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Auditory Neuropathy is a Factor in All Cases of Cochlear Hearing Loss

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As this current edition of *Canadian Audiologist* is published, the ENT World Congress has just wrapped up in Paris. A large portion of that meeting is devoted to audiology because, worldwide, audiology and otolaryngology are very closely associated (much more so than in Canada). At this congress, I had the privilege (task) of presenting an audiology keynote lecture entitled: *"Central auditory changes in sensorineural hearing loss."* I had a message to get out that I will share with you here.

While it is convenient to classify hearing loss according to the most obvious site of lesion (e.g., conductive, cochlear, central) there is likely no "peripheral" hearing loss that does not cause changes to central auditory pathways. We have become accustomed to thinking about these hearing loss types as separate entities, which of course they are not. Let me start with the most dramatic example of how a cochlear hearing loss can have an effect the central auditory system. When there is a total lack of cochlear function from birth, an auditory cortex will not develop! We have brain-imaging evidence that in profound congenital loss, the cortical areas normally associated with hearing become used for other sensory purposes (resulting from cross-modal plasticity). This lack of auditory cortical substrate is confirmed by scores (if not hundreds) of examples of congenitally deaf adults who get zero benefit from cochlear implantation.

The normal development of the central auditory brain depends on stimulation by an adequate pattern of neural activity from the cochlea. In infants, when the auditory pathways are still developing and in a plastic state, a cochlear hearing loss will permanently change central auditory networks. I have published extensively on reorganization of cortical frequency mapping resulting from neonatal high frequency hearing loss.[1] Even when cochlear lesion occurs in adults there can be some modification to cortical frequency mapping.[2]

But let us consider more common etiologies of peripheral hearing loss, such as noise exposure,

infection, ototoxicity, aging. A large number of human and animal studies have demonstrated that no matter whether the initial site of cochlear damage is the stria or the haircells there is ultimately a degeneration of the inner haircell synapse. This is arguably the most vulnerable structure of the cochlea. When this cochlear afferent link is broken, there is always some degree of degeneration of spiral ganglion neurons. We have known this sequence of neural degeneration for decades from temporal bone histology studies. We are reminded of this spiral ganglion cell degeneration in every case of cochlear implantation, where the effectiveness of electrical stimulation of the cochlea depends on the pattern of spiral ganglion cell survival.

We have recently become accustomed to thinking more about auditory neuropathy spectrum disorder and "hidden hearing loss."[3,4] Both of these hearing loss types make us question whether there is any real boundary between the inner ear and cochlear afferent neurons and central auditory pathways.

As a final provocative note, I will add that our increased understanding the very intimate links between cochlear function and central auditory pathway integrity should lead us to consider whether so called central auditory processing disorders are indeed "central" problems. It is likely that many such cases have a peripheral cause. I trust that you will now see the verity of the title for this column: ***"Auditory neuropathy is a factor in all cases of cochlear hearing loss."***

References

1. Harrison, Nagasawa, Smith, et al. Hear Res 1991;54:11–19.
2. Robertson and Irvine. J Comp Neurol 1989;282:456–71.
3. Starr, Picton, Sininger, et al. Brain 1996;119:741–53.
4. Kujawa and Liberman. J Neurosci 2009;29:14077–85.