

## Hearing Loss

### Definition / Supporting Information

Hearing loss is described as being either conductive or sensorineural. It may range in severity from mild to profound.

The UK Newborn Hearing Screening Programme (NHSP), which aims to screen babies between birth and 5 weeks, has defined referral and management protocols for hearing loss in childhood.

Even mild patterns of hearing loss are far from benign and may cause academic, communicative, social, and emotional difficulty for children of any age.

### Essential History

#### Ask about:

- Parental concerns
  - The parent-held personal child health record (PCHR or “Red book”) contains pointers for parents about what to look and listen out for
- Previous newborn hearing screening or hearing tests
  - Hearing loss can occur or progress after screening
- Significant speech and language delays
- Behavioural problems, social difficulties, or academic difficulties
- Balance problems and / or clumsiness
- Congenital malformations
- Family history of hearing loss

### Differential Diagnosis / Conditions

In the UK, all children should have newborn screening and should also be monitored through the [Healthy Child Programme; Pregnancy and the first five years of life](#). Children with the following conditions have a higher risk of hearing loss and should be monitored and referred appropriately:

- Otitis media / otitis media with effusion (OME)
  - This is the most common cause of hearing loss in childhood.
  - It causes most problems when persistent or recurrent.
  - OME should be bilateral and persist for 3 months before intervention is considered.

- Congenital hearing loss
  - Mostly hereditary, although a negative family history is common
    - 80% autosomal recessive transmission
    - 18% autosomal dominant transmission
    - 2% X-linked recessive transmission
  - Most children with inherited hearing loss are non-syndromic.
- Syndromes
  - Down's syndrome
    - High risk for conductive hearing loss and higher than average risk for sensorineural loss
  - Alport's syndrome
  - Neurofibromatosis
  - Usher's syndrome
  - Waardenburg's syndrome
- Neurodegenerative disorders
  - Charcot–Marie–Tooth disease
  - Friedreich's ataxia
- Neonatal intensive care Indicators associated with progressive or late onset loss
  - Hyperbilirubinaemia requiring exchange transfusion
  - Persistent pulmonary hypertension of the newborn associated with mechanical ventilation
  - Use of extracorporeal membrane oxygenation (ECMO)
- In utero infections
  - Syphilis
  - Toxoplasmosis
  - Rubella (see PHE Green Book Chapter 28)
  - Cytomegalovirus
- Postnatal infections
  - Meningitis
  - Measles (see PHE Green Book Chapter 21)
  - Mumps (see PHE Green Book Chapter 23)
  - Herpes simplex
- Head trauma
- Treatment with potentially ototoxic drugs (eg, aminoglycosides)

## Investigations

To be undertaken by non-specialist practitioners (eg, General Practitioner (GP) Team):

- Confirm that newborn hearing screening is done for all infants before 5 weeks of age

- Ensure that by the age of 5 years children have undergone their pre-school hearing screening ([Healthy Child Programme](#); [Pregnancy and the first five years of life](#)).
  - Hearing screening should be carried out using an agreed, quality-assured protocol in appropriate surroundings.
  - Parental concern about hearing should always be noted and acted upon.

To be undertaken by specialist practitioners (eg, Paediatric / Paediatric Audiology / Ear, Nose and Throat (ENT) Team(s)):

- Define the degree and type (conductive or sensorineural) of hearing loss.
- Use a combination of physiological and behavioural measures to evaluate hearing in infants and young children, allowing for chronological age and development level.
- Investigate the aetiology of the hearing loss.

## Treatment Approach

To be undertaken by non-specialist practitioners (eg, GP Team), or by specialist practitioners (eg, Paediatric / Paediatric Audiology\* / ENT Team(s)):

- Monitor infants who have failed the newborn hearing screening to ensure that they receive appropriate referral and treatment.
- Communicate with the primary healthcare team (health visitor / school nurse / community paediatrician).
- Ensure that the child and family are supported and enrolled in appropriate services as defined under the specialist treatment approach below.
- Make parents / carers aware of behavioural and educational consequences associated with even mild hearing loss and suggest ways to help.
- Treat conditions such as wax, otitis externa, and otitis media.
- Patients with a significant conductive hearing loss due to otitis media with effusion, should be seen and retested at 6–12 weeks and referred to a specialist if the hearing loss is present on two separate occasions (see Otitis media with effusion [[NICE clinical knowledge summary](#)]).

To be undertaken by specialist practitioners (eg, Paediatric / Paediatric Audiology\* / ENT Team(s)):

- Be sure that the child has received a thorough medical evaluation (including genetics consultation and visual assessment) to determine the cause of the hearing loss.
- Approximately 30–40% of children with hearing loss have additional disabilities.
  - These children should have periodic developmental screening and surveillance as an integral part of their management.
- The specialist team should work together with the appropriate services, including educational support services and specialist social care.

\*Professional make-up of Paediatric Audiology Teams varies across the UK.

## Specific Treatment

- Communication, hearing aids, and language development
  - Although many children have conductive and sensorineural hearing losses not amenable to medical treatment, they can benefit from amplification, educational support, speech and language therapy, and sign language.
  - Traditional hearing aids are designed to pick up sounds, convert them to electrical signals, amplify, filter, and then convert signals back to acoustic signals for a receiver.
  - In noisy settings, frequency-modulated systems can be used alone or in combination with hearing aids (eg, a microphone worn by a teacher to amplify only the teacher's voice while minimising interfering background noise).
- Cochlear implants
  - Bilateral cochlear implants (both implanted during the same operation) are recommended for children with severe or profound deafness if they do not get enough benefit from hearing aids after 3 months (see Cochlear implants for children and adults with severe to profound deafness [[NICE technology appraisal guidance TA166](#)]).
    - The system takes sound from the environment and converts it to an electronic stimulus which is delivered to the spiral ganglia / auditory nerve.
      - This provides a sensation of hearing but does not restore "normal" hearing.
    - Cochlear implants have been shown to be beneficial in:
      - Auditory and speech perception
      - Speech production
      - Localisation of sound (also increasing personal safety)
      - Quality of life

## When to Refer

Refer to specialist practitioners (eg, Paediatric / Paediatric Audiology\* / ENT Team(s)) for a full audiological evaluation if:

- An infant or child fails a hearing, speech, or language screening measure in primary care
- A child with a known loss shows signs of deterioration
- The patient or caregiver is concerned about hearing, speech, or language development
- Family history of permanent hearing loss (especially if there is concern)
- Postnatal infections associated with sensorineural hearing loss (eg, meningitis)

- History of in utero infections
  - Syphilis
  - Toxoplasmosis
  - Rubella (see PHE Green Book Chapter 28)
  - Cytomegalovirus
  - Herpes
- Neonatal indicators associated with progressive or late onset hearing loss
  - Hyperbilirubinaemia requiring exchange transfusion
  - Persistent pulmonary hypertension of the newborn associated with mechanical ventilation
  - Use of extracorporeal membrane oxygenation
- Syndromes associated with hearing loss
  - Alport's syndrome
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- Head trauma

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## **'Safety Netting' Advice**

- The personal child health record (PCHR or "Red Book") contains a useful guide to age-appropriate development of hearing and communication that may help to alert parents to potential problems.
- Support parents / carers and children to seek help if they suspect hearing loss or deterioration of a known loss.
- Warn parents that children with hearing loss are especially vulnerable to the effects of otitis media with effusion, because additional hearing loss can negatively affect the audibility of speech through hearing aids.
  - If they suspect ear or upper respiratory tract infections, these should be evaluated promptly by the primary care practitioner.

## Patient / Carer Information

***\*Please note: whilst these resources have been developed to a high standard they may not be specific to children.***

- [TA166 Hearing impairment – cochlear Implants: understanding NICE guidance](#) (Web page), National Institute for Health and Care Excellence
- [Deafness in Children](#) (Web page), Patient.co.uk
- [Hearing Loss](#) (Web page), NHS Choices

## Resources

### National Clinical Guidance

[Surgical management of otitis media with effusion](#) (Web page), NICE clinical guideline CG60, National Institute for Health and Care Excellence.

[Cochlear implants for children and adults with severe to profound deafness](#)(Web page), NICE technology appraisal guidance TA166, National Institute for Health and Care Excellence.

### Medical Decision Support

[Rubella](#) (Web page), Public Health England's Green Book

[Measles](#) (Web page), Public Health England's Green Book

[Mumps](#) (Web page), Public Health England's Green Book

### Suggested Resources

***\*Please note: these resources include links to external websites. These resources may not have national accreditation and therefore PCO UK cannot guarantee the accuracy of the content.***

[Otitis media with effusion](#) (Web page), NICE clinical knowledge summary, National Institute for Health and Care Excellence.

[NHS Newborn Hearing Screening Programme](#) (Web page), Public Health England (including [Reactions to sounds checklist](#) (pdf) and [Leaflets](#) (Web page) relating to neonatal hearing test and hearing loss).

[Hearing Loss](#) (Web page), NHS Choices.

Shribman S, Billingham K. [The Healthy Child Programme: Pregnancy and the first five years of life](#), London: Department of Health; 2004 (overview pp23-24; detail pp 31-57).

[National Deaf Children's Society](#) (Website), The National Deaf Children's Society.

[Glue ear: A guide for parents](#) (Web page), The National Deaf Children's Society.

[British Association of Paediatricians in Audiology \(Website\)](#), British Association of Paediatricians in Audiology.

## Acknowledgements

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