



Universal Screening in Newborn

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Abstract

Hearing impairment is a primary communication problem limiting an infant's access to spoken language. The objective of the study was to study the incidence of hearing impairment in "NO Risk" and "At Risk" newborn using Transient evoked otoacoustic Emission (TEOAE) and to know the feasibility of using TEOAE as Universal screening procedure for detecting hearing impairment in newborns in developing countries

Methods: A prospective observational study conducted in R.C.S.M. Govt. Medical College & Chhatrapati Pramila Rajee Hospital Kolhapur during the period of December 2014 to April 2016. All enrolled newborn are screened by TEOAE within first 3 days of life followed by screening at the age of 4 to 6 weeks. Babies of "At Risk" group & babies of "No risk" group who failed the first hearing test, confirmation of hearing impairment was done at 3 months of age by Auditory brain stem response (ABR)

Results: A total of 2638 neonates were screened of which 217 babies were from "At Risk" group & 2421 babies were from "No risk" group. 15 neonates from total 2638 had hearing impairment confirmed by ABR. The incidence of hearing impairment is 5.6/1000 screened with 95% confidence interval between 3.4 -9.4. incidence of hearing impairment in the "No Risk" group was 2.89 with 95% confidence interval between 1.4 - 6 where as incidence of 36.8 per 1000 with 95% confidence interval between 16.0 - 71.3 was seen in "at Risk:" group.

This study has shown that two stage TEOAE hearing screening can be successfully implemented as newborn screening method for early detection of impaired hearing on large scale. This study also brought out the fact that, though the incidence of hearing impaired in "AT Risk" newborn is higher than "No Risk" newborns, universal hearing screening is essential to detect hearing impairment in "No Risk" Population also. Two stage TEOAE proves to be feasible method for early detection of congenital hearing loss in India.

Key Words: TEOAE, Universal hearing screening, Auditory brain stem response (ABR).

Introduction

Deafness is one of the most common congenital anomaly in the newborn. Incidence of congenital sensorineural hearing loss (SNHL), averages approximately 3/1000 (hearing loss of 30db or

more).^{1,2} This incidence in the newborn population is greater than the combined incidence of all the metabolic conditions that are currently screened for at birth, such as phenylketonuria, hypothyroidism and galactossemia.³ The

incidence dramatically increases with presence of any risk factors for hearing loss in children and is 10 times greater (2-5%) for infants with one or more risk factors included in “High Risk Registry (HRR)” of Joint committee for infant hearing (JCIH,2000)⁴. Studies done in India using different protocols have estimated the prevalence of neonatal hearing loss to vary between 1-8 per 1000 babies screened.^{5,6}

This stud has been done to know the actual incidence of hearing impairment and feasibility of hearing screening methods for early diagnosis of impaired hearing in newborn with aim of early intervention.

Material and Methods

All newborns born in Chhatrapati Pramila Raje Hospital Kolhapur were enrolled in the study during period of December 2014 to April 2016 with prior informed verbal consent obtained from parents . Enrolled babies were grouped into “At risk” and “No risk” group based on presence or absence of risk factors included in the High Risk Registry (HRR) of Joint Committee on Infant Hearing (JCIH) in 2007⁷.

Risk indicators included are

1. Caregiver concern regarding hearing, speech, language, or developmental delay.
2. Family history of permanent childhood hearing loss.
3. Neonatal intensive care of more than 5 days or any of the following regardless of length of stay: Extracorporeal Membrane Oxygenation (ECMO) therapy, assisted ventilation, exposure to Ototoxic medications or Loop Diuretics and hyperbilirubinemia that requires exchange transfusion.
4. In utero infections, such as Cytomegalovirus (CMV), herpes, rubella, syphilis, and toxoplasmosis.
5. Craniofacial anomalies, including those that involve the pinna, ear canal, ear tags, ear pits, and temporal bone anomalies.

6. Physical findings, such as white forelock, that is associated with a syndrome known to include a sensorineural or permanent conductive hearing loss.
7. Syndromes associated with hearing loss or progressive or late-onset hearing loss, such as Neurofibromatosis, Osteopetrosis, and Usher syndrome; other frequently identified syndromes include Waardenburg, Alport, Pendred.
8. Neurodegenerative disorders, such as Hunter syndrome, or sensory motor neuropathies,
9. Culture-positive postnatal infections associated with sensorineural hearing loss, including confirmed bacterial and viral (especially herpes viruses and varicella) meningitis.
10. Head trauma, especially basal skull/temporal bone fracture that requires hospitalization.

“At risk” group included neonates who had distinct and significant associations with risk factors included int the HRR of as JCIH 2007. “No risk” group included neonates who did not fulfill the criteria mentioned in the HRR of JCIH 2007.

Technique and Tool

Handheld TEOAE device , “ NEURO –AUDIO” OAE Screener Manufactured by ‘ NEUROSOFT’ made in Russia, was used for Initial Screening & Follow-UP Screening.

It is hand held battery operated screening device that can be used for newborns, children and adults. The NEURO-AUDIO OAE detection scheme is based upon signal statistical analysis which guarantees high specificity and sensitivity with minimal impact of background noise and recording conditions. It has a clinical sensitivity of more than 99 % without requiring decision or equipment adjustment by the user. It has a TEOAE testing frequency range from 0.7 to 4 kHz. The instrument does not permit beginning the OAE test until a proper seal of the probe is obtained. A single button push initiates OAE

searching which last for approximately 3 min (maximum time depends on environmental noise conditions.)

The display shows statistical waveform. Measurement progress, TEOAE detection level, noise level, and the results- PASS or REFER. PASS, determined by a statistical algorithm, based on binomial statistics, indicates that the patient has normal outer hair cell function at the time of testing. A REFER result diagnostic hearing evaluation. It also show 'A' value greater than 20% indicate a noisy test and the 'S' value less than 80% indicates the ear probe mal-position. When test result show a 'A' value > 20% and 'S' value < 80% a repeat test is advocated. The screening was conducted in noiseless room with babies in sleeping state after confirmation of non-obstructive external auditory canal.

Screening /Re-screening Protocol – as per flow chart -1

The study protocol was carried out in three steps.

- 1. Initial Screening** – All newborns enrolled into study were screened by

Table - 1

	Total Number screened	Refer in initial screening (Refer Rate)	Refer in 1 st follow – up Screening (Refer Rate)	Refer in 2 nd follow-up Screening (Refer Rate)	Incidence of Hearing Impaired
Total Screened	2638	289 (10.95%)	53.(2.009%)	15(0.56%)	5.6/1000
AT Risk	217	102 (47%)	37 (17.05%)	08(3.36%)	36.8/1000
No Risk	2421	187(7.12%)	16.(0.66%)	07(0.28%)	2.89/1000

Statistical Analysis

The data was entered into MS Excel spreadsheet and was analyzed by using Epi-Info version 7.2 .Appropriate tests of significance were used.

In the initial screening 289 of the 2638 study cohort screened, failed the initial TEOAE test, accounting to a referral rate of 10.95% and pass rate of 89.05%. Of the 289 who failed, 102 belonged to “at risk” group and 187 were of “no risk” group (Flow Chart 2). The referral rate in “at risk” neonates was 47% and that in “no risk” neonates was 7.72% (Table 1)

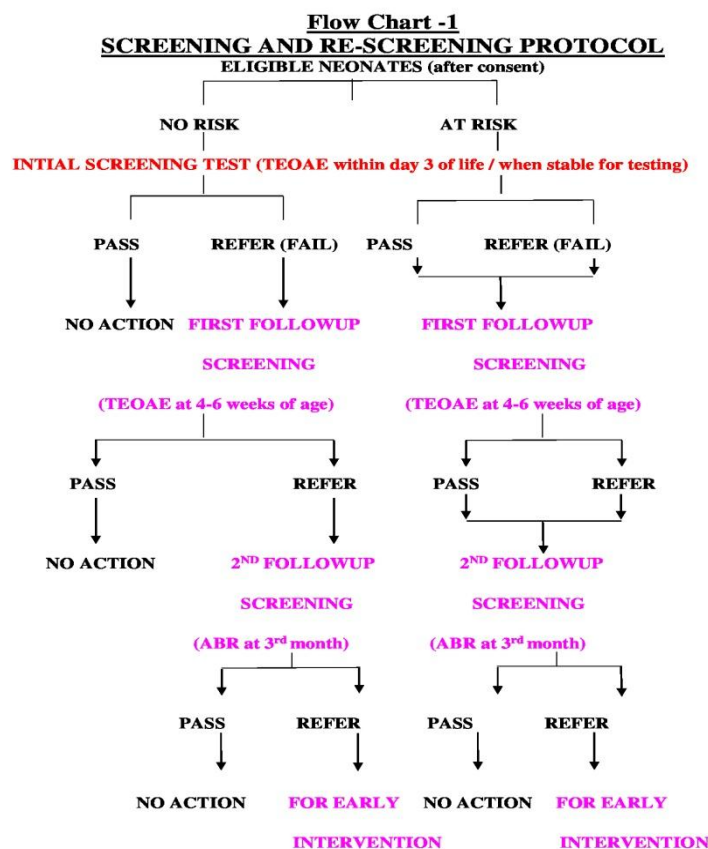
TEOAE within first 3 days of life / as soon as the babies were fit enough to undergo the test in case of very sick babies .

- 2. First follow- up screening** – was done at 4 to 6 weeks of age by TEOAE for –
 - All babies of “At risk” group
 - Babies of “No risk” group who failed the first test screening (‘refer category)
- 3. Second follow –up screening** - was done at 3 months age to confirm the hearing impairment by BERA test for.
 - All babies of “ At risk” group
 - Babies of “No risk” group who failed the first follow-up screening (refer’ category)

Results

A total of 2638 neonates were included into the study , of which 217 (8.22%) had risk factors for hearing impairment as per “HRR” of as JCIH 2007 (“at risk group”) . Results at different stages of the study are shown in table 1 and flow chart 1.

In the 1st follow-up TEOAE 53 neonates failed the TEOAE for the 2nd time of which 37 belonged to “at risk” group and remaining 16 belonged to “no risk” group. Among the “at risk” group though whole group was subjected to TEOAE screening for 2nd time, no failures were found among the infants who had already passed the initial screening (Flow Chart 2). The referral rate in first follow-up screening (end of 2staged TEOAE) was 2.00% of the total study cohort, with 17.5% referral rate among the “at risk” group and 0.66% referral among the “no risk”. (Table 1)



The 2nd follow-up screening which was done to confirm hearing deficit, using BERA showed 15 neonates with hearing impairment among the total study population of 2638. Here again though whole of ‘at risk’ group were subjected for BERA no failures were found among the infants who had already passed the TEOAE (Flow Chart 2). Of the 15 who failed BERA, 8 newborn had risk factor for hearing loss as per JICH and the rest 7 had no risk factor for hearing impairment.

Incidence of hearing impaired in the total study cohort – 15 newborn among the study cohort of 2638 screened had hearing impairment confirmed by BERA. The overall incidence of hearing impairment is 5.6 / 1000 screened with a

95% confidence interval between 3.4-9.4. (Table 2) .

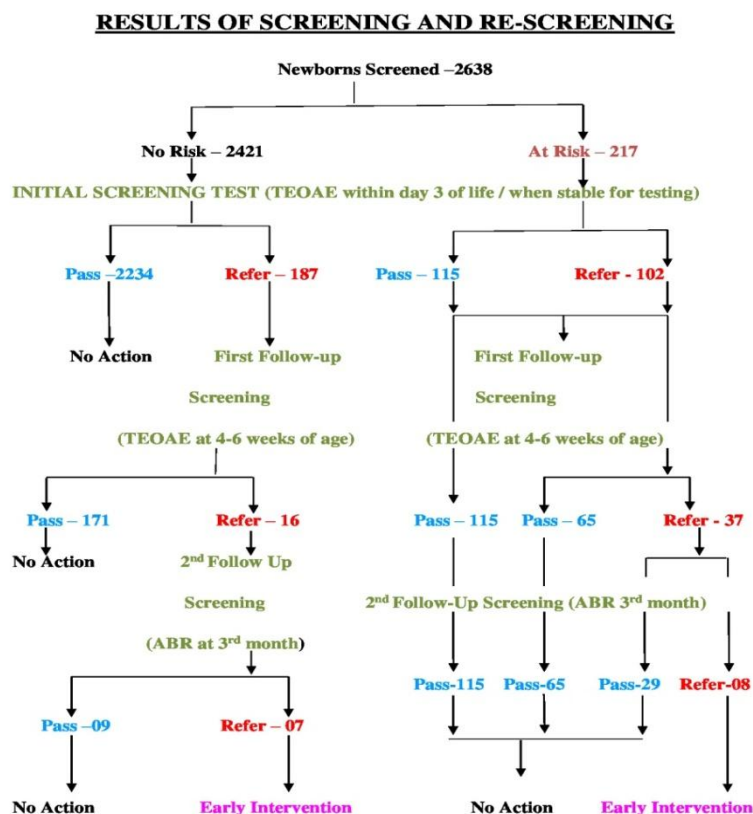
Incidence of hearing impaired in no risk newborns – Among 2421 infants with no risk factors screened only 7 had hearing impairment, showing an incidence of 2.89/1000 in the no risk group with a 95% confidence interval is between 1.4-6.0 (Table 2)

Incidence of hearing impaired in at risk newborns – 217 at risk neonates were screened and 8 were detected to be hearing impaired, which is an incidence of 36.8/1000 (95% confidence interval is between 16.0-71.3). (Table 2)

Table 2 – Incidence of Hearing Impaired

Children Screened	Incidence in the cohort	Incidence expressed / 1000 screened	95% confidence interval per 1000 screened
Total Screened	15 / 2638	5.6	3.4 – 9.4
At Risk	8 / 217	36.8	16.0 – 71.3
No Risk	7 / 2421	2.89	1.4 – 6.0

Flow Chart – 2



The distribution of “at risk” infants screened as per their risk factors and the hearing impaired in various group of infants with risk factors is shown in (Table 3).

In the study eight hearing impaired infants were detected in the at risk group. One newborn suffered congenital rubella syndrome and sepsis in early neonatal period. One newborn suffered from extremely high levels of bilirubin due to

Rh-hearing impairment, two newborns suffered from birth asphyxia with low APGAR score & rest three had NICU stay more than 5 day , out of these , two had birth weight <1.5 kg with respiratory distress requiring mechanical ventilation and the other one had an history of ototoxic drug exposure No hearing impaired cases were detected in newborns with other risk factors.

Table 3

Risk Factor	Number Screened	No. of infants with hearing impairment
Family history of childhood hearing loss.	01	0
Hyperbilirubinemia exchange level	03	01
In utero infections	16	01
Craniofacial anomalies	2	Expired
Syndromes associated	3	1
Culture positive postnatal infections	53	0
Birth asphyxia (APGAR at 1 min<4/5min<6)	65	2
NICU stay> 5day / Mechanical ventilation/ Birth Wt. <1.5/Ototoxic medication	74	3
Total	217	8

Discussion

This study is one of the many steps towards evaluating the need and applicability of universal hearing screening in a developing nation like India. Screening for hearing loss at birth with TEOAEs and later confirming it at three to six months was taken as the standard based on the reports of study by R.C. Deka Et al¹¹. We have tried to look into the incidence of hearing impairment in at risk and no risk group using two staged TEOAE followed by confirmation by BERA, as per the recommendations of National Institutes of Health Consensus (NIHC) Development Conference Statement.¹⁰ TEOAE was preferred as screening tool as it is cost effective, convenient, easy to use and time saving. ABR was used to confirm the hearing defect in TEOAE failed infants to decrease the false alarm and unnecessary intervention. ABR was also done for all the at risk infants with the aim of identifying false negative TEOAE (eg. Auditory neuropathy or auditory dyssynchrony). The incidence of hearing impairment in this cohort is 5.6/1000 with a 95% confidence interval is between 3.4-9.4. As per most of the western studies, incidence of congenital sensorineural hearing loss (SNHL), averages approximately 3/1000.^{1,2} There are few surveys showing incidence of hearing impairment in India. In one such study, by P.Nagapoornima, et al in 2006 an incidence of hearing impairment of 5.6/1000 was demonstrated.¹² The incidence of hearing impairment in our study (5.6 per 1000) is similar to previous studies and much higher than national average of 4/1000.⁸ This may be because our hospital being a tertiary care centre has large number of high risk deliveries leading to larger case load of at risk group. The incidence of hearing impaired 5.6/1000 is high in relation to other congenital defects for which cure can be provided⁹ advocating for an early implementation of hearing screening in our nation. In this study a high incidence of hearing impairment of 36.8/1000 is seen in at risk group when compared 2.89/1000 in no risk group. A

huge disparity has been noticed in the incidence of hearing impairment in at risk and no risk groups, with incidence in at risk group being 12.7 times more than the no risk group. This finding is at par with the literature reports, which state, the incidence in at risk infants being approximately 10 times greater than the incidence in normal population.⁴

It's worthwhile to note that among the 15 hearing impaired detected in the study 7 didn't have any risk factor. Hence just an 'at risk' hearing screen would have missed detection of 7 of the 15 hearing impaired (46.6% of total hearing impaired in the study cohort would be missed). Although the incidence of hearing impaired in no risk group (2.89/1000) is much less than the incidence in the at risk group (36.8/1000), the magnanimity of newborn population in 'no risk' group is huge, leading to a large number hearing impaired missed by high risk screening. Hence universal hearing screening is the ideal strategy of hearing screening for neonates.

The referral rate of 10.95% after the initial screening with TEOAE and a confirmed congenital hearing loss (CHL) of 0.56% in 2nd follow-up screening by BERA, shows a false positive rate of 10.39% with single TEOAE evaluation. The false positivity of the OAE decreased to 2.009% with 2nd screening with TEOAE, showing the importance of two-staged TEOAE screening followed by confirmatory BERA in reducing the unnecessary intervention and parental alarm and concern.

It is necessary and high time to implement and incorporate universal neonatal screening in our country to secure normal, social and holistic development of the child by detecting hearing loss at birth and providing remedial services at the earliest. National policies in these lines have to be made for neonatal hearing screening in all national health care facilities in India. Universal newborn hearing screening can yield high returns, and the two staged hearing screening programme is cost effective and feasible. A child who receives early interventions for hearing loss

requires less expensive special education in later part of life and has better chance to have a normal social life and improved quality of life.

All the 15 hearing impaired infants (8 at risk infants and 7 no risk infants) in our study, are being followed up by an audiologist and at high risk neonate clinic. The hearing impaired infants with no risk factors, were worked up for identifying the etiology and no conclusive causative factor is identified till date (genetic analysis pending). One at risk infant with congenital rubella syndrome was noted to have global developmental delay and is being treated with hearing aid for the hearing impairment. All the remaining are developmentally appropriate for age with no further complications till now. All the 15 infants are under constant follow-up at high risk neonatal clinic and audiologist with aim of early intervention and appropriate care.

Conclusion

This study has shown that two-stage TEOAE hearing screening can be successfully implemented as newborn hearing screening method, for early detection of hearing impaired, on a large scale, in hospital, to achieve the high quality standard of screening programs. This study has also brought out the fact that, though the incidence of hearing impaired in 'at risk' newborns is higher than the 'no risk' newborns, universal hearing screening is essential to detect the large number of hearing impaired in the 'no risk' newborn population also. Universal newborn hearing screening using two-stage TEOAEs proves to be a feasible method for early identification of congenital hearing loss in India.

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