Chapter 16: Sensorineural hearing loss

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The clinician finds himself faced with a difficult diagnostic prospect each time he is confronted by a patient with sensorineural deafness. In this chapter an attempt is made to devise a practical approach to this problem in the everyday setting of the otolaryngology clinic.

Usually the patient has been aware of a hearing loss, but sometimes is not until attention has been drawn to it as, for example, in a pre-employment or some other routine medical examination. Not infrequently the remarks of family or friends will be the stimulus to seek an otological opinion, often because of a tendency to turn up the television volume to the annoyance of the family.

Our aim is to devise a method which will chart a direct and useful route through the symptoms and signs of sensorineural deafness towards the identification and management of the underlying pathology, a means by which the benefits of knowledge, experience and intuition, may be put to best use. Unfortunately, a diagnosis of the pathology will frequently elude us and, in some instances, the most important objective will be a negative one, that is to find out what is not wrong.

In the outpatient clinic

An initial long and detailed history may be wasteful of time and a brief and general otological history is taken first. This is followed by clinical examination of the ear, including an assessment of the hearing by tuning fork tests. A pure tone audiogram is then carried out with the measurement of speech discrimination ability in each ear.

With the information gleaned from these initial steps, a more specifically directed history, examination and investigations may then be indicated.

History

Initially, enquiry is directed towards obtaining the patient's outline of his symptoms. When did he first notice hearing impairment? Was it of sudden or gradual onset? Is it unilateral or bilateral? Which ear does he consider to be the better? Is the hearing loss progressive, static or fluctuant? Is there or has there been otorrhoea? Were any incidents or specific circumstances associated with the onset of the deafness? How does the hearing loss affect him in his everyday life? Has he any tinnitus or vertigo?

Examination

A first requirement is the adequate visualization of both tympanic membranes. When the external auditory meatus is occluded by wax, hairs or debris, there may be a temptation to proceed with audiometry on the assumption that the bone conduction curve will at least give an idea of the severity of the sensorineural component of the deafness. This is not necessarily so. A small conductive component can convert a severe sensorineural deafness
into an apparently profound deafness. It is imperative that the ear canals are clear before carrying out audiometry. It is at this time that one ought to detect, and indicate to the audiology technician, an ear canal that may collapse under the pressure of the earphone.

An assessment of the overall ability to communicate will be made at this time and it is easy to eliminate any possibility of lip reading, by talking to the patient while cleaning out the ears and inspecting the tympanic membranes. Such an assessment is of importance both clinically and in medico-legal cases.

In clinical assessment of speech discrimination, it may be essential to mask the contralateral ear with the Barany noise box.

Routine use of tuning fork tests is an important clinical discipline. Patients not infrequently have uncharacteristic difficulty when presented with choices and allowance must be made for incorrect tuning fork responses occurring in patients who are unable to accept what to them is conflicting information from their senses, for example the lateralization to the deaf ear in conductive hearing loss. It may be helpful to use both the 256 Hz and 512 Hz tuning forks. In compensation cases the responses to the tuning fork tests sometimes suggest a lack of cooperation.

**Audiometry**

Simple pure tone and speech audiometry (PBmax), with masking, have become the anchor of the clinical approach to sensorineural hearing losses. These routine measurements are generally very reliable, although constant vigilance should be maintained with regard to spurious audiometric responses.

The frequencies normally measured are 250 and 500 Hz, 1, 2, 4 and 8 kHz. In many situations it is desirable to include the 3 and 6 kHz frequencies. Measurement of these latter frequencies is required for certain pre-employment examinations including military service and also may be useful in cases of mild noise-induced hearing loss.

In our everyday practice we consider the lower limit of normal hearing to be 20 dB.

Speech scores are measured, based on 25 phonetically-balanced words, presented at approximately 40 dB above the average pure tone threshold for 500 Hz, 1 and 2 kHz (Kerr and Smyth, 1972). There is no necessity at this stage for a time-consuming speech discrimination curve.

**Evaluation of the audiogram**

In the absence of a conductive loss there will be three groups:

1. bilateral hearing loss
2. unilateral hearing loss
3. those found to have apparently normal hearing.
In each group, it is important to consider two points:

(1) is the recorded pure tone loss consistent with the clinical assessment?
(2) is the speech discrimination score consistent with the clinical assessment?

In unilateral hearing loss, with normal hearing in one ear, there should be no difficulty in communicating in the normal clinic situation.

**Bilateral sensorineural hearing loss**

In bilateral sensorineural deafness one should note whether the loss is symmetrical or nearly so, and how good or otherwise, the speech discrimination is in each ear.

In general clinical practice, the commonest cause of bilateral sensorineural deafness is presbyacusis. Schuknecht (1974) has described four types.

*Sensory presbyacusis* is due to loss of hair cells, possibly secondary to initial loss of supporting cells. This starts at the base of the cochlea and slowly progresses apically. Consequently the low frequencies are untouched initially with a steep fall off in hearing in the high frequencies. Speech discrimination in quiet surroundings remains good unless the speech frequencies become affected.

*Neural presbyacusis* is due to loss of auditory neurons. The whole length of the spiral ganglion is affected, but this is more marked at the basal turn. All the frequencies tend to be involved, but the higher frequencies are usually more affected. The prominent feature is a disproportionately severe loss of speech discrimination.

*Atrophy of the stria vascularis* gives a flat audiogram with good speech discrimination. (Although this is a degenerative condition it may also occur in younger people.)

*Inner ear 'conductive' deafness* gives the well known ski-slope audiogram with only slightly impaired speech discrimination.

It is not uncommon to find combinations of two or more of these degenerative processes in the one patient.

It is worth noting that the factors influencing speech discrimination scores are the severity of the loss for the speech frequencies, the angle of the audiometric curve, the presence or absence of recruitment and the number of available neurons in the auditory nerve.

*Industrial noise-induced deafness* is also common in many practices.

Before making a diagnosis of industrial noise-induced hearing loss, a careful work history should be taken. This should include some assessment of the noise exposure, its duration and probable levels. If the person has consistently to shout to communicate with colleagues close by, then there is a strong likelihood that the ambient noise is 90 dB(A) or above.
It is important that a diagnosis of noise-induced deafness should not be made simply on the patient's statement that his work is noisy. 'Noisy' is a relative term, but hair-cell damage does not usually occur in exposure to levels of less than 85-90 dB(A) for an average 40-hour week. A number of patients are likely to engage in litigation against their employers in respect of noise damage. A careful history is important and one must be circumspect in what one says lest a patient is induced to set off on a spurious and potentially embittering claim.

Frequent mention is found in the recent literature to losses arising from the leisure activities of young people, including acoustic trauma from fireworks and noise damage from personal stereos, discos and rock concerts. However, as long as they are not working in noisy environments, the risk to the audience is probably small because of the relative shortness of the periods of exposure. There is a definite risk for the performers, particularly if they engage in long practice sessions in small rooms.

*Ototoxic hearing loss* is usually bilateral and symmetrical. In those drugs which cause irreversible hair cell damage, the diagnosis usually becomes apparent from the history. However, in the reversible types, such as salicylate deafness, probing may be necessary because some patients fail to admit to the consumption of aspirin or related drugs.

Salicylate deafness characteristically produces a 'flattish' hearing curve accompanied by good speech discrimination. It will usually reverse on withdrawal of the drug.

A *dish-shaped audiogram* is occasionally found, in which the curve exhibits a moderately severe loss for the middle frequencies and good hearing for the high and low frequencies. It is usually without obvious cause and is, by custom, attributed to heredity.

*Asymmetrical bilateral hearing loss* is probably most commonly found following *weapon firing*, particularly one which is fired from the shoulder. The worse hearing is usually in the ear closer to the muzzle. Enquiry in these cases should be made into the type of weapons fired, for example, high or low velocity, whether or not fired from the shoulder, the frequency of use and number of rounds. Other weapons such as anti-tank weapons, rocket launchers, and mortars should also be enquired about. A history of tinnitus or temporary threshold shift immediately after firing, will indicate ears at risk. When patients are asked about weapons they will sometimes forget to mention sporting weapons including shotguns. Members of shooting teams are particularly at risk. It is useful to remember that some earplugs or earmuffs may not provide adequate ear protection against high velocity weapons.

Asymmetry of high frequency loss may be associated with *head injury*. Hearing loss is more probable in these cases if there is a history of unconsciousness following the injury. In general, the longer the period of unconsciousness, the greater the likelihood of a consequent hearing loss. Unconsciousness, bleeding or cerebrospinal fluid leak should be enquired about in deafness associated with *head injury*.

Hearing loss in late *Ménière's disease* may be bilateral and non-fluctuant. Speech discrimination will, at that stage, usually be significantly reduced. Hearing loss associated
with congenital or late syphilis is usually bilateral and the speech discrimination tends to be reduced and to fluctuate.

Bilateral acoustic neuromata are rare but must be kept in mind and the deafness may or may not be asymmetrical. Generalized neurofibromatosis or a positive family history increase suspicion of this condition.

Each of the causes of unilateral deafness may, of course, occur either bilaterally, or in combination with each other, or with any of the causes of bilateral deafness, resulting in a bilateral sensorineural deafness which is usually asymmetrical.

Unilateral sensorineural hearing loss

Unilateral hearing loss is frequently of sudden onset. Trauma including head injury, acoustic accident, blast injury, and damage at surgery, will come to mind early in consideration of unilateral hearing loss. There continue to be the sporadic cases of sudden hearing loss which are attributed to some interference with the cochlear blood supply, a viral infection, Reissner's membrane rupture, or perilymph leak. Some cases may, with careful questioning, be found to be due to childhood mumps, half-forgotten head injury, or possible perinatal causes. A significant number of cases will be seen for which no cause can be determined.

It is in the unilateral group that one is most commonly placed in the position of excluding an acoustic neuroma. Obviously it would not be practical to perform a computerized tomographic (CT) scan and evoked response audiometry on every patient with asymmetrical or unilateral deafness. Hence it is important to make, where possible, an accurate diagnosis of the pathology so as to limit the number of cases where expensive investigations are required.

Deafness with normal pure tone audiometry

Not infrequently, patients complain of hearing loss where an audiogram shows normal pure tone hearing and excellent speech discrimination. There probably are two categories of this syndrome. The first, and more common, could be called the 'auditory inferiority complex' group. These patients are insistent that they have difficulty in hearing which is usually more marked in background noise. Once they have decided that their hearing is impaired, they tend to blame their ears for hearing difficulties rather than the speaker or the background noise. The clinician will reassure these patients that they have 'normal hearing' and in most cases that is usually all that is needed. The reassurance often produces a dramatic improvement in their ability to hear.

The second group is one described by Pick and Evans (1983). They described patients who have abnormal difficulty in hearing in background noise despite having a normal pure tone audiogram. The problem is due to impairment of frequency resolution and the diagnosis depends on a 'comb-filtered noise test' which, at present, is not in general clinical use. They considered that the condition represents an early stage of hearing damage.
A diagnosis of neurosis in a patient should be made very reluctantly. Pressure on the central auditory pathways by a tumour has been known to cause severe reduction in speech discrimination in the presence of satisfactory or minimally reduced pure tone hearing loss. Exotica of this type are extremely rare but will arise from time to time so as to remind us of the dangers of attaching to patients the label 'neurotic'.

**Fluctuating hearing loss**

In taking a history of hearing loss it is important to ask specifically of the patient as to whether the hearing is fluctuant. Care must be taken in assessment of this symptom as occasionally the patient may be describing the variation in ability to hear in quiet and noisy surroundings.

Fluctuating hearing loss usually results from a small number of clear-cut pathological entities. The most common of these is middle ear pressure change, resulting in minor degrees of conductive hearing loss.

The most common cause of fluctuation of inner ear function is endolymphatic hydrops which occurs in Ménière's disease and syphilitic labyrinthitis. Perilymph fistula is a rare cause of fluctuating inner ear function.

In Ménière's disease, the history of tinnitus and associated episodic rotatory vertigo will determine the 'true' Ménière's, but this leaves a number of cases of low frequency loss which may or may not have tinnitus or dizziness. These may be 'early' cases of Ménière's disease. The disease is usually unilateral in the early stages and speech discrimination is generally well preserved until the later stages.

In congenital or late syphilitic labyrinthitis, the hearing loss fluctuates and the associated vertigo is episodic and rotatory in the early stages. In the later stages with destruction of vestibular function, it becomes constant and is described simply as unsteadiness. A history in early life of treatment, usually injections, for 'eye trouble' (interstitial keratitis), will suggest the diagnosis. Wassermann and Kahn tests will frequently be negative and the fluorescent treponemal antibody absorption (FTA abs) test will be of most help. The loss is usually bilateral and asymmetrical. In established cases, speech discrimination tends to fluctuate more than the pure tone hearing.

Perilymph fistula presents a difficult diagnostic problem and is discussed in Chapter 7.

**Mixed sensorineural/conductive hearing loss**

Mixed sensorineural and conductive deafness presents a challenge to the physiological measurement technician and it is here that one most commonly encounters spurious audiometry. It is always important to ensure that the audiometric findings and clinical judgement are in agreement, especially if there is any question of surgery. A speech discrimination test (PBmax), with adequate masking of the other ear, will provide confirmation that the ear is serviceable or otherwise, and should not be overlooked.
The measurement of bone conduction is a rather artificial concept since it is not necessarily a true reflection of the function of the inner ear. It is well known that the middle ear makes a contribution to bone conduction and that correction of a middle ear conductive lesion causes an apparent improvement in inner ear function. The best known example is the Carhart notch in otosclerosis, but this may also be seen in chronic suppurative otitis media and secretory otitis media. The apparent inner ear hearing loss caused in this way may be reversible.

It has long been accepted that chronic suppurative otitis media is often accompanied by sensorineural hearing loss related to the chronic suppurative otitis media but not due to the effect of conductive deafness on bone conduction. Toxins, it has been said, have damaged the inner ear. Walby, Barrera and Schuknecht (1983) have confirmed, in a clinical study of 87 patients with unilateral uncomplicated chronic otitis media, that an abnormality of bone conduction does exist. However, in a study of 12 pairs of temporal bones with unilateral chronic otitis media, there was no evidence that the disease resulted in damage to the inner ear. They concluded that the sensorineural loss is due to altered mechanics of sound transmission.

There has been debate about the cause of the sensorineural deafness that is often seen with otosclerosis. Schuknecht (1974) has put forward good evidence that otosclerosis only rarely results in sensorineural loss in the absence of a conductive loss. He also has shown that there is no consistent histological explanation for the sensorineural loss found in ears with otosclerosis.

However, all the other causes of sensorineural hearing loss also may occur in association with any of the conductive lesions. One cannot reasonably assume that inner ear function will necessarily improve with correction of the conductive component. Indeed, this carries the risk of surgical trauma with an increase in the sensorineural deafness.

In cases of industrial noise exposure, a concomitant conductive hearing loss may afford some protection to the inner ear. This is supported by unilateral cases of conductive hearing loss where inner ear function in the 'protected' ear is better than in the 'unprotected' one. However, not all investigations have confirmed this concept of protection for the inner ear by conductive deafness.

**Suspected malingering or feigned hearing loss**

One must always be aware of the possibility of non-organic hearing loss. This, in the main, will arise in the litigant and less frequently is psychogenic. Public awareness of excessive noise as a cause of hearing loss has resulted in increased interest in civil action in this respect. The individual concerned may occasionally succumb to the temptation to exaggerate the condition.

Suspicion will usually arise in the first few minutes of the interview with the patient. Not infrequently he will, in a rather obvious manner, fail to hear his name being called. In the initial stages of the interview each question may have to be repeated but will usually be heard on the second time, despite the clinician keeping the volume of his voice at the same level. Later, as the interview comes to include questions which the patient feels are important
to his case, such as enquiries designed to confirm the absence of other causes, he will tend to hear on the first occasion!

In psychogenic cases, suspicion may also arise when the patient appears to have a relative lack of concern about an apparently severe hearing loss.

At audiometry, attempts are made to exaggerate the audiometric results. Suspicion of this will arise when the recorded hearing levels are inconsistent with his ability to hear the spoken word.

**Sudden hearing loss**

Sudden hearing loss presents a therapeutic dilemma. There are two schools of thought, broadly represented by nihilism and those who advocate simultaneous multiple drug therapy. The fact that early treatment appears to achieve better hearing need only mean that there is a high rate of spontaneous recovery and that the inclusion of early cases boosts the results! Controlled trials have failed to produce convincing evidence of success and often have produced conflicting conclusions.

If one considers the potential aetiological factors in sudden deafness, then certain treatments, for example steroids, could aggravate the problem they were intended to help; the deafness of viral labyrinthitis may be exacerbated by steroids. For those with a compulsion to prescribe active treatment there is little to be said against bed-rest accompanied by carbon dioxide inhalations in an effort to improve cochlear blood flow.

In cases of acoustic incident a period of avoidance of noise exposure is advocated.

**Immune sensorineural deafness**

There is increasing interest in inner ear autoimmune disease as a cause of sensorineural hearing loss. There is no doubt that immune disorders can cause deafness which may reverse with steroid therapy. However, increasing numbers of alleged cases of immune sensorineural deafness are being reported. Many of these may be due to immune reactions but, so far, no clear pattern of clinical presentation has emerged and much more work will be required. Meanwhile one must guard against spurious claims and poorly controlled trials of expensive but dubious treatment regimens.

**General management**

There is no doubt that demands and expectations with regard to hearing vary from person to person. The person with a hearing loss will, in the main, have five concerns:

1. how bad is his hearing loss?
2. can it be reversed?
3. will it be progressive?
4. how will it affect his future?
5. what can be done to help cope with the handicap?
The extent of the deafness is clearly explained to the patient. He is frequently relieved to hear that he has not been imagining things and that his family have been justified in their complaints. The simple explanation that with a high tone loss one expects increased difficulties in noisy places is found by many patients to be reassuring.

The prognosis of the loss has then to be considered. Unfortunately, most sensorineural deafness is irreversible in our present knowledge, and this should be explained. Treatment is obviously required for certain conditions such as an acoustic neuroma or syphilitic deafness. Most cases will not improve. Indeed, although some may be static, most will progress slowly. Thus it has usually to be clearly explained that the loss will be permanent, but that any progression in the deterioration of hearing will be very gradual and that the patient will not become completely deaf.

Very occasionally young adults present with hearing problems where the prognosis will affect career choices. Care must be taken to avoid both the optimism that can leave him in a blind alley in middle life, and the pessimism that can put him there instantly. The question of the patient's future must be given serious consideration whether or not the matter is raised.

The adult audiology volume (Volume 2) contains chapters on rehabilitation and hearing aids in the management of sensorineural deafness. All that need be said here is that the main factors in success with a hearing aid are the ability to discriminate speech and the motivation to receive help from the aid. When both are poor the outlook is bleak. However, a positive approach from the otologist is of immense help. Constructive advice should be given on the selection and use of an aid, of avoiding if possible, communication in noisy places and of the importance of non-auditory clues. Finally, it is reassuring for the patient to know that almost everyone with deafness experiences frustration with himself and irritation from his family.