Cochlear dead regions and their implications for hearing aid rehabilitation

This article describes what is meant by a dead region, describes a simple test for diagnosing dead regions in the cochlea and discusses treatment options for people with dead regions.

Sounds entering the ear give rise to vibration patterns on the basilar membrane within the cochlea. Each place on the basilar membrane is tuned to respond best to a specific small range of frequencies; high-frequency sounds produce maximum vibration towards the base and low-frequency sounds produce maximal vibration towards the apex. The frequency that leads to a maximal vibration at a given place on the basilar membrane is called the characteristic frequency (CF) for that place. In an ear with normal hearing, the patterns of vibration on the basilar membrane are strongly influenced by the activity of the outer hair cells (OHCs), which are minute sensory cells forming rows along the length of the basilar membrane. The OHCs play a role in what is called the “active mechanism” in the cochlea (Moore, 2007). They do this by changing their stiffness and length in response to the vibrations. This activity of the OHCs enhances the response to weak sounds (increasing the amplitude of vibration) and sharpens the tuning on the basilar membrane. This sharpening increases the frequency selectivity of the auditory system, i.e., its ability to separate out the different frequencies that are present in complex sounds such as speech and music. The amplified vibrations are then detected by the inner hair cells (IHCs), which form a single row running along the length of the basilar membrane. In response to vibrations on the basilar membrane, the IHCs generate electrical signals and release neurotransmitter, and this in turn leads to neural activity in the auditory nerve.

Cochlear hearing loss is often associated with damage to the hair cells within the cochlea (Schuknecht, 1993; Moore, 2007). This damage can give rise to raised hearing thresholds (i.e., hearing loss as measured by the audiogram) in two main ways. Firstly, damage to the OHCs impairs the active mechanism in the cochlea, resulting in reduced basilar membrane vibration for a given low sound level (Ruggiero & Rich, 1991). Hence, the sound level must be larger than normal to give a just-detectable amount of vibration. Secondly, IHC damage can result in less efficient stimulation of the auditory nerve. As a result, the amount of basilar membrane vibration needed to reach the hearing threshold is larger than normal (Liberman & Dodds, 1984). A cochlear hearing loss up to about 55 dB may be caused by damage to IHCs alone. A hearing loss greater than 65 dB nearly always involves some loss of function of IHCs as well as OHCs.

In some cases, the IHCs at certain places along the basilar membrane may be completely non-functioning. In addition, the auditory neurones making contact with those places may be non-functioning. Places with non-functioning IHCs and/or neurones have been referred to as “lacunae” (Troland, 1929) and “holes in hearing” (Shannon et al., 2002), but I have used the blunt phrase “dead regions” (Moore & Glasberg, 1997; Moore et al., 2000; Moore, 2001; Moore, 2004) and that phrase seems to be catching on. Figure 1 shows the dissected cochlea of a person who had been exposed to intense impact sounds (gunshots) before dying in an incident unrelated to gunshots! The dark lines are the neurones that would eventually get together and form the auditory nerve. There are no neurones coming from the basal part of the cochlea, indicating a dead region.

Figure 1: Cochlea from a 25-year old man who had been exposed to gunshots. The dark lines show auditory neurones. There are no neurones coming from the basal part of the cochlea, indicating a dead region.
Basilar-membrane vibration that occurs within a dead region is not detected by the neurones connected to that region. Say, for example, that the IHCs at the basal (high-frequency) end of the cochlea are non-functioning. Neurones connected to the basal end, that would normally have high CFs, will not respond. However, if a high-frequency pure tone is presented, it may be detected if it produces sufficient basilar-membrane vibration at a region closer to the low-frequency, apical end. In other words, a high-frequency sound may be detected via neurones that are tuned to lower frequencies. Similarly, if there are no functioning IHCs in an apical region of the cochlea, a low-frequency tone may be detected via neurones that are tuned to higher frequencies. Because of this possibility, the "true" hearing loss at a given frequency may be greater than suggested by the audiometric threshold at that frequency. Also, for this reason, dead regions are not easy to diagnose from the pure-tone audiogram, although a hearing loss greater than 70 dB is often associated with a dead region (Aazh & Moore, 2007; Viray & Moore, 2007a).

A dead region can be characterised in terms of the range of CFs that would normally be associated with that region. In other words, a frequency-to-place map is used to relate the cochlear location at each edge of the dead region to frequency. This is illustrated in Figure 2.

Figure 2: Illustration of how the edge of a dead region can be related to frequency, using a frequency-to-place map of the cochlea. In this example, the dead region starts at an edge frequency of about 2500 Hz, and extends upwards towards higher frequencies.

Say, for example, that the IHCs are non-functioning over a region of the basilar membrane having CFs in the range 2500 to 20000 Hz. One might describe this as a dead region extending from 2500 Hz upwards. The lower "edge frequency" of the dead region in this example is 2500 Hz. Henceforth, the edge frequency is denoted fe.

We have developed a test for diagnosing dead regions which is quick and easy to administer, and which is suitable for use in clinical practice. The development and validation of the first version of the test are described in Moore et al. (2000). The test involves measuring the threshold for detecting a pure tone presented in a background noise called "threshold-equalizing noise". Hence the test is called the "TEN" test. The noise was synthesised in such a way that the threshold for detecting a tone in the noise, specified in dB SPL, was approximately the same for all tone frequencies in the range 250 Hz to 10 kHz, for people with normal hearing. The masked threshold is approximately equal to the nominal level of the noise specified in dB SPL.

When the pure-tone signal frequency falls in a dead region, the signal will only be detected when it produces sufficient basilar membrane vibration at a remote region in the cochlea where there are surviving IHCs and neurones. The amount of vibration at this remote region will be less than in the dead region, and so the noise will be very effective in masking it. Thus, the signal threshold is expected to be markedly higher than normal. Moore et al. (2000) proposed the following rule: a dead region at a particular frequency is indicated by a masked threshold that is at least 10 dB above the absolute threshold and 10 dB above the nominal noise level.

To make the test easy to administer, the TEN test was recorded on a CD; the noise is on one channel and test tones are on the other channel. It is intended that the signals from the CD are fed through a two-channel audiometer. The methods used to conduct the test are similar to those used for conventional pure-tone audiometry, except that the signal threshold is measured in the presence of a continuous background noise.

A problem with the first version of the TEN test is that the clinician using the test had to measure absolute thresholds (audiometric thresholds) twice, once using the tones generated by the audiometer, with level specified in dB HL, and once using the tones from the CD, with level specified in dB SPL. This is inconvenient for the clinician. To overcome this problem, a second version of the TEN test was developed in which the noise was designed to give equal masked thresholds in dB HL for all
frequencies from 500 to 4000 Hz, for normally hearing people (Moore et al, 2004). This version is called the "TEN(HL)" test. As all calibrations are in dB HL, absolute thresholds can be measured either using the tones generated by the audiometer, or using the test tones from the CD; the results should be very similar.

Another advantage of the second version of the test is that the noise has been designed to have minimal amplitude fluctuations; such noise is called "low-noise noise" (Pumpkin, 1985). Figure 3 shows the spectrum of the noise (top) and a sample of the waveform (bottom). Note that all peaks and dips in the waveform are of similar magnitude. This characteristic allows high noise levels to be used without significant distortion being produced by the audiometer or earphone. Hence, testing is possible for more severe hearing losses than could be assessed with the earlier version, without any special equipment being required.

Figure 3: Spectrum (top) and a segment of the waveform (bottom) of the noise used for the TEN(HL) test.

The criteria for diagnosing a dead region are similar to those for the earlier test: a dead region at a particular frequency is indicated by a masked threshold that is at least 10 dB above the absolute threshold and 10 dB above the nominal noise level in dB HL. When used as recommended in the CD booklet, the test takes about 5 minutes per ear, if testing is conducted for all possible frequencies.

The presence or absence of dead regions can have important implications for fitting hearing aids and for predicting the likely benefit of hearing aids. When a person has a dead region, there may be little or no benefit in applying amplification (via a hearing aid) for frequencies well inside the dead region. However, for people with high-frequency dead regions, there may be some benefit in applying amplification for frequencies up to about 1.7 kHz (Vickers et al, 2001; Baer et al, 2002). For example, if a person has a dead region which starts at 1000 Hz and extends upwards from there, there may be some benefit in amplifying frequencies up to 1700 Hz. However, there will probably be no benefit of applying amplification for frequencies above about 1700 Hz. Trying to achieve sufficient gain for frequencies above 1700 Hz might lead to problems with distortion and acoustic feedback. For a person with an extensive high-frequency dead region, a hearing aid incorporating frequency transposition might be a viable option (Simpson et al, 2005; Robinson et al, 2007). For people with low-frequency dead regions, there appears to be some benefit of amplifying frequencies above 0.57 kHz, but not of amplifying frequencies below 0.57 kHz (Vinay & Moore, 2007b; 2008).

Details about how to obtain the TEN(HL) CD can be obtained from:
http://hearing.psychol.cam.ac.uk/dead/dead.html

References


