Progress in Documented Early Identification and Intervention for Deaf and Hard of Hearing Infants: CDC’s Hearing Screening and Follow-up Survey, United States, 2006–2016

Krishnaveni Subbiah, MPH1,3
Craig A. Mason, MPH2
Marcus Gaffney, MPH3
Scott D. Grosse, PhD3

1Oak Ridge Institute for Science and Education, Oak Ridge, Tennessee
2University of Maine College of Education and Human Development, Orono, Maine
3National Center on Birth Defects and Developmental Disabilities, Centers for Disease Control and Prevention, Atlanta, Georgia

Abstract: The national EHDI 1-3-6 goals state that all infants should be screened for hearing loss before 1 month of age; with diagnostic testing before 3 months of age for those who do not pass screening; and early intervention (EI) services before 6 months of age for those with permanent hearing loss. This report updates previous summaries of progress on these goals by U.S. states and territories. Data are based on the Hearing Screening and Follow-up Survey (HSFS) conducted annually by the Centers for Disease Control and Prevention for the years 2006–2016. Trends were assessed using 3-year moving averages, with rates of newborns lost to follow-up or lost to documentation (LTF/D) also examined. During this period, the percentage of infants screened before one month increased from 85.1% to 95.3%, while the percentage receiving diagnostic testing before three months increased from 19.8% to 36.6%, and the percentage of infants identified with permanent hearing loss enrolled in early intervention (EI) before six months increased from 25.1% to 47.2%. Percentages of infants who ultimately received screening, diagnostic testing, and early intervention services – regardless of timing – were higher. During this period, LTF/D declined from 42.1% to 31.3% for diagnostic testing, and 39.4% to 20.3% for EI services. Diagnoses of hearing loss recorded increased from 0.9 to 1.7 per 1,000 infants screened, likely reflecting improved data.

Acronyms: AAA = American Academy of Audiology; ASHA = American Speech-Language-Hearing Association; CDC = Centers for Disease Control and Prevention; DHH = deaf or hard of hearing; EI = early intervention; DSHPSHWA = Directors of Speech and Hearing Programs in State Health and Welfare Agencies; EHDI = Early Hearing Detection and Intervention; EHDI-PALS = EHDI Pediatric Audiology Links to Services; HL = hearing loss; HRSA = Health Resources and Services Administration; HSFS = Hearing Screening and Follow-up Survey; LTF/D = lost to follow-up or lost to documentation; NCHAM = National Center for Hearing Assessment and Management; NICHQ = National Institute for Children’s Health Quality; UNHS = universal newborn hearing screening

Acknowledgements: This report is based on data reported by jurisdictional EHDI programs in U.S. states and the District of Columbia.

Correspondence concerning this article should be addressed to: Marcus Gaffney, MPH, Division of Human Development and Disability, National Center on Birth Defects and Developmental Disabilities, Centers for Disease Control and Prevention, Atlanta, Georgia. Phone: 404-498-3031; Email: MGaffney@cdc.gov

Introduction

Congenital hearing loss (HL) affects 1.5 to 3 per 1,000 infants in the United States (Grosse et al., 2017). Children who are born deaf or hard of hearing (DHH) are at increased risk for delays in nonverbal communication skills and speech and language development (Caskey & Vohr, 2013). In particular, in the absence of universal newborn hearing screening (UNHS), many children are not diagnosed as DHH until 2 years of age or later (Elssmann, Matkin, & Sabo, 1987), at which point delays in language development are more difficult to remediate (Yoshinaga-Itano & Apuzzo, 1998; Yoshinaga-Itano, Sedey, Coulter, & Mehl, 1998). Early identification facilitated by UNHS accompanied by prompt initiation of early intervention (EI) services has been shown to directly benefit infants who are DHH by reducing deficits in their language and vocabulary (Kennedy et al., 2006; Nelson, Bougatsos, & Nygren, 2008; Vohr et al., 2011; Yoshinaga-Itano, Sedey, Wiggins, & Chung, 2017).

We used data collected through the Centers for Disease Control and Prevention (CDC) Early Hearing Detection and Intervention (EHDI) Hearing Screening and Follow-
up Survey (HSFS) for the years 2006–2016 to assess progress towards meeting the national “1-3-6” EHDI goals or benchmarks. The goals, which together constitute the 1-3-6 EHDI plan, have been agreed upon by EHDI partners since the early 2000s: (a) all infants be screened for HL before 1 month of age; (b) those not passing the screening receive diagnostic testing before 3 months of age; and (c) those confirmed as DHH begin receiving appropriate early intervention services before 6 months of age (CDC, 2003; White, 2003; White, Forsman, Eichwald, & Munoz, 2010). This report updates previous summaries of HSFS data by including additional survey years and using a different analytical approach (Gaffney, Eichwald, Gaffney, Alam, & CDC, 2014; Gaffney, Green, & Gaffney, 2010; Williams, Alam, & Gaffney, 2015).

Previous studies have demonstrated that the vast majority of U.S. infants are screened for HL soon after birth. From 1996 to 2000, the estimated percentage of U.S. infants screened for hearing loss prior to hospital discharge increased from roughly 10% to roughly 50% (White, 2003). Using annual state screening estimates collected by the Directors of Speech and Hearing Programs in State Health and Welfare Agencies (DSHPHSHA), screening rates in participating states increased from 53% in 2000 to 92% in 2003 (Green, Gaffney, Devine, & Grosse, 2007). However, although almost all (> 97%) U.S. infants now undergo hearing screening soon after birth, those who fail to pass screening do not necessarily receive timely diagnostic evaluations or timely intervention services once diagnosed with permanent HL (Grosse et al., 2017). The key challenges facing EHDI programs are to increase the percentage of infants who meet the 3-month diagnostic evaluation and 6-month early intervention goals and to document that those goals are met.

This can be challenging because there are multiple, diverse reasons why the 1-3-6 goals are not met. Parents face competing demands on their time and resources as well as possess different levels of confidence in the healthcare system. Those who have low resources and/or trust levels may be less likely to keep appointments or respond as expected by providers. They may be classified as refusing services or lost to follow-up (LTF) or they may go on to simply receive services at a later age. However, greater efforts by providers or program staff may increase the likelihood of the infants in those families receiving timely services. In other cases, families may be engaged and supportive of follow-up, yet be stymied by external factors, such as reduced access to services—either due to limited availability of diagnostic or EI providers, or lack of insurance coverage. In addition, infants may meet the goals but that information is not reported by service providers to the EHDI program, resulting in loss to documentation (LTD; Mason, Gaffney, Green, & Grosse, 2008). In practice, it can be difficult or impossible to distinguish cases of LTD from LTF, and so the two are often examined together (LTF/D).

Method

In 2007, CDC began using the HSFS to collect annual aggregate EHDI data from states based on births from the calendar year two years prior (i.e., all infants born during 2005) to ensure that they had sufficient time to complete the EHDI process. This report uses HSFS data submitted for 2006–2016 to describe the progress of EHDI programs in the 50 states and District of Columbia toward the early identification and treatment of DHH infants, including meeting the 1-3-6 goals. The number of jurisdictions submitting data varied each year due to the inability of some jurisdictions to provide empirical estimates for one or more reporting years.

To better assess progress, jurisdictions were assigned a population weight based on the total number of occurrent live births each year. Trends in meeting the three goals of EHDI were assessed by determining the percentage of infants reported as (a) screened among the total reported occurrent births; (b) having received diagnostic testing among the total reported as not passing the hearing screening; and (c) enrolled in EI among the total reported as diagnosed with permanent HL. Percentages were calculated for screening, diagnostic, and EI services, both overall, regardless of timing, and in accordance with the 1-3-6 goals.

Finally, progress in identification was determined by comparing the percentage of infants classified as LTF/D for diagnosis and EI to the prevalence of HL in each year. CDC defines LTF/D as not having received or not documented as having received follow-up diagnostic and intervention services. Infants are classified as LTF/D if the EHDI program was unable to contact their family, or if the child’s status was otherwise unknown. Cases were also classified as LTF/D if the parents/family were contacted by the EHDI program but unresponsive—choosing not to engage in the diagnostic or early intervention systems—for reasons (possibly those described previously) not conveyed to the EHDI program. To account for year-to-year fluctuations, a 3-year moving average of these weighted percentages was calculated.

Results

The percentage of infants screened by one month of age increased from an average of 85.1% during 2006–2008 to 95.3% during 2014–2016 (see Table 1). This change reflects a two-thirds reduction in the number of children not screened in the first month of life (from 14.9% to 4.7%). When the time frame is expanded to include infants not screened before one month of age, the overall percentage of infants screened remained consistently high, increasing slightly from 97.0% to 98.4%.
Table 1
Weighted Percentages of Documented Infants Receiving Screening and Diagnostic Testing and Enrolled in Early Intervention Services, (CDC EHDI HSFS*, United States, 2006-2016†§)

<table>
<thead>
<tr>
<th>Data Point†</th>
<th>EHDI Stage &amp; 1-3-6 Goal</th>
<th>Screening¶</th>
<th>Diagnostic Testing¶</th>
<th>Screening¶</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Total Births</td>
<td>Total % Screen</td>
<td>By 1 month**</td>
<td>Not Pass Screen N</td>
</tr>
<tr>
<td>2007‡</td>
<td>3,388,056</td>
<td>97.0%</td>
<td>85.1%</td>
<td>68,212</td>
</tr>
<tr>
<td>2008</td>
<td>3,648,433</td>
<td>97.6%</td>
<td>87.4%</td>
<td>65,588</td>
</tr>
<tr>
<td>2009</td>
<td>3,819,961</td>
<td>96.0%</td>
<td>92.1%</td>
<td>65,374</td>
</tr>
<tr>
<td>2010</td>
<td>3,913,705</td>
<td>98.2%</td>
<td>94.4%</td>
<td>63,841</td>
</tr>
<tr>
<td>2011</td>
<td>3,728,962</td>
<td>97.6%</td>
<td>95.1%</td>
<td>62,346</td>
</tr>
<tr>
<td>2012</td>
<td>3,547,267</td>
<td>97.0%</td>
<td>94.6%</td>
<td>60,404</td>
</tr>
<tr>
<td>2013</td>
<td>3,603,806</td>
<td>97.0%</td>
<td>94.4%</td>
<td>59,872</td>
</tr>
<tr>
<td>2014</td>
<td>3,732,653</td>
<td>97.8%</td>
<td>95.3%</td>
<td>61,931</td>
</tr>
<tr>
<td>2015§</td>
<td>3,859,270</td>
<td>98.4%</td>
<td>95.3%</td>
<td>63,718</td>
</tr>
</tbody>
</table>

Note. Annual data for individual jurisdictions is available at www.cdc.gov/nchbidd/earrhi/earlyinterventions/earlyinterventions-data.html; CDC = Center for Disease Control and Prevention; EHDI = Early Hearing Detection and Intervention; EI = Early Intervention; HSFS = Hearing Screening and Follow-up Survey.

* Hearing Screening and Follow-up Survey (HSFS) is an annual, voluntary, Office of Management and Budget approved data survey (OMB No. 0920-05AA) administered by CDC.
§ This analysis does not include U.S. territories that may have responded to the survey. Data are analyzed as population weighted, three-year moving averages, with 2007 as the first data point (the average of 2006-2008 data) and 2015 as the final data point (the average of 2014-2016 data).
¶ Percent screened and screened by 1 month of age, among total live births as reported in HSFS.
†† For 2012 and 2013 annual data, one jurisdiction was excluded for the inability to report complete data for the number of infants screened by 1 month of age, due to temporary reporting issues with their data system. This jurisdiction was able to report all other measures.
‡‡ Percent diagnostically tested and diagnostically tested by 3 months of age, among total newborns not passing hearing screenings.
§§ For 2015 annual data, one jurisdiction was excluded for the inability to report complete data for the number of infants diagnosed by 3 months of age, due to temporary reporting issues with their data system. This jurisdiction was able to report all other measures.
¶¶ Percent enrolled in EI and enrolled by 6 months of age, among total diagnosed with permanent HL.

Figure 1. Prevalence of HL* and LTF/D for Diagnostic Testing† and Enrollment in Early Intervention Services§ (CDC EHDI HSFS* United States, 2006-2016**)

* Prevalence of HL is calculated using the following formula: (# Diagnosed with HL / # Total Screened) * 100.
† Percent LTF/D for Diagnostic Testing is calculated using the following formula: (# No Diagnostic Data due to Unable to Contact + # No Diagnostic Data due to Parents/Family Contacted but Unresponsive + # No Diagnostic Data Due to Unknown Reason) / # Total Not Passed Screening * 100.
‡ Percent LTF/D for Enrollment in EI is calculated using the following formula: (# No EI Data due to Unable to Contact + # No EI Data due to Parents/Family Contacted but Unresponsive + # No EI Data due to Unknown Reason) / # Total Diagnosed with HL) * 100.
§ Hearing Screening and Follow-up Survey (HSFS) is an annual, voluntary, Office of Management and Budget approved data survey (OMB No. 0920-05AA) administered by CDC.
** This analysis does not include U.S. territories that may have responded to the survey. Data are analyzed as population weighted, three-year moving averages, with 2007 as the first data point (the average of 2006-2008 data) and 2015 as the final data point (the average of 2014-2016 data).
The percentage of infants who did not pass screening and who received diagnostic testing by three months of age increased from 19.8% during 2006–2008 to 36.6% during 2014–2016 (see Table 1). Including those diagnosed after the 3-month target date, the overall percentage of infants who did not pass screening but who received diagnostic testing nearly doubled—increasing from 30.2% to 58.6%. These increased numbers were also associated with reductions in the percentage of infants who were LTF/D, another indicator of progress. An average of 42.1% of infants who did not pass screening in the 2006–2008 period were classified as LTF/D for diagnostic testing, which declined to 31.3% in the 2014–2016 period (see Figure 1).

Finally, during the same timeframe, the percentage of DHH infants enrolled in EI before six months of age increased from 25.1% to 47.2%. When the time frame was expanded to include those who were enrolled in EI but did not meet the six-month goal, the percentages of DHH infants reported as enrolled in EI increased from 54.1% to 67.9% (see Table 1). The corresponding decrease in LTF/D for enrollment in EI was greater, dropping from 39.4% to 20.3% — a nearly fifty percent reduction in LTF/D (Figure 1).

Given these changes, it was not surprising that diagnoses of infants as DHH increased from 0.9 to 1.7 per 1,000 infants screened between these data points (see Figure 1). This increase likely reflects the improvement in early identification along with decreased LTF/D for diagnosis and EI. Although there continues to be jurisdictional-level variation in early identification and enrollment in EI (see Figure 2), these overall trends reflect progress in the reporting and documentation of recommended services among EHDI programs.

Figure 2. Weighted Percentages of Infants Receiving Diagnostic Testing and Enrolled in Early Intervention Services Among the 5 Lowest Performing Jurisdictions*, the National Average†, and 5 Highest Performing Jurisdictions‡, for the 2007 versus 2015 data point (CDC EHDI HSFS¶, United States, 2006-2016**).
Substantial progress has been made since 2007, especially in the delivery and reporting by providers to EHDI programs of diagnostic testing before age 3 months and of enrollment of DHH infants in EI before age 6 months. However, the rate of overall progress has slowed since 2011 and there is variation in progress between jurisdictions. In particular, the fluctuating trend of LTF/D rates for early diagnoses and the recent plateau of LTF/D rates for EI indicate that challenges remain. For the most recent data points, 2011–2015, the percentages of infants reported as completing the three EHDI stages and meeting the 1-3-6 goals show smaller yearly improvements compared to 2007–2010 (see Table 1). There are also wide discrepancies at the jurisdictional level in early identification and enrollment in EI services, with some programs performing well above the national average, whereas others have not been as successful (see Figure 2). Reasons for some states having less success in meeting the 1-3-6 goals could potentially include lack of comprehensive follow-up strategies to ensure receipt of diagnostic and EI services; reductions in resources available to some programs; and differences in state laws, regulations, or policies. Differences in patterns over time across states could also reflect changes in reporting, data systems, reporting capacity, and best practice policies for audiologists and EI providers.

The relatively low absolute percentages of children documented as receiving timely diagnosis and initiation of EI highlight the need for continued efforts to ensure all DHH infants are identified early and able to reach their full potential. The observed variability in progress by goal and across states can be used to focus additional efforts to improve the delivery and documentation of essential EHDI follow-up diagnostic and EI services and to reduce variability in access to needed services (Liu, Farrell, MacNeil, Stone, & Barfield, 2008).

Calculations using population-weighted, 3-year moving averages allow for a more standardized comparison of data that has varying respondents between years. Nonetheless, the findings in this report are subject to at least four limitations. First, the use of moving averages minimizes fluctuations associated with random variation, which can indicate no overall trend despite large differences between adjacent years. Second, some jurisdictions did not report data for one or more years. Third, the HSFS is a voluntary survey and although there are standardized data definitions, the data reported are subject to different interpretations. For example, the question of what constitutes an “in process” diagnostic evaluation was clarified and refined to improve consistency. Fourth, incomplete reporting of services could understate the receipt of services and overstate rates of LTF/D.

Despite smaller improvements at the national level in recent years, some high performing state EHDI programs have shown continued progress through implementation of innovative strategies. For example, the Kentucky and Louisiana EHDI programs have reported that scheduling follow-up appointments at the time a hearing screening is not passed prior to hospital discharge is associated with increased receipt of follow-up services, and the programs encourage and track this practice (Lester, 2017; Ye et al., 2014). Louisiana has also created a system for the routine linkage of Medicaid data, which is used by the EHDI program to verify initiation of follow-up and improve communication between EHDI coordinators and clinical providers (Tran et al., 2016). However, relatively few children can be tracked through that linkage. During 2012–2013, of 682 infants classified as LTF/D in Louisiana, 57 had Medicaid records, and 38 of those had records that could be retrieved and matched. Of those 38 infants, 25 were reclassified as having received follow-up services (Tran et al., 2016).

The EHDI program in Georgia has shown that texting parents after an unsuccessful attempt to contact them by telephone can improve families’ response to a reminder of audiological follow-up (Hermanns, Currie, LaVell, & Lo, 2016). The program recommends incorporating texting into the follow-up protocol for all EHDI programs. Other states have focused efforts on encouraging pediatric audiologists to report diagnostic results and provide technical assistance with electronic reporting of diagnostic results.

In 2011, Wisconsin started providing varying levels of assistance to families, hospitals, and providers to reduce LTF/D rates (Wisconsin Sound Beginnings, 2016). These changes included in-home and in-community, infant-specific outreach to families reluctant to or unable to access follow-up services, and training and technical support to health care systems. These state-implemented strategies involved a team approach including families, state EHDI staff, and providers. The various initiatives helped further improve the receipt of follow-up services. Among 1,819 infants who did not pass initial screening in Wisconsin in 2015, 138 never received further services, mostly because parents refused (n = 38) or were unresponsive (n = 49); just 9 infants were LTF/D. The primary remaining challenge in Wisconsin is assuring timely intervention. Of 133 infants diagnosed with permanent hearing loss, 122 were referred to EI, but just 44 were enrolled by 6 months of age.

One strategy that might help reduce LTF/D is to more closely integrate EHDI activities into other newborn health and development services. For example, in 2012 to 2014, Ohio tested an intervention that involved a partnership between EHDI and WIC, in which WIC infants who did not pass initial newborn hearing screening received an outpatient rescreen at their WIC office (Hunter et al., 2016). Combining co-location of services with timely scheduling and contact with families reduced LTF/D rates from 33.3% to 9.6%, while the mean age of diagnosis dropped from 68 days to 34.8 days for children in the study.
In addition to LTF that reflects children not receiving a diagnostic evaluation or services, LTD can occur if providers fail to report information to their state EHDI program. For example, 13.6% of a national sample of 1,024 pediatric audiology facilities indicated that they reported less than two-thirds of their results to their state EHDI program—with 8.6% reporting none of their results (Chung, Beauchaine, Grimes, et al., 2017). Furthermore, among facilities that do report data, 14.5% indicated that they did not report normal hearing results. This gap in reporting and documentation will inevitably impact overall LTF/D rates and lead to underestimating true EHDI program coverage and impact.

Beyond state-level strategies designed to reduce LTF/D, at the national level CDC, the National Center for Hearing Assessment and Management (NICHAM), the American Speech-Language-Hearing Association (ASHA), the American Academy of Audiology (AAA), and other partners collaborated on the creation of EHDI Pediatric Audiology Links to Services (EHDI-PALS). EHDI-PALS is a web-based, geocoded national directory of facilities that offer pediatric audiology services to children who are younger than five years of age (Chung, Beauchaine, Hoffman, et al., 2017). EHDI-PALS is designed to help parents find pediatric audiologists with the training and tools necessary to provide evaluation services for young children, and who also report data to state EHDI programs. Parents can enter the age and other relevant information about their child and, based on their zip code, be given a highly detailed list of facilities in their area or region.

Also at the national level, the National Institute for Children’s Health Quality (NICHQ), with support from the Health Resources and Services Administration (HRSA), conducted a Learning Collaborative from 2010 to 2013 to seek out ways to reduce LTF/D. In 2016 it published an Action Kit for audiologists that summarized lessons from the Collaborative (NICHQ, 2016). For example, NICHQ emphasizes the importance of communication with families prior to the first diagnostic appointment regarding what to expect and how to prepare their infant as well as the logistics of getting to the appointment to reduce the frequency of “no-show” appointments. EHDI programs can partner with peer support organizations such as Hands & Voices to facilitate the communication process and hopefully reduce LTF/D at the diagnostic evaluation stage, although we are not aware of formal evaluations.

Conclusion

Meeting the 1-3-6 EHDI goals helps DHH infants improve vocabulary outcomes and minimizes developmental delays that can last a lifetime. Although screening by 1 month of age is necessary and is routinely occurring, it is not sufficient to improve outcomes without timely diagnosis and enrollment in EI services. Although progress in the receipt of diagnostic testing and EI has been made, as illustrated in this report, further progress will require strengthening current practices. Continued efforts in these areas will help ensure all infants who are DHH are identified early while supporting improved developmental outcomes.

References


Engaging Parents in System Design to Reduce Loss to Follow-Up

Dana Yarbrough, MA
Kathleen Watts, MA
Daphne Miller, BS
Susan J. Murdock, PhD

1Virginia Department of Health
2Virginia Commonwealth University

Abstract: This article provides an overview of Virginia’s 1-3-6 Family Educator Project involving the induction and integration of parents of children who are deaf or hard of hearing as leaders in systems change, and the investment of Virginia’s Early Hearing Detection & Intervention program in the project. The article describes the role of the 1-3-6 Family Educators and their success in collaborating with audiologists and hospitals, helping parents get timelier rescreenings of infants who did not pass the newborn hearing screening, and providing peer-to-peer support to parents. Details are provided about the 1-3-6 Family Educator Project to allow other states to consider replicating Virginia’s efforts to empower and support parents as an integral part of system change.

Key Words: EHDI, deaf or hard of hearing, family involvement, family support, leadership

Acronyms: CFI = Center for Family Involvement; CYSHCN = children and youth with special health care needs; DCFH = Division of Child and Family Health; EHDI = Early Hearing Detection & Intervention; ENT = Otolaryngologist (Ear, Nose, Throat Doctor); FE = Family Educator; F2F = Family to Family Network of Virginia; GBYS = Guide By Your Side©; LTF = loss to follow-up; OFHS = Office of Family Health Services; VDH = Virginia Department of Health; VEHDIP = Virginia’s Early Hearing Detection and Intervention program; VISITS = Virginia Infant Screening and Infant Tracking System.

Correspondence concerning this article should be addressed to: Daphne Miller, EHDI Coordinator, Virginia Department of Health, daphne.miller@vdh.virginia.gov or to Dana Yarbrough, Director, Center for Family Involvement, dvyarbrough@vcu.edu

Introduction

There is an adage in the disability field of “nothing about us, without us” that speaks to the conviction that people with disabilities and their families know what is best for them when planning for their personal supports and services and as stakeholders in designing the systems that support them (Bartha & Smith, 2017; Charlton, 2000). This sentiment of end-users contributing as active participants in systems design is not new. The result of their participation yields richer understanding of a system from different perspectives, helps organizations build practical empathy—a mindset of serious listening—that influences its design, and provides invaluable feedback on what works and what doesn’t (Sloan, 2016; Young, 2015).

Early Hearing Detection and Intervention (EHDI) programs have a history of including parents of children who are deaf or hard of hearing on their advisory groups and sometimes as staff assisting with follow-up from newborn hearing screening or parent involvement issues. However, true family engagement goes beyond activities to a process of induction and integration. Pulling family members from the sidelines to a seat at the table in a way that is not tokenism takes demonstrating the values for authentically engaging parents and for co-powering with parents. It requires commitment to transparency about the results we find from their work and pledging to provide funding that supports parents paid as professionals. It also takes acknowledging that there are systemic barriers (Yarbrough, 2017a; Frank, 2016). In the case of EHDI programs, the barriers may perpetuate loss to follow-up.

Parents of children diagnosed as deaf or hard of hearing are information seekers, not only when they first receive their child’s diagnosis, but throughout their child’s life. From the beginning, parents and caregivers have to learn about communication options including communicating with their family and others. They also must decide how to help their child fit into society. All these decisions are made while parents are encountering new systems and dealing with often-difficult funding streams. Parents want their children to have the same opportunities as other children, though there may be challenges, so they develop a level of advocacy. As these parents become advocates for their own children, many develop a desire to help and support other parents of children with hearing loss. They may also aspire to help others avoid some of the more challenging
experiences their family encountered. And, some of these parents want to share their stories and ideas with policymakers to improve the system for those that follow.

Meaningful relationships, shared leadership, and power are at the core of genuine parent engagement. The process of family engagement called induction occurs when families move from the periphery to the center of the system. They begin to take notice, speak up, and intervene in ways that change the nature of the parental relationships with the system. Rather than viewing professionals as final authorities, families begin to see them as partners. In addition, professionals and systems benefit from viewing parents as assets, not liabilities. This takes letting go of the established agenda and creating a shared agenda with families. This takes co-powering. When we co-power, we acknowledge the power families innately carry—power that too often has been dismissed. Co-powering recognizes the power of families in telling their own stories. Co-powering shifts perspectives about what families are capable of doing so they are seen as assets and professionals meet them where they are (Vargas, 2008; Yarbrough, 2017b).

This article provides an overview of the innovative use of parents of children who are deaf or hard of hearing as leaders in systems change within Virginia’s EHDI program, the collaboration with a statewide family-led program (Center for Family Involvement) which facilitated the induction and integration of these parents, and the Virginia EHDI program’s perspective on this investment.

**Virginia’s EHDI Program**

Virginia’s Early Hearing Detection & Intervention program (VEHDIP) resides within the Virginia Department of Health’s Division of Child and Family Health (DCFH) in the Office of Family Health Services (OFHS). The DCFH operates numerous programs serving the maternal and child health populations including, but not limited to, the children and youth with special health care needs (CYSHCN) programs, home visiting, newborn blood spot screening, and child development services. DCFH is the hub of the Title V Block Grant.

The VEHDIP is committed to the 1-3-6 national EHDI goals—all newborns receive a hearing screening by 1 month of age; all newborns who do not pass their screen receive a diagnostic audiological evaluation by 3 months of age; and all infants with hearing loss receive early intervention services by 6 months of age. VEHDIP staff follow-up with parents during each stage in the EHDI process: screening, diagnosis, and early intervention. With over 100,000 births per year, VEHDIP tracks children who need follow-up and/or who have risk indicators through almost 500 letters and approximately 100 phone calls per week to parents or providers. There are currently six VEHDIP staff: a full-time EHDI program manager, follow-up coordinator, and follow-up specialist; and two part-time follow-up specialists and a part-time quality improvement coordinator. All the staff members are in Richmond, the capital of Virginia. In their work, the VEHDIP staff collaborate with approximately 70 hospitals and birthing facilities, over 120 audiologists, and 40 local early intervention systems throughout the state.

Improvements in data collection and the Virginia Infant Screening and Infant Tracking System (VISITS) have led to a decrease in Virginia’s loss to follow-up (LTF) rate. In 2009, Virginia’s LTF rate was 78%, but it steadily decreased to 34.1% in 2015. VEHDIP staff have long been committed to authentically engaging with families and providers to improve these rates. The staff make visits to those hospitals and audiologists that appear to be having the most difficulty with LTF after a failed screen or throughout the diagnostic process. In 2014, the staff recognized there was a need to visit more hospitals and audiologists than staff alone could accomplish in getting to the desired decrease in LTF rates. This recognition led to the inception of the 1-3-6 Family Educator project.

**The Center for Family Involvement**

Under the federal Developmental Disabilities Assistance and Bill of Rights Act, Virginia Commonwealth University’s Partnership for People with Disabilities has received funding since 1985 from the U.S. Administration on Intellectual and Developmental Disabilities to act as the Commonwealth’s university center for excellence in developmental disabilities research, training, technical assistance, and dissemination. One of the largest projects at the Partnership is the Center for Family Involvement, or CFI. The CFI has a mission of working with families to increase their skills as advocates, mentors, and leaders so that family members with disabilities can lead the lives they want. The CFI has grown from one full time and three part-time staff operating two grants ($500,000) in 2005 to a little over $1.1 million in funding in 2017 that supports one full-time and 19 part-time staff running 12 initiatives backed by four state and two federal agencies. The CFI staff are all parents or family members of CYSHCN or are themselves people with disabilities. Annually, the CFI provides one-to-one enhanced emotional, informational, and systems navigational support to approximately 750 diverse families of CYSHCN and participates in informational and educational opportunities that reach over 8,000 families and professionals.

The CFI’s largest initiative is the Family to Family (F2F) Network of Virginia that provides evidence-informed parent to parent support through eight local coordinators, five cultural brokers, three disability liaisons, five 1-3-6 Family Educators, and over 100 volunteer Family Navigators. The F2F Network is Virginia’s federally recognized Family to Family Health Information Center and a nationally recognized Parent to Parent USA alliance member.
VEHDIP and CFI Collaboration

In 2007, the VEHDIP collaborated with the Partnership for People with Disabilities and CFI to pilot a family to family support program using the Guide By Your Side© (GBYS) model from Hands & Voices. The program connects parents of children who have been newly diagnosed with hearing loss to other parents who have already had that experience. Data from the CFI’s three-year pilot of GBYS demonstrated the need for such services:

- Approximately 250 families requested emotional support and unbiased informational support on communication options from another parent.
- Ninety-four percent of the families who were matched with a family guide and responded to a survey reported being satisfied or highly satisfied with the support received from the CFI/Virginia GBYS program.
- Eighty-two percent of parents found the information they received useful.
- Ninety-five percent found the information on communication options helpful in making their decisions.

Success of this pilot indicated the continued need for family to family support with the same objectives to connect newly diagnosed families with experienced parents and to expand supports to culturally diverse families. From 2011 to 2014, the EHDI program contracted with the CFI for evidence informed parent to parent support from its F2F Network. It is important to note that during this timeframe, families in Virginia were trying to establish a sustainable chapter of Hands & Voices and that some funds in the contract with CFI supported those efforts.

The scope of work between the VEHDIP and the CFI was expanded in 2015 to focus on families participating in data collection, policy discussions, and leading stakeholder learning. Prior to this expanded contract, the challenges of distance and time limited the VEHDIP staff to only completing about six hospital visits and six audiology visits each year. The VEHDIP needed boots on the ground or ambassadors who lived in various parts of the state and could extend VDH’s outreach to EHDI stakeholders (i.e., hospitals, audiologists, early intervention programs). Six parents were hired to educate stakeholders about EHDI programs, processes, and best practices. Additionally, and more importantly, these parents were able to share personal experiences of their child’s hearing journey with the stakeholders, as well as with other parents in need of support. This initiative became known as the 1-3-6 Family Educator (FE) project.

As shown in Figure 1, the VEHDIP has progressed from 2000 to 2018 in its efforts involving families. It began with families attending EHDI Advisory Committee meetings to receive information for themselves, evolved into a system of paying it forward by providing peer support to other families, which in turn led to families actively participating in systems change efforts.

Virginia’s 1-3-6 Family Educator Project

1-3-6 FEs are parents of children and young adults diagnosed as deaf or hard of hearing who can commit 16 to 20 hours per month to the 1-3-6 FE project. They receive compensation as contractors. The FE role is three-pronged with the following responsibilities:

- Visit local hospitals and audiology practices to conduct short surveys and discuss the processes used for testing and for sharing hearing testing results with families, and maintain contact with the stakeholders following the initial visit;
- Acquire information about and distribute resources regarding hearing loss and services to professionals and families within their region; and
- Provide emotional, informational, and systems navigational support to families of children recently diagnosed with a hearing loss.

The parents chosen as FEs participate in an interview, submit to a criminal background check, and are required to have transportation to visit hospitals, audiology practices, and other providers. In 2016, the CFI contracted with six parents for the role of a 1-3-6 FE. Two resigned during the first year; one of them was replaced in 2017. The red stars highlight the FEs current geographic distribution in Virginia (Figure 2).

As shown in Figure 1, the VEHDIP has progressed from 2000 to 2018 in its efforts involving families. It began with families attending EHDI Advisory Committee meetings to receive information for themselves, evolved into a system of paying it forward by providing peer support to other families, which in turn led to families actively participating in systems change efforts.
FEs complete the eight-hour CFI training required of volunteer F2F Family Navigators. An additional eight hours of training is also required for the FEs. This training includes:

- An overview of the history of EHDI and the importance of a systematic way to identify infants with hearing loss;
- Screening equipment used for infants;
- Expected procedures for screening, diagnosis, and referrals;
- The types of hearing loss (including deaf-blindness);
- An overview of the EHDI program in Virginia and a review of recent data;
- The role of a 1-3-6 FE;
- How to use various EHDI/CFI products with families, hospitals, and audiologists;
- The process used to match a 1-3-6 FE with a referred family;
- Role-playing providing unbiased family support; and
- Role-playing hospital and audiology site visits (including conducting a survey and sharing of resources).

A CFI staff person (a woman who is hard of hearing and is herself the parent of a young adult who is hard of hearing) and a VEHDIP staff person provide ongoing support for the FEs. These two staff members conduct the initial and ongoing training, regularly provide coaching and mentoring to the FEs, and are available when questions or concerns arise about their role or assignments.

### 1-3-6 Family Educator Site Visits

In February 2016, FEs began visiting hospitals and in May 2016, the first audiology site visit was conducted. Prior to each visit, the VEHDIP staff emails a letter to the hospital or audiology practice that describes the 1-3-6 FE project and introduces the FE who will visit that site. At the same time the letter is sent, VEHDIP staff provide the FE with the pertinent information about the facility, including who to contact and how to best reach that person. The FE schedules the visit through emails or phone calls to the facility; a letter is mailed if there is no response. The visits typically last between 20 and 60 minutes, depending on the availability of the provider. In general, the hospitals have been able to schedule more time to meet with a FE than audiologists have. The average length of time between the FE receiving the hospital information and completing a visit was 18 days while the average time for audiology practice visits was 41 days from time of referral.

Over a 22-month span in 2016 and 2017, FEs conducted site visits with 35 hospital newborn screening teams and 32 audiology practices. They also assisted 89 families seeking support. Additionally, they participated in 10 meetings and trainings with VEHDIP and CFI staff and presented at or participated in 20 community or educational events. All the FEs are members of the EHDI Advisory Committee and participate regularly in quarterly meetings.

Visits by FEs to the first 12 hospital newborn hearing screening teams occurred between March and September of 2016. For these hospitals, the average 2015 LTF rate was 51%, as compared to the overall state LTF rate of 34.1%. For 2016, the average LTF rate for these 12 hospitals was 46.6%; a decrease of 4.4%. The six hospitals that showed the greatest improvement in LTF received visits by FEs between March and June 2016, allowing more time for the effect of the visit to influence the LTF rate.

The information gathered from the FE site visits goes beyond the surveys they complete. Because of the FE site visits, the VEHDIP has learned about pockets of need that they would not know of otherwise. And, hospital staff have witnessed how FEs can impact parents, hospital staff, and even the community. For example, one hospital shared concerns about their role or assignments. In another case, a hospital hearing screening coordinator shared that staff were afraid of giving bad results to a family and that it would help them be more willing to let parents know their child did not pass the hearing screen if the staff were to hear the success story only the FE could share.

The hospitals anecdotally reported that they value the FE site visits. One hospital hearing screening coordinator has requested that the FE return to their hospital to share her experiences with her own child with the nursery staff. The screening coordinator shared that staff were afraid of giving bad results to a family and that it would help them be more willing to let parents know their child did not pass the hearing screen if the staff were to hear the success story only the FE could share.

The VEHDIP has also learned useful information about audiology practices from the site visits, including:

- Audiologists found the follow-up calls from the EHDI team to be helpful.
- Parents come to their appointments with CFI follow-up postcards that were designed by the FEs and shared with hospitals.
- Audiologists are frustrated because they lose money when parents cancel at the last minute.
- Some audiologists were not aware of the VDH-funded Hearing Aid Loan Bank.
- If a child is diagnosed with hearing loss, audiologists are most likely to refer to an otolaryngologist (ENT), early intervention, genetics, and ophthalmology in that order.
- The average number of infants seen is one per week at most audiology practices.

A preliminary survey conducted of the 1-3-6 Family Educator project in April 2017 (Murdock & Yarbrough, 2017)
found that the project was successful for six reasons. (a) The nine responding audiologists agreed that they were satisfied with the FEs and understood their purpose (89% strongly agreed; 11% agreed). (b) FEs felt they were prepared for their role (84% felt well- or very well-prepared; 17% felt prepared). (c) FEs conducted considerably more hospital and audiology clinic visits than EHDI staff were able to do. (d) FEs were pleased with their role and enjoyed nearly all aspects of their job, particularly improving the EHDI system, making a contribution, learning information, providing awareness to other families, visiting newborn screening teams, and providing support and information to families (100% of FEs strongly agreed that they enjoyed each of these roles). (e) FEs understood VEHDIP requirements, knew who to contact at VEHDIP and CFI, and felt supported in their role (83% of FEs had a large gain in knowledge and 100% rated the training/support very good or excellent). (f) When the average age of rescreen was determined for the three-month period prior to initiation of FE visits and for the same three-month period one year later, all but one hospital had decreased the length of time it took to have infants return for follow-up. The average age among all the hospitals visited had decreased from over three months of age to about two months (Figure 3).

The VEHDIP found that both the qualitative and the quantitative results from site visits provide information useful for short- and long-range planning, including where outreach or education would be beneficial, where there are pockets with specific needs or where there are specific requests, and which programs have successful processes that might be duplicated at other facilities. Though the preliminary data looks promising, it is too early to determine whether LTF has declined as the result of the FE visits. The usefulness of the information gathered is significant and has been helpful to enrich VEHDIP’s understanding of individual facility concerns and strengths.

Because the VEHDIP had a long history partnering with the CFI and trusted their ability to recruit, train, and mentor family members for the FE role, there were very few challenges encountered piloting the FE program. One helpful strategy to address the physical distance of FEs to the central CFI office was to continually state and reinforce expectations of the FE role. This was done through quarterly face to face meetings, email, video conferencing, and CFI/VEHDIP staff coaching sessions.

The CFI and VEHDIP recommend that state EHDI programs interested in replicating Virginia’s 1-3-6 Family Educator project invest in (a) an EHDI program liaison to the FEs; (b) on-going face to face meetings with FEs for clarifying processes and brainstorming solutions to challenges they encounter in their work; (c) a parent or a deaf person to coordinate the project and provide on-going coaching; (d) tools for the FE role that are available in multiple formats (i.e., hard copy, as Word documents, and as Google forms) for various learning styles; and (e) connections of FEs to larger family leadership and family engagement efforts so they are not isolated, but joined to other family leaders (who may or may not have children with the same disability/special health care need diagnosis).

Next steps for the CFI and VEHDIP include recruiting additional parents as FEs (with an emphasis on diversity—cultural and diagnosis, including deaf-blindness); establishing five regional EHDI Learning Collaboratives to support facilitation between VEHDIP staff and FEs; and expanding FE site visits to early intervention programs and pediatrician practices.

**Conclusion**

Embedding the 1-3-6 FE initiative within a well-established family-led program supported the VEHDIP’s goal of engaging parents in systems change efforts targeting Virginia’s LTF rate. FEs were supported by other parents of CYSHCN. The director of the CFI had the wisdom and expertise in building leadership behaviors in parents who may not have had lengthy employment histories or post-secondary education. And, on-going professional development for the FEs, particularly the use of technology (i.e., iPads, google products, cloud-based databases), was ensured through their integration with the CFI whose home was within a university.
References


Demographic Considerations in Serving Children Who are Hard of Hearing or Deaf

Karl R. White, PhD

Abstract: People involved with Early Hearing Detection and Intervention (EHDI) programs should understand that fewer than 15% the babies identified by EHDI programs have bilateral profound hearing loss and more than 50% have mild bilateral or unilateral hearing loss. Further, less than 5% of newborns with congenital hearing loss have two parents who are hard of hearing or deaf. It is important that EHDI program managers and staff ensure that educational, audiological, and medical care are tailored to the needs and circumstances of the child and family. Achieving this goal requires that participants in the EHDI system recognize and respect the heterogeneity of this population and the many options that families have for educating and communicating with their child who is hard of hearing or deaf.

Acronyms: ASL = American Sign Language; CDC = Centers for Disease Control and Prevention; HH/D = hard of hearing or deaf; EHDI = Early Hearing Detection and Intervention; HRSA = Health Resources and Services Administration; NCHAM = National Center for Hearing Assessment and Management; SEE = Signed Exact English; UHL = unilateral hearing loss

Correspondence concerning this article should be addressed to: Karl R. White, PhD, National Center for Hearing Assessment and Management, USU, 2615 Old Main Hill, Logan, Utah 84322. Phone: 435.797.3584; Email: karl.white@usu.edu

Introduction

Every U.S. state now has an Early Hearing Detection and Intervention (EHDI) program that is responsible for ensuring that all newborns are screened for hearing loss, that those who do not pass the screen receive timely audiolingual diagnosis, and those diagnosed with permanent hearing loss are enrolled in appropriate early intervention programs (White, 2014). The success of the screening portion of EHDI programs is demonstrated by the fact that the Centers for Disease Control and Prevention (CDC, 2018) reported that 98.0% of newborns are now screened for hearing loss. However, there is still much work to be done informing and supporting families as they (a) make decisions about how they will communicate with their child who is hard of hearing or deaf (HH/D), and (b) learn how to effectively engage in new skills important for language learning.

Most EHDI stakeholders agree that to be effective, early intervention services need to be tailored to the child’s characteristics and the needs of their families (Joint Committee on Infant Hearing, 2013). It is clear that one size does not fit all and interventions are more effective when they reflect the needs of individuals (Zolnierek & DiMatteo, 2009). Even though it may seem intuitive that services for children who are HH/D should be designed and delivered in a way that accounts for differences in parent/family factors (e.g., ethnicity, education, religious and cultural beliefs), other child variables can be overlooked and can powerfully affect service delivery decisions.

What is the Child’s Hearing Status?

Congenital hearing loss for any given child ranges from mild unilateral to profound bilateral. Even though the implications for a child’s development are significantly different depending on the degree of hearing loss, all childhood hearing loss has important developmental consequences as noted by the Department of Health and Human Services in their landmark 1990 document establishing National Health Promotion and Disease Promotion Objectives:

It is difficult, if not impossible, for many [children with congenital hearing loss] to acquire the fundamental language, social, and cognitive skills that provide the foundation for later schooling and success in society. When early identification and intervention occur, [children who are HH/D] make dramatic progress, are more successful in school, and become more productive members of society. The earlier intervention and habilitation begin, the more dramatic the benefits. (p. 460)
Many people do not realize that infants with unilateral hearing loss are by far the largest group of children identified in newborn hearing screening programs. As shown in Figure 1, based on data reported by state-based EHDI programs to CDC (2018) for the years 2012–2014, almost 40% of all babies who were reported by EHDI programs to the CDC during this time period had unilateral hearing loss (UHL). According to Lieu (2018),

School-aged children with UHL score lower on standardized tests of language and cognition and need increased assistance in school for educational and behavioral issues than siblings with normal hearing, and report lower hearing-related quality of life, similar to children with bilateral hearing loss. (p. 74)

Even though the needs of a child with unilateral hearing loss are much different than a child with bilateral moderate or profound hearing loss, all children who are HH/D, and their families, need assistance if they are to reach their full potential.

Hearing Status of Parents

For children who are HH/D to learn language, they need consistent access to fluent language models as early as possible. We have known for decades that which specific language is used is not nearly as important as consistently exposing children to a rich language environment from the time they are born (Hart and Risley, 1995). According to Gallaudet University’s 2011–2012 Annual Survey of Deaf and Hard of Hearing Children and Youth (Gallaudet Research Institute [GRI], 2013), less than 5% of children and youth sampled had parents who were HH/D (see Figure 2).

Thus, for the vast majority of children who are HH/D, their earliest language environment is one of spoken language (usually English, but not always). For families who choose to use a visual language such as American Sign Language (ASL) or Signed Exact English (SEE), or a visual support system such as cued speech, it is important that as many family members as possible become fluent in that choice so that the child has consistent language models during this important developmental period. Although research is sparse, there is evidence that most children who are deaf and are raised with fluent ASL do just as well as children who are deaf and are raised with a spoken language such as English or Spanish (Giezen, Baker, & Escudero, 2014; Marschark, Sarchet, Rhoten & Fabich, 2010).

Communication Modalities Used by Families of Children Who Are HH/D

One of the most frequently discussed issues among people involved in EHDI programs is how families and others will communicate with the child who is HH/D. Will the child communicate using Listening and Spoken Language, ASL, Cued Speech, SEE, or some combination (Gardiner-Walsh & Lenihan, 2017)? Questions about communication modality can be confusing for parents to navigate, particularly as they encounter conflicting information and controversy on this topic, and this can interfere with their ability to make informed decisions about the educational and audiological services they want to have for their child. Discussions about the pros and cons
of various communication modalities (Fitzpatrick et al., 2016; Humphries et al., 2017) can consume considerable time and energy on the part of people managing EHDI programs.

Based on currently available data, there is no “best way” for a child who is HH/D to communicate. Families choose to communicate with their children who are HH/D in a variety of ways, based on an array of factors that are important to their family. EHDI programs, early intervention staff, and health care providers have an important role in assisting families in learning about and considering options, and in helping them access the best possible services and support for their communication choice. According to a recent national survey by the National Center for Hearing Assessment and Management (NCHAM, 2018), a significant number of families explore multiple options during the child’s early years, and many change their approach or combine options from time to time. Table 1 shows the percentage of families using various communication options based on that national survey. These findings are consistent with Gallaudet’s 2011–2012 Annual Survey of Deaf and Hard of Hearing Children and Youth (GRI, 2013) which reported that 51.8% of the respondents were taught using only spoken language, 15.5% were taught using spoken language with cues, 15.2% were taught using sign language only, 13.2% were taught with sign language and spoken language, and 4.2% were taught with “other.”

Table 1
Family Report of Child’s Primary Communication Modality

<table>
<thead>
<tr>
<th>Communication Modality</th>
<th>Percentage of Families</th>
</tr>
</thead>
<tbody>
<tr>
<td>Listening and Spoken Language only</td>
<td>49%</td>
</tr>
<tr>
<td>Sign Language only</td>
<td>3%</td>
</tr>
<tr>
<td>Mostly Listening and Spoken Language (supplemented by sign language, cued speech, or other)</td>
<td>17%</td>
</tr>
<tr>
<td>Mostly Sign Language (supplemented by listening and spoken language, cued speech, or other)</td>
<td>3%</td>
</tr>
<tr>
<td>Mostly Cued Speech (supplemented by listening and spoken language, or other)</td>
<td>12%</td>
</tr>
<tr>
<td>Equal Parts Sign Language and Listening and Spoken Language (including total communication)</td>
<td>14%</td>
</tr>
<tr>
<td>Other</td>
<td>1%</td>
</tr>
</tbody>
</table>

Hearing Status Demographics for Serving Children Who Are HH/D

The vast majority of families who have an infant or young child diagnosed as HH/D have no experience and often have never met a person who has been HH/D since childhood. Thus, those responsible for managing EHDI systems have responsibilities far beyond just making families aware of the various communication options. Instead, EHDI program managers and staff need to ensure that families have opportunities to interact with people who have used various communication options, including those who have used multiple options.

Current federal funding guidelines have begun to address this issue, but are not as broad as they need to be. For example, Health Resources and Services Administration (HRSA) Funding Opportunity Announcement 17-061 (HRSA, 2017, p. 3) encouraged . . . the establishment of Deaf Mentor programs for families with deaf or hard of hearing infants and children in all states/territories. Deaf Mentor programs should provide families regular opportunities with a Deaf Mentor, a qualified deaf or hard of hearing adult, to interact with their child using American Sign Language (ASL) and effective visual communication strategies, and to guide understanding of deafness and Deaf Culture.

Ensuring that families of newly identified children who are HH/D have opportunities to interact with ASL-using adults is important, but it is not enough given that only 6–15% of children who are HH/D being identified in EHDI programs are using ASL as their primary mode of communication (GRI, 2013; NCHAM, 2018). The EHDI system also needs to provide families with opportunities to interact with adults who are HH/D and use Listening and Spoken Language, cued speech, SEE, and other communication modalities.

It is important to remember that many adults who are HH/D were children learning language before EHDI existed. Thus, the experiences that most adults who are HH/D had as children are often radically different than what is currently happening. It is equally important for families of children who are HH/D to have opportunities to interact with families using various communication modalities. Additionally, parents of newly identified children benefit from interacting with other parents of children who are HH/D (Henderson, Johnson, & Moodie, 2016).

An oft-repeated slogan among adults who are profoundly deaf and use ASL, is, “Nothing about us, without us.” It is a good guideline, but it is important to be thoughtful about who “us” is. Including people in EHDI programs who are profoundly deaf and use ASL is important. But it is equally important to include people with varying degrees of hearing loss who use Listening and Spoken Language, cued speech, SEE, and other communication options. It must be remembered that decisions that families make are influenced by multiple factors. Respect and support for their family-specific context should be paramount. Families have the best opportunity to thrive when intervention and support are aligned with their values and needs, and this ultimately helps children who are HH/D reach their full potential.

Conclusion

Identifying children who are HH/D is only the first step in helping these children reach their full potential. Providing appropriate educational, audiological, and medical care requires that services are tailored to the needs and circumstances of the child and family. Achieving this goal
requires that participants in the EHDI system recognize and respect the heterogeneity of this population and the many options that families have for educating and communicating with their child who is HH/D.

References


Signing Exact English: A Simultaneously Spoken and Signed Communication Option in Deaf Education

Kabian Rendel, MS1
Jill Bargones, PhD1
Britnee Blake, MEd1
Barbara Luetke, PhD1
Deborah S. Stryker, PhD2

1Northwest School for Deaf and Hard-of-Hearing Children
2Bloomsburg University of Pennsylvania, Education of the Deaf and Hard of Hearing

Abstract: Current reviews of the literature continue to demonstrate that even with modern assistive listening technology, many children who are deaf or hard-of-hearing (DHH) have English language and literacy gaps compared to hearing peers (e.g., C. Mayer, 2016; C. Mayer & Trezek, 2018). For example, Geers, Tobey, Moog, and Brenner (2008) reported that “early cochlear implantation had a long-term positive impact on auditory and verbal development, but did not result in age-appropriate reading levels in high school for the majority of students” (p. S21).

Given the continually-reported variability of results about language and reading outcomes for children with cochlear implants (e.g., Harris, 2016; C. Mayer & Trezek, 2018), alternative approaches for promoting better language and reading outcomes should be considered. Signing Exact English (S.E.E.), a system designed and demonstrated to encode grammatically-accurate English, is an option to support the development of speech, listening, English language, and literacy. In this article, S.E.E. as it is used in the United States, is contrasted with the many terms that have been used to describe the practice of simultaneously speaking and signing (e.g., total communication, simultaneous communication, sign supported speech, etc.). Research-based responses to common concerns about S.E.E. are provided.

Acronyms: ASL = American Sign Language; CALP = cognitive-academic language proficiency; CASE = Conceptually Accurate Signed English; CI = cochlear implant; CS = cued speech; DHH = deaf or hard of hearing; DLC = Developmental Language Curriculum; IEP = individualized education program; LSL = Listening and Spoken Language; MCE = Manually-Coded English; MSS = Morphemic Sign System; PSE = Pidgin Signed English; NWSDHH = Northwest School for Deaf and Hard-of-Hearing Children; SC = simultaneous communication; S.E.E. 1 = Seeing Essential English; S.E.E. 2 = Signing Exact English; SE = Signed English; SSE = Sign Supported English; SSS = Sign Supported Speech; TC = total communication; TOD = teacher of the deaf

Correspondence concerning this article should be addressed to: Barbara Luetke, Northwest School for Deaf and Hard-of-Hearing Children, 15303 Westminster Way N., Shoreline, WA 98133. Phone: 206-364-4605; Email: b.luetke@northwestschool.com

A Brief History of Total or Simultaneous Communication

Marschark, Schick, & Spencer (2006, p. 9) noted there is still a “continuing concern about low levels of literacy and other academic skills attained by most deaf students” as well as “an attempt to teach deaf children the language [English] that would be used in schools.” When David Denton became superintendent of Maryland School for the Deaf in the late 1960s, he promoted sign language and fingerspelling only after instruction in speech and speechreading was tried. He encouraged “speech then sign” as an alternative to an “oral only” instructional method for students who were deaf or hard-of-hearing (DHH). Around this same time, the philosophy of simultaneous use of speech and sign was introduced by Roy Holcomb, a deaf man with two deaf sons and a supervisor of a program for deaf students in California. Labeled as Total Communication (TC), it involved a multi-sensory approach that included speech, speechreading, signs, fingerspelling, gesture, and pantomime—all of which could be used by adults and students in educational settings (Beck, 2005). Today, most people do not distinguish between TC as a philosophy or TC as a method of communication. In practice, there is no empirical basis to suggest that TC differs from Simultaneous Communication (SC; SimCom). Both SC and TC are umbrella terms used to generally describe speaking and signing simultaneously.

Cued Speech (CS) was proposed in 1966 by Dr. R. Orin Cornett at Gallaudet College to aid speechreading without
the use of signs. It is defined by the National Cued Speech Association as “a visual mode of communication that uses handshapes and placements in combination with the mouth movements of speech to make the phonemes of a spoken language look different from each other.” (http://www.cuedspeech.org/cued-speech-definition). Because CS is based on making phonemes visible and there is little current research on the system available, it is not discussed further in this article. The interested reader is referred to http://www.cuedspeech.org for more information about CS.

Also in the late 1960s, at least three systems of English signing were being developed. These included Seeing Essential English (S.E.E. 1), Signing Exact English (S.E.E. 2), and Signed English (SE). Each of these systems are described below. In all three systems, speech was paired with signs, creating bimodal input that transferred English spoken with the mouth to English communicated to some degree by both the mouth and hands. In situations where interpreting is being done, the systems are technically transliterated—changing the English language from one form to another and not from one language to another.

Seeing Essential English or S.E.E. 1 was introduced in 1966 by David Anthony (1971), a deaf man who was a teacher of the deaf (TOD). It is referred to today as the Morphemic Sign System (MSS). The system uses separate signs for most syllables of words and is often signed by “root words.” For example, gene is the root for genetic, general, and generous (Gustason, 1997). Today, MSS is used exclusively in Amarillo, Texas, where a dictionary can be found on the school website (http://aisd-web.amaisd.org/sites/mss/). Luetke-Stahlman and Milburn (1996) reported that students in the Amarillo program scored higher than most other students who were DHH in Texas on state reading tests. Signing Exact English (originally referred to as S.E.E. 2, but known today as S.E.E.) was developed in the late 1960s by Gerilee Gustason, a deaf woman, Esther Zawolkow, the daughter of deaf parents, and Donna Pfeitzing, the mother of a deaf child. Both MSS and S.E.E. are signed in a grammatically-accurate manner in which users attempt to include every morpheme of what is said (see Figure 1 below).

Deciding that S.E.E. 1 and S.E.E. 2 were too complicated for young children, Harry Bornstein and a team at Los Alamitos High School (e.g., Bornstein & Saulnier, 1984; both hearing) developed Signed English (SE) and published The Comprehensive Signed English Dictionary that is often referred to as “the blue book,” (Bornstein, Saulnier, & Hamilton, 1983). The system included both invented signs and those borrowed from American Sign Language (ASL) as well as 14 affix markers for bound morphemes (e.g., -ed, -ing, -s). The authors intended the system to be used with young children and Gallaudet University published at least three series of children’s stories and reference materials for this purpose. Within a short time period, whole programs adopted SE. Luetke-Stahlman (1988a) found variable recommendations about how SE should be signed. In the preschool storybooks some SE was signed conceptually (i.e., signing the concept of the message rather than single words) so that the grammar of English was not always apparent, while other phrases were signed literally (i.e., signing one sign for each English word). Luetke-Stahlman provided examples of morphemic inconsistently illustrated in the Dictionary. For example, the sign for emotionally retarded does not require the affix markers -ly or -ed but the signs for emotionally disturbed do. In language samples filmed by Luetke-Stahlman, users sometimes created plurals by repeating the signs for nouns rather than using the /s/ marker. Although signs are illustrated in the Dictionary for verbs such as know and don’t know, there is no explanation available as to how to sign inflections such as didn’t know or known. Because of these factors, SE cannot be signed in a grammatically-accurate manner (see Figure 1 below).

As signing in English became more frequent in school programs for children who were DHH, many terms were used to label it. Among these were Pidgin Signed English (PSE), sometimes referred to as contact signing (Baker-Shenk & Cokely, 1996) and Conceptually Accurate Signed English (CASE; http://www.cdc.gov/nccddd/hearingloss/parentsguide/building/case.html), described as a naturally-occurring result when people who do not know the complete grammar of the other’s language want to communicate with each other. PSE and CASE are synonyms for the same method of signing; both combine parts of two languages, thus they do not completely represent the grammar of either one (see Figure 1 below). In addition to these terms, a number of other labels are used, such as Manually-Coded English (MCE; defined on Wikipedia as “a variety of visual communication methods expressed through the hands which attempt to represent the English language”), Manual English, s/Signed English (the lower-case form signifying a generic term; and the upper-case form signifying the system developed by Bornstein, Saulnier, & Hamilton, 1983). These terms have been used over the last 60 years to label variations of simultaneous speech and sign (Stewart, Bonkowski, & Benet, 1990) that by design do not completely represent the grammar of English (see Figure 1 below).

Sign Supported English (SSE) and Sign Supported Speech (SSS) are terms used more recently, borrowed from British authors. These methods are defined as the use of British Sign Language vocabulary using English sentence structure and grammar (Sutton-Spence & Woll, 2004; Nielsen, Luetke, & Stryker, 2011). Signed Speech (Johnson & Durieux-Smith (n.d.) is another term used in Great Britain to mean SC.

Current data could not be located as to the popularity of any of the above-mentioned forms of simultaneously speaking and signing, but survey data from the Gallaudet Research Institute (2013) indicated that approximately 13% of children who were DHH in the United States received instruction in some type of simultaneous
speech and sign. Specifics as to which sign methods were included in the percentage were not mentioned and because the survey data has been discontinued, more recent data are not available. Given the confusion in sign system labels described by Dan Diffee (personal communication, May 16, 2018), below, it is also unlikely that the percentage is accurate. Still, it is included here to indicate that there are a significant number of families, professionals, researchers, and children who use a communication method that is not Listening and Spoken Language (LSL) or ASL.

What is Signing Exact English (S.E.E.)?

Signing Exact English was developed in the late 1960s by Gelrilee Gustason, a deaf woman and teacher of the deaf; Esther Zawolkow, the daughter of deaf parents and an educational interpreter; and Donna Pfetzing, an educational interpreter and the mother of a deaf child. By the early 1970s, the three colleagues started their own company and published “the yellow book,” the S.E.E. Dictionary (Gustason, Pfetzing, & Zawolkow, 1973). Signing Exact English was initially referred to as S.E.E. 2 or (S.E.E. II) because MSS was originally referred to as S.E.E. 1 (or S.E.E. I), but today it is simply referred to as S.E.E.

Gustason (1990) explained that the authors of S.E.E. were motivated to invent the system for use by both parents and professionals for the following reasons:

1. Increased knowledge about how hearing children develop English language;
2. A desire to have children who were DHH use English as it was used by hearing teachers in school programs;
3. Dissatisfaction with the educational achievement of children who were DHH (e.g., compared to their hearing peers, children who were DHH typically lagged significantly behind their hearing peers in reading and writing skills; had smaller English vocabularies; used simpler and more rule-bound clauses; had a weaker grasp of the morphological and syntactical rules of English; used fewer adverbs, auxiliaries, conjunctions, and figurative words and phrases; and made many errors of omission of necessary words); and

4. Research had shown that access to the morphology of spoken English was impossible via speechreading alone. As noted by Gustason (1990, p. 109), “Research on speechreading indicated that 40 to 60% of the sounds of English looked like other sounds on the lips (e.g., interest, interesting, interests, and interested are nearly impossible to distinguish) and the best speechreaders used their knowledge of English to fill in the gaps... otherwise bright and capable deaf children caught only 5% of what was said though speechreading.”

To address the need to visually represent words and grammar fully and accurately, S.E.E. was designed so that the signs corresponded with the number of morphemes of the English utterance (Gustason et al., 1973; Gustason & Zawolkow, 1993) and represented age-appropriate, complete, grammatically-accurate, proficient, whole English (Luetke-Stahlman, 1993b). Signs in S.E.E are provided for root words and affix markers (e.g., re-, un-, -ing, -ity, -ness) including articles, conjunctions, pronouns, and so forth, so that all words and word parts (i.e., bound morphemes) are made visually obvious, resulting in the potential for children to acquire morphemic awareness, which Gustason claimed was necessary to read proficiently. In S.E.E. there are different signs for different words, so that it is possible to sign electric, electrical, electrician, electricity, and non-electrical all as they are said and written.

S.E.E. uses a two out of three rule: If a word is spelled with the same letters and sounds the same, it is signed in the same way, even if the meaning of the two words are different. Out of respect for some ASL signs, there are exceptions (G. Gustason, personal communication, June 25, 2018). As Schick and Moeller (1992) explained, S.E.E. “attempts to represent English literally, and it purports to follow a strict criterion of one sign for one English free morpheme or ‘word’” (1992, pp. 318–319). The researchers also noted that S.E.E. follows English semantics and does not borrow from ASL semantics (i.e., conceptual signs), unlike some other sign modes (i.e., TC, SC, CASE, etc.). Schick and Moeller (1992, p. 319) gave the example in S.E.E. for the word run which “would appear as the same sign in the following phrases even through a different sign for each phrase would be used in ASL: ‘a home run’; ‘a runny nose’; ‘run for office’; and ‘a run on the bank.’” S.E.E. includes many ASL signs that have only one English translation and roughly 75% of the signs are common to ASL, S.E.E., and PSE/CASE. To illustrate how S.E.E. signs are initialized, Schwarz,
Guajardo, and Hart (2018) described the S.E.E. sign for the word *ant*, made with an A-hand-shape using the dominant hand on a wiggling base 5-hand (see Figure 2).

![Image of S.E.E. sign for the word “ant.”](image_url)

When the handshape on the dominant hand is changed (using a B, C, I, P, R, or T), the signs for other words are created (i.e., BEETLE, CRICKET, INSECT, PEST, ROACH, TERMITE; respectively). Groups of signs like these are often referred to as sign families, that assist in the retention of sign formation.

S.E.E. uses the visual features of directionality/movement, change in location based on semantics, facial expression (e.g., questioning, surprise) sign emphasis, body referencing, eye gaze, sign directionality, and use of space as explained by the authors in the first edition of the S.E.E. dictionary (Gustason et al., 1973) and again in Gustason and Zawolkow (1993).

Luetke-Stahlman (1988a, 1988b, 1988c, 1988d) showed how MSS, S.E.E., and Cued Speech use inputs that **completely** encode the morphology and syntax of English as compared to TC, SC, PSE, CASE, and MCE which **incompletely** encode spoken English (see Figure 1). In her research, Luetke-Stahlman (1988a, 1988b, 1988c, 1988d) found that large numbers of students exposed to grammatically-complete English codes (i.e., MSS, S.E.E., and Cued Speech) scored higher on tests of English language and reading achievement than students exposed to systems using grammatically incomplete English codes (i.e., TC, SC, PSE, CASE, and MCE).

Stryker, Nelson, and Luetke (2015) noted that because PSE/CASE and SE lack sufficient affixes to fully represent English morphology **through the air**, PSE/CASE and SE-users “inevitably provide DHH students with ungrammatical English input when morphologically complex words are communicated, such as unworkable, irreplaceable, and foolishness” (Schwarz et al., 2018; p. 11). S.E.E. advocates believe that as students are developing their speech and English language skills, it is better to sign each affix in such multi-morphemic words. Once a student pronounces or signs all of the syllables of the word, it is no longer necessary to sign all the affixes (e.g., characteristically could be signed as CHARACTER + LY).

Luetke (1988a, 1988c, 1991, 1993a) and Luetke-Stahlman and Tyrrell (1995) completed several studies in which language samples were taken and coded to determine a sign-to-voice ratio following procedures described by Luetke-Stahlman (1982). The results of this work are depicted in Table 1 and explained later in this article.

**Table 1**  
*The Degree of English Necessary to Encode the Semantics and Grammar of English in Various Sign Methods*

<table>
<thead>
<tr>
<th>Language Component</th>
<th>Pidgin Signed English (PSE)</th>
<th>Conceptually-Accurate Signed English (CASE)</th>
<th>Signed English</th>
<th>Manual English</th>
<th>Sign Supported English or Sign Supported Speech, Signed Speech</th>
<th>Morphemic Sign System (MSS)</th>
<th>Signing Exact English (SEE)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Semantics (meaning) encoded</td>
<td>Some</td>
<td>Some</td>
<td>Some</td>
<td>Some</td>
<td>Some</td>
<td>Yes</td>
<td>Yes</td>
</tr>
<tr>
<td>English morphology and syntax encoded</td>
<td>Some</td>
<td>Some</td>
<td>Some</td>
<td>Some</td>
<td>Some</td>
<td>Yes</td>
<td>Yes</td>
</tr>
</tbody>
</table>


Appelman, Callahan, Mayer, Luetke, and Stryker (2012) demonstrated that when compared to a national sample of post-secondary deaf adults, graduates of a program that used simultaneous speech and S.E.E. had higher graduation rates from high school (100% of S.E.E. graduates compared to 93% nationwide), earned more college degrees (67% of S.E.E. graduates compared to 30% nationwide) and had higher employment rates (85% compared to 65%). M. Mayer (2013) and Nielsen, Stryker, Luetke, and McLean (2016) empirically demonstrated how S.E.E. can be used to successfully support the development of speech articulation, listening skills, English language development, and literacy abilities in children who are DHH.

Information regarding S.E.E. materials, virtual S.E.E. courses, a S.E.E. smart phone application, and the dates
A number of school programs that are successfully using S.E.E. exist around the United States. These include NWSDHH, where IEP teams in about 20 school districts have found S.E.E. to be the appropriate method of communication for about 45 children, preschool through 8th grade. S.E.E. is paired with spoken English and all children wear assistive listening devices which are checked daily. In addition, about 17 regional day school programs in Texas (D. Diffee, personal communication, May 16, 2018) use this method. S.E.E. is used with individual students in California, Kansas, Kentucky, Missouri, Nebraska, and Wisconsin. S.E.E. Skillshops are held in many of these states. For more detailed information about ASL and SC (including S.E.E.) see Stewart and Luetke-Stahlman (1998).

Confusion Regarding Sign Systems

Confusion among parents, interpreters, professionals, and researchers concerning the type of signing being used is common. As exemplified in a recent email from a consultant in Texas (D. Diffee, personal communication, May 16, 2018), “many who purport to use Signed English (SE) actually use PSE…Those who use simultaneous communication in Texas, use PSE although they might call it Signed English, Manual English, Manually Coded English, Sign Supported Speech, and so forth. Some report that they try to sign in English word order (i.e., the words are in the correct order in a sentence but not all bound morphemes or function words are signed) and not in grammatically-accurate English (i.e., all morphemes are signed) or ASL during English class, Language Arts, or reading and writing instruction.”

D. Diffee (personal communication, May 16, 2018) added that the “state of TX interpreter certification board misunderstands Signed English, which they define as ‘the ability to watch and understand information and ideas presented through signs, gestures, classifiers and fingerspelling in an English-like structure and to communicate information and ideas through signs, gestures, classifiers, and fingerspelling in an English-like structure so that others will understand’” (Texas Department of Assistive and Rehabilitative Services, 2012, p. 16). Classifiers, handshapes and/or rule-grounded body pantomime used to represent nouns and verbs for the purpose of providing additional information (e.g., location, kind of action, size, shape, and manner) do not exist in English. It is rare that the authors of published studies involving signing in English by adults and/or children film and analyze the degree to which the morphology and syntax of English is signed so that descriptions are empirically compared one method to another.

Not only is there confusion about the labels and use of simultaneous speech and sign, but standard practice in research studies involving these methods is to collapse both grammatically-complete and grammatically-incomplete methods into one group for analysis (e.g., Geers, 2003; Geers, Brenner, & Tobey, 2011; Geers, Brenner, Nicholas, Tye-Murray, & Tobey, 2003; Knoors & Marschark, 2012; C. Mayer & Akamatsu, 1999; Niparko & Geers, 2004). Giezen, Baker and Escudero (2014, p. 107) noted that “a large number of studies have compared children in Oral Communication (sometimes referred to as LSL) settings, where only spoken language is used, to children in Total Communication (TC) settings, where both spoken language and some form of signed communication are used” (italics added). Clarification as to the degree to which the morphology and syntax of English were used by parents and/or teachers are not typically described in the research methodology of these studies (see Schwarz et al., 2018, as an exception). Because of confusions such as those just mentioned, it is important to have definitions and examples whenever simultaneous English signing is discussed.

A Rationale for Providing S.E.E. as an Option in Deaf Education

It is not surprising that when hearing levels are first identified most parents and professionals focus on obtaining assistive listening devices and parent-child communication. The acquisition of age-appropriate English or literacy skills is often not an immediate concern. However, later, if the toddler or preschooler does not begin to use language like his or her hearing peers, concerns may arise. In addition, even if the child uses intelligible speech, Archbold and Mayer (2012, p. 3) cautioned that “excellent levels of speech intelligibility…may mask the young person’s language delay or difficulty,” especially with regard to cognitive-academic language proficiency (CALP) as it is spoken, read, or written.

A fudge sundae analogy can be used to explain CALP, as well as to discriminate speech from language development. The analogy represents the acquisition of linguistic competence using terms first proposed by Cummins (1980, 1984). In the analogy, a child’s speech articulation ability is represented by fudge because most parents and professionals strive for children to achieve their best possible speech intelligibility. Basic interpersonal communication, or the degree of English that is used routinely and includes known vocabulary and simple grammar, is represented by ice cream.
CALP as defined by Archbold and Mayer (2012) is represented by the bowl. CALP is the English language needed in school to understand and express abstract, decontextualized thinking, as well as to comprehend what is read, and to write similar to their hearing peers (Luetke-Stahlman, 1998). Research by Nielsen et al. (2016) demonstrated that in a program where S.E.E. was used to develop English and literacy, most students achieved CALP as measured on standardized tests of English language and reading ability. The study did not include a comparison group of students using a different method of communication.

When children who are DHH are eligible for preschool, choice of communication method and other classroom variables are often discussed. Eisenberg, Shannon, Martinez, Wygonski, and Boothroyd (2000) demonstrated empirically that even with amplification, access to acoustic information (e.g., the teacher’s instruction, peers’ social language) is degraded for children who are DHH compared with the information acquired by hearing peers, and the vocabulary of most children with hearing loss is compromised. Even for students with mild hearing losses, the result can be underdeveloped communication abilities (Tomblin, Harrison, Ambrose, Walker, Oleson, & Moeller, 2015; Yoshinaga-Itano, Sedey, Wiggin, & Chung, 2017). In research by Mukari, Ling, & Ghani (2007), general classroom teachers using an informal language checklist rated children with cochlear implants (CI) interacting in their mainstreamed classrooms as poor in their LSL communication abilities. Yet, many children who are DHH develop articulate speech and age-appropriate language. Relevant in this article are those who do not. For example, when only CI-using children have been studied, Geers et al. (2008) and Gregory and Hindley (1996), among others, emphasized the need for communication alternatives because some learners continue to lag significantly behind their hearing peers (see also Geers & Hayes, 2011; and C. Mayer & Trezek, 2018; Traxler, 2000). Knoors and Marschark (2014) suggested that CI-using children as a group rarely reach levels of academic achievement comparable to hearing peers regardless of whether children are enrolled in LSL or ASL programming. This is an outcome that has remained remarkably persistent over time (C. Mayer & Trezek, 2018, p. 1). Geers, Mitchell, Wareener-Czyz, Wang, & Eisenberg, 2017, in a study based on children recruited between 2002 and 2004, “found no advantage to parents’ use of sign language (ASL) either before or after CI” (p.1). However, data was collected by parent report “just before surgery and at 12, 24, and 36 months postimplant” (p. 2). Long-term sign use was defined as “positive for sign language use at baseline and/or 12 months and at 24 and 36 months postimplant (N = 36)” (p. 2). Parents were also asked to estimate how much of the day they used sign language. These responses were divided into two categories for analysis (less than 50% and more than 50% of the day in the home). The authors did not provide a definition for the term “sign language,” did not verify the use of parental sign via filming and analysis of voice-to-sign ratio, and did not include an analysis of children who use sign in their educational program as is discussed in this article.

C. Mayer & Trezek (2018) reviewed 21 studies of primarily school-age students who used CIs and were assessed on standardized measures of reading and writing. The researchers found that most children achieved in the average range, although there was a wide variation in their scores. This finding of variability has been found repeatedly (see also Harris, 2016; Marschark, Sarchet, Rhoten, & Fabich, 2010). In addition, CI users may evidence age-appropriate literacy outcomes in the earlier school years but not maintain them in higher grades (Arfé, Ghiselli, & Montino, 2016; Harris, 2016; C. Mayer & Trezek, 2018). Especially when children who are DHH are young and still acquiring English, they can have difficulty hearing all the parts (morphemes) of English syntax and grammar. Relevant research to this point is provided below:

- Cannon & Kirby (2013) studied 26 children who were DHH, 5–12 years of age, with whom sign was not used and found that the children had difficulty with regular noun singular/plural; accusative first- and second-person singular; noun/verb agreement copular be; accusative third-person number/gender; locative pronominal; auxiliary be/regular past—ed; prenominal determiners plural and inconsistently producing tense markers.
- Spencer, Tye-Murray, and Tomblin (1998) found that 32% of the aural/oral children studied could not produce the third person s and 51% did not produce the past tense ed. They concluded that the amount of comprehensible input that the children with CIs can perceive influences the acquisition of the inflectional morphology, such as tense markers. As a group, initial speech sound improvement after implantation declined between the fourth and sixth year of CI use.
- Tomblin, Peng, Spencer, and Lu (2008) as well as Ramirez Inscoe and Nikolopoulos (2009) reported similar findings regarding the stabilization and plateauing of speech intelligibility trajectories.
- Koehler, Van Home, and Moeller (2013) described the risk of undeveloped grammatical morphology when children are hard of hearing if input is “inconsistent and distorted” (p. 1702). They found that children who are hard of hearing may have difficulty processing and storing grammatical morphemes that have “low phonetic substance,” such as “verb-related morphemes” (e.g., contracted forms of be, third-person singular -s, and regular past tense –ed; p. 1702–1703).

The ability to both perceive and produce all the morphemes (i.e., including bound morphemes) of English is also important for attaining literacy. As Easterbrooks and Stephenson (2006, p. 390) explained, “word meanings are expanded, modified, and changed routinely by affixing
single and multiple morphemes to the front or end of a root word...If students who are DHH are to read and write well, they must have facility with the morphemic system." For example, the Kindergarten level of National Geographic Windows On Literacy (McGough, 2001) contains 11 bound morphemes (i.e., -ed, -en, -er, -ing, -ist, plural -s, third person -s, the suffix -self, and -y, as well as the irregular past tense and the contraction 's (what's). One word, bakery, contained three morphemes. Luetke (2013) reviewed readers for first graders published by Harcourt and found that to read and comprehend first grade selections a student would need to comprehend and express at least 10 bound morphemes (i.e., di-s, -ed, -en, -ly, -ful, -ing, plural -s, possessive -s, third person -s, and -y. These are understood and used by hearing six-year-olds (de Villiers & de Villiers, 1978) who are prepared linguistically to read them. Third grade level selections included words with the bound morphemes: -able, -an, -ant, -er, -ible, -ic, -ice, -in, -ion, -ious, -its, -ity, -ment, mis-, -or, -re, -sion, -th, -tion, and un- and fifth grade readers were found to include -age, -al, -ch, -est, -ify, -ish, -ite, and -ize (Luetke, 2013). Many readers who are DHH do not have access to these bound morphemes because they do not use a grammatically accurate sign system, or their assistive listening device does not provide a clear enough signal.

There is possibly a relationship between the finding that most children who are DHH read at the fourth-grade level when they graduate from high school (Traxler, 2000) and the lack of exposure to single morphemic words (e.g., a, the, to) and bound morphemes (e.g., -ible, -ic, -ice, etc.). Luetke-Stahlman (1988b, 1990b) found that S.E.E.-using children had better English language and reading abilities than children who use PSE. Nielsen et al. (2016) found that most students in a S.E.E. program had intelligible speech articulation, average English language ability, and read at or above grade level.

S.E.E. has proven to be an appropriate option when compared to other methods of simultaneous communication for some children who are DHH when English is the desired language of instruction and social interaction. However, the research on S.E.E. is often ignored and the method is frequently not offered as a communication option for families and professionals. Although more research on S.E.E. is warranted, parents deserve to be given information on all methods of communication that might assist their family.

Responses to Common Concerns About S.E.E.

It is not unusual for current articles written by advocates of LSL or ASL to include concerns about sign systems (Gardiner-Walsh & Lenihan, 2018). Below are some of the most often stated criticisms about S.E.E. with additional information about each one.

Sign Use Hinders Speech Development

As noted by Knoors and Marschark (2012, p. 294), this topic “remains a ‘hot button’ for many people.” However, there is empirical evidence that sign can support both the understanding of speech and speech production itself. Giezen et al. (2014) reviewed the literature related to children who used CIs and were exposed to sign and also conducted experiments of their own. They found that for the children they studied, sign “did not interfere with spoken word processing and may even have provided a benefit when children were trying to perceive perceptually confusable words” (p.118–119).

In contrast, Fink, Wang, Visaya, and the CDaCI Investigative Team (2007), and later Geers, Mitchell, Warner-Czyk, Wang, Eisenberg, and the CDaCI Investigative Team (2017), found that when parents signed to their child during early childhood, the child’s use of speech and listening strategies was compromised compared to when parents did not use sign. These researchers (a) defined “sign” to include ASL and “baby sign” as well as other simultaneous communication methods, (b) used parent report to determine whether sign was used, and (c) analyzed ASL and all simultaneous communication methods as one group. They found that children who did not sign had better speech perception and articulation abilities than those children whose parents reported that they used some form of sign communication between 12 and 36 months after implantation. Some parents used sign until their child was 12 months post-implant and others used it for 12, 24, or 36 months post-implant. Sign might have been used for less than half the day or more than half of the day, the two rating choices surveyed. The authors did not verify the use of parental sign via filming and analysis of voice-to-sign ratio and did not include an analysis of children who use sign in their educational program as is discussed in this article.

M. Mayer (2013) noted that children developing English language learn best when difficult productions are taught in small steps, encouraged, and reinforced. As Schick (1997) stated: Children learn what is modeled for them and by the kind of production elicited (and facilitated) from them. To partially illustrate this point, Luetke-Stahlman and Tyrrell (1995) asked adults who purported to use SE or PSE to transcribe the vocabulary and English grammar of sentences that they themselves had signed two years previously (e.g., “The cars in the lot were lined up in rows; Time is fleeting;” Luetke-Stahlman, 1993a). Professionals using SE and PSE could not retrieve the semantics and grammar of the original utterances when they attempted to transcribe them. The researchers wondered how educators could expect students, the recipients of their input, to understand their “English” if they themselves couldn’t extract English vocabulary and/or grammar from what they had signed.

M. Mayer (2013) demonstrated an outcomes-based approach whereby morphemic aspects of English that a child was not using were facilitated by intentional
strategies. In doing so, she noted that adults who use Sign Supported Speech typically sign the main words in phrases, ones already being expressed by the child, and not the words or word parts (e.g., bound morphemes) that the child is missing. Speaking of using grammatically-incomplete simultaneous communication methods, Mayer noted that the parts of the English language that are hardest to hear and in the most need of sign support are the ones that adults do not sign. In her work, Mayer provided spontaneous language samples to illustrate the kind of signing in English that is possible when adults are motivated to sign accurate grammar and reinforced for doing so (Nielsen et al., 2016). In the following sample from her study, the TOD simultaneously signed 93% of what she said, omitting words like "oh" and those involved with a listening only condition as she facilitated the listening, speech, and English language development of a Kindergartener (intentional strategies are underlined).

Spontaneous Language Sample from M. Mayer (2013)

G. is interested in a small plastic toy he thinks represents apples and shows the toy to his TCD. She follows his lead and uses parallel talk to express what she believes he is trying to communicate to her (Luetke-Stahlman, 1996). The TOD speaks and signs simultaneously in a slow and deliberate manner, using a distinct sign for every morpheme—including an affix marker on the word "apples." "Oh, you think those are apples?" the TOD asks. She exaggerates the /s/ on the word "apples" in both speech and sign, holding the -s handshape for longer than is typical in normal conversation (i.e., both signed and spoken highlighting; Fleming, 1974). She clarifies that they are both looking at little, plastic coconuts (e.g., "Oh, you know what? Those are coconuts!") and then counts the coconuts, repeating and emphasizing the -s on the word coconuts, and informing G. "with a -s on the end." Next the teacher models the target word only. "Coconuts," she says, elongating the -s in both speech and S.E.E. This is followed by a direct request for the desired speech sound or morpheme that the child is omitting: "Can you tell me the -s on the end of coconuts?" G. does so and his teacher specifically labels his effort in her reinforcement "Good. Good -s sound on the end of coconuts." She continues with the conversation by asking, "You love coconuts?" (again exaggerating the -s sound in her speech and sign), but G. has switched topics on her and asks if he can have a toy squirrel that they have played with in the past. The TOD again follows his lead and repeats his request, modeling, "May I have the squirrel?" (emphasizing the word "have") that he has omitted in both speech and S.E.E. sign. G. attempts to imitate her model. "Again," she repeats (so that correct speech articulation and grammar are habitualized, from Appelman et al., 1980). G. repeats his query but omits the word "the." "Listen again from the word the..." the TOD requests as she picks up a speech hoop. "Listen." Using the hoop to prevent G. from speechreading, she models in speech only, "May I have the squirrel." G. attempts to repeat the sentence and his teacher positively reinforces him, "Oh, you are so close!" she encourages. She asks him to listen again for "May I" and again she uses the speech hoop to encourage G. to listen for the desired morpheme he has omitted. "May I have the squirrel," she models, emphasizing the word "the." This time G. repeats the request correctly and his teacher reinforces him before explaining that he can have the squirrel after they work on vocabulary. She emphasizes the word "after." When G. doesn't respond, she models conversational turn taking, saying "Okay...Say, 'Ok'" from behind the speech hoop. G. takes his turn in the conversation: "Ok" he says, imitating her prosody.

Signing Exact English is Not a Language

S.E.E. is not a language; it is an invented system that encodes English; however, as Mitchell (1982) stated, "there is no logically implicit reason why contrived systems of communication should be considered less functional than 'natural' languages" (p. 332). There is no evidence that parents are troubled by the fact that S.E.E. is not a language. Parents are more concerned that family members have a way to communicate to their child (Luetke-Stahlman, 1996). Any concept that can be said via LSL or signed via ASL, can be expressed using S.E.E., no matter how abstract or complex. Parents who use S.E.E. want their child to learn proficient English—the door to literacy, high school and college graduation, and employment (Appelman et al., 2012).

S.E.E. Cannot be Signed in a Manner that Represents Grammatically-Accurate English

Some linguists, educators, and parents believe that S.E.E. cannot be signed accurately because of a widely-quoted study by Marmor and Petitto (1979) or because they have seen people sign one of the other sign systems and not proficient S.E.E. However, in the Marmor and Petitto (1979) study, communication samples were analyzed from only two users of PSE who were teachers of the deaf and employed at a residential school. Given these variables, it is not surprising that the finding was that only 10% of the grammar of English was signed because PSE users don't attempt to completely encode the grammar of English. By its very definition, PSE is an unstandardized mixture of English and ASL. Still, the results of the Marmor and Petitto study were widely quoted in the late 80s, at a time when a bilingual approach using ASL was introduced into the educational arena. Today, it is rare that those who have read the available research literature on the topic do not acknowledge that S.E.E. can convey English on the hands. A few of the relevant studies to this point are summarized below:

- Luetke-Stahlman (1988a, 1988c, 1991, 1993a, 1993b) and Luetke-Stahlman and Tyrrell (1995) conducted several studies in which language samples were taken and coded to determine a sign-to-voice ratio following procedures described by Luetke-Stahlman (1982). This calculation shows the degree to which specific vocabulary as well as the morphology and syntax of English are represented simultaneously. The ratio is figured as a percentage of the free and bound English morphemes that are signed compared to those that are spoken. The results showed that S.E.E.-users were able to sign English to a more complete degree than those who used SE and PSE. It was recommended that parents and professionals attempt to sign 100% of what they are saying, which typically results in a high ratio; film themselves; calculate their sign-to-voice ratio; and set goals for improvement.

- As a part of the Nielsen et al. (2016) study, spontaneous language samples of teachers as
they were interacting with children were filmed and analyzed. TODs were unaware that sign-to-voice percentages would be calculated. The average sign-to-voice ratio across TODs was 94.4%, demonstrating that S.E.E. can be signed in a manner that accurately represents the grammar of English.

- M. Mayer & Lowenbraun (1990) found that people were able to sign grammatically-correct English via S.E.E. if they learned the vocabulary and grammar of the system, were motivated to sign proficiently, and were regularly observed and coached. These results were supported by similar research by Leigh (1995) who studied the Australian English used by TODs in Australia. Like Mayer and Lowenbraun, Leigh advocated that positive steps be taken to ensure consistent and accurate English input via sign, including: (a) adequate training and experience in the use of the system, as exemplified by appropriate assessment; (b) a positive attitude toward the method; (c) accurate knowledge of the encoding principles and specific rules; (d) commitment to use at all times; and (e) monitoring and coaching.

Children Who Use S.E.E. will be Unable to Converse with Members of the Deaf Community

Because S.E.E. was invented in the early 1970s and has been in use for almost 50 years, there are hundreds of deaf adults who were raised using S.E.E. and are capable of using it as a part of the array of communication options available to them, including speech, S.E.E., PSE, and ASL. Children who grew up using S.E.E. often learn to code-switch depending on the person with whom they are conversing. It is inconceivable that the “Deaf Community” is a single entity that converses using one method of communication.

In addition, in the only research article the authors could find on the topic, Luetke-Stahlman (1990a) demonstrated that elementary-aged children of differing communication backgrounds comprehended ASL to the same degree. In that study, 12 deaf children who were enrolled in a public-school program that used S.E.E. and 14 deaf students enrolled in a residential program that used a combination of PSE and ASL watched commercially-available ASL stories and answered comprehension questions about them. There was no difference in the ability of S.E.E. students as compared to residential peers in the number of correct answers they supplied to written comprehension questions.

S.E.E. is Unnecessarily Complex

Contrary to the claims made by Bornstein (1990), S.E.E. is no more complex than the English being used by the person. The authors could find no research support for the notion that it is beneficial to use simplified English with children who are DHH or that S.E.E. is too complex for children to acquire, no matter their age.

Children who are DHH deserve the same level of instructional and social language input as hearing children receive. If adults do not sign grammatically-correct English, children who are DHH do not have access to the vocabulary and grammar that hearing children do. Examples of age-appropriate English can be found in developmental lists of vocabulary and grammar. One such source is the Developmental Language Curriculum (DLC; Luetke-Stahlman, 1998, reprinted from Cheney, Compton, & Harder, 1988) which lists parts of English such as pronouns, possessives, helping verbs, and bound morphemes that are typically acquired by hearing toddlers and preschoolers (and children up through 8 years of age). According to the DLC, a child who is 24 to 30 months old should use more than a dozen verbs; use the present progressive verb form (-ing); name at least one color; use the pronouns it, this, that, and you; and the preposition in; and, have a vocabulary of 300 to 500 words. Before three years of age, the child should use 2- to 3-word combinations; use what, where, and why in question forms; use two dozen verbs; begin to use regular past tense (-ed); use about a dozen adjectives; use at least four pronouns; use contractions for negatives such as can’t and don’t; use the prepositions on, under, and off, use determiners such as a, the, this, and that; use the conjunction and; use the possessive marker ‘s as well as plural -s; and, have a vocabulary of 500 to 700 words. Children who are DHH and are not exposed to these parts of English in an accessible manner, have great difficulty acquiring these parts of English, many of which are difficult to hear (Guo, Spencer, & Tomblin, 2013).

Children Who are DHH Don’t Learn Grammatically-Correct English or Literacy Via S.E.E

In the most recent study about this concern, Nielsen et al. (2016) found that most students enrolled in a program that used simultaneous speech and S.E.E. acquired English proficiency and read on grade level. Participants were 17 children who were from English speaking homes, old enough to be tested on the standardized language and reading tools used, had no additional significant disabilities, and were granted permission by their parents to participate. Extensive information about the 17 students’ background (i.e., ethnicity, age of identification, age of obtaining of assistive listening equipment, unaided pure tone average, pure tone average while wearing equipment, social economic status, and parent signing ability) was published in McLean, Nielsen, Stryker, and Luetke (2015). All the students wore amplification at home and school; all came from families where English was the language of the home (and no family used another method of simultaneous communication or ASL).

Students were administered both informal and formal (standardized) tests of language and reading annually. The study found a developmental trend (and no plateau) for both English language and reading achievement (i.e., students improved in their abilities each year).

Morphological awareness, made possible via S.E.E., was found to be a prerequisite to high language and reading
scores. Nielsen et al. (2011) reviewed the research on reading acquisition, including the importance of morphemic awareness. They showed how the construction of S.E.E. facilitates morphemic awareness. In the Nielsen et al. (2016) study, neither speech ability nor CI use was significantly correlated with English acquisition or reading ability. Instead, English language proficiency predicted reading achievement. That is, the more proficient a student’s English, the more age-appropriate their reading ability was.

Schick and Moeller (1992) conducted a series of studies involving children enrolled in a S.E.E. program and Schick (1997) reported that they internalized and produced some of the most complex rules of the syntactic structures in English as measured by embedded clause, conjunctions, and modal. Further, an increase in cognitive complexity resulted in more complex use of English structures but not an increase in errors.

**Conclusion**

This article has distinguished the invented system of Signing Exact English (S.E.E.) from the commonly-confused array of terms used to describe various ways of speaking and signing at the same time (e.g., total communication, simultaneous communication, PSE, CASE, MCE, S/signed English, Manual English, Sign Supported English or Sign Supported Speech, etc.). These various methods are often discussed without definition and without information as to the extent to which English grammar is accurately signed. There is a lack of research and analysis of these communication methods to clarify whether and if so, how they differ from each other in actual use.

S.E.E. is a viable option to Listening and Spoken Language (LSL) or American Sign Language (ASL). The rationale, examples, and research have been summarized and show that S.E.E. can be an effective primary method of communication for many children who are DHH and their families. The fact that an estimated 13% of children who are DHH use S.E.E. is further evidence that it can be an effective communication method. Research has demonstrated that S.E.E. differs in the accuracy of English grammar conveyed when compared to PSE.

Responses to common concerns about S.E.E. were provided so that this information is available to parents and professionals who are considering communication options for children, discussing equal representation on panels and committees, and crafting public policy. Because S.E.E. is being used successfully by many families across the United States, research on the system as well as other methods of simultaneous communication is warranted.

**References**


Luetke, B. (April, 2013). Research update: Sign supported speech. Workshop sponsored by the Center for Childhood Deafness and Hearing Loss: Using Sign Supported Speech to Improve Speech and English Grammar, Tacoma, WA.


Mayer, M. (2013, April). *Strategies to support listening, speech, and English with Signing Exact English*. Workshop presented by the Center for Childhood Deafness and Hearing Loss: Using Sign Supported Speech to Improve Speech and English Grammar, Tacoma, WA.


Hispanic Parents’ Beliefs, Attitudes, and Perceptions Toward Pediatric Hearing Loss: A Comprehensive Literature Review

Ana Caballero, MD, AuD¹,²
Karen Muñoz, EdD¹,²
Jared Schultz, PhD³
Laurel Graham, MEd¹
Alex Meibos, AuD, PhD¹

¹Department of Communicative Disorders and Deaf Education, Utah State University, Logan, UT
²National Center for Hearing Assessment and Management, Utah State University, Logan, UT
³Department of Special Education and Rehabilitation, Utah State University, Logan, UT

Abstract: Objective: The purpose of this review was two-fold: (a) to understand research findings about Hispanic parents’ beliefs, attitudes, and perceptions related to hearing loss after having children who are deaf or hard of hearing, and (b) to inform future research needs that could expand audiologists’ ability to provide patient-centered care with this population.

Design: A comprehensive review of the literature was used to identify relevant articles for the review.

Study Sample: Five research articles met the inclusion criteria.

Results: Three primary themes emerged: (1) deafness causality, parents tended to describe the cause of the hearing loss in terms of religion and folk beliefs; (2) cultural attitudes, parents reported having paternalistic views related to the care of their child and experiencing community stigma, and (3) cultural values, parents described how personalism, familism, fatalism, and respect informed their perspectives.

Conclusions: This comprehensive literature review found that limited research has been done to understand beliefs, attitudes, and perceptions of Hispanic parents toward pediatric hearing loss. Synthesis of five studies revealed important cultural factors for audiologists to consider in the provision of patient-centered care. Beliefs related to the cause of hearing loss, cultural values, and integration of children within the Hispanic community, may be critical elements for audiologists to address when promoting parental engagement.

Key Words: Hispanic, beliefs, attitudes, perceptions, hearing loss

Acronyms: ASHA = American Speech-Language-Hearing Association; DHH = deaf or hard of hearing; JCIH = Joint Committee on Infant Hearing; PRISMA = Preferred Reporting Items for Systematic Reviews and Meta-Analyses

Correspondence concerning this article should be addressed to: Ana Caballero, Department of Communicative Disorders and Deaf Education, Utah State University, 2620 Old Main Hill, Logan, UT 84322, USA.
E-mail: ana.caballero@usu.edu

Introduction

Hispanic people constitute 18% (58 million) of the population in the United States. The Hispanic population increased by over 40% between the years 2000 and 2010 and is expected to be at 119 million by 2060 (Colby & Ortman, 2015). Many (72%) Hispanics report Spanish as the primary language they use at home (U.S. Census Bureau, 2017), an important factor when considering educational supports for parents with a child who is deaf or hard of hearing (DHH). Based on the shift in demographics, it can be anticipated that audiologists will be offering services to this population more frequently.

Best practice guidelines for working with children who are DHH indicate the importance of providing services that are family-centered and culturally sensitive (Joint Committee on Infant Hearing [JCIH], 2013) to support family engagement in all aspects of service delivery (American Speech-Language-Hearing Association [ASHA], 2004). Language barriers and cultural differences between the provider and patient can compromise patient satisfaction, and possibly treatment outcomes, if patients feel their needs are not being addressed in a manner that acknowledges their cultural perspective (Abreu, Adriático, & DePierro, 2011). It is estimated that 13% of families living in the United States speak Spanish in the home.
as a primary language, but only 3.7% of audiologists in
the United States report being Spanish-English bilingual
(ASHA, 2017). Audiologists may find it challenging to
navigate cultural differences in addition to language
barriers in the delivery of services, requiring intentional
efforts on the part of audiologists to adequately address
the needs of Hispanic families.

Few studies have explored perspectives, experiences,
and needs of Hispanic parents of children who are DHH.
Research in pediatric audiology has primarily focused on
the experiences of families from the majority culture with
higher socioeconomic status (Muñoz, Blaiser, & Barwick,
2013; Muñoz, Preston, & Hicken, 2014; Muñoz et al.,
2015; Moeller, Hoover, Peterson, & Stelmachowicz, 2009;
Sjoblad, Harrison, Roush, & McWilliam, 2001; Walker
et al., 2013). One study, conducted by Caballero et al.
(2017), explored hearing aid management challenges and
support needs of Hispanic parents (N = 42) of children
birth to five years of age. Findings revealed that parents
wanted more support from their audiologist than they were
receiving, wanted more information and training, and that
they experienced challenges that interfered with how much
their child used their hearing aids. Given the importance
of family-centered care and the projected growth of the
Hispanic population in the United States, understanding
research findings related to the perspectives, beliefs, and
attitudes of Hispanic parents would benefit audiologists.
The purpose of this review was two-fold: (a) to understand
research findings about Hispanic parents’ beliefs,
attitudes, and perceptions related to hearing loss after
having children who are deaf or hard of hearing, and (b) to inform future research needs that could expand
audiologists’ ability to provide patient-centered care with
this population.

Method

A comprehensive review of the literature was conducted
to identify peer-reviewed journal articles published
on the beliefs, attitudes, and perceptions of Hispanic
parents related to pediatric hearing loss. The search
for relevant studies followed the PRISMA (Preferred
Reporting Items for Systematic Reviews and Meta-
Analyses) approach (Liberati et al., 2009), which
consists of four phases: (a) identification of research
papers through database searching; (b) screening of the
articles identified in the first phase; (c) eligibility of the
articles after full-text readings, and (d) inclusion in the
review of the articles that fit the criteria.

Procedures

Database searches were completed (CINAHL, ERIC,
PsycINFO, Medline, Psychology and Behavioral
Science, MedicLatina, and Fuente Académica) using
English search terms (“Hispanic OR Spanish OR Latin**”)
AND (“believe OR belief* OR attitude* OR perception*”)
AND (“Hearing loss OR deaf* OR hearing impair* OR hard of hearing”), and Spanish search terms:

(*“Hispanic* OR Latin***) AND (“creencia OR percepción* OR actitud*”) AND (“pérdida auditiva OR sordera OR trastorno auditivo”). Articles were included if they met the following criteria: (a) peer-reviewed research published from 1985 through 2017, (b) participant sample included Hispanic parents of children who are deaf or hard of hearing, and (c) research addressed attitudes, beliefs, or perceptions about hearing loss.

The database search identified 170 unduplicated journal
article abstracts. Title and abstract screening excluded
136, and full-text review excluded an additional 29
articles. Of those excluded by full-text review, 10 did
not include Hispanic participants, 7 did not include
hearing loss, 8 were not research studies, and 4 did not
address cultural beliefs, attitudes, or perceptions. The
complete search resulted in five peer-reviewed articles
that met the inclusion criteria. A qualitative assessment
was completed for all eligible articles, emergent
themes related to beliefs, attitudes, and perceptions of
Hispanic parents related to pediatric hearing loss were
synthesized, and a narrative summary was generated.

Results

All of the studies were published in English, and the
same author (Steinberg) wrote three of the five articles.
Three primary themes emerged (see Table 1): (a)
deafness causality, parents tended to describe the
cause of the hearing loss in terms of religion and folk
beliefs; (b) cultural attitudes, parents reported having
paternalistic views related to the care of their child and
experiencing community stigma, and (c) cultural values,
parents described how personalism, familism, fatalism,
and respect informed their perspectives. A discussion of
the five articles that met our inclusion criteria follows.

Steinberg and colleagues (1997)
The authors interviewed nine Hispanic families (the
majority were of Puerto Rican origin) to identify their
perceptions, attitudes, and beliefs about deafness and
disability. The interviews were conducted orally, in either
Spanish or English, based on the participant’s language
preference; most reported coming from large families
and having family support to raise their children. Sixty-
seven percent (n = 6) of the families referred to God
when explaining why their child or other children were
deaf. Other beliefs were related to the fact that God had
chosen their family to raise a child with hearing loss or
that God would restore the child’s hearing whenever
he might think it is the right time. Besides the belief
that God caused the child’s deafness, other causes
of hearing loss reported were noise from a plane or
hereditary causes when another diagnosis was provided
by the physician.

Parents described experiencing public stigma, feeling
as though their child was losing their identity in the
Hispanic community because others would refer to
their child as the “deaf one,” the “mute one,” or use the diminutive “little deaf one”. Some mothers reported that deaf children were viewed with pity by the Hispanic people they encounter (e.g., attaching the diminutive “-ito” or “-ita” when referring to the deaf child: “sordita”, “pobrecito” [little deaf one, poor little one]). Another element shared by the parents was that other Hispanics in the community tend to assume that hearing loss is associated with intellectual disabilities: “there are people who think that he is retarded because they don’t know him…” (p. 211).

Steinberg and colleagues (2003)
The researchers interviewed 29 Hispanic families living in four states (i.e., Pennsylvania, Texas, Central Florida, and Northern California) to explore their decision-making process after the identification of deafness. All parents had at least some high school education. Eighty percent of the children ($n = 23$) had severe to profound hearing loss, and three had known hereditary deafness. Four children (14%) had cochlear implants; technology use was not reported for the remaining children. The authors explored the impact of culture, language, and access to available information and services. Approximately half were given written materials about communication opportunities. The authors found that Hispanic parents’ decisions were often complicated by language and cultural barriers, as well as limited access to information in Spanish at an appropriate educational level. Parents reported that even when they expressed their desire to have their children learn Spanish, most followed the professional’s recommendations when making decisions about communication, with a combination of sign language and spoken English most often recommended by professionals. The study also found that mothers were the most involved in the decision-making process, followed by fathers, family doctor, grandparents, and friends. Findings related to
attitudes and beliefs about deafness showed 96% of the Hispanic families agreed with the statement, “Children should be taught sign language so they can talk to deaf people” (p. 6). Also, less than half of the participants agreed with the statement: “Deaf children should go to ordinary schools” (p. 6).

Palmer and colleagues (2008)
The authors presented baseline data collected from 139 parents of deaf children participating in a longitudinal, prospective study on genetic testing for connexin-related deafness. Forty-two percent \( (n = 59) \) of the parents were Hispanic living in Southern California, with the majority having a high school diploma or higher. The purpose of this study was to examine ethnic differences in parental perceptions of genetic testing for deaf infants. All parents in this study consented to genetic testing to understand the cause of their child’s deafness. Asians and Hispanic parents were more likely to perceive genetic testing in harmful terms (e.g., harmful effects on family, harmful to child, emotionally difficult to receive genetic information). Hispanic participants reported the most common reasons for genetic testing were related to etiology: to learn more about the baby’s deafness, to know the cause of the baby’s deafness, and to find out if the cause of the baby’s deafness was genetic. Furthermore, Hispanic parents attached more importance to family planning as a reason for genetic testing than did Caucasian parents, with the majority of the Hispanic parents (60%) feeling that a “very important” reason for their child to have genetic testing was for making decisions about whether to have more children in the future.

Steinberg and colleagues (2007)
The authors interviewed 24 parents whose children had been referred for genetic testing (but had not yet been tested) for hearing loss. Parents recruited for this study included 16 hearing, English-speaking parents; 5 hearing, Spanish-speaking parents; and three deaf parents. Participants represented a diverse range of ethnic, racial, and socioeconomic groups. The Spanish-speaking parents recruited for this study had a lower educational level compared to the English-speaking parents. The interview included asking parents questions related to their knowledge of genetics (e.g., “What did you think was the reason for getting genetic testing?”), questions specific to hearing loss and experiences with healthcare professionals (e.g., “What were you told at the time about the results of the testing?”), values regarding the presence of hearing loss in their children (e.g., “How do you think being deaf or hard of hearing is different from having a different kind of disability?”). Several themes emerged from parental narratives, such as misconceptions and limited knowledge about genetics and the nature of hearing loss being transmitted genetically, and perspectives on genetic testing. Parents also reported wanting to know the cause of their children’s hearing loss (e.g., feeling relief or responsible). All of the Hispanic parents in the study speculated divine intervention as the cause of hearing loss. Some parents considered their children’s hearing loss as a punishment from God while others considered the belief that God had chosen them as parents of a child with hearing loss, for a special purpose.

Guiberson (2013)
The author surveyed 71 highly educated Hispanic parents of children who are DHH from Spain, to obtain information related to four specific areas: (a) family supportiveness and involvement, (b) accessibility to services and information; (c) professional involvement and supportiveness, and (d) bilingual variables. The majority of children had severe-to-profound hearing loss, were using cochlear implants, and their parents had selected an oral-only mode of communication; approximately one-third of the parents chose to raise their children to be spoken-language bilingual (English/Spanish). Most parents indicated they believed it was beneficial for their child to be bilingual, and that children who are DHH are capable of becoming bilingual in spoken languages. Parents reported that their partners/spouses and the extended family were their major support system and were involved in the decision-making process. Regarding professional support and involvement, parents indicated that the highest level of support and involvement was from speech-language pathologists, audiologists, and deaf educators. Half of the parents reported they struggled and had to work hard to obtain information about their children’s options, and more than half said they wished professionals had provided more resources and offered more communication opportunities (e.g., oral vs. American Sign Language vs. cued speech) for their child.

Discussion
Given the importance of patient and family-centered care and the projected growth of the Hispanic population in the United States, audiologists can benefit from understanding how Hispanic parents of children who are DHH view hearing loss. This comprehensive literature review identified five studies that explored Hispanic parents’ beliefs, attitudes, and perceptions toward pediatric hearing loss. Three primary themes emerged from the review: deafness causality (cause of the hearing loss in terms of religion and folk beliefs); cultural attitudes (e.g., paternalistic views and community stigma); and cultural values (e.g., personalism, familism, fatalism, and respect). These themes offer insights that can help audiologists consider questions that may be important to raise with Hispanic parents of children who are DHH.

Deafness Causality: Religious and Folk Beliefs
How the cause of a chronic illness or disability is perceived culturally can have a significant impact on other aspects of the family’s and the community’s
opportunities to learn how to support their children and engagement in the process. Their willingness to explore process, including feeling respected, can influence their between the majority and minority cultures on the 1997). These perspectives may represent a mismatch or insulted by others (Groce, 1987; Steinberg et al., 2003) found evidence of paternalistic attitudes, studies in this review (Steinberg et al., 1997; Steinberg, Davilla, Collazo, Loew, & Fischgrund, 1997; Steinberg et al., 2007). Inherited disorders are frequently seen as being caused by a family curse, and a clinician’s suggestion to determine who is the carrier for a particular gene, could be interpreted as an attempt to identify who is at fault. Such interpretations from families, may further complicate attempts to encourage families to continue getting assistance (Steinberg et al., 2007). In this literature review, however, two studies (Palmer et al., 2008; Steinberg et al., 2007) found that parents wanted to identify the cause of the disability, and the authors indicated the importance parents placed on understanding their children’s cause of deafness, regardless of their religion, hearing status, or ethnic background.

Understanding parent perspectives on genetic testing for deafness is an important consideration within a patient-centered care model (Palmer et al., 2008). Furthermore, Steinberg et al. (2007) emphasized the need for audiologists to be sensitive to parents’ personal and sociocultural backgrounds when discussing genetic testing. Audiologists should provide emotional and educational support when parents are facing the possibility of having their child identified with a genetic cause for the hearing loss.

**Cultural Attitudes**

**Paternalistic views.** In Hispanic families, parents often overprotect their children with a disability, even when the person with a disability wants independence and to be included in society (Steinberg et al., 2003). Two studies in this review (Steinberg et al., 1997; Steinberg et al., 2003) found evidence of paternalistic attitudes, in which parents feel the disability is unacceptable, or they experience negative social pressure. Such feelings and experiences can interfere with their participation in programs and their engagement in the intervention process. Hispanic parents of young children may prefer to keep the child at home, unseen even by close family and neighbors, for various reasons such as embarrassment about their children’s condition or the desire to protect their children from being teased or insulted by others (Groce, 1987; Steinberg et al., 1997). These perspectives may represent a mismatch between the majority and minority cultures on the views of disabilities. How parents are supported in this process, including feeling respected, can influence their engagement in the process. Their willingness to explore opportunities to learn how to support their children and how to advocate for their children should be addressed in a way that incorporates their potential paternalistic tendencies.

**Community Stigma.** Within the Hispanic community, disability can be seen as a divine punishment, an inherited stigma, or as a result of family impurity. This can cause the family to feel deeply ashamed or even interfere with their ability to cope (Steinberg et al., 1997). Two of the studies in this review found parents experienced difficulty dealing with the community’s stigmatization (e.g., children referred to as “sordo” [the deaf one] rather than using their name), and cases in which the child’s own identity was lost due to the existence of the hearing loss (Steinberg, et al., 1997; Guiberson, 2013).

**Cultural Values**

**Familism (Familismo).** The Hispanic community places a strong importance on close family relationships. The extended family plays an important role in decision-making, whereas within the U.S. society, the nuclear family is more often primarily involved in making treatment decisions for their child. Hispanic parents, conversely, often make key decisions in consultation with older relatives, grandparents, uncles, and aunts. This is a critical factor to consider within service delivery, as it is also common for extended family members to have a primary role in the day-to-day care of the child, including emotional support and financial assistance (Groce, 1987). Four of the studies in this literature review found the family and extended family members were an important part of their social support (Guiberson, 2013; Palmer et al., 2008; Steinberg et al., 1997; Steinberg et al., 2003).

**Personalism (Personalismo).** Development of a personal relationship with the health provider (e.g., audiologists) is often desired, and also includes the physical space between patient and provider—body language is important (Christensen, 1992). A desire for this relationship preference was described by parents in relation to the management of their children’s hearing aids (Caballero et al., 2017). Guiberson (2013) detailed how parents reported the professional involvement; specifically, how the speech-language pathologists, audiologists, and deaf/special educators were highly involved, supportive, and their personal relationship helped them engage in making shared decisions regarding the children’s treatment.

**Fatalism (Fatalismo).** Hispanics are more likely than the majority culture to believe that all events are predetermined and inevitable because it is part of their destiny. When parents hold this belief, they may think chronic illnesses are determined by God, and therefore must be accepted and considered as a castigo divino (punishment) for personal or family members’ sins (Baquet & Hunter, 1995). Fatalism was a common
cultural value found in three of the articles included in this review (Steinberg et al., 1997; Steinberg et al., 2003; Steinberg et al., 2007). Audiologists need to take into consideration this cultural value because it might interfere with treatment adherence.

**Respect (Respeto).** Hispanics place importance on showing respect to authorities or knowledgeable figures, including healthcare providers. They are often presumed to know the answers, and because of this they will tend to follow whatever recommendations are suggested, even when it goes against their will. Two of the articles included in this review (Steinberg et al., 1997; Steinberg et al., 2003) reported on the importance of the value of respect within the treatment process. Steinberg et al. (2003) reported that even when parents had expressed their desire to have their children learn Spanish, most followed the professional's recommendations (respect) when making decisions about communication, agreeing to intervention that included spoken English and sign language. When audiologists are working with parents, they can support parent engagement by acknowledging and discussing what values are important to the family and letting those values guide the process.

There are limitations to this review; because of the scant research on this topic, findings should be interpreted with caution. The results of all the studies together represent 173 Hispanic families and cannot be generalized. Another limitation to highlight is that one study was primarily Puerto Rican parents, and another study was conducted in Spain. Furthermore, two of the studies were published more than ten years ago, and younger Hispanic parents' views may be different from older generations.

**Implications for Practice and Future Research Needs**

Familiarization with characteristics of the Hispanic culture can offer benefits to audiologists in development of culturally competent practices. Even though differences exist among Hispanic families, there are four values that are common within the Hispanic culture that may be helpful to consider when providing audiology services to this population (Antshel, 2002; Calzada, Tamis-LeMonda, & Yoshikawa, 2013; Inclan, 1990; Irving, Benjamin, & San-Pedro, 1999; Rhoades, Price, & Perigo, 2004; Zea, Quezada, & Belgrave, 1994).

Health communication is an essential component of service delivery, and patient-centered care requires audiologists to consider needs on an individual basis. Patient and family factors can differ in multiple ways, such as culture, English proficiency, health literacy, socioeconomic status, or education level. There is a difference between the U.S. general population and Hispanics in educational attainment for individuals 25 years and older. According to the U.S. Census Bureau (2017), 66% of the population 25 years and over of Hispanic origin had at least a high school diploma; compared to 93% of Non-Hispanic Whites alone. In the Caballero et al. study (2017), 72% of the primary caregivers did not complete high school. In this review, three studies reported participants had a low educational level (less than high school) compared to other ethnicities. This has important implications regarding how professionals provide information and support parent learning.

Culturally competent practices can help reduce disparities through culturally sensitive and unbiased care. This involves understanding and respecting the language, religion, beliefs, and cultural values (Anderson, Scrimshaw, Fullilove, Fielding, & Normand, 2003); and taking time to learn more about each family’s cultural background. The following suggestions may be helpful for audiologists when engaging families coming from a Hispanic background:

- **Ask families their thoughts on the cause of their children’s hearing loss:** Sensitively challenge misconceptions, take time during the appointment to address the causes of hearing loss and compare it to the families’ beliefs, and potentially incorporate information about the myths related to disabilities among the Hispanic community.
- **Involve extended families and friends:** Ask families how they would like their family members to be involved in the process (familism) and adjust the practices, to the extent possible, to reflect respect for their cultural preferences. For example, welcome extended family members with permission to the appointment, appointments can also be confirmed with extended family members (get their phone numbers as well) or teach other family members how to manage the children’s hearing devices (e.g., how to change a hearing aid battery).
- **Provide emotional support:** For Hispanic families, close and warm relationships are important, even at a professional level. Address parents’ emotional response to the hearing loss and treatment process. Families need to feel safe and free from judgment to share their challenges, beliefs, and thoughts.
- **Education opportunities:** Families vary in the extent they are aware of, know how to, and desire to take initiative to access educational resources. Having culturally appropriate resources in their native language can help families better understand the nature of hearing loss, treatment options, and communication opportunities available to their children; facilitating their ability to move forward.
Further research is needed that describes Hispanics beliefs, attitudes, and perceptions toward pediatric hearing loss in the United States. This information is important and can help audiologists improve service delivery for this population by providing a better understanding of parental experiences, beliefs about hearing loss, and important factors for making decisions for their children. Further, it would be beneficial to explore how audiologists are prepared to provide culturally competent services and opportunities to enhance training related to serving culturally and linguistically diverse families.

Conclusion

This comprehensive literature review found that limited research has been done to understand beliefs, attitudes, and perceptions of Hispanic parents toward pediatric hearing loss. Synthesis of five studies revealed important cultural factors for audiologists to consider in the provision of patient-centered care. Beliefs related to the cause of hearing loss, cultural values, and integration of children within the Hispanic community, may be critical elements for audiologists to address when promoting parental engagement. Audiologists need to consider the patients’ cultural background, and their level of education to implement shared treatment decisions.

References

Use of Hearing Protection in Neonatal Intensive Care Unit Patients: A Systematic Review of the Evidence

Nathalie Chouery, BS, BA1
Kathleen T. Dunckley, PhD1
1Rush University Medical Center

Introduction

Infants who require dedicated medical attention after birth are routinely admitted into a neonatal intensive care unit (NICU) due to maternal risk factors, delivery complications, and active disease. Often premature, these patients are not ready to cope with the external environment, let alone with stressors present in medical settings. Although necessary, the very medical care designed to sustain life may also pose a threat to the neurodevelopment and physiologic stability of neonates. Life-sustaining care is often accompanied by excessive levels of noise, which can lead to elevated stress responses in critically-ill infants. Developmentally supportive care has been proposed as a means to decrease negative effects on infants and optimize their development (Aucott, Donohue, Atkins & Allen, 2002) by regularly assessing the neurodevelopmental condition of an infant, minimizing environmental stressors, promoting infant rest, and maintaining a positive energy balance (Hamilton & Redshaw, 2009). Individual infant hearing protection has been proposed as a minimally-invasive and cost-effective way to reduce the effect of noise on infant stress response.

This systematic review was designed to assess the effectiveness of using individual infant hearing protection to reduce stress responses. The results of this analysis are particularly relevant to hearing healthcare providers due to their role as consultants on matters relating to noise, hearing protection devices (HPD), and creating NICU protocols. Audiologists perform follow up assessments for failed newborn hearing screens and to monitor for hearing changes in at-risk infants; as such, they will provide information to caregivers and pediatricians regarding ongoing risks.

Noise in the NICU is a pervasive environmental stressor to infants. For an infant with compromised medical status, noise exposure can be a hazard to their global health. Neonates in the NICU depend on healthcare professionals to identify and manage sources of stress, including noise. The American Academy of Pediatrics (AAP) lists validated and reliable behavioral indicators (e.g., body movement, crying, sleep) and physiologic indicators (e.g., changes in heart rate, respiratory rate, blood pressure, oxygen saturation, and cortisol levels) that can be used to assess and manage infant pain and stress (Lemons et al., 2000).

Abstract: Neonatal intensive care unit (NICU) settings present neonates with many environmental hazards, including exposure to dangerous sound intensity levels. Noise levels in NICUs worldwide overwhelmingly exceed the recommendations for safe exposure by the American Academy of Pediatrics. Environmental modifications and staff behavioral changes have proved ineffective to sufficiently reduce infant noise exposure. A systematic review of the literature was undertaken to answer if earmuffs improve physiologic stability, behavioral response, and sleep behavior, which are markers of stress response in NICU patients. Seven studies met the review's inclusion criteria and were examined for qualitative synthesis. This review supports using earmuffs to reduce neonate exposure to noise in the NICU as a viable intervention to improve physiologic stability and sleep and behavioral responses. Earmuffs are a minimally invasive, affordable, and effective option for attempting to comply with recommended noise guidelines. Moreover, earmuff use by NICU patients should be considered as a component of routine evidence-based practices when implementing development-centered care to minimize over-stimulation of NICU patients.

Key Words: NICU, noise, hearing protection, earmuff, infant, stress

Acronyms: AAP = American Academy of Pediatrics; ABR = auditory brainstem responses; ABSS = Anderson Behavioral State Scoring; HPD = hearing protection devices; NICU = Neonatal intensive care unit

Correspondence concerning this article should be addressed to: Kathleen Dunckley, PhD, Rush University Medical Center, Department of Communication Disorders and Sciences, Armour Academic Building, 600 S. Paulina St., Suite 1000, Chicago, IL 60612. Phone: 312-942-9787; Email: kathleen_t_dunckley@rush.edu
They recommend minimizing noxious environmental stimuli for all neonates and continuous use of pulse oximetry and frequently monitoring vital signs to detect stress. These sequelae of toxic stress in a newborn are all the more serious in NICU populations who are already at risk for developmental complications. Audiologists, in particular those working alongside NICU nursing staff, should be familiar with the noise stressors this environment presents in order to make recommendations that can mitigate infant noise exposure and associated stress responses.

Noise is one of the major iatrogenic environmental hazards neonates face in the NICU (Lai and Bearer, 2008). Sound levels measured in NICUs between 1979 and 2005 range from 70 dB up to 117 dB* (Brown, 2009). According to the U.S. Environmental Protection Agency (1974) the sound levels in hospital environments should not exceed 45 dB* in the daytime and 35 dB* at night. Although recently reported sound pressure levels vary tremendously (Pinheiro, Guinsburg, Nabuco, & Kakehashi, 2011) they overwhelmingly and consistently surpass the levels recommended by the AAP. Parra, de Suremain, Audeoud, Ego, and Debillon (2017), concluded that NICU noise exceeds recommended limits within incubators. This may seem counterintuitive given that incubators are viewed as safe spaces, but may be explained by reverberation, noise causing peak sounds inside the incubator, and equipment and infant noise. Despite several decades of continuous efforts to control sounds in NICU environments, the noise levels remain at levels that pose a threat to infants with compromised health.

The link between exposure to excessive noise and resulting stress responses is well known. Heart rate increased in both full-term and preterm infants in response to a 90 dB, 2.5-second buzzer (Field, Dempsey, Hatch, Ting, & Clifton, 1979), with only the full-term infants demonstrating an ability to habituate to the buzzer over time. Closely related to heart rate, oxygen saturation is the percentage of oxygen available in blood. According to the clinical guidelines of The Royal Children’s Hospital Melbourne (2016), oxygen saturation levels should be targeted within the range of 91–95% in both preterm and term neonates in order for the body and internal organs to perform essential functions. Noise may cause alteration in oxygen saturation and increased oxygen consumption secondary to elevated heart and respiratory rates (Morris & Bose, 2000; Wachman & Lahav, 2011), and may lead to a decrease in the amount of calories available for growth. Mean oxygen saturation in premature newborns during a daily, designated silence period (94.22%) were significantly higher than before (92.80%) the silence period (Taheri, Abbasi, Abdeyazdan and Fathizadeh, 2010). Infants housed within incubators causing high environmental noise (Parra et al., 2017) had increased heart rates and decreased oxygen saturation (Cardoso, Kozlowski, Lacerda, Marques, & Ribas, 2015). Slevin, Farrington, Duffy, Daly, & Murphy (2000) reported that neonates’ median diastolic blood pressure, mean arterial pressure, and infant movements were all reduced when the NICU environment was deliberately altered by reducing light, noise, infant handling, and staff activity for a specified time period. Lastly, weight gain is monitored closely in the NICU because promoting growth is a crucial aspect of managing these patients’ care and development. It is generally accepted that neonatal stress leads to energy expenditure, which in turn may result in altered growth and delayed discharge from the NICU (Farrell & Nicoteri, 2007). Availability of an intervention to reduce at least one source of infant stress and that could be used while the infant is housed in an incubator would lead to more stable physiologic response and increased weight gain. Given the obstacles to implementing silent periods and operating under the recommended noise levels, HPDs such as earmuffs can be considered a comparatively simple alternative. When NICU nurses were polled on the subject of earmuff use, 72% reported that NICU noise is too loud and 100% believed this intervention was not hazardous to the infants (Abdeyazdan, Ghassemi, & Marofi, 2014).

Compared to full term infants, the behavior of preterm infants is notable for signs of stress including motor responses (tone, activity, and posture) and states of central nervous system arousal (drowsiness, alertness, and crying; Mulligan LaRossa, 2018). The Anderson Behavioral State Scoring (ABSS) system is a measure that evaluates preterm infants’ behavioral states and provides information on infant sleep so sleep quality can be measured quantitatively (Burroughs, Asonye, Anderson, Shanklin, & Vidyasagar, 1978; Parmelee and Stern, 1972). Essential brain functions related to neonatal neurodevelopment take place during non-rapid eye movement period of sleep (NREM; Peirano, Algarin, & Uauy, 2003). Therefore, a variable of interest to monitor neurodevelopment is the total time spent in NREM sleep as measured by electroencephalograms.

In adults, when noise is combined with other factors, such as ototoxic medications, its potential to cause damage to hearing organs increases (Cone et al., 2017). This is true for infants as well. Bernard (1981) detected hearing loss using auditory brainstem responses (ABR) in preterm neonates exposed to aminoglycosides. The amount of shift in ABR wave V was correlated to the aminoglycoside dose administered per kilogram of body weight. In 2007, Rees confirmed that preterm infants in NICU who received aminoglycosides 7 or more days while exposed to noise levels produced by mechanical ventilation (> 80 dBA for > 30 minutes) had a 68% probability of developing hearing loss.

The direct connections between exposure to excessive environmental noise and increased stress in newborns highlight the need to implement sound reduction practices in NICUs. One tactic targets the professionals providing care to critically ill neonates. However, a dedicated education program for NICU staff failed to produce a...
sufficient or lasting change in NICU noise level (Degorre et al., 2017). Educating nurses about the link between elevated sound pressure levels and infant stress and implementing dedicated quiet time for high-risk preterm infants failed to reduce noise to within recommended levels (Laubach, Wilhelm, & Carter, 2014) by the AAP (Miquel, 2015). Practical changes have ranged from making alarms visual instead of auditory (Chang, Pan, Lin, Chang, & Lin, 2006), using single-patient room designs (Walsh, McCullough & White, 2006), to building entirely new NICUs with a focus on sound abatement (Williams, van Drongelen & Lasky, 2007). These attempts have not successfully reduced sound pressure levels to below < 45 dB across settings and in a consistent manner. Infants receiving care in the NICU are subject to environmental stressors, including exposure to sounds of high intensities that exceed the acceptable level of 45 dB recommended by the AAP (Committee on Environmental Health, 1997). Stressors can impact the neonate’s ability to maintain physiologic stability and can be deleterious to their behavioral responses and sleep. The efforts to reduce environmental and behavioral modification in NICUs have not been sufficient, consistent, or lasting and infants still need to be protected.

An individualized option to reduce infant exposure to noise is the use of personal hearing protection such as earmuffs. The most widely used HPD in routine infant care and research are the MiniMuffs® (Natus Medical Incorporated), supra-aural noise attenuators designed specifically for premature infants. Natus Medical Inc. states these attenuators reduce sound levels by at least 7 dB. An alternative to a supra-aural HPD is a silicone earplug that must be molded to the shape of the ear and inserted in the concha and auditory canal.

A systematic review of the literature was undertaken to determine if personal hearing protection use by NICU patients improves physiologic stability, behavioral response, and sleep behavior, which are markers of stress response. Initial review of the available literature identified one study using silicone earplugs in NICU infants (Turk, Williams, & Lasky, 2009). However, this study’s outcome measure was restricted to infant weight gain during the NICU stay and performance on developmental tests at 18–22 months of age. We wished to determine if evidence supports the use of HPD based on physiologic measures, which could justify the use of developmentally supportive care in NICUs. Therefore, Turk et al.’s conclusions were not included in further analyses.

**Method**

**Time Frame**
This review only included studies published between 1997 and April 2017. According to the Global Marketing department of Natus Medical Incorporated, the MiniMuff® has been on the market since 1993, without any major design changes since its release (G. Accetturo, personal communication, May 10, 2017).

**Search Strategy**
A systematic search of records was completed using six databases: CINAHL Complete, MEDLINE, Scopus, ComDisDome, Access Medicine, and Nursing Reference Center. The search queries were:

1. “(NICU) AND (noise reduction)"
2. “(NICU) AND (earmuff)"
3. “(NICU) AND (earplug)"

Secondly, the references cited by articles that met inclusion criteria (see Table 1) were searched to identify additional records, which yielded three results. Figure 1 depicts the complete search process. Study selection was guided by the “Preferred Reporting Items for Systematic Reviews and Meta-Analyses: The PRISMA Statement," by Moher, Liberati, Tetzlaff, Altman, & Prisma Group (2009). Article titles and abstracts were subjected to the inclusion criteria. The record screening was followed by reading the full texts to identify required inclusion criteria.

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![Figure 1. Identification of included studies.](image-url)
1997, use of MiniMuffs®, and published in peer-reviewed sources. Studies had to include either a physiologic and/or behavioral measure as an outcome measure. Physiologic measures included heart rate, respiratory rate, blood pressure, oxygen saturation, body temperature, weight gain, time in rapid eye movement (REM) sleep, and time in non-rapid eye movement sleep (NREM). Behavioral outcome measures included frequency of motor responses and scores on the ABSS (Burroughs et al., 1978; Parmelee & Stern, 1972). All articles were screened and reviewed by the first author to ensure all inclusion criteria were met.

**Level of Evidence and Quality Assessment**
Each study meeting inclusion criteria was assigned a level of evidence and study quality. Level of evidence was determined based upon the guideline published by The Oxford Centre for Evidence-Based Medicine (Phillips et al., 1998). The included studies were each categorized as a Level 1b for individual randomized controlled trial. Study quality was determined using the scheme developed by the American Speech-Language-Hearing Association (ASHA) National Center for Evidence-Based Practice in Communication Disorders (Cherney, Patterson, Raymer, Frymark, & Schooling, 2008). Records received a point for each quality indicator: assessor blinding, random sampling, group participant comparability, treatment fidelity, valid outcomes, significance, precision, and intent-to-treat. All included studies received 7 points, missing one point for assessor blinding.

### Table 1
**Systematic Review Inclusion and Exclusion Criteria**

<table>
<thead>
<tr>
<th>Component</th>
<th>Inclusion</th>
<th>Exclusion</th>
</tr>
</thead>
<tbody>
<tr>
<td>Population</td>
<td>Neonatal intensive care unit patients</td>
<td>Well-baby nursery patients, non-infant patients</td>
</tr>
<tr>
<td>Language</td>
<td>English</td>
<td>All other languages</td>
</tr>
<tr>
<td>Publication date</td>
<td>On or after 1997</td>
<td>1996 or earlier</td>
</tr>
<tr>
<td>Intervention</td>
<td>Noise exposure reduction using MiniMuffs® by Natus Medical Incorporated</td>
<td>Silicone ear plugs, other environmental or behavioral noise reduction strategies</td>
</tr>
<tr>
<td>Method</td>
<td>Published in peer-reviewed sources, meta-analyses, randomized controlled trials, cohort studies, case control, cross-sectional, retrospective and prospective studies</td>
<td>Theoretical papers, opinion-based editorials, reviews, qualitative studies, case studies, records with no statistical data reported, theses, and dissertations</td>
</tr>
<tr>
<td>Physiologic Outcome Measures</td>
<td>heart rate, respiratory rate, blood pressure, O₂ saturation, temperature, weight gain</td>
<td>Any physiologic or behavioral outcome measure obtained more than 1 day after intervention. Pain index.</td>
</tr>
<tr>
<td>Behavioral Outcome Measures</td>
<td>frequency of motor response, ABSS score, time in REM &amp; NREM sleep.</td>
<td></td>
</tr>
</tbody>
</table>

*Note: ABSS = Anderson Behavioral State Scoring; NREM = Non-rapid eye movement, REM = Rapid eye movement*

### Table 2
**Patient Characteristics**

<table>
<thead>
<tr>
<th>Authors</th>
<th>N</th>
<th>Gestational Age</th>
<th>Gender</th>
<th>Premature</th>
<th>APGAR at 5 min</th>
</tr>
</thead>
<tbody>
<tr>
<td>Abdeyazdan, Ghassemi, Marofi, &amp; Banjis (2014)</td>
<td>96</td>
<td>29–36 weeks</td>
<td>NR</td>
<td>Yes</td>
<td>≥ 7</td>
</tr>
<tr>
<td>Abdeyazdan, Ghassemi, &amp; Marofi (2014)</td>
<td>64</td>
<td>28–37 weeks</td>
<td>41 M</td>
<td>23 F</td>
<td>≥ 7</td>
</tr>
<tr>
<td>Abujarir et al. (2012)</td>
<td>100</td>
<td>&gt; 26 weeks</td>
<td>54 M</td>
<td>46 F</td>
<td>NR</td>
</tr>
<tr>
<td>Aita et al. (2013)</td>
<td>54</td>
<td>28–32 weeks</td>
<td>26 M</td>
<td>28 F</td>
<td>≥ 6</td>
</tr>
<tr>
<td>Duran et al. (2012)</td>
<td>20</td>
<td>29.9 ± 2.1 weeks</td>
<td>6 M</td>
<td>14 F</td>
<td>NR</td>
</tr>
<tr>
<td>Khalesi et al. (2017)</td>
<td>36</td>
<td>28–32 weeks</td>
<td>22 M</td>
<td>14 F</td>
<td>≥ 9</td>
</tr>
<tr>
<td>Varvara et al. (2016)</td>
<td>32</td>
<td>≥ 31</td>
<td>22 M</td>
<td>10 F</td>
<td>Not all</td>
</tr>
</tbody>
</table>

*Note: NR = not reported*
Data Synthesis
All studies in this review used the MiniMuff® as the HPD. Methods, outcome measures, and statistical reporting varied across the selected studies, which prevented the completion of a meta-analysis. Prematurity status was not homogeneous, intervention duration varied, noise level and total exposure differed, and dB weighing was not provided. Data were entered into summary tables and a narrative synthesis was used to determine if clinical use of earmuffs on NICU patients should be recommended. In particular, it was noted whether the authors of each study recommended or did not recommend the use of earmuffs.

Results
Seven studies met inclusion criteria. Table 2 summarizes patient characteristics (e.g., gestational age, gender) and Table 3 summarizes intervention variables and results (e.g., intervention schedule, outcome measures). Six

Table 3
**Intervention Variables and Results**

<table>
<thead>
<tr>
<th>Authors</th>
<th>Intervention Program</th>
<th>Intervention Schedule</th>
<th>Duration</th>
<th>Outcomes Measures</th>
<th>Results</th>
</tr>
</thead>
<tbody>
<tr>
<td>Abdeyazdan, Ghassemi, &amp; Marofi</td>
<td>1. Earmuffs on 2. Control 3. Silence (staff behavior modification)</td>
<td>9–11 AM and 4–6 PM for two consecutive days; 11 PM–5 AM for ten consecutive nights</td>
<td>68 hours</td>
<td>Weight gain; Frequency of MR (tremor, twitch, startle reflex)</td>
<td>1. 83.7 g weight gain; reduction in MR during and 1 hour after earmuffs</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>2. 7.94 g weight gain; increased MR</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>3. 59.1 g weight gain; reduction of MR only during silence period</td>
</tr>
<tr>
<td>Abdeyazdan, Ghassemi, &amp; Marofi</td>
<td>1. Earmuffs on 2. Control</td>
<td>9–11 AM and 4–6 PM for two consecutive days</td>
<td>8 hours</td>
<td>HR, arterial O₂ saturation, RR, and MR frequency</td>
<td>Intervention group had increased O₂ saturation, decreased RR and HR, and a reduction of MR. Control group had decreased O₂ saturation, increased HR, and increased number of MR.</td>
</tr>
<tr>
<td>Abujarir et al. (2012)</td>
<td>1. Earmuffs on 2. Control</td>
<td>From time of admission until 72 hours</td>
<td>72 hours</td>
<td>HR, systolic BP, diastolic BP, RR, temperature and O₂ saturation</td>
<td>Earmuff group had significantly lower HR and RR, and higher O₂ saturation and systolic BP. No significant difference in temperature control and diastolic BP.</td>
</tr>
<tr>
<td>Aita et al. (2013)</td>
<td>Eye goggles and earmuffs applied to infants in a crossover trial with a 20-hour wash out</td>
<td>6 AM–12 PM</td>
<td>8 hours</td>
<td>Mean/min/max HR, HR variability, O₂ saturation, ECG low frequency power (sympathetic activation), high frequency power (parasympathetic activation, RR)</td>
<td>Significantly higher maximum HR and lower HR power during intervention. No difference in mean HR, minimum HR, and O₂ saturation during control period.</td>
</tr>
<tr>
<td>Duran et al. (2012)</td>
<td>Earmuffs applied in crossover trial</td>
<td>2 days with and 2 days without earmuffs for 4 consecutive days</td>
<td>36 hours (48 hours in each condition)</td>
<td>Body temperature, BP, HR, RR, O₂ saturation and ABSS</td>
<td>Mean body temperature, HR, RR, BP, and O₂ saturation not statistically different between conditions. Infants with earmuffs were more frequently in quiet sleep and less frequently in active sleep. Infants without earmuffs were more frequently in awake or fussy/cry state.</td>
</tr>
<tr>
<td>Khalesi et al. (2017)</td>
<td>Earmuffs applied in crossover trial, infants served as own control.</td>
<td>8 AM–4 PM during two consecutive days</td>
<td>16 hours (8 hours in each condition)</td>
<td>Body temperature, HR, RR, systolic BP, diastolic BP, O₂ saturation, and ABSS</td>
<td>When wearing earmuffs infants had significantly lower RR and HR, higher O₂ saturation, and lower ABSS. More infants with earmuffs were in quiet sleep.</td>
</tr>
<tr>
<td>Varvara et al. (2016)</td>
<td>Day 1: baseline conditions  Day 2: earmuffs Day 3: incubator cover</td>
<td>8 AM–12 AM for three consecutive days</td>
<td>48 hours (16 hours each condition)</td>
<td>Time in Non-REM, REM, and total sleep duration.</td>
<td>Total time of Non-REM sleep was significantly higher with earmuffs than without. No significant difference in REM time or total sleep duration.</td>
</tr>
</tbody>
</table>

Note: MR = motor response; HR = heart rate; RR = respiratory rate; BP = blood pressure; ECG = electrocardiogram; ABSS = Anderson Behavioral State Scoring System; REM = rapid eye movement
studies reported physiologic outcome measures and five studies reported behavioral outcome measures. All but two studies (Abdeyazdan, Ghassemi, & Marofi, 2014; Abdeyazdan, Ghassemi, Marofi, & Berjis, 2014) reported that infants were cared for in incubators during the study periods, but did specify that infants were in the NICU of the hospital, not in individual rooms. Based on this information we presume the environment was similar for infants in all studies. The majority of studies made sound pressure level recordings (Abdeyazdan, Ghassemi, & Marofi, 2014; Abujarir, Salama, Greer, Al Thani, & Visda, 2012; Aita, Johnston, Goulet, Oberlander, & Snider, 2013; Duran et al., 2012; Varvara, Effrossine, Despoina, Konstantinos, & Matziou, 2016) all of which were above the recommended value of 45 dB SPL (Committee on Environmental Health, 1997). Khalesi, Khosravi, Ranjbar, Godarzi, & Karimi (2017) did not make sound pressure level recordings. Each of the included studies supported the use of earmuffs to protect infants in the NICU from excessive noise levels (Table 4).

Table 4
Evidence of Intervention Benefit and Recommendation

<table>
<thead>
<tr>
<th>Authors</th>
<th>Conclusions</th>
<th>Support/Reject interventions</th>
</tr>
</thead>
<tbody>
<tr>
<td>Abdeyazdan, Ghassemi, Marofi, &amp; Berjis (2014)</td>
<td>Both interventions led to fewer motor responses and improved weight gain. This effect was more pronounced in the earmuff group.</td>
<td>Support, particularly in wards facing executive problems with implementing silence</td>
</tr>
<tr>
<td>Abujarir et al. (2012)</td>
<td>Positive effect of wearing earmuffs in four common vital signs in sick newborn infants.</td>
<td>Support as routine care in the NICU.</td>
</tr>
<tr>
<td>Aita et al. (2013)</td>
<td>More stress responses when infants were wearing eye goggles and earmuffs than when not wearing them.</td>
<td>Reject wearing eye goggles and earmuffs as an intervention to reduce light and noise exposure.</td>
</tr>
<tr>
<td>Duran et al. (2012)</td>
<td>Noise reduction with earmuffs was associated with significant improvement in behavioral state of ABSS.</td>
<td>Support earmuff use to improve sleep efficiency and increase time of quiet sleep.</td>
</tr>
<tr>
<td>Varvara et al. (2016)</td>
<td>Providing low noise levels within the NICU will improve structural organization of sleep with more prolonged NREM periods.</td>
<td>Partially support intervention. Authors believe earmuffs are not practical clinical approach and that more research on sleep is needed.</td>
</tr>
</tbody>
</table>

Note: ABSS = Anderson Behavioral State Scoring System; NICU = neonatal intensive care unit; NREM = non-rapid eye movement

Discussion

Technological innovation and improvement in modern neonatal medicine have allowed for a reduction in the mortality rates of newborns that receive intensive care (Chow et al., 2015). Although overwhelmingly positive, these interventions have made the NICU a high sensory environment that may increase infants’ stress, placing their neurological development at increased risk. This systematic review focused on evaluating the use of HPDs to reduce the harmful effects of noise on the developing infant in the NICU. The seven studies included in this review reported physiologic stability and behavioral and sleep data during controlled periods of HPD (specifically earmuffs) use by NICU patients.

Physiologic Stability

Heart rate. Five out of the seven studies reported heart rate as an outcome measure. Out of those, three reported significantly lower heart rates during earmuff use compared to control periods without earmuffs (Abdeyazdan, Ghassemi, & Marofi, 2014; Abdeyazdan, Ghassemi, Marofi, & Berjis, 2014; Khalesi, Khosravi, Ranjbar, Godarzi, & Karimi, 2017). The remaining two studies found no difference in heart rate with and without earmuff use (Aita et al., 2013; Duran et al., 2012). Given that infants with lower Apgar scores (≤ 5 at 5 minutes) have more variable heart rate responses to sound, an intervention that stabilizes heart rate by reducing stress response to sounds is promising (White-Traut et al., 2009).

Respiratory rate. In the presence of persisting, inappropriately intense or complex stimulation, an increase in respiratory rate serves as an autonomic sign of stress (Emory University, n.d.). The same three studies listed reporting heart rate (Abdeyazdan, Ghassemi, & Marofi, 2014; Abdeyazdan, Ghassemi, Marofi, & Berjis, 2014; Khalesi, Khosravi, Ranjbar, Godarzi, & Karimi, 2017), reported significantly decreased respiratory rates when infants were protected from noise using earmuffs. A fourth study, Duran et al. (2012), found no difference between with and without earmuff conditions. Acutely ill infants, when exposed to continuous
stressors, experience increased heart and respiratory rates. Implementing earmuff use can significantly lower respiratory rates.

**Oxygen saturation.** Abdeyazdan, Ghassemi, & Marofi (2014a), Abdeyazdan, Ghassemi, Marofi, & Berjis (2014), and Khalesi et al. (2017) reported significantly increased oxygen saturation levels when infants were using earmuffs compared to no-earmuff conditions. Aita et al. (2013) and Duran et al. (2012) did not find significant differences in oxygen saturation levels with earmuff use.

**Blood pressure.** Three out of the seven selected articles measured infant blood pressure. One found a significant improvement (i.e., a decrease) in mean blood pressure, but not in diastolic blood pressure (Abujarir et al., 2012). Duran et al. (2012) and Khalesi et al. (2017) failed to find a significant effect on this vital sign with the use of earmuffs. One record is insufficient to validate blood pressure measurement as an acceptable and useful method of tracking physiologic stability as a result of earmuff use. Blood pressure change is not a good method of screening for illness and can be difficult to track reliably on neonates as artifact from movement and crying is common (Puchalski, 2011). Furthermore, the accuracy of selecting the appropriate upper arm, forearm, and calf cuff size for neonates by visual assessment is low (Devinck, Keukelier, Savoye, Desmet, & Smets, 2013).

**Temperature.** Three studies included body temperature as an outcome measure, yet none found this to be significantly impacted by earmuff use (Abujarir et al., 2012; Duran et al., 2012; Khalesi et al., 2017). Although body temperature is one of the fundamental variables for health monitoring of premature NICU infants, infant body temperatures are not yet locked to time of day as are adults’, and they vary with sleeping and feeding patterns (Anderson, Petersen, & Wailoo, 1990). As such, body temperature would not be a reliable indicator of improved physiologic stability as a result of earmuff use.

**Weight gain.** Abdeyazdan, Ghassemi, Marofi, & Berjis (2014) have been the only ones to document the effect of noise reduction by earmuff on weight gain in NICU patients. They demonstrated a meaningful and statistically significant increase in weight gain in the earmuff-wearing group when compared to non earmuff-wearing controls. It is important to note that this difference was between the first and the tenth day of the study. Although weight and weight gain are recorded accurately and often in NICUs, not all patients remain in intensive care for such a long duration. More research is needed to compare weight gain during shorter periods of time and to determine if this effect can be attributed to earmuff use.

**Electrocardiogram findings.** Aita et al. (2013) reported the low- and high-frequency power from the spectral analyses of electrocardiograms as an outcome measure. Preterm infants had lower high-frequency power while wearing earmuffs and eye goggles, suggesting that they had more stress responses, which was interpreted as higher parasympathetic activation. However, this relationship may be due to handling of the infants during the four-hour study periods. Touching the infants with their vision occluded may have exacerbated their stress response. According to the authors, high-frequency power is synchronous with respiratory rate. Respiratory and heart rate are more reliably measured and easier to obtain without the need of electrocardiogram equipment, so low- and high-frequency power would not be recommended as effective indicators of physiologic stability as a result of earmuff use.

**Summary of physiologic markers.** Heart rate, respiratory rate, and oxygen saturation level are good markers of the effect of HPD use and are vital signs routinely monitored in the NICU. Blood pressure and body temperature are unreliable indicators of stress minimization as an outcome of HPD use.

**Non-Vital and Behavioral Responses**

**Frequency of motor response.** Infants manifest behavioral stress responses to noise by moving, fussing and crying. Abdeyazdan, Ghassemi, & Marofi (2014) and Abdeyazdan, Ghassemi, Marofi, & Berjis (2014) found a significant reduction in motor responses (startle, tremor and twitch) during the use of earmuffs as compared to before use. Abdeyazdan, Ghassemi, & Marofi (2014) reported that this reduction continued immediately after and for one hour after earmuff use. Abdeyazdan, Ghassemi, Marofi, & Berjis (2014) reported a reduction in behavioral responses during the use of earmuffs. Reduced frequency of motor responses is an indicator of fewer stress reactions and decreased energy consumption.

**ABSS.** Two studies used the ABSS system to more formally describe infant behavioral responses. Duran et al. (2012) and Khalesi et al. (2017) reported that preterm infants using earmuffs were more frequently observed in the quiet sleep state of ABSS compared to those without earmuffs. Khalesi et al. (2017) concluded that the ABSS was lower for infants with earmuffs as compared to those without.

**NREM and REM data.** Varvara et al. (2016) employed a quantitative approach to measuring infant sleep quality. Infants’ time in NREM sleep increased when sound exposure was reduced, which in turn likely results in better neurodevelopmental outcomes for the patient and improved central nervous system maturation.

**Non-vital and behavioral summary.** Earmuff use by NICU patients leads to reductions in the frequency of motor responses, lower (i.e., improved) ABSS scores, and increased sleep efficiency.

**Aggregated Conclusions**

None of the studies using only earmuffs reported detrimental effects on the infants. Neonates showed improvement in physiologic and/or behavioral responses...
when earmuffs alone were used as an intervention to decrease stress responses. Earmuffs most often had a beneficial effect on heart rate, respiratory rate, and oxygen saturation levels. Aggregated conclusions support use of earmuffs to protect infants from complications of noise pollution in NICU. Furthermore, infants not wearing earmuffs were observed in some instances to have poor and/or unstable physiologic behavioral responses throughout the day. Abujarir et al. (2012) note that some vital signs do not differ significantly in the first six to twelve hours of earmuff use which may be because infants are handled more often upon NICU admission and that infants need more time for adaptation to the extra-uterine environment.

The study that used both eye goggles and earmuffs to reduce preterm infants’ exposure to light and noise does not recommend this intervention for neonatal practice (Aita et al., 2013). The authors note that controlling these patients’ exposure to noise and light remains an essential part of developmental care, but the combined use of goggles and earmuffs actually adds to infants’ stress.

Duran et al. (2012) state that earmuffs were helpful to improve sleep efficiency, which is important for the healthy neurodevelopment of this population in question. To that, Varvara et al. (2016) add that lower noise improves the structural organization of sleep, particularly NREM sleep, during which synapses are formed for specific functions and which constitutes an integral part of memory and learning process.

**Implementation**
Clinical interventions must be strongly guided by evidence that supports patient benefit as well as the practicality of its implementation. Abdeyazdan, Ghassemi, & Marofi (2014) measured NICU nurses’ attitude about the use of earmuffs in premature infants in order to lower the stress imposed by noise. The majority of nurses surveyed (72%) perceived the noise in the NICU to be too loud, believed earmuffs were beneficial to the infants (64%), and all of the staff agreed that earmuffs only interfered with routine care sometimes and posed no hazard to the infants.

Any change to the NICU environment requires time and teamwork. NICU staff may be more likely to accept and implement change when they have contributed to the change process (Bremmer, Byers, & Kiehl, 2003). Staff may also benefit from evidence-based education programs in order to promote behaviors and interventions that prevent over-stimulating NICU patients (Aita & Goulet, 2003).

**Conclusion**
Analysis of the seven studies that examined the efficacy of using neonatal earmuffs to reduce infant stress in the NICU support this as a viable intervention. None of the studies using only earmuffs reported detrimental effects on the infants. Neonates had improved physiologic stability, sleep, and behavioral responses. Earmuffs are a minimally invasive, affordable, and effective option for complying with recommended noise guidelines (Committee on Environmental Health, 1997). Moreover, earmuff use by NICU patients should be considered as part of the routine evidence-based practices when implementing development centered care. This endorsement is in line with recommendations by the American Academy of Pediatrics to evaluate and reduce the stress experienced by neonates using validated measures and appropriate environmental and nonpharmaceutical (behavioral) interventions (Lemon et al., 2000).

When striving to provide a developmentally supportive environment, facilities should consider the following physiologic measures as reliable indicators of stability: heart rate, respiratory rate, and oxygen saturation levels. Stable or improved responses should not be expected until at least six (but up to thirteen) hours after earmuffs are placed on the infant due to increased handling and need for habituation. Measurements should be made during earmuff use, not immediately after or hours later to accurately reflect changes in vital signs.

The ABSS can reliably track behavior and sleep information in infants using earmuffs, particularly when NICU staff adheres to its recommended schedule and administration instructions. Simultaneous earmuff and goggle use is not recommended until further research elaborates on the presumed increased stress response by infants of this dual stimulus reduction technique. However, incubator covers can be used in combination with earmuffs when it is necessary to minimize exposure to light.

Infants with the greatest number of risk factors for hearing loss are coincidentally the ones receiving specialized care in the NICU. A longer NICU stay means a longer duration to excessive noise. Currently, no studies have investigated whether a relationship exists between NICU noise, hearing loss, and auditory development.

Further research is needed to evaluate parameters beyond the benefit of earmuffs on infants’ immediate physiological and behavioral stability. Implementation in different facilities and across NICU levels would further refine the present recommendation. NICU staff perceptions of feasibility, practicality, job duty interruption and ability to adequately secure earmuffs on newborns should be examined.

A physiologic measure not evaluated by any of the seven included studies is salivary cortisol level. This biomarker of stress and/or diurnal rhythms can be collected from infants non-invasively and without causing additional stress (Neu, Goldstein, Gao & Laudenslager, 2007). Investigating cortisol level differences and stability during earmuff use would add to the existing knowledge of physiologic status. Finally, controlled trials should be conducted to assess the long-term effects of using earmuffs in the NICU on neurodevelopmental outcomes.


The Journal of Early Hearing Detection and Intervention (JEHDI) publishes peer-reviewed articles that describe current research, evidence-based practice, and standards of care specifically focused on newborn and early childhood hearing screening, diagnosis, support, early intervention, the medical home, information management, financing, and quality improvement that contribute to improving Early Hearing Detection and Intervention (EHDI) systems.

JEHDI is the only journal that focuses exclusively on improving EHDI systems, but many other journals include articles relevant to JEHDI's aim as part of their journal's broader focus. To help JEHDI readers stay up-to-date about current research and practices related to improving EHDI programs, we provide titles and abstracts of recent publications that JEHDI editors think are relevant to improving EHDI programs. Titles of all articles are hyperlinked to the source.

EHDI continues to be a global phenomenon with dozens of articles reporting on the status and progress of newborn hearing screening programs—from a systematic analysis of how Poland’s national universal newborn hearing screening program has evolved over the last 15 years to reports of emerging newborn hearing screening programs in Iran, Malaysia, and Botswana, among others. These reports emphasize the importance of tailoring screening and follow-up programs to local conditions and practices. There continues to be a major focus on targeted and universal screening for congenital cytomegolavirus and genetic mutations associated with hearing loss. A number of articles reported on successful efforts to screen and identify hearing loss in young children, recognizing that there are as many or more children who acquire hearing loss after the neonatal period as there are children with congenital hearing loss. The broadening of screening and identification programs to other conditions that are associated with hearing loss and to young children beyond the newborn period has important implications for how newborn hearing screening programs are best implemented and managed. Staying current with EHDI’s processes and accomplishments requires staying informed about what is happening in these closely related areas.

Below are some of the interesting findings from recently reported research studies from around the world.

- Lantos et al., in a health disparities study conducted in the United States, found that urban, low-income neighborhoods had a higher prevalence of infant hearing loss compared with more affluent surrounding communities, particularly among minorities.
- ElAlfy et al., in a study conducted in Egypt, found that iron deficiency anemia during late pregnancy adversely affected the newborn’s hearing status. They recommended antenatal screening of pregnant mothers to improve fetal iron status and prevent abnormal auditory maturation for the newborn.
- Rahimi et al., in a five-year study conducted in Iran with 4,729 children, reported that doing hearing screening during a neonatal thyroid screening program done during the third to fifth day after birth, significantly improved hearing screening outcomes. They concluded that this approach could be used as an alternative to hospital-based newborn hearing screening in many countries.
- Banda et al., demonstrated the importance of screening children for hearing loss after the newborn period by reporting on the high incidence of hearing loss among children 10 years of age and younger who were referred to a public audiology clinic in Botswana.
- Hao et al., based on a cohort of 142,417 neonates in China who were screened for common genetic mutations associated with hearing loss, concluded that genetic screening for hearing loss was practical in a large-scale community setting.
- Cejas et al., reported on a longitudinal study of 147 children with cochlear implants and 75 typically hearing peers. They found that children using cochlear implants perform similarly to hearing peers on measures of intelligence, but those with severe comorbidities are at-risk for cognitive deficits.
- Fowler and Boppana provide an excellent overview of congenital cytomegolavirus, how it is related to childhood hearing loss, and why it should be carefully considered as a part of early hearing detections and intervention systems.

Abstracts of many more articles with results that are important for continuing to improve EHDI programs are listed below.

**BACKGROUND:** Congenital cytomegalovirus (CMV) infection (cCMV) is an important cause of hearing loss and cognitive impairment. Prior studies suggest that HIV-exposed children are at higher risk of acquiring cCMV. We assessed the presence, magnitude and risk factors associated with cCMV among infants born to HIV-infected women, who were not receiving antiretrovirals during pregnancy.

**METHODS:** cCMV and urinary CMV load were determined in a cohort of infants born to HIV-infected women not receiving antiretrovirals during pregnancy. Neonatal urines obtained at birth were tested for CMV DNA by qualitative and reflex quantitative real-time polymerase chain reaction.

**RESULTS:** Urine specimens were available for 992 (58.9%) of 1684 infants; 64 (6.5%) were CMV-positive. Mean CMV load (VL) was 470,276 copies/ml (range: < 200-2,000,000 copies/ml). Among 89 HIV-infected infants, 16 (18%) had cCMV versus 42 (4.9%) of 858 HIV-exposed, uninfected infants (P < 0.0001). cCMV was present in 23.2% of infants with in utero and 9.1% infants with intrapartum HIV infection (P < 0.0001). Rates of cCMV among HIV-infected infants were 4-fold greater (adjusted OR, 4.4; 95% CI: 2.3-8.2) and 6-fold greater among HIV in utero-infected infants (adjusted OR, 6; 95% CI: 3-12.1) compared with HIV-exposed, uninfected infants. cCMV was not associated with mode of delivery, gestational age, Apgar scores, 6-month infant mortality, maternal age, race/ethnicity, HIV viral load or CD4 count. Primary cCMV risk factors included infant HIV-infection, particularly in utero infection.

**CONCLUSION:** High rates of cCMV with high urinary CMV VL were observed in HIV-exposed infants. In utero HIV infection appears to be a major risk factor for cCMV in infants whose mothers have not received combination antiretroviral therapy in pregnancy.


**OBJECTIVES:** Preschool children hearing impairment is a common otologic diseases worldwide. The burdens of this preventable condition can be reduced in developing country. This study was carried out to determine the prevalence, diagnosis and management of hearing impairment among children less than 5 years seen in our center.

**METHOD:** This was a prospective hospital based study of preschool children with complaints of hearing impairment in Ekiti state university teaching hospital, Nigeria. This study was conducted over a period of two years (February 2016 to January 2018). Data was obtained from consented patients by using pretested interviewers assisted questionnaire. Data obtained was collated and analyzed by using SPSS software version 18.0. Descriptive statistics was used to present and expressed the data as simple tables and charts.

**RESULTS:** The prevalence of hearing impairment in this study was 5.9% with peak value of 37.6% at preschool age 3 years. There were 42.6% males and 57.4% females. Majority (51.5%) of the patients were urban dwellers. The aetiological factors of hearing impairment include; 15.8% neonatal jaundice, 13.9% febrile illness, 12.9% otitis media and 11.9% birth asphyxia. Prelingual presentation was commonest in 57.4%. Common clinical features includes yet to speak, restlessness/stubborn and not responding to command in 60.4%, 48.5% and 46.5% respectively. Commonest type of hearing impairment was sensorineural hearing loss is 61.4%. Type A tympanometry (normal) was the commonest findings in 82.2%.

**CONCLUSION:** Preschool children hearing impairment with late presentation is a common otologic diseases burden to parents and otorhinolaryngologist in our center. The common causes were preventable with resulting permanent auditory damage.


**BACKGROUND:** Congenital deafness is the commonest birth defect and it affects 2-4 neonates among 1000 live births. Detection and intervention especially before 6 months of age prevents severe linguistic, educational and psychosocial repercussions and helps the deaf child in the development of normal speech and language. Children who are identified after 6 months of age experience great difficulties in attaining speech and language.

**METHODS:** To find out the frequency of hearing loss in neonates, a hospital based observational study was conducted in Combined Military Hospital Abbottabad from June to December 2014. One thousand new-borns selected by consecutive sampling within a specified period of time were subjected to Otoacoustic Emission (OAE) testing. Brain Evoked Response Audiometry (BERA) evaluation was performed in all those who failed OAE testing to confirm the hearing loss. Children born with microtia, mental stenosis, cleft palate, craniofacial abnormalities and syndormic inllnesses were excluded from the study.

**RESULTS:** Of 1000 newborns screened, 465 were males and 535 were females whereas 632 (63.2%) were delivered through C-section and 368 (36.8%) were born via SVD. Four hundred and ninety-one (49%) babies had a positive history of consanguinity among the parents. Out of 1000 infants 13 were having hearing loss which was later on confirmed on BERA evaluation. Among them 7 were males and 6 females, 9 (69%) were born through SVD and 4 (31%) through C-section and 8 (61.5%) new-borns had a positive history of consangunuity among their parents. In all these 13 patients only 2 (15%) patients had profound while the remaining 11 (85%) had moderate to severe hearing loss.

**CONCLUSIONS:** Frequency of hearing loss in neonates is much higher in our study (13 per 1000) as compared to other parts of the world and demands that more studies should be undertaken on this subject to confirm this.
Almosnino G, Anne S, Schwartz SR.

**OBJECTIVES:** Evaluate usage trends of neural response telemetry (NRT) in cochlear implant centers across the nation and assess reported benefits of intraoperative NRT for pediatric cochlear implant recipients.

**STUDY DESIGN:** Survey.

**STUDY PARTICIPANTS:** All US cochlear implant centers (n = 110).

**METHODS:** A 15-question multiple-choice survey was distributed electronically to all centers. The survey captured demographic information of all centers, practice patterns surrounding the use of NRT, and the extent to which intraoperative NRT is of benefit.

**RESULTS:** Thirty-two invited participants (29%) completed the survey. A majority of participants reported practicing in an academic center (66%), followed by a hospital setting (19%) and private practice (16%). Seventy-two percent of survey participants reported using NRT for pediatric cochlear implant recipients. Sixty-three percent felt it improved the ability to program at initial activation, and 50% of participants felt that NRT improves satisfaction at initial activation.

**CONCLUSION:** This study suggests that a majority of surgeons use intraoperative NRT for pediatric cochlear implantation as an additional measure to ensure appropriate electrode placement and improve device activation. Larger studies are needed to better establish the relationship between intraoperative NRT and postoperative outcomes and justify the additional costs associated with intraoperative NRT.

Banda FM, Powis KM, Mokoka AB, Mmapetla M, Westmoreland KD, David T, Steenhoff AP.

**Objective.** To describe and quantify hearing impairment among children referred to the audiology clinic in Princess Marina Hospital, a public referral hospital in Botswana. **Methods.** In a retrospective case series, we reviewed medical records of children aged 10 years and younger whose hearing was assessed between January 2006 and December 2015 at the audiology clinic of Princess Marina Hospital in Gaborone, Botswana. **Results.** Of 622 children, 50% were male, and median age was 6.7 years (interquartile range = 5.0-8.3). Hearing impairment was diagnosed in 32% of clinic attendees, comprising sensorineural (23%), conductive (25%), and mixed (11%) hearing loss, while 41% of children with diagnosed hearing impairment did not have a classification type. Hearing impairment was mild in 22.9%, moderate in 22.4%, severe in 19.4%, profound in 16.9%, and of undocumented severity in 18.4%. Children younger than 5 years were 2.7 times (95% confidence interval = 1.29-5.49; \( P = .008 \)) more likely to be diagnosed with sensorineural hearing impairment compared with those older than 5 years. By contrast, children older than 5 years were 9.6 times (95% confidence interval = 2.22-41.0; \( P = .002 \)) more likely to be diagnosed with conductive hearing loss compared with those under 5 years. **Conclusion.** Hearing impairment was common among children referred to this audiology clinic in Botswana. Of those with hearing impairment, more than a third had moderate or severe deficits, suggesting that referrals for hearing assessments are not occurring early enough. Hearing awareness programs individually tailored to parents, educators, and health care workers are needed. Neonatal and school hearing screening programs would also be beneficial.

Bartlett AW, Hall BM, Palasanthiran P, McMullan B, Shand AW, Rawlinson WD.

**BACKGROUND AND OBJECTIVES:** Australian national surveillance data was used to assess recognition, sequelae, and antiviral therapy for congenital cytomegalovirus (CMV) cases.

**STUDY DESIGN:** Data from congenital CMV cases reported through the Australian Paediatric Surveillance Unit born January 1999 to December 2016 were described and Chi-square tests used to characterise trends and associations in case reporting, maternal CMV serology testing, and antiviral therapy. Descriptive analyses for hearing loss and developmental delay were reported for cases born ≥2004, following introduction of universal neonatal hearing screening.

**RESULTS:** There were 302 congenital CMV cases (214 symptomatic, 88 asymptomatic). Congenital CMV was suspected in 70.6% by 30 days of age, with no differences across birth cohorts. Maternal CMV serology testing was associated with maternal illness during pregnancy but not birth cohort. There was increasing antiviral use for symptomatic cases, being used in 14% born 1999-2004, 19.6% born 2005-2010, and 44.4% born 2011-2016 (\( p <0.001 \)). For those born ≥2004, hearing loss was reported in 42.1% of symptomatic and 26.6% of asymptomatic cases; while developmental delay was reported in 16.9% of symptomatic and 1.3% of asymptomatic cases.

**CONCLUSION:** There appears to be under-reporting and under-recognition of congenital CMV despite increasing use of antiviral therapy. Universal newborn CMV screening should be considered to facilitate follow-up of affected children and targeted linkage into hearing and developmental services, and to provide population-level infant CMV epidemiology to support research and evaluation of antiviral and adjunctive therapies.
Bhind A, Carpineta L, Qassabi B, Waissbluth S, Ywakim R, Manoukian JJ, Nguyen LHP. 


**INTRODUCTION:** Temporal bone fractures (TBF) are traditionally classified by their angle of fracture relative to the petrous ridge, and more recently by whether or not they violate the otic-capsule. This study compared rates of hearing loss (HL) and signs of otologic dysfunction among fracture types of both classification systems, within the pediatric population.

**METHODS:** Pediatric patients were retrospectively characterized from a previously identified cohort of TBF patients, diagnosed from 2000 to 2014. CT scans were reviewed and TBFs were classified first as longitudinal (L), transverse (T) or mixed (M), and then as otic-capsule sparing (OCS) or otic-capsule violating (OCV). Medical records were reviewed, and rates of HL and presenting signs were compared among L, T and M fractures, and OCS and OCV fractures.

**RESULTS:** Forty-three patients with 47 TBFs met the inclusion criteria. Eighteen, 4 and 25 TBFs were classified as L, T and M fractures, respectively. Thirty-three and 9 were classified as OCS, and OCV, respectively. Among 24 cases of HL: 20, 3, and 1 were conductive HL (CHL), sensorineural HL (SNHL) and mixed HL, respectively. Two cases of SNHL were found among OCV fractures, with none in OCS fractures (estimated difference 0.22; 95% confidence interval 0.01-0.60). Similar rates of CHL were found across L, T and M fractures (range 36-50%), and across OCV and OCS fractures (range 42-44%). Hemotympanum was the most common presenting sign, found in 68% of TBFs and 80% of CHL cases. There were no significant differences in the incidence of signs or symptoms between fracture types.

**CONCLUSIONS:** In our cohort, both the traditional and otic-capsule radiographic classification systems failed to predict the incidence of CHL and other otologic signs in the pediatric population. Though OCV fractures conferred an increased risk for developing SNHL, we found a lower incidence than anticipated given violation to the bony labyrinth.

Boskabadi H, Zakerihamidi M, Moradi A, Bakhshaee M. 


**Introduction:** Hyperbilirubinemia is a common neonatal problem with toxic effects on the nervous system that can cause hearing impairment. This study was conducted to assess the risk factors for sensorineural hearing loss and other coexisting problems in icteric infants.

**Materials and Methods:** In a case-control study, 200 term infants with bilirubin levels higher than 20 mg/dl admitted to the neonatal intensive care unit of Ghaem Hospital, Mashhad during 2007-2015 were investigated. Profiles of infants with hearing impairment (n=60) were compared with those of icteric newborns with normal hearing (140 newborns) as the control group. After confirming the clinical diagnosis of jaundice by laboratory findings, a validated questionnaire containing mother and infant profiles were used for data collection. The auditory brainstem response test was used for assessment of infant hearing status after discharge.

**Results:** Sensorineural hearing loss among infants with severe hyperbilirubinemia was found to be 4.8%. Serum total bilirubin (P=0.001), creatinine levels (P=0.002), direct Coombs test results (P=0.001), etiology (P=0.000) and treatment for jaundice (P=0.000), eye movement disorders (P=0.001), opisthotonos (P=0.001), and microcephaly (P=0.001) were found to be significantly different between the two groups (P<0.005). The prognostic predictability of sensorineural hearing loss based on total bilirubin level was found to be 82%.

**Conclusion:** Hearing impairment occurs about 10-50 times more frequently in neonates with severe jaundice. Total bilirubin level has the highest predictability for infant hearing status. Blood group and Rhesus (Rh) incompatibilities between mother and child and G6PD deficiency are important known causes for hearing impairment due to jaundice.

Chakrabarti S. 


**OBJECTIVE:** Prelingual deafness in children demands urgent action as best outcome is dependent on earliest possible diagnosis and intervention. Objective of this study was to determine age of suspicion, diagnosis, intervention, and outcome in a representative group of deaf children in West Bengal, India, and suggest ways of improving these parameters.

**METHODS:** In this cross-sectional study, ages of suspicion, diagnosis, intervention and outcome of 303 randomly selected deaf children were elicited from a cohort of 1316 children with deafness identified in an earlier study.

**RESULTS:** Median ages of suspicion, diagnosis and amplification were 18, 72 and 84 months respectively. Age of suspicion was significantly related to parental education (p < 0.05); age of diagnosis to parental education and socio-economic status (p < 0.001) and children's geographic location (p < 0.01). Following diagnosis, 86% of children received hearing aids but only 6% used their aids consistently; 86% were non-verbal, 12% could communicate with a mixture of speech and gesture and only 2% with speech alone.

**CONCLUSION:** Current situation of deaf children in West Bengal, and evidence indicates, in much of India, is insupportable. However, widely diverse socio-economic conditions and scarcity of public health infrastructure preclude one solution of the problem for the whole country. In absence of the ideal universal newborn hearing screening, rigorously monitored and costed pilot programs of different models of early detection and intervention using newborn hearing screening, targeted screening and trial of calibrated noisemakers by primary care workers should be tried to see which works best where, so that successful programs can be scaled up over time.
Chen JY, Yang J.
International consensus (ICON) on audiological assessment of hearing loss in children. Lin Chng Er Bi Yan Hou Tou Jing Wai Ke Za Zhi.

Summary: The prevalence of hearing loss in newborns and infants is estimated between 1 to 3.47 cases per 1000 live births. Neonatal screening for hearing loss and audiological evaluation are becoming more extensively carried out. However, there is no consensus regarding the use of audiometry and other electrophysiological tests in current practices. This article is intended to provide professionals with recommendations about the “best practice” based on consensus opinion of the session’s speakers, and a review of the literature on the efficacy of various assessment options for children with hearing loss.

Cevizci R, Dilci A, Celenk F, Karamert R, Bayazit Y.
Surgical considerations and safety of cochlear implantation in otitis media with effusion. Auris Nasus Larynx.

OBJECTIVE: To evaluate the effects of otitis media with effusion on surgical parameters, patient safety, perioperative and postoperative complications.

METHODS: Total 890 children who underwent cochlear implantation between 2006 and 2015 were included. The ages ranged from 12 months to 63 months (mean: 32 months). The patients were divided into two groups according to the presence or absence of otitis media with effusion: otitis media with effusion group and non-otitis media group.

RESULTS: Of 890 children, 105 had otitis media with effusion prior to surgery. In non-otitis media group, there were 785 children. The average duration of surgery was 60min (ranged from 28 to 75min) in non-otitis media group, and 90min (ranged from 50 to 135min) in otitis media with effusion group (p<0.05). Granulation tissue and edematous middle ear and mastoid mucosa were observed in all cases of otitis media with effusion during the surgery. There was no significant difference between the complications of groups with or without otitis media with effusion (p>0.05). In 5 of 105 patients, there was a ventilation tube inserted before cochlear implantation, which did not change the outcome of implantation.

CONCLUSION: There is no need for surgical treatment for otitis media with effusion before implantation since otitis media with effusion does not increase the risks associated with cochlear implantation. Operation duration is longer in the presence of otitis media with effusion. However, otitis media with effusion leads to intraoperative difficulties like longer operation duration, bleeding, visualization of the round window membrane, cleansing the middle ear granulations as well as mastoid and petrous air cells.

Chiong CM, Reyes-Quintos MRT, Yarza TK, Tobias-Grasso CAM, Acharya A, Leal SM, Mohlke KL, Mayol NL, Cutiongco-de la Paz EM, Santos-Cortez RLP.
The SLC26A4 c.706C>G (p.Leu236Val) Variant is a Frequent Cause of Hearing Impairment in Filipino Cochlear Implantees. Otol Neurotol.

HYPOTHESIS: Variants in SLC26A4 are an important cause of congenital hearing impairment in the Philippines.

BACKGROUND: Cochlear implantation is a standard rehabilitation option for congenital hearing impairment worldwide, but places a huge cost burden in lower-income countries. The study of risk factors such as genetic variants that may help determine genetic etiology of hearing loss and also predict cochlear implant outcomes is therefore beneficial.

METHODS: DNA samples from 29 GJB2-negative Filipino cochlear implantees were Sanger-sequenced for the coding exons of SLC26A4. Exome sequencing was performed to confirm results.

RESULTS: Four cochlear implantees with bilaterally enlarged vestibular aqueducts (EVA) were homozygous for the pathogenic SLC26A4 c.706C>G (p.Leu236Val) variant, which has a minor allele frequency of 0.0015 in Filipino controls. In patients with the SLC26A4 variant there was no association between cochlear implant outcome and age at implantation or duration of implant. There was also no association between the occurrence of the SLC26A4 variant and postsurgical audiometric thresholds and parents’ evaluation of aural/oral performance of children (PEACH) scores. On the other hand, the SLC26A4 variant increased presurgical median audiometric thresholds (p=0.01), particularly at 500 to 2000Hz.

CONCLUSION: The SLC26A4 c.706C>G (p.Leu236Val) variant is a frequent cause of congenital hearing impairment in Filipinos and is associated with bilateral EVA and increased presurgical audiometric thresholds, but does not adversely affect post-implant outcomes.

Chung YS, Park SK.
2018 Sep 14. doi: 10.4178/epih.e2018044. [Epub ahead of print]

Objectives: To analyze the current status and problems of hearing screening tests for newborns of low income class in the southeastern region of Korea.

Methods: This study analyzed the data of the Ministry of Health and Welfare’s project on the early detection of hearing loss in low income class newborns from the southeastern region of Korea (2011 to 2015).

Results: The referral rate was 1.33%, 1.69%, and 1.27% in Daegu, Gyeongbuk, and Ulsan, respectively. The confirmation test rate was 36.09%, 23.38%, and 52.94% in Daegu, Gyeongbuk, and Ulsan, respectively. The incidence of hearing loss (adjusted) was 0.41%, 0.62%, and 0.41% in Daegu, Gyeongbuk, and Ulsan, respectively. After confirming the hearing loss, newborns with hearing handicaps were mostly lost to follow up, and rehabilitation, such as hearing aids or cochlear implants, were not used. The screening tests were performed within one month of birth, and the confirmation tests were performed within three months of birth. On the other hand, the groups with the risk factor for hearing loss took more than three months to reach the

**OBJECTIVES:** To evaluate the auditory performance and speech production outcome in children with auditory neuropathy spectrum disorder (ANSD). The effect of age on the outcomes of the surgery at the time of implantation was also evaluated.

**METHODS:** Cochlear implantation was performed in 136 children with bilateral severe-to-profound hearing loss due to ANSD, at four tertiary academic centers. The patients were divided into two groups based on the age at the time of implantation; Group I: Children ≤24 months, and Group II: subjects >24 months. The categories of auditory performance (CAP) and speech intelligibility rating (SIR) scores were evaluated after the first and second years of implantation. The differences between the CAP and SIR scores in the two groups were assessed.

**RESULTS:** The median CAP scores improved significantly after the cochlear implantation in all the patients (p value < 0.001). The improvement in the CAP scores during the first year in Group II was greater than Group I (p value: 0.007), but the improvement in CAP scores tended to be significantly higher in patients who were implanted at ≤24 months (p value < 0.001). There was no significant difference between two groups in SIR scores at first-year and second-year follow-ups. The evaluation of the SIR improvement revealed significantly higher values for Group I during the second-year follow-up (p value: 0.003).

**CONCLUSION:** The auditory performance and speech production skills of the children with ANSD improved significantly after cochlear implantation, and this improvement was affected by age at the time of implantation.

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Deshpande AK, Tan L, Lu LJ, Altaye M, Holland SK.


**BACKGROUND:** The trends in cochlear implantation candidacy and benefit have changed rapidly in the last two decades. It is now widely accepted that early implantation leads to better postimplant outcomes. Although some generalizations can be made about postimplant auditory and language performance, neural mechanisms need to be studied to predict individual prognosis.

**PURPOSE:** The aim of this study was to use functional magnetic resonance imaging (fMRI) to identify preimplant neuroimaging biomarkers that predict children’s postimplant auditory and language outcomes as measured by parental observation/reports.

**RESEARCH DESIGN:** This is a pre-post correlational measures study.

**STUDY SAMPLE:** Twelve possible cochlear implant candidates with bilateral severe to profound hearing loss were recruited via referrals for a clinical magnetic resonance imaging to ensure structural integrity of the auditory nerve for implantation.

**INTERVENTION:** Participants underwent cochlear implantation at a mean age of 19.4 mo. All children used the advanced combination encoder strategy (ACE, Cochlear Corporation™, Nucleus® Freedom cochlear implants). Three participants received an implant in the right ear; one in the left ear whereas eight participants received bilateral implants. Participants’ preimplant neuronal activation in response to two auditory stimuli was studied using an event-related fMRI method.

**DATA COLLECTION AND ANALYSIS:** Blood oxygen level dependent contrast maps were calculated for speech and noise stimuli. The general linear model was used to create z-maps. The Auditory Skills Checklist (ASC) and the SKI-HI Language Development Scale (SKI-HI LDS) were administered to the parents 2 yr after implantation. A nonparametric correlation analysis was implemented between preimplant fMRI activation and postimplant auditory and language outcomes based on ASC and SKI-HI LDS. Statistical Parametric Mapping software was used to create regression maps between fMRI activation and scores on the aforementioned tests. Regression maps were overlaid on the Imaging Research Center infant template and visualized in MRicror.

**RESULTS:** Regression maps revealed two clusters of brain activation for the speech versus silence contrast and five clusters for the noise versus silence contrast that were significantly correlated with the parental reports. These clusters included auditory and extra-auditory regions such as the middle temporal gyrus, supramarginal gyrus, precuneus, cingulate gyrus, middle frontal gyrus, subgyral, and middle occipital gyrus. Both positive and negative correlations were observed. Correlation values for the different clusters ranged from -0.90 to 0.95 and were significant at a corrected p value of <0.05. Correlations suggest that postimplant performance may be predicted by activation in specific brain regions.

**CONCLUSIONS:** The results of the present study suggest that (1) fMRI can be used to identify neuroimaging biomarkers of auditory and language performance before implantation and (2) activation in certain brain regions may be predictive of postimplant auditory and language performance as measured by parental observation/reports.

**BACKGROUND:** Seven hundred children were recalled for hearing screening at age 2-3 years due to a problem with their newborn hearing screen. They had all been well babies with no identified risk factors for hearing loss and hence were not scheduled for targeted follow-up to retest hearing.

**METHODS:** There were 485 children (69%) that attended the recall. The average age was 36 months (SD 3.7). Family ethnicity was Pacific Island (36%), Asian (26%), NZ European (13%), and Māori (11%), and there was a high level of deprivation in the study population. Children were screened using distortion product otoacoustic emission (DPOAE) and a parent or caregiver completed a 14-item questionnaire about ear health. The children that did not pass screening were given appointments for audiology testing. Children with hearing loss and/or middle ear problems were referred for otolaryngology review and further hearing assessments.

**RESULTS:** About one third (36%; n = 176) of children did not pass DPOAE screening; 82 (17%) had abnormal type B tympanograms and hearing loss; 29 underwent insertion of ventilation tubes, and one had a perforated tympanic membrane. There was a significant association between failed tympanometry and hearing loss (Chi-squared = 16.67, p < .001). Five children had permanent sensorineural hearing loss (SNHL), two of whom required cochlear implants for idiopathic hearing loss, with no specific risk factors. Overall 380 of 485 children screened were deemed to have normal hearing (i.e. 22% failed hearing). From the questionnaire, 15% of the caregivers with no suspicion of hearing problems did have children with significant hearing loss. Regression analysis showed that Pacific/Māori ethnicity was significantly associated with risk of hearing loss, together with questionnaire items identifying hearing problems and breathing problems.

**CONCLUSIONS:** There is a high proportion of children in South Auckland with unsuspected hearing loss; a different approach to hearing screening is warranted for this population with high rates of middle ear disease at age 3.


**BACKGROUND:** Iron is crucial for fetal brain development; however, there are insufficient data regarding the effects of maternal iron deficiency anemia (IDA) on auditory neural maturation.

**AIM:** We evaluated the effect of maternal IDA on auditory brainstem response (ABR) in full term neonates.

**METHODS:** Out of 223 pregnant women, 50 were diagnosed as having IDA and 50 healthy mothers were enrolled as controls. ABR test was done for the studied neonates within 48 hours after birth and at 3 months.

**RESULTS:** We found that hemoglobin and iron profile were lower in neonates born to anemic mothers compared with controls. Of 100 neonates screened for ABR, 25 failed the test (all of them were born to anemic mothers). The majority of neonates who failed the screening ABR test (88%) had latent iron deficiency (cord blood ferritin 11-75 µg/L). After 3 months, 85 neonates underwent diagnostic ABR test which revealed significantly prolonged interpeak latencies I-III, III-V, and I-V among neonates born to IDA mothers compared with the control group. Within the IDA group, all interpeak latencies were more prolonged in neonates with latent iron deficiency and in those born to mothers with serum ferritin < 15 µg/L. Logistic regression analysis showed that maternal hemoglobin and mean corpuscular volume could predict neonatal ABR results.

**CONCLUSIONS:** IDA during late pregnancy adversely affects cord blood iron and hearing status. ABR results are closely related to the severity of maternal and neonatal iron status. Antenatal screening of pregnant mothers is needed to improve fetal iron status and prevent abnormal auditory maturation.


Newborn screening programs aim to achieve presymptomatic diagnosis of treatable disorders allowing for early initiation of medical care to prevent or reduce significant morbidity and mortality. Many of the conditions included in the newborn screening panels are inborn errors of metabolism; however, screening for endocrine, hematologic, immunologic, and cardiovascular diseases, and hearing loss is also included in many panels. Newborn screening tests are not diagnostic and therefore diagnostic testing is needed to confirm or exclude the suspected diagnosis. Further advancement in technology is expected to allow continuous expansion of newborn screening.


**Objective:** The objectives of this study were to describe the findings of the auditory screening in children of mothers with ZIKV during pregnancy or suspicious of congenital ZIKV, and to determine whether hearing loss was in the first 2 years in life, regardless of whether microcephaly was also present.

**Methods:** This is a cases report. The information was collected and recorded in a database between January 2016 and April 2018. We perform two auditory tests to 3 and 24 months of life. The study was developed in Aguachica (Cesar, Colombia). It is considered a high-risk area for ZIKV infection. Participants included children of mothers with confirmed ZIKV during pregnancy.
or suspicious of congenital ZIKV exposure of ZIKV infection during an epidemic period in a tropical area. We defined a positive case according to the epidemiological definition and clinical criteria based on maternal symptoms. However, other children of mothers without clinical signs of Zika were evaluated at the same time. The main outcome was the presence of sensorineural hearing loss.

**Results:** The median age in the study group (n=43) was 3.5 months (rank: 0-6) and the comparison group (n: 23, children of mothers without clinical signs of ZIKV) was 3 months (rank: 0-12). Screening hearing test was done using distortion product otoacoustic emissions. At 3 months follow-up, children were evaluated using distortion product otoacoustic emissions and automated auditory brainstem response. None of the patients evaluated in this study were found to have sensorineural hearing loss.

**Conclusions:** We did not find hearing loss during the first 2 years in the children whose mother showed Zika during pregnancy. We recommend these children must be assessed to closed because there is a high risk the hearing loss as it usually may occur with CMV.

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Fasunla AJ.


**BACKGROUND:** The long-term effect of maternal HIV and antiretroviral medication on the hearing of HIV-exposed infants has not been well studied. We determined and compared the hearing thresholds of HIV-exposed infants with that of HIV-unexposed infants within the first month of life and at three, six and nine months of life.

**MATERIALS AND METHODS:** This was a Case control study of 126 HIV-exposed newborns and 121 HIV-unexposed newborns. Data collected included Socio-demographic, clinical characteristics and risk factors for hearing loss. Hearing was evaluated at newborn, 3, 6, and 9 months of life. Within and between groups analyses were done with appropriate statistics. Level of significance was P<0.05.

**RESULTS:** In both ears, the mean hearing thresholds of HIV-exposed infants were greater than those of the HIV-unexposed infants at baseline, 3, 6 and 9 months (P>0.05). In both groups, there was a decline in the mean hearing thresholds from baseline (new born) till 6 months of age. The highest mean threshold was recorded at 9 months. The mean hearing thresholds of infants at 3, 6, and 9 months were lower for HIV-exposed infants and higher for HIV-unexposed infants than the corresponding mean hearing thresholds measured at baseline. There was a significant strong correlation among hearing thresholds at 3, 6 and 9 months but weakly correlated with hearing thresholds at baseline.

**CONCLUSION:** There was a tendency towards higher hearing thresholds in HIV-exposed infants than the HIV-unexposed infants throughout the infancy period. This appears to have association with in-utero exposure to HIV.

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Findlen UM, Hounam GM, Alexy E, Adunka OF.

**Early Hearing Detection and Intervention: Timely Diagnosis, Timely Management.** *Ear Hear.*


**OBJECTIVE:** A quality improvement study was completed to assess the impact of three clinical practice changes on the timing of diagnosis and intervention for congenital hearing loss.

**DESIGN:** A retrospective chart review was conducted for 800 infants evaluated for congenital hearing loss before and after implementing three clinical practice changes: the use of Kalman-weighted signal averaging for auditory brainstem response testing, a tone burst-prioritized testing protocol, and expediting scheduling of initial assessment. The impact of middle ear involvement on age at diagnosis and history of neonatal intensive care unit stay on age at treatment was also examined.

**RESULTS:** The use of Kalman-weighted signal averaging for auditory brainstem response testing, a tone burst-prioritized testing protocol, and expediting scheduling of initial assessment each resulted in a decrease of age at diagnosis. Ultimately, the age at initial assessment was the only significant predictor related to decreased timeline for diagnosis. Middle ear pathology significantly increased age at diagnosis, while history of time in the neonatal intensive care unit significantly increased the age at provision of amplification as a treatment for permanent hearing loss.

**CONCLUSIONS:** The technology used for assessment, clinical protocol, and timing of assessment of infants can impact the timeline for diagnosis and treatment of congenital hearing impairment. Given the significant sequelae of delayed or missed diagnosis of hearing loss in infancy, implementing clinical practice changes should be considered at pediatric diagnostic centers.

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Fitzpatrick EM, Coyle D, Gaboury I, Durieux-Smith A, Whittingham J, Grandpierre V, Na E, Salamatmanesh M.

**Service Preferences of Parents of Children With Mild Bilateral or Unilateral Hearing Loss: A Conjoint Analysis Study.** *Ear Hear.*


**OBJECTIVE:** Universal newborn hearing screening results in substantially more children with mild bilateral and unilateral hearing loss identified in the early years of life. While intervention services for children with moderate loss and greater are generally well-established, considerable uncertainty and variation surrounds the need for intervention services for children with milder losses. This study was undertaken with parents of young children with permanent mild bilateral and unilateral hearing loss to examine their preferences for characteristics associated with intervention services.

**DESIGN:** Conjoint analysis, a preference-based technique, was employed to study parents’ strength of preferences. Using a cross-sectional survey that consisted of eight hypothetical clinic scenarios, we invited parents to make a discrete choice (to select one of two or more different services) between available services with different characteristics. The survey was informed by qualitative interviews conducted for this purpose. The questionnaire was administered to parents receiving intervention services in the province of Ontario, Canada, who were enrolled in a mixed-methods longitudinal study examining outcomes in early-identified children with mild bilateral/unilateral hearing loss. Data were analyzed using a generalized linear model (probit
INTRODUCTION: Hearing loss is the most common congenital disease and the prevalence of neonatal deafness can be estimated between 1 and 2 cases per 1000 live births. Infant deafness must be diagnosed as early as possible and an effective therapeutic intervention needs to be carried out in order to avoid the serious consequences of hearing deprivation during the evolutionary period: alterations in the development of central auditory pathways and lack of language acquisition. The cochlear implant (CI) has proved to be the best instrument to solve the problem of auditory deprivation. In particular, the bilateral CI gives the patient access to binaural hearing which results in benefits in terms of sound localisation and discrimination. The optimal age of application of the CI is a widely discussed topic in the scientific community and the current guidelines indicate a period between 12 and 24 months of age, even though the supporters of the application before 12 months of age are nowadays increasing.

MATERIALS AND METHODS: The study is observational, retrospective, monocentric. 49 paediatric patients (<18 years) with simultaneous bilateral CIs were included. The audiometric threshold and speech tests were carried out during the follow-up 3, 6 and 12 months after the CIs activation and when the patient reached 2 years of age.

RESULTS: The statistical analysis showed that undergoing bilateral implantation surgery before 2 years of age allows a satisfactory audiometric performance, while there are no particular benefits in performing the surgery before 1 year of age. As far as the speech outcome is concerned, the statistical analysis didn’t show significant correlation between the earlier age of implantation and better speech performance if the operation is carried out before 2.5 years of age.

CONCLUSIONS: The results of the study indicate that the optimal age to perform the simultaneous bilateral CIs surgery is between 12 and 24 months, without demonstrating any particular benefit in carrying out the procedure before 1 year of age. This may be clinically relevant in terms of avoiding the risks of diagnostic mistakes and reducing the related surgical risk in children under 1 year of age.

Gardner PA, Shanley R, Perry BP.

INTRODUCTION: In a discussion of the risks and benefits of pediatric cochlear implantation, device failure and the need for revision surgery is often overlooked. The failure rate has not been investigated extensively for this population of patients. Hearing results are under-reported following revision surgery as well. We will review our experience with cochlear implant failure, revision, and hearing results when available to better guide the preoperative counseling of families considering cochlear implantation.

METHODS: Retrospective chart review of all children undergoing cochlear implantation from 2004 to 2014.

RESULTS: In this review of 579 cases of pediatric cochlear implantation, a 4.7% device failure rate was identified. Additionally, there was a 0.3% device infection rate, as well as a 0.3% electrode extrusion rate. 10 patients had audiometric data prior to and following revision surgery. These data demonstrate similar pre-failure and post revision surgery pure tone average.

CONCLUSION: Cochlear implant device failure is the most common long-term complication of surgery; fortunately, hearing outcomes following revision surgery, evaluated with pure tone average, revealed no decline in auditory performance.
above 5 years of age. SPSS software was used for data analysis. Chi-square test was used to analyze difference between proportions.

**RESULTS:** Overall prevalence of hearing loss was 25.1%. Conductive hearing loss was present among 61 (10.3%) subjects, mixed hearing loss was found among 5 (0.8%) subjects, and sensorineural hearing loss among 94 (15.8%) subjects. On OAE, 62 (89.9%) children passed the test, and 7 (10.1%) were referred. Increasing age, female gender, and low education were significantly associated with hearing loss.

**CONCLUSION:** There was high prevalence of hearing loss in the study sample. Urgent interventions are required to identify individuals with hearing loss so that its serious complications can be reduced.

Goh LC, Azman A, Siti HBK, Khoo WV, Muthukumarasamy PA, Thong MK, Abu Bakar Z, Manuel AM. 
**Perinatol.** An audiological evaluation of syndromic and non-syndromic craniosynostosis in pre-school going children. Goh LC, Azman A, Siti HBK, Khoo WV, Muthukumarasamy PA, Thong MK, Abu Bakar Z, Manuel AM.

**Implementation of Updated Hearing Screen Guidelines in a Level IV NICU-A Quality Improvement Project.** George L, Patel JB, Park N, Manimtim WM.

**Monitoring neonates for ototoxicity.** Garinis AC, Kemph A, Tharpe AM, Weitkamp JH, McEvoy C, Steyger PS.

**Monitoring neonates for ototoxicity.** Garinis AC, Kemph A, Tharpe AM, Weitkamp JH, McEvoy C, Steyger PS.

**Monitoring neonates for ototoxicity.** Garinis AC, Kemph A, Tharpe AM, Weitkamp JH, McEvoy C, Steyger PS.

**Monitoring neonates for ototoxicity.** Garinis AC, Kemph A, Tharpe AM, Weitkamp JH, McEvoy C, Steyger PS.

**OBJECTIVES:** Neonates admitted to the neonatal intensive care unit (NICU) are at greater risk of permanent hearing loss compared to infants in well mother and baby units. Several factors have been associated with this increased prevalence of hearing loss, including congenital infections (e.g. cytomegalovirus or syphilis), ototoxic drugs (such as aminoglycoside or glycopeptide antibiotics), low birth weight, hypoxia and length of stay. The aetiology of this increased prevalence of hearing loss remains poorly understood.

**DESIGN:** Here we review current practice and discuss the feasibility of designing improved ototoxicity screening and monitoring protocols to better identify acquired, drug-induced hearing loss in NICU neonates.

**STUDY SAMPLE:** A review of published literature.

**CONCLUSIONS:** We conclude that current audiological screening or monitoring protocols for neonates are not designed to adequately detect early onset of ototoxicity. This paper offers a detailed review of evidence-based research, and offers recommendations for developing and implementing an ototoxicity monitoring protocol for young infants, before and after discharge from the hospital.

Goh LC, Azman A, Siti HBK, Khoo WV, Muthukumarasamy PA, Thong MK, Abu Bakar Z, Manuel AM.

**An audiological evaluation of syndromic and non-syndromic craniosynostosis in pre-school going children.** Goh LC, Azman A, Siti HBK, Khoo WV, Muthukumarasamy PA, Thong MK, Abu Bakar Z, Manuel AM.

**OBJECTIVE:** To study the audiological outcome and early screening of pre-school going children with craniosynostosis under follow-up at the University of Malaya Medical Center (UMMC), Kuala Lumpur, Malaysia over a 10 year period.

**METHODS:** A retrospective descriptive cohort study on the audiological findings detected during the first hearing assessment done on a child with craniosynostosis using otoacoustic emissions, pure tone audiometry or auditory brainstem response examination. The main aim of this study was to evaluate the type and severity of hearing loss when compared between syndromic and non-syndromic craniosynostosis, and other associated contributory factors.

**RESULTS:** A total of 31 patients with 62 ears consisting of 14 male patients and 17 female patients were evaluated. Twenty two patients (71%) were syndromic and 9 (29%) were non-syndromic craniosynostosis. Amongst the syndromic craniosynostosis, 9 (41%) had Apert syndrome, 7 (32%) had Crouzon syndrome, 5 (23%) had Pfeiffer syndrome and 1 (4%) had Shaethre Shutzen syndrome. Patients with syndromic craniosynostosis were more likely to present with all types and severity of hearing loss, including severe to profound sensorineural hearing loss while children with non-syndromic craniosynostosis were more likely to present with normal hearing (p < 0.05). In addition, when the first hearing test was done at a later age, a hearing loss including sensorineural hearing loss is more likely to be present in a child with syndromic craniosynostosis (p < 0.05).

**CONCLUSION:** Our study suggested that children who are born with syndromic craniosynostosis are more likely to suffer from a hearing loss, including that of a severe to profound degree compared to children with non-syndromic craniosynostosis. In addition to that, hearing loss is more likely to be detected when the first hearing test is done at a later age, and this can be an irreversible sensorineural hearing loss. We would like to advocate the need for early audiological screening and follow up in children with syndromic craniosynostosis.
Universal newborn hearing screening (UNHS), when accompanied by timely access to intervention services, can improve language outcomes for children born deaf or hard of hearing (D/HH) and result in economic benefits to society. Early Hearing Detection and Intervention (EHDI) programs promote UNHS and using information systems support access to follow-up diagnostic and early intervention services so that infants can be screened no later than 1 month of age, with those who do not pass their screen receiving diagnostic evaluation no later than 3 months of age, and those with diagnosed hearing loss receiving intervention services no later than 6 months of age. In this paper, we first document the rapid roll-out of UNHS/EHDI policies and programs at the national and state/territorial levels in the United States between 1997 and 2005. We then review cost analyses and economic arguments that were made in advancing those policies in the United States. Finally, we examine evidence on language and educational outcomes that pertain to the economic benefits of UNHS/EHDI. In conclusion, although formal cost-effectiveness analyses do not appear to have played a decisive role, informal economic assessments of costs and benefits appear to have contributed to the adoption of UNHS policies in the United States.

Hansashree YS, Bhatt SH, Nimbalkar S, Mishra G.
**Non-compliance With Neonatal Hearing Screening Follow-up in Rural Western India.** *Indian Pediatr.* 2018 Jun 15;55(6):482-484.

**OBJECTIVE:** The reasons of failure to follow-up for the Universal Neonatal Hearing Screening (UNHS) program were delineated.

**METHODS:** Review of case records for data related to follow-up of neonates who underwent the UNHS between February 2012 - January 2015.

**RESULTS:** 2534 neonates underwent primary screening with Distortion Product Oto-acoustic Emission (DPOAE). 14 (26.9%) were lost to follow-up between the first and second DPOAE screenings. 275 neonates (including high-risk cases) were to undergo confirmatory Brain Evoked Response Audiometry testing out of which 201 (73.4%) came for follow-up. Out of 74 who failed to follow-up (including those lost between first and second DPOAE screenings), unwillingness and non-compliance was the commonest reason.

**CONCLUSION:** Increasing awareness and counseling of the caretaker are important interventions for ensuring good follow-up in hearing screening programs.


Almost one third of the three million people in China suffering severe deafness are children, and 50% of these cases are believed to have genetic components to their etiology. Newborn hearing genetic screening can complement Universal Neonatal Hearing Screening for the diagnosis of congenital hearing loss as well as identifying children at risk for late-onset and progressive hearing impairment. The aim of this joint academic and Ministry of Health project was to prototype a cost-effective newborn genetic screen in a community health setting on a city-wide level, and to ascertain the prevalence of variation at loci that have been associated with non-syndromic hearing loss. With the participation of 143 local hospitals in the city of Wuhan, China we screened 142,417 neonates born between May 2014 and Dec. 2015. The variants GJB2 c.235delC, SLC26A4 c.919-2A>G, and mitochondrial variants m.1555A>G and m.1494C>T were assayed using real time PCR. Newborns found to carry a variant were re-assayed by sequencing in duplicate. Within a subset of 707 newborns we assayed using real-time PCR and ARMS-PCR to compare cost, sensitivity and operating procedure. The most frequent hearing loss associated allele detected in this population was the 235delC variant in GJB2 gene. In total, 4289 (3.01%) newborns were found to carry a variant were re-assayed by sequencing in duplicate. Within a subset of 707 newborns we assayed using real-time PCR and ARMS-PCR to compare cost, sensitivity and operating procedure. The most frequent hearing loss associated allele detected in this population was the 235delC variant in GJB2 gene. In total, 4289 (3.01%) newborns were found to carry at least one allele of either GJB2 c.235delC, SLC26A4 c.919-2A>G or two assayed MT-RNR1 variants. There was complete accordance between the real-time PCR and the ARMS PCR, though the real-time PCR had a much lower failure rate. Real-time PCR had a lower cost and operating time than ARMS PCR. Ongoing collaboration with the participating hospitals will determine the specificity and sensitivity of the association of the variants with hearing loss at birth and arising in early childhood, allowing an estimation of the benefits of newborn hearing genetic screening in a large-scale community setting.

Heffernan CB, McKeon MG, Molony S, Kawai K, Stiles DJ, Lachenauer CS, Kenna MA, Watters K.

**OBJECTIVE(S):** The objective was to describe the characteristics of hearing losses documented in patients treated with clarithromycin alone for nontuberculous mycobacterial NTM lymphadenitis in a pediatric tertiary care center over a 12-year period.

**METHODS:** An institutional review board (IRB) approval was obtained. A database search was performed using the ICD-10 diagnosis codes 31.0, 31.1, and 31.8 between January 2004 and January 2017. A REDCap database was created to record variables. Patients were included if they received clarithromycin alone and had, at the minimum, a baseline audiometry assessment, and 1 further evaluation during treatment. Fisher’s exact test was used to analyze categorical variables, and Wilcoxon rank sum test was used to analyze continuous variables.

**RESULTS:** A total of 167 patients with cervicofacial NTM were identified. Of them, 42 patients fulfilled inclusion criteria. Three children (7%) developed a hearing loss (HL) between 25 and 63 days after starting treatment. HL was unilateral in 2 children. HL persisted in 1 child following cessation of treatment. However, this patient had Rubinstein Taybi syndrome, limiting our ability to attribute the HL solely to clarithromycin.
CONCLUSION: We noted a 7% hearing loss rate in our series. Confounding issues, such as 1 patient with a syndrome potentially contributing to HL, and limitations to this study, including retrospective design and loss to follow-up, temper our ability to conclude that clarithromycin was the sole cause of these HL. However, enough supporting data for a role in clarithromycin causing HL exist that testing should be considered for patients undergoing long-term clarithromycin treatment.

Hilditch C1, Liersch B, Spurrier N, Callander EJ, Cooper C, Keir AK.
Currently, the diagnosis of congenital cytomegalovirus (cCMV) infection in most highly resourced countries is based on clinical suspicion alone. This means only a small proportion of cCMV infections are diagnosed. Identification, through either universal or targeted screening of asymptomatic newborns with cCMV, would who previously have gone undiagnosed, would allow for potential early treatment with antiviral therapy, ongoing audiological surveillance and early intervention if sensorineural hearing loss (SNHL) is identified. This paper systematically reviews published papers examining the potential benefits of targeted and universal screening for newborn infants with cCMV. We found that the treatment of these infants with antiviral therapy remains controversial, and clinical trials are currently underway to provide further answers. The potential benefit of earlier identification and intervention (eg, amplification and speech therapy) of children at risk of later-onset SNHL identified through universal screening is, however, clearer.

Huang B, Han M, Wang G, Huang S, Zeng J, Yuan Y, Dai P.
OBJECTIVES: To provide appropriate genetic testing and counseling for non-syndromic hearing impairment patients in Hainan Province, an island in the South China Sea.
METHODS: 299 unrelated students with non-syndromic hearing loss who attended a special education school in Hainan Province were enrolled in this study. Three common deafness-related genes (GJB2, SLC26A4, and mtDNA 12S rRNA) were analyzed using Sanger sequencing.
RESULTS: GJB2 mutations were detected in 32.78% (98/299) of the entire cohort; however, only 5.69% (17/299) had two confirmed pathogenic mutations. The most common mutation observed in this population was c.109G > A in the GJB2 gene, with an allelic frequency of 15.05% (90/598), which is significantly higher than that reported in previous cohorts. A total of 16 patients had two confirmed pathogenic SLC26A4 gene mutations, and 16 patients had one. The IVS7-2A > G mutation was the most commonly observed, with an allelic frequency of 3.51% (21/598). Three patients had a m.1555A > G mutation in the mtDNA 12S rRNA gene.
CONCLUSIONS: These results reveal that genetic etiology occurred in 11.71% (35/299) of patients, suggesting that Hainan province have a different mutational spectrum compared to Mainland China in non-syndromic deafness patients, which provide useful information to genetic counseling in Hainan province.

Huang Z, Gordish-Dressman H, Preciado D, Reilly BK.
OBJECTIVES/HYPOTHESIS: Our objectives were to investigate pediatric cochlear implantation (PCI) across representative states within the United States and analyze any geographical differences in age, median household income, race, insurance, and total medical charges.
STUDY DESIGN: Cross-sectional.
METHODS: Data from children (aged 0.5-18 years) who received cochlear implantation surgery were collected from the 2011 State Ambulatory Surgery and Services Databases from California (CA), Florida (FL), Maryland (MD), New York (NY), and Kentucky (KY) as a part of the Healthcare Cost and Utilization Project. We performed data analysis using a combination of Kruskal-Wallis and Wilcoxon rank sum tests, as well as nominal logistic regression.
RESULTS: Five hundred twelve cases of PCI were performed during 2011 across the five states. The overall mean and median age of implantation were 5.6 years and 4 years, respectively. There was no statistical difference in age of implantation across states (P = .85). However, there were statistical differences in primary payer (P < .001), median household income quartiles of patients who received an implant (P < .006), race (P < .001), and total median hospital charges for four of the states, with the exception of CA (P = .001).
CONCLUSIONS: Age of PCI appears to be similar across the five states in cross-sectional analysis. Geographic variations in charges, payer, race, and median household income occur with statistical significance in PCI. Further analysis of contributing factors at each state level may help elucidate the root cause of these disparities and improve and justify a uniform approach to healthcare delivery and standards of care.

Hunter LL, Blankenship CM, Gunter RG, Keefe DH, Feeney MP, Brown DK, Baroch K.
BACKGROUND: Examination of cochlear and neural potentials is necessary to assess sensory and neural status in infants, especially those cared for in neonatal intensive care units (NICU) who have high rates of hyperbilirubinemia and thus are at
Ismail AI, Abdul Majid AH, Zakaria MN, Abdullah NAC, Hamzah S4, Mukari SZS.


OBJECTIVE: The current study aims to examine the effects of human resource (measured with the perception of health workers’ perception towards UNHS), screening equipment, program layout and screening techniques on healthcare practitioners’ awareness (measured with knowledge) of universal newborn hearing screening (UNHS) in Malaysian non-public hospitals.

METHODS: Via cross sectional approach, the current study collected data using a validated questionnaire to obtain information on the awareness of UNHS program among the health practitioners and to test the formulated hypotheses. 51, representing 81% response rate, out of 63 questionnaires distributed to the health professionals were returned and usable for statistical analysis. The survey instruments involving healthcare practitioners’ awareness, human resource, program layout, screening instrument, and screening techniques instruments were adapted and scaled with 7-point Likert scale ranging from 1 (little) to 7 (many). Partial Least Squares (PLS) algorithm and bootstrapping techniques were employed to test the hypotheses of the study.

RESULTS: With the result involving beta values, t-values and p-values (i.e. β=0.478, t=1.904, p<0.10; β=0.809, t=3.921, p<0.01; β=-0.436, t=1.870, p<0.10), human resource, measured with training, functional equipment and program layout, are held to be significant predictors of enhanced knowledge of health practitioners. Likewise, program layout, human resource, screening technique and screening instrument explain 71% variance in health practitioners’ awareness. Health practitioners’ awareness is explained by program layout, human resource, and screening instrument with effect size (f2) of 0.065, 0.621, and 0.211 respectively, indicating that program layout, human resource, and screening instrument have small, large and medium effect size on health practitioners’ awareness respectively. However, screening technique has zero effect on health practitioners’ awareness, indicating the reason why T-statistics is not significant.

CONCLUSION: Having started the UNHS program in 2003, non-public hospitals have more experienced and well-trained employees dealing with the screening tools and instrument, and the program layout is well structured in the hospitals. Yet, the issue of homogeneity exists. Non-public hospitals charge for the service they render, and, in turn, they would ensure quality service, given that they are profit-driven and/or profit-making establishments, and that they would have no option other than provision of value-added and innovative services. The employees in the non-public hospitals have less screening to carry out, given the low number of babies delivered in the private hospitals. In addition, non-significant relationship between screening techniques and healthcare practitioners’ awareness of UNHS program is connected with the fact that the techniques that are practiced among public and non-public hospital are similar and standardized. Limitations and suggestions were discussed.

**Purpose:** The aims of the study were to examine the acoustic reflex screening and threshold in healthy neonates and those at risk of hearing loss and to determine the effect of birth weight and gestational age on acoustic stapedial reflex (ASR).

**Method:** We assessed 18 healthy neonates (Group I) and 16 with at least 1 risk factor for hearing loss (Group II); all of them passed the transient evoked otoacoustic emission test that assessed neonatal hearing. The test battery included an acoustic reflex screening with activators of 0.5, 1, 2, and 4 kHz and broadband noise and an acoustic reflex threshold test with all of them, except for the broadband noise activator.

**Results:** In the evaluated neonates, the main risk factors were the gestational age at birth and a low birth weight; hence, these were further analyzed. The lower the gestational age at birth and birth weight, the less likely that an acoustic reflex would be elicited by pure-tone activators. This effect was significant at the frequencies of 0.5, 1, and 2 kHz for gestational age at birth and at the frequencies of 1 and 2 kHz for birth weight. When the broadband noise stimulus was used, a response was elicited in all neonates in both groups. When the pure-tone stimulus was used, the Group II showed the highest acoustic reflex thresholds and the highest percentage of cases with an absent ASR. The ASR threshold varied from 50 to 100 dB HL in both groups. Group II presented higher mean ASR thresholds than Group I, this difference being significant at frequencies of 1, 2, and 4 kHz.

**Conclusions:** Birth weight and gestational age at birth were related to the elicitation of the acoustic reflex. Neonates with these risk factors for hearing impairment were less likely to exhibit the acoustic reflex and had higher thresholds.


**BACKGROUND:** Fetal MRI at 3 T is associated with increased acoustic noise relative to 1.5 T.

**OBJECTIVE:** The goal of this study is to determine if there is an increased prevalence of congenital hearing loss in neonates who had a 3-T prenatal MR vs. those who had it at 1.5 T.

**MATERIALS AND METHODS:** We retrospectively identified all subjects who had 3-T fetal MRI between 2012 and 2016 and also underwent universal neonatal hearing screening within 60 days of birth. Fetuses with incomplete hearing screening, magnetic resonance imaging (MRI) studies at both field strengths or fetuses affected by conditions associated with hearing loss were excluded. A random group of controls scanned at 1.5 T was identified. Five subjects had repeat same-strength MRIs (one at 3 T and four at 1.5 T). The pass/fail rate of the transient otoacoustic emissions test and auditory brainstem response test were compared using the Fisher exact test. A logistic regression was performed to assess the effects of other known risk factors for congenital hearing loss.

**RESULTS:** Three hundred forty fetal MRI examinations were performed at 3 T, of which 62 met inclusion criteria. A control population of 1.5-T fetal MRI patients was created using the same exclusion criteria, with 62 patients randomly selected from the eligible population. The fail rates of transient otoacoustic emissions test for the 1.5-T and 3-T groups were 9.7% and 6.5%, respectively, and for the auditory brainstem response test were 3.2% and 1.6%, respectively. There was no significant difference in the fail rate of either test between groups (P=0.74 for transient otoacoustic emissions test, and P=0.8 for auditory brainstem response test). The median gestational age of the 3-T group was 30 weeks, 1 day, significantly higher (P<0.001) than the 1.5-T group (median gestational age: 20 weeks, 2 days).

**CONCLUSION:** Our findings suggest that the increase in noise associated with 3 T does not increase the rate of clinically detectable hearing abnormalities.


**OBJECTIVE:** To examine maturation of the central auditory pathway, using P1 cortical auditory evoked potential (CAEP), in children who had received unilateral or bilateral cochlear implantation (CI).

**STUDY DESIGN:** Prospective study.

**SETTING:** Tertiary referral hospital.

**METHODS:** Twenty children who had received CI due to congenital, or prelingual, deafness participated in the study. Participants had received the 1st implant at a mean age of 3.4±0.7 years; 16 had also received a 2nd CI for the contralateral ear, at a mean age of 11.1±2.1 years. P1 CAEP was recorded while using the 1st implant and, for those who received contralateral CI, within 2 weeks of switching on the 2nd implant. Relations between P1 latency and duration with the 1st implant, and between age at 1st CI and P1 latency, were investigated. Relations between P1 latency with the 1st and 2nd implants, and between the interstage interval and difference between P1 latencies with the 1st and 2nd implants, were also examined.

**RESULTS:** P1 CAEP with the 1st implant was present in 16 of the 20 children. Mean P1 latency was shorter in the early CI group compared with the late CI group, but this difference was not statistically significant (p=0.154). There was a significant negative correlation between the duration with the 1st implant and P1 latency (r = -0.783, p<0.001). Among the 16 children with bilateral CI, P1 CAEP with the 2nd implant was present in 10. There was a significant negative correlation...
between the duration with the 1st implant before receiving the 2nd implant and P1 latency with the 2nd implant ($r = -0.710, p = 0.021$); there was also a significant positive correlation between P1 latency with the 1st and 2nd implants ($r = 0.722, p = 0.018$). There was not a significant correlation between interstage interval and the difference between the two P1 latencies ($r = -0.430, p = 0.248$).

**CONCLUSION:** Longer cochlear implant use is associated with shorter P1 latency. Unilateral hearing with the 1st implant may positively affect P1 latency with the 2nd CI ear. These findings imply that increased auditory experience may influence central auditory pathway maturation and that the degree of central auditory pathway maturation before the 2nd CI, rather than the timing when the surgery is received, may influence 2nd CI outcome in children with sequential bilateral cochlear implants.

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**Jones AL, Lambert AW, Barnett M.**

**Nursing students: Training and maintaining universal newborn hearing screening knowledge.** *Nurse Educ Pract.*


The purpose of the study was to obtain information concerning the effectiveness of a training program to equip nursing students in administering universal newborn hearing screening procedures, correctly recording results and clearly explaining outcomes to parent/guardians. First semester and third semester nursing students completed the National Center for Hearing Assessment and Management (NCHAM) Utah State University™ Universal Newborn Hearing Screening (UNHS) training as an in-class assignment. Students were surveyed and knowledge assessed prior to and post training. Overall results showed that all student benefited from the training.

**Kanji A1, Khoza-Shangase K1.**

**Objective Hearing Screening Measures: An Exploration of a Suitable Combination for Risk-Based Newborn Hearing Screening.** *J Am Acad Audiol.*


**BACKGROUND:** The ideal hearing screening measure is yet to be defined, with various newborn hearing screening protocols currently being recommended for different contexts. Such diverse recommendations call for further exploration and definition of feasible and context-specific protocols.

**PURPOSE:** The aim of the study was to establish which combinations of audiological screening measures provide both true-positive (TP) and true-negative (TN) results for risk-based hearing screening, at and across time.

**RESEARCH DESIGN:** A longitudinal, repeated-measures design was employed.

**STUDY SAMPLE:** Three-hundred and twenty-five participants comprised the initial study sample. These participants comprised newborns and infants who were discharged from the neonatal intensive care unit and high care wards to "step down" wards at two public sector hospitals within an academic hospital complex.

**DATA COLLECTION AND ANALYSIS:** Transient evoked otoacoustic emissions (TEOAEs), distortion product otoacoustic emissions (DPOAEs), and automated auditory brainstem response (AABR) were conducted at the initial and repeat hearing screening. Diagnostic audiological assessments were also conducted. Results from combinations of audiological screening measures at the initial and repeat hearing screening were analyzed in relation to the final diagnostic outcome ($n = 91$). Participants were classified as presenting with an overall "refer" if the outcome for any one test was "refer." The overall screening outcomes for different test combinations were compared using McNemar's test for paired data. Proportions across different test combinations were compared by the z-test for proportions.

**RESULTS:** Because of the absence of participants with hearing loss in the current study sample, analysis could only be conducted in relation to TN findings (specificity) and not TP findings (sensitivity). The percentage of TN findings was highest at the repeat hearing screening using any test or combination of tests when compared with findings from the initial hearing screening. TEOAE combined with AABR (TEOAE/AABR) ($p < 0.0001$), DPOAE combined with AABR (DPOAE/AABR) ($p < 0.0001$), and the combination of all three screening measures ($p < 0.0001$) yielded the highest percentage specificity at the repeat hearing screening when compared with the initial hearing screening.

**CONCLUSIONS:** The best specificity was noted at the repeat hearing screening. Within a resource stricken context, where availability of all screening measures options may not be feasible, current study findings suggest the use of a two-stage AABR protocol or TEOAE/AABR protocol.

**Kanji A.**

**Early hearing detection and intervention: Reflections from the South African context.** *S Afr J Commun Disord.*


For researchers and clinicians in developing contexts like South Africa, the establishment of universal newborn hearing screening (UNHS) programmes is something which we have strive to achieve. However, we need to ask the question as to whether we have attempted to view our ultimate goal of achieving mandated UNHS programmes from the perspective of the South African healthcare system as a whole. The current manuscript is aimed at providing an overview of audiological services within a broader context, with reflections from a South African perspective, and a suggestion to consider alternatives to UNHS, particularly in the South African public health care sector.

**Kanona H, Stephenson K, D’Arco F, Rajput K, Cochrane L, Jephson C.**

**Computed tomography versus magnetic resonance imaging in paediatric cochlear implant assessment: a pilot study and our experience at Great Ormond Street Hospital.** *J Laryngol Otol.*


**BACKGROUND:** To date, there is a lack of consensus regarding the use of both computed tomography and magnetic reso-
nance imaging in the pre-operative assessment of cochlear implant candidates.

**METHODS:** Twenty-five patients underwent high-resolution computed tomography and magnetic resonance imaging. ‘Control scores’ describing the expected visualisation of specific features by computed tomography and magnetic resonance imaging were established. An independent radiological review of all computed tomography and magnetic resonance imaging scan features was then compared to the control scores and the findings recorded.

**RESULTS:** Agreement with control scores occurred in 83 per cent (20 out of 24) of computed tomography scans and 91 per cent (21 out of 23) of magnetic resonance imaging scans. Radiological abnormalities were demonstrated in 16 per cent of brain scans and 18 per cent of temporal bone investigations.

Kaspar A, Newton O, Kei J, Driscoll C, Swanepoel W, Goulios H.


**OBJECTIVES:** The present study aimed to assess the prevalence of otitis media and risk-factors for sensorineural hearing loss among infants in the Solomon Islands, in order to recommend an Infant Ear and Hearing Program that would be suitable to the Pacific Island context.

**METHODS:** Ear examinations and the JCIH Risk-Factor Questionnaire were administered to 288 infants attending Child Welfare Clinics in the Solomon Islands.

**RESULTS:** Overall, 150 infants (52.08%) presented with bilateral normal ear examinations and no risk-factors for SNHL. There were 73 infants (25.34%) with ear pathology in at least one ear, 13 (4.5%) of whom required referral to the ENT Clinic for medical management. The most common pathology was otitis media with effusion (OME) (21.87%). Infants aged 7-12 months were significantly more likely to present with OME (p<0.001) and a history of otitis media (p<0.017) than infants aged 0-6 months. There were 71 infants (24.65%) with at least one risk-factor for sensorineural hearing loss. The most common risk-factors were ototoxicity (8.3%), non-elective caesarean delivery (6.59%), and possible in-utero syphilis infection (5.55%).

**CONCLUSIONS:** The prevalence of otitis media and risk-factors for sensorineural hearing loss indicate the importance of initiating Infant Ear and Hearing Programs in the Solomon Islands. Program should facilitate early education on prevention of ear disease, as well as early diagnosis and management of children with hearing loss.

**CONCLUSION:** Assessment in the paediatric setting constitutes a special situation given the likelihood of congenital temporal bone abnormalities and associated co-morbidities that may be relevant to surgery and prognosis following cochlear implantation. Both computed tomography and magnetic resonance imaging contribute valuable information and remain necessary in paediatric cochlear implant pre-operative assessment.


**Risk Factors for Hearing Loss and Its Prevalence in Neonates Older than 6 Months with History of Hospitalization in Intensive Care Unit.** Iran J Child Neurol.


**Objectives:** Hearing loss is one of the most important disabilities in neonates. Delay in the detection of hearing loss leads to impaired development and may prevent the acquisition of speech. We aimed to determine the risk factors associated with hearing loss in neonatal patients aged more than 6 months with a history of hospitalization in Neonatal Intensive Care Unit (NICU).

**Methods:** In this case-control study, screening for hearing loss was carried out on 325 neonates aged 6-12 months referred to Pediatric Neurology Office of Vali-e-Asr Hospital, Tehran, Iran up to 2011. Hearing loss was confirmed using Auditory Brainstem Response screening test (ABR).

**Results:** The prevalence of mildly and moderately hearing loss in neonates was determined as 3.6%. The most significant risk factors for hearing loss in neonates were neonatal icterus associated with phototherapy, respiratory distress syndrome (RDS) and lower Apgar score.

**Conclusion:** It seems to quantitative auditory system screening using ABR is necessary for all neonates; because rehabilitation support such as speech therapy and hearing training in this age period is more effective than older ages.

Khairy MA, Abuelhamed WA, Ahmed RS, El Fouly HES, Elhawary IM.

**Hearing loss among high-risk newborns admitted to a tertiary Neonatal Intensive Care Unit.** J Matern Fetal Neonatal Med.


**PURPOSE:** The aim of this work is to identify the most significant risk factors for hearing impairment in high risk neonates hospitalized at our Neonatal Intensive Care Unit (NICU) and to assess the sensitivity of hearing screening tests.

**METHODS:** This study involved 260 neonates admitted to a tertiary NICU; they were classified into two groups; 150 preterm and 110 full term neonates. The hearing screening tests performed were transient evoked otoacoustic emissions (TEOAEs) and the automated auditory brainstem response (AABR).

**RESULTS:** Forty-eight preterm neonates (32%) and 30 full term neonates (27.3%) had pathological AABR. In preterm group, mechanical ventilation more than five days, sepsis, usage of aminoglycosides, loop diuretics, vancomycin alone or in combination with aminoglycosides and prolonged duration of admission were considered risk factors of hearing affection whereas in full term group mechanical ventilation more than five days was the risk factor of hearing affection (p<0.05).

**CONCLUSIONS:** The prevalence of hearing loss is highest among high risk neonates and TEOAE and AABR were found to be reliable screening tools. Use of ototoxic drugs and mechanical ventilation for more than five days were significant risk factors for hearing loss in our study population.

BACKGROUND: In South Africa, primary health care is the first point of contact with the health system for at least 85% of the population, yet early hearing detection and intervention continues to be elusive in these settings. Nurses at community level may, therefore, be missing an opportunity to identify prelingual infants with hearing losses and alter their developmental trajectory.

AIM: To determine primary health care nurses’ experiences, practices and beliefs regarding hearing loss in infants.

SETTING: The study was conducted in the eThekwini District of KwaZulu-Natal, South Africa.

METHODS: A descriptive survey was used with quantitative methods of analysis. Fourteen primary health care clinics from the eThekwini district were selected, from which 75 nurses participated by completing a self-administered questionnaire.

RESULTS: At least one-third of primary health care nurses had never screened a child for hearing loss, and most clinics did not have access to basic hearing screening equipment or materials. Only 49% of nurses had access to an otoscope, while 31% used the Road to Health Development screener to check for hearing loss. None of the clinics had access to an otowacoum emission screener or the Swart questionnaire. Although nurses reported that they would refer to audiology services for some of the risk factors, as indicated on the Joint Committee on Infant Hearing (JCIH) 2007 list, they were less likely to refer if the child was in a neonatal intensive care unit (ICU) longer than five days, had neurodegenerative disorders, meningitis, hyperbilirubinaemia requiring blood transfusion or were undergoing chemotherapy. Less than a third of nurses always referred if the child displayed additional non-JCIH risk factors or those pertinent to the South African context. Approximately 38% reported that communities believed that hearing loss could be because of some form of spiritual or supernatural causes.

CONCLUSION: This study demonstrates that hearing screening and referral practices at primary health care clinics need to be strengthened. Nurses need to be capacitated to conduct basic screening, make necessary referrals, provide information to caregivers and understand community beliefs about hearing loss in order to counsel caregivers appropriately and facilitate the process of early hearing detection and intervention.


Congenital cytomegalovirus (cCMV) infection is a common congenital infection that causes sensorineural hearing loss (SNHL). Despite its substantial impact on public health and cost burden, epidemiology and clinical features of CMV-related SNHL have never been reported in the Korean populations. This study investigated the detailed audiologic phenotypes of cCMV infection to see if a specific SNHL pattern is associated with a particular clinical setting. A total of 38 patients with cCMV infection were studied retrospectively. Patients were classified into three groups with distinct demographics: clinically driven diagnosis (n=17), routine newborn CMV screening according to the NICU protocols (n=10), or referral to ENT for cochlear implant (CI) (n=11). The incidence of cCMV infection was 3.6%, showing 33.3% of SNHL among cCMV patients, 38% of asymmetric hearing loss, 29% of late-onset hearing loss, and diverse severity spectrum in patients with CMV-related SNHL. CI recipients with CMV-related SNHL showed a significantly improved speech perception. Surprisingly, in 36.4% of CI implantees, initial audiological manifestation was significant asymmetry of hearing thresholds between both ears, with better ear retaining significant residual hearing up to 50dB. CMV turns out to be a significant etiology of SNHL, first to date reported in the Korean pediatric population. Analysis of audiologic phenotypes showed a very wide spectrum of SNHL and favorable CI outcomes in case of profound deafness. Especially for the patients with asymmetric hearing loss, close surveillance of hearing should be warranted and CI could be considered on the worse side first, based on the observation of rapid progression to profound deafness of better side.


BACKGROUND: Congenital cytomegalovirus (cCMV) infections are the leading nongenetic cause of congenital sensorineural hearing loss (SNHL); however the true impact of cCMV infections remains unknown.

AIMS OF THE STUDY: (1) To identify the number of asymptomatic and symptomatic cCMV infections diagnosed between 1999 and 2014 at the Lausanne University Hospital; (2) to describe the audiological and neurodevelopmental outcomes of infants with cCMV infection; and (3) to compare clinical outcomes between infants born to mothers with primary versus nonprimary infection.

METHODS: This was a single-centre, observational, exploratory, retrospective study of newborns diagnosed with cCMV infection at the Lausanne University Hospital between 1999 and 2014.

RESULTS: Fifty newborns with cCMV infection were identified, 39 (78%) were symptomatic at birth, of whom 29 (74%) were neurologically symptomatic. Twelve children (24%) presented with subsequent abnormal audiological and/or neurodevelopmental outcomes. Newborns born to mothers with a nonprimary infection were more often symptomatic at birth than those born to mothers with a primary infection.

CONCLUSIONS: All infants with subsequent SNHL or abnormal neurodevelopment were symptomatic at birth. Similar long-term neurodevelopmental and audiological outcomes were observed in infants born to mothers with a primary and nonprimary infection.
An average number of $10 \times 2$ threshold measurements were obtained during ABR testing with age-appropriate sedation, thus allowing for the evaluation of the degree, type and configuration of the hearing loss.

### Geographic and Racial Disparities in Infant Hearing Loss


**Objective**

Approximately 1 to 2 of every 1000 American newborns have hearing loss identified by newborn screening. This study was designed to determine if infant hearing loss is more common in socioeconomically disadvantaged communities.

**Study Design**

In this retrospective study, we analyzed electronic medical record data using geostatistical models. Setting

Infants were residents of Durham County, North Carolina, born in 2 hospitals of the Duke University Health System. This county includes the city of Durham and surrounding suburban and rural communities. Subjects and Methods

Subjects were hearing-screened newborns, born between 2005 and 2016, whose residential address was in Durham County, North Carolina. This was a retrospective study using medical record data. We used Bayesian regression models with smoothing of coordinate date to identify both spatial and nonspatial predictors of infant hearing loss. Results

We identified 19,348 infants from Durham County, of whom 675 had failed initial hearing screening and 191 had hearing loss confirmed on follow-up. Hearing loss was significantly associated with minority race (odds ratio [OR], 2.45; 95% confidence interval, 1.97-3.06), as well as lower gestational age and maternal sexually transmitted infections. We identified significant geographic heterogeneity, with a higher probability of hearing loss in poorer urban neighborhoods (local OR range, 0.59-1.39). Neighborhood disadvantage was a significant predictor of hearing loss, as was high local seroprevalence of cytomegalovirus (CMV) among pregnant women. Conclusions

Urban, low-income neighborhoods have a high prevalence of infant hearing loss compared with more affluent surrounding communities, particularly among minorities. This distribution may be attributable to congenital CMV infection.

### Frequency-specific auditory brainstem response testing with age-appropriate sedation

**Levit Y, Mandel D, Matot I.**


**Objective:** Auditory brainstem response (ABR) testing is the gold-standard procedure for hearing evaluation in pediatric patients who cannot complete a behavioral hearing test. The amount of audiological information obtained depends on the quality of the patient’s sleep during the test. In this retrospective database review, we aimed to assess the amount and the characteristics of the audiological information obtained in ABR testing in pediatric patients with age-appropriate sedation.

**Methods:** A retrospective chart review was conducted on 501 consecutive ABR sedation sessions performed between January 2014 and June 2016 at the Tel Aviv Medical Center. Oral triclofos was used for the sedation of younger patients (<24 months) and intravenous propofol for older patients (>24 months). The dataset included 370 triclofos sessions (in 337 patients) and 131 propofol sessions (in 126 patients).

**Results:** None of the children developed complications, and all were discharged on the same day of the evaluation. Among the hearing-impaired children, a mean of 10 (1.8 SD) ABR threshold measurements was obtained from propofol-sedated patients and 9.4 (2.8 SD) measurements from those sedated with triclofos (P = 0.039). The major characteristics of the hearing loss, including its degree, type, and configuration, were obtained from all propofol-sedated patients and from 95% of those sedated with triclofos.

**Conclusions:** A comprehensive evaluation of hearing status can be obtained in ABR testing with age-appropriate sedation. An average number of $\sim 10$ threshold measurements were obtained during ABR testing with age-appropriate sedation, thus allowing for the evaluation of the degree, type and configuration of the hearing loss.

OBJECTIVE: To evaluate the feasibility and potential benefits of incorporating genetic and cytomegalovirus (CMV) screenings into the current newborn hearing screening (NHS) programs.

STUDY DESIGN: Newborns were recruited prospectively from a tertiary hospital and a maternity clinic between May 2016 and December 2016 and were subjected to hearing screening, CMV screening, and genetic screening for 4 common mutations in deafness genes (p.V37I and c.235delC of GJB2 gene, c.919-2A>G of SLC26A4 gene, and the mitochondrial m.1555A>G). Infants with homozygous nuclear mutations or homoplasmic/heteroplasmic mitochondrial mutation (referred to as “conclusively positive genotypes”) and those who tested positive for CMV received diagnostic audiologic evaluations.

RESULTS: Of the total 1716 newborns enrolled, we identified 20 (1.2%) newborns with conclusively positive genotypes on genetic screening, comprising 15 newborns (0.9%) with GJB2 p.V37I/p.V37I and 5 newborns (0.3%) with m.1555A>G. Three (0.2%) newborns tested positive on CMV screening. Twelve of the 20 newborns (60%) with conclusively positive genotypes and all 3 newborns who tested positive for CMV (100%) passed NHS at birth. Diagnostic audiologic evaluations conducted at 3 months confirmed hearing impairment in 6 of the 20 infants (30%) with conclusively positive genotypes.

CONCLUSIONS: This study confirms the feasibility of performing hearing, genetic, and CMV screenings concurrently in newborns and provides evidence that the incorporation of these screening tests could potentially identify an additional subgroup of infants with impaired hearing that might not be detected by the NHS programs.


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INTRODUCTION: Some studies have demonstrated a parallelism between the extent of hearing loss and the frequency of vestibular dysfunction in children with sensorineural hearing loss (SNHL). Despite this, little is known about the repercussion of degrees of hearing loss and etiological factors on the balance performance in this children.

OBJECTIVE: Compare the balance performance between normal hearing (NH) children and those with SNHL, considering the sex and age range of the sample, and analyze balance performance according to the degrees of hearing loss and etiological factors in the latter group.

METHODS: Cross-sectional study that assessed 96 children (48 NH and 48 with SNHL), aged between 7 and 18 years old. The balance performance was assessed by the Brazilian version of the Pediatric Balance Scale, validated for Brazilian child population and the Mann-Whitney test used for statistical analysis.

RESULTS: The group with SNHL showed lower average balance performance compared to NH (p = 0.000). This was also observed when the children were grouped by sex: female and male (p = 0.001). The same difference occurred when the children were stratified by age group: 7-14 years old (p = 0.000). There were no differences between the balance performance of the groups according to the degrees of hearing loss (p = 0.236) and the children with prematurity or post-natal meningitis as an etiological factor demonstrated the worst balance performance.

CONCLUSION: The children with SNHL showed worse balance performance compared to NH of the same sex and age range between seven to fourteen years. There were no differences between balance performance and hearing loss degrees, and those children with prematurity or post-natal meningitis as an etiological factor demonstrated the worst balance performances.

Musacchia G, Ortiz-Mantilla S, Roesler CP, Rajendran S, Morgan-Byrne J, Benasich AA.

OBJECTIVE: Background noise makes hearing speech difficult for people of all ages. This difficulty can be exacerbated by co-occurring developmental deficits that often emerge in childhood. Sentence-type speech-in-noise (SIN) tests are available clinically but cannot be administered to very young individuals. Our objective was to examine the use of an electrophysiological test of SIN, suitable for infants, to track developmental trajectories.

METHODS: Speech-evoked brainstem potentials were recorded from 30 typically-developing infants in quiet and +10 dB SNR background noise. Infants were divided into two age groups (7-12 and 18-24 months) and examined across development. Spectral power of the frequency following response (FFR) was computed using a fast Fourier Transform. Cross-correlations between quiet and noise responses were computed to measure encoding resistance to noise.

RESULTS: Older infants had more robust FFR encoding in noise and had higher quiet-noise correlations than their younger counterparts. No group differences were observed in the quiet condition.

CONCLUSIONS: By two years of age, infants show less vulnerability to the disruptive effects of background noise, compared to infants under 12 months.

SIGNIFICANCE: Speech-in-noise electrophysiology can be easily recorded across infancy and provides unique insights into developmental differences that tests conducted in quiet may miss.

Netten AP, Rieffe C, Ketelaar L, Soede W, Gadow KD, Frijns JHM.
Terrible Twos or Early Signs of Psychopathology? Developmental Patterns in Early Identified Preschoolers With Cochlear Implants Compared With Hearing Controls. Ear Hear.

OBJECTIVE: Cochlear implants (CIs) have dramatically improved the lives of children who are deaf or hard of hearing; however, little is known about its implications for preventing the development of psychiatric symptoms in this at-risk population. This is the first longitudinal study to examine the early manifestation of emotional and behavioral disorders and associated risk and protective factors in early identified preschoolers with CIs compared with hearing peers.

DESIGN: Participants were 74 children with CIs and 190 hearing controls between ages 1 and 5 years (mean age, 3.8 years). Hearing loss was detected using the Newborn Hearing Screening in The Netherlands and Flanders. Parents completed the Early Childhood Inventory-4, a well-validated measure, to evaluate the symptoms of DSM-IV-defined psychiatric disorders, during three consecutive years. Language scores were derived from each child's medical notes.

RESULTS: Children with CIs and hearing controls evidenced comparable levels of disruptive behavior and anxiety/depression (which increased with age in both groups). Greater proficiency in language skills was associated with lower levels of psychopathology. Early CI and longer duration of CI use resulted in better language development. In turn, higher early language skills served as a protective factor against the development of disruptive behavior symptoms.

CONCLUSIONS: This longitudinal study uniquely shows that improvement in language skills mitigates the development of early signs of psychopathology. Early identification of hearing loss and CIs help children improve their language skills.
Improving universal newborn hearing screening outcomes by conducting it with thyroid screening. Int J Pediatr Otorhinolaryngol.

Rahimi V, Mohammadkhani G, Javadi F.

Objective: One of the most important factors that can improve hearing screening indicators is testing infants after 48 h of birth. The neonatal thyroid screening program is done during the third to fifth day after birth in many countries. So this screening is done at the appropriate time for hearing screening. The aim of the present study was to evaluate hearing screen-
ing outcomes (the referral rate, false positive rate, and positive predictive value) conducted with the thyroid screening at the healthcare centers and compare the results with hospital before discharge the infant.

**METHODS:**
This was a prospective exploratory cohort study. The study population included all the newborns at a hospital (group 1) and newborns who were referred to healthcare centers for thyroid screening (group 2), except for infants with risk factors, from March 2012 to December 2017. Transient evoked otoacoustic emissions (TEOAE) and automatic auditory brainstem response (AABR) were used for the evaluation. The results were compared between the two groups.

**RESULTS:** Of the 4729 newborns, who participated in the study, 3001 were referred from a hospital (group 1) and 1728 from two healthcare centers (group 2). The referral rate in group 1 and 2 was 16.1% and 7.6%, respectively. Also, the false positive rate in group 1 and 2 was 15.9% and 7.6%, respectively. Our study showed that the referral rate and false positive rate of hearing screening in group 2 were significantly lower than that in group 1 (p < 0.001). The positive predictive value in group 1 was significantly higher than that in group 2 (p < 0.05). There was no significant sex difference in any of the variables.

**CONCLUSIONS:** Our results showed that performing the hearing screening during the thyroid program, instead of the hospital could be significantly improved screening outcomes and suggest that hearing and thyroid screening together after discharge from the hospital could be a good opportunity to introduce new framework for hearing screening in many countries.


**PURPOSE:** To assess the potential association between psychological risk and limited auditory pathway maturation.

**METHODS:** In this longitudinal cohort study, 54 infants (31 non-risk and 23 at-risk) were assessed from age 1 to 12 months. All had normal hearing and underwent assessment of auditory maturation through cortical auditory evoked potentials testing. Psychological risk was assessed with the Child Development Risk Indicators (CDRIs) and PREAUT signs. A variety of statistical methods were used for analysis of results.

**RESULTS:** Analysis of P1 and N1 latencies showed that responses were similar in the both groups. Statistically significant differences between-groups were observed only for the variables N1 latency and amplitude at 1 month. Significant maturation occurred in both groups (p<0.05). There was moderate correlation between P1 latency and Phase II CDRIs, which demonstrates that children with longer latencies at age 12 months were more likely to exhibit absence of these indicators in Phase II and, therefore, were at greater psychological risk. The Phase II CDRIs also correlated moderately with P1 and N1 latencies at 6 months and N1 latencies at 1 month; again, children with longer latency were at increased risk.

**CONCLUSION:** Less auditory pathway maturation correlated with presence of psychological risk. Problems in the mother-infant relationship during the first 6 months of life are detrimental not only to cognitive development, but also to hearing. A fragile relationship may reflect decreased auditory and linguistic stimulation.


**INTRODUCTION:** Zika virus infection during pregnancy causes serious birth defects and might be associated with neurodevelopmental abnormalities in children. Early identification of and intervention for neurodevelopmental problems can improve cognitive, social, and behavioral functioning.

**METHODS:** Pregnancies with laboratory evidence of confirmed or possible Zika virus infection and infants resulting from these pregnancies are included in the U.S. Zika Pregnancy and Infant Registry (USZPIR) and followed through active surveillance methods. This report includes data on children aged ≥1 year born in U.S. territories and freely associated states. Receipt of reported follow-up care was assessed, and data were reviewed to identify Zika-associated birth defects and neurodevelopmental abnormalities possibly associated with congenital Zika virus infection.

**RESULTS:** Among 1,450 children of mothers with laboratory evidence of confirmed or possible Zika virus infection during pregnancy and with reported follow-up care, 76% had developmental screening or evaluation, 60% had postnatal neuroimaging, 48% had automated auditory brainstem response-based hearing screen or evaluation, and 36% had an ophthalmologic evaluation. Among evaluated children, 6% had at least one Zika-associated birth defect identified, 9% had at least one neurodevelopmental abnormality possibly associated with congenital Zika virus infection identified, and 1% had both. Our results showed that responses were similar in the both groups. Statistically significant differences between-groups were observed only for the variables N1 latency and amplitude at 1 month. Significant maturation occurred in both groups (p<0.05). There was moderate correlation between P1 latency and Phase II CDRIs, which demonstrates that children with longer latencies at age 12 months were more likely to exhibit absence of these indicators in Phase II and, therefore, were at greater psychological risk. The Phase II CDRIs also correlated moderately with P1 and N1 latencies at 6 months and N1 latencies at 1 month; again, children with longer latency were at increased risk.

**CONCLUSION:** Less auditory pathway maturation correlated with presence of psychological risk. Problems in the mother-infant relationship during the first 6 months of life are detrimental not only to cognitive development, but also to hearing. A fragile relationship may reflect decreased auditory and linguistic stimulation.
**da Silva LS, Ribeiro GE, Montovani JC, Silva DPCD.**


**OBJECTIVE:** To verify the effect of peri-intraventricular hemorrhage on the auditory pathway of preterm infants.

**METHOD:** It is a non-concurrent cohort study. This study was conducted in a tertiary public. Preterm infants with peri-intraventricular hemorrhage comprised the study group, and preterm infants without peri-intraventricular hemorrhage were included as a comparison group, both were similar in relation to gestational age and risk indicators for hearing loss. Participants had to meet the following inclusion criteria: have been born at the study site, presence of otoacoustic emissions by transient stimulus in both ears and brainstem auditory evoked potentials with all components bilaterally identified.

**RESULTS:** 44 infants with an average age of 3 months with peri-intraventricular hemorrhage and 2,6 months without peri-intraventricular hemorrhage met the inclusion criteria. Regarding the brainstem auditory evoked potentials results, a significant increase was observed in absolute latency values of waves I, III and V, as well as in the interpeak intervals I-III and I-V, bilaterally, in infants with peri-intraventricular hemorrhage.

**CONCLUSION:** This study concluded that infants with peri-intraventricular hemorrhage presented a delay in the neural conduction of sound, which justifies the monitoring of the auditory function in these infants during the period of language development.
Sunwoo W, Lee WW, Choi BY.  
**Extremely common radiographic finding of cochlear nerve deficiency among infants with prelingual single-sided deafness and its clinical implications.** Int J Pediatr Otorhinolaryngol.  
**OBJECTIVES:** To clarify the common radiographic findings of audiologically documented prelingual single-sided deafness (SSD) and identify the prevalence of cochlear nerve deficiency (CND) in SSD infants referred from the newborn hearing screening program.  
**METHODS:** Between March 2012 and March 2017, the records of all infants referred to our otology clinic after undergoing newborn hearing screening program were retrospectively reviewed. Twenty-four consecutive well infants without risk factors who had a confirmed diagnosis of prelingual SSD under the age of 1 year and who underwent internal auditory canal (IAC) magnetic resonance imaging (MRI) were included. The sizes of cochlear nerve (CN), IAC, and cochlear nerve canal (CNC) were measured on MRI. The presence of CND was visually determined by comparing the CN size to the ipsilateral facial nerve (FN) in the affected side via an oblique sagittal view of IAC MRI and defined when CN was absent or smaller than FN.  
**RESULTS:** CND was seen in all 24 deaf ears (100%) on MRI. There was one with incomplete partition type I, and another with combined cochleovestibular nerve absence. Twenty-four subjects demonstrated either an absent (20/24, 83.3%) or small (4/24, 16.7%) CN. When the absent and small CN groups were compared, the former group had a higher prevalence of narrow CNC and narrow IAC. Of the 20 infants without identifiable CN on the affected side, 17 (85%) had narrow IAC and 17 (85%) had narrow CNC. In the 20 ears with absent CN, only one had both normal-sized IAC and CNC.  
**CONCLUSION:** The contribution of CND to prelingual SSD in Korean infants reached 100%, according to IAC MRI alone.  

Suskind DL, Leung CYY, Webber RJ, Hundertmark AC, Leffel KR, Fuenmayor Rivas IE1, Grobman WA.  
**Educating Parents About Infant Language Development: A Randomized Controlled Trial.**  
A total of 427 women (aged 18-45 years) who delivered a singleton neonate without serious medical complications were randomized to watch either an educational intervention (n = 225) or the sudden infant death syndrome (n = 202) video. Linear mixed models showed that the intervention women significantly gained knowledge over time. Knowledge gain was largest among high-socioeconomic status (high-SES) and middle-SES English-speaking, smaller among low-SES Spanish-speaking, and nonsignificant among low-SES English-speaking women. Analysis of deviance revealed that the intervention women of all SES learned strategies fostering secure attachment and language acquisition. Participants considered watching an educational video alongside the universal newborn hearing screening (UNHS) conveniently timed. The intervention women were more likely than the control women to recognize the importance of timely UNHS follow-up.  

**Cochlear volume as a predictive factor for residual-hearing preservation after conventional cochlear implantation.** Acta Otolaryngol.  
**OBJECTIVE:** The preservation of residual hearing after conventional cochlear implantation (CI) is frequently observed when atrumatic soft surgery is adopted. The purpose of this study was to elucidate the predictive factors for residual hearing preservation after atrumatic CI.  
**PATIENTS:** This study included 46 patients who underwent CI based on an atrumatic technique using a standard-length flexible electrode implant through a round window approach.  
**MAIN OUTCOME MEASURE:** Cochlear volume was measured using magnetic resonance imaging (MRI). Cochlear duct length (CDL) was taken as the length of the scala media measured using computed tomography (CT). The association between residual hearing preservation and cochlear volume/CDL was then examined.  
**RESULT:** Cochlear volume and CDL were significantly larger in patients with complete hearing preservation than in those with hearing loss. Multivariate logistic regression analysis revealed that cochlear volume was a significant predictive factor for residual hearing preservation.  
**CONCLUSION:** Residual hearing preservation after conventional CI was observed in patients with a larger cochlear volume and longer CDL. Cochlear volume could be a predictive factor for residual hearing preservation after conventional CI.  

Thomson V, Yoshinaga-Itano C.  
**The Role of Audiologists in Assuring Follow-Up to Outpatient Screening in Early Hearing Detection and Intervention Systems.** Am J Audiol.  
**Purpose:** The purpose of this study was to investigate the role of audiology involvement and other factors associated with failure to follow through from the initial hearing screening to the second outpatient screen.  
**Method:** Linear regression, logistical regression, and descriptive analyses were used across demographic and hospital variables associated with infants who did not receive a follow-up outpatient screen.  
**Results:** The results included birthing hospital outpatient rescreen rates from January 1, 2005, through December 31, 2005. Variables were collected from the birth certificate and hospital surveys. Results showed higher loss to follow-up/documentation to outpatient screen for (a) infants born in hospitals with low rates for returning for follow-up, (b) infants born in hospitals that did not have an audiologist involved, (c) infants who were Hispanic, (d) infants who were born to mothers who were not married, (e) infants with mother’s with < 12 years of education, and (f) infants with Apgar scores of 7 or below.
**Conclusions:** The findings were used to identify quality improvement strategies to decrease the loss to follow-up. Strategies included ensuring audiology support, providing information in the parent’s native language, educating personnel in the newborn intensive care units, developing and disseminating information in Spanish in written form, and educating hospitals on the importance of scheduling the outpatient rescreening before hospital discharge.

**Tokat T, Catli T, Bayrak F, Bozkurt EB, Olgun L.**


**PURPOSE:** The aim of this study is to evaluate long-term outcomes of cochlear implantation (CI) in patients with postmeningitic deafness.

**METHODS:** Twenty-seven patients with severe to profound hearing loss due to bacterial meningitis and received CI were the subjects of this study. Surgical findings and long-term audiological performances were evaluated. Speech perception and speech intelligibility of the implanted patients were evaluated with the categories of auditory performance-II (CAP-II) test and speech intelligibility rating (SIR) test, respectively.

**RESULTS:** Eighteen of the 27 patients had received full electrode insertion through the patent cochlear lumen. Remaining 9 patients had varying degrees of ossification throughout the cochlea and needed to be drilled to achieve partial electrode insertion. None of the patients exhibited surgical complication. Scores in both test batteries (CAP-II and SIR) were comparable between patients who received full or partial electrode insertion (P>0.05).

**CONCLUSION:** Cochlear implantation after postmeningitic deafness has favorable outcomes especially in long term. Although this type of inner ear pathology may require special considerations during surgery, it is a relatively safe procedure.

**Turchetta R, Conti G, Marsella P, Orlando MP, Picciotti PM, Fresza S, Russo FY, Scorpecci A, Cammeresi MG, Gianantonio S, Greco A, Ralli M.**


**BACKGROUND:** The introduction of Universal Newborn Hearing Screening (UNHS) programs has drastically contributed to the early diagnosis of hearing loss in children, allowing prompt intervention with significant results on speech and language development in affected children. UNHS in the Lazio region has been initially deliberated in 2012; however, the program has been performed on a universal basis only from 2015. The aim of this retrospective study is to present and discuss the preliminary results of the UNHS program in the Lazio region for the year 2016, highlighting the strengths and weaknesses of the program.

**METHODS:** Data from screening facilities in the Lazio region for year 2016 were retrospectively analyzed. Data for Level I centers were supplied by the Lazio regional offices; data for Level II and III centers were provided by units that participated to the study.

**RESULTS:** During 2016, a total of 44,805 babies were born in the Lazio region. First stage screening was performed on 41,821 children in 37 different birth centers, with a coverage rate of 93.3%. Of these, 38,977 (93.2%) obtained a “pass” response; children with a “refer” result in at least one ear were 2844 (6.8%). Data from Level II facilities are incomplete due to missing reporting, one of the key issues in Lazio UNHS. Third stage evaluation was performed on 365 children in the three level III centers of the region, allowing identification of 70 children with unilateral (40%) or bilateral (60%) hearing loss, with a prevalence of 1.6/1000.

**CONCLUSIONS:** The analysis of 2016 UNHS in the Lazio region allowed identification of several strengths and weaknesses of the initial phase of the program. The strengths include a correct spread and monitoring of UNHS among Level I facilities, with an adequate coverage rate, and the proper execution of audiological monitoring and diagnosis among Level II facilities. Weakness, instead, mainly consisted in lack of an efficient and automated central process for collecting, monitoring and reporting of data and information.

**Uddén F, Filipe M, Reimer Å, Paul M, Matuschek E, Thegerström J, Hammerschmidt S, Pelkonen T, Riesbeck K.**


**BACKGROUND:** Chronic suppurative otitis media (CSOM) is an important cause of hearing loss in children and constitutes a serious health problem globally with a strong association to resource-limited living conditions. Topical antibiotics combined with aural toilet is the first-hand treatment for CSOM but antimicrobial resistance and limited availability to antibiotics are obstacles in some areas. The goal of this study was to define aerobic pathogens associated with CSOM in Angola with the overall aim to provide a background for local treatment recommendations.

**METHODS:** Samples from ear discharge and the nasopharynx were collected and cultured from 152 patients with ear discharge and perforation of the tympanic membrane. Identification of bacterial species was performed with matrix-assisted laser desorption/ionization-time of flight mass spectrometry and pneumococci were serotyped using multiplex polymerase chain reactions. Antimicrobial susceptibility testing was done according to EUCAST.

**RESULTS:** One hundred eighty-four samples from ear discharge and 151 nasopharyngeal swabs were collected and yielded 534 and 289 individual isolates, respectively. In all patients, correspondence rate of isolates from 2 ears in patients with bilateral disease was 27.3% and 9.3% comparing isolates from the nasopharynx and ear discharge, respectively. Proteus spp. (14.7%), Pseudomonas aeruginosa (13.2%) and Enterococcus spp. (8.8%) were dominating pathogens isolated from ear discharge. A large part of the remaining species belonged to Enterobacteriaceae (23.5%). Pneumococci and Staphylococcus aureus were detected in approximately 10% of nasopharyngeal samples. Resistance rates to quinolones exceeded 10% among Enterobacteriaceae and was 30.8% in S. aureus, whereas 6.3% of P. aeruginosa were resistant.
CONCLUSIONS: The infection of the middle ear in CSOM is highly polymicrobial, and isolates found in nasopharynx do not correspond well with those found in ear discharge. Pathogens associated with CSOM in Angola are dominated by gram-negative including Enterobacteriaceae and P. aeruginosa, while gram-positive enterococci also are common. Based on the results of antimicrobial susceptibility testing topical quinolones would be the preferred antibiotic therapy of CSOM in Angola. Topical antiseptics such as aluminum acetate, acetic acid or boric acid, however, may be more feasible options due to a possibly emerging antimicrobial resistance.


OBJECTIVE: To evaluate the clinically relevant abnormalities as visualized on CT and MR imaging in children with symmetric and asymmetric bilateral sensorineural hearing loss (SNHL), in relation to age and the severity of hearing loss.

STUDY DESIGN: Retrospective cohort study.

SETTING: Tertiary referral otology and audiology center.

PATIENTS AND DIAGNOSTIC INTERVENTIONS: From January 2006 until January 2016, a total of 207 children diagnosed with symmetric and asymmetric bilateral SNHL were included. They underwent CT and/or MR imaging for the evaluation of the etiology of their hearing loss.

MAIN OUTCOME MEASURES: Radiologic abnormalities associated with SNHL.

RESULTS: 302 scans were performed in 207 children (median age of 0.8 years old) with bilateral SNHL. The most frequently identified cause of bilateral SNHL was a malformation of the labyrinth. The combined diagnostic yield of CT and MR imaging was 32%. The diagnostic yield of MR (34%) was considerably higher than that of CT (20%). We found a higher rate of abnormalities in children with profound hearing loss (41%) compared to milder hearing loss (8-29%), and in asymmetric SNHL (52%) compared to symmetric SNHL (30%).

CONCLUSION: Imaging is essential in the etiologic evaluation of children with bilateral SNHL. The highest diagnostic yield is found in children with bilateral asymmetric SNHL or profound SNHL. Based on our findings, MR is the primary imaging modality of choice in the etiologic evaluation of children with bilateral SNHL because of its high diagnostic yield.


OBJECTIVES: Platinum-based chemotherapy is effective against a variety of pediatric malignancies. Unfortunately, the use of cisplatin and carboplatin can lead to permanent and progressive sensorineural hearing loss which can affect the quality of life of cancer survivors. The objectives of this study were to evaluate the incidence of platinum-induced ototoxicity in children and analyze potential risk factors.

METHODS: Prospective cohort study. All pediatric patients receiving chemotherapy with cisplatin and/or carboplatin from 01/2012 until 10/2017 were included. Hearing evaluations were performed before every chemotherapy cycle, and following the end of chemotherapy, with auditory brainstem response, otoacoustic emissions and/or audiometry. Demographics, cumulative doses, cranial irradiation and exposure to other ototoxic agents were analyzed.

RESULTS: Twenty-eight patients were included, with a mean age of 7.2 years at the beginning of chemotherapy (range 5 months-15 years 2 months); twenty-one patients received cisplatin, four received carboplatin, and three received both agents. Twelve patients had cranial irradiation and seven received another ototoxic medication. The most frequent malignancies were germ cell tumors, medulloblastoma and gliomas. Sensorineural hearing loss occurred in 28.6% of the patients with a median follow-up period of 21.5 months (range: 1-53 months). All patients evaluated with audiometry had ≥ Chang 2b ototoxicity. Risk factors include age less than 5 years, cranial irradiation, and cisplatin cumulative dose greater than 400 mg/m².

CONCLUSION: Sensorineural hearing loss is a potential side effect of platinum-based chemotherapy. Pediatric patients receiving cisplatin chemotherapy with a cumulative dose exceeding 400 mg/m², cranial irradiation as well as patients younger than 5 years are at greater risk of developing hearing loss.


BACKGROUND: At very high doses, furosemide is linked to ototoxicity in adults, but little is known about the risk of hearing loss in premature infants exposed to furosemide.

AIMS: Evaluate the association between prolonged furosemide exposure and abnormal hearing screening in premature infants.

STUDY DESIGN: Using propensity scoring, infants with prolonged (≥28 days) exposure to furosemide were matched to infants never exposed. The matched sample was used to estimate the impact of prolonged furosemide exposure on the probability of an abnormal hearing screen prior to hospital discharge.


OUTCOME MEASURES: We defined abnormal hearing screen as a result of either “fail” or “refer” for either ear.
RESULTS: Altogether, 1020 infants exposed to furosemide for ≥28 days were matched to 790 unique infants never exposed, yielding a total of 1042 matches due to sampling with replacement and propensity score ties. Matching resulted in a population similar in baseline characteristics. After adjusting for covariates, the proportion of infants with an abnormal hearing screen in the furosemide-exposed group was not significantly higher than the never-exposed group (absolute difference 3.0% [95% CI -0.2-6.2%], P = 0.07).

CONCLUSIONS: Prolonged furosemide exposure was associated with a positive, but not statistically significant, difference in abnormal hearing screening in premature infants. Additional studies with post-hospital discharge audiology follow-up are needed to further evaluate the safety of furosemide in this population.


The current study retrospectively investigated variations in audiological phenotypes in children with GJB2 gene mutations. Subjects were 128 infants and young children who were seen as outpatients by Otology at Beijing Tongren Hospital from 2012 to 2018. Of the 128 subjects, 99 had biallelic truncating (T/T) mutations and 29 had truncating/nontruncating (T/NT) mutations. Genotypes, results of universal newborn hearing screening (UNHS), and the degree and symmetry of hearing loss were examined in the two groups. Twenty-two subjects (20.37%, 22/108) passed UNHS, including 13 children with T/T mutations and 9 with T/NT mutations. Of the 128 subjects, 22 had normal hearing, 2 had unilateral hearing loss, and 115 had bilateral hearing loss. Severe-to-profound hearing loss was the most prevalent phenotype in children with T/T mutations (73.23%), while normal hearing was prevalent in children with T/NT mutations (41.38%). Symmetrical hearing loss was the main phenotype in both groups, and the number of subjects with symmetrical hearing loss did not differ significantly between the two groups. Therefore, children with GJB2 gene mutations have phenotypic variability in terms of their results of UNHS and their degree and symmetry of hearing loss. Subjects with T/NT mutations of the GJB2 gene were more likely to pass UNHS and had milder hearing loss compared to those with T/T mutations. Symmetrical hearing loss was the main phenotype in the two groups, but 36.53% of children had bilateral asymmetric hearing loss. Parents of all subjects with sensorineural hearing loss were informed that their children may have a GJB2 mutation.


OBJECTIVES: To evaluate the prevalence of middle ear disease in infants referred for failed newborn hearing screening (NBHS) and to review patient outcomes after intervention in order to propose an evidence-based protocol for management of newborns with otitis media with effusion (OME) who fail NBHS.

METHODS: 85 infants with suspected middle ear pathology were retrospectively reviewed after referral for failed NBHS. All subjects underwent a diagnostic microscopic exam with myringotomy with or without placement of a ventilation tube in the presence of a middle ear effusion and had intra-operative auditory brainstem response (ABR) testing or testing at a later date.

RESULTS: At the initial office visit, a normal middle ear space bilaterally was documented in 5 babies (6%), 29/85 (34%) had an equivocal exam while 51/85 (60%) had at least a unilateral OME. Myringotomy with or without tube placement due to presence of an effusion was performed on 65/85 (76%) neonates. Normal hearing was established in 1785 (20%) after intervention, avoiding the need for any further audiologic workup. Bilateral or unilateral sensorineural hearing loss (SNHL) or mixed hearing loss was noted in 54/85 (64%) and these children were referred for amplification. Initially observation with follow up outpatient visits was initiated in 27/85 (32%) however, only 3/27 (11%) resolved with watchful waiting and 24/27 (89%) ultimately required at least unilateral tube placement due to OME and 14/24 (59%) were found to have at least a unilateral mixed or SNHL.

CONCLUSIONS: An effective initial management plan for children with suspected middle ear pathology and failed NBHS is diagnostic operative microscopy with placement of a ventilation tube in the presence of a MEE along with either intra-operative ABR or close follow-up ABR. This allows for the identification and treatment of babies with a conductive component due to OME, accurate diagnosing of an underlying SNHL component and for prompt aural rehabilitation.


BACKGROUND/OBJECTIVE: Sensorineural hearing loss is a common diagnosis among children. The diagnostic workup varies widely among practitioners. This study's aim was to assess the utilization of diagnostic testing for SNHL and determine the yield of each test.

STUDY DESIGN: Retrospective chart review.

SETTING: Tertiary care center.

SUBJECTS: 827 patients with a diagnosis of SNHL from January 1, 2011 to January 1, 2015.

RESULTS: 746 patients met inclusion criteria. Temporal bone imaging was performed on 561 (75%) of patients with 224 (40%) having positive results that explained the etiology of the SNHL. Congenital SNHL was more likely to be associated with abnormal imaging than acquired SNHL (109/299 versus 106/316 respectively) (p = 0.001). Unilateral SNHL was more likely to be associated imaging abnormalities than bilateral SNHL (101/221 and 123/340 respectively) (p = 0.028). Genetic testing was performed on 244 (33%) patients, of which 94 (39%) had abnormalities. Positive genetics results were more common with bilateral than unilateral SNHL (82/191 and 12/53 respectively) (p = 0.007). There was no statistically significant difference in
the utility of genetic testing for congenital and acquired SNHL (p = 0.0836). Cytomegalovirus (CMV) testing was available for 104 (14%) of patients with 13 (12.5%) being positive and consistent with congenital CMV. Electrocardiogram, urinalysis, and Lyme titers were less useful.

CONCLUSIONS: Imaging and genetic testing had the highest yield in the evaluation of children with SNHL and were the most commonly performed. CMV testing was valuable in neonates who failed newborn hearing screening.

Zahed Pasha Y, Zamani M, Hashemi Fard A, Zahed Pasha E.
**Screening of Hearing in Newborn Infants: Follow-Up and Outcome After 40930 Births in Babol, Northern Iran.** Arch Iran Med.
2018 Sep 1;21(9):382-386.

**BACKGROUND:** The purpose of this study was to investigate the results of hearing screening in all newborn infants, and their follow-up in Babol, northern Iran.

**METHODS:** Between 2006 and 2014, all healthy neonates delivered in 3 hospitals were included in this cross-sectional study. Newborns were screened using the transient evoked otoacoustic emissions test before discharge. Those who failed to pass the examination were tested for auditory brainstem response (ABR) by the age of one month. The infants referred from the previous level underwent tests of auditory steady state response, ABR and impedanced audiology before the age of 3 months. For infants with the diagnosis of bilateral hearing impairment, it was recommended to use a hearing aid in 3 months. Then, their parents were recommended to take infants again to the hearing testing centers within next 6 months. If the infant's hearing was not improved, he/she was advised to undergo cochlear implantation.

**RESULTS:** In total, 40930 newborns were screened. Out of them, 62 (1.5 per 1000 live births) were finally diagnosed to have hearing impairment, of whom 14 had unilateral and 48 had bilateral disorders (candidate for supportive measures). Overall, 986 (2.4%) were lost to follow-up and 11 (0.03%) died over the first 3 months of age. At the end of the 6-month follow-up for supportive stage, 15 out of 48 infants received a hearing aid and 18 (0.4 per 1000 children) underwent cochlear implant surgery. Fourteen out of 48 cases were lost to follow-up over supportive stage.

**CONCLUSION:** It is recommended that all newborns undergo hearing screening test before hospital discharge, and those with impairment receive supportive measures from 3 months of age, and be re-examined at 12 months of age.

Zhao P, Lin L, Lan L.
**Analysis of mutation spectrum of common deafness-causing genes in Hakka newborns in southern China by semiconductor sequencing.** Medicine (Baltimore).
2018 Sep;97(38):e12285. doi: 10.1097/MD.0000000000012285.

Hearing loss is a common neurosensory disorder, approximately half of the cases are caused by genetic factors, and approximately 70% of hereditary hearing impairments are nonsyndromic hearing loss (NSHL). The mutations of GJB2 (gap junction beta-2 protein), GJB3 (gap junction beta-3 protein), SLC26A4 (solute carrier family 26 member 4), and MT-RNR1 (mitochondrionally encoded 12S RNA) are the most common inherited causes of NSHL. Because of different genetic backgrounds, the mutation spectrum of these common deafness-causing genes varies among different regions in China. Because no data are known on these mutations among the Hakka population of Southern China, we aim to investigate the mutation spectrum to add these to neonatal screening and genetic counseling. A total of 1252 blood samples from newborns have been detected by semiconductor sequencing for 100 mutations loci of 18 deafness-causing genes. Of the participants, 95 subjects carried deafness-causing genes mutations with the carrier rate of 7.59%. The mutation frequencies of GJB2, SLC26A4, GJB3, and mitochondrial genes were 3.04%, 3.51%, 0.16%, and 0.88%, respectively. We followed up subjects with single-gene homozygous or compound heterozygous mutations. Our study firstly analyzed deafness-causing genes mutation spectrum in Hakka population, providing evidence for future neonatal screening and genetic counseling in this area.

Zhao XL, Huang LH, Wang XY, DU Y, Wang X, Cheng XH, Zhao LP, Li Y.
**Analysis of genotypes and audiological characteristics of children with SLC26A4 gene pathogenic mutations.** Lin Chung Er Bi Yan Hou Jing Wai Ke Za Zhi.

**Objective:** To explore the correlation of SLC26A4 genotype and audiology. **Method:** The subjects were 70 children aged 0 to 7 years old, who were admitted to otological outpatient department. All subjects received nine crystal hereditary deafness gene chip and confirmed by (or)SLC26A4 gene full coding region detection. The patients were diagnosed as homozygous or compound heterozygous mutations. At the same time, acoustic immittance, auditory brainstem response, auditory steady state response and pediatric behavior audiometry, newborn hearing screening and other audiological tests were displayed. According to the genotype, the subjects were divided into two groups: group A (SLC26A4 gene homozygous mutation) in 40 cases, group B (SLC26A4 gene compound heterozygous mutation) in 30 cases. The frequency of SLC26A4 gene mutation, the two groups of genotypes and hearing screening results, the degree of hearing loss and audiometric configurations were analyzed statistically.

**Result:** In 70 patients, the top 4 of the 70 patients with high frequency of mutations were IVS7-2A> G (76.43%), 2168A>G (15.00%), 1226G> A (2.86%) and 2000T>C (2.16%), respectively. 34.29% of newborns passed hearing screening with single or double ears, among which group A and group B were 32.50% and 36.67%, respectively. There was no statistically significant difference between two groups in hearing screening. The degree of hearing loss in group A (56.25%) and group B (48.33%) were mainly profound and there was no significant difference between them. The audiometric configurations: group A (60.00%) was mainly high frequency loss type, while group B (55.00%) was mainly flat type. The difference between them was statistically significant. **Conclusion:** The mutation sites of SLC26A4 gene were mainly IVS7-2A> G, and the degree of hearing loss was mostly profound. To the audiometric configurations, SLC26A4 gene homozygous mutant were mainly high frequency loss type, while SLC26A4 gene compound heterozygous mutant were mainly flat type. 34.29% children passed universal newborn hearing screening with one ear at least, which indicates SLC26A4 gene mutations can result in late-onset hearing loss, so those patients should be attached great importance.