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

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Abstract

This study describes the epidemiology of infants' hearing loss (IHL) among patients under 3 months of age at Caritas Baby Hospital, the only pediatric hospital in Palestine. It was aimed to demonstrate that IHL is a major health problem in Palestine and to assess the first available data of the newborn hearing screening program conducted 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. As to their origin, 72% (5886) came from the Bethlehem district, 25% (2044) from the Hebron district, while 3% (214) from the other Palestinian districts (Jericho, Ramallah, Nablus, Jenin and Jerusalem). The transient evoked otoacoustic emissions (TEOAEs) and the automated auditory brainstem response were used according to the manufacturer guidelines. The results were interpreted according to the indications of 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%) did not pass the 1st test, 477 (32.8%) of these 1507 infants failed retesting, while 498 (33%) patients were lost to follow-up. Only 152 (31.9%) patients that failed retesting went to an audiologist. The audiologist evaluation revealed that 101 (66.4%) patients presented with a mild-moderate or profound hearing loss according to the *Bureau International of Audiophonologie* standards, 44 (28.9%) patients had otitis media, whereas 7 cases (4.7%) had no hearing disorders. The overall unadjusted percentage of hearing loss was 1.24%, and the adjusted overall percentage was 1.85%. The 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 deserves immediate attention on the part of the Palestinian government. Meanwhile, Caritas Baby Hospital undertook to set up a newborn hearing screening unit utilizing the TEOAE method.

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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, who wish to transform Caritas Baby Hospital into a center of excellence, ensuring patient and staff safety and quality across all activities.

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Introduction

In August 2006, Caritas Baby Hospital (CBH) welcomed a proposal from the medical audiologist to implement a universal neonatal hearing screening program. 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 a high prevalence of severe hearing loss in the Palestinian population during a study performed in 2006.[2] In that study, we concluded that a deafness screening program should be extended to all newborns, while parents should be informed about the potential problems their children might face because of hearing loss. The screening program started at the Department of Neonatology on the basis of the otoacoustic emissions (OAEs) method. Hereditary deafness affects

approximately 8‰ of the Palestinian population, and up to 3-15% of these individuals live in isolated villages.[1] Hereditary deafness has several sociological implications, including isolation and abandonment for those who are deaf, and lack of educational strategies for those who lose their hearing in their 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 gene 26 (*GJB2*) 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 in poorer socio-economic conditions its prevalence appears to be twice as high compared to other developed countries (1.3-2.7 per 1000).[3]

The American Joint Committee on Infant Hearing Screening established a list of risk factors and created a register for babies at risk for deafness.[4] This protocol identifies only 50% of the children with hearing impairment. Infants with no health problems during pregnancy or at birth may have a normal test result, although they are actually suffering from deafness. Therefore it was suggested by the American Joint Committee of Infant Hearing Screening to apply the neonatal screening program to the all babies under three months of age and not only to those at high risk.

OAEs are considered an ideal tool for universal screening by several institutions as the American Academy of Pediatrics,[4] the National Institutes of Health, and the European Consensus Development Conference on Neonatal Hearing Screening. The test requires 3-5 min per infant and, because it is easy to perform, it can be done after the first hours of the child's life. The sensitivity of OAEs is slightly less than 100% compared to 95% of the auditory brainstem response (ABR).[5] The limitation of the OAE testing is that it is a peripheral testing and does not evaluate the full functionality of the ears. Childhood deafness and a potential delay in language acquisition can lead to problems in terms of cognitive, emotional-affective and social development.[5]

In this manuscript, we report the epidemiology of childhood diseases that lead to deafness in Palestinian patients under the age of 3 months across the West Bank. Also, the manuscript highlights that infant hearing loss is a major health problem in Palestine.

Materials and Methods

Study location and study period

Caritas Baby Hospital (CBH) has 82 beds and is located in the district of Bethlehem, Palestine. Patients seen in the various hospital departments come from all over Palestine, in particular, from the Southern districts of Bethlehem and Hebron. A total of 8144 patients were included in the study, 4812 (59%) were males and 3332 (41%) were females; they were either admitted to or visited in the outpatient clinics between September 25, 2006 and December 31, 2011. The patient distribution by district was as follows: 72% (5886) of the infants were from the Bethlehem district, 25% (2044) from the Hebron district, while 3% (214) were from the other Palestinian districts (Jericho, Ramallah, Nablus, Jenin and Jerusalem).

In 2009, the service was expanded to include *normal newborn babies*, particularly from the districts of Bethlehem and Hebron.

Data collection

This study was a retrospective study on patients referred to CBH Outpatient clinics for hip ultrasound, hearing test, or follow-up. This study was approved by the CBH Executive Committee and the Medical Research committee and it was supported by the local staff. The study was carried out utilizing the data from children who underwent the hearing 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 result of the retest, medical diagnosis for those who failed the 1st test, audiologist's report.

Disease groups

Given the variety of medical diagnosis among the patients, it was necessary to identify the most common diseases, which were the following seven: jaundice, prematurity, infection, lung disease, cardiac disease, gastrointestinal disease and others.

Data analysis

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

Otoacoustic emissions test

This test consists of low-intensity sounds emitted by the outer hair cells of the cochlea 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 is inserted just inside both ear canals of the child. These foam tips are connected to a handheld device that does the recording and gives a pass/fail result. The child will not experience any pain since the probe will be placed just inside the ears. This test requires no longer than 10 min.

The sensitivity of OAEs is slightly less than 100% compared to 95% of ABR. The limitation of this test is that it tests the periphery only without assessing the acoustic full functionality of the street noise.[1] The transient evoked otoacoustic emissions (TEOAEs) 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 in speech and its etiology can unknown, genetic or due to jaundice, anoxia or prematurity.

Stem auditory evoked potentials (auditory brainstem response)

It is a method for hearing screening of infants at risk. It has the highest sensitivity and reliability and a very low number of false negatives and false positives.

The examination is performed during spontaneous sleep of the newborn. In case of positivity, another check is performed before the infant is discharged from hospital. The advantages of this method are the accuracy and the competence of the examiners.[1]

The use of ABR screening procedures has been developed to detect automatically the presence or absence of response, and to reduce the cost and the time of the examination.[6] The automated ABR is a more sophisticated form of hearing test than TEOAE and involves the use of 3 electrodes, which are placed 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 the probe are attached to a handheld device, which does the reading. This test takes no longer than 20 min. However, it may be much faster (less than 10 min), if the child is very calm. Also this test is painless for the child.

Tympanometry

Tympanometry is a test that is performed to detect the presence of

any fluid in the ear, which can cause hearing loss. It is done by inserting tiny probes (foam tips) just inside the ear canal that send a pressure signal toward the ear drum and receive the signal back. An attached handheld device records the outcome of the test. It is painless for the child and can require no longer than 10 min.

We started using tympanometry if the patient failed either the TEOAE or the Automated ABR tests.

Study procedures

Within the neonatal hearing screening program, the child is assessed to identify risk factors for hearing loss. A risk factor questionnaire is administered to the parents and can take up to 10 minutes to complete. This questionnaire is used to determine whether the child has any existing risk factors associated with hearing loss. The questionnaire includes questions related to: i) illness or condition requiring admission to and a stay of 48 h or longer at a Neonatal Intensive Care Unit (NICU); ii) stigmata or other findings of a syndrome known to include sensorial neural hearing loss or conductive hearing loss; iii) family history of permanent childhood hearing loss; iv) craniofacial abnormalities including morphological abnormalities of the pinna or ear canal; and v) evidence of *in utero* infections such as cytomegalovirus, herpes, toxoplasmosis, or rubella.

If no risk factors are identified, the child 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, it means that, at the time of the 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 has risk factors for hearing loss, he/she will be asked to undergo another hearing test called Automated ABR. If the child passes the Automated ABR in both ears, he/she is sent home.

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

If the child's tympanometry test results are normal, he/she will be asked to see the closest Audiologist to his/her town to undergo a diagnostic ABR test. This is a more sophisticated and time-consuming diagnostic hearing test than the previous ones, *i.e.* TOAE and Automated ABR. It is very similar to the Automated ABR, but it is more complete and shows exactly how much and what kind of hearing loss the child has. This diagnostic hearing test can require up to 2-3 h and will not cause any pain to the child.

If the child's tympanometry results are not normal, he/she will be referred to an Ear Nose Throat specialist to check if he/she is suffering from any ear disease. If the doctor identifies an ear disease, he/she will recommend the most appropriate standard of medical or surgical care for the child. After the treatment of the 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 a standard of care and reveals whether the child has a nerve-related hearing loss that may occur due to problems of the inner ear.

Results

The sample consisted of 8144 infants, including 4812 (59%) males and 3332 (41%) females (Appendix Table 1). The distribution of the sample is summarized in Appendix Table 2. Out of the 8144 tested infants, 1507 (14.6%) did not pass the 1st OAE screening test and, out of them, 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 happened because this service was made available to other maternity facilities for normal newborns. As a result, the rate of hearing loss was expected to decrease with the inclusion of babies without risk factors for hearing loss. Only 152 (31.9%) patients that failed retesting saw to an audiologist (Table 2).

The audiologist's evaluation revealed that 101 (66.4%) patients presented with mild-moderate or profound hearing loss according to the *Bureau International of Audiophonologie* standards, 44 (28.9%) patients had otitis media, while the remaining 7 (4.6%) were normal.

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

Besides summarizing the outcome in these 6 years, the comparison of data between the first group (2006-2008) and the second group (2009-2011) showed a significant reduction in the percentage of chil-

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

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

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

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

dren who failed the first test (from 36.2% to 14.1% with a P-value <0.001), because the first group mainly included sick infants, whereas the second also comprised many healthy infants.

In addition, the overall hearing loss rate was statistically significant (from 2.26% to 1% with a P-value <0.001).

The slight increase in the patients who went to see the audiologist (from 30.6% to 32.7%) showed a greater commitment on the part of the families who followed all the foreseen steps.

The significant increase in the percentage of hearing loss cases among the patients (from 58.3% to 71.7%) showed that normal newborns were, also, affected by hearing loss and demonstrated the importance of providing a universal newborn hearing screening program (Table 3).

The overall unadjusted hearing loss rate throughout the 6 years (2006-2011) was 1.24% (101/8144). If one considers that 10% (101/1009) of the children at follow up had a hearing loss and that the same percentage of those who were not followed up had also a hearing loss, the final hearing loss rate must be increased by 0.1 x (1507-1009), *i.e.* 50 patients. The total adjusted number of patients with hearing over the 6 years was 151, while the total adjusted hearing loss rate was 1.85% (151/8144).

As the prevalence of hearing loss was high, and there was only one neonatal screening unit in the entire area, it was necessary to implement guidelines for early hearing loss detection and intervention centers and to extend the program to other local healthcare facilities.

Discussion and Conclusions

The study enabled us to identify the type of infants with positive screening audiology and to demonstrate that risk factors for hearing loss are not only diseases like jaundice, infections, prematurity and lung disease, because also normal newborns can be affected by this disorder.

Moreover, the study showed a 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% of the patients did not return for a retest. Unfortunately this high proportion of infants can bias severely the final hearing loss rate and this is a limitation of the study.

In addition, only 31.9% of the infants who tested positive at the second screening underwent a specialist visit. This may be due to the lack of parents' awareness about the importance of the follow-up. Therefore, it is highly recommended to improve the educational activities for families and the collaboration between the staff and the audiologist. The survey demonstrated that a high prevalence of children with hearing loss (66.4%) who were taken to the audiologist were affected by a mild-moderate or profound hearing loss.

This study has some significant limitations. We used different testers over several years. We started training the staff nurse and the practical nurse, then after 2 years another practical nurse was involved and subsequently another one in charge of the daily routine.

Also, we started with a OZ screen machine and later we used a wireless Otoport otoacoustic machine, therefore we used 2 machines, namely one for inpatients and one for outpatients. We used two different machines and, even if both the calibration data and the TEOAE study were the same, the results can be slightly different. Furthermore the patients who were tested during the first 3 years were inpatients at high risk of hearing loss and were hospitalized for longer than 48 h. We opened the service to normal infants only in 2009.

Also, most of the participants were from 2 districts only (Bethlehem and Hebron), which was a significant limitation for this study.

Given the high prevalence of this disorder, it was recommended to disseminate this information and extend research in all branches of neonatology, improve the intensive care units, and keep a database to be updated annually. It was also highlighted that it is important to create a network and to foster proactive cooperation between the audiology service and the University of Bethlehem that conducts research on genetic diseases including deafness in children. It is known that connexin 26 (GJB2) mutations lead to hearing loss, although at least 50 more genes are also associated with this disorder. The entire coding region of connexin 26 was sequenced in 75 hearing-impaired children and adults in Israel in order to determine the percentage of hearing loss cases ascribable to connexin 26 and the mutations in this population. Almost 39% of all persons tested had GJB2 mutations.[7] All the staff must be trained to listen, reassure, discuss and support the families, so that they can accept this disorder and be encouraged to continue the treatment.

A study conducted 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. It showed that the prevalence and severity of hearing loss amongst Jordanian infants (1.37%) was remarkably higher compared to the Israeli infants (0.48%).[8] The study included 17,000 infants with and without risk factors for hearing loss from both countries. The hearing screening protocol included distortion product OAE, followed in case of repeated OAE referral or high-risk infants by diagnostic ABR. The results, also, indicated that the overall prevalence of bilateral sensorineural hearing loss was seven times greater in Jordanian infants, 18 times greater than in the population with no risks and three times greater than in the high-risk group compared to Israeli infants. The risk factors, which included family history, hyperbilirubinemia, bacterial meningitis, and associated syndromes, were more prevalent among Jordanian infants. This unique study underscored the importance of sharing and exchanging information in order to gather empirical data to guide healthcare providers in adapting protocols to the local constraints in the developing countries. In our study we also demonstrated that the overall hearing loss rate was very high, therefore a prevention program and family education were highly recommended, in order to increase the aware-

Table 3. Comparison data among the years.

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

ness around this significant problem, to ensure adequate and complete follow-up and to provide a more accurate estimate of the infant hearing loss rate in Palestine.

References

1. Kanaan MWT, Rayan AA, et al. The genetic bases of hearing loss in the Palestinian population. Bethlehem: Eftah Shool for Deaf; 2002. Available from: http://www.bethlehem.edu/researches Accessed: 20/07/2007.
2. Corradin L, Fanton E. [A survey and childhood deafness in a neonatology unit of Caritas Baby Hospital in Bethlehem]. Assist Inferm Ric 2008;27:85-90. [In Italian].
3. Epidemiologia della sordità infantile. Available from: http://www.sestocircoloss.it/servizi/formazione/materiale_corso_succ_integra_scol/sordita'20infantile.pdf Accessed: 09/07/2007.
4. Wroblewska-Seniuk K, Chojnacka K, Pucher B, et al. The results of newborn hearing screening by means of transient evoked otoacoustic emissions. Int J Pediatr Otorhinolaryngol 2005;69:1351-7.
5. Ambrosetti U, Del Bo L. Apparecchi acustici, acufeni, orecchio, sordità infantile. Available from: http://www.sordita.it /load.php?page=sorditainfantile Accessed: 10/07/2007.
6. Mason JA, Herrmann KR. Universal infant hearing screening by automated auditory brainstem response measurement. Pediatrics 1998;101:221-8.
7. Sobe T, Vreugde S, Shahin H, et al. The prevalence and expression of inherited connexin 26 mutations associated with nonsyndromic hearing loss in the Israeli population. Hum Genet 2000;106:50-7.
8. Attias J, Al-Masri M, Abukader L, et al. The prevalence of congenital and early-onset hearing loss in Jordanian and Israeli infants. Int J Audiol 2006;45:528-36.