Development of hearing is usually completed by the first year of life. Usually, the mother of a congenitally deaf child begins to suspect early that her child is unable to hear. The mother’s instinct senses this, and she usually tries to come to a conclusion with simple distraction tests such as ringing of loud bells, clapping of hands, etc. or other signs that the child is able to hear the sound. Such tests are however are not standardised scientifically and hence a negative result is not conclusive of deaf-mutism.

Distraction tests or behavioural audiology is of limited value in children less than 6 months. Before this age, infants have inadequate upper body and neck control to reinforce a positive result. Given this lack of inadequate response and problem in determining ear specific threshold information, several researches were conducted to develop objective measure of hearing taking advantage of known physiological responses. In 1977, Kemp described Otoacoustic Emissions (OAE) as a means to determine cochlear outer hair cell function. These can be recorded with a small microphone placed

**ABSTRACT**

**Introduction**
Congenital deafness in a child is often missed. Several distraction tests have evolved over time to diagnose congenital deafness. However, these are of limited value in the era of Evoked Response Audiometry. The study was conducted to compare the result of universal neonatal hearing screening (UNHS) in high risk and non-high risk neonates using Otoacoustic Emission (OAE) and Brainstem Evoked Response Audiometry (BERA).

**Materials And Methods**
A study was conducted over a time period of three months at a tertiary care institute to screen all live neonates for congenital hearing impairment using OAE, followed up by BERA, if required.

**Result**
1182 neonates were screened for hearing impairment. 336 were in high risk group and the rest in non-high risk group. Nine neonates turned out to have abnormal BERA results (absence of wave V). Six of them were high risk babies and the rest three were non-high risk ones.

**Discussion**
33.33% of congenital deaf population detected by UNHS belong to the Non High Risk group. Studies across the world suggest at least 50% chance of missing out a congenital deaf child if Universal Neonatal Hearing Screening is not practiced.

**Conclusion**
In order to ensure that early detection and effective intervention are possible for all neonates with hearing impairment, UNHS should be performed. Three stage UNHS protocol using OAE and BERA showed that the implementation of UNHS for congenital childhood hearing loss for all neonates in India would be beneficial.

**Keywords**
Deafness; Infant, Newborn; Hearing Tests; Evoked Potentials, Auditory, Brain Stem; Audiometry, Evoked Response; Otoacoustic Emissions, Spontaneous
in the ear canal just lateral to the tympanic membrane and confirm the integrity of the cochlea. Two types of evoked OAEs are in use, the Transient Evoked Otoacoustic Emissions (TEOAEs) and Distortion Product Otoacoustic Emissions (DPOAEs). Automated Auditory Brainstem Response (AABR) is frequently used for newborn hearing screening as it provides accurate information in a fairly short space of time. It provides an electrophysiological measure of the auditory pathway along the auditory nerve. To administer the AABR test, electrodes are placed on the infant’s forehead and the mastoid of the test ear to record the brain activity of the auditory brainstem in response to sound.

Globally, it has been estimated that 1.5 to 6 per 1000 newborns have congenital hearing impairment. Presently, its diagnosis is markedly delayed. At this progressed age, rehabilitation procedures (like hearing aids, cochlear implant, speech therapy, psychological intervention for family) are unable to ensure complete development of speech, thus preventing the full participation of the deaf child in social living. This has brought in the concept of Neonatal Hearing Screening. Auropalpebral reflex, Moro reflex, combined head and limb movement and their relationship to acoustic stimulation formed the basis for screening for deafness in the early 1960s. Nowadays, Neonatal Hearing Screening usually makes use of OAE and BERA. Early recommendations by Joint Committee of Infant Hearing encouraged screening of neonates with high risk factors. Unfortunately, several estimates have suggested a proportionately significant number of children in non-high risk groups to have congenital hearing impairment. The present study compares the result of Universal Neonatal Hearing Screening in High Risk and Non-high Risk Neonates.

Materials And Methods:
The study was conducted between, 1st July, 2014 and 30th September, 2014 in a tertiary care hospital. All babies born in the institute within the study period and available for screening at 24 to 48 hours of birth on two specific days of the week were included in the study. Classification of the neonates as high risk group was performed using the Newborn Hearing Screening Programme (NHSP) Risk Factor Screening guidelines (Table I). Presence of one or more risk factor(s) resulted in the newborn being placed in the high risk group.

Verbal explanation of the screening process was given to the mothers. Additionally, a leaflet describing the screening procedure was also made available. Before screening, proper inspection of the external auditory canal was done with an otoscope. Any vernix or fluid in the external auditory canal was cleaned. A three stage newborn hearing screening protocol was implemented.

All babies were initially screened for hearing by TEOAE equipment using neonatal probes. The result of the test was interpreted as “Valid Response or Pass” or an “Invalid Response or Refer.” Babies having ‘Refer’ response were sent for second stage screening.

The Second Stage Screening was performed with TEOAE at 4 weeks interval from the first screening. Results were interpreted as in first stage. Babies having a ‘Refer’ response in the second stage were sent for the third stage screening immediately.

Brain Stem Evoked Response Audiometry (BERA) was done in the third stage of screening and an abnormal BERA result (absence of wave V) was taken as confirmation of congenital hearing loss.

Results:
The study was conducted on 1182 neonates after institutional delivery. 336 were High risk neonates and the rest non-high risk (Table II). Out of the total population available for first stage screening, 91 had ‘Refer’ result in TEOAE; 25 were from the high risk and 66 from the non-high risk group. Amongst these refer cases, 81 were available for second stage screening. 15 of them had a ‘Refer’ result in the second TEOAE screening with 10 babies in the high risk and 5 in the non-high risk group. Amongst these refer cases, 81 were available for second stage screening.

15 of them had a ‘Refer’ result in the second TEOAE screening with 10 babies in the high risk and 5 in the non-high risk group. These cases were followed up with BERA as a third stage screening. 9 turned out to have abnormal BERA result (absence of wave V); 6 of them were high risk babies and the rest 3 were non-high risk babies.
ones (Table III).

Thus, to put it into perspective, in the non-high risk group, 1st stage TEOAE screening yielded ‘Refer’ result in 7.8% neonates which subsequently declined to 0.59% in the 2nd stage TEOAE screening, and was ultimately initial OAE screening. The second stage screening showed a significant improvement with ‘Pass’ results in both the groups although the proportion of ‘Refer’ results was more in the high risk group. 3 infants out of the total non-high risk population was confirmed to have hearing impairment in comparison to 6 from the high risk group. Thus, out of total 9 congenitally deaf children, 3 were in the non-high risk group showing that 33.33% of the congenitally deaf population as detected by UNHS in the study belonged to the non-high risk group (Fig. 1).

Table I: Newborn Hearing Screening Programme (NHSP) Risk Factor Screening guidelines

<table>
<thead>
<tr>
<th>RISK FACTORS IN NEONATES</th>
<th>YES</th>
<th>NO</th>
</tr>
</thead>
<tbody>
<tr>
<td>Confirmed or strongly suspected bacterial meningitis (any organism) or meningococcal septicaemia.</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Microtia/external ear canal atresia in one or both ears.</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Confirmed congenital infection due to Toxoplasmosis, Rubella, CMV or Herpes as determined by TORCH screen.</td>
<td></td>
<td></td>
</tr>
<tr>
<td>A noticeable craniofacial anomaly.</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Confirmed syndrome relating to hearing loss.</td>
<td></td>
<td></td>
</tr>
<tr>
<td>SCBU/NICU &gt; 48hrs with no clear response A0AEs both ears but clear response on AABR.</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Family history of hearing loss.</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Family history of hearing loss in parents or siblings.</td>
<td></td>
<td></td>
</tr>
<tr>
<td>SCBU/NICU child who had IPPV &gt; 5 days or who underwent Extra-Corporeal Membrane Oxygenation (ECMO).</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Jaundiced at exchange transfusion level?</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Developmental delay associated with a neurological disorder?</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

confirmed by BERA in 0.35% of the neonates. On the other hand, in the high risk group, 1st stage TEOAE screening yielded 7.40% ‘Refer’, declining to 2.97% in the 2nd stage TEOAE screening and was confirmed in 1.79% neonates by BERA.

Thus, almost equal percentage of neonates failed

Table II: Distribution According to Risk Group and Sex

<table>
<thead>
<tr>
<th>SCREENED NEONATES</th>
<th>NON HIGH RISK</th>
<th>HIGH RISK</th>
<th>TOTAL</th>
</tr>
</thead>
<tbody>
<tr>
<td>Male</td>
<td>398 (33.7%)</td>
<td>171 (14.5%)</td>
<td>569 (48.1%)</td>
</tr>
<tr>
<td>Female</td>
<td>448 (37.9%)</td>
<td>165 (14.0%)</td>
<td>613 (51.9%)</td>
</tr>
<tr>
<td>Total</td>
<td>846 (71.6%)</td>
<td>336 (28.4%)</td>
<td>1182 (100.0%)</td>
</tr>
</tbody>
</table>
Discussion:
Studies across the world suggest at least 50% chance\textsuperscript{6,7} exists of missing out a congenitally deaf child if Universal Neonatal Hearing Screening is not practised. Globally it is estimated that 1.5 to 6 per 1000 newborns have congenital hearing impairment. However, there is a significant delay in diagnosis. Average age of diagnosis of congenital hearing impairment in United States is 18 months.\textsuperscript{5} The Joint Committee of Infant Hearing (JCIH) has provided guidelines in 1994, 2000 and 2007. The standard recommendation is that the mean age of diagnosis of congenital hearing impairment in a child should be 3 months, and intervention should be done as early as six months.\textsuperscript{5,10} In India, it is estimated that at least 4 per 1000 newborns have congenital hearing impairment.\textsuperscript{11,12,13,14} India faces a stiff challenge in this field owing to its high birth rate, large number of births in rural India, lack of contact with developed healthcare facilities and lack of proper knowledge.

UNHS implementation initiative should include:
- Development of screening protocol and screening method
- Staff training and monitoring

<table>
<thead>
<tr>
<th></th>
<th>TEOAE 1 (NO. OF ‘REFER’ RESULTS)</th>
<th>TEOAE 2 (NO. OF ‘REFER’ RESULTS)</th>
<th>BERA (NO. OF ABSENCE OF WAVE V RESULTS)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Non-High Risk Neonate</td>
<td>66 (7.80%)</td>
<td>5 (0.59%)</td>
<td>3 (0.35%)</td>
</tr>
<tr>
<td>High Risk Neonate</td>
<td>25 (7.40%)</td>
<td>10 (2.97%)</td>
<td>6 (1.79%)</td>
</tr>
<tr>
<td>Total</td>
<td>91 (7.70%)</td>
<td>15 (1.27%)</td>
<td>9 (0.76%)</td>
</tr>
</tbody>
</table>

**Table III: Result of Hearing Screening**

![Fig. 1 Venn Diagram Showing Results of Hearing Screening](image) *not to scale.
NRH- non high risk group, HR- high risk group, R- refer in OAE, C- confirmation by BERA.
Information to parents or guardians
General hospital consent obtained at the time of admission should include universal neonatal hearing screening
Implementation of UNHS by home visits and immunisation clinic visits
Documentation of results in a standard prescribed format
Assurance of a long term secure funding for the programme

Otoacoustic Emission as a screening tool has been an excellent indicator of cochlear health. The instrument uses pre-programmed algorithms to receive, analyse and interpret the data in the form of a ‘Valid Response or Pass’ (Fig. 2) or a ‘Invalid response or Refer’ (suggestion to move on to the next set of tests). OAE is simple, quick, portable, noninvasive, highly sensitive, reproducible, cheap and has high compliance.

Like OAE, BERA is an indirect measure of hearing. It is highly specific and sensitive, but time consuming and costly. Thus it qualifies as an excellent confirmation tool (Fig. 3).

Conclusion:
In order to ensure that early detection and effective intervention are possible for all newborns with hearing impairment, UNHS should be performed. UNHS is becoming the accepted standard of care in many developed countries. Our current 3 stage UNHS protocol using TEOAE and BERA showed that the implementation of UNHS for congenital childhood hearing loss among all newborns in India is accurate, feasible, effective and above all, necessary. Identification of all newborns with congenital hearing loss can become an attainable realistic goal in India. There is no reason why any child born in India should experience anything other than normal acquisition of communicating skills as a result of early and appropriate intervention when required. Thus, the implementation of a UNHS programme is strongly recommended from an Indian perspective.

References:


