Newborn hearing screening: importance and characteristics of a high-risk register in a neonatal intensive care unit

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Background
Hearing loss is one of the most common congenital anomalies, occurring in approximately two to four infants per 1000. Children whose hearing loss is identified, and who receive appropriate intervention before 6 months of age, develop significantly better language ability than those who are identified later.

Objectives
To assess the failure rate of neonatal hearing screening in a high-risk register group of neonates and the relative importance of these risk factors in hearing impairment in the neonatal ICU (NICU) of the El-Mounira Children Hospital and identify the incidence of hearing loss.

Patients and methods
The study was carried out on 100 neonates in El-Mounira Children Hospital (NICU) and on 30 control neonates (15 boys and 15girls) chosen from Kasr El-Eini Maternity Hospital. All control cases aged 1–4 days had normal birth weight (2000–4000 g) and gestational age (35–40 weeks) and were delivered normally or by cesarean section without complications. The children in the study group were 3–23 days old (53 boys and 47 girls) and had birth weight ranging from 900 to 4000 g and gestational age ranging from 28 to 41 weeks. All the cases were screened for hearing loss using the transient evoked otoacoustic emission device (Echo-Screen), followed by a second-stage screening for those who failed the test with the transient evoked otoacoustic emission device. Those cases given a Refer were then made to undergo an Automated Auditory Brainstem Response test after 3–4 weeks.

Results
In the NICU group, 55% had hyperbilirubinemia, requiring exchange transfusion, 13% were of low birth weight (≤1500 g), and 13% were on mechanical ventilation. Other risk factors such as cranio-facial anomalies showed a combined effect. In the first screening phase, 71% were given a Pass response and 29% were given a Refer response. In the second screening phase, 28% were given a Pass, 31% were given a Refer, and 41% were dropouts, as they had passed their critical stage and had been discharged. The highest referral rates were in neonates with multiple risk factors.

Conclusion
A comprehensive intervention and management program must be an integral part of the screening program in the postnatal period. Public awareness about the value of hearing screening is important for follow-up to be more effective. Monitoring of ototoxic drug administration and further assessment of the high prevalence of hyperbilirubinemia are needed. A team of obstetricians, pediatricians, and audiologists is needed to identify and assess risk factors.

Keywords:
care unit neonates, hearing loss, high-risk register, hyperbilirubinemia, low birth weight-mechanical ventilation-ototoxic drugs, neonatal hearing screening, neonatal intensive

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Introduction

Hearing impairment is 20 times more prevalent in neonates than are other disorders that are routinely screened for, including hypothyroidism, sickle cell anemia, and phenylketonuria [1]. Screening only those infants who meet the high-risk register (HRR) criteria is not enough, because as many as 50% of infants born with hearing loss have no known risk factors. Reliable screening tests that minimize referral rates and maximize sensitivity and specificity are available [2].

Mass screening of hearing in children is based on the concept of secondary prevention. In recognition of

congenital or early acquired (i.e. neonatal) hearing disability, numerous hearing screening programs have been introduced worldwide [3]. In contrast to the recommendations of the Joint Committee on Infant Hearing (JCIH), neonatal hearing screening programs are still not universally available, and many countries implement elective screening in high-risk newborns [4]. The reasons for such elective screening are practical: prolonged stay of several newborns in the same hospital; and issues of cost, as at least half of the congenitally deaf newborns are detected in neonatal ICU (NICU) screening [5].

It was noted that early detection of hearing loss will improve the success of programs. Moreover, the recent studies supported by the National Institute of Health have concluded that children whose hearing loss is identified and who receive appropriate intervention before 6 months of age develop significantly better language ability than those who are identified later [6]. The same observations were stated by Hall [7], regardless of the degree of hearing loss. In contrast, delayed identification and management of severe to profound hearing impairment may impede the child's ability to adapt to life in a hearing-abled world or in the deaf community [8].

The primary goal of this study was to assess the failure rate of neonatal hearing screening in a HRR group of neonates and the relative importance of these risk factors in hearing impairment in the NICU of the El-Mounira Children Hospital and identify the incidence of hearing loss.

Patients and methods

The study was conducted on 130 neonates (260 ears) classified into two main groups: the control group and the study group.

The control group included 30 neonates (60 ears were examined), 15 boys and 15 girls, delivered naturally or by cesarean section at the Maternity Hospital, Kasr Al-Aini, Cairo University. Gestational age ranged from 35 to 40 weeks. Birth weight ranged from 2000 to 4000 g, and they had no history or signs of risk for hearing impairment. All the newborns were examined by transient evoked otoacoustic emission (TEOAE) devices between the first and fourth day of life.

The study group included 100 cases (200 ears), 53 boys and 47 girls, selected from the NICU, El-Mounira Children Hospital, Cairo University, between January 2007 and May 2007.The screening was carried out over 2–3 days in a week. The cases were between the ages of 3 and 23 days, and gestational age ranged from 29 to 40 weeks. These cases fulfilled the selection criteria of the HRR of the Joint Committee of Infant Hearing (1994). This group was further subdivided into two groups according to the number of risk factors: the group with two risk factors and the group with multiple risk factors (more than two risk factors).

A portable TEOAE screener (Echo-Screen) was used for the first-stage screening, which gave a Pass or Refer response. A second-stage screening after 3–4 weeks was carried out for cases given a Refer response, using the same equipment. Cases given a Refer response underwent an Automated Auditory Brainstem Response (AABR) test using the same portable Echo-Screen device immediately after the second TEOAE screening test.

The newborns were tested with AABR after they failed the second-stage TEOAE tests, using three surface electrodes. The newborn was tested first at 35 and 55 dBnHL.

Both the TEOAE and AABR screening tests were conducted during natural sleep with no sedation.

Statistical analysis

All statistical calculations and analyses were carried out using the computer program SPSS version 11 (Statistical Package for Social Science; SPSS Inc., Chicago, Illinois, USA).

The basic statistical analysis included arithmetic mean, SD, range, median, frequencies (number of cases), and relative frequencies (percentages) of age and sex. Comparison between different groups in the present study was made using Student's *t*-test to compare continuous data when normally distributed, and the χ^2-test was applied to compare the frequency of qualitative variables among different groups.

All *P*-values are two-sided; *P*-value less than 0.05 was considered significant.

Results

From the 30 full-term neonates in the well-baby nursery, 26 (86.7%) passed the screening test and 4 (13.3%) failed. In the initial (first) screening of the 100 neonates in the NICU group using TEOAE devices, 71% were given a Pass response and 29% were given a Refer. The distribution of the risk factors is shown in Table 1.

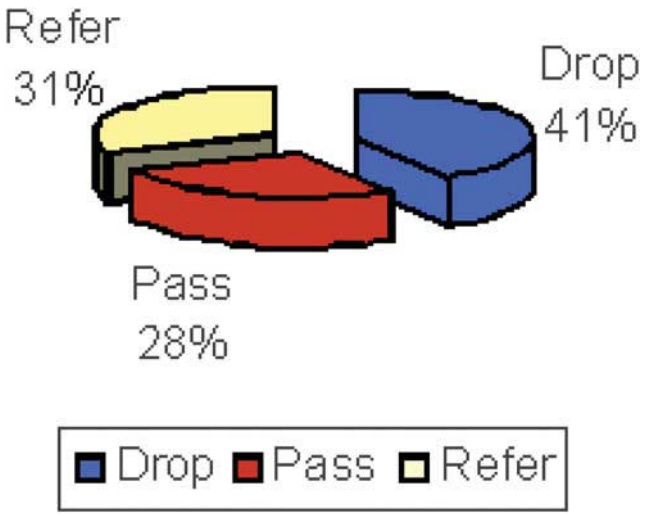
From Table 1, the most frequent risk factor in the NICU is observed to be hyperbilirubinemia, followed by low birth weight and mechanical ventilation. However, it is difficult to assess the magnitude of ototoxicity because of the empirical use of these drugs in the NICU and lack of plasma level monitoring.

Table 1 Distribution of risk factors in the neonatal ICU

		Number	Percent
Valid	0, 1	55	55.0
	0, 1, 2	10	10.0
	0, 1, 3	3	3.0
	0, 1, 4	1	1.0
	0, 2	13	13.0
	0, 2, 3	6	6.0
	0, 3	12	12.0
	Total	100	100.0

Key of risk factors: 0, ototoxicity; 1, hyperbilirubinemia; 2, very low birth weight; 3, mechanical ventilation; 4, craniofacial anomalies.

Figure 1



Pie chart showing the outcome of the second-stage screening.

A follow-up investigation was planned for all neonates who did not pass the initial screening by TEOAE. A second screening or rescreening was carried out in the follow-up clinic in El-Monira Hospital, but out of the 29 cases that were given a Refer response 12 were not brought (dropped out) for follow-up (41.4%). The 17 children who were brought for follow-up were reassessed by TEOAE, and an AABR was performed on the group that was given a Refer. AABR was applied in 10 cases, of which nine were given a Refer response and one was given a Pass (Fig. 1).

Because of the relatively high dropout rate, the estimation of hearing loss incidence adopts an extrapolation policy. In other words, the neonates who underwent the second screening were considered as a random sample for the total neonates who failed the initial screening in order to overcome this dropout rate. This method of calculation was also reported by Finitzo *et al.* [9].

To estimate the true referral rate in this group, the identified number in the second screening [9] was divided by the number of cases who failed the initial screening and were brought for rescreening [10]. This percentage (52.9%) was multiplied by the number of neonates who required rescreening (29). By dividing the total number screened, the probability of referral was 15.4% in this sample, after adjusting for dropout cases (Figs 2–4).

Figure 2 shows that a statistically significant (χ^2-test) difference was found in the group with multiple risk factors (more than two risk factors) and that the probability of referral increased by more than two-fold when risk factors were combined ($\chi^2 = 16.4$, $P < 0.05$).

Discussion

The JCIH recommends a '1–3–6' plan: all infants should be screened before 1 month of age, preferably before discharge; diagnostics should be completed by 3 months of age for those referred after rescreening; for those with confirmed hearing loss, intervention should be initiated by 6 months of age [11].

The present study was carried out on 130 newborns, of whom 30 had no risk factors for hearing loss (60 ears). In this screening, 26 (86.7%) neonates passed the test and 4 (13.3%) had a Refer result. This prevalence of failure was higher than cited by many authors [12,13]; they found prevalence rates of hearing loss ranging from 0 to 5%.

However, all the previous authors had screened their cases after the second day of life, whereas in our study the cases were screened in the first 48 h of life, as reported by Levi *et al.* [14], who screened in the first 10–48 h of life and reported a 22% referral rate. When the test was repeated after 108 h of life, the failure rate dropped to about 1%. This could be explained by the presence of vernix caseosa in the external canal or effusion in the middle ear, which could have been residual amniotic fluid.

The 100 neonates who formed the case group were screened in the NICU. In the present study, 29% of infants obtained a Refer result in the initial screening (Fig. 4) and the final percentage of failure (after adjustment for dropouts) was 15.4%. Among the Egyptian NICU neonates, El Danasoury *et al.* [15] reported the presence of hearing impairment in 19.1% of neonates belonging to the HHR group. Makky and Rashid [16] found 24% hearing impairment in those belonging to the HHR group. In addition, El Gamal *et al.* [17] reported hearing loss in 20% of NICU neonates. Gupta *et al.* [10] demonstrated a 30.88% hearing loss among children who have been successfully discharged from NICUs in India. In contrast, a lower prevalence of hearing loss was reported in the HHRs: it varied between 1.5 and 7% [4].

Although risk factors for hearing loss have been used for screening in many countries worldwide, these factors are constantly refined by the JCIH. However, we should not consider these factors as the 'gold standard' and assign the same relative importance, because the situation in different countries or time periods may vary considerably. For example, a developing country may use more ototoxic agents without assessing drug blood levels because of the lack of resources, whereas another developed country may not use such agents. Therefore, continuous investigation of the relative importance of high-risk factors of hearing loss is essential to assess, refine these factors, and modify according to the current clinical practice. Moreover, such studies are very useful when funding is limited, and screening should be targeted to the most vulnerable neonates [4].

In this work, the high-risk newborns were categorized according to the number of risk factors into the following: groups with two risk factors and groups with multiple risk factors. The prevalence of hearing loss in those with two risk factors was 11.1%, whereas those with multiple risk factors had a referral rate of about 44.45%. Therefore, it is clear that infants who suffer many clinical adverse effects during their neonatal course are more liable to develop hearing impairment. This was supported by several

Figure 2

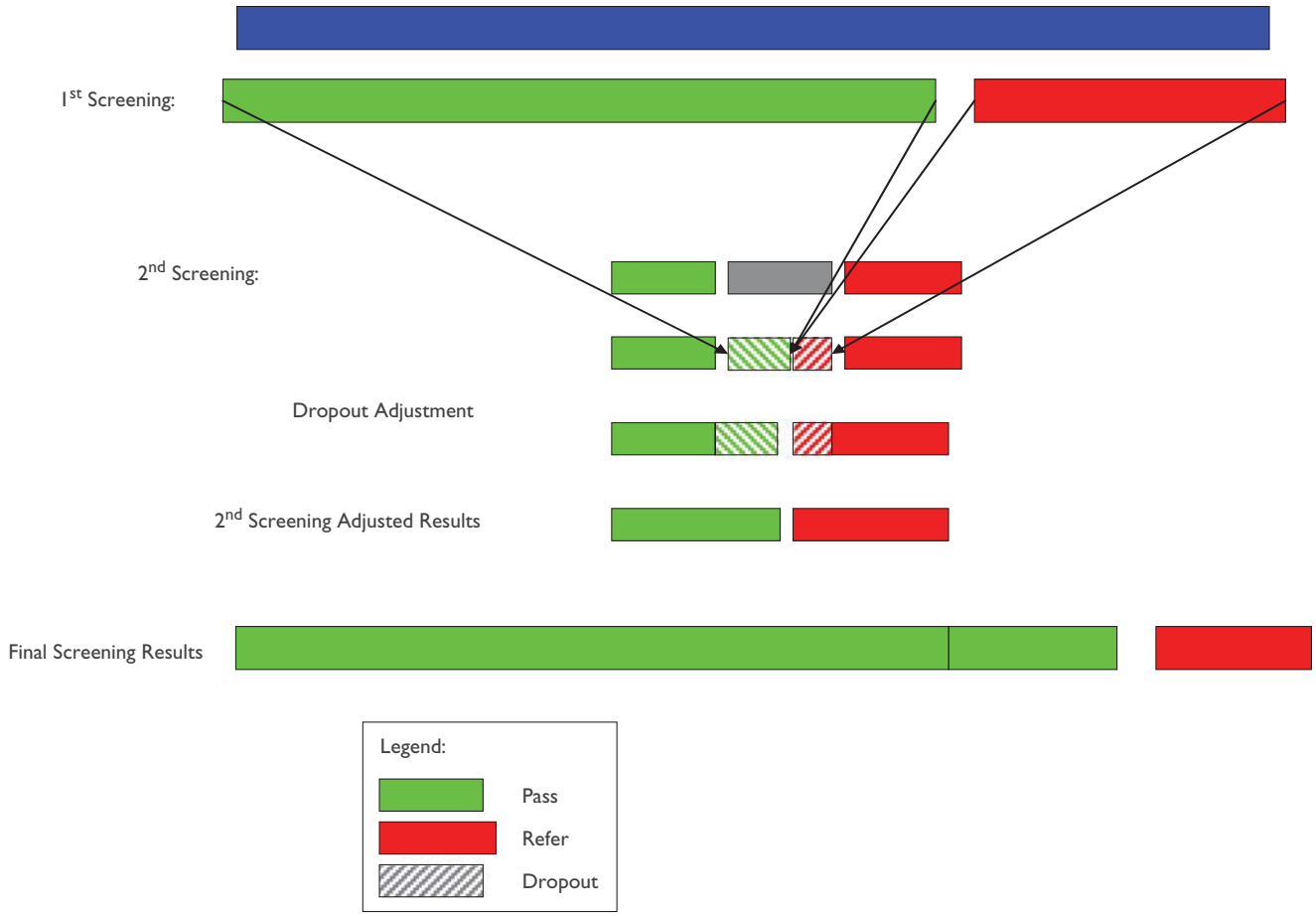
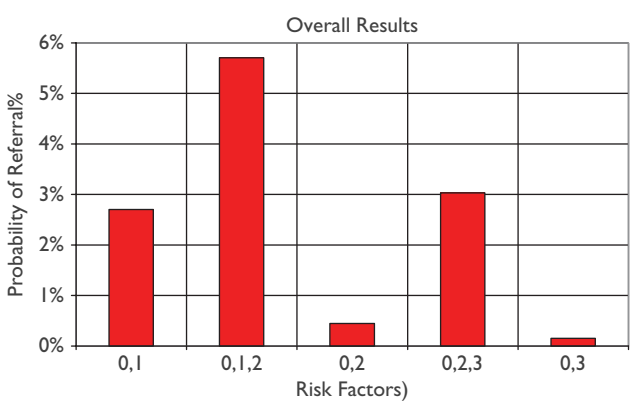


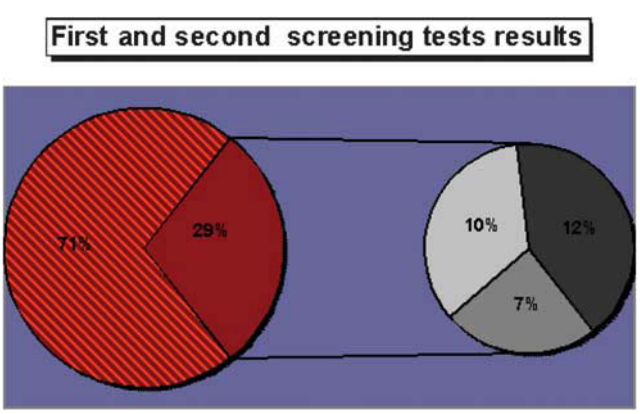
Diagram showing different steps of screening and the procedure of dropout adjustment.

Figure 3



Probability of Referral in the neonatal ICU group (after dropout adjustment). Key of risk factors: 0, ototoxicity; 1, hyperbilirubinemia; 2, very low birth weight; 3, mechanical ventilation; 4, craniofacial anomalies.

Figure 4



Two pie charts showing first and second screening results.

studies; El Gamal *et al.* [17] studied 90 cases selected from the NICU. They reported a failure rate of 54% in the group with multiple risk factors and 20% in the group with a single risk factor. A lower rate was found by El Danasoury *et al.* [15], which was 27.7% in the multiple risk group and 14.3 in the single risk group; such discrepancy was due to lack of sharp demarcation between the groups, which was due to defective data for the newborn.

The most frequent risk factor encountered in the NICU was ototoxicity (100%), followed by hyperbilirubinemia (55%), low birth weight (14.5%), mechanical ventilation for more than 5 days (11.5%), and finally craniofacial anomalies (1%) (Table 1). Vohr *et al.* [18] found that the

four most frequent risk factors in the NICU were ototoxic drugs, low birth weight, mechanical ventilation for more than 5 days, and a low Apgar score. Korres *et al.* [4] found that toxic levels of ototoxic drugs, mechanical ventilation for more than 24 h, prematurity, and low birth weight were the four frequent risk factors. Although there is a slight difference between the two studies, ototoxic drugs, mechanical ventilation, and low birth weight were still the three most frequent risk factors in both studies.

In contrast, hyperbilirubinemia was the most frequent factor encountered in our study and in other studies carried out in Egypt [15,17]. This is a point that needs further research in coordination with pediatricians to assess its magnitude and effect, as neonatal jaundice is more likely to cause central rather than peripheral hearing loss. This necessitates the combination of TEOAE and ABR in cases with neonatal jaundice [19,20].

Infants who failed the initial screening with TEOAE were scheduled for rescreening after 3–4 weeks in the follow-up clinic. However, this second screening was not satisfactory because of the high dropout rate (41.4%; Fig. 1). However, defective follow-up was because of lack of awareness of the public about the drawbacks of undiscovered hearing loss.

A high dropout rate was reported in different countries: 32% in Egypt [21], 40% in South Africa [22], and 56% in Malaysia [23]. Similarly, in the United States, the dropout rate after the initial screening ranged from 25 to 80% at the beginning of establishment of hearing screening programs. The loss to follow-up is due to lack of resources, inadequate data management systems, inconsistent reporting requirements, and lack of communication with medical professionals with regard to follow-up testing [24].

Finally, hearing screening programs based on identification of neonates from an HRR are strongly needed in developing countries, keeping in mind the 'feasibility' of screening. Despite a relatively high prevalence of hearing loss and its serious impact on spoken language, neonatal hearing screening programs are still not universally available [25]. A compromise between no screening and UNHS has been the implementation of screening in NICU newborns under risk for hearing loss.

Conclusion and recommendations

All NICU candidates (>48 h) should be screened for hearing loss before discharge. TEOAE devices can be used successfully for screening in the NICU population.

High-risk neonates who pass the test should have regular and continuous follow-up until the age of 3 years so that the cases that might develop late-onset hearing loss can be identified. Plasma levels should be monitored and the use of ototoxic drugs in the NICUs, especially in the HRRs, must be minimized. It is also necessary to find substitutes other than ototoxic drugs and determine whether its administration is mandatory. Public awareness about the value of hearing screening is important for follow-up to be more effective.

HRRs are good detectors of hearing loss in places where the program is difficult to implement. A team of obstetricians, pediatricians, and audiologists is needed to identify and assess risk factors.

Acknowledgements

Conflicts of interest

There are no conflicts of interest.

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