The physiology of hearing impairment 1


Hearing impairment can affect each of the steps in auditory transduction

**Conductive** - damage (eardrum puncture), pathological processes (bone growth), or blockage of external and middle ear (not shown above).

**Metabolic** - damage or degeneration of the stria vascularis (SV) or other components of cochlear homeostasis, indirectly affecting hair-cell transduction.

**Sensorineural** - damage to, degeneration of, or decreased gain of hair cells (IHC, OHC), degeneration of auditory nerve fibers (SGN, AN), or central auditory neurons and circuits.
These lecture focuses on sensorineural hearing impairment (SNHL) and the functional consequences of damage to hair cells or auditory nerve fibers.

SNHL has four well-studied causes:

- **Acoustic trauma** - exposure to very loud sounds for a long enough time to damage hair cells or auditory-nerve fibers.

- **Ototoxic substances** - some antibiotics (gentamycin), cancer drugs (cisplatin), and various other toxins specifically damage hair cells or other components of the cochlea.

- **Genetic defects** - any genetic defect that affects an essential functional element of cochlear physiology can cause deafness (e.g. Usher’s syndrome, caused by a defect in a myosin or in one of four other genes).

- **Aging** - which appears to operate indirectly via degeneration of the stria vascularis, leading to a reduction in the endolymphatic potential (EP) and a decrease in gain of hair cells.

The perceptual deficits of SNHL:

- Loss of audibility
- Loudness recruitment
- Degraded frequency tuning
- Auditory neuropathy, degraded temporal precision
- Tinnitus
To illustrate two of the primary behavioral effects of hearing impairment, look at **equal-loudness contours**, along which tones of different frequencies sound equally loud.

**Two problems:**

1. Soft sounds can't be heard
2. Loudness growth is faster than normal (recruitment)
The largest problem in hearing impairment is **loss of audibility**, sounds cannot be heard. The extent to which audibility alone can account for the degraded performance of hearing impaired listeners is evaluated with the **articulation index**.

\[ AI = P \int_{f_0}^{f_1} I(f) W(f) \, df \]

- \( P \) - proficiency (fudge factor)
- \( I(f) \) - relative importance of different frequencies for understanding speech, empirically determined in normal listeners
- \( W(f) \) - fraction of the speech that is audible above noise masking or elevated threshold (roughly the red region at right)

AI analysis shows that, with parameters \( P \) and \( I(f) \) set for normal listeners, the AI accurately predicts loss of performance in mild to moderately impaired listeners (<50 dB), but not in moderate-to-severe group (>55 dB). Apparently there are additional problems in such subjects.

The task was recognition of spoken word lists with 9 different degrees of LP, HP, and BP filtering.

The difference is usually worse in older persons.

Pavlovic, 1984
The second problem, **recruitment**: loudness grows more rapidly than normal over some range, so that sounds of ≈80-100 dB are equally loud in a normal and an impaired listener.

The behavior of impaired loudness near threshold is controversial. Some (Buus and Florentine, 2001) claim that loudness is elevated right at threshold, but the preponderance of evidence indicates that this is not so and that loudness growth is more like the blue curve (Moore, 2004).

Recruitment is important in hearing-aid design, because the reduced dynamic range requires compressor circuits in aids. These circuits are problematic in that they are nonlinear and distort the sound.

Buus and Florentine, 2001

The third problem: **psychophysical auditory filters are wider** in hearing-impaired ears. This means the ability to analyze sound by its frequency content is impaired.

Glasberg and Moore, 1986
The implications of broadened auditory filters:

A vowel spectrum

The same spectrum on the cochlea’s frequency scale (in units of auditory filter bandwidths)

The theoretical excitation pattern on the basilar membrane for this stimulus in a normal ear (the spectrum smoothed by auditory filters)

Note the loss of spectral detail, e.g. the locations of the formant peaks, with decreased cochlear filtering. This is one likely explanation of below-AI performance in speech perception and suggests problems with background noise.

Moore, 1995

The fourth problem: **auditory neuropathy** in which the thresholds are normal or show mild impairment, but subjects have considerable difficulty with complex perception, like speech.

Thresholds of a person at two ages (19 and 32) show mild impairment (<60 dB) in both ears.

However, the speech perception performance (% of words on an open set test) is much worse than expected from the hearing loss (green and orange symbols).

The other data are from a population of similar subjects.

Starr et al. 1996
Auditory neuropathy subjects show little or no ABR, meaning a lack of synchronous activity in the CNS. However, their DPOAEs (distortion product otoacoustic emissions) are normal, showing no deficit in outer hair cell function.

Starr et al. 1996

The physiological correlates of perceptual problems in hearing impairment. **Inner** and **outer** hair cells: outer hair cells amplify BM displacement; inner hair cells are the transducers for ANFs

C. Smith
Hair cell damage has been studied experimentally with acoustic trauma. These data show auditory nerve tuning and thresholds after such trauma (temporary threshold shift has been allowed to resolve).

Note the
1. threshold shifts and
2. broadening of tuning when compared to normal tuning curves (gray).

(The exposure was a 50 Hz noise band centered at 2 kHz, 115 dB for 2 hr, 54 day recovery)

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Another example of hair cell loss from acoustic trauma.

In this case there is a dead region marked by the red band. At these frequencies, no auditory nerve fibers were found and there was substantial hair cell damage.

Dead regions have been described in psychophysical tests. Sophisticated masking methods can detect such regions (see Moore, 2007).

Importantly, amplifying sounds at frequencies in a dead region does not help speech perception, even though it should increase audibility (from the AI).

The reason is that sounds in a dead region are being heard through the spread of tuning curves from adjacent regions with existing IHCs and ANFs.

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What about the regions where hair cells remain, but substantial threshold shifts are still observed (the blue rectangle)?

A view of normal hair cells from above, showing the orderly rows of cilia.
Cilia in the normal guinea pig cochlea, showing tip lengths (arrows) between the tips of shorter cilia and the sides of their longer neighbors.

But intact hair cells may have damaged cilia after acoustic trauma.

Inner hair cell after acoustic trauma; damage is most severe to tallest cilia. Arrows show intact tip links, arrowheads show broken ones.

Outer hair cell after acoustic trauma; cilia are fused and missing tip links.
Generally there was a good correlation between threshold shift and ciliary damage, in ears with surviving hair cells but substantial threshold shifts.
Acoustic trauma produces a complex and variable mixture of IHC and OHC damage.

Ototoxic antibiotics (kanamycin) produce a simpler lesion in which regions with only OHC damage can be found.

Unit 50 was localized using HRP injection to the middle of such a region (dashed line). This example shows the effects of a severe outer hair cell lesion, with little inner hair cell damage, including minimal cilia damage.

Summary of the effects of different types of hair-cell damage

IHC damage produces threshold shift with no loss of tuning.

OHC damage produces threshold shift and broadened tuning.
Auditory nerve fibers are apparently driven by two opposite-phase processes, called C1 and C2; examples are shown below for two fibers from normal animals.

Acoustic trauma seems to specifically affect C1 without modifying C2, shown below in two fibers from traumatized ears.
Hypersensitive tails in W-shaped tuning curves occur when there is no sign of a C1-C2 transition (no phase shift), presumably in the absence of C1.

Liberman and Kiang correlated hypersensitive tails with cochlear regions where the IHCs are missing the tall row of stereocilia. Maybe C1 is the IHC driven through its tall stereocilia by sharply-tuned (due to the OHC) BM motion and C2 is the IHC driven through its short stereocilia.


Liberman and Kiang, 1984