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Pathway to Amplification in Children who Passed their Universal Newborn Hearing Screening Bilaterally

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Abstract

Purpose: The aim of this study was to investigate the pathway to amplification technologies for children who passed their universal newborn hearing screening (UNHS) bilaterally with the intent of revealing effective strategies to identify children with acquired or progressive hearing losses. Additionally, the degrees, types, and causes of hearing loss, as well as the types of amplification used by the patients were investigated.

Methodology: Medical records were reviewed for 102 children who passed their UNHS bilaterally and who are enrolled in the Boston Children's Hospital Amplification or Cochlear Implant Programs. Of the 204 total ears, 177 ears were identified with hearing loss and were included in the study.

Conclusion: More than half of new hearing loss identifications in children over 11 years and approximately one third of all new hearing loss identifications resulted from a referred hearing screening. For children under age three, a speech-language delay was the most common reason for referral leading to identification of a permanent, postnatal hearing loss. This study emphasizes the importance of routine hearing screenings in school-aged children as well as highlights the need for audiological evaluations when signs of childhood hearing loss arise, such as a speech-language delay.

Keywords: UNHS, hearing aid, cochlear implant, acquired hearing loss

Acronyms: AABR = automated auditory brainstem response; BCH = Boston Children's Hospital; CMV = cytomegalovirus; cCMV = congenital cytomegalovirus; EVA = enlarged vestibular aqueduct; OAE = otoacoustic emission; UNHS = universal newborn hearing screening

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Universal Newborn Hearing Screening (UNHS) has remarkable value in decreasing the average age of hearing loss identification (Dalzell et al., 2000; Vohr et al., 1998); however, it is possible for a child to pass the newborn hearing screening with a mild, congenital hearing loss. Current automated auditory brainstem response (AABR) testing and otoacoustic emissions (OAEs) screening tools frequently use a 30–35 dB criterion level, which would fail to capture newborns with a slight to mild hearing loss. Johnson et al. (2005) estimates that approximately 23% of newborns who have a permanent hearing loss would pass a UNHS conducted via AABR as a result of the chosen screening level.

In addition, there are many causes of delayed-onset congenital or acquired hearing loss that can occur in childhood, including hearing loss associated with genetic mutations, infectious diseases, anatomic abnormalities, trauma, and ototoxicity (Kenna, 2015). By age nine, 25% of permanent childhood hearing loss is postnatal in nature, suggesting that while the UNHS is playing a significant role in the identification of permanent childhood hearing loss, provisions must also be in place to identify children who acquire hearing loss postnatally (Weichbold et al., 2006; Watkin & Baldwin, 2011). Among the cases of permanent childhood hearing loss identified through post-neonatal care pathways, hearing loss is most commonly identified due to school hearing screenings and parental concerns regarding hearing (Dedhia et al., 2013; Watkin & Baldwin, 2011). Once identified, Walker et al. (2014) observed significantly longer delays from hearing loss identification to intervention for children with postnatal hearing loss compared to children who were identified in the newborn period. The same investigation revealed that degree of hearing loss predicted age at follow-up clinical services

for children with postnatally identified hearing loss, such that children with more severe losses received services at younger ages compared to children with milder hearing loss.

Approximately 40% of patients in the Amplification Program at Boston Children's Hospital (BCH) passed their UNHS bilaterally. In the BCH Cochlear Implant Program, 18% of patients with known UNHS outcomes passed in both ears. We designed this study to investigate the pathway to amplification technologies for children who passed their UNHS with the aim of revealing the factors that led to the later identification of children with hearing loss. This study addresses the average age of hearing loss identification and the average time between hearing loss identification and amplification fittings in this population. Additionally, we describe the degree, type, and causes of hearing loss observed. Based on previous studies described above, we hypothesized that most children would be identified through childhood hearing screening programs and that more severe hearing losses would have a shorter time between identification and intervention.

Methods

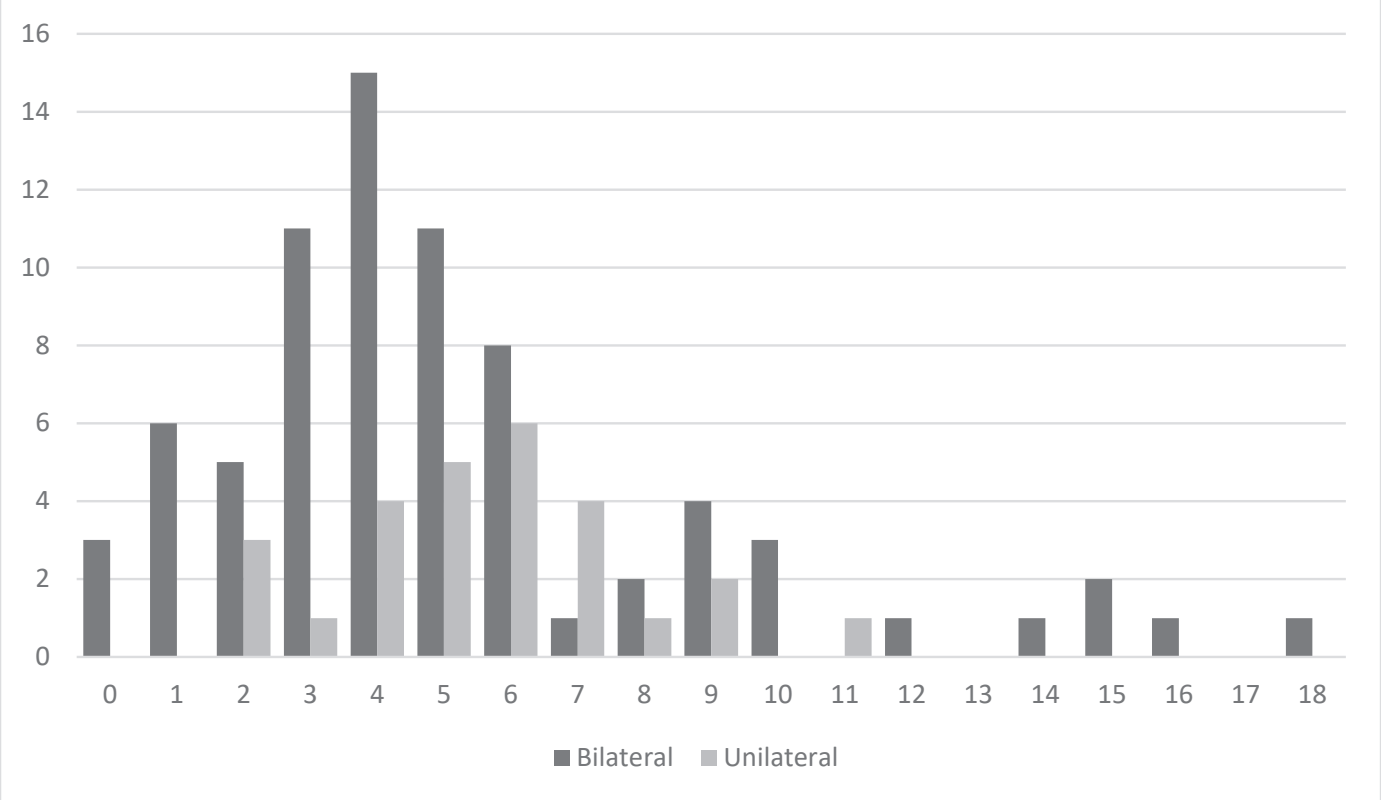
We reviewed medical records of 102 children who passed their UNHS and who are enrolled in the BCH Amplification and/or Cochlear Implant Programs. Medical records were included for review from July 1999 through July 2018. Participants were included in this study if they were (a) identified with hearing loss between 0–22 years of age, (b) had known outcomes of their UNHS, and (c) were users of amplification technologies including hearing aids, cochlear implants, or bone anchored hearing systems. Of the 204 ears, 177 ears were identified with permanent hearing loss and were included in the study. Table 1 indicates the breakdown of participants by sex and by whether the hearing loss was unilateral or bilateral at initial diagnosis.

Table 1
Participant Breakdown by Sex and Number of Ears with Hearing Loss (Unilateral vs. Bilateral)

	Male	Female	Total
Bilateral	37 (36.3%)	38 (37.3%)	75 (73.5%)
Unilateral	16 (15.7%)	11 (10.8%)	27 (26.5%)
Total	53 (52.0%)	49 (48.0%)	102 (100%)

Figures 1 and 2 respectively display the age of identification broken down by laterality of hearing loss and by sex. Note that race/ethnicity data are not reliably coded in the hospital medical record and are not included.

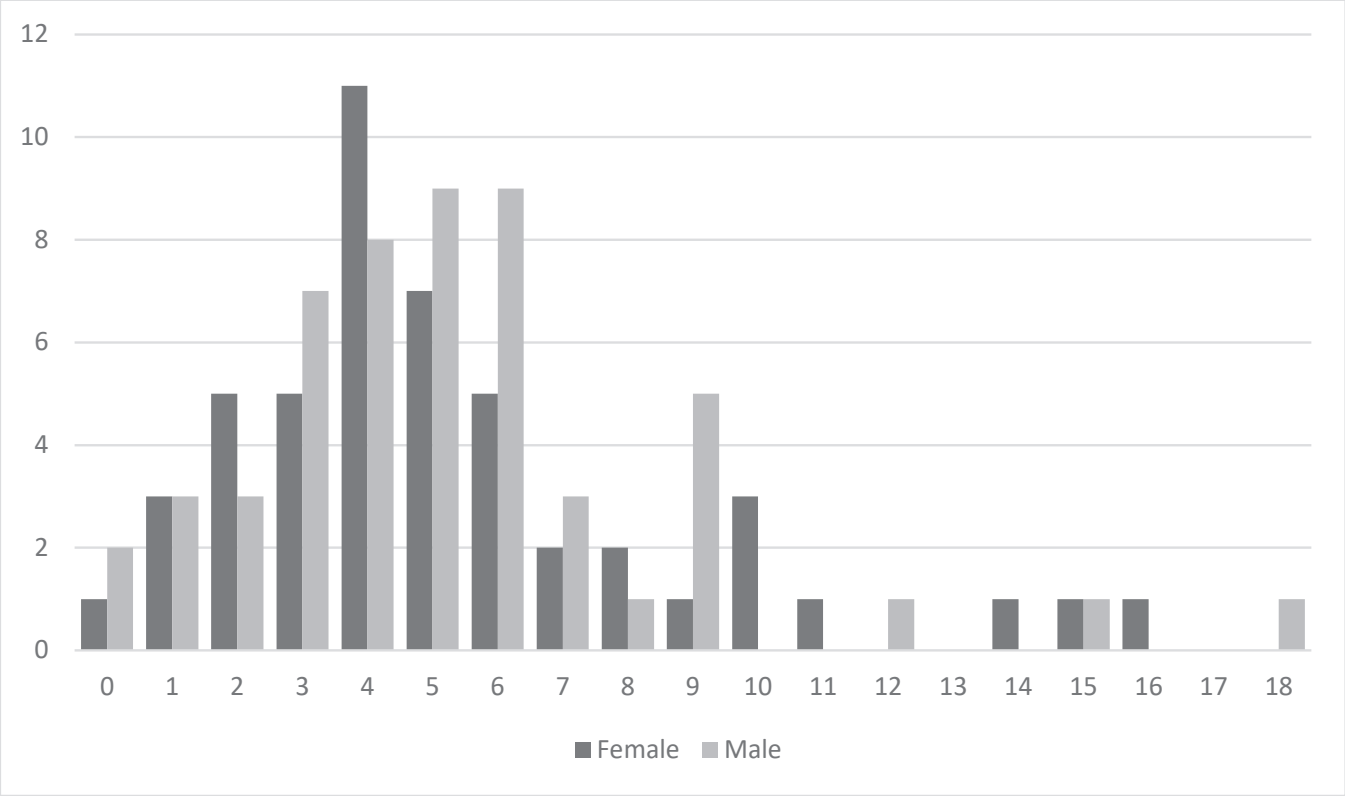
Figure 1
Histogram of Age (in Years) of Identification of Hearing Loss Split by Unilateral Versus Bilateral Hearing Loss



Note. Age of identification was not significantly different between these two groups (t_{78} = -0.6, p = 0.5).

Figure 2

Histogram of Age (in years) of Identification of Hearing Loss Split by Sex of Participant



Note. Age of identification was not significantly different between these two groups (t_{98} = -0.08, p = 0.9).

When reviewing the medical records, we investigated certain criteria to describe the type, degree, and configuration of the hearing losses. The types of hearing loss were determined to be sensorineural, conductive, or mixed. We categorized the patients' hearing loss configurations using the terms flat, rising, sloping, cookie bite, reverse cookie bite, notched, or unconventional. The patients' best threshold degrees and worst threshold degrees were documented to fully capture their hearing loss and to not exclude those with irregular configurations. Additionally, the patients' 2000 Hz pure tone threshold degrees were documented due to the importance of 2000 Hz in speech recognition.

To capture the patients' timeline to amplification technologies, we looked at the month and year of initial hearing loss diagnosis and calculated the years between birth and hearing loss identification to find the average age of identification. We then investigated the month and year of initial hearing aid fitting and calculated the years between hearing loss identification and hearing aid fitting. For patients who use cochlear implants, we documented the date of initiation for their pre-surgical hearing aid trial, if known; if there was no documented hearing aid trial, the date of intervention was marked as the implant surgery date. With this information we were able to calculate the average amount of time between initial hearing loss diagnosis and amplification fitting across all of our patients.

We were also interested in capturing identifiable reasons for the referral for audiological evaluations. These fields included a referred hearing screening at the pediatrician or school, speech-language delay, pediatrician concern, parental concern, suspected or confirmed diagnosis of autism spectrum disorder, or other medical referral from a specialist. These reasons were not mutually exclusive, and, for some patients, more than one reason was selected. In our records, it was not always clear whether the hearing screening was performed at the doctor's office or the school; hence these are combined. Tier 1 and 2 risk factors for childhood hearing loss outlined by the Massachusetts Department of Public Health (Table 2) were investigated as possible predictors for late-onset childhood hearing loss. The risk factor tier indicates when an infant would receive follow-up hearing testing. Infants born with Tier 1 risk factors are recommended to receive a diagnostic ABR by 3 months of age. This appointment is scheduled by the birth hospital prior to discharge. Infants born with Tier 2 risk factors are recommended to receive a diagnostic hearing assessment at 6–9 months of age (Stewart, 2017). This is coordinated by the medical home.

Knowing the etiology of hearing loss was important in the determination of whether the participant's hearing loss was acquired, presumably congenital missed by the UNHS, or delayed-onset congenital. If the etiology of the hearing loss was known, we categorized them as

Table 2

Massachusetts Department of Public Health Tier 1 and Tier 2 Risk Factors for Hearing Loss

Tier 1	Tier 2
• Maternal CMV • Down Syndrome • Cleft lip/palate • Bacterial meningitis • Craniofacial anomalies • Syndromes associated with hearing loss • Perinatal asphyxia • ECMO • Hyperbilirubinemia (> 20 mg/dL bilirubin) • Permanent hearing loss in immediate family • Parental or medical provider concern	• > 10 days mechanical ventilation • ≤ 32 weeks gestational age • < 1500 grams birth weight • Permanent hearing loss in extended family • Herpes, rubella, syphilis, or toxoplasmosis • Head trauma • Ear pits with preauricular tags • Ototoxic medications (> 7 day course in conjunction with loop diuretics) • NICU stay > 5 days

Note. CMV = cytomegalovirus, ECMO = Extracorporeal membrane oxygenation, NICU = Newborn Intensive Care Unit.

genetic, such as connexin-26 or related with a syndrome; anatomical, such as enlarged vestibular aqueduct (EVA); caused by infection, such as cytomegalovirus (CMV); caused by ototoxic medications, such as chemotherapy; or due to another cause. We further wanted to investigate whether the patients had a coexisting diagnosis related to neurologic status, such as Autism Spectrum Disorder or intellectual disability.

Results

Approximately half (52.0%) of hearing losses were sensorineural in nature. Conductive hearing loss comprised 30.5% of hearing losses in our cohort and the remaining 17.5% of hearing losses were mixed in nature. Figure 3 illustrates degree of hearing loss for the 177 ears in the study based on the 2000 Hz threshold, the best threshold, and the poorest threshold. For 63.8% of ears, the 2000 Hz threshold at hearing loss identification was in the normal hearing or mild hearing loss range. 2000 Hz thresholds were observed in the moderate or moderately-severe hearing loss range for 26.0% of ears and in the severe to profound range for the remaining 10.1% of ears. At initial identification, more than 80% of ears had at least one pure-tone threshold in the normal to mild loss range and more than 60% of ears had at least one pure-tone threshold in the moderate to profound range. The majority (90.2%) of participants wore hearing aids; 8.8% used cochlear implants exclusively or as a bimodal solution. The rest of the participants (1%) used a bone-anchored device.

Etiologies of hearing loss varied greatly across participants. Unknown etiology accounted for 37.3% of participants, often despite the use of temporal bone imaging and genetic testing under management by an otolaryngologist. Acquired conditions accounted for 31.4% of hearing loss, including conditions such as chronic otitis media (53.1%), cholesteatoma (25.0%), or ototoxicity (18.8%). Syndrome related losses accounted for 16.7% of participants, of which the most common was Down syndrome (58.8%). Enlarged vestibular aqueducts accounted for 10.8% of participants. Connexin-26 genetic mutations accounted for 3.9% of participants. Congenital CMV (cCMV) accounted for 2.9% of participants. Incidentally, 5.9% of participants had a comorbid diagnosis of Autism Spectrum Disorder and 4.9% of participants had a comorbid diagnosis of intellectual disability.

Table 2 summarizes hearing loss identification and amplification fitting timelines by type of hearing loss. The average age of hearing loss identification was 5.7 years (SD = 3.6 years). Group means for type of hearing loss were evaluated for differences using one-way Analysis of Variance (ANOVA) testing. No significant difference for age of hearing loss identification was observed based on hearing loss type, $F(2, 174) = 2.79$, $p = 0.06$. Once identified with hearing loss, the average time from diagnosis to amplification fitting was 2.0 years (SD = 2.8 years). A significant main effect of type of hearing loss was observed for the time from hearing loss diagnosis to amplification fitting, $F(2, 174) = 6.45$, $p < 0.01$. A Tukey test for multiple comparison of means, using a 99% confidence level, revealed that children with sensorineural hearing loss had a significantly shorter time from hearing loss diagnosis to amplification fitting than children with conductive hearing loss ($p < 0.01$). No difference was observed when comparing children with mixed hearing loss to those with either sensorineural ($p = 0.15$) or conductive hearing losses ($p = 0.63$).

Tier 1 and 2 risk factors for hearing loss were investigated as possible predictors for late-onset childhood hearing

loss. At least one Tier 1 or 2 risk factor for hearing loss in the neonatal period was present for 40.2% of our cohort. The average age of hearing loss identification for those with at least one risk factor was 5.6 years (*SD* = 4.2 years) compared to 5.8 years (*SD* = 2.8 years) for those without a risk factor. A Tier 1 risk factor for hearing loss was present in 24.5% of participants. The most frequent Tier 1 risk factor was an immediate family history of hearing loss (*n* = 9) followed by cCMV (*n* = 3). 19.6% of participants had a Tier 2 risk factor for hearing loss. Among Tier 2 risk factors, the most commonly observed was a neonatal intensive care unit stay of greater than 5 days (*n* = 11). Six participants had an extended family history of hearing loss. Five participants were given ototoxic medication in the neonatal period. Five participants had a gestational age of less than 32 weeks.

Table 3 shows reasons for audiological referral by age group. Approximately 1 in 4 patients did not have an identifiable reason for audiological evaluation. For children older than 3, a hearing screening was the primary reason for referral for diagnostic hearing testing. For children under age three, a speech-language delay was the most common reason for referral leading to identification of a permanent, postnatal hearing loss. A referral from a specialist (e.g., geneticist, developmental pediatrician, cardiologist) led to diagnosis for 22.1% of patients.

Table 3

Reason for Referral for Audiological Evaluation by Age Group (Age of Diagnosis)

	Infant/Toddler (0-3 Years)	Preschool (4-5 Years)	Early School (6-10 Years)	Later School (11+ Years)	All Ages
n	25	31	39	7	102
Referred Screening	0.0%	38.1%	35.5%	57.1%	27.5%
Speech-Language Delay	37.9%	22.9%	16.1%	14.3%	25.5%
Referral from Specialist	27.6%	17.1%	22.6%	0.0%	20.6%
Parent Concern	17.2%	22.9%	9.7%	14.3%	16.7%
Primary Care Provider Referral	0.0%	5.7%	6.5%	14.3%	4.9%
No Known Reason	34.5%	14.3%	25.8%	14.3%	23.5%

ANOVA was performed to determine whether severity of hearing loss was related to identification of hearing loss. The analysis indicated no significant relationship between severity of hearing loss, either based on best hearing threshold or best threshold at 2 kHz, and number of months between identification of hearing loss and first fitting with amplification. The average time between identification and fitting was 25.5 months (*SD* = 34.9 months).

Discussion

The implementation of the UNHS has made a significant impact on early hearing detection and intervention. However, UNHS cannot stand alone in detection of childhood hearing loss. As observed by Walker et al. (2014), this study indicates that children identified with hearing loss through post-natal pathways experience long delays between hearing loss identification and the implementation of hearing loss interventions.

Documented risk factors for hearing loss fall into two tiers, which then determines the timeline for initial diagnostic testing. Children with a Tier 1 or Tier 2 risk factor requiring diagnostic testing may have not developed hearing loss by the time of initial appointment despite the possibility of later-onset hearing loss. This supports routine monitoring and screening of hearing to document any changes in a prompt manner. However, the risk factors do not capture every child who may develop a delayed-onset congenital or acquired hearing loss. The list of risk factors increases the number of children being diagnostically monitored for potential hearing loss in childhood but cannot encompass or predict all children that will require audiological evaluations. This is supported by our cohort as children with and without risk factors were included.

Children who pass their UNHS, but experience signs of hearing loss during childhood must be appropriately referred to an audiologist trained to evaluate hearing in pediatric patients. The most frequent catalyst for hearing loss identification in our cohort was referring on a routine hearing screening, consistent with published data (Dedhia et al., 2013; Watkin & Baldwin, 2011). Our data demonstrate the importance and necessity of school- and primary care provider-based hearing screenings in the process of identifying and treating children with hearing loss. There may have been delays that we could not capture in this study. For instance, if a patient referred their

school screening and then went to their physician for a repeat screening and then was referred to our clinic, this may have caused added delay to the time of diagnosis.

Additionally, our data show the importance of referring children with speech delays for hearing evaluations, even if they passed the newborn hearing screening. This was the primary route to identification for children under 3 years of age. Speech-language pathologists and Early Intervention staff should not assume hearing is normal if a child passed their newborn hearing screening and should include a hearing test as part of the work-up when a child is exhibiting speech and language delays.

We found that the average duration between diagnosis and fitting is greater than one year. This suggests there is a lesser sense of urgency for these older children than there is for children who refer newborn hearing screening and are fit with amplification by 6 months. Boston Children's Hospital does abide by the EHDI 1-3-6 guideline for newborns, it being tied to a state mandate. These data suggest that Boston Children's may benefit from an initiative to fit later-diagnosed children with hearing aids within 3 months of diagnosis.

Our data also demonstrate a relative greater average time from diagnosis to fitting of children with conductive hearing losses. This is not surprising given the time it takes to evaluate candidacy for the greater number of medical and surgical treatments available for conductive hearing loss. Future research may evaluate whether efforts to quickly determine the etiology of conductive hearing loss may lead to earlier fitting of amplification. Future research may evaluate whether there are benefits to fitting amplification synchronously with the medical evaluation process instead of waiting for the physicians to complete their assessments prior to fitting amplification. This finding raises the question as to whether the addition of new options for medically treating sensorineural hearing loss (e.g., gene therapy) may increase time between diagnosis and fitting in the coming years.

Conclusions

It is critical to reinforce the importance of regular childhood hearing screenings through later school-age years. These efforts provide opportunities for earlier identification of childhood hearing loss allowing for earlier intervention options. Family members, educational professionals and clinicians alike should be aware of and pay attention to signs of childhood hearing loss, such as speech-language delay, academic difficulties, and increased exhaustion at the end of a school day to ensure proper referrals lead to early diagnosis. Pediatric medical centers should ensure that, once diagnosed with hearing loss, older children are being fit with amplification with as little delay as possible, similar to the 1-3-6 guidelines for newborns.

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