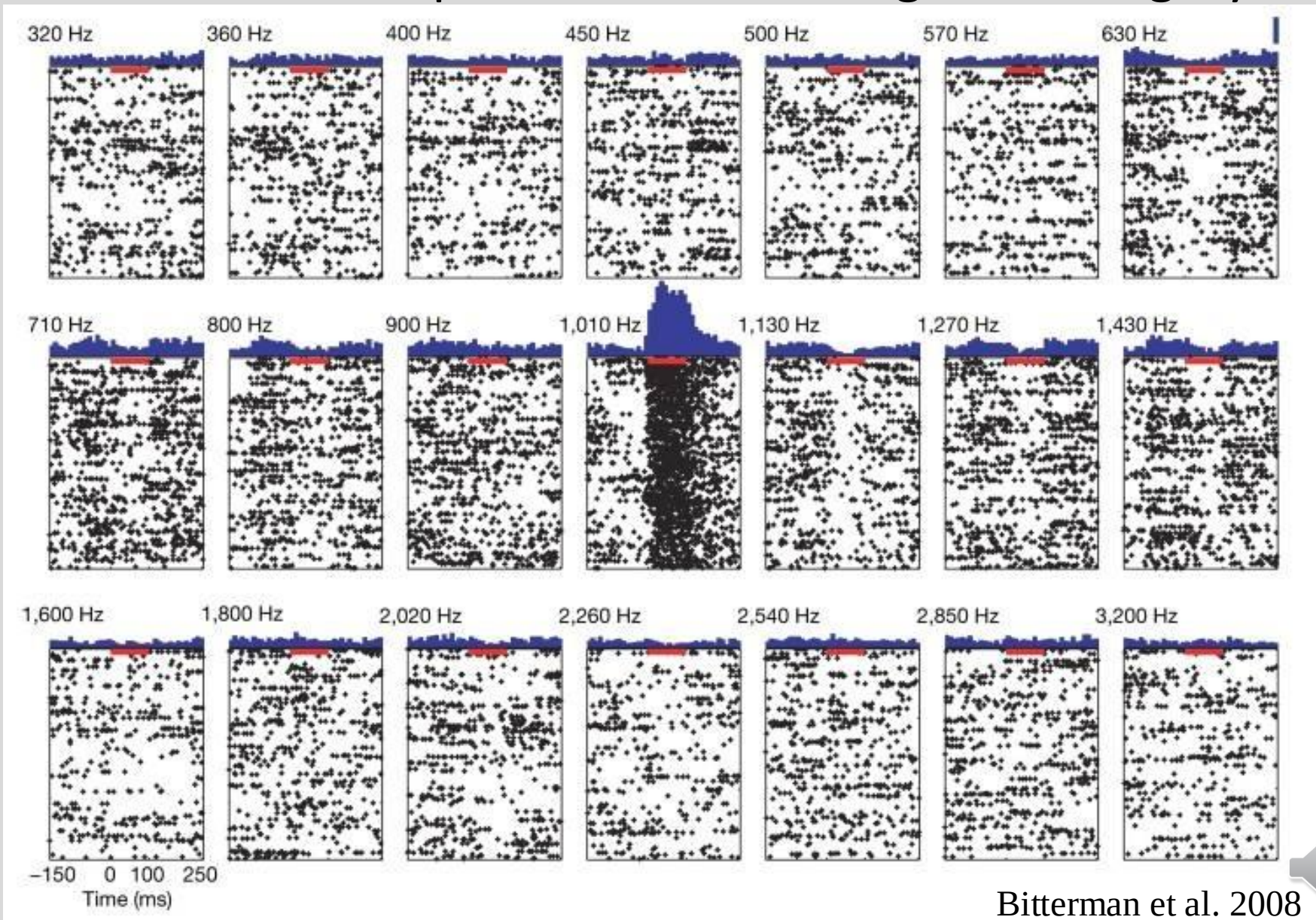

Bitterman et al. 2008

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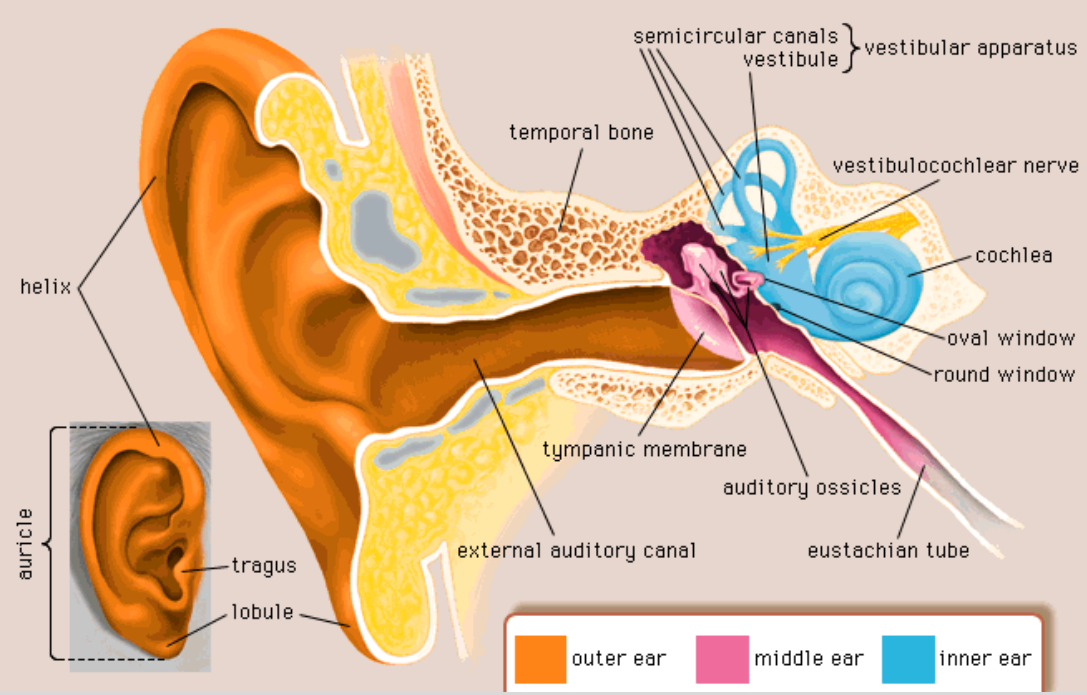
Hearing Loss

- Conductive: involves anything that blocks the conduction of sound from the external and middle ear
  - This is often correctable
- Sensorineural: involves the inner ear, cochlea, or auditory nerve
  - Usually caused by hair cell pathology
- Mixed Loss: Combination of the two



Causes of Conductive Hearing Loss

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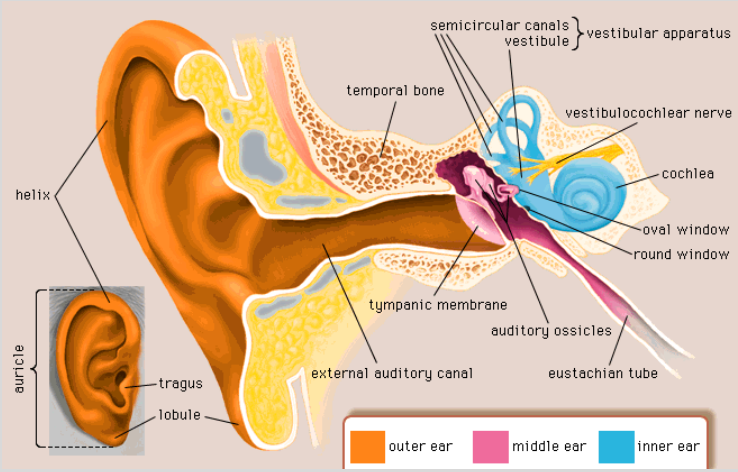
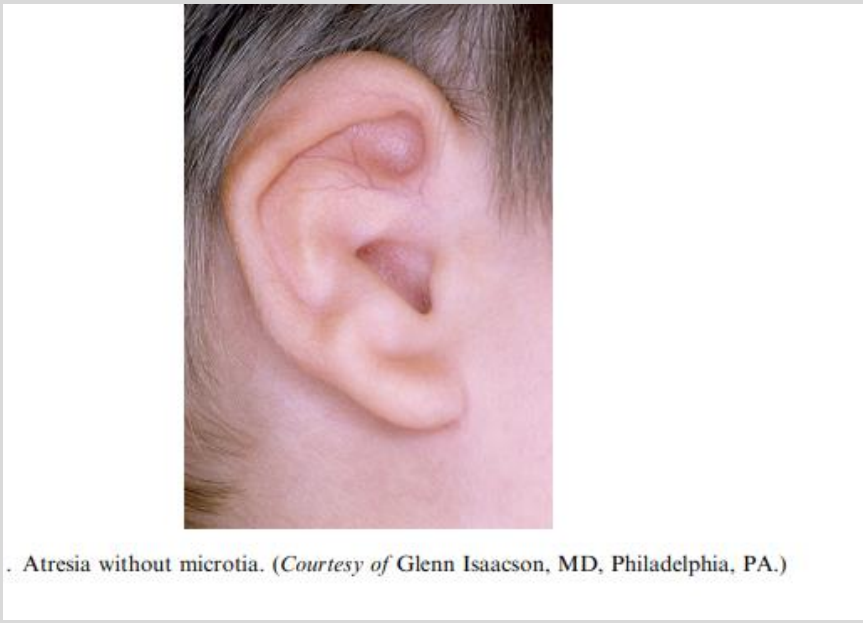
- Congenital – atresia or malformation of the external auditory canal



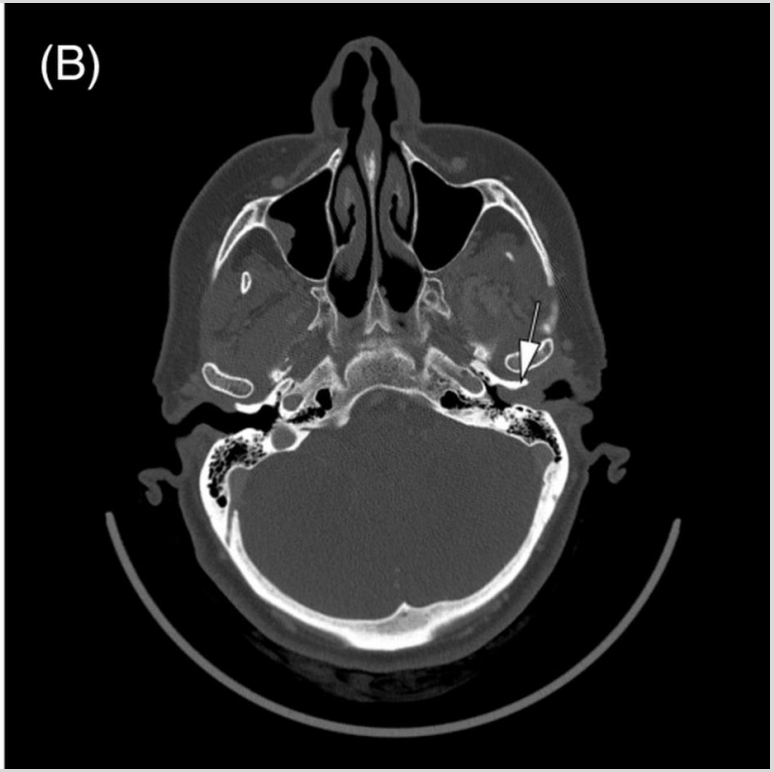


. Atresia without microtia. (*Courtesy of* Glenn Isaacson, MD, Philadelphia, PA.)

- It is unilateral in a majority of cases
- Children with unilateral aural atresia typically have normal speech development so long as the other ear is unaffected and typically have normal hearing and ear development in the unaffected ear.
- Possibility of malformation of the ossicles
- Treatment is surgical
- They are at increased risk of delayed language development due to functional mono-aural hearing, and early identification of this is essential.

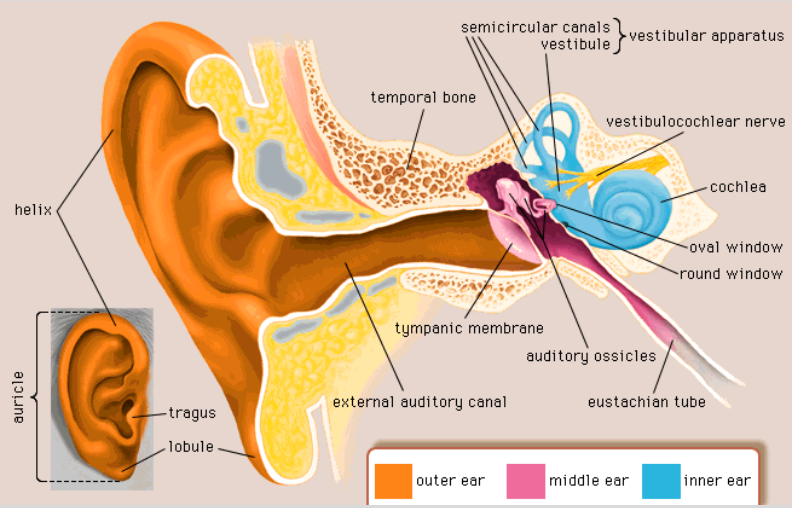
Causes of Conductive Hearing Loss

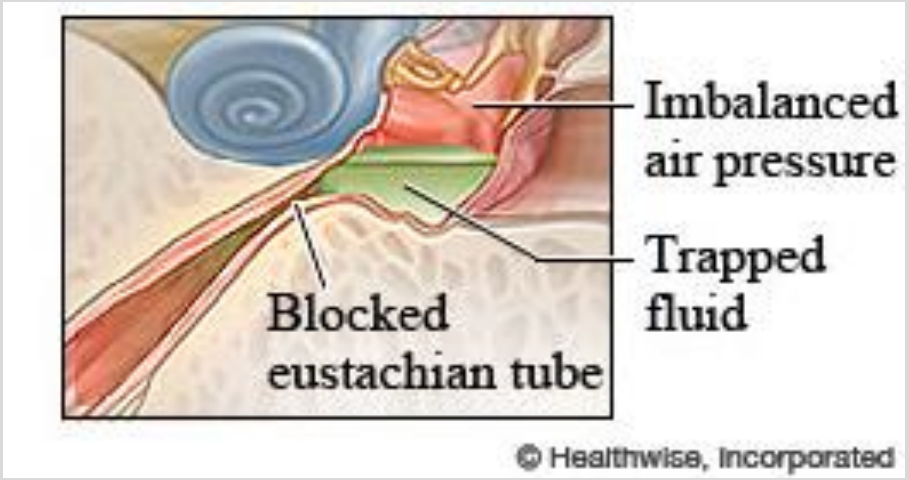
- Squamous cell carcinoma can block the ear canal or affect the middle ear



Causes of Conductive Hearing Loss

- Eustachian tube dysfunction – usually with viral URI or sinusitis





- Blocked eustachian tube leads to the accumulation of fluid in the middle ear.
- The fluid accumulation affects the transmission of vibration thorough the ossicles.
- Middle ear barotrauma – filling of middle ear with serous fluid or blood

Causes of Conductive Hearing Loss

- Osteoma – at tympanosquamous suture line
- Malformation of the ossicles
- Otosclerosis – over stapes footplate
- Benign polyps and cholesteatomas
- Cerumen impaction

Causes of Sensorineural Loss

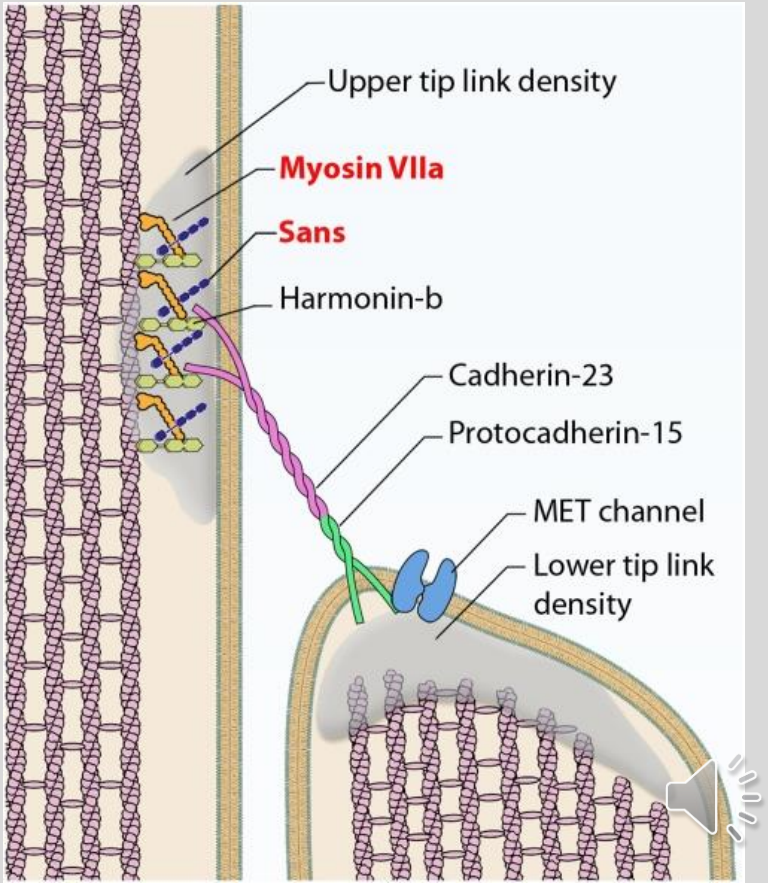
- Congenital
  - Can be hereditary, autosomal dominant or recessive (Usher syndrome)
  - Non-hereditary caused by infections, recreational drug use, alcohol, and retinoic acid
- Presbycusis – age related hearing loss (specifically high frequencies, only 3500 inner hair cells)
- Infections such as meningitis and measles which destroy inner ear hair cells
- Noise exposure – damages cochlear structures (especially high frequencies)
- Tumor – most commonly acoustic neuroma
- Arnold Chiari malformations (tonsils) that impinge on CN VIII
- Medications – antibiotics, chemotherapeutics, high dose aspirin, anti-malarial agents

Usher syndrome

	Type 1	Type 2	Type 3
Hearing	Profound deafness in both ears from birth	Moderate to severe hearing loss from birth	Normal at birth; progressive loss in childhood or early teens
Vision	Decreased night vision before age 10	Decreased night vision begins in late childhood or teens	Varies in severity; night vision problems often begin in teens
Vestibular function (balance)	Balance problems from birth	Normal	Normal to near-normal, chance of later problems

Usher syndrome, type 1:

- Associated gene mutations: *MY07A*, *USH1C*, *CDH23*, *PCHD15*, *SANS*
- These genes play a role in development and maintenance of stereocilia



Epilepsy, Ataxia, Sensorineural Deafness, and Tubulopathy (EAST syndrome):*KCNJ10* Mutations

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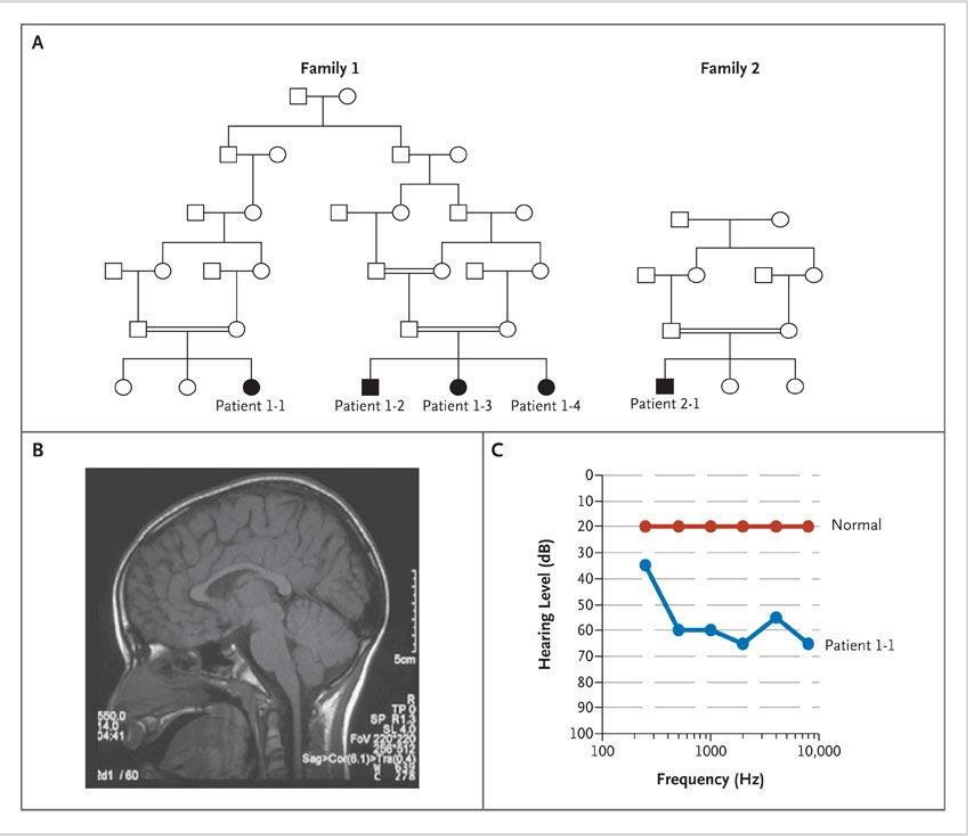
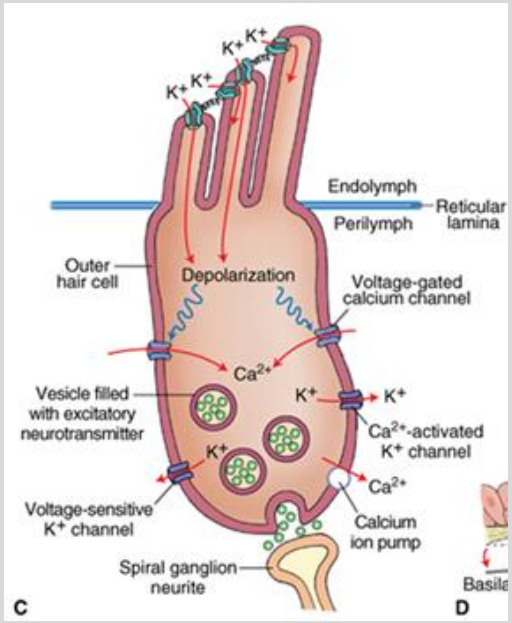
Recessive genetic disorder



- Kir4.1 Potassium channel expressed in brain, inner ear, and kidney

- Kir4.1 participates in the generation and maintenance high K^+ concentration in the endolymph.

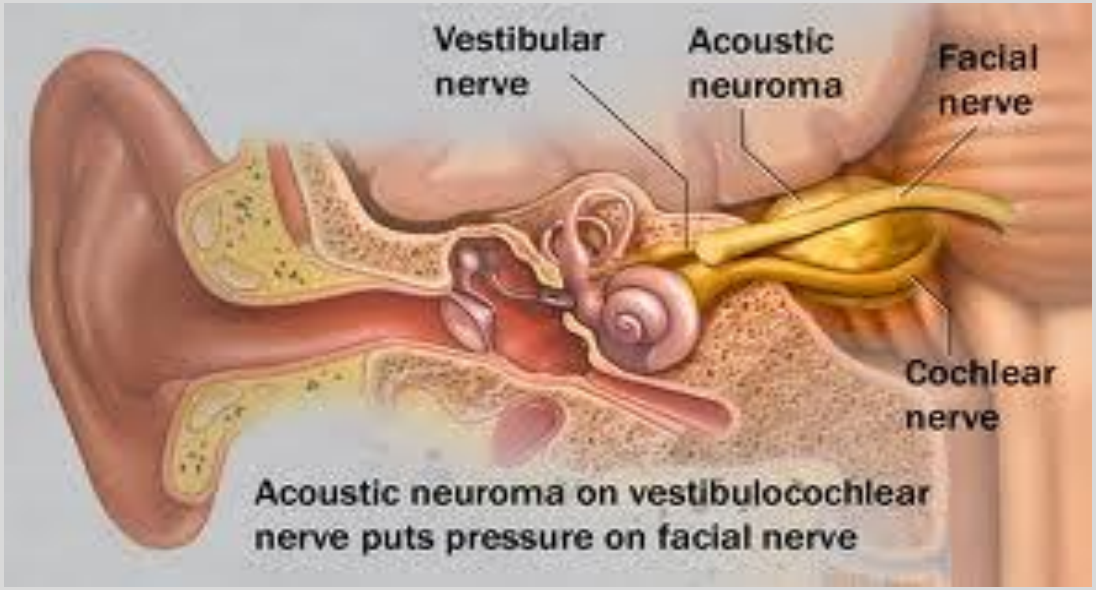
-Without the high extracellular potassium, the inner hair cells will not depolarize.



Tinnitus

- Most common description is a high pitched continuous tone
- Caused by hyperactivity of cochlear amplification/outer hair cells
- Frequently is the first sign of a lesion that is causing hearing loss (esp. sensorineural)

Vestibular schwannoma 4/100,000

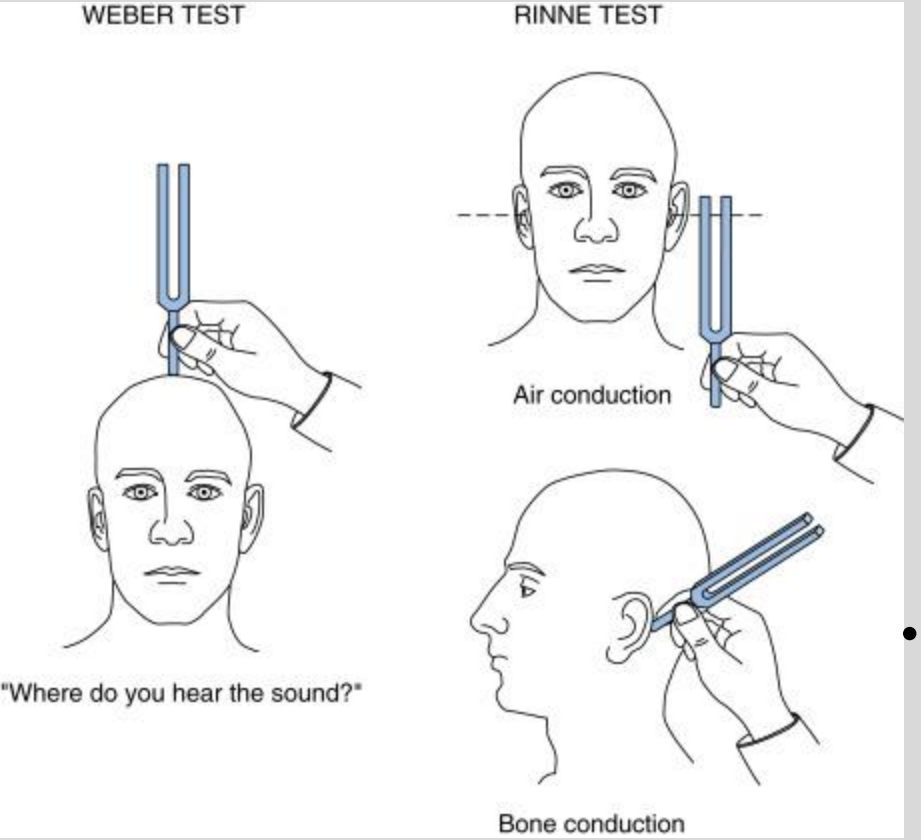


- Slow growing tumor of the vestibular cochlear nerve
- Benign tumor, but can cause damage to surrounding structures as it grows including facial nerve
- Most common symptoms: vertigo, hearing loss, tinnitus
- Most useful test to diagnose: MRI of the brain
- Surgery is the treatment
  - Complications include hearing loss and paralysis of the facial muscles

Ear & Auditory testing

- Otoscopoic exam
- Pure-Tone Testing
- Speech Testing
- Tests of the Middle Ear
  - tympanometry, acoustic reflex testing
- Auditory Brainstem Response
- Otoacoustic Emissions
- Weber & Rinne tests

Ear & Auditory testing



Evidence-Based Physical Diagnosis (Fourth Edition), 2018

- **Rinne test.** Tuning fork is placed against the mastoid bone. Once the subject cannot hear the bone conducted sound, the tuning fork is moved close to the ear, therefore using air conduction. Air conduction should more efficient than bone conduction, so the patient should report that the sound has restarted. If not, that would indicate conductive hearing loss.
- **Weber test: If sound of the tuning fork is localized to affected ear.** This means that bone conduction is unaffected. Therefore, the patient has conductive hearing loss on that ear.
- **If the sound is localized to non-affected ear,** it means there is sensorineural hearing loss in that ear.

Summary slide

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- The anatomy of the auditory system includes the ear (outer, middle, inner) as well as diverse central nuclei of the brainstem and thalamus and finally the neocortex.
- Audition emerges from mechanotransduction and synaptic transmission along the auditory pathway.
- Dysfunction of mechanotransduction or synaptic transmission broadly causes hearing loss.

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