HEARING IMPAIRMENT

Introduction

Hearing impairment is a relative term, ranging from a mild loss of hearing to complete deafness. Hearing loss has a profound effect on the human being. Total deafness isolates the individual from this environment. Even mild hearing loss may severely impacts a child's development. Hearing impairment may occur at any age, from the neonate to the geriatric patient.

Hearing loss in the elderly

In our setting, the commonest cause is presbycusis which translates to hearing loss due to age. All people will have worsening of hearing thresholds as they get older. In general, most people will lose on average 1 dB / year after the age of 35 years. The approach is therefore to do an ENT examination, tuning fork tests, and to exclude other simple, reversible causes.

If your patient complains of progressive bilateral hearing loss usually with constant tinnitus, your ENT examination is normal and Rinne tests are positive, you have confirmed the diagnosis of presbycusis. Typically, one can differentiate the patient who continues to struggle despite an increase in the loudness versus the patient who struggles to hear and then says don't speak so load. The former group is due to retro-cochlear loss, and they would typically have poor speech discrimination. The latter group is due to cochlear loss and the complaint is known as recruitment.

Remember that the three inner ear symptoms are hearing loss, tinnitus, and vertigo. The presence of all three should raise the suspicion of a more sinister underlying problem, especially when the symptoms are unilateral, the tinnitus pulsatile in nature, and there is presence of vertigo. These patients should be referred to an ENT specialist.

Once the diagnosis of presbycusis is established, provide the patient counselling and refer for a hearing aid trial. It is important to establish a working relationship with your audiologist. Frequently, older patients are fitted with hearing aids, but they struggle to find benefit in real world situations. Rather have them pay for the ear mould and use a demo hearing aid for 2-4 weeks before committing to pay for them. Recently, older patients are also considered for cochlear implants, especially if their thresholds at 1 kHz exceed 70dB.

Other common causes are wax impaction and middle ear effusions. REMEMBER that in adults who present with a new onset middle ear disease / effusion, nasopharyngeal pathology should be excluded. This is usually done by an ENT specialist by performing a nasal endoscopy. Nasopharyngeal carcinoma typically presents with middle ear disease in an adult, as well as cervical lymphadenopathy.

Hearing loss in the child

The early diagnosis of hearing impairments in children is vital so that the resultant speech, communication, educational and psychological handicap can be minimised through amplification and intensive education. It is better to over-suspect hearing loss in a child rather than to miss the diagnosis. The testing of hearing is very difficult in young children and increasing reliance is being placed on Otoacoustic Emissions (OAEs) and Auditory Brainstem Response Audiometry (ABR), which are objective tests.

The capacity of the human brain to learn to interpret sound and speech (plasticity) is unique to the child below 6 years of age. If they are not exposed to sound before the age of 18-24 months, they can never learn to understand speech. If hearing loss is corrected before the age of 18 months, educational outcomes are the same as normal hearing children. However, if hearing remains reduced or not corrected at all, they will never catch up with normal hearing children. Cochlear implant outcomes correlate directly with the age of hearing correction, so much so

that implanting a completely deaf child after the age of 2-3 years will not lead to speech development. Therefore, hearing loss in a child, especially < 18 months, is an EMERGENCY and should be referred immediately! Ideally every child should undergo newborn hearing screening.

Approach to hearing loss – all ages

Milestones and history
Normal speech milestones are given in the table one below.

Table 1. Speech milestones.

Age	Task
0 – 4 months	Startled by sounds (even Moro reflex) Quietens when hearing sounds ("listen")
4 – 6 months	Turns to localise sounds once neck control is achieved
9 months	Listens Respond to name Mimics environmental sounds
12 months	Uses one word – girls a bit earlier, boys a bit latter
18 months	Babble rhythmically Understands instructions
24 months	Uses 20 words

When taking the history, always believe the mother (caretaker)! An easy acronym to assess risk factors for hearing loss in a young child is HEARING. It stands for;
H – Heredity
E – Ear abnormalities / syndromes
A – Asphyxia
R – Intra-venous or other ototoxic medications
I – Infections such as TORCH / Meningitis
N – Neonatal ICU (also ask about oxygen requirement and jaundice)
G – Growth. Babies below 1500 grams.

Older children might have other risk factors, and ones that influence the Eustachian tube are the most important. These include the adenoids, skull base deformities (Down syndrome), passive smoking, and structural problems with the muscles of the soft palate and Eustachian tube (cleft palates). Otherwise, ear problems are more common in boys, winter months, family history of ear disease, and primary and secondary immune diseases.

The doctor would need to think if it is congenital or acquired and whether it is sensori-neural, conductive, or mixed hearing loss. Congenital and acquired is discussed under aetiology below in more detail. When referring to a deafness as sensori-neural, conductive or mixed one can use the following approach:

- Sensori-neural
    - Cochlea
        - Outer hair cells / Inner hair cells / Synapse
        - Cochlear / Retro-cochlear
    - Nerve
        - Cochlear nerve
    - Brain
        - Cochlear nuclei / Olivary complex / Lateral lemniscus / Inferior colliculus / Medial geniculate body / Cortex (ECOLI mnemonic)
    - Auditory nerve spectrum disorder
        - Inner hair cells / Synapse / NVIII
    - Mechanical inner ear deafness
        - Acoustic neuroma

- Conductive
    - EAC
        - Obstructions / Wax / Exostosis / Etc
    - TM
        - Perforation / Stiffness / Anterior blunting / Myringo-sclerosis
    - Middle ear
        - Ossicles
            - Tympanosclerosis / Dislocation / Subluxation / Erosion / Fixation / Fracture
            - Congenital (Dysplasia, Hypoplasia, Aplasia)
            - Increased mass – Paget's disease / Osteopetrosis
        - ME cavity
            - Pressure / Fluid / Mass / Fibrosis / Atelectasis
        - Oval / Round window obliteration
            - Congenital / Otosclerosis / Tympanosclerosis
        - Third window
            - Superior SSC / Lateral SSC dehiscence / Disease associated (cholesteatoma / TB / Syphilis)
- Mixed

While taking a history, it is important to note factors mentioned in table two.

Table 2. Characteristics of hearing loss.

Characteristics		
Onset	Pre-lingual	
	Post-lingual	
Severity	Mild	21-40 dB
	Moderate	41-60 dB
	Moderately-severe	61-80 dB
	Severe	81-100 dB
	Profound	>100 dB
Frequency	Low	<500 Hz
	Middle	501-2000 Hz
	High	>2000 Hz
Pattern	Sloping	Typical pattern in presbycusis
	Rising	Meniere's
	Cookie-bite	Think congenital
	Notched	Noise induced hearing loss if at 4 kHz
	Saucer	
	Falling	Acute idiopathic nerve deafness
	Flat	Metabolic / Metabolic presbycusis
Clinical	Progressive / Acute idiopathic nerve deafness / Fluctuating / Stable	

Aetiology

A differential diagnosis for congenital hearing loss is given in table three. It will never be expected of you at GP level to known this and it only serves for reference value.

Table 3. Congenital hearing loss

Congenital hearing loss				
Genetic (50%)	Syndromic (30%)	Mono-gene	Autosomal dominant (AD)	Waardenburg Stickler NFII Treacher Collins
			Autosomal recessive (AR)	Pendred Usher Jervell
			X-linked	Alport Norries
			Other	Osteogenesis imperfecta Crouzon Apert
		Chromosomal		Down syndrome Turner syndrome
		Mitochondrial		
		Heterogeneous		Goldenhaar Klippel-Feil
	Non-syndromic (70%)	Mono-gene	AD (18%)	
			AR (80%)	Connexin 26 (50%) Connexin 30,31,43 Other
			X-linked (<2%)	Usually with Stapes fixation
			Other	
		Chromosomal		
		Mitochondrial		Aminoglycosides DM
		Heterogeneous		
Acquired (25%)	Pre-natal	TORCH – Toxoplasmosis, Rubella, Cytomegalovirus, Herpes		
	Natal	Asphyxia / Medication / Infections (meningitis) / NICU / Low birth weight / Jaundice		
	(Post-natal)	See later for full differential diagnosis		
Idiopathic (25%)				

A differential diagnosis for acquired hearing loss is given in table four. This can also be applied to adults.

Table 4. Acquired hearing loss.

Acquired hearing loss					
Infections	Viral – HZ / VZ / Measles / Mumps / CMV / EBV / Influenza / Rubella / Hep / HIV	Bacterial – Meningitis / AOM Strep / Staph / H Infl	Labyrinthitis – Viral / Via AOM	Other – Syphilis / Lyme / Rocky Mountain spotted fever / Rickettsia / Malaria / Parasites / Toxoplasmosis	
Granulomatous	TB / Histiocytosis / Fungal / Sarcoidosis / Syphilis / Wegener's (GPA)				
Auto immune	Cogan / PAN / Relapsing polychondritis / GPA / Primary inner ear AI disease / Para-neoplastic / Temporal arthritis / SLE / RA / Sarcoidosis / Scleroderma / HIV				
Vascular / Haematological	Emboli / Anaemia / Coagulation problems / Migraine / Vertebrobasilar occlusions / Waldenstrom / Cryoglobulinemia / Sickle cell / Leukaemia / Lymphoma / Loops				
Noise	Constant / NIHL / Blast trauma				
Neurological	MS / Friedreich ataxia / ALS /				
Medication	Aminoglycoside / Loop diuretics / Quinine / Salicylate / NSAIDS / Vanco / Erythromycin / Cisplatin / Vincristine / Vinblastine / Eflornithine / Deferoxamine				
Metabolic	DM / Hypothyroidism / Mucopolysaccharidosis				
Tumours	Acoustic neuroma / Meningioma				
Trauma	Head (fractures, concussion, penetrating) / NIHL / Barotrauma / Perilymph fistulas / Radiation				
Toxins	Mustard gas / Heavy metals				
Temporal bone diseases	Bone diseases / Metabolic / Granulomatous / Auto immune / Neoplastic				
Presbycusis	Neural – Normal STO and reduced SD	Sensory – Classical high tone loss, and later progress to other frequencies	Metabolic – Flat HL in young	Mechanical – Inner ear conductive component	
Inner ear diseases	Meniere's				
Oxygen	Hypoxia				

Congenital ear abnormalities

The inner ear embryologically develops separate from the middle ear and external ear canal. In roughly 25% of cases inner ear congenital abnormalities will overlap with middle ear and ear canal abnormalities. Isolated middle ear abnormalities are extremely rare and occur in less than 1% of cases. Any congenital abnormality can be divided into:

- Isolated (75%)
    - Teratogens
- Genetic
    - Syndromic
    - Non-syndromic

Table five below gives a more detailed description of congenital abnormalities, but this is just for reference and too detailed for a general practitioner.

Table 5. Congenital ear abnormalities

<table>
<tr><th colspan="6">Congenital ear abnormalities</th></tr>
<tr><td rowspan="11">Inner ear</td><td rowspan="2">Membranous</td><td>Complete</td><td>(Siebenmann-Bing)</td><td colspan="2">Jahrsdorfer 10 points:
Stapes – 2
Oval window 2 mm – 1
Round window 1 mm – 1
ME space > 3mm from prom to atretic plate – 1
NVII – 1
Malleus-incus – 1
Incus-stapes – 1
Mastoid pneumatisation – 1
EAC – 1</td></tr>
<tr><td>Limited</td><td colspan="3">Cochleo-saccular (Scheibe) / Cochlear basal turn</td></tr>
<tr><td rowspan="9">Membranous and osseus</td><td>Complete</td><td colspan="3">(Michel)</td></tr>
<tr><td>Cochlear</td><td colspan="3">Aplasia / Hypoplasia / Incomplete partition (Mondini) / Common cavity</td></tr>
<tr><td>Labyrinthine</td><td colspan="3">Aplasia / Hypoplasia / Dysplasia</td></tr>
<tr><td rowspan="2">Aqueduct</td><td>Vest aqueduct (VA)</td><td>Measure against post SSC >1.5-2mm</td><td>Associated with:
Isolated – Large VA syndrome
With cochlear abnormalities (Mondini)
With syndromes – Pendreds / BOR</td></tr>
<tr><td colspan="3">Cochlear aqueduct</td></tr>
<tr><td>IAM</td><td colspan="3">Narrow / Wide</td></tr>
<tr><td>NVII</td><td colspan="3">Aplasia / Hypoplasia</td></tr>
<tr><td>ME</td><td>25% also inner ear abnormalities 25% due to syndrome <1% in isolation</td><td>Stapes, Incus, Malleus, oval or round window – hypo, dys-, aplasia</td><td>Abnormalities of stapedial artery, carotid, jugular, NVII</td><td colspan="2">Types of stapes fixation:
Genetic
Syndromic – Treacher Collins / Branchio-Oto-Renal / Klippel-Feil / Crouzon
X-linked
Isolated
Trauma / Post surgery / Tympanosclerosis / Infections</td></tr>
<tr><td>EAC</td><td colspan="5">Schuknechts classification A – Fibro-cartilage narrow, B – A + TM / Malleus abnormalities, C – Total atresia, no TM, Malleus-Incus fixed, Stapes mobile, D – A + B+ C + extreme ossicle abnormalities, NVII abnormalities, Poor pneumatisation</td></tr>
<tr><td>Pinna</td><td colspan="5">Minor (Bat, Cupped, Lop, Constricted) / Microtia / Anotia</td></tr>
<tr><td>Hillocks</td><td colspan="5">Pre-auricular sinus / fistulas / pits</td></tr>
<tr><td>Branchial cleft</td><td colspan="5">Works I – Anterior to lobule, parallel to EAC, blind ending, only skin. Works II – Mandible angle, curves superior, true duplication because contain skin and cartilage</td></tr>
</table>

In practice

Take a thorough history, do a proper ENT examination which includes the tuning fork tests. In children always think about other neuro-developmental milestones and record that in your notes. Be careful of the very young child with pre-lingual hearing loss. Arrange for urgent otoacoustic emission (OAEs) to confirm or exclude congenital sensori-neural hearing loss and when in doubt rather refer to an ENT specialist / Audiologist. Children between the ages of 2 – 9 years of age, with post-lingual onset deafness is usually due to middle ear conditions (OME / AOM / Perforation). Remember that AOM has two peak incidences namely between 6 – 24 months and between 4 – 7 years of age. The first peak coincides with the immaturity of the ET due to its fairly horizontal orientation in the young skull. The latter peak is due to the adenoid and / or tonsil lymphoid tissue being at its most active. This then leads to middle ear conditions and mostly conductive hearing losses. Lastly, you will frequently see older people, and this has been discussed above.

Special investigations

Hearing tests

The following can be used:

- PTA
- Tympanometry
- Acoustic reflexes
- OAEs
- ABR
- In children also
    - Rattle test
    - Behavioural observational audiometry (0-6 months of age)
    - Visual reinforcement audiometry (6 months – 2 years)
    - Play – conditioning audiometry (2 years – 5 years)

Bloods

Genetic testing is becoming more available and also as panel tests. However, the clinical implications are still limited. Recently the first congenital genetically deaf children were given a reverse transcriptase viral vector to restore hair cells in the cochlea with positive results.

Radiology

The following can by useful

- MRI for pulsatile tinnitus, AIND, cerebro-pontine angle pathology, pre-cochlear implantation, congenital inner ear abnormalities
- CT for tinnitus, congenital inner ear abnormalities