Survey on infant hearing loss at Caritas Baby Hospital in Bethlehem-Palestine

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Dedication: the article is dedicated to the patients evaluated at Caritas Baby Hospital and their mothers and to the hospital staff, whom have the dream to make Caritas Baby Hospital a Center of excellence, ensuring patient and staff safety and quality in all activities.
Abstract

The research describes the epidemiology of the infants’ hearing loss (IHL) of patients less than 3 months of age in the only pediatric hospital in Palestine. The purpose of this study was to highlight IHL loss as a major health problem in Palestine and to assess the first available data about newborn hearing screening between September 25, 2006 and December 31, 2011. Data was uploaded and analyzed using Microsoft Excel and the Statistical Package for the Social Sciences software (SPSS version 21). A total of 8144 infants were tested, 4812 (59%) were males and 3332 (41%) were females. A percentage of 72 (5886) of the infants were from Bethlehem district, 25% (2044) from Hebron district, while 3% (214) from the other Palestinian districts (Jericho, Ramallah, Nablus, Jenin and Jerusalem). The Transient Evoked Otoacoustic Emissions (TEOAE) and Automated Auditory Brainstem Response were used according to the manufacturer guidelines. The results were interpreted according to the American Academy of Pediatrics, the National Institutes of Health, and the European Consensus Development Conference on Neonatal Hearing Screening. Out of the 8144 infants tested, 1507 (14.6%) infants did not pass the 1st test 477 (32.8%) of the 1507 failed retesting, while 498 (33%) patients did not come for follow-up. Only 152 (31.9%) of the patients that failed retesting went to an audiologist. The audiologist evaluation revealed that 101 (66.4%) patients presented with mild-moderate or profound hearing loss according to the Bureau International of Audiophonologie, while 44 (28.9%) patients presented Otitis media and 7 cases (4.7%) had no problems. The overall unadjusted percent of hearing loss is 1.24%, and the adjusted overall percent is 1.85%. Chart review showed that jaundice, sepsis, prematurity, lung disease were more common among the affected patients. The high prevalence of childhood deafness in Palestine is of utmost importance and needs the immediate attention of the Palestinian government to combat this problem. Meanwhile, Caritas Baby Hospital took the initiative to start a newborn hearing-screening unit utilizing the TEOAE method.
Introduction

In August 2006, Caritas Baby Hospital (CBH) welcomed a proposal by the medical audiologist for implementing a universal neonatal hearing screening. This was, in part, due to the high prevalence of childhood deafness in Palestine, as was noted by our initial report, and to the lack of any screening method in Bethlehem.\(^1\) We initially reported high prevalence of severe hearing loss in the Palestinian population during a study performed in the year 2006.\(^2\)

In that study, we concluded that screening for deafness should be extended to all newborns and parents should be educated on potential problems their children might face because of hearing loss. It started in the Department of Neonatology adopting the method of otoacoustic emissions (OAE). Hereditary deafness affects approximately 8‰ of the Palestinian population, and up to 3-15% in isolated villages.\(^1\) There are several sociological implications of hereditary deafness, including isolation and abandonment for those who are deaf, and lack of educational strategies for those who lose their hearing in early life. In some Palestinian villages, the prevalence of infantile congenital deafness is among the highest in the world due to genetic mutations, in particular in the gene 26 (GJB2) site on the long arm of chromosome 13.\(^1\) When compared to other countries, severe sensor neural hearing loss has a prevalence rate of 0.4 (Japan) and 1.48 (Denmark) per 1000 children. In other countries with low socioeconomically status the prevalence appear to be doubled than other developed countries (1.3-2.7 per 1000).\(^3\)

The American Joint Committee on Infant Hearing Screening established a list of risk factors that make the baby fall in *at risk register* for deafness.\(^4\) This protocol identifies only 50% of children with hearing impairment. Infants with no health problems during pregnancy or at birth may give a normal test result, although they are suffering from deafness. For these reasons, it was suggested by the American Joint Committee of Infant Hearing Screening to apply the neonatal screening to the entire population within three months of age, not only
those with high risk.

The OAE are considered an ideal tool for universal screening by several institutions as the American Academy of Pediatrics, the National Institutes of Health, and the European Consensus Development Conference on Neonatal Hearing Screening. The test can be performed within 3-5 minutes per infant and because of the ease of the testing; it can be performed after the first hours of child’s life. The sensitivity of OAEs is slightly less than 100% compared to 95% of Auditory Brainstem Response (ABR). The limitation of the OAE testing is that it is a periphery testing and does not evaluate the ears full functionality. The childhood deafness, as well as the risk of delay in language acquisition, also, determines problems in cognitive, emotional-affective and social development.

In this manuscript, we report the epidemiology of childhood diseases that lead to deafness in Palestinian patient less than 3 months of age in the entire West Bank. Also, the manuscript highlights infant hearing loss as a major health problem in Palestine.

**Materials and Methods**

**Study location and study period**

CBH is an 82-beds hospital located in the district of Bethlehem, Palestine. Patients seen at the different medical services of the hospital come from all over Palestine, in particular, from the Southern districts; Bethlehem and Hebron districts. A total of 8144 patients were included in the study, 4812 (59%) were males and 3332 (41%) were females; either admitted or visited the outpatient clinics between September 25, 2006 and December 31, 201. The distribution of the patients by districts was as follows: 72% (5886) of the infants were from Bethlehem district, 25% (2044) from Hebron district, while 3% (214) from the
other Palestinian districts (Jericho, Ramallah, Nablus, Jenin and Jerusalem).

In 2009, the service was expanded to include *Normal newborns*, particularly from the two districts: Bethlehem and Hebron.

**Data collection**

This study was a retrospective study on patients referred to CBH Outpatient clinics for Hip ultrasound, Hearing test, or for follow-up. The study was approved by the CBH Executive Committee, Medical Research committee and supported by the local staff. The study was carried out by utilizing the data from children who underwent audiological screening and other clinical documentation. The collected data were: hospital identification number, date of birth, sex, place of origin, district of origin, the date of testing, results of the screening and the result of the retest, medical diagnosis for whom failed the 1st test, audiologist doctor’s report.

**Disease groups**

Given the variety of range of medical diagnosis that the patients had, it was necessary to identify the most common diseases. Seven diseases were chosen (jaundice, prematurity, infection, lung disease, cardiac, gastrointestinal and others).

**Data analysis**

Data was collected and analyzed using Microsoft Excel with the methods of descriptive statistics and Statistical Package for the Social Sciences software (SPSS version 21). Data was represented as counts and percentages. Chi-square test or Fisher’s exact test was used to compare percentages. A P-value of <0.05 indicates statistical significance.
**Otoacoustic emissions test**

This test consists of low-intensity sounds emitted by the outer hair cells of the cochlea, produced spontaneously or evoked by auditory stimulation and recorded by a tiny microphone inserted into the ear canal. It is a quick and basic form of hearing test that involves the use of a foam tip that will be inserted just inside both of your child’s ear canals. These foam tips will be connected to a handheld device that will make the recording and give a pass/fail result. The child will not experience any pain since the probe will be placed just inside the ear. This test will take up to 10 minutes.

The sensitivity of OAEs is slightly less than 100% compared to 95% of ABR. The limitation of this test is testing the periphery only without assessing the acoustic full functionality of the street noise.\(^1\) The TEOAE are obtained in response to transient stimuli of 80 decibel. Their presence is determined by units of acoustic energy transmitted from the cochlea in a retrograde fashion through the middle ear. The limit of this method is the high rate of false negative results, especially in cases of auditory neuropathy. This disease is characterized by a delay of language. The etiology of this disease is assumed to be unknown, genetic, due to jaundice, anoxia or prematurity.

**Stem auditory evoked potentials (auditory brainstem response)**

It is a method for audiological screening of infants at risk. It is characterized with the highest sensitivity and reliability and presenting a very low number of false negatives and false positives.

The examination is performed during spontaneous sleep in the newborn. In case of positivity, another check has to be performed before dismissing the infant. The advantages of this method is the accuracy and competence of the examiners.\(^1\)

The use of ABR screening procedures has been developed to automatically detect the
presence or absence of response, and to reduce cost and time of the examination. Automated ABR is a more sophisticated form of hearing test than TEOAE and involves placing 3 electrodes: 1 on the forehead, 1 behind the neck and 1 on the shoulder. A small probe is placed on the outer side of the ear canal of each ear. The electrodes and probe are attached to a handheld device, which takes the reading. This test takes up to 20 minutes. It may, however, be done much quicker (less than 10 minutes) if the child is very calm. This test will not cause any pain to your child.

**Tympanometry**

Tympanometry is a test that is performed to detect the presence of any fluid in the ear, which can be a cause for hearing loss. It involves inserting tiny probes (foam tips) just inside the ear canal that send a pressure signal toward the ear drum and receive the signal back from it and making a recording on an attached handheld device. This test will not cause any pain to your child and can take up to 10 minutes.

We started to use the Tympanometry if the patient failed either TEOAE or the Automated ABR.

**Study procedures**

For the Neonatal hearing screening, the child will be assessed for risk factors for hearing loss. A risk factor survey will be provided to the parents for this purpose and could take up to 10 minutes of their time to complete. This survey will be used to determine whether the child has any existing risk factors that are associated with hearing loss. The survey includes questions related to: i) illness or condition requiring admission of 48 hours or greater to a Neonatal Intensive Care Unit (NICU); ii) stigmata or other findings of a syndrome known to include Sensorial neural HL or conductive HL; iii) family history of
permanent childhood HL; iv) craniofacial abnormalities including those with morphological abnormalities of the pinna or ear canal; and v) evidence of in utero infections such as cytomegalovirus, herpes, toxoplasmosis, or rubella.

If the child is NOT identified with any risk factors, he/she will be asked to undergo an initial screening hearing test. The test is called TEOAE. If the child passes the hearing screening in both ears, means that at the time of screening he/she is unlikely to have a hearing loss.

If the child fails the initial TEOAE screening hearing test in one or both ears, or he/she has risk factors for hearing loss, he/she will be asked to undergo further hearing test called Automated ABR. If the child passes the Automated ABR in both ears he/she will go home.

If the child fails the Automated ABR test one or both ears, he/ she will you will undergo Tympanometry test.

If the child’s Tympanometry test results are normal, he/she will be asked to undergo a diagnostic ABR test, going to the Audiologist nearest to his/her town. Diagnostic ABR is a diagnostic hearing test that is more sophisticated and time-consuming than the screening hearing tests - TOAE and Automated ABR. It is very similar to the Automated ABR but this test is more complete and tells us exactly how much and what kind of hearing loss the child has. This diagnostic hearing test can take up to 2 to 3 hours and will not cause any pain to your child.

If the child’s Tympanometry results are NOT normal, he/she will be referred to an Ear Nose Throat specialist doctor to examine if he/she is suffering from any ear disease. If the doctor identifies ear disease, the doctor will recommend standard of care medical or surgical care, as necessary. Following treatment of ear disease, the child will be asked to repeat a diagnostic ABR hearing test to check if he/she still has any hearing loss.
The diagnostic ABR hearing test is standard of care and will reveal whether the child has a nerve-related hearing loss that occurs due to problems of the inner ear.

**Results**

The sample consists of 8144 infants, including 4812 (59%) males and 3332 (41%) females (Appendix Table 1). The distribution of the sample of population is summarized in Appendix Table 2. Out of the 8144 tested infants, 1507 (14.6%) infants did not pass the 1st OAE screening test, out of those, 498 (33%) did not come for retesting and 477 (31.6%) failed the retest (Table 1). In 2009 there was a significant increase in the total number of patients screened (from 829 in 2008 to 1694) (Appendix Table 1). This was due to opening the service to other maternity facilities for Normal newborns. In this case the rate of hearing loss is expected to decrease with the inclusion of babies without risk factors for hearing loss. Only 152 (31.9%) of the patients that failed retesting went to an audiologist (Table 2).

The audiologist evaluation revealed that 101 (66.4%) patients presented with mild-moderate or profound hearing loss according to the Bureau International of Audiophonologie (9), while 44 (28.9%) patients presented with Otitis media and the rest 7 (4.6%) are normal.

Chart review of the affected patients revealed that high proportion of infants failed the screening test having one of the following diseases: jaundice, sepsis, prematurity, lung disease (Appendix Tables 3 and 4).

Besides summarizing the outcome in these 6 years, and comparing the data between the two groups (2006-2008) and (2009-2011) show that the percent of failure in the first test reduced significantly (from 36.2% to 14.1% with P-value <0.001). That is because the first group is mostly sick infants but the second group includes a lot of healthy infants.

Also, the overall percent of hearing loss is statistically significant (from 2.26% to 1%.
with P-value <0.001).

The slight increase of the percent of patients going to the audiologist (from 30.6% to 32.7%) shows more commitment by the family to follow all the steps.

Because it was large increased in the percent of hearing loss discovered to the patients (from 58.3% to 71.7%) showing that normal newborns were, also, affected by hearing loss and demonstrating the importance in providing an universal newborn hearing screening (Table 3).

The overall unadjusted percent of hearing loss in all 6 years (2006-2011) is 1.24% (101/8144). A percentage of 10 (101/1009) of those who follow up have hearing loss. If we assume that the same percent of those who did not follow up will have hearing loss, this will add 0.1 x (1507 – 1009) = 50 patients to the hearing loss result. The adjusted hearing loss total number in the 6 years will be 151, and the adjusted overall percent of hearing loss will be 1.85% (151/8144).

As the prevalence of hearing loss is high, and there was only one neonatal screening location in the entire area, it is necessary to implement the Early Hearing detection and intervention action center guidelines and extend the program to other local health facilities.

**Discussion and Conclusions**

The study has enabled us to identify the type of infants with positive screening audiology. Not only diseases like jaundice, infections, prematurity and lung disease are risk factors for hearing loss, but also the normal newborns are affected by hearing loss.

Moreover, the study showed high referral rate (14.6%) in the first screening, especially, among premature infants suffering from infections and jaundice. The study also highlighted the lack of follow up as 33% did not come back for the retest. Unfortunately this
high number of infants not returning for the confirmatory test can bias severely the index of hearing loss and this is a limitation of the study.

In addition, only 31.9% of the infants tested positive for the second screening underwent a specialist visit. This may be due to the lack of parents’ knowledge about the importance of continuing the follow-up. Therefore, it is highly recommended to improve the educational activities for families and collaboration among the staff and the specialist audiologist. The survey demonstrated a high prevalence of Hearing loss as 66.4% of the infants visited the audiologist were affected by mild-moderate or profound hearing loss.

We can identify some significant limitations in this study. We used different testers over several years. We started training staff nurse and practical nurse, then after 2 years another practical nurse and then another one dealing daily with the service.

Also, we started with OZ screen machine and then with Otoacoustic machine Otoport wireless, using so 2 machines: one for Inpatients and 1 for Outpatients. We used two different machines and, even if the calibration data were the same, using the same type of study TEOAE, the results can be slightly different. Our patients in the first 3 years were with high risk of hearing loss as inpatients and staying for more than 48 hours of hospitalization. Only in 2009 we opened the service for Normal newborns.

Also, most of the participants were from 2 districts only: Bethlehem and Hebron limiting significantly the study.

Given the high prevalence of the phenomenon, it was recommended to disseminate this information and extend research in all branches of Neonatology, improve the Intensive Care Units, and keep annually updated database. Also, it is important to create a link network and active cooperation between the audiology service and the University of Bethlehem dedicated to the research of genetic diseases including deafness in children. Connexin 26 (GJB2) mutations lead to hearing loss, despite the fact that at least 50 other genes are also
associated with hearing loss. The entire coding region of connexin26 was sequenced in 75 hearing impaired children and adults in Israel in order to determine the percentage of hearing loss attributed to Connexin 26 and the types of mutations in this population. Almost 39% of all persons tested harbored GJB2 mutations.\textsuperscript{7} Listening, dialogue, security and proximity are key elements that all the staff must ensure in order to support the family to accept the reality and to encourage the family to continue their treatment.

A study done in 2006, investigated the prevalence of congenital and early-onset hearing loss, and the influence of the known risk factors for hearing loss on infants in Jordan and Israel shows that the prevalence and severity of hearing loss amongst Jordanian infants (1.37%) is remarkably higher compared to the Israeli infants (0.48%).\textsuperscript{8} The study included 17,000 infants from both countries, infants with and without risk factors for hearing loss. The hearing screening protocol included distortion product OAE, followed in case of repeated OAE referral or high-risk infant by diagnostic ABR. The results, also, indicate that the overall prevalence of bilateral sensorineural hearing loss was seven times more in the Jordanian infants, 18 times in non-risk, and three times in the high-risk infants relative to the Israeli infants. Risk factors including family history, hyperbilirubinemia, bacterial meningitis, and associated syndromes were more prevalent amongst Jordanian infants. This unique study underscored the importance of sharing and exchanging information to create empirical data to guide health-care providers in adapting protocols to the local constraints in developing countries. In our study we demonstrated that the overall percent of hearing loss is very high and a prevention program and family education is highly recommended, in order to increase awareness of this significant problem, to guarantee a correct and complete follow-up and to provide a more accurate estimate infant hearing loss in Palestine.
References


Table 1. Distribution of the first screening results and the follow-up results across the years.

<table>
<thead>
<tr>
<th>Years</th>
<th>Failed 1st screening %</th>
<th>Failed 2nd screening %</th>
<th>No retest %</th>
</tr>
</thead>
<tbody>
<tr>
<td>2006-2007</td>
<td>33.6 (n=242)</td>
<td>34.7 (n= 84)</td>
<td>34.7 (n=84)</td>
</tr>
<tr>
<td>2008</td>
<td>40.7 (n=338)</td>
<td>33.1 (n=112)</td>
<td>31 (n=109)</td>
</tr>
<tr>
<td>2009</td>
<td>29.1 (n=493)</td>
<td>42.2 (n=208)</td>
<td>13.8 (n=68)</td>
</tr>
<tr>
<td>2010</td>
<td>10.7 (n=231)</td>
<td>16.5 (n=38)</td>
<td>58 (n=134)</td>
</tr>
<tr>
<td>2011</td>
<td>7.3 (n=203)</td>
<td>17.2 (n=35)</td>
<td>50.8 (n=103)</td>
</tr>
<tr>
<td>Total number</td>
<td>1507</td>
<td>477</td>
<td>498</td>
</tr>
</tbody>
</table>

Table 2. Distribution of retest and hearing loss results across the years.

<table>
<thead>
<tr>
<th>Years</th>
<th>Failed 2nd screening %</th>
<th>Seen by audiologist %</th>
<th>Positive hearing loss %</th>
</tr>
</thead>
<tbody>
<tr>
<td>2006-2007</td>
<td>34.7 (n=84)</td>
<td>9.5 (n=8)</td>
<td>75 (n= 6)</td>
</tr>
<tr>
<td>2008</td>
<td>33.1 (n=112)</td>
<td>46.4 (n=52)</td>
<td>55.8 (n=29)</td>
</tr>
<tr>
<td>2009</td>
<td>42.2 (n=208)</td>
<td>22.6 (n=47)</td>
<td>72.3 (n=34)</td>
</tr>
<tr>
<td>2010</td>
<td>16.5 (n=38)</td>
<td>65.8 (n=25)</td>
<td>56 (n=14)</td>
</tr>
<tr>
<td>2011</td>
<td>17.2 (n=35)</td>
<td>57.1 (n=20)</td>
<td>90 (n=18)</td>
</tr>
<tr>
<td>Total number</td>
<td>477</td>
<td>152</td>
<td>101</td>
</tr>
</tbody>
</table>

Table 3. Comparison data among the years.

<table>
<thead>
<tr>
<th>Items</th>
<th>2006-2008</th>
<th>2009-2011</th>
<th>P-value</th>
<th>Total number</th>
</tr>
</thead>
<tbody>
<tr>
<td>Number of infants</td>
<td>1550</td>
<td>6594</td>
<td>-</td>
<td>8144</td>
</tr>
<tr>
<td>Percent not passing 1st test</td>
<td>36.2% (n=580)</td>
<td>14.1% (n=927)</td>
<td>&lt;0.001</td>
<td>1507</td>
</tr>
<tr>
<td>Percent of follow-up</td>
<td>66.7% (n=387)</td>
<td>67.1% (n=622)</td>
<td>0.88</td>
<td>1009</td>
</tr>
<tr>
<td>Percent did not pass 2nd test</td>
<td>50.6% (n=196)</td>
<td>45.2% (n=281)</td>
<td>0.09</td>
<td>477</td>
</tr>
<tr>
<td>Percent went to the audiologist</td>
<td>30.6% (n=60)</td>
<td>32.7% (n=92)</td>
<td>0.62</td>
<td>152</td>
</tr>
<tr>
<td>Percent of hearing loss</td>
<td>58.3% (n=35)</td>
<td>71.7% (n=66)</td>
<td>0.11</td>
<td>101</td>
</tr>
<tr>
<td>Overall percent of hearing loss</td>
<td>2.26% (35/1550)</td>
<td>1% (66/6594)</td>
<td>&lt;0.001</td>
<td>-</td>
</tr>
</tbody>
</table>