Presbyacusis A Review

Balasubramanian Thiagarajan

Stanley Medical College

Abstract:

Life expectancy of Humans is on the rise. World’s human population is aging rapidly. According to a report of US census Bureau nearly 24% of current world population is above the age of 50. This is a phenomenal number. Hearing loss due to degenerative process is rather common in old age. Presbyacusis is defined as a progressive bilateral symmetrical age related sensorineural hearing loss. The hearing loss is confined to higher frequencies. This causes a greater burden on them considering the fact that they already have failing eyesight. This article attempts to review the current scenario pertaining to Presbyacusis with a review of current published literature on the subject.

Introduction:

Presbyacusis is defined as a progressive bilateral symmetrical age related sensorineural hearing loss. The hearing loss is confined to higher frequencies. Presbyacusis is an added problem for the elderly who have a tendency to compensate for their loss of vision through their intact sense of hearing. They even tend to get isolated and become a social recluse due to this problem. Among the changes that occur with progressing age, deterioration of hearing is the most expected and a most accepted decline in the quality of life. Impact of decline in hearing could really be very profound with consequences involving social, functional and psychological well being of the person. Some of these problems can still persist despite the use of appropriate hearing aid. Studies reveal that nearly 75% above the age of 60 manifested with greater than 45 dB sensorineural hearing loss.

Factors responsible for Presbyacusis:

Various factors have been postulated as causes of Presbyacusis. They are:

1. Hereditary: Features like early aging of the cochlea and susceptibility of the cochlea for drug insults are genetically determined.

2. Atherosclerosis: May diminish vascularity of the cochlea there by reducing its oxygen supply.
3. Dietary habits: Increased intake of fatty diet may accelerate atherosclerotic changes in old age.

4. Diabetes: May cause vasculitis and endothelial proliferation in the blood vessels of the cochlea thereby reducing its blood supply.

5. Noise trauma: Exposure to loud noise on a continuing basis stresses the already hypoxic cochlea hastening the Presbyacusis process.

6. Smoking: Is postulated to accentuate atherosclerotic changes in blood vessels aggravating Presbyacusis.

7. Hypertension: Causes potent vascular changes, like reduction in blood supply to the cochlea thereby aggravating Presbyacusis.

8. Ototoxic drugs: Ingestion of ototoxic drugs like aspirin may hasten the process of Presbyacusis.

Presbyacusis traditionally has been divided into 4 groups depending on the location of pathology and patterns of hearing loss. This has amply been proved by temporal bone analysis.

1. Sensory hearing loss (outer hair cell loss)
2. Neural hearing loss (Ganglion cell hearing loss)
3. Metabolic (atrophy of stria vascularis)
4. Cochlear conductive (stiffness of basilar membrane) 

This classification is not absolutely rigid. Alternate theories suggest a combination of these patterns associated with central causes.

Hereditary causes of Presbyacusis:

Many genes and genetic pathways have been implicated in the etiopathogenesis of Presbyacusis. These include:

1. Mitochondrial dysfunction
2. Mitochondrial DNA Mutations
3. Mitochondrial haplotypes

It has been postulated that these genetic causes promotes age related sensori neural hearing loss.
Pathophysiology of age related hearing loss:

The auditory system conducts sound and converts it into electrophysiological signals that could be interpreted by the brain. There are a number of stages that are involved in this process of transduction of sound energy into electrical stimulus. Ageing process can affect any stage from stimulus to perception.

Figure showing the auditory pathway

- External ear
- Tympanic membrane and middle ear
- Inner ear
- Cochlear nerve
- Cochlear nuclei (medulla)
- Olivary nuclei
- Inferior colliculus (midbrain)
- Medial geniculate body
- Primary auditory cortex (sylvian fissure)
Classically hearing sensitivity in presbyacusis decreases in the highest frequencies.

Cochlear pathology seen in Presbyacusis:

Depending on the pathology seen in the cochlea, 4 different types of Presbyacusis have been identified. They are Sensory Presbyacusis, neural Presbyacusis, strial presbyacusis and cochlear conductive presbyacusis. The aging cochlea present disorders that are symmetric in paired ears; but the extent of involvement at the cellular level may be uneven. Hence presbyacusis can occur in 4 differing pathological types, or in combination thereof. A study of pure tone audiograms and cytocochleograms show:

1. Abrupt high frequency hearing loss (attributed to sensory cell pathology (loss)).

2. Flat threshold hearing loss (seen in cases with strial atrophy)

3. Diminised speech discrimination (due to loss of cochlear neurons)

4. Gradual descending audiometric pattern (due to inner ear conductive disorder)

Patients with presbyacusis uniformly have poor threshold for frequencies in 8 Khz range. In fact the threshold was as low as 60% in most of the patients.

Sensory Presbyacusis: is caused by loss of hair cells at the basal end of the cochlea. This commonly occurs in an aging cochlea. The area of involvement may extend to involve even the speech frequency area of the cochlea. These changes cause a rapid decrease in the threshold for high frequency sounds.
The earliest changes occurring in the cochlea is the loss of stereocilia, which can be identified only on electron microscopy. The second change to occur is distortion or flattening of the organ of corti followed by loss of supporting cells. Finally the organ of corti appears as an undifferentiated mound of tissue on a basement membrane. There is a gradual reduction in the number of outer hair cells in the elderly more so in the basal area of the cochlea. This occurs to a lesser extent at the apex of the cochlea. The apical loss of outer hair cells is seen only in individuals of more than 70 years of age. The loss of inner hair cells is less marked, but follows the same pattern as the outer hair cells.

The wear and tear pigment lipofuscin is known to accumulate in the apical cytoplasm of the hair cells. The lipofuscin is assumed to be a waste product of lysosomal activity.

Neural presbyacusis: Is caused by a loss in the population of cochlear neurons, but the end organs are still functional causing severe loss in speech discrimination. Pure tone thresholds are nearly normal. Gaeth used the term Phonemic regression to describe this phenomenon. Studies have shown that speech discrimination scores are slightly better in the left ear when compared to the right, this has been attributed to the left cerebral dominance becoming manifest due to the degenerative changes affecting the auditory pathway. The loss of cochlear neurons is the most consistent pathologic change seen in these patients. It has been calculated that loss of cochlear neurons occurs at the rate of 2,100 (Schuknecht) neurons every decade. There are roughly about 35,000 cochlear neurons in a normal ear. The loss of cochlear neurons may be genetically determined. The atrophy occurs throughout the cochlea, but is more pronounced in the basal turn of the cochlea.

Strial Presbyacusis : (Also known as metabolic presbyacusis) Atrophy of stria vascularis is commonly seen in this condition. Hearing loss in these patients is insidious in onset occurring during the 3rd - 6th decades of life. It progresses slowly. The clinical feature that identifies this condition from the other types of presbyacusis is the presence of a flat or a slightly descending audiometric curve. These patients respond well to the amplification produced by hearing aids. This type of presbyacusis carries the best prognosis because of this feature. Takahashi demonstrated two types of atrophy in these patients:

Type I: a patchy type more severe in the apical and extreme basal regions of the cochlea.
Type II: A diffuse type often showing normal strial thickness with large intercellular spaces that may not be visible under light microscopy.

All 3 layers of stria are involved in various degrees. The loss of strial tissue may cause changes in the composition of the endolymphatic fluid causing further damage to the cochlear hair cells.

Pure tone audiometry shows a flat curve because the pathology involves the whole of the cochlea. Speech discrimination is preserved. This type of presbyacusis is considered by many to be familial.

Cochlear conductive (Mechanical) presbyacusis: This type of presbyacusis is differentiated from others by a linear descending audiogram. This is postulated to be caused due to stiffening of the basilar membrane of the cochlea. The thickening has been found to be more severe in the basilar turn of the cochlea where the basilar membrane is thin. Speech discrimination is average for the given frequency.

Mixed presbyacusis: Has been recently introduced to describe conditions which features of either two or all of the types of presbyacusis discussed above. These patients have been clubbed under this mixed category to account for their varied manifestations.

Presbyacusis is a diagnosis of exclusion. All the other causes of sensorineural hearing loss must be ruled out before declaring the patient to be suffering from presbyacusis.

References: