# Identification and management of childhood hearing loss

Most cases of hearing loss in children are not associated with syndromes.

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Hearing loss is one of the most common conditions that affects newborn babies worldwide. The spectrum of hearing loss ranges from mild loss of hearing affecting one ear, to profound hearing loss in both ears. Identification of hearing loss in young children is difficult because most cases are non-syndromic (i.e. there are no other dysmorphic features). Awareness of risk factors for hearing loss will contribute to the identification of only 50% of neonatal cases. Universal newborn hearing screening is now the accepted international standard. Management of these cases involves a multi-professional team and must be initiated as early as possible. In cases of severe to profound hearing loss, there is a limited window of opportunity to allow for development and integration of central auditory function. One of the major hurdles in the management of childhood hearing loss is the scarcity of schools that can educate children with hearing impairments.

#### Scale of the problem

The incidence ranges worldwide from 0.5 to 6.0 per 1 000 live births for severe to profound hearing loss, and developing countries such as South Africa probably have an incidence at the upper end of that spectrum.1-5 It is estimated that annually over 6 000 babies in South Africa are born with bilateral, permanent hearing loss.6 The incidence climbs even higher during early childhood, mainly owing to middle-ear infections and meningitis. A recent review estimated that the prevalence of hearing loss in children aged 5 - 14 years in sub-Saharan Africa is 1.9%.3 South Africa has a population of 10.6 million children in this age bracket, and a further 5.2 million below 5 years of age.7,8 Using a conservative prevalence of 1% for the combined group

one can estimate that 1.5 million children under the age of 15 years in South Africa have some form of hearing impairment.

### **Hearing loss**

Disabling hearing impairment in children under the age of 15 years has been defined by the World Health Organization (WHO) as a permanent, unaided hearing threshold for the better hearing ear of 31 dB or greater.9 In this case, the hearing threshold is the average threshold of hearing at 0.5 kHz, 1 kHz, 2 kHz and 4 kHz (Fig. 1). A relatively recent diagnostic term, i.e. auditory neuropathy/dyssynchrony (AN/ DS), underscores the fact that hearing loss encompasses a spectrum of disorders causing a disruption or distortion of auditory information reaching or being processed by the central nervous system. Hearing loss is categorised as conductive (CHL) or sensorineural (SNHL), with SNHL being further subdivided as cochlear or retrocochlear in origin.

# Hearing loss is one of the most common conditions that affects newborn babies worldwide.

### Actiology

CHL in children may be due to congenital malformations of the outer/middle ear, wax impaction, persistent middle-ear effusions (Fig. 2), chronic infectious conditions, i.e. chronic suppurative otitis media (CSOM) or cholesteatoma. These conditions all impede the transmission of sound energy from the outside to the inner ear where it can be converted into neural impulses. CSOM and cholesteatoma may also result in damage to the inner ear or central nervous system. The degree of hearing loss caused by these conditions may range from a few decibels up to 60 decibels. However, this may be misleading because it is a very crude measure of hearing – it does not take into account the frequencies involved or whether the pathology is unilateral or bilateral. These factors have a major influence on fine auditory cues that our brains use to process auditory information.

The majority of cases of congenital hearing loss are caused by genetic malformations (55 -70%<sup>10</sup>). The remainder are either infectious (with congenital cytomegalovirus infection being the leading culprit), caused by maternal ototoxic drug use during pregnancy, or idiopathic. Acquired childhood hearing loss is usually caused by infections, trauma, ototoxic medication or missed/progressive congenital cases. AN/ DS has been associated with numerous different possible aetiological factors. Neonatal jaundice and congenital infections feature prominently on this list.

Seventy per cent of genetic cases are non-syndromic. This lack of additional clinical signs helps to explain the often late diagnosis of hearing loss. Eighty per cent of genetic cases are autosomal recessive. Thus, the affected child has no family history of hearing loss to alert clinicians to the possibility of hearing loss.

#### Identification

### Importance of early identification

The neural connections between the inner ear and primary auditory cortex of the brain are intact in virtually all cases of disabling hearing impairment. However, functional connections between the primary auditory

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Fig. 1. Audiogram showing the frequencies and amplitudes of common sounds and the 'speech banana' (adapted from: www. brentbolthousephotography.com – accessed via Google images, 16 March 2012).



Fig. 2. A chronic middle-ear effusion.

cortex and secondary auditory centres develop over time owing to stimulation. These secondary or association areas are involved in the interpretation or understanding of sound stimuli, with subsequent development of connections, with other areas of the central nervous system (CNS) involved with communication, learning, emotional response, etc. These connections are also essential for the development of one's internal communication framework ('inner speech'). There is a limited window of opportunity in which these connections may be formed. This process is greatest in the first two years of a child's life and then tapers off until about four years, with minimal to no development after the age of six. A process of 'synaptic pruning' takes place from the age of about two years, in which the CNS reorganises itself to sever unused connections and re-allocate unused functional areas of the brain. The secondary auditory areas are usually re-allocated to vision, proprioception and vibration detection ('use it or lose it'). This has been determined through studies using functional MRI techniques and PET scanning.<sup>11</sup>

A lack of infantile and early childhood auditory stimulation will result in a permanent functional communication handicap with associated learning difficulties, impaired cognitive development, and emotional/psychological issues.<sup>5</sup>

In addition to the degree of hearing loss and the absolute age of the child when the hearing loss is reversed, the actual delay in receptive

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and expressive language development is also crucial. Most children in whom profound hearing loss has been corrected will acquire these skills at the same rate as a child with 'normal hearing'. To correct this delay requires years of speech therapy, implying that the earlier a child is identified and managed, the better the brain's ability to use the auditory stimulation and the shorter the delay in language development that needs to be overcome.

Awareness of risk factors for hearing loss will contribute to the identification of only 50% of neonatal cases. Universal newborn hearing screening is now the accepted international standard.

#### Hearing screening

Prior to the implementation of widespread oto-acoustic emission (OAE) screening in developed countries, the average age of diagnosis of congenital hearing loss was approximately 20 months. Children were evaluated based on the presence of known risk factors for hearing loss or as a result of delayed speech development. Targeted screening based on the presence of risk factors is no longer advocated as it resulted in the detection of only half of the cases of congenital hearing loss. Universal newborn hearing screening (UNHS) by means of OAE is the gold standard for hearing screening worldwide. The goals of UNHS are to screen all babies before the age of 1 month, to complete diagnostic testing by 3 months, and to implement communication intervention strategies by 6 months. The USA has the most comprehensive national screening programme, with 94% of all newborns being screened, and 64% of eligible infants being enrolled in early intervention programmes.1

Despite the endorsement of UNHS by the HPCSA as far back as 2002, little has been

done locally to meet these goals. The vast majority of newborns nationally are not screened for hearing loss. Less than 1% of public hospitals in South Africa offer UNHS<sup>6</sup> and hearing screening in private hospitals is voluntary. One local study in a private hospital setting found a screening rate of 20%.<sup>4</sup> Hospital management and paediatric health services must prioritise hearing screening as part of the standard of care for newborns.<sup>5</sup>

#### Hearing evaluation in children

Accurate hearing evaluation in children is difficult and time consuming. Audiologists use a test battery to evaluate children, and a diagnosis should be based on the collective results of all tests in the test battery. The various tests often evaluate different functional areas of the auditory system, so results are complementary as well as confirmatory. The younger the child, the less able she/he is to actively participate in the process and the greater the reliance on objective tests for accurate threshold determination. However, objective test results must always be interpreted in conjunction with behavioural test results.

Objective tests include:

- tympanometry
- auditory reflex testing
- OAE testing
- auditory evoked potential (AEP) testing
  - auditory brainstem response (ABR)
  - auditory steady-state response (ASSR).

Objective tests have the advantage that they can be performed from birth, while not relying on any active participation from the child. A disadvantage is that children need to be asleep for all forms of AEP testing and be quiet for other forms of testing. These tests only test the auditory pathway up to brainstem level; hence the need for subjective testing.

Subjective testing includes:

- visual reinforced audiometry (VRA)
- play audiometry
- pure tone audiometry
- speech discrimination.

VRA is performed in a soundproof room with two audiologists. Calibrated sounds are presented by speakers around the room and conditioned and unconditioned behavioural responses are used as indicators for hearing. The results of this test reflect free-field responses to sound and do not localise hearing to a specific ear. Play audiometry provides ear-specific results. The two lastmentioned subjective tests would only be possible in older children. In experienced hands, reliable subjective test results are possible from around 6 months of age.

#### Management

Treatment for childhood CHL may involve medical and surgical management, ranging from myringotomy and ventilation tube (grommet) insertion for persistent middleear effusions, tympanomastoidectomy for cholesteatoma with recon- struction of the ossicular chain, to bone-anchored hearing systems for congenital aural atresia. Conservative management in the form of temporary or permanent hearing aids may be advisable in cases where the likelihood of successful surgery is low owing to various patient-related factors.



Fig. 3. A CT image of a child with congenital inner-ear malformations. The white arrows indicate enlarged vestibular aqueducts, and the black arrow indicates a cystic space formed by the middle and apical cochlear turns.

### **Childhood hearing loss**

The management of a child with congenital hearing loss involves a multi-disciplinary team that includes otorhinolaryngologists, audiologists, speech therapists, parents (and other family members), teachers, paediatricians, radiologists (Fig. 3), child psychologists and occupational therapists. This reflects the central role of hearing in a child's development.

The benefits of early rehabilitation of hearing loss affect more than speech development. The educational and socio-economical benefits for the child are well established, and the economic benefit for society has also been proven.<sup>2,4-6,11,12</sup>

It is known that at least a third of children with profound hearing loss have additional neurological pathology. These include motor delay, global developmental delay, behavioural disorders (including attentiondeficit disorders and depression) and language disorders (including autismspectrum disorders and language-learning disorders). These may only become apparent after initiation of auditory rehabilitation and at an older age.

Good communication is essential in the multi-disciplinary professional team. Rehabilitation needs to be structured and well monitored and lack of progress in receptive and/or expressive language skills should prompt referral to a cochlear implant programme for further evaluation. The small window of opportunity for intervention in early childhood hearing loss should always be respected

## Acquired childhood hearing loss is usually caused by infections, trauma, ototoxic medication or missed/ progressive congenital cases.

All children with hearing loss should be evaluated by an otorhinolaryngologist. Hearing loss is a medical condition, the cause of which can range from wax impaction to complex inner-ear malformations secondary to genetic defects. The otorhinolaryngologist can arrange appropriate special investigations and manage the majority of these cases. The remainder should be referred to tertiary academic centres/cochlear implant programmes for further management.

A close working relationship between audiologist and speech therapist is essential

in all cases. Despite advances in diagnostic audiology and hearing aid technology, the response to amplification is measured in a subjective manner – this can be difficult in a 1-year-old child. The speech therapist needs to keep the audiologist advised on indicators of insufficient amplification – the hearing aid programme can then be modified accordingly.

Parents or primary caregivers form the backbone of the rehabilitation process. The speech therapist's role is to guide the parents and child through the rehabilitation process. This usually involves a consultation of 1 - 2 hours per week. The parents, who spend the majority of the week communicating with the child, will determine the success or failure of a rehabilitation programme. The greater the hearing loss, the more intensive the rehabilitation process needs to be. A child with severe to profound hearing loss and who has hearing aids or a cochlear implant will require daily focused one-on-one therapy with a caregiver for at least 2 - 3 hours. Undoubtedly, this has a huge impact on the family dynamics. Support groups and parent guidance sessions are available throughout the country to assist parents.

References available at www.cmej.org.za

### **IN A NUTSHELL**

- Childhood hearing loss has a high prevalence in South Africa.
- Hearing loss encompasses a spectrum of disorders causing a disruption or distortion of auditory information reaching or being processed by the central nervous system.
- Childhood hearing loss may be congenital or acquired.
- Congenital hearing loss is mostly genetic and non-syndromic, so there will not be any visible clue to suggest the hearing loss.
- Congenital hearing loss has a profound impact on the development of all communication skills, which in turn has a severe impact on most areas of cognitive development.
- Universal newborn hearing screening is the gold standard for early detection of hearing loss and needs to be implemented in South Africa.
- There is a limited window of opportunity in which congenital or early childhood deafness may be corrected.
- Childhood hearing loss is evaluated by means of a test battery as many tests evaluate different parts of the auditory system.
- The management of congenital hearing loss requires a multi-professional team as well as a motivated family.