Original Article

Neonatal Screening for Congenital Hearing Loss in the North of Jordan; Findings and Implications

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Abstract

Background: Congenital hearing loss is one of the important illnesses that affect newborns. Early diagnosis and treatment are a challenge for medical authorities in developing countries to improve children's functional, intellectual, emotional, and social abilities. We aimed to study the prevalence of congenital hearing loss in northern Jordan community and identify factors that could affect hearing screening protocol. **Methods:** Prospective cross-sectional study of 1595 infants born in our hospital underwent hearing screening tests. Totally, 104 were tested in NICU and the rest examined in the nursery room using Otoacoustic emission (OAE) test as a primary testing tool. The patients were followed in the three hearing screening phases. Factors affecting screening results were studied and analyzed. **Results:** The total number of newborns who didn't pass the first OAE test in one or both ears were 90 (5.6%); 69 from the nursery group and 21 from the NICU group. In the 2nd screening phase 21 (23.3%) didn't attend the appointment. Sixty-four passed the second screening OAE test. Five newborns (5.6%) had a second refer result in one or both ears and referred for a diagnostic ABR test. Three infants passed the test and two found to have bilateral hearing loss. **Conclusions:** Hearing screening test is conducted via a 3-phases-protocol. OAE is used in the first two phases and ABR in the third phase. Hearing results is significantly affected for infants admitted to NICU. The following factors increase OAE fail response: mechanical ventilation for more than 5 days, Hyperbilirubinemia, associated congenital anomalies. Mode of delivery doesn't have statistical significance on hearing screening results.

Keywords: *Deafness, hearing, hearing test, otoacoustic emission, screening*

Introduction

Congenital hearing loss has started to become one of the foremost medical problems to gain attention in the last decades due to the advancement in management options which improves children's functional, intellectual, emotional, and social abilities. Congenital hearing loss is one of the major birth abnormalities in newborns where the incidence is estimated to be 1-3 per 1000 live births in well-baby nursery newborns and nearly 2-4 per 100 infants in neonatal intensive care unit (NICU) babies.[1-6]

Congenital hearing loss can be classified according to the etiology, half of the cases are suggested to be genetic in origin, and 25% are acquired (perinatal infections e.g., torch, hyperbilirubinemia, birth complications, etc.) leaving the rest for idiopathic reasons.[7,8] Risk factors for hearing loss include a previous family history of hearing loss, craniofacial anomalies, complex congenital anomalies associated with congenital hearing loss, congenital infections (TORCH infections), low birth weight (<1500 g), prematurity (<33 weeks), hyperbilirubinemia, ototoxic medications, bacterial meningitis, low APGAR score in 1 min (0-4), low APGAR score in 5 minutes (0-6), mechanical ventilation for 5 days or more and NICU admission for more than 7 days.[9]

Neonatal hearing loss can be suspected by parents or caregivers by them noticing the absence of the baby's usual reflex to loud noise, which may be late in some families leading to critical effects on the development of language and the development of social and intellectual abilities.[10] Hence, early identification of deafness can avoid these defects, that why a Universal hearing screening program is important to catch these cases.

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Hearing screening can be implemented on different levels according to the Joint Committee on infant hearing 2000.[11] First level screening using otoacoustic emissions (OAE) testing and passing a questionnaire that aims to identify described risk factors in all newborns before discharge. In the second screening level, infants with "Refer" test results in one or two ears or having high-risk factors should be referred to Auditory Brain stem Response (ABR) Testing in specialized audiology centers. The third screening level is transferring infants with definitive hearing loss to either auditory or otology centers for hearing aids trial or surgical management.

In this study, we tried to get initial information about the prevalence of congenital hearing loss among newborns in our community by screening newborns in our hospital's nursery and Neonatal Intensive Care Unit (NICU). Moreover, as a secondary goal, we are trying to re-evaluate our screening protocol to decrease the failure rate among the OAE test to reduce repeated testing and to minimize costs and parents' anxiety.

Methods

This prospective cross-sectional study took place over one year, between the 1st of September 2017 and the 31st of July 2018, in King Abdullah University Hospital a tertiary referral hospital in north of Jordan. The study was approved by the Institutional Review Board (IRB approval number 237-2017). The screening process took place in both units the nursery and the NICU. All newborns and their mother's medical records were reviewed, and history was taken from the parents to check for possible hearing loss risk factors. The following data were recorded:

1. Family history of hearing loss and parents consanguinity.
2. In-utero infection, such as cytomegalovirus, rubella, syphilis, herpes, and toxoplasmosis
3. Craniofacial anomalies including those with morphological abnormalities of the pinna and ear canal.
4. Birth weight less than 1500 g (1.5 kg).
5. Hyperbilirubinemia not requiring exchange transfusion.
6. Ototoxic medications during pregnancy and during neonatal period.
7. APGAR scores of 0–4 at 1 min or 0–6 at 5 min.
8. Mechanical ventilation lasting 5 days or longer.
9. Stigmata or other findings associated with a syndrome known to include a sensorineural hearing loss.
10. Prematurity (gestational age <37 weeks).
11. Suffering from bacterial meningitis.

The hearing screening was obtained using both transients evoked otoacoustic emission (TEOAE) and distortion product otoacoustic emissions (DPOAE). The screening was conducted within the first 48 hours after delivery for healthy newborns and just before discharge for newborns in the NICU. The examination rooms were the offices inside the nursery or NICU which are not perfectly soundproofed but away from the noise.

Phases of hearing screening

Three phases of the screening were implemented within three months of delivery. During the first phase, newborns were screened before discharge, where both ears were screened separately. Results of screening were either "Pass" or Refer". All babies who had "refer" results were given an appointment for a second OAE test in our audiology department 2-4 weeks after delivery depending on the mother's gynecology clinic visit or the infants' pediatrics clinic visit to increase compliance. If the baby failed the second OAE test, a diagnostic Auditory Brain stem Response (ABR) test appointment is given.

Results

The total number of infants enrolled in the screening process was 1595, 836 (52.4%) males and 759 (47.6%). Totally, 1491 (779 male, 712 female) newborns were examined in the nursery unit and 104 (57 male, 47 female) were NICU babies. The two groups were studied separately and then compared Table 1.

The total number of newborns who didn't pass the first OAE both (TEOAE) and (DPOAE) test in one or both ears was 90 (5.6%); 69 from the nursery group and 21 from the NICU group [Table 2]. Totally, 64 of the total 90 newborns with referring results (71.1%) had a repeat OAE and passed the second screening level, 5 newborns (5.6%) had another refer result in one or both ears and subsequently were being referred for a diagnostic ABR test. Totally, 2 of the 5 failed the ABR testing and were diagnosed with

Table 1: Description of infants included in the study

	Nursery		NICU		Total	
	n	%	*n*	%	*n*	%
Gender						
Male	779	52.2	57	54.8	836	52.4
Female	712	47.8	47	45.2	759	47.6
Gestational Age (Weeks)						
<37	152	10.2	54	51.9	206	12.9
>=37	1339	89.8	50	48.1	1389	87.1
Mode of delivery						
Caesarian	878	58.9	73	70.2	951	59.6
Vaginal	613	41.1	31	29.8	644	40.4
Smoking during pregnancy						
Yes	40	2.7	0	0	40	2.5
No	1450	97.3	104	100	1555	97.5
Congenital abnormalities						
Yes	67	4.5	10	9.6	77	4.8
No	1424	95.5	94	90.4	1518	95.2
Family history of hearing loss						
Yes	60	4	0	0	60	4
No	1431	96	104	100	1535	96
Auricular deformities						
Yes	10	0.7	0	0	10	0.6
No	1481	99.3	104	100	1585	99.4

n: number of infants, NICU: Neonatal Intensive Care Unit

Table 2: Screening results per screening phase for the 1595 infants enrolled in the study

	Nursery N		NICU N		Total
	TEOAE	DPOAE	TEOAE	DPOAE	*n*
First phase					
Total screened	1491	1491	104	104	1595
Pass	1422	1422	83	83	1505
Refer	69	69	21	21	90
Second phase					
Total screened	54	54	15	15	69
Pass	52	52	12	12	64
Refer	2	2	3	3	5
Not attend	15	15	6	6	21

n: Number of infants, NICU: Neonatal Intensive Care Unit. TEOAE: Transients evoked otoacoustic emission. DPOAE: Distortion product otoacoustic emissions

bilateral hearing loss. Twenty-one of the total 90 newborns with referring results (23.3%) didn't attend the appointment one of them did not attend because he passed away, others did not attend due to multiple factors including, change in the contact phone number (7 newborns) and non-convinced with test results due to negative family history of hearing problems (13 newborns).

Among the nursery babies [Table 2], 1422 passed the first screening level in both ears [Had a "pass" OAE result in both ears] and no further follow up was needed. Totally, 69 newborns failed the first screening level [Had a "refer" OAE result in one or both ears]; 46 had a "refer" OAE result in both ears, 10 had a "refer" OAE results in the right ear only and a "pass" in the left ear and 13 had a "refer" OAE results in the left ear only and a "pass" result in the right ear. All 69 newborns were referred for the second screening level. Newborns who had a "refer" OAE result in one or both ears were given an appointment to repeat the OAE test in our audiology department 2-4 weeks after delivery and were included in the second screening level. Fifteen didn't attend the appointment (one of them because he passed away) while the remaining had the second screening level; 52 passed, 2 failed and were referred for the third screening level with a diagnostic ABR test. One was found to have bilateral hearing loss.

A total of 104 NICU babies [Table 2] were included in the first screening level of the study. Totally, 83 newborns passed the first screening level and didn't require any further follow-up hearing screening. A total of, 21 NICU babies failed the first screening level with a "refer" OAE results in one or both ears; 16 had a "refer" result in both ears, 3 had a "refer" result in the right ear only, and 2 had a "refer" result in the left ear only. The 21 NICU newborns who had a "refer" OAE result in one or both ears were referred for the second screening level; 6 didn't attend the second OAE appointment, 12 passed the second screening level, 3 failed the second screening level and were referred for diagnostic ABR testing; one out of the three was found to have hearing loss.

According to our results, the prevalence of SNHL among north X infants was (0.12%) taking into consideration all infants who underwent the OAE test excluding those who did not follow the regular screening protocol. (21 infants from total 1595 infants).

The prevalence of infants who failed the first OAE was (5.6% in total). Among infants who were admitted to the nursery unit, the prevalence was 4.6% while for those admitted to the NICU unit, the prevalence was 20.1%. The difference was obvious with *P* value of less than 0.0001 which is considered as marked statistical significance.

Regarding the infants who were admitted to the NICU unit, 83 infants pass the first screening test while 21 infants did not pass the test. Regarding these results there was a statistically significant correlation between mechanical ventilation for more than 5 days (*P*-value 0.03), Hyperbilirubinemia (*P*-value 0.01), infants with other congenital anomalies (*P*-value 0.02), and the failure of the first screening test by the OAE [Table 3].

Discussion

It is widely agreeable that the screening of congenital hearing loss is critical, and the implementation of a comprehensive screening program for all neonates is more beneficial than screening just those who admitted to the NICU unit.[12] One of the crucial points is that early detection and treatment of neonates with congenital hearing loss has a great value[13,14] as hearing plays a substantial role in developing speech and language, cognitive development and socialization.[15] Delayed identification of congenital hearing loss can gravely influence the future life of the child with subsequent significant disability and related huge social expenditures.[16,17] One of the most important and popular screening tests is the Otoacoustic emissions that considered to be the most acceptable tests for a hearing screening. However, it has some limitations with false results in few situations that require more reliable testing or retesting. On the other hand, Brainstem Evoked Response Audiometry (BERA) is highly reliable, but it is more expensive as compared to TEOAE.

In our study, the prevalence of congenital SNHL among infants was 0.12% which is almost similar to the global rate of neonatal hearing impairment which is between 0.1% and 0.3%.[18] the prevalence of congenital SNHL among nursery group was 0.07% which is lower than the prevalence among other reported studies in different countries which ranged from 0.09 to 0.13%.[19,20] On the other hand, the prevalence of congenital SNHL was 1% among neonates with high-risk groups. Connolly *et al*. found that 1 of every 811 infants had hearing loss among those with low-risk groups compared to 1 of every 75 infants with high-risk groups.[21] Erenberg *et al*. stated that the prevalence

Table 3: Characteristics of the 104 newborns that were admitted to the NICU unit

	n		
	NICU Pass screening	**NICU Fail screening**	**Total**
Sex			
Male	42	15	57
Female	41	6	47
Congenital abnormalities			
Yes	5	5	10
No	78	16	94
Family history of hearing loss			
Yes	0	0	0
No	83	21	104
Auricular deformities			
Yes	0	0	0
No	83	21	104
Apgar Score 0-4 at 1 min			
Yes	11	2	13
No	72	19	91
Apgar Score 0-6 at 5 min			
Yes	4	1	5
No	79	20	99
Mechanical ventilation lasting 5 days or longer			
Yes	13	8	21
No	70	13	83
Hyperbilirubinemia			
Yes	20	11	31
No	63	10	73

of bilateral hearing loss is approximately 1 to 3 per 1000 newborns in the well-baby nursery population and it approximately 2 to 4 per 1000 infants in the intensive care unit population.[22]

The newborns' hearing screening (NHS) schedule can be divided into one-stage and two-stage types depending on the number of screening tests performed before a diagnostic ABR. The guidelines recommend one-stage or two-stages NHS relying on the hospital system and protocols, where In a one-stage NHS, newborns who do not pass the first step screening are referred for a diagnostic ABR test without an extra hearing screening test while In a two-stages NHS, a second step screening is performed for newborns who do not pass the first step screening; only newborns who do not pass both steps are referred for a diagnostic ABR test.[23,24]

In our study, the prevalence of infants who failed the first screening phase of screening through OAE was 5.6% where it was 4.6% among infants who were admitted to the nursery unit and 20.1% among infants who were admitted to the NICU unit. The difference between both groups was statistically significant where the *P* value was less than 0.0001. With the second trial of OAE, the referral rate was 3.7% among infants who were admitted to the nursery unit and 20% among infants who were admitted to the NICU unit. Vohr *et al*. reported a referral rate 10% through the TEOAE-based program in Rhode Island.[25] Colella-Santos *et al*.[26] reported the referral rates in Brazil were 18.6% in the 1-stage test and 4.1% in the 2-stage test. For the 1-stage test, the referral rates were 9.2% in the Netherlands[26] and 4.9% in the USA.[19]

In this paper, we try to correlate the risk factors of NICU newborns among infants who were admitted to the NICU unit, 83 infants pass the first screening test while 21 infants did not pass the test with a referral rate 20.1%. Regarding these results there was a statistically significant correlation between mechanical ventilation for more than 5 days, Hyperbilirubinemia, infants with other congenital anomalies and the failure of the first screening test by the OAE; the *P* values were 0.03, 0.01, and 0.02, respectively. In our study, we did not find a statistical correlation between the mode of delivery and the failure rate of OAE. In contrast to our findings, Smolkin *et al*. found that differences were observed between the two delivery modes.[27]

Conclusions

Our study suggests that the prevalence of permanent SNHL in the north of Jordan is similar to the global rates for both nursery and NICU babies. Also, the study suggests a higher referral rate for babies from the NICU with failed hearing screening compared to the babies from nursery settings. Moreover, mechanical ventilation for more than 5 days, hyperbilirubinemia, infants with other congenital anomalies had higher possibility to fail the first screening test by the OAE test.

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Conflicts of interest

There are no conflicts of interest.

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