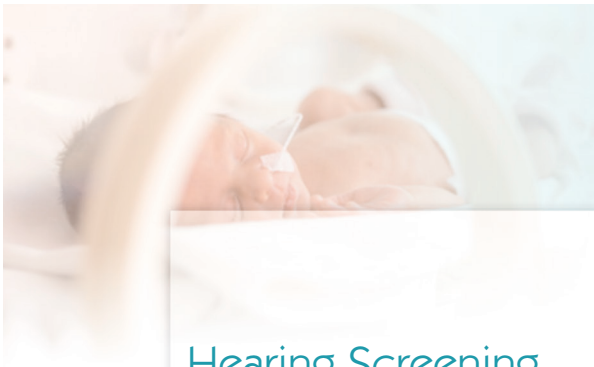

CHAPTER

36

Hearing Screening

The hearing threshold of more than 25 decibels (dB) at the frequencies (500, 1000, 2000 and 4000 Hertz) which are important for speech recognition can result in significant auditory deficits. Hearing loss can be slight, moderate, severe or profound hearing loss based on the average threshold (Table 36.1).

Hearing loss may be conductive, sensorineural or mixed based on the site of lesion.

1. Conductive hearing loss presents in the outer or middle ear restricting the sound waves to propagate to the inner ear. It can be due to deformity in outer/middle ear such as micotia/anotia, atresia, ossicular deformity etc or due to transient obstruction of middle ear with fluid.
2. Sensorineural hearing loss ceases the conduction and processing of acoustic signals in the inner ear secondary to damage to cochlea, auditory nerve or central auditory pathway itself. It can be further subdivided into sensory loss wherein the hair cells in the cochlea are damaged and neural loss in which the auditory nerve and the neural pathway are affected.
3. Auditory neuropathy occurs when the auditory signal is either absent or processed abnormally in the auditory nerve, brainstem, or cerebral cortex.

Table 36.1: Grade of hearing impairment

Grade of Impairment	*Corresponding audiometric value in decibels (dB)*
0. No impairment	25 or better
1. Slight impairment	26–40
2. Moderate impairment	41–60
3. Severe impairment	61–80
4. Profound impairment including deafness	81 or greater

4. Mixed hearing loss is the condition when conductive component is present along with the sensorineural component.

HEARING SCREENING[1–3]

Implementation of hearing screening programme ensures early diagnosis of hearing impairment. The average age of identification of bilateral profound hearing loss has been estimated to be 24 months and impairment of lesser degree has been noted to be 48 months. This can be significantly reduced to 12 months or lesser with execution of neonatal hearing screening. It can be either "universal" or "high risk". However, high risk focused screening programmes have shown to miss out almost 50% of the congenital hearing loss cases. Therefore, several authorities have proposed to implement universal hearing screening protocol.

Two techniques are used for screening:[4]

1. **Otoacoustic emissions:** Otoacoustic emissions (OAE) are an objective, efficient and noninvasive tool to assess cochlear function. They are the reflected acoustic vibrations from the cochlea in response to the stimulus given through the testing probe. They are obtained as a result of cochlear sensory cells, primarily outer hair cell movement, and are obtained at frequencies essential for speech acquisition.

   The OAE screener consists of a probe that is placed inside the ear. The probe houses a small microphone along with the speakers. The click stimuli in case of transient evoked otoacoustic emissions (TEOAEs) or tones in case of distortion product otoacoustic emissions (DPOAE) are given through speakers and the microphone receives the reflected sound from the cochlea for SNR (Signal to noise ratio). It gives the result as 'PASS' or 'REFER'.
2. **Automated auditory brainstem response:** Automated auditory brainstem response (AABR) is a screening tool to record electrophysiological response against the auditory stimulus given at the threshold level. The AABR method produces a simple 'PASS' or 'REFER' result without requiring interpretation. It is important to note that both AABR and auditory brainstem response (ABR) are different. ABR being a diagnostic test gives the detailed information such as type and degree of hearing loss. However, AABR is a screening test that measures the response and gives the result as 'PASS' or 'REFER'. In high risk infants, OAEs alone

can assess the sound conduction through middle ear and cochlear function but will miss the condition such as Auditory Neuropathy (AN). Therefore, it is recommended to perform AABR along with OAE in such cases.

AABR screening device measures the surface signals by placing electrodes on the forehead, mastoid, and the nape of the neck. For all the high-risk infants, AABR is essentially to be used as part of screening.

Execution of screening programme can give targeted results if it follows the 1-3-6 principle (Fig. 36.1). States who meet the 1-3-6 benchmark (screening completed by 1 month, audiologic diagnosis by 3 months, enrollment in early intervention by 6 months) should strive to meet a 1-2-3 month timeline.[5,6]

- The hearing screening should be performed by one month of age either before or after discharge for all the newborns.
- The babies who fail initial and follow-up screening should undergo diagnostic audiological evaluation to confirm the degree and type of hearing loss by the age of three months.
- After the diagnosis of hearing loss, the children identified with congenital hearing loss should be intervened with appropriate medical management or amplification devices such as hearing aids or cochlear implants by the age of 6 months.

SCREENING PROTOCOL[5,7]

A two-step screening programme is the most effective and widely accepted arrangement to ensure high sensitivity of the testing protocol (Fig. 36.2). In this the babies are screened initially within 24–48 hours of birth and in case they get REFER result, they are re-screened preferably before discharge or on subsequent visit to the hospital. This reduces the number of false-positive cases due to presence of fluid in the ear during initial hours of birth.

In high-risk babies, initial screening should be carried out with both OAE and AABR. Considering the greater incidences of hearing impairment in children falling under the high-risk category, it is imperative to perform OAE as well as AABR to ensure neural hearing loss is not missed in these children. Also, to decrease false-positive results it is advisable that hearing screening is done after 34

Fig. 36.1: 1-3-6 Principle for hearing screening

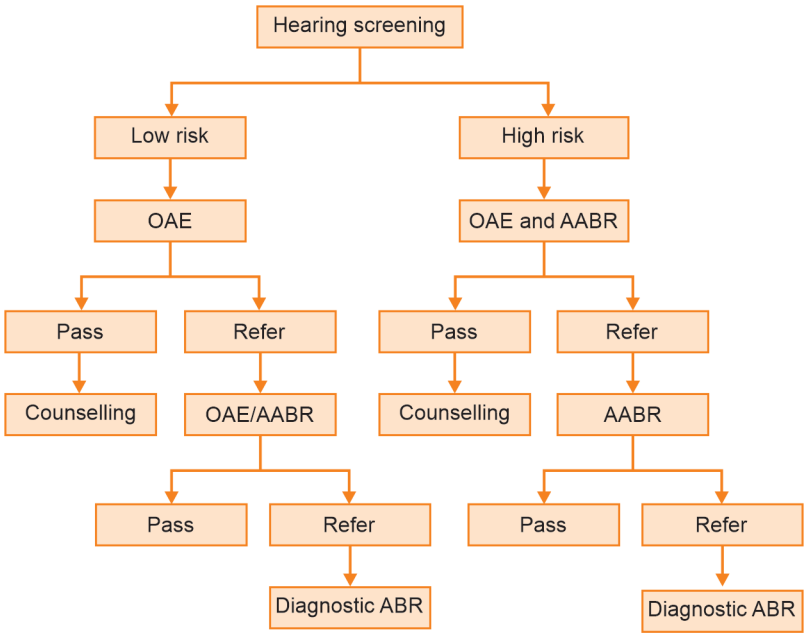

Fig. 36.2: Screening protocol for low-risk and high-risk babies

ABP: Auditory brainstem response; *AABR:* Automated ABR; *OAE:* Otoacoustic emission

weeks postmenstrual age for premature infants born at <34 weeks of gestation age (Fig. 36.2).

Evidence of two-step screening protocol and AABR for high-risk infants[4]

A retrospective study has advocated the use of two step screening protocol including Automated ABR (auditory brainstem response)/OAE (otoacoustic emissions) for well babies and inclusion of AABR along with OAE for high-risk babies.

IMPLEMENTATION[6]

The hearing screening programme needs resources such as appropriate instrumentation, isolated and less-noisy area for testing and manpower for successful implementation. Also, the execution of UNHS is majorly dependent on attitude and belief of physicians at the medical facilities involved in neonatal care. They should ensure hearing screening of all the neonates along with the appropriate test battery for well babies and high-risk infants. Counselling of parent/caregivers to increase awareness regarding importance of hearing screening and its implication is imperative to warrant the success of screening programme.

FOLLOW-UP

All the children failing two-stage hearing screening should undergo diagnostic evaluation by 3 months of age. The test battery includes:

1. Medical evaluation: To examine the outer and middle ear and check for the presence of impacted wax or fluid.
2. Tympanometry to assess the middle ear function.
3. Otoacoustic emissions to differentially diagnose sensorineural hearing loss and auditory neuropathy.
4. Auditory brainstem responses (ABR): It is the gold standard test for threshold estimation and site of lesion analysis and also to help to determine the management option suitable.

Along with ear examination and audiological evaluation, bilateral congenital hearing loss should also prompt for genetic work-up. The genetic testing may give the opportunity to predict the possible course of impairment. It can help to establish the connection of auditory impairment with other congenital anomalies and differentiate between underlying syndrome or an isolated hearing loss.

LIMITATIONS

One of the major limitations of UNHS that poses a great challenge for the medical professionals is delayed-onset hearing loss. The late-onset progressive hearing loss, mostly associated with the known risk factors such as congenital cytomegalovirus (CMV) infection, hyperbilirubinemia, etc. can be identified with parental awareness and follow-up. However, the impairment with delayed onset and without associated risk factor can fail the purpose of screening as they cannot be captured with UNHS. The pass UNHS results may give false reassurance to parents resulting in missed or delayed identification.

How to Manage[8]

In accordance with the recommendation of WHO the rehabilitation for hearing impairment should be initiated by six months of age. It should be a multidisciplinary approach including otorhinolaryngologists, audiologists and parents. Based on the type and degree of loss, appropriate management options should be communicated to parents or caregiver. The habilitation process may include speech and language therapy along with the following options (Fig. 36.3):

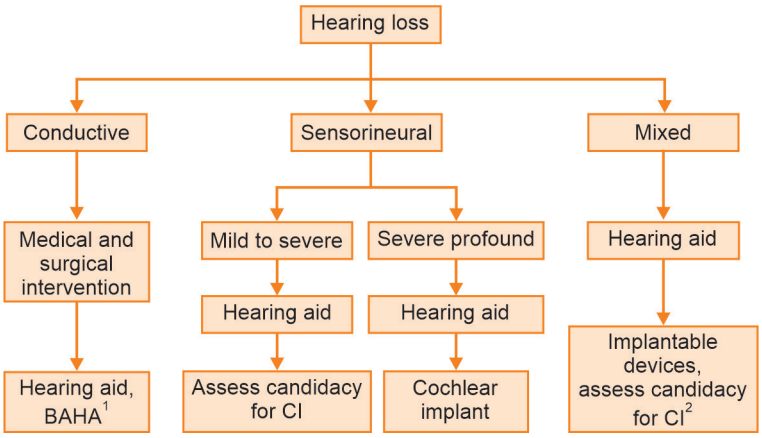

Fig. 36.3: Management of hearing loss post-diagnostic evaluation
BAHA: Bone-anchored hearing aid; *CI:* Cochlear implant

1. **Medical and surgical intervention:** Effective medical treatments are available for variety of conditions causing conductive hearing loss like middle ear effusion, chronic otitis media and acute otitis media. Some conditions like chronic tympanic membrane perforation, ear atresia and congenital ossicle abnormalities can be effectively managed by surgical correction. Immediate attention to these conditions and intervention can help to avoid delay in speech development.
2. **Hearing aids:** They can be suggested in cases of hearing loss with sensorineural component. Aids can be prescribed for hearing impairment ranging from mild to severe degree with the adequate amplification and appropriate programme. A strong follow-up and re-evaluation are critical to ensure the benefit.
3. **Bone-anchored hearing aid (BAHA):** In cases of permanent conductive hearing loss, BAHA can be suggested. It is most suitable in cases of congenital malformations of middle and external ears or chronically discharging ear. Surgical implantation should be planned considering the age of the child.
4. **Cochlear implant (CI):** In cases of severe to profound hearing loss cases, cochlear implants are the most suitable intervention option. Children with no or inadequate benefits with amplification devices can be surgically implanted with cochlear implants. The surgery performed by twelve months of age or during the critical period results in significant improvement and near normal development of speech and language skills.

Evidence of benefits from universal neonatal hearing screening

A prospective study of large cohort of children indicated towards significant benefit from early detection and intervention of hearing impairment followed by improved language outcomes.

OUTCOMES[6]

Hearing impairment when diagnosed and treated in early years of development can minimize its effects on communication skills of the child. The timely medical intervention and use of appropriate amplification device will assist children with hearing loss in achieving speech and language skills at par with their normal hearing counterparts. It will also be helpful in attaining normal cognitive development and social integration.

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