Limitations of Universal Newborn Hearing Screening in Early Identification of Pediatric Cochlear Implant Candidates

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Objectives: To determine whether implementation of universal newborn hearing screening (UNHS) in the state of Illinois has affected the ages at diagnosis of hearing loss and implantation in children receiving cochlear implants and to determine how often children undergoing implantation had UNHS results with no indication of hearing loss (pass).

Design: Retrospective case review of 417 randomly selected pediatric implant recipients born before and after UNHS was mandated by law in Illinois. Data analyzed included hearing screening status, ages at initial diagnosis of sensorineural hearing loss (SNHL) and severe to profound SNHL, and age at implantation.

Setting: Tertiary care medical center.


Main Outcome Measures: Ages at diagnosis of SNHL and implantation.

Results: Children born after legally mandated UNHS had significantly younger ages at diagnosis and implantation. However, a younger age at diagnosis of SNHL was not achieved in children who had passed UNHS or who were not screened. Approximately 30% of pediatric implant recipients passed UNHS, regardless of the cause of hearing loss or the presence or absence of known risk factors.

Conclusions: Almost one-third of our pediatric implant recipients pass UNHS and are older at the time of initial diagnosis and implantation than their peers who fail UNHS. Delayed onset of SNHL limits our ability to achieve early diagnosis and implantation of a significant number of deaf children. This problem will not be solved by the current design of universal hearing screening programs.


In the past, late-onset progressive hearing loss in children was thought to occur most commonly in those with a history of neonatal risk factors such as prolonged assisted ventilation, administration of ototoxic medication, congenital cytomegalovirus (CMV) infection, or hyperbilirubinemia. However, the number of children with onset of significant permanent hearing loss after the newborn period, the timing of onset, and the stability of hearing loss are not well understood. The authors of an Australian study estimated that approximately 24% of significant permanent childhood hearing losses diagnosed by 9 years of age begin in the postnatal period and are thus undetected by UNHS.1

Before the implementation of newborn hearing screening programs, children with profound bilateral SNHL were usually identified at 17 to 24 months of age.2-4 When UNHS was initially conceived, it was presumed that most hearing-impaired children, especially those without risk factors for progressive hearing loss, would fail the objective screening performed during the newborn period, thus making early diagnosis and intervention possible on a widespread basis.

In 1999, the state of Illinois passed the Hearing Screening for Newborns Act (410 ILCS 213/1), which legislatively mandated UNHS by all birthing hospitals to become effective in Illinois by December 31, 2002 (ie, at the start of 2003).5 During the years between the passage of this legislation and date of required implementation...
tion, many hospitals implemented UNHS programs using a 2-step screening process with objective measures (otoacoustic emissions or automated auditory brainstem response testing), as specified in the legislation. Before passage of this legislation, only children with known risk factors were typically screened before hospital discharge.

Despite significant success in lowering the age at which many children with hearing loss are identified, our experience finds it not uncommon for parents of cochlear implant candidates to report that newborn hearing screening did not identify hearing loss in their child (ie, the child passed screening). This situation is concerning because pediatric implant candidates who pass UNHS only to lose their hearing during infancy or early childhood may be particularly disadvantaged. It is reasonable to assume that passing UNHS, especially in the absence of known risk factors, may falsely reassure parents and professionals. Therefore, hearing loss in this subpopulation of children may not be diagnosed, and these children may not undergo implantation at as young an age as those who fail UNHS. Because UNHS programs, by definition, are structured to screen for hearing loss in newborns, it is important to understand how often delayed-onset hearing loss may limit our ability to provide early identification and implantation among infants and young children.

**METHODS**

We performed a retrospective institutional review board-approved study of children undergoing implantation at our institution from 1991 through 2008. We selected 417 children at random from a pool of 660 pediatric implant recipients. After review of the medical records, all children with postnatally acquired deafness were excluded. As a consequence, 26 children with acquired deafness due to bacterial meningitis and chemotherapy were eliminated, leaving 391 implant recipients. Information abstracted from the medical records included history and results of newborn hearing screening reported by the parent or the audiologist, the age at which hearing loss was diagnosed, the age at which severe to profound loss was diagnosed, and the age at implantation. The following neonatal risk factors for SNHL were recorded: neonatal intensive care unit stay of longer than 5 days, use of assisted ventilation, hyperbilirubinemia, use of ototoxic medications or loop diuretics, extracorporeal membrane oxygenation, craniofacial abnormalities, family history of hearing loss (siblings and parents), and in utero infection. The cause of hearing loss was recorded if known and included auditory neuropathy, presence of cochlear malformations based on temporal bone imaging, mutations of the connexin genes, history of in utero infection such as rubella and CMV, and syndromic SNHL.

**PATIENT CHARACTERISTICS**

The overall group of 391 pediatric implant recipients had an even sex distribution (51.3% male and 48.7% female). Hearing screening results were available for 49.6% of the recipients. For some children who underwent implantation elsewhere (and were followed up by our institution), were adopted, or were born in other countries, it was not known whether screening might have been performed. Mean (SD) age at implantation of the overall group was 4.2 (3.2) years. The number of children with risk factors or a diagnosed cause of SNHL is summarized in the following tabulation:

**RESULTS**

The implant recipients were divided between those born before (n = 264) and after (n = 127) UNHS became a mandatory legal requirement in 2003. Of the 264 children born before 2003, 86 (32.6%) had screening results compared with 108 of the 127 (85.0%) born during and after 2003. The Figure shows the distribution of hearing screening by year of birth. The premandate and postmandate groups did not differ by distribution of sex, risk factors, or diagnoses. Thirty-two of 86 children (37.2%) in the premandate group passed hearing screening, as did 32 of 108 children (29.6%) in the postmandate group. Ten of 24 children (41.7%) in the premandate group had no risk factors or cause of hearing loss identifiable at birth and passed hearing screening, as did 11 of 34 children (32.4%) in the postmandate group.

Table 1 shows screening status and result by cause of hearing loss or presence of risk factors in the group of children born after mandated UNHS. Of the 127 children in this group, 83 (65.4%) had a known cause of hearing loss or at least 1 risk factor. Twenty-one of the 83 children (25.3%) with a diagnosis or with risk factors passed hearing screening. Twenty percent to 33% of children in each diagnostic/risk category passed UNHS. Of the 7 children with cochlear malformations who passed screening, 2 had wide vestibular aqueducts with poor partition of the apical and middle turns, and 5 had a variety of malformations with a relatively well-formed basal and middle turn of the cochlea. One child with congenital CMV infection passed screening. Two children with syndromic SNHL, one with Usher syndrome and the other with Waardenburg syndrome, also passed.

**Diagnosis/Risk Factor**

<table>
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<tr>
<th>No. (%) of Children (N=391)</th>
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<tr>
<td>Auditory neuropathy</td>
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<tr>
<td>Cochlear malformation</td>
</tr>
<tr>
<td>Connexin mutation</td>
</tr>
<tr>
<td>Congenital infection</td>
</tr>
<tr>
<td>Syndrome</td>
</tr>
<tr>
<td>Neonatal risk factors</td>
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Neonatal risk factors were identified in 34.8% of the children, with a NICU stay of longer than 5 days (19.3%) and family history (14.1%) being the most common risk factors. The single most common diagnosis was the presence of a cochlear malformation that consisted of wide vestibular aqueducts in 38 of 63 children (60.3%). The next most common diagnosis was mutation of connexin genes 26 (OMIM *121011) and 30 (OMIM *604418). Of the 37 children diagnosed as having syndromic SNHL, 14 (37.8%) had Usher syndrome and 4 (10.8%) had Waardenburg syndrome. Congenital CMV accounted for 17 of 24 diagnosed congenital infections (70.8%). Overall, 63.3% of the children had at least 1 risk factor or diagnosis associated with SNHL.

**DATA ANALYSIS**

Data were entered into a spreadsheet (Excel; Microsoft Corporation, Redmond, Washington) and then imported into a statistical program (SPSS 11.5; SPSS, Inc, Chicago, Illinois) for analysis. Descriptive and inferential analyses were performed. A χ² test was computed for comparison of categorical variables between groups. The nonparametric Mann-Whitney test was used to compare age at onset of hearing loss and age at implantation between groups because of the large and unequal variability of these measures within each group. The criterion for statistical significance was set at P < .05 (2-tailed).
Figure. Graph demonstrating increase in hearing screening before and after universal newborn hearing screening (UNHS) in the state of Illinois was legislatively mandated in 1999 to be effective by December 31, 2002. First year in which UNHS was legally required for all children born in Illinois.

Table 1. Screening Result by Cause of Hearing Loss or Presence of Risk Factors in Mandatory UNHS Groupa

<table>
<thead>
<tr>
<th>Diagnosis/Risk Factor</th>
<th>Children Screened, No. (%)</th>
<th>Pass, No. (%)</th>
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<tbody>
<tr>
<td>Auditory neuropathy</td>
<td>3/6 (50.0)</td>
<td>1/3 (33.3)</td>
</tr>
<tr>
<td>Cochlear malformation</td>
<td>19/22 (86.4)</td>
<td>7/19 (36.8)</td>
</tr>
<tr>
<td>Connexin mutation</td>
<td>23/24 (95.8)</td>
<td>6/23 (26.1)</td>
</tr>
<tr>
<td>Congenital infection</td>
<td>5/5 (100)</td>
<td>1/5 (20.0)</td>
</tr>
<tr>
<td>Syndrome</td>
<td>10/15 (66.7)</td>
<td>2/10 (20.0)</td>
</tr>
<tr>
<td>Neonatal risk factors</td>
<td>42/45 (93.3)</td>
<td>12/42 (28.6)</td>
</tr>
</tbody>
</table>

Abbreviation: UNHS, universal newborn hearing screening.

For all comparisons between groups, *P < .001 by the Mann-Whitney test.

Table 2 shows the mean age at diagnosis and implantation in the premandate and postmandate groups. Although there was considerable variability within each group, age at diagnosis of hearing loss, age at diagnosis of severe to profound hearing loss, and age at implantation were all significantly younger in the postmandate group (*P < .001 for all). Age at diagnosis of hearing loss and implantation in the mandated UNHS group was further examined by UNHS result (pass, fail, or not screened) (Table 3). Children who failed UNHS were diagnosed on average at 5.9 months of age and underwent implantation at 1.7 years of age, whereas those not screened or who passed UNHS were significantly older at diagnosis and implantation. There was no significant difference in age at diagnosis or at implantation between those children who were not screened and those who passed UNHS screening.

Table 2. Mean Age at Diagnosis of Hearing Loss and Implantation Before and After Implementation of Mandatory UNHSa

<table>
<thead>
<tr>
<th>Age Evaluated</th>
<th>Premandate Group</th>
<th>Postmandate Group</th>
</tr>
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<tbody>
<tr>
<td>At diagnosis, mo</td>
<td>21.4 (19.9)</td>
<td>11.1 (11.7)</td>
</tr>
<tr>
<td>At severe to profound hearing loss, mo</td>
<td>34.8 (37.6)</td>
<td>12.4 (12.3)</td>
</tr>
<tr>
<td>At implantation, y</td>
<td>5.1 (3.5)</td>
<td>2.1 (1.2)</td>
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Abbreviation: UNHS, universal newborn hearing screening.
aFor all comparisons between groups, *P = .001 by the Mann-Whitney test.

The committee suggested use of a high-risk registry, based on risk factors for hearing loss, to determine which infants should be referred for audiological evaluation. As a result of this position statement, many hospitals instituted hearing screening programs using auditory evoked potentials for graduates of the NICU but rarely for graduates of the well-infant nursery. It became clear by the early 1990s that most children with known risk factors for hearing loss never required NICU admission and thus these infants were not screened before hospital discharge. They were identified at an older age than infants screened in the NICU. The degree of success achieved by using risk factors as the criteria for hearing screening was also recognized to be limited by the significant number of infants lacking a history of risk factors. To address these issues, in 1994 JCIH first declared that universal early identification and management of congenital hearing loss should be a national goal. Universal newborn hearing screening was endorsed by the American Academy of Pediatrics in 1999 on the basis of their assessment that there was adequate research demonstrating newborn hearing screening met criteria necessary to justify implementation of a universal program. At present, UNHS is defined as all infants having access to hearing screening performed before hospital discharge for infants graduating from the well-infant nursery and NICU and referral and access to screening during the neonatal period for infants born at home or in alternative birthing facilities. In our study, although the mean ages at diag-

Table 3. Mean Age at Diagnosis of Hearing Loss and Implantation by Screening Category in Mandatory UNHS Groupa

<table>
<thead>
<tr>
<th>Age Evaluated</th>
<th>No Screening</th>
<th>Passed</th>
<th>Failed</th>
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<tbody>
<tr>
<td>At diagnosis, mo</td>
<td>20.2 (13.9)</td>
<td>18.5 (12.3)</td>
<td>5.9 (6.4)</td>
</tr>
<tr>
<td>At severe to profound hearing loss, mo</td>
<td>22.8 (15.5)</td>
<td>20.5 (12.8)</td>
<td>6.7 (7.0)</td>
</tr>
<tr>
<td>At implantation, y</td>
<td>2.6 (1.3)</td>
<td>2.6 (1.2)</td>
<td>1.7 (1.0)</td>
</tr>
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</table>

Abbreviation: UNHS, universal newborn hearing screening.
aN = 127. For all comparisons between those who failed vs those who passed screening and those who failed screening vs those who did not undergo screening, *P < .001 by the Mann-Whitney test. Differences were not significant between those who passed screening vs those who did not undergo screening.
nosis and implantation in children born during and after 2003, when UNHS was legally required, were significantly younger, the benefit was primarily incurred by those who had failed UNHS. The subgroup that failed UNHS was diagnosed at a mean age of 5.9 months compared with the overall group’s mean age of 11.1 months. Children who passed UNHS during the period of mandated UNHS were not significantly younger at diagnosis than those children who were not screened. This finding was true when we compared the groups of children born before and after UNHS was legally required who did not undergo screening. Children in the mandated UNHS group who passed UNHS or were not screened were not diagnosed at a younger age than that reported in the literature before the establishment of UNHS programs. Younger age at diagnosis was not achieved in these children despite ongoing and widespread education of primary care physicians and the public about the benefits of early identification of childhood hearing loss.

This finding highlights the difficulty of identifying hearing loss in young children and underscores the reason for development of universal screening programs using objective test measures. The decline in age at implantation among children born after legally mandated UNHS in 2003 compared with those born before 2003 likely occurred in large part because of the belief that early implantation in children in whom amplification does not provide significant access to spoken language is advantageous. The benefits of younger age at implantation are supported by many studies demonstrating a strong relationship between a shorter period of auditory deprivation and improved speech perception and language outcomes.11-14 However, the advantage conferred by having failed UNHS is clearly evident in the group of children who were born subsequent to mandatory UNHS in Illinois. The children who failed UNHS underwent implantation on average at 1.7 years of age compared with 2.6 years for those who were not screened and those who passed UNHS. The earlier age at implantation of those who failed UNHS was facilitated by this group achieving definitive diagnosis of hearing loss, on average, by 5.9 months of age and of severe to profound loss by 6.7 months, thus permitting time for the medical and audiological evaluation needed to confirm implant candidacy and for completion of the insurance process.

Approximately one-third of our implant candidates had at least 1 neonatal risk factor for hearing loss, with the most common risk factor other than a NICU stay of longer than 5 days being a family history of hearing loss (14.1%). Family history, typically in a sibling, has been previously reported as the most common risk factor.8 The premandate and postmandate groups did not differ in distribution of causes of hearing loss or of the presence of specific risk factors. Among those born after legally mandated UNHS, a history of passing UNHS occurred in approximately 30% of children, whether they were known to have risk factors at birth (21 of 83) or not (11 of 34). This finding is interesting when one considers that, before widespread implementation of UNHS, it was expected that most children with congenital hearing loss would be identified at birth if objective testing was performed. It was thought that the children most likely to initially pass UNHS only to experience progressive hear-
tional information confirming the screening test modality, the result, and the name of the birthing hospital would be helpful to confirm the accuracy of our results and would help us to investigate the possibility that issues related to screening equipment, testing methods, or reporting systems at birthing hospitals might be causing false-negative results. However, we believe the latter issue is unlikely to have significantly affected our results because our implant candidates come from a large geographic area served by many different birthing hospitals.

This study also lacked sufficient numbers of implant recipients to draw conclusions regarding the frequency with which specific causes are associated with delayed onset of hearing loss. Plans are under way to implement a prospective study of children with bilateral SNHL of varying degrees to address these issues and to obtain a larger database for analysis.

CONCLUSIONS

One-third of our cochlear implant population born after UNHS was legally mandated passed the screening. In light of their older age at diagnosis and implantation compared with their peers who failed UNHS, these children may not have received the potential benefit of improved outcomes conferred by early identification and intervention.

At present, delayed onset of SNHL limits our ability to achieve early implantation of a significant number of deaf children. The current UNHS programs, by their very nature and design, will not solve this dilemma. There is little reason to be optimistic that most children who pass UNHS will be referred in a timely manner for further testing based on concerns of parents or primary care physicians. This situation raises the issue of whether repeating mandatory hearing screening for all children before 1 year of age would be beneficial. The issue of additional mandatory testing to identify a smaller but significant number of hearing-impaired children is a public policy issue that requires further research and consideration. More information about the age at onset and the natural history of early childhood SNHL of delayed onset is needed. Before implementation, issues of consideration include age and location of additional screening, as well as funding and cost-effectiveness.

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Author Contributions: Dr Young had full access to all the data in the study and takes responsibility for the integrity of the data and the accuracy of the data analysis.

Study concept and design: Young. Acquisition of data: Young, Reilly, and Burke. Analysis and interpretation of data: Young and Reilly. Drafting of the manuscript: Young. Critical revision of the manuscript for important intellectual content: Young, Reilly, and Burke. Obtained funding: Young. Administrative, technical, and material support: Young, Reilly, and Burke. Study supervision: Young.

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REFERENCES