Assessing and treating adult patients with hearing loss

ABSTRACT

Hearing loss is a very common presentation both in primary and secondary care. In addition to causing significant morbidity, it can make communication with patients very challenging, and thus impact other aspects of their care. This article presents an approach to the assessment and management of this condition.

Almost 9 million people in the UK suffer from significant hearing impairment, with over 2 million people using hearing aids. Hearing loss has many causes, some of which can be curatively treated, and some only managed. It may cause significant social stigmata, and make communication almost impossible.

The majority of hearing loss is slowly progressive and seen in an elderly population. A thoughtful and considered consultation is required to delineate treatable aetiologies and help patients manage their hearing impairment. This article gives a guide to the hospital-based assessment, investigation and management of these patients.

Questions to cover in the history

Risk factors for hearing loss

Age is the principal risk factor for hearing loss, accounting for more than half the patients with this complaint. There is evidence that hearing begins to deteriorate at the beginning of the third decade (Robinson and Sutton, 1979), although it is unusual for this to become clinically evident before the fifth or sixth decade of life. Hearing loss usually continues progressively through the eighth and ninth decade (Pedersen et al, 1989). A number of mechanisms are thought to be involved including atrophy of sensory hair cells and neurons, homeostatic mechanisms within the cochlea, and thickening of the basilar membrane of the cochlea.

The second most important cause of hearing loss is noise exposure. Exposure to noise causes ‘temporary threshold shift’, or decreased hearing, that recovers after the noise exposure, usually in a period of hours to days. Prolonged or continual exposure leads to a permanent threshold shift or hearing loss, which is a product of the sound intensity, length of exposure and frequency of sound. The cochlea is most vulnerable to sounds at frequencies between 3 and 6 kHz (Mahendra Prashanth and Venugopalachar, 2011), and this is manifested in a characteristic diminished hearing around 4 kHz during hearing testing.

It is thought that noise may lead to approximately a third of cases of adult onset hearing loss (Kurmis and Apps, 2007). The European Union has published strict limits on occupational noise exposure (European Agency for Health and Safety at Work, 2003). This gives an upper noise limit of 87 dB, although the exposure action values are 80 dB and 85 dB (lower and upper limits respectively), and these values exclude the protective effect of noise protectors. Nevertheless, occupation remains the principal cause of noise-induced hearing loss. Occupations that have been implicated in noise-induced hearing loss include both classical and contemporary musicians, machinists and assemblers, and agricultural workers. Outside the workplace, recreational shooting and woodworking have been associated with hearing loss. Also, those living near airports have been shown to have higher hearing thresholds than controls (Chen et al, 1997). Recreational music exposure is controversial, with no clear consensus on whether it is a significant cause or not (Zhao et al, 2010).

Past medical history and medications

Ototoxic medications are another important cause to consider – 30% of patients presenting with hearing loss are taking potentially ototoxic medications (Lim and Stephens, 1991), although it is difficult to establish causality in these patients, as they frequently have other causes for hearing loss such as increased age. Ototoxic medications include aminoglycosides, furosemide, chemotherapeutic agents and salicylates. These drugs may cause reversible hearing loss (in the case of salicylates) or irreversible hearing loss (with drugs such as gentamicin). For this reason it is important to establish whether there has been prior, as well as current, exposure to these medications.

Past medical history is important, notably if there is a history of ear complaints or operations on the ear. Autoimmune conditions, such as systemic lupus erythematosus, can cause hearing loss. Furthermore, cardiovascular disease is associated with hearing loss (Agrawal et al, 2009).

Onset of hearing loss after meningitis should be rapidly addressed. The inner ear is continuous with CSF, and meningitis may lead to inflammation, fibrosis and ossification in the cochlea. Early intervention can be crucial to maintain options for hearing rehabilitation.

Mr Robert Almeyda, Ear, Nose and Throat Consultant, Royal Berkshire Hospital, Reading, Berkshire

Mr Robert Nash, Consultant Paediatric Otolaryngologist and Cochlear Implant Surgeon, Great Ormond Street Hospital, London WC1N 3JH

Correspondence to: Mr R Nash (mr.robert.nash@gmail.com)

Epidemiological associations include male sex, white race (Helzner et al, 2005), smoking (Nakanishi et al, 2000), and obesity (Fransen et al, 2008).

The nature of hearing loss and associated ear symptoms

The onset and laterality of the hearing loss may provide information about possible causes. Unilateral hearing loss can suggest a localized, treatable aetiology for hearing loss. Bilateral hearing loss may be more disabling.

Sudden onset hearing loss can be important to address. In the context of an upper respiratory tract infection, it is more likely to represent middle ear effusion. In the context of head injury, it may indicate temporal bone fracture. Sudden sensorineural hearing loss requires treatment with oral steroid medication. The timing of the hearing loss in relation to ototoxic medication or noise exposure may give an indication to the aetiology, and may be relevant should the patient seek to make a claim for damages. There is a recognized condition known as 'acoustic shock', in which exposure to a single short duration, high frequency, high intensity and unanticipated noise via a telephone headset may produce decreased hearing, otalgia, tinnitus, aural fullness and anxiety or depression (McFerran and Baguley, 2007).

Symptoms such as otorrhoea are likely to indicate middle ear pathology. Vertigo may indicate an inner ear or central (intracranial) illness. Tinnitus, if unilateral or pulsatile, may require investigation, but is otherwise non-specific and frequently seen in hearing loss.

Points to cover in the examination

Free field vocal testing

Free field vocal testing is performed by asking the patient to repeat combinations of numbers and letters that are in turn whispered and spoken at arm's length (approx 60 cm), and close to the ear (approx 15 cm) (*Figure 1*). The tragus is blocked in the non-test ear. If a whisper can be heard at arms length, the chance of there being a significant clinical hearing loss is very low (Bagai et al, 2006). This is useful as a quick, simple screening examination.

Otoscopy

Otoscopy (*Figure 2*) can identify a number of treatable causes of hearing loss. Wax can cause hearing loss if impacted. Other occlusions of the external auditory meatus, such as stenosis or false fundus formation, may also be visualized. The tympanic membrane should also be examined. This may demonstrate pathologies such as chronic otitis media, glue ear or perforation.

Tuning fork tests

The most commonly used tuning fork tests, Rinne and Weber, work together to identify conductive and sensorineural hearing loss. The Rinne test is performed by asking a patient to say whether a tuning fork is louder when held next to the ear, or when placed onto the mastoid process. The Weber test asks a patient to say in which (if any) ear a tuning fork placed on the forehead is heard (*Figure 3*). Patients with conductive hearing losses (which

Figure 1. Free field testing. A voice ranging from a whisper to a shout is presented to the patient **(a)** at arm's length (approx 60 cm), and **(b)** near to the ear (approx 15 cm). Bisyllabic words or numbers are used and the patient must repeat them. A hand covers the mouth to prevent the patient from lipreading. If a whisper can be heard at 60 cm then the patient's hearing thresholds will be better than 15 dB, if a shout at 60 cm is not audible, then the threshold will be worse than 75 dB.

Figure 2. Otoscopy is performed with an appropriate sized (>4 mm) speculum. **a.** The pinna is held slightly posteriorly and slightly superiorly to straighten the ear canal, and the otoscope is introduced into the ear. The little finger of the hand holding the otoscope lies against the patient's cheek for stability. **b.** All four quadrants of the tympanic membrane are visualized.

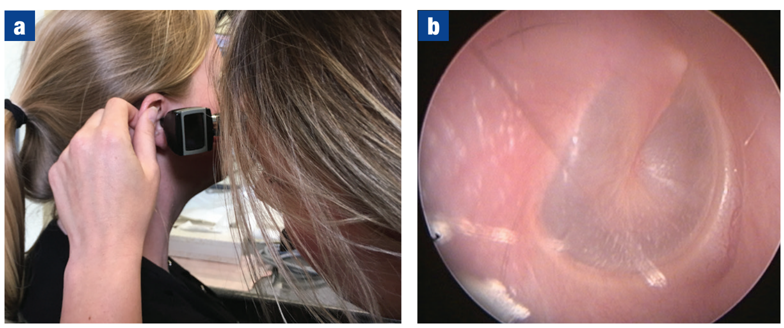


Figure 3. Tuning fork tests. **a.** A 512 Hz tuning fork is struck onto the clinician's knee, and then placed on the forehead of the patient. The patient is asked which ear the sound is loudest in (Weber test). **b.** The fork is struck again and presented next to the ear, and then **(c)** pressed against the mastoid process behind the ear. The patient is asked whether the sound is loudest when the fork is next to the ear or when pressed to the bone behind the ear (Rinne test). The Rinne test is considered positive when the sound is louder through air – this is a normal result. When sound is louder through bone it is either the result of a conductive hearing loss in that ear, or of a complete sensorineural hearing loss – sound presented to bone may travel across the skull and be heard by the other ear in these cases. The Weber test is used to distinguish these two situations (when there is a conductive loss the Weber test would be loudest in the tested ear, and when there is a sensorineural loss it would be louder in the other ear).

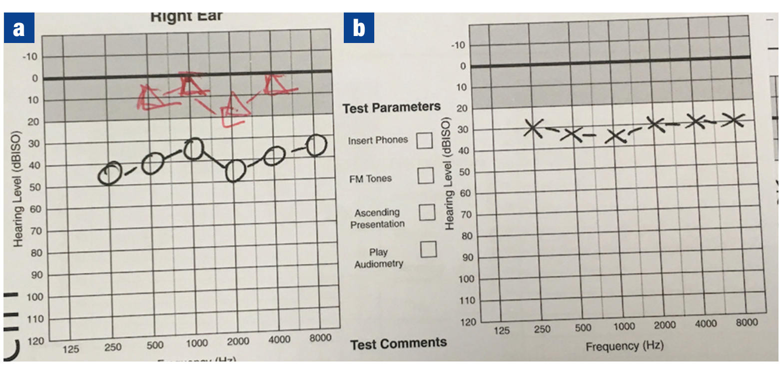
are usually seen in middle or external ear pathologies) often report better hearing when sound is presented through bone (the mastoid process) than air. Indeed, the side with a conductive hearing loss usually hears a sound presented through bone in the midline (Weber test) more loudly than the other side.

In adults an air–bone gap of 25 dB (a mild conductive hearing loss) is detected in 87% of patients (Browning and Swan, 1988). Tuning forks can also be used to test for suspected non-organic unilateral hearing loss (Durmaz et al, 2009).

Cranial nerves

Not only does the facial nerve run through the middle ear, but it also exits the brainstem with the vestibulocochlear nerve at the cerebellopontine angle. Facial nerve weakness in the context of hearing loss is highly significant, and suggests pathologies such as cholesteatoma or acoustic neuroma.

Figure 4. Pure tone audiograms. Pure tones are presented to the patient in either ear (with the other ear 'masked' if necessary). Tones are presented at volumes standardized to 'normal' hearing, and can be presented through air and through bone. **a.** In the right ear, there is a conductive hearing loss – with sound heard normally when presented through bone (the red marks), and only sounds of around 40 dB heard though air. **b.** The left ear has a sensorineural hearing loss, as the quietest sounds that can be heard are around 30 dB HL (a mild hearing loss). (0–20 dB is considered normal, 21–40 dB is a mild hearing loss, 41–70dB is a moderate hearing loss, 71–90 dB is a severe hearing loss and >91 dB is a profound hearing loss.)



What investigations are appropriate?

Audiometry

A pure tone audiogram can be performed by audiologists in sound-proofed booths, and demonstrates a patient's capacity to detect tonal sounds presented at differing frequencies, relative to a population average. It presents these sounds using bone and air conduction, allowing conductive and sensorineural components of a hearing loss to be delineated. High thresholds indicate worse hearing, and are generally shown lower down a graph plotting hearing thresholds (*Figure 4*).

The pure tone audiogram is subjective – it relies on a patient's responses – but it is reliable and reproducible (Lemkens et al, 2002), and correlate well with the purely objective electrocochleography (Schoonhoven et al, 1996). Generally, the pure tone audiogram correlates well with functional hearing deficit, although there is a population of patients suffering from a significant functional hearing deficit, but who have a normal pure tone audiogram (Middelweerd et al, 1990).

Tympanograms

Tympanograms demonstrate resonance of the tympanic membrane at different pressures (*Figure 5*), and calculate the volume of the ear canal. They can therefore be used to help diagnose pathologies like glue ear or tympanic membrane perforation.

Cross-sectional imaging

A computed tomography scan is used to investigate the structural components of the middle ear. This is relevant to

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Figure 5. Tympanograms measure the ability of the ear drum to resonate at different pressures. **a.** Result from the left ear is normal – 'type A'. **b.** Result from the right ear is flat – 'type B'. The ear canal volume (ECV) is also measured, and is 5.9 cm^3. This is much greater than the volume of an ear canal indicating a perforation in the ear drum. When a normal ECV is seen (as in the left ear – 1.2 cm^3) the most likely cause of a flat trace would be middle ear fluid.

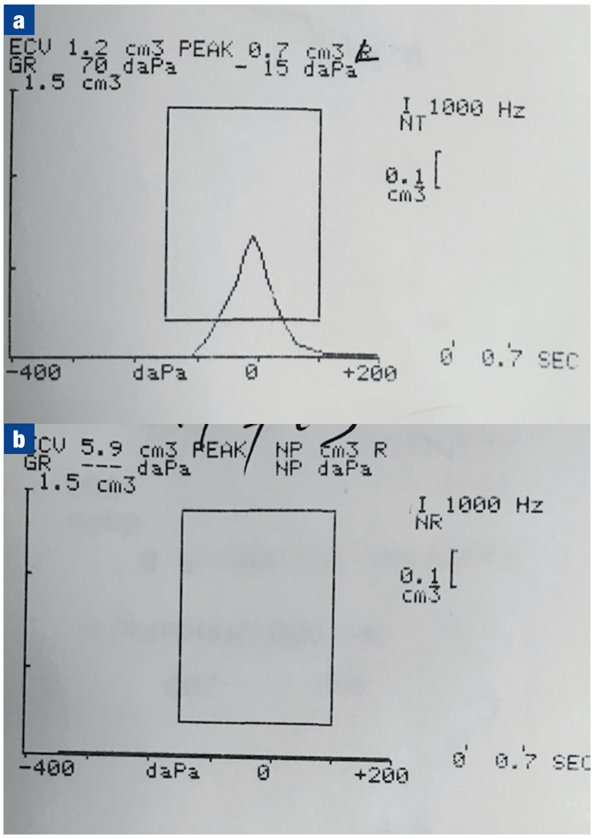


Figure 6. Coronal computed tomography scans of two right ears. **a.** The scan shows an aerated, normal ear with no signs of bone erosion. **b.** Soft tissue has accumulated in the middle ear, and there are signs of erosion of the ear canal wall and the ossicles, consistent with cholesteatoma.

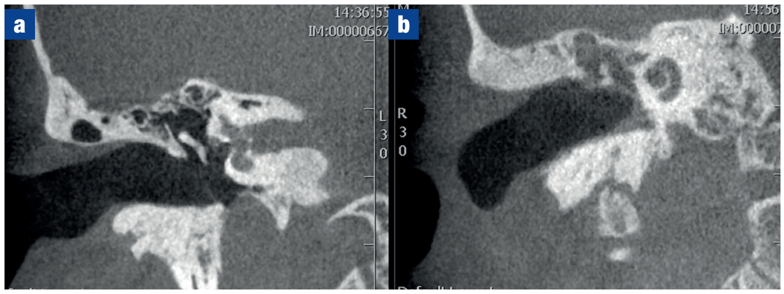
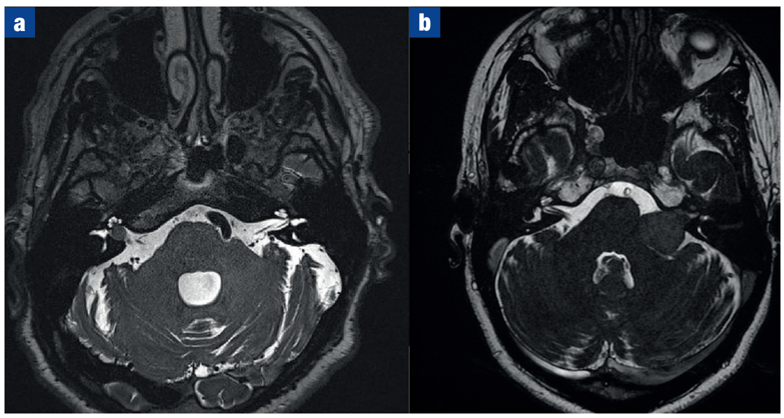


Figure 7. T2 weighted axial magnetic resonance imaging scans used to investigate asymmetrical audiovestibular symptoms. These can be caused by vestibular schwannomas – benign tumours growing on parts of the vestibulocochlear nerve. **a.** A small vestibular schwannoma, which might be left untreated and observed. **b.** A much larger tumour, pressing into the cerebellopontine angle. This tumour is likely to require treatment.



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conductive hearing loss when glue ear and uncomplicated perforations have been excluded, particularly when surgery is considered. It also plays an important role in the assessment of cholesteatoma – and can demonstrate the extent of bony erosion and complications caused by the disease (*Figure 6*).

The imaging modality of choice for unilateral or asymmetrical sensorineural hearing loss is magnetic resonance imaging of the internal auditory meatus. There are a number of published criteria for selecting those patients with asymmetrical symptoms, for example an interaural difference in hearing threshold of 15 dB in unilateral hearing loss, and 20 dB in bilateral hearing loss (Obholzer et al, 2004). It is worth noting that retrospective case series suggest that when magnetic resonance imaging of the internal auditory meatus is performed, vestibular schwannomas are rarely seen (around 1%) (*Figure 7*), and incidental findings of other intracranial pathologies are much more common (around 20–30% of cases) (Chisholm et al, 2006).

Management plans

Lifestyle changes

Exposure to noise should be reduced where possible. Simple techniques such as basic lip reading, asking people to speak clearly and slowly, and standing close with the better hearing ear towards the speaker can increase speech comprehension (Woodhouse et al, 2009). For some patients, a formal programme of aural rehabilitation may be useful (Boothroyd, 2007).

Medications

The decision to withdraw potentially ototoxic medications should be made cautiously, as it is likely that only a proportion of these patients' hearing will be significantly affected by these potentially life-saving medications.

Hearing aids

Hearing aids have consistently been shown to improve quality of life. Digital hearing aids may be programmed to fit an individual's hearing loss. A review by the American Academy of Audiology concluded that hearing aids improve quality of life by decreasing the psychological, social and emotional sequelae of sensorineural hearing loss

KEY POINTS

- Hearing loss is a frequently encountered condition.
- The history should help the clinician to identify those patients who may have a preventable deterioration in hearing.
- The aim of the examination should be to identify patients with a treatable cause of hearing loss.
- Having assessed an adult patient with progressive hearing loss, it is appropriate to arrange investigations including an audiogram.
- For the majority of patients with hearing loss, hearing aids will be beneficial.

(Chisolm et al, 2007). Furthermore, hearing aid fitting improves the quality of life of the partner of the patient with hearing loss.

Bone anchored hearing aid

The use of bone anchored hearing aids is recognized in patients with single-sided deafness and in cases where a conventional hearing aid may not be used (e.g. persistently discharging ears). Sound is conducted through bone directly to the cochlea through an abutment osseointegrated into the temporal bone. Sound can easily pass to the contralateral side, providing a method of treating unilateral profound hearing loss.

Implantable middle ear devices

Fully or semi-implantable middle ear implants are relatively new devices that transduce sound into the middle ear. Their safety and efficacy has been demonstrated, but they are significantly more expensive than conventional hearing aids, and may not provide significant audiological advantages (Verhaegen et al, 2008). The NHS recommends their use in cases where conventional hearing aids are not suitable (e.g. patients with recurrent otitis externa) (NHS Commissioning Board, 2013).

Figure 8. A cochlear implant – this works in a similar fashion to a normal hearing aid, but requires a stimulator receiver package to be implanted beneath the skin with an electrode that goes to the cochlea. Sound is detected by the hearing aid part worn around the ear, converted by the processor, and sent through the transmitter – held to the inner part with a magnet – to the internal part. This internal package sends these electrical signals directly to the spiral ganglion of the cochlear nerve.

Cochlear implants

Cochlear implantation is available for patients with severe to profound hearing loss that can not be adequately treated with hearing aids. By inserting an electrode into the cochlea, the spiral ganglion and consequently cochlea nerve can be directly stimulated by the cochlear implant package. A processor is worn over the ear which detects sound and transmits it to the electrode via an implanted stimulator receiver package (*Figure 8*).

In the UK, the National Institute for Health and Care Excellence (2009) has approved unilateral cochlear implants in adults with severe, profound hearing loss who do not receive adequate benefit from conventional hearing aids. Adequate benefit is defined as a score of 50% or greater in the Bamford–Kowal–Bench sentence test at 70 dB.

There is a great deal of evidence for improvement in functional hearing, quality of life and depression with cochlear implantation (Eshraghi et al, 2009). However, cochlear implants are more effective for speech perception than music appreciation – even in patients referred for cochlear implantation, music perception is better with conventional hearing aids (Looi et al, 2008).

Conclusions

Using a structured approach, patients with hearing loss can be effectively and efficiently managed. The majority of these patients will be best managed with a hearing aid, although a proportion will benefit from surgical intervention. **BJHM**

Conflict of interest: none.

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