Consensus Statement of the Indian Academy of Pediatrics on Newborn Hearing Screening

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Justification: Hearing impairment is one of the most critical sensory impairments with significant social and psychological consequences. Evidence-based, standardized national guidelines are needed for professionals to screen for hearing impairment during the neonatal period.

Process: The meeting on formulation of national consensus guidelines on developmental disorders was organized by Indian Academy of Pediatrics in Mumbai, on 18th and 19th December, 2015. The invited experts included Pediatricians, Developmental Pediatricians, Pediatric Neurologists and Clinical Psychologists. The participants framed guidelines after extensive discussions.

Objective: To provide guidelines on newborn hearing screening in India.

Recommendations: The first screening should be conducted before the neonate's discharge from the hospital – if it 'fails', then it should be repeated after four weeks, or at first immunization visit. If it 'fails' again, then Auditory Brainstem Response (ABR) audiometry should be conducted. All babies admitted to intensive care unit should be screened *via* ABR. All babies with abnormal ABR should undergo detailed evaluation, hearing aid fitting and auditory rehabilitation, before six months of age. The goal is to screen newborn babies before one month of age, diagnose hearing loss before three months of age and start intervention before six months of age.

Keywords: *Assessment, Auditory brainstem response audiometry, Deafness, Hearing loss, Otoacoustic emission, Prevention.*

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Hearing impairment is one of the most critical sensory impairments with significant social and psychological consequences. Failure to detect children with congenital or acquired hearing loss may result in lifelong deficits in speech and language acquisition, poor academic performance and personal-social and behavior problems [1,2]. Deficits in speech and language lead to lack of stimulation, which adversely affects the structure of the synaptic junction. Lack of auditory stimulation leads to retrograde degeneration in the cell body and axon [3].

Apart from the biological evidence, the data on congenital disabilities indicate that hearing loss has a substantially high incidence with congenital hearing loss affecting 30 per 10,000 children [4]. Significant hearing loss is the most common disorder, occurring in 1 to 2 newborns per 1000 in the general population, and 24% to 46% of newborns admitted to neonatal intensive care unit [5,6]. Vocabulary of a 3-year-old child with hearing impairment if remediated at birth is 300-700 words; if remediated at 6 months is 150-300 words and if remediated at 2 years is 0-50 words, respectively; as compared to vocabulary of a 3-year-old child with typical hearing which is 500-900 words.

In view of the above, standard guidelines for screening newborns for hearing loss are urgently needed. The meeting on formulation of National consensus guidelines on developmental disorders was organized by the Indian Academy of Pediatrics in Mumbai, on 18th and 19th December, 2015. The invited experts included Pediatricians, Developmental Pediatricians, Pediatric Neurologists and Clinical Psychologists. The participants framed guidelines after extensive discussions and review of literature. Thereafter, a committee was established to review and finalize the points discussed in the meeting.

Subsequent sections include the points of consensus on screening of newborn hearing.

RECOMMENDATIONS

Early Screening

Critical period for identification and remediation of hearing loss is before the age of 6 months. Since the pediatrician is the primary care provider for the child during the first few days of life, it is the sole responsibility of the pediatrician (or the primary physician) to evaluate the child for hearing loss (or ensure referral for the same). It has been observed that practice of neonatal screening has dramatically lowered the age of diagnosis of deafness from 1½ - 3 years to less than 6 months of age. Screening should ideally be 'universal' i.e., everybody is screened and at a minimum, screening should be 'targeted' i.e., 'high risk' babies are screened.

Causes of hearing loss are summarized in ***Box* 1**. These can be classified as: Conductive, Cochlear (i.e. Sensory: defect in the cochlea and Neural: defect in the 8th cranial/ auditory nerve), Retrocochlear (i.e. defect at the level of auditory nerve, brainstem auditory pathway or both) and Central (i.e. defect in the auditory area in cerebral cortex).

With respect to current guidelines, sensorineural hearing loss is most relevant and cochlear causes of sensorineural hearing loss are more common. Many risk factors for hearing loss have been identified and are listed in ***Box* 2**.

Congenital rubella syndrome, Usher syndrome and Jervell and Lange-Nielsen (JLN) syndrome have been noted to be associated with hearing loss; few other syndromes include Treacher-Collins syndrome, Apert syndrome, Alport syndrome, Neurofibromatosis syndrome, Achondroplasia, CHARGE syndrome, Brachio Oto Renal syndrome, Chudley McCullough syndrome and Golden Har syndrome [8].

BOX 1 CAUSES OF HEARING LOSS

- Causes in ear canal/Conductive (*e.g.*, congenital atresia, wax, foreign body, trauma, external otitis, stenosis
- Causes in middle ear/Conductive (*e.g.*, acute and chronic otitis media, perforation of tympanic membrane, congenital defects, trauma, malformations either hereditary or familial)
- Causes in the cochlea/Cochlear (*e.g.*, ototoxic drugs, stay in neonatal intensive care unit due to jaundice or other causes, neonatal infections, head injury, noise); and
- Causes in auditory nerve/Retrocochlear (*e.g.,* problems in cochlear nerve, auditory pathway or cortex like tumors, trauma, de myelination).
- Intrauterine infections (tetanus, toxoplasma, rubella, cytomegalovirus and herpes or TORCH group of infections) can be classified as cochlear or retrocochlear causes of Sensorineural hearing loss.

Screening for Newborn Hearing Loss

In India, majority of hospitals do not conduct universal or high risk screening. In such a situation, a centralized facility catering to all hospitals in a city is a practical option. A two-stage screening protocol can be made, in which infants are screened first with otoacoustic emissions (OAE). Infants who fail the OAE are screened with auditory brainstem response (ABR). In this two tier screening program, the second tier being ABR (which is more expensive) is required only for a select few, making the program more practical and viable.

The Child Health Screening and Early Intervention Services Program (Rashtriya Bal Swasthya Karyakram) under National Rural Health Mission initiated by the Ministry of Health and Family Welfare of Government of India has included congenital deafness as one of the conditions to be included for early identification and remediation. It involves screening of infants and children under age 18 years by a mobile team and provision of appropriate treatment at District Early Intervention Centres (DEICs). This ambitious scheme is likely to streamline the management of hearing disabilities [9].

Otoacoustic emissions (OAEs) are quicker methods (as compared to electrophysiologic methods like ABR) for assessing hearing in newborns *via* a simple set-up. Otoacoustic emissions (OAEs) are sounds of cochlear origin recorded in the auditory meatus (ear canal), produced by the action of healthy outer hair cells. The

BOX 2 RISK FACTORS FOR HEARING LOSS [7]

- Family history of hereditary childhood sensorineural hearing impairment
- Intrauterine infection (TORCH)
- Craniofacial anomalies
- Birth weight less than 1500 gram
- Hyperbilirubinemia at a serum level requiring exchange transfusion
- Ototoxic medications used in multiple courses, or in combination with loop diuretics.
- Bacterial meningitis
- APGAR scores 0-4 at 1 minute or 0-6 at 5 minute
- Mechanical ventilation for 5 days or longer
- Stigmata of other findings associated with a syndrome known to include sensorineural and/or conductive hearing loss.

emissions themselves serve no purpose and are simply a leakage of energy from the ear. Hearing is facilitated by hair cell activity in the cochlea and more specifically, the activity of outer hair cells. There are three rows of outer hair cells (OHCs) and one row of inner hair cells that sit on the basilar membrane, sandwiched by the tectorial membrane on top. This forms the organ of Corti. There are around 12000 motile OHCs working together to provide mechanical assistance to sound energy, amplifying the travelling wave to overcome the viscous nature of the cochlear fluid. As the 'W' shaped steriocilia are stimulated by fluid moving over them, it causes the cells to alternately contract and release, providing a pumping action. This mechanical system provides the frequency tuning within the cochlea. The inner hair cells are also stimulated and deflected by fluid flow; and at a specific threshold, the inner cells release a neurotransmitter which causes the auditory nerve to transmit a signal to the brain.

Cochlear damage is almost always apparent in the loss of outer hair cells. This is true regardless of the etiology – congential progressive hearing loss, ototoxic drugs, presbyacusis (Sensorineural hearing loss with aging), as well as noise-induced hearing loss. With damaged OHCs, there is no amplification or frequency tuning, thus the child will not only suffer a threshold shift but also have problems with frequency discrimination.

OAE test is performed *via* a small probe placed in the child's ear canal; click sounds are delivered and response is detected (***Web Appendix* 1**). The child must be quiet [10].

Recommendations on Screening

- A two-stage screening protocol with OAE as the first screen, followed by ABR for those who fail the OAE screen [11].
- It is advisable that all hospitals with level-3 neonatal care have OAE and ABR facilities. If not feasible, a centralized hearing screening with a portable OAE is suggested and all abnormal cases can be referred for ABR to the nearest centre.
- The program is to be coordinated by an audiologist and weekly assessment meeting is to be convened with the staff to discuss and sort out the issues, if any (held by the convenor). Usual issues could include non-compliance by parents to bring the child for repeat OAE or ABR. This usually can be tackled by phone calls made by screening personnel, coordinator, or in rare instances by the convenor himself. A medical social worker can be involved for problem-solving.
- Personnel with basic knowledge in computer and good communication skills are chosen. They should be provided basic training in hearing screening and also skills to gather information on high-risk criteria, if any, from parents/hospital staff/hospital records. This training is to be conducted over one day.
- The screening personnel should visit each hospital daily/on alternate days/twice a week/weekly depending upon the number of births in that particular hospital. Daily screening may be carried out in hospitals which have more than 200 births, alternate day screening in hospitals with 100-200 births and twice weekly or weekly screening in hospitals with births less than 100 per month.
- All screeners should maintain a register of all cases screened and those with abnormal results. Neonates with abnormal screening results should be evaluated. It is the duty of the screeners to call back all abnormal cases for follow up, with the help of a coordinator. (Number of hospitals covered by a screener depends on the number of cases in a particular hospital and proximity of the hospitals)
- If abnormal OAE is detected, it is repeated at 6 weeks on the 1st immunization visit. If again abnormal, ABR is done for confirmation followed by full audiological evaluation and remediation with hearing aids (cochlear implant may be required in cases of profound hearing loss or poor response to hearing aids).
- All NICU babies undergo ABR testing to rule out auditory dyssynchrony/ auditory neuropathy.
- In babies with abnormal ABR, detailed enquiry is made to identify and record any risk factors. Any baby missing screening before hospital discharge is called for OAE test on the first immunization visit.
- All babies with abnormal ABR should undergo detailed ENT evaluation hearing- aid fitting and auditory rehabilitation before 6 months of age. Systematic evaluation for ruling out syndromic associations such as ophthalmic, paediatric and cardiac assessments should be conducted.
- Children with neonatal meningitis should be treated as a special category and need investigations including imaging and intervention like cochlear implant (if needed) on a semi-emergency basis. Delay can result in cochlear ossification which may preclude subsequent intervention like a cochlear implant.

The goal is to screen newborn babies before 1 month of age, diagnose hearing loss before 3 months of age and start intervention before 6 months of age. Hurdles

KEY MESSAGES

- Hearing loss should be screened preferably before 1 month of age.
- Universal neonatal screening rather than targeted 'high risk' screening is ideal.
- If abnormal OAE detected, it is repeated at 6 weeks or on the first immunization visit. If again abnormal, ABR is done for confirmation followed by full audiological evaluation and remediation with hearing aids.
- All NICU babies should undergo ABR testing to rule out auditory dys-synchrony/ auditory neuropathy.

experienced in the screening process include: less motivated pediatricians; lack of awareness among parents/ community; non-compliance by the family for evaluation, and stigma attached to hearing aids.

CONCLUSIONS

As normal hearing is critical for speech and language development, it is recommended that during first 6 months of life, clinicians identify infants with hearing loss, preferably before 3 months of age. Other important issues are:

- Evaluate infants before discharge from nursery, especially high risk babies
- Universal neonatal screening and not targeted 'high risk' screening is ideal since about 50% of infants with hearing loss have no known risk factors for hearing loss and are discharged from well-baby nursery
- Delayed onset hearing loss should be considered and followed up (if presence of language delays, infections, head trauma, stigmata of syndromes, ototoxic medications, recurrent otitis media, intrauterine infections, neurofibromatosis type II)
- Prevalence of hearing loss is more than twice that of the other newborn disorders combined, which can be screened
- Never delay hearing assessment in a suspected case; no child is too young to be tested or too young to be evaluated
- Never resort to rudimentary tests of hearing (like clapping hands) as confirmatory tests, and reassure parents that their child's hearing is normal.

Universal Newborn Hearing Screening (UNHS) has become a standard practice in most developed countries. The identification of all newborns with hearing loss before six months has now become an attainable and realistic goal. A concept of a centralized newborn hearing screening model existing in Ernakulam District - Kerala to cater to all hospitals in the district is worth replicating [12]. It takes away the financial burden of each hospital investing for the screening equipment. Follow up of positive cases and drop-outs are made easier with the central reporting and monitoring system. With unified strength of pediatricians, IAP city/ district branches could take initiative to replicate this model in their respective towns or districts and by collaborating with government agencies involved in implementation of Rashtriya Bal Swasthya Karyakram.

Newborn hearing screening will help to identify hearing loss at an earlier age and alleviate the double tragedy of inability to hear and speak. Forming a consensus and national level guidelines for hearing screening is very important to construct a healthy independent society. Early intervention is mandatory for best prognostic outcomes.

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ANNEXURE I

Participants of the National Consultative Meet for Development of IAP National Consensus Guidelines on Newborn Hearing Screening

Convener: Dr Samir Dalwai, Mumbai.

Experts: (In alphabetical order) Abraham Paul, Cochin; Anjan Bhattacharya, Mumbai; Anuradha Sovani, Mumbai; Bakul Parekh, Mumbai; Chhaya Prasad, Chandigarh; Deepti Kanade, Mumbai; Kate Currawalla, Mumbai; Kersi Chavda, Mumbai; Madhuri Kulkarni, Mumbai; Monica Juneja, New Delhi; Monidipa Banerjee, Kolkata; Mamta Muranjan, Mumbai; Nandini Mundkar, Bangalore; Neeta Naik, Mumbai; P Hanumantha Rao, Telangana; Pravin J Mehta, Mumbai; SS Kamath,Cochin; Sandhya Kulkarni, Mumbai; Shabina Ahmed, Assam; S Sitaraman, Jaipur; Sohini Chatterjee, Mumbai; Uday Bodhankar, Nagpur; V Sivaprakasan, Tamil Nadu; Veena Kalra, New Delhi; Vrajesh Udani, Mumbai; Zafar Meenai, Bhopal.

Rapporteur: Leena Deshpande, Mumbai; Leena Shrivastava, Pune.

Invited but could not attend the meeting: MKC Nair, Thrissur; Pratibha Singhi, Chandigarh; Jeeson Unni, Ernakulam, Cochin; Manoj Bhatvadekar, Mumbai.