RESEARCH PAPER | Medical Science | Volume : 5 | Issue : 3 | March 2015 | ISSN - 2249-555X

Hearing loss in the newborn infant

KEYWORDS: OAE:Otoacoustic emission , ABR;Auditory brainstem response

Dr Arvind Saili	Dr Savita Jha	Dr Babu Madarkar
Director Professor & HOD Deptt of Neonatology LHMC New Delhi	Professor of Obstetrics & Gynaecology Deptt of Obstetrics & Gynaecology DJ Institute of Medical Sciences & Research UP,	DM Resident Deptt of Neonatology Kalawati Saran Children's Hospital New Delhi

ABSTRACT *Hearing impairment in children is a serious hurdle against their optimal development. Hearing-impaired infants may suffer from delayed development of speech, language and cognitive skills, resulting in learning disability at the school. Auditory development has a critical stage during the early childhood when it has to be nurtured. Adequate support has to be provide during this period to prevent the impairment of auditory neuropathway of children at a later developmental stage . Hence, early detection is very important element in providing appropriate support the hearing-impaired babies that will help them enjoy equal opportunities in society .*

A two-stage screening protocol is often followed, in which otoacoustic emissions (OAE) is used to screen the babies .Those who fail the OAE are screened with auditory brainstem response (ABR). This two stage screening program (the second stage being ABR, which is costly) is required only for minimal few children .Thus this method makes the program more practical .

The "Consensus Statement on Early Identification of Hearing Impairment in Infants and Young Children in 1993." states that all babies admitted to the neonatal intensive care unit (NICU) should be screened for hearing loss before discharge and implementation of universal hearing screening should be done in initial three months of life. A nearly 2%(0.3%-6.5%) of neonates fail the hearing screen (Abdala,1997). Of these about 2 per thousand babies suffer from permanent hearing loss. Congenital hearing loss is more common than acquired hearing loss (Coplan J,1993) .If the hearing loss not identified in time it adversely affects development of two way communication ,comprehension academics , social and emotional development,

(Mayne AM,1998) ,(Mayne AM,1998) when hearing loss is identified and intervened in right time especially within first six months ,it provides definitive benefit in overall development(Yoshinaga-Itano C,2004) The current evidence says that the central nervous system is highly responsive to language input early part of life (Sharma A,2002), (Sharma A,2002).

The Joint Committee on Infant Hearing 2007 recommends that to improve the outcomes in babies with any grade of hearing loss

- All neonates should undergo a hearing screen in the first month of life.
- Those babies who fail in the first screen should undergo detailed evaluation by an audiology expert before the completion of three months.
- Those babies who have confirmed hearing loss, should receive expert intervention before six months of age .

Normal hearing

The ear is divided in to outer, middle, and inner parts. The outer ear is composed of the pinna and ear canal. Sound passes through the outer ear canal and hits the tympanic membrane. The vibrations from the tympanic membrane travel in to middle ear and get amplified and then transmitted within the ossicles to reach the fluid inside the cochlea (inner ear). Sound stimulates hair cells within the cochlea. The outer hair cells in response ,produce otoacoustic emissions which are basically an echo of sounds, Inner hair cells convert mechanical energy into electrical energy which gets transmitted through the cochlear branch of the 8th cranial nerve, then through the brainstem, ultimately in to cerebral cortex for comprehension of words.

Hearing loss

Any interruption in the transmission of sounds from outer ear to inner ear results may lead to a temporary (fluid or debris) or permanent (anotia or microtia) conductive hearing loss. When sound is not able to get transmitted through cochlea, outer and inner hair cells, and eighth cranial nerve ,then it leads to sensorineural hearing loss. The term 'Auditory dyssynchrony' is used to describe a situation where in inner hair cells and 8th cranial nereve suffer from pathology but the outer hair cells are functioning intact

Type	Characteristics
Sensorineural hearing loss	The sound energy is not able to reach the brain stem due to abnormality in outer and inner hair cells and the 8th cranial nerve
Permanent conductive hearing loss	Failure of transmission of sounds due to structural obstruction of outer ear(anotia,microtia) or the middle ear(fusion of ossicles)
Auditory dyssynchrony	The outer hair cells are functionally normal The abnormality in the 8th cranial nerve and the inner hair cell prevent the transmission of sounds to the brain stem causing hearing loss..
Transient conductive	Secretions and the solid debris prevent the movement of the sound in to the inner ear
Mixed hearing loss	It is a combination of sensorineural or neural hearing loss

Volume : 5 | Issue : 3 | March 2015 | ISSN - 2249-555X

Tests for hearing loss

The two most preferred test are otoacaustic emissions (OAE)and auditory brain stem response(ABR) .These tests measure electrical potential difference. The advantage of these tests are that they do not need an active response from the baby and they can be done on a sleeping baby. In OAE hearing screen, a sound stimulus is given to the baby which reaches the inner ear, in response of which outer hair cells produce certain vibrations which are further picked up by the highly sensitive microphone (present within the probe) The two main interferences with OAE recordings are conduction system blockage and back ground noise. In Auditory brainstem response (ABR) a sound stimulus in the form of a click is given to the ears. The surface electrodes placed over the scalp pick up neuroelectrical activity in a sequential manner from cochlea, outer hair cells ,inner hair cells ,auditory nerve, and brain stem . In automated auditory brain stem response (AABR),there is preset algorithm which decides whether a screening test is pass or fail based on presence or absence of wave 5 respectively.. Both OAE and ABR can detect both sensorineural and conductive hearing loss. A false-positive fail screen for permanent hearing loss may result from non functioning outer or middle ear ,and any noise obstruction . Babies with sensory hearing loss fail the ABR screening but pass the OAE screening. In sensorineural hearing loss there is defect in either of inner hair cells, 8th Cranial nerve, or brain stem but the outer hair cells are normal, hence OAEs should not be used to screen the sensory hearing loss.

Table-2.Screening and Diagnostic methods for hearing loss

Method	Mechanism	Detection
Otoacoustic emissions (OAE) screen	THE Echo like response produced by the outer hair cells to the measured sound stimulus is picked by a microphone placed within the probe. An automated version of equipment is also available .	Sensorineural Conductive
Automated auditory brainstem response screen	These tests have become standards currently for detection of hearing loss. They are dependent on wave5 based threshold aldorithms	Sensorineural Conductive Neural
Battery of diagnosisfor detection of hearing loss:		
Auditory brainstem response diagnostic	Electrodes placed over the scalp detect the neuronal electrical activity of 8th cranial nerve ,auditory pathway,	Sensorineural Conductive Neural
Tympanometry battery	This test measures function of middle ear. For babies smaller than six months 1000 Hz is used	Conductive
Vision reinforcement audiometry (>6 months of age) Conditioned audiometry response (>2.5 years of age)	Sysytematic observation of babysbehavioral responses to various sound stimulus	Sensorineural Conductive Neural
Standard audiometry (>4.5 years of age)	Observation of the child's behavioral responses to a task in response to sounds	Sensorineural Conductive Neural

Tympanometry testing also is called as immittance testing assesses functionality, structural adequacy, , and movable property of the tympanum , middle ear pressure, and the ossicle continuity and mobility.. A probe is placed in the inner ear, and air pressure is changed to assess the movement of the tympanic membrane. The tympanogram shows the response of the tympanic membrane in response to the pressure stimulus: a type A curve is a normal response. A flat curve response is due to presence of fluid in the middle ear or due to structural discontinuation of tympanic membrane. Tympanometry is not used for screening.

Behavioral tests like vision reinforcement audiometry (VRA), are usually tested for infants more than 6 months of age. The baby should be able to turn to sound. During the VRA testing, the baby rests comfortably in the mothers lap in a testing booth. There are animated toys placed either side of the baby and the baby is conditioned to the sound stimulus earphones are placed, and the baby's response of turning to sound stimulus from the animated toys is noted

Early intervention services

Currently there is strong evidence which supports the importance of early screening and treatment, improving the outcome of hearing loss in children. Before the implementation of universal hearing screening, the children with more than severe hearing loss were identified at two to three years of age to have delay in in communication, language, and literacy (Mayne AM,1998),(Mayne A M,1998). The Colorado study demonstrated that if the intervention is done before six months of age, it improves the outcome in the form of better speaking, sign, or total communication scores when assessed at 3 years of age. (Yoshinaga-Itano C,2004),(Vohr B,2008) The Joint Committee on Infant Hearing 2007 recommends that infants with all degrees of unilateral or bilateral hearing loss need to be referred to Early Intervention Services at the time of diagnosis, before 6 months of age.

Etiology of hearing loss

It is estimated that at least 50% of congenital hearing loss is hereditary. There are nearly 400 syndromes and hundreds of genes associated with hearing loss that have been identified(Nance WE,2006).Genetic hearing loss is about 30% syndromic and 70% nonsyndromic. Among children with nonsyndromic hearing loss, 75% to 85% of cases are autosomal recessive (DFNB), 15% to 24% are autosomal dominant (DFNA), and 1% to 2% are X-linked (DNF). Therefore, most infants with hearing loss have nonsyndromic autosomal recessive hearing loss and are born to hearing parents. A single gene, GJB2, which encodes connexin 26, a gap-junction protein expressed in the connective tissues of the cochlea, accounts for up to 50% of all cases of profound nonsyndromic hearing loss. More than 100 mutations of GJB2 have been identified. A single GJB2mutation, 35delG, accounts for up to 70% of the mutations.

Risk Factors Associated with Permanent Congenital, Delayed Onset, or Progressive Hearing Loss

1. 1.Caregiver concerns regarding , speech, language, or developmental delay
2. 2.Positive Family history of hearing loss
3. 3.Neonatal intensive care stay for >5 days, which includes ECMO, ventilation, ototoxic medications .Eg,gentamicin and Hyperbilirubinemia

Volume : 5 | Issue : 3 | March 2015 | ISSN - 2249-555X

4. 4.Neurodegenerative disorders -Hunter syndrome ,Friedrich ataxia or Charcot-Marie-Tooth disease
5. 5.postnatal infections (herpesviruses and varicella) meningitis.
6. 6.Head trauma, basal skull or temporal bone fracture, requiring hospital admission
7. 7.Cancer Chemotherapy
8. 8.In utero infections, such as rubella, syphilis, and toxoplasmosis cytomegalovirus,herpesvirus,
9. 9.Anatomical anomalies of ear, and temporal bone
10. 10.Physical findings, of syndromes causing hearing loss
11. 11. Syndromes associated with hearing loss or progressive or late-onset hearing loss, such as neurofibromatosis, osteopetrosis, and Usher syndrome Waardenburg, Alport, Pendred, and Jervell and Lange-Nielson syndromes.

Risk factors

It is also often seen that the parents themselves are not aware of the positive family history of hearing loss. In circumstances where there is positive family history of hearing loss, but the baby passes the hearing screen, it is advisable to continue surveillance with minimum one audiologic assessment by two to three years of age . The second risk factor which is care giver concern , regarding hearing, speech,social, language, delay in the initial three years of life,(Centers for Disease Control and Prevention,2010) is also associated with an increased risk for progressive hearing loss. The rate of hearing loss almost triples between birth and school age from 2 to 3 per 1000 in newborns to about 7 per 1000 at school age,(Roizen NJ,1999)Cytomegalovirus is the most common cause of hearing loss in infants and children(Johnson JL,2005). Most infants with congenital cytomegalovirus infection have no clinical findings at birth, and the diagnosis often goes unrecognized.

Medical workup for hearing loss and care coordination

The primary care physician, needs to be aware of community resources and support the family choice of early intervention program and mode of communication.

Every infant with confirmed hearing loss should be evaluated by an otolaryngologist with knowledge of pediatric hearing loss. The otolaryngologist conducts a comprehensive assessment to determine the etiology of hearing loss and provides recommendations and information to the family, audiologist, and primary care provider on candidacy for amplification, assistive devices, and surgical intervention, including reconstruction, bone-anchored hearing aids, and cochlear implantation.

Because of the prevalence of hereditary hearing loss, all families of children with confirmed hearing loss should be offered a genetics evaluation and counseling. This evaluation can provide families with information on etiology, prognosis, associated disorders, and the likelihood of hearing loss in future offspring. The geneticist will review the family history for specific genetic disorders or syndromes, examine the child, and complete genetic testing for syndromes or gene mutations for nonsyndromic hearing loss such as GJB2(connexin 26)(Fowler KB,1997).

Because 30% to 40% of children with confirmed hearing loss have comorbidities or other disabilities, the primary care physician should closely monitor developmental milestones and initiate referrals related to suspected disabilities as needed(Santos RL,2005) Because of the association of hearing loss with vision impairments and the importance of vision for children with hearing loss, it is recommended that each child with a permanent hearing loss have at least one examination to assess visual acuity by an ophthalmologist experienced in evaluating infants.

Middle ear disease

Otitis media with effusion (OME) is highly prevalent among young children, and about 90% of children have an episode of OME before starting school(Karchmer MA, 1999). Middle ear status should be monitored closely in children with permanent hearing loss because the presence of middle ear effusion can further compromise hearing. Recommendations related to diagnosis of OME include examination with a pneumatic otoscope and documentation of laterality, duration of effusion, and severity of symptoms. Although about 40% to 50% of children with OME do not have symptoms(Tos M,1984), some children may have associated balance problems(Rosenfeld RM,1984). Medical management of OME in children with permanent hearing loss may include hearing testing, amplification adjustment, and tympanostomy tubes. Most OME is self-limited(Golz A,1984), and 75% to 90% of cases spontaneously resolve in 3 months. Therefore, a 3-month period of observation is recommended. Evidence suggests that no benefit is derived from the use of antihistamines or decongestants in children.

Hearing AIDS, frequency modulated systems, and cochlear implants

Hearing AIDS

Hearing aids are compact and worn either in-the-ear (ITE) or behind-the-ear (BTE), and can be fitted on an infant in the first month of life. The main components are the microphone that picks up sounds and the amplifier. If the child has different degrees of hearing loss at different frequencies, the audiologist adjusts the gain (loudness) by frequency. Normal speech range is from 500 to 2000 Hz. Ear molds are made from an impression of the child's ear. As a young infant grows, the ear molds may need to be replaced every 6-8 weeks.

Frequency modulated systems

Frequency modulated (FM) systems are designed to be used in patients with hearing loss to hear better in noisy environments. AFM system is made up of a microphone and a receiver. A small radio transmitter is attached to a microphone and a small radio receiver. the guardian wears the FM transmitter and microphone while the baby wears the FM receiver. The FM transmitter sends a low-power radio signal to the FM receiver that needs to be within 50 feet of the transmitter. The FM receiver gets the signal from the microphone and sends it to a personal hearing aid or cochlear implant. Listening to the FM signal is similar to listening to speech only inches away.

Cochlear implants

Candidacy criteria for pediatric cochlear implantation currently is 18 months or older for children with severe to profound bilateral sensorineural hearing loss and 12 to 18 months for children with profound hearing loss. In cases of deafness due to meningitis, implants may be placed early in the first year of life. Children up to 7 years of age appear to derive the greatest benefit from a cochlear implant for the development of speech(Rosenfeld RM,2003).

Volume : 5 | Issue : 3 | March 2015 | ISSN - 2249-555X

Continued surveillance

Regular surveillance of developmental milestones, auditory skills, parental concerns, and middle ear status should be performed in the medical home.1 All infants should have an objective standardized screen of global development with a validated screening tool at 9, 18, and 24 to 30 months of age or at any time if the health care professional or family has concern. Language screens used in the primary care setting include the Early Language Milestone Scale (Coplan J,1993), the MacArthur Communicative Development Inventory(American Academy of Pediatrics,2006) the Language Development Survey (Rescoria L,1989) and Ages and Stages(Bricker D, 1999).

Stress and impact on the family

Parents perceive varying degrees of stress when they learn that their infant has failed a newborn hearing screen. Although the screen result may be either a false-positive or a true fail, most parents will have some increase in worry until their infant is rescreened. In one study (Vohr BR,2001) of well-baby nursery infants, parents reported increased "worry" at 2 to 8 weeks of age when they returned for the rescreen. Physicians who understand the screening process can support the family whose infant fails the screen, encourage the family to return for the rescreen, and follow-up with the family about the rescreen results. A second study(Vohr BR,2008) reported that mothers of infants with a false-positive screen did not report increased levels of stress or impact at 12 to 16 months or at 18 to 24 months. In addition, greater family resources were protective against persistent stress, whereas NICU stay contributed to prolonged stress(Vohr BR,2008).

REFERENCE

1. Abdala,deUzcategu C, Yoshinaga-Itano C(1997): Parents' reactions to newborn hearing screening. Audiology Today.9 (1):24 . | 2. Coplan J(1993): Early language milestone scale. 2nd ed ,Pro-ed Austin, TX. | 3. Mayne AM(1998): Expressive vocabulary development of infants and toddlers who are deaf or hard of hearing. Volta Rev. 100:1 . | 4. Mayne AM(1998): Receptive vocabulary development of infants and toddlers who are deaf or hard of hearing. Volta Rev. 100:29 . | 5. Yoshinaga-Itano C(2004): Levels of evidence. universal newborn hearing screening (UNHS) and early hearing detection and intervention systems (EHDI) J Commun Disord. 37:4. | 6. Sharma A(2002): A sensitive period for the development of the central auditory system in children with cochlear implants. implications for age of implantation Ear Hear.23:532. | 7. Sharma A(2004): Central auditory maturation and babbling development in infants with cochlear implants. Arch Otolaryngol Head Neck Sur. 130:511. | 8. Joint Committee on Infant Hearing (2007): Year 2007 position statement. principles and guidelines for early hearing detection and intervention programs Pediatrics. 120:898. | 9. Vohr B (2008): Early language outcomes of early-identified infants with permanent hearing loss at 12 to 16 months of age. Pediatrics. 122:535 . | 10. Nance WE (2006): Importance of congenital cytomegalovirus infections as a cause for pre-lingual hearing loss. J Clin Virol. 35:221 . | 11. Centers for Disease Control and Prevention(2010): Early Hearing Detection and Intervention (EHDI) | Program.(website) www.cdc.gov/ncbddd/ehdiMay, 2010Accessed | 12. Roizen NJ(1999): Etiology of hearing loss in children. Nongenetic causes. Pediatr Clin North Am. 46:49. | 13. Johnson JL(2005): A multisite study to examine the efficacy of the otoacoustic emission/automated auditory brainstem response newborn hearing screening protocol.introduction and overview of the study Am J Audiol.14:S178 | 14. Fowler KB(1997): Progressive and fluctuating sensorineural hearing loss in children with asymptomatic congenital cytomegalovirus infection. J Pediatr. 130:624. | 15. Santos RL(2005).: Hearing impairment in Dutch patients with connexin 26 (GJB2) and connexin 30 (GJB6) mutations. Int J Pediatr Otorhinolaryngol. 69:165 . | 16. Karchmer MA, Allen TE(1999): The functional assessment of deaf and hard of hearing students. Am Ann Deaf. 144:68 | 17. Tos M(1984): Epidemiology and natural history of secretory otitis. Am J Otol. 5:459 . . | 18. Rosenfeld RM(1997): Quality of life for children with otitis media. Arch Otolaryngol Head Neck Surg. 123:1049. | 19. Golz A(1998) : Evaluation of balance disturbances in children with middle ear effusion. Int J Pediatr Otorhinolaryngol. 43:21. | 20. Rosenfeld RM, Kay D(2003): Natural history of untreated otitis media. Laryngoscope. 113:1645 . | 21. Sharma A(2005): The influence of a sensitive period on central auditory development in children with unilateral and bilateral cochlear implants. Hear Res. 203:134. | 22. American Academy of Pediatrics(2006): Policy statement.identifying infants and young children with developmental disorders in the medical homean algorithm for developmental surveillance and screening Pediatrics.118:405 . | 23. Fenson L, Dale PS, Reznick JS(1993): The McArthur communicative development inventories. user's guide and technical manual Singular: Thomson Learning San Diego. | 24. Rescoria L(1989): The Language Development Survey. a screening tool for delayed language in toddlers J Speech Hear Dis. 54:587 . | 25. Bricker D, Squires J(1999): Ages and stages questionnaires. a parent-completed, child-monitoring system Paul H. Brookes Baltimore. | 26. Vohr BR(2001): Maternal worry about neonatal hearing screening. J Perinatol. 21:15 . | 27. Vohr BR(2008): Results of newborn screening for hearing loss. effects on the family in the first 2 years of life Arch Pediatr Adolesc Med. 162:205.