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# We are Not Progressing in Diagnosis of Progressive Hearing Loss

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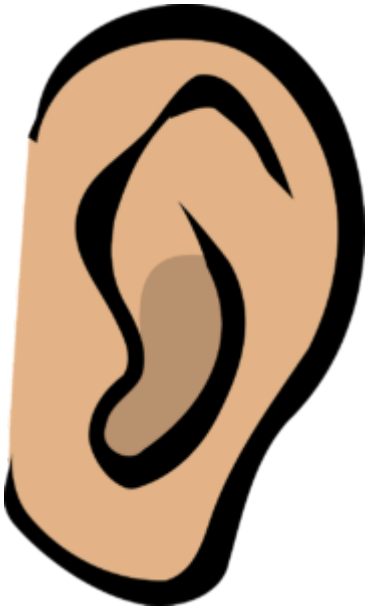

The most prevalent type of hearing loss is a progressive hearing loss. I can make that statement confidently because presbyacusis is progressive, and everybody will have age-related hearing loss, unless they pass away before age 30. However, it is difficult to find good evidence or reliable statistics if you ask the question about the prevalence of progressive hearing loss from other causes.

I have carried out basic science studies on hearing loss initiated by a lesion to one cochlear area or structure, such as the stria vascularis or outer haircells. That damage can progress to other haircells, and, in turn, inner haircells can degenerate. When this occurs, spiral ganglion cell degeneration can result.[1-4] I have used the term “cascading effect” to refer to this sequential degeneration. The steps of this cascading can be explained by known pathological mechanisms. For example, strial damage causes the endocochlear potential loss, affecting haircell metabolism. Haircells that degenerate release toxic chemicals including cytokines, necrosis factors, and reactive oxygen species that can further damage adjacent cells. Inner haircell damage causes excess glutamate transmitter release that results in excitotoxicity and injury to spiral ganglion cells.

Basic science studies inform us that an acquired hearing loss caused by an initial cochlear insult such as viral infection, noise exposure, drug ototoxicity, or cochlear ischemia/hypoxia, can theoretically progress over time. However, searching the published medical and scientific literature, there is scant clinical evidence about how widespread progressive hearing is for most

etiologies. We do have some very particular and often rare genetic causes of progressive loss (e.g., one symptom of Usher syndrome), but for most common, acute etiologies (such those mentioned above) there is very little data on the prevalence hearing loss progression, and the rate of such progression.

This has led to the impression, even amongst hearing healthcare professionals, that progressive hearing loss is not common and that most acquired hearing loss rapidly stabilizes after an initial insult. The fact is that there are very few clinical studies that have specifically explored the progression of hearing loss other than the age-related variety. Various practical issues contribute to this lack of data. In the clinical "case study" papers, there tends to be no routine follow-up, especially over many months and years.[5] Epidemiological studies do not seem to capture serial assessments of hearing loss.

So, what is my point here? It is just to say that progressive hearing loss after initial insult is much more common than is suggested by the clinical evidence available. Knowing the many pathogenic mechanisms that can occur in the cochlea, tissue injury spread and the resulting hearing loss progression is likely the norm rather than the exception. Information about any hearing loss progression is important; verification will require a routine follow-up.

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## References


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