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ORIGINAL ARTICLE

Assessment of Hearing in High Risk Infants, Using Brainstem Evoked Response Audiometry

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Abstract Hearing plays a basic and important role in language, speech and intellectual development. A hearing impaired child develops psychological, social, educational and cognitive problems. The aim is to evaluate the hearing in high-risk neonates using BERA. 50 High risk admissions in the NICU and high- risk infants presenting to the OPD of our hospital for a period of 2 years were included in the study. Of the 50 high-risk infants, 44 (88%) cases had impaired BERA, remaining 6 (12%) had normal hearing threshold. LBW and hyperbilirubinemia were found to have strong relationship with hearing impairment (p value 0.04 and 0.03 respectively) and OR of 1.98 and 1.75 respectively. Elevated auditory threshold was found more frequently in infants with multiple clinical adverse factors than in those having single risk factor (36/50 vs. 8/50, $p < 0.009$). Proportion of infants with impaired BERA was high in infants with low birth weight and hyperbilirubinemia in exchange range. Screening by BERA at an early age is beneficial for early diagnosis of hearing impairment, so that possible interventions can be used as early as possible and prevent developmental delays in newborns.

Keywords BERA · High risk infants · LBW · Hyperbilirubinemia

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Introduction

Hearing is necessary to learn languages and speech and to develop cognitive skills. It helps the developing child to learn to recognize sounds, identify objects and internalize concepts. Hearing loss has the highest incidence amongst any pediatric disability and should be detected as early as possible. The importance of timely referral of children with suspected hearing loss for a full hearing assessment cannot be overemphasized. As hearing is important for normal educational and social development, hearing loss can be devastating [1]. In India 5–15% of high risk neonates are prone to hearing loss as compared to 1–2 out of 1000 normal newborns [2]. Hearing impairment is one of the important causes of developmental delay in children and, if not diagnosed and treated promptly, it could result in speech delay.

The use of informal methods of hearing assessment, such as the whisper test, can lead to late diagnosis of hearing loss and should be discouraged. With modern screening and diagnostic equipment, hearing can and should be quantified. This can be accomplished in a screening and/or a diagnostic hearing assessment [3]. Implementing high-risk neonatal screening, detecting hearing loss prior to 3 months and intervention prior to 6 months will result in a better speech performance of neonates [4].

The aim of the our study is to evaluate the performance of brainstem evoked response audiometry (BERA), an electro-physiological test which studies the electrical potential generated at the various levels of the auditory system starting from cochlea to cortex, in detecting hearing loss in high risk neonates. BERA has expanded the possibility of objective testing of hearing. This is an effective and simple method that requires less cooperation of the

patient and measures the specific part of the auditory pathway. It is not significantly affected by the state of consciousness, drugs and environmental factors like the sensory input to the cortex.

Materials and Methods

Aim of the study is to evaluate the hearing in high-risk neonates using brainstem evoked response audiometry (BERA) and to confirm the hearing impairment and its magnitude in the abnormal study subjects using a follow up BERA at the third month.

A hospital based prospective study was conducted on 50 NICU stabilized and discharged neonates at neonatal intensive care unit (NICU) and high- risk infants presenting to the OPD at our Hospital.

High Risk Neonates with Gestational age < 37 weeks, Birth weight < 1.5 kg, Hypoxic Ischemic Encephalopathy Grade II/III, Hyperbilirubinemia, Bacterial meningitis, those having family history of hearing impairment, consanguinity, congenital perinatal infection, Anatomical malformations of head and neck, Birth trauma at the time of delivery, Ototoxic drug intake during pregnancy of mother, Exanthematous fever to mother during pregnancy were included in the study. All NICU babies were considered at risk, and hence were included in the study. Healthy term neonates were excluded from the study.

The following information was recorded before performing BERA: gestational age, sex, birthweight, final diagnosis, the period of hospitalization and possible risk factors for hearing impairment. A complete ENT check-up was done to rule out wax, ear infection, middle ear problems.

Procedure of BERA

Brainstem evoked response audiometry (BERA) checks how the brainstem(the part of the nerve that carries sound from the ear to the brain) and the brain respond to the sound. It is performed in a dust free, sound free and air conditioned room free of electromagnetic disturbances. The forehead, vertex and both mastoid regions are cleaned with spirit. External auditory canals are cleaned and the infant is made to wear earphones, and surface electrodes are placed on the head and ears.(Fig. 1) A mild sedative (Syrup Trichlofos (Pedichloryl)0.5–1 mg/kg body weight), if required, will be given to keep the infant calm and quiet during the test. The neck is slightly flexed to minimize any myogenic activity. Auditory click stimuli delivered at the rate of 28/s. The test takes about 30–45 min under optimum conditions. The sounds are sent through the earphones and the electrical activity in the infant's brain is recorded when he or she perceives the sound. The hearing impairment is graded as mild, moderate, severe and profound.

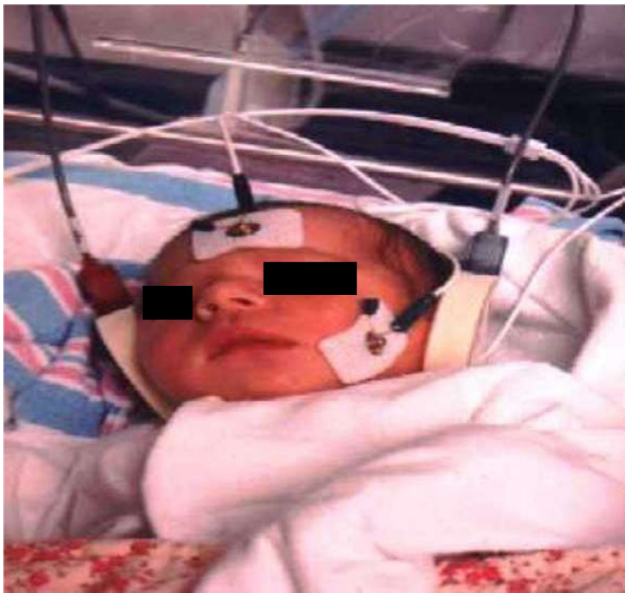

Fig. 1 BERA in progress in a newborn

Statistical Analysis

Statistical package for social sciences (SPSS) for Windows Version 22.0 Released2013. Armonk, NY: IBM Corp., will be used to perform statistical analysis. Descriptive analysis of all the explanatory and outcome parameters was done using frequency and proportions. Chi Square test was used to observe for the association between BERA and gestational maturity, birth weight, presence of different risk factors. Multivariate logistic regression analysis was performed to assess risk factors (Odd's Ratio) of different factors of BERA positive among study subjects. The level of significance was set at $p < 0.05$.

Observation and Results

The present study was conducted at Dr. B.R. Ambedkar Medical College and Hospital, Bangalore. Fifty high-risk infants were considered for the study. All the infants had undergone screening with BERA. The study subjects were divided into four groups based on their age. Among the 50 study subjects, 34 (68%) were males and 16 (32%) were females.

Among the 50 high-risk infants subjected to BERA, 44(80%) had hearing loss and only 6(12%) had normal hearing. Infants with hearing impairment were classified into mild, moderate, severe and profound.

Out of the 50 high risk infants, 22 (44%) were preterms, 23 (46%) had low birth weight, 16 (32%) had hyperbilirubinemia, 18 (36%) were born out of consanguineous marriage, 6 (12%) had family history of hearing loss, 11 (22%) had a history of exposure to ototoxic drugs, 2 (4%) had perinatal infections, 3 (6%) suffered from Hypoxic-Ischemic Encephalopathy and 1 (2%) had anatomical malformation of the ear.

Among the high-risk infants with hearing impairment, 19 (43.2%) were preterms and 25 (56.8%) were term births. Among the high-risk infants with hearing impairment, 33 (75%) were low birth weight and 11 (25%) were > 1.5 kgs, which was statistically significant ($p < 0.05$).

Among the high-risk infants with hearing impairment, 14 (31.8%) had hyperbilirubinemia with hearing impairment, 15 (34.1%) were born out of consanguineous marriage and had some degree of hearing impairment, 5 (5%) had hearing impairment with a history of family history of hearing loss, 10 (22.7%) with hearing impairment had a history of exposure to ototoxic drugs, 2 (4.5%) with a history of perinatal infections had hearing impairment, 3 (6.8%) with Hypoxic Ischemic Encephalopathy had hearing impairment and 1 (2.3%) with anatomical variation had hearing impairment. (Table 1).

Out of the 50 high-risk infants analyzed in the study, 8 (18.2%) who had a single risk factor had hearing impairment and 36 (81.8%) who had multiple risk factors had hearing impairment. (Table 2).

From the above mentioned data, it was inferred that infants with birth weight < 1.5 kg were 1.98 times more likely to develop hearing loss and infants with hyperbilirubinemia in the neonatal period were 1.75 times more likely to develop hearing loss. (Table 3).

Discussion

Hearing loss is one of the most common congenital defects occurring in approximately 2–4 infants per thousand live births [5]. When children are identified with hearing loss at birth and receive intervention before 6 months of age, they catch up with their normal hearing peers and develop essentially normal language by five years of age [6, 7]. Conversely, children who are identified with hearing loss later in life and receive intervention after 6 months of age, especially those with severe to profound hearing loss and with multiple handicaps struggle to catch up with their normal hearing peers.

The screening strategies of young infants, specially, universal screening versus selective screening (high risk targeted approach) is a debate especially in resource limited settings. The fact that selective screening may miss considerable number of cases is the justification for universal screening in less developed countries also.

According to different studies, the incidence of hearing loss in normal newborn population is 1–2 in 1,000 live births. According to one study, the speech performance of the infants whose hearing problem was detected and treated before they were six months old, was the same as normal age-matched children [4].

Table 1 Association between different risk factors and BERA using Chi Square test

Risk factors	Categories	Positive		Negative		Chi^2 value	p value
		n	%	n	%		
Hyperbilirubinemia	Present	28	63.6	1	16.7	4.782	0.03*
	Absent	16	36.4	5	83.3		
Consanguinity	Present	15	34.1	3	50.0	0.580	0.45
	Absent	29	65.9	3	50.0		
Family h/o hearing loss	Present	5	11.4	1	16.7	0.141	0.71
	Absent	39	88.6	5	83.3		
H/o drug intake	Present	10	22.7	1	16.7	0.113	0.74
	Absent	34	77.3	5	83.3		
Perinatal infections	Present	2	4.5	0	0.0	0.284	0.59
	Absent	42	95.5	6	100.0		
HIE	Present	3	6.8	0	0.0	0.435	0.51
	Absent	41	93.2	6	100.0		
Anatomical malformation	Present	1	2.3	0	0.0	0.139	0.71
	Absent	43	97.7	6	100.0		

* p value < 0.05

Table 2 Association between BERA positive and presence of multiple risk factors using Chi square test

Risk factors	Positive		Negative		Chi^2 value	*P* value
	n	%	n	%		
Single	8	18.2	4	66.7	6.805	0.009*
Multiple	36	81.8	2	33.3		

Table 3 Multivariate logistic regression analysis to assess the risk factors (Odds ratio) of different factors of BERA Positive among study subjects

Risk factors	OR	95% C.I. for OR		*p* value
		Lower	Upper	
Preterm	1.20	0.14	10.46	0.87
Low birth-weight	1.98	0.67	2.18	0.04*
Hyperbilirubinemia	1.75	0.53	1.95	0.04*
Consanguinity	2.05	0.17	25.15	0.57
Family history	3.15	0.10	102.95	0.52
Ototoxic drug exposure	1.10	0.04	29.94	0.96
Perinatal infection	1.23	0.11	10.59	1.00
HIE	1.18	0.14	12.96	1.00
Anatomical malformations	1.58	0.00	30.58	1.00

* p value < 0.05

Hyperbilirubinemia is one of the major risk factor accounting for hearing loss. Sharma et al. [8] conducted a study on BERA in hyperbilirubinemia neonates and concluded that BERA is a simple, reliable and effective technique for determining auditory functions in the neonates especially changes of early bilirubin toxicity.

Zamani et al. carried out a study to estimate the hearing loss in high risk neonates, concluding that BERA is one of the most important methods for surveying hearing loss in neonates and is standard test for screening hearing loss in neonates, with high sensitivity and specificity. They found that, out of 230 high risk cases, 18 (8%) had sensorineural hearing loss [8, 9]. Chadha and Bias [10] studied brainstem responses of high risk neonates from NICU and suggested that all high risk neonates should undergoscreening for hearing impairment. Aiyer and Parikh [11], in their study, concluded that retesting of infants with abnormal initial BERA within 3 months is important.

All of these studies show the importance of early detection and treatment of hearing impairment in children. Unfortunately the age of detection of hearing loss is delayed even in developed countries if the screening for hearing impairment is not performed during the first few months of life. Early detection and rehabilitation of hearing loss produces worthwhile benefit in terms of improved speech and language provides the rationale for universal screening of neonates and infants across the world and also maximizes linguistic competence and literacy development for children who are deaf or hard of hearing.

BERA being rapid, easy and cheap test gives the electrophysiological response of hearing without any need for assessment of newborn behavior. The result of this test is not affected by sedatives, which was used during the test. It is thus a useful and non-invasive tool for hearing assessment in newborn and infants [12].

The reported sensitivity of the BERA for hearing assessment was 100% and specificity around 86% [13]. We observed that infants exposed to high risk factors are prone for developing hearing abnormality. Among all the risk factors included in our study, low birth weight and neonatal hyperbilirubinaemia carry a much higher risk of hearing abnormality. The risk indicators associated with permanent congenital, delayed-onset or progressive hearing loss in childhood as defined by the 2007 JCIH position statement [14] (Table 4).

Absolute latencies of wave III, wave V and IPL of I-V were increased among the high risk group. Absolute latency of wave V is a consistent and stable parameter, which has received primary attention as a valuable factor in response evaluation. It is suggested that all the risk factors which bring the neonate under intensive care, induce a certain amount of hypoxia of the cochlea and brainstem

Table 4 The risk indicators associated with permanent congenital, delayed-onset or progressive hearing loss in childhood as defined by the 2007 JCIH position statement

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 regardlessof length of stay: ECMO, assisted ventilation, exposure to ototoxic medcations (Gentamycin and Tobramycin) or loop diuretics (Furosemide/Lasix) and hyperbilirubinemia that requires exchange transfusion
4	In-utero infections such as 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 are 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 like Waardenburg, Alport, Pendred and Jervell and Lange-Nielson
8	Neurodegenerative disorders such as Hunter syndrome or sensory motor neuropathies, such as Friedreich ataxia and Charcot-Marie- Tooth syndrome
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
11	Chemotherapy

which leads to various cellular changes such as edema, degeneration and necrosis. Hence, they predispose to hearing impairment which may be reversible following reversal of hypoxic changes. Hypoxemia has been identified as a possible ototoxin according to Duara et al. [15], Leech and Alvird [16] concluded that brainstem auditory nuclei are particularly susceptible to acute hypoxic insults in the neonates.

The I-V IPL is a reflection of neural conduction time between the auditory nerve and brainstem nuclei and reflects upon the efficiency of the auditory pathway. Prolonged I-V IPL is a feature of neurological and is an indication of delay in neural conduction within the brainstem [17]. Thus hearing assessment by BERA at an early age in all high risk infants is very beneficial and can reduce morbidity associated with hearing impairment.

Conclusion

In our study, low birth weight and hyperbilirubinemia were found to be the significant risk factors. Family history of hearing loss, craniofacial anomalies and intrauterine infection administration of ototoxic medication, NICU admissions for more than 5 days and maternal diabetes mellitus, hypothyroidism are other risk factors associated with neonatal hearing loss.

Universal neonatal hearing screening (UNHS) has become a national practice in most developed countries. The identification of all newborns with hearing loss before 6 months of age has now become a realistic and attainable goal.

This study was also able to determine the additional risk factors in a hearing screening program can provide an efficacious alternative to the use of UNHS. The full potential of technological advances that have been made in diagnostic audiology and rehabilitation and treatment of hearing impaired can only be rightfully exploited if early identification of hearing impaired babies is meticulously carried out.

Compliance with Ethical Standards

Conflict of interest The authors declare that they have no conflict of interest.

Ethical Approval Obtained from the ethical committee of our institution.

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