Newborn hearing loss

Early diagnosis is vital

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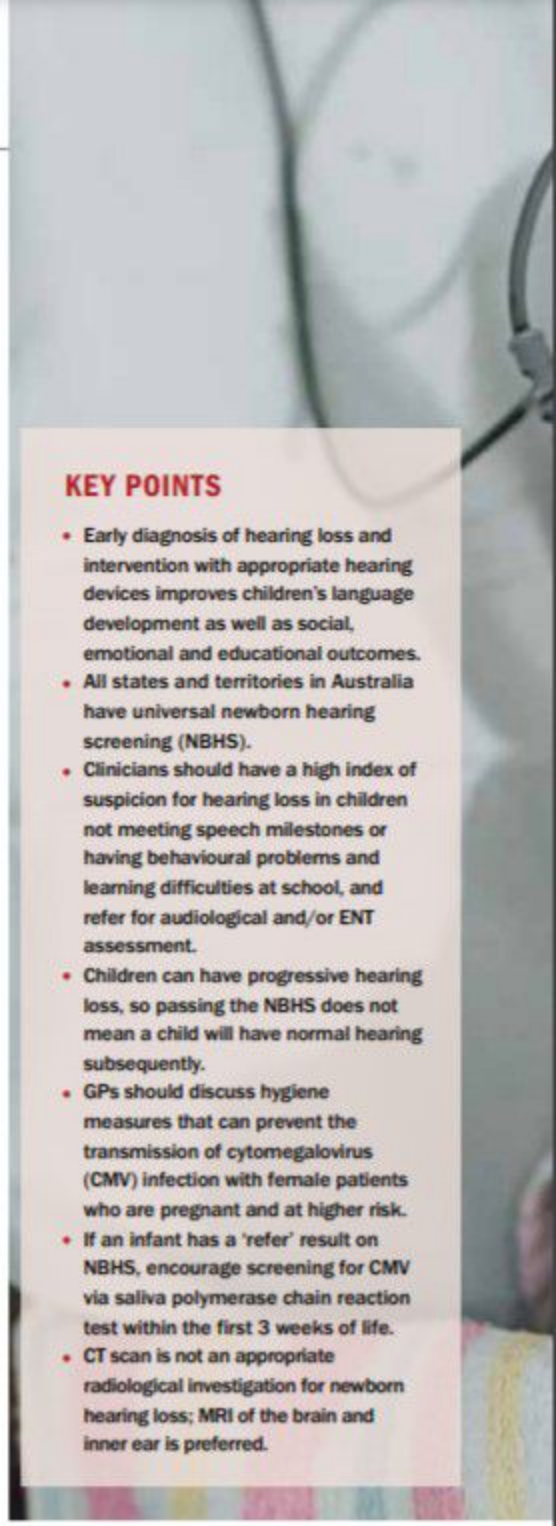
In Australia, three in every 1000 children are born with some degree of hearing loss. Early diagnosis is the key to early intervention and improved outcomes from hearing rehabilitation.



KEY POINTS

- Early diagnosis of hearing loss and intervention with appropriate hearing devices improves children's language development as well as social, emotional and educational outcomes.
- All states and territories in Australia have universal newborn hearing screening (NBHS).
- Clinicians should have a high index of suspicion for hearing loss in children not meeting speech milestones or having behavioural problems and learning difficulties at school, and refer for audiological and/or ENT assessment.
- Children can have progressive hearing loss, so passing the NBHS does not mean a child will have normal hearing subsequently.
- GPs should discuss hygiene measures that can prevent the transmission of cytomegalovirus (CMV) infection with female patients who are pregnant and at higher risk.
- If an infant has a 'refer' result on NBHS, encourage screening for CMV via saliva polymerase chain reaction test within the first 3 weeks of life.
- CT scan is not an appropriate radiological investigation for newborn hearing loss; MRI of the brain and inner ear is preferred.

In Australia, one in every 1000 children is born with bilateral hearing loss in the moderate to profound range. Three children in every 1000 are born with some degree of hearing loss.

Most permanent hearing loss detected at birth (92%) is due to sensorineural causes including auditory neuropathy spectrum disorder (ANSD). A small proportion of cases (8%) are related to conductive hearing loss. Otitis media with effusion is a common cause of temporary conductive hearing loss in children and the most common cause of hearing loss worldwide. Congenital permanent conductive hearing loss can be due to abnormalities of the pinna, ear canal or middle ear resulting in microtia, canal atresia or ossicular chain abnormalities.

Early diagnosis of hearing loss and appropriate intervention are important in improving children's language development and social and educational outcomes.

This article focuses on the investigation and management of sensorineural hearing loss.

Newborn hearing screening

Universal newborn hearing screening (NBHS) has revolutionised the diagnosis and management of newborn hearing loss. Testing is carried out using the automated auditory brainstem response (AABR), an electrophysiological test where electrodes are placed on the forehead and behind the ears of the sleeping baby. The results are binary: pass or refer (fail). A refer result requires a repeat AABR test within the next 48 hours. If the refer result is confirmed by the repeat test, then the newborn is referred for diagnostic audiology at a specialist centre (usually a children's hospital).

Diagnostic audiology should be performed promptly after a refer result on the repeat AABR test and this can be carried out with the infant asleep, often after feeding. The diagnostic tests include acoustic auditory brainstem response, otoacoustic emissions and tympanometry.

Like any investigative tool, NBHS is not perfect. There are rare instances when hearing loss at birth may be missed or, more commonly, the child may have normal hearing at birth but progressive hearing

MedicineToday 2020; 21(8): 12-17

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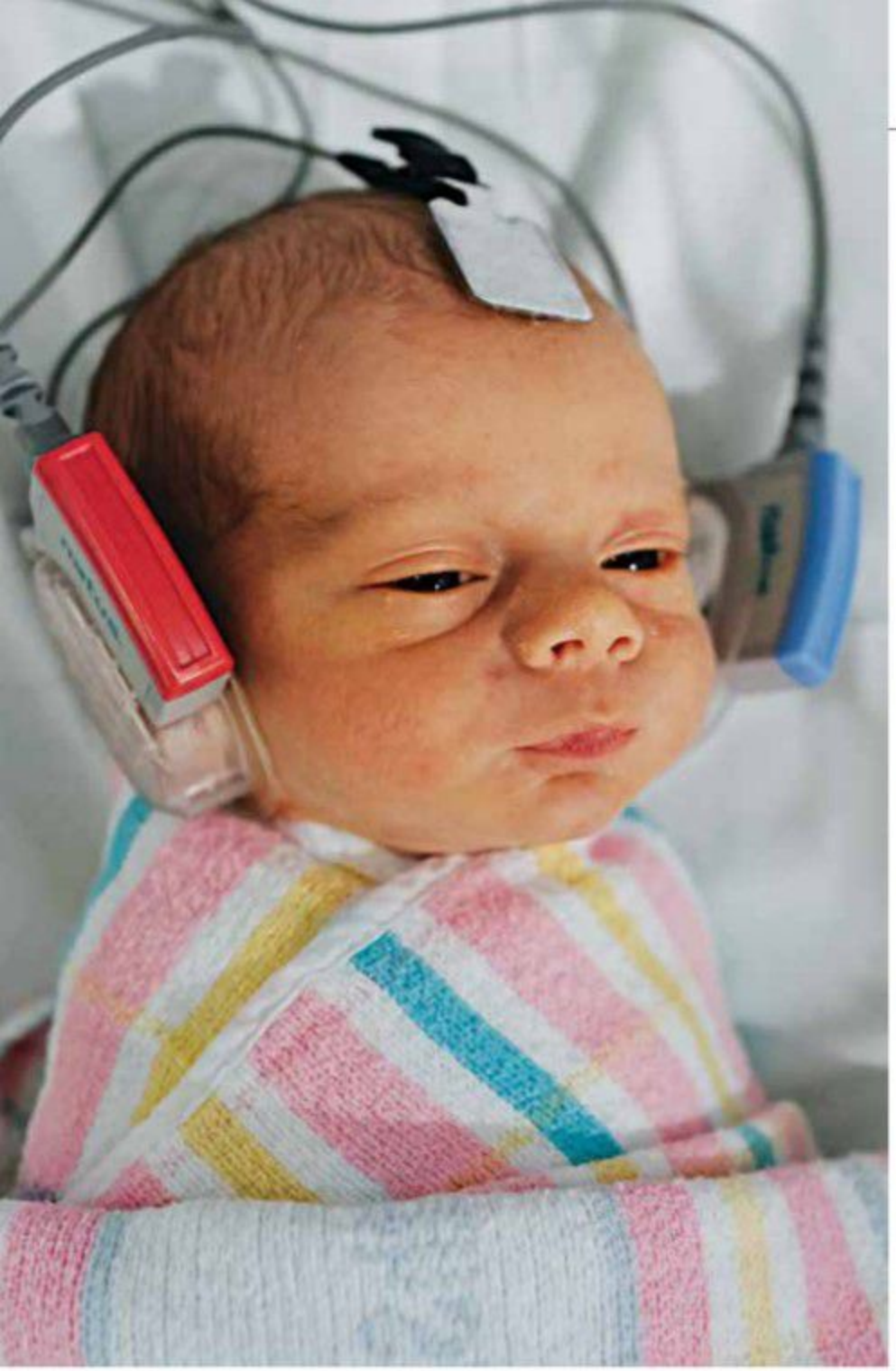

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loss over the following months and years.

Parents may notice children increasing the television volume or repeatedly missing verbal cues. Clinicians should have a high index of suspicion for hearing loss in children not meeting speech milestones, or having behavioural problems and learning difficulties in school or a diagnosis of attention deficit hyperactivity disorder. For these children, consider referral for audiological and/or ENT assessment as indicated.

Sensorineural hearing loss

The cause of hearing loss can be determined through a series of investigations including MRI scanning, blood and urine tests, as well as saliva tests for cytomegalovirus (CMV) by polymerase chain reaction (PCR). Supplementary investigations include ECG and ophthalmological examination when indicated.[1]

MRI of the brain, inner ear and internal auditory meatus is the most appropriate radiological investigation for newborn hearing loss. CT scan is not a first-line investigation and should not be ordered unless specified by an otolaryngologist or paediatrician with expertise in the assessment and management of newborn hearing loss.

With history, examination and investigation, an aetiology can be identified in two-thirds to three-quarters of children.

Causes of sensorineural hearing loss

Congenital bilateral sensorineural hearing loss is due to environmental, genetic and structural causes. Environmental causes account for approximately one-third of cases and include pre- and perinatal events such as congenital infections; extreme prematurity and risk factors associated with neonatal intensive care admission; ototoxic medications; meningitis; and jaundice requiring exchange transfusion. Congenital CMV infection is the most common environmental cause.

Genetic forms of hearing loss account for approximately half of bilateral sensorineural hearing loss. One-third of these will be syndromic (e.g. trisomy 21, Waardenburg syndrome, Usher syndrome). The remainder are nonsyndromic and mostly inherited recessively (85%). Up to 10% are transmitted in an autosomal dominant fashion and 2 to 5% through an X-linked or mitochondrial pattern of inheritance. More than 150 genes that cause hearing loss have been identified.

Structural causes of bilateral sensorineural hearing loss can be sporadic, environmental or genetic. Structural problems include cochlear dysplasia, cochlear nerve hypoplasia and large vestibular aqueduct syndrome.

In unilateral congenital sensorineural hearing loss, approximately 40% of children will have a structural abnormality on MRI such as cochlear nerve deficiency, cochlear dysplasia and enlarged vestibular aqueduct.[2] Congenital CMV infection is another common cause. Genetic causes are uncommon in unilateral hearing loss and are usually syndromic (e.g. trisomy 21, branchio-oto-renal syndrome, Waardenburg syndrome).