

Review Article

Neonatal hearing screening

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Introduction:

Significant hearing impairment is an important & common birth defect that occurs 1 to 3 per 100 live births and 2 to 4 per 100 neonatal intensive Care units^{1,2,3}. South East Asia has the largest number of hearing impairment in the world & houses one third of the worlds hearing impaired population. WHO estimates that every year about 38,000 deaf children are born in this region. In Bangladesh 2600 babies born deaf annually where the population is almost 13,000,000. Unfortunately, for many babies, this disability remains undetected until it is too late to prevent undesirable & often irreversible damage.

Early detection and the treatment of childhood hearing loss have remained very important health and social issues. It has been well established that early detection and treatment are essential for the acquisition of communication competence, important social skills, emotional well-being and positive self-

esteem. Most language development occurs during the first few years of life and inadequate auditory input during this critical period irreversibly delays the development of language skills for communication and reading. It also has a profound effect on receptive and expressive speech and language development, which is likely to adversely affect academic and vocational achievements. This ultimately affects successful integration into society and the prospects of leading a productive life⁴.

Despite Advances in hearing Aid technology, improved educational techniques & intensive intervention services available to children to hearing impairment (HI), there has been little advancement in their language development & academic performance^{5,6}. This may be due to late diagnosis. Several prospective studies have consistently demonstrated that early diagnosis of HI & intervention can improve intellectual, language & speech development^{7,8}. One study reported that the only significant variable to affect development of language skills are the age at which HI was diagnosed⁹. Children in whom hearing loss was identified by 6 months of age demonstrated significantly better language scores than those in whom it was diagnosed latter.

It has also been established that traditional methods of behavioral childhood hearing screening are subjective & deficient¹⁰. Using these behavioral methods of screening, the average age of identification of children with hearing loss has been shown to be between 18 and 30 months. This falls far short of the

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recommended standard, that hearing loss in newborns be detected by 3 months of age & intervention implemented by 6 months of age.

Recent advances in the area of hearing screening have facilitated the availability of more sensitive & easy to use screening tools that can effectively & reliably test hearing soon after birth.

Many developed countries have adopted universal newborn hearing screening (UNHS) program using subjective methods. UNHS has in fact, become standard practice in medical care & many Asian countries have begun to develop & report studies of different models of newborn screening & rehabilitation programs^{11,12}. Unfortunately they are very costly, technically sophisticated and beyond the affordability of the poor & developing countries of the world where vast majority of the people live¹³.

High – Risk newborn hearing Screening :

The joint committee on infant hearing in the US (JCIH) has recommended that every newborn infant should be screened¹⁴. The average age of diagnosis of hearing impairment where universal hearing screening is utilized has been reported to be as low as 3 months. Where universal screening is not available, “At Risk” screening may be conducted utilizing criteria to determine screening. Registration of risk babies & periodical check-up of them also helps in the early diagnosis of hearing impairment. It may be more cost-effective to continue screening to the 6% to 8% of babies who are at high risk of developing hearing loss¹⁵. In 1994, Position Statement of the American Academy of Pediatrics Joint Committee on Infant Hearing¹⁶ recommended the maintenance of a role for high-risk indicators associated with sensorineural and/or conductive hearing loss in newborns and infants and modified the list of indicators described in the 1990

position statement. The committee recommends a specific hearing protocol for high-risk infants when universal screening is unavailable. The indicators associated with hearing loss for use with neonates are:

- (1) Family history of hereditary childhood sensorineural hearing loss;
- (2) In-utero infection, such as cytomegalovirus, rubella, syphilis, herpes, and toxoplasmosis;
- (3) Craniofacial anomalies, including those with morphological abnormalities of the pinna and ear canal;
- (4) Birth weight of less than 1500 g (3.3lb);
- (5) Hyperbilirubinaemia at a serum level requiring exchange transfusion;
- (6) Ototoxic medications, including but not limited to aminoglycosides used in multiple courses or in combination with loop diuretics;
- (7) Bacterial meningitis;
- (8) Apgar scores of 0 to 4 at 1 minute or 0 to 6 at 5 minutes;
- (9) Mechanical ventilation lasting 5 days or longer; and
- (10) Stigmata or other findings associated with a syndrome known to include sensorineural and/or conductive hearing loss¹.

The 2007 guidelines were developed to update the 2000 JCIH position statement principles and Infants in NICU also included in high risk target population because research data have indicated that this population is at highest risk of having neural hearing loss.

However, this argument is not tenable because about 50% of infants with hearing loss do not fall within the high-risk category^{17,18}. To identify these children, as well as those in the high-risk group, it is necessary to screen all newborns.

Universal Newborn Screening:

Historically, moderate-to severe hearing loss in young children was not detected until well beyond the newborn period, and it was not unusual for diagnosis of milder hearing loss and unilateral hearing loss to be delayed until children reached school age.

In the late 1980s, Dr. C. Everett Koop, then US Surgeon General, on learning of new technology, encouraged detection of hearing loss to be included in the Healthy People 2000 goals for the nation. In 1988, the Maternal and Child Health Bureau (MCHB), a division of the US Health Resources and Services Administration (HRSA), funded pilot projects in Rhode Island, Utah, and Hawaii to test the feasibility of a universal statewide screening program to screen newborn infants for hearing loss before hospital discharge. The National Institutes of Health, through the National Institute on Deafness and Other Communication Disorders (NIDCD), issued in 1993 a consensus statement on early identification of hearing impairment in infants and young children¹⁹. In the statement the authors concluded that all infants admitted to the NICU should be screened for hearing loss before hospital discharge and that universal screening should be implemented for all infants within the first 3 months of life⁴. In its 1994 position statement, the JCIH endorsed the goal of universal detection of infants with hearing loss and encouraged continuing research and development to improve methods for identification of and intervention for hearing loss. In 2000, citing advances in screening technology, the JCIH endorsed the universal screening of all infants through an integrated, interdisciplinary system of EHDI³. The Healthy People 2010 goals included an objective to "increase the proportion of newborns who are screened for hearing loss by one month, have audiological evaluation by 3 months, and are enrolled in

appropriate intervention services by 6 months"²⁰.

The purpose of early detection of hearing problems and intervention is to maximize linguistic and communicative competence and liberal development for children who are hearing impaired. The American Academy of Pediatrics Task Force on Newborn and Infant Hearing recommended that universal detection of infant hearing loss requires universal screening of all infants. Reliance on a physician's observation or parental recognition has not been very successful. At least five criteria must be fulfilled before universal screening is justified: (1) the availability of an easy-to-use test that possesses a high degree of sensitivity and specificity to minimize referral for additional assessment; (2) the condition being screened for is otherwise undetectable by clinical parameters; (3) there are interventions available to correct the conditions detected by screening; (4) early screening, detection, and intervention result in improved outcome; and (5) the screening program is documented to be cost-effective. Current available evidence confirms that a newborn hearing screening program fulfills most of these criteria. The American Academy of Pediatrics also recommended five essential elements of an effective UNHS program: screening, tracking and follow-up, identification, intervention, and evaluation^{21,22}.

Methods of Screening:

Evaluation of hearing should include a demonstration of a behavior in response to a measured stimulus; this cannot be reliably performed on a child younger than eight to nine months. The screener should be trained to do the observational screening and have appropriate materials.

The expected response is some sort of head turn toward the sound. An exact description

of the head turn and accompanying eye movement should be noted. A normal-hearing child's orientation to sound will progress as follows:

- Newborn – arousal from sleep, or eye widening, eye blinking
- 3-4 mos.- rudimentary head turn, a wobble of the head even slightly toward the sound
- 4-7 mos.- localization to side only
- 7-9 mos.- localization to side and indirectly below
- 9-13 mos.- localized to side and below
- 13-16 mos.- localized directly to all signals to side, below, and above
- 21-24 mos.- locates directly to a sound at any angle.

When there appears to be no response, the screener should report the use of a particular stimulus at his discretion until the observer is satisfied that the failure to respond is genuine. Two repetitions should be adequate to establish this fact. It is important to conduct the screening when the infant is otherwise alert and calm.

Failure of the child to locate the sound does not always indicate that the child did not hear it. So the child who deviates markedly in these behaviors should be referred for electrophysiological measures.

At present the choice of device for newborn hearing screening is between Otoacoustic emission (OAE) & Automated Auditory brain stem response (AABR), or a combination of the two. Both are non-invasive, quick & easy to perform on newborns, although each assesses different hearing mechanisms.

OAE measures sound waves (emissions) generated by the motion of the outer hair cells in the cochlea. It detects peripheral hearing loss. Emissions are not detected in an infant who cannot hear. The OAE technology can

have a sensitivity of 95% and a specificity of 91%. The referral rate is 5% to 20% when screening is performed within the first 24 hours of life²³.

AABR provides an electrophysiological measure of the hearing pathway along the auditory nerve. Three small sensors are placed on the infant's head to record the brain wave activity of the auditory brainstem in response to sound. AABR judges the response against a "normal" template in order to determine the presence/absence of waveforms. The false positive rates range from 0.3% to 2.5%²¹. The referral rate is less than 3% when screening is performed during the first 24 to 48 hours after birth²³.

Draw backs of Universal Newborn Hearing Screening:

Universal newborn hearing screening produces a large number of false-positive test results. Both AABR and OAE can be influenced by motion artifact and therefore are more specific if performed on a sleeping child in a quiet room. OAE may also be affected by debris or moisture in the ear canal, or the presence of middle ear fluid.

The rate of false positives ranges from more than 30 percent for one-step programs using OAE to less than 1 percent with a two-step process, such as retesting a child before discharge if the initial test is positive²⁴.

Increased parental anxiety may result from a false-positive test, although this finding has not been demonstrated consistently in all studies^{25,26}. Qualitative studies indicate that negative parental emotions may be addressed with more systematic education before and after screening²⁶.

Despite these concerns, the consensus of multiple organizations that develop children's health guidelines is that the potential benefits of universal newborn hearing screening outweigh its adverse effects²⁷.

Hearing Screening Programs Throughout the World:

Currently in the United States, where there are universal hearing screening programs in 40 states, 86.5% of neonates are tested. This may be compared with only 25% of infants who were screening in 1999. Across the UK, over 100,000 children have been tested and similar programs have commenced throughout Europe, as well as in Canada and Australia.

In the **South East Asia Region**, a Guideline is going to be setup for Infant Hearing Screening. In consideration of the indigenous problems of this region, screening protocol should be divided into two phases. **The first phase** shall to screen all infants and identify those with bilateral, severe to profound hearing loss by the age of six months and to ensure the prompt initiation of rehabilitation measures soon thereafter. To fulfill the objective Institutional based screening and community based screening would be followed.

Conclusion:

UNHS is becoming standard medical care in developed countries. There is an urgent need to incorporate universal neonatal hearing screening in all the neonatal health care facilities in Bangladesh. Cost effective and appropriate behavioral methods may be used if resources are limited and use OAE and AABR test to confirm. Any UNHS is a multi disciplinary program and will involve pediatricians, audiologists, otolaryngologists, nurses, speech therapists, community health workers, and education specialists.

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