International Journal of Otorhinolaryngology and Head and Neck Surgery
Munjal M et al. Int J Otorhinolaryngol Head Neck Surg. 2025 Aug;11(4):469-474
http://www.ijorl.com
pISSN 2454-5929 | eISSN 2454-5937
DOI: https://dx.doi.org/10.18203/issn.2454-5929.ijohns20252265

Review Article

The physiological and pathophysiological aspects of the human cochlea: a review

Manish Munjal[1], Shubham Munjal[1]*, Hardeep Kaur[1], Vineeta Arora[2], Aditi Randev[3], Sakshi Jeriwal[1], Prachi Bharadwaj[1], Namit Mittal[1], Navya Mittal[1], Anjana Pillai[4]

[1]Department of ENT-Head and Neck Surgery, Dayanand Medical College, Ludhiana, Punjab, India
[2]Guru Teg Bahadur Hospital, Ludhiana, Punjab, India
[3]Department of Medicine, Dayanand Medical College, Ludhiana, Punjab, India
[4]Department of Dental Surgery, BJS Dental College, Ludhiana, Punjab, India

Received: 02 May 2025
Revised: 15 May 2025
Accepted: 22 May 2025

***Correspondence:**
Dr. Shubham Munjal,
E-mail: manishmunjaldr@yahoo.com

ABSTRACT

The fluids of the membranous labyrinth are in a state of dynamic equilibrium with the intracranial cerebrospinal fluid. Mechanical shifts in the ear canal, middle and inner ear due to tympanic, ossicular, and basilar membrane displacements are likely to result in electrolyte alterations with the generation of nerve impulses. Sound wave, depending on its frequency, attains maximum amplitude at a particular site on the basilar membrane and stimulates that very segment. Higher frequencies are represented at the basal turn of the cochlea, and the progressively lower ones towards the apex. The disturbance in the milieu interior is consequent to auditory and vestibular impairments. Physiology of the cochlea is altered in varied conditions like hypertension, renal disease, syphilis, chronic suppurative otitis media, diabetes, and noise trauma. The unique physio-pathology of the membranous labyrinth shall be elaborated.

Keywords: Cochlea, Physiology, Pathology, Pathophysiology

INTRODUCTION

The human cochlea and the semicircular canals are protected in the bony labyrinth and have the perilymph, endolymph, and the cortilymph with different anionic and cationic compositions. Shifts in the electrolytes from one compartment to another due to mechanical transmission from the external to the middle to the inner ear are the basis of the generation of nerve impulses that are finally received by the auditory cerebral cortex. The physiology and patho-physiology of the cochlea need introspection.

REVIEW OF LITERATURE

Physiological aspects

Transduction of mechanical energy occurs to generate electrical impulses. The movements of the footplate of stapes, which are transmitted to the cochlear fluids, move the basilar membrane and set up a shearing force between the tectorial membrane and the hair cells. The distortion of hair cells is the origin of cochlear microphonics, which initiate a nerve impulse. A sound wave, depending on its frequency, attains maximum amplitude at a particular site on the basilar membrane and stimulates that very segment. Higher frequencies are represented at the basal turn of the cochlea and the progressively lower ones towards the apex.[1]

Neural pathways

Hair cells get innervations from the bipolar cells of spiral ganglion.

Central axons of these cells collect to form the cochlear nerve, which goes to the ventral and dorsal cochlear nuclei.

From there, both crossed and uncrossed fibers travel to the superior olivary nucleus, lateral lemniscus, inferior colliculus, and medial geniculate body, and finally reach the auditory cortex of the temporal lobe.[1]

Electrical potentials of the cochlea and cranial nerve VIII

Four types of potentials have been recorded: three from the cochlea and one from cranial nerve VIII fibers - endocochlear potential, cochlear microphonic, summating potential from cochlea, and compound action potential from nerve fibers.

Endocochlear potential

It is a direct current potential recorded from scala media. It is +80 mV and is generated from the stria vascularis by Na^+/K^+-ATPase pump and provides source of energy for cochlear transduction. It is present at rest and does not require sound stimulus. This potential provides a sort of "battery" to drive the current through hair cells when they move in response to a sound stimulus.

Cochlear microphonic

When basilar membrane moves in response to sound stimulus, electrical resistance at the tips of hair cells changes allowing flow of K^+ through hair cells and produces voltage fluctuations called cochlear microphonic. It is an alternating current potential.[2]

Summating potential

It is a direct current potential and follows "envelope" of stimulating sound. It is produced by hair cells. It may be negative or positive.[17] Summating potential has been used in diagnosis of Ménière's disease. It is superimposed on VIII nerve action potential. Both cochlear microphonics and summating potential are receptor potentials as seen in other sensory end-organs.

They differ from action potentials in that: they are graded rather than all or none phenomenon, have no latency, are not propagated, and have no post response refractory period.[2]

Compound action potential

It is an all or none response of auditory nerve fibers.

Vestibular nerve

Vestibular or Scarpa's ganglion is situated in the lateral part of the internal acoustic meatus. It contains bipolar cells. The distal processes of bipolar cells innervate the sensory. epithelium of the labyrinth while its central processes aggregate to form the vestibular nerve.

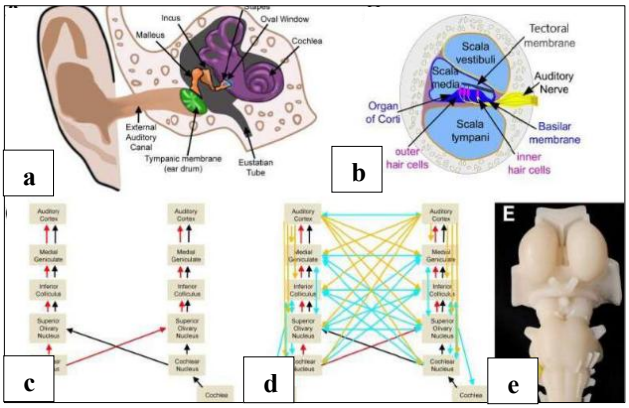

Figure 1: Auditory circuits, (a) outer, middle, and inner ear, (b) cross section of the cochlea, (c) ascending auditory pathway, (d) ascending (red/black), descending (cortical: orange; brain-stem: blue), and crossed (blue) auditory circuits, and (e) model representation of an acoustic neuroma.[3]

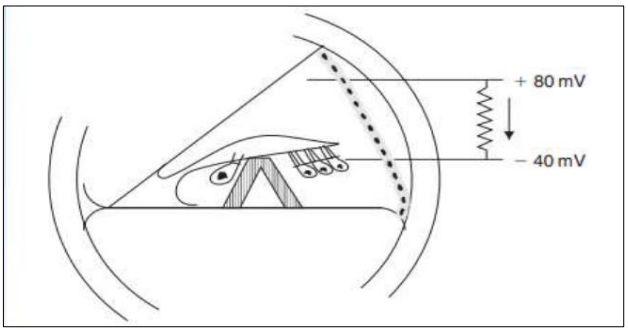

Figure 2: Davis battery model of transduction.[2]

Inner ear fluids and their circulation

Perilymph resembles extracellular fluid and is rich in sodium ions. It fills the space between the bony and the membranous labyrinth. It communicates with CSF through the aqueduct of cochlea which opens into the scala tympani near the round window. In fact, this duct is not a direct communication but contains connective tissue resembling arachnoid through which perilymph percolates. There are two views regarding the formation of perilymph: it is a filtrate of blood serum and is formed by capillaries of the spiral ligament, and it is a direct continuation of cerebrospinal fluid and reaches the labyrinth via aqueduct of cochlea.[4]

Endolymph fills the entire membranous labyrinth and resembles intracellular fluid, being rich in potassium ions. It is secreted by the secretory cells of the stria vascularis of the cochlea and by the dark cells (present in the utricle and also near the ampullated ends of semicircular ducts). There are two views regarding its flow: longitudinal, i.e. endolymph from the cochlea reaches saccule, utricle and endolymphatic duct and gets absorbed through endolymphatic sac, which lies in the subdural space, and

radial, i.e. endolymph is secreted by stria vascularis and also gets absorbed by the stria vascularis.[4]

Physiology of outer hair cells

The groups of cells that are responsible for hearing are the outer hair cells located in the inner ear. These hair cells are sensitive to low sound pressure levels. This property of outer hair cells is defined as the process of "cochlear amplification". Failure of function of and loss of outer hair cells leads to hearing loss in old age or due to chronological medical conditions such as diabetes mellitus.[5]

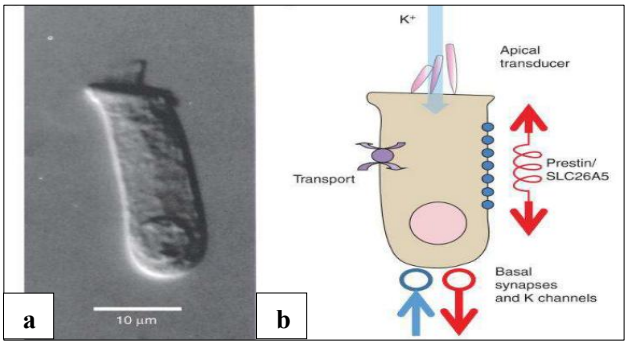

Figure 3: The elements of the mammalian outer hair cell (OHC), (a) an isolated OHC from turn 2 (~5–10 kHz region) of the guinea pig cochlea, the apical stereocilia are apparent at the apical surface, and (b) schematic OHC showing the location of prestin/SLC26A down the basolateral surface, generating longitudinal forces. Transport to regulate pH is presumed to be collocated as a parallel property of prestin. K+ ions from the scala media enter through the mechanoelectric transducer channels at the apex and exit through the basally located K+ channels. Both afferent (red) and efferent (blue) terminals are located at the base of the cell.[5]

Mammalian outer hair cells possess a very characteristic V-shaped bundle of stereocilia, serving to inject current into the cells when deflected. As sensors of the movement of the basilar membrane, outer hair cells thus form part of a local mechanical-electrical-mechanical feedback loop to control basilar membrane tuning.

There are several constraining observations: the lateral membrane of the outer hair cells is packed with a particle about 8 nm in diameter; a change in the outer hair cell membrane potential is accompanied by a gating charge movement, or equivalently the cell membrane capacitance is voltage dependent; the charge movement is blocked by the amphiphilic anion salicylate (aspirin, with an additional methyl group has the same effect); and outer hair cells only acquire motile properties progressively during a short period of development. The identification of prestin by using a subtracted complementary DNA (cDNA) library prepared from the messenger RNA (mRNA) of isolated hair cells in principle, solves most of these problems.[6]

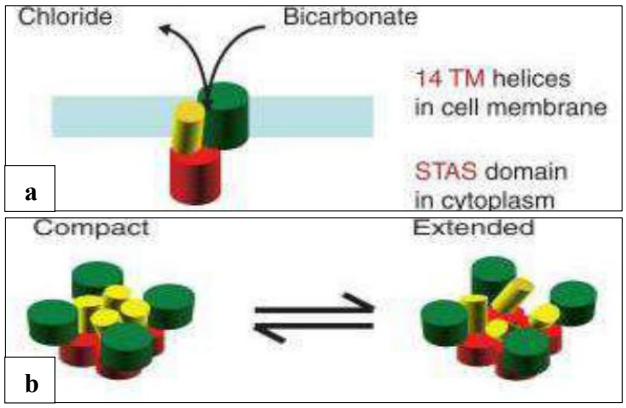

Figure 4: Models for prestin, (a) organization of prestin/SLC26A5 in the membrane as a monomer. Two mobile components of the protein are placed in the membrane, whereas the carboxy-terminal region, containing the sulfate transporter and anti-sigma factor antagonist (STAS) domain, is cytoplasmic, and (b) Hypothetical model for co-assembly into a tetramer. The carboxy-terminal region (red) forms a base against which the oligomer can deform, rapidly shifting between a compact (cell depolarized) and an extended configuration (cell hyperpolarized).

PATHOLOGICAL ASPECTS

Effect of hypertension on the cochlea

Kirbac et al study on the effects of primary arterial hypertension on cochlear function included 34 patients with primary hypertension and 17 healthy adults. The Cochlear function was assessed with conventional audiometry (0.125-8 kHz), ultra-high frequency audiometry (10-16 kHz), the transient evoked otoacoustic emission (TEOAE) test, and the distortion product otoacoustic emission (DPOAE) test. Hearing thresholds at 8, 10, 12.5, 14, and 16 kHz were significantly poorer in the hypertensive group than in the control group. There was no significant difference in the mean conventional thresholds between the groups. Compared to the control group, the patient group exhibited statistically significantly lower amplitudes of TEOAE and DPOAE.[7]

Effect of renal disease on cochlea

Bergstrom et al study on renal disease with respect to pathology, treatment, and effects on the ear included twenty-six hemodialysis and renal transplant patients who were studied for the effects of age, duration of kidney failure, amounts and types of treatment, disturbed calcium metabolism, and vascular disease on clinical and pathologic inner ear manifestations. Three subgroups of patients were compared with each other: those having hearing loss of unknown etiology, those having strial deposits, and those having neither, and severity of

treatment was not necessarily predictive of severity of inner ear disease. Within each subgroup, there was wide variability and almost complete overlap for the factors studied when the three groups were compared. Cochlear strial deposits seem to bear little relationship to disturbed calcium metabolism or vascular disease, but their size may be related to the presence of hearing loss.[8]

Effect of syphilis on the cochlea

Hızlı et al study on stria vascularis and changes in the cochlear hair cells in syphilis included 13 human temporal bone samples from 8 patients with syphilis (syphilis group), as well as 12 histopathologically normal samples from 9 age-matched patients without syphilis (control group) comparison was made between the two groups, the mean area of the stria vascularis (measured with conventional light microscopy connected to a personal computer) and the mean percentage of cochlear hair cell loss. It was concluded that syphilis led either to complete loss of the organ of Corti or to significant loss of cochlear hair cells. But the area of the stria vascularis did not show any change.[9]

Effect of chronic suppurative otitis media on the cochlea

Cureoglu et al retrospective study on cochlear changes in chronic otitis media was conducted on fifteen temporal bones with unilateral chronic otitis media and compared with contralateral normal temporal bones. The loss of outer and inner hair cells was common in the basal turn of the cochlea in temporal bones with chronic otitis media compared with control ears. There was no difference in the number of spiral ganglion cells in the chronic otitis media and contralateral ears. The areas of stria vascularis and spiral ligament in the basal turn decreased significantly in the ears with chronic otitis media compared with control ears. There were no significant differences between the ears with chronic otitis media and the contralateral ears for any of the regions characterized by the presence of types I-IV fibrocytes.[10]

Effect of noise on the cochlea

In the Le et al study on noise-induced hearing loss, degeneration of auditory nerve fibers, along with loss of outer hair cells both was seen in temporal bone histopathology in a mouse model, while damage to inner hair cells was limited. Destruction of the organ of Corti is consequent to two mechanisms: 23 mechanical destruction by short exposure to extreme noise intensity or metabolic decompensation after noise exposure over a longer period of time. Noise intensity levels above 130 dB sound pressure cause to mechanical destruction of the organ of Corti, disruption of cell junctions, and mixing of endolymph with perilymph. The pathology observed as a result of metabolic decompensation includes stereocilia disruption, swollen nuclei, swollen mitochondria, cytoplasmic vesiculation, and vacuolization. Current theories of metabolic damage center on the formation of free radicals or reactive oxygen species (ROS), emerge immediately after noise exposure and persist for 7–10 days thereafter, spreading apically from the basal end of the organ of Corti, thus widening the area of necrosis and apoptosis.[11]

Effect of diabetes on cochlea

Akinpelu et al analysed the effects of type 2 diabetes mellitus on hearing function. They concluded that the incidence of hearing loss ranged between 44% and 69.7% for type 2 diabetics, significantly higher than in controls. The mean pure tone audiometric thresholds were greater in diabetics than in controls, and auditory brainstem response (ABR) wave V latencies were also statistically significantly longer in diabetics when compared to control groups.[12]

Hao et al conducted a study on the early detection of hearing impairment in patients with diabetes mellitus utilizing otoacoustic emissions. The study concluded that the distortion product otoacoustic emission (DPOAE) amplitudes in diabetics were significantly lower than those in controls.[13] Hou et al study on auditory impairment in young type 1 diabetics concluded that type 1 diabetics exhibited higher auditory threshold, slower auditory conduction time, and cochlear impairment.[14] Cho et al interpreted auditory brainstem evoked responses and distortion product otoacoustic emissions in 24 diabetic patients with normal hearing. 45 ears in patients with type 2 diabetes mellitus and 85 ears in non-diabetic patients. The study concluded that Diabetic subjects showed significantly more prolonged absolute peak

Latencies (I, III, V) and inter-peak latencies (I-V, III-V) than non-diabetic subjects.[15]

Spankovich et al DPOAE study in young adults with type-1 diabetes revealed a reduced cochlear function.[16] Botelho et al study in young patients with type 1 diabetes concluded that abnormal DPOAEs responses were found more frequently than the alterations in TEOAEs and pure-tone audiometry, suggesting that DPOAEs evaluation is the most sensitive and it could be used for monitoring the progression of cochlear damage during the early stages of hearing impairment.[17]

Di Nardo et al study concluded that IDDM patients show an early abnormality of the micromechanical properties of the outer hair cells and damage is limited to the higher frequencies and can be detected only by DPOAEs.[18]

Lisowska et al analysis concluded that the mean amplitudes of various DPOAEs were significantly reduced in the diabetic group compared with control subjects. ABR latencies were longer in diabetic patients when compared with those of control subjects.[19]

Ottaviani et al otoacoustic emission study concluded that the mean intensity of the response was lower in diabetic

subjects than in controls.[20] Akcay et al study concluded significant decrease was also found in distortion product otoacoustic emission amplitudes at 4, 6, 8, and 10 kHz in the high blood glucose and diabetes mellitus groups compared to the control group.[21]

DISCUSSION

The know-how of the physiology of the cochlea is essential for cochlear implantation. The latter utilizes electricity to directly stimulate the cells of the spiral ganglion of the auditory nerve, thereby circumventing the damaged peripheral organ of hearing and hence providing hearing in individuals with sensorineural hearing impairment. The cochlear implant electrode is inserted via the round window and is retained in the scala tympani. The electrode directly stimulates the VIIIth nerve, facilitating transmission of neural signals to the central auditory cortex of the brain. A cochlear implant is only effective in a relatively anatomically preserved cochlea and the auditory nerve.[22]

Most loop diuretics are ototoxic, as they inhibit the Na/K/2Cl co-transporter in the stria vascularis. They affect the patho-physiology of the cochlea by the formation of edematous spaces in the stria vascularis epithelium, with consequent reduced cochlear electrical potential and, thereby, a long-term use, temporary or permanent sensorineural hearing deficit.[23]

Clinically, disturbances in normal cochlear physiology present as hearing impairment. Cochlear dysfunction reigns topmost should be in the differential diagnosis in individuals with reduced hearing of different grades.[24]

CONCLUSION

On Vis a Vis, the physiology of the cochlea is altered in varied conditions like hypertension, renal disease, syphilis, chronic suppurative otitis media, diabetes, and noise trauma. The auditory impairment detectable by the stepladder audiological test battery, can affect different frequencies depending on the site of involvement in the cochlea.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

REFERENCES

1. Ahmed ZM, Goodyear R, Riazuddin S, Lagziel A, Legan PK, Behra M, et al. The tip-link antigen, a protein associated with the transduction complex of sensory hair cells, is protocadherin-15. J Neurosci. 2006;26(26):7022-34.
2. Miller JD, Rothenberg SJ, Eldredge DH. Preliminary observations on the effects of exposure to noise for seven days on the hearing and inner ear of the chinchilla. J Acoustical Soc Am. 1971;50(4B):1199-203.
3. Peterson DC, Reddy V, Launico MV, Hamel RN. Neuroanatomy, Auditory Pathway. In: StatPearls. Treasure Island (FL): StatPearls Publishing. 2023.
4. Martin P. Active hair-bundle motility of the hair cells of vestibular and auditory organs. Inactive processes and Otoacoustic emissions in hearing. New York, NY: Springer New York. 2008;93-143.
5. Ashmore J. Outer Hair Cells and Electromotility. Cold Spring Harb Perspect Med. 2019;9(7):a033522.
6. Dallos P. Cochlear amplification, outer hair cells and prestin. Curr Opin Neurobiol. 2008;18(4):370-6.
7. Kirbac A, Boke B. Effects of primary arterial hypertension on cochlear function. Acta Oto-Laryngologica. 2021;141(2):158-62.
8. Bergstrom L, Thompson P, Sando I, Wood RP. Renal disease: Its pathology, treatment, and effects on the ear. Arch Otolaryngol. 1980;106(9):567-72.
9. Hızlı Ö, Kaya S, Hızlı P, Paparella MM, Cureoglu S. Stria vascularis and cochlear hair cell changes in syphilis: A human temporal bone study. Auris Nasus Larynx. 2016;43(6):614-9.
10. Cureoglu S, Schachern PA, Paparella MM, Lindgren BR. Cochlear changes in chronic otitis media. The Laryngoscope. 2004;114(4):622-6.
11. Le TN, Straatman LV, Lea J, Westerberg B. Current insights in noise-induced hearing loss: a literature review of the underlying mechanism, pathophysiology, asymmetry, and management options. J Otolaryngol-Head Neck Surg. 2017;46(1):41.
12. Akinpelu OV, Mujica-Mota M, Daniel SJ. Is type 2 diabetes mellitus associated with alterations in hearing? A systematic review and meta-analysis. The Laryngoscope. 2014;124(3):767-76.
13. Hao J, Hua L, Fu X, Zhang X, Zou Q, Li Y. Genome-wide DNA methylation analysis of human peripheral blood reveals susceptibility loci of diabetes-related hearing loss. J Hum Genet. 2018;63(12):1241-50.
14. Hou Y, Xiao X, Ren J, Wang Y, Zhao F. Auditory impairment in young type 1 diabetics. Arch Med Res. 2015;46(7):539-45.
15. Cho WK, Kang WS, Lee JB, Park HJ, Chung JW, Ahn JH. Interpreting auditory brainstem evoked responses and distortion product otoacoustic emissions in diabetic patients with normal hearing. Auris Nasus Larynx. 2021;48(2):227-34.
16. Spankovich C, Long GR, Hood LJ. Early indices of reduced cochlear function in young adults with type-1 diabetes revealed by DPOAE fine structure. J Am Acad Audiol. 2019;30(06):459-71.
17. Botelho CT, Carvalho SA, Silva IN. Increased prevalence of early cochlear damage in young patients with type 1 diabetes detected by distortion product otoacoustic emissions. Int J Audiol. 2014;53(6):402-8.
18. Nardo WD, Ghirlanda G, Paludetti G, Cercone S, Saponara C, Ninno MD, et al. Distortion-product

otoacoustic emissions and selective sensorineural loss in IDDM. Diabetes Care. 1998;21(8):1317-21.
19. Lisowska G, Namysłowski G, Morawski K, Strojek K. Otoacoustic emissions and auditory brain stem responses in insulin dependent diabetic patients. Polish Otolaryngol. 2002;56(2):217-25.
20. Ottaviani F, Dozio N, Neglia CB, Riccio S, Scavini M. Absence of otoacoustic emissions in insulin-dependent diabetic patients: is there evidence for diabetic cochleopathy? J Diabetes Complic. 2002;16(5):338-43.
21. Akcay G, Danısman B, Basaranlar G, Guzel P, Derin N, Derin AT. The effect of increase in blood glucose level on hearing loss. Braz J Otorhinolaryngol. 2022;88(Suppl 3):95-102.
22. Gillihan N, Singh B, Jones W, Alexander M, Cader A. CSF Gusher Complication: Literature review on Cochlear Implants and Outcomes. Available at: https://digitalcommons.pcom.edu/cgi/viewcontent.cgi?article=2008&context=research_day. Accessed on 12 May 2025.
23. Ding D, Liu H, Qi W, Jiang H, Li Y, Wu X, et al. Ototoxic effects and mechanisms of loop diuretics. J Otol. 2016;11(4):145-56.
24. Lim R, Brichta AM. Anatomical and physiological development of the human inner ear. Hear Res. 2016;338:9-21.

Cite this article as: Munjal M, Munjal S, Kaur H, Arora V, Randev A, Jeriwal S, et al. The physiological and pathophysiological aspects of the human cochlea: a review. Int J Otorhinolaryngol Head Neck Surg 2025;11:469-74.