RESEARCH PAPER

Neonatal Hearing Screening – Experience from a Tertiary Care Hospital in Southern India

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Objective: To implement a neonatal hearing screening program using automated auditory brainstem response audiometry in a tertiary care set-up and assess the prevalence of neonatal hearing loss.

Design: Descriptive study.

Setting: Tertiary care hospital in Southern India.

Participants: 9448 babies born in the hospital over a period of 11 months.

Intervention: The neonates were subjected to a two stage sequential screening using the BERAphone. Neonates suspected of hearing loss underwent confirmatory testing using auditory steady state response audiometry. In addition, serological testing for TORCH infections, and *connexin* 26 gene was done.

Main outcome measures: Feasibility of the screening program, prevalence of neonatal hearing loss and risk factors found in association with neonatal hearing loss.

Results: 164 babies were identified as suspected for hearing loss, but of which, only 58 visited the audiovestibular clinic. Among 45 babies who had confirmatory testing, 39 were confirmed to have hearing loss and were rehabilitated appropriately. 30 babies had one or more risk factors; 6 had evidence of TORCH infection and 1 had *connexin 26* gene mutation.

Conclusion: Neonatal hearing screening using BERAphone is a feasible service. The estimated prevalence of confirmed hearing loss was comparable to that in literature. Overcoming the large numbers of loss to follow-up proves to be a challenge in the implementation of such a program.

Keywords: *BERA phone, Neonate, Screening, Outcome.*

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Neonatal hearing loss has a prevalence that is more than twice that of other newborn disorders amenable to screening such as congenital hypothyroidism and phenyl-ketonuria [1,2]. Congenital, bilateral hearing impairment occurs in approximately 1 to 5 per 1000 live births and when permanent unilateral hearing loss is included, the incidence increases to 8 per 1000 live births [3-5]. Studies done in India using different hearing screening protocols have estimated the prevalence of neonatal hearing loss to vary between 1 and 8 per 1000 babies screened [6-8]. Early identification and intervention for hearing loss by 6 months of age provides better prognosis in language development, academic success, social integration and successful participation in the society [5].

The effectiveness and need for univeral hearing screening in neonates has previously been well proven [9,10]. Although hearing screening programs using different screening protocols have been set up in some centres, procedures for systematic identification and rehabilitation on a large scale are yet to be tested and implemented in the Indian setting.

Accompanying Editorials: Pages 173-5.

Tests used for screening newborns for hearing loss include Otoacoustic emissions (OAE) and automated Auditory Brainstem Response audiometry (aABR). While OAE is cheap, quick, simple and reliable with a sensitivity of 100% and specificity of 99 % [11-13], aABR has the additional advantage of identifying neonates with auditory neuropathy unlike testing for OAE. The other advantages of aABR include rapidity, easy-to-use and high sensitivity (0.99) and specificity (0.87) [14,15]. The Maico MB11 BERAphone is an aABR system employing a special headphone [16]. It consists of a hand-held headphone unit which incorporates a set of three fixed reusable electrodes. It has been tested and found to have a sensitivity of 99.9% and specificity of 97.9% when used in a two-stage screening protocol which is comparable to that of OAE. The test is also

seldom affected by ambient noise making it suitable for use in the postnatal ward [17].

This study was undertaken with the primary objective of exploring the feasibility of setting up a universal neonatal hearing screening program in a tertiary care hospital (handling an average of 10 000 deliveries/year), using the BERAphone (Two-stage sequential screening protocol). The secondary objectives included estimating the prevalence of neonatal hearing loss in a tertiary care setting, and assessing the associated risk factors in those identified with hearing loss.

METHODS

This descriptive study was conducted between January and November of 2010 at our tertiary care center after institutional research and ethical committee clearance was obtained. Four graduates in biological sciences were trained for the study, and their knowledge, ability to obtain informed consent, counsel parents and perform the screening test was assessed formally at the end of the training period.

The BERAphone consists of a handheld headphone unit which is positioned on the babies head after application of electrode gel at the points of contact with the electrodes (vertex and mastoid). An optimized chirp stimulus is used at 35dB and the system automatically detects the presence of an auditory brainstem response based on an implemented statistical test algorithm. If response is detected the test produces a 'Pass' result while failure to detect a response within 180 seconds produces a 'Refer' result.

All normal newborn babies delivered in our hospital were screened by the trained technicians using BERAphone between 24 hours and 72 hours after birth. Newborns admitted in the neonatal intensive care unit (NICU) were screened prior to discharge from the NICU (once their general condition was stable). Mothers of all babies born in the tertiary care hospital were counseled regarding the benefits of hearing screening, procedure of the screening test, need for follow-up and further tests if the neonate failed the screening test, and the interventions available if hearing loss was confirmed. The first screening test was done in the postnatal wards or NICU after obtaining informed consent from the mother. Parents of babies who failed ('refer') the screening test were counseled and asked to return after 1 week for second screening. These babies underwent a second testing in a quiet room. Those who passed on the second screening were discharged from the study while those who failed a second time were referred for further evaluation in the audiovestibular clinic (AVC) at the same centre, where a detailed history for risk factors [10] was obtained, the babies were examined, parents were counseled and diagnostic testing using Auditory Steady State Response Audiometry (ASSR) was done. Repeated phone calls and letters were used to contact parents of babies who failed to return for follow-up.

ASSR was used as the diagnostic procedure to confirm hearing loss, as well as to obtain frequency specific thresholds to enable more effective and appropriate hearing aid fitting. Distortion product otoacoustic emission (DPOAE) testing was used in addition, to detect those with auditory neuropathy. Those confirmed with hearing loss were followed up in the AVC for further evaluation and appropriate rehabilitation.

The babies who were referred after screening twice with BERA phone and whose parents consented for blood tests also underwent serological tests for known infective causes of hearing loss (Toxoplasma, Rubella, Cytomegalovirus and Herpes simplex virus) and genetic testing for the *connexin* 26 gene mutation. Data obtained was analysed using SPSS. Rates, ratios and proportions were calculated.

RESULTS

Among 9671 neonates born between 1st January and 30th November 2010, 9448 (97.7%) were screened for hearing deficit. 223 babies could not be screened since they were critically ill in the nursery and later died or were discharged at request.

A total of 863 babies were referred on first screening which implies a discharge rate of 90.9% with single screening. 713 (82.6%) came for second screening and 164 of them were referred again. The discharge rate after the 2-stage sequential screening with BERAphone was 98.2% (***Fig*.1**). Of the 9448 babies screened, 2339 were NICU graduates (***Table* I**).

Among 164 babies referred to the AVC, only 58 (35.4%) registered in the clinic. The remaining 106 babies failed to come for follow-up despite repeated attempts (phone calls and letters) to contact the families. Eleven of these children were lost to further follow up and did not come back for confirmatory tests despite repeatedly contacting them. One child had died and therefore 46 children underwent confirmatory testing. The ASSR was done between 1 and 3 months of age. Thirty nine were confirmed to have hearing loss and 7 had bilateral normal hearing by ASSR (***Fig*.2**).

***Table* II** shows the associated risk factors [10] identified in the screened babies who had been 'referred'

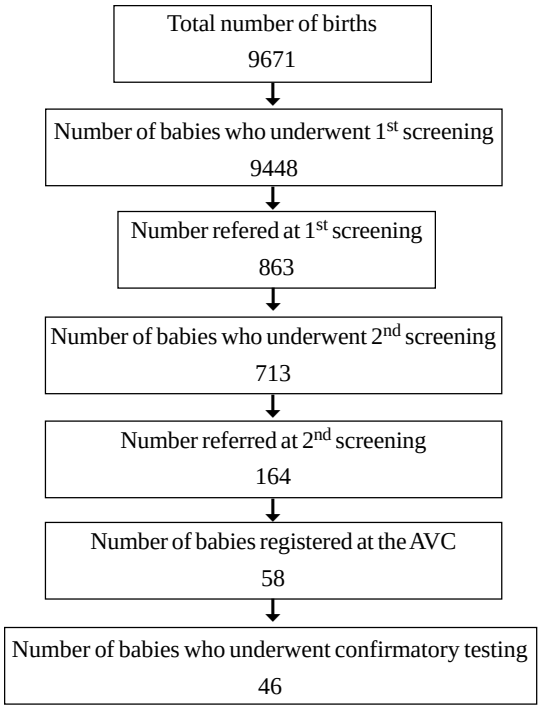

FIG. 1 *Number of babies at each stage of the screening program.*

after the second screening. Among 58 neonates, 30 had one or more risk factors. Three neonates had other congenital anomalies *viz.* Down's syndrome; hydrocephalus, ventricular septal defect and ectopic left kidney; and microcephaly, thrombocytopenia, hepato-splenomegaly and patent ductus arteriosus. Parents of 34 neonates consented for blood tests: screening for TORCH infections and Connexin 26 gene was done. Six neonates were positive for TORCH infections: 5 were positive for Cytomegalovirus while one was positive for Rubella. Of these 6 neonates, 3 had severe to profound hearing loss, 2 had mild to moderate hearing loss and one had normal hearing on ASSR. One neonate out of the 34 was positive for *connexin* 26 gene mutation and the ASSR showed severe to profound hearing loss. The mutation found in this neonate was the common founder mutation W24X.

All children with confirmed bilateral hearing loss of moderate degree or more have been fitted with hearing aids and are on follow-up. Those with severe to profound hearing loss have been advised cochlear implant. One child has undergone bilateral cochlear implant and has joined regular school.

DISCUSSION

The selection, training of staff and establishing procedures for screening were found to be feasible and can be effectively done in any secondary or tertiary level hospital provided adequate knowledge about the importance of the program, the procedure and equipment is available with the supervisory staff. The screening program required intense supervisory input from the primary investigator as well as an audiologist. Frequent evaluation of test procedures, entry of data and supervision of the technicians is required. It is possible, however, to train a non-ENT surgeon for the supervisory role in the screening procedure, maintenance of equipment and interpretation of results.

TABLE II ASSOCIATED RISK FACTORS IN 58 BABIES REFERRED TWICE ON SCREENING

Risk Factor	*No. (%)*
Consanguineous marriage	12 (20.7)
Family history of hearing loss	5 (8.6)
H/o in utero infection	6 (10.3)
Family H/o craniofacial anomalies	2 (3.4)
Family H/o syndromes	3 (5.2)
Hyperbilirubinemia (> 20mg/dL)	3 (5.2)
Very low birth weight <1500g	3 (5.2)
Prematurity (gestation <37weeks)	6 (10.3)
H/o Meningitis	2 (3.4)
Low Apgar score (≥4 at 1 min or ≥6 at 5 min)	2 (3.4)
Mechanical ventilation (> 5 days)	1 (1.7)
Ototoxic drugs (gentamicin)	2 (3.4)
Other congenital diseases	3 (5.2)

TABLE I SCREENING RESULTS OF NORMAL BABIES AND NICU GRADUATES

	Babies screened	*Babies refered on 1st screening*	*Babies who underwent 2nd screening*	*Babies refered on 2nd screening*	*Babies who underwent confirmatory tests*	*Babies with confirmed hearing loss*
Normal	7109	713	563	150	32	31
NICU graduates	2339	150	150	14	14	8
Total	9448	863	713	164	46	39

The BERAphones were quite easy to use and worked very well in high ambient noise surroundings. Under ideal conditions (sound proof room and a quiet sleeping child) the BERAphone screening test takes five minutes to complete. On an average, screening took 10 to 15 minutes to complete in the postnatal ward since the ambient noise in the ward was more than 50 dB (as recorded in the wards with a sound level meter). However, the equipment required frequent servicing by the company and the software required frequent reinstallation. High usage was the reason attributed. The laptop required constant recharging of batteries which added to delays and disruption in work and consequently limited the number of children who could be screened on a given day.

Follow-up (after failing the test the first time) was intended at 6 weeks after birth. In practice, it was found that the follow up was poor at 6 weeks and IgM testing for infective causes required an early sample. Hence the follow up appointment had to be advanced to one week after discharge. Parents were more likely to come a week after discharge from the hospital for a checkup hence decreasing the dropouts. This also had the advantage that those children who failed the test the second time could be referred for the diagnostic test earlier.

The waiting time for confirmatory testing was between 1 to 3 months. This was because of the availability of only one testing facility for both the routine diagnostic testing of patients attending tertiary care and the neonates identified during the study. Often babies required multiple attempts to obtain a satisfactory result because of artifacts produced by upper respiratory tract infections and failure of the baby to achieve deep sleep. Frequently, patients did not keep appointments and so had to be rescheduled for another date. Babies with confirmed hearing loss could be fitted with appropriate hearing aids by 6 months of age and started on auditory verbal therapy thereby initiating the process of early rehabilitation.

The estimated prevalence of hearing loss among neonates in this study was 4.1 per 1000 babies screened. Although this value is similar to that obtained in other studies done in India [6-8], it is still an underestimation considering the large number of babies who were lost to follow-up. Nearly 50% of neonates who attended the AVC after failing the screening test twice had one or more risk factors for hearing loss. However, babies with risk factors are more likely to be brought back for follow-up as these children require frequent hospital visits for various other reasons. The causal association of the identified risk factors is also difficult to ascertain.

The fact that nearly 98% of the babies born in the hospital were recruited for the first screening and more than 80% of those identified on the first screening completed the 2nd stage of screening establishes the feasibility of a 2 stage sequential hearing screening protocol using automated ABR (BERAphone) in a tertiary care set-up. However, ensuring follow up of children who were referred twice proved to be the biggest hurdle. Most parents required repeated counseling and multiple telephone calls to return for confirmatory tests. In spite of these measures, our study showed a large attrition of patients. Only 46 of the 164 neonates identified on screening underwent confirmatory tests. The problem of a huge loss to follow-up is a reality even in developed countries which have established universal neonatal hearing screening programs. In the United States, where nearly 95% of neonates are screened only half of those who do not pass the initial screening undergo confirmatory testing and rehabilitation [18,19]. Measures to increase awareness regarding neonatal hearing loss, its effect on the individual and society, available rehabilitation modalities, and the effectiveness of early identification and rehabilitation are essential for the successful implementation of such a program.

We conclude that the BERAphone-based two-step screening is easy to use effectively by trained technicians for the implementation of a screening program. The

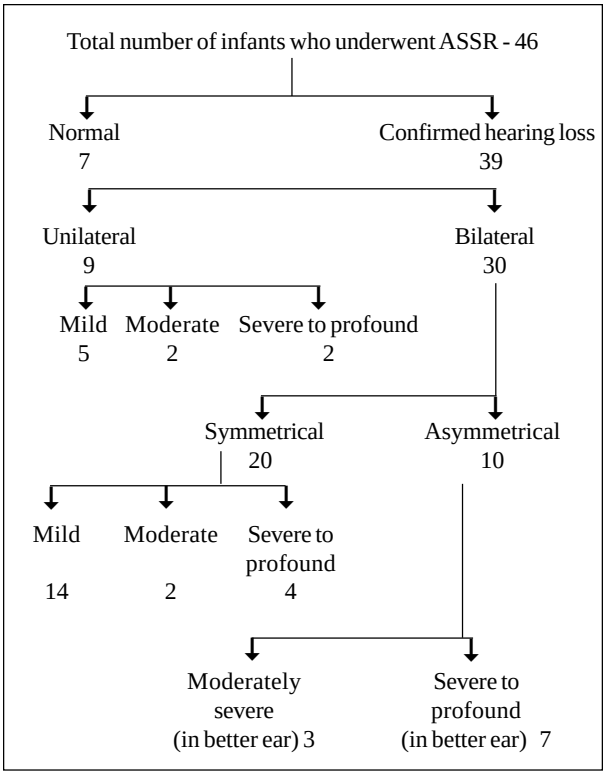

FIG. 2 *Results of ASSR in study subjects.*

WHAT IS ALREADY KNOWN?

- Universal neonatal hearing screening has been widely instituted in most developed countries.

WHAT THIS STUDY ADDS?

- The feasibility of a universal neonatal hearing screening program at a tertiary care set up in a developing country using automated ABR has been emphasized and the potential hurdles including a large number of loss to follow-up have been highlighted.

sensitivity and specificity of the equipment in the test setting however, are to be ascertained. A large loss to follow-up is the biggest hurdle in the implementation of such a program.

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