Age-Related Hearing Loss

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Age-related hearing loss (ARHL) is the most common neurosensory deficit associated with aging. It presents with a predictable pattern of sensorineural hearing loss, causing problems with communication that have been associated with depression and social isolation. Recent studies have improved our understanding of the etiology of ARHL on a molecular level. While treatment options exist with hearing aids and cochlear implants, prevention by identification and avoidance of key risk factors remains the best strategy for dealing with this disease.

Key words: *presbycusis, age-related hearing loss, deafness, hearing aids, aging*

Introduction

Presbycusis, or age-related hearing loss (ARHL), has been recognized for centuries; however, it is only recently that we have begun to understand the etiology of ARHL. This understanding is fortunate as the numbers of people predicted to experience ARHL in the coming decades is immense. Between the year 2000 and 2050 the number of people aged 65 years and older is predicted to double in the U.S. to more than 86 million.[1] The prevalence of hearing loss in people older than 50 years has been estimated at 50%, and in those over 80 at 90%.[2] This makes hearing loss the most common neurosensory deficit associated with aging, and, while common, it is not benign.[3] A study by Cacciatore *et al.* found that older adults with hearing loss were more likely to have depression, were less likely to participate in meaningful activities, and had lower cognitive scores.[4] The challenges have been to understand why our hearing worsens with age and what we can do to either prevent or ameliorate that degradation.

Types of Hearing Loss

Hearing loss is divided into two broad categories: conductive and sensorineural. Conductive hearing losses are caused by alterations in the structure or function lateral to the cochlea. This may be due to diseases of the pinna, external ear canal, tympanic membrane, middle ear space, or the ossicles. It is easily distinguished from a sensorineural loss on physical examination by using the Weber tuning fork test. In the setting of a pure conductive loss, the tuning fork sounds louder in the affected ear. The earliest theory for the etiology of ARHL was that it was a problem of conduction. In 1870, Toynbee noted thickening of the drum among older patients, which he proposed was responsible for the hearing loss in older age.[5] While conductive losses occur among older adults as they do in every age group, it has been shown that the aging process has no inherent effect on the conductive capacity of the middle ear.[5] Sound transmission is unabated by the changes in the drum and ossicles that occur in later life.

The classic audiometric finding among people with ARHL is a symmetrical sensorineural loss that is worse in the higher frequencies. Sensorineural losses represent a defect in the medial hearing pathways, between the cochlea and the auditory processing cortex. The symmetrical pattern of ARHL is important to recognize as individuals with asymmetrical hearing loss need to be screened aggressively for other etiologies, such as a vestibular schwannoma. Age-related hearing loss is initially frequency specific, with worse hearing in higher frequencies, creating the down-sloping pattern on an audiogram. This down-sloping pattern is similar to the audiometric pattern seen among people with noise-induced hearing loss. Individuals with a history of otoacoustic trauma, such as exposure to fire arms, classically demonstrate a "noise notch," with elevated hearing thresholds in the 3,000–6,000 Hertz range.[6] Age-related and noise-induced hearing loss have traditionally been thought of as distinct entities; however, recent theories of ARHL have stressed the interplay between environmental and genetic causes.[3]

Etiology of Hearing Loss

The cochlea is organized tonotopically: higher frequencies stimulate the basal turn, while low-frequency sounds stimulate the apical cochlea. Pathological studies of the temporal bones of people with ARHL and noise-induced hearing loss have shown similar changes in similar sites of the cochlea, fitting with the similar audiometric findings in these disease processes. Both groups show a loss of hair cells in the basal turn, degradation of the spiral ligament and spiral limbus, and pathology of the stria vascularis.[7] There is also clinical evidence of interplay between these disease processes. Those with acoustic trauma and a noise notch are more likely to develop hearing loss in other frequencies as they age, and at a faster rate.[6] People in jobs with significant noise exposure are more likely to develop hearing loss in older age, even years after the cessation of significant exposure.[8]

Nevertheless, noise exposure alone does not explain ARHL. A cross-sectional study of the population in the town of Beaver Dam, Wisconsin, showed that hearing thresholds dropped predictably

with older age regardless of noise exposure.[8] A longitudinal study in South Carolina followed up older adults for a mean period of 6 years and found that age was an independent risk factor for hearing loss, independent of noise exposure history.[9] A Dutch study of paternal and fraternal twins showed a genetic predisposition for hearing loss in older age independent of noise exposure history.[10]

Other factors that likely affect ARHL include oxidative stress and genetic predilection. Animal studies have shown that reactive oxygen species or free radicals—which are the inevitable product of cellular respiration—degrade proteins, lipids, and deoxyribonucleic acid in the cochlea over the lifetime of an organism.[11] A study of older dogs showed a decrease in the degradation of the stria vascularis and a preservation of neuronal density in animals fed a diet rich in antioxidants compared with those in a control group.[11] Preliminary data from our own institution have showed improved hearing thresholds in older mice fed a diet of antioxidants compared with those in a control group. In addition to the findings from the Dutch twins study cited above, there is strong evidence from mouse studies for a genetic component to ARHL. The first gene demonstrated to cause ARHL-like pathology in mice was ah1. This gene was recently determined to code for cadherin 23, which is expressed in hair cell stereocilia. Defects in the production of cadherin result in the histological hallmarks of all major forms of ARHL.[7] While this mechanism has not yet been confirmed in humans, genetic homology between human deafness and mouse deafness has been established in other mouse strains.[5] Other established risk factors for ARHL besides family history, diet, and noise exposure include smoking, ototoxic medications, and in some studies hypertension and heart disease.[12]

Diagnosis

Individuals with ARHL present with the predictable sequela of progressive high-frequency loss. Often the first problem that the patient or his or her family notices is an increased difficulty understanding speech in noisy environments. Age-related hearing loss is a progressive problem: in the Beaver Dam study, patients lost an average of 1 dB a year.[8] As hearing loss progresses, patients notice impaired localization of sounds. Once the deficit extends to the 2,000–4,000 Hz range, speech understanding in any setting is impaired.[12] Not uncommonly, the deficit is most obvious to those around the patient, while the patient insists that the problem lies with family or friends who mumble or do not speak clearly. In a society that puts a premium on youthfulness, hearing loss is seen as a marker of older age, and patients often minimize their loss. To combat this phenomenon, the U.S. Preventive Services Task Force has recommended routine screening of older adults for hearing impairment.[13]

Key Points

- Age-related hearing loss (ARHL) is common and has been associated with depression and isolation.
- ARHL is typically a sensorineural loss that initially affects the high frequencies.
- Genetic predisposition and sound exposure are probably the most important risk factors.
- Treatment of ARHL involves ear protection, hearing aids, and cochlear implantation.
- Stem cell research may one day allow regrowth of lost hair cells in the inner ear of people with ARHL.

Treatment

The mainstay of therapy for ARHL has been hearing aids. Hearing aids work by amplifying sound to overcome elevated hearing level thresholds. Recent advances in hearing aid technology, such as improved signal compression and directional microphones, have improved patient satisfaction with aids.[14] New digital aids are much better at amplifying the frequencies and even the specific sounds that users want to hear, while filtering out extraneous noise. Problems with signal distortion, feedback, and sound fidelity, which were commonplace with analogue aids, are better addressed with digital sound processing. However, despite these advances, approximately 80% of the people who could benefit from amplification do not seek assistance from hearing aids.[15] In addition to the negative stigma discussed earlier, there are issues of cost, comfort, and clarity. Loss of word recognition, which is common in sensorineural loss, is not addressed by an aid. The aid can amplify soft sounds, but it cannot replace lost hair cells.

For people whose hearing loss progresses to a point at which hearing aids cannot re-establish communication, cochlear implants become an option. These devices stimulate the spiral ganglion cells of the cochlear nerve through an electrode surgically advanced into the cochlea. Current candidacy guidelines include patients whose word discrimination score has dropped below 50% in best hearing conditions. Outcomes for implant users are excellent, with 70% of users able to use the telephone for communication after implantation.[16] The implant is a communication tool, however, and not a cure for deafness.

Conclusion

At this time no therapy can reverse the changes seen in the cochlea with aging; therefore, prevention is critical. Acoustic trauma in youth manifests itself many years later. Hearing loss among people with a history of acoustic trauma progresses at a faster rate, even years after that exposure has ceased.[8] Regulations from the Occupational Safety and Health Administration allow for 8 hours of exposure to work noise of 85 dB, but that drops in half with every 3 dB increase. Ear plugs can provide 15–25 dB of noise attenuation and can therefore make a

dramatic difference for industrial workers over a lifetime.

As mentioned earlier, there is growing enthusiasm for the possibilities of antioxidants to provide protection against ARHL. Researchers at our institution are testing a combination of orally administered antioxidants in an ARHL mouse model in an attempt to reduce the degree of ARHL; the goal is to develop an antioxidant supplement for human use.

Preliminary studies using stem cells to ameliorate the degree of ARHL have also been done. Once stem cells are localized to the cochlea, they could be stimulated with local growth factors to encourage differentiation into either hair cells or lateral wall cells.[17]

Breakthroughs in the treatment of ARHL have the capacity to affect millions of individuals. An intervention that would allow us to connect with the people around us easily and effectively throughout our entire life would be a sea change in human history and worthy of significant research commitment.

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