

Hearing Loss and the Aging Ear

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Multiple factors lead to the cause and progression of presbycusis or hearing loss secondary to increased age. Noise trauma, genetic predisposition and oxidative damage all have been implicated in its development. Tinnitus, a frequent side effect of hearing loss, often has its origin within the central auditory system. A classification scheme for hearing loss and treatment options for tinnitus are discussed.

Key words: hearing loss, presbycusis, tinnitus, free radicals, central auditory system.

Presbycusis, or hearing loss secondary to increased age, has multiple factors leading to its cause and progression. Hearing loss, depending on its severity, can be debilitating and isolating. Unfortunately, auditory dysfunction increases in prevalence as one ages. Data have shown that hearing begins to deteriorate more rapidly for both men and women after the fourth decade, and many theories explain this increased loss with age, each with reliable data to support its claim. This leads to the conclusion that it is not just one factor but an interaction of multiple causes, some more influential than others, that leads to a gradual decline in hearing sensitivity with age.

Etiological Factors

Noise Exposure

Multiple auditory insults presented to us by our environment are contributing factors to presbycusis. This repeated exposure can traumatise the auditory system and lead to a slow progressive hearing loss. In an industrialised society, one is bombarded repeatedly throughout life with loud sounds that traumatise the inner ear by damaging hair cells and causing small membrane breaks, thus causing temporary or permanent threshold shifts. Depending on one's occupation, hobbies and place of residence, this repeated trauma may be more relevant in certain individuals. A classic investigation established that

there was remarkably little hearing impairment in old age among the isolated Mabaan tribe in southern Sudan.¹ This lack of progressive hearing loss with age in isolated, non-industrialised populations was further demonstrated in a tribe in India.² These studies of isolated populations give credence to the theory that presbycusis is a result of accumulated cochlear trauma sustained through one's lifetime. However, although environmental trauma seems to play an important role in the development of presbycusis, it is naïve to view this as the only cause. Free radical damage and genetic inheritance may play equally important roles in the development of hearing loss.

Mitochondria and Free Radicals

The free radical theory of aging may have potential implications for hearing impairment. In an experimental study, McFadden, *et al.* found a premature development of hearing loss when superoxide dismutase, an antioxidant enzyme, was expressed in low concentrations.³ Furthermore, the hearing loss the animals sustained was more severe. Free radicals are highly reactive molecules with one unpaired electron. They are produced during oxidative phosphorylation in the mitochondria. Mitochondrial DNA (mtDNA), with its simple circular structure and lack of sophisticated repair mechanisms, are

more prone to damage than other DNA. The free radicals usually are neutralised by scavenging enzymes before they can cause any damage, but the body's normal defense against these damaging molecules becomes less efficient as an individual ages.⁴ This loss of efficiency allows the free radicals to accumulate and cause damaging reactions to the surrounding molecules. One theory proposes that if ingestion of an exogenous antioxidant increases, one could potentially ward off the inevitable advancement of presbycusis. The ingestion of antioxidants like lecithin, a polyunsaturated phosphatidylcholine, has been shown to reduce free radical damage to mtDNA.⁵

Mitochondrial mutations have been shown to contribute to several human diseases and play an integral role in the aging process. Certain mitochondrial mutations also have been associated with the development of presbycusis. Bai, *et al.* found the 4,977 mtDNA deletion and its association with presbycusis.⁶ Fischel-Ghodsian and colleagues also discovered that people with presbycusis have a disproportionately high rate of mtDNA mutations when compared to age-matched controls.⁷

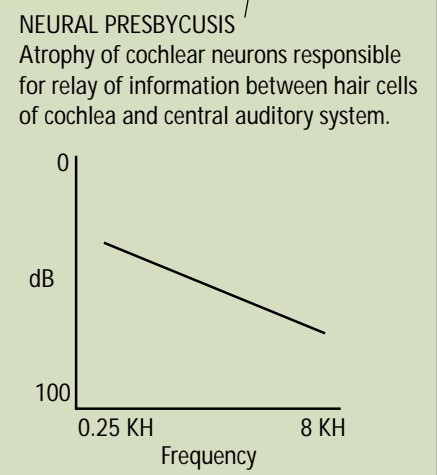
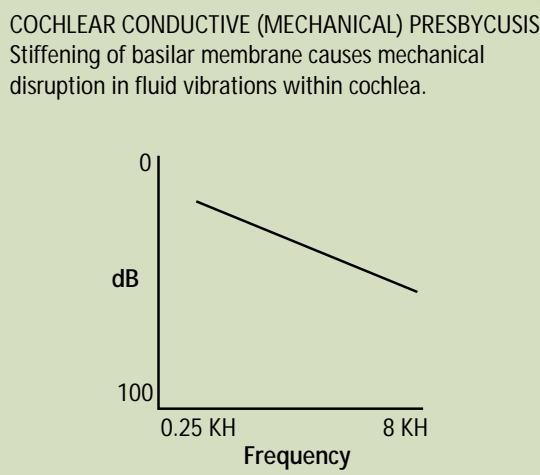
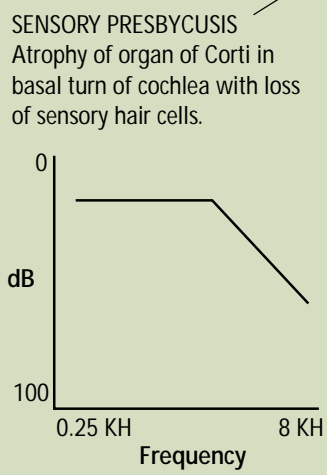
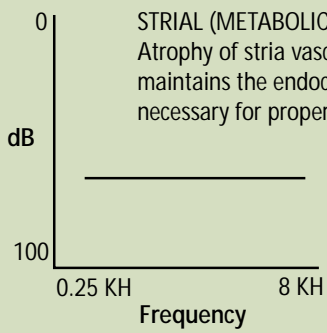
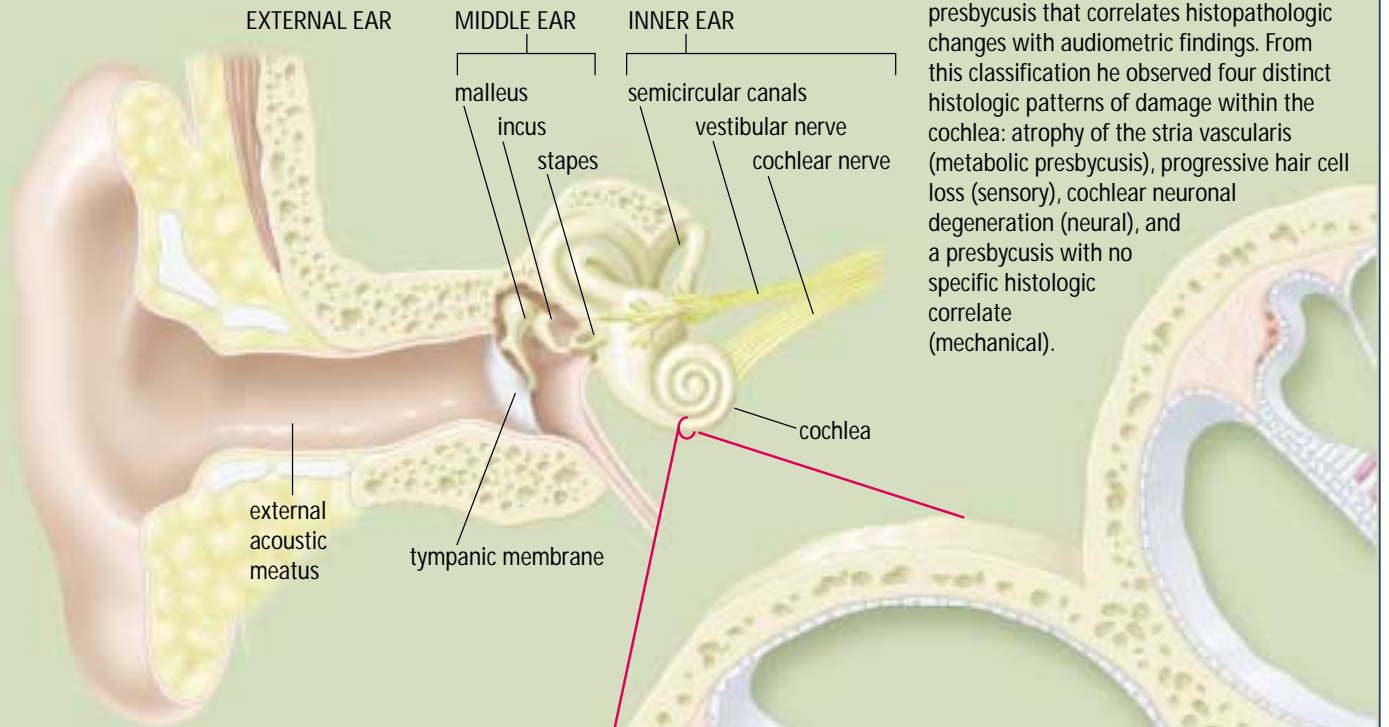
Genetic Factors

Certain hereditary traits may be involved in the development of presbycusis. Historically, this inherited predisposition to age-related hearing loss has been thought to play a role in the development of presbycusis, but only recently has there been data to support this hypothesis. DeStefano, *et al.* recently published a genomewide linkage analysis that suggests hearing loss in the low-to-mid-frequencies is heritable and possibly connected to certain types of congenital hearing loss.⁸

Noise trauma, mitochondrial DNA damage from oxidative insults and genetic predispositions all play a theoret-

Classification of Presbycusis

Schuknecht devised a classification of presbycusis that correlates histopathologic changes with audiometric findings. From this classification he observed four distinct histologic patterns of damage within the cochlea: atrophy of the stria vascularis (metabolic presbycusis), progressive hair cell loss (sensory), cochlear neuronal degeneration (neural), and a presbycusis with no specific histologic correlate (mechanical).



ical role in the acquisition of presbycusis. These potential causes manifest in different areas of the inner ear, enabling the classification of presbycusis into different subtypes.

Classification of Presbycusis

One of the most commonly used classification systems for presbycusis is one that was developed by Harold F. Schuknecht. Schuknecht, a renowned otopathologist, characterised presbycusis by coordinating audiometric findings with pathologic changes in the ear.⁹ Based on these pathologic changes, he observed four distinct histologic patterns of damage within the cochlea (Figure). Atrophy was noted in the organ of Corti, stria vascularis and spiral ganglia. The fourth change is attributed to an alteration within the basal membrane. The criteria for the diagnosis of presbycusis is “sensorineural hearing loss characterised by insidious onset, bilateral symmetry, and progression into old age without any clinical evidence of other ear disorders.”⁹

Sensory Presbycusis

Sensory presbycusis is characterised by atrophy of the organ of Corti in the extreme basal end of the cochlea. This damage is very common in the aged cochlea but rarely progresses far enough to reach the speech frequencies.⁹ This loss of hair cells in the basal end of the cochlea is manifested clinically by good hearing in the low- to mid-frequencies followed by a precipitous loss in the high frequencies; speech discrimination is rarely affected. The loss is indolent, with a progression of months to years. The genetic implications of presbycusis are just beginning to undergo investigation. Currently, no definite genetic cause can be attributed to this subclass of hearing loss.

Strial (Metabolic) Presbycusis

Strial (metabolic) presbycusis affects the stria vascularis, which is important in maintaining the endocochlear

potential necessary for proper hair cell function. Typically, there is patchy atrophy of the stria vascularis in the middle and apical turns of the cochlea,⁹ and there may be partial or complete loss of strial cells.

Strial atrophy has been seen to follow a familial pattern of inheritance but no gene can be directly attributed to its cause. There seems to be a direct correlation between the extent of strial loss and the magnitude of hearing impairment.⁹ Again, the loss is insidious and usually affects patients in the third to sixth decades. All frequencies are affected equally so that strial presbycusis is represented on the audiogram as a flat line with a threshold shift that does not typically go beyond 60–70dB. The ability to discern speech remains high.

Neural Presbycusis

Histologically, neural presbycusis is caused by an atrophy of the cochlear neurons. It has been shown that cochlear neurons slowly degenerate as an individual ages. On average, 37,000 neurons are identified in the cochlea in the first decade of life, dropping to approximately 18,000 by the ninth decade.¹⁰ This conglomeration of neural tissue, specifically called the spiral ganglion, acts as the relay between the hair cells of the cochlea and the central auditory system. Elderly individuals with rapid neural hearing loss will often manifest other general neurodegenerative symptoms, such as motor weakness, lack of coordination and tremor.

Neural presbycusis is manifested audiometrically by a down-sloping hearing loss similar to that seen with mechanical presbycusis, but with a disproportionate decrease in speech discrimination. Of the four groups of hearing loss, this is the only one with poor speech recognition scores. The unique finding of poor speech recognition scores and relatively good pure tone thresholds represents the lack of functional neural tissue in light of a functioning inner ear hair cell system.

Cochlear Conductive (Mechanical) Presbycusis

It is hypothesised that cochlear conductive (mechanical) presbycusis is caused by a stiffening of the basilar membrane and lacks any specific histologic correlate. This stiffening is thought to cause a mechanical disruption in the fluid vibrations within the cochlea. Hyalinisation and deposition of calcium salts have been found within the aging basilar membrane and may contribute to its stiffening. Audiometric findings show a straight down-sloping hearing loss, with the higher frequencies more affected than the lower frequencies that is slowly progressive. Speech discrimination is good.

Central Auditory System

One should not to be lulled into believing that cochlear pathology is the primary cause of hearing difficulty in the elderly. Presbycusis is a combination of deterioration of function in the central as well as the peripheral auditory pathways.¹¹ By testing cochlear function directly with different types of otoacoustic emissions (OAE), and by testing the central auditory pathways with speech audiometry, intensity discrimination and temporal resolution tests, Mazelova, *et al.* was able to support the view that the peripheral and central auditory systems are equally affected by presbycusis.^{12,13} Kirikae found several signs of deterioration in the medial geniculate body and auditory cortex among patients with presbycusis, demonstrating the role the central nervous system has in the development of age-related hearing loss.^{12,14} GABA production in the central nucleus of the inferior colliculus also has been shown to decrease with age.¹⁵

As the ear ages, there are changes that take place in the cochlea as well as in the central auditory pathways. These changes are most frequently manifested by increasing hearing impairment, but also may be accompanied by an equally, if not more, distressing symptom—tinnitus.

Tinnitus

Tinnitus is a common complaint that often accompanies presbycusis. There are many causes, ranging from drug toxicity, aberrant middle ear blood flow and stapedial muscle spasm.^{16,17} Tinnitus may be classified as objective (able to be heard by others) or subjective (only heard by the patient). For the purposes of this article, we will concern ourselves only with subjective tinnitus. It is often believed that the origin of tinnitus lies within the cochlea. However, it is not uncommon for individuals who have had the auditory nerve cut to also complain of ringing. Several studies have suggested a central cause of tinnitus. Lack of sensory input will often lead to a reorganisation of the central pathways. This theory of neural reorganisation, which has been described for phantom pain phenomena, also has been adopted for explanations of tinnitus. Lack of sensory input causes a lack of negative feedback to the dorsal cochlear nucleus, and thus increases activity in the central auditory system. This increase in activity leads to an auditory perception that the patient describes as hissing, blowing or ringing.^{16,17} Of note, if the tinnitus is not symmetric but more prominent in one ear, it may be representative of an acoustic tumour and further evaluation is warranted.¹⁶

Several medications have been studied in hopes of finding a remedy to help patients cope with this annoyance that can be quite disabling. Benzodiazepines have been shown to be effective, especially when trying to sleep. We usually start a patient on 0.5mg of alprazolam at bedtime, and slowly increase frequency to three times a day until symptoms are controlled. Benzodiazepines, however, have addictive properties and symptoms tend to recur when treatment is stopped. Another treatment option is with tricyclic antidepressants, and nortriptyline (50–150mg per day) has shown efficacy.¹⁷ Non-pharmacologic options include a masking device, provided by the audiologist, that provides

an extrinsic white noise to drown out the intrinsic tinnitus.¹⁷ Biofeedback techniques, often taught through psychology departments, also have demonstrated some success in helping patients cope with the annoyance of tinnitus.

Conclusion

Unfortunately, there is little we can do to modulate the course of presbycusis. Aural amplification currently is our only option to rehabilitate individuals with hearing loss (see article, page 19). Through continued research, we hope to be able to halt the progression or reverse the course of age-related changes in the inner ear. ♦

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