NEUROANATOMIC BASIS OF HEARING IMPAIRMENT AND ITS MANAGEMENT.

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(ENT, HEAD AND NECK SURGERY)

OUTLINE

- Introduction
- Definition
- Epidemiology
- Anatomy
- Physiology
- Central auditory pathway
- Management
  - History
  - Examination
  - Investigation
  - Treatment
  - Follow-up
  - prevention
- Conclusion

INTRODUCTION

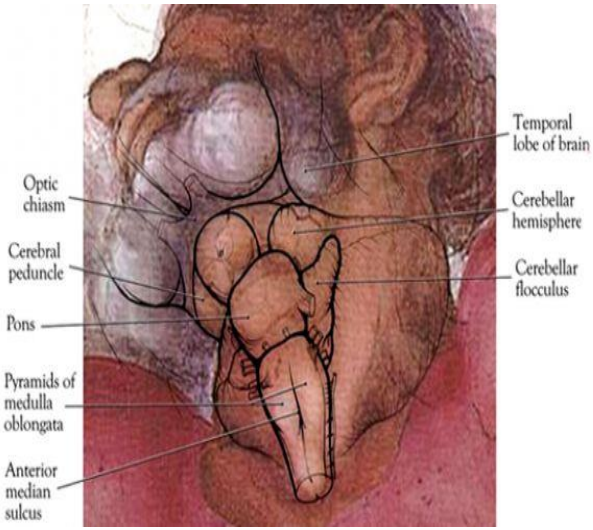
- Hearing loss is more prevalent than diabetes mellitus, myelomeningocele, all pediatric cancers, and numerous other medical conditions.

- Many medical professionals learn little about hearing impairment, how to advise parents of children who are deaf or hard of hearing, or about the special considerations needed in the care of children with hearing loss.

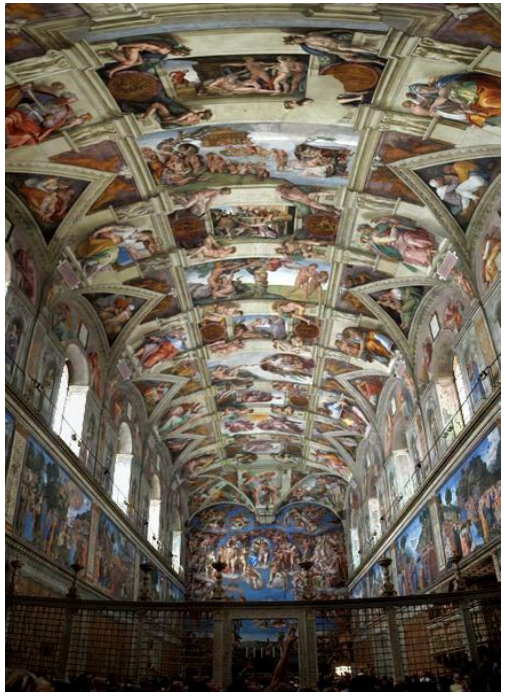
Introduction

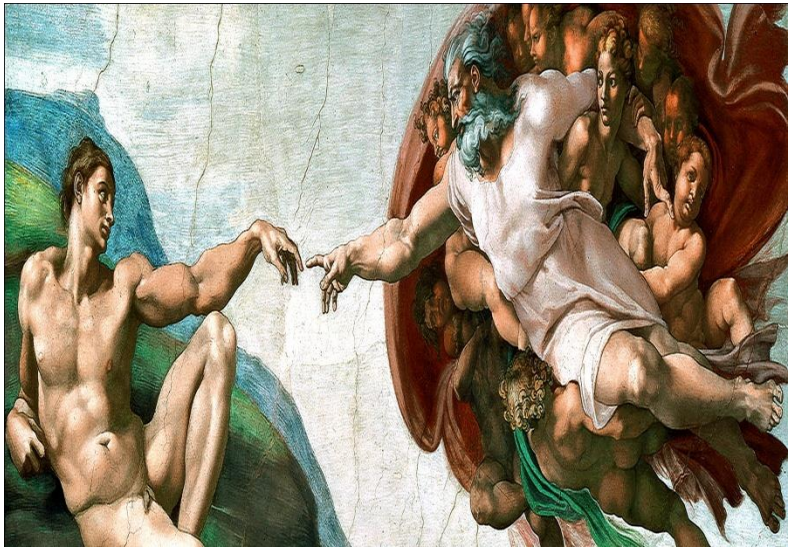
- The ancient Egyptian document the Edwin Smith Papyrus
- The Greek Alcmaeon determined that the brain and not the heart ruled the body and that the senses were dependent on the brain.
- In 1664, Thomas Willis, a physician and professor at Oxford University, coined the term neurology when he published his text Cerebri anatome- considered the foundation of neuroanatomy.

History of neuroanatomy; Michelangelo?



History of neuroanatomy; Michelangelo?





Definitions

- Hearing is a conscious appreciation of vibration perceived as sound.

- Hearing is the process by which the ear transforms sound vibrations into nerve impulses that are conveyed to the brain, and interpreted as sounds.

- Sounds are produced when vibrating objects, produce pressure pulses of vibrating air molecules, better known as sound waves

Definitions

- Hearing loss exists when there is diminished sensitivity to the sounds normally heard.

- The terms hearing impairment or hard of hearing are usually reserved for people who have relative insensitivity to sound in the speech frequencies.

EPIDEMIOLOGY

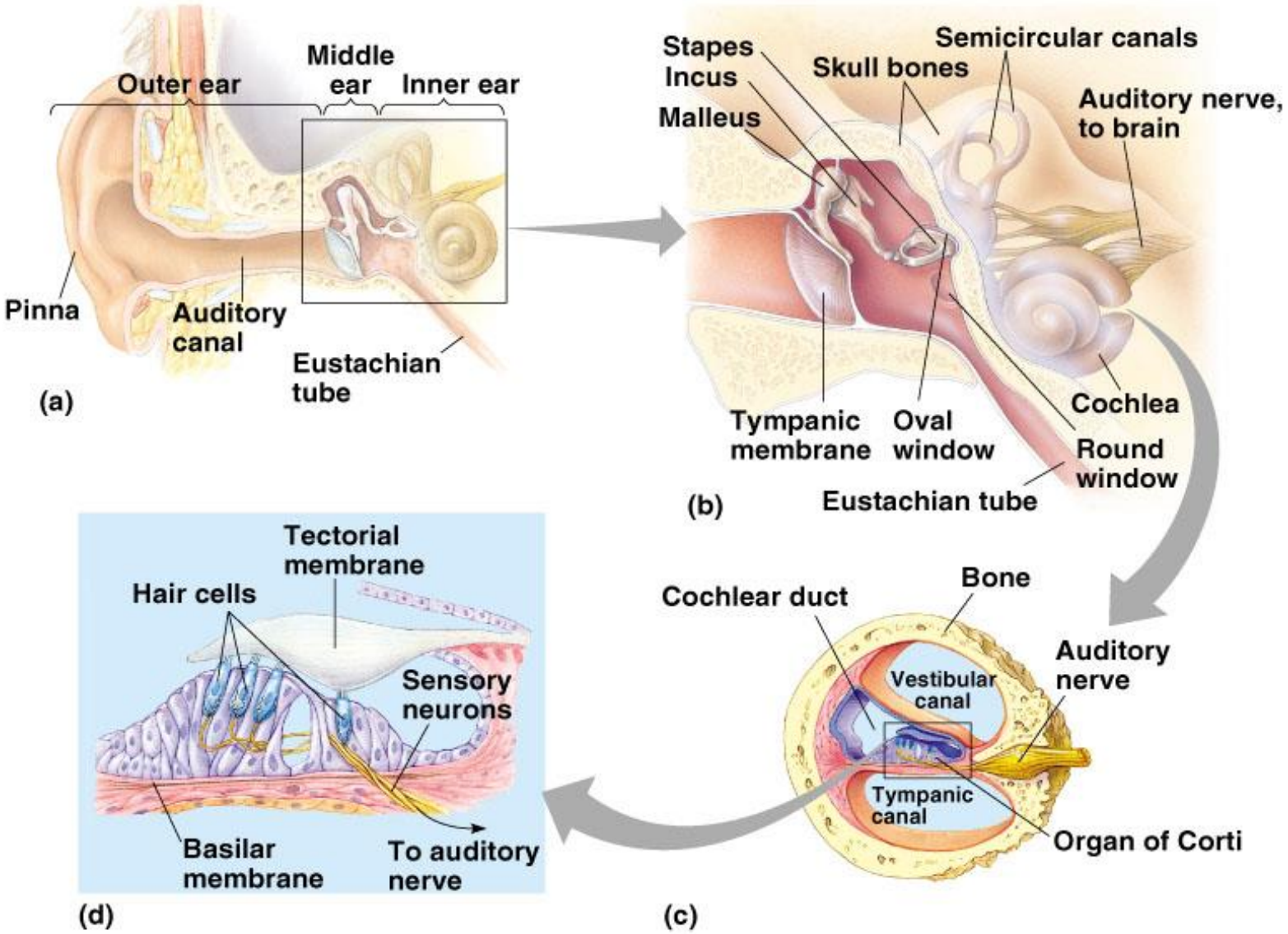
United states

- Hearing loss occurs in approximately 5-10 per 1000 children
- Approximately 1 in 20 adolescents has significant hearing loss.
- Almost 3% of the population in the workforce reports having some hearing loss.

International

- SNHL occurs in 9-27 per 1000 children worldwide.
- **Sex**
- No sex predilection is known.
- **Age**
- Hearing loss may occur at any age

ANATOMY



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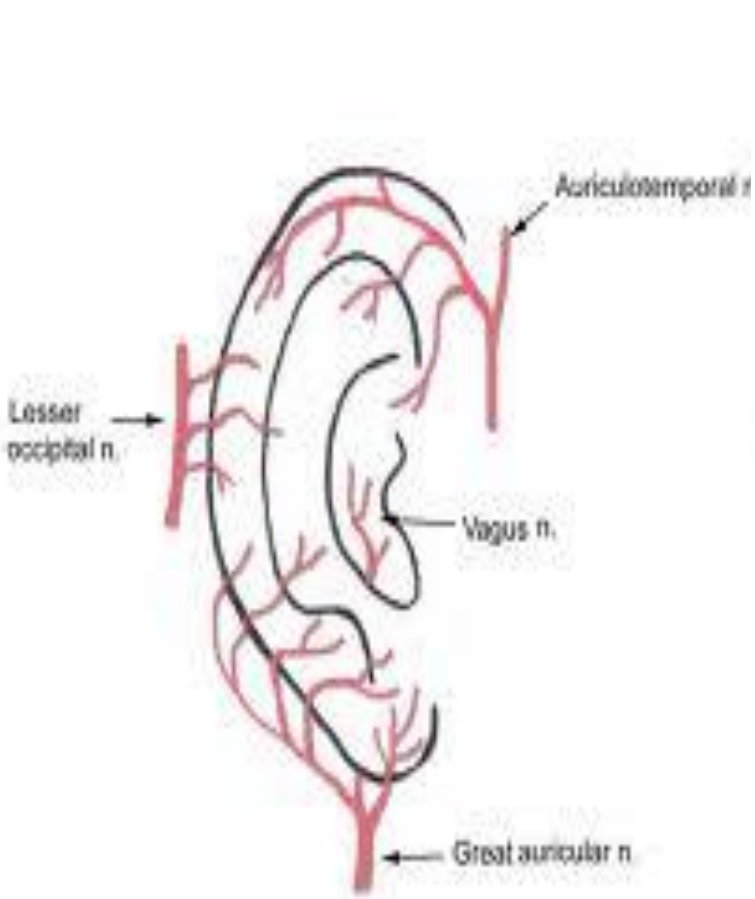
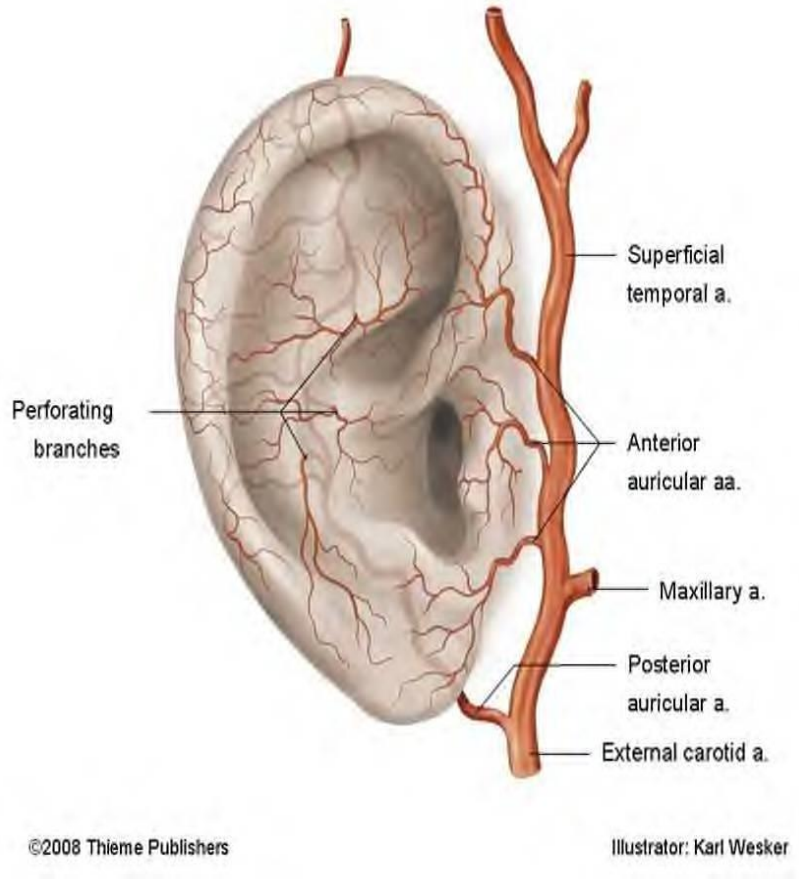

Auriculotemporal
Lesser occipital n.
Vagus n.
Great auricular n


Superficial temporal a.
Perforating branches
Anterior auricular aa.
Maxillary a.
Posterior auricular a.
External carotid a.
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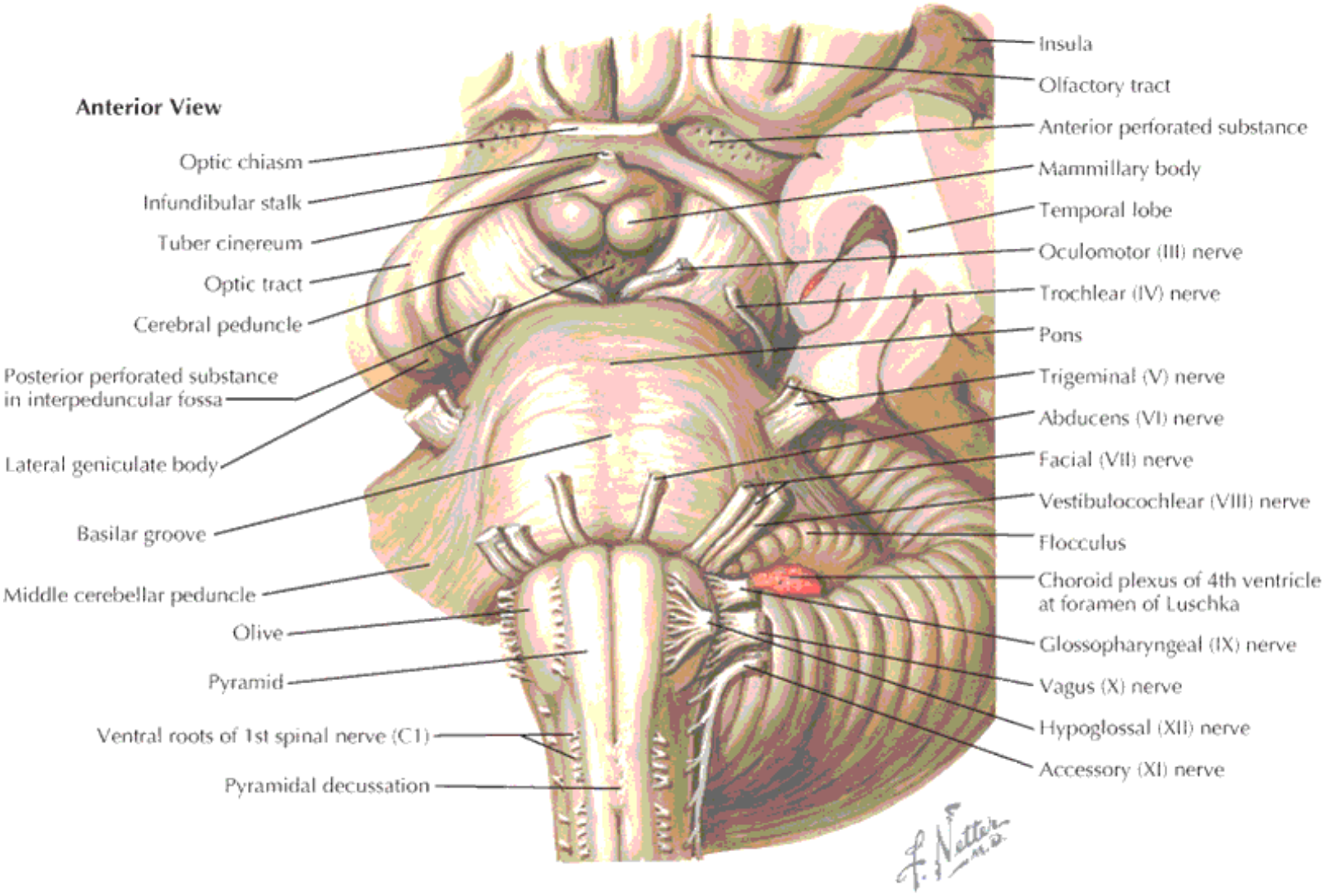
Illustrator: Karl Wesker

Anatomy

- Middle ear;
- Tympanic branch of maxillary
- Mastoid branch of occipital or post auricular
- Veins-pterygoid plexus or superior petrosal sinus
- Nvs-glossopharyngeal, vagus, facial
- Inner ear;
- Labyrinthine artery
- Veins-
- Nerve-vestibulocochlear

ANATOMY



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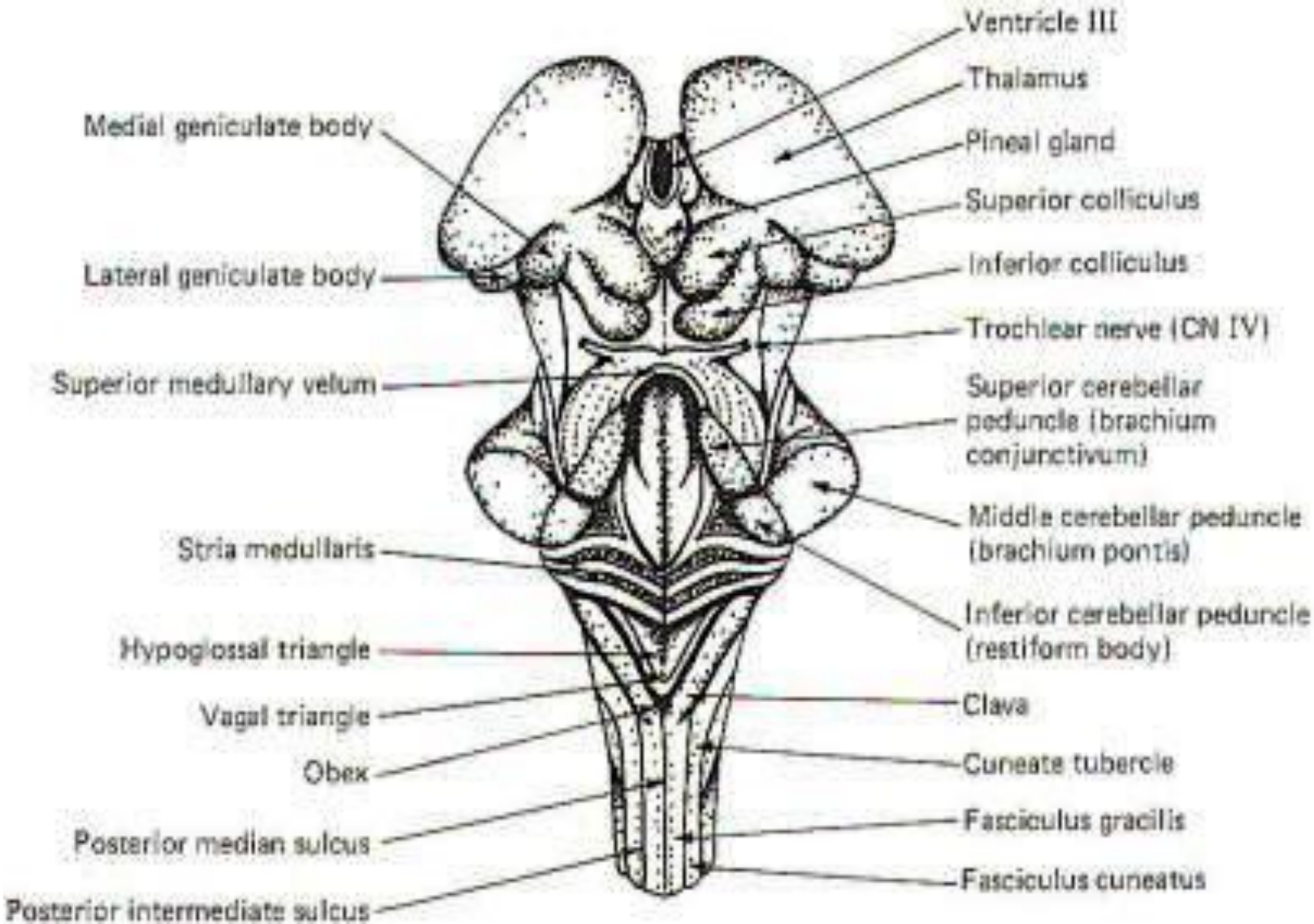
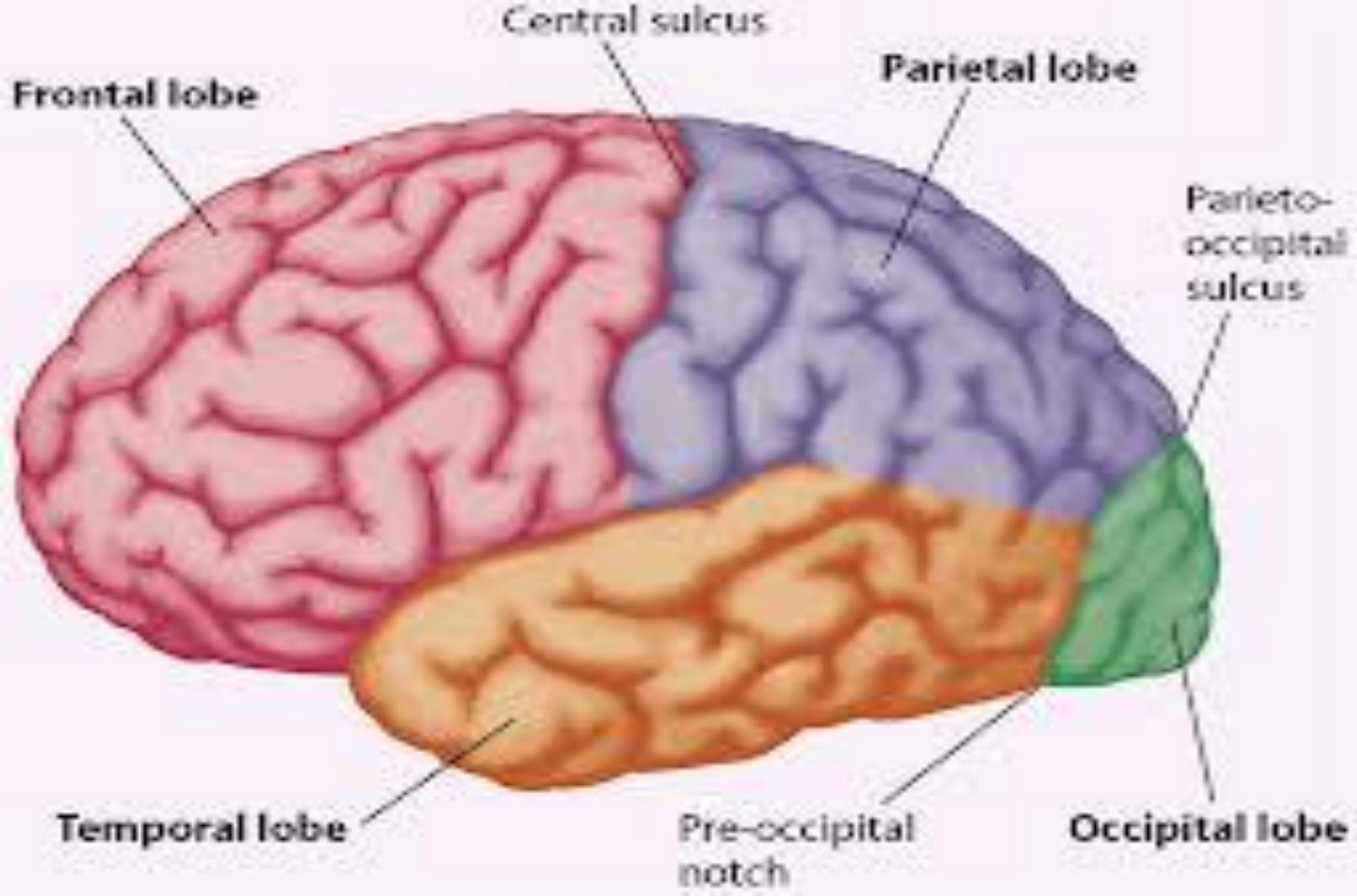

Ventricle III
Thalamus
Medial geniculate body
Pineal gland
Superior colliculus
Lateral geniculate body
Inferior colliculus
Trochlear nerve (CN IV)
Superior medullary velum
Superior cerebellar peduncle (brachium conjunctivum)
Middle cerebellar peduncle (brachium pontis)
Stria medullaris
Inferior cerebellar peduncle (restiform body)
Hypoglossal triangle
Clava
Vagal triangle
Cuneate tubercle
Obex
Fasciculus gracilis
Posterior median sulcus
Fasciculus cuneatus
Posterior intermediate sulcus

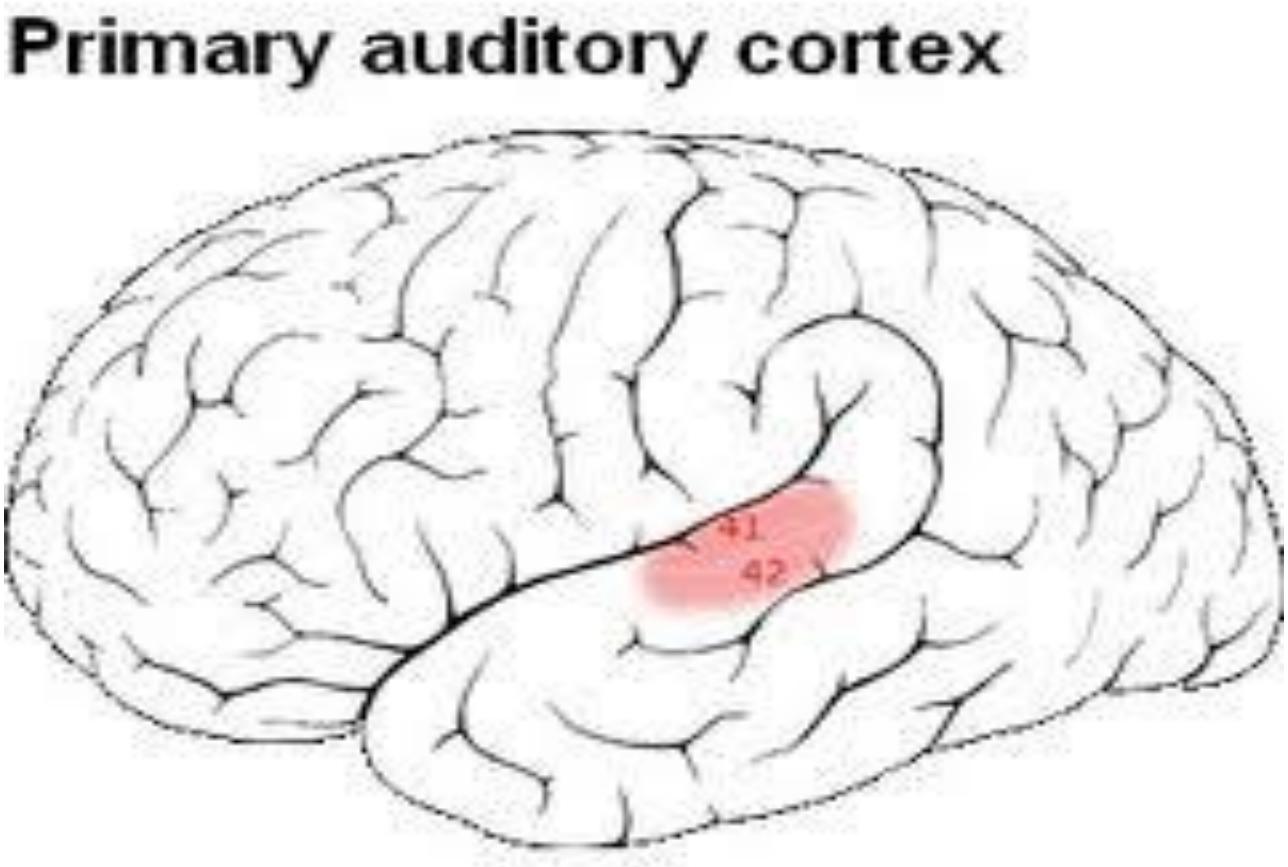
ANATOMY

- The cerebellopontine angle is a potential space in the posterior cranial fossa. Its boundaries are as follows:
- Anteriorly: Posterior fossa of the temporal bone
- Posteriorly: Anterior surface of the cerebellum
- Medially: Inferior olive
- Superiorly: Inferior border of the pons and cerebellar peduncle
- Inferiorly: The cerebellar tonsil
- The trigeminal nerve is visible superior to the cerebellopontine angle, whereas the IXth, Xth, and XIth nerves course inferiorly. Other important structures within the cerebellopontine angle include the anterior inferior cerebellar artery (AICA), flocculus, and lateral aperture of the fourth ventricle (foramen of Luschka).

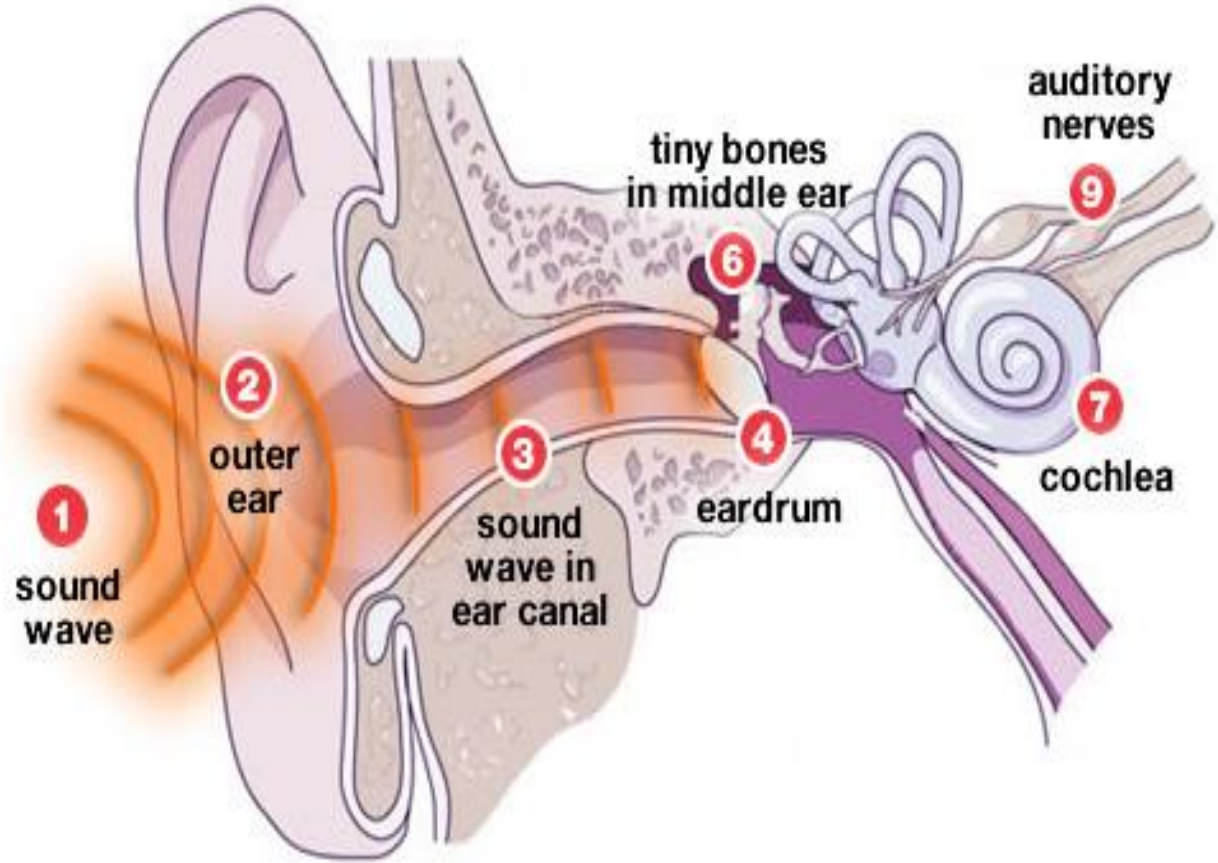
ANATOMY

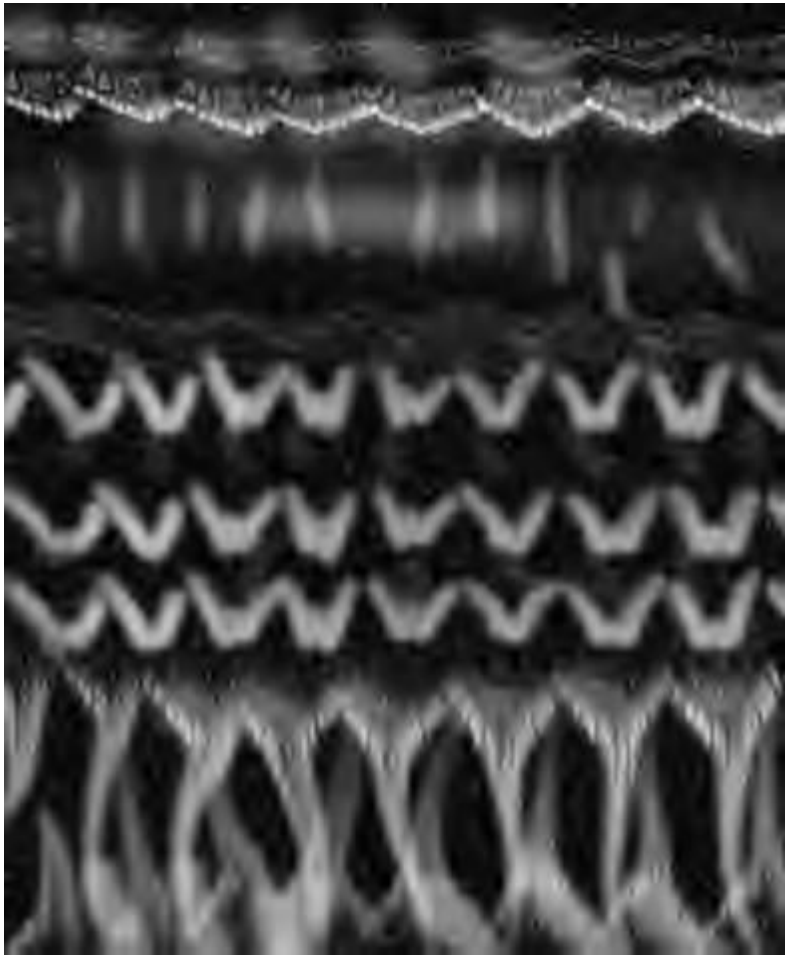

Central sulcus
Frontal lobe
Parietal lobe
Parieto-occipital sulcus
Temporal lobe
Pre-occipital notch
Occipital lobe

ANATOMY



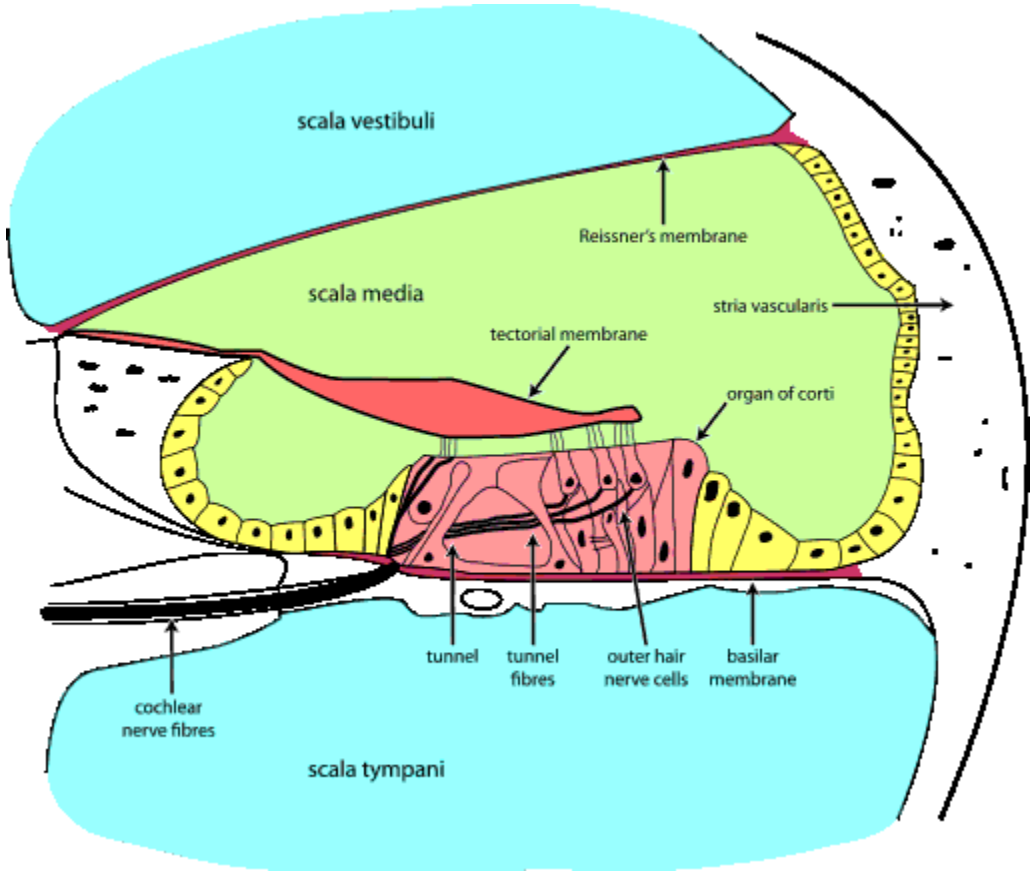
PHYSIOLOGY


auditory nerves
tiny bones in middle ear
9
6
2
outer ear
3
4
7
cochlea
1
sound wave
sound wave in ear canal
eardrum



Scanning electron micrograph of the upper surface of the organ of Corti after removal of the tectorial membrane. There are 3 rows of outer hair cells with their characteristic V-arranged stereocilia. A single row of inner hair cells with a slightly curved row of stereocilia is also shown.

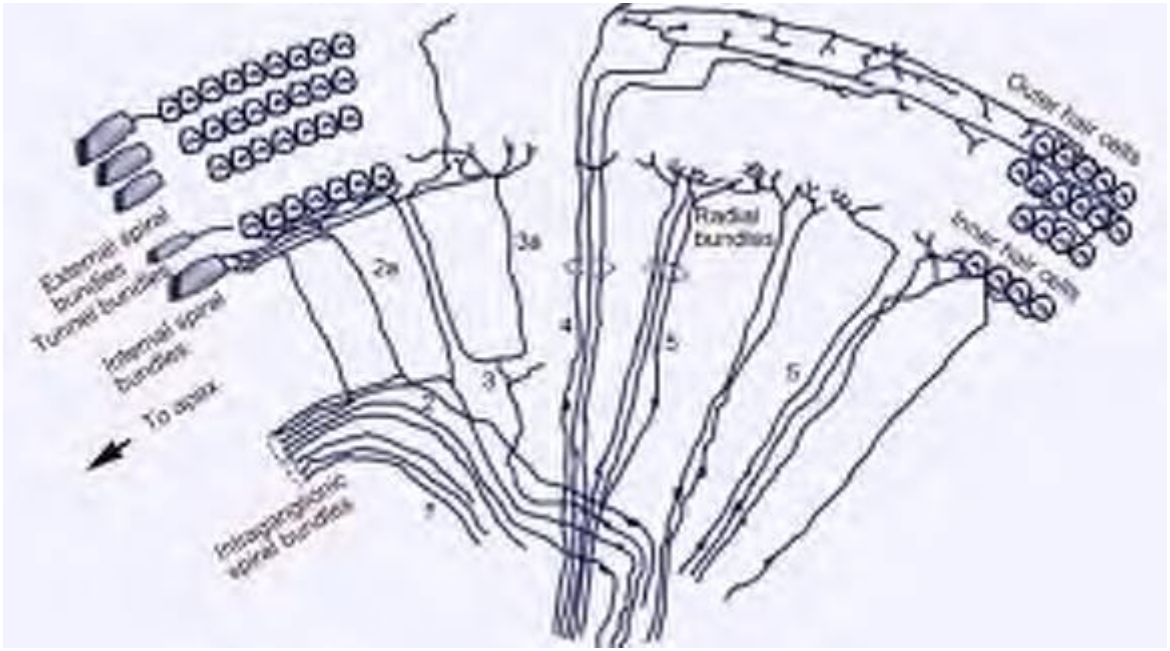
PHYSIOLOGY


scala vestibuli
Reissner's membrane
scala media
stria vascularis
tectorial membrane
organ of corti
tunnel
tunnel fibres
outer hair nerve cells
basilar membrane
cochlear nerve fibres
scala tympani

PHYSIOLOGY

- The fibers of the cochlear nerve originate from an aggregation of nerve cell bodies in the spiral ganglion, located in the modiolus of the cochlea.
- They are bipolar cells
- The longer central fibers form the cochlear nerve, and the shorter, peripheral fibers extend to the bases of the inner and outer hair cells.
- These primary auditory fibers(cochlear nv) exit the modiolus through the internal meatus and enter the medulla oblongata.

PHYSIOLOGY



- The cochlea's afferent innervation pattern. The illustration is shown through the vestibular (Reissner's) membrane, looking "down" on the spiral organ (of Corti). The principal fiber bundles are 1 and 2; 2a and 3a are internal spiral fibers; 4 are external spiral fibers; 5 and 6 are radial fibers innervating the inner hair cells.

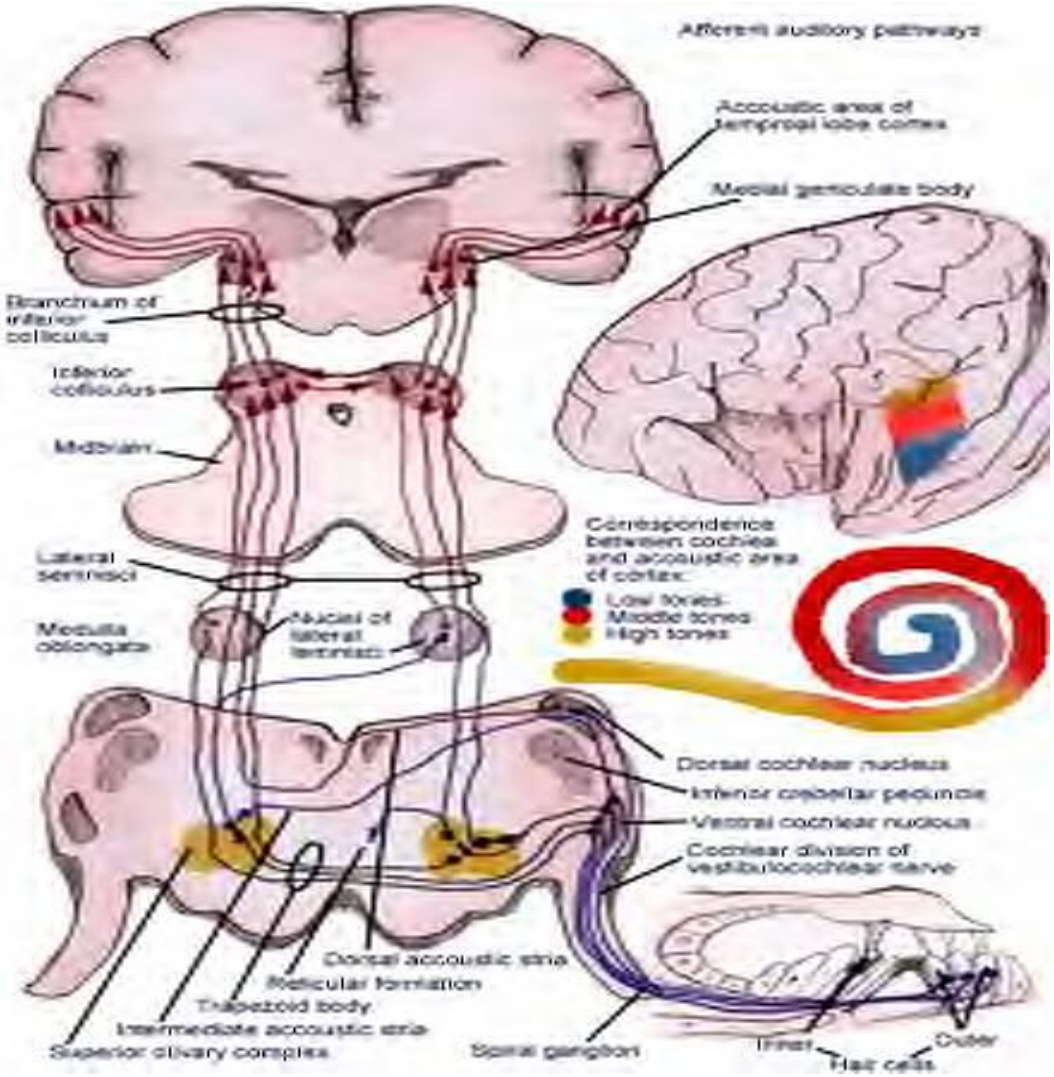


Illustration of the afferent auditory reflex pathway

Central auditory pathway

- E-COLI MA

- Eighth nerve →Cochlear nucleus →Superior olivary complex→ Lateral lemniscus →Medial geniculate body →Auditory cortex.

Central auditory pathway

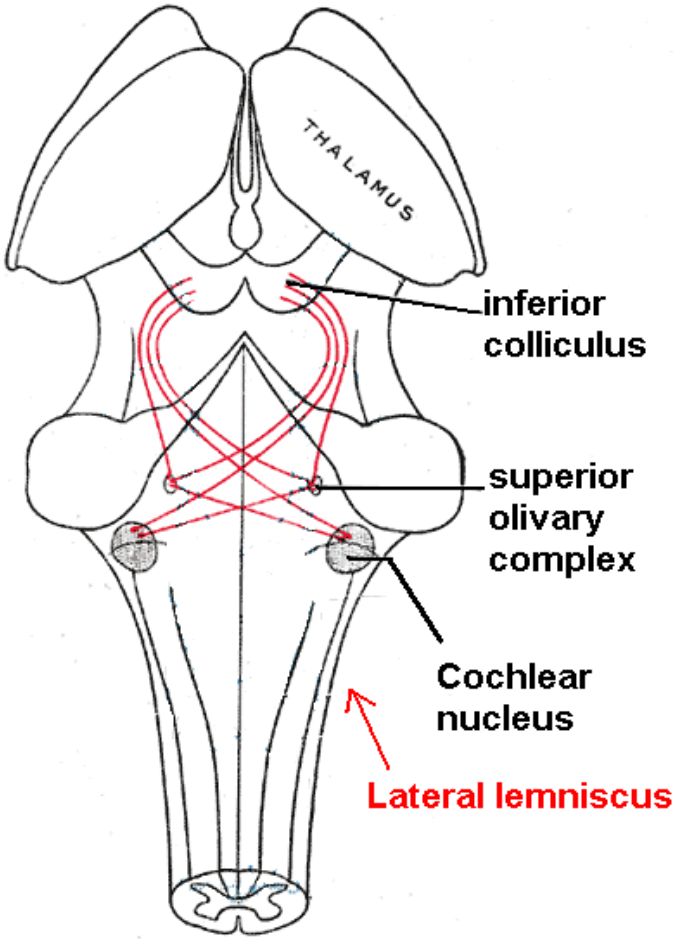
- After entering the medulla, the cochlear nerve fibers proceed to the cochlear nucleus.
- The cochlear nucleus is divided into the dorsal and ventral parts.
- The cochlear fibers divide into 2 main bundles: One group passes lateral and dorsal to the restiform body; the other group remains slightly ventral and medial to the restiform body and terminates in the ventral cochlear nucleus.

Central auditory pathway

- Some fibers from the ventral cochlear nucleus cross the midline to the cells of the superior olivary complex, whereas others make connection with the olivary cells of the same side.

- Fibers from the dorsal cochlear nucleus cross the midline to end on the cells of the nuclei of the lateral lemniscus. There they are joined by the fibers from the ventral cochlear nuclei of both sides and from the olivary complex.

Central auditory pathway



The lemniscus is a major tract, most of the fibers of which end in the inferior colliculus.

Central auditory pathway

- Some fibers may bypass the colliculus and end, together with the fibers from the colliculus at the medial geniculate body.

- From the medial geniculate body, there is an orderly projection of fibers to a portion of the cortex of the temporal lobe.

Central auditory pathway

- Primary acoustic area in the cerebral cortex is the superior transverse temporal gyri of Heschl.

- About half of the fibers of the auditory pathways cross the midline whereas others ascend on the same side of the brain, therefore each ear is represented in both the right and left cortex.

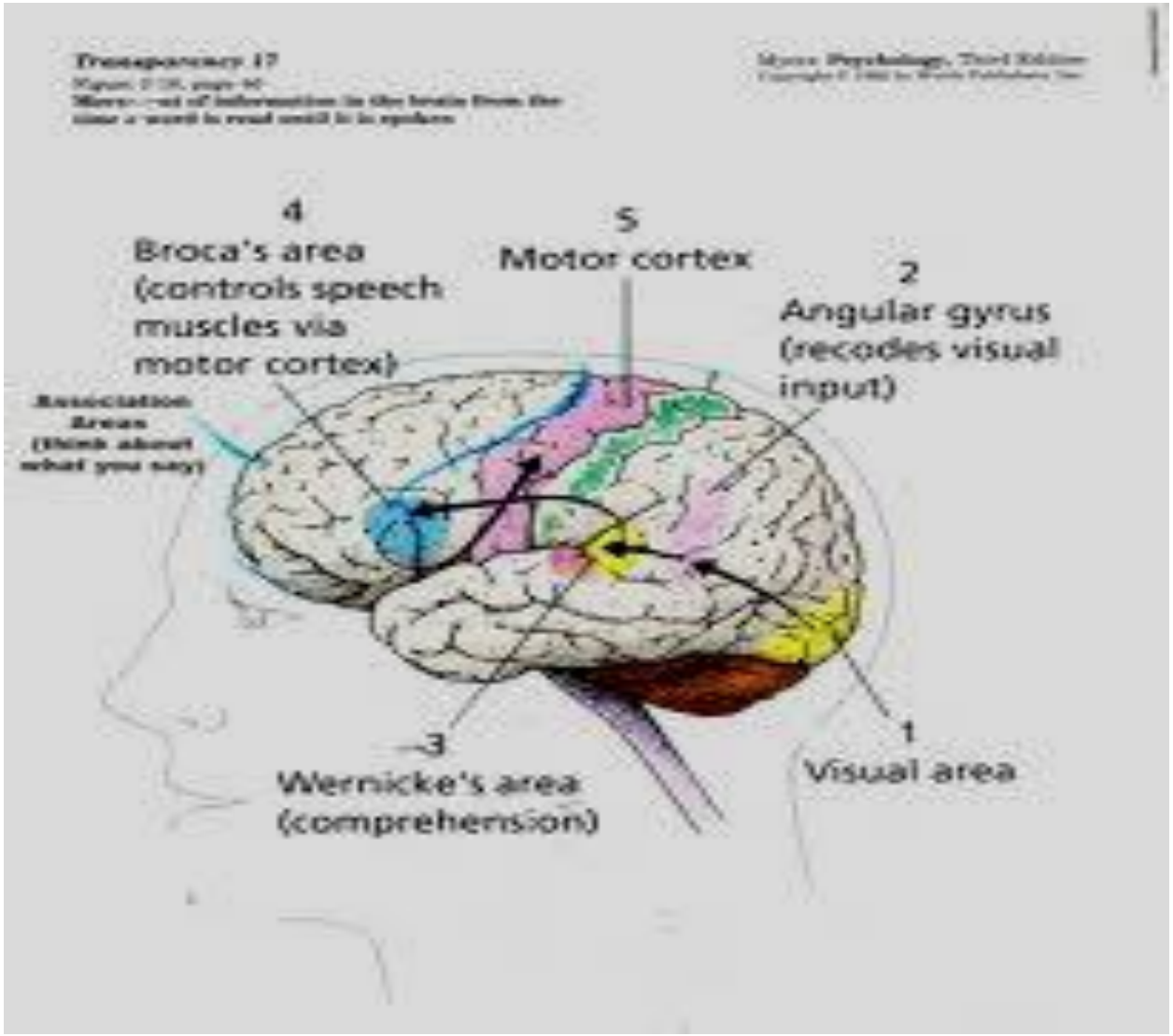
Central auditory pathway

- Brodmann 41 and 42, marking the location of the primary auditory cortex -responsible for the sensation of basic characteristics of sound such as pitch and rhythm.

- The auditory association area is located in Wernicke area or area 22. An important region for the processing of acoustic signals so that they can be distinguished as speech, music, or noise.

- Primary auditory cortex is surrounded by secondary auditory cortex, and interconnects with it.
- These secondary areas interconnect with further processing areas in the superior temporal gyrus, in the dorsal bank of the superior temporal sulcus, and in the frontal lobe.
- In humans, connections of these regions with the middle temporal gyrus are probably important for speech perception.

Central auditory pathway



Key Points about the Central Auditory pathway

- Summarized below are the concepts that are most important to know about the central auditory system:

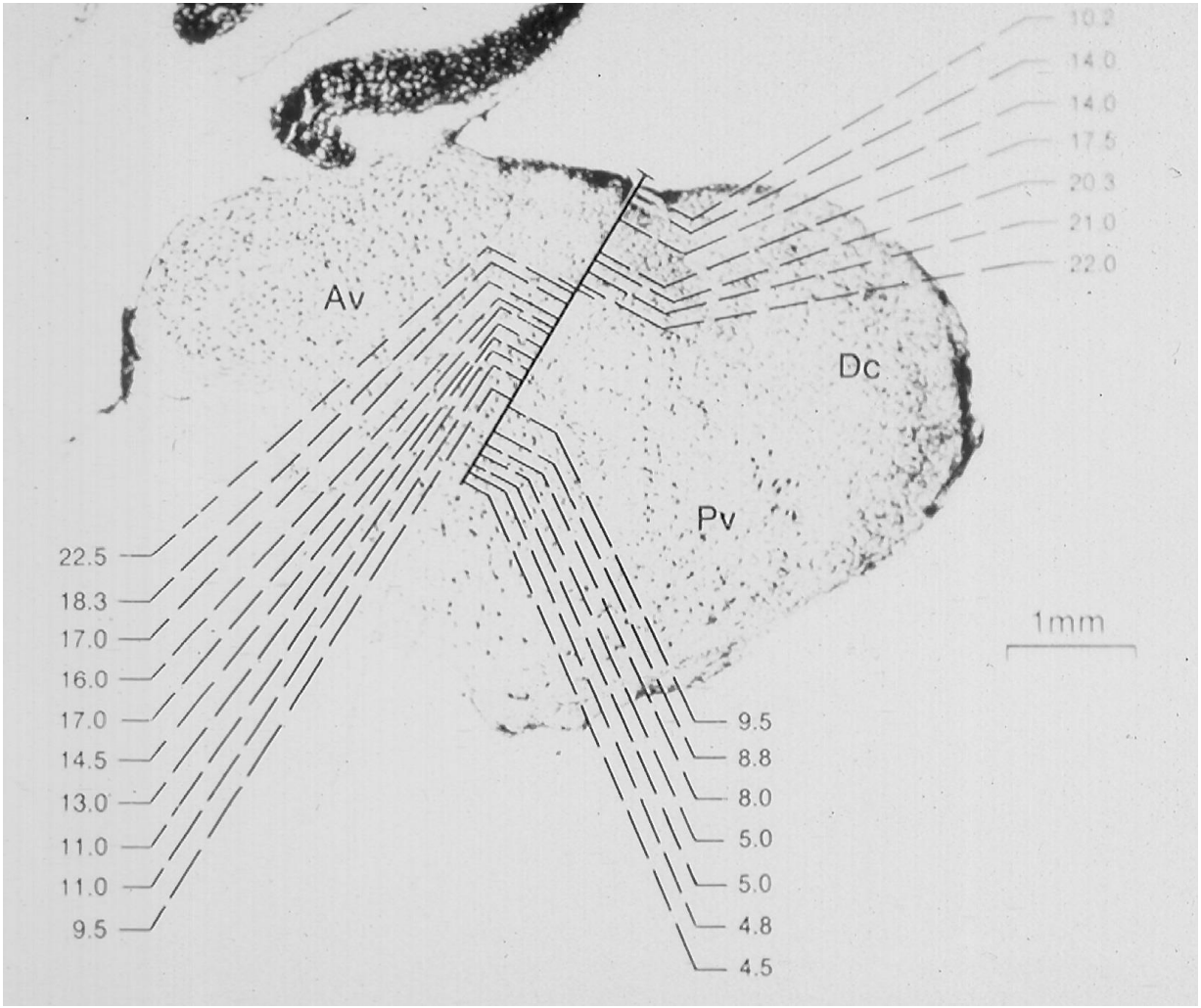
- ***1. Endpoints of the Auditory Pathway***. The pathway begins in the ***cochlear nucleus*** of the ***medulla*** and ends with the ***auditory radiations***, which run from the ***thalamus*** to ***auditory cortex***.

- ***2. General Architectural Features***

  (a) ***decussation*** **(crossing over)**
  (b) ***tonotopic organization***
  (c) ***specialization***

- a. Decussation. This is important for our ability to localize sound. Sound localization depends (in part) on differences between the two ears in time of arrival and intensity.

- b. Tonotopic Organization. This term refers to the relationship between the spatial location of nerve fibers and the characteristic frequency (CF) or best frequency (BF) of the fiber. Tonotopic organization is not just for 8th N fibers, but throughout the entire auditory pathway.



- Tonotopic organization in the central auditory pathway. The numbers below are CFs (in kHz) for a slice of tissue in the brain stem. The simple point to be made here is that the CFs are not randomly distributed – there is a relationship between CF and the physical location of the fiber, just like there is for 8th N fibers in the cochlea.

- c. Specialization
- Auditory nerve fibers (these are in the auditory periphery, not the CNS) show very little specialization; i.e., they're nearly interchangeable with one another, with one exception: Some have low thresholds (and high spontaneous rates), some have high thresholds (and low spontaneous rates). That's about it for specialization. Neurons in central auditory system, on the other hand, are specialized.

Four Different cell types in the cochlear nucleus

Structurally, the cells look different.

They also respond differently to input from other neurons.

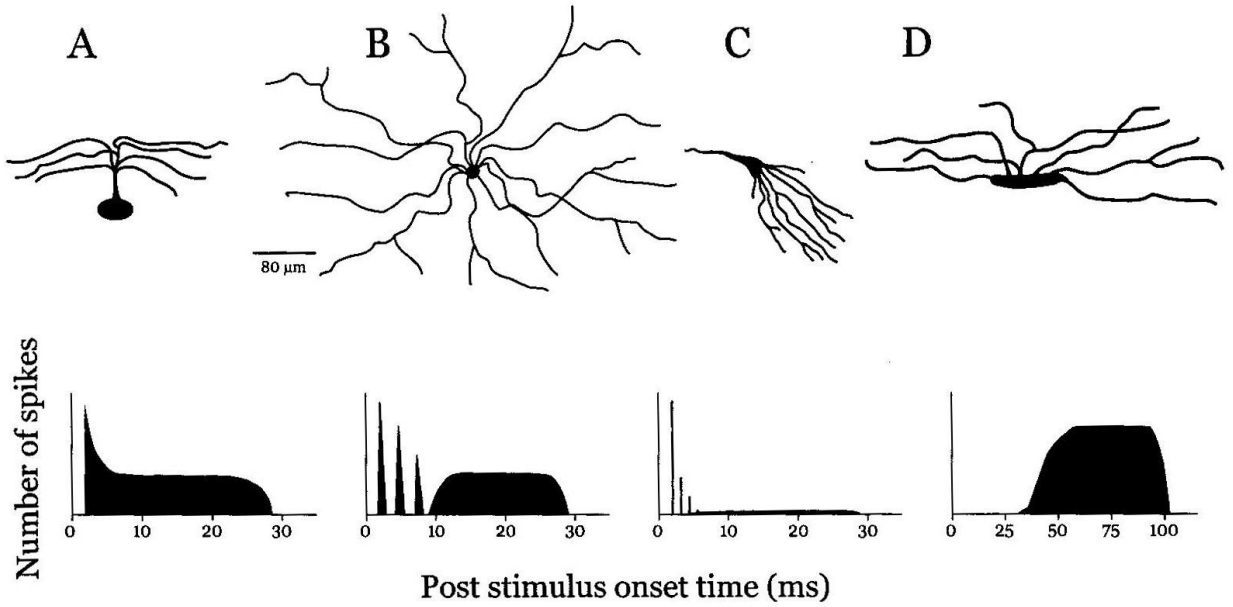


Figure 26-1. Upper drawings are stylized impressions of the morphology of 4 cell types of the cochlear nucleus, prompted by the descriptions of Oertel et al[21] and Rhode.[20] Lower panels are schematic, stylized impressions of the poststimulus time histograms of the responses to CF tones that might be expected from the neurons depicted (prompted by those presented by Rhode and Greenberg[18]).

Descending pathways of the auditory nerve

- The descending pathways have an inhibiting effect upon the ascending fibers.
- From the superior olivary complex a fiber tract called the olivocochlear bundle originates. It constitutes a feedback loop, by which nerve impulses, thought to be inhibitory, reach the hair cells.
- Efferent auditory pathways modulate the outer hair cells of the cochlea, protect against noise, and improve the detection of sound sources in noisy environments.

Physiology

- Following birth, the brain of a newborn is flooded with information from the baby's sense organs.
- At birth, each neuron in the cerebral cortex has approximately 2,500 synapses.
- By the time an infant is two or three years old, the number of approximately 15,000 synapses per neuron.
- This is about twice as many as an adult brain

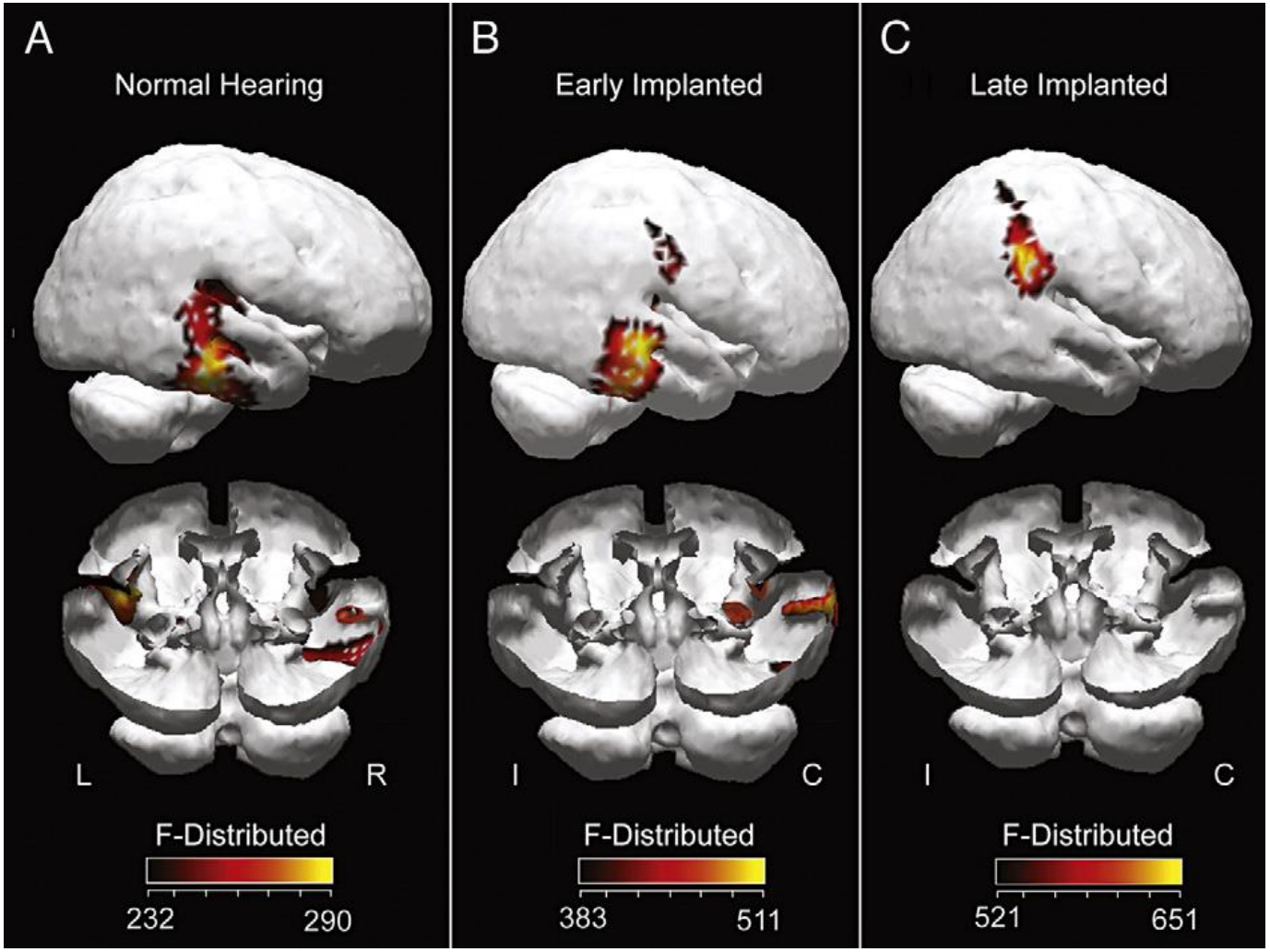
Synaptic pruning

- Synaptic pruning eliminates weaker synaptic contacts while stronger connections are maintained and strengthened.
- Experience determines which connections will be strengthened and which will be pruned; connections that have been activated most frequently are preserved.
- Ineffective or weak connections are "pruned" It is plasticity that enables the process of developing and pruning connections, allowing the brain to adapt itself to its environment.

Physiology

- Stimulation of the auditory centres of the brain influence the actual growth and organisation of the auditory pathways

Physiology


A
Normal Hearing
L
R
F-Distributed
232
290
B
Early Implanted
I
C
F-Distributed
383
511
C
Late Implanted
I
C
F-Distributed
521
651

Management

Presentation

- Poor/No response to sound
- Poor/No response to call
- Reduction in hearing
- Incidental
- Following screening

History

- Hearing loss- onset , course , side , exacerbating and relieving factors.
- Ear sympts-Tinnitus, Vertigo ,Ear discharge, Ear pain, Aural fullness.
- Nasal symptoms-Nasal discharge, Nasal obstruction
- Noise exposure
- Recent trauma
- Family history
- Medical history
- Drug history
- Other cranial nerve deficits

Examination

- General examination
- Ear
  - Pneumatic Otoscopy
  - Tuning fork tests
    - Webers
    - Rinnes
    - Schwabach's
- Nose
  - Anterior rhinoscopy
  - Nasopharyngscopy

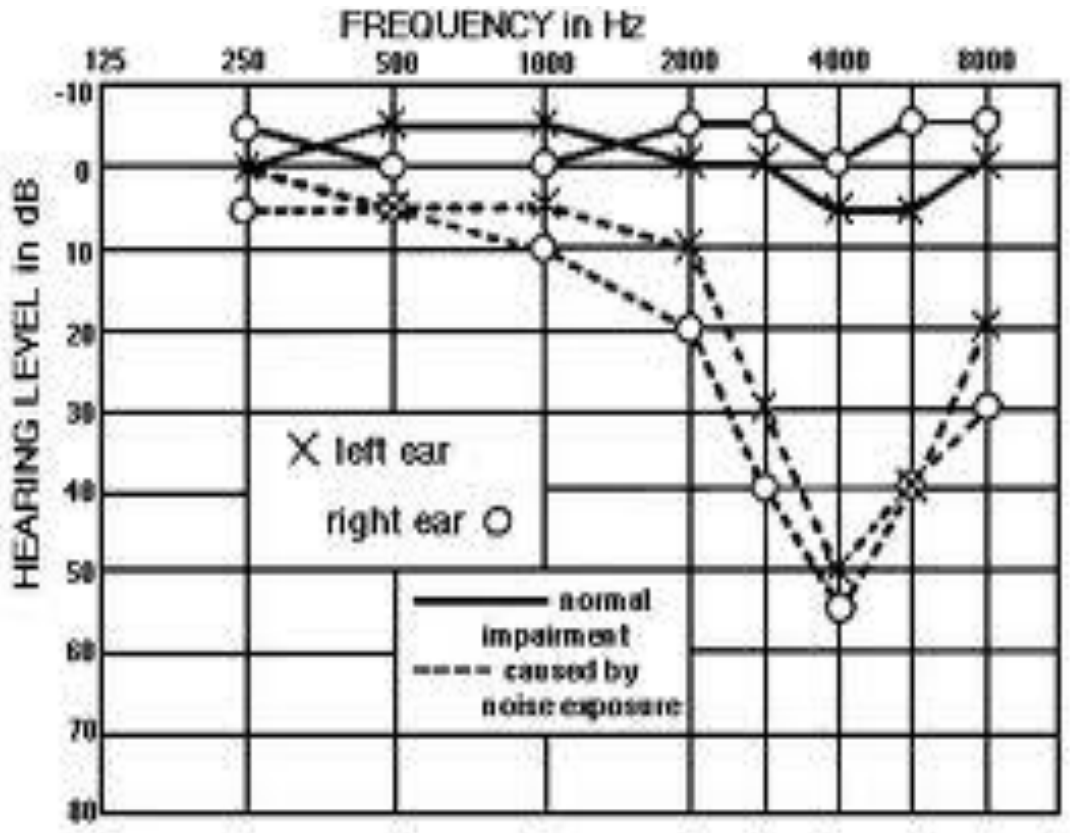
Examination

- Neck – goitre
- CVS
- CNS

Investigations ; audiological

- Tympanometry + acoustic reflex
- Pure tone audiometry
- Speech reception threshold test
- Speech discrimination test
- Auditory brainstem response
- Oto-acoustic emissions

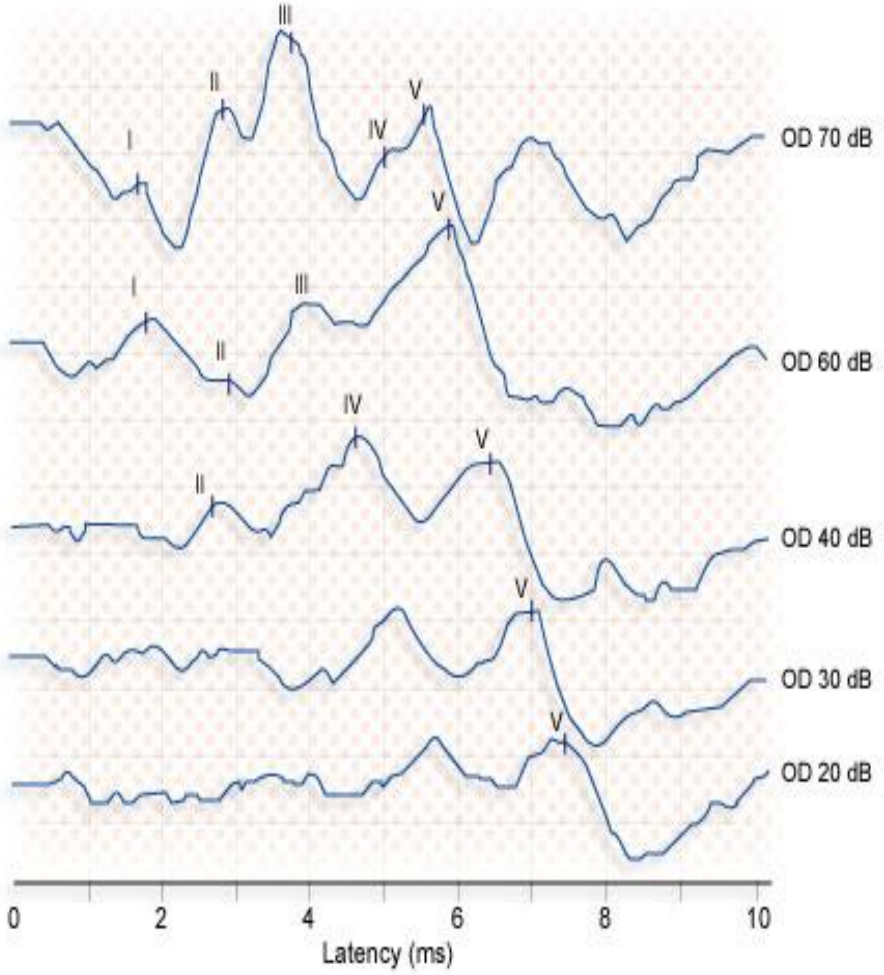
Pure tone audiometry



Auditory brainstem response

- When a hearing ear is given a stimulus, the resulting electrographic activity can be followed from the ear to central areas of the brain.
- CHL cannot be distinguished from SNHL with the screening test
- The sensitivity and specificity of this testing are near 100%.
- Requires sedation, they take time and are expensive. Abnormal brain-wave activity (eg, seizure activity, significant prematurity, movement artifact) can render the results uninterpretable.

Auditory brainstem response



ECOLI

- Wave I :Eighth nerve
- Wave II: Cochlear nucleus
- Wave III: Superior olive
- Wave IV: lateral lemniscus
- Wave V: Inferior colliculus

Oto-Acoustic Emissions

- Sounds generated by the inner ear can be recorded.
- The sounds may be spontaneous or evoked.
- The sensitivity and specificity reported with evoked OAE are 100% and 82%, respectively.
- Cannot be used to distinguish CHL from SNHL.
- High failure rate in the neonatal ICU.
- Quick and inexpensive by personnel with relatively little training.

Childhood Audiometry

- **Behavioural observation audiometry**: 0-6months; change in activity in response to sound
- **Visual reinforcement audiometry (VRA):**6 mths-2½ years. Child is asked to turn toward any sound and is rewarded with an entertaining visual image for responding.
- **Conditioned play audiometry (CPA):** In CPA, children between 2½ and 4 years of age are asked to perform a simple play activity (like placing a ring on a peg) when they hear a sound.

Investigations

- FBC
- E/U/Cr
- ECG
- Connexin-26
- Markers of general inflammatory disease (eg, ESR, RF)
- specific markers for autoimmune inner ear disease (e.g, 68-kd protein).
- CT scan
- MRI

OAE/ABR Setup

Classification

- Type
  - conductive
  - Sensorineural
  - mixed
- Severity
  - Mild 20-40 dB
  - Moderate 41-60 dB
  - Severe 61-80 dB
  - Profound >80 dB
- Age of onset
  - prelingual
  - Post-lingual
- No of ear(s)
  - unilateral
  - bilateral
- Genetic
  - Syndromic
  - non-syndromic

Syndromic hearing loss

External ear

- DiGeorge sequelae
- Branchio-oto-facial syndrome
- Townes-Brocks syndrome
- Miller syndrome
- Bixler syndrome

Cardiac

- Coloboma, heart disease, atresia choanae, retarded growth, and ear anomalies (CHARGE) syndrome
- Jervell Lange-Nielson syndrome
- Limb-oto-cardiac syndrome

Renal

- Alport syndrome
- Branchio-oto-renal syndrome
- Kearns-Sayre syndrome
- Epstein syndrome
- Barakat syndrome

Mental (retardation)

- Noonan syndrome
- Killian/Teschler-Nicola syndrome
- Cockayne syndrome, type I
- Gustavson syndrome

Dermatologic

- Waardenburg syndrome
- Lentigines, ECG, ocular, pulmonary, abnormal, retardation, and deafness (LEOPARD) syndrome
- Senter syndrome
- Black locks with albinism and deafness (BADS) syndrome
- Davenport syndrome

Syndromic hearing loss

Endocrine and/or metabolic

- Pendred syndrome
- Johanson-Blizzard syndrome
- Refetoff syndrome
- Wolfram syndrome
- Kallmann syndrome

Facial

- Goldenhar syndrome
- Frontometaphyseal dysplasia
- Escher-Hirt syndrome
- Levy-Hollister syndrome

Ophthalmologic

- Usher syndrome
- Marshall syndrome
- Alström syndrome
- Harboyan syndrome
- Fraser syndrome
- Jensen syndrome

Orthopedic

- Klippel-Feil sequelae
- Stickler syndrome

Craniometaphyseal dysplasia

- Oto-spondylo-megaepiphyseal dysplasia (OSMED) syndrome

Causes of hearing impairment

External ear

- Impacted cerumen
- Foreign bodies
- Otitis externa
- Congenital stenosis
- Bony exostosis
- Osteomas
- Tumors
- Cholesteatoma

Middle ear

- TM peforations
- Tympanosclerosis
- Ossicular malformations
- Ossicular discontinuity
- Otosclerosis
- Labyrintine fistula
- Haemotympanum
- Otitis media
- Tumors
- ETD
- Temporal bone fractures

Causes of hearing impairment

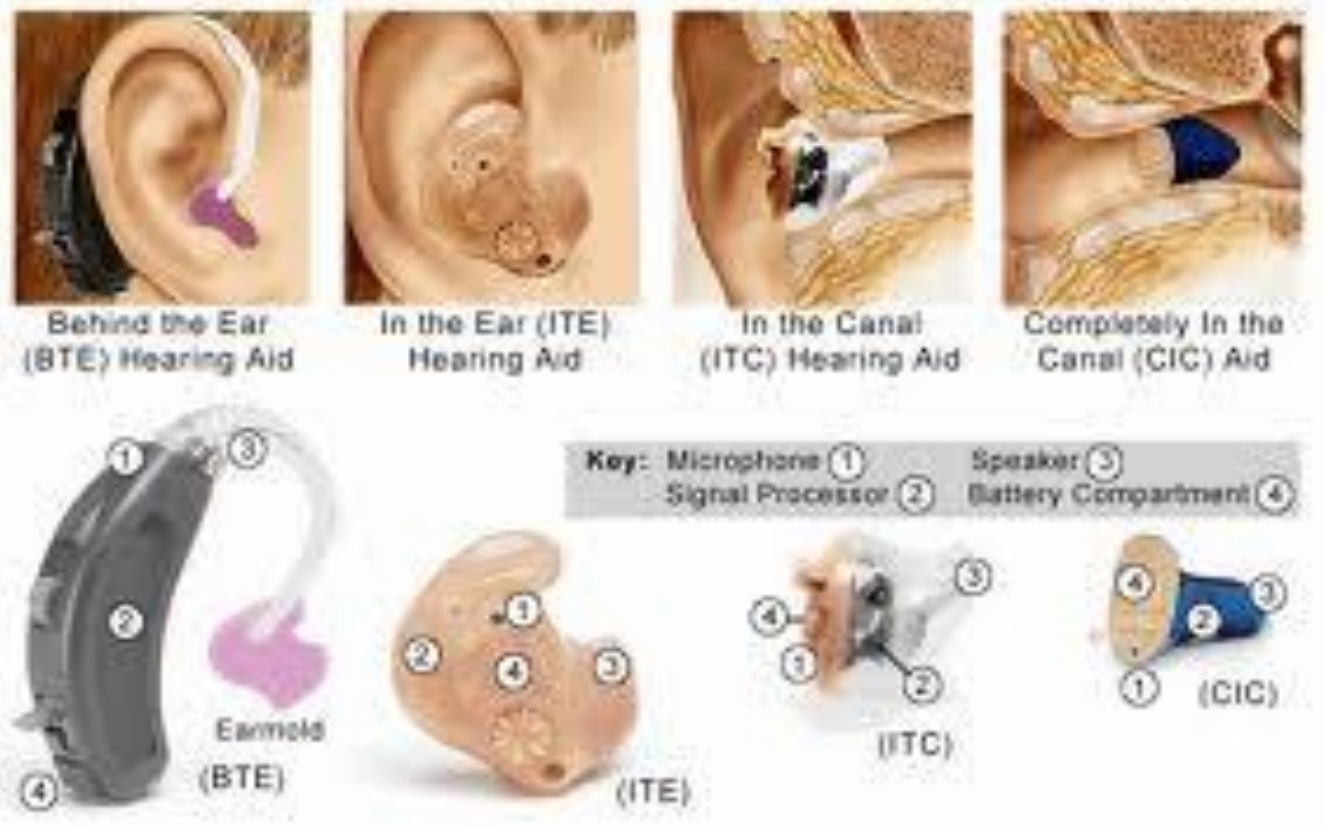
Inner ear

- Presbycusis
- Heredity
- Noise exposure
- Acoustic trauma
- Ototoxicity
- Infection
- Immune related
- Meniere's disease
- Genetic
- Sudden Idiopathic hearing loss
- Neurologic diseases e.g MS
- Tumors

Treatment

- Treat underlying cause
  - Medical- Antibiotics, Steroids, NSAIDS
  - Surgical- Myringotomy , Gromet insertion , Tympanoplasty , Ossiculoplasty , Stapedectomy , Mastoidectomy.
- Amplification
- Cochlear implantation
- Auditory brainstem implantation
- Deaf culture

Amplification



Cochlear implants(CI) and Auditory brainstem implants(ABI)

- CI- external part implanted under the skin above and behind the ear digitally encodes sound then sends them via FM to an implanted receiver in the cochlea which then stimulates the auditory nerve.

- ABI -bypasses the cochlea altogether and attaches its electrode directly to the brainstem. Provides sound information by direct stimulation of the cochlear nucleus to patients with dysfunctional or absent cranial nerve VIII.

Lip-reading (oralism) and sign language

- American Sign Language (ASL) or Signed English (SE) with Signing Exact English/Seeing Essential English (SEE). PSE (Pidgin Signed English) is used functionally.

- Almost half of the consonants in English look identical on the lips but are distinguishable to hearing people when spoken. E.g /ch/, /j/, and /sh/, making chew, Jew, and shoe indistinguishable from each other

Not deaf and dumb!

- Deaf and hard-of-hearing people who use either lip-reading or cued speech must learn to speak for themselves. They have varying success, but many can make themselves understood in most situations.

Screening

- The Centers for Disease Control and Prevention (CDC) recommends that all babies be screened for hearing impairment before 1 month of age, preferably before they leave the hospital after birth.

- Without newborn screening- not diagnosed until 2 to 3 years

Screening

- The American Academy of Pediatrics advises that children should have their hearing tested several times throughout their schooling:
- When they enter school
- At ages 6, 8, and 10,
- At least once during middle school
- At least once during high school

Screening

High-risk criteria for neonates (birth to 28 d) are as follows:

- Family history of congenital or early SNHL
- Congenital infection known to be associated with SNHL
- Craniofacial anomalies
- Birth weight of more than 1500 g (< 3.3 lb)
- Hyperbilirubinemia over the exchange level
- Exposure to ototoxic medications
- Bacterial meningitis
- Low Apgar scores at birth
- Prolonged mechanical ventilation
- Findings of a syndrome associated with SNHL

Screening

High-risk criteria for infants (29 d to 2 y) are as follows:

- Concern about hearing, speech, language, and/or developmental delay
- Bacterial meningitis
- Neonatal risk factors associated with SNHL
- Head trauma, especially with fracture of the temporal bone
- Findings of a syndrome associated with SNHL
- Exposure to ototoxic medications
- Neurodegenerative disorders
- Infectious diseases associated with SNHL

Follow-up

- Monitor the child's linguistic and social development.

- Children who cannot communicate with those around them may be frustrated and therefore, act out or display withdrawal behaviors. These behaviors may be misinterpreted as being a behavioral or psychological problem rather than being reactions to the child's environment or situation.

Follow-up

- **Children who are deaf or hard of hearing are at particular risk for abuse.** Physical abuse may be inflicted by parents who are frustrated because their child is not acting as they expect. Also, children who perpetrators perceive as being unable to report misconduct are at high risk for sexual abuse.

Follow-up

- The family should be encouraged to obtain devices such as strobe lights connected to doorbells, timers, alarm clocks, and fire alarms. Telecommunication Devices for the Deaf (TDDs) and teletypewriters (TTYs) are machines than enable deaf people to use the phone.

Follow-up

- Computers with modems or video/webcam, cell phones with text messaging or instant messaging, and other hand-held devices all permit children to communicate using modern technologies

- All new televisions are equipped with closed captioning, which decodes the captioning of dialogue and action provided with most television shows, videotapes, and DVDs.

Prevention

- It is estimated that half of cases of hearing impairment and deafness are preventable.
- A number of preventative strategies are effective including: immunization against rubella to reduce congenital infections, immunization against *H. influenza* and *S. pneumoniae* to reduce cases of otitis media, and avoiding or protecting against excessive noise exposure.
- Advocacy and Support groups.

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Thank you