# The Cognitive and Behavioral Consequences of Hearing Loss, Part 1

Evidence suggests a causal link between hearing decline and cognitive impairment.

#### By Ronald Devere, MD

ecently, a good friend and medical colleague of mine who is 70 years old, went to see his ear, nose, and throat (ENT) specialist due to a decline in his hearing. He noted he could no longer have conversations at meetings or restaurants with loud ambient noise. The ENT physician noted a decline in hearing on testing and recommended a hearing aid. He also told my friend that untreated hearing loss can increase one's risk of developing dementia. My first thought was that poor hearing and loss of verbal communication due to hearing loss could be interpreted by others as an obvious cause of memory and cognitive impairment. Having no awareness of any literature on the topic, I spoke to the ENT physician, and he told me that scientific studies have shown that hearing loss can impair auditory central processing and can cause dementia. This article digs deeper into some of these studies and explores how hearing loss influences cognition.

### The Connection Between Hearing Loss and Cognitive Decline

In general, aging (65 and older) and other risk factors often leads to decreased hearing and very mild cognitive decline. Significant hearing loss (>20dB), elevation of the threshold for pure tone detection affects 40% of those over the age of 65 and 80% over the age of 85 and has links to cognitive impairment and dementia.<sup>1</sup> Hearing loss greater than 25dB has an effect on cognitive deterioration equivalent to seven years of aging.<sup>2</sup> Age-related hearing loss (presbycusis) commonly results from cochlea dysfunction. Agerelated alteration in more central auditory pathways has been under-recognized.<sup>3</sup> The good news about the association between hearing loss and cognition is that hearing loss is considered treatable. If hearing loss is treatable by hearing aids, cochlear implants, etc., are we missing a very simple therapy that can delay or improve cognitive decline, which, "It appears that if hearing loss is discovered as early as possible and treated, it could possibly improve or delay cognitive decline."

until now, with some exceptions, has not been very successful other than slowing it down? More importantly, however, is that only a small proportion of older adults who would potentially benefit from hearing loss treatment seek help.<sup>4</sup> The effects of hearing loss on auditory processing, speech communication, and psychosocial well-being have been well studied for many years,<sup>5,6</sup> but the negative impact of hearing loss and cognition has only been studied in more detail in the last three years.<sup>7</sup>

Hearing has two major domains:

- 1. **Peripheral hearing**, which is the transmission of sound through the auditory periphery and includes the outer ear, middle ear, and sound encoding by the cochlea
- 2. Central auditory processing (CAP), which includes the brainstem, midbrain, and auditory cortex.

Auditory cognition processing beyond sound detection leading to auditory perception and understanding is mediated by networks involving the auditory cortex and its cerebral connections. Auditory objects that are present in our daily life include speech, voices, music, and environmental noises that must be separated from the auditory background and organized with coherent representation. The processing of these sound objects includes encoding of acoustic features such as pitch, rhythm, and timbre, leading to the extraction of the meaning of this information to sound recognition. The CAP network includes the temporal, parietal, and frontal networks, as well as the subcortical circuitry, which also plays a role in cognitive function. Central auditory processing disorders affect 23% to 76% of older adults.<sup>8,9</sup> Decreased central auditory processing has been associated with poor cognitive performance and decline in small study samples. Mild cognitive impairment (MCI) and Alzheimer's disease (AD) have been associated with decreased performance on tests of central auditory performance such as speech in noise.<sup>10,11</sup> One study examining peripheral hearing, CAP, and multiple cognitive skills in 120 adults with a mean age of 70 found that cognitive and CAP abilities were significantly related to speech understanding in noise but not peripheral hearing loss.<sup>11</sup> Another study examining pure tone thresholds (250-8000Hz), optoacoustic emissions, performance of completing sentence tasks, and scores on clinical dementia rating scale showed significant relation between CAP and cognitive function but not between peripheral hearing loss and cognitive function.<sup>12</sup>

Other studies with large populations have suggested that peripheral hearing loss is independently associated with poor cognitive function,<sup>13</sup> and when using measures of memory, executive function, and speed of processing.<sup>2,14,15</sup> One study showed that cognitive decline has been associated with faster rates of peripheral hearing loss.<sup>16</sup> The main criticisms of these studies is that they are cross-sectional, and the cause-and-effect direction is not clear. Moreover, other studies have found no relationship between peripheral hearing loss and cognitive impairment mostly because insensitive measures such as only using the Mini-Mental State Examination for cognitive testing and using limited hearing tests were the likely factors.<sup>17</sup>

In a very detailed study on hearing loss and dementia, researchers evaluated 4,545 individuals with a mean age of 75 and no known dementia.<sup>13</sup> Of note, 836 patients had baseline hearing loss, determined by history, examination, and family input. The remainder of the individuals did not have hearing deficits. The researchers then followed these individuals for more than 12 years. Of those with hearing loss, 16% developed dementia and 12% developed dementia without hearing loss. The mean time to reach dementia was 10.3 years for the hearing loss group, as compared to 11.9 years for those without hearing loss. Additionally, 10% of patients with dementia who had hearing loss. Sex, presence of ApoE 4, education, and baseline age were controlled.<sup>13</sup> The researchers concluded that older people

with baseline hearing loss develop dementia at a faster and higher rate than those without hearing impairment. They also mentioned that subjective hearing assessments have been reliably valid within 3% when compared to standard audiometry.<sup>18,19</sup>

Findings from another study showed that individuals with hearing loss developed between 30% and 40% acceleration of cognitive decline and 24% increased risk for incident cognitive impairment during a six-year period compared to those with normal hearing. They also found that individuals with hearing loss would require 7.7 years to decline five points on the cognitive scale versus 11 years in individuals with normal hearing.<sup>17</sup> The risk of all-cause dementia increases with severity of baseline hearing loss,<sup>20</sup> and decreased hearing is independently associated with lower scores on tests of memory and executive dysfunction.<sup>21,22</sup> Moreover, an Australian study on aging suggested that cognitive impairment can hasten the degree of sensorineural hearing loss.<sup>16</sup>

#### Brain Changes Observed With Age-Associated Hearing Loss

There have been a number of tests used to measure anatomical brain changes observed with age-associated hearing loss, including MRI voxel-based morphmetry, which measures thickening, concentration, and volume of the gray matter, and diffusion tensor imaging, which estimates the integrity of white matter by diffusion of water molecules. The unit used to measure white matter tract integrity is called *fractional anisotropy* and can range in value from 0 to 1, with higher measurements corresponding to healthier myelin and fiber density.

Studies have shown that poor hearing is associated with reduced gray matter volume in the auditory cortex bilaterally when age-related structural brain changes were controlled.<sup>23,24</sup> Other brain areas such as the prefrontal cortex gray matter volume and thickness were reduced in patients with poor ability to perceive speech in noisy environments when age effects were taken into an account.<sup>25</sup> In one 2011 study,<sup>26</sup> patients with mild to moderate hearing loss (<70dB HL) showed reduced gray matter volume in the right anterior cingulate, bilateral medial frontal gyrus, and superficial temporal vertex when compared to age-matched controls. These studies, however, were not longitudinal; therefore, it wasn't clear whether these observations on voxel-based morphometry occurred before or after the hearing loss. A similar study over a six-year period looking at age, hearing loss, and brain atrophy found an accelerated process of decline in whole brain volume in those with decreased hearing.27

Hearing loss has also been associated with decreased white matter integrity leading into and out of the auditory cortex (inferior colliculus to primary auditory cortex).<sup>28</sup> This indicates that changes in the brain noted with hearing loss cannot be due to age alone, suggesting that loss of hearing leads to sensory deprivation (auditory), information degradation, and changes in brain anatomy. The big question is whether treatment of hearing loss changes these anatomical abnormalities discovered in the brain.

Tests used to study functional brain changes are functional MRI (FMRI), which measures brain activity by detecting brain blood flow, and event-related potentials (ERPs), which measure electrical activity of the brain that correlates with a cognitive process.

Hussain et al examined FMRI on the effects of hearing loss on emotional processing using age-matched controls with normal hearing.<sup>26</sup> Participants were charged with rating affective stimuli from a digital sound database as pleasant, unpleasant, or neutral. The normal hearing group employed the expected limbic and auditory regions compared to the hearing loss participants, who had an altered dorsal attention network, suggesting evidence supporting functional alteration beyond the auditory cortex. Peele et al used FMRI and found that differences in hearing ability predicted the degree of neural recruitment in bilateral superior temporal gyri, thalamus, and brainstem during sentence comprehension.<sup>23</sup> In particular, those with poor hearing showed less language-driven brain activity, even when age was controlled.

A number of studies have also evaluated ERP function. In general, ERP is less subjective and measures neural transmission time (latency) and strength of response (amplitude) in various stimuli including auditory and cognitive stimulation. In one study, researchers examined auditory evoked potentials in individuals with mild to moderate high-frequency hearing loss and normal controls.<sup>29</sup> The auditory stimuli involved the speech stimuli. Those with hearing loss showed increase in latency and amplitude of the P2 auditory evoked potentials relative to controls. Cortical source localization revealed decreased activation in the temporal cortex and increased activation in the frontal cortical areas in individuals with hearing loss compared to controls, suggesting potential changes in allocation of cortical resources.<sup>29</sup>

In a study of visual evoked potentials in middle-aged adults with normal and impaired hearing, the amplitudes of the visual evoked potentials (P1, N1, and P2) were significantly larger in those with hearing loss.<sup>30</sup> Additionally, the latency was decreased in individuals with problems in the N1 complex compared to controls and correlated with speech perception performance in noise. Cortical source localization revealed increased activation of auditory processing temporal areas with visual stimulation in adults with hearing impairment, suggesting visual cross-modal reorganization.<sup>30</sup> This finding suggests that visual stimulation "Older adults often pretend they understand verbal instructions and comments rather than request clarification. This may reduce performance on cognitive testing of any kind and masquerade as cognitive impairment."

is trying to compensate with central processes in the brain with impaired hearing input. Of note, recent studies have shown that among older adults with high-frequency hearing loss, the P1, N1, and P2 complex is longer in latency and lower in amplitude in those individuals with MCI compared to no MCI,<sup>31</sup> and the hemisphere activation pattern is more diffuse in patients with MCI. This suggests that the auditory signal is degraded in individuals with MCI as compared to those without. Golub et al found that ERPs may be useful in emerging MCI, and the N1 latency was longer in individuals with gene mutations consistent with familial AD than in individuals without the gene mutation in the absence of any behavioral or cognitive concerns.<sup>32</sup>

#### **Possible Mechanisms**

Importantly, many of the structural and/or physiologic brain changes associated with hearing loss cannot be explained by aging only. Simply put, it has not been proven that cognitive impairment (including dementia) is caused by hearing loss or that cognitive impairment causes hearing decline. A 2015 report thoroughly examined this topic and explored possible underlying mechanisms of hearing loss and cognitive impairment.<sup>33</sup> The study divided mechanisms into five categories:

1. Overdiagnosis. As previously stated, the majority of older adults do not seek a diagnosis and treatment for hearing loss, and many do not routinely use hearing aids when prescribed. Older adults often pretend they understand verbal instructions and comments rather than request clarification. This may reduce performance on cognitive testing of any kind and masquerade as cognitive impairment. In one study examining the effects of simulated hearing loss on the MMSE score in 125 adults of various ages, researchers used five audibility levels of hearing; normal, mild to moderately severe, mild to severe, moderate to severe and severe to profound hearing loss.34 MMSE performance was significantly affected by audibility. At the highest audible level used (in patients with mild to moderate severe hearing loss), 16% of the young

adults were misdiagnosed as having dementia. The rate of misdiagnosis increased with lower levels of audibility. The authors recommended that if hearing impairment is suggested by history, especially from caregivers, family members, friends, or the patient, a hearing assessment should be performed prior to cognitive assessment, to reduce the chance of a misdiagnosis of cognitive impairment.

- 2. Widespread neural degeneration. Some studies have found that age, speed, vision, and hearing loss comprise a common factor in age-related changes in cognition and suggest that these age-related changes have a central neural component in addition to any peripheral or end-organ source.35,36 In neurodegenerative cognitive disorders, matching of incoming sound information that has been stored in a neural "template" based on past experience of the auditory world may be an important general operating principle of the auditory brain. Disruption of this process in patients with neurodegenerative disorders may result in impaired perception of sounds and lead to abnormal behavior to sounds. After all, deficient sound perception not attributed to peripheral or subcortical hearing loss is called auditory agnosia or word deafness, a cortical disorder. Excessive auditory processing can lead to hallucinations, another nonperipheral disorder. Genetic disorders can also cause hearing disorders and other neurological symptoms. For instance, one study found a gene mutation of DNA methyltransferase 1 that caused hearing loss and sensory neuropathy in young patients (mid-teens to 30 years old) and cognitive decline later in life (30-50 years of age), suggesting widespread failing of neural function first evidenced by hearing loss and only later by cognitive decline.37
- 3. Sensory degradation/deprivation. Hearing loss results in degradation and loss of input to the brain. Peripheral hearing loss impacts the audibility and clarity of sound, meaning that information reaching the cortex is degraded and deprived of quality sensory information.<sup>38</sup> This leads in time to changes in structure and function of the central auditory and cognitive systems. As described previously, one study found neuroimaging evidence of brain atrophy and volume decline in the right temporal lobe in individuals with hearing loss compared to individuals with normal hearing across six years of evaluation.<sup>2</sup> The real proof of this concept clinically is whether treating hearing loss with amplification improves cognitive impairment. Studies have shown

that hearing aid use improves signal detection and speech comprehension, but no pertinent studies have looked at long-term cognitive changes.<sup>39,40</sup> Palmer et al assessed behavioral changes in those with MCI or Dementia after short-term use of sound amplification with hearing aids.<sup>41</sup> They found that this strategy improved compliance of hearing aid use and reduced behavioral problems after two months of use. They also noted better results in those individuals who were diagnosed early in their hearing loss.

- 4. Cognitive resource allocation and depletion. When incoming auditory signals are degraded due to hearing loss or external noise, fewer resources are available for higher-level cognitive processing (e.g., working memory, long-term recall).<sup>23</sup> Decreased gray matter density has been shown in the primary auditory areas, as previously mentioned, suggesting a reorganization of the brain processing systems when auditory stimulation is decreased. In a study examining the cognitive spare capacity of 24 older individuals (mean age, 69) with hearing loss, researchers found no differences in the cognitive spare capacity test between groups when the listening conditions were optimal.<sup>42</sup> When these tests were given in background noise, older individuals performed much worse. This finding suggests but does not prove that cognitive function may be decreased in older persons due to reallocation of valuable resources, in an effort to process degraded auditory signals.
- 5. Social isolation and depression. As we know, hearing loss places increased demands on cognitive processes and resources. Social gatherings are usually held in places with a high level of background noise, and this environment increases the effort necessary to communicate for any listener. Many individuals with decreased hearing withdraw from these situations and develop social isolation. Older adults with hearing loss are more likely to experience depression, more feelings of loneliness, and enter a much smaller social network. We are also aware that social isolation is a risk factor for cognitive decline.<sup>43,44</sup>

#### **To Be Continued**

The link between cognitive decline and hearing loss is no doubt complicated, with much more to be discovered. Based on what we know now, however, it appears that if hearing loss is discovered as early as possible and treated, it could possibly improve or delay cognitive decline.

Part 2 of this article series (to appear in the November/ December 2017 edition of *Practical Neurology*®), will review evaluations of patients with cognitive symptoms with or without hearing complaints and will also explore options for management.

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