Deafness in Children

Deafness is a partial or complete loss of hearing, also known as hearing impairment. The normal threshold range is 0-20 decibels (dB), where 0 dB is the threshold for the perception of sound at a given frequency for people with normal hearing.

Typical dB levels are around 30 dB for a whisper, 50 dB for average home noises and 60 dB for conversational speech. The pain threshold is at about 140 dB (which approximates to the sound of a jet engine).

Hearing loss is measured in decibels hearing loss (dB HL). It can be graded as follows:[1]

- 40-69 dB HL: moderate, cannot hear conversational speech.
- 70-94 dB HL: severe, cannot hear shouting.
- >95 dB HL: profound, cannot hear sounds that would be painful for a person with normal hearing.

There are two types of hearing loss:

- Conductive hearing loss: affects 4% of all school children. Almost all cases are due to glue ear. Sounds perceived by the brain are diminished but are generally not distorted.
- Sensorineural hearing loss: affects 0.3% of all schoolchildren. Sounds perceived by the brain are both diminished and distorted. The degree of distortion is independent of the degree of hearing loss (eg, a very mild hearing loss but very poor speech discrimination is possible).
- Mixed hearing loss has components of both conductive and sensorineural hearing loss.

Although conductive hearing loss is more common, the majority of permanent childhood hearing impairment is sensorineural.

Epidemiology

Deafness to some degree is a very common problem affecting more than 10 million people in the UK. Of these, 45,000 are children. About half of all deaf children are born deaf; about as many children again suffer from temporary deafness due to such conditions as glue ear.[2]

- Temporary deafness, due to Eustachian tube blockage and middle ear infections, is common.
- Most sensorineural hearing loss in children is congenital or acquired perinatally, but may present at any age. Approximately 10-20% of all deafness is acquired postnatally, although some genetic causes of deafness result in hearing loss that begins during childhood or adolescence.

Risk factors

- Family history of deafness.
- Infection: congenital (eg, rubella), mumps, meningitis.
- Ototoxic medications: in utero or postnatal.
- Low birth weight, prematurity, low birth Apgar scores, prolonged mechanical ventilation.
- Craniofacial anomalies or any syndrome associated with sensorineural hearing loss.
- Severe neonatal hyperbilirubinaemia.
- Head injury.
- Neurodegenerative disorders.
- Concern about hearing, speech, language or developmental delay.
Causes

- Genetic causes (50%): may or may not be part of a recognised genetic syndrome - eg, Turner syndrome, Klinefelter’s syndrome.
- Intrauterine (8%): congenital infection - eg, TORCH (toxoplasmosis, rubella, CMV, herpes), HIV; maternal drugs/toxins - eg, alcohol, cocaine, streptomycin.
- Perinatal causes (12%): examples include prematurity and/or low birth weight, birth asphyxia, severe hyperbilirubinaemia and sepsis.
- Postnatal causes (30%): childhood infections - eg, meningitis, encephalitis, head injury.
- Unknown causes: 20-30% of deaf children have no definite known aetiology.

Presentation

Presentation of hearing loss is dependent on the degree and patient age at onset of the hearing loss, the threshold for suspicion of parents and health care providers, and the presence of other identifiable risk factors. Always take parents seriously if they suspect that their child is deaf.

- Children with congenital or perinatally acquired profound sensorineural hearing loss (>90 dB) may present by age 6-9 months, whereas those with lesser degrees of hearing loss may present with minor speech impediments, language delay, behavioural problems or problems at school.
- Children with congenital hearing loss may have other physical stigmata.
- Children with conductive hearing loss should be assessed for serous otitis media and adenoidal hypertrophy - eg, mouth breathing, persistent cough (postnasal drip).
- Wax may be a cause of conductive hearing loss but this is not a common cause in children.
- Examination of the ear may reveal evidence of chronic infections (eg, perforation or scarring of the tympanic membrane, cholesteatoma, fluid behind the tympanic membrane).

Differential diagnosis

- The differential diagnosis of children presenting with language, behavioural or school difficulties should include hearing loss.
- Conductive hearing loss may be due to chronic serous otitis media, foreign body, wax.
- The precise aetiology of sensorineural hearing loss may be obvious (eg, post-meningitis) or remain uncertain.

Screening

See the separate article Hearing Testing and Screening in Young Children.

Investigations

- Specific tests for hearing loss include auditory brainstem response (formally the Brainstem Audio-evoked Response or the automated Auditory Brainstem Response), otoacoustic emissions and audiometry.
- Tympanometry.
- MRI or CT scan: may be indicated in investigation of any underlying cause of hearing loss.
- Chromosomal studies may be of benefit in seeking particular genetic syndromes.
Management

Management of moderate to profound permanent hearing impairment includes:

- Family support, advice and information.
- Communication support (spoken and/or signed), including:
  - Auditory-oral approaches maintain that many deaf children can develop their listening skills and a spoken language. Approaches to maximise the hearing a deaf child may have include:
    - Hearing aids
    - Radio aids (e.g., the teacher wears a transmitter and the child with hearing loss wears a receiver)
    - Cochlear implants
  - Lip reading/speech reading.
  - British Sign Language (BSL) is a visual language using hand shapes, facial expressions, gestures and body language to communicate. It is an independent and complete language with a unique vocabulary.
  - Sign bilingualism describes an approach that encourages the learning and using of two languages at the same time: a sign language and a spoken/written language.
  - Finger spelling: each letter of the alphabet is indicated by using the fingers and palm of the hand. It is used for spelling names, places, or for words that don’t have a sign.

- Pre-school education support.
- School education needs assessment and provision.

Conductive hearing loss

- Conductive hearing loss: grommet insertion and/or adenoidectomy may be required. If the hearing loss continues, amplification with a hearing aid may be needed. Speech therapy may also be required.
- Auto-inflation may be considered during the active observation period for children with otitis media with effusion who are likely to co-operate with the procedure. Autoinflation reopens the Eustachian tube by raising pressure in the nose - e.g., by forced exhalation with closed mouth and nose, blowing up a balloon through each nostril or using an anaesthetic mask. Hearing aids should be offered to children with persistent bilateral otitis media with effusion and hearing loss as an alternative to surgical intervention where surgery is contra-indicated or not acceptable. If otitis results in the destruction or fixation of the ossicles, surgery may help improve ossicular function.
- Cholesteatomas should be treated surgically.

Hearing aids

Externally worn aids
These are devices that increase the volume of the sound reaching the ear (effectively, amplifiers). They sit either behind the ear (although these devices are not powerful enough for patients with severe impairment) or just inside. Hearing aids that are placed right inside the external auditory meatus are available for patients with mild hearing loss. Bone conduction hearing aids for patients with conductive hearing loss are available in the form of headbands.

Analogue versus digital aids
Digital aids often (but not always) improve the quality of sound. Aids available on the NHS usually have a digital component to them.
Implantable aids

- **Cochlear implants**: simultaneous bilateral cochlear implantation is recommended as an option for children with severe-to-profound deafness (hearing only sounds that are louder than 90 dB HL at frequencies of 2 and 4 kHz without acoustic hearing aids) who do not receive adequate benefit from acoustic hearing aids.\(^5\)

- **Bone-anchored hearing aids (BAHAs).\(^6\)**
  - This type of surgery is reserved for patients with conductive and mixed hearing loss. It involves the fixing of a titanium implant just behind the ear, to which is connected an external abutment and a sound processor. Thus, it allows sound to be conducted through the bone rather than through the middle ear ('direct bone conduction').
  - The advantages are a better quality of sound and improved cosmesis (as they are less bulky than the headband system) but, as with cochlear implant surgery, there is a risk of unrealistic patient expectation about outcome.
  - There is also a risk of soft tissue reactions and loss of the fixture from its position in the skull.

Other equipment

There are various other products on the market, to assist with hearing. These range from hearing loops to vibrating pagers, visual trigger units for different situations (baby monitors, doorbells, fire alarms) and specialised alarm clocks and telephones.

Complications

There are huge personal, educational and social ramifications for the child born with hearing impairment.\(^7\) An holistic approach to managing these patients and their families is necessary. They may encounter problems stemming from lack of understanding by people around them and therefore educating the family is essential in order to help them pave the way for their child to live a fulfilled life.

**Bacterial meningitis and cochlear implants\(^8\)**

Patients with any type of cochlear implants are at a low risk of developing bacterial meningitis but this risk is slightly higher than for the normal population. It is for this reason that the Department of Health included this group of patients in the list of patients who should be immunised against pneumococcal infection, in 2002. Since 2006, babies will have received this immunisation anyway if they have followed the recommended immunisation programme but, for older patients, existing and prospective cochlear implant recipients should have their immunisation status checked, and be immunised accordingly at least two weeks before the implant.

Prognosis

- Conductive hearing loss: 50% of cases of glue ear resolve within three months and 95% within one year. Problems rarely persist beyond childhood.
- Early identification of permanent hearing loss is associated with better language, speech, social and emotional development.

Prevention

- **Rubella immunisation** programmes aiming at eradication.
- Early diagnosis and treatment of potential causes - eg. meningitis.
- Cautious prescribing of medications for pregnant and possibly pregnant women as well as children.

Further reading & references

1. Facts and figures on hearing loss and tinnitus; Action on Hearing Loss
2. Statistics; Action on Hearing Loss
3. Surgical management of children with otitis media with effusion (OME); NICE Clinical Guideline (February 2008)
5. Hearing impairment - cochlear implants; NICE Technology Appraisal Guidance, January 2009
6. Quality Standards in Bone Anchored Hearing Aids for Children and Young People; National Deaf Children's Society, Published March 2010.
8. Cochlear Implants; Medicines and Healthcare products Regulatory Agency (MHRA), July 2010

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