# Hearing loss in children

DR NICHOLAS LEITH and DR NIRMAL PATEL investigate the latest thinking on this relatively common problem.

HEARING loss in children is a common problem that encompasses a broad differential diagnosis. Knowledge of normal auditory developmental milestones, risk factors and common causes of hearing loss are essential in managing the child with hearing loss. Early intervention is important in preventing delays in speech and language development.

The incidence of hearing loss (moderate or greater) in Australia is estimated to be between nine and 12 children per 10,000 live births. Furthermore, 23 per 10,000 children will acquire a hearing impairment severe enough to require hearing aids by the age of 17.

Indigenous children in Australia experience a greater burden of ear and hearing problems than non-Indigenous children. Indigenous infants less than one year of age are four times as likely as non-Indigenous infants to develop acute otitis media (AOM) and three times as likely to have otitis media with effusion (OME).

Hearing loss in children may be classified by the child's age and type of the hearing loss.

Conductive hearing loss (CHL) occurs when there is a problem with the conduction of sound waves from the environment to the inner ear. Sensorineural hearing loss (SNHL) occurs when there is a problem with the transduction of soundwaves into neural impulses, and the conduction and interpretation of these by higher centres. Mixed hearing loss occurs when there is both a conductive and sensorineural component to the hearing loss.

In infants with congenital hearing loss, the cause is often sensorineural in nature. About 60% of cases of congenital SNHL will have a genetic cause and of these, 15-30% are syndromic, with over 400 syndromes described to include hearing loss. In the infant with hearing loss, a thorough perinatal history to exclude in-utero infections known as TORCH (toxoplasmosis, other [such as syphilis, varicella, mumps, parvovirus and HIV], rubella, cytomegalovirus and herpes simplex virus) is required. Other important risk factors include: a family history of congenital hearing loss, perinatal ototoxic exposure, hyperbilirubinaemia requiring exchange transfusion and admission to neonatal ICU. In children, infection is the most common cause of hearing loss, predominantly due to AOM and OME which result in a conductive hearing loss. Table 1 summarises the risk factors and red flags for hearing loss in children.

The differential diagnosis of hearing loss in children is extensive.

## Assessing a child's hearing

Normal hearing is within the range of 20-20,000Hz. Most human speech lies within the range of 250-8000Hz and the loudness of conversational speech is about 65dB.

There are various methods used to measure hearing in children. Newborn screening uses auditory brainstem response, which involves measuring action potential responses of the eighth cranial nerve to an auditory stimulus.

For infants who are unable to understand verbal instruction, visual reinforcement audiometry (VRA) can be used as a modification of standard audiometry. VRA involves operant conditioning by rewarding the child when they turn their head towards the direction of a sound. VRA is used for children between the ages of six months and two years. In children aged between two and five years, play audiometry may be used to assess hearing. Children over the age of five may be assessed using conventional pure tone audiometry. Tympanometry can also be used as an adjunct to assess middle ear function when assessing hearing loss.

Based on pure tone audiogram measurements, normal hearing is classified as thresholds that fall within the range of 0-25dB.

**Mild hearing loss 25-40dBHL:** Children with mild hearing loss usually have normal speech, but will have trouble in the school setting because it may be difficult to hear speech from more than 4m away, or when there is background noise. Much of the meaning in English is contained in the voiceless consonants that are high-pitched and soft: s, sh, t, p, k, ch and th.

**Moderate hearing loss 40-55dBHL:** Children with moderate hearing loss can clearly hear speech only when the speaker is very close. They need hearing aids to hear the softest sounds and to acquire understandable speech.

**Severe hearing loss 70 -90dBHL:** Children with a severe hearing loss do not perceive speech, no matter how close they are to the speaker. They will not learn to speak intelligibly without hearing aids and special help.

**Profound >90dBHL:** Children with profound hearing loss do not hear speech or other sounds. The younger they are fitted with hearing aids or cochlear implants, the greater the likelihood they will eventually speak intelligibly.

Having a good understanding of the normal developmental milestones for hearing and speech is essential; it enables early detection of a child that may have hearing loss that is affecting their speech and language development. Normal developmental milestones for hearing and speech are summarised in table 2 (next page). It is important to note that a child who is not talking by 18 months is not considered normal.

## Two rarer but important conditions of hearing loss in children

### Enlarged vestibular aqueduct syndrome (EVAS)

The vestibular aqueduct is a bony canal that runs from the vestibule of the inner ear through the temporal bone, opening into the posterior cranial fossa.

Enlargement of the vestibular aqueduct is the most common radiologically detectable malformation of the inner ear. EVAS is usually bilateral and affected children will be born with normal to mild hearing impairment that deteriorates as the child ages in a stepwise progression, often associated with head trauma. At least 40% of those affected eventually develop profound SNHL. Vestibular symptoms, such as vertigo attacks lasting 15 minutes to three hours, may be precipitated by either head injury or vigorous spinning.

### Congenital cholesteatoma

This is a temporal bone epidermoid cyst, thought to be caused by the retention of epithelial cells in the middle ear during fetal development.

Typically patients with congenital cholesteatoma will not have a history of otitis media. Middle ear cholesteatomas may present either with conductive hearing loss or may be detected incidentally on routine otoscopic examination as a bulging whitish mass behind an intact tympanic membrane, most often in the anterosuperior quadrant. Congenital cholesteatomas are managed surgically, with the aim to create a clean dry ear and avoid the complications associated with expan-

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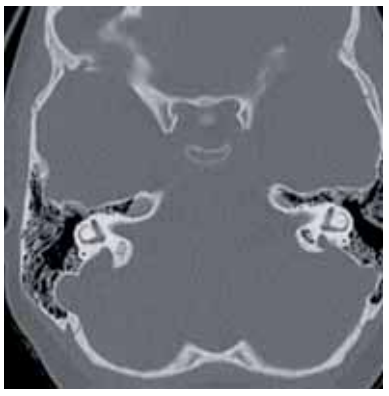
**TABLE 1. Risk factors and red flags for hearing loss in children**

- Family history of permanent childhood hearing loss
- In-utero infections: toxoplasmosis, other rubella, cytomegalovirus, and herpes (TORCH)
- Perinatal factors:
  - Low birthweight (<1.5kg)
  - Ear and other craniofacial abnormalities (cleft palate, microtia*)
  - Apgar 0-3 at five minutes and 0-6 at 10 minutes
  - Respiratory distress
  - Admission to neonatal ICU
  - Mechanical ventilation for more than 10 days
  - Hyperbilirubinaemia requiring transfusions
  - Ototoxic medication administered for more than five days (eg, gentamicin) or used in combination with loop diuretics
  - Syndromes known to included hearing loss (Down syndrome, Waardenburg, Alport and Pendred syndromes)
- Parental/caregiver concern regarding speech, language or developmental delay
- Recurrent or persistent otitis media with effusion (OME) for at least three months
- Head trauma with fracture of the temporal bone
- Chemotherapy
- Neurodegenerative disorders (eg, Hunter syndrome) or sensory motor neuropathies (eg, Charcot–Marie–Tooth syndrome, Friedreich ataxia)

*underdevelopment of the pinna

Adapted from the Joint Committee on Infant Hearing. Year 2000 Position Statement: Principles and Guidelines for Early Hearing Detection and Intervention Programs. *Pediatrics* 2000; 106:798-817.



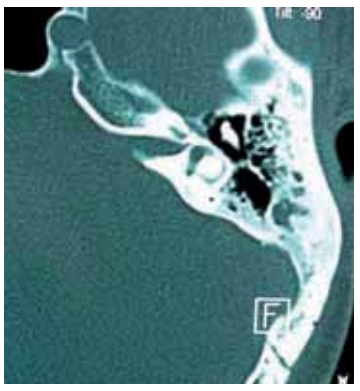


**Figure 1. Axial CT scan showing bilateral enlargement of the vestibular aqueducts in the image on the left, compared with normal anatomy in the image on the right.**

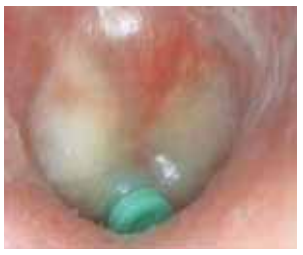


**Figure 2. Congenital cholesteatoma. This patient was referred after squamous debris was noted in the middle ear following grommet insertion for hearing loss. A white mass lesion is seen behind the tympanic membrane along with the grommet.**

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sion of the cholesteatoma including facial nerve damage, sensorineural hearing loss, vertigo and intracranial extension.

## Management of hearing loss

The benefit of early intervention in children with hearing loss has been well established.

In a study of children with SNHL, diagnosis by six months of age, followed by immediate intervention (within two months) resulted in significantly better language, speech and social–emotional development when compared with children with hearing loss identified after six months of age. This was the case even when diagnosis after six months of age was followed by intervention.

In addition, parents with early identified children reported less parental stress and maternal depression than those of children identified after six months of age.

## Mild to moderate sensorineural hearing loss

Hearing aids may be required in children with mild to moderate hearing loss to improve speech and language development.

## Severe to profound sensorineural hearing loss

If hearing aids are not providing adequate speech and language development, cochlear implants provide a way of treating severe to profound sensorineural hearing loss by stimulating auditory nerve cells.

The components of a cochlear implant, as seen in figure 3 (right) include: A microphone (1) for sensing environmental sounds and a speech processor to transform the sounds into digital signals. A transcutaneous link (2) for transmission of the digital signal to the implanted receiver–stimulator (3). This transmits the digital signal to an electrode array that is placed into the scala tympani of the cochlea, to stimulate the spiral ganglion cells of the auditory nerve (4).

## Update on the treatment of otitis media with effusion

OME is the most common cause of acquired conductive hearing loss in children. It is defined as the presence of fluid in the middle ear in the absence of acute inflammation. There has been recent debate regarding the use of intranasal steroids in the treatment of OME. A systematic review investigating the use of intranasal steroids in improving hearing in children with OME and hearing loss did not find any evidence of benefit. National Institute for Clinical Excellence (NICE) guidelines for the surgical management of OME in children suggest that children with persistent bilateral OME over a period of three months, with a hearing level in the better ear of 25-30dBHL or worse should be considered for surgical management.

Antibiotics, antihistamines, decongestants and steroids are not recommended in the NICE guidelines for management of OME. In children with bilateral OME persisting for three months, there is evidence to suggest that ventilation tubes improve hearing at six months. However, the effect diminished after this time .

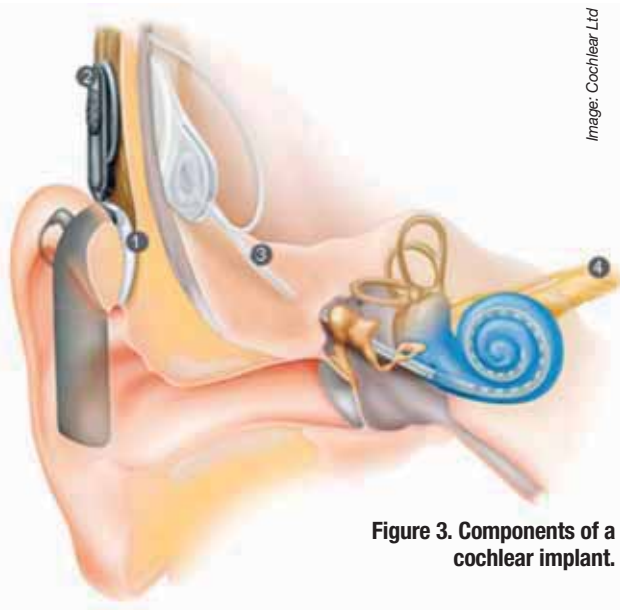


Image: Cochlear Ltd

**Figure 3. Components of a cochlear implant.**

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## References

Available on request from jeannie.yoo@reedbusiness.com.au

**TABLE 2. Normal developmental milestones for hearing and speech**

| Age | Milestones |
|---|---|
| Newborn | Variable response to sound.<br>Startles to sudden loud noises |
| 1 month | Soft guttural noises when content |
| 6 weeks | Quiets in response to<br>soft sound 15cm from ear |
| 3 months | Turns head to sound at ear level |
| 6 months | Visually locates soft sounds<br>at 40-60cm at ear level |
| 9 months | Tries to communicate vocally<br>Localises soft sounds above<br>and below ear at 1m |
| 12 months | Says 2-3 wvords with meaning |
| 18 months | Says 5 -20 recognisable words,<br>understands many more |
| 2 years | Two- and three-word phrases |
| 3 years | Gives full name, uses plurals<br>3-5 word sentences |
| 4 years | Asks many questions<br>Gives name and address<br>Names four primary colours |
| 5 years | Speech fluent, with good articulation |

Adapted from *Paediatrics Manual: The Children's Hospital at Westmead Handbook*

### When to refer to ENT

- Children with suspected hearing loss
- OME persisting for greater than three months, or less than three months in the presence of speech delay
- Recurrent AOM (more than three infections in six months)
- Complications of AOM (including persistent headache, surrounding abscesses, facial nerve weakness, non-resolving infection)
- Suspicious-appearing tympanic membrane (suspected cholesteatoma)