

Abdulsalam Al-Qahtani, Reni K. Chandran,
Khaled Abdulhadi, and Zaid Altamimi

66.1 Introduction

The etiology of childhood hearing impairment can be congenital (at birth) or acquired (after birth). It can affect the child in the pre-lingual or post-lingual stage, depending on whether the child has acquired the ability to speak. Early detection and prevention of childhood hearing impairment play a major role in the management of this condition.

Permanent childhood hearing impairment (PCHI) defined as a confirmed permanent bilateral hearing impairment ≥ 40 dB HL (hearing level) averaged over the frequencies of 500 Hz, 1 KHz, 2 KHz, and 4 kHz in the better hearing ear. It can result from a structural abnormality in the outer ear, middle ear as well as the inner ear. Hence PCHI can be a conductive, sensorineural, or mixed hearing loss [1].

Congenital hearing loss occurs in approximately 1 to 2 per 1000 live births. Genetic factors account for at least 60% of childhood hearing loss, among children with PCHI, about 70% are non-syndromic, and the remaining 30% are syn-

dromic [2]. The onset of the hearing loss can be prenatal, perinatal, or postnatal in onset. Syndromic or non-syndromic is based on the presence or absence of other clinical abnormalities and the pattern of inheritance (autosomal, X-linked, dominant, recessive, mitochondrial). Eighty percent of cases are autosomal recessive typically of pre-lingual onset or 20% autosomal dominant typically progressive or post-lingual [3]. X-linked or mitochondrial inheritance trait (1%) is passed through the maternal lineage [4]. Table 66.1 shows the most common syndromes with hearing loss.

Table 66.1 Syndromic hearing loss

<i>Autosomal dominant</i>	
1.	Branchio-oto-renal syndrome
2.	Neurofibromatosis
3.	Otosclerosis
4.	Stickler syndrome
5.	Treacher Collins syndrome
6.	Waardenburg syndrome
<i>Autosomal recessive</i>	
7.	Jervell and Lange-Nielsen syndrome
8.	Pendred syndrome
9.	Usher syndrome
<i>X-linked</i>	
11.	Alport syndrome
12.	Norrie syndrome
13.	Otopalatodigital syndrome
14.	Wildervanck syndrome
15.	Mohr-Tranebjaerg syndrome
16.	X-linked Charcot-Marie-Tooth

A. Al-Qahtani · Z. Altamimi
Hamad Medical Corporation, Doha, Qatar
e-mail: aalqahtani@hamad.qa ; Zaltamimi@hamad.qa

R. K. Chandran (✉) · K. Abdulhadi
Audiology Department, Hamad Medical Corporation,
Doha, Qatar
e-mail: rchandran1@hamad.qa; Khadi@hamad.qa

For more about congenital hearing loss syndromes, risk factors for permanent congenital, delayed-onset, or progressive childhood hearing loss, please go to the congenital hearing loss chapter.

Acquired causes of childhood hearing loss can be due to: otitis media with effusion (OME), ear infections, meningitis, measles, encephalitis, chickenpox, flu, mumps, ototoxic medications, head injury, and noise exposure.

Otitis media with effusion (OME) is an inflammatory condition, defined by the accumulation of fluid in the middle ear with an intact tympanic membrane without acute infection manifestation. It is the most common cause of pediatric acquired hearing loss and speech delay. Several risk factors associated with an increased incidence of OME include younger age, eustachian tube dysfunction, craniofacial abnormalities, adenoid hypertrophy, recurrent otitis media, daycare attendance, gastroesophageal reflux, and passive smoking. Usually, it resolves spontaneously in >80% of cases; in persistent OME cases with significant hearing loss more than 25 dB, a myringotomy with ventilation tube insertion is indicated.

66.2 Pediatric Hearing Assessment

The goals of a pediatric evaluation are:

1. identify the existence of an auditory disorder.
2. identify the nature of the disorder.
3. identify the extent of the hearing impairment caused by the disorder.

Hearing assessment in infants and young children presents a special challenge in reaching a diagnosis. Infants are usually referred because of a known risk factor for hearing loss or on failing neonatal hearing screening. In contrast, young children are usually referred due to a delay in speech and language or because they have an otologic disease with an effect on hearing. Older children are usually referred because of an oto-

logic problem, failing a school screening, or suspected auditory processing disorder.

66.2.1 Newborn Hearing Screening Program

The newborn hearing screening program aims to identify all children with a moderate–profound PCHI in the better hearing ear [5]. Current screening tests available for newborn hearing screening are automated otoacoustic emissions (AOAEs) using either transient or distortion product otoacoustic emissions or automated auditory brainstem response (AABR) [6]. Most screening programs use a two-stage protocol (either OAE followed by AABR if required or two-stage AABR) [5–7]. A pass in both ears using AOAE, or a pass in both ears on AABR, constitutes an overall screen pass. Though OAE is quick and minimally invasive, the disadvantage is that it can be affected by the presence of fluid/debris in the outer or middle ear. If used alone, it could miss neonates with auditory neuropathy. Babies cared for in NICU for more than 48 hours undergo a transient AOAE screen and an AABR screen. Those that fail the AABR screen in one or both ears are referred for audiological assessment [7].

All cases of bilateral severe to profound sensorineural hearing loss should have a full assessment which includes: *pediatric history* (detailed history of pregnancy, delivery and postnatal period, developmental milestones including speech and language and motor milestones, pre- and postnatal noise exposure, history of ototoxic medications, head injuries, ear disease, meningitis, viral illness, and immunization status), *family history* (deafness or risk factors associated with hearing loss in first- and second-degree relatives), *clinical examination*, *audiological assessment* (age-appropriate tests as mentioned below), *imaging studies* (magnetic resonance imaging and/or computed tomography of inner ears), *referral to clinical geneticist* (connexin 26 and 30 mutations testing with access to clinical genetics service for counseling), *ophthalmic assessment*, *vestibular investigations*, and *other investiga-*

tions (electrocardiograph, viral serology (TORCH), renal ultrasound, thyroid tests).

66.2.2 Pediatric Audiological Test Battery

The 2007 Joint Committee on Infant Hearing 1-3-6 guidelines recommend diagnosis of hearing loss by 3 months of age and intervention by 6 months of age [8]. The Pediatric Hearing Assessment Task Force has delineated the following areas that make up the pediatric audiology assessment test battery:

- History.
- Clinical examination.
- Behavioral observation audiometry (BOA).
- Visual reinforcement audiometry (VRA).
- Conditioned play audiometry (CPA).
 - Frequency-specific stimuli.
 - Speech audiometry.
- Physiologic assessments:
 - Acoustic immittance, including tympanometry and acoustic reflex testing.
 - Otoacoustic Emission (OAE) testing.
- Electrophysiologic audiometry:
 - Auditory Brainstem Response (ABR).
 - At least one electrophysiological measure of threshold prediction should be completed before fitting hearing aids and/or cochlear implant, for any infant under the age of 3 years [8].

Hearing assessment for children can be objective or subjective and it varies according to age. Figure 66.1 shows the hearing tests according to the age group, while Table 66.2 shows the comparison between the subjective and objective hearing tests.

66.2.2.1 Immittance Audiometry

Acoustic impedance describes the resistance to energy transmission while acoustic admittance describes the ease of energy transmission through the auditory system. The term “acoustic immittance” has been introduced to express either acoustic condition. Acoustic immittance

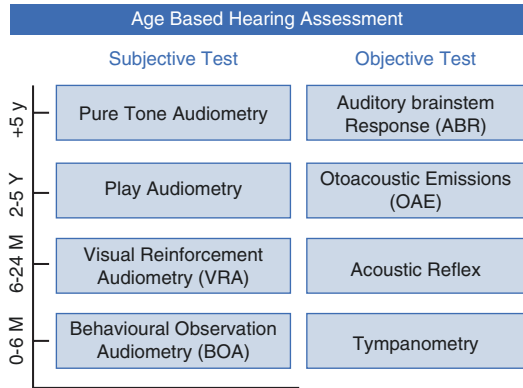


Fig. 66.1 Age-based hearing assessment

Table 66.2 Comparison between subjective and objective hearing

Subjective tests	Objective tests
• Requires reliable and consistent response to sound from the child	• No voluntary response required from the child
• Depends on developmental age	• Can be done on infants, young children, and non-compliant subjects with developmental delay
• Not used in newborn screening	• Used in newborn hearing screening
• Done when the child is awake	• Done mostly in deep sleep/quiet

audiometry serves at least three functions in audiological assessment [9]:

1. Detects middle ear disorder.
2. Differentiates cochlear from retro-cochlear disorder.
3. Cross-checks pure-tone audiometry.

Three immittance measures of middle ear function commonly used are: tympanometry, static immittance, acoustic reflex thresholds. It should be a routine component of the audiological evaluation; the benefit of starting with immittance audiometry is that it provides a useful indication of what to expect from pure-tone audiometry.

The probe tube has three apertures [10, 11]:

1. Transducer which emits a low-frequency probe tone (226 Hz) of approximately 85 dB

Table 66.3 Tympanometry parameters

	Peak compliance (mmho or cc)	Ear canal volume (cm ³)	Middle ear pressure (daPa)
<i>Pediatric age</i>			
Mean	0.5	0.7	
Range	0.2–0.9	0.4–1.0	–50 to +50
<i>Adult age</i>			
Mean	0.8	1.1	
Range	0.3–1.4	0.6–1.5	–100 to +50

Table 66.4 Age-based threshold assessment audiometry

Age group	Subjective hearing test
0–6 months	Behavioral observation audiometry (BOA)
6–24 months	Visual reinforcement audiometry (VRA)
2–5 years	Conditioning play audiometry (CPA)
>5 years	Pure-tone audiometry

66.2.2.2 Audiometry

Audiometry can be challenging for the pediatric age especially when it is a subjective test. Table 66.4 shows the age-based audiometry methods.

Behavioral observation audiometry (BOA): used for the age group between 0 and 6 months where it observes the changes in the child’s behavior in response to a sound stimulus (eye widening or blink, alteration in sucking response or arousal from sleep, startle or shudder of the body, definite movement of the arms, legs or body). The sound stimulus may be presented for <2 s, in a horizontal plane, 15 cm from the child’s ear, out of peripheral vision. The response is observed by the distractor.

Visual reinforcement audiometry (VRA): method of choice for screening children between 6 months and 2 years. Young children are trained by operant conditioning to produce a response to sound. A clear response is a 90° spontaneous head turn within 2–3 s of the sound stimulus reinforced with a visual stimulus, so the child learns to respond to the stimulus to seek a reward. It is considered normal when the response is acquired in the sound field at 25 dB HL.

Conditioned Play Audiometry (CPA): used for the age group between 2 and 5 years. It helps in determining ear-specific and frequency-specific hearing sensitivity. The patient is seated at a child-sized table in an appropriately sized chair. Conditioning is a training session to train the child for the play task test. The transducers can be an insert earphone, supra-aural earphones, bone vibrator, or sound field speakers. Play tasks include placing or tossing a block in a box, stacking blocks in response to an auditory stimulus (speech or frequency-specific).

Pure-tone audiometry (PTA): used for the children above 5 years old. This test provides

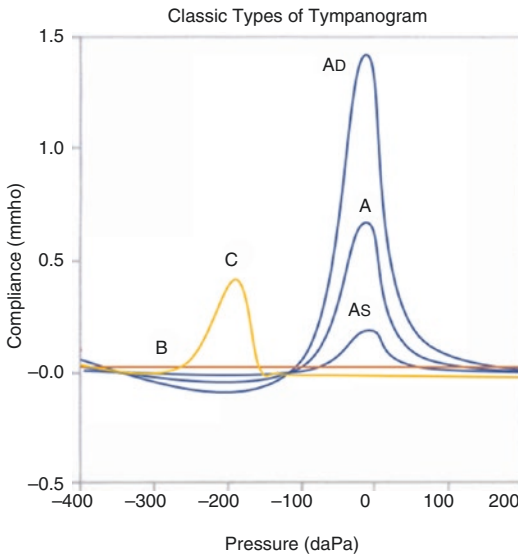


Fig. 66.2 Tympanogram types. *Type A:* normal patients, type AS: otosclerosis, otitis media, type AD: flaccid ear drums, ossicular interruptions. *Type B:* middle ear fluid, eardrum perforations, or impacted cerumen. *Type C:* Eustachian tube disorders, middle ear fluid

sound pressure level (SPL) in adult, but in newborns and infants less than 6 months corrected age 1000 Hz frequency probe tone is recommended.

2. Microphone.
3. Manometer which controls air pressure change within the ear canal.

Parameters in tympanometry include: static compliance, middle ear pressure, and ear canal volume. The values are reviewed in Table 66.3.

Three major types of tympanometry are identified: Type A, Type B, and Type C which are demonstrated in Fig. 66.2.

Table 66.5 Classification of hearing loss based on hearing threshold (dB HL)

Four commonly used systems of hearing loss classification			
System	WHO	BSA	ASHA
Normal	≤25	<20	-10 to 15
Slight	26-40		16-25
Mild		20-40	26-40
Moderate	41-60	41-70	41-55
Moderately severe			56-70
Severe	61-80	71-95	71-90
Severe to profound	≥81	>95	>90

WHO World Health Organization, BSA British Society Audiology, ASHA American Speech-Language Hearing Association

quantitative measures of air conduction (AC) and bone conduction (BC) thresholds. AC thresholds assess the entire auditory pathway using headphones or insert phones. BC testing stimulates the cochlea directly, thus bypassing the outer and middle ears. BC thresholds are measured by placing a bone vibrator on the skull (mostly mastoid). AC and BC thresholds are compared to get an idea about the status of conductive and sensory/neural systems. If AC and BC thresholds are elevated equally then the loss is sensorineural, while if AC thresholds are worse than BC thresholds with a gap more than 10 dB, then hearing loss is conductive. The pure-tone average is the mean of thresholds at 500, 1000, and 2000 Hz, which is the same frequencies referred to the speech frequencies. Tables 66.5 and 66.6 show the classification of hearing loss severity and its configuration [12].

Speech audiometry for pediatric age helps to determine the ability to perceive speech and check pure-tone threshold reliability. It includes speech detection threshold (SDT) and speech reception threshold (SRT) using developmentally appropriate spondee words or body-part identification.

66.2.2.3 Otoacoustic Emissions (OAE)

OAE are low-intensity sounds generated by the cochlea either spontaneously or evoked by an auditory stimulus and measured in the ear canal by-product of the active processes of the cochlear outer hair cells, which indicates mechanically active outer hair cells. They are frequency specific

Table 66.6 Criteria for classifying audiometric configurations

Term	Description
Flat	Less than 5 dB rise or fall per octave
Gradually falling	5-12 dB increase per octave
Sharply falling	15-20 dB increase per octave
Precipitously falling	Flat or gradually sloping, then threshold increasing at 25 dB or more per octave
Rising	Less than 5 dB decrease in threshold per octave
Peaked or saucer	20 dB or greater loss at the extreme frequencies, but not at the mid frequencies
Trough	20 dB or greater loss in the mid frequencies (1000-2000 Hz), but not at the extreme frequencies (500 or 4000 Hz)
Notched	20 dB or greater loss at one frequency with complete or near-complete recovery at adjacent octave frequencies

arising from the place on the cochlea’s basilar membrane responsible for processing that frequency.

Types of otoacoustic emissions:

- Spontaneous OAEs
 - Sounds emitted spontaneously without an acoustic stimulus.
 - Prevalence estimates vary (found in 40-50% of normal hearing).
 - Not generally used in clinical settings.
- Evoked OAEs
 - *Transient Evoked OAEs (TEOAEs)*: Sounds emitted in response to an acoustic stimulus, usually clicks but also tone bursts can be used.
 - *Distortion Product OAEs (DPOAEs)*: Sounds emitted in response to two simultaneous tones (F1, F2) of different frequencies and the response is a third tone, usually at 2F1-F2.
 - *Sustained Frequency OAEs (SFOAEs)*: Sounds emitted in response to a continuous tone. It is not much used for clinical purposes except for research.

OAEs are primarily analyzed in terms of signal-to-noise ratio (SNR), and the OAE

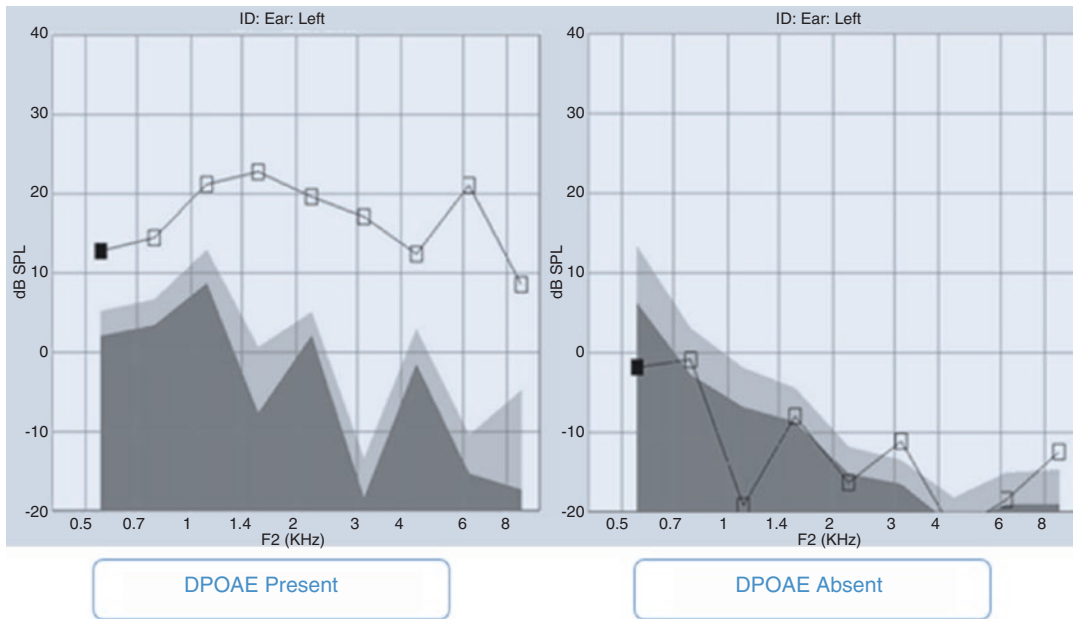


Fig. 66.3 Distortion Product OAEs (DPOAEs) response

response is how many dB above the “noise floor.” It is considered present if the response is 3–6 dB above the noise floor (Fig. 66.3). It is also considered present with regard to reproducibility, >70% reproducibility for TEOAEs.

The presence of the response indicates that the outer hair cells function is normal. Normal OAEs generally imply normal hearing, except in cases of auditory neuropathy and give indirect clue about middle ear function, while absent OAE suggests cochlear hearing loss >25–30 dB HL.

The advantages of otoacoustic emission test: It is an objective test, highly sensitive, highly dependent on outer hair cell function, relatively inexpensive; offers immediate response, ear-specific results; and does not require a sound-treated booth. On the other hand, it cannot estimate the type and degree of hearing loss, as OAEs are not a test of hearing.

A rule of thumb for comparing OAEs to hearing thresholds is that OAEs are present if thresholds are better than about 30 dB HL and absent if thresholds are poorer than 30 dB HL.

Clinical applications of otoacoustic emission test:

1. Newborns and infant screening.
2. Pediatric assessment.
3. Cochlear function monitoring.
4. Certain cases like ototoxicity and noise-induced hearing loss.

66.2.2.4 Auditory Brainstem Response (ABR)

An auditory evoked potential (AEP) is a waveform that reflects the electrophysiologic function of a certain portion of the central auditory nervous system in response to sound. An ABR is an objective test that elicits brain stem potentials in response to click or tone burst stimuli.

In response to the stimulus onset, the ABR waveform appears as several narrow peaks and troughs within 1–10 ms. The main positive peaks are labeled, in Roman numerals for Waves I, II, III, IV, V, VI, and VII. The following are the neural generators of the waves (Table 66.7).

ABRs can be performed via air conduction or bone conduction. ABRs can be affected by the subjects’ gender, age, temperature, and degree of hearing loss but are not acutely affected by the state of arousal or most sedatives anesthesia

Table 66.7 Auditory brainstem response waves

dWave number	Site
I	Distal VIIIth cranial nerve
II	Proximal VIIIth cranial nerve
III	Superior olivary complex
IV	Pons, lateral lemniscus
V	Midbrain, lateral lemniscus, and inferior colliculus
VI and VII	Undetermined (probably medical geniculate body and auditory cortex)

drugs. The ABR should be used in combination with other audiological procedures.

ABR can be used in different methods:

- *Neurologic ABR*: Evaluates the integrity of the auditory neural pathways, used as a diagnostic and screening tool to indicate auditory nerve and brainstem lesions and to rule out acoustic neuromas/vestibular schwannoma. It requires a high stimulus level of 80–90 dBnHL. A click stimulus generates a response from the 2–4 K Hz region of the basal portion of the cochlea [13]. Click ABR has a specificity of eighty percent for eighth nerve tumor detection [14]. ABRs are not known to be sensitive to small eighth nerve tumors.
- *Stacked ABR*: Enhances small eighth nerve tumor detection up to 95% sensitivity with 88% specificity.
- *Threshold ABR*: Assess the hearing thresholds in children, difficult-to-test patients, and non-organic hearing loss. The wave V peak is identified at each intensity level in a descending method with either a click or tone bursts stimuli until it is no longer identifiable. The click stimuli give an estimated hearing threshold from 1000 Hz through 8000 Hz frequency regions of the cochlea's greatest energy at 3000 Hz [15] while tone burst ABR uses a brief tone stimulus and gives frequency-specific information at 500, 1000, 2000, and 4000 Hz. The thresholds estimated from the ABR are typically higher when compared to behavioral thresholds. They can be as much as 20–30 dB higher, depending on the frequency; this why the corrected ABR is referred to as

estimated Hearing Level (eHL) to distinguish it from a typical ABR referenced in normalized Hearing Level (nHL).

A complete ABR test will provide the following information about each ear:

1. Absolute wave and interpeak latencies of all identifiable Waves I–V at different intensities.
2. Wave amplitudes (absolute and relative).
3. Threshold of Wave V if hearing threshold estimation was the purpose of the test.
4. A comparative response with a higher click rate when the ABR was for neurological assessment.

66.2.2.5 The Cochlear Microphonic (CM)

Pre-neural response from the cochlear outer hair cells (OHC) indicating normal OHC function. It follows the characteristics of the external stimulus. The direction of the cochlear microphonic will reverse with changes in the polarity of the stimulus. Responses obtained with positive (condensation) and negative (rarefaction) polarity stimuli show an inversion of the peaks of the cochlear microphonic waveform. CM testing is usually not necessary if OAE is reliably present. However, if OAE may be absent due to a conductive pathology, it is important to consider CM testing to exclude Auditory Neuropathy Spectrum Disorder (ANSO). CM cannot be used to estimate the hearing threshold.

66.3 Management of Hearing Loss

Early detection and prevention is the key to curb the effects of hearing loss. When the results are disclosed to the parents, they may experience a range of emotions, including shock, confusion, depression, anger, and guilt, while parents are still adjusting to being parents. Parents and healthcare professionals are faced with a series of dilemmas and challenges when it is established that a child is deaf.

Medical management consists of

- Developmental assessment
- Identification of coexisting medical conditions
- Extensive investigation to find the underlying cause of deafness

For the best child and family outcomes, an experienced empathic multidisciplinary team should work in a coordinated way with the family. Early intervention should be provided. Exposure to family support groups is very beneficial.

Early intervention services at a very early stage include hearing aids and a cochlear implant in the first 6 months of life, along with audio-verbal therapy with speech and language therapists and teachers of the deaf. Early intervention is a strong predictor of later outcomes, with appropriate early intervention, children with hearing loss can be mainstreamed in regular elementary and secondary education classrooms.

Cochlear implantation: Candidacy for cochlear implantation relies mainly on the audiological assessment. The basic criteria for implanting children include:

- Bilateral severe to profound sensorineural hearing loss (unaided PTA thresholds of ≥ 90 dB HL)
- Minimal benefit after at least 3–6 months of rehabilitation with well-fitted hearing aids
- No evidence of auditory nerve absence or central auditory lesions
- Have the strong support of his/her family for using oral language with a realistic expectation, and have access to a postoperative rehabilitation program for oral/aural language training [16, 17]

Communication methodologies in these cases include:

1. Auditory-Verbal Approach—based entirely on the use of audition and early amplification with hearing aids/cochlear implant for the development of spoken, receptive and expressive communication skills.

2. American Sign Language (ASL).
3. Aural/Oral Method—child's residual hearing is amplified along with auditory and speech reading training. The child's output is expected to be speech.

66.4 Strategies for Prevention of Hearing Loss

1. Strengthening relevant programs and organizations: Immunization programs to prevent many of the infections and maternal and child health programs.
2. Implementation of screening and intervention programs
 - A newborn hearing screening program
 - Implement school-based hearing screening
3. Training primary-level physicians and health-care workers about the need for early intervention to address hearing loss
4. Making appropriate technologies accessible
5. Regulation and monitoring of ototoxic medication use
6. Raising awareness via programs targeting children community awareness

Take Home Messages

- Early detection and prevention is the key to curb the effects of hearing loss.
- The newborn hearing screening program aims to identify all children with a moderate–profound PCHI in the better hearing ear.
- Hearing assessment for children can be objective or subjective, and it varies according to age.
- Absent otoacoustic emissions (OAE) suggests cochlear hearing loss >25 – 30 dB HL.
- Auditory brainstem response (ABR) audiometry is not acutely affected by the state of arousal or most sedatives anesthesia drugs.

References

1. Tharpe AM, Seewald R. *Comprehensive handbook of pediatric audiology*. 2nd ed. San Diego, CA: Plural Publishing, Inc.; 2016.
2. Shearer AE, Smith RJH. Genetics: advances in genetic testing for deafness. *Curr Opin Pediatr*. 2012;24(6):679–86.
3. Hilgert N, Smith RJH, Van Camp G. Function and expression pattern of nonsyndromic deafness genes. *Curr Mol Med*. 2009;9(5):546–64.
4. Saloojee HS, Velaphi Y, Goga N. Congenital syphilis: an overview and recommendations. *Bull World Health Organ*. 2004;82(6):424–30.
5. Bamford J, Ankjell H, Crockett R et al.. Evaluation of the newborn hearing screening programme in England: studies, results and recommendations. Report to Department of Health and National Screening Committee. 2005.
6. Public Health England. NHS Screening Programmes in England: April 2015 to 31 March 2016. <https://www.gov.uk/government/publications/nhs-screening-programmes-annual-report>.
7. British Society of Audiology. NHSP audiology protocols and guidance. <http://www.thebsa.org.uk/resources/>.
8. Joint Committee on Infant Hearing. Year 2007 position statement: principles and guidelines for early hearing detection and intervention programs. *Pediatrics*. 2007;102:893–921.
9. Martin FN. *Introduction to audiology*. 11th ed. New York: Pearson; 2017.
10. Hunter LL, Margolis RH. Multifrequency tympanometry. *Am J Audiol*. 1992;1(3):33–43.
11. Paradise JL. Tympanometry controversy and middle ear effusion: too much concern or too little? *Pediatrics*. 1976;58(4):626–7.
12. Katz J, Chasin M, English K, Hood LJ, Tillery KL, editors. *Handbook of clinical audiology*. 7th ed. New York: Wolters Kluwer Health. 2014.
13. Hall JW, Mueller HG. *Audiologists' desk reference*. Vol. I: Diagnostic audiology principles, procedures, and practices. San Diego, CA: Singular Publishing Group; 1997. p. 395.
14. Burkard RF, Don M, Eggermont JJ, editors. *Auditory evoked potentials: basic principles and clinical application*. Baltimore, MD: Lippincott Williams & Wilkins; 2007.
15. Hall III. *New handbook of auditory evoked responses*. New York: Pearson; 2007.
16. Zyfter W, Karlik M, Sekula A, Harris S, Gawęcki W. Current indications for cochlear implantation in adults and children. *Otolaryngol Pol*. 2019;73(3):1–5.
17. Wackym PA, Tran A. Chapter 15: Cochlear implantation: patient evaluation and device selection. In: Flint PW, Haughey BH, editors. *Cummings otolaryngology: head & neck surgery*. 6th ed. Philadelphia: Elsevier Inc.; 2015. p. 2428–43.