Presbycusis: A Brain Disorder?
“The Ears Listen, the Brain Hears”

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ABSTRACT

Age-related hearing loss (presbycusis) is the most common cause of hearing loss. The audiometric profile of peripheral presbycusis is well known, but little attention has been paid to the role of central auditory dysfunction in both the diagnosis and management of presbycusis. Central presbycusis is typified by difficulty understanding speech in noise. Central presbycusis appears to involve or arise from decrements in executive functioning, which may be age-related or dementia-related. Tests employing speech in noise are appropriate measures for identifying patients who understand speech relatively normally in quiet but have undue difficulty understanding in background noise. The message of this discussion is that such testing should be done routinely for patients complaining of this problem. Identification of central presbycusis with such testing is important for proper auditory rehabilitation.

KEYWORDS: Hearing loss, dementia, working memory, executive function, central auditory processing

Learning Outcomes: As a result of this activity, (1) the participant will be able to describe the associations between auditory processing and cognitive impairment; (2) participants will be able to implement recommendations for assessing auditory processing in older patients.

Age-related hearing loss—presbycusis—is the most common type of hearing loss. Although the earliest signs have been observed in 20-year-olds, presbycusis generally begins to materially impact hearing in our 60s and gets progressively worse thereafter. Although hearing aids help, dissatisfaction with these devices is common. Emerging evidence points to

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changes in brain function as a major component of presbycusis, commonly labeled as "central presbycusis." This evidence has two aspects: (1) the brain changes in response to loss of information from cochlear dysfunction (age-related or otherwise), and (2) the aging brain is less and less capable of processing speech, especially in difficult situations (e.g., as with competing speech at a social reception). It is presumed but unproven that loss of neurons in the auditory system and association areas as well as decreased processing speed with aging or disease are the major pathological elements in presbycusis. Given the increasing numbers of senior citizens with hearing loss and difficulty hearing in noise—the hallmark of age-related auditory brain dysfunction—greater understanding of this important component of presbycusis is needed. Moreover, the putative links between central presbycusis and Alzheimer’s dementia (AD) raise issues about causation that have only recently been addressed.

PERIPHERAL PRESBYCUSIS
Elevation of pure-tone thresholds in presbycusis begins in the highest frequencies and works its way down the scale. This "top-down" progression mirrors the way the inner ear works and reflects the complex organization of the cochlea. As every student of hearing learns, the ear has 30 different cell types, loss of any one of which may cause hearing loss. The pattern of loss usually gives a clue as to the cells involved, but different pathological processes may have similar audiometric profiles. The most common cell loss in the cochlea is that of the outer hair cells (OHCs), a loss that is generally ascribed to noise damage. For decades, it has been assumed that loss of OHCs is the major pathogenic factor in presbycusis. However, recent evidence suggests that loss of OHC function secondary to strial atrophy is an important element in the development of age-related hearing loss.

The strial vascularis is the “battery” of the inner ear, providing metabolic energy to power OHC function. The fact that more OHC motility is required to hear in the high than the low frequencies offers an explanation for why presbycusis begins in the high frequencies. Also, declining endocochlear potential affects the high more than the low frequencies, which is the classic audiometric signature of presbycusis. Eventually, low-frequency thresholds also decline, producing the pattern of flat hearing loss that is typical of advanced cases.

The cause of declining strial function is unknown, but a familial tendency is more evident in mothers and daughters than in fathers and sons. Gates et al demonstrated an association between low-frequency hearing loss and cardiovascular disease events in women that suggests an underlying vascular mechanism for some forms of presbycusis. The role of diabetes in age-related hearing loss is uncertain, with some studies showing a significant relationship and others not. These studies compared various measures of audition in groups of people with and without diabetes to determine a statistical association. However, none of the studies I was able to review examined the signature of strial dysfunction in their analyses; further, none reported improvement in auditory function with diabetes treatment and glycemic control. Clearly, there is much work to be done to understand strial dysfunction, how to diagnose it, and how to treat it. I believe we have only scratched the surface of this important aspect of presbycusis.

It is suspected that hearing aids, which help to compensate for reduced OHC function, replace, in a metaphorical sense, the “lost” energy provided by the strial vascularis. Such a supposition is unproven but logical. Clinicians know that people with the strial loss pattern of presbycusis do reasonably well with hearing aids as opposed to those who have lost OHCs from other causes, such as loud noise exposure. It can be hypothesized that in strial loss the OHC are present but functioning at a reduced level, which can be compensated by amplification. However, with OHC absence, no amount of amplification will restore lost function. The endocochlear potential maintained by the stria powers OHC motility, which is the source of the cochlear “amplifier.” That this nonlinear cochlear amplifier, which shuts down with high-level inputs, is seemingly substituted for by external amplification remains hypothetical but is worthy of continued investigation.

Owing to our inability to measure human endocochlear potentials, it cannot be stated
with surety how pervasive strial dysfunction might be in human presbycusis. Some animal strains (e.g., Mongolian gerbil) show clear atrophy of the stria with preservation of OHCs. Nonetheless, strial microvascular disease remains as a likely candidate for the pathophysiology of human peripheral presbycusis. As a parenthetical comment, there is continued interest in hair cell regeneration, as occurs spontaneously in birds and fishes, as a potential therapy for human hearing loss. Precise identification of the types and degree of cellular loss will be a necessary step in that research. Unfortunately, we have only scratched the surface of identifying human phenotypes of presbycusis.

HEARING IN NOISE (CENTRAL PRESBYCUSIS)
The number one clinical problem in presbycusis is not understanding speech in quiet, but understanding speech in noise. This deficit increases in frequency and severity as we age. Of course, people with poor cochlear function may have difficulty hearing both in noise and in quiet, depending on the degree and pattern of hearing loss. By contrast, this discussion will focus on people with presbycusis who understand speech in quiet relatively well but who have a disproportionate decrease in understanding speech in the presence of competing sounds. For purposes of this discussion, I refer to this phenomenon as “central presbycusis,” a condition that adversely affects hearing aid acceptance and communication ability. For decades, many investigators have studied central auditory function as if it were a noncognitive (i.e., purely auditory) phenomenon, usually taking care to exclude people with evidence of cognitive dysfunction from these investigations. In contrast, my colleagues and I at the University of Washington published a series of articles that clarify some of the relationships between elements of brain function and presbycusis. In these publications, we make the case that understanding speech is a cognitive process and that loss of the cognitive elements subsumed under the rubric of executive functioning is a likely explanation for our observations. Craik reached a similar conclusion in his report.

Before outlining these findings, it should be remembered that without the ability to process rapidly changing speech signals, spoken words would sound like gibberish. The wonderfully complex auditory system relies on association areas in the brain to store speech elements and, as we listen, the brain compares incoming sounds with those stored in working (i.e., short-term) memory. The brain rapidly judges the sound, its meaning, and its linguistic relationships, allowing us to make “sense” of incoming stimuli. This process becomes more difficult when pathways between the primary auditory cortex and the association areas or these areas themselves become degraded. Because speech processing is so rapid, considerable brain resources are used in separating the speech signals from background noise. Current thinking suggests that even a minor decline in cortical brain function can have a measurable impact on understanding speech in noise.

CENTRAL AUDITORY PROCESSING
Elements of so-called central auditory processing disorders also are included in the term “executive functioning.” Tasks such as working memory, selective and divided attention, concept generation, and the ability to plan, initiate, maintain, switch, or inhibit behavioral responses are categorized as the human brain’s executive function. As we age, loss of nerve cells and neural networks in these critical areas impact on executive functioning. Loss of executive functioning is also a very early finding in AD. Therefore, we wondered if people with early AD or its preclinical equivalent might also have central auditory processing dysfunction. To this end, my colleagues and I conducted a long-term epidemiology research program with observations from three cohorts: the Alzheimer’s Center at Washington University in St. Louis, the Framingham Heart Study in Framingham Massachusetts, and the Adult Change of Thought (ACT) study in Seattle. We obtained and studied a battery of hearing tests in these groups of older people, including tests of central auditory processing function. The results of these tests showed a significant decrease in central auditory function with age (as expected) in each of these cohorts, with even
poorer scores in people with early memory loss, and even more so in those with early AD. We found these associations of central auditory test results and cognitive status to be highly significant, even after adjusting for age and gender. In the ACT cohort, we determined that the majority of pathways between the inner ear and the primary auditory cortex were intact. This finding led us to conclude that patients' auditory processing dysfunction probably involved their brain association areas more than primary auditory cortex or ascending pathways. This conclusion agreed with the archetype of AD as a “top-down” disorder. Although mid-brain plaques and tangles are common in late stage AD, this is not the case in the early stages. We then compared the association of central auditory function and executive functioning. Once again, the correlation was statistically significant. Finally, we examined the data to determine if patients with severe central auditory dysfunction but normal cognitive function might receive an AD diagnosis sooner than those without central auditory dysfunction. We also found this to be the case, as it was in the Framingham Heart Study, in which patients whose performance on the Synthetic Sentence Identification with Ipsilateral Competing Message (SSI-ICM) test was below 50% correct (normal is 80% or more correct) exhibited a ninefold risk of receiving a future diagnosis (3 to 12 years later) of AD.

In these studies, we used a different criterion for central auditory abnormality than that used by the developers of the tests. For our analyses, we set the fail rate at 40% or fewer correct in either ear to minimize false-positive results. We also required our subjects to have normal word recognition scores in quiet of 70% or better to control for audibility.

One might be tempted to conclude that central auditory dysfunction is a precursor to cognitive dysfunction in general and to executive dysfunction in particular. Although the evidence points in that direction, I believe this issue needs further study before such a conclusion is warranted. We simply do not know enough about this complex area to understand all of the cause-and-effect relationships. However, it is clear that evaluation of central auditory function should be part of the routine hearing evaluation of seniors complaining of difficulty hearing in noise because we cannot treat a condition that is not diagnosed. Strouse et al reached a similar conclusion and recommended central auditory screening in people with AD to enhance psychological assessment and rehabilitation of peripheral hearing loss.

**CLINICAL IMPLICATIONS**

Problems associated with implementing the recommendation for central auditory assessment include which test to use, testing time, test materials, and reimbursement. The SSI-ICM takes 20 minutes or less in most people, which includes instruction and a training trial at +10-dB signal-to-noise ratio. We recommend that the test is conducted using the standard 10 presentations at 0-dB signal-to-noise ratio because we discovered that using slower presentations and up to 30 presentations adversely affects the predictive power of the test in terms of executive function. The Dichotic Sentence Identification in free report mode (in which the patient points to two of the six sentences that were spoken) also can be used to assess central presbycusis and to predict cognitive status. Given that assessment of central auditory function has immediate relevance to rehabilitation methodology, these tests should be performed routinely for seniors complaining of difficulty hearing speech in noise. Evaluation of central auditory function is a billable procedure (Current Procedural Terminology Code 92620) that establishes the extent to which auditory perception versus central processing is the problem. Such baseline evaluations can be repeated later for those with increasing difficulty.

Some people with both peripheral and central auditory dysfunction prefer to use one hearing aid (in the better ear) rather than the traditional binaural fitting. Thus, identifying central auditory dysfunction has immediate practical value. Other strategies to improve communication for people with central presbycusis include face-to-face communication, reduction of background noise, use of assistive listening devices, and efficient communication style (avoid speaking too quickly, pronounce words clearly, repeat the message, and ask for confirmation of what was heard). A little effort
on both the speaker’s and listener’s parts can help to maximize the use of residual hearing. Auditory training programs may be useful for some patients, but these are not standardized nor widely available. Nonetheless, interest in this possibility is growing. A recent Internet search for “auditory training” yielded five commercially available software packages. It is beyond the purview of this article to evaluate theses programs, but, caveat emptor aside, this concept seems like it could be a fruitful addition to standard auditory rehabilitative offerings. Certainly, this is a ripe area for clinical research.

AD is increasing in prevalence as our society ages.24 There is no treatment available now that can reverse the pathology; current medications are aimed at slowing the progression of decline. Logic would suggest that early detection of AD is key to reducing the burden upon those afflicted, their families, and society. The earliest changes in brain function often precede the clinical diagnosis of AD by many years. Thus, it seems reasonable to investigate methods for early detection of this disease. Given that the onset of AD can be insidious and variable, utilization of auditory testing for early detection of cognitive decline may be warranted. Certainly, research into this possibility is justified. The studies cited in this article should help to persuade AD investigators to add central auditory testing to their research protocols.

CONCLUSION

Central presbycusis is defined by disproportionately greater difficulty understanding speech in noise than in quiet and is a common factor in hearing aid rejection. Tests of central auditory function that involve speech, such as the Dichotic Sentence Identification or the SSI-ICM, should be performed routinely in senior patients who complain of difficulty hearing in noise; those with poor performance should be considered for customized rehabilitation measures. Research in central presbycusis should include assessment of auditory training methods. Use of central auditory tests as methods to screen for preclinical dementia are not warranted yet, but future research may open avenues for greater understanding of the complex role of cognitive elements in understanding speech.

REFERENCES