Frequency of Profound Congenital Hearing Loss in Healthy Newborn Infants in Fars Province

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Abstract

Introduction
Deafness is one of the most important sensory disturbances at birth. Hearing loss can affect the development of speaking and learning during life. Early diagnosis and intervention improve language outcome. The current study aimed to determine the frequency of profound congenital hearing loss in healthy newborn infants in Fars province.

Methods
In a multicenter prospective study, from August 2010 to August 2011, 12573 newborns were screened for hearing loss prior to discharge from the wellborn nursery at nine teaching and private hospitals in Fars province. A three-stage hearing screening protocol using transient evoked otoacoustic emissions (TEOAE) screening with referral for diagnostic second TEOAE and auditory brainstem evoked response (ABER) assessment was employed. All neonates with abnormal initial TEOAE screening, followed with phone call for language outcome after one year. Newborns with hospital admission or drug administration excluded from the study.

Results
The overall pass rate in the initial screening was 91.8%, thus 1019(8.1/1000) neonates referred for diagnostic audiological assessments. Out of 1019 infants scheduled for follow-up study, only 619 neonates returned. Using follow-up letters and phone calls, it was managed to improve the response rate for language outcome after one year. Two infants were identified with profound bilateral congenital hearing impairment. These infants were immediately referred for cochlear implantation.

Conclusion
Findings of the current study showed that the frequency of profound congenital hearing loss is 0.159/1000 normal newborn infants in Fars province. The initial TEOAE has high false positive that may be due to occlusion of external ear canal by vernix in the early postnatal period.

Key words
Congenital hearing loss, Newborns, Prevalence, Neonatal screening

Introduction

Significant hearing loss is one of the most important disorders at birth. Hearing is a key sense to develop communication and communication is the basis of learning and education. Hearing loss leads to delayed language development, psychosocial disturbance, and poor academic achievement. Early diagnosis and intervention with speech therapy, amplification (e.g., hearing aid) and interventions such as cochlear implantation improve language outcomes (1, 2). Universal newborn hearing screening identifies congenital hearing loss at an earlier age, allowing for earlier intervention (3). Two electro physiologic techniques meet the criteria of the American Academy of Pediatrics to screen hearing loss (4).

1.Otoacoustic emission (OAE) measures the presence or absence of sound wave generated

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by the cochlear outer hair cells of the inner ear in response to sound stimuli. The transient OAE (TOAE) utilizes a click stimulus that results in the emission of several frequencies at the same time.
2.Auditory brainstem response (ABR), measures the summation of action potentials from the cochlear nerve to midbrain in response to a click stimulus.
The prevalence of hearing loss is different among studies. Thompson et al (5) reported that the frequency of moderate, severe, and profound bilateral permanent hearing loss is estimated at 1 in 900 to 2500 newborns. The frequency of congenital hearing loss has been 4.97/1000, 1.3/1000, (1-4)/1000, 1/1000 in Turkey (6), Mashhad (7) and Tehran (8, 9) respectively.

Objectives

The purpose of this current study was aimed to determine the frequency of profound congenital hearing loss in healthy newborn infants in Fars province.

Methods

It was a multicenter prospective study in healthy newborn infants. All neonates born at nine teaching and private hospitals of Fars province (Hafez, Hazrat Zeinab, Dena, Ordibehesht, Shafa, Moslemin, Alavi, and Pars hospitals in Shiraz, and Motahari hospital in Marvdasht) from August 2010 through August 2011 were enrolled in this study. Newborns with hospital admission or drug administration were excluded from the study. Neonates were screened for hearing loss prior to discharge from the wellborn nursery. A three-stage hearing screening protocol was employed. Neonates with abnormal initial transient evoked otoacoustic emission (TEOAE) were referred for diagnostic second TEOAE and auditory brain stem evoked response (ABER) assessment. TEOAE technique : The apparatus for OAE screening consists of a miniature microphone placed into the infant's outer ear canal. The microphone produces a stimulus (clicks or tones) and detects sound waves as they arise from the cochlea. The device also measures the signal-to-noise ratio to ensure accuracy. ABER technique: The screening ABR utilizes click stimuli presented at 35 dB. Three surface electrodes placed on the forehead, nape, and mastoid detect waveform recordings generated by the auditory brainstem response to the click stimuli. The morphology and latency of the waveforms are compared to those of normal and a pass or fail reading is generated. Delayed or absent waves suggest a neurologic or cochlear deficit. Screening ABR requires 4 to 15 minutes for testing. All neonates with abnormal initial OAE screening, were followed with phone call for language outcome after one year. They must say a few words bedsides “mama” “dada” at the age of one year.
Analysis of data was done by descriptive statistic analysis. SPSS (version 16, USA) was employed for data analysis. The protocol of the study was approved by the local ethics committee of the university. The informed consent was obtained verbally from the parents.

Results

According to inclusion and exclusion criteria, 12573 healthy neonates were screened prior to discharge from hospital.

Table I. Number and sex of neonates who screened and failed initial TEOAE in each hospital

Hospital	Number of neonates who screened	Number of neonate with abnormal first TEOAE
Hazrat Zeinab	2608 (M) 2594 (F)	20 (2%)
Hafez	1471 (M) 1374 (F)	479 (47 %)
Moslemin	425 (M) 426 (F)	16 (1.6 %)
Ordibehesht	108 (M) 110 (F)	32(3.1 %)
Shafa	21 (M) 15 (F)	14 (1.4 %)
Pars	395 (M) 328 (F)	96 (9.4 %)
Dena	755 (M) 723 (F)	212 (20.8 %)
Alavi	69 (M) /59 (F)	58 (5.7%)
Marvdasht	546 (M)/ 546 (F)	92(9 %)
Total	12573	1019(100 %)

Table I showed the number and sex of neonates who were screened and failed initial TEOAE in each hospital. The overall pass rate in the initial screening was 91.8%. 1019 (8.2/1000) neonates failed the initial TEOAE test in one or both ears who were referred for diagnostic second

TEOAE and ABER assessment. Table II showed the characteristics of neonate with abnormal first TEOAE. Out of 1019 infants scheduled for follow-up study, only 619 neonates returned. Using follow up letters and phone calls, it was managed to improve the response rate for language outcome after one year. Two infants were identified with profound bilateral congenital hearing impairment. Both of them were male, full term and had family history of congenital hearing loss. These infants were referred for cochlear implantation.

Table II. Characteristics of neonates with abnormal first TEOAE.

Variables	Number (percent)
Male Female	514 (50.4%) 505 (49.6%)
Term Preterm Unknown	868 (85.20%) 113 (11.08%) 38 (3.72%)
Positive family history of hearing loss Negative family history of hearing loss Unknown	39 (3.82%) 973 (95.48%) 7 (0.69%)
Facial congenital anomalies present Facial congenital anomaly absents	0 (0%) 1019 (100%)

Discussion

Significant hearing loss is one of the most common disorders at birth. Screening neonates for hearing loss leads to earlier detection and intervention. There are two approaches to identity hearing impairment: selective screening to test newborns that are at increased risk for hearing loss. Major risk factors include: family history of hereditary SNHL, neonatal intensive care unit admission, craniofacial anomaly, congenital infection, severe hyperbilirubinemia and syndrome associated with SNHL (5, 10). Approximately 10-30% of neonates have one or more of these risk factors (10). In universal screening, all newborns must be screened for hearing during birth hospitalization. With the widespread adaption of universal screening, the age of identification of hearing loss has decreased from a range of 24-30 months to 2-3 months of age (11).

The current study findings showed that the frequency of profound congenital hearing loss is 0.159/1000 normal newborn infants in Fars province. Frequency of congenital hearing loss was more in other studies (6-9). In the current study 400 neonates with initial abnormal TEOAE did not refer for audiological assessment. This problem was managed by evaluation of language outcome after one year. So the cases with mild, moderate or unilateral hearing loss were missed.

The limitation of the current study was absence of second TEOAT and ABER in 400 neonates but these neonates were followed with phone calls to detect the cases of profound bilateral congenital hearing impairment with delay in speech and language.

Findings of the current study also showed that only two neonates with abnormal initial TEOAE had abnormal second TEOAE and ABER. So, initial TEOAE had high false positive rate which was similar to the results of headley et al (12) and Stewart et al (13) . They reported that more infants appear to have hearing loss by TEOAE than by ABER during the first three days of life. These false positive results are due to occlusion of the external ear canal by vernix (12, 13). This can occur in 19-25% of neonates with OAE screening (14). Dolye et al (14) showed that clearing vernix can increase the pass rates.

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Conflict of interest:

None declared.

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