NIHL - understanding audiograms and medical causation

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How the ear works

The human outer ear performs rather like a funnel, channelling the sounds around us along the ear canal.

The sound then strikes and vibrates the tympanic membrane, more commonly referred to as the ear drum, a thin, delicate membrane stretched across the opening of the middle ear cavity. This is an air-filled cavity maintained at atmospheric pressure through the Eustachian tube, which connects with the upper respiratory tract behind the nose. The eardrum converts the acoustic disturbances of sound into mechanical vibrations.

Three tiny bones behind the eardrum, known as the malleus, incus and stapes (colloquially known as the hammer, anvil and stirrup), or collectively as the ossicles, transmit these vibrations via the fluid filled canal of the inner ear to the cochlea.

The cochlea contains very many intricate, highly sensitive, hair-like cells, suspended in fluid. These react to the vibrations, passing electrical and chemically generated impulses along the auditory nerve to the brain, where they are perceived as the sensation of sound.

Noise-induced hearing loss (NIHL)

Imagine, for a moment, the effects of a violent wind beating down on a field of corn until eventually all is flattened and destroyed, unable to recover – ever. We have all seen the situation to varying degrees, usually just before harvesting!

This analogy is apt to describe the process by which noise damages the hearing. Exposure to excessive noise, in the form of the energy of the vibrations of sound (the wind), progressively damages the fine, sensitive, hair-like cells (the corn) in the cochlea of the inner ear, until they are damaged beyond recovery.

It is important to stress that any noise-induced hearing loss is irreversible. It cannot be remedied by surgery and hearing aids are of little or no value.
In the majority of cases, noise damage increases with regular exposure to excessive noise. It is insidious and cumulative, so that when a person starts to become aware of hearing difficulties, the damage process will already have been occurring for some time. Subjective recognition comes too late to permit recovery of the hearing ability, which has already been lost.

The extent of NIHL is dependent upon the period of exposure to noise, the loudness of the noise and the individuals own particular susceptibility to noise damage. It is impossible to identify individuals susceptibility other than by comparison of the results of regular hearing tests undertaken on a group all exposed to similar noise levels.

Hearing loss occurring within the cochlea of the inner ear is what is known as sensori-neural loss, though the term also includes malfunction of the auditory nerve and brain. The sensory system within the cochlea is continuous with that in the vestibular organs which control balance, so that anything causing injury to the inner ear may, in addition to producing a loss of hearing, also disturb the balance, although there is no evidence that noise can affect the balance of organs.

Sensori-neural hearing loss must be distinguished from what is known as 'conductive' loss. The latter is a loss resulting from malfunction of the middle ear and will not be noise induced (unless as the result of a single noise trauma e.g. an explosion which perforates the ear drum.)

Whilst NIHL is sensori-neural in type, a sensori-neural hearing loss may not be noise induced, but can be congenital or can arise from head injuries, electric shock, exposure to industrial poisons and the use of OTO-toxic drugs such as streptomycin used in the treatment of tuberculosis. Indeed, the loss of hearing which results from the ageing process (known as ‘presbyacusis’) is a form of sensori-neural deafness, even though by definition it is not noise induced.

In the case of head injuries, a blow to the head is the equivalent of a single severe impulse noise exposure transmitted to both ears more or less equally by bone conduction through the skull. The result may be a sensori-neural hearing loss of a temporary or permanent nature. In a more severe case involving fracture of the skull, if the fracture passes through the cochlea, a severe and permanent sensori-neural hearing loss results. Hearing loss in that case is usually unilateral.

**The medical examination**

An ear, nose and throat specialist can be instructed to establish the nature of the complaints both in regard to hearing ability and exposure to noise. The expert should discuss past medical history, to ascertain the possibility of any other causes for any hearing loss and enquire about any family history of deafness.

The examination requires that the claimant be away from noise at work (or from other sources) for a minimum of 12 hours. This is because of what is known as ‘Temporary Threshold Shift’, which in the context of noise means temporary deafness resulting from noise exposure, though the term generally can be applied to temporary hearing loss from any cause.

Clinical examination will usually involve a visual examination of the eardrum (tympanic membrane) to ensure that there is no obstruction (eg by wax or foreign bodies) of the ear canal. It may also involve two tuning fork tests. The Weber Test which indicates asymmetry in the hearing between the ears and the Rinnes Test differentiates between conductive (middle ear) and sensori-neural (inner ear) hearing losses. The tests are not infallible, but they take very little time and may give the Expert a useful feel for the case at the outset. They have no quantitative value.
Note may also be made of the patient’s apparent ability to hear and understand softly spoken conversation.

**The pure tone audiogram**

The major investigation is then normally pursued by means of what is known as manual pure-tone audiometry. This measures the patient's voluntary responses to pure-tone signals relayed to him by headphones. The results are plotted on a graph called an audiogram.

Normal hearing is shown by a relatively flat line with up to 20dB loss over all test frequencies.

![Figure 1 – normal hearing](image)

NIHL is usually bilateral and characterised by high frequency hearing loss which usually occurs first at 4 kHz and then progresses into adjacent frequencies. The loss at 4 kHz progresses over the first 10 years and then tends to stabilise.

NIHL is usually maximal at 4kHz and so there may be a characteristic dip or ‘notch’ usually at this frequency but which can alternatively occur at 3 or 6kHz – but it is of note that notching is not unique to NIHL and may be due to another cause. Equally, whilst a ‘notch’ is a good diagnostic indicator, it does not have to be present for a diagnosis of NIHL to be made.

Although NIHL is maximal at between 3-6 kHz it probably affects all frequencies between 1-8 kHz given sufficient dose/duration of exposure.

Once exposure to noise ceases the NIHL does not progress further.

Any low frequency loss below around 1 kHz is generally not related to noise.

![Figure 2 – Noise-induced hearing loss](image)

Other causes of sensori-neural (inner ear) hearing loss include:

- ototoxic damage
- genetic damage
- German measles in pregnancy
premature birth
high cholesterol levels
diabetes
rheumatoid arthritis/MS
heart disease
trauma
Ageing (see the Age related loss table).

Age-related hearing loss (AAHL) or presbyacusis starts from around age 20 and accelerates with age generally affecting all frequencies causing maximal losses above 6 kHz.

Conductive hearing loss is demonstrated by an ‘air-bone’ gap i.e. loss arising from a defect in the sound conducting mechanism in outer/middle ear. This can arise from:

- a perforated ear drum
- otosclerosis
- outer/middle ear infections
- Head injury.

A diagnosis of NIHL may be relatively straightforward where there is a history of exposure to excessive noise, a typical audiometric notch between 3-6 kHz and no complicating factors/diagnostic competitors.

In many cases diagnosis is far more difficult and the audiometric presence of NIHL, if present, may be obscured by AAHL and/or other causes of hearing loss. Many experts now follow the Coles Guidelines in determining diagnosis. (For guidelines on the diagnosis of NIHL for medico legal purposes see Clin. Otolaryngol 2000, 25, 264-273, R Coles et al.)
ERA testing

Whilst pure tone audiometry is the most accurate method of assessing hearing levels it is susceptible to an element of manipulation as it is subjective. This can result in variable and confusing results suggesting very high levels of hearing loss.

Where there are concerns with the accuracy of pure tone audiometry then a more objective test is Cortical Evoked Response Audiometry (CERA).

This can be arranged by, but is not undertaken by the ENT specialist. The most likely explanation of the inconsistencies will be deliberate exaggeration by the patient.

There are several different methods of ERA, but a typical examination has the patient in a quiet room, e.g. reading a book/magazine. Sound signals are introduced into the ear and the brain’s response monitored by means of electrodes placed on the scalp. Those responses are in turn analysed by computer. The important points are that the patient himself is quite unable consciously to influence the responses; it is his brain’s activity and not his perception, which is measured.

The disadvantage of ERA is that it does not permit so precise a means of quantifying hearing loss as pure-tone audiometry with a fully co-operative Patient. The results may (e.g. because of misleading neural activity in a tired or tense patient) exaggerate the apparent hearing loss by up to 10dB or more.

ERA can be taken over both air and bone conduction readings, but is normally restricted to air conduction readings unless there is an obvious air/bone gap. Unfortunately, an ERA takes time and the more frequencies that are covered, the more tired the subject gets and the more confused and irregular become his brain patterns.

In short, ERA, results by themselves should never be accepted as an accurate picture of the patient’s degree of hearing loss. However, the technique does provide a very useful means of identifying the patient who is, for whatever reason, disposed to exaggerate his hearing loss.

Management of hearing loss including hearing aid provision

The mainstay of treatment for hearing loss is the use of a hearing aid, or hearing aids. Such devices work on the basis of amplifying sounds to a level that in theory will allow the listener to discriminate between the different sounds. In practice this depends on numerous factors. These include motivation, expectation and the willingness on the part of the wearer. However, it is generally accepted that a subject with a significant hearing loss will do better with a hearing aid than without.

Typically digital hearing aids are widely issued both by the NHS and in the private sector. This is a direct consequence of the government’s Modernising Hearing Aid Service Initiative in April 2000. Since April 2005 all NHS hearing aid centres in England have provided modern behind the ear (BTE) digital aids.

Types of hearing aid

All digital aids work in the same way and have three basic parts: a microphone, amplifier and speaker. The aid receives sound through the microphone and then coverts the sound wave to electrical signals which are amplified and then relayed to the ear via the speaker.
BTE (Behind the Ear)

These have an earmould or a soft tip that fits snugly inside the ear. The hearing aid rests behind the ear and a soft plastic tube connects it to the earmould/soft tip and channels sound from the aid into the ear. Some models have twin microphones which allow a user to switch between all-round sound and more directional setting that helps focus on wanted sound amongst a noisy environment. They can be programmed in a precise way to suit the hearing loss and everyday needs. The aids generally have a lifespan of around five years.

ITE (In the Ear)

These fit entirely into the ear. The working parts are either in a small compartment clipped to the earmould or inside the mould itself. ITE aids tend to need repairing more often than BTE aids.

ITC (In the Canal)

These are even smaller than ITE aids and less visible. They are unsuitable for severe hearing losses.

The advantages and disadvantages of the different types of aid are summarised in the table below.

<table>
<thead>
<tr>
<th>Type of aid</th>
<th>Advantages</th>
<th>Disadvantages</th>
</tr>
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</table>
| **BTE**     | • Suitable for mild-profound hearing losses.  
• More amplification than ITE and ITC aids. A must for profound loss.  
• Size permits complex circuitry with numerous options.  
• Can be less visible than ITE aids if an ‘open fit’ aid is used—here there is a small BTE aid with a narrow tube inserted into the ear canal.  
• Open fit aids good for people with build up of ear wax and are less occlusive so sound not ‘plugged up’.  
• Longest battery life. | • More visible although with normal length hair often covered. |
| **ITE**     | • Suitable for mild to severe hearing loss.  
• Easy to insert and remove. | • Not suitable for profound hearing loss.  
• Less reliable than BTE aids.  
• Shorter battery life.  
• More visible than ITC aids and sometimes BTE aids.  
• Prone to damage by ear wax. |
The management of hearing loss may include other measures which operate either independently or in conjunction with hearing aid use. These may include the use of environmental devices, or training to maximise the use of residual hearing e.g. lip-reading. Some subjects may have a hearing loss that is too slight and only at certain frequencies which would not be suitable for amplification. At the other end of the range may be subjects whose hearing loss is so severe that they would receive no benefit from a hearing aid as they have no hearing ability to amplify. Such patients may require cochlea implants.

**Tinnitus**

This term describes the subject’s perception of noise in the ears or head. Medical text books typically define this as “the sensation of sound not brought about by mechano-acoustic or electrical means”. It is a symptom and not a disease. It is subjective i.e. a condition the patient experiences, and is not tangible. Most cases arise as a result of cochlear damage, and consequently tinnitus is usually seen in patients with sensorineural hearing loss. However, it can arise where there is no hearing loss and is not present with many patients who have a hearing loss.

The prevalence of tinnitus is said to be as high as 35% for intermittent non-troublesome tinnitus, while severe disabling tinnitus is said to be presenting 0.5% of the population.

Tinnitus may be intermittent or permanent. Noise exposure is the second most common cause of tinnitus in the UK and National Study of Hearing results have shown the incidence of tinnitus increases by up to 50% in persons exposed to noise compared to those with no previous noise exposure.

Another common cause of tinnitus is head or neck injury and several medical studies show a possible causative link between injury and the onset of tinnitus. Generally, whatever causes the cochlea damage will also be responsible for the tinnitus.

Tinnitus typically arises with a gradual onset although it may develop rapidly over a matter of minutes or hours. Cases of rapid onset are usually attributable to a specific disorder e.g. viral infection, sudden noise exposure, vascular accident or head injury. The impact of tinnitus on individuals varies widely and it is difficult to quantify. The effects may include sleep disturbance, effects on mood and concentration.

The level of tinnitus is often graded on an arbitrary scale:

TRIVIAL-MILD-MODERATE-MODERATE/SEVERE/SEVERE-SEVERE.

This follows various descriptions of the symptoms and impact and referred to in the medical practitioners’ text “Assessment of Hearing Disability (King et al, 1992).
Tinnitus is difficult to treat and there is a host of options which include surgery, medication, homeopathic remedies, psychological input and prosthetic devices. One treatment which is typically adopted is tinnitus retraining therapy (TRT). This is a form of habituation and conditioning of the patient to the condition.

Age associated hearing loss (dB average over frequencies 1, 2, 3 kHz based on ISO 7029)

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Calculating hearing loss

Claimant is a 55 year old man. The audiogram shows the following reading for both ears:

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<th>Frequency (kHz)</th>
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<td>2</td>
<td>50</td>
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<td>3</td>
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</table>

1. Add 3 levels together and divide by 3 = total hearing loss

\[35 + 50 + 65 = 150\]

150 divided by 3 = 50 dB total hearing loss

2. With reference to ISO 7029 take off hearing loss due to age = 10dB

50dB – 10dB = total loss due to noise exposure

= 40dB

Claimant is 52 year old woman. The audiogram shows the following readings:

Left Ear

<table>
<thead>
<tr>
<th>Frequency (kHz)</th>
<th>Level (dB)</th>
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<tr>
<td>2</td>
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<td>40</td>
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Right Ear

<table>
<thead>
<tr>
<th>Frequency (kHz)</th>
<th>Level (dB)</th>
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<tbody>
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<td>1</td>
<td>5</td>
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<tr>
<td>2</td>
<td>10</td>
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</table>

1. Repeat step 1 in example 1 above for each ear this time which gives the following readings:

Left ear 33.3 dB
Right ear 8.3 dB

2. Calculate average binaural using the DSS formula by multiplying the reading for the better ear by 4, adding it to the reading for the worse ear and dividing the total by 5

\[8.33 \times 4 + 33.33 = 66.65 \text{ divided by } 5 = 13.33\]

Thus 13.33 dB = total hearing loss

3. Repeat step 2 in example 1 above

13.33 – 6.66 = 6.67dB

Total loss due to noise = 6.67dB
Common definitions and terms

- **Acoustic trauma**
  The term used to refer to damage to the inner ear, usually from noise exposure.

- **Auditory brainstem response audiometry (ABR)**
  Also known as brainstem evoked response audiometry (BSERA) or brainstem auditory evoked potentials (BAEP), this is an objective test of hearing usually over the frequency range of 2-4 kHz. The threshold over this range is identifiable, but not the threshold at specific frequencies.

- **Auditory nerve**
  Also known as the cochlear nerve or part of the VIII cranial nerve, this links the cochlea to the brainstem in which are located the cochlear nuclei.

- **Audiometry**
  The science of hearing assessment.

- **Air conduction levels**
  The result from measurement of hearing with headphones.

- **Bone conduction levels**
  The result from hearing measurement using a bone conductor.

- **Cochlea**
  The part of the inner ear containing the organ of hearing.

- **Cortical evoked response audiometry (CERA)**
  Objective test of hearing allowing identification of thresholds at specific frequencies.

- **Decibel (dB)**
  Unit of measurement of hearing. When used on pure tone audiograms the scale is in dBHL.

- **Free field speech audiometry**
  The testing of subjects to speech (usually spoken words) in the free field environment.

- **Hair cells**
  The sensory component at the beginning of the neural auditory pathway. These are located in the Organ of Corti, found in the cochlea.

- **Headphone pure tone audiometry (or pure tone audiometry)**
  The testing of hearing with the presentation of pure tones through headphones.

- **Hearing aids**
  Amplification devices, using either the older analogue technology, or the more modern digital technology.

- **Hertz (Hz)**
  Unit of measurement of frequency. One thousand hertz is a kilohertz (kHz).

- **Nonorganic hearing loss**
  A hearing loss that is not present, or in excess of a genuine hearing loss.

- **Obscure auditory dysfunction**
  A term used to describe the condition where hearing is within clinical range of normality, but the subject presents with difficulties discriminating sounds, usually in noisy conditions.
- **Oto acoustic emissions**
The response arising from outer hair cells of the cochlea following click stimulation which can be measured and displayed.

- **Ototoxicity**
Damage to the inner ear as a result of drug ingestion.

- **Presbyacusis**
Hearing loss related to the ageing process.

- **Pure tone audiogram**
The graph used to represent hearing.

- **Rinne test**
A clinical test of hearing with a tuning fork. Best used for identification of a conductive hearing loss.

- **Tympanometry**
The procedure in which the middle ear function is assessed objectively.

- **Vestibular system**
The part of the inner ear dealing with balance.

- **Weber test**
A clinical test of hearing with a tuning fork used for identifying the better functioning of the two inner ears.

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