Screening for hearing impairment in neonatal period is pivotal for timely detection and appropriate treatment. Early diagnosis provides the opportunity for early intervention in hearing impaired children, thus enhancing their overall quality of life.

**Definition**
Hearing loss is defined by measuring the hearing threshold decibels (dB) at different frequencies. Normal hearing has a threshold of 10 to 15 dB. In individuals with bilateral hearing loss, the severity of loss is based on the better-functioning ear. Hearing loss can be categorized as

- No hearing loss: 10 to 15 dB
- Slight: 16 to 25 dB
- Mild: 26 to 40 dB
- Moderate: 41 to 55 dB
- Moderately severe: 56 to 70 dB
- Severe: 71 to 90 dB, or 61 to 80 dB based on the World Health Organization (WHO) definition
- Profound: >91 dB, or >80 dB based on WHO definition

All newborns irrespective of risk factors should be screened for hearing impairment.

**Rationale of screening**
Automated auditory brain stem response (AABR) and otoacoustic emissions (OAE) are recommended for newborn hearing screening. Either or both may be used, depending upon the number and profile of the deliveries at any centre, and expertise and equipment available. Both AABR and OAE techniques are portable, inexpensive, automated and reproducible.
Otoacoustic emissions (OAE)

**Principle:** Transient evoked otoacoustic emissions (TEOAEs) are frequency-dispersive responses arising within the cochlea. Since OAE evaluates hearing from the middle ear to the outer hair cells of the inner ear, it is used to screen for sensorineural hearing loss (SNHL) but cannot detect auditory neuropathy (AN).

**Technique:** The OAE screener consists of small microphone that is placed in the ear canal of the infant. The screener sends stimuli in form of clicks or tones and also receives the reflected sound from the cochlea. The device measures the signal-to-noise ratio to make sure that recordings are accurate.

Automated Auditory Brainstem Response (AABR)

**Principle:** It is an electro-physiologic measurement that is used to assess auditory function from the eighth nerve to the inferior colliculus of the mid brain in response to a click stimulus. The AABR method produces a simple ‘pass’ or ‘fail’ result without requiring interpretation. It is important to note the difference between AABR and ABR; ABR being a diagnostic test which provides quantitative data (e.g, waveforms) that must be interpreted by a trained audiologist, thereby determining the degree and the site of the hearing loss.

**Technique:** AABR equipment measures the surface signals by placing electrodes on the forehead and the mastoid, and on the nape of the neck. Most commercially available devices effectively screen infants younger than 6 months.

Two-step screening

A two-step screening means that if any ear fails the first screen, a repeat screening should be done on both ears within a specified time frame. The repeat screening in such cases should be done prior to discharge of the infant from the hospital. For all infants (both with and without risk factors), a two-step screening should be done (two-step OAE, two-step AABR or TEOAE followed by AABR) (Figure 42.1).
Two-step screening: What is the evidence?
A large community-based trial showed that a 2-step screening approach (OAEs followed by ABR for those who failed the first test) yielded a sensitivity of 0.92 and specificity of 0.98.

- In infants without risk factors, OAEs may be used at both steps, but for infants with risk factors (Table 39.1), it is imperative to use an AABR for the initial screen so that neural hearing loss will not be missed.
- For premature infants (born at <34 weeks of gestation), hearing screening should ideally be done after they reach 34 wks postmenstrual age to reduce false positive results.
- For readmissions in the first month of life in infants who are at high risk (e.g., hyperbilirubinemia requiring exchange transfusion or meningitis), a repeat hearing screening is
recommended before discharge.

- Any infant missing initial screen should be instructed to return for screen after 6 weeks, which may coincide with the immunization visit (to minimize loss to follow-up).

**Table 42.1: Infants with risk factors**

| 1. | Caregiver concern regarding hearing, speech, language, or developmental delay |
| 2. | Family history of permanent childhood hearing loss |
| 3. | Admission in Neonatal intensive care unit of more than 5 days or history of any of the following irrespective duration of stay in NICU: ECMO, assisted ventilation, ototoxic medications (gentamycin and tobramycin) or loop diuretics (furosemide), and neonatal hyperbilirubinemia requiring exchange transfusion |
| 4. | Intrauterine infections, such as CMV, herpes, rubella, syphilis, and toxoplasmosis |
| 5. | Craniofacial anomalies, particularly those that involve the pinna, ear canal, ear tags, ear pits, and temporal bone anomalies |
| 6. | Syndromes associated with hearing loss or progressive or late-onset hearing loss |
| 7. | Careful evaluation for neurodegenerative disorders such as Hunter syndrome, or sensory motor neuropathies, such as Friedreich ataxia and Charcot-Marie-Tooth syndrome |
| 8. | History of culture-positive postnatal infections particularly with meningitis (including confirmed bacterial and viral) |

**Follow up**

The neonates who do not pass screening should have a comprehensive audiological evaluation by 3 months of age. It is mandatory for infants with confirmed hearing loss to receive appropriate intervention within 6 months of age from health care and education professionals with expertise in hearing loss.

**References**