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Odds of Autism at 5 to 10 Years of Age for Children Who Did Not Pass Their Automated Auditory Brainstem Response Newborn Hearing Screen, but were Diagnosed with Normal Hearing

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Abstract

Background: Research has found atypical auditory brainstem response (ABR) activity in some children with Autism Spectrum Disorder (ASD). The current study examined whether an association may also be found between ASD and pass/refer results obtained via automated auditory brainstem response (AABR) screening. As stewards of large-scale AABR data, an AABR–ASD association may be of interest to EHDI programs.

Methods: State EHDI data for children born in Maine between 2003 and 2005 were linked with education records, including special education status, for the 2010-2011 and 2013-2014 school years.

Results: Children who did not pass their AABR screen but were later documented to have typical hearing were at more than eight times the odds of being identified with ASD at 5 to 7 years of age, and over six times the odds at 8 to 10 years of age.

Conclusion: Newborns who did not pass their AABR screen but were subsequently diagnosed with typical hearing, experienced higher rates of ASD 5 to 10 years later. With further research evidence, this may create opportunities for EHDI programs to support and facilitate the work of colleagues in the ASD community, as well as further assist families already touched by EHDI systems.

Keywords: Automated Auditory Brainstem Response, Autism Spectrum Disorder, Special Education, Newborn Hearing Screening, ASD, AABR

Acronyms: AABR = automatic auditory brainstem response; ABR = auditory brainstem response; ASD = autism spectrum disorder; EHDI = Early Hearing Detection and Intervention

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Auditory Brainstem Response (ABR) and the more limited Automated Auditory Brainstem Response (AABR) are familiar to many in the Early Hearing Detection and Intervention (EHDI) community as tools for screening (AABR) and diagnosing (ABR) hearing loss in children. Although commonly used for audiological evaluations, prior research (e.g., Cohen et al., 2013; Rosenhall et al., 2003; Roth et al., 2011) has shown that some individuals diagnosed with or suspected to have Autism Spectrum Disorder (ASD) demonstrate atypical results for ABR testing. The purpose of this study was to examine whether the evidence of a possible association between ASD and ABR using ABR testing—which provides detailed data regarding ABR activity—may be detectable using AABR screening that only provides *pass* or *refer* results. Although AABR screening provides more limited data than ABR testing, it is used in many EHDI programs and thus already available for many young infants.

Autism Spectrum Disorder

ASD is a neurodevelopmental disorder characterized by persistent communication impairments related to social communication and social interaction; and behavioral symptomatology described as restricted, repetitive patterns of behaviors, interests, and activities (American Psychiatric Association, 2013). Children diagnosed with ASD typically demonstrate functional performance deficits directly related to these characteristics in the areas of adaptive skills, communication and social engagement with peers and adults, and behavioral regulation (Volkmar et al., 2014).

A particular concern with ASD is the steadily increasing number of cases that have been identified over the last few decades—an observation that has received widespread attention by families, health care professionals, and policy makers (Maenner et al., 2020; Shaw et al., 2020). Since 2007, the American Academy of Pediatrics has

recommended that all children be screened for ASD at 18 and 24 months of age (Johnson et al., 2007). In the past decade, the age for a reliable diagnosis of ASD has decreased to as early as 14 months with the recommended age for early diagnosis at 18 months (Hyman et al., 2020; Pierce et al., 2019). This trend leads to an increased demand for early intervention services for children as young as 12 months who demonstrate ASD symptomatology (Chawarska et al., 2014). Barriers to screening for ASD include physician time and resources to screen, as well as lack of confidence in screening tools (Khowaja et al., 2018; Siu & the U.S. Preventive Services Task Force, 2016).

Automated Brainstem Response and Autism Spectrum Disorder

Multiple studies have observed atypical ABR results in children with ASD. For example, 101 Swedish children with typical (i.e., *normal*) hearing who were diagnosed with ASD (mean age = 8.38 years, range = 4 to 20 years) were found to have abnormal ABR results when compared to a typically hearing control group (Rosenhall et al., 2003). In another study of younger children with suspected ASD and typical hearing, 26 Israeli youth (mean age = 32.5 months, range = 24 to 45 months) exhibited abnormal ABRs when compared to a matched sample of children with language delay, as well as when compared to clinical norms (Roth et al., 2011). More recently Miron and colleagues (2016) examined ABRs of infants (mean adjusted age of 1.6 months) who were later diagnosed with ASD. Compared to ABRs from a case matched control group, the ASD diagnosis group had increased interpeak latency I-V and wave V latency. When the same authors compared ABRs from 1.5 to 3.5 year olds with ASD to clinical norms, increased interpeak latencies were seen in I-III, III-V, and I-V along with increased latencies in I, II, and V.

Such differences may vary based on age (Miron et al., 2018; Roth et al., 2011), and may be evident in the latency and amplitude of the waveform. For example, increased latencies have been observed in children with ASD suggesting slower conduction and/or longer conduction pathways, particularly in waves I, II and V (Miron et al., 2018; Miron et al., 2016; Rosenhall et al., 2003; Roth et al., 2011; Talge et al., 2018) and in those children under 8 years of age (Miron et al., 2018; Miron et al., 2016; Talge et al., 2018). Children with ASD may also be more likely than children with other language delays to have increased interpeak latencies with I-III, III-V, and I-V (Miron et al., 2016; Rosenhall et al., 2003; Roth et al., 2011; Talge et al., 2018). In addition to latency, other studies have examined wave amplitude and found greater amplitude in waves I and III among children with ASD, versus age-matched controls (Claesdotter-Knutsson et al., 2019; Santos et al., 2017). It is unknown whether such atypical ABR activity existed at birth or developed over time for these individuals. However, based on a small histopathology study of 2-year to 36-year-old decedents with ASD that showed changes in the auditory brainstem nuclei, some have proposed that it may be possible to use ABR testing to screen for ASD (Smith et al., 2019).

Of course, an association between ASD and AABR may reflect other mechanisms or processes. For example, in studies of infants referred due to atypical newborn hearing screening tests, 39%–60% had middle ear effusions (Adachi et al., 2010; Weber et al., 2018). The effusions occurred along with other sensorineural hearing loss in many infants, while others had typical hearing once the effusion was cleared. (Adachi et al., 2010; Weber et al., 2018). A slightly increased frequency of otitis media with effusion was seen in children with typical hearing and ASD (Adams et al., 2016; Myne & Kennedy, 2018), suggesting that an association with ASD may also reflect other, more fundamental mechanisms that are also related to middle ear effusions at birth.

Automated Auditory Brainstem Response Screening

As a screening tool for hearing loss, AABR does not provide the breadth and depth of information available in ABR diagnostic testing. Nevertheless, these findings based on ABR data raise the question of whether similar associations may be seen between the pass/refer results obtained via AABR newborn screening and subsequent identification of ASD. For clarity, AABR screening technology used by the equipment employed in this study

> …delivers thousands of soft click sounds at 35 dB nHL ('normal hearing level' scale) to a newborn's ears through disposable earphones. Each click evokes a series of identifiable brain waves from a special area of the baby's brain called the auditory brainstem. This brain wave activity is called the auditory brainstem response (ABR)…The instance in which the screener delivers a click and receives a response to that click is called a sweep. Sensors on the baby's skin pick up the brain wave signals and transmit the signals to the screener. The screener uses advanced signal processing technology to separate the ABR waves from background noise and other brain activity. These brain waves are averaged and checked to see if they are consistent with a pattern called a template. The template is derived from ABRs of normal-hearing infants. The screener must detect the ABR waveform with high statistical confidence to determine that a response is present…The screener will generate a PASS result when it collects sufficient data to establish with > 99% statistical confidence that an ABR signal is present and consistent with the template at a minimum of 1000 sweeps…If it has not established with > 99% statistical confidence that the ABR signal is present at 15,000 sweeps, the screener will generate a REFER result. (Natus Medical Incorporated, 2014, p. 9)

An association between AABR screening results and ASD would potentially be valuable given the use of AABR in many EHDI programs across the United States and other countries. In 1993, the U.S. National Institutes of

Health recommended that all newborns be screened for hearing loss. Subsequent position statements by the Joint Committee on Infant Hearing (Joint Committee on Infant Hearing, 1995, 2000, 2007, 2019) and Healthy People 2010 (U.S. Department of Health and Human Services & Office of Disease Prevention and Health Promotion, 2000) and 2020 (U.S. Department of Health and Human Services & Office of Disease Prevention and Health Promotion) called for universal screening of all newborns by one month of age—preferably prior to hospital discharge. AABR is widely used in many EHDI programs, and as such provides access and data for a large portion of births.

To that end, we conducted two sets of population-based archival analyses by linking newborn hearing screening results at birth, with public school records from kindergarten through fifth grade. Specifically, we were interested in those children who did not pass their newborn hearing screen using AABR but were subsequently diagnosed with typical (i.e., normal) hearing. The goal was to determine the prevalence rate of ASD among these children and compare it to overall rates. Given that prior studies (Cohen et al., 2013; Rosenhall et al., 2003; Roth et al., 2011) found that children who were diagnosed with or suspected to have ASD were more likely to show abnormal ABR activity, we hypothesized that newborns *with typical hearing* who nevertheless did not pass their AABR hearing screen would be more likely to be identified with ASD in elementary school. Although we anticipated such an association would also exist among children with diagnosed hearing loss, we focused solely on those with typical hearing to avoid any confounds with hearing loss, such as a possible inflated risk of being identified with ASD due to a child with hearing loss receiving a more careful evaluation upon school entry.

Method

This archival study was based on statewide newborn hearing screening and diagnostic data obtained from the Maine Newborn Hearing Program (EHDI), and statewide education data obtained from the Maine Department of Education. The Maine Newborn Hearing Program was established in 2000 and has been collecting newborn hearing screening and diagnostic evaluation data for all children born in the state since 2003. Coincidentally, all birthing hospitals in Maine used AABR for screening from the inception of the Maine Newborn Hearing Program, with all equipment provided by a single supplier (Natus). Relevant for this study, it is worth noting that the Maine Newborn Hearing Program data also includes information obtained from the electronic birth certificate, the Maine Birth Defects Program, and the Maine Newborn Bloodspot Screening Program.

The Maine Department of Education maintains the State Longitudinal Data System, which stores educational data for all children attending public school (and many large private schools) from preschool through the 12th grade. In addition to educational outcome data, the system includes the disability identification, such as ASD, for children receiving special education services. The existence of these two independent data systems creates a unique opportunity to investigate the potential correlation between the newborn hearing screening results and identification of ASD at a later age.

Measures

Eight childhood characteristics or variables were examined including child sex, age, reported birth defect, NICU status, birth weight, AABR/hearing status, special education status, and ASD status. Definitions for each variable can be found in Table 1.

Sample

Within the newborn hearing screening data, we identified all births in Maine from 2003 to 2005 (N = 41,493). Given that special education identification may change over time, these records were then linked to the Maine Department of Education records for the 2010 and 2013 school years (Time 1 and Time 2, respectively). By examining two different time-periods corresponding to early and later elementary school years, it would be possible to observe age-related variation within the same cohort of children. Record linkage was based on the child's name (first, middle, and last) and date of birth using an iterative, probabilistic linkage algorithm (Tu & Mason, 2004; Tu, Mason, & Song, 2007). Summaries of the data-flow from the original birth and school records, through data linkage, to special education enrollment and ASD identification are presented in Figure 1 (for Time 1) and Figure 2 (for Time 2), as well as in the following section.

All analyses were conducted using a de-identified data set, and the project was approved by the University of Maine Institutional Review Board (IRB), the Maine Center for Disease Control and Prevention (Maine CDC), and the Maine Department of Education.

Results

Time 1: Automated Auditory Brainstem Response at Birth Predicting Autism Spectrum Disorder at Age 5–7 Years

Record Linkage

Newborn records (AABR screening, diagnostic evaluation, birth data) for 41,493 children born in Maine from 2003 to 2005 were electronically linked to 2010/2011 school records for 37,730 children born in 2003 to 2005. A total of 30,226 matches were found, reflecting 72.8% of the newborn and 80.1% of the 2010/2011 school records. Non-matched birth records included children who moved out of state or were not attending public school in 2010/2011, as well as those who died or had a name change. Non-matched school records included children born out of state as well as those with a name change. A summary of the data-flow from birth and school records to special education enrollment and ASD identification is presented in Figure 1.

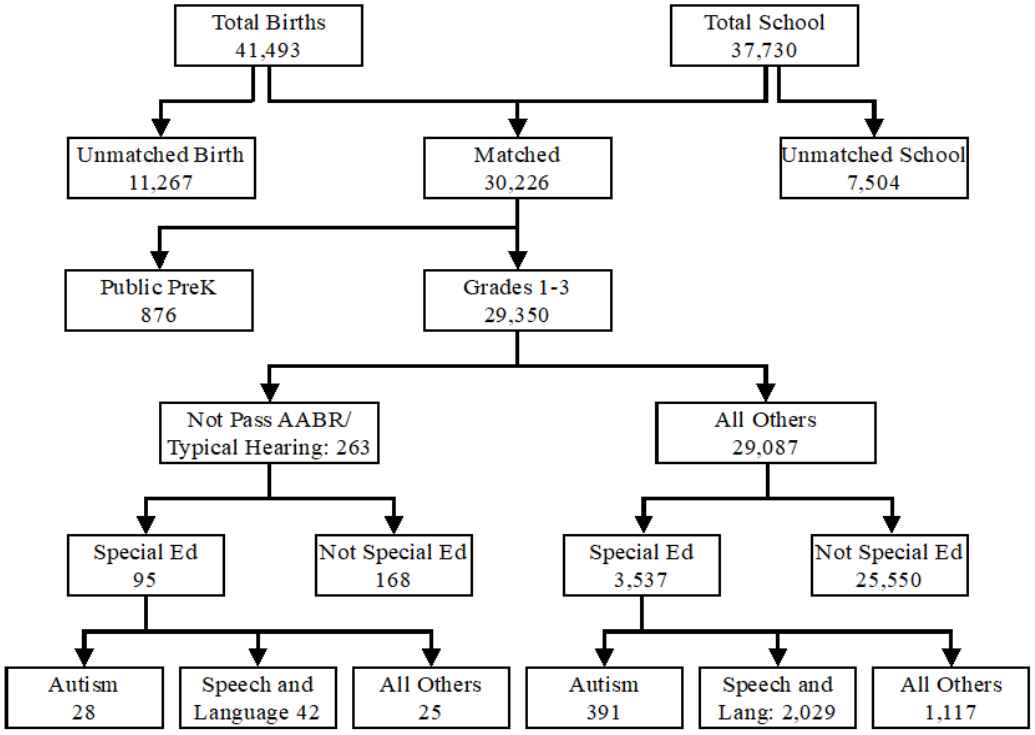
Table 1

Measures Used to Investigate Potential Correlation between Automated Auditory Brainstem Response (AABR) and Autism Spectrum Disorder (ASD)

Variable	Definition
Child Sex	A dummy variable indicating child sex (Female = 0, Male = 1).
Age	Child age in years.
Reported Birth Defect	A dummy variable indicating the documented presence of any of the 57 birth defects covered by the Maine Birth Defects program (0 = No documented birth defect, 1 = Documented birth defect).
NICU Status	A dummy variable indicating that a child's birth hospitalization included time in the neonatal intensive care unit (0 = No NICU placement at the birth hospitalization, 1 = NICU placement at the birth hospitalization).
Birth Weight	Birth weight, as recorded on a child's electronic birth certificate, was coded as one of four categories—extremely low birth weight (< 1000g), very low birth weight (1000g up to 1500g), low birth weight (1500g up to 2500g), and normal birth weight (2500+g). Note that although all analyses used the 4-category birth weight variable, due to suppression rules, birth weight is reported in tables as < 2500g and 2500+g.
AABR/Hearing Status	A dummy variable coded "1" if a child had a final AABR newborn screening result of "refer" for one or both ears **and** a formal diagnosis of typical/normal hearing reported to the state EHDI program. Children with diagnostic testing that was in process, missing, or unknown were not considered to have a diagnosis of typical hearing and were coded as "0".
Special Education Status	A dummy variable indicating whether a child was enrolled in special education during the specified academic year (0 = Not enrolled in special education, 1 = Enrolled in special education).
ASD Status	A dummy variable indicating whether a child was identified as having ASD based on their special education category (0 = Not enrolled in special education *or* enrolled in special education with a category other than ASD, 1 = Enrolled in special education with the category of ASD).

Figure 1

Case-Flow from Birth and School Records. Through Data Linkage and Time 1 Special Education Status



Note. Information regarding the counts for children enrolled in special education (Special Ed) under the specific category of speech/language impairment is provided for context, but not analyzed separately. AABR = automated auditory brainstem response.

Sample Characteristics

Of the 30,226 matched records, 876 were for children enrolled in public preschool at that time. Most children do not attend public preschool, which is neither required nor offered uniformly across the state; therefore, these children were excluded to avoid potential sampling bias. This resulted in a final sample of 29,350 matched records, for whom 8,080 were five years of age, 10,577 were six years of age, and 10,693 were seven years of age. Slightly more than half were male (n = 15,134) and 6.5% (n = 1,903) were placed in the NICU during their birth hospitalization. Eighty-one were extremely low birth weight (< 1000g), 161 were very low birth weight (1000g up to 1500g), and 1,615 were low birth weight (1500g up to 2500g) when born. In addition, 1,038 had a known birth defect.

Characteristics of Children Who Did Not Pass Their AABR Screen, But Were Diagnosed with Normal/ Typical Hearing

Of the 29,350 matched records, 263 were children who did not pass their AABR hearing screening, but were later documented to have normal/typical hearing. As summarized in the first pair of columns in Table 2, they were nevertheless more likely to have a birth defect (OR = 2.40, 95% CI: 1.53–3.76; $\chi^2(1, N = 29,350) = 15.39$, $p < .001$), be in a lower birth weight category ($\chi^2(3, N = 29,337) = 17.55$, $p = .001$), have been in the NICU at birth (OR = 2.02, 95% CI: 1.39–2.93; $\chi^2(1, N = 29,350) = 14.14$, $p < .001$), and be male (OR = 2.05, 95% CI: 1.58–2.66; $\chi^2(1, N = 29,350) = 30.27$, $p < .001$).

Table 2

Frequencies of Various Child Characteristics at Time 1 Based on Automated Auditory Brainstem Response (AABR) Status and Autism Spectrum Disorder (ASD) Status

	All Others	Not Pass AABR Typical Hearing	Not ASD	ASD
No Birth Defect	28,070	242	27,906	406
Birth Defect	1,017	21	1,025	13
Normal BW	27,247	233	27,095	385
Low BW	1,827	30	1,824	33
Not NICU Birth	27,216	231	27,060	387
NICU Birth	1,871	32	1,871	32
Female	14,133	83	14,151	65
Male	14,954	180	14,780	354

Note. BW = birthweight; NICU = Newborn Intensive Care Unit.

Birth Factors Related to ASD at 5–7 Years Age

Of the 29,350 children in the final dataset, 3,632 (12.4%) were enrolled in special education, and 419 (1.4%) were specifically identified as having ASD at five to seven years of age. As summarized in the second pair of columns in Table 2, males were at higher risk for ASD during this age period (OR = 5.21, 95% CI: 4.00–6.80; $\chi^2(1, N = 29,350) = 184.48$, $p < .001$), as were older children ($\chi^2(2, N = 29,350) = 23.22$, $p < .001$), with rates of 0.9% for five-year-olds, 1.6% for six-year-olds, and 1.7% for seven-year-olds. Presence of a birth defect ($\chi^2(1, N = 29,350) = 0.23$, $p = .63$), birth weight category ($\chi^2(3, N = 29,337) = 2.32$, $p = .51$), and NICU status ($\chi^2(1, N = 29,350) = 0.93$, $p = .33$) were unrelated to ASD at five to seven years of age.

Preliminary Analyses: Predicting Age 5–7 Special Education Placement Based on AABR Screening Results and Hearing Status

As a preliminary test, analyses first examined the overall rate of special education placement—any special education category—among children who did not pass an AABR screen, but were diagnosed with typical hearing. Results found that the 263 children who did not pass their AABR hearing screen but had documented typical hearing experienced higher rates of special education five to seven years later—36.1% versus 12.2% for all other children (OR = 4.08, 95% CI: 3.17–5.27; $\chi^2(1, N = 29,350) = 138.01$, $p < .001$).

To address additional possible confounds, a logistic regression examined this same relationship controlling for sex, age, reported birth defect, birth weight category, and NICU status. As summarized in Table 3, children who did not pass their AABR newborn hearing screen, but were subsequently diagnosed with typical hearing continued to exhibit higher levels of enrollment in special education when five to seven years of age (OR = 3.35, 95% CI: 2.58–4.35), even after controlling for these other factors. Although not presented in Table 3, results were similar when controlling for school grade-level instead of age (OR = 3.49, 95% CI: 2.70–4.53).

Primary Analyses: Predicting Age 5–7 ASD Identification Based on AABR Screening Results and Hearing Status

These same analyses were then repeated, specifically focusing on ASD classification at age 5 to 7 years. The 263 children who did not pass their AABR hearing screen but had documented typical hearing were again found to experience higher rates of ASD five to seven years later—10.6% versus 1.3% for all other children (OR = 8.74, 95% CI: 5.84–13.10; $\chi^2(1, N = 29,350) = 160.27$, $p < .001$).

As summarized in Table 4, this result remained even after controlling for sex, age, reported birth defect, birth weight category, and NICU status. Children who did not pass their AABR newborn hearing screen but were diagnosed with typical hearing continued to exhibit higher levels of ASD when five to seven years old (OR = 6.94, 95% CI: 4.59–10.48), even after controlling for these other factors. Although not presented in Table 4, similar results were found controlling for school grade-level instead of age (OR = 7.34, 95% CI: 4.86–11.07).

Table 3

Logistic Regression Predicting Special Education Status (S.E.) at 5–7 Years of Age Based on Newborn Automated Auditory Brainstem Response (AABR) Screen and Child Characteristics

Variable	*b*	S.E.	Wald	*p*	OR [95% CI]
Constant	-4.103	0.148	764.69	< .001	0.017
Male	0.838	0.038	476.86	< .001	2.311 [2.144, 2.491]
Age	0.253	0.023	117.54	< .001	1.287 [1.230, 1.347]
Any Birth Defect	0.514	0.095	29.08	< .001	1.671 [1.387, 2.014]
ELBW	0.280	0.275	1.03	0.310	1.323 [0771, 2.269]
VLBW	-0.005	0.216	0.00	0.982	0.995 [0.651, 1.521]
LBW	0.383	0.075	26.13	< .001	1.466 [1.266, 1.698]
NICU	0.357	0.072	24.53	< .001	1.429 [1.241, 1.646]
Not Pass AABR w/TH	1.209	0.133	82.91	< .001	3.351 [2.583, 4.347]

Note. Special Education Status (0 = Not enrolled in special education, 1 = Enrolled in special education); Male (0 = Female, 1 = Male); Any Birth Defect (0 = No record of monitored birth defect, 1 = Presence of a monitored birth defect); ELBW (Extremely low birth weight under 1000g, 0 = No, 1 = Yes); VLBW (Very low birth weight, 1000g to 1500g, 0 = No, 1 = Yes); LBW (Low birth weight, 1500g to 2500g, 0 = No, 1 = Yes); NICU (Presence in NICU during birth hospitalization, 0 = No, 1 = Yes); Not Pass AABR w/TH (Child with typical hearing who did not pass their newborn AABR screening, 0 = Passed screening, 1 = Did not pass screen but later diagnosed with typical hearing). All Wald tests have one degree of freedom.

Table 4

Logistic Regression Predicting Autism Spectrum Disorder (ASD) Identification at 5–7 Years of Age Based on Newborn Automated Auditory Brainstem Response (AABR) Screen and Child Characteristics

Variable	*b*	S.E.	Wald	*p*	OR [95% CI]
Constant	-6.966	0.424	270.37	< .001	0.001
Male	1.620	0.136	142.23	< .001	5.052 [3.871, 6.593]
Age	0.249	0.065	14.71	< .001	1.283 [1.129, 1.456]
Any Birth Defect	-2.30	0.332	0.48	0.487	0.794 [0.415, 1.521]
ELBW	0.114	1.069	0.01	0.915	1.120 [0.138, 9.103]
VLBW	0.081	0.794	0.01	0.919	1.084 [0.229, 5.140]
LBW	0.320	0.211	2.30	0.129	1.377 [0.911, 2.083]
NICU	0.006	0.213	0.00	0.979	1.006 [0.662, 1.527]
Not Pass AABR w/TH	1.937	0.210	84.80	< .001	6.940 [4.595, 10.481]

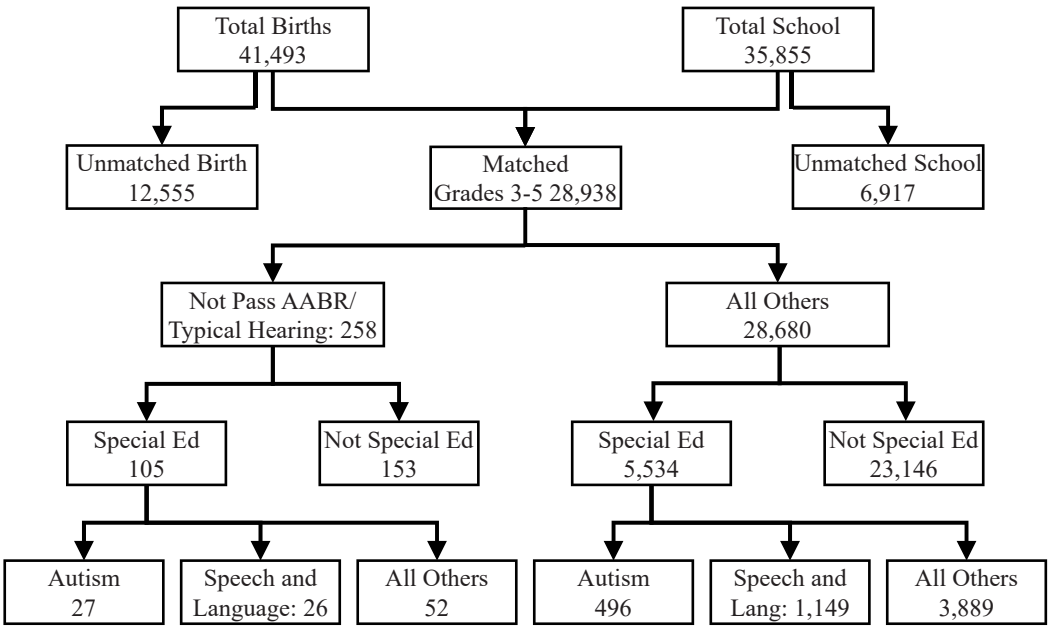
Note. ASD Identification (0 = Not identified as having ASD, 1 = Identified as having ASD); Male (0 = Female, 1 = Male); Any Birth Defect (0 = No record of monitored birth defect, 1 = Presence of a monitored birth defect); ELBW (Extremely low birth weight under 1000g, 0 = No, 1 = Yes); VLBW (Very low birth weight, 1000g to 1500g, 0 = No, 1 = Yes); LBW (Low birth weight, 1500g to 2500g, 0 = No, 1 = Yes); NICU (Presence in NICU during birth hospitalization, 0 = No, 1 = Yes); Not Pass AABR w/TH (Child with typical hearing who did not pass their newborn AABR screening, 0 = Passed screening, 1 = Did not pass screen but were later diagnosed with typical hearing). All Wald tests have one degree of freedom.

Time 2: Automated Auditory Brainstem Response at Birth Predicting Autism Spectrum Disorder at Age 8–10 Years

As children age, more in any given cohort will tend to be identified with ASD. Consequently, ASD and special education rates also change with age throughout the elementary school years. Therefore, the previous analyses were repeated using the same birth cohort linked with school records at a later point in time. Although largely overlapping the children included in Time 1, the underlying samples are not identical due to factors such as out-migration (children leaving the state) and sample-specific in-migration (children who were born in Maine, but not enrolled in public school at Time 1). A summary of the data-flow from birth and school records to special education enrollment and ASD identification using Time 2 data is presented in Figure 2.

Figure 2

Case-Flow from Birth and School Records, Through Data Linkage and Time 2 Special Education Status



Note. Information regarding the counts for children enrolled in special education (Special Ed) under the specific category of speech/language impairment is provided for context, but not analyzed separately. AABR = automated auditory brainstem response.

Record Linkage

Newborn records for the 41,493 children born from 2003 to 2005 were electronically matched to 2013-2014 school records for 35,855 children born those same years. Second grade students who were in preschool in 2010-2011 and not included in Time 1 analyses were excluded to continue with the same potential cohort. A total of 28,938 matches were found, reflecting 69.7% of the newborn records and 80.7% of the 2013-2014 school records. Non-matched birth records included children who moved out of state or were not attending public school in 2013-2014, as well as those who died or had a name change. Non-matched school records included children born out of state as well as those with a name change.

Sample Characteristics

Among matched records, 8,066 were eight years of age, 10,395 were nine years of age, and 10,477 were ten years of age. Slightly more than half were male (n = 14,984) and 6.5% (n = 1,890) had been in the NICU during their birth hospitalization. Seventy-nine were extremely low birth weight (< 1000g), 157 were very low birth weight (1000g up to 1500g), and 1,601 were low birth weight (1500g up to 2500g) when born. In addition, 1,013 had a known birth defect.

Characteristics of Children Who Did Not Pass Their AABR Screen, But Were Diagnosed with Normal/ Typical Hearing

Two hundred fifty-eight children who did not pass their AABR hearing screen were later found to have documented normal/typical hearing. As summarized in the first two columns of Table 5 they were also more likely to have a birth defect (OR = 2.47, 95% CI: 1.58–3.88; ($\chi^2(1, N = 28{,}938) = 16.58$, $p < .001$), be in a lower birth weight category ($\chi^2(3, N = 28{,}927) = 18.56$, $p < .001$), have spent time in the NICU at birth (OR = 2.04, 95% CI: 1.41–2.97; $\chi^2(1, N = 28{,}938) = 14.70$, $p < .001$), and be male (OR = 2.08, 95% CI: 1.60–2.72; $\chi^2(1, N = 28{,}938) = 30.89$, $p < .001$). This is generally consistent with the results from Time 1.

Birth Factors Related to ASD at 8–10 Years Age

Of the 28,938 children in the final dataset, 523 (1.8%) were identified as having ASD at eight to ten years of age. As summarized in the second pair of columns of Table 5, males

Table 5

Frequencies of Various Child Characteristics at Time 2 Based on Automated Auditory Brainstem Response (AABR) Status and Autism Spectrum Disorder (ASD) Status

	All Others	Not Pass AABR Typical Hearing	Not ASD	ASD
No Birth Defect	27,688	237	27,424	501
Birth Defect	922	21	991	22
Normal BW	26,862	228	26,619	471
Low BW	1,807	30	1,786	51
Not NICU Birth	26,822	226	26,574	474
NICU Birth	1,858	32	1,841	49
Female	13,874	80	13,864	90
Male	14,806	178	14,551	433

Note. BW = birthweight; NICU = Newborn Intensive Care Unit.

continued to have higher rates of ASD during this age period (OR = 4.58, 95% CI: 3.65–5.76; $\chi^2(1, N = 28{,}938) = 205.16$, $p < .001$) and presence of a birth defect continued to be unrelated at this later age ($\chi^2(1, N = 28{,}938) = 0.79$, $p = .38$). In contrast to results three years earlier, higher rates of ASD at eight to ten years of age were observed among those born below normal birth weight (2.8%), compared to those born at normal birth weight (1.7%; $\chi^2(3, N = 28{,}927) = 14.17$, $p = .003$). NICU births also had higher rates of ASD (OR = 1.49, 95% CI: 1.11–2.01; $\chi^2(1, N = 28{,}938) = 7.03$, $p = .008$) compared to non-NICU births (2.6% vs. 1.8%). In contrast, while age continued to be related to ASD, rates now *declined* slightly with age ($\chi^2(2, N = 28{,}938) = 7.87$, $p = .02$) reflecting a potential peak rate of 2.2% around eight years of age, versus 1.6% for nine-year olds, and 1.7% for ten-year olds.

Preliminary Analyses: Predicting Age 8–10 Special Education Placement Based on AABR Screening Results and Hearing Status

A preliminary pair of analyses first examined the overall rate of special education placement—in any special education category—among children age 8 to 10 years, who did not pass an AABR screen, but were diagnosed with typical hearing. Results found that the 258 children who did not pass their AABR hearing screen but had documented typical hearing experienced higher rates of enrollment in special education eight to ten years later—40.7% versus 19.3% for all other children (OR = 2.87, 95% CI: 2.24–3.69; $\chi^2(1, N = 28{,}938) = 74.65$, $p < .001$).

This effect continued to be present in a logistic regression controlling for sex, age, reported birth defect, birth weight category, and NICU status. As summarized in Table 6, children who did not pass their AABR newborn hearing screen and were nevertheless diagnosed with typical hearing continued to exhibit higher levels of enrollment in special education when eight to ten years of age (OR = 2.52, 95% CI: 1.95–3.25), even after controlling for these other factors. Although not presented in Table 6, results were similar using grade-level in school in place of age (OR = 2.69, 95% CI: 2.07–3.49).

Table 6

Logistic Regression Predicting Special Education Status at 8–10 Years of Age Based on Newborn Automated Auditory Brainstem Response (AABR) Screen and Child Characteristics

Variable	*b*	S.E.	Wald	*p*	OR [95% CI]
Constant	-0.994	0.174	32.77	< .001	0.370
Male	0.794	0.031	635.27	< .001	2.21 [2.079, 2.352]
Age	-0.107	0.019	31.65	< .001	.0899 [.0866, 0.933]
Any Birth Defect	0.392	0.084	21.83	< .001	1.480 [1.255, 1.744]
ELBW	0.807	0.247	10.67	0.001	2.242 [1.381, 3.638]
VLBW	0.119	0.194	0.38	0.539	1.27 [0.770, 1.648]
LBW	0.446	0.064	48.68	< .001	1.562 [1.378, 1.771]
NICU	0.339	0.063	29.28	< .001	1.403 [1.241, 1.587]
Not Pass AABR w/TH	0.923	0.131	49.94	< .001	2.516 [1.948, 3.249]

Note. Special Education Status (S.E.; 0 = Not enrolled in special education, 1 = Enrolled in special education); Male (0 = Female, 1 = Male); Any Birth Defect (0 = No record of monitored birth defect, 1 = Presence of a monitored birth defect); ELBW (Extremely low birth weight under 1000g, 0 = No, 1 = Yes); VLBW (Very low birth weight, 1000g to 1500g, 0 = No, 1 = Yes); LBW (Low birth weight, 1500g to 2500g, 0 = No, 1 = Yes); NICU (Presence in Neonatal Intensive Care Unit during birth hospitalization, 0 = No, 1 = Yes); Not Pass AABR w/TH (Child with typical hearing who did not pass their newborn AABR screening, 0 = Passed screening, 1 = Did not pass screen but later diagnosed with typical hearing). All Wald tests have one degree of freedom.

Primary Analyses: Predicting Age 8–10 ASD Identification Based on AABR Screening Results and Hearing Status

The 258 children who did not pass their AABR newborn hearing screen but had documented typical hearing continued to exhibit higher rates of ASD at eight to ten years of age—10.5% versus 1.7% for all other children (OR = 6.64, 95% CI: 4.41–9.99; $\chi^2(1, N = 28{,}938) = 109.95$, $p < .001$). The decrease in the odds-ratio reflects the relative increase in the overall number of identified cases of ASD as children grew older.

Finally, a logistic regression examined this same relationship controlling for sex, age, reported birth defect, birth weight category, and NICU status. As summarized in Table 7, children who did not pass their AABR newborn hearing screen but were diagnosed with typical hearing continued to exhibit higher rates of ASD when eight to ten years old (OR = 5.70, 95% CI: 3.76–8.63), even after controlling for these other factors. Results were similar when substituting school grade-level for age (OR = 5.98, 95% CI: 3.85–9.28).

Table 7

Logistic Regression Predicting Autism Spectrum Disorder (ASD) Identification at 8–10 Years of Age Based on Newborn Automated Auditory Brainstem Response (AABR) Screen and Child Characteristics

Variable	*b*	S.E.	Wald	*p*	OR [95% CI]
Constant	-3.784	0.513	54.42	< .001	0.023
Male	1.508	0.117	166.90	< .001	4.519 [3.595, 5.681]
Age	-0.146	0.056	6.85	0.009	0.864 [0.775, 0.964]
Any Birth Defect	-0.434	0.303	2.05	0.152	0.648 [0.358, 1.174]
ELBW	1.434	0.609	5.55	0.019	4.195 [1.272, 13.836]
VLBW	1.132	0.524	4.68	0.031	3.102 [1.112, 8.656]
LBW	0.395	0.183	4.68	0.031	1.484 [1.038, 2.122]
NICU	0.131	0.182	0.51	0.473	1.140 [0.797, 1.629]
Not Pass AABR w/TH	1.740	0.212	67.34	< .001	5.696 [3.759, 8.631]

Note. ASD Identification (0 = Not identified as having ASD, 1 = Identified as having ASD); Male (0 = Female, 1 = Male); Any Birth Defect (0 = No record of monitored birth defect, 1 = Presence of a monitored birth defect); ELBW (Extremely low birth weight under 1000g, 0 = No, 1 = Yes); VLBW (Very low birth weight, 1000g to 1500g, 0 = No, 1 = Yes); LBW (Low birth weight, 1500g to 2500g, 0 = No, 1 = Yes); NICU (Presence in Neonatal Intensive Care Unit during birth hospitalization, 0 = No, 1 = Yes); Not Pass AABR w/TH (Child with typical hearing who did not pass their newborn AABR screening, 0 = Passed screening, 1 = Did not pass screen but later diagnosed with typical hearing). All Wald tests have one degree of freedom.

Discussion

Based on previous research that found atypical ABR results among some children with ASD (Miron et al., 2018; Rosenhall et al., 2003; Roth et al., 2011; Talge et al., 2018), this study examined whether state-wide, child-level AABR pass/refer results collected by an EHDI program would be related to identification as having ASD at 5 to 10 years of age. By linking newborn hearing screening records and educational records, we were able to identify a cohort and explore the relationship between newborn hearing screening results and identification of ASD at a later age. Results found that newborns who did not pass their AABR hearing screen but were diagnosed with normal/typical hearing were at more than eight times the odds of being identified with ASD at 5 to 7 years of age, and over six times the odds at 8 to 10 years of age.

This study adds to the existing research base in several key ways. First, previous research involved older, clinical-based samples of children and young adults with ASD. In this study we have extended the age-range down to newborn infants. Second, this is the first study to use a population-based sample, suggesting the possibility to further examine a relationship between ABR activity—albeit as more limited pass/refer results—and ASD on an epidemiological, population-level, using data from existing EHDI programs in the United States or elsewhere. Third, while prior research drew on the more rich and detailed data available through ABR testing, this study found a statistically significant association was evident even with the more limited information available in simple pass/refer results provided by AABR screening.

The results are particularly noteworthy because the newborn AABR data and data on ASD status were collected independently and years apart by two different systems, health and education, that do not usually share information. Furthermore, the five to ten year delay between AABR screening and ASD identification, as well

as the focus on children with typical hearing, minimizes the potential for some variation of confirmation bias (i.e., schools were somehow aware that a child with normal/typical hearing did not pass an AABR at birth, and this knowledge influenced their decision to identify the child as having ASD).

It should be noted that this effect continued to be observed after controlling for various early childhood characteristics, including child sex, age, presence of another known birth defect, birth weight, and presence in a NICU during birth hospitalization (which served as a marker for other high-risk birth factors that may be related to both hearing loss and ASD). Although large, the effect size did decrease with age from an adjusted odds ratio of 6.94 at 5 to 7 years of age to an adjusted odds ratio of 5.70 at 8 to 10 years of age. This reflects the relative increase in the number of children identified with ASD as they became older, but it is also possible that these specific children exhibited more clear or severe ASD-related behaviors that resulted in earlier identification. Furthermore, although this study focused on ASD as an outcome, analyses also examined whether an association was seen more broadly based on whether a child was or was not enrolled in any special education classification when 5 to 10 years old. Consistent with the ASD findings, children who did not pass their newborn AABR but were subsequently diagnosed with typical hearing, were significantly more likely to be enrolled in special education in elementary school. This further suggests that although AABR screening compares ABR activity against a template derived from normal-hearing infants, there may be additional signal in the *noise* associated with an AABR refer/pass result that goes beyond hearing loss and may potentially tap into other important areas of child development.

As we note throughout this paper, AABR only provides binary results of pass or refer, and does not provide detailed information regarding wave forms that is available through ABR testing. Clearly, additional research that examines specific waveform patterns in connection with subsequent ASD identification would be valuable. Furthermore, the current study cannot shed light on specific mechanisms or processes through which the observed association between AABR screening at birth and ASD five to ten years later operates. Additional research examining such possible mechanisms would also be valuable.

The widespread availability of AABR screening data via EHDI programs may have a role in these efforts—for example, this may create opportunities for EHDI programs to support and facilitate the work of colleagues in the ASD community, as well as further assist families already touched by EHDI systems. In this regard, we must be perfectly clear that we are not suggesting a change in practice or policy based on a single study, and we are certainly not suggesting that AABR be seen as a diagnostic tool for ASD. However, when a child who did not pass an AABR screen is subsequently diagnosed with typical hearing, it is currently standard practice within the EHDI community to *close the case* and move on. If the findings in this study are supported by additional research, parents and primary health care providers may want to continue to monitor language, behavioral, and cognitive developmental milestones for these children, even after they are diagnosed with typical hearing.

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Private Insurance Reimbursements for Newborn Hearing Screening in the United States, 2013–2014 Birth Cohort

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Abstract

The purpose of this study was to describe private insurance reimbursements for newborn hearing screening (NBHS) in the United States. Data from the MarketScan® Commercial Databases were used to estimate itemized reimbursements for privately insured infants born between January 1, 2013–December 31, 2014. Estimates were based on billed claims for hearing screening services during infancy among 456,407 infants with birth hospitalization claims (71,820 infants with inpatient NBHS and 1,104 infants with outpatient NBHS). The median reimbursement for NBHS was almost three times greater when performed in an inpatient setting than outpatient setting. Median reimbursement for NBHS performed in a hospital and billed as inpatient service was $148.00 (interquartile range [IQR] $99.52–$210.00) and $57.53 (IQR $34.40–$120.91) when billed as an outpatient service. The mean reimbursement for NBHS performed in an outpatient hospital setting was $136.48 (IQR $86.08–$220.15) and $41.60 (IQR $28.15–$57.52) for NBHS billed in conjunction with an office visit (e.g., performed in an audiology clinic, an audiologist's office, or physician's office during a routine check-up). No NBHS claims were filed for 84.3% of infants (384,587/456,407), as NBHS is generally included as a covered service bundled along with delivery and newborn care.

Acronyms: ABR = auditory brainstem response; CPT = current procedural terminology, ICD-9-CM = International Classification of Diseases, Ninth Revision, Clinical Modification; IQR = interquartile range; NBHS = newborn hearing screening; OAE = otoacoustic emissions

Keywords: newborn hearing screening, private insurance reimbursement, hearing loss

Disclaimer: The findings and conclusions in this report are those of the authors and do not necessarily represent the official position of the Centers for Disease Control and Prevention.

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Almost all infants in the United States are screened soon after birth for hearing loss using automated auditory brainstem response (automated ABR) and/or otoacoustic emissions (OAE). Both OAE and automated ABR tests provide non-invasive recordings of physiologic activity underlying normal auditory function for the purpose of confirming the presence or absence of a hearing loss (Wroblewska-Seniuk, Dabrowski, Szyfter, & Mazela, 2017). These reliable and objective methods of testing and screening can be easily performed in newborns and infants, either used alone or in sequence (Joint Committee on Infant Health [JCIH], 2007; Wroblewska-Seniuk et al., 2017).

Little is known about the healthcare cost of newborn hearing screening in the United States. Estimates of the resource cost of hospital-based NBHS in terms of staff time, instruments, and consumables in U.S. hospitals published between 1995 and 2002 ranged from $25 to $50 per infant screened, adjusted for inflation to 2016 U.S. dollars, but more current estimates are lacking (Grosse, Mason, Gaffney, Thomson, & White, 2018). In any case, there may be little relation between resource costs, charges, and reimbursements for hospital services. When NBHS is conducted by hospital staff, there is usually no separate bill and it is bundled in the overall labor and delivery charge (Winston-Gerson & Rousch, 2016). Some hospitals outsource hearing screening services to a contractor, who can bill families and insurers separately. Based on anecdotal parent reports, Winston-Gerson and Rousch (2016) reported a typical charge for NBHS by a contractor is $250 and could be in excess of $500.

An analysis of 2004 insurance claims data reported the average private-sector payer cost of screening for hearing loss in the hospital was $84 (95% confidence interval [CI]: $0–$200) when billed and paid separately from the labor and delivery charge (Grosse, 2006). McManus et al. (2010) reported proprietary estimates of typical direct provider payments by an employer health plan in 2005 was $82.01 for an OAE test with limited evaluation (current procedural terminology [CPT] code 92587) for the sole purpose of confirming the presence or absence of a hearing loss (McManus et al., 2010). The authors of that study did not include the other OAE screening CPT code (92558) in their estimates. The purpose of this

analysis was to provide more up-to-date information on reimbursement rates for privately insured infants who are individually billed for NBHS during infancy in both inpatient and outpatient settings.

Method

Data Source

This retrospective analysis used claims data from the IBM® MarketScan® Commercial Research Databases from 2013 through 2015. The commercial databases include employer-sponsored insurance claims data for approximately 30 to 40 million employees and their beneficiaries each year from all U.S. states. The databases contain fully integrated, de-identified, individual-level data across the entire continuum of care (e.g., inpatient, outpatient, outpatient pharmacy, laboratory) that capture real-world treatment patterns and expenditures (Truven Health Analytics, 2017). Each enrollee is assigned a de-identified unique number, allowing linkage across claims over time. MarketScan data is de-identified and their analysis is not classified by the Centers for Disease Control and Prevention as human subjects research and has been determined not to require an Institutional Review Board.

Claims were identified using the International Classification of Diseases, Ninth Revision, Clinical Modification (ICD-9-CM) codes (Table 1). Inpatient and outpatient data were extracted from MarketScan Research Databases (2013–2015) for infants born between January 1, 2013 and December 31, 2014, who were individually billed for NBHS, did not die during the study period, and had a first claim with a delivery code. The analysis included the following information: birth year, gender of patient (male/female), setting (inpatient/outpatient), outpatient place of service, census division, diagnoses, procedures, service date, procedure age (days), net payment, and health plan type. An algorithm (Figure 1 and Table 1) was used to create a proxy birth date using the admission date of the first inpatient claim for the baby containing a delivery code (ICD-9-CM: V30-31, V33-V34, V36-V37, and V39). We analyzed three CPT codes typically used for hearing screening (American Academy of Pediatrics, 2016): 92586 (automated ABR), 92558 (screening OAE), and 92587 (distortion product evoked OAE or OAE with limited evaluation). See Table 1 for detailed descriptions. OAE comprehensive diagnostic evaluation code 92588, used to bill for a test to determine the amplitude level of an otoacoustic emission output at each discrete frequency and not to determine the presence or absence of a hearing loss, was not examined. Service date was the date when the procedure or service occurred.

Table 1
List of Newborn Birth and Hearing Screening Codes

Code(s)	Code Description
Newborn ICD-9-CM Delivery Codes	
V30-31, V33-V34, V36-V37, V39	Live birth
Newborn Hearing Screening CPT Codes	
92586 Automated ABR	Auditory evoked potentials for evoked response audiometry and/or testing of the central nervous system; limited
92558 Screening OAE	Evoked otoacoustic emissions, screening; qualitative measurement of distortion product or transient evoked otoacoustic emissions, automated analysis
92587 OAE Limited Evaluation	Distortion product evoked otoacoustic emissions; limited evaluation (to confirm the presence or absence of hearing disorder, 3–6 frequencies) or transient evoked otoacoustic emissions, with interpretation and report

Note. ABR = automated auditory brainstem response; CPT = current procedural terminology codes; ICD-9-CM = International Classification of Diseases, 9th Revision, Clinical Modification; OAE = otoacoustic emissions.

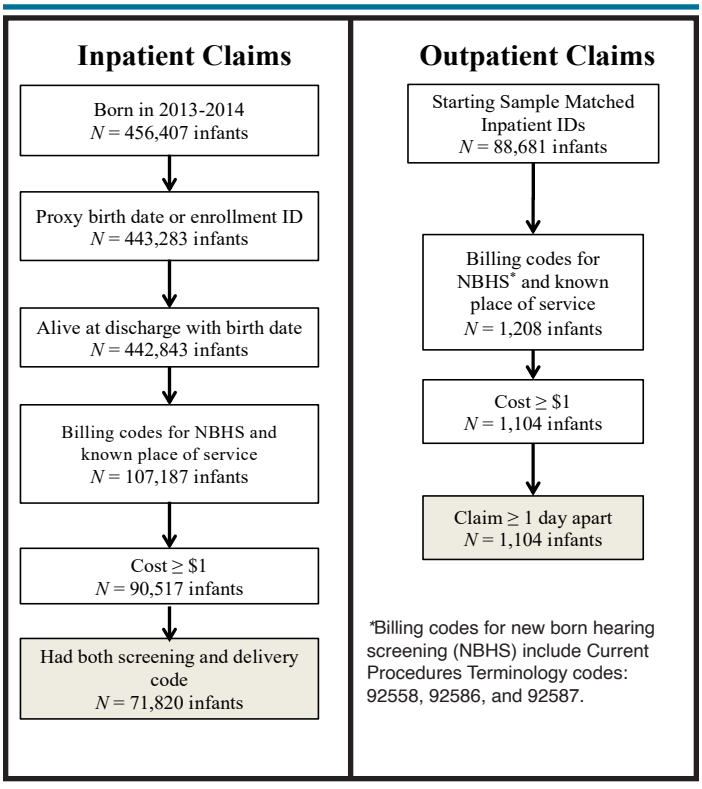

Figure 1. Flowchart depicting the selection process of inpatient (left) and outpatient claims (right) included in the present study. CPT = current procedural terminology.

Procedure age (days) was estimated using the difference between service date and proxy birth date. Net payment is defined as the payment received by the provider, excluding patient out-of-pocket and coordination of benefits. Claims were categorized as inpatient or outpatient using the place of service code. For inpatient claims, the place of service codes included inpatient hospital, hospital emergency room, and birthing center. Inpatient claims represent billing as occurring in the hospital inpatient setting when a patient was admitted into the hospital and a service was provided during the hospital stay. The outpatient place of service codes included outpatient hospital and office. Outpatient services can occur after an infant has been discharged from a hospital or birthing center. In the case of NBHS, the outpatient service can be a repeat or an initial screen. When place of service is coded as an office visit, the service can occur in an audiology clinic, an audiologist's office, or a physician's office during a routine well child visit. When the place of service is coded as outpatient hospital, the infant is receiving the service as an outpatient at a hospital-owned facility. Claims were categorized as nine census divisions defined by the U.S. Census Bureau (New England, Middle and South Atlantic, East and West North Central, East and West South Central, Mountain, and Pacific), and unknown region.

Data Analysis

In this descriptive analysis, all claims for services that an individual received on a given service date are assumed to refer to a single encounter. The proxy birth date was used to limit claims to the first year of life (infancy), that is, difference in days between service date and birth date (< 365 days). Mean, median, range, and interquartile range (IQR) of the net payments were calculated by summing each claim. Medical expenditures were adjusted for inflation to 2014 dollars and reported by care setting and place of service.

Claims were excluded if (a) enrollment ID was missing; (b) the infant died before discharge; (c) the difference between the service date and proxy birth date was a negative number (i.e., screening occurred before proxy birth date in which proxy birth date could not be determined); (d) infant was not individually billed for NBHS or place of service was unspecified; and (e) sum of the net payment for a single encounter was equal to or less than $1 irrespective if the claim was denied or reimbursed. Claims presumed to be duplicates of the initial claim were also excluded (claims with similar dates and billing codes). Inpatient claims were limited to those occurring during birth hospitalization (containing both NBHS and delivery codes). All analyses were conducted using SAS software version 9.4 (SAS Institute Inc., Cary, NC). Descriptive statistics (frequency counts and percentages) were used to compare mean and median reimbursement rates and IQRs by setting (inpatient/outpatient), outpatient place of service, and census division.

Results

Among 456,407 privately insured infants born during 2013–2014, 71,820 (15.7%) had inpatient claims for NBHS. Of those infants, 1,104 (1.5%) also had outpatient claims for NBHS (Table 2 and Figure 1). Mean reimbursement rates for NBHS were higher than the median reimbursement rates (Table 2). Median reimbursement for NBHS (IQR) performed in a hospital setting was $148.00 ($99.52–$210.00) billed as an inpatient service, and $57.53 ($34.40–$120.91) billed as an outpatient service. The median reimbursement for NBHS (IQR) was $136.48 ($86.08–$220.15) for an outpatient service in a hospital facility and $41.60 ($28.15–$57.52) for NBHS billed in conjunction with an office visit (Table 2).

Table 2

Descriptive Summary of Individually Billed Newborn Hearing Screening Claims for Infants Born 2013–2014

Variable	Newborn Hearing Screening	
	Inpatient	Outpatient
	n (%)	*n* (%)
Total Claims	72,146	1,300
Total Enrollees	71,820	1,104
Mean number of Claims (Range)	1.0 (1.0–1.0)	1.2 (1.0–4.0)
Net Payment Reimbursements		
Mean Net Payment (Range)	$159.46 ($1.04–$1580.10)	$96.89 ($2.03–$1320.78)
Median Net Payment (IQR)	$148.00 ($99.52–$210.00)	$57.53 ($34.40–$120.91)
Mean Net Payment for Outpatient Place of Service (Range)		
Office	N/A	$50.68 ($4.11–$714.00)
Outpatient Hospital	N/A	$169.87 ($2.03–$1320.78)
Median Net Payment for Outpatient Place of Service (IQR)		
Office	N/A	$41.60 ($28.15–$57.52)
Outpatient Hospital	N/A	$136.48 ($86.08–$220.15)
Gender of Patient		
Male	37,403 (52.1)	608 (55.1)
Female	34,417 (47.9)	496 (44.9)

Note. IQR = Interquartile Range.

Reimbursement rates for NBHS varied significantly by procedure and setting (Table 3). For inpatient NBHS and outpatient office visit NBHS, mean and median reimbursements for automated ABR (CPT 92586) were substantially higher than OAE hearing screening (CPT 92558 or 92587). The same was true for outpatient hospital-based claims, with reimbursements for automated ABR (CPT 92586) slightly higher than for OAE hearing screening (CPT 92558 or 92587). About half of the inpatient claims for NBHS (49.4%, 249/504) were for automated ABR. Most of outpatient claims for NBHS (94.2%, 750/796) were for OAE hearing screening services. The median reimbursement (IQR) for automated ABR was $150.00 ($104.40–$210.68) when billed as an inpatient screen, $102.18 ($75.81–$169.13) as an office hearing screen, and $164.34 ($94.02–$254.00) as an outpatient hospital screen. The median reimbursement (IQR) for screening OAE tests (CPT 92558) or OAE with limited evaluation (CPT 92587) was $57.80 ($29.37–$108.68) when billed as an inpatient service, $39.74 ($27.63–$52.54) as an office screening service, and $116.90 ($78.22–$178.27) as a hospital outpatient service.

In the outpatient setting, reimbursement rates were higher for OAE hearing screening (CPT 92558 or 92587) and automated ABR (CPT 92586) occurring as an outpatient hospital visit than an office visit, where hearing screens were performed in an audiology clinic, an audiologist's office, or a physician's office (Table 3). Irrespective of outpatient place of service, reimbursement for automated ABR (CPT 92586) was higher than OAE hearing screening (CPT 92558 or 92587).

Table 3

Unweighted Inpatient and Outpatient Hearing Screening Reimbursement Rates for Newborns Born between 2013 and 2014*

Inpatient Hearing Screen (n = 72,176 claims)			
CPT Codes	***n* (%)**	**Mean (Range)**	**Median (IQR)**
92558 or 92587 OAE	2,228 (3.1)	$73.38 ($2.25–$1121.48)	$57.80 ($29.37–$108.68)
92558 only	66 (0.1)	$63.86 ($8.40–$293.61)	$60.35 ($21.92–$95.85)
92587 only	2,162 (3.0)	$73.67 ($2.25–$1121.48)	$57.14 ($29.65–$109.24)
92586 Automated ABR	69,948 (96.9)	$162.20 ($1.04–$1580.10)	$150.00 ($104.40–$210.68)
Outpatient Newborn Hearing Screen (n = 1,300 claims)			
CPT Codes	***n* (%)**	**Mean (Range)**	**Median (IQR)**
Office			
92558 or 92587	255 (19.6)	$45.42 ($4.11–$360.00)	$39.74 ($27.63–$52.54)
92586	249 (19.2)	$136.33 ($31.71–$714.00)	$102.18 ($75.81–$169.13)
Outpatient Hospital			
92558 or 92587	750 (57.7)	$155.98 ($2.03–$1320.78)	$116.90 ($78.22–$178.27)
92586	46 (3.5)	$184.09 ($2.43–$650.00)	$164.34 ($94.02–$254.00)
Irrespective of outpatient place of service			
92558	39 (3.0)	$41.98 ($6.85–$176.27)	$33.00 ($12.78–$60.00)
92587	966 (74.3)	$74.75 ($2.03–$1320.78)	$47.64 ($30.74–$83.98)
92586	295 (22.7)	$176.65 ($2.43–$714.00)	$156.90 ($87.23–$250.00)

Note. CPT = Current Procedural Terminology; IQR = Interquartile Range.

*IBM® MarketScan® Commercial Databases for 2013–2015

As shown in Tables 4 and 5, the average reimbursement rates varied by census division. For inpatient hearing screens, the South Atlantic had the highest median reimbursement rate and East South Central had the lowest median reimbursement rate ($196.02, IQR $98.74–$239.14 and $107.93, IQR $85.22–$160.00, respectively; Table 4). For outpatient hearing screens, the lowest median reimbursement rates ranged from $32.02 (IQR $24.00–$58.28) in the West South Central to the highest $158.56 (IQR $57.52–$210.00) in the Pacific (Table 5). The census division with the highest median reimbursement for an outpatient service in a hospital facility and office visit were Middle Atlantic ($195.57, IQR $105.20–$254.00) and Pacific ($49.25, IQR $31.71–$57.52; Table 5).

Discussion

Our estimates of average reimbursement for NBHS by private insurers for screening conducted in birth hospitals are substantially greater than published estimates of the resource costs of providing such services. Published U.S. cost estimates for pre-discharge hospital screening have generally been in the range of $27 to $47 per infant

Table 4

*Summary of Inpatient Net Payment Reimbursement by Census Division**

Census Division	Inpatient Newborn Hearing Screening (*n* = 72,146 claims)		
	No. of Claims	Mean (Range)	Median (IQR)
New England	1,208	$175.11 ($2.57–$980.70)	$120.17 ($109.09–$215.10)
Middle Atlantic	6,493	$177.17 ($2.52–$1106.50)	$144.00 ($109.60–$239.00)
East North Central	6,680	$127.27 ($1.63–$840.00)	$116.00 ($104.21–$139.82)
West North Central	2,377	$117.54 ($2.80–$714.00)	$113.51 ($90.00–$135.00)
South Atlantic	9,735	$178.18 ($1.23–$1121.48)	$196.02 ($98.74–$239.14)
East South Central	5,718	$123.99 ($1.41–$490.04)	$107.93 ($85.22–$160.00)
West South Central	19,273	$185.79 ($1.15–$1580.10)	$185.00 ($148.00–$246.46)
Mountain	12,506	$139.02 ($1.32–$478.00)	$136.18 ($90.19–$179.25)
Pacific	7,505	$152.90 ($1.04–$576.78)	$143.40 ($81.42–$215.00)
Unknown Region	681	$157.17 ($5.31–$714.00)	$148.00 ($104.49–$204.30)

Note. IQR = Interquartile Range.

* IBM® MarketScan® Commercial Databases for 2013–2015

Table 5

*Summary of Outpatient Net Payment Reimbursement by Census Division**

Census Division	Newborn Hearing Screening Outpatient (*n* = 1,300 claims)				
	All Outpatient Claims			Outpatient by Place of Service	
	No. of Claims	Outpatient Mean (Range)	Outpatient Median (IQR)	Outpatient Hospital Median (IQR)	Office Median (IQR)
New England	25	$99.04 ($21.59–$482.11)	$66.40 ($40.23–$145.09)	$151.18 ($69.27–$164.21)	$40.23 ($40.23–$60.56)
Middle Atlantic	289	$100.92 ($2.43–$734.25)	$54.92 ($39.50–$121.50)	$195.57 ($105.20–$254.00)	$45.18 ($34.00–$54.92)
East North Central	95	$96.56 ($17.99–$1320.78)	$57.60 ($42.88–$93.33)	$102.66 ($81.48–$182.47)	$44.80 ($32.42–$57.60)
West North Central	26	$65.51 ($12.78–$176.27)	$47.37 ($39.65–$91.50)	$82.35 ($39.65–$105.00)	$45.00 ($21.00–$78.00)
South Atlantic	227	$113.32 ($4.11–$714.00)	$69.59 ($37.75–$140.18)	$169.06 ($93.25–$293.78)	$47.59 ($32.36–$81.23)
East South Central	92	$88.45 ($4.69–$640.80)	$51.82 ($33.90–$97.87)	$128.21 ($98.09–$287.66)	$38.57 ($33.90–$55.90)
West South Central	230	$62.04 ($4.43–$1122.66)	$32.02 ($24.00–$58.28)	$103.41 ($60.91–$182.59)	$30.48 ($21.34–$43.59)
Mountain	181	$78.67 ($2.03–$339.08)	$72.56 ($41.92–$106.77)	$94.02 ($72.56–$131.08)	$41.97 ($24.97–$61.10)
Pacific	126	$78.67 ($22.71–$550.00)	$158.56 ($57.52–$210.00)	$175.00 ($138.53–$281.86)	$49.25 ($31.71–$57.52)

Note. CPT = Current Procedural Terminology; IQR = Interquartile Range. Results for unknown region (*n* = 9) are not shown because of small numbers.

*IBM® MarketScan® Commercial Databases for 2013-2015

screened, adjusted for inflation to 2016 U.S. dollars (Grosse et al., 2018). In contrast, average inpatient NBHS reimbursements reported here, with IQR from $100 to $210 (Table 2), are several times as high.

Our retrospective analysis of the private insurance reimbursements rate for NBHS services using IBM® MarketScan® Commercial Research Databases (2013–2015) complements previous analyses (Grosse, 2006; McManus et al., 2010). There are a limited number of NBHS cost studies specifically looking at the reimbursement rate using the procedure codes. Whereas McManus et al. (2010) investigated the Medicaid reimbursement rates for all types of hearing services for infants and young children, our study provides reimbursement estimates by setting and type of screening services for privately insured infants. McManus et al. (2010) reported mean Medicaid reimbursement rates of $106.30 for automated ABR (CPT 92586) and $99.40 for OAE with limited evaluation (CPT 92587 adjusted for inflation to 2014 dollars) irrespective of inpatient or outpatient setting. Our mean estimates for automated ABR (CPT 92586: $162.26, range $1.04–$1580.10) and OAE with limited evaluation (CPT 92587: $74.00, range $2.03–$1320.78) irrespective of inpatient or outpatient setting

were higher for privately insured infants than for infants with Medicaid (results not shown).

It should be emphasized that the vast majority (84.3%) of privately insured infants who received a hospital-based NBHS were not separately billed for the service because the cost of providing a hearing screen for a newborn is typically bundled under the newborn delivery care charge. Consequently, the reimbursements reported here do not characterize how much hospitals are reimbursed for NBHS. The reimbursement rate reported here in most, if not all, cases reflect reimbursements to independent providers or contractors contracted to perform NBHS.

We were unable to find studies on the estimated resource cost of conducting screening by an independent provider or contractor contracted to perform NBHS. In contrast, we found several older studies that have reported resource cost estimates associated with NBHS conducted by hospital staff (Kezirian, White, Yueh, & Sullivan, 2001; Maxon, White, Behrens, & Vohr, 1995; Mehl & Thomson, 1998; Vohr et al., 2001). Kezirian et al. and Vohr et al. estimates were based on direct cost of the equipment, overhead, and all personnel cost including clerical administrative assistance cost. Kezirian et al. reported the cost of providing an OAE hearing screen was $13 per infant and the cost for an automated ABR hearing screen was $25 per infant. Vohr et al. reported $28.69 for an OAE hearing screen and $32.81 for an automated ABR hearing screen. Adjusting to 2014 dollars, the cost of providing an OAE hearing screen would range from $17.38 to $38.25 and the cost of an automated ABR screen would range from $33.42 to $43.86. These costs would not accurately describe the cost for independent providers or contractors contracted to perform NBHS. Since those cost estimates are very old, it is not clear that adjustment for inflation is sufficient. It would be helpful to have estimates from new hearing screening cost studies.

Reimbursement rates appeared to be dependent on the type of hearing screening service and place of service. Unlike previous studies that reported only mean reimbursements, this analysis provided means, medians, range, and IQRs for reimbursements. The median, unlike the mean, is not influenced by a small number of extremely large or small values. Therefore, the median net payment may provide a better estimate of the *typical* inpatient reimbursement.

This study provides new cost information on how the two screening methods were used across places of service, OAE, and automated ABR. Almost half (49.4%) of privately insured infants who were individually billed for NBHS as an inpatient received an automated ABR screen, while almost all (94.2%) infants who were individually billed for outpatient NBHS received an OAE screen (Table 3). The decision to use ABR screening equipment by a hospital for inpatient screening could be driven by both best practice considerations and the higher reimbursement rate relative to OAE hearing screen. On average, the claim for an automated ABR screen performed as an inpatient service (median payment) was reimbursed 2.6 times higher than for an OAE hearing screen performed in the same setting. The ratio of reimbursements between the two types of service was also the same for office visit claims, yet only half of inpatient visit claims were for OAE.

We found the median net payment per claim for NBHS was almost three times as high for inpatient as for outpatient claims (Table 2). This appears to largely reflect differences in the relative shares of automated ABR and OAE screening types between inpatient and office visits. Within those settings there were much smaller differences in reimbursements by service type. In hospital outpatient claims, reimbursements were similarly high for both service types (Table 3). Separately reporting outpatient and inpatient reimbursements provides a more comprehensive and accurate summary of the variability in reimbursement rates by type of service.

The higher average reimbursement for automated ABR than OAE hearing screening services performed as an inpatient service in the hospital is consistent with some published estimates of resource costs (Kezirian et al., 2001; Lin et al., 2005; Lin, Shu, Lee, Lin, & Lin, 2007). Performing automated ABR requires the use of disposable electrodes, which is not required for an OAE hearing screening procedure. The electrode supply adds to the total cost of providing an automated ABR hearing screen. However, a few studies reported little cost difference between automated ABR and OAE hearing screening services (Lemons et al., 2002; Vohr et al., 2001).

The median and mean reimbursements for an automated ABR screen performed for an outpatient hospital service, $164.34 and $184.09, were higher than the reimbursement rate in an office setting, $102.18 and $136.33, but similar to the inpatient hospital reimbursement rate of $150.00 and $162.20. For OAE hearing screens, the median and mean reimbursements were lower when conducted in an office setting, $39.74 and $45.42, than in an inpatient setting, $57.80 and $73.38. The highest reimbursement rate for OAE screening service took place in a hospital setting as an outpatient service, $116.90 and $155.98. In the inpatient and outpatient settings, reimbursements were lower for OAE than automated ABR hearing screen.

We were unable to find any previously published cost study specifically looking at the cost of providing hearing screening in an office as the place of service after infants have been discharged from the hospital. We were able to find the cost for providing *post-discharge* hearing screens in five hospitals in one study (Vohr et al., 2001). Vohr et al. reported the cost for providing an OAE screen as $66.87. According to our analysis, an outpatient OAE screen performed in a hospital setting was reimbursed at a median rate of $116.90 and a mean of $155.98 for privately insured infants. The cost for providing an ABR screen was reported as $95.04 (adjusted to 2014 dollars) by Vohr et al. and we found the median and mean reimbursement rates for an automated ABR screen (CPT 92856) were $102.18 and $136.33 respectively.

This analysis has several limitations. First, billing codes are subject to coding errors (O'Malley et al., 2005), which

means that some claims for what appear to be NBHS may actually be for a different service. Second, we examined claims data from 2013 to 2015 for the 2013–2014 birth cohort using ICD-9-CM codes to avoid the coding transition to ICD-10 on October 1, 2015. However, the claims data are now more than 4 years old and may be a bit dated. The estimates may have changed since the study was completed in 2017. Finally, the data used in this study comes from employer-based plans and cannot be generalized to other types of private payers. The MarketScan Commercial data have been found to be comparable in demographics to the U.S. population with employer-sponsored insurance (Aizcorbe et al., 2012), which in turn comprises more than 90% of the U.S. population with private insurance. However, MarketScan data cannot be generalized to populations with public insurance or no insurance.

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Neonatal Abstinence Syndrome and Infant Hearing Assessment: A Kids' Inpatient Database Review

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Abstract

Objective: Neonatal abstinence syndrome (NAS) has become an epidemic. This study assesses documented rates of failed newborn hearing screening (NBHS) or hearing loss (HL) diagnosis in NAS infants, and sociodemographic factors associated with abnormal inpatient hearing results.

Method: The 2016 HCUP/KID national database was used to identify a weighted sample of infants with failed NBHS/HL during birth hospitalization. Independent variables included diagnoses of NAS/in-utero opioid exposure, HL risk factor presence, and sociodemographic data. Univariate analyses and multivariate logistic regression were used to determine associations between NAS and abnormal hearing assessment.

Results: NAS infants had lower odds ratio (OR) of documented failed NBHS (OR = 0.76, $p < 0.05$) than controls, but a higher rate of HL diagnosis (OR = 2.17, $p < 0.01$). Certain sociodemographic factors had higher OR of abnormal hearing results, including race ($p < 0.001$) (Black, OR = 1.48 and Native American, OR = 1.83), and Medicaid coverage (OR = 1.45, $p < 0.001$). A lower OR of HL diagnosis was observed in females (OR = 0.84, $p < 0.001$) and infants with higher household income (OR = 0.53, $p < 0.01$).

Conclusion: NAS children have lower rates of inpatient documented failed NBHS and higher rates of HL diagnosis. The complex medical care of these infants could complicate NBHS, documentation, and subsequent follow-up. Certain sociodemographic factors result in a higher risk of hearing loss.

Acronyms: EHDI = Early Hearing Detection and Intervention; HCUP = Healthcare Cost and Utilization Project; HL = hearing loss; NAS = neonatal abstinence syndrome; KID = Kids' Inpatient Database; NBHS = newborn hearing screening; OR = odds ratio

Keywords: Neonatal abstinence syndrome, hearing loss, newborn hearing screen

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Neonatal abstinence syndrome (NAS) is a condition of the newborn in which withdrawal signs and symptoms are displayed following exposure to an offending medication and/or drug of abuse (McQueen & Murphy-Oikonen, 2016). The vast majority of cases are due to *in-utero* exposure from maternal use of opioids during pregnancy, and like all problems stemming from the opioid epidemic, the incidence of NAS has increased (Patrick et al., 2015). This has placed strain on not only an increasing number of patients, families, and caretakers, but has also been responsible for a large economic burden within the healthcare system, estimated at $1.5 billion in 2012 (Patrick et al., 2015).

There has been little to no focus on this population within the otolaryngology literature. As such, the needs of an NAS patient within this specialty are not well defined. Prior studies have shown NAS patients to be at risk for poor use of prophylactic and specialty care (Fang et al., 2015; Gill et al., 2007; Kivisto et al., 2014; Payot & Berner, 2000). Poor healthcare utilization and being lost to follow-up is a concern for any condition, but this is especially true with newborn hearing loss. There have been no reports of

clinically significant teratogenic effects of opioids leading to hearing loss in a newborn, nor has the rate of hearing loss in the NAS population ever been specifically assessed. However, there have been reports in adults of opioid use causing sensorineural hearing loss (Friedman et al., 2000; Ho et al., 2007; Rigby & Parnes, 2008; Vorasubin et al., 2013). Infants with NAS are a significantly vulnerable population and deserve special attention as they may face barriers to hearing healthcare after birth.

The prevalence and the lifelong effects of unrecognized hearing impairment in a newborn have been the driving force to support universal newborn hearing screening. The Early Hearing Detection and Intervention (EHDI) guidelines state that children should undergo hearing screening by 1 month of age, receive audiologic testing and diagnosis by 3 months if testing indicates hearing impairment, and have an intervention as indicated by 6 months. These recommendations were made by the U.S. Preventive Task Force and Joint Committee on Infant Hearing (JCIH) Loss over a decade ago and have been underscored by studies demonstrating improved speech and language outcomes in children whose care benefitted from earlier detection as advised by EHDI guidelines (JCIH, 2007; Kennedy et al., 2006; U.S. Preventive Services Task Force, 2008; Yoshinaga-Itano, 2017). Recently JCIH updated their position statement, recommending that EHDI programs consider a new target where hearing screening would occur by 1 month of age, audiologic testing and diagnosis by 2 months of age, and intervention as indicated by 3 months (JCIH, 2019). Achieving this goal can prove difficult, especially in patient populations at risk for poor follow-up and healthcare use. Newborn hearing screening and subsequent follow-up testing can be complicated by many barriers, making it difficult for patients and families to navigate. Over 98% of children within the United States undergo NBHS (CDC, 2015); however, nearly 60% of infants fail to obtain a timely diagnosis after abnormal screening (CDC, n.d.). Certain sociodemographic factors, such as insurance status and parental education level, have been associated with decreased use of audiologic services following a failed newborn hearing screening (Folsom, 2000; Liu, 2008; Oghalai, 2002; Spivak, 2009). Communication of failed NBHS is important for continuity of care of timely diagnosis of infant hearing loss, however, many primary care providers and parents are either uninformed or misinformed about NBHS results (Bush, Alexander, et al., 2015, Bush, Hardin, et al., 2015). Birthing hospitals are mandated to report NBHS results to state Early Hearing Detection and Intervention (EHDI) agencies; however, the documentation and subsequent billing of abnormal NBHS on birth inpatient records on a local level is largely unknown. A disconnect between the NBHS results and inpatient records could influence continuity of care for the infant after hospital discharge. The relationship of NAS and infant hearing screening results and/or documented diagnosis of hearing loss has not been previously described. The primary aim of this study was to assess the documented rate of failed newborn hearing screenings and diagnoses of hearing loss in NAS patients during their birth hospitalizations. Furthermore, by using a large national inpatient admissions database, we aimed to assess the association of NBHS screening results and patient demographics and socioeconomic factors.

Materials and Method

The study uses publicly available data that is deemed by the Institutional Review Board (IRB) as not involving human subjects and not requiring IRB review and approval.

Study Sample

The study examines the association between NAS and NBHS screening results in 2016 using the Kids' Inpatient Database (KID), Healthcare Cost and Utilization Project (HCUP), Agency for Healthcare Research and Quality. The database includes relevant diagnostic and procedure codes, as well as demographic data, for a national sample of pediatric inpatient hospitalizations. Pediatric inpatient admissions were included in the study sample if the admission was associated with a hospital birth, where NBHS is expected to be performed and where NAS could be detected. Admissions associated with patients at risk for iatrogenic NAS were excluded using methods described in prior studies (Patrick et al., 2012). The study also excludes admissions with Medicare as primary payer and where data on demographic variables of interest were missing.

Measures

NAS was identified using the International Classification of Diseases, 10th ed. [ICD-10] code P96.1. The study uses two measures of potential hearing loss, a failed NBHS or a diagnosis of hearing loss during the inpatient birth admission. A failed NBHS was identified using ICD-10 codes R94.120, R94.8, and Z01.110. Diagnosed hearing loss was determined using ICD-10 codes H91.90, H90.3, H90.41, H90.42, H90.71, H90.72, H90.6, H90.2, H90.11, H90.12, H90.0, H90, H90.1, H90.4, H90.5, H90.7, H90.8, H90.A, H90.A1, H90.A11, H90.A12, H90.A2, H90.A21, H90.A22, H90.A3, H90.A31, H90.A32, H91, H91.0, H91.01, H91.02, H91.03, H91.09, H91.8, H91.8X, H91.8X1, H91.8X2, H91.8X3, H91.8X9, H91.9, H91.91, H91.92, H91.93, and H91.3.

To control for the potential effects of known risk factors for hearing loss on a failed NBHS or hearing loss diagnosis, a variable indicating the presence of any known risk factor was developed. This indicator variable denotes whether or not sepsis, bacterial meningitis, jaundice, cytomegalovirus, syphilis, rubella, herpes, craniofacial anomalies, or persistent pulmonary hypertension were present diagnoses in the birth admission. ICD-10 codes were used to identify these diagnoses and are available upon request. There are other risk factors, including family history of hearing loss, not used to construct these variables due to lack of a diagnostic code to identify that the risk factor was present. Additional demographic measures included in the study include payer type, race, gender, urban/rural residence, and median household income, all of which are available in the KID.

Statistical Approach

All statistical analyses were performed using Stata 15 (StataCorp LL, College Station, Texas). The construction

of the KID and its sampling approach are described on the HCUP website (https://www.hcup-us.ahrq.gov/tech_assist/sampledesign/508_compliance/index508_2018.jsp). Statistical analyses use sampling weights to account for the KID's complex design and to calculate accurate standard errors. We performed descriptive univariate analyses to summarize the characteristics of the study sample, using chi-square tests to assess differences in demographic variables for the groups with and without a failed NBHS. We used multivariate logistic regression to test for associations between NAS diagnosis and either a failed NBHS or hearing loss diagnosis, controlling for demographic characteristics and the presence of any risk factors for hearing loss. We further examined, again using logistic regression, the association between NAS and, separately, each measure of hearing loss: a failed NBHS and a diagnosis of hearing loss. We report odds ratios (OR) with 95% confidence interval (CI) and a level of significance at alpha = 0.05.

Results

The weighted study sample included 1,113,150 observations, of which 0.67% (n = 21,888) had a diagnosis of NAS. Approximately 0.71% (n = 23,185) of all infants had ICD-10 codes indicating either abnormal NBHS or diagnosis of hearing loss on inpatient birth records. Among those infants with NAS, 117 had a failed NBHS and 15 had a HL diagnosis; none had both. The incidence of documented failed NBHS/hearing loss diagnosis in the NAS cohort was 0.6% (n = 133), and not statistically different, compared to 0.7% (n = 23,051) in the unexposed cohort (p = 0.23). This is summarized in Table 1 along with other patient-specific factors including race and sex. Higher rates of documentation of failed NBHS were seen in males ($p < 0.001$) and Black and Native American infants ($p < 0.001$). When assessing for an association between documented inpatient failed NBHS or hearing loss diagnosis and socioeconomic factors, statistically significant differences were seen based on patient insurance status, primary place of residence, and familial income levels. These findings are shown in Table 2.

Table 1

Diagnosis of NAS and Patient Demographics and Association with Failed Newborn Hearing Screen or Hearing Loss Diagnosis During Birth Hospitalization (Weighted Estimates)

		Abnormal Auditory Function Diagnosis or Hearing Loss Diagnosis				
		No		Yes		
		n	%	*n*	%	*p*-value
NAS	No	3,234,872	99.3%	23,051	0.7%	0.2329
	Yes	21,754	99.4%	133	0.6%	
Risk †	No	3,214,278	99.3%	22,943	0.7%	< 0.05
	Yes	42,348	99.4%	241	0.6%	
Sex	Male	1,665,967	99.2%	12,857	0.8%	< 0.001
	Female	1,590,659	99.4%	10,328	0.6%	
Race	White	1,693,045	99.4%	10,916	0.6%	< 0.001
	Black	466,282	99.1%	4,410	0.9%	
	Hispanic	655,020	99.2%	4,968	0.8%	
	Asian or Pacific Islander	201,572	99.5%	1,044	0.5%	
	Native American	22,442	98.7%	291	1.3%	
	Other	218,262	99.3%	1,553	0.7%	

Note. NAS = neonatal abstinence syndrome.
†Risk=Presence of known medical risk factor for hearing loss

Multivariate logistic regression analyses reveal several findings. When a failed NBHS and hearing loss diagnosis are combined as the outcome variable, we identify no statistically significant association between NAS and hearing loss (OR = 0.82, p = 0.11, data not shown). This analysis includes a weighted sample of 22,327 infants with a failed NBHS, 844 with a HL diagnosis, and 12 with both.

However, when separate regressions are performed for each measure, this study reveals more meaningful associations. Patient demographics and socioeconomic factors and odds of a failed NBHS (i.e. abnormal auditory function diagnosis) or hearing loss diagnosis are summarized in Tables 3 & 4, respectively. When controlling for confounding variables, infants with NAS had a lower odds ratio of documentation of abnormal NBHS (OR = 0.76, $p < 0.05$) compared with non-NAS infants. There is also a statistically significant difference in the odds of

Table 2

Socioeconomic Characteristics and Association with Failed Newborn Hearing Screen or Hearing Loss Diagnosis During Birth Hospitalization (Weighted Estimates)

		Abnormal Auditory Function Diagnosis or Hearing Loss Diagnosis				
		No		Yes		
		n	%	*n*	%	*p*-value
Payer Type	Medicaid	1,518,324	99.2%	12,204	0.8%	< 0.001
	Private Insurance	1,499,297	99.4%	9,325	0.6%	
	Self-Pay	146,617	99.3%	1,083	0.7%	
	No Charge	1,590	98.6%	23	1.4%	
	Other	90,796	99.4%	548	0.6%	
Patient Geography †	Central metro	1,123,029	99.4%	7,017	0.6%	< 0.001
	Fringe metro	781,982	99.4%	5,110	0.6%	
	Mid-metro	643,885	99.2%	5,149	0.8%	
	Small metro	270,852	99.2%	2,274	0.8%	
	Micropolitan	262,985	99.1%	2,273	0.9%	
	Not metro- or micropolitan	173,890	99.2%	1,359	0.8%	
Income Quartile	1st (< $25,000)	953,931	99.2%	7,640	0.8%	< 0.001
	2nd ($25,000–$34,999)	803,257	99.3%	5,958	0.7%	
	3rd ($35,000–$44,999)	797,211	99.3%	5,719	0.7%	
	4th (> $44,999)	702,226	99.5%	3,866	0.5%	

†Patient Geography: Central metro = county population > 1 million; Fringe metro = co. pop. > 1 million; Mid-metro = co. pop. 250,000–999,999; Small-metro = co. pop. 50,000–249,999; Micropolitan = co. pop. 49,999–10,000; Not metro- or micropolitan = co. pop. < 10,000.

diagnosed hearing loss between NAS infants and non-NAS infants (OR = 2.17, $p < 0.01$). Sociodemographic factors with higher odds of abnormal NBHS results included Medicaid insurance status (OR = 1.27, $p < 0.001$), Black race (OR = 1.48, $p < 0.001$), Native American race (OR = 1.83, $p < 0.01$), and smaller metropolitan residence (OR = 1.33–1.44, $p < 0.05$). Factors with lower odds ratio of abnormal NBHS results included female gender (OR = 0.85, $p < 0.001$) and presence of a medical risk factor for hearing loss (OR = 0.69, $p < 0.001$). There are no observed associations between family income and an abnormal NBHS. Sociodemographic factors with higher odds ratio of diagnosis of hearing loss during birth admission records included Medicaid as the primary payer (OR = 1.45, $p < 0.001$) and presence of a medical risk factor for hearing loss (OR = 3.02, $p < 0.001$). Other factors with lower odds ratio of diagnosis of hearing loss were female gender (OR = 0.84, $p < 0.001$) and family income over $45,000 (OR = 0.53, $p < 0.01$).

Discussion

The most recently available data from the Centers for Disease Control and Prevention estimates a rate of failed newborn hearing screen at 1.7% with the prevalence of newborn hearing loss at 1.7 per 1,000 infants screened (CDC, 2018b). These data are reported to the CDC from each state EHDI program as collected from birthing hospitals. These data are collected from hospitals outside the medical record through reporting systems that are distinct from hospital records and billing. From an epidemiological standpoint, it is valuable to have data on the incidence and prevalence of infant hearing loss on a national level; however, these data are detached from the medical record of infants, which may limit progress in large scale research regarding other medical or sociodemographic factors associated with abnormal NBHS and infant hearing loss, when those factors are not captured in the EHDI program. With current EHDI data, it is impossible to investigate for links between medical conditions such as NAS and infant hearing loss, thus, other research tools and databases must be used. Unlike hospital EHDI data, there is no mandate or requirement of reporting abnormal NBHS results or hearing loss diagnosis in administrative records and it is possible that diagnoses related to abnormal NBHS and infant hearing loss may be underreported or may go unreported.

Table 3

Logistic Regression Analysis: Likelihood an Infant Failed Their Hearing Screen or was Given a Diagnosis of Hearing Loss Based on Patient-Specific Factors

Logistic Regression Analysis, Likelihood of Abnormal Hearing Assessment			
		Abnormal Auditory Function	**Hearing Loss Diagnosis**
		Odds Ratio (95% CI, *p*)	**Odds Ratio (95% CI, *p*)**
	NAS	0.76 (0.58–0.98, < 0.05)	2.17 (1.23–3.85, < 0.01)
	Risk†	0.69 (0.58–0.82, < 0.001)	3.02 (1.85–4.95, < 0.001)
	Female	0.84 (0.79–0.90, < 0.001)	0.84 (0.73–0.96, 0.01)
Race	White	1.00	1.00
	Black	1.48 (1.30–1.69, < 0.001)	0.77 (0.48–1.24, 0.29)
	Hispanic	1.18 (0.99–1.41, 0.06)	1.14 (0.73–1.79, 0.56)
	Asian or Pacific Islander	0.90 (0.74–1.10, 0.31)	0.98 (0.70–1.39, 0.92)
	Native American	1.83 (1.19–2.81, < 0.01)	1.94 (0.93–4.03, 0.08)
	Other	1.19 (0.94–1.49, 0.138)	0.68 (0.36–1.26, 0.22)

Note. NAS = neonatal abstinence syndrome. N = 951,437
Risk = Presence of known medical risk factor for hearing loss.

Table 4

Logistic Regression Analysis: Likelihood an Infant Failed Their Hearing Screen or was Given a Diagnosis of Hearing Loss Based on Socioeconomic Characteristics

Logistic Regression Analysis, Likelihood of Abnormal Hearing Assessment			
		Abnormal Auditory Function	**Hearing Loss Diagnosis**
		Odds Ratio (95% CI, *p*)	**Odds Ratio (95% CI, *p*)**
Payer Type	Private	1.00	1.00
	Medicaid	1.13 (1.02–1.25, < 0.05)	1.45 (1.19–1.77, < 0.001)
	Self-Pay	1.10 (0.91–1.33, 0.34)	1.21 (0.80–1.81, 0.36)
	No Charge	2.24 (0.72–6.85, 0.16)	n/a‡
	Other	0.87 (0.66–1.16, 0.35)	1.41 (0.66–3.01, 0.37)
Patient Geography†	Central metro	1.00	1.00
	Fringe metro	1.10 (0.89–1.35, 0.37)	1.59 (0.87–2.93, 0.14)
	Metro of 250,000–999,999	1.33 (1.03–1.71, < 0.05)	0.58 (0.32–1.07, 0.08)
	Metro of 50,000–249,999	1.39 (1.04–1.86, < 0.05)	0.93 (0.40–2.15, 0.87)
	Micropolitan	1.44 (1.08–1.93, < 0.05)	0.79 (0.40–1.58, 0.51)
	Not metro- or micropolitan	1.26 (0.95–1.68, 0.11)	1.53 (0.59–4.0, 0.385)
Income Quartile	1st (< $25,000)	1.00	1.00
	2nd ($25,000–$34,999)	1.0 (0.88–1.13, 0.95)	0.83 (0.56–1.25, 0.38)
	3rd ($35,000–$44,999)	1.03 (0.90–1.18, 0.68)	0.96 (0.72–1.29, 0.0.80)
	4th ($45,000 and above)	0.89 (0.75–1.04, 0.15)	0.53 (0.34–0.81, < 0.01)

Note. N = 951,437. †Patient Geography: Central metro = county population > 1 million; Fringe metro = co. pop. > 1 million; Mid-metro = co. pop. 250,000–999,999; Small-metro = co. pop. 50,000–249,999; Micropolitan = co. pop. 10,000–49,999; Not metro- or micropolitan = co. pop. < 10,000.
‡Excluded from analysis due to small sample size and perfect failure prediction.

Our data are discordant with CDC findings, yielding a lower overall rate of failed hearing screen or hearing loss diagnosis of 0.71% in this inpatient sample of birth hospitalizations. This study found, using uni- and multivariate analysis, that infants with NAS have a lower odds ratio of reported abnormal NBHS results on inpatient discharge records than non-NAS infants. These findings could be due, simply, to an actually lower incidence of abnormal NBHS in NAS infants. There is no other evidence that would suggest that neonatal substance exposure is protective against hearing loss.

Conversely, we hypothesize that abnormal NBHS is underreported in the inpatient records and hospital billing of NAS infants which could account for the lower odds ratio found in this data. The complexity of medical care and multi-disciplinary discharge follow-up of NAS infants could influence the reporting of abnormal NBHS on inpatient hospital records. If proven true, this hypothesis is significant as it indicates that complex medical conditions in infants, such as NAS, could negatively influence the reporting of NBHS results. This could lead to delays in the diagnosis and treatment of hearing loss within the local medical community if NAS is indeed a risk factor for hearing loss. This hypothesis is further supported by this data which found a significantly lower odds ratio of documented abnormal NBHS in infants with known risk factors for hearing loss. These factors include complex medical conditions such as perinatal maternal infections, craniofacial abnormalities, ototoxic drug exposure, NICU admission, prematurity, and hyperbilirubinemia. These complex medical conditions along with other conditions in the infant would be prioritized in the inpatient and early outpatient care of infants which could influence the reporting of NBHS results, along with other relevant clinical findings. This study also finds a difference in the actual diagnosis of hearing loss of NAS-infants and non-NAS infants based on inpatient data. Although the overwhelming majority of infants who are diagnosed with hearing loss receive that diagnosis after multiple audiological evaluations on an outpatient basis, this study suggests that infants with NAS and congenital hearing loss may receive definitive audiological evaluation in addition to NBHS due to their prolonged inpatient stays in the hospital. Although this study does not test for the causal relationship between NAS and hearing loss diagnosis, it is the first to identify a relationship between the two in an infant population.

Although few cases have been reported in the adult literature of hearing loss from opioid use, there is not strong evidence to suggest ototoxicity with *in-utero* opiate exposure. Our findings, based on birth hospitalization data, found there was no significant difference in hearing loss incidence between the exposed and unexposed cohorts. More research is needed to assess the relationship between NAS and infant hearing loss as the complex care and increased length of stay required by these patients can make identification of hearing loss a difficult task. Subsequent work should be completed to follow these patients into childhood to ensure longevity of hearing health or recognize later needs, as well as improve detection of delayed onset or progressive hearing loss not observable in the birth admission. It is also important to stress the need for thorough discharge planning for these patients and confirmation of audiologic follow-up in the event of a failed NBHS given their risk of poor use of prophylactic and specialty healthcare services (Fang et al., 2015; Gill et al., 2007; Kivisto et al., 2014).

In this study, we also assessed patient and socioeconomic factors associated with documented abnormal NBHS/infant hearing loss. In doing so, increased rates of failed hearing screens and hearing loss diagnoses were noted in vulnerable patient populations. Medicaid insurance status had higher odds of abnormal hearing assessments and diagnoses compared to patients with private insurance. Place of residence was also associated with differences in hearing assessment. Patients from outside a central metropolitan area were at increased odds of failed NBHS. The greatest likelihood was seen in micropolitan (county population 10,000-49,999), OR = 1.44. Compared to the lowest earning families, patients whose family incomes were in the 4th quartile had decreased odds ratios of HL, OR = 0.53 ($p < 0.01$). By using data from the National Health Interview Survey, Boss et al., 2011 also described increased rates of hearing loss in children of lower socioeconomic status. Increased rates of failed NBHS or HL diagnoses from national inpatient data in children covered by Medicaid, and in those from smaller communities are novel findings not yet reported in the literature. The possibility of failed NBHS or HL diagnoses is concerning given these are populations already at-risk for worse audiologic follow-up or decreased access to care following a failed NBHS. Prior studies with small samples have shown that loss to follow up and decreased care access are common for children from rural areas or outside a central metropolitan area, who are uninsured or covered by public insurance, and come from families with lower incomes and lower parental education levels (Bush et al., 2014; Liu et al., 2008; Oghalai et al., 2002; Ravi et al., 2016; Zeitlin et al., 2017). Individuals who identified as Black, OR =1.48 ($p < 0.001$) or Native American, OR = 1.24 ($p = 0.02$), were more likely to have abnormal hearing assessments. Unfortunately, racial and ethnic minorities have been noted to be at higher risk for loss to follow-up after a failed NBHS (CDC, 2018a; Liu et al., 2008; Zeitlin et al., 2017). This again highlights patient populations not only at risk for increased rates of hearing loss but also worse use of subsequent care.

This study is limited most notably by its retrospective nature and reliance on administrative data, which may not document all clinically-relevant information. Although 98% of newborns received hearing screening in 2015 (CDC, 2015), it must be noted that differing techniques of screening and reporting mechanisms are used throughout the country. As EHDI and KID data rely on reporting from national samples, testing and diagnostic homogeneity cannot be assumed for this study. Likewise, NAS is a clinical diagnosis made based on a constellation of signs and symptoms, and there is no uniform evaluation

mechanism to make this diagnosis (McQueen & Oikonen, 2016). Finally, our study uses imprecise measures of the outcome of an abnormal NBHS. We rely on ICD-10 codes for "abnormal auditory function" that may not be consistently coded in billing programs when an infant fails their screen.

Conclusion

NAS children have a lower rate of inpatient documented failed NBHS and a higher odds of HL diagnosis during the birth admission. The complex medical care of these infants could complicate NBHS and subsequent follow-up. Certain sociodemographic factors including some racial and ethnic minorities, lower income level, residence outside a metropolitan center, and Medicaid insurance are associated with higher risk of hearing loss. Further research is needed to assess hearing screening and diagnoses of hearing loss in vulnerable populations such as NAS infants.

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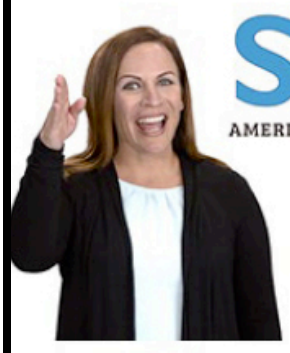
EHDInfo



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Audiologic Clinical Practice Patterns: Infant Assessment

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Abstract
The purpose of the present study was to report the current clinical practice patterns for assessment of infants after a referred newborn hearing screening within the context of available guidelines and to examine how the advent of newer stimuli, technology, and/or instrumentation has changed clinical practice patterns for audiologic infant assessment. A mixed-method survey that included both quantitative and qualitative questions was disseminated to pediatric audiologists in 2017. Quantitative data were analyzed via descriptive statistics while qualitative questions were analyzed via content analysis and combined with associated quantitative data. Lastly, infant assessment test battery categorization was completed to ascertain the extent to which providers were using recommended protocols. Results revealed appreciable variability in the test batteries employed by facilities evaluating infants. Additionally, a sizable portion of facilities are not using test batteries recommended by sources of guidance for evidence-based practice, suggesting a possible need for adopting a standardized protocol in the United States. Factors that potentially contribute to these results are reviewed as well as proposed next steps toward improving adherence to recommended guidelines.

Acronyms: ABR = automated brainstem response; ANSD = auditory neuropathy spectrum disorder; ASSR = auditory steady state response testing; DPOAE = distortion product otoacoustic emissions; EHDI = Early Hearing Detection and Intervention; JCIH = Joint Committee on Infant Hearing; OAE = otoacoustic emissions

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Over the past decade, the rate of infants screened for hearing loss at birth, receiving diagnostic testing, and enrolled in EI services have all increased significantly (Subbiah, Mason, Gaffney, & Grosse, 2018). Although the screening rate quickly approached ceiling levels shortly after newborn hearing screening became universal in most states in 2005, successful completion of diagnostic testing and enrollment in early intervention services for children with confirmed hearing loss continues to lag behind (Grosse et al., 2017). One factor that might contribute to differences in follow-up rates across early hearing detection and identification (EHDI) programs is variability in how programs are executed across the United States. For screenings, each individual state mandates when testing occurs (solely as inpatient or allowing an outpatient screening) and the type of testing that occurs, which typically depends upon risk factors for hearing loss. Decisions for screening protocols are often based on recommendations from the Joint Committee on Infant Hearing, which allows for some variability in screening depending upon certain factors (JCIH, 2007). Despite the variability in how screening occurs from both a logistical and testing paradigm perspective, state EHDI systems have successfully achieved a high rate of screening prior to one month of age, with national data increasing from 85.1% in 2006 to 98.6% in 2016 (Subbiah et al., 2018). A potential reason for these success rates may be that defined screening procedures and protocols merely exist.

However, the high level of success seen at the screening step of EHDI programs has not translated to the diagnostic step of the process. Within the same time period, the percentage of infants receiving diagnostic assessment prior to three months of age increased from 19.8% in 2006 to 36.6% in 2016 (Subbiah et al., 2018). Although the overall percentage of infants receiving diagnostic assessment in general reached a high of 56.6% in 2016, state EHDI programs continue to struggle with executing the diagnostic step of the EHDI process. Reasons for delays between initial diagnostic testing and confirmation of hearing loss have included a need for multiple tests to confirm hearing status, recurrent middle ear issues, and near-normal hearing at initial testing or fluctuant hearing loss noted on serial tests (Fitzpatrick, dos Santos, Grandpierre, & Whittingham, 2017; Holte et al., 2012). Parents who have gone through the EHDI system have reported that multiple tests were needed for confirmation

of hearing loss and 29% of families reported a need to go to multiple locations for a complete testing (Larsen, Muñoz, DesGeorges, Nelson, & Kennedy, 2012). The need for multiple tests to confirm hearing status has been attributed to additional multiple factors, including inadequate sleep state limiting the number of threshold measures obtained, noisy test results precluding conclusive results, and the presence of chronic middle ear fluid (Muñoz, Nelson, Goldgewicht, & Odell, 2011).

An additional explanation for the need for multiple tests may be the lack of a defined expectation of diagnostic centers in terms of testing protocols or adoption of an expected protocol. Although some states have defined diagnostic protocols for infant assessment, many do not, and of those who have recommended protocols available to review there is significant variability in the level of detail provided to guide clinicians (Hunter, Steuerwald, Hounam, & Kothari, 2016). In contrast, diagnostic programs outside of the United States often have published protocols to define necessary testing procedures for diagnosing hearing loss in infancy at either the national or province-level (Hatton, Hyde, & Stapells, 2012; Hyde et al., 2016; Sutton et al., 2013). Although some guidance has been offered in the United States through governing body guideline statements (American Academy of Audiology [AAA], 2012; JCIH, 2007; JCIH, 2019) and by practitioners providing guidance articles (Smith & Wolfe, 2014), there continues to be no specific protocols mandated by a majority of EHDI programs.

The limited adoption of recommended, evidence-based protocols across the United States has led to significant variability in the provision of services. Munoz et al. (2011) systematically studied clinical practice patterns for infant assessment through a national survey. Findings of this survey revealed that only 9.4% of respondents were using an infant assessment battery consistent with JCIH (2007) recommendations, with the remaining 90.6% of facilities reporting assessment batteries of varying thoroughness (Muñoz et al., 2011). At that time, 16.9% of respondents reported using no frequency-specific electrophysiologic measures of hearing (i.e., automated brainstem response [ABR] using tone burst stimuli), which is considered to be essential given that the fitting of amplification for those children who are diagnosed with permanent hearing loss will be the next step in the process. Consequently, evaluations completed after a newborn hearing screening referral appear to vary considerably across facilities and states in general, which may significantly impact the national EHDI program effort to diagnose hearing loss in infants by three months of age.

An update to the JCIH statement was just released and continues to provide guidelines for diagnostic testing of infants and young children along with substantial evidence to support those guidelines (JCIH, 2019). Although this updated statement does not outline which diagnostic tests should take place within specific age ranges in the same manner as previous iterations, the statement outlines the key aspects of audiologic assessment for infants and young children as including the following: (a) auditory brainstem response testing to estimate ear- and frequency- specific thresholds to define type, degree, and configuration of hearing level, (b) tympanometry or wideband reflectance to assess middle ear function, (c) acoustic reflexes to evaluate middle ear and auditory brainstem pathway integrity, (d) otoacoustic emissions (OAE) to evaluate the integrity of the outer hair cell function of the cochlear, and lastly, (e) behavioral evaluation via visual response audiometry or conditioned play audiometry as soon as developmentally appropriate.

The purpose of the present study was to report the current clinical practice patterns for assessment of infants after a referred newborn hearing screening within the context of available guidelines. Additionally, we sought to examine whether the advent of newer stimuli, technology, and/or instrumentation has changed clinical practice patterns for audiologic infant assessment.

Method

This survey study was deemed exempt from review by the Nationwide Children's Hospital Institutional Review Board. The study was designed as a mixed-model survey that included both quantitative and qualitative questions collected electronically through REDCap (Harris et al., 2009). Survey development was modeled after a previously published clinical practice survey (Muñoz et al., 2011) after obtaining permission from the lead author (personal communication). Survey questions included information regarding tests completed as a part of assessment of both infants and young children, as well as testing conditions and logistics of scheduling wherever applicable. Survey questions were updated to provide choices that included modern assessment stimuli (chirp) and testing paradigms (auditory steady state response testing; ASSR) for the electrophysiologic questions. This paper will describe the infant assessment data only, focusing on diagnosis of hearing loss in children birth to six months of age. Once survey formulation was completed by the study team, questions were piloted with ten clinical audiologists currently engaged in assessment of infants and young children to evaluate whether questions were straight forward and answerable. The final survey is available for review in the Appendix.

Survey dissemination was completed over a two month time period from October to November 2017. Surveys were disseminated by direct email to 345 pediatric audiologists known to be currently providing care for infants and young children, social media posts on specialized pediatric audiology groups, and through communication via two EHDI program coordinators who were willing to provide the survey link to audiologists in their diagnostic networks. One EHDI coordinator also offered to post the survey announcement on an EHDI coordinator listserv for the United States to encourage other coordinators to disseminate the survey. During the course of the survey period, audiologists who were directly emailed were invited to participate in the survey twice (10/17/2017 and 11/1/2017) to facilitate completion of the

survey. The survey announcement was also posted twice during this time period on social media outlets (10/18/2017 and 11/1/2017). Because of the use of social media and listservs for dissemination, the total number of audiologists the survey reached cannot be calculated.

A total of 272 surveys were submitted during the data collection period; 187 (68.8%) were completed in full. Respondents reported practicing in 39 states and Washington, D.C. Most respondents reported they were female (n = 173, 92.0%) practicing in a hospital setting (n = 101, 54.1%). Other settings represented in the dataset included: private-practice (n = 17, 9.0%), college/university clinic (n = 13, 7.0%), ENT office (n = 18, 9.6%), school (n = 19, 102%), and other (n = 19, 10.2%). Most of the respondents reported having an AuD degree (n = 146, 78.6%) while 20 (10.6%) reported having a Master's degree, 17 (9.1%) reported having a PhD, one (0.5%) reported having ScD degree, and three (1.6%) declined to respond to this question. Most of the respondents reported having between one and five years (n = 64, 34.8%) or over 20 years (n = 37, 20.1%) of clinical experience. Respondents were also asked to report how many years of clinical experience they have specifically evaluating infants and children. Of the 187 respondents who provided this information, 27 (14%) reported that they had not spent their entire clinical career seeing pediatric patients, and all but five reported at least 1–5 years of experience evaluating children. The remaining five (2.6%) respondents did not choose to report their years of clinical experience with pediatric patients.

Once the survey period ended, all variables were exported into Microsoft Excel files for analysis. Quantitative questions were analyzed through descriptive statistics using IBM SPSS Statistics, Version 24 (IBM Corp; Armonk, NY). Qualitative responses, predominantly in the form of free-field comments throughout the survey, were individually analyzed using content analysis (Hsieh & Shannon, 2005; Krippendorff, 1980) to derive themes that could supplement the quantitative results. Quantitative and qualitative results were then merged for each section of the survey. Percentages were calculated for each diagnostic test reportedly performed by respondents completing the infant assessment portion of the survey. Test batteries that were reported for assessment of infants between birth to six months of age were classified as either meeting or not meeting the JCIH (2007) guidelines, which outlines the following tests should be completed in infants ages birth to six months: (a) Child and family history; (b) frequency-specific assessment of the ABR using air-conduction and bone-conduction tone bursts; (c) Click-evoked ABR testing using both condensation and rarefaction single-polarity stimulus, if there are risk factors for neural hearing loss or if there is no response on tone burst ABR; (d) distortion product otoacoustic emissions (DPOAEs); and (e) Tympanometry using 1000-Hz probe tone. Because of the advent of additional frequency-specific testing stimuli and procedures since the publication of the JCIH (2007) guidelines, respondents who reported doing frequency-specific chirp ABR or ASSR testing were included as being adherent to the guidelines. Additionally, data were analyzed in light of the newly released JCIH (2019) statement which adds acoustic reflex testing as a key part of a diagnostic test battery in infants and children.

Lastly, a logistic regression was completed to evaluate the effects of geographical location, years of clinical experience, and appointment length allowed for completing a natural sleep ABR on the likelihood that providers are adherent to recommended guidelines for diagnostic assessment in infants. These specific factors were chosen for analysis due to their potential impact on whether a provider would follow recommended guidelines. For instance, depending upon the state in which the respondent is located and the presence of their specific EHDI program, some respondents may have more support or higher visibility of JCIH guidelines than others. For this analysis, due to variance in the number of respondents from individual states, location was collapsed from state-level to regional-level, including Northeast (n = 22), South (n = 34), Midwest (n = 55), and West (n = 8) regions consistent with the United States Census Bureau Regions and Divisions (U.S. Census Bureau, Geography Division, 2000). For the purposes of categorization, one respondent from Hawaii was included in the West region. Eight respondents declined to report their location and had to be excluded from the analysis. Years of clinical experience may impact the confidence of providers executing different aspects of a test battery or alternatively may impact which tests are completed depending upon provider bias for specific tests. Lastly, appointment length may impact a provider's decision process for which aspects of a test battery should be completed given the allotted time. Analysis was completed with adherence to the JCIH (2007) guidelines (categorical yes/no) as the dependent variable with two-sided p-values < 0.05 considered significant.

Results

A total of 162 survey respondents recorded which tests they typically complete as a part of a test battery assessing infants birth to six months of age. Table 1 provides the number and percentage of respondents reporting they complete each test. Overall, a vast majority of respondents are performing a case history (100%), 1000 Hz tympanometry (93.8%), DPOAES (94.4%), frequency specific ABR (74.0%), and click ABR (85.19%). Alternative frequency-specific electrophysiologic testing was also reported by some respondents: chirp ABR (8%), tone burst ASSR (14.2%), or chirp ASSR (4.3%). Overall, these data suggest that there is variability among clinicians in what they include in a test battery to assess hearing for infants after a referred newborn hearing screening.

Responses were further categorized into whether the test battery meets or does not meet JCIH (2007) guidelines. Results showed that 88 (54%) were adherent to the JCIH (2007) recommendations. Among the 74 respondents who were not meeting recommendations, a variety of tests were omitted: 36 (48.6%) omitted bone conduction

Table 1
Number and Percent of Respondents who Perform Each Test Measure as a Part of their Infant Diagnostic Test Battery

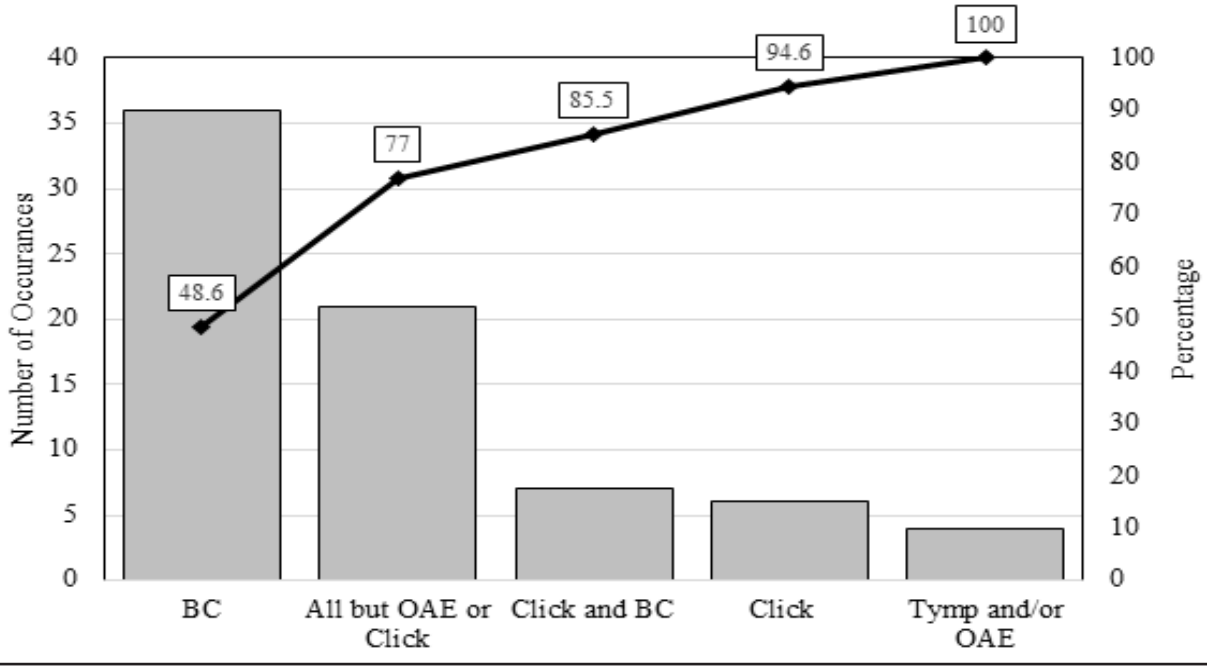
Test Measure	Number	Percent
Otoscopy	145	89.51
Case History	162	100
1000 Hz Tympanometry	152	93.83
226 Hz Tympanometry	25	15.43
Acoustic Reflex Testing	40	24.69
DPOAEs	153	94.44
TEOAEs	19	11.73
Click ABR	138	85.19
Tone Burst ABR	120	74.07
Chirp ABR	13	8.02
Bone Conduction ABR	98	60.49
Chirp ASSR	7	4.32
Tone Burst ASSR	23	14.20

Note. DPOAEs = distortion product otoacoustic emissions; TEOAEs = transient evoked otoacoustic emissions; ABR = auditory brainstem response; ASSR = auditory steady state response.

testing, 21 (28.4%) omitted all but OAE and Click testing, 7 (9.5%) omitted click and bone conduction testing, 6 (8.1%) omitted click testing, and 4 (5.4%) omitted tympanometry and/or OAE testing (Figure 1). Of note, 21 (12.9%) of respondents reported using no frequency-specific electrophysiologic testing in their test battery. The recent publication of the 2019 JCIH statement additionally includes acoustic reflex testing as a key aspect of pediatric assessment and provides evidence to support its use in infants. It should be noted that based on the results of this survey, over 75% of respondents would be non-adherent to the updated guidelines based on excluding acoustic reflex testing from their test battery alone.

Respondents were asked whether their individual state provides a protocol or guidance for the assessment diagnostic test battery. Of the 162 respondents, 111 (68.5%) reported that their state does provide either a protocol or guidance. Qualitative responses revealed significant variability in the types of guidance offered, including anything from recommending that both ears are tested as the only recommendation to referring providers to national organization best practice statements for guidance on test battery formulation. Additionally, multiple respondents commented that although a guidance statement from their state EHDI program exists, the recommendations are dated and in need of updating due to not being consistent with current best practice statements. The logistic regression to evaluate the potential effects of region, years of clinical experience, and appointment length on the likelihood that a provider is adherent to recommended guidelines was not significant (X^2 (10) = 5.353, p = 0.866).

Figure 1. *Pareto chart of omitted test battery items leading to a determination of non-adherence to the JCIH (2007) recommended guidelines for assessment of infants birth to six months of age.*



Note. BC = bone conduction; OAE = otoacoustic emissions; Tymp = tympanometry.

Test Conditions

In addition to respondents reporting which tests they performed as a part of their test battery, respondents were also asked a number of questions regarding test conditions or logistics. Parents were provided instructions for the test at 98.7% of facilities, but instructions varied and sometimes multiple channels were used. Respondents reported providing verbal instructions on the phone at the time of appointment scheduling (n = 123; 76.4%) and on the phone at the time of appointment confirmation (n = 72; 44.7%), or via a letter prior to the appointment (n = 110, 69.3%). Instructions included a number of different strategies to maximize sleep state (Table 2), with most respondents reporting they instruct families to bring the infant sleep deprived (n = 153, 95.6%) and hungry (n = 150, 93.8%).

Table 2

Number and Percent of Respondents Providing Specific Instructions to Parents for Preparation of Infant Natural Sleep Electrophysiologic Testing

Parental Instructions Provided	*N* (%)
Bring infant sleep deprived	153 (96.6)
Bring infant hungry	150 (93.8)
Bring items that comfort the infants (bottle, blanket, pacifier, etc.)	132 (82.5)
Bring an additional adult if planning on bringing additional children (older siblings) to the appointment	105 (65.6)
Bring an additional adult to help keep the infant awake during the car ride	95 (59.4)
Bring the car seat for them to sleep in for testing	45 (28.1)
Do not put lotion on the infant's face	40 (25.0)
Our facility provides no instructions prior to the appointment	2 (1.3)

A variety of appointment lengths were reported by respondents for performing a diagnostic ABR in natural sleep. Of the respondents who provided a response to this question (n = 161), 12 (7.4%) reported having a 60-minute appointment length, 28 (17.4%) reported 90 minutes, 93 (57.8%) reported 120 minutes, and the remaining 28 (17.4%) reported having 180–240 minutes to complete the test battery. Many respondents qualitatively added that this appointment length includes the time it takes for the infant to fall asleep for testing.

For test administration, a variety of starting points were reported for electrophysiologic measures, with most respondents reporting they start with click stimuli (n = 94, 62.3%) while others reported a variety of tone burst ABR or ASSR stimuli (Table 3). Comments included for this question indicated that some respondents start with a click to rule out auditory neuropathy spectrum disorder (ANSD) at the onset of the evaluation depending upon birth history or if the ABR was being conducted as a sedated

Table 3

Number and Percent of Respondents Reporting the Initial Stimulus for Electrophysiologic Testing of Infants

Stimulus	*N* (%)
Click ABR	94 (62.3)
2000 Hz tone burst ABR	36 (23.8)
4000 Hz tone burst ABR	11 (7.3)
1000 Hz tone burst ABR	4 (2.6)
2000 Hz chirp ABR	3 (1.9)
500 Hz chirp ABR	1 (0.7)
4000 Hz chirp ABR	1 (0.7)
Tone burst ASSR	1 (0.7)

Note. ABR = auditory brainstem response; ASSR = auditory steady state response

procedure, while using a 2K Hz stimulus for their starting point for non-sedated ABRs. Most respondents (n = 156, 98.7%) reported routinely using insert ear phones for their transducer versus standard/supra-aural TDH headphones (n = 2, 1.3%). Narrative comments included caveats for using supra-aural only for infants presenting with aural atresia/microtia. All respondents reported testing both ears regardless of screening results. In the case of unilateral referrals, 82.9% of respondents start testing in the ear that referred while 17.1% start testing in the ear that passed the newborn hearing screening.

Respondents were asked to report the top three factors that presented the most common challenges for completing a diagnostic evaluation in one appointment session (Table 4). The most common challenges were reported to be as follows: patient sleep state (n = 157, 98.7%), electrical noise interference during testing (n = 67, 42.1%), and equipment issues (n = 61, 38.4%). Narrative comments for this question included that it is rare to not complete testing within the allotted time (n = 5), the primary issue is the infant sleep state (n = 5), and additional factors were offered, including late arrival for the appointment (n = 5), neurologic issues leading to poor replicability (n = 1), a high no-show rate (n = 1) and parents not following directions for optimal testing (n = 1).

Table 4

Factors Related to an Inability to Complete a Diagnostic Evaluation Within One Appointment Session

Factors for Incomplete Tests	*N* (%)
Patient sleep state/waking up	157 (98.7)
Electrical noise interference	67 (42.1)
Equipment issues	61 (38.4)
Appointment time too short	44 (27.7)
Parent request to discontinue testing	27 (17.0)

Note. Respondents were requested to report the top three reasons

Discussion

The purpose of this clinical practice survey was to report the current clinical practice patterns for assessment of infants after a referred newborn hearing screening within the context of available guidelines. Results indicate that more clinicians report completing an infant test battery consistent with JCIH (2007) recommendations than previously reported on similar surveys conducted in a similar cohort of audiologists who complete assessments for infants who refer the newborn hearing screening (Muñoz et al., 2011). This is promising as EHDI programs across the United States strive to improve outcomes for children with congenital hearing loss by implementing interventions to increase adoption of recommended diagnostic follow-up and decrease loss-to-follow-up in this population. Despite the increase in evidence-based practice, significant variability in testing batteries and practices remain. Although there will always be patient-specific factors that exist which necessitate some flexibility in practice, having a consistent approach to diagnosis across test centers will reduce variability and increase equity of care for infants who refer on the Universal Newborn Hearing Screening. This survey indicates that there are several areas of commonality within assessment approach but also several areas of variability which may require further consideration for a unified approach across test centers.

Most respondents (98.7%) reported that they provide parental instructions for testing prior to the test day to optimize testing conditions. This is consistent with the previous data suggesting that clinicians recommend a variety of instructions to have parents prepare infants for optimal testing (Muñoz et al., 2011). Additionally, all respondents reported that they evaluate both ears during a diagnostic appointment regardless of the screening results (i.e., bilateral refer vs. unilateral refer). This finding is a positive practice considering hearing status might change in the time between screening and diagnostic testing and that human error could contribute to reporting results of ears erroneously. Both of these factors were mentioned by respondents in the narrative comments provided as a rationale for always testing both ears.

Despite improvements in evidence-based practice engagement, almost half of the respondents have not adopted recommended test batteries, and 12.9% of respondents report they do not use any frequency-specific electrophysiologic testing for their diagnostic assessments. Although the survey instructions were specific to diagnostic testing of infants birth to six months of age after a referred newborn hearing screening, results showed a large number of facilities engaging in re-screening approaches when perhaps a diagnostic evaluation was indicated. It is unclear as to whether these particular responses came from facilities within states that allow re-screening as an outpatient, or whether clinicians engage in re-screening despite state guidelines mandating a diagnostic after a pre-determined number of referred screens regardless of whether screenings were completed inpatient or in a hybrid approach of one inpatient and one outpatient screening. Regardless of the source, results suggest a fair amount of re-screening in this population which may suggest a need for standardization in the definition of *diagnostic* assessment of hearing loss in infants. Although JCIH (2007), JCIH (2019), and the AAA Audiologic Guidelines for Assessment of Infants and Young Children (2012) Clinical Practice Guideline all state that there is a need for both a test battery approach and the use of frequency-specific electrophysiologic measures to infant assessment, it does appears that a number of clinicians who assess infants do not heed these recommendations. This is troubling given that another finding of this study was that emerging stimuli (chirp) and assessment methods (ASSR) are being employed by clinicians which would presumably give providers more flexibility in how they assess infants. Specifically, these newer testing approaches have been found to reduce test time due to elicitation of larger responses and concurrent measurement of multiple frequencies (Ferm, Lightfoot, & Stevens, 2013; Rodrigues, Ramos, & Lewis, 2013; Sininger, Hunter, Hayes, Roush, & Uhler, 2018).

Additionally, survey results revealed that clinicians are often starting their assessment using click stimuli despite the main objective of the assessment being to establish frequency-specific hearing sensitivity to evaluate whether intervention via amplification is necessary. Both JCIH (2007) and JCIH (2019) advocate for the prioritization of frequency-specific ABR assessment to establish frequency-specific hearing levels to guide fitting of amplification. Although assessment for neural integrity is important, especially for children with risk factors associated with possible neural involvement, less than 1% of the greater population will have findings of ANSD and only between 5 and 13% of children with permanent hearing loss will have results consistent with ANSD (Berlin et al., 2010; Vignesh, Jaya, & Muraleedharan, 2016; Rance, 2005; Sanyelbhaa, Kabel, Sammy, & Elbadry, 2009). Consequently, the assessment of neural integrity in cases in which there is a concern for ANSD is recommended by JCIH *after* risk factors and/or a no-response ABR has been established. Results of this clinical practice survey suggest that a majority of clinicians are not following clinical guidelines specific to which test among an infant test battery should be prioritized.

A lack of adherence to evidence-based practice is not a novel finding in our field. Other clinical practice surveys have indicated that clinicians are not following evidence-based practice guidelines specifically for the provision and management of amplification in children (Moodie et al., 2016). The current study continues to indicate that there is a significant need for improving adherence to recommended guidelines for evidence-based practice in the United States to ensure infants and young children are provided the hearing healthcare they need to optimize their outcomes in the presence of congenital hearing loss. To that end, there has been a recent push for more standardization at the state level (Hunter et al., 2018; Silver, 2019) and at the national level with continued revision of guidelines from national associations and

the formulation of the Audiology Standards Practice Organization. Although multiple factors can contribute to loss-to-follow-up after a referred newborn hearing screening, having a unified approach to assessment in infants can at the very least aid in increasing diagnostic follow-up. In countries where standards are set, follow-up for newborn hearing screening is considerably higher. Wood, Sutton, and Davis (2015) reported the advances made by the newborn hearing screening program in the United Kingdom between 2006 and 2013. Results showed that follow-up rates reached 82.5% for follow-up testing by 4 weeks of age and 95% follow-up testing prior to six months of age for the cohort of children born in late 2013 (Wood et al., 2015). Loss to follow-up rates are also lower in U.S. states that have established clinical protocols and/or state approval for diagnostic centers capable of providing infant assessment via ABR. California, Florida, Kansas, Massachusetts, Rhode Island, Vermont, Wisconsin, and Wyoming all have loss-to-follow-up rates less than 10% as of 2016 and have either a detailed state protocol or a system for state approval to be a diagnostic center specifically for ABR assessment (Centers for Disease Control and Prevention, 2018). Although there are many interventions that could be instituted to improve follow-up rates in the United States, until adoption of a unified approach to assessment in infants can be established it is unlikely that diagnostic follow-up rates after referral on newborn hearing screening will improve to meet peer-nation standards.

Although the data presented here reflect what pediatric audiologists reported as their diagnostic test battery for infants, one limitation of this study is the relatively small number of respondents which may not be reflective of the entire field. An attempt was made to evaluate whether specific factors affect the likelihood of a provider engaging in evidence-based practice as recommended by JCIH (2007) through logistic regression modeling; however, that analysis was not significant. It cannot be ruled out that this analysis was impacted by the small number of respondents or the variability in demographics and circumstances under which audiologists reportedly execute diagnostic testing. Additionally, direct comparisons with previous studies cannot be made due to potential differences in sample. In future studies, additional efforts should be made to ensure more consistent sampling across the United States through a structured, prospective, longitudinal study that would allow for direct comparison and evaluation of change across time.

Conclusion

Although engagement in evidence-based practice for infant hearing assessment has increased over the past several years, variability in testing protocols still exists. Facilitating the adoption of test batteries consistent with recommended national guidelines, especially if it is facilitated at the state-level in a similar fashion to screening procedures, may reduce this variability and serve to increase diagnostic rates after referral on the newborn hearing screening.

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Appendix

Audiology Infant Assessment Clinical Practice Survey

The Audiology Department at Nationwide Children's Hospital is conducting a survey of common clinical practices for infant assessment in the United States. The purpose of this survey is to explore how children are evaluated via electrophysiological and behavioral testing within the first 36 months of life.

This survey will take approximately 5-10 minutes to complete. Survey responses are anonymous and cannot be traced to individuals. This information will provide our field with important insight as to how we are providing services to this population. This study has been approved by the NCH Institutional Review Board (IRB 017- 00859).

For additional information about this survey, please feel free to contact the Principal Investigator, Dr. Ursula Findlen, for a Research Summary at ursula.findlen@nationwidechildrens.org.

Thank you for your consideration and time in completing this survey.

General Questions

Do you or does your facility provide assessment services to infants via electrophysiological (i.e.: ABR, ASSR, etc.) Testing?	◯ Yes ◯ No
Do you or does your facility provide assessment services for infants and young children via Visual Reinforcement Audiometry (VRA)?	◯ Yes ◯ No

State Early Hearing Detection and Intervention (EHDI) Programs

From the following choices, choose the response that best describes how much control that you feel you have/had on the development of your practice's protocol for testing infants and young children:

- ◯ I have a lot of control over the protocol.
- ◯ I can influence the protocol but ultimately the decision is out of my hands.
- ◯ I have little/no influence on the protocol that is used in this practice.

Comment: _____

Does your state Early Hearing Detection and Intervention (EHDI) program provide protocol recommendations for the following ages?

- ☐ For Testing children 0-6 months old
- ☐ For Testing children 6-12 months old
- ☐ For Testing children 12+ months old
- ☐ No recommendations are provided
- ☐ Unsure

Comment: _____

If your state EHDI program provides a recommended protocol, does your practice's clinical protocol reflect the state recommended protocol?

- ◯ Yes
- ◯ No
- ◯ Unsure
- ◯ Not applicable

If your state EHDI program provides a recommended protocol, choose the response that best describes how much control that you feel you have/had on the development of that protocol:

- ◯ I have a lot of control over the protocol.
- ◯ I can influence the protocol but ultimately the decision is out of my hands.
- ◯ I have little/no influence on the protocol that is used in this practice.
- ◯ Not applicable

Comment: _____

Electrophysiological Testing

Currently what is the length of appointment you have to complete an ABR/ASSR in natural sleep?

◯ 30 minutes
◯ 45 minutes
◯ 60 minutes
◯ 90 minutes
◯ 120 minutes
◯ Other (include length in comment section)

Comment: _____

Currently what is the length of appointment you have to compete a sedated ABR/ASSR in your department an/or the procedure center/OR?

◯ 30 minutes
◯ 45 minutes
◯ 60 minutes
◯ 90 minutes
◯ 120 minutes
◯ Other (include length in comment section)

Comment: _____

If an infant (0-6 months) comes to my office after referring the newborn hearing screening I complete the following: (check all that apply)

☐ Otoscopy
☐ Case history
☐ 1000 Hz Tympanometry
☐ 226 Hz Tympanometry
☐ Acoustic reflexes
☐ Distortion Product Otoacoustic Emissions
☐ Transient Evoked Otoacoustic Emissions
☐ Click ABR
☐ Tone burst ABR
☐ Chirp ABR
☐ Bone conduction ABR
☐ Chirp ASSR
☐ Tone burst ASSR
☐ Behavioral Observation
☐ Visual Reinforcement Audiometry
☐ Other (list in comments section below)

Comment: _____

For natural sleep or sedated electrophysiological testing on a new patient (with no previous testing completed), which test stimulus do you start with when testing air-conduction thresholds?

◯ Click ABR
◯ 250 Hz tone burst ABR
◯ 500 Hz tone burst ABR
◯ 1000 Hz tone burst ABR
◯ 2000 Hz tone burst ABR
◯ 4000 Hz tone burst ABR
◯ 250 Hz Chirp ABR
◯ 500 Hz Chirp ABR
◯ 1000 Hz Chirp ABR
◯ 2000 Hz Chirp ABR
◯ 4000 Hz Chirp ABR
◯ Chirp ASSR
◯ Tone burst ASSR
◯ Other (list in comment section)

Comment: _____

If an infant (6-12 months) comes to my office after referring the newborn hearing screening I complete the following: (check all that apply)

- ☐ Otoscopy
- ☐ Case history
- ☐ 1000 Hz Tympanometry
- ☐ 226 Hz Tympanometry
- ☐ Acoustic reflexes
- ☐ Distortion Product Otoacoustic Emissions
- ☐ Transient Evoked Otoacoustic Emissions
- ☐ Click ABR
- ☐ Tone burst ABR
- ☐ Chirp ABR
- ☐ Bone conduction ABR
- ☐ Chirp ASSR
- ☐ Tone burst ASSR
- ☐ Behavioral Observation
- ☐ Visual Reinforcement Audiometry
- ☐ Other (list in comments section below)

Comment: _____

When measuring a child's hearing thresholds via ABR/ASSR methods, I use the following audiometric transducer most of the time:

- ◯ Insert earphones
- ◯ Standard or supra-aural headphones

Comment: _____

If an infant comes to my office after referring the newborn hearing screening in one ear and passing in the other, I complete testing in:

- ◯ Only the ear that referred the screening
- ◯ Both ears

Comment: _____

If an infant comes to my office after referring the newborn hearing screening in one ear and passing in the other, I complete testing in this order:

- ◯ In the referred ear first followed by the passed ear
- ◯ In the passed ear first followed by the referred ear

Does your facility routinely provide re-screening of infants who refer on the newborn hearing screening for both their initial and repeat screening at their birthing hospital?

- ◯ Yes
- ◯ No
- ◯ Unsure

Comment: _____

Does your facility have a limited protocol (ie. Tymps, OAEs, and/or Click ABR only) for otherwise well babies with no risk factors who refer on the newborn hearing screening at their birth hospital?

- ◯ Yes we complete limited testing (tymps, OAEs and/or click ABR only)
- ◯ No we complete a full diagnostic evaluation
- ◯ Unsure

Comment: _____

My facility has a separate diagnostic protocol for babies who are referred from well-baby nurseries vs NICU babies admitted for greater than 5 days.

- ◯ Yes
- ◯ No
- ◯ Unsure

Comment: _____

If an infant/young child has a confirmed hearing loss I refer to the following professionals: (select all that apply)

- ☐ ENT for medical clearance
- ☐ PCP for medical clearance
- ☐ State early intervention program for services
- ☐ Audiologist for amplification
- ☐ Private speech-pathologist for evaluation
- ☐ Other (please specify)

Comment: _____

At your facility what risk factors require additional follow up testing? Select all that apply

- ☐ Ototoxic medication
- ☐ Meningitis
- ☐ Family history of hearing loss
- ☐ Intrauterine infections (including CMV, rubella, and herpes simplex virus)
- ☐ Prematurity
- ☐ Maternal diabetes
- ☐ Anoxia
- ☐ Malformations of the ear, nose or throat
- ☐ Apgar score from 0-3
- ☐ Low birth weight
- ☐ Hyperbilirubinemia
- ☐ Prolonged mechanical ventilation and/or severe respiratory distress
- ☐ Intensive care stay greater than 5 days
- ☐ Other (please specify)

Comment: _____

How many days until your next available natural sleep ABR?

- ◯ 0-5 days
- ◯ 6-10 days
- ◯ 11-15 days
- ◯ 15+ days (please specify if over 15 days in comment section)
- ◯ unsure

Comment: _____

How many days until your next available sedated ABR?

- ◯ 0-10 days
- ◯ 11-20 days
- ◯ 21-30 days
- ◯ 30+ days (please specify if over 30 days in comment section)
- ◯ unsure

Comment: _____

Out of the following factors, please select the top three reasons as to why it may be difficult to complete ABR testing within one appointment:

- ☐ Patient sleep state/waking up
- ☐ Electrical noise interference
- ☐ Appointment time too short
- ☐ Equipment issues
- ☐ Parent request to discontinue testing

Other/Comment: _____

During the past six months approximately what percentage of natural sleep ABRs could not be completed due to the infant sleep state/waking up?

- ◯ 0-25%
- ◯ 25-50%
- ◯ 50-75%
- ◯ 75-100%

During the past six months approximately what percentage of natural sleep ABRs could not be completed due to electrical noise/interference?

- ◯ 0-25%
- ◯ 25-50%
- ◯ 50-75%
- ◯ 75-100%

During the past six months approximately what percentage of natural sleep ABRs could not be completed due to not enough time in the appointment?

- ◯ 0-25%
- ◯ 25-50%
- ◯ 50-75%
- ◯ 75-100%

During the past six months approximately what percentage of natural sleep ABRs could not be completed due to equipment issues?

- ◯ 0-25%
- ◯ 25-50%
- ◯ 50-75%
- ◯ 75-100%

During the past six months approximately what percentage of natural sleep ABRs could not be completed due to parental request to discontinue testing?

- ◯ 0-25%
- ◯ 25-50%
- ◯ 50-75%
- ◯ 75-100%

Which of the following instructions do you provide to families prior to a natural sleep ABR appointment? (select all that apply)

- ☐ Bring infant sleep deprived
- ☐ Bring infant hungry
- ☐ Bring items that comfort the infants (bottle, blanket, pacifier, etc.)
- ☐ Bring the babies car seat for them to sleep in for testing.
- ☐ Do not put lotion on the infant's face
- ☐ Bring an additional adult to help keep the infant awake during the car ride
- ☐ Bring an additional adult if planning on bringing additional children (older siblings) to the appointment.
- ☐ Other (please specify)
- ☐ Our facility provides no instructions prior to the appointment

Other/Comment: _____

How do you provide families with instructions prior to a natural sleep ABR? (select all that apply)

- ☐ Over the phone when they schedule the appointment
- ☐ Over the phone via a confirmation call a few days before/or day before appointment
- ☐ A letter in the mail prior to the appointment
- ☐ I do not provide families with instructions

Behavioral Testing

Currently what is the length of appointment you have to complete an outpatient behavioral appointment for a child 6-36 months?

- ◯ 30 minutes
- ◯ 45 minutes
- ◯ 60 minutes
- ◯ 90 minutes
- ◯ Other (include length in comment section)

Comment: _____

When measuring a child's hearing thresholds who is 6-12 months of age, I use the following audiometric transducer most of the time

- ◯ Insert earphones
- ◯ Standard or supra-aural headphones
- ◯ Soundfield with loudspeakers/reinforcers at 0 degrees azimuth
- ◯ Soundfield with loudspeakers/reinforcers at 45 degrees azimuth
- ◯ Soundfield with loudspeakers/reinforcers at 90 degrees azimuth

Other/Comment: _____

When measuring a child's hearing thresholds who is 12-36 months of age, I use the following audiometric transducer most of the time

- ◯ Insert earphones
- ◯ Standard or supra-aural headphones
- ◯ Soundfield with loudspeakers/reinforcers at 0 degrees azimuth
- ◯ Soundfield with loudspeakers/reinforcers at 45 degrees azimuth
- ◯ Soundfield with loudspeakers/reinforcers at 90 degrees azimuth

Other/Comment: _____

For VRA testing what is your preferred position of patient?

- ◯ In a high chair
- ◯ On a caregiver's lap

Other/Comment: _____

Do you routinely use a high chair?

- ◯ Yes
- ◯ No

Comment: _____

Do you routinely use a test assist?

- ◯ Yes
- ◯ No

Comment: _____

What stimulus type do you routinely use? (select all that apply)

- ☐ Pure tones
- ☐ Warbled tone
- ☐ Narrowband noise
- ☐ Pediatric noise/FRESH noise
- ☐ Other (please specify)

Comment: _____

For VRA testing on a new patient (with no previous testing completed), which test stimulus do you start with when testing air-conduction thresholds?

- ◯ Speech
- ◯ Frequency specific stimuli (warble tones or noise)
- ◯ Other (comments)

Comment: _____

At what frequency do you typically begin conditioning? (select one)

- ◯ 250 Hz
- ◯ 500 Hz
- ◯ 1000 Hz
- ◯ 2000 Hz
- ◯ 4000 Hz
- ◯ 8000 Hz
- ◯ Other (please specify)

Comment: _____

What do you consider a normal VRA response? (select all that apply)

- ☐ 45 degree head turn
- ☐ 90 degree head turn
- ☐ eye shift
- ☐ look up
- ☐ other (please specify)

Other/Comment: _____

Do you use bone conduction for VRA testing?

- ◯ Yes
- ◯ No

Comment: _____

What are the top three pitfalls of VRA testing?

- ☐ Inadequate setup precluding the consistent judgement of head turns
- ☐ Inadequate communication between tester and test assist
- ☐ Attempting to condition with sub-threshold stimuli
- ☐ Not establishing clear responses at supra-threshold levels before descending to threshold
- ☐ Incorrect scoring due to false positive responses
- ☐ Rhythmical phasing that gives response clues to patient
- ☐ Use of toys/distractors that provides too little or too much engagement for the child
- ☐ Other (please specify)

Comment: _____

Do you have a lower limit stop criteria for testing threshold in children 6-36 months of age (ie. Do you not test below a certain intensity level)?

- ◯ Yes
- ◯ No

If you have a lower limit stop criteria for testing children 6-36 months what is the lowest level you stop at?

- ◯ 20
- ◯ 15
- ◯ 10
- ◯ 5
- ◯ 0
- ◯ Other

Do you consider the responses you record to be a minimal response level (MRL) or threshold?

- ◯ MRL
- ◯ Threshold
- ◯ Other (please specify)

Comment: _____

What is considered a normal hearing threshold or MRL for an infant 6-36 months of age?

- ◯ 15 dB HL or better
- ◯ 20 dB HL or better
- ◯ 25 dB HL or better
- ◯ Other (please specify)

Comment: _____

What are some factors that can potentially impact the reliability of the test results?

- ☐ State of alertness
- ☐ Patient attention
- ☐ Parental interference
- ☐ Presence of developmental/cognitive delay
- ☐ Other (Please specify)

Comment: _____

Demographics

Current state where you practice (select one):

- ◯ Alabama
- ◯ Alaska
- ◯ Arizona
- ◯ Arkansas
- ◯ California
- ◯ Colorado
- ◯ Connecticut
- ◯ Delaware
- ◯ Florida
- ◯ Georgia
- ◯ Hawaii
- ◯ Idaho
- ◯ Illinois
- ◯ Indiana
- ◯ Iowa
- ◯ Kansas
- ◯ Kentucky
- ◯ Louisiana
- ◯ Maine
- ◯ Maryland
- ◯ Massachusetts
- ◯ Michigan
- ◯ Minnesota
- ◯ Mississippi
- ◯ Missouri
- ◯ Montana
- ◯ Nebraska
- ◯ Nevada
- ◯ New Hampshire
- ◯ New Jersey
- ◯ New Mexico
- ◯ New York
- ◯ North Carolina
- ◯ North Dakota
- ◯ Ohio
- ◯ Oklahoma
- ◯ Oregon
- ◯ Pennsylvania
- ◯ Rhode Island
- ◯ South Carolina
- ◯ South Dakota
- ◯ Tennessee
- ◯ Texas
- ◯ Utah
- ◯ Vermont
- ◯ Virginia
- ◯ Washington
- ◯ West Virginia
- ◯ Wisconsin
- ◯ Wyoming

Current degree designation (please select most recent degree completed)

- ◯ AuD
- ◯ Master Degree
- ◯ PhD
- ◯ Other (please specify)

What is your gender?

- ◯ Female
- ◯ Male
- ◯ Non-binary
- ◯ Do not wish to respond

Are you now employed

- ◯ full time
- ◯ part time
- ◯ not employed
- ◯ retired
- ◯ other (please specify)

Comment: _____

State the number of years you have been working as an audiologist:

- ◯ 1-5 years
- ◯ 6-10 years
- ◯ 11-15 years
- ◯ 16-20 years
- ◯ +20 years

Of your number of years of experience, State the number of years you have been routinely seeing children:

- ◯ 1-5 years
- ◯ 6-10 years
- ◯ 11-15 years
- ◯ 16-20 years
- ◯ +20 years

Please choose the best terms to describe your current pediatric audiology work setting:

- ◯ private practice- owner
- ◯ private practice- employee
- ◯ hospital
- ◯ college/university
- ◯ ENT office
- ◯ department/warehouse store
- ◯ school
- ◯ other (please specify)

Comment: _____

How many audiologists in your facility/practice see children routinely?

- ◯ 1-3
- ◯ 4-7
- ◯ 8-10
- ◯ Over 10

What is the average number of diagnostic evaluations your facility performs each month for children age birth-6 months?

- ◯ 0-5
- ◯ 6-10
- ◯ 11-15
- ◯ 16+
- ◯ unsure

What is the average number of diagnostic evaluations your facility performs each month for children age 7 months to 2.11 years?

- ◯ 0-5
- ◯ 6-10
- ◯ 11-15
- ◯ 16+
- ◯ unsure

What is the average number of diagnostic evaluations your facility performs each month for children age 3-5 years?

- ◯ 0-5
- ◯ 6-10
- ◯ 11-15
- ◯ 16+
- ◯ unsure

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South Dakota Early Hearing Detection and Intervention Program: Using Teleaudiology to Conduct Infant Diagnostic Assessments

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Abstract

Teleaudiology allows patients and providers to bypass several economic and geographic barriers that impede the delivery and accessibility of audiological services. The South Dakota Early Hearing Detection and Intervention (EHDI) program recognized this benefit and created a teleaudiology infrastructure for the diagnostic assessment of infants. Using a hub-and-spoke model, a certified pediatric audiologist at the hub site assesses infants located at two spoke sites in South Dakota. Remote control software applications are used to provide a synchronous method of service delivery. The audiologist's test battery includes video otoscopy, tympanometry, and auditory brainstem response (ABR) testing. Since establishing the teleaudiology program, nine infant assessments have been completed. The South Dakota EHDI program will continue improving the teleaudiology project to ensure all infants in the state have access to pediatric audiological services.

Acronyms: AABR = automated auditory brainstem response; ABR = auditory brainstem response; ASHA = American-Speech-Language-Hearing Association; CDC = Centers for Disease Control and Prevention; DHH = deaf or hard of hearing; DPOAE = distortion product otoacoustic emissions; EHDI = Early Hearing Detection and Intervention; HRSA = Health Resources and Services Administration; LTF/D = lost-to-follow-up/lost-to-documentation; SDDOH = South Dakota Department of Health

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Telepractice enables clinicians to offer health services at a distance by linking clinician and patient or clinician and clinician via technology (American-Speech-Language-Hearing Association [ASHA], 2001). In the mid-1900s, researchers studied the application of telepractice to the field of audiology. Though slow in its initial stages of development, the availability of low-cost web cameras, broad-band connectivity, and highly computerized equipment catalyzed the growth of teleaudiology (Krumm & Syms, 2011).

Teleaudiology allows clinicians and patients to circumvent both geographic and economic barriers. Such barriers include long distances, detrimental weather conditions, travel expenses, and impaired mobility (ASHA, 2005b; Krumm et al., 2002). The challenges these barriers create are heightened by a worldwide shortage of audiologists (Hayes, 2012). Although this shortage disproportionally affects developing countries, rural areas of the United States are not immune to a lack of specialists. In response to these barriers, Swanepoel et al. (2010) said, "The majority of children and adults with hearing loss are isolated from the very services which may improve hearing and communication and reduce the potential negative effects of hearing loss on social interaction, education, and vocational opportunity" (p. 197).

Delayed diagnosis of adults who are deaf or hard of hearing (DHH) may adversely affect their activities of daily living. Within the pediatric population, untreated hearing loss can affect a child's speech, language, cognitive, and social development (ASHA, n.d.). As such, the timely diagnosis of hearing loss and enrollment in intervention services are of paramount importance.

In its position statement on telepractice, ASHA (2005a) stated that telepractice is an appropriate model of service delivery. ASHA subsequently indicated that such services must be of the same quality as face-to-face services. This quality can be achieved through use of a synchronous (real-time) method of service delivery, where a clinician at one location directly tests a patient at a distant location (ASHA, 2005b). A key component of this method is the presence of a facilitator at the patient's location. The facilitator is trained on video otoscopy, electrode and insert placement, and observation of the patient's response patterns (Krumm, 2007). Remote control computing allows the clinician to control equipment at the testing site (Krumm et al., 2002).

Several audiological services have been delivered via telepractice, and research studies validate the accuracy and feasibility of such services. Edwards et al. (2012) summarized the literature pertaining to the use of

telepractice in speech-language pathology and audiology; all studies reviewed in the meta-analysis denoted telepractice as an effective medium for the diagnosis and treatment of children and adults with communication and/or hearing limitations. Another systematic review of teleaudiology validated its use for screening, diagnostic, and intervention services (Swanepoel & Hall, 2010). These systematic reviews cite findings by the following researchers: Lancaster et al. (2008), who found real-time otoscopy and immittance testing to be feasible and reliable; Givens & Elangovan (2003), who used remote control software applications to provide real-time diagnostic audiometry services; and Krumm et al. (2008), who conducted a study with 30 infants and found that results obtained by telemedicine and by conventional face-to-face methods were essentially equal for both distortion product otoacoustic emissions (DPOAEs) and automated auditory brainstem response (AABR) testing.

Teleaudiology applications have also been used by several state Early Hearing Detection and Intervention (EHDI) programs. The Joint Committee on Infant Hearing (JCIH, 2019) endorses the early detection and intervention of children who are DHH to "to maximize [their] language and communication competence, literacy development, and psychosocial well-being" (p. 3). This goal is achieved by following EHDI's 1-3-6 benchmarks: all newborns should be screened for hearing loss no later than one month of age; newborns who refer on their initial screening should receive a diagnostic evaluation no later than three months of age; and infants who are identified as DHH should enroll in early intervention services no later than six months of age (JCIH, 2019). Several projects have demonstrated the success of telehealth's application to the EHDI program. For example, Hayes (2012) reported that Children's Hospital Colorado established connections with Guam's EHDI program 7,000 miles away. Due to a shortage of audiologists on the U.S. island territory, Children's Hospital Colorado worked with professionals in Guam to create a teleaudiology infrastructure for assessing infants. With appropriate technology, acceptable test protocols, and a suitable test environment, the Guam EHDI project demonstrated the viability of using remote control software to conduct infant diagnostic assessments.

As demonstrated by the aforementioned research studies and pilot project, telepractice is an effective medium for the delivery of audiological services. Both increasing internet connectivity and improvements in technology are bridging the gap between patients and providers separated by geographic and economic barriers (Swanepoel & Hall, 2010). Telepractice and its associated benefits will create both global and local improvements in the delivery of audiological services. Givens & Elangovan (2003) argued that teleaudiology is not so much an alternative method for diagnostic testing, as this definition portrays telehealth as an inferior mode of service delivery; rather, teleaudiology has become a wise, cost-effective, and convenient method for both clinicians and patients alike. Recognizing these benefits, researchers and professionals working with the South Dakota EHDI program adopted teleaudiology for the provision of infant diagnostic evaluations.

History of South Dakota EHDI Program

South Dakota's EHDI program was established in 2001 after the state received funding from the Centers for Disease Control and Prevention (CDC). Additional funding was provided by the Health Resources and Services Administration (HRSA) in 2015 as part of a nationwide effort to develop additional EHDI programs; recruit and train staff on EHDI goals; ensure families have accurate information on their child's hearing status; and foster family-to-family support after a child has been identified as DHH (HRSA, 2019). This funding led to the creation of the South Dakota EHDI Collaborative. The Collaborative is a partnership between the University of South Dakota (Department of Communication Sciences and Disorders; Department of Nursing) and the Department of Health's State EHDI program, in addition to other partners, including the South Dakota School for the Deaf.

High Lost-to-Follow-Up/Lost-to-Documentation Rates

South Dakota is one of six states lacking a legislative mandate for a newborn hearing screening program (Messersmith et al., 2014). Despite this fact, South Dakota implements universal newborn hearing screening. In 2016, 98% of newborns in the state were screened for hearing loss (CDC, 2016). However, high lost-to-follow-up/lost-to-documentation (LTF/D) rates remain a priority for the South Dakota EHDI program (HRSA, 2019). These rates are highest among American Indian families and infants born to low-income families living in western and central South Dakota. Several reasons account for the state's high LTF/D rates such as limited pediatric audiological services, rurality/geographic isolation, and high poverty levels.

Limited Pediatric Audiological Services

As is common in other states and countries, South Dakota has a shortage of pediatric audiologists. There are five pediatric diagnostic follow-up sites in South Dakota. Four sites are located in the southeastern corner of the state, and one is located on the far western side of the state. Families located in central and northern South Dakota would need to drive three to four hours to receive testing at one of these follow-up sites.

Rurality/Geographic Isolation

Another challenge facing residents is South Dakota's classification as a frontier state. Of the 66 counties in South Dakota, 34 are considered frontier, having a population density of less than six people per square mile. In addition, geographic isolation prevents many families from seeking services at tertiary healthcare centers due to transportation difficulties and/or financial limitations. Detrimental weather conditions can also hinder a family's ability to travel.

High Poverty Levels

Poverty is a major factor contributing to South Dakota's high LTF/D rates. In 2018, South Dakota's poverty rate

was 13.1% (compared to the national average of 11.8%). This percentage equates to 115,572 individuals living in poverty based on the state's estimated population of 882,235 residents in 2018 (United States Census Bureau, 2018).

Solution to High LTF/D Rates

The South Dakota EHDI program aims to lower these high LTF/D rates and ensure infants who are DHH receive a timely diagnosis and early intervention services. Based on the estimate that three to four of every 1,000 babies are born with some level of hearing loss in the United States, approximately 33 to 44 babies are identified as DHH in South Dakota each year (South Dakota Department of Health, 2019). Determined to diagnose all infants who are DHH and overcome the previously mentioned barriers, the Collaborative established two teleaudiology sites in South Dakota. A description of how South Dakota EHDI created a teleaudiology infrastructure, in addition to the equipment required for synchronous diagnostic evaluations, will be provided in the remainder of this article.

Creation of Teleaudiology Infrastructure

From 2016 to 2017, the Collaborative established two teleaudiology sites in South Dakota. An outside consultant with expertise in teleaudiology assisted the Collaborative in developing the program's infrastructure.

Method

Using a hub-and-spoke paradigm, synchronous (real-time) methods are used to assess infants for hearing loss. A hub-and-spoke model allows healthcare professionals (located at a centralized *hub* site) to assess patients located at distant *spoke* sites via telepractice. The infant and family receive testing at the spoke site location, where trained medical personnel place equipment on the infant (e.g., otoscope speculum, electrodes, insert earphones) and assist the family in preparing the infant for sleep. The pediatric audiologist performs testing and evaluates test results at the hub location via remote control software applications. Routine maintenance and annual calibration of equipment is performed at the spoke site locations.

The University of South Dakota Speech Language and Hearing Clinic, located in Vermillion, South Dakota, serves as the hub site. The first spoke site is located at the Sanford Health Winner Regional Hospital in Winner, South Dakota (approximately 180 miles from the hub location). The second spoke site is located at Avera Saint Luke's Hospital in Aberdeen, South Dakota (approximately 260 miles from the hub location).

At the Aberdeen spoke site, both the initial screen and rescreen are performed prior to diagnostic testing. The protocol for the Winner spoke site is slightly different. If the infant refers on the initial screen, the family is referred to diagnostic testing. The spoke site assistant begins the appointment by performing the rescreen, and the audiologist only moves forward with diagnostic testing if the infant refers on this second screen.

A certified pediatric audiologist at the University of South Dakota clinic (hub site) remotely performs the diagnostic evaluations. The audiologist's test battery includes video otoscopy, tympanometry, and ABR testing. As mentioned in the introduction, completing these assessments via teleaudiology is proven to be a reliable and valid method; results obtained through conventional face-to-face methods and through telemedicine are essentially equal (Krumm & Syms, 2011; Lancaster et al., 2008).

Currently, these services are being provided through the HRSA grant, and no entity (patient or third party) is billed for the diagnostic testing. When the teleaudiology program transitions out of the pilot phase, services will be billed to the responsible entity, which may be the patient and/or a third-party provider (e.g., Medicaid, Medicare, private insurance).

Training

Before teleaudiology appointments were scheduled, medical personnel at the spoke site locations were trained on proper procedures for placing equipment and interacting with family members. Providing this in-person training was necessary to guarantee that spoke site assistants were well prepared.

The South Dakota EHDI Collaborative also created toolkits for personnel at the spoke sites. These toolkits explain how to complete otoscopy, ABR testing, otoacoustic emissions (OAE) testing, and tympanometry. They also include scripts for personnel to use when discussing information with parents.

In addition to toolkits and in-person training, PowerPoint presentations and video trainings were provided to spoke site assistants. Medical personnel can visit the YouTube channel titled "Communication Support through Aids and Technology" to see a list of training videos uploaded by the Collaborative. Such videos offer training on swaddling infants, completing otoscopy, scrubbing for electrode placement, placing electrodes, removing electrodes, placing insert earphones, and preparing the infant for bone conduction testing. An example of a training video can be viewed at https://www.youtube.com/watch?v=P9CltdLNLG4.

Equipment

The spoke site locations must have specific test equipment for assessments to be completed. A list of supplies and additional requirements is shown in Table 1. Necessary equipment made available to the spoke sites' trained personnel included the following items: video otoscope, ABR equipment, OAE equipment, tympanometry equipment, a computer to operate hardware and software programs, web camera, and ancillary supplies (e.g., specula and probe tips). The spoke site must also have an adequate upstream speed (at least 3 megabit) and permissible ambient noise levels.

Two types of software are necessary for completing synchronous testing: 1) software allowing remote access to the spoke site computer and 2) software allowing video and audio connection between the hub site and spoke sites. The South Dakota EHDI Collaborative uses

Table 1
Necessary Supplies for Teleaudiology Infrastructure

Equipment	*Software*	*Additional Requirements*
Video otoscope	Software allowing remote access to spoke site computer	Adequate upstream speed at spoke site (must be at least 3 megabit)
ABR equipment	Software allowing video and audio connection between hub and spoke sites	Permissible ambient noise levels at spoke site
OAE equipment		Internet connection at spoke site
Tympanometry equipment		Trained technicians at spoke site
Computer to run hardware and software programs		
Web camera		
Ancillary supplies (probe tips, specula, etc.)		

Note. Establishing a teleaudiology program requires standard audiology equipment, specific software programs, and additional standards required of the spoke site itself. ABR = auditory brainstem response; OAE = otoacoustic emissions

TeamViewer to obtain remote access to both spoke site computers. For video and audio connection, the Collaborative has tested two types of software programs, with a different program being used at each spoke site.

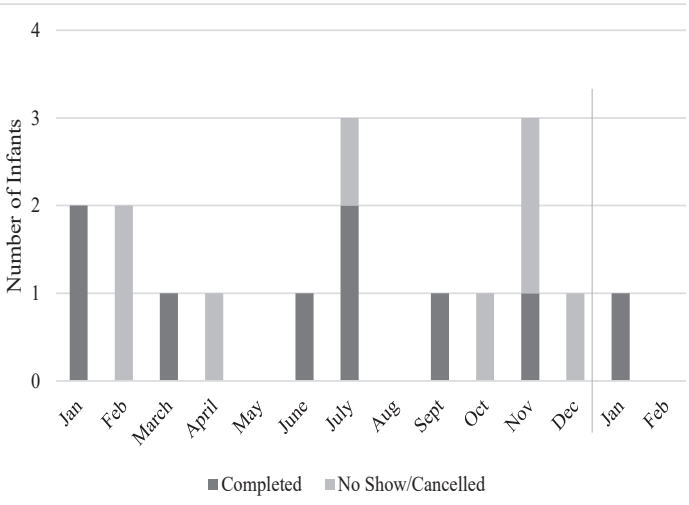
For appointments with Sanford Health Winner Regional Hospital, *Skype for Business* is used for video and audio connection. Although this program is HIPAA compliant, cost effective, and user friendly, it provides a somewhat informal connection between the audiologist and family. For appointments with Avera Saint Luke's Hospital, *Cisco Systems* is being used. Compared to *Skype for Business*, this program offers a more formal connection between the patient and provider. *Cisco Systems* is also HIPAA compliant and allows for clearer imaging. However, *Cisco Systems* is a more expensive software program, and both the hub site and spoke site need to purchase the program. Both *Skype for Business* and *Cisco Systems* have their advantages and disadvantages, and one program is not necessarily superior to the other.

At the time these software programs and equipment items were purchased, the HRSA grant was held by the South Dakota Department of Health (SDDOH). As such, the SDDOH purchased the teleaudiology equipment (subject to HRSA approval) before subcontracting the grant to the University of South Dakota. The EHDI Collaborative, cognizant of decreased funding opportunities and the expense of audiology equipment, did its best to minimize cost by taking advantage of cost-effective or previously-held software programs (e.g., using the *Skype for Business* program with a HIPAA certificate and business affiliation agreement; using a preexisting electronic medical records system for data entry).

Results

Testing at the teleaudiology sites began in January 2019. As of February 2020, a total of nine infants have received diagnostic assessments. Eight additional appointments were classified as *no show* or *cancelled*. See Figure 1 for a timeline of assessments from 2019 to 2020.

Figure 1
Appointments Completed and No Show/Cancelled Appointments at Both Spoke Sites from January 2019 to February 2020



Although one spoke site was consistently referring infants to their teleaudiology location, the other spoke site was facing challenges with its referral process. As a result, assistants with the EHDI Collaborative spread awareness of the teleaudiology program to additional pediatricians, obstetricians/gynecologists, and family care physicians nearest this spoke site through postcards, emails, and presentations. In addition, contact information for the teleaudiology sites was sent to the South Dakota Department of Health, which now lists both the Winner and Aberdeen spoke sites on its website. It is expected that the number of infants tested via teleaudiology will increase as more healthcare providers and families become aware of the program.

Counseling

Following a conventional face-to-face assessment, the audiologist immediately provides the family with results. When testing via teleaudiology, discussing results with parents can differ based upon the audiologist's and family's preferences.

The South Dakota EHDI Collaborative has determined its preferred method for delivering results. When no hearing loss is identified, the audiologist provides the family with results at the time of testing. When a hearing loss is identified, the audiologist either conducts a virtual meeting with the family or determines another appropriate route for conveying these results. Krumm (2007) stressed the need for future research on proper counseling procedures for telehealth appointments. Research should focus on counseling methods in the event that a parent experiences denial upon discovering his or her child has been identified as DHH.

Collecting Feedback from Spoke Sites

After diagnostic testing had been performed at both teleaudiology spoke sites, the Collaborative collected feedback from the spoke sites' trained assistants. The Collaborative wanted to understand the assistants' experience with the teleaudiology program and identify the need for potential improvement in training. Results of the formal feedback survey are displayed in Table 2.

Table 2
Formal Feedback Results from Teleaudiology Spoke Sites

Question	***Winner Regional Hospital*** *Date: 10/09/2019*	***Aberdeen's Avera St. Luke's Hospital*** *Date: 11/11/2019*
1. Has the teleaudiology spoke site been a useful resource since it's been established? Why or why not?	"Yes - it has saved families a lot of driving by allowing them to do the testing closer to home."	Respondent 1: "The training was great when we started but then we didn't have any [additional trainings], so we set up practice trainings a couple different times, but it took 3 hours out of our day."
2. Was the training you received sufficient to prepare you for the teleaudiology sessions? If not, what could be improved?	"Yes, it was sufficient. Additional information about how the testing works would have been helpful."	Respondent 1: "Maybe we should set up a refresher [course] to go through the equipment briefly."
3. Would a refresher training course be beneficial?	"Not for me, but possibly for others who could fill in for me but do not regularly assist with the testing."	Respondent 2: "I think a yearly competency [training] would be good. Step-by-step visuals are great."
4. What improvements could be made to the teleaudiology spoke site?	"None"	Respondent 1: "Trying to get the word out and trying to get more clientele."

Overall, feedback from both spoke sites was positive. The assistants believed the teleaudiology program was a useful resource for families with limited access to audiological services. Two opportunities for improvement were suggested in the formal feedback survey. First, the assistants commented on the need for refresher training courses once or twice a year, especially if new assistants join the teleaudiology team. Second, the personnel recommended that information on the teleaudiology spoke sites be made available to more healthcare providers in their respected locations.

Since the survey was completed, the assistants' suggestions were reviewed by the Collaborative and progress has been made to improve the teleaudiology program. The hub site's pediatric audiologist agreed with the recommendation to present refresher training courses. In addition, the previously mentioned training videos and PowerPoint presentations created by the Collaborative (see "Training") have been placed in the medical facilities' continuing education platforms and are available for review at any time by spoke site personnel. To address the second suggestion, information regarding the

teleaudiology program has been sent to nearby physicians who may contribute to the spoke sites' referral processes.

Conclusion

With technological advances and increasing Internet connectivity, telepractice proves to be an effective avenue for delivering healthcare services. Teleaudiology, though slow in its initial stages of development, has gained increasing attention. Audiological services delivered via technology allow patients and providers to bypass several barriers—both geographic and economic—that too often separate individuals from the very services that could improve their hearing and communication.

The South Dakota EHDI Collaborative's teleaudiology program and its adoption of a hub-and-spoke model has demonstrated the feasibility of using remote control software applications to complete video otoscopy, tympanometry, and ABR testing. Infants born in the western and central portions of South Dakota can now receive diagnostic audiological testing that may have been challenging or nearly impossible to attain prior to the development of the two spoke sites.

Future research on teleaudiology should focus on patient satisfaction with the teleaudiology program. Although feedback from spoke site assistants has been positive, formal feedback should also be collected from families whose children have undergone testing at the spoke sites. Additional research should be conducted on how best to counsel families whose children have been identified as DHH following a teleaudiology evaluation.

Regardless of where children live, whether it be in a rural area of the United States or a developing country, they deserve access to audiological services—services that could largely impact their speech, language, cognitive, and social development. The way in which to broaden their access to these services is no enigma; countless studies (Edwards et al., 2012; Swanepoel & Hall, 2010; Givens & Elangovan, 2003; Krumm et al., 2008) corroborate teleaudiology's status as a valid and reliable method of service delivery. By choosing to welcome the advent of teleaudiology and embrace its benefits, barriers to audiological services will become a challenge of the past.

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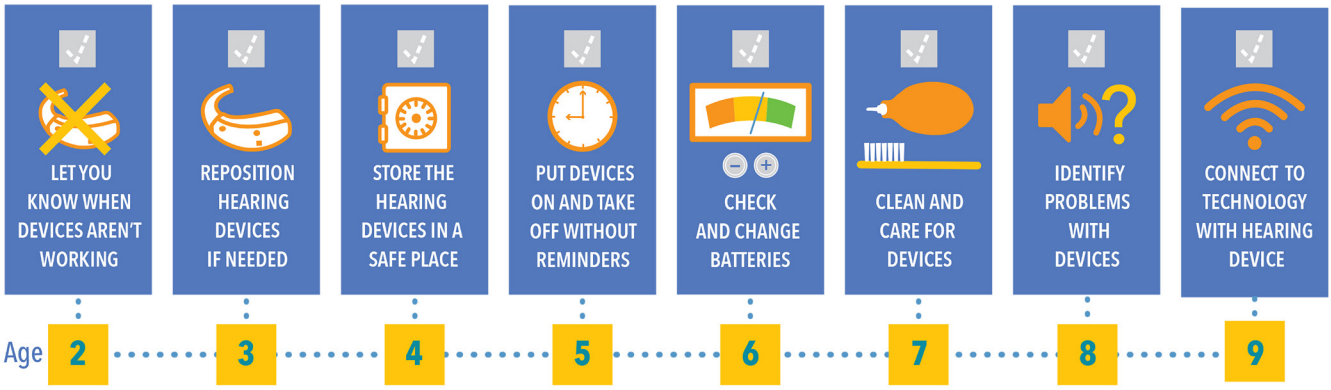
EHDInfo

HEAR to LEARN
Resources for Parents of Children Who are Deaf or Hard of Hearing

http://heartolearn.org

HELP CHILDREN USE AND CARE FOR HEARING DEVICES

Practice these tasks together until you are confident your child can:



Not all children may be ready to do these skills by the ages listed in this guideline. Talk to your child's audiologist for ideas on how to teach your child to independently care for their hearing devices.

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EHDI System Effectiveness: The Impact of Community Collaboration

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Abstract

Collaboration between statewide stakeholders is integral to ensuring that families who have children who are deaf or hard of hearing successfully access the resources of Early Hearing Detection and Intervention systems. However, collaboration between stakeholders takes time, resources, and common goals. The Idaho Community Collaboration (ICC) project brought statewide state and non-state agencies together to assess the Early Hearing Detection and Intervention system in Idaho through data collection and survey. With the objective data obtained from these data sources, the ICC was able to take first steps in meeting the needs of the state's family and children through collaborative decision making and resource development.

Acronyms: DHH = deaf or hard of hearing; EHDI = Early Hearing Detection and Intervention; EI= early intervention; ICC = Idaho Community Collaboration; IESDB = Idaho Educational Services for the Deaf and Blind; IFSP = Individual Family Service Plan; ISB = Idaho Sound Beginnings; ITP = Infant Toddler Program; NCHAM = National Center for Hearing Assessment and Management

Keywords: EHDI, collaboration, stakeholders, assessment

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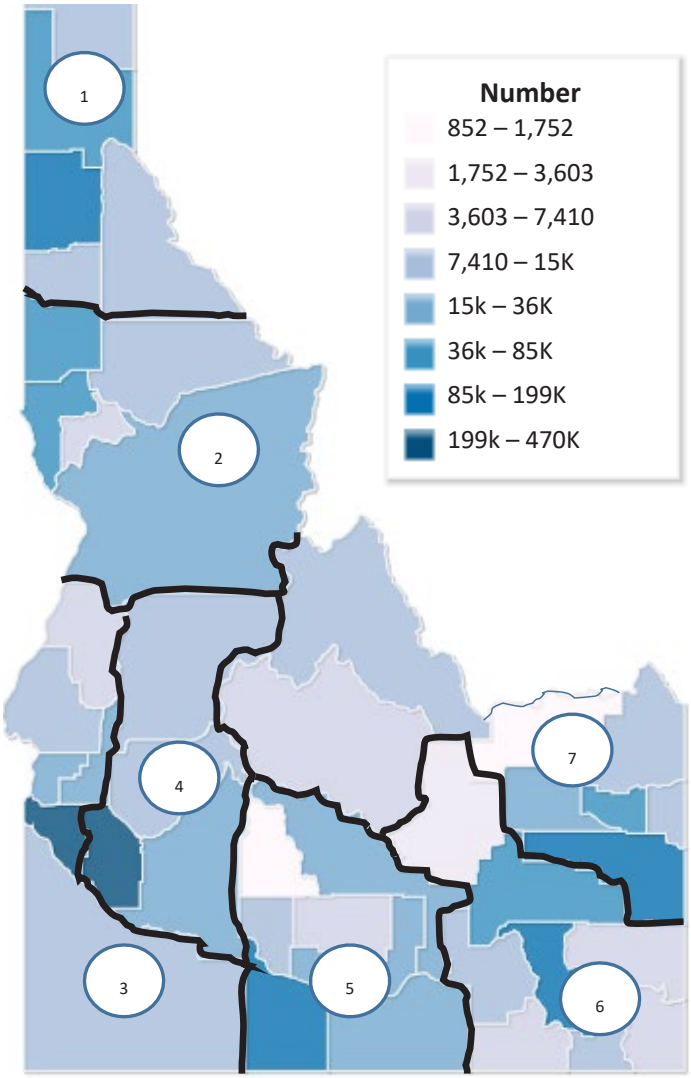
Early Hearing Detection and Intervention (EHDI) systems include stakeholders from varying state and non-state agencies including newborn screening programs, Part C providers, educational services for the Deaf and Blind, parents, and parent support agencies. As EHDI systems strive to adhere to best practice guidelines, successful implementation depends on multiple providers and coordinated systems (Brown et al., 2019). For example, if an infant is screened at birth, successful follow-up is dependent on factors such as how the information is presented, if/how scheduling for diagnostics takes place, families' understanding of importance of diagnosis, and timeliness of assessment and initiation of intervention.

In 2009, the National Center for Hearing Assessment and Management (NCHAM) supported strategic planning activities to help state EHDI systems strengthen their programs and identify challenges (White & Blaiser, 2011), including collaboration as one key component of the strategic planning analysis. Although collaboration is often touted as an integral aspect of the EHDI system, in actuality, communication may be limited to periodic interactions about common factors and processes with little integrated engagement focused on systematic improvement of outcomes for children and families. Many factors can influence collaboration such as turf (i.e., feeling that a child *belongs* to one entity more than another), time (i.e., barriers related to caseload size, amount of time allocated to communication), and trust (i.e., a mutual feeling of respect between stakeholders). See Himmelman (1996) for a review.

Collaboration and coordination can be even more challenging in a state with substantial rural or remote areas. The state of Idaho is divided into seven public health regions used by multiple entities including the Department of Health and Welfare, containing Idaho Sound Beginnings and Idaho Infant Toddler Program (ITP), and Idaho Educational Services for the Deaf and Blind (IESDB; Figure 1). As shown in Figure 1, a substantial portion of Idaho is considered *rural*: the panhandle of Idaho (Regions 1 and 2), most of southwest Idaho (Region 3), and south central Idaho (Region 5). Region 4, while one of the most populated regions in the state (therefore counted as *suburban/urban* for the purposes of this project) still contains two counties that are classified as

rural. Similarly, in Regions 6 and 7, located in southeastern Idaho and eastern Idaho respectively, two to three of the eight counties are classified as rural. In fact, because of some of the low population density, many of Idaho's rural populations are considered *frontier* because of their isolation from population centers and services (Idaho Department of Health & Welfare, 2018). In these regions, there are limited resources specific to EHDI systems, such as pediatric audiologists, otolaryngology specialists, and early intervention providers with experience serving children who are deaf or hard of hearing (DHH). To serve families and ensure adhesion to EHDI system best practice, statewide teams must look at outcomes and processes regionally as well as statewide to better decipher the specific needs of the families who reside in more remote locations.

Figure 1
Idaho 2019 Population Estimates Based on U.S. Census Bureau 2010 Census Data



In addition to challenges faced through reduced population and access to services, Idaho is one of three states in the United States that does not mandate newborn hearing screening (NCHAM, 2020). Although there is an active newborn hearing screening advisory committee, the need for more formalized collaboration and discussion is particularly important when there is no legislation or state funding to support these processes.

A foundation grant was awarded to faculty at Idaho State University with the primary goal of improving outcomes for children who are DHH and their families across the state of Idaho through enhanced stakeholder collaboration. This paper outlines the process that was followed in developing the Idaho Community Collaboration with stakeholders who are involved with families of children who are DHH from newborn hearing screening to the child's enrollment in the Part B system.

Identify Key Stakeholders

The first step in developing the Idaho Community Collaboration (ICC) was to identify key stakeholders in Idaho's EHDI system. Although there are many entities involved with families of children who are DHH within the state of Idaho, the focus of this group was to include stakeholders who represent various aspects of the statewide systems. As shown in Table 1, five stakeholders were identified: the newborn hearing screening program (Idaho Sound Beginnings, ISB), the primary state Part C provider (the Infant Toddler Program, ITP), the state school services for the deaf and blind (Idaho Educational Services for the Deaf and Blind, IESDB), a statewide hospital system that provides clinical audiological and speech-language pathology services (St. Luke's Hearing and Balance Center), and the family advocacy and support organization specific to children who are DHH (Idaho Hands and Voices). Two faculty members from Idaho State University's Speech-Language Pathology and Audiology programs participated in the ICC with the primary roles of facilitating discussions, coordinating processes, engaging graduate students in communication sciences and disorders, and disseminating findings. There were ten participants in the meetings but many of these participants held more than one role, specifically, in addition to their professional position, they were parents of children or adults who are DHH.

Define the Process

Prior to the first meeting, each stakeholder (or stakeholder group) was asked to develop a map based on their understanding of the current process from newborn hearing screening to enrollment in Part B services. Existing examples were provided, such as the EHDI Guidelines for Pediatric Medical Home Providers (https://www.aap.org/en-us/advocacy-and-policy/aap-health-initiatives/PEHDIC/Documents/Algorithm1_2010.pdf). At the first meeting, each stakeholder shared their map and included questions related to their own perspectives on the strengths, opportunities, and points of clarification needed for each step of the process. As a group, each of the maps

Table 1
Participants and Roles of Idaho Community Collaboration Members

Entity	Position	Role	Secondary Role
St. Luke's Hearing and Balance Center	Pediatric audiologist	Pediatric audiologist	Mother of child who is DHH
	Speech-Language Pathologist	Speech-Language Pathologist	Mother of adult who is DHH
Idaho Sound Beginnings	Administrator	Administrator	
	Parent follow-up consultant	Parent follow-up consultant	Mother of children who are DHH
Infant Toddler Program	Administrator	Administrator	
Idaho Hands and Voices	Director of Parent Support Organization	Director of Parent Support Organization	Mother of child who is DHH
Idaho Educational Services for the Deaf and Blind	Administrator, Director of Outreach	Administrator	
	Director, Part C	Deaf Educator	Interpreter
Idaho State University	Faculty member, Primary Investigator	Coordinator	Speech-Language Pathologist
	Faculty member, Co-Primary Investigator	Co-Coordinator	Audiologist

Note. DHH = deaf or hard of hearing

were discussed and compared to determine the common questions or points of clarification needed for each part of the process. As a result of these discussions, it was decided there was a need for coordinated data collection to distinguish what was actually occurring in practice and to examine the perceptions of what might be occurring at different parts of the process.

An unintended benefit of the mapping process was an opportunity for partners to learn about resources and options that were available to providers and families. For example, information about funding resources for hearing aids was not universally known across the stakeholders (e.g., IESDB and/or ITP were not aware of the same funding resources as hospital-based audiologists). Having this opportunity to discuss the processes and resources in place, as well as how they may differ from region to region was beneficial.

Gather Information

Following the stakeholder discussion, a data collection system was developed to cross-check the information between ISB, ITP, and IESDB. Existing data sharing agreements facilitated this process. The system was created and cross-referenced by administrators for each of the stakeholders (ITP, ISB, and IESDB). Based on the stakeholder maps that were created in the first step, data collected included 21 data points:

- Child identification number
- Region
- Screening date
- Screening result
- Diagnosis date
- Hearing status
- Language spoken
- Early intervention status
- Idaho Educational Services for the Deaf and Blind (IESDB) phone contact date
- IESDB assessment date
- Primary interventionist
- Discipline of interventionist
- Secondary interventionist
- Discipline of secondary interventionist
- Individual family service plan start date
- Scheduled visits
- Completed visits
- Infant toddler speech-language pathologist start date
- If closed during intake, why?
- If not in services why?
- Notes (indicate if not eligible for specific service)

In addition to the state-based system, a survey was sent to all of the families who had gone through a newborn hearing screening for a three-year period. This data was used as a way to cross-reference and compare the data that is collected within state systems and families' perceptions of the processes that had occurred.

The parent survey incorporated key concepts from Bush et al. (2014). An electronic survey via Qualtrics was distributed to 591 families via email addresses collected by ISB's newborn hearing screening form. Because of the collaboration, the email was distributed by ISB so no personal health information was shared with the investigators. Fifty surveys were returned due to wrong email addresses in the EHDI system. Surveys were

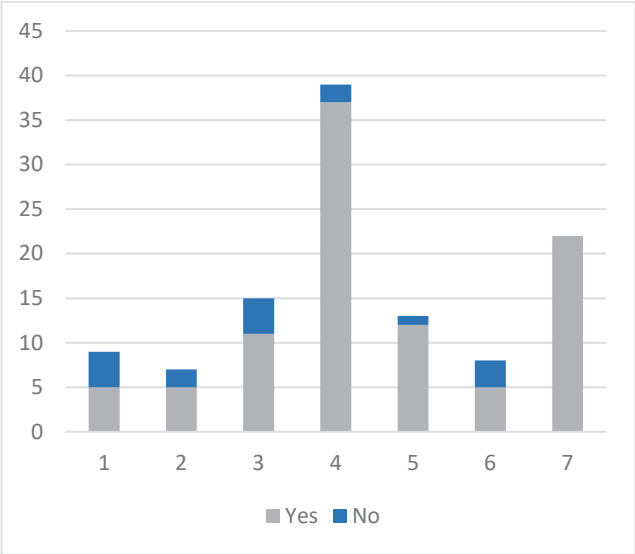
completed by families in all regions, with 116 surveys completed, yielding a 21.4% return rate. Broken down by region, 7.7% (*n* = 9) of the responses came from Region 1, 6.0% (*n* = 7) from Region 2, 12.9% (*n* = 15) from Region 4, 33.6% (*n* = 39) from Region 4, 12.1% (*n* = 14) from Region 5, 7.7% (*n* = 9) from Region 6, and 19.8% (*n* = 23) from Region 7. Having responses from each region is particularly important in Idaho, where there are regional discrepancies in terms of access to pediatric services in more urban/suburban areas (i.e., Regions 3, 4, and 6) and those in more rural/remote areas (i.e., Regions 1, 2, 5, and 7).

Of the 111 families who responded to the question, "When were you told the results of the hearing screening?" 63.1% (*n* = 70) received the results of the screening right after the screening was performed and 30.6% (*n* = 34) were told the results before they left the hospital. One family reported that they were not given the results of the hearing test. Of the 113 families who answered a related but separate question, "Who made the follow-up hearing appointment?", most families (*n* = 61, 54%) made the follow-up appointments themselves, followed by the hearing screener (*n* = 15) as part of a regional pilot program.

According to 113 responses to the question, "Were you able to follow up within 3 months of age?", 85% of the families (*n* = 97) reported that they were able to follow up within three months of age, 16 (14.1%) families reported they were not able to follow up in this timeframe (Figure 2). Distance and home responsibilities were identified as the primary factors that made follow-up challenging for families, followed by health insurance and scheduling.

Figure 2

Parent Response to "Were You able to Follow-up Within 3 Months of Age?" by Region



Of the 112 families who responded to the question about the importance of follow-up testing, over half of the families (56.3%, *n* = 63) felt that follow-up testing related to their child's hearing was extremely important. It is important to note that 10 families (8.9% of the sample) were unsure of the importance or thought follow-up testing was not very important. Of the 114 families that responded to the overall survey, 45.6% (*n* = 52) had children diagnosed with hearing loss, 50.9% (*n* = 58) did not have children diagnosed with hearing loss, and 3.5% (*n* = 4) did not know if their child had a hearing loss or not. Approximately 37.5% (*n* = 42) of the 112 families who responded were told to go to an audiologist (non-specified) for the follow-up appointment, while 32 (28.6%) families were told to follow up with a pediatric audiologist. The average age of identification/diagnosis of hearing loss was 3.16 months of age; however, this varied from region to region (Figure 3).

Fit with Hearing Aids

The average age children received hearing aids was 9.86 months, with a range of 5.3 to 14 months (Figure 3). More than half (71.4%) of the 56 families who responded, reported that the amount of time required to be fit with hearing aids was what they expected (*n* = 28) or faster (*n* = 12). Approximately 19.6% (*n* = 11) of the families reported that it took longer than expected.

Enrollment in Early Intervention

Families were asked to answer questions about who provides early intervention services and what types of services they received. According to the families who responded to "who provides early intervention services to your family" (with a check all that apply response), families reported that they receive services from IESDB (*n* = 47; 52.2%) and the ITP (*n* = 53; 58.9%) while five (5.6%) were not sure and 34 (37.8%) indicated some other service provider.

When asked what type of services their child received (with a check all that apply response), parents that responded (*n* = 85) reported audiology as the most commonly received type of service (*n* = 42; 49.4%), followed by early intervention (*n* = 38; 44.7%), and speech-language pathology (*n* = 32, 37.6%). This was slightly different than the information that was gained from the state system database. The differences between these two data sources indicates, perhaps, that families are often unsure of the types of services that they are receiving, particularly in a home-based, coaching model where a provider or multiple providers may overlap in the services that are offered (i.e., language or cognitive development). Per the state system database, 2% (*n* = 3) of families received early intervention services four times per month by their early intervention provider (Figure 4). The majority of Idaho families who are enrolled in Part C Early Intervention (EI) receive services twice per month or less (Figure 4).

Parents were asked to report the communication approaches (with a check all that apply response) used by their child. Listening and Spoken Language (*n* = 50; 48.5%) and Total Communication (*n* = 45, 43.7%) were the most commonly used communication modalities of the families who responded to the survey. American Sign Language (ASL) was used by 35.9% of families (*n* = 37) who participated in the study. It should be noted that this is a higher level of sign language/total communication use than other states typically report (e.g., Brown, 2006).

Figure 3

Average Age (in Months) of Child When the Hearing Loss Diagnosis was Made and Age (in Months) Child was Fit with Hearing Technology by Region Based on Parent Survey Results

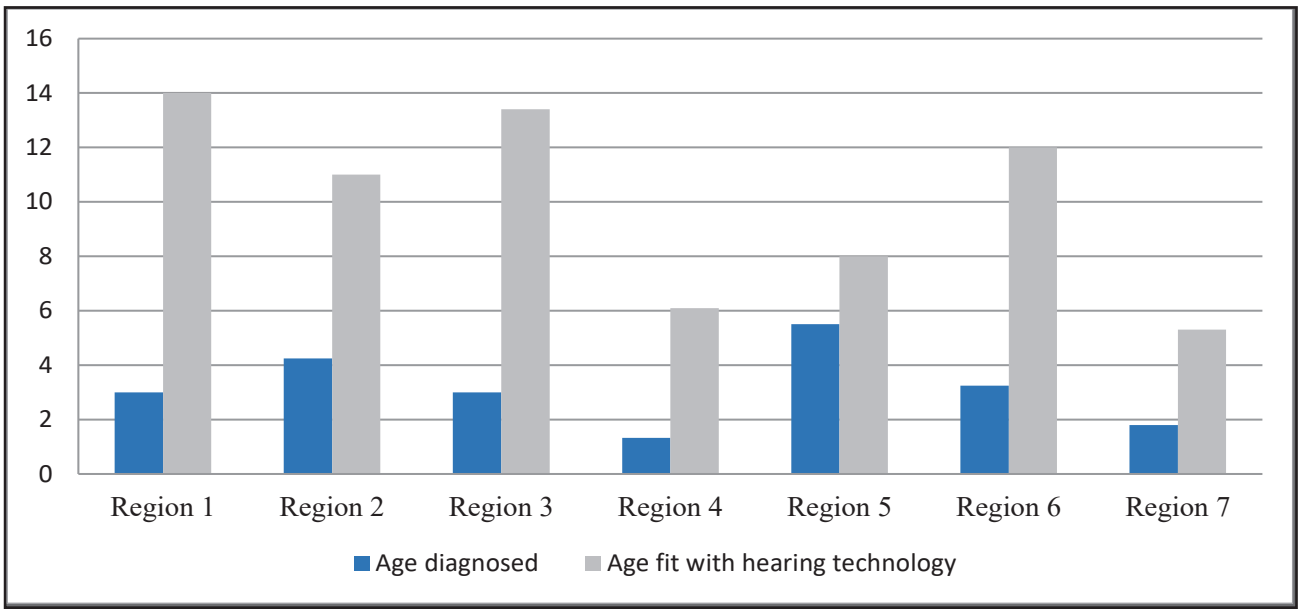
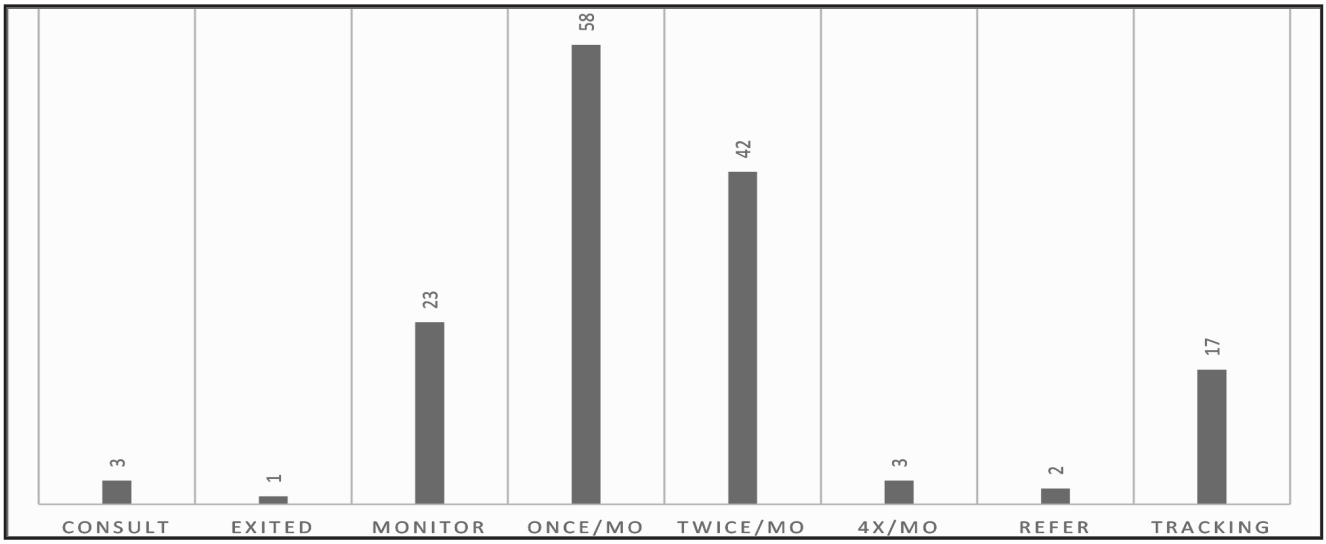


Figure 4

Amount of Early Intervention Services on Individual Family Service Plan Per Month based on Statewide Collaborative Data Tracking



Note. *Consult* refers to providers seeing a family on regular/consistent schedule with another provider. *Exited* refers to families who have left the system. *Monitor* refers to families who want support but not on a consistent basis. Families may have other children who are deaf or hard of hearing and only want resources or the opportunity to meet occasionally for assessment and/or strategies. *Refer* means there are concerns or providers are waiting to see if they are identified with hearing loss. *Tracking* are families who do not want services. Part C providers send newsletters, invitations to community events and check-in to see if things have changed and if they are ready for service.

Use Data to Identify Needs

The ICC met as a team on a monthly basis to discuss processes, questions, and opportunities for improvement. The parent survey results (shared here) were one aspect of data collection. This was supplemented by a provider survey (Bargen et al., 2017) and ongoing discussion of statewide needs identified by the team. These data sources and discussions lead to clear opportunities for improvement. For example, by having key stakeholders coordinate a data collection effort, it was clear that there were gaps in the communication between entities. For example, 66 children were identified as needing and wanting services who were not identified by the other partner. This gap was not a result of parents' choice or

refusal of services, but gaps in the data collection and sharing. To address this need, EI administrators from ITP and IESDB now meet on a monthly basis to ensure that all families identified as having a child who is DHH have been made aware of all of the services that exist ensuring that they have not been missed by one provider or another. In addition, a protocol and training for ITP providers was developed to ensure all providers who serve children who are DHH offer IESDB participation in the Individual Family Service Plan development process.

One of the goals of this project was to develop a collaborative process that could be shared with other states. Development and implementation of the Idaho Community Collaboration was a learning process and helped us to better identify the needs and opportunities within our state. [See Brown et al. (2019) for additional benefits of public health program collaboration]. The group has since presented the development of the ICC at the national EHDI conference, the American Speech-Language and Hearing Association (ASHA) convention, and the American Academy of Audiology (AAA) conference. Perhaps, more importantly, this information has been shared with statewide providers through the ITP, IESDB, and the statewide Pediatric Audiology Conference. Through dissemination within our state, we have found other opportunities to improve our systems and engage providers to ensure that this journey is transparent and facilitated.

The ICC used the Hogue (1994), Community Based Collaborations framework to assess the level and movement of the collaboration over the two-year collaboration period (Table 2). As the group reflected on the process of the first year, there was definite movement in the relationship of the collaborators. At the beginning of the collaboration the stakeholders were somewhere between the *Networking* and *Cooperation or Alliance* levels of collaboration (Table 2). Stakeholders collaborated and communicated, tasks were completed, processes

Table 2

Community Based Collaboration

Community Linkages - Choices and Decisions			
Levels	**Purpose**	**Structure**	**Process**
Networking	* Dialog and common understanding * Clearinghouse for information * Create base of support	* Loose/flexible link * Roles loosely defined * Community action is primary link among members	* Low key leadership * Minimal decision making * Little conflict * Informal communication
Cooperation or Alliance	* Match needs and provide coordination * Limit duplication of services * Ensure tasks are done	* Central body of people as communication hub * Semi-formal links * Roles somewhat defined * Links are advisory * Group leverages/raises money	* Facilitative leaders * Complex decision making * Some conflict * Formal communications within the central group
Coordination or Partnership	* Share resources to address common issues * Merge resource base to create something new	* Central body of people consists of decision makers * Roles defined * Links formalized * Group develops new resources and joint budget	* Autonomous leadership but focus in on issue * Group decision making in central and subgroups * Communication is frequent and clear
Coalition	* Share ideas and be willing to pull resources from existing systems * Develop commitment for a minimum of three years	* All members involved in decision making * Roles and time defined * Links formal with written agreement * Group develops new resources and joint budget	* Shared leadership * Decision making formal with all members * Communication is common and prioritized
Collaboration	* Accomplish shared vision and impact benchmarks * Build interdependent system to address issues and opportunities	* Consensus used in shared decision making * Roles, time and evaluation formalized * Links are formal and written in work assignments	* Leadership high, trust level high, productivity high * Ideas and decisions equally shared * Highly developed communication

Note. Adapted from "Community Based Collaborations: Wellness Multiplied," by T. Hogue, 1994, Oregon Center for Community Leadership and Ohio State University.

were in place, and roles were somewhat defined. Children were identified with hearing loss and there was a general process for ensuring that the partners were aware of the child, invited to meetings, and clinical audiologists were identified. Conflicts did not exist explicitly and communication was formal (generally presented in meetings). At the end of the first year, the stakeholders had moved to a *Coordination or Partnership* or *Coalition* level of collaboration, with data points to discuss and questions about effectiveness of current processes being examined. If a child is identified with hearing loss, how do we make sure that we are all aware of the child? Do providers have the resources needed to support spoken language as a choice for parents? How do we work to ensure consistency of services across regions with less access to professionals?

Informal discussions with existing partners or relying on status quo communication does not lead to systems change or meeting the collective vision of the providers. Instead, collective work on task-oriented projects was a more effective way to engage stakeholders with productive, constructive discussion. In particular, starting the collaborative with each individual's understanding of the current system was a positive way to engage in the discussion and to identify processes that were unclear or varied from provider to provider and region to region.

Collective data collection and comparison of this data was a very effective way for partners to clearly see gaps in the system as well as discuss and identify ways to address these gaps. An advantage of coordinated data collection was that ICC partners were actively engaged in the numbers and in discussing surprises when they arose. Data provided objective ways of starting discussions and was effective in dissecting perceptions that existed.

The information gained from the parent survey, paired with the information obtained from the state early intervention systems (ITP and IESDB) provides an important first-step to defining the EHDI system in Idaho. The improved collaboration and communication between entities is important for starting objective conversations about ways to address the needs of the state. It will be important to better understand why families did not receive information about their child's hearing loss, to address if and how families are getting information about a variety of communication options such as Listening and Spoken Language, and to understand and address the reasons the majority of families are getting services twice a month or less.

In the last two years, Idaho has made significant strides in terms of its EHDI services. These changes have been, in part, because collaborative partners have had increased awareness, and in turn, more engagement in statewide efforts to support families with children who are DHH. With this engagement, collaborative efforts have included:

- In 2019, Idaho passed a rule that insurance companies cover pediatric hearing aids and 45 hours of speech-language pathology visits during the first year after the child who is DHH is fit with the amplification. A team led by IESDB facilitated a change in terminology for eligibility of services (now there is one category "Deaf/Hard-of-Hearing replacing the two categories "Deaf" or "Hearing Impaired"). Collaborative members were actively engaged in these changes and participated in statewide presentations to educational providers.
- Because of the collaborative relationship, stakeholders started conversations about the comfort level and experience of providers serving families with children who are DHH. As a result, a survey was developed for EI teams asking what basic questions existed about serving young children who are DHH. The results indicated more developmental specialists needed increased understanding of hearing loss, hearing technology, how to assist with hearing aid retention, and interpretation of audiograms. In response to this need, a website was developed to share information and resources related to these specific topics. This website was developed in partnership with the pediatric audiologist and ISU graduate students and distributed to families and providers across Idaho.
- Additionally, because of the needs identified in rural areas, ICC partners are examining the role of telepractice for collaboration and to increase intensity of services to families who live in rural/remote areas. The state has also initiated a statewide early intervention assessment process to examine child outcomes and to use these outcomes as a starting point for professional development opportunities.

Lessons Learned

Student involvement was excellent, not only from an assistance perspective, but also from the opportunity to engage future professionals in the important discussions related to the EHDI system. ISU students from the audiology and speech-language pathology programs were involved in every part of the ICC process: helping with scheduling meetings, taking minutes, data collection, entry and analysis, and development and presentation of talks at regional and national conferences. This increased students' awareness of the EHDI system, challenges, and opportunities for growth and specialization.

Monthly attendance was attainable for most of the participants. Zoom (or teleconferencing software) was very helpful for connecting all participants, particularly when administrators were traveling to satellite offices or at conferences. In hindsight, occasional in-person meetings would be recommended (even on a quarterly basis), as in-person meetings did allow for more informal communication and discussion, which led to productive outcomes.

Technology applications (such as Doodle, Zoom, and Padlet) were effective tools for communicating and scheduling between stakeholders. Some of these tools were not able to be used by all participants due to firewalls within state systems; however, these were able to be

addressed by using home email addresses when needed. Having all of the partners participate was essential. After the first ICC cycle, it was felt that the right partners were involved in the process. The ICC was fortunate to have willing and engaged partners in this collaborative effort to aid the progress or accomplishments of the project.

Conclusion

Idaho is a frontier state with many families living in rural areas. There are a limited number of pediatric audiologists and newborn hearing screening is not mandated. It is surmised that the challenges discovered during this ICC process were not unique to Idaho. Collaboration between systems is challenging for a variety of reasons. Having a grant provided an opportunity to bring together stakeholders and was a driving force to initiate the group with a specific focus of collaborative development. However, once the collaboration was established, all members of the ICC realized the importance of working together to improve the EHDI system within Idaho.

The Idaho stakeholders involved with the ICC were positive, eager to participate, and willing to reflect on their own opportunities for improvement. This was seen as a significant advantage for the state of Idaho, but may limit the generalization to other states with less willing community partners. At the conclusion of the first year, the ICC partners came together to determine the vision of the group moving forward. The collective vision was summarized as:

"In five years, Idaho will be nationally recognized as a leader in DHH education, supports, resources, and partnerships. This includes:

- An easily accessible clearinghouse of information, resources, and support for providers and families
- A cohesive team and streamlined process from screening to enrollment in early intervention through transition to Part B
- Well-established use of technology to ensure access to high quality resources and support across the state."

This shared purpose and goal helped to solidify the accomplished work and create a pathway for the next steps. Given these clear goals, Idaho is better able to leverage resources, training, and support to the families and providers in the regions that demonstrate the most need.

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Using Technology to Monitor Hearing Device Use and Linguistic Environments: Early Intervention Providers' Perspectives

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Abstract

Early intervention professionals must work with families to optimize children's hearing device use and the linguistic and auditory features of children's environments to improve outcomes for children with hearing loss. Two technologies with potential use in monitoring these domains are data logging and Language Environment Analysis (LENA) technology. This study, which surveyed early intervention providers, had two objectives: (a) to determine whether providers' experiences, perspectives, and current practices indicated there was a need for tools to better monitor these domains, and (b) to gain a better understanding of providers' experiences with and perspectives on use of the two technologies. Most providers reported that they used informal, subjective methods to monitor functioning in the two domains. The providers also felt confident that their methods showed how consistently children on their caseloads were wearing their hearing devices and what their environments were like between intervention visits. Most providers reported limited personal experience with accessing data logging information and with LENA technology. However, many providers reported receiving data logging information from children's audiologists. Providers generally believed access to the technologies could be beneficial, but only if coupled with proper funding for the technology, appropriate training, and supportive administrative policies.

Acronyms: CI = cochlear implant; DLP = digital language processor; HA = hearing aid; LENA = Language Environment Analysis; OCHL = Outcomes of Children with Hearing Loss

Keywords: cochlear implants, data logging, deaf and hard of hearing, early intervention, hearing aids, LENA technology

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Children with hearing loss are at risk for experiencing delays in spoken language development due to limitations in their ability to fully access the linguistic input in their environments (Moeller et al., 2007). Given recent improvements in early identification of children with hearing loss and in hearing assistive technologies (e.g., hearing aids [HAs] and cochlear implants [CIs]), children with hearing loss should be experiencing consistently improved outcomes. Although this has proven true for many children, the language outcomes of children with hearing loss continue to be widely variable (Geers et al., 2009; Tomblin, Walker, et al., 2015).

Recent findings from the Outcomes of Children with Hearing Loss (OCHL) study indicate that one contributor to the variance in outcomes may be variability in children's access to linguistic input. The research team developed and validated a model in which access to linguistic input was affected by children's aided audibility (access to speech with their hearing aids), duration and consistency of hearing aid use, and characteristics of the caregiver input in their environment. In turn, access to input influenced linguistic uptake and thus, language outcomes (Moeller & Tomblin, 2015). Although children's aided audibility is limited by aspects of their hearing loss and falls within the domain of the audiologist's influence, the other factors are potentially malleable within the context of early intervention. For early intervention providers to support families' efforts to establish consistent device use and optimize the child's linguistic environment, providers must be able to assess, monitor, and provide families with feedback on their progress in each domain. There are two technologies that may be particularly useful in supporting providers in completing these tasks: data logging and Language Environment Analysis (LENA) technology. In this study, we sought to understand how providers were currently monitoring children's device use and the linguistic and auditory features of their environments, including whether they were making use of these technologies. Additionally, we queried providers on their experiences with and perspectives on use of the technologies.

Consistent Hearing Device Use

The evidence tying amount of device use to outcomes is robust. Results from the OCHL study indicated that children who are hard of hearing who wore their hearing aids (HA) at least 10 hours a day were more likely to develop age-appropriate language skills than children

who wore their HAs less than 10 hours a day (Tomblin, Harrison, et al., 2015). Similarly, research indicates that for children who use CIs, quantity of device use is positively related to language outcomes (Gagnon et al., 2019; Wang et al., 2011; Wie et al., 2007).

Despite evidence regarding the positive contributions of device use to children's language outcomes, many families struggle in their efforts to establish consistent hearing device use, especially when children are young (Marnane & Ching, 2015; Muñoz et al., 2017; Walker et al., 2013). Studies using objective data logging information from children's HAs indicate that infants and toddlers aged 6 to 24 months wear their HAs an average of less than 4.5 hours per day (Walker, McCreery, et al., 2015). This differs from parent reports, which overestimated child use by an average of 2.43 hours per day. Similarly poor device use has been observed for young children who use CIs (Marnane & Ching, 2015; Wiseman & Warner-Czyz, 2018). Studies that use data logging report that the average amount of time hearing devices are used generally increases with age and degree of hearing loss (Walker et al., 2013). However, results from these objective measures show that few children reach full time device use in the first 3 years of life or in the first year after cochlear implantation (Gagnon et al., 2019; Walker, McCreery, et al., 2015; Walker, Van Voorst, et al., 2015). Potential barriers to device use include caregivers not believing in the importance of device use, situation-specific barriers (e.g., safety of wearing devices when children cannot be closely monitored in the backseat of a car), child behaviors (e.g., children removing the devices frequently), and low caregiver self-efficacy with managing the technology (Moeller et al., 2009; Muñoz et al., 2015, 2016).

Linguistic Environments: Linguistic and Auditory Features

For both children with normal hearing and children with hearing loss, the quantity and quality of linguistic input to which they are exposed during interactions with their caregivers has a strong positive relationship with later language outcomes (Ambrose et al., 2014; Ambrose et al., 2015; Hoff, 2006). However, exposure to linguist input alone does not ensure uptake by the child, especially if the child has limited auditory access to the input. Thus, to optimize the environments of young children with hearing loss, early intervention providers and families must ensure not only that children are exposed to high rates of quality linguistic input, but also that they can access that linguistic input.

Although the use of hearing devices improves children's access to linguistic information, the amount of access is often still not optimal, especially when listening in complex auditory environments (Ambrose et al., 2014). For the purpose of this study, we defined children's linguistic environments as being characterized by both the linguistic input provided by the family and the acoustic characteristics of the environment that may affect a child's ability to access linguistic input (e.g. reverberation, distance between the speaker and listener, and background noise). Auditory characteristics of the environments of infants and toddlers may be modified to improve access to linguistic input through changes in the physical environment (e.g., additions of carpeting and curtains, closing doors to other areas of the home that are noisy). Parents may also be able to use specific strategies during interactions to improve their child's access to linguistic information (e.g., gaining children's attention prior to speaking to them and being close to children when talking to them). Furthermore, the auditory characteristics of the environment can be improved through addressing sources of background noise in the home, including electronic media (e.g., turning off televisions and radios). Reducing exposure to electronic media may be one of the most accessible and impactful ways of modifying the auditory characteristics of children's linguistic environments. Not only does linguistic input become more audible to children with hearing loss, but caregivers may be able to increase and improve their interactions with their children when electronic media is not in use. Ambrose et al. (2014) found that children with hearing loss who were exposed to more electronic media had lower receptive language scores than children with hearing loss who were exposed to less electronic media. The relationship between electronic media exposure and language outcomes was mediated by the number of conversational turns between caregivers and children, indicating that parents and children had fewer successful language interactions when in the presence of electronic media.

In addition to supporting families in modifying the auditory characteristics of children's environments in ways that reduce barriers to accessing spoken language, early intervention providers must help families optimize the linguistic input they provide to their child. It is especially important for children with hearing loss to be exposed to high rates of quality linguistic input given that their inconsistent access to the input in their environments places them at risk for delays in spoken language development. Optimized input includes being engaged in frequent, high-quality conversations. Additionally, children with hearing loss learn best from interactions in which the parent adopts a responsive, as opposed to directive, interaction style and in which parents use diverse vocabulary and grammatical structures (Ambrose et al., 2015).

Assessment, Monitoring, and Feedback Technologies

For early intervention providers to support families' efforts to establish consistent device use and optimize their child's environments, providers must be able to assess, monitor, and provide families with feedback on their progress toward each goal. Little is known about how early intervention providers currently achieve these tasks.

Specifically, in this study we were interested in the use of two technologies that might support these efforts: (a) data logging in HAs and CIs, and (b) LENA technology. Use of these tools may allow early intervention providers to offer better feedback to parents about their progress

toward the goals. When given access to the information provided by these technologies, as well as coaching regarding the behavior, parents may be able to better set and make progress toward relevant goals.

Data Logging

In their efforts to determine whether children are consistently wearing their devices, both audiologists and early intervention providers frequently ask parents to estimate how many hours per day their children wear their HAs. However, evidence indicates that parents frequently overestimate their children's device use (Moeller et al., 2009; Walker, McCreery, et al., 2015; Walker, Van Voorst, et al., 2015). Data logging is a feature available in most contemporary HAs and CIs. Data logging information is accessed through each manufacturer's proprietary programming software and serves as a tracking tool for device use, including the average number of hours per day that the device was in use since the last programming session. With the advent of data logging, providers have the potential to access objective information regarding children's device use, rather than relying on the subjective information provided by parents. Audiologists have access to data logging information during programming of the devices and may share this information with families to increase awareness of how many hours the child is wearing his or her devices, help the family set and monitor progress toward goals for increased device use, or support maintenance of current use trends. At least one study has demonstrated that audiologists' use of data logging information during counseling can be effective in helping families improve device use (Muñoz et al., 2017). However, traditional counseling sessions with audiologists only occur approximately every 3 to 6 months in the first few years of a child's life. Early intervention sessions are often more frequent and place early intervention providers in a better position than audiologists to continuously monitor and support parents' efforts to establish consistent device use.

LENA Technology

The second monitoring and feedback technology is the LENA system (LENA Foundation, Boulder, Colorado). The system comprises a Digital Language Processor (DLP) and a related software program. The DLP is a digital recording device that can be worn by a child in a pocket on a specially designed piece of clothing to capture up to 16 hours of audio from the child's environment. After the recording is complete, the audio from the DLP can be transferred to the computer for analysis using the associated software. The software analyzes the audio recording to quantify information about the child's environment, including linguistic input (e.g., number of adult words and conversational turns) and presence of specific acoustic characteristics (e.g., background noise and sound from electronic media). LENA technology has been used successfully as a feedback tool with families of children with hearing loss to improve parent-child interactions in intervention studies (Sacks et al., 2014; Suskind et al., 2016).

Research Questions

Although there is evidence of the potential benefits of using data logging and LENA technology as intervention tools, it is unclear the extent to which these technologies are being used in clinical practice. It often takes many years to translate research into clinical practice, which is known as the research to practice gap. This gap is known to be higher in special education than in many other fields (Greenwood & Abbott, 2001). Currently, we know little about how early intervention providers are assessing children's device use and if they are able to access data logging information. Moreover, it is unclear if early intervention providers see utility in having increased access to data logging information for use in early intervention sessions. Similarly, we know little about how providers are monitoring the linguistic and auditory features of children's environments, whether they are using LENA technology, or if they see use of LENA technology as being potentially beneficial for families on their caseloads.

In this study, early intervention providers were surveyed regarding their practices and perspectives regarding monitoring children's device use and linguistic environments. The study posed two research questions:

1) Do early intervention providers' experiences, perspectives, and current practices indicate there is a need for tools to better monitor children's hearing device use and environments?

We queried whether providers believed families on their caseloads were already (a) optimizing children's device use and the linguistic and auditory features of their environments, (b) if providers felt confident in their ability to monitor families' progress in these domains, and (c) what tools providers were using to monitor functioning. If providers reported families were already achieving relevant goals and if providers felt confident in their ability to monitor families' functioning with tools already readily accessible to them, providers might be unlikely to see the need for data logging and LENA technology in their practice.

2) What experiences with and perspectives on use of data logging and LENA technology do early intervention providers have?

We queried whether providers had first- or second-hand experience with the technologies, what those with experience with the technologies perceived the benefits and barriers of using the technologies to be, the reasons providers had not used the technologies, and whether providers were interested in using the technologies.

Methods

Early intervention providers across the country were recruited to complete an online questionnaire. The questionnaire queried their experiences with and perspectives on monitoring the hearing device use and linguistic environments of children with hearing loss on their caseloads.

Study Procedures

The questionnaire was hosted on Qualtrics, an online survey software tool. Information about the study and a link to the questionnaire was sent directly to early intervention providers who had participated in the OCHL study and agreed to be contacted for future studies. Additionally, study information and the recruitment link were posted in several social media sites geared toward speech-language pathologists and deaf educators (e.g., the early intervention special interest group of the American Speech-Language-Hearing Association). Recruitment materials invited professionals who were currently providing early intervention services to at least one child with hearing loss to participate. Upon completion of the questionnaire, if participants wanted to be compensated for their time, they could provide a name and physical address and they were sent a $15.00 Target gift card. The survey remained open for completion from June to October 2016, when the link was closed as the total number of responses desired had been obtained. The project was approved by the Internal Review Board for Boys Town National Research Hospital.

Study Participants

A total of 163 potential participants began the online survey. Respondents were asked to confirm that they were currently serving at least one family of a child with hearing loss. Two respondents indicated that they were not, and thus were not provided with survey questions. Survey responses were also excluded from the analysis if they were not fully completed; 71 surveys were not completed and therefore excluded from the subsequent analysis. Finally, responses were excluded if participants indicated that their location or professional role was outside the scope of the purpose of the questionnaire. Two surveys were excluded for this reason (one completed by a professional from outside the United States and one completed by an individual who identified their professional role as a president of a state chapter of a parent support organization). Ultimately, 88 questionnaires were completed and included in analyses.

Participants provided early intervention services in 32 states and one U.S. territory. Of the 88 participants, 38 identified as teachers of the deaf, 34 as speech-language pathologists, five as early childhood educators, three as audiologists, and two participants did not indicate how they identified professionally. Additionally, six participants selected the "other" option. The professional identity of these participants was listed as an early childhood special educator, a dual speech-language pathologist and audiologist, a Listening and Spoken Language Specialist certified audiologist, a Listening and Spoken Language Specialist certified Auditory Verbal Educator, a dual speech-language pathology assistant and itinerate teacher of the deaf, and a teacher consultant for children who are deaf or hard of hearing. Participants had an average of 16 years of experience (range 1–50 years).

Participants were employed by a variety of agencies: 34 respondents indicated they worked for a state agency, 17 worked for a school district, 17 worked in an Option school program (a school that is a member of the international, non-profit organization designed to provide programs to educate children with hearing loss in listening and spoken language), 12 worked for private early intervention agencies, and 12 selected "other" and provided an individual response to describe their employment. There were eight settings represented in the 12 responses: hospital (n = 3), infant-toddler program provider contracted with the state (n = 1), pediatric audiology (n = 1), self-employed and private (n = 1), hospital home health (n = 1), university clinic (n = 2), pediatric rehabilitation (n = 1), and university and children's hospital (n = 2). Participants indicated that their caseload comprised between 1% and 100% children who have a hearing loss, with an average of 75%.

Questionnaire

The questionnaire included questions about (a) providers' educational preparation and current employment, (b) the hearing device use and linguistic environments of children with hearing loss on the provider's early intervention caseload, (c) providers' perceptions of the barriers families experienced in establishing consistent hearing device use and optimizing children's linguistic environments, (d) providers' experiences with or barriers to using data logging and LENA technology with families of children with hearing loss in an early intervention setting, (e) providers' opinions on the potential benefits or barriers to the use of these technologies in their current practice, and (f) other aspects of early intervention service delivery that were beyond the scope of the current manuscript. Questions used Likert scale, yes/no, multiple choice, or open-ended responses. After the questionnaire was developed, it was reviewed by research scientists and clinicians with expertise in deaf education and early intervention. The survey was piloted with current early intervention providers and the feedback was used to make changes in the wording and formatting of the questionnaire to ensure clarity of the questions.

Analysis

Each submitted survey was reviewed to confirm it was complete and not fraudulent. The results were summarized descriptively. Participants' responses to the open-ended questions were reviewed line by line and coded. For example, if a participant indicated that they had not used a technology in their practice due to the high cost of the system and the lack of training to use the device, these two components of the response were coded with two separate codes (cost and training) under the barriers to use for the technology. Once all the short answer responses were coded, the categories were reviewed and individual codes were collapsed when appropriate (e.g., codes for cost of system and lack of personal funds were combined to be represented under one code for cost). The coding process was inductive and reductive. Both authors reviewed the responses under each code to ensure that the coding system reflected the responses of all participants. Definitions for each code were developed and all responses were re-coded. Results are presented below.

Results

Research Question 1: Do Early Intervention Providers' Experiences, Perspectives, and Current Practices Indicate there is a Need for Tools to Better Monitor Children's Hearing Device Use and Environments?

Providers' Perceptions of Families' Functioning

The survey included four questions about providers' perceptions of families' functioning. Providers were asked to consider all families they had served in early intervention over the past five years, but to respond separately for families of children with HAs and families of children with CIs. Responses indicating that the provider did not serve children who used a particular hearing device or did not answer a survey question were not included in the result calculations. The results were calculated based on the number of individuals who provided answers to the question. Many providers believed that less than 20% of the HA and CI users that they have provided services to over the last five years were unable to establish full-day use in the first year after fitting. Despite reporting that the families they serve experience limited difficulty overall, providers reported that slightly more of the families of children with hearing loss they have served over the past five years had trouble establishing HA use than CI use. Providers shared that most children accepted hearing devices, with a higher percentage of providers reporting that more than 20% of families had more trouble with children accepting HAs than CIs. Providers reported that both parents of children with HAs and parents of children with CIs believed that full-day use of hearing devices was necessary for their child. Providers generally reported a higher percentage of their caseload not believing full-day HA use was important. See Table 1 for detailed results.

Table 1

Percent of Providers who Selected that Each Device Use Item was Applicable to 0-20%, 20-40%, or Greater Than 40% of Their Caseload Over the Past Five Years

	Families of Children with Cochlear Implants			Families of Children with Hearing Aids		
Item	0–20%	20–40%	> 40%	0–20%	20–40%	> 40%
Family was unable to establish full-day use in the first year after fitting	72	13	15	53	24	23
Family was unable to establish full-day device use by transition out of early intervention	77	13	10	65	21	14
Child did not consistently accept the device	84	11	5	67	21	12

Note. Eight participants indicated they did not serve children who use cochlear implants and one participant did not provide answers regarding families of children with cochlear implants for unknown reasons; therefore, for families of children with cochlear implants, percentages for the four questions were calculated based on responses from 79 participants. Additionally, one participant did not provide answers to the second and third questions for families of children with hearing aids for unknown reasons; therefore, percentages for those two questions for families of children with hearing aids were calculated based on responses from 87 participants.

Early intervention providers were asked to indicate the percent of the families on their caseload who had "room to improve" on four aspects of the linguistic environment that are positively associated with child outcomes. More providers reported substantial room for improvement on "responding to children's verbal and/or nonverbal communication attempts" than other behaviors. The fewest providers noted substantial room for improvement on "becoming less directive with their child and following their child's lead." See Table 2 for detailed results.

Providers were also asked to report the percentage of families they have served over the past 5 years who, despite the provider's counsel, continued to have their child experience one of three less-than-ideal auditory characteristics: noisy home or childcare environments, 30 minutes or more of electronic media per day, and communicating with the child without first getting close to the child. In each case, at least 50% of the providers responded that more than 40% of the children on their caseload experienced the queried characteristic. Results are displayed in Table 2.

Providers' Perceptions of Their Knowledge of Families' Functioning

Providers were asked to report their level of knowledge regarding the functioning of families on their caseloads with respect to device use and two characteristics of the environment: quantity of linguistic input and the auditory environment. Descriptive statistics are reported in Table 3. Most providers reported high confidence with regard to knowing how much each family uses their child's hearing device(s) on a daily basis, how much each family talks to and interacts with their child between visits, and what each child's auditory environment is like between visits.

Table 2

Percent of Providers who Selected that each Linguistic Environment Item was Applicable to 0–20%, 20–40%, or Greater Than 40% of Their Caseload over the Past Five Years

	Families on Caseload		
Item	0–20%	20–40%	> 40%
Family had room to improve with regard to responding to the children's verbal and/or nonverbal communication attempts	31%	27%	42%
Family had room to improve with regard to increasing and varying their language input to their children	22%	27%	51%
Family had room to improve with regard to increasing their engagement and quantity of interactions with their children	24%	26%	50%
Family had room to improve with regard to becoming less directive with their child and following their child's lead	14%	37%	49%
Family had their child spend substantial time in noisy home or childcare environments	18%	30%	52%
Family had their child view or listen to 30 minutes or more of electronic media (e.g., TV) per day	3%	23%	74%
Family attempted to communicate with their child without first getting close to the child	17%	33%	50%

Note. One participant did not answer this set of questions for unknown reasons; therefore, the percentages are calculated based on responses from 87 participants.

Table 3

Percent of Providers Indicating Each Level of Agreement Regarding Their Knowledge of the Functioning of Families on Their Caseload over the Past Five Years

	Strongly agree/ agree	Neither agree or disagree	Strongly disagree/ disagree
I know how much each family uses his/ her child's hearing device(s) on a daily basis.	83%	8%	9%
I know how much each family talks to and interacts with their child between visits.	79%	14%	7%
I know what each child's auditory environment is like between visits.	66%	23%	11%

Note. One participant did not answer this set of questions for unknown reasons; therefore, the percentages are calculated based on responses from 87 participants.

Methods Used by Providers to Monitor Functioning

Providers were asked about the current methods they used to monitor and provide feedback to parents regarding the domains of interest. Responses to the closed-set items are found in Tables 4, 5, and 6 for children's hearing device use, linguistic input in parent-child interactions, and features of children's auditory environments, respectively. Additionally, providers were asked to indicate if they used any other methods to assess these domains and, if so, to describe the method. Few alternate responses were gathered from the open-ended option. The responses regarding device use were "lack of progress in data," "comparing a child's progress to others," "asking other teachers," "daycare checks," and "judging performance." The responses regarding linguistic input in parent-child interactions were "engaging in reflection with the parent at the end of the early intervention session," "participating in role playing activities with the parent," "providing real-time or direct coaching to the parent during an interaction," and "teaching parents how to self-monitor their involvement with their children." Participants did not indicate that they used any additional methods to assess features of children's auditory environments.

Research Question 2: What Experiences with and Perspectives on Use of Data Logging and LENA Technology do Early Intervention Providers have?

Experience with the Technologies

Early intervention providers were asked about their use of data logging and LENA technology. Only 14% of the providers reported they had personal experience with

Table 4
Percent of Providers Indicating That They Never, Rarely, Sometimes*, or* Often *Used the Specified Methods to Monitor Hearing Device Use*

Technique	Never	Rarely	Sometimes	Often
Using data logging software	61%	11%	14%	14%
Asking the child's audiologist for his/her impressions	11%	11%	30%	48%
Asking the child's audiologist for results from data logging	17%	10%	38%	35%
Having the family keep a regular use log	25%	26%	34%	15%
Asking the family about the child's use[a]	2%	4%	10%	84%
Observing the child's use during sessions	2%	3%	5%	90%

[a]One participant did not answer this question for unknown reasons; therefore, the percentages are calculated based on responses from 87 participants.

Table 5
Percent of Providers Indicating That They Never, Rarely, Sometimes*, or* Often *Used the Specified Methods for Measuring and Providing Feedback on Linguistic Input in Parent-Child Interactions*

Measures	Never	Rarely	Sometimes	Often
Complete and discuss the results of a formal observational measure of parent-child interaction	49%	20%	16%	15%
Informally watch parent-child interaction and take written or mental notes to share with the family[a]	2%	5%	14%	79%
Use LENA technology	76%	13%	9%	2%
Video record the parent and child interacting for co-viewing with the parent	42%	34%	21%	3%

[a]One participant did not answer this question for unknown reasons; therefore, the percentages are calculated based on responses from 87 participants.

Table 6
Percent of Providers Indicating that They Never, Rarely, Sometimes*, or* Often *Used the Specified Methods for Measuring and Providing Feedback on Features of Children's Auditory Environments*

	Never	Rarely	Sometimes	Often
Complete and discuss the results of a formal observational measure of the child's auditory environment	64%	15%	11%	10%
Discuss my impressions of the auditory environment from my informal observations during early intervention sessions	8%	5%	23%	64%
Use LENA technology[a]	79%	9%	10%	2%
Use a sound level meter or other device to measure the noise level in the child's environment and provide results to family[b]	57%	19%	22%	2%

[a]One participant did not answer this question for unknown reasons; therefore, the percentages are calculated based on responses from 87 participants.

[b]Two participants did not answer this question for unknown reasons; therefore, the percentages are calculated based on response from 86 participants.

data logging software. However, 73% of the respondents indicated that they had received data logging information from audiologists. Only 21% of participants reported any experience with LENA technology.

Experienced Providers' Perceptions of the Benefits and Barriers to Use of the Technologies

Providers who had used either of the two technologies were asked to indicate the benefits and barriers they experienced during use. Providers with experience receiving data logging information on a first-hand or

second-hand basis listed benefits as: information to begin a discussion with the parent regarding amount of hearing device use and barriers to device use, use of the data to inform the clinician about how long hearing devices were in use and in what kinds of auditory environments, and use of the information for tracking hearing device use over time. Providers with first-hand experience with data logging technology shared several barriers to use of the technology in early intervention: "incorrect results due to improper hearing aid use," "challenging conversations between parents and providers regarding results," and "lack of correct results."

Participants with experience using LENA technology shared that they felt the information provided a platform to identify behavioral changes that could be made in the home to support language development. For example, one participant stated, "It's helpful. It is a visual way to show exactly what's going on in the home and where the parent could make improvements. It's a useful tool for helping the parent get a clear understanding of how much time needs to be devoted to achieve the target amount of daily interactions." Professionals who had used LENA technology also noted some barriers to using the technology, including parents' fear of being recorded and the need for parents to remember to put the device on their child and to turn it on daily. One participant stated, "Many parents are afraid their family interactions are being recorded and listened to by strangers, being stored on some database, and report that they cannot behave normally when the LENA is there."

Non-Experienced Providers' Reasons for Non-Use of the Technologies

Participants who reported they did not use data logging or LENA technology were asked to provide a reason. Several reasons were provided for non-use of data logging: outside of their professional responsibilities, lack of access to the technology or software, the information was obtained from another source, lack of benefit, and lack of knowledge. Reported reasons for non-use of LENA technology were lack of access, cost, lack of personal knowledge of the technology, and lack of clear benefit to current practice.

Non-Experienced Providers' Interest in Use of the Technologies

Providers who did not use the technologies were also asked if they were interested in using these technologies in their current early intervention practices. Of the 32 participants who responded about their level of interest in using data logging, 30 participants expressed interest in being able to use data logging themselves and two participants stated that they had no interest in using data logging, citing that the information was available through children's audiologists. Of the 35 participants who responded about their interest in using LENA technology in the future, 27 participants expressed interest in using the technology and eight participants expressed interest in using the technology if specific conditions were met (e.g., funding, training, increased information concerning the product).

Discussion

Recent research indicates that hearing device use and the linguistic and auditory features of children's environments contribute to the outcomes of children who are deaf or hard of hearing. However, we know little about how early intervention providers assess, monitor, and provide feedback within these areas for the families they serve, including whether they see a need for access to additional tools for monitoring families' functioning in these domains. We were specifically interested in providers' perspectives on two potential tools that could be used to objectively measure functioning in these domains: data logging and LENA technology. This study had two objectives: (a) to determine whether providers' experiences, perspectives, and current practices indicate there is a need for tools to better monitor these domains, and (b) to gain a better understanding of providers' experiences with and perspectives on use of data logging and LENA technology.

Hearing Device Use and Data Logging

The majority of providers indicated that they believed that 80% or more of the children on their caseloads were able to establish full-day CI or HA use in the first year after device fitting. This result is in stark contrast with the findings of recent research using data logging to objectively measure device use, which indicate that on average, both young children who use HAs and young children who use CIs wear their devices 5 hours a day or less (Walker, McCreery, et al., 2015; Walker, Van Voorst, et al., 2015). The primary techniques providers reported using for monitoring device use were observing use during sessions and asking parents about device use. Device use during sessions may not be representative of use between sessions. Additionally, research indicates that it is difficult for parents to estimate how much their children wear their devices, with parents having a strong tendency to overestimate use (Walker, McCreery, et al., 2015; Walker, Van Voorst, et al., 2015). Thus, the findings of the current study indicate that providers may benefit from increased access to data logging as a means of ensuring their perceptions of the device use of families on their caseload is accurate. Similarly, given how difficult it is for parents to accurately estimate device use, parents might also benefit from their early intervention providers being able to provide them with objective data on how much their child is wearing his or her devices.

Providers generally felt that data logging information had the potential to be beneficial. However, they typically reported receiving this information from audiologists, as opposed to collecting it themselves, which is likely at least in part due to the numerous barriers that exist to using data logging technology in early intervention settings. Early intervention providers' familiarity with data logging spoke to the collaborative nature of early intervention services. However, given that audiology visits only occur approximately every 3 to 6 months in the first few years of a child's life, the frequency with which this data can be attained is limited if the audiologist is the only one who can

access the data. At least one study indicates that access to data logging information between regularly scheduled audiology appointments can be useful in supporting families' efforts to increase HA use (Muñoz et al., 2016). This finding, paired with early intervention providers' interest in collaborating with audiologists to obtain data logging information, indicates that there may be benefits in increasing the accessibility of data logging information. If there was increased access to data logging information, families of children with hearing loss, early intervention providers, and audiologists might have improved capacity to collaboratively develop strategies to help children increase their hearing device use.

Linguistic and Auditory Features of Children's Environments and LENA Technology

Most early intervention providers believed that families had room to improve the linguistic and auditory features of their children's environments. However, the majority also indicated that they were relatively confident that, through use of observation and other objective measures, they were aware of how much each family talked to and interacted with their child between visits and what each child's auditory environment was like between visits. Although use of LENA technology is one potential means of gathering a more objective perspective, most providers reported limited experience with the technology. In addition to providing a tool for monitoring children's environments, LENA technology has potential to be used as a coaching tool. Indeed, in one study by Suskind et al. (2013) the authors reported that after a one-time educational intervention using LENA technology, the number of words spoken by adults in the environments of children with hearing loss increased an average of 31%. The ability for the technology to quantify key aspects of both parent-child interactions and auditory environments, including exposure to electronic media, may be especially valuable, given the interaction between these factors. For example, Ambrose et al. (2014) found that conversational turns between parents and children were less frequent in households with a high degree of electronic media usage than in households with less electronic media usage. Access to objective information regarding the amount of electronic media and conversational interactions in households may allow early intervention providers to identify families with whom having a conversation about the relationships between the auditory environment and parent-child interactions is most important and may allow parents to become more aware of their child's auditory and linguistic environment. Access to this information may also support parents in their efforts to set and monitor their progress toward goals related to media use and their interactions with their child.

Implications for Clinical Service

Many providers stated that data logging and LENA technology could improve their current practice with families. They reported that it could provide data to begin discussions with families about their barriers to behavioral changes. They also reported that data logging and LENA technology could provide families with a tool for tracking their progress toward consistent device use or optimizing the acoustic or linguistic features of their child's environment. Despite the perceived benefits, providers identified several potential barriers to their use of these technologies; such as lack of access and training, concerns regarding confidentiality and administrative policies or infrastructure, and parents' comfort. To increase use of these technologies in early intervention services, substantial effort will be needed to increase providers' access to the necessary technologies. This will include gathering more evidence on the effectiveness of these technologies, as attaining funding for technology is often dependent upon the evidence base for the technology. Additionally, providers will need support in how to think through issues of privacy, confidentiality, and access to private information. Furthermore, providers will need training in how to talk with parents about the use of these technologies.

Currently, the barriers to directly accessing data logging in early intervention are high. However, children's audiologists are able to easily access this information. Thus, administrators may want to consider methods of ensuring early intervention providers are able to easily communicate with children's audiologists to get this information. Additionally, increased communication will allow audiologists and providers to collaborate on methods for supporting the family in increasing hearing device use. Further, HA and CI manufactures should consider making this information available to parents through apps or other portals so that parents can monitor their children's device use and share this information with providers as they wish.

Conclusions

Results suggest that these monitoring and feedback technologies have the potential to improve service provision to families of children who are deaf or hard of hearing, but also suggest that they are not currently being used to their full potential in the early intervention setting. Although providers identified potential benefits to incorporating these technologies into their practices, they also identified educational, procedural, and administrative barriers to use of these technologies in early intervention services. These barriers will need to be addressed prior to widespread acceptance and integration of the technologies into early intervention services. Future transition to common use of these technologies may help bridge the research to practice gap and increase the number of effective practices documented for working with children who are deaf and hard of hearing and their families.

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Beliefs and Self-Efficacy of Parents of Young Children with Hearing Loss

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Abstract

The purpose of this study was to learn more about the beliefs and self-efficacy of parents of young children with hearing loss. Seventy-two parents completed the Scale of Parental Involvement and Self-Efficacy–Revised (SPISE-R), which queries parents about their child's hearing device use and their perceptions of their own beliefs, knowledge, confidence, and actions pertaining to supporting their child's auditory access and spoken language development. Two beliefs were identified that related to parents' action scores and one belief was identified that related to children's hearing device use. Knowledge and confidence scores were significantly correlated with action scores and children's hearing device use, whereas only confidence scores were related to scores on a measure of children's spoken language abilities. Results indicate the SPISE-R is a promising tool for use in early intervention to better understand parents' strengths and needs pertaining to supporting their young child's auditory access and spoken language development.

Acronyms: ABR = auditory brainstem response; DP-3 = Developmental Profile 3; OCHL = Outcomes of Children with Hearing Loss; PSE = parental self-efficacy; SPISE = Scale of Parental Involvement and Self-Efficacy; SPISE-R = Scale of Parental Involvement and Self-Efficacy–Revised

Keywords: cochlear implants, early intervention, hearing aids, hearing loss, parental self-efficacy

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The widespread implementation of universal newborn hearing screening has lowered the age at which children with hearing loss are identified and begin receiving intervention services (Durieux-Smith et al., 2008; Harrison et al., 2003). Younger ages at initiation of intervention services, including the fitting of hearing aids and receipt of cochlear implants, are associated with improved spoken language outcomes (Ching et al., 2013; Harrington et al., 2009; Moeller, 2000; Niparko et al., 2010; Yoshinaga-Itano et al., 1998). However, great variability in children's spoken language abilities still exists within populations of children who are fit with hearing devices and enrolled in early intervention at young ages (Geers et al., 2009; Tomblin et al., 2015).

For young children with hearing loss, spoken language outcomes are best when children have optimal auditory access through the consistent use of appropriately fitted hearing devices and are exposed to high-quality linguistic input in their environments (Ambrose et al., 2014; Ambrose et al., 2015; DesJardin & Eisenberg, 2007; Tomblin et al., 2015; Walker, Holte, et al., 2015). However, there is high variability for both these factors. For example, Walker and colleagues (2015) reported that, on average, infants in the Outcomes of Children with Hearing Loss (OCHL) study wore their hearing aids 4.36 hours per day, but device use ranged from less than 1 hour per day to almost 9 hours per day (SD = 3.17). Similarly, high variability was found in the quantity and quality of the linguistic input children in the OCHL study were exposed to, with some children engaged in fewer than 20 conversational turns an hour and others engaged in more than 100 (Ambrose et al., 2014).

Parental Self-Efficacy and Involvement

Parents can play a large role in facilitating their children's use of hearing devices and supporting their language development. Grounded in social learning theory, parental self-efficacy (PSE) describes parents' beliefs in their ability to perform a parenting task successfully (Bandura, 1977;

Wittkowski et al., 2017). PSE can also be defined as parents' estimations of their own competence in parental roles (Coleman & Karraker, 2003). Competent parents select goals, monitor their own and their child's needs and behaviors, implement strategies, and evaluate the effectiveness of their parenting behaviors (Sanders et al., 2003). PSE has been shown to be related to a wide range of parenting and child outcomes in young children with normal hearing (Albanese et al., 2019; Benedetto & Ingrassia, 2018; Jones & Prinz, 2005) and children with hearing loss (DesJardin & Eisenberg, 2007; DesJardin, 2017b; Joulaie et al., 2019). In this study, we examine PSE in parents of young children with hearing loss as it pertains to supporting their children's auditory access and spoken language development.

PSE is of special interest because it is malleable; experimental studies have indicated that interventions can successfully increase PSE (Benedetto & Ingrassia, 2018). The potential for interventions to alter PSE is important, given that PSE has been tied to parent characteristics that are, in turn, associated with child outcomes (Mouton et al., 2018). For example, parents with high PSE are more likely than parents with lower PSE to use a responsive, stimulating, and non-punitive care taking approach and to have positive maternal health (Kwok & Wong, 2000; Unger & Wandersman, 1985). In contrast, parents with lower PSE are more likely than parents with higher PSE to experience maternal depression and to report perceiving their child to be difficult to parent (Coleman & Karraker, 1997; Teti & Gelfand, 1991).

DesJardin and her colleagues were the first to examine PSE as it relates to parents of children with hearing loss (DesJardin, 2003, 2005, 2017b; DesJardin & Eisenberg, 2007). They used the Scale of Parental Involvement and Self-Efficacy (SPISE), which DesJardin designed specifically for use with families of children with hearing loss (2003). Findings from research using the SPISE have shown that parents of children with cochlear implants and parents of children with hearing aids differ significantly in terms of PSE, with parents of children with cochlear implants perceiving higher self-efficacy in the care of their children's hearing device and more involvement in developing their children's spoken language abilities than parents of children with hearing aids (DesJardin, 2005). Findings also indicate that, overall, parents report higher self-efficacy in managing their children's auditory device use than in supporting their children's language development (DesJardin, 2005; DesJardin & Eisenberg, 2007; Joulaie et al., 2019). Additionally, DesJardin (2003) found that parents' self-efficacy pertaining to supporting their child's speech and language development was positively related to the frequency with which parents reported they engaged in activities designed to support their child's speech and language development at home. In a more recent longitudinal study, parental self-efficacy and involvement in auditory device use when children were 12 months old was positively related to children's receptive language skills when children were 36 months old, whereas parent involvement in language development when children were 12 months old was positively related to children's expressive language skills when children were 36 months old (DesJardin, 2017b).

Further support for the relationship between PSE and parental use of strategies to support speech and language development was found in a study in which the research group observed mothers and their children with hearing loss play and engage in a shared book reading (DesJardin & Eisenberg, 2007). Mothers who reported high self-efficacy pertaining to supporting their children's language development were observed to provide their children with higher-level language strategies than mothers who reported lower self-efficacy, and those same higher-level techniques were positively related to children's spoken language skills. Lastly, for mothers of children who used hearing aids, but not mothers of children with cochlear implants, their perceptions of their involvement and self-efficacy pertaining to their child's hearing device use were negatively related to age at receipt of the hearing device and age at enrollment in early intervention (DesJardin, 2005).

Supporting Parents

To ensure best outcomes for children, parental involvement is critical in facilitating auditory access and supporting language development (Moeller, 2000; Sarant et al., 2009; Yanbay et al., 2014). Today, early intervention providers increasingly coach and collaborate with caregivers, using a model that seeks to build PSE, which in turn supports children's development. The coaching model is a method of family-centered practices that embraces the parents and professionals as equal members of the team, whereby parents and professionals learn from each other and work together to support the child (DesJardin, 2017a). The coaching model also focuses on strengthening families' knowledge and interactions with their children to support children's language development and should include providing parents with information they can use as part of their everyday routines (Campbell & Sawyer, 2007; Division for Early Childhood, 2014; Friedman et al., 2012).

Although the goal of the coaching model is to provide parents with the necessary skills to support their child's development, there is limited research to show if parents of children with hearing loss perceive they possess adequate knowledge and confidence to carry out the necessary tasks within their home activities. Recently, through in-depth interviews, Decker and Vallotton (2016) examined parents' reports of information received from early intervention providers about ways to promote the language development of their children with hearing loss. Findings suggested that the parents obtained some knowledge about the importance of frequent communication with their children during everyday activities. However, in this same study, parents indicated they felt the need for additional specific information about how to promote their children's language skills during daily interactions. In the recent DesJardin (2017b) study, longitudinal findings indicated that parents' perceived self-efficacy in terms of supporting both their children's auditory and language

skills was relatively high when children were 12 months of age and increased over time between when children were 12 and 36 months of age. However, during this same time period, parents' levels of involvement in supporting their children's language development decreased. The magnitude of the decrease was influenced by children's language skill level and parents' perceived guidance or support from professionals during those early years. Given that parents' sense of involvement may change over time and parents may need additional and varied support as their children's development progresses, professionals working with families of very young children with hearing loss may need better ways to recognize the kinds of support parents require throughout their years in early intervention.

Scale of Parental Involvement and Self-Efficacy–Revised (SPISE-R)

Having a better understanding of parents' beliefs, knowledge, confidence, and actions can provide professionals in early intervention with information regarding parents' areas of strengths (areas in which they are most knowledgeable and confident) and areas in which to provide additional support or guidance. To obtain a clearer view of these specific constructs, a revised version of the SPISE was developed: the Scale of Parental Involvement and Self-Efficacy–Revised or SPISE-R (Ambrose et al., 2019). The SPISE-R queries parents about their child's hearing device use and their perceptions of their own beliefs, knowledge, confidence, and actions pertaining to supporting their child's auditory access and language development. (See Appendix for the complete questionnaire.)

Research Questions

The purpose of this study was to learn more about the beliefs and self-efficacy of parents of infants and toddlers with hearing loss who wear at least one cochlear implant or hearing aid. This study addressed three research questions.

1. What does the SPISE-R tell us about parents' beliefs and self-efficacy?
2. Are demographic characteristics (i.e., parent gender, parent education level, immediate family member with a hearing loss, child age, better-ear hearing category, age at hearing loss confirmation, and type of hearing device) associated with parents' beliefs, knowledge, or confidence?
3. Are parents' perceptions of their beliefs, knowledge, or confidence related to their perceptions of their actions, children's hearing device use, or children's spoken language abilities?

Method

Data were included from two sets of participants: (a) parents who participated in a local study about their child's hearing device use and its relationship to self-efficacy and (b) parents who participated in an online survey study designed to examine the relationships between self-efficacy, hearing device use, and spoken language development. Both sets of participants completed the SPISE-R. Additionally, both sets of participants answered demographic questions. Audiologic data for children of participants in the local study, including the child's most recent audiogram or auditory brainstem response (ABR) results and other audiologic details, were retrieved from the child's medical records. Audiologic data for children of participants in the online study were collected from parents, who were asked to upload their child's latest audiogram or ABR results. Online participants also answered questions about their child's spoken language development.

Participants

Inclusion criteria required the participating adult to be the parent of a child who (a) was 36 months of age or younger, (b) wore at least one hearing aid or cochlear implant, (c) was learning spoken language, and (d) had no known conditions other than hearing loss that would affect language development. Additionally, all participants had to live in the United States. Data were available for 72 unique parents and children. Ten of the parents were participants in the local study. Sixty-two of the parents completed the online survey, with 49 of those parents submitting additional documentation regarding the child's hearing thresholds. See Tables 1 and 2 for demographic information.

Recruitment and Procedures

Local Study

Local participants were recruited by their audiologist at Boys Town National Research Hospital (Omaha, Nebraska) to participate in a longitudinal study on device use. Procedures included having parents complete a demographic questionnaire, a questionnaire about device use, and the SPISE-R at their child's first audiologic visit after enrollment in the study. Parents also consented for the research staff to access their child's medical records. At each subsequent audiologic appointment, parents completed another questionnaire about device use and, at 6-month intervals, completed the SPISE-R again and updated their demographic information. Parents were compensated for their time with a Target gift card at each visit. All 18 participants in the local study who met the inclusion criteria were invited either to complete the online study or have their existing data used in this study. Eight parents completed the online study. For the remaining 10 participants, data from the first SPISE-R they completed, along with information from the demographic questionnaire and their child's audiologic records, were included in this study.

Online Study

Online participants were recruited via a flyer that included information about the study and a link to the survey. The

Table 1
Demographic Characteristics for Responding Parents and Children's Environments

Characteristic	*n*	% group	Mean	*SD*	Range
Gender					
Mother	63	87.50			
Father	9	12.50			
Age (years)[a]			32.62	4.91	23–50
Ethnicity					
Hispanic or Latino	12	16.67			
Not Hispanic or Latino	60	83.33			
Race					
Asian	1	1.39			
Black or African American	3	4.17			
White	66	91.67			
Other	2	2.78			
Education level					
Elementary, junior high, high school, GED	7	9.72			
Some college, technical school, associate's degree	13	18.06			
Bachelor's degree	27	37.50			
One or more years of graduate education	25	34.72			
Number of children in the home			2.25	1.20	1–6
Immediate family member with hearing loss					
Yes	14	19.44			
No	58	80.56			

[a]Parent age is missing for one parent due to a discrepancy in the parent's birthdate.

flyer was posted on social media sites geared toward parents of children with hearing loss and sent to parents who participated in previous studies in the Communication Development Lab at Boys Town National Research Hospital. The flyer was also posted on social media sites geared toward professionals in the field and sent to professional contacts (e.g., early interventionists, early intervention service coordinators, and audiologists) with a request that they share it with appropriate families on their caseloads. The survey was hosted by REDCap. Participants could stop taking the survey at any time and had the ability to access a partially completed survey via a unique URL and code by selecting the "save and exit" option on the survey. The survey took approximately 30 minutes to complete.

The first portion of the survey asked participants five questions to determine if they met the inclusion criteria. If the inclusion criteria were met, the participants were presented with consent information on the following screen. If they agreed to the consent statement, they were then directed to the full survey. The survey included a demographic questionnaire, the SPISE-R, and questions from the communication subscale of the Developmental Profile 3 (DP-3; Alpern, 2007). Additionally, after all the survey questions were completed, participants were prompted to upload their child's most recent ABR report or audiogram or, if they did not have the document available to upload at that time, email the document to the lab. To increase the number of complete responses, reminder emails were sent to participants who had provided their email. Additionally, reminders were sent to participants who finished the survey, but had not uploaded or emailed their child's audiologic results. If a parent responded that they did not have access to an ABR report or audiogram, they were asked to explain their child's hearing loss in detail and given example descriptors. If the participant completed the entire survey and provided audiologic results and a mailing address, they were compensated with a $15 Target gift card.

Measures

Demographic Questionnaire

Demographic questions queried a variety of information about the responding parent, their child, and the child's environment (e.g., parent gender [i.e., mother, father], parent education levels, whether the child had any immediate family members [parents or siblings] with hearing loss, race, ethnicity, and age). For the online study, this portion also queried information specific to the child's hearing loss, including questions about the age at hearing

Table 2
Demographic Characteristics for Children

Characteristic	*n*	% group	Mean	*SD*	Range
Age (months)			21.52	9.74	4–36
Better-ear hearing threshold category					
Normal	4 (4 HA, 0 CI)	5.56			
Mild	16 (16 HA, 0 CI)	22.22			
Moderate	9 (7 HA, 2 CI)	12.50			
Moderate-severe	5 (5 HA, 0 CI)	6.94			
Severe	7 (4 HA, 3 CI)	9.72			
Profound	18 (2 HA, 16 CI)	25.00			
Device type					
Hearing aid only	48	66.76			
Cochlear implant[a]	24	33.33			
Age at confirmation of hearing loss (months)			3.56	5.00	0–25
Age at hearing aid fit (months)			5.74	5.05	1–28
Age at receipt of first cochlear implant (months)			12.88	4.12	5–25
Device use (percent of waking hours)			74.08	23.04	4–100
DP-3 Communication subscale (standard score)			97.00	21.20	50–130

Note. HA = hearing aid, CI = cochlear implant, DP-3 = Developmental Profile 3. Due to a discrepancy in reported birthdate, data is missing for one child for age, age at confirmation of hearing loss, age at hearing aid fit, and DP-3 Communication subscale score. One additional child did not have data for age at hearing aid fit due to the child not receiving a hearing aid and 10 additional children did not have data for DP-3 Communication subscale score because they were in the longitudinal study. Data are also missing for 13 children for better-ear hearing threshold category (10 HA, 3 CI) whose parents did not provide that data and one child for device use due to the parent providing incomplete data.
[a]One child used both a cochlear implant and a hearing aid and was included in the cochlear implant group for analyses of device type.

loss confirmation, age at which hearing devices were fit, and the early intervention services the child received. For the local study, this latter information was gathered from a review of the child's audiologic records. See Tables 1 and 2.

Scale of Parental Involvement and Self Efficacy–Revised (SPISE-R)

The SPISE-R (Ambrose et al., 2019) is the revised version of the SPISE (DesJardin, 2003). The SPISE-R comprises five sections. The first four use a 7-point Likert scale to query parents' beliefs, knowledge, confidence, and actions relevant to supporting their child's auditory access and spoken language development. The belief section does not yield a summary score. However, the knowledge, confidence, and action sections each yield three summary scores: average score for auditory access items, average score for language development items, and average score for the full section. In the final section, parents are asked about their child's hearing device use. See Appendix for the complete questionnaire.

Beliefs Section. The beliefs section consists of seven statements, which parents rate on a Likert scale ranging from 1 (*not at all*) to 7 (*a great deal*), with a midpoint of 4 (*somewhat*) to indicate how much they share the belief. Three items are positively-keyed, meaning that agreement is more optimal than disagreement: (a) if children are given the right supports, they can overcome the effects of hearing loss, (b) how my family talks to and interacts with my child will have a big impact on how my child develops, and (c) my child's hearing devices help him/her communicate. Four items are negatively-keyed, meaning that disagreement is more optimal than agreement: (d) no matter what we do as a family, my child's development will be delayed compared to children with normal hearing, (e) if people see my child wearing his/her hearing devices, they will judge my child or family, (f) if I keep my home too quiet, my child won't learn to listen in noise, and (g) if children wear their hearing devices all the time, they will become overly dependent on them. After parents complete the measure, the negatively-keyed items are reverse scored by recoding the responses (e.g., a 1 on the Likert scale is replaced with a score of 7 and a 7 on the Likert scale is replaced with a score of 1). The process of reverse scoring results in higher scores representing more optimal scores for all items in the section, thus allowing for comparisons between items.

Knowledge Section. The knowledge section asks parents to indicate how much they know about 10 topics: five topics related to facilitating their child's auditory access and five topics related to supporting their child's language development. The response format is a 7-point Likert scale ranging from 1 (*a little*) to 7 (*a great deal*) with a midpoint of 4 (*some*). Sample items include (a) how to manage my child's hearing devices, (b) how to share a book with my child in a way that helps him/her learn to communicate, and (c) strategies the interventionist recommends using to help my child learn to communicate.

Confidence Section. The confidence section asks parents to indicate how confident they feel in their ability to do ten tasks: five tasks related to facilitating their child's auditory access and five tasks related to supporting their child's language development. A 7-point Likert scale ranging from 1 (*not at all*) to 7 (*very*), with a midpoint of 4 (*somewhat*) is used for responses. Examples include (a) put and keep my child's hearing devices on him/her, (b) help my child hear by making changes in his/her environment, and (c) help my child learn to say new sounds, words, or sentences.

Actions Section. The instructions for the actions section ask parents to indicate how often they do fifteen tasks: seven tasks related to facilitating their child's auditory access, five tasks related to supporting their child's language development, and three tasks related to involvement in their child's intervention services. Responses were reported on a 7-point Likert scale ranging from 1 (*never*) to 7 (*always*), with a midpoint of 4 (*sometimes*), thus higher scores are more optimal. Examples from this section are (a) draw my child's attention to sounds in speech or the environment that he/she is still learning or might not have heard, (b) make sure other people caring for my child know how to help my child learn to communicate, and (c) advocate for my child's needs in intervention sessions and IFSP/IEP [Individualized Family Service Plan/Individualized Education Program] meetings.

Device Use Section. The device use section includes questions related to the child's use of his or her hearing devices. Questions query how much the child wears his or her hearing devices while sleeping, how many hours a day the child is awake, and how many hours the child wears his or her hearing devices in total. Although data logging information was not collected for this project, the first question is standardly included in the SPISE-R for situations in which a comparison is being made to objective data logging stored in the hearing devices, as data logging will capture time the devices were turned on while children are sleeping. The responses are used to calculate the percent of the day the child wears his or her hearing devices while awake, which is the value used in this study. The section also asks parents to use a scale ranging from *never* to *always* to report how often their child wears their hearing devices in different environments.

Developmental Profile 3 (DP-3)

The DP-3 is a general development screener with physical, adaptive behavior, social-emotional, cognitive, and communication scales, the latter of which was used for this study. It was developed for ages birth to 12 years, 11 months (Alpern, 2007). It was not included in the methods for the longitudinal study, thus was only completed by parents in the online study. The communication scale asks whether the child has completed 29 language milestones ranging from "does your child usually look toward the source of a sound when it starts, such as a person beginning to talk?" to "does your child write or print from memory at least 20 words with correct spellings?" Responses were transferred to hard copies of the parent/caregiver checklist. Raw scores were used to calculate age-normed standard scores with a normative mean of 100 and standard deviation of 15.

Statistical Analysis

Prior to conducting the analyses for the three research questions, we examined whether the three sections of the SPISE-R that were designed to yield summary scores (knowledge, confidence, and actions) had sufficient internal consistency. Cronbach's alpha indicated acceptable levels of reliability with $\alpha = 0.89$, 0.92, and 0.92 for the three sections, respectively (DeVellis, 2003). Most items resulted in a decrease in the alpha if deleted, which indicated they should be retained in the measure. For the small number of items that would result in an increase if deleted, the change would be minimal (in all cases, less than 0.01).

Our first research question queried what parents' responses on the SPISE-R tell us about their beliefs and self-efficacy. To answer this question, the data were summarized descriptively. In addition, paired-samples *t*-tests were used to examine whether there were differences between average scores for the knowledge and confidence sections and, within each section, whether there were differences between average scores for the auditory access and language development subsections. Additionally, Pearson correlations were calculated between the knowledge total mean score and the confidence total mean score.

Our second research question queried whether demographic characteristics (i.e., parent gender [mother, father], parent education level, immediate family member with a hearing loss [yes, no], child age, better-ear hearing category [normal, mild, moderate, severe, profound], age at hearing loss confirmation, and type of hearing device [hearing aid, cochlear implant]) were associated with parents' beliefs, knowledge, or confidence. For device type, the one child who used both a cochlear implant and a hearing aid was represented as a cochlear implant user. To address this question, we first examined relationships between the seven demographic characteristics. Device type was significantly related to better-ear hearing category ($r_s = 0.73$, $p < .01$), reflecting that the children with cochlear implants had more hearing loss than the children with hearing aids. Device type was also significantly related to child age ($r_{pb} = 0.27$, $p = .02$), reflecting that the children with cochlear implants were older than the children with hearing aids. Given these relationships and the limited

variability in better-ear hearing category for the children with cochlear implants, better-ear hearing category was only examined for the hearing aid users and child age was examined separately for hearing aid and cochlear implant users. No other demographic characteristics were significantly related to one another. Because the belief data were not designed to be summarized, analyses were conducted separately for each belief. Spearman correlations were used for the belief analyses, due to violations in the assumptions for parametric analyses. The knowledge and confidence sections were each represented by the respective section score. Pearson product moment correlations were calculated to examine the relationships between the section scores and the child's current age. Point-biserial correlations were calculated to examine the relationships between the section scores and the three binomial variables: parent gender, immediate family member with a hearing loss, and device type. Lastly, Spearman correlations were calculated to examine the relationships between the section scores and the three variables that did not meet the assumptions for use of parametric analyses (parent education level, better-ear hearing category, and age at hearing loss confirmation). Data were missing for one child with a hearing aid for the two age related variables due to a discrepancy in reported birthdate and for 10 children with hearing aids for better-ear hearing category due to parents not submitting audiologic information.

The third research question queried whether parents' perceptions of their beliefs, knowledge, or confidence were related to their perceived actions, children's hearing device use, or children's spoken language abilities. Spearman correlations were calculated to examine the relationship of each belief with action scores, language scores, and device use. Pearson product moment correlations were calculated to determine whether knowledge and confidence scores were associated with action scores and language scores. Spearman rank order correlations were calculated to determine whether knowledge scores and confidence scores were associated with device use, due to the device use variable violating the assumption for parametric tests. Data was missing for one child for hearing device use (due to incomplete data) and 11 children for language abilities (10 children in the longitudinal study and one child for whom a discrepancy in the reported birthdate made it impossible to calculate a standard score on the DP-3). For the one child who wore both a hearing aid and a cochlear implant, the parent reported identical wear time for the two devices.

Results

Parents' Beliefs and Self-Efficacy

The first research question examined what the SPISE-R results indicated about the beliefs and self-efficacy of parents of infants and toddlers with hearing loss. The beliefs section consisted of seven items querying how strongly a parent agrees with the belief. See Table 3 for individual item data. After reverse scoring the four negatively-keyed items, low scores represent less desirable levels of agreement. For all seven items, the average scores were above the mid-point of four on the scale (range of 4.76 to 6.49). Although average scores for each belief were generally high, there was a wide range in parent responses, with five of the seven beliefs having scores ranging from 1–7 and the two remaining beliefs having scores ranging from 2–7.

The knowledge and confidence sections each consisted of 10 items: five related to auditory access and five related

Table 3
Agreement Level for Belief Items

Belief	Agreement Level		
	M	*SD*	Range
1. If children are given the right supports, they can overcome the effects of hearing loss.	5.83	1.52	1–7
2. How my family talks to and interacts with my child will have a big impact on how my child develops.	6.49	0.95	2–7
3. No matter what we do as a family, my child's development will be delayed compared to children with normal hearing.[a]	5.53	1.51	1–7
4. My child's hearing devices help him/her learn to communicate.	5.89	1.62	1–7
5. If people see my child wearing his/her hearing device(s), they will judge my child or family.[a]	4.76	1.66	1–7
6. If I keep my home too quiet, my child won't learn to listen in noise.[a]	4.97	1.66	1–7
7. If children wear their hearing device(s) all the time, they will become overly dependent on them.[a]	6.21	1.21	2–7

[a]Reverse scoring rules applied.

Table 4
Descriptive Data for the Knowledge and Confidence Sections and the Corresponding Subsections

	Knowledge			Confidence		
Score	M	SD	Range	M	SD	Range
Auditory access subsection score	5.31	1.07	3.40–7	5.29	1.12	3.00–7
Language development subsection score	5.30	1.26	2.40–7	5.50	1.19	2.00–7
Total section score	5.30	1.08	3.00–7	5.40	1.08	2.50–7

to language development. See Table 4 for descriptive data for each section and subsection. Average scores were relatively high for both knowledge and confidence (M = 5.30, SD = 1.08 and M = 5.40, SD = 1.08, respectively), with no significant difference between average scores for the two sections (t = -1.33, p = .19). For the knowledge section, there was not a significant difference between average scores for the auditory access items and average scores for the language development items (auditory access M = 5.31, SD = 1.07 and language development M = 5.30, SD = 1.26; t = 0.78, p = .94). However, for the confidence section, average scores for the auditory access items were significantly lower than average scores for the language development items (auditory access M = 5.29, SD = 1.12 and language development M = 5.50, SD = 1.19; t = -2.23, p = .03). On both the knowledge and confidence scales, the item with the lowest score was the item pertaining to the Ling 6-Sound test (knowledge item #5 M = 4.75, SD = 2.21; confidence item #5 M = 4.42, SD = 2.17). Of the twenty total items on the knowledge and confidence scales, only one item had an average score above 6 (knowledge item #1 M = 6.21, SD = 0.83): "how to manage my child's hearing device(s)."

The average knowledge and confidence scores were strongly correlated (r = 0.85, p < .01) indicating that parents who self-reported being highly knowledgeable were also likely to self-report being highly confident and vice versa. However, despite the strong correlation between knowledge and confidence for the full group, inspection of the individual data for each item indicated that some parents reported large differences between their perceived knowledge and confidence scores for individual skills, with differences as high as five points.

Associations with Demographic Characteristics

The second research question examined whether demographic characteristics (i.e., parent gender, parent education level, child having an immediate family member with a hearing loss, child age, better-ear hearing category, age at hearing loss confirmation, and type of hearing device) were associated with parents' beliefs, knowledge, and confidence.

Beliefs

Spearman correlations indicated that scores for the belief that "how my family talks to and interacts with my child will have a big impact on how my child develops" were significantly correlated with parent gender (r_s = -0.37, p < .01), indicating that mothers agreed more strongly with this statement than fathers. Scores for this belief were also negatively correlated with age at hearing loss confirmation (r_s = -0.27, p = .03), indicating that the earlier a child was identified with hearing loss, the more likely their parent was to strongly agree with this statement. Additionally, after reverse scoring, scores for the belief that "no matter what we do as a family, my child's development will be delayed compared to children with normal hearing" were significantly correlated with parent gender (r_s = -0.24, p = .04), indicating that fathers agreed more strongly with this statement than mothers. Scores for this belief were also negatively correlated with better-ear hearing category for hearing aid users (r_s = -0.52, p = < .01) and age at hearing loss confirmation (r_s = -0.24, p = .04), indicating that the later a child was identified with hearing loss and/or the greater the child's hearing loss, the more likely the parent was to strongly agree with this statement. Finally, after reverse scoring, scores for the belief that "if children wear their hearing device(s) all the time, they will become overly dependent on them" were significantly correlated with parent gender (r_s = -0.33, p < .01) and the child having an immediate family member with hearing loss (r_s = -0.24, p < .04) indicating that fathers agreed more strongly with this statement than mothers and parents whose children had no immediate family members with a hearing loss agreed more strongly with this statement than parents whose children did have an immediate family member with hearing loss. No significant relationships were identified between the remaining beliefs and demographic characteristics (all ps > .05).

Knowledge and Confidence

Knowledge scores were significantly related to hearing device type (r_{pb} = 0.30, p = .01), with parents of children with cochlear implants reporting higher knowledge scores than parents of children with hearing aids. Confidence scores were significantly related to parent gender (r_{pb} = -0.30, p = .01), indicating mothers reported higher levels of confidence than fathers. No other significant relationships were identified between the demographic characteristics and knowledge or confidence (all ps > .05).

Relationships with Parents' Perceived Actions, Children's Hearing Device Use, and Children's Spoken Language Abilities

The third research question queried whether parents' perceptions of their beliefs, knowledge, or confidence were related to their perceived actions, children's hearing device use, or children's spoken language abilities. The belief that "how my family talks to and interacts with my

child will have a big impact on how my child develops" was significantly related to action scores ($r_s = 0.40$, $p < .01$). The belief that "my child's hearing devices help him/her learn to communicate" was significantly related to both action scores ($r_s = 0.34$, $p < .01$) and hearing device use ($r_s = 0.33$, $p < .01$). No other belief scores were significantly correlated with action scores, hearing device use, or language scores (all ps > .05).

Both knowledge and confidence scores were significantly correlated with action scores (knowledge $r = 0.64$, $p < .01$; confidence $r = 0.69$, $p < .01$) and hearing device use (knowledge $r_s = 0.33$, $p < .01$; confidence $r_s = 0.25$, $p = .04$). Confidence scores were also significantly related to language scores ($r = 0.34$, $p = .01$), whereas knowledge scores were not ($r = 0.23$, $p = .08$).

Discussion

Recommended practices for early intervention include an intervention model that seeks to enhance the family's ability to meet the unique needs of their child (American Speech-Language-Hearing Association, 2008; Division for Early Childhood, 2014). For families of children with hearing loss who are learning spoken language through audition, this typically entails ensuring families have the skills necessary to support their child's auditory access and language development within daily activities and routines in their home (DesJardin, 2017a). However, we know little about whether families participating in early intervention hold beliefs and self-efficacy levels that are likely to facilitate their ability to carry out actions that facilitate their children's auditory access and spoken language development. To fully support families, it is imperative not only to monitor children's hearing device use and language development, but also to assess parents' perceptions of their beliefs, knowledge, confidence, and actions, all of which can affect how they facilitate their child's auditory access and language skills. When early intervention professionals obtain parents' perceptions of these constructs, professionals can identify parents' strengths and areas in which they may need additional support and guidance.

Results of this study indicate the SPISE-R has promise for use in early intervention to better understand parents' strengths and needs. The knowledge, confidence, and actions sections all had high levels of internal consistency, with item analyses indicating no items should be excluded. The items within each section were created to ensure professionals using the measure could collect meaningful information about the most relevant aspects of parents' roles in supporting their child's auditory access and language development. Although, on average, parents reported desirable agreement levels with the beliefs, as well as relatively high levels of knowledge and confidence, there was individual variability, with some families having several beliefs with undesirable agreement levels and low levels of knowledge or confidence for multiple items. Thus, the tool may be helpful in identifying families who need additional educational counseling pertaining to their beliefs or additional support to feel knowledgeable and confident enough to carry out actions that will facilitate their child's auditory access and language development.

The tool may also be useful in identifying parents with gaps between their perceived knowledge and confidence levels. Overall, parents' knowledge and confidence scores were strongly correlated, indicating that parents who self-report being highly knowledgeable are also likely to self-report being highly confident. However, knowledge does not always translate to confidence, as some parents reported gaps between their perceived knowledge and confidence levels for individual skills. For example, a parent may indicate a high score in knowledge relating to strategies for keeping the child's hearing devices on, but a low score in his or her confidence in their ability to do so. When professionals note such gaps, it may be an indication that parents need more support to practice a skill, as suggested in an early intervention coaching model.

Beliefs

Results from this study indicate that parents may vary in terms of their beliefs about children's hearing device use and language development. Of the seven belief items, the two with the lowest scores were (a) that others judge the child or family when they see the child's hearing devices and (b) that if the child's home is too quiet, the child won't learn to listen in noise. Neither belief is concerning if it does not affect parents' behavior. In the former case, the concern would arise if a parent's belief that their family will be judged when others see the hearing device results in their having the child use the device less frequently in public settings. In the latter case, if the belief leads to parents not reducing background noise in their home, it may put the child at higher risk for spoken language delays than children whose parents attempt to provide them with an optimal listening environment (Erickson & Newman, 2017).

Findings regarding the relationships between the beliefs and demographic characteristics indicate that both the later a child was identified with hearing loss and the greater the hearing loss a child with hearing aids had, the more likely their parent was to strongly agree with the idea that their child's development would inevitably be delayed. Fathers also expressed a stronger agreement with this belief than mothers. If parents believe that their child's language development will be delayed regardless of their own efforts, parents may have little incentive to take actions that could positively impact their child's learning, including providing their child with high rates of quality linguistic input to further support their child's spoken language skills.

Agreement with the belief that how the family talks to and interacts with the child will impact the child's development was negatively related to the age at which the child's hearing loss was confirmed and was weaker for fathers than mothers. Similar to findings in the DesJardin 2017b study, it could be that the earlier children are identified with hearing loss, the more time they spend in early intervention where the importance of high-quality interactions with their child is continuously emphasized. Additionally, multiple studies point to generally lower

involvement of fathers in early intervention services (Erbasi et al., 2018; Ingber & Most, 2018), which if true for fathers in this study, could be a source of the differences between mothers' and fathers' beliefs.

Parents whose children did not have an immediate family member with hearing loss expressed stronger agreement than parents of children who had an immediate family member with hearing loss with the belief that children can become too dependent on their devices if they wear them all the time. Fathers also expressed a stronger agreement with this belief than mothers. The difference between mothers and fathers may be tied to potential differences in participation in early intervention services. The differences between parents whose children did and did not have an immediate family member with hearing loss might indicate that more extensive experience with hearing loss helps parents understand the benefits and lack of negative consequences presented by consistent hearing aid use.

There were two beliefs that were significantly associated with parents' perceived actions. First, scores for the belief regarding the potential positive impact of how the family talks to and interacts with the child were positively related to action scores. Thus, although it was uncommon for parents to disagree with this belief, when observed, it may warrant further educational counseling by early intervention providers. Additionally, scores for the belief that their child's hearing devices help their child learn to communicate were positively related to action scores and children's hearing device use. These relationships indicate the importance of families believing in the benefits of hearing device use, a belief that can be targeted through a variety of strategies, including simulations of the child's hearing loss (Ambrose et al., 2020). Although agreement with the remaining beliefs was not significantly related to actions, hearing device use, or spoken language scores, when providers find that parents hold a belief, they should monitor how that belief affects how the parent supports their child's auditory access and language development on a case-by-case basis.

Self-Efficacy

Parents generally reported high levels of knowledge and confidence pertaining to supporting their child's auditory access and language development. However, variability across parents and between skills was high, indicating these are important constructs to measure and monitor. Similar to prior research (DesJardin, 2005), parents of children with cochlear implants reported higher knowledge scores than parents of children with hearing aids. Fathers reported lower levels of confidence than mothers. Contrary to prior research using the SPISE (DesJardin, 2005; DesJardin & Eisenberg, 2007; Joulaie et al., 2019), confidence scores were slightly lower for supporting children's auditory access as compared to language development, indicating that families with young children may benefit from coaching strategies on topics pertaining to use of hearing devices and creating an optimal listening environment. In particular, given the relatively low levels of knowledge and confidence parents reported for the items pertaining to the Ling 6-Sound test, families may especially benefit from coaching pertaining to conducting the test, which is a valuable tool for monitoring children's auditory access with their hearing devices (Ling, 1976).

Unlike prior research (DesJardin, 2005), knowledge and confidence were not associated with demographic characteristics, with the exception of parents of children with cochlear implants reporting significantly higher knowledge levels (but not confidence levels) than parents of children with hearing aids and fathers reporting lower confidence levels than mothers. The relationship of device type with knowledge levels may be a result of children with cochlear implants often receiving more intensive intervention services than children with hearing aids. However, if the differences in intervention lead to increased knowledge levels, but not confidence levels, the intervention efforts may need to be reexamined to ensure the efforts influence children's outcomes. The differences in confidence between mothers and fathers may be related to possible differences in the involvement of mothers versus fathers in early intervention.

Results also indicated that both perceived knowledge and confidence levels were positively associated with self-reported action levels and hearing device use. Additionally, perceived confidence levels were associated with children's spoken language scores. This aligns with findings from studies using the SPISE (DesJardin & Eisenberg, 2007; Stika et al., 2015), as well as more general findings indicating that levels of PSE are related to a range of parenting and child outcomes (Benedetto & Ingrassia, 2018; DesJardin, 2017b; Jones & Prinz, 2005; Joulaie et al., 2019). This finding also indicates that early intervention professionals should seek to boost parents' PSE as an intervention strategy that may ultimately affect children's outcomes.

Limitations and Future Research

Although the results of this study are promising, additional research is needed to further establish the validity of the SPISE-R, including the construct and content validity of the measure. Future investigations should also examine the predictive validity of the SPISE-R, as it is possible that beliefs and self-efficacy have bi-directional relationships with outcomes. One weakness of the current work was that the data were highly reliant on parent report, not only of parents' perceptions of their own knowledge and confidence, but also their actions, their child's device use, and their child's language skills. Although self-reports of how people perceive their knowledge and confidence may be reasonably valid, self-reports of action behaviors may have lower validity (Wittkowski et al., 2017) and parents are known to typically overestimate how much their children use their hearing devices (Walker, McCreery, et al., 2015). Future studies should use objective outcome measures when possible and, when not, also collect information on providers' perceptions of relevant outcomes. Additionally, early intervention characteristics (e.g., frequency, provider type, proportion of intervention time spent coaching the parent, etc.) need to be explored to investigate the variability in beliefs and self-efficacy.

Lastly, given that prior studies of families with children with typical hearing found PSE to be related to gender, socioeconomic status, and cultural variability (Coleman & Karraker, 2003; Dumka et al., 1996), it will be important for future studies investigating beliefs, PSE, and involvement of families of young children with hearing loss to include participants who are more culturally diverse, as well as more fathers as the sample of parents in this study were primarily mothers, Caucasian, and of relatively high socio-economic status.

Summary

The revised SPISE-R is a promising tool for use in early intervention to better understand parents' beliefs and their areas of strength and needs pertaining to supporting their young child's auditory access and spoken language development. Early intervention professionals should ensure their intervention services use a coaching model that helps parents understand their potential to influence their child's outcomes, builds PSE, and supports parents' involvement in facilitating their child's development. Additionally, professionals should monitor how parents' beliefs and PSE change over time and how beliefs and PSE may relate to how parents are involved in their young children's early intervention.

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Well-being of Parents of Children Who Are Deaf or Hard of Hearing

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Abstract

Purpose: The purpose of this study was to explore factors influencing the well-being of parents who have children who are deaf or hard of hearing (DHH) and to compare their experiences to non-clinical samples.

Method: A cross-sectional online survey was used to collect data ($N = 296$).

Results: Data analyses revealed the majority of parents of children who are DHH were functioning similarly to or better than the non-clinical samples in our comparison and within the non-clinical range for the included measures. No relationship was found between factors related to child age or timing of services (age at diagnosis, time between diagnosis and amplification fitting, age fit with hearing technology, child's current age) and parent psychosocial functioning.

Conclusions: Although most parents are likely to be functioning well, knowing when a parent is experiencing challenges has important implications for clinical practice, including supporting parents in finding solutions when sub-optimal daily intervention practices are occurring. Audiologists can incorporate strategies to identify parents that may be experiencing challenges into their routine practice.

Keywords: counseling, pediatrics, psychosocial impact

Acronyms: DASS-21 = Depression, Anxiety, and Stress Scale; DHH = deaf or hard of hearing; GSES = Generalized Self-Efficacy Scale; MHC-SF = Mental Health Continuum Short Form; PCC = person-centered care; SF-36 = RAND 36-Item Health Survey; WSAS = Work and Social Adjustment Scale

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Hearing loss affects 34 million children worldwide (World Health Organization [WHO], 2018). In the United States, two to three out of every 1,000 children are born with permanent hearing loss (Centers for Disease Control and Prevention, 2018a). Parents are central to the intervention process and instrumental in supporting language development; however, parents can experience challenges incorporating intervention tasks (e.g., hearing aid care and use) for a variety of reasons, that can change over time. For example, initially many parents are unprepared for the news when their child is identified with hearing loss as most parents of children who are deaf or hard of hearing (DHH) have normal hearing (Mitchell & Karchmer, 2004). Furthermore, life variables can be unpredictable, interfering with parent engagement and how effectively they are able to manage intervention tasks. Understanding parental well-being can help audiologists consider the support parents may need as they implement person-centered care (PCC) within their clinical encounters with families. Well-being (emotions and functioning) is a concept that encompasses physical and mental health and provides insights into perceptions on how people feel their lives are going (CDC, 2018b). When people have higher levels of well-being, they are better able to manage typical daily routines (Healthy People, 2020).

When children are identified with hearing loss, the demands of intervention represent a new layer in the daily lives of families and consideration of how parents are managing hearing care is an important part of the intervention process. The concept of family quality of life is used to discuss the degree to which the family members' needs are met as well as the extent to which family members enjoy their time together and are able to do things that are important to them (Poston et al., 2003). Research has highlighted the need to support parents of children who are DHH related to their emotional well-being and intervention management challenges (Hintermair, 2006; Lederberg & Golbach, 2002; Most & Zaidman-Zait, 2003; Muñoz et al., 2015; Jean et al., 2018). For example, parents of young children who are DHH have reported significantly higher levels of context-specific stress (e.g., language development, hearing devices, child behavior) compared to parents of children with typical hearing (Quittner et al., 2010). Studies have also found young

children inconsistently wear their hearing aids (Jones & Launer, 2010; Muñoz et al., 2014; Walker et al., 2013), which hinders spoken language development (Tomblin et al., 2015).

Parents are required to change their behaviors to add new elements to their daily routines to provide effective day-to-day hearing care management. Audiologists have an important role in helping parents adjust and gain new skills, and how audiologists communicate with parents is a critical consideration. For example, in a meta-analysis patient adherence was found to be highly correlated to physician communication (Zolnierek & DiMatteo, 2009). Communication plays an important role in behavior change and adherence in the treatment of chronic pediatric health conditions (DiMatteo, 2004). In addition to how audiologists communicate, they need to understand challenges parents are experiencing that may interfere with effective hearing care management, as having this information allows audiologists to better support parents. Therefore, the purpose of this study was to explore factors influencing the well-being of parents who have children who are DHH, and to compare their experiences to non-clinical samples.

Method

Participants and Procedures

This study met ethical approval by the Utah State University Institutional Review Board. Parents of children who are DHH were recruited to participate via flyers posted on social media, on parent organization websites, and in pediatric audiology facilities across the United States. Participants were eligible to participate if they were proficient in English and a parent of a child with hearing loss. Participants completed an online survey in Qualtrics from June to August 2018. As an incentive, participants were eligible to enter a drawing for one of ten $50 Amazon gift cards by providing their contact information in a separate window after completion of the study, ensuring anonymity of survey responses.

The study was designed to reach participants broadly, therefore, it is not possible to calculate a response rate. Three hundred and eighteen survey submissions were started, and 296 were subsequently analyzed for demographic data. Responses from 22 participants were dropped entirely, as they appeared to have been opened by participants; however, no items in these 22 surveys were completed. Responses to individual survey questions were not forced, thus leaving a variable amount of responses for each item. For participant demographic information see Table 1. The majority of respondents were mothers (94%; 277/296), were White (83%; 248/296), had a college degree (75%; 222/296), and reported an annual income of more than $81,000 (58%; 172/295).

Instruments

Demographic Questionnaire

The demographic questionnaire included 10 items related to the child, six items related to the parent, and two items on the impact of the hearing loss. The first question on the impact of hearing loss asked participants to rate how their child was currently doing as a result of the treatment they have received/are receiving for their hearing loss on a seven-point scale of improvement/decline from *much improved* to *very much worse*, along with an option for *my child does not receive treatment for hearing loss*. The second question asked participants to indicate, in a *Yes*/*No* format, the areas that they or their child have received help in 11 categories (i.e., Friends/Social, Relationship/Family, Marriage/Intimate Relationship, Parenting, Financial, Academic/Education, Communication Confidence, Self-identity/Stigma, Recreation, Self Care, Bullying).

Depression, Anxiety, and Stress Scale (DASS-21)

The DASS-21 is a 21-item self-report questionnaire measuring psychological distress. It includes three subscales for depression, anxiety, and stress. Items are scored from 0 (*never*) to 3 (*always*) with higher scores indicating more distress. The questions for this measure are time-bound to the past week and include a four-point scale (i.e., *did not apply to me at all, applied to me some degree, applied to me a considerable degree, applied to me very much*). An example question is "I was intolerant of anything that kept me from getting on with what I was doing." The scale has high total reliability (Cronbach's α = .88), high item reliability for depression (Cronbach's α = .82), anxiety (Cronbach's α = .90) and stress (Cronbach's α = .93), and has adequate construct validity (Henry & Crawford, 2005). Internal consistency for the DASS-21 in the current study was good for depression (Cronbach's α =.89), anxiety (Cronbach's α =.83), and stress (Cronbach's α =.89).

The Generalized Self-Efficacy Scale (GSES)

The GSES is a 10-item self-report questionnaire measuring an individual's perception of his or her ability to respond to new or challenging situations. The questions for this measure include a four-point scale (i.e., *not at all true*; *hardly true*; *moderately true*; *exactly true*). An example question is "I can always manage to solve difficult problems if I try hard enough." The measure has a maximum score of 40, with a higher score indicating more self-efficacy. The scale has high internal consistencies reported, ranging from Cronbach's α = .82–.93 (Schwarzer & Jerusalem, 1995). Internal consistency for the GSES in the current study was good (Cronbach's α =.88).

The RAND 36-Item Health Survey (SF-36)

The SF-36 is a 36-item scale that measures individual functioning based on eight elements which include: (a) physical functioning (Cronbach's α = .93), (b) role limitations due to physical health (Cronbach's α = .84), (c) role limitations due to emotional health (Cronbach's α = .83), (d) energy and fatigue (Cronbach's α = .86), (e) emotional well-being (Cronbach's α = .90), (f) social functioning (Cronbach's α = .85), (g) pain (Cronbach's α = .78), and (h) general health (Cronbach's α = .78). A higher score overall and in each subscale defines a more favorable health state. The SF-36 has been used to

Table 1
Participant Demographic Information

Demographic Variables	% (n)	*M* (SD)	Median	Range
Parent				
Race (*N* = 296)				
White	83 (248)			
Latino/a	4 (14)			
Asian	4 (11)			
Black/African American	3 (9)			
Multiracial	3 (8)			
Other	2 (5)			
Native/Indigenous	1 (1)			
Age (*N* = 296)		39 (8)	38	45
Education Level (*N* = 296)				
Graduate degree	34 (101)			
College education	41 (121)			
Partial college	15 (44)			
High school diploma/GED	7 (20)			
Less than high school	3 (10)			
Annual Income (*N* = 295)				
More than $81,000	58 (172)			
$41–80,000	26 (78)			
$21–40,000	10 (28)			
Less than $20,000	6 (17)			
Relation to Child (*N* = 296)				
Mother	94 (277)			
Father	5 (14)			
Other caregiver	1 (5)			
Child				
Race (*N* = 288)				
White	80 (230)			
Multiracial	8 (24)			
Latino/a	5 (14)			
Asian	3 (9)			
Black/African American	2 (6)			
Other	2 (5)			
Current Age in years (*N* = 292)		7 (6)	6	30
Age Identified in months (*N* = 286)		20 (30)	3	168
Degree of Hearing Loss (*N* = 296)				
Mild-moderate	25 (74)			
Severe-profound	74 (219)			
Unsure	1 (3)			
Unilateral or Bilateral (*N* = 296)				
Unilateral	22 (64)			
Bilateral	78 (232)			
Age fit with technology in months (*N* = 239)		26 (31)	15	168
Technology Type (*N* = 296)				
Hearing aid (HA)	43 (127)			
Cochlear implant (CI)	32 (96)			
Bimodal (HA+CI)	8 (24)			
Other (did not write in response)	8 (24)			
Bone conduction hearing aid	5 (15)			
FM system only	2 (5)			
Does not use technology	2 (5)			
Parent-reported hours of device use (*N* = 169)		12 (3.5)	12	23
Other comorbidities (*N* = 296)	32 (95)			
Primary mode of communication (*N* = 286)				
Spoken language	87 (250)			
Sign language	13 (36)			
Language spoken in the home (*N* = 288)				
English only	85 (244)			
English plus another language	14 (40)			
Other	1 (4)			

measure functioning in a variety of individuals representing a wide range of health conditions. The questions for this measure are time-bound and have varying scales (e.g., *limited a lot*, *limited a little*, *not limited at all*). Example questions include "Does your health now limit you in climbing several flights of stairs?" (physical functioning), "During the past 4 weeks, have you had any of the following problems with your work or other regular daily activities as a result of your physical health—accomplished less than you would like?" (role limitations due to physical functioning), and "During the past 4 weeks, have you had any of the following problems with your work or other regular daily activities as a result of any emotional problems—cut down the amount of time you spent on work or other activities?" (role of emotional health). The scale has been validated to accurately distinguish impacts of health conditions on physical and mental health (Hays & Sherbourne, 1993; Hays & Stewart, 1990). Internal consistency for the SF-36 in the current study ranged from acceptable to excellent: (a) physical functioning (Cronbach's α = .93), (b) role limitations due to physical health (Cronbach's α = .90), (c) role limitations due to emotional health (Cronbach's α = .85), (d) energy and fatigue (Cronbach's α = .77), (e) emotional well-being (Cronbach's α = .83), (f) social functioning (Cronbach's α = .86), (g) pain (Cronbach's α = .85), and (h) general health (Cronbach's α = .82).

Mental Health Continuum Short Form (MHC-SF)

The MHC-SF is a 14-item self-report questionnaire measuring facets of emotional, psychological, and social well-being. It measures the frequency which respondents experience symptoms of positive mental health, providing clear standards for assessment and categorization of three levels of mental health (flourishing, languishing, and moderately mentally healthy). The questions for this measure are time-bound to the past month and include a six-point scale (i.e., *never, once or twice, about once a week, about 2 or 3 times a week, almost every day, every day*). An example question is "During the past month, how often did you feel good at managing the responsibilities of your daily life?" Total scores can range from 0–70 with a higher score indicating a higher level of emotional well-being. The MHC-SF has demonstrated excellent internal consistency (> .80) and validity (Cronbach's α = .88; Keyes et al., 2008; Westerhof & Keyes, 2009). Internal consistency for the MHC-SF in the current study was excellent (Cronbach's α = .91).

The Work and Social Adjustment Scale (WSAS)

The WSAS is a 5-item self-report questionnaire that assesses the impact of a person's psychological difficulties on functioning in terms of work, home management, social leisure, private leisure, and personal/family relationships. It allows for comparisons of functional impairment across studies and disorders and was modified in this study by placing the carrier phrase "Because of my child's hearing loss…" at the start of each item. The questions for this measure include an eight-point scale (e.g., *not at all impaired* to *very severely impaired*). An example question is "Because of my child's hearing loss, my ability to work is impaired." Scoring is continuous up to a maximum score of 40. The higher the score, the more an individual sees their disability or disorder as an impairment to functioning. The scale has high internal consistencies reported (Cronbach's α = .70–.94; Mundt et al., 2002). Internal consistency for the WSAS in the current study was excellent (Cronbach's α = .90).

Analyses

The IBM Statistical Package SPSS v25 was used for data analyses (IBM SPSS Statistics for Macintosh, Version 25.0). Prior to analyses, data were checked for normality using measures of skewness and kurtosis (absolute values that fall within 1 suggest normality). Central tendency (i.e., means, medians) and variability were calculated to provide sample descriptives. One sample *t*-tests (for continuous independent variables) were used to compare the present sample to non-clinical score samples, defined as individuals who do not require psychological intervention based on normed scale scores, drawn from previous research studies. In addition, effect sizes (Cohen's *d*) were calculated to provide an estimate of the magnitude of between-group differences. Regression analyses were conducted to examine the relationship among age of diagnosis, time between diagnosis, age fit with technology, and all outcomes of interest.

Results

Parents rated how their children have responded to the intervention they have received for hearing loss on a seven-point scale of improvement/decline (i.e., *very much improved, much improved, minimally improved, unchanged, minimally worse, much worse, very much worse*). Parent responses ($N = 296$) indicated 73% reported *very much* or *much improved* (see Figure 1), less than 1% ($n = 1$) reported *much worse*, and 5% ($n = 14$) reported their child had never received treatment for their hearing loss.

Parents also indicated types of support from a list of 11 services they have sought for themselves and/or their child (see Figure 2). Almost half (49%; 144/296) indicated they have attended a hearing loss support group. Over half of the respondents reported seeking two types of support services—Academic/Educational (63%; 186/296) and Social/Friends (52%; 155/296).

Outcomes of Interest

The scores for parents of children who are DHH were compared to non-clinical samples. The results of the comparisons are described below and can be found in Table 2.

Psychological Distress (DASS-21)

Compared to a non-clinical sample (Henry & Crawford, 2005), the current sample did not report higher levels of distress. The majority of the present sample fell within the normal range for clinical cut-offs (Depression: 77%, 188/243; Anxiety: 80%, 195/244; Stress: 77%, 185/241);

20 to 23% of parents reported experiencing depression, anxiety, and/or stress ranging from mild to extremely severe. All questions in each subscale required completion to obtain accurate scores. Scores and participant breakdowns can be found in Table 3.

Sense of Self-Efficacy (GSE)

There was a statistically significant difference between our sample and the non-clinical sample ($p < .0001$; $d = .94$; Schwarzer & Jerusalem, 1995). Parents of children who are DHH self-reported a greater sense of self-efficacy (belief that they have an innate ability to achieve goals) than the non-clinical sample. The authors of the scale recommended a dichotomous split for scoring, using the median as a cut-off point. Therefore, our sample was categorized into scores of 0–29 (moderate self-efficacy) and 30–40 (high self-efficacy). Eighty-nine percent (231/261) of the current sample reported high self-efficacy. Results of this measure can be found in Table 3.

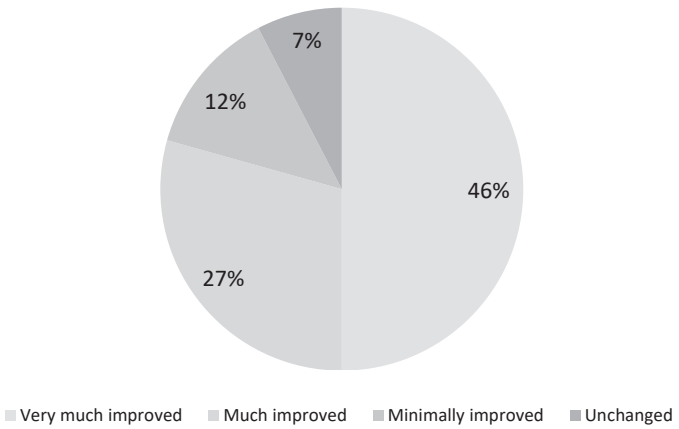
Quality of Life (SF-36)

Parents in our sample had statistically significantly better scores (see Table 2) than the non-clinical sample for measurements of physical functioning ($p \leq .0001$), the role limitations due to physical functioning ($p \leq .0001$), the role of emotional health ($p \leq .001$), pain ($p \leq .0001$), and general health ($p \leq .0001$). Some participants fell below the mean (see Table 4), in particular in the area of energy/fatigue, (21% 1–2 *SD* and 7% > 2 *SD*) and emotional health (8% 1–2 *SD* and 15% >2 *SD*).

Overall Well-Being (MHC-SF)

The majority of participants fell into the *flourishing* category (66%; 167/254) meaning they frequently (i.e., *every day* or *almost every day*) experience symptoms of positive mental health. Thirty-three percent (84/254) fell into the *moderate* group (categorized as neither *languishing* or *flourishing*) and 1% (3/254) were in the *languishing* group (i.e., *never* or *once or twice* during the past month have experienced positive mental health). Participant results can be found in Table 3.

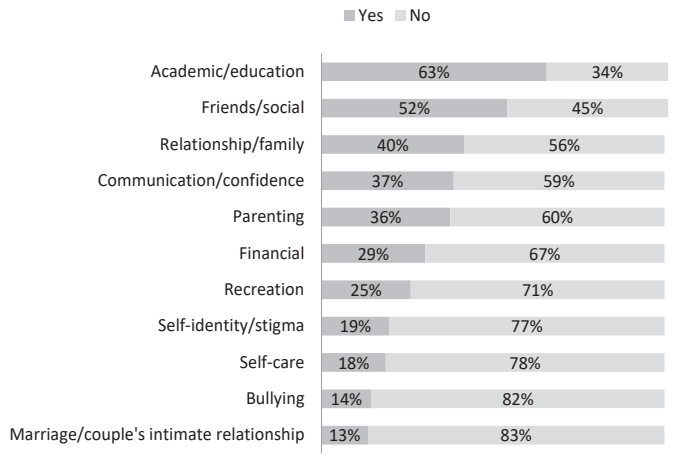
Figure 1
Parent Perceived Response to Intervention (N=296)



Functional Impairment (WSAS)

The majority of the current sample (70%; 171/246) reported subclinical scores (< 10 points) meaning they do not perceive their child's hearing loss as impeding their ability to work or socially interact with others in meaningful ways. Scores above 20 suggest moderately severe or worse psychopathology, scores between 10 and 20 have been associated with significant functional impairment but less severe clinical symptomology, and scores below 10 reveal typical functioning (Mundt et al., 2002). Participant responses can be found in Table 3.

Figure 2
Areas of Support Sought (N = 296)



Regression Analysis

Preliminary regression analyses were completed to see if there was any relationship between degree of psychosocial functioning and age of diagnosis, time between diagnosis and amplification fitting, age fit with technology, and current age. There were no significant relationships between predictors tested (i.e., age of diagnosis, time between diagnosis, age fit with technology, current age) and outcomes (e.g., psychological distress, sense of self-efficacy, quality of life, overall well-being, functional impairment).

Discussion

The purpose of this study was to explore factors related to the well-being of parents who have children who are DHH, and to compare their experiences to non-clinical samples. The majority of parents in this study were functioning similarly to or better than the non-clinical samples in our comparison. Furthermore, there was no relationship between factors related to child age or timing of services (age at diagnosis, time between diagnosis and amplification fitting, age fit with hearing technology, child's current age) and parent psychosocial functioning. The finding that parents reported positive indicators for well-being is encouraging and may be influenced by multiple factors, such as the type of support and services they are receiving. Recruitment for our study included social media and parent support organizations, and this may

Table 2
Group Comparison of Means on Outcomes of Interest

	Non-clinical Sample	Study Sample		
	X (*SD*)	X̄ (*SD*)	*p*	*d*
DASS-21 (*N* = 245)				
Total score	9.43 (9.66)	10.02 (9.67)	.37	
Anxiety (*N* = 244)		2.05 (3.07)		
Depression (*N* = 243)		2.76 (3.44)		
Stress (*N* = 241)		5.26 (4.3)		
GSES (*N* = 261)				
Total score	29.46 (5.33)	33.9 (4.07)	< .0001	.94
SF-36				
Physical functioning (*n* = 230)	70.61 (27.42)	88.35 (20.19)	< .0001	.74
Role limitations due to physical functioning (*n* = 232)	52.97 (40.78)	85.35 (30.61)	< .0001	.90
Role of emotional health (*n* = 232)	65.78 (40.71)	75.00 (37.45)	< .001	.24
Energy fatigue (*n* = 230)	52.15 (22.39)	49.98 (20.07)	.1564	
Emotional well-being (*n* = 230)	70.38 (21.97)	73.23 (18.05)	.0565	
Social functioning (*n* = 228)	78.77 (25.43)	81.30 (24.60)	.1496	
Pain (*n* = 232)	70.77 (25.46)	79.25 (21.75)	< .0001	.36
General health (n = 230)	56.99 (21.11)	69.54 (19.97)	< .0001	.61
MHC-SF (*N* = 254)				
Total score	3.98 (.85)	3.74 (.83)	< .0001	.29
WSAS (*N* = 246)				
Total score	10.8 (8.8)	6.89 (8.62)	< .0001	.45

Note. Normed Sample Populations differ per test. Depression, Anxiety and Stress Scale (DASS-21): 1,794 (Henry & Crawford, 2005); Generalized Self-efficacy Scale (GSES): 17,553 (Schwarzer & Jerusalem, 1995); RAND 36-Item Health Survey (SF-36): 2,471 (Hays & Sherbourne, 1993); Mental Health Continuum Short Form (MHC-SF): 1,662 (Lamers et al., 2011); Work and Social Adjustment Scale (WSAS): 365 (Mundt et al., 2002).
DASS-21: Higher score indicates more distress. GSES: Higher score indicates more self-efficacy. SF-36: Higher score indicates more favorable health state. MHC-SF: Higher score indicates a higher level of emotional well-being. WSAS: Higher score indicates more impairment to functioning.

have influenced the number of participants connected and supported by other parents. Parents have reported that an important source of support and information is other parents of children who are DHH (Jackson, 2011).

Although most parents are likely to be functioning well, knowing when a parent is experiencing challenges has important implications for clinical practice, including supporting parents in finding solutions when sub-optimal daily intervention practices are occurring (e.g., low hours of hearing aid use). It is important to keep in mind sample characteristics when interpreting comparisons to a non-clinical sample (e.g., non-clinical samples are obtained at a different time). The analysis does not represent a true comparison as our study had different population characteristics given the design of our study (e.g., cross-sectional design and measures not normed for a population related to hearing disorders), and caution should be taken to guard against over-interpretation.

Although our study looked at psychological functioning overall, our findings corroborate other research. For example, Dyson (1996) stated that families of children with learning disabilities are similar to families of normally achieving children in that they have a positive and cohesive family relationship and use rules for operating the family routine, despite experiencing higher levels of parenting stress in relation to their child's learning disability. Furthermore, Hayes & Watson (2013) found parents of children with autism spectrum disorder experience higher parenting stress than parents of typically developing children; however, research also shows positive parental characteristics and early intervention may reduce the impact that stress has on the family. These findings, in addition to research related to parents of children who are DHH (Hintermair, 2006; Jean et al., 2018; Quittner et al., 2010), reveal parents of children with chronic conditions may experience more challenges related to that particular condition. However, research

Table 3

Clinical Cut-off Statistics

	% (*n*)
DASS-21 (*N* = 245)	
Depression (*N* = 243)	
Normal (0–9)	77 (188)
Mild (10–13)	11 (26)
Moderate (14–20)	8 (19)
Severe (21–27)	2 (4)
Extremely severe (28+)	2 (6)
Anxiety (*N* = 244)	
Normal (0–7)	80 (195)
Mild (8–9)	5 (12)
Moderate (10–14)	8 (20)
Severe (15–19)	4 (9)
Extremely severe (20+)	3 (8)
Stress (*N* = 241)	
Normal (0–14)	77 (185)
Mild (15–18)	8 (20)
Moderate (19–25)	8 (18)
Severe (26–33)	6 (15)
Extremely severe (34+)	1 (3)
GSES (*N* = 261) *Dichotomous Split	
Moderate self-efficacy (0–29)	11 (30)
High self-efficacy (30–40)	89 (231)
MHC-SF (*N* = 254)	
Flourishing	66 (167)
Moderately mentally healthy	33 (84)
Languishing	1 (3)
WSAS (*N* = 246)	
Normal (< 10)	70 (171)
Significant functional impact (10–20)	20 (49)
Moderately severe psychopathology (> 20)	11 (26)

Note. DASS-21 = Depression, Anxiety and Stress Scale; GSES = Generalized Self-efficacy Scale; SF-36 = RAND 36-Item Health Survey; MHC-SF = Mental Health Continuum Short Form; WSAS = Work and Social Adjustment Scale.

*The author of this measure does not endorse clinical cut-offs but does state that a median split/dichotomous split can be used to show how many fall above or below a median score of 30.

also shows intervention helps reduce the level of negative psychosocial impact on the family unit.

Clinical Implications

The majority of parents in our study had a high level of well-being, underscoring the importance for audiologists to explore multiple life variables (e.g., other caregiver involvement, child factors) when challenges related to treatment adherence arise. When audiologists create a safe space to comprehensively understand parent concerns and respond to parent emotions, they are better able to determine underlying challenges. Furthermore, talking with parents about their struggles and their emotions is therapeutic and may reduce the power of negative emotions, opening the parent up to exploring solutions to problematic behaviors (e.g., not putting on their child's hearing aids).

Parents often will not initiate sharing their emotions. Having a prompt, such as use of a mental health screening tool from a caring professional, can be a welcome opportunity (Muñoz et al., 2017), and parents have reported it can help with recall, validating their concerns, reframing issues that may not have been seen as relevant, and in raising new questions (Fothergill et al., 2013). Additionally, Fothergill reported physicians felt that the screening tool helped open the conversation to sensitive issues while providing more comprehensive care. If significant emotional challenges are identified, for example on a screening tool such as the DASS-21, referral to a mental health professional can be facilitated.

Limitations and Future Research

The study was conducted exclusively online and that may have deterred responses from parents less comfortable with this format (e.g., several people opened the survey but did not complete it). The majority of our sample consisted of White mothers with a college education. This is not reflective of the multicultural population that makes up the United States. Additionally, the majority of parents reported their children had a severe-profound degree of hearing loss. The demographic composition of our sample is not inclusive of the heterogeneity of parents of children who are deaf or hard of hearing. Early Hearing Detection and Intervention programs have found that more than 50% of infants identified with hearing loss have a mild bilateral loss or a unilateral loss (White, 2018). Furthermore, the results of our study reflect parent perceptions at a single point in time; it is not possible to know the relationship between variables or the causes. Life variables change and can influence parent well-being in an unpredictable manner.

Further research is needed to explore experiences of a more diverse sample of parents, parents of younger children, as well as parents with children who have mild to moderate and unilateral hearing loss. Research is also needed to understand factors that may predict parents who are more likely to experience challenges, as well as supports that can mitigate problems to improve hearing management and child outcomes.

Table 4

Rand 36-Item Health Survey (SF-36) Analyses of Distribution

Scale	> 2 *SD*s below mean % (*n*)	1–2 *SD*s below mean % (*n*)	0–1 *SD*s below mean % (*n*)	0–1 *SD*s above mean % (*n*)	1–2 *SD* above mean % (*n*)	> 2 *SD*s above mean % (*n*)
Physical functioning (*n* = 230)	7 (16)	5 (12)	12 (27)	76 (175)		
Role limitations due to physical functioning (*n* = 232)	9 (20)	6 (13)	10 (24)	75 (175)		
Role of emotional health (*n* = 232)	15 (35)	8 (19)	13 (31)	64 (147)		
Energy/fatigue (*n* = 230)	7 (16)	21 (49)	27 (60)	31 (72)	12 (28)	2 (5)
Emotional well-being (*n* = 230)	6 (14)	11 (25)	26 (61)	39 (89)	18 (41)	
Social functioning (*n* = 228)	6 (14)	9 (20)	24 (54)	61 (140)		
Pain (*n* = 232)	4 (10)	13 (31)	31 (71)	24 (56)	28 (64)	
General health (*n* = 230)	6 (14)	8 (19)	24 (55)	46 (106)	16 (36)	

Conclusion

This study sampled parents of children who are deaf or hard of hearing to explore how they were doing in various domains related to their well-being. The majority of parents in this study were functioning similarly to or better than the non-clinical samples in our comparison. Although most parents are likely to be functioning well, knowing when a parent is experiencing challenges has important implications for clinical practice, including supporting parents in finding solutions when sub-optimal daily intervention practices are occurring. Audiologists can incorporate strategies to identify parents that may be experiencing challenges in their routine practice.

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Appendix

SCALE OF PARENTAL INVOLVEMENT AND SELF-EFFICACY-REVISED (SPISE-R)

Directions: Circle a number to answer each question. The phrase "hearing devices" is used to refer to both hearing aids and cochlear implants. "Parents" is used to refer to children's main caregivers.

A. BELIEFS: These items describe things that some parents of children with hearing loss may believe or be concerned about. Please indicate how much YOU share these beliefs or concerns.

		Not at all			Somewhat			A great deal
1.	"If children are given the right supports, they can overcome the effects of hearing loss."	1	2	3	4	5	6	7
2.	"How my family talks to and interacts with my child will have a big impact on how my child develops."	1	2	3	4	5	6	7
3.	"No matter what we do as a family, my child's development will be delayed compared to children with normal hearing."	1	2	3	4	5	6	7
4.	"My child's hearing device(s) help him/her learn to communicate."	1	2	3	4	5	6	7
5.	"If people see my child wearing his/her hearing device(s), they will judge my child or family."	1	2	3	4	5	6	7
6.	"If I keep my home too quiet, my child won't learn to listen in noise."	1	2	3	4	5	6	7
7.	"If children wear their hearing device(s) all the time, they will become overly dependent on them."	1	2	3	4	5	6	7

B. KNOWLEDGE: Parents must learn a lot of new information and skills when their child has a hearing loss. This process takes time. We are interested in how much you currently **know** about each topic.

		Not at all			Some			A great deal
1.	How to manage my child's hearing device(s)	1	2	3	4	5	6	7
2.	Strageties to use to keep my child's hearing device(s) on him/her	1	2	3	4	5	6	7
3.	What my child can and cannot hear without his/her hearing device(s)	1	2	3	4	5	6	7
4.	What my child can and cannot hear with his/her hearing device(s)	1	2	3	4	5	6	7
5.	How to do the Ling 6-Sound test (ah, ee, oo, m, sh, s)	1	2	3	4	5	6	7
6.	The sounds, words, or sentence types my child should be learning to say	1	2	3	4	5	6	7
7.	How to help my child learn to communicate	1	2	3	4	5	6	7
8.	How my child's learning is affected by his/her hearing loss	1	2	3	4	5	6	7
9.	How to share a book with my child in a way that helps him/her learn to communicate	1	2	3	4	5	6	7
10.	Strategies the interventionist recommends using to help my child learn to communicate	1	2	3	4	5	6	7

C. CONFIDENCE: Knowledge alone doesn't always make us confident or comfortable doing something. We may need more time or practice to build confidence. Please indicate how **confident** you are in your ability to do each thing.

	Not at all			Somewhat			Very
1. Determine if my child's hearing device(s) are working okay	1	2	3	4	5	6	7
2. Put and keep my child's hearing device(s) on him/her	1	2	3	4	5	6	7
3. Help my child hear by making changes in his/her environment	1	2	3	4	5	6	7
4. Help my child hear and understand new speech sounds or sounds in his/her environment	1	2	3	4	5	6	7
5. Find out if my child is hearing okay by using the Ling 6-Sound test (ah, ee, oo, m, sh, s)	1	2	3	4	5	6	7
6. Help my child learn to say new sounds, words, or sentences	1	2	3	4	5	6	7
7. Help my child communicate what he/she wants and needs	1	2	3	4	5	6	7
8. Communicate with my child in a way that is appropriate to address his/her hearing needs	1	2	3	4	5	6	7
9. Share books with my child in a way that helps him/her learn to communicate	1	2	3	4	5	6	7
10. Do the things I learned during intervention sessions when the professional is not there to help me	1	2	3	4	5	6	7

D. ACTIONS: We know daily lives are busy. There are many responsibilities that parents have. It is not possible to always do everything we would like to do each day. Given other responsibilities, we are interested in how often you are able to **do** the following things.

	Never			Sometimes			Always
1. Daily listening checks on my child's hearing device(s)	1	2	3	4	5	6	7
2. Make sure other people caring for my child know how to manage my child's hearing device(s)	1	2	3	4	5	6	7
3. Make sure I, or someone else, puts my child's hearing device(s) on immediately after he/she wakes up	1	2	3	4	5	6	7
4. Make sure I, or someone else, puts my child's hearing device(s) on immediately if they fall off or my child takes them off	1	2	3	4	5	6	7
5. Make sure my child's environment makes it as easy as possible for him/her to hear	1	2	3	4	5	6	7
6. Draw my child's attention to sounds in speech or the environment that he/she is still learning or might not have heard	1	2	3	4	5	6	7
7. Daily check of my child's listening with the Ling 6-Sound test (ah, ee, oo, m, sh, s)	1	2	3	4	5	6	7
8. Use strategies during our daily activities to help my child learn to say new sounds, words, or sentences	1	2	3	4	5	6	7
9. Use strategies to help my child communicate his/her wants and needs	1	2	3	4	5	6	7
10. Make sure other people caring for my child know how to help my child learn to communicate	1	2	3	4	5	6	7
11. Share books with my child at least one time a day	1	2	3	4	5	6	7
12. Use the strategies I learned during intervention sessions to help my child learn to communicate.	1	2	3	4	5	6	7
13. Advocate for my child's needs in intervention sessions and IFSP/IEP	1	2	3	4	5	6	7

14. Get my child to the audiologist as soon as a visit is needed	1	2	3	4	5	6	7
15. Attend and be involved in my child's intervention sessions (instead of having to do other things during that time, such as prepare meals or take care of siblings)	1	2	3	4	5	6	7

E. DEVICE USE: We are intereseted in how much your child wears his/her hearing device(s) when he/she is awake on an average day. If your child has one hearing aid and one cochlear implant and there are differences in how you would answer the questions for each device, please answer separately for each device. (In the table, please use "CI" and "HA" if needed.)

1. How many hours a day is your child usually awake? _____
2. How many hours a day does your child usually wear his/hear hearing device(s) while awake? _____
3. If your child ever wears his/her hearing devices (turned on) while sleeping, please indicate the average number of hours per day this occurs._____
4. How often does your child usually wear his/her hearing device(s) when he/she is awake in these situations?

	Never	Rarely	Sometimes	Often	Always	Doesn't Apply to us
a) At home						
b) In the car						
c) In daycare or school						
d) When cared for by family or friends outside the home						
e) Playing outside						
f) On outings (e.g., store, zoo, children's museum)						

Note. Please cite instrument as: Ambrose, S. E., Appenzeller, M., & DesJardin, J. L. (2019). Scale of Parental Involvement and Self-Efficacy – Revised [Assessment Instrument]. Boys Town National Research Hospital.

JEHDI
The Journal of Early Hearing Detection and Intervention
NCHAM National Center for Hearing Assessment and Management Utah State University
2020; 5(1): 98-138

Others' Publications about EHDI: December 2019 through April 2020

The *Journal of Early Hearing Detection and Intervention (JEHDI)* focuses on improving Early Hearing Detection and Intervention (EHDI) systems by publishing peer-reviewed articles describing current research, evidence-based practice, and standards of care that are relevant for newborn and early childhood hearing screening, diagnosis, support, early intervention, the medical home, information management, financing, and quality improvement.

Even though *JEHDI* is the only journal that focuses exclusively on improving EHDI systems, many other journals include some articles relevant to *JEHDI's* aim. To help *JEHDI* readers stay up-to-date about information in other journals about improving EHDI programs, we provide titles and abstracts of recent publications that are relevant to EHDI systems. Articles are listed in alphabetical order by the last name of the first author and titles of all articles are hyperlinked to the source.

The EHDI-relevant articles described in the following abstracts are from all over the world – demonstrating the global relevance of EHDI systems. Many of the following abstracts focus on screening and diagnosis, suggesting that the fundamentals of the EHDI system still need to be improved. Many other articles are breaking new ground and suggesting creative innovations. For example:

- DeForte et al. described an app called Hear Me Read (HMR) which uses enhanced digital stories as therapy tools for speech, language, and literacy for children with hearing loss. The study evaluated the user experience of the HMR app through a focus group study with caregivers and their children. The findings suggest that such educational apps can be valuable for those with hearing loss who are pursuing listening and spoken language as a communication outcome.
- Diener et al. surveyed 365 caregivers whose children were being seen in an otolaryngology clinic at a tertiary pediatric hospital about their knowledge of and attitudes toward congenital Cytomegalovirus (cCMV) and cCMV screening. Caregivers frequently were unaware of cCMV and its implications. Attitudes toward cCMV screening generally were positive. A majority wanted to know if their child had cCMV even if asymptomatic and were willing to pay $20 for cCMV screening. The results suggested that education on epidemiology and impact of cCMV may benefit both prevention of infection and attitudes toward screening.
- Guo et al. evaluated the efficacy of concurrent hearing and genetic screening in a general 239,636 eligible infants. They found 548 infants with hearing loss based on the physiological hearing screening, 41 infants who passed the hearing screening but likely had hearing loss based on the genetic screen, and 570 infants at risk for ototoxicity which is undetectable by hearing screening. They concluded that genetic screening complements newborn hearing screening by improving the detection of infants at risk of hereditary hearing loss and ototoxicity, and by informing genotype-based clinical management for affected infants and their family members.
- Kruyt et al. evaluated the efficacy of Bone-Anchored Hearing implants (BAHIs) in children based on 20 articles published between 2000 and 2017, encompassing 952 children with implants. They concluded that BAHIs are a safe method for hearing rehabilitation in children, although large differences between studies are observed. The outcomes of new surgical techniques and implant designs in the pediatric population seem promising, but more research is needed before definitive conclusions can be drawn.
- Le et al. examined the health related quality of life (HRQoL) in young children with low language or congenital hearing loss. Based on a sample of 108 children in Australia, they found that children with low language and with hearing loss had lower HRQoL than children with normal language; the worst HRQoL was experienced by children with both. They concluded that children with low language and congenital hearing loss might benefit from interventions targeting overall health and well-being, not just their impairments.
- Rabiço-Costa et al. in a study conducted in Portugal assessed the incidence of hearing loss in 51 children after the exposure to platinum drugs used to treat central nervous system tumors. They found ototoxicity in 23.5% of the children. Even though the use of chemotherapy for such tumors has significantly improved cure and survival rates, the ototoxicity resulting from platinum-derived chemotherapy may accompany patients for the rest of their lives (see related article by van As et al.).

- Tarhun examined the smoking habits of family members of 75 children with serious otitis media (SOM) and 50 healthy controls. The correlation between SOM and passive smoke was statistically significantly positive. They concluded that the effect of passive smoking is a preventable and controllable risk factor in the etiology of the SOM.

Abstracts for all 100 articles are listed below.

Genes (Basel). 2020 Jan 27;11(2). pii: E132. doi: 10.3390/genes11020132.

Enhancing Genetic Medicine: Rapid and Cost-Effective Molecular Diagnosis for a *GJB2* Founder Mutation for Hearing Impairment in Ghana.

Adadey SM, Tingang Wonkam E, Twumasi Aboagye E, Quansah D, Asante-Poku A, Quaye O, Amedofu GK, Awandare GA, Wonkam A.

ABSTRACT: In Ghana, gap-junction protein β 2 (*GJB2*) variants account for about 25.9% of familial hearing impairment (HI) cases. The *GJB2*-p.Arg143Trp (NM_004004.6:c.427C>T/OMIM: 121011.0009/rs80338948) variant remains the most frequent variant associated with congenital HI in Ghana, but has not yet been investigated in clinical practice. We therefore sought to design a rapid and cost-effective test to detect this variant. We sampled 20 hearing-impaired and 10 normal hearing family members from 8 families segregating autosomal recessive non syndromic HI. In addition, a total of 111 unrelated isolated individuals with HI were selected, as well as 50 normal hearing control participants. A restriction fragment length polymorphism (RFLP) test was designed, using the restriction enzyme NciI optimized and validated with Sanger sequencing, for rapid genotyping of the common *GJB2*-p.Arg143Trp variant. All hearing-impaired participants from 7/8 families were homozygous positive for the *GJB2*-p.Arg143Trp mutation using the NciI-RFLP test, which was confirmed with Sanger sequencing. The investigation of 111 individuals with isolated non-syndromic HI that were previously Sanger sequenced found that the sensitivity of the *GJB2*-p.Arg143Trp NciI-RFLP testing was 100%. All the 50 control subjects with normal hearing were found to be negative for the variant. Although the test is extremely valuable, it is not 100% specific because it cannot differentiate between other mutations at the recognition site of the restriction enzyme. The *GJB2*-p.Arg143Trp NciI-RFLP-based diagnostic test had a high sensitivity for genotyping the most common *GJB2* pathogenic and founder variant (p.Arg143Trp) within the Ghanaian populations. We recommend the adoption and implementation of this test for hearing impairment genetic clinical investigations to complement the newborn hearing screening

Iran J Child Neurol. 2020 Winter;14(1):21-30.

Well-Being and Coping Capacities of Adolescent Students with Hearing Loss in Mainstream Schools.

Adibsereshki N, Hatamizadeh N, Sajedi F, Kazemnejad A.

OBJECTIVES: Coping strategies used by adolescents has an important role in preventing or decreasing their stresses and also increasing their well-beings. This study aimed at evaluating the coping capacity and well-being of adolescent students with hearing loss in mainstream schools and also the correlations between their coping strategies and positive characteristics of well-being (engagement, perseverance, optimism, connectedness and happiness (EPOCH).

MATERIALS & METHODS: In this correlational study, 122 adolescent students with hearing loss were randomly selected from mainstream schools. Data collection was done by EPOCH Measure of Adolescent Well-Being and the Ways of Coping Questionnaire (WAYS). The Spearman correlation coefficient was used for determining the correlations between variables.

RESULTS: The mean scores of using different coping strategies varied from 1.36 in problem solving to 1.44 in seeking support. Among the positive characteristics of well-being, happiness had the lowest (11.04) and connectedness showed the highest score (12.33). The findings also showed a significant correlation between all coping strategies and EPOCH, however there was a strong positive correlation between total coping strategy score and perseverance (0.648) and happiness (0.629).

CONCLUSION: Based on the results, the score of happiness in students with hearing loss was the lowest among positive characteristics of well-being and also happiness showed a strong association with total scores in coping strategies. Accordingly, interventional studies are needed to examine whether training students with hearing loss to use coping strategies is effective in increasing their happiness and overall well-being.

BMC Pediatr. 2020 Apr 20;20(1):175. doi: 10.1186/s12887-020-02080-2.

Parental knowledge and attitudes to childhood hearing loss and hearing services in Qassim, Saudi Arabia.

Alsudays AM, Alharbi AA, Althunayyan FS, Alsudays AA, Alanazy SM, Al-Wutay O, Alenezi MM.

BACKGROUND: Successful audiology service delivery depends on support from the community, and agreement

to utilize hearing healthcare programs. Assessment of parents' awareness regarding hearing loss (HL) and audiology services is necessary for the development of suitable hearing programs for children. Previous studies reported that early detection and intervention for hearing problems are typically strongly supported by parents. The current study sought to evaluate parents' knowledge and attitudes regarding childhood HL and hearing services.

METHODS: A cross-sectional study conducted at five centers in Qassim region of Saudi Arabia. A self-report questionnaire was administered to collect demographic data in addition to 31 questions regarding the knowledge and attitudes of parents toward HL. IBM SPSS Statistics for Windows, Version 21 was used for data analysis. A p-value cut-off point of 0.05 at 95% CI was used to determine statistical significance. The analyses examined the association between socio-demographic characteristics and knowledge and attitudes toward HL using chi-square tests.

RESULTS:

Overall, participants included in this study were 243 participants. Of these, 105 (43.2%) were fathers, and 138 (56.8%) were mothers. Ages ranged from 21 to 60+ years. Assessment of the prevalence of various aspects of knowledge and attitudes among parents toward childhood HL revealed that 103 participants (42.4%) possessed good knowledge, while 140 participants (57.6%) possessed poor knowledge. In contrast, the attitude analysis revealed that 224 participants (92.2%) expressed positive attitudes, while only 19 participants (07.8%) showed a negative attitude regarding audiology services. We found a significant association between age group and knowledge ($p = 0.002$).

CONCLUSION: Most parents in our sample possessed poor knowledge regarding childhood HL. However, most parents expressed positive attitudes regarding audiology services. The current findings suggest a need to increase awareness among parents regarding childhood HL.

Indian J Pediatr. 2020 Apr 13. doi: 10.1007/s12098-020-03260-9

Congenital Cytomegalovirus and Zika Infections.

Angueyra C, Abou Hatab H, Pathak A.

ABSTRACT: Congenital infections affecting newborn infants can have potentially devastating clinical outcomes. They are usually caused by viruses that infect mothers during pregnancy and are transmitted to the fetus or newborn during the prenatal, perinatal or postnatal periods. Congenital cytomegalovirus (cCMV) is the most common congenital infection affecting up to 2.5% of all live births. Even though most infected infants are asymptomatic at birth, cCMV is an important cause of neurodevelopmental impairment and represents the main cause of non-hereditary sensorineural hearing loss. Also, congenital Zika infection has emerged in recent years as a cause of microcephaly and neurodevelopmental delays. Currently, universal screening is not recommended for either infection in pregnant women or newborn infants. Therefore, screening for both conditions is based on multiple factors such as maternal immune status, exposure, and clinical manifestations of the infant. Use of antiviral medications on symptomatic cCMV has shown improvement in outcomes, in contrast with congenital Zika for which there are no therapeutic options available. Even though both viruses can be present in breast milk, there are no recommendations against breastfeeding in full-term infants. Close follow-up for affected infants is necessary to monitor for developmental delays and sensory impairments to implement interventional therapies at the earliest time possible.

Lang Speech Hear Serv Sch. 2020 Jan 8;51(1):68-73. doi: 10.1044/2019_LSHSS-OCHL-19-0025.

Audiological Considerations for Managing Mild Bilateral or Unilateral Hearing Loss in Infants and Young Children.

Bagatto M.

PURPOSE: This clinical focus article describes considerations for recommending assistive hearing technology to infants and young children who have mild bilateral or unilateral hearing loss. These conditions present special challenges compared to bilateral permanent hearing losses that are moderate to profound in their degree in that the recommendation to proceed with technology is not as clear.

CONCLUSION: Current clinical practice guidelines and protocols for pediatric hearing aid fitting recommend managing these conditions on a case-by-case basis. Descriptions of key considerations for recommending assistive hearing technology for infants and young children with mild bilateral hearing loss or unilateral hearing loss are offered herein.

Ear Hear. 2020 Mar 6. doi: 10.1097/AUD.0000000000000862.

Perception of Child-Directed Versus Adult-Directed Emotional Speech in Pediatric Cochlear Implant Users.

Barrett KC, Chatterjee M, Caldwell MT, Deroche MLD, Jiradejvong P, Kulkarni AM, Limb CJ.

OBJECTIVES: Cochlear implants (CIs) are remarkable in allowing individuals with severe to profound hearing loss to perceive speech. Despite these gains in speech understanding, however, CI users often struggle to perceive elements such as vocal emotion and prosody, as CIs are unable to transmit the spectro-temporal detail needed to decode affective cues. This issue becomes particularly important for children with CIs, but little is known about their emotional development. In a previous study, pediatric CI users showed deficits in voice emotion recognition with child-directed stimuli featuring exaggerated prosody. However, the large intersubject variability and differential developmental trajectory known in this population incited us to question the extent to which exaggerated prosody would facilitate performance in this task. Thus, the authors revisited the question with both adult-directed and child-directed stimuli.

DESIGN: Vocal emotion recognition was measured using both child-directed (CDS) and adult-directed (ADS) speech conditions. Pediatric CI users, aged 7-19 years old, with no cognitive or visual impairments and who communicated through oral communication with English as the primary language participated in the experiment (n = 27). Stimuli comprised 12 sentences selected from the HINT database. The sentences were spoken by male and female talkers in a CDS or ADS manner, in each of the five target emotions (happy, sad, neutral, scared, and angry). The chosen sentences were semantically emotion-neutral. Percent correct emotion recognition scores were analyzed for each participant in each condition (CDS vs. ADS). Children also completed cognitive tests of nonverbal IQ and receptive vocabulary, while parents completed questionnaires of CI and hearing history. It was predicted that the reduced prosodic variations found in the ADS condition would result in lower vocal emotion recognition scores compared with the CDS condition. Moreover, it was hypothesized that cognitive factors, perceptual sensitivity to complex pitch changes, and elements of each child's hearing history may serve as predictors of performance on vocal emotion recognition.

RESULTS: Consistent with our hypothesis, pediatric CI users scored higher on CDS compared with ADS speech stimuli, suggesting that speaking with an exaggerated prosody-akin to "motherese"-may be a viable way to convey emotional content. Significant talker effects were also observed in that higher scores were found for the female talker for both conditions. Multiple regression analysis showed that nonverbal IQ was a significant predictor of CDS emotion recognition scores while Years using CI was a significant predictor of ADS scores. Confusion matrix analyses revealed a dependence of results on specific emotions; for the CDS condition's female talker, participants had high sensitivity (d' scores) to happy and low sensitivity to the neutral sentences while for the ADS condition, low sensitivity was found for the scared sentences.

CONCLUSIONS: In general, participants had higher vocal emotion recognition to the CDS condition which also had more variability in pitch and intensity and thus more exaggerated prosody, in comparison to the ADS condition. Results suggest that pediatric CI users struggle with vocal emotion perception in general, particularly to adult-directed speech. The authors believe these results have broad implications for understanding how CI users perceive emotions both from an auditory communication standpoint and a socio-developmental perspective.

Int J Pediatr Otorhinolaryngol. 2020 Mar 24;134:110017. doi: 10.1016/j.ijporl.2020.110017.

Audiologic testing in children with Down Syndrome: Are current guidelines optimal?

Basonbul RA, Ronner EA, Rong A, Rong G, Cohen MS.

INTRODUCTION: Down Syndrome (DS) is a Tier 1 risk factor for hearing loss. Guidelines exist to ensure close monitoring of children with DS for hearing loss. It is important to consider the timing of testing in order to obtain meaningful audiologic data in this high-risk population. The purpose of this study is to present hearing outcomes for children with DS during the first 8 years of life and to assess these outcomes in the context of current screening guidelines.

METHODS: Retrospective review of audiometric outcomes was conducted for children with DS age 8 or younger who presented to a multidisciplinary DS clinic between January 2014 to June 2017. Age at the time of testing, as well as test success rate and hearing loss type and severity were noted.

RESULTS: 131 patients were included in the study, 52% of which were male. 36% of the patients failed their newborn hearing screening and only 9% of those subjects had normal hearing on subsequent testing. Most hearing loss identified was mild and conductive in nature. Inconclusive results were most likely to be obtained at 6-10 months of age.

CONCLUSION: Hearing loss is common among children with DS. To optimize the quality of testing and avoid the need for sedation in followup testing, routine follow-up hearing screening should be performed either before 6 months of age or after 10 months of age.

Fetal Pediatr Pathol. 2020 Jan 25:1-10. doi: 10.1080/15513815.2019.1710788.

Negative Effects of Noise on NICU Graduates' Cochlear Functions.

Beken S, Önal E, Gündüz B, Çakir U, Karagöz İ, Kemaloğlu YK.

AIM: To evaluate the adverse effects of noise on hearing.

METHDOS: Thirty-two infants that had been admitted to neonatal intensive care unit (NICU) and 25 healthy controls were included in this study. Noise levels were recorded continously during the hospitalization period.

RESULTS: All healthy controls passed the hearing screening tests before discharge and on the sixth-month follow up. Hospitalized infants had lower "Distortion Product Auto Acoustic Emission Signal Noise Ratio" (DPOAE SNR) amplitudes (dB) at five frequencies (1001, 1501, 3003, 4004, 6006 Hz in both ears). DPOAE fail rates at 1001 Hz and 1501 Hz were higher than in hospitalized infants (81.8% and 50.0% vs 20.0% and 4.0%). Infants who failed the test at 1001 and 1501 Hz were exposed to noise above the recommended maximum level for longer periods of time.

CONCLUSION: Hearing tests performed at sixth-months of life were adversely affected in NICU graduates.

J Speech Lang Hear Res. 2020 Jan 15;63(1):321-333. doi: 10.1044/2019_JSLHR-19-00230.

Associations Between Parenting Stress, Language Comprehension, and Inhibitory Control in Children With Hearing Loss.

Blank A, Frush Holt R, Pisoni DB, Kronenberger WG.

PURPOSE: Parenting stress has been studied as a potential predictor of developmental outcomes in children with normal hearing and children who are deaf and hard of hearing. However, it is unclear how parenting stress might underlie at-risk spoken language and neurocognitive outcomes in this clinical pediatric population. We investigated parenting stress levels and the shared relations between parenting stress, language comprehension, and inhibitory control skills in children with and without hearing loss (HL) using a cross-sectional design.

METHOD: Families of children with HL ($n = 39$) and with normal hearing ($n = 41$) were tested. Children completed an age-appropriate version of the Concepts & Following Directions subtest of the Clinical Evaluation of Language Fundamentals and the NIH Toolbox Flanker Test of Attention and Inhibitory control. Caregivers completed the Parenting Stress Index-Short Form 4.

RESULTS: Parenting stress levels were not significantly different between parents of children with and without HL. A significant negative association was observed between parenting stress and our measure of language comprehension in children with HL. A negative association between parenting stress and inhibitory control skills was also found in families of children with HL, but not hearing children. The parenting stress-inhibitory control relationship was indirectly accounted for by delayed language comprehension skills in children with HL.

CONCLUSION: Even at moderate levels of parenting stress similar to parents of children with normal hearing, increases in parenting stress were associated with lower scores on our measures of language comprehension and inhibitory control in children with HL. Thus, parenting stress may underlie some of the variability in at-risk pediatric HL outcomes.

Int J Pediatr Otorhinolaryngol. 2020 Apr 8;134:110039. doi: 10.1016/j.ijporl.2020.110039.

Implementation of a neonatal hearing screening programme in three provinces in Albania.

Bussé AM, Qirjazi B, Goedegebure A, Toll M, Hoeve HL, Toçi E, Roshi E, Carr G, Simonsz HJ.

OBJECTIVES: The EUSCREEN study compares the cost-effectiveness of paediatric hearing screening programmes and aims to develop a cost-effectiveness model for this purpose. Alongside and informed by the development of the model, neonatal hearing screening (NHS) is implemented in Albania. We report on the first year.

METHODS: An implementation plan was made addressing objectives, target population, screening protocol, screener training, screening devices, care pathways and follow up. NHS started January 1st, 2018 in four maternity hospitals: two in Tirana, one in Pogradec and one in Kukës, representing both urban and rural areas. OAE-OAE-aABR was used to screen well infants in maternity hospitals, whereas aABR-aABR was used in neonatal intensive care units and in mountainous Kukës for all infants. Screeners' uptake and attitudes towards screening and quality of screening were assessed by distributing questionnaires and visiting the maternity hospitals. The result of screening, diagnostics, follow up and entry into early intervention were registered in a database and monitored.

RESULTS: Screeners were keen to improve their skills in screening and considered NHS valuable for Albanian health care. The number of "fail" outcomes after the first screen was high initially but decreased to less than 10% after eight months. In 2018, 11,507 infants were born in the four participating maternity hospitals, 10,925 (94.9%) of whom were screened in the first step. For 486 infants the result of screening was not registered. For the first screen, ten parents declined, eight infants died and one infant was discharged before screening could be performed. In 1115 (10.2%) infants the test either could not be performed or the threshold was not reached; 361 (32,4%) of these did not attend the second screen. For the third screen 31 (34.4%) out of 90 did not attend. Reasons given were: parents declined (124), lived too far from screening location (95), their infant died (11), had

other health issues (7), or was screened in private clinic (17), no reason given (138).
CONCLUSIONS: Implementation of NHS in Albania is feasible despite continuing challenges. Acceptance was high for the first screen. However, 32.4% of 1115 infants did not attend the second screen, after a "fail" outcome for the first test.

Arch Dis Child. 2020 Feb;105(2):187-189. doi: 10.1136/archdischild-2018-315866.

Risk factors for permanent childhood hearing impairment.

Butcher E, Dezateux C, Knowles RL.

OBJECTIVE: While several perinatal risk factors for permanent childhood hearing impairment (PCHI) are known, association with gestational length remains unclear. We hypothesised that shorter gestational length predicts higher PCHI risk.
DESIGN: 19 504 participants from the UK Millennium Cohort Study (born 2000-2002, prior to newborn screening).
METHODS: Multivariable discrete-time survival analysis to examine associations between parent-reported PCHI by age 11 years and gestational length, plus other prespecified factors.
RESULTS: PCHI affected 2.1 per 1000 children (95% CI 1.5 to 3.0) by age 11; however, gestational length did not predict PCHI risk (HR, 95% CI 1.00, 0.98 to 1.03 per day increase). Risk was increased in those with neonatal illness, with or without admission to neonatal care (6.33, 2.27 to 17.63 and 2.62, 1.15 to 5.97, respectively), of Bangladeshi or Pakistani ethnicity (2.78, 1.06 to 7.31) or born to younger mothers (0.92, 0.87 to 0.97 per year).
CONCLUSION: Neonatal illness, rather than gestational length, predicts PCHI risk. Further research should explore associations with ethnicity.

Int J Pediatr Otorhinolaryngol. 2020 Apr 10;134:110043. doi: 10.1016/j.ijporl.2020.110043.

Etiological profile of hearing loss amongst Lithuanian pediatric cochlear implant users.

Byckova J, Mikstiene V, Kiveryte S, Mickeviciene V, Gromova M, Cernyte G, Mataityte-Dirziene J, Stumbrys D, Utkus A, Lesinskas E.

INTRODUCTION: Congenital sensorineural hearing loss is a heterogeneous disorder; its etiological profile varies between populations. Pathogenic variants of GJB2 gene are the major cause of non-syndromic hearing loss. Congenital cytomegalovirus infection (cCMV) is the most important prenatal etiological factor causing hearing loss and other disorders. Perinatal events, syndromes, postnatal infections or traumas are less common. Causes of the remaining one third of hearing loss cases are unknown.
OBJECTIVES: To determine the etiological profile of hearing loss in pediatric cochlear implant users in Lithuanian population.
METHODS: The data of 122 children (70 male/52 female; aged 7.6 ± 3.3 years) cochlear implant users were analysed. Medical records of all children recruited in Santaros Clinics (Vilnius, Lithuania) were analysed to identify prenatal, perinatal, or postnatal risk factors based on the adapted list proposed by the Joint Committee of Infant Hearing. Genetic counselling and testing according to the scheme were performed to 101 children. DNA of 117 children was extracted from the DBS on Guthrie cards and CMV DNA detected using real time PCR.
RESULTS: Non-syndromic hearing loss was diagnosed in 65 cases (53.3%), 58 of which were GJB2 gene-associated; syndromic hearing loss was diagnosed to 8 children (6.6%). Perinatal (prematurity, low birth weight, hypoxia, hyperbilirubinemia, sepsis, ototoxicity, and meningitis) and postnatal (meningitis) risk factors were associated with hearing loss in 16 (13.1%) and 4 (3.3%) study participants respectively. CMV DNA was detected in 12 samples (9.8%). The cause of hearing loss remained unknown only for 17 (13.9%) children.
CONCLUSIONS: The major cause of HL in the current study was GJB2 gene alterations. Only 14% of the cohort had congenital hearing loss of unknown origin.

Am J Audiol. 2020 Mar 24:1-5. doi: 10.1044/2020_AJA-19-00094.

Deafness Gene Mutations in Newborns in the Foshan Area of South China With Bloodspot-Based Genetic Screening Tests.

Cao S, Sha Y, Ke P, Li T, Yuan W, Huang X.

PURPOSE: The aim of this study was to determine the rate of deafness gene mutations in the Foshan area of South China.
METHOD: We enrolled the infants delivered in Foshan Maternity and Children's Healthcare Hospital. Deafness gene mutation was detected by HibriMax method. Our study tested 47,538 newborns within 3 days after birth, including 13 sites in four genes: *GJB2* (c.35 del G, c.176 del 16, c.235 del C, c.299 del AT, c.155 del TCTG), *GJB3* (c.583 C>T), *SLC26A4* (c.2168 A>G, c.919-2 A>G, c.1299 C>T), and *mtDNA 12S rRNA* (m.1555 A>G, m.1494 C>T, m.12201 T>C, m.7445 A>G). The birth condition of infants was collected, including sex, low or high birth weight, twins, and premature delivery.
RESULTS: In a total of 47,538 newborns, 1,415 were positively identified with deafness gene mutations. The

total rate of the deafness gene mutation was 2.976%. The carrier rates of *GJB2* (c.35 del G, c.176 del 16, c.235 del C, c.299 del AT, c.155 del TCTG), *GJB3* (c.583 C>T), *SLC26A4* (c.2168 A>G, c.919-2 A>G, c.1299 C>T), and *mtDNA 12S rRNA* (m.1555 A>G, m.1494 C>T, m.12201 T>C, m.7445 A>G) mutations were 0.000%, 0.048%, 1.422%, 0.185%, 0.000%, 0.076%, 0.116%, 0.755%, 0.160%, 0.187%, 0.021%, 0.000%, and 0.006%, respectively.

CONCLUSIONS: Our study showed that the c.235 del C *GJB2* mutation was the leading deafness-related mutation in the Foshan area of South China. Deafness gene mutations screening in newborns detected by bloodspot-based genetic screening tests can help the diagnosis of newborn congenital hearing loss.

Int J Environ Res Public Health. 2020 Mar 17;17(6). pii: E1969. doi: 10.3390/ijerph17061969.

Increased Risk of Sensorineural Hearing Loss as a Result of Exposure to Air Pollution.

Chang KH, Tsai SC, Lee CY, Chou RH, Fan HC, Lin FC, Lin CL, Hsu YC.

ABSTRACT: Whether exposure to air pollution is associated with developing sensorineural hearing loss (SHL) remains controversial. Using data from the National Health Insurance Research Database, we recruited a total of 75,767 subjects aged older than 20 years with no history of SHL from 1998 to 2010, and they were followed up until SHL was observed, they withdrew from the National Health Insurance program, or the study ended. The subjects were evenly exposed to low-level, mid-level, and high-level carbon monoxide (CO) and nitrogen dioxide (NO_2). The incidence rate ratio of SHL for patients exposed to high-level CO was 1.24 (95% confidence interval (CI) = 1.14-1.36). The NO_2 pollutants increased the incidence rate ratios of SHL in mid-level NO_2 and high-level NO_2 exposures by 1.10 (95% CI = 1.10-1.32) and 1.36 (95% CI = 1.24-1.49) times, respectively. The adjusted hazard ratio (adj. HR) of SHL in patients exposed to high-level CO was 1.45 (95% CI = 1.31-1.59), relative to that of patients exposed to low-level CO. Compared to patients exposed to low-level NO_2, patients exposed to mid-level NO_2 (adj. HR = 1.40, 95% CI = 1.27-1.54) and high-level NO_2 (adj. HR = 1.63, 95% CI = 1.48-1.81) had a higher risk of developing SHL. The increased risk of SHL following the increased concentrations of air pollutants (CO and NO_2) was statistically significant in this study. In conclusion, the subjects' exposure to air pollution exhibited a significantly higher risk of developing SHL in Taiwan.

Mol Biol Rep. 2020 Apr 22. doi: 10.1007/s11033-020-05460-0.

Mesenchymal stem cells for sensorineural hearing loss: a systematic review of preclinical studies.

Chorath K, Willis M, Morton-Gonzaba N, Moreira A.

ABSTRACT: Sensorineural hearing loss (SNHL) is the most common form of hearing loss that is routinely treated with hearing aids or cochlear implants. Advances in regenerative medicine have now led to animal studies examining the possibility of restoring injured hair cells with mesenchymal stem/stromal cell (MSC) administration. We conducted a systematic review and meta-analysis to collate the existing preclinical literature evaluating MSCs as a treatment for SNHL and quantify the effect of MSCs on functional hearing. Our protocol was published online on CAMARADES. Searches were conducted in four medical databases by two independent investigators. Twelve studies met inclusion and were evaluated for risk of bias using SYRCLE. Rodent models were commonly used (n = 8, 66%), while auditory brainstem response (ABR) and distortion product otoacoustic emissions (DPOAE) were the most frequent measures assessing hearing loss. MSCs were derived from multiple tissue sources, including bone marrow, adipose tissue, and umbilical cord blood and the dose ranged from 4×10^3 to 1×10^7 cells. Treatment with MSCs resulted in an improvement in ABR and DPOAE (mean difference-15.22, + 9.10, respectively). Despite high heterogeneity and multiple "unclear" domains in the risk of bias, this review provides evidence that MSCs may have a beneficial effect in hearing function.

Pediatr Infect Dis J. 2020 Apr;39(4):273-276. doi: 10.1097/INF.0000000000002564.

Middle Ear Effusion in Children With Congenital Cytomegalovirus Infection.

Chung W, Leung J, Lanzieri TM, Blum P, Demmler-Harrison G, Ahmed S, Baer H, Bhatt AR, Brown F, Catlin F, Caviness AC, Coats DK, Edmonds JC, Flores M, Franklin D, Gandaria C, Greer J, Griesser C, Hussein MA, Iovino I, Istas A, Jin HD, Kelinske MK, Klingen JT, Laurente A, Littman T, Murphy M, Miller J, Nelson C, Noyola D, Paysse EA, Percy A, Reis S, Reynolds A, Rozelle J, Smith O, Steinkuller P, Turcich M, Vinson SS, Voigt RG, Walmus B, Williams J, Williamson D, Yen KG, Yow MD; Congenital Cytomegalovirus Longitudinal Study Group; Congenital Cytomegalovirus Longitudinal Study Group.

BACKGROUND: Sensorineural hearing loss (SNHL) is well described in children with congenital cytomegalovirus (CMV) infection, but limited data are available on middle ear effusion (MEE) occurrence in this population. We assessed the prevalence of MEE and the degree of transient hearing change associated with MEE among

children with congenital CMV infection.
METHODS: Children with congenital CMV infection enrolled in a longitudinal study received hearing and tympanometric testing during scheduled follow-up visits annually up to 6 years of age. We used a generalized linear mixed-effect logistic regression model to compare the odds of MEE, defined as type B tympanogram (normal ear canal volume with little tympanic membrane movement) among patients categorized as symptomatic or asymptomatic based on the presence of congenital CMV-associated signs in the newborn period.
RESULTS: Forty-four (61%) of 72 symptomatic and 24 (28%) of 87 asymptomatic patients had ≥1 visit with MEE. After controlling for the number of visits, symptomatic patients had significantly higher odds of MEE (odds ratio: 2.09; 95% confidence interval: 1.39-3.14) than asymptomatic patients. Transient hearing decrease associated with a type B tympanogram ranged from 10 to 40 dB, as measured by audiometric air-bone gap in 11 patients.
CONCLUSIONS: Among children with congenital CMV, MEE can result in transient hearing decrease, which can reduce the efficacy of a hearing aid in those with SNHL. It is warranted that children with congenital CMV infection and SNHL receive routine audiologic and tympanometric testing to better manage hearing aid amplification levels.

Otol Neurotol. 2019 Dec;40(10):1278-1286. doi: 10.1097/MAO.0000000000002410.

Long-term Outcomes in Down Syndrome Children After Cochlear Implantation: Particular Issues and Considerations.

Clarós P, Remjasz A, Clarós-Pujol A, Pujol C, Clarós A, Wiatrow A.

OBJECTIVE: The aim of the study was to analyze the long-term outcomes after cochlear implantation in deaf children with Down syndrome (DS) regarding age at the first implantation and refer the results to preoperative radiological findings as well as postoperative auditory and speech performance. Additionally, the influence of the age at implantation and duration of CI use on postoperative hearing and language skills were closely analyzed in children with DS.
STUDY DESIGN: Retrospective analysis.
SETTING: Referral center (Cochlear Implant Center).
MATERIALS AND METHODS: Nine children with Down syndrome were compared with 220 pediatric patients without additional mental disorders or genetic mutations. Patients were divided into four categories depending on the age of the first implantation: CAT1 (0-3 yr), CAT2 (4-5 yr), CAT3 (6-7 yr), and CAT4 (8-17 yr). The auditory performance was assessed with the meaningful auditory integration scales (MAIS) and categories of auditory performance (CAP) scales. The speech and language development were further evaluated with meaningful use of speech scale (MUSS) and speech intelligibility rating (SIR). The postoperative speech skills were analyzed and compared between the study group and the reference group by using nonparametric statistical tests. Anatomic abnormalities of the inner ear were examined using magnetic resonance imaging (MRI) and high-resolution computed tomography of the temporal bones (HRCT).
RESULTS: The mean follow-up time was 14.9 years (range, 13.1-18.3 yr). Patients with DS received a multichannel implant at a mean age of 75.3 months (SD 27.9; ranging from 21 to 127 mo) and 220 non-syndromic children from reference group at a mean age of 51.4 months (SD 34.2; ranging from 9 to 167 mo). The intraoperative neural response was present in all cases. The auditory and speech performance improved in each DS child. The postoperative mean CAP and SIR scores were 4.4 (SD 0.8) and 3.2 (SD 0.6), respectively. The average of scores in MUSS and MAIS/IT-MAIS scales was 59.8% (SD 0.1) and 76.9% (SD 0.1), respectively. Gathered data indicates that children with DS implanted with CI at a younger age (<6 years of age) benefited from the CI more than children implanted later in life, similarly in a control group. There were additional anomalies of the temporal bone, external, middle, or inner ear observed in 90% of DS children, basing on MRI or HRCT.
CONCLUSIONS: The early cochlear implantation in children with DS is a similarly useful method in treating severe to profound sensorineural hearing loss (SNHL) as in non-syndromic patients, although the development of speech skills present differently. Due to a higher prevalence of ear and temporal bone malformations, detailed diagnostic imaging should be taken into account before the CI qualification. Better postoperative outcomes may be achieved through comprehensive care from parents/guardians and speech therapists thanks to intensive and systematic rehabilitation.

Eur J Radiol. 2020 Feb;123:108803. doi: 10.1016/j.ejrad.2019.108803. Epub 2019 Dec 26.

Temporal bone and intracranial abnormalities in syndromic causes of hearing loss: an updated guide.

D'Arco F, Youssef A, Ioannidou E, Bisdas S, Pinelli L, Caro-Dominguez P, Nash R, Siddiqui A, Talenti G.

PURPOSE: To describe in detail the temporal bone and brain findings in both common and rare syndromic causes of hearing loss, with the purpose of broadening among radiologists and enhance the current understanding of distinct imaging features in paediatric patients with syndromic hearing loss.
METHODS: A detailed search of electronic databases has been conducted, including PubMed, Ovid Medline,

Scopus, Cochrane Library, Google Scholar, National Institute for Health and Care Excellence (NICE), Embase, and PsycINFO.

RESULTS: Syndromic causes of hearing loss are characterised by different and sometimes specific abnormalities in the temporal bone.

CONCLUSION: A complete knowledge of the image findings in the temporal bones, brain, skull and other body regions is critical for the optimal assessment and management of these patients.

Int J Pediatr Otorhinolaryngol. 2020 Apr 10;134:110036. doi: 10.1016/j.ijporl.2020.110036.

Nature and extent of hearing loss in HIV-infected children: A scoping review.

Dawood G, Klop D, Olivier E, Elliott H, Pillay M, Grimmer K.

INTRODUCTION: Antiretroviral therapy (ART) has had a major impact on life expectancy from HIV as many people now live with it as a chronic disease. Chronic HIV has been associated with a range of comorbid disabilities and health conditions, one of which is hearing loss. Undiagnosed and untreated hearing loss, particularly in children, has been linked to poorer spoken language skills, with subsequent effects on academic performance.

METHODS: This systematic scoping review aimed to summarize the available peer-reviewed literature on hearing loss in HIV-infected children, specifically to describe its extent and nature. The review followed the framework proposed by Arksey and O'Malley. Key search terms included hearing loss (and synonyms), child (and synonyms), and HIV. Electronic databases (EBSCOhost Research Platform, PubMed, Web of Science and Scopus databases) were searched for any relevant articles published from January 1, 2000 to June 30, 2019. Reference lists of included articles were pearled for additional relevant articles not already identified. Each stage of the selection process was conducted independently by two authors. The results were then collated by a third author who also resolved any discrepancies. Extracted data included sample descriptors, audiologic tests, hearing loss prevalence, hearing loss descripts, and factors associated with hearing loss.

RESULTS: Seventeen articles were included; 10 from Africa, four from South America, two from North America and the remaining article from Asia. Although most of the articles reported on pure tone audiometry, the samples as well as the cut-off criteria for normal hearing were heterogenous. Prevalence of hearing loss varied across articles (from 6% to 84%). Conductive hearing loss occurred more frequently than sensorineural or mixed hearing loss. ART use and ear infection were reported as significant in three of five articles that reported on significant associates of HIV-related hearing loss.

CONCLUSION: There was a modest volume of research from a limited number of countries. Heterogeneity in sampling and audiometric methods precluded a clear understanding of potential associations between chronic HIV-related hearing loss and contributing factors.

Laryngoscope. 2020 Feb 17. doi: 10.1002/lary.28561.

Cochlear Implantation in Children with Single-Sided Deafness.

Deep NL, Gordon SA, Shapiro WH, Waltzman SB, Roland JT Jr, Friedmann DR.

OBJECTIVE: To describe our experience with children undergoing unilateral cochlear implantation (CI) for treatment of single-sided deafness (SSD).

STUDY DESIGN: Retrospective case series.

METHODS: A retrospective case review from a tertiary referral center involving 14 pediatric patients (<18 years) with SSD who underwent unilateral CI. Speech perception testing in quiet and noise in the CI-only and bimodal conditions with at least 1 year of device use and device usage from data logs represent the main outcome measures.

RESULTS: The mean age at CI was 5.0 years (median 4.4, range 1.0-11.8 years). The mean duration of deafness was 3.0 years (median 2.4, range 0.6-7.0 years). Mean follow-up was 3.4 years. Speech perception testing with a minimum of 1 year post-CI was available in eight patients. The mean word recognition scores (WRS) in the CI-only condition was 56%; a significant improvement from baseline. Testing in background noise with spatially separated speech and noise revealed that patients scored as well or better with the CI-on versus CI-off in all conditions and in no cases was interference from the CI noted. Data logs were reviewed for device usage which revealed an average use of 6.5 hr/d.

CONCLUSION: Cochlear implantation is a viable treatment option for pediatric SSD in this self-selected cohort. Open-set speech and improvement in background noise can be achieved. Careful patient selection and thorough counseling on expectations is paramount to achieving successful outcomes.

JMIR Hum Factors. 2020 Mar 10. doi: 10.2196/16310.

Usability of a Mobile App for Improving Literacy in Children with Hearing Impairment: A Focus Group Study.

DeForte S, Sezgin E, Huefner J, Lucius S, Luna J, Satyapriya AA, Malhotra P.

BACKGROUND: Children with hearing loss, even those identified early and who are using hearing aids or cochlear implants, may face challenges in developing spoken language and literacy. This can lead to academic, behavioral, and social difficulties. There are apps for healthy children to improve their spoken language and literacy and apps that focus on sign language proficiency for children with hearing loss, but these apps are limited for children with hearing loss. We have therefore developed an app called Hear Me Read (HMR) which uses enhanced digital stories as therapy tools for speech, language, and literacy for children with hearing loss. The platform has therapist and parent/child modes that allows 1) selection of high quality, illustrated digital stories by a speech-language pathologist (SLP), parent, or child 2) modification of digital stories for a multitude of speech and language targets, and 3) assignment of stories by therapist to facilitate individualized speech and language goals. Additionally, HMR makes the caregiver a core partner in engagement through functionality whereby the caregiver can record video and audio of themselves to be played back by the child.

OBJECTIVE: The objective of this study was to evaluate the user experience of the HMR app through a focus group study with caregivers and their children.

METHODS: We recruited 16 participants (8 children with and without hearing loss and 8 caregivers) to participate in one-hour focus groups. Caregivers and children interacted with the app and discussed their experience through a semi-structured group interview. We employed thematic analysis methods and analyzed the data. We used feedback from the focus group to improve elements of the app for a larger clinical trial assessing the impact of the app on outcomes.

RESULTS: We identified 3 themes: default needs, specific needs and family needs. Participants found the app to be aesthetically pleasing and easy to use. Findings helped us to identify usability attributes and to amend app functionalities to best fit user needs. Caregivers and children appreciated the enhancements, such as parts of speech highlighting and video playback of caregivers reading, that were made possible by the digital format. Participants expressed that the app could be used to enhance family reading sessions and family interaction.

CONCLUSIONS: The findings from this focus group study are promising for the use of educational apps designed specifically for those with hearing loss who are pursuing listening and spoken language as a communication outcome. Further investigation is needed with larger sample sizes in order to understand the clinical impact on relevant language and literacy outcomes in this population.

J Pediatr. 2020 Mar;218:151-156.e2. doi: 10.1016/j.jpeds.2019.12.005. Epub 2020 Jan 14.

A Cross-Sectional Study of Caregiver Perceptions of Congenital Cytomegalovirus Infection: Knowledge and Attitudes about Screening.

Diener ML, Shi K, Park AH.

OBJECTIVES: To understand caregiver knowledge of and attitudes toward congenital cytomegalovirus (cCMV) testing in Utah.

STUDY DESIGN: We surveyed 365 caregivers whose children were being seen in an otolaryngology clinic at a tertiary pediatric hospital about their knowledge of and attitudes toward cCMV and cCMV screening. Descriptive statistics and cluster analysis were used to examine their responses.

RESULTS: The majority of caregivers were unsure how cCMV was spread, the symptoms of cCMV, and why cCMV screening of infants was important. Most caregivers did not know that cCMV screening was required by law in Utah if an infant is referred after newborn hearing screening. A majority wanted to know if their child had cCMV even if asymptomatic and were willing to pay $20 for cCMV screening. Caregivers of children who had been tested for cCMV were significantly more likely to be strongly in favor of cCMV screening than expected by chance. Caregivers in the highly knowledgeable cluster were more likely to be strongly in favor of cCMV screening.

CONCLUSIONS: Caregivers frequently were unaware of cCMV and its implications. Attitudes toward cCMV screening generally were positive. Education on epidemiology and impact of cCMV may benefit both prevention of infection and attitudes toward screening.

Eur J Pediatr. 2020 May;179(5):807-812. doi: 10.1007/s00431-019-03558-7. Epub 2020 Jan 11.

Treatment of congenital cytomegalovirus beyond the neonatal period: an observational study.

Dorfman L, Amir J, Attias J, Bilavsky E.

ABSTRACT: Recently, valganciclovir treatment of symptomatic congenital cytomegalovirus (cCMV) disease, commenced during the neonatal period (≤4 weeks), was found to improve hearing and developmental outcome. However, many children (symptomatic or asymptomatic at birth) present only after 4 weeks of age. The purpose of this observational retrospective study was to describe the outcome and safety of valganciclovir therapy in infants with cCMV who started treatment >4 weeks of life. Of the 91children who started antiviral treatment

>4 weeks of age, 66/298 (22.2%) were symptomatic at birth; 25/217 (11.5%) were asymptomatic at birth. Treatment was initiated on average at 14 weeks of age (range 5-77 weeks) and at 53.3 weeks (range 12-156 weeks), respectively. Of the 45 affected ears in the symptomatic group, 30 (66.7%) improved and only 2 (4.4%) deteriorated, with most of the improved ears (27/30, 90%) returning to normal. In the asymptomatic group, late-onset treatment was initiated and out of the 42 deteriorated ears, 38 (90.5%) improved after at least 1 year of follow-up. Hematological adverse events, i.e., neutropenia, were noted in a minority of cases (4.4%).Conclusion: Our study demonstrates the benefits and safety aspects of treating symptomatic and asymptomatic children with cCMV even beyond the recommended neonatal period.What is Known:• Valganciclovir treatment of symptomatic congenital cytomegalovirus (cCMV) disease, commenced during the neonatal period, is beneficial in improving hearing and developmental outcome.• However, data of treatment started beyond the neonatal period is lacking. What is New:• Our study demonstrates the benefits of treating symptomatic children with cCMV as well as asymptomatic children that develop late-onset hearing loss even beyond the recommended neonatal period.• This was true for symptomatic children who presented >4 weeks as well as to those were asymptomatic at birth but experienced late hearing deterioration.

Eur J Hum Genet. 2020 May;28(5):587-596. doi: 10.1038/s41431-019-0553-8. Epub 2019 Dec 12.

Exome sequencing in infants with congenital hearing impairment: a population-based cohort study.

Downie L, Halliday J, Burt R, Lunke S, Lynch E, Martyn M, Poulakis Z, Gaff C, Sung V, Wake M, Hunter MF, Saunders K, Rose E, Lewis S, Jarmolowicz A, Phelan D, Rehm HL; Melbourne Genomics Health Alliance, Amor DJ.

ABSTRACT: Congenital hearing impairment (HI) is the most common sensory impairment and can be isolated or part of a syndrome. Diagnosis through newborn hearing screening and management through early intervention, hearing aids and cochlear implantation is well established in the Australian setting; however understanding the genetic basis of congenital HI has been missing. This population-derived cohort comprised infants with moderate-profound bilateral HI born in the 2016-2017 calendar years, detected through newborn hearing screening. Participants were recruited through an integrated paediatric, otolaryngology and genetics HI clinic and offered whole exome sequencing (WES) on a HiSeq4000 or NextSeq500 (Illumina) platform with a targeted average sequencing depth of 100x and chromosome microarray on the Illumina Infinium core exome-24v1.2 platform. Of those approached, 68% (106/156) consented to participate. The rate of genetic diagnosis was 56% (59/106), significantly higher than standard of care (GJB2/6 sequencing only), 21% (22/106). There were clinical implications for the 106 participants: 36% required no further screening, 9% had tailored screening initiated, 2% were offered treatment and 4% had informed care for a complex neurodevelopmental syndrome. WES in this cohort demonstrates the range of diagnoses associated with congenital HI and confirms the genetic heterogeneity of congenital HI. The high diagnostic yield and clinical implications emphasises the need for genomic sequencing to become standard of care.

Genet Med. 2020 Jan 24. doi: 10.1038/s41436-019-0745-1.

Exome sequencing in newborns with congenital deafness as a model for genomic newborn screening: the Baby Beyond Hearing project.

Downie L, Halliday J, Lewis S, Lunke S, Lynch E, Martyn M, Gaff C, Jarmolowicz A, Amor DJ.

PURPOSE: Genomic newborn screening raises practical and ethical issues. Evidence is required to build a framework to introduce this technology safely and effectively. We investigated the choices made by a diverse group of parents with newborns when offered tiered genomic information from exome sequencing.
METHODS: This population-derived cohort comprised infants with congenital deafness. Parents were offered exome sequencing and choice regarding the scope of analysis. Options were choice A, diagnostic analysis only; choice B, diagnostic analysis plus childhood-onset diseases with medical actionability; or choice C, diagnostic analysis plus childhood-onset diseases with or without medical actionability.
RESULTS: Of the 106 participants, 72 (68%) consented to receive additional findings with 29 (27.4%) selecting choice B and 43 (40.6%) opting for choice C. Family size, ethnicity, and age of infant at time of recruitment were the significant predictors of choice. Parents who opted to have additional findings analysis demonstrated less anxiety and decisional conflict.
CONCLUSIONS: These data provide evidence from a culturally diverse population that choice around additional findings is important and the age of the infant when this choice is offered impacts on their decision. We found no evidence that offering different levels of genomic information to parents of newborns has a negative psychological impact.

Int J Pediatr Otorhinolaryngol. 2020 Apr;131:109864. doi: 10.1016/j.ijporl.2020.109864. Epub 2020 Jan 7.

Comparison of ABR and ASSR using NB-chirp-stimuli in children with severe and profound hearing loss.

Eder K, Schuster ME, Polterauer D, Neuling M, Hoster E, Hempel JM, Semmelbauer S.

INTRODUCTION: Objective techniques for hearing threshold estimation in infants and children with profound or severe hearing loss play a key role in pediatric audiology to prevent speech acquisition disorders by choosing the adequate therapy. Auditory brainstem responses and auditory steady-state responses are available for frequency-dependent hearing threshold estimations and both techniques show strong correlations. However, various systems and stimuli are available, which is one reason why comparison is challenging, and, so far, no single "gold standard" could be established for hearing threshold estimation in children suffering from profound or severe hearing loss. The aim of the study was to compare hearing threshold estimations in children with profound or severe hearing loss derived with narrow-band CE-chirps evoked auditory brainstem responses and auditory steady-state response.

SUBJECTS and METHODS: 71 children (121 ears) with an age from 3 month to 15 years were measured with the Interacoustics Eclipse EP25 ABR system® (Denmark) with narrow-band CE-chirps® at 500, 1000, 2000 and 4000 Hz under identical conditions.

RESULTS: Auditory brainstem responses and auditory steady-state responses highly correlate ($r = 0.694$, $p < 0.001$). Correlation coefficients differ depending on the center frequency and patient age. Generally, auditory steady-state responses show a better hearing threshold than auditory brainstem responses or a remaining hearing threshold when auditory brainstem responses could not be obtained. In approximately 15% of cases this would have affected the therapeutic strategy when only taking one technique into account.

CONCLUSION: Auditory brainstem responses and auditory steady-state responses should be jointly used in the diagnostic approach in children with suspected profound or severe hearing loss.

Int J Pediatr Otorhinolaryngol. 2019 Dec;127:109681. doi: 10.1016/j.ijporl.2019.109681. Epub 2019 Sep 13.

Evaluation and therapy outcome in children with auditory neuropathy spectrum disorder (ANSD).

Ehrmann-Müller D, Cebulla M, Rak K, Scheich M, Back D, Hagen R, Shehata-Dieler W.

OBJECTIVES: The aims of the present study are to: describe diagnostic findings in patients with auditory neuropathy spectrum disorder (ANSD); and demonstrate the outcomes of different therapies like hearing aids (HAs) or cochlear implantation.

METHODS: 32 children were diagnosed and treated at our tertiary referral center and provided with HAs or cochlear implants (CIs). All of them underwent free-field or pure-tone audiometry. Additionally, otoacoustic emissions (OAEs), impedance measurements, auditory brainstem responses (ABRs), auditory steady-state responses (ASSR), electrocochleography, and cranial magnetic resonance imaging (cMRI) were all performed. Some patients also underwent genetic evaluation. Following suitable provision pediatric audiological tests, psychological developmental diagnostic and speech and language assessments were carried out at regular intervals in all the children.

RESULTS: OAEs could initially be recorded in most of the children; 17 had no ABRs. The other eight children had a poor ABR morphology. Most of the children had typical, long-oscillating cochlear microphonics (CMs) in their ABRs, which was also observed in all of those who underwent electrocochleography. Eight children were provided with a HA and 17 received a CI. The functional gain was between 32 and 65 decibel (dB) with HAs and between 32 and 50 dB with CI. A speech discrimination level between 35 and 100% was achieved during open-set monosyllabic word tests in quiet with HA or CI. With the Hochmair-Schulz-Moser (HSM) sentence test at 65 dB SPL (sound pressure level), 75% of the children with a CI achieved a speech discrimination in noise score of at least 60% at a signal to noise ratio (SNR) of 5, and four scored 80% or higher. Most of the children (72%) were full-time users of their devices. All the children with a CI used it on a regular basis.

CONCLUSION: Only a few case reports are available in the literature regarding the long-term outcomes of ANSD therapy. The present study reveals satisfactory outcomes with respect to hearing and speech discrimination in children with CIs or HAs. The nearly permanent use of the devices reflects a subjective benefit for the children. Provision with a suitable hearing device depends on audiological results, the speech and language development of an individual child, and any accompanying disorders. Repeated audiological evaluations, interdisciplinary diagnostics, and intensive hearing and speech therapy are essential for adequate rehabilitation of this group of children.

Blood Coagul Fibrinolysis. 2020 Apr 22. doi: 10.1097/MBC.0000000000000911.

Thrombosis risk of Alport syndrome patients: evaluation of cardiological, clinical,

biochemical, genetic and possible causes of inherited thrombophilia and identification of a novel COL4A3 variant.

Eroz R, Damar İH, Kılıçaslan O.

ABSTRACT: To evaluate cases with Alport syndrome for laboratory, radiological, ophthalmological, auditory tests, cardiological and inherited thrombophilia risk. Laboratory findings, abdominal and urinary ultrasonography, ophthalmological and auditory tests and cardiological examination of 21 Alport syndrome suspicious cases were performed. Also, collagen type IV alpha three chain (COL4A3) gene, four chain (COL4A4) gene and five chain (COL4A5) genes were sequenced by next-generation sequencing system. In addition, possible causes of inherited thrombophilia were evaluated. A novel (c.2806C>T/p.Gln936Ter) variation in COL4A3 gene was detected in three cases. Also c.221G>A/p.Arg74Gln variation in COL4A5 gene of two cases, c.4421C>T/p.Thr1474Met variation in COL4A4 gene of one case, c.665C>T/p.Pro222Leu variation in COL4A4 gene of one case and compound heterozygous c.4421C>T/(p.Thr1474Met) and c.665C>T/p.Pro222Leu variation in COL4A4 gene of one case were detected. Although 10 (47.6%) cases had microscopic hematuria, six (28.6%) cases had macroscopic hematuria, but there were not hematuria in five (23.8%) of cases. Three cases with variation carrier in COL4A genes and one case without variation carrier had vision problem. Also, one case with variation carrier in COL4A gene had hearing loss. All cases with variation carrier in COL4A genes exclude one had at least one cardiac problems. Also, all cases with variation carrier in COL4A genes had possible causes of inherited thrombophilia risk. In addition to developing risk of progressive kidney failure, sensorineural hearing loss and ocular abnormalities, Alport syndrome cases may have increasing cardiac problems and possible causes of inherited thrombophilia risk. Therefore, these cases should be regularly evaluated and followed for cardiac problems and inherited thrombophilia risk.

World J Pediatr. 2020 Jan 7. doi: 10.1007/s12519-019-00325-4.

Etiology of newborn hearing impairment in Guangdong province: 10-year experience with screening, diagnosis, and follow-up.

Fang BX, Cen JT, Yuan T, Yin GD, Gu J, Zhang SQ, Li ZC, Liang YF, Zeng XL.

BACKGROUND: Hearing impairment is one of the most common birth defects in children. Universal newborn hearing screenings have been performed for 19 years in Guangdong province, China. A screening/diagnosis/intervention system has gradually been put in place. Over the past 10 years, a relatively complete data management system had been established. In the present study, an etiological analysis of newborn cases that failed the initial and follow-up screenings was performed.

METHODS: The nature and degree of hearing impairment in newborns were confirmed by a set of procedures performed at the time of initial hearing screening, rescreening and final hearing diagnosis. Then, multiple examinations were performed to explore the associated etiology.

RESULTS: Over a period of 10 years, 720 children were diagnosed with newborn hearing loss. Among these children, 445 (61.81%) children had a clearly identified cause, which included genetic factor(s) (30.56%), secretory otitis media (13.30%), maternal rubella virus infection during pregnancy (5.83%), inner ear malformations (4.86%), maternal human cytomegalovirus infection during pregnancy (2.92%), malformation of the middle ear ossicular chain (2.50%) and auditory neuropathy (1.81%). In addition, 275 cases of sensorineural hearing loss of unknown etiology accounted for 38.19% of the children surveyed.

CONCLUSIONS: Long-term follow-up is needed to detect delayed hearing impairment and auditory development in children. The need for long-term follow-up should be taken into account when designing an intervention strategy. Furthermore, the use of the deafness gene chip should further elucidate the etiology of neonatal hearing impairment.

Am J Audiol. 2019 Dec 16;28(4):1025-1045. doi: 10.1044/2019_AJA-19-0061. Epub 2019 Dec 12.

Candidacy for Amplification in Children With Hearing Loss: A Review of Guidelines and Recommendations.

Fitzpatrick EM, Cologrosso E, Sikora L.

PURPOSE: The 1st point in the intervention process for the majority of children is the fitting of hearing devices. The objective of this review was to compile guidelines and recommendations for candidacy criteria for children with hearing loss.

METHOD: Electronic databases (e.g., MEDLINE, Embase, and Cumulative Index of Nursing and Allied Health Literature) and websites were searched. Any document referring to children with hearing loss that discussed amplification guidelines or protocols was included. Documents specific to implantable devices or addressing only remote microphone systems were excluded. One reviewer screened all potentially relevant documents, and a subset was screened by a 2nd reviewer. Guidelines/recommendations referring to pediatric amplification candidacy were extracted.

RESULTS: A total of 40 documents were included for data extraction. Studies were categorized according to hearing loss of any degree, with separate categories for documents providing specific criteria for mild bilateral, unilateral, and auditory neuropathy spectrum disorders. Guidelines ranged from generic statements about the need for amplification to criteria based on specific audiometric thresholds. In guidelines recommending audiometric cut-points, the majority considered > 25 dB HL as a criterion for consideration for amplification. Overall, guidelines for children with mild bilateral and unilateral loss remain more ambiguous, and there was some variation across the recommendations. Guidelines for auditory neuropathy spectrum disorder stressed the need to obtain results from behavioral audiometry before considering amplification.
CONCLUSIONS: Numerous organizations have established candidacy guidelines for pediatric amplification. Most guidelines specify criteria for amplification as audiometric threshold levels. There is considerable variation in the guidelines for mild bilateral and unilateral hearing loss with candidacy criteria ranging from 15 to 30 dB HL, and many guidelines recommend a case-by-case decision approach.

Int J Pediatr Otorhinolaryngol. 2020 Feb 27;133:109975. doi: 10.1016/j.ijporl.2020.109975.

Impact of Universal Newborn Hearing Screening on cochlear implanted children in Ireland.

Gabriel MM, Geyer L, McHugh C, Thapa J, Glynn F, Walshe P, Simoes-Franklin C, Viani L.

OBJECTIVES: Cochlear Implant (CI) is an established treatment for severe to profound hearing loss (HL). Early diagnosis and intervention in HL are crucial in order to provide access to sound and increase the likelihood of spoken language development in pre-lingually deaf children. In April 2011, the Health Service Executive (HSE) implemented the Universal Newborn Hearing Screening (UNHS) in a phased regional basis in Ireland. This study aimed to investigate the general clinical pathway for UNHS referrals to the CI service and to evaluate the impact of earlier referrals via UNHS on functional outcomes in children.
METHODS: The first part of this study constituted a retrospective review of 100 children referred to the National Hearing Implant and Research Centre (NHIRC) via UNHS from November 2011 to December 2016. Implanted children referred via UNHS were categorised into three groups according to their medical status. Their clinical pathway to cochlear implantation was evaluated. Functional outcomes were investigated based on medical and developmental status, respectively. In the second part of this study, developmentally healthy implanted children referred post-UNHS were compared with medically healthy children referred pre-UNHS under the age of four, from January 2005 to June 2011. Current implant status of children, age at referral and functional outcomes were investigated.
RESULTS: Medically healthy children were referred to the NHIRC at an earlier age than the medically complex children (2.8 months vs 5.2 months, $p < 0.01$) and the children presenting with auditory neuropathy spectrum disorder (ANSD) (2.8 months vs 5.3 months, $p < 0.01$). On average they attended their first appointment and were implanted at a younger age than the ANSD group (6.1 months vs 10.1 months, $p < 0.01$; 16.3 months vs 29.4 months, $p < 0.001$, respectively). Developmentally healthy children had significantly better functional outcomes than children with developmental delays. Children referred via UNHS were referred and implanted at a younger age than those referred pre-UNHS. The former group achieved better Categories of Auditory Performance (CAP) and Speech Intelligibility Rating (SIR) scores 2 years post-implantation.
CONCLUSION: UNHS in Ireland is an important platform for earlier diagnosis and management of congenital HL and our results show that early intervention has a positive impact on functional outcomes in children.

J Clin Microbiol. 2020 Mar 25;58(4). pii: e01951-19. doi: 10.1128/JCM.01951-19. Print 2020 Mar 25.

Performance of the Alethia CMV Assay for Detection of Cytomegalovirus by Use of Neonatal Saliva Swabs.

Gantt S, Goldfarb DM, Park A, Rawlinson W, Boppana SB, Lazzarotto T, Mertz LM.

ABSTRACT: Congenital cytomegalovirus (cCMV) infection is a major cause of childhood hearing loss and neurodevelopmental delay. Identification of newborns with cCMV infection allows provision of beneficial interventions. However, most infants with cCMV infection have subclinical infection and go undiagnosed. Thus, expanded neonatal CMV testing is increasingly recommended. Saliva is an attractive sample type for CMV testing of newborns, because it is easier to collect than urine and more sensitive for CMV detection than dried blood spots. We evaluated the Alethia CMV assay, a rapid, easy-to-use loop-mediated isothermal amplification method for qualitative detection of CMV DNA in neonatal saliva samples. Saliva swabs were collected prospectively from newborns <21 days old and tested by the Alethia assay according to the manufacturer's instructions. Archived saliva swabs from newborns with cCMV infection were also tested retrospectively. A composite reference method (CRM; two validated PCR assays followed by bidirectional sequencing of amplicons) was performed on all samples as the reference standard comparator. Of 1,480 prospectively collected saliva swabs, 1,472 (99.5%) were negative by both the Alethia assay and CRM, 5 (0.34%) were positive by both the Alethia assay and CRM, and 3 (0.20%) were positive only by the Alethia assay. All 34 (100%) archived swabs from newborns with cCMV infection were positive by both the CRM and the Alethia assay. Overall, the Alethia assay showed 100% and

99.8% positive and negative agreement with the CRM, respectively. The Alethia CMV assay is an accurate method for identifying neonates with cCMV infection and, given its simplicity, appears suitable for CMV testing using neonatal saliva outside a reference laboratory, including remote and resource-limited settings.

Medicine (Baltimore). 2020 Mar;99(13):e19373. doi: 10.1097/MD.0000000000019373.

Screening for mitochondrial 12S rRNA C1494T mutation in 655 patients with non-syndromic hearing loss: An observational study.

Gao Z, Yuan YS.

ABSTRACT: Mutations in mitochondrial DNA, especially in 12S rRNA gene, are the most important causes for hearing loss. In particular, the A1555G and C1494T mutations have been found to be associated with both aminoglycoside-induced and non-syndromic hearing loss in many families worldwide. To determine the frequency of C1494T mutation in deaf patients, in the current study, we screened this mutation in 655 patients with non-syndromic hearing loss and 300 control subjects. After PCR amplification of mitochondrial 12S rRNA gene and direct sequence analysis, we found that there were 2 patients carrying the C1494T mutation; however, this mutation was not detected in 300 healthy subjects. Further genetic counseling suggested that only 1 patient had an obvious family history of hearing impairment. Clinical evaluation showed that 3 of 10 matrilineal relatives suffered from hearing loss, with different age at onset of hearing loss. Molecular analysis revealed the presence of homoplasmic 12S rRNA C1494T and ND5 T12338C mutations, together with a set of polymorphisms belonging to human mitochondrial haplogroup F2. Interestingly, T12338C mutation resulted in the replacement of the first amino acid, a translation-initiating methionine with a threonine, shortening 2 amino acids of ND5 polypeptide. Moreover, this mutation is located in 2 nucleotides adjacent to the 3' end of the mt-tRNALeu(CUN) gene. Therefore, this mutation may alter ND5 mRNA metabolism and the processing of RNA precursors. Thus, the combination of T12338C and C1494T mutations may contribute to deafness expression in this family. Taken together, our data suggested that the C1494T mutation was the molecular basis for hearing loss, screening for the mitochondrial DNA pathogenic mutations was recommended for early detection, prevention, and diagnosis of mitochondrial deafness.

J Perinatol. 2020 May;40(5):774-780. doi: 10.1038/s41372-020-0628-y. Epub 2020 Feb 26.

Treatment for hypotension in the first 24 postnatal hours and the risk of hearing loss among extremely low birth weight infants.

Gogcu S, Washburn L, O'Shea TM.

OBJECTIVE: To evaluate whether treated hypotension in the first 24 postnatal hours is associated with hearing loss in extremely low birth weight (ELBW) infants.
STUDY DESIGN: In a cohort of 735 ELBW infants, we identified 25 with sensorineural hearing loss (SNHL) at 12-24 months adjusted age. For each case, we selected three controls with normal hearing. Logistic regression models were used to adjust for confounding variables.
RESULTS: Sixty percent of cases and 25% of controls were treated for hypotension. After adjusting for confounding variables (gestational age, antenatal glucocorticoids, 5 min Apgar < 6, insertion of an umbilical catheter, treatment with high frequency ventilation, and major cranial ultrasound abnormality), treated hypotension was associated with an increased risk of SNHL (adjusted odds ratio: 3.6; 95% confidence interval: 1.3-9.7).
CONCLUSIONS: Treated hypotension in ELBW infants in the first 24 h of life is associated with an increased risk of SNHL.

Cureus. 2020 Jan 4;12(1):e6566. doi: 10.7759/cureus.6566.

Prevalence of Sensorineural Hearing Loss in Children with Palliated or Repaired Congenital Heart Disease.

Gopineti L, Paulpillai M, Rosenquist A, Van Bergen AH.

BACKGROUND: Children with congenital heart disease (CHD) are at increased risk of neurodevelopmental deficits, and the presence of sensorineural hearing loss (SNHL) may further lead to poor language skills acquisition and speech delays. Prevalence of SNHL in the general pediatric population is estimated to be 0.2% at birth to 0.35% during adolescence. Very few studies have attempted to estimate SNHL prevalence in children who have undergone congenital heart surgery.
METHODS: This retrospective study aimed to estimate SNHL prevalence in children who underwent congenital heart surgery in our institution and were followed up in our high-risk pediatric cardiology clinics for four years from 2009 to 2013. Data were collected on demographics, preoperative variables, surgical variables, and post-operative variables.
RESULTS: SNHL prevalence in asymptomatic, palliated/repaired CHD patients followed in our high-risk clinics and undergoing routine surveillance was 11.6% (20 of 172 patients with hearing impairment). SNHL prevalence

was not statistically higher in single-ventricle patients (17.2%) compared to biventricular patients (14.7%). Inotropic score in the first 24 hours of postoperative period (p=0.05), lowest arterial PaO2 (p=0.003), duration of Lasix drip (p=0), and bolus dose in days (p=0.03) were all found to be statistically significant in the hearing-impaired group. However, using logistic regression, we identified no statistically significant predictors for hearing loss.
CONCLUSION: The results suggest the need for routine audiology screening of all patients with complex CHD, especially those who have undergone neonatal cardiac repair/palliation at less than one year of age, irrespective of risk factors.

Hum Genet. 2020 Apr;139(4):521-530. doi: 10.1007/s00439-020-02118-6. Epub 2020 Jan 30.

Concurrent hearing and genetic screening in a general newborn population.

Guo L, Xiang J, Sun L, Yan X, Yang J, Wu H, Guo K, Peng J, Xie X, Yin Y, Wang J, Yang H, Shen J, Zhao L, Peng Z.

ABSTRACT: Newborn hearing screening is not designed to detect delayed-onset prelingual hearing loss or aminoglycoside-antibiotic-induced ototoxicity. Cases with severe to profound hearing loss have been reported to have been missed by newborn hearing screens. The aim of this study was to evaluate the efficacy of concurrent hearing and genetic screening in the general population and demonstrate its benefits in practice. Enrolled newborns received concurrent hearing and genetic screens between September 1, 2015 and January 31, 2018. Of the 239,636 eligible infants (median age, 19 months), 548 (0.23%) had prelingual hearing loss. Genetic screening identified 14 hearing loss patients with positive genotypes and 27 patients with inconclusive genotypes who had passed the hearing screens. In addition, the genetic screen identified 0.23% (570/239,636) of the newborns and their family members as at-risk for ototoxicity, which is undetectable by hearing screens. In conclusion, genetic screening complements newborn hearing screening by improving the detection of infants at risk of hereditary hearing loss and ototoxicity, and by informing genotype-based clinical management for affected infants and their family members. Our findings suggest that the practice should be further validated in other populations and rigorous cost-effectiveness analyses are warranted.

Laryngorhinootologie. 2020 Mar 4. doi: 10.1055/a-1114-6452. [Article in German]

Evaluation of Universal Newborn Hearing Screening and follow-up.

Hall V, Brosch S, Hoffmann TK.

BACKGROUND: Universal newborn hearing screening (UNHS) was established in Germany in 2009. Even compliance was tested in early studies, there is little knowledge regarding the follow-up examination of children with suspected hearing disorder.
METHODS: A retrospective evaluation was performed in 570 cases of children who failed newborn hearing screening for the years between 2009-2016. Hearing deficiency was defined as having a hearing threshold ≥35dB. Compliance with national guidelines was checked. Every child received brainstem evoked response audiometry (BERA).
RESULTS: Permanent hearing disorder was found in 24%, of whom about half (51%) had an inner ear hearing loss (of these in 73% bilateral). Only 27% of high risk children born in peripheral hospitals were tested immediately by the envisaged automated auditory brainstem response (AABR) method. They often presented tardy, leading to a delayed diagnosis and therapy. Children tracked by the Bavarian health office presented little earlier but had less cases who were lost to follow-up.
DISCUSSION: In 93% a diagnosis was made during first examination and therapy (e.g. prescription of hearing aids) initiated on average within four months age. The rate of deafness corresponded with national averages. The quality of primary screenings is crucial in revealing problems and avoiding delay in dealing with them.

J Neurooncol. 2020 Jan;146(1):147-156. doi: 10.1007/s11060-019-03356-z. Epub 2019 Nov 28.

Effect of sensorineural hearing loss on neurocognitive and adaptive functioning in survivors of pediatric embryonal brain tumor.

Heitzer AM, Villagran AM, Raghubar K, Brown AL, Camet ML, Ris MD, Hanning JH, Okcu MF, Paulino AC, Chintagumpala M, Kahalley LS.

PURPOSE: Survivors of pediatric embryonal brain tumors (BT) are at high risk for sensorineural hearing loss (SNHL) associated with neurocognitive decline. However, previous studies have not assessed the relationship between SNHL and adaptive functioning. We examined neurocognitive and adaptive functioning in patients with and without SNHL.
METHODS: Participants included 36 patients treated for an embryonal BT with craniospinal irradiation (CSI) and cisplatin chemotherapy who were assessed 6.7 years post-treatment on average. The impact of SNHL on neurocognitive performance and parent-rated adaptive functioning was assessed in univariate and multivariate analyses.

RESULTS: There were 17 cases with SNHL (mean age at evaluation = 14.4) and 19 cases with NH (mean age at evaluation = 13.8). After accounting for age at diagnosis and additional covariates in multivariable analyses, SNHL was associated with worse overall intellectual functioning (p = 0.027) and perceptual reasoning (p = 0.016) performance. There was no effect of SNHL on adaptive functioning in multivariable models. Age at diagnosis and sex were associated with performance on neurocognitive measures.
CONCLUSIONS: SNHL in pediatric embryonal BT is associated with increased risk for neurocognitive deficits in conjunction with other demographic and treatment-related factors.

Cleft Palate Craniofac J. 2019 Dec 23:1055665619895635. doi: 10.1177/1055665619895635.

Eustachian Tube Dysfunction in Children With Unilateral Cleft Lip and Palate: Differences Between Ipsilateral and Contralateral Ears.

Hu A, Shaffer AD, Jabbour N.

OBJECTIVE: To evaluate Eustachian tube dysfunction in the ipsilateral and contralateral ears, in children with unilateral cleft lip and palate (UCLP).
DESIGN: Retrospective chart review.
SETTING: Tertiary care children's hospital.
PATIENTS: Seventy-four consecutive patients with UCLP born between 2005 and 2011 and treated at UPMC Children's Hospital of Pittsburgh Cleft-Craniofacial Center were included.
MAIN OUTCOME MEASURES: Conductive hearing loss, tympanogram type, number of middle ear effusions, tympanostomy tubes, and complications. Hypothesis was formulated prior to data collection.
RESULTS: Conductive hearing loss was nearly twice as common in the ipsilateral ear (43.2%) compared with contralateral (23.0%; *P* = .001, McNemar test). There were no significant differences in the frequency of each type of tympanogram between the contralateral and ipsilateral ears. The proportions of ipsilateral (90.5%) and contralateral (91.9%) ears with effusion were not significantly different. The total number of tubes received was not significantly different between the 2 ears (median of 2 bilaterally). When combined, complications (retractions, perforations, and cholesteatomas) were significantly more common in the ipsilateral ear (29.7%) compared with the contralateral ear (18.9%; *P* = .039, McNemar test).
CONCLUSION: In children with UCLP, there were significantly more instances of conductive hearing loss and complications on the cleft side compared to the noncleft side. This suggests that Eustachian tube dysfunction may indeed be more severe on the cleft side. Considering this information, clinicians may need to be especially observant of the ipsilateral ear.

Am J Audiol. 2020 Mar 5;29(1):23-34. doi: 10.1044/2019_AJA-19-00054. Epub 2020 Jan 14.

Auditory Detection Thresholds and Cochlear Resistivity Differ Between Pediatric Cochlear Implant Listeners With Enlarged Vestibular Aqueduct and Those With Connexin-26 Mutations.

Jahn KN, Bergan MD, Arenberg JG.

PURPOSE: The goal of this study was to evaluate differences in the electrode-neuron interface as a function of hearing loss etiology in pediatric cochlear implant (CI) listeners with enlarged vestibular aqueduct (EVA) syndrome and in those with autosomal recessive connexin-26 mutations (DFNB1).
METHOD: Fifteen implanted ears (9 participants, 5 ears with EVA, 10 ears with DFNB1) were assessed. Single-channel auditory detection thresholds were measured using broad and spatially focused electrode configurations (steered quadrupolar; focusing coefficients = 0 and 0.9). Cochlear resistivity estimates were obtained via electrode impedances and electrical field imaging. Between-group differences were evaluated using linear mixed-effects models.
RESULTS: Children with EVA had significantly higher auditory detection thresholds than children with DFNB1, irrespective of electrode configuration. Between-group differences in thresholds were more pronounced on apical electrodes than on basal electrodes. In the apex, electrode impedances and electrical field imaging values were higher for children with EVA than for those with DFNB1.
CONCLUSIONS: The electrode-neuron interface differs between pediatric CI listeners with DFNB1 and those with EVA. It is possible that optimal clinical interventions may depend, in part, on hearing loss etiology. Future investigations with large samples should investigate individualized CI programming strategies for listeners with EVA and DFNB1.

Iran J Otorhinolaryngol. 2020 Mar;32(109):85-92. doi: 10.22038/ijorl.2019.36090.2191.

Prevalence of Hearing Loss among School-Age Children in the North of Iran.

Jalali MM, Nezamdoust F, Ramezani H, Pastadast M.

INTRODUCTION: The present study aimed to investigate the audiological profiles of elementary school-age children in Rasht, Iran, and estimate the prevalence of hearing impairments in this population.

MATERIALS AND METHODS: In this cross-sectional descriptive-analytical study, the hearing threshold was screened using pure tone audiometry (PTA). Hearing impairment was defined as equal to or higher than 20 dB HL. Results of the hearing thresholds were separately reported in the left or right ears and better or worse ears. Logistic regression tests were used to investigate the association between hearing loss and possible risk factors. In this study, all the analyses were conducted using SPSS software (version 21).
RESULTS: The present study was carried out on a total of 2019 children. Mean age of the participants was reported as 9.66±1.66 years. Based on low-frequency pure-tone average, the prevalence rates of hearing loss > 15 dB in the right and left ears were reported as 1.94% and 1.68%, respectively. The high-frequency hearing loss > 15 dB in the right and left ears was obtained at 1.14% and 1.04%, respectively. Prevalence rate of hearing loss (in all frequencies) in boys was higher than that in girls. There was a strong association between a history of otitis media and sensorineural or conductive hearing loss (adjusted odds ratio reported as 12.2 and 8.1, respectively).
CONCLUSION: In this study, the rate of hearing loss in the participants was approximately 2%. It was concluded that the screening of hearing loss in children is necessary for the identification and management of these children as early as possible. It is recommended to perform further trials to investigate the impact of different causes on childhood hearing impairment.

J Infect Dis. 2020 Mar 5;221(Supplement_1):S9-S14. doi: 10.1093/infdis/jiz446.

Congenital Cytomegalovirus Infection.

Kabani N, Ross SA.

ABSTRACT: Congenital cytomegalovirus (cCMV) infection is a leading cause of hearing loss and neurological disabilities in children, with the disease burden and disabilities due to cCMV greater than many other well recognized childhood conditions. A minority of infants with cCMV will have symptoms at birth. Infants with symptomatic cCMV are at higher risk for sequelae than those born without symptoms. The majority of infants with cCMV are asymptomatic at birth, but 10%-15% will develop hearing loss. Although clinical symptoms can help predict which infants will have sensorineural hearing loss, among asymptomatic cCMV there are currently no predictors of adverse outcome. The identification of a biomarker to identify those at highest risk of sequelae is highly desirable to target interventions to those who could potentially benefit. Because there is increasing rationale for establishing both targeted and universal screening programs for cCMV in the United States and worldwide, this is an urgent priority.

Lancet Infect Dis. 2020 Feb;20(2):220-229. doi: 10.1016/S1473-3099(19)30416-5. Epub 2019 Nov 7.

Congenital viral infections in England over five decades: a population-based observational study.

Kadambari S, Pollard AJ, Goldacre MJ, Goldacre R.

BACKGROUND: Congenital viral infections cause substantial long-term morbidity but population-based data about diagnosis rates are scarce. The aim of this study was to assess the long-term trends in congenital viral infections in England and to report on how the rates of these infections might have changed with improved methods for detection, the introduction of the two-dose measles-mumps-rubella (MMR) vaccine in 1996, and the implementation of the Newborn Hearing Screening Programme (NHSP) in 2006.
METHODS: For this population-based, observational cohort study, we used national and regional hospitalisation data from 1968 to 2016 in England (Hospital In-Patient Enquiry, Hospital Episode Statistics, and Oxford Record Linkage Study) to calculate annual rates of hospital discharges coded with-and individuals aged younger than 1 month diagnosed with-congenital cytomegalovirus, herpes simplex virus (HSV), varicella zoster virus (VZV), and rubella. We investigated associations of congenital cytomegalovirus, HSV, and VZV with perinatal and maternal factors (sex, mother's ethnicity, mode of delivery, gestational age, birthweight, mother's age, mother's index of multiple deprivation, and number of previous pregnancies).
FINDINGS: In 2016, discharge rates per 100 000 infant population were 22·3 (95% CI 18·8-26·1) for congenital cytomegalovirus, 17·6 (14·6-21·1) for HSV, 32·6 (28·4-37·2) for VZV, and 0·15 (0·0-0·8) for rubella. Compared with earlier years of the study, the discharge rate in 2016 was higher for congenital cytomegalovirus, HSV, and VZV, whereas it was lower for rubella. For congenital cytomegalovirus, there was a significant step-increase between 2006 and 2007 following implementation of the NHSP (rate ratio comparing the trend line post-NHSP with that pre-NHSP 1·55 [95% CI 1·12-2·14], p=0·0072). Congenital cytomegalovirus infection was associated with birthweight less than 1 kg, maternal age younger than 25 years, socioeconomically deprived households, casearean section, and mothers of black ethnicity. Congenital HSV infection was associated with maternal age younger than 20 years, gestational age less than 32 weeks, and vaginal and emergency caesarean section deliveries, while VZV infection was associated with increased parity and black and south Asian ethnicities.
INTERPRETATION: The increase in hospital discharges coded with congenital cytomegalovirus is most likely due to the introduction of sensitive diagnostic techniques and retrospective diagnoses made in infants after implementation of the NHSP. Public health strategies to improve prevention and treatment of congenital viral

infections are urgently warranted. The decrease in discharges for rubella is most likely due to the MMR vaccine.

Laryngoscope. 2020 Jan;130(1):212-216. doi: 10.1002/lary.27722. Epub 2018 Dec 8.

Clinical Guidelines in Pediatric Hearing Loss: Systemic Review Using the Appraisal of Guidelines for Research and Evaluation II Instrument.

Kanabur P, Hubbard C, Jeyakumar A.

OBJECTIVES: Despite the importance, impact, and prevalence of pediatric hearing loss (HL), there are very few published clinical practice guidelines (CPG) supporting the evaluation and management of pediatric patients with HL. Our objective was to appraise existing CPGs to ensure safe and effective practices.
METHODS: A literature search was conducted in PubMed, Google Scholar, EBSCO, as well as a manual Google search. Three independent assessors using the Appraisal of Guidelines for Research and Evaluation II (AGREE II) instrument evaluated CPGs related to HL in children. Standardized domain scores were calculated for each guideline.
RESULTS: A total of four guidelines met the inclusion criteria and were appraised. Scope and purpose achieved a high median score of 83%. Stakeholder involvement, clarity of presentation, and editorial independence achieved intermediate scores of 67%, 54%, and 50%, respectively. The areas that required most improvement and achieved low scores were rigor of development and applicability, with scores of 22% and 38%, respectively. Based on the AGREE II measures, the four guidelines had domain scores less than 60% for each domain, and without modification no guideline could be recommended.
CONCLUSIONS: Based on the AGREE II, the qualities of CPGs for pediatric HL have several shortcomings, and the need for a comprehensive CPG remains. Rigor of development and applicability present the greatest opportunities for improvement of these CPGs.

J Turk Ger Gynecol Assoc. 2020 Jan 13. doi: 10.4274/jtgga.galenos.2019.2019.0070.

Does Antenatal Magnesium Sulphate improve hearing function in premature newborns?

Kasapoğlu I, Çetinkaya Demir B, Atalay MA, Orhan A, Özkan H, Çakır SC, Tütüncü Toker R, Kasapoğlu F, Özerkan K.

OBJECTIVE: To evaluate whether antenatal magnesium sulphate (MgSO4) exposure has a neuroprotective effect against hearing impairment in premature newborns.
MATERIALS AND METHODS: Retrospective cohort study performed with prematurely (<37 weeks) delivered newborns at a tertiary university hospital. Newborns of 92 women who received MgSO4 infusions (study group) for various indications were compared to newborns of 147 women who did not receive MgSO4 infusions (control group). Every eligible premature newborn underwent hearing screening by auditory brainstem response (ABR) testing before being discharged from the hospital.
RESULTS: The fail rate in ABR hearing screening was 3.3% (n=3) in the study group and 10.9% (n=16) in the control group (p=0.034). The rate of concurrent use of betamethasone was higher in the study group (72.8%; n= 67) compared to control group (29.2%; n=43) (p<0.001). Other neonatal parameters such as the number of neonates who are small for gestational age and the rate of microcephaly were similar between the groups (p=0.54, p=0.48, respectively). After adjusting for co-variates including the use of betamethasone and gestational age at delivery, we did not find any statistically significant association between antenatal administration of MgSO4 and fail rates in hearing screening by ABR testing (p=0.07).
CONCLUSION: Our results do not suggest a clear and definite benefit from antenatal MgSO4 infusion in respect of hearing impairment in premature newborns.

Int J Environ Res Public Health. 2020 Apr 10;17(7). pii: E2613. doi: 10.3390/ijerph17072613.

What Are the Current Audiological Practices for Ototoxicity Assessment and Management in the South African Healthcare Context?

Khoza-Shangase K, Masondo N.

ABSTRACT: The study was an initial exploration of the current ototoxicity assessment and management practices by audiologists in South Africa. An exploratory survey research methodology through a cross-sectional research design was adopted where audiologists were recruited from professional associations' databases in South Africa, using specific inclusion criteria. The study made use of an 18-item web-based survey guided by the Health Professions Council of South Africa (HPCSA) (2018) guidelines which were developed from reviewing international guidelines such as the American Speech-Language-Hearing Association (ASHA,1994) and the American Academy of Audiology (AAA, 2009). The study surveyed 31 audiologists from across the country. Data were analyzed through descriptive statistics. Findings implied significant gaps between knowledge and translation of this knowledge into practice. Over two thirds of the participants engage with ototoxicity monitoring and management, but the practices adopted by them do not align with international standards nor with the national

HPCSA guidelines on assessment and management of patients on ototoxic medications. Most participants do not conduct baseline assessments, and the frequency of monitoring is irregular and reduced from the recommended; thus influencing ability for early detection and intervention of ototoxicity within this context. Non-standard assessment battery is used for assessment and monitoring, raising questions about the reliability and validity of the data used to make preventive treatment decisions. Lack of collaborative work between audiologists and the rest of the clinical team involved in the treatment of patients on ototoxic medications was found to be an important contributing factor to the less than optimal ototoxicity management practices. Of factors potentially influencing adherence to guidelines, the institution of employment, specifically employment in a tuberculosis hospital, seemed to have a positive influence, possibly due to the focused nature of the audiologists' scope of practice there as well as availability of resources. The level of education appeared to have no influence. Current findings provide contextually relevant evidence on ototoxicity assessment and management within this context. They raise important implications for guidelines adherence and translating knowledge, policies and guidelines into practice, clinical assessment and management protocols followed, appropriate resource allocation per programme, as well as strategic planning for national ototoxicity assessment and management programmes in context. The findings also raise important implications for low- and middle-income countries, in terms of adopting international guidelines without considering context.

Genet Med. 2020 Mar 17. doi: 10.1038/s41436-020-0774-9.

Significant Mendelian genetic contribution to pediatric mild-to-moderate hearing loss and its comprehensive diagnostic approach.

Kim BJ, Oh DY, Han JH, Oh J, Kim MY, Park HR, Seok J, Cho SD, Lee SY, Kim Y, Carandang M, Kwon IS, Lee S, Jang JH, Choung YH, Lee S, Lee H, Hwang SM, Choi BY.

PURPOSE: Timely diagnosis and identification of etiology of pediatric mild-to-moderate sensorineural hearing loss (SNHL) are both medically and socioeconomically important. However, the exact etiologic spectrum remains uncertain. We aimed to establish a genetic etiological spectrum, including copy-number variations (CNVs) and efficient genetic testing pipeline, of this defect.

METHODS: A cohort of prospectively recruited pediatric patients with mild-to-moderate nonsyndromic SNHL from 2014 through 2018 (n = 110) was established. Exome sequencing, multiplex ligation-dependent probe amplification (MLPA), and nested customized polymerase chain reaction (PCR) for exclusion of a pseudogene, STRCP, from a subset (n = 83) of the cohort, were performed. Semen analysis was also performed to determine infertility (n = 2).

RESULTS: Genetic etiology was confirmed in nearly two-thirds (52/83 = 62.7%) of subjects, with STRC-related deafness (n = 29, 34.9%) being the most prevalent, followed by MPZL2-related deafness (n = 9, 10.8%). This strikingly high proportion of Mendelian genetic contribution was due particularly to the frequent detection of CNVs involving STRC in one-third (27/83) of our subjects. We also questioned the association of homozygous continuous gene deletion of STRC and CATSPER2 with deafness-infertility syndrome (MIM61102).

CONCLUSION: Approximately two-thirds of sporadic pediatric mild-to-moderate SNHL have a clear Mendelian genetic etiology, and one-third is associated with CNVs involving STRC. Based on this, we propose a new guideline for molecular diagnosis of these children.

Clin Exp Otorhinolaryngol. 2020 Jan 14. doi: 10.21053/ceo.2019.01144.

A Retrospective Review of Temporal Bone Computed Tomography to Present Safe Guideline for Bone-Anchored Hearing Aids.

Kim S, Cho YS, Cho YS, Moon IJ.

OBJECTIVES: Bone-anchored hearing device (BAHD) is contraindicated in patients younger than 5 years because their calvarial bones are not thick enough to be implanted site. However, it has not been studied in the Korean population. This study was not only to establish a safe guideline for depth of implant device in all age groups who undergo BAHD implant surgery, but also to investigate whether implantation of currently used BAHDs could be done safely in Korean children, especially those younger than 5.

METHODS: Two hundred eighty patients, who underwent high-resolution temporal bone computed tomography (TBCT) images between August 2010 and October 2018 were randomly enrolled in all ages. We retrospectively reviewed TBCT imaging to measure skull bone thickness at the recommended BAHD implant site.

RESULTS: The average skull bone thickness was 2.87 mm in patients younger than 5 years and 6.72 mm in patients older than 5 years, respectively, which conforms to the current guideline. The results indicate nearly 50% of calvarial bone thicknesses were less than 3 mm in patients under 5 years old, while 92.78% of the patients older than 5 years of age showed bone thickness greater than 4 mm. Of note, calvarial bone thickness was thicker than 3 mm in all patients who are older than 6 years.

CONCLUSION: This study confirms that the currently approved BAHD implantation guideline is suitable in the

Korean population. For safety, we suggest taking TBCTs prior to surgery, especially in pediatric patients. Besides, noninvasive applications are recommended for patients younger than 5.

Int Arch Otorhinolaryngol. 2020 Apr;24(2):e198-e205. doi: 10.1055/s-0039-1698775. Epub 2020 Jan 28.

An Investigation of Hearing (250-20,000 Hz) in Children with Endocrine Diseases and Evaluation of Tinnitus and Vertigo Symptoms.

Kocyigit M, Bezgin SU, Cakabay T, Ortekin SG, Yıldız M, Ozkaya G, Aydın B.

INTRODUCTION: Despite much advancement in medicine, endocrine and metabolic diseases remain an important cause of morbidity and even mortality in children.
OBJECTIVE: The present study was planned to investigate the evaluation of hearing that also includes high frequencies, and the presence and degree of vertigo and tinnitus symptoms in pediatric patients diagnosed with endocrine diseases such as type 1 diabetes mellitus (DM), growth hormone deficiency (GHD), obesity, idiopathic short stature, and precocious puberty
METHODS: The present study included a patient group of 207 children patients diagnosed with endocrine disease (95 males, 112 females; mean age 9.71 years old [range 6-16 years old]) and a control group including 55 healthy children who do not have any kind of chronic disease (26 males, 29 females; mean age 9.33 years old [range 6-16 years old]). The subjects underwent a hearing test with frequencies between 250 and 20,000 Hz. The vestibular and tinnitus symptoms were evaluated with the Pediatric Vestibular Symptom Questionnaire.
RESULTS: Out of 207 patients in the patient group, 5 (2.4%) had hearing loss in pure tones, 10 (4.8%) had it in high frequencies, 40 (19.3%) had tinnitus symptoms, and 18 (8.7%) had vertigo symptoms. A total of 4 out of 207 patients in the study group (1.9%), 2 out of 59 with type 1 DM patients (3.4%), 1 out of 46 with GHD (2.2%), and 1 out of 43 obesity patients (2.3%) had hearing loss, vertigo, and tinnitus symptoms.
CONCLUSIONS: Our results suggest that some childhood endocrine diseases can cause some changes in the inner ear, although the exact cause is unknown. Perhaps, a detailed hearing and balance examination should be a routine in a child diagnosed with an endocrine disease. We think it is necessary to work on more comprehensive patient groups and tests in the future.

Int J Pediatr Otorhinolaryngol. 2020 Feb 3;132:109926. doi: 10.1016/j.ijporl.2020.109926.

Limitations and drawbacks of the hospital-based universal neonatal hearing screening program: First report from the Arabian Peninsula and insights.

Kolethekkat AA, Al Abri R, Hlaiwah O, Al Harasi Z, Al Omrani A, Sulaiman AA, Al Bahlani H, Al Jaradi M, Mathew J.

OBJECTIVES: To assess the efficacy of the current universal neonatal hearing screening program in a tertiary medical institution in Oman, identify its limitations and drawbacks, and explore their causative factors.
METHODS: A retrospective review was carried out to analyse the hearing screening of 12,743 live babies born between January 2016 and December 2018. Screen coverage, drop outs, follow up rate, and age at completion of screening, diagnosis, and intervention were analysed. The results were compared with the Joint Committee on Infant Hearing (JCIH) performance quality indices. Prospective questionnaire-based telephonic interviews were then conducted with the parents or caregivers of neonates with hearing loss. Finally, the causes of loss to follow up or delays in hearing screenings, diagnosis, and/or early intervention were studied.
RESULTS: The true prevalence of hearing loss was 4.0 in 1000. The coverage of first-stage screening was 90% whereas the compliance with the second stage was 88.04%. 22.8% of the patients eventually obtained final diagnostic confirmation. The overall compliance with amplification was 30.2%. The completion ages of primary screening and final confirmation were 7.98 and 17.3 weeks respectively. The importance of hearing screening is well received by parents, but problems related to communication, delays in the appointment system, and inefficient follow up tracking were identified as the main limitations and drawbacks of the program.
CONCLUSION: The coverage of the neonatal hearing screening program had not yet reached the required goal of 95%. The performance indicators also fell below the international benchmark. There is a need to address the identified causative factors. Effective communication and well-maintained tracking systems need to be implemented.

Int J Pediatr Otorhinolaryngol. 2020 Jan 28;132:109906. doi: 10.1016/j.ijporl.2020.109906.

The efficacy of bone-anchored hearing implant surgery in children: A systematic review.

Kruyt IJ, Bakkum KHE, Caspers CJI, Hol MKS.

OBJECTIVE: To evaluate the efficacy of Bone-Anchored Hearing implants (BAHIs) in children and to elucidate the usage and outcomes of new surgical techniques and implants in this specific population. **DATA SOURCES:** Embase and PubMed.
STUDY SELECTION: We identified studies evaluating surgical outcomes of BAHIs in children. Retrieved articles

were screened using predefined inclusion and exclusion criteria. Critical appraisal included directness of evidence and risk of bias. Studies that successfully passed critical appraisal were included.
DATA EXTRACTION: Outcome measures included patient demographics, follow-up time, surgical technique (one-versus two-stage surgery), tissue handling technique (reduction versus preservation), type of implant used, and complications.
DATA SYNTHESIS: We selected 20 articles published between 2000 and 2017 for data extraction, encompassing 952 implanted BAHIs. The overall mean age at implantation was 8.6 years (range, 2-21 years). Adverse soft-tissue reactions occurred in 251 of the 952 implants (26.4%; range 0%-89% across studies). Revision surgery was performed in 16.8% (142 of the 845) of the implants. The total rate of implant loss, i.e. caused by OIF (n = 61), trauma (n = 33), recurrent infection (n = 15), elective removal due to insufficient benefit (n = 1), cosmetic reasons (n = 1), or unknown reason (n = 16), was 13.3% of the implants (127 out of 952; range 0%-40% across studies). Differences are seen in the type of implants used; wide-diameter implants seem to be superior in terms of implant survival, and similar in terms of adverse skin reactions, while one-stage surgery and soft-tissue preservation do not seem to result in higher implant loss rates or increased adverse skin reactions based upon limited amounts of literature.
CONCLUSION: In general, BAHIs are a safe method for hearing rehabilitation in children, although large differences between studies are observed. The outcomes of new surgical techniques and implant designs in the pediatric population seem promising, but more research is needed before definitive conclusions can be drawn.

Front Pediatr. 2020 Jan 31;8:13. doi: 10.3389/fped.2020.00013. eCollection 2020.

Congenital Cytomegalovirus Infection: A Narrative Review of the Issues in Screening and Management From a Panel of European Experts.

Lazzarotto T, Blázquez-Gamero D, Delforge ML, Foulon I, Luck S, Modrow S, Leruez-Ville M.

ABSTRACT: Maternal primary and non-primary cytomegalovirus (CMV) infection during pregnancy can result in *in utero* transmission to the developing fetus. Congenital CMV (cCMV) can result in significant morbidity, mortality or long-term sequelae, including sensorineural hearing loss, the most common sequela. As a leading cause of congenital infections worldwide, cCMV infection meets many of the criteria for screening. However, currently there are no universal programs that offer maternal or neonatal screening to identify infected mothers and infants, no vaccines to prevent infection, and no efficacious and safe therapies available for the treatment of maternal or fetal CMV infection. Data has shown that there are several maternal and neonatal screening strategies, and diagnostic methodologies, that allow the identification of those at risk of developing sequelae and adequately detect cCMV. Nevertheless, many questions remain unanswered in this field. Well-designed clinical trials to address several facets of CMV treatment (in pregnant women, CMV-infected fetuses and both symptomatic and asymptomatic neonates and children) are required. Prevention (vaccines), biology and transmission factors associated with non-primary CMV, and the cost-effectiveness of universal screening, all demand further exploration to fully realize the ultimate goal of preventing cCMV. In the meantime, prevention of primary infection during pregnancy should be championed to all by means of hygiene education.

Value Health. 2020 Feb;23(2):164-170. doi: 10.1016/j.jval.2019.07.019. Epub 2019 Oct 10.

Health-Related Quality of Life in Children With Low Language or Congenital Hearing Loss, as Measured by the PedsQL and Health Utility Index Mark 3.

Le HND, Petersen S, Mensah F, Gold L, Wake M, Reilly S.

OBJECTIVES: To examine health-related quality of life (HRQoL) in young children with low language or congenital hearing loss and to explore
the value of assessing HRQoL by concurrently administering 2 HRQoL instruments in populations of children.
METHODS: Data were from 2 Australian community-based studies: Language for Learning (children with typical and low language at age 4 years, n = 1012) and the Statewide Comparison of Outcomes study (children with hearing loss, n = 108). HRQoL was measured using the parent-reported Health Utilities Index Mark 3 (HUI3) and the Pediatrics Quality of Life Inventory 4.0 (PedsQL) generic core scale. Agreement between the HRQoL instruments was assessed using intraclass correlation and Bland-Altman plots.
RESULTS: Children with low language and with hearing loss had lower HRQoL than children with normal language; the worst HRQoL was experienced by children with both. The lower HRQoL was mainly due to impaired school functioning (PedsQL) and speech and cognition (HUI3). Children with hearing loss also had impaired physical and social functioning (PedsQL), vision, hearing, dexterity, and ambulation (HUI3). Correlations between instruments were poor to moderate, with low agreement.
CONCLUSIONS: Children with low language and congenital hearing loss might benefit from interventions targeting overall health and well-being, not just their impairments. The HUI3 and PedsQL each seemed to provide unique information and thus may supplement each other in assessing HRQoL of young children, including those with low language or congenital hearing loss.

Am J Obstet Gynecol. 2020 Feb 24. pii: S0002-9378(20)30198-8. doi: 10.1016/j.ajog.2020.02.018.

Cytomegalovirus infection during pregnancy: state of the science.

Leruez-Ville M, Foulon I, Pass R, Ville Y.

ABSTRACT: Cytomegalovirus is the most common congenital infection, affecting 0.5-2% of all live births and the main nongenetic cause of congenital sensorineural hearing loss and neurological damage. Congenital cytomegalovirus can follow maternal primary infection or nonprimary infection. Sensorineurological morbidity is confined to the first trimester with up to 40-50% of infected neonates developing sequelae after first-trimester primary infection. Serological testing before 14 weeks is critical to identify primary infection within 3 months around conception but is not informative in women already immune before pregnancy. In Europe and the United States, primary infection in the first trimester are mainly seen in young parous women with a previous child younger than 3 years. Congenital cytomegalovirus should be evoked on prenatal ultrasound when the fetus is small for gestation and shows echogenic bowel, effusions, or any cerebral anomaly. Although the sensitivity of routine ultrasound in predicting neonatal symptoms is around 25%, serial targeted ultrasound and magnetic resonance imaging of known infected fetuses show greater than 95% sensitivity for brain anomalies. Fetal diagnosis is done by amniocentesis from 17 weeks. Prevention consists of both parents avoiding contact with body fluids from infected individuals, especially toddlers, from before conception until 14 weeks. Candidate vaccines failed to provide more than 75% protection for >2 years in preventing cytomegalovirus infection. Medical therapies such as cytomegalovirus hyperimmune globulins aim to reduce the risk of vertical transmission but 2 randomized controlled trials have not found any benefit. Valaciclovir given from the diagnosis of primary infection up to amniocentesis decreased vertical transmission rates from 29.8% to 11.1% in the treatment group in a randomized controlled trial of 90 pregnant women. In a phase II open-label trial, oral valaciclovir (8 g/d) given to pregnant women with a mildly symptomatic fetus was associated with a higher chance of delivering an asymptomatic neonate (82%), compared with an untreated historical cohort (43%). Valganciclovir given to symptomatic neonates is likely to improve hearing and neurological symptoms, the extent of which and the duration of treatment are still debated. In conclusion, congenital cytomegalovirus infection is a public health challenge. In view of recent knowledge on diagnosis and pre- and postnatal management, health care providers should reevaluate screening programs in early pregnancy and at birth.

Lang Speech Hear Serv Sch. 2020 Jan 8;51(1):98-102. doi: 10.1044/2019_LSHSS-19-00075. Epub 2020 Jan 8.

Where Do We Go From Here? Some Messages to Take Forward Regarding Children With Mild Bilateral and Unilateral Hearing Loss.

Lewis DE.

ABSTRACT: This epilogue discusses messages that we can take forward from the articles in the forum. A common theme throughout the forum is the ongoing need for research. The forum begins with evidence of potential progressive hearing loss in infants with mild bilateral hearing loss, who may be missed by current newborn hearing screening protocols, and supports the need for consensus regarding early identification in this population. Consensus regarding management similarly is a continuing need. Three studies add to the growing body of evidence that children with mild bilateral or unilateral hearing loss are at risk for difficulties in speech understanding in adverse environments, as well as delays in language and cognition, and that difficulties may persist beyond early childhood. Ambivalence regarding if and when children with mild bilateral or unilateral hearing loss should be fitted with personal amplification also impacts management decisions. Two articles address current evidence and support the need for further research into factors influencing decisions regarding amplification in these populations. A third article examines new criteria to determine hearing aid candidacy in children with mild hearing loss. The final contribution in this forum discusses listening-related fatigue in children with unilateral hearing loss. The absence of research specific to this population is evidence for the need for further investigation. Ongoing research that addresses difficulties experienced by children with mild bilateral and unilateral hearing loss and potential management options can help guide us toward interventions that are specific for the needs of these children.

Zhonghua Er Bi Yan Hou Tou Jing Wai Ke Za Zhi. 2019 Dec 7;54(12):881-887. doi: 10.3760/cma.j.issn.1673-0860.2019.12.001. [Article in Chinese]

A follow-up study of abnormal mutation in neonatal deafness gene screening.

Liu QM, Tian Y, Yu JJ, He QQ, Peng L, Guo XQ, Li DY, Chen T.

OBJECTIVE: To screen, diagnose and follow up the abnormal mutation in the gene screening of neonatal deafness.
METHODS: A total of 24161 newborns born in Zhuhai Maternal and Child Health Hospital from February 1, 2015 to January 31, 2008 were screened for hearing and deafness genes, and audiological screening, diagnosis and 1-3 years follow-up were carried out for the newborns with positive gene screening.

RESULTS: There were 991 cases of deafness gene mutation (533 males and 458 females), and the rate of abnormal mutation was 4.10%(991/24 161). Among them, 921 cases were single heterozygous mutation, 130 cases were failed in primary hearing screening, 11 cases were failed in secondary hearing screening, 8 cases were abnormal in audiological diagnosis finally. In these 8 cases, 3 were diagnosed as otitis media and passed audiological follow-up after cure, 2 cases of single ear sensorineural injury caused by high-risk factors, passed after close audiological follow-up, and the other 3 cases were closely audiological follow-up while none of them were successfully sequenced. All of them were moderate to severe sensorineural deafness, 1 case was heterozygous mutation at 3 loci of *GJB2*(c.235delC,c.408C>A,c.134G>A), 1 case was heterozygous mutation at 2 loci of *GJB2*(c.235delC, c.109G>A), and 1 case was single heterozygous mutation of *GJB2*(c.235delC). The remaining 913 cases who passed the primary screening, secondary screening or hearing diagnosis were followed up for 1 to 3 years. Three cases of multiple heterozygous mutation were found in gene screening(2 cases were *SLC26A4* 2168A>G, IVS7-2A>G, 1 case was *GJB2* c.176_191del 16bp, c.299_300del AT), all of them passed both primary and secondary hearing screening. In these 3 cases, the final audiological diagnosis was moderate sensorineural deafness in both ears, with no improvement in the follow-up of 1-3 years. There were 9 monogenic homozygous mutations, 7 failed in primary hearing screening, 3 failed in secondary hearing screening and also failed in audiological diagnosis and 1-3 years' audiological follow-up, all of whom were *GJB2* c.235 del C homozygous mutations, and one of whom had a definite family history of deafness. The remaining 6 cases of homozygous mutation diagnosed by primary screening, secondary screening or hearing diagnosis were *GJB2* c109G>A homozygous mutation, and passed the 1-3 years' hearing follow-up. 58 children with mtDNA mutations, including 2 with 12S rRNA 1494C>T homozygous mutation, 47 with 1555A>G homozygous mutation, and 9 with 1555A>G heterozygous mutation, all passed the primary or secondary hearing screening, and were instructed to ban ototoxic drugs for the whole life, and passed the 1-3 years' hearing follow-up.
CONCLUSIONS: The audiological follow-up of children with monogenic heterozygous mutations in deafness gene screening is generally normal. In case of abnormality, the influencing factors such as otitis media should be excluded at first. In case of unexplained moderate to severe sensorineural deafness, the whole-gene sequencing should be performed to find possible pathogenic factors. The children with homozygous mutation or compound heterozygous mutation in gene screening, most of whom show different degrees of hearing loss, should be followed up for a long time, and provide parents with scientific and reasonable genetic counseling according to the mutation genes and loci,. The hearing of drug-induced deafness gene carriers is normal after birth. Parents should be advised to strengthen prevention and follow-up is generally enough.

Int J Pediatr Otorhinolaryngol. 2020 Mar 12;133:109999. doi: 10.1016/j.ijporl.2020.109999.

Implementation of auricular malformation screenings in the newborn population.

Liu YC, Kini S, Barton G, Pham T, Marcet-Gonzalez J, Novak B.

BACKGROUND AND OBJECTIVE: Research has shown that it is important to initiate ear molding early for children with auricular malformations in order to achieve the best results. Currently our institute relies on the traditional primary care physician (PCP) referral system, which does not recognize the time sensitivity of the visit in patients with auricular malformations. The purpose of the current research is to implement a new screening protocol for identifying auricular malformations in the newborn population and thus expedite the clinic visit and necessary intervention.
METHODS: The hearing screen technicians (HSTs) were trained to identify some of the most common auricular malformations. A picture guide of 11 types of auricular malformations were given to the HSTs to use as a reference. At the time of the newborn hearing screen, the HSTs examined the pinnas of each baby. When an auricular malformation was identified, the auricular malformation team was immediately alerted and a bedside consultation with ENT occurred.
RESULTS: Comparison was made of the referral rate between pre- and post-implementation of the protocol which showed an increased rate of identification (five referrals in the 12-month period pre-implementation versus eighteen referrals in the 15-month period post-implementation).
CONCLUSION: We successfully implemented an auricular malformation screening protocol that was linked to newborn hearing screenings. The frequency of identification has increased with the implementation of the new screening protocol and has resulted in earlier initial ENT consultations for ear molding with the goal of improving patient satisfaction and results.

Cleft Palate Craniofac J. 2020 Feb 4:1055665619899743. doi: 10.1177/1055665619899743.

Parental Judgement of Hearing Loss in Infants With Cleft Palate.

McAndrew L.

OBJECTIVE: To investigate whether reported parental concern is supported by hearing assessment findings in children with cleft palate. To describe this population by examining the relationship between cleft type, middle ear status, and hearing loss.
DESIGN: Retrospective consecutive case note review.

SETTING: Tertiary institutional regional cleft center.
PATIENTS: Consecutive cases of 194 babies born with cleft palate and referred to the specialist center from January 2009 and December 2013. Following exclusions, data from 155 infants were included for analysis.
INTERVENTIONS: Documented parental concern in ear, nose and throat (ENT) and speech and language therapy case notes were compared to hearing assessment findings. Findings from otoscopic examination, tympanometry, and hearing assessment were analyzed with respect to cleft type.
RESULTS: Parental concern is not always accurately reflected by objective assessment particularly when no concern is reported. Analysis of the cohort examined suggests that cleft type is not related to middle ear findings or hearing.
CONCLUSIONS: It is helpful to be aware of parental concern and clinicians should consider that parental reports may not be accurately reflected by test results. As cleft type was not found to substantially influence middle ear status or hearing it is not recommended to adapt speech and language advice offered to families according to cleft type. Follow-up studies to increase participant numbers would support a statistical analysis.

Ear Hear. 2020 Feb 12. doi: 10.1097/AUD.0000000000000829. [Epub ahead of print]
Prelinguistic Vocal Development in Children With Cochlear Implants: A Systematic Review.
McDaniel J, Gifford RH.

OBJECTIVES: This systematic review is designed to (a) describe measures used to quantify vocal development in pediatric cochlear implant (CI) users, (b) synthesize the evidence on prelinguistic vocal development in young children before and after cochlear implantation, and (c) analyze the application of the current evidence for evaluating change in vocal development before and after cochlear implantation for young children. Investigations of prelinguistic vocal development after cochlear implantation are only beginning to uncover the expected course of prelinguistic vocal development in children with CIs and what factors influence that course, which varies substantially across pediatric CI users. A deeper understanding of prelinguistic vocal development will improve professionals' abilities to determine whether a child with a CI is exhibiting sufficient progress soon after implantation and to adjust intervention as needed.
DESIGN: We systematically searched PubMed, ProQuest, and CINAHL databases for primary reports of children who received a CI before 5 years 0 months of age that included at least one measure of nonword, nonvegetative vocalizations. We also completed supplementary searches.
RESULTS: Of the 1916 identified records, 59 met inclusion criteria. The included records included 1125 total participants, which came from 36 unique samples. Records included a median of 8 participants and rarely included children with disabilities other than hearing loss. Nearly all of the records met criteria for level 3 for quality of evidence on a scale of 1 (highest) to 4 (lowest). Records utilized a wide variety of vocalization measures but often incorporated features related to canonical babbling. The limited evidence from pediatric CI candidates before implantation suggests that they are likely to exhibit deficits in canonical syllables, a critical vocal development skill, and phonetic inventory size. Following cochlear implantation, multiple studies report similar patterns of growth, but faster rates producing canonical syllables in children with CIs than peers with comparable durations of robust hearing. However, caution is warranted because these demonstrated vocal development skills still occur at older chronological ages for children with CIs than chronological age peers with typical hearing.
CONCLUSIONS: Despite including a relatively large number of records, the evidence in this review regarding changes in vocal development before and after cochlear implantation in young children remains limited. A deeper understanding of when prelinguistic skills are expected to develop, factors that explain deviation from that course, and the long-term impacts of variations in vocal prelinguistic development is needed. The diverse and dynamic nature of the relatively small population of pediatric CI users as well as relatively new vocal development measures present challenges for documenting and predicting vocal development in pediatric CI users before and after cochlear implantation. Synthesizing results across multiple institutions and completing rigorous studies with theoretically motivated, falsifiable research questions will address a number of challenges for understanding prelinguistic vocal development in children with CIs and its relations with other current and future skills. Clinical implications include the need to measure prelinguistic vocalizations regularly and systematically to inform intervention planning.

Int J Pediatr Otorhinolaryngol. 2020 Jan 22;132:109900. doi: 10.1016/j.ijporl.2020.109900.
Adherence to follow-up recommendations for babies at risk for pediatric hearing loss.
McInerney M, Scheperle R, Zeitlin W, Bodkin K, Uhl B.

OBJECTIVE: The purpose of this retrospective study was to evaluate the families' compliance with recommendations for continued monitoring of babies with high-risk factors for hearing loss.
METHODS: Hearing screening and follow-up results from 604 babies were tracked across a five-year period. Bivariate analysis, including chi-square analysis, t-tests, and one-way analyses of variance were conducted to test whether various factors predicted likelihood of follow up.

RESULTS: Although 86% of the babies returned for the initial follow-up appointment, few completed the protocol or were diagnosed with hearing loss (10.3%). Excluding the babies who never returned, the average age for initial assessment was near the recommended 3-month target (3.5 months). However, babies were last seen at 9.4 months on average, which is earlier than recommended. Some factors positively predicted follow-up: receipt of ototoxic medication, hyperbilirubinemia requiring transfusion, ECMO, syndromes associated with hearing loss, craniofacial anomalies, and passing the newborn hearing screening. Others were negatively predictive: NICU stay >5 days, younger maternal age, and failing the newborn screening. There was no relationship between the results of the last test and whether the families continued with monitoring. Babies with risks categorized as more likely to be associated with delayed onset hearing loss were more often late to the initial follow up, but also followed up for a longer period of time.
CONCLUSIONS: These results demonstrate the need to focus on the barriers unique to babies with risk factors for late onset/progressive hearing loss in addition to those barriers that generally affect loss to follow up. Tools for parental engagement are recommended.

Am J Audiol. 2019 Dec 16;28(4):823-833. doi: 10.1044/2019_AJA-19-0047. Epub 2019 Nov 5.

Using Visual Supports to Facilitate Audiological Testing for Children With Autism Spectrum Disorder.

McTee HM, Mood D, Fredrickson T, Thrasher A, Bonino AY.

PURPOSE: One in 59 children is diagnosed with autism spectrum disorder (ASD). Due to overlapping symptoms between hearing loss and ASD, children who are suspected of having ASD require an audiological evaluation to determine their hearing status for the purpose of differential diagnosis. The purpose of this article is twofold: (a) to increase audiologists' knowledge of ASD by discussing the challenges associated with testing and interpreting clinical data for children with ASD or suspected ASD and (b) to provide visual supports that can be used to facilitate audiological assessment.
METHOD: Eight children (ages 4-12 years) were recruited as video model participants. Videos were filmed using scripts that used concise and concrete language while portraying common clinical procedures. Using the video models, corresponding visual schedules were also created.
CONCLUSION: Although obtaining reliable hearing data from children with ASD is challenging, incorporating visual supports may facilitate testing. Video models and visual schedules have been created and made freely available for download online under a Creative Commons License (Creative Commons-Attribution-NonCommercial-ShareAlike 4.0 International License). Incorporating visual supports during clinical testing has the potential to reduce the child's and family's stress, as well as to increase the probability of obtaining a reliable and comprehensive audiological evaluation. Future research is warranted to determine the effectiveness and feasibility of implementing these tools in audiology clinics.

Gait Posture. 2020 Mar;77:144-155. doi: 10.1016/j.gaitpost.2020.02.001. Epub 2020 Feb 3.

Does the practice of sports or recreational activities improve the balance and gait of children and adolescents with sensorineural hearing loss? A systematic review.

Melo RS, Tavares-Netto AR, Delgado A, Wiesiolek CC, Ferraz KM, Belian RB.

BACKGROUND: Balance and gait disorders have been observed in children and adolescents with sensorineural hearing loss (SNHL), justified by vestibular dysfunctions that these children may present, due to the injury to the inner ear. Therefore, some investigations have suggested that the practice of sports or recreational activities can improve the balance and gait of this population.
OBJECTIVE: Assess the evidence quality from randomized or quasi-randomized controlled trials that used sports or recreational activities as an intervention to improve the balance and /or gait of children and/or adolescents with SNHL.
METHODS: Systematic review that surveyed articles in nine databases, published up to January 10, 2019, in any language, using the following inclusion criteria: (1) Randomized or quasi-randomized controlled trials. (2) Participants from both groups with the clinical diagnosis of SNHL, aged 6-19 years old, without physical problems, cognitive or neurological deficits, except the vestibular dysfunction. (3) Using the practice of sports or recreational activities as an intervention, to improve the balance and/or gait outcomes.
RESULTS: 4732 articles were identified in the searches, after the removal of the duplicates articles and the reading of the titles and their abstracts, remained 16 articles for reading in full, being 5 trials eligible for this systematic review. Of the five eligible trials, three used sports activities and two recreational activities as intervention and presented very low-quality evidence for balance and gait outcomes.
SIGNIFICANCE: Sports and recreational practices seem to represent promising modalities to improve the balance and gait of children and adolescents with SNHL. However, due to the methodological limitations of the trials and the low quality of the current evidence on the topic, the results of the trials should be interpreted with caution. Due to the low quality of evidence observed, we suggest that new trials be proposed on this topic, with

greater methodological rigor, to provide high-quality evidence on the effectiveness of sports and recreational practices to improve the balance and gait of children and adolescents with SNHL.

MMWR Morb Mortal Wkly Rep. 2020 Mar 20;69(11):303-306. doi: 10.15585/mmwr.mm6911a6.

Delayed Identification of Infants Who Are Deaf or Hard of Hearing - Minnesota, 2012-2016.

Meyer AC, Marsolek M, Brown N, Coverstone K.

ABSTRACT: Few studies have examined factors associated with the timing of identification of hearing loss within a cohort of infants identified as deaf or hard of hearing (DHH) and what factors are associated with delayed identification. Minnesota Early Hearing Detection and Intervention (EHDI) personnel studied deidentified data from 729 infants with confirmed congenital hearing loss (i.e., hearing loss identification after not passing newborn hearing screening) born in Minnesota during 2012-2016. Differences in likelihood of delayed identification of congenital hearing loss (defined as not passing newborn hearing screening and age >3 months at the time of identification as DHH) based on multiple variables were analyzed. Overall, 222 (30.4%) infants identified as DHH had delayed identification. Multivariate regression showed that infants identified as DHH were significantly more likely to have delayed identification if they had 1) low birthweight, 2) public insurance, 3) a residence outside the metropolitan area, 4) a mother with a lower level of education, 5) a mother aged <25 years, or 6) a mother who was Hmong. Despite achievements of EHDI programs, disparities exist in timely identification of hearing loss. Using this information to develop public health initiatives that target certain populations could improve timely identification, reduce the risk for language delay, and enhance outcomes in children who are DHH.

J Perinat Med. 2020 Mar 26;48(3):234-241. doi: 10.1515/jpm-2019-0331.

Prenatal findings, neonatal symptoms and neurodevelopmental outcome of congenital cytomegalovirus infection in a university hospital in Montreal, Quebec.

Minsart AF, Rypens F, Smiljkovic M, Kakkar F, Renaud C, Lamarre V, Boucher M, Boucoiran I.

BACKGROUND: Outcome of congenital cytomegalovirus (cCMV) infection in the absence of routine CMV screening and third-trimester scan in North America is scarcely documented. The aim of this study was to assess the severe outcomes related to cCMV according to the indication for screening.

METHODS: This was a retrospective study of 84 mother-child pairs followed for cCMV between 2003 and 2017 at CHU Sainte-Justine in Montreal, Canada. Prenatal ultrasound, neonatal symptoms, neuroimaging and severe outcomes (cerebral palsy, severe cognitive impairment, bilateral hearing loss or neonatal death) were reviewed.

RESULTS: Among 38 cases with abnormal prenatal ultrasound, 41.9% of live-born infants developed severe outcomes. Sixteen (42.1%) were detected in the third trimester. Among 16 cases diagnosed prenatally because of maternal history, all had normal prenatal ultrasound, and none developed severe outcomes. Among cases diagnosed postnatally because of neonatal symptoms, 25% developed severe outcomes. All infants who developed severe outcomes had moderate/severe neonatal symptoms.

CONCLUSION: Outcome of cCMV infection varies according to the reason for screening and timing of diagnosis. Any prenatal ultrasound anomaly might indicate a risk of severe outcome, and warrants a detailed ultrasound scan. However, late detection, or postnatal diagnosis, represented more than half of the cases, and awareness of this will help ensuring optimal management.

Am J Perinatol. 2020 Apr 10. doi: 10.1055/s-0040-1709467.

Auditory Brainstem Evoked Response Patterns in the Neonatal Intensive Care Unit.

Mohammed ST, El-Farrash RA, Taha HM, Moustafa OA.

OBJECTIVE: Delayed maturation of auditory brainstem pathway in neonates admitted to the neonatal intensive care unit (NICU) may lead to misdiagnosis of children with normal peripheral hearing and inappropriate use of amplification devices. The aim of this study is to determine the pattern of auditory brain stem response in neonates admitted to the NICU for proper hearing assessment in this high-risk population.

STUDY DESIGN: This prospective study was conducted on 1,469 infants who were admitted to the NICU, of which 1,423 had one or more risk factors for permanent congenital hearing loss and were screened with automated auditory brain stem response (AABR). A total of 60 infants were referred for diagnostic ABR analysis after failure on AABR screening. The control group comprised 60 well-baby nursery neonates with no risk factors for PCHL.

RESULTS: Mean values of absolute latencies of waves III and V; interpeak latencies I-III, III-V, and I-V; amplitude of waves I, and V; and I/V amplitude ratio at 90 dBnHL measured for the right and left ears at 1 and 3 months of age show significant difference in NICU neonates compared with controls ($p < 0.05$). All the diagnostic ABR measurements significantly improved at the age of 3 months ($p < 0.001$) except wave I absolute latency of both groups ($p > 0.05$). Significant correlations were found between ABR readings at the age of 1 and 3 months and the gestational age of the NICU neonates ($p < 0.05$).

CONCLUSION: Diagnostic ABR findings in NICU neonates suggested delayed maturation of the auditory brainstem pathway with a great impact of gestational age on this maturation. Auditory maturational changes were observed at 3 months of age of patient and control groups.

J Infect Dis. 2020 Mar 5;221(Supplement_1):S15-S22. doi: 10.1093/infdis/jiz443.

Natural History of Congenital Cytomegalovirus Infection in Highly Seropositive Populations.

Mussi-Pinhata MM, Yamamoto AY.

ABSTRACT: Maternal preconceptional cytomegalovirus (CMV) immunity does not protect the fetus from acquiring congenital CMV infection (cCMV). Nonprimary infections due to recurrence of latent infections or reinfection with new virus strains during pregnancy can result in fetal infection. Because the prevalence of cCMV increases with increasing maternal CMV seroprevalence, the vast majority of the cases of cCMV throughout the world follow nonprimary maternal infections and is more common in individuals of lower socioeconomic background. Horizontal exposures to persons shedding virus in bodily secretions (young children, sexual activity, household crowding, low income) probably increase the risk of acquisition of an exogenous nonprimary CMV infection and fetal transmission. In addition, more frequent acquisition of new antibody reactivities in transmitter mothers suggest that maternal reinfection by new viral strains could be a major source of congenital infection in such populations. However, the exact frequency of CMV nonprimary infection in seroimmune women during pregnancy and the rate of intrauterine transmission in these women are yet to be defined. Usually, the birth prevalence of cCMV is high (≥7:1000) in highly seropositive populations. There is increasing evidence that the frequency and severity of the clinical and laboratory abnormalities in infants with congenital CMV infection born to mothers with nonprimary CMV infection are similar to infants born after a primary maternal infection. This is particularly true for sensorineural hearing loss, which contributes to one third of all early-onset hearing loss in seropositive populations. This brief overview will discuss the need for more research to better clarify the natural history of cCMV in highly seropositive populations, which, in almost all populations, remains incompletely defined.

Iran J Otorhinolaryngol. 2020 Jan;32(108):3-10. doi: 10.22038/ijorl.2019.37313.2219.

Comparison of the Pediatric Cochlear Implantation Using Round Window and Cochleostomy.

Naderpour M, Aminzadeh Z, Jabbari Moghaddam Y, Pourshiri B, Ariafar A, Akhondi A.

INTRODUCTION: Cochlear implantation (CI) is now regarded as a standard treatment for children with severe to profound sensor neural hearing loss. This study aimed to compare the efficacy of the round window approach (RWA) and standard cochleostomy approach (SCA) in the preservation of residual hearing after CI in pediatric patients.

MATERIALS AND METHODS: This double-blind randomized controlled trial was conducted on 97 pediatric patients receiving CI with 12-month follow-up. The study population was divided into two groups according to the surgical approaches they received, including RWA and SCA. Consequently, the patients were evaluated based on the Categories of Auditory Performance scale (CAP) and Speech Intelligibility Rating (SIR) test 45-60 days and 3, 6, 9, and 12 months post-surgery.

RESULTS: The CAP and SIR mean scores increased in both groups during the 12-month follow-up. This upward trend was significant in both groups ($P<0.001$). There was no significant difference between the two treatment groups in any of the follow-up stages regarding the CAP mean score. The mean SIR score ($P=1.14\pm0.40$) was significantly higher in the RWA group 3($P=0.001$), 6($P=0.008$), and 9($P=0.006$) months after the surgery. However, there was no significant difference between the RWA and SCA groups, regarding 1-year SIR ($P=0.258$).

CONCLUSION: The CI with either RWA or SCA could improve hearing and speech performance in pediatric patients. Although mid-term speech intelligibility was better for RWA, there was no significant difference in the 1-year outcome between these two methods.

PLoS One. 2020 Jan 9;15(1):e0227143. doi: 10.1371/journal.pone.0227143. eCollection 2020.

Characterization of a universal screening approach for congenital CMV infection based on a highly-sensitive, quantitative, multiplex real-time PCR assay.

Nagel A, Dimitrakopoulou E, Teig N, Kern P, Lücke T, Michna D, Korn K, Steininger P, Shahada K, Neumann K, Überla K.

ABSTRACT: The majority of congenital cytomegalovirus (cCMV) infections are asymptomatic at birth and therefore not diagnosed. Approximately 10-15% of these infants develop late-onset hearing loss and other developmental disorders. Implementation of a universal screening approach at birth may allow early initiation of symptomatic interventions due to a closer follow-up of infants at risk and offers the opportunity to consider treatment of late-onset disease. Real-time PCR assays for the detection of CMV DNA in buccal swab samples demonstrated feasibility and good clinical sensitivity in comparison to a rapid culture screening assay. Because most cCMV infections remain asymptomatic, a universal screening assay that stratifies CMV infected infants

according to low and high risk of late-onset cCMV disease could limit the parental anxiety and reduce follow-up costs. We therefore developed and characterized a screening algorithm based on a highly-sensitive quantitative real-time PCR assay that is compatible with centralized testing of samples from universal screening and allows to determine CMV DNA load of saliva samples either as International Units (IU)/ml saliva or IU/105 cell equivalents. 18 of 34 saliva samples of newborns that tested positively by the screening algorithm were confirmed by detection of CMV DNA in blood and/or urine samples obtained during the first weeks of life. All screening samples that could not be confirmed had viral loads of <2.3x105 IU/ml saliva (median: 6.8x103) or 1.3x105 IU/105 cell equivalents (median: 4.0x102). The viral load of screening samples with confirmed cCMV infection ranged from 7.5x102 to 8.2x109 IU/ml saliva (median: 9.3x107) or 1.5x102 to 5.6x1010 IU/105 cell equivalents (median: 3.5x106). Clinical follow-up of these newborns with confirmed cCMV infection should reveal whether the risk of late-onset cCMV disease correlates with CMV DNA load in early life saliva samples and whether a cut-off can be defined identifying cCMV infected infants with or without risk for late-onset cCMV disease.

Otolaryngol Head Neck Surg. 2020 Mar;162(3):319-321. doi: 10.1177/0194599819900492. Epub 2020 Jan 21.

Letters to the Deaf: Present-Day Relevance of History's Earliest Social Analysis of Deafness.

Naples J, Valdez TA.

ABSTRACT: Harriet Martineau was a 19th-century sociologist who had a progressive form of deafness. Her 1834 essay, *Letters to the Deaf*, was the earliest historical document depicting the social challenges of hearing loss. Martineau details complex situations that hard-of-hearing people experienced in the 19th century such as social isolation due to frustrations with communication, physician shortcomings, limited music appreciation, and the stigma of hearing amplification devices. Her descriptions of these experiences are commonly faced by hard-of-hearing people in present-day society. Advancements in technology and recognition of the negative social impact of hearing loss have improved the social experience for the hard of hearing; however, social challenges remain relevant. In this article, we review *Letters to the Deaf* and note the ways in which this essay provides a dual perspective regarding how much we have advanced as a society and how much we still have to overcome in addressing the social challenges of hearing loss.

Acta Otorrinolaringol Esp. 2020 Jan - Feb;71(1):45-55. doi: 10.1016/j.otorri.2018.09.004. Epub 2018 Dec 19.

Early diagnosis and treatment of unilateral or asymmetrical hearing loss in children: CODEPEH recommendations.

Núñez-Batalla F, Jáudenes-Casaubón C, Sequí-Canet JM, Vivanco-Allende A, Zubicaray-Ugarteche J.

ABSTRACT: The aim of this document is to improve the management and the treatment of unilateral or asymmetrical hearing loss in children. One in one thousand newborn infants has unilateral hearing loss and this prevalence increases with age, due to cases of acquired and delayed-onset hearing loss. Although the impact on the development and learning processes of children of these kinds of hearing loss have usually been minimized, if they are not treated they will impact on language and speech development, as well as overall development, affecting the quality of life of the child and his/her family. The outcomes of the review are expressed as recommendations aimed at clinical diagnosis and therapeutic improvement for unilateral or asymmetrical hearing loss.

Acta Med Port. 2019 Dec 2;32(12):767-775. doi: 10.20344/amp.11880. Epub 2019 Dec 2. [Article in Portuguese]

Congenital or Early Acquired Deafness: An Overview of the Portuguese Situation, from Diagnosis to Follow-Up.

Oliveira C, Machado M, Zenha R, Azevedo L, Monteiro L, Bicho A.

INTRODUCTION: Congenital deafness or early acquired deafness affects 1 to 3 out of 1000 newborns without risk factors and 20 to 40 out of 1000 newborns with risk factors. The universal newborn hearing screening enables its early identification. Children with congenital deafness/early acquired deafness have a higher prevalence of other conditions, especially ophthalmologic and neurodevelopmental ones, and at least 30% to 40% have at least one associated comorbidity.

MATERIAL AND METHODS: We carried out a cross-sectional, multicenter study in which 83% (n = 30) of the hospitals/maternity hospitals of the National Health Service participated.

RESULTS: All surveyed hospitals/maternity hospitals routinely performed universal newborn hearing screening to all newborns before discharge; 63% referred children with risk factors for hearing loss to Otorhinolaryngology. All children with congenital deafness/early acquired deafness are referred to: Pediatrics in 23% hospitals/maternity hospitals. In 23 hospitals/maternity hospitals, all children with congenital deafness/early acquired deafness are referred to: Speech Therapy in 44% hospitals/ maternity hospitals; Ophthalmology in 17% hospitals/maternity hospitals; National System of Early Intervention in Childhood in 30% hospitals/maternity hospitals; 22% of hospitals/maternity hospitals refer all children with congenital deafness/early acquired deafness, with no identified cause, to Clinical Genetics clinics. The number of diagnoses of deafness in the years 2014 and 2015 was 2.5 and

1.5 per 1000 newborns, respectively, in 15 hospitals/maternity hospitals.
DISCUSSION: Awareness of universal newborn hearing screening seems to be widely spread in the National Health Service. The number of children with SC / SPA, as well as the percentage of different types of deafness diagnosed, were identical to those found in other studies and shows its importance. The assessment / follow-up of these children by specialties other than the otolaryngology was heterogeneous in different health entities and revealed that not all children with risk factors for deafness follow up advised by existing standards.
CONCLUSION: Results show that Portugal made an important path in the screening and follow-up of children with SC / SPA. It is important, with the ultimate aim of continually improving the care of these children, to reflect on the involvement of specialties other than otolaryngology, such as the National Early Childhood Intervention System in the follow-up of these children.

Indian J Otolaryngol Head Neck Surg. 2020 Mar;72(1):30-35. doi: 10.1007/s12070-019-01723-w. Epub 2019 Jul 31.

A Prospective Study on Temporal Bone Involvement in Polytrauma Patients and the Effect of Early Diagnosis on Hearing Loss.

Padmakumar V, Ramesh Kumar E, Ramakrishnan VR.

ABSTRACT: As polytrauma cases are on the rise, a large number of patients presents with temporal bone fractures, which can result in various types of injuries varying from trivial to more serious injuries. Early diagnosis and appropriate management in required in case of serious injuries for a better outcome. The aim of my study is to study the incidence, the different injuries occurring and the effect of early diagnosis on hearing loss. Patients coming to our emergency department with polytrauma are studied and clinically evaluated for any temporal bone injuries. Based on the type of injuries audiological and radiological studies are done. And if required, biochemical tests like CSF analysis will be done. Also hearing assessment will be done as early as possible and appropriate treatment required will be started. The outcome is then assessed and followed up on a regular basis. In our study there were 90 patients with temporal bone fracture out of the 2748 polytrauma cases. The incidence was calculated to be 32 per 1000 cases. 69 patients (76.7%) had longitudinal fracture of temporal bone; 13 patients (14.4%) had transverse fracture; 2 patients (2.2%) had oblique fractures and 6 patients (6.6%) had comminuted fractures. Hearing loss was found to be the most common injury seen in 56 patients (62.2%). Of which 30 (53.5%) had conductive hearing loss (CHL); 9 (16%) had sensorineural hearing loss (SNHL); 17 had mixed hearing loss (MHL). 27 (90%) out of 30 patients with CHL showed improvement in hearing. Out of the 26 patients with SNHL and MHL, 22 patients (84.61%) showed improvement. 5 out of 6 with immediate onset facial palsy and 6 out of 8 with late onset facial palsy showed improvement. The hearing outcome in our study was found to be much better than the previous year which shows that the difference might be due to the early diagnosis and management. In our study hearing improvement was noted in most patients with hearing loss when compared to the previous year, which may have been due to the detection of the injuries at the earliest and managing the same with appropriate treatment modalities.

Mol Genet Genomic Med. 2020 Feb 17:e1171. doi: 10.1002/mgg3.1171.

Analyses of del(GJB6-D13S1830) and del(GJB6-D13S1834) deletions in a large cohort with hearing loss: Caveats to interpretation of molecular test results in multiplex families.

Pandya A[1], O'Brien A[1], Kovasala M[1], Bademci G[2], Tekin M[2], Arnos KS[3].

BACKGROUND: Mutations involving the closely linked GJB2 and GJB6 at the DFNB1 locus are a common genetic cause of profound congenital hearing loss in many populations. In some deaf GJB2 heterozygotes, a 309 kb deletion involving the GJB6 has been found to be the cause for hearing loss when inherited in trans to a GJB2 mutation.
METHODS: We screened 2,376 probands from a National DNA Repository of deaf individuals.
RESULTS: Fifty-two of 318 heterozygous probands with pathogenic GJB2 sequence variants had a GJB6 deletion. Additionally, eight probands had an isolated heterozygous GJB6 deletion that did not explain their hearing loss. In two deaf subjects, including one proband, a homozygous GJB6 deletion was the cause for their hearing loss, a rare occurrence not reported to date.
CONCLUSION: This study represents the largest US cohort of deaf individuals harboring GJB2 and GJB6 variants, including unique subsets of families with deaf parents. Testing additional members to clarify the phase of GJB2/GJB6 variants in multiplex families was crucial in interpreting clinical significance of the variants in the proband. It highlights the importance of determining the phase of GJB2/GJB6 variants when interpreting molecular test results especially in multiplex families with assortative mating.

BMC Infect Dis. 2020 Mar 12;20(1):217. doi: 10.1186/s12879-020-4941-z.

Diagnosing congenital Cytomegalovirus infection: don't get rid of dried blood spots.

Pellegrinelli L, Alberti L, Pariani E, Barbi M, Binda S.

BACKGROUND: Congenital Cytomegalovirus (cCMV) is a serious global public health issue that can cause irreversible fetal and neonatal congenital defects in symptomatic or asymptomatic newborns at birth. In absence of universal cCMV screening, the retrospective diagnosis of cCMV infection in children is only possible by examining Dried Blood Spot (DBS) samples routinely collected at birth and stored for different time spans depending on the newborn screening regulations in force in different countries. In this article, we summarize the arguments in favor of long-term DBS sample storage for detecting cCMV infection.

MAIN TEXT: CMV infection is the most common cause of congenital infection resulting in severe defects and anomalies that can be apparent at birth or develop in early childhood. Sensorineural hearing loss is the most frequent consequence of cCMV infection and may have a late onset and progress in the first years of life. The virological diagnosis of cCMV is essential for clinical research and public health practices. In fact, in order to assess the natural history of CMV infection and distinguish between congenital or acquired infection, children should be diagnosed early by analyzing biological samples collected in the first weeks of life (3 weeks by using viral culture and 2 weeks by molecular assays), which, unfortunately, are not always available for asymptomatic or mildly symptomatic children. It now seems possible to overcome this problem since the CMV-DNA present in the blood of congenitally infected newborns can be easily retrieved from the DBS samples on the Guthrie cards routinely collected and stored within 3 days from birth in the neonatal screening program for genetic and congenital diseases. Early collection and long-term storage are inexpensive methods for long-term bio-banking and are the key points of DBS testing for the detection of cCMV.

CONCLUSION: DBS sampling is a reliable and inexpensive method for long-term bio-banking, which enables to diagnose known infectious diseases - including cCMV - as well as diseases not jet recognized, therefore their storage sites and long-term storage conditions and durations should be the subject of political decision-making.

Laryngoscope. 2020 Jan 27. doi: 10.1002/lary.28536.

Genetic Testing for Congenital Bilateral Hearing Loss in the Context of Targeted Cytomegalovirus Screening.

Peterson J, Nishimura C, Smith RJH.

OBJECTIVES/HYPOTHESIS: To determine the prevalence of children with genetic hearing loss who are cytomegalovirus (CMV) positive at birth and the relative proportion of genetic and CMV etiology among children with congenital bilateral hearing loss.

STUDY DESIGN: Database review.

METHODS: We performed a review of clinical test results for patients undergoing comprehensive genetic testing for all known hearing loss-associated genes from January 2012 to January 2019. This population was reviewed for reported CMV status and genetic causes of congenital bilateral hearing loss.

RESULTS: In the OtoSCOPE database, 61/4,282 patients were found to have a documented CMV status, and 661/4282 had documented bilateral congenital hearing loss. Two patients were identified who had both a positive CMV result and a genetic cause for their hearing loss. Forty-eight percent of patients with bilateral congenital hearing loss (320/661) were found to have a genetic etiology. In 62% (198/320), the hearing loss was associated with pathogenic variants in GJB2, STRC, SLC26A4 or an Usher syndrome-associated gene.

CONCLUSIONS: We estimate that ~2% of CMV-positive newborns with hearing loss have a known genetic variant as a cause. The subcohort of CMV-positive newborns with symmetric mild-to-moderate bilateral hearing loss will have at least a 7% chance of having pathogenic gene variants associated with hearing loss. In a CMV-positive neonate who failed their newborn hearing screen bilaterally, genetic screening needs to be considered for accurate diagnosis and possible deferment of antiviral treatment.

J Pediatr Hematol Oncol. 2020 Jan;42(1):e25-e31. doi: 10.1097/MPH.0000000000001637.

Platinum-drugs Ototoxicity in Pediatric Patients With Brain Tumors: A 10-Year Review.

Rabiço-Costa D, Gil-da-Costa MJ, Barbosa JP, Bom-Sucesso M, Spratley J.

PURPOSE: Platinum-derived chemotherapy is one of the cornerstones in the treatment of central nervous system tumors in children. We aimed to assess the incidence of hearing loss in children after the exposure to platinum drugs.

MATERIAL AND METHODS: Retrospective study of prospectively collected data on children consecutively diagnosed with brain tumors and treated with platinum derivatives at a tertiary referral hospital between January 2006 and December 2015. We analyzed multiples variables, such as: age at diagnosis, tumor location, hydrocephalus, platinum drug type, radiotherapy, and follow-up time. The final sample size was 51 patients.

RESULTS: The median age at diagnosis was 6 years. The median overall follow-up time was 75 months. The incidence of ototoxicity was 23.5%. Rates of hearing loss with carboplatinum were lower than with cisplatinum. A statistically significant association occurred between the presence of hydrocephalus, radiotherapy exposure, infratentorial tumor location, and ototoxicity after treatment with platinum derivatives.

CONCLUSIONS: Childhood central nervous system tumors nowadays exhibit improved cure and survival rates. However, the ototoxicity resulting from the chemotherapy treatment may accompany patients for the rest of their lives. This study reveals that this occurrence is not negligible, and the association of radiotherapy and the presence of hydrocephalus can be potentiating factors.

Am J Med Genet B Neuropsychiatr Genet. 2020 Apr;183(3):172-180. doi: 10.1002/ajmg.b.32774. Epub 2019 Dec 19.

Identification of TMC1 as a relatively common cause for nonsyndromic hearing loss in the Saudi population.

Ramzan K, Al-Owain M, Al-Numair NS, Afzal S, Al-Ageel S, Al-Amer S, Al-Baik L, Al-Otaibi GF, Hashem A, Al-Mashharawi E, Basit S, Al-Mazroea AH, Softah A, Sogaty S, Imtiaz F.

ABSTRACT: Hearing loss (HL) is the most common sensory disorder worldwide and genetic factors contribute to approximately half of congenital HL cases. HL is subject to extensive genetic heterogeneity, rendering molecular diagnosis difficult. Mutations of the transmembrane channel-like 1 (TMC1) gene cause hearing defects in humans and mice. The precise function of TMC1 protein in the inner ear is unknown, although it is predicted to be involved in functional maturation of cochlear hair cells. TMC1 mutations result in autosomal recessive (DFNB7/11) and sometimes dominant (DFNA36) nonsyndromic HL. Mutations in TMC1 are responsible for a significant portion of HL, particularly in consanguineous populations. To evaluate the importance of TMC1 mutations in the Saudi population, we used a combination of autozygome-guided candidate gene mutation analysis and targeted next generation sequencing in 366 families with HL previously shown to lack mutations in GJB2. We identified 12 families that carried five causative TMC1 mutations; including three novel (c.362+3A>G; c.758C>T [p.Ser253Phe]; c.1396_1398delACC [p.Asn466del]) and two reported mutations (c.100C>T [p.Arg34Ter]; c.1714G>A [p.Asp572Asn]). Each of the identified recessive mutation was classified as severe, by both age of onset and severity of HL. Similarly, consistent with the previously reported dominant variant p.Asp572Asn, the HL phenotype was progressive. Eight families in our cohort were found to share the pathogenic p.Arg34Ter mutation and linkage disequilibrium was observed between p.Arg34Ter and SNPs investigated. Our results indicate that TMC1 mutations account for about 3.3% (12/366) of Saudi HL cases and that the recurrent TMC1 mutation p.Arg34Ter is likely to be a founder mutation.

Wiley Interdiscip Rev Syst Biol Med. 2020 Mar;12(2):e1469. doi: 10.1002/wsbm.1469. Epub 2019 Dec 4.

Structural neuroimaging of the altered brain stemming from pediatric and adolescent hearing loss-Scientific and clinical challenges.

Ratnanather JT.

ABSTRACT: There has been a spurt in structural neuroimaging studies of the effect of hearing loss on the brain. Specifically, magnetic resonance imaging (MRI) and diffusion tensor imaging (DTI) technologies provide an opportunity to quantify changes in gray and white matter structures at the macroscopic scale. To date, there have been 32 MRI and 23 DTI studies that have analyzed structural differences accruing from pre- or peri-lingual pediatric hearing loss with congenital or early onset etiology and postlingual hearing loss in pre-to-late adolescence. Additionally, there have been 15 prospective clinical structural neuroimaging studies of children and adolescents being evaluated for cochlear implants. The results of the 70 studies are summarized in two figures and three tables. Plastic changes in the brain are seen to be multifocal rather than diffuse, that is, differences are consistent across regions implicated in the hearing, speech and language networks regardless of modes of communication and amplification. Structures in that play an important role in cognition are affected to a lesser extent. A limitation of these studies is the emphasis on volumetric measures and on homogeneous groups of subjects with hearing loss. It is suggested that additional measures of morphometry and connectivity could contribute to a greater understanding of the effect of hearing loss on the brain. Then an interpretation of the observed macroscopic structural differences is given. This is followed by discussion of how structural imaging can be combined with functional imaging to provide biomarkers for longitudinal tracking of amplification.

J Infect Dis. 2020 Mar 5;221(Supplement_1):S74-S85. doi: 10.1093/infdis/jiz601.

Clinical Diagnostic Testing for Human Cytomegalovirus Infections.

Razonable RR, Inoue N, Pinninti SG, Boppana SB, Lazzarotto T, Gabrielli L, Simonazzi G, Pellett PE, Schmid DS.

ABSTRACT: Human cytomegalovirus (HCMV) infections are among the most common complications arising in transplant patients, elevating the risk of various complications including loss of graft and death. HCMV infections are also responsible for more congenital infections worldwide than any other agent. Congenital HCMV (cCMV) infections are the leading nongenetic cause of sensorineural hearing loss and a source of significant neurological disabilities in children. While there is overlap in the clinical and laboratory approaches to diagnosis of HCMV infections in these settings, the management, follow-up, treatment, and diagnostic strategies differ considerably.

As yet, no country has implemented a universal screening program for cCMV. Here, we summarize the issues, limitations, and application of diagnostic strategies for transplant recipients and congenital infection, including examples of screening programs for congenital HCMV that have been implemented at several centers in Japan, Italy, and the United States.

Am J Otolaryngol. 2020 Mar - Apr;41(2):102372. doi: 10.1016/j.amjoto.2019.102372. Epub 2019 Dec 10.

Impact of cochlear abnormalities on hearing outcomes for children with cochlear implants.

Ronner E, Basonbul R, Bhakta R, Mankarious L, Lee DJ, Cohen MS.

OBJECTIVE: Evaluate the impact of cochlear anomalies on hearing outcomes for pediatric patients with cochlear implants.
STUDY DESIGN: Retrospective chart review.
SETTING: Tertiary care center.
SUBJECTS AND METHODS: Charts were retrospectively reviewed for cases where pediatric cochlear implant surgery was performed between 2002 and 2018 at a single, tertiary care institution. Patients were divided into groups based on the presence or absence of radiological cochlear abnormalities, which were further classified as low or high risk anomalies. Hearing outcomes were evaluated by measuring pure tone averages and word recognition scores preoperatively, 3 and 12 months postoperatively, in addition to the most recent test results.
RESULTS: There were 154 ears implanted in our cohort of 100 patients. 107 ears had normal cochlear anatomy, 31 had low risk, and 16 had high risk abnormalities. The most common modality of preoperative imaging was CT scan. Postoperative mean pure tone average (PTA) was significantly higher in patients with inner ear anomalies compared to those with normal anatomy. No significant difference in PTA was noted between low versus high risk patients. <50% of patients had word recognition scores available within the first year following surgery.
CONCLUSION: Abnormalities of the inner ear significantly influenced hearing outcomes over time following cochlear implant surgery when compared to pediatric patients with normal anatomy. Obtaining hearing testing can be difficult in very young children and therefore future studies are warranted to further investigate the impact that cochlear abnormalities may have on hearing outcomes following cochlear implant surgery.

Int J Pediatr Otorhinolaryngol. 2020 Apr;131:109870. doi: 10.1016/j.ijporl.2020.109870. Epub 2020 Jan 10.

Outcomes of regional-based newborn hearing screening for 35,461 newborns for 5 years in Akita, Japan.

Sato T, Nakazawa M, Takahashi S, Mizuno T, Ishikawa K, Yamada T.

OBJECTIVES: Newborn hearing screening (NHS) has been actively performed since 2001 in Akita, Japan. The NHS coverage rate has increased yearly, and performance has been consistently >90% since 2012. The purpose of this study was to summarize NHS outcomes in the Akita prefecture of Japan and to obtain new insights for from our summarized data for the future.
METHODS: A total of 35,461 newborns in hospitals and clinics where hearing screening was performed in Akita from 2012 to 2016 were included. The outcome data of NHS were collected for analysis.
RESULTS: The overall screening coverage rate for hearing loss was 94.7%. Of the screened infants, 0.53% received a referral on the 2-stage automated auditory brainstem response (ABR), and 80.4% of referred infants had a check-up at the hospital to receive a diagnostic hearing examination. Finally, the prevalence of bilateral congenital hearing loss was 0.14%, that of bilateral moderate to profound hearing loss was 0.12%, and that of unilateral congenital hearing loss was 0.10%. Furthermore, the average consultation period in infants with risk factors was significantly later than that in infants without risk factors ($p = 0.0015$). Follow-up for infants diagnosed with normal hearing after diagnostic hearing examination revealed that 4.7% suffered bilateral moderate to profound hearing loss later. This percentage is significantly higher than that of the general group ($p < 0.001$).
CONCLUSION: The prevalence of bilateral congenital hearing loss was 0.14% in Akita and 0.12% of infants were diagnosed with bilateral moderate to severe hearing loss. Medical personnel should be enlightened regarding the importance of performing hearing diagnostic examinations until 3 months of age. Even if infants were diagnosed with normal hearing after a diagnostic examination, we strongly suggest continuing follow-up until they are able to perform pure tone audiometry with accuracy.

HNO. 2020 Mar 5. doi: 10.1007/s00106-020-00825-0. [Epub ahead of print] [Article in German]

Guideline: Auditory processing and perception disorders: Proposal for treatment and management of APD : S1 guideline of the German Society of Phoniatrics and Pediatric Audiology.

Schönweiler R, Kiese-Himmel C, Plotz K, Nickisch A, Am Zehnhoff-Dinnesen A.

ABSTRACT: Despite normal hearing thresholds in pure-tone audiometry, 0.5-1% of children have difficulty understanding what they hear. An auditory processing disorder (APD) can be assumed, which should be clarified

and treated. In patients with hearing loss, this must first be compensated or resolved. Only hereafter can a suspected APD be confirmed or excluded. Diagnosis of APD requires that a clear discrepancy between the child's performance in individual auditory functions and other cognitive abilities be demonstrated. Combination of therapeutical modalities is considered particularly more beneficial in APD patients than a single modality. Treatment modalities should consider linguistic and cognitive processes (top-down), e.g., metacognitive knowledge of learning strategies or vocabulary expansion, but also address underlying auditory deficits (bottom-up). Almost 50% of children with APD also have a language development disorder requiring treatment and/or dyslexia. Therefore, each therapeutic intervention for a child with APD must be individually adapted according to the diagnosed impairments. Musical training can improve phonologic and reading abilities. Changes and adaptations in the classroom are helpful to support the weak auditory system of children with APD. Architectural planning of classrooms can be a means of ensuring that direct sound is masked by as little diffuse sound as possible. For example, acoustic ceiling tiles are suitable for reducing reverberant and diffuse sound.

Int J Pediatr Otorhinolaryngol. 2020 Mar 9;133:109984. doi: 10.1016/j.ijporl.2020.109984.

Hearing and speech benefits of cochlear implantation in children: A review of the literature.

Sharma SD, Cushing SL, Papsin BC, Gordon KA.

ABSTRACT: Cochlear implantation is a safe and reliable treatment for children with severe to profound hearing loss. The primary benefit of these medical devices in children is the acquisition of hearing, which promotes development of spoken language. The present paper reviews published literature demonstrating predictive effects of a number of factors on acquisition of hearing development and speech recognition. Of the many variables that contribute to an individual child's development after implantation, age at implantation, the presence of medical comorbidities, social determinants of health, and the provision of bilateral versus unilateral hearing are those that can vary widely and have consistently shown clear impacts. Specifically, age of implantation is crucial to reduce effects of deafness on the developing auditory system and capture the remarkable plasticity of early development. Language development after cochlear implantation requires therapy emphasizing hearing and oral communication, education, and other support which can be influenced by known social determinants of health; specifically, outcomes in children decline with reductions in socioeconomic status and levels of parental education. Medical co-morbidities also slow rates of progress after cochlear implantation. On the other hand, benefits of implantation increase in children who are provided with access to hearing from both ears. In sum, cochlear implants promote development of hearing in children and the best outcomes are achieved by providing early access to sound in both ears. These benefits can be limited by known social determinants of health which restrict access to needed support and medical comorbidities which add further complexity in care and outcome.

Int J Pediatr Otorhinolaryngol. 2020 Apr;131:109881. doi: 10.1016/j.ijporl.2020.109881. Epub 2020 Jan 16.

Comprehensive hearing care network for early identification and intervention in children with congenital and late-onset/acquired hearing loss: 8 years' experience in Miyazaki.

Shirane M, Ganaha A, Nakashima T, Shimoara S, Yasunaga T, Ichihara S, Kageyama S, Matsuda Y, Tono T.

OBJECTIVE: In 2010, we established the Miyazaki Comprehensive Hearing Care Network (MCHCN) for early identification and intervention in children with congenital and late-onset/acquired hearing loss with the cooperation of related administrative bodies in Miyazaki prefecture. The central roles of the MCHCN program are played by the Hearing Care Center (HCC) at the University of Miyazaki Hospital established in 2010 to facilitate audiological diagnoses, hearing aid interventions, and educational efforts, as well as linkage with the Department of Otolaryngology for surgical interventions. Herein, we aimed to present the main outcomes of the MCHCN program organized by the HCC at the University of Miyazaki Hospital.

METHODS: The MCHCN consists of two different networks, the Newborn Hearing Screening Network (NHSN) and the Pediatric Hearing Care Network (PHCN). All children suspected of having hearing loss by Newborn Hearing Screening (NHS) are referred to the HCC via the NHSN. In addition, children suspected of late-onset/acquired hearing loss by municipality-led health checkups, pediatricians, public health nurses, and childcare workers are referred to the HCC via the PHCN. Children who were born in Miyazaki prefecture between January 2010 and December 2017 and referred to the HCC for detailed hearing examination were included in this study.

RESULTS: Within the study period, 89,390 infants were born in Miyazaki prefecture, and 84,737 (94.9%) of them underwent NHS. A total of 698 infants and 182 children with suspected hearing loss were referred to the HCC via the NHSN and PHCN, respectively. Of the 880 referrals, 169 were diagnosed with hearing loss, which included 80 children with bilateral hearing loss and 89 children with unilateral hearing loss. Of the 80 children with bilateral hearing loss, 76 began wearing hearing aids and 15 had cochlear implants in the follow-up period. In children with bilateral conductive hearing loss, 4 children with bilateral middle ear anomalies underwent ossiculoplasty, following which two of these children no longer required hearing aids. Imaging assessments performed on 71 of the 89 children with unilateral hearing loss revealed that 20 of the 30 (66%) children who underwent CT exhibited

ossicular anomalies and 28 out of the 48 (58%) children who underwent MRI were found to have ipsilateral cochlear nerve hypoplasia. Among the 169 children with hearing loss, no follow-up loss was observed during the period of this study.

CONCLUSION: The MCHCN that was organized at the initiative of the HCC at the University of Miyazaki Hospital has enabled the provision of comprehensive and continuous support, ranging from diagnosis to intervention, not only for children with suspected hearing loss referred based on their NHS results but also for those who pass the screening. Via this system, children with late-onset/acquired hearing loss can be identified early and can receive medical interventions tailored to the cause of their hearing loss while simultaneously avoiding a loss to follow-up.

Int J Pediatr Otorhinolaryngol. 2020 Mar 6;133:109983. doi: 10.1016/j.ijporl.2020.109983.

Endoscopic findings and long-term hearing results for pediatric unilateral conductive hearing loss.

Silvola JT.

OBJECTIVES: Analyze reasons for unilateral conductive hearing loss (CHL) with unknown etiology in children.

INTRODUCTION: Unilateral conductive hearing loss (HL) without known etiology can be undiagnosed despite of hearing screening programs. It can be difficult to find the reason for HL and to make a treatment plan. Middle ear endoscopy gives hard-evidence diagnosis and basis for an individual treatment plan.

METHODS AND MATERIAL: Prospective clinical follow-up study for a cohort of generally healthy elementary school age children with unilateral conductive HL with unknown etiology. The study population was 192 children, of which 46 had a HL of at least 25 dB with more than 10 dB conductive component. Mean age was 8.7 years. Preoperative tests included otomicroscopy, bone- and air-conduction audiogram, tympanometry, stapes reflex tests, Rinne and Weber test and Otoacoustic emissions. The children underwent endoscopy of the middle ear with an individual treatment plan and long-term follow-up. The aim was to explore etiology and to give a treatment plan for hearing loss. Follow-up included air- and bone conduction hearing tests annually or every other year. Mean follow-up was 5.2 years.

RESULTS: A clear etiological finding was found in 36 (78%) ears, stapes anomaly (23) as the most common (64%) finding. Other findings were two cholesteatomas, 2 status after trauma, 5 middle ear anomalies, 5 incus fixations and one incus erosion. Air conduction hearing improved spontaneously during follow-up in 81% (17/21, 2 dropouts) of the stapes anomaly ears (mean 11,3 dB, range 4-32 dB), and none of these ears showed hearing deterioration. In the incus fixation group, one ear showed hearing deterioration. There were no major complications for exploration, and 5 minor postoperative infections.

CONCLUSIONS: The most common reason for pediatric unilateral conductive hearing loss was stapes anomaly/fixation. The HL does not deteriorate. Hearing loss in stapes anomalies shows a tendency for spontaneous recovery. Stapes surgery can be postponed or avoided.

Aust J Prim Health. 2020 Jan 20. doi: 10.1071/PY18162.

Developmental vulnerability of Australian school-entry children with hearing loss.

Simpson A, Šarkić B, Enticott JC, Richardson Z, Buck K.

ABSTRACT: National data from the Australian Early Development Census (AEDC) was used to describe the sociodemographic and developmental characteristics of a cohort of Australian children entering their first year of primary school in 2012. Results, together with sociodemographic variables were reported for two groups: children with and without reported hearing loss. Data on 285232 children were analysed, with just over 1% of these children identified with hearing loss. Logistic regression analysis found that children with reported hearing loss had over double the odds than their hearing peers of being developmentally 'vulnerable' on one or more domains of the AEDC. Covariates of interest included Aboriginal and Torres Strait Islander heritage, as well as high rates of school absenteeism. Retrospective longitudinal research linking developmental outcomes with intervention efforts, such as newborn hearing screening, would be beneficial in future research.

Am J Otolaryngol. 2020 Jan 10:102398. doi: 10.1016/j.amjoto.2020.102398.

The effect of passive smoking on the etiology of serous otitis media in children.

Tarhun YM.

ABSTRACT: Serous otitis media (SOM) is a disease mostly seen in the pediatric age group and characterized by serous effusion in the middle ear. The disease which is mostly silent can cause permanent hearing loss if it is not diagnosed and treated early. Passive smoking is one of the environmental factors in the etiopathology of the disease and risk factors for SOM formation in children. In our study, smoking habits of family members of 75 children with SOM and 50 healthy controls were investigated. At the end of the study, the correlation between SOM and passive smoke exposed was statistically significant in children ($p < 0.01$). In this study, the effect of passive smoking, which is a preventable and controllable risk factor in the etiology of the SOM in children is emphasized.

BMC Infect Dis. 2020 Mar 17;20(1):225. doi: 10.1186/s12879-020-4950-y.

Bacterial otitis media in sub-Saharan Africa: a systematic review and meta-analysis.

Tesfa T, Mitiku H, Sisay M, Weldegebreal F, Ataro Z, Motbaynor B, Marami D, Teklemariam Z.

BACKGROUND: Otitis media is inflammation of the middle ear, comprising a spectrum of diseases. It is the commonest episode of infection in children, which often occurs after an acute upper respiratory tract infection. Otitis media is ranked as the second most important cause of hearing loss and the fifth global burden of disease with a higher incidence in developing worlds like Sub-Saharan Africa and South Asia. Therefore, this systematic review is aimed to quantitatively estimate the current status of bacterial otitis media, bacterial etiology and their susceptibility profile in sub-Saharan Africa.

METHODS: A literature search was conducted from major databases and indexing services including EMBASE (Ovid interface), PubMed/MEDLINE, Google Scholar, ScienceDirect, Cochrane Library, WHO African Index-Medicus and others. All studies (published and unpublished) addressing the prevalence of otitis media and clinical isolates conducted in sub-Saharan Africa were included. Format prepared in Microsoft Excel was used to extract the data and data was exported to Stata version 15 software for the analyses. Der-Simonian-Laird random-effects model at a 95% confidence level was used for pooled estimation of outcomes. The degree of heterogeneity was presented with I^2 statistics. Publication bias was presented with funnel plots of standard error supplemented by Begg's and Egger's tests. The study protocol is registered on PROSPERO with reference number ID: CRD42018102485 and the published methodology is available from http://www.crd.york.ac.uk/CRD42018102485.

RESULTS: A total of 33 studies with 6034 patients were included in this study. All studies have collected ear swab/discharge samples for bacterial isolation. The pooled isolation rate of bacterial agents from the CSOM subgroup was 98%, patients with otitis media subgroup 87% and pediatric otitis media 86%. A univariate meta-regression analysis indicated the type of otitis media was a possible source of heterogeneity (p-value = 0.001). The commonest isolates were P. aeruginosa (23-25%), S. aureus (18-27%), Proteus species (11-19%) and Klebsiella species. High level of resistance was observed against Ampicillin, Amoxicillin-clavulanate, Cotrimoxazole, Amoxicillin, and Cefuroxime.

CONCLUSION: The analysis revealed that bacterial pathogens like P. aeruginosa and S. aureus are majorly responsible for otitis media in sub-Saharan Africa. The isolates have a high level of resistance to commonly used drugs for the management of otitis media.

Otolaryngol Head Neck Surg. 2020 Jan;162(1):114-120. doi: 10.1177/0194599819880348. Epub 2019 Oct 8.

Should You Follow the Better-Hearing Ear for Congenital Cytomegalovirus Infection and Isolated Sensorineural Hearing Loss?

Torrecillas V, Allen CM, Greene T, Park A, Chung W, Lanzieri TM, Demmler-Harrison G.

OBJECTIVE: To describe the progression of sensorineural hearing loss (SNHL) in the better- and poorer-hearing ears in children with asymptomatic congenital cytomegalovirus (CMV) infection with isolated SNHL. **STUDY DESIGN:** Longitudinal prospective cohort study.

SETTING: Tertiary medical center.

SUBJECTS AND METHODS: We analyzed hearing thresholds of the better- and poorer-hearing ears of 16 CMV-infected patients with isolated congenital/early-onset or delayed-onset SNHL identified through hospital-based CMV screening of >30,000 newborns from 1982 to 1992.

RESULTS: By 12 months of age, 4 of 7 patients with congenital/early-onset SNHL developed worsening thresholds in the poorer-hearing ear, and 1 had an improvement in the better-hearing ear. By 18 years of age, all 7 patients had worsening thresholds in the poorer-hearing ear and 3 patients had worsening thresholds in the better-hearing ear. plHearing loss first worsened at a mean age of 2 and 6 years in the poorer- and better-hearing ears, respectively. Nine patients were diagnosed with delayed-onset SNHL (mean age of 9 years vs 12 years for the poorer- and better-hearing ears), 6 of whom had worsening thresholds in the poorer-hearing ear and 1 in both ears.

CONCLUSION: In most children with congenital CMV infection and isolated SNHL, the poorer-hearing ear worsened earlier and more precipitously than the better-hearing ear. This study suggests that monitoring individual hearing thresholds in both ears is important for appropriate interventions and future evaluation of efficacy of antiviral treatment.

Eur Arch Otorhinolaryngol. 2020 Mar 27. doi: 10.1007/s00405-020-05935-7.

Assessment of temporal processing functions in early period cochlear implantation.

Tuz D, Aslan F, Böke B, Yücel E.

PURPOSE: The purpose of this study is to compare the temporal processing performance of children with cochlear implant (CI) according to the age of implantation and to determine their relation with auditory perception scores.

METHODS: In this study, 30 cochlear implant users and ten normal hearing children at 9 and 10 years were included. Children with cochlear implants are divided into two groups according to the age of implantation: group I includes participants whose implantation age is between 13 and 35 months (20 children), group II includes

participants whose implantation age is between 36 and 45 months (10 children). Individuals were evaluated with random gap detection test (RGDT), duration pattern test (DPT), frequency pattern test (FPT), the Mr. Potato Head task, word recognition, and sentence recognition test.
RESULTS: A significant difference was found between the control and CI groups in temporal processing performance. The temporal processing ability of CI groups was significantly worse than those of normal hearing. Although there was no significant difference among the groups with cochlear implant in terms of temporal processing performance, children who started to use CI at an earlier age showed a tendency of better performance on temporal processing tasks. There was a significant relationship between Daily Sentence Test and FPT, and the Mr. Potato Head task and FPT rev (the score calculated by accepting the reverse patterns correctly). There was a significant relationship between duration of implant use and temporal ordering performance
CONCLUSION: In this study, children with CI cannot perform as well as normal-hearing peers on temporal processing tasks, even if they had started to use their CIs at an early age. It is important to evaluate temporal processing in implanted individuals and to guide auditory training considering the evaluation results.

Am J Otolaryngol. 2020 Mar - Apr;41(2):102379. doi: 10.1016/j.amjoto.2019.102379. Epub 2019 Dec 19.

Experience with cholesteatoma behind an intact tympanic membrane in children.

Urík M, Kaliariková A, Machač J, Jurajda M.

INTRODUCTION: To systematically investigate all surgeries for cholesteatoma behind an intact tympanic membrane at our department. To identify predictive factors that can help the surgeon to plan surgery, surgical techniques, and follow-up treatment.
MATERIAL AND METHODS: This retrospective study evaluates 21 child patients, who were operated in the period 2007-2017 on for cholesteatoma behind an intact tympanic membrane.
RESULTS: A total of 202 primary operations were performed for cholesteatoma. In 21 cases (10,4%) there was a cholesteatoma behind an intact tympanic membrane and in 11 (5,45%) cases of it there was the congenital cholesteatoma. The most frequently affected area was the anterior-superior quadrant. The preoperative hearing loss increased significantly with disease severity (I-IV by Potsic).
CONCLUSIONS: The classification system according to Potsic is sufficient and fully corresponds to the surgeon's needs. It has been clearly shown that a higher CC stage is associated with worse postoperative hearing results.

Cochrane Database Syst Rev. 2020 Jan 21;1:CD010885. doi: 10.1002/14651858.CD010885.pub5.

Different infusion durations for preventing platinum-induced hearing loss in children with cancer.

van As JW, van den Berg H, van Dalen EC.

BACKGROUND: Platinum-based therapy, including cisplatin, carboplatin or oxaliplatin, or a combination of these, is used to treat a variety of paediatric malignancies. Unfortunately, one of the most important adverse effects is the occurrence of hearing loss or ototoxicity. In an effort to prevent this ototoxicity, different platinum infusion durations have been studied. This review is the third update of a previously published Cochrane Review.
OBJECTIVES: To assess the effects of different durations of platinum infusion to prevent hearing loss or tinnitus, or both, in children with cancer. Secondary objectives were to assess possible effects of these infusion durations on: a) anti-tumour efficacy of platinum-based therapy, b) adverse effects other than hearing loss or tinnitus, and c) quality of life.
SEARCH METHODS: We searched the electronic databases Cochrane Central Register of Controlled Trials (CENTRAL; the Cochrane Library 14 November 2019), MEDLINE (PubMed) (1945 to 14 November 2019) and Embase (Ovid) (1980 to 14 November 2019). In addition, we handsearched reference lists of relevant articles and we assessed the conference proceedings of the International Society for Paediatric Oncology (2009 up to and including 2019) and the American Society of Pediatric Hematology/Oncology (2014 up to and including 2019). We scanned ClinicalTrials.gov and the World Health Organization International Clinical Trials Registry Platform (WHO ICTRP; apps.who.int/trialsearch) for ongoing trials (both searched on 4 November 2019).
SELECTION CRITERIA: Randomised controlled trials (RCTs) or controlled clinical trials (CCTs) comparing different platinum infusion durations in children with cancer. Only the platinum infusion duration could differ between the treatment groups.
DATA COLLECTION AND ANALYSIS: Two review authors independently performed the study selection, 'Risk of bias' assessment and GRADE assessment of included studies, and data extraction including adverse effects. Analyses were performed according to the guidelines of the Cochrane Handbook for Systematic Reviews of Interventions.
MAIN RESULTS: We identified one RCT and no CCTs; in this update no additional eligible studies were identified. The RCT (total number of children = 91) evaluated the use of a continuous cisplatin infusion (N = 43) versus a one-hour bolus cisplatin infusion (N = 48) in children with neuroblastoma. For the continuous infusion, cisplatin was administered on days one to five of the cycle, but it is unclear if the infusion duration was a total of five days. Risk of bias was present. Only results from shortly after induction therapy were provided. No clear evidence of a

difference in hearing loss (defined as asymptomatic and symptomatic disease combined) between the different infusion durations was identified as results were imprecise (risk ratio (RR) 1.39, 95% confidence interval (CI) 0.47 to 4.13, low-quality evidence). Although the numbers of children were not provided, it was stated that tumour response was equivalent in both treatment arms. With regard to adverse effects other than ototoxicity, we were only able to assess toxic deaths. Again, the confidence interval of the estimated effect was too wide to exclude differences between the treatment groups (RR 1.12, 95% CI 0.07 to 17.31, low-quality evidence). No data were available for the other outcomes of interest (i.e. tinnitus, overall survival, event-free survival and quality of life) or for other (combinations of) infusion durations or other platinum analogues.

AUTHORS' CONCLUSIONS: Since only one eligible RCT evaluating the use of a continuous cisplatin infusion versus a one-hour bolus cisplatin infusion was found, and that had methodological limitations, no definitive conclusions can be made. It should be noted that 'no evidence of effect', as identified in this review, is not the same as 'evidence of no effect'. For other (combinations of) infusion durations and other platinum analogues no eligible studies were identified. More high-quality research is needed.

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Correlation of air-bone gap and size of Enlarged Vestibular Aqueduct in children.

Van Beck J, Chinnadurai S, Morrison AK, Zuniga MG, Smith B, Lohse CM, McCaslin D.

OBJECTIVE: Enlarged vestibular aqueduct (EVA) is an inner ear malformation that represents an important cause of pediatric hearing loss. While certain elements in the history or audiogram may suggest EVA, it is most often diagnosed using computed tomography (CT). The present investigation was conducted to determine if the size of the audiometric air-bone gap (ABG) is correlated with the size of the vestibular aqueduct in the pediatric population using three vestibular aqueduct measurements. These included the fundus, midpoint, and porous widths of the vestibular aqueduct.

STUDY DESIGN: This is a retrospective cohort study.

SETTING: This study took place at a tertiary care referral center.

PATIENTS: Fifty-five children (33 female; 22 male) with a confirmed diagnosis of unilateral or bilateral EVA as determined by prior imaging of the inner ear were included in the study.

MAIN OUTCOME MEASURES: Associations of EVA measurements with ABGs at 0.5 and 1 kHz were evaluated using Pearson correlation coefficients.

RESULTS: All of the correlation coefficients were positive, indicating that as EVA measurements increased so did the ABG. Only the correlation between fundus width and ABG at 1 kHz was not statistically significant.

CONCLUSIONS: ABGs measured during audiometric testing correlate with the size of the EVA and ABGs can be clinical predictors of the severity of the bony abnormality. These data support the third window theory of conductive hearing loss in pediatric EVA.

BMJ Case Rep. 2019 Dec 1;12(11). pii: e231978. doi: 10.1136/bcr-2019-231978.

Neonatal cholestasis, hyperferritinemia, hypoglycemia and deafness: a diagnostic challenge.

van Westering-Kroon E, Heijligers M, Hütten MC.

ABSTRACT: Neonatal conjugated hyperbilirubinemia is a diagnostic challenge. A full term, small for gestational age boy presented with cholestasis, hypoglycemia, hyperferritinemia and severe bilateral deafness. Diagnostic work-up revealed two hereditary diseases: alpha-1-antitrypsin deficiency (PI*ZZ genotype) and autosomal recessive deafness type 3 (compound heterozygous MYO15A gene mutation). In addition, we found late hypoglycemia on full enteral feeding which complicated this case. Hyperferritinemia is an uncommon finding in newborn cholestasis without liver failure.

Lin Chung Er Bi Yan Hou Tou Jing Wai Ke Za Zhi. 2020 Feb;34(2):113-118. doi: 10.13201/j.issn.1001-1781.2020.02.004. [Article in Chinese]

Analysis of genotypes and hearing phenotypes of mutation infants with deafness.

Wang X, Zhao X, Huang L, Wen C, Wang X, Cheng X.

OBJECTIVE: The aim of this study is to explore the genotype and hearing phenotype of deaf infants with mutation of *GJB2* gene.

METHOD: Subjects were 121 infants with *GJB2* gene mutations who were treated in the Children's Hearing Diagnosis Center of Beijing Tongren hospital. All subjects were accepted to undertake the universal newborns hearing screening (UNHS) and series of objective audiometry, including auditory brainstem response, distortion product otoacoustic emission, auditory steady-state response and other audiological tests. All subjects were screened for nine pathogenic variants in four genes or all exons of the *GJB2* gene, and then were diagnosed as infants with *GJB2* gene mutations. Initially, analyzing their genotypes and hearing phenotypes generally. Then, the subjects were divided into two groups according to the genotypes: T/T group (truncated/truncated mutations, 89

cases) and T/NT group (truncated/non-truncated mutations, 32 cases). Chi-square test was used to analyze the results of UNHS, hearing degree, audiogram patterns and symmetry/asymmetry of binaural hearing phenotype. Eventually, analyzing the results of UNHS.

RESULT: The most common truncated mutation was c.235delC (64.88%, 157/242 and the most common non-truncated mutation was c.109G>A (11.16%, 27/242. The homozygous mutation of c.235delC/c.235delC was the dominant in T/T group (38.84%, 47/121, and the compound heterozygous mutation of c.235delC/c.109G>A was the dominant in T/NT group (18.18%, 22/121. 81.82% (99/121 of subjects failed in UNHS, including 74.38% (90/121 with bilateral reference, 7.44% (9/121 with a single pass. The refer rate of UNHS of group T/T and T/NT were 86.52% (77/89 and 68.75%, respectively. There was a statistically significant difference between the two groups (P<0.05. 85.95% (104/121 of subjects were diagnosed as hearing loss and 14.05% (17/121 of subjects were diagnosed as normal hearing. The degree of hearing loss: profound, severe, moderate and mild were 31.40% (38/121, 19.01% (23/121, 24.79% (30/121 and 10.74% (13/121, respectively. There was no subjects with normal hearing in T/T group and individuals with severe and profound hearing loss accounted for the highest proportion (65.17%, 58/89, while in T/NT group, normal hearing accounted for 53.13% (17/32 and mild and moderate hearing loss accounted for the highest proportion (37.5%, 12/32. There was statistically significant difference between the two groups (P<0.05. Of 104 patients (208 ears with hearing loss, the audiogram patterns: flat, descending, ascending, residual, Valley and other types were 49.03% (102/208, 12.02% (25/208, 8.65% (18/208, 7.69% (16/204, 3.36% (7/204 and 19.23% (40/204, respectively. The two most common types in T/T group were flat (47.19%, 84/178 and other types (20.22%, 36/178, while in T/NT group were flat (60.00%, 18/30 and ascending (20.00%, 6/30. There was statistically significant difference between the two groups (P<0.05. There were 50 cases (48.07% with symmetrical hearing phenotype and 54 cases (51.93%) with asymmetrical hearing phenotype. Asymmetry was predominant in T/T group (53.93%, 48/89, and symmetry was predominant in T/NT group (60.00%, 9/15. There was no statistically significant difference between the two groups (P>0.05.

CONCLUSION: In this study, c.235delC/c.235delC homozygous mutation was dominant in T/T group and c.235delC/c.109G>A heterozygous mutation was dominant in T/NT Group. The hearing phenotypes in T/T group were mostly bilateral asymmetric severe hearing loss, and those in T/NT Group were bilateral symmetric mild to moderate hearing loss, special attention should be paid to the audiological characteristics of different genotypes.

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Cognitive and Behavioral Functioning in Hearing-Impaired Children with and without Language Delay.

Williams A, Pulsifer M, Tissera K, Mankarious LA.

ABSTRACT: Poor language development in patients with sensorineural hearing loss (SNHL) may be related to an auditory deficit and/or other neurologic condition that influences the ability to communicate. A retrospective chart review of children (mean age = 4.0 years) with congenital, bilateral SNHL was performed to assess for linguistic and nonlinguistic neurodevelopmental differences between those who were language-impaired (LI) versus non-language-impaired (NLI). Language, neurodevelopmental functioning, and behavior were assessed. Twenty-two patients were identified: 12 were LI and 10 were NLI. Average pure-tone thresholds and nonverbal intelligence were not different between the language groups, but the LI group demonstrated significantly lower median overall adaptive skills, personal living skills, and motor skills. Behavioral dysregulation was significantly higher in the LI versus NLI group (58% vs 10%; P = .031), although the median neurodevelopmental scores did not differ significantly. These findings introduce the possibility that nonlinguistic processing deficit(s) may be confounding the ability to develop language.

Zhonghua Yi Xue Yi Chuan Xue Za Zhi. 2020 Mar 10;37(3):269-276. doi: 10.3760/cma.j.issn.1003-9406.2020.03.008. [Article in Chinese]

Clinical practice guidelines for hereditary non-syndromic deafness.

Writing Group For Practice Guidelines For Diagnosis And Treatment Of Genetic Diseases Medical Genetics Branch Of Chinese Medical Association, Yuan H, Dai P, Liu Y, Yang T.

ABSTRACT: Genetic factors are a common cause for non-syndromic hearing loss (NSHL). Along with the development and maturity of molecular techniques, genetic diagnosis and counseling is increasingly affecting the clinical practice of NSHL. Newborn hearing screening has facilitated early detection of affected children, whilst genetic screening has enabled identification of the cause of NSHL, and genetic diagnosis and consultation can promote early intervention of deafness. So far 110 pathogenic genes of NSHL have been discovered, though there are still many challenges lying in its clinical identification. The development of genetic counseling and prenatal diagnosis has put forward greater requirements for genetic testing and data interpretation. This guideline has summarized the incidence, mutational spectrum, inheritance mode, pathogenesis, clinical manifestation, genotype - phenotype correlation, genetic testing, treatment and intervention, as well as risk assessment for

NSHL, with an aim to provide a reference for genetic consultants, clinical otologists and professionals engaged in genetic testing.

Clin Infect Dis. 2020 Mar 17;70(7):1379-1384. doi: 10.1093/cid/ciz413.

Contribution of Congenital Cytomegalovirus Infection to Permanent Hearing Loss in a Highly Seropositive Population: The Brazilian Cytomegalovirus Hearing and Maternal Secondary Infection Study.

Yamamoto AY, Anastasio ART, Massuda ET, Isaac ML, Manfredi AKS, Cavalcante JMS, Carnevale-Silva A, Fowler KB, Boppana SB, Britt WJ, Mussi-Pinhata MM.

BACKGROUND: The exact contribution of congenital cytomegalovirus infection (cCMVI) to permanent hearing loss (HL) in highly seropositive populations is unknown. We determined the contribution of cCMVI to HL and estimated the effectiveness of newborn hearing screening (HS) in identifying neonates with CMV-related HL.
METHODS: A total of 11 900 neonates born from a population with ≥97% maternal seroprevalence were screened for cCMVI and HL. cCMVI was confirmed by detection of CMV-DNA in saliva and urine at age <3 weeks.
RESULTS: Overall, 68 (0.6%; 95% confidence interval [CI], 0.4-0.7) neonates were identified with cCMVI. Of the 91 (0.8%) newborns who failed the HS, 24 (26.4%) were confirmed with HL, including 7 (29.2%; 95% CI, 17.2-59.3) with cCMVI. Another newborn with cCMVI passed the HS but was confirmed with HL at age 21 days. Of the 62 neonates with cCMVI who underwent a complete hearing evaluation, 8 (12.9%; 95% CI, 6.7-23.4) had HL and most (7/8; 87.5%; 95% CI, 46.6-99.7) were identified by HS. The rate of CMV-related HL was 8 per 11 887 neonates (0.7 per 1000 live births). The prevalence ratio of HL among neonates with cCMVI compared to CMV-uninfected neonates was 89.5 (95% CI, 39.7-202.0). No late-onset cCMVI-related HL was detected during a median follow-up of 36 months.
CONCLUSIONS: cCMVI is an important cause of HL in childhood in all settings. Integrating targeted cCMVI screening among neonates who fail a HS could be a reasonable, cost-effective strategy to identify newborns with early-onset cCMVI-related HL.

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Cost-effectiveness of School Hearing Screening Programs: A Scoping Review.

Yong M, Liang J, Ballreich J, Lea J, Westerberg BD, Emmett SD.

OBJECTIVE: School hearing screening is a public health intervention that can improve care for children who experience hearing loss that is not detected on or develops after newborn screening. However, implementation of school hearing screening is sporadic and supported by mixed evidence to its economic benefit. This scoping review provides a summary of all published cost-effectiveness studies regarding school hearing screening programs globally. At the time of this review, there were no previously published reviews of a similar nature.
DATA SOURCES: A structured search was applied to 4 databases: PubMed (Medline), Embase, CINAHL, and Cochrane Library.
REVIEW METHODS: The database search was carried out by 2 independent researchers, and results were reported in accordance with the PRISMA-ScR checklist and the JBI methodology for scoping reviews. Studies that included a cost analysis of screening programs for school-aged children in the school environment were eligible for inclusion. Studies that involved evaluations of only neonatal or preschool programs were excluded.
RESULTS: Four of the 5 studies that conducted a cost-effectiveness analysis reported that school hearing screening was cost-effective through the calculation of incremental cost-effectiveness ratios (ICERs) via either quality- or disability-adjusted life years. One study reported that a new school hearing screening program dominated the existing program; 2 studies reported ICERs ranging from 1079 to 4304 international dollars; and 1 study reported an ICER of £2445. One study reported that school-entry hearing screening was not cost-effective versus no screening.
CONCLUSION: The majority of studies concluded that school hearing screening was cost-effective. However, significant differences in methodology and region-specific estimates of model inputs limit the generalizability of these findings.

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Parents' satisfaction with a trial of a newborn hearing screening programme in Jordan.

Zaitoun M, Nuseir A.

OBJECTIVE: This study examines parents' satisfaction level toward a trial of a newborn hearing screening programme (NHSP) that was applied in King Abdullah II University Hospital (KAUH) in Jordan over one year. This is the first study that investigated parents' satisfaction toward a hearing screening programme in the Arab countries, and the results will improve any future screening programmes in the Arabian region.
METHOD: The main tool for this study was a questionnaire that was translated and modified from the original

version of the Parental Satisfaction with the Newborn Hearing Screening Programme (PSQ-NHSPs1). The questionnaire consisted of 19 items covering five main aspects of the NHSP. The parents' responses were not anonymously given where the parents whose children had undergone the hearing screening were contacted by phone using the data record of the hospital.

RESULTS: The majority of the parents were very satisfied with the programme overall and showed great support and appreciation for the effort in testing their babies and increasing their awareness. The satisfaction levels varied among the specific aspects of the programme. Good portion of the parents did not receive the brochure containing information about the screening, and almost half of them did not know the results of the hearing screening.

CONCLUSION: Parents were overall satisfied with neonatal hearing screening programme that was conducted at KAUH. However, parents were less satisfied with information related to the test procedure and results. Parents' responses in this study could be used to improve any future hearing screening program in Jordan or in the Arab countries.