

Early Identification of Hearing Loss and Centralized Newborn Hearing Screening Facility-The Cochin Experience

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Significant hearing loss is one of the most common major abnormalities present at birth. If undetected, it will impede speech, language and cognitive development. Significant bilateral hearing loss is present in 1 to 3 per 1000 new born infants in the well-baby nursery population and in 2 to 4 per 100 infants in the intensive care unit population. It is an established fact that if hearing loss is present it should be detected and remediated before the baby is 6 months old. Neither universal screening nor a high risk screening, exists in majority of the hospitals in our country. In such a situation, a centralized facility catering to all hospitals in the city is a practical option. A two-stage screening protocol is projected, in which infants are screened first with otoacoustic emissions (OAE). Infants who fail the OAE are screened with auditory brainstem response (ABR). This two tier screening program (the second tier being ABR, which is more expensive) is required only for a selected few, making the program more practical and viable. It is the practicability of this program that makes it relevant for replication in other cities of the country, making it a model screening program for any developing country.

Key words: *Disability, Hearing loss, India, Prevention, Universal newborn hearing screening.*

Significant hearing loss present at birth is one of the most common major disabilities. Early detection can prevent further disabilities in speech, language and cognition in the child's development. It is established that hearing loss, if present, should be detected before the infant is 6 months old. In a developing country like India, simple and practical preventive measures for universal screening need to be developed for this purpose. In majority of the hospitals in India, there is neither a universal neonatal screening nor a high risk screening for hearing, as a routine.

The Indian studies mostly cite screening facility available to newborns brought in to tertiary hospitals. It is in this context that a program to provide a centralized screening facility to every hospital in the city at its doorstep for universal screening was conceptualized and established in Cochin.

A two stage screening protocol with otoacoustic emission (OAE) as the first screen, followed by

auditory brainstem response (ABR) for those who fail the first screen was introduced. This two tier screening program (the second tier being more expensive) is required only for a selected few – thus making the program practical and viable. Trained personnel report to each particular hospital at a particular time and day of the week. This protocol makes the screening cost-effective and acceptable; it avoids the need for transporting the neonates to a screening facility and thus significantly prevents dropouts. This overall practicability makes it relevant for our country; making it a potential model screening program.

BACKGROUND

It is well recognized that unidentified hearing loss can adversely affect optimal speech and language development, acquisition of literacy skills, and academic, social and emotional development. The risk is even more in a country like India where infrastructure is abysmally inadequate for prevention and remediation. Neonatal hearing loss and its developmental consequences are measurable

before the age of 3 years [1-3]. If these are not remediated through early intervention, they impact the child for life [4]. There is robust evidence that identification and remediation of hearing loss, when done before 6 months of age for newborn infants who are hard of hearing, enables them to perform significantly higher on vocabulary, communication, intelligence, social skills and behavior necessary for a successful later life [5-8]. American Academy of Pediatrics (AAP) in 1999 advocated universal newborn hearing screening programme (UNHSP) and remedial intervention which is being practiced in most of the developed countries. In a developing country like India, the risk of infants to develop these disabilities is obviously more [9,10]. In interventional programs, the Indian studies mostly cite the screening facilities available to newborns brought in to tertiary referral hospitals [11,12]. A hearing screening equipment facility in every hospital with a maternity unit today may not be a viable proposition. In this background, a practical interventional model was conceived for the city of Cochin (which has 20 hospitals with maternity units) in January 2003. At that time, there was no hearing screening facility for neonates in most of the hospitals in the city. It was felt that if successful, this centralized screening facility on a cost-effective template can go on to become a solution and model for the country.

CENTRALIZED NEWBORN HEARING SCREENING

The AAP Task Force on newborn and infant hearing recommends UNHS by 3 months of age with intervention by 6 months of age. The Joint Committee on Infant Hearing (JCIH) position statement provides guidelines that include Newborn Hearing Screening (NHS) soon after birth, before discharge from hospital, or before 1 month of age, diagnosis of hearing loss through audiological and medical evaluation before 3 months, and intervention through interdisciplinary programme for infants with confirmed hearing loss before 6 months of age [13]. This screening involves all newborns, with special attention to the high risk group which include the following:

- (i) family history of hereditary childhood sensorineural hearing loss;

- (ii) *in utero* infection such as cytomegalovirus, rubella, syphilis, herpes and toxoplasmosis;
- (iii) craniofacial anomalies, including those with morphological abnormalities of the pinna and ear canal;
- (iv) birth weight less than 1500 gms;
- (v) hyperbilirubinemia at a serum level requiring exchange transfusion;
- (vi) ototoxic medications, including but not limited to the aminoglycosides;
- (vii) bacterial meningitis;
- (viii) apgar score 0 to 4 at 1 minute, or 0 to 6 at 5 minutes;
- (ix) mechanical ventilation lasting 5 days or more; and
- (x) stigmata or other findings associated in the sensorineural and/or conductive hearing loss.

The Indian Academy of Pediatrics (IAP) Cochin Branch mooted a centralized screening facility, initially for high risk screening but quickly decided to choose the two-tier UNHS for all newborns. Despite several neonatal intensive care units in the private and government sectors in Cochin city, adequate screening facility was lacking. Awareness programs and brainstorming were done with pediatricians and obstetricians catering to those units to design an acceptable protocol. Twenty major hospitals with maternity units catering to various socioeconomic strata came on-board, and the program for centralized newborn hearing screening (CNHS) facility was launched in January 2003.

COCHIN EXPERIENCE

Screening facility operates out of Child Care Centre, which is also the secretariat of IAP Cochin Branch. We have acquired 3 portable screening machines. Personnel with basic knowledge in computer and good communication skills were chosen, given basic training in hearing screening. At present there are 3 screening machines and 3 screeners.

The co-ordination with the medical and nursing staff in the neonatal units is done by the screener. She is also in-charge of the screening register. The respective pediatrician apprises the parents of the infant about the procedure and its need. As per guidelines, the neonate gets screened before the day

of discharge. If there is an abnormal result, repeat test is done on the day of the infant's next scheduled visit. If the repeat test also is abnormal, the baby is referred for ABR (available in most hospitals in the city). Babies with abnormal ABR are referred to National Institute of Speech and Hearing (NISH), Trivandrum, for comprehensive evaluation and remediation. The evaluator records the medical history from the discharge card/inpatient file of the infant, to identify the high risk factors recorded. High risk infants who miss screening are screened on subsequent scheduled follow up visit. Screening takes only about 3-4 minutes, if the baby is in natural sleep. Older babies may require sedation. Babies requiring repeat test are given specific dates by the hospitals for the scheduled follow up visits, so that it will coincide with the visit of the screener.

OAE, ABR and Automated ABR (AABR) testing have all been used in newborn hearing screening programs. ABR assesses auditory function from the eighth nerve through the auditory brainstem. OAEs are used to assess structural integrity and are physiologic measurements of the response of outer hair cells of the cochlea to acoustic stimuli. They serve as a fast objective screening test for normal cochlear function.

OAE tests are used to assess the outer, middle and inner ear portions of the auditory system. ABR testing helps in assessing the whole system, from periphery to the auditory nerve and brainstem. If an infant has normal OAE and abnormal ABR, he may be having auditory neuropathy or auditory dyssynchrony. The ABR and OAE are tests of structural integrity of the auditory pathway and not tests of hearing; therefore, even if ABR and OAE test results are normal, hearing cannot be considered definitely normal until a child is mature enough for a reliable behavioral audiogram. All infants, regardless of newborn hearing screening outcome are recommended to receive ongoing monitoring for development of age-appropriate auditory behaviors and communication skills. Any infant who demonstrates delayed auditory and/or communication skill development, even if he or she passed newborn hearing screening should receive an audiological evaluation to rule out hearing loss. These are also communicated to the parents by the pediatrician.

Over the period of 7 years from January 2003 till December 2009, we screened a total of 10,165 babies and the incidence of hearing loss as per our observation in the high risk group is 10.3 per 1000 and 0.98 per 1000 in the well baby group (**Fig. 1**).

The relative advantages and disadvantages of a 2 stage (OAE/ABR) protocol for newborn hearing screening need to be considered carefully for individual circumstances. In locations where getting infants to return for out patient screening and testing is very difficult, the substantially lower failure rate that will likely be achieved by using both OAE and ABR at the same sitting has significant advantages. In a setting like ours, this may not be very practical, but has to be considered wherever possible.

PROBLEMS FACED AND SOLUTIONS

One challenge we initially faced was getting the infant who failed the first screen for retest after 2 weeks (AAP guidelines). This was solved by coinciding the immunization visit with that of screening. Performing test at that age period was a little time consuming because one has to wait for the baby to go to natural sleep. Another challenge was convincing parents (in some cases grandparents) the need for ABR in babies with abnormal OAE. It required counseling and cajoling, which was time consuming. The biggest hurdle was convincing the need for fixing a hearing aid in ABR abnormal babies, probably due to the stigma attached to having a hearing aid. As the programme is gaining roots, these are becoming easier.

CONCLUSIONS

UNHS has become a national practice in most developed countries. The identification of all newborns with hearing loss before 6 months has now become an attainable realistic goal, as our program of UNHS in Cochin now crosses 7 years and over 10 thousand cases. With our limited data, it is too early to arrive at any conclusions or definite interpretations yet. Our unique experience is still evolving. Nevertheless, the detected cases represent a rate of about 1% hearing loss among high risk newborns. This correlates well with national and international figures and therefore calls for UNHS to be made a national practice.

Algorithm of Babies Screened for Hearing Loss

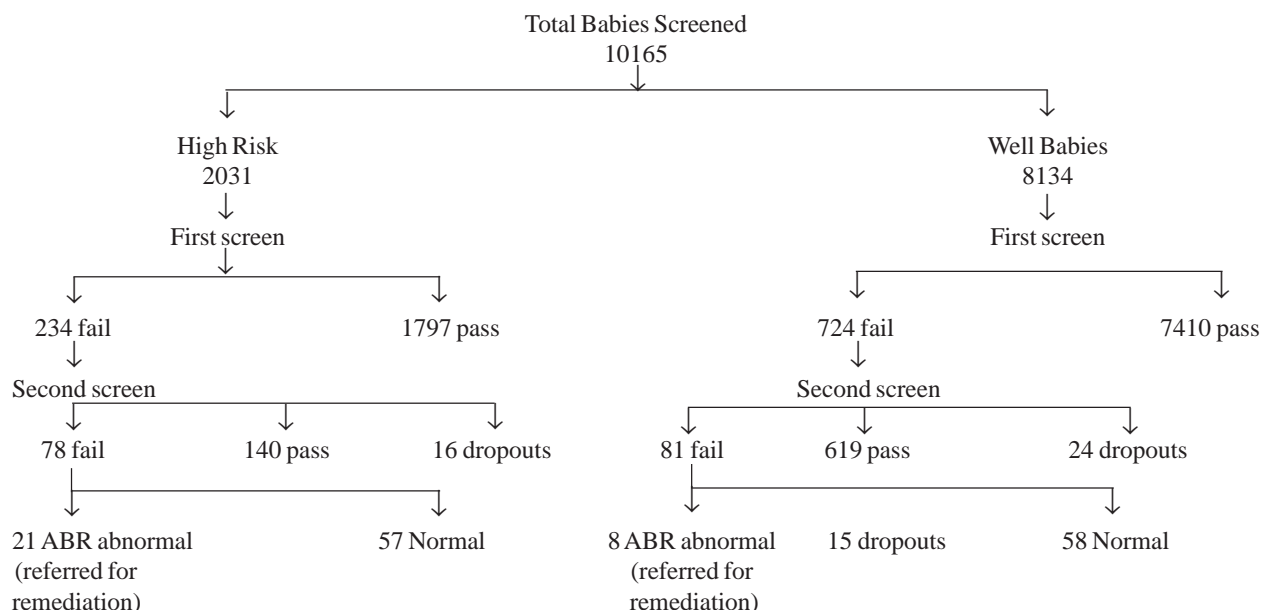


FIG. 1 Result of newborn hearing screening for high risk and well babies.

UNHS need not detect all cases of congenital hearing loss - it only provides an indication of the baby's hearing at the time of the screening. Mild hearing loss and hearing loss outside the main speech frequencies may not be detected. Hearing impairment may develop after the neonatal period and therefore, it is crucial for the pediatrician to encourage parents to continue to have their child's hearing checked. The pediatrician should maintain a high index of suspicion if there are manifestations of hearing loss such as speech and language delay. Any parental concern regarding a child's hearing should also be thoroughly investigated. At Cochin, we have instituted a practical model and a cost effective protocol for early identification of hearing loss through a CNHS facility. This can be replicated in other parts of the country with the unified strength of pediatricians in every town.

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