Objective: This article reports the results of newborn hearing screening which has been performed between March 2009-2010 in Cizre State Hospital. The results of the first step hearing screening were discussed together with the risk factors for hearing loss.

Materials and Methods: All 1840 babies that were born in Cizre State Hospital between March 2009 and March 2010 were enrolled in the study. They all underwent TEOAE (transient otoacoustic emissions) test as the first step of hearing screening.

Results: Of the 1840 babies who were screened, 1441 (78.3%) passed the first test and remaining 399 (21.7%) were called again for the second test. Hundred and fourteen (6.2%) of these babies were not brought for control. Among the latter patients 29 had various risk factors for hearing loss. 249 (13.5%) of the 285 (15.5%) babies who were re-evaluated, passed the test and remaining 36 (1.96%) were referred to the second step institutions.

Conclusion: In rural areas more intensive protocols should be employed. Those babies delivered during weekends and nights should be screened before discharge and a member of the hearing screening team should be present during this time period as well. AABR (Automated Auditory Brainstem Response) and TEAOE should be done together in these high risk areas. More efficient way of follow-up of these babies is also necessary.

Submitted: 5 April 2011 Revised: 15 July 2011 Accepted: 11 August 2011

Introduction
Speech and language development of babies improves quickly in the first years, especially in the first months of life. A six-month-old baby shows more interest to speaking voice than the other sound sources. The baby can make simple sentences when he/she is 18-month-old [12]. Having a normal hearing in early infancy is crucial for social, emotional and mental development along with speech and language acquisition [13]. Thus early recognition of hearing loss, that is among the common congenital problems, is quite important [4-5].

Ratio of congenital hearing loss among healthy newborns varies between 0.1-0.6% and detection in early stages is of great importance [4-10]. In Turkey, data obtained from NNHSP (National newborn hearing screening program in Turkey) showed that 17 out of every 10 000 have hearing loss in healthy newborns. [11] Newborn hearing screening should be performed for early intervention of hearing loss. American Academy of Pediatrics recommends screening all newborns in the first month following delivery [12]. Hearing loss that may vary from slight to severe, leads to impairment of communication skills along with sensory deprivation and learning difficulties [13]. In order to prevent these developmental issues the hearing loss should be confirmed within three months after birth and hearing aids should be fitted within six months [12,14].

Newborn hearing screening program aims to diagnose babies with severe and profound hearing loss early, definitely and in the least expensive way [13]. Some
studies recommend that all universal newborn hearing screening programs should consider revising their protocols to decrease expenses in terms of time and money \[16\]. However, a successful newborn hearing screening program is expected to diagnose hearing loss that could interfere with linguistic, emotional, social and cognitive development \[8, 17, 18\].

Methods that are used in hearing screenings are not used as definitive diagnostic tools although they are fast, cheap, practical and safe. Screening results reduce the population that should be assessed in more detail in terms of possible hearing loss. Babies who fail the screening test are directed towards further tests. If hearing loss is confirmed, the patient is enrolled in a rehabilitation program \[8,19\]. The goal of screening programs is to apply hearing screening to at least 95% of newborn population \[20\].

At present, otoacoustic emissions (EOAEs: Evoked Otoacoustic Emissions) and Auditory Brainstem Response (ABR) measurements are used together or alone in newborn hearing screening programs \[6, 7, 21\]. In Turkey, newborn hearing screening was first begun in 2000 by two audiology departments with collaboration of Turkish Prime Ministry. On December 2004, newborn hearing screening campaign was started by means of the protocol signed out by Prime Ministry, Head-Office of Disabled People and Ministry of Health \[22\]. Cizre State Hospital was involved in this screening procedure on the last days of January 2009. The present study gives one-year data of screening test in a region with high birth rate. It also provides data about the region's general social structure, demographic information, and puts forward the effect of regional resources on screening results.

Materials and Methods

A total of 1840 full-term babies who were born between March 2009 and March 2010 in Cizre State Hospital and referred to Department of ENT Newborn Hearing Screening Unit were included in the study. All newborns who came to the Department of Otorhinolaryngology Newborn Hearing Screening Unit underwent hearing screening by a full time working audiologist with master of science degree and an audiology technician who has been also involved in in-service training on newborn screening. Hearing screening was applied when the baby was sleeping or still in a quiet room separated for this purpose.

Transient Evoked Otoacoustic Emissions (TEOAE) method was employed using Bio-logic Systems Corp. brand AuDX-Pro Model device. Obtaining an automatic result of 'pass' was accepted as the criteria for the presence of hearing.

Screening results were reported to the families in written forms. Screening results were entered to web based Newborn Hearing Screening Program of the Ministry of Health and to Hearing Screening Surveillance Form.

All families received questions about the risk factors involving the baby and family history which may predispose to hearing loss. Risk factors for hearing loss are accepted as follows; family history of hearing loss, intrauterine infections, craniofacial anomalies, birth weight <1500 gram, critical hyperbilirubinemia, ototoxic medications, bacterial meningitis, postnatal asphyxia, mechanical ventilation >5 days, stigmata, or syndromes associated with hearing loss, prematurity (gestational age lower than 37 weeks), APGAR score of 0-4 in first minute or 0-6 in fifth minute \[23\].

Results

The newborn hearing screening program was analyzed according to the data gathered from the families of 1840 newborns: 194 (10.5%) of the mothers were 20 years old and younger, 961 (52.2%) were between 21-30 years, 604 (32.8%) were between 31-40 years and 81 (4.4%) were 40 years and older. Mean age of the mothers was 27.98±6.12 (min:14- max:45). Forty two (2.3%) of the fathers were 20 years old and younger, 767 (41.7%) were between 21-30 years, 735 (39.9%) were between 31-40 years and 296 (16.1%) were 40 years old and older. Mean age of the fathers was 31.44±6.99 (min:17-max:60).

When families were analyzed in terms of number of children: 133 (61.6%) families had 4 or less offsprings, 649 (35.3%) had 5-10 offsprings, 57 (3.1%) had 10 or more offsprings. Mean number of children was 4.01±2.67 (min:1-max:14).

When families were analyzed in terms of number of children: 133 (61.6%) families had 4 or less offsprings, 649 (35.3%) had 5-10 offsprings, 57 (3.1%) had 10 or more offsprings. Mean number of children was 4.01±2.67 (min:1-max:14).

When families were analyzed in terms of health coverage: 1394 (75.6 %) had green cards (no social security but taken care by the government directly), 289 (15.7 %) were members of social security fund, 60 (3.3%) were members of superannuation fund, 22 (1.2) % were members of occupational pension fund and 75 (4.1%) had no health coverages. While 76.5% of the
families were residents of Cizre, 23.5 % were living in neighbourhood.

When mothers and fathers were analyzed in terms of relationship: 611 (33.2%) had consanguity; 440 (23.9%) were first degree, 171 (9.3%) were second degree relatives. Number of family members with hearing loss was 54 (2.9%) and of them, 38 (2.1%) were first degree relatives, 16 (0.8%) were second degree relatives of the babies (Table 1).

Mean body weight of the babies was 3,170.91±413.62 gr. Babies who had less than 1500 gr weight at birth, were referred to university hospital during their routine pregnancy follow-up and they were not included in the present data.

Table 1. Findings regarding with risk factors.

<table>
<thead>
<tr>
<th>N</th>
<th>Passed</th>
<th>Not brought for second control</th>
<th>Referral</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>N:1690</td>
<td>N:114</td>
<td>N:36</td>
</tr>
<tr>
<td>Consanguity(611)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>First degree</td>
<td>440</td>
<td>398 (23.5%)</td>
<td>29 (25.4%)</td>
</tr>
<tr>
<td>Second degree</td>
<td>171</td>
<td>150 (8.9%)</td>
<td>19 (16.7%)</td>
</tr>
<tr>
<td>Family history of hearing loss(54)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>First degree</td>
<td>38</td>
<td>33 (2%)</td>
<td>5 (4.4%)</td>
</tr>
<tr>
<td>Second degree</td>
<td>16</td>
<td>15 (0.9%)</td>
<td>1 (2.8%)</td>
</tr>
<tr>
<td>Phototherapy due to hyperbilirubinemia</td>
<td>81</td>
<td>12 (0.7%)</td>
<td>3 (2.6%)</td>
</tr>
<tr>
<td>History of intensive care unit stay</td>
<td>28</td>
<td>21 (1.2%)</td>
<td>1 (0.9%)</td>
</tr>
<tr>
<td>Total</td>
<td>629 (37.2%)</td>
<td>57 (50 %)</td>
<td>26 (72.2%)</td>
</tr>
</tbody>
</table>

833 of the infants were girls (45.3%), 1007 were boys (54.7%) and 1340 (72.8%) were normal vaginally delivered and 500 (27.2%) were delivered with caesarean sections. 81 (4.4%) of newborns received phototherapy because of hyperbilirubinemia and of these babies the ones who failed only TEOAE were referred to second step institutions for ABR. 28 (1.5%) newborns had been hospitalized in intensive care unit (Table 2).

Out of 1840 1441 (78.3%) newborns passed the initial test. 114 (6.2%) babies out of 399 (21.7%) who were invited for re-evaluation were not brought for control, 249 (13.5%) out of 285 (15.5%) babies who were brought for control passed the test. 36 (1.96%) newborns who failed the second test were referred to a second step institution for ABR test (Table 3).

Table 2. Demographic data of newborns (n=1840)

<table>
<thead>
<tr>
<th>Gender</th>
<th>N</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td>Girl</td>
<td>833</td>
<td>45.3</td>
</tr>
<tr>
<td>Boy</td>
<td>1007</td>
<td>54.7</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Delivery type</th>
<th>N</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td>Normal vaginal</td>
<td>1340</td>
<td>72.8</td>
</tr>
<tr>
<td>Caesarean section</td>
<td>500</td>
<td>27.2</td>
</tr>
</tbody>
</table>

| Phototherapy due to hyperbilirubinemia | 81 | 4.4 |

| History of intensive care unit stay | 28 | 1.5 |
| Atresic ear | 2 | 0.1 |
| Down’ syndrome | 2 | 0.1 |
| Microcephaly | 1 | 0.05 |
| Osteogenesis imperfecta | 1 | 0.05 |

Table 3. Results of the babies included in the study

<table>
<thead>
<tr>
<th>Tested</th>
<th>Passed</th>
<th>Invited for control</th>
<th>Not brought for control</th>
<th>Referral</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. Test</td>
<td>1840 (100%)</td>
<td>1441 (78.3%)</td>
<td>399 (21.7%)</td>
<td>-</td>
</tr>
<tr>
<td>2. Test</td>
<td>285 (15.5%)</td>
<td>249 (13.5%)</td>
<td>-</td>
<td>114 (6.2%)</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>36 (1.96%)</td>
</tr>
</tbody>
</table>
The parents of the 36 babies who were referred, 13 (36.1%) were first degree relatives and 2 (5.6%) were second degree relatives. Additionally, they had a history of hearing loss in their family. Only one family (2.8%) parents had no consanguinity. 4 (11.1%) out of 36 babies who were referred had a history of hyperbilirubinemia (Table 1).

Twenty nine (25.4%) out of 114 cases who failed the initial test and were not brought for the second test were first degree relatives, 19 (16.7%) were second degree relatives and 5 (4.4%) had hearing loss within other family members. In 1 of these 5 cases, parents were second degree relatives (Table 1).

Nineteen (23.5%) out of 81 (4.4%) babies who received phototherapy due to hyperbilirubinemia failed the initial test. Of them, 12 (14.8%) passed the second test, 3 (3.7%) were not brought for the second test, 4 (5%) failed the second test, too and referred. 352 (19.13%) out of 399 babies who failed the first step screening, failed bilaterally and 47 (2.55%) failed unilaterally. 33 out of 36 babies who were referred for second step screening were referred as they had failed bilaterally and 3 were referred as they had failed unilaterally. Regarding the babies with craniofacial anomalies 2 babies had external ear canal atresia, 2 had Down’s syndrome, 1 had microcephaly and 1 had osteogenesis imperfecta (Table 2).

A total of 70 (3.80%) twins were evaluated in the course of screening. Twenty one (30%) out of 70 twin babies failed the first test. Of these babies, 11 (15.9%) passed the second test but 10 (14.1%) were not brought for the second test. Also within the 114 babies who were not brought for the second control, 58 had at least one of the risk factors. 26 from 36 babies who were referred for the second step of hearing evaluation had risk factors. Within the 1690 babies who passed from the hearing screening 629 of them had also one of the risk factors.

In the first step of hearing screening the TEOAE test was accepted as a “pass” when he/she got ‘pass’ result for both ears. Babies who could pass the test for only one ear or the ones who could not pass for any of the ears, were recalled fifteen days later and both ears were evaluated again. Babies who could pass the test for both ears on the second test were accepted as a “pass”. Otoscopic examinations of the babies who failed the test unilaterally or bilaterally were done by an ENT specialist. In case of presence of a problem involving external auditory canal and/or middle ear like debris or otitis media, treatment and recommendations were given. If a response could not be obtained after the problem had been resolved, the baby was referred to Cizre State Hospital that is the second step for ABR. If the babies reapplied to the hospital with negative ABR result, they were referred to third step institutions for further investigation.

The usual trend worldwide is performing hearing tests prior to baby’s discharge from the hospital [21]. However, in this hospital, not all babies who are delivered stayed long enough at the hospital to be screened before discharge. Babies who are delivered with caesarean sections are discharged in average two days. Those with normal vaginal delivery are discharged at the same day. Even babies, who are delivered at night, are sometimes discharged within 3-4 hours. Only newborn babies who are delivered during working hours were screened before discharge and the families of the babies who are delivered at the weekend are informed about bringing the babies to the hospital within 10 days following discharge and a written document is given to the first degree relatives of the babies.

Usually TEOAE, which is among the evoked otoacoustic emission forms and AABR are used for hearing screening. In different studies in literature, different protocols are seen that TEOAE was used alone [19,25], TEOAE test was used as two-step or TEOAE and AABR tests were used together, DPOAE (Distortion Product Otoacoustic Emissions) and AABR tests were used together also AABR test was used alone [19,26]. In the present study, TEOAE test was used alone that takes place in the first step of three-step screening protocol. Some studies indicate that protocols involving both TEOAE and AABR are more sensitive than using either modality. It is more effective to use the protocol which consists of

Discussion

The goal of hearing screening protocol is to diagnose hearing loss before the baby reaches 3 months of age and initiate treatment when the baby is six-months old [12,18]. Using the most appropriate, effective and the least expensive method for hearing screening protocol would provide early diagnosis [24].
TEOAE, and AABR in combination with tympanometric measurements.\cite{27}

In literature most of the studies mainly focused on the screening outcomes of either healthy newborns with no risk factors or newborns only with risk factors. This study provides hearing screening outcomes of all newborns both with or without risk factors\cite{21,24}.

In previous studies, hearing screening results are reported to be performed by audiologists, health technicians, nurses, students of nursing and audiology or volunteers\cite{28,29}. In this study, although the screenings were performed by an experienced audiologist and by an audiology technician using TEOAE only, certain pathologies such as auditory neuropathy spectrum disorder might be overlooked. Characteristics of the target population, birth rate, false positive rate of TEOAE, the cost of device and consumption, distance to diagnostic audiology centers must be taken into consideration in deciding the protocols.

In this study, 36 (1.96\%) babies were referred to a second step institution for diagnostic ABR test. However, whether those babies reached that institution or not, was not reported to the clinic along with the result. Socioeconomic and natural conditions of the region make newborn hearing screening difficult to reach its exact goal. Therefore, combined use of both TEOAE and AABR in similar rural areas may decrease the number of referrals.

Genc et al.,\cite{22}, analysed the babies who were referred to Hacettepe University Department of Ear-Nose-Throat Audiology Section from Ankara Zubeyde Hanum Maternity Hospital and Ankara Zekai Tahir Burak Maternity Hospital for further investigation. They reported that 39 out of 5832 (0.67\%) babies were referred from Ankara Zubeyde Hanum Maternity Hospital (September 2000-September 2001) and 108 out of 12 665 (0.85\%) babies were referred from Ankara Zekai Tahir Burak Maternity Hospital. In another study carried out by Yıran et al., they reported that they recalled 16 out of 8052 (0.19\%) babies for control three times. In our study, 36 out of 1840 (1.96\%) babies were referred to a second step institution which represents 2 to 3 times more referrals than a metropolitan city. This shows us that there may be more risk factors namely the consanguinity in rural areas.

In the studies conducted in our country, Genc et al.\cite{22} reported the ratio of hearing loss to be 0.20 \% as a result of hearing screening. Kayıran et al.\cite{30} reported ratios of unilateral and bilateral hearing loss as 0.06\% for each. They concluded that the reason for lower ratios compared to literature were socioeconomic status of the families being moderate and high, mothers’ gestation period being healthy and ratio of consanguineous marriages being nearly none. In the light of these ratios, they stated that ratio of hearing loss was consistent with literature\cite{18, 19, 26, 29}. In this study, only first step results are given. The reason for not being able to give ratio of hearing loss is that we could not obtain the results of the babies who were referred to second step and tertiary step institutions. In the present study the ratio of consanguineous marriages that is among the major causes of hearing loss was detected as 33.2\%, ratio of familial history of a hearing loss was detected as 2.9\%. While 76.5\% of the families of the tested babies were residents of Cizre, 23.5\% were living in neighbourhood and most of the babies who failed the test were not brought for control. We consider that the reasons of this high ratio of babies that were not brought for control are the follows: high ratio of consanguineous marriages depending on the family structure of the region, low socioeconomic level of most of the families, high number of gestations and irregular follow-ups during gestation.

Consanguineous marriage is a tradition which is commonly practiced among Asian, African, and Latin American communities. While ratio is about 1-2\% in United States, Canada, Northern and Western Europe, it was found as 23-54\% in Egypt, Jordan, Lebanon, Kuwait, India and Turkey\cite{31}. These communities, in addition to their custom of interrelated marriage, have large families and are a rapidly growing population. The siblings of consanguineous marriages have a significantly higher incidence of autosomal recessive diseases including hearing impairment. The rate of consanguinity was 45 per cent. The incidence of hereditary hearing impairment is very high in developing countries compared to developed countries\cite{32}. In our country, many factors like education level of the community, traditions, religious faiths, size of that particular region, geographic location, socioeconomic level are seen to affect the frequency of consanguineous marriages\cite{33,34}.
In this study region, girls are married when they are very young. Age of giving birth begins with 14 and 10.5% of the mothers are aged 20 and younger. Additionally, families are not willing to send their daughters to school, especially in rural areas. Families marry their children when they are young as their social, cultural and economic levels are low (79.7% of the families had green cards or no health coverage). So marriages at an early age increase the frequency of intermarriages. Researches about consanguineous marriages conducted in different regions support this data [32,34,35].

In the studies held in our country, ratio of consanguineous marriages varies between 11.5% and 46.9% [32-35]. The difference between these ratios can be explained by cultural and socioeconomic conditions. This ratio was found as 33.2% in the present study which was held in Southeastern Anatolia region. This is higher than the mean ratio for Turkey and it is similar to previous studies.

Reddy et al [36] reported that among various etiological factors described for deafness, consanguinity is an established high risk etiological factor. A prospective study was carried out in 1076 children in the age group of 0-14 years attending Government ENT hospital and schools for deaf in and around Hyderabad. The results showed that 41.73% (449) of the cases were the products of intermarriage and 58.27% (627) were born to non consanguineous parents. Further analysis revealed a high rate of consanguinity (44.53%) in children with non syndromic deafness. These percentages are more than our results.

Hundred and fourteen babies failed the first screening, and did not come for control. Twenty nine of these had consanguinity and positive history of familial hearing loss. Although many risk factors were identified, familial hearing loss, bacterial infections and craniofacial abnormalities are reported to be the most significant ones. Other perinatal complications did not significantly influence screening results indicating improved perinatal handling in a neonatal population at risk for hearing disorders [37].

Twenty eight babies who were followed in the intensive care unit and the 81 babies with hyperbilirubinemia had high risk of hearing loss and should have been screened with ABR. However, the current unit was newly opened and was being used for monitorization of hyperbilirubinemia only. There was no ABR facility at that time in that unit. After the hyperbilirubinemia treatment, they were referred to other centers for ABR screening.

When we analysed the ratio of factors that take place in etiology of hearing loss: 2 out of 1840 babies had Down’s syndrome. There were 2 atretic ears in our study (2/1840); one was unilateral (in the right ear) and the other was bilateral. The presence of atresia prevents normal speech and language development and special treatment is necessary. One baby had osteogenesis imperfecta that is a genetic disorder. In addition, one baby had microcephaly.

One of the most important indicators of the success of newborn hearing screening, is a well planned follow-up program. It is necessary to establish audiology diagnostic centers that further investigations could be done for babies who fail hearing screening. These centers should be easily reached and have an effective service [39]. In present study, mean number of children for families was 4, 75.6% of newborn babies had green cards, 4.1% had no health coverage, second and third step institutions were remote so reaching them was difficult. Those were among the reasons for insufficient success of hearing screening.

Although the protocol that we applied in our study is widespread worldwide, in recent years AABR test was seen to be used alone or together with TEOAE test. In the presence of factors that affect TEOAE test negatively, if a response could be obtained with AABR test, hearing screening is accepted as a “pass” and the baby was not requested for a control test. Based from the results obtained from the present study it is important to have an AABR at each first step hospital to be used together with TEOAE. In addition, in rural areas the team should be organized to screen the babies that were delivered during weekend and also night. The reason for this is that only half of the babies who were requested to come to be tested came for screening. In future, we plan to reach more newborns by taking the emission device in delivery room and screening the babies who are delivered at night and in the weekend. Another plan is to buy an AABR device for the rural area hospital in order to follow up the babies who are not brought for control.

Number of the hospitals that newborn hearing screening is performed is increasing also in Turkey.
similarly to many countries. Families are informed about suspicion of hearing loss early by means of screening protocol however difficulties for reaching related institutions for further investigation make the families disappear. More effective screening and follow up programs should be developed by taking into consideration the socio cultural and economic level of the region that hearing loss risk criteria are intensive.

References


