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Introduction

Hearing plays a basic role in language, speech and intellectual development. The importance of early detection and rehabilitation of infants with hearing impairment cannot be overstated. Unfortunately, the average time between birth and the detection of congenital sensorineural (SN) hearing loss is 2.5 years. The American Joint Committee on Infant Hearing recommended that audiological rehabilitation should begin within the first 6 months of life (1). The only way of detecting congenital deafness at this age is by neonatal screening. Measurement of the auditory brain stem response (ABR) is considered the most sensitive method of assessing the auditory activity of neonates.

In this report, we summarize our experience of the evaluation of high risk neonates for hearing impairment by ABR during a 6-year period.

Materials and Methods

The patients included in this study were 154 newborns admitted to the neonatal intensive care unit of

Auditory Brain Stem Response Screening for Hearing Loss in High Risk Neonates

Abstract: Hearing screening of neonates is the key to preventing the most severe consequences of congenital hearing loss. In this paper, the auditory brain stem responses (ABR) of neonates at high risk for hearing impairment at Marmara University Hospital Neonatal Intensive Care Unit are reported. One hundred fifty-four high-risk neonates underwent screening by ABR carried out according to the US Joint Committee on Hearing 1994 Position Statement. One hundred and fourteen of them passed the first ABR test while 40 patients failed. The number of patients with sensorineural and conductive hearing deficit was 5 and 35, respectively. Fifteen of the infants who failed had bilateral hearing loss. During follow-up, 6 of these 40 infants (15%) failed subsequent tests. Two of these babies had bilateral involvement, and the rest had unilateral involvement. The infant with bilateral sensorineural hearing loss had a cochlear implantation. Seventeen of the 40 infants passed the second ABR (42%). Three patients did not survive and 11 (27%) were lost from the follow-up. Three patients who failed the first ABR were scheduled to have the second ABR soon afterwards. It is concluded that early diagnosis of hearing loss in high risk neonates is important for the implementation of hearing aids early in infancy so as to prevent acoustic deprivation.

Key Words: Auditory brain stem response, newborn, hearing loss

Marmara University Hospital between 1991 and 1997. The patients were identified as being at high risk for hearing impairment according to the US Joint Committee on Infant Hearing 1994 Position Statement (2).

The subjects were screened close to the time of discharge from the hospital. The infants were tested either during natural sleep or after sedation with chloral hydrate in a sound-treated room. An otoscopic examination was performed before the test. All infants who failed an initial test were referred to an audiologist. ABRs were recorded using Amplaid MK 15 equipment. Electrodes were placed on the vertex (Cz, positive electrode), both mastoids (Ai and Ac, negative electrodes) and on the forehead (ground). The responses were filtered with a bandpass of 100 to 2500 Hz. The analysis time was 15 ms. The stimuli consisted of 100-ms, alternating clicks presented monaurally at rates of 11/s and 21/s. Two intensity levels were used: 60 dB nHL and 30 dB nHL, summing to 2000. In the case of no response at 60dB nHL, the stimuli were increased to maximum intensity levels. Each ear was tested separately. All the responses were replicated.

The major criterion for a normal response was a clearly reproducible Wave V at 30 dB nHL at each ear (3). For the degree of hearing loss, Goodman's categorization chart was used (4). Infants who failed the first test were retested after a month, when bone conduction ABR was performed as well. A great effort was made to retest all high-risk infants at 3 to 4 months of age. Sedation was performed at that time with chloral hydrate if necessary. Subsequent follow-up evaluations and provision of further services e.g., hearing aid evaluations, medical treatment and enrollment in a speech therapy program were recommended when appropriate.

Results

Patient characteristics: One hundred fifty-four neonates at high risk for hearing impairment underwent screening by ABR according to the United States Joint Committee on Infant Hearing Position Statement. The patient population consisted of 82 males and 72 females. The number of patients born before 32 weeks', at 33-37 weeks' and after 38 weeks' gestation were 41, 46 and 67, respectively. The birth weight was less than 1500 gram in 32 patients. Forty-six patients weighed 1500-2500 grams whereas 76 weighed more than 2500 grams. The distribution of neonates according to risk factor is shown in Table 1. The highest bilirubin level determined in babies with hyperbilirubinemia was 40 mg/dl while the duration of exposure to high bilirubin levels had a median level of five days (range:1-15). The most commonly used autotoxic antimicrobial agents were tobramycin and amikacin. The duration of treatment ranged from seven to twenty-eight days, with a median

of 10 days. Unfortunately, we were not able to measure the serum levels of these agents.

ABR results: The flow diagram of the neonatal screening program and outcome is shown in the Figure. Of the 154 infants tested, 114 (74%) patients passed the first ABR test while 40 patients failed. The number of patients with sensorineural and conductive hearing deficits was 5 and 35, respectively. Three out of 5 babies with SN hearing impairment died (1 of them had meningitis, the others had encephalocele and kernicterus) while 1 was lost during follow-up. The last infant with sensorineural hearing loss had behind-the-ear (BTE) hearing aids but there was no progress in her speech or comprehension. At the age of 4 she had a cochlear implantation. During the follow-up of the 40 infants who failed the initial test, 6 (15%) failed subsequent tests. The results for these infants are shown in Table 2. Five

Table 1. Risk factors for hearing impairment in high risk infants.

Risk factors	Number of Infants	%
Birth weight <1500 gram	32	20
Gestational age <32 weeks	41	27
Low Apgar score (5'< 3)	17	11
Hyperbilirubinemia	36	23
Craniofacial anomalies	20	13
Autotoxic drug use	60	39
History of meningitis	11	7
Mechanical ventilation>5 days	10	6
Family history of deafness	2	1
Congenital infection	-	-



Flow diagram of the neonatal screening program and outcome

Patient no.	Risk group	Age at diagnosis	Severity (Left/Right)	Туре	Outcome
1	Fraser syndrome	5 weeks	R-L/severe	Conductive	Died
2	$^{\epsilon}$ Craniofacial anomaly	3 days	R/severe	Conductive	*
3	$^{\epsilon}$ Craniofacial anomaly	8 weeks	R/severe	Conductive	*
4	Mondini syndrome	3 weeks	R-L/severe	β_{SN}	Cochlear implantation
5	⁹ Craniofacial anomaly	4 weeks	L/severe	Conductive	*
6	⁹ Craniofacial anomaly	4 weeks	L/mild	Conductive	*
7	Ventriculitis	12 weeks	R-L/severe	SN	Died
8	Ventriculitis	5 weeks	R-L/severe	SN	Lost
9	Kernicterus	2 weeks	R-L/severe	SN	Died
10	Encephalocele	5 weeks	R/severe	SN	Died
External ear canal atresia Skin tag *Normal unilateral bearing		ßSensorineural			

Table 2. Results for* all hearing-impaired infants.

 ϵ External ear canal atresia \rightarrow Skin tag *Normal unilateral hearing

* Including the infants who failed at the initial test and died afterwards.

of these babies had bilateral involvement, and the rest had unilateral involvement. The mean age at diagnosis of the hearing impairment was 5.1 weeks. Seventeen of the 40 infants passed the second ABR (43%) and 11 (27%) were lost from the follow-up. The overall follow-up rate for those who did not pass the initial ABR test was about 73%.

Discussion

The incidence of congenital bilateral hearing loss is 1 per 1000 live births (5,6). Infants who require admission to neonatal intensive care are reported to be at 20 times greater risk for auditory impairment (7,8). In our study of 154 high risk neonates, 6 infants with hearing impairment were identified, which represents 3.9% of the total population tested with ABR. Of these 6 infants, 1 had severe SN involvement. If one considers that only 73% of the infants who failed the initial ABR test were able to return for follow-up evaluation, 5.2% would be a more accurate incidence of hearing loss in our high-risk population. Our predicted incidence of severe SN hearing impairment would be 1 %, which is lower than the 2-4 % range reported by other investigators (6,7). One reason for this difference might be the fact that 3 of our patients who had severe SN hearing deficit at initial ABR testing did not survive and 1 was lost during follow-up. If it had been possible to follow them up, the incidence of SN hearing deficit would have been 4.2 %.

The Joint Committee on Infant Hearing (1982) recommended the screening of infants at risk on the basis of several risk criteria (1). However, selective screening based on high-risk criteria fails to identify approximately 50% of these children born with significant hearing impairment. The National institute of Health consensus statement (1993) and The Joint Committee on Infant Hearing (1994) recommended that universal hearing screening be performed before hospital discharge for all newborns as well as high risk neonates (2,9). After these statements we revised our policy by screening healthy newborns as well as high risk neonates. The click-evoked otoacoustic emission (EOAE) recording procedure is simple and quick and can be used as a screening test. However, one shortcoming of the EOAE is a high false positive rate in newborns, which is attributed to ear canal debris (10).

Different methods have been evaluated in the search for a reliable and effective technique for determining auditory function in newborns. Among these have been behavioral audiometry and electrophysiologic testing. Behavioral tests identify only infants who are profoundly deaf and miss mild or moderate hearing loss (11,12,13).

Auditory Brain Stem Response Screening for Hearing Loss in High Risk Neonates

Auditory evoked potentials are electroencephalographic responses to auditory stimuli. ABR is an important clinical tool in the identification and quantification of hearing impairment. It is objective, noninvasive and unaffected by sleep or drugs. In the automated version of ABR (AABR), a machine-based decision on the presence or absence of waveforms is made, resulting in a pass/refer decision. AABR screening does not require interpretation by an audiologist (14).

Transient conductive hearing loss was the most common cause of initial ABR screening failure in those of our patients who were normal on retesting. It has been reported that estimates of the incidence of middle ear effusion in neonatal intensive care, especially if intubated, are as high as 30% (15). Other possible causes of transient ABR failures could be transient neurologic anomalies.

One of the primary goals of a neonatal screening program is to identify and initiate follow-up treatment of hearing-impaired infants within the first 6 months of life. It has been shown that sensory deprivation in the early years of life can result in permanent neurologic changes. Hearing-impaired children who are subject to intervention at an early age have significantly better communicative skills than children who receive similar intervention at a later age. In our study of 4 infants with severe SN hearing impairment, 3 did not survive long enough for rehabilitation, and 1 had a cochlear implantation.

In conclusion, it is important to screen all high risk infants with ABR for early diagnosis and the implementation of hearing aid early in infancy in order to prevent acoustic deprivation.

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