Potential Pitfalls of Initiating a Newborn Hearing Screening Program

Theodore J. Kanne, MD, MPH; Lynne Schaefer, MA; Jonathan A. Perkins, DO

Objective: To examine the efficacy of a universal screening program for infant hearing.

Design: Retrospective analysis.

Setting: Tertiary care center.

Patients: We examined 2289 (90.2%) of 2537 infants born at our institution from April 1, 1995, to June 30, 1996, for hearing loss (HL).

Interventions: We used a 3-stage protocol with transient evoked otoacoustic emissions (TEOAE) in stages 1 and 2 and diagnostic evaluation in stage 3. Infants without reproducible TEOAEs in either ear after stage 2 were referred to stage 3.


Results: Of the infants undergoing TEOAE screening, 91.1% passed stage 1. Of infants needing repeated testing, 73.7% passed and 26.2% failed. Of the 43 infants referred for diagnostic evaluation, 5 (11.6%) had HL. The combined incidence of conductive and sensorineural HL was 2.18 per 1000 newborns. The prevalence of at least 1 risk factor for HL was 10.4%. The estimated cost of TEOAE screening was $24.48 per infant. Ten percent of infants did not undergo screening due to program deficiencies.

Conclusions: Screening with TEOAE was sufficiently specific for universal screening. However, we were unable to achieve truly universal screening. This is probably the reason for our lower incidence of sensorineural HL. We were unable to continue our universal newborn screening program, due to lack of funding, difficulties with program implementation, and our low incidence of detected HL.


IN 1993, the National Institutes of Health (NIH) convened a consensus development conference on the early identification of hearing impairment in infants and young children. They formulated a consensus statement, recommending universal screening of infant hearing in the first 3 months of life, using evoked otoacoustic emissions followed by auditory brainstem response (ABR) when necessary, and screening of hearing of all infants admitted to the neonatal intensive care unit (NICU) before discharge.1 In the past, universal screening of infant hearing was not possible because the existing testing modalities were labor-intensive or lacked sensitivity and specificity. Before the NIH consensus statement, screening of infant hearing targeted high-risk infants and used ABR.2 However, this method misses approximately one half of all infants with hearing loss (HL), because they lack known risk factors.3 Furthermore, time requirements and screening costs associated with ABR have hampered its widespread use.4 The result is delayed diagnosis of HL in many infants and children.

Delayed diagnosis results in loss of a crucial period in speech and language development, secondarily affecting cognitive, emotional, and social development.5 Early HL identification allows for intervention with hearing amplification and special education during the first 3 years of life.6

The discovery in 1978 of otoacoustic emissions created a new screening modality.7 Otoacoustic emissions are low-energy sound waves emitted by cochlear

This article is also available on our Web site: www.ama-assn.org/oto.
MATERIALS AND METHODS

We reviewed the results of TEOAE for screening of infant hearing from April 1, 1995, to June 30, 1996, at MAMC. During this time we identified 2337 infants born at MAMC. All records were available for review. Infants transferred from other facilities were excluded. The following study cohorts were identified: infants from the well-baby nursery (WBN) and infants admitted to the NICU. Records were analyzed for results of hearing screening, presence of risk factors for HL, and reasons for an infant not undergoing screening.

SCREENING PROCEDURE

After informed consent was obtained, infants underwent screening in a secluded room in the NICU or WBN by an audiologist using a commercially available analyzer (ILO88 Otodynamic Analyzer; Otodynamics Ltd, Herts, England) during the work week. Infants from the NICU underwent screening 24 to 72 hours before their discharge (age range, 2 days to 3 months). Infants from the WBN underwent screening at the first outpatient visit, at approximately 2 weeks of age. All infants underwent assessment for known risk factors for HL at the time of their initial TEOAE screen through parent interview and/or review of the medical record.16 Testing with TEOAE was performed according to established standards.4,7 Results were categorized as pass or fail; a partial-pass category was not included. A pass score was given when an emission spectrum of significant gain was demonstrated across the testing frequency range.17 All others were considered failure scores and were referred for repeated testing. Time to administer each test was recorded.

Infants with failure scores in the first 2 stages were referred for diagnostic evaluation with ABR. If merited on the basis of these results, the infants were referred to a physician for evaluation and treatment.

STATISTICAL ANALYSIS

Statistical analysis was performed using descriptive statistics and power analysis of the existing data to predict the number of missed subjects with HL.

RESULTS

Of 2537 infants born at MAMC, 2208 (87.0%) were admitted to the WBN and 329 (13.0%) were admitted to the NICU. A total of 2289 infants (92.5%) from the WBN and 247 (75.1%) from the NICU underwent screening, 2042 (92.5%) from the WBN and 247 (75.1%) from the NICU.

Data regarding the prevalence of HL risk factors in each cohort are presented in the Table. Two-hundred sixty-three (10.4%) of the 2537 infants had at least 1 risk factor for HL that would merit hearing screening with ABR based on the high-risk register.18 At least 1 risk factor for HL was present in 6.6% of the infants from the WBN and 35.6% of the infants admitted to the NICU.

For infants not undergoing screening in the WBN (n = 166), the following reasons were identified: no referral for hearing screening (34 [20.5%]), loss of eli-
Hundred seventy-nine infants underwent repeated TEOAE sumed to have normal hearing thresholds. One-(91.1%) passed the initial TEOAE test and were as-

ing. Of the 2289 infants undergoing screening, 2085 our 3-stage protocol for universal screening of infant hear-

ing and lack of timely audiology support.

In the NICU, the reasons for infants not un-

port unavailable (61 [36.7%]), and indeterminate (20 [12.0%]). In the NICU, the reasons for infants not un-

going screening (n = 82) were lack of referral for screen-

ing stage 3. Thus, the estimated number of infants with HL could have been missed in the 25 infants not under-

ing retesting after failing stage 1. Based on an HL prevalence of 5/179, approximately 0.21 infants would be missed

at stage 2 screening due to false-negative results. Based on an HL prevalence of 2.8% at stage 2, 0.70 infants with HL would

have not been detected at stage 1. Assuming a TEOAE sensitivity of 0.95, a specificity of 0.90, and an HL prevalence of 0.2%, approximately 0.21 infants would have

false-negative results and be missed in stage 1 screening. Assuming an HL prevalence in stage 2 testing of 2.8% (5 of 179), approximately 0.21 infants would be missed at stage 2 screening due to false-negative results. Based on an HL prevalence of 2.8% at stage 2, 0.70 infants with HL could have been missed in the 25 infants not undergoing retesting after failing stage 1. Based on an HL prevalence of 11.6% (5 of 43) in the infants needing stage 3 testing, 0.46 infants with HL could have been missed among the 4 infants who could not undergo retesting during stage 3. Thus, the estimated number of infants with HL missed in all 3 stages of testing is 2.13 infants. This would increase the overall HL prevalence only slightly from 0.2% to 0.3%.

Admittedly, 1 weakness of our observations was our inability to screen for HL in all infants born at MAMC. Assuming the HL prevalence of the infants not undergoing screening in stage 1 was the same as those undergoing screening (5/2260 [0.2%]), 0.55 infants with HL would

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From Joint Committee on Infant Hearing.16

<table>
<thead>
<tr>
<th>Risk factors*</th>
<th>Well-Baby Nursery</th>
<th>Neonatal Intensive Care Unit</th>
</tr>
</thead>
<tbody>
<tr>
<td>Relative with hearing loss, parental report</td>
<td>33 (1.5)</td>
<td>3 (0.9)</td>
</tr>
<tr>
<td>In utero infection</td>
<td>4 (0.2)</td>
<td>8 (2.4)</td>
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<tr>
<td>Malformations of the head and/or neck</td>
<td>21 (1.9)</td>
<td>9 (2.7)</td>
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<tr>
<td>Birth weight &lt;1500 g</td>
<td>0 (0)</td>
<td>25 (7.6)</td>
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<tr>
<td>Hyperbilirubinemia requiring exchange transfusion</td>
<td>0 (0)</td>
<td>0 (0)</td>
</tr>
<tr>
<td>Ototoxic medications ≥5 d</td>
<td>0 (0)</td>
<td>62 (18.8)</td>
</tr>
<tr>
<td>Meningitis with positive results of cultures</td>
<td>0 (0)</td>
<td>1 (0.3)</td>
</tr>
<tr>
<td>Apgar scores of 0-4 at 1 min or 0-6 at 5 min</td>
<td>90 (4.1)</td>
<td>59 (17.9)</td>
</tr>
<tr>
<td>Mechanical ventilation lasting ≥5 d</td>
<td>0 (0)</td>
<td>35 (10.6)</td>
</tr>
<tr>
<td>Stigmata of syndromes associated with deafness</td>
<td>0 (0)</td>
<td>0 (0)</td>
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</tbody>
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<table>
<thead>
<tr>
<th>Stage 1</th>
<th>TEOAE N=2208</th>
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<tbody>
<tr>
<td>Discharged</td>
<td>Pass n=2085 (91.1%)</td>
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<tr>
<td>Fall n=204 (8.9%)</td>
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<table>
<thead>
<tr>
<th>Stage 2</th>
<th>Retesting With TEOAE n=179 (87.7%)</th>
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<tbody>
<tr>
<td>Discharged</td>
<td>Pass n=132 (73.7%)</td>
</tr>
<tr>
<td>Fall n=47 (26.2%)</td>
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</tbody>
</table>

<table>
<thead>
<tr>
<th>Stage 3</th>
<th>Diagnostic Evaluation n=43 (91.5%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Discharged</td>
<td>Pass n=38 (88.4%)</td>
</tr>
<tr>
<td>Fall n=5 (11.6%)</td>
<td></td>
</tr>
</tbody>
</table>

Three-stage screening protocol for infant hearing at Madigan Army Medical Center, Tacoma, Wash. TEOAE indicates transient evoked otoacoustic emissions.

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Our experience using TEOAE for universal infant hearing screening in a 2-stage fashion suggests that it is cost-effective, confirming results of other published reports.\textsuperscript{14,17,18} When administered by trained personnel, it was reproducible, objective, easily performed, minimally invasive, and quicker than ABR. The 8.7\% rate of false-positive results after stage 1 screening is consistently found with findings of other reports and indicates that it is sufficiently specific for universal screening.\textsuperscript{19} A second TEOAE screening decreased the need for ABR. In our study, only 2.3\% of the infants required diagnostic ABR after the second TEOAE screening. In comparison, 8.9\% would have required ABR if referred directly after the first TEOAE failure, and 10.4\% would have required ABR based on the high-risk register.\textsuperscript{16}

Howerver, universal infant hearing screening at our institution was not particularly efficacious compared with targeted screening according to the high-risk register. No significant interventions took place as a direct result of universal hearing screening that would not have occurred with high-risk screening only. The only infant identified with SNHL would have been identified with targeted screening because of multiple risk factors. Furthermore, the infant’s early death prevented any intervention. Screening with TEOAE identified 4 infants with CHL but no risk factors for HL. All had HL secondary to middle ear effusions, and their respective therapies did not change as a result of universal screening.

The prevalence of HL in our study was lower than that reported by other investigators.\textsuperscript{4} We found 2.18 per 1000 infants undergoing screening had a mild to profound CHL or SNHL in at least 1 ear. The prevalence of significant SNHL was 0.44 per 1000 infants, and 1.75 per 1000 infants had mild to moderate CHL in at least 1 ear. This is in contrast to the rates reported by the Rhode Island Hearing Assessment Project of 5.95 per 1000 infants with an SNHL and 20 per 1000 infants with a CHL.\textsuperscript{4} The rate of SNHL detected in our population using TEOAE is below the generally accepted estimates of 1 to 2 per 1000 infants with moderate to profound SNHL.\textsuperscript{4} This may be due to the characteristics of our sample population and/or too small a sample size. Also, some infants with HL were probably missed due to incomplete screening, especially in the NICU cohort, where 24.9\% did not undergo screening. However, the calculated prevalence of risk factors for HL in our population was similar to other estimates for the general population.\textsuperscript{4}

Since the HL prevalence in our population was low, the cost of HL identification was high. We spent $56 045 for screening in 2289 infants during the 64-week inclusion period and identified 1 infant with a bilateral profound SNHL and 4 infants with mild to moderate CHL. We estimate the cost of hearing screening was $24.48 per infant, which is comparable to other estimates.\textsuperscript{20} However, it cost $11 209 to identify each infant with HL due to the low prevalence. If only SNHL is considered, we spent $56 045 to identify 1 case. This estimate is considerably more than $4378 per infant identified with an SNHL by Maxon et al.\textsuperscript{20} However, our data analysis shows that we potentially missed 2 to 3 infants with HL in our population, because true universal screening was not accomplished. This would have reduced the costs of HL identification to $7000 to $8000 per infant.

These figures can be compared with the estimated costs for other mandated infant screening programs already in existence, such as for phenylketonuria, hypothyroidism, or sickle cell anemia ($20.00 per child and $40 960 per case identified).\textsuperscript{14} The increased costs per child for these other types of screening programs are due to their lower prevalence within the population when compared with HL. This makes screening for HL look rather cost-effective, especially when one considers the benefits of early HL identification.

Although the notion of universal screening of infant hearing for the early identification of HL is good in theory, there are a number of barriers to its effective implementation. We found 3 important prerequisites to an effective universal screening program. First, primary care providers (pediatricians, family practitioners, and nursing staff) must appreciate the importance of early HL identification and prompt auditory intervention in hearing-impaired infants to help ensure early language development. At our institution, we had difficulty attaining complete access to the NICU population due to inconsistent referrals from the staff. Too often, this resulted in infants in the NICU not undergoing screening during the window between clinical stability and hospital discharge. Second, dedicated personnel and funding are necessary for hearing screening. In an increasing number of states, mandated screening and governmental funding have enabled hiring of additional personnel specifically for hearing screening. We believe that outside funding is necessary to maintain effective screening programs and to ensure appropriate auditory intervention in those infants identified with HL. In our program, the audiologists performed all screening because no other staff was available. This was not cost-effective, but without an external or internal source of funding, our institution could not justify hiring additional personnel to provide staffing of the screening program 7 d/wk. The TEOAE screening can be performed by nonaudiologists, provided they are adequately trained and supervised.\textsuperscript{12} We recognize the audiologist’s role in a screening program is better served as supervisor and consultant. Third, uniform access to newborns at some point within the first 3 months of life is needed. Preferably, screening should be performed before discharge. At our institution, the 2-week well-infant outpatient appointment provided the best venue for access to infants from the WBN, because a well-established clinic system was already in place. Early discharge of mothers and healthy newborns would have made in-patient screening logistically and technically more difficult. Conversely, access before discharge of infants in the NICU was necessary, because no reliable system existed for outpatient screening.

Any attempt to institute a universal screening program for infant hearing without the cooperation of the primary care provider will be fraught with difficulty. More education is needed concerning the adverse effects of delayed HL identification to ensure cooperation in such an undertaking. We agree with the comments by Bess and Paradise\textsuperscript{21,22} concerning the efficacy of efforts directed at “educating both primary health care providers and par-
Universal newborn hearing screening is logistically difficult. For universal screening of infant hearing to be successful on their part will help in the identification of delayed-onset or acquired HL that universal infant screening would not detect and ensure improved cooperation with universal screening. Universal hearing screening for infants requires a dedicated, proficient staff and appropriate financial resources to ensure quality control. In our program, we had to discontinue TEOAE screening in the WBN, because there was no clear source of funding to hire more personnel. Instead, we have concentrated our efforts on more comprehensive screening with TEOAE for the infants from the NICU, where the prevalence of HL risk factors is highest.

Testing with TEOAE is a good device for universal screening of infant hearing. Two-staged screening significantly decreased the rate of ABR use compared with ABR testing based on the high-risk register. Our study cohort demonstrated a lower HL prevalence than that seen in other published results. This may have been the result of our inability to achieve truly universal screening. Universal screening of infant hearing is logistically difficult. It requires a dedicated staff and the full cooperation of an educated primary care community to be effective. A screening program is effective only if the ability for appropriate intervention is in place once HL is identified. For universal screening of infant hearing to become commonplace in our practice setting, an external source of funding is necessary, and/or legislation must mandate it.

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The views and assertions contained herein reflect the private opinions of the authors. The content does not represent the position of the Department of the Army or the Department of Defense.


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REFERENCES


