**INTRODUCTION**

Congenital hearing loss was found in 1–3/1000 newborns in the well-baby population and in 2–4/100 newborns in the intensive care unit population. Early identification and intervention of hearing loss is crucial for hearing-impaired newborns in terms of their speech and language development. If they are not identified early, hearing impairment may have negative impacts on their language, cognitive, social, emotional, and academic development [1-3].

Several reports have demonstrated that early identification and intervention before 6 months of age is correlated with better development of language in hearing-impaired children [4, 5]. National Institutes of Health concluded in 1993 that universal hearing screening should be practiced for every infant within the first 3 months of life [6]. In 1994, the Joint Committee on Infant Hearing (JCIH) released a position statement approving the goal of the universal detection of newborns with hearing loss and stated that all infants with hearing loss should be identified before 3 months of age and intervened by 6 months [7]. Many investigations have provided evidence that universal newborn hearing screening (UNHS) is feasible, cost effective, and successful in identifying hearing-impaired newborns [8-10]. After the American Academy of Pediatrics in 1999 and European Consensus Conference in 1998 recommended UNHS, it started to become widespread in USA and Europe [3, 11]. The efficacy of UNHS has also been demonstrated by the decrease in the age of diagnosis and age of hearing aid fitting since implementing newborn hearing screening [12-15].

Newborn hearing screening had first begun to be applied in Turkey in 1994 and 1998 at the Marmara and Hacettepe University hospitals, respectively. The UNHS program began to be implemented in some pilot hospitals, including the Gazi, Hacettepe, Marmara, and Dokuz Eylül University hospitals, at the end of 2003 in Turkey and spread to all 81 provinces in 2012 [16, 17]. Currently, the hearing screening is performed using transient evoked otoacoustic emission (TEOAE) and automated auditory brainstem response (ABR) tests, which are non-invasive and quick [18].

This study aimed to present the results of UNHS at a Training and Research Hospital in Istanbul with respect to literature and to determine the ages of diagnosis, hearing aid fitting, and cochlear implantation in newborns with hearing loss.

**OBJECTIVE:** This study aimed to present the follow-up results of newborns after universal newborn hearing screening at a Training and Research Hospital in Istanbul and to determine the ages of diagnosis, hearing aid fitting, and cochlear implantation in newborns with hearing loss.

**MATERIALS and METHODS:** A total of 5985 newborns were screened between December 2009 and August 2011 using the transient evoked otoacoustic emission test as the first two steps and automated auditory brainstem response (ABR) test as the third step. Newborns who failed the screening tests were referred to a tertiary hospital for clinic ABR and were followed up at least for 2 years.

**RESULTS:** Of 5985 newborns, 5116 (85.5%) completed the screening. Of 53 newborns who were referred to a tertiary hospital, 13 (0.25%) had a hearing impairment. The mean age of diagnosis, hearing aid fitting, and cochlear implantation were 6.1, 9.5, and 24.5 months, respectively. Among the risk factors for hearing impairment, neonatal intensive care (60%) and consanguineous marriage (50%) were the most common ones that were encountered.

**CONCLUSION:** Our results were consistent with the national literature. Consanguineous marriage may be a risk factor for hearing impairment where it is commonly practiced because consanguineous marriage is significantly high in parents of deaf children. The ages of diagnosis and hearing aid fitting are still beyond the recommended ages by the Joint Committee on Infant Hearing.

**KEYWORDS:** Universal newborn hearing screening, age, diagnosis, hearing aid, cochlear implantation
MATERIALS and METHODS

This study was performed between December 1, 2009 and August 31, 2011 at the Bakirköy Dr. Sadi Konuk Training and Research Hospital in Istanbul, which is a second-stage hospital, as a part of the Turkish National Newborn Hearing Screening Program. The study was approved by the local ethics committee. A total of 5985 newborns who were born in or referred to our hospital were included. Most of the risk factors for hearing impairment that were defined by JCIH in 2007 were recorded in a questionnaire (Table 1) [19]. The risk factors that were not included in our UNHS were the concern of the caregiver regarding hearing, language, speech, or developmental delay; chemotherapy; and head trauma. In addition, consanguineous marriage (first and second cousins) was evaluated as a risk factor. The screening program was performed by four audiometrists and was composed of three steps using the TEOAE test as the first two steps and automated ABR test as the third step. The newborns were tested when they were in a sleeping state in a quiet room. The screening equipment used was the AccuScreen PRO (MADSEN-GN Otometrics; Taastrup, Denmark), which can perform both TEOAE and automated ABR tests. The TEOAE test gives a “pass” or “refer” result. Newborns born in our hospital were tested using TEOAE on the first or second day of life before discharge, and newborns who were referred to our hospital from first-stage hospitals were screened on the same day. Newborns were divided into the following two groups: healthy newborns and high-risk newborns. In healthy newborns, when a “pass” response was achieved for both ears, it was accepted that the newborn had met the pass criteria. The parents of newborns who had met the pass criteria were informed regarding the delayed-onset hearing impairment and follow-up of their children regarding hearing loss was recommended. They were not called back for screening again as this implementation did not exist in our screening program. If the newborn did not have the “pass” response in one or both ears, the test was repeated after 15 days. The newborns who failed in the second step were examined by an ENT specialist. The newborns having debris or cerumen in their external ear canal or otitis media were treated. After treatment, the TEOAE test was performed, and the newborns who failed again were screened using automated ABR. If the ear examination of the newborn was normal, automated ABR was directly performed. Healthy newborns who failed the automated ABR test were referred to a tertiary hospital for clinic ABR. High-risk newborns were screened using TEOAE before discharge and called back after 1 week for automated ABR test, independent of whether they had a normal TEOAE screen or not. Because our hospital is a second-stage hospital, the newborns who failed the screening tests were referred to a tertiary hospital for clinic ABR. The referred newborns were followed up for at least 2 years, and the data regarding the age of diagnosis, level of hearing loss, and ages of hearing aid fitting and cochlear implantation were obtained through phone interviews of the family and hospital records. In this study, all descriptive statistical analysis were performed using NCSS (Number Cruncher Statistical System) 2007 Statistical Software (Utah, USA).

RESULTS

Between December 2009 and August 2011, 5985 newborns were screened. There were two groups: the first group comprised 4111 (68.7%) healthy newborns, while the second group comprised 1874 (31.3%) high-risk newborns (with risk factors of hearing impairment).
SNHL. Hearing aids were fit in all of the 12 newborns with bilateral SNHL, but the newborn with unilateral profound SNHL was decided to be followed-up. The age of hearing aid fitting for the 12 newborns with bilateral SNHL ranged from 4 to 18 months, with a median and mean age of 9.5 months. Of the 12 newborns with bilateral SNHL, 4 had received cochlear implant, 4 were lost to follow-up, and 4 were not suitable for cochlear implant. The age of cochlear implantation for the 4 newborns with bilateral profound SNHL ranged from 18 to 30 months, with a mean age of 24.5 months. Because four newborns’ parents changed their phone numbers, they were lost to follow-up. Three of the newborns who had moderate and severe bilateral SNHL and one newborn with hydrocephalus were not considered as good candidates for cochlear implantation. Of the 10 newborns with risk factors for hearing impairment, 6 (60%) had prolonged mechanical ventilation or neonatal intensive care, 5 (50%) had parents with consanguineous marriage, 1 (10%) had low birth weight, 1 (10%) had a family history of hearing impairment, and 1 (10%) had hyperbilirubinemia. The results of the 13 newborns with hearing impairment are shown in Table 3.

**DISCUSSION**

Congenital hearing loss, if not identified early and treated, affects a child’s skill to develop normal speech, language, and social skills. Therefore, early identification and intervention of congenital hearing loss are mandatory for a child’s linguistic and cognitive development. To achieve these goals, in 1994, JCIH suggested screening of high-risk newborns for hearing loss. Thereafter, many studies demonstrated that up to 50% of all newborns with congenital hearing loss do not have any risk factors. Hence, UNHS became the accepted strategy for the early identification and intervention of congenital hearing impairment in 1999. In Turkey, UNHS started to be implemented in some pilot hospitals at the end of 2003. Finally, in 2007, JCIH released a position statement approving the goals of calling for screening within 1 month of age, diagnosis within 3 months, and intervention within 6 months for all diagnosed infants. Furthermore, JCIH recommended benchmarks for the achievement of these goals. Screening before 1 month of age should occur for 95% of all newborns. Diagnosis within 3 months of age should occur for 90% of infants who were referred from the screening program. Hear-

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*Figure 1. Flowchart representing the results of the newborn hearing screening program*
The healthy and high-risk newborns, respectively. Because the newborn referral rate before comprehensive audiological evaluation should be <4% [19]. The referral rates after screening tests in the healthy and high-risk newborns were 0.1% and 3.4%, respectively. The overall referral rate was 1.0% (53 newborns). Our results were consistent with this goal. Of the 5985 newborns, 869 (14.5%) were lost to follow-up and 5116 (85.5%) were screened. Our rate of screening before 1 month of age was below 95%. The high rates of newborns lost to follow-up may be associated with the introduction of more measures in the screening program to reduce referral rates [20]. Other reasons for the high number of newborns lost to follow-up in our study may be the absence of a secretariat system to follow-up every failed newborn and remind parents regarding their future appointments, low degree of awareness regarding hearing loss in families and healthcare providers, and presentation of failed newborns’ anxious parents to the tertiary hospitals because of the absence of psychological support after screening. The tracking system of the program can be improved by a dedicated secretariat system, and detailed information and psychological support could be provided to the parents of failed newborns after screening to decrease the number of newborns lost to follow-up. Moreover, the awareness regarding hearing loss in families and healthcare providers should be increased with the assistance of publicity.

In our study, of 4111 healthy newborns, 3677 were screened and 3 (0.08%) were diagnosed with bilateral SNHL. Of the 1874 high-risk newborns, 1435 were screened and 9 (0.6%) were diagnosed with bilateral SNHL. However, in the literature, bilateral SNHL is present in 0.1%–0.3% of healthy newborns and 2%–4% of newborns in the intensive care unit population [18]. The low rate of SNHL among high-risk newborns may be associated with many reasons, such as few sample size, and a significant number of newborns lost to follow-up.

The prevalence of bilateral congenital hearing loss is estimated to vary from 1.5 to 6 per 1000 live births [7]. In this study, the prevalence of bilateral SNHL in newborns was 0.23% (12 in 5116 screened newborns), and the prevalence of unilateral SNHL was 0.02% (1 in 5116 newborns). In total, 13 (0.25%) newborns were found to have hearing loss. Bolat et al. [18] reported the first national data regarding the Turkish Newborn Hearing Screening Program with 764,352 newborns screened between 2004 and 2008. They stated that the prevalence of hearing loss had been 0.17%, ranging between 0.05% and 0.23% in 2004 and 2008, respectively. In 2008, the prevalence of bilateral and unilateral SNHL was found to be 0.12% and 0.09%, respectively, among 337,690 newborns. Our results were consistent with the national literature.

According to the benchmarks as set by JCIH, diagnosis by 3 months of age should occur for 90% of infants who were referred from the screening program and hearing aid fitting by 6 months of age should occur for 95% of infants with permanent hearing loss [19]. JCIH’s position statement in 2007 is based on for guiding principles, such as definitions, protocols, screening goal, audiological evaluation, and roles and responsibilities, in the UNHS program in Turkey. In addition, some updates were conducted according to the special conditions of our country by the UNHS Program Science Committee [17]. Some risk factors that were associated with hearing impairment, such as head trauma, and chemotherapy were excluded and consanguineous marriage was added as a risk factor [21].

<table>
<thead>
<tr>
<th>No</th>
<th>Degree of hearing loss</th>
<th>Age of Dx (mo)</th>
<th>Age of HAF (mo)</th>
<th>Age of Cl (mo)</th>
<th>Risk factors</th>
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<tbody>
<tr>
<td>1</td>
<td>Bilateral profound</td>
<td>9</td>
<td>10</td>
<td>Lost to F/U</td>
<td>None</td>
</tr>
<tr>
<td>2</td>
<td>Bilateral profound</td>
<td>5</td>
<td>6</td>
<td>Lost to F/U</td>
<td>None</td>
</tr>
<tr>
<td>3</td>
<td>Bilateral moderate</td>
<td>6</td>
<td>7</td>
<td>Not suitable</td>
<td>None</td>
</tr>
<tr>
<td>4</td>
<td>Bilateral profound</td>
<td>8</td>
<td>9</td>
<td>30</td>
<td>CM, NIC, CM, HB, FHHI</td>
</tr>
<tr>
<td>5</td>
<td>Bilateral profound</td>
<td>4.5</td>
<td>6</td>
<td>20</td>
<td>CM</td>
</tr>
<tr>
<td>6</td>
<td>Bilateral profound</td>
<td>3.5</td>
<td>6</td>
<td>30</td>
<td>NIC</td>
</tr>
<tr>
<td>7</td>
<td>Bilateral profound</td>
<td>4.5</td>
<td>15</td>
<td>Not suitable</td>
<td>NIC</td>
</tr>
<tr>
<td>8</td>
<td>Bilateral profound</td>
<td>3</td>
<td>4</td>
<td>18</td>
<td>CM</td>
</tr>
<tr>
<td>9</td>
<td>Bilateral severe</td>
<td>3.5</td>
<td>18</td>
<td>Not suitable</td>
<td>NIC</td>
</tr>
<tr>
<td>10</td>
<td>Bilateral profound</td>
<td>12</td>
<td>13</td>
<td>Lost to F/U</td>
<td>NIC</td>
</tr>
<tr>
<td>11</td>
<td>Bilateral severe</td>
<td>10</td>
<td>11</td>
<td>Not suitable</td>
<td>CM</td>
</tr>
<tr>
<td>12</td>
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<td>Lost to F/U</td>
<td>LBW, NIC</td>
</tr>
<tr>
<td>13</td>
<td>Unilateral profound</td>
<td>3</td>
<td>3</td>
<td>Not suitable</td>
<td>CM</td>
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</table>


Different newborn hearing screening programs have been used in different countries, but the most acceptable methods for UNHS include both the TEOAE and automated ABR tests [2, 24]. Therefore, we used the TEOAE and automated ABR tests in our UNHS program. The TEOAE test is non-invasive and quick. It is easy to perform and has high sensitivity. The test is adversely affected by ear canal debris or middle ear fluid, resulting in referral percentages of 5%–20% when it is performed in the first 24 h of life. The automated ABR test requires newborns to be in a quiet state or asleep at the time of testing, and it may be affected by middle ear fluid or external ear debris as well [15].

The referral rates were high in the first step with 31.6% and 28% in the healthy and high-risk newborns, respectively. Because the newborns are generally discharged from the hospital within the first 24 h, we usually performed the first step of screening using TEOAE in the first 24 h, and thus, this could lead to high referral rates. According to the American Academy of Pediatrics guidelines, screening by 1 month of age should occur for 95% of all newborns and the referral rate before comprehensive audiological evaluation should be <4% [19]. The referral rates after screening tests in the healthy and high-risk newborns were 0.1% and 3.4%, respectively. The overall referral rate was 1.0% (53 newborns). Our results were consistent with this goal. Of the 5985 newborns, 869 (14.5%) were lost to follow-up and
loss between 2001 and 2006 in New York was 8.7 weeks. Moreover, they found that hearing aid fitting by 6 months of age occurred for 39% of newborns and 61% were fit late or were lost to follow-up. Although we had a limited number of newborns with hearing loss compared with other studies, the results of our study demonstrate that there are significant improvements at the ages of diagnosis of hearing loss, hearing aid fitting, and cochlear implantation after the UNHS program started to be implemented in Turkey. Although these better results appear promising, the ages of diagnosis and hearing aid fitting are still beyond the recommended ages by JCIH. However, as Spivak et al. [24] demonstrated, it is not easy to maintain all of the benchmarks recommended by JCIH. There are many factors, such as low degree of awareness in families and healthcare providers, low socioeconomic situation of families, and few number of diagnosis and intervention services, that might contribute to the delays in diagnosis and hearing aid fitting [25].

Among the risk factors for hearing impairment, neonatal intensive care (60%) and consanguineous marriage (50%) were the most common ones that were encountered. Consanguineous marriage is a custom that is commonly practiced among the African, Asian, and Latin American communities, whether they live in their own countries or are settled in USA or Europe. It is also common in Turkey, and many studies in Turkey and other countries have reported that the siblings of consanguineous marriages have a notably higher incidence of autosomal recessive diseases comprising hearing impairment [27-31]. Sajjad et al. [28] conducted a study among 140 deaf school pupils and 221 non-hearing-impaired children in Pakistan. They found that parental consanguinity (i.e., first and second cousins) constituted 86.4% of deaf school children and 59.7% of non-hearing-impaired children. They concluded that the prevalence of parental consanguinity was notably higher in hearing-impaired children than in non-hearing-impaired children. Derekoy [31] performed a study among 130 deaf students in Turkey and stated that parental consanguinity was noted among the parents of 64 (49.2%) children in the school, and this rate was found to be significantly higher than the average of Turkey.

Our results were consistent with the national literature. Consanguineous marriage may be considered as a risk factor for hearing impairment where it is commonly practiced because consanguineous marriage is significantly high in parents of deaf children. After the UNHS program started to be implemented in Turkey, this is the first study that determined the ages of diagnosis, hearing aid fitting, and cochlear implantation in newborns with hearing loss. Although the improvements demonstrated by the results of our study appear promising, the ages of diagnosis and hearing aid fitting are still beyond the recommended ages by JCIH. As our study had a few sample size, a large number of patients must be studied to clearly determine whether the goals endorsed by JCIH are achieved in Turkey.

Ethics Committee Approval: Ethics committee approval was received for this study from the ethics committee of Istanbul Bakirköy Dr. Sadi Konuk Training and Research Hospital (06/10/2010 with a protocol number 55).

Informed Consent: Written informed consent was obtained from the parents of the patients who participated in this study.

Peer-review: Externally peer-reviewed.


Conflict of Interest: No conflict of interest was declared by the authors.

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REFERENCES


