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SCREENING OF DEAF CHILDREN- AN EXPERIENCE AT RAMAKRISHNA MISSION SEVA PRATISHTHAN HOSPITAL IN WEST BENGAL

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ABSTRACT

Background: Hearing impairment is the most frequent sensory deficit in human populations, affecting more than 250 million people in the world. One in 2,600 newborns has Non syndromic Hearing Loss (NSHL). The incidence of hearing loss in India is found to be 1 to 6 per 1000 newborns screened. Screening only the high risk neonates misses 50% of babies with hearing loss, hence a cost effective universal screening is the viable option to sustain such a program. **Methods:** In our study, the possible burden of hearing disability was evaluated in babies born at Ramakrishna Mission Seva Pratishthan Hospital, Kolkata, India. Total 222 newborn children were screened by Otoacoustic Emissions (OAE). Audiometry was performed in 288 cases from 1225 cases who attended the ENT outdoor for various problems. **Results:** Out of 288 cases 22 were diagnosed as deaf in both ears since birth, after audiometry test. All the cases chosen for our study were up to the age of 15 years. The new born babies undergo of Otoacoustic Emissions (OAE) test (222 cases) 88% had pass report in both ears, 6.75% cases in right ear and 5.85% cases in left ear had refer report. The refer cases were asked to reattend after six months.

KEYWORD: Hearing loss, Screening, Nonsyndromic, Otoacoustic Emission.

INTRODUCTION

Hearing impairment in children across the world constitutes a particularly serious obstacle to their optimal development and education, including language acquisition. According to a range of studies and surveys conducted in different countries, around 0.5 to 5 in every 1000 neonates and infants have congenital or early childhood onset sensorineural deafness or severe-to-profound hearing impairment. Deaf and hearing-impaired children often experience delayed development of speech, language and cognitive skills, which may result in slow learning and difficulty progressing in school. The partial or complete inability to perceive sounds is a common disorder in humans. Approximately one in thousand newborn is affected by severe to profound hearing loss (HL) either at birth or during early childhood[1] Genetic causes account for half of cases of childhood deafness and the remainder are attributed to environmental factors.[2] Hearing loss is expected to increase to 25% by 2020 along with increased life expectancy.[3] Hearing impairment is the world's third leading chronic disease.[4] Hearing loss prevalence in isolated communities is almost 15% in newborns.[5]

Hereditary hearing loss is categorized as non syndromic where hearing loss is the only symptom and syndromic where deafness co-segregates with some other clinical manifestations.[6] A disruption of different classes of proteins involved may cause hearing impairment with or without associated syndromic features. Initially the genes involved in syndromic deafness were identified, as individuals with syndromic mode of deafness share other clinical problems in addition to hearing loss that helps in recognition of a distinct entity. In contrast, non-syndromic hearing loss requires linkage analysis in single large families. Non-syndromic autosomal recessive deafness is usually clinically heterogeneous, non-progressive in nature, and exhibits a high degree of genetic heterogeneity.[7] Non-syndromic hearing loss (NSHL) with autosomal recessive mode of inheritance is responsible for 70% of congenital deafness.[8] In some countries, newborn and infant hearing screening has become a widespread tool for the early detection of hearing impairment, while in other countries such screening is considered to be too costly and its value is questioned. Even when it is available, there is no

consistent approach to newborn and infant hearing screening, and there is often great variation within individual countries. The reasons for this are not always financial – some wealthy countries have fragmented and ineffective programmes while a number of less-wealthy countries have very successful programmes.[8]

For many countries, one major challenge is the lack of contact between the majority of mothers and their babies and the health system – with about half of all global births occurring at home without skilled care. In many settings there is no continuum of care from pregnancy and birth to the neonatal period and childhood, and globally only a quarter of all neonates receive any postnatal care. Although efforts to increase the coverage of antenatal and postnatal services do provide opportunities to expand newborn and infant hearing screening, the global situation is complex. In urban Nigeria, for example, the typical delivery model has changed from "at-home" to private "maternity homes" but still not to a hospital-based system. Conversely, recent trends in Canada have seen increasing numbers of children born at home or during only very brief contact with health services. Any WHO guidance on newborn and infant hearing screening models must take into account variations in national, cultural and economic contexts.[6-8]

This study was undertaken in order to detect the frequency of congenital hearing loss among neonates. The study also identified the challenge in implementing a screening programme in normal neonates in Ramakrishna Mission Seva Pratishthan hospital in West Bengal.

MATERIALS AND METHODS

Ethical clearance

The study protocol was reviewed and approved by the Ethical Committee of Ramakrishna Mission Seva Prathisthan, Kolkata, India.

An informed consent was taken from the parent/guardian. The study was conducted prospectively on all neonates born in Ramakrishna Mission Seva Prathisthan Hospital, Kolkata from 1st January, 2014 to 31st August, 2014. Parents of the neonates were informed about the study and motivated to undergo the screening program.

A questionnaire, which included question on previous history of deafness among family, birth history with any kind of complication, illness or complication during pregnancy of mother, age of their parents, and any kind of genetic anomaly was obtained.

OAE is the recording of sounds that the ear produces itself. Otoacoustic emissions were first reported by Kemp in 1978. They appear to be generated by motile elements in the cochlear outer hair cells.[9]

There are 2 types of otoacoustic emissions in clinical use.

- Transient otoacoustic emissions (TOAEs) or transient evoked otoacoustic emissions (TEOAEs) - Sounds emitted in response to an acoustic stimuli of very short duration; usually clicks but can be tone-bursts
- Distortion product otoacoustic emissions (DPOAEs) - Sounds emitted in response to 2 simultaneous tones of different frequencies.

Both the normal and high-risk neonates underwent hearing assessment age between 48 hours to 5 days after birth using TOAE as the first level of screening. Neonates who failed the initial screening were subjected to repeat testing with TOAE after six month (Screening Algorithm, Figure I & II). This was done in the Department of Pediatrics at Ramakrishna Mission Seva Prathisthan hospital using a GSI Audio Screener, which is a completely automated analysis system that gives a "PASS" or "REFER" result. Absence of emissions for 2 out of the 3 frequencies tested (2 kHz, 3 kHz and 4 kHz) was given a "REFER" result. Infants who failed the screening twice were referred. A total of 222 newborn babies from Pediatric Department were tested by OAE to know whether the hearing loss was normal or patients.

Pure tone audiometry was performed on 288 cases out of 1225 attending the ENT outdoor. Out of 288 cases 22 were diagnosed as deaf in both ears since birth. All the cases chosen for our study were up to the age of 15 years. Detailed history of these 22 cases was taken.

RESULTS

New born baby undergoing Otoacoustic Emission (OAE) test (222 cases) showed 90.09% pass report in both ear. In 4.05% cases in only right ear, 3.15% cases in only left ear and 2.7% cases in both ear had refer and they were advised to come after six months for further testing (Table 1).

Detailed history like age, sex, family history of deafness, birth history, speech problem, presence of other congenital anomaly, age of mother, age of father of those deaf cases were taken by filling up questionnaire (Table 2).

Out of 288 OPD attendant 22 selected patients doing pure tone audiometry (PTA) or Brainstem evoked response audiometry (BERA) three cases were diagnosed with syndromic deaf (Down syndrome baby and child with cerebral palsy) and nineteen cases were nonsyndromic deaf children.

Table 1: Screening of newborn by Otoacoustic emissions method.

	No. of Cases	Only Right Ear Refer (Not detectable)	Only Left Ear Refer (Not detectable)	Both Ear Refer (Not detectable)	Both Ear Pass (Normal)
Male	117	3	5	4	105
Female	105	6	2	2	95

Table 2: Detailed history of deaf children.

Sl. No.	Sex	Age	Family History of Deafness	Birth History	Speech Problem	Other Congenital Anomaly	Age of Mother	Age of Father	Type of deafness
1	M	3 Years 4 Months	Nil	Full term gestation caesarean delivery	Say many words but no meaningful word	No	30 Years	32 Years	Non-syndromic
2	M	1 Years 2 Months	Nil	Full term gestation caesarean delivery	Normal	Mental disability	29 Years	30 Years	Non-syndromic
3	M	2 Years	-	Full term gestation caesarean delivery	Normal	Referred to pscychriatist	40 Years	40 years	Non-syndromic
4	F	4 Years 6 months	Cousin brother is deaf	Full term gestation Normal delivery in hospital	Speak 4-5 Words	Delayed development milestone	25 Years	30 Years	Non-syndromic
5	F	4 Years	Causing sister is deaf and dumb.	Full term gestation and Caesarea delivery	Speak 2-3 Words	Cerebral palsy	30 Years	46 Years	Syndromic
6	F	5 Years	Nil	Full term gestation and Normal delivery at home, Birth asphyxia of baby	Speak 1-2 Words	Down Syndrome	30 Years	35 Years	Syndromic
7	F	1 Years 7 Months	Nil	Full term gestation and Normal delivery at primary health center baby did not cry during birth	Problem at 1-2 year. After treatment it is normal	Delayed development al milestone	21 Years	30 Years	Non-syndromic
8	M	1 Years 3 Months	Nil	Full term gestation and Normal delivery at hospital	Dumb	Delayed mental and development al milestone	25 Years	30 Years	Non-syndromic
9	M	1 Years 6 Months	Nil	Prenatal birth at 38 weeks Caesar at hospital	Normal	No	28 Years	33 Years	Non-syndromic
10	M	2 Years 1½ Months	Nil	Prenatal delivery 7 Month, birth weight 1 kg 300 g	Disyllable words	No	27 Years	31 Years	Non-syndromic
11	M	4 Years	Nil	Full term gestation and Normal delivery at hospital	Dumb	No	33 Years	44 Years	Non-syndromic
12	M	4 Years	Not known cause patient is orphan	Not known cause patient is orphan	Monosyllable word from 1½ Year	Down Syndrome	Not Known	Not Known	Syndromic

13	M	1 Years 2 Months	Nil	Full term gestation and Normal delivery at hospital	Dumb	No	19 Years	25 Years	Non-syndromic
14	F	2 Years 3 Months	Maternal grandfather is deaf.	Full term gestation and Normal delivery at hospital. Cried late after birth	Speak very few words 3-4 words	Delayed development al milestone	28 Years	30 Years	Non-syndromic
15	F	4 Years 6 Months	Nil	Full term gestation and Normal delivery at hospital.	Dumb	No	36 Years	36 Years	Non-syndromic
16	M	3 Years 6 Months	Nil	Full term gestation and Normal delivery at hospital but admitted NICU immediately after delivery & stay 2-3 Days	Speak 4-5 words but nothing significant	No	25 Years	28 Years	Non-syndromic
17	F	4 Years	Nil	Prenatal delivery in 7 months and CS delivery in Hospital	Speak very few words	No	40 Years	43 Years	Non-syndromic
18	M	3 Years 9 Months	Nil	Full term gestation normal delivery in hospital. Patient had retinopathy during birth	Normal after treatment	Delayed development al milestone	-	-	Non-syndromic
19	M	3 Years 6 Months	Nil	Full term gestation normal delivery in hospital.	Normal	No	35 Years	41 Years	Non-syndromic
20	M	2 Years 3 Months	Nil	Full term gestation Caesarea delivery in hospital.	Normal	No	-	-	Non-syndromic
21	M	3 Years 4 Months	Nil	Many Complications at the time of births, Meningitis at 1½ month pregnancy. In the time of delivery hydronephrosis was present in baby	Normal	No	-	-	Non-syndromic
22	F	3 Years 8 Months	Nil	Full term gestation caesarean delivery	Speak few words	No	40 Years	41 Years	Non-syndromic

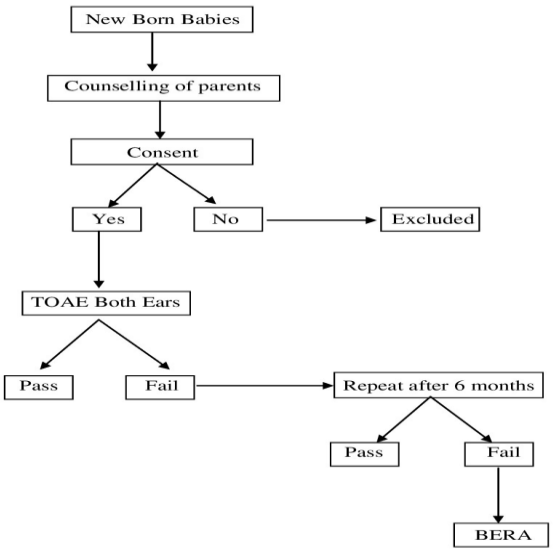

Figure: I screening of new born babies

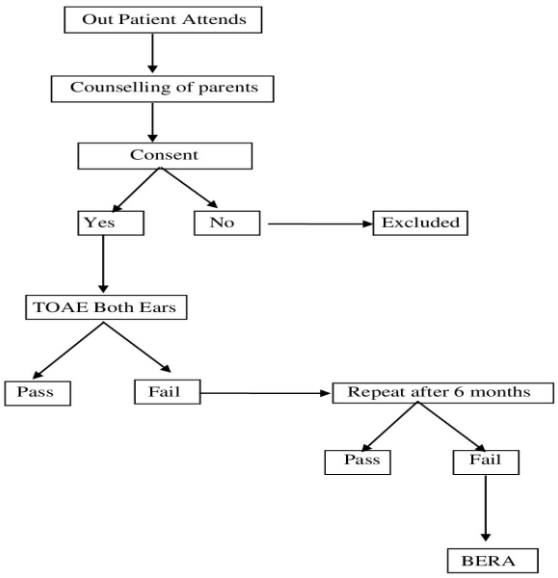

Figure: II Patient attending on E.N.T Dept.

DISCUSSION

Studies have demonstrated that when hearing loss of any degree, including mild bilateral or unilateral hearing loss, is not adequately diagnosed and addressed, the hearing loss can adversely affect the speech, language, academic, emotional, and psychosocial development of young children. Although efforts to identify and evaluate hearing loss in young children have improved, there is still anecdotal evidence to suggest that many young children with hearing loss may not be receiving the early intervention or other services they need in a timely manner that will enable them to enter preschool and school ready to succeed.

It is well recognized that unidentified hearing loss can adversely affect optimal speech and language development, acquisition of literacy skills, academic, social and emotional development. There is robust evidence that the identification and remediation of hearing loss, when done before six months of age for newborn infants who are hard of hearing, enable them to perform significantly higher on vocabulary, communication, intelligence, social skills and behaviour necessary for success in later life.[10] In 1994, the Joint Committee on Infant Hearing (JCIH) established in the United States recommended screening of high risk babies for hearing loss using High Risk Registry.[11] Several studies thereafter suggested that up to 50% of all the children with congenital hearing loss have no risk factors and would be missed by screen- ing only those at high risk.[12] American Academy of Paediatrics (AAP) in 1999 advocated universal newborn hearing screening programme (UNHSP) and remedial intervention, which is being practiced in most of the developed countries. 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 one month of age, diagnosis of hearing loss through audiological and medical evaluation before three months, and intervention through interdisciplinary pro- grams for infants with confirmed hearing loss before six months of age.[13]

Otoacoustic emissions are low intensity sounds generated from the outer hair cells of the cochlea in response to audible sounds. There are two main types of automated otoacoustic emissions (AOAEs) namely, TEOAE and distortion-product otoacoustic emissions (DPOAE). TEOAE, also known as cochlear echoes, are low intensity sounds originating from the active amplification of the outer hair cells and can be elicited in response to clicks or tone bursts presented to the ear through a light weight probe that houses both a transducer and microphone/receiver. The emissions are then matched through advanced digital processing technology with a standard template before giving a 'pass' or 'refer' result. A typical TEOAE instrument is light, portable and powered by an inbuilt rechargeable battery that can last many hours of continuous use. The recording often takes seconds and can be administered without audiological expertise. The sensitivity and specificity are greater than 90%. One disadvantage with this test in newborns is that it is sensitive to peripheral hearing impairment such as mild conductive hearing loss resulting from debris associated with vernix caseosa and amniotic fluid in the external ear canal, in the first day of life. The test is sensitive to excessive internal noise from patient or ambient noise in the test environment and will not detect any retrocochlear dysfunction of the inner hair cells and beyond such as auditory neuropathy/ dyssynchrony. DPOAE differs from TEOAE because they are generated by two continuous pure tones introduced to the ear simultaneously. Because DPOAEs are evoked by frequencyspecific signals, it is possible to use the response to predict frequency-specific hearing sensitivity across the frequency range of 500 to 8,000Hz. DPOAE amplitude and pure-tone audiograms are somewhat but imperfectly comparable in the frequency

region above 1,500Hz. However, this advantage is not critical for screening infants and young children which is perhaps why TEOAE is the most widely used in infant screening programmes. Nonetheless, initial refer rates above 10% are not uncommon with TEOAE when conducted in babies prior to hospital discharge which reduces with subsequent test over time.[14]

In other countries where newborn hearing screening is conducted it is assumed that the vast percentage of babies born deaf can be helped and their futures immeasurably improved. However, issues such as quality control, screening methods, follow-up and cost effectiveness need to be thoroughly discussed and reviewed. Quality assurance issues in particular are vital to successful newborn and infant hearing screening and related interventions – in some settings it is estimated that the poor training and performance of screeners renders up to 80% of screening useless.[15]

In Paediatric Dept., otoacoustic emission (OAE) test were done on all the newborn cases at first. After that those referred at first time them called after six months and OAE test performed again. Those who were referred, second time then under were for BERA test to confirm deafness. After screening of 222 newborn children from paediatric department it was found that 90.09% babies are normal in both ears and 2.7% babies are refer in both ear.
In ENT outdoor, otoacoustic emission (OAE) test will be done on the suspected deaf cases, ages below 5 years. Then referred cases are undergoing for BERA test to confirm deafness. Puretone audiometry test will be done on the suspected deaf cases ages 5 to 15 years. Then referred cases are undergoing for BERA test to confirm deafness. Out of twenty two cases 3 cases had syndromic deafness i.e they had Down syndrome and cerebral palsy. Remaining 19 cases were diagnosed as non-syndromic deaf children.

Another study showed that a multi-step OAE screening protocol led to the identification of children who were ultimately diagnosed with a wide range of hearing-health conditions warranting monitoring and treatment. The 5.7% fail/refer rate compares favorably with rates reported by effective hospital-based newborn hearing screening programs[16] given that the fail/refer rate for infants and toddlers is expected to be higher than newborns due to transient middle-ear conditions that are more prevalent in this population. The data-derived positive predictive value of negative 67.3% indicates that over-referral was not occurring.[16]

An objective screening tool, OAE technology holds great promise for health and early education care providers in reliably screening infants and toddlers for hearing loss during the critical language-learning years. As educational program directors and individual practitioners make decisions about how to meet children's hearing-health needs.

One problem we faced was getting a noiseless surrounding in the nursery setting. The babies had hence to be transported to the audiology room for testing which increased the discomfort for the relatives. Some babies woke up during transit, increasing the time taken for the test. To improve the follow-up rate, we coincided the immunization visit with that of screening. Performing a test on that day was a little time consuming because one has to wait for the baby to go to natural sleep.[17]

A hearing screening equipment facility in every hospital with a maternity unit today may not be an economically viable proposition. A program with centralized screening facility, where a screener would operate out of one hospital, to cater to the different hospitals of the city was success- fully implemented with the co-operation of IAP.[18] This is a viable and cost effective model for the whole country.

CONCLUSION

With our limited data, it is too early to arrive at any conclusions or definite interpretations yet. Our unique experience is still evolving. Our Hospital needs to 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 should be detected. Hearing impairment may develop after the neonatal period and therefore, it is crucial for the paediatrician to encourage parents to continue to have their child's hearing checked. The paediatrician 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.

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