The results of national newborn hearing screening (NNHS) data of 11,575 newborns from west part of Turkey

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Abstract. OBJECTIVE: In this study, we aimed to review the National Newborn Hearing Screening (NNHS) programme data obtained from Corlu, located in west part of Turkey for the last 4 years.

SUBJECTS AND METHODS: A total 11,575 newborns that were either born in Çorlu State Hospital or referred from other Health Care Centers, between September 2009 and November 2012 were included into the study. Automated-Transient Evoked Otoacoustic Emission (A-TEOAE) test and Automated-Auditory Brainstem Response (A-ABR) were used as screening tests. When the newborn had failed at the initial A-TEOAE test, then the test was repeated after 15 days. If the same result was obtained at the second test; the newborns were referred for ENT examination. A-TEOAE and A-ABR screening tests were performed as the third stage evaluation. The failed newborns were referred for clinical ABR test.

RESULTS: Out of 11,575 newborns, 593 (5.12%) had failed the test and they were referred for clinical ABR. Out of these 593 neonates, 470 had passed the diagnostic ABR test at the referral center. Bilateral and unilateral sensori-neural hearing loss (SNHL) was detected at 15 and 7 babies respectively, 10 of 22 of these babies had risk factors such as family history of hearing loss, and parental consanguinity.

CONCLUSIONS: The final hearing impairment and risk factor rates of our study was similar with literature. Although referral rates and the number of default babies gradually decreased in the last 4 years; definitive diagnosis and hearing aid initiation times need to improvement.

Key Words:
Hearing loss, The National Newborn Hearing Screening (NNHS) programme, Newborn.

Introduction

Hearing loss (HL) is among the most common congenital anomalies with the rate of 0.1-0.3%1,2. This rate is much higher than the total of all the congenital metabolic diseases such as hypothyroid and phenylketonuria for which routine screening tests have been performed for many years3. Furthermore, the cost of newborn screening metabolic are quite economical compared to the other routine screening tests that have been conducted for many years for congenital disorders4.

Early diagnosis is very important for the children with HL, and it plays an essential role in reducing its destructive results. Lingual development of babies during the initial period is rather fast. It is important for the babies to have a normal hearing capability from the points of social, emotional and mental progress along with speech development. In case the infant’s hearing loss is not detected at an early stage, this leads to a disability which will have a lifelong influence in the person such as impediment in speech, intellectual performance weakness, social inadaptability and emotional disorders5.

In 1994, the American Joint Committee on Infant Hearing (AICIH) recommended the universal detection of hearing loss within the first 3 months of age and an appropriate intervention should be made no later than 6 months of age6. The average age at which hearing loss is detected decreased from 12 to 24 months, to 3 to 6 months after the introduction of screening programs for hearing in newborns in United States. In addition, the average age at which children receive hearing aids has been reduced from 13-16 months to 5-7 months7. In subsequent years many countries formed their own National Newborn Hearing Screening (NNHS) programmes.
In Turkey, NNHS was first introduced in two of the university hospitals in 2003. In the following years it became to be used in many hospitals across the country. NNHS can be executed in three different types of hospitals which are distinguished depending on the available type of diagnostic equipment: only execute Automated-Transient Evoked Otoacoustic Emissions (A-TEOAE) tests (Level-1 center); execute both Automatic A-TEOAE and Auditory Brainstem Response (A-ABR) tests (Level-2 center); and diagnostic clinical ABR (Level 3 center).

We aimed to present the results of NNHS that was applied to 11,575 infants at Corlu State Hospital (CSH), a reference hospital in the Thrace region in Turkey, between September 2009 and November 2012 in comparison with existing literature.

Subjects and Method

This study was conducted in Çorlu State Hospital (CSH), 2nd Step Newborn Hearing Screening Center (2nd SNHSC), between September 2009 and November 2012. All steps of the study were planned and continued according to the principles outlined in the Declaration of Helsinki. This study was presented in The Meeting of American Academy of Otolaryngology-Head and Neck Surgery, 2013; and summary was printed in Otolaryngology-Head and Neck Surgery.

Newborn Hearing Screening (NHS) Programme Details of Health Ministry of Turkey

A. Centers

Newborn Hearing Screening Programme was conducted by Screening Centers of 1st-3rd Level in Turkey. In diagnostic center; it was aimed to diagnose around 3 months by electrophysiological and behavioral tests; and then hearing aid fitting and orientation training started. Sometimes, process of diagnosis are delayed to 5-6 months in practice due to some problems such as an appointment system, families’ fail to bring the baby to the center, difficulties on conclusion of the diagnostic tests, etc. These centers are classified as below:

1. 1st Level Newborn Hearing Screening Center (1st LNHSC): The first screening test was performed by A-TEOAE before discharge after the birth. These centers may be Maternity Hospital, State Hospitals or University Hospitals.

2. 2nd Level Newborn Hearing Screening Center (2nd LNHSC): In this center, A-ABR equipment and ENT doctor (for ENT examination) were present. In this center, A-TEOAE and A-ABR tests were performed together.

3. 3rd Level Newborn Hearing Screening Center (3rd LNHSC): Clinical audiologist and equipment for audiological diagnosis were present in the comprehensive Audiology Clinic. In this center, diagnostic clinical ABR was performed.

B. Screening test method in the centers: Screening tests with the sequence of the algorithm are as follows:

1. At 1st LNHSC: At the hospital just after 2-3 days of the birth, hearing screening before discharge in the hospital where the birth was performed: Automated otoacoustic emission screening (AOAE) was performed initially.
   - If the baby passed bilaterally, the family was given information.
   - If the baby cannot pass the A-TEOAE, he/she will be invited to control screening test after 15 days.
   - If there is A-ABR device in 1st SNHSC, A-ABR is done to the baby who cannot pass the second A-TEOAE. If there is not A-ABR device in 1st LNHSC, the baby who cannot pass the second A-TEOAE, will be referred to 2nd LNHSC.

2. At 2nd LNHSC: 15-day after the birth: If the baby cannot pass the 15th day-second screening test, A-ABR and ENT examination will be performed at 2nd LNHSC. Babies with risk factors and babies to not have passed A-TEOAE must be screened by A-ABR and ENT examination before being routed to the center (3rd LNHSC).
   - If the baby passed bilaterally, the family was given information.
   - If the baby cannot pass from one or two ears, he/she will be directed to 3rd LNHSC. The latest time to go to 3rd LNHSC was the babies’ 2-month age.

3. At the comprehensive 3rd LNHSC, by using behavioral and electrophysiological test methods, the diagnosis of HL was performed. During diagnosis period, it was noticed that auditory stimuli thresholds vary depending on age of the child. On the basis of these criteria, HL of the child were diagnosed and classified.
Subjects

The research was conducted on 11575 infants who were born in CSH or referred by the medical centers in the vicinity between September 2009 and November 2012. Hearing Screening Test (HST) was performed just before the baby was discharged from the hospital by 2 audiometrists and 1 nurse with relevant certificate in a quite room exclusively designed for this purpose.

The babies who were able to pass one of the tests were considered to have passed. Otherwise, they were referred to another city for a diagnostic clinical ABR, called the third level NHS center (3rd LNHS C) in Turkey. Written screening test results were given to the families. Screening data of the babies were recorded in the NNHS follow-up form. Transferred babies who didn’t give any feedback to parents or the guardians were called by phone, and the flow of information was conducted.

Instrumentations

Automated-Transient Evoked Otoacoustic Emissions (A-TEOAE)

All A-TEOAE screenings were performed by Otoport Lite OAE System (Odyodynamics, Liverpool, UK). Tests were done while baby was asleep or when being calmly held by mother or nursemaid. Probes were selected in proportion with the size of the external auditory canal. The babies were given a bilateral A-TEOAE checking and the results were evaluated as “passed” or “failed” according to the test results.

Automatic Auditory Brainstem Response (A-ABR)

 Babies who couldn’t pass the second TEOAE tests until January 2012 were transferred to another medical center in our city for A-ABR tests. Following January 2012 our hospital obtained an A-ABR device, which enabled us to do the measurements in the last one-year. The A-ABR device was of AccuScreen brand (Madsen, Denmark).

Tympanometric Test

At 15-day after the birth: If the baby can not pass the 15th day-second screening test, A-ABR and ENT examination will be performed. For these babies in which ENT examination was done, tympanometric tests were also performed and acoustic reflex measurements were made using the Interacoustic AZ 26 impedance device (Interacoustic A/S, Denmark) to get information about the presence of effusion in the middle air, the state of the eardrum, middle ear pressure and acoustic reflex arch in the babies.

Results

NNHS results were shown on Table I, Figure 1:

- Out of the total number of 11575 infants who underwent hearing screening by A-TEOAE, 8652 of them (74.74%) passed the first test.
- 2923 (25.26%) of the infants were invited for the second A-TEOAE test though 38 (0.32 %) of them did not attend to the hospital. Therefore, 2847 (24.59%) infant examined at the second stage and 1272 (10.98 %) of them passed the second screening by A-TEOAE: 1575 (13.60%) failed at this stage. An Otolaryngologist examined those who failed the second A-TEOAE, and any pathological problems detected were cured if possible and other patients (1548 babies-13.37%) were invited for the third stage.
- We should mention that we purchased the A-ABR device and thus introduced its use in our hospital after January 2012. Up to that date the

Table I. NNHS Results*.

<table>
<thead>
<tr>
<th>Applied tests</th>
<th>Passed</th>
<th>Failed</th>
<th>Defaults</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>N</td>
<td>%</td>
<td>N</td>
<td>%</td>
</tr>
<tr>
<td>1st A-TEOAE</td>
<td>8652</td>
<td>74.74</td>
<td>2923</td>
<td>25.26</td>
</tr>
<tr>
<td>2nd A-TEOAE</td>
<td>1272</td>
<td>10.98</td>
<td>1575</td>
<td>13.60</td>
</tr>
<tr>
<td>A-TEOAE+A-ABR</td>
<td>928</td>
<td>8.01</td>
<td>593</td>
<td>5.12</td>
</tr>
<tr>
<td>Clinical ABR</td>
<td>470</td>
<td>4.06</td>
<td>22</td>
<td>0.19</td>
</tr>
</tbody>
</table>

*NNHS: National Newborn Hearing Screening Programme.
babies who could not pass the first two TEOAE tests in our practice would be referred to another medical center within the province. In the third stage screening, A-TEOAE and A-ABR tests were performed together on 1548 (13.37%) babies though 27 (0.23%) of the babies were not attended to the hospital for screening. In this stage, 928 (8.01%) babies passed; and 593 (5.12%) babies who failed the tests in one or two ears were referred to third level, for their diagnostic clinical ABR tests (Table I).

– Of these referred 593 (5.12%) babies, the diagnostic clinical ABR records of 101 (0.87%) could not be reached in the following period. One reason of this failure was the fact that the telephone numbers of the families had changed. The families to be communicated said that they either did not bother to take the baby to the referred clinic or they could not do it due to socioeconomically inconveniences. It was found out that 470 of the babies referred for clinical ABR, 22 of them were diagnosed as sensori-neural hearing loss (SNHL). The age when hearing impairment was detected using diagnostic ABR ranged from 2 to 9 months (mean: 4.45 months).

**Side of Sensori-neural Hearing Loss (Unilateral or Bilateral)**

Fifteen (68.18%) of the 22 babies with SNHL had bilateral HL whereas 7 (31.82%) of them had unilateral hearing loss (Table II). 14 of the babies with unilateral hearing loss began to use hearing aid at 8.86 months (mean); one of the newborns received cochlear implant at the 12th month. Finally, 1 baby has not been able to have any hearing aid yet because of socioeconomical inconveniences although the baby has reached 18 months of age.

**Risk Factors for Sensorineural Hearing Loss**

In 10,852 babies who passed screening tests (A-TEOAE or A-TEOAE+A-ABR), there were 549 risk factors for hearing loss. In 593 babies who had been referred for clinical ABR tests, there were 167 risk factors. In 22 babies who had diagnosed as SNHL by clinical ABR, there were 12 risk factors which was detected in 10 of 22 babies with HL. In 4 of the babies who had hearing loss there was a familial HL history, 3 of them had parental consanguinity. In 2 of the babies, there were both of these problems. One baby had been taken to intensive care unit and

<table>
<thead>
<tr>
<th>N</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td>Bilateral SNHL</td>
<td>15</td>
</tr>
<tr>
<td>Unilateral SNHL</td>
<td>7</td>
</tr>
<tr>
<td><strong>Total</strong></td>
<td><strong>22</strong></td>
</tr>
</tbody>
</table>

*SNHL: Sensorineural hearing loss.
stayed there for 11 days due to umbilical cord problem and meconium aspiration causing Respiratory Distress Syndrome (RDS).

Head and face anomalies were detected in 2 babies and 1 baby was diagnosed as Trisomy 21. Another baby had been kept in the intensive care unit for 14 days due to newborn hyperbilirubinemia. All this can be seen in Table III. No risk factor was detected in 12 of the 22 babies with hearing loss.

Table IV and Figure 2 displays screening tests by A-TEOAE; or ATEOAE+ABR and clinical ABR referrals in 2009-2012. According to the data displayed, in the years of 2009, 2010 and 2011 the rates of referred cases were 246 of 3263 cases in 2009; 158 of 3014 cases in 2010, 157 of 3181 cases in 2011 and 32 of 2117 cases in 2012.

Discussion

Universal newborn hearing screening (NNHS) has been widely accepted and applied worldwide. UNHS is designed to identify congenital or acquired hearing loss in a short time. Early identification of hearing loss is essential to avoid problems related to speech, language, social life and education to occur at a later stage of life. This program requires that all babies be tested before leaving the hospital. Two-level TEOAE or the two level TEOAE then AABR protocols or by itself AABR are the most frequently applied UNHS methods.

TEOAE was used as an initial screening tool because it is technically easier and quicker to carry out and can accommodate a large number of patients. Furthermore, TEOAE is cheaper than the

Table III. Risk factors for hearing loss*.

<table>
<thead>
<tr>
<th>Risk factors</th>
<th>Passed screening tests by A-TEOAE or A-TEOAE+A-ABR (n=10852)</th>
<th>Failed at hearing screening tests and referred for diagnostic ABR (n=593)</th>
<th>Diagnosed as SNHL (n=22)**</th>
</tr>
</thead>
<tbody>
<tr>
<td>Family history of hearing loss</td>
<td>173</td>
<td>56</td>
<td>4</td>
</tr>
<tr>
<td>Parental consanguinity</td>
<td>252</td>
<td>50</td>
<td>3</td>
</tr>
<tr>
<td>Low birth weight (low 1500 gr)</td>
<td>61</td>
<td>27</td>
<td>0</td>
</tr>
<tr>
<td>RDS</td>
<td>13</td>
<td>5</td>
<td>1</td>
</tr>
<tr>
<td>Hyperbilirubinemia</td>
<td>24</td>
<td>13</td>
<td>1</td>
</tr>
<tr>
<td>Head and face anomalies</td>
<td>15</td>
<td>7</td>
<td>2</td>
</tr>
<tr>
<td>Congenital genetic disorders</td>
<td>11</td>
<td>9</td>
<td>1</td>
</tr>
<tr>
<td>Total number of risk factors</td>
<td>549</td>
<td>167</td>
<td>12</td>
</tr>
</tbody>
</table>

* RDS: Respiratory Distress Syndrome, SNHL: Sensorineural hearing loss; **12 risk factors were detected in 10 newborns with SNHL.

Table IV. Hearing screening tests and clinical ABR referrals in 2009-2012.

<table>
<thead>
<tr>
<th>Years</th>
<th>Screened by A-TEOAE; or A-TEOAE+A-ABR</th>
<th>Referred for clinical ABR</th>
<th>Default</th>
</tr>
</thead>
<tbody>
<tr>
<td>2009</td>
<td>3263 (28.19%)</td>
<td>246 (2.12% of total, 7.96% of 2009 year screening)</td>
<td>53 (0.45%)</td>
</tr>
<tr>
<td>2010</td>
<td>3014 (26.03%)</td>
<td>158 (1.36% of total, 5.24% of 2010 year screening)</td>
<td>24 (0.20%)</td>
</tr>
<tr>
<td>2011</td>
<td>3181 (27.48%)</td>
<td>157 (1.35% of total, 4.93% of 2011 year screening)</td>
<td>21 (0.18%)</td>
</tr>
<tr>
<td>2012</td>
<td>2117 (18.28%)</td>
<td>32 (0.27% of total, 1.51% of 2012-year screening)</td>
<td>3 (0.02%)</td>
</tr>
<tr>
<td>Total</td>
<td>11575 (100.0)</td>
<td>593 (5.12% of total)</td>
<td>101 (0.87%)</td>
</tr>
</tbody>
</table>
auditory brainstem response, and this makes it more cost-effective as a screening method. Disadvantages of TEOAE are that the effusion or debris accumulated in the middle ear and external ear canal may mislead the diagnosis. Also the eighth nerve and auditory brain stem dysfunction cannot be detected with TEOAE. AABR is not affected by above mentioned middle ear and external ear canal pathologies and it also provides us with valuable information in highlighting the problems concerning central auditory processes. In the two-step detection, infants with auditory neuropathy cannot be detected by single TEOAE. Newborns affected by auditory neuropathy can be diagnosed only through automated or diagnostic ABR test execution. Korver et al found a review study prevalence of auditory neuropathy in children to vary between 0.006% and 0.03%. Auditory neuropathy is characterized by normal TEOAE and fluctuating AABR. Nevertheless, the need to place electrodes and that the process takes longer time to be implemented and finally the necessity to do the screening only while the baby is asleep are among the disadvantages of AABR.

It has been emphasized in various studies that having false positive results at minimal levels saves time and unnecessary further tests as well as it protects the families from unnecessary anxiety thus increasing the success of the program. Tatlı et al reported the percentage of false positive rates in their studies applied the two-level of TEOAE as 0.3% at Izmir; and Ghirri et al underwent both TEOAE and AABR screening together upon 8113 neonates have found newborn hearing screening false-positives rates 0.54% at Pisa.

In our study, we found that 74.74% of the newborns passed first screening step by A-TEOAE. This is very close to Ahmad et al and Kucur et al studies, they had found 74.5% and 76.9% respectively. The fact that the probe used especially for premature infants cannot exactly fit into the External Auditory Canal (EAC) increases the false positive cases. Because the amnion fluid that can be found in EAC in the newborns in the first 24 hours increases the false cases of TEOAE. It has been suggested in a number of publications that the ideal test time should be between 24 hours after the birth and immediately before the baby is discharged (in the studies about the cases when babies are kept in the hospital for a few days). Furthermore, baby becomes more active and starts to utter sounds at the end of the first month. Due to this reason it has been suggested and advised that the hearing screening test should be done within the first month. It is not always possible to conduct the newborn screening procedure especially in the developing countries where births are not often given in hospitals and, even if the birth is given in a hospital, the baby is discharged in a few hours after the birth. This can explain another reason for having high rates of false positive cases of the first TEOAE test mentioned in our study too.

Adding the AABR tests to the routinely done A-TEOAE in the screening program increases the success and efficiency of the NNHS, and it
further decreases the number of babies transferred to other centers as well and contributes positively to the elimination of the family stress.

Ahmet et al.24 found prevalence of default for second and third screening was 33.9% and 40.7%, respectively. Compared with Ahmet et al.24 our default rates of second and third screening tests was very low, we thought about the reason of this might be easy to transport and communication to our NNHS team depends on close distant of our province. Even though our low default rates at our hospital, our ratio for diagnostic ABR referrals was not high. Korres et al.25 had screening of 76560 neonates their large series and reported 98% pass rate at first screening with TEOAE. Whereas their follow-up defaults was 981 (62%) TEOAE and AABR stage and 249 (69%) at clinic diagnostic ABR tests. Default rate of our study was second and third screening 0.32% and 0.23% respectively; it was 17.03% (101/593) of newborns that referred for diagnostic ABR to other provinces for third level definitive diagnosis. We found our default rates resemble with Korres et al.25.

One of the possible causes of variation in the level of hearing can be related to inflammation of the middle ear. It has been reported middle ear opacity perform with high resolution computed tomography (HRCT) in about 40% of children with bilateral hearing impairment greater than 50 dB identified by newborn hearing screening15. The differential diagnosis of temporary hearing loss due to middle ear effusion or SNHL need to complete evaluation for middle ear pathology. We used otoscopic examination and 226 Hz tympanometry. Examination of the tympanic membrane in newborns may be inaccurate due to technical challenges, such as narrow ear canals, the presence of cerumen, and the lack of communication, and may also contribute to a misdiagnosis of middle ear effusion25,26. Another possible reason of false positive results and extention diagnost ic process could be due to delayed hearing auditory pathway maturation. Such cases have been reported by several authors as well17,27.

Paradise et al28 in a study of infants less than 7 month old demonstrated that the result of tympanometry is often normal in the presence of middle ear effusion. Furthermore, AJCIH has recommended that these infants be screened by 1,000 Hz tympanometry instead of standard 226 Hz tympanometry26,29. In the future we hope to use 1,000 Hz tympanometry to prevent these type false results.

After January 2012, referral rates and the number of default babies dropped significantly. This indicates that our experience in screening tests has, in time, increased considerably; and that we were able to apply both A-TEOAE and A-ABR together even in the early stages in our programme.

We concluded that not having a diagnostic ABR device in our province is another reason of the high number of failed follow-up newborns. Certainly, conducting the diagnostic ABR tests in nearby centers on the babies who could not pass the screening tests would increase our success in the future.

Despite the overall benefits of UNHS is increasingly clear, boundaries exist. Less severe congenital hearing loss (less than 30 dB or 40 dB) is not detected in most programs UNHS. Some progressive or late-onset hearing impairment is also not detected by a newborn screening program. AJCIH in the 2007 Position Statement has identified the problem of late onset hearing loss and the risk factors identified audiology requires monitoring during the first years life29. In recent years, there has been an increased focus on late onset hearing loss17. Progressive or late onset hearing impairment may have different causes (genetic predisposition infections, etc.) and represents a relatively large percentage (20-30%) of hearing loss in children16,17. In our study, 15 of 22 infants (68.18%) were diagnosed as bilateral SNHL and 7 of 22 infants (31.82%) were diagnosed as unilateral hearing loss. We aimed subsequent studies, passed our screening programs, investigated our newborns’ late onset of hearing loss.

We had a prevalence rate of SNHL 1.9 in 1000 live births; this is quite close to the rate abroad17,24,25 and domestic5,8,24,33 relevant literature. Hearing impairment in Turkey is estimated to be at least as high as in other countries. In a hearing test programme between the school children, hearing loss was found at a rate of 2.5/1000 among the 8.7 million students in Turkey33. We concluded that our results have been in accordance with the results of Johnson et al when their 20-30% late onset hearing loss is added to this ratio of our 0.19% SNHL.

During the full 8 year study period, from 1992 to 1999, 148240 newborns were screened by Mehl et al34 and it was found that the presence of high-risk factors was 47% at totally permanent hearing loss. Another study reported that an admission to intensive care for more than two days
increased the risk of getting the hearing impairment to 10-fold. Meanwhile there is no consensus in the literature about the most common etiologic causes.

Whereas Ghirri et al. found neonatal respiratory distress as 20.58%, Botelho et al. reported 33% hyperbilirubinemia as most frequently risk causes. In accordance with the relevant literature our 10 out of 22 newborns with SNHL had at least one risk factors. A total 10 newborns with SNHL had risk factors of family history of HL, Parental consanguinity, head and face anomalies, hyperbilirubinemia and Respiratory Distress Syndrome.

In case at least 95% of the newborns who failed the hearing screening test go through further audiological test of diagnostic ABR which is considered to be an efficient screening program by the AJCHI. In this study, the age when hearing loss was detected using diagnostic ABR ranged from 2 to 9 months (mean: 4.45 months). The age at hearing aid-fitting in this study ranged from 5 months to 16 months (mean: 8.86).

Conclusions

Our findings are longer than recommended AJCHI recommendations about detection of hearing loss within the first 3 months of age and an appropriate intervention should be made no later than 6 months of age. We aimed to improve our program in accordance with the Committee in the future.

Acknowledgements

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Conflict of Interest

The Authors declare that there are no conflicts of interest.

References


The results of national newborn hearing screening (NNHS)


