CHAPTER FOUR

Central Auditory Processing in Presbycusis: an Epidemiologic Perspective

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Background

Presbycusis (literally elder hearing) or age-related hearing loss is the inevitable deterioration in hearing that occurs as people get older. While loss of the highest frequencies can be detected in young adulthood, it is not until the 6th decade and beyond that clinically significant hearing loss is evident. Presbycusis is a multifactorial process that affects people in their senior years in degrees ranging from mild to profound. It is conceptually useful to consider presbycusis as a mixture of acquired auditory stresses, trauma, and otologic diseases that affect hearing over time superimposed upon an intrinsic, genetically controlled, aging process.[1] Separating aging effects from age effects and aging changes from age-related diseases complicates the study of presbycusis.

Presbycusis has both peripheral and central components. Although the latter is the primary focus of this review, peripheral conditions that may confound central auditory processing are included. The review is based primarily on my epidemiologic investigations of the Framingham Heart Study cohort and the Adult Change of Thought Cohort (Seattle). These investigations provide a more controlled and representative overview of auditory aging than is seen in clinic populations. Unlike clinical studies, however, trends in groups can only be described with a broad brush. The clinical picture thus obtained focuses on central tendencies, which provide valuable insight (but not proof) into possible processes involved. By addressing the effects of brain dysfunction (dementia, e.g.) on central auditory processing in these people, a fuller picture of the central presbycusis may be obtained.

The high prevalence of presbycusis, which is a consequence of our aging population, leads to hearing difficulty as a common social and health problem. Overall, 10% of the population has a hearing loss great enough to impair communication, and this rate increases to 40% in the population over 65 years.[2] Eighty percent of hearing loss cases occur in the elderly.[3] Although hearing worsens increasingly with age, the magnitude of the hearing problem at any given age varies greatly. It is rare to find a person over 70 years of age who has no hearing impairment or whose hearing sensitivity has not declined from youthful levels.

Cardiovascular disease as well as cardiovascular disease risk-factors affect hearing to some extent. Stroke, myocardial infarction, claudication, hypertension, hyperlipidemia, and diabetes mellitus have all been associated with excessive hearing loss. Therefore, it is logical that maintenance of good general health and fitness would minimize the risk of hearing loss due to systemic disease. High lipid diets are associated with poorer hearing.[4] There is inconclusive evidence that low-caloric diets, which clearly prolong life in laboratory animals, have an effect on presbycusis.[5] While free radical accumulation is presumed to be involved in presbycusis, research into anti-oxidant agents is have not been shown to counter auditory aging.

Dividing the auditory system into peripheral and central components is useful for didactic purposes but one must bear in mind the integrated nature of system and the overlapping attributes of peripheral and central auditory function. For example, deficits in speech perception may occur at any level of the system. Therefore, peripheral changes must be taken into account when central problems are being considered.

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Peripheral (Cochlear) Presbycusis

There are three classic peripheral histologic archetypes seen in human temporal bones: sensory, strial, and neural, occurring singly or in combination, but most cases show mixed histopathologic changes and 25% show no light-microscopic abnormalities.[6] A fourth category, cochlear conductive presbycusis, which was postulated as a gradually descending hearing loss in the absence of pathology under light microscopy, remains controversial and may well reflect the limitations of light microscopy. It is my view that this category reflects early strial degeneration because the audiometric pattern is typical of experimental strial loss.

The decrease in hearing sensitivity in presbycusis begins in the highest frequencies. This has an adverse effect on understanding speech in noisy or reverberant places. Once the loss progresses to the 2–4 kHz range, which is important in understanding the voiceless consonants (*t, p, k, f, s,* and *ch*), speech understanding in any situation is affected. The most common complaint in presbycusis is not that "I can't hear"; rather, it is "I can't understand". For example, people will confuse "mash", "math", "map" and "mat", or "Sunday" with "someday". Even such minor misperceptions, left uncorrected, may lead to communication gaffes or worse. The hearing worsens into old age with the loss extending into the lower frequencies. From the 70's onwards, however, central auditory processing worsens at a faster rate than peripheral processing[7].

The primary peripheral effect of aging *per se* is a progressive decline in cochlear metabolic function (strial presbycusis)[8], which is characterized by a moderately high sloping audiogram and relatively good word recognition ability. Examination of the inner ears of animals raised in quiet shows that degeneration of the stria vascularis is the dominant element.[9–11] The degeneration usually originates in both the base and apex of the cochlea, extending to mid-cochlear regions as age increases. The prevalence of strial/metabolic presbycusis may prove to be substantially higher in humans when the appropriate immunohistochemical techniques (e.g. Na, KATPase) are applied to the study of temporal bones of older humans.

Strial degeneration has a profound influence on the basic physiology of the cochlea, especially on the endocochlear potential (EP), which provides a voltage to the cochlear amplifier. When the EP is reduced significantly, the operation of the cochlear amplifier is affected. Indeed, when the EP decreases to values of 20 mV or lower the cochlear amplifier is considered to be "voltage starved" with a maximum reduction in its gain. The gain ranges from 20 dB in the apex of the cochlea and increases to as much as 60 dB in the base. As much as 20–30% of the stria may degenerate with only a 20 mV reduction in the EP. As strial degeneration exceeds 50%, EP values drop substantially.

The audiometric pattern resulting from strial degeneration in animals is also the most typical finding in cohort studies of the elderly.[12–16] In contrast, the steeply sloping audiogram of people with confirmed noise-induced hearing loss differs perceptably from the strial pattern and coincides with Schuknecht's sensory presbycusis pattern[17], which can occur at any age. Indeed, many of his cases had clear histories of noise exposure. In the absence of any clinical method to measure the endocochlear potential in humans, audiometric patterns are the only way to infer the probable pathophysiology. Available clinical tests are unable to determine whether hearing loss is due to inner hair cell dysfunction, synaptic abnormalities, spiral ganglion atrophy, or auditory neuropathy. Recent quantitative morphometric studies of the cochlea shows loss of strial volume, loss of outer and inner hair cells and ganglion cells in people with classic down-sloping threshold loss.[18]

Central Auditory Processing Dysfunction

The classic clinical finding in what is called "nerve deafness" in the lay literature is difficulty understanding speech. While loss of function at any level in the auditory system may contribute to poor speech understanding, for the purposes of this report, reduced speech understanding in quiet is considered mainly an audibility (peripheral) issue whereas difficulty understanding speech in noise but not in quiet is a sign of central auditory processing dysfunction. For example, large numbers of missing hair cells from noise overexposure affect audibility. The missing sensory cells limit the benefits of hearing aids because optimal speech comprehension depends on adequate frequency tuning within the cochlea. Similarly, loss of spiral ganglion cells typically produces more loss of word recognition ability in quiet than would be expected from the pure-tone thresholds. Central projections to the primary auditory cortex appear to be maintained with age[7] but loss of association area function appears to be a common, age-related phe-

nomenon. Those persons with central presbycusis may have normal or near normal word recognition in quiet but have considerable difficulty understanding speech in noise.

We now know that the pure-tone threshold audiogram is a poor indicator of neural loss, since any pattern from normal to anacousis may be seen. The sine qua non of neural lesions is reduced word recognition in quiet. Better clinical assessment techniques, such as auditory brainstem response audiometry and otoacoustic emission testing, may provide additional detail about the neural function. Primary dysfunction of the auditory nerve (auditory neuropathy)[19] is more prevalent in children than seniors. In this condition, outer hair cell function, as measured by otoacoustic emissions, is normal but the auditory brainstem responses are reduced or absent. The term, auditory dyssynchrony, is now used to describe the condition. It remains difficult to separate malfunctions of the auditory nerve, caused by a reduced EP, from malfunctions caused by degeneration of spiral ganglion cells[20].

The remainder of this review focuses on the causes of reduced speech identification in noise. Other methods, such as compressed or degraded speech, are outside the scope of this report.

Central auditory dysfunction (central presbycusis, i.e. age-related auditory processing dysfunction) is a common element of presbycusis that is typified by difficulty understanding speech in noise or other difficult listening situations. Central auditory processing problems are usually superimposed on peripheral cochlear dysfunction and in late cases central problems dominate treatment challenges. Because speech understanding may be affected at both the peripheral and central levels, an adequate assessment protocol should evaluate both.

Given the complex neural connections through which auditory stimuli pass to be perceived in the auditory cortex, it is logical to ask whether lesions of these pathways would affect hearing. Indeed, secondary degeneration of central pathways after loss of sensory cells in the cochlea, although slow, is a limiting factor in cochlear implantation results in people with long-standing deafness. Primary neural lesions of the auditory pathways are uncommon, however. We have demonstrated with electrophysiologic measures that near-normal responses from the VIIIth nerve, midbrain, and primary auditory cortex are the norm even in people with significant impairment of the central auditory processing system or cognitive deficit.[7]

Central Auditory Testing

Because the ultimate perception of speech is in the brain, it is not unexpected that age-related brain dysfunction could affect hearing. Central presbycusis (i.e. age-related processing dysfunction) limits rehabilitation, increasingly so with advanced age, yet few centers include central testing in the routine evaluation of people for amplification candidacy. A large discrepancy between the history and the results of standard peripheral auditory tests may prompt an evaluation of central auditory function. For example, when a patient has reasonable speech understanding in quiet but has severe speech problems in noise or difficult listening environments, central presbycusis is likely to be present. The most frequently used central tests evaluate speech recognition in relation to noise or other speech sounds.

A widely available and standardized central auditory test is the Synthetic Sentence Identification with either an Ipsilateral Competing Message (SSI-ICM) or a Contralateral Competing Message (SSI-CCM).[21] The patient listens to an interesting narrative and is instructed to identify which one of 10 sentences superimposed on the narrative by the same speaker. Each sentence has no meaning even though each series of 3 words makes syntactical sense. The test is quite easy for most people even when the signal and the message are presented at the same intensity level, usually 40 dB above threshold. Since audibility is not an issue, the test requires the listener to attend to the message and ignore the narrative. The SSI-ICM appears to be more sensitive to dementia than the contralateral form[22]. The Dichotic Sentence Identification test[23] uses the same sentences with one being presented to each ear simultaneously. Performance is generally better when the subject is asked to focus on one ear at a time (directed mode) as opposed to the free format where what was heared in either ear is reported.

My colleagues and I have shown that poor performance on the SSI-ICM is common in people with Alzheimer's disease (AD) as contrasted to age-matched controls[24]. In this study, 40 non-demented controls, 22 people with a Clinical Dementia Rating (CDR) score of 0.5 (equivalent to early dementia), and 20 persons with a CDR score of 1 (mild, definitive dementia) were tested. The participants were members of the Washington University Alzheimer's Disease Center cohort. Peripheral hearing, word recognition and ABR test results did not vary by cognitive group. Ninety-three percent of the CDR1 cases had a 20 point difference in the SSI-ICM and word-recognition score in quiet, as did 50% of the

CDR 0.5 cases. Very poor performance (< 50% correct) in both ears occurred in 8%, 27% and 66% of the CDR groups 0, 0.5, and 1 respectively (X^2 = 20.2, $p < 0.0001$). Neither hearing aid use, educational level, nor age affected these results.

In another population – older people from the Framingham Heart Study with normal cognitive screening test results on the Mini Mental State Examination – we identified a subset of people where very poor performance on the SSI-ICM (< 50% correct) in either ear and normal word recognition predicted the subsequent diagnosis of AD by several years[25]. The odds ratio for dementia onset was over 12 for those with grossly abnormal SSI-ICM and normal word recognition in either ear. This finding suggested a common mechanism for AD and central auditory dysfunction. We reasoned that because executive functioning is abnormal in people with AD and that many of the elements involved in central auditory processing, such as short term memory, attention to task, and inhibition of unwanted signals, might involve executive functioning (or at least be affected by it) and undertook, therefore, an examination of executive functioning and central auditory processing in another cohort.

Comprehensive audiometry, including three measures of central auditory function (the SSI-ICM, DSI in free report mode and the Dichotic Digits test (DDT)), and standard neuropsychiatric tests (Trails A and B, the Stroop color test, and clock drawing) were performed in group of 313 older volunteers from a dementia surveillance study (The Adult Change of Thought (ACT) group in Seattle)[26]. The participants were selected on the basis of symmetric pure-tone thresholds (i.e. + 25 dB) and a word recognition score in quiet of 70% or better at most comfortable loudness. Participants were grouped on the basis of 1) normal screening cognitive tests (N = 233), 2) mild memory impairment without other signs of dementia (N = 60), or 3) an established diagnosis of AD and a Clinical Dementia Rating of 0.5 or 1 (i.e. early stage AD). There was a significant relation of central auditory test results and the neuropsychiatric tests even adjusted for age, education, and pure-tone hearing thresholds that persisted after excluding the AD cases (Gates, et al. in press). Of interest, the pure-tone thresholds were associated with cognitive test results showing poorer thresholds in those with poorer executive function scores. This suggests a cognitive component for threshold testing (i.e. "did I hear the tone or not"). The prevalence of a poor central auditory test (i.e. < 60% on the SSI-ICM, DSI, or DDT in the poorer ear) was 33% for the cognitively normal group, 80% for the memory impaired group, and 90% for the AD group.

These studies prompt speculation about the relations of central auditory testing, executive functioning, and dementia. First, it is clear that in people with AD there is a notable loss of the ability to extract speech out of a background of competing speech and that loss is greatest for those persons with the poorest dementia status. Second, because central auditory tests may be abnormal years before the dementia becomes clinically evident, it may be the case that the performance burden of central auditory tests upon diminishing executive functioning and cortical association structures is greater than the burden upon usual and customary tasks of daily living, such that one might theorize that abnormal central auditory tests in the elderly are a sign of preclinical dementia. Third, the strong associations between executive function and central auditory function suggest a common mechanism. Whether central auditory testing abnormalities are indicators of decline of executive function, or whether a third factor is responsible for the association, can not be determined on the basis of these data. Nonetheless, the high prevalence of both central auditory dysfunction and mild cognitive deficits in the elderly argues for continued research into the mechanism(s) involved. It appears prudent to me to include central auditory testing in the evaluation of older people with hearing problems and consider referral of those cases with substantial loss of auditory processing ability for neuropsychiatric evaluation. With the possibility of treatments to forestall the progression of Alzheimer's disease on the horizon, early identification assumes great importance, and these data suggest that auditory processing deficits may be a risk factor for the subsequent onset of clinical dementia.

Similarly, auditory processing testing should be included in the evaluation of older people with hearing difficulty in order to tailor their rehabilitation program. For example, degraded speech, such as occurs in noise, reverberant halls, or with rapid speakers, is more difficult to understand. Therefore, optimizing the listening environment, often by simple means (e.g. turning off the television, speaking slower), has positive effects on speech comprehension. Enhancing the signal-to-noise ratio is an important consideration. Patients with central processing disorders generally do better with a single aid in the better ear.[27;28]

Aural communication is enhanced by viewing the speaker's face. Given that speech is often redundant, facial expressions and lip contours provide assistance in

filling the gaps resulting from unheard speech sounds. Formal speech reading classes are available at a few centers. Unfortunately, not enough centers are available to provide adequate speech reading training. Materials are now becoming available on the World Wide Web (search engine phrase "speech reading").

For people with severe losses, auditory training is likely to be of benefit. The hearing impaired listener is trained to identify speech sounds and key words with amplification in place. Such training is has seldom available outside of centers, but now, the widespread availability of personal computers and internet access has facilitated the development of automated programs for training. The Listening and Communication Enhancement (L.A.C.E.) is one of the available programs. See http://www.neurotone.com. While training has the promise of improved speech comprehension for people with central presbycusis, the use of training in this condition is in its early stages.

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