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This issue features an excellent range of articles reflecting our goal of publishing current research, evidence based practice, and standards of care. It is our intention to disseminate timely information reflecting the broad range of topics typically associated with Early Hearing Detection and Intervention programs.

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Thanks for making our first issue such a resounding success. We look forward to continuing to be THE go-to source for information relevant to all areas of Early Hearing Detection and Intervention.

Les R. Schmeltz, Au.D.
Editor-In-Chief
Progress in Standardization of Reporting and Analysis of Data from Early Hearing Detection and Intervention (EHDI) Programs

Suhana Alam, MPH.,1 Ashley Satterfield, MPH.,2 Craig A. Mason, Ph.D.,3 Xidong Deng, Ph.D.,1

1Centers for Disease Control and Prevention, Atlanta, GA
2Oak Ridge Institute for Science and Education, Oak Ridge, TN
3University of Maine, Orono, ME

Abstract
Congenital hearing loss affects one to three of every 1,000 live born infants. If left undetected, it may negatively impact children through delayed speech and language development. To help avoid developmental delays and ensure that deaf or hard of hearing (DHH) infants are identified and receiving services as early as possible, complete and accurate data are crucial. Despite substantial progress made over the years, some children are still delayed in identification and/or lost to the early hearing detection and intervention (EHDI) surveillance and tracking systems. Lack of standardization in data reporting contributes to this issue. This article discusses reasons for lack of standardization in data reporting and gives suggestions for how the situation could be improved.

Acronyms: CDC = Centers for Disease Control and Prevention; CMS = Centers for Medicare & Medicaid Services; DHH = Deaf or Hard of Hearing; EHDI = Early Hearing Detection and Intervention; EHDI-IS = EHDI Information System; EI = Early Intervention; HSFS = Hearing Screening and Follow-up Survey; JCIH = Joint Committee on Infant Hearing; LFU/LTD = Lost to Follow-Up or Lost To Documentation; NQF = National Quality Forum

Corresponding author’s contact information: Suhana Alam, MPH. Early Hearing Detection and Intervention, National Center on Birth Defects and Developmental Disabilities, Centers for Disease Control and Prevention. 4770 Buford Highway, MS E-88, Atlanta Georgia 30341-3717, United States. Phone: 404.498.6084 Fax: 404.498.3060; Email: SAlam1@cdc.gov

Introduction
Implementation of routine newborn hearing screening, known as universal newborn hearing screening, has provided the opportunity for infants who are deaf or hard of hearing (DHH) to be identified shortly after birth, and as a result, the age of identification for most babies in the United States has decreased from 2½ years to 2-3 months of age (White, Forsman, Eichwald, & Munoz, 2010). When late identified and therefore delayed in opportunities to acquire language and communication skills, these children will likely fall behind their hearing peers in communication, cognition, reading, and social-emotional development (Pimperton & Kennedy, 2012). With the widespread implementation of early hearing detection and intervention (EHDI) programs across the United States, more than 95% of newborns now receive a hearing screening, usually before hospital discharge (Centers for Disease Control and Prevention [CDC], 2016a).

However, providing a hearing screening is only the first step in the process for infants who do not pass the screening. To maximize the benefits of screening and to ensure early identification, it is essential that infants who do not pass the screening receive timely follow-up testing to confirm their hearing status. Infants diagnosed as DHH should receive early intervention services that meet the needs and preferences of the child and family. Timing from screening to enrollment into early intervention is crucial, so the Joint Committee on Infant Hearing (JCIH) recommends: (a) hearing screening no later than one month of age; (b) a diagnostic evaluation before three months of age for those who did not pass the newborn hearing screening; and (c) enrollment into early intervention services before six months of age for those who are diagnosed with hearing loss (JCIH, 2007). These recommendations are commonly referred to as the 1-3-6 benchmarks.

To ensure that DHH infants are receiving timely services, complete and accurate data reporting from hospitals, audiologists, and other providers to the state or territorial EHDI program1 is crucial (Mason, Gaffney, Greene, & Gross, 2008). To help assess progress toward the 1-3-6 benchmarks the CDC EHDI program developed the Hearing Screening and Follow-up Survey (HSFS). This voluntary survey is completed by EHDI program staff and was designed in collaboration with partners that included Directors of Speech and Hearing Programs in State Health and Welfare Agencies, the Health Resources and Services Administration, and other stakeholders. The survey gathers non-estimated data related to the receipt of hearing screening, diagnostic testing, and enrollment into early intervention for all occurrent births within a jurisdiction in a given year.

1Throughout the remainder of this article, “jurisdiction” will be used to refer to states, territories, and other political jurisdictions that operate screening programs such as Washington, D.C., Puerto Rico, the Virgin Islands, etc.
In addition to monitoring progress toward the 1-3-6 benchmarks, the HSFS also allows the CDC to monitor progress in other areas, such as the number of infants not receiving or not documented to have received recommended follow-up services. These infants are referred to as being lost to follow-up or lost to documentation (LFU/LTD). Information gathered through the HSFS also allows CDC to collaborate and provide technical assistance to EHDI programs that need assistance. Since 2005, jurisdictions have been asked to complete and submit the survey annually and because the survey is voluntary, the response rate varies from year to year. For the year 2014, 57 of 59 (97%) jurisdictions completed the HSFS. Despite the significant progress in screening rates made over the years and improved efforts of public health programs and health care providers to ensure that all infants and children receive their recommended follow-up services, the LFU/LTD rate is still high in some jurisdictions and some children still fall through the cracks and are lost to the EHDI tracking and surveillance systems.

Reasons for Lack of Standardization in EHDI

Lack of standardization in reporting data regarding screening and diagnostic follow-up testing has contributed to some infants becoming LFU/LTD. Nationally, it is difficult to monitor children needing follow-up services and to accurately assess progress toward the 1-3-6 benchmarks when local data are incomplete and/or inconsistent. JCIH recognized the need for standardization of data definitions and reporting practices and their 2007 position statement noted that standardized reporting is crucial and that all federal and state agencies should standardize data definitions for higher quality and more reliable data (JCIH, 2007).

Three primary reasons contribute to the lack of standardized data for EHDI programs across the nation. First, there are variations in the degree of completeness of data that jurisdictions report each year, which impacts national estimates. This is illustrated in Table 1 where a hypothetical country X is comprised of three jurisdictions: A, B, and C. Theoretically, the most accurate percentage of children with a confirmed hearing loss enrolled in early intervention (EI) is 67.2%, which includes all three jurisdictions (Equation 1). However, if Jurisdiction A did not report early intervention data, the percentage of children enrolled in EI would be reduced from the accurate 67.2% to 59.7% (Equation 2). When a jurisdiction is not able to report information on enrollment in EI or other data items, it impacts the representativeness of the national estimates. This could be due to the EHDI program not being linked with the EI program, which can occur when there is no data sharing agreement in place or the privacy laws within the jurisdiction disallow it. It could also be due to limitations with the functionality of the jurisdiction’s EHDI Information System (EHDI-IS) that affects their ability to report all data. Limitations occur because although every jurisdiction currently has an EHDI-IS, the design and capabilities of these systems range from basic to advanced, impacting what can be reported. It is also possible that the jurisdiction is directed to only report certain data.

Second, despite substantial progress made in development and use of the EHDI-IS, challenges remain in ensuring complete documentation of services for the entire newborn

<table>
<thead>
<tr>
<th>Jurisdiction</th>
<th>Screening Not Pass Screen</th>
<th>Diagnostics Diagnosed</th>
<th>Early Intervention Hearing Loss Confirmed</th>
<th>Enrolled</th>
</tr>
</thead>
<tbody>
<tr>
<td>A</td>
<td>1,625</td>
<td>750</td>
<td>225</td>
<td>167</td>
</tr>
<tr>
<td>B</td>
<td>2,364</td>
<td>1,911</td>
<td>145</td>
<td>83</td>
</tr>
<tr>
<td>C</td>
<td>3,404</td>
<td>2,328</td>
<td>66</td>
<td>43</td>
</tr>
<tr>
<td><strong>Total</strong></td>
<td><strong>7,393</strong></td>
<td><strong>4,989</strong></td>
<td><strong>436</strong></td>
<td><strong>293</strong></td>
</tr>
</tbody>
</table>

Correct percent of children enrolled in EI 67.2% or 293/436  
Reported percentage of children enrolled in EI 59.7% or (83+43)/(145+66)

*Note. EI = Early Intervention.*
population. This makes it difficult to ensure all infants are receiving recommended services and to generate accurate national estimates. Currently, screening results are consistently reported to the jurisdictional EHDI programs; however, the same does not apply to diagnostic test results and enrollment in EI. Reporting of EHDI data is not mandated by law in some jurisdictions. Infants who are referred for diagnostic evaluation and/or EI but did not receive recommended diagnostic and/or intervention services are commonly classified as LFU. Situations where an infant received the recommended diagnostic evaluation and/or intervention, but was never reported to the EHDI program, are referred to as LTD. Because it is difficult for EHDI programs to differentiate between infants who are LFU and those who are LTD, terms are typically used together. Table 2, which focuses on the screening stage, illustrates how a lack of documentation affects national estimates, using a hypothetical cohort of 100,000 births. Theoretically, the true overall screening rate is 95.6% (Equation 3), which includes all children who were screened, both documented and undocumented. However, the reported screening rate would be 86.1% (Equation 4), which is based on only those infants with a documented screen. This is an underestimate in comparison to the correct 95.6%. In addition, 11.7% of infants are LFU/LTD (Equation 5). Because of LTD, any reported LFU/LTD rate may not necessarily reflect the true burden of LFU/LTD. Time and resources could be unnecessarily expended on tracking those LFU/LTD children who already received services, subsequently reducing the efficiency of the jurisdictional EHDI program. Missing data is also a problem for infants who do not pass the hearing screening but are later found to have a normal hearing because they artificially inflate the estimated rate of hearing loss among infants who did not pass the screening.

Third, jurisdictions may define and calculate LFU/LTD in different, non-standardized ways. Despite formulas being provided and multiple instructional sessions about how to use the specified formulas, not all jurisdictions follow the guidance for the HSFS. The CDC defines LFU/LTD on the HSFS based on infants who are referred for follow-up but are not documented as having received it for one of the following three specific reasons: (a) unable to contact the family, (b) the family was contacted but unresponsive, or (c) reason unknown. Reasons such as the infant deceased, the family moved, the parents declined, or the physician did not refer the infant, are not counted in LFU/LTD because the status of these infants is known to the EHDI program. The percentage of infants who are LFU/LTD for diagnostics is calculated by taking the number of infants LFU/LTD for diagnostics divided by the total number of infants not passing screening, then multiplying by 100%. The percentage LFU/LTD for early intervention is calculated by taking the number of infants LFU/LTD for EI divided by the total number of infants confirmed to have a permanent hearing loss, then multiplying by 100%. Table 3 reflects variation in calculating LFU/LTD for diagnosis, using a hypothetical cohort of 800 infants who did not pass the hearing screening and needed a diagnostic evaluation. For this scenario, according to the CDC guidance, the LFU/LTD for diagnosis would be 39.1% (Equation 6). Jurisdiction A, however, may calculate and report LFU/LTD differently in their reports and include all reasons except infant death, arriving at 48.1% (Equation 7). In contrast, Jurisdiction B may exclude from the LFU/LTD category infants whose families were unresponsive. This would bring their LFU/LTD estimate to 2.3% (Equation 8), an underestimate in comparison to the CDC’s recommended formula of 39.1%. As can be seen, adopting definitions not in accordance to the HSFS guidance contributes to lack of standardization.

Table 2. Effect of Documentation Status on Data

<table>
<thead>
<tr>
<th>Actual Screening status</th>
<th>N</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td>Screening documented</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Completed Screening</td>
<td>86,126</td>
<td>85.1</td>
</tr>
<tr>
<td>Incomplete Screening</td>
<td>2,154</td>
<td>10.3</td>
</tr>
<tr>
<td>Screening not documented</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Screening did in fact occur (LTD)</td>
<td>9,512</td>
<td>1.3</td>
</tr>
<tr>
<td>Screening did not in fact occur (LFU)</td>
<td>2,208</td>
<td>3.3</td>
</tr>
<tr>
<td>Total</td>
<td>100,000</td>
<td></td>
</tr>
</tbody>
</table>

Note. LFU/LTD=lost to follow-up/lost to documentation
Lack of standardization occurs for many reasons. It can occur due to data programming within the EHDI-IS that collects and stores information in varying degrees of detail and granularity. It can occur at the local hospital/provider level with differences in what information is reported. It can occur at the jurisdictional level if EHDI programs calculate rates differently. And it can occur at the national level when jurisdictions change how they classify and/or report data in different years. Consequently, it is difficult to estimate the true number of children who are DHH and are not receiving follow-up services and to compare the data across jurisdictions and years. In response, the CDC and its partners have made a number of efforts to address this issue.

**What has been done to address lack of standardization?**

To help increase the standardization of data the CDC has made several updates to the HSFS since its inception based on feedback from jurisdictions and analyses of the reported data. One example is the updating of the definition for “In Process” for diagnostic evaluation, which has been revised twice to allow for more accurate reporting on the HSFS. In Process can occur when additional testing is needed to make a definitive diagnosis for an infant that did not pass the hearing screening. The revisions in the definition were made due to a higher than expected number of infants being reported in this category. The initial definition referred to infants that did not pass a hearing screening and did not yet have a confirmed diagnosis. In 2007, the definition was narrowed to specify that the infants reported in this category must have been seen by an audiologist for diagnostic evaluation at least once. If the infant was diagnosed as having a permanent hearing loss, it was to be reported as a “confirmed hearing loss,” even if the degree of hearing loss (e.g., moderate or severe) was still undetermined. The definition clarified that scheduling an appointment for an initial evaluation or only making a referral to an audiologist was not considered as In Process. In 2009, the definition was further refined and required that infants reported in this category not only must have been seen by an audiologist for diagnostic evaluation at least once, but also must have a follow-up appointment already scheduled. The change in definition has improved the accuracy of this data and the percent of infants reported as In Process has decreased from 16.0% in 2006 to 1.8% in 2013.

As another example, in 2015 the CDC collaborated with the EHDI Data Committee, which includes representatives from jurisdictional EHDI programs, the National Center for Hearing Assessment and Management, and other stakeholders to revise the HSFS’s “Unresponsive” definition. Unresponsive is one of the three categories used by the CDC to calculate rates of LFU/LTD for diagnosis and intervention. The previous definition, “Parents or family of an infant who did not pass the screening were contacted but there was no documented response” was considered broad and contributed to a lack of standardization in data reported in the HSFS. The revised definition specified that for a case to be identified as “Unresponsive” the EHDI program or healthcare provider must have a documented two-way conversation or written communication with the child’s legal parent or guardian in which the parent or guardian acknowledged awareness of the corresponding 1-3-6 recommendation and had nevertheless not obtained

### Table 3. Adopting Different Definitions of Lost to Follow Up/Loss to Documentation

<table>
<thead>
<tr>
<th>Actual Screening status</th>
<th>N</th>
</tr>
</thead>
<tbody>
<tr>
<td>In process</td>
<td>5</td>
</tr>
<tr>
<td>Non-resident</td>
<td>29</td>
</tr>
<tr>
<td>Moved out of jurisdiction</td>
<td>15</td>
</tr>
<tr>
<td>Medical reason</td>
<td>0</td>
</tr>
<tr>
<td>Physician did not refer</td>
<td>1</td>
</tr>
<tr>
<td>Infant died</td>
<td>8</td>
</tr>
<tr>
<td>Parents/family declined</td>
<td>22</td>
</tr>
<tr>
<td>Parents contacted but unresponsive</td>
<td>295</td>
</tr>
<tr>
<td>Unable to contact</td>
<td>4</td>
</tr>
<tr>
<td>Unknown</td>
<td>14</td>
</tr>
<tr>
<td>Percentage of LFU/LTD, according to the CDC</td>
<td>39.1% or (295+4+14)/800</td>
</tr>
<tr>
<td>Percentage of LFU/LTD, according to Jurisdiction A</td>
<td>48% or (5+29+15+0+1+22+295+4+14)/800</td>
</tr>
<tr>
<td>Percentage of LFU/LTD, according to Jurisdiction B</td>
<td>2.3% or (4+14)/800</td>
</tr>
</tbody>
</table>

Note. LFU/LTD=lost to follow-up/lost to documentation; CDC = Centers for Disease Control and Prevention
In 2015, the CDC convened the EHDI Functional Standard Working Group, which included EHDI program staff from several states, and created the EHDI-IS Functional Standards (CDC, 2016b). These standards provide jurisdictions with guidance on the technical and functional requirements for a complete EHDI-IS and are intended to identify the operational, programmatic, and technical criteria that all jurisdictional EHDI programs should implement when developing, using, and evaluating an EHDI-IS. The Functional Standards also define a set of data items that are considered to be essential for the EHDI tracking and surveillance process and aims to set the standard for minimum data collection at the jurisdictional level. Having an EHDI-IS that meets these requirements will better enable jurisdictions to collect, use, and provide complete and accurate data.

In addition to the above mentioned efforts to address the lack of standardization, the CDC has supported national standardization initiatives to improve interoperability between clinical electronic health records and public health information systems. Interoperability describes the extent to which systems and devices can exchange data and interpret that shared data. Within health care it refers to the ability for systems to work together and exchange information within and across organizational boundaries to advance the delivery of health care services. CDC EHDI is working to leverage advances in health information technology to ensure infants receive recommended services and improve standardization by helping connect public health and clinical services. As part of this effort the CDC is: (a) establishing national standards on information exchange and electronic quality measures (eMeasures), (b) promoting the use of standards to support data exchange with electronic health records, and (c) developing standards-based tools to support clinical care coordination to help ensure infants receive recommended follow-up services. These standards and tools are designed to improve how data are collected, analyzed, and used, as well as strengthening service coordination between public health and early intervention providers.

To better utilize the surveillance data and to assess the performance of the EHDI process in a standard manner, CDC has developed three EHDI-related quality measures that were re-endorsed in 2015 by the National Quality Forum (NQF): Hearing screening prior to hospital discharge (NQF#1354), diagnostic evaluation no later than 3 months of age (NQF#1360), and signed Part C Individual Family Service Plan before 6 months of age (NQF#1361). The NQF is a not-for-profit, nonpartisan, membership-based organization that works to make improvements in health care by setting standards, recommending measures for public programs, identifying and accelerating quality improvement, advancing electronic measurement, and providing information and tools to aid health care workers in decision-making. An NQF endorsement reflects scientific, evidence-based review, patient and family input, and the perspectives of the health care industry.

The EHDI quality measures have been well received and adopted by a number of healthcare organizations. For example, the Joint Commission (formerly the Joint Commission on Accreditation of Healthcare Organizations) — the nation’s oldest and largest standards-setting accrediting body in healthcare — has adopted NQF#1354 “Hearing Screening Prior to Hospital Discharge” for their 2016 data reporting. Any accredited hospital may choose this measure set as one of their six required sets to satisfy their accreditation requirements. Similarly, an eMeasure version of this same measure is included as one of the 29 hospital measures in the 2017 reporting period for Stage 2 of the Centers for Medicare & Medicaid Services (CMS) Meaningful Use Incentive Program. NQF#1360 (audiology evaluation no later than 3 months of age), was recently included in the CMS’s 2016 Core Set of Children’s Health Care Quality Measures (Child Core Set). Implementation of a standardized Child Core Set is helping the CMS and states move toward a national system for quality measurement, reporting, and improvement.

To help jurisdictions understand these standards and measures, the CDC regularly holds webinars and meetings to educate and discuss with EHDI program staff ideas about how to improve reporting and documentation. Members of the EHDI Data Committee hold monthly conference calls to discuss methods to report more standardized data and to further improve quality. CDC EHDI staff members also participated in the standard development committee meetings and have recorded educational webinars on the interoperability standards that have been developed.

Conclusion/Next Steps

Lack of standardization for EHDI data occurs for several reasons and adversely affects the quality and accuracy of data. This makes it difficult to capture the true number of infants who are DHH and in need of services and to accurately assess progress toward the 1-3-6 benchmarks. It also makes it difficult to evaluate an EHDI program’s effectiveness and overall success. The consistent availability of standardized data will better enable EHDI programs to ensure that all infants who are DHH are identified early and receive the services they need in a timely manner. However, improving and maintaining data standardization requires continuous commitment and collaboration around the collection and reporting of complete and accurate data among jurisdictional EHDI programs, providers, the CDC, and other stakeholders.
This can be accomplished by increasing awareness of the need for data standardization and improved reporting practices. Generating and assessing the data in a timely manner will also support this ongoing progress. The CDC EHDI program will continue to collaborate with and provide technical assistance to jurisdictional EHDI programs to strengthen their EHDI-IS, which will in turn expand capacity to collect and report complete and accurate data. Other efforts include updating and promoting the use of national standards on information exchange and electronic quality measures and supporting research to study the impact of complete and accurate data on the success of EHDI programs.

References


Tyson S. Barrett, B.S., 1,2 Karl R. White, Ph.D.,1,2

1National Center for Hearing Assessment and Management (NCHAM)
2Department of Psychology, Utah State University

Abstract
A recent highly cited publication, using data from the National Health and Nutrition Examination Survey (NHANES), concluded that the prevalence of childhood hearing loss in the United States is increasing (Shargorodsky, Curan, Curhan, & Eavey, 2010). This article examines the accuracy of that conclusion based on additional data from three nationally-representative surveys of childhood health. Using data from NHANES, the National Survey of Children's Health (NSCH), and the National Health Interview Survey (NHIS), logistic regression was used to assess trends from audiometry-measured and parent-reported childhood hearing loss.

In contrast to prior research, the results were highly conflicting. NHANES suggested both an increasing (audiometry) and decreasing (parent-report) trend, NSCH (parent-report) suggested no trend, and NHIS (parent-report) suggested a possible increasing trend. Given the disagreements among these federally funded national surveys, administrators and policy makers should be very cautious about conclusions drawn from these surveys regarding prevalence and trends related to childhood hearing loss in the United States.

Key Words: hearing loss; prevalence; trend; NHANES; NHIS; NSCH

Acronyms: NHANES = National Health and Nutrition Examination Survey; NHIS = National Health Interview Survey; NSCH = National Survey of Children’s Health; P/S Report = Parent/Self Report; PTA = pure tone averages

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Corresponding author’s contact information: Tyson S. Barrett; Utah State University, 2615 Old Main, Logan, UT, 84322; Email: t.barrett@aggiemail.usu.edu

Introduction
Hearing loss frequently has serious negative consequences, especially for children (Smith, Bale, & White, 2005). Childhood hearing loss impacts many aspects of the child’s life. It hinders a child’s development including speech, language, and social development (Theunissen et al., 2014; Tomblin, Oleson, Ambrose, Walker, & Moeller, 2014; Warner-Czyz, Loy, Roland, Tong, & Tobey, 2008; Yoshinaga-Itano, 2003). Even a mild loss in hearing for a child can seriously hamper the child’s ability to develop language and succeed in school (Bess, Dodd-Murphy, & Parker, 1998; Blair, Peterson, & Viehweg, 1985; Davis, 1989; Davis, Elfenbein, Schum, & Bentler, 1986; Festen & Plomp, 1990), whether that loss is bilateral (both ears) or unilateral (one ear; Bess & Tharpe, 1986; Brookhouser, Worthington, & Kelly, 1991; Lieu, 2004; Lieu, Tye-Murray, & Fu, 2012).

Research has shown that early diagnosis of hearing loss (preferably before 6 months of age) and subsequent enrollment in intervention services improved the speech, language, and social-emotional development of the child (Moeller, 2000; Pimperton & Kennedy, 2012; White, 2004; Yoshinaga-Itano, 2003). Further, school self-esteem was positively associated with earlier identification and intervention in children with hearing loss (Leigh, Maxwell-McCaw, Bat-Chava, & Christiansen, 2009).

Interventions to alleviate the negative consequences of childhood hearing loss are more likely to be implemented when policy makers and program administrators have correct information about the prevalence of hearing loss and whether prevalence is increasing or decreasing over time. For example, policies in the late 1990s and early 2000s advanced opportunities to help children with hearing loss (White, 2003). But, in order to continue to allocate the proper amount of resources, to assess recent policy efforts, and to study the epidemiology of childhood hearing loss, accurate estimation of prevalence and the temporal trend of childhood hearing loss is necessary. Otherwise, resources are unlikely to be appropriately allocated and the effects of policies and programs are unlikely to be well understood.

The United States federal government expends considerable money and effort to collect data about national prevalence and trends of various health-related variables. Probably the most well known and highly respected nationally representative data collection efforts related to children’s health in the United States are the National Health and Nutrition Examination Survey (NHANES; Curtin, Mohajder, & Dohrmann, 2010; Zipf, Chiappa, Porter,
Ostchega, Lewis, & Dostal, 2013), the National Survey of Children’s Health (NSCH, 2012), and the National Health Interview Survey (NHIS, 1997). Each is a systematically collected, well-documented survey collecting data on many health issues that affect the population of the United States. These cross-sectional surveys are designed to be nationally representative. Due to the high costs, both in time and resources, the federal government is likely the only entity capable of conducting such endeavors.

The way in which data from these federally-sponsored surveys are used to make important policy and programmatic decisions was highlighted in a recent article by Shargorodsky, Curan, Curhan, & Eavey (2010). Shargorodsky et al. used NHANES data to conclude that there had been a 31% increase in the prevalence of hearing loss in 2005–2006 compared with 1988–1994. Using the NHANES data, Shargorodsky et al. (2010) also concluded that there is higher prevalence of hearing loss among males compared to females, a positive correlation between income and childhood hearing loss, and that “vaccination against Haemophilus influenzae and Streptococcus pneumoniae, as well as greater awareness of music-induced hearing loss,” had not led to “…a reduction in the prevalence of hearing loss” (p. 776). They concluded that, “Further studies are needed to determine reasons for this increase and to identify potential modifiable risk factors to prevent the development of hearing loss” (p. 777).

There are many other cases where governmental, academic, and professional entities have used these federally-sponsored surveys to address important policy and administrative questions. For example, the Social Security Administration recently commissioned the Health and Medicine Division of the National Academies of Sciences, Engineering, and Medicine (NASEM; formerly known as the Institute of Medicine or IOM) to “identify past and current trends in the prevalence and persistence of speech and language disorders among the general U.S. population under 18 and compare those trends with trends among the SSI [Supplemental Security Income] childhood disability population (National Academies of Sciences, Engineering, and Medicine, 2016, p. 2). The report’s conclusions about prevalence relied heavily on the NHANES, NSCH, and NHIS data sets.

The NHANES, NSCH, and NHIS data sets have been used extensively to study health and well-being among children in the United States (e.g., Bitsko, Holbrook, Robinson, Kaminski, & Ghandour, 2016; Cprek, Williams, Asaolu, Alexander, & Vanderpool, 2015), including the prevalence of hearing loss (e.g., Boulet, Boyle, & Schieve, 2009; Nisker et al., 1998). Yet, even though the individual data sets have been used frequently to study childhood hearing loss, no studies that compared prevalence and trend results from the NHANES, NSCH, and NHIS data sets could be located. Reports using these data sources independently have apparently assumed that each source would likely give similar results; therefore, only one source was referenced. This assumption needs to be tested to know if these sources are a reliable way to estimate childhood hearing loss. In addition, it is important to point out that Shargorodsky et al.’s (2010) widely cited conclusion that childhood hearing loss in the United States is increasing was based on only two points in time (1988–1994 compared to 2005–2006) and only one data set (NHANES). The fact that more data are available from NHANES and that data on prevalence are available from other nationally-collected data sets means that questions about prevalence and trends in childhood hearing loss can be addressed more comprehensively than has been previously reported.

The present study, therefore, aims to answer two important questions. First, do these nationally representative surveys (NHANES, NSCH, and NHIS) agree on the prevalence and the direction/magnitude of the temporal trend of childhood hearing loss? Second, if they do agree, is childhood hearing loss increasing in the United States? To answer these questions, publically available data from the NHANES, the NSCH, and the NHIS were analyzed.

Method

Data

Data from three major national surveys were used: the NHANES across the years 1994 to 2010, the NSCH across the years 2007 to 2012, and the NHIS across the years 2005 to 2013. These years for each survey were chosen due to their availability, having data on childhood hearing loss, and having questions that are identical across years. For simplicity, we refer to each release by its final year (e.g., 2005–2006 is referred to as 2006). Analyses were performed in the survey package in the R statistical software environment developed for analyses of complex survey designs (Lumley, 2010). Table 1 presents descriptive statistics of the samples stratified by each survey. Note that in drawing conclusions about prevalence and trends in childhood hearing loss, the clustering and the non-random probability-sample were taken into account and, consequently, the proportions of the demographics are adjusted to be representative of the United States.

National Health and Nutrition Examination Survey (NHANES).

Releases of the NHANES data in 1994, 2006, 2008, and 2010 were used for the study because these are the only recent years with data on childhood hearing loss. The NHANES data set contains data on children ages 5–19 (although the 1994 NHANES data have information only on children ages 5–11). Although it would appear to be beneficial to include children up to age 17 as both the NSCH and the NHIS only include children 17 years old or younger (see descriptions of the NSCH and NHIS data sets below), NHANES stipulates that stratifying by age levels not predefined by the survey administrators can adversely affect the weighting scheme. Results were compared based on
Table 1: Descriptive Statistics of the Samples Stratified by Data Set.

<table>
<thead>
<tr>
<th></th>
<th>NHANES</th>
<th>NSCH</th>
<th>NHIS</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>(n = 10,542)</td>
<td>(n = 187,085)</td>
<td>(n = 300,844)</td>
</tr>
<tr>
<td><strong>Count (%)</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Parent/Self Report (Moderate +)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>No Loss</td>
<td>4,672 (98.9%)</td>
<td>186,050 (99.4%)</td>
<td>299,463 (99.5%)</td>
</tr>
<tr>
<td>Loss</td>
<td>54 (1.1%)</td>
<td>1,035 (0.6%)</td>
<td>1,381 (0.5%)</td>
</tr>
<tr>
<td>Examination (40dB+)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>No Loss</td>
<td>10,477 (99.1%)</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>Loss</td>
<td>95 (0.9%)</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>dB Threshold, right ear, mean (SD)</td>
<td>5.90 (7.22)</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>dB Threshold, left ear, mean (SD)</td>
<td>6.01 (7.13)</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>Age, mean (SD)</td>
<td>13.40 (3.79)</td>
<td>9.00 (5.29)</td>
<td>8.51 (5.2)</td>
</tr>
<tr>
<td>Sex</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Male</td>
<td>5,262 (49.9%)</td>
<td>96,744 (51.7%)</td>
<td>154,176 (51.2%)</td>
</tr>
<tr>
<td>Female</td>
<td>5,280 (50.1%)</td>
<td>90,341 (48.3%)</td>
<td>146,668 (48.8%)</td>
</tr>
<tr>
<td>Race</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>White</td>
<td>2,904 (27.4%)</td>
<td>136,143 (72.8%)</td>
<td>223,256 (74.2%)</td>
</tr>
<tr>
<td>Black</td>
<td>3,374 (32.0%)</td>
<td>18,877 (10.1%)</td>
<td>53,352 (17.7%)</td>
</tr>
<tr>
<td>Mexican American</td>
<td>3,389 (32.1%)</td>
<td>19,664 (10.5%)</td>
<td>-</td>
</tr>
<tr>
<td>Other</td>
<td>875 (8.3%)</td>
<td>4,441 (2.4%)</td>
<td>24,136 (8.0%)</td>
</tr>
<tr>
<td>Unknown</td>
<td>-</td>
<td>7,960 (4.3%)</td>
<td>-</td>
</tr>
<tr>
<td>End-Year Data Collected</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>1994</td>
<td>6,166 (58.5%)</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>...</td>
<td>...</td>
<td>...</td>
<td>...</td>
</tr>
<tr>
<td>1998</td>
<td>-</td>
<td>-</td>
<td>13,634 (4.5%)</td>
</tr>
<tr>
<td>1999</td>
<td>-</td>
<td>-</td>
<td>12,895 (4.3%)</td>
</tr>
<tr>
<td>2000</td>
<td>-</td>
<td>-</td>
<td>13,365 (4.4%)</td>
</tr>
<tr>
<td>2001</td>
<td>-</td>
<td>-</td>
<td>13,565 (4.5%)</td>
</tr>
<tr>
<td>2002</td>
<td>-</td>
<td>-</td>
<td>12,509 (4.2%)</td>
</tr>
<tr>
<td>2003</td>
<td>-</td>
<td>-</td>
<td>12,239 (4.1%)</td>
</tr>
<tr>
<td>2004</td>
<td>-</td>
<td>-</td>
<td>24,313 (8.1%)</td>
</tr>
<tr>
<td>2005</td>
<td>-</td>
<td>-</td>
<td>24,321 (8.1%)</td>
</tr>
<tr>
<td>2006</td>
<td>-</td>
<td>-</td>
<td>19,188 (6.4%)</td>
</tr>
<tr>
<td>2007</td>
<td>2,003 (19.0%)</td>
<td>-</td>
<td>18,535 (6.2%)</td>
</tr>
<tr>
<td>2008</td>
<td>1,134 (10.8%)</td>
<td>91,524 (48.9%)</td>
<td>17,185 (5.7%)</td>
</tr>
<tr>
<td>2009</td>
<td>-</td>
<td>-</td>
<td>21,732 (7.2%)</td>
</tr>
<tr>
<td>2010</td>
<td>1,239 (11.8%)</td>
<td>-</td>
<td>21,878 (7.3%)</td>
</tr>
<tr>
<td>2011</td>
<td>-</td>
<td>-</td>
<td>24,724 (8.2%)</td>
</tr>
<tr>
<td>2012</td>
<td>-</td>
<td>95,561 (51.1%)</td>
<td>25,922 (8.6%)</td>
</tr>
<tr>
<td>2013</td>
<td>-</td>
<td>-</td>
<td>24,839 (8.3%)</td>
</tr>
</tbody>
</table>

*NHIS 4 is a lot of trouble or more instead of moderate or more.

Note. NHANES = National Health and Nutrition Examination Survey; NHIS = National Health Interview Survey; NSCH = National Survey of Children’s Health
the suggested method and without 18 and 19 year olds, which demonstrated large differences in the estimated prevalence. Since this is likely due to the sampling design, we, therefore, followed the recommendations on age groups.

NHANES is a unique data set because results were collected using audiometry examinations and parent/self-report. The audiometry examination measured hearing loss based on an examination by a trained professional. Pure tone averages (PTA) were calculated using the decibel level the child was able to detect averaged over 500Hz, 1,000Hz, and 2,000Hz. As per American Speech-Language-Hearing Association recommendations (Clark, 1981), slight hearing loss was defined as bilateral or unilateral PTA ≥ 16 dB, mild loss as PTA ≥ 25 dB, and moderate loss as PTA ≥ 40 dB. There were 12,410 children between the ages 6 and 19 in the data set. After excluding individuals with missing data on audiometry measures (n = 1,868), 10,542 children remained in the audiometry analyses.

The parent/self-report measure was collected during an interview with the parent and/or child. As noted in the documentation for the NHANES (National Health and Nutrition Examination Survey, 2016) participants under 16 years of age, unless there was no one living in the household who was older than 16, were interviewed via a proxy (generally the participant’s parent or guardian); otherwise children reported for themselves. There were no significant differences in responses by parent or child report from what could be ascertained from this guideline.

Since the question asked in the 1994 release of NHANES in the interview differed significantly from those asked from 2006–2010, only those from 2006–2010 were used for the analyses based on parent/self report. After removing any individuals with missing data (n = 1), n = 4,726 children were included in the analyses. To assess hearing loss, the interviewer asked: “Which statement best describes [the child’s] hearing (without a hearing aid)? Would you say [his/her] hearing is excellent, good, that [the child] has a little trouble, moderate trouble, a lot of trouble, or is [the child] deaf?” Hearing loss was defined as moderate trouble, a lot of trouble or deaf. These definitions were used because they most closely resembled that of the other surveys, both in theoretical meaning and in overall prevalence. This question closely follows both the NHANES and the NHIS interview questions, making for fairly simple comparisons between the three parent/self-report measures. After removing individuals with missing data on hearing loss (n = 15) or sex (n = 219), n = 187,085 children remained in the NSCH data set for analyses.

National Health Interview Survey (NHIS). Sixteen releases of the NHIS data (1998–2013) that contained data on childhood hearing loss for ages 0–17 were used for the analyses reported in this article. An adult in the home answered the interview questions. Between 1998 and 2007, the interview specifically asked: “Which statement best describes the child’s hearing without a hearing aid: good, a little trouble, a lot of trouble, or deaf?” From 2008 to 2013, the question is identical but additional options are included, namely excellent and moderate trouble. Due to these additions, we cannot combine the two versions without introducing a spurious trend due to changes in the response options. Thus, the data for 1998–2007 (referred to as NHIS 4) are reported separately from the data for 2008–2013 (referred to as NHIS 6) with the number referring to the amount of options available. For NHIS 4, hearing loss was defined in these analyses as a lot of trouble or deaf. For NHIS 6, loss was defined as moderate trouble, a lot of trouble, or deaf. These definitions were used because they most closely resembled that of the other surveys, both in theoretical meaning and in overall prevalence.

Note that the NHANES parent/self-report measure question is nearly identical to that of the parent report measure in NHIS 6 data (both in the question and the options available) and only differs from NHIS 4 by the number of hearing loss options. After removing individuals with missing data on hearing loss (n = 357), n = 300,844 children from the NHIS data set remained for the analyses (NHIS 4, n = 164,564; NHIS 6, n = 136,280).

Data Analysis

Results of descriptive statistics for each survey are shown in Table 1, including counts on hearing loss (whether examined audiometrically or parent/self-report), age, sex, race, and year of data collection. However, these descriptive statistics do not take into account the non-random sampling and the weighting that can be used to make the estimates nationally representative. Nonetheless, these descriptive statistics do provide information that is useful in understanding some of the factors that may be contributing to differences among the results of the surveys.

1The missing data in the race variable was produced as an unknown category in the analyses. This resulted in an unknown race of n = 7,960 children.
To address the question of the temporal trend in childhood hearing loss by data set, two strategies were used. First, model-based parameters were estimated. Second, prevalence by year was plotted. The first used seven design-based logistic regressions (Lumley, 2010), four for NHANES (PTA ≥ 16 dB, PTA ≥ 25 dB, PTA ≥ 40 dB, and parent/self-report), one for NSCH (parent/self-report), one for NHIS 4 (parent report), and one for NHIS 6 (parent report). The basic model is shown in the following equation, \(2\) where \(i\) is the \(i\)th individual, \(\text{Prob}(Y_{i} = 1)\) is the probability that the \(i\)th individual has hearing loss as measured by either audiologic examination or parent/self-report:

\[
\text{Logit} \left( \frac{\text{Prob}(Y_{i} = 1)}{1 - \text{Prob}(Y_{i} = 1)} \right) = \beta_0 + \beta_1 \text{Year}_i + \beta_2 \text{Sex}_i + \beta_3 \text{Race}_i + \beta_4 \text{Age}_i + \varepsilon_i
\]

The estimated \(\beta\)'s were then transformed to odds ratios via a simple exponentiation to make the interpretation of the model more straightforward. As odds ratios, the resulting interpretation of the year variable (i.e., the estimated trend in childhood hearing loss) becomes the change in the odds of childhood hearing loss given a one-year increase controlling for sex, race, and age. For example, an odds ratio greater than 1 means the odds of hearing loss is increasing over time; an odds ratio less than 1 suggests a decrease in the odds of any given child having hearing loss over time.

Additionally, prevalence by year was displayed graphically as depicted in Figure 1 to show the overall pattern across time for each of the three surveys. This shows the variability within each survey and the agreement among the surveys with regard to the trend in childhood hearing loss in addition to the parametric modeling.

### Results

In Table 1, unadjusted proportions are shown for both the parent/self-report measures and for the audiometry examination. These vary between 0.5–1.1%. However, these proportions do not account for the complex survey design (i.e., the clustering and non-random sampling of specific demographics) and are therefore not representative of the United States population. Each survey has similar demographics, although both NSCH and NHIS have high proportions of white children participating in the survey whereas NHANES is similar across the included race categories.

The results of the seven logistic regressions are shown in Table 2. The NHANES audiometrically measured estimate of the prevalence of hearing loss at a PTA ≥ 16 dB demonstrated a statistically significant increasing trend (OR = 1.022, \(p = .035\)). Similarly, at PTA ≥ 25 dB the odds are increasing over time although it is not statistically significant (\(p = .218\)). Hearing loss measured by NHANES audiometric data at PTA ≥ 40 dB showed decreasing prevalence estimates across time, although this is not statistically significant (\(p = .590\)). Parent/self-reported hearing loss in NHANES showed a statistically significant decreasing trend (OR = 0.772, \(p = .002\)). The parent-report in NSCH leans negative but is not statistically significant (\(p = .827\)). NHIS 4 had a statistically significant downward trend at 7.3% per year (\(p < .001\)). NHIS 6 showed a positive trend with the odds of hearing loss in children increasing 7.1% per year, although this is not statistically significant at the .05 level (\(p = .113\)).
Table 2: The Results of the Modeling of Hearing Loss (or Hearing Threshold) on the Year of the Survey (i.e., the Estimated Trend), the Sex, Race/Ethnicity and the Age of the Child.

<table>
<thead>
<tr>
<th>Variable</th>
<th>PTA ≥ 16 dB</th>
<th>PTA ≥ 25 dB</th>
<th>PTA ≥ 40 dB</th>
<th>Parent Report</th>
<th>Parent Reporta</th>
<th>Parent Reportb</th>
</tr>
</thead>
<tbody>
<tr>
<td>Year</td>
<td>1.022*</td>
<td>1.02</td>
<td>0.989</td>
<td>0.772**</td>
<td>0.994</td>
<td>0.927***</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>1.071</td>
<td>1.029</td>
</tr>
<tr>
<td>Covariates</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Sex</td>
<td>0.958</td>
<td>1.062</td>
<td>1.241</td>
<td>1.281</td>
<td>0.650*</td>
<td>0.665***</td>
</tr>
<tr>
<td>Female</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>1.029</td>
<td></td>
</tr>
<tr>
<td>Race/Ethnicity (White)c</td>
<td>0.959</td>
<td>1.124</td>
<td>1.025</td>
<td>0.797</td>
<td>1.111</td>
<td>1.007</td>
</tr>
<tr>
<td>Black</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>1.23</td>
<td>0.611</td>
</tr>
<tr>
<td>Mexican American</td>
<td>1.084</td>
<td>0.84</td>
<td>1.172</td>
<td>0.423*</td>
<td>1.117</td>
<td>-</td>
</tr>
<tr>
<td>Other</td>
<td>0.973</td>
<td>1.121</td>
<td>1.141</td>
<td>0.214*</td>
<td>1.106</td>
<td>0.611</td>
</tr>
<tr>
<td>Unknown</td>
<td>-</td>
<td>-</td>
<td>-</td>
<td>-</td>
<td>-</td>
<td>0.906</td>
</tr>
<tr>
<td>Age</td>
<td>0.988</td>
<td>0.987</td>
<td>1.028</td>
<td>1.089</td>
<td>1.062***</td>
<td>1.044***</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>1.028</td>
<td></td>
</tr>
<tr>
<td>N</td>
<td>8,812</td>
<td>8,812</td>
<td>8,812</td>
<td>3,577</td>
<td>187,085</td>
<td>164,564</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>136,280</td>
<td></td>
</tr>
</tbody>
</table>

* significant at 0.05 level, ** significant at the 0.01 level, *** significant at the 0.001 level.

Note. NHANES has ages 12–19, NSCH has ages 0–17, and NHIS has ages 0–17. Examination and parent reported measures were modeled using a Generalized Linear Model with a logit link and a binomial distribution (i.e., logistic regression). The results are reported in odds ratios. The effects are adjusted for the complex survey design. All parent-reported rates are at Moderate or more loss except where noted. NHANES = National Health and Nutrition Examination Survey; NHIS = National Health Interview Survey; NSCH = National Survey of Children’s Health; PTA = pure tone averages.

aParent Report between 1998–2007 which only had 4 categories: good, a little trouble, a lot of trouble, deaf. Loss was defined at a lot of trouble or deaf.
bParent Report between 2008–2013 which had 6 categories: excellent, good, a little trouble, moderate trouble, a lot of trouble, deaf. Loss was defined at moderate trouble, a lot of trouble, or deaf.
cNHIS did not have a Mexican American category.

The conflicting results for whether the prevalence of childhood hearing loss is increasing or decreasing are shown graphically in Figure 1. Not only do the prevalence estimates vary substantially across time between surveys, there is also a great deal of variation within some of the surveys across years. Even though the prevalence estimates are within the same general range with the lowest at about 2 per 1,000 and the highest at about 16 per 1,000, it is important to note that this is an eight-fold difference in prevalence. (Note that the prevalence and the 95% confidence interval for each survey at each point in time are shown in Table 3 for reference on the precise values.)

The vertical error bars for each point in Figure 1 show the 95% confidence interval around each estimate of prevalence. These bars emphasize the differences between the prevalence estimates. For example, in 2007, there is no overlap between NSCH and the NHIS error bars suggesting very different estimates of prevalence. Additionally, there is no overlap in the error bars for the 2008 estimates of prevalence based on the NHANES and the NHIS parent/self report even though the parent/self-report questions are essentially identical for both surveys (see Methods section). In 2006, NHANES audiometry and NHANES parent/self report are very different, even though both are at moderate or greater levels of loss, with parent/self report at 15.7 and audiometry at 9.2 per 1,000 children.

Consistent with the data from the logistic regression models in Table 2, it is also clear from Figure 1 that the temporal trends among the surveys do not agree either in direction or magnitude. The NHANES measures show a noteworthy drop from 2008 to 2010 while NHIS 6 has a generally upward trend. NSCH holds relatively steady during the time that NHIS increases and NHANES drops. These varying results could have been affected by the relatively low number of children with hearing loss in the NHANES sample where there were only 95 children with hearing loss at PTA ≥ 40 dB summed across the four years available for the audiometry measure in NHANES. Similarly, only 54 children had hearing loss according to the parent/self-report in NHANES across the three time points.
Table 3: Prevalence per 1,000 by data set and year as shown in Figure 1 for reference.

<table>
<thead>
<tr>
<th>Year</th>
<th>NHANES Audiometry</th>
<th>NHANES PS</th>
<th>NHIS4</th>
<th>NHIS6</th>
<th>NSCH</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Prevalence</td>
<td>95% CI</td>
<td>Prevalence</td>
<td>95% CI</td>
<td>Prevalence</td>
</tr>
<tr>
<td>1994</td>
<td>7.184</td>
<td>(4.98,9.39)</td>
<td>4.104</td>
<td>(2.90,5.31)</td>
<td></td>
</tr>
<tr>
<td>1995</td>
<td>9.198</td>
<td>(2.59,15.8)</td>
<td>4.751</td>
<td>(2.56,6.30)</td>
<td></td>
</tr>
<tr>
<td>1996</td>
<td>10.463</td>
<td>(2.80,16.0)</td>
<td>4.259</td>
<td>(2.91,5.61)</td>
<td></td>
</tr>
<tr>
<td>1997</td>
<td>2.893</td>
<td>(0.00,5.90)</td>
<td>3.762</td>
<td>(2.64,4.88)</td>
<td></td>
</tr>
<tr>
<td>1998</td>
<td>2.855</td>
<td>(2.55,5.16)</td>
<td>4.196</td>
<td>(2.82,5.58)</td>
<td></td>
</tr>
<tr>
<td>1999</td>
<td>5.174</td>
<td>(2.75,7.59)</td>
<td>2.226</td>
<td>(1.20,3.25)</td>
<td></td>
</tr>
<tr>
<td>2000</td>
<td>9.425</td>
<td>(2.71,6.14)</td>
<td>2.578</td>
<td>(0.96,4.19)</td>
<td></td>
</tr>
<tr>
<td>2001</td>
<td>15.747</td>
<td>(11.28,20.21)</td>
<td>4.060</td>
<td>(2.90,5.31)</td>
<td></td>
</tr>
<tr>
<td>2002</td>
<td>15.976</td>
<td>(11.28,20.21)</td>
<td>4.425</td>
<td>(2.71,6.14)</td>
<td></td>
</tr>
<tr>
<td>2003</td>
<td>3.759</td>
<td>(2.90,5.31)</td>
<td>6.339</td>
<td>(3.60,9.07)</td>
<td></td>
</tr>
<tr>
<td>2004</td>
<td>2.140</td>
<td>(1.16,3.12)</td>
<td>7.149</td>
<td>(4.78,9.52)</td>
<td></td>
</tr>
<tr>
<td>2005</td>
<td>5.818</td>
<td>(4.46,7.18)</td>
<td>5.103</td>
<td>(3.10,7.11)</td>
<td></td>
</tr>
<tr>
<td>2006</td>
<td>5.572</td>
<td>(4.65,6.50)</td>
<td>5.841</td>
<td>(3.60,8.08)</td>
<td></td>
</tr>
<tr>
<td>2007</td>
<td>8.091</td>
<td>(5.14,11.04)</td>
<td>95% CI</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>


Discussion

The primary question addressed by these analyses was whether estimates from the different surveys had similar estimates of prevalence for childhood hearing loss and the direction and magnitude of the temporal trend. Although all of the estimates are in a range of 2–16 children per 1,000 from 1994 to 2013, there are noteworthy differences within this range. For example, estimates ranged between 4.4 per 1,000 to 16.0 per 1,000 in 2008 alone with essentially identical questions. Based on these estimates, there would be somewhere between 326,040 children and 1,185,600 children with hearing loss in the United States in 2010. The resources needed to provide diagnostic and habilitation services for 326,040 children are very different than what would be needed for 1,185,600 children. Such a wide range in estimates indicates that funders, administrators, and policy makers do not have the precise information they need to make decisions.

Information about childhood hearing loss are similarly problematic with the trend. Similar to the estimates of prevalence, the estimates of the trend vary greatly between surveys. For example, audiometry measures at PTA ≥ 40 dB and the parent/self-report measures in the NHANES data suggested a decreasing trend of hearing loss, while PTA ≥16 dB and ≥ 25 dB in NHANES and the NHIS 6 (parent report) suggested an increasing prevalence across time (although only PTA ≥16 dB was increasing at a statistically significant level). Further, the parent-report in NSCH showed no change in the prevalence across time. This high degree of variability shown in Figure 1 is striking and has important implications for administrative, policy, and resource allocation decisions.

Considering data from all three surveys at the same time raises fundamental questions about the accuracy of prevalence and trend data from these surveys. The results suggest that there must be some aspect of the measures that are not reliable. For a start, no well-documented research has addressed whether parent/self-report measures of childhood hearing loss are aligned with audiometry measures. Future research should address this important question. Additionally, for the parent/self-report measures, the phrasing is likely important. Although giving the parent the freedom to rate their child’s hearing loss may seem advantageous, it appears that such a rating may not be reliable. Research needs to examine whether the way in which questions are worded affects the accuracy of parent/self-report.

A second research question was whether there was agreement between the surveys about the trend in the prevalence of hearing loss in the United States. In the report by Shargorodsky et al. (2010), the trend appeared to be steady and consistent. However, subsequent data from NHANES results in a less clear answer. Instead of a steady increase, there appears to be a sizable increase and then an even larger decrease in prevalence thereafter. This is especially true at PTA ≥ 40 dB, but a similar pattern is also found at PTA ≥ 16 and PTA ≥ 25 dB. In light of those next data points, and the results from the other surveys, there is no clear answer from federally funded surveys about whether the prevalence of childhood hearing loss is increasing or decreasing.

3There were an estimated 74.1 million children in the United States in 2010 (America’s Children: Key National Indicators of Well-Being, 2016).
Limitations
In interpreting the results of this study, it should be noted that each of these surveys was designed for slightly different purposes. Whereas the NHANES and NHIS are designed for researching both adults and children, the NSCH is designed specifically for children. This may explain some of the more stable estimates for children. The analyses were reliant on the surveys’ designs and weighting information. Factors such as missing data could obstruct the resulting weighting scheme from maximally being nationally representative. However, none of the surveys had a high rate of missing values in any of the variables of interest.

Each survey, in an attempt to accommodate the needs of the country’s health research, occasionally changed which questions were used or how responses about hearing loss were worded. This limited some of the analyses to specific years (e.g., NHANES parent/self-report from 1994 could not be combined with 2006–2010 and NHIS data from 1998–2007 could not be combined with 2008–2013).

Finally, the results bring into question the use of the data for prevalence and trend analyses and measurement (especially parent report) in regards to childhood hearing loss. The results do not indicate whether a similar pattern would be found for other health factors. Further, the results do not indicate that the data cannot be used for other purposes (e.g., testing relationships among the data without reference to being nationally representative).

Conclusions
The NHANES, NSCH, and NHIS data sets are arguably the best data available about children’s health in the United States. They are widely respected because of the systematic and state-of-the-art way in which information is collected, and data from each of these surveys have been used frequently to make important policy and administrative decisions. Given this, it is troubling how much disagreement there is among these three data sets about the prevalence and trends of childhood hearing loss in the United States. While all of the surveys suggest that childhood hearing loss is a substantial problem, affecting somewhere between 2 to 16 children per 1,000 over the last two decades, these large, federally-funded surveys do not provide good enough data to be confident about estimates of either prevalence or trend. Thus, until additional research is done to explain why there is so much disagreement within and between the data sets, we only have a rough estimate of the prevalence of childhood hearing loss in the United States and we do not know whether the trend is increasing or decreasing.

References


Time Trend and Factors Associated with Late Enrollment in Early Intervention among Children with Permanent Hearing Loss in Louisiana 2008-2013

Tri Tran, MD, MPH1,2
Yao Wang, MPH3
Mary Jo Smith, B.S.2
Brenda Sharp, MA4
Terri Ibieta, M.Ed2
Jeanette Webb, M.Ed2
Wendy Jumonville, AUD, MS, CCCA2
Melinda Peat, MCD2
Susan Berry, MD, MPH1,2

1Louisiana State University Health Sciences Center School of Medicine, Department of Pediatrics
2Louisiana Department of Health and Hospitals (DHH) Office of Public Health, Children with Special Health Services Program
3Tulane School of Public Health and Tropical Medicine, Department of Epidemiology
4Our Lady of the Lake Regional Medical Center in Baton Rouge, Louisiana

Abstract
This study aimed to identify the time trend of and factors associated with late enrollment in early intervention (EI) services among children with permanent hearing loss (HL) born between 2008 and 2013 in Louisiana. 2008-2013 linked Louisiana Early Hearing Detection and Intervention, birth records, EarlySteps (IDEA, Part C), Parent-Pupil Education Program, and Medicaid data were analyzed. Logistic regression models were used to evaluate the trend and associations of mother and child's demographic and hearing loss characteristics with late EI enrollment. Results of data analyses did not show any trend of late enrollment in EI services from 2008 to 2013. Delayed diagnosis and mild or unilateral HL were strongly associated with late enrollment. Appropriate strategies to resolve problems relating to missed diagnosis during newborn hearing screening and to convince parents of children with HL to enroll soon after diagnosis of HL will contribute to success of early EI enrollment in the state.

Key Words: Early Hearing Detection and Intervention, newborn hearing screening, early intervention, hearing loss

Acronyms: EI = Early Intervention, HL = Hearing Loss, IDEA = Individuals with Disabilities Education Act, JCIH = Joint Committee on Infant Hearing, D/HH = Deaf and Hard of Hearing, CDC = Centers for Disease Control and Prevention, EHDI – IS = Early Hearing Detection and Intervention – Information System, PPEP = Parent – Pupil Education Program, DSHPSHWA = Directors of Speech and Hearing Programs in State Health and Welfare Agencies, CI = Confidence Interval, UHL = Unilateral Hearing Loss, MBHL = Mild Bilateral Hearing Loss, NHS = Newborn Hearing Screening

Disclosures
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Corresponding author’s contact information: Tri Tran, Email: tri.tran@la.gov or ttran@lsuhsc.edu
Introduction

It is known that hearing loss is strongly associated with delayed development of speech, language, and cognition in early childhood (Holt & Svirskey 2008; Kennedy et al., 2006; Moeller, 2000; Nicholas & Geers, 2006). Previous researchers have suggested the significant value of receiving early intervention services before six months of age for improved academic achievement as well as language and social-emotional development among children with permanent hearing loss (Pipp-Siegel, Sedey, VanLeeuwen, & Yosinahga-Itano, 2003; Yosinahga-Itano, 2003, 2004; Yosinahga-Itano, Coulter, & Thomson, 2000). Although the Joint Committee on Infant Hearing (JCIH) recommends that all newborns diagnosed with hearing loss receive early intervention services no later than six months of age (JCIH, 2007), many deaf and hard of hearing (DHH) children still do not enroll or enroll late in early intervention programs in the United States. Based on 2013 National Centers for Disease Control and Prevention’s (CDC) Early Hearing Detection and Intervention (EHDI) Summary Data Report, only 63.9% of children diagnosed with permanent hearing loss are enrolled in early intervention programs. Of children who enrolled in early intervention programs, 68.8% enrolled before six months of age (CDC, 2013).

Recent studies have identified risks factors related to discrepancies in early intervention enrollment timing and/or service provision among DHH children such as rural residential area (Bush, Burton, Loan, & Shinn, 2013), low socioeconomic status (Boss, Niparko, Gaskin, & Levinson, 2011), a shortage of healthcare insurance (Sommers, 2005), missed newborn hearing screening, lack of parent and primary care provider education on the importance of early intervention (Lester, Dawson, Gantz, & Hansen, 2011), lack of family involvement (Harrison et al., 2016), and late age at diagnosis of hearing loss (Alyami, Soer, Swanepool, & Pottas, 2016; Walker et al., 2014).

Although the Louisiana (LA) EHDI Program has seen an increase in the number of children reported with hearing loss since 2002 when universal newborn hearing screening began, enrollment in early intervention services among children with hearing loss has never been evaluated. Using Louisiana Newborn Hearing Screening, birth records, EarlySteps (IDEA, Part C), Parent-Pupil Education Program, and Medicaid data, we aimed to identify the time trend of late enrollment in early intervention services and associated factors among children ages 0–3 years with permanent hearing loss born between 2008 and 2013. Research factors included mother and child’s demographic and geographic characteristics, time of diagnosis of hearing loss, and characteristics of hearing loss (i.e., type, degree, and laterality). The findings of the study may facilitate improvements in EHDI program implementation and policy making to ensure all affected children have equal access to and benefit from the early intervention services in Louisiana and other states.

Method

Study Population, Data Sources and Linkages

The study included children ages 0–3 years who were born in Louisiana between 2008 and 2013 and were diagnosed with permanent hearing loss. The following children were excluded from the study: children whose mothers were not Louisiana residents at birth; who moved out of state after birth; or who died after diagnosis of hearing loss regardless of receiving any early intervention services.

Four datasets were used for data analyses including birth certificates, LA EHDI-Information System (IS), EarlySteps (i.e., the state’s IDEA, Part C early intervention program), and Medicaid. LA EHDI-IS consisted of newborn hearing screening, diagnosis, and audiological and early intervention data. Only records of children diagnosed with permanent hearing loss were selected and used for data linkages and analyses. The early intervention data in LA EHDI-IS were provided directly from the LA Parent-Pupil Education Program (PPEP), a statewide outreach program provided by the Louisiana School for the Deaf at no cost to families with children ages 0–3 who are deaf or hard of hearing. The LA EHDI Tracking Specialist received data from the PPEP and entered it into the LA EHDI-IS monthly. Louisiana Bureau of Health Statistics and Vital Records provided birth certificate data. Medicaid data included only records with Current Procedural Terminology (CPT) codes of 92507 and 92508 (treatment of speech, language, voice, communication, and/or auditory processing disorder; 92507 for individual, and 92508 for group). The project was deemed exempt by Louisiana State University Institutional Review Board because it did not meet the federal definition of human subjects research.

SAS 9.4 and LinkPro 3.0 were used for data linkages. First, LA EHDI-IS data including only children with hearing loss were linked to birth certificates. Only records matched with birth records were kept and used in the next linkage (552 matched records in total 559 records with hearing loss). Second, matched LA EHDI-IS and birth data were linked to EarlySteps data; and last, matched LA EHDI-IS, birth, and EarlySteps data were linked to Medicaid data. The linking variables included child’s date of birth, first name, and last name with soundex codes (i.e., codes of names based on the phonetic spelling of the name). In each stage of linkages, linked records were reviewed manually to define true matches using linking variables and some of the following variables when available: mother’s last name, first name, maiden name; address of residence at birth or most updated address of residence; and birthing hospital. Of 552 records of children with hearing loss matched with birth certificates, 351 (63.5%) records contained PPEP data, 412 matched with EarlySteps data (74.5%), and 240 (43.5%) matched with Medicaid data. Thus, EarlySteps contributed the most data of documented enrollment in EI in this study. A total of 492 (89.1%) records of children in the final matched data were included in PPEP, and/or EarlySteps, and/or Medicaid data. Those children were defined as enrolled in early intervention programs and used for data analysis.
**Analysis Variables-Outcome Variables.**

**Enrollment in early intervention (EI)**
As mentioned above, only children found in PPEP, EarlySteps, or Medicaid data were defined as enrolled in early intervention programs. Children who enrolled in intervention programs may have received services (i.e., PPEP, EarlySteps, or Medicaid) or were monitored by audiologists (PPEP). Intervention services included any type of habilitative, rehabilitative, or educational service provided to children with hearing loss (JCIH, 2007).

**Late/early enrollment in early intervention**
Of those who enrolled in early intervention programs, children who began services or were monitored before six months of age were classified as enrolled early in early intervention; otherwise they were classified as enrolled late. The earliest date of enrollment in the three programs was used to estimate the time of enrollment.

**Independent variables**
Factors used to evaluate associations with late enrollment in EI included mother’s child’s demographic and geographic characteristics, time of diagnosis of hearing loss, and characteristics of hearing loss (i.e., type, severity degree, and laterality).

All demographic and geographic variables were derived from birth certificate data and defined as categorical variables. They included birth weight (i.e., low birth weight, < 2,500 grams vs. normal weight, > 2,500 grams), race (i.e., white, black, and other), ethnicity (i.e., Hispanic vs. non-Hispanic), geographic area of residence (i.e., urban vs. rural), maternal age (i.e., < 20, 20-34, and 35+ years old), maternal education (i.e., not completed, completed high school, and completed some college), number previous live births (i.e., none, one, and two or more), and sex (i.e., male vs. female).

Hearing loss (HL) was classified into different levels of severity, types, and laterality. The Directors of Speech and Hearing Programs in State Health and Welfare Agencies system for degree of hearing loss was used to categorize severity of hearing loss as follows: mild (21–40 decibels hearing level [dBHL]), moderate (41–40 dBHL), severe (71–90 dBHL), and profound (> 91 dBHL; Curry & Gaffney, 2010). For bilateral HL, the ear with more severity was used to categorize severity degree. Laterality of hearing was categorized as unilateral versus bilateral. Four types of hearing loss were defined as sensorineural, conductive, mixed, and auditory neuropathy/dysynchrony. Age at diagnosis of HL was calculated using date of birth and date when hearing loss was diagnosed and confirmed by an audiologist, and categorized as 0–2, 3–5, and 6+ months of age.

**Data analysis**
Rate of late enrollment in EI was calculated using the following formula: Number of children with hearing loss who enrolled in EI at six months of age or older/total children with hearing loss who enrolled in EI)*100. Trend of late enrollment in EI was analyzed from the 2008 to 2013 birth years. Both unadjusted and adjusted annual percent change of odds of late enrollment was estimated by using logistic regression models. Birth year was treated as a continuous variable when estimating the trend of late enrollment in regression models. Multiple regression models used to estimate adjusted annual percent change of odds of late enrollment included birth year and all study factor variables.

To identify associations of independent variables with late enrollment in EI, only data including children with hearing loss diagnosed before six months of age were analyzed (267 of total 492 children defined as enrolled in EI). Logistic regression models were used to analyze data, and adjusted models included all independent variables. All final models included only variables with p value < 0.05. Data analyses were conducted in SAS 9.4.

**Results**

**Study population description**
The study included 492 children ages 0–3 years old who were born between 2008 and 2013 in Louisiana, were diagnosed with permanent hearing loss, and enrolled in EI. Approximately 54% of children were white, 96% non-Hispanic, 55% male, and 27% low birth weight (< 2,500 grams). The earliest date of enrollment in the three programs was used to estimate the time of enrollment.

**Table 1: Late Enrollment in Early Intervention (%) among Children with Hearing Loss Born between 2008 and 2013, Louisiana (N = 492)**

<table>
<thead>
<tr>
<th>Demographic and hearing loss characteristics</th>
<th>Percent (C95%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Total</td>
<td>48.8, 44.4 – 53.2</td>
</tr>
<tr>
<td>Race</td>
<td></td>
</tr>
<tr>
<td>White</td>
<td>43.3, 37.1 – 49.4</td>
</tr>
<tr>
<td>Black</td>
<td>55.7, 47.4 – 62.8</td>
</tr>
<tr>
<td>Other</td>
<td>50.0, 30.0 – 70.0</td>
</tr>
<tr>
<td>Ethnicity</td>
<td></td>
</tr>
<tr>
<td>Non-Hispanic</td>
<td>48.5, 43.9 – 53.2</td>
</tr>
<tr>
<td>Hispanic</td>
<td>52.4, 31.0 – 73.7</td>
</tr>
<tr>
<td>Geographical area at birth</td>
<td></td>
</tr>
<tr>
<td>Rural</td>
<td>48.2, 41.6 – 54.8</td>
</tr>
<tr>
<td>Urban</td>
<td>49.2, 42.9 – 55.4</td>
</tr>
<tr>
<td>Maternal age</td>
<td></td>
</tr>
<tr>
<td>&lt; 20 (11%)</td>
<td>56.9, 43.3 – 70.5</td>
</tr>
<tr>
<td>20-34 (78%)</td>
<td>48.6, 43.5 – 53.8</td>
</tr>
<tr>
<td>35+ (11%)</td>
<td>41.2, 27.7 – 54.7</td>
</tr>
<tr>
<td>Maternal education</td>
<td></td>
</tr>
<tr>
<td>&lt; High school (19%)</td>
<td>57.3, 47.0 – 67.6</td>
</tr>
<tr>
<td>High school (33%)</td>
<td>51.7, 43.7 – 59.6</td>
</tr>
<tr>
<td>&gt; High school (48%)</td>
<td>43.4, 37.0 – 49.9</td>
</tr>
<tr>
<td># Previous live births</td>
<td></td>
</tr>
<tr>
<td>None (41%)</td>
<td>45.1, 38.1 – 52.1</td>
</tr>
<tr>
<td>One (33%)</td>
<td>49.7, 41.8 – 57.6</td>
</tr>
<tr>
<td>Two+ (26%)</td>
<td>53.3, 44.4 – 62.1</td>
</tr>
<tr>
<td>Birth weight</td>
<td></td>
</tr>
<tr>
<td>&lt;2,500 grams (27%)</td>
<td>51.2, 2.4 – 60.0</td>
</tr>
<tr>
<td>2,500 grams (73%)</td>
<td>47.8, 42.5 – 53.1</td>
</tr>
<tr>
<td>Sex</td>
<td></td>
</tr>
<tr>
<td>Male (65%)</td>
<td>51.0, 44.9 – 57.1</td>
</tr>
<tr>
<td>Female (45%)</td>
<td>46.0, 39.2 – 52.7</td>
</tr>
<tr>
<td>Type of HL</td>
<td></td>
</tr>
<tr>
<td>Conductive (6%)</td>
<td>44.4, 25.7 – 63.2</td>
</tr>
<tr>
<td>Sensorineural (81%)</td>
<td>51.3, 46.3 – 56.4</td>
</tr>
<tr>
<td>Mixed (5%)</td>
<td>38.5, 19.8 – 57.2</td>
</tr>
<tr>
<td>Auditory Neuropathy (8%)</td>
<td>32.5, 18.0 – 47.0</td>
</tr>
<tr>
<td>Severity of HL</td>
<td></td>
</tr>
<tr>
<td>Mild (18%)</td>
<td>55.0, 44.1 – 65.9</td>
</tr>
<tr>
<td>Moderate (30%)</td>
<td>44.7, 36.2 – 53.2</td>
</tr>
<tr>
<td>Severe (18%)</td>
<td>53.8, 42.8 – 64.7</td>
</tr>
<tr>
<td>Profound (34%)</td>
<td>50.0, 42.0 – 58.0</td>
</tr>
<tr>
<td>Laterality of HL</td>
<td></td>
</tr>
<tr>
<td>Bilateral (75%)</td>
<td>44.9, 35.9 – 53.9</td>
</tr>
<tr>
<td>Unilateral (25%)</td>
<td>50.1, 45.0 – 55.3</td>
</tr>
<tr>
<td>Age at diagnosis of HL</td>
<td></td>
</tr>
<tr>
<td>0-2 (40%)</td>
<td>21.1, 15.3 – 26.8</td>
</tr>
<tr>
<td>3-5 (15%)</td>
<td>29.0, 18.3 – 39.7</td>
</tr>
<tr>
<td>6+ (45%)</td>
<td>100.0</td>
</tr>
</tbody>
</table>
Enrollment in Early Intervention among Children with Hearing Loss (HL) between 2008 and 2013, Louisiana

Of those who enrolled in EI, 48.8% enrolled late. The rate of late enrollment was fairly stable during the study time period with the rate of 44.8% in 2008 and 45.5% in 2013 (Figure 1). Unadjusted annual percent change of odds of late enrollment was 10.0% (Odds Ratio [OR]: 1.1, Cl: 0.9-1.2, p = 0.1967). Adjusted logistic regression models did not show any trend of late enrollment from 2008 to 2013 (p > .05). Birth year was not statistically significant and excluded from the final model; therefore, the value of the odds ratio was not shown.

One of the main reasons for late enrollment in EI was diagnosis made at six months of age or older. Of children who enrolled late in EI programs (240), 74.5% of them were diagnosed with HL at six months of age or older. Limited to children diagnosed with hearing loss before six months of age (267), the rate of late enrollment was 19.5%. The final adjusted regression model showed odds of late enrollment were statistically higher in children with mild HL (Mild: OR: 12.2, Cl: 3.9-38.6; Moderate: 4.4, Cl: 1.5-12.6; Severe: 5.4, Cl: 1.6-18.0), unilateral HL (OR: 2.5, Cl: 1.1-5.7), or those with HL diagnosed after two months of age (OR: 3.2, Cl: 1.5-7.0). There was no statistically significant association of late enrollment with birth weight, race, ethnicity, geographic area of residence, maternal age, maternal education, number of previous live births, or sex (Table 2).

**Discussion**

Results of data analyses indicated that of those who enrolled in EI, the rate of late enrollment (after six months of age) was 48.8%. The rate of late enrollment was steady and a trend was not found during 2008–2013. One of the main reasons for late enrollment was late diagnosis, made at six months of age or older. It contributed 74.5% of total late enrollment. Among those whose HL were diagnosed before six months of age, children with mild HL had the highest risk of late enrollment. In addition, children with unilateral HL or diagnosis after two months of age were more likely to enroll late.

Based on results of the study, children with mild or unilateral HL were potentially at risk for late enrollment in EI programs. Findings from previous studies indicated that unilateral or mild HL can adversely affect a child’s development. Bess and Tharpe (1984, 1986) found that approximately one-third of children with permanent unilateral HL experienced significant language and academic delays. Madell and Flexer (2008) showed that children with unilateral HL or mild bilateral HL can be at risk for academic, speech-language, and social-emotional difficulties. Tharpe (2008) also found children with mild HL were not performing at expected academic levels. Thus, late EI enrollment can negatively impact developmental outcomes for children with unilateral or mild HL. Some researchers have indicated one of the main reasons leading to late enrollment was difficulty in obtaining EI services, wherein children with unilateral hearing loss and mild bilateral hearing loss were not qualifying for the EI services (Holstrom, Gaffney, Gravel, Oyler, & Ross, 2008; JCIH 2007). However, this reason was not applicable to...
Louisiana where all children with any degree of unilateral or bilateral HL are eligible for both EarlySteps (IDEA, Part C) and PPEP. Parents of children with unilateral or mild HL often declined services and those children often enrolled in EI programs later when developmental delays, specifically language delay, were evidenced. The study data showed that of children diagnosed with HL before six months of age, those with unilateral-mild HL had the highest rate of late enrollment (40.0%), followed by bilateral-mild HL (31.4%), and other laterality-severity HL (<25%). In fact, children with unilateral or mild HL may appear to have “normal” hearing, making it difficult to convince parents of the necessity of enrolling early in early intervention programs (Haggard & Primus, 1999). Thus, it is very important to help parents understand difficulties of hearing for children with unilateral and/or mild HL. To do so, audiologists may educate parents to use hearing loss simulation, via software such as NIOSH Hearing Loss Simulator (CDC, 2002), which is useful to help parents listen to what the hearing loss sounds like, and also to demonstrate the challenges of distance and noise in speech recognition for a hearing loss child.

The findings of this study also indicated that delayed diagnosis as a strong factor related to late EI enrollment. Delayed diagnosis could be caused by no newborn hearing screening (NHS) or missed diagnosis through NHS. The study data indicated 33 (6.0%) children with HL were not screened with NHS. Of those, 5 (4.8%) and 21 children (63.6%) were diagnosed with HL after three months and six months of age, respectively. The data also found that 104 children (18.8%) passed NHS but were diagnosed with HL later. Of those, 9 (8.7%) and 77 (74.0%) were diagnosed with HL after three months and six months of age, correspondingly. Missed diagnosis may be due to some forms of HL (mild, auditory neuropathy, or delayed-onset HL) or quality of NHS services so that HL could not be detected through NHS. Studies by Cone-Wesson and Johnson et al. have indicated that current NHS technologies fail to detect some infants with mild hearing loss (Cone-Wesson et al., 2000; Johnson et al., 2005). Other studies also showed that newborns with auditory neuropathy HL may not be detected through NHS when otoacoustic emission (OAE) method is used alone (D’Agostino & Austin, 2004). In our study, among 104 newborns who passed NHS but had HL later, 35 (33.7%) of those were diagnosed with mild and/or auditory neuropathy HL. Thus, 69 (66.3%) of children with other levels and types of HL were still not detected through NHS. Delayed onset HL or quality of screening services may relate to missed diagnosis among those children. Closely monitoring passed-NHS newborns with risk factors of mild, auditory neuropathy, and delayed onset HL is recommended by the JCIH to capture HL missed through NHS. The JCIH developed a list of risk factors and time frames to monitor children with increased risk of these forms of HL. However, with the current recommended time frames of monitoring for delayed onset HL with an audiological evaluation at least once by 24 to 30 months of age, early detection of this form of HL is challenging. In order to improve missed diagnosis of auditory neuropathy HL, the JCIH recommends using automated auditory brainstem response to screen newborns who require NICU care and who are at high risk for this type of HL (JCIH, 2007). More research is needed regarding problems related to the quality of NHS services that cause missed diagnosis.

The findings from this study were consistent with previous studies which indicated that earlier diagnosis was effective in decreasing the age at entry into EI (Alyami et al., 2016; Harrison, Roush, & Wallace, 2003). Although data analysis in our study showed children with mild HL were inversely related to early enrollment in EI programs, this finding was contrary to earlier studies. For instance, Walker et al. (2014) did not find any association between the severity of HL and age at entry into early intervention. Note that the analysis of Walker et al. was conducted with a very small sample size of only 20 children who enrolled in early intervention following HL confirmation, which may attenuate the power of the statistical tests. Recent studies have found that socioeconomic status is an important effect on enrollment timing of EI (Boss et al., 2011). However, this information was not well captured in the study data. Although Medicaid coverage can be used as a proxy of low family income, and linkage with Medicaid data was conducted in the study, the definition of Medicaid children may be underestimated because Medicaid data did not include children who may have been qualified for Medicaid but only enrolled in EarlySteps and/or PPEP, not Medicaid.

**Strengths and Limitations**

This study had two major strengths. First, the study used three data sources (EarlySteps, PPEP, and Medicaid) that covered nearly all early intervention services in the state. About 90% of total children with hearing loss reported by LA EHDI were found in these data sources. Use of all three data sources improved both quality of LA EHDI program reports and research in EI enrollment. Second, high accuracy of the data linkages was ensured by using multiple identifiers for both child and mother for the linkages and matched case review.

The findings in this study were subject to three limitations. First, the study did not capture data of early intervention services provided through other data sources such as private health insurance. However, with an estimate of 10% of children with HL from those data sources, bias in results of data analyses was not expected. Second, some other factors (study independent variables) that may be significant were excluded from the final adjusted regression model when data analyses were limited to the small sample of children with a HL diagnosis before six months after birth (267). Last, the study did not include newborns who failed the newborn hearing screening and were lost to follow-up (about 32%) meaning their diagnosis of HL and enrollment in EI are unknown. Exclusion of those newborns from the study may affect both data validity and reliability of analyzed results.
Conclusions

Among birth cohorts from 2008 to 2013, about 90% of HL children were found enrolled in EI programs in Louisiana. Of those, approximately 50% enrolled late, and this rate was not seen to improve during the study time period. Efforts targeted on high-risk populations defined in the study may enhance early enrollment in EI services. Delayed diagnosis and mild or unilateral HL were strongly associated with late enrollment. Appropriate strategies to resolve problems relating to missed diagnosis during NHS and to encourage parents of children with HL to enroll soon after diagnosis of HL will contribute to success of early EI enrollment in the state.

References


Home Visiting Programs for Families of Children who are Deaf or Hard of Hearing: A Systematic Review

Nannette Nicholson, Ph.D.,1, 2, 3 Patti Martin, Ph.D.,1, 2, 3, 4 Abby Smith, B.S.,1, 2, 4 Sheila Thomas, M.A. (L.S.), M.Ed.,1 Ahmad A. Alanazi, M.Aud.,1,2,5

1University of Arkansas for Medical Sciences, Little Rock, AR
2University of Arkansas at Little Rock, Little Rock, AR
3Arkansas Children’s Hospital, Little Rock, AR
4National Center for Hearing Assessment and Management, Logan, UT
5King Saud bin Abdulaziz University for Health Sciences, Riyadh, Saudi Arabia

Abstract
Prelingual hearing loss greatly restricts a child’s language development, hindering his or her behavioral, cognitive, and social functioning. Although technology such as hearing aids and cochlear implants provide access to sound, infants and children also need habilitation to develop skills. These skills include learning to listen, or attend, to process language (whether visual or spoken), and to produce language and communicate. Home visiting is widely recognized as a cost-effective intervention service delivery model. Home visiting programs for promoting language development in children who are diagnosed as deaf or hard of hearing have been in existence for over 50 years, yet there is limited evidence of their effectiveness. This review was undertaken to assess the evidence of effectiveness of home visiting in children with prelingual hearing loss. While many studies have examined early intervention for children who are deaf or hard of hearing, few are published from specific home visiting programs meeting the criteria for inclusion in this review. Studies from specific home visiting program models designed to meet the needs of the target population are needed to examine the effectiveness of promoting language development within the context of a home visiting program for children who are deaf or hard of hearing and their families.

Keywords: deaf, hearing loss, hard of hearing, home visiting, home visits, home visitors, home visitation, in-home, family counseling, family health, early intervention, early interventions, language development, literacy development, evidence based, evidence of effectiveness, evidence-based practice, systematic review.

Acronyms: AHQR = Agency of Healthcare Quality and Research, CDI = Child Development Inventory, CHIP = Colorado Home Intervention Program, CHTP = Counseling and Home Training Program, DHHS = Department of Health and Human Services, EAS = Emotional Availability Scales, ECHI = Early Childhood Home Instruction Program, EHDI = Early Hearing Detection and Intervention, EDWPVT4 = Expressive One-Word Picture Vocabulary Test, HomVEE = Home Visiting Evidence of Effectiveness Program, JCIH = Joint Committee on Infant Hearing, MCDI – EV = MacArthur Communication Development Inventory: Expressive Vocabulary, MCDI – RV = MacArthur Communication Development Inventory: Receptive Vocabulary, PICO = participants or population, interventions, comparisons, and outcomes, PRISMA = preferred reporting items for systematic reviews and meta-analyses, Project ASPIRE = Achieving Superior Parental Involvement for Rehabilitative Excellence, SK*HI, TACL4 = Test of Auditory Comprehension of Language

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Acknowledgements: We thank Carol MacEach, MLS, Associate Professor, Research and Scholarly Communications Coordinator, Ottenheimer Library, University of Arkansas at Little Rock, for her assistance with the systematic review. Sincere appreciation is extended to Mary Ellen Nevins, Ed.D., Professor, Department of Audiology and Speech Pathology, University of Arkansas for Medical Sciences, for her review of the initial drafts of this manuscript. In addition, the authors extend their appreciation to Kelsey Kitchens, B.S., for her assistance with the references.

Corresponding author’s contact information: Nannette Nicholson, Ph.D.; UALR Speech and Hearing Clinic; 2801 S. University Ave., Suite 600 UP, Little Rock, AR 72204; 501.569.8909; nn@uams.edu
Introduction

Background
Use of evidence-based practices in health and education as the basis for quality improvement and accountability are at the forefront of federal policy in the United States. Without early intervention, children with congenital or early childhood hearing loss, are at risk for social, emotional, cognitive, and other developmental delays impacting language, literacy, learning, and overall academic performance (Joint Committee on Infant Hearing [JCIH], 2007; JCIH, 2013; Moeller, Tomblin, Yoshinaga-Itano, Connor, & Jerger, 2007; Yoshinaga-Itano & Apuzzo, 1998). Estimates of the lifetime educational costs for prelingual hearing loss are very high (Keren, Helfand, Homer, McPhillips, & Lieu, 2002; Schroeder, 2006); thus even modest models estimate a cost/benefit ratio for newborn hearing screening programs of more than 25:1 (Gorga & Neely, 2003; Porter, Neely, & Gorga, 2009). The World Health Organization (2010) identifies cost-effective newborn and early childhood screening programs as the first step in the process leading to diagnosis and treatment.

Early Hearing Detection and Intervention (EHDI) Initiative
Implicit in the terminology used to describe the EHDI initiative promoted by JCIH is the notion that hearing screening programs are linked to effective diagnostic and treatment programs (White, 2016). This continuum of care from early detection to intervention for children who are diagnosed as deaf or hard of hearing is multifaceted and requires a multidisciplinary approach to intervention (JCIH, 2013). The origins of EHDI programs share this multidisciplinary approach to family-centered early intervention programs, recognizing the importance of the family as a system on outcomes of intervention services (JCIH, 2007, 2013; White, 2016). The current challenge of EHDI programs is the implementation of existing evidence-based policies and practices in ways that will enable and empower families by increasing individual family and child capabilities and strengths (White, Forsman, Eichwald, & Munoz, 2010). Home visiting is one of the early intervention options available to families of young children with hearing loss (Adirim & Supplee, 2013; Doggett, 2013; Duggan et al., 2013; Korfmacher, Laszewski, Sparr, & Hammel, 2012; Sacks et al., 2003).

Home Visiting
Home visiting is grounded as an early intervention approach to service delivery in a number of disciplines. It is based on the notion that early intervention makes a difference in child and family outcomes (Division for Early Childhood & National Association for Education of Young Children, 2009). As a result of this philosophy, numerous home visiting programs have been developed including prenatal care, parenting support, child maltreatment prevention, and early intervention for children with disabilities. The origins of home visiting programs can be traced back to three main movements that began in the 1800s: (a) early childhood education, (b) public health nursing, and (c) social advocacy and prevention efforts (Boller, Strong, & Daro, 2010). Evidence-based home visiting programs embrace the concept that family and child outcomes are improved when family-centered principles are embedded within program activities (Bailey, Raspa, Humphreys, & Sam, 2011; Llewelyn, McConnell, Honey, Mayes, & Russo, 2003). Family-centered principles are evidence-based and form the foundation of federal legislation for the Part C (birth to three) services specified in the Individuals with Disabilities Education Act (IDEA, 2004). These guidelines are based on family-centered values and include a shared philosophy of families as partners, a focus on family strengths, family choice of goals and services, collaboration and coordination of services, effective communication, and flexibility (JCIH, 2007, 2013; IDEA, 2004).

The Maternal and Child Health Bureau’s (MCHB) Home Visiting Program builds upon decades of research demonstrating that home visits by nurse, social worker, or early childhood educators during pregnancy and in the first years of life improves child and family outcomes (Adirim & Supplee, 2013). By equipping parents with the skills needed to support the cognitive, socio-emotional, and physical health development of their children, the MCHB Home Visiting Program works with other parts of the early learning initiative to optimize outcomes for children and families and to help each attain their full potential (Adirim & Supplee, 2013). These goals are closely aligned with best practices promoted by JCIH (2007, 2013). The Home Visiting Evidence of Effectiveness Program (HomVEE) was developed by MCHB to conduct rigorous ongoing systematic reviews to evaluate the evidence of effectiveness of home visiting programs with published outcomes demonstrating positive outcomes for children and families (Avellar et al., 2016; Paulsell, Boller, Hallgren, & Esposito, 2010). The results of the systematic reviews conducted annually by HomVEE are published on their website (http://homvee.acf.hhs.gov/).

Over the past few years, HomVEE (2016) has designated 19 named home visiting models as “national models” meeting specific criteria set forth by the U.S. Department of Health and Human Services (DHHS) for their rigorous review process (Avellar et al., 2016). Interestingly, although early intervention home visiting programs for children who are deaf or hard of hearing and their families have existed for decades, none of the national home visiting models designated by HomVEE include those developed specifically for application in EHDI programs (Avellar et al., 2016; Sacks et al., 2003). Therefore, the purpose of this research project was to use the HomVEE research design and inclusion criteria (see Table 1) to identify programs specifically serving children who are deaf or hard of hearing and their families. The research question addressed was: Using the method and criteria employed by HomVEE to evaluate specific home visiting models (i.e., national models), do any home visiting programs specifically serving

1Throughout the remainder of this article, “jurisdiction” will be used to refer to states, territories, and other political jurisdictions that operate screening programs such as Washington, D.C., Puerto Rico, the Virgin Islands, etc.
children who are deaf or hard of hearing meet the DHHS criteria for evidence-based or emerging practice?

**Systematic Review Methodology**

HomVEE employed a systematic review methodology to evaluate the quality and strength of evidence available for named home visiting models which consisted of (a) conducting a broad literature search, (b) screening studies for relevance, (c) critically appraising the studies, (d) comparing the appraisals to predetermined criteria (see methods for this study), and (e) extracting the data to evidence tables. We used the same criteria that HomVEE used for inclusion and exclusion in this study. The quality of each study with an eligible design was rated as high, moderate, or low. The home visiting model was rated as an “evidence-based early childhood home visiting service delivery model” if there was one randomized controlled trial (high quality) or two moderate quality studies with statistically significant findings and demonstrated sustainability over 6 months or more (Avellar et al., 2016, p. 5). If the home visiting model met the criteria without demonstrated sustainability, it was designated as a promising practice.

Similar to HomVEE, the authors used the Cochrane Handbook for Systematic Reviews of Interventions (Higgins & Green, 2011) as a guide for developing the methodology for this project. In accordance with steps outlined in this handbook, the systematic review question specifies the types of population (participants) included in the reviewed studies, types of intervention (and comparisons), and the types of outcomes of interest. The acronym PICO (participants or population, interventions, comparisons, and outcomes) serves as a reminder of these components (Cousine, 1997; O’Connor, Green, & Higgins, 2011). According to these authors, the research question is typically stated as an objective using the PICO framework and includes the question components, which are used to generate search terms and search term strings developed for execution of the systematic review. The components of the question, with the specification of the types of studies included in the review are determined a priori, serving as the basis for the eligibility criteria included in the review.

The target populations of the early intervention home visiting models are children who are deaf or hard of hearing (aged birth to three years or birth to five years) and their parents and/or caregivers. The target intervention is home visiting to promote language, listening, and literacy development for infants and young children who are diagnosed, or at risk for prelingual childhood hearing loss. We limited our study to outcome measures in the child development and school readiness domain, which most closely aligns with the JCIH domains of interest. The outcome measures relevant to the target domain included auditory, speech, language, and literacy developmental assessments and/or assessment tools. HomVEE used a similar process in their evaluation of home visiting models, but included eight domains (Avellar et al., 2016). Outcome domains in the HomVEE review and excluded from our study were: (a) child health; (b) maternal health; (c) reductions in child maltreatment; (d) reductions in juvenile delinquency, family violence, or crime; (e) family economic factors; (f) positive parenting factors; and (g) linkage and referrals.

We used the flow diagram reporting method recommended by Higgins and Green (2011) known as PRISMA, the acronym for Preferred Reporting Items for Systematic Reviews and Meta-Analyses (Liberati et al., 2009; Moher et al., 2009). We used the same criteria that HomVEE used to critically appraise each study. In addition, we used evidence summary tables to present the findings of the study as recommended in the Cochrane Handbook.

**Objectives**

The objective of this systematic review was to assess the effects of home visiting for children who are deaf or hard of hearing and their families in the child development and school readiness domain.

**Method**

This study was submitted to and approved by the Institutional Review Board (IRB) at the University of Arkansas for Medical Sciences with exempt status (Protocol #205394).

**Criteria for Considering Studies for this Review**

Eligible study designs were prospective randomized controlled trials or quasi-experimental studies. Retrospective quasi-experimental research designs were also eligible for inclusion.

**Types of Studies**

Eligible study designs were prospective randomized controlled trials or quasi-experimental studies. Retrospective quasi-experimental research designs were also eligible for inclusion.

**Types of Participants**

Children from birth to five years of age with congenital or early acquired (before age five years of age) deafness. Type, degree, configuration, and laterality of hearing loss were not considerations. Children with known cognitive, social-emotional, or behavioral disorders were not excluded.

**Types of Interventions**

We included specific, named home visiting programs (i.e., national models designed for children who are deaf or hard of hearing and their families). We did not require a minimum period of intervention. We did not expect to find studies using treatment-as-usual control groups, different dose control groups, or adverse effects from intervention.

**Types of Outcome Measures**

Child outcomes were considered primary and parent report measures were considered secondary. Outcome measures included receptive language, expressive language, developmental language, auditory development, pre-literacy language development, listening development, social-emotional development, and other developmental
outcome measures indicative of child development and school readiness. We did not limit inclusion of the study based on the developmental outcome measure. Table 1 summarizes the inclusion and exclusion criteria that were used for considering studies for this systematic review.

Criteria for Rating Studies
We used the HomVEE criteria for rating the quality and impact of studies (HomVEE, 2016). Study rating options included high, moderate, or low.
1. High—random assignment studies with low attrition of sample members and no reassignment of sample members after the original random assignments.
2. Moderate—random assignment studies that, due to flaws in the study design, execution, or analysis, do not meet all the criteria for the high rating; matched comparison group designs that establish baseline equivalence on selected measures; and single case and regression discontinuity designs.
3. Low—other studies that do not meet the criteria for high or moderate.

Criteria for Designation as an Evidence-Based Early Intervention Home Visiting Model or as Promising Practice
To meet the criteria for an evidence-based early childhood home visiting service delivery model, program models must meet at least one of the following criteria (HomVEE, 2016): 1. At least one high- or moderate-quality impact study of the model finds favorable, statistically significant impacts in the outcome domain of child development and school readiness. 2. At least two high- or moderate-quality impact studies of the model using non-overlapping analytic study samples with one or more favorable, statistically significant impacts in the target domain.

Home visiting models with at least one moderate-quality impact analytic study sample with one favorable statistically significant impact that had not yet demonstrated sustainability were designated as promising practices.

Search Methods for Identification of Studies
Electronic Searches
Databases available through the University of Arkansas for Medical Sciences (UAMS) and the University of Arkansas at Little Rock (UALR) searched for this systematic review using the search terms generated from the PICO framework are shown in Appendix A.
Table 2. Search Terms and Filters

<table>
<thead>
<tr>
<th>Concept</th>
<th>Terms</th>
<th>Notes</th>
</tr>
</thead>
<tbody>
<tr>
<td>P = Hearing Loss</td>
<td>All databases: deaf, deafness, &quot;hearing impaired&quot;, &quot;hearing loss&quot;&lt;br&gt;MeSH terms Deafness and Persons With Hearing Impairments also used in PubMed</td>
<td>Combined with OR</td>
</tr>
<tr>
<td>P = Age Groups</td>
<td>Terms used in resources without age filters: preschool OR infant OR infants OR baby OR babies</td>
<td>Age filters used in CINAHL, PsycINFO, and PubMed to limit to birth to 5 years old</td>
</tr>
<tr>
<td>I = Home Visits</td>
<td>All databases: “home visit”, “home visits”, “home visitors”, “home visitation”, “in-home”, “family counseling”&lt;br&gt;MeSH term Family Health also used in PubMed</td>
<td>Combined with OR</td>
</tr>
<tr>
<td>I = Intervention</td>
<td>All databases: “early intervention”, “early interventions”</td>
<td>Combined with OR</td>
</tr>
</tbody>
</table>

*Note. P = Participant or Population search terms (children who are deaf or hard of hearing aged birth to five and their families) or variables; I = Intervention search terms (home visiting intervention) or variables; CINAHL = Cumulative Index of Nursing and Allied Health Literature; MeSH = Medical Subject Headings.*

Search Strategy
Table 2 summarizes the search terms and filters that were used to retrieve relevant items from the databases. Searches were limited to English language items; no publication date limits were used in any database.

Other Searches
In addition to the database search, the literature search for this study included two additional activities:
1. Search results were compared against the bibliographies of recent literature reviews and meta-analyses of home visiting models for children who are deaf or hard of hearing, and relevant missing citations were added to our search results.
2. Google was used to search relevant government, state, university, research, and nonprofit websites for unpublished reports and papers.

Data Collection and Analysis
We identified studies by employing a systematic search strategy in electronic databases, screened identified studies for relevance, compared each study to the eligibility criteria for program models and research design, and summarized data from included publications into evidence tables (see Results and Figure 1). One member of the research team designed and executed the systematic search. Two of the authors screened the titles and abstracts for relevance. Three members of the research team served as reviewers and critically appraised the research design, assessing the evidence for each model. One author summarized the findings in evidence tables. All members of the research team contributed to writing and editing the final manuscript.

Selection of Studies
After removal of duplicates, two review authors independently screened titles and abstracts of studies identified in the searches and selected all potentially relevant studies. The titles and abstracts were reviewed for relevancy. Those deemed irrelevant were eliminated from further consideration. Studies that examined variables not integral to the home intervention (i.e., demographic report), conducted in underdeveloped countries (i.e., some countries in Africa), and unpublished demonstration project reports were excluded. We obtained copies of relevant articles, which were then evaluated independently by the same review authors against the inclusion criteria. Review authors were not blinded to author names or institutions nor to journals of publication of potential studies.

Full-text electronic versions of the studies qualifying for inclusion were downloaded, printed, and organized with a study review data extraction form that was created by authors and attached to each study (see Appendix B). Three copies of each article and review form were made available to the review authors. Three review authors independently reviewed the articles to determine if they met inclusion criteria, and then met to resolve differences of opinion. For example, if one author indicated the study met the inclusion criteria and two authors excluded it based on the exclusion criteria, the characteristics of the study were discussed at length prior to making a decision. Exclusion criteria for this study were consistent with the HomVEE criteria (see Table 1).
Data Extraction and Management
One review author performed data extraction using standardized forms, which was checked by two additional review authors. We extracted data on study characteristics (i.e., study design, number of patients enrolled in the study, number of patients fulfilling the review's inclusion criteria), participant characteristics (i.e., age, sex, hearing loss, groups), interventions (i.e., information, resources, indirect services, direct services), outcome measures (i.e., names of receptive language, expressive language, etc. including designation as primary or secondary outcome measure), compliance, number of visits, and length of follow-up. We resolved discrepancies between authors by discussion.

At the top of each form, the full citation for the study was recorded. The program model name, target program population, and a brief program description were recorded. Each study was identified and categorized as a randomized control trial or a quasi-experimental study design and examined for validity and reliability of outcome measure choice. Key features of each program model were extracted from each study and recorded. Key features extracted were consistent with those identified in the HomVEE reviews: education requirement for home visitors, minimum number of visits, outcomes (favorable or unfavorable), and demonstrated sustainability for six months or more. Authors used the standardized form when completing critical appraisals and assessing the impact of the evidence (see Appendix B).

Critical Appraisal
Three review authors independently rated each study as high, moderate, or low based on the HomVEE criteria. Each review author independently synthesized the information, identified named program models, and determined if the program model met the criteria for designation as an evidence-based home visiting model or as a promising practice. Critical appraisal ratings were compared at a face-to-face meeting and differences of opinion were resolved through discussion. One review author organized the data into evidence summary tables.

Results

Literature Search Results
Results of the literature search are shown in Figure 1. The number of studies identified, screened for relevance, eligible for inclusion, and included in the final analysis are shown in the PRISMA flowchart. Seven hundred and sixty-three peer-reviewed articles were identified in electronic database searches. An additional 37 articles were identified by other means (checking reference lists, website searches, etc.). Seven hundred and two articles remained after removing duplicates. A total of 127 articles remained after the title screen. Eighty-seven studies were deemed irrelevant based on the abstract review, and 44 studies were identified as viable options for full review. Twenty-two studies were excluded on the basis of the exclusion criteria, leaving 22 publications for inclusion in the final analysis.

Home Visiting Models
Twenty-two publications met the inclusion criteria for systematic review. Within those publications, five (n = 5) home visiting intervention models, designed for children who were deaf or hard of hearing and their families, targeting an outcome in the child development and school readiness domain were identified.

1. Colorado Home Intervention Program (1969; CHIP)
2. Counseling and Home Training Program (CHTP)
3. Early Childhood Home Instruction Program (ECHI)
4. Project ASPIRE
5. SKI’HI

Evidence Tables
We assessed the effectiveness of each home visiting model and the outcome domain as well as each model’s implementation guidelines, if available. This section provides a summary of evidence of effectiveness by model and outcomes. Evidence tables (3, 4, & 5) show summary data for the five identified home visiting program models specific to children who are deaf or hard of hearing and their families. Table 3 shows the EHDI program model name, target population for the program, and brief published program model description. Table 4 shows a summary of the key features of the three remaining home visiting models from children who are deaf or hard of hearing and their families. Table 5 shows the program model name, the number of studies for each early intervention home visiting model, critical appraisal rating (i.e., high, moderate, low), outcome domain measure used, and full reference citation by program model for each of the publications.

Table 3. Program Model, Target Population, and Brief Descriptions of Home Visiting Programs for Children who are Deaf or Hard of Hearing and Their Families in the Child Development and School Readiness Outcome Domain

<table>
<thead>
<tr>
<th>Program Model</th>
<th>Target Population (in months)</th>
<th>Program Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>Colorado Home Intervention Program (CHIP)</td>
<td>Birth-11; 12-23; 24-35; 36-47; 48+</td>
<td>The Colorado Home Intervention Program (CHIP) started in 1969 was established through the U.S. Department of Education demonstration grant awarded through the University of Denver. The program is now administered through the Colorado Department of Education. The early intervention providers are trained professionals, deaf educators, speech/language pathologists, audiologists, early childhood special educators, bilingual educators, and social workers/psychologists who typically have earned graduate degrees in their field of expertise. Information (e.g., resources, strategies, development, methods of communication) is provided to parents through 1 or 1.5 hour sessions each week. Direct services to the child are not provided (Yoshinaga-Itano, 2003).</td>
</tr>
<tr>
<td>Counseling and Home Training Program (CHTP)</td>
<td>Birth-11; 12-23; 24-35</td>
<td>The goals of the family-focused Counseling and Home Training Program (CHTP) were to: (a) encourage rich and natural communication between children who are deaf and their families by using all possible modes of communication; (b) support the families’ realistic adaptation to deafness through counseling and supportive contacts with other parents and people who are deaf; and (c) to build a sense of competence and esteem for children who are deaf by developing an understanding and secure family context. This home-visiting model used a total communication philosophy and included six specific program components including services provided by a multidisciplinary team. At the time of publication, this program was offered through the Vancouver Children’s Hospital and served families in the Lower Mainland of British Columbia (Greenberg, 1984).</td>
</tr>
<tr>
<td>Early Childhood Home Instruction Program (ECHI)</td>
<td>Birth-11; 12-23; 24-35</td>
<td>The Early Childhood Home Instruction program emphasized the child’s language and communication development using auditory and speech training and manual communication within a family, home-based intervention model. ECHI used a total communication approach with Signing Exact English as the manual mode of communication. The intervention program also made available a parent support group and a center-based playgroup to promote language development in play environments and interaction among toddlers who are deaf. At this time of publication, this program was operated out of Children’s Hospital and Regional Medical Center, Seattle, WA (Calderon &amp; Low, 1998; Calderon &amp; Naidu, 2000).</td>
</tr>
</tbody>
</table>
2 randomized control trials. The critical appraisals are 1 moderate and 2 high ratings. SKI*HI, the fifth model identified, has been in operation for 44 years and has one publication critically appraised as a low rating.

Key Features by Program Model
Table 4 shows a summary of the key features of three EHDI home visiting models: CHIP, Project ASPIRE, and SKI*HI. Key features include the target population in months; minimum required education for home visiting personnel; minimum required reported visit frequency; number of research studies reporting favorable primary outcomes (direct observation, direct assessment, administrative records); number of research studies reporting favorable secondary outcomes (parent report); and sustainable outcomes, replication, and number of unfavorable outcomes reported by program model.

The review process revealed 14 publications meeting criteria for the CHIP program, 3 for Project ASPIRE, and 1 for SKI*HI. Project ASPIRE targets the birth to three population through parent education while CHIP and SKI*HI target children aged birth to five. CHIP and Project ASPIRE report a training requirement for home visiting personnel, SKI*HI does not. Favorable outcomes are reported in all 14 publications for CHIP, in 3 publications

<table>
<thead>
<tr>
<th>Program Model</th>
<th>Targeted Population (in months)</th>
<th>Minimum HV Staff Education Required</th>
<th>Minimum Required Visit Frequency</th>
<th>Number Favorable Primary Outcomes</th>
<th>Number Favorable Secondary Outcomes</th>
<th>Favorable Outcome Sustained</th>
<th>Favorable Impact/Replicated</th>
<th>Number of Unfavorable Outcomes</th>
</tr>
</thead>
<tbody>
<tr>
<td>Colorado Home Visiting Program (CHIP)</td>
<td>Birth-11; 12-23; 24-35; 36-47; 48+</td>
<td>Yes</td>
<td>Yes</td>
<td>14</td>
<td>14</td>
<td>Yes</td>
<td>Yes</td>
<td>0</td>
</tr>
<tr>
<td>Project ASPIRE</td>
<td>Birth-11; 12-23; 24-35</td>
<td>Yes</td>
<td>Yes</td>
<td>3</td>
<td>3</td>
<td>Not Reported</td>
<td>No</td>
<td>0</td>
</tr>
<tr>
<td>SKI*HI</td>
<td>Birth-11; 12-23; 24-35; 36-47; 48+</td>
<td>Not Reported</td>
<td>Not Reported</td>
<td>1</td>
<td>1</td>
<td>Not Reported</td>
<td>No</td>
<td>0</td>
</tr>
</tbody>
</table>
for Project ASPIRE and in 1 publication for SKI*HI, with no unfavorable outcomes. CHIP studies demonstrated replication and sustainability for 6 months or more. Replication and sustainability was not reported for Project ASPIRE or for SKI*HI.

Evidence-Based Home Visiting Model or Promising Practice

Three program models were assessed to determine if they met the HomVEE criteria for designation as an evidence-based model or a promising practice. One program model met the criteria for designation as an EHDI evidence-based home visiting model and one program model met the criteria for designation as an EHDI promising practice. One program model did not meet the designation criteria for either category.

The program model name, the number of studies for each EHDI home visiting model, critical appraisal rating (i.e., high, moderate, low), outcome domain measure used, and full reference citation by program model for each of the publications are shown in Table 5. References are organized chronologically. The outcome domain measure is the instrument or test tool that was used to collect data relevant to auditory, speech, language, listening, literacy, and other developmental outcomes relevant to the outcome domain of child development and school readiness.

The CHIP model meets the HomVEE criteria as an evidence-based home visiting model. Data for CHIP showed 14 published impact studies over the past 20 years, each with a critical appraisal rating of moderate for evidence of effectiveness. All studies were high quality quasi-experimental research designs with no randomization or comparison group. Outcome measures used in these studies were standardized, reliable, valid instruments and included the Emotional Availability Scales (EAS; Biringen, Robinson, & Emde, 1998), Expressive One-Word Picture Vocabulary Test (EOWPVT4; Martin & Brownell, 2010), Child Development Inventory (CDI; Ireton, 1992), MacArthur Communication Development Inventory: Expressive Vocabulary (MCDI – EV) and Receptive Vocabulary (MCDI – RV) subtests (Fenson et al., 1993), and the Test of Auditory Comprehension of Language (TACL4; Carrow-Woolfolk, 2013).

Project ASPIRE meets the HomVEE criteria as a promising practice. Data showed three published studies over the past five years. One study was a high quality quasi-experimental study with a critical appraisal rating of moderate. The other two studies employed a randomized control design and received a high critical appraisal rating. Outcome measures included a developmental questionnaire, video language sample analysis, and sub-analyses of the Language ENvironment Analysis (LENA) system. This program model is considered a “promising practice” until evidence of sustainability has been demonstrated.

The SKI*HI model did not meet the HomVEE criteria as either a promising practice or as an evidence-based model. Data showed one publication that did not meet the critical appraisal criteria rating as high (randomized control trial) or moderate (high quality quasi-experimental study design).

Table 5. Critical Appraisal, Outcome Measure and Full Reference By Program Model for the Child Development and School Readiness Outcome Domain for Families of Children who are Deaf or Hard of Hearing

<table>
<thead>
<tr>
<th>Program Model</th>
<th>Number of Studies</th>
<th>Study Rating</th>
<th>Outcome Assessment Measure</th>
<th>Reference</th>
</tr>
</thead>
<tbody>
<tr>
<td>Colorado Home Visiting Program (CHIP)</td>
<td>14</td>
<td>Mod</td>
<td>Minnesota Child Development Inventory</td>
<td>Apuzzo, M., &amp; Yoshinaga-Itano, C. (1995). Early identification of infants with significant</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Minnesota Child Development Inventory</td>
<td>hearing loss and the Minnesota Child Development Inventory. <em>Seminars in Hearing</em>, 16, 124–139.</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>MacArthur Communication Development Inventory: Expressive Vocabulary</td>
<td>Yoshinaga-Itano, C., &amp; Snyder, L. (1998). The relationship of language and symbolic play in</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>MacArthur Communication Development Inventory: Expressive Vocabulary</td>
<td>Mayne, A. (1998a). Expressive vocabulary development of infants and toddlers who are</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>MacArthur Communication Development Inventory: Expressive Vocabulary</td>
<td>deaf or hard of hearing. In C. Yoshinaga-Itano &amp; A. L. Sedey (Eds), Language, speech and</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>MacArthur Communication Development Inventory: Expressive Vocabulary</td>
<td>and social-emotional development of children who are deaf and hard of hearing: The early years.</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>MacArthur Communication Development Inventory: Expressive Vocabulary</td>
<td><em>Volta Review</em>, 100, 29–52.</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>MacArthur Communication Development Inventory: Expressive Vocabulary</td>
<td>Mayne, A. (1998b). Receptive vocabulary development of infants and toddlers who are deaf or</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>MacArthur Communication Development Inventory: Expressive Vocabulary</td>
<td>hard of hearing. In C. Yoshinaga-Itano &amp; A. L. Sedey (Eds), Language, speech and</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>MacArthur Communication Development Inventory: Expressive Vocabulary</td>
<td>social-emotional development of children who are deaf and hard of hearing: The early years.</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>MacArthur Communication Development Inventory: Expressive Vocabulary</td>
<td>comparison of the links between emotional availability and language gains in young children</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>MacArthur Communication Development Inventory: Expressive Vocabulary</td>
<td>Touch and emotional availability in hearing and deaf or hard of hearing toddlers and their</td>
</tr>
</tbody>
</table>
and was therefore rated as low. The SKI*HI Language Development Scale was used as the outcome measure. This scale is standardized or normed on children who are deaf and hard of hearing, and not on their typically hearing peers, thus – would not be valid as a language assessment tool for children using spoken language.

Summary and Conclusions

Summary

Results of our study revealed 22 publications from which five EHDI home visiting programs were identified. CHIP met the criteria for designation as an EHDI Evidence Based Home Visiting Model and Project ASPIRE was identified as an EHDI Promising Practice. These results are important and demonstrate consistency with the purpose of EHDI articulated by JCIH (2013). Implications of these results are provided for practice, policy, and future research efforts.

El services represent the purpose and goal of the entire EHDI process. Screening and confirmation that a child is DHH [deaf or hard of hearing] are largely meaningless without appropriate, individualized, targeted, and high-quality intervention. For the infant or young child who is DHH to reach his or her full potential, carefully designed individualized intervention must be implemented promptly, utilizing service providers with optimal knowledge and skill levels and providing services on the basis of research, best practices, and proven models (JCIH, 2013, p. e1324).

Overall Completeness and Applicability of Evidence

One issue that HomVEE does not differentiate or comment on in their studies is the difference between a home visiting program model and a home visiting curriculum model. This is a very important distinction that we want to draw attention to as it has very different implications for practicing EHDI professionals. The two EHDI home visiting models identified in this study are very different types of home visiting models.

Project ASPIRE is a home intervention curriculum program model currently in development that is not yet commercially available. It has a specific set of objectives related to listening and spoken language, specific materials for use in parent training sessions, and a specific goal of facilitating listening and spoken language. Therefore, it is most appropriate for hearing parents choosing the aggressive use of technology to access auditory sounds. It is an innovative, well-designed, technology-based, culturally sensitive, active engagement curriculum targeting the needs of adult learners developed by a multidisciplinary team. For practicing professionals, this is a curriculum that one might choose to provide indirect services in the form of parent education. It is also the only curriculum developed specifically for children who are deaf or hard of hearing and their families with a rigorous and robust research agenda guiding the development. It is the only curriculum the authors are aware of in which prospective research with randomized group treatment has demonstrated

<table>
<thead>
<tr>
<th>Project ASPIRE</th>
<th>Mod</th>
<th>Overall Completeness and Applicability of Evidence</th>
</tr>
</thead>
<tbody>
<tr>
<td>SKI*HI</td>
<td>3</td>
<td>High</td>
</tr>
<tr>
<td></td>
<td>Low</td>
<td></td>
</tr>
</tbody>
</table>

Note: Mod = Moderate
evidence of effectiveness. This is very similar in structure and philosophy to one of the national home visiting models designated by HomVEE known as PALS (Play and Learn Strategies; Landry et al., 2012; Landry, Smith, & Swank, 2006; Roggman & Cardia, 2014). This program is a curriculum developed to facilitate language development through parent training. It is supported by rigorous and robust research following a focused research agenda appropriate to the target population and target audience.

In contrast, CHIP is part of a multidisciplinary integrated statewide EHDI system designed to meet the needs of a diverse population in a geographically diverse state. As such, CHIP does not subscribe to one specific curriculum with targeted communication goals, but instead, offers a continuum of services from which families can choose to best meet their individual needs. Statewide data is warehoused at the University of Colorado, Boulder and serves as a rich repository from which retrospective analyses can be done. Prospective randomized controlled trials are not part of this system and probably never will be. However, the components of this early intervention home visiting program are consistent with the JCIH (2007, 2013) guidelines. It is the only statewide EHDI home visiting program with published outcome data and serves as the standard for program development and implementation.

Another important consideration in the completeness and applicability of our study is telepractice. HomVEE does not address this issue and did not include telepractice services in their definition of home visiting programs. Although by nature, telepractice is a home-based service, we chose to follow the HomVEE definition and did not include studies using telepractice as a service delivery method in this systematic review.

Quality of Evidence
The quality of the studies included in this systematic review was high overall. Randomized controlled trials were well designed and rated as high impact, and despite the moderate impact rating for the Colorado studies, they employed a repeatable and replicable methodology to facilitate developmental outcomes. These Colorado outcome studies were well-designed quasi-experimental studies using matched designs, multi-variate analyses, and covariance statistic designs and multiple regression techniques using both step-wise and block designs (Yoshinaga-Itano, 2004). The internal validity of the studies was high with little selection, attrition, or detection bias. Confounding variables were limited or controlled by research design. In addition, external validity was high with well-described participant populations in all studies.

Potential Biases in the Review Process
This systematic review used a very broad search strategy for identifying eligible studies, although it is unlikely that eligible studies were missed, it is never possible to rule out reporting bias.

Conclusions of Evidence-Based Review
The CHIP EHDI model should be submitted to HomVEE for consideration as a designated national evidence-based home visiting model specific to children who are deaf or hard of hearing and their families. The Project ASPIRE home visiting curriculum model should be submitted to HomVEE for consideration of designation as a promising practice for facilitating listening and spoken language development. Studies of the SKI*HI program are insufficient to recommend inclusion as an evidence-based model at this time.

Implications for Practice
High-quality, cost-effective services resulting in the best possible patient outcomes are at the heart of the national conversation regarding health care and education reform (Nicholson, Shapley, & Martin, 2012). Although the concept of healthcare and service quality assessment has been around for most of a century, it has been a hot topic in the healthcare and education arena for the past decade. Quality in healthcare has been defined by the Agency of Healthcare Quality and Research (AHQR, 2003) as safe, timely, patient-centered, efficient, and equitable service delivery with full consideration of a patient’s preference and values. This definition can be viewed in a broad sense, encompassing intervention services provided by audioligists, speech pathologists, early interventionists, etc. No one would deny that the ultimate goal for any diagnostic and/or intervention service in the field of communication disorders is to achieve the best possible results or outcomes by providing the right services, at the right time, in the right way (Nicholson et al., 2012). Although home visiting services have been provided as a service model for decades to children who are deaf or hard of hearing and their families, there is little high quality outcome data to support this practice, and the data that exists, is largely from one state. Child developmental outcomes (social emotions, language, and literacy) are the foundation for school readiness and school success, and the literature supports the use of home visiting as one cost-effective method of achieving these goals.

This article serves as a wake-up call to clinicians and researchers practicing in the field of deafness and early intervention to reach beyond disciplinary knowledge and skills and to continue to work together to achieve better parent and child outcomes, and to recognize the value of using evidence-based clinical protocols implemented systematically with outcome data collected, documented, databased, and studied at the group level. Increased awareness, cooperation with, collaboration among, partnerships between, and integration of systems in early intervention, medicine, public health, and education are one potential solution to the complex challenges posed by the families in need of these services.

Home visiting is one of the services on the continuum that should be available in every state as an option to meet the needs of the families of children who are deaf or hard of hearing. Home visiting and medical home initiatives share
goals of promoting the health and development of children, often through trusting longitudinal relationships (Tschudy, Toomey, & Cheng, 2013). Both provide children and their families with social support and anticipatory guidance (e.g., development, safety), and linkage to community resources and services. To fully capitalize on these synergies, the systems should be integrated, whenever possible prioritizing the particular strengths of each service and needs of the family (IDEA, 2004). State systems are challenged to do more than play together nicely in the sandbox, and instead to dig deep and join forces though thoughtful efforts in joint consideration, communication, cooperation, and collaboration to solve problems and to publish meaningful outcome data. These aspirations are not new, however, practitioners are challenged to come up with new and innovative solutions to help reduce barriers to high quality services which generate outcome data in a retrievable format. This is the approach taken by Suskind and colleagues in the conceptualization and development of Project ASPIRE (2014, 2016).

The decision for a parent to choose home visiting intervention should balance the benefits and downsides and integrate the parent/child’s values and preferences (Haynes, Devereaux, & Guyatt, 2002; JCIH 2007, 2013). Parents with a high preference for home based services may find that the advantages with regard to costs associated with time, travel, and transportation far outweigh the disadvantages. What authors found missing from the home visiting outcome literature was the parent perspective. Surprisingly, secondary outcomes were not considered that may have related not only to increased knowledge and skills on the part of the parent, but also to confidence, self-efficacy, and satisfaction with services. In addition, parent preferences about choice of the preferred method of learning (reading, watching video, listening, etc.) were not available in the studies reviewed. These aspects of home visiting intervention are data that could be collected by home visitors or at the program level to use in the development of programming and in quality improvement efforts.

Implications for Research

Results of this systematic review highlight the need for a systematic interdisciplinary outcomes-based approach to program evaluation to support and/or inform best practices. The current state of evidence for home visiting models for children who are deaf or hard of hearing and their families has been described. This is the right time to join the conversation of the Home Visiting Research Network (Duggan et al., 2013). This network was established in July 2012 to meet 3 objectives, (a) develop a national home visiting research agenda, (b) advance the use of innovative research methods to carry out this agenda, and (c) provide a research environment supportive of the professional development of emerging home visiting researchers (Duggan et al., 2013). The stated overarching goal of this organization is to promote the translation of research into policy and practice. They have developed a conceptual model of home visiting service delivery and outcomes, characteristics of families and providers, variables relevant to family and home visitor relationships and demographic variables such as psycho-social well-being; cognitive capacity and attitudes; and perceived norms, personal agency, knowledge, skills, and dispositions (Duggan et al., 2013). The National Home Visiting Research Network (2013) priorities, a multidisciplinary collaboration, include the following:

1. Strengthen and broaden home visiting effectiveness
2. Identify core elements of home visiting
3. Promote successful adoption of home visiting innovations
4. Promote successful adaptation of home visiting innovations
5. Promote fidelity in implementing home visiting innovations
6. Build a stable, competent home visiting workforce
7. Promote family engagement in home visiting
8. Promote home visiting coordination with other services for families
9. Promote the sustainment of effective home visiting

Contributions that researchers can make, specific to children who are deaf or hard of hearing include child population variables (e.g., age of diagnosis, age of enrollment in early intervention), intervention variables (frequency of intervention, dose per week, number of visits, home versus clinic, qualifications of providers, etc.), comparison groups (prospective or retrospective, randomized or matched), and outcome variables (auditory development, listening skills, etc.). It is incumbent upon current and future researchers in the fields of communication sciences and disorders, deafness, and early intervention to design, implement, and study voluntary home visiting programs for children who are deaf or hard of hearing, and participate in longitudinal interdisciplinary data collection.

Furthermore, collaborative efforts in tracking child and family outcomes, and adherence to robust program evaluation designs are needed and provide an adequate level and quality of evidence for effectiveness (Korfmaner et al., 2012). These authors provide an invaluable practical tool for use in the cross disciplinary assessment of home visiting with common components of quality programming and specific operational anchors for measurement across multiple program models. Program evidence like this, coupled with primary (child) and secondary (parent) outcome data, can be used to guide program development, design decisions in EHDI programs, plan quality improvement initiatives, and influence policy.
## Appendix A

### Systematic Review Databases, Vendor, and Support

<table>
<thead>
<tr>
<th>Databases Available for Systematic Review Search</th>
<th>Vendor</th>
<th>Supported by</th>
</tr>
</thead>
<tbody>
<tr>
<td>CINAHL ® Plus with full text</td>
<td>EBSCO</td>
<td>UAMS</td>
</tr>
<tr>
<td>Education Research Complete</td>
<td>EBSCO</td>
<td>UALR</td>
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<tr>
<td>Education Resources Information Center</td>
<td>EBSCO</td>
<td>UAMS</td>
</tr>
<tr>
<td>JSTOR ®</td>
<td>ITHAKA</td>
<td>UALR</td>
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<td>PsycINFO ®</td>
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<td>Psychology and Behavioral Sciences Collection</td>
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<td>UAMS</td>
</tr>
<tr>
<td>PubMed (MEDLINE)</td>
<td>National Library of Medicine</td>
<td>UAMS</td>
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<tr>
<td>SociINDEX™</td>
<td>EBSCO</td>
<td>UAMS</td>
</tr>
<tr>
<td>Web of Science ™</td>
<td>Thomson Reuters</td>
<td>UAMS</td>
</tr>
</tbody>
</table>

*Note: CINAHL = Cumulative Index to Nursing and Allied Health Literature; EBM = Evidence-based Medicine; UALR = University of Arkansas at Little Rock; UAMS = University of Arkansas for Medical Sciences*
### Study Full Citation:


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Database: 
Reviewer: 

### (1) Study Screen Details

<table>
<thead>
<tr>
<th>Screening Decision</th>
<th>Screening Conclusion</th>
</tr>
</thead>
<tbody>
<tr>
<td>Study Passes Screens</td>
<td>Yes</td>
</tr>
<tr>
<td>Eligible for Review</td>
<td>Yes</td>
</tr>
</tbody>
</table>

### (2) Study Design Details (Circle Appropriate Indicator)

<table>
<thead>
<tr>
<th>Rating</th>
<th>Study Design</th>
<th>Outcomes</th>
<th>Threats to Validity</th>
<th>Outcome Effect</th>
</tr>
</thead>
<tbody>
<tr>
<td>High</td>
<td>Randomized Control Trial (RCT)</td>
<td>Primary Child Outcomes</td>
<td>Number of Subjects Number of Groups Instrumentation</td>
<td>Favorable No Effect Unfavorable</td>
</tr>
<tr>
<td>Mod</td>
<td>Quasi Experimental Cross Sectional Cohort</td>
<td>Secondary Parent Report Parent Outcomes</td>
<td>Differences between participants</td>
<td></td>
</tr>
<tr>
<td>Low</td>
<td>Parent Outcomes</td>
<td></td>
<td></td>
<td>Not Measured</td>
</tr>
</tbody>
</table>

### (3) Study Characteristics

#### Population
Child who is deaf or hard of hearing age birth to five and parent/caregiver

#### Intervention
Home Visiting Program
Name:

#### Outcomes Targeted
Child Development and School Readiness

Outcome Measures
(specify title of test measurement for assessment)

- Receptive Language (spoken or sign)
- Expressive Language (spoken or sign)
- Auditory Skill Development
- Listening Skills
- Literacy Development
- Speech Development


Losing Ground: Awareness of Congenital Cytomegalovirus in the United States

Sara M. Doutre,1,2 Tyson S. Barrett,1,2 Janelle Greenlee,3 Karl R. White,1,2

1National Center for Hearing Assessment and Management, Utah State University
2Department of Psychology, Utah State University
3National CMV Foundation

Abstract

One in 150 infants is born with cytomegalovirus (CMV) and one in 750 will have lifelong disabilities due to CMV. Even though congenital CMV is the leading viral cause of congenital disabilities and the leading non-genetic cause of childhood hearing loss, most adults have never heard of it. Data from the 2015 and 2016 HealthStylesTM surveys were analyzed and compared to data from similar studies and show an awareness rate of 7% for U.S. adults (5% for men and 9% for women), a statistically significant decrease from 2005 and 2010 HealthStylesTM surveys. Predictors of awareness include gender and education level. The presence of a child ages 0–5 in the household does not increase the chance that an adult in the household is aware of CMV. CMV is a large public health burden and further research needs to be focused on awareness and prevention of the negative sequela associated with congenital CMV.

Acronyms: CDC = Centers for Disease Control and Prevention, CMV = Cytomegalovirus, IOM = Institutes of Medicine (now known as National Academies of Sciences, Engineering, and Medicine), STD = Sexually Transmitted Disease

Corresponding author’s contact information: Sara M. Doutre; National Center for Hearing Assessment and Management, Utah State University; Phone: 801.620.0932; Email: saradoutre@gmail.com

Introduction

Cytomegalovirus (CMV) is a member of the herpes family of viruses, spread through bodily fluids including saliva, blood, genital secretions, urine, and breast milk. Ninety percent of the U.S. population has had CMV by the time they are 80 years old and most do not experience any symptoms (Fowler & Boppana, 2006; Staras, Dollard, & Radford, 2006).

Nonetheless, CMV has a very heavy disability burden when acquired congenitally. It is the leading cause of non-genetic hearing loss (Fowler & Boppana, 2006) with 15–20% of bilateral moderate to profound sensorineural hearing loss caused by CMV (Grosse, Ross, & Dollard, 2008). Congenital CMV also causes mental retardation, cerebral palsy, and many other disabilities (Dollard, Grosse, & Ross, 2007). Approximately 0.7% of infants are estimated to be born with congenital CMV in the United States, leading to 30,000 annual cases. About 90% of these babies are referred to as asymptomatic because there are no clinically apparent symptoms of the infection, and 10% are referred to as symptomatic because there are obvious clinical abnormalities (Boppana, Ross, & Fowler, 2013). Approximately 6,000, or one in five of those newborns with congenital infection will go on to develop permanent disabilities such as microcephaly, hearing loss, vision loss, cerebral palsy, seizure disorders, or cognitive impairment (Cannon, 2009).

According to the Institute of Medicine (IOM, 2000), the cost of medical and educational care for children with disabilities known to be due to congenital CMV in the United States is $1.9 billion per year. Given such high costs associated with congenital CMV, the IOM identified the development of a vaccine to prevent congenital CMV as a top priority. However, a vaccine appears to be years, if not decades away (Adler & Nigro, 2013).

There is no FDA-approved treatment for congenital CMV, but recent research by Kimberlin et al. (2015) on the use of Valganciclovir to treat symptomatic congenital CMV infections is promising. Even if an approved treatment or a vaccine can be developed and becomes widely available, the best alternative for reducing the incidence of congenital CMV at the present time appears to be more widespread use of basic hygiene practices among pregnant women to avoid transmission via saliva or urine from young children (Adler & Nigro, 2013; Pass & Anderson, 2014; Swanson & Schleiss, 2013). The first step in improving hygiene practices that will likely lead to reducing the incidence of congenital CMV, is ensuring that the general public, especially men and women of childbearing age, knows about the existence and consequences of CMV.

This paper presents previously unpublished findings from the 2015 and 2016 HealthStylesTM surveys about the awareness of CMV among adults in the United States. It uses the results of analyses to identify further research
needs and guidance for policymakers and public health programs on where to focus efforts to increase awareness of congenital CMV.

CMV Transmission
Pregnant women are most likely to contract CMV from young children and intimate partners (Fowler & Pass, 2006). CMV is transmitted from young children to pregnant women through urine or saliva during diaper changes, sharing of eating utensils, or exchanging saliva when kissing. CMV can also be sexually transmitted.

Johnson, Anderson, and Pass (2012) documented a number of factors associated with acquisition of CMV infections, often referred to in the literature as seroconversion. Seroconversion is when a person transitions from seronegative (has never had a CMV infection) to seropositive or seroconverted (has had a CMV infection). Low income pregnant women have almost three times the incidence (6.8%) of CMV infection compared to middle income pregnant women (2.5%). Thirty-seven percent of women in sexually transmitted disease (STD) clinics and 7.9–10% of daycare workers contract CMV infections each year. At the highest risk for CMV infection are parents of children who have recently had an active CMV infection and have CMV in their saliva, urine, and other bodily fluids that could be passed to another person (often referred to as shedding the virus).

CMV Prevention
Stowell, et al. (2014) found that while the CMV virus can live for up to 15 minutes on hard plastic and up to 5 minutes on crackers, no viable virus was recovered after washing hands with soap, sanitizer, or even just with water. The Centers for Disease Control and Prevention (CDC) recommend that an effective way of reducing exposure to and the incidence of CMV infection is by “regular hand washing, particularly after changing diapers.” (CDC, n.d.). Research studies have demonstrated that this and other preventative steps are effective. For example, Adler and Nigro (2013) found that only 3% (one of 37) of CMV-seronegative pregnant women with an infected young child who were educated about using simple hygiene practices in their daily routines seroconverted to CMV during pregnancy, while 42% (65 of 154 women) of pregnant women who were not educated seroconverted.

Other studies support the implementation of preventative hygienic precautions. Revello et al. (2015) found that only 1.2% of women who were given hygiene information and prospectively tested until delivery acquired a CMV infection, compared to 7.6% in a comparison group that were neither tested nor informed about CMV during pregnancy. Importantly, 93% of these women felt hygiene recommendations were worth suggesting to all pregnant women at risk for infection. In an earlier study, Vauloup-Fellous et al. (2009) found, for 2,595 seronegative women, that the incidence of maternal CMV conversion was reduced from 0.035% per woman-week to 0.008% per woman-week (P = .0005) following an educational intervention. Women were less than 25% as likely to acquire a CMV infection when the woman and her partner were given detailed information on preventative hygienic measures verbally and in writing.

Previous Assessments of CMV Awareness
The public health impact of congenital CMV infection is substantial and under-recognized. (Swanson & Schleiss, 2013). While congenital CMV is one of the most common causes of congenitally acquired childhood disabilities and is preventable, most women of childbearing age have never heard of it (Cannon, 2009; Jeon et al., 2006; Ross, Victor, Sumartojo, & Cannon, 2008).

Three surveys of public CMV awareness in the United States have been conducted in the past decade. Jeon et al. (2006) surveyed 643 women at seven geographic locations (Atlanta, GA; Birmingham, AL; Cleveland, OH; Provo, UT; Richmond, VA; Chicago, IL; and Houston, TX) and found that only 142, or 22%, of women surveyed had heard of congenital CMV. Women’s awareness statistically significantly increased with higher levels of education, older age, and previous employment in a healthcare profession. When multiple regression analyses were used to adjust for other covariates, age no longer predicted awareness, but higher levels of education (high-school diploma or less, OR = 1.0; some college, OR = 1.5; bachelor’s degree or more, OR = 2.1; p = .03) and employment in a healthcare profession (no, OR = 1.0; yes, OR = 6.8, p < .0001) remained statistically significantly related. The study found no statistically significant differences by income, race and ethnicity, or between women who had been pregnant and who had never been pregnant. The study also found that employment in a daycare setting did not impact awareness (no, 21%, OR = 1.0; yes, 27%, OR = 1.4; p = .18).

Jeon et al. (2006) also found that most women, even those who had heard of CMV, could not identify modes of CMV transmission or prevention and 23% (83 of 137) incorrectly stated that CMV could be prevented by avoiding cat litter. One strength of the study was that it compared awareness about CMV with awareness about other birth defects and childhood illnesses and first reported the disparity between awareness and incidence rates of various childhood conditions. Jeon et al. (2006) noted that 53% of respondents were aware of congenital rubella syndrome, which had been eradicated in the United States, compared to 22% who were aware of CMV. Comparing CMV awareness with other diseases and conditions provides context for the results and makes them more relevant for decision- and policymakers.

A limitation of Jeon et al.’s (2006) study was that participants were recruited from pediatric outpatient clinic waiting rooms (4 sites), an obstetrics/gynecology clinic, a university’s student center, and medical students and support staff in a hospital. The fact that the survey was a convenience sample administered in mainly healthcare settings means that it may not be representative of all women in the United States (for example, women with
knowledge about CMV were likely oversampled given that the survey was conducted in health care settings).

Awareness of congenital CMV was also queried in the 2005 and 2010 HealthStyles™ survey, a subset of a consumer mail survey of U.S. adults over 18 years of age commonly used by the CDC for public health planning (Ross et al., 2008). HealthStyles™ surveys oversample certain demographic groups to enable more precise estimates about responses from people in those groups, but then the data are weighted to create a nationally representative sample with respect to age, sex, race/ethnicity, income, and household size. In the 2005 HealthStyles™ survey, 2,656 females and 2,163 males responded to four CMV-related questions, but the analyses reported by Ross et al. (2008) only focused on women because they are at risk for transmitting CMV to an unborn child. The potential role of a sexual partner in spreading CMV to a pregnant woman was not considered. Four questions asked whether participants had heard of CMV, where they learned about CMV, knowledge about the effects of CMV, and whether they would willingly adopt measures to prevent CMV while pregnant. The survey also collected demographic variables including sex, age, income, race and ethnicity, level of education, and household size.

Ross et al. (2008) reported that 14% of women had heard of CMV, and consistent with Jeon et al. (2006), knowledge increased with level of education (did not graduate high school, 10%, OR = 1.0; graduated high school, 6%, OR = 0.6; attended college, 13%, OR = 1.4; graduated college, 22%, OR = 2.6; 5–8 years of graduate school, 23%, OR = 2.7; p < 0.001). Knowledge also increased with household income, but not when other covariates were controlled using multiple regression analyses.

Ross et al. (2008) also found that the preventative hygiene measures previously recommended by the CDC were judged to be easy to adopt by a large majority of participants, regardless of whether participants had heard of CMV. For example, 90% reported that washing hands would be very easy to adopt and 65% reported that it would be easy to adopt the recommendation to not share eating utensils with a young child. Fewer participants, 48%, reported that not kissing a young child on the mouth would be very easy, but 20% reported it would be somewhat easy.

The 2010 HealthStyles™ survey, with a sample of 2,181 women and 2,003 men, showed 13% of women and 7% of men had heard of CMV (Cannon et al., 2012). As with the 2005 survey, Cannon et al. (2012) only reported analysis results for women. Congenital CMV awareness varied by age, race/ethnicity, educational attainment, geographic region, and household income, with the strongest association between CMV awareness and the educational level of the respondent, even though awareness among women with post-graduate education was only 21%. Because only linear trend data were reported, odds ratios cannot be compared to previous surveys. The 2010 survey did not repeat questions related to the ease of implementing the CDC’s recommended precautions, but added questions regarding the number of times women with children under age 19 engaged in risk and preventative behaviors while their youngest child was still in diapers. The study found that both risk and preventative behaviors are common (e.g., 69% of women reported kissing young children on the lips, 42% reported sharing utensils with young children, 95% reported washing hands after diaper changing, and 65% reported washing hands after wiping a child’s nose).

Recently, Thackeray and Magnusson (2016) assessed childcare provider awareness of CMV and other infectious diseases by asking a random sample of licensed family and residential childcare providers in Utah to complete a 29-item questionnaire on awareness of CMV and other infectious diseases. The study focused on awareness as well as knowledge of how to prevent diseases in childcare settings. Thackeray and Magnusson found that 18.5% of 306 respondent childcare providers had heard of CMV. For comparison, 99.4% were aware of influenza, 67.2% of giardia, 24.9% of toxoplasmosis, and 23.2% of enterovirus. Because childcare providers are at higher risk for CMV infections and may be serving infants and young children with asymptomatic CMV infections, it is particularly important that they are aware of CMV (Thackeray & Magnusson, 2015). While targeted information has been provided to licensed childcare providers in Utah (Utah Department of Health, n.d.), public awareness efforts should reach both licensed and unlicensed childcare providers everywhere.

Finally, a 2014 survey of congenital CMV knowledge among medical students (Baer, McBride, Caviness & Demmler-Harrison, 2014) found that 34% of first year medical students and 100% of second through fourth year medical students at Baylor University, were somewhat or very familiar with CMV. Self-reported awareness by these students who were enrolled at a university with a history of significant research conducted on congenital CMV, was confirmed based on second through fourth year students’ knowledge of modes of CMV transmission and signs and symptoms of CMV. Similar studies have not been conducted at other institutions where CMV research is not a priority. Consistent with results from the 2005 HealthStyles™ survey of CMV awareness completed by the CDC showing correlation with employment in a medical field, students’ awareness in this study was strongly correlated with level of medical education (p < .0001).

In summary, CMV awareness among the general population is low and appears to be declining over time. While there are some predictors of CMV awareness, even those factors only raise CMV awareness levels among the general population slightly. This article uses data from more recent HealthStyles™ surveys to evaluate whether CMV awareness rates are declining and discusses potential research and public health policy mechanisms that could be used to increase awareness about CMV.
CMV Awareness Programs

Recently, there have been a number of public health efforts to increase awareness about CMV. In 2013, the Utah Legislature unanimously passed the first CMV public health initiative law (McVicar, 2014). Utah’s law mandates that the Utah Department of Health implement a public health education campaign to inform women who are pregnant or might become pregnant about CMV, the risks associated with CMV, and the recommended prevention measures. The law also mandates an education campaign for medical and child-care professionals. The charge for implementation was given to the Early Hearing Detection and Intervention (EHDI) program within the state’s Department of Health.

Utah’s law was the first of its kind and appears to have spurred action in several other states. As of 2015, five states had enacted CMV laws (Doutre, 2015). Based on enactment of these laws, multiple programs have been initiated by state Departments of Health to educate women about CMV. In addition to legislatively-mandated public awareness programs, other EHDI programs are leading efforts to raise awareness of CMV (Mirizzi et al, 2015).

A number of non-profit organizations are also working to raise awareness of CMV. The National CMV Foundation (2015) was founded when four non-profit CMV organizations joined forces with an aim to “empower women, parents, families, and local community networks through grassroots engagement to facilitate conversations about CMV and to champion the cause against congenital CMV” (http://www.nationalcmv.org).

As public health programs and non-profit organizations work to increase awareness about CMV, it is important to document how people’s awareness of CMV is changing. Such efforts will help focus educational efforts, identify factors that influence likelihood of CMV awareness, evaluate effectiveness of approaches to increasing CMV awareness, and determine areas of need and opportunity for the greatest impact. This article combines results from the previously-reported 2005 and 2010 HealthStyles™ survey data (Ross et al., 2008; Cannon et al., 2012) with previously unreported analyses from the 2015 and 2016 HealthStyles™ survey data to examine whether public awareness of CMV is increasing, decreasing, or staying the same.

Methodology

Data Set

The National CMV Foundation contracted with Porter Novelli to include the same awareness question about CMV in the 2015 and 2016 Summer HealthStyles™ surveys that had been asked in the 2005 and 2010 versions of the survey. Have you heard of the following: congenital rubella syndrome, beta strep (Group B strep), HIV/AIDS, congenital cytomegalovirus (CMV), Down syndrome, sudden infant death syndrome (SIDS), fetal alcohol syndrome, autism, spina bifida, congenital toxoplasmosis, and parvovirus B19? The resulting data were provided to Utah State University for analysis. Both data sets were collected by Porter Novelli Public Services via GfK’s KnowledgePanel® (a national, probability-based panel that is representative of the entire U.S. population). GfK’s KnowledgePanel® consists of 55,000 panel members who are randomly recruited from a sample frame of residential addresses “including households that: have unlisted telephone numbers, do not have landline telephones, are cell phone only, do not have current internet access, and do not have devices to access the internet” (GfK, 2013). GfK provides household without phone and or internet with a laptop computer and internet access. The panel of 55,000 is continuously replenished and respondents for individual surveys are selected from the larger panel to ensure a representative sample.

The summer 2015 HealthStyles™ survey was conducted from June 11 to June 29, 2015 with 4,127 adults completing the survey (a response rate of 67%). All respondents received compensation for completing the survey in the form of cash-equivalent reward points worth approximately $10. Respondents with incomplete responses (who did not answer at least half of the questions, n = 7) and speeders (who completed the survey in 7 minutes or less, n = 33) were removed from the data.

The summer 2016 survey was conducted from June 24 to July 11, 2016 using the same procedures and had a response rate of 68% with 4,203 of 6,166 adults completing the survey. Participants received the same compensation as that provided in 2015. Incomplete (n = 10) and speeder (n = 39) responses were removed from the data set.

Participants responded to a question asking if they had heard of the following conditions: congenital rubella syndrome, beta strep (Group B strep), HIV/AIDS, congenital cytomegalovirus (CMV), Down syndrome, sudden infant death syndrome (SIDS), fetal alcohol syndrome, autism, spina bifida, congenital toxoplasmosis, and parvovirus B19. The question asked for each condition was “Have you heard of [condition]?”. Response choices were Yes, No, or the participant could refuse to answer the question. Respondents’ awareness of CMV compared to awareness of other conditions provides context to policy and decision makers and allows for analysis of awareness compared to disease burden, making a case for the potential impact of CMV awareness initiatives.

Data were also available about each respondent’s race/ethnicity, gender, zip code, whether the respondent currently had children under age 18, ages of the respondent’s three youngest children, age, education (highest degree received and categorical), household size, household income, marital status, metro status (metro or non-metro), census region, employment status, housing status (own, rent, or occupied without payment of rent), and state of residence. Weights were provided so that survey responses could be matched to U.S. Current Population
Survey proportions using 9 factors: gender, age, household income, race/ethnicity, household size, education, census region, metro status, and prior internet access.

Data Analysis
An analysis of descriptive statistics was conducted for all study variables for both the 2015 and 2016 HealthStyles™ data using the R statistical software program. Rates of awareness for CMV were computed using data weighted for representativeness and stratified by demographic characteristics. In addition, CMV awareness rates were compared to awareness rates for other conditions queried in the survey.

A total of five logistic regression models were used to assess both the trend and characteristics related to CMV awareness. One model tests the trend across time, using the year as the independent variable. Two logistic regression models per year were used to determine the association of demographic conditions with CMV awareness, where CMV awareness was the binary outcome for both models. The first model examined basic demographic predictor variables: age, race, gender, education, and household income. The second model added two additional predictor variables to the model to examine parenthood and age of children: household presence of children under ages 0–1 and household presence of children ages 2–5. These variables were chosen based on the relativity of CMV awareness to families experiencing pregnancy and the increased risk of acquiring CMV from a young child.

Results
The 2015 and 2016 HealthStyles™ CMV awareness rates are 6.79% and 6.70% in the overall U.S. population when weighted for representativeness. Awareness rates for all levels of the various demographic characteristics are similarly low as shown in Table 1. Females have a higher rate of awareness than males (9.08% and 9.17% in 2015 and 2016 compared to 5.72% and 4.92%), but the number of females reporting awareness of congenital CMV has decreased from 14% and 13% in 2005 and 2010. Figure 1 is a summary of HealthStyles™ survey data from 2005, 2010, 2015 and 2016, showing a decrease over 11 years for women from 14% to 9% and for men a decrease from 2010 to 2016 from 7% to 5%. Data from the 2005 survey were not reported for men.

Table 1. U.S. Congenital Cytomegalovirus Awareness By Demographic Characteristics, 2015 and 2016

<table>
<thead>
<tr>
<th></th>
<th>2015 (N = 4121)</th>
<th>2016 (N = 4197)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Overall Awareness (weighted)</td>
<td>6.79% (310)</td>
<td>6.70% (300)</td>
</tr>
<tr>
<td>Gender</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Male</td>
<td>5.72% (109)</td>
<td>4.92% (98)</td>
</tr>
<tr>
<td>Female</td>
<td>8.08% (201)</td>
<td>9.17% (202)</td>
</tr>
<tr>
<td>Race</td>
<td></td>
<td></td>
</tr>
<tr>
<td>White</td>
<td>7.41% (256)</td>
<td>7.11% (250)</td>
</tr>
<tr>
<td>Black/African-American</td>
<td>7.67% (32)</td>
<td>7.69% (34)</td>
</tr>
<tr>
<td>Hispanic</td>
<td>2.92% (13)</td>
<td>5.13% (24)</td>
</tr>
<tr>
<td>American Indian or Alaska Native</td>
<td>12.12% (4)</td>
<td>0.00% (0)</td>
</tr>
<tr>
<td>Asian</td>
<td>10.81% (12)</td>
<td>6.14% (7)</td>
</tr>
<tr>
<td>Hawaiian/Pacific Islander</td>
<td>0.00% (0)</td>
<td>0.00% (0)</td>
</tr>
<tr>
<td>2+ Races</td>
<td>5.89% (6)</td>
<td>9.00% (9)</td>
</tr>
<tr>
<td>Currently have children under Age 18?</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Yes</td>
<td>10.23% (134)</td>
<td>9.77% (136)</td>
</tr>
<tr>
<td>No</td>
<td>6.25% (175)</td>
<td>5.86% (164)</td>
</tr>
<tr>
<td>Age</td>
<td></td>
<td></td>
</tr>
<tr>
<td>18–29</td>
<td>7.52% (41)</td>
<td>7.49% (494)</td>
</tr>
<tr>
<td>30–44</td>
<td>8.88% (82)</td>
<td>8.75% (86)</td>
</tr>
<tr>
<td>45–59</td>
<td>8.08% (111)</td>
<td>8.25% (111)</td>
</tr>
<tr>
<td>60+</td>
<td>5.94% (76)</td>
<td>4.80% (66)</td>
</tr>
<tr>
<td>Education</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Less than High School</td>
<td>3.77% (11)</td>
<td>3.62% (10)</td>
</tr>
<tr>
<td>High School</td>
<td>3.82% (47)</td>
<td>5.06% (63)</td>
</tr>
<tr>
<td>Some College</td>
<td>8.45% (106)</td>
<td>6.37% (81)</td>
</tr>
<tr>
<td>Bachelor’s Degree or Higher</td>
<td>10.86% (146)</td>
<td>10.40% (146)</td>
</tr>
<tr>
<td>Professional or Doctorate Degree</td>
<td>20.16% (26)</td>
<td>20.41% (30)</td>
</tr>
<tr>
<td>Marital Status</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Married</td>
<td>8.13% (188)</td>
<td>7.29% (179)</td>
</tr>
<tr>
<td>Divorced</td>
<td>7.88% (34)</td>
<td>7.46% (37)</td>
</tr>
<tr>
<td>Never Married</td>
<td>6.17% (52)</td>
<td>7.20% (58)</td>
</tr>
<tr>
<td>Living with Partner</td>
<td>7.09% (19)</td>
<td>6.02% (10)</td>
</tr>
<tr>
<td>Metro Status</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Metro</td>
<td>7.73% (267)</td>
<td>7.39% (265)</td>
</tr>
<tr>
<td>Non-Metro</td>
<td>6.44% (43)</td>
<td>5.71% (35)</td>
</tr>
</tbody>
</table>
A logistic regression analysis of awareness rates across years from 2005 – 2016 shows that the decrease in awareness among U.S. women is statistically significant (OR = 0.94, 95% CI = [0.93, 0.95], p < .0001). The lack of a combined rate reported in previous analyses does not allow for exploration of the statistical significance of the decline of overall awareness.

CMV awareness was compared to awareness of other congenital conditions associated with negative developmental outcomes, up to and including death. Respondents are least aware of CMV compared to the 10 other conditions. Table 2 presents 2015 and 2016 awareness rates of the 10 comparable conditions to congenital CMV and the estimated annual frequency, in number of congenital or infant cases that result in long-term disabilities for each condition. Figure 2 shows the disparity between awareness using 2016 results and frequency of the 11 surveyed conditions. Although other diseases with low awareness have relatively low occurrences, the difference between CMV’s relatively high occurrence and its low awareness is contrasted with Down syndrome, which has a similar occurrence rate (6,000 babies born with Down syndrome each year) but 85% to 89% report awareness of Down syndrome compared to 7% awareness for CMV.

Table 3 presents the results of each multivariate logistic regression model, reported as adjusted odds ratios (i.e., each odds ratio has been statistically adjusted for all other variables in the model), and their confidence intervals. In the first model, which included basic demographic predictor variables (age, race, gender, education, and household income), both gender and education level are statistically significant predictors of CMV awareness. In that model, women had an odds of awareness of one and a half to two times greater than men (2015: AOR = 1.56, 95% CI = [1.22, 2.00], p < .001; 2016: AOR = 1.94, 95% CI = [1.52, 2.51], p < .001) and Hispanic adults (men and women) were less than half as likely to be aware of CMV as white adults (2015: AOR = 0.37, 95% CI = [0.19, 0.67], p < .001). Education also was statistically significantly associated with awareness about CMV. For each increase in unit of education, the odds of awareness increased by 1.5 times (2015: AOR = 1.50, 95% CI = [1.29, 1.74], p < .001; 2016: AOR = 1.43, 95% CI = [1.23, 1.66], p < .001).

The second model included the presence of children ages 0–1 or ages 2–5 in the household, which was used as an indicator of whether the household had recently experienced a pregnancy. Adding these factors did not change the relationships seen in Model 1 for either year. Further, the additional variables (the presence of household members ages 0–1 or ages 2–5) does not statistically significantly predict CMV awareness.
### Table 2. Percentage of U.S. Adult Awareness of Childhood Conditions Comparable to Congenital Cytomegalovirus

<table>
<thead>
<tr>
<th>Condition</th>
<th>2015 Awareness</th>
<th>2016 Awareness</th>
<th>Approximate Annual U.S. Frequency</th>
</tr>
</thead>
<tbody>
<tr>
<td>Congenital Cytomegalovirus (CMV)</td>
<td>6.79%</td>
<td>6.70%</td>
<td>6,000c</td>
</tr>
<tr>
<td>Congenital Toxoplasmosis</td>
<td>8.27%</td>
<td>8.53%</td>
<td>400c</td>
</tr>
<tr>
<td>Congenital Rubella Syndrome</td>
<td>16.80%</td>
<td>13.27%</td>
<td>&lt; 3e</td>
</tr>
<tr>
<td>Beta Strep (Group B Strep)</td>
<td>17.87%</td>
<td>16.91%</td>
<td>380c</td>
</tr>
<tr>
<td>Parvovirus B19 (Fifth Disease)</td>
<td>22.52%</td>
<td>19.63%</td>
<td>1045f</td>
</tr>
<tr>
<td>Fetal Alcohol Syndrome</td>
<td>65.56%</td>
<td>61.04%</td>
<td>1200g</td>
</tr>
<tr>
<td>Spina Bifida</td>
<td>69.42%</td>
<td>64.54%</td>
<td>1500h</td>
</tr>
<tr>
<td>Sudden Infant Death Syndrome (SIDS)</td>
<td>83.96%</td>
<td>78.70%</td>
<td>1500i</td>
</tr>
<tr>
<td>Autism</td>
<td>88.59%</td>
<td>84.28%</td>
<td>60,000j</td>
</tr>
<tr>
<td>Down Syndrome</td>
<td>89.57%</td>
<td>85.44%</td>
<td>6,000k</td>
</tr>
<tr>
<td>HIV/AIDS</td>
<td>91.13%</td>
<td>86.33%</td>
<td>30l</td>
</tr>
</tbody>
</table>

Note: Awareness data taken from the 2015 and 2016 HealthStyles™ surveys.  
* Approximate frequency of infants affected with long-term disabilities, including death, by each condition.  
3 https://www.cdc.gov/mmwr/preview/mmwrhtml/mm6212a3.htm#fig  
5 http://www.cdc.gov/abcs/reports-findings/surveireports/gbs10.html  
7 http://www.cdc.gov/mmwr/preview/mmwrhtml/mm6212a3.htm#fig  
9 http://www.cdc.gov/abcs/reports-findings/surveireports/gbs10.html  
11 http://www.cdc.gov/ncbddd/spinabifida/data.html  
12 http://www.cdc.gov/ncbddd/spinabifida/data.html  
13 http://www.cdc.gov/birthdefects/downsyndrome/data.html  
14 HIV diagnosis at < 1 year, http://www.cdc.gov/mmwr/preview/mmwrhtml/mm6253a1.htm

**Figure 2.** U.S. adult awareness of childhood conditions from the 2016 HealthStyles™ surveys with approximate annual U.S. incidence of disability due to each condition.
Table 3. Results of Logistic Regression Analyses Identifying Factors That Do and Do Not Predict CMV Awareness (2015 and 2016)

<table>
<thead>
<tr>
<th></th>
<th>2015 Model 1</th>
<th>2015 Model 2</th>
<th>2016 Model 1</th>
<th>2016 Model 2</th>
</tr>
</thead>
<tbody>
<tr>
<td>Gender</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Male</td>
<td>1.00 [Reference]</td>
<td>1.00 [Reference]</td>
<td>1.00 [Reference]</td>
<td>1.00 [Reference]</td>
</tr>
<tr>
<td>Female</td>
<td>1.56 (1.22, 2.00)*</td>
<td>1.56 (1.22, 2.00)*</td>
<td>1.94 (1.52, 2.51)*</td>
<td>1.94 (1.52, 2.51)*</td>
</tr>
<tr>
<td>Education</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>White</td>
<td>1.50 (1.29, 1.74)*</td>
<td>1.50 (1.30, 1.74)*</td>
<td>1.43 (1.23, 1.66)*</td>
<td>1.43 (1.23, 1.66)*</td>
</tr>
<tr>
<td>Non-Hispanic</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Hispanic</td>
<td>0.37 (0.19, 0.67)*</td>
<td>0.37 (0.19, 0.67)*</td>
<td>0.64 (0.40, 1.04)</td>
<td>0.66 (0.40, 1.03)</td>
</tr>
<tr>
<td>Presence of Household Members age 0-1</td>
<td>0.89 (0.37, 1.82)</td>
<td>1.18 (0.67, 1.98)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Presence of Household Members age 2-5</td>
<td>1.02 (0.69, 1.46)</td>
<td>1.09 (0.78, 1.48)</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

* (p < .001)

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<th>2016 Model 1</th>
<th>2016 Model 2</th>
</tr>
</thead>
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<td>Gender</td>
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<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Male</td>
<td>1.00 [Reference]</td>
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<td>1.00 [Reference]</td>
<td>1.00 [Reference]</td>
</tr>
<tr>
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</tr>
<tr>
<td>Education</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>White</td>
<td>1.50 (1.29, 1.74)*</td>
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<td></td>
<td></td>
</tr>
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<td>1.09 (0.78, 1.48)</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

* (p < .001)

Table 3 presents the results of each multivariate logistic regression model, reported as adjusted odds ratios (i.e., each odds ratio has been statistically adjusted for all other variables in the model), and their confidence intervals. In the first model, which included basic demographic predictor variables (age, race, gender, education, and household income), both gender and education level are statistically significant predictors of CMV awareness. In that model, women had an odds of awareness of one and a half to two times greater than men (2015: AOR = 1.56, 95% CI = [1.22, 2.00], p < .001; 2016: AOR = 1.94, 95% CI = [1.52, 2.51], p < .001) and Hispanic adults (men and women) were less than half as likely to be aware of CMV as white adults (2015: AOR = 0.37, 95% CI = [0.19, 0.67], p < .001). Education also was statistically significantly associated with awareness about CMV. For each increase in unit of education, the odds of awareness increased by 1.5 times (2015: AOR = 1.50, 95% CI = [1.29, 1.74], p < .001; 2016: AOR = 1.43, 95% CI = [1.23, 1.66], p < .001).

The second model included the presence of children ages 0–1 or ages 2–5 in the household, which was used as an indicator of whether the household had recently experienced a pregnancy. Adding these factors did not change the relationships seen in Model 1 for either year. Further, the additional variables (the presence of household members ages 0–1 or ages 2–5) does not statistically significantly predict CMV awareness.

Discussion

Awareness of congenital CMV decreased by nearly 50% from 2010 to 2015 and 2016 despite the large disease burden and high frequency of infections. The 2015 and 2016 HealthStyles™ survey data showed lower awareness rates despite increased attention to congenital CMV in the public health and policy arenas. It is noteworthy that CMV awareness is even lower than congenital rubella syndrome, which has been eradicated, and lower than other less common conditions. Most of the documented efforts by public health entities to increase CMV awareness (e.g., Doutre, 2015; Mirizzi et al., 2015) have taken place since 2013 so it may be too early to see the impact of those activities, but the fact that CMV awareness appears to be declining is a serious concern.

Consistent with previous research, analyses of the HealthStyles™ survey data across multiple years showed that women are more likely to be aware of CMV than men. This difference is expected as congenital CMV is most relevant to pregnant women but the odds ratios of 1.56 (2015) and 1.94 (2016) are lower than desirable in order to promote prevention of transmission from mother to fetus during pregnancy. These data suggest that women’s doctors may not be counseling them on CMV despite its prevalence and the associated disability burden.

The significance of the differences by demographic factor are further explored in the logistic regression models. In addition to CMV awareness being higher among women
in 2015 and 2016 than for men, CMV also varied by race/ethnicity. Adult respondents reporting Hispanic ethnicity reported lower CMV awareness (2.92% in 2015 and 5.13% in 2016) than adults in general and adults reporting any other race or ethnicity category. If an adult currently has children under age 18 he or she is more likely to be aware of CMV, but awareness in this group remains low (10.23% in 2015 and 9.77% in 2016).

CMV awareness increases with increasing levels of education as reported in previous studies. The correlation of awareness with education level is concerning. Women with low socioeconomic status have almost three times the incidence of CMV infection compared to middle income pregnant women (Johnson et al., 2012) and it appears that awareness of CMV is often associated with higher education levels that may not be accessible to women of lower socioeconomic status. But, even in the most aware group (those with a professional or doctorate degree, n = 131), only 20% of the respondents to the 2015 and 2016 HealthStyles™ surveys had heard of CMV. Public awareness and education initiatives are needed at all levels.

At the highest risk for CMV infection are parents of children who have recently had an active infection and are shedding the virus in bodily fluids (Johnson et al., 2012). The surrogate measures to this variable are the measures of adults who report the presence of children ages 0–1 or the presence of children ages 2–5 in their household. The presence of children of these ages in the household were not statistically significantly related to CMV awareness. A large majority of respondents, 89.5%, did not report having any children ages 0–1.

**Conclusions**

Analyses of the 2015 and 2016 HealthStyles™ survey data shows that awareness of CMV is decreasing among adults in the United States. Because of the high burden of disease associated with congenital CMV, it is alarming that the virus is relatively unknown. There is good evidence that preventative hygienic measures taken by women and their partners can reduce the risk of CMV infection and thus the risk of transmitting CMV to a fetus (Adler and Nigro, 2013; Revello et al., 2015; Vauloup-Fellous et al., 2009). A logical precursor to the wider implementation of preventative hygienic measures is increased awareness of CMV. Therefore, the decreasing trend in CMV awareness documented by the HealthStyles™ survey data from 2005 to 2016 is of great concern.

CMV awareness is low for all subsets of the U.S. population, but it is especially low for Hispanic adults. Even though awareness is higher for women and those with higher education levels, awareness in those groups remains alarmingly low considering CMV’s disease burden and incidence rate. Furthermore, because CMV can be transmitted through sexual relations, it is important for men to be aware of what CMV is and how to prevent it.

The fact that CMV awareness is so low and is decreasing will hopefully help public health policymakers and program officials prioritize and focus their efforts to increase CMV awareness and prevention efforts. The data from the HealthStyles™ surveys also provide baseline data for beginning to evaluate CMV public health programs and specific initiatives, whether mandated by legislation or prioritized by stakeholders. Continued resources must be dedicated to increase awareness and prevention of this harmful virus.

**Limitations**
The greatest limitation of this study is the narrow definition of CMV awareness. Survey respondents responded to one yes/no question: “Have you ever heard of congenital cytomegalovirus (CMV)?” Ideally, additional questions would be asked to validate respondents’ awareness of CMV, such as how CMV is acquired, what the symptoms of CMV are, what measures may be taken to prevent CMV, or if there is a CMV vaccine available. Responses to these questions would allow policy makers and public health officials to better target their efforts to increase CMV awareness and prevention initiatives.

Another limitation of this study was the inability to evaluate CMV awareness by state and set a baseline for CMV awareness for states working to increase CMV awareness. Although state data were provided for each participant, sample sizes from most states were too small to establish awareness rates by state. These data would be useful in planning for CMV awareness programs.

**Implications for Further Research**

Although the data collected through the HealthStyles™ survey are useful in establishing the need for CMV awareness campaigns and education, further research is needed in many areas related to CMV awareness. First, no reported research has been conducted on the efficacy of different methods of raising public awareness and whether raising awareness of CMV leads to behavior changes in pregnant women.

Research has established the reasonableness and efficacy of recommended hygienic measures for reducing risk of acquiring a CMV infection during pregnancy (Adler and Nigro, 2013; Revello et al., 2015; Vauloup-Fellous et al., 2009). However, further research is needed on how to best educate women about hygienic practices and when is the most appropriate. For example, it would be useful to know if high school health education programs can effectively reach women who are just reaching child-bearing age. The correlation between education level and CMV awareness suggests a need for further research to study public health programs including those for high school students and other young adults. It would be useful to examine high school and undergraduate health education curriculum to determine whether information about CMV is currently included. In addition, further research should be conducted to determine whether health care providers are informing women planning to become pregnant and their sexual partners of CMV.
Implications for Policy and Public Health Programs

As state EHDI programs, other state agencies, and non-profit organizations embark on public awareness programs, consideration should be given to the fact that CMV awareness seems to be declining. There is a great need for general awareness and all populations are in need of education about congenital CMV including low socioeconomic and Hispanic populations. Consideration should be given to educating young adults at the beginning of their childbearing age. Programs should also ensure that educational materials are available to adults of all races and ethnicities, especially those with Hispanic ethnicity.

Although women are more likely than men to know about congenital CMV, it should be the goal of public education campaigns to raise awareness of both men and women. Because CMV can be spread through sexual activity (Fowler & Boppana, 2006; Staras et al., 2006), men should also be aware of and exercise hygienic precautions during a partner’s pregnancy to reduce the risk of obtaining a CMV infection.

CMV awareness rates are alarmingly low and there is a significant need for CMV education programs. As more states and other organizations pursue CMV awareness programs, further work will be needed to establish measures of the effectiveness of the public health and policy actions related to CMV. More detailed data, with larger sample sizes on a local scale, are needed to evaluate efforts of state stakeholders and non-profit organizations in developing policy and public information programs for CMV.

References

Late Newborn Hearing Screening, Late Follow-up, and Multiple Follow-Ups Increase the Risk of Incomplete Audiologic Diagnosis Evaluation

Tri Tran, MD, MPH,1,2 Ian Ng, MPH Thira Choojitarom, MD,4 Jeanette Webb, M. Ed,2 Wendy Jumonville, AUD, MS, CCCA,2 Mary Jo Smith, BS,2 Terri Ibieta, M. Ed,2 Melinda Peat, MCD,2 Susan Berry, MD, MPH,1,2

1Louisiana State University Health Sciences Center School of Medicine, Department of Pediatrics
2Louisiana Department of Health and Hospitals (DHH) Office of Public Health, Children with Special Health Services Program
3Tulane School of Public Health and Tropical Medicine, Department of Epidemiology
4Our Lady of the Lake Regional Medical Center in Baton Rouge, Louisiana

Abstract
This study aimed to determine whether the following factors were associated with an incomplete audiologic diagnosis evaluation (IAD): age at newborn screening, length of time between newborn hearing screening (NHS) and first follow-up, and total number of follow-ups. 2011-2013 linked Louisiana Early Hearing Detection and Intervention data and birth records were analyzed. Logistic regression models were used to evaluate different effects of the predictors on IAD among birth weight groups. In very low birth weight newborns, there were no statistical associations of IAD with age at NHS or length of time between NHS and first follow-up, but there was with the number of follow-up appointments. Among low birth weight or normal weight newborns, risk of IAD was significantly increased in babies with NHS > 30 days of age; length of time between NHS and first follow-up > 30 days; and having more than one follow-up. In order to reduce the number of infants who fail to complete the audiologic diagnosis evaluation, it is necessary to conduct NHS early, expedite follow-up, and decrease the number of follow-ups.

Key Words: Early Hearing Detection and Intervention, newborn hearing screening, audiologic diagnosis, lost to follow-up

Acronyms: ABR = auditory brainstem response, CDC = Centers for Disease Control and Prevention, EHDI = Early Hearing Detection and Intervention, IAD = incomplete audiologic diagnosis evaluation, JCIH = Joint Committee on Infant Hearing, LBW = low birth weight, LFU = lost to follow-up, LTD = lost to documentation, NICU = neonatal intensive care unit, NHS = newborn hearing screening, OAE = otoacoustic emissions, VLBW = very low birth weight

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Corresponding author’s contact information: Tri Tran, Email: tri.tran@la.gov or ttran@lsuhsc.edu

Introduction
Hearing loss plays a crucial role in delayed development of speech, language, and cognition in early childhood (Bess, Dodd-Murphy, & Parker, 1998; Holt & Svirsky, 2008; Moeller, 2000; Nicholas & Geers, 2006). Previous studies showed that children with hearing loss who received intervention services before 6 months of age had significantly better academic achievement and language development than those who received them after 6 months of age (Yoshinaga-Itano, 2003, 2004). Early Hearing Detection and Intervention (EHDI) programs in the United States are designed to detect congenital and early acquired hearing loss and link infants and their families to appropriate intervention services. The Joint Committee on Infant Hearing (JCIH) recommends that all newborns be screened at no later than 1 month of age; diagnosis be completed at no later than 3 months of age for infants who do not pass screening; and appropriate intervention be received at no later than 6 months of age for infants identified with hearing loss (JCIH, 2007). Hearing screenings and diagnoses completed after recommended timelines are considered barriers to the effectiveness of EHDI programs (White & Blaiser, 2011). Although the programs have effectively identified many children with early childhood hearing loss in recent years (Centers for Disease Control and Prevention [CDC], 2010; Muñoz, Blaiser, & Barwick, 2013), results of diagnostic tests for
children who fail hearing screening are not consistently reported to the EHDI programs (Williams, Alam, & Gaffney, 2015). Based on 2013 National CDC EHDI data, the rate of undocumented audiologic diagnosis was 41.0% among infants who did not pass the newborn hearing screening (CDC, 2013).

In Louisiana, all children who do not pass the final newborn hearing screening before hospital discharge are referred for hearing rescreening by audiologists or physicians at outpatient clinics. Of those who do not pass hearing rescreening, a referral is made for further evaluations to complete the audiologic diagnosis. Figure 1 presents a detailed process of newborn hearing screening, diagnosis, and intervention in Louisiana. In this study, a timeline for an audiologic diagnostic evaluation was defined as the time from the hearing rescreening at an outpatient clinic to the time when the audiologic diagnosis was completed. In Louisiana, in fact, many children undergo a prolonged and incomplete audiologic diagnosis process—in particular children with late newborn hearing screening, late follow-up, and many follow-ups. With a hypothesis that late newborn hearing screening, late follow-up, and multiple follow-up visits may increase the risk of incomplete audiologic diagnosis, this study was conducted to identify associations between incomplete audiologic diagnosis and (a) age at final newborn hearing screening prior to discharge, (b) length of time between final newborn hearing screening prior to discharge and first follow-up, and (c) total number of follow-ups among newborns who failed newborn hearing screening prior to hospital discharge. To our knowledge there are no published studies evaluating these associations.

Method

Study population
The study included children who were born in Louisiana between 2011 and 2013; had newborn hearing screening prior to hospital discharge, but did not pass; and completed at least one follow-up at an outpatient clinic. All follow-ups mentioned in the study were conducted at outpatient clinics by audiologists or physicians if the follow-up was for rescreening, and only by audiologists for audiologic diagnosis. The first follow-up was always for the hearing rescreening. The term screening in the study refers to hearing screening conducted before hospital discharge.

As mentioned previously, the timeline for an audiologic diagnostic evaluation was defined as the time from the hearing rescreening at an outpatient clinic to the time when the audiologic diagnosis was completed. The following children were excluded from the study: children whose mothers were not Louisiana residents at birth, children who died after hearing screening regardless of receiving any follow-up, or children who were reported as lost to follow-up (LTF; i.e., testing providers reported children did not show up at the time of the scheduled follow-up appointment, the family was unable to be contacted, or was contacted but unresponsive) or lost to documentation (LTD; i.e., the Louisiana [LA] EHDI program did not receive any report or documentation of follow-up or LTF from audiologists or physicians).

Figure 1: Louisiana Early Hearing Detection and Intervention (LA EHDI) Process of Screening, Diagnosis, and Intervention

Note. ENT = Ear Nose Throat; GBYS = Guide By Your Side (a program providing family support services); LA EHDI = Louisiana early hearing detection and intervention; PCP = Primary Care Physician.
Data sources and linkages

Three data sets were linked and used for analyses including: 2011-2013 birth certificates, 2011–2013 newborn hearing screening (NHS), and 2011–Sept 2014 hearing screening follow-up (after NHS). NHS data provided screening status, screening date, and screening methods. Screenings defined as failed or passed were dependent upon results of final tests before hospital discharge using either auditory brainstem response (ABR) or otoacoustic emissions (OAE). Follow-up data provided follow-up status, time at follow-up, and number of follow-ups.

The data linkages were conducted using SAS 9.3 and LinkPro 3.0. First, failed NHS data were linked to birth certificates by child’s date of birth, first name, and last name with Sounex codes (codes of names based on the phonetic spelling of the name). Linked records were reviewed manually to define true matches using linking variables and the following variables: mother’s last name, first name, maiden name, address of residence at birth or most updated address of residence, and birthing hospital. Second, the NHS-birth records matched data were linked to follow-up data by a unique identification number assigned by the LA EHDI database system. The match rate was 99.2% and 100% for the first and second linkage, respectively.

Analysis variables

Outcome variable. The outcome variable was classified as incomplete or complete audiologic diagnosis. An incomplete audiologic diagnosis (IAD) was defined if an infant (a) failed all newborn hearing screenings before hospital discharge, (b) completed at least one follow-up conducted by an audiologist or physician, and (c) did not have a conclusive audiologic diagnosis confirmed by an audiologist at the time of the study. As mentioned before, all follow-ups mentioned in the study were conducted at outpatient clinics by audiologists or physicians if the follow-up was for rescreening, and only by audiologists for audiological diagnosis. The first follow-up was always a hearing rescreening which marked the beginning of the diagnostic evaluation in this study. A complete audiologic diagnosis was defined, at the last follow-up, if an infant passed both ears with rescreening tests, if diagnostic findings indicated hearing threshold levels within normal limits, or if permanent hearing loss was confirmed. There was no specific time limit applied in definition of the study outcome variable. The LA EHDI program follows hearing status from birth to five years of age. At the time the study was conducted (September 2014), children with IAD were still in process of hearing loss diagnosis but had not yet had a conclusive audiologic diagnosis from the last follow-up between 2011 and 2014.

Predictor variables. There were three predictor variables used: age at NHS, time between NHS and first follow-up, and total number of follow-ups.

Age at NHS. Age in days was calculated using date of birth and date of newborn hearing screening prior to hospital discharge. If there was more than one screening, the date of the last screening was used for calculation. Age was categorized into < 30 days and > 30 days.

Time between NHS and first follow-up. The length of time between date of NHS and date of first follow-up was calculated. If there was more than one screening prior to hospital discharge, the date of the last screening was used for calculation. The time was grouped into < 30 days and > 30 days.

Total number of follow-ups. A sum of all follow-ups that an infant completed. The variable was grouped into one and more than one follow-up.

Covariates

All covariates were derived from birth certificates and defined as categorical variables. Birth weight was categorized into very low birth weight (VLBW, < 1,500 g), low birth weight (LBW, 1,500 g–2,499 g), and normal birth weight (> 2,500 g). Child’s neonatal intensive care unit (NICU) admission was not included as another covariate in the study. NICU admission was considered underreported and the length of time of stay was not reported in birth certificates. However, this variable had a strong collinear relationship with birth weight because

<table>
<thead>
<tr>
<th>Table 1. Population Characteristics (%) by Age at Newborn Hearing Screening (NHS), Time Between NHS and First Follow-up, and Total Number of Follow-ups</th>
<th>Age at Newborn hearing screening (days)</th>
<th>Time between NHS and first follow-up (days)</th>
<th>Total Number of follow-ups</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>&lt;30</td>
<td>30+</td>
<td>p value</td>
</tr>
<tr>
<td>Race/Ethnicity</td>
<td>NH White</td>
<td>45.9</td>
<td>50.6</td>
</tr>
<tr>
<td>NH Black</td>
<td>42.2</td>
<td>56.6</td>
<td>.0001</td>
</tr>
<tr>
<td>NH Other</td>
<td>4.0</td>
<td>2.3</td>
<td>3.7</td>
</tr>
<tr>
<td>Hispanic</td>
<td>7.9</td>
<td>3.8</td>
<td>7.2</td>
</tr>
<tr>
<td>Maternal age</td>
<td>&lt;20</td>
<td>11.6</td>
<td>9.4</td>
</tr>
<tr>
<td>20-34</td>
<td>79.1</td>
<td>78.1</td>
<td></td>
</tr>
<tr>
<td>&gt;34</td>
<td>9.3</td>
<td>12.5</td>
<td></td>
</tr>
<tr>
<td>Maternal Education</td>
<td>&lt; High school (33%)</td>
<td>21.1</td>
<td>21.9</td>
</tr>
<tr>
<td>High school (33%)</td>
<td>31.5</td>
<td>29.8</td>
<td>30.8</td>
</tr>
<tr>
<td>&gt;High school (33%)</td>
<td>47.4</td>
<td>48.3</td>
<td>49.3</td>
</tr>
<tr>
<td>Married at birth</td>
<td>No</td>
<td>58.3</td>
<td>55.8</td>
</tr>
<tr>
<td>Yes</td>
<td>41.7</td>
<td>44.2</td>
<td>42.8</td>
</tr>
<tr>
<td>Delivery method</td>
<td>Vaginal</td>
<td>66.3</td>
<td>30.6</td>
</tr>
<tr>
<td>C-Section</td>
<td>33.7</td>
<td>69.4</td>
<td>33.4</td>
</tr>
<tr>
<td>Delivery pay- ment method</td>
<td>Non-Medicaid Medicaid</td>
<td>29.3</td>
<td>32.5</td>
</tr>
<tr>
<td>70.7</td>
<td>67.5</td>
<td>69.9</td>
<td>72.2</td>
</tr>
<tr>
<td>Area of residence</td>
<td>Urban</td>
<td>33.5</td>
<td>30.4</td>
</tr>
<tr>
<td>Rural</td>
<td>64.7</td>
<td>66.0</td>
<td>63.5</td>
</tr>
<tr>
<td>Previous live birth</td>
<td>None</td>
<td>41.2</td>
<td>44.2</td>
</tr>
<tr>
<td>One</td>
<td>31.1</td>
<td>28.5</td>
<td>30.4</td>
</tr>
<tr>
<td>Two+</td>
<td>27.7</td>
<td>27.3</td>
<td>26.4</td>
</tr>
<tr>
<td>Sex</td>
<td>Female</td>
<td>58.3</td>
<td>54.3</td>
</tr>
<tr>
<td>Male</td>
<td>41.7</td>
<td>45.7</td>
<td>41.4</td>
</tr>
<tr>
<td>Plurality</td>
<td>Singleton</td>
<td>97.2</td>
<td>82.6</td>
</tr>
<tr>
<td>Twin</td>
<td>2.8</td>
<td>17.4</td>
<td>2.6</td>
</tr>
<tr>
<td>Birth weight</td>
<td>VLBW</td>
<td>0.7</td>
<td>0.6</td>
</tr>
<tr>
<td>LBW</td>
<td>8.7</td>
<td>10.6</td>
<td>7.6</td>
</tr>
<tr>
<td>Normal Weight</td>
<td>90.7</td>
<td>9.8</td>
<td>90.1</td>
</tr>
<tr>
<td>Age at newborn screening</td>
<td>&lt;30 Days</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>30+ Days</td>
<td>-</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>Time between NHS and first follow-up</td>
<td>&lt;30 Days</td>
<td>68.3</td>
<td>44.0</td>
</tr>
<tr>
<td>30 Days</td>
<td>31.7</td>
<td>56.0</td>
<td></td>
</tr>
<tr>
<td>Total number of follow-ups</td>
<td>One</td>
<td>86.9</td>
<td>63.4</td>
</tr>
<tr>
<td>Two+</td>
<td>13.1</td>
<td>36.6</td>
<td>11.5</td>
</tr>
</tbody>
</table>

*pChi-square p value. Note: NH: Non-Hispanic; VLBW = very low birth weight; LBW = low birth weight
All VLBW babies were admitted to NICU. Thus, presence of NICU admission in adjusted regressions of data analyses was not necessary. Table 1 shows distributions of all covariates by age at NHS, time between NHS and first follow-up, and total number of follow-ups.

**Data analysis**

Percentages and 95% confidence intervals of IAD by predictors were calculated. Confidence intervals were estimated by using the normal approximation method of the binomial confidence interval. Logistic regression models were used to determine associations between IAD and predictors. To address confounding in adjusted regression models all covariates were controlled. In fact, all VLBW babies are admitted into the NICU, typically for extended stays, and therefore have late newborn hearing screening. Because of VLBW newborns’ long-term NICU stay and medical characteristics that are very different from other groups (low birth weight and normal birth weight), effects of predictors on IAD were evaluated in each group and also compared together among different groups of birth weight by including interaction terms between birth weight and predictors in models. Specifically, there were three analyses using logistic regression models to assess the associations described as follows:

**Association of IAD with age at NHS.** In the unadjusted model, the independent variables consisted of age at NHS, birth weight, and the interaction between age at NHS and birth weight. All covariates were added in adjusted model.

**Association of IAD with length of time between NHS and first follow-up.** In the unadjusted model, the independent variables consisted of time between NHS and first follow-up, birth weight, and the interaction between time between NHS and first follow-up and birth weight. All covariates plus age at NHS were added in the adjusted model.

**Association of IAD with number of follow-up.** In the unadjusted model, the independent variables consisted of number of follow-ups, birth weight, and the interaction between number of follow-ups and birth weight. All covariates plus age at NHS and time between NHS and first follow-up were added in the adjusted model.

All final adjusted models included only variables with p value < 0.05. Data analyses were conducted in SAS 9.3.

The project was deemed exempt by the Louisiana State University Institutional Review Board because it did not meet the federal definition of human subjects research.

**Results**

There were 6,970 children included in the study. A majority of children (96.2%) completed NHS before 30 days of age and completed one follow-up (86.1%). The percent of children who completed the first follow-up before 30 days after NHS was 67.4%.

The overall rate of IAD was 6.9% (CI: 6.3–7.5). The rate was very high among newborns with NHS at 30 days of age or older (25.7%) compared to those with NHS within 30 days of age (6.1%). Stratified by birth weight, this difference was also seen among newborns with low birth weight or normal weight (LBW: 8.1% for age at NHS < 30 days vs. 25.0% for age at NHS > 30 days; normal weight: 5.8% for age at NHS < 30 days vs. 26.9% for age at NHS > 30 days). However, among newborns with VLBW, the rate was very high in both age groups and was not statistically different (20.5% for age at NHS < 30 days vs. 25.6% for age at NHS > 30 days; t(6959) = 0.72, p = 0.4734).

For the length of time between NHS and first follow-up, the rate of IAD was 5.0% with the length < 30 days and it doubled with the length > 30 days (10.5%). Stratified by birth weight, this difference was seen among newborns with LBW and normal weight (LBW: 6.8% for the length < 30 days vs. 11.9% for the length > 30 days; normal weight: 4.3% for the length < 30 days vs. 9.4% for the length > 30 days). Similar to age at NHS, among babies with VLBW the rate was very high in both groups and was not statistically different (26.4% for the length < 30 days vs. 23.2% for the length > 30 days; t(6,929) = -0.57, p = 0.5682).

For the number of follow-ups, the rate of IAD was 4.8% among newborns with one follow-up and it was almost four times higher among those who had more than one follow-up (19.6%). Stratified by birth weight, the rate was high and statistically different between groups among newborns with VLBW, LBW, and normal weight: VLBW: 20.1% for one vs.

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**Table 2. Percentage of Incomplete Audiologic Diagnosis by Age at Newborn Hearing Screening (NHS), Time between NHS and First Follow-up, and Total Number of Follow-ups Stratified by Birth Weight.**

<table>
<thead>
<tr>
<th>Birth Weight</th>
<th>Age at newborn hearing screening (days)</th>
<th>Time between NHS and first follow-up (days)</th>
<th>Total number of follow-ups</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>n</td>
<td>Percent, 95% CI</td>
<td>n</td>
</tr>
<tr>
<td>VLBW</td>
<td>&lt;30</td>
<td>9</td>
<td>29.5, 8.5–32.4</td>
</tr>
<tr>
<td></td>
<td>30+</td>
<td>54</td>
<td>25.6, 19.7–31.5</td>
</tr>
<tr>
<td></td>
<td>Total</td>
<td>63</td>
<td>24.7, 19.4–30.0</td>
</tr>
<tr>
<td>LBW</td>
<td>&lt;30</td>
<td>47</td>
<td>8.1, 5.9–10.3</td>
</tr>
<tr>
<td></td>
<td>30+</td>
<td>7</td>
<td>25.0, 9.0–41.0</td>
</tr>
<tr>
<td></td>
<td>Total</td>
<td>54</td>
<td>9.8, 6.6–11.1</td>
</tr>
<tr>
<td>Normal Weight</td>
<td>&lt;30</td>
<td>354</td>
<td>9.8, 5.2–6.4</td>
</tr>
<tr>
<td></td>
<td>30+</td>
<td>7</td>
<td>26.9, 9.9–44.0</td>
</tr>
<tr>
<td></td>
<td>Total</td>
<td>361</td>
<td>5.9, 5.3–6.5</td>
</tr>
</tbody>
</table>

Note: VLBW = very low birth weight; LBW = low birth weight.
31.4% for more than one follow-up ($t(6,962) = 2.03, \ p = 0.0423$); LBW: 6.8% for one vs. 18.5% for more than one follow-up ($t(6,962) = 3.74, \ p = 0.0002$); and normal weight: 4.2% for one vs. 8.1% for more than one follow-up ($t(6,962) = 14.0, \ p < 0.0001$). Table 2 presents percentage of IAD by age at NHS, time between NHS and first follow-up, and total number of follow-ups stratified by birth weight.

Adjusted regression models showed that associations of IAD with the predictors varied among birthweight groups. The interactions were significant between birthweight and age at NHS ($F(2, 6,863) = 3.13, \ p = 0.0439$); length of time between NHS and first follow-up ($F(2, 6,859) = 5.37, \ p = 0.0047$); and number of follow-ups ($F(2, 6,858) = 4.59, \ p = 0.0101$). Among VLBW newborns, there were no statistical associations of IAD with age at NHS (Odds Ratio [OR]: 1.3, CI: 0.6–3.0) or the length between NHS and first follow-up (OR: 0.8, CI: 0.5–1.2); however, the association of IAD with number of follow-ups was found (OR: 1.9, CI: 1.0–3.4). Among newborns with LBW, odds of IAD was significantly higher in babies with NHS > 30 days of age (OR: 3.8, CI: 1.5–9.4); the length of time between NHS and first follow-up > 30 days (OR: 2.3, CI: 1.9–2.9); and more than one follow-up (OR: 4.7, CI: 3.4–6.0). See Table 3.

### Table 3: Odds Ratio (OR) Estimates and 95% Confidence Interval (CI) for Associations between Incomplete Audiologic Diagnosis and Age at Newborn Hearing Screening (NHS), Time between NHS and First Follow-up, and Total Number of Follow-ups

<table>
<thead>
<tr>
<th>Age at newborn hearing screening (days)</th>
<th>Unadjusted</th>
<th>Adjusted</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>OR, 95% CI</td>
<td>t statistic</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Very low birth weight &lt;30</td>
<td>1.0</td>
<td>0.72</td>
</tr>
<tr>
<td>Very low birth weight 30+</td>
<td>1.3, 0.6–3.0</td>
<td>0.54</td>
</tr>
<tr>
<td>Low birth weight &lt;30</td>
<td>1.0</td>
<td>4.00</td>
</tr>
<tr>
<td>Low birth weight 30+</td>
<td>3.8, 1.5–9.4</td>
<td>2.99</td>
</tr>
<tr>
<td>Normal birth weight &lt;30</td>
<td>1.0</td>
<td>6.0, 2.5–14.3</td>
</tr>
<tr>
<td>Normal birth weight 30+</td>
<td>6.0, 2.5–14.3</td>
<td>3.49</td>
</tr>
<tr>
<td>Time between NHS and first follow-up</td>
<td>Unadjusted</td>
<td>Adjusted</td>
</tr>
<tr>
<td></td>
<td>OR, 95% CI</td>
<td>t statistic</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Very low birth weight &lt;30</td>
<td>1.0</td>
<td>0.57</td>
</tr>
<tr>
<td>Very low birth weight 30+</td>
<td>0.8, 0.5–1.5</td>
<td>0.8, 0.4–1.5</td>
</tr>
<tr>
<td>Low birth weight &lt;30</td>
<td>1.0</td>
<td>2.14</td>
</tr>
<tr>
<td>Low birth weight 30+</td>
<td>1.9, 1.1–3.2</td>
<td>2.00</td>
</tr>
<tr>
<td>Normal birth weight &lt;30</td>
<td>1.0</td>
<td>7.61</td>
</tr>
<tr>
<td>Normal birth weight 30+</td>
<td>2.3, 1.9–2.9</td>
<td>7.54</td>
</tr>
<tr>
<td>Total number of follow-ups</td>
<td>Unadjusted</td>
<td>Adjusted</td>
</tr>
<tr>
<td></td>
<td>OR, 95% CI</td>
<td>t statistic</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Very low birth weight One</td>
<td>1.0</td>
<td>2.03</td>
</tr>
<tr>
<td>Very low birth weight Two+</td>
<td>1.8, 1.0–3.2</td>
<td>2.04</td>
</tr>
<tr>
<td>Low birth weight One</td>
<td>1.0</td>
<td>3.74</td>
</tr>
<tr>
<td>Low birth weight Two+</td>
<td>3.1, 1.7–5.7</td>
<td>3.39</td>
</tr>
<tr>
<td>Normal birth weight One</td>
<td>1.0</td>
<td>14.00</td>
</tr>
<tr>
<td>Normal birth weight Two+</td>
<td>5.1, 4.0–6.4</td>
<td>13.00</td>
</tr>
</tbody>
</table>

Discussion

This study showed that children with late NHS (>30 days of age), late follow-up (>30 days after NHS), and multiple follow-ups were more likely not to complete the audiologic diagnosis process. Effects of age at NHS, timing of follow-up, and number of follow-ups on IAD varied among birth weight groups. With the presence of VLBW, the rate of IAD was very high (>20%) regardless of time when NHS and first follow-up were completed or number of follow-ups, and associations of IAD with age at NHS and length of time between NHS and first follow-up did not exist. However, the association was seen with number of follow-ups. For LBW or normal weight groups, the rate was consistent between two groups and higher among those who had late NHS, late follow-up, and multiple follow-ups. The risk difference of IAD between groups of the predictors was fairly similar among LBW and normal weight newborns and larger than one among VLBW newborns.

It was clear that VLBW had a strong effect on IAD as well as late NHS, late follow-up, and multiple follow-ups. The main reason of late NHS was VLBW. The data showed that more than 80% of VLBW babies had NHS after 30 days. VLBW babies who normally have severe medical conditions often have long-term hospital stays, particularly in the NICU where procedures of medical stabilization are required (Berry, Shah, Brouillette, & Hellmann, 2008), thus the NHS is delayed until just prior to initial discharge. For parents of those babies, appointments for medical conditions may take priority over hearing follow-up appointments. Thus, hearing follow-up appointments could be missed, ignored, or delayed which leads to IAD or late diagnosis. In fact, VLBW newborns are more likely to get many follow-ups. Those babies are harder to test due to very small ear canals. In addition, a very small head sometimes makes it harder to obtain results on bone conduction testing. Thus, more tests are needed before confirming the diagnosis. Table 2 presents the difference of late NHS, late follow-up, and number of follow-ups by birth weight.

The study indicated that when stratified by birth weight, age at NHS, timing of follow-up, and number of follow-ups had different effects on IAD. Specifically, LBW or normal weight babies with late NHS, late follow-up, and multiple follow-ups had a higher risk of IAD. Although the associations were well defined in LBW and normal weight babies, their underlying mechanisms were not clear. Therefore, further research is needed to understand the
mechanisms of associations as well as reasons of late NHS, late follow-up, and multiple follow-ups among non-VLBW babies, particularly among normal weight babies which consisted of about 90% of the total study population. A large reduction of late follow-up and multiple follow-ups in normal weight babies would have a significant impact on a decrease of late follow-up and multiple follow-ups as well as an improvement of IAD in the whole study population. The following factors may be some of the possible reasons of late follow-up in normal weight babies who have fewer medical conditions: inaccessibility to follow-up facilities or providers; lack of transportation, particularly in rural areas; lack of health insurance; lack of parents’ knowledge and awareness of the importance of early diagnosis of hearing loss (Shulman et al., 2010); and overwhelming parental responsibilities (Folsom et al., 2000; Lui, Farrell, MacNeil, Stone, & Barfield, 2008). Some of the main reasons for multiple follow-ups may be a lack of well-trained pediatric audiologists and physicians who provide follow-up testing or a lack of facilities that provide sedated diagnostic testing (Shulman et al., 2010). In fact, the sleeping or quiet state of the infant, particularly for those younger than three months old, is necessary in the early stages of testing and diagnosis to avoid the need for sedation (National Center for Hearing Assessment and Management, 2012). If the sleeping or quiet state is not attained sufficiently, untrained or inexperienced providers may recommend rescheduling another visit or referring to another facility that can conduct sedated hearing testing for diagnosis. Parents of babies with many follow-ups may become frustrated with the continual re-testing with no conclusion and lose confidence in the follow-up provider and the facility. This assumption may explain why some parents did not follow through with subsequent appointments and the audiologic diagnosis evaluation was not completed. Figure 2 summarizes possible reasons of late newborn hearing screening, late follow-ups, and multiple follow-ups as pathways leading to incomplete audiologic diagnosis.

Strengths and Limitations
This study had three major strengths: First, covariates including mother and child characteristics collected in birth certificates were captured through data linkages. These characteristics were controlled for in adjusted regression models to evaluate independent effects of predictors. Second, the study displayed an important role of VLBW, a strong confounder, in contributing to late NHS, late follow-up, and multiple follow-up appointments as well as IAD. Last, independent effects of the predictors were evaluated among different birth weight categories, which excluded a direct effect of VLBW on predictors in evaluating associations among LBW and normal weight newborns.

The study included two major limitations: First, underlying mechanisms to explain associations were limited, particularly among LBW and normal weight newborns; therefore, more studies are needed. Second, the incomplete audiologic diagnosis status was not verified through contacting parents or follow-up facilities. Verification may improve underreporting problems and avoid misclassification of the study outcome as well as bias of study results.

Conclusions
In order to reduce IAD, it may be necessary to conduct NHS early, expedite follow-up, and decrease the number of follow-up visits. Severe medical conditions, particularly VLBW, majorly contributed to late NHS that increased risk of IAD. Efforts to reduce severe medical conditions by enhancing the quality of prenatal and obstetrical care could help prevent both NICU admission and prolonged hospitalization, and thus reduce late NHS prior to hospital discharge (Gregory, Jackson, Korst, & Fridman, 2012; Lu, Kotelchuck, Hogan, Johnson, & Reyes, 2010; Newnham et al., 2014; Sakala, Yang, & Corry, 2013). To reduce the risk of IAD due to late NHS, screening should be conducted as early as possible during the NICU stay as medical conditions allow instead of waiting until hospital discharge.
Currently, based on the JCIH Position Statement 2007, all infants admitted to the NICU should be screened for hearing loss before hospital discharge. Although only 10-15% of the newborn population spends time in the NICU, this population has a higher risk of hearing loss and in particular, neural hearing loss (auditory neuropathy spectrum disorder; D’Agostino & Austin, 2004; Starr, Sininger, & Pratt, 2000). Therefore, not only screening but also the diagnostic process should be completed prior to discharge for newborns with severe medical conditions or those with prolonged hospitalizations, particularly in NICU, if at all possible. An increased number of sedated hearing diagnostic testing facilities and follow-up providers with significant pediatric experience, may reduce referral to other facilities and the number of follow-up appointments.

To understand mechanisms of the associations and reasons of late NHS, late follow-up, and multiple follow-ups, particularly among non-VLBW newborns, further in-depth quality improvement studies are needed. Through such studies, both parents and follow-up facilities should be contacted. Specifically, the studies might target the following: parents’ knowledge and awareness of the importance of early diagnosis of hearing loss, providers and audiologists’ experience or skill in screening young infants, referral scheduling relying on the parents instead of staff of referring facilities, and miscommunications and unclear referral protocols between referring and receiving facilities.

**References**


How Many Babies with Hearing Loss Will Be Missed by Repeated Newborn Hearing Screening with Transient Evoked Otoacoustic Emissions Due to Statistical Artifact?

Karl R. White¹
Lauri H. Nelson²
Karen Munoz²

¹National Center for Hearing Assessment and Management, Department of Psychology, Utah State University, Logan Utah, USA
²Department of Communicative Disorders and Deaf Education, Utah State University, Logan Utah, USA

Abstract

Objective: It is often said that repeating OAE hearing screening more than two or three times per ear creates statistical artifacts that unacceptably increase false-negatives (i.e., passing babies who have permanent hearing loss). This study evaluated the accuracy of that recommendation for screening with transient evoked otoacoustic emissions (TEOAE).

Design: The false negative rate was estimated using a 2.0 cc coupler and three human ears with moderate or worse hearing loss. Using those results and the prevalence of hearing loss among newborns, the number of babies with hearing loss that would be missed due to repeated testing was calculated.

Results: Only 1% of ears with moderate or worse hearing loss will be missed due to statistical probability of false-negatives resulting from repeated testing.

Conclusions: Excessive repeated testing in a newborn hearing screening program wastes time; raises questions about accuracy of screening; and may disturb the infant, family, or hospital staff. Repeated TEAOE testing does not cause statistical artifacts that result in a significant number of babies with hearing loss to pass the screening test. Not repeating screening tests often enough may needlessly inflate the number of babies referred for diagnostic testing and create financial burdens and worry for families.

Acronyms: ABR = Auditory Brainstem Response, EHDI = Early Hearing Detection and Intervention, JCIH = Joint Committee on Infant Hearing, NBHS = Newborn Hearing Screening, OAE = Otoacoustic Emissions, TEOAE = Transient-Evoked Otoacoustic Emissions

Corresponding author’s contact information: Karl R. White, National Center for Hearing Assessment and Management, Utah State University, 2615 Old Main Hill, Logan, Ut 84322; Email: karl.white@usu.edu

Introduction

Otoacoustic emissions (OAE) testing is used worldwide in hospitals and clinics to test cochlear function of individuals in all age groups. The most common use of OAE testing is in hospital-based newborn hearing screening programs as an objective measure to identify infants who require additional diagnostic audiologic testing to confirm the presence or absence of hearing loss. The screening test is often performed with a hand-held unit that measures the presence or absence of an OAE in response to an auditory stimulus (Kemp, 1978). Screening is done by placing a small probe in the ear canal that delivers a low-intensity signal to the structures of the cochlea in the inner ear. If the cochlea is functioning normally, the outer hair cells of the cochlea respond by producing an otoacoustic emission, sometimes described as an echo, that travels back through the middle ear and the ear canal and is detected by the screening unit (NCHAM, 2011). There is widespread agreement that doing hearing screening with OAE testing is reliable, harmless, and effective (e.g., ASHA, 2004; JCIH, 2007; Keppler, Dhooge, & Maes, 2010; White, 2014).

Currently, every state in the United States has implemented either a mandatory or voluntary newborn hearing screening (NBHS) program. Many of these programs use OAE screening equipment (White, 2014) due to the safety and ease-of-use. Every state-based Early Hearing Detection and Intervention (EHDI) program has a coordinator who collaborates with stakeholders in the state to implement and support effective newborn hearing screening programs. In 1995, the percentage of newborns screened for hearing loss was just 3%. A decade later that number had increased to 95% (White, 2006; White, Forsman, Eichwald, & Munoz, 2010), largely owing to the ease with which screening could be done and the wide acceptance of reliable and objective screening tools. The Centers for Disease Control and Prevention (CDC; 2015) report that 98% of newborns in the United States are currently screened for hearing loss.

Recommendations Regarding Repeating Newborn Hearing Screening Tests

The Joint Committee on Infant Hearing (JCIH, 2007) provides guidelines for all aspects of pediatric audiological services, such as screening and diagnostic testing protocols and hearing technology management. Many hospital-based newborn hearing screening program coordinators rely on the JCIH recommendations for guidance in developing and managing their programs. Commenting on how often the newborn hearing screening
The recommendation to limit the number of OAE screening tests performed in NBHS programs due to the potential of passing babies who have hearing loss because of the statistical probability of obtaining a false negative response appears to have become accepted as best-practice.

Materials and Method

To estimate how many babies with moderate or worse hearing loss are likely to be missed because of repeated newborn hearing screening tests, it is necessary to estimate the false negative rate of OAE screening (i.e., the probability of passing an ear with known hearing loss). Unfortunately, none of the manufacturers of the equipment used for newborn hearing screening provide such information. Consequently, this study estimated the false negative rate for a single test using the Biologic AuDx® Pro OAE Screener. Because the false negative rate could be different for other brands and types of screening equipment (e.g., Biologic versus Otodynamics, or OAE versus automated auditory brainstem response, or transient evoked versus distortion product otoacoustic emissions), the results reported here represent a starting point for addressing questions about the frequency of false-negatives attributable to statistical artifact in hearing screening programs, but these results are not the complete answer. We have demonstrated the consequences of repeated newborn hearing screening tests using TEOAEs with one of the most frequently used OAE screeners. Making similar estimates for other brands or types of screening instruments will require additional data collection. The data collection described below was approved by the Institutional Review Board at Utah State University.

Participants

To estimate the false negative rate of TEOAE testing, two participants with bilateral moderate sloping to severe-profound hearing loss provided informed consent to have repeated TEOAE tests. Audiograms for each of three ears are shown in Figure 1. One thousand transient evoked OAE (TEOA) tests were obtained from the left ear of the first participant and in both ears of the second participant for a total of 3,000. Additionally, 1,000 TEOAEs were collected using a 2.0 cc coupler.

Equipment and Procedures

Using the Biologic AuDx® Pro OAE screener, all screening tests were completed with the TEOAE screening default test parameters (see Table 2). TEOAEs were selected for this study due to their common usage in NBHS programs and their high sensitivity and specificity in detecting outer hair cell dysfunction (Cunningham, 2011; Keppler et al., 2010; Lapsley-Miller & Marshall, 2001).

Data were collected in a quiet room on each ear over a 2-week period, averaging approximately 200 tests per day. Within each data collection time period, the probe was securely placed into the canal of the ear being tested and remained in place throughout the test session. The probe was not removed and then re-fitted after each individual TEOAE test. Data were collected under the supervision of a licensed audiologist.

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Table 1. Examples of Statements from State EHDI Programs and Others about Repeating Newborn Hearing Screening Tests

<table>
<thead>
<tr>
<th>Statement</th>
<th>Source</th>
</tr>
</thead>
<tbody>
<tr>
<td>The initial hearing screening . . . should consist of no more than 2 attempts using the same screening technique on each ear.</td>
<td>(Washington State EHDI Program Guidelines, 2015)</td>
</tr>
<tr>
<td>For infants who fail the initial screen, hospitals should attempt to re-screen the infant prior to discharge. Inpatient hearing screening will consist of no more than two attempts using the same screening technique on each ear, assuming the infant is in an appropriate state for testing and there are neither equipment problems nor environmental interference during the test. The likelihood of obtaining a pass by chance alone is increased when screening is performed repeatedly.</td>
<td>(Minnesota State EHDI Program Guidelines, 2015)</td>
</tr>
<tr>
<td>… take caution to avoid over-screening newborns! Although there may be factors that require the screen to be repeated, it is not recommended that babies be screened more than three times.</td>
<td>(Connecticut Department of Health, 2015)</td>
</tr>
<tr>
<td>… excessive re-screening can increase the false negative rate (passing babies with actual hearing loss) … Two screening sessions of no more than three screens per ear are recommended, for a total of six screens per ear.</td>
<td>(Iowa EHDI Program Guidelines, 2015)</td>
</tr>
<tr>
<td>“Do not screen patient more than three times per ear. Over screening can result in a false negative result.”</td>
<td>(Welch Allyn OAE Hearing Screener Quick Reference Guide, 2015)</td>
</tr>
<tr>
<td>Screening too many times isn’t recommended and it can lead to false results, … Your goal is not to pass every baby. “With multiple screenings, babies with hearing loss may falsely pass.”</td>
<td>(Newborn Hearing Screening Training Curriculum, NCHAM, 2015)</td>
</tr>
</tbody>
</table>

Note: Emphasis added. EHDI = Early Hearing Detection and Intervention, OAE = optoacoustic emissions.
Figure 1. Audiograms for Ears Used to Estimate False-Negative Rate of Transient-Evoked Otoacoustic Emission (TEOAE) Hearing Screening Tests. Different subscripts indicate different people.

Table 2. Bio-logic AuDx® TEOAE System Default Protocol

<table>
<thead>
<tr>
<th>Parameter</th>
<th>Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Number of frequencies for overall pass</td>
<td>0</td>
</tr>
<tr>
<td>Checkfit trials</td>
<td>10</td>
</tr>
<tr>
<td>Number of successes to pass</td>
<td>1</td>
</tr>
<tr>
<td>Number of checkfit failures until refit</td>
<td>7</td>
</tr>
<tr>
<td>Checkfit/calibration artifact rejection</td>
<td>250</td>
</tr>
<tr>
<td>Minimum percent probe stability</td>
<td>95</td>
</tr>
<tr>
<td>Start time (ms)</td>
<td>3.50</td>
</tr>
<tr>
<td>Ramp time (ms)</td>
<td>0.98</td>
</tr>
<tr>
<td>Number of samples per set</td>
<td>3</td>
</tr>
<tr>
<td>Calibration trials</td>
<td>10</td>
</tr>
<tr>
<td>Number of calibration successes to pass</td>
<td>1</td>
</tr>
<tr>
<td>Number of calibration failures until refit</td>
<td>7</td>
</tr>
<tr>
<td>Maximum number of samples</td>
<td>512</td>
</tr>
<tr>
<td>Target amplitude (dB SPL)</td>
<td>80</td>
</tr>
<tr>
<td>End time (ms)</td>
<td>12.0</td>
</tr>
<tr>
<td>Artifact reject (mPa)</td>
<td>20</td>
</tr>
</tbody>
</table>

*Note.* TEOAE = transient-evoked otoacoustic emission.
Analysis and Results
To estimate the number of babies with moderate or worse hearing loss that would be missed due to repeated testing two pieces of information are needed: a) an estimate of the false negative rate of OAE testing due to statistical artifact; and, b) the incidence of congenital hearing loss.

False Negative Rate of OAE Testing
The false negative rate for OAE screening is the number of times a pass result is obtained for an ear that has hearing loss. Of the three ears with hearing loss that were tested 1,000 times, one ear had 999 fails, a second had 1,000 fails, and the third had 998 fails. Testing with the 2.0 cc coupler had similar results with 1,000 fails. Based on these results, the false negative rate for this piece of TEOAE screening equipment was estimated to be 1 per 1,000. The fact that the false negative rate was based on adult ears instead of infant ears is a limitation. However, the authors of the study decided that it was not practical or appropriate to repeat a screening test 1,000 times on a newborn. If the false negative rate for newborns is substantially higher for newborns than for adults, the results would be different. However, as discussed below, even in the unlikely event that the false negative rate for newborns is ten times as high as the rate estimated for adults, it does not change the basic conclusions of this study.

Prevalence of Congenital Hearing Loss
In the latest data available, staff at state-based EHDI programs reported an average of 1.5 babies per 1,000 with permanent hearing loss (CDC, 2015). However, as noted by White (2014) this number is likely a low estimate of the number of babies with congenital hearing loss due to high rates of loss to follow-up in many states and inefficient newborn hearing screening programs and/or poor documentation in some states. White (2014) suggested that a better estimate is 3.0 per 1,000 births. For this study, the higher number for the incidence of congenital hearing loss was used to estimate a worst case scenario of how many babies with hearing loss were likely to be missed due to repeated TEOAE testing.

Analyses
In calculating the number of ears with permanent hearing loss that are likely to be missed due to repeated screening, we must first focus on only those ears that have hearing loss, because it is impossible to “miss” ears that have normal hearing. If 10,000 ears with hearing loss were tested with the probability of an accurate test being 0.9990 as estimated above, ten ears with permanent hearing loss would be missed as shown in the first row of Table 3.

Table 3. Number of Ears with Permanent Hearing Loss in a General Population Sample of 100,000 that are Missed Due to Repeated Screening Tests if False Negative Rate is 1 per 1,000.

<table>
<thead>
<tr>
<th>Probability of an ear with hearing loss failing the test</th>
<th># of screening tests</th>
<th>False negatives per 10,000 ears with hearing loss</th>
<th>False negatives per 10,000,000 newborns ears in the general population</th>
<th>False negatives per 100,000 newborn ears in the general population</th>
<th>% of “missed” newborn ears with hearing loss</th>
</tr>
</thead>
<tbody>
<tr>
<td>0.9990</td>
<td>1</td>
<td>10.0</td>
<td>30.0</td>
<td>0.30</td>
<td>0.10%</td>
</tr>
<tr>
<td>0.9980</td>
<td>2</td>
<td>20.0</td>
<td>60.0</td>
<td>0.60</td>
<td>0.20%</td>
</tr>
<tr>
<td>0.9970</td>
<td>3</td>
<td>30.0</td>
<td>89.9</td>
<td>0.90</td>
<td>0.30%</td>
</tr>
<tr>
<td>0.9960</td>
<td>4</td>
<td>39.9</td>
<td>119.8</td>
<td>1.20</td>
<td>0.40%</td>
</tr>
<tr>
<td>0.9950</td>
<td>5</td>
<td>49.9</td>
<td>149.7</td>
<td>1.50</td>
<td>0.50%</td>
</tr>
<tr>
<td>0.9900</td>
<td>10</td>
<td>99.6</td>
<td>298.7</td>
<td>2.99</td>
<td>1.00%</td>
</tr>
<tr>
<td>0.9851</td>
<td>15</td>
<td>149.0</td>
<td>446.9</td>
<td>4.47</td>
<td>1.49%</td>
</tr>
<tr>
<td>0.9802</td>
<td>20</td>
<td>198.1</td>
<td>594.3</td>
<td>5.94</td>
<td>1.98%</td>
</tr>
<tr>
<td>0.9753</td>
<td>25</td>
<td>247.0</td>
<td>741.1</td>
<td>7.41</td>
<td>2.47%</td>
</tr>
</tbody>
</table>
The incidence of a missed hearing loss if every ear is tested once is obtained by multiplying the incidence of a false negative in the population of ears with hearing loss (10 per 10,000) by the incidence of hearing loss in the general population (3 per 1,000). Thus, the incidence of missed ears if only one screening test were done is 30 per 10,000,000. Converting this to a number that is more realistic for state-based EHDI programs, 0.30 ears with hearing loss would be missed for every 100,000 ears in the general population as shown in the first row of the right-most column in Table 3.

But what happens if the screening test is repeated multiple times? As noted in the JCIH (2007, p. 903) position statement, when a test is less than 100% accurate, “the likelihood of obtaining a pass outcome by chance alone is increased when screening is performed repeatedly.” In this case, it was estimated that the test is only 99.9% accurate, so there is no question that the likelihood of a false negative will be increased—but by how much and is it enough to be concerned?

The probability of a false negative result due to statistical artifacts of repeated testing is estimated by multiplying the accuracy of each test in the series and subtracting the result from 1.0. Thus, the probability of a false negative for two tests is:

\[ 1 - (0.999 \times 0.999) = 0.998. \]

The probability when three tests are given is:

\[ 1 - (0.999 \times 0.999 \times 0.999) = 0.997. \]

Similar calculations can be done for however many tests are given and selected results are shown in Table 3. The number of ears that would be missed due to statistical artifact if every ear were tested from 2–25 times is very small because the false negative rate of each individual test is only 1 per 1,000 and the incidence of hearing loss among babies is only 3 per 1,000. For example, in a population of 50,000 babies (or 100,000 ears), we would expect 150 babies with permanent hearing loss (3 babies per 1,000 × 50,000 babies). But, if every one of these 50,000 babies were tested ten times in each ear, only 2.99 ears (or about 1.0% of the 300 ears with permanent hearing loss) would be missed due to statistical artifact.

Table 4 shows the number of babies’ ears that would be missed due to statistical artifact if there were 10 times as many false negatives (i.e., 1 per 100 instead of 1 per 1,000). Table 4 is provided to emphasize how unlikely it is that a mistake in estimating the false negative rate per 1,000 would change the basic conclusions of this analysis.

Calculating the number of babies that would be missed due to statistical artifact in the birth cohort of 50,000 requires differentiating between babies with unilateral hearing loss and babies with bilateral hearing loss. According to the CDC (2015), 40% of babies reported in 2013 as having congenital hearing loss were unilateral. Thus, if there were 300 ears with permanent hearing loss missed in the population of 100,000 ears tested, there would be 113 babies with bilateral losses (226 ears) and 74 babies with unilateral losses (74 ears) for a total of 187 babies and 300 ears. If 1% of these ears were missed, it would be one baby with bilateral loss and one with unilateral loss. However, the probability of missing both ears in a baby with bilateral loss due to statistical artifact when one ear is tested right after the other is 1 in 1,000,000 instead of 1 in 1,000 because the probability of two independent events happening in sequence is the product of the probabilities of each of those events happening independently. Thus, the chance of a baby with bilateral hearing loss being missed due to statistical artifact approaches zero because one or the other of the ears would fail the testing and both ears would be identified during follow-up diagnostic testing. Therefore, the only baby missed would be the one with unilateral loss. To summarize, in a birth cohort of 50,000 babies, there would be 150 babies with congenital hearing loss, and 1 baby with unilateral loss (0.67%) would be missed due to the statistical artifact of repeated testing.

<table>
<thead>
<tr>
<th>Probability of an ear with hearing loss failing a test</th>
<th># of screening tests</th>
<th>False negatives per 10,000 ears with hearing loss</th>
<th>False negatives per 10,000,000 newborns ears in the general population</th>
<th>False negatives per 100,000 newborns in the general population</th>
<th>% of “missed” newborns with hearing loss</th>
</tr>
</thead>
<tbody>
<tr>
<td>0.9900</td>
<td>1</td>
<td>100.0</td>
<td>300.0</td>
<td>3.00</td>
<td>1.00%</td>
</tr>
<tr>
<td>0.9801</td>
<td>2</td>
<td>199.0</td>
<td>597.0</td>
<td>5.97</td>
<td>1.99%</td>
</tr>
<tr>
<td>0.9791</td>
<td>3</td>
<td>208.8</td>
<td>626.4</td>
<td>6.26</td>
<td>2.09%</td>
</tr>
<tr>
<td>0.9781</td>
<td>4</td>
<td>218.6</td>
<td>655.8</td>
<td>6.56</td>
<td>2.19%</td>
</tr>
<tr>
<td>0.9772</td>
<td>5</td>
<td>228.4</td>
<td>685.1</td>
<td>6.85</td>
<td>2.28%</td>
</tr>
<tr>
<td>0.9723</td>
<td>10</td>
<td>277.1</td>
<td>831.4</td>
<td>8.31</td>
<td>2.77%</td>
</tr>
<tr>
<td>0.9674</td>
<td>15</td>
<td>325.7</td>
<td>977.0</td>
<td>9.77</td>
<td>3.26%</td>
</tr>
<tr>
<td>0.9626</td>
<td>20</td>
<td>373.9</td>
<td>1121.8</td>
<td>11.22</td>
<td>3.74%</td>
</tr>
<tr>
<td>0.9578</td>
<td>25</td>
<td>422.0</td>
<td>1265.9</td>
<td>12.66</td>
<td>4.22%</td>
</tr>
</tbody>
</table>
Discussion

The positive impact of effective NBHS programs on the linguistic and academic development of children who are deaf or hard of hearing has been well documented (Kennedy et al., 2006; Marge & Marge, 2005; Moeller, 2000; Yshinaga-Itano, Sedey, Coulter, & Mehl, 1998). Although programs should seek to improve screening methods and minimize false OAE test results, the goal of a screening program should not be to pass every baby tested. Instead, programs should identify and implement effective screening protocols and procedures with well-trained personnel so that the results of screening tests are reliable and accurate.

Even though it is estimated that less than 1% of the babies with moderate or worse permanent hearing would be missed due to statistical artifacts, there are a number of other potentially adverse effects if OAE screening tests are repeated too often in a newborn hearing screening program. For example, frequent repetition of OAE screening may:
1. Be an inefficient use of resources because staff are often repeating tests that are unlikely to have different results.
2. Decrease the confidence that health care providers (e.g., nurses, physicians, etc.) and parents have about the efficacy of the NBHS program or the testing process because tests are being repeated so frequently.

Conversely, not repeating the OAE test enough times to get an accurate result can result in an excessive number of false positive results where infants with normal hearing are discharged from the hospital with a failed OAE test result. For example, it is well known that cerumen or other debris in the ear canal of newborns can cause a fail screening result for babies with normal hearing (White, 2014). Such debris often clears after a few hours and a baby with normal hearing who has failed the initial screening will often pass a subsequent screening. Similarly, a baby with normal hearing who is very agitated during a screening test may fail because the probe is not positioned correctly or there is too much noise in the screening environment. Retesting at a later time will often result in an accurate pass result.

If too many babies with normal hearing have failed the screening test when they leave the hospital, overall screening costs increase due to a large number of babies who must be followed and brought back for additional testing. Doing follow-up testing with an unnecessarily high number of infants not only increases costs, but it may cause parents undue alarm and anxiety, undermining confidence in the screening program among all stakeholders (Clemens, Davis, & Bailey, 2000).

The ramifications of over-testing or under-testing illustrate the importance of effective and appropriate screening protocols (Wada, Kubo, Aiba, & Yamane, 2004). In addressing potential program improvements to increase the accuracy of hearing screening procedures, program administrators may benefit from re-evaluating their procedures, including clarifications for when to test, how to test, and providing a clear protocol for what constitutes a testing attempt. For example, attempting to test when the baby is agitated or when the test environment is excessively noisy will often result in a failed screening result even if the baby has normal hearing. Debris in the test probe, excessive cerumen in the infant’s ear canal, or the probe tip blocked against the canal wall also should be identified so that effective adjustments can be made prior to attempting the OAE test. Well-trained screeners can readily identify adverse test conditions, ensure proper probe fit, and proceed with testing only when conditions are conducive to obtaining an accurate test result.

There are a number of resources that can guide NBHS program administrators to evaluate their current program procedures and identify potential areas of improvement. For example, NCHAM offers free online training modules for newborn hearing screening programs (http://www.infanthearing.org/nhstc). Even those who believe their screening programs are highly effective may benefit from regularly evaluating program processes to ensure the screening follows best-practice recommendations.

Conclusions

It is appropriate for administrators of newborn hearing screening programs to be concerned about how often OAE screening should be repeated—but not because repeated screening prior to discharge will result in a high number of false negative results due to statistical artifacts. As shown in this article, very few babies with permanent hearing loss are likely to pass a newborn hearing screen test because the test was repeated multiple times. Even if a TEOAE screening test were repeated ten times for every baby, fewer than 1% of those with permanent hearing loss would pass because of repeated testing.

It should be noted that once a baby has failed the newborn hearing screening test, diagnostic assessment to determine the baby’s hearing status should be done as soon as possible. The results of this study should not be used to justify repeated OAE screening after the baby is discharged from the hospital as a prerequisite for doing the diagnostic evaluation. Such a practice has nothing to do with false negatives as a result of statistical artifacts of repeated testing and will only delay diagnosis and commencement of appropriate early intervention.

It is important for administrators of NBHS programs to be thoughtful about how often newborn hearing screening tests are repeated and to train their screeners accordingly. Not repeating the test often enough will lead to inappropriately high numbers of babies with normal hearing who fail a screen. This will lead to higher costs for follow-up screening and diagnostic testing. Repeating screening tests too often is also an inefficient use of staff time and may undermine the credibility of the program.
Newborn hearing screening programs should have well-trained screeners who recognize when to attempt testing and when to repeat OAE testing to obtain an accurate test result rather than focusing on the number of tests performed. To do otherwise can undermine the success of the screening program by wasting time, disturbing the baby, and upsetting parents and health care providers.

References


Infant Diagnostic Evaluation via Teleaudiology Following Newborn Screening in Eastern North Carolina

Andrew Stuart

Department of Communication Sciences and Disorders
East Carolina University

Abstract

Universal newborn hearing screening in North Carolina began in 2000 under the auspices of the North Carolina Department of Health and Human Services Early Hearing Detection and Intervention Program (NC-EHDI). Despite initial success, lost to follow-up/lost to documentation for diagnostic testing was problematic. To address this, the NC-EHDI received U.S. Department of Health and Human Services Health Resources and Services Administration lost to follow-up funding to fund, in part, a pilot “Teleaudiology Project” in 2010 to provide services for infants in eastern North Carolina. This part of the state is a traditionally underserved area. The project involved a partnership with East Carolina University. The project’s goals were to provide infant diagnostic evaluations in rural eastern counties and to establish a coordinated system for the delivery of audiological evaluations for infants whose families experience economic and geographic barriers to service. Project planning preparation and preliminaries, project service model, and outcome data are presented. From 2011 to 2015, outcome data provide positive proof-of-concept for a teleaudiology model in meeting national recommendations for providing diagnostic testing of infants following screening referral in a timely manner. In addition, the endeavor provides graduate audiology students with a unique didactic and clinical experience in teleaudiology.

Introduction

The North Carolina General Assembly passed the General Statutes Chapter 130A-125 (Screening of newborns for metabolic and other hereditary and congenital disorders) in the fall of 1999. Implemented on August 1, 2000, it mandated a newborn screening program and universal newborn hearing screening in the state of North Carolina. Specifically, it authorized each newborn to undergo physiological screening in each ear for the presence of permanent hearing loss. Presently, the North Carolina’s Early Hearing Detection and Intervention Program (NC-EHDI) provides screen-rescreen-diagnosis-intervention. NC-EHDI is organizationally located in the North Carolina Department of Health and Human Services, Division of Public Health, Women’s and Children’s Health Section, Children and Youth Branch as part of the state Title V Maternal and Child Health Services Program.

Initial newborn hearing screening rates across North Carolina have been excellent. For example, 98.2% of infants born in 2006 were screened for the presence of permanent hearing loss (Williams et al., 2015). In 2012, the percentage of newborns receiving hearing screening remained high (i.e., 99.1%). From those that received diagnostic testing, prevalence of permanent hearing loss per 1000 screened was estimated as 1.8 and 1.6 in 2006 and 2012, respectively.

Despite initial success with universal hearing screening of newborns, lost to follow-up (LFU)/lost to documentation (LTD) for diagnostic testing following the screening phase was problematic. For example, 53.7% (808 of 1,505) of infants, who did not pass the newborn hearing screening and were referred in 2006, were LFU/LTD and did not undergo audiological diagnostic testing (Williams et al., 2015). Although improved in 2012, a similar pattern of performance was evidenced in 2012: More than one-third of 854 newborn infants referred following newborn hearing screening (37.8%, n = 323) were LFU/LTD and did not complete a diagnostic evaluation.

To address the LFU/LTD for diagnostic testing, the NC-EHDI sought and received U.S. Department of Health and Human Services Health Resources and Services Administration lost to follow-up funding in September 2009. A portion of the funds was used to develop a pilot Teleaudiology Project in 2010 to provide services for infants in 38 counties in the eastern part of North Carolina. The targeted eastern North Carolina catchment area is unique for several reasons.

Corresponding author’s contact information: Andrew Stuart, Ph.D., Department of Communication Sciences and Disorders, Mail Stop 668, East Carolina University, Greenville, NC 27858-4353, USA. Email: stuartad@ecu.edu

Acronyms: ABR = auditory brainstem response; BRI = basic rate interface; CSDI = Department of Communication Science and Disorders; DPOAE = distortion product otoacoustic emissions; ECU = East Carolina University; EHDI = Early Hearing Detection and Intervention; HIPPA = Health Insurance Portability Act; HL = hearing loss; ISDN = Integrated Services Digital Network; LFU = lost to follow-up; LTD = lost to documentation; NC = North Carolina; TM = telemedicine

Acknowledgements: This work was presented in part at the 2012 American Speech-Language-Hearing Association Annual Convention, Chicago, IL; 2015 American Speech-Language-Hearing Association Annual Convention, Denver, CO; and the Early Hearing Detection and Intervention 2012 Annual Meeting, St. Louis, MO. The assistance of Kathleen Watts, Program Manager (retired), North Carolina, Early Hearing Detection and Intervention, and Gloria Jones, Clinical Telehealth Manager (retired), East Carolina University Telemedicine Center, was invaluable toward this project.
relative to the rest of the state (see Figure 1). Eastern North Carolina is primarily rural farmland. Traditionally, the population in these eastern counties has a median income lower than that of the rest of the state and a larger percentage of people living below the poverty level (http://quickfacts.census.gov/qfd/maps/north_carolina_map.html). In addition to poverty, this area has an unusually high teen pregnancy rate, greater percentage of Medicaid births, higher percentage of mothers who have not completed high school, and a larger percentage of minority births in comparison to the remainder of the state. Geographically, many inland bodies of water complicate travel over much of the region as well as travel from the Outer Banks to the mainland. Travel for diagnostic audiologic services can be as long as five hours and involve marine routes.

The goal of the NC-EHDI Teleaudiology Project was to provide infant diagnostic evaluations in rural eastern counties and to establish a coordinated system for the delivery of audiological evaluations for infants whose families experience economic and geographic barriers to service. The driving objectives were to reduce the number of infants in eastern North Carolina who are LFU/LTD for diagnostic testing or have delayed follow-up after referring on a hearing rescreen and to provide diagnostic evaluations. These objectives were in tune with the 1-3-6 Plan Joint Committee on Infant Hearing (2007) recommendation of providing comprehensive audiological evaluations no later than three months of age. Diagnostic testing began in June 2011. What follows is a description of the project development and implementation. Outcome data from June 2011 to July 1, 2015 is also presented.

Method

Participants
The catchment area included 22 of 98 birthing centers in North Carolina. Approximately 15% of live births ($n = 68,494$) in the state occurred at these birthing centers. Of those infants, 98.9% were screened prior to hospital discharge. Following initial screening, 2.3% ($n = 1559$) were referred for rescreen. Prior to discharge, the parent(s)/caregiver(s) was/were given information and an appointment for outpatient rescreen. Of those referred for rescreening, approximately 86% were evaluated ($n = 1339$). Rescreening tests were conducted following discharge at the birth hospital or at North Carolina Division of Public Health (NCDPH) local county health clinics. The percentage of infants in the catchment area that were screened and referred for a diagnostic test was similar to those in the rest of the state (see Figure 2).

Approximately 12% of those rescreened were referred for further diagnostic testing ($n = 157$). This represented approximately 0.2% of live births (see Figure 3) and was similar to the rest of the state. The total number of infants referred for diagnostic testing was also similar to those in the rest of the state (see Figure 3).

One hundred and fifty-seven infants were referred for diagnostic testing in the catchment area. Parent(s)/guardian(s) of 18 infants declined diagnostic testing. Approximately 29% of the remaining infants ($n = 40$) were referred for diagnostic testing, 40% were female. Slightly more than one-half (i.e., 56%) were referred for unilateral diagnostic testing.
Materials and Procedures

Project planning preparation and preliminaries. Preparation and implementation of preliminary project processes by NC-EHDI took 18 months. The project initiation involved establishing a partnership with East Carolina University (ECU). This was a logical first step as the university had an established Telemedicine Program (ECU-TM) and experienced infant audiological diagnosticians in the Department of Communication Science and Disorders (ECU-CSDI). In addition, ECU-CSDI was an early pioneer in examining the provision of audiology services in a telehealth environment (Givens et al., 2003). In fact, the term “teleaudiology” was coined at ECU (Givens & Elangovan, 2003). A contract for services was developed and put in place between NC-EHDI and ECU. Following consultation with ECU-CSDI, necessary diagnostic audiologic equipment (i.e., evoked potential/otoacoustic emission systems and middle ear analyzers) was purchased. NC-EHDI developed protocols, guidelines, and training for their staff. It was also necessary to seek approval from North Carolina Board of Examiners for Speech-Language Pathologists and Audiologists for telepractice (North Carolina Administrative Code Title 21 64.0219; effective July 1, 2010).

The ECU-TM has been in continuous operation since its inception in 1992, making it one of the longest running clinical telemedicine operations in the world. The Clinical Telehealth Manager of ECU-TM initially undertook a number of preliminary steps such as determining equipment/network needs, defining technical and user support, transferring protected health information, medical records, establishing lines for referring, defining scheduling responsibilities and coordination, deciding and establishing immediate assistance protocol for teleaudiology delivery, and establishing a call center for field assistance. The ECU-TM also configured both patient end units at remote sites (See Figure 4) and the ECU-CSDI provider site. The remote sites established the ECU-TM network, which comprises heterogeneous communications links, including full and fractional T-1 (1.54 Mbps) and Integrated Services Digital Network (ISDN), which is typically aggregated at 3 Basic Rate Interface (BRI; 384 kbps). The remote sites were examination rooms in regional hospitals and typically equipped with a general view camera with pan, zoom, and tilt capability mounted on a mobile cart. These units used Advanced Encryption Standard encryption for Health Insurance and Portability Act (HIPAA) compliancy, video switching for auxiliary inputs, content sharing with laptop/audiology test equipment, user profile synchronization provisioned for wireless, and network/power connectivity. The ECU-CSDI provider site was equipped with a Polycom HDX 4000 HD video conferencing system. This system allowed for pan, zoom, and tilt camera far-end control in the remote site room; picture-in-picture layout control; directory dialing; up to 6 Mbps calls; mobile desktop capability; and dual audio/microphone capability.


The implementation of the diagnostic audiologic component of this project was the responsibility of the lead audiologist at ECU-CSDI. It was a four-fold process that included test protocol development, training NC-EHDI site staff, dual site preparation (i.e., remote test site and ECU-CSDI), and continuing evaluation/changes of protocol. The development of the diagnostic protocol was consistent with existing guiding diagnostic principles (American Speech-Language-Hearing Association, 2004; Joint Committee on Infant Hearing, 2007; Ontario Infant Hearing Program, 2008; British Columbia Early Hearing Program, 2008). The objective was to determine the presence or absence of permanent childhood hearing impairment with a target impairment of hearing threshold ≥ 30 dB HL in 500 to 4000 Hz range. The diagnostic protocol included patient history, cursory otoscopy, middle-ear analysis, distortion product otoacoustic emissions (DPOAEs), and auditory brainstem response (ABR). Onsite training at ECU-CSDI for NC-EHDI staff testing at remote sites was undertaken. This training included classroom instruction for test equipment and diagnostic protocols, provision of a protocol handbook, and lab instruction and exercises with test equipment and diagnostic protocols. Continued consulting support was ongoing with audiologists/technicians and the lead audiologist at ECU. Site preparation began at ECU with consultation between the lead audiologist and the Clinical Telehealth Manager at the ECU-TM. Equipment setup and training on Polycom systems was foremost. The remote audiologists/technicians in conjunction with the ECU-CSDI lead audiologist undertook site preparation at the remote sites.

Teleaudiology project service model. The Teleaudiology Project’s diagnostic service delivery is a hybrid model. That is, it uses synchronous services to clients in real time and asynchronous store-and-forward
of audiometric data. Initial communication with parent(s)/caregiver(s) was with direct telephone contact by NC-EHDI staff. Diagnostic testing options via teleaudiology or at a diagnostic test center closest to their geographic location was offered. When parent(s)/caregiver(s) chose the teleaudiology option, an appointment was arranged by NC-EHDI staff. The family was informed of test date, time, and site location. Information was provided about preparations for the testing and the length of test as well. The day prior to the test, a reminder call was made to confirm the appointment, review the preparation instructions, and answer any last-minute questions.

NC-EHDI staff arrived at the remote site to set equipment up and connect with the ECU-CSDI lead audiologist prior to testing. After arrival of the infant and family, at the beginning of the testing session, an introduction to the lead audiologist at the provider site occurred via the Polycom video hardware. The infants were prepared for testing by NC-EHDI staff and the lead audiologist at the ECU-CSDI site who oversaw testing once the infant was settled. The diagnostic test battery was consistent with the Joint Committee on Infant Hearing (2007) position statement. The protocols were consistent with the Ontario Infant Hearing Program (2008) and the British Columbia Early Hearing Program (2008). The main goal of assessment was to determine the presence or absence of permanent childhood hearing impairment. The nominal target permanent childhood hearing impairment includes any hearing threshold \( \geq 30 \) dB HL at any frequency in the range of 500 to 4000 Hz, in either ear. The target permanent childhood hearing impairment includes conductive impairment associated with structural anomalies of the ear but does not include impairment attributable to non-structural middle ear conditions. The target also includes auditory neuropathy/auditory dys-synchrony.

All testing was attempted while the infant was in natural sleep or resting quietly. Wherever feasible, bilateral assessment included all of the procedures listed in Table 1. Except for the initial otoscopy, the order of procedures was discretionary. The order of testing proceeded on the basis of obtaining the most important/most useful information first, the next most important next, et cetera for diagnostic, management, and parent/caregiver information purposes. The sequence-of-testing within a procedure (e.g., within ABR assessment) follows the same underlying principle—thus, most infants would undergo the same sequence. DPOAE and ABR testing was conducted with a GSI Audera AEP system (Version 2.67). Middle-ear analysis

### Table 1. Diagnostic Test Protocol Components.

<p>| | |</p>
<table>
<thead>
<tr>
<th></th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>1.</td>
<td>History taking.</td>
</tr>
<tr>
<td>2.</td>
<td>Cursory otoscopy.</td>
</tr>
<tr>
<td>3.</td>
<td>DPOAE amplitude and noise floor measurements at f2 frequencies of 1500, 2000, 3000 and 4000 Hz. The f2/f1 ratio was 1.2, with L1 and L2 levels of 65 and 55 dB SPL (Gorga et al., 1997).</td>
</tr>
<tr>
<td>4.</td>
<td>Middle-ear analysis, which will include admittance tympanometry using a probe frequency of 1000 Hz and ipsilateral middle-ear muscle reflex testing using a 2000 Hz stimulus with a probe frequency of 1000 Hz (Margolis et al., 2003).</td>
</tr>
<tr>
<td>5.</td>
<td>ABR threshold estimation by air conduction at 2000 Hz and 500 Hz with tonal stimuli. If time permits it would be desirable to also obtain ABR threshold estimation at 4000 Hz and 1000 Hz (Stapells, Gravel, &amp; Martin, 1995).</td>
</tr>
<tr>
<td>6.</td>
<td>Tonal stimulus ABR threshold estimation by bone conduction, where indicated, at 500 Hz and 2000 Hz (British Columbia Early Hearing Program, 2008).</td>
</tr>
<tr>
<td>7.</td>
<td>In special circumstances, where indicated, high-intensity click-ABR measurement for auditory neuropathy/auditory dys-synchrony, including cochlear microphonic potentials and stimulus artifact analysis (British Columbia Early Hearing Program, 2008).</td>
</tr>
</tbody>
</table>

*Note. DPOAE = Distortion Product Otoacoustic Emission ; ABR = Auditory Brainstem Response

*Parent(s)/guardian(s) were advised their infant be sleep deprived and arrive at the test site hungry. This means that the night prior to testing, the infant should not be allowed to get his/her normal amount of sleep. Also, it is normally appropriate to deny sleep and food for at least an hour before testing unless medically contraindicated. If the child is being brought to the test by car, it is important that every reasonable effort be made (consistent with safety) to keep the child awake on the journey. Because of the soporific effect of car journeys on infants, it was advised another person in addition to the driver is usually necessary.*
was conducted with a GSI 39 Auto Tymp system. Detailed test protocols are presented in the Appendix. In the cases of unilateral referrals, the referred ear was tested first. If the infant was cooperative, the other ear was also tested. Four infants were recalled when diagnostic testing was not completed.

Diagnosis of infant hearing status was based on a general approach of audiologic inference with an integration and critical evaluation of all test findings (British Columbia Early Hearing Program, 2008). An infant was considered as audiometrically normal if air-conduction estimated hearing thresholds were 25 dB HL or better for all frequencies and/or DPOAE amplitudes exceeded the 5th percentile of the normal population and the 95th percentile of the impaired population at all frequencies (Gorga et al., 1997). An infant was considered to have a sensorineural impairment if air-conduction estimated hearing thresholds were > 25 dB HL; ABRs to bone-conducted stimuli exceeded the minimum test levels (i.e., elevated threshold); and/or DPOAE amplitudes were absent; and/or peak compensated static acoustic admittance fell below the 5th percentile of the normal population (Margolis, Bass-Ringdahl, Hanks, Holte, & Zapala, 2003). Auditory neuropathy/dys-synchrony was considered if the infant presented with OAEs and cochlear microphonics, abnormal ABRs, and absent middle ear acoustic reflexes.

Following the assessment, the parent(s)/caregiver(s) was/were counseled, via video, regarding test results by the lead audiologist. In the case where test results were pending, due to offline analysis in detail following asynchronous store-and-forward of audiometric data, parent(s)/caregiver(s) were contacted via telephone. The lead audiologist at ECU also reported diagnostic outcomes and recommendations via mail to the primary care physician/referring source within five business days of the diagnostic assessment. In addition, diagnostic outcomes and recommendations were entered into the North Carolina Division of Public Health Woman and Children Services Web (Hearing Link) website within five business days of the diagnostic assessment for state data tracking of hearing screening/diagnostic outcomes. Infants that presented with conductive hearing loss/abnormal middle ear function were referred to an otolaryngologist and recommended for retest following any medical management. Infants that presented with sensorineural hearing loss were also referred to an otolaryngologist, as well as back to the NC-EHDI staff for habilitation referral and family support services.

**Results**

The proportion of diagnostic tests performed via teleaudiology is illustrated in Figure 5. Diagnostic outcomes proportions are illustrated in Figure 6. The degree of sensorineural hearing loss ranged from mild to profound. Five percent (n = 2) of infants had an undetermined status (i.e., testing was incomplete to determine etiology). Boxplots of age of infants at screening, rescreening, and diagnostic test are shown in Figure 7.

The mean ages of infants at each test were 8.8 (SD = 27.6), 27.4 (SD = 25.5), and 73.3 (SD = 47.3) days for screening, rescreening, and diagnostic tests, respectively. The median ages of infants at each test were 1, 21, and 60 days for screening, rescreening, and diagnostic tests, respectively. Two infants who spent considerable time in the neonatal intensive care unit prior to hospital discharge mainly drove the variability in the distributions. Those two infants did not receive their initial screening until 89 and 154 days. All other infants received their initial screening in their first month. One infant relocated out of the state after the rescreen referral and before diagnostic testing could be completed. Approximately 77% of infants referred for diagnostic testing were evaluated in the first three months after birth.
Discerning Electrical Transmission andContinuing Evaluation/Changes of Dual Site Preparation (i.e., Remote Test Site and Observation/Direct Participation in Protocol Development, Involved in the Program Setup from the Beginning Including: Training of Audiology Graduate Students in Training. Students were Another positive of the project has been the involvement in a rural area like eastern North Carolina. The catchment area presents with a number of socioeconomic challenges including poverty, lower education level, high teen pregnancies and Medicaid births, and a large percentage of minority births in comparison to the remainder of the state. Additional geographical challenges compound the socioeconomic challenges including complicated and lengthy travel over much of the catchment area.

Another positive of the project has been the involvement of audiology graduate students in training. Students were involved in the program setup from the beginning including: observation/direct participation in protocol development, dual site preparation (i.e., remote test site and teleaudiology clinic), and continuing evaluation/changes of protocol. Students placed in the teleaudiology “clinic block” also gain a unique clinical experience. That is, there are few opportunities for students to participate in teleaudiology clinical placements. For example, the Telepractice Special Interest Group of the American Speech-Language-Hearing Association (2014) in a recent survey found that of 52% of audiologists who provide services through telepractice, only 11% are in college/university facilities. Wilson and Seal (2015) reported that less than one-half of current program directors, who responded to a survey of telepractice in university AuD programs, reported they offer teleaudiology course work or clinical training. Finally, only 4% of training programs used this technology to deliver audiology services (Grogan-Johnson, Meehan, McCormick, & Miller, 2015). The Teleaudiology Project’s experience and outcome data are also included in the AuD didactic curriculum for diagnostic testing models for infant hearing.

As with all programs, there remain some discouraging observations. First, there still remain a number of infants LFU/LTD for diagnostic testing (see Figure 6). Approximately 13% of infants referred were LFU/LTD. The issue of infants LFU/LTD has been identified in numerous programs (Alam, Gaffney, & Eichwald, 2014; Cockfield, Garner, & Borders, 2012; Krishnan, 2009; Liu, Farrell, MacNeil, Stone, & Barfield, 2008; Nikolopoulos, 2015; Spivak, Sokol, Auerbach, & Gershkovitch, 2009). It remains a continuing concern for clinicians and program administrators. There are also a number of parent(s)/caregiver(s) whose infants were referred for diagnostic testing who declined. Unfortunately, 11% of parent(s)/guardian(s) declined diagnostic hearing testing for their infant in this catchment area. The audiologic status of these infants is unknown. The reason(s) for the parental/guardian decline is unknown. This parental/guardian noncompliance is similar to that found following preschool hearing screening referrals in the same catchment area (Allen, Stuart, Everett, & Elangovan, 2004). These findings point to the necessity of hearing health care professionals to improve public education, for both parent(s)/guardian(s) and physicians, concerning the importance of identification and habilitation of hearing loss. Of those referred for diagnostic testing, approximately 29% were seen via teleaudiology. The status of the remaining 71% is unknown. It is likely that some were LFU/LTD and did not undergo audiological diagnostic testing as was previously found in North Carolina (Williams et al., 2015). It is speculated that the majority of these infants were seen at the major birthing facilities located in the higher population areas/cities (e.g., Greenville and Jacksonville, NC).

Numerous studies have demonstrated the technical and clinical feasibility of providing audiologic services via teleaudiology. They include audiometric testing (Givens & Elangovan, 2003; Givens et al., 2003; Margolis, Killion, Bratt, & Saly, 2016), hearing screening (Krumm, Huffman, Dick, & Klich, 2008; Lancaster, Krumm, Ribera, & Klich, 2008), hearing aid fitting (Blamey, Blamey, & Saunders, 2008), and medical referral and retest were recommended, is not reported.
2015; Penteado, Bento, Battistella, Silva, & Sooful, 2014), cochlear implant candidacy assessment (Aiello & Ferrari, 2015), and cochlear implant programming (Hughes et al., 2012). To date, however, there are no studies that have looked at an economic evaluation of teleaudiology services including the provision of infant diagnostic testing following newborn hearing screening. Remarkably, more than a decade ago, Suri, Dowling, Laxminarayan, and Singh (2005) presented a framework for an economic evaluation of telemedicine services both in terms of clinical effectiveness and cost-benefit. They identified a number of challenges for economic assessment including technological changes, sustainability of applications, availability of outcomes and other patient data, and generalizability of evaluation results. These same challenges face teleaudiology and specifically infant diagnostic testing following newborn hearing screening. As with their example in teleradiology, a significant barrier is the absence of a solid model for telemedicine cost analysis (i.e., how do you compare between two alternatives of teleaudiology and conventional service) and a lack of credible data sets with sufficient sample sizes. In addition, there is the need for randomized clinical trials of telemedicine. Suri et al. (2005) pointed out that studies might be driven by “technology push” rather than “clinical pull.” Studies should focus on three fundamental aspects: define what services are provided and the speed of such services; identify whom the clinical service is benefitting (i.e., the clinician or the patient); and determine what outcome measures (e.g., patient and/or parent/guardian satisfaction, compliance, and outcomes) should be used.

In summary, the Teleaudiology Project developed jointly by the NC-EHDI and ECU has demonstrated positive proof-of-concept for teleaudiology in meeting the recommendations for providing diagnostic testing of infants following screening referral in a timely manner. In addition, with the project located at a university site that provides clinical training of graduate audiologist students, it provides a distinctive opportunity for curriculum and clinical experiences in teleaudiology and stays current with developments in the field of audiology. Future studies are needed to evaluate the economic impact of teleaudiology services including the delivery of infant diagnostic testing following newborn hearing screening.

References


The distortion product otoacoustic emissions (DPOAE) protocol followed that of Gorga et al. (1997). Primary tones had an $f_2/f_1$ ratio of 1.22. $L_1$, $L_2$ levels were 65, 55 dB SPL. The $f_2$ frequencies were 1500, 2000, 3000, 4000, and 6000 Hz. A sequential signal presentation and time domain averaging was employed for data collection. The minimum and maximum averages that were acquired for each data point were 10 and 375, respectively. Frame rejection ensued if $L_1$ and $L_2$ were out of tolerance by ±5 dB and/or ambient noise levels exceeded 25 dB SPL. DPOAE collection terminated when either of the following occurred: test time exceeded 32 s or 1500 frames; 30% occurrences of frame rejection due to excessive ambient noise; and/or 20 occurrences of $L_1$ or $L_2$ being out of tolerance. The test was accepted when 32 frames were averaged and the average noise level was less than -12 dB SPL plus either of the following conditions were met: the DPOAE was 3 dB above the noise floor or the absolute noise level was less than -20 dB SPL.

For admittance tympanometry, the pressure sweep began at the starting pressure of +200 daPa and proceeded to -400 daPa at a rate of 600 daPa/s. The probe frequency was 1000 Hz. Peak compensated static acoustic admittance was determined from the negative tail at -400 daPa (Margolis et al., 2003). Ipsilateral middle-ear muscle reflex testing employed a 2000 Hz evoking stimulus. Reflex stimulus level should begin at 85 dB hearing level (HL) and increase in 5 dB steps up to no greater than 100 dB HL.

For behavioral hearing threshold estimation, ABR stimuli were air- and bone-conducted linear ramped 2-1-2 tone bursts. In the case of suspected auditory neuropathy/auditory dys-synchrony, 75 dB nHL 100 μs air-conducted clicks were used at a rate of 8.7/s. A total of 1026 samples were averaged and replicated. Tone bursts were centered at 500, 1000, 2000, and/or 4000 Hz. Stimuli were presented through a GSI TIP-50 insert earphone or a Radioear B-71 bone vibrator at a rate of 37.7/s. A total of 2014 samples were averaged and replicated. Reference threshold levels for air- and bone-conducted clicks were adopted from Yang, Stuart, Mencher, Mencher, & Vincer (1993). Reference threshold levels for tone burst stimuli were adopted from Stapells (2000). An ipsilateral recording montage was used with the noninverting electrode on the high-forehead (Fpz), inverting electrode on the ipsilateral postauricular area (M1/2), and one common to the contralateral inferior postauricular area (M2/1). Interelectrode impedances were maintained below 5000 Ω. The recorded electroencephalogram was amplified 105 and bandpass filtered (30 to 3000 Hz). Electroencephalogram samples exceeding ± 25 μV were rejected. Analysis times were 13 ms post-stimulus for click and 25 ms post-stimulus for tone bursts. The bone vibrator was placed in a supero-posterior temporal position during bone conducted stimuli delivery (Stuart, Yang, & Stenstrom, 1990). An elastic band with Velcro was used to hold the bone vibrator with a coupling force of 425 ± 25 g (Yang & Stuart, 1990). Coupling force was verified with a spring scale (Ohaus 8014) that manually pulled the bone vibrator away from the skull by a nylon monofilament attached to the bone vibrator. The coupling force was measured at the point the vibrator cleared and became flush with the scalp.
A Review of Internet Resources Related to Spoken Language Intervention for Spanish-Speaking Parents of Children who are Deaf or Hard of Hearing

Karen Muñoz,¹,²
Lauri Nelson,¹
Brittan A. Barker,¹

¹Department of Communicative Disorders and Deaf Education, Utah State University
²National Center for Hearing Assessment and Management, Utah State University

Abstract

Objective: The purpose of this study was to identify website information related to hearing loss, hearing technology, and spoken language development available to Spanish-speaking parents of children who are deaf or hard of hearing (DHH).

Design: An exploratory, descriptive design was used to determine the presence or absence of parent education information on a variety of websites.

Study Sample: The study explored Internet resources provided by national, state, and parent support organizations in the United States.

Results: A total of 53 organization websites were identified that had information for parents of children who are DHH and learning spoken language, eight of which were international. Fifteen content areas were reviewed for each website. Of the 53 websites, 25 had information in Spanish.

Conclusions: Results of the current study revealed website resources are often fragmented and less in-depth for Spanish speaking parents with children who are DHH and learning spoken language.

Key Words: hearing, website, Internet, children, Spanish

Acronyms: DHH = Deaf or Hard of Hearing; EHDI = Early Hearing Detection and Intervention; FM = Frequency Modulation; JCIH = Joint Committee on Infant Hearing

Corresponding author’s contact information: Karen Muñoz, Utah State University, 2620 Old Main Hill, Logan UT 84322, Email: karen.munoz@usu.edu

Introduction

Hearing loss in early childhood is often unexpected and 95% of children who are deaf or hard of hearing (DHH) have hearing parents (Mitchell & Karchmer, 2004) who want their child to learn spoken language (Alberg, Wilson, & Roush, 2006). For many parents, identification of hearing loss not only alters their vision of their child’s future, but their confidence in how to support their child’s development and overcome obstacles that may impact their access to or understanding of pertinent information related to their child’s diagnosis (Cole & Flexer, 2015). Upon learning their child is DHH, parents often seek information about hearing loss (e.g., cause, type, degree) and hearing technology (e.g., hearing aids, cochlear implants, assistive technology); many parents are unfamiliar with available services and have financial questions or concerns (DesGeorges, 2003). In addition, social and emotional support, including accessing parent-to-parent connections can be critical for many families (Henderson et al., 2014).

When hearing loss is identified, and early intervention provided, child developmental outcomes can be optimized. Tomblin, Oleson, Ambrose, Walker, and Moeller (2014) found that when children had early and consistent audibility with hearing aids, their language outcomes were better than children without access to consistent intervention. Parents, however, face numerous challenges in learning how to secure and navigate daily management of hearing technology and language intervention (Moeller, Hoover, Peterson, & Stelmachowicz, 2009; Muñoz et al., 2015; Muñoz, Preston, & Hicken, 2014; Muñoz, Blaiser, & Barwick, 2013; Sjoblad, Harrison, Roush, & McWilliam, 2001). In fact, studies have shown significant variability in hearing aid use for young children (Jones, 2013; Jones & Launer, 2010; Muñoz et al., 2014; Walker et al., 2013), a factor that can influence spoken language development. Parents face a steep learning curve related to intervention, and access to information (e.g., various Internet resources) can offer opportunities for parents and other caregivers to gain essential knowledge.

The importance of culturally and linguistically family centered services for effective intervention is identified in professional practice guidelines (Joint Committee on Infant Hearing [JCIH] Supplement, 2013), and includes provision of materials for families in their home language. In the United States, by 2050, it is projected that 82% of population growth will be from immigrants and U.S.-born minorities, with Hispanics representing one of the fastest growing segments of the population (Passel & Cohn, 2008). Given these data, the number of potential Spanish speakers would rise to about 62 million people in the U.S. (Instituto Cervantes, 2015). In a country where English is the dominant language it is important to ensure that health care information is available and accessible to all families—

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regardless of their home language. A language barrier can increase the risk of having limited access to information and resources (Steinberg, Bain, Li, Delgado, & Ruperto, 2003), reducing a person’s ability to clearly understand information that is important for making educated decisions about health care. Furthermore, parents can experience challenges accessing information and support from experienced providers. In the U.S., state Early Hearing Detection and Intervention (EHDI) coordinators have reported significant shortages in pediatric audiologists (Muñoz, Bradham, & Nelson, 2011). There are also shortages of speech-language pathologists and deaf educators with expertise in listening and spoken language (Nelson, Lenihan, & White, 2014). In addition, hearing loss is relatively low incidence and many families live in disperse geographical locations. Together these factors further increase challenges for Spanish-speaking families in the U.S.

Research shows that when people are faced with a new diagnosis, such as hearing loss, they turn to the Internet for education (Rice, 2006). In fact, the Internet is now nearly ubiquitous in the U.S., with 90% of people ages 25 years and older (U.S. Department of Commerce, 2014) utilizing the Internet as a primary source of information. Having access to accurate and complete information via the Internet can help parents of children who are DHH effectively understand and manage their child’s hearing loss, support their child’s developmental progress, and identify potential emotional and financial support services.

Given the disperse population, challenges accessing professionals with specialized expertise in pediatric hearing loss, and the increase in Spanish language families in the U.S., there is a need for Internet resources in Spanish to support parent learning related to spoken language development for children who are DHH. For these reasons, this study was completed to determine what information is available electronically via the Internet by Spanish-speaking parents of children who are DHH and communicate using spoken language.

Method

National, state, and parent support organization websites in the U.S. that address pediatric hearing loss were included in the review if the scope of their website addressed topics related to intervention for spoken language acquisition in children who are DHH. Websites from private clinics and hospitals, and websites that addressed only newborn hearing screening were excluded from the review. The first author and an assistant jointly conducted the website review in June 2015 in two phases.

Procedure

Phase 1 included a broad Internet search of national, state, and parent support organizations in the U.S. using the Google Chrome search engine. State EHDI websites that included information related to intervention for spoken language development after diagnosis of hearing loss were also included. Links embedded within the initial sites and hyperlinked to outside websites were also evaluated.

Content Areas

The content area topics deemed important for families seeking information about hearing loss, hearing technology, and spoken language intervention were determined using an iterative process. As websites were reviewed, main topics were identified and added to a list. As new topics were identified, previously reviewed websites were checked again to see if the topic area was overlooked during the initial review of content. The sites were searched for presence or absence of content areas; 15 areas were identified (see Table 1).

Website Features and Social Media

Various social media and website features were noted as present or absent. The features were determined using an iterative process. As websites were reviewed, features (e.g., newsletters, blogs, videos) and social media were added to a list as they were identified. As new features and social media were identified, previously reviewed websites were checked again to see if the feature was overlooked during the initial review.

Table 1. Content Areas Included in the Website Review

| Foundational knowledge (e.g., type of hearing loss) | Advocacy and education |
| Hearing aids | Hearing assessment |
| Cochlear implants | Parent-to-parent support |
| Early intervention | Glossary |
| Medical referrals after diagnosis (e.g., otolaryngology) | Social-emotional |
| Assistive technology (generally) | Financial assistance |
| FM systems | Additional disabilities |
| Parent-professional collaboration (working with professionals) | |

Note. FM = Frequency Modulation

Table 2. Social Media and Website Features included in the Website Review.

| Google+ | Videos | Newsletter | Contact us |
| Facebook | Blog | Rich Site Summary (RSS) | Chat |
| Twitter | YouTube | Pinterest | Parent distance education |
Twelve features and social media options were identified (see Table 2). Phase 2 included an in-depth review of the written content on the websites that included information in Spanish. Websites from Phase 1 that only had information in English were excluded. The content in Spanish for each website was subjectively rated by the first author and the research assistant jointly using a 3-point scale (1 = no information; 2 = some information; 3 = extensive information). This scale was used to broadly categorize websites rather than to determine specific differences between them.

For example, www.babyhearing.org, provides in-depth information (e.g., hearing aids and cochlear implants) in English and Spanish, and was categorized as having extensive information, whereas, http://www.parentcenterhub.org/repository/auditiva provides basic descriptions on some topics (e.g., Early Intervention and educational considerations) and was categorized as having some information.

Analysis
The assistant entered ratings into an Excel spreadsheet. For Phase 1, a code was entered to indicate the presence or absence for each content area. When information was present the assistant indicated whether it was in English, Spanish, or both English and Spanish. For Phase 2, the same content areas (Table 1) were reviewed using the 3-point Likert scale for the websites that provided information in Spanish. The ratings data from Phase 2 were also entered into the spreadsheet. Descriptive statistics were used to identify frequencies and trends noted across the websites.

Results
The website review revealed a total of 53 websites with information about hearing loss, hearing technology, and spoken language intervention. For the 15 content areas identified (Table 1), five were found on more than 50% of the websites: cochlear implants, foundational knowledge (e.g., type of hearing loss, causes of hearing loss), early intervention, hearing aids, and hearing assessment. Less than 20% of the websites addressed parent-professional collaboration and additional disabilities (see Figure 1).

The 12 website features (e.g., FaceBook, Twitter, blog), embedded in the 53 websites, were evaluated to identify if they were present or absent (see Figure 2). More than 50% of the websites contained contact information, a FaceBook link, and Twitter account link. Approximately one-third of the websites offered a newsletter. Less than 10% of the websites offered educational modules or a chat feature to ask a question.

Of the 53 websites, 25 had information in Spanish in at least one of the 15 content areas (see Figure 3). The websites with Spanish language information included four national, seven state, six parent support, and eight international websites (for a list, see Appendix). More than 50% of the websites had some information in Spanish on the following eight content areas: foundational knowledge, cochlear implants, early intervention, hearing aids, hearing assessment, Frequency Modulated (FM) systems, assistive technology, and advocacy/education. Approximately one-third of the websites had extensive information in three areas: foundational knowledge, hearing aids, and a glossary. Less than 10% of the websites had extensive information in Spanish related to parent-to-parent support,
parent-professional collaboration, additional disabilities, and FM systems.

**Discussion**

This website review investigated the extent of healthcare information about hearing loss, hearing technology, and spoken language intervention available for families of children who are DHH and speak Spanish. Findings from this study revealed 53 websites with healthcare information in English and/or Spanish. Of those, 25 websites had information in Spanish, and very few provided in-depth information for parents. Information was often fragmented on websites with only some content areas (Table 1) included. For websites that provided information in both English and Spanish, the information in Spanish was often more limited than that provided in English; for example, FaceBook pages were only in English and videos were often only in English. The gaps that exist for Spanish-language Internet resources further limit how families can gain needed knowledge to help their children.

Hearing loss identification and subsequent intervention can be overwhelming for parents. Parents and other caregivers are faced with learning new information and skills, as well as how to apply new learning in their daily lives. To compund the problem, when English is not the primary language in the home, parents may require an interpreter when communicating with the audiologist and other intervention providers. Having access to health information in a variety of formats (e.g., verbal, written, demonstration, video) can aid in retention of information and support the development of effective self-management in patients and their families (Rogo, 2014). Both mothers and fathers of children who are DHH reported that they want information in a variety of formats (verbal, written, video), and that access to accurate information is important for learning to integrate new skills into daily routines with their children (Muñoz et al., 2015). Hispanic parents of young children who are DHH specifically reported that they want more concrete resources (Caballero & Muñoz, 2015).

Children often have caregivers other than the mother, including the father, grandparents, other family members, and day care providers. Audiologists have reported that they most frequently instruct mothers (n = 332/343; 97%) on how to manage hearing aids (Melbos et al., 2015), yet mothers have reported that other individuals care for their children who are DHH during the day (Rusk & Muñoz, 2015). The mother then is often in the position of instructing other caregivers how to manage the child’s hearing devices and auditory environment. Internet resources can aid instruction of other caregivers, reinforce parent learning, and support parents in gaining confidence with new knowledge and skills.

Access to accurate information via the Internet in the parents’ primary language offers a mechanism to support parent learning in a flexible manner that can serve to reinforce and supplement information provided by the audiologist and other intervention providers. For information to be accessible for learning, factors such as health literacy, computer access, readability of information on other devices (e.g. smart phones), and cultural sensitivity that can influence how effectively information reaches the intended audience should be considered (Cotton & Gupta, 2004). Additional research is needed to better understand factors that influence Hispanic parent access of information on the Internet, how it may need to be tailored to meet their access needs, and how delivery of information can provide action-oriented learning support.

**Conclusions**

Results of the current study revealed website resources are often fragmented and less in-depth for Spanish speaking parents with children who are DHH and learning spoken language. Current available resources in Spanish on national, state, U.S. parent organizations, and international websites primarily support awareness for the content in the hearing healthcare areas identified in this review. The present data reflect a need for more complete, in-depth information that is easily accessible on the Internet, to support parent learning and enhance parent confidence for managing their children’s hearing loss effectively on a daily basis. Parents need access to complete and accurate information, regardless of their primary language.

**References**


# Appendix

Websites with Spanish language information related to hearing loss, hearing technology, and spoken language development for parents of children who are deaf or hard of hearing (DHH) as of June 2015.

<table>
<thead>
<tr>
<th>Organization</th>
<th>Website URL</th>
</tr>
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<tbody>
<tr>
<td>National (U.S.)</td>
<td></td>
</tr>
<tr>
<td>Center for Disease Control and Prevention</td>
<td><a href="http://www.cdc.gov/ncbdd/hearingloss/">www.cdc.gov/ncbdd/hearingloss/</a></td>
</tr>
<tr>
<td>Center for Parent Information and Resources</td>
<td><a href="http://www.parentcenterhub.org/repository/auditiva">www.parentcenterhub.org/repository/auditiva</a></td>
</tr>
<tr>
<td>March of Dimes</td>
<td><a href="http://nacersano.marchofdimes.org/complicaciones/perdida-de-la-audicion.aspx">http://nacersano.marchofdimes.org/complicaciones/perdida-de-la-audicion.aspx</a></td>
</tr>
<tr>
<td>State Early Hearing Detection and Intervention</td>
<td></td>
</tr>
<tr>
<td>Iowa</td>
<td><a href="http://idph.iowa.gov/ehdi/families">http://idph.iowa.gov/ehdi/families</a></td>
</tr>
<tr>
<td>Kansas</td>
<td><a href="http://www.soundbeginnings.org/">http://www.soundbeginnings.org/</a></td>
</tr>
<tr>
<td>Wyoming</td>
<td><a href="http://www.wyomingehdi.org/">http://www.wyomingehdi.org/</a></td>
</tr>
<tr>
<td>Michigan</td>
<td><a href="http://www.michigan.gov/mdch/0,1607,7-132-2942_4911_21429---,00.html">http://www.michigan.gov/mdch/0,1607,7-132-2942_4911_21429---,00.html</a></td>
</tr>
<tr>
<td>New Mexico</td>
<td><a href="http://archive.nmhealth.org/nbhs/">http://archive.nmhealth.org/nbhs/</a></td>
</tr>
<tr>
<td>Parent</td>
<td></td>
</tr>
<tr>
<td>Baby Hearing</td>
<td><a href="http://www.babyhearing.org">www.babyhearing.org</a></td>
</tr>
<tr>
<td>AG Bell Listening and Spoken Language</td>
<td><a href="http://www.listeningandspokenlanguage.org">www.listeningandspokenlanguage.org</a></td>
</tr>
<tr>
<td>Center</td>
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<tr>
<td>Beginnings</td>
<td><a href="http://www.ncbegin.org">www.ncbegin.org</a></td>
</tr>
<tr>
<td>Hands &amp; Voices</td>
<td><a href="http://www.handsandvoices.org">www.handsandvoices.org</a></td>
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<tr>
<td>John Tracey</td>
<td><a href="http://www.jtc.org">http://www.jtc.org</a></td>
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<tr>
<td>Hear-It</td>
<td><a href="http://www.hear-it.org">www.hear-it.org</a></td>
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<tr>
<td>International</td>
<td></td>
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<td>FIAPAS</td>
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</tr>
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<td>Fundacion ECO</td>
<td><a href="http://www.ecodepadres.org">www.ecodepadres.org</a></td>
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<td>t-oigo</td>
<td><a href="http://www.t-oigo.com">www.t-oigo.com</a></td>
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<tr>
<td>Mas Que Padres</td>
<td><a href="http://www.masquepadres.arrakis.es">www.masquepadres.arrakis.es</a></td>
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<tr>
<td>Fundacion Amaoir</td>
<td><a href="http://www.amaoir.org">www.amaoir.org</a></td>
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<tr>
<td>Mi Hijo Sordo</td>
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What Are Others Publishing About Early Hearing Detection and Intervention?

The aim of the Journal of Early Hearing Detection and Intervention (JEHDI) is to promote access to evidence-based practice, standards of care, and research focused on all aspects of Early Hearing Detection and Intervention. Taking a broad systems perspective, 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, family support, early intervention, the medical home, information management, financing, quality improvement and other issues that contribute to improving EHDI systems.

Whereas JEHDI is the only journal that focuses specifically on improving EHDI systems, many other journals publish relevant articles as a part their journal’s broader focus. To help JEHDI readers stay up-to-date about recently published material, we provide titles and abstracts of what has been published in the last 9 months that JEHDI editors think are most relevant to improving EHDI programs. Titles of all articles are hyperlinked to the source.


Background and Methods: The study participants included 41 full term hyperbilirubinemic infants (mean age 39.24 days) with normal birth weight (3,200-3,700 grams) that admitted in hospital for hyperbilirubinemia and 39 normal infants (mean age 35.54 days) without any hyperbilirubinemia or other hearing loss risk factors for ruling out maturational changes. All infants in hyperbilirubinemic group had serum bilirubin level more than 20 milligram per deciliter and undergone one blood exchange transfusion. Hearing evaluation for each infant was conducted twice: the first one after hyperbilirubinemia treatment and before leaving hospital and the second one three months after the first hearing evaluation. Hearing evaluations included transient evoked otoacoustic emission (TEOAE) screening and auditory brainstem response (ABR) threshold tracing.

Results: The TEOAE and ABR results of control group and TEOAE results of the hyperbilirubinemic group did not change significantly from the first to the second evaluation. However, the ABR results of the hyperbilirubinemic group improved significantly from the first to the second assessment (p=0.025).

Conclusion: The results suggest that the bilirubin induced auditory neuronal damage can be reversible over time so we suggest that infants with hyperbilirubinemia who fail the first hearing tests should be reevaluated after 3 months of treatment.


Objectives: Deterioration in hearing thresholds in children is of concern due to the effect on language development. Before universal newborn hearing screening (UNHS), accurate information on the progression of hearing loss was difficult to obtain due to limited information on hearing loss onset. The objective of this population-based study was to document the proportion of children who experienced progressive loss in a cohort followed through a UNHS program in one region of Canada. We explored risk factors for progression including risk indicators, audiologic, and clinical characteristics of children. We also investigated deterioration in hearing as a function of age. For this study, two working definitions of progressive hearing loss were adopted: (1) a change of ≥20 dB in the frequencies (500, 1000, and 2000 Hz) pure-tone average, and (2) a decrease of ≥10 dB at two or more adjacent frequencies between 500 and 4000 Hz or a decrease in 15 dB at one octave frequency in the same frequency range.

Design: Population-based data were collected prospectively on a cohort of children identified from 2003 to 2013 after the implementation of UNHS. Clinical characteristics including risk indicators (as per Joint Committee on Infant Hearing), age at diagnosis, type and severity of hearing loss, and initial audiologic information were recorded when children were first identified with hearing loss. Serial audiometric results were extracted from the medical charts for this study. Differences between children with progressive and stable hearing loss were explored using χ tests. Association between risk indicators and progressive hearing loss was assessed through logistic regression. The cumulative amount of deterioration in hearing from 1 to 4 years of age was also examined.

Results: Our analysis of 330 children (251 exposed to screening) with detailed audiologic records showed that 158 (47.9%) children had some deterioration (at least ≥10 dB and) in hearing thresholds in at least one ear. The 158 children included 76 (48.1%) with ≥20 dB loss in pure-tone average in at least one ear and 82 (51.9%) with less deterioration in hearing levels (≥10 but <20 dB). In the children with progressive hearing loss, of 131 children initially diagnosed with bilateral loss, 75 (57.3%) experienced deterioration in 1 ear and 56 (112 ears; 42.7%) in both ears (total of 187 ears). Of 27 children with an initial diagnosis of unilateral loss, 25 experienced deterioration in the impaired ear and 5 in the normal-hearing ear, progressing to bilateral hearing loss. Within 4 years after diagnosis, the mean decrease in hearing for children with progressive loss was 25.9 dB (SD: 16.4) in the right ear and 28.3 dB (SD: 12.9) in the left ear. We explored the risk factors for hearing loss identified by Joint Committee on Infant Hearing where there were sufficient numbers in our sample. On multivariate analysis, there was no statistically significant relationship between most risk indicators examined (neonatal intensive care unit admission, family history, syndromes, and postnatal infections) and the likelihood of progressive loss. However, the presence of craniofacial anomalies was inversely associated with risk of progressive hearing loss (odds ratio = 0.27; 95% confidence interval: 0.10, 0.71; p = 0.01), that is, these children were more likely to have stable hearing.

Conclusion: The results suggest that

Purpose: Patients treated with cranial radiation therapy (RT) are at risk for sensorineural hearing loss (SNHL). Although SNHL is often characterized as a delayed consequence of anticancer therapy, longitudinal reports of SNHL in childhood cancer survivors treated with contemporary RT are limited. We report the incidence, onset, severity, and long-term trajectory of SNHL among children receiving RT. Potential risk factors for SNHL were also identified.

Patients and Methods: Serial audiologic testing was conducted on 235 pediatric patients who were treated with conformal or intensity-modulated RT as part of an institutional phase II trial for localized primary brain tumors, including craniopharyngioma, ependymoma, and juvenile pilocytic astrocytoma. All but one patient had measurable cochlear radiation dose (CRD) greater than 0 Gy. The median follow-up from RT initiation to latest audiogram was 9 years with a median of 11 post-RT audiograms per patient. Audiograms were classified by the Chang Ototoxicity Grading Scale. Progression was defined by an increase in Chang grade from SNHL onset to the most recent evaluation.

Results: At last evaluation, SNHL was prevalent in 14% of patients: 2.1% had mild and 11.9% had significant SNHL requiring hearing aids. Median time from RT to SNHL onset was 3.6 years (range, 0.4 to 13.2 years). Among 29 patients with follow-up evaluations after SNHL onset, 65.5% experienced continued decline in hearing sensitivity in either ear and 34.5% had no change. Younger age at RT initiation (hazard ratio [HR], 2.32; 95% CI, 1.21 to 4.46), higher CRD (HR, 1.07; 95% CI, 1.03 to 1.11), and cerebrospinal fluid shunting (HR, 2.02; 95% CI, 1.07 to 3.78) were associated with SNHL.

Conclusions: SNHL is a late effect of RT that likely worsens over time. Long-term audiologic follow-up for a minimum of 10 years post-RT is recommended.


Abstract: Diagnosis of child permanent hearing impairment (PHI) can be made with extreme timeliness compared to the past thanks to improvements in PHI identification through newborn hearing screening programmes. It now becomes essential to provide an effective amplification as quickly as possible in order to restore auditory function and favour speech and language development. The early fitting of hearing aids and possible later cochlear implantation indeed prompts the development of central auditory pathways, connections with secondary sensory brain areas, as well as with motor and articulatory cortex. The aim of this paper is to report the results of a strategic analysis that involves identification of strengths, weaknesses, opportunities and threats regarding the process of achieving early amplification in all cases of significant childhood PHI. The analysis is focused on the Italian situation and is part of the Italian Ministry of Health project CCM 2013 “Preventing Communication Disorders: a Regional Program for Early Identification, Intervention and Care of Hearing Impaired Children”.


Abstract: New-born screening programs for congenital disorders and chronic disease are expanding worldwide and children “at risk” are identified by nationwide tracking systems at the earliest possible stage. These practices are never neutral and raise important social and ethical questions. An emergent concern is that a reflexive professionalism should interrogate the ever earlier interference in children’s lives. The Flemish community of Belgium was among the first to generalize the screening for hearing loss in young children and is an interesting case to study the public justification of early interventions for families with deaf children. This article uses a critical lens to study the archive of the government child healthcare organization in Flanders in order to uncover underlying constructions of childhood, deafness, and preventive health. We focus on two interrelated themes. The first is the notion of exclusion of the human factor through the mediation of technology. The second is the idea of deafness as endangering a healthy development, an impairment that can nevertheless be treated if detected early enough. It is argued that, since deafness cannot be viewed as a life-threatening condition, the public interest which is implicitly defended is not the rescue of deaf children rather the exclusion of otherness.

DOI: 10.1007/s11673-016-9752-y


Abstract: Auditory neuropathy spectrum disorder (ANSD) is a particular kind of hearing disorder characterised by normal outer hair cell function and abnormal or absent auditory brain stem responses. Little data are available regarding the prevalence of this condition in healthy newborns. We performed a retrospective medical records review of 791 referrals from universal neonatal hearing screening (UNHS) at a well-baby clinic to investigate the prevalence of ANSD. Hearing screening was performed by automated auditory brain stem response (ABR) testing. Diagnosis of ANSD was established when ABR tracings were absent in the presence of otoacoustic emissions and/or a cochlear microphonics. Amongst 201 infants with confirmed congenital hearing loss, 13 infants were diagnosed with ANSD. The condition was unilateral in six and bilateral in seven infants. A risk factor for hearing loss could be identified in three infants. Abnormalities on magnetic resonance imaging were found in six infants; five of them had cochlear nerve deficiency.

Conclusion: The prevalence of ANSD was 6.5 % amongst well babies with confirmed congenital hearing loss identified through UNHS. The estimated incidence of ANSD in our population of newborns at the well-baby clinic was 0.09/1000 live births. Magnetic resonance revealed an underlying anatomical abnormality in about half of the patients.

What is Known: Auditory neuropathy dys synchrony spectrum disorder (ANSD) is a particular form of hearing loss, mostly encountered in neonatal intensive care unit (NICU) graduates. • Little data are available on the prevalence and risk factors for ANSD in healthy newborns. What is new: • The estimated prevalence of ANSD in healthy newborns is 0.09/1000 live births. • In about half of the healthy newborns with ANSD, a structural abnormality was detected on magnetic resonance imaging of the posterior fossa/brain.
**Calcutt TL, Dornan D, Beswicke R, Tudehope DI.**

**Newborn Hearing Screening in Queensland 2009-2011: Comparison of Hearing Screening and Diagnostic Audiological Assessment between Term and Preterm Infants.**


**Aim:** This study compares rates and timing of newborn hearing screening outcomes, audiological assessment and hearing loss diagnosis between infants of different gestational age groups. Early identification and management of sensorineural hearing loss (SNHL), ideally by 3-6 months of age, facilitates speech and language optimisation. Literature stratifying hearing screening and diagnostic audiology assessment by gestational age groups is lacking.

**Methods:** Subjects were infants with recorded gestational ages receiving newborn hearing screening in Queensland between 2009 and 2011. Data were provided through the Queensland Healthy Hearing database. Infants were analysed in <34 weeks, 34-36+6 weeks, 37-38+6 weeks and ≥39 weeks gestational age groups.

**Results:** Infants (175 911) were eligible for analysis, 7.9% being preterm. Per 1000 infants analysed, bilateral SNHL of >40 dB occurred in 2.4 for <34, 1.4 for 34-36+6, 0.7 for 37-38+6 and 0.7 for ≥39 weeks gestation. Diagnoses attributable to newborn hearing screening direct referral were 93.1% for bilateral >40 dB SNHL and 88.2% for other hearing loss. Relative to term, preterm infants had a higher incidence of direct and targeted surveillance referrals, audiology assessment and hearing loss diagnosis. Preterm infants were screened later after birth.

**Conclusions:** Specific hearing screening and diagnostic characteristics differed between preterm infants <34 and 34-36+6 weeks gestation, and term infants. Consideration of unique gestational age strata characteristics supports care individualisation. Preterm infants represent a diagnostic challenge, with higher rates of bilateral >40 dB SNHL than term but correspondingly higher false positive results on screening, justifying vigilant monitoring. Focused research into specific risk factors in preterm infants is warranted.

**Carey JC, Palumbos JC.**


**Abstract:** Hearing loss represents the most common sensorineural disability of children. Remarkable advances in the identification of genes underlying nonsyndromic and syndromic hearing loss in just the last 2 decades have led to the ability to determine the specific genetic cause of hearing loss in many children. Surprisingly one gene, GJB2, encoding the protein connexin-26, accounts for about 20% of sensorineural hearing loss (including in India) and is considered the first tier in evaluating an infant with unexplained congenital hearing loss. Using the knowledge of the etiology of hearing loss, the authors propose a diagnostic reasoning process for the assessment of a child in the pediatric setting. Second tier testing consists of the multiple gene panels using whole exome sequencing strategies, and is becoming available in some regions of the world including the US. Referral to medical genetics is always a consideration in a child with no explanation for the hearing loss and in families with questions about recurrence risk.

**Chiou ST, Lung HL, Chen LS, Yen AM, Fann JC, Chiu SY, Chen HH.**


**Objective:** Little is known about the long-term efficacious and economic impacts of universal newborn hearing screening (UNHS).

**Design:** An analytical Markov decision model was framed with two screening strategies: UNHS with transient evoked otoacoustic emission (TEOAE) test and automatic acoustic brainstem response (aABR) test against no screening. By estimating intervention and long-term costs on treatment and productivity losses and the utility of life years determined by the status of hearing loss, we computed base-case estimates of the incremental cost-utility ratios (ICURs). The scattered plot of ICUR and acceptability curve was used to assess the economic results of aABR versus TEOAE or both versus no screening.

**Study Sample:** A hypothetical cohort of 200,000 Taiwanese newborns.

**Results:** TEOAE and aABR dominated over no screening strategy (ICUR=$-4800.89 and $-4111.23, indicating less cost and more utility). Given $20,000 of willingness to pay (WTP), the probability of being cost-effective of aABR against TEOAE was up to 90%.

**Conclusions:** UNHS for hearing loss with aABR is the most economic option and supported by economically evidence-based evaluation from societal perspective.

**DOI:** 10.1080/14992027.2016.1219777

**Costich JF, Durst AL.**

**The Impact of the Affordable Care Act on Funding for Newborn Screening Services.** *Public Health Rep.* 2016 Jan-Feb;131(1):160-6.

**Objective:** The Affordable Care Act requires most health plans to cover the federal Recommended Uniform Screening Panel of newborn screening (NBS) tests with no cost sharing. However, state NBS programs vary widely in both the number of mandated tests and their funding mechanisms, including a combination of state laboratory fees, third-party billing, and other federal and state funding. We assessed the potential impact of the Affordable Care Act coverage mandate on states' NBS funding.

**Method:** An analytical Markov decision model was framed with two screening strategies: UNHS with transient evoked otoacoustic emission (TEOAE) test and We performed an extensive review of the refereed literature, federal and state agency reports, relevant organizations’ websites, and applicable state laws and regulations; interviewed 28 state and federal officials from August to December 2014; and then assessed the interview findings manually.

**Results:** Although a majority of states had well-established systems for including laboratory-based NBS tests in bundled charges for newborn care, billing practices for critical congenital heart disease and newborn hearing tests were less uniform. Most commonly, birthing facilities either prepaid the costs of laboratory-based tests when acquiring the filter paper kits, or the facilities paid for the tests when the kits were submitted. Some states had separate arrangements for billing Medicaid, and smaller facilities sometimes contracted with hearing test vendors that billed families separately.

**Conclusions:** Although the Affordable Care Act coverage mandate may offset some state NBS funding for the screenings themselves, federal support is still required to assure access to the full range of NBS program services. Limiting reimbursement to the costs of screening tests alone would undermine the common practice of using screening charges to fund follow-up services counseling, and medical food or formula, particularly for low-income families.

The Assessment of the Newborn Hearing Screening Program in the Region of Murcia from 2004 to 2012.


Objective: Newborn (NB) auditory deficit has a prevalence of 1-2% in the world. Since the 1990s different screening programs have been put into practice. The Newborn Hearing Screening Program has been in operation since 2002 in our hospital (HCUVA) in Murcia (Spain) and two years later it was introduced into the whole of the Autonomous Community of the Region of Murcia as part of universal healthcare. The objective of this study was to analyze and assess its results.

Method: The newborn (NB) population is divided into two groups: not-at-risk NBs and at-risk NBs. In the first case we carry out acoustic otoemissions (AOEs) 48 h after birth and depending on the result the child is either discharged or, in negative cases, the infant undergoes a series of tests in a period of 30-45 days to confirm or rule out the existence of hearing anomalies. In the at-risk group we combine AOE with brain trunk potentials (BERA) following the stages in a decision tree diagram similar to the ones for not-at-risk children in order to provide a clinical diagnosis in the first three months of life.

Results: The screening performance was assessed for the 156,122 children studied, of which 151,258 belonged to the group of not-at-risk children; and 4664 to the group at risk of hypacusis. As a result of the screening only 410 (0.26%) were sent to consultation, 213 in the not-at-risk group (0.14%) and 197 (24.7%) in the at-risk group. A total of 7452 false positives were identified (4.7%), 6951 (4.5%) in the not-at-risk group; and 501 (10.3%) in the group with risk factors; and there were 53 false negatives (0.3%). Sensitivity in the screening program was 88.5%, with a specificity of 95%.

Conclusions: The Region of Murcia has a Newborn Hearing Screening Program with tests that provide a high level of sensitivity and specificity in accordance with the findings of the literature. Our results endorse the program and the patients were treated in a way that met the objective of providing a correct diagnosis and the appropriate therapeutic action.

de Kock T, Swanepoel D, Hall JW 3rd.

Newborn Hearing Screening at a Community-based Obstetric Unit: Screening and Diagnostic Outcomes.


Objective: Postnatal visits at community-based midwife obstetric units (MOUs) have been proposed as an alternative primary healthcare screening platform in South Africa. This study evaluated the outcomes of distortion product otoacoustic emissions (DPOAEs) and automated auditory brainstem response (AABR) screening conducted by dedicated non-professional screener at a community-based MOU in the Western Cape, South Africa.

Method: Universal newborn hearing screening (UNHS) at a community-based MOU was evaluated over a 16-month period. A dedicated non-professional screener was trained to follow a two-stage screening protocol targeting bilateral hearing loss. A two group comparative design was used alternating AABR (Maico MB11 BERaphone®) and DPOAE (Bio-logic AuDX I) technology on a daily basis. Infants referring the initial screen received a follow-up appointment in two days' time and were re-screened with the same technology used at their first screen. Those referring the second stage were booked for diagnostic assessments.

Results: 7452 infants were screened including 47.9% (n=3573) with DPOAE and 52.1% (n=3879) with AABR technology. Mean age at first stage screen was 6.1 days. The initial bilateral referral rate was significantly lower for AABR (4.6%) compared to DPOAE (7.0%) and dropped to 0.3% and 0.7% respectively following the second stage screenings. First rescreen and initial diagnostic follow-up rates of 90% and 92.3% were obtained for the DPOAE group and 86.6% and 90% for the AABR group. Follow-up rates showed no significant difference between technology groups. Diagnostic assessment revealed a higher prevalence rate for bilateral SNHL among the AABR group (1/1000) compared to the DPOAE group (0.3/1000).

Screening technology had no significant influence on daily screening capacity (23 AABR/day; 24 DPOAE/day).

Conclusions: Postnatal visits at community-based MOUs create a useful platform for hearing screening and follow-up. AABR technology with negligible disposable costs provides opportunity for AABR screening to be utilised in community-based programmes. AABR screening offers lower initial referral rates and a higher true positive rate compared to DPOAE.

Dettman S, Cho D, Dowell R.

Barriers to Early Cochlear Implantation.


Objective: Identify variables associated with paediatric access to cochlear implants (CIs).

Design: Part 1. Trends over time for age at CI surgery (N = 802) and age at hearing aid (HA) fitting (n = 487) were examined with regard to periods before, during, and after newborn hearing screening (NHS). Part 2. Demographic factors were explored for 417 children implanted under 3 years of age. Part 3. Pre-implant steps for the first 20 children to receive CIs under 12 months were examined.

Results: Part 1. Age at HA fitting and CI surgery reduced over time, and were associated with NHS implementation. Part 2. Demographic factors were explored for 417 children implanted under 3 years of age. Part 3. Pre-implant steps for the first 20 children to receive CIs under 12 months were examined.

Conclusions: NHS implementation was associated with reductions in age at device intervention in this cohort.

Eipers J, Lester C, Shinn JB, Bush ML.


Abstract: Infant hearing loss has the potential to cause significant communication impairment. Timely diagnosis and intervention is essential to preventing permanent deficits. Many infants from rural regions are delayed in diagnosis and treatment of hearing loss. The purpose of this study is to characterize the barriers in timely infant hearing healthcare for rural families following newborn newborn hearing screening (NHS) testing. Using stratified purposeful sampling, the study design involved semi-structured phone interviews with parents/guardians of children who failed NHS testing in the Appalachian region of Kentucky between 2012 and 2014 to describe their experiences with early hearing detection and intervention program. Thematic qualitative analysis was performed on interview transcripts to identify common recurring themes in content. 40 parents/guardians participated in the study and consisted primarily of mothers. Demographic data revealed limited educational levels of the participants and 70 % had state-funded insurance coverage. Participants reported barriers in timely infant hearing healthcare that included poor communication of hearing screening results, difficulty in obtaining outpatient testing, inconsistencies in healthcare information from primary care providers, lack of local resources, insurance-related healthcare delays, and conflict with family and work responsibilities. Most participants expressed a great desire to obtain timely hearing healthcare for their children and expressed a willingness to use resources such as telemedicine to obtain that care. There are multiple barriers to timely rural infant hearing healthcare. Minimizing misinformation and improving access to care are priorities to prevent delayed diagnosis.
Farzal Z, Kou YF, St John R, Shah GB, Mitchell RB.
The Role of Routine Hearing Screening in Children with Cystic Fibrosis on Aminoglycosides: A Systematic Review.
Objective: To review the role of routine hearing screening for sensorineural hearing loss (SNHL) in children with cystic fibrosis (CF) who have been on aminoglycoside therapy.
Data Sources: PubMed, Cochrane, Scopus, and Ovid databases.
Review Methods: A systematic review of the literature was performed in accordance with PRISMA (Preferred Reporting Items for Systematic Reviews and Meta-Analyses) guidelines. A comprehensive search was performed from 1970 to 2014. Randomized controlled trials, case-control studies, cohort studies, and case series including pediatric subjects with baseline auditory evaluations were included.
Results: Twelve studies (1979-2014) were reviewed. The study population included 762 children (5 months-20 years). Hearing screening measures included pure-tone audiometry (PTA) at standard ± high frequency threshold (HFPTA) (12/12), distortion product otoacoustic emissions (DPOAE) (4/12), transient-evoked otoacoustic emissions (1/12), and automated auditory brainstem response (1/12). The overall prevalence of SNHL ranged from 0% to 29%. However, on subset analysis of children with greater than 10 courses of intravenous (IV) aminoglycosides, up to 44% had SNHL. Eight studies recommended hearing screening in CF children on aminoglycosides; of these, two studies recommended screening even without aminoglycoside exposure, and four studies made no recommendations. HFPTA was the most commonly recommended screening measure followed by DPOAEs.
Conclusions: This systematic review supports a recommendation for clinicians to perform routine hearing screening in children with CF during and after aminoglycoside exposure based on the high prevalence of SNHL in this population. Future studies should define the optimal timing for hearing screening during and after aminoglycoside therapy in children with CF.

Fitzpatrick EM, Grandpierre V, Durieux-Smith A, Gaboury I, Coyle D, Na E, Sallam N.
Abstract: Children with mild bilateral and unilateral hearing loss are now commonly identified early through newborn hearing screening initiatives. There remains considerable uncertainty about how to support parents and about which services to provide for children with mild bilateral and unilateral hearing loss. The goal of this study was to learn about parents’ experiences and understand, from their perspectives, the impact of hearing loss in the mild range on the child’s functioning. Parents of 20 children in Ontario, Canada, participated in the study. The median age of identification of hearing loss was 4.6 months (interquartile range: 3.6, 10.8). Parents appreciated learning early about hearing loss, but their experiences with the early process were mixed. Parents felt that professionals minimized the importance of milder hearing loss. There was substantial uncertainty about the need for hearing aids and the findings suggest that parents need specific guidance. Parents expressed concerns about the potential impact of hearing loss on their child’s development, particularly at later ages.

Fitzpatrick EM, Hamel C, Stevens A, Pratt M, Moher D, Doucet SP, Neuss D, Bernstein A, Na E.
Sign Language and Spoken Language for Children With Hearing Loss: A Systematic Review.
Context: Permanent hearing loss affects 1 to 3 per 1000 children and interferes with typical communication development. Early detection through newborn hearing screening and hearing technology provide most children with the option of spoken language acquisition. However, no consensus exists on optimal interventions for spoken language development.
Objective: To conduct a systematic review of the effectiveness of early sign and oral language intervention compared with oral language intervention only for children with permanent hearing loss.
Data Sources: An a priori protocol was developed. Electronic databases (eg, Medline, Embase, CINAHL) from 1995 to June 2013 and gray literature sources were searched. Studies in English and French were included.
Study Selection: Two reviewers screened potentially relevant articles.
Data Extraction: Outcomes of interest were measures of auditory, vocabulary, language, and speech production skills. All data collection and risk of bias assessments were completed and then verified by a second person. Grades of Recommendation, Assessment, Development, and Evaluation (GRADE) was used to judge the strength of evidence.
Results: Eleven cohort studies met inclusion criteria, of which 8 included only children with severe to profound hearing loss with cochlear implants. Language development was the most frequently reported outcome. Other reported outcomes included speech and speech perception.
Limitations: Several measures and metrics were reported across studies, and descriptions of interventions were sometimes unclear.
Conclusions: Very limited, and hence insufficient, high-quality evidence exists to determine whether sign language in combination with oral language is more effective than oral language therapy alone. More research is needed to supplement the evidence base.

A Programme of Studies including Assessment of Diagnostic Accuracy of School Hearing Screening Tests and a Cost-effectsiveness Model of School Entry Hearing Screening Programmes.
Background: Identification of permanent hearing impairment at the earliest possible age is crucial to maximise the development of speech and language. Universal newborn hearing screening identifies the majority of the 1 in 1000 children born with a hearing impairment, but later onset can occur at any time and there is no optimum time for further screening. A universal but non-standardised school entry screening (SES) programme is in place in many parts of the UK but its value is questioned.
Objectives: To evaluate the diagnostic accuracy of hearing screening tests and the cost-effectiveness of the SES programme in the UK.
Design: Systematic review, case-control diagnostic accuracy study, comparison of routinely collected data for services with and without a SES programme, parental questionnaires, observation of practical implementation and cost-effectiveness modelling.
Setting: Second- and third-tier audiology services; community.
Participants: Children aged 4-6 years and their parents.
Main Outcome Measures: Diagnostic accuracy of two hearing screening devices, referral rate and source, yield, age at referral and cost per quality-adjusted life-year.
Results: The review of diagnostic accuracy studies concluded that research to date demonstrates marked variability in the design, methodological quality and results. The pure-tone screen (PTS) (Amplivox, Eynsham, UK) and HearCheck (HC) screener (Siemens, Frimley, UK) devices had high
sensitivity (PTS ≥89%, HC ≥83%) and specificity (PTS ≥78%, HC ≥83%) for identifying hearing impairment. The rate of referral for hearing problems was 36% lower with SES (Nottingham) relative to no SES (Cambridge) [rate ratio 0.64, 95% confidence interval (CI) 0.59 to 0.69; p<0.001]. The yield of confirmed cases did not differ between areas with and without SES (rate ratio 0.82, 95% CI 0.63 to 1.06; p=0.12). The mean age of referral did not differ between areas with and without SES for all referrals but children with confirmed hearing impairment were older at referral in the site with SES (mean age difference 0.47 years, 95% CI 0.24 to 0.70 years; p<0.001). Parental responses revealed that the consequences to the family of the referral process are minor. A SES programme is unlikely to be cost-effective and, using base-case assumptions, is dominated by a no screening strategy. A SES programme could be cost-effective if there are fewer referrals associated with SES programmes or if referrals occur more quickly with SES programmes.

Conclusions: A SES programme using the PTS or HC screener is unlikely to be effective in increasing the identified number of cases with hearing impairment and lowering the average age at identification and is therefore unlikely to represent good value for money. This finding is, however, critically dependent on the results of the observational study comparing Nottingham and Cambridge, which has limitations. The following are suggested: systematic reviews of the accuracy of devices used to measure hearing at school entry; characterisation and measurement of the cost-effectiveness of different approaches to the ad-hoc referral system; examination of programme specificity as opposed to test specificity; further observational comparative studies of different programmes; and opportunistic trials of withdrawal of SES programmes.

Gantt S, Dionne F, Kozak FK, Goshen O, Goldfarb DM, Park AH, Boppana SB, Fowler K.
Cost-effectiveness of Universal and Targeted Newborn Screening for Congenital Cytomegalovirus Infection.

Importance: Congenital cytomegalovirus (cCMV) infection is a major cause of childhood deafness. Most cCMV infections are not diagnosed without newborn screening, resulting in missed opportunities for directed care.

Objective: To estimate the cost-effectiveness of universal and targeted newborn cCMV screening programs compared with no cCMV screening.

Design, Setting, and Participants: Models were constructed using rates and outcomes from prospective cohort studies of newborn cCMV screening in US postpartum care and early hearing programs. Costs of laboratory testing, treatment, and hearing loss were drawn from Medicaid data and published estimates. The benefits of cCMV screening were assumed to come from antiviral therapy for affected newborns to reduce hearing loss and from earlier identification of hearing loss with postnatal onset. Analyses were performed from July 2014 to March 2016.

Interventions: Models compared universal or targeted cCMV screening of newborns with a failed hearing screen, with standard care for cCMV infection.

Main Outcome Measures: The incremental costs of identifying 1 cCMV infection, identifying 1 case of cCMV-related hearing loss, and preventing 1 cochlear implant; the incremental reduction in cases of severe to profound hearing loss; and the differences in costs per infant screened by universal or targeted strategies under different assumptions about the effectiveness of antiviral treatment.

Results: Among all infants born in the United States, identification of 1 case of cCMV infection by universal screening was estimated to cost $2000 to $10 000; by targeted screening, $566 to $2832. The cost of identifying 1 case of hearing loss due to cCMV was as little as $27 460 by universal screening or $975 by targeted screening. Assuming a modest benefit of antiviral treatment, screening programs were estimated to reduce severe to profound hearing loss by 4.2% to 13% and result in direct costs of $10.86 per newborn screened. However, savings of up to $37.97 per newborn screened were estimated when costs related to functionality were included.

Conclusions and Revelance: Newborn screening for cCMV infection appears to be cost-effective under a wide range of assumptions. Universal screening offers larger net savings and the greatest opportunity to provide directed care. Targeted screening also appears to be cost-effective and requires testing for fewer newborns. These findings suggest that implementation of newborn cCMV screening programs is warranted.

Gardner-Berry K, Chang H, Ching TY, Hou S.
Detection Rates of Cortical Auditory Evoked Potentials at Different Sensation Levels in Infants with Sensory/ Neural Hearing Loss and Auditory Neuropathy Spectrum Disorder.

Abstract: With the introduction of newborn hearing screening, infants are being diagnosed with hearing loss during the first few months of life. For infants with a sensory/neural hearing loss (SNHL), the audiogram can be estimated objectively using auditory brainstem response (ABR) testing and hearing aids prescribed accordingly. However, for infants with auditory neuropathy spectrum disorder (ANSD) due to the abnormal/absent ABR waveforms, alternative measures of auditory function are needed to assess the need for amplification and evaluate whether aided benefit has been achieved. Cortical auditory evoked potentials (CAEPs) are used to assess aided benefit in infants with hearing loss; however, there is insufficient information regarding the relationship between stimulus audibility and CAEP detection rates. It is also not clear whether CAEP detection rates differ between infants with SNHL and infants with ANSD. This study involved retrospective collection of CAEP, hearing threshold, and hearing aid gain data to investigate the relationship between stimulus audibility and CAEP detection rates. The results demonstrate that increases in stimulus audibility result in an increase in detection rate. For the same range of sensation levels, there was no difference in the detection rates between infants with SNHL and ANSD.

Giuntini G, Forli F, Nicoastro R, Ciabotti A, Bruschini L, Berrettini S.
Early Care in Children with Permanent Hearing Impairment.

Abstract: The implementation of regional protocols for newborn hearing screening and early audiologic diagnosis represent the first step of the entire diagnostic, rehabilitative and prosthetic programme for children with permanent hearing impairment. The maximum benefit of early diagnosis can indeed be obtained only by prompt rehabilitation aimed at fostering the child's communicative, linguistic and cognitive development. Within the framework of the CMM 2013 project of the Ministry of Health entitled "Preventing Communication Disorders: a Regional Program for Early Identification, Intervention and Care of Hearing Impaired Children", the problems concerning the promotion of the global development of children with PHI through an early rehabilitation project based on shared knowledge and scientific evidence. In this project, our specific aim was to define the features and modes of access to a precise and specialised rehabilitation project for the small hearing-impaired child within three months from the identification of later hearing loss.

Holzinger D, Weishaupt A, Fellinger P, Beitel C, Fellinger J.
Prevalence of 2.2 Per Mille of Significant Hearing Loss at School Age Suggests Rescreening After NHS.

Objectives: The study aims to determine the prevalence of different degrees of significant hearing loss in a complete sample of Austrian school-age children born between 1997 and 2001 living in the federal state of Carinthia and to evaluate the role of Newborn Hearing Screening (NHS) in the identification of later hearing loss.
Methods: In Carinthia, all school-age children with significant hearing loss (mean pure tone average in the better ear above 40 dB) are registered by the Department of Education. From five complete birth cohorts from 1997 to 2001 (n = 28,171) all the children with sensorineural hearing loss (n = 61, mean age 10.5, age range 7.5-13.6 years) were assessed for their hearing threshold and level of cognitive functioning. Socio-demographic data, including information about NHS and amplification with hearing devices, were collected from parents and teachers using structured interviews.

Results: 22 children per thousand (49.2% male) were found to be affected by significant bilateral hearing loss at school age, with 36.1% of them having a moderate hearing loss, 34.4% severe, and 29.5% profound. Fourteen children (23.0%) used cochlear implants. Their mean nonverbal IQ was 93.4 (SD 23.1), including 13.1% of children with intellectual disabilities (IQ < 70). Of those who had undergone NHS (85.2%), 50.0% had passed the screening according to parents' reports.

Conclusion: A rate of significant hearing loss in school-age children was observed which was twice the rate found in newborns. Ongoing awareness of late-onset hearing loss to improve identification and hearing screening at school entry are recommended.

Hu T, Stead K, Fu T, Papsin B.
A Program Evaluation of Kids2Hear, a Student-run Hearing Screening Program for School Children.

Background: Hearing deficits in children are demonstrably negatively associated with language acquisition and cognition. Although universal neonatal hearing screening exists, it is not offered equally across Canada. Additionally, children emigrating from other countries are often not assessed. The objective of this study is to evaluate Kids2Hear, a free hearing screening program run by medical students at elementary schools, and to determine the rate of hearing deficits that were identified and referred for evaluation.

Methods: Retrospective analysis of screening program data from 228 participants seen at three inner-city elementary schools over six months.

Results: In our sample, the mean age was 5.8 ± 1.0 years with 48% males. Approximately 21 participants (9.3%) were screened positive for a hearing deficit and required referral for supplementary audiological evaluation. About 44 participants (19.3%) were referred to a family physician for otoscopic abnormalities. Females were significantly more likely to be identified for both hearing deficits and otoscopic abnormalities.

Conclusions: Hearing deficits and otoscopic abnormalities are common among young children. Female children may be at higher risk for developing hearing issues or otoscopic abnormalities compared to males. Additional research is needed to determine the effectiveness of hearing screening programs.

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Hunter LL, Keefe DH, Feeney MP, Fitzpatrick DF, Lin L.
Longitudinal Development of Wideband Reflectance Tympanometry in Normal and At-risk Infants.

Purpose: The goals of this study were to measure normal characteristics of ambient and tympanometric wideband acoustic reflectance, which was parameterized by absorbance and group delay, in newborns cared for in well-baby and Neonatal Intensive Care Unit (NICU) nurseries, and to characterize the normal development of reflectance over the first year after birth in a group of infants with clinically normal hearing status followed longitudinally from birth to one year of age.

Methods: Infants were recruited from a well-baby and NICU nursery, passed newborn otoacoustic emissions (OAE) and automated auditory brainstem response (ABR) tests as well as follow-up diagnostic ABR and audiometry. They were tested longitudinally for up to one year using a wideband middle ear acoustic test battery consisting of tympanometry and ambient-pressure tests. Results were analyzed for ambient reflectance across frequency and tympanometric reflectance across frequency and pressure.

Results: Wideband absorbance and group delay showed large effects of age in the first 6 months. Immature absorbance and group delay patterns were apparent in the low frequencies at birth and one month, but changed substantially to a more adult-like pattern by age 6 months for both ambient and tympanometric variables. Area and length of the ear canal estimated acoustically increased up to age 1 year. Effects of race (African American and others compared to Caucasian) were found in combination with age effects. Mean and confidence intervals are provided for use as a normative longitudinal database for newborns and infants up to one year of age, for both well-baby and NICU infants.

Hunter LL, Meinzen-Derr J, Wiley S, Horvath CL, Kothari R, Wexelblatt S.
Influence of the WIC Program on Loss to Follow-up for Newborn Hearing Screening.

Background: Newborn hearing screening has a high participation rate of ~97% of infants nationally, but a high loss to follow-up of ~32% limits the effectiveness of the program. This study tested an intervention of targeted outpatient rescreening of infants through collaboration with the Women, Infants, and Children (WIC) program to improve follow-up rates for newborn hearing screen referrals.

Methods: Controlled intervention study of WIC-eligible infants who referred on newborn hearing screens at target hospitals. Hearing rescreens were performed by using screening auditory brainstem response testing by trained research assistants, coordinated with the infant's WIC appointment. Loss to follow-up rates and age at follow-up were compared with non-WIC infants tracked via the Ohio Department of Health during the same time periods at the same hospitals and at nonintervention hospitals.

Results: During a 2-year period, there were 1493 hearing screen referrals at 6 hospitals in the Cincinnati region recorded by the Ohio Department of Health. Of these, 260 WIC-eligible infants were referred to the study. Among WIC-eligible intervention infants, the lost to follow-up rate over 2 years was 9.6%, compared with 26.7% for nonintervention infants in the same hospital and 18.1% for nonintervention hospitals. The average age of hearing confirmation for the WIC intervention group was 34.8 days, compared with 63.6 days in non-WIC infants. One-third of mothers reported barriers to follow-up.

Conclusions: Collaborating with WIC to provide targeted follow-up for newborn hearing screening improved loss to follow-up rates, decreased the age at hearing confirmation by 1 month, and addressed reported care barriers.

Jeong SW, Kang MY, Kim JR, Kim LS.
Delayed-onset Hearing Loss in Pediatric Candidates for Cochlear Implantation.

Abstract: The objective of this study was to evaluate the clinical significance of delayed-onset hearing loss in children. Seventy-three children who underwent cochlear implantation (CI) were included. They were divided into a congenital hearing loss group (n = 50) and a delayed-onset hearing loss group (n = 23). The age at diagnosis of hearing loss, age at the beginning of auditory habilitation, the age at CI, and the postimplant speech perception abilities were compared between the two groups. Children in the congenital hearing loss group were confirmed to have hearing loss at a mean age of 0.3 years, and those in the delayed-onset hearing loss group were diagnosed with hearing loss at a mean age of 2.0 years. Auditory habilitation began at a mean age of 0.4 and 2.0 years, and CI was performed at a mean age of 1.4 and 2.6 years, respectively. Children in the congenital hearing loss group had better scores on speech perception tests than those in the delayed-onset hearing loss group, but the differences
were not significant. About half of the children with delayed-onset hearing loss (57 %) had risk factors associated with delayed-onset hearing loss. A high prevalence of delayed-onset hearing loss was noted in the group of children who underwent CI. Risk factors for hearing loss were not found in 43 % of children with delayed-onset hearing loss. Universal screening for delayed-onset hearing loss needs to be performed during early childhood.

Jiang ZD, Xu ZM, Wilkinson AR.

Comparison of Maturational Process of Hearing Threshold in Early Life Between At-risk and Low-risk Preterm Infants.


AIM: To detect any abnormality in the maturational process of hearing threshold during the early life in at-risk preterm infants.

Study Design: The threshold of brainstem auditory evoked response was recorded and analyzed longitudinally from 30 to 42weeks of postconceptual age in 357 at-risk infants born at 23-36weeks of gestation. The results were compared with those in 82 low-risk infants born at 30-42weeks at various postconceptual ages.

Results: From 31 to 42weeks, the response threshold in the at-risk infants was consistently slightly higher than that in the low-risk infants. No statistically significant difference was found between the two groups of infants at any designated postconceptual ages. The threshold in the at-risk infants born at 23-29weeks of gestation tended to be higher than those born at 30-36weeks at various postconceptual ages, but the difference did not reach statistical significance. There was also no significant difference in the slope of BAER threshold-age function between the at-risk infants, irrespective of gestational ages, and the low-risk infants.

Conclusions: During the early life, hearing threshold in at-risk preterm, mainly very preterm, infants is marginally elevated, but the maturational process of the threshold is generally similar to that in low-risk infants, without notable abnormality.

Kaga K.

Auditory Nerve Disease and Auditory Neuropathy Spectrum Disorders.


Abstract: In 1996, a new type of bilateral hearing disorder was discerned and published almost simultaneously by Kaga et al. [1] and Starr et al. [2]. Although the pathophysiology of this disorder as reported by each author was essentially identical, Kaga used the term “auditory nerve disease” and Starr used the term “auditory neuropathy.” Auditory neuropathy (AN) in adults is an acquired disorder characterized by mild-to-moderate pure-tone hearing loss, poor speech discrimination, and absence of the auditory brainstem response (ABR) all in the presence of normal cochlear outer hair cell function as indicated by normal distortion product otoacoustic emissions (DPOAEs) and evoked summing potentials (SPs) by electrocochleography (ECoG). A variety of processes and etiologies are thought to be involved in its pathophysiology including mutations of the OTOF and/or OPA1 genes. Most of the subsequent reports in the literature discuss the various auditory profiles of patients with AN [3,4] and in this report we present the profiles of an additional 17 cases of adult AN. Cochlear implants are useful for the reacquisition of hearing in adult AN although hearing aids are ineffective. In 2008, the new term of Auditory Neuropathy Spectrum Disorders (ANSD) was proposed by the Colorado Children’s Hospital group following a comprehensive study of newborn hearing test results. When ABRs were absent and DPOAEs were present in particular cases during newborn screening they were classified as ANSD. In 2013, our group in the Tokyo Medical Center classified ANSD into three types by following changes in ABRs and DPOAEs over time with development. In Type I, there is normalization of hearing over time. Type II shows a change into profound hearing loss and Type III is true auditory neuropathy (AN). We emphasize that, in adults, ANSD is not the same as AN.

Kanji A, Khoza-Shangase K.

Feasibility of Newborn Hearing Screening in a Public Hospital Setting in South Africa: A Pilot Study.


Objectives: The current pilot study aimed to explore the feasibility of newborn hearing screening (NHS) in a hospital setting with clinical significance for the implementation of NHS. Context-specific Objectives included determining the average time required to screen each neonate or infant; the most suitable time for initial hearing screening in the wards; as well as the ambient noise levels in the wards and at the neonatal follow-up clinic where screening would be conducted.

Method: A descriptive, longitudinal, repeated measures, within-subjects design was employed. The pilot study comprised 11 participants who underwent hearing screening. Data were analysed using descriptive statistics.

Results: The average time taken to conduct hearing screening using otoacoustic emissions and automated auditory brainstem response was 18.4 minutes, with transient evoked otoacoustic emissions taking the least time. Ambient noise levels differed between wards and clinics with the sound level readings ranging between 50 dBA and 70 dBA. The most suitable screening time was found to be the afternoons, after feeding times.

Conclusions: Findings highlight important considerations when embarking on larger scale NHS studies or when planning a hospital NHS programme. Current findings suggest that NHS can be efficiently and effectively conducted in public sector hospitals in South Africa, provided that test time is considered in addition to sensitivity and specificity when deciding on a screening protocol; bar recognised personnel challenges.

Kim SH, Choi BY, Park J, Jung EY, Cho SH, Park KH.

Maternal and Placental Factors Associated with Congenital Hearing Loss in Very Preterm Neonates.


Background: Sensorineural hearing loss (SNHL) is a multifactorial disease that more frequently affects preterm newborns. Although a number of maternal conditions have been reported to be associated with preterm birth, little information is available concerning maternal risk factors for the development of SNHL. We aimed to identify maternal and placental risk factors associated with a “refer” result on the newborn hearing screening (NHS) test and subsequently confirmed SNHL in very preterm neonates.

Methods: This retrospective cohort study included 267 singleton neonates who were born alive after ≤32 weeks. Histopathologic examination of the placenta was performed, and clinical data were retrieved from a computerized perinatal database. Cases with two abnormal findings, “refer” on the NHS test, and presence of SNHL on the confirmation test were retrospectively reviewed based on electronic medical records.

Results: Forty-two neonates (15.7%) showed a “refer” result, and, on the confirmation test, permanent SNHL was identified in 1.87% (5/267) of all neonates. Multivariate regression analysis revealed that the presence of funisitis was independently associated with a “refer” on the NHS test, whereas use of antenatal corticosteroids was statistically significantly associated with a reduced incidence of “refer” on the screening test. Neither histologic choorioamnionitis nor prematurity (as defined by low gestational age and birth weight) was associated with a “refer” on the NHS test. By contrast, multivariate analysis with occurrence of SNHL as a dependent variable identified no significant associations with the parameters studied, probably owing to the small total number of neonates with permanent SNHL.

Conclusions: Presence of funisitis was significantly and independently associated with increased risk of abnormal NHS results, while administration of antenatal corticosteroids was related to a normal NHS result. These findings support the hypothesis that a systemic fetal inflammatory response, manifested as funisitis, might play a role in the pathogenesis of SNHL in preterm neonates.
Introduction: It is assumed that the critical period for diagnosis of hearing disorders is the baby’s first three months of life and that appropriate course and implementation of treatment and/or rehabilitation should begin before a child is six months old. However various kinds of problems may occur during auditory screening of a child may exceed this interval. This problem is particularly pronounced among children with development and health problems and leads to unreliable and varied results.

Aim: The aim of this study was an analysis of prevalence of difficulties occurring during the first year of auditory screening among groups of children with congenital hearing impairment.

Material and Methods: Patients were examined in The Universal Newborn Hearing Screening Program in the years 2012 - 2013 in Level III NICUs in Krakow. Results from 250 cases were analyzed retrospectively. Medical exam results of patients with high risk of hearing loss were also included in our analysis. The groups of children included in our study were: children with Down Syndrome, children with nervous system disorders, children with cleft palate or both cleft palate and lip and children with congenital CMV.

Results: In the group of children with cleft palate or both cleft palate and lip the most frequent cause of not conducting objective audiometric tests was bad health condition of a child which precluded his arrival for administering the tests. The most common cause of difficulties in performing hearing tests was the emotional state of children from groups with Down Syndrome. In the group of children with congenital CMV the most common cause of difficulties was a lack of availability of their parents.

Conclusions: 1. We encountered the greatest diagnostic difficulties during the child’s first year of life in chosen high-risk groups of children with congenital hearing loss in children with cleft palate or both cleft palate and lip. 2. The highest prevalence of not finished tests was in III and IV interval for all chosen high-risk groups with congenital hearing loss.

Ferrán de la Cierva L, García-Purriños FJ.

Comparison of Newborn Hearing Screening in Well-Baby Nursery and NICU: A Study Applied to Reduce Referral Rate in NICU.


Objectives: To determine whether newborn hearing screening in a well-baby nursery (WBN) and neonatal intensive care unit (NICU) nursery: 1) meet three targeted, screening, referral, and diagnostic follow-up rates; 2) compare the average age of diagnosis for infants admitted to the WIN and NICU; and 3) determine prevalence of hearing loss in neonatal population; and 4) try to find a practical newborn hearing screening time algorithm to reduce refer rate in NICU.

Material and Methods: The screening rates were 99.8% and 99.6% in the WBN and NICU groups, respectively, without significant difference. The referral rates were 0.7% and 2.8% in the WBN and NICU groups, with significant difference. Furthermore, the diagnostic follow-up rates were 76.7% and 89.1% in the WBN and NICU groups without significant difference. The average initial diagnostic ages were 1.9 months and 3.8 months in the WBN and NICU groups, with significant difference. The prevalence of congenital bilateral hearing loss were 0.27% and 1.6% in the WBN and NICU groups with significant difference.

Conclusions: The screening, referral and follow-up rate in the WBN and NICU groups were equivalent to the quality indicators. For NICU group, screening and diagnostic follow up were performed later than those in WBN group; however the lower referral rate in our NICU group was successfully achieved in this study and can be applied clinically. The prevalence of congenital bilateral hearing loss was higher in the NICU group than in the WBN group.

Luz I, Ribas A, Kozlowski L, Willig M, Berberian AP.

Newborn Hearing Screening in a Public Maternity Ward in Curitiba, Brazil: Determining Factors for Not Retesting.


Introduction: Law 12.303/10 requires hearing screening in newborns before hospital discharge to detect possible hearing problems within the first three months after birth. If the newborn fails the test or presents signs of risk for hearing loss, it must undergo a retest and monitoring during the first year of life. In practice, this often does not happen.

Objective: To identify, in a group of mothers of children with risk factors for hearing loss, the determining reasons for non-compliance with the auditory retest.

Method: This is a cross-sectional quantitative study. For data collection, we handed a semi-structured questionnaire to 60 mothers of babies at risk for hearing loss who did not attend the hearing retest after hospital discharge. The questionnaire investigated their age, education, marital status, level of knowledge about the hearing screening, and reasons for non-compliance with the retest. We compared and analyzed data using the Chi-square test at a significance level of 0.05%.

Results: Our study found that 63% of the respondents were unaware of the hearing screening and most did not receive guidance on testing during prenatal care; 30% of participants stated forgetting as the reason for not attending the retest. There was no significant relationship between age, education, and marital status regarding knowledge about the test and the non-compliance with the retest.

Conclusion: Identified as the most significant determining factors for non-compliance with the newborn hearing screening retest were the surveyed mothers’ forgetting the date, and their ignorance as to the importance of retesting.

Martínez-Pacheco MC, Ferrán de la Cierva L, García-Purriños FJ.

Delayed Diagnosis of Childhood Deafness: The Value of False Negatives in the Programme for Early Detection of Neonatal Hearing Loss.


Introduction: Despite its importance, the existence of false negatives (patients who are told they hear well, but they have some degree of hipacusia) is rarely evaluated in programs for early detection of hearing loss. The aim of this study is to determine the variables that can lead to a delayed diagnosis, especially the existence of false negatives and the lack of registration of risk factors.

Method: A retrospective study of prevalence has been carried out, in which the medical records of children diagnosed with sensorineural hearing loss born within 2005 and 2012 in the health centers of study have been analyzed.

Results: Of the 32 children with sensorineural hearing loss, 16 passed the OAE, 12 did not pass the OAE, and in four they were not carried out. Of the children who passed the OAE, 57% have severe hearing loss. 66% of children with hearing loss presented a risk factor for hearing loss at birth, being the most frequent family history of hearing loss, but only 7% of those with family history of hearing loss were included in the risk group.

Conclusion: The results of the study indicate that the late diagnosis of hearing loss is related to the presence of false negatives to the OAE and the non-registration of risk factors.
Two-Way Radio Modem Data Transfer for Newborn Hearing Screening Devices.

Introduction: The success of a newborn hearing screening program depends on successful tracking and follow-up to ensure that children who have had positive screening results in the first few days of life receive appropriate and timely diagnostic and intervention services. The easy availability, through a suitable infrastructure, of the data necessary for the tracking, diagnosis, and care of children concerned is a major key to enhancing the quality and efficiency of newborn hearing screening programs.

Materials and Methods: Two systems for the automated two-way transmission of newborn hearing screening and configuration data, based on mobile communication technology, for the screening devices MADSEN AccuScreen® and Natus Echo-Screen® were developed and tested in a field study. Radio modem connections were compared with conventional analogue modem transmissions from Natus Echo-Screen devices for duration, transmission rate, number of lost connections, and frequency of use.

Results: The average session duration was significantly lower with the MADSEN AccuScreen (12 s) and Natus Echo-Screen both with radio modem (15 s) than the Natus Echo-Screen with analogue modem (108 s). The transmission rate was significantly higher (898 and 1,758 vs. 181 bytes/s) for the devices with radio modems. Both radio modem devices had significantly lower rates of broken connections after initial connection (2.1 and 0.9 vs. 5.5%). An increase in the frequency of data transmission from the clinics with mobile radio devices was found.

Conclusion: The use of mobile communication technology in newborn hearing screening devices offers improvements in the average session duration, transmission rate, and reliability of the connection over analogue solutions. We observed a behavioral change in clinical staff using the new technology: the data exchange with the tracking center is more often used. The requirements for on-site support were reduced. These savings outweigh the small increase in costs for the Internet service provider.


Abstract: Hearing loss is a relatively common condition in children, occurring in approximately 2 out of every 1,000 births with approximately 50% of reported diagnoses having a primary genetic etiology. Given the prevalence and genetic component of hearing loss, coupled with a trend toward early diagnosis with the institution of universal newborn hearing screening, The Genetics of Hearing Loss Clinic was established at The Children's Hospital of Philadelphia to manage the diagnosis, testing, and genetic counseling for individuals and families. This paper described a cohort of 660 individuals with a diagnosis of hearing loss evaluated between July 2008 and July 2015 in the Genetics of Hearing Loss Clinic. To elucidate the cause of hearing loss in this cohort for better management and prognostication, testing included single nucleotide polymorphism chromosomal microarray, hearing loss next generation sequencing panel, and additional clinical tests inclusive of thyroid and renal function studies, temporal bone magnetic resonance imaging, and electrocardiogram. Of those evaluated, most had bilateral sensorineural hearing loss, occurring in 489/660 (74%). Additionally, 612/660 (93%) of patients presented with a nonsyndromic form of hearing loss (no other observed clinical findings at the time of exam), of which pathogenic mutations in GJB2 were most prevalent. Of the individuals with syndromic manifestations (48/660), Usher and Waardenburg syndrome were most commonly observed. A family history of hearing loss (first degree relative) was present in 12.6% of families with available information. Through molecular analyses, clinical examination, and laboratory testing, a definitive etiologic diagnosis was established in 157/660 (23.8%) of individuals. © 2016 Wiley Periodicals, Inc.

Mena-Domínguez EA, Benito-Orejas JI, Ramírez-Cano B, Morais-Pérez D, Muñoz-Moreno MF. High Frequency Tympanometry (1000Hz) in Young Infants and its Comparison with Otoacoustic Emissions, Otomicroscopy and 226Hz Tympanometry.


Introduction and Objective: In the first 6 months of life, 226Hz tympanometry is considered an ineffective procedure for the diagnosis of otitis media with effusion. With the introduction of universal hearing screening, the use of high frequency 1000Hz (1kHz) tympanometry has been recommended. To optimize the diagnosis of neonatal hearing loss, we present this comparison, from the clinical point of view, of the results of 226Hz and 1kHz tympanometry in infants.

Materials and Methods: We designed a prospective study of 100 children under 9 months of age proceeding from our hearing screening program. We compare the result of tympanometry with binocular microscopy and transient evoked otoacoustic emissions.

Results: The application of transient otoacoustic emissions, otomicroscopy and 226Hz and 1kHz tympanometry has shown its usefulness in the management of otitis media with effusion of young infants, with a similar effectiveness between the 4 tests.

Conclusion: The joint use of otomicroscopy, transient otoacoustic emissions and 226Hz and 1kHz tympanometry, has allowed us to diagnose otitis media with effusion in young infants more accurately than each test separately. We recommend initial use of 1kHz tympanometry, at least in children younger than 7 months, but in the presence of hearing loss or an unclear result, 226Hz tympanometry is a good diagnostic complement.


Abstract: Human cytomegalovirus (CMV) is under-recognised, despite being the leading infectious cause of congenital malformation, affecting ~0.3% of Australian live births. Approximately 11% of infants born with congenital CMV infection are asymptomatic, resulting in clinical manifestations, including jaundice, hepatosplenomegaly, petechiae, microcephaly, intrauterine growth restriction and death. Congenital CMV infection may cause severe long-term sequelae, including progressive sensorineural hearing loss and developmental delay in 40-58% of symptomatic neonates, and ~14% of initially asymptomatic infected neonates. Up to 50% of maternal CMV infections have nonspecific clinical manifestations, and most remain undetected unless specific serological testing is undertaken. The combination of serology tests for CMV-specific IgM, IgG and IgG avidity provide improved distinction between primary and secondary maternal infections. In pregnancies with confirmed primary maternal CMV infection, amniocentesis with CMV-PCR performed on amniotic fluid, undertaken after 21-22 weeks gestation, may determine whether maternalfetal virus transmission has occurred. Ultrasound and, to a lesser extent, magnetic resonance imaging are valuable tools to assess fetal structural and growth abnormalities, although the absence of fetal abnormalities does not exclude fetal damage. Diagnosis of congenital CMV infection at birth or in the first 3 weeks of an infant's life is crucial, as this should prompt interventions for prevention of delayed-onset hearing loss and neurodevelopmental delay in affected infants. Prevention strategies should also target mothers because increased awareness and hygiene measures may reduce maternal infection. Recognition of the importance of CMV in pregnancy and in neonates is increasingly needed, particularly as therapeutic and preventive interventions expand for this serious problem.
Nittrouer S, Lowenstein JH, Holloman C.

Beyond Early Intervention: Supporting Children With CIs Through Elementary School.


Background: The development of cochlear implants (CIs) and the broader availability of early intervention, made possible by newborn hearing screening, have raised prospects that deaf children can be mainstreamed at the start of elementary school and fare well with minimal support. This report examines the veracity of that perspective.

Methods: This report specifically: (1) reviews progress made by deaf children in spoken language acquisition over the past 25 years; (2) presents data collected from 104 children in the early elementary grades (49 with normal hearing (NH) and 55 with severe-to-profound hearing loss who use CIs); (3) describes language acquisition that typically occurs in elementary school; and (4) highlights intervention strategies for school-age deaf children with CIs.

Results: The spoken language skills of deaf children have improved thanks to CIs and early intervention, but remain below those of children with NH. The amount of deficit varies across the language construct examined, with the greatest deficit found for skills dependent upon phonological (speech-sound) sensitivity, and the mildest associated with morphosyntactic (grammatical) skills. There is substantial development in both phonological and morphosyntactic skills that typically occurs during the elementary school years.

Conclusion: Both the data and theoretical models of language acquisition indicate that even with the availability of CIs and early intervention, deaf children are behind their peers with NH when they enter school. And there is much language learning that lies ahead for them. Thus, there is a need for us to enhance.

Núñez-Batalla F, Jáudenes-Casaubón C, Zubicaray-Ugarteche J, Cabanillas-Farpón R.

Aetiological Diagnosis of Child Deafness: CODEPEH Recommendations.


Abstract: Important progress in the fields of molecular genetics (principally) and diagnostic imaging, together with the lack of a consensus protocol for guiding the diagnostic process after confirming deafness by neonatal screening, have led to this new work document drafted by the Spanish Commission for the Early Detection of Child Deafness (Spanish acronym: CODEPEH). This 2015 Recommendations Document, which is based on the most recent scientific evidence, provides guidance to professionals to support them in making decisions regarding aetiological diagnosis. Such diagnosis should be performed without delay and without impeding early intervention. Early identification of the causes of deafness offers many advantages: it prevents unnecessary trouble for the families, reduces health system expenses caused by performing different tests, and provides prognostic information that may guide therapeutic actions.
Concurrent Genetic and Standard Screening for Hearing Impairment in 9317 Southern Chinese Newborns.


Objective: The goal of this study was to investigate the use of concurrent genetic screening together with standard newborn hearing screening (NHS) in an effort to provide a scientific basis for the beneficial use of concurrent genetic hearing screening in newborns. Our aim was to improve the neonatal detection rate of hearing impairment and the potential for hearing loss, allowing for increased early intervention and potentially allowing for prevention of later onset hearing loss. This information could also be used to increase the effectiveness of genetic counseling regarding hearing impairment.

Methods: A total of 9317 neonates from Children’s Hospital of Dongguan and Dongguan People’s Hospital were included in this study between January 2015 and October 2015. Twenty hotspot hearing-associated mutations of four common deafness-susceptibility genes (GJB2, GJB3, SLC26A4, and MTRNR1) were analyzed by matrix-assisted laser desorption-ionization time-of-flight mass spectrometry (MALDI-TOF-MS). The results of genetic screening and NHS were concurrently analyzed.

Results: Seventy-one midwifery education program directors were contacted, and the response rate was 38% (27 surveys). Most respondents reported that universal newborn hearing screening is discussed in the program, with the amount of time spent covering these topics varying considerably. Programs provide information about the midwife’s role in universal newborn hearing screening, legal obligation to provide hearing screening information, and tests used to complete universal newborn hearing screening. How to complete the hearing screening, counseling for parents about results, and follow-up after a newborn does not pass the screening are topics that were not often discussed. There was no influence of program type or program length on the universal newborn hearing screening content discussed.

Conclusion: The majority of midwifery education program directors that responded indicated that their programs include information about universal newborn hearing screening to midwifery students. There is a need for further information and resources specific to universal newborn hearing screening. Providing additional information to midwifery students about newborn hearing screening may result in increased awareness and education for families.
Pheian E, Pal R, Henderson L, Green KM, Bruce IA.

Introduction: Although, the association between Down syndrome (DS) and conductive hearing loss is well recognized, the fact that a small proportion of these children may have a severe to profound sensorineural hearing loss that could benefit from cochlear implantation (CI) is less well understood. The management of significant co-morbidities in children with DS can delay initial diagnosis of hearing impairment and assessment of suitability for CI can likewise be challenging, due to difficulties conditioning to behavioural hearing tests.

Methods: We performed a retrospective case note review of three children with DS referred to the Manchester Cochlear Implant Programme.

Results: Three illustrative cases are described including CI in a 4 years old. Using conventional outcome measurement instruments, the outcome could be considered to be suboptimal with a Categories of Auditory Performance score of 4 at 6 months post-op and at last follow up. In part, this is likely to reflect the delay in implantation, but the role of cognitive impairment must be considered. The cases described emphasize the importance of comprehensive radiological and audiological assessment in children with DS being considered for CI.

Conclusion: The influence of cognitive impairment upon outcome of CI must be taken into account, but should not be considered a contra-indication to implantation in children with DS. Benefit that might be considered limited when quantified using existing general outcome measurement instruments, may have a significant impact upon psychosocial development and quality of life in children with significant cognitive impairment, or other additional needs.

The Impact of Universal Newborn Hearing Screening on Long-Term Literacy Outcomes: A Prospective Cohort Study.

Objective: To determine whether the benefits of universal newborn hearing screening (UNHS) seen at age 8 years persist through the second decade.

Design: Prospective cohort study of a population sample of children with permanent childhood hearing impairment (PCHI) followed up for 17 years since birth in periods with (or without) UNHS.

Setting: Birth cohort of 100 000 in southern England.

Participants: 114 teenagers aged 13-19 years, 76 with PCHI and 38 with normal hearing. All had previously their reading assessed aged 6-10 years.

Interventions: Birth in periods with and without UNHS; confirmation of PCHI before and after age 9 months.

Main Outcome Measure: Reading comprehension ability. Regression modelling took account of severity of hearing loss, non-verbal ability, maternal education and main language.

Results: Confirmation of PCHI by age 9 months was associated with significantly higher mean z-scores for reading comprehension (adjusted mean difference 1.17, 95% CI 0.36 to 1.97) although birth during periods with UNHS was not (adjusted mean difference 0.15, 95% CI -0.75 to 1.06). The gap between the reading comprehension z-scores of teenagers with early compared with late confirmed PCHI had widened at an adjusted mean rate of 0.06 per year (95% CI 0.01-0.13) during the 9.2-year mean interval since the previous assessment.

Conclusion: The benefit to reading comprehension of confirmation of PCHI by age 9 months increases during the teenage years. This strengthens the case for UNHS programmes that lead to early confirmation of permanent hearing loss.

Pitaro J, Al Masaoudi L, Motallebzadeh H, Funnell WR, Daniel SJ.
Wideband Reflectance Measurements in Newborns: Relationship to Otoscopic Findings.

Objectives: Newborn hearing screening includes testing with otoacoustic emissions and the auditory brainstem response. Unfortunately, both tests are affected by the presence of material in the ear canal and middle ear such as vernix, meconium, and amniotic fluid. The objective of this study was to determine to what extent occlusion of the ear canal as seen on otoscopy affects wideband energy reflectance measurements in newborns. A secondary objective was to obtain additional normative wideband reflectance data in newborns.

Methods: Newborns from a well-baby nursery were enrolled. Wideband energy reflectance measurements and otoscopy were done immediately after the hearing screening. Occlusion of the ear canal as seen on otoscopy was described on a scale of 0-100.

Results: A total of 156 babies were enrolled (mean age = 25 hours). A statistically significant difference in the reflectance at ambient pressure was found between the 0-70% and 80-100% occlusion groups. There was no significant difference in reflectance between the right and the left ears. The median reflectance pattern generally followed that of previous studies but in certain frequency regions the present reflectance values were higher.

Conclusion: A significant increase in reflectance occurs when 70%-80% of the ear-canal diameter is occluded. Taking otoscopy findings into account may improve the interpretation of reflectance measurements. However, further studies are required to better establish the relationship between canal occlusion and reflectance.

Poonual W, Navacharoen N, Kangsanaraj J, Namwongprom S.
Risk Factors for Hearing Loss in Infants Under Universal Hearing Screening Program in Northern Thailand.

Objectives: To define the risk factors for hearing loss in infants (aged 3 months) under universal hearing screening program.

Materials and Methods: A total of 3,120 infants (aged 3 months) who underwent hearing screening using a universal hearing screening program using automated otoacoustic emission test between November 1, 2010 and May 31, 2012 in Uttaradit Hospital, Buddhachinaraj Hospital, and Sawanprachar Hospital (tertiary hospitals) located in Northern Thailand were included in this prospective cohort study.

Results: Of the 3,120 infants, 135 (4.3%) were confirmed to have hearing loss with the conventional otoacoustic emission test. Five of these 135 infants (3.7%) with hearing loss showed test results consistent with auditory brainstem responses. From the univariable analysis, there were eleven potential risk factors associated with hearing deterioration. On multivariable analysis, the risk factors independently associated with hearing loss at 3 months were birth weight 1,500-2,500 g (risk ratio [RR] 1.6, 95% confidence interval [CI] 1.1-2.6), APGAR score <5 at 5 minutes (RR 2.2, 95% CI 1.1-4.4), craniofacial anomalies (RR 2.6, 95% CI 1.6-4.2), sepsis (RR 1.8, 95% CI 1.0-3.2), and ototoxic exposure (RR 4.1, 95% CI 1.9-8.6).

Conclusion: This study concluded that low birth weight, APGAR score <5 at 5 minutes, craniofacial anomalies, sepsis, and ototoxic exposure are the risk factors for bilateral hearing loss in infants (aged 3 months) and proper tests should be performed to identify these risk factors. As an outcome, under the present circumstances, it is suggested that inpatient/physicians/general practitioners/health action centers/polyclinics should carry out universal hearing screening in all infants before 36 weeks. The public health policy of Thailand regarding a universal hearing screening program is important for the prevention of disability and to enhance people's quality of life.
Psarros C, Love S.
The Role of the World Health Organization’s International Classification of Functioning, Health and Disability in Models of Infant Cochlear Implant Management.

Abstract: Newborn hearing screening has led to the early diagnosis of hearing loss in neonates and early device fitting is common, based primarily on electrophysiologic and radiologic information, with some supplementary behavioral measures. Such early fitting of hearing devices, in particular cochlear implants (CIs), has been beneficial to the majority of children implanted under the age of 12 months who meet the cochlear implant candidacy criteria. Comorbidities are common in children with hearing loss, although they may not be evident in neonates and may not emerge until later in infants. Evidence suggests that the child’s outcomes are strongly influenced by a range of environmental factors including emotional and social support from the immediate and extended family. Consequently, such factors are important in service planning and service delivery for babies and children receiving CIs. The World Health Organization’s International Classification of Functioning, Health and Disability (ICF) can provide a framework to facilitate the holistic management of pediatric cochlear implant recipients. The ICF also can be used to map the progress of recipients over time to highlight emerging issues that require intervention. This article will discuss our preliminary use of the ICF to establish clinical practice; develop advocacy skills among clients and their families; identify eligibility for services such as support in educational settings; enable access to modes of service delivery such as telepractice; provide a conceptual framework for policy and program development for pediatric cochlear implant recipients (i.e., in both disability and health services); and, most importantly, establish a clear pathway for the longitudinal management of the cochlear implant in a child’s future. It is anticipated that this model will be applied to other populations receiving cochlear implants through our program.

Raine C, Atkinson H, Strachan DR, Martin JM.
Access to Cochlear Implants: Time to reflect.

Abstract: Cochlear implant (CI) intervention is expensive and accessed mainly by developed countries. The introduction of Universal Newborn Hearing Screening and funding via a public health service give children better access to CIs. However, for adults large disparities exist between utilization and estimated prevalence. In the UK CI selection criteria are restrictive compared with many other countries. Improved audiological awareness and screening programmes for adults would improve access to hearing technologies that would improve health and quality of life. Hearing loss itself has significant medical and financial burden on society and by investing in early intervention and using best technology this would mitigate some of the rising associated medical costs.

Raveh E, Ulanovski D, Attias J, Shkedy Y, Sokolov M.
Acute Mastoiditis in Children with a Cochlear Implant.

Objectives: Cochlear implantation is performed at a young age, when children are prone to acute otitis media. Acute mastoiditis is the most common complication of otitis media, but data on its management in the presence of a cochlear implant are sparse. The objective of this study was to assess the characteristics, treatment, and outcome of acute mastoiditis in children with a cochlear implant.

Methods: The medical files of all children who underwent cochlear implantation at a pediatric tertiary medical center in 2000-2014 were retrospectively reviewed. Those diagnosed with acute mastoiditis after implantation were identified, and data were collected on demographics, history, presentation, method of treatment, complications, association with untreated otitis media with effusion, and long-term middle-ear sequelae.

Results: Of the 370 children (490 ears) who underwent cochlear implantation, 13 (3.5%) were treated for acute mastoiditis (median age at acute mastoiditis, 32 months). Nine had a pre-implantation history of chronic secretory or acute recurrent otitis media, and 5 had been previously treated with ventilation tubes. In all 9 children who had unilateral cochlear implant, the acute mastoiditis episode occurred in the implanted ear. The time from implantation to mastoiditis was 5-61 months. The same treatment protocol as for normal-hearing children was followed, with special attention to the risk of central nervous system complications. Primary treatment consisted of myringotomy with intravenous administration of wide-spectrum antibiotics. Surgical drainage was performed in 8 out of 13 patients, with (n=7) or without (n=1) ventilation-tube insertion, to treat subperiosteal abscess or because of lack of symptomatic improvement. There were no cases of intracranial complications or implant involvement or need for a wider surgical approach. No middle-ear pathology was documented during the average 3.8-year follow-up.

Conclusion: The relatively high rate of acute mastoiditis and subperiosteal abscess in children with a cochlear implant, predominantly involving the implanted ear, supports the suggestion that recent masticatory may be a risk factor for these complications. Despite the frequent need for drainage, more extensive surgery is usually unnecessary, and recovery is complete and rapid. As infections can occur even years after cochlear implantation, children with otitis media should be closely followed, with possible re-introduction of ventilation tubes.
Ravi R, Gunjawate DR, Yerraguntla K, Lewis LE, Driscoll C, Rajashekhar B.
Follow-up in newborn hearing screening - A systematic review.

Introduction: The quality and efficiency of newborn hearing screening programs (NHS) rely heavily on appropriate follow-up. The Joint Committee on Infant Hearing recommends a follow-up rate of more than 95% of infants who fail the initial hearing screening. However, a 70% benchmark is considered to be more feasible. This high loss to follow-up (LTF) rate acts as a threat to the overall success of NHS programs. The objective of the study was to identify and examine the reported rates of LTF, attributed reasons for LTF and strategies undertaken to reduce LTF.

Methods: Using a systematic search, articles published between 2005 to December 2015 were identified from PubMed/Medline, Cumulative Index to Nursing and Allied Health Literature (CINAHL), Educational Resources Information Center (ERIC), Scopus,ovid, ProQuest, and Cochrane Library. To be included in the review, the study should be exploring the loss to follow-up or drop-out rate in newborn hearing screening programs and be published in an indexed peer-reviewed journal in the English language. The main outcome measures were overall rate of LTF, factors leading to LTF and measures adopted to overcome LTF.

Results: 53 articles were short-listed for data extraction. Out of these, 27 were single-centre studies, 19 were multi-centre, 3 compared multiple databases, and 4 used survey-based methods. Overall LTF rates of 20% in single-centre and 21% in multiple-centre studies were observed. Educational disparity and lack of adequate knowledge among parents were associated with LTF. The most commonly used strategy to overcome LTF suggested by studies was the use of an adequate data management system.

Conclusion: This review is a novel attempt to explore the LTF among NHS studies, reasons for LTF and strategies to reduce LTF. This review can act as a basis for planning and execution of effective NHS programs.

Ravi R, Yerraguntla K, Gunjawate DR, Rajashekhar B, Lewis LE, Guddattu V.
Knowledge and Attitude (KA) Survey Regarding Infant Hearing Loss in Karnataka, India.

Introduction: The support provided and the decisions taken by mothers determine the success of Universal Newborn Hearing Screening (UNHS). Attempts at exploring the existing knowledge-attitude among mothers is crucial to create/modify the existing screening programs. The present study attempts to explore the knowledge and attitude toward infant hearing loss (HL) among mothers of newborns in the Indian state of Karnataka.

Methods: A cross-sectional survey was conducted among 219 mothers of newborns in Karnataka, India. The questionnaire was framed from existing literature and consisted of 19 questions assessing knowledge and attitude toward infant HL to be rated on a three-point scale (no, not sure, yes). Descriptive statistics and Cronbach’s α were used to analyze the data.

Results: Mothers exhibited good knowledge of risk factors; noise (70.3%) and ear discharge (54.3%). More than 75% agreed that treatment for HL is available and that these children can attend school. The questions of superstitions and cultural beliefs yielded mixed responses. A large number of mothers expressed desire to have their children tested at birth (84.9%) and were concerned about their children’s hearing (87.7%). Yet only 54.3% stated that they would allow their children to wear hearing aids.

Summary and Conclusion: The present study is an attempt to understand the knowledge and attitude of mothers toward infant HL in Karnataka and facilitate identification of potential areas of less knowledge as a reference for endeavors of enhancement. It further highlights the need for implementing public awareness programs to improve knowledge and attitude of mothers toward infant HL for better implementation of UNHS.

Ravi R, Gunjawate DR, Yerraguntla K, Rajashekhar B, Lewis LE.
Knowledge and Attitude of Parents/Caregivers Towards Hearing Loss and Screening in Newborns - A Systematic Review.

Objective: The parents/caregivers of a newborn play a pivotal role in the process of hearing screening and intervention. The decisions taken by them depend on their knowledge and attitude. The purpose of this study was to review the literature systematically on knowledge and attitude of parents/caregivers towards infant hearing loss and newborn hearing screening.

Design: A systematic search was conducted using electronic databases for the periods from 1990 to March 2016. Two authors scrutinized the studies and extracted the data based on predetermined criteria.

Study Sample: Ten studies.

Results: Ear discharge was correctly identified as a risk factor for hearing loss along with measles, drugs/medication, family history, congenital causes and noise exposure. The studies revealed mixed results for knowledge about newborn hearing screening. Overall, the parents/caregivers showed positive attitudes towards hearing screening and intervention options. However, due to heterogeneity in the studies, it’s hard to derive a conclusion.

Summary and Conclusion: The present review sheds light on the common areas of misconception among parents/caregivers about risk factors of infant hearing loss and newborn hearing screening. The review also draws attention to the need to have more studies exploring this knowledge and attitude of parents/caregivers among diverse populations.

Ribeiro GE, Silva DP, Montovani JC.
Transient Evoked Otoacoustic Emissions and Auditory Brainstem Response in Infants with Perinatal Asphyxia.

Objective: The objective of this study was to verify the effects of perinatal asphyxia on different parts of the auditory system.

Methods: This was a non-concurrent cohort study conducted on a fixed population in a tertiary public hospital. Participants included 181 infants born at term who underwent the transient evoked otoacoustic emission test as a part of a neonatal hearing screening program, with a “pass” result in both ears, and by auditory brainstem response testing. The infants were divided into 3 groups: G1, 20 infants who had perinatal asphyxia; G2, 111 infants with an Apgar score lower than 4 in the first minute and/or lower than 6 in the fifth minute (called “low Apgar” at birth); and G3, 50 infants with first- and fifth-minute Apgar scores ≥7.

Results: The signal-to-noise ratio of transient evoked otoacoustic emissions was greater in G3 compared with G1 and G2 at 4 kHz frequency for males. An increased latency of waves I and III in the auditory brainstem response of male infants in G1 was observed.

Conclusion: This study demonstrated that alterations occurred in both the cochlear and the neural components in male infants who had perinatal asphyxia.
Objective: This update of a 2004 guideline co-developed by the American Academy of Otolaryngology-Head and Neck Surgery Foundation, the American Academy of Pediatrics, and the American Academy of Family Physicians, provides evidence-based recommendations to manage otitis media with effusion (OME), defined as the presence of fluid in the middle ear without signs or symptoms of acute ear infection. Changes from the prior guideline include consumer advocates added to the update group, evidence from 4 new clinical practice guidelines, 20 new systematic reviews, and 49 randomized control trials, enhanced emphasis on patient education and shared decision making, a new algorithm to clarify action statement relationships, and new and expanded recommendations for the diagnosis and management of OME.

Purpose: The purpose of this multidisciplinary guideline is to identify quality improvement opportunities in managing OME and to create explicit and actionable recommendations to implement these opportunities in clinical practice. Specifically, the goals are to improve diagnostic accuracy, identify children who are most susceptible to developmental sequelae from OME, and educate clinicians and patients regarding the favorable natural history of most OME and the clinical benefits for medical therapy (eg, steroids, antihistamines, decongestants). Additional goals relate to OME surveillance, hearing and language evaluation, and management of OME detected by newborn screening. The target patient for the guideline is a child aged 2 months through 12 years with OME, with or without developmental disabilities or underlying conditions that predispose to OME and its sequelae. The guideline is intended for all clinicians who are likely to diagnose and manage children with OME, and it applies to any setting in which OME would be identified, monitored, or managed. This guideline, however, does not apply to patients <2 months or >12 years old.

Action Statements: The update group made strong recommendations that clinicians (1) should document the presence of middle ear effusion with pneumatic otoscopy when diagnosing OME in a child; (2) should perform pneumatic otoscopy to assess for OME in a child with otalgia, hearing loss, or both; (3) should obtain tympanometry in children with suspected OME for whom the diagnosis is uncertain after performing (or attempting) pneumatic otoscopy; (4) should manage the child with OME who is not at risk with watchful waiting for 3 months from the date of effusion onset (if known) or 3 months from the date of diagnosis (if onset is unknown); (5) should recommend against using intranasal or systemic steroids for treating OME; (6) should recommend against using systemic antibiotics for treating OME; and (7) should recommend against using antihistamines, decongestants, or both for treating OME.

The update group made recommendations that clinicians (1) should document in the medical record counseling of parents of infants with OME who fail a newborn screening regarding the importance of follow-up to ensure that hearing is normal when OME resolves and to exclude an underlying sensorineural hearing loss; (2) should determine if a child with OME is at increased risk for speech, language, or learning problems from middle ear effusion because of baseline sensory, physical, cognitive, or behavioral factors; (3) should evaluate at-risk children for OME at the time of diagnosis of an at-risk condition and at 12 to 18 months of age (if diagnosed as being at risk prior to this time); (4) should not routinely screen children for OME who are not at risk and do not have symptoms that may be attributable to OME, such as hearing difficulties, balance (vestibular) problems, poor school performance, behavioral problems, or ear discomfort; (5) should educate children with OME and their families regarding the natural history of OME, need for follow-up, and the possible sequelae; (6) should obtain an age-appropriate hearing test if OME persists for 3 months or longer OR for OME of any duration in an at-risk child; (7) should counsel families of children with bilateral OME and documented hearing loss about the potential impact on speech and language development; (8) should reevaluate, at 3- to 6-month intervals, children with chronic OME until the effusion is no longer present, significant hearing loss is identified, or structural abnormalities of the eardrum or middle ear are suspected; (9) should recommend tympanostomy tubes when surgery is performed for OME in a child <4 years old; adenoidectomy should not be performed unless a distinct indication exists (nasal obstruction, chronic adenoiditis); (10) should recommend tympanostomy tubes, adenoidectomy, or both when surgery is performed for OME in a child ≥4 years old; and (11) should document resolution of OME, improved hearing, or improved quality of life when managing a child with OME.
Abstract: Hearing loss is the most common sensory deficit in humans, affecting 1 in 500 newborns. Due to its genetic heterogeneity, comprehensive diagnostic testing has not previously been completed in a large multiethnic cohort. To determine the aggregate contribution inheritance makes to nonsyndromic hearing loss, we performed comprehensive clinical genetic testing with targeted genomic enrichment and massively parallel sequencing on 1119 sequentially accrued patients. No patient was excluded based on phenotype, inheritance or previous testing. Testing resulted in identification of the underlying genetic cause for hearing loss in 440 patients (39%). Pathogenic variants were found in 49 genes and included missense variants (49%), large copy number changes (18%), small insertions and deletions (18%), nonsense variants (8%), splice-site alterations (6%), and promoter variants (<1%). The diagnostic rate varied considerably based on phenotype and was highest for patients with a positive family history of hearing loss or when the loss was congenital and symmetric. The spectrum of implicated genes showed wide ethnic variability. These findings support the more efficient utilization of medical resources through the development of evidence-based algorithms for the diagnosis of hearing loss.

Results: Brain MRI investigations are important for making a diagnosis and formulating an intellectual prognosis. Analysis of umbilical cord tissue represents a unique and useful way to retrospectively diagnose congenital CMV infection.
Van As JW, Van den Berg H, Van Dalen EC.

**Platinum-Induced Hearing Loss After Treatment for Childhood Cancer.**


**Background:** Platinum-based therapy, including cisplatin, carboplatin, oxaliplatin or a combination of these, is used to treat a variety of paediatric malignancies. Unfortunately, one of the most important adverse effects is the occurrence of hearing loss or ototoxicity. There is a wide variation in the reported prevalence of platinum-induced ototoxicity and the associated risk factors. More insight into the prevalence of and risk factors for platinum-induced hearing loss is essential in order to develop less ototoxic treatment protocols for the future treatment of children with cancer and to develop adequate follow-up protocols for childhood cancer survivors treated with platinum-based therapy.

**Objective:** To evaluate the existing evidence on the association between childhood cancer treatment including platinum analogues and the occurrence of hearing loss.

**Search Methods:** We searched the Cochrane Central Register of Controlled Trials (CENTRAL) (2015, Issue 8), MEDLINE (PubMed) (1945 to 23 September 2015) and EMBASE (Ovid) (1980 to 23 September 2015). In addition, we searched reference lists of relevant articles and the conference proceedings of the International Society for Paediatric Oncology (2008 to 2014), the American Society of Pediatric Hematology/Oncology (2008 to 2015) and the International Conference on Long-Term Complications of Treatment of Children and Adolescents for Cancer (2010 to 2015). Experts in the field provided information on additional studies.

**Selection Criteria:** All study designs, except case reports, case series (i.e. a description of non-consecutive participants) and studies including fewer than 100 participants treated with platinum-based therapy who had an ototoxicity assessment, examining the association between childhood cancer treatment including platinum analogues and the occurrence of hearing loss.

**Data Collection and Analysis:** Two review authors independently performed the study selection. One review author performed data extraction and risk of bias assessment, which was checked by another review author.

**Main Results:** We identified 13 eligible cohort studies including 2837 participants with a hearing test after treatment with a platinum analogue for different types of childhood cancers. All studies had methodological limitations, with regard to both internal (risk of bias) and external validity. Participants were treated with cisplatin, carboplatin or both, in varying doses. The reported prevalence of hearing loss varied considerably between 0% and 101%. None of the studies provided data on tinnitus. Three studies reported a prevalence of 0%, but none of these studies provided a definition for hearing loss and there might be substantial or even complete overlap in included participants between these three studies. When only studies that did provide a definition for hearing loss were included, the prevalence of hearing loss still varied widely between 1.7% and 90.1%. All studies were very heterogeneous with regard to, for example, definitions of hearing loss, used diagnostic tests, participant characteristics, (prior) anti-tumour treatment, other ototoxic drugs and length of follow-up. Therefore, pooling of results was not possible. Only two studies included a control group of people who had not received platinum treatment. In one study, the prevalence of hearing loss was 67.1% (95% confidence interval (CI) 59.3% to 74.1%) in platinum-treated participants, while in the control participants it was 7.4% (95% CI 6.2% to 8.8%). However, hearing loss was detected by screening in survivors treated with platinum analogues and by clinical presentation in control participants. It is uncertain what the effect of this difference in follow-up/diagnostic testing was. In the other study, the prevalence of hearing loss was 20.1% (95% CI 17.4% to 23.2%) in platinum-treated participants and 0.4% (95% CI 0.12% to 1.6%) in control participants. As neither study was a randomized controlled trial or controlled clinical trial, the calculation of a risk ratio was not feasible as it is very likely that both groups differed more than only the platinum treatment. Only two studies evaluated possible risk factors using multivariable analysis. One study identified a significantly higher risk of hearing loss in people treated with cisplatin 400 mg/m(2) plus carboplatin 1700 mg/m(2) as compared to treatment with cisplatin 400 mg/m(2) or less, irrespective of the definition of hearing loss. They also identified a significantly higher risk of hearing loss in people treated with non-anthracycline aminoglycosides antibiotics (using a surrogate marker) as compared to people not administered with these, for three out of four definitions of hearing loss. The other study reported that age at treatment (odds ratio less than 1 for each single-unit increase) and single maximum cisplatin dose (odds ratio greater than 1 for each single-unit increase) were significant predictors for hearing loss, while gender was not.

**Authors’ Conclusions:** This systematic review shows that children treated with platinum analogues are at risk for developing hearing loss, but the exact prevalence and risk factors remain unclear. There were no data available for tinnitus. Based on the currently available evidence we can only advise that children treated with platinum analogues are screened for ototoxicity in order to make it possible to diagnose hearing loss early and to take appropriate measures. However, we are unable to give recommendations for specific follow-up protocols including frequency of testing. Counselling regarding the prevention of noise pollution can be considered, such as the use of noise-limiting equipment, avoiding careers with excess noise and ototoxic medication. Before definitive conclusions on the prevalence and associated risk factors of platinum-induced ototoxicity can be made, more high-quality research is needed. Accurate and transparent reporting of findings will make it possible for readers to appraise the results of these studies critically.

Vashistha I, Aseri Y, Singh BK, Verma PC.

**Prevalence of Hearing Impairment in High Risk Infants.**


**Abstract:** Hearing impairment is prevalent in the general population, early intervention facilitates proper development. The aim of this study was to establish the prevalence of hearing impairment in high-risk infants born between 2013 and 2014. 100 newborns were evaluated using evoked otoacoustic emissions and distortion produce and auditory behavior. Tests were reported if the results were altered. If altered results persisted, the child was referred for impedance testing and when necessary for medical evaluation. Infants referred for BOA and OAE undergone Brainstem auditory evoked potential testing. Of 100 children, 85 children have hearing within normal limits. Hearing impairment was found in 15 out of which 7 had unilateral hearing loss and 8 had bilateral hearing loss. The high prevalence of hearing impairment in this population underlines the importance of early audiological testing.

Vos B, Senterre C, Lagasse R, Tognola G, Levêque A.

**Organization of Newborn Hearing Screening Programmes in the European Union: Widely Implemented, Differently Performed.**


**Background:** Implementation of newborn hearing screening programmes is widely recommended and programme organisational designs may differ in practice. The objective of this article was to establish an overview of the newborn hearing screening programmes in the 28 countries of the European Union on four topics (policy-decision, financing, general designs, organisational features).

**Methods:** National or regional programme coordinators completed an online self-administered questionnaire focusing on protocol description and programme organization.

**Results:** Thirty-nine key informants, representing 24 countries, from national or regional levels completed the questionnaire. Newborn hearing screening programmes are or will be shortly implemented largely in the European Union countries. Levels of policy decision-making and organisational decisions are diverse (national, regional or combined). Designs of the programmes (number of steps before diagnosis referral, single
or dual target group protocol) highly varied. However, common organisational elements were observed: hearing screening tests are often performed by nursing staff, in hospitals and early in life. This pattern does not apply when a screening protocol is specifically implemented for newborns with risk factor(s) for hearing impairment or admitted to neonatal intensive care units. Hearing test financing frequently involved public sources, including government and public health funds.

Conclusion: Despite the same goal of early identification of hearing-impaired children, there is a high level of diversity in programmes, including policy decisions, financing, general designs and pragmatic organisational choices (e.g. professionals involved, location or time for screening, number of steps in the protocol). Further investigations should analyse these differences in relation to the programmes’ contexts and outcomes.

Voss SE, Herrmann BS, Horton NJ, Amadei EA, Kujawa SG.


Objective: The objective is to develop methods to utilize newborn reflectance measures for the identification of middle-ear transient conditions (e.g., middle-ear fluid) during the newborn period and ultimately during the first few months of life. Transient middle-ear conditions are a suspected source of failure to pass a newborn hearing screening. The ability to identify a conductive loss during the screening procedure could enable the referred ear to be either (1) cleared of a middle-ear condition and recommended for more extensive hearing assessment as soon as possible, or (2) suspected of a transient middle-ear condition, and if desired, be rescreeened before more extensive hearing assessment.

Design: Reflectance measurements are reported from full-term, healthy, newborn babies in which one ear referred and one ear passed an initial auditory brainstem response newborn hearing screening and a subsequent distortion product otoacoustic emission screening on the same day. These same subjects returned for a detailed follow-up evaluation at age 1 month (range 14 to 35 days). In total, measurements were made on 30 subjects who had a unilateral refer near birth (during their first 2 days of life) and bilateral normal hearing at follow-up (about 1 month old). Three specific comparisons were made: (1) Association of ear’s state with power reflectance near birth (referred versus passed ear), (2) Changes in power reflectance of normal ears between newborn and 1 month old (maturation effects), and (3) Association of ear’s newborn state (referred versus passed) with ear’s power reflectance at 1 month. In addition to these measurements, a set of preliminary data selection criteria were developed to ensure that analyzed data were not corrupted by acoustic leaks and other measurement problems.

Results: Within 2 days of birth, the power reflectance measured in newborn ears with transient middle-ear conditions (referred newborn hearing screening and passed hearing assessment at age 1 month) was significantly greater than power reflectance on newborn ears that passed the newborn hearing screening across all frequencies (500 to 6000 Hz). Changes in power reflectance in normal ears from newborn to 1 month appear in approximately the 2000 to 5000 Hz range but are not present at other frequencies. The power reflectance at age 1 month does not depend significantly on the ear’s state near birth (refer or pass hearing screening) for frequencies above 700 Hz; there might be small differences at lower frequencies.

Conclusion: Power reflectance measurements are significantly different for ears that pass newborn hearing screening and ears that refer with middle-ear transient conditions. At age 1 month, about 90% of ears that referred at birth passed an auditory brainstem response hearing evaluation; within these ears the power reflectance at 1 month did not differ between the ear that initially referred at birth and the ear that passed the hearing screening at birth for frequencies above 700 Hz. This study also proposes a preliminary set of criteria for determining when reflectance measures on young babies are corrupted by acoustic leaks, probes against the ear canal, or other measurement problems. Specifically proposed are “data selection criteria” that depend on the power reflectance, impedance magnitude, and impedance angle. Additional data collected in the future are needed to improve and test these proposed criteria.

Wake M, Ching TY, Wirth K, Poulakis Z, Mensah FK, Gold L, King A, Bryson HE, Reilly S, Rickards F.


Background: Universal newborn hearing screening was implemented worldwide largely on modeled, not measured, long-term benefits. Comparative quantification of population benefits would justify its high cost.

Methods: Natural experiment comparing 3 population approaches to detecting bilateral congenital hearing loss (>25 dB, better ear) in Australian states with similar demographics and services: (1) universal newborn hearing screening, New South Wales 2003-2005, n = 69; (2) Risk factor screening (neonatal intensive care screening + universal risk factor referral), Victoria 2003-2005, n = 65; and (3) largely opportunistic detection, Victoria 1991-1993, n = 86. Children in (1) and (2) were followed at age 5 to 6 years and in (3) at 7 to 8 years. Outcomes were compared between states using adjusted linear regression.

Results: Children were diagnosed younger with universal than risk factor screening (adjusted mean difference -8.0 months, 95% confidence interval -12.3 to -3.7). For children without intellectual disability, moving from opportunistic to risk factor to universal screening incrementally improved age of diagnosis (22.5 vs 16.2 vs 8.1 months, P < .001), receptive (81.8 vs 83.0 vs 88.9, P = .05) and expressive (74.9 vs 80.7 vs 89.3, P < .001) language and receptive vocabulary (79.4 vs 83.8 vs 91.5, P < .001); these nonetheless remained well short of cognition (mean 103.4, SD 15.2). Behavior and health-related quality of life were unaffected.

Conclusion: With new randomized trials unlikely, this may represent the most definitive population-based evidence supporting universal newborn hearing screening. Although outperforming risk factor screening, school entry language still lagged cognitive abilities by nearly a SD. Prompt intervention and efficacy research are needed for children to reach their potential.

Walker E, McCrery R, Spratford M, Roush P.


Background: Up to 15% of children with permanent hearing loss (HL) have auditory neuropathy spectrum disorder (ANSd), which involves normal outer hair cell function and disordered afferent neural activity in the auditory nerve or brainstem. Given the varying presentations of ANSd in children, there is a need for more evidence-based research on appropriate clinical interventions for this population.

Purpose: This study compared the speech production, speech perception, and language outcomes of children with ANSd, who are hard of hearing, to children with similar degrees of mild-to-moderately severe sensorineural hearing loss (SNHL), all of whom were fitted with bilateral hearing aids (HAs) based on the American Academy of Audiology pediatric amplification guidelines.

Research Design: Speech perception and communication outcomes data were gathered in a prospective accelerated longitudinal design, with entry into the programme between 6 mo and second birthday. Three children involved in participant recruitment: Boys Town National Research Hospital, the University of North Carolina at Chapel Hill, and the University of Iowa.

Study Sample: The sample consisted of 12 children with ANSd and 22 children with SNHL. The groups were matched based on better-ear pure-tone average, better-ear aided speech intelligibility index, gender, maternal education level, and newborn hearing screening result (i.e., pass or refer).
**Data Collection and Analysis:** Children and their families participated in an initial baseline visit, followed by visits twice a year for children <2 yr of age and once a yr for children ≥2 yr of age. Paired-sample t-tests were used to compare children with ANSD to children with SNHL.

**Results:** Paired t-tests indicated no significant differences between the ANSD and SNHL groups on language and articulation measures. Children with ANSD displayed functional speech perception skills in quiet. Although the number of participants was too small to conduct statistical analyses for speech perception testing, there appeared to be a trend in which the ANSD group performed more poorly in background noise with HAs, compared to the SNHL group.

**Conclusion:** The American Academy of Audiology Pediatric Amplification Guidelines recommend that children with ANSD receive an HA trial if their behavioral thresholds are sufficiently high enough to impede speech perception at conversational levels. For children with ANSD in the mild-to-severe HL range, the current results support this recommendation, as children with ANSD can achieve functional outcomes similar to peers with SNHL.

**Wu CC, Tsai CH, Hung CC, Lin YH, Lin YH, Huang FL, Tsao PN, Su YN, Lee YL, Hsieh WS, Hsu CJ.**

**Newborn Genetic Screening for Hearing Impairment: A Population-Based Longitudinal Study.**


**Purpose:** The feasibility of genetic screening for deafness-causing mutations in newborns has been reported in several studies. The aim of this study was to investigate the long-term results in those who screened positive for deafness mutations; these results are crucial to determine the cost-effectiveness to justify population-wide genetic screening.

**Methods:** We performed simultaneous hearing screening and genetic screening targeting four common deafness mutations (p.V37I and c.235delC of GJB2, c.919-2A>G of SLC26A4, and the mitochondrial m.1555A>G) in 5173 newborns at a tertiary hospital between 2009 and 2015. Serial audiometric results up to 6 years old were then analyzed in children with conclusive genotypes.

**Results:** Newborn genetic screening identified 82 (1.6%) babies with conclusive genotypes, comprising 62 (1.2%) with GJB2 p.V37I/p.V37I, 16 (0.3%) with GJB2 p.V37I/c.235delC, and 4 (0.1%) with m.1555A>G. Of these, 46 (56.1%) passed hearing screening at birth. Long-term follow-up demonstrated progressive hearing loss in children with the GJB2 p.V37I/p.V37I and p.V37I/c.235delC genotypes; this hearing loss deteriorated by approximately 1 decibel hearing level (dBHL) per year.

**Conclusion:** We delineated the longitudinal auditory features of the highly prevalent GJB2 p.V37I mutation on a general population basis and confirmed the utility of newborn genetic screening in identifying infants with late-onset or progressive hearing impairment undetectable by newborn hearing screening. Genet Med advance online publication 16 June 2016 Genetics in Medicine (2016); doi:10.1038/gim.2016.66

**Yoshinaga-Itano C, Wiggin M.**

**A Look into the Crystal Ball for Children Who Are Deaf or Hard of Hearing: Needs, Opportunities, and Challenges.**


**Abstract:** Hearing is essential for the development of speech, spoken language, and listening skills. Children previously went undiagnosed with hearing loss until they were 2.5 or 3 years of age. The auditory deprivation during this critical period of development significantly impacted long-term listening and spoken language outcomes. Due to the advent of universal newborn hearing screening, the average age of diagnosis has dropped to the first few months of life, which sets the stage for outcomes that include children with speech, spoken language, and auditory skill testing in the normal range. However, our work is not finished. The future holds even greater possibilities for children with hearing loss.

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**Yılmazer R, Yazıcı MZ, Erdim İ, Kaya HK, Özcan Dalbudak Ş, Kayhan TF.**

**Follow-Up Results of Newborns after Hearing Screening at a Training and Research Hospital in Turkey.**


**Objective:** This study aimed to present the follow-up results of newborns after universal newborn hearing screening at a Training and Research Hospital in Istanbul and to determine the ages of diagnosis, hearing aid fitting, and cochlear implantation in newborns with hearing loss.

**Materials and Methods:** A total of 5985 newborns were screened between December 2009 and August 2011 using the transient evoked otoacoustic emission test as the first two steps and automated auditory brainstem response (ABR) test as the third step. Newborns who failed the screening tests were referred to a tertiary hospital for clinic ABR and were followed up at least for 2 years.

**Results:** Of 5985 newborns, 5116 (85.5%) completed the screening. Of 53 newborns who were referred to a tertiary hospital, 13 (0.25%) had a hearing impairment. The mean age of diagnosis, hearing aid fitting, and cochlear implantation were 6.1, 9.5, and 24.5 months, respectively. Among the risk factors for hearing impairment, neonatal intensive care (60%) and consanguineous marriage (50%) were the most common ones that were encountered.

**Conclusion:** Our results were consistent with the national literature. Consanguineous marriage may be a risk factor for hearing impairment where it is commonly practiced because consanguineous marriage is significantly high in parents of deaf children. The ages of diagnosis and hearing aid fitting are still beyond the recommended ages by the Joint Committee on Infant Hearing.