A STUDY OF NEWBORN HEARING SCREENING USING OTOACOUSTIC EMISSIONS.

[1]Nikita Chaudhari, [2]R.G.Aiyer, [3]Rahul Gupta, [4]Jayman Raval

[1]Resident Doctor, [2]Professor & Head, [3]Associate Professor, [4]Assistant Professor, Department of Otorhinolaryngology and Head-Neck Surgery, Medical College, Baroda, Gujarat, India

ABSTRACT

Background: Congenital hearing loss has recently been recognized as one of the most common birth defect present in newborns, with a prevalence of permanent hearing loss ranging from 2-3/1000 live births.

Aims & Objectives: The present study was undertaken to know the prevalence of congenital hearing loss in normal neonates as well as in high risk neonates using otoacoustic emissions.

Methods: The present study was prospective, observational study, which was carried out in the Department of Otorhinolaryngology and Head-Neck Surgery, Medical College and S.S.G Hospital, Baroda from December 2014 to December 2016. A total of 709 Newborn babies were included in this study. These 709 Newborn babies were subjected to 2 stages DPOAE (Distortion Product Otoacoustic Emissions). Newborn babies were subjected to a 1st DPOAE screening within the first week of life if they don't have any high risk factors and those Newborn babies who were admitted in NICU for more than 5 days were examined after being discharged from NICU. For those babies who pass 1st DPOAE, no further testing was done. For those babies who refer 1st DPOAE, repeat DPOAE testing was done after 15 days, failing which such newborn baby was subjected to BERA (Brainstem Evoked Response testing to confirm hearing loss).

Results: Seven hundred and nine newborn babies were screened by DPOAE. 19 newborn babies had refer result for 1st DPOAE hearing screen and for these infants repeat DPOAE screens was done after 15 days. On repeat DPOAE testing, 2 infant gave refer result. Amongst 2 newborns who failed the final OAE test, one newborn had normal hearing in BERA testing and another one had moderately severe sensorineural hearing loss in right ear and mild sensorineural hearing loss in left ear. Hence the prevalence of hearing, loss of 1.41 per thousand was detected in newborn babies examined.

Conclusion: Initial DPOAE screening followed by BERA examination in referingr cases is helpful for early identification of infants with hearing loss, hence allowing for timely intervention. It is necessary to secure the holistic development of the child by detecting hearing loss at birth and providing remedial measures at the earliest. At present there are no national policies to this effect although under NPPCD there is a planned provision of hearing aids for children. We need to identify those with mild to moderate hearing loss that are amenable to treatment through a universal Newborn Screening Program.

Key-words: Distortion product otoacoustic emission (DPOAE), Newborn Hearing Screening, Otoacoustic Emissions (OAE).

Corresponding Address: Dr. Nikita G. Chaudhari, Resident, Department of Otorhinolaryngology and Head-Neck Surgery, Medical College, Baroda-390001, Gujarat, India. E-mail: nikitachaudhari7631@gmail.com

INTRODUCTION

Congenital hearing loss has recently been recognized as one of the most common birth defect present in newborns, with a prevalence of permanent hearing loss ranging from 2-3/1000 live births.[1] The reported prevalence of permanent bilateral hearing loss identified by newborn hearing screening programs was 1.61/1000 of at-risk infants in India, & 1.83/1000 in USA (Washington DC).[2] Congenital Cholesteatoma, Ossicular discontinuity, fluid in the middle ear are causes for congenital conductive Hearing Loss**.** Sensorineural hearing loss is divided into Nonsyndromic Sensorineural Hearing Loss and Syndromic Sensorineural Hearing Loss. Two third of the congenital hearing loss are non syndromic. Nonsyndromic SNHL is further classified by the mode of inheritance. Rare modes of transmission include X-linked and mitochondrial transmission, which account for the remaining 2% of hearing impairment.

Alport's syndrome Branchio-oto-renal syndrome, Jervell and Lange-Nielsen syndrome, Pendred's syndrome, Stickler's syndrome, Treacher Collins, Usher' syndrome, Waardenburg's syndrome, & Congenital Rubella Syndrome are the causes of Syndromic Sensorineural Hearing Loss. Perinatally acquired causes of hearing loss includes Hyperbilirubinemia, Ototoxic Drug Usage, Meningitis, Hypoxic Encephalopathy, Sepsis, Head Trauma, Mechanical Ventilation and Extra Corporal Membrane Oxygenation.[3] The permanent hearing loss identified in newborn screening programs varies from a minimum level of 40dBHL in the United Kingdom to 35dBHL in the United States. The Joint Committee on Infant Hearing (JCIH, 2000) define the target population for infant screening programs as unilateral or bilateral permanent hearing loss averaging 30-40dB in the speech frequency range. Conductive hearing loss, as a result of anomalies to the outer or middle ear, is also included in the targeted screening population.[4]

A number of neonates identified through early universal newborn hearing screening programs provided the evidence to demonstrate that early identification and intervention of newborns that were deaf or hard of hearing could actually achieve nearly normal language acquisition by three years of age. The researchers analyzed many demographic factors (e.g. degree of hearing loss, race/ethnicity, economic status, gender, and mode of communication) and found early identification was the key to improved language outcomes. Six months of age were the critical cut off period for early identification that would achieve normal speech and language development. Neonatal screening with Otoacoustic emission helps in early identification of congenital hearing loss and hence early intervention in cases of newborns who

were found deaf or hard of hearing. This helps them to achieve nearly normal language.[5]

The origins of newborn hearing screening can be traced to Sweden. In 1956, Wedenberg reported that the most easily observable response to sound is the auropalpebral reflex, i.e. a rapid and distinctive closing of eyelids when they are open or screwing their eyes if they are closed.[6] Marion Downs approached the American Speech and Hearing Association (ASHA) with a request that a national joint committee be formed for the evaluation of the status of the newborn hearing screening programme in 1969. This committee came to be known as the Joint Committee on Infant Hearing (JCIH).[7]

Earlier studies aimed at detection of hearing loss only in high risk groups. But studies showed that by screening only at-risk population, 30-50% of hearing loss will be missed as they can occur in well babies also.[8] JCIH recommended that the hearing of all infants should be screened at no later than 1 month of age, those that do not pass screening should have a comprehensive audiological evaluation at no later than 3 months of age. They had also recommended that Infants with confirmed hearing loss should receive appropriate intervention latest by 6 months of age. Regardless of previous hearing-screening outcomes, all infants with or without risk factors should receive ongoing surveillance of communicative development beginning at 2 months of age during well-child visits in the medical home.[9]

MATERIAL & METHODS

The present study was a prospective and observational study, which was carried out in the Department of Otorhinolaryngology and Head-Neck Surgery, Medical College and S.S.G Hospital, Baroda from December 2014 to December 2016. Approval from Scientific and Ethical Research Committee of the institute was taken and informed written consent about the study was taken from either parent of each newborn before enrolling them in to study. All parents were provided information about the disease, treatment modalities, and importance of regular follow up in their local language. A total of 709 Newborn babies were included in our study.

Inclusion criteria

1) Babies who, delivered in S.S.G. Hospital
2) Those babies who required intensive care management were not included in the study during the acute phase. However, they were included after stabilization or before discharge.
3) Babies whose parents gave written and informed consent.

Exclusion criteria

1) Babies whose parents did not give consent for enrollment in the study.

Procedure of the test: The parents were counseled regarding congenital hearing loss and the need for early diagnosis and intervention prior to the test. Written informed consent was obtained from the parents. The babies underwent a routine ENT examination consisting of inspection

of the pre-aural, pinna, and post aural region. Occluding wax or debris were gently cleaned using cotton tipped swab and otoscopic examination of the tympanic membrane was conducted using Heine 3000 series otoscope with plastic speculums and findings were noted in predesigned proforma containing newborn's details (gestational age, birth weight, date of birth, Duration of labour, Presentation, Mode of delivery, APGAR Score, Meconium aspiration, NICU Admission, Post Natal infections, CNS Diseases, Hyperbilirubinemia, Birth Trauma) as well as mother's details (H/O anaemia, Diabetes Mellitus, Thyroid Dysfunction, HIV, VDRL, TORCH, PIH, Hydramnios, Chorioamnionitis). 2 stage OAE done on the newborns with 2, 3, 4, and 6 kHz frequency in both ears. Newborns who refer 1st OAE in any ear, examined for 2nd OAE in both ears. Newborns who refer, 2nd OAE examined for BERA.

Testing environment: The babies were then tested in a sound treated room in the audiology department. The babies were tested in a supine position, preferably on the guardian's lap and preferably when the child was asleep. The test was conducted by a qualified audiologist.

Instrumentation: The machine used for this test was of Otodynamics Company with model DPECOPort and software containing ILO292USB. The software was connected to a computer for data collection and data analysis. The system was calibrated using the calibration mode in the software. Daily calibration of the Otoacoustic emission probe was performed to ensure the infants were screened with a functioning probe. During the measurement, two pure tone stimuli (f1 and f2), where f2 was higher than f1were presented with f2/ f1 ratio at approximately 1.22 (range 1.21 to 1.23) to obtain a robust DPOAE response in human's ears.

The f2 frequencies were tested on a 2 point per octave manner, from 2 kHz to 6 kHz. Two stimuli were presented at an asymmetrical intensity level of L1= 65 dBSPL and the second intensity, L2=55dBSPL (such that L1>L2) with the probe tip in place and the check fit procedure passed, DPOAEs were initiated. The DPOAE amplitude and noise floor adjacent frequency regions of distortion product 2f1-f2 were recorded.

Sequence of the testing: The first test was done using distortion product otoacoustic emissions. The probe was fitted with a standardized infant ear tip kit. These probes are made of soft rubber.

The ear tip was gently inserted into the right ear by a gentle traction on the pinna in a backward and downward direction. Once the probe tip was in place the test was started. First the probe fit and seal was checked, followed by any extrinsic noise levels in a systematic computerized manner preloaded in the software.

Procedure: The test was carried out in a sound treated room (Audiology Room, Ward 19, Department of Otorhinolaryngology and Head & Neck Surgery). The baby was observed for a short period prior to the presentation of the stimulus. All the newborns were checked with DPOAE. Those newborns that

responded favorably to DPOAE testing were labeled as normal hearers, but those who failed, underwent a 2nd DPOAE testing after 15 days. Newborns that responded positively to 2nd DPOAE testing were labeled as normal hearers. Those babies who failed a second DPOAE underwent confirmative BERA tests. All results were recorded in proforma and were analyzed.

RESULTS

In this study Male (53.03%) / Female (46.97%) ratio was 1.13. This difference in sex ratio was statistically insignificant. In this study most of the newborns (66.99 %) were having weight between 2 to 3 kg followed by 1 to 2 kg (16.93%), 3 to 4 kg (15.51%) and 0 to 1 kg as well as 4 to 5 kg (0.28%). In this study most of the newborns (80.54%) were having a full term gestation. In this study, a majority of the newborns (52.60%) was having no high risk factors. The most common high risk factor in our study (42.26%) was low birth weight (<2.5 kg) followed by (30.05 %) low birth weight along with preterm delivery.

In the present study most common high risk factors were LBW and Preterm deliveries in both the sexes and the majority of the newborns 690 (97.32%) had passed the 1st OAE test. Amongst 19 newborns who had failed 1st OAE test, most of the newborns 17 (89.47%) had passed the 2nd OAE test in this study. Amongst 2 newborns who failed the 2nd OAE test, one newborn had normal hearing in BERA testing and another one had moderately severe sensorineural hearing loss in right ear and mild sensorineural hearing loss in left ear. Both the newborns who failed even a 2nd OAE test were having LBW as a risk factor. Maternal Thyroid dysfunction, Neonatal Hyperbilirubinemia and maternal PIH were the additional risk factors apart from the LBW in one of these babies who had failed the 2nd OAE test.

Table No: 1 High risk factors

Sr. No.	High Risk Factor	No. of Newborns
1	Maternal Anemia	03
2	Maternal Thyroid Dysfunction	02
3	Maternal PIH	01
4	Maternal Hydramnios	05
5	CPD	02
6	Preterm	17
7	LBW	142
8	Meconium Aspiration	01
9	Neonatal Hyperbilirubinemia	03
10	Neonatal Respiratory Distress	17
11	Neonatal Pyogenic Meningitis	01

12	Maternal Anemia + Maternal PIH	01
13	LBW + Maternal Anemia	01
14	Maternal DM + Preterm	01
15	Maternal Thyroid dysfunction + LBW	01
16	Maternal Herpes + LBW	01
17	CPD + LBW	01
18	LBW + Preterm	101
19	Preterm + Maternal PIH	01
20	LBW + Maternal PIH	02
21	LBW + Maternal Hydramnios	04
22	LBW + Neonatal Respiratory Distress	01
23	LBW + Neonatal Hyperbilirubinemia	03
24	Maternal Hydramnios + Neonatal Respiratory Distress	02
25	Maternal HIV + Preterm + LBW	01
26	Maternal Herpes + LBW + Preterm	01
27	LBW + Preterm + Maternal PIH	01
28	LBW + preterm + Neonatal Respiratory Distress	03
29	LBW + Preterm + Maternal Anemia	02
30	LBW + Maternal Anemia + cervical polyp	01
31	LBW + preterm + Neonatal Hyperbilirubinemia	07
32	LBW + Neonatal Respiratory Distress + maternal hydramnios	01
33	LBW + Preterm + Maternal Anemia+ Maternal PIH	01
34	LBW + Preterm + Maternal Thyroid dysfunction + maternal hydramnios	01
35	LBW + preterm + Neonatal Respiratory Distress + Maternal PIH	01
36	LBW + Neonatal Respiratory Distress + Neonatal Hyperbilirubinemia + pneumonia	01
37	LBW + Maternal Thyroid dysfunction + Maternal PIH + Neonatal Hyperbilirubinemia	01
	Total	**336**

Table No: 2 Sex distribution in babies with high risk factor

Sr. No.	High Risk Factor	Male	Female
1	Maternal Anemia	01	02
2	Maternal Thyroid Dysfunction	00	02
3	Maternal PIH	01	00
4	Maternal Hydramnios	03	02
5	CPD	00	02
6	Preterm	09	08

7	LBW	67	75
8	Meconium Aspiration	00	01
9	Neonatal Hyperbilirubinemia	03	00
10	Neonatal Respiratory Distress	11	06
11	Neonatal Pyogenic Meningitis	01	00
12	Maternal Anemia + Maternal PIH	01	00
13	LBW + Maternal Anemia	01	00
14	Maternal DM + Preterm	00	01
15	Maternal Thyroid dysfunction + LBW	00	01
16	Maternal Herpes + LBW	00	01
17	CPD + LBW	00	01
18	LBW + Preterm	48	53
19	Preterm + Maternal PIH	00	01
20	LBW + Maternal PIH	02	00
21	LBW + Maternal Hydramnios	02	02
22	LBW + Neonatal Respiratory Distress	01	00
23	LBW + Neonatal Hyperbilirubinemia	01	02
24	Maternal Hydramnios + Neonatal Respiratory Distress	02	00
25	Maternal HIV + Preterm + LBW	01	00
26	Maternal Herpes + LBW + Preterm	01	00
27	LBW + Preterm + Maternal PIH	01	00
28	LBW + preterm + Neonatal Respiratory Distress	01	02
29	LBW + Preterm + Maternal Anemia	01	01
30	LBW + Maternal Anemia + cervical polyp	00	01
31	LBW + preterm + Neonatal Hyperbilirubinemia	02	05
32	LBW + Neonatal Respiratory Distress + maternal hydramnios	01	00
33	LBW + Preterm + Maternal Anemia+ Maternal PIH	00	01
34	LBW + Preterm + Maternal Thyroid dysfunction + maternal hydramnios	01	00
35	LBW + preterm + Neonatal Respiratory Distress + Maternal PIH	01	00
36	LBW + Neonatal Respiratory Distress + Neonatal Hyperbilirubinemia + pneumonia	00	01
37	LBW + Maternal Thyroid dysfunction + Maternal PIH + Neonatal Hyperbilirubinemia	00	01
	Total	**164**	**172**

Table No: 3 High risk babies posted for 2nd OAE

Sr. No.	High Risk Factor (If Present)	Pass	Refer
1	Maternal Thyroid Dysfunction	01	00
2	Neonatal Hyperbilirubinemia	02	00
3	LBW	01	01
4	Meconium Aspiration	01	00
5	LBW + Maternal Hydramnios	01	00
6	LBW + Preterm	03	00
7	LBW + Neonatal Hyperbilirubinemia	02	00
8	LBW + Maternal Thyroid dysfunction + Maternal PIH + Neonatal Hyperbilirubinemia	00	01
9	LBW + Preterm + Maternal HIV	01	00
10	LBW + Neonatal Hyperbilirubinemia + Preterm	03	00
11	LBW + Maternal Thyroid dysfunction + Preterm + Maternal Hydramnios	01	00
12	NO HIGH RISK FACTOR	01	00
	Total	**17**	**02**

Table No: 4 Profile of both babies who had failed 2nd OAE test:

	Birth weight (Both were LBW)	Maternal Thyroid Dysfunction	Maternal PIH	Neonatal Hyperbilirubinemia
1st Baby	1880 gm	No	No	No
2nd Baby	1300 gm	Hypothyroidism	Yes	Total Billirubin : 4 Indirect Billirubin:1.6 Direct Billirubin: 8.8

DISCUSSION

In this study, we found that most of the newborns (53.03%) were male. Study by Shreeya Kulkarni et al [10], Christie Ohl et al [11], Girish Mishra et al [12], Boo N Y et al [13], & Joyce Pascal Rozario et al [14] also favored this study. In this study, we found that most of the newborns (66.99 %) were having weight between 2 to 3 kg. Study by Shreeya Kulkarni et al [10], & C R Kennedy et al [13] also favored this study. In this study, we found that most common high risk factor (42.26%) was low birth weight (<2.5 kg). Study by Shreeya Kulkarni et al [10], Joyce Pascal Rozario et al [14] and Jean L. Jhonson et al [15] also favored this study.

In this study most of the newborns (84.86%) were having term gestation. Study by Shreeya Kulkarni et al [31, 10], B. De Capua et al [16], & Boo N Y et al [13] also favored this study.

In this study, we found that most of the newborns (97.32%) had passed the 1st OAE test. Study by Shreeya Kulkarni et al [10], Glen et al [17], Joyce Pascal Rozario et al [14] & B. De Capua et al[16] also favored this study. Amongst 17 newborns who had failed 1st OAE test, most of the newborns

(89.47%) had passed the 2nd OAE test in this study. Study by Shreeya Kulkarni et al[16] , A.Ciorba et al [17], Joyce Pascal Rozario et al [14] & B. De Capua et al [16] also favored this study.

In this study, we found that prevalence rate of hearing loss in high risk patient was 1.41 / 1000 newborns. Study by Fortnum et al [18]. found the prevalence rate of 1.1, Cynthia C. Morton et al [19] found it 1.33, Kumar A et al[20] found it 1.61, Renitha R et al[21] found it 1 and Kriek F et al[22] found it 1.86.

CONCLUSION

Newborn hearing screening is vital in recognizing babies born with congenital hearing loss. Distortion product otoacoustic emissin (DPOAE) is an excellent mass screening device for large country like India & Universal newborn screening should be implemented in each and every institution. Indian association of Paediatrics (IAP) also recommends that if hearing loss is screened by OAE and confirmed by BERA, next step should be hearing aid using at the earliest. Hearing aids may be fitted for infants as early as 2 months of age. This should be followed with auditory training and speech therapy. Children with profound deafness who drive negligible benefit from conventional amplification wiith hearing aids may be considered for chochlear implants.

JCIH recommended that the hearing of all infants should be screened at no later than 1 month of age, those that do not pass screening should have a comprehensive audiological evaluation at no later than 3 months of age. They had also recommended that infants with confirmed hearing loss should receive appropriate intervention latest by 6 months of age. Regardless of previous hearing screening outcomes, all infants with or without risk factors should receive ongoing surveillance of communicative development beginning at 2 months of age during well child visits in the medical home.

To conclude, it is necessary to secure holistic development of the child by detecting hearing loss at birth and providing remedial measures at the earliest. At present there are no national policies to this effect although under NPPCD there is a planned provision for hearing aids to children. We need to identify those with mild to moderate hearing loss that are amenable to treatment through a universal Newborn Screening Program.

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