INCIDENCE EVALUATION OF SNHL IN HIGH RISK NEONATES

Shilpi Arora¹, L. K. Kochhar²

ABSTRACT : *Objective : to evaluate hearing in high-risk neonates to find the incidence of congenital and early acquired sensory-neural hearing loss.*
Study Design and Setting : The study was designed as a consecutive cases, hospital based study in a tertiary care hospital.
Patients : The subjects were 70 normal born neonates and 70 high-risk neonates. The 70 neonates with various high risks included in the study had family history of deafness, prematurity, IUGR, asphyxia, perinatal infections, hyperbilirubinemia, neonatal sepsis, meningitis, ototoxicity and fetal malformations.
Interventions : The study was conducted over a period of two years from Sep 2000 to Aug 2002. BERA using Octavus neuro-otological computer and a main unit BERA module with integrated pre-amplifier was performed within first 28 days of life and at 6 months follow up.
Results : 44 out of 140 neonates showed abnormalities on initial BERA testing. 28 ears out of 67 ears (44 neonates) had losses confirmed at repeat BERA. Wave V was considered to determine the threshold of hearing and was found to be consistently present in neonates and also the wave . Hyperbilirubinemia (18 cases) and prematurity (29 cases) were the most commonly observed. The cases of Hyperbilirubinemia showed raised thresholds, absolute latencies and Wave I abnormalities (suggesting affection of eighth nerve). In the cases of Prematurity, 8 ears showed raised threshold at follow up. Asphyxia was found in 5 cases and is known to affect the auditory nerve. Family history of deafness found in 3 cases showed X-linked inheritance in one case and autosomal recessive inheritance in the others. Perinatal Rubella was observed in two cases, which showed hearing loss. Congenital malformations were seen in 7 cases (Waardenberg, Downs, Hydrocephalus and T-O Fistula). Neonatal Sepsis was observed in 3 cases and is known to affect the eighth nerve. IUGR was observed in 3 cases, two had normal hearing while one had no waveforms.
Conclusion : BERA should be used as a screening tool to test the auditory function in all high-risk neonates.

The estimated incidence of hearing loss in newborn infants ranges from 0.001-0.5% and may be as high as 1-5% in high-risk infants (Anand 1990)¹. An estimated one in every 750 children is born deaf or hearing impaired according to studies in the country. Even a mild hearing loss can significantly impair the normal development of speech and language. Thus early identification of infants with hearing impairment becomes necessary so that initiation of rehabilitation can be done at a time when the brain is sensitive to the development of speech and language preferably by the age of six months.

METHODS

Brainstem Evoked Responses Audiometry (BERA) was performed for 140 neonates (70 high risk and 70 normal neonates) during the first 28 days of life of the newborn and repeated at one to six month's duration for those

with abnormal responses. The study was performed over a period of two years from Sept 2000 to Aug. 2002.

The Auditory Brainstem Responses (ABR) of the high-risk group were compared with the normative group of 70 normal neonates (without any of the high risks) using the same protocol.

High risk factors considered were ; Family History of Deafness,, Prematurity, Perinatal Infections, Asphyxia, IUGR, Meningitis, Hyperbilirubinemia, Neonatal Sepsis, Fetal Malformations and Ototoxicity.

Far field scalp averaging technique was employed using an Octavus neuro-otological computer and a main unit BERA module with integrated pre amplifier for amplifying and averaging the ABR responses. Skin and electrode

Table -I : Results of Follow-up

Sl. No.	High Risk Factors Observed	No.of case	No. of Ears	Number of Affected		
				With Raised Threshold	Reported for followup	Still showed raised Threshold
1.	Prematurity	29	58	29	10	8
2.	Hyperbilirubinemia	18	36	18	10	6
3.	Malformation	7	14	10	9	8
4.	Asphyxia/Low APGAR	5	10	5	1	0
5.	Family H/O SNHL	3	6	5	5	4
6.	IUGR	3	6	2	2	2
7.	Neonatal Sepsis	3	6	4	0	0
8.	Infections	2	4	3	0	0

impedance was kept below 5 Kohm. Octavus D 748 earphones were used. 1024-2000 clicks of 100-microsecond duration having 2-4 kHz frequencies with intensities of 30-80 dB were used for each trial. Stimulus rate was 24/sec.

The intensity was gradually reduced to determine threshold of response i.e. minimum intensity to elicit wave V. Filter setting of 300-3000 Hz with click spectrum encompassing 1-4 KHz frequency range were taken. The testing was done with parental consent and a parent in attendance, while the neonate slept in an air-conditioned, quiet and acoustically shielded electro-neurodiagnostic laboratory of the department of Otorhinolaryngology.

ABR parameters analyzed were, the presence or absence of identifiable and reliable Waves I, III, V morphology and interpeak latencies, acquisition of absolute threshold levels in dBnHL for Wave V. BERA recordings were classified as abnormal when no Wave V was detectable from 30 to 80 dB intensities (i.e. increased auditory threshold) or I-V interpeak interval greater than the mean $\pm$ 2 SD for age matched, healthy control subjects as established in our laboratory by the above described technique and instrumentation. Elevated BERA threshold, more than 30 dB, was considered abnormal suggesting hearing loss.

Statistical Analysis

Summary data are represented as means with standard deviation or as means with ranges. Statistical analysis consisted of the use of chi-square analysis with automatic corrections for small sample size where appropriate and

the student 't' test for continuous variables that demonstrated a normal distribution. Single and multiple BERA variables as predictors of peripheral hearing status were evaluated by calculating sensitivity and specificity by using standard ABCD formulae.

RESULTS

140 neonates were tested. 70 were normal neonates (40 males, 30 females) and 70 were high-risk (44 males, 26 females). The various high risks included in the study were Family history of deafness (3), Prematurity (29), IUGR (3), Asphyxia (5), Perinatal infections (2), Hyperbilirubinemia (18), Neonatal sepsis (3) and Fetal malformations (7). Of the 70 high-risk cases, 44 neonates i.e., 64% of high risk (70) and 32% of all neonates (140) studied showed abnormal results (i.e., increased Wave V threshold) on neonatal Brainstem Auditory Evoked Potential (BAEP) recordings obtained before hospital discharge.

There was no difference between the control and test group with regards to clinical variables analyzed, specifically fever, excessive crying, refusal to feed, history of fever, exchange transfusion, history of family deafness, perinatal infections, and APGAR score. Additionally there were no differences between the two groups in gender and socio-economic status. The difference between the control and high-risk group was significant for Icterus, Respiratory distress, Asphyxia, IUGR, Neonatal sepsis, Fetal malformations, Phototherapy, Birth weight, Head circumference, Prematurity, and Hyperbilirubinemia. Table I compares the frequencies of hearing loss (demonstrated on follow-up audiometric assessment) for high-risk neonates whose predischarge BAEP showed an

Table -II : Distribution of Auditory Brainstem Evoked Response Threshold in High-risk Neonates.

Ser No.	High Risk Factors Observed	No.of case	No. of Ears	Auditory Threshold in dB (in number of ears) at BERA Recordings						
				30 dB	31-40 dB	41-50 dB	51-60 dB	61-71 dB	71-80 dB	No response
1.	Prematurity	29	58	29	10	6	8	4	0	1
2.	Hyperbilirubinemia	18	36	18	6	6	1	1	1	3
3.	Malformation	7	14	4	3	2	1	1	0	3
4.	Asphyxia/Low APGAR	5	10	5	3	1	1	0	0	0
5.	Family H/O SNHL	3	6	1	1	0	0	0	2	2
6.	IUGR	3	6	4	0	0	0	0	1	1
7.	Neonatal Sepsis	3	6	2	0	1	1	2	0	0
8.	Infections	2	4	1	0	3	0	0	0	0

Table -III : Normal Values at 80 dB

Wave	Latency	SD
Wave I	1.98 ms	0.11
Wave III	4.75 ms	0.28
Wave V	6.91 ms	0.33
Wave I-III	2.76 ms	0.23
Interpeak III-V	2.16 ms	0.26
Interpeak I-V	4.92 ms	0.32

increased Wave V threshold that suggests hearing impairment.

Auditory evoked responses using BERA were studied in 18 neonates with serum bilirubin $\geq 15\text{mg\%}$. Of the 18 neonates i.e., 36 ears tested, 18 ears showed thresholds of 40 dB and above. One neonate with 60 dB threshold in one ear and 40 dB in the second ear who was lost to follow up showed 40-50 dB threshold in the affected ear. Two neonates with serum bilirubin count $>20\text{ mg \%}$ showed no waveforms. Auditory threshold was observed to be raised in 8 neonates. Wave I was not recorded in 3 neonates. Absolute latencies were prolonged in 11, Inter peak latencies were also observed to be raised suggesting prolonged central conduction time. Exchange transfusion resulted in marked improvements in ABR findings.

Three neonates, (two females and one male) born to three sets of normal parents, had a family history of deafness. Two had no waveforms and one showed some response on follow-up.

Of the 5 neonates with Asphyxia i.e., 10 ears, 5 ears had raised thresholds, four were lost at follow up.

Prematurity was observed in 29 neonates (i.e., 58 ears, 19 ears failed initial testing at 40 dB). In 7 neonates i.e., 14 ears, with gestational age of 30 weeks, 13 ears had thresholds above 30 dB with increased absolute latencies. At 2 months follow up, 3 neonates had thresholds above 40 dB but all passed at 70 dB. They also had associated low birth weight. One neonate, at 26 weeks post-conception age, passed both ears at 50 dB. In 3 neonates, at 32 week of gestation, all showed increased latencies. Four ears had threshold of $> 40\text{ dB}$. Two were lost to follow-up. In 18 neonates, with gestational age of 34 weeks, 5 had thresholds $> 40\text{ dB}$. All passed at 70 dB. Three at follow-up had normal thresholds.

Perinatal infection (Rubella) was observed in the case of two neonates (4 ears). Three ears showed raised thresholds at 40 dB.

Out of the three neonates having IUGR, two had normal thresholds and one had no waveforms. This case had associated history of maternal rubella and prematurity.

There were 7 cases of congenital malformations. Meningomyelocele with hydrocephalus, post-shunting, was observed in two cases. They showed threshold of 30-40 dB. One case of Waardenberg syndrome (heterochromia irides, without dystopia canthorum) showed no waveforms bilaterally which was confirmed

at follow-up. One neonate with hydrocephalus with craniocynostosis showed thresholds at 70 dB bilaterally which was confirmed at six months follow up and was also diagnosed as a case of Crousans. One neonate with tracheo-oesophageal fistula showed normal threshold bilaterally. Of the two cases of Downs syndrome (4 ears), two ears passed at 50 dB and one showed no response.

In the 3 cases of septicemia, one ear passed at 40-50 dB and one showed 60 dB. Two neonates had associated history of prematurity. All three were lost to follow up.

Univariate analysis shows Prematurity, Hyperbilirubinemia, Neonatal sepsis, Fetal malformations and Asphyxia to be statistically significant with the occurrence of hearing loss. There were no cases of Meningitis, Intracranial hemorrhage and Ototoxicity and as such no inference was given for these factors. Thus five significant factors as above were recognized.

Results in the study indicate that BERA is a good predictor of normal hearing as seen in the cases that reported for follow up. We did not find any neonate with previously normal BERA to have reported with reduced hearing in the follow up. BERA had reported 44 neonates as having hearing loss. 28 ears (out of 67 ears) had the loss confirmed at repeat BERA follow up. Six neonates with unilateral hearing loss on follow up showed normal threshold (false positive). They were mostly from the premature group, where at least one year follow up is required. Thus calculating sensitivity $A/(A+c) \times 100 = 100\%$. Table II shows the Distribution of Auditory Brainstem Evoked Response threshold in High-risk neonates.

DISCUSSION

BERA is a very valuable audiological investigation for neonates and the otherwise difficult to test child. It is best performed in a formal audiological setting or in a room removed from high levels of acoustic and electrical interference. Most neonates, with parents in attendance, will settle and sleep sufficiently to allow testing without sedation. A detailed history including the presence of risk factors was taken into account and then the child was subjected to Evoked Response Audiometry. Comparable number of males and females were included in the control group for the establishment of normative data as shown in Table III.

There is no universal standardization for variables (Stockard 1979)² such as filter settings, stimulus characteristics, electrode montage, test locations etc. Of the 7 component waves I, III and V were most consistently found in neonates. Rarity of other waves may be due to incomplete myelination and maturation of brainstem. Absolute latency of Wave I was found to be 1.9 ms and Wave V 6.9 ms at 80 dB comparable to Mandal and Deorari^{3 4}. Interpeak latencies I-III, III-V and I-V were also calculated and found to be 2.73 ms, 2.23ms and 4.92 respectively at 60 dB comparable to Deka 1986⁵.

Of the total number of 140 neonates (control group and high risk group), 44 showed threshold greater than 30 dB i.e., 32% or 64% of the high-risk group. Eden and Ford⁶ in their study of 72 neonates had reported an abnormality in 84% of the neonates tested.

In the cases of Hyperbilirubinemia, auditory threshold was observed to be raised in 8 neonates. Wave I was not recorded in 3 neonates. Hyperbilirubinemia probably affects the auditory nerve rather than the cochlear hair cells. Prolongation of absolute latencies was observed in 11 neonates. Interpeak latencies were also observed raised, suggesting prolonged conduction time. Exchange transfusion resulted in marked improvement in ABR responses. BERA abnormalities though transient in some patients were significantly related to the level of serum bilirubin and duration of jaundice (Agarwal 1998)⁷. With exchange transfusion, bilirubin is removed from the brainstem nuclei resulting in improvement in BERA responses, Nwaelsi 1984⁸. Serum bilirubin >20 mg% increases the risk of neurological abnormalities, as does longer duration of jaundice, Yilmaz, 2001⁹.

In the family history cases, deafness was possibly transmitted as an autosomal recessive trait. In the family of one neonate, one female sibling was normal and one male sibling was deaf. Probably in this case X linked transmission lead to one female carrier, deafness in one male child and one male neonate.

Neonatal tissue hypoxia, in cases of asphyxia, can interfere with cochlear oxygenation and function as studied by Haupt 1993¹⁰. Konkle¹¹, reports late onset of hypoxia related hearing loss and hence the need for at least one year follow up even if initial BERA was normal.

Generally BERA latencies decrease and amplitude increases with advancing age. BERA can be recorded in infants as young as 28-30 weeks gestation (Starr, Schulman)^{12,13}. Between 36-40 weeks latencies of Wave I decreases by 0.1 ms and Wave V by 0.4 ms. At term, latency of Waves I-V is 5 ms.

There is a special vulnerability of eye, ear, heart of the developing fetus to maternal Rubella, greatest during first trimester. Infection in 2nd and 3rd trimester can cause deafness. Microscopy of the cochlea is known to show partial collapse of the Reissner's membrane and small granulomas between stria vascularis and Reissner's membrane. These granulomatous lesions described by many authors are known to have appeared when organ of Corti has reached morphological maturity, interpreted as consistent with the degeneration of the preformed neuroepithelial structures reflecting continued viral infection, as suggested by other stigmata of the Rubella syndrome, Friedman 1966¹⁴. However, the histopathology remains fragmentary.

Waardenberg syndrome is an autosomal dominant condition with deafness in both ears in 50% of those with type II i.e., without dystopia canthorum. In hydrocephalous BERA latencies following shunt insertion show improvement (Mc Pherson 1985)¹⁵. Patients with Down's syndrome are known to have deafness.

Sepsis is a known risk factor for hearing loss as described by Jacobson 1985¹⁶. It is attributed to dysfunction secondary to fibrous reaction with adhesions comprising the eighth nerve. Some suggest vasculitis of inner ear vessels. Many of the central abnormalities show reversibility.

BERA is thus a useful, non-invasive tool to test the difficult to test neonates. For neonates with high risk factors testing of the auditory function should be made mandatory prior to discharge and those with abnormalities kept on regular follow up so that remedial methods are applied at the earliest.

ACKNOWLEDGEMENT

Indian Council of Medical Research (ICMR), New Delhi, Supported this work through a grant from Sep 2000 to Aug, 2002.

REFERENCES

1. Anand N. K., Gupta A. K., Hans Raj. (1990) : Bera –A diagnostic Tool in Neonatology. Indian Pediatrics Volume 27: 1039-44.
2. Stockard J. E.; Stockard J. J., Westmoreland B., Corfits J. L. (1979) : Brainstem Auditory Evoked Responses : 'Normal Variation as a Function of Stimulus and Subject Characteristics : Arch Neurol : 36-823-831.
3. Mandal A. K., Mehra Y. N., Narang A., Raghunath M., Walia B. N. (1989) : Brainstem Evoked Response Audiometry in Normal Neonates : Indian Pediatric : 26:566-570.
4. Deorari A. K. Garg R., Bist M. S. (1989) : Auditory Brainstem Evoked Response in Normal Neonates and Infants : Indian Pediatric : 26: 981-986.
5. Deka R. C., Deka D., Kacker S. K. (1986) : Maturation of Cochlea, Auditory Nerve and Brainstem as observed by ABEP in Human Infants and Children : Indian Journal of Otolaryngology : Vol 38 : No. 2.
6. Eden D., Ford R. P., Hunter M. F., Malpas T. J., Darlow B., Gourley J. (2000) : Audiological Screening of Neonatal Intensive Care Unit Graduates At High Risk of Sensorineural Hearing Loss : NZ Med J. : 113(1110) : 182-3
7. Agarwal V. K., Shukla R., Mishra P. K., Kapoor R. K., Malik G. K. (1998) : Brainstem Auditory Evoked Response in Newborns with Hyperbilirubinemia : Indian Pediatrics ; 35 : 513-518.
8. Nwaelsi C. C., Aerde J. V., Boyden M., Perlman M., Chukwuma G. (1984) : Changes in Auditory Brainstem Responses in Hyperbilirubinemic Infants before and after Exchange Transfusions : Pediatrics : 74 : 800-803.
9. Yilmaz Y, Say, Yildiz (2001) : Indian Pediatrics; 38 : 165-168.
10. Haupt H., Seneib F., Ludwig C. (1993) : Changes in Cochlear Oxygenation, Microcirculation and Auditory Functions during prolonged General Hypoxia : Ear Arch Otol : 250 : 396-400.
11. Konkle D. F., Knightly C. A. (1993) : Delayed onset Hearing Loss in Respiratory distress Syndrome Case Reports. J. Am Acad Audiol; 4 : 351-4.
12. Starr A., Amlie R. R., Martin W. H., Sanders S. (1977) : Development of Auditory Functions in New born Infants Revealed by Auditory Brainstem Potential, Pediatrics : 60-831-839.
13. Schulman –Galambos C., Galambos R (1975) : BERA in Premature Infants ; J. speech. Hear Res : 18 : 456-465.
14. Friedman and Wright, (1966) : Histopathological Changes in the Fetal and Infantile Inner Ear by Maternal Rubella. British Medical Journal, 2 : 20-23.
15. Mc Pherson D. L., Amlie R., Foltz E. (1985) : ABR in Infantile Hydrocephalus, Childs Nerve Sys, 1 : 70-76.
16. Jacobson J. T. (1985) : Normative Aspects of the Pediatric Auditory Brainstem Response : J Otolaryngol Suppl, 14 : 7-11.