Key Questions - Hearing Problems and cochlear implantation

Black print – Mr Harry R F Powell, ENT SpR, Royal National Throat, Nose and Ear Hospital, London.

Question 4 and 8 written by: Dr Rachel Booth, Principal Clinical Scientist, Head of Paediatric Audiology, Central Manchester University Hospitals NHS Foundation Trust

Question 6,7 and 9 written by: Ms Deborah Mawman, Audiologist/Hearing Therapist, Coordinator of Adult Cochlear Implant Team, Central Manchester University Hospitals NHS Foundation Trust

1. What proportion of the population develop hearing problems as they get older? What is the usual age of onset of presbyacusis? How might we identify those patients who would benefit from amplification?

Our hearing is at it's best in the late teenage years and early twenties, from then on there is progressive decline. The prevalence of hearing impairment is about 53% of the population between 71-80 years of age. Ultimately we would all experience some age related hearing loss if we lived long enough. The age of onset of a typical sloping sensorineural loss is incredibly variable and it is likely to be influenced by accelerated ageing and some form of hereditary/ genetic degenerative process.
Patients describe difficulty hearing conversation, particularly when there is background noise. They may have to ask others to repeat themselves or watch television with the volume louder than necessary for those with normal hearing. The onset is typically slow and insidious. The symptoms are often a problem for other members of the family/household.
Assessment for any patient who complains of hearing loss consists of an otological history and examination. If there are no suspicious features and normal tympanic membranes then some audiology services will accept direct referral for pure tone audiometry and consideration of hearing aids. If there is any doubt then a referral to the local ENT service is appropriate for specialist review.

2. A) The use of cochlear implants seems to be becoming more widespread, even for adult patients. How effective are they? I always assumed that they would be more beneficial for children but is that not the case?

In the UK the first single channel cochlear implant was inserted at UCL in 1984. Cochlear implantation has become established over the last 15 years. The device itself is very expensive (about £20,000) and for this reason cochlear implantation surgery is the most expensive operation done in the NHS. Consequently the PCTs were reticent for quite some time about providing funding for patients to have an implant. Following publication of NICE guidelines on cochlear implantation in January 2009 it has become more widely accepted.

Deafness can be subdivided in to pre-lingual and post-lingual depending on the age of onset of deafness and whether the patient has developed speech prior to becoming deaf. Initially it was thought that patients with post lingual deafness would gain more benefit from implantation, as they knew what it was like to have some hearing; therefore more adults were implanted in the early days. With advances in research and understanding of the aural pathways it has been established that the brain has incredible neural plasticity and that binaural hearing (with 2 ears) confers significant advantages. Hence pre-lingually deaf infants that meet implantation criteria now receive bilateral implants as early as possible.
Any patient with severe to profound sensorineural hearing loss who does not gain adequate benefit from hearing aids may be a candidate for cochlear implantation. A cochlear implant does not restore normal hearing levels but provides access to sound, enabling patients to acquire and understand spoken language and speak intelligibly.

2. B) What problems are associated with the use of cochlear implants?

The process of cochlear implantation is very involved. The assessment for suitability requires a number of visits to hospital and includes various audiological tests including speech discrimination. Both CT and MRI scanning is undertaken to check the cochlear anatomy and presence of auditory nerves. Patients *and parents* need to be psychologically suitable. There is a team of surgeons, audiologists and rehabilitation professionals who are all involved in the selection process.
Patients with an implant are at an increased risk of pneumococcal meningitis and therefore all patients should receive a pneumococcal vaccination prior to surgery.
There are inherent risks of surgery and general anaesthetic. There are intra-operative risks including damage to the facial nerve and the post-operative risk of infection.
The post-operative outpatient care including switch on and monitoring is intensive and requires an ongoing commitment to attend regular appointments to help patients to get maximum benefit from their implant(s) and check their progress.
Hearing with a cochlear implant will never be the same as a normal ear but the way the brain is able to interpret the sounds presented to it enable patients to lead a normal life, where those with best results can enjoy music and manage well in background noise.
We do not know what technologies will be available in the future for patients with severe to profound deafness but currently cochlear implantation is the best available method of stimulation of the auditory pathway for these patients.

3. What is the association between tinnitus and hearing problems? Does improving someone's hearing help with the management of their tinnitus or not?

Tinnitus is a symptom of many different diseases, the vast majority of which involve the ear. The prevalence of tinnitus has been reported variably in the literature. The most common pathologies associated with tinnitus are acoustic trauma leading to noise induced sensorineural hearing loss, Menière's disease and presbyacusis (see table 1). The prevalence of tinnitus in patients with normal hearing is 15-35%. The figures vary considerably when looking at patients with severe to profound hearing loss awaiting cochlear implantation, from 27% to 81% reporting tinnitus.
It has been reported that psychological issues, anxiety and stress can lead to the onset or exacerbation of tinnitus and it is important to take a sympathetic and attentive approach when managing these patients. There are no controlled trials of tinnitus therapies. According to some reports reassurance, education and counseling are the most important management strategies. Hearing aids are the 1st line treatment for patients with tinnitus and hearing loss, the rationale being that amplification of external sounds may reduce awareness of tinnitus. The efficacy as a treatment for the tinnitus varies between individuals.

Condition	Prevalence of Tinnitus
Sudden hearing loss	50%
Acute noise trauma	100%
Chronic noise induced H loss	50-90%
Vestibular Schwannoma	70%
Menière's attack	100%
Presbyacusis	70%
Normal hearing	15-35%

Table 1. Prevalence figures in common conditions associated with tinnitus.

4. What would make you suspicious that a baby has hearing difficulties? What is the current screening programme for newborns – is it still just for those judged to be at high risk of problems?

It is difficult to tell whether a baby has hearing difficulties by observation or informal testing in your surgery. Babies and young children can appear to startle and react to sounds even if they have significant hearing problems that will impact on their ability to learn to talk. You should always be concerned if the parents are.

ALL babies are offered a hearing screen shortly after birth. Useful information for professionals about the screening programme is available at http://hearing.screening.nhs.uk.

In most areas the screen is performed in the hospital before the baby is discharged although this can be difficult with early discharges from maternity units. If the screen

cannot be done in hospital, an outpatient appointment is arranged. Babies who do not pass the hearing screen are at high risk of hearing loss and are referred to the Audiology department for diagnostic testing.

Unfortunately some families do not attend appointments, even when their baby has not passed the screen. You should be informed when this happens and we would ask you to discuss the importance of the screen with the family. Services are keen for you to refer babies back in after you have spoken to the family.

5. How useful are Weber's and Rinnes' tests in clinical practice? Are they still used routinely?

The tuning forks are used to distinguish between a conductive and a sensorineural hearing loss but they have low sensitivity and specificity. Either a 256Hz or a 512Hz fork should be used, (forks with higher frequencies may not be heard by those with hearing impairment and with a 128Hz fork it is difficult to distinguish between hearing it and feeling the vibration).
The tuning fork should be struck 1/3 of the distance from its tip against a firm but elastic object (elbow) to produce a relatively pure tone. In the Rinne test the fork should be held 2-3cm from the external meatus with its acoustic axis in line with the EAC (See figure 1). The tests are based on two principles:

a) The inner ear is normally twice as sensitive to sound conducted by air rather than bone.
b) In a purely conductive hearing loss the ear is subject to less environmental sound and is therefore more sensitive to bone conduction.

In general practice the tests can be a simple and quick way to establish the type of hearing loss. We routinely use the Weber test post-operatively. In the ENT outpatient setting pure tone audiometry is used to make the distinction between conductive and sensorineural loss and quantify their magnitude.

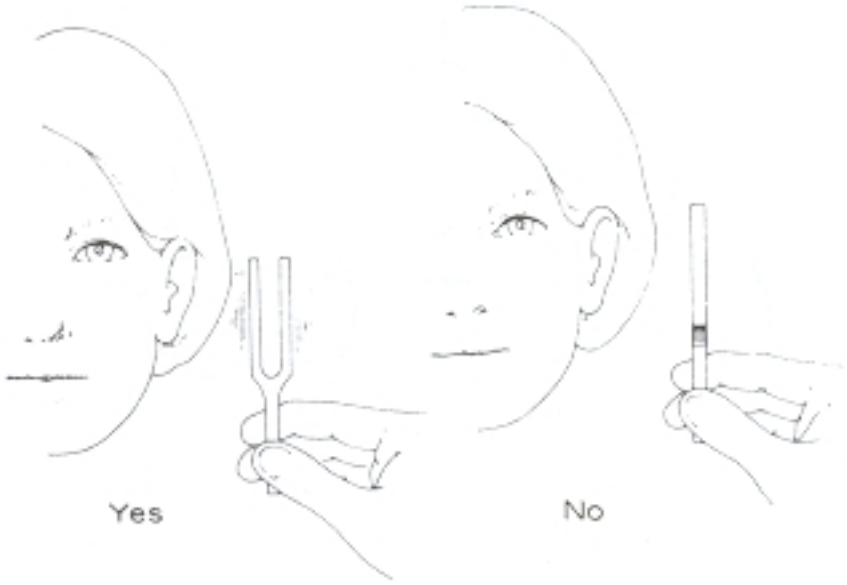

Figure 1.

6. Modern hearing aids, particularly those provided by private providers, seem to be getting much smaller. This often means that they have no T setting for use with induction loops. Does this matter? Are there particular patients who particularly benefit from the use of induction loops? How do they work?

Induction loop systems help people with all degrees of hearing loss who wear a hearing aid or cochlear implant to hear sounds more clearly by reducing the effect of background noise. The loop system has to be used together with a hearing aid or a cochlear implant switched on to the "T" setting. Some of the smaller hearing aids like the In The Ear (ITE) or In The Canal (ITC) hearing aids may not have a T setting because they are too small to house the induction pick up coil. Some patients prefer to trade off the benefit of improved hearing in noise for cosmetic reasons.

The induction loop is a cable that goes around the listening area, for example in the TV room at home or in a public area such as a theatre, cinema, place of worship, meeting room, conference hall, lecture room, airport, bank, shopping centre, and bus or train station. It works by transmitting a sound signal in the form of a magnetic field, which is picked up by the T setting on the hearing aid. No matter how much background noise there is the hearing aid or implant user will hear the speech signal through the loop.

7. What communication strategies should we, as GPs, use to maximise the benefit of consultations with patients with hearing difficulties? What should we avoid?

It's important to remember that even if someone is wearing a hearing aid it doesn't mean they will hear everything you say clearly. Ask if they need to lip-read or even if they would prefer you to type out what you are saying on your PC.

Make sure you have face-to-face or contact with the hearing impaired patient and that you talk directly to them and not to their interpreter or significant other. Always make sure the patient is looking at you before you start to speak and that you have their attention throughout their consultation.

Speak clearly but not too slowly and don't shout! Shouting can be uncomfortable for a hearing aid user and it can make you look aggressive. Don't exaggerate your lip movements it's much easier to follow what someone is saying if they are using natural facial expressions and gestures. If the patient doesn't understand what you've said, don't just keep repeating the same thing over and over. Try saying the same thing it in a different way.

Make sure your consultation room has good lighting and that your face is towards the window. The room should be away from noise and distractions. Finally try to use plain language and avoid jargon or difficult medical terminology. If necessary write things down if the patient is unable to understand what you're saying.

8. How reliable are the childhood screening tests for hearing problems? Should we be referring all children for whom there is parental concern? At what age can children reliably undertake an audiogram?

Childhood screening tests occur at various intervals and the tests used depend on the age of the child, their intellect and motor ability.
Neonatal: Otoacoustic emissions.
7 months: Distraction techniques.
2-4 years: Visual reinforcement audiometry or conditioned reflex audiology.
5 years and over: Pure tone audiogram.

Reliability increases with selection of an appropriate test and a co-operative child.
As with all consultations there is an element of clinical judgment when deciding whether a child needs referral for further assessment. Any parental concern should be taken seriously and a decision made after thorough history and examination. Referral to the local community audiology service should enable problems to be flagged up so that further investigations and specialist opinion can be saught as required.

Hearing screening is very reliable at identifying children who need further testing and who may have a hearing loss. The newborn screen will only identify a problem present at birth and aims to identify problems that will affect development of spoken language. Children with milder hearing problems may pass the screen but development can still be affected. Some children can develop permanent hearing loss later on and there is also hearing loss due to persistent glue ear.

You should always refer children for hearing assessment if the parents are concerned, you may still want to refer if they're not concerned! Often when directly asked parents will say they are not concerned but will comment 'they ignore me' or

'their speech isn't clear'. This can be because quieter sounds are not heard and can indicate a possible milder hearing loss which still requires help.

The best place to refer for a hearing assessment is to your local community audiology service –ENT is typically not the most appropriate place for initial assessment. A reliable hearing test can be performed at any age. There are different tests depending on the development of the child and all provide accurate information about the hearing.
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9. What sort of level of hearing loss means that patients might benefit from specialist adaptations such as text relay when using the phone? How would patients find out about these and other services available to them?

Generally patients who are unable to have an easy interactive conversation on the telephone will have a severe hearing loss or worse. These patients normally have difficulty or are unable to follow conversations in a quiet room with one other person if they are unable to see the speakers face. A textphone is a telephone device that has a keyboard and a display screen. It is used to communicate text in real-time and the user types what they want to say rather than speaking into a mouthpiece. Textphones connect to the telephone network. In the UK, companies often use the word 'Minicom' rather than textphone. In fact, Minicom is a widely used brand of textphone.

Text phones and other specialist adaptations for hearing loss can be purchased from the RNID or social services may help to pay for or provide equipment. Information about text phones and other specialist adaptations for hearing loss can be found on the RNID website at www.rnid.org.uk or by contacting the social worker for deaf adults at the local social services department. Information about text phones can also be obtained from:

Text Relay Customer Service Team
PO Box 284, Liverpool L69 3UZ
Telephone 0800 7311 888 Textphone 18001 0800 500 888

10. We sometimes hear comments that the use of personal electronic devices, such as MP3 players has increased the incidence of hearing problems later in life. Is this true? How does exposure to excessive noise cause hearing problems? Are there other possible aetiologies for hearing loss that we could be anticipating and educating patients about as a preventative measure?

Noise exposure is not only the main avoidable cause of hearing loss both recreationally and occupationally but also a major cause of tinnitus. Listening to personal music players with excessive volume can cause long-term damage to the cochlear. Power tools, shooting and nightclubbing also expose people to excess noise (See figure 2). There is no evidence

that listening to personal music players at reasonable volume levels leads to hearing loss later in life.
Noise induced hearing damage is related to the volume and duration of exposure. Occupationally the safe exposure limit is 85 decibels for 8 hours a day. Biological variability means that individuals are susceptible to damage at different noise levels. Generally noise louder than 90 decibels will cause pathological changes. Permanent mechanical and metabolic damage will occur first to the outer hair cells then the inner hair cells, and subsequently the supporting pillar cells and stria vascularis depending on duration and intensity. Noise damage will show on an audiogram as a dip at 4-6KHz. Damage to the cochlear is cumulative, therefore adequate protective measures and avoidance of further noise exposure is crucial.

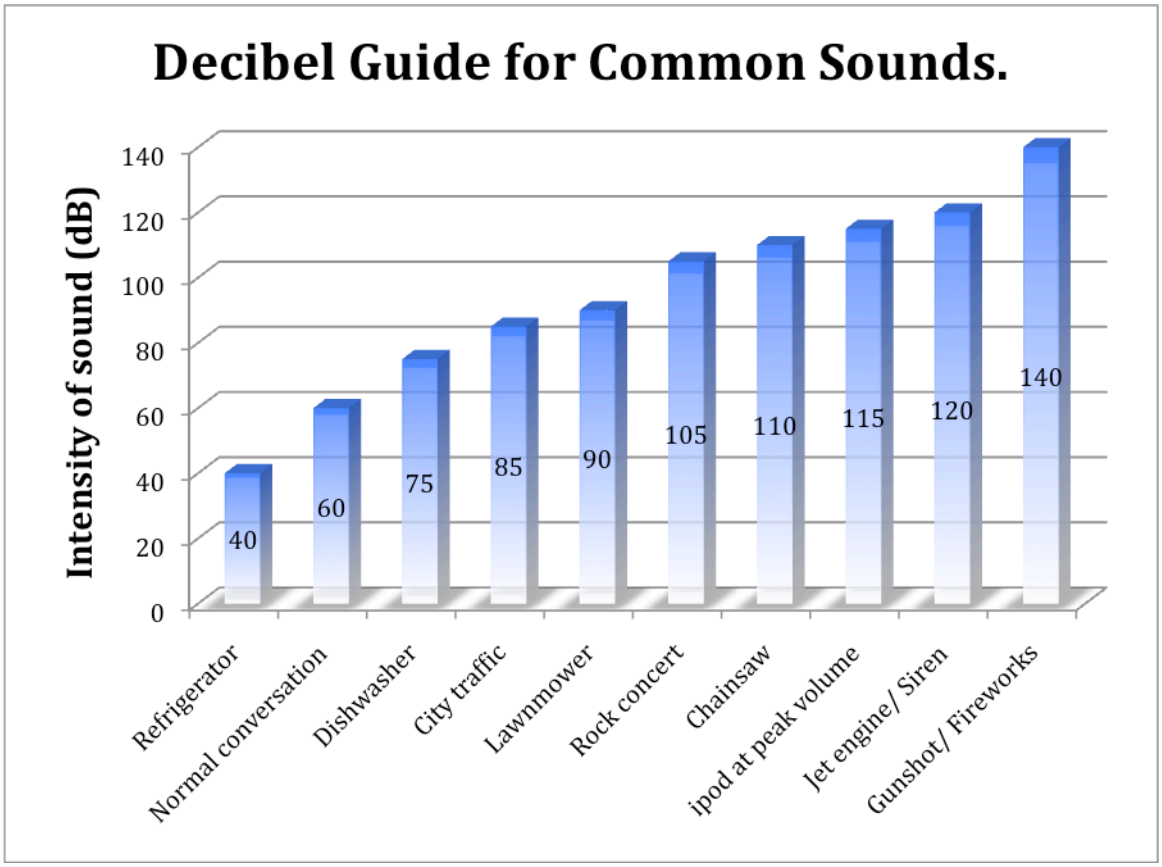

Figure 2.