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Announcing the new EHDI Learning Center eBook for Professionals and Families

In early September, the National Center for Hearing Assessment and Management (NCHAM) posted a new eBook, Preparing to Teach, Committing to Learn: An Introduction to Educating Students Who Are Deaf/Hard of Hearing, edited by Susan Lenihan, Professor and Director of Deaf Education at Fontbonne University. Lenihan was motivated to create the eBook by the dramatic changes in deaf education over the last 50 years. Advances in technology, especially newborn hearing screening and cochlear implants, changed the educational experiences of many children who are deaf or hard of hearing (DHH). The Individuals with Disabilities Education Act (IDEA) changed the way teachers and families plan for the education of students who are DHH. Research on practices for developing communication skills, literacy, and academic achievement provided evidence-based strategies for teaching these students and coaching their caregivers. These changes require that professionals prepare to teach by studying current knowledge and strategies and applying that learning under the guidance of skilled mentors. Lenihan invited colleagues to submit chapters on important topics in deaf education to provide an overview of current educational services for children who are DHH.

The goal for the text is to provide an open source eBook on deaf education that is available to students and faculty in professional preparation programs, practitioners in deaf education, and families of children who are DHH. Although the focus is on deaf-education teachers who are providing services to students who are DHH, the text is useful to students and faculty in related fields including speech-language pathology, audiology, and special education. Because the text is introductory in nature it covers a broad range of topics and does not replace texts that go into more depth on a particular aspect of deaf education such as literacy. The text includes valuable references and additional recommended readings and resources for further study. The text addresses the range of communication options used by students in deaf education, however, the emphasis is on listening and spoken language approaches and strategies which have often been only minimally addressed in introductory texts. References and resources for further study of approaches that are primarily visual are included.

There are many benefits to an eBook. Including:

- The free, digital medium allows students, professionals and families access to the content, reducing barriers to receiving information and implementing evidence-based strategies across a variety of settings and regions.
- The chapters are flexible in that they may be used as a whole text or individually for particular topics in a course.
- To meet the preferences of students and professionals in the digital age, the content may be downloaded to a computer thus providing a “green” alternative to paper.
- Since the text is in eBook format, the goal is to update the text regularly to reflect new research and changes in technology and services.
- This eBook also allows readers to explore certain topics in more depth through embedded links. These links will be regularly monitored and updated as needed.
- The eBook format also allows for the addition of appendices as needed and for the inclusion of video in the future which will provide examples of many of the strategies and concepts.
- The eBook format also allows for an interactive component through which readers may share ideas and comments for new content and updates.

Ultimately, relevant resources and applicable knowledge drive the preparation of well-rounded, collaborative professionals. Without access to current content, future teachers and practitioners cannot engage in a dialogue about how to best serve children who are DHH. This eBook can meet those needs by providing additional locations to seek further knowledge.

The collaborative model of *Preparing to Teach, Committing to Learn* allowed top academics and professionals in the field to share their expertise about the chapter topics. The authors include faculty in higher education programs, researchers, and practitioners in deaf education. Five of the contributors are DHH and three more have family members who are DHH. Most of the authors are professionals with many years of experience but a number of young professionals also contributed to the project. To learn more about the contributors, see the author information section of the text.

The eBook provides valuable content and resources for EHDI professionals and families of children who are DHH. Here are somehighlights from the different chapters.

- Chapter 1: Stacy Lim and Don Goldberg provide an overview of audiology with a user-friendly resource, “alphabet soup of audiology”. Stephanie Gardiner-Walsh and Susan Lenihan describe.
- Chapter 2: Resources such as The Radical Middle and the Common Ground Project to encourage awareness of options in communication modality and stress that the choice should be individualized and based on the multifaceted needs and values of the family and the child.
- Chapter 3: Uma Soman describes the latest research on brain development and executive function and how cognitive skills are impacted by deafness.
- Chapter 4: Joni Alberg explores the importance of social and emotional development. She writes, “This is our goal for all children—to become socially accepted and participating members of the communities in which they live and learn.”
- Chapter 5: Janice Gatty, who has supported families and children for many years through family-centered services contributed a chapter on families with a developmental and literary perspective.
- Chapter 6: Colleagues from Galluadet University, Heather Zimmerman and Thomas Horejes collaborated to describe the history of deaf education in Europe and the United States with a focus on the implications of language and culture (or what they call languaculture).

Chapters 7 through 9 address strategies and approaches for early intervention and for educating young children who are DHH.

- Chapter 7: Sherri Fickenscher and Dan Salvucci provide a set of strategies used to develop listening and spoken language for children whose families have chosen this approach.
- Chapter 8: Jenna Voss and Arlene Stredler-Brown address the ways in which practitioners provide early intervention services with a family-centered focus, including specific examples of evidence-based practices in coaching caregivers.
- Chapter 9: Ellie White describes a model preschool using a listening and spoken language approach including detailed information about curriculum programming to support language, literacy, speech, and listening.

Educational placement decisions, instructional planning, and the more prevalent role of itinerant teaching are addressed in Chapters 10, 11 and 12.

- Chapter 10: The Individuals with Disabilities Education Act (IDEA) and the variety of educational settings are described by practitioner Dawn Gettemeier.
- Chapter 11: Lauri Nelson and Blane Trautwein explain the importance of effective assessment and planning and provide numerous examples.
- Chapter 12: John Luckner describes the challenges and joys of being an itinerant teacher, stating, “Accepting change as a constant, not allowing oneself to get irritated and seeking to be a collaborative problem solver are essential behaviors.”

Chapters 13 through 16 address key topics for educators in deaf education: literacy development, students with disabilities who are also deaf/hard of hearing, the importance of ongoing professional learning, and career development.

- Chapter 13: Paula Gross and Lyn Robertson describe the ways in which students who are DHH develop the critical literacy skills of language comprehension and word recognition which will allow students to succeed academically.
- Chapter 14: Christy Borders and colleagues from Illinois State University use case studies to encourage readers to apply a framework and strategies for optimizing the development of students who are deaf with disabilities.
- Chapter 15: Sarah Ammerman and Mary Ellen Nevins guide the reader in developing the dispositions needed to be a reflective practitioner who is continually learning.
- Chapter 16: Megan Reister shares her personal story of growing up and finding her career path as a person who is deaf while also explaining the laws that support employment for individuals who are DHH.

The authors hope that professionals and families find current, relevant, and useful content in Preparing to Teach, Committing to Learn: An Introduction to Educating Students Who Are Deaf/Hard of Hearing and will return to the eBook for updates and new resources. This great resource can be found on the NCHAM website: http://www.infanthearing.org/ebook-educating-children-dhh/index.html

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Using Data to Improve Services for Infants with Hearing Loss: Linking Newborn Hearing Screening Records with Early Intervention Records

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Abstract: The purpose of this study was to match records of infants with permanent hearing loss from the New York Early Hearing Detection and Intervention Information System (NYEHDI-IS) to records of infants with permanent hearing loss receiving early intervention services from the New York State Early Intervention Program (NYSEIP) to identify areas in the state where hearing screening, diagnostic evaluations, and referrals to the NYSEIP were not being made or documented in a timely manner. Data from 2014–2016 NYEHDI-IS and New York Early Intervention System (NYEIS) information systems were matched using The Link King. There were 274 infants documented in NYEIS Information System as having received early intervention services, but not having documentation of failed hearing screening (n = 103) or a diagnostic evaluation confirming hearing loss (n = 171) in NYEHDI-IS. There were 40 infants with hearing loss in NYEHDI-IS who were not referred to NYSEIP, and 19 of these infants' providers documented in NYEHDI-IS that a referral to NYSEIP was made. The results from these analyses were used to direct targeted technical assistance to audiologists to educate them about the importance of early identification and referral and the reporting requirements to the New York State Department of Health with the goal of improving NYSEIP and the NYEHDI Program.

Key Words: data match, EHDI, early intervention, loss to follow-up, hearing loss

Acronyms: CDC = Centers for Disease Control and Prevention; EHDI = Early Hearing Detection and Intervention; MOU = Memorandum of Understanding; NYEHDI-IS = New York Early Hearing Detection and Intervention Information System; NYEIS = New York Early Intervention System; NYS = New York State; NYSEIP = New York State Early Intervention Program;

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Background

Hearing loss has been demonstrated to have a negative impact on a young child's speech and language development. Without early diagnosis and enrollment in early intervention services, a child may have persistent delays in social-emotional development and learning that could impact school readiness (Moeller, 2000; Yoshinaga-Itano & Gravel, 2001; Yoshinaga-Itano, Sedey, Coulter, & Mehl, 1998).

State Early Hearing Detection and Intervention (EHDI) programs are tasked with ensuring the timely identification of young children and access to appropriate diagnostic and therapeutic supports and services. Each year the states' performance in achieving the Joint Committee on Infant Hearing guidelines is published on the Center for Disease Control and Prevention's (CDC, 2017) website. State performance in 2014 ranged greatly across these measures, with the greatest variation in documentation of diagnostic evaluation before three months of age (0 to 100%) and enrollment in early intervention services (0 to 100%; CDC, 2016a, 2016b).

Risk factors such as having a low socioeconomic status, missed newborn hearing screening, lack of parent and primary care provider education on the importance of early intervention, lack of family involvement, hearing

loss diagnosed at a late age, or living in a rural residential area have shown to be related to discrepancies in early intervention enrollment, service provision among deaf or hard of hearing children, or both (Boss, Niparko, Gaskin, & Levinson, 2011; Bush, Burton, Loan, & Shinn, 2013; Lester, Dawson, Gantz, & Hansen, 2011; Moeller, 2000).

Moeller (2000), Yoshinaga-Itano & Gravel (2001), and Yoshinaga-Itano et al. (1998) found that the earlier children enroll in early intervention programs, the better their development of vocabulary, verbal reasoning, speech, and social-emotional skills than later enrolled children. Since the most intensive period of acquiring these skills occurs during a child's first three years of life, children whose hearing loss goes undetected often have significant language, speech, and social delays (American Speech-Language-Hearing Association, n.d.; Culbertson & Gilbert, 1986; Moeller, 2000; National Institute on Deafness and Other Communication Disorders (2017). Before the implementation of universal newborn hearing screening, the average age of hearing loss detection for many children was two to three years of age (Shulman et al., 2010).

In states with large populations served by many professionals, the task of directly engaging all primary care clinicians, early intervention providers, and audiologists is significant and may not be feasible within the existing infrastructure. In New York, approximately 240,000 babies are born each year. There are over 5,200 family physicians; 6,700 pediatricians; 1,300 audiologists; and 14,000 Early Intervention Program providers. The New York State Early Hearing Detection (NYEHDI) Program identified the need to better use existing data sources, such as the state's early intervention program, to identify gaps and target technical assistance to address those gaps.

Data matching has been widely used by public health researchers to conduct routine assessments, evaluations, and research studies, such as reviewing causes of infant deaths, identifying people with co-infections for case management and epidemiologic surveillance, evaluating immunization compliance for children with Medicaid, monitoring pregnancy-associated hospitalizations, and evaluation of Medicaid program coverage and impact (Qayad & Zhang, 2009; Xia, Braunstein, Stadelmann, Pathela, & Torian, 2014). Matching has also been used to improve the completeness of registry data by adding demographic, clinical, or behavioral information obtained from other registries on cases of HIV co-infection (Xia et al., 2014). There is very little information related to the linkage of newborn hearing screening and early intervention programs.

The objective of this study was to demonstrate the feasibility of the match between the NYEHDI program and the New York State Early Intervention Program (NYSEIP) data, and demonstrate the potential to direct targeted technical assistance to improve the state system of identification of hearing loss and access to early intervention services.

Method

Data Use Agreement

A data use agreement and Memorandum of Understanding (MOU) are in place with the state's two vital record systems, Statewide Perinatal Data System and Electronic Vital Events Registration System, and NYEHDI-IS. The vital records systems include pertinent information for all newborns recorded on the birth certificate including the hearing screening results. The NYEHDI Program and NYSEIP are administered by the Bureau of Early Intervention in the New York State Department of Health (NYSDOH), so data use agreements were not required for the data match.

Data Systems

NYEHDI-IS was built by the NYSDOH as an application that is connected to and leverages the New York State (NYS) Immunization Information System and integrates the state's two vital record systems. The NYEHDI-IS was deployed statewide in June 2014. All infant records are included in NYEHDI-IS, regardless of whether they had a hearing screening, follow-up, or diagnostic evaluation recorded. Providers who conduct hearing tests can enter follow-up hearing screening results, diagnostic evaluation results, and NYSEIP referrals directly into NYEHDI-IS.

The New York Early Intervention System (NYEIS) is the repository for pertinent data related to the provision of early intervention services including referrals, eligibility, evaluations, individualized family service plan details, services provided, and providers.

Dataset Criteria

SAS 9.4 software was used to prepare the datasets for the data match. The criteria for compiling the NYEHDI-IS dataset included all infants born in calendar years 2014–2016 who had a hearing loss documented by an audiologist. The hearing loss diagnosis included bilateral, unilateral, conductive, sensorineural, and auditory neuropathy. For calendar years 2014 and 2015, the datasets included infants who had transient conductive hearing loss. Based on the CDC's Hearing Screening and Follow-up Survey hearing loss definition, transient conductive hearing loss was excluded for the dataset compiled for the 2016 data match.

The criteria for compiling the NYEIS dataset included infants who were born in the calendar year who were referred and enrolled in the NYSEIP with a diagnosis of hearing loss as specified in Table 1. In New York, a diagnosis of transient hearing loss does not establish eligibility for the NYSEIP, so this diagnosis was not included in the criterion for selection of records from NYEIS.

Table 1
International Classification of Diseases (ICD) Codes Included in the NYEIS Dataset

ICD9 Diagnosis	ICD10 Diagnosis
315.34 Speech and language developmental delay due to hearing loss	F80.4 Speech and language development delay due to hearing loss
389.00 Conductive Hearing Loss	H90.2 Conductive hearing loss, unspecified
389.01 Conductive hearing loss, external ear	
389.02 Conductive hearing loss, tympanic membrane	
389.03 Conductive hearing loss, middle ear	
389.08 Conductive hearing loss of combined types	
	H90.42 Sensorineural hearing loss, unilateral, left ear, with unrestricted hearing on the contralateral side
389.14 Central hearing loss	H90.5 Unspecified sensorineural hearing loss
389.15 Sensorineural hearing loss, unilateral	H90.41 Sensorineural hearing loss, unilateral, right ear, with unrestricted hearing on the contralateral side
	H90.42 Sensorineural hearing loss, unilateral, left ear, with unrestricted hearing on the contralateral side
389.18 Sensorineural hearing loss, bilateral	H90.3 Sensorineural hearing loss, bilateral
389.20 Mixed Conductive and Sensorineural Hearing Loss	H90.8 Mixed conductive and sensorineural hearing loss, unspecified
389.21 Mixed hearing loss, unilateral	H90.71 Mixed conductive and sensorineural hearing loss, unilateral, right ear, with unrestricted hearing on the contralateral side
	H90.72 Mixed conductive and sensorineural hearing loss, unilateral, left ear, with unrestricted hearing on the contralateral side
389.22 Mixed hearing loss, bilateral	H90.6 Mixed conductive and sensorineural hearing loss, bilateral
744.00 Unspecified anomalies of the ear with hearing impairment	Q16.9 Congenital malformation of ear causing impairment of hearing, unspecified
744.02 Congenital absence, atresia, and stricture of auditory canal (external)	Q16.1 Congenital absence, atresia, and stricture of auditory canal (external)
744.09 Other anomalies of ear causing impairment of hearing	Q16.9 Congenital malformation of ear causing impairment of hearing, unspecified

Note. NYEIS = New York Early Intervention System

NYEIS data were matched to New York Early Hearing Detection and Intervention Information System (NYEHDI-IS) data to identify infants documented in NYEIS who were not documented with a hearing loss diagnosis in NYEHDI-IS. In addition, NYEHDI-IS data were matched to NYEIS data to identify infants with hearing loss in NYEHDI-IS who were not documented in NYEIS.

The Link King

Matching was completed using The Link King, a free record linkage software that uses a combination of probabilistic and deterministic algorithms to link and de-duplicate records (Campbell, 2005). The Link King program allows the user to select one of three blocking levels (low, medium, high) and must be specified prior to the blocking of the data. In this analysis, a medium blocking level was used and the first and last name, date of birth, and gender variables were used to block and match the datasets.

Paired records were included in the blocked dataset if any one of the following criteria matched in a pair: date of birth and last name; date of birth, gender, and first name; gender and first and last name; first and last names and either birth month and year, birth month and day, or birth day and year match (The Link King, 2004).

Manual Review

A paired record was considered a match if it had a perfect match on all four variables and no further review was conducted. All non-matched pairs were manually reviewed and researched in both the NYEHDI-IS and NYEIS systems. Unmatched records were considered matched pairs if they had an obvious typographical error or misspelling in one of the matching variables, the first and last name or first and middle name were found to be transposed, the last name changed or was missing a hyphen, and both records had the same residence address and maternal information.

Results

Record linkage results are shown in Table 2. For the NYEIS to NYEHDI-IS match, Link King identified 84 direct matched pairs and 313 non-matched records for 2014, 94 direct matched pairs and 167 non-matched records for 2015, and 66 direct matched pairs and 94 non-matched records for 2016 in the NYEIS to NYEHDI-IS data match process. In the NYEHDI-IS to NYEIS data match process, the Link King identified 84 direct matched pairs and 130 non-matched records for 2014, 94 direct matched pairs and 122 non-matched records for 2015, and 66 direct matched pairs and 79 non-matched records for 2016 (Table 2).

Table 2

New York Early Intervention and Early Hearing Detection and Intervention Data Match Results, 2014-2016

Data Match		2014	2015	2016
NYEIS to NYEHDI-IS	**NYEIS sample size**	397	261	160
	Direct matched pairs from Link King	84	94	66
	Results needing further review	313	167	94
	No permanent hearing loss diagnosis	270	150	84
	Late onset*	155	31	24
	Lost to follow-up†	87	35	49
	No results recorded‡	27	72	4
	Transient conductive	1	12	7
	Infant born out of state or country	30	11	4
	Parent refusal of screening, follow-up, or diagnostic testing	3	0	0
	Changes in infant DOB or name	6	2	3
	Other§	4	0	1
	True matched records missed	0	4	2
NYEHDI-IS to NYEIS	**NYEHDI sample size**	214	216	145
	Direct matched pairs from Link King	84	94	66
	Results needing further review	130	122	79
	No hearing loss diagnosis documented in NYEIS	93	93	45
	Infant moved out of state or country	0	0	2
	Parent refused referral to Early Intervention	8	9	6
	Changes in infant DOB or name	5	9	4
	Record not found in Early Intervention	13	6	21
	Not referred to Early Intervention	6	2	13
	Provider did not complete referral process	7	4	8
	True matched records missed	11	5	1

Note. DOB = date of birth; NYEHDI-IS = New York Early Hearing Detection and Intervention Information System; NYEIS = New York Early Intervention System.
* Infant passed their initial, follow-up, or diagnostic evaluation and were later found to have permanent hearing loss.
† Infant failed their initial or follow-up screening and never received a diagnostic evaluation.
‡ Infant did not have any results recorded.
§ Infant cannot be found in NYEHDI-IS.

Based on manual reviews, reasons for records from NYEIS not matching records from NYEHDI-IS include the infant not having a hearing loss diagnosis documented in NYEHDI-IS; the infant was born out of state or country; parent refusal of screening, diagnostic evaluation, or referral to NYSEIP; or there were major changes to any one of the matching variables. Those changes included data entry corrections made to the birthdate or gender, name changes such as "Twin A" or "Baby Boy X" to a permanent name, or last name changes from a maternal last name to a paternal last name that were not reflected in the other system. The "Other" category contains infants that were not found in NYEHDI-IS and there is insufficient information in NYEIS to determine a specific reason. This resulted in a total of six true matched pairs missed (99% matching success rate) for 2014–2016. Complete results for the 2014–2016 NYEIS to NYEHDI-IS data match can be seen in Table 2 and Table 3.

Table 3
Degree of Accuracy for Link King Data Match between NYEIS and NYEHDI-IS Datasets, 2014-2016*

Data Match	2014	2015	2016
NYEIS to NYEHDI-IS	100%	98%	99%
NYEHDI-IS to NYEIS	95%	98%	99%

Note. NYEHDI-IS = New York Early Hearing Detection and Intervention Information System; NYEIS = New York Early Intervention System.

* $\frac{\text{true matched records}}{\text{Dataset sample size}}$

Non-matched records from NYEHDI-IS to NYEIS datasets were also manually reviewed. Reasons for records from NYEHDI-IS not matching records from NYEIS include infant not having a hearing loss diagnosis documented in NYEIS, infant moved out of state or the country, the parent refused referral to NYSEIP, changes were made to an infant's date of birth or name, or there was no record of the infant in NYEIS. This resulted in a total of 17 true matched pairs missed (97% matching success rate) for 2014–2016. Complete results for the 2014–2016 NYEHDI-IS to NYEIS data match can be seen in Table 2 and Table 3.

As a result of the match, there were also 504 infants identified who had no permanent hearing loss recorded in NYEHDI-IS, but had a permanent hearing loss diagnosis recorded in NYEIS by an NYSEIP evaluator for 2014–2016. Among those infants, there were 210 infants with late-onset hearing loss, which was defined as infants with a diagnosis of hearing loss in NYSEIP as determined by the evaluator but who had passed their initial or follow-up hearing screening or diagnostic evaluation in NYEHDI-IS.

Discussion

The purpose of this study was to match records from NYEHDI-IS and NYEIS to identify areas in the state where hearing screening, follow-up services, and referrals to the NYSEIP were not documented.

By matching NYEIS data to NYEHDI-IS data, 274 infants were identified as having a diagnosis of permanent hearing loss documented in NYEIS and receiving early intervention services, but not documented as having a diagnostic evaluation in NYEHDI-IS (Table 2). These children received services from 72 audiologists who had not documented diagnostic evaluations in NYEHDI-IS. NYEHDI Program staff provided education on NYS Public Health Law reporting requirements and ensured that the audiologists had access to NYEHDI-IS to report. The average amount of time needed for education and training of these providers ranged from 30 minutes to two hours, depending on provider technical skill and previous experience with data reporting to the NYS Department of Health.

Conversely, by matching NYEHDI-IS data to NYEIS data, 40 infants with a documented diagnosis of hearing loss in NYEHDI-IS did not have a documented referral to NYSEIP. Nineteen of these infant's providers documented the referral in NYEHDI-IS, but did not complete the referral process (Table 2). The infant referral process is not automated in NYS so the provider must contact the municipal Early Intervention Official based on the infant's county of residence (NYSDOH, 2017). These infants were served by 22 audiologists. The NYEHDI program provided information about the benefits of referring children for early intervention services, as well as training on the referral process to the audiologists to ensure infants diagnosed with hearing loss were referred to NYSEIP in a timely manner. The time spent providing education and training to these providers ranged from 10 to 30 minutes depending on their knowledge of NYSEIP and NYEHDI-IS.

Strengths

The results of this study indicate that the Link King can successfully match newborn hearing screening and early intervention records with a high degree of accuracy (Table 3). The initial number of non-matched records in the NYEIS to NYEHDI-IS data match was reduced from 574 total non-matched records for 2014–2016 to six missed matched records after manual review. The initial number of non-matched records for the 2014–2016 NYEHDI-IS to NYEIS data match was also reduced from 331 total non-matched records to 17 missed matched pairs. Upon manual review of the initial non-matched records, it was found that the high number of non-matched records in 2014 was due to the broad criteria used to prepare the datasets which included infants referred to the Early Intervention Child

Find Program as a result of a missed initial hearing screening. The criteria were revised for subsequent years' data, which resulted in better matching.

The Link King software was user friendly and the program's website contains informative walk-through videos, a user manual, and extensive online help that makes it easy for the user to familiarize themselves very well with the program (The Link King, 2016). The Link King program is free to download, but does require a SAS license to run.

The efforts implemented as a result of this analysis have also improved the timeliness, accuracy, and completeness of data reporting to the CDC in the annual Hearing Screening and Follow-Up Survey.

Limitations

More questions were raised than answered by these analyses. There was a noticeable increase between 2014 and 2015 in the number of infants with no results recorded. It is not evident to the NYEHDI program as to why this may have happened.

Both birth record data and data entered into NYEHDI-IS by providers were used for the data match process. For infants with late-onset hearing loss, hearing screening results submitted on the birth certificate were used. However, the hearing screening results obtained from the birth certificates were not verified individually at the hospital. The matching process relies upon accurate data reporting and although a review of original hospital and NYSEIP records would provide quality assurance, this was not feasible for the current study.

Similarly, the data match relied on complete and accurate information entered into NYEIS. Infants may be determined eligible for NYSEIP for reasons other than hearing loss and depending on local data entry practices, hearing loss diagnosis codes may not have been recorded.

The time needed to research the unmatched infant records depended on the completeness of documentation and accessibility of the diagnostic hearing result in both information systems, the service provider, and the need for higher level staff to assist in the research, which varied from five minutes to two hours. Two part-time temporary staff were hired by the NYEHDI Program to assist with the research of unmatched infants' records. The process of reviewing unmatched records from three birth cohort years took staff approximately 250 hours to complete.

However, documentation of the data match procedures and subsequently conducting the data match on a monthly basis helped streamline the review process. The amount of time spent reviewing unmatched infant records decreased to approximately two hours per month.

Identifying, contacting, and educating providers so they comply with NYS Public Health Law requires staff time. Working with providers and ensuring they are following reporting requirements is an ongoing process. Approximately eight hours were spent providing education and training about the importance of referral to NYSEIP and explaining the process for referring infants after confirmed hearing loss diagnosis. Approximately 90 hours were spent educating providers about the need to report diagnostic hearing results in NYEHDI-IS and providing access to and training of NYEHDI-IS.

The data match is not a one-time analysis to validate the data. The match must be repeated on an ongoing basis. The initial set-up of the data match required development of SAS code to extract data from both information systems. Furthermore, setting up a blocking algorithm in the Link King software takes technical skill and time. Once set up is complete, the SAS code and matching algorithms can be used for future matches and the amount of time staff spend reviewing unmatched infant records will decrease significantly.

Conclusion

Data matching is an effective and evidence-based method to identify gaps in data and in states' systems of screening, diagnosing, and referring infants for Early Intervention Program services. The NYEHDI Program has integrated the process of linking data from the two data systems into routine monthly processes. By conducting a monthly data match, the NYEHDI program can continue to identify gaps in reporting and target technical assistance to areas in need of improvement.

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Effectiveness of Dexmedetomidine for Sedation in Auditory Brainstem Response Testing

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Abstract: Sedation is widely used for auditory brainstem response (ABR) testing for infants or young children who are unable to sleep or remain adequately quiet for testing. Because chloral hydrate is no longer readily available, dexmedetomidine has been proposed as an alternative medicinal agent to achieve moderate levels of sedation without risk of respiratory depression. The purpose of the study was to assess the effectiveness of dexmedetomidine in terms of the completeness of the audiologic data obtained in achieving moderate levels of sedation for auditory brainstem response testing. A retrospective chart review was conducted on 99 patients at Kennedy Krieger Institute. Participants were administered either chloral hydrate or dexmedetomidine prior to ABR testing. Effectiveness was defined as having obtained thresholds for click and tone burst stimuli centered at 500, 2000, and 4000 Hz for both ears. Complete audiological data were obtained on 92.2% of patients sedated with dexmedetomidine whereas complete audiological data were obtained on 91% of patients sedated with choral hydrate in a period prior to the use of dexmedetomidine. It was concluded that dexmedetomidine is as effective as chloral hydrate in producing an appropriate state for sedated auditory brainstem response testing.

Key Words: dexmedetomidine, sedation, auditory brainstem response

Acronyms: ABR = Auditory Brainstem Response; IHS = Intelligent Hearing Systems; KKI = Kennedy Krieger Institute; MRI = Magnetic Resonance Imaging

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Introduction

The auditory brainstem response (ABR) is used to delineate the auditory status of infants and children who are unwilling or unable to cooperate for developmentally appropriate behavioral test procedures or to confirm behavioral test findings obtained when a hearing loss is suspected. ABRs are bioelectric signals generated by the auditory nerve and units in the brainstem in response to auditory stimuli. The ABR is an onset response and requires neural synchrony in order to effectively record a response from electrodes which are placed on the scalp. ABR testing is a non-invasive procedure but requires the patient to remain quiet and relaxed for approximately 45 to 60 minutes. Sedation is widely used to complete ABR testing for children who are not able to sleep naturally or remain sufficiently quiet for the duration of testing (American Academy of Audiology, 2012). Infants and older children with developmental disabilities, behavior disorder, autism, or intellectual disabilities often require sedation for successful completion of ABR testing. Using general anesthesia will achieve an adequate state for testing; however, the testing facility must arrange for anesthesiology, which may introduce delays in early identification of hearing loss. Consequently, use of sedation or general anesthesia could compromise adherence with Joint Committee on Infant Hearing Guidelines for early detection and management of hearing loss (Joint Committee on Infant Hearing, 2007). General anesthesia also introduces health risks such as respiratory and cardiac complications (Jenkins & Baker, 2003).

There are alternatives to general anesthesia for obtaining threshold-finding ABR results in infants and young children. For example, several studies have reported on the efficacy of melatonin as an alternative to sedation

for ABR testing (Guerlain et al., 2016; Marseglia et al., 2015; Schmidt, Knief, Deuster, Matulat, & am Zehnhoff-Dinnesen, 2007) and other procedures in children typically requiring the use of sedation (Marseglia et al., 2015). The efficacy of melatonin was assessed in a large sample of 250 children assessed with both click and tone burst stimuli in notched noise (Schmidt et al., 2007). Although the use of melatonin significantly reduced the need for general anesthesia by more than 80%, thresholds for a click, and three or more tone burst stimuli were only obtained in 57% of children assessed with melatonin-aided sleep (Schmidt et al., 2007). However, success rates were markedly better for children less than one year of age than for older children (Schmidt et al., 2007). More recently, use of melatonin was assessed in 56 children from 1 year to 14.5 years who were administered ABR testing with a protocol that included only thresholds for click stimuli (Guerlain et al., 2016). The click-evoked ABR was completed among 43 patients. In the patients successfully tested, the mean delay to achieving sleep was 35 minutes and the mean duration of sleep only 23 minutes (Guerlain et al., 2016).

Another alternative to sedation use for ABR testing involves using sophisticated response collection algorithms which are more robust in the presence of patient-generated motion artifact (e.g., Kalman adaptive processing; Cone & Norrix, 2015). Kalman adaptive averaging has been used to improve the signal to noise ratio in noisy recordings and has been shown to reduce the averaging time needed for resolution of ABR responses by 75% as compared to more traditional averaging techniques (Chan, Lam, Poon, & Qiu, 1995). In the presence of motoric activity, Kalman-weighted averaging coupled with in situ bio-amplifiers results in ABR threshold estimates that are 6 to 7 dB lower than when conventional ABR averaging methods are used (Cone & Norrix, 2015). It was suggested that the methodology may make it possible to test some infants in an awake state without the associated added costs and potential delays inherent with use of sedation or general anesthesia (Cone & Norrix, 2015). The effectiveness the commercial application of Kalman-weighted averaging coupled with in situ bio-amplifiers was undertaken in 103 children who were administered click and tone burst evoked ABR testing without use of sedation (Hall, 2010). It was not possible to record any interpretable ABR results on 6% of the children (Hall, 2010). Click evoked ABR thresholds were obtained on 94% of children. However, the instrumentation was less effective in obtaining thresholds for 500 Hz (24% of Ss), 1000 Hz (36% of Ss), 2000 Hz (40% of Ss), and 4000 Hz (40% of Ss) for the tone burst stimuli (Hall, 2010).

Use of conscious or moderate sedation is also an alternative to general anesthesia for achieving an adequate level of cooperation for ABR testing (Reynolds, Rogers, Medellin, Guzman, & Watcha, 2016). Dexmedetomidine has been used in recent years to achieve moderate levels of sedation for ABR and other procedures (Ambi, Joshi, Ganeshnavar, & Adarsh, 2012; Reynolds, Rogers, Medellin et al., 2016). Dexmedetomidine achieves rapid onset of sedation effects with a relatively short half-life which lends itself to non-invasive outpatient procedures such as magnetic resonance imaging (MRI) and ABR studies (Phan & Nahata, 2008). In addition, intranasal dexmedetomidine has some potentially beneficial analgesic and anxiolytic side effects which might be significant for some patients (Phan & Nahata, 2008). Use of dexmedetomidine was approved by the Food and Drug Administration for use in non-intubated adult patients in 2008 and has been used in pediatric applications for procedures which are minimally invasive off-label since that time (Shukry & Miller, 2010). It is not a controlled substance (Drug Enforcement Administration, 2017). Dexmedetomidine is an anxiolytic and sedative medication which is used in the intensive care setting for light to moderate sedation (Phan & Nahata, 2008). It is an agonist of alpha2-adrenergic receptors in certain parts of the brain and is similar to the medication clonidine, which is often prescribed in children with attention deficit hyperactivity disorder (Sallee, Connor, & Newcorn, 2013). It can provide sedative effects without risk of respiratory depression, unlike other commonly used sedatives such as propofol, fentanyl, and midazolam (Phan & Nahata, 2008). Dexmedetomidine is absorbed through the olfactory mucosa, allowing for intranasal administration (Iirola et al., 2011). Dexmedetomidine does have potential adverse side effects which include lowered blood pressure and decreased oxygen in tissues or blood (Cravero, Anderson, & Wolf, 2015). Although use of dexmedetomidine has been shown to reduce heart rate and blood pressure, vital signs remain within safe physiologic limits and no serious adverse effects have been reported (Surendar, Pandey, Saksena, Kumar, & Chandra, 2014). Intranasal dexmedetomidine produces considerably less impact upon respiration than alternative forms of sedation and is regarded as safer than chloral hydrate or other alternative drugs used to achieve conscious sedation (Cozzi, Norbedo, & Barbi, 2017). Intranasal dexmedetomidine has been used to achieve moderate levels of sedation for MRI studies (Ambi, Joshi, Ganeshnavar, & Adarsh, 2012; Zhang et al., 2016) and for electroencephalogram studies (Baier, Mendez, Kimm, Velazquez, & Schroeder, 2016) as well as auditory brainstem response measurements in children (Baier et al., 2016; Reynolds, Rogers, Capehart, Manyang, & Watcha, 2016; Reynolds, Rogers, Medellin et al., 2016).

Moderate sedation is an alternative to general anesthesia which requires less intensive medical supervision and can be undertaken outside of an operating room environment. Chloral hydrate was widely used to achieve moderate sedation for ABR studies but is no longer used for several reasons. Respiratory depression, vomiting, and paradoxical hyperactivity are potential side effects of chloral hydrate sedation (Greenberg, Faerber, & Aspinall, 1991). Complications such as vomiting, hyperactivity, or rash have been reported in 20.7% of children tested with chloral hydrate (Avlonitou et al., 2011). Additionally, chloral hydrate is often administered orally, which requires

cooperation of the patient to swallow the medication in order to ensure accurate dosing. There is also a heightened risk of respiratory compromise with the possibility of neurologic injury in patients with certain conditions (e.g., cerebral palsy, obstructive sleep apnea, hypertrophic tonsils and adenoids, and tracheostomy; Phan & Nahata, 2008). Finally, chloral hydrate is difficult to obtain, as the sole remaining pharmaceutical manufacturer in the United States has ceased production of the medication (Mason, 2014). Choral hydrate is still in use in other countries (Valenzuela et al., 2016). However, it has not been recommended for use in pediatric sedation for a number of years (Cote, Karl, Notterman, Weinberg, & McCloskey, 2000). Use of chloral hydrate is now banned in France and Italy due to evidence of carcinogenicity and genotoxicity (Cozzi et al., 2017).

In anticipation that chloral hydrate would no longer be available, a team with representatives from pediatrics, pediatric anesthesiology, nursing, and pharmacy considered available medications for moderate sedation and selected intranasal dexmedetomidine as the most suitable option for the Kennedy Krieger Institute (KKI) because of its safety profile, lack of respiratory depression effects, and ease of administration. Intranasal dexmedetomidine has been shown to achieve moderate levels of sedation in significantly less time than chloral hydrate with no occurrences of hypoxemia (Reynolds, Rogers, Medellin et al., 2016; Zhang et al., 2016). First dose success rates for intranasal dexmedetomidine have been shown to be higher than those achieved previously in patients sedated with chloral hydrate (Baier et al., 2016). The injectable formulation of dexmedetomidine is administered into the nose using an atomizer. This method of administration has been used safely in previous investigations evaluating the sedative effects of dexmedetomidine in children (Baier et al., 2016; Cravero et al., 2015; Reynolds, Rogers, Medellin et al., 2016). Onset of sedation is typically about 20 to 30 minutes and lasts 60 to 90 minutes (Reynolds, Rogers, Medellin et al., 2016).

The purpose of the present retrospective study was to compare the effectiveness of intranasal dexmedetomidine with chloral hydrate in achieving an adequate state for the auditory brainstem response testing protocol used at Kennedy Krieger Institute. Consecutive medical records were examined with no attempt made to control for subject variables. This protocol requires obtaining responses to click stimuli and tone bursts centered at 500, 2000, and 4000 Hz for both ears. The tone bursts were presented in the background of notched noise. Tympanometry and measurements of otoacoustic emissions are also performed. Few studies have investigated the effectiveness of intranasal dexmedetomidine for sedation in ABR studies (Reynolds, Rogers, Medellin et al., 2016). No studies have quantified the effectiveness of dexmedetomidine for a range of stimulus conditions, including both click stimuli and frequency specific tone burst stimuli. Previous studies investigating the efficiency of intranasal dexmedetomidine in sedation for ABR studies have included only a single stimulus condition (Baier et al., 2016; Reynolds, Rogers, Capehart et al., 2016; Reynolds, Rogers, Medellin et al., 2016). Such data do not permit the degree, configuration, and etiology of a hearing loss to be determined. It is important that effectiveness of intranasal dexmedetomidine be evaluated for a range of stimulus conditions because the completeness of this data informs treatment decisions and the need for further evaluation and/or intervention.

Procedures

This investigation was approved by the Johns Hopkins Medicine Institutional Review Board. A retrospective chart review was conducted for patients at Kennedy Krieger Institute who underwent sedated ABR testing. Data were available for 64 patients sedated with intranasal dexmedetomidine and 35 patients sedated with chloral hydrate. The subjects in the dexmedetomidine group ranged from 6 months of age to 10 years and 7 months of age (mean age = 3.30 years). The subjects in the chloral hydrate group ranged in age from 7 months to 5 years and 11 months (mean age = 2.80 years). Sedated ABR testing is typically not performed on children younger than six months because these children are usually able to sleep without sedation for the duration of testing. The subjects typically had multiple diagnoses including speech and language delay, history of otitis media, behavior disorders, autism spectrum disorder, ADHD, global developmental delay, and hypotonia. All subjects were medically evaluated by a Kennedy Krieger Institute developmental pediatrician as well as by their own pediatrician to ensure candidacy for sedation. A sedation referral form, shown in Table 1, was completed by each child's pediatrician prior to scheduling an appointment for a sedated ABR. Because dexmedetomidine lowers heart rate and blood pressure, it is contraindicated for use in children with known bradycardia, hypotension, or other cardiac problems. When children were screened for sedation, it was decided that dexmedetomidine would not be used in children who were taking other medications that lower heart rate or blood pressure. Dosing and response to dexmedetomidine may be affected if children are prescribed other alpha-2 agonists. Guanfacine and clonidine are the two alpha-2 agonists that are commonly prescribed. Guanfacine, also known as Intuniv, is approved for treatment of attention deficit hyperactivity disorder (ADHD). Clonidine can be used for treatment of ADHD and high blood pressure. Tizanidine is also a related medication. The protocol at KKI specifies that patients discontinue these medications for 2 weeks prior to sedation, as dexmedetomidine will be more effective if patients do not have tolerance to that class of medications.

Table 1
Pre-Sedation Referral Form

Name: _____ DOB: _____

KKI#: _____ AEP DATE: _____

The above named patient has been referred for sedated AEP testing. Intranasal Dexmedetomidine will be administered.

Medical Problem List	Medications	Allergies (or sensitivities, especially to meds)	Past Surgeries

Any prior complications of anesthesia or sedation? [ ] yes [ ] no

Pertinent Past Medical History (elaborate on any Yes responses):

History of:	Yes	No	History of:	Yes	No	History of:	Yes	No
Premature birth			Obstructive sleep apnea			Congenital anomaly		
Apnea			Aspiration			Anemia		
ENT Problems			Asthma			History of transfusion		
Feeding/Swallowing problems			Other pulmonary disease			Other blood disorder		
Gastroesophageal reflux			Congenital heart disease			Seizures/epilepsy		
Frequent vomiting			Need for SBE prophylaxis			Hydrocephalus/Shunt		
Liver disease			Arrhythmia			Cerebral palsy		
Kidney disease			Other heart disease			Other neurologic problem		

Additional Info: _____

_____

Physical Exam: Normal (N) / Abnormal (A) Abnormal (describe)

HEENT _____ Abdomen _____

Lungs _____ Other _____

Cardiovascular _____ Weight: _____ Height: _____

I evaluated the above patient on _____ and find the patient to be in his/her usual state of health and find no contraindications to doing AEP testing under moderate sedation.

Printed Name Provider Signature Date/Time

_____ _____ _____

Note. AEP = Auditory Evoked Potential; ENT = Ear, Nose, and Throat; HEENT = Head, Ear, Eyes, Nose, and Throat; SBE = Subacute Bacterial Endocarditis Prophylaxis

Dexmedetomidine is administered through intranasal administration by contact with the olfactory mucosa (Iirola et al., 2011). This was achieved with use of a tuberculin syringe with an atomizer device. Guidelines promulgated by the Committee on Drugs of the American Academy of Pediatrics pertaining to dietary precautions, as well as monitoring and management of pediatric patients for conscious sedation were followed (American Academy of Pediatrics, 1992). Patients received an examination from a developmental pediatrician and a health history was taken along with a baseline weight and vital signs. Due to the possible effects of decreased blood pressure and pulse, the patients were monitored by a nurse throughout the procedure. Blood oxygen saturation was measured by continuous pulse oximetry. Blood pressure measurements were taken every 15 minutes. The available dosing of dexmedetomidine at KKI was in a concentration of 100mcg/1cc. The dosing was usually split between each nostril to decrease runoff and maximize absorptive tissue area. The ideal volume per nostril is 0.2–0.3cc and maximum recommended dose is 0.5–1cc per nostril (Barclay & Lie, 2010). When administering small doses to younger patients, it was necessary to dilute dexmedetomidine to a concentration of 50mcg/1cc to allow adequate volume for intranasal use. On the day following the appointment, a telephone call was placed to each patient's parent/guardian to inquire about recovery and any adverse effects. Based on continued evaluation of patient response intranasal dexmedetomidine at KKI, the current dosing protocol is to give 2 mcg/kg for the initial dose. Previous research provides evidence that a 2 mcg/kg dose is effective for sedation in pediatric populations (Yuen et al., 2012). If the patient is not sufficiently sedated by 25 minutes after the initial dose, an additional 1 mcg/kg dose of intranasal dexmedetomidine is given. A third dose of 1 mcg/kg is given if the patient remains awake 25 minutes after the second dose. Dosing remains individualized but this protocol has provided consistent results for the majority of our sedated auditory evoked potential and electroencephalogram procedures. The dosing for chloral hydrate was 75 mg/kg. When vomiting occurred with chloral hydrate administration, re-administration of the medication was required. In some such cases, sedation was ineffective.

After ensuring the patient had reached an adequate state for testing, ABR data were collected using an Intelligent Hearing System's (IHS) Smart Evoked Potential System. The skin was prepped using NuPrep solution and standard EEG disk electrodes were affixed to the following locations: Fpz (ground), Fz (non-inverting), A1 and A2 (inverting for ipsilateral and contralateral ear). Click stimuli and tone burst stimuli centered at 500, 2000, and 4000 Hz were presented to each ear via insert earphones. Notched noise was used for the tone burst stimuli. If any degree of hearing loss was present, testing by bone conduction was completed with the delivery of appropriate contralateral masking noise. Tympanometry and measurements of otoacoustic emissions were also performed following ABR testing.

Results

Complete audiological data was defined as having obtained thresholds for click stimuli and for tone burst stimuli centered at 500, 2000, and 4000 Hz for both the left and the right ear. Complete audiological data were obtained on 92.2% of patients sedated with dexmedetomidine. Complete audiological data were obtained on 91% of patients sedated with choral hydrate. A chi-square analysis revealed that there was no significant difference in outcome between the two forms of sedation ($\chi 2(1, N = 99) = 0.0175$, $p = 0.89$). Complete data were obtained for at least one ear on all patients.

Table 2
Participant Demographics

Sedation Type	Effective	Not effective
Dexmedetomidine ($n = 64$)	92% ($n = 59$)	8% ($n = 5$)
Sedation Type: Chloral Hydrate ($n = 35$)	91% ($n = 32$)	9% ($n = 3$)

Most of the children required two (50%) or three doses (20%) of dexmedetomidine. Only 30% of patients sedated with a single dose. The average times before sedation were 25 minutes, 39.4 minutes, and 57.5 minutes for patients requiring 1, 2 or 3 doses of dexmedetomidine respectively.

The mean age for patients sedated with dexmedetomidine who were adequately sedated with only one dose was 3 years. The range of ages of children receiving a single dose was 6 months to 10.5 years. The mean age for those receiving more than 1 dose was 4.4 years, with a range of 9 months to 10.6 years. The mean total examination time across all participants who received dexmedetomidine was 53.1 minutes. No patients in the dexmedetomidine group required supplemental oxygen. All patients returned to baseline functioning and were discharged on the same day as the procedure. No patients receiving dexmedetomidine experienced nausea or vomiting that was either observed or reported after discharge.

Discussion

The results of this investigation indicate that intranasal dexmedetomidine is as effective as chloral hydrate in achieving moderate levels of sedation for ABR testing. No sentinel events occurred due to the use of dexmedetomidine. None of the adverse side effects associated with chloral hydrate including vomiting, hyperactivity, or respiratory depression occurred during

the use of intranasal dexmedetomidine. Intranasal dexmedetomidine was noted by all members of the team to be far easier to administer than oral chloral hydrate. Intranasal administration requires minimal cooperation from the patient. A team member or family member gently tilts the patient's head posteriorly and immobilizes the head while the nasal spray is being administered. The patient remains in this position for approximately 20 seconds and can then resume normal activities. In contrast, administration of chloral hydrate required staff to induce patients to swallow an ill-tasting medication which was for some patients difficult to achieve without uncertain amounts of the medication being spit out or vomited. This study supports previous studies suggesting that intranasal dexmedetomidine is an appropriate alternative to chloral hydrate or general anesthesia for ABR testing for most patients (Baier et al., 2016; Reynolds, Rogers, Medellin et al., 2016). The effectiveness of dexmedetomidine appears to be superior to that of alternatives such as administration of melatonin (Schmidt et al., 2007) or use of Kalman averaging (Hall, 2010) which previous studies have shown to be less effective than results achieved in the present study.

Several limitations of the present investigation were noted. The retrospective design of the investigation did not allow for variables such as patient age, the presence of health conditions, or time of testing to be controlled or systematically examined. Additionally, the participants in this investigation were limited to the patients who chose to seek care at KKI. Use of dexmedetomidine requires medical monitoring and continuous physiological monitoring with the attendant expense just as when using chloral hydrate. Further investigations should be completed to include larger sample sizes and diverse populations. As noted earlier, there are alternatives to moderate sedation or general anesthesia for obtaining threshold-finding ABR results in infants and young children. For example, several studies have reported on the efficacy of melatonin as an alternative to sedation for ABR testing with few adverse side effects (Guerlain et al., 2016; Marseglia et al., 2015; Schmidt et al., 2007). Future research comparing the effectiveness and safety of melatonin to dexmedetomidine for obtaining ABR measurements should be completed to determine if either medication is more effective. Additionally, Kalman adaptive averaging has been used to improve the signal to noise ratio in noisy recordings and has been shown to reduce the averaging time needed for resolution of ABR responses by 75% as compared to more traditional averaging techniques (Chan et al., 1995). Incorporating the use of this technology may improve the effectiveness of sedation medication by shortening the length of test time and allowing a greater amount of patient movement. Future investigations could evaluate the effectiveness of dexmedetomidine and/or other sedation medications while incorporating this signal averaging technology.

Conclusions

Measurement of ABRs is an important tool for the delineation of hearing status, as the ABR allows the auditory status of infants and children to be assessed when other methods are inappropriate or unsuccessful. Although sedation is commonly used for ABR testing, no medication has yet replaced chloral hydrate as the clinical standard since its large-scale production was ceased. The results of this study suggest that intranasal dexmedetomidine is an acceptable form of sedation for ABR testing in pediatric patients. Complete audiological data was achieved in 92% of patients. This was accomplished without any of the patients experiencing breathing difficulty, vomiting, or other sentinel effects during or after the procedure. Dexmedetomidine demonstrated similar rates of effectiveness as chloral hydrate with fewer side effects. This study contributes to the body of literature supporting the use of dexmedetomidine for clinical use in facilities performing sedated ABR measurements.

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Are Audiologists Directly Referring children Who are Deaf or Hard of Hearing to Early Intervention?

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Abstract:
Purpose. This study investigated audiologists' perceived roles and responsibilities when making direct referrals to Early Intervention (EI) upon an infant's initial diagnosis of being deaf or hard of hearing.

Method. A national survey was distributed via email and social media networks to pediatric audiologists. A total of 132 anonymous surveys were completed.

Results. 94% of respondents reported that it is within audiologists' scope of practice to directly refer children who are deaf or hard of hearing to EI, however, only 78% of respondents reported ever making this direct referral. Direct referral methods varied across states. Audiologists identified parent resistance and being unsure of the EI eligibility criteria in their state as potential barriers to making direct referrals. Additional analysis was completed on results obtained from Arizona, Illinois, Massachusetts, Ohio, Pennsylvania, and Tennessee, which highlight the various systems used for direct referrals across states. Suggestions for improving the direct referral system include creating universal guidelines across states and an online referral system.

Conclusion. There is variation in how audiologists refer children who are deaf or hard of hearing to EI. Systematic changes to the direct referral system may improve EI enrollment of children who are deaf or hard of hearing prior to 6 months of age.

Key Words: EHDI, Early Intervention, deaf or hard of hearing, direct referral

Acronyms: ASHA = American Speech-Language-Hearing Association; CDC = Centers for Disease Control and Prevention; DHH = deaf or hard of hearing; EHDI= Early Hearing Detection and Intervention; EI = Early Intervention; JCIH = Joint Committee on Infant Hearing

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Introduction

The first universal newborn hearing screening programs were established in the early 1990s. Prior to this, the average age of identification of hearing loss was between 2 and 3 years of age, when children demonstrated limited receptive and expressive language. The universal newborn screening movement built momentum with evidence that children identified as deaf or hard of hearing (DHH) before age 6 months could match language development of their hearing peers (Yoshinaga-Itano, Sedey, Coulter, & Mehl, 1998). In 2000, early hearing detection and intervention (EDHI) legislation was passed to develop newborn hearing screening follow-up services. Since 2000, the EHDI Act has been reauthorized and expanded to include diagnostic services and to require federal administration to recruit, retain, educate, and train qualified personnel to implement the program (Joint Committee on Infant Hearing [JCIH], 2007).

Currently, the primary goals of the EHDI system focus on timely screening, identification, and intervention. These are often called the "1-3-6 goals" as they promote newborn hearing screening by 1 month of age, diagnosis of hearing loss by 3 months of age, and implementation of appropriate early intervention by 6 months of age. Early intervention (EI) services consist of evaluations and therapies for infants and toddlers with developmental delays and established risk conditions, and provision of support to families during the first three years of their

child's life. Services may include the provision of assistive technology, audiology services, speech and language therapy, special education services by a teacher of the deaf or hard of hearing, counseling for the family, medical assistance, nursing services, nutrition services, occupational therapies, physical therapies, or physiological services. Infants who are diagnosed as DHH, have age-appropriate cognitive abilities, and begin receiving EI services before 6 months of age have significantly better outcomes in language, speech, and social-emotional development (Yoshinaga-Itano, 2003) compared to children who begin receiving therapies after 6 months of age. They also have significantly better scores in receptive IQ and have age-appropriate expressive language quotient (Meinzen-Derr, Wiley, & Choo, 2011) compared to children who begin receiving therapies after 6 months of age.

EI eligibility criteria for children who are deaf and hard of hearing varies by state. In some states, unilateral or mild hearing losses do not qualify. In addition, depending on the state, families may be charged for EI services. However, many state programs provide services to children with any type or degree of permanent hearing loss, including unilateral and mild hearing loss, free of charge.

Most recent national EHDI statistics as reported by the CDC indicate that, although 96.1% of all newborns were screened before 1 month of age for hearing loss in 2014, only 67.9% of newborns identified as DHH received EI before 6 months of age (CDC, 2016). There are multiple barriers that may impede enrollment in EI, such as lack of availability of service providers, geographical location of families to EI centers, family refusal of services, and lack of provider referral. It is the belief of these authors that the pathway between the frontline professional (i.e., diagnosing audiologist) and the EI system is the shortest and least susceptible to loss of referral. Our clinical experience indicated that not all audiologists were making direct referrals to EI. The discrepancy noted between the number of infants diagnosed as DHH during diagnostic follow-up and the number of these same infants enrolled in EI before 6 months of age may also reflect children falling through the cracks or obtaining delayed enrollment because the audiologist did not make a direct referral. The purpose of this study was to survey practicing pediatric audiologists about their current practices and perceptions of direct referrals to EI for children who are DHH.

Methods

Participants

Participants in this national study included pediatric audiologists who performed diagnostic evaluations for children ages 0 to 3. Participants were excluded if they were not performing diagnostic evaluations and if their caseload included less than 25% of diagnostic testing for children ages 0 to 3. A total of 132 respondents from 29 U.S. states successfully completed the survey.

The Survey

The instrument used in this study was developed by the audiology externs at Boston Children's Hospital with input from the Director of Audiology and five audiology site managers. The survey included 19 questions and was designed to be completed in less than ten minutes. The questions were related to audiologists' perceived roles and responsibilities when directly referring to EI. A direct referral was defined as a direct contact between the audiologist and the EI provider (with parents' consent). This direct contact would include the audiologist beginning the enrollment process for any child diagnosed as deaf or hard of hearing. An indirect referral was defined as instructions, brochures, or a verbal/written recommendation to the parent or physician to initiate enrollment in EI. The survey was comprised of a variety of question types including free response sections where respondents described the protocol for referring to EI in their state and provided suggestions for improvement to the direct referral system. The survey questions are shown in Appendix A.

Procedure

Research Electronic Data Capture (RedCAP), a secure web application for building and managing online surveys and databases (Harris et al., 2009), was used to develop and track the results of the survey. A specific link to the survey was generated through RedCAP and was distributed via email to various pediatric audiology contacts at hospitals and institutions, social media outlets including Facebook audiology groups, and forwarded via email or word of mouth. All responses from participants were voluntary and anonymous. The survey was available for completion from December 2016 to February 2017. The survey met the Boston Children's Hospital Institutional Review Board's guidelines for exemption from the requirements of 45 CFR 46.101(b).

Results

A total of 151 participants began the survey, but only 132 completed the survey because of eligibility criteria or other factors. Completion rate was 86% and only completed surveys were included in the analysis. Responses were collected from pediatric audiologists practicing in 29 different states. Respondents varied in experience, ranging from 10 or more years of experience (46%) to 6 to 10 years of experience (17%) and 0 to 5 years of experience (36%). Respondents worked in a variety of settings including hospital (72%), clinical (29%), academic (12%), private practice (7%), and educational (6%).

Ninety-four percent of respondents stated that they believed audiologists had a role in directly referring children to EI. However, only 78% of those respondents

reported ever making a direct referral to EI upon initial diagnosis of a hearing loss. The audiologist was rated as the most important referral source for EI by 74% of respondents, the otolaryngologist by 31% of respondents, and parents by 12% of respondents.

Direct referrals were reportedly made using fax (48%), phone (34%), email (15%), and other methods (14%). Other methods for making a direct referral included an online referral form available for the audiologist to complete and a direct referral generated through the Electronic Medical Record. Respondents were asked to rate the direct referral process on a 10-point scale (Figure 1). The direct referral process was rated as extremely easy by 36% of respondents and extremely time efficient by 15% of respondents. Mean ratings were 7.99 for ease of direct referral process and 7.02 for time efficiency. Additionally, 74% of respondents reported ever making an indirect referral to EI upon initial diagnosis of a hearing loss. Indirect referrals were reportedly made by providing verbal instructions to the parents (88%), providing the family with the EI brochure (70%), and writing a recommendation for a referral to EI that was included in the report to the physician (73%).

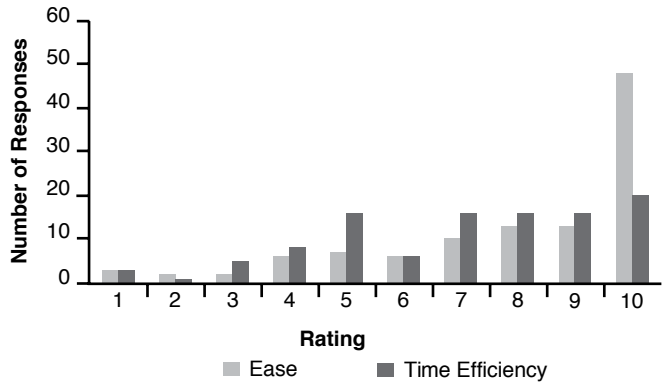

Figure 1. **Ratings of direct referral process for ease and time efficiency. A rating of 1 indicates most difficult/least efficient; a rating of 10 indicates easiest/most efficient.**

Of the 132 respondents, 22% reported not ever making a direct referral to EI upon initial diagnosis of a hearing loss. Barriers to making a direct referral were identified as parent resistance to the referral (33%), audiologist unsure of state EI referral protocol (23%), the direct referral was too time consuming (18%), audiologist unsure of the eligibility criteria for EI in his/her state (8%), direct referrals out of the scope of practice for an audiologist (7%), and other barriers (45%; see Figure 2). Respondents noted other barriers to directly referring to EI including patient already enrolled or referred by another professional and belief that it is the parent's responsibility to initiate EI services. Seventy percent of all responding audiologists who are not directly referring to EI reported relying on the parents to self-enroll in EI.

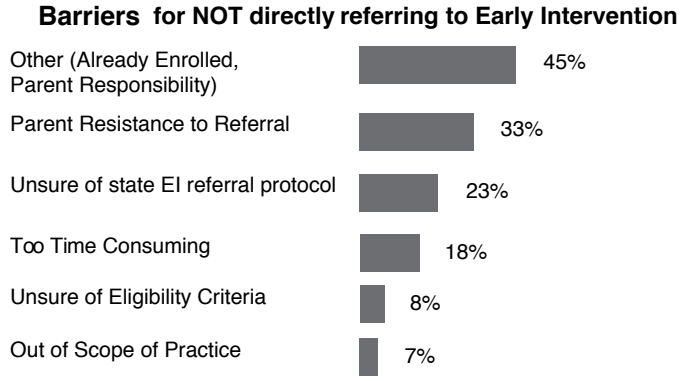

Figure 2. **Barriers to direct referrals to Early Intervention (EI). Audiologists were able to select more than one barrier for this question. Total *n* = 132.**

Additional analyses were performed on the 6 states from where the majority of responses came (64%): Arizona, Illinois, Massachusetts, Ohio, Pennsylvania, and Tennessee. Table 1 highlights the number of pediatric audiologists in the aforementioned states who reported ever making a direct referral to EI upon initial diagnosis of hearing loss. The methods for direct referral varied per state. The methods included a faxed Department of Public Health form to the state Parent-Infant Program (Arizona), a designated Aural Rehabilitation provider who assisted parents to initiate services (Illinois), a regionally based telephone number for the audiologist to call and begin the EI intake process (Massachusetts), Electronic Medical Record referral through the EPIC system and a care coordinator to initiate services (Ohio), a written report and direction to the parent (Pennsylvania), and a faxed form directly to EI (Tennessee).

Table 1
Percentage of Direct Referrals Reported by Respondents in Top Six Responding States

State	Percentage of direct referrals reported
Tennessee	90%
Pennsylvania	88%
Ohio	85%
Arizona	83%
Illinois	62%
Massachusetts	54%

When asked how the respondent decided whether to make a direct or indirect referral for a family, 52 of the 132 audiologists provided a written response. Of those, 11 of the 52 audiologists stated that the decision to make a direct or indirect referral is reliant on the parent's understanding of the hearing loss and perceived capability of the family to initiate services. Ten of the 52 respondents noted that direct referrals are made consistently based on state policy and center protocol.

When asked what methods are used to ensure the child who is DHH is enrolled in EI, 17 audiologists provided a written response. Consistent family follow-up was noted by 6 of the 17 audiologists and communication with the care coordinator or social worker was noted by 5 of the 17 audiologists as methods for ensuring the child who is DHH is enrolled in EI.

In response to a request for suggestions for improving the EI direct referral system, 57 respondents provided an answer. The top suggestions were an online referral system (30%); universal guidelines across states, particularly in locations where providers are working with families from multiple states (12%); and a directory of regionally based EI providers as a reference (12%).

Discussion

In this survey, the majority of respondents (94%) believe that audiologists have a role in directly referring children who are DHH to EI. A direct referral was defined as the audiologist directly contacting the family's local EI program with parental consent. This direct contact would include the audiologist beginning the enrollment process for any child diagnosed as DHH. According to American Speech-Language-Hearing Association (ASHA; 2004), audiologists are responsible for "provision of comprehensive audiologic rehabilitation services, including management procedures for speech and language habilitation and/or rehabilitation for persons with hearing loss or other auditory dysfunction, including but not exclusive to speechreading, auditory training, communication strategies, manual communication, and counseling for psychosocial adjustment for persons with hearing loss or other auditory dysfunction and their families/caregivers." According to the ASHA directive, a direct referral from the audiologist to EI is appropriate and within the scope of practice. However, only 78% of audiologists reported ever making a direct referral to EI upon initial diagnosis of a hearing loss. This reveals a discrepancy between audiologists' perceived roles and responsibilities and current clinical practice.

Audiologists reported certain barriers to making direct referrals to EI. Some audiologists felt that the direct referral process was too time consuming and may be neglected to allow completion of more pressing responsibilities. Audiologists reported parent resistance to the direct referral to EI or that the referral to EI had previously been made by another physician. Lastly, a small number of audiologists do not believe directly referring to EI falls within their scope of practice and should be the responsibility of the child's pediatrician and/or parent. 70% of audiologists who are not making consistent direct referrals reported relying on parents to make the referral. It can be argued that this method empowers the parent in helping them learn to be an advocate for their child. However, this approach may put a child at risk as it creates an additional step in the referral process where enrollment may become delayed, or fail to occur at all. Additionally, at initial diagnosis, parents are often feeling overwhelmed and adding an additional responsibility to the parent to contact EI may cause more burden than empowerment. Considering the national investment in early enrollment into EI, audiologists should consider making the direct referral and find alternative ways for parents to develop their advocacy skills. An audiologist making direct referrals for children who are DHH to EI is particularly advisable for families who have limited income, education, or have a minority status, as this population is more likely to experience delays in enrollment (Bailey, Hebbeler, Scarborough, Spiker, & Mallik, 2004).

By initiating EI services for children who are DHH, the audiologist is acknowledging the benefits EI has on the child's speech and language development and social-emotional outcomes. It can be argued that some families may be resistant to the direct referral to EI if the family and child are DHH and do not wish to pursue amplification for their child. However, the initiation of these services can further support the family in making decisions about the appropriate communication mode for their child. In Massachusetts, there are multiple specialty programs for children who are DHH that the family can choose to access. These specialty programs include oral, total communication, and American Sign Language based programs. Information about these Massachusetts specialty programs are provided in a document titled Specialty Services for Children who are Deaf or Hard of Hearing through the Universal Newborn Hearing Screening Program (2016). Audiologists should be aware of the specialty services available for children who are DHH in their state as these services may vary. By making a direct referral to EI, the audiologist is connecting the family to important resources that can educate them about the options available in their state and can support them in choosing a communication mode for their child.

Overall results indicate that direct referrals to EI programs vary from state to state. With 78% of audiologists reporting experience making direct referrals, it can be concluded that most states already have a means for direct referral by audiologists. The subgroup analysis further supported this notion. Each state reported different rates and methods of direct referrals based on protocol and the structure of their EI system. For example, the states that reported the highest rate of direct referrals were faxing the patient information either directly to EI or to the state EHDI program or making the direct referral to the EI program through the electronic medical record. Eligibility for EI is different across states and 8% of the respondents reported being unsure of the eligibility criteria for the state in which they practice. In Massachusetts, the Early Intervention Operational Standards is a guideline that outlines eligibility criteria for EI and states that permanent hearing loss of any degree deems an infant

eligible for services. This guideline is available through the Massachusetts Department of Public Health (2013) and is accessible on the Massachusetts government website. In light of EI programs varying at the state level, providers should contact their state EHDI or EI program to determine where further information related to EI eligibility for children who are deaf and hard of hearing can be obtained.

In the survey audiologists were asked to provide suggestions for improvement of current direct referral processes. Suggestions included an online referral system, universal guidelines across states, and a directory of regionally based EI providers. An online referral system would reduce paperwork, increase efficiency in making the direct referral, and eliminate referral losses that may take place with faxing, mailing, and other methods currently in practice. This system could be available to the referring audiologist 24 hours per day. This would have an advantage over telephone referral systems as an audiologist could make a referral when their busy schedule permits, without concern of whether an EI employee is available to answer a phone. An online system also has the potential to easily track data for the state on referral trends and support quality assurance initiatives around enrollment in EI. A website hosting the referral system could also provide information related to state laws and guidelines related to EHDI and EI eligibility criteria and references to better connect families with the appropriate EI program or provider. Online referral systems would require capital to create and the direction of state funding toward development of such a system may require advocacy and lobbying.

Although a universal guideline would be ideal, some barriers to such a system would exist. Because EI is funded and operated at the state level, creating a system that could easily be adopted across state lines would require negotiation and buy-in from all states. Collaboration between bordering states, specific to region, could be a more feasible solution. For example, having a collaborative system between nearby states such as Massachusetts, New Hampshire, Connecticut, Maine, New York, and Rhode Island could allow audiologists to provide direct referrals for the majority of patients. This collaborative system could be implemented in states where providers are seeing out-of-state patients on a regular basis.

An additional barrier to an electronic universal system is the potential to violate protected patient health information, either inadvertently or through hacking. The system would need to be a secure site with access only to appropriate and accredited healthcare and EI programs. Such a system would need to be continuously monitored and secured to prevent breaches of private health information.

This study demonstrates that there are systemic changes that could be implemented to support direct referrals for children who are DHH from their diagnosing audiologist to EI. Some barriers reported by respondents in this study could be alleviated through education and technology. An online, universal referral system was the most popular suggestion for improving the direct referral process. Direct referral to EI is within the audiologist's scope of practice. It is the belief of these authors that the most direct route for enrollment into EI for children who are DHH is directly from the diagnosing audiologist to the EI program. Improvements in enrollment processes and audiology education may help states reach their target of enrolling infants with hearing loss in EI by six months of age.

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Appendix A

In this survey, we define direct referral as a direct contact between the audiologist and the early intervention provider (with parents' consent). This direct contact would include the audiologist beginning the enrollment process for any child diagnosed with a hearing loss.

In this survey, we define indirect referral as either instructions, brochures, or a verbal/written recommendation to the parent or physician to initiate enrollment in Early Intervention.

1. When you initially diagnose or confirm a hearing loss are you ever making a direct referral to early intervention?
a. Yes
b. No

2. When you initially diagnose or confirm a hearing loss are you ever making an indirect referral for early intervention services?
a. Yes
b. No

3. Do you feel it's appropriate for audiologists to directly refer to early intervention?
a. Yes
b. No

4. When diagnosing a hearing loss for children ages 0-3, what percentage of the time are you making a direct referral to early intervention?
a. 0% - 100% (Place a mark on the scale above)

5. How are you making the direct referral to early intervention?
a. Phone call
b. Email
c. Fax
d. Other
e. I am not directly referring to early intervention (Check all that apply)

6. Please specify other way(s) you are making direct referrals.
a. (Write In Option)

7. How are you making the indirect referral to early intervention?
a. Verbal instructions to parents
b. Early Intervention brochure
c. Other written material provided to parents
d. Referral included in report of physician
e. Other (Check all that apply)

8. Please specify other way(s) you are making indirect referrals.
a. (Write In Option)

9. On a scale of 1-10, rate the amount of difficulty for making a direct referral to early intervention in your state.
a. 1 (extremely difficult) - 10 (extremely easy)

10. On a scale of 1-10, rate the amount of difficulty for making a direct referral to early intervention in your state.
a. 1 (extremely difficult) - 10 (extremely easy)

11. In your opinion, who should be the primary person to make a referral to early intervention upon initial diagnosis of hearing loss in children? Please rank the following from most appropriate to least appropriate. 1 = most important 5 = least important
a. Audiologist 1-5
b. Otolaryngologist 1-5
c. Primary Care Physician 1-5
d. Parent/Guardian 1-5
e. Speech Language Pathologist 1-5

12. If you are not making direct referrals to early intervention 100% of the time, what are some reasons/barriers?
a. Unsure of eligibility criteria
b. Unsure of the early intervention system protocol for referrals in my state
c. Directly referring to early intervention is out of my scope of practice
d. Too time consuming
e. Parent resistance to the referral
f. Other (Check all that apply)

13. Please indicate other reasons/barriers for not making direct referrals to early intervention.
a. (Write In Option)

14. If you are not making direct referrals to early intervention 100% of the time, how are you ensuring the child receives early intervention?
a. Rely on parents to call/email the early intervention program in their area
b. Rely on Primary Care Physician to make referral
c. Rely on Otolaryngologist to make referral
d. Rely on Department of Public Health to make referral
e. Other (Check all that apply)

15. Please indicate other ways in which you are ensuring children with hearing loss are getting referred to early intervention.
a. (Write In Option)

16. How do you decide whether to make a direct or indirect referral for families?
a. (Write in Option)

17. Please share any suggestions you have for improving audiologists direct referral to early intervention.
a. (Write In Option)

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Scheduling Hearing Appointments Prior to Hospital Discharge Improves Follow-up After Failed Newborn Hearing Screening

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Abstract: The study aimed to identify if there was a relationship between a follow-up hearing appointment scheduled prior to hospital discharge (hospital scheduled appointment) and follow-up status, including loss to follow-up or loss to documentation (LTF/LTD); early follow-up initiation; and early completion of audiological diagnosis. The study included 4,597 children who were born between January 2015 and June 2016 in Louisiana birthing hospitals and failed newborn hearing screening (NHS) prior to hospital discharge. Of the study population, 56.1% of children were scheduled for a follow-up hearing appointment prior to hospital discharge. The LTF/LTD among children without a hospital scheduled appointment was 52% higher than children with a hospital scheduled appointment. The rate of early follow-up initiation with a hospital scheduled appointment was 25% higher than if an appointment was not scheduled while in the hospital. There was no statistical association of early completion of audiological diagnosis with a hospital scheduled appointment. Thus, a hospital scheduled appointment improved LTF/LTD and early follow-up initiation among newborns who failed NHS.

Key Words: Newborn hearing screening, loss to follow-up, hearing appointment, hospital discharge, hospital scheduled appointment

Acronyms: CDC = Centers for Disease Control and Prevention; IS = Information System; LA EHDI = Louisiana Early Hearing Detection and Intervention program; LTD = lost to documentation; LTF = lost to follow-up; LTS = lost to system; NHS = newborn hearing screening; NICHQ = National Initiative for Children's Healthcare Quality; PCP = primary care physician; UNHS = Universal Newborn Hearing Screening

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Introduction

Universal newborn hearing screening (UNHS) has been implemented in all states and some U.S. territories. All newborns with positive screening are advised to get further testing to confirm hearing status, preferably by three months of age (Joint Committee on Infant Hearing, 2007). In fact, follow-up status of many newborns who failed hearing screening is still unknown. According to a 2016 Centers for Disease Control and Prevention (CDC) report using 2014 U.S. data, 42.4% of infants who did not pass the newborn hearing screening (NHS) had no documented diagnosis and 34.4% were lost to follow-up (LTF) or lost to documentation (LTD) for diagnosis. Previous studies have indicated risk factors of LTF/LTD such as government funded health insurance (Liu, Farrell, MacNeil, Stone, & Barfield, 2008); inadequate service-system capacity; lack of provider's knowledge (Moeller, Eiten, White, & Shisler, 2006; Shulman et al., 2010); lack of medical home-state NHS program communication (Kim, Lloyd-Puryear, &

[1]Per CDC data (CDC, 2016); parents of LFU patients are either unresponsive or cannot be contacted.

Tonniges, 2003); late NHS; late follow-up; and multiple follow-ups (Tran, Ng, et al., 2016); long commuter distance (Ravi et al., 2016); mother's lack of knowledge about retesting (Alvarenga, Gadret, Araujo, & Bevilacqua, 2012; Luz, Ribas, Kozlowski, Willig, & Berberian, 2016); rural physicians' limited training in hearing detection; and lack of confidence to direct further care (Bush, Alexander, Noblitt, Lester, & Shinn, 2015). Studies have also shown improvement in follow-up with education of parents before discharge (Spivak & Sokol, 2005; Cockfield, Garner, & Borders, 2012); use of multidisciplinary teams focusing on public awareness (Ravi et al., 2016); collaboration with the Woman, Infants, and Children (WIC) Program (Hunter et al., 2016); partnerships with pediatric otolaryngologists and audiologists (Danhauer et al., 2006); scripting messages given to remind families of follow-up appointments (Russ, Hanna, DesGeorges, & Forsman, 2010); and use of repeated hearing screens to reduce false-positive rates (Shoup, Owens, Jackson, & Laptook, 2005).

Louisiana Early Hearing Detection and Intervention program (LA EHDI) implemented UNHS in 2002. Newborns who fail initial hearing screening prior to hospital discharge are advised to have a follow-up hearing appointment for further testing within one month of discharge. A follow-up hearing appointment can be scheduled either by hospital staff prior to hospital discharge, which is recommended by LA EHDI, or by hospital staff, primary care physician (PCP), or parents after discharge. In Louisiana, scheduling an appointment prior to hospital discharge varies among birthing hospitals and its effect on follow-up status is still unknown. To our knowledge very limited studies evaluating the effect have been published. The objective of this study was to identify if there was any relationship between a follow-up hearing appointment scheduled prior to hospital discharge and follow-up status including (a) loss to follow-up or loss to documentation (LTF/LTD), (b) early follow-up initiation, and (c) early completion of audiological diagnosis. The study findings may help EHDI programs enhance follow-up compliance of newborns who fail newborn hearing screening by developing strategies to improve the rate of scheduling follow-up hearing appointments prior to hospital discharge.

Methods

Data Source and Study Population

The data from LA EHDI-Information System (IS) were used for analysis. The LA EHDI-IS is a web-based database that collects and manages LA EHDI surveillance data including NHS, hearing follow-up/diagnosis, and early intervention. The database system is integrated with birth and death certificates provided by Louisiana Center for Health Statistics and Vital Records. The study included 4,597 children who were born between January 2015 and June 2016 in Louisiana birthing hospitals and had hearing screening prior to hospital discharge but did not pass. Babies born to non-Louisiana residents at birth or those who died at any time after birth were excluded from the study.

Analysis Variables

Study outcomes. Three study outcomes of follow-up status were analyzed: LTF/LTD, early follow-up initiation, and early completion of audiological diagnosis.

LTF/LTD. LTF was defined if a child was reported as LTF to LA EHDI by the follow-up provider. LTD was defined if a child did not have any follow-up reported to LA EHDI by a follow-up provider. LTD newborns had no follow-up indicated in the database, while LTF newborns either had no follow-up indicated or had at least one documented follow-up and still needed additional follow-up to determine audiological diagnosis.

Early follow-up initiation. Early follow-up initiation was defined if follow-up was started within 30 days after NHS, that is, the length of time between NHS and first follow-up was < 30 days. If a child had more than one NHS, time of the last screening was used to calculate the length of time between NHS and first follow-up. Late follow-up initiation was determined if follow-up was started more than 30 days after NHS.

Early completion of audiological diagnosis. Early completion of audiological diagnosis was defined if a child had at least one follow-up and audiological diagnosis was completed by three months of age, that is, age at last follow-up was less than or equal to 90 days. Late completion of audiological diagnosis was determined if the last follow-up for audiological diagnosis was completed after three months of age.

Predictor. The predictor was follow-up hearing appointment scheduled prior to hospital discharge, categorized as yes or no. The predictor was yes if both date and audiologist/facility for follow-up hearing appointment were indicated in the NHS Report Form; otherwise, the predictor was categorized as no.

Covariates. Eight covariates derived from birth certificates included race/ethnicity, child sex, geography of residence at birth, maternal age and education, Medicaid paid delivery, birth weight, and number of previous live births. Five covariates derived from NHS and follow-up/diagnosis reports included number of failed screening ears, age at NHS, screening place, length of time between NHS and first follow-up, and total number of follow-ups. All covariates were defined as categorical variables. Table 1 shows distributions of all covariates in detail.

Data analysis

Three analyses were conducted for the three study outcomes separately. All children who failed NHS prior to hospital discharge (4,597) were included in the LTF/LTD analysis; only children who failed NHS and had follow-up (3,480) were included in the early follow-up initiation analysis; and only children who failed NHS and

completed follow-up (3,404) were included in analysis of early completion of audiological diagnosis. Generalized linear models using PROC GENMOD in SAS were used to determine associations between study outcomes and the predictor. In adjusted models, covariates were included to control for confounding effects. Specifically, for models of LTF/LTD and early follow-up initiation, the following covariates were adjusted in multiple regression models: race/ethnicity, sex, geography of residence at birth, maternal age and education, Medicaid paid for delivery, birth weight, number of previous live births, number of failed screening ears, age at NHS, and screening place. Two additional variables (total number of follow-ups and length of time between NHS and first follow-up) were included in adjusted models of early completion of audiological diagnosis. A backward elimination procedure was used in multiple regression models. All final adjusted models included only variables with p value < 0.05 for statistical significance. 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.

Table 1. **Hearing Appointment Scheduled Prior to Hospital Discharge (%) among Newborns Who Failed Newborn Hearing Screening (N = 4,597)**

Demographic, hearing screening, and follow-up characteristics		Number	Percent (CI 95%)
	Total	**2,579**	**56.1 (54.7-57.5)**
Race/Ethnicity	Non-Hispanic White (42%)	1,087	56.8 (54.5-59.0)
	Non-Hispanic Black (45%)	1,187	57.1 (55.0-59.3)
	Non-Hispanic Other (4%)	95	50.8 (43.6-58.0)
	Hispanic (9%)	210	50.4 (45.6-55.2)
Child sex	Female (43%)	1,114	56.3 (54.2-58.5)
	Male (57%)	1,465	55.9 (54.0-57.8)
Medicaid-paid delivery	No (29%)	637	47.7 (45.0-50.4)
	Yes (71%)	1,942	59.5 (57.8-61.2)
Residence at birth	Rural (38%)	1,124	64.6 (62.4-66.9)
	Urban (62%)	1,455	50.9 (49.1-52.7)
Maternal age	< 20 (9%)	243	58.3 (53.5-63.0)
	20-34 (81%)	2,093	56.1 (54.5-57.7)
	35+ (10%)	243	54.2 (49.6-58.9)
Maternal Education	< High school (22%)	554	56.4 (53.3-59.5)
	High school (33%)	885	57.9 (55.4-60.4)
	> High school (45%)	1,140	54.7 (52.5-56.8)
Previous live births	None (39%)	922	52.0 (49.7-54.4)
	One (29%)	779	58.0 (55.3-60.6)
	Two+ (32%)	878	59.3 (56.8-61.8)
Birth weight (grams)	< 1,500 (4%)	62	34.8 (27.8-41.8)
	1,500 - 2,499 (10%)	216	48.3 (43.7-53.0)
	2,500+ (86%)	2,301	57.9 (56.4-59.5)
Age at newborn hearing screening (NHS)	< 30 days old (96%)	2,504	56.9 (55.5-58.4)
	30+ days old (4%)	75	37.7 (31.0-44.4)
Number of failed screening ears	One (64%)	1,633	55.9 (54.1-57.7)
	Two (36%)	946	56.4 (54.0-58.8)
Screening Place	Well-baby nursey (88%)	2,383	59.2 (57.7-60.7)
	NICU (12%)	196	34.2 (30.3-38.1)
Time between NHS and first follow-up	No follow-up (24%)	506	45.3 (42.4-48.2)
	< 30 days (44%)	1,322	66.0 (64.0-68.1)
	30+days (32%)	751	50.8 (48.3-53.4)
Total number of follow-ups	No follow-up (24%)	506	45.3 (42.4-48.2)
	One (67%)	1,860	60.6 (58.9-62.4)
	Two (7%)	166	52.0 (46.6-57.5)
	Three (2%)	47	50.5 (40.4-60.7)

Results

Characteristics of Follow-Up Hearing Appointment Scheduled Before Discharge

Of 93,996 children born to Louisiana residents between January 2015 and June 2016, 98.8% had NHS at birthing hospitals before discharge. Among newborns with NHS, 4,597 (5.0%) did not pass screening and needed further testing. Of those, 56.1% (2,579) were scheduled for follow-up hearing appointments prior to hospital discharge. The percentage was higher among newborns whose screening was conducted in the well-baby nursery (59.2% vs. 34.2% in NICU), those with screening before 30 days of age (57.0% vs. 37.7% for NHS after 30 days of age), normal weight babies (58.0% vs. 34.8% for very low birth weight and 48.3% for low birth weight), children living in rural areas (64.6% vs. 50.9% in urban areas), and those whose delivery was paid for by Medicaid (59.5% vs. 47.8% non-Medicaid). Table 1 indicates distributions of mother and child demographic characteristics and rate of follow-up hearing appointment scheduled prior to hospital discharge, henceforth referred to as hospital scheduled appointment. LTF/LTD, early follow-up initiation, and early completion of audiological diagnosis will be referred to as lost to system (LTS; Beauchaine & Hoffman, 2008), early follow-up, and early diagnosis, respectively.

Associations between Hospital Scheduled Appointment with Study Outcomes

Among newborns who failed NHS, rate of LTS (1,193 including 554 LTD and 639 LTF) after screening was 26.0% (95% confidence interval [CI]: 24.7–27.2). This rate was 20.9% (CI: 19.4–22.5) and 32.4% (CI: 30.3–34.4) among newborns with and without a hospital scheduled appointment, respectively. The adjusted regression model showed that the rate of LTS was 52% higher in children without a hospital scheduled appointment than in counterparts with a hospital scheduled appointment (adjusted prevalence ratio [PR] = 1.52, CI: 1.38–1.68; Table 2). Reasons for LTF were different between those with and without a hospital scheduled appointment. For the 312 LTF children with a hospital scheduled appointment, the most common LTF reason was missed appointment (85.6%), while no appointment made (59.0%) and missed appointment (32.7%) were the most common reasons for LTF among the 327 LTF children without a hospital scheduled appointment (Table 3).

In children who failed NHS and had follow-up, rate of early follow-up after screening was 57.5% (CI: 55.9–59.2). The rate was 63.8% (CI: 61.7–65.8) and 48.3% (CI: 45.7–50.9) among children with and without a hospital scheduled appointment, respectively. The rate of early follow-up was 25% higher among newborns with a hospital scheduled appointment than those without one (PR = 1.25, CI: 1.17–1.33; Table 2).

For children who failed NHS and completed hearing follow-up after screening, the rate of early diagnosis was 87.9% (CI: 86.9–89.0). The rate was 90.8% (CI: 89.5–92.0) and

83.6% (CI: 81.6–85.6) among newborns with and without a hospital scheduled appointment, respectively. There was no statistical association of early diagnosis with hospital scheduled appointment (PR = 1.02, CI: 0.98–1.05, p = 0.3620; Table 2).

Table 2. **Percent and Prevalence Ratio (PR) to Follow-Up Status after Newborn Hearing Screening**

	Percent (CI 95%)	Unadjusted PR (CI 95%)	*P* value	Adjusted PR (CI 95%)	*P* value
Loss to follow-up or loss to documentation, *N* = 4,597					
Follow-up not scheduled before discharge	32.4 (30.3-34.4)	1.55 (1.40-1.70)	< .0001	1.52 (1.38-1.68)	< .0001
Follow-up scheduled before discharge	20.9 (19.4-22.5)	1.0		1.0	
Total	26.0 (24.7-27.2)				
Early follow-up initiation, *N* = 3,480					
Follow-up not scheduled before discharge	48.3 (45.7-50.9)	1.0		1.0	
Follow-up scheduled before discharge	63.8 (61.7-65.8)	1.32 (1.24-1.41)	< .0001	1.25 (1.17-1.33)	< .0001
Total	57.5 (55.9-59.2)				
Early completion of audiological diagnosis, *N* = 3,404					
Follow-up not scheduled before discharge	83.6 (81.6-85.6)	1.0		1.0	
Follow-up scheduled before discharge	90.8 (89.5-92.0)	1.09 (1.06-1.12)	< .0001	1.02 (0.98-1.05)	0.36250
Total	87.9 (86.8-89.0)				

*Models of loss to follow-up/loss to documentation and early follow-up initiation were adjusted for race/ethnicity, sex, geography of residence at birth, maternal age and education, Medicaid-paid delivery, birth weight, number of previous live births, number of failed screening ears, age at newborn hearing screening and screening place. Two additional variables including total number of follow-ups and length of time between newborn hearing screening and first follow-up were adjusted for the model fo early completion of audiological diagnosis.

Table 3. **Reasons for Loss to Follow-Up among Children Who Failed Newborn Hearing Screening and Were Reported as Loss to Follow-Up after Screen (*N* = 639)**

	Follow-up not scheduled before discharge	Follow-up scheduled before discharge	Total
Reasons for loss to follow-up	Percent (number)	Percent (number)	Percent (number)
Missed/canceled appointment	32.7 (107)	85.6 (267)	58.5 (374)
Unable to contact parents	2.4 (8)	5.1 (16)	3.8 (24)
Rescreened by other provider	3.1 (10)	5.1 (16)	41.(26)
Moved out of state	0.6 (2)	2.2 (7)	1.4 (9)
No appointment made	59.0 (193)	0.0 (0)	30.2 (193)
Other	2.1 (7)	1.9 (6)	2.0 (13)
Total	100.0 (327)	100 (312)	100.0 (639)

Discussion

This study indicated that only 56% of newborns who did not pass NHS were scheduled for a follow-up hearing appointment prior to hospital discharge. The hospital scheduled appointment improved both LTF/LTD and early follow-up, but not early diagnosis. Newborns with a hospital scheduled appointment were less likely to be lost to the system and more likely to start follow-up early. No appointment made and missed appointment were the most common reasons for LTF among children without a hospital scheduled appointment, while missed appointment was the most common reason for LTF among children with a hospital scheduled appointment.

In Louisiana, hospital staff schedule a follow-up hearing appointment before discharge by contacting audiologists to schedule the appointment, and then notifying the parent of the appointment time and audiology facility. However, in some instances, the appointment cannot be scheduled due to discharge on a weekend when the audiology office is not open or due to a hospital policy that all follow-up appointments are scheduled after discharge. In these instances, post-discharge follow-up appointments are scheduled in different ways, and reasons for not making an appointment can be potentially explained by the following:

1. Hospital staff contact an audiologist to make the appointment, and then contact the parent to notify them of the appointment made. In this scenario, hospital staff may be unable to contact parents to inform them of the time and location of the follow-up.

2. Upon discharge, parents are provided with a list of select audiologists or even just a specific audiologist and instructed to make the appointment themselves. In many cases, the list provided by the hospital may not provide a viable choice for the parent because of location, hours, or other barriers. Furthermore, an appointment may not occur when the parent ignores the importance of re-screening, forgets to schedule an appointment, or could not make an appointment due to a language barrier (Holte et al., 2012).

3. Some physicians adopt a wait-and-see attitude and may not refer babies who failed NHS to audiology facilities for further testing because they may not realize the urgency of early intervention (Luz et al., 2016; Ravi et al., 2016; Shulman et al., 2010; Tran, Wang, et al., 2016).

Missed appointment was identified as the second most common reason for LTF among children without a

hospital scheduled appointment and the most common among children with a hospital scheduled appointment. The barriers leading to missed appointments have been reported in previous studies such as inaccessibility of follow-up facilities (Park, Warner, Sturgill, & Alder, 2006); 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 (which may be because screening results were not explained well or the importance of follow-up evaluation was not stressed), and overwhelming parent responsibilities (Shulman et al., 2010; Russ et al., 2010; Ravi et al., 2016; Liu et al., 2008).

Regarding loss to follow-up, results of our study were similar to previous studies. A study by Borders, Vess, Dumas, and Edlund (2016) indicated improvement of follow-up visit with a hospital scheduled appointment among Emirati infants who failed newborn hearing screening. Borders et al.'s study focused on nurse-led interventions, which included delivery of culturally specific educational counseling, prearranged follow-up appointments, and automated message reminders to parents whose newborns failed hearing screening at a hospital. Prearranged appointments were scheduled by nurses prior to discharge, and parents were registered for a follow-up phone message reminder 24 hours before the scheduled appointment. The rate of follow-up compliance within three months from birth improved from 25% to 75%. Aprahamian, Coats, Paysse, and Brady-McCreery (2000) also found that newborns with appointments scheduled by hospital personnel before discharge were more likely to be brought to a follow-up examination. Aprahamian et al.'s study included newborns who were at risk of retinopathy of prematurity and needed follow-up after discharge. Results of the analysis showed that 73% of patients with appointments scheduled by hospital personnel before discharge were brought to their follow-up appointment, compared with 37% of patients with appointment scheduled by parents after discharge.

Through this study and previous ones it is evident that follow-up improves when strategies implement a hospital scheduled appointment prior to discharge that includes a clear and universal protocol for appointment scheduling during discharge planning. A multidisciplinary team of nurses, physicians, audiologists, technicians, and administrative staff should be used to ensure that parents whose infants fail NHS receive proper counseling about the test results, clear communication with PCPs and follow-up audiologist, and a follow-up appointment for rescreening and/or diagnosis scheduled prior to hospital discharge (Ravi et al., 2016; Russ et al., 2010). The use of automated reminders is an effective strategy to ensure parents remember appointments once they are scheduled. Telephone calls or text messages 24 hours before an appointment will help to increase compliance for follow-up (Borders et al., 2016). The National Initiative for Children's Healthcare Quality (NICHQ), a nonprofit organization dedicated to bettering children's health and healthcare (NICHQ, 2013), has identified several strategies to decrease LTF for infants who do not pass screening. These include the following: (a) scripting the communication with parents after a failed NHS; (b) standardizing procedures for collecting contact information including alternate phone numbers or contacts and verifying the PCP prior to discharge; (c) scheduling the follow-up appointment before the family leaves the hospital and stressing the importance to the family; (d) calling the family to verify the follow-up appointment and provide assistance with transportation if needed; and (e) using fax-back forms between all parts of the care team, including at the time of the diagnostic evaluation to alert the PCP of the results and need for follow-up. (Russ et al., 2010; Spivak & Sokol, 2005).

Strengths and Limitations

This study had three major strengths: First, mother and child demographics and important characteristics of NHS and follow-up were controlled for in adjusted regression models to evaluate the independent effect of the study predictor—hospital scheduled appointments. Second, the study indicated most common reasons of LTF/LTD so that interventions to improve LTF/LTD were clearly delineated. Last, to date few peer-reviewed published papers have addressed the important effect of a hospital scheduled appointment on follow-up status among newborns who failed NHS.

The study included one major limitation—true follow-up status of children reported as LTD and verification of LTF. Verification of LTF/LTD can be done through contacting parents or follow-up facilities. This may help identify over-reporting problems and avoid misclassification of follow-up status (Tran, Wang, et al., 2016).

Conclusion

Scheduling follow-up hearing appointments prior to hospital discharge is strongly recommended. The study showed that hospital scheduled appointments improved not only LTF/LTD but also early follow-up initiation among newborns who failed NHS. The most common reason for LTF/LTD in children whose appointment was not scheduled before hospital discharge was no appointment made after discharge. Missed appointment was a main reason for LTF for children whose appointments were scheduled before or after discharge. Follow-up providers' lack of reporting of follow-up status among children whose appointments were scheduled after discharge made LTS worse for these children. Effective strategies focusing on improvement of hospital scheduled follow up appointments, missed appointments, and follow-up providers' reporting of results will help reduce both LTF/LTD and late follow-up.

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Parental Satisfaction and Objective Test Measurements Associated with Post-Partum versus Nursery Newborn Hearing Screening

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Abstract: Beth Israel Deaconess Medical Center (BIDMC; Boston, MA) initiated a change to the newborn hearing screening program in 2013 to encourage increased parental presence at the time of screening and support a more family-centered hospital environment. Newborn hearing screening program technicians were encouraged to conduct all hearing screens in the parent's post-partum rooms instead of in the nursery. To measure the effect of this change on the families' experience and screening measures, satisfaction surveys and retrospective data were collected over a 2-year period and compared. Newborn hearing screening program technicians and mother-baby nursing staff were surveyed to determine influence of this new process on their work flow. Results suggest post-partum room testing leads to an increase in family satisfaction without a resulting change in pass rates or decrease in the efficiency of screening activities or staff work flow.

Key Words: Parental satisfaction, hearing screening, auditory brainstem response, rooming-in, newborn

Acronyms: ABR = Auditory Brainstem Response; AABR = Automated Auditory Brainstem Response; BIDMC = Beth Israel Deaconess Medical Center; CCI = Committee on Clinical Investigations; NICU = Newborn Intensive Care Unit

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Background

Universal newborn hearing screening is performed in all 50 states as an effort to identify infants at risk for congenital hearing loss (Centers for Disease Control and Prevention, 2014). The recommendation from the Joint Committee on Infant Hearing (2007) to screen all infants prior to one month of age or before discharge is aimed at obtaining quicker diagnoses and earlier initiation of intervention. In the most recent CDC national data from 2014, over 97% of newborns are screened, with approximately 1.6% not passing their final screen. Although many infants who do not pass the hearing screening will go on to have a normal diagnostic test, approximately 1 to 2 per every 1000 babies are diagnosed with permanent hearing loss each year (CDC, 2014).

Many hospitals screen infants within a few hours or days of birth and past studies investigating parental feelings around the newborn hearing screening have highlighted the need to determine the best methods and practices to minimize worry and stress. Although most families express a positive view of the hearing screening process, those parents who express worry or skepticism often report feeling less informed (Vohr, Letourneau, & McDermott, 2001; Weichbold, Welzl-Mueller, & Mussbacher, 2001). To better educate families, many programs provide parents with written information regarding newborn hearing screening either on registration, with post-partum documentation, or at the time of testing. Weichbold et al. (2001) showed that parental presence at the time of hearing screening decreased skepticism and that mothers who attended the hearing screen had a more positive view of the program. A newborn hearing screening survey distributed to parents in Massachusetts regarding satisfaction with the Early Hearing Detection and Intervention program in 2007 highlighted parental presence at the time of the screening as the most frequent suggestion for program improvement (MacNeil, Liu, Stone, & Farrell, 2007).

Today, many birthing hospitals are moving toward increased direct parental care of the infant during hospitalization. Studies have shown that rooming-in, the practice of keeping newborns in the mother's post-partum room instead of in a nursery, provides numerous

benefits to families and babies, including improved sleep, better bonding, and more successful breastfeeding with increases in milk production and duration of nursing (Crenshaw, 2007). Svensson, Matthiesen, and Widstrom (2005) found that staff attitudes on rooming-in can subsequently influence the parent's attitude. In their study, mothers who did not room-in with their babies were more likely to feel that hospital staff believed the baby should stay in the nursery. Beth Israel Deaconess Medical Center (BIDMC), located in Massachusetts, increased efforts to promote a more family-centered environment in 2012 and to support family-centered initiatives, the BIDMC Newborn Hearing Screening Program started performing screenings in the post-partum rooms in May 2013.

The Massachusetts Universal Newborn Hearing Program Birth Facility Guidelines (2012) requires measures of quality assurance when evaluating and monitoring the success of a program. Satisfaction surveys are one of a variety of methods used in health care services to assist with the assessment of outcomes and provide information to plan quality improvement (Castle, Brown, Hepner, & Hayes, 2005). Parent surveys have proven to be a reliable and informative way to assess family satisfaction with newborn hearing screening and diagnostic follow-up programs (Mazlan, Hickson, & Driscoll, 2006). Mazlan et al.'s study of 80 families found high test–retest reliability with their survey suggesting parental feelings associated with the newborn hearing program did not vary significantly over time. The study also indicated that the majority of parents, when surveyed, expressed high levels of satisfaction with the quality of services they received.

This project was aimed at determining the effects of a change in hearing screening location on the satisfaction levels of families, staff workflow, and efficiency of the screening process and outcomes.

Method

This quality improvement project was presented to the Director of Operations for Committee on Clinical Investigations (CCI) at BIDMC in accordance with CCI policy and deemed not to constitute human subjects research.

Equipment

Newborn hearing screening program technicians conducted an automated auditory brainstem response (AABR) screening on all babies using the Natus Algo 5 Newborn Hearing Screener. Testing was conducted using automated parameters with a 35 dBnHL click stimulus, a 60 Hz Notch Filter, and a rate of 37 clicks/second. The screening protocol used was consistent with the Massachusetts Universal Newborn Hearing Screen Program (2012) guidelines. The ALGO 5 screening parameters are as follows: (a) Equipment produces a "passing" result when at a minimum of 1000 clicks, it establishes a > 99% statistical confidence that the auditory brainstem response (ABR) signal is present and matches the internal template. (b) A "refer" result is produced if the equipment reaches 15,000 clicks and has not established with a > 99% statistical confidence the presence of an ABR signal that matches the internal template (Natus Medical Incorporated, 2011). (c) Impedance levels of the electrodes must be below 12 kOhms individually and within 5 kOhms of each other for testing to commence. Natus Medical (2011) designates excessive myogenic interference at greater than 50% and excessive ambient noise at greater than 30%.

Protocol

Hearing screens were completed at least 12 hours after vaginal birth and 24 hours after cesarean birth. At the onset of the project, the technicians were educated about the benefits of rooming-in and were encouraged to begin screening all infants in the family's post-partum room. Screening in the nursery was discouraged unless the baby was not allowed to be in the post-partum room due to medical concern or family request. Babies who were in the nursery due to blood draw, circumcision check or due to family wishes had testing delayed until they were re-united with their parents. When a screening was conducted in the nursery, families were not present. Although initial post-partum room screening rates were low, these continued to rise throughout the project period. All other protocols regarding screening procedure remained unchanged. At the completion of the screenings in either setting, the technicians verbally shared the screening results with all parents, answered any questions, and provided them with written information on the final results, follow-up and hearing and language developmental milestones. All surveys were distributed to families after the infant received a final screen result (first screen pass; second screen pass; second screen refer).

Survey Data Collection Procedures and Parent Participants

Instrument. A five-item survey, developed by Beth Israel Deaconess Medical Center Newborn Hearing Screening Program, was used to obtain parental feedback about their satisfaction with the newborn hearing screening (see Appendix A). Responses to questions were anonymous, completed using paper and pencil, and satisfaction was rated using a 5-point Likert scale (1 = *not at all satisfied*; 5 = *extremely satisfied*). The survey queried parent perceptions of the information they received prior to testing, the test process, the results, information regarding follow-up, and the overall process.

Data collection. Surveys were collected over two time periods. The first data collection period was from September 2013 to December 2013. Surveys were distributed and collected daily during those months (n = 201; 103 nursery, 98 post-partum room). Due to time required to distribute and collect surveys and technician availability the collection of surveys was discontinued

for two months. The second data collection period was from March 2014 to December 2015. During this time, surveys were distributed and collected on the last day of each month (n = 164: 36 in nursery; 128 in post-partum room). A total of 365 completed surveys were obtained (n = 139 nursery; n = 226 post-partum room). There were a total of 9,861 infants screened during the two collection periods, indicating 4% of the population surveyed. Hospital demographic data revealed average maternal age of 32 and 45% of mothers reporting their race as White. Survey data were analyzed using Wilcoxon Rank Sum test.

From March 2014 to March 2015, the technicians tracked the number of surveys they distributed and collected. When comparing locations, there was a small difference in percentage of surveys returned (75% in nursery, n = 9/12; 79% post-partum room, n = 50/63). Return rate data was not analyzed for significance due to small sample size of nursery surveys.

Survey Data Collection Procedures and Staff Participants

Instrument. A five-item survey (nurses) and six-item survey (technicians), developed by Beth Israel Deaconess Medical Center Newborn Hearing Screening Program was used to obtain feedback from the mother-baby nursing staff and newborn hearing screening technicians in March 2015. Responses to the survey were obtained using Qualtrics, an online survey generator and rated using a 5-point Likert scale (1 = *not at all*; 5 = *extremely*). The survey queried staff perceptions about the effect of the post-partum room testing process on comfort level with the program and influences on work process (see Appendix B and C). Completion of the survey was voluntary.

Participants. BIDMC employs part-time technicians to perform hearing screenings on the well-baby units. Anywhere from two to five technicians are employed at one time and each work between two to four days per week. Technicians in the program are typically pursuing education in the healthcare field but may have limited occupational experience in the medical setting and with handling newborns. The survey was distributed to both current and previous employees. At the time of this project the technicians' average amount of time working with the program was 11 months (range 1–26 months). A total of eight hearing screening technicians were emailed the link to the survey with a 100% response rate (n = 8).

The mother-baby nursing staff does not perform hearing screenings, but are responsible for completing paperwork regarding risk factors for late onset hearing loss. Mother-baby nurses are in close contact with families and newborn hearing screening technicians and are usually the first to know when a family has a concern or question. A total of 120 mother-baby nurses were emailed the link to the survey with a 20% response rate (n = 24).

Objective Data Collection and Participants

Objective hearing screening outcome data were collected retrospectively. All infant hearing screenings between September 2013 and December 2015 with a final result of "pass both", "refer both", "refer right", or "refer left" were eligible for inclusion in the study. Per hospital policy, infants that did not pass the first screen received a second hearing screen prior to discharge. Eleven percent of infants required a second hearing screening. Infants were not screened more than twice and infants that did not pass the first screen had only their second (final) screen included in the data analysis. Screens completed on infants in the neonatal intensive care unit (NICU) were excluded due to inconsistencies in time of screening and parental presence. Total number of infant screens included in this measure was 10,538 (7,588 post-partum room; 2,950 nursery).

Objective data was downloaded biweekly from the ALGO 5 between September 2013 and December 2015 and was analyzed using the t-test procedure and Satterthwaite method for unequal variances. Test parameters such as duration of screen, myogenic interference (muscular or electrical interference), ambient noise, and screening results, were compared to determine if the change in location resulted in any objective differences. Screen duration (total time in seconds the ALGO device was actively screening) was assessed because significant increases would decrease technician efficiency and increase cost of program. Screening results were compared to determine consistency with national, state, and program pass rates.

Myogenic interference (time during the test that the equipment was not accumulating data due to myogenic interference) and ambient noise were reported by the equipment in percentages. Myogenic interference and ambient noise percentages were assessed because changes in either of these parameters may lead to changes in screen accuracy. Along with infant activity level, myogenic interference can also be created by electrical interference. It is possible that minimal electrical activity was present in both the post-partum room and in the nursery.

Results

Survey Data

A total of 365 family satisfaction surveys were collected which included 226 surveys from families with screening in the post-partum room and 139 surveys from families with screening in the nursery. Figure 1 shows an analysis of satisfaction measures that reveals parents report higher satisfaction levels for post-partum room screening compared to nursery screening for information prior to screen ($p < .0001$); testing process

($p < .0001$); information on results ($p < .01$); information on follow-up ($p < .01$); and overall program ($p < .0001$). Newborn hearing screening technician survey results indicated the technicians' comfort in screening was

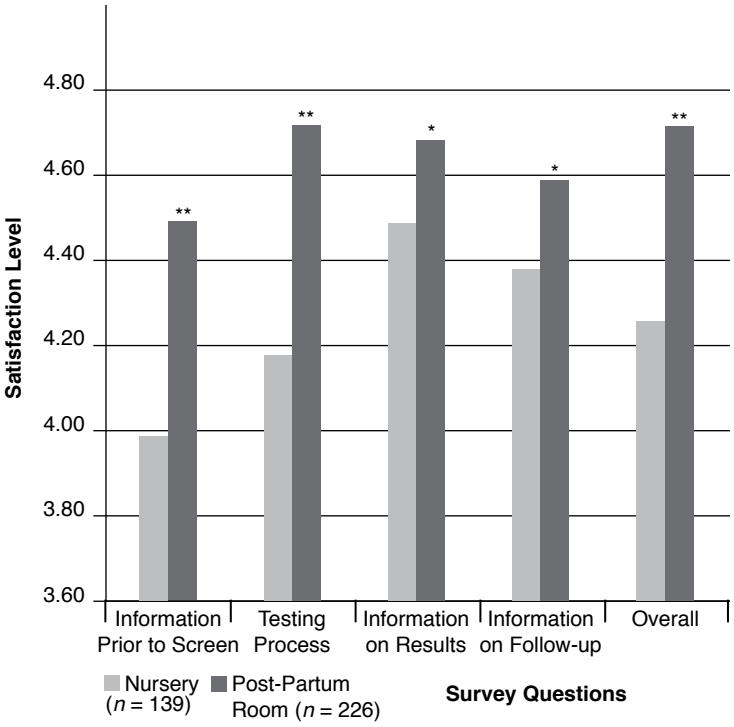

Figure 1. **Mean satisfication rating for families with post-partum room or nursery hearing screening**
***$p < .01$, **$p < .0001$**

higher when testing was completed in the nursery (5) compared to in the post-partum room (4.25). Five of the technicians were equally comfortable quieting the infant in both settings, but three were more comfortable quieting the infant in the nursery. Issues relating to hearing screens such as equipment handling, missing paperwork, parental involvement, and disruptions from other staff members or family did not appear to create problems in either setting for the technicians. Data were not analyzed for significance due to small sample size.

The mother-baby nurse survey results indicated they are very comfortable regarding the hearing screening process in both locations (4.3 in the post-partum room; 4 in the nursery). Some nurses reported disruption in their work flow when the hearing screens were done in the post-partum room (15%), but the majority (85%) felt the change in screen location caused either no issues or allowed the nurses to improve their work flow.

Objective Data

The objective measure analysis in Table 1 shows the comparison of percent of myogenic interference, percent of ambient noise, and screen duration between screens conducted in the post-partum room and screens conducted in the nursery. Analysis revealed the percent of myogenic interference present during screens was always slightly higher in the post-partum room. There were significantly higher rates of myogenic interference during post-partum room testing compared to nursery testing for 2013 ($p = .007$) and 2014 ($p < .0001$). In 2013 there was also a significant difference in both the percent of ambient noise and duration of the screen ($p = .03$ and $p = .005$ respectively). By 2015 no significant differences remained in any measure between the two locations.

Hearing screen results were similar in both locations. Post-partum room pass rates for 2013, 2014, and 2015 were 98.2%, 98.2% and 98.7% respectively. Nursery pass rates for 2013, 2014, and 2015 were 97.3%, 97.7% and 98.5% respectively.

Table 1
Average Myogenic and Duration Measures across Location and Year

	Post-Partum Room ($N = 7588$)	Nursery ($N = 2950$)
2013	$n = 508$	$n = 925$
Myogenic (%)	2.6**	22.4
Ambient (%)	2.2**	1.6
Duration (sec)	322.1*	284.7
2014	$n = 3176$	$n = 1287$
Myogenic (%)	27.5**	24.3
Ambient (%)	1.6	1.4
Duration (sec)	307.4	295.2
2015	$n = 3904$	$n = 738$
Myogenic (%)	27.6	26.9
Ambient (%)	1.7	1.9
Duration (sec)	307	302.4

*$p < .05$; **$p < .01$

Discussion

The primary purpose of the project was to determine if there was an increase in family satisfaction when screenings were moved from the nursery to the post-partum rooms without negatively impacting passing rates or markedly decreasing the efficiency of the screening.

Subjective satisfaction survey results indicated that families were satisfied with both post-partum room and nursery hearing screenings which is consistent with Mazlan et al. (2006), who showed that greater than 95% of parents are highly satisfied with hearing screening programs. This project determined that although both settings are highly rated, parents were significantly more satisfied with all measured aspects of the post-partum room hearing screening program including the information they received prior to testing, the test process, the results, information regarding follow-up, and the overall program. Parental presence, direct observation of the testing procedure and immediate access to technicians to answer questions, could

lead to an increased comfort level and understanding of information resulting in higher satisfaction levels. Weichbold et al. (2001) reported mothers who were present at the hearing screening gained some impressions from the situation and these impressions along with information received most likely added to their positive views of hearing screening.

Staff surveys suggest that the technicians have a slight preference for testing in the nursery. This is possibly influenced by their rating of comfort level in quieting the baby in parents' presence which was lower than comfort level in the nursery. Based on personal discussions with staff, comfort level has the potential to improve with increased length of employment and experience. Technicians were equally comfortable answering parent questions in both settings and although there was some reported increase in perceived complexity associated with testing in the post-partum rooms, it was not reported as problematic.

Mother-baby nurses were enthusiastic about the post-partum room hearing screening process and rated their feelings toward it slightly higher than nursery screenings. Nurses felt that the location of testing did not drastically affect their part of the hearing screening process and although there are some changes in work flow, the nurses felt that the change to post-partum room screening did not negatively impact their other work processes.

Objective data analyzed consisted of percent of myogenic interference, ambient noise, screen duration, and screen results. Screening results remained similar in both locations over the project period and was consistent with Massachusetts' pass rate of 98.2% (Massachusetts Universal Newborn Hearing Screening Program, 2014). Data analysis of myogenic interference, ambient noise, and screen duration, showed significant differences between post-partum room and nursery screenings in 2013, but by 2015 those differences no longer persisted and screenings from both locations demonstrated similar levels for each measure.

Limitations

Several limitations of the project were identified. Though our survey response rate was good (75–79% completion of parents offered survey), the responses reflect a sample of only 4% of our total population during the period assessed. Sample size was restricted by limitations on hearing screening technician time and resources. Non-English speaking families were not surveyed therefore the survey population may not be representative of the total family population (88% of BIDMC's maternal population is English speaking). Surveys were anonymous so obtaining demographic data to assist in comparing differences in who completed the survey (maternal vs. partner), ethnicity, or maternal age were unable to be evaluated.

Future directions

Future direction includes plans to determine if the location of the screening had any influence on follow-up rates for infants who do not pass the screening and those with identified risk factors for late onset hearing loss.

Conclusion

The purpose of the project was to examine the association between family satisfaction, objective test measurements, and location of newborn hearing screenings. After performing a survey of 365 families and reviewing objective test data from 10,538 infant screens, the project demonstrated that conducting hearing screenings in the post-partum room increased family satisfaction while not negatively influencing objective test measurements. Changes in program process may result in small impacts on staff work flow.

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Appendix A

BIDMC Newborn Hearing Screening Program Satisfaction Survey

Thank you for giving us the opportunity to find out more about the effectiveness of our hearing screening program. Your answers below will be kept confidential and we will not be collecting any patient information.

Where was your infant's hearing screened? ☐ In your room ☐ In the nursery

Thinking back on your baby's hearing screening at BIDMC, please circle the number that shows how satisfied you were with each part of the process.

1. How satisfied were you with the information you received about hearing screening **prior** to your infant's testing?	Not at all Satisfied 1	Slightly Satisfied 2	Moderately Satisfied 3	Very Satisfied 4	Extremely Satisfied 5
2. How satisfied were you with the **testing process**?	Not at all Satisfied 1	Slightly Satisfied 2	Moderately Satisfied 3	Very Satisfied 4	Extremely Satisfied 5
3. How satisfied were you with the information regarding the **results** of the hearing screening?	Not at all Satisfied 1	Slightly Satisfied 2	Moderately Satisfied 3	Very Satisfied 4	Extremely Satisfied 5
4. How satisfied were you with the information you received regarding **follow-up**?	Not at all Satisfied 1	Slightly Satisfied 2	Moderately Satisfied 3	Very Satisfied 4	Extremely Satisfied 5
5. **Overall**, how satisfied were you with the hearing screening services provided to your baby and family?	Not at all Satisfied 1	Slightly Satisfied 2	Moderately Satisfied 3	Very Satisfied 4	Extremely Satisfied 5

6. Comments: _____

_____

_____

_____

Please place the completed survey back in the envelope and seal closed. It will be collected by the hearing screening technician later today.

Thank you,
Beth Israel Deaconess Medical Center Newborn Hearing Screening Program

Appendix B

In Room Hearing Screenings: Mother-Baby Nurse Survey

In the past year we have transitioned to performing the majority of our hearing screenings in the parent's post-partum room as opposed to the nursery. While we are continuously assessing the satisfaction levels of our families, our program was interested in the opinions of our staff as well. Please take a few moments and complete the following survey. In order to keep this anonymous, please feel free to drop it in my mailbox or on my desk in a sealed envelope.

Thank you,

Newborn Hearing Screening Program

1) How comfortable are you with the hearing screening process when done in the...

nursery?	Not at all	Slightly	Moderately	Very	Extremely
	1	2	3	4	5
post-partum room	Not at all	Slightly	Moderately	Very	Extremely
	1	2	3	4	5

2) How problematic are these issues to the <u>in room</u> hearing screening process?

A - Finding out test results	Not at all	Slightly	Moderately	Very	Extremely
	1	2	3	4	5
B - Contacting the technician	Not at all	Slightly	Moderately	Very	Extremely
	1	2	3	4	5
C - Questions from parents regarding the test	Not at all	Slightly	Moderately	Very	Extremely
	1	2	3	4	5
D - Completing hearing screening forms on test day	Not at all	Slightly	Moderately	Very	Extremely
	1	2	3	4	5

3) How problematic are these issues to the <u>nursery</u> hearing screening process?

A - Finding out test results	Not at all	Slightly	Moderately	Very	Extremely
	1	2	3	4	5
B - Contacting the technician	Not at all	Slightly	Moderately	Very	Extremely
	1	2	3	4	5
C - Questions from parents regarding the test	Not at all	Slightly	Moderately	Very	Extremely
	1	2	3	4	5
D - Completing hearing screening forms on test day	Not at all	Slightly	Moderately	Very	Extremely
	1	2	3	4	5

4) Does having the hearing screening done in the room influence your work in the <u>nursery</u>?

A - Yes — Improves it

B - Yes — Hinders it

C - No — Neither improves nor hinders it

4) Does having the hearing screening done in the room influence your work in the <u>post-partum room</u>?

A - Yes — Improves it

B - Yes — Hinders it

C - No — Neither improves nor hinders it

Appendix C

In Room Hearing Screenings: Hearing Screening Technician Survey

In the past year we have transitioned to performing the majority of our hearing screenings in the parent's post-partum room as opposed to the nursery. While we are continuously assessing the satisfaction levels of our families, our program was interested in the opinions of our staff as well. Please take a few moments and complete the following survey

Thank you!

1) How comfortable are you...

screening babies in the nursery?	Not at all 1	Slightly 2	Moderately 3	Very 4	Extremely 5
screening babies in the room?	Not at all 1	Slightly 2	Moderately 3	Very 4	Extremely 5

2) Do you feel comfortable taking the necessary steps to quiet/calm baby in both settings?

A - Yes, just as comfortable in room as in nursery

B - No, more comfortable in nursery

C - No, more comfortable in room

3) Is parental presence helpful in ensuring baby stayed quiet/calm during testing?

No, never	Rarely	Occasionally	Regularly	All the time
1	2	3	4	5

4) Do you feel as comfortable answering questions in both settings?

A - Yes, just as comfortable when baby tested in room as in nursery

B - No, more comfortable explaining when parents **have not** seen test (nursery)

C - No, more comfortable explaining after parents **have** seen test (room)

5) How problematic are these issues to the in room hearing screening process?

A - Equipment handling	Not at all 1	Slightly 2	Moderately 3	Very 4	Extremely 5
B - Missing paperwork	Not at all 1	Slightly 2	Moderately 3	Very 4	Extremely 5
C - Parental involvement	Not at all 1	Slightly 2	Moderately 3	Very 4	Extremely 5
D - Disruptions from other hospital staff members	Not at all 1	Slightly 2	Moderately 3	Very 4	Extremely 5
E - Disruptions from family (TV, other kids, talking)	Not at all 1	Slightly 2	Moderately 3	Very 4	Extremely 5

6) How problematic are these issues to the nursery hearing screening process?

A - Equipment handling	Not at all 1	Slightly 2	Moderately 3	Very 4	Extremely 5
B - Missing paperwork	Not at all 1	Slightly 2	Moderately 3	Very 4	Extremely 5
C - Parental involvement	Not at all 1	Slightly 2	Moderately 3	Very 4	Extremely 5
D - Disruptions from other hospital staff members	Not at all 1	Slightly 2	Moderately 3	Very 4	Extremely 5
E - Disruptions from family (TV, other kids, talking)	Not at all 1	Slightly 2	Moderately 3	Very 4	Extremely 5

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Measuring Nurses' Knowledge and Understanding of Universal Newborn Hearing Screenings

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Abstract: The present study was conducted to investigate the knowledge and understanding of Universal Newborn Hearing Screening (UNHS) in nursing professionals. A group of 15 adult, licensed nurses with varied professional experience participated in the study. Participants completed both objective and subjective measurements to evaluate their current knowledge of training procedures. A free online UNHS training program offered through the National Center for Hearing Assessment and Management (NCHAM) served as both the objective measure and training module. Participants completed pre- and post-surveys as the subjective measure of their UNHS training. Results of these surveys were compared across the participants to determine significant outcomes. Results of this study suggest nursing professionals are not confident in their current level of training concerning UNHS. Additionally, the participants' scores on the online training module testing showed improvement from pre- to post-testing.

Key Words: Universal Newborn Hearing Screening (UNHS), National Center for Hearing Assessment and Management (NCHAM)

Acronyms: AAA = Academy of Audiology; ADPH = Alabama Department of Health; ASHA = American Speech-Language-Hearing Association; EHDI = Early Hearing Detection and Intervention; NICU = Neonatal Intensive Care Unit; NIDCD = National Institute on Deafness and other Communication Disorders; NIH = National Institutes of Health; UNHS = Universal Newborn Hearing Screening

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Introduction

The most critical period of speech and language development occurs during the first three years of life while the child's brain changes and develops. According to the National Institute on Deafness and other Communication Disorders (NIDCD), if a child is not immersed in language and communication during this time due to hearing loss, other difficulties arise involving the development of speech, language, and reading abilities (2014). Children with undetected hearing loss often fall behind their peers in school concerning the development of language, cognitive, and social skills. It is difficult to interact, learn, and follow social cues when a hearing loss is present. Academic performance, problem solving skills, and even long-term job opportunities can be affected by hearing loss in infancy or early childhood (National Institutes of Health, 2010).

Prior to discharge, a baby's hearing is screened to determine if further testing is needed to verify if the child is deaf or hard of hearing (DHH; Nierengarten, 2016). Normal hearing is vital for the typical development of speech and language in children. Children who are DHH often have delayed speech and language development when the hearing loss is left unidentified. Further, hearing loss can be one of many symptoms associated with genetic, congenital disorders (Morten, 1991). According to the National Institutes of Health (NIH), "2 to 3 of every 1000 children in the United States are born deaf or hard-of-hearing." This does not include children with milder hearing impairments or those who develop hearing loss later in childhood (NIH, 2010).

Universal Newborn Hearing Screening (UNHS) programs are now established in all states and U.S. jurisdictions (American Speech-Language-Hearing Association [ASHA], n.d.). Across most of the country, health care workers complete, document, and report the baby's hearing results to the state. Typically, the state early hearing and intervention coordinator will then follow a baby's hearing results until a passing result is documented for each ear or a hearing loss is identified and an early intervention plan is determined.

In February 2001, the Alabama Department of Health (ADPH) initiated a state-wide universal newborn hearing screening (UNHS) program, known as Alabama's Listening!. The main goals of the initiative were to establish statewide UNHS programs to ensure early identification of children who are DHH, identify the appropriate treatment of each individual child identified with hearing loss, and propagate the necessary early intervention services to aid families of children identified as DHH (ADPH, 2014). Since that time, UNHS programs have developed under the guidelines of the Joint Committee of Infant Hearing (JCIH) Position Statement (2007) and the regulation and documentation of the ADPH. Across the state, hospitals conduct hearing screenings on all newborns to evaluate the status of their hearing sensitivity at birth. Although this process has shown progress in the number of babies screened, professionals who conduct the screens are often not up-to-date on current procedures, equipment, or documenting the results of such testing (Gallagher, Easterbrooks, & Malone, 2006).

UNHS programs aim to promote the identification of congenital hearing loss as early as possible in order to initiate appropriate early intervention services and supports. Although the UNHS program has been successful in lowering the age at which children who are DHH are identified, the immense variation of training procedures used across the state leaves confusion concerning the effectiveness of training offered at different locations (Pallarito, 2012; Parving & Salomon, 1996). The purpose of this project was to obtain information concerning the knowledge and understanding of nurses in administering UNHS procedures, correctly documenting results, and clearly explaining outcomes to parent/guardians.

This research aims to aid in identifying areas in which current training is lacking. The information obtained can be used by hospitals across the state to better equip their personnel to conduct UNHS and deliver appropriate results to families. A better understanding of UNHS serves to aid parents and professionals in initiating the appropriate treatment for children identified as DHH (DesGeorges, 2003).

Literature Review

In the past, only babies born into circumstances that put them at a higher-than-normal risk for hearing loss were screened. These risk factors included low birth weight, a stay in the Neonatal Intensive Care Unit (NICU), and other difficulty at time of birth (NIH, 2010). On average, children were identified as being DHH around 30 months (JCIH, 2000). This was simply not good enough to ensure the appropriate intervention and management of hearing loss in young children. The need for a better screening protocol was as clear as the detrimental effects of unidentified hearing loss were prevalent. Finally, in 1993, the National Institutes of Health (NIH) held a conference that endorsed the screening of all newborns for hearing loss before the child left the hospital. This consensus bolstered a state-by-state effort to promote mandatory screenings of newborns. In addition, Congress passed the Newborn and Infant Hearing Screening and Intervention Act of 1999, which helped coordinate and fund statewide programs while promoting awareness (NIH, 2010). Increased recognition sparked new research. Study after study found that children who are DHH and received intervention at an earlier age demonstrated language skills comparable to their age-matched peers regardless of degree of hearing loss (Moeller, 2000; White & White, 1987; Yoshinaga-Itano, Sedey, Coulter, & Mehl, 1998).

Across the country, all states have implemented Early Hearing Detection and Intervention (EHDI) programs to aid in the identification of children who are DHH. The EHDI guidelines are based on a "1-3-6" goal: newborns should receive a hearing screen no later than 1 month of age, infants that did not pass their initial hearing screening should have appropriate audiological and medical evaluations to confirm the presence of hearing loss no later than 3 months of age, and all infants with confirmed permanent hearing loss should receive early intervention services as soon as possible after diagnosis but no later than 6 months of age. Further, use of amplification should begin within 1 month of diagnosis and ongoing audiological management should not exceed 3 month intervals (Ditty, 2007; JCIH, 2007). Early intervention is associated with attaining language abilities of age-matched peers, which is one of the main goals of programs that support children who are DHH (JCIH, 2007). UNHS has become a routine practice in hospitals across most states. This practice provides a significant benefit to children identified as being DHH at birth because early intervention can be initiated earlier (NIH, 2010; Pallarito, 2012). Although significant progress has been made, not all newborns are screened after birth. In some cases parents refuse testing, a qualified screener is not available, or a more pressing medical condition may take priority over hearing screenings (DesGeorges, 2003). Additionally, of the babies who are screened but did not pass the hearing screen, only 34.4% return for follow-up testing to confirm if a hearing loss is present (Centers for Disease Control and Prevention [CDC], 2014). Reasons for lack of follow-up may be due to some families being unable to afford the cost to travel to audiologists (especially in rural settings), some parents may be in denial about the presence of a hearing loss, and some families may not be aware of the need for a follow-up appointment (ASHA, n.d.; Ditty, 2007). These children continue to risk late identification and delay intervention, leading to similar outcomes seen before the implementation of UNHS.

Although UNHS programs are in place at hospitals around the country, training provided to those employees and volunteers who screen newborns is widely varied.

The American Academy of Audiology (AAA) and the American Speech-Language-Hearing Association (ASHA) recommend training programs that are under the direction of a supervising audiologist with content that exceeds the simple understanding of how to use the equipment and addresses the competency of screening personnel. Additionally, it is advised that personnel should be retrained every two years with continuing assessment (AAA, 2014; ASHA, n.d.). Often other factors, such as finances and available staffing, are the main factors in determining what personnel, training, equipment, and procedures are implemented. Although most states are required to submit documentation of hearing screen results to their state EHDI for follow-up, even this process is not uniform among hospitals within and across individual states (Pallarito, 2012).

Despite the progress that has been made with the implementation of UNHS, several challenges complicate the goal of early detection of children who are DHH, including the number of infants born annually and personnel available for screening in newborn nurseries (AAA, 2014). In 2010, President Obama signed legislation that reauthorized and expanded EHDI law and emphasized the importance of appropriate and efficient diagnosis of hearing loss by a trained professional so that children who are DHH may receive appropriate services and are provided access to intervention earlier (Pallarito, 2012). Even with increased funding and governmental support, establishing and maintaining programs with uniform practices has proven difficult due to a lack of awareness of health providers and the lack of training of personnel (Gallagher et al., 2006; Houston, Bradham, & Guignard, 2011). Appropriate training of the screeners and their competence with giving correct information to parents is essential to the early intervention process (Laugen, 2013).

Ultimately, hearing loss remains the most prevalent developmental disorder that is identifiable at birth. Even though it is clear this situation demands a solution to better serve children and their families, a lack of identification and appropriate management continues to have significant impact on children's educational, cognitive, and social development (AAA, 2011). Understanding the clinical effectiveness (and shortcomings) of the current knowledge of hearing screening providers will assist in improving current training programs. This information provides feedback concerning how training programs are currently functioning, and how the programs in place can be made better for both professionals and patients (Gallagher et al., 2006; Houston et al., 2011).

Methods

Participants

Participants in this study were required to be employed and licensed as a nurse and 19 years of age or older. Additional participant demographics are seen in Table 1. For this study, the participants included 15 nursing professionals with varied career experience ranging from 0–1 year to more than 5 years of experience, with a scope of practice that includes newborn hearing screening. All participants were licensed in the nursing field at the time of this study. The majority of the participants have worked in their current position for 5 or more years. All participants completed 0–5 newborn hearing screenings per day, and no more than 10 per week. Of those who participated, half had never interpreted the results of the hearing tests they completed, while the other nurses interpreted 0–5 of the newborn hearing screenings they conducted. The majority of the participants have never counseled parents or guardians on the results of the newborn hearing screenings.

Table 1
Participant Demographics

	0-1 Years	1-3 Years	3-5 Years	5 or More Years
Length of Time in Current Positon	2	5	1	7
Years of Experience	8	0	1	6

	0-6 Months	6-12 Months	1-2 Years	2-5 Years	5 or More Years	Never
Time Elapsed Since Initial Training	10	0	1	1	3	0
Most Recent Continuing Education on UNHS	3	0	0	0	0	0

	0-5 Screenings	5-7 Screenings	7-10 Screenings	15 or more Screenings	Never
UNHS Conducted Per Day	15	0	0	0	0
UNHS Conducted Per Week	14	1	0	0	0
UNHS Interpreted Per Day	7	0	0	0	8
UNHS Interpreted Per Week	5	1	0	1	8
	Never	Rarely	Sometime	Often	Always
UNHS Counseling Per Day	12	2	1	0	0
UNHS Counseling Per Week	11	3	1	0	0

Recruitment

This study recruited participants from local hospitals, through posts on social media, and across university nursing school departments. Participants were sent individual packets of information containing the consent forms and project instructions. Additionally, packets contained directions for completing the online UNHS training, how to submit online test scores, and

the three hard-copy surveys they were required to complete (participant demographics, pre-training, and post-training surveys). Contact information for the investigators was included in each of the information packets. Participants were required to sign an Institutional Review Board (IRB) approved informed consent form outlining the study, their requirements, and their potential benefits and risks before completing the project.

Procedures

In order to obtain information concerning the effectiveness of UNHS training procedures, participants were asked to complete both subjective and objective measures. Three subjective surveys were used to collect information: a participant demographics survey with 11 questions, a pre-survey with 15 questions, and a post-survey with the same 15 questions. These were all presented in the same hard-copy format.

The pre-survey measured their knowledge and understanding of UNHS according to their previous level of training in UNHS. This information provided a baseline for how the participants believed their current understanding of UNHS was and showed which areas they had less confidence in their training. The National Center for Hearing Assessment and Management (NCHAM) online training modules (Interactive Newborn Hearing Screening Training Curriculum at http://www.infanthearing.org/nhstc/index.html) were used as both an objective measure of participants' knowledge and the continuing education course to update professionals on the current best practices for UNHS. See Table 2 for the list of topics covered in the Interactive Newborn Hearing Screening Training Curriculum. The participants were asked to complete the provided pre-test within the Interactive Newborn Hearing Screening online course and then complete the training modules. Next, the post-test within the Interactive Newborn Hearing Screening online course was completed to assess their knowledge after completing the training modules. The post-survey asked the same questions and provided the same answer choices as the pre-survey but was completed after finishing the Interactive Newborn Hearing Screening online course training modules. The participants printed their pre- and post-test scores and submitted them to the investigators. Data was collected by the investigators and de-identified before any analysis was completed. Participant responses to the subjective pre- and post-surveys were coded in order to conduct statistical analyses. The results of the pre- and post-surveys of both the subjective and objective measures were compared across the participants to determine any significant outcomes.

Table 2
List of topics in the Interactive Newborn Hearing Screening Training Curriculum

Introduction to screening
Preparing to screen
Screening with otoacoustic emissions (OAEs)
Screening with automated auditory brainstem response (A-ABR)
Communicating with parents and providers
Completing the screening process
Screening babies with risk indicators
Outpatient screening and re-screening

Participants completed the surveys and online training in their free time. The participants were not compensated for the time they spent completing the project. Those who took part in the study could use this training to fulfill some of their continuing education hour requirements for their licensure based on their guidelines from work. The only risk to the participants who completed the study was breach of confidentiality. To minimize this risk for those in the current study, their information was de-identified and coded using a numbering system. There were no costs to the participants to complete the research. Participation was completely voluntary and any participant could choose to withdraw from the study at any time, at which point their information (if, identifiable) would be destroyed.

Results

Subjective Survey Responses

All participants completed surveys pre- and post-training. Not all participants completed every question; answers that were not recorded by the participant were deleted from the analysis. Responses to each question can be found in Figures 1–14. Within subject one-way repeated measures ANOVAs were conducted to assess if there were significant differences between the participants' pre- and post-training responses. Responses were statistically significant for questions 1–2, 4–5, 7–8, 10, and 12–14.

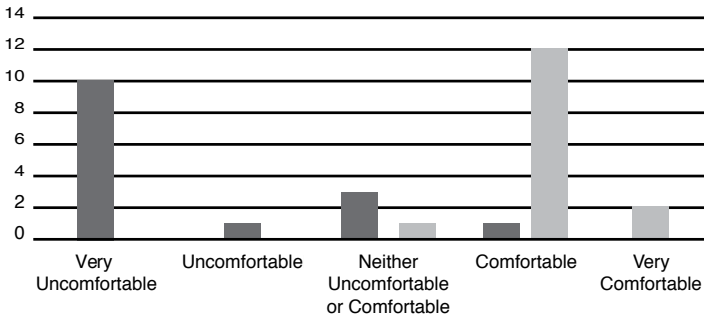

Figure 1. **Response to Q1: How comfortable do you feel performing UNHS?**

Figure 1 shows the majority of the subjects reported feeling uncomfortable prior to completing UNHS training. Conversely, after completing the training, most participants reported feeling *comfortable* with how to perform UNHS screenings. One way repeated ANOVA showed ($F [1, 12] = 39.76$, $p < 0.01$). This was statistically significant.

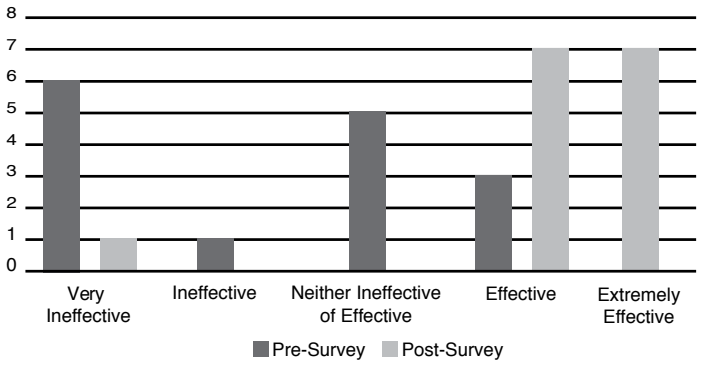

Figure 2. **Response to Q2: How effective do you believe your training concerning UNHS has been?**

Figure 2 indicates many participants did not believe their training concerning UNHS was *effective*. After completing the NCHAM module training 14 of 15 participants reported their training was effective or *extremely effective*. One way repeated ANOVA showed ($F [1, 12] = 14.52$, $p < 0.01$). This was statistically significant.

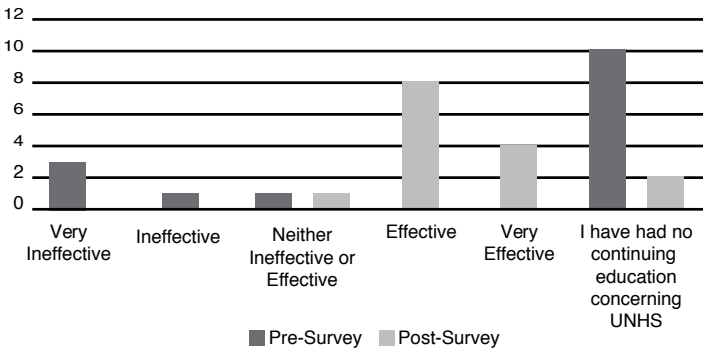

Figure 3. **Response to Q3: How effective do you believe your continuing education concerning UNHS has been?**

In Figure 3, pre-training, most participants reported they had no continuing education at all concerning UNHS. One way repeated ANOVA showed ($F [1, 12] = 2.69$, $p > 0.05$). This was not statistically significant even though most of the participants indicated that the continuing education was *effective* post-training.

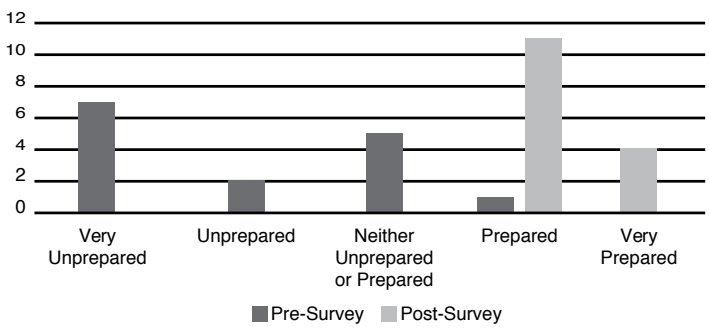

Figure 4. **Response to Q4: Do you feel your training has prepared you to complete UNHS using the most up-to-date methods?**

Figure 4 indicates that the majority of the participants did not feel their training had prepared them to complete UNHS using the most updated methods. Post-training survey responses showed significant differences. All participants felt they were either *prepared* or *very prepared*. One way repeated ANOVA showed ($F [1, 12] = 94.08$, $p < 0.00$). This was statistically significant.

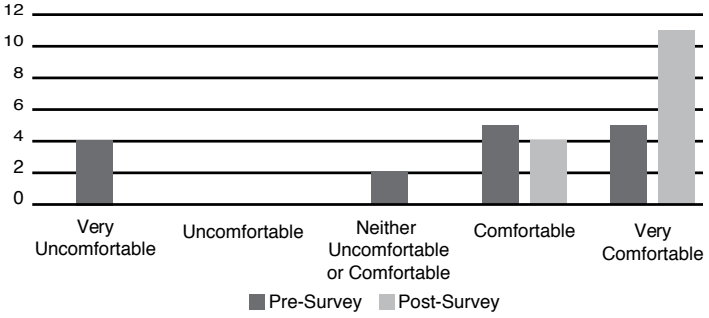

Figure 5. **Response to Q5: If you have a question concerning UNHS testing methods, how comfortable do you feel asking another professional?**

In Figure 5, the majority of participants were already comfortable asking another professional for help to complete UNHS testing if necessary. Post survey, all participants were either *comfortable* or *very comfortable*. One way repeated ANOVA showed ($F [1, 12] = 7.89$, $p < 0.05$). This was statistically significant.

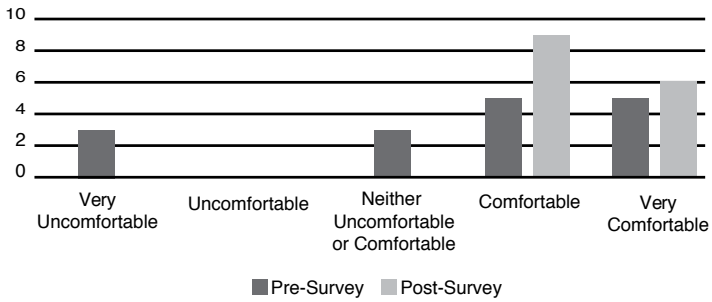

Figure 6. **Response to Q6: If you have a question concerning UNHS testing methods, and there is no other professional available at the time of your screening, how comfortable do you feel rescheduling the UNHS for another time?**

Pre-training responses, reported in Figure 6, showed that a majority of participants were already *comfortable* rescheduling UNHS for another time if they had a question concerning testing methods and another professional was not available at the time of the screening. Post-training survey responses showed that all participants were *comfortable* or *very comfortable*. One way repeated ANOVA showed (F [1, 12] = 2.18, $p > 0.05$). This was not statistically significant.

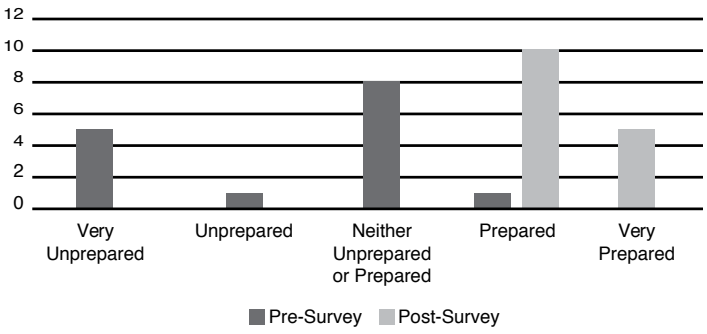

Figure 7. **Response to Q7: Do you feel your training has prepared you to complete UNHS using the most up-to-date equipment?**

Figure 7 indicates that a majority of the participants did not feel prepared to complete UNHS using the most updated equipment before they completed the training module. After completing the online training, all participants reported feeling *prepared* or *very prepared*. One way repeated ANOVA showed (F [1, 12] = 45.47, $p < 0.01$). This was statistically significant.

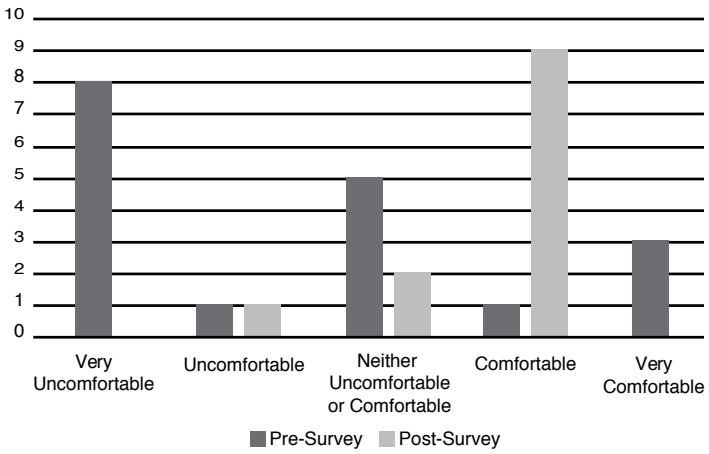

Figure 8. **Response to Q8: If you have trouble with the testing equipment, how comfortable are you performing troubleshooting?**

As shown in Figure 8, prior to completing the online training, the majority of test subjects reported feeling *very uncomfortable* with conducting troubleshooting if a problem occurs with the testing equipment. Post-training survey responses show the majority of subjects were *comfortable* completing troubleshooting. One way repeated ANOVA showed (F [1, 12] = 27.57, $p < 0.01$). This was statistically significant.

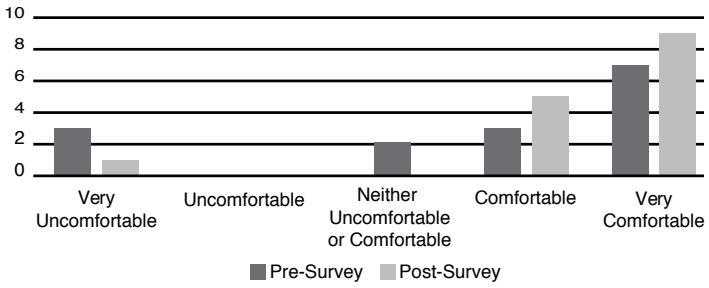

Figure 9. **Response to Q9: If you perform troubleshooting and it does not correct the problem you are experiencing with the testing equipment, how comfortable are you asking another professional for help?**

In Figure 9, the majority of participants indicated they were either *comfortable* or *very comfortable* asking another professional for help if performing troubleshooting did not correct an equipment problem that occurred during UNHS testing. The total number of participants who were either *comfortable* or *very comfortable* increased across post-survey responses. One way repeated ANOVA showed (F [1, 12] = 1.21, $p > 0.05$). This was not statistically significant.

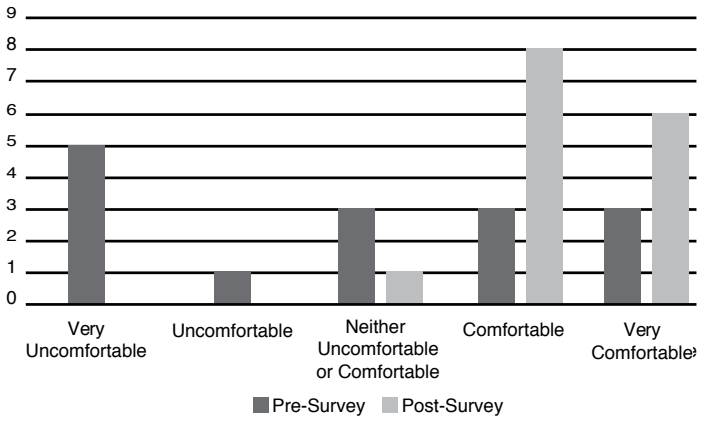

Figure 10. **Response to Q10: If the patient you are testing has a failing result, how comfortable do you feel documenting the result?**

Figure 10 shows that a majority of participants were not comfortable documenting the results of UNHS testing if the newborn has a failing result. After completing the online training module, most participants responded that they felt comfortable documenting this type of result. One way repeated ANOVA showed ($F [1, 27] = 5.85$, $p < 0.02$). This was statistically significant.

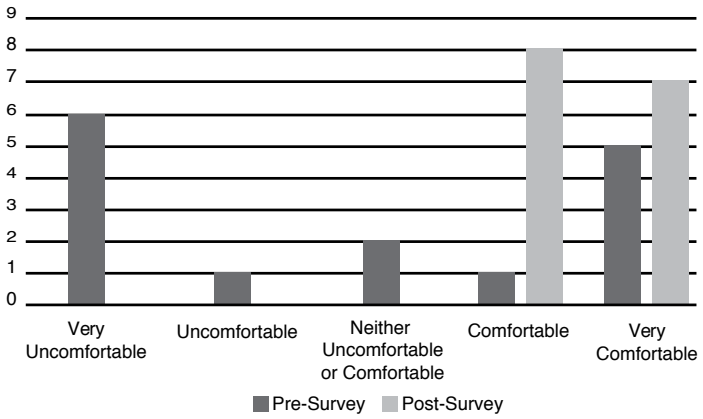

Figure 11. **Response to Q11: If the patient you are testing has a passing result, how comfortable do you feel documenting the result?**

In Figure 11, most subjects responded that they were very uncomfortable documenting a passing newborn hearing screening result. After training, post-survey responses show that all participants felt either comfortable or very comfortable with documenting a passing test result. One way repeated ANOVA showed ($F [1, 10] = 4.23$, $p > 0.05$). This was not statistically significant.

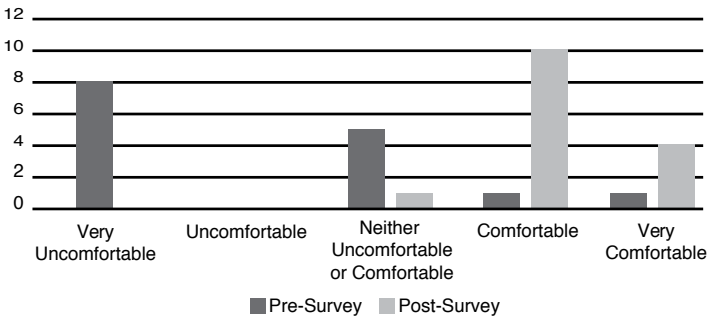

Figure 12. **Response to Q12: How comfortable do you feel interpreting the results of UNHS?**

Figure 12 indicates that most participants felt *very uncomfortable* interpreting the results of UNHS. Post-survey responses show that the majority of subjects felt *comfortable* interpreting the results of UNHS testing after completing the online training module. One way repeated ANOVA showed ($F [1, 11] = 25.63$, $p < 0.01$). This was statistically significant.

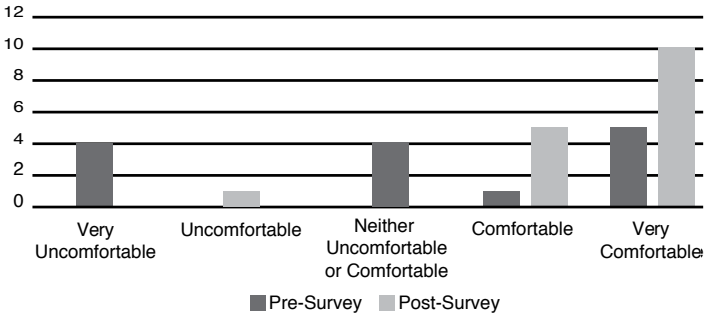

Figure 13. **Response to Q13: How comfortable do you feel relaying the results of UNHS to another professional?**

Concerning how comfortable participants felt relaying the results of UNHS to another professional, Figure 13 shows that there was a wide variety of responses prior to completing the online training. However, post-survey responses indicate that all subjects felt *comfortable* or *very comfortable* completing this task. One way repeated ANOVA showed ($F [1, 11] = 6.22$, $p < 0.05$). This was statistically significant.

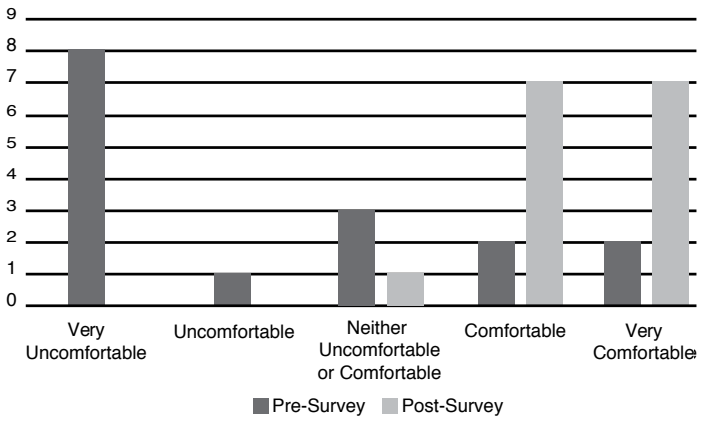

Figure 14. **Response to Q14: How comfortable do you feel counseling parents on the results on UNHS?**

As shown in Figure 14, when asked how comfortable they felt counseling parents on the results of UNHS, the majority of participants indicated that they felt *very uncomfortable*. Conversely, after completing the online training module, almost all participants indicated that they felt either comfortable or very comfortable completing this task. One way repeated ANOVA showed ($F [1, 11] = 18.75$, $p < 0.01$). This was statistically significant.

Participants completed both pre- and post-surveys as a subjective measure of their level of comfort and preparedness to complete different aspects of UNHS testing. The main goal of this study was to see if the nurse's knowledge and understanding improved following this online training program. In Figure 15, investigators compared the number of participants who responded that they were *comfortable* or *very comfortable* (or, a similar response such as *prepared* or *very prepared*) on each survey question asked both pre- and post-training.

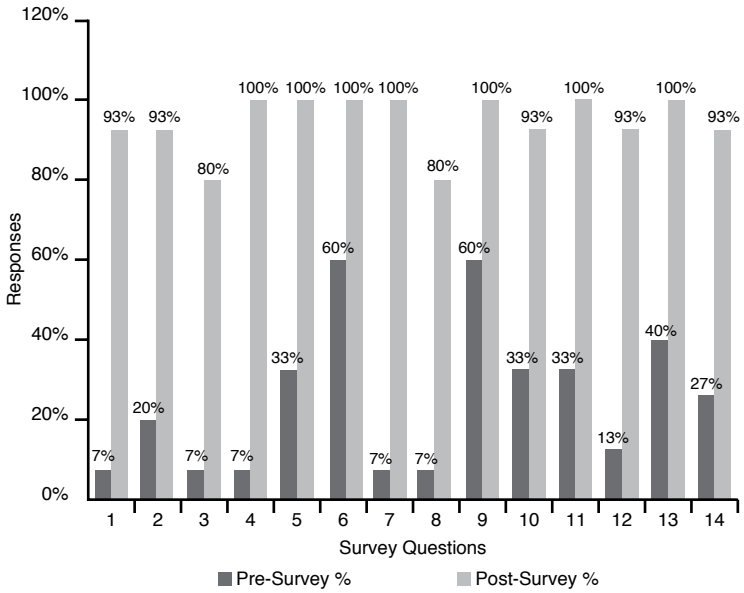

Figure 15. **Comparison of Pre- and Post-Survey Responses.**

Participants who responded *comfortable* or *very comfortable* (or, an equivalent answer) improved on all questions asked between pre- and post-survey questions.

Test Scores

Participants completed pre- and post-testing through the NCHAM online training module. Mean test scores for the pre-test was 81 with a standard deviation of 6 and the mean test scores for the post-test was 92 with a standard deviation of 6. Figure 16 shows the comparison of subjects' test scores prior to training and after completing the training module. A one-way repeated measures ANOVA was completed, and showed ($F [1, 14] = 33.27$, $p < 0.01$). This was statistically significant. Thirteen of fifteen participants made improvements to their test scores between pre- and post-training. Two subjects' test scores remained the same between measures.

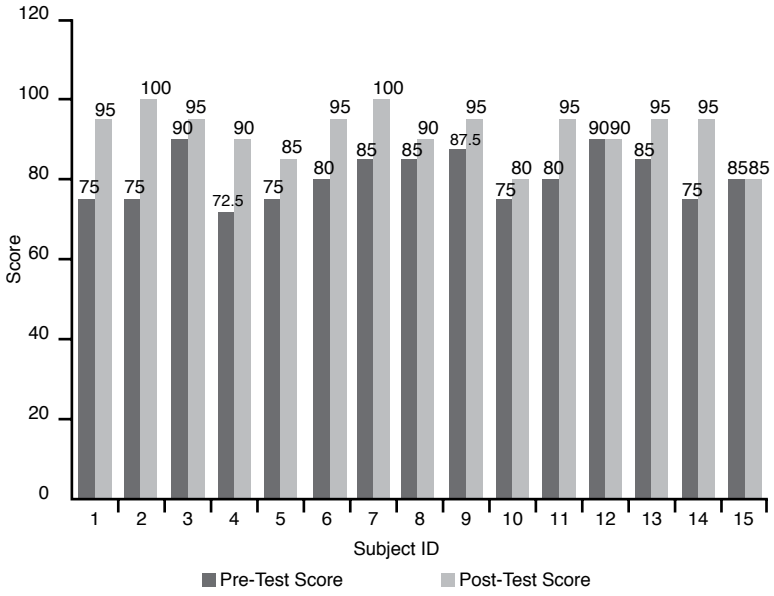

Figure 16. **Comparison of Pre- and Post-Test Scores.**

Discussion

In the present study, several aspects included in UNHS were evaluated to determine whether or not nursing professionals were currently comfortable with and prepared to perform UNHS. Participants initially reported that they were uncomfortable performing UNHS evaluations. As similarly reported (Parving & Saloman, 1996), they did not believe their previous training was effective, or that it prepared them to complete UNHS using the most up-to-date methods. After completing the online training, participants' perception of their comfort in completing UNHS was much improved. This subjective improvement, coupled with the improvement on the participants' pre- and post-training test scores, indicates that the online training program was effective in increasing the subject's perception of their ability to complete UNHS.

The results show that, as a whole, the participants do not feel comfortable asking other professionals for help concerning testing methods, troubleshooting the testing equipment, or documenting passing or failing results. Our findings agree with those of Ditty (2007) who found that communication between medical professionals and with patients' families is sometimes neglected. Further, participants did not feel they were prepared to interpret the results of UNHS, relay the information from the screening to another professional, or counsel parents concerning test results. After completing training, the majority of participants showed an improvement concerning their perception of their ability to ask other professionals for help and complete testing on their own. This was confirmed in 13 out of 15 participants who improved their test scores post-training. The remaining two participants who did not have improvement in post-training test scores had already achieved high scores on the pre-test measure.

When asked how effective their continuing education has been concerning UNHS, many participants reported that they have never had any additional training. That is concerning since everyone included in the project has a scope of practice that includes conducting UNHS and even training other professionals how to complete testing and record UNHS results. As similarly addressed in Pallarito's (2012) article, the importance of sufficient training and adequate documentation of screening information is of the utmost importance in positive outcomes. After completing this study, the majority of participants reported feeling their continuing education using NCHAM's Interactive Newborn Hearing Screening Curriculum was either *effective* or *extremely effective*. This shows that this method is sufficient to further train and equip professionals who are required to complete UNHS. Continuing education improves awareness of healthcare providers and improves their ability to perform the screenings (Gallagher et al. 2006; Houston et al., 2011).

Conclusions and Future Study

In general, the findings of this study suggest that nursing professionals do not feel they are adequately up-to-date with newborn hearing screenings and documenting screening results. Healthcare professionals are trusted with completing and documenting results that may affect how their patients develop and learn. It is imperative these professionals are adequately trained to manage the amount of responsibility entrusted to them. Objectively, study participants who completed this specific online training made improvements in their pre- and post-testing across both objective and subjective measures. This indicates that the training model used is an effective way to update professionals' current knowledge while expanding their overall understanding.

This model should be considered to adequately train professionals who complete UNHS in order to ensure timely diagnosis of infants and education of parents. Additional continuing education for professionals that complete UNHS should include the modules included in this training, such as how to complete hearing screens, what current equipment looks like, how to document and record results, and how to answer questions concerning screening, results, and further follow-up. Further, training for professionals working in the NICU nursery, should be expanded to cover more in-depth topics that may not be addressed by general training such as risk factors. Routine evaluation of healthcare professionals' understanding of such topics would underscore the importance of the measures and would serve to identify any areas for needed training.

Several of the participants stated that they received initial training several years prior to completing the training included in this project. Further, the majority of participants indicated that they had never received continuing education concerning UNHS even though this is included within their scope of practice. This is not an acceptable standard for health care professionals, especially concerning such an important measure of how a child will grow, develop, and learn. Professionals should be adequately prepared to complete the job they are certified and licensed to complete. In order to most effectively and efficiently serve patients and their families, UNHS training should continue to evolve and retraining should be completed on a scheduled basis.

The present study had limitations that could be improved to more adequately investigate current procedures to develop the best-suited training program. Most notably, the sample size of this research includes only 15 participants. Although results are promising concerning the effectiveness of this model, further research should focus on replicating this project on a larger scale to determine whether or not the outcomes apply to larger, more diverse populations as well. This project focused on nursing professionals. A future study including allied health professionals and non-nursing staff would be beneficial as well as including a hands-on module.

Finally, research should evaluate how including a hands-on training component, or lab, for participants would affect their perception and performance concerning overall preparedness to complete UNHS. Although the online training module provides both verbal and visual components to aid in participant's learning, it did not provide an actual person to ask questions of or demonstrate procedures directly. This experience would allow for more in-depth education for professionals and promote active training.

UNHS is one of many tests and screenings that newborns undergo when they are first born. Screening does not take a long time and is non-invasive. However,

for as quick and easy a test as hearing screening is, the outcome of the screening is vital. The need to effectively screen infants' hearing is crucial. Early identification of hearing loss allows for earlier referral to early interventions services and support systems for the child and their families. Conversely, missing a hearing loss or not identifying a loss until there are developmental delays eliminates the possible gains of early intervention. Healthcare professionals are tasked with serving patients and their families to the best of their abilities. Adequately training professionals and volunteers to complete hearing screening is necessary to provide important information to parents to assist them in appropriate and timely follow-up (DesGeorges, 2003).

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Some Permanent Hearing Loss is Missed When "Switched Ear" Passes are Combined for Determining Screening Results

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Abstract: In practice, some Newborn Hearing Screening (NHS) programs designate an infant as having a bilateral "pass" by using non-simultaneous results obtained in each ear during different screening sessions (referred to in this article as switched ears or SW_EAR). This study aims to obtain evidence for determining the screening outcome of infants with SW_EAR, using data from MEDNAX-Pediatrix's nationwide NHS program. From January 2009 to December 2012, infants with SW_EAR passes were referred for audiological evaluation. The audiological evaluations of 13,044 infants who referred (1,907 due to SW_EAR passes) out of the total infant population screened (2,212,107) were analyzed. Of the 2,816 infants identified with permanent hearing loss (PHL), 150 (5.3%) were from the group of infants with SW_EAR passes. Most of these infants (116/150, 77%) had bilateral PHL, with documented hearing aids in 89 infants and 7 infants who received cochlear implants. By not using SW_EAR passes (i.e., by not combining non-simultaneous ear passes from different screening sessions) to determine that the infant had passed the newborn screen, and instead referring those infants for audiological evaluation, a significant number of infants with PHL were identified, while maintaining program performance within benchmarks.

Key Words: Hearing screening, false-negative, infant, sensorineural hearing loss

Acronyms: AABR = automated auditory brainstem response; ANSD = auditory neuropathy spectrum disorder; ASHA = American Speech-Language-Hearing Association; CDC = Centers for Disease Control and Prevention; CHL = conductive hearing loss; FCHL = fluctuating conductive hearing loss; IP = inpatient; JCIH = Joint Committee on Infant Hearing; NHS = Newborn Hearing Screening; OAE = otoacoustic emissions; OP = outpatient; PCHL = permanent conductive hearing loss; PDX = MEDNEX-Pediatrix; PHL = permanent hearing loss; ROC = receiver operating characteristics; SNHL = sensorineural hearing loss; SW_EAR = non-simultaneous results obtained in each ear during different screening sessions

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Introduction

Over the last two decades Newborn Hearing Screening (NHS) programs have evolved and are being developed in many countries worldwide. As a result, the focus and challenges of NHS have shifted from implementing programs to creating more efficient and effective screening practices (Choo & Meinzen-Derr, 2010; Nelson, Bougatsos, & Nygren, 2008; White, Forsman, Eichwald, & Muñoz, 2010).

Major challenges for successful and cost-efficient screening practices include the need to maintain sufficiently low referral and false positive rates. The use of repeat screening tests/sessions with automated auditory brainstem response (AABR) and/or otoacoustic emissions (OAE) technology has proven to be a useful approach to achieve these goals (Clemens & Davis, 2001; Gravel et al., 2005; Vohr et al., 2001; White et al., 2005). A number of well-controlled studies have demonstrated that the commonly used two-step/two-technology NHS protocols can effectively reduce the overall referral rates to ≤ 4% of the total infants screened or even lower for those protocols that combine inpatient (IP) and outpatient (OP) screens (Thompson et al., 2001). However, while repeated automated screens help to enhance specificity by reducing false positive results (Clemens & Davis, 2001), they could take a toll on the protocol sensitivity and cause more

infants with hearing loss to pass the screen (an increase of false negatives) due to inherent problems of statistical artifacts associated with repeated testing (JCIH, 2007, p. 903).

It is important to systematically evaluate the various factors that could impact the overall performance of repeat testing NHS protocols. Recent studies by Turner (2013a, 2013b), using mathematical modeling and receiver operating characteristics (ROC) methodology, identified complex interactions between three basic factors that could enhance or reduce below optimum the overall performance of NHS protocols using repeat automated screens: (a) the accuracy of the screening test/technology in use, (b) the internal test correlation measuring the likelihood that repeated screens identify the same individuals as positive (refer) or negative (pass), and (c) the protocol's *stopping* criterion by which the results from repeat screens are combined to make the final pass/refer decision for the infant.

Past studies of NHS have focused on the overall efficiency/effectiveness of the most commonly implemented protocols (Gravel et al., 2005; White et al., 2005) or compared the accuracy of the screening method (OAE vs. AABR) in use (Norton et al., 2000). However, there is limited data available from NHS programs regarding the consequences of repeated automated screens or the criteria by which the results from multiple screens are used to determine the pass/refer outcome for an infant. As a result, there is a limited understanding of the impact these internal decisions may have on the overall performance of the screening protocol being used. This knowledge, when available, may provide a rational basis for further enhancements of NHS programs.

A key aspect in the implementation of NHS protocols, regardless of the technology being used, is how results from multiple automated screens are used to make a final pass/refer decision for an infant. The Expert Panel Recommendations on Newborn Hearing Screening of the American Speech-Language-Hearing Association (ASHA, 2013) state: "…. the infant must pass the screening in both ears to be considered a "pass". The Recommendations also state "… If the newborn fails one ear, both ears must be screened during the re-screening"… (ASHA, 2013).

In this context, the interpretation of passing results in both ears, which were obtained at a different test session or on a different day, poses an interesting problem that needs further investigation. There are no agreed-upon criteria for deciding how to use ear passes in both ears that were not obtained within the same test time or session.

Some Newborn Hearing Screening (NHS) programs combine a pass result obtained for the left ear during one test session with a pass result obtained for the right ear during a different test session to conclude that the infant has a bilateral pass for the hearing screen even though both ears did not pass during the same test session.

Unfortunately, it is not known how many infants with PHL may be missed by considering non-simultaneous passes obtained in each ear during repeated screens (referred to in this article as switched ears or SW_EAR passes) as a bilateral "pass." By this practice, an infant who passes only one ear (left or right) during a screen session and then during a repeat screen performed at a different test time or on a different day passes the ear that previously referred, would be given a "pass-pass" or screen negative outcome. However, there is no systematic research to whether the use of such SW_EAR passes may result in infants with permanent hearing loss being missed.

Method

Study Design

A retrospective cross-sectional study compared hearing screen results and audiological outcome data collected from January 2009 to December 2012 by MEDNAX-Pediatrix's nationwide NHS program (PDX_NHS) using AABR technology. Out of the total infant population screened (2,212,107), infants who received a refer status at discharge (13,044) were categorized into two groups: (a) those with SW_EAR passes (1,907) and (b) those without SW_EAR passes (11,137).

Inclusion/Exclusion Criteria. During the four years in which data were collected for this study, PDX_NHS programs referred all infants with SW_EAR passes for a complete audiological follow-up. The hearing screen data and audiological evaluations were categorized into two groups (with and without SW_EAR passes) as described below.

Infants with SW_EAR Passes. This group included those infants who had non-simultaneous passing results which were obtained in each ear during a repeat screen performed at a different time or on a different day. The "switching" between left ear and right ears passes could have occurred during any of the screens performed prior to discharge (inpatient) or when recalled as an outpatient. These infants would have been considered a "pass" (screen negative result) prior to this study. Figure 1 illustrates an example of an SW_EAR pass result for an infant with three repeat inpatient screens.

Infants without SW_EAR Passes. The group included those infants who failed one or both ears during the final AABR screen performed, prior to discharge as an inpatient (IP) and/or when recalled as an outpatient (OP), who had no "switching" between left and right ear passes during any combination of the screens or test sessions performed.

Participants. The study included all infants who received a refer status (13,044 in total; 1,907 with SW_EAR passes) during the four-year study period (2009–2012). The total number of infants screened by PDX_NHS programs during this time was 2,212,107, which represented 99.9% of all eligible births from 320 hospitals in 29 states.

Hearing Screen #1 (1/27/2011)
Right Ear: **PASS**
Left Ear: **REFER**
Decibel Level: 35

Hearing Screen #2 (1/27/2011)
Right Ear: **REFER**
Left Ear: **REFER**
Decibel Level: 35

Hearing Screen #3 (1/29/2011)
Right Ear: **REFER**
Left Ear: **PASS**
Decibel Level: 35

Figure 1. **A typical example of SW_EAR hearing screen results. Three screens were performed, each at a different time or test session. Conflicting passing results in each ear (SW_EAR) were obtained during screen #1 and screen #3. These non-simultaneous passes for the Left and Right ear would be combined as a Pass-Pass or screen negative outcome for the infant if SW_EAR pass results are allowed.**

Screening Protocol. The protocol combined IP and OP AABR hearing screens (when allowable per state specific guidelines) in most facilities. During the study period, the PDX_NHS screening protocol limited the number of AABR screens that could be performed for any infant to a maximum of three repeat screens during the IP stage (prior to discharge) and no more than two additional screens if recalled for OP testing session.

Equipment. All PDX_NHS programs used AABR as the method for screening with equipment manufactured, and approved for use, in the USA. However, the specific AABR testing device/model varied across hospitals from 2009–2011 and included ALGO® screeners (models ALGO 2E®, ALGO 2EC®, AlGO3®) and Bio-Logic ABaer® systems manufactured by Natus Medical Inc., as well as, Smart Screener-Plus 2® manufactured by Intelligent Hearing Systems. Specifications for each product are provided in the Hearing Review Products Technology Guides (2012) on the National Center for Hearing Assessment and Management (NCHAM) website. To facilitate program and operational standardization, a conversion to a single manufacturer of automated screening devices (Intelligent Hearing Systems, Smart Screener-Plus 2®) was initiated beginning in 2010 and completed by the end of 2011.

Data Collection. Demographic information of all infants screened by PDX_NHS program during the study, each infant's screening results, audiological evaluations, and information about use of hearing technology for all infants who were referred from the hearing screening in each group (with and without SW_EAR passes) was maintained in a web-based tracking and database management system (Soundata®). Referred infants who failed the audiological testing were followed for two years to capture as much diagnostic and hearing technology data as possible.

Data Audit. The diagnoses/outcomes data maintained in Soundata® for the infants who were referred at follow-up were audited independently by two authors to validate the audiological evaluations data used in this study. The authors specifically focused on the manual entry of the results from different Audiology/ENT reports. Different queries were posed to cross-check the data for inconsistencies in the results and/or inconclusive outcomes/diagnoses. Any detected cases were reviewed and corrected prior to final data analysis. Since a separate diagnostic category for fluctuating or temporary hearing loss due to middle ear pathology was not available for data categorization during the initial stage of the study, a full case-by-case review was conducted of all referred infants categorized as conductive hearing loss and/or middle ear disorder(s). Lastly, all cases in the SW_EAR group with a diagnosis of PHL, as well as those documented as receiving hearing technology (e.g., cochlear implants and/or hearing aids) were reviewed case-by-case and updated/corrected as needed.

Data Analysis. Upon completion of the data auditing process, the audiological outcome data of all referred infants in the study sample (with and without SW_EAR passes) were analyzed. Data analyses included descriptive and nonparametric statistics.

Audiological outcomes data. Infants who were referred during screening (with and without SW_EAR) were categorized as follows:

Permanent Hearing Loss (PHL). Included infants with unilateral or bilateral hearing loss of any of the following diagnosis/types: Sensorineural hearing losses (SNHL); Auditory Neuropathy Spectrum Disorder (ANSD); Permanent Conductive hearing loss (PCHL), and mixed hearing loss.

Fluctuating Conductive Hearing Losses (FCHL). Included infants whose only hearing loss was attributable to temporary or fluctuating unilateral or bilateral conductive hearing loss, due to middle ear pathology which was evidenced through repeat audiological testing and/or following medical intervention (e.g., pressure equalization tubes or medical treatment).

Inconclusive Diagnosis. Included infants who failed follow-up testing with abnormal diagnostic tests and/or rescreen results, but had insufficient data to reach a definitive audiological diagnosis (i.e., type and/or degree of hearing loss in each ear).

No Hearing Loss. Included all infants who passed the follow-up audiological testing in both ears. Passing results could be obtained with either automated screening tests alone (e.g., OAE, AABR) or were produced via a complete or incomplete diagnostic test battery (e.g., diagnostic ABR

thresholds and/or behavioral testing) as well as other audiological tests.

Program Performance Metrics. Appropriate actions for SW_EAR results within PDX_NHS program were determined by calculating the following metrics of the program during the study period and expressed as percentages:

Referral Rates: number of refers in each group divided by the number of infants screened

Permanent Hearing Loss Rate among Referrals: number of infants who had a definitive diagnosis of PHL (e.g. sensorineural, conductive, mixed and ANSD) unilateral or bilateral in each group, divided by the number of infants referred for audiological follow-up

No Hearing Loss Rate among Referrals: number of refer infants who had passing results during the audiological follow-up testing and no temporary hearing loss evidenced during the follow-up period in each group, divided by the total infants who were referred for audiological follow-up

Diagnosed PHL: number of infants who had a definitive diagnosis of PHL in each group (e.g., SNHL, PCHL, mixed, and ANSD) unilateral or bilateral, divided by the total number of infants screened

False Positives: number of infants determined to have normal hearing who failed the hearing screening, divided by the number of infants screened

Results

Figure 2 shows the results for hearing screen data and audiological diagnostic evaluation of infants who received a refer status in both groups: (a) without SW_EAR passes and (b) with SW_EAR passes. Seventy-seven percent of the infants who referred in each group during the period of the study (2009–2012) were successfully tracked and had Audiology/ENT reports in Soundata®.

Most of the infants who were successfully tracked had sufficient follow-up data (e.g., Audiology/ENT reports, test results, and/or information about use of hearing technology) for a definitive diagnosis and could be categorized as either: (a) PHL (including SNHL, PCHL, mixed or ANSD), (b) FCHL due to transient or chronic middle ear pathology, or (c) no hearing loss. However, there were a few infants who failed the initial diagnostic testing but had insufficient follow-up data for determining the nature of the hearing loss and were therefore categorized as inconclusive and omitted from further analysis (4.7% of those with SW_EAR passes).

Of the 1,907 infants in the group of infants with SW_EAR passes (Figure 2), 150 infants (7.9%) were diagnosed with PHL including SNHL, ANSD, PCHL, or mixed hearing loss. Note that the infants with SW_EAR passes constituted 14.6% (1907/13,044) of the total infants referred for audiological follow-up. The infants with SW_EAR passes who were diagnosed with PHL represented 5.3% (150/2,816) of those diagnosed with PHL in the population of 2,212,107 infants that were screened. Interestingly, in the group of infants with SW_EAR passes (1,907 infants) the proportion of PHL identified (150/1907, 7.9%) was higher than the PHL diagnosed in those infants screened who had no SW_EAR passes (2,666/2,210,200, 0.12%).

To further validate the audiological diagnosis of PHL, data were analyzed for the 183 infants in the group with SW_EAR passes who had documented use of hearing technology in Soundata® during the follow-up period. There were 89 infants diagnosed with PHL in the group with SW_EAR passes who were fit with hearing aids and 7 infants (6 SNHL, 1 ANSD) who received cochlear implants.

The type and degree of hearing loss was reviewed for each of the infants diagnosed with PHL in the group with SW_EAR passes. Three quarters of these had both ears affected (116/150, 77.3%). The severity of PHL for the total number of ears affected ($N = 266$ ears) is shown in Figure 3.

Note that about half of the infants' ears with PHL (52.6%, 140/266) had moderate-to-severe or severe-to-profound hearing loss. Also, 61% (42/70 ears) of the total ears which were classified as mild or mild-to-moderate PHL were fit with hearing aids. The frequency distribution by type of hearing loss diagnosed in both groups (with and without SW_EAR passes) is shown in Figure 4.

Fluctuating conductive hearing loss due to temporary and/or chronic middle ear disorders was more frequently diagnosed in the group with SW_EAR passes than in the group without SW_EAR passes (Chi square = 71.65; $p < 0.000$). Also, the proportion of PCHL was lower in the group with SW_EAR passes compared to the group without SW_EAR passes (Chi square = 16.59; $p < 0.000$). Given that PDX_NHS policies stipulated that infants with ear atresia should not be screened, but should be referred directly for audiological follow-up, PCHL secondary to ear atresia/microtia was not represented in the group with SW_EAR passes. The remaining types of PHL showed similar frequency distributions for refer infants with and without SW_EAR passes (SNHL: Chi Square = 3.19; $p < 0.07$; ANSD: Chi Square = 1.56; $p < 0.21$; Mixed: Chi Square = 1.10; $p < 0.29$).

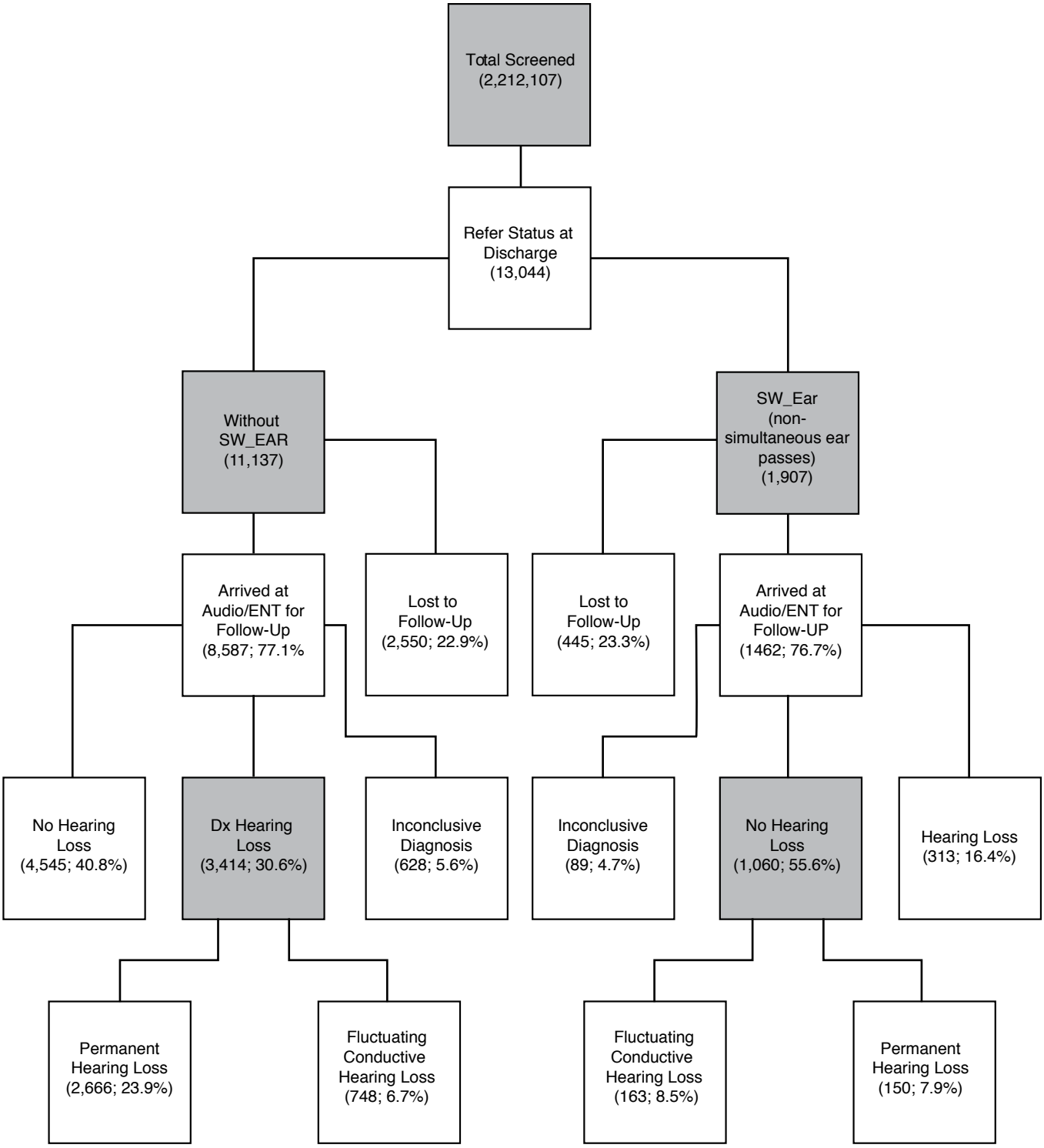

Figure 2. **Flowchart summarizing the hearing screen data and audiological diagnostic outcomes of the infants who received a refer status in both groups: (a) without SW_EAR and (b) with SW_EAR passes. Lost to Follow-up category includes all refer infants that were lost (no audiological follow-up) including those unsuccessfully tracked as well as the parents/physician refusals, ineligibles due to medical constraints, etc.**

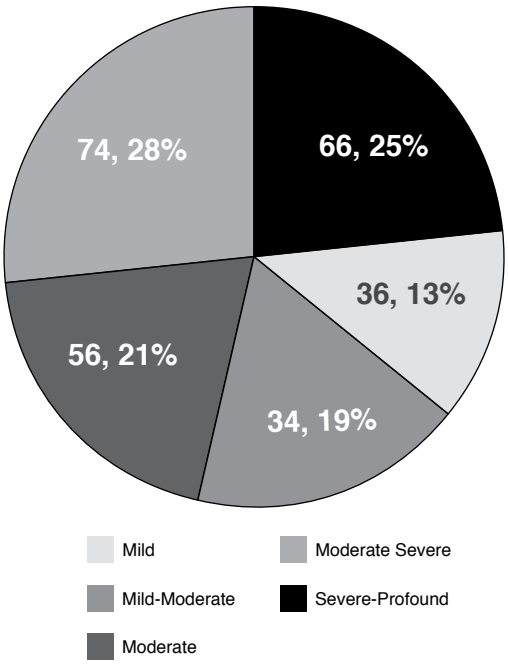

Figure 3. **Severity of permanent hearing loss (PHL) diagnosed in infants with SW_EAR passes. (Total number of ears affected (N = 266).**

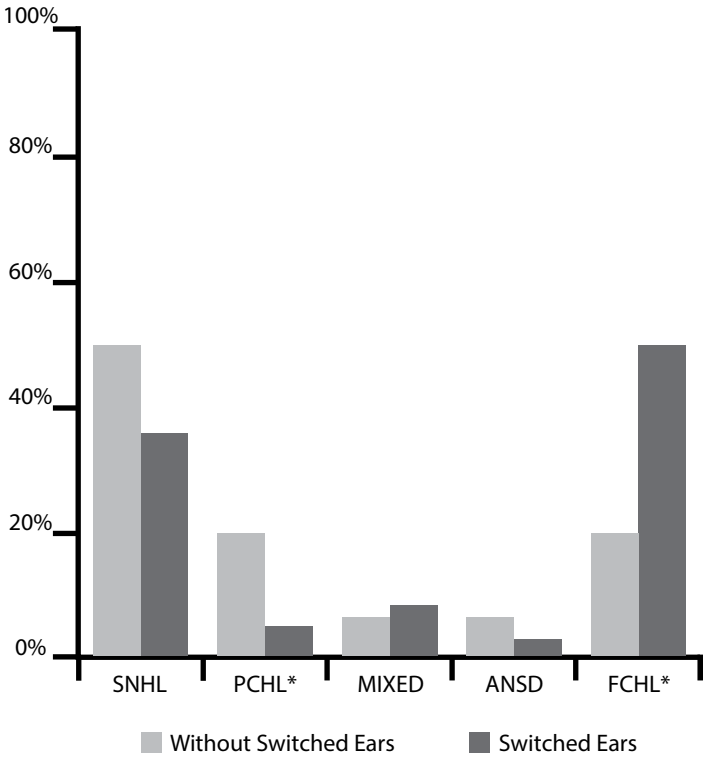

Figure 4. **Type of permanent hearing loss (PHL) identified in infants with and without non-simultaneous passes obtained in each ear during different screening sessions (SW_EAR).**

Note. ANSD = auditory neuropathy spectrum disorder; FCHL = fluctuating conductive hearing loss; MIXED = mixed hearing loss; PCHL = permanent conductive hearing loss; SNHL = sensorineural hearing loss.

*** Indicates statistical significance (p < 0.05) between infants referred with SW_EAR and without SW_EAR.**

Finally, selected metrics of program performance were analyzed for the group of infants without SW_EAR passes and for all infants in the sample (see Table 1). Note that the addition of the group with SW_EAR passes allowed the identification of an average of one more infant with PHL in every hundred with positive screening results (PHL rates increased from 21.7% for infants without SW_EAR to 22.9% when infants with SW_EAR passes were referred for audiological follow-up). Also, there was a slight increase in the referral rates (from 0.50% to 0.58%, as well as in the proportion of infants with no HL (from 34.8% to 42.9%).

Discussion

The need for systematic evaluation and monitoring of NHS program performance has been recognized as an important area for clinical research (White, Forsman, Eichwald, & Muñoz, 2010, ASHA, 2013). This population-based study, conducted within the context of PDX_NHS nationwide program provides convincing evidence that a significant number of infants with permanent hearing loss will be missed if infants with SW_EAR passes are not referred for audiological evaluation. Furthermore, results constitute strong empirical support for the current NHS recommendations (ASHA, 2013) that both ears must pass the screening for an infant to be screened negative. In addition, it supports clarification of the recommendation that both ear passes must be obtained during the same test time or during the same session.

Evidence from the retrospective analysis of diagnostic audiological evaluations collected during a four-year period (2009–2012) for 13,044 referred infants (of which 1,907 infants had SW_EAR passes) out of the

Table 1

Impact of referring infants with SW_EAR passes on the performance of PDX_NHS program

Screening Metrics	Infants without SW_EAR passes (n = 2,210,200)	All infants in sample (N = 2,212,107)
Referral Rates	0.50 [0.44-0.55]	0.58 [0.50-0.65]
No HL Rate	33.8 [31.8-35.9]	41.2 [37.8-44.6]
PHL Rate	21.7 [19.5-23.7]	22.9 [20.7-25.1]
False Positive	0.20 [0.16-0.23]	0.24 [0.2-0.3]
Permanent HL	0.12 [0.11-0.12]	0.13 [0.12-0.14]

Note. Data based on quarterly estimates for each metric calculated across sites (320 hospitals in 29 states) during the four years study (2009–2012). HL = hearing loss; PHL = permanent hearing loss; PDX_NHS = MEDNEX-Pediatrix newborn hearing screening; SW_EAR passes = non-simultaneous passes obtained in each ear during different screening sessions.

total population screened (2,212,107) showed that by completing the audiological follow-up on infants with SW_EAR passes, PDX_NHS program identified one more infant with PHL in every hundred infants who were referred from the newborn hearing screening program. The infants in group of SW_EAR passes who were diagnosed at follow-up with permanent hearing loss (150/1,907, 7.3%) represent 5.3% of all infants identified with PHL in this sample of 2,212,107 infants who were screened. It is also important to note that the program maintained very low referral rates (0.58%) even though additional infants were being referred.

The hearing loss diagnosed in the group of children with SW_EAR passes should be further analyzed. Most of these infants diagnosed with PHL (116/150, 77%) had bilateral hearing losses and about half of these infants' ears (52%, 140/266) had moderate-to-severe or severe-to-profound hearing losses. In addition, a high proportion of infants with SW_EAR passes (136/1,907, 8.5%) were diagnosed with fluctuating conductive hearing losses (FCHL) due to middle ear effusion. This type of dysfunction could "switch" from one ear to the other, and be reflected in non-simultaneous ear passing results at different test times. An elevated incidence of temporary middle ear dysfunction in the neonatal period associated with the development of middle ear pathology has been well documented by many authors (Doyle, Kong, Strobel, Dallaire, & Ray, 2004; Doyle, Rodgers, Fujikawa, & Newman, 2000). The fact that the relative proportion of FCHL was significantly higher in the infants with SW_EAR passes compared to those without SW_EAR passes is consistent with the hypothesis that middle ear pathology may be a plausible explanation for part of the hearing loss diagnosed in the SW_EAR group.

Another possible explanation for the hearing losses identified in infants with SW_EAR passes that should be analyzed is the problems associated with the use of automated screening technology. There are many operational factors that may affect screening results (e.g., accuracy of earphone placement, artifacts due to baby movement, environmental noise, etc.) as well as technical issues (e.g., problems of repeat screening attempts and lack of standardization of automated screening technology). Although these issues have been mentioned in the literature (JCIH, 2007; ASHA, 2013), they have not been adequately explored.

One limitation of this study is that different types of AABR equipment were used during the study. Given that each type of equipment/manufacturer uses different algorithms for determining the pass/refer decision in any single screen performed, the likelihood of an infant having SW_EAR passes might vary for the different devices. This possibility needs to be explored with all types of AABR manufactured equipment. Also, the possibility that similar results would be obtained with OAE equipment needs to be investigated.

Another limitation of the study is the number of repeat automated screens that were performed for determining the final outcome for an infant. During this period of the study, up to five screens (3 IP + 2 OP) were allowed. As more screens are performed, the statistical problems of sequential testing (Stürzebecher, Cebulla, & Elberling, 2005; Stürzebecher & Cebulla, 2013) may increase the probability of falsely passing PHL, but this needs to be investigated.

The implications of this study for clinical practice are important. Current best practice guidelines state that both ears should pass for an infant to pass the screen (ASHA, 2013). Also, the recommendations state that both ears must be tested during re-screening. The empirical data provided in this study supports the above recommendations and indicates that both ears should pass within the same screening session for an infant to be considered a pass (screen negative outcome). The fact that PHL, mostly bilateral and of significant magnitude, was diagnosed in this group, suggests that infants with non-simultaneous ear passes should be referred and tracked for audiological follow-up with the same urgency as repeat "non-switching" unilateral or bilateral refers.

The results of this study also demand refocused attention on how parents are counseled regarding SW_EAR results by screeners, pediatricians, and audiologists. Providers should not suggest that because a pass result was obtained for both ears, albeit at different times, that the diagnostic evaluation is likely to result in a conclusion that the infant has normal hearing. Indeed only 55.6% of the 1,907 infants who had SW_EAR passes were determined to have normal hearing, with the remainder being diagnosed with PHL (7.9%) or conductive hearing loss (CHL; 8.5%), having inconclusive results (4.7%), or not returning for the audiological evaluation (23.3%). These data suggest that if providers minimize the importance of parents completing diagnostic follow-up testing, there is a real possibility of missing infants with permanent and conductive hearing loss with a consequential detrimental effect for infant development (Yoshinaga-Itano, Coulter, & Thomson, 2001).

Conclusion

This retrospective study of 2,212,107 screened infants, 1,907 of whom had SW_EAR passes, provides evidence for eliminating the practice of passing infants by combining "switched ear" passes from repeat screens and therefore missing potential permanent hearing loss. Results support the current ASHA best practice recommendation which requires both ears to pass the screening for an infant to be screened negative with the added specification that both ear passes should be obtained within the same screening session. Furthermore, all hearing health care providers

involved in clinical follow-up care of refer infants should be cautious about concluding that an infant has normal hearing based on non-simultaneous passes on each ear from repeat screens.

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JEHDI
The Journal of Early Hearing Detection and Intervention
NCHAM National Center for Hearing Assessment and Management Utah State University
2017; 2(2): 56–77

Others' Publications About EHDI: May through October 2017

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

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

It is interesting to note that of the 80 articles published in other journals from May through October 2017, 59 articles (74%) reported on research conducted in countries other than the United States. Clearly, EHDI has become a global phenomenon and keeping up with EHDI requires staying informed about what is happening in other countries. As an indicator of what is trending in the literature:

- 23 articles discuss issues related to cochlear implants.
- 11 articles explore issues related to the genetics of hearing loss.
- 8 articles discuss the identification or treatment of cytomegalovirus (CMV) and its relation to childhood hearing loss.
- 7 articles report on issues related to protocols and procedures used in newborn hearing screening.

Noted below are just some of the interesting findings from around the world.

- Chung et. al surveyed 1024 audiology facilities in the United States and found that more than 90% reported results of diagnostic evaluations following newborn hearing screening to their state EHDI program.
- Fitzpatrick et. al reported that 16.5% of children identified with hearing loss in their sample from Canada experienced more than 3 months delay from the initial audiologic assessment to confirmation of their hearing disorder.
- Martinez-Cruz et. al found that 40% of the Mexican infants from neonatal intensive care units with sensorineural hearing loss had progressive hearing loss.
- Netten et. al reported that children from the Netherlands and Belgium who were identified early and received cochlear implants had higher early language skills, which served as a protective factor against the development of disruptive behavior.
- Sanyelbhaa et. al found a 76% increased risk for sensorineural hearing loss among children in Egypt who were progeny of consanguineous marriages compared to similar children from non-consanguineous marriages.
- Guerzoni and Cuda used data logging from cochlear implant speech processors to show that the amount of time the cochlear implant was worn positively predicted early linguistic and auditory outcomes.

Listed below are many more articles with interesting and relevant findings that can be used to improve EHDI programs wherever you live.

Abdul Majid AH, Zakaria MN, Abdullah NAC, Hamzah S, Mukari SZS.
Determinants of caregivers' awareness of Universal Newborn Hearing Screening in Malaysia.
Int J Pediatr Otorhinolaryngol. 2017 Oct;101:107-111. doi: 10.1016/j.ijporl.2017.07.036. Epub 2017 Jul 27.

OBJECTIVE: This paper aims to investigate the effects of perceived attitude and anxiety on awareness of UNHS among caregivers in Malaysia.
METHODS: Using cross sectional research approach, data were collected and some 46 out of 87 questionnaires distributed to caregivers attending UNHS programs at selected public hospitals were usable for analysis (response rate of 52.8%). Partial Least Squares Method (PLS) algorithm and bootstrapping technique were employed to test the hypotheses of the study.
RESULTS: R square value is 0.205, and it implies that exogenous latent variables explained 21% of the variance of the endogenous latent variable. This value indicates moderate and acceptable level of R-squared values. Findings from PLS structural model evaluation revealed that anxiety has no significant influence ($\beta = -0.091$, $t = 0.753$, $p > 0.10$) on caregivers' awareness; but perceived attitude has significant effect ($\beta = -0.444$, $t = 3.434$, $p < 0.01$) on caregivers' awareness.
CONCLUSION: Caregivers' awareness of UNHS is influenced by their perceived attitude while anxiety is not associated with caregivers' awareness. This implies that caregivers may not believe in early detection of hearing impairment in children, thinking that their babies are too young to be tested

for hearing loss. Moreover, socio-economic situation of the caregivers may have contributed to their failure to honor UNHS screening appointments as some of them may need to work to earn a living while some may perceive it a waste of time honoring such appointments. Non-significant relationship between anxiety and caregivers' awareness may be due to religious beliefs of caregivers. Limitations and suggestions were discussed.

Aimoni C, Ciorba A, Cerritelli L, Ceruti S, Skarżyński PH, Hatzopoulos S.

Enlarged vestibular aqueduct: Audiological and genetical features in children and adolescents.

Int J Pediatr Otorhinolaryngol. 2017 Oct;101:254-258. doi: 10.1016/j.ijporl.2017.07.042. Epub 2017 Jul 29.

BACKGROUND: Enlarged Vestibular Aqueduct (EVA) is one of the most common congenital malformations associated with sensorineural or mixed hearing loss. The association between hearing loss and EVA is described in syndromic (i.e. Pendred Syndrome, BOR, Waardenburg) and non-syndromic disorders, as isolate or familiar mutations of the SLC26A4 gene. The audiological phenotype of the EVA syndrome is heterogeneous, the type and entity of hearing loss may vary and vertigo episodes might also be present.
OBJECTIVE: The aim of this retrospective study was to describe the clinical and genetic features of a group of adolescent subjects presenting an EVA clinical profile, considering the presence of SLC26A4 gene mutations.
METHODS: 14 Caucasian patients were assessed (24 ears in total; 4 patients presented a monolateral EVA), 10 females and 4 males. Their age at the time of diagnosis was between 1 and 6 years (mean age 2.5 years). Subjects were assessed by an ENT microscopy evaluation with a complete audiometric assessment, CT & MRI scans and genetic tests for the evaluation of the pendrin gene mutations (SLC26A4).
RESULTS: Considering the presence of SLC26A4 mutations and thyroid function, we could identify three sub-groups of patients: group 1, non syndromic EVA (ns EVA, no SLC26A4 mutation and no thyroid dysfunction); group 2, EVA with DFNB4 (single SLC26A4 gene mutation and no thyroid dysfunction); group 3, EVA with Pendred Syndrome (two pathological mutation of SLC26A4 and thyromegaly with thyroid dysfunction). Patients of group 1 (ns-EVA) showed various degrees of hearing loss from mild (55%) to severe-profound (45%). In groups 2 (DFNB4) and 3 (PDS), the degree of hearing loss is severe to profound in 70-75% of the cases; middle and high frequencies are mainly involved.
CONCLUSIONS: The phenotypic expressions associated with the EVA clinical profile are heterogeneous. From the available data, it was not possible to identify a representative audiological profile, in any of the three sub-groups. The data suggest that: (i) a later onset of hearing loss is usually related to EVA, in absence of SLC26A4 gene mutations; and (ii) hearing loss is more severe in patients with SLC26A4 gene mutations (groups 2 and 3 of this study).

Al-Sayed AA, AlSanosi A.

Cochlear implants in children: A cross-sectional investigation on the influence of geographic location in Saudi Arabia

J Family Community Med. 2017 May-Aug;24(2):118-121. doi: 10.4103/jfcm.JFCM_142_15.

INTRODUCTION: The role of the family in detecting a child's hearing difficulty and the age at which an implantation is done have been identified as strong predictors of the outcomes of pediatric cochlear implantation. In the absence of screening programs for hearing loss in Saudi neonates, the family's role is of paramount importance. The aim of this study was to investigate the influence of geographic location on the course of identification, examination, and cochlear implantation in children in Saudi Arabia.
MATERIALS AND METHODS: Pediatric patients who had received either unilateral orbilateral cochlear implantation at King Abdulaziz University Hospital in Riyadh, Saudi Arabia, between January 1, 2012, and December 31, 2014, were surveyed.
RESULTS: A total of 156 pediatric patients have had a cochlear implant between January 1, 2012, and December 31, 2014. The one-way analysis of variance test to compare the means of the independent sample groups in various geographic zones showed that with a hundred percent access to primary health care, the geographic location of the population had an influence on the detection of hearing loss but not on the cochlear implantation.
CONCLUSION: This study found that the geographic location of the population has an influence on the time of detection of hearing loss in children but not on the time of cochlear implantation. Raising parental awareness of the importance of early detection of hearing loss is necessary. Further research is also required to define the role of factors such as the income and the educational level of parents on the early detection of neonatal hearing loss.

Alkowari MK, Vozzi D, Bhagat S, Krishnamoorthy N, Morgan A, Hayder Y, Logendra B, Najjar N, Gandin I, Gasparini P, Badii R, Girotto G, Abdulhadi K.

Targeted sequencing identifies novel variants involved in autosomal recessive hereditary hearing loss in Qatari families.

Mutat Res. 2017 Aug;800-802:29-36. doi: 10.1016/j.mrfmmm.2017.05.001. Epub 2017 May 4.

Hereditary hearing loss is characterized by a very high genetic heterogeneity. In the Qatari population the role of GJB2, the worldwide HHL major player, seems to be quite limited compared to Caucasian populations. In this study we analysed 18 Qatari families affected by non-syndromic hearing loss using a targeted sequencing approach that allowed us to analyse 81 genes simultaneously. Thanks to this approach, 50% of these families (9 out of 18) resulted positive for the presence of likely causative alleles in 6 different genes: CDH23, MYO6, GJB6, OTOF, TMC1 and OTOA. In particular, 4 novel alleles were detected while the remaining ones were already described to be associated to HHL in other ethnic groups. Molecular modelling has been used to further investigate the role of novel alleles identified in CDH23 and TMC1 genes demonstrating their crucial role in Ca2+ binding and therefore possible functional role in proteins. Present study showed that an accurate molecular diagnosis based on next generation sequencing technologies might largely improve molecular diagnostics outcome leading to benefits for both genetic counseling and definition of recurrence risk.

Amin SB, Saluja S, Saili A, Orlando M, Wang H, Laroia N, Agarwal A.

Chronic auditory toxicity in late preterm and term infants with significant hyperbilirubinemia.

Pediatrics. 2017 Oct;140(4). pii: e20164009. doi: 10.1542/peds.2016-4009.

BACKGROUND AND OBJECTIVES: Significant hyperbilirubinemia (SHB) may cause chronic auditory toxicity (auditory neuropathy spectrum disorder and/or sensorineural hearing loss); however, total serum bilirubin (TSB) does not discriminate neonates at risk for auditory toxicity. Our objective was to compare TSB, bilirubin albumin molar ratio (BAMR), and unbound bilirubin (UB) for their association with chronic auditory toxicity in neonates with SHB (TSB ≥20 mg/dL or TSB that met criteria for exchange transfusion).
METHODS: Infants ≥34 weeks' gestational age (GA) with SHB during the first 2 postnatal weeks were eligible for a prospective longitudinal study in India. Comprehensive auditory evaluations were performed at 2 to 3 months of age by using auditory brainstem response, tympanometry, and an otoacoustic emission test and at 9 to 12 months of age by using audiometry. The evaluations were performed by an audiologist unaware of the degree of jaundice.
RESULTS: A total of 93 out of 100 infants (mean GA of 37.4 weeks; 55 boys, 38 girls) who were enrolled with SHB were evaluated for auditory toxicity. Of those, 12 infants (13%) had auditory toxicity. On regression analysis controlling for covariates, peak UB (but not peak TSB or peak BAMR), was

associated with auditory toxicity (odds ratio 2.41; 95% confidence interval: 1.43-4.07; P = .001). There was significant difference in the area under the receiver operating characteristic curves between UB (0.866), TSB (0.775), and BAMR (0.724) for auditory toxicity (P = .03) after controlling for covariates.
CONCLUSIONS: Unconjugated hyperbilirubinemia indexed by UB (but not TSB or BAMR) is associated with chronic auditory toxicity in infants ≥34 weeks' GA with SHB.

Ari-Even Roth D, Lubin D, Kuint J, Teperberg-Oikawa M, Mendelson E, Strauss T, Barkai G.
Contribution of targeted saliva screening for congenital CMV-related hearing loss in newborns who fail hearing screening.
Arch Dis Child Fetal Neonatal Ed. 2017 Nov;102(6):F519-F524. doi: 10.1136/archdischild-2016-311859. Epub 2017 May 3.
BACKGROUND: We previously reported a 2.2% rate of infants born with sensorineural hearing loss (SNHL) due to congenital cytomegalovirus (cCMV) infection identified by universal neonatal screen for cCMV using saliva.
OBJECTIVE: To evaluate the contribution of targeted saliva screening for cCMV to the detection of infants born with cCMV-related SNHL who failed universal newborn hearing screening (UNHS).
METHODS: We retrospectively reviewed the audiological and medical records of infants who failed UNHS and were tested for cCMV using saliva sample prior to discharge at Sheba Medical Center between 2014 and 2015. Positive cases were confirmed by urine sample.
RESULTS: Two hundred (1%) of the 19 830 infants tested during the study period failed in-hospital hearing screening. A saliva specimen was obtained prior to discharge in 187 infants (93.5% of those who failed UNHS). In 178 infants saliva testing was performed at ≤21 days of chronological age and yielded results. cCMV nfection was identified in 4/178 tested infants (2.25%, 95% CI 0.8% to 5.3%), of whom three were diagnosed with SNHL (1.7%, 95% CI 0.5% to 4.4%) and offered antiviral treatment. Two of the tested infants (1.12%, 95% CI 0.2% to 3.6%) were diagnosed with cCMV solely due to failure in UNHS. Occult central nervous system (CNS) symptoms of cCMV infection were detected in 2/4 infants following targeted investigation.
CONCLUSIONS: Targeted cCMV screening in newborns who failed UNHS contributed to the early detection of infants born with cCMV-related isolated SNHL or with occult CNS symptoms who could potentially benefit from antiviral treatment.

Bernardi GF, Pires CTF, Oliveira NP, Nisihara R.
Prevalence of pressure equalization tube placement and hearing loss in children with Down Syndrome.
Int J Pediatr Otorhinolaryngol. 2017 Jul;98:48-52. doi: 10.1016/j.ijporl.2017.04.041. Epub 2017 Apr 27.
OBJECTIVE: To determine the prevalence of pressure equalization tube (PET) placement and hearing loss in children with Down syndrome (DS).
MATERIAL AND METHODS: We evaluated 90 DS children births between 1 and 11 years old and compared to 90 children without DS paired in sex and age. Medical records were analyzed consecutively. Were collected data about proceedings PET placement, age of the patient at each PET, adenoidectomy, tonsillectomy and results for audiometry and tympanometry.
RESULTS: Among the 90 patients with DS, 49 (54.4%) were male, median age of 58 months (15-143 months). In this group, 75 PET were placed in 26/90 children (28.9%) mostly between 3 and 5 years old. In 10/26 (38.5%) was necessary PET replaced. When compared to the control group- 6/90 (6.7%)- children with DS presented OR = 13.7 (95% CI 4.0-47.3) times more likely to use PET. Adenoidectomy and tonsillectomy (44.4% and 42.2% respectively) were significantly more frequent in DS group. The prevalence of hearing loss was 32.1% in the right ear and 26.9% in the left ear. Type B timpanometry was found in more than half of the patients with DS.
CONCLUSION: We found a 13-fold higher risk of PET in DS children, especially between the ages of 3-5 years. The high prevalence of hearing loss and PET placement in patients with DS reinforcing the importance of early and regular follow-up for hearing screening in this population, mostly in preschool-aged children.

Berrettini S, Ghirri P, Lazzerini F, Lenzi G, Forli F.
Newborn hearing screening protocol in Tuscany region.
Ital J Pediatr. 2017 Sep 20;43(1):82. doi: 10.1186/s13052-017-0397-1.
BACKGROUND: Newborn hearing screening has to be considered the first step of a program for the identification, diagnosis, treatment and habilitation/rehabilitation of children with hearing impairment.
MAIN PART: In Tuscany Region of Italy, the universal newborn hearing screening is mandatory since November 2007. The first guidelines for the execution of the screening have been released in June 2008; then many other Italian regions partially or totally adopted these guidelines. On the basis of the experience from 2008 and according to the recent evidences in the scientific literature, a new screening protocol was released in Tuscany region. The new protocol is an evolution of the previous one. Some issues reported in the previous protocol and in the Joint Committee on Infant Hearing statement published in 2007 were revised, such as the risk factors for auditory neuropathy and for late onset, progressive or acquired hearing loss. The new updated guidelines were submitted to the Sanitary Regional Council and then they have been approved in August 2016. The updated screening protocol is mainly aimed to identify newborns with a congenital moderate-to-profound hearing loss, but it also provides indications for the audiological follow-up of children with risk's factor for progressive or late onset hearing loss; further it provides indications for the audiological surveillance of children at risk for acquired hearing impairment. Then, in the new guidelines the role of the family paediatrician in the newborn hearing screening and audiological follow-up and surveillance is underscored. Finally the new guidelines provide indications for the treatment with hearing aids and cochlear implant, in accordance with the recent Italian Health Technology Assessment (HTA) guidelines.
CONCLUSIONS: In the paper we report the modality of execution of the universal newborn hearing screening in the Tuscany Region, according to the recently updated protocol. The main features of the protocol and the critical issues are discussed.

Blázquez-Gamero D, Galindo Izquierdo A, Del Rosal T, Baquero-Artigao F, Izquierdo Méndez N, Soriano-Ramos M, Rojo Conejo P, González-Tomé MI, García-Burguillo A, Pérez Pérez N, Sánchez V, Ramos-Amador JT, De la Calle M.
Prevention and treatment of fetal cytomegalovirus infection with cytomegalovirus hyperimmune globulin: a multicenter study in Madrid.
J Matern Fetal Neonatal Med. 2017 Oct 4:1-211. doi: 10.1080/14767058.2017.1387890. [Epub ahead of print]
INTRODUCTION: Cytomegalovirus (CMV) is the leading cause of congenital infection worldwide. Data about the management of CMV infection in pregnant women are scarce, and treatment options are very limited. The aim of the study is to investigate the effectiveness of cytomegalovirus hyperimmune globulin (CMV-HIG) for the prevention and treatment of congenital CMV (cCMV) infection.
MATERIALS AND METHODS: A retrospective observational study was conducted in three tertiary hospitals in Madrid. In the period 2009-2015, CMV-HIG (Cytotect® CP Biotest, Biotest) treatment was offered to all pregnant women with primary CMV infection and/or detection of CMV-DNA in amniotic fluid in participating centers. Women were divided into prevention and treatment groups (PG and TG, respectively). Those with primary CMV infection who had not undergone amniocentesis comprised the PG and received monthly CMV-HIG (100 UI/kg). If CMV-DNA was subsequently detected in

amniotic fluid, one extra dose of CMV-HIG (200 UI/kg) was given 4 weeks after the last dose. Those women were considered to be part of the PG group despite detection of CMV-DNA in amniotic fluid. In the case of a negative result in CMV-DNA detection in amniotic fluid or if amniocentesis was not performed, monthly HIG was given up to the end of the pregnancy.
RESULTS: Thirty-six pregnant women were included. Median gestational age at birth was 39 weeks [interquartile range (IQR): 38-40] and 2 children (5.5%) were premature (born at 28 and 34 weeks' gestation). Amniocentesis was performed in 30/36 (83.4%) pregnancies and CMV PCR was positive in 21 of them (70%). One fetus with a positive PCR in amniotic fluid that received one dose of HIG after amniocentesis presented a negative CMV-PCR in urine at birth, and was asymptomatic at 12 months of age. Twenty-four children were infected at birth, and 16/21 (76.2%) presented no sequelae at 12 months, while 2 (9.5%) had mild unilateral hearing loss and three (14.3%) severe hearing loss or neurological sequelae. Seventeen women were included in the PG and 19 in the TG. In the PG 7/17 (41%) fetuses were infected, one pregnancy was terminated due to abnormalities in cordocentesis and one showed mild hearing loss at 12 months of age. In the TG, 18/19 children (95%) were diagnosed with cCMV, while the remaining neonate had negative urine CMV at birth. Eight out of the 19 fetuses (42.1%) showed CMV related abnormalities in fetal US before HIG treatment. Complete clinical assessment in the neonatal period and at 12 months of age was available in 16 and 15 children, respectively. At birth 50% were symptomatic and at 12 months of age, 4/15 (26.7%) showed hearing loss and 3/15 (20%) neurologic impairment. Fetuses with abnormalities in ultrasonography before HIG presented a high risk of sequelae (odds ratios (OR): 60 95%CI: 3-1185; p = 0.007).
DISCUSSION: Prophylactic HIG administration in pregnant women after CMV primary infection seems not to reduce significantly the rate of congenital infection, but is safe and it could have a favorable effect on the symptoms and sequelae of infected fetuses. The risk of long-term sequelae in fetuses without US abnormalities before HIG is low, so it could be an option in infected fetuses with normal imaging. On the other hand, the risk of sequelae among infected fetuses with abnormalities in fetal ultrasonography (FUS) before HIG despite treatment is high.

Bruijnzeel H, Bezdjian A, Lesinski-Schiedat A, Illg A, Tzifa
Evaluation of pediatric cochlear implant care throughout Europe: Is European pediatric cochlear implant care performed according to guidelines?
Cochlear Implants Int. 2017 Nov;18(6):287-296. doi: 10.1080/14670100.2017.1375238. Epub 2017 Sep 19.
OBJECTIVES: International guidelines indicate that children with profound hearing loss should receive a cochlear implant (CI) soon after diagnosis in order to optimize speech and language rehabilitation. Although prompt rehabilitation is encouraged by current guidelines, delays in cochlear implantation are still present. This study investigated whether European countries establish timely pediatric CI care based on epidemiological, commercial, and clinical data.
METHODS: An estimation of the number of pediatric CI candidates in European countries was performed and compared to epidemiological (Euro-CIU), commercial (Cochlear(®)), and clinical (institutional) age-at-implantation data. The ages at implantation of pediatric patients in eight countries (the Netherlands, Belgium, Germany, the United Kingdom, France, Turkey, Portugal, and Italy) between 2005 and 2015 were evaluated.
RESULTS: From 2010 onwards, over 30% of the pediatric CI candidates were implanted before 24 months of age. Northern European institutions implanted children on average around 12 months of age, whereas southern European institutions implanted children after 18 months of age. The Netherlands and Germany implanted earliest (between 6 and 11 months).
DISCUSSION: Implemented newborn hearing screening programs and reimbursement rates of CIs vary greatly within Europe due to local, social, financial, and political differences. However, internationally accepted recommendations are applicable to this heterogeneous European CI practice. Although consensus on early pediatric cochlear implantation exists, this study identified marked delays in European care.
CONCLUSION: Regardless of the great heterogeneity in European practice, reasons for latency should be identified on a national level and possibilities to prevent avoidable future implantation delays should be explored to provide national recommendations.

Bush ML, Taylor ZR, Noblitt B, Shackleford T, Gal TJ, Shinn JB, Creel LM, Lester C, Westgate PM, Jacobs JA, Studts CR.
Promotion of early pediatric hearing detection through patient navigation: A randomized controlled clinical trial.
Laryngoscope 2017 Nov;127 Suppl 7:S1-S13. doi: 10.1002/lary.26822. Epub 2017 Sep 20.
OBJECTIVES/HYPOTHESIS: To assess the efficacy of a patient navigator intervention to decrease nonadherence to obtain audiological testing following failed screening, compared to those receiving the standard of care.
METHODS: Using a randomized controlled design, guardian-infant dyads, in which the infants had abnormal newborn hearing screening, were recruited within the first week after birth. All participants were referred for definitive audiological diagnostic testing. Dyads were randomized into a patient navigator study arm or standard of care arm. The primary outcome was the percentage of patients with follow-up nonadherence to obtain diagnostic testing. Secondary outcomes were parental knowledge of infant hearing testing recommendations and barriers in obtaining follow-up testing.
RESULTS: Sixty-one dyads were enrolled in the study (patient navigator arm = 27, standard of care arm = 34). The percentage of participants nonadherent to diagnostic follow-up during the first 6 months after birth was significantly lower in the patient navigator arm compared with the standard of care arm (7.4% vs. 38.2%) (P = .005). The timing of initial follow-up was significantly lower in the navigator arm compared with the standard of care arm (67.9 days after birth vs. 105.9 days, P = .010). Patient navigation increased baseline knowledge regarding infant hearing loss diagnosis recommendations compared with the standard of care (P = .004).
CONCLUSIONS: Patient navigation decreases nonadherence rates following abnormal infant hearing screening and improves knowledge of follow-up recommendations. This intervention has the potential to improve the timeliness of delivery of infant hearing healthcare; future research is needed to assess the cost and feasibility of larger scale implementation.

Carew P, Mensah FK, Rance G, Flynn T, Poulakis Z, Wake M.
Mild-moderate congenital hearing loss: Secular trends in outcomes across four systems of detection.
Child Care Health Dev. 2017 Jun 14. doi: 10.1111/cch.12477. [Epub ahead of print]
BACKGROUND: Universal newborn hearing screening (UNHS) targets moderate or greater hearing loss. However, UNHS also frequently detects children with mild loss that results in many receiving early treatment. The benefits of this approach are not yet established. We aimed to (i) compare language and psychosocial outcomes between four hearing loss detection systems for children aged 5-8 years with congenital mild-moderate hearing loss; (ii) determine whether age of detection predicts outcomes; and (iii) compare outcomes between children identified via well-established UNHS and the general population.
METHODS: Linear regression adjusted for potential confounding factors was used throughout. Via a quasi-experimental design, language and psychosocial outcomes were compared across four population-based Australian systems of hearing loss detection: opportunistic detection, born 1991-1993, n = 50; universal risk factor referral, born 2003-2005, n = 34; newly established UNHS, born 2003-2005, n = 41; and well-established UNHS, born 2007-2010, n = 21. In pooled analyses, we examined whether age of detection predicted outcomes. Outcomes were similarly compared between

the current well-established UNHS system and typically developing children in the Early Language in Victoria Study, born 2003, n = 1217.
RESULTS: Age at diagnosis and hearing aid fitting fell steadily across the four systems. For moderate losses, mean expressive language (P for trend .05) and receptive vocabulary (P for trend .06) improved across the four systems, but benefit was not obvious for mild losses. In pooled analyses, diagnosis before age six months predicted better language outcomes for moderate losses. Children with mild-moderate losses exposed to well-established UNHS continue to experience expressive language scores well below children in the general population (adjusted mean difference -8.9 points, 95% CI -14.7 to -3.1).
CONCLUSIONS: Treatment arising from UNHS appears to be clearly benefitting children with moderate hearing losses. However, rigorous trials are needed to quantify benefits, versus costs and potential harms, of early aiding of children with mild losses.

Chan KH, Gao D, Jensen EL, Allen GC, Cass SP.
Complications and parent satisfaction in pediatric osseointegrated bone-conduction hearing implants.
Laryngoscope. 2017 Sep;127(9):2165-2170. doi: 10.1002/lary.26469. Epub 2017 Jan16.
OBJECTIVE: To assess long-term complication rate and parental satisfaction of osseointegrated bone conduction hearing implants (OBCHIs).
STUDY DESIGN: Retrospective chart review of children undergoing OBCHIs.
METHODS: A retrospective chart review of children undergoing OBCHIs for the treatment of conductive, mixed, and single-sided sensorineural hearing loss in children.
RESULTS: Forty-five subjects were identified with 0.3 to 10.4 years of follow-up. The mean/median age and age range at implant were 9.0/7.8 and 1.7 to 19.1 years. The underlying hearing loss for the cohort included conductive (N = 30), sensorineural (N = 7), and mixed (N = 8) hearing loss. Conductive hearing loss, caused by aural atresia (62.9), was the most common indication for implantation. Fifty-eight complications occurred in 29 subjects, most related to skin infection or overgrowth. Seventeen events required revision surgery, and 18 required oral antibiotics and/or office-based cauterization. Children under the age of 5 years were more likely to have failure of osseointegration or require revision surgery. Parents of 33 subjects underwent a phone interview; 76% rated the overall satisfaction as satisfied or very satisfied.
CONCLUSION: A large percentage of children undergoing OBCHI develop postoperative complications, and up to 44% require revision surgery-a figure higher than generally reported and higher than in adults. No factors were found to adequately explain the higher complication rates in children compared to adults. Despite the occurrence of complications, parents viewed this device as satisfactory from many perspectives.

Chen X, Yuan M, Lu J, Zhang Q, Sun M, Chang F.
Assessment of universal newborn hearing screening and intervention in Shanghai, China.
Int J Technol Assess Health Care. 2017 Jan;33(2):206-214. doi:10.1017/S0266462317000344. Epub 2017 Jun 6.
OBJECTIVES: The aim of this study was to evaluate the universal newborn hearing screening (UNHS) and intervention program in Shanghai, China.
METHODS: This study included the quantitative analyses of the UNHS-Shanghai database in 2002-12 and qualitative assessment of the program. The Otoacoustic Emissions and the Automated Auditory Brainstem Evoked Responses tests were conducted in screening. The costs and benefits were calculated based on the number of participants in each stage. The short-term and long-term periods were defined as from birth to 15 years of age or to death (82-year-olds), respectively. Sensitivity analyses were conducted.
RESULTS: A total 1,574,380 newborns were included, representing 93.6 percent of all eligible babies in Shanghai during the study period. The prevalence of newborn hearing loss was 1.66‰. The short-term/long-term program costs were ¥488.5 million (US$75.52 million)/¥1.08 billion (US$167.12 million), and the short-term/long-term program benefit was ¥980.1 million (US$151.53 million)/¥8.13 billion (US$1.26 billion). The program benefit was greater than its cost if the proportion of hearing-loss children enrolled in regular schools was no less than 41.4 percent of all hearing impaired children, as well as if the wage growth rate ranged from 3 percent to 8 percent. Qualitative results also suggested that stakeholders strongly supported this program.
CONCLUSIONS: The universal newborn hearing screening and intervention program in Shanghai is justified in terms of the resource input in the long run, although there is still room for further improvement with respect to educational rehabilitation and a better infrastructure system.

Ching TYC, Dillon H, Button L, Seeto M, Van Buynder P, Marnane V, Cupples L, Leigh G.
Age at intervention for permanent hearing loss and 5-year language outcomes.
Pediatrics. 2017 Sep;140(3). pii: e20164274. doi: 10.1542/peds.2016-4274. Epub 2017 Aug 3.
OBJECTIVES: Universal newborn hearing screening has been implemented to detect permanent childhood hearing loss (PCHL) early, with the ultimate goal of improving outcomes through early treatment. However, there is disagreement between studies on the size of this benefit and in some cases whether it is significantly different from 0. There have been no studies of sufficient size in which researchers have determined reliably whether the effect varies with degree of PCHL. We aimed to explore how intervention timing influences 5-year language in children with PCHL.
METHODS: Via a prospective study of 350 children, we used standard multiple regression analyses to investigate the effect of age at intervention or hearing screening on language outcomes after allowing for the effects of nonverbal IQ, degree of PCHL, sex, birth weight, maternal education, additional disabilities, and communication mode.
RESULTS: The benefit of early intervention for language development increased as hearing loss increased. Children whose amplification started at age 24 months had poorer language than those whose amplification started at 3 months. The difference was larger for 70-dB HL (-11.8 score points; 95% confidence interval [95% CI]: -18.7 to -4.8) than for 50-dB HL (-6.8; 95% CI: -10.8 to -2.8). Children who received cochlear implants at 24 months had poorer language than those implanted at 6 months (-21.4; 95% CI: -33.8 to -9.0). There was no significant effect of screening on outcomes.
CONCLUSIONS: Early intervention improves language outcomes, thereby lending support to streamlining clinical pathways to ensure early amplification and cochlear implantation after diagnosis.

Chiossi JSC, Hyppolito MA.
Effects of residual hearing on cochlear implant outcomes in children: A systematic-review.
Int J Pediatr Otorhinolaryngol. 2017 Sep;100:119-127. doi: 10.1016/j.ijporl.2017.06.036. Epub 2017 Jul 1.
OBJECTIVES: to investigate if preoperative residual hearing in prelingually deafened children can interfere on cochlear implant indication and outcomes.
METHODS: a systematic-review was conducted in five international databases up to November-2016, to locate articles that evaluated cochlear implantation in children with some degree of preoperative residual hearing. Outcomes were auditory, language and cognition performances after cochlear implant. The quality of the studies was assessed and classified according to the Oxford Levels of Evidence table - 2011. Risk of biases were also described.
RESULTS: From the 30 articles reviewed, two types of questions were identified: (a) what are the benefits of cochlear implantation in children with residual hearing? (b) is the preoperative residual hearing a predictor of cochlear implant outcome? Studies ranged from 04 to 188 subjects, evaluating populations between 1.8 and 10.3 years old. The definition of residual hearing varied between studies. The majority of articles (n = 22) evaluated speech perception as the outcome and 14 also assessed language and speech production.

CONCLUSION: There is evidence that cochlear implant is beneficial to children with residual hearing. Preoperative residual hearing seems to be valuable to predict speech perception outcomes after cochlear implantation, even though the mechanism of how it happens is not clear. More extensive researches must be conducted in order to make recommendations and to set prognosis for cochlear implants based on children preoperative residual hearing.

Chung W, Beauchaine KL, Grimes A, O'Hollearn T, Mason C, Ringwalt S.
Reporting newborn audiologic results to state EHDI programs.
Ear Hear. 2017 Sep/Oct;38(5):638-642. doi: 10.1097/AUD.0000000000000443.
OBJECTIVES: All US states and territories have an Early Hearing Detection and Intervention (EHDI) program to facilitate early hearing evaluation and intervention for infants who are deaf or hard of hearing. To ensure efficient coordination of care, the state EHDI programs rely heavily on audiologists' prompt reporting of a newborn's hearing status. Several states have regulations requiring mandatory reporting of a newborn's hearing status. This is an important public health responsibility of pediatric audiologists. Reasons for failing to report vary.
DESIGN: The Early Hearing Detection and Intervention-Pediatric Audiology Links to Services (EHDI) facility survey was used to inform reporting compliance of audiology facilities throughout the United States. The survey was disseminated via articles, newsletters, and call-to-action notices to audiologists.
RESULTS: Among 1024 facilities surveyed, 88 (8.6%) reported that they did not report newborn's hearing findings to their state EHDI program. Not knowing how to report to the state EHDI program was the most frequently chosen reason (60%). However, among the 936 facilities that were compliant with the reporting requirements, 51 estimated that they reported less than two-third of all hearing evaluation results (5.4%). Some facilities did not report a normal-hearing result and some failed to report because they assumed another facility would report the hearing results.
CONCLUSIONS: Survey results indicated that audiologists were compliant reporting hearing results to the state EHDI programs. However, there is room for improvement. Regular provider outreach and training by the state EHDI program is necessary to ensure those who are not reporting will comply and to clarify reporting requirements for those who are already compliant.

Cicuto Ferreira Rocha NA, de Campos AC, Cicuto Ferreira Rocha F, Pereira Dos Santos Silva F.
Microcephaly and Zika virus: Neuroradiological aspects, clinical findings and a proposed framework for early evaluation of child development.
Infant Behav Dev. 2017 Jul 26;49:70-82. doi: 10.1016/j.infbeh.2017.07.002. [Epub ahead of print]
BACKGROUND AND AIMS: As the recent outbreak of microcephaly cases caused by Zika virus has been declared a global health emergency, providing assessment guidelines for multidisciplinary teams providing early developmental screening and stimulation to infants with microcephaly is much needed. Thus, the aim of this manuscript is to provide an overview on what is known about neuroradiological aspects and clinical findings in infants with microcephaly caused by Zika virus and to propose a framework for early evaluation of child development.
METHODS: The keywords "Zika virus" and "microcephaly" were searched in PubMed database for articles published from incept to May 2017. These texts were reviewed, and the ones addressing neuroradiological and clinical findings in infants were selected. Recommendations for early assessment were made based on the International Classification of Functionality Disability and Health (ICF) model.
OUTCOMES AND RESULTS: The database search yielded 599 publications and 36 were selected. The studies detected microcephaly with diffuse brain malformations and calcifications, ventriculomegaly, optic nerve hypoplasia, macular atrophy, cataracts, impaired visual and hearing function, arthrogryposis, spasticity, hyperreflexia, irritability, tremors, and seizures, but very little is known about early development. Early assessments were described based on the ICF domains (Body Function and Structures, Activities and Participation and Contextual factors).
CONCLUSION AND IMPLICATIONS: Studies published showed abnormal brain, optic, neurologic and orthopedic findings, but very little is known about other aspects of functioning in infants with microcephaly caused by Zika virus. The biopsychosocial model based on the ICF paradigm provides an adequate framework to describe the condition of the infant with microcephaly receiving rehabilitative efforts to minimize disability. Efforts towards early identification of developmental delays should be taken within the first six months of life.

Cunningham M, Thomson V, McKiever E, Dickinson LM, Furniss A, Allison MA.
Infant, maternal, and hospital factors' role in loss to follow-up after failed newborn hearing screening.
Acad Pediatr. 2017 May 23. pii: S1876-2859(17)30321-2. doi: 10.1016/j.acap.2017.05.005. [Epub ahead of print]
OBJECTIVE: Completion of newborn hearing screening (NBHS) is recommended by 1 month old. Delays and loss to follow-up and documentation (LTF/LTD) after failed NBHS are common. Committees of experts have established hospital guidelines to reduce LTF/LTD. We aimed to identify maternal and infant factors associated with LTF/LTD and determine if adherence to hospital guidelines is associated with timely completion of follow-up screening.
METHODS: We conducted a retrospective study of all infants born in Colorado hospitals who failed the newborn admission hearing screening from 2007 to 2012 and a cross-sectional survey of NBHS coordinators at Colorado birthing hospitals. Neonatal intensive care unit infants were excluded. Outcomes included documented completion of the follow-up NBHS and completion by 1 month. Data sources comprised the electronic birth record, infant hearing integrated data system, and NBHS coordinator survey. Data were analyzed by logistic regression.
RESULTS: A total of 13,904 newborns did not pass the newborn admission hearing screening from 2007 to 2012, and 11,422 (82%) had documentation of a completed follow-up screening. A total of 10,558 (76%) completed follow-up screening by 1 month. All 53 NBHS coordinators completed the survey. Maternal age, education, smoking, and birth country; and payer, race, birth order, and population density were associated with completion of follow-up hearing screening. Maternal education, payer, population density, birth weight, and cleft lip were associated with completion by 1 month of age. Only birth in a facility that charges a rescreening fee was associated with completion of follow-up screening.
CONCLUSIONS: Low-income, rural, and minority infants are at risk for LTF. Further studies are needed to determine if adherence to guidelines can overcome barriers to follow-up.

Dejaco D, Aregger FC, Hurth HV, Kegele J, Muigg V, Oberhammer L, Bunk S, Fischer N, Pinggera L, Riedl D, Otieno A, Agbenyega T, Adegnika AA, Riechelmann H, Lackner P, Zorowka P, Kremsner P, Schmutzhard J.
Evaluation of transient-evoked otoacoustic emissions in a healthy 1 to 10 year pediatric

cohort in Sub-Saharan Africa.

Int J Pediatr Otorhinolaryngol. 2017 Oct;101:65-69. doi: 10.1016/j.ijporl.2017.07.029. Epub 2017 Jul 24.

OBJECTIVE: Transient-evoked otoacoustic emissions (TEOAEs) monitor cochlear function. High pass rates have been reported for industrialized countries. Pass rates in low and middle income countries such as Sub-Saharan Africa are rare, essentially lower and available for children up to 4 years of age and frequently based on hospital recruitments. This study aims at providing additional TEOAE pass rates of a healthy Sub-Saharan cohort aged 1-10 years with data from Gabon, Ghana and Kenya. Potentially confounding factors (recruitment site, age) are taken into consideration.
METHODS: Healthy children were recruited in hospitals, schools and kindergartens. Inclusion criteria were age 1-10 years and normal otoscopic findings. Exclusion criteria were any sickness or physical ailment potentially impairing the hearing capacity. Five measurements per ear were performed with Capella Cochlear Emission Analyzer (MADSEN, Germany). An overall wave reproducibility of above 60% served as pass-criterion. Pass rates were compared between recruitment sites and age groups (1-5 and 6-10 years).
RESULTS: Overall pass rate was 87.5% (n = 264; 231 passes vs. 33 fails). Of these 84.0% of hospital recruited children passed (n = 156; 131 passes vs. 25 fails), compared to 92.6% of community recruitments (n = 108; 100 passes vs. 8 fails), which was significantly different p = 0.039). If analyzed by age groups, this difference was only observed in children younger than 6 years (p = 0.007).
CONCLUSION: Hospitals as recruitment sites for healthy controls seem to affect TEOAE pass rates. We advise for a cautious approach when recruiting healthy TEOAE control collectives under the age of 6 in a hospital setting. In children older than 6 years conventional pure-tone audiometry remains the standard method for hearing screening.

Dobbie AM.

Evaluation and management of cytomegalovirus-associated congenital hearing loss.

Curr Opin Otolaryngol Head Neck Surg. 2017 Oct;25(5):390-395. doi: 10.1097/MOO.0000000000000401.

PURPOSE OF REVIEW: The current article reviews the current literature related to congenital cytomegalovirus (CMV)-related hearing loss. The discussion will focus on the epidemiology, pathogenesis, and clinical presentation of human CMV infection as it pertains to hearing loss. Current methods of CMV diagnosis with a focus on the evolving trend toward broader neonatal screening protocols will also be explored. discussion of medical, surgical, and audiologic management of the condition will also be addressed.
RECENT FINDINGS: Much of the current research on this topic is focused on improving detection of CMV through screening programs. Some advances in understanding cochlear pathogenesis have also been made.
SUMMARY: Congenital CMV infection remains an important cause of hearing loss in infants and children. Early detection of CMV infection can broaden treatment options and allow for improved hearing and language outcome for patients with CMV-associated sensorineural hearing loss.

Du Y, Huang L, Wang X, Cui Q, Cheng X, Zhao L, Ni T.

Clinical data analysis of genotypes and phenotypes of deafness gene mutations in newborns: A retrospective study.

Biosci Trends. 2017 Sep 12;11(4):460-468. doi: 10.5582/bst.2017.01070. Epub 2017 Jul 17.

We retrospectively analyzed newborns with deafness gene mutations and summarized the relationship between genotype and phenotype to provide a basis for genetic counseling. We studied 582 subjects positive for deafness gene mutations that were treated in the otology outpatient department of Beijing Tongren Hospital, Capital Medical University, between April 2012 and April 2016. The subjects were divided into 3 categories: a diagnosed group (group A), which was further subdivided into subgroups A1 (homozygous and compound heterozygous GJB2 mutations) and A2 (homozygous and compound heterozygous SLC26A4 mutations); a drug-induced deafness group (group B, mitochondrial (Mt) gene mutations); and a mutation carrier group (group C), which was further subdivided into the subgroups C1 (GJB2 heterozygous mutations), C2 (SLC26A4 heterozygous mutations), C3 (GJB3 heterozygous mutations), and C4 (double gene mutations). Partial sequences positive for GJB2 or SLC26A4 were sequenced and analyzed for mutations. Subjects underwent otoscopic examination and comprehensive audiological evaluation, and temporal bone computerized tomography and/or inner ear magnetic resonance imaging were performed. GJB2 235delC was the most common mutation locus. The highest proportion of deafness detected during universal newborn hearing screening was for drug-induced deafness, whereas the lowest was for the diagnosed group. GJB2 gene mutations mainly resulted in flat-type, profound-to-severe sensorineural hearing loss (SNHL). SLC26A4 gene mutation was mainly associated with high-frequency drop-type and profound-severe SNHL and was closely related to enlargement of the vestibular aqueduct.

Dudda R, Muniyappa HP, Puttaraju S, Lakshmi MS.

A qualitative study on knowledge and attitude towards risk factors, early identification and intervention of infant hearing loss among puerperal mothers- a short survey.

J Clin Diagn Res. 2017 Jul;11(7):MC01-MC05. doi: 10.7860/JCDR/2017/25837.10238. Epub 2017 Jul 1.

INTRODUCTION: Maternal active participation and their support are critical for the success of early hearing loss detection program. Erroneous maternal decisions may have large life long consequences on the infant's life. The mothers' knowledge and their attitudes towards infant hearing loss is the basis for their decisions.
AIM: The present study was done to determine the mothers' knowledge and their attitude towards risk factors of infant hearing loss, its early identification and intervention and also awareness of effect of consanguinity on hearing loss.
MATERIALS AND METHODS: In this cross-sectional questionnaire survey study, a total of 100 mothers were interviewed using the questionnaire which consisted of three sections namely risk factors, early identification and early intervention of hearing loss. Chi-square test was used to establish relationship between consanguineous and non-consanguineous mother's responses to its effect on hearing loss. A p-value < 0.05 was considered as significant.
RESULTS: Mothers' awareness was significantly high for visible causes (ear pain/discharge, head injury and slap to ear) of hearing loss. Positive attitude was seen for importance of screening programs and follow up testing. Moderate level of awareness was found on hazards of consanguinity and benefits of early identification. However, mothers were least aware of neonatal jaundice, NICU admission (>5 days), signs of late-onset and neural hearing loss, management of hearing loss, hearing aid fitting and therapy necessity, which might interfere in early detection and intervention of hearing loss.
CONCLUSION: It is crucial to educate mothers on few risk factors and management of hearing loss to reduce its consequences.

Fengler I, Nava E, Villwock AK, Büchner A, Lenarz T, Röder B.

Multisensory emotion perception in congenitally, early, and late deaf CI users.

PLoS One. 2017 Oct 12;12(10):e0185821. doi: 10.1371/journal.pone.0185821. eCollection 2017.

Emotions are commonly recognized by combining auditory and visual signals (i.e., vocal and facial expressions). Yet it is unknown whether the ability to link emotional signals across modalities depends on early experience with audio-visual stimuli. In the present study, we investigated the role of auditory experience at different stages of development for auditory, visual, and multisensory emotion recognition abilities in three groups of adolescent and adult cochlear implant (CI) users. CI users had a different deafness onset and were compared to three groups of age- and gender-matched hearing control participants. We hypothesized that congenitally deaf (CD) but not early deaf (ED) and late deaf (LD) CI users would show reduced multisensory interactions and a higher visual dominance in emotion perception than their hearing controls. The CD (n = 7), ED (deafness onset: <3 years of age; n = 7), and LD (deafness onset: >3 years; n = 13) CI users and the control participants performed an emotion recognition task with auditory, visual, and audio-visual emotionally congruent and incongruent nonsense speech stimuli. In different blocks, participants judged either the vocal (Voice task) or the facial expressions (Face task). In the Voice task, all three CI groups performed overall less efficiently than their respective controls and experienced higher interference from incongruent facial information. Furthermore, the ED CI users benefitted more than their controls from congruent faces and the CD CI users showed an analogous trend. In the Face task, recognition efficiency of the CI users and controls did not differ. Our results suggest that CI users acquire multisensory interactions to some degree, even after congenital deafness. When judging affective prosody they appear impaired and more strongly biased by concurrent facial information than typically hearing individuals. We speculate that limitations inherent to the CI contribute to these group differences.

Fitzpatrick EM, Dos Santos JC, Grandpierre V, Whittingham J.

Exploring reasons for late identification of children with early-onset hearing loss.

Int J Pediatr Otorhinolaryngol. 2017 Sep;100:160-167. doi:10.1016/j.ijporl.2017.06.039. Epub 2017 Jul 1.

INTRODUCTION: Several studies have shown that early identification of childhood hearing loss leads to better language outcomes. However, delays in the confirmation of hearing loss persist even in the presence of well-established universal newborn hearing screening programs (UNHS). The objective of this population-based study was to document the proportion of children who experienced delayed confirmation of congenital and early onset hearing loss in a UNHS program in one region of Canada. The study also sought to determine the reasons for delayed confirmation of hearing loss in children.

METHODS: Population level data related to age of first assessment, age of identification and clinical characteristics were collected prospectively for all children identified through the UNHS program. We documented the number of children who experienced delay (defined as more than 3 months) from initial audiologic assessment to confirmation of hearing loss. A detailed chart review was subsequently performed to examine the reasons for delay to confirmation.

RESULTS: Of 418 children identified from 2003 to 2013, 182 (43.5%) presented with congenital or early onset hearing loss, of whom 30 (16.5%) experienced more than 3 months delay from initial audiologic assessment to confirmation of their hearing disorder. The median age of first assessment and confirmation of hearing loss for these 30 children was 3.7 months (IQR: 2.0, 7.6) and 13.8 months (IQR: 9.7, 26.1) respectively. Close examination of the factors related to delay to confirmation revealed that for the overwhelming majority of children, a constellation of factors contributed to late diagnosis. Several children (n = 22; 73.3%) presented with developmental/medical issues, 15 of whom also had middle ear dysfunction at assessment, and 9 of whom had documented family follow-up concerns. For the remaining eight children, additional reasons included ongoing middle ear dysfunction for five children, complicated by family follow-up concerns (n = 3) and mild hearing loss (n = 1) and the remaining three children had isolated reasons related to family follow-up (n = 1) or mild hearing loss (n = 2).

CONCLUSION: Despite the progress made in the early detection of pediatric hearing loss since UNHS, a substantial number of children referred for early assessment can experience late confirmation and intervention. In particular, infants with developmental and/or medical issues including middle ear disorders are at particular risk for longer time to confirmation of hearing loss.

Fitzpatrick EM, Al-Essa RS, Whittingham J, Fitzpatrick J.

Characteristics of children with unilateral hearing loss.

Int J Audiol. 2017 Nov;56(11):819-828. doi: 10.1080/14992027.2017.1337938. Epub 2017 Jun 22.

OBJECTIVE: The purpose of this study was to describe the clinical characteristics of children with unilateral hearing loss (UHL), examine deterioration in hearing, and explore amplification decisions.

DESIGN: Population-based data were collected prospectively from time of diagnosis. Serial audiograms and amplification details were retrospectively extracted from clinical charts to document the trajectory and management of hearing loss.

SAMPLE: The study included all children identified with UHL in one region of Canada over a 13-year period (2003-2015) after implementation of universal newborn hearing screening.

RESULTS: Of 537 children with permanent hearing loss, 20.1% (108) presented with UHL at diagnosis. They were identified at a median age of 13.9 months (IQR: 2.8, 49.0). Children with congenital loss were identified at 2.8 months (IQR: 2.0, 3.6) and made up 47.2% (n=51), reflecting that a substantial portion had late-onset, acquired or late-identified loss. A total of 42.4% (n=39) showed deterioration in hearing, including 16 (17.4%) who developed bilateral loss. By study end, 73.1% (79/108) of children had received amplification recommendations.

CONCLUSIONS: Up to 20% of children with permanent HL are first diagnosed with UHL. About 40% are at risk for deterioration in hearing either in the impaired ear and/or in the normal hearing ear.

Fontenot TE, Giardina CK, Teagle HF, Park LR, Adunka OF, Buchman CA, Brown KD, Fitzpatrick DC.

Clinical role of electrocochleography in children with auditory neuropathy spectrum disorder.

Int J Pediatr Otorhinolaryngol. 2017 Aug;99:120-127. doi:10.1016/j.ijporl.2017.05.026. Epub 2017 Jun 5.

OBJECTIVES: To assess electrocochleography (ECochG) to tones as an instrument to account for CI speech perception outcomes in children with auditory neuropathy spectrum disorder (ANSD).

MATERIALS & METHODS: Children (<18 years) receiving CIs for ANSD (n = 30) and non-ANSD (n = 74) etiologies of hearing loss were evaluated with ECochG using tone bursts (0.25-4 kHz). The total response (TR) is the sum of spectral peaks of responses across frequencies. The compound action potential (CAP) and the auditory nerve neurophonic (ANN) in ECochG waveforms were used to estimate nerve activity and calculate nerve score. Performance on open-set monosyllabic word tests was the outcome measure. Standard statistical methods were applied.

RESULTS: On average, TR was larger in ANSD than in non-ANSD subjects. Most ANSD (73.3%) and non-ANSD (87.8%) subjects achieved open-set speech perception; TR accounted for 33% and 20% of variability in the outcomes, respectively. In the ANSD group, the PTA accounted for 69.3% of the variability, but there was no relationship with outcomes in the non-ANSD group. In both populations, nerve score was sensitive in identifying subjects at risk for not acquiring open-set speech perception, while the CAP and the ANN were more specific.

CONCLUSION: In both subject groups, the TRs correlated with outcomes but these measures were notably larger in the ANSD group. There was also strong correlation between PTA and speech perception outcome in ANSD group. In both subject populations, weaker evidence of neural activity was related to failure to achieve open-set speech perception.

Franck C, Vorwerk W, Köhn A, Rißmann A, Vorwerk U.
Prevalence, risk factors and diagnostics of hearing impairment in preterm infants.
[Article in German; Abstract available in German from the publisher]
Laryngorhinootologie. 2017 Jun;96(6):354-360. doi: 10.1055/s-0043-109512. Epub 2017 Jul 11.

INTRODUCTION: The preterm birth is clearly associated with increased risk of developing congenital hearing impairment. Therefore, special attention must be paid to the postnatal control of auditory function in all preterm infants. The present work investigates if the latest scientific findings regarding prevalence, clinical diagnostics, therapy and risk factors of hearing impairment in premature infants are regularly implemented in daily practice.
METHODS: At the department of phoniatrics and pediatric audiology of the University m of Magdeburg, the treatment data of 126 preterm children born between 2006 and 2011 were evaluated retrospectively. The additional analysis of all records available at the screening center (n=67 640) covering this period enables drawing conclusions on the total number and prevalence of hearing impairment in preterm infants in Saxony-Anhalt.
RESULTS: Almost all premature babies, like mature newborns, underwent postnatal hearing screening of both ears. The data analysis shows that the practical implementation often does not comply with the guideline of the G-BA (Gemeinsamer Bundesausschuss) in all details. For example, the recommended screening method for preterm infants (AABR) or the screening and treatment timing are not always applied in accordance with the guidelines of the G-BA.
DISCUSSION: Assessment of the practical implementation of universal newborn hearing screening was planned at the time of the introduction of the hearing screening program by the G-BA. As a part of this investigation, the practical care of vulnerable groups such as preterm infants must be given special attention. Based on the collected data, the diagnostics and therapy should be unified. Regardless of the maternity clinic where the infants were born, there should be the same opportunity for early diagnosis and thus for prognostically better treatment of congenital hearing impairment. Rapid postnatal fitting with hearing aid can stimulate the maturation of the central auditory system and potentially help to avoid problems of hearing and speech development.

Garinis AC, Kemph A, Tharpe AM, Weitkamp JH, McEvoy C, Steyger PS.
Monitoring neonates for ototoxicity.
Int J Audiol. 2017 Jun 22:1-8. doi: 10.1080/14992027.2017.1339130. [Epub ahead of print]

OBJECTIVES: Neonates admitted to the neonatal intensive care unit (NICU) are at greater risk of permanent hearing loss compared to infants in well mother and baby units. Several factors have been associated with this increased prevalence of hearing loss, including congenital infections (e.g. cytomegalovirus or syphilis), ototoxic drugs (such as aminoglycoside or glycopeptide antibiotics), low birth weight, hypoxia and length of stay. The aetiology of this increased prevalence of hearing loss remains poorly understood.
DESIGN: Here we review current practice and discuss the feasibility of designing improved ototoxicity screening and monitoring protocols to better identify acquired, drug-induced hearing loss in NICU neonates.
STUDY SAMPLE: A review of published literature.
CONCLUSIONS: We conclude that current audiological screening or monitoring protocols for neonates are not designed to adequately detect early onset of ototoxicity. This paper offers a detailed review of evidence-based research, and offers recommendations for developing and implementing an ototoxicity monitoring protocol for young infants, before and after discharge from the hospital.

Garinis AC, Liao S, Cross CP, Galati J, Middaugh JL, Mace JC, Wood AM, McEvoy L, Moneta L, Lubianski T, Coopersmith N, Vigo N, Hart C, Riddle A, Ettinger O, Nold C, Durham H, MacArthur C, McEvoy C, Steyger PS.
Effect of gentamicin and levels of ambient sound on hearing screening outcomes in the neonatal intensive care unit: A pilot study.
Int J Pediatr Otorhinolaryngol. 2017 Jun;97:42-50. doi: 10.1016/j.ijporl.2017.03.025. Epub

OBJECTIVE: Hearing loss rates in infants admitted to neonatal intensive care units (NICU) run at 2-15%, compared to 0.3% in full-term births. The etiology of this difference remains poorly understood. We examined whether the level of ambient sound and/or cumulative gentamicin (an aminoglycoside) exposure affect NICU hearing screening results, as either exposure can cause acquired, permanent hearing loss. We hypothesized that higher levels of ambient sound in the NICU, and/or gentamicin dosing, increase the risk of referral on the distortion product otoacoustic emission (DPOAE) assessments and/or automated auditory brainstem response (AABR) screens.
METHODS: This was a prospective pilot outcomes study of 82 infants (<37 weeks gestational age) admitted to the NICU at Oregon Health & Science University. An ER-200D sound pressure level dosimeter was used to collect daily sound exposure in the NICU for each neonate. Gentamicin dosing was also calculated for each infant, including the total daily dose based on body mass (mg/kg/day), as well as the total number of treatment days. DPOAE and AABR assessments were conducted prior to discharge to evaluate hearing status. Exclusion criteria included congenital infections associated with hearing loss, and congenital craniofacial or otologic abnormalities.
RESULTS: The mean level of ambient sound was 62.9 dBA (range 51.8-70.6 dBA), greatly exceeding American Academy of Pediatrics (AAP) recommendation of <45.0 dBA. More than 80% of subjects received gentamicin treatment. The referral rate for (i) AABRs, (frequency range: □1000-4000 Hz), was 5%; (ii) DPOAEs with a broad F2 frequency range (2063-10031 Hz) was 39%; (iii) DPOAEs with a low-frequency F2 range (<4172 Hz) was 29%, and (iv) DPOAEs with a high-frequency F2 range (>4172 Hz) was 44%. DPOAE referrals were significantly greater for infants receiving >2 days of gentamicin dosing compared to fewer doses ($p = 0.004$). The effect of sound exposure and gentamicin treatment on hearing could not be determined due to the low number of NICU infants without gentamicin exposure (for control comparisons).
CONCLUSION: All infants were exposed to higher levels of ambient sound that substantially exceed AAP guidelines. More referrals were generated by DPOAE assessments than with AABR screens, with significantly more DPOAE referrals with a high-frequency F2 range, consistent with sound- and/or gentamicin-induced cochlear dysfunction. Adding higher frequency DPOAE assessments to existing NICU hearing screening protocols could better identify infants at-risk for ototoxicity.

Geers AE, Mitchell CM, Warner-Czyz A, Wang NY, Eisenberg LS; CDaCI Investigative Team.
Early sign language exposure and cochlear implantation benefits.
Pediatrics. 2017 Jul;140(1). pii: e20163489. doi: 10.1542/peds.2016-3489. Epub 2017 Jun 12.

BACKGROUND: Most children with hearing loss who receive cochlear implants (CI) learn spoken language, and parents must choose early on whether to use sign language to accompany speech at home. We address whether parents' use of sign language before and after CI positively influences auditory-only speech recognition, speech intelligibility, spoken language, and reading outcomes.
METHODS: Three groups of children with CIs from a nationwide database who differed in the duration of early sign language exposure provided in their homes were compared in their progress through elementary grades. The groups did not differ in demographic, auditory, or linguistic

characteristics before implantation.
RESULTS: Children without early sign language exposure achieved better speech recognition skills over the first 3 years postimplant and exhibited a statistically significant advantage in spoken language and reading near the end of elementary grades over children exposed to sign language. Over 70% of children without sign language exposure achieved age-appropriate spoken language compared with only 39% of those exposed for 3 or more years. Early speech perception predicted speech intelligibility in middle elementary grades. Children without sign language exposure produced speech that was more intelligible (mean = 70%) than those exposed to sign language (mean = 51%).
CONCLUSIONS: This study provides the most compelling support yet available in CI literature for the benefits of spoken language input for promoting verbal development in children implanted by 3 years of age. Contrary to earlier published assertions, there was no advantage to parents' use of sign language either before or after CI.

Gouws N, Swanepoel W, De Jager LB.
Wideband acoustic immittance for assessing middle ear functioning for preterm neonates in the neonatal intensive care unit.
S Afr J Commun Disord. 2017 Jun 28;64(1):e1-e11. doi: 10.4102/sajcd.v64i1.182.
BACKGROUND: The primary aim of newborn hearing screening is to detect permanent hearing loss. Because otoacoustic emissions (OAEs) and automated auditory brainstem response (AABR) are sensitive to hearing loss, they are often used as screening tools. On the other hand, false-positive results are most often because of transient outer- and middle ear conditions. Wideband acoustic immittance (WAI), which includes physical measures known as reflectance and absorbance, has shown potential for accurate assessment of middle ear function in young infants.
OBJECTIVE: The main objective of this study was to determine the feasibility of WAI as a diagnostic tool for assessing middle ear functioning in preterm neonates in the neonatal intensive care unit (NICU) designed for premature and ill neonates. A further objective was to indicate the difference between the reflectance values of tones and click stimuli.
METHOD: Fifty-six at-risk neonates (30 male and 26 female), with a mean age at testing of 35.6 weeks (range: 32-37 weeks) and a standard deviation of 1.6 from three private hospitals, who passed both the distortion product optoacoustic emission (DPOAE) and AABR tests, were evaluated prior to discharge from the NICU. Neonates who presented with abnormal DPOAE and AABR results were excluded from the study. WAI was measured by using chirp and tone stimuli. In addition to reflectance, the reflectance area index (RAI) values were calculated.
RESULTS: Both tone and chirp stimuli indicated high-power reflectance values below a frequency of 1.5 kHz. Median reflectance reached a minimum of 0.67 at 1 kHz - 2 kHz but increased to 0.7 below 1 kHz and 0.72 above 2 kHz for the tone stimuli. For chirp stimuli, the median reflectance reached a minimum of 0.51 at 1 kHz - 2 kHz but increased to 0.68 below 1 kHz and decreased to 0.5 above 2 kHz. A comparison between the present study and previous studies on WAI indicated a substantial variability across all frequency ranges.
CONCLUSION: These WAI measurements conducted on at-risk preterm NICU neonates (mean age at testing: 35.6 weeks, range: 32-37 weeks) identified WAI patterns not previously reported in the literature. High reflective values were obtained across all frequency ranges. The age of the neonates when tested might have influenced the results. The neonates included in the present study were very young preterm neonates compared to the ages of neonates in previous studies. WAI measured in at-risk preterm neonates in the NICU was variable with environmental and internal noise influences. Transient conditions affecting the sound-conduction pathway might have influenced the results. Additional research is required to investigate WAI testing in ears with and without middle ear dysfunction. The findings of the current study imply that in preterm neonates it was not possible to determine the feasibility of WAI as a diagnostic tool to differentiate between ears with and without middle ear pathology.

Govender SM, Khan NB.
Knowledge and cultural beliefs of mothers regarding the risk factors of infant hearing loss and awareness of audiology services.
J Public Health Afr. 2017 Sep 4;8(1):557. doi: 10.4081/jphia.2017.557.eCollection 2017 Jun 23.
The aim of the paper is to describe the knowledge of mothers in Durban, South Africa, regarding risk factors of hearing loss in infants and their awareness of audiology services, and to describe their cultural beliefs about the risk factors for hearing loss in infants. A descriptive survey design with quantitative **METHODS** of analysis were used. Conveniently sampled mothers (n=102) receiving postnatal care for their infants from eight provincial clinics within Durban consented to participate, yielding a response rate of 48%. A questionnaire was used to collect the data and the Cronbach α was calculated yielding a score of 0.835, indicating good internal consistency and reliability of the questionnaire. Sixty percent of the mothers were aware of risk factors, such as middle ear infections, ototoxic medication and consumption of alcohol during pregnancy. Seventy percent were unaware that NICU/mechanical ventilation for more than 5 days, prematurity, rubella and jaundice are considered risk factors for hearing loss, implying a need to create awareness amongst mothers regarding such risk factors. Sixty percent (n=62) believed that bewitchment and ancestral curses can cause hearing loss. Cultural beliefs were associated with hearing loss, therefore, health professionals need to demonstrate cultural competence when providing audiology services, especially in a culturally and linguistically diverse countries such as South Africa. Although the mothers had an average knowledge about risk factors, two thirds did not know which professional to seek help from. There is a need to create awareness amongst mothers regarding the risk factors of infant hearing loss as well as audiology services in order to facilitate early detection and intervention. There is a need for health professionals to demonstrate cultural competence when working with their patients.

Grandori F, Hayes D.
Reflections on Lake Como Conferences (2000-2016).
Am J Audiol. 2017 Oct 12;26(3S):467-468. doi:10.1044/2017_AJA-17-0062.
PURPOSE: We present an overview of the conceptualization and development of the Newborn Hearing Screening and Hearing Across the Lifespan (Lake Como) conferences from 2000 to 2016.
Grosse SD, Riehle-Colarusso T, Gaffney M, Mason CA, Shapira SK, Sontag MK, Braun KVN, Iskander J.
CDC grand rounds: Newborn screening for hearing loss and critical congenital heart disease.
MMWR Morb Mortal Wkly Rep. 2017 Aug 25;66(33):888-890. doi:10.15585/mmwr.mm6633a4.

Newborn screening is a public health program that benefits 4 million U.S. infants every year by enabling early detection of serious conditions, thus affording the opportunity for timely intervention to optimize outcomes (1). States and other U.S. jurisdictions decide whether and how to regulate newborn screening practices. Most newborn screening is done through laboratory analyses of dried bloodspot specimens collected from newborns. Point-of-care newborn screening is typically performed before discharge from the birthing facility. The Recommended Uniform Screening Panel includes two point-of-care conditions for newborn screening: hearing loss and critical congenital heart disease (CCHD). The objectives of point-of-care screening for these two conditions are early identification and intervention to improve neurodevelopment, most notably language and related skills among infants with permanent hearing loss, and to prevent death or severe disability resulting from delayed diagnosis of CCHD. Universal screening for hearing loss using otoacoustic emissions or automated auditory brainstem response was endorsed by the Joint Committee on Infant Hearing in 2000 and 2007* and was incorporated in the first Recommended Uniform Screening Panel in 2005. Screening for CCHD using pulse oximetry was recommended by the Advisory Committee on Heritable Disorders in Newborns and Children in 2010 based on an evidence review(†) and was added to the Recommended Uniform Screening Panel in 2011.(§).

Guerzoni L, Cuda D.

Speech processor data logging helps in predicting early linguistic outcomes in implanted children.

Int J Pediatr Otorhinolaryngol. 2017 Oct;101:81-86. doi: 10.1016/j.ijporl.2017.07.026. Epub 2017 Jul 25.

OBJECTIVE: To analyse the value of listening-data logged in the speech processor on the prediction of the early auditory and linguistic skills in children who received a cochlear implant in their first 2 years of life.
STUDY DESIGN: Prospective observational non-randomized study.
METHODS: Ten children with profound congenital sensorineural hearing loss were included in the study. The mean age at CI activation was 16.9 months (SD ± 7.2; range 10-24). The auditory skills were evaluated with the Infant Toddler Meaningful Inventory Scale and the Category of Auditory Performance. Lexical level was assessed with the MacArthur-Bates Communicative Development Inventory. The overall data of average daily use and acoustic scene-analyses were extracted from Data Logging system. The effect of the one-year cumulative listening time to speech (in quiet) and speech-in-noise on the auditory and lexical scores was analysed.
RESULTS: A significant positive correlation was found between speech in quiet exposure time at low loudness level (<70 dB) and lexical quotient after one year of CI use. Infant Toddler Meaningful Inventory Scale was negatively correlated with the highest speech-in-noise loudness levels (>80 dB). The Category of Auditory Performance was not related to the logged data.
CONCLUSION: The listening environment can influence the early functional outcomes in younger implanted children. In this perspective, the data logging system is a promising tool in predicting early linguistic and auditory outcomes.

Hall WC.

What you don't know can hurt you: The risk of language deprivation by impairing sign language development in deaf children.

Matern Child Health J. 2017 May;21(5):961-965. doi: 10.1007/s10995-017-2287-y.

A long-standing belief is that sign language interferes with spoken language development in deaf children, despite a chronic lack of evidence supporting this belief. This deserves discussion as poor life outcomes continue to be seen in the deaf population. This commentary synthesizes research outcomes with signing and non-signing children and highlights fully accessible language as a protective factor for healthy development. Brain changes associated with language deprivation may be misrepresented as sign language interfering with spoken language outcomes of cochlear implants. This may lead to professionals and organizations advocating for preventing sign language exposure before implantation and spreading misinformation. The existence of one-time-sensitive-language acquisition window means a strong possibility of permanent brain changes when spoken language is not fully accessible to the deaf child and sign language exposure is delayed, as is often standard practice. There is no empirical evidence for the harm of sign language exposure but there is some evidence for its benefits, and there is growing evidence that lack of language access has negative implications. This includes cognitive delays, mental health difficulties, lower quality of life, higher trauma, and limited health literacy. Claims of cochlear implant- and spoken language-only approaches being more effective than sign language-inclusive approaches are not empirically supported. Cochlear implants are an unreliable standalone first-language intervention for deaf children. Priorities of deaf child development should focus on healthy growth of all developmental domains through a fully-accessible first language foundation such as sign language, rather than auditory deprivation and speech skills.

Han R, Li L, Duan L, Xia Y, Kuyaxi P, Zhao J, Zhao Q, Zhang H, Chen Y.

Efficiency of microarray and SNPscan for the detection of hearing loss gene in 71 cases with nonsyndromic hearing loss.

Medicine (Baltimore). 2017 Jun;96(25):e7149. doi: 10.1097/MD.0000000000007149.

We aim to screen the mutations of 3 hearing loss (HL) genes (GJB2, SLC26A4, and 12S rRNA) in 71 cases with nonsyndromic hearing loss (NSHL) using microarray and SNPscan, and identify the roles of nonhotspot mutation of these genes in the screening of NSHL. Seventy-one cases with moderate or severe neurosensory deafness confirmed in our department from July 2014 to December 2015 including 25 Uyghur minorities and 46 Han Chinese were included in this study. The type of mutations in GJB2, SLC26A4, and 12S rRNA genes were detected using microarray and SNPscan, respectively. Statistical difference was noticed in the detection rate of the HL genes in 71 cases. Using microassay, deafness genes were identified in 10 subjects (14.08%), while 22 cases (30.98%) were confirmed with the presence of deafness genes using the SNPscan. Compared with the microarray, remarkable difference was noticed in the detection rate of SNPscan ($P<.05$). Nonhotspot mutation in GJB2, SLC26A4, and 12S rRNA genes played a crucial role in the pathogenesis of NSHL. SNPscan contributed to elevation of detection rate of NSHL in clinical practice.

Harris MS, Dodson EE.

Hearing health access in developing countries.

Curr Opin Otolaryngol Head Neck Surg. 2017 Oct;25(5):353-358. doi:10.1097/MOO.0000000000000392.

PURPOSE OF REVIEW: The developing world carries a disproportionate burden of hearing loss. Individuals with hearing loss in austere settings worldwide are also potentially impacted by their impairment to a greater extent owing to underdeveloped or nonexistent hearing health infrastructure. The purpose of this review is to examine the state of the literature on hearing health access in developing countries and identify areas for improvement.
RECENT FINDINGS: Over the last 10 years progress has been made in some areas, whereas other aspects of hearing health in developing countries have changed very little. There has been expansion of efforts to train primary care and local hearing healthcare providers to recognize and appropriately treat preventable causes of hearing loss in the developing world. Applications of telehealth to connect providers and patients in rural

locales have grown. Adaptions of newborn hearing screening programs that better fit local resources and customs have been reported. There has been a considerable increase in interest, including cost-benefit analyses, with regard to the use of cochlear implants in the developing world.
SUMMARY: In spite of progress, the developing world still shoulders a disproportionate amount of the world's hearing loss, in particular chronic ear disease, and there is a paucity of well trained local hearing healthcare professionals to deal with the challenge. The role of international humanitarian efforts, telemedicine, and education is highlighted.

Hoey AW, Pai I, Driver S, Connor S, Wraige E, Jiang D.
Management and outcomes of cochlear implantation in patients with congenital cytomegalovirus (cCMV)-related deafness.
Cochlear Implants Int. 2017 Jul;18(4):216-225. doi:10.1080/14670100.2017.1315510. Epub 2017 May 9.
OBJECTIVE: Congenital Cytomegalovirus (cCMV) is a well-defined cause for neonatal mortality and morbidity, particularly sensorineural hearing loss and other neurodevelopmental disruption. We present a retrospective study which provides an overview of the assessment and preoperative work-up for patients diagnosed with cCMV and their cochlear implant (CI) outcomes.
METHOD: This was a retrospective case series study of all children with a confirmed diagnosis of cCMV who underwent cochlear implantation at St Thomas' Hospital from 2003 to 2015. Data were collected on the preoperative audiology, imaging findings, and neurological assessment. CI outcomes were measured using the Speech Intelligibility Rating (SIR), Category of Auditory Performance (CAP), and Infant-Toddler Meaningful Auditory Integration Scale (IT-MAIS).
RESULTS: Eleven patients underwent cochlear implantation, 45% had severe-to-profound hearing loss, and 55% had bilateral profound hearing loss. The mean age at initial assessment was 2.1 years (median 1.7, range 0.6-7.5) and the mean age of implantation was 4.0 years (median 2.5, range 0.9-11.8). The mean length of follow-up was 4.8 years (median 2.3, range 1.5-14). Six patients had bilateral simultaneous implantation (55%), four bilateral sequential (36%), and one unilateral (9%). Nine patients had white matter changes on magnetic resonance imaging, largely in the periventricular and cortical regions. Of the 11 patients, 4 (36%) had associated neurological comorbidities and 3 (27%) had additional neurocognitive developmental delay of varying severity. The majority of patients showed improvement in auditory outcomes. No statistically significant correlation was found between age of implantation, neurocognitive, and neurological comorbidities or length of follow-up and hearing outcomes.
CONCLUSION: While the overall outcomes were mixed, most children in our cohort were found to benefit from cochlear implantation. Our data also highlight the significant neurodevelopmental comorbidities associated with cCMV and their negative impact on CI outcomes. With the recent advances in medical treatment, this underlines the importance of multidisciplinary management of these patients.

Kasakura-Kimura N, Masuda M, Mutai H, Masuda S, Morimoto N, Ogahara N, Misawa H, Sakamoto H, Saito K, Matsunaga T.
WFS1 and GJB2 mutations in patients with bilateral low-frequency sensorineural hearing loss.
Laryngoscope. 2017 Sep;127(9):E324-E329. doi: 10.1002/lary.26528. Epub 2017 Mar8.
OBJECTIVE: Evaluating the prevalence of specific gene mutations associated with a certain audiometric configuration facilitates clinical assessment of patients with sensorineural hearing loss (SNHL). WFS1 is responsible for autosomal dominant nonsyndromic deafness 6/14/38 and is the most frequent genetic cause of low-frequency SNHL (LFSNHL); however, the exact prevalence of WFS1 mutations in LFSNHL is unknown. Therefore, we evaluated genetic mutations and clinical features in patients with nonsyndromic bilateral LFSNHL, focusing on the WFS1.
STUDY DESIGN: Retrospective case series from 2002 to 2013 at the National Hospital Organization Tokyo Medical Center and collaborating hospitals.
METHODS: WFS1, GJB2, and mitochondrial DNA mutation screening was carried out for 74 of 1,007 Japanese probands with bilateral LFSNHL.
RESULTS: WFS1 and GJB2 mutations were identified in eight of 74 cases (10.8%). Four cases had heterozygous WFS1 mutations; one case had a heterozygous WFS1 mutation and a heterozygous GJB2 mutation; and three cases had biallelic GJB2 mutations. Three cases with WFS1 mutations were sporadic; two of them were confirmed to be caused by a de novo mutation based on the genetic analysis of their parents. In the case with mutations in both WFS1 and GJB2, a de novo mutation of WFS1 was confirmed in the proband's mother by genetic screening of the mother's parents.
CONCLUSION: Genetic screening focusing on LFSNHL has not been conducted. The present study first revealed the prevalence of specific gene mutations. WFS1 autosomal dominant mutations were identified even in sporadic cases. Our results also suggested a mutational hotspot in WFS1.

Khairy MA, Abuelhamed WA, Ahmed RS, El Fouly HES, Elhawary IM.
Hearing loss among high-risk newborns admitted to a tertiary Neonatal Intensive Care Unit.
J Matern Fetal Neonatal Med. 2017 May 22:1-6. doi: 10.1080/14767058.2017.1326902. [Epub ahead of print]
PURPOSE: The aim of this work is to identify the most significant risk factors for hearing impairment in high risk neonates hospitalized at our Neonatal Intensive Care Unit (NICU) and to assess the sensitivity of hearing screening tests.
METHODS: This study involved 260 neonates admitted to a tertiary NICU; they were classified into two groups; 150 preterm and 110 full terms with risk factors for hearing loss. The hearing screening tests performed were transient evoked otoacoustic emissions (TEOAEs) and the automated auditory brainstem response (AABR).
RESULTS: Forty-eight preterm neonates (32%) and 30 full term neonates (27.3%) had pathological AABR. In preterm group, mechanical ventilation more than five days, sepsis, usage of aminoglycosides, loop diuretics, vancomycin alone or in combination with aminoglycosides and prolonged duration of admission were considered risk factors of hearing affection whereas in full term group mechanical ventilation more than five days was the risk factor of hearing affection ($p<.05$).
CONCLUSIONS: The prevalence of hearing loss is highest among high risk neonates and TEOAE and AABR were found to be reliable screening tools. Use of ototoxic drugs and mechanical ventilation for more than five days were significant risk factors for hearing loss in our study population.

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.
Pediatr Neonato. 2017 Jun;58(3):236-244. doi: 10.1016/j.pedneo.2016.05.003. Epub 2016 Aug 9.
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 chorioamnionitis 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.
CONCLUSION: 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.

Kim SH, Lim JH, Han JJ, Jin YJ, Kim SK, Kim JY, Song JJ, Choi BY, Koo JW.
Outcomes and limitations of hospital-based newborn hearing screening.
Int J Pediatr Otorhinolaryngol. 2017 Jul;98:53-58. doi:10.1016/j.ijporl.2017.04.030. Epub 2017 Apr 20.
OBJECTIVES: Globally, newborn hearing screening (NHS) is variably incorporated into national healthcare systems. The authors reviewed the set-up and evolution process of a hospital-based NHS program in South Korea, where screening costs for low-income families are paid by the National Health Authority.
METHODS: The NHS process for 13805 newborns delivered in a tertiary referral center of South Korea from 2005 through 2014 was reviewed. Hearing screening was conducted using automated auditory brainstem response (AABR); hearing loss was confirmed by auditory brainstem response for newborns who did not pass the screening test.
RESULTS: The mean screening rate for hearing loss was 53.6% (7403 of 13805 newborns), which plateaued at 79.6% over time. Of the 14806 ears (7403 newborns), 1030 (7.0%) were assessed as “refer” on the first AABR, with 204 (1.4%) being assessed as “refer” on the second AABR. In hearing confirmation tests, 74 infants (1.0% of 7403 newborns) were diagnosed with hearing loss, including 13 infants (0.2%) with bilateral moderate to profound sensorineural hearing loss (SNHL). Hearing rehabilitation with long-term follow-up was confirmed in 11 infants.
CONCLUSIONS: In this hospital-based NHS program, the screening rate plateaued at □50% when the National Health Authority was not involved, but increased to □70% when the cost for low-income families was covered by the government. Among infants needing active hearing rehabilitation due to bilateral moderate to profound SNHL, 15% were lost to follow-up. These results demonstrate the need for a universal, mandatory NHS program to systematically register hearing-impaired infants within the government-sponsored public healthcare system.

Komínek P, Chrobok V, Zeleník K, Dršata J.
[Newborn hearing screening - importance, current state in the Czech Republic]. [Article in Czech]
Cas Lek Cesk. 2017 Summer;156(4):173-177.
The importance of early detection of hearing impairment in newborns and children and the early rehabilitation of hearing disorder with hearing aid or cochlear implant was demonstrated in a number of papers. As a result, newborn hearing screening was introduced in many countries around the world. The incidence of congenital hearing impairment has been underestimated for a long time, empirically determined incidence was 1:1000 neonates. Thanks to newborn hearing screening was revealed that incidence of congenital hearing impairment is 3 times higher. One out of 300 newborns has profound hearing loss (deafness) and 1 in 300 newborns has mild to moderate hearing loss. Moreover, in 1 out of 300 children hearing impairment would develop before the age of 18. In the Czech Republic, the screening of all newborns is still systematical and multilevel conducted in only three regions (Moravskoslezský, Královéhradecký, Pardubický). In these regions, statistics as well as assessments of individual stages of screening are carried out. In other regions, records of the number of screened children, number of rescreened children and detailed statistics of hearing impairment is missing. The authors summarize the basic information about the importance of screening, the history of screening and its organization. Provided information is based on experience with the gradual introduction of screening over recent years in the regions in which they work. Newborn hearing screening should be organized in several stages (1 - screening at maternity hospitals, 2 - rescreening on collaborating otolaryngology/phoniatric workplaces and 3 - detailed hearing examination in centers in children whose screening was negative). The authors focus on problems related to the organization of screening and offer practical advice (e.g. implementation of screening coordinators). They consider it is essential to have statistical evidence of examination at all levels. Introducing of newborn hearing screening is not a short-term task but a long-term (many years) challenge.

Koyano S, Morioka I, Oka A, Moriuchi H, Asano K, Ito Y, Yoshikawa T, Yamada H, Suzutani T, Inoue N; Japanese Congenital Cytomegalovirus Study Group.
More than two years follow-up of infants with congenital cytomegalovirus infection in Japan.
Pediatr Int. 2017 Oct 15. doi:10.1111/ped.13433. [Epub ahead of print]
BACKGROUND: The aim of the study was to evaluate outcomes of congenital cytomegalovirus (CMV) cases identified by our urine-filter screening assay after follow-up for more than 2 years, and to observe the clinical outcomes after anti-CMV therapies.
METHODS: Sixty of the 72 congenital CMV cases were enrolled. Forty-three cases asymptomatic at birth, 7 cases symptomatic at birth but untreated with anti-CMV drugs, and 10 symptomatic cases treated with anti-CMV drugs were clinically observed for more than 2 years.
RESULTS: Among the 43 asymptomatic cases, 3 cases developed hearing loss or language disabilities for which association with congenital CMV has been repeatedly reported and 2 cases demonstrated neurological sequelae of which etiology was unclear, indicating that the rate of CMV-associated late-onset sequelae was 7~12%. All 7 symptomatic infants without treatment developed sequelae, while 3 of the 10 treated cases were free from any sequelae.
CONCLUSIONS: The rate of late-onset sequelae observed in Japan is similar to that reported for cases in the US and European countries. The treatment of symptomatic cases with antiviral agents results in favorable clinical outcomes. Thus, newborn screening of congenital CMV infection by the collection of urine on filter paper is warranted.

Kubba H, Smyth A, Wong SC, Mason A.
Ear health and hearing surveillance in girls and women with Turner’s syndrome: recommendations from the Turner’s Syndrome Support Society.
Clin Otolaryngol. 2017 Jun;42(3):503-507. doi: 10.1111/coa.12750. Epub 2016 Sep28.

BACKGROUND: Turner's syndrome (TS) is a common chromosomal disorder, affecting one in 2000 newborn girls, in which part or all of one X chromosome is missing. Ear and hearing problems are very common in girls and women with TS. The aim of this review was to review the published literature to suggest recommendations for otological health surveillance.
METHOD: A keyword search of Ovid Medline was performed for published literature on the subject and evidence rated according to the GRADE criteria.
RESULTS: Middle ear disorders are very common and persistent in girls and women with TS as are progressive sensorineural hearing loss and balance disorders.
CONCLUSIONS: Otolaryngologists should be aware of the high prevalence and challenging nature of all forms of ear disease in individuals with TS. Early intervention may offer benefits to health and education, and we advocate routine lifelong annual hearing screening in this group.

Kumar A, Gupta SC, Sinha VR.

Universal hearing screening in newborns using otoacoustic emissions and brainstem evoked response in Eastern Uttar Pradesh.

Indian J Otolaryngol Head Neck Surg. 2017 Sep;69(3):296-299. doi: 10.1007/s12070-017-1081-x. Epub 2017 Feb 3.

The objectives were to determine the incidence of hearing impairment in a standardized population of neonates and to determine the significance of association of epidemiological and risk factors with neonatal hearing loss. A cohort of 600 newborns was selected for study and divided into two groups-525 in 'No Risk' group and remaining 75 in 'At Risk' group. The study protocol was carried out in three steps: (a) Screening of Hearing Loss with TOAE, done from 36 h after birth to 28 days of life, (b) Re-screening of hearing loss in newborns (of 4-12 weeks of age), who were tested positive for hearing loss in the first screening, done with DPOAE, (c) Confirmation of hearing loss with BERA, in those who were tested positive in both the first and second screening. In the study the incidence of hearing impairment in 600 infants screened was 6.67 per 1000 screened; 3.81 per 1000 screened in the Not at Risk group and 26.67 per 1000 screened in At Risk group. In At Risk group, admitted to the NICU, severe birth asphyxia and hyperbilirubinemia were found to be major risk factors. Loss to follow up was more in Not at Risk group and False Positive cases with TEOAE were more than DPOAE. BERA was found to be must for confirmation of hearing loss. Neonatal Hearing Screening of only At Risk population is likely to miss some hearing loss. Universal Hearing Screening should be the preferred strategy. Good follow up in the 'At Risk' group suggests that initial interventions in NICU had sensitized the parents for the possibility of hearing loss. This study recommends the introduction of two stage neonatal screening-rescreening protocol, using OAE and BERA, in the country in phased manner.

Li JN, Chen S, Zhai L, Han DY, Eshraghi AA, Feng Y, Yang SM, Liu XZ.

The advances in hearing rehabilitation and cochlear implants in China.

Ear Hear. 2017 May 3. doi: 10.1097/AUD.0000000000000441. [Epub ahead of print]

Hearing loss (HL) is a common sensory impairment in humans, with significant economic and social impacts. With nearly 20% of the world's population, China has focused on economic development and health awareness to improve the care for its hearing-impaired population. Recently, the Chinese government has initiated national programs such as the China Disabled Persons Federation to fund prevention, treatment, and rehabilitation of hearing impairment. Newborn hearing screening and auditory rehabilitation programs in China have expanded exponentially with government support. While facing many challenges and overcoming obstacles, cochlear implantation (CI) programs in China have also experienced considerable growth. This review discusses the implementation of CI programs for HL in China and presents current HL data including epidemiology, newborn hearing screening, and determination of genetic etiologies. Sharing the experience in Chinese auditory rehabilitation and CI programs will shine a light on the developmental pathway of healthcare infrastructure to meet emerging needs of the hearing-impaired population in other developing countries.

Mack I, Burckhardt MA, Heininger U, Prüfer F, Schulzke S, Wellmann S.

Symptomatic congenital cytomegalovirus infection in children of seropositive women.

Front Pediatr. 2017 Jun 9;5:134. doi: 10.3389/fped.2017.00134. eCollection 2017.

Cytomegalovirus (CMV) is the most frequent congenital virus infection worldwide. The risk of congenital CMV (cCMV) transmission is highest in seronegative women who acquire primary CMV infection during pregnancy. A growing body of evidence indicates that secondary CMV infections in pregnant women with preconceptual immunity (either through reactivation of latent virus or re-infection with a new strain of CMV) contribute to a much greater proportion of symptomatic cCMV than was previously thought. Here, we describe a case of symptomatic cCMV infection in the newborn of a woman with proven immunity prior to pregnancy. Diagnosis was confirmed by CMV PCR from amniotic fluid and fetal MR imaging. The newborn presented with typical cCMV symptoms including jaundice, hepatosplenomegaly, cholestasis, petechiae, small head circumference, and sensorineural hearing loss, the most common neurologic sequela. CMV was detected in infant blood and urine by PCR, and intravenous ganciclovir was initiated and continued orally for 6 weeks totally. Apart from persisting right-sided deafness, the child exhibited normal neurological development up through the last follow-up at 4.5 years. To date, the most effective strategy to prevent vertical CMV transmission is hygiene counseling for women of childbearing age, which, in our case, and in concordance with recent literature, applies to seronegative, as well as seropositive, women. Once an expecting mother shows seroconversion or signs of an active CMV infection, there are no established procedures to reduce the risk of transmission, or therapeutic options for the fetus with signs of infection. After birth, symptomatic infants can be treated with ganciclovir to inhibit viral replication and improve hearing ability and neurodevelopmental outcome. A comprehensive review of the literature, including our case study, reveals the most current and significant diagnostic and treatment options available. In conclusion, the triad of maternal hygiene counseling, postnatal hearing screening of all newborns, followed by CMV PCR in symptomatic infants, and antiviral therapy of infants with symptomatic cCMV provides an outline of best practice to reduce the burden of CMV transmission sequelae.

Martínez-Cruz CF, Poblano A, García-Alonso Themann P.

Changes in tonal audiometry in children with progressive sensorineural hearing loss and history of Neonatal Intensive Care Unit discharge. A 20 year long-term follow-up.

Int J Pediatr Otorhinolaryngol. 2017 Oct;101:235-240. doi: 10.1016/j.ijporl.2017.08.022. Epub 2017 Aug 29.

OBJECTIVE: Newborns from Neonatal intensive care units (NICU) are at high-risk for sensorineural hearing loss (SNHL) a follow-up is needed for early diagnosis and intervention. Our objective here was to describe the features and changes of SNHL at different periods during a follow-up of almost 20 years.
METHODS: Risk factors for SNHL during development were analyzed. The audiological examination included: Brainstem auditory evoked potentials (BAEP), and Transient evoked otoacoustic emissions (TEOAE). At birth; tonal audiometry (between 125 and 8000 Hz), and tympanometry were performed at 5, 10, 15, and 20 years of age.
RESULTS: Sixty-five percent of cases presented bilateral absence of BAEP. At 5 years of age, the most frequent SNHL level was severe (42.5%), followed by moderate (22.5%), and profound level (20%), in all cases, the SNHL was symmetrical with a predominance of lesion for the high frequencies. Exchange transfusion was associated with a higher degree of SNHL (OR = 6.00, CI = 1.11-32.28, $p < 0.02$). In 55%, SNHL remained stable, but in 40% of the cases it was progressive. At the end of the study six cases with moderate loss progressed to the severe level and seven cases with severe level progressed to profound.
CONCLUSIONS: Forty percent of infants with SNHL discharged from NICU may present a progression in the hearing loss. Exchange transfusion was associated with a higher degree of SNHL. NICU graduates with SNHL merit a long-term audiological follow-up throughout their lifespan.

Matulat P, Stroe S, Am Zehnhoff-Dinnesen A.
Transregional tracking in newborn hearing screening.
HNO. 2017 Sep 22. doi: 10.1007/s00106-017-0424-y. [Epub ahead of print]
[Article in German]
BACKGROUND AND OBJECTIVE: When patients in the universal newborn hearing screening program move from one geographical area to another between initial screening and medical follow-up, the responsibility for their tracking also moves from one screening center to another. As a result, these patients are lost to follow-up according to the center which had initial responsibility. In cooperation with the Association of German Hearing Screening Centers ("Verband Deutscher Hörscreening-Zentralen e. V.," VDHZ) as an offer to the developers of tracking software, a concept for nationwide tracking including a reference implementation and evaluation is described.
METHODS: On the basis of error analysis of real screening data, techniques for preprocessing data, the technical background of the interface, and details regarding integration of the interface into tracking software are presented. Data from a stress test are shown.
RESULTS: In a simulation stress test with six hearing screening centers and 54,551 children, all requests were answered within an average response time of 637 ms (standard deviation, SD = 266 ms; median 613 ms). Anonymized surnames (n = 675/1.24%) and duplicate entries in the database (n = 49/0.01%) were detected.
CONCLUSION: A transregional tracking procedure using heterogeneous tracking software is possible without the use of a standardized screening ID. The presented approach seems conceptually and technically suitable.

Meyer L, Sharon B, Huang TC, Meyer AC, Gravel KE, Schimmenti LA, Swanson EC, Herd HE, Hernandez-Alvarado N, Coverstone KR, McCann M, Schleiss MR.
Analysis of archived newborn dried blood spots (DBS) identifies congenital cytomegalovirus as a major cause of unexplained pediatric sensorineural hearing loss.
Am J Otolaryngol. 2017 Sep - Oct;38(5):565-570. doi:10.1016/j.amjoto.2017.06.002. Epub 2017 Jun 7.
PURPOSE: Congenital cytomegalovirus (cCMV) infection is the most common non-genetic cause of sensorineural hearing loss (SNHL). However, accurate diagnosis of cCMV as the etiology of SNHL is problematic beyond the neonatal period. This study therefore examined whether cCMV infection could be identified retrospectively in children presenting with unexplained SNHL to a multidisciplinary diagnostic outpatient otolaryngology clinic at an academic medical center in Minnesota.
METHODS: Over a 4-year period, 57 patients with an age range of 3months to 10years with unexplained SNHL were recruited to participate in this study. Informed consent was obtained to test the archived dried blood spots (DBS) of these patients for cCMV infection by real-time PCR, targeting a highly conserved region of the CMV UL83 gene. RESULTS were normalized to recovery of an NRAS gene control. Chart review was conducted to identify subjects who underwent genetic testing and/or neurodiagnostic imaging to investigate possible genetic, syndromic, or anatomical causes of SNHL.
RESULTS: In total, 15 of the 57 children with unexplained SNHL tested positive for CMV DNA in their DBS (26%). A mean viral load of 8.3×10(4) (±4.1×10(4)) [range, 1×10(3)-6×10(5)] copies/μg DNA was observed in subjects retrospectively diagnosed with cCMV. No statistically significant correlation was found between viral load and SNHL severity.
CONCLUSIONS: A retrospective DBS analysis demonstrated that 26% of patients presenting with unexplained SNHL in childhood had cCMV. DBS testing is useful in the retrospective diagnosis of cCMV, and may provide definitive diagnostic information about the etiology of SNHL.

Müller J, Fechner H, Köhn A, Rißmann A.
Newborn hearing screening – Results of a parental survey in Saxony-Anhalt.
[Article in German]
Gesundheitswesen. 2017 May;79(5):388-393. doi: 10.1055/s-0035-1549969. Epub 2015 Jun 25.
BACKGROUND: In recent years quality assurance has become an essential part of today's health-care system in the wake of the modern patient-oriented quality management. With the statutory introduction of newborn hearing screening (NHS) in 2009, a quality assurance of these early detection methods has become necessary. The aim of the study was to determine patient satisfaction in relation to the NHS in Saxony-Anhalt.
PATIENTS/METHODS: During the period from November 2013 to April 2014, 394 parents were retrospectively interviewed about their experiences and expectations in relation to the NHS, using a standardised questionnaire. In total, 21 child care centres and 6 paediatric primary care centres from all over Saxony-Anhalt were involved.
RESULTS: It turns out that the majority of parents are satisfied with the NHS and 97.7% are in favour of the offer of an NHS. Of the surveyed parents, 69.3% felt the information as sufficient. However, only 66.2% of parents took a closer look at the leaflet issued by the G-BA. In addition, 17.7% of respondents are dissatisfied with the professional competence of the examining staff.
CONCLUSION: The study shows that the general attitude among parents towards newborn hearing screening was very positive. They felt reassured by it although there are some aspects still open to criticism.

Netten AP, Rieffe C, Ketelaar L, Soede W, Gadow KD, Frijns JHM.
Terrible twos or early signs of psychopathology? Developmental patterns in early identified preschoolers with cochlear implants compared with hearing controls.
Ear Hear. 2017 Oct 4. doi: 10.1097/AUD.0000000000000500. [Epub ahead of print]

OBJECTIVE: Cochlear implants (CIs) have dramatically improved the lives of children who are deaf or hard of hearing; however, little is known about its implications for preventing the development of psychiatric symptoms in this at-risk population. This is the first longitudinal study to examine the early manifestation of emotional and behavioral disorders and associated risk and protective factors in early identified preschoolers with CIs compared with hearing peers.
DESIGN: Participants were 74 children with CIs and 190 hearing controls between ages 1 and 5 years (mean age, 3.8 years). Hearing loss was detected using the Newborn Hearing Screening in The Netherlands and Flanders. Parents completed the Early Childhood Inventory-4, a well-validated measure, to evaluate the symptoms of DSM-IV-defined psychiatric disorders, during three consecutive years. Language scores were derived from each child's medical notes.
RESULTS: Children with CIs and hearing controls evidenced comparable levels of disruptive behavior and anxiety/depression (which increased with age in both groups). Greater proficiency in language skills was associated with lower levels of psychopathology. Early CI and longer duration of CI use resulted in better language development. In turn, higher early language skills served as a protective factor against the development of disruptive behavior symptoms.
CONCLUSIONS: This longitudinal study uniquely shows that improvement in language skills mitigates the development of early signs of psychopathology. Early identification of hearing loss and CIs help children improve their language skills.

Pan J, Xu P, Tang W, Cui Z, Feng M, Wang C.
Mutation analysis of common GJB2, SCL26A4 and 12S rRNA genes among 380 deafness patients in northern China.
Int J Pediatr Otorhinolaryngol. 2017 Jul;98:39-42. doi: 10.1016/j.ijporl.2017.04.018. Epub 2017 Apr 12.
OBJECTIVES: The molecular etiology of nonsyndromic deafness in Chinese population has not been investigated systematically, our study is aim to investigate the molecular etiology of nonsyndromic deafness patients from Northern China (Heilongjiang province), in order to provide genetic test and counseling to families.
METHODS: 380 unrelated patients with hearing loss who attended to the Department of Otolaryngology, The Fourth Affiliated Hospital of Harbin Medical University were enrolled to our study. All patients were diagnosed with nonsyndromic deafness by audiologic evaluation, 202 normal-hearing individuals were taken as controls. Mutations in three common deafness-causing genes (GJB2, SLC26A4 and 12S rRNA) were screened by direct sequencing.
RESULTS: Mutations (homozygote or compound heterozygote) in GJB2 accounted for 8.9% (34/380) of the patients, mutations in SLC26A4 accounted for 10.0% (38/380) of the patients screened. Only one case was found to carry 12S rRNA 1555A > G (1/380, 0.26%). Five types of mutations in GJB2 were identified, GJB2 235delC was the most prevalent mutation in our patient group (76/380, 20.0%), followed by 299-300delAT with a frequency of 7.4% (28/380). Two types of mutations in SLC26A4 were detected in our patient group (IVS7-2A > G and 2168A > G). IVS7-2A > G was identified in 27 patients (27/380, 7.1%) and 2168A > G was identified in 14 patients (14/380, 3.7%).
CONCLUSIONS: Our results demonstrate that 19.2% patients with nonsyndromic deafness were caused by mutations in three common deafness genes (GJB2, SLC26A4 and 12S rRNA) in our northern China patient group. GJB2 235delC was the most prevalent mutation, same as in the most Asian populations. These data enrich the database of deafness mutations and provide the standard for clinical diagnose, treatment and genetic counseling in Northern China population.

Paul A, Prasad C, Kamath SS, Dalwai S, C Nair MK, Pagarkar W.
Consensus statement of the Indian Academy of Pediatrics on Newborn Hearing Screening.
Indian Pediatr. 2017 Aug 15;54(8):647-651. Epub 2017 Jun 4.
JUSTIFICATION: Hearing impairment is one of the most critical sensory impairments with significant social and psychological consequences. Evidence-based, standardized national guidelines are needed for professionals to screen for hearing impairment during the neonatal period.
PROCESS: The meeting on formulation of national consensus guidelines on developmental disorders was organized by Indian Academy of Pediatrics in Mumbai, on 18th and 19th December, 2015. The invited experts included Pediatricians, Developmental Pediatricians, Pediatric Neurologists and Clinical Psychologists. The participants framed guidelines after extensive discussions.
OBJECTIVE: To provide guidelines on newborn hearing screening in India.
RECOMMENDATIONS: The first screening should be conducted before the neonate's discharge from the hospital - if it 'fails', then it should be repeated after four weeks, or at first immunization visit. If it 'fails' again, then Auditory Brainstem Response (ABR) audiometry should be conducted. All babies admitted to intensive care unit should be screened via ABR. All babies with abnormal ABR should undergo detailed evaluation, hearing aid fitting and auditory rehabilitation, before six months of age. The goal is to screen newborn babies before one month of age, diagnose hearing loss before three months of age and start intervention before six months of age.

Pimperton H, Kreppner J, Mahon M, Stevenson J, Terlektsi E, Worsfold S, Yuen HM, Kennedy CR.
Language outcomes in deaf or hard of hearing teenagers who are spoken language users: Effects of universal newborn hearing screening and early confirmation.
Ear Hear. 2017 Sep/Oct;38(5):598-610. doi: 10.1097/AUD.0000000000000434.
OBJECTIVES: This study aimed to examine whether (a) exposure to universal newborn hearing screening (UNHS) and b) early confirmation of hearing loss were associated with benefits to expressive and receptive language outcomes in the teenage years for a cohort of spoken language users. It also aimed to determine whether either of these two variables was associated with benefits to relative language gain from middle childhood to adolescence within this cohort.
DESIGN: The participants were drawn from a prospective cohort study of a population sample of children with bilateral permanent childhood hearing loss, who varied in their exposure to UNHS and who had previously had their language skills assessed at 6-10 years. Sixty deaf or hard of hearing teenagers who were spoken language users and a comparison group of 38 teenagers with normal hearing completed standardized measures of their receptive and expressive language ability at 13-19 years.
RESULTS: Teenagers exposed to UNHS did not show significantly better expressive (adjusted mean difference, 0.40; 95% confidence interval [CI], -0.26 to 1.05; d = 0.32) or receptive (adjusted mean difference, 0.68; 95% CI, -0.56 to 1.93; d = 0.28) language skills than those who were not. Those who had their hearing loss confirmed by 9 months of age did not show significantly better expressive (adjusted mean difference, 0.43; 95% CI, -0.20 to 1.05; d = 0.35) or receptive (adjusted mean difference, 0.95; 95% CI, -0.22 to 2.11; d = 0.42) language skills than those who had it confirmed later. In all cases, effect sizes were of small size and in favor of those exposed to UNHS or confirmed by 9 months. Subgroup analysis indicated larger beneficial effects of early confirmation for those deaf or hard of hearing teenagers without cochlear implants (N = 48; 80% of the sample), and these benefits were significant in the case of receptive language outcomes (adjusted mean difference, 1.55; 95% CI, 0.38 to 2.71; d = 0.78). Exposure to UNHS did not account for significant unique variance in any of the three language scores at 13-19 years beyond that accounted for by existing

language scores at 6-10 years. Early confirmation accounted for significant unique variance in the expressive language information score at 13-19 years after adjusting for the corresponding score at 6-10 years (R change = 0.08, p = 0.03).
CONCLUSIONS: This study found that while adolescent language scores were higher for deaf or hard of hearing teenagers exposed to UNHS and those who had their hearing loss confirmed by 9 months, these group differences were not significant within the whole sample. There was some evidence of a beneficial effect of early confirmation of hearing loss on relative expressive language gain from childhood to adolescence. Further examination of the effect of these variables on adolescent language outcomes in other cohorts would be valuable.

Quevedo ALA, Leotti VB, Goulart BNG.
Analysis of prevalence of self-reported hearing loss and associated factors: Primary versus proxy informant. [Article in Portuguese]
Cad Saude Publica. 2017 Jul 3;33(6):e0076216. doi: 10.1590/0102-311X00076216.
The objective was to evaluate differences between prevalence rates for self-reported hearing loss and associated factors, obtained from responses by primary and proxy informants in a Population-Based Study on Human Communication Disorders (DCH-POP in Portuguese). This was a study on epidemiological methods using data from a cross-sectional household survey with a sample of 1,253 individuals from Southern Brazil. To verify differences between prevalence rates comparing primary informants and proxy informants, we used the chi-square or Fisher's exact test for categorical variables and Mann-Whitney for continuous variables. The log-binomial model was adjusted for hearing loss as the dependent variable, considering three datasets: the entire sample, only primary informants, and only proxy informants, estimating association by prevalence ratios. In the final models, only the independent variables age and dizziness were associated with hearing loss, independently of the dataset that was used. Proxy informants generally underestimated the prevalence rates for the target outcomes, when compared to primary informants.

Ricalde RR, Chiong CM, Labra PJP.
Current assessment of newborn hearing screening protocols.
Curr Opin Otolaryngol Head Neck Surg. 2017 Oct;25(5):370-377.doi:10.1097/ MOO.0000000000000389.
PURPOSE OF REVIEW: The objective of this article is to assess current newborn hearing screening protocols. We will focus on technologies or modalities used, protocol steps, training of screeners, timing of first screen, and loss to follow-up. A summary of program reports focusing on protocols from Greece, China, South Africa, France, Spain, South Korea, Denmark, Italy, Turkey, Taiwan, South Korea, Poland and Iran as they are recently reported will also be presented.
RECENT FINDINGS: Community-based hearing screening programs in South Africa and efforts in the Asian region are being reported. The use of automated auditory brainstem response and staged procedures are gaining popularity because of low refer rates. However, follow-up issues remain a problem. The importance of having trained nonprofessional screeners and an efficient database is becoming more evident as the number of newborns screened for hearing loss increase each year.
SUMMARY: There are many reported protocols using different technologies, involving several stages, implemented in different settings which should not confuse but rather guide stakeholders so that programs may attain certain benchmarks and ultimately help the hard-at-hearing child in achieving his or her full potential.

Robertson VS, von Hapsburg D, Hay JS.
The effect of hearing loss on novel word learning in infant- and adult-directed speech.
Ear Hear. 2017 Jun 22. doi: 10.1097/AUD.0000000000000455. [Epub ahead of print]
OBJECTIVES: Relatively little is known about how young children with hearing impairment (HI) learn novel words in infant- and adult-directed speech (ADS). Infant-directed speech (IDS) supports word learning in typically developing infants relative to ADS. This study examined how children with normal hearing (NH) and children with HI learn novel words in IDS and ADS. It was predicted that IDS would support novel word learning in both groups of children. In addition, children with HI were expected to be less proficient word learners as compared with their NH peers.
DESIGN: A looking-while-listening paradigm was used to measure novel word learning in 16 children with sensorineural HI (age range 23.2 to 42.1 months) who wore either bilateral hearing aids (n = 10) or bilateral cochlear implants (n = 6) and 16 children with NH (age range 23.1 to 42.1 months) who were matched for gender, chronological age, and maternal education level. Two measures of word learning were assessed (accuracy and reaction time). Each child participated in two experiments approximately 1 week apart, one in IDS and one in ADS.
RESULTS: Both groups successfully learned the novel words in both speech type conditions, as evidenced by children looking at the correct picture significantly above chance. As a group, children with NH outperformed children with HI in the novel word learning task; however, there were no significant differences between performance on IDS versus ADS. More fine-grained time course analyses revealed that children with HI, and particularly children who use hearing aids, had more difficulty learning novel words in ADS, compared with children with NH.
CONCLUSIONS: The pattern of results observed in the children with HI suggests that they may need extended support from clinicians and caregivers, through the use of IDS, during novel word learning. Future research should continue to focus on understanding the factors (e.g., device type and use, age of intervention, audibility, acoustic characteristics of input, etc.) that may influence word learning in children with HI in both IDS and ADS.

Rütten H, Rissmann A, Brett B, Costa SD, Doßow B, Färber J, Fest S, Fritzsch C, Lux A, Päge I, Spillner C, Redlich A.
Congenital cytomegalovirus infection in Central Germany: An underestimated risk.
Arch Gynecol Obstet. 2017 Aug;296(2):231-240. doi: 10.1007/s00404-017-4435-4. Epub 2017 Jun 17.
PURPOSE: This is the first study to determine the cytomegalovirus (CMV) seronegativity rate for women of childbearing age in Saxony-Anhalt and to determine the prevalence of clinically relevant congenital CMV (cCMV) infection in Central Germany, because there are no valid data available.
METHODS: The retrospective study was undertaken between January 2005 and December 2015. For the first time in Germany, the following seven data sources were used to analyze the prevalence of clinically relevant cCMV infection and the rate of CMV seronegative women of childbearing age: CMV Screening in maternity unit, University Women's Hospital, Social Paediatrics Centre (SPC), Malformation Monitoring Centre (MMC), Newborn Hearing Screening (NHS), Neonatal Intensive Care Unit (NICU), and In-house Doctor Department. Key parameters were anti-CMV IgG and IgM, CMV PCR of urine, and clinically relevant symptoms caused by CMV.
RESULTS: Between 46 and 52% of women of childbearing age were CMV seronegative. The prevalence of clinically relevant cCMV infection was between 0.008 and 0.04%.
CONCLUSIONS: The CMV seronegativity rate of women of childbearing age was confirmed to be in the middle range of estimated data from other sources in Germany. Data from the NICU, SPC, NHS, and MMC show the prevalence of clinically relevant cCMV infection. The risk of all cCMV

infections is underestimated. Thus, the true prevalence of clinically relevant and subclinical cCMV infections is >0.04%.

Sabbag JC, Lacerda ABM.

Neonatal hearing screening in primary health care and family health care. [Article in English, Portuguese]

Codas. 2017 Aug 10;29(4):e20160102. doi: 10.1590/2317-1782/20172016102.

PURPOSE: The Universal Newborn Hearing Screening (UNHS) looks for early diagnosis and rehabilitation of newborns at risk or not of hearing impairment. The purpose is analyze the flow of Universal Newborn Hearing Screening in the family health care strategy unit through the tracking and monitoring of children.
METHODS: This is a quantitative and retrospective study. The trace begins with the third copy of the Live Newborn Declaration, filled in at the maternity ward. An interview with parents and guardians was made by a community agent at the Health Unit or at the home of the newborn. Monitoring was conducted by live birth declaration and the information collected by the interviewer from maternal and child health booklet and the follow-up at high complexity services.
RESULTS: The sample was made up of 50 neonates. 52% were between 30 and 89 days and 54% were male. 12% of newborns presented a risk factor for hearing loss and the neonatal screening was performed in 86% of cases. Hearing health measures show integrality in hearing impairment care at the basic health unit to high
complexity hospital.
CONCLUSION: The flow of care for newborn hearing screening is in agreement with the child health care guidelines in Curitiba, however, it is not yet universal. In conclusion, the participation of the family health strategy unit in the tracking and monitoring of children submitted to the Universal newborn hearing screening program is feasible and recommended.

Sachdeva K, Sao T.

Outcomes of newborn hearing screening program: A hospital based study.

Indian J Otolaryngol Head Neck Surg. 2017 Jun;69(2):194-198. doi: 10.1007/s12070-017-1062-0. Epub 2017 Jan 17.

Hearing loss is hidden disability and second most common congenital pathology. Prevention, early identification and early intervention of hearing loss can prevent further disability in development of speech, language, cognition and other developmental domains. The prevalence of congenital hearing loss has been estimated to be 1.2-5.7 per thousand in neonates. In these contexts, the aim of study was to determine outcomes of neonates hearing screening program in Hospital. It is a clinical cross-sectional study which was conducted in tertiary care centre from 8th July, 2015 to 31th May, 2016. Total no of 2254 cases were screened. Details case history including high risk register, Pediatric Audiometry, Otoacoustic Emission tests were performed followed brainstem evoked response audiometry. The Prevalence of hearing loss among high risk babies confirmation by BERA was 8.8% per 1000 babies and 16 cases were recommended for Cochlear Implant. The screening protocol with objective test i.e. Distortion Product Otoacoustic Emission and confirmation by Brainstem Evoked Response Audiometry is very useful tool in early identification of congenital hearing loss in neonates. Hence, the results of this study will be used to initiate universal newborn hearing screening in other hospitals. Moreover, this study highlights the relevance of neonatal hearing screening in other states of India and country where this screening is not performed routinely in all hospitals and creating awareness to identify neonatal risk factors associated with hearing loss and understand the importance of early identification and early intervention and among health care professionals.

Saki N, Bayat A, Hoseinabadi R, Nikakhlagh S, Karimi M, Dashti R.

Universal newborn hearing screening in southwestern Iran.

Int J Pediatr Otorhinolaryngol. 2017 Jun;97:89-92. doi: 10.1016/j.ijporl.2017.03.038. Epub 2017 Mar 31.

OBJECTIVES: The implementation of Neonatal Hearing Screening (NHS) program is still at the preliminary stage particularly in developing countries despite the burden of permanent congenital and early-onset hearing impairment. The purpose of this study was to report results for universal newborn NHS in a cohort of children born in the southwestern region of Iran, as part of a national screening program set up by the Iranian National Health System.
METHODS: During this cross-sectional study, which took place between March 2013 and April 2016, healthy newborns were screened using transient evoked optoacoustic emissions (TEOAEs) and automated auditory brainstem responses (AABRs) methods at several points in time as early as possible after birth. Screening followed a two-stage strategy and newborns referred after the second-stage screening were scheduled for diagnostic evaluation.
RESULTS: A total of 92,521 newborns were screened in the urban (n = 67,780) and rural (n = 24,741) regions. Hearing impairment was confirmed in 223 (2.41 per 1000) newborns. One hundred forty-one (1.52 per 1000) of these newborns were affected bilaterally. More than 87% of these infants (195/223) showed a sensorineural hearing loss, while the defect was found to be conductive in 12 cases ($P < 0.001$). Of the 223 cases with hearing loss, 28 (12.5%) infants had auditory neuropathy. The majority of the infants, in both urban and rural regions, showed severe hearing impairment. We did not observe any significant difference among the incidences associated with gender ($p = 0.29$).
CONCLUSION: Our results demonstrated that universal newborn hearing screening program is an adequate program for southwestern of Iran with high coverage, low referral rate, and good follow-up rate.

Sanyelbhaa H, Kabel A, Abo El-Naga HAE, Sanyelbhaa A, Salem H.

The risk ratio for development of hereditary sensorineural hearing loss in consanguineous marriage offspring.

Int J Pediatr Otorhinolaryngol. 2017 Oct;101:7-10. doi: 10.1016/j.ijporl.2017.07.020. Epub 2017 Jul 19.

OBJECTIVES: This study aims to define the relative risk of development of hearing loss in offspring of consanguineous marriages.
MATERIALS AND METHODS: This is a retrospective case-control study conducted in a tertiary referral center in Jeddah, KSA. The study group included 1600 probands (848 males, 752 females), with age range 0.5-12 years (6.6 ± 3.6). The study group comprised of two equal, age and sex matched subgroups; Hearing Loss (HL) group and Normal Hearing (NH) group. The children included in the HL group should have idiopathic or non syndromic genetic sensorineural hearing loss.
RESULTS: The HL Group comprised 800 children with variable degrees of sensorineural hearing loss. Profound and severe degrees of hearing loss were the most prevalent degrees ($P < 0.05\%$). The prevalence of consanguineous marriage offspring in the NH group was 42.5%, while in the HL group it was 68.9% ($P < 0.05$). The differences between both study subgroups regarding the distribution of different degrees of parental consanguinity (first, second, double first, and first once removed cousins) were insignificant ($P > 0.05$). The relative risk and 95% confidence interval (RR, 95% CI) for development of hearing loss in offspring of consanguineous marriage was 1.76 (95% CI 1.57-1.97, $P < 0.001$).
CONCLUSIONS: There was 76% increased risk for consanguineous marriage progeny to develop SNHL when compared to non consanguineous progeny.

Serra A, Spinato G, Cocuzza S, Licciardello L, Pavone P, Maiolino L.
Adaptive psychological structure in childhood hearing impairment: audiological correlations.
Acta Otorhinolaryngol Ital. 2017 Jun;37(3):175-179. doi: 10.14639/0392-100X-1291.

The present research deals with the clinical and social problems present during linguistic and cognitive development of deaf children. Currently, the development of Theory of Mind represents an important research field in deafness studies. These international studies highlighted a significant alteration in the development of Theory of Mind in deaf children compared to normal hearing children, especially in cases of congenital or preverbal hearing loss. In particular, the research focuses on the skills of deaf children in recognising emotions and desires, through both perceptive and cognitive methods, by evaluation of psycho-cognitive skills of children with severe hearing loss using a set of questions to be administered to hearing loss patients. The experiment was performed on a group composed of 10 children (5 males and 5 females) aged 4 to 9 years and 54 to 108 months, affected by bilateral congenital hearing loss (severe to total), or hearing loss that developed in preverbal children the year before entering elementary school, or during the fourth year of elementary school. The selection criteria were based on: audiologic evaluation, neuro-psychological tests administered to assess general, cognitive as well as praxis and perceptive abilities, and clinical observations performed to assess psychopathology using tests that assess development of both visual perceptive (Coloured Progressive Matrices) and graphic representational abilities (Test of Human Figure Drawings and the Family Drawing Test). The instrument "cognitive" was the "Deaf Children Series", arranged by us, that consists of a mental status examination (MSE) that evaluates: level of cognitive (knowledge-related) ability, emotional mood, and speech and thought patterns at the time of evaluation. Deaf children show a reduced responsiveness to the expressions of sadness on the perceptive side. Through the test, we observed a psychodynamic defense mechanism considering perceptive understanding performance. On the contrary, in normal hearing children, the emotion 'fear' is the most difficult to identify. Deaf children seem to be more susceptible to recognition of visual emotions. Furthermore, deaf children present significant problem-solving skills and emotional recognition skills, possibly as a result of their hearing impairment.

Su BM, Park JS, Chan DK.
Impact of primary language and insurance on pediatric hearing health care in a multidisciplinary clinic.
Otolaryngol Head Neck Surg. 2017 Oct;157(4):722-730. doi: 10.1177/0194599817725695. Epub 2017 Sep 12.

OBJECTIVE: This study aims to describe the effects of primary language and insurance status on care utilization among deaf or hard-of-hearing children under active otolaryngologic and audiologic care. Study Design Cross-sectional analysis. Setting Multidisciplinary hearing loss clinic at a tertiary center.
SUBJECTS AND METHODS: Demographics, hearing loss data, and validated survey responses were collected from 206 patients aged 0 to 19 years. Two-sided t tests and $\chi(2)$ tests were used to obtain descriptive statistics and hypothesis testing.
RESULTS: Of the sample, 52.4% spoke primarily English at home. Non-English-speaking children and families were less likely to receive psychiatric counseling (12.2% vs 35.2% in the English group, $P < .001$) and reported more difficulty obtaining educational interventions ($P = .016$), and 68.9% had public insurance. Parents of publicly insured children were less likely to know the type or degree of their child's hearing loss (56.9% vs 75.4%, $P = .022$), and these children were older on presentation to the clinic (8.5 vs 6.5 years of age, $P = .01$) compared to privately insured children. Publicly insured children were less likely to receive cochlear implants ($P = .046$) and reported increased difficulty obtaining hearing aids ($P = .047$). While all patients reported impairment in hearing-related quality of life, publicly insured children aged 2 to 7 years were more likely to perform below minimum thresholds on measures of auditory/oral functioning.
CONCLUSION: Even when under active care, deaf or hard-of-hearing children from families who do not speak English at home or with public insurance face more difficulty obtaining educational services, cochlear implants, and hearing aids. These findings represent significant disparities in access to necessary interventions.

van Beeck Calkoen EA, Sanchez Aliaga E, Merkus P, Smit CF, van de Kamp JM, Mulder MF, Goverts ST, Hensen EF.
High prevalence of abnormalities on CT and MR imaging in children with unilateral sensorineural hearing loss irrespective of age or degree of hearing loss.
Int J Pediatr Otorhinolaryngol. 2017 Jun;97:185-191. doi:10.1016/j.ijporl.2017.04.002. Epub 2017 Apr 4.

OBJECTIVE: Evaluation of causal abnormalities identified on CT and MR imaging in children with unilateral sensorineural hearing loss (USNHL), and the association with age and severity of hearing loss.
STUDY DESIGN: Retrospective cohort study.
SETTING: Tertiary referral otology/audiology center.
PATIENTS AND DIAGNOSTIC INTERVENTIONS: 102 children diagnosed with USNHL between 2006 and 2016 were included. They underwent CT and/or MR imaging for the evaluation of the etiology of their hearing loss.
MAIN OUTCOME MEASURES: Radiologic abnormalities of the inner ear and brain associated with USNHL.
RESULTS: Using CT and/or MR imaging, causal abnormalities were identified in 49%, which is higher than previously reported (25-40%). The most frequently affected site was the labyrinth (29%), followed by the cochlear nerve (9%) and brain (7%). No significant difference in the number or type of abnormalities was found for the degree of hearing loss or age categories.
CONCLUSIONS: Imaging is essential in the etiologic analysis of USNHL because of the high prevalence of causative abnormalities that can be identified with radiology, irrespective of the patients' age or degree of hearing loss. CT and MR imaging are complementary imaging options. The ideal imaging algorithm is controversial. Based on our findings, we conclude that there is limited additional diagnostic value of simultaneous dual modality imaging over sequential diagnostics. We therefore perform a stepwise radiological workup in order to maximize the diagnostic yield while minimizing impact and costs. If the primary imaging modality does not identify a cause for USNHL, performing the alternative imaging modality should be considered.

van Noort-van der Spek IL, Goedegebure A, Hartwig NG, Kornelisse RF, Franken MJP, Weisglas-Kuperus N.
Normal neonatal hearing screening did not preclude sensorineural hearing loss in two-year-old very preterm infants.
Acta Paediatr. 2017 Oct;106(10):1569-1575. doi: 10.1111/apa.13960. Epub 2017 Jul 13.

AIM: Very preterm infants are at risk of neonatal hearing loss. However, it is unknown whether infants with a normal neonatal hearing screening result risk sensorineural hearing loss (SNHL) at a later age.
METHODS: This cohort study was conducted at the Erasmus Medical University Center Rotterdam, the Netherlands, on 77 very preterm infants born between October 2005 and September 2008. All infants underwent auditory brainstem response audiometry during neonatal hearing screening and at two years of corrected age. The frequency of SNHL in infants with a normal neonatal hearing screening was analysed and the risk factors associated with newly diagnosed SNHL in these infants were examined.
RESULTS: We found that 3.9% (3/77) of the very preterm infants showed permanent hearing loss during their neonatal hearing screening. In addition, a relatively high prevalence of newly diagnosed SNHL (4.3%) was found in three of the 70 infants followed up at the age of two. The total prevalence rate of permanent hearing loss in the cohort was approximately 8%.
CONCLUSION: A normal outcome of neonatal hearing screening did not guarantee normal hearing at two years of age in this very preterm cohort and paediatricians should be alert to the possibility of late-onset SNHL.

Vila PM, Ghogomu NT, Odom-John AR, Hullar TE, Hirose K.
Infectious complications of pediatric cochlear implants are highly influenced by otitis media.
Int J Pediatr Otorhinolaryngol. 2017 Jun;97:76-82. doi: 10.1016/j.ijporl.2017.02.026. Epub 2017 Mar 12.
OBJECTIVE: Determine the incidence of ear infections in cochlear implant patients, evaluate the contribution of otitis media to complications, describe the bacteriology of otitis media in the cochlear implant population, the treatment provided at our center, and the long term outcome.
METHODS: Data collected included age at implantation, history of otitis media or ear tubes, etiology of hearing loss, inner ear anatomy, postoperative infections, time to infection, route of antibiotic administration, and interventions for infections. Categories of infection were acute otitis media, otitis media with effusion, tube otorrhea, meningitis, scalp cellulitis, and infection at the implant site.
RESULTS: Middle ear infections were diagnosed in 37% of implanted ears. Extension of middle ear infections into the implant site occurred in 2.8% of all implants (n = 16). Of the 16 infected devices, 10 were successfully treated with antibiotic therapy and did not require explantation. The retained implant group and explanted group both included some middle ear microbes such as Haemophilus influenzae and Streptococcus pneumoniae, as well as skin flora such as Staphylococcus aureus.
CONCLUSION: Otitis media in pediatric cochlear implant patients is a common event and usually does not lead to complications of the cochlear implant. However, when the ear infection spreads to the scalp and the implant site, it is still possible to eliminate the infection using antibiotic therapy, particularly when treatment is directed to the specific organism that is recovered from the infected space and the duration and route of antibiotic treatment is carefully considered.

Välimaa T, Kunnari S, Laukkanen-Nevala P, Lonka E; National Clinical Research Team.
Early vocabulary development in children with bilateral cochlear implants.
Int J Lang Commun Disord. 2017 Jun 16. doi: 10.1111/1460-6984.12322. [Epub ahead of print]
BACKGROUND: Children with unilateral cochlear implants (CIs) may have delayed vocabulary development for an extended period after implantation. Bilateral cochlear implantation is reported to be associated with improved sound localization and enhanced speech perception in noise. This study proposed that bilateral implantation might also promote early vocabulary development. Knowledge regarding vocabulary growth and composition in children with bilateral CIs and factors associated with it may lead to improvements in the content of early speech and language intervention and family counselling.
AIMS: To analyse the growth of early vocabulary and its composition during the first year after CI activation and to investigate factors associated with vocabulary growth.
METHODS & PROCEDURES: The participants were 20 children with bilateral CIs (12 boys; eight girls; mean age at CI activation = 12.9 months). Vocabulary size was assessed with the Finnish version of the MacArthur Communicative Development Inventories (CDI) Infant Form and compared with normative data. Vocabulary composition was analysed in relation to vocabulary size. Growth curve modelling was implemented using a linear mixed model to analyse the effects of the following variables on early vocabulary growth: time, gender, maternal education, residual hearing with hearing aids, age at first hearing aid fitting and age at CI activation.
OUTCOMES & RESULTS: Despite clear vocabulary growth over time, children with bilateral CIs lagged behind their age norms in receptive vocabulary during the first 12 months after CI activation. In expressive vocabulary, 35% of the children were able to catch up with their age norms, but 55% of the children lagged behind them. In receptive and expressive vocabularies of 1-20 words, analysis of different semantic categories indicated that social terms constituted the highest proportion. Nouns constituted the highest proportion in vocabularies of 101-400 words. The proportion of verbs remained below 20% and the proportion of function words and adjectives remained below 10% in the vocabularies of 1-400 words. There was a significant main effect of time, gender, maternal education and residual hearing with hearing aids before implantation on early receptive vocabulary growth. Time and residual hearing with hearing aids had a significant main effect also on expressive vocabulary growth.
CONCLUSIONS & IMPLICATIONS: Vocabulary development of children with bilateral Cis may be delayed. Thus, early vocabulary development needs to be assessed carefully in order to provide children and families with timely and targeted early intervention for vocabulary acquisition.

Wang L, Wang Z, Gao F, Peng KA.
Modiolar ossification in paediatric patients with auditory neuropathy.
J Laryngol Otol. 2017 Jul;131(7):598-601. doi: 10.1017/S0022215117001037. Epub 2017 May 15.
OBJECTIVE: To describe our finding of increased ossification of the modiolus in paediatric patients with auditory neuropathy who met criteria for cochlear implantation.
METHODS: A retrospective case series with a comparison group at a tertiary referral centre is described. Seven paediatric patients with auditory neuropathy who met criteria for and underwent cochlear implantation were identified. Fifteen paediatric implantees with bilateral profound sensorineural hearing loss were included as the comparison group. All patients underwent pre-operative computed tomography. Attenuation at the modiolus was measured in all subjects by a neuroradiologist blinded to clinical information.
RESULTS: Attenuation values in the modiolus in the auditory neuropathy patients (mean ± standard deviation = 796.2 ± 53.0 HU) was statistically significantly higher than in the comparison sensorineural hearing loss patients (267.1 ± 45.6 HU; $p < 0.05$, t-test).
CONCLUSION: Patients with auditory neuropathy who meet criteria for cochlear implantation demonstrate significantly higher modiolar attenuation on computed tomography imaging, consistent with increased ossification at the modiolus.

Wang X, Wu D, Zhao Y, Li D, He D.
Knowledge and attitude of mothers regarding infant hearing loss in Changsha, Hunan province, China.
Int J Audiol. 2017 Oct 16:1-6. doi: 10.1080/14992027.2017.1366671. [Epub ahead of print]
OBJECTIVE: The objective of this study was to explore the knowledge and attitude among mothers of newborns regarding infant hearing loss (HL) in

Changsha, Hunan province, China.
DESIGN: A questionnaire including 18 items was given to mothers.
STUDY SAMPLE: A total of 115 mothers participated in the study.
RESULTS: Seven risk factors for hearing loss were identified correctly by above 60% of respondents and the top three were prolonged noise (88.7%), high fever (82.6%) and ear discharge (82.6%). Poor knowledge was demonstrated on risk factors jaundice (20.0%), measles (22.6%), convulsion (33.0%) and traditional Chinese medicine (39.1%). Maternal knowledge scores in identification and intervention (2.68±0.31) was slightly higher than the score in risk factors (2.47±0.34). Ninety-nine per cent of the mothers expressed the willingness to test baby's hearing soon after birth and concern about hearing.
CONCLUSIONS: Mothers were concerned about baby's hearing and the attitude was positive. However, the correct recognition rate towards some risk factors for HL was low. Action needs to be taken to raise awareness about ear and hearing care, prevent HL caused by preventable causes and prompt early identification, early diagnosis and intervention of HL.

Wang XY, Huang LH, Du YT.
Research progress on the etiology of delayed-onset hearing loss in children. [Article in Chinese]
Zhonghua Er Bi Yan Hou Tou Jing Wai Ke Za Zhi. 2017 Oct 7;52(10):787-791. doi: 10.3760/cma.j.issn.1673-0860.2017.10.017.
Newborn hearing screening is an effective method for early detection of hearing loss, however, it is not able to detect delayed-onset hearing loss. By exploring the etiology of delayed-onset hearing loss in children, it can provide a clinical basis for early detection of delayed-onset hearing loss. Mutations in SLC26A4, mitochondrial, GJB2 and other genes, enlarged vestibular aqueduct, congenital cytomegalovirus infection, extracorporeal membrane oxygenation, and auditory neuropathy et al were more commonly reported risk factors. In this paper, the risk factors related to delayed-onset hearing loss, which are divided into 5 categories: genetic mutation, abnormal inner ear malformation, perinatal factors, auditory neuropathy and no identifiable cause, are reviewed and analyzed.

Wien MA, Whitehead MT.
The association among prematurity, cochlear hyperintensity, and hearing loss.
Neuroradiol J. 2017 Oct;30(5):448-453. doi: 10.1177/1971400917709623. Epub 2017 Jun 20.
BACKGROUND Prematurity is a major risk factor for neonatal hearing loss. Recent advancements in magnetic resonance imaging (MRI) have made it possible to evaluate structural details of the membranous labyrinths in premature infants that have heretofore been inaccessible.
OBJECTIVE We compared the prevalence of abnormal cochlear signal intensity in premature and term neonates and evaluated for a potential link with hearing loss.
MATERIALS AND METHODS We retrospectively reviewed 148 consecutive MR exams performed in premature (< 37 weeks' gestation) and term neonates performed over a 30-month period. Cochlear signal alteration was evaluated on three-dimensional T1-weighted imaging (T1WI) sequences. Each patient's electronic medical record was reviewed to document demographics, symptomatology, physical exam findings, and potential medical variables that could contribute to cochlear signal alteration.
RESULTS Cochlear hyperintensity on T1WI was present in 6.8% patients (n = 10) overall, but was much more common in preterm than term patients (12.2% (9 of 74) vs. 1.4% (1 of 74), respectively; p value < 0.05; Fisher's exact test). Overall, 14.9% (n = 15) of the patients with hearing test results failed the screening Auditory Brainstem Response exam. However, failure was much more common among patients with cochlear hyperintensity on T1W1 than those with normal findings (56% (5 of 9) vs. 11% (10 of 92), respectively; p value < 0.01; Fisher's exact test).
CONCLUSION Cochlear hyperintensity on T1WI is more common in preterm than term neonates, and potentially associated with hearing loss. Cochleae should be closely scrutinized in all premature infants; signal alterations should prompt further diagnostic inquiry and possible early otolaryngology referral.

Wong LY, Espinoza F, Alvarez KM, Molter D, Saunders JE.
Otoacoustic emissions in rural Nicaragua: Cost analysis and implications for newborn hearing screening.
Otolaryngol Head Neck Surg. 2017 May;156(5):877-885. doi:10.1177/0194599817696306. Epub 2017 Feb 1.
OBJECTIVE (1) Determine the incidence and risk factors for congenital hearing loss. (2) Perform cost analysis of screening programs. Study Design Proportionally distributed cross-sectional survey. Setting Jinotega, Nicaragua.
SUBJECTS & METHODS Otoacoustic emissions (OAEs) were used to screen 640 infants <6 months of age from neonatal intensive care unit, institutional, and home birth settings. Data on 15 risk factors were analyzed. Cost of 4 implementation strategies was studied: universal screening, screening at the regional health center (RHC), targeted screening, and screening at the RHC plus targeted screening. Cost-effectiveness analysis over 10 years was based on disability-adjusted life year estimates, with the World Health Organization standard of cost-effectiveness ratio (CER) / gross domestic product (GDP) <3, with GDP set at $4884.15.
RESULTS Thirty-eight infants failed the initial OAE (5.94%). In terms of births, 325 (50.8%) were in the RHC, 69 (10.8%) in the neonatal intensive care unit, and 29 (4.5%) at home. Family history and birth defect were significant in univariate analysis; birth defect was significant in multivariate analysis. Cost-effectiveness analysis demonstrated that OAE screening is cost-effective without treatment (CER/GDP = 0.06-2.00) and with treatment (CER/GDP = 0.58-2.52).
CONCLUSIONS Our rate of OAE failures was comparable to those of developed countries and lower than hearing loss rates noted among Nicaraguan schoolchildren, suggesting acquired or progressive etiology in the latter. Birth defects and familial hearing loss correlated with OAE failure. OAE screening of infants is feasible and cost-effective in rural Nicaragua, although highly influenced by estimated hearing loss severity in identified infants and the high travel costs incurred in a targeted screening strategy.

Wroblewska-Seniuk K, Greczka G, Dabrowski P, Szyfter-Harris J, Mazela J.
Hearing impairment in premature newborns: Analysis based on the national hearing screening database in Poland.
PLoS One. 2017 Sep 14;12(9):e0184359. doi: 10.1371/journal.pone.0184359. eCollection 2017.
OBJECTIVES: The incidence of sensorineural hearing loss is between 1 and 3 per 1000 in healthy neonates and 2-4 per 100 in high-risk infants.

The national universal neonatal hearing screening carried out in Poland since 2002 enables selection of infants with suspicion and/or risk factors of hearing loss. In this study, we assessed the incidence and risk factors of hearing impairment in infants ≤33 weeks' gestational age (wga).
METHODS: We analyzed the database of the Polish Universal Newborns Hearing Screening Program from 2010 to 2013. The study group involved 11438 infants born before 33 wga, the control group-1487730 infants. Screening was performed by means of transient evoked otoacoustic emissions. The risk factors of hearing loss were recorded. Infants who failed the screening test and/or had risk factors were referred for further audiological evaluation.
RESULTS: Hearing deficit was diagnosed in 11% of infants ≤25 wga, 5% at 26-27 wga, 3.46% at 28 wga and 2-3% at 29-32 wga. In the control group the incidence of hearing deficit was 0.2% (2.87% with risk factors). The most important risk factors were craniofacial malformations, very low birth weight, low Apgar score and mechanical ventilation. Hearing screening was positive in 22.42% newborns ≤28 wga and 10% at 29-32 wga and in the control group.
CONCLUSIONS: Hearing impairment is a severe consequence of prematurity. Its prevalence is inversely related to the maturity of the baby. Premature infants have many concomitant risk factors which influence the occurrence of hearing deficit.

Yoshinaga-Itano C, Sedey AL, Wiggin M, Chung W.
Early hearing detection and vocabulary of children with hearing loss.
Pediatrics. 2017 Aug;140(2). pii: e20162964. doi: 10.1542/peds.2016-2964. Epub 2017 Jul 8.
BACKGROUND AND OBJECTIVES: To date, no studies have examined vocabulary outcomes of children meeting all 3 components of the Early Hearing Detection and Intervention (EHDI) guidelines (hearing screening by 1 month, diagnosis of hearing loss by 3 months, and intervention by 6 months of age). The primary purpose of the current study was to examine the impact of the current EHDI 1-3-6
policy on vocabulary outcomes across a wide geographic area. A secondary goal was to confirm the impact of other demographic variables previously reported to be related to language outcomes.
METHODS: This was a cross-sectional study of 448 children with bilateral hearing loss between 8 and 39 months of age (mean = 25.3 months, SD = 7.5 months). The children lived in 12 different states and were participating in the National Early Childhood Assessment Project.
RESULTS: The combination of 6 factors in a regression analysis accounted for 41% of the variance in vocabulary outcomes. Vocabulary quotients were significantly higher for children who met the EHDI guidelines, were younger, had no additional disabilities, had mild to moderate hearing loss, had parents who were deaf or hard of hearing, and had mothers with higher levels of education.
CONCLUSIONS: Vocabulary learning may be enhanced with system improvements that increase the number of children meeting the current early identification and intervention guidelines. In addition, intervention efforts need to focus on preventing widening delays with chronological age, assisting mothers with lower levels of education, and incorporating adults who are deaf/hard-of-hearing in the intervention process.

Yuan EF, Xia W, Huang JT, Hu L, Liao X, Dai X, Liu SM.
A sensitive and convenient method for clinical detection of non-syndromic hearing loss-associated common mutations.
Gene. 2017 Sep 10;628:322-328. doi: 10.1016/j.gene.2017.07.045. Epub 2017 Jul 19.
BACKGROUND: The majority of non-syndromic hearing loss (NSHL) patients result from causative mutations in GJB2, SLC26A4 and mitochondrial 12S rRNA genes. Accurate detection of these genetic mutations is increasingly recognized for its clinical significance to reduce incidence and guide individual treatment of NSHL. Current methods for clinical practice are labor intensive, expensive or of low sensitivity.
METHODS: Genomic DNA from 7 newborns not passing the hearing screening and 94newborns passing the hearing screening were analyzed for the common mutations using high resolution melting analysis (HRMA) and Sanger sequencing.
RESULTS: Our newly developed HRMA allowed the hot-spot mutations of GJB2 c.176_191del16 and c.235delC, SLC26A4 IVS7-2A>G and mitochondrial 12S rRNA 1494C>T and 1555A>G to be detected by melting profiles based on small amplicons. HRMA can distinguish different content mutant DNA from wildtype DNA, with a detection limit of 5%. Moreover, the results were highly concordant between HRMA
and Sanger sequencing.
CONCLUSIONS: These results indicate that HRMA could be used as a routine clinical method for prenatal diagnosis and newborn genetic screening due to its accuracy, sensitivity, and rapid, low-cost and less laborious workflows.