

We are pleased to present Volume 3, No.1 of JEHDI. The wide variety of disciplines represented in Early Hearing Detection and Intervention is reflected in the articles we are publishing and the astounding number of downloads of each issue. Our authors, editors, reviewers, and the NCHAM staff have done an exceptional job of assembling relevant and current information on the many topics necessary to accomplish the goals of EHD. We encourage you to share your expertise with this vibrant and exciting community of professionals.

Les R. Schmeltz, AuD  
Editor-in-Chief

# Pediatric Hearing Device Management: Professional Practices for Monitoring Aided Audibility

Karen Muñoz, EdD<sup>1,2</sup>  
Lauri Nelson, PhD<sup>1</sup>  
Kacy Herald, BS<sup>1</sup>

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

**Abstract: Objective.** The purpose of this study was to explore professional practices for monitoring aided audibility for children who are deaf or hard of hearing (DHH).

**Design.** A cross-sectional survey design was used to identify providers' self-reported practice patterns for monitoring aided audibility for children who use hearing aids, cochlear implants, and bone-conduction hearing aids. Three surveys were used.

**Study Sample.** Practicing audiologists, speech-language pathologists, and deaf educators providing services to children birth to six years of age who use hearing technology were recruited to participate. A total of 184 surveys were included in the analysis (96 hearing aid; 47 cochlear implant; 41 bone conduction hearing aid).

**Results.** Practice gaps were identified, including infrequent use of parent questionnaires to explore how children are hearing at home and in other environments, lack of loaner equipment for some children when hearing devices were being repaired, and inconsistent monitoring of data logging to identify challenges with hearing aid use.

**Conclusion.** Children who are DHH and their parents rely on professionals to provide evidence-based practices. This study revealed practice gaps related to monitoring audibility, suggesting opportunities for training to address provider confidence and consistent implementation of monitoring practices.

**Key Words:** hearing device, audibility, monitoring, professional practices

**Acronyms:** BCHA = Bone Conduction Hearing Aids; CI = Cochlear Implants; DHH = Deaf or Hard of Hearing; HA = Hearing Aids

**Correspondence concerning this article should be addressed to:** Karen Muñoz, Utah State University, 2620 Old Main Hill, Logan, UT 84322. Email: [karen.munoz@usu.edu](mailto:karen.munoz@usu.edu)

Hearing loss affects one to six of every 1000 newborns in the United States (American Speech-Language-Hearing Association, n.d.) and approximately 32 million children worldwide (World Health Organization, 2016). In the United States, 98% of newborns receive a hearing screening (Centers for Disease Control and Prevention, 2015), and this early identification offers important developmental advantages by providing opportunities to begin intervention within the first few weeks of life. Early identification coupled with advanced hearing technology and specialized intervention services provides opportunities for children to learn to communicate using spoken language, regardless of the degree of their hearing loss. Early identification, however, also presents challenges for parents and professionals. Most parents of children who are deaf or hard of hearing (DHH) have normal hearing (Mitchell

and Karchmer, 2004), and need complete and accurate information as they make decisions to support their child's development. For spoken language development, access to sound using hearing technology, also described as aided audibility (i.e., hearing aids, cochlear implants, bone-conduction hearing aids) is an important factor. Children have better outcomes when they receive amplification early, have their devices optimally programmed for their hearing loss, and wear their devices consistently (Tomblin, Oleson, Ambrose, Walker, & Moeller, 2014; McCreery, Bentler, & Roush, 2013).

There are parent and professional factors that can influence how consistently children receive adequate audibility. Parents have reported experiencing an array of challenges related to daily management of their child's

hearing needs (e.g., child behavior, parent emotions, lack of confidence) that negatively influence hours of hearing aid use (Muñoz et al., 2016; Caballero et al., 2017; Muñoz, Preston, & Hicken, 2014; Walker et al., 2013). Parents are central to the intervention process and practices to support and educate parents are vital. When professionals engage parents within a therapeutic relationship, they can help manage challenges that arise, so the parents effectively implement skills into their daily life.

In addition to parent education, it is critical that professionals employ practices to ensure children who use hearing technology experience consistent and effective audibility. All professionals do not have the same level of expertise in working with young children who are DHH, and it cannot be assumed that best practices for this population are occurring. For example, McCreery et al. (2013) investigated the proximity of pediatric fittings to prescriptive targets for hearing aid gain and found that more than half of the children had at least one ear that did not meet prescribed targets for audibility. Identifying professionals' practice patterns for monitoring aided audibility can provide insights into gaps and areas that could benefit from a focused approach to training, for both pre-service and continuing education. Therefore, the purpose of this study was to better understand professional practices related to factors that influence audibility for young children using hearing devices, and to explore factors that influence practices for teaching skills to parents.

## Method

This study used a cross-sectional survey design. There were three surveys to explore professional practice patterns for monitoring aided audibility for children who use hearing aids (HA), cochlear implants (CI), and bone-conduction hearing aids (BCHA). Survey responses were anonymous. Institutional Review Board approval was obtained through Utah State University.

### Participants and Procedures

Audiologists, speech-language pathologists, and deaf educators providing services to children who are DHH, birth to six years of age, were recruited to participate in the study February to December 2017. Participants were recruited through websites (i.e., Hear to Learn, Hands & Voices, AG Bell Association, Hearing First) and social media posts. Survey data were collected online using Qualtrics through a link posted on the website; a flyer was used to invite professionals to participate. Two-hundred and twenty-five surveys were submitted. Of those, 41 were incomplete and excluded, resulting in 184 surveys included in the analysis (96 HA; 47 CI; 41 BCHA). There were participants from 33 states and 5 countries. The majority of the participants were audiologists ( $n = 139$ ) and the remainder were intervention professionals, such as speech-language pathologists, teachers, or early interventionists ( $n = 45$ ). Most professionals had more

than 10 years of experience (audiologists [54%,  $n = 75$ ]; interventionists [51%;  $n = 23$ ]), approximately one-third had less than five years of experience (audiologists [32%,  $n = 45$ ]; interventionists [29%;  $n = 13$ ]), and the remainder had six to ten years of experience (audiologists [14%,  $n = 19$ ]; interventionists [20%;  $n = 9$ ]). Ninety-one percent of the audiologists ( $n = 126$ ) and 69% of the interventionists ( $n = 31$ ) reported working in urban areas, with the remainder working in rural areas.

### Survey Instruments

Three survey instruments (HA [17 items]; CI [16 items]; BCHA [16 items]) were developed by the first and second authors. Items were developed based on professional guidelines (e.g., American Academy of Audiology, 2013) in order to capture fundamental practices for hearing technology monitoring. Each survey had three sections: Information About You, Device Use, and Monitoring (instruments available in Appendix).

### Data Analysis

Data were analyzed in SPSS to calculate descriptive statistics to report characteristics of professional practices related to monitoring aided audibility for children who use hearing devices. To observe differences between audiologists and interventionists, data were split and analyzed separately. Participants were not required to answer each item to continue responding to the survey; therefore, the total number of responses per item varies. Percentages are reported based on the number of responses for each item. For survey items that were the same for each survey, regardless of device type (i.e., HA, CI, BCHA), responses were combined. For items that were unique to the device type, responses were reported separately. Item analysis examined practices related to teaching skills to parents that are important regardless of device type, specifically:

- Ask parents about the number of hours their child wears device(s);
- Ask parents about challenges with device use;
- Help parents resolve challenges with device use;
- Talk to parents about data logging results;
- Talk to parents about how to do a speech sound check;
- Talk to parents about difficulties their child may have hearing in different environments;
- Talk to parents about benefits of personal assistive device use in addition to device; and
- Talk to parents about monitoring personal assistive device use.

The result suggested good internal consistency across the items ( $n = 184$ ; Cronbach's  $\alpha = .765$ ). Differences in teaching practices for the eight items listed above were explored using analysis of variance for professional type (i.e., audiologist; interventionist), device type (i.e., HA; CI; BCHA), and years practicing (i.e., less than 10 years; 10 or more years).

## Results

Participants were queried about hearing device use to explore their perceptions about how often children they serve in their practice are using their hearing devices, and the professionals they think should be talking about device use with parents (see Table 1). Participants reported, on average, that approximately two-thirds of the children birth to six years of age whom they serve are using their hearing devices all waking hours; however, only 29% of audiologists and 13% of interventionists reported that they always know hours of device use. The majority of audiologists and interventionists reported that each professional (i.e., audiologist; speech-language pathologist; teacher; early interventionist) should talk with parents about device use. The participants also felt that physicians, deaf mentors, counseling professionals (e.g., social worker), and other families of children who are DHH should talk with parents about device use.

### Monitoring Practices

Participants indicated how often (i.e., *never, sometimes, often, always*) three practices for monitoring aided audibility were provided (see Table 2). Two practices were applicable to all hearing devices: *During audiology appointments, how often is speech understanding tested while children are wearing their hearing device?* and *How often do you ask parents to complete a questionnaire about how their child is responding to sounds in their daily life (e.g., hearing in quiet, hearing in noise)?* One item was applicable to hearing aids: *When the children you work with get new earmolds, how often are hearing aid settings checked to make sure sounds are being appropriately amplified?*

The majority of the audiologists (85%) reported performing aided speech testing *often* or *always*, and 53% of interventionists reported the children they work with receive aided speech testing. Just over one-third of audiologists and fewer than one-fourth of interventionists reported that they ask parents to complete questionnaires *often* or *always* in order to monitor their child's responses to sounds in daily life (audiologists 38%; interventionists 21%). When questionnaires are completed, participants reported using a variety of instruments: LittlEARS (Audiologist 63%; Interventionist 45%); PEACH ([Parent's Evaluation of Aural/Oral Performance of Children]; Audiologist 57%; Interventionist 10%); IT MAIS ([Infant-Toddler Meaningful Auditory Integration Scale]; Audiologist 42%; Interventionist 57%); SIFTER ([Preschool Screening Instrument For Targeting Educational Risk]; Audiologist 30%; Interventionist 29%); CHILD ([Children's Home Inventory for Listening Difficulties]; Audiologist 27%; Interventionist 14%); ELF ([Early Listening Function]; Audiologist 25%; Interventionist 17%); TEACH ([Teachers' Evaluation of Aural/oral performance of Children]; Audiologist 9%; Interventionist 7%); COW ([Children's Outcome Worksheets]; Audiologist 8%; Interventionist 0%). Other questionnaires used (ranging from < 1% to 8%) included: CASLLS (Cottage Acquisition Scales for Listening, Language, and Speech), SSQ (Speech Spatial Qualities), LIFE R (Listening Inventory For Education – Revised), ASC (Auditory Skills Checklist), ALG (Auditory Learning Guide), Starting School LIFE (Starting School Listening Inventory For Education), Sanders Questionnaire, FLI (Functional Listening Index) MAIS (Meaningful Auditory Integration Scale), and MUSS (Meaningful Use of Speech Scale). Participants reported hearing aid settings are checked *often* or *always* when new earmolds are obtained 82% of the time as reported by audiologists and 45% as reported by interventionists.

**Table 1**  
*Provider Perceptions about Hearing Device Use*

	Audiologist N = 141		Interventionist N = 147	
	% (n)	M (SD)	% (n)	M (SD)
Percent of children I serve who use devices all waking hours		64 (25.24)		64 (32.98)
I don't know		9 (.28)		7 (.25)
How often hours of device use known by professional				
Never		1 (2)		0
Sometimes		20 (28)		24 (11)
Often		49 (68)		62 (28)
Always		30 (41)		13 (6)
Professionals you think should talk about device use with parents*				
Audiologist		100 (139)		94 (44)
Speech-language pathologist		94 (130)		84 (38)
Teacher		91 (127)		98 (44)
Early Interventionist		94 (130)		93 (42)
Other		48 (67)		24 (11)

\*More than one answer allowed

**Table 2**  
**How Often Practices for Monitoring Aided Audibility are Provided**

Monitoring Practices	% (n)				
	N	S	O	A	U
<b>All Devices</b>					
Aided speech understanding is tested in audiology appointment					
Audiologist	0	14 (19)	39 (52)	46 (60)	<1 (1)
Interventionist	5 (2)	22 (9)	24 (10)	29 (12)	20 (8)
Parents complete a questionnaire about child's auditory development					
Audiologist	21 (27)	42 (55)	29 (38)	9 (12)	
Interventionist	24 (10)	55 (23)	14 (6)	7 (3)	
<b>Hearing Aid Only</b>					
Hearing aid settings are checked when new ear mold obtained					
Audiologist	3 (2)	10 (7)	42 (28)	40 (27)	5 (93)
Interventionist	0	18 (4)	27 (6)	18 (4)	36 (8)

N = Never, S = Sometimes, O = Often, A = Always, U = I don't know

**Table 3**  
**Loaner Hearing Devices Provided when Child's Device Sent for Repair**

Device Type	% (n)				
	N	S	O	A	U
Hearing Aid	7 (7)	26 (25)	35 (34)	27 (26)	4 (4)
Bone Conduction Hearing Aids	15 (6)	32 (13)	29 (12)	20 (8)	5 (2)
Cochlear Implant	15 (7)	30 (14)	11 (5)	40 (19)	4 (2)

N = Never, S = Sometimes, O = Often, A = Always, U = I don't know

**Table 4**  
**Professional's Level of Confidence**

Level of confidence in knowing how to...	M (SD)	
	Audiologist	Interventionist
<b>Hearing Aids</b>		
Recognize when earmolds need to be replaced	97 (10.15)	83 (14.44)
Verify hearing aid settings are appropriate	95 (14.59)	42 (29.23)
Conduct a speech sound check	98 (5.40)	93 (17.80)
Determine hearing aids are functioning properly	98 (7.64)	88 (11.31)
<b>Bone Conduction Hearing Aid (BCHA)</b>		
Tell when programming adjustments are needed	84 (22.68)	51 (34.50)
Conduct a speech sound check	97 (7.48)	99 (3.78)
Check BCHA function	92 (15.41)	65 (35.05)
<b>Cochlear Implant</b>		
Tell when programming adjustments are needed	93 (8.62)	75 (19.74)
Conduct a speech sound check	99 (3.81)	96 (9.32)
Check cochlear implant function	99 (2.61)	84 (16.81)



**Table 5**  
**Frequency Audiologists and Interventionists Teach Parents Monitoring Skills**

How often the following is addressed with parents...	Audiologists				Interventionists			
	N	S	O	A	N	S	O	A
<b>All Devices</b>								
Ask about the number of hours their child wears hearing device	0	5 (7)	20 (28)	75 (104)	0	16 (7)	47 (21)	38 (17)
Ask about challenges with hearing device use	0	<1 (1)	26 (36)	73 (102)	0	11 (5)	47 (21)	42 (19)
Help parents resolve challenges with hearing device use	<1 (1)	2 (3)	23 (32)	74 (103)	0	7 (3)	44 (20)	49 (22)
Talk about hearing device data logging results	4 (6)	20 (28)	32 (44)	44 (61)	29 (13)	33 (15)	29 (13)	9 (4)
How to do a speech sound check	2 (3)	16 (21)	35 (46)	47 (61)	0	17 (7)	26 (11)	57 (24)
Difficulties child may have hearing in different environments	0	4 (5)	40 (53)	56 (74)	0	17 (7)	36 (15)	48 (20)
Benefits of personal assistive device in addition to hearing device	<1 (1)	8 (11)	38 (50)	53 (70)	5 (2)	21 (9)	38 (16)	36 (15)
Monitoring personal assistive device use	3 (4)	20 (26)	41 (54)	36 (48)	12 (5)	36 (15)	24 (10)	29 (12)
<b>Hearing Aids</b>								
How often earmolds need to be replaced	2 (1)	2 (1)	39 (26)	58 (39)	0	55 (12)	32 (7)	14 (3)
Why earmolds need to be replaced	0	8 (5)	31 (21)	61 (41)	0	50 (11)	32 (7)	18 (4)
How to tell when earmolds need to be replaced	3 (2)	6 (4)	27 (18)	64 (43)	5 (1)	55 (12)	23 (5)	18 (4)
How to know settings are at level needed for their child to hear well	3 (2)	20 (13)	39 (26)	38 (25)	23 (5)	46 (10)	14 (3)	18 (4)
How to determine if their child's devices are functioning properly	3 (2)	0	18 (12)	79 (52)	0	23 (5)	55 (12)	23 (5)
<b>Bone Conduction Hearing Aid and Cochlear Implant</b>								
How to change batteries	0	9 (6)	22 (14)	69 (45)	15 (3)	25 (5)	20 (4)	40 (48)
How to interpret the indicator lights and beeps	2 (1)	9 (6)	31 (20)	59 (38)	15 (3)	20 (4)	20 (4)	45 (9)
How to monitor the condition of external equipment	0	12 (8)	25 (16)	63 (41)	10 (2)	20 (4)	35 (7)	35 (7)
<b>Cochlear Implant</b>								
How to listen to cochlear implant microphone	0	6 (2)	44 (14)	50 (16)	23 (3)	23 (3)	31 (4)	23 (3)

N = Never, S = Sometimes, O = Often, A = Always, U = I don't know

### Loaner Devices

Participants reported how often loaner hearing devices are provided when a child's device needs to be sent in for repair (See Table 3). Loaners were reported as always being provided for 27% of children using HAs, 20% using BCHAs, and 40% of children using CIs.

### Professionals' Confidence Levels

Participants indicated how confident they were in four practices related to monitoring audibility (see Table 4), on a scale from *not confident at all* (0) to *very confident* (100). Audiologists overall reported high levels of confidence ( $M = 84$  to  $99$ ) for all devices (i.e., HA, BCHA, CI). The lowest rating was in being able to tell when programming adjustments are needed for BCHA; responses for this

item also had the greatest variance ( $M = 84$ ;  $SD = 22.68$ ). Interventionists reported a wider range of confidence ratings ( $M = 42$  to  $99$ ). The highest confidence was reported for performing a speech sound test for all devices (HA [ $M = 93$ ;  $SD = 17.80$ ]; BCHA [ $M = 99$ ;  $SD = 3.78$ ]; CI [ $M = 96$ ;  $SD = 9.32$ ]). The lowest confidence was reported for knowing how to verify that hearing aid settings are appropriate ( $M = 42$ ;  $SD = 29.23$ ) and knowing how to tell when programming/mapping adjustments are needed (BCHA [ $M = 51$ ;  $SD = 34.50$ ]; CI [ $M = 75$ ;  $SD = 19.74$ ]).

### Teaching Parents

Participants indicated how frequently (i.e., *never*, *sometimes*, *often*, *always*) they address a variety of topics with parents that are important for monitoring audibility

(see Table 5). Eight topics queried were applicable to all devices (i.e., HA; BCHA; CI), three were applicable to BCHA and CI, and one was applicable to only children who use CIs. Responses revealed variability in practices for teaching parents for all topics.

For teaching practices related to all devices, the top three most frequently taught topics audiologists reported they *always* address included: asking parents about the number of hours their child wears their hearing device (75%), helping parents resolve challenges with hearing device use (74%), and asking parents about challenges with hearing device use (73%). The top three most frequently taught topics interventionists reported they *always* address included: teaching parents how to do a speech sound check (57%), helping parents resolve challenges with hearing device use (49%), and talking with parents about difficulties child may have hearing in different environments (48%). Few professionals reported always talking with parents about data logging results (audiologists 44%; interventionists 9%).

For teaching practices specific to HAs, the most frequently taught topic participants reported they *always* address is talking with parents about how to determine if their child's devices are functioning properly (audiologists 79%; interventionists 23%); remaining topics (how often earmolds need to be replaced; why earmolds need to be replaced; how to tell when earmolds need to be replaced; how to know their child's hearing aid settings are at a level needed to hear well) were addressed less frequently. For teaching practices specific to BCHA and/or CI only, the most frequently taught topic audiologists reported they always address is talking with parents about how to change batteries (69%) and for interventionists, how to interpret device indicator lights (45%). The remaining topics (how to monitor condition of external equipment; how to check CI microphone) were addressed less frequently.

### Factors Influencing Teaching

Professional type (i.e., audiologist; interventionist), device type (i.e., HA; BCHA; CI), and years practicing (i.e., less than 10 years; 10 or more years) were explored to investigate their influence on eight practices for teaching parents applicable to all hearing devices. Analysis of variance showed no statistically significant effects for professional type  $F(1, 154) = 1.233, p = .269$ , device type  $F(2, 183) = 1.095, p = .337$ , or years practicing  $F(1, 154) = 1.089, p = .298$ .

## Discussion

Monitoring aided audibility for children who are DHH is critical for supporting spoken language outcomes. This study explored professionals' perspectives on hearing device use and practices they include in their services for monitoring children's aided hearing. The findings from this study revealed practice gaps and opportunities for improvement.

### Practice Gaps

Research findings have shown that consistent use of well-functioning hearing devices positively contributes to child outcomes, and that children who use hearing aids 10 hours or more per day have better language outcomes (Tomblin et al., 2014). The professionals in this study indicated multiple professionals have the responsibility to talk with parents about hearing device use; however, approximately one-quarter of the audiologists and two-thirds of the interventionists never or only sometimes talk about hearing device data logging with parents. Studies have found that parent report of hearing aid use often over-estimates use when compared to device data logging (Walker et al., 2013; Muñoz et al., 2014), suggesting that parent report alone is insufficient for monitoring how consistently children wear their hearing devices. Hearing device malfunction can also disrupt audibility. When hearing devices are sent to the manufacturer for repair, audibility is compromised if children are not provided with loaner equipment. Participants in this study indicated at least one-quarter of children they work with never or only sometimes have access to loaner devices.

Practices to monitor audibility can be incorporated within routine interactions at home and at school. Verification and validation are important components to include and are indicated in practice guidelines (American Academy of Audiology, 2013). Asking parents to periodically complete a questionnaire about how their child responds to sounds in their daily life (validation) is an important way to understand how children are hearing in various environments. Participants in this study reported rarely using this mechanism to monitor audibility; two-thirds of audiologists and three-fourths of interventionists reported never or only sometimes asking parents to complete a questionnaire. Most of the audiologists reported checking hearing aid settings (verification) after new earmolds are obtained; however, 18% reported they do not verify settings. As children grow their earmolds need to be replaced and hearing aid settings adjusted to maintain audibility. When hearing aid settings are not monitored, children are at risk for under amplification as they grow. Both audiologists and interventionists reported lacking confidence in knowing when programming adjustments are needed for children who use BCHAs.

Teaching parents about monitoring for audibility is critical as parents are with their children every day and are central to the intervention process. Participants in this study reported talking with parents about many aspects important for monitoring audibility; however, one-quarter of the audiologists and two-thirds of the interventionists reported never or only sometimes talking with parents about how to know that their child's hearing devices are set at a level needed for their child to hear well. Professionals reported more frequently talking with parents about how to do a speech sound test. Additionally, approximately one-quarter of audiologists and one-half of the interventionists never or only sometimes talk with parents about how to monitor the function or performance

of personal assistive devices. For parents to advocate effectively for their children they need information about factors critical to audibility and support as they gain confidence implementing routine monitoring tasks.

### Opportunities

Several opportunities to enhance monitoring of aided audibility exist. For example, inter-professional collaboration can improve communication among service providers, improving continuity of care and reinforcement of and support for parent learning. In this study, audiologists reported always knowing hours of device use more often than did interventionists. Regularly sharing key information, such as hearing device data logging, results of aided speech understanding, and verification of device settings, offers opportunities to improve quality of care (Muñoz & Blaiser, 2011). Audiologists and interventionists would benefit from training opportunities to increase their confidence levels. For audiologists, training related to determining when BCHA settings need to be adjusted would be particularly beneficial. For interventionists, training related to knowing how to determine if programming adjustments are needed for all types of devices would be particularly beneficial. Improving access to loaner devices when repairs are necessary would improve consistent audibility for children. Additionally, asking parents to complete questionnaires reporting how their child is hearing at home and in other environments not only provides important information, but is also an opportunity to engage parents in the monitoring process.

### Limitations and Future Research

This survey study was self-report and may not reflect actual practices. Self-report provides insights into perspectives and perceptions of practices; however, it may also be misleading due to conscious bias by the participants to look good (Baldwin, 2000). Self-selection to complete the survey instrument may also introduce bias, artificially inflating frequency of practices reported. Participants were recruited through sources targeting professionals who work with pediatric populations; however, the extent of their case load specific to pediatric hearing loss was not explored. The response rate for intervention professionals was low and findings cannot be generalized to the broader population of speech-language pathologists, early interventionists, and deaf educators.

Further research is needed to identify how to increase professionals' implementation of practices for monitoring aided audibility. Important questions include exploring to what extent audiologists and interventionists are prepared for this aspect of practice within graduate training; what barriers, both internal (e.g., confidence) and external (e.g., equipment access) exist for routine implementation of monitoring practices; how to increase inter-professional collaboration for monitoring aided audibility; and professionals' attitudes related to monitoring aided audibility.

## Conclusion

Children who are DHH and their parents rely on professionals to provide evidence-based practices. This study revealed practice gaps related to monitoring audibility, including infrequent use of parent questionnaires to explore how children are hearing at home and in other environments, lack of loaner equipment for some children when hearing devices are being repaired, and inconsistent monitoring of data logging to identify challenges with hearing aid use. Training opportunities exist to address provider confidence and implementation of monitoring practices.

## References

- American Academy of Audiology. (2013). *Clinical practice guidelines: Pediatric amplification*. Retrieved from <http://galster.net/wp-content/uploads/2013/07/AAA-2013-Pediatric-Amp-Guidelines.pdf>
- American Speech-Language-Hearing Association. (n.d.). *The prevalence and incidence of hearing loss in children*. Retrieved from <http://www.asha.org/public/hearing/Prevalence-and-Incidence-of-Hearing-Loss-in-Children/>
- Baldwin, W. (2000). Information no one else knows: The value of self-report. In A. A. Stone, J. S. Turkkan, C. A. Bachrach, J. B. Jobe, H. S. Kurtzman, & V. S. Cain (Eds.), *The Science of Self-Report: Implications for Research and Practice* (pp. 3–7). Mahwah, NJ: Taylor & Francis e-Library.
- Caballero, A., Muñoz, K., White, K. R., Nelson, L., Domenech-Rodriguez, M., & Twohig, M. (2017). Pediatric hearing aid management: Challenges among hispanic families. *Journal of the American Academy of Audiology, 28*(8), 718–730.
- Centers for Disease Control and Prevention. (2015). *Hearing loss in children: 2015 annual data early hearing detection and intervention (EHDI) program*. Retrieved from <https://www.cdc.gov/ncbddd/hearingloss/2015-data/01-data-summary.html>
- McCreery, R. W., Bentler, R. A., & Roush, P. A. (2013). Characteristics of hearing aid fittings in infants and young children. *Ear and Hearing, 34*(6), 701–710. doi: 10.1097/AUD.0b013e31828f1033
- Mitchell, R. E. & Karchmer, M. A. (2004). Chasing the mythical ten percent: Parental hearing status of deaf and hard of hearing students in the United States. *Sign Language Studies, 4*(2), 138–163.
- Muñoz, K. & Blaiser, K. (2011, Dec.). Audiologists and speech-language pathologists: Making critical cross-disciplinary connections for quality care in EHDI. *Perspectives on Audiology, 7*(1), 34–42.
- Muñoz, K., Rusk, S., Nelson, L., Preston, E., White, K., Barrett, T., & Twohig, M. (2016). Pediatric hearing aid management: Parent reported needs for learning support. *Ear and Hearing, 37*(6), 703–709. doi:10.1097/AUD.0000000000000338
- Muñoz, K., Preston, E., & Hicken, S. (2014). Pediatric hearing aid use: How can audiologists support parents to increase consistency? *Journal of the American Academy of Audiology, 25*, 380–387. doi:10.3766/jaaa.25.4.9
- Tomblin, J. B., Oleson, J. J., Ambrose, S. E., Walker, E., & Moeller, M. P. (2014). The influence of hearing aids on the speech and language development of children with hearing loss. *JAMA Otolaryngology Head & Neck Surgery, 140*, 403–409.
- Walker, E. A., Sprattford, M., Moeller, M. P., Oleson, J., Ou, H., Roush, P., & Jacobs, S. (2013). Predictors of hearing aid use time in children with mild-to-severe hearing loss. *Language Speech and Hearing Services in Schools, 44*(1), 73–88. doi:10.1044/0161-1461(2012)12-0005
- World Health Organization. (2016). Childhood hearing loss. Retrieved from [http://www.who.int/pbd/deafness/world-hearing-day/WHDD2016\\_Brochure\\_EN\\_2.pdf](http://www.who.int/pbd/deafness/world-hearing-day/WHDD2016_Brochure_EN_2.pdf)



**Appendix**  
Hearing Aid Survey

**Pediatric Hearing Aid Management: Professional Practices for Monitoring Children's Aided Hearing**

The purpose of this survey is to better understand how professionals monitor audibility for children ages birth to six years who use hearing aids. Completing the survey should take about 5 minutes.

**Your experiences are important!**

**Information About You:**

1. My profession is:

- Audiologist
- Speech-Language Pathologist
- Teacher
- Early Interventionist
- Other (specify) \_\_\_\_\_

2. I have been working with children with hearing loss for:

- Less than 5 years
- 6–10 years
- More than 10 years

3. Approximately what percent of young children that you work with wear their hearing aids all waking hours?

- \_\_\_\_\_%
- I don't know

4. How often do you know the hours of hearing aid use for the children with whom you work?

- Never
- Sometimes
- Often
- Always

5. I work in:

- An urban area
- A rural area

6. I practice in:

- United States \_\_\_\_\_  
indicate state
- Country \_\_\_\_\_  
indicate country

### Hearing Aid Use

Consistent use of hearing aids is important for children to learn to speak. Having children wear their hearing aids consistently can be hard for many different reasons. Addressing hearing aid use can help identify problems.

7. Indicate each of the professionals you think should talk about hearing aid use with parents: (mark all that apply)

- Audiologist
- Speech-Language Pathologist
- Teacher
- Early Interventionist
- Other (specify) \_\_\_\_\_

8. Indicate how often *you* address each of the following when you talk with parents:

	Never	Sometimes	Often	Always
Ask about the number of hours their child wears hearing aids	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Ask about challenges with hearing aid use	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Help parents resolve challenges with hearing aid use	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Talk about hearing aid data logging results	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>

9. When the children you work with need to have their hearing aids repaired by the company, how often are they typically provided with loaner hearing aids?

- Never
- Sometimes
- Often
- Always
- I don't know

### Monitoring

Monitoring helps you know if children are hearing well with their hearing aids or if there are problems with settings, device function, how earmolds fit, and hearing in noise.

10. When the children you work with get new earmolds, how often are hearing aid settings checked to make sure sounds are being appropriately amplified?

- Never
- Sometimes
- Often
- Always
- I don't know

11. During audiology appointments, how often is speech understanding tested while children are wearing their hearing aid(s)?

- Never
- Sometimes
- Often
- Always
- I don't know

12. How often do you ask parents to complete a questionnaire about how their child is responding to sounds in their daily life (e.g., hearing in quiet, hearing in noise)?

- Never
- Sometimes
- Often
- Always
- I don't know

13. Indicate the questionnaires you use to monitor how children, birth to six years of age, are functioning with their hearing aids in daily life (mark all that apply):

- LittleEARS Auditory Questionnaire
- Parent's Evaluation of Aural/Oral Performance of Children (PEACH)
- Infant-Toddler Meaningful Auditory Integration Scale (IT-MAIS)
- Early Listening Function (ELF)
- Teachers' Evaluation of Aural/Oral performance of Children (TEACH)
- Preschool SIFTER: Preschool Screening Instrument For Targeting Educational Risk
- Children's Home Inventory for Listening Difficulties (CHILD)
- Children's Outcome Worksheets (COW)
- Other (specify): \_\_\_\_\_
- I do not use any questionnaires

14. Indicate how often you talk with parents about each of the following:

	Never	Sometimes	Often	Always
How often earmolds need to be replaced	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Why earmolds need to be replaced	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
How to tell when earmolds need to be replaced	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
How to know the hearing aid settings are at the level needed for their child to hear well	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
How to determine if their child's hearing aids are functioning properly (e.g., sound quality, batteries)	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
How to do a speech sound check (ah, ee, oo, mm, sh, s)	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>

15. Indicate how confident you are in your ability to:

- Recognize when earmolds need to be replaced \_\_\_\_\_ (0-100)
- Verify hearing aid settings are appropriate \_\_\_\_\_ (0-100)
- Conduct a speech sound check \_\_\_\_\_ (0-100)
- Determine hearing aids are functioning properly \_\_\_\_\_ (0-100)

There are personal assistive devices that can help children hear better in noise. The devices work wirelessly (e.g., FM system, remote mic, mini mic) to send the speech signal from a transmitter worn by the speaker, directly to the child's hearing aids.

17. Indicate how often you talk with parents about each of the following:

	Never	Sometimes	Often	Always
Difficulties their child may have hearing in different environments	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Benefits of personal assistive device use in addition to the hearing aids	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Monitoring personal assistive device use	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>

18. What is challenging for you in monitoring audibility for children who wear hearing aids?

**Thank You!**



## Bone Conduction Hearing Aid Survey

### **Pediatric Bone-Conduction Hearing Aid Management (BCHA): Professional Practices for Monitoring Children's Aided Hearing**

The purpose of this survey is to better understand how professionals monitor audibility for children ages birth to six years who use a bone-conduction hearing aid (BCHA). Completing the survey should take about 5 minutes.

**Your experiences are important!**

#### **Information About You:**

1. My profession is:

- Audiologist
- Speech-Language Pathologist
- Teacher
- Early Interventionist
- Other (specify) \_\_\_\_\_

2. I have been working with children with hearing loss for:

- Less than 5 years
- 6–10 years
- More than 10 years

3. Approximately what percent of young children that you work with wear their BCHA(s) all waking hours?

- \_\_\_\_\_%
- I don't know

4. How often do you know the hours of BCHA use for the children with whom you work?

- Never
- Sometimes
- Often
- Always

5. I work in:

- An urban area
- A rural area

6. I practice in:

- United States \_\_\_\_\_  
indicate state
- Country \_\_\_\_\_  
indicate country

### Bone Conduction Hearing Aid Use

Consistent use of BCHA(s) is important for children to learn to speak. Having children wear their device(s) consistently can be hard for many different reasons. Addressing BCHA use can help identify problems.

7. Indicate each of the professionals you think should talk about BCHA use with parents: (mark all that apply)

- Audiologist
- Speech-Language Pathologist
- Teacher
- Early Interventionist
- Other (specify) \_\_\_\_\_

8. Indicate how often you address each of the following when you talk with parents:

	Never	Sometimes	Often	Always
Ask about the number of hours their child wears the BCHA	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Ask about challenges with BCHA use	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Help parents resolve challenges with BCHA use	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Talk about BCHA data logging results	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>

9. When the children you work with need to have their BCHA repaired by the company, how often are they typically provided with loaner equipment?

- Never
- Sometimes
- Often
- Always
- I don't know

### Monitoring

Monitoring helps you know if children are hearing well with their hearing aids or if there are problems with settings, device function, how earmolds fit, and hearing in noise.

10. During audiology appointments, how often is speech understanding tested while children are wearing their BCHA?

- Never
- Sometimes
- Often
- Always
- I don't know

11. How often do you ask parents to complete a questionnaire about how their child is responding to sounds in their daily life (e.g., hearing in quiet, hearing in noise)?

- Never
- Sometimes
- Often
- Always
- I don't know

12. How often do you ask parents to complete a questionnaire about how their child is responding to sounds in their daily life (e.g., hearing in quiet, hearing in noise)?

- Never
- Sometimes
- Often
- Always
- I don't know

13. Indicate the questionnaires you use to monitor how children, birth to six years of age, are functioning with their BCHA in daily life: (mark all that apply):

- LittleEARS Auditory Questionnaire
- Parent's Evaluation of Aural/Oral Performance of Children (PEACH)
- Infant-Toddler Meaningful Auditory Integration Scale (IT-MAIS)
- Early Listening Function (ELF)
- Teachers' Evaluation of Aural/Oral performance of Children (TEACH)
- Preschool SIFTER: Preschool Screening Instrument For Targeting Educational Risk
- Children's Home Inventory for Listening Difficulties (CHILD)
- Children's Outcome Worksheets (COW)
- Other (specify): \_\_\_\_\_
- I do not use any questionnaires

14. Indicate how often you talk with parents about each of the following:

	Never	Sometimes	Often	Always
How to change batteries	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
How to interpret the indicator lights and beeps	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
How to monitor the condition of the external equipment	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
How to do a speech sound check (ah, ee, oo, mm, sh, s)	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>

15. Indicate how confident you are in your ability to:

- Tell when programming adjustments are needed \_\_\_\_\_ (0-100)
- Conduct a speech sound check \_\_\_\_\_ (0-100)
- Check BCHA function \_\_\_\_\_ (0-100)

There are personal assistive devices that can help children hear better in noise. The devices work wirelessly (e.g., FM system, remote mic, mini mic) to send the speech signal from a transmitter worn by the speaker, directly to the child's BCHA.

17. Indicate how often you talk with parents about each of the following:

	Never	Sometimes	Often	Always
Difficulties their child may have hearing in different environments	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Benefits of personal assistive device use in addition to the BCHA	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Monitoring personal assistive device use	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>

18. What is challenging for you in monitoring audibility for children who wear BCHA(s)?

**Thank You!**



**Pediatric Cochlear Implant Management: Professional Practices for Monitoring Children's Aided Hearing**

The purpose of this survey is to better understand how professionals monitor audibility for children ages birth to six years who use cochlear implants. Completing the survey should take about 5 minutes.

**Your experiences are important!**

**Information About You:**

1. My profession is:

- Audiologist
- Speech-Language Pathologist
- Teacher
- Early Interventionist
- Other (specify) \_\_\_\_\_

2. I have been working with children with hearing loss for:

- Less than 5 years
- 6–10 years
- More than 10 years

3. Approximately what percent of young children that you work with wear their cochlear implant(s) all waking hours?

- \_\_\_\_\_%
- I don't know

4. How often do you know the hours of cochlear implant use for the children with whom you work?

- Never
- Sometimes
- Often
- Always

5. I work in:

- An urban area
- A rural area

6. I practice in:

- United States \_\_\_\_\_  
indicate state
- Country \_\_\_\_\_  
indicate country

### Cochlear Implant Use

Consistent use of cochlear implants is important for children to learn to speak. Having children wear their device(s) consistently can be hard for many different reasons. Addressing cochlear implant use can help identify problems.

7. Indicate each of the professionals you think should talk about BCHA use with parents: (mark all that apply)

- Audiologist
- Speech-Language Pathologist
- Teacher
- Early Interventionist
- Other (specify) \_\_\_\_\_

8. Indicate how often you address each of the following when you talk with parents:

	Never	Sometimes	Often	Always
Ask about the number of hours their child wears the cochlear implant(s)	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Ask about challenges with cochlear implant use	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Help parents resolve challenges with cochlear implant use	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Talk about cochlear implant data logging results	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>

9. When the children you work with need to have their cochlear implant repaired by the company, how often are they typically provided with loaner equipment?

- Never
- Sometimes
- Often
- Always
- I don't know

### Monitoring

Monitoring helps you know if children are hearing well with their cochlear implant(s) or if there are problems with settings, device function, and hearing in noise.

10. During audiology appointments, how often is speech understanding tested while children are wearing their cochlear implants?

- Never
- Sometimes
- Often
- Always
- I don't know

11. How often do you ask parents to complete a questionnaire about how their child is responding to sounds in their daily life (e.g., hearing in quiet, hearing in noise)?

- Never
- Sometimes
- Often
- Always
- I don't know

12. How often do you ask parents to complete a questionnaire about how their child is responding to sounds in their daily life while wearing their cochlear implant (e.g., hearing in quiet, hearing in noise)?

- Never
- Sometimes
- Often
- Always
- I don't know

13. Indicate the questionnaires you use to monitor how children, birth to six years of age, are functioning with their cochlear implants in daily life (mark all that apply):

- LittleEARS Auditory Questionnaire
- Parent's Evaluation of Aural/Oral Performance of Children (PEACH)
- Infant-Toddler Meaningful Auditory Integration Scale (IT-MAIS)
- Early Listening Function (ELF)
- Teachers' Evaluation of Aural/Oral performance of Children (TEACH)
- Preschool SIFTER: Preschool Screening Instrument For Targeting Educational Risk
- Children's Home Inventory for Listening Difficulties (CHILD)
- Children's Outcome Worksheets (COW)
- Other (specify): \_\_\_\_\_
- I do not use any questionnaires

14. Indicate how often you talk with parents about each of the following:

	Never	Sometimes	Often	Always
How to change batteries	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
How to listen to the cochlear implant microphone	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
How to interpret the indicator lights and beeps	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
How to monitor the condition of the external equipment, such as cables and headpiece	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
How to do a speech sound check (ah, ee, oo, mm, sh, s)	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>

15. Indicate how confident you are in your ability to:

- Tell when programming/mapping adjustments are needed \_\_\_\_ (0-100)
- Conduct a speech sound check \_\_\_\_ (0-100)
- Check cochlear implant function \_\_\_\_ (0-100)

There are personal assistive devices that can help children hear better in noise. The devices work wirelessly (e.g., FM system, remote mic, mini mic) to send the speech signal from a transmitter worn by the speaker, directly to the child's cochlear implant.

17. Indicate how often you talk with parents about each of the following:

	Never	Sometimes	Often	Always
Difficulties their child may have hearing in different environments	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Benefits of personal assistive device use in addition to the cochlear implant	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>

18. What is challenging for you in monitoring audibility for children who wear cochlear implants?

**Thank You!**



## Identification of Oral Clefts as a Risk Factor for Hearing Loss During Newborn Hearing Screening

Patricia L. Purcell MD, MPH<sup>1</sup>  
Kathleen C. Y. Sie, MD<sup>2,3</sup>  
Todd C. Edwards, PhD<sup>3</sup>  
Debra Lochner Doyle, MS<sup>4</sup>  
Karin Neidt, MPH<sup>4</sup>

<sup>1</sup>Department of Otolaryngology–Head & Neck Surgery, University of Washington, Seattle, WA

<sup>2</sup>Division of Pediatric Otolaryngology, Seattle Children's Hospital, Seattle, WA

<sup>3</sup>Department of Health Services, University of Washington, Seattle, WA

<sup>4</sup>Screening and Genetics Unit, Washington State Department of Health, Shoreline, WA

**Abstract: Objective.** This study assessed whether children with oral clefts are appropriately classified as at-risk for hearing loss at the time of newborn hearing screening and describes their screening and diagnostic results.

**Design.** Birth certificates were used to identify children with cleft lip and palate or isolated cleft palate born in Washington State from 2008–2013. These were cross-referenced with the state's Early Hearing Detection, Diagnosis and Intervention (EHDDI) database. Multivariate logistic regression was used to examine associations.

**Results.** Birth records identified 235 children with cleft lip and palate and 116 with isolated cleft palate. Six children were listed as having both diagnoses. Only 138 (39%) of these children were designated as having a craniofacial anomaly in the EHDDI database. Children who were misclassified were less likely to have referred on initial hearing screening, OR 0.3, 95% CI [0.2, 0.5]. Misclassification of risk factor status was also associated with delayed hearing screening past 30 days of age or unknown age at screening, OR 4.4, 95% CI [1.5, 13.3],  $p = 0.008$ . Of 50 children with diagnostic results; 25 (50%) had hearing loss: 18 conductive, 2 mixed, and 5 unspecified.

**Conclusion.** A majority of children with oral clefts were misclassified regarding risk factor for hearing loss in the EHDDI database.

**Acronyms:** EHDI = Early Hearing Detection and Intervention; EHDDI = Early Hearing-loss Detection, Diagnosis and Intervention; DHH = deaf or hard of hearing; JCIH = Joint Committee on Infant Hearing; NICU = neonatal intensive care unit

**Acknowledgements:** The authors would like to acknowledge the contributions of Marcie Rider, AuD, of Washington State Department of Health Screening and Genetics Unit, and Laura Steinmetz, AuD, pediatric audiologist and consultant to Washington State EHDDI program for their contributions to the conceptualization and design of this study.

**Funding Sources:** All phases of this study were supported by Project T76 MC 00011 and Project #H61MC00084 from the Maternal and Child Health Bureau (Title V, Social Security Act), Health Resources and Services Administration. Patricia Purcell is an otolaryngology resident and research fellow who was supported by Grant 2T32DC000018, Institutional National Research Service Award for Research Training in Otolaryngology from the National Institute on Deafness and Other Communication Disorders.

**Correspondence concerning this article should be addressed to:** Kathleen Sie, MD, Division of Pediatric Otolaryngology, Seattle Children's Hospital, 4800 Sand Point Way NE, Seattle, WA 98115. Phone: 206-987-9562; Email: [kathysie@uw.edu](mailto:kathysie@uw.edu)

### Introduction

The Joint Committee on Infant Hearing (JCIH) has established the 1-3-6 guideline for detection, diagnosis, and intervention for congenital hearing loss (JCIH, 2007;

Bower & St. John, 2014). All children should undergo hearing screening by 1 month of age, diagnostic audiology assessment by 3 months of age for those who do not pass screening, and enrollment in early intervention by 6 months of age for those with hearing loss. In addition,

children with certain risk factors for hearing loss should undergo diagnostic audiology assessment by 24 to 30 months of age, even if they pass their initial hearing screen (JCIH, 2007; Beswick, Driscoll, & Kei, 2012). To promote adherence to these guidelines, states have created Early Hearing Detection and Intervention (EHDI) programs; in Washington State, the program has been titled, “Early Hearing Detection, Diagnosis and Intervention” (EHDDI). These programs can monitor follow-up for children who do not pass hearing screening or who have risk factors for hearing loss (White, 2014; Gracey, 2003).

The EHDDI program relies on newborn hearing screeners to report hearing screening results and indicate which children have risk factors for delayed onset or progressive hearing loss. In Washington State, newborn hearing screenings can be performed by nursing staff, obstetric technicians, audiologists, midwives, and staff from contracted newborn hearing screening companies. Individuals do not need certification to conduct newborn hearing screenings, but EHDDI program staff provide training and outreach. This training includes information about risk factors for delayed onset and progressive hearing loss and how to report risk factors to the EHDDI program. Newborn hearing screeners may also receive training from the hospital, clinic, or company where they work. Washington State does not have legislation that mandates newborn hearing screening or reporting results to the EHDDI program. Although screening is voluntary, it is estimated that 96% of all infants born in Washington State receive a newborn hearing screening (Weisman, 2014).

There are 29 audiology clinics throughout the state that provide comprehensive diagnostic audiological assessment for infants. These clinics meet Washington State’s Protocol for Diagnostic Audiological Assessment: Follow-up for Newborn Hearing Screening. This protocol is based on the JCIH 2007 position statement and was created by a workgroup of 22 audiologists with expertise in the screening and diagnosis of hearing loss in newborns and infants (Washington State Department of Health, 2011).

Children with craniofacial anomalies are at greater risk of hearing loss (Lieu, Ratnaraj, & Ead, 2013; Yelverton et al., 2013; Beswick, Driscoll, Kei, Khan, & Glennon, 2013). Most commonly, children with these anomalies will experience conductive hearing loss due to anatomic abnormalities affecting middle ear function, although sensorineural hearing loss can occur as well (Swibel Rosenthal, Caballero, & Drake, 2012). The most common craniofacial anomaly is cleft lip and palate, which occurs in approximately 10 per 10,000 live births; isolated cleft palate is also relatively common with an incidence of 6.5 cases per 10,000 births (National Birth Defects Prevention Network, 2010). These craniofacial anomalies can be grouped together under the term oral clefts. Children with oral clefts have high rates of conductive hearing loss, most commonly associated with Eustachian tube dysfunction (Kuo et al., 2014).

The objective of this study was to determine what proportion of children with cleft lip and palate or isolated cleft palate were correctly identified as having a craniofacial anomaly at the time of newborn hearing screening, and to determine if there was an association between correct identification of risk factor status and adherence to guidelines for newborn hearing screening.

## Materials and Method

Prior to investigation, approval for this study was obtained from the Washington State Department of Social and Health Services’ Human Research Review Section. In Washington State, the long form birth certificate records from January 1, 2008 to December 31, 2013 were electronically queried. The long form birth certificate includes a field for designating whether a child is born with an oral cleft, which includes either cleft lip and palate or isolated cleft palate.

Birth record numbers were then cross-referenced with the state’s EHDDI database to identify the cohort. The EHDDI database was queried to obtain information regarding birth weight, race and ethnicity, maternal age, and presence of JCIH hearing loss risk factors. The Washington State newborn hearing screening card includes data regarding five risk factors for delayed onset or progressive hearing loss including the following: (a) neonatal intensive care unit (NICU) stay greater than 5 days, (b) syndrome with stigmata of hearing loss, (c) family history of hearing loss, (d) presence of craniofacial anomalies, and (e) in-utero infection. In addition, screeners can denote that a child has no risk factors present. Hearing screening and diagnostic audiology test results were recorded. The birth hospital location and site of diagnostic audiology assessment were also recorded for each child. The demographic data were collected because factors such as birthweight, socioeconomic status, and geographic location may impact the potential association between risk factor misclassification and adherence to newborn hearing screening guidelines.

After data collection was completed, cases were reviewed to determine the number of children who underwent hearing screening and diagnostic testing, and at what age the testing occurred. Diagnostic testing results were then reviewed to determine the types and configuration of hearing loss.

The cohort was divided into two groups based on whether the child had been appropriately identified as having a craniofacial anomaly (risk factor 4) on EHDDI screening card. One group consisted of children with oral clefts correctly classified as having risk factor 4, while the other consisted of children with oral clefts who were misclassified by not having risk factor 4 noted at time of screening. Comparisons were made between the two groups regarding demographic characteristics, presence

of other risk factors, and adherence to 1-3-6 guidelines for screening and diagnostic testing.

All children who had completed newborn hearing screening by 30 days of age were considered to have met the guideline for screening. Children who did not have record of screening were noted, but excluded from further analysis. Among children who did not pass their hearing screen, those who completed diagnostic testing by 90 days of age were considered to have met the guideline for diagnostic testing.

### Analysis

Based on the presence or absence of craniofacial risk factor designation, univariate analysis was performed to calculate descriptive statistics, including means and proportions, for the two groups within the cohort. For continuous variables, an unpaired *t*-test was used for inferential testing; for binary variables, chi-square testing was used to determine significance.

Multivariate logistic regression was then used to investigate association between risk factor status and adherence to screening and diagnostic guidelines. Risk estimates were expressed as odds ratios (OR) and 95% confidence intervals (CI). The following variables were considered as potential confounding covariates: maternal age, birth weight, race or ethnicity, and distance from birth hospital to site of diagnostic audiology assessment. Data that met  $p < 0.05$  were considered statistically significant. Stata 13.1 (Stata Inc, College Station, TX) statistical software was used for all analyses.

### Results

Total number of resident births in Washington State from January 2008 to December 2013 was 526,774. Birth certificate records identified 357 children with oral cleft malformations born during this time period. There were 235 children with cleft lip and palate and 116 with isolated cleft palate for a birth prevalence of 4.5 per 10,000 births and 2.2 per 10,000 births, respectively. There were 6 children who had been erroneously designated as having both diagnoses. Of 357 children, only 138 (39%) were designated as having a craniofacial anomaly in EHDDI database, while 130 (36.4%) were erroneously designated as having no risk factor for hearing loss. The other 89 children were not provided with risk factor classification at time of screening.

Table 1 contains the characteristics of the cohort based on whether or not they were designated as having craniofacial risk factor. Sixty-six percent of children with isolated cleft palate were misclassified compared to 59% of children with cleft lip and palate. Children who were correctly classified had a mean birthweight of 3318.9 g ( $SD = 600.7$  g), which was higher than the mean birthweight among

children who were misclassified 3189.4 g ( $SD = 670.5$  g), but this finding did not achieve significance with unpaired *t*-test,  $p$ -value = 0.07.

**Table 1**  
*Characteristics of Children with Oral Clefts by Craniofacial Risk Factor Identification*

	Craniofacial risk factor identified at hearing screening	Craniofacial risk factor not identified at hearing screening
Total number (%)	138 (38.7%)	219 (61.3%)
Craniofacial anomaly*		
Cleft lip & palate (%) ( <i>n</i> = 235)	96 (40.9%)	139 (59.1%)
Isolated cleft palate (%) ( <i>n</i> = 116)	40 (34.5%)	76 (65.5%)
Mean birthweight in grams, (SD)	3318.9 (600.7)	3189.4 (670.5)
Racial/ethnic minority (%)** ( <i>n</i> = 118)	56 (47.5%)	62 (52.5%)
Mean maternal age in years, (SD)	28.4 (6.2)	28.1 (5.7)
Presence of other risk factors for hearing loss (%):		
Neonatal ICU ( <i>n</i> = 59)	21 (35.6%)	38 (64.4%)
Stigmata of syndrome ( <i>n</i> = 10)	5 (50%)	5 (50%)
Family history ( <i>n</i> = 11)	4 (36.4%)	7 (63.6%)
In-utero infection ( <i>n</i> = 0)	--	--

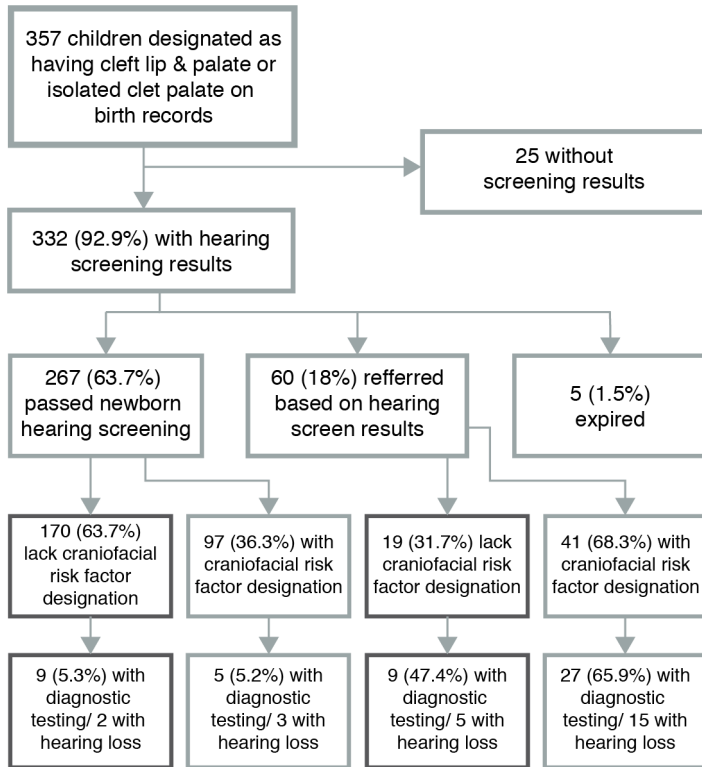
\* 6 children were listed as having both malformation types

\*\*Not all children had information available regarding race or ethnicity

There were no significant differences between the groups based on race and ethnicity, maternal age, or presence of other risk factors. Of the 357 children, 59 children (16.5%) also had NICU risk factor. No child had a risk factor for in-utero infection, and only a small proportion of children had any of the other risk factors.

Mean age at first hearing screening was 5.4 days (range 0–135 days); 217 (60.1%) underwent screening by the second day of life. There were 25 children (7%) who did not undergo newborn hearing screening; none of these children were correctly classified as having a craniofacial risk factor. Twelve children (3.6%) underwent screening after 30 days of age, and 16 children were of unknown age at time of screening.

Of the 332 children who underwent hearing screening, 60 (18%) did not pass their hearing screen (see Figure 1) and 36 (60%) underwent diagnostic testing. The mean age at diagnostic testing for these children was 74.8 days (range 8–232 days). Thirty of the 60 (50%) who referred underwent diagnostic testing by 90 days of age.



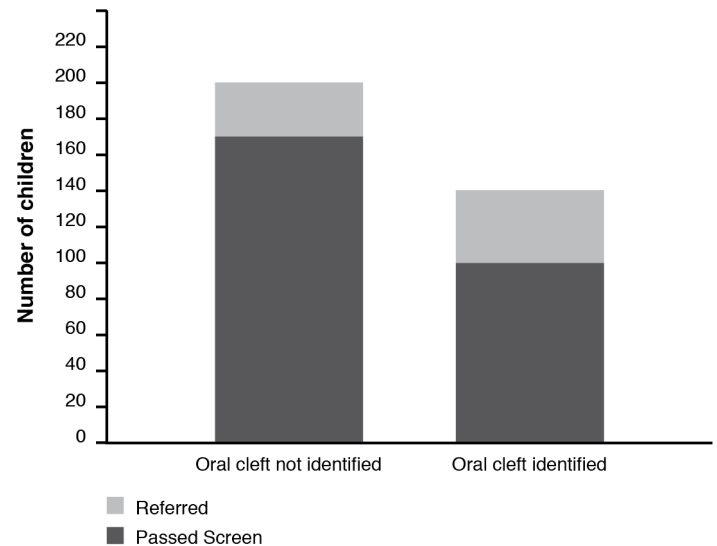
**Figure 1.** Flowchart of screening and diagnostic characteristics of children with oral clefts.

Diagnostic results were available for an additional 14 children who had passed their hearing screen, so that a total of 50 children had diagnostic results. Of these, 25 (50%) had a final diagnosis of hearing loss: 18 conductive, 2 mixed, and 5 unspecified.

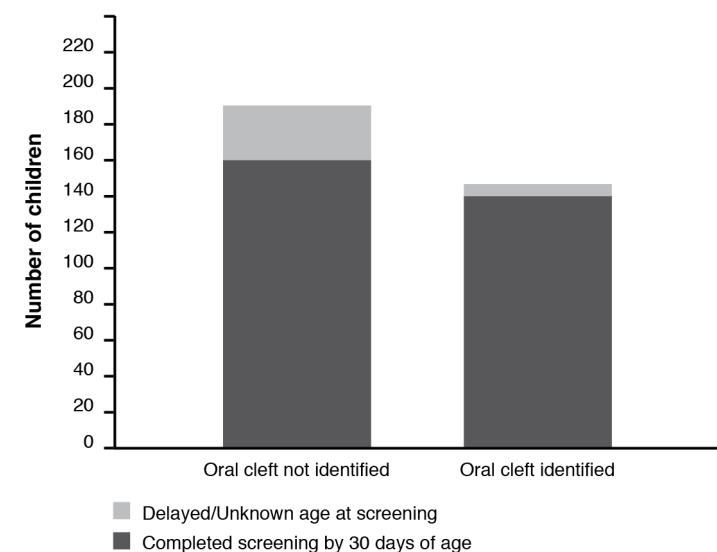
Logistic regression was performed to investigate associations between risk factor status and not passing the newborn hearing screen. There were not significant differences in race/ethnicity or maternal age between children who were correctly classified compared with those who were not, so these covariates were not included in the regression model. Distance from a child’s birth hospital to the audiology center of referral was calculated and explored as a covariate, but it was not significant.

Low birth-weight was found to be significantly associated with likelihood of delayed screening. Of 12 children who underwent screening after 30 days of age, 6 (50%) weighed less than 2500 g at birth,  $p$ -value = 0.001. In addition, as noted above, there was a difference in the mean birth weight between children who were correctly classified on risk factor status and those who were

not, although this difference did not achieve statistical significance. Therefore, birth-weight was included in the model as a binary variable based on a child’s birth weight being less than or greater than 2500 g. After adjustment for birth-weight status, children who were misclassified were less likely to have referred on hearing screening, OR 0.3, 95% CI [0.2, 0.5],  $p < 0.001$ , see Figure 2.



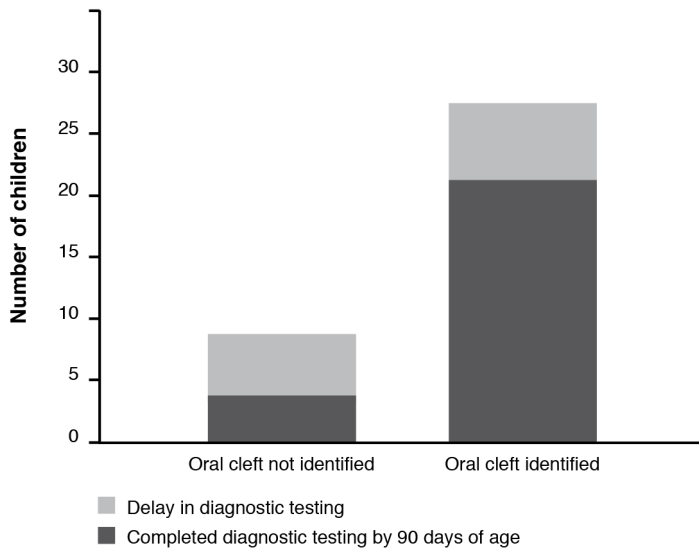
**Figure 2.** Among children identified as having oral clefts on birth certificate, those who were not identified as having a craniofacial risk factor at time of hearing screening were less likely to have referred on initial hearing screening, OR 0.3, 95% CI [0.2, 0.5],  $p < 0.001$ .



**Figure 3.** Stacked bar chart compares adherence to hearing screening guidelines among children with oral clefts. Those who were not identified as having a craniofacial risk factor at time of hearing screening were more likely to also have delayed or unknown age at screening, OR 4.4, 95% CI [1.5, 13.3],  $p = 0.008$ .



When association between risk factor status and adherence to 1-3-6 guidelines was investigated, misclassification of risk factor status was associated with delayed hearing screening past 30 days of age or unknown age at screening, OR 4.4, 95% CI [1.5, 13.3],  $p$ -value = 0.008, see Figure 3. This study also found that misclassification may be associated with delayed diagnostic testing past 90 days of age or unknown age at testing, OR 5.7, 95% CI [0.9, 38],  $p$  = 0.07, see Figure 4. However, this result did not reach statistical significance. The results of the logistic regression models are also summarized in Table 2.



**Figure 4.** Stacked bar chart compares adherence to diagnostic testing guidelines among children with oral clefts who did not pass their hearing screen. Those who were not identified as having a craniofacial risk factor at time of hearing screening may be more likely to also have delayed or unknown age at diagnostic testing, OR 5.7, 95% CI [0.9, 38],  $p$  = 0.07; however, this result did not reach statistical significance.

**Table 2**  
*Logistic regression models of association between risk factor misclassification and newborn hearing outcome*

Outcome	Coefficient	Coefficient Confidence Interval	Odds Ratio	Odds Ratio Confidence Interval	$p$ -value
Child refers on 1 <sup>st</sup> screen	-1.2	-1.7, -0.6	0.3	0.2, 0.5	<0.001
Child has delayed or unknown age at hearing screening	1.5	0.4, 2.6	4.4	1.5, 13.3	0.01
Child has delayed or unknown age at diagnostic assessment	1.4	0.5, 2.7	5.7	0.9, 38	0.07

## Discussion

In 2013, JCIH issued a statement emphasizing the importance of accurate data management systems for newborn hearing screening, stating that such systems are critical to facilitate timely, well-coordinated entry into early intervention for all children who are deaf or hard of hearing (DHH). Other studies have come to similar conclusions (Shulman et al., 2010).

States have dedicated substantial resources to early identification of hearing loss through the establishment of EHDI programs; however, debate remains as to the role that risk factor designation should play in hearing screening and surveillance. Not all states track risk factors for hearing loss as part of a hearing screening program. In a recent national survey, about 65% of states had databases containing information about risk factors for hearing loss (Houston, Behl, White, & Forsman, 2010).

Evidence supports universal screening as preferable to screening based upon risk factors. Universal newborn hearing screening is associated with improved language and literacy outcomes among children who are DHH because it facilitates their enrollment in early intervention services (Yoshinaga-Itano, 2003). Lack of intervention at a critical period in development may have long-lasting impact. In a prospective cohort study of children who were DHH, Pimperton and colleagues found that disparity in reading comprehension between those enrolled in early intervention and those who were not continued to widen with age (Pimperton et al., 2016). Wake and colleagues (2016) recently performed a population-based investigation of Australian states with similar demographic characteristics, finding that universal screening was associated with greater improvement in language outcomes than risk-factor based screening. Other studies have raised questions as to which risk factors for hearing loss are important to monitor. For example, a retrospective study of risk factor registry in Queensland, Australia, found two risk factors, family history and craniofacial anomalies, to predict the occurrence of postnatal hearing loss (Beswick et al., 2013). However, a subsequent study found family history of hearing loss to be of low yield in predicting development of hearing loss (Driscoll, Beswick, Doherty, D'Silva, & Cross, 2015), and it is no longer monitored as a risk factor in the United Kingdom (Sutton et al., 2012). A more recent study found that approximately 10% of children have a risk factor for delayed onset or progressive hearing loss, and of those children, 2.3% develop a permanent hearing loss by age 3 (Dumanch et al., 2017). This study also found the presence of craniofacial anomalies to be among the factors that placed a child at the highest risk for permanent postnatal hearing loss.

However, risk factor identification may be a useful adjunct to universal screening in that it could help target limited resources to those at greatest risk. EHDI programs across the nation continue to face challenges,

including incomplete data reporting and lack of follow-up (Nikolopoulos, 2015). Using a nationwide survey, Gaffney, Green, & Gaffney (2010) found that two-thirds of children who did not pass their final hearing screening in 2005 did not have documentation of a diagnostic test result, primarily due to loss to follow-up. In 2014, a similar study found that more than one-third of children referred lacked a diagnostic result (Alam, Gaffney, & Eichwald, 2014). A recent nationwide survey of parents found that more than 35% could not recall whether their child had undergone screening, and many remained unsure about guidelines for follow-up (Pynnonen et al., 2016). If we acknowledge there are limited resources available for counseling families regarding screening, testing, and surveillance; then perhaps there is a role for proper identification of risk factors as a way to assist efforts for targeted counseling.

The current study used birth certificate records to determine that only 39% of children with oral clefts were correctly classified as having a craniofacial risk factor at the time of newborn hearing screening. Children who were misclassified were less likely to have met 1-3-6 guidelines for screening and diagnosis. Unfortunately, early intervention data were not available at the time of this study. JCIH guidelines call for all children with craniofacial anomalies to complete one diagnostic assessment by 24 to 30 months of age. However, this study found only 50 (14%) of 357 children with oral clefts to have diagnostic results in the state's EHDDI database. Of the children who did not pass screening, 60% had a diagnostic test result, which is consistent with the national percentage (Alam et al., 2014).

Oral clefts are associated with conductive hearing loss (Schönmeyer & Sadhu, 2014); this increased risk is thought to be due to abnormality of the tensor veli palatini muscle, which inserts onto the membranous portion of the Eustachian tube to equilibrate the middle ear space. Children with cleft palate, or even submucous cleft, will often have persistent middle ear effusion requiring tympanostomy tube placement (Reiter, Brosch, Wefel, Schlömer, & Haase, 2011; Smillie, Robertson, Yule, Wynne, & Russell, 2014; Szabo 2010). Of the 25 children found to have hearing loss in this study, most had a conductive hearing loss; these findings appear similar to previous studies (Viswanathan, Vidler, & Richard, 2008).

Tympanostomy tube placement is frequently performed at the same time as cleft palate repair, often around 1 year of age (Kosowski, Weathers, Wolfswinkel, & Ridgway, 2012). Craniofacial centers typically offer pediatric audiology services and diagnostic assessment, especially in the setting of tympanostomy tube placement. Therefore, we need to investigate how well these results are being reported to EHDDI. A next step for this investigation would be to longitudinally track clinical records for the children within this cohort, perhaps at the state's largest tertiary care facility, Seattle Children's Hospital, to determine how many children have records of evaluation and intervention.

Given the frequency of middle ear effusion among patients with cleft palate, recent studies have also questioned whether children with oral clefts should undergo diagnostic audiology testing prior to tympanostomy tube placement (Jordan & Sidman, 2014). Perhaps provider uncertainty regarding the utility of current guidelines could also be contributing to lack of adherence to 1-3-6 guidelines among children with oral clefts. More qualitative data gathering from pediatric otolaryngologists and audiologists might help to clarify this point further.

As an observational cohort study, this investigation had several limitations. It was a population-based study, but the frequency of certain events was quite low, which can make it difficult to determine statistical significance. In addition, a number of children had missing data. There is also the potential for additional confounding by unmeasured factors. Previous studies found that children from rural settings are more likely to have delayed screening or diagnosis (Bush et al., 2015), but this dataset did not include home address information. Distance from birth hospital to audiology center was explored as a covariate, but it was found to not be significant.

If states choose to allocate resources for documentation of risk factors, it is important for this process to be accurate. This study found oral clefts, the most common craniofacial risk factor, to be widely under-reported. If craniofacial risk factors are being under-reported, it is likely that other risk factors are as well. These findings prompt the question of what can be done to improve the system.

Nationwide, efforts are underway to improve integration of electronic health records among clinical providers. Perhaps integration could expand to include public health programs, such as EHDI, although privacy concerns would have to be addressed (Uhler, Thomson, Cyr, Gabbard, & Yoshinaga-Itano, C., 2014). To our knowledge, efforts are underway to implement linkage models in Utah and Nebraska between birth certificate and other vital records and newborn screening registries (McVicar, 2014; Northrop, 2014). It will be important to monitor how effective these systems are at correctly identifying risk factors and whether this change has any effect on rate of enrollment in early intervention programs.

In the meantime, additional education could be provided to those who perform newborn hearing screening regarding the importance of correctly identifying an infant's risk factor status. Newborn hearing screeners are often clinical staff or technicians who receive training by either hospitals or contractor companies. One commonly used curriculum is provided through the National Center for Hearing Assessment and Management (NCHAM), but there are not standardized methods for assessment of screener competencies in the vast majority of states.

In Washington State, EHDDI staff conduct site visits and hold annual meetings to train screeners. About half of hospital newborn hearing screening programs

in Washington require that newborn hearing screeners achieve annual competencies in screening. It might be helpful to develop a training curriculum for hearing screeners that includes education related to identification of risk factors for hearing loss, including oral clefts. Regulations could be set that require hospital screeners to participate in training or meet certain competencies in order for institutions to receive certification from states. For example, California is now requiring that inpatient hearing screening be administered by certified facilities, and certification is required for reimbursement for hearing screening services provided to MediCal eligible infants (California Newborn Hearing Screening Program, 2016). If more states had a certification process that required particular training elements, there may be improvement in screening and reporting of risk factors.

## Conclusion

Using birth certificate records, this population-based study found that a majority of children with oral clefts were misclassified regarding hearing loss risk in the Washington state EHDDI database. Children who were misclassified may be less likely to meet 1-3-6 guidelines for screening and diagnostic audiology assessments. As states take steps to improve data systems and standards for hearing screening certification, it is important to investigate accuracy and effectiveness of newborn hearing screening systems to improve care and services for children who are DHH.

## References

- Alam, S., Gaffney, M., & Eichwald, J. (2014). Improved hearing screening follow-up results in more infants identified. *Journal of Public Health Management and Practice, 20*(2), 220–223.
- Beswick, R., Driscoll, C., & Kei, J. (2012). Monitoring for postnatal hearing loss using risk factors: A systematic literature review. *Ear and Hearing, 33*(6), 745–756.
- Beswick, R., Driscoll, C., Kei, J., Khan, A., & Glennon, S. (2013). Which risk factors predict postnatal hearing loss in children? *Journal of the American Academy of Audiology, 24*(3), 205–213.
- Bower, C. M., & St. John, R. (2014). The otolaryngologist's role in newborn hearing screening and early intervention. *Otolaryngologic Clinics of North America, 47*(5), 631–649.
- Bush, M. L., Hardin, B., Rayle, C., Lester, C., Studts, C. R., & Shinn, J. B. (2015). Rural barriers to early diagnosis and treatment of infant hearing loss in Appalachia. *Otology and Neurotology, 36*(1), 93–98.
- California Newborn Hearing Screening Program. (2016). Health & Safety Code: Regarding NHSP. Retrieved from <http://www.dhcs.ca.gov/services/nhsp/Pages/default.aspx>
- Driscoll, C., Beswick, R., Doherty, E., D'Silva, R., & Cross, A. (2015). The validity of family history as a risk factor in pediatric hearing loss. *International Journal of Pediatric Otorhinolaryngology, 79*, 654–659.
- Dumanch, K. A., Holte, L., O'Hollearn, T., Walker, E., Clark, J., & Oleson, J. (2017). High risk factors associated with early childhood hearing loss: A 3-year review. *American Journal of Audiology, 26*(2), 129–142.
- Gaffney, M., Green, D. R., & Gaffney, C. (2010). Newborn hearing screening and follow-up: Are children receiving recommended services? *Public Health Reports, 125*(2), 199–207.
- Gracey, K. (2003). Current concepts in universal newborn hearing screening and early hearing detection and intervention programs. *Advances in Neonatal Care, 3*(6), 308–317.
- Houston, K. T., Behl, D. D., White, K. R., & Forsman, I. (2010). Federal privacy regulations and the provision of Early Hearing Detection and Intervention programs. *Pediatrics, 126*(Supp. 1), S28–S33.
- Joint Committee on Infant Hearing of the American Academy of Pediatrics. (2007). Year 2007 position statement: Principles and guidelines for early hearing detection and intervention programs. *Pediatrics, 124*(4), 898–921.
- Joint Committee on Infant Hearing of the American Academy of Pediatrics. (2013). Supplement to the JCIH 2007 position statement: Principles and guidelines for early intervention after confirmation that a child is deaf or hard of hearing. *Pediatrics, 131*(4), e1324–e1349.
- Jordan, V. A., & Sidman, J. D. (2014). Hearing outcomes in children with cleft palate and referred newborn hearing screen. *The Laryngoscope, 124*(9), E384–E388.
- Kosowski, T. R., Weathers, W. M., Wolfswinkel, E. M., & Ridgway, E. B. (2012). Cleft palate. *Seminars in Plastic Surgery, 26*(4), 164–169.
- Kuo, C. L., Tsao, Y. H., Cheng, H. M., Lien, C. F., Hsu, C. H., Huang, C. Y., & Shiao, A. S. (2014). Grommets for otitis media with effusion in children with cleft palate: A systematic review. *Pediatrics, 134*(5), 983–994.
- Lieu, J. E., Ratnaraj, F., & Ead, B. (2013). Evaluating a prediction model for infant hearing loss. *The Laryngoscope, 123*(11), 2873–2879.
- McVicar, S. B. (2014). 2013 Utah HRSA Grant. Retrieved from <http://www.infanthearing.org/stategrants/index.php>
- National Birth Defects Prevention Network. (2010). Updated national birth prevalence estimates for selected birth defects in the United States, 2004–2006. *Birth Defects Research Part A: Clinical and Molecular Teratology, 88*(12), 1008–1016.
- Nikolopoulos, T. P. (2015). Neonatal hearing screening: What we have achieved and what needs to be improved. *International Journal of Pediatric Otorhinolaryngology, 79*(5), 635–637.
- Northrop, K. (2014). 2014 Nebraska HRSA Grant. Retrieved from <http://www.infanthearing.org/stategrants/index.php>
- Pimperton, H., Blythe, H., Kreppner, J., Mahon, M., Peacock, J. L., Stevenson, J., . . . Kennedy, C. R. (2016). The impact of universal newborn hearing screening on long-term literacy outcomes: A prospective cohort study. *Archives of Disease in Childhood, 101*, 9–15.
- Pynnonen, M. A., Handelsman, J. A., King, E. F., Singer, D. C., Davis, M. M., & Lesperance, M. M. (2016). Parent perception of newborn hearing screening: Results of a US national survey. *JAMA Otolaryngology Head Neck Surgery, 142*(6), 538–543.
- Reiter, R., Brosch, S., Wefel, H., Schlömer, G., & Haase, S. (2011). The submucous cleft palate: Diagnosis and therapy. *International Journal of Pediatric Otorhinolaryngology, 75*(1), 85–88.
- Schönmeier, B., & Sadhu, P. (2014). A review of the tensor veli palatine function and its relevance to palatoplasty. *Journal of Plastic Surgery and Hand Surgery, 48*(1), 5–9.
- Shulman, S., Besculides, M., Saltzman, A., Ireys, H., White, K. R., & Forsman, I. (2010). Evaluation of the universal newborn hearing screening and intervention program. *Pediatrics, 126*(Supp. 1), S19–S27.
- Smillie, I., Robertson, S., Yule, A., Wynne, D. M., & Russell, C. J. (2014). Complications of ventilation tube insertion in children with and without cleft palate: A nested case-control comparison. *JAMA Otolaryngology Head Neck Surgery, 140*(10), 940–943.
- Sutton, G., Wood, S., Feirn, R., Minchom, S., Parker, G., & Sirimanna, T. (2012). Newborn hearing screening and assessment: Guidelines for surveillance and audiological referral of infants & children following the newborn hearing screen. Retrieved from <http://abrpeerreview.co.uk/onewebmedia/NHSP%20Surveillance%20guidelines%20v5-1%20290612.pdf>
- Swibel Rosenthal, L. H., Caballero, N., & Drake, A. F. (2012). Otolaryngologic manifestations of craniofacial syndromes. *Otolaryngology Clinics of North America, 45*(3), 557–577.
- Szabo, C., Langevin, K., Schoem, S., & Mabry, K. (2010). Treatment of persistent middle ear effusion in cleft palate patients. *International Journal of Pediatric Otorhinolaryngology, 74*(8), 874–877.

- Uhler, K., Thomson, V., Cyr, N., Gabbard, S. A., & Yoshinaga-Itano, C. (2014). State and territory EHCI databases: What we do and don't know about the hearing or audiological data from identified children. *American Journal of Audiology*, *23*(1), 34–43.
- Viswanathan, N., Vidler, M., & Richard, B. (2008). Hearing thresholds in newborns with cleft palate assessed by auditory brain stem response. *The Cleft Palate-Craniofacial Journal*, *45*(2), 187–192.
- Wake, M., Ching, T. Y., Wirth, K., Poulakis, Z., Mensah, F. K., Gold, L., . . . Rickards, F. (2016). Population outcomes of three approaches to detection of congenital hearing loss. *Pediatrics*, *137*(1), e2015-1722.
- Washington State Department of Health. (2011). Protocol for diagnostic audiological assessment: Follow-up for newborn hearing screening. Retrieved from [https://www.doh.wa.gov/Portals/1/Documents/Pubs/344-016\\_EHDDIAudioProtocol.pdf](https://www.doh.wa.gov/Portals/1/Documents/Pubs/344-016_EHDDIAudioProtocol.pdf)
- Weisman, J. (2014). Universal newborn hearing screening 2012. Washington State Department of Health. Retrieved from [https://www.doh.wa.gov/Portals/1/Documents/Pubs/344-022\\_EHDDISBOHReport.pdf](https://www.doh.wa.gov/Portals/1/Documents/Pubs/344-022_EHDDISBOHReport.pdf)
- White, K. R. (2014). The evolution of EHCI from concept to standard of care. In L. Schmeltz (Ed.), *A Resource Guide for Early Hearing Detection and Intervention*. Logan, Ut: National Center for Hearing Assessment and Management. Retrieved from <http://www.infantheating.org/ehdi-ebook/>
- Yelverton, J. C., Dominguez, L. M., Chapman, D. A., Wang, S., Pandya, A., & Dodson, K. M. (2013). Risk factors associated with unilateral hearing loss. *JAMA Otolaryngology Head Neck Surgery*, *139*(1), 59–63.
- Yoshinaga-Itano, C. (2003). Early intervention after universal neonatal hearing screening: Impact on outcomes. *Mental Retardation and Developmental Disabilities Research Reviews*, *9*(4), 252–266.



## Perceptions of Cochlear Implant Audiologists Regarding Sequential Versus Simultaneous Bilateral Cochlear Implants for Children

Frayne Poeting, BA<sup>1</sup>  
Donald M. Goldberg, PhD<sup>1,2</sup>

<sup>1</sup>College of Wooster  
<sup>2</sup>Cleveland Clinic Foundation

**Abstract:** This study examined the opinions of cochlear implant audiologists in the United States regarding sequential versus simultaneous bilateral cochlear implants for children. Audiologists were asked about the most important factors they consider when choosing between sequential or simultaneous bilateral cochlear implants for pediatric candidates. All of the responding audiologists valued binaural hearing, but most worked with sequentially implanted patients. For these sequentially implanted recipients, the most common interval between the first and second surgeries was less than one year. Overall, the audiologists were more likely to recommend bilateral sequential cochlear implants to their patients, but many believed that bilateral simultaneous cochlear implants had value. More research is needed to identify the best interval for sequential cochlear implant recipients, along with the need to further investigate the frequency and types of therapy being used for the training of the second ear for these children.

**Key Words:** bilateral cochlear implants, simultaneous cochlear implants, sequential cochlear implants

**Acronyms:** ACIA = American Cochlear Implant Alliance; ANSD = Auditory Neuropathy Spectrum Disorder; CI = cochlear implant

**Correspondence concerning this article should be addressed to:** Donald M. Goldberg, PhD, College of Wooster, 203 Wishart, 1189 Beall Avenue, Wooster, Ohio 44691. Phone: 330-263-2029; Email: [dgoldberg@wooster.edu](mailto:dgoldberg@wooster.edu)

In our changing landscape for children who are deaf or hard of hearing, some make use of personal hearing aids, but sound may not be adequately amplified to allow for the audibility of the entire speech spectrum. Cochlear implants (CIs) have therefore become an efficacious option, especially for young children (Hammes, Novak, Rotz, Willis, Edmonson, & Thomas, 2002; Vohr, Jodoin-Krauzk, Tucker, Johnson, Topol, & Ahlgren, 2008). There is substantial evidence that children have better listening and language outcomes when they receive early cochlear implantation and participate in early intervention programs (Dornan, Hickson, Murdoch, & Houston, 2007, 2008; Eriks-Brophy, 2004; Eriks-Brophy, Durieux-Smith, Olds, Fitzpatrick, Duquette, & Whittingham, 2006; Fitzpatrick, Rhoades, Dornan, Thomas, & Goldberg, 2012; Hogan, Stokes, White, Tyskiewicz, & Woolgar, 2008; Rhoades, 2001, 2006; Rhoades & Chisholm, 2001; Yoshinaga-Itano, Sedey, Coulter, & Mehl, 1998).

### Background

Since the introduction of CIs in the United States for adults in 1985 and for children in 1990, there have been many changes in CI technology and surgical techniques and practices. In the early years, unilateral cochlear

implantation was the only option. By the early 2000s, a number of patients had received bilateral CIs in an effort to improve their “ability to localize sound and to understand speech in noise” (Litovsky et al., 2004, p. 648).

Initially there were professionals who questioned the notion that bilateral CIs could surpass “the impressive improvements in perception and linguistic development accrued from monaural implantation” (Papsin & Gordon, 2008, p. 69). But, as evidence has accumulated about the benefits of bilateral CIs, they have become a common treatment for patients with profound hearing loss in both ears (Bichey & Miyamoto, 2008; Scherf et al., 2009). Some patients receive bilateral sequential CIs, in which one ear is fitted with a CI and then several months (or years) later, in a separate surgery, the patient is fitted with a CI in the contralateral ear (Steffens et al., 2008). An even more recent development is bilateral simultaneous cochlear implantation, in which the patient receives CIs in both ears during a single surgery.

Interestingly, in the earliest years of pediatric cochlear implantation, many children did not continue to wear a hearing aid in the contralateral ear. Over time though, more patients became “bimodal” (i.e., one ear had a CI and the contralateral ear was fitted with a hearing aid). A

concern about sequential implantation has been that the ears are not receiving consistent auditory stimulation in tandem and a lag can develop between the function of the ears (and the brain) and that lag can become more pronounced over time (Bichey & Miyamoto, 2008).

Luntz, Shpak, and Weiss (2005) have stated, “there is a natural tendency for the contralateral residual acoustic hearing in CI candidates to diminish, possibly to the point of complete deafness” (p. 863). Another concern when an interval is created by delaying implantation of the second ear is that there may be negative effects on the patient’s binaural processing (Gordon, Valero, & Papsin, 2007; Papsin & Gordon, 2008). According to Gordon et al. (2007), most clinicians on CI teams in the United States attempt to minimize the interval between the first and second sequential implants.

With bilateral simultaneous cochlear implant surgery, lag time of auditory stimulation is not an issue. Additional benefits of bilateral hearing include the absence of binaural processing challenges, improved speech perception in noise, the summative effect of two ears, and the feeling of balance (Johnson, 2012).

A host of negatives has also been raised regarding bilateral simultaneous CIs, including: the potential negative sequelae of being under anesthesia for a longer time; possible surgical complications (no longer considered a viable complication, according to Gantz et al., 2002; Grainger, Jonas, & Cochrane, 2012; Ramsden, Papsin, Leung, James, & Gordon, 2009); and negative vestibular side effects (Mick, Friesen, Shipp, & Chen, 2012; Papsin & Gordon, 2008). Additionally, some parents want to “save one ear” in the hope that more advanced technologies, such as stem cells or better hearing technology might become available in the future (Zeitler et al., 2008).

It should also be mentioned that some pediatric patients do not want to be without sound for any period of time. For others, including patients with Auditory Neuropathy Spectrum Disorder (ANSD), some CI team members want to assess the outcomes of implanting a first CI prior to proceeding to a second CI (see Roush, Frymark, Venediktov, & Wang, 2011; Roush, 2011). Therefore, some patients with bilateral ANSD are only initially “approved” for a unilateral CI.

The purpose of this research investigation was to explore the opinions and practices of CI audiologists about sequential or simultaneous bilateral cochlear implants for children.

## Method

Audiologists working with pediatric cochlear implant candidates at cochlear implant centers throughout the United States were recruited for participation. The audiologists were contacted by email after using the

American Cochlear Implant Alliance (ACIA) institution membership listing along with the “Find a Clinician” search bar for each of the three U.S.-based CI manufacturers. A Territory Manager for Cochlear America also circulated the survey participation request to other managers in the United States.

The electronic, on-line survey was designed to collect information about audiologists’ opinions and practices regarding bilateral cochlear implantation with children. The survey was developed using Qualtrics. A total of 20 questions were set up using Likert-type scales and multiple-choice responses (4 demographic questions and 16 questions about past clinical experiences). A final open-ended question was also included. Prior to any data collection, the research project was reviewed and approved by the Human Subject Research Committee at the College of Wooster.

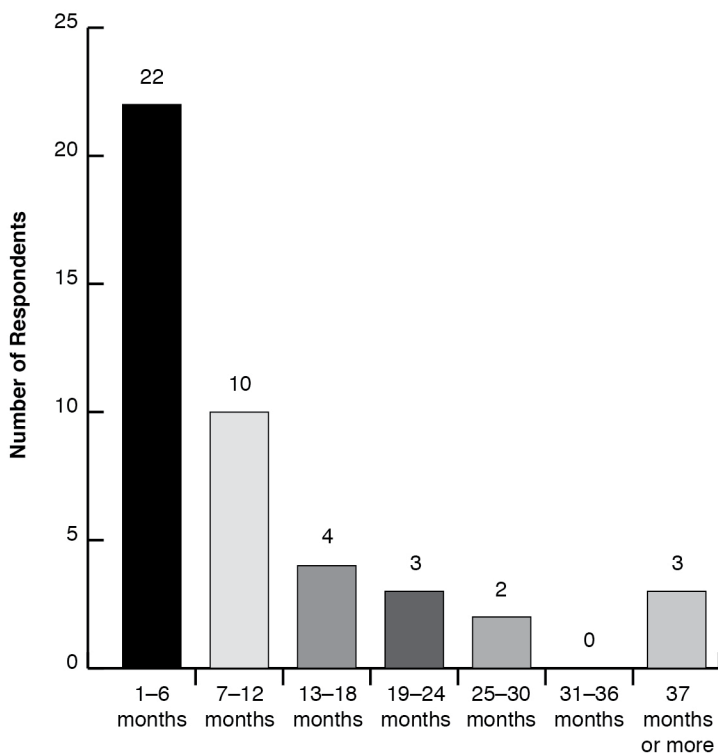
## Results

A total of 57 audiologists responded, although not all of the questions were answered by all of the respondents. All of the respondents held a Master’s degree (6%), a doctorate in Audiology (82%), or PhD or other doctoral degree (13%). Respondents varied in their years of experience: 31% with 6–10 years, 26% having 1–5 years, 20% with 11–15 years, 9% with 16–20 years, and 11% having more than 20 years of experience. Participants came from 21 different states and the most common work site was a hospital-based CI center (reported by 74% of the respondents). The number of pediatric bilateral recipients the audiologists had worked with over the last 12 months ranged from 10 respondents with 1–5 recipients, 13 having seen 6–10 patients, 4 with 11–15 recipients, and 17 who had worked with 16 or more recipients. Most of the patients seen by these audiologists had sequential cochlear implants (73%) and only 27% had simultaneous implants.

The most common interval between cochlear implants for the bilateral sequential patients was quite variable as shown in Figure 1. A total of 73% waited 1–6 months and 23% waited 7–12 months between the first and second implants. The shortest interval reported was 3 months.

The respondents felt that the most important benefits of bilateral hearing for children included localization, the summation effect, better hearing in noise, and reduced listening effort, as shown in Figure 2. Better hearing in noise was noted most frequently followed by reduced listening effort.

When asked why parents reported selecting sequential CIs over simultaneous CIs, the most common reasons



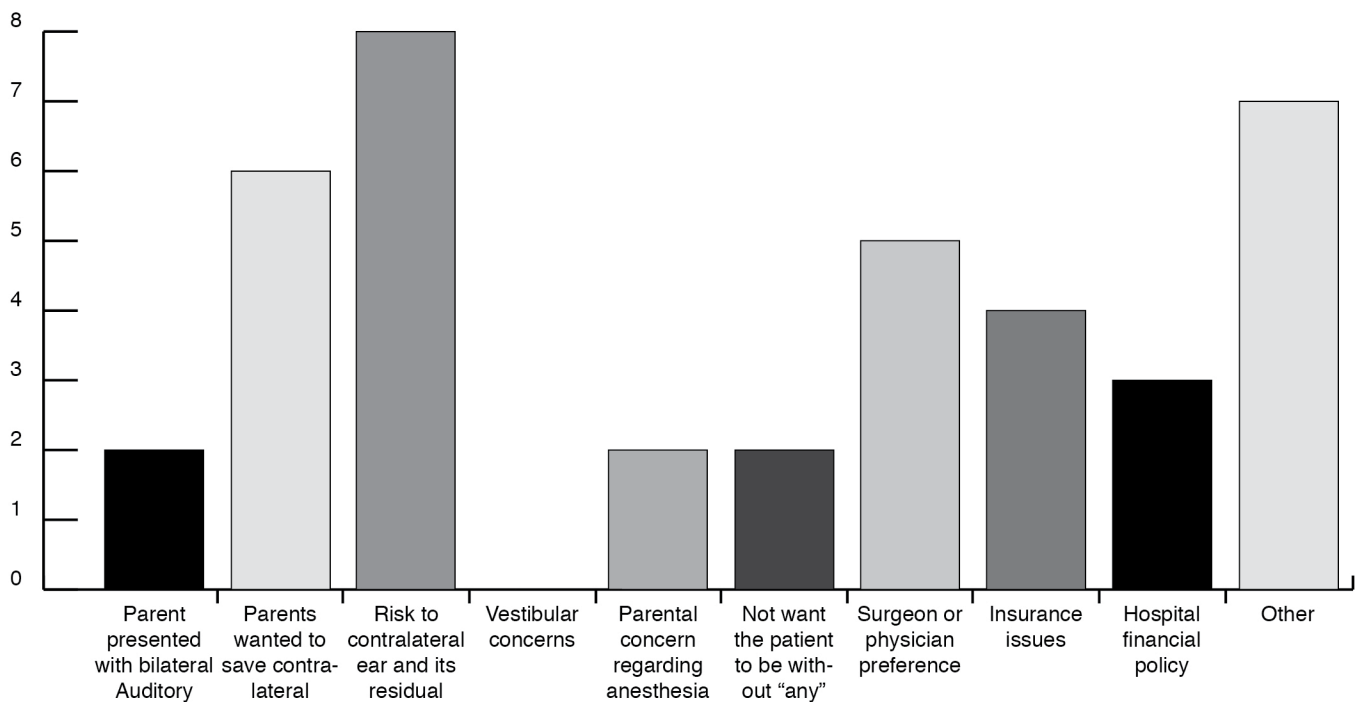
**Figure 1. Surgery interval between first and second cochlear implants for bilateral sequential patients.**

were concerns about risk to the contralateral ear (21%), parents wanting to save the contralateral ear for future technology (15%), surgeon or physician preference (13%), insurance issues (10%), and the fact that their facility simply did not do simultaneous surgeries (8%; see Figure 3).

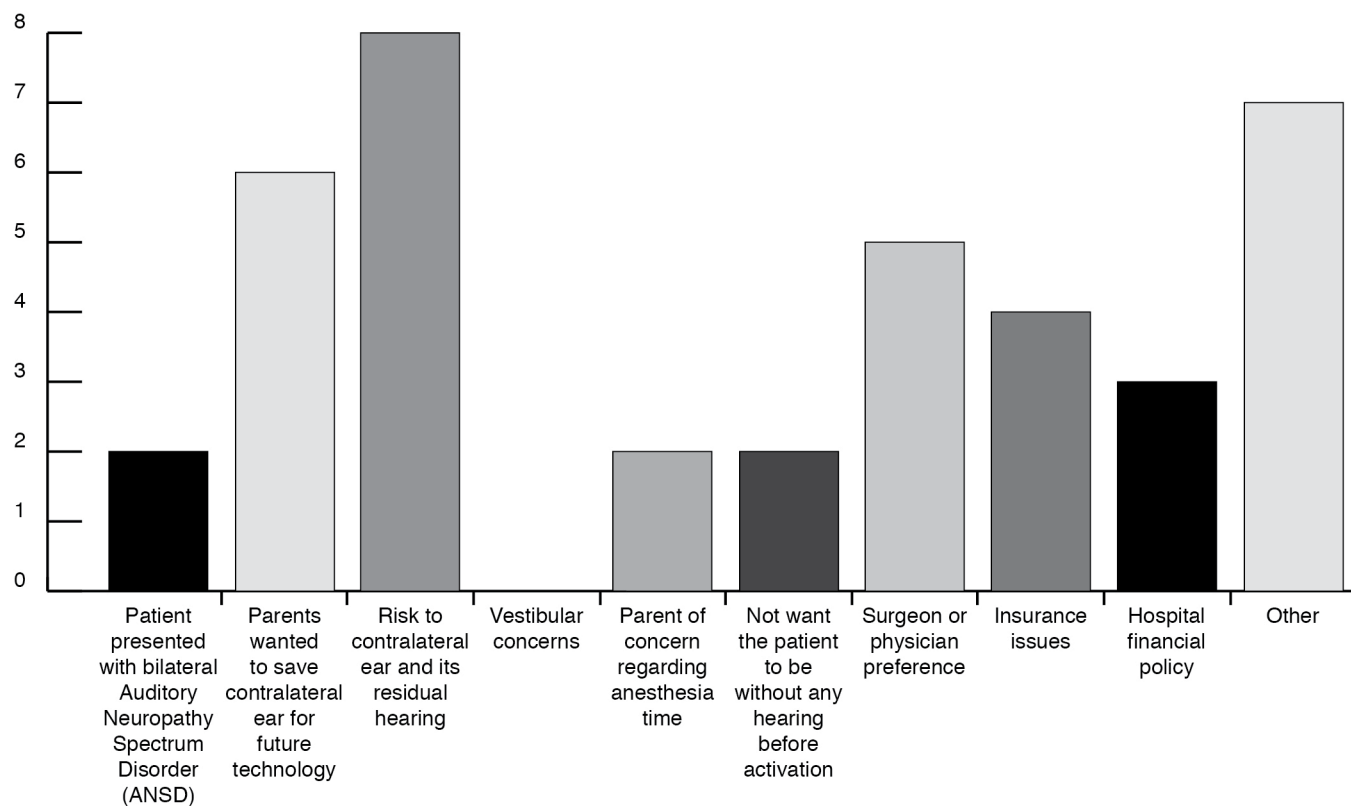
Table 1 shows Likert-type scale ratings (with 1 reflecting *least important* through 7 for *most important*) about why sequential implants might be preferred over simultaneous implants. The highest rated item was concern about the risk to the contralateral ear (mean rating of 5.1), followed by concern for patients with Auditory Neuropathy Spectrum Disorders (mean rating of 4.1).

As shown in Table 2, the most important factors impacting the decision-making process about whether to do a bilateral simultaneous implantation, included minimal benefit with hearing aids use (mean rating of 6.1) and that the child was profoundly deaf in both ears (mean rating of 6.0).

Respondents were also asked to evaluate the primary disadvantages of sequential versus simultaneous CIs. As seen in Figure 4, the most common disadvantage cited for sequential CIs, was *negatively impacts future binaural processing*.



**Figure 2. Audiologists' opinions about the primary benefits associated with binaural hearing.**



**Figure 3. Audiologists' opinions about why parents select sequential implantation over simultaneous implantation.**

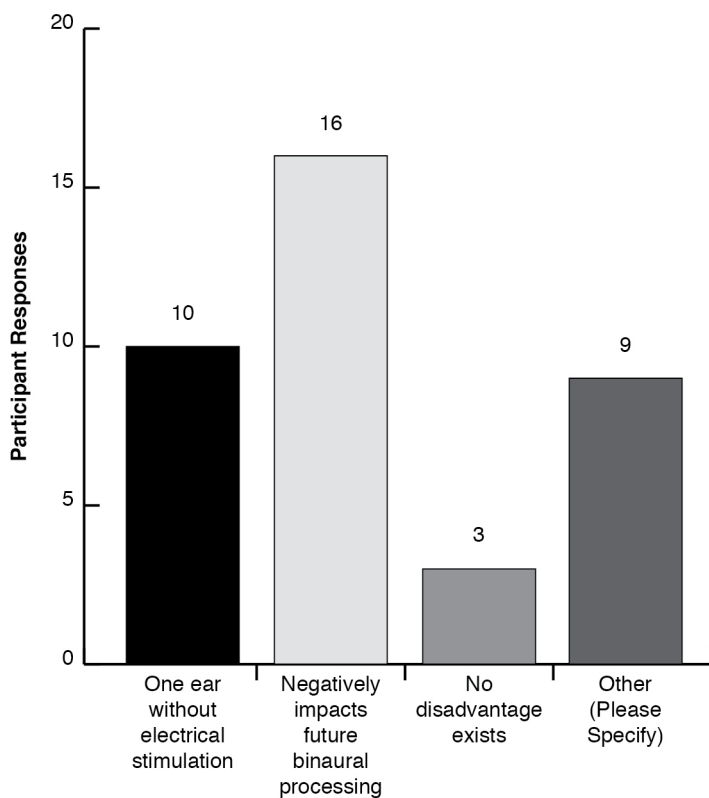
**Table 1**  
**Factors Impacting the Decision-Making Process for Bilateral sequential**

Factor	Least Important							Most Important	Mean	Standard Deviation
	1	2	3	4	5	6	7			
ANSD (both ears) (n=39)	10.2% (n=4)	20.5% (n=8)	15.4% (n=6)	10.2% (n=4)	7.7% (n=3)	17.9% (n=7)	17.9% (n=7)	4.10	2.10	
Save contralateral ear (n=39)	17.9% (n=7)	20.5% (n=8)	15.4% (n=6)	10.2% (n=4)	17.9% (n=7)	15.4% (n=5)	5.1% (n=2)	3.49	1.89	
Risk to contralateral ear (n=39)	0% (n=0)	10.2% (n=4)	10.2% (n=4)	12.8% (n=5)	20.5% (n=8)	17.9% (n=7)	28.2% (n=11)	5.10	1.68	
Vestibular concerns (n=39)	15.4% (n=6)	28.2% (n=11)	17.9% (n=7)	20.5% (n=8)	5.1% (n=2)	10.2% (n=4)	2.6% (n=1)	3.13	1.64	
Anesthesia (n=39)	10.2% (n=4)	15.4% (n=6)	20.5% (n=8)	17.9% (n=7)	7.7% (n=3)	25.6% (n=10)	5.1% (n=1)	3.85	1.77	
Being without hearing (n=39)	7.7% (n=3)	28.2% (n=11)	12.5% (n=5)	15.4% (n=6)	17.9% (n=7)	12.5% (n=4)	7.7% (n=3)	3.69	1.79	

Note. ANSD = Auditory Neuropathy Spectrum Disorder.

**Table 2**  
**Factors Impacting the Decision-Making Process for Bilateral sequential**

Factor	Least Important							Most Important	Mean	Standard Deviation
	1	2	3	4	5	6	7			
Minimal benefit with hearing aids (n=39)	2.6% (n=1)	2.6% (n=1)	2.6% (n=1)	2.6% (n=1)	7.7% (n=3)	25.6% (n=10)	56.4% (n=22)	6.13	1.44	
Profoundly deaf (both ears) (n=39)	5.1% (n=2)	5.1% (n=2)	0% (n=0)	2.6% (n=1)	7.7% (n=3)	17.9% (n=7)	61.5% (n=24)	6.03	1.72	



**Figure 4. Primary disadvantages of bilateral sequential cochlear implants.**

Respondents rated how frequently they recommended that their pediatric CI candidates should obtain bilateral sequential CIs, using a Likert-type scale of 1 (*never*) through 7 (*always*). The mean rating was 4.5 compared to a mean rating of 4.0 when asked about how frequently they recommended bilateral simultaneous CIs.

Additional questions focused on simultaneous CIs, beginning with a question about why clinicians definitely do not recommend the bilateral simultaneous option. *Asymmetrical hearing loss or contralateral ear had acoustically aid-able hearing* were cited by 40.5% as the main reasons for not proceeding immediately

to simultaneous CIs. Another 18.9% indicated that simultaneous CIs were not recommended when a patient presents with ANSD.

The option of having children wear a hearing aid on the non-implanted ear was almost universally endorsed, with 89.2% of the audiologists responding with yes for their patients going through the sequential route.

A final question asked if there were any questions or concerns not addressed in the survey. Although only 16 participants provided a response, the common theme of 7 audiologists was that they would recommend either bilateral sequential or bilateral simultaneous CIs on a case-by-case basis; and 3 respondents indicated that insurance approval dictates whether sequential or simultaneous implants are done.

### Discussion

All of the audiologists responding to the survey supported the use of bilateral versus unilateral hearing, with the most important identified value of binaural hearing being improved listening in noise, followed by reduced listening effort. The latter finding supports the work of Hughes and Galvin (2013) who similarly reported on reduced listening effort with binaural hearing.

Although the number of bilateral simultaneous CIs for the pediatric population have increased (Bichey & Miyamoto, 2008; Papsin & Gordon, 2008; Scherf et al., 2009), the majority of audiologists who responded to this survey were seeing and/or recommending more sequential versus simultaneous procedures for bilateral cochlear implants. But, in keeping with the long-held value of truly binaural hearing, the average interval between the patients receiving their first CI and their second CI was under 12 months.

The most frequent explanations for why the majority of the bilateral CI recipients were sequential instead of simultaneous was that many patients had only obtained



a unilateral CI when they were younger and now wanted to obtain a second ear. Other frequently mentioned rationales included concerns about the risk to the contralateral ear and its residual hearing, or parents who desired to save the non-implanted contralateral ear for future technological improvements. Additional reasons for the sequential CIs were asymmetrical hearing losses, need for insurance approval, and surgeon preference. Interestingly, a number of sequential recipients presented with ANSD, and the CI team members typically opted for sequential versus simultaneous CI management for these patients (see also Roush et al., 2011).

For those patients who received a unilateral CI, almost all continued to wear a hearing aid on the contralateral ear. This bimodal option presumably ensures that the non-implanted ear still receives some auditory stimulation and the potential for binaural hearing (Luntz et al., 2005). Ultimately, more and more of these bimodal patients, in a fairly short period of time, become bilateral CI candidates, and proceed to having a second cochlear implant so they are able to enjoy the benefits of binaural hearing.

Patients who received bilateral simultaneous CIs, most typically presented with an early diagnosis of a significant hearing loss and then embarked on a hearing aid trial. Other bilateral simultaneous CI recipients had a bilateral profound degree of hearing loss and were demonstrating minimal or no benefit from hearing aids, and soon were approved for CIs for each ear during one surgical procedure.

## Conclusions

Although the sample for this study was small and may not be representative of all cochlear implant audiologists in the country, it begins to explore some important issues that need to be evaluated by additional research with larger and more representative samples. Four preliminary conclusions can be drawn from this study:

1. All of the audiologists surveyed valued binaural hearing.
2. The majority of these audiologists most often recommended and worked with sequential patients. This finding must be considered with some caution, especially due to changing CI candidacy criteria through the years. Although most of the respondents were treating both bilateral sequential and simultaneous CI recipients, there were those who treated sequential CI recipients exclusively. Despite all the noted benefits of binaural hearing, sequential CIs were still the preferred method of implantation for the respondents to this survey.
3. The most common interval reported for the sequential CI patients was less than a year.

4. There were other outside influences and factors that often eliminated bilateral simultaneous CIs as an option—notably insurance and hospital policies. In addition, some parents continued to report that they were interested in saving the contralateral ear for future technological improvements.

The findings noted above suggest a number of issues that should be investigated further. One important issue is the amount of time between the first and second surgeries in sequential cochlear implants. Future studies should probe this topic with more discrete time frames regarding the interval; so instead of providing only the 1–6 months and 7–12 month interval choices, more options might include smaller interval lengths. In addition, a larger sample size should be strongly considered for future investigations. And finally, future studies should investigate evidence-based practices regarding the therapy options for bilateral simultaneous and sequential cochlear implants (Kuhn-Inacker, Shehata-Dieler, Muller, & Helms, 2004).

As we consider the changing landscape of deafness, the introduction of bilateral CIs appears to be a most important and positive development. If the sky is the limit for children who are deaf and hard of hearing, gaining access to truly binaural hearing will help in their journey to hear from both sides; and in so doing, optimize their speech, language, and auditory outcomes.

## References

- Bichey, B., & Miyamoto, R. (2008). Outcomes in bilateral cochlear implantation. *Otolaryngology-Head and Neck Surgery, 135*(5), 655–661.
- Dornan, D., Hickson, L., Murdoch, B., & Houston, T. (2007). Outcomes of an auditory-verbal program for children with hearing loss: A comparative study with a matched group of children with normal hearing. *The Volta Review, 107*, 37–54.
- Dornan, D., Hickson, L., Murdoch, B., & Houston, T. (2008). Speech and language outcomes for children with hearing loss educated in auditory-verbal therapy programs: A review of the evidence. *Communicative Disorders Review, 2*, 157–172.
- Eriks-Brophy, A. (2004). Outcomes of auditory-verbal therapy: A review of the evidence and a call for action. *The Volta Review, 104*, 21–35.
- Eriks-Brophy, A., Durieux-Smith, A., Olds, J., Fitzpatrick, E., Duquette, C., & Whittingham, J. (2006). Facilitators and barriers to the inclusion of orally educated children and youth with hearing loss: Promoting partnerships to support inclusion. *The Volta Review, 106*, 53–88.
- Fitzpatrick, E., Rhoades, E., Dornan, D., Thomas E., & Goldberg D., (2012). What are some of the evidence-based outcomes of auditory-verbal practice? In W. Estabrooks (Ed.), *101 frequently asked questions about auditory-verbal practice*. Washington, DC: AG Bell Association for the Deaf and Hard of Hearing.
- Gantz, B., Tyler, R., Rubinstein, J., Wolaver, A., Lowder, M., Abbas, P., ... & Preece, J. (2002). Binaural cochlear implants placed during the same operation. *Otology & Neurotology, 23*(2), 169–180.
- Gordon, K., Valero, J., & Papsin, B. (2007). Auditory brainstem activity in children with 9–30 months of bilateral cochlear implant use. *Hearing Research, 233*(1), 97–107.
- Grainger, J., Jonas, N., & Cochrane, L. (2012). Simultaneous cochlear implantation in children: The Great Ormond Street experience. *Cochlear Implants International, 13*(3), 137–141.

- Hammes, D., Novak, M., Rotz, L. A., Willis, M., Edmondson, D., & Thomas, J. (2002). Early identification and cochlea implantation: Critical factors for spoken language development. *Annals of Otolaryngology Rhinology Laryngology*, *111*, 74–78.
- Hogan, S., Stokes, J., White, C., Tyskiewicz, E., & Woolgar, A. (2008). An evaluation of auditory-verbal therapy using rate of early language development as an outcome measure. *Deafness and Education International*, *10*, 143–167.
- Hughes, K., & Galvin, K. (2013). Measuring listening effort expended by adolescents and young adults with unilateral or bilateral cochlear implants or normal hearing. *Cochlear Implants International*, *14*(3), 121–129.
- Johnson, C. (2012). Introduction to auditory rehabilitation: A contemporary issues approach. New York: Pearson.
- Kuhn-Inacker, H., Shehata-Dieler, W., Muller, J., & Helms, J. (2004). Bilateral cochlear implants: A way to optimize auditory perception abilities in deaf children? *International Journal of Pediatric Otorhinolaryngology*, *68*(10), 1257–1266.
- Litovsky, R., Parkinson, A., Arcaroli, J., Peters, R., Lake, J., Johnstone, P., & Yu, G. (2004). Bilateral cochlear implants in adults and children. *Archives of Otolaryngology—Head & Neck Surgery*, *130*(5), 648–655.
- Luntz, M., Shpak, T., & Weiss, H. (2005). Binaural-bimodal hearing: Concomitant use of unilateral cochlear implant and a contralateral hearing aid. *Acta Oto-Laryngologica*, *125*(8), 863–869.
- Mick, P., Friesen, L., Shipp, D., & Chen, J. (2012). Treatment outcomes of adult cochlear implantation. In M. Ruckenstein (Ed.), *Cochlear implants and other implantable hearing devices* (pp. 185–199). San Diego, CA: Plural Publishing.
- Papsin, B., & Gordon, K. (2008). Bilateral cochlear implants should be the standard for children with bilateral sensorineural deafness. *Current Opinion in Otolaryngology & Head and Neck Surgery*, *16*(1), 69–74.
- Ramsden, J., Papsin, B., Leung, R., James, A., & Gordon, K. (2009). Bilateral simultaneous cochlear implantation in children: Our first 50 cases. *The Laryngoscope*, *119*(12), 2444–2448.
- Rhoades, E. (2001). Language progress with an auditory-verbal approach for young children with hearing loss. *International Pediatrics*, *16*, 41–47.
- Rhoades, E. (2006). Research outcomes of auditory-verbal intervention: Is the approach justified? *Deafness and Education International*, *8*, 125–143.
- Rhoades, E., & Chisholm, T. (2001). Global language progress with an auditory-verbal approach for children who are deaf or hard of hearing. *The Volta Review*, *102*, 2–24.
- Roush, J., Frymark, T., Venediktov, R., & Wang, B. (2011). Audiologic management of auditory neuropathy spectrum disorder in children: A systematic review of the literature. *American Journal of Audiology*, *20*(2), 159–170.
- Roush, P. (2011). Children with Auditory Neuropathy Spectrum Disorders (ANSO). In R. Seewald & A. M. Tharpe (Eds.), *Comprehensive handbook of pediatric audiology* (pp. 731–750). San Diego, CA: Plural Publishing.
- Scherf, F., Van Deun, L., Van Wieringen, A., Wouters, J., Desloovere, C., Dhooge, I., ... & Van de Heyning, P. (2009). Functional outcome of sequential bilateral cochlear implantation in young children: 36 months postoperative results. *International Journal of Pediatric Otolaryngology*, *73*(5), 723–730.
- Steffens, T., Lesinski-Schiedat, A., Strutz, J., Aschendorff, A., Klenzer, T., Ruhl, S., ... & Lenarz, T. (2008). The benefits of sequential bilateral cochlear implantation for hearing-impaired children. *Acta Oto-Laryngologica*, *128*(2), 164–176.
- Vohr, B., Jodoin-Kruzak, J., Tucker, R., Johnson, C., Topol, D., & Ahlgren, M. (2008). Early language outcomes of early-identified infants with permanent hearing loss at 12 to 16 months of age. *Pediatrics*, *122*(3), 535–544.
- Yoshinaga-Itano, C., Sedey, A., Coulter, D. K., & Mehl, A. L. (1998). Language of early and later-identified children with hearing loss. *Pediatrics*, *102*, 1161–1171.
- Zeitler, D., Kessler, M., Terushkin, V., Roland Jr., J., Svirsky, M., Lalwani, A., & Waltzman, S. (2008). Speech perception benefits of sequential bilateral cochlear implantation in children and adults: A retrospective analysis. *Otology & Neurotology*, *29*(3), 324–325.

## Where Do We Go From Here? The Need for Genetic Referrals in Patients who are Deaf or Hard of Hearing: Findings from a Regional Survey

Tyson S. Barrett, PhD<sup>1</sup>  
Amanda Wylie, MSPH<sup>2</sup>  
Beth Vogel, MS<sup>3</sup>  
Kunal Sanghavi, MS<sup>4</sup>  
Melissa Raspa, PhD<sup>2</sup>  
Kathryn Aveni, RN, MPH<sup>5</sup>  
Kathryn Tullis, PhD<sup>6</sup>  
Michele Caggana, ScD<sup>3</sup>  
Karl White, PhD<sup>1,7</sup>  
Joann Bodurtha, MD<sup>8</sup>

<sup>1</sup>Utah State University

<sup>2</sup>RTI International

<sup>3</sup>Wadsworth Center, NYS Department of Health

<sup>4</sup>The Jackson Laboratory for Genomic Medicine

<sup>5</sup>New Jersey Department of Health

<sup>6</sup>Delaware Division of Public Health

<sup>7</sup>National Center for Hearing Assessment and Management

<sup>8</sup>Johns Hopkins University

**Abstract: Purpose.** The purpose of this study was to assess primary health care providers' knowledge and use of genetic services for children whose hearing screening indicates they may be deaf or hard of hearing (DHH) and identify areas in which health care providers can be supported to increase family education and referral of families for genetic consultation.

**Methodology.** A survey was developed on current practices, knowledge, and perceived beliefs regarding genetic education and referrals for deafness. The surveys were distributed to pediatricians, family medicine physicians, nurse practitioners, and physician assistants in Washington DC, Delaware, Maryland, New Jersey, New York, Pennsylvania, Virginia, and West Virginia.

**Results.** Among 266 respondents, 80% were uninformed about Early Hearing Detection Intervention (EHDI) 1-3-6 guidelines prior to taking the survey. Approximately 55% were not confident about the genetic causes of deafness, 44% rarely consulted genetics professionals, 41% had not referred families to genetics, and 37% were not confident about the importance of genetic referrals.

**Conclusions.** Integrated, targeted, and user-friendly genetics education strategies in the existing EHDI framework are needed to ensure adequate awareness and delivery of genetics services for children who are DHH.

**Key Words:** genetics, hearing loss, deafness, EHDI, provider survey, needs assessment

**Acronyms:** AAP = American Academy of Pediatrics; ACMG = American College of Medical Genetics and Genomics; CDC = Centers for Disease Control & Prevention; DHH = deaf or hard of hearing, EHDI = Early Hearing Detection and Intervention; EI = early intervention; HL = hearing loss; HRSA = Health Resources and Services Administration; LEND = Leadership Education in Neurodevelopmental and Related Disabilities; MCHB = Maternal & Child Health Bureau; MMS = Medical Marketing Services, Inc.; NBHS = Newborn Hearing Screening; NCHAM = National Center for Hearing Assessment and Management; NYMAC = New York–Mid-Atlantic Consortium for Genetic & Newborn Screening Services

**Acknowledgements:** This project was supported by the Health Resources and Services Administration (HRSA) of the U.S. Department of Health and Human Services (HHS) under Grant #H46MC24094 (6/1/12–5/31/17), New York–Mid-Atlantic Consortium for Genetic and Newborn Screening Services for \$600,000 per year, and under Cooperative Agreement # UH7MC30773 (6/1/17–5/31/20), Regional Genetics Networks, for \$600,000 per year. This information or content and conclusions are those of the authors and should not be construed as the official position or policy of, nor should any endorsements be inferred by HRSA, HHS, or the U.S. Government.

**Correspondence concerning this article should be addressed to:** Tyson S. Barrett, 2810 Old Main Hill, Logan, UT 84341. Email: tyson.barrett@usu.edu



Approximately 3 to 4 per 1000 infants are born each year in the United States whose hearing thresholds indicate they are moderately, severely, or profoundly deaf or hard of hearing (DHH; Mercer, 2015). Early Hearing Detection and Intervention (EHDI) systems work to ensure timely identification and intervention for infants and toddlers who are DHH and include public health surveillance of newborn hearing screening (NBHS). The EHDI system has promoted 1-3-6 guidelines, recommending newborn screening no later than 1 month of age, evaluation of those infants who do not pass their hearing screening by 3 months of age, and enrollment in early intervention by 6 months of age. 1-3-6 has been shown to positively impact speech and language development for those identified as DHH (Moeller, White, & Shisler, 2006). More than 95% of all newborns<sup>1</sup> in the United States have their hearing screened at birth through NBHS with federal and state support (Muñoz, Shisler, Moeller, & White, 2009).

Progress in genetics has led to the identification of multiple genes causing non-syndromic and syndromic impacts on hearing levels, with over 400 genes now identified (Toriello, Reardon, & Gorlin, 2004). The majority of genetic causes (about 70%) are non-syndromic with more than half of identifiable variants or gene changes in two genes, *GJB2* and *GJB6*. These genes are associated with moderate to profound bilateral, sensorineural, and non-progressive impacts on hearing levels (Shearer, Hildebrand, & Smith, 2017). About 30% of children who are DHH with a genetic component have associated physical and clinical features such as retinitis pigmentosa (Usher syndrome), inner ear deformities and thyroid goiter (Pendred syndrome), cardiac arrhythmias (Jervell and Lange-Nelson syndrome), and renal malformations (Branchiootorenal syndrome; Shearer et al., 2017). The co-morbidities associated with these syndromes warrant additional medical assessments in newborns who are DHH. Previous studies have described how genetics evaluations can be incorporated into the EHDI process and benefit the parents of children with hearing impairment (Mercer, 2015; Muñoz et al., 2009; Schimmenti et al. 2004; White, 2004).

The benefits of an early genetic evaluation of newborns who are identified as DHH are numerous. First, a genetic evaluation can help families understand why their child is DHH, potentially reducing unnecessary concerns. Second, genetic evaluation can provide families with additional information about the services that might be helpful for their child and family. Given this information, families may consider reaching out to other families who have children with similar genetic backgrounds, building a stronger support system. Third, genetic evaluation can support a more thorough formation of a personalized medical care plan, thereby empowering families to obtain better care. This formation of a medical care plan can also provide additional connections to necessary medical and psychosocial support services. Further, an early genetic evaluation may help families better understand the link between hearing loss and genetics, and can

provide an opportunity to discuss recurrence risk with genetic professionals.

Recent literature indicates that genetic services are under-utilized. A 2005 survey conducted by the National Center for Hearing Assessment and Management (NCHAM) indicated only 8.8% of physicians ( $n = 1,968$ ) routinely refer a child who is DHH to a geneticist (Moeller et al., 2006). The referral rate for audiologists and otolaryngologists was higher, with 56% of audiologists reporting that they would *often* or *always* refer a hearing couple with one child who is DHH for genetic counseling (Connelly, 2010). When asked what initial set of tests they use in an infant with confirmed nonsyndromic sensorineural hearing loss, 49% of otolaryngologists reported they order a genetics evaluation (Duncan, Prucka, Wiatrak, Smith, & Robin, 2007). A simulation-based survey found that 37% of otolaryngologists and geneticists ordered a genetic consultation on the first encounter of sensorineural hearing loss and 30% did so on the second encounter (Jayawardena, Shearer, & Smith, 2015). Although physicians recognize the importance of genetics evaluation for children that are DHH, various challenges continue to persist in making physician referrals to genetics services. The 2005 NCHAM survey found that 90% of physicians perceived there being *somewhat of a need* or *a great need* for training and/or resources on genetics and DHH (Moeller et al., 2006). Furthermore, physicians have cited lack of appropriate education or training to make referrals and uncertainty about the usefulness of genetics (Connelly, 2010).

Because of the known under-utilization of genetic services by primary care providers caring for individuals who are DHH, we sought to assess primary health care providers' knowledge and use of genetic services for children who are DHH. We further sought to identify areas in which health care providers can be supported in increasing family education and referral of families for genetic consultation. The long-term goal of this study is to improve services for infants and children who are DHH as well as their families by integrating genetic services into the management of patients who are DHH.

## Method

### Instrument

The hearing loss needs assessment was developed through a collaboration between the New York–Mid-Atlantic Consortium for Genetic & Newborn Screening Services (NYMAC) and the National Center for Hearing Assessment and Management (NCHAM). NYMAC is one of seven regional genetics collaboratives funded by the Health Resources and Services Administration (HRSA). The region encompasses the District of Columbia, Delaware, Maryland, New Jersey, New York, Pennsylvania, Virginia, and West Virginia. Two previous NCHAM surveys (distributed in 2005 and 2012),

<sup>1</sup>An updated percentage of newborns receiving hearing screening can be found at <https://www.cdc.gov/ncbddd/hearingloss/ehdi-data2015.html>

literature reviews, and ongoing discussions with the NYMAC region EHDI coordinators and the Leadership Education in Neurodevelopmental and Related Disabilities (LEND) faculty guided the design of the survey. The survey was designed to identify the unmet needs of children, families, and professionals regarding use of genetics in hearing loss screening, diagnosis, and referral.

The 10–20 minute survey was available in paper and electronic format and consisted of 32 questions divided into four sections: (a) demographic information, (b) knowledge and beliefs about genetic referrals for children with hearing loss, (c) current practice regarding referrals for children with hearing loss, and (d) resources and strategies needed (see Appendix for a copy of the survey). The survey also included links to available resources, including the American College of Medical Genetics and Genomics (ACMG) Action Sheet on patients who are DHH and genetics. Notably, DHH was defined as permanent, bilateral or unilateral, sensorineural or conductive hearing loss of an average loss of 30 decibels or more in the frequency range important for speech recognition.

The Johns Hopkins University and the Utah State University Institutional Review Boards approved the study.

### Participants and Procedures

EHDI coordinators in the NYMAC region were contacted by NCHAM with study information, the paper survey, a pre-addressed and stamped envelope, instructions for completion, and a URL to the electronic version of the survey. EHDI coordinators then contacted pediatricians, family medicine physicians, nurse practitioners, and physician assistants within their states to invite them to complete the needs assessment. Respondents were encouraged to share the survey link with fellow providers to enhance uptake. Respondents were asked to complete only one survey (paper or electronic). Contact information for NYMAC and NCHAM was provided.

A follow-up email reminder to the target population was sent via Medical Marketing Services, Inc. (MMS), a professional service provider. MMS estimated that 17,974 physicians, 4,837 advance practice nurse or nurse practitioners, and 1,373 physician assistants received the email blast. Two written reminders and one e-mailed reminder were sent to EHDI coordinators to encourage providers in their states to complete the needs assessment. The paper survey was re-sent to providers in the state of Delaware only.

### Analytic Strategy

Responses to demographic questions, questions related to knowledge and beliefs about genetic hearing loss referrals, current practices for hearing loss referrals, and

resources and strategies are reported. All comparisons across groups (i.e., disciplines) herein were carried out using chi-square tests of independence. All analyses were conducted using the R statistical environment version 3.4.2 (R Core Team, 2018).<sup>2</sup>

Differences in the number of genetic referrals, timing of genetic referrals, and reasons for genetic referrals were examined by professional discipline. Differences in the perceived frequencies of challenges (*not a challenge, sometimes a challenge, always a challenge, not applicable*) relating to typical challenges experienced by clinical professionals were examined by reasons for the genetic referrals as well.

## Results

### Sample Characteristics

A total of 266 participants across 8 states and the District of Columbia completed the survey. Participants were allowed to skip questions; therefore, the response rate varied per question. Table 1 presents demographic information regarding the participants, including their specialty, experience, and practice information. Of the respondents, 47% were pediatricians, 53% worked in private practice, most (68%) worked in either a large or small metropolitan area, and 42% had more than 20 years of experience.

**Table 1**  
*Demographic Information about the Participants*

	Percent Total N = 266
Specialty	<i>n</i> = 247
Family Medicine Physician	18%
Neonatologist	3%
Nurse Practitioner	17%
Pediatrician	47%
Physician Assistant	11%
Other	4%
Practice Setting	<i>n</i> = 244
Community clinic	13%
Federally Qualified Health Centers	7%
Hospital setting	15%
Medical school or parent university	3%
Private practice	53%
Other	9%
Practice Location	<i>n</i> = 240
Large metropolitan area	34%
Small metropolitan area	34%
Rural area	11%
Small town	20%
Years of Experience	<i>n</i> = 239
Less than 1 year	5%
1-2 years	7%
3-5 years	9%
6-9 years	13%
10-19 years	25%
More than 20 years	42%

<sup>2</sup>All code and data used in the present study are provided at: <https://osf.io/8thwv/>

### Provider Knowledge and Beliefs

A total of 266 participants across 8 states and the District of Columbia completed the survey. Participants were allowed to skip questions; therefore, the response rate varied per question. Table 1 presents demographic information regarding the participants, including their specialty, experience, and practice information. Of the respondents, 47% were pediatricians, 53% worked in private practice, most (68%) worked in either a large or small metropolitan area, and 42% had more than 20 years of experience.

Table 2 shows the responses regarding the participants' levels of confidence in speaking with parents of a child with permanent hearing loss about the genetic causes (52% not confident), the importance of genetic referrals (33% not confident), the logistics of genetic referrals (3% not confident), and the significance of genetics in hearing loss due to ototoxic medication exposure (43% not confident).

### Current Practice

Approximately 41% of the participants have referred a family to a genetics specialist because there was a family history of hearing loss, 4.3% because the parents

**Table 2**  
*Responses about the Confidence Level in Speaking with Parents of a Child with Permanent Hearing Loss about Genetic Causes of Hearing Loss and Genetic Referrals*

	Percent Total N = 266
Genetic causes of hearing loss	n = 225
Not confident	52%
Somewhat confident	40%
Very confident	8%
The importance of genetic referrals	n = 222
Not confident	33%
Somewhat confident	44%
Very confident	23%
The logistics of genetic referral for hearing loss	n = 221
Not confident	39%
Somewhat confident	43%
Very confident	18%
The significance of genetics in hearing loss due to ototoxic medication exposure	n = 221
Not confident	42%
Somewhat confident	39%
Very confident	17%

**Table 3**  
*Referrals by Discipline*

	Pediatrician n = 116	Nurse Practitioner n = 41	Family Medicine Physician n = 45	Physician Assistant n = 27	Neo- natologist n = 8	Other n = 10	P-value
How many genetic referrals have been made							0.323
0-10	97%	100%	100%	100%	75%	100%	0-10
11-20	1%	0%	0%	0%	25%	0%	11-20
50+	1%	0%	0%	0%	0%	0%	50+
When genetic referral is made							0.178
At diagnosis	6%	22%	43%	0%	0%	0%	
Enroll in EI	71%	67%	29%	50%	100%	100%	
Screening	14%	0%	29%	50%	0%	0%	
Other	9%	11%	0%	0%	0%	0%	
Reason for Genetic Referral							<.001
I have not referred families to a genetics professional for hearing loss	16%	49%	54%	88%	14%	50%	
Parents are discussing another pregnancy	5%	5%	5%	0%	0%	0%	
Parents ask for a referral	3%	5%	7%	4%	0%	0%	
There is a family history of hearing loss (parents, siblings, cousins, etc.)	58%	33%	22%	4%	43%	38%	
Other	18%	8%	12%	4%	43%	13%	

Note. EI = Early Intervention. Due to missing values in responding discipline, this table includes 247 responses.

were discussing another pregnancy, and 3.9% because the parents asked for a referral. However over a third (37.1%) of the participants stated that they had not made a genetic referral. Nearly 40% of all participants indicated they have not consulted with a genetics specialist without a formal referral, with another 39% rarely consulting with a genetics specialist. The majority of participants (59%) had never ordered eConnexin 26 testing in infants with nonsyndromic hearing loss.

Table 3 shows the number of referrals, when referrals are made, and for what reason by discipline. Although there was a statistically significant difference of the reason for referral by discipline ( $p < .001$ ), further analyses showed that physician assistants (12%), family medicine physicians (46%), and nurse practitioners

(51%) were less likely to have made a referral as compared to pediatricians (84%;  $p < .001$ ). Beyond this, there were no differences between disciplines regarding reasons for a referral ( $p = .435$ ).

In addition, several participants indicated that they did not have tracking in place for failed newborn hearing screening (30%), failed newborn blood spot screening (26%), delayed developmental milestones (31%), and follow-up after referrals (32%). Of those that received reports of the newborn hearing screening, nearly 80% say they sometimes or often refer children to a genetics referral. Notably, however, this differed by discipline with family medicine physicians rarely making a genetic referral in these situations compared to other professionals ( $p = .002$ ).

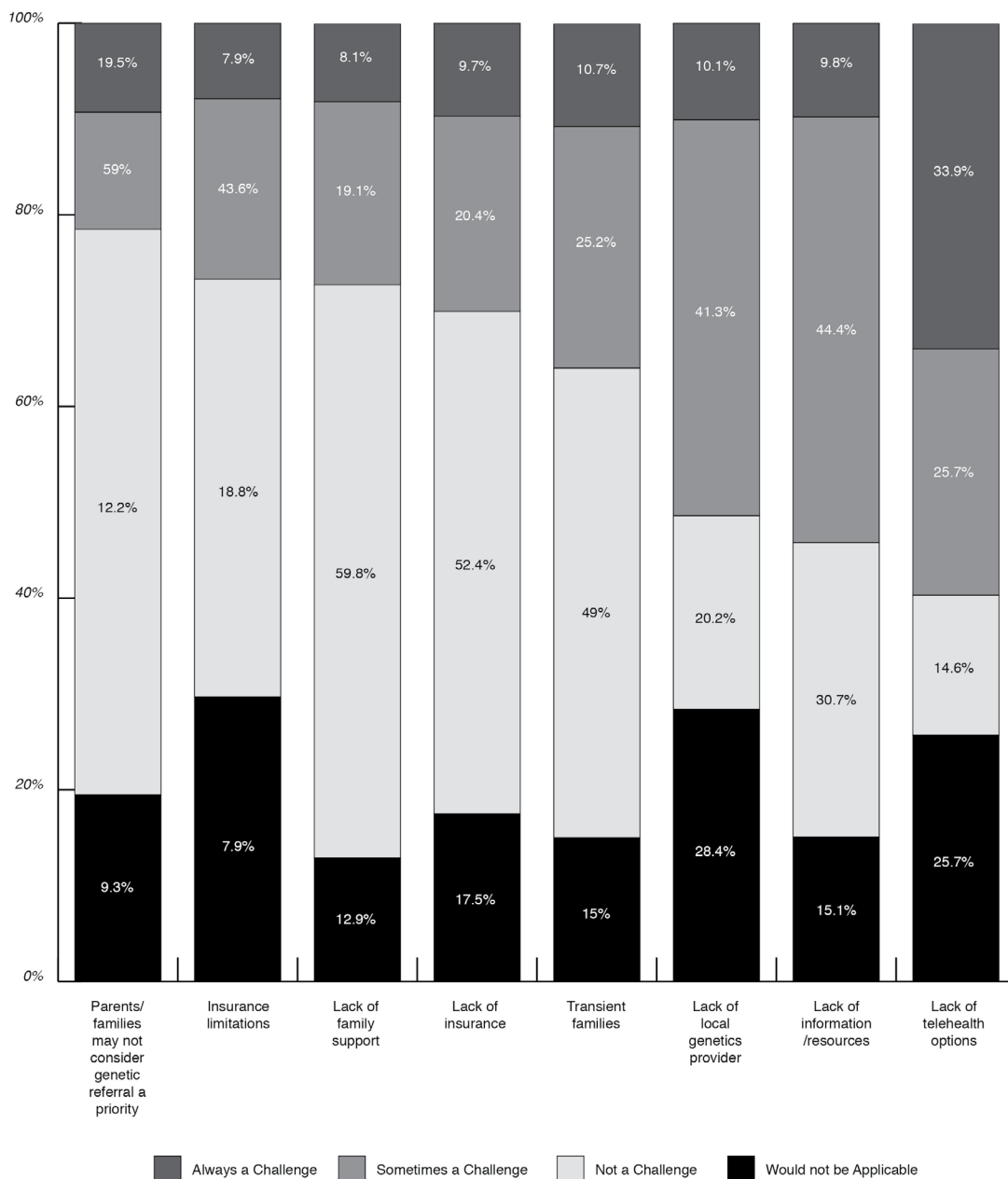


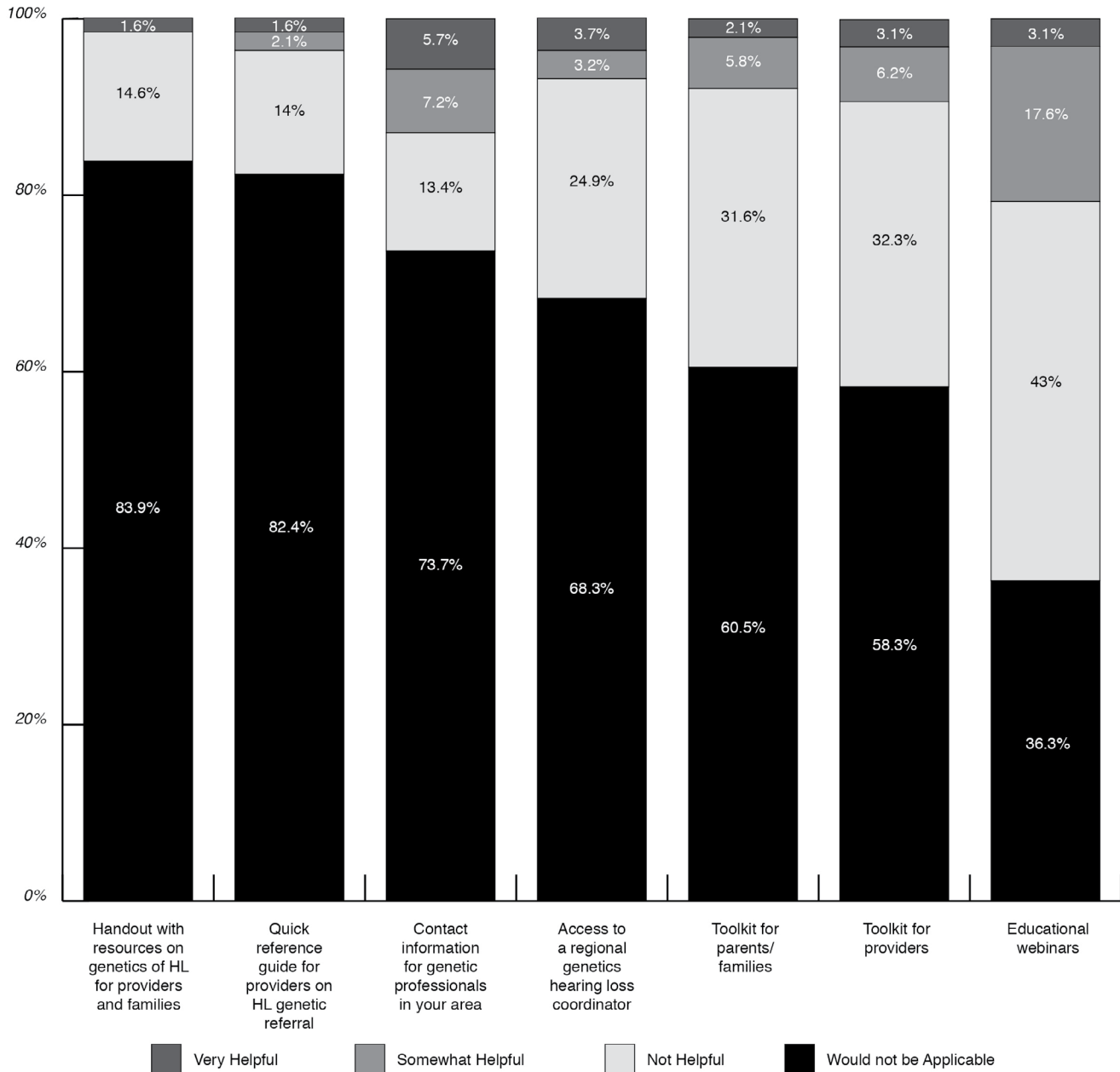
Figure 1. Responses regarding challenges faced about the genetics of hearing loss.

## Challenges and Opportunities

A final question was asked regarding challenges professionals face in respect to making genetic referrals for hearing loss (see Figure 1). The greatest challenge perceived by the survey participants when making genetic referrals were the parent/family priorities about genetic referrals (78.5% indicating *always or sometimes a challenge*) followed by the family's inability to attend an appointment due to logistics, such as transportation or child care coverage (72.7% saying *always or sometimes a challenge*). More than half of participants indicated that lack of information to make the referral was a challenge as well. Neonatologists (83%) and

pediatricians (65%) said lack of information/resources was not a challenge compared to family medicine physicians (11%), nurse practitioners (21%), and physician assistants (24%;  $p < .001$ ).

A majority (84%) reported that a handout with resources on genetics of hearing loss for providers and families would be very helpful (Figure 2). Similar responses were given for both a quick reference guide about genetic referrals (82%) and contact information about genetic professionals in their area (74%). Educational webinars were far less popular with only 36.3% of participants indicating that webinars would be very helpful.



**Figure 2. Perceived needs of the participants regarding training, information, and other resources about genetics and hearing loss (HL).**



## Discussion

These results strongly support the need for education on genetics and patients who are DHH for primary care providers, as 43% of respondents indicated they lack information on this topic. Data from the current study supports a prior physician survey on congenital hearing loss, in which approximately 40% of respondents perceived a great need for training and/or resources on genetics and DHH (Moeller et al., 2006).

In addition to education on genetics and patients who are DHH, there is a need for public health surveillance to ensure newborns identified as DHH through hearing screening have a genetic evaluation. As noted in our results, many participants do not have tracking mechanisms in place for NBHS and other infant health processes. However, several states in the NYMAC region do have existing mechanisms for incorporating genetics into their EHDI program activities. In Virginia, parents are called after a newborn is identified as DHH to verify enrollment in early intervention services and the completion of the diagnostic work-up, including a genetics evaluation. In New Jersey, primary care providers are sent a checklist of necessary post-diagnosis evaluations including genetics after identification. In Delaware, all newborns are referred to audiology at a single site, which allows for tracking of the post-diagnostic work-up. Going forward, HRSA-funded EHDI programs will be held responsible for improving care coordination through the patient/family-centered medical home model. Programs are required to report the number of care coordination plans developed with the parent or family and the number of care coordination plans that are shared across providers. This new, funded activity provides EHDI programs an opportunity to incorporate genetic evaluations into care plans and to include the sharing of care plans with the genetics provider.

Primary care provider education and public health surveillance are key to improving access to genetic services for newborns that are DHH, but based on our results, other barriers exist. Although we found that primary care providers perceive family acceptance as a barrier, a previous survey of parents of children who are DHH indicated that about 96% of parents—of whom none were DHH and about a quarter reported a family member born deaf—had a positive attitude toward genetic evaluation. A broader community of hearing, deaf, and hard-of-hearing adults supported genetic evaluation for newborns and expressed their willingness to engage in genetic counseling. Most families of children who are DHH value the availability of genetic testing as a means of better understanding the cause of the hearing loss and promoting discussion of the condition. (Geelhoed, Harrison, Davey, & Walpole, 2009).

## Response to Needs

In response to the identified need for genetics education, NYMAC conducted an educational campaign using professional marketing (Figure 3) from May 1, 2017 through May 29, 2017. The campaign targeted pediatricians from New York, Delaware, Washington DC, Maryland, New Jersey, Pennsylvania, Virginia, and West Virginia. Standard desktop and tablet banners were placed on the American Academy of Pediatrics and AAP Publications websites. The campaign reached 215,949 pediatricians. This campaign represents one approach to provide education on genetics and hearing loss, but novel methods for ongoing education is needed—not only for this topic, but for genetics as a whole. In the survey, providers indicated a preference for handouts (83.9%), a quick reference guide (82.4%), and contact information about genetic professionals (73.7%). Although preparation of these materials is straightforward, incorporating them into practice is likely to be more challenging.



*Figure 3. Ad used for educational campaign on the effect of genetics on being deaf or hard of hearing in the New York–Mid-Atlantic Consortium for Genetic & Newborn Screening Services Region.*

Results also showed that providers perceive appointment logistics as a barrier to families receiving genetics evaluations. NYMAC is piloting several methods to reduce barriers related to appointment logistics. These methods include a phone line for help identifying a genetic service provider, expansion of telegenetics services, primary care provider education, and a formal relationship between primary care providers and a geneticist to review cases. The HRSA-funded Regional Genetics Networks are piloting different approaches to improve access to genetic services. Individuals with a variety of genetic conditions, including children who are DHH, will benefit from these nationwide activities.

Notably, there are a number of limitations to the study. First, the survey response rate was low, with only 266 of more than 24,000 professionals returning the survey. This indicates that the sampling may be biased toward certain groups. It is therefore difficult to assess the generalizability of this single sample. However, this may not be a major limitation given the corroboration between these results and prior results mentioned earlier. Second, there were missing values for many questions. For all tables, the number of participants that answered each question was noted.

## Conclusion

Ultimately, this study may highlight the challenges and needs for healthcare professionals in their work with children who are DHH. Results indicate that there is a need for integrated, targeted, and user-friendly genetics education strategies for providers of children who are DHH, to ensure adequate awareness and delivery of genetics services for these children. This could include early intervention providers, as they may be able to encourage families to learn more about the genetic evaluation process if they have not pursued this. With recommendations coming from multiple sources, parents/family members may progressively become more interested in understanding their child's genetic background.

## References

- Connelly, A. (2010). Practicing Audiologists' Confidence in Their Understanding of Genetics and the Genetic Counseling Process. Retrieved from ProQuest Dissertations & Theses. (AAT 3436849)
- Duncan R. D., Prucka, S., & Wiatrak, B. J. (2007). Pediatric otolaryngologists' use of genetic testing. *Archives of Otolaryngology— Head & Neck Surgery*, 133(3), 231–236.
- Geelhoed, E. A., Harrison, K., Davey, A., & Wolpole, I. R. (2009). Parental perspective of the benefits of genetic testing in children with congenital deafness. *Public Health Genomics*, 12(4), 245–250.
- Jayawardena, A. D. L., Shearer, A. E., Smith, R. J. H. (2015). Sensorineural hearing loss—A changing paradigm for its evaluation. *Archives of Otolaryngology— Head & Neck Surgery*, 153(5), 843–850.
- Mercer, D. (2015). Guidelines for audiologists on the benefits and limitations of genetic testing. *American Journal of Audiology*, 24, 451–461.
- Moeller, M. P., White, K. R., & Shisler, L. (2006). Primary care physicians' knowledge, attitudes and practices related to newborn hearing screening. *Pediatrics*, 118(4), 1357–1370.
- Muñoz, K., Shisler, L., Moeller, M., & White, K. (2009). Improving the quality of early hearing detection and intervention services through physician outreach. *Seminars in Hearing*, 30(3), 184–192.
- R Core Team. (2018). R: A language and environment for statistical computing. R Foundation for Statistical Computing, Vienna, Austria. Retrieved from <https://www.R-project.org/>
- Schimmenti, L. A., Martinez, A., Fox, M., Crandall, B., Shapiro, N., Telatar, M. ... Palmer, G. (2004). Genetic testing as part of the Early Hearing Detection and Intervention (EHDI) process. *Genetics in Medicine*, 6(6), 521–525.
- Shearer, A. E., Hildebrand, M. S., & Smith, R. J. H. (2017). Hereditary hearing loss and deafness overview. In M. P. Adam, H. H. Ardinger, R. A. Pagon, & S. E. Wallace (Eds.), GeneReviews® [Internet]. Seattle (WA): University of Washington, Seattle; 1993–2017. Retrieved from <https://www.ncbi.nlm.nih.gov/books/NBK1116/?term=hereditary+hearing+loss>
- Toriello, H. V., Reardon, W., & Gorlin, R. J. (Eds.) (2004). *Hereditary Hearing Loss and its Syndromes*. New York: Oxford University Press.
- White, K. R. (2004). Early hearing detection and intervention programs: Opportunities for genetic services. *American Journal of Medical Genetics*, 130A: 29–36.

## Appendix

The following is what was presented to the online surveys. The paper surveys are extremely similar in appearance.

Each year, 3 in 1,000 infants are born in the US with moderate, severe, or profound hearing loss (HL). By age 19, 15% of adolescents have HL in one or both ears. Newborn hearing screening (NBHS) is included in the Recommended Universal Screening Panel for newborns. The national and state Early Hearing Detection and Intervention (EHDI) programs funded by the Centers for Disease Control & Prevention (CDC) and the Maternal & Child Health Bureau (MCHB) of the Health Resources and Services Administration (HRSA), in cooperation with professional societies, support families of children with HL and their providers based on the EHDI 1-3-6 guidelines:

- Screening by 1 month of age
- Diagnosis of HL by 3 months of age
- Entry into early intervention (EI) services by 6 months of age

The objectives of this research survey are to

- Determine the unmet needs of physicians and health care providers related to genetics services for children in the HL screening, diagnosis, and referral continuum and
- Identify areas where appropriate assistance can be provided to support physicians to increase family education about and genetic referrals for HL.

This research survey's long-term goal is to use the findings to improve services for infants and children with HL as well as their families by integrating genetic services into the management of patients and families with HL.

Your completion of this survey or questionnaire will serve as your consent to be in this research study. Please take about 10–20 minutes to tell us about your experiences. Your responses are completely confidential and will be used to improve services for infants and young children with hearing loss. Your participation is greatly appreciated.

1. Please indicate your profession:

- Pediatrician
- Family Medicine Physician
- Otolaryngologist (ENT)
- Neonatologist
- Nurse Practitioner
- Physician Assistant
- Resident/Fellow (specify) \_\_\_\_\_
- Other \_\_\_\_\_

2. Practice setting where you spend most of your time:

- Private Practice
- Community clinic
- Hospital setting
- Medical school/parent university
- Federally Qualified Health Centers
- Other \_\_\_\_\_



3. Practice Location:

- Small metropolitan area
- Large metropolitan area
- Small town
- Rural Areas

4. State/District:

---

5. Year(s) of practice with pediatric population:

- Less than a year
- 1–2
- 3–5
- 6–9
- 10–19
- More than 20 years

**Hearing Screening Genetics**

*Hearing Loss* that is permanent, bilateral or unilateral, sensorineural or conductive, and averaging loss of 30 decibels or more in the frequency range important for speech recognition. The following questions are about children who were identified through newborn hearing screening (NBHS) as having hearing loss.

6. Do you receive reports about children who have failed their newborn hearing screening (NBHS)?

- Yes
- No

7. Have you referred parents who have a child with hearing loss identified through NBHS to genetics professionals? (*If you answered "No" to this question, you will skip to Question #11*)

- Yes
- No

8. If/When you have a child with hearing loss identified through NBHS, how often do you refer the parents to genetics professionals?

- Rarely
- Sometimes
- Often
- Unsure
- Not applicable

9. In the last year, approximately how many patients did you refer for genetic evaluation of hearing loss after an abnormal NBHS result:

- 0–10
- 11–20
- 21–50
- >50

10. For infants identified through NBHS as having hearing loss, what is your best estimate of the earliest stage at which:

	At the time of screening	At the time of diagnosis	At the time of enrollment in early intervention	Other (please specify)
Parents/family need to be informed about genetic referrals	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Genetic referral should be made	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>

**Other Identification and Genetics**

The following questions are about children who were identified as having hearing loss via means OTHER THAN newborn hearing screening (NBHS).

11. Have you referred parents who have a child with hearing loss identified through means other than NBHS to genetics professionals? (If you answered “No” to this question, you will skip to Q15)

- Yes
- No

12. If/When you have a child with hearing loss identified through means other than NBHS, how often do you refer the parents to genetics professionals?

- Rarely
- Sometimes
- Often
- Unsure
- Not applicable

13. In the last year, approximately how many patients did you refer for genetic evaluation of hearing loss after identification through a means other than NBHS?

- 0–10
- 11–20
- 21–50
- >50

14. For patients identified through a means other than NBHS as having hearing loss what is your best estimate of the earliest stage at which:

	At the time of screening	At the time of diagnosis	At the time of enrollment in early intervention	Other (please specify)
Parents/family need to be informed about genetic referrals	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Genetic referral should be made	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>

**All Children with Hearing Loss and Genetics**

The following questions are about ALL children with hearing loss, regardless of how they were identified.

15. Which is the primary reason why you refer families to a genetics professional for hearing loss?

- Parents ask for a referral
- There is a family history of hearing loss
- Parents are discussing another pregnancy
- I have not referred to a genetics professional for hearing loss
- Other \_\_\_\_\_

16. How often do you consult with (i.e., do not make a formal referral) geneticists and genetic counselors regarding hearing loss?

- Never (1)
- Rarely (2)
- Sometimes (3)
- Often (4)
- Unsure (5)

17. What challenges have you experienced when referring parents/families for a genetic evaluation of the infant/child?

	Not a Challenge	Sometimes a Challenge	Always a Challenge	Not Applicable
Lack of information/resources to make referrals	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Lack of insurance	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Insurance limitations	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Lack of family support	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Transient families	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Parents/families may not consider genetic referral a priority	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Lack of local genetics provider	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Lack of telehealth options	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Other	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Other _____				

18. What reimbursement challenges have your patients encountered regarding hearing loss genetic referrals?

	Not a Challenge	Sometimes a Challenge	Always a Challenge	Not Applicable
Lack of insurance	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Medicaid constraints and/or cost limitations	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Preexisting or other regulations and policies	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Preauthorization	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Other	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Other _____				

19. For parents/families who already have a diagnosis of hearing loss, do you discuss with them the genetics of hearing loss?

- No
- Yes, sometimes
- Yes, always
- Not applicable

20. It is possible that infants with nonsyndromic hearing loss have indentifiable gene changes in Connexin 26 and/or 30 (GJB2/GJB6). At what age (in months) do you think connexin testing should be offered in a failed newborn hearing screen workup when there are not dysmorphic features/anomalies or a known genetic condition?

- Age (in months) \_\_\_\_\_
- Don't think connexin testing should be ordered
- Don't know/not familiar

21. Do you order connexin testing in infants with nonsyndromic hearing loss?

- Never
- Rarely
- Yes, sometimes
- Yes, almost always
- Not applicable

22. How often do you test for Cytomegalorvirus (CMV) when HL is identified on NBHS?

- Never
- Rarely
- Sometimes
- Always
- Unsure
- Not applicable

23. Do you have a designated system (i.e., person or computer system/database) for tracking...

	No	Yes	I don't know	Not Applicable
Failed newborn metabolic screening	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Failed NBHS	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Failed newborn blood spot screening	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Delayed developmental milestones	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Preauthorization for HL genetic testing	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Follow up after referrals	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>

24. Which specialist would you routinely refer the family of a child with confirmed hearing loss? Please select all that apply.

- Genetic specialist
- Audiologist
- ENT
- Other \_\_\_\_\_

25. How informed do you think you are about...

	Uninformed	Somewhat Informed	Very Informed
The genetics of HL	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
The importance of genetic referrals	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>

26. Do you have a designated system (i.e., person or computer system/database) for tracking...

	Uninformed	Somewhat Informed	Very Informed
Genetic causes of HL	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
The importance of genetic referrals	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
The logistics of genetic referral for HL	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
The significance of genetics in HL due to ototoxic medication exposure	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>

27. How important do you think it is to refer children with hearing loss to genetics professionals?

- Unimportant
- Somewhat important
- Very important

28. Prior to receiving this questionnaire were you familiar with the “EhDI 1-3-6” guidelines?

- No
- Somewhat
- Yes

29. Would you implement the following strategies to facilitate (or ensure) tracking of genetic referrals?

	Yes	No
Implement a system to follow up with patients and families	<input type="radio"/>	<input type="radio"/>
Engage case managers in the EhDI programs	<input type="radio"/>	<input type="radio"/>
Increase genetics education efforts	<input type="radio"/>	<input type="radio"/>
Implement a system to follow up with other providers	<input type="radio"/>	<input type="radio"/>
Other	<input type="radio"/>	<input type="radio"/>
Other _____		

30. Have you used the following strategies to facilitate (or ensure) tracking of genetics referrals?

	Yes	No
Implement a system to follow up with patients and families	<input type="radio"/>	<input type="radio"/>
Engage case managers in the EhDI programs	<input type="radio"/>	<input type="radio"/>
Increase genetics education efforts	<input type="radio"/>	<input type="radio"/>
Implement a system to follow up with other providers	<input type="radio"/>	<input type="radio"/>
Other	<input type="radio"/>	<input type="radio"/>
Other _____		

31. If you have used the following strategies to facilitate (or ensure) tracking of genetic referrals did it work?

	No	Somewhat	Yes	Have Not Used
Implement a system to follow up with patients and families	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Engage case managers in the EHDl programs	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Increase genetics education efforts	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Implement a system to follow up with other providers	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Other	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Other _____				

32. Would you implement the following strategies to enhance collaborations and communication with EHDl/EI programs and primary care providers regarding genetic referrals and follow-up?

	No	Yes
Genetics of HL Reference guide	<input type="radio"/>	<input type="radio"/>
Handout of HL genetic testing	<input type="radio"/>	<input type="radio"/>
List of available resources on HL genetics	<input type="radio"/>	<input type="radio"/>
State/district-specific contact information on EHDl programs and genetics centers	<input type="radio"/>	<input type="radio"/>
Other	<input type="radio"/>	<input type="radio"/>
Other _____		



33. Have you used the following strategies to enhance collaboration and communication with EHD/El programs and primary care providers regarding genetic referrals and follow-up?

	No	Yes
Genetics of HL Reference guide	<input type="radio"/>	<input type="radio"/>
Handout of HL genetic testing	<input type="radio"/>	<input type="radio"/>
List of available resources on HL genetics	<input type="radio"/>	<input type="radio"/>
State/district-specific contact information on EHD/El programs and genetics centers	<input type="radio"/>	<input type="radio"/>
Other	<input type="radio"/>	<input type="radio"/>
Other _____		

34. If you have used the following strategies to enhance collaboration and communication with EHD/El programs and primary care providers regarding genetic referrals and follow-up, did it work?

	No	Somewhat	Yes	Have Not Used
Genetics of HL reference guide	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Handout of HL genetics testing	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
List of available resources on HL genetics	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
State/district-specific contact information on EHD/El programs and genetics centers	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Other	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>	<input type="radio"/>
Other _____				

## The Past, Present, and Future of Universal Newborn Hearing Screening in Taiwan

Hung-Ching Lin, MD, MSc<sup>1,2,3</sup>  
 Hsiu-Wen Chang, PhD<sup>1</sup>  
 Wen-Hui Hsieh, BS<sup>1</sup>

<sup>1</sup>Department of Audiology & Speech Language Pathology, Mackay Medical College, New Taipei City, Taiwan

<sup>2</sup>Department of Otolaryngology, Mackay Memorial Hospital, Taipei, Taiwan

<sup>3</sup>Department of Medicine, Mackay Medical College, New Taipei City, Taiwan

**Abstract:** The Taipei Mackay Memorial Hospital in Taiwan, in collaboration with the Children’s Hearing Foundation, initiated a free newborn hearing screening program using transient evoked otoacoustic emissions (TEOAE) in 1998. During 2003-2013, there were four major phases of implementation and promotion of Taiwan’s universal newborn hearing screening program (UNHS) initiated by the government’s Bureau of Health Promotion. These included (a) establishment of the 2004 Taiwan Guidance of Newborn Hearing Screening Program, (b) completion of the 2008 Taiwan Consensus Statement on Newborn Hearing Screening, (c) implementation of a national government-funded UNHS program in 2012, and (d) the 2014 Taiwan UNHS Revised Guidelines. In summary, in 2016, Taiwan national statistics indicated that for a total of nearly 200,000 babies the coverage rate of newborn hearing screening was up to 98.2%, the refer rate was 1.13%, the return rate of ordinary patient department (OPD) referral was 86.10%, and the incidence of congenital deafness was estimated to be 0.455%.

**Key Words:** universal newborn hearing screening (UNHS), aABR, congenital deafness

**Acronyms:** aABR = automated auditory brainstem response; AAP = American Academy of Pediatrics; JCIH = Joint Committee on Infant Hearing; NIH = National Institutes of Health; OAE = otoacoustic emissions; OPD = ordinary patient department; TEOAE = Transient evoked otoacoustic emissions; UNHS = universal newborn hearing screening

**Acknowledgements:** The authors acknowledge Health Promotion Administration, Ministry of Health and Welfare, Taiwan. This paper’s results were supported by many studies from a National Research Project (grants DOH96-HP-1312, DOH99-HP-1201, C1031229) commissioned by the Health Promotion Administration, Minister of Health and Welfare, Taiwan.

**Correspondence concerning this article should be addressed to:** Hung-Ching Lin, MD, MSc, Department of Audiology and Speech Language Pathology, Mackay Medical College, New Taipei City, Taiwan, No. 46, Sec. 3, Zhongzheng Rd., Sanzhi Dist., New Taipei City 252, Taiwan. Phone: +886 2 25433535/2209; Fax: +886 2 25433642; Email: hclin59@ms29.hinet.net

### Introduction

In March 1993, the U.S. National Institutes of Health (NIH) concluded, “that all infants should be screened for hearing impairment... This will be accomplished most efficiently by screening prior to discharge (from the birth hospital). Infants who fail hearing screening should have a comprehensive diagnostic hearing evaluation no later than 6 months of age” (p. 215-227). Subsequently, the Joint Committee on Infant Hearing (JCIH, 2007) and the American Academy of Pediatrics (AAP, 2008) both recommended that newborn hearing screening should be conducted by the first month of life, confirmation of congenital hearing loss should be made by three months of age, and intervention and rehabilitation plans should be in place by six months of age. The fact that early diagnosis and intervention of hearing loss has resulted in significant

impacts on the development of children with hearing impairment has been well studied and documented in the United States since 1998 (Downs & Yoshinaga-Itano, 1999; Yoshinaga-Itano, Sedey, Coulter, & Mehl, 1998).

Based on this, the Taipei Mackay Memorial Hospital in Taiwan, in collaboration with the Children’s Hearing Foundation, initiated a free newborn hearing screening program using transient evoked otoacoustic emissions (TEOAE) in 1998 (Lin, Shu, Chang, & Bruna, 2002). This was followed in 2002 by a call from the Taiwan Ministry of Health and Welfare for research proposals related to the administration of screening procedures and policies as a way of improving the welfare of infant hearing health. As a result, Taipei Mackay Memorial Hospital began to take part in these projects. During 2003–2013, there were four major phases of implementation and promotion of the

universal newborn hearing screening program (UNHS) in Taiwan initiated by the government's Bureau of Health Promotion. First, in 2003, Chang Gung Medical Hospital was commissioned by the Taiwan Ministry of Health and Welfare to evaluate the feasibility of implementing a newborn hearing screening program in Taiwan. The purpose of the project included understanding the feasibility of conducting newborn hearing screening in maternity hospitals and planning the screening procedures during 2003 and 2004. Since 2004, Dr. Hung-Ching Lin of Taipei Mackay Memorial Hospital has played a major role in these 4 major Taiwan UNHS promotion projects, including (a) establishment of the 2004 Taiwan Guidance of Newborn Hearing Screening Program (Bureau of Health Promotion, 2004), (b) completion of the 2008 Taiwan Consensus Statement on Newborn Hearing Screening (Bureau of Health Promotion, 2008), (c) implementation of a national government-funded UNHS program in 2012 (Yoshinaga-Itano et al., 1998), and the 2014 Taiwan UNHS Revised Guidelines (Bureau of Health Promotion, 2014).

In addition, in order to raise public awareness of the importance of UNHS, the 2004 Taiwan Guidance of Newborn Hearing Screening Program (using otoacoustic emissions [OAE] or automated auditory brainstem response [aABR] at parent's own expense) was completed with support from many speech and hearing professionals (Bureau of Health Promotion, 2004). In the same year, the Taiwan Speech-Language-Hearing Association implemented several training courses in the basic concepts of UNHS for speech and hearing professionals.

Nevertheless, by 2007, Taiwan national statistics showed that only 28.7% of medical institutions offered newborn hearing screening services in Taiwan (Bureau of Health Promotion, 2007–2008). Consequently, a national research project commissioned by the Ministry of Health and Welfare was conducted by Taipei Mackay Memorial Hospital with the goal of promoting and studying the effectiveness of UNHS in an effort to increase the coverage rate of newborn hearing screening in Taiwan. A promotion center in Taipei and four hearing screening service centers located in the northern, central, southern, and eastern parts of Taiwan were established to recruit more medical institutions to conduct newborn hearing screening. By 2008, the percentages of medical institutions offering newborn hearing screening service had increased to 39.4% (Bureau of Health Promotion, 2014). Meanwhile, the Taiwan Consensus Statement on Newborn Hearing Screening, 2008 (using aABR at parent's own expense) was completed and endorsed by a panel of international experts from Singapore and Hong Kong (Bureau of Health Promotion, 2008). The four service centers, in collaboration with other associated organizations, began to hold seminars and workshops to facilitate the communication between hearing screening personnel and speech and hearing professionals, and also to improve their clinical skills and knowledge in the area of newborn hearing screening.

Through these efforts, 87% of medical institutions in Taiwan implemented newborn hearing screening programs by 2011–2012 (Bureau of Health Promotion, 2011–2012). At that time, some counties and cities had been implementing newborn hearing screening using either OAE or automated auditory brainstem response aABR; however research by the authors indicated that aABR may be more beneficial and more practical than TEOAE (Lin, Shu, Lee, Lin, & Lin, 2007). Consequently, the Taiwan Ministry of Health and Welfare issued regulations in February 2012 that provided government funding for universal newborn hearing screening using aABR, free of charge to parents, with the policy initiated on March 15, 2012. At the same time, the maternal and child health data management and tracking system to monitor the hearing status of newborns was activated.

After implementation of universal newborn screening in Taiwan in 2012, a monitoring center was established to work together with the four promotion centers across the island to offer consultation and quality control of hearing screening and diagnosis. By 2013, the coverage rate of Taiwan's newborn hearing screening program reached 97.3% (Bureau of Health Promotion, 2011–2012); comparable with the status in the United Kingdom, United States, and Australia. In addition, the 2013 confirmation rate of hearing diagnosis in Taiwan was 81.6%, higher than the 54.1% in the United States in 2012 (Muse et al., 2013). A revised consensus conference for newborn hearing screening, diagnosis, and intervention held and confirmed by U.S. professor Christine Yoshinaga-Itano and Australian professor Joseph Kei in 2014 sought to improve the distribution of medical resources and the referring procedures of early intervention with the goal of improving quality and outcomes of newborn hearing screening programs in Taiwan (Bureau of Health Promotion, 2014).

In summary, the 2016 Taiwan national statistics indicate that for a total of nearly 200,000 babies, the coverage rate of newborn hearing screening was up to 98.2%, the refer rate was 1.13%, the return rate of OPD referral was 86.10%, and the incidence of congenital deafness was estimated to be 0.445% (Bureau of Health Promotion, 2015–2016). This shows that the overall outcome of the Taiwan newborn hearing screening program has approached international performance levels (Huang et al., 2014). In the future, we will be devoted to leading and sharing our experience in establishing infant hearing screening programs to neighboring countries. From our experience in promotion of UNHS in Taiwan, there are five main points we would suggest for others creating a UNHS program.

1. Establish a national guidance and consensus statement for newborn hearing screening in order to bring professional experts together for the work.
2. Publish domestic UNHS related results in international papers to raise your government's attention of its importance and to gain government funding.

3. In order to increase the coverage rate, the UNHS program must be supported with national funding so that parents do not pay for screening. In Taiwan the coverage rate was initially only 70% when parents paid, later increasing to 98.3% with national free UNHS.

4. Monitor UNHS quality to confirm higher coverage and diagnosis rates through projects such as Taiwan's quality control improvement project for newborn hearing screening and confirmed diagnosis (Bureau of Health Promotion, 2015–2016).

5. Create a national UNHS data tracking system. In Taiwan, the national UNHS data tracking system was monitored by Health Promotion Administration, Ministry of Health and Welfare, Taiwan. Our Mackay Memorial Hospital team was commissioned by the government to help audit and promote its qualities, to reduce inappropriately higher referral rates, and to increase diagnostic follow up rates and early intervention rates via this data tracking system.

hearing. *Pediatrics*, 131(4), e1324–1349. doi:10.1542/peds.2013-0008

National Institutes of Health Consensus Development Conference Statement. (1993). Early identification of hearing impairment in infants and young children. *International Journal of Pediatric Otorhinolaryngology*, 27(3), 215–227.

U.S. Preventive Services Task Force. (2008). Recommendation statement: Universal screening for hearing loss in newborns. *Pediatrics*, 122(1), 143–148. doi:10.1542/peds.2007-2210

Yoshinaga-Itano, C., Sedey, A. L., Coulter, D. K., & Mehl, A. L. (1998). Language of early- and later-identified children with hearing loss. *Pediatrics*, 102(5), 1161–1171.

## References

- Bureau of Health Promotion, Department of Health, Taiwan, Republic of China (ROC). (2004). *Guidelines of newborn hearing screening*. Retrieved from <https://drive.google.com/file/d/0Bxxznrg0Qv68MGRySXpQNzR1Vzg/view>
- Bureau of Health Promotion, Department of Health, Taiwan, ROC. (2008). *Manual of consensus of newborn hearing screening*. Retrieved from <https://drive.google.com/file/d/0Bxxznrg0Qv68Y080WEI3SFipMjg/view>
- Bureau of Health Promotion, Department of Health, Taiwan, ROC. (2014). *Manual of newborn hearing screening and diagnosis*. Retrieved from <https://drive.google.com/file/d/0Bxxznrg0Qv68XzJpWDdMTW5mSmc/view>
- Bureau of Health Promotion, Department of Health, Taiwan, ROC. (2007–2008). *Promotion of newborn hearing screening and its effectiveness evaluation 2007–2008* (Project Grant DOH96-HP-1312). Taipei City, Taiwan, ROC: Mackey Memorial Hospital.
- Bureau of Health Promotion, Department of Health, Taiwan, ROC. (2010–2012). *Service network of newborn hearing screening and evaluation of its efficacy, 2010–2012* (Project Grant DOH99-HP-1301). Taipei City, Taiwan, ROC: Mackey Memorial Hospital.
- Bureau of Health Promotion, Department of Health, Taiwan, ROC. (2015–2016). *Quality control improvement project for newborn hearing screening and confirmed diagnosis 2015–2016* (Project Grant C1031229). Taipei City, Taiwan, ROC: Mackey Memorial Hospital.
- Centers for Disease Control and Prevention. (2007). Retrieved from <https://www.cdc.gov/ncbddd/hearingloss/recommendations.html>
- Downs, M. P., & Yoshinaga-Itano, C. (1999). The efficacy of early identification and intervention for children with hearing impairment. *Pediatric Clinics of North America*, 46(1), 79–87.
- Huang, C. M., Yang, I. Y., Ma, Y. C., Lin, G. S., Yang, C. C., Tsai, H. T., & Lin, H. C. (2014). The effectiveness of the promotion of newborn hearing screening in Taiwan. *International Journal of Pediatric Otorhinolaryngology*, 78(1), 14–18. doi:10.1016/j.ijporl.2013.10.005
- Lin, H. C., Shu, M. T., Chang, K. C., & Bruna, S. M. (2002). A universal newborn hearing screening program in Taiwan. *International Journal of Pediatric Otorhinolaryngology*, 63(3), 209–218.
- Lin, H. C., Shu, M. T., Lee, K. S., Lin, H. Y., & Lin, G. (2007). Reducing false positives in newborn hearing screening program: How and why. *Otology & Neurotology*, 28(6), 788–792.
- Muse, C., Harrison, J., Yoshinaga-Itano, C., Grimes, A., Brookhouser, P. E., Epstein, S., . . . Martin, B. (2013). Supplement to the JCIH 2007 position statement: Principles and guidelines for early intervention after confirmation that a child is deaf or hard of

## Restructuring Data Reported from Jurisdictional Early Hearing Detection and Intervention (EHDI) Programs: A Pilot Study

Suhana Alam, MPH<sup>1</sup>  
Winnie Chung, AuD<sup>1</sup>  
Xidong Deng, PhD<sup>1</sup>  
Tammy O'Holearn, LBSW<sup>2</sup>  
Jim Beavers, BGS<sup>3</sup>  
Rebekah F. Cunningham, PhD<sup>4</sup>  
Alyssa K. Rex, AuD<sup>4</sup>  
ThuyQuynh N. Do, PhD, MPH<sup>1</sup>

<sup>1</sup>National Center on Birth Defects and Developmental Disabilities, Centers for Disease Control and Prevention, Atlanta, GA

<sup>2</sup>Iowa Department of Public Health, Des Moines, Iowa

<sup>3</sup>Nebraska Department of Health and Human Services, Lincoln, Nebraska

<sup>4</sup>Indiana State Department of Health, Indianapolis, Indiana

**Abstract: Objective.** To assess the feasibility, benefits, and challenges surrounding individual-level versus aggregate data reporting by jurisdictional EHDI programs to the Centers for Disease Control and Prevention (CDC).

**Method.** Using data reported to CDC by three jurisdictions in 2011, descriptive statistics were used to assess the feasibility of collecting and reporting individual-level data. Comparisons were made on what can be learned from individual-level data as opposed to CDC's aggregate survey data.

**Results.** Individual-level data provided a detailed overview of the population served, services received, and variations across jurisdictions in data collection, reporting, and quality monitoring practices. Several challenges and areas needing improvement were identified: variations in (1) data standardization; (2) data collection and reporting procedures; and (3) protocols for recommended follow-up services.

**Conclusions.** Using individual-level data, CDC was able to perform in-depth statistical analyses and learn more about each jurisdiction's population, their EHDI process, and challenges to data collection, tracking, and surveillance efforts. As a result, CDC was able to provide more targeted technical assistance. All of the above would not be feasible using aggregate survey data. The pilot study demonstrated that individual-level data reporting to CDC is feasible and offers many opportunities for both CDC and jurisdictional EHDI programs.

**Key Words:** Early hearing detection and intervention, newborn hearing screen, surveillance and tracking

**Acronyms:** CDC = Centers for Disease Control and Prevention; EHDI = Early Hearing Detection and Intervention; EI = early intervention; HL = hearing loss; HSFS = Hearing Screening and Follow-up Survey; iEHDI = individual EHDI; NBHS = Newborn hearing screening; NQF = National Quality Forum; SDN = Secure Data Network

**Acknowledgements:** We would like to extend special thanks to the Indiana, Iowa, and Nebraska EHDI programs for participating in the iEHDI pilot study. This study would not be feasible without their involvement and support.

**Correspondence concerning this article should be addressed to:** Suhana Alam, MPH, National Center on Birth Defects and Developmental Disabilities, Centers for Disease Control and Prevention, 4770 Buford Highway, MS-E88, Atlanta, GA 30341. Phone: 404-498-0984; Fax: 770-488-0270; Email: [SAlam1@cdc.gov](mailto:SAlam1@cdc.gov)

### Introduction

Newborn hearing screening (NBHS) is one of the 31 primary conditions included on the Recommended Universal Screening Panel (Health Research & Services Administration, 2017). However, NBHS alone does not

ensure that a child with hearing loss (HL) is identified (Winston-Gerson & Hoffman, 2017). Early diagnosis of HL involves a series of steps and services through multiple providers. If an infant does not pass NBHS, it is crucial to determine if the infant received appropriate and timely follow-up diagnostic services. If HL is present,



the infant needs to receive recommended intervention services as early as possible to reduce the likelihood for developmental delays (Vohr, 2003). Most U.S. states and territories have an EHDI program with goals to screen infants for HL no later than 1 month of age, diagnose HL no later than 3 months of age for infants who did not pass the hearing screening, and enroll infants identified with permanent HL into early intervention (EI) no later than 6 months of age. EHDI programs accomplish these 1-3-6 goals through active tracking, surveillance, and coordination with clinical service providers and families (Williams, Alam, & Gaffney, 2015).

CDC supports jurisdictional EHDI programs by providing programs with funding and assistance to develop, maintain, and enhance the collection of hearing screening, diagnosis, and EI data. Through the Hearing Screening and Follow-up Survey (HSFS), CDC collects aggregate data based on individually identifiable records from jurisdictional EHDI programs about NBHS, which allows for evaluation of the timeliness of receipt of hearing screening, diagnosis, and enrollment in EI services. This survey tool helps assess and monitor EHDI progress nationally and assists states and territories in strengthening their programs by identifying data gaps and areas of need (Alam, Gaffney, & Eichwald, 2014; CDC, 2017). The voluntary survey is sent annually to each EHDI program.

Although the HSFS allows CDC to generate national reports about the number of infants screened, diagnosed, and enrolled in EI and to assess progress toward the 1-3-6 goals, several limitations and questions cannot be addressed by the survey's data. The use of aggregate data can lead to an ecological fallacy where inferences are incorrectly generalized to the whole jurisdictional population (i.e., using aggregate data to infer individual-level relationships; King, 2013; Stewart & Tierney, 2002). Detailed data quality checks are not possible using aggregate data. Although CDC provides definitions for each HSFS data item, some respondents may quantify and aggregate their data differently when they participate in the survey (Alam, Satterfield, Mason, & Deng, 2016). Improving data standardization is not possible without seeing individual-level data. It is difficult to provide a descriptive summary of the individual services when data are aggregated.

Aggregate data do not allow for in-depth analyses of infant and family sociodemographic characteristics and the receipt of EHDI-related services. Aggregate data do not allow for answering key questions, such as the average age when an infant is diagnosed with HL. As a result, it is often not possible for CDC to use HSFS to identify potential program gaps and needs that would help provide more targeted technical assistance. To address these limitations, CDC implemented a pilot study in September 2010 known as individual EHDI (iEHDI), in which the participating jurisdictions assembled and transmitted limited sets of de-identified, individual-level data to CDC. The objective of this article is to describe the feasibility,

benefits, and challenges surrounding the reporting and use of individual-level data compared to HSFS data for EHDI.

## Method

### iEHDI Pilot Study

To participate, jurisdictions were required to have a comprehensive EHDI tracking and surveillance system in place and to routinely collect and maintain non-aggregated, individual-level data on all infants born in the jurisdiction, as well as the hearing screening and follow-up services they received. Three jurisdictions—Indiana, Iowa, and Nebraska—were selected and awarded funds to provide de-identified sets of specified data items to CDC for infants born in 2010. Two jurisdictions (Iowa and Nebraska) voluntarily provided these data to CDC for infants born in 2012.

Quarterly data sets were transmitted to CDC via a Secure Data Network (SDN). Jurisdictional participants and CDC jointly reviewed and finalized the list and format of data items to be transmitted. The list was based on items included in the HSFS and additional information already collected by the jurisdictional programs. Prior to transmission, participants were required to perform a data validation and verification check to identify and correct data format and logic errors. Format errors refer to errors in the type, value, or range of a single data item (e.g., an infant's residence zip code coded in character string instead of numeric format). Logic errors occur when an illogical relationship is discovered when the data item is validated with another data item. For example, crosschecking the infant's date of birth shows that the NBHS occurred before birth.

To maintain the data integrity and privacy, jurisdictions assigned each infant record a new identifier consisting of a 2-digit jurisdictional ID followed by a 13-digit record ID. The 13-digit record ID could not contain any direct personal identifiers or information that may indirectly identify the infant. The infant's pseudonym was used to link records across the study period. Participating jurisdictions transmitted the data through an SDN operated by CDC Public Health Informatics and Technology Program Office. The data were stored in a stand-alone Microsoft Access database maintained by CDC Information Technology Services Office. Access was restricted to approved CDC EHDI program staff who had signed a data user agreement. CDC EHDI program staff performed an additional data review, validation, and verification check. All identified data errors were listed in a data quality report and shared with the jurisdictions to correct before re-transmission.

When the datasets were in acceptable format and clear of obvious format or logic errors (e.g., an infant's date of hearing screening occurring before the infant's date of birth), in-depth statistical analyses were conducted to demonstrate the value of having individual-level data



as opposed to HSFS (aggregate) data. A summary of 2010 and 2012 EHDI tracking and surveillance efforts was assembled. This information included the number of newborns not passing the final hearing screening, the status and results of diagnostic evaluation, the EI enrollment status, and infant and maternal characteristics for those diagnosed with permanent HL.

### **Demographic, Clinical, and Sociodemographic Variables**

Descriptive variables collected from vital records were reported for the infant and parents, such as infant gender (male/female), marital status (married: yes/no). Maternal Age was calculated as the difference in years between the mother's date of birth and the child's date of birth, and categorized as  $\leq 19$  years, 20-34 years,  $\geq 35$  years. Ethnicity for mother and father were each categorized as Hispanic (Mexican/Mexican American/Chicana, Puerto Rican, Cuban, unspecified Hispanic, or other Spanish/Hispanic/Latina) or Non-Hispanic. Maternal and Paternal Race were each categorized as White, Black or African American, or Other.

Infant clinical measures from birth certificates from vital records, including birth weight, low Appearance, Pulse, Grimace response, Activity, and Respiration (APGAR) score (score  $< 6$  at 5 minutes: Yes/No), neonatal intensive care  $> 5$  days (Yes/No), number of prenatal visits, and family history of permanent HL (Yes/No) were reported. Birth weight was categorized as Low ( $< 2,500$  grams), Normal (2,500–4,000 grams), and High ( $\geq 4,001$  grams). A low APGAR score is a potential risk factor that can be used for identifying HL in infants (Biswas, Goswami, Baruah, & Tripathy, 2012; Lin & Oghalai, 2011).

Socioeconomic variables included maternal education, principal source of payment, and receipt of women, infants and children (WIC) food & nutrition services (Yes/No). Maternal Education was categorized as Less than High School or Unknown (8th grade or less, 9th to 12th grade without a diploma, or unknown), Completed High School or General Education Development (GED), Some College or Associate's Degree, and Bachelor's Degree and Above (i.e., Bachelor's, Master's, Doctorate or professional degree). Principal source of payment included Private Insurance, Medicaid, and Other.

### **EHDI Screening, Diagnostic, and EI Variables**

Tracking and surveillance variables included: screening methods, results of initial hearing screen, rescreen results, dates and results of diagnostic evaluation, and EI enrollment status. Table 1 provides a detailed summary of the EHDI screening, diagnostic, and EI variables. Permanent HL was described by laterality (bilateral/unilateral), type of HL (Sensorineural, Permanent Conductive, Mixed, Auditory Neuropathy, Unknown Type), and severity (degree of HL: Mild (26-40 decibels, dB), Moderate (41-55 dB), Moderately Severe (56-70 dB), Severe (71-90 dB), Profound (91+ dB), and Unknown or Missing) for each ear (American Speech-Language-Hearing, 2017a, 2017b).

### **Statistical Analysis**

Descriptive statistics (frequency counts and percentages) were used to analyze infant and parental sociodemographic characteristics of the newborn hearing screening population and of the infants with permanent HL, and key indicators for EHDI tracking and surveillance efforts for infants born in 2010 and 2012. Median age and standard deviation were calculated for maternal age (years) and infant age at first diagnostic evaluation (days). All analyses were performed using SAS Version 9.3 (SAS Institute Inc., Cary, NC) and validated by two of the authors.

### **Results**

Compared to HSFS data, it was feasible to receive more data items through the iEHDI pilot. Additional infant and family information not currently collected by the HSFS, such as maternal and paternal sociodemographic variables, infant birth characteristics, and risk factors for HL, were available through the pilot study. Table 2 compares the data items collected by the HSFS and iEHDI. With an increase in the range and depth of individual-level data, a comparison of individual infant characteristics at each benchmark was feasible (e.g., maternal characteristics of infants screened or diagnosed with HL).

Table 3 provides a summary of the infant and parental characteristics of each jurisdiction's infant population by year. Compared to HSFS data, Table 3 provides a more comprehensive description of the infant population in each jurisdiction and examples of the iEHDI information collected (e.g., birth weight of infant, family history of permanent childhood HL, and low APGAR score). As reflected in Table 3, birth cohort size varied across the three jurisdictions, however the infants had similar characteristics. There were more male than female births and the average birth weight was in the normal range. Across all three jurisdictions, more mothers were aged between 20-34 years, White, non-Hispanic, and had private insurance. Approximately 40% of the mothers received WIC food and nutrition services. Maternal education level varied by jurisdiction and birth year. A higher percentage of the fathers were White and non-Hispanic.

Table 4 provides summary statistics of key EHDI tracking and surveillance efforts by jurisdiction and birth year. The results of hearing screen, diagnostic evaluation for those not passing the screen, and the status of EI enrollment for those diagnosed with permanent HL revealed variations across the jurisdictions by cohort size and screening method. For example, Indiana had the largest birth cohort ( $n = 84,866$ ) and the lowest rate of not passing the final hearing screen (3.0%) in 2010. The percentage of infants diagnosed with permanent HL varied across jurisdictions in 2010. Of those infants documented with permanent HL, 23.2% of Indiana and 28.2% of Iowa infants were not documented as receiving EI services in 2010. EI data were unavailable from Nebraska (Table 4).

**Table 1****Summary of the Definitions Used for Early Hearing Detection and Intervention Screening, Diagnostic, and Early Intervention Variables.**

<b>Variable</b>	<b>Definition</b>
<b>Initial Hearing Screen Results</b>	
Pass	Infants who passed the initial hearing screening
Not Pass	Infants who did not pass the initial hearing screening
Not Screened	Infants who did not receive the initial hearing screening
Unknown Status	Infants whose initial hearing screening status was unknown
<b>Infants Receiving Rescreen</b>	
Pass	Infants who did not pass the initial hearing screen and passed the rescreen
Not Pass	Infants who did not pass the initial hearing screen and did not pass the rescreen
Not Screened	Infants who did not pass the initial hearing screen and did not receive the rescreen
<b>Final Hearing Screen Results</b>	
Pass	Infants who (1) Passed the initial screen and rescreen (2) Passed the initial screen and did not receive a rescreen; (3) Passed the initial screen and were not documented to have received a rescreen; (4) Did not pass the initial screen and passed the rescreen; or (5) Did not receive the initial screen but passed the rescreen
Not Pass	Infants who (1) Passed the initial screen but not the rescreen; (2) Did not pass both the initial and the rescreen; (3) Did not pass the initial and did not receive a rescreen; (4) Did not pass the initial and not documented as having received a rescreen; or (5) Did not receive the initial screen and did not pass the rescreen
Not Screened	Infants who were not screened or not documented as having received any hearing screening
<b>Diagnostic Evaluation Status</b>	
Diagnosed	Infants who did not pass the hearing screen as the final result and received a diagnostic evaluation, regardless of the diagnostic result being conclusive or inconclusive
Not Diagnosed	Infants who did not pass the hearing screen as the final result and did not receive a diagnostic evaluation
Died or Moved	Infants who did not pass the hearing screen as the final result and did not receive a diagnostic evaluation because the infant expired or the family moved
Refused	Infants who did not pass the hearing screen as the final result and did not receive a diagnostic evaluation because the family has refused the evaluation
<b>Diagnostic Results</b>	
No Hearing Loss	Infants diagnosed as having no hearing loss
Permanent Hearing Loss	Infants diagnosed with permanent hearing loss
Non-Permanent Hearing Loss	Infants diagnosed with non-permanent hearing loss
In Process	Infants who did not pass the hearing screening as the final result and are still in the evaluation process for a conclusive diagnosis
Unknown Status	Infants who did not pass the hearing screening as the final result and received a diagnostic evaluation but the diagnosis was not known to the EHDI program
<b>Status of EI Service for those Diagnosed with Permanent Hearing Loss</b>	
Receiving EI	Infants who did not pass the hearing screen as the final result, diagnosed as having a permanent hearing loss, and enrolled into early intervention services
Not Receiving EI or Unknown	Infants who did not pass the hearing screen as the final result, diagnosed as having a permanent hearing loss, and not enrolled into EI services or the enrollment status is unknown to the EHDI program
Died or Moved	Infants who did not pass the hearing screen as the final result, diagnosed with a permanent hearing loss, and not enrolled into EI services because the infant expired or the family moved
Refused	Infants who did not pass the hearing screen as the final result, diagnosed with a permanent hearing loss, and not enrolled into EI services due to family refusal

**Table 2*****Comparison of Data Collected Between Hearing Screening and Follow-up Survey (HSFS) and Individual Early Hearing Detection and Intervention (iEHDI)***

<b>Data Item</b>	<b>HSFS</b>	<b>iEHDI</b>
Individual-level infant birth information (e.g., birth date, birth county, time, birth weight, birth order, birthing facility, transfer)		x
Gender of infant	x	x
Birth jurisdiction	x	x
Maternal race/ethnicity	x	x
Maternal education	x	x
Maternal residence (jurisdiction and/or zip code)		x
Maternal age	x	x
Marital status of mother		x
Plurality		x
Prenatal visits		x
WIC enrollment status		x
Paternal race/ethnicity		x
Paternal education		x
Principle source of payment		x
Number of hearing screening received		x
Initial hearing screening information		x
Initial screening results for each ear		x
Final initial hearing screening result		x
Reason for no initial screen		x
Rescreen result for each ear		x
Follow-up screening information (date and location)		x
Reason for no rescreen		x
Date and location of initial diagnostic visit		x
Date and location of most recent diagnostic visit		x
Result of most recent diagnostic visit		x
Diagnosis date	x	x
Number of documented visits during diagnostic process		x
Laterality	x	x
Severity		x
Reason for no intervention	x	x
Referred to receive other services (i.e., genetic counseling, ophthalmology, otolaryngology, other provider)		x
Early intervention enrollment information	x	x
Part C eligibility	x	x
Hearing aid, implant, or FM system		x
Risk factors and comorbidities (e.g., family history of hearing loss, birth defects, medical conditions)		x

Furthermore, iEHDI allows for comparing trends of key tracking and surveillance indicators within a jurisdiction (Table 4). Between 2010 and 2012, the percentage of infants who did not pass the final hearing screen decreased from 1.7% to 1.2% for Iowa, and from 1.0% to 0.4% for Nebraska. This may be a direct result of an increase in the percentage of infants passing the initial hearing screen. The decrease in the percentage of infants who did not pass the final hearing screening subsequently yielded a smaller cohort of infants in need of a diagnostic evaluation in 2012. Between 2010 and 2012, infants in Iowa who were not documented as receiving a diagnostic evaluation decreased from 56.7% to 44.4%. Likewise, a decrease from 28.2% to 17.0% was also seen for infants in Iowa who were not documented as receiving EI. For Nebraska, there was a decrease from 46.4% to 37.5% for infants who were not documented as receiving a diagnostic evaluation. EI enrollment data were not available for Nebraska in 2010. Altogether, Table 4 shows that it is feasible to track each infant's EHDl process and to perform subset analyses (e.g., assess EI enrollment

status among infants diagnosed with permanent HL, using individual-level data). In addition, individual-level data allow for detailed understanding of each jurisdiction's EHDl process, which was otherwise not possible using HSFS data.

As shown in Table 4, it was feasible to calculate the median age of infants who did not pass the hearing screen and received a diagnostic evaluation. The median age varied across years for each jurisdiction. Between 2010 and 2012, the median age when infants received a diagnostic evaluation decreased for Iowa (74 days vs. 48 days) and increased for Nebraska (49 days vs. 65 days). For Indiana, the median age was younger (48 days) in 2010. Currently, the HSFS does not gather this information.

Table 5 shows the summary of infant and maternal characteristics for infants who were a diagnosed with permanent HL in 2010 and 2012. Across all jurisdictions, regardless of the birth cohort size, 2.0 per 1,000 live born

Table 3

Summary of Infant and Parental Characteristics of the Newborn Hearing Screening Population, iEHDI 2010 and 2012.

Characteristic	Indiana <sup>a</sup>	Iowa		Nebraska	
	2010 n (%)	2010 n (%)	2012 n (%)	2010 n (%)	2012 n (%)
<b>Total Number of Births</b>	<b>84,866</b>	<b>38,572</b>	<b>38,427</b>	<b>26,247</b>	<b>26,284</b>
<b>Gender of Infant</b>					
Male	43,759 (51.6)	19,646 (50.9)	19,701 (51.3)	13,333 (50.8)	13,500 (51.4)
Female	41,106 (48.4)	18,926 (49.1)	18,726 (48.7)	12,914 (49.2)	12,777 (48.6)
<b>Birth weight categories(g)</b>					
Average infant birth weight ± SD	3,273.4 ± 592.0	3,345.6 ± 581.7	3,355.7 ± 577.0		
Low (< 2500)	6,749 (8.0)	2,535 (6.6)	2,429 (6.3)		
Normal (2500–4000)	71,469 (84.2)	32,141 (83.3)	32,022 (83.3)		
High (≥ 4001)	6,569 (7.7)	3,879 (10.7)	3,965 (10.3)		
Unknown	79 (0.1)	17 (0.0)	11 (0.0)	N/A <sup>d</sup>	
<b>Family History of Permanent Childhood Hearing Loss</b>	613 (0.7)	948 (2.5)	766 (2.0)		
<b>Neonatal intensive care &gt; 5 days</b>	199 (0.2)	2,126 (5.5)	1,915 (5.0)		
<b>Low APGAR score(&lt;6) at 5 minutes</b>	1,464 (1.7)	306 (0.8)	317 (0.8)		
<b>Average number of prenatal visits</b>	10.7 ± 3.7	11.8 ± 3.2	11.9 ± 3.2		
<b>Maternal Age (years)</b>					
Median Age ± SD	27.0 ± 5.9	27.7 ± 5.6	28.1 ± 5.5	28.0 ± 5.0	28.4 ± 5.5
≤ 19	8,716 (10.3)	3,139 (8.1)	2,567 (6.7)	1,943 (7.4)	1,678 (6.4)
20–34	66,994 (78.9)	31,334 (81.2)	31,629 (82.3)	21,305 (81.2)	21,435 (81.6)
≥ 35	9,115 (10.7)	4,097 (12.1)	4,230 (11.0)	2,999 (11.4)	3,171 (12.1)
<b>Maternal Ethnicity</b>					
Hispanic	6,739 (7.9)	3,309 (8.6)	3,373 (8.8)	3,776 (14.4)	3,648 (13.9)
Non-Hispanic	78,127 (92.1)	35,263 (91.4)	35,054 (91.2)	22,471 (85.6)	22,636 (86.1)
<b>Maternal Race</b>					
White	69,725 (82.2)	33,301 (86.3)	33,020 (85.9)	21,106 (80.4)	21,059 (80.1)
Black or African American	9,553 (11.3)	1,743 (4.5)	1,911 (5.0)	1,561 (6.0)	1,579 (6.0)
Other race <sup>b</sup>	5,588 (6.6)	3,528 (9.1)	3,496 (9.1)	3,580 (13.6)	3,646 (16.4)
<b>Maternal Education</b>					
< High School or Unknown	17,651 (20.8)	5,271 (13.7)	4,890 (12.7)	3,763 (14.3)	3,308 (12.6)
Completed High School or GED	22,460 (26.5)	7,780 (20.2)	7,446 (19.4)	4,933 (18.8)	4,812 (18.3)
Some College or Associate's Degree	24,038 (28.3)	8,373 (21.7)	13,549 (35.3)	9,048 (34.5)	6,336 (24.1)
≥ Bachelor's Degree	20,717 (24.4)	11,987 (30.8)	12,542 (32.6)	8,503 (32.4)	11,828 (45.0)
<b>Marital Status</b>					
Married	48,720 (57.4)	25,154 (65.2)	24,875 (64.7)	17,558 (66.9)	17,664 (67.2)
Not Married/Unknown/Missing	36,146 (42.6)	13,418 (34.8)	13,552 (35.3)	8,689 (33.1)	8,620 (32.8)
<b>Receipt of Women, Infants and Children (WIC) Food &amp; Nutrition Services</b>	39,746 (46.8)	14,879 (38.6)	14,380 (37.4)		
<b>Paternal Ethnicity</b>					
Hispanic	36,629 (43.2)	8,726 (22.6)	8,807 (22.9)		
Non-Hispanic	48,237 (56.8)	29,846 (77.4)	29,620 (77.1)	N/A <sup>d</sup>	
<b>Paternal Race</b>					
White	56,551 (66.6)	28,305 (73.4)	27,822 (72.4)		
Black or African American	7,080 (8.3)	1,511 (3.9)	1,776 (4.6)		
Other race <sup>b</sup>	21,235 (25.0)	8,576 (22.7)	8,829 (23.0)		
<b>Principal Source of Payment</b>					
Private Insurance	39,762 (46.8)	21,495 (55.7)	21,745 (56.6)	14,940 (56.9)	15,408 (58.6)
Medicaid	39,480 (46.5)	15,530 (40.3)	15,130 (39.4)	7,951 (30.3)	8,772 (33.4)
Other <sup>c</sup>	5,714 (6.7)	1,547 (4.0)	1,552 (4.0)	3,356 (12.8)	2,104 (8.0)

Note. iEHDI = Individual Early Hearing Detection and Intervention pilot study

<sup>a</sup>2012 data for Indiana were not available<sup>b</sup>Other race includes American Indian/Alaska Native, Asian Indian, Chinese, Filipino, Japanese, Korean, Vietnamese, Other Asian, Native Hawaiian, Guamanian or Chamorro, Aamoan, other Pacific Islander, other race, multiracial (not specified), and unknown<sup>c</sup>Other principal source of payment includes CHAMPUS/Tricare, Indian Healthcare, self-pay, other, and missing.<sup>d</sup>Data were not available.

Table 4

Summary of EHDI Tracking and Surveillance Efforts, iEHDI 2010 and 2012.

Characteristic	Indiana <sup>b</sup>	Iowa		Nebraska	
	2010 n (%)	2010 n (%)	2012 n (%)	2010 n (%)	2012 n (%)
<b>Total</b>	<b>84,866</b>	<b>38,572</b>	<b>38,427</b>	<b>26,247</b>	<b>26,284</b>
<b>Screening Method</b>					
Auditory Brainstem Response or Automated Auditory Brainstem Response	72,985 (86.0)	7,870 (20.4)	10,164 (26.4)	19,260 (73.4)	18,615 (71.5)
Otoacoustic Emissions, Transient-Evoked Otoacoustic Emissions or Distortion Product Otoacoustic Emissions Tests	11,881 (14.0)	29,935 (77.6)	27,657 (72.0)	3,127 (11.9)	2,760 (10.5)
Unknown		767 (2.0)	606 (1.6)	3,860 (14.7)	4,909 (18.7)
<b>Initial Hearing Screen Results</b>					
Pass	71,185 (83.9)	35,444 (91.9)	35,837 (93.3)	24,805 (94.5)	25,185 (95.8)
Not Pass	2,511 (3.0)	2,361 (6.1)	1,957 (5.0)	1,126 (4.3)	837 (3.2)
Not Screened	2,157 (2.5)	767 (2.0)	633 (1.7)	316 (1.2)	262 (1.0)
Unknown Status	9,013 (10.6)	0 (0.0)	0 (0.0)	0 (0.0)	0 (0.0)
<b>Infants Receiving Rescreen (Total)</b>	<b>N/A<sup>a</sup></b>	<b>2,361</b>	<b>1,957</b>	<b>1,126</b>	<b>837</b>
Pass		1,735 (73.5)	1,504 (76.8)	873 (77.5)	722 (86.3)
Not Pass		303 (12.8)	219 (11.2)	48 (4.3)	58 (6.9)
Not Screened		323 (13.7)	234 (12.0)	205 (18.2)	57 (6.8)
<b>Final Hearing Screen Results (Total)</b>	<b>N/A<sup>a</sup></b>	<b>38,572</b>	<b>38,427</b>	<b>26,247</b>	<b>26,284</b>
Pass		37,404 (97.0)	37,461 (97.5)	25,783 (98.2)	26,009 (99.0)
Not Pass		645 (1.7)	462 (1.2)	263 (1.0)	120 (0.4)
Not Screened		523 (1.4)	504 (1.3)	201 (0.8)	155 (0.6)
<b>Diagnostic Evaluations Status (Total)</b>	<b>2,511</b>	<b>645</b>	<b>462</b>	<b>263</b>	<b>120</b>
Diagnosed	1,726 (68.7)	265 (41.1)	237 (51.3)	129 (49.0)	55 (45.8)
Not Diagnosed	631 (25.1)	366 (56.7)	205 (44.4)	122 (46.4)	45 (37.5)
Died or Moved	73 (2.9)	11 (1.7)	15 (3.3)	4 (1.5)	9 (7.5)
Refused	81 (3.2)	3 (0.5)	5 (0.1)	8 (3.0)	11 (9.2)
<b>Median Age Receiving First Diagnostic Evaluation ± SD (days)</b>	<b>48.0 ± 90.9</b>	<b>74.0 ± 130.8</b>	<b>48.0 ± 126.1</b>	<b>49.0 ± 105.2</b>	<b>65.0 ± 119.7</b>
<b>Diagnostic Results (Total)</b>	<b>1,726</b>	<b>265</b>	<b>237</b>	<b>129</b>	<b>55</b>
No Hearing Loss	1,441 (83.5)	142 (53.6)	181 (76.4)	70 (54.2)	17 (30.9)
Permanent Hearing Loss	138 (8.0)	78 (29.4)	53 (22.4)	49 (38.0)	38 (69.1)
Non-Permanent Hearing Loss	90 (5.2)	0 (0.0)	0 (0.0)	8 (6.2)	0 (0.0)
In Process	5 (0.3)	45 (17.0)	3 (1.3)	0 (0.0)	0 (0.0)
Unknown Status	52 (3.0)	0 (0.0)	0 (0.0)	2 (1.6)	0 (0.0)
<b>Status of EI Service for those Diagnosed with Permanent Hearing Loss (Total)</b>	<b>138</b>	<b>78</b>	<b>53</b>	<b>49</b>	<b>38</b>
Receiving EI	49 (62.8)	95 (68.8)	38 (71.7)	0 (0.0)	29 (76.3)
Not Receiving EI or Unknown	22 (28.2)	32 (23.2)	9 (17.0)	49 (100.0) <sup>c</sup>	3 (7.9)
Died or Moved	1 (1.3)	8 (7.4)	2 (3.8)	0 (0.0)	4 (10.5)
Refused	7 (9.0)	3 (2.8)	4 (7.6)	0 (0.0)	2 (5.3)

Note. EHDI = Early Hearing Detection and Intervention; EI = Early Intervention; iEHDI = Individual EHDI.

<sup>a</sup>Indiana screening protocol does not require a hearing rescreen. Screening method numbers were estimated by average percent.

<sup>b</sup>2012 data for Indiana were not available.

<sup>c</sup>EI data for Nebraska were not available for 2010.

infants had permanent HL, reflecting combined data for 2010 and 2012 for Iowa and Nebraska and only 2010 data for Indiana. This prevalence rate of HL is higher than the national prevalence rate, which is 1.3 per 1,000 live born infants in 2010 and 1.4 per 1,000 live born infants for 2012 (CDC, 2017). Table 5 also shows that more than half of the infants diagnosed with permanent HL ( $\geq 70\%$  in each jurisdiction) had bilateral HL, and most infants were born to married mothers and mothers who are White. Maternal education varied across jurisdictions. Regardless of laterality or jurisdiction, most infants had mild ( $\geq 48\%$  in each jurisdiction), sensorineural ( $\geq 60\%$  in each

jurisdiction) HL. Although, it is feasible to estimate the prevalence of HL using the HSFS data, the ability to better understand both the infant and maternal characteristics of infants diagnosed with permanent HL is not feasible using current HSFS data.

## Discussion

As learned from the iEHDI pilot, individual-level data offered many opportunities for CDC. The pilot study allowed CDC and jurisdictional EHDI programs to



**Table 5**

*Summary of Infant and Maternal Characteristics for Infants Diagnosed with Permanent Hearing Loss by Jurisdiction, Individual Early Hearing and Detection Intervention (iEHDI) Pilot Study 2010 and 2012*

Characteristic	Indiana <sup>a</sup>	Iowa	Nebraska
	n (%)	n (%)	n (%)
<b>Infants with Permanent Hearing Loss</b>	<b>138 (0.2)</b>	<b>131 (0.2)</b>	<b>87 (0.2)</b>
<b>Gender of Infant</b>			
Male	75 (54.3)	70 (53.4)	45 (51.7)
Female	63 (45.7)	61 (46.6)	42 (48.3)
<b>Laterality</b>			
Bilateral Hearing Loss	96 (69.6)	97 (74.0)	38 (71.7)
Unilateral Hearing Loss	42 (30.4)	34 (26.0)	15 (28.3)
<b>Type of hearing loss, right ear</b>			
Sensorineural	93 (67.4)	100 (76.3)	53 (60.9)
Permanent Conductive	7 (5.1)	1 (0.8)	7 (8.0)
Mixed	9 (6.5)	7 (5.3)	12 (13.8)
Auditory Neuropathy	6 (4.4)	1 (0.8)	3 (3.4)
Unknown Type	0 (0.0)	0 (0.0)	6 (6.9)
No Hearing Loss	23 (16.7)	22 (16.8)	6 (6.9)
<b>Type of hearing loss, left ear</b>			
Sensorineural	93 (67.4)	110 (84.0)	51 (58.6)
Permanent Conductive	7 (5.1)	3 (0.2)	3 (3.4)
Mixed	12 (8.7)	5 (3.8)	10 (11.5)
Auditory Neuropathy	7 (5.1)	1 (0.8)	1 (1.1)
Unknown Type	0 (0.0)	0 (0.0)	9 (10.30)
No Hearing Loss	19 (13.8)	12 (9.2)	13 (14.9)
<b>Severity of hearing loss, right ear</b>			
Mild	77 (55.8)	64 (48.9)	44 (50.6)
Severe	14 (10.1)	17 (13.0)	15 (17.2)
Profound	16 (11.6)	12 (9.2)	20 (23.0)
Unknown or Missing	31 (22.5)	38 (29.0)	8 (9.2)
<b>Severity of hearing loss, left ear</b>			
Mild	78 (56.5)	65 (49.6)	42 (48.3)
Severe	16 (11.6)	22 (16.8)	9 (10.3)
Profound	16 (11.6)	13 (1.0)	23 (26.4)
Unknown or Missing	28 (20.3)	31 (23.7)	13 (14.9)
<b>Birth weights (g)</b>			
Low (< 2500)	25 (18.1)	13 (9.9)	
Normal (2500-4000)	103 (74.6)	109 (83.2)	
High (≥ 4001)	10 (7.3)	9 (6.9)	
			N/A <sup>c</sup>
<b>Neonatal Intensive care &gt; 5 days</b>	35 (25.4)	28 (21.4)	
<b>Low APGAR score (&lt; 6) at 5 minutes</b>	7 (5.1)	10 (7.6)	
<b>Family History of Permanent Child Hearing Loss</b>	36 (26.1)	12 (9.2)	
<b>Maternal Average Age ± SE (years)</b>	28.5 ± 6.7	27.6 ± 5.5	28.3 ± 6.8
<b>Maternal Hispanic Ethnicity</b>	18 (13.0)	4 (3.1)	19 (21.8)
<b>Maternal Race</b>			
White	86 (62.3)	114 (87.0)	64 (73.6)
Black or African American	11 (8.0)	9 (6.9)	6 (6.9)
Other <sup>b</sup>	41 (29.7)	8 (6.1)	17 (19.5)
<b>Maternal Education</b>			
< High School or Unknown	33 (23.9)	16 (12.2)	18 (20.7)
Completed High School	35 (25.4)	26 (19.8)	18 (20.7)
Some College or Associate's Degree	30 (21.7)	51 (38.9)	27 (31.0)
≥ Bachelor's Degree	40 (29.0)	38 (29.0)	24 (27.6)
<b>Marital Status</b>			
Married	80 (58.0)	79 (60.3)	59 (67.8)
Not Married/Unknown/Missing	58 (42.0)	52 (39.7)	28 (32.2)

<sup>a</sup>2012 data for Indiana were not available

<sup>b</sup>Other race/ethnicity includes American Indian/Alaska Native, Asian, Native Hawaiian/Pacific Islander, other, multiracial not specified, and unknown.

<sup>c</sup>Data were not available.



collaborate and identify data quality issues (e.g., an infant's date of hearing screening occurring before the infant's date of birth and inconsistent screening and diagnostic results for a baby diagnosed with no HL) and implement procedures to correct them. It also highlighted inconsistencies in data standardization, which can adversely affect the quality and accuracy of data (King, 2013). For instance, the definition of passing the hearing screen varied from jurisdiction to jurisdiction, depending on the screening protocol used, which also differed between jurisdictions. According to Indiana's mandate, an infant is referred directly for a diagnostic evaluation after not passing two inpatient screenings. Alternatively, Iowa and Nebraska require an outpatient screen only if the infant did not pass the initial inpatient screen. In addition, the pilot study revealed that the data collection and reporting procedures vary from jurisdiction to jurisdiction. The processes by which infants receive recommended follow-up services vary in each jurisdiction. The data collection and process issues would not have been identified if the CDC EHDI program had relied only on HSFS data.

The study also allowed CDC to gain a better understanding of the challenges unique to each jurisdiction in terms of data collection and reporting. For example, EHDI data collected in one jurisdiction were captured from multiple sources and the relationship between discrete data items from the multiple sources were not always consistent. One data source might have documented a newborn passing the hearing screen for both ears while in another data source the same newborn was documented as failing hearing screen in one ear. Another challenge for jurisdictions was the time-consuming process of matching the newborn screening record report with the vital records report to create a final record with all variables for the iEHDI pilot. Due to the iEHDI partnership, a matching algorithm was used to automate this matching process. The algorithm enabled the jurisdictional EHDI program staff to match different iterations of the data or record by deterministic, probabilistic, or other types of similar measures and led to improvements in efficiency. This challenge would not have come to light without the pilot study. In terms of reporting data to CDC, one jurisdiction experienced the challenge of converting several data items in the jurisdictional database from text to numeric format to fulfill the iEHDI data requirements. They had to import certain data items from other sources (i.e., the Federal Information Processing Standard county code from the birth record into the jurisdictional database before transmitting the data to CDC). Another challenge noted in this pilot study was the increased costs for the participating jurisdictional EHDI programs to provide limited, de-identified datasets to CDC. The increased costs were due to the amount of personnel time and effort for the data management and collection required for this study, which were substantial for the jurisdictions. The increased costs were also due to upgrades made to the tracking and surveillance system, which in some cases, were necessary to make the pilot study feasible. The upgrades, while beneficial to the programs, are often times costly and the

jurisdictions were challenged to find the financial means to make the upgrades feasible. This collaboration allowed for CDC to understand the challenges and the substantial efforts required from the participating jurisdictions to report individual-level data. Through this collaboration, CDC recognized that data standardization and more refined definitions are needed.

A major benefit seen in the pilot study is the availability of far more data items compared to HSFS (Table 2). Unlike HSFS, the iEHDI pilot gathered data on WIC enrollment status, paternal characteristics, infant birth characteristics, and risk factors for HL. Although these data items are already gathered at the jurisdictional level, the availability of these data items in the pilot study allowed for CDC to further understand each jurisdiction's infant population and their EHDI process. It also allowed for more research opportunities.

Individual-level data allow for in-depth statistical analyses, which is another benefit seen in the pilot study. In addition to learning more about each jurisdiction's infant population and their EHDI process, the individual-level data also allowed for more discussions between CDC and the jurisdictional EHDI programs. For instance, analyses revealed that Indiana had the largest birth cohort, yet a lower than expected proportion of newborns underwent initial newborn hearing screening. The analyses also revealed that even though the jurisdictions varied in birth cohort, the number and percentage of infants receiving newborn hearing screening and diagnostic evaluation were wide-ranging. This prompted questions about why the percentages seen are different and provided opportunity for discussions between CDC and jurisdictional EHDI programs, which is currently not feasible using HSFS data.

In addition, individual-level data allowed for identification and tracking of infants at different stages of the EHDI process and ability to assess the demographic and socioeconomic characteristics that may be associated with the receipt of recommended screening, diagnostic, and/or intervention services. It was feasible to look at subsets of interests in further detail. For instance, we learned that for Indiana in 2010, 25.1% of the infants who did not pass the hearing screen as final result were not documented as having received a diagnostic evaluation (Table 4). Also for Indiana in 2010, we learned that 26.1% of the infants with permanent HL have family history of permanent childhood HL and 69.6% of the infants with permanent HL have bilateral HL (Table 5). The ability to assess subgroups in detail is not feasible using the current HSFS data. This pilot study demonstrated that key measures using individual-level data could be calculated at the national level which is not currently feasible using HSFS data (e.g., median age at first diagnostic visit, median age at referral, and median age when enrolled into early intervention). The ability to calculate these key measures allowed for assessing progress toward meeting the 1-3-6 goals which are measured by Healthy People 2020 Objective ENT-VSL-1 and three child health quality measures that were

endorsed by the National Quality Forum (NQF) in August 2011 (NQF #1354: hearing screening before discharge from the hospital, NQF #1360: audiological evaluation no later than age 3 months [for those failing the screening], and NQF #1361: intervention no later than age 6 months [for those identified with a HL]; U.S. Department of Health and Human Services, 2018; National Quality Forum, 2018). The ability to calculate key measures allowed for more opportunities for improvement through targeted technical assistance from CDC.

## Conclusions

Because of the limitations of HSFS data, the iEHDI pilot study was implemented to explore the feasibility, benefits, and challenges surrounding reporting of individual-level data from the jurisdictional EHDI programs to CDC. Findings of the pilot study demonstrated that reporting of individual-level data to CDC is feasible and more in-depth analyses benefit both CDC and jurisdictional EHDI programs. More importantly, it offered an opportunity for CDC and jurisdictional EHDI programs to collaborate to identify, discuss, and implement procedures to improve the quality and usefulness of data in ensuring infants receive recommended screening, diagnostic, and EI services. In-depth analyses also increased CDC's understanding of each jurisdiction's EHDI process, making it possible to detail EHDI tracking and surveillance efforts and for CDC to better understand the gaps and needs of each jurisdictional EHDI program. This in turn allows for CDC to provide more targeted and relevant technical assistance to the jurisdictions. All of the above are not feasible using the currently reported HSFS data. Although there were challenges in reporting individual-level data, benefits seen in this pilot study outweighed the challenges. Lessons learned from this iEHDI pilot were used to inform and guide current activities and procedures for expanding EHDI data collection at CDC. This includes refining data definitions and incorporating activities from the pilot study into the ten jurisdictional EHDI programs currently funded to gather and report individual-level data.

## References

- Alam, S., Gaffney, M., & Eichwald, J. (2014, Mar-Apr). Improved newborn hearing screening follow-up results in more infants identified. *Journal of Public Health Management and Practice*, 20(2), 220–223. doi: [10.1097/PHH.0b013e31829d7b57](https://doi.org/10.1097/PHH.0b013e31829d7b57)
- Alam, S., Satterfield, A., Mason, C. A., & Deng, X. (2016). Progress in standardization of reporting and analysis of data from Early Hearing Detection and Intervention (EHDI) Programs. *Journal of Early Hearing Detection and Intervention*, 1(2), 2–7.
- American Speech-Language-Hearing Association. (2017a). Degree of Hearing Loss. Retrieved from <http://www.asha.org/public/hearing/Degree-of-Hearing-Loss/>
- American Speech-Language-Hearing Association. (2017b). Types of Hearing Loss. Retrieved from <http://www.asha.org/public/hearing/Types-of-Hearing-Loss/>
- Biswas, A. K., Goswami, S. C., Baruah, D. K., & Tripathy, R. (2012). The potential risk factors and the identification of hearing loss in infants. *Indian Journal of Otolaryngology and Head & Neck Surgery*, 64(3), 214–217. doi: [10.1007/s12070-011-0307-6](https://doi.org/10.1007/s12070-011-0307-6)
- Centers for Disease Control and Prevention. (2017). Annual Data Early Hearing Detection and Intervention (EHDI) Program. Retrieved from <https://www.cdc.gov/ncbddd/hearingloss/ehdi-data.html>
- Health Research & Services Administration. (2016). Federal Advisory Committees: Recommended Uniform Screening Panel. Retrieved from <https://www.hrsa.gov/advisory-committees/heritable-disorders/rusp/index.html>
- King, G. (2013). *A solution to the ecological inference problem: Reconstructing individual behavior from aggregate data*. Princeton, NJ: Princeton University Press.
- Lin, J. & Oghalai, J. S. (2011). Towards an etiologic diagnosis: Assessing the patient with hearing loss. *Advances in Oto-rhino-laryngology*, 70, 28–36. doi: [10.1159/000322468](https://doi.org/10.1159/000322468)
- National Quality Forum. (2018). Measures, Reports & Tools. Retrieved from [http://www.qualityforum.org/Measures\\_Reports\\_Tools.aspx](http://www.qualityforum.org/Measures_Reports_Tools.aspx)
- Stewart, L. A. & Tierney, J. F. (2002, Mar). To IPD or not to IPD? Advantages and disadvantages of systematic reviews using individual patient data. *Evaluation & the Health Professions*, 25(1), 76–97.
- U.S. Department of Health and Human Services. (2018). Healthy people 2010: Hearing and other sensory or communication disorders. Retrieved from [www.healthypeople.gov/2020/topicsobjectives2020/objectiveslist.aspx?topicid=20](http://www.healthypeople.gov/2020/topicsobjectives2020/objectiveslist.aspx?topicid=20)
- Vohr B. (2003). Overview: Infants and children with hearing loss—part I. *Mental Retardation and Developmental Disabilities Research Reviews*, 9(2), 62–64. doi: [10.1002/mrdd.10070](https://doi.org/10.1002/mrdd.10070)
- Williams, T., Alam, S., & Gaffney, M. (2015, April). Progress in identifying infants with hearing loss—United States, 2006–2012. *Morbidity Mortality Weekly Report*, 64(13), 351–356.
- Winston-Gerson, R. & Hoffman, J. (2017). Chapter 3: Tracking, reporting, & follow-up. In *NCHAM eBook: A Resource Guide for Early Hearing Detection and Intervention*. Retrieved from [http://www.infantheating.org/ehdi-ebook/2017\\_ebook/3%20Chapter3Tracking2017.pdf](http://www.infantheating.org/ehdi-ebook/2017_ebook/3%20Chapter3Tracking2017.pdf) Logan, UT: National Center for Hearing Assessment and Management.

## Others' Publications About EHDl: October 2017 through April 2018

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

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

Of the 72 articles published in other journals from October 2017 through April 2018, most reported on research conducted in countries other than the United States. Clearly, EHDl continues to be a global phenomenon. Staying current with EHDl's processes and accomplishments requires staying informed about what is happening in other countries. During this time period, the topics most frequently addressed included issues related to the identification or treatment of congenital cytomegalovirus (cCMV) and its relation to childhood hearing loss, the genetics of hearing loss, the use of cochlear implants, language outcomes for children identified in newborn hearing screening programs, and issues related to protocols and procedures used in newborn hearing screening, follow-up, and diagnosis.

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

- Cejas et al. reported that children using cochlear implants perform similarly to hearing peers on measures of intelligence, but those with severe comorbidities are at-risk for cognitive deficits.
- Fitzpatrick et al. in a cross-sectional analysis of 120 children, found that children with unilateral hearing loss tended to perform poorer than those in the mild bilateral hearing loss or normal hearing.
- Grasty et al. followed 348 children who underwent repair of congenital heart disease and found that that the prevalence of hearing loss in preschool children after heart surgery in infancy was 20-fold higher than in the 1% prevalence seen in the general population.
- Hao et al. demonstrated that newborn hearing genetic screening could be used in conjunction with newborn hearing screening to identify four genetic mutations frequently associated with congenital or late-onset hearing loss. Genetic mutations were found for 3.01% of the 142,417 neonates screened.
- Rawlinson et al. screened infants for cCMV using PCR of urine and saliva and found ~6% of the large sample of children who had failed a newborn hearing screening test were positive for cCMV. In contrast, Vancor et al., found only 2 of 171 (1.2%) infants who failed newborn hearing screening had confirmed cCMV based on PCR of saliva.
- Zych et al. reported on 14 years of data from the Polish Universal neonatal Hearing Screening Program.

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

**Adebanjo T, Godfred-Cato S, Viens L, Fischer M, Staples JE, Kuhnert-Tallman W, Walke H, Oduyibo T, Polen K, Peacock G, Meaney-Delman D, Honein MA, Rasmussen SA, Moore CA** [Update: Interim Guidance for the Diagnosis, Evaluation, and Management of Infants with Possible Congenital Zika Virus Infection - United States, October 2017.](#) *MMWR Morb Mortal Wkly Rep.* 2017 Oct 20;66(41):1089-1099. doi: 10.15585/mmwr.mm6641a1.

CDC has updated its interim guidance for U.S. health care providers caring for infants with possible congenital Zika virus infection (1) in response to recently published updated guidance for health care providers caring for pregnant women with possible Zika virus exposure (2), unknown sensitivity and specificity of currently available diagnostic tests for congenital Zika virus infection, and recognition of additional clinical findings associated with congenital Zika virus infection. All infants born to mothers with possible Zika virus exposure\* during pregnancy should receive a standard evaluation at birth and at each subsequent well-child visit including a comprehensive physical examination, age-appropriate vision screening and developmental monitoring and screening using validated tools (3-5), and newborn hearing screen at birth, preferably using auditory brainstem response (ABR) methodology (6). Specific guidance for laboratory testing and clinical evaluation are provided for three clinical scenarios in the setting of possible maternal Zika virus exposure: 1) infants with clinical findings consistent with congenital Zika syndrome regardless of maternal testing results, 2) infants without clinical

findings consistent with congenital Zika syndrome who were born to mothers with laboratory evidence of possible Zika virus infection,† and 3) infants without clinical findings consistent with congenital Zika syndrome who were born to mothers without laboratory evidence of possible Zika virus infection. Infants in the first two scenarios should receive further testing and evaluation for Zika virus, whereas for the third group, further testing and clinical evaluation for Zika virus are not recommended. Health care providers should remain alert for abnormal findings (e.g., postnatal-onset microcephaly and eye abnormalities without microcephaly) in infants with possible congenital Zika virus exposure without apparent abnormalities at birth.

**Appelbaum EN, Howell JB, Chapman D, Pandya A, Dodson KM.**

[Analysis of risk factors associated with unilateral hearing loss in children who initially passed newborn hearing screening.](#) *Int J Pediatr Otorhinolaryngol.* 2018 Mar;106:100-104. doi: 10.1016/j.ijporl.2018.01.024. Epub 2018 Feb 2.

**OBJECTIVE:** To analyze 2007 Joint Committee on Infant Hearing (JCIH) risk factors in children with confirmed unilateral hearing loss (UHL) who initially passed newborn hearing screening.

**METHODS:** Retrospective record review of 16,108 infants who passed newborn hearing screening but had one or more JCIH risk factors prompting subsequent follow-up through the universal newborn hearing screening (UNHS) program in Virginia from 2010 to 2012. The study was reviewed and qualified as exempt by the Virginia Commonwealth University Institutional Review Board (IRB) and the Virginia Department of Health.

**RESULTS:** Over the 2-year study period, 14896 (4.9% of total births) children passed UNHS but had the presence of one or more JCIH risk factor. Ultimately, we identified 121 babies from this group with confirmed hearing loss (0.7%), with 48 babies (0.2%) showing UHL. The most common risk factors associated with the development of confirmed UHL after passing the initial screen were neonatal indicators, craniofacial anomalies, family history, and stigmata of syndrome associated with hearing loss.

**CONCLUSION:** Neonatal indicators and craniofacial anomalies were the categories most often found in children with confirmed unilateral hearing loss who initially passed their newborn hearing screen. While neonatal indicators were also the most common associated risk factor in all hearing loss, craniofacial abnormalities are relatively more common in children with UHL who initially passed newborn hearing screening. Further studies assessing the etiology underlying the hearing loss and risk factor associations are warranted.

**Bianchin G, Tribi L, Formigoni P, Russo C, Polizzi V.**

[Sequential pediatric bilateral cochlear implantation: The effect of time interval between implants.](#) *Int J Pediatr Otorhinolaryngol.* 2017 Nov;102:10-14. doi: 10.1016/j.ijporl.2017.08.025. Epub 2017 Aug 25.

**OBJECTIVE:** To examine speech intelligibility in children subjected to sequential bilateral cochlear implants (CI) surgery and to assess the influence of the inter-stage interval duration.

**INTRODUCTION:** Binaural hearing recovery can have additional benefits, especially in speech and language development in patients with congenital profound sensorineural hearing loss; so recently there has been an increase in the number of children receiving bilateral CI.

**METHODS:** Twenty-seven children who underwent sequential bilateral cochlear implant (SBCI) with a short (1-3 yrs), medium (4-6 yrs) and long (7-12 yrs) range interval between both implantations, respectively, were evaluated. All patients underwent periodic speech perception test in quiet and noise after second implant activation in three conditions: with the first or second implant alone and with both implants. Results were examined according to the inter-stage interval.

**RESULTS:** Speech intelligibility in noise was significantly better under bilateral conditions than either ear alone, in all three groups. Small improvements were seen in quiet, especially in the third group (6-12 yrs).

**CONCLUSION:** Benefits of second implant in the early-implanted children and after a short inter-implant delay are more evident. However our study support that, even after a long period of deafness and despite a prolonged inter-stage interval, sequential bilateral cochlear implantation should be considered.

**LEVEL OF EVIDENCE:** Level 4.

**DISCUSSION:** Prophylactic HIG administration in pregnant women after CMV primary infection seems not to reduce significantly the rate of congenital infection, but is safe and it could have a favorable effect on the symptoms and sequelae of infected fetuses. The risk of long-term sequelae in fetuses without US abnormalities before HIG is low, so it could be an option in infected fetuses with normal imaging. On the other hand, the risk of sequelae among infected fetuses with abnormalities in fetal ultrasonography before HIG despite treatment is high.

**Bostic K, Lewis RM, Chai B, Manganella JL, Barrett DL, Kawai K, Kenna MA, Stiles DJ, Clark T.**

[Enlarged Vestibular Aqueduct and Cochlear Implants: The Effect of Early Counseling on the Length of Time Between Candidacy and Implantation.](#) *Otol Neurotol.* 2018 Feb;39(2):e90-e95. doi: 10.1097/MAO.0000000000001663.

**OBJECTIVE:** To determine if discussing cochlear implantation (CI) with patients with enlarged vestibular aqueducts (EVA) and their families before reaching audiological criteria for CI candidacy effects the length of time between reaching audiological candidacy and CI surgery, and to describe the universal newborn hearing screening (UNHS) results and communication modality in this sample. **PATIENTS:** Forty-two patients (25 females) with confirmed EVA and cochlear implants.



**INTERVENTION(S):** Diagnostic CI visit.

**MAIN OUTCOME MEASURES:** The primary outcome measure is the difference in length of time between reaching audiological candidacy for CI and surgical implantation between those who had preliminary discussions regarding CI with their medical and healthcare providers before reaching audiological candidacy versus who had discussions after reaching candidacy. The secondary outcome measure is the result of the UNHS and primary mode of communication used by each patient.

**RESULTS:** Discussing CI before reaching audiological candidacy was associated with a significantly shorter duration between reaching audiological candidacy and receiving CI (median=3.1 mo; interquartile range [IQR]=1.7-5.4) as compared with discussing CI after reaching candidacy (median=5.8 mo; IQR=3.2-11.2;  $p=0.012$ ). Participants born after the implementation of the UNHS, 16 of 24 patients referred on one or both ears. Communication modalities were evenly divided between utilizing sign-support English and oral/aural communicators only.

**CONCLUSIONS:** Discussion of CI in patients with EVA before reaching audiological candidacy reduces the amount of time the child is without adequate auditory access and contributes to a constructive and interactive preparatory experience.

**Bright T, Mulwafu W, Thindwa R, Zuurmond M, Polack S.**

[Reasons for low uptake of referrals to ear and hearing services for children in Malawi.](#)

*PLoS One.* 2017 Dec 19;12(12):e0188703. doi: 10.1371/journal.pone.0188703. eCollection 2017.

**BACKGROUND:** Early detection and appropriate intervention for children with hearing impairment is important for maximizing functioning and quality of life. The lack of ear and hearing services in low income countries is a significant challenge, however, evidence suggests that even where such services are available, and children are referred to them, uptake is low. The aim of this study was to assess uptake of and barriers to referrals to ear and hearing services for children in Thyolo District, Malawi.

**METHODS:** This was a mixed methods study. A survey was conducted with 170 caregivers of children who were referred for ear and hearing services during community-based screening camps to assess whether they had attended their referral and reasons for non-attendance. Semi-structured interviews were conducted with 23 caregivers of children who did not take up their referral to explore in-depth the reasons for non-uptake. In addition, 15 stakeholders were interviewed. Thematic analysis of the interview data was conducted and emerging trends were analysed.

**RESULTS:** Referral uptake was very low with only 5 out of 150 (3%) children attending. Seven main interacting themes for non-uptake of referral were identified in the semi-structured interviews: location of the hospital, lack of transport, other indirect costs of seeking care, fear and uncertainty about the referral hospital, procedural problems within the camps, awareness and understanding of hearing loss, and lack of visibility and availability of services.

**CONCLUSION:** This study has highlighted a range of interacting challenges faced by families in accessing ear and hearing services in this setting. Understanding these context specific barriers to non-uptake of ear and hearing services is important for designing appropriate interventions to increase uptake.

**LEVEL OF EVIDENCE:** 1b. *Laryngoscope*, 127:S1-S13, 2017.

**Cejas I, Mitchell CM, Hoffman M, Quittner AL; and the CDaCI Investigative Team.**

[Comparisons of IQ in Children With and Without Cochlear Implants: Longitudinal Findings and Associations With Language.](#)

*Ear Hear.* 2018 Apr 5. doi: 10.1097/AUD.0000000000000578. [Epub ahead of print]

**OBJECTIVES:** To make longitudinal comparisons of intelligence quotient (IQ) in children with cochlear implants (CIs) and typical hearing peers from early in development to the school-age period. Children with additional comorbidities and CIs were also evaluated. To estimate the impact of socioeconomic status and oral language on school-age cognitive performance.

**DESIGN:** This longitudinal study evaluated nonverbal IQ in a multicenter, national sample of 147 children with CIs and 75 typically hearing peers. IQ was evaluated at baseline, prior to cochlear implantation, using the Bayley Scales of Infant and Toddler Development and the Leiter International Performance Scale. School-age IQ was assessed using the Wechsler Intelligence Scales for Children. For the current study, only the Perceptual Reasoning and Processing Speed indices were administered. Oral language was evaluated using the Comprehensive Assessment of Spoken Language.

**RESULTS:** Children in the CI group scored within the normal range of intelligence at both time points. However, children with additional comorbidities scored significantly worse on the Processing Speed, but not the Perceptual Reasoning Index. Maternal education and language were significantly related to school-age IQ in both groups. Importantly, language was the strongest predictor of intellectual functioning in both children with CIs and normal hearing.

**CONCLUSION:** These results suggest that children using cochlear implants perform similarly to hearing peers on measures of intelligence, but those with severe comorbidities are at-risk for cognitive deficits. Despite the strong link between socioeconomic status and intelligence, this association was no longer significant once spoken language performance was accounted for. These results reveal the important contributions that early intervention programs, which emphasize language and parent training, contribute to cognitive functioning in school-age children with CIs. For families from economically disadvantaged backgrounds, who are at-risk for suboptimal outcomes, these early intervention programs are critical to improve overall functioning.

**Chorozoglou M, Mahon M, Pimperton H, Worsfold S, Kennedy CR.**

[Societal costs of permanent childhood hearing loss at teen age: a cross-sectional cohort follow-up study of universal newborn hearing screening.](#)

*BMJ Paediatr Open.* 2018 Feb 24;2(1):e000228. doi: 10.1136/bmjpo-2017-000228. eCollection 2018.

**Objective:** To investigate the effects in adolescence of bilateral permanent childhood hearing loss (PCHL) > 40 dB and of exposure to universal newborn hearing screening (UNHS) on societal costs accrued over the preceding 12 months.

**Design setting participants:** An observational cohort study of a sample of 110 adolescents aged 13–20 years, 73 with PCHL and 37 in a normally hearing comparison group (HCG) closely similar in respect of place and date of birth to those with PCHL, drawn from a 1992–1997 cohort of 157 000 births in Southern England, half of whom had been exposed to a UNHS programme.

**Intervention:** Birth in periods with and without UNHS. Outcome measures: Resource use and costs in the preceding 12-month period, estimated from interview at a mean age of 16.9 years and review of medical records. Effects on costs were examined in regression models.

**Results:** Mean total costs for participants with PCHL and the HCG were £15 914 and £5883, respectively (difference £10 031, 95% CI £6460 to £13 603), primarily driven by a difference in educational costs. Compared with the HCG, additional mean costs associated with PCHL of moderate, severe and profound severity were £5916, £6605 and £18437, respectively. The presence of PCHL and an additional medical condition (AMC) increased costs by £15 385 (95% CI £8532 to £22 238). An increase of one unit in receptive language z-score was associated with £1616 (95% CI £842 to £2389) lower costs. Birth during periods of UNHS was not associated with significantly lower overall costs (difference £3594, 95% CI -£2918 to £10 106).

**Conclusions:** The societal cost of PCHL was greater with more severe losses and in the presence of AMC and was lower in children with superior language scores. There was no statistically significant reduction in costs associated with birth in periods with UNHS. Trial registration number: ISRCTN03307358, pre-results.

**Daub O, Bagatto MP, Johnson AM, Cardy JO.**

[Language Outcomes in Children Who Are Deaf and Hard of Hearing: The Role of Language Ability Before Hearing Aid Intervention.](#)

*J Speech Lang Hear Res.* 2017 Nov 9;60(11):3310–3320. doi: 10.1044/2017\_JSLHR-L-16-0222.

**Purpose:** Early auditory experiences are fundamental in infant language acquisition. Research consistently demonstrates the benefits of early intervention (i.e., hearing aids) to language outcomes in children who are deaf and hard of hearing. The nature of these benefits and their relation with prefitting development are, however, not well understood.

**Method:** This study examined Ontario Infant Hearing Program birth cohorts to explore predictors of performance on the Preschool Language Scale–Fourth Edition at the time of (N = 47) and after (N = 19) initial hearing aid intervention.

**Results:** Regression analyses revealed that, before the hearing aid fitting, severity of hearing loss negatively predicted 19% and 10% of the variance in auditory comprehension and expressive communication, respectively. After hearing aid fitting, children's standard scores on language measures remained stable, but they made significant improvement in their progress values, which represent individual skills acquired on the test, rather than standing relative to same-age peers. Magnitude of change in progress values was predicted by a negative interaction of prefitting language ability and severity of hearing loss for the Auditory Comprehension scale.

**Conclusions:** These findings highlight the importance of considering a child's prefitting language ability in interpreting eventual language outcomes. Possible mechanisms of hearing aid benefit are discussed.

**Supplemental Materials:** <https://doi.org/10.23641/asha.5538868>.

**Dejaco D, Aregger FC, Hurth HV, Kegele J, Muigg V, Oberhammer L, Bunk S, Fischer N, Pinggera L, Riedl D, Otieno A, Agbenyega T, Adegnikaa AA, Riechelmann H, Lackner P, Zorowka P, Kremsner P, Schmutzhard.**

[Evaluation of transient-evoked otoacoustic emissions in a healthy 1 to 10 year pediatric cohort in Sub-Saharan Africa.](#)

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

**OBJECTIVE:** Transient-evoked otoacoustic emissions (TEOAEs) monitor cochlear function. High pass rates have been reported for industrialized countries. Pass rates in low and middle income countries such as Sub-Saharan Africa are rare, essentially lower and available for children up to 4 years of age and frequently based on hospital recruitments. This study aims at providing additional TEOAE pass rates of a healthy Sub-Saharan cohort aged 1–10 years with data from Gabon, Ghana and Kenya. Potentially confounding factors (recruitment site, age) are taken into consideration.

**METHODS:** Healthy children were recruited in hospitals, schools and kindergartens. Inclusion criteria were age 1–10 years and normal otoscopic findings. Exclusion criteria were any sickness or physical ailment potentially impairing the hearing capacity. Five measurements per ear were performed with Capella Cochlear Emission Analyzer (MADSEN, Germany). An overall wave reproducibility of above 60% served as pass-criterion. Pass rates were compared between recruitment sites and age groups (1–5 and 6–10 years).

**RESULTS:** Overall pass rate was 87.5% (n = 264; 231 passes vs. 33 fails). Of these 84.0% of hospital recruited children passed (n = 156; 131 passes vs. 25 fails), compared to 92.6% of community recruitments (n = 108; 100 passes vs. 8 fails), which was significantly different p = 0.039). If analyzed by age groups, this difference was only observed in children younger than 6 years (p = 0.007).



**CONCLUSION:** Hospitals as recruitment sites for healthy controls seem to affect TEOAE pass rates. We advise for a cautious approach when recruiting healthy TEOAE control collectives under the age of 6 in a hospital setting. In children older than 6 years conventional pure-tone audiometry remains the standard method for hearing screening.

**Dhondt CMC, Swinnen FKR, Dhooge IJM.**

[Bilateral cochlear implantation or bimodal listening in the paediatric population: Retrospective analysis of decisive criteria.](#)

*Int J Pediatr Otorhinolaryngol.* 2018 Jan;104:170-177. doi: 10.1016/j.ijporl.2017.10.043. Epub 2017 Nov 7.

**INTRODUCTION:** In children with bilateral severe to profound hearing loss, bilateral hearing can be achieved by either bimodal stimulation (CIHA) or bilateral cochlear implantation (BICI). The aim of this study was to analyse the audiologic test protocol that is currently applied to make decisions regarding the bilateral hearing modality in the paediatric population.

**METHODS:** Pre- and postoperative audiologic test results of 21 CIHA, 19 sequential BICI and 12 simultaneous BICI children were examined retrospectively.

**RESULTS:** Deciding between either simultaneous BICI or unilateral implantation was mainly based on the infant's pre-operative Auditory Brainstem Response thresholds. Evolution from CIHA to sequential BICI was mainly based on the audiometric test results in the contralateral (hearing aid) ear after unilateral cochlear implantation. Preoperative audiometric thresholds in the hearing aid ear were significantly better in CIHA versus sequential BICI children ( $p < 0.001$  and  $p = 0.001$  in unaided and aided condition, respectively). Decisive values obtained in the hearing aid ear in favour of BICI were: An average hearing threshold measured at 0.5, 1, 2 and 4 kHz of at least 93 dB HL without, and at least 52 dB HL with hearing aid together with a 40% aided speech recognition score and a 70% aided score on the phoneme discrimination subtest of the Auditory Speech Sounds Evaluation test battery.

**CONCLUSIONS:** Although pure tone audiometry offers no information about bimodal benefit, it remains the most obvious audiometric evaluation in the decision process on the mode of bilateral stimulation in the paediatric population. A theoretical test protocol for adequate evaluation of bimodal benefit in the paediatric population is proposed.

**Fitzpatrick EM, Gaboury I, Durieux-Smith A, Coyle D, Whittingham J, Nassrallah F.**

[Auditory and language outcomes in children with unilateral hearing loss.](#)

*Hear Res.* 2018 Mar 13. pii: S0378-5955(17)30363-5. doi: 10.1016/j.heares.2018.03.015. [Epub ahead of print]

**OBJECTIVES:** Children with unilateral hearing loss (UHL) are being diagnosed at younger ages because of newborn hearing screening. Historically, they have been considered at risk for difficulties in listening and language development. Little information is available on contemporary cohorts of children identified in the early months of life. We examined auditory and language acquisition outcomes in a contemporary cohort of early-identified children with UHL and compared their outcomes at preschool age with peers with mild bilateral loss and with normal hearing.

**DESIGN:** As part of the Mild and Unilateral Hearing Loss in Children Study, we collected auditory and spoken language outcomes on children with unilateral, bilateral hearing loss and with normal hearing over a four-year period. This report provides a cross-sectional analysis of results at age 48 months. A total of 120 children (38 unilateral and 31 bilateral mild, 51 normal hearing) were enrolled in the study from 2010 to 2015. Children started the study at varying ages between 12 and 36 months of age and were followed until age 36-48 months. The median age of identification of hearing loss was 3.4 months (IQR: 2.0, 5.5) for unilateral and 3.6 months (IQR: 2.7, 5.9) for the mild bilateral group. Families completed an intake form at enrolment to provide baseline child and family-related characteristics. Data on amplification fitting and use were collected via parent questionnaires at each annual assessment interval. This study involved a range of auditory development and language measures. For this report, we focus on the end of follow-up results from two auditory development questionnaires and three standardized speech-language assessments. Assessments included in this report were completed at a median age of 47.8 months (IQR: 38.8, 48.5). Using ANOVA, we examined auditory and language outcomes in children with UHL and compared their scores to children with mild bilateral hearing loss and those with normal hearing.

**RESULTS:** On most measures, children with UHL performed poorer than those in the mild bilateral and normal hearing study groups. All children with hearing loss performed at lower levels compared to the normal hearing control group. However, mean standard scores for the normal hearing group in this study were above normative means for the language measures. In particular, children with UHL showed gaps compared to the normal hearing control group in functional auditory listening and in receptive and expressive language skills (three quarters of one standard deviation below) at age 48 months. Their performance in receptive vocabulary and speech production was not significantly different from that of their hearing peers.

**CONCLUSIONS:** Even when identified in the first months of life, children with UHL show a tendency to lag behind their normal hearing peers in functional auditory listening and in receptive and expressive language development.

**Forli F, Giuntini G, Ciabotti A, Bruschini L, Löfkvist U, Berrettini S.**

[How does a bilingual environment affect the results in children with cochlear implants compared to monolingual-matched children? An Italian follow-up study.](#)

*Int J Pediatr Otorhinolaryngol.* 2018 Feb;105:56-62. doi: 10.1016/j.ijporl.2017.12.006. Epub 2017 Dec 8.

**OBJECTIVES:** To compare the results after cochlear implantation achieved by monolingual and bilingual deaf children implanted at our Institution, with the aim of understanding if there are any differences between the two groups and if there is a correlation between the outcomes and some patients' variables.

**METHODS:** The study group was composed by 14 bilingual deaf children and the control group by the same number of monolingual children implanted at our Institution. The control group was obtained by matching to each bilingual child a monolingual one with a similar clinical history regarding age at hearing loss diagnosis, age at first hearing-aids fitting and age at CI procedure. Children received a speech perception and linguistic development evaluation through specific structured tests. The linguistic competence of the patients both in mainstream and native language was determined by the Student Oral Language Observation Matrix (SOLOM).

**RESULTS:** We did not find any statistically significant differences between bilingual and monolingual children in speech perception outcomes. Nevertheless, we obtained different results concerning language skills: bilingual implanted children scored lower at structured language tests, even if the difference was not statistically relevant. Bilingual children scored significantly lower than monolingual ones at the SOLOM scale for linguistic competence.

**CONCLUSION:** The results reported in the present study show better language skills after cochlear implant in Italian monolingual cases than in bilingual ones. This seems to be related to the condition of bilingualism in Italy, mainly related to immigration, and frequently associated with low socio-economic levels, poor competence in the mainstream language and poor social integration, with a suboptimal exposure to the mainstream language and difficulties in following the rehabilitative program.

**Fowler KB, Boppana SB.**

[Congenital cytomegalovirus infection.](#)

*Semin Perinatol.* 2018 Mar 1. pii: S0146-0005(18)30008-9. doi: 10.1053/j.semperi.2018.02.002. [Epub ahead of print]

Each year, thousands of children are born with or develop permanent disabilities such as hearing loss, vision loss, motor and cognitive deficits from congenital CMV infection (cCMV). However, awareness of cCMV and its associated sequelae is very low in pregnant women and healthcare providers. Both targeted and universal approaches to screen newborns for CMV infection are now achievable due to recent scientific advances including the development of a rapid, high-throughput method for detecting CMV in saliva, the efficacy of antiviral treatment in symptomatic infants, and the demonstration of cost effectiveness of CMV screening. Future studies are needed to address gaps in our understanding on the role of non-primary maternal CMV infections, the evaluation of antiviral treatment in asymptomatic infants, and the implementation of prevention strategies for cCMV.

**Funamura JL.**

[Evaluation and management of nonsyndromic congenital hearing loss.](#)

*Curr Opin Otolaryngol Head Neck Surg.* 2017 Oct;25(5):385-389. doi: 10.1097/MOO.0000000000000398.

**PURPOSE OF REVIEW:** Nonsyndromic congenital hearing loss represents the largest proportion of paediatric sensorineural hearing loss. The optimal evaluation and management of affected patients remains clinically challenging. Current controversies in the diagnostic work-up of nonsyndromic congenital hearing loss are presented in this review.

**RECENT FINDINGS:** The improved diagnostic yield of comprehensive genetic testing due to new sequencing technologies is changing the diagnostic for congenital hearing loss. Concerns for both ionizing radiation and general anaesthetic exposure are also driving shifts in imaging modality preferences for infants and toddlers.

**SUMMARY:** A thoughtful systematic, targeted approach taking into consideration the audiologic phenotype of the patient is recommended for the work-up of nonsyndromic congenital hearing loss.

**Ghadersohi S, Ida JB, Bhushan B, Billings KR.**

[Outcomes of tympanoplasty in children with down syndrome.](#)

*Int J Pediatr Otorhinolaryngol.* 2017 Dec;103:36-40. doi: 10.1016/j.ijporl.2017.10.004. Epub 2017 Oct 5.

**INTRODUCTION:** The prevalence of chronic otitis media with effusion (COME), and Eustachian tube dysfunction (ETD) is high in Down syndrome (DS) patients. This often necessitates multiple tympanostomy tube (TT) placements resulting in a higher rate of persistent tympanic membrane (TM) perforation requiring tympanoplasty for repair.

**OBJECTIVES:** To assess risk factors for persistent perforation and outcomes of tympanoplasty in DS patients.

**METHODS:** Retrospective case series of 91 ears in 69 DS patients with TM perforations, who were either observed or underwent tympanoplasty. Clinical features, surgical outcomes, and hearing outcomes were assessed.

**RESULTS:** 91 ears were evaluated. Sixty perforations were observed, and 31 perforations were repaired. The closure rate was 54.8% for primary surgery, and 70.9% after secondary surgical interventions in the Tympanoplasty Group, compared to 33.0% spontaneous closure rate in the Observation Group ( $p < 0.001$ ). The only risk factor for failed tympanoplasty repair was persistent COME/ETD (OR 27.2,  $p = 0.001$ ). In the Observation Group perforations diagnosed at an

older age, with >3 TT insertions, and with persistent COME/ETD were less likely to close spontaneously. Patients undergoing tympanoplasty had worse preoperative pure tone averages than those being observed, but significant improvement in air-bone gaps were noted in the Tympanoplasty Group ( $p = 0.02$ ) post-operatively. Patients were often rehabilitated with hearing aids regardless of intervention (53.3% Observation Group, 48.4% Tympanoplasty Group).

**CONCLUSIONS:** Persistent TM perforation in children with Down syndrome was associated with a history of COME/ETD, and multiple prior TT insertions. Tympanoplasty was successful for repair in most patients who underwent surgical intervention, but residual hearing loss was common.

**Giannattasio A, Di Costanzo P, De Matteis A, Milite P, De Martino D, Bucci L, Augurio MR, Bravaccio C, Ferrara T, Capasso L, Raimondi F.**

[Outcomes of congenital cytomegalovirus disease following maternal primary and non-primary infection.](#)

*J Clin Virol.* 2017 Nov;96:32-36. doi: 10.1016/j.jcv.2017.09.006. Epub 2017 Sep 14.

**BACKGROUND:** Natural history and long term prognosis of congenital cytomegalovirus (CMV) disease according to maternal primary versus non-primary infection are not clearly documented.

**OBJECTIVE:** To investigate clinical, laboratory and neuroimaging features at onset and long term outcome of congenitally CMV-infected patients born to mothers with non-primary infection compared with a group of patients born to mothers with primary infection.

**STUDY DESIGN:** Consecutive neonates born from 2002 to 2015 were considered eligible for the study. Patients underwent clinical, laboratory and instrumental investigation, and audiologic and neurodevelopmental evaluation at diagnosis and during the follow up.

**RESULTS:** A cohort of 158 congenitally infected children was analyzed. Ninety-three were born to mothers with primary CMV infection (Group 1) and 65 to mothers with a non-primary infection (Group 2). Eighty-eight infants had a symptomatic congenital CMV disease: 49 (46.2%) in Group 1 and 39 (60%) in Group 2. Maternal and demographic characteristics of patients of Group 1 and Group 2 were comparable, with the exception of prematurity and a 1-min Apgar score less than 7, which were more frequent in Group 2 compared to Group 1. Prevalence of neuroimaging findings did not significantly differ between the two groups. An impaired neurodevelopmental outcome was observed in 23.7% of patients of Group 1 and in 24.6% cases of Group 2. Similarly, the frequency of hearing loss did not differ between the two groups (25.8% versus 26.2%, respectively).

**CONCLUSIONS:** Neurodevelopmental and hearing sequelae are not affected by the type of maternal CMV infection. Preventing strategies should be developed for both primary and non-primary infections.

**Goh BS, Fadzilah N, Abdullah A, Othman BF, Umat C.**

[Long-term outcomes of Universiti Kebangsaan Malaysia Cochlear Implant Program among pediatric implantees.](#)

*Int J Pediatr Otorhinolaryngol.* 2018 Feb;105:27-32. doi: 10.1016/j.ijporl.2017.11.024. Epub 2017 Nov 24.

**OBJECTIVES:** Cochlear implant (CI) greatly enhances auditory performance as compared to hearing aids and has dramatically affected the educational and communication outcomes for profoundly deaf children. Universiti Kebangsaan Malaysia (UKM) pioneered CI program in 1995 in the South East Asia. We would like to report the long-term outcomes of UKM paediatric cochlear implantation in terms of: the proportion of children who were implanted and still using the device, the children's modes of communication, their educational placements, and their functional auditory/oral performance. We also examined the factors that affected the outcomes measured.

**STUDY DESIGN:** This was a cross sectional observational study. **METHODS:** Two sets of questionnaires were given to 126 parents or primary caregivers of the implantees. The first set of questionnaire contained questions to assess the children's usage of CI, their types of education placement, and their modes of communication. The second set of questionnaire was the Parent's Evaluation Of Aural/Oral Performance of Children (PEACH) to evaluate the children's auditory functionality.

**RESULTS:** Our study showed that among the implantees, 97.6% are still using their CI, 69.8% communicating orally, and 58.5% attending mainstream education. For implantees that use oral communication and attend mainstream education, their mean age of implantation is 38 months. This is significantly lower compared to the mean age of implantation of implantees that use non-oral communication and attend non-mainstream education. Simple logistic regression analysis shows age of implantation reliably predicts implantees ( $N = 126$ ) would communicate using oral communication with odds ratio of 0.974, and also predict mainstream education ( $N = 118$ ) with odds ratio of 0.967. The median score of PEACH rating scale is 87.5% in quiet, and this significantly correlates with an earlier age of implantation ( $r = -0.235$   $p = 0.048$ ).

**CONCLUSIONS:** UKM Cochlear Implant Program has achieved reasonable success among the pediatric implantees, with better outcomes seen in those implanted at the age of less than 4 years old.

**Grandpierre V, Fitzpatrick EM, Na E, Mendonca O.**

[School-aged Children with Mild Bilateral and Unilateral Hearing Loss: Parents' Reflections on Services, Experiences, and Outcomes.](#)

*J Deaf Stud Deaf Educ.* 2018 Apr 1;23(2):140-147. doi: 10.1093/deafed/enx049.

Following the establishment of newborn hearing screening programs, age of identification and length of time before receiving interventions has been reduced for children, including those with milder degrees of hearing loss who were previously not identified until school age. This population of early-identified children requires new support programs for parents. Although literature is emerging on how parents experience the initial years, there is limited information on support needs during early school years. The objectives were to gain insights into parents' experiences with services during the early period of identification until early school years, as well as their perceptions of the consequences of hearing loss on their child's overall development. A qualitative research design informed by Interpretive Description was employed. Individual semi-structured interviews were conducted with 12 parents of children identified with mild hearing loss. Transcripts were analyzed using a constant comparative method. Four themes emerged from the data: early experiences with services and hearing technology, effects of hearing loss on social functioning, effects of hearing loss on language and academics, and experiences in early school years. From parents' perspectives, more support during the early school years is needed to help ensure academic success.

**Grasty MA, Ittenbach RF, Knightly C, Solot CB, Gerdes M, Bernbaum JC, Wernovsky G, Spray TL, Nicolson SC, Clancy RR, Licht DJ, Zackai E, Gaynor JW, Burnham NB.**

[Hearing Loss after Cardiac Surgery in Infancy: An Unintended Consequence of Life-Saving Care.](#)

*J Pediatr.* 2018 Jan;192:144-151.e1. doi: 10.1016/j.jpeds.2017.09.049.

**OBJECTIVES:** To investigate the prevalence of hearing loss after cardiac surgery in infancy, patient and operative factors associated with hearing loss, and the relationship of hearing loss to neurodevelopmental outcomes.

**STUDY DESIGN:** Audiologic and neurodevelopmental evaluations were conducted on 348 children who underwent repair of congenital heart disease at the Children's Hospital of Philadelphia as part of a prospective study evaluating neurodevelopmental outcomes at 4 years of age. A prevalence estimate was calculated based on presence and type of hearing loss. Potential risk factors and the impact of hearing loss on neurodevelopmental outcomes were evaluated.

**RESULTS:** The prevalence of hearing loss was 21.6% (95% CI, 17.2-25.9). The prevalence of conductive hearing loss, sensorineural hearing loss, and indeterminate hearing loss were 12.4% (95% CI, 8.8-16.0), 6.9% (95% CI, 4.1-9.7), and 2.3% (95% CI, 0.6-4.0), respectively. Only 18 of 348 subjects (5.2%) had screened positive for hearing loss before this study and 10 used a hearing aid. After adjusting for patient and operative covariates, younger gestational age, longer postoperative duration of stay, and a confirmed genetic anomaly were associated with hearing loss (all  $P < .01$ ). The presence of hearing loss was associated with worse language, cognition and attention ( $P < .01$ ).

**CONCLUSIONS:** These findings suggest that the prevalence of hearing loss in preschool children after heart surgery in infancy may be 20-fold higher than in the 1% prevalence seen in the general population. Younger gestational age, presence of a genetic anomaly, and longer postoperative duration of stay were associated with hearing loss. Hearing loss was associated with worse neurodevelopmental outcomes.

**Greczka G, Zych M, Wróbel M, Dąbrowski P, Szyfter Harris J, Szyfter W.**

[Analysis of follow-up at the diagnostic level in the Polish Universal Neonatal Hearing Screening Programme.](#)

*J Med Screen.* 2018 Mar;25(1):13-16. doi: 10.1177/0969141317695848. Epub 2017 Mar 21.

**Objectives** Routine analysis showed that between 1 June and 30 November 2014, only 47.6% of expected follow-up visits at the diagnostic level were registered in the Polish Universal Neonatal Hearing Screening Programme central database. We attempted to detect and analyse the reasons for this low percentage.

**Methods** A telephone survey questionnaire was developed for parents whose children had not registered for consultation at the diagnostic level, or had not received a final diagnosis according to the programme database. Questions aimed to verify the database records and compare these with information received from and given to parents. From the 7888 children not registered at the diagnostic level, 3239 records were randomly selected, i.e. 52.4% of those who had been expected to attend.

**Results** Questions were answered by 1950 parents (60.2% of the selected group). Of these, 52.1% ( $n = 734$ ) had attended for diagnostic tests, but this was not recorded in the database. The most common reasons for not attending were the long waiting time for the visit (36.09%), lack of referral to a visit (25.9%) and conscious parent decision (16.35%).

**Conclusion** The telephone survey disclosed omissions in database registration, and that in fact 83.6% of children had attended at the diagnostic level.

**Hao Z, Fu D, Ming Y, Yang J, Huang Q, Lin W, Zhang H, Zhang B, Zhou A, Hu X, Yao C, Dong Y, Ring HZ, Ring BZ.**

[Large scale newborn deafness genetic screening of 142,417 neonates in Wuhan, China.](#)

*PLoS One.* 2018 Apr 10;13(4):e0195740. doi: 10.1371/journal.pone.0195740. eCollection 2018.

Almost one third of the three million people in China suffering severe deafness are children, and 50% of these cases are believed to have genetic components to their etiology. Newborn hearing genetic screening can complement Universal Neonatal Hearing Screening for the diagnosis of congenital hearing loss as well as identifying children at risk for late-on-



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

**He RP, Zheng Y.**

[\[Application of multiple acoustic immittance tests in evaluation of middle ear function in infants\].](#)

*Zhonghua Er Bi Yan Hou Tou Jing Wai Ke Za Zhi.* 2018 Mar 7;53(3):227-231. doi: 10.3760/cma.j.issn.1673-0860.2018.03.013.

Evaluation of middle ear function in infants is the key to distinguish sensorineural hearing loss and conductive hearing loss, and acoustic immittance test is the routine audiological evaluation of middle ear function. Because of the characteristics of middle ear in infants, middle ear examination parameters of adults are not suitable for infants. This article reviewed the current multiple acoustic immittance methods for detecting middle ear function in infants, and summarized the advantages and disadvantages of these methods.

**He X, Li X, Guo Y, Zhao Y, Dong H, Dong J, Zhong L, Shi Z, Zhang Y, Soliman M, Song C, Zhao Z.**

[Newborn Screening of Genetic Mutations in Common Deafness Genes With Bloodspot-Based Gene Chip Array.](#)

*Am J Audiol.* 2018 Mar 8;27(1):57-66. doi: 10.1044/2017\_AJA-17-0042.

**Purpose:** This study screens for deafness gene mutations in newborns in the Northwest China population.

**Method:** The 9 sites of 4 common deafness genes (GJB2, GJB3, SLC26A4, and mt 12S rRNA) were detected by blood-spot-based gene chip array in 2,500 newborns.

**Results:** We detected mutations of the 4 genes in 101 (4.04%) newborns; particularly, 0.20% detected the double mutations. In the Hui population, 4.58% of the newborns tested positive for mutations, whereas 4.01% of Han newborns tested positive for mutations. The detective rates are as follows: 1.44% for GJB2 235delC, 1.08% for SLC26A4 IVS7-2A>G, 0.48% for GJB2 299\_300delAT, 0.28% for SLC26A4 2168A>G, 0.2% for mt 12S rRNA 1555A>G, and 0.16% for GJB3 538C>T. The 31.25% (5/16) of infants with GJB2 235delC, 50% (3/6) with GJB2 299\_300delAT, and 25% (3/12) with SLC26A4 IVS7-2A>G showed abnormal hearing when tested; only 1 double mutation case received the hearing test, and this infant showed abnormality in both ears on the hearing test.

**Conclusions:** High mutation rates in the common deafness genes were detected in newborns in Northwest China. Our study is helpful in understanding the deafness genomic epidemiology and also provides evidence for prenatal and post-natal care as well as policy making on population health in the region.

**Hrnčić N.**

[Identification of risk factors for hearing impairment in newborns: a hospital based study.](#)

*Med Glas (Zenica).* 2018 Feb 1;15(1):29-36. doi: 10.17392/943-18.

**Aim** To identify risk factors for hearing impairment presented in neonates born in Cantonal Hospital Zenica (CHZ) and to estimate their influence on outcome of hearing tests in Newborn Hearing Screening (NHS).

**Methods** Retrospective-prospective study was done at the Department of Gynaecology and Maternity. The NHS was performed with transitory evoked otoacoustic emissions (TEOAE) during a six-month period using "Titan" device (Inter-acoustics, Denmark). The questionnaire was written for the purpose of getting more structured basic information about every newborn and to identify risk factors for hearing impairment. Chi-square test was used to investigate the difference between experimental and control group refer incidence.

**Results** A total of 1217 newborns was screened for hearing impairment of which 259 (21.28%) with one or more known risk factors for hearing impairment. The following risk factors for hearing impairment were identified during the study period: family history of permanent childhood hearing impairment in 42 (3.45%) newborns, prematurity in 39 (3.21%), low APGAR scores in 29 (2.40%), asphyxia in 31 (2.55%), hyperbilirubinemia in 41 (3.37%), admission of ototoxic medication (aminoglycosides) after birth in 155 (12.74%).

**Conclusion** There were many serious risk factors for hearing loss identified in this study. Identification of risk factors for hearing impairment in neonates is necessary because a follow up of the children with risk factors is very important.

**Huang MP, Sheng HB, Ren Y, Li Y, Huang ZW, Wu H.**

[\[Effects of bimodal intervention on the development of auditory and speech ability in infants with unilateral cochlear implantation\].](#)

*Zhonghua Er Bi Yan Hou Tou Jing Wai Ke Za Zhi.* 2018 Mar 7;53(3):203-208. doi: 10.3760/cma.j.isn.1673-0860.2018.03.007.

**Objective:** To explore the effects of bimodal intervention on the development of auditory and speech ability in the infants with unilateral cochlear implantation(CI).

**Methods:** Total 35 bilateral profound sensorineural hearing loss infants with unilateral CI, aged 0.7 to 2.8 years old, were selected. The subjects were divided into two groups: the group with unilateral CI(cochlear implant alone, n= 15), and the bimodal group with CI and contralateral fitting hearing aid(n= 20). Their auditory and speech abilities were estimated at the different time points after switch-on(the 0th, 0.5th, 1st, 3rd, 6th, 12th, 18th, and 24th month, respectively) using Infant Toddler-Meaningful Auditory Integration Scale(IT-MAIS), Meaningful Use of Speech Scale(MUSS), Categories of Auditory Performance(CAP), and Speech Intelligibility Rating(SIR) scores.

**Results:** The IT-MAIS scores of bimodal group after switch-on were higher than unilateral CI group(the 0.5th, 1st, 3rd, 6th, 12th, and 18th month), the statistical significances were identified at the 0.5th, 1st, 3rd, 6th, and 12th month, respectively( $P<0.05$ ). The CAP scores of bimodal group before CI operation and after switch-on(the 0.5th, 1st, 3rd, 6th, 12th, 18th and 24th month)were higher than unilateral CI group, the statistical significances were seen at the 3rd, 6th, 12th, 18th and 24th month after switch-on( $P<0.05$ ). The MUSS scores of bimodal group after switch-on were higher than unilateral CI group(the 1st, 3rd, 6th, 12th, 18th and 24th month), the statistical significances were found at the 12th, 18th and 24th month, respectively( $P<0.05$ ). The SIR scores of bimodal group after switch-on were higher than unilateral CI group(the 3rd, 6th, 12th, 18th and 24th month), and significant differences appeared at the 12th, 18th and 24th month after switch-on( $P<0.05$ ).

**Conclusion:** Bimodal intervention could be helpful to the development of auditory and speech ability of infants.

**Huang S, Huang B, Wang G, Kang DY, Zhang X, Meng X, Dai P.**

[The relationship between the GJB3 c.538C>T variant and hearing phenotype in the Chinese population.](#)

*Int J Pediatr Otorhinolaryngol.* 2017 Nov;102:67-70. doi: 10.1016/j.ijporl.2017.09.001. Epub 2017 Sep 7.

**BACKGROUND:** Mutations in GJB3 were originally shown to underlie an autosomal dominant form of non-syndromic deafness in Chinese patients and the c.538C>T (p.R180\*) variants caused high-frequency hearing loss. But after that, few reports have reported this mutation. This study investigated the relationship between the GJB3 c.538C>T variant and hearing phenotype in Chinese to assist with risk assessment and genetic counseling for hearing loss patients and their families.

**METHOD:** The study enrolled 5700 patients with hearing loss and 4600 normal subjects. Deafness gene mutations were distinguished using a gene chip. The GJB3 c.538C>T variant rate was calculated from the results. **RESULT:** Of the 5700 patients, 23 (0.40%) carried a GJB3 c.538C>T heterozygous variant; of these, 11 patients had other gene (GJB2/SLC26A4) mutations simultaneously. Most patients had moderate to profound hearing loss. All 23 patients were sporadic cases and had no family history of deafness. Of the 4600 normal individuals, 11 (0.24%) had GJB3 c.538C>T heterozygous variant. There was no statistical difference in incidence between the two groups.

**CONCLUSIONS:** Our results showed that the GJB3 c.538C>T variant has a very low incidence in the Chinese population, and there was no clear evidence to support a role of the GJB3 c.538C>T variant in the autosomal dominant form of non-syndromic deafness. Our findings suggested that GJB3 c.538C>T does not contribute to hearing loss, and this conclusion will assist with genetic counseling and risk prediction for deafness related to the GJB3 c.538C>T variant.

**Johnson LC, Toro M, Vishnja E, Berish A, Mills B, Lu Z, Lieberman E.**

[Age and Other Factors Affecting the Outcome of AABR Screening in Neonates.](#)

*Hosp Pediatr.* 2018 Mar;8(3):141-147. doi: 10.1542/hpeds.2017-0060.

**BACKGROUND:** Although the utility of universal newborn hearing screening is undisputed, testing protocols vary. In particular, the impact of the infant's age at the time of automated auditory brainstem response (AABR) screening has not been well studied.

**METHODS:** We conducted a retrospective review of newborn hearing screening data in 6817 low-risk, term and late-preterm newborns at our large, urban, academic medical center for a 1-year period to analyze the impact of age and other factors on the screening failure rate and referral for diagnostic testing.

**RESULTS:** AABR screening failure rates decreased with postnatal age over the first 48 hours; 13.3% failed at <24 hours versus 3.8% at  $\geq 48$  hours ( $P < .0001$ ). Infants who were initially tested at  $\geq 36$  hours failed repeat testing more often than those who were tested at <36 hours (11.5% vs 18.9%;  $P = .03$ ). Other factors that were associated with failure included being a boy and of a race other than white. Sensorineural hearing loss (SNHL) was diagnosed in 18.6% of infants who failed their final screening at  $\geq 48$  hours compared with 2.8% of those whose final screening occurred earlier ( $P = .03$ ). SNHL was more likely in infants who failed their first screening bilaterally (21.2%) than unilaterally (4.4%);  $P = .03$ .

**CONCLUSIONS:** Among healthy newborns, delaying AABR screening in the first 48 hours minimized failure rates. SNHL was 6 times as likely in infants who failed their final screening at  $\geq 48$  hours compared with those who were screened at



<48 hours of age. In our study, we offer guidance for nursery directors and audiologists who determine hearing screening protocols and counsel families about results.

**Conflict of interest statement:** POTENTIAL CONFLICT OF INTEREST: The authors have indicated they have no potential conflicts of interest to disclose.

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

[Parental knowledge and attitudes to childhood hearing loss and hearing services in the Solomon Islands.](#)

*Int J Pediatr Otorhinolaryngol.* 2017 Dec;103:87-92. doi: 10.1016/j.ijporl.2017.10.003. Epub 2017 Oct 5.

**OBJECTIVE:** An understanding of parental knowledge and attitudes towards childhood hearing loss is essential to the successful implementation of audiology services. The present study aimed to investigate parental knowledge and attitudes among parents in the Solomon Islands.

**METHODS AND MATERIALS:** A total of 100 mothers and 50 fathers were administered a questionnaire via semi-structured interviews.

**RESULTS:** Highest parental awareness of aetiology of childhood hearing loss was noted for otitis media (94%), noise exposure (87.3%), and family history (72.7%). The highest parental awareness concerning public health initiatives to reduce/prevent otitis media was noted for routine childhood immunizations (84%) and breast-feeding (76%). Higher rates of knowledge in fathers than in mothers included otitis media ( $p = 0.038$ ), noise exposure ( $p = 0.007$ ), and breast-feeding ( $p = 0.031$ ). Approximately half of parents (56%) agreed that curses may cause hearing loss. Overall parental responses showed positive support for infant hearing screening programs (96%) and school-based ear and hearing health examinations (99.3%).

**CONCLUSIONS:** High levels of parental readiness and support for childhood hearing services in the Solomon Islands was evident. Knowledge of aetiology of childhood hearing loss was highest for otitis media, noise exposure, and family history. Knowledge and attitudes of fathers to childhood hearing loss and hearing services was either the same or better than that of mothers.

**Keilmann A, Friese B, Lässig A, Hoffmann V.**

[Validation of the second version of the LittIEARS® Early Speech Production Questionnaire \(LEESPQ\) in German-speaking children with normal hearing.](#)

*Eur Arch Otorhinolaryngol.* 2018 Apr;275(4):883-888. doi: 10.1007/s00405-018-4889-4. Epub 2018 Feb 7.

**INTRODUCTION:** The introduction of neonatal hearing screening and the increasingly early age at which children can receive a cochlear implant has intensified the need for a validated questionnaire to assess the speech production of children aged 0-18. Such a questionnaire has been created, the LittIEARS® Early Speech Production Questionnaire (LEESPQ). This study aimed to validate a second, revised edition of the LEESPQ.

**METHODS AND MATERIALS:** Questionnaires were returned for 362 children with normal hearing. Completed questionnaires were analysed to determine if the LEESPQ is reliable, prognostically accurate, internally consistent, and if gender or multilingualism affects total scores.

**RESULTS:** Total scores correlated positively with age. The LEESPQ is reliable, accurate, and consistent, and independent of gender or lingual status. A norm curve was created. **DISCUSSION:** This second version of the LEESPQ is a valid tool to assess the speech production development of children with normal hearing, aged 0-18, regardless of their gender. As such, the LEESPQ may be a useful tool to monitor the development of paediatric hearing device users.

**CONCLUSION:** The second version of the LEESPQ is a valid instrument for assessing early speech production of children aged 0-18 months.

**Kim SY, Choi BY, Jung EY, Park H, Yoo HN, Park KH.**

Risk factors for failure in the newborn hearing screen test in very preterm twins.

*Pediatr Neonatol.* 2018 Jan 31. pii: S1875-9572(17)30122-5. doi: 10.1016/j.pedneo.2018.01.014. [Epub ahead of print]

**BACKGROUND:** We aimed to identify prenatal and postnatal risk factors associated with abnormal newborn hearing screen (NHS) results and subsequently confirmed sensorineural hearing loss (SNHL) in preterm twin neonates.

**METHODS:** Electronic medical records of 159 twin neonates who were born alive after  $\leq 32$  weeks were retrospectively reviewed for hearing loss in both ears. Histopathologic examination of the placenta was performed and clinical data, including method of conception and factors specific to twins, were retrieved from a computerized perinatal database. The main outcome measure was failure to pass the NHS test. The generalized estimation equations model was used for twins.

**RESULTS:** Thirty-two neonates (20.1%) had a “refer” result, and, on the confirmation test, permanent SNHL was identified in 4.4% (7/159) of all neonates. Neonates who had a “refer” result on the NHS test were more likely to be of lower birth weight, more likely to have been conceived with the use of in vitro fertilization (IVF), and more likely to have higher rates of intraventricular hemorrhage (IVH) and bronchopulmonary dysplasia. However, monochorionic placentation, death of the co-twin, or being born first was not associated with a “refer” result on the NHS test. Multivariable logistic regression revealed that conception after IVF and the presence of IVH were the only variables to be statistically significantly associated with “refer” on the NHS test. No parameters studied were found to be significantly different between the SNHL and no SNHL groups, probably because of the relatively small number of cases of SNHL. **CONCLUSION:** In

preterm twin newborns, IVF and the presence of IVH were independently associated with an increased risk of abnormal NHS results, whereas the factors specific to twins were not associated with abnormal NHS results.

**Konukseven O, Kaya S, Genc A, Muluk NB, Basar FS, Kirkim G, Tuncer U, Karatas E, Topcu C, Bolat H, Dincol I.**  
[Regional differences of Turkey in risk factors of newborn hearing loss.](#)

*Int J Pediatr Otorhinolaryngol.* 2017 Nov;102:49-55. doi: 10.1016/j.ijporl.2017.08.028. Epub 2017 Sep 1.

**OBJECTIVE:** The aim of this study was to discover Turkish regional differences in the risk factors of newborn hearing loss.

**METHOD:** A multi-centered retrospective design was used. A total of 443 children, registered to the national newborn hearing screening programme, with bilateral hearing loss, from five different regions of Turkey, were evaluated in terms of the types of hearing loss, the degree of hearing loss, the types of risk factors, parental consanguinity, age at diagnosis and age of auditory intervention, respectively.

**RESULTS:** There was no significant difference in the prevalence of hearing loss between regions ( $\chi^2 = 3.210$ ,  $P = 0.523$ ). Symmetric Sensorineural Hearing Loss (SSHL) was the most common type of HL in all regions (91.8%). Profound HL was the most common degree of HL in all regions (46.2%). There were statistically significant differences between regions in terms of types of HL ( $\chi^2 = 14.151$ ,  $P = 0.000$ ). As a total, 323 (72.9%) of subjects did not have any risk factors. There were statistically significant differences between regions in terms of the types of risk factors (pre, peri and post-natal) for SSNHL ( $\chi^2 = 16.095$ ,  $P = 0.000$ ). For all regions, the age of diagnosis was convenient with the JCIH criteria. However the age of hearing aid application was prolonged in some regions. There were statistically significant differences between regions in terms of the age of diagnosis ( $\chi^2 = 93.570$ ,  $P = 0.000$ ) and the age of auditory intervention ( $\chi^2 = 47.323$ ,  $P = 0.000$ ). The confounding effects of gender, age of diagnosis, age of hearing aids applications, HL in the family, types of risk factors for HL on SSNHL were detected. **CONCLUSION:** To reach the goal of a high quality newborn hearing screening, there is a need to develop an evidence-based standard for follow up guideline. In addition, risk factors should be re-evaluated according to regional differences and all regions should take their own precautions according to their evidence based data.

**Kosaner J, Van Dun B, Yigit O, Gultekin M, Bayguzina S.**

Clinically recorded cortical auditory evoked potentials from paediatric cochlear implant users fitted with electrically elicited stapedius reflex thresholds.

*Int J Pediatr Otorhinolaryngol.* 2018 May;108:100-112. doi: 10.1016/j.ijporl.2018.02.033. Epub 2018 Feb 24.

**OBJECTIVES:** This study aimed to objectively evaluate access to soft sounds (55 dB SPL) in paediatric CI users, all wearing MED-EL (Innsbruck, Austria) devices who were fitted with the objective electrically elicited stapedius reflex threshold (eSRT) fitting method, to track their cortical auditory evoked potential (CAEP) presence and latency, and to compare their CAEPs to those of normal-hearing peers.

**METHODS:** Forty-five unilaterally implanted, pre-lingually deafened MED-EL CI users, aged 12-48 months, underwent CAEP testing in the clinic at regular monthly intervals post switch-on. CAEPs were recorded in response to short speech tokens /m/, /g/ and /t/ presented in the free field at 55 dB SPL. Twenty children with normal hearing (NH), similarly aged, underwent CAEP testing once.

**RESULTS:** The proportion of present CAEPs increased and CAEP P1 latencies reduced significantly with post-implantation duration. CAEPs were scored based on their presence and age-appropriate P1 latency. These CAEP scores increased significantly with post-implantation duration. CAEP scores were significantly worse for the /m/ speech token compared to the other two tokens. Compared to the NH group, CAEP scores were significantly smaller for all post-implantation test intervals.

**CONCLUSIONS:** This study provides clinicians with a first step towards typical ranges of CAEP presence, latency, and derived CAEP score over the first months of MED-EL CI use. CAEPs within these typical ranges could validate intervention whereas less than optimum CAEPs could prompt clinicians to seek solutions in a timely manner. CAEPs could clinically validate whether a CI provides adequate access to soft sounds. This approach could form an alternative to behavioural soft sound access verification.

**Lam MYY, Wong ECM, Law CW, Lee HHL, McPherson B.**

[Maternal knowledge and attitudes to universal newborn hearing screening: Reviewing an established program.](#)

*Int J Pediatr Otorhinolaryngol.* 2018 Feb;105:146-153. doi: 10.1016/j.ijporl.2017.12.021. Epub 2017 Dec 23.

**OBJECTIVES:** To facilitate early diagnosis of infants with hearing loss, a universal newborn hearing screening program (UNHS) has been implemented in Hong Kong's public hospitals for over a decade. However, there have been no known studies investigating parent attitudes to, and satisfaction with, UNHS since its launch in Hong Kong. The present study aimed to investigate knowledge of UNHS as well as infant hearing development, and attitudes and satisfaction with UNHS, in Hong Kong mothers with newborns. The study was designed to help evaluate and improve an established UNHS public hospital program, based on the perspectives of service users.

**METHODS:** A researcher-developed questionnaire was administered to 102 mothers whose newborn had received UNHS in the postnatal wards of a large public hospital in Hong Kong. The questionnaire considered parental knowledge

of UNHS and infant hearing development, attitudes and satisfaction toward public hospital UNHS. In the knowledge dimension, parents' preferred time and location for pre-test information delivery, interpretation of screening results, and knowledge of hearing developmental milestones were surveyed. In addition, maternal attitudes to and satisfaction with UNHS screening services, the potential impact of UNHS on parent emotions and parent-baby bonding, attitudes toward informed consent, and willingness to comply with diagnostic assessment referral were also surveyed.

**RESULTS:** Mean participant scores on knowledge of infant hearing development were relatively low ( $M = 2.59/6.0$ ,  $SD = 0.90$ ). Many mothers also underestimated the potential ongoing risks of hearing impairment in babies. Around 80% of mothers thought an infant could not have hearing impairment after passing the screening. In addition, one-third of mothers thought a baby could not later develop hearing impairment in infancy or childhood. In terms of attitudes and satisfaction, participants gave somewhat negative ratings for questions regarding receiving sufficient information about the screening ( $M = 2.90/5.0$ ,  $SD = 1.27$ ), screening procedure ( $M = 2.20/5.0$ ,  $SD = 1.08$ ), and sufficiency of information about results ( $M = 2.87/5.0$ ,  $SD = 1.14$ ). Nonetheless, participants gave positive ratings concerning whether screening could lead to early diagnosis ( $M = 4.61/5.0$ ,  $SD = 0.57$ ) and over 95% of mothers supported UNHS despite potential for false positive results. Mothers reported a high willingness to bring their baby to follow-up assessments if required ( $M = 4.53/5.0$ ,  $SD = 0.56$ ). Participants gave positive ratings for their level of satisfaction with the time and location of first UNHS information provision ( $M = 4.34/5.0$ ,  $SD = 0.80$ ) and the way permission was asked for screening the baby ( $M = 4.04/5.0$ ,  $SD = 0.97$ ) but alternative procedures were also recommended. Most recommendations focused on providing more information about the test and a more detailed explanation of screening results.

**CONCLUSIONS:** The survey results highlighted the need to provide more information to parents about infant hearing development to support home monitoring for signs of hearing loss after UNHS, as well as more detailed explanation and information regarding hearing screening and the implications of results to parents. Regardless of location, surveys of this type may provide valuable support for UNHS program quality assurance.

**Lanzieri TM, Chung W, Leung J, Caviness AC, Baumgardner JL, Blum P, Bialek SR, Demmler-Harrison G; Congenital Cytomegalovirus Longitudinal Study Group.**

[Hearing Trajectory in Children with Congenital Cytomegalovirus Infection.](#)

*Otolaryngol Head Neck Surg.* 2018 Apr;158(4):736-744. doi: 10.1177/0194599818758247. Epub 2018 Mar 20.

**Objectives** To compare hearing trajectories among children with symptomatic and asymptomatic congenital cytomegalovirus infection through age 18 years and to identify brain abnormalities associated with sensorineural hearing loss (SNHL) in asymptomatic case patients.

**Study Design** Longitudinal prospective cohort study.

**Setting** Tertiary medical center.

**Subjects and Methods** The study included 96 case patients (4 symptomatic and 92 asymptomatic) identified through hospital-based newborn cytomegalovirus screening from 1982 to 1992 and 72 symptomatic case patients identified through referrals from 1993 to 2005. We used growth curve modeling to analyze hearing thresholds (0.5-8 kHz) by ear with increasing age and Cox regression to determine abnormal findings on head computed tomography scan associated with SNHL (hearing threshold  $\geq 25$  dB in any audiometric frequency) among asymptomatic case patients.

**Results** Fifty-six (74%) symptomatic and 20 (22%) asymptomatic case patients had SNHL: congenital/early-onset SNHL was diagnosed in 78 (51%) and 10 (5%) ears, respectively, and delayed-onset SNHL in 25 (17%) and 20 (11%) ears; 49 (32%) and 154 (84%) ears had normal hearing. In affected ears, all frequency-specific hearing thresholds worsened with age. Congenital/early-onset SNHL was significantly worse (severe-profound range,  $>70$  dB) than delayed-onset SNHL (mild-moderate range, 26-55 db). Frequency-specific hearing thresholds were significantly different between symptomatic and asymptomatic case patients at 0.5 to 1 kHz but not at higher frequencies (2-8 kHz). Among asymptomatic case patients, white matter lucency was significantly associated with SNHL by age 5 years (hazard ratio, 4.4; 95% CI, 1.3-15.6).

**Conclusion** Congenital/early-onset SNHL frequently resulted in severe to profound loss in symptomatic and asymptomatic case patients. White matter lucency in asymptomatic case patients was significantly associated with SNHL by age 5 years.

**Lim CH, Lim JH, Kim D, Choi HS, Lee DH, Kim DK.**

[Bony cochlear nerve canal stenosis in pediatric unilateral sensorineural hearing loss.](#)

*Int J Pediatr Otorhinolaryngol.* 2018 Mar;106:72-74. doi: 10.1016/j.ijporl.2018.01.015. Epub 2018 Feb 2.

**OBJECTIVES:** This study was performed to evaluate the frequency of bony cochlear nerve canal (BCNC) stenosis and its clinical significance in pediatric patients with unilateral sensorineural hearing loss (SNHL) of unknown etiology.

**MATERIALS AND METHODS:** We analyzed the medical records and temporal bone computed tomography (CT) results of patients less than 13 years of age with a diagnosis of unilateral SNHL of unknown etiology between July 2007 and July 2017. We compared the BCNC diameter between both sides and analyzed the age at diagnosis, degree of hearing loss, and accompanying inner ear anomalies.

**RESULTS:** In 42 patients, the mean age at diagnosis was  $7.4 \pm 3.6$  years, and the average hearing level in the affected ear was  $87.9 \pm 20.0$  dB HL (decibels hearing level). The average diameter of the BCNC was  $1.22 \pm 0.75$  mm on the

affected side and  $1.96 \pm 0.52$  mm on the normal side. The most suitable criterion for BCNC stenosis appeared to be a diameter of 1.2 mm by the recursive partitioning procedure. With application of this criterion, the rate of BCNC stenosis was significantly greater on the affected side than on the normal side (52.4% vs. 4.8%, respectively;  $P < 0.05$ ). A narrow internal acoustic canal was found in two patients, and vestibular and cochlear anomalies were found in three patients each.

**CONCLUSIONS:** Our results suggest that it is reasonable to set a diameter of 1.2 mm as a cutoff for BCNC stenosis, and also that BCNC stenosis is a common cause of unilateral SNHL of unknown etiology in childhood.

**Lopez AS, Lanzieri TM, Claussen AH, Vinson SS, Turcich MR, Iovino IR, Voigt RG, Caviness AC, Miller JA, Williamson WD, Hales CM, Bialek SR, Demmler-Harrison G; Congenital Cytomegalovirus Longitudinal Study Group.** [Intelligence and Academic Achievement With Asymptomatic Congenital Cytomegalovirus Infection.](#) *Pediatrics.* 2017 Nov;140(5). pii: e20171517. doi: 10.1542/peds.2017-1517.

**OBJECTIVES:** To examine intelligence, language, and academic achievement through 18 years of age among children with congenital cytomegalovirus infection identified through hospital-based newborn screening who were asymptomatic at birth compared with uninfected infants.

**METHODS:** We used growth curve modeling to analyze trends in IQ (full-scale, verbal, and nonverbal intelligence), receptive and expressive vocabulary, and academic achievement in math and reading. Separate models were fit for each outcome, modeling the change in overall scores with increasing age for patients with normal hearing ( $n = 78$ ) or with sensorineural hearing loss (SNHL) diagnosed by 2 years of age ( $n = 11$ ) and controls ( $n = 40$ ).

**RESULTS:** Patients with SNHL had full-scale intelligence and receptive vocabulary scores that were 7.0 and 13.1 points lower, respectively, compared with controls, but no significant differences were noted in these scores among patients with normal hearing and controls. No significant differences were noted in scores for verbal and nonverbal intelligence, expressive vocabulary, and academic achievement in math and reading among patients with normal hearing or with SNHL and controls.

**CONCLUSIONS:** Infants with asymptomatic congenital cytomegalovirus infection identified through newborn screening with normal hearing by age 2 years do not appear to have differences in IQ, vocabulary or academic achievement scores during childhood, or adolescence compared with uninfected children.

Copyright © 2017 by the American Academy of Pediatrics.

DOI: 10.1542/peds.2017-1517 PMID: 29066580 [Indexed for MEDLINE]

**Conflict of interest statement: POTENTIAL CONFLICT OF INTEREST:** Dr Demmler-Harrison's institution has received funding from Merck Sharpe & Dohme Corporation since July 2016 to assist with salary support for further analysis on long-term outcomes of congenital cytomegalovirus infection not included in this report; the other authors have indicated they have no potential conflicts of interest to disclose.

**Lu X, Qin Z.**

[Auditory and language development in Mandarin-speaking children after cochlear implantation.](#)

*Int J Pediatr Otorhinolaryngol.* 2018 Apr;107:183-189. doi: 10.1016/j.ijporl.2018.02.006. Epub 2018 Feb 7.

**OBJECTIVE:** To evaluate early auditory performance, speech perception and language skills in Mandarin-speaking pre-lingual deaf children in the first two years after they received a cochlear implant (CI) and analyse the effects of possible associated factors.

**METHODS:** The Infant-Toddler Meaningful Auditory Integration Scale (ITMAIS)/Meaningful Auditory Integration Scale (MAIS), Mandarin Early Speech Perception (MESP) test and Putonghua Communicative Development Inventory (PCDI) were used to assess auditory and language outcomes in 132 Mandarin-speaking children pre- and post-implantation.

**RESULTS:** Children with CIs exhibited an ITMAIS/MAIS and PCDI developmental trajectory similar to that of children with normal hearing. The increased number of participants who achieved MESP categories 1-6 at each test interval showed a significant improvement in speech perception by paediatric CI recipients. Age at implantation and socioeconomic status were consistently associated with both auditory and language outcomes in the first two years post-implantation.

**CONCLUSION:** Mandarin-speaking children with CIs exhibit significant improvements in early auditory and language development. Though these improvements followed the normative developmental trajectories, they still exhibited a gap compared with normative values. Earlier implantation and higher socioeconomic status are consistent predictors of greater auditory and language skills in the early stage.

**Maas RR, Iwanicka-Pronicka K, Kalkan Ucar S, Alhaddad B, AlSayed M, Al-Owain MA, Al-Zaidan HI, Balasubramaniam S, Barić I, Bubshait DK, Burlina A, Christodoulou J, Chung WK, Colombo R, Darin N, Freisinger, Garcia Silva MT, Grunewald S, Haack TB, van Hasselt PM, Hikmat O, Hörster F, Isohanni P, Ramzan K, Kovacs-Nagy R, Krumina Z, Martin-Hernandez E, Mayr JA, McClean P, De Meirleir L, Naess K, Ngu LH, Pajdowska M, Rahman S, Riordan G, Riley L, Roeben B, Rutsch F, Santer R, Schiff M, Seders M, Sequeira S, Sperl W, Staufner C, Synofzik M, Taylor RW, Trubicka J, Tsiakas K, Unal, Wassmer E, Wedatilake Y, Wolff T, Prokisch H, Morava E, Pronicka E, Wevers RA, de Brouwer AP, Wortmann SB.**



Progressive deafness-dystonia due to SERAC1 mutations: A study of 67 cases.

*Ann Neurol.* 2017 Dec;82(6):1004-1015. doi: 10.1002/ana.25110.

**OBJECTIVE:** 3-Methylglutaconic aciduria, dystonia-deafness, hepatopathy, encephalopathy, Leigh-like syndrome (MEGDHEL) syndrome is caused by biallelic variants in SERAC1.

**METHODS:** This multicenter study addressed the course of disease for each organ system. Metabolic, neuroradiological, and genetic findings are reported.

**RESULTS:** Sixty-seven individuals (39 previously unreported) from 59 families were included (age range=5 days-33.4 years, median age=9 years). A total of 41 different SERAC1 variants were identified, including 20 that have not been reported before. With the exception of 2 families with a milder phenotype, all affected individuals showed a strikingly homogeneous phenotype and time course. Severe, reversible neonatal liver dysfunction and hypoglycemia were seen in >40% of all cases. Starting at a median age of 6 months, muscular hypotonia (91%) was seen, followed by progressive spasticity (82%, median onset=15 months) and dystonia (82%, 18 months). The majority of affected individuals never learned to walk (68%). Seventy-nine percent suffered hearing loss, 58% never learned to speak, and nearly all had significant intellectual disability (88%). Magnetic resonance imaging features were accordingly homogenous, with bilateral basal ganglia involvement (98%); the characteristic “putaminal eye” was seen in 53%. The urinary marker 3-methylglutaconic aciduria was present in virtually all patients (98%). Supportive treatment focused on spasticity and drooling, and was effective in the individuals treated; hearing aids or cochlear implants did not improve communication skills.

**INTERPRETATION:** MEGDHEL syndrome is a progressive deafness-dystonia syndrome with frequent and reversible neonatal liver involvement and a strikingly homogenous course of disease. *Ann Neurol* 2017;82:1004-1015.

**Mallen JR, Hunter JB, Auerbach C, Wexler L, Vambutas A.**

[Characterization of newborn hearing screening failures in multigestational births.](#)

*Int J Pediatr Otorhinolaryngol.* 2018 Feb;105:158-162. doi: 10.1016/j.ijporl.2017.12.017. Epub 2017 Dec 15.

**OBJECTIVE:** To define the rate and characterize the type of newborn hearing screening failures in multigestational births.

**METHODS:** Retrospective chart review of all multigestational births that occurred in a 10-year period (2002-2012) in which at least one newborn failed newborn hearing screening at two tertiary care hospitals in the Northwell Health System.

**RESULTS:** Out of 125,405 total births, we identified 2961 multigestational births, of which 59 (2.0%) newborns failed newborn hearing screening. None of their 66 twin/triplet siblings failed their newborn hearing screens. Of 43 newborns that returned for follow-up, 56.0% (24/43) had confirmed hearing loss, resulting in an overall rate of 0.81% in all multigestational newborns with hearing loss. Of 19 infants that passed repeat testing, two were judged to need myringotomy tube placement. Twenty-four infants had a confirmed hearing loss, 11 of which had sensorineural hearing loss (0.37%), and 13 with a conductive or mixed hearing loss (0.44%).

**CONCLUSIONS:** We identified a greater than expected risk of conductive hearing loss, not attributable to otitis media, than sensorineural hearing loss in this population. These observations are consistent with the increased risk of birth defects in multigestational births.

**Matsushima K, Nakano A, Arimoto Y, Mutai H, Yamazawa K, Murayama K, Matsunaga T.**

[High-level heteroplasmy for the m.7445A>G mitochondrial DNA mutation can cause progressive sensorineural hearing loss in infancy.](#)

*Int J Pediatr Otorhinolaryngol.* 2018 May;108:125-131. doi: 10.1016/j.ijporl.2018.02.037. Epub 2018 Feb 27.

**OBJECTIVE:** Hearing loss caused by mutation of mitochondrial DNA typically develops in late childhood or early adulthood, but rarely in infancy. We report the investigation of a patient to determine the cause of his early onset hearing loss.

**MATERIALS AND METHODS:** The proband was a boy aged 1 year and 2 months at presentation. Newborn hearing screening test by automated auditory brainstem response generated “pass” results for both ears. His reaction to sound deteriorated by 9 months. Average pure tone threshold at 0.5, 1, and 2 kHz was 55 dB by conditioned orientation response audiometry. His father had congenital hearing loss, and his mother had progressive hearing loss since childhood. Invader assays and Sanger sequencing were performed to investigate genetic causes of the hearing loss in the proband, and heteroplasmy was assessed by PCR-restriction fragment length polymorphism, Sanger sequencing, and pyrosequencing. Additionally, mitochondrial function was evaluated by measurement of the oxygen consumption rate of patient skin fibroblasts.

**RESULTS:** An m.7445A > G mitochondrial DNA mutation and a heterozygous c.235delC (p.L79Cfs\*3) mutation of GJB2 were detected in the proband. His mother carried the m.7445A > G mitochondrial DNA mutation, and his father was a compound heterozygote for GJB2 mutations (c.[235delC]; [134G > A; 408C > A]). Tissue samples from both the proband and his mother exhibited a high degree of heteroplasmy. Fibroblasts from the proband exhibited markedly reduced oxygen consumption rates. These data indicate that the proband had impaired mitochondrial function, resulting in hearing loss.

**CONCLUSION:** This research demonstrates that hearing loss in a proband who presented in infancy and that of his mother resulted from a high level of heteroplasmy for the m.7445A > G mitochondrial DNA mutation, indicating that this alteration can cause hearing loss in infancy.



**McKinney S.**

[Cochlear implantation in children under 12 months of age.](#)

*Curr Opin Otolaryngol Head Neck Surg.* 2017 Oct;25(5):400-404. doi: 10.1097/MOO.0000000000000400.

**PURPOSE OF REVIEW:** Children with congenital hearing loss are being identified earlier, leading to earlier intervention. Current US Food and Drug Administration (FDA) criteria states a child must be 12 months or older for cochlear implantation. The purpose of this article is to review recent publications regarding the benefits of implanting infants under 12 months of age. Topics include: safety and efficacy of surgery, speech and language acquisition outcomes, audiologic components, and limitations.

**RECENT FINDINGS:** Since the early 1990s, the candidacy criteria evolved drastically. However, the FDA criteria for cochlear implantation in children has remained at 12 months of age or older since 2000. Recent research indicates implanting below 12 months of age a safe and effective procedure. Speech and language outcomes showed better speech and language advantages. In addition, infants implanted earlier showed normal auditory skills as early as 3 months post cochlear implant activation. This article will also address recent findings on the limitations of earlier implantation.

**SUMMARY:** Recent research demonstrates positive outcomes in children implanted under 12 months of age. Developing research on earlier implantation could lead to a change in the current FDA criteria allowing infants to reach their speech and hearing potential faster.

**Montino S, Agostinelli A, Trevisi P, Martini A, Ghiselli S.**

[Check-list for the assessment of functional impairment in children with congenital aural atresia.](#)

*Int J Pediatr Otorhinolaryngol.* 2017 Nov;102:174-179. doi: 10.1016/j.ijporl.2017.09.024. Epub 2017 Sep 28.

**OBJECTIVES:** Congenital Aural Atresia (CAA) is a deformity of the external ear and it is commonly associated with malformations of middle and inner ear and, in some cases, with other facial deformities. Very few assessment measures exist for evaluating the functional impairment in children with CAA. Purpose of this study is to introduce and describe an assessment Checklist, (nominated FOS Checklist) that covers feeding abilities (F), oralmotor skills (O), communication/ language development (S) in children with CAA. FOS wants to offer a range of assessment providing a profile of the child in comparison to hearing peers and it aims to make clinicians able to identify additional problems and areas of difficulties as well as specific abilities and skills. Secondary, we want to investigate the presence of correlations between disorders and side of CAA.

**METHODS:** a new Checklist (FOS Checklist) was administered to 68 children with CAA. **RESULTS:** Feeding abilities are age-adequate in 94,3% of all patients. 54,4% of all patients are in need for further assessment of their oral-motor skills; delays in language development were found in 44,1% of cases. Orofacial development delays have been observed in 57.2% of subjects among the bilateral CAA group, in 53.9% among the right CAA group and in 53.4% among the left CAA group. Patients referred for further language evaluation were 42,9% in the bilateral CAA group, 33.3% in the right CAA group and 33.3% in the left CAA group. According to the  $\chi^2$  analysis, referral for further assessment is independent from side of aural atresia.

**CONCLUSIONS:** Subjects with bilateral CAA are more likely to be referred for further assessment, both for oral motor aspects and for speech perception and language development. However, there is not a significant statistical difference between the performances of children with bilateral or unilateral CAA. FOS Checklist is simple, reliable and time effective and can be used in everyday clinical practice. FOS enable clinicians to identify additional problems and areas of difficulties as well as specific abilities and skills; moreover, FOS allows to determine appropriate referrals and intervention strategies.

**Myne S, Kennedy V.**

[Hyperacusis in children: A clinical profile.](#)

*Int J Pediatr Otorhinolaryngol.* 2018 Apr;107:80-85. doi: 10.1016/j.ijporl.2018.01.004. Epub 2018 Jan 31.

**OBJECTIVE:** Hyperacusis is commonly seen in clinical paediatric practice and can be distressing for the children and their families. This paper looks at the clinical profile of children seen for hyperacusis in a paediatric audiology service and reviews the possible underlying mechanisms.

**METHODS:** Retrospective study of case notes of 61 children with troublesome hyperacusis seen in the paediatric audiology service, looking at their clinical presentation and presence of other medical conditions.

**RESULTS:** Hyperacusis was the main presenting complaint in more than half of the cases ( $n = 31$ , 51%). The commonest age at presentation with this problem was 3-4 years ( $n = 33$ , 54%). Hearing was normal in the majority of these children ( $n = 41$ , 67%). An active middle ear problem was observed in nearly half ( $n = 29$ , 48%) of all the children, of which glue ear (otitis media with effusion, OME) was the commonest. Presence of a neurodevelopmental condition was found in almost half ( $n = 28$ , 46%) of these patients of which autistic spectrum disorder was the commonest (8/61, 13%). In nearly one-fourth of the children (23%), presence of both middle ear problems and neurodevelopmental was noted. Tinnitus was an accompanying symptom reported in 11% of all the patients.

**CONCLUSION:** Hyperacusis may commonly present at a very young age. Awareness of different clinical presentations, presence of other medical conditions and possible underlying pathomechanisms in children with hyperacusis can be helpful for clinicians in informing prognosis, counselling and in individualising management plan.

**Norrix LW, Velenovsky D.**

[Clinicians' Guide to Obtaining a Valid Auditory Brainstem Response to Determine Hearing Status: Signal, Noise, and Cross-Checks.](#)

*Am J Audiol.* 2018 Mar 8;27(1):25-36. doi: 10.1044/2017\_AJA-17-0074.

**Purpose:** The auditory brainstem response (ABR) is a powerful tool for making clinical decisions about the presence, degree, and type of hearing loss in individuals in whom behavioral hearing thresholds cannot be obtained or are not reliable. Although the test is objective, interpretation of the results is subjective.

**Method:** This review provides information about evidence-based criteria, suggested by the 2013 Newborn Hearing Screening Program guidelines, and the use of cross-check methods for making valid interpretations about hearing status from ABR recordings.

**Results:** The use of an appropriate display scale setting, templates of expected response properties, and objective criteria to estimate the residual noise, signal level, and signal-to-noise ratio will provide quality data for determining ABR thresholds. Cross-checks (e.g., immittance measures, otoacoustic emissions testing, functional indications of a child's hearing) are also needed to accurately interpret the ABR.

**Conclusions:** Using evidence-based ABR signal detection criteria and considering the results within the context of other physiologic tests and assessments of hearing function will improve the clinician's accuracy for detecting hearing loss and, when present, the degree of hearing loss. Diagnostic accuracy will ensure that appropriate remediation is initiated and that children or infants with normal hearing are not subjected to unnecessary intervention.

**Oliveira LS, Didoné DD, Durante AS.**

[Automated cortical auditory evoked potentials threshold estimation in neonates.](#)

*Braz J Otorhinolaryngol.* 2018 Feb 2. pii: S1808-8694(18)30002-8. doi: 10.1016/j.bjorl.2018.01.001. [Epub ahead of print]

**INTRODUCTION:** The evaluation of Cortical Auditory Evoked Potential has been the focus of scientific studies in infants. Some authors have reported that automated response detection is effective in exploring these potentials in infants, but few have reported their efficacy in the search for thresholds.

**OBJECTIVE:** To analyze the latency, amplitude and thresholds of Cortical Auditory Evoked Potential using an automatic response detection device in a neonatal population.

**METHODS:** This is a cross-sectional, observational study. Cortical Auditory Evoked Potentials were recorded in response to pure-tone stimuli of the frequencies 500, 1000, 2000 and 4000Hz presented in an intensity range between 0 and 80dB HL using a single channel recording. P1 was performed in an exclusively automated fashion, using Hotelling's T2 statistical test. The latency and amplitude were obtained manually by three examiners. The study comprised 39 neonates up to 28 days old of both sexes with presence of otoacoustic emissions and no risk factors for hearing loss.

**RESULTS:** With the protocol used, Cortical Auditory Evoked Potential responses were detected in all subjects at high intensity and thresholds. The mean thresholds were 24.8±10.4dB NA, 25±9.0dB NA, 28±7.8dB NA and 29.4±6.6dB HL for 500, 1000, 2000 and 4000Hz, respectively.

**CONCLUSION:** Reliable responses were obtained in the assessment of cortical auditory potentials in the neonates assessed with a device for automatic response detection.

**Poonual W, Navacharoen N, Kangsanarak J, Namwongprom S, Saokaew S.**

Hearing loss screening tool (COBRA score) for newborns in primary care setting.

*Korean J Pediatr.* 2017 Nov;60(11):353-358. doi: 10.3345/kjp.2017.60.11.353. Epub 2017 Nov 27.

**Purpose:** To develop and evaluate a simple screening tool to assess hearing loss in newborns. A derived score was compared with the standard clinical practice tool.

**Methods:** This cohort study was designed to screen the hearing of newborns using transiently evoked otoacoustic emission and auditory brain stem response, and to determine the risk factors associated with hearing loss of newborns in 3 tertiary hospitals in Northern Thailand. Data were prospectively collected from November 1, 2010 to May 31, 2012. To develop the risk score, clinical-risk indicators were measured by Poisson risk regression. The regression coefficients were transformed into item scores dividing each regression-coefficient with the smallest coefficient in the model, rounding the number to its nearest integer, and adding up to a total score.

**Results:** Five clinical risk factors (Craniofacial anomaly, Ototoxicity, Birth weight, family history [Relative] of congenital sensorineural hearing loss, and Apgar score) were included in our COBRA score. The screening tool detected, by area under the receiver operating characteristic curve, more than 80% of existing hearing loss. The positive-likelihood ratio of hearing loss in patients with scores of 4, 6, and 8 were 25.21 (95% confidence interval [CI], 14.69-43.26), 58.52 (95% CI, 36.26-94.44), and 51.56 (95% CI, 33.74-78.82), respectively. This result was similar to the standard tool (The Joint Committee on Infant Hearing) of 26.72 (95% CI, 20.59-34.66). **Conclusion:** A simple screening tool of five predictors provides good prediction indices for newborn hearing loss, which may motivate parents to bring children for further appropriate testing and investigations.

**Conflict of interest statement:** Conflicts of interest: No potential conflict of interest relevant to this article was reported.

**Puhakka L, Lappalainen M, Lönnqvist T, Niemensivu R, Lindahl P, Nieminen T, Seuri R, Nupponen I, Pati S, Boppana S, Saxen H.**

[The Burden of Congenital Cytomegalovirus Infection: A Prospective Cohort Study of 20 000 Infants in Finland.](#)

*J Pediatric Infect Dis Soc.* 2018 Mar 15. doi: 10.1093/jpids/piy027. [Epub ahead of print]

**Background:** Congenital cytomegalovirus (cCMV) infection is the most common congenital infection and causes significant morbidity. This study was undertaken to evaluate the benefits of screening newborns for cCMV and to understand the cCMV disease burden in Finland.

**Methods:** Infants born in Helsinki area hospitals were screened for CMV by testing their saliva with a real-time polymerase chain reaction assay. The CMV-positive infants and matched controls were monitored to determine their neurodevelopmental, audiological, and ophthalmological outcomes at 18 months of age. Griffiths Mental Development Scales, otoacoustic emission and sound field audiometry, and ophthalmologic examination were performed.

**Results:** Of the 19868 infants screened, 40 had confirmed cCMV infection (prevalence, 2 in 1000 [95% confidence interval, 1.4-2.6 in 1000]). Four (10%) infants had symptomatic cCMV. Griffiths general quotients did not differ significantly between the CMV-positive (mean, 101.0) and control (mean, 101.6) infants ( $P = .557$ ), nor did quotients for any of the Griffiths subscales (locomotion, personal-social, hearing and language, eye and hand, performance) ( $P = .173-.721$ ). Four of 54 CMV-positive ears and 6 of 80 CMV-negative ears failed otoacoustic emission testing ( $P = 1.000$ ). The mean minimal response levels over the frequencies 500 Hz to 4 kHz in the sound field audiometry did not differ between CMV-positive (mean, 34.31-dB hearing level) and control (mean, 32.73-dB hearing level) infants ( $P = .338$ ). No CMV-related ophthalmologic findings were observed.

**Conclusions:** The prevalence of cCMV was low, and outcomes at 18 months of age did not differ between the infected infants and healthy control infants. With such a low burden in Finland, universal newborn screening for cCMV seems unwarranted.

**Pérez-Martín J, Artaso MA, Díez FJ.**

[Cost-effectiveness of pediatric bilateral cochlear implantation in Spain.](#)

*Laryngoscope.* 2017 Dec;127(12):2866-2872. doi: 10.1002/lary.26765. Epub 2017 Aug 4.

**OBJECTIVES/HYPOTHESIS:** To determine the incremental cost-effectiveness of bilateral versus unilateral cochlear implantation for 1-year-old children suffering from bilateral sensorineural severe to profound hearing loss from the perspective of the Spanish public health system.

**STUDY DESIGN:** Cost-utility analysis.

**METHODS:** We conducted a general-population survey to estimate the quality-of-life increase contributed by the second implant. We built a Markov influence diagram and evaluated it for a life-long time horizon with a 3% discount rate in the base case.

**RESULTS:** The incremental cost-effectiveness ratio of simultaneous bilateral implantation with respect to unilateral implantation for 1-year-old children with severe to profound deafness is €10,323 per quality-adjusted life year (QALY). For sequential bilateral implantation, it rises to €11,733/QALY. Both options are cost-effective for the Spanish health system, whose willingness to pay is estimated at around €30,000/QALY. The probabilistic sensitivity analysis shows that the probability of bilateral implantation being cost-effective reaches 100% for that cost-effectiveness threshold.

**CONCLUSIONS:** Bilateral implantation is clearly cost-effective for the population considered. If possible, it should be done simultaneously (i.e., in one surgical operation), because it is as safe and effective as sequential implantation, and saves costs for the system and for users and their families. Sequential implantation is also cost-effective for children who have received the first implant recently, but it is difficult to determine when it ceases to be so because of the lack of detailed data. These results are specific for Spain, but the model can easily be adapted to other countries.

**LEVEL OF EVIDENCE:** 2C. *Laryngoscope*, 127:2866-2872, 2017.

**Ravi R, Gunjawate DR, Yerraguntla K, Rajashekhar B.**

*Int J Pediatr Otorhinolaryngol.* 2018 Jan;104:138-144. doi: 10.1016/j.ijporl.2017.11.004. Epub 2017 Nov 9.

[Systematic review of knowledge of, attitudes towards, and practices for newborn hearing screening among healthcare professionals.](#)

**INTRODUCTION:** The success of newborn hearing screening programs lies in the timely identification, diagnosis, and management of children with hearing loss accomplished via a multidisciplinary newborn hearing screening (NHS) team. The team is typically comprised of various healthcare professionals who act as decision makers as well as facilitators for different stages in the screening process. Team members' knowledge of, attitudes towards, and practices for early hearing detection and intervention programs are critical for success and prevention of loss to follow up. In this context, it becomes crucial to understand their knowledge of, attitudes towards, and practices for towards newborn hearing screening.

**METHODS:** A systematic review was conducted on the following databases; PubMed/Medline, Cumulative Index to Nursing and Allied Health Literature (CINAHL), Scopus, Web of Science, Science Direct and Cochrane Library. This search was carried out using various keywords such as practitioners, newborn hearing screening, knowledge, attitudes, and practices in different combinations. The review was conducted based on Preferred Reporting Items for Systematic

Reviews and Meta-analyses statement guidelines.

**RESULTS:** A total of 271 hits were obtained of which 20 articles were found suitable for inclusion in the final review. Overall, similar results were found regarding team members' knowledge of NHS programs, regardless of country of origin. Similarly, attitudes toward NHS programs were positive. Team members' experiences with NHS programs varied from country-to-country and across healthcare professionals. Results consistently showed gaps in team members' knowledge suggesting the need for outreach and professional education programs on NHS.

**CONCLUSION:** NHS teams members from different countries, healthcare systems, and early hearing detection and intervention programs show gaps in critical knowledge warranting outreach and educational programs.

**Rawlinson WD, Palasanthiran P, Hall B, Al Yazidi L, Cannon MJ, Cottier C, van Zuylen WJ, Wilkinson M.**

[Neonates with congenital Cytomegalovirus and hearing loss identified via the universal newborn hearing screening program.](#)

*J Clin Virol.* 2018 May;102:110-115. doi: 10.1016/j.jcv.2018.03.006. Epub 2018 Mar 15.

**BACKGROUND:** Congenital cytomegalovirus (CMV) is the most common non-genetic cause of sensorineural hearing loss. Currently, there are no universal CMV screening programs for newborns or routine CMV testing of neonates with hearing loss in Australia, or elsewhere. **OBJECTIVES:** This study was undertaken to determine the prevalence of congenital CMV infection in infants with hearing loss identified using routine resources via the Australian universal neonatal hearing screening (UNHS) program.

**STUDY DESIGN:** Infants who failed UNHS, referred for audiological testing and found to have permanent hearing loss were screened for CMV via PCR of urine and saliva. Congenital CMV was diagnosed if CMV was detected in infants  $\leq 30$  days of age, or using retrospective testing on stored newborn screening cards, retrospective testing, or using clinical criteria if  $>30$  days of age. The cohort was analyzed for time of testing and prevalence of congenital CMV determined. **RESULTS:** The Audiology Department reviewed 1669 infants who failed UNHS between 2009 and 2016. Thirty percent (502/1669) had permanent hearing loss confirmed, of whom 336/502 were offered CMV testing. A definite ( $n = 11$ ) or probable ( $n = 8$ ) diagnosis of congenital CMV occurred in 19/323 (5.9%), of whom definite diagnoses were made in 4/19 on tests positive prior to 21 days of life, in 5/19 who were positive on neonatal blood screening card (NBSC) testing, in 2/19 who were positive on placental testing. In 8/19 probable diagnoses were made based on positive testing between ages 23-42 days and a consistent clinical syndrome in the absence of another cause for hearing loss after genetic and other testing. CMV testing mirrored the timing of audiological testing, with ~40% completing audiology and CMV testing by 21 days, and 64% by 30 days.

**CONCLUSION:** This program, utilizing existing clinical services identified probable congenital CMV in ~6% of a large cohort failing UNHS with permanent hearing loss, of whom more than half were definite diagnoses. No additional assets were required to those already existing in this tertiary referral pediatric centre, whilst providing useful and timely data for clinical and audiological management.

**Razza S, Zaccone M, Meli A, Cristofari E.**

[Evaluation of speech reception threshold in noise in young Cochlear™ Nucleus® system 6 implant recipients using two different digital remote microphone technologies and a speech enhancement sound processing algorithm.](#)

*Int J Pediatr Otorhinolaryngol.* 2017 Dec;103:71-75. doi: 10.1016/j.ijporl.2017.10.002. Epub 2017 Oct 5.

**OBJECTIVE:** Children affected by hearing loss can experience difficulties in challenging and noisy environments even when deafness is corrected by Cochlear implant (CI) devices. These patients have a selective attention deficit in multiple listening conditions. At present, the most effective ways to improve the performance of speech recognition in noise consists of providing CI processors with noise reduction algorithms and of providing patients with bilateral CIs. The aim of this study was to compare speech performances in noise, across increasing noise levels, in CI recipients using two kinds of wireless remote-microphone radio systems that use digital radio frequency transmission: the Roger Inspiro accessory and the Cochlear Wireless Mini Microphone accessory.

**METHODS:** Eleven Nucleus Cochlear CP910 CI young user subjects were studied. The signal/noise ratio, at a speech reception threshold (SRT) value of 50%, was measured in different conditions for each patient: with CI only, with the Roger or with the MiniMic accessory. The effect of the application of the SNR-noise reduction algorithm in each of these conditions was also assessed. The tests were performed with the subject positioned in front of the main speaker, at a distance of 2.5 m. Another two speakers were positioned at 3.50 m. The main speaker at 65 dB issued disyllabic words. Babble noise signal was delivered through the other speakers, with variable intensity.

**RESULTS:** The use of both wireless remote microphones improved the SRT results. Both systems improved gain of speech performances. The gain was higher with the Mini Mic system (SRT = -4.76) than the Roger system (SRT = -3.01). The addition of the NR algorithm did not statistically further improve the results.

**CONCLUSION:** There is significant improvement in speech recognition results with both wireless digital remote microphone accessories, in particular with the Mini Mic system when used with the CP910 processor. The use of a remote microphone accessory surpasses the benefit of application of NR algorithm.



**Rissmann A, Koehn A, Loderstedt M, Schwemmler C, Goetze G, Bartel S, Plontke SK, Langer J, Begall K, Matulat P, Roehl FW, Vorwerk U.**

[Population-based cross-sectional study to assess newborn hearing screening program in central Germany.](#)

*Int J Pediatr Otorhinolaryngol.* 2018 Apr;107:110-120. doi: 10.1016/j.ijporl.2018.01.035. Epub 2018 Jan 31.

**OBJECTIVES:** Airway management during adenoidectomy is traditionally performed through endotracheal intubation (ETT). Laryngeal mask airway (LMA) may be less stimulating to the airway and allow for shorter overall operating room time. Previous studies report LMA use during adenotonsillectomy with conversion rates to ETT of up to 17%. There has been no prior evaluation of LMA use during adenoidectomy alone. In this study, we attempt to identify the rate and contributing factors of LMA failure during adenoidectomy.

**METHODS:** All pediatric patients undergoing adenoidectomy between January 1, 2016 and June 30, 2017 were reviewed. Demographic and clinical data were collected and analysed to determine the need for conversion to ETT and the occurrence of any complications.

**RESULTS:** Our study revealed 139 pediatric patients who underwent adenoidectomy during the study period. 110 patients had adenoidectomy performed with LMA and 27 patients had ETT. Two patients (1.8%) required conversion to ETT because of difficulty with ventilation when the mouth gag was in place. There were no complications. Mean operating room time was 20 min less in the LMA group ( $P < 0.05$ ).

**CONCLUSIONS:** The use of an LMA in adenoidectomy may be a safe and effective alternative to ETT. More study is required to determine overall complication rates.

**Sato T, Nakazawa M, Takahashi S, Mizuno T, Sato A, Noguchi A, Sato M, Katagiri S, Yamada T.**

[Leaflets and continual educational offerings led to increased coverage rate of newborn hearing screening in Akita.](#)

*Auris Nasus Larynx.* 2017 Nov 28. pii: S0385-8146(17)30685-5. doi: 10.1016/j.anl.2017.11.014.

**OBJECTIVE:** Newborn hearing screening (NHS) has been actively performed in Japan since 2001. The NHS coverage rate has increased each year in Akita Prefecture. We analyzed the details of the NHS program and how the Akita leaflets and the many educational offerings about the importance of NHS led to the high NHS coverage rate

**METHODS:** A retrospective study was conducted in liveborn newborns in hospitals and in clinics where hearing screening was performed from the program's beginning in 2001 through the end of 2015. We describe the chronological history of NHS. The outcome data of NHS were collected from our department and analyzed.

**RESULTS:** From the founding of the program in 2001 to 2015, the live birth rate in Akita continually declined. Nevertheless, the number of infants receiving NHS rose each year. Since 2012, the coverage rate of NHS has been over 90%. From 2001 to 2015, 75,331 newborns constituted the eligible population for the NHS program. Since 2012, the number of NHS tests has stabilized. We prepared educational leaflets for Akita Prefecture early in 2002. We also provided many educational classes about the importance of NHS for not only pregnant women but also professionals including obstetricians and gynecologists, pediatricians and municipal staff members. The NHS program received the complete endorsement of the Akita Association of Obstetricians and Gynecologists in 2010. The largest increase in the NHS coverage rate occurred from 2001 to 2002, and the second largest increase occurred from 2009 to 2010. The number of participating institutions increased the coverage rate. The coverage rate is strongly correlated with the number of participating institutions ( $r_s=0.843$ ,  $p<0.001$ , Spearman's rank correlation coefficient). Comparing the coverage rate for 5 years before and after the Akita Association of Obstetricians and Gynecologists reached their consensus on the importance of NHS, the coverage rate after 2010 was significantly higher than before 2010 ( $p<0.001$ , paired sample t-test).

**CONCLUSION:** The NHS coverage rate ultimately reached 95.4% without need for legislation or subsidization. The number of participating institutions increased each year, and the number of NHS tests and the coverage rate increased proportionately. The number of participating institutions statistically has a strong correlation with the number of NHS tests and the coverage rate. Our research indicates that the Akita leaflets and the provision of educational sessions about the importance of NHS were the most significant factors in establishing the high NHS coverage rate.

**Satterfield-Nash A, Kotzky K, Allen J, Bertolli J, Moore CA, Pereira IO, Pessoa A, Melo F, Santelli ACFES, Boyle CA, Peacock G.**

[Health and Development at Age 19-24 Months of 19 Children Who Were Born with Microcephaly and Laboratory Evidence of Congenital Zika Virus Infection During the 2015 Zika Virus Outbreak - Brazil, 2017.](#)

*MMWR Morb Mortal Wkly Rep.* 2017 Dec 15;66(49):1347-1351. doi: 10.15585/mmwr.mm6649a2.

In November 2015, the Brazilian Ministry of Health (MOH) declared the Zika virus outbreak a public health emergency after an increase in microcephaly cases was reported in the northeast region of the country (1). During 2015-2016, 15 states in Brazil with laboratory-confirmed Zika virus transmission reported an increase in birth prevalence of microcephaly (2.8 cases per 10,000 live births), significantly exceeding prevalence in four states without confirmed transmission (0.6 per 10,000) (2). Although children with microcephaly and laboratory evidence of Zika virus infection have been described in early infancy (3), their subsequent health and development have not been well characterized, constraining planning for the care and support of these children and their families. The Brazilian MOH, the State Health Secretariat of Paraíba, and CDC collaborated on a follow-up investigation of the health and development of children in northeastern Brazil who were reported to national surveillance with microcephaly at birth. Nineteen children with microcephaly at birth and laboratory



evidence of Zika virus infection were assessed through clinical evaluations, caregiver interviews, and review of medical records. At follow-up (ages 19-24 months), most of these children had severe motor impairment, seizure disorders, hearing and vision abnormalities, and sleep difficulties. Children with microcephaly and laboratory evidence of Zika virus infection have severe functional limitations and will require specialized care from clinicians and caregivers as they age.

**Schwarz Y, Kaufman GN, Daniel SJ.**

[Newborn hearing screening failure and maternal factors during pregnancy.](#)

*Int J Pediatr Otorhinolaryngol.* 2017 Dec;103:65-70. doi: 10.1016/j.ijporl.2017.09.027. Epub 2017 Sep 28.

**OBJECTIVE:** Temporary conductive hearing loss due to amniotic fluid accumulation in the middle ear cavity may lead to failure (false positive) in newborn hearing screening tests. The aim of this study was to identify whether amniotic fluid index has association with failure of the initial newborn otoacoustic emission (OAE) screening test.

**METHODS:** A cohort study in a tertiary hospital center (Royal Victoria Hospital, Montréal) was constructed from 70 newborns that failed the OAE test, but passed a subsequent auditory brainstem response (ABR) test, and 75 randomly selected newborns that passed initial otoacoustic emission testing. Maternal (including the amniotic fluid index in the third trimester) and newborn clinical data were extracted from medical records. Statistical association models were built to determine variables that influenced hearing screen passage or failure.

**RESULTS:** The two arms of the cohort had no significant differences in maternal or child clinical indices, including in amniotic fluid index. Calculated as individual odds ratios, maternal tobacco [95% CI of odds ratio: 0.04, 0.59,  $p = 0.0078$ ], and drug use [95% CI of odds ratio: 0.0065, 0.72,  $p = 0.058$ ] [borderline significance] were associated with failing the otoacoustic emission testing.

**CONCLUSIONS:** Amniotic fluid index was not found to be associated with failure of otoacoustic emission screening in newborns. However, our study unveiled an interesting unexpected association of OAE failure with maternal smoking and/or drug use. This finding can help alleviate some of the time, cost and parental anxiety related to failed OAE screening. In selected cases of maternal smoking or drug use we might want to replace or add OAE to the ABR test in newborn hearing screening protocols, that don't perform both tests before discharge.

**Smith A, O'Connor A, Hennessy S, O'Sullivan PG, Gibson L.**

[Permanent Childhood Hearing Impairment: Aetiological Evaluation of Infants identified through the Irish Newborn Hearing Screening Programme.](#)

*Ir Med J.* 2017 Dec 18;110(10):651.

The Newborn Hearing Screening Programme (NHSP) was established in Cork University Maternity Hospital (CUMH) in April 2011. Between April 2011 and July 2014, 42 infants were identified with a Permanent Childhood Hearing Impairment (PCHI). Following this diagnosis, infants underwent a paediatric assessment according to recognised guidelines with the intention of identifying the underlying aetiology of the PCHI. The aim of this study was to assess the findings of this aetiological workup via retrospective chart review. PCHI data was obtained from the eSP database. This is a web based information system (eSP) used to track each baby through the screening and referral process. A retrospective chart review of these patients was performed. Sixteen (38%) infants were diagnosed with a bilateral sensorineural hearing loss. Two infants had congenital CMV infection. A Connexin 26 gene mutation was detected in one infant. Two infants were diagnosed with Waardenburg syndrome, One with Pendred syndrome and one with Pfeiffer syndrome. Five babies underwent cochlear implantation. Through adherence to the recommended protocol a possible cause of PCHI may be determined. This study has identified areas of future improvement for this service in Ireland.

**Suskind DL, Leung CYY, Webber RJ, Hundertmark AC, Leffel KR, Fuenmayor Rivas IE, Grobman WA.**

[Educating Parents About Infant Language Development: A Randomized Controlled Trial.](#)

*Clin Pediatr (Phila).* 2017 Oct 1:9922817737079. doi: 10.1177/0009922817737079. [Epub ahead of print]

A total of 427 women (aged 18-45 years) who delivered a singleton neonate without serious medical complications were randomized to watch either an educational intervention ( $n = 225$ ) or the sudden infant death syndrome ( $n = 202$ ) video. Linear mixed models showed that the intervention women significantly gained knowledge over time. Knowledge gain was largest among high-socioeconomic status (high-SES) and middle-SES English-speaking, smaller among low-SES Spanish-speaking, and nonsignificant among low-SES English-speaking women. Analysis of deviance revealed that the intervention women of all SES learned strategies fostering secure attachment and language acquisition. Participants considered watching an educational video alongside the universal newborn hearing screening (UNHS) conveniently timed. The intervention women were more likely than the control women to recognize the importance of timely UNHS follow-up.

**van Beeck Calkoen EA, Merkus P, Goverts ST, van de Kamp JM, Mulder MF, Sanchez Aliaga E, Hensen EF.**

[Evaluation of the outcome of CT and MR imaging in pediatric patients with bilateral sensorineural hearing loss.](#)

*Int J Pediatr Otorhinolaryngol.* 2018 May;108:180-185. doi: 10.1016/j.ijporl.2018.02.022. Epub 2018 Feb 15.

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

**STUDY DESIGN:** Retrospective cohort study.

**SETTING:** Tertiary referral otology and audiology center.

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

**MAIN OUTCOME MEASURES:** Radiologic abnormalities associated with SNHL.

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

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

**Vancor E, Shapiro ED, Loyal J.**

[Results of a Targeted Screening Program for Congenital Cytomegalovirus Infection in Infants Who Fail Newborn Hearing Screening.](#)

*J Pediatric Infect Dis Soc.* 2018 Jan 24. doi: 10.1093/jpids/pix105. [Epub ahead of print]

**Background:** Congenital cytomegalovirus (CMV) infection is a major cause of sensorineural hearing loss. By law, newborns in Connecticut who fail newborn hearing screening are tested for infection with CMV. This targeted screening is controversial, because most children with congenital CMV infection are asymptomatic, and CMV-related hearing loss can have a delayed onset. Our hospital uses a saliva polymerase chain reaction (PCR) assay (confirmed by a urine PCR assay) to detect CMV. Here, we report the results of the first year of our screening program.

**Methods:** We reviewed the medical records of newborns in the Yale New Haven Health System who failed the newborn hearing screening test between January 1 and December 31, 2016.

**Results:** Of 10964 newborns, 171 failed newborn hearing screening, and 3 of these newborns had positive saliva CMV PCR test results. Of these 3 newborns, 2 had positive results on the confirmatory test (for 1 of them the confirmatory test was not performed until the infant was 10 weeks old), and 1 had a negative result on the confirmatory test. Three additional newborns with congenital CMV infection were tested because of clinical indications (1 for ventriculomegaly on prenatal ultrasound and 2 for CMV infection of the mother). Results of audiology follow-up were available for 149 (87.1%) of the 171 newborns who failed newborn hearing screening; 127 (85.2%) had normal results.

**Conclusion:** Our targeted screening program for congenital CMV infection had a low yield. Consideration should be given to other strategies for identifying children at risk of hearing loss as a result of congenital CMV infection.

**Verkerk MM, Wagner R, Fishchuk R, Fagan JJ.**

[Survey of otolaryngology services in Ukraine and neighbouring Central and Eastern European countries.](#)

*J Laryngol Otol.* 2017 Nov;131(11):1002-1009. doi: 10.1017/S0022215117002134. Epub 2017 Oct 30.

**OBJECTIVE:** The present humanitarian crisis in Ukraine is putting strains on its healthcare system. This study aimed to assess services and training in otolaryngology, audiology and speech therapy in Ukraine and its geographical neighbours.

**METHOD:** Survey study of 327 otolaryngologists from 19 countries.

**RESULTS:** Fifty-six otolaryngologists (17 per cent) from 15 countries responded. Numbers of otolaryngologists varied from 3.6 to 12.3 per 100 000 population (Ukraine = 7.8). Numbers of audiologists varied from 0, in Ukraine, to 2.8 per 100 000, in Slovakia, and numbers of speech therapists varied from 0, in Bulgaria, to 4.0 per 100 000, in Slovenia (Ukraine = 0.1). Ukraine lacks newborn and school hearing screening, good availability of otological drills and microscopes, and a cochlear implant programme.

**CONCLUSION:** There is wide variation in otolaryngology services in Central and Eastern Europe. All countries surveyed had more otolaryngologists per capita than the UK, but availability of audiology and speech and language therapy is poor. Further research on otolaryngology health outcomes in the region will guide service improvement.

**Vo QT, Pham D, Choi KJ, Nguyen UTT, Le L, Shanewise T, Tran L, Nguyen N, Lee WT.**

[Solar-powered hearing aids for children with impaired hearing in Vietnam: a pilot study.](#)

*Paediatr Int Child Health.* 2018 Feb;38(1):40-45. doi: 10.1080/20469047.2016.1276119. Epub 2017 Jan 25.

**BACKGROUND:** Hearing loss is a barrier to speech and social and cognitive development. This can be especially pronounced in children living in low- and middle-income countries with limited resources. AIM: To determine the feasibility, durability and social impact of ComCare GLW solar-powered hearing aids provided for Vietnamese children with hearing impairment.

**METHODS:** A retrospective review of data from an international, multi-discipline humanitarian visit was performed. Hearing aids were given to 28 children enrolled at the Khoai Chau Functional Rehabilitation School, Hung Yen Province, Vietnam. Device inspection and observational assessments were performed by teachers using a modified Parents' Evaluation of Aural/Oral Performance of Children and an Infant Hearing Program Amplification Benefit Questionnaire. Qualitative interviews were undertaken to assess the study aims.

**RESULTS:** Hearing aids were well tolerated for use during regular school hours. All units remained functional during the study period (12 months). Teachers noted increased student awareness and responsiveness to surrounding sounds, but the degree of response to amplification varied between children. There was no significant improvement in speech development as all subjects had prelingual deafness. Teachers felt confident in troubleshooting any potential device malfunction.

**CONCLUSIONS:** A solar-powered hearing aid may be a viable option for children in low- and middle-income countries. This study demonstrates that device distribution, maintenance and function can be established in countries with limited resources, while providing feasibility data to support future studies investigating how similar devices may improve the quality of life of those with hearing loss.

**Vos B, Senterre C, Boutsen M, Lagasse R, Levêque A.**

[Improving early audiological intervention via newborn hearing screening in Belgium.](#)

*BMC Health Serv Res.* 2018 Jan 30;18(1):56. doi: 10.1186/s12913-018-2878-3.

**BACKGROUND:** Newborn hearing screening programs aim to lower the ages at audiological intervention among hearing-impaired children. In Wallonia and Brussels (Belgium), audiological intervention data are not collected in the screening program, and the ages at initiating audiological care have never been assessed. This study aimed to assess the evolution in the ages at initiating audiological intervention in the context of a newborn hearing screening program implementation.

**METHODS:** This population-based descriptive study used data from the Belgian healthcare billing database. The main outcomes were the children's ages at the initial audiological assessment, hearing-aid fitting, and cochlear implantation. Results were compared to the same outcomes from another Belgian regional program (Flanders) that was implemented one decade earlier. Annual birth cohorts from 2006 to 2011 were included in the study.

**RESULTS:** In Wallonia-Brussels, the median ages for all outcomes tended to decrease over time but remained higher than in Flanders for each birth cohort. For all outcomes except the hearing-aid fitting, differences in median ages between the two regions became less pronounced during the study period. In 2006, <23% of the children from Wallonia-Brussels received any audiological care before the age of 12 months and these proportions were approximately 2-fold greater in the subsequent birth cohorts. For all outcomes, early care (< 12 months) was typically delivered less frequently in Wallonia-Brussels, compared to the delivery in Flanders. These region-specific differences exhibited a decreasing trend over time, and statistically significant differences were less common in the later birth cohorts.

**CONCLUSIONS:** We conclude that the hearing screening program in Wallonia and Brussels promoted earlier audiological intervention among hearing-impaired children. However, milestones recommended by experts for an early intervention were not totally encountered. We also recommend collecting audiological intervention data as part of this program, which can facilitate more accurate and regular program evaluation.

**Wallis KE, Blum NJ, Waryasz SA, Augustyn M.**

[Bilateral Cochlear Implants: Maximizing Expected Outcomes.](#)

*J Dev Behav Pediatr.* 2018 Feb/Mar;39(2):177-179. doi: 10.1097/DBP.0000000000000547.

**CASE:** Sonia is a 4 years 1 month-year-old girl with Waardenburg syndrome and bilateral sensorineural hearing loss who had bilateral cochlear implants at 2 years 7 months years of age. She is referred to Developmental-Behavioral Pediatrics by her speech/language pathologist because of concerns that her language skills are not progressing as expected after the cochlear implant. At the time of the implant, she communicated using approximately 20 signs and 1 spoken word (mama). At the time of the evaluation (18 months after the implant) she had approximately 70 spoken words (English and Spanish) and innumerable signs that she used to communicate. She could follow 1-step directions in English but had more difficulty after 2-step directions. Sonia was born in Puerto Rico at 40 weeks gestation after an uncomplicated pregnancy. She failed her newborn hearing test and was given hearing aids that did not seem to help. At age 2 years, Sonia, her mother, and younger sister moved to the United States where she was diagnosed with bilateral severe-to-profound hearing loss. Genetic testing led to a diagnosis of Waardenburg syndrome (group of genetic conditions that can cause hearing loss and changes in coloring [pigmentation] of the hair, skin, and eyes). She received bilateral cochlear implants 6 months later. Sonia's mother is primarily Spanish-speaking and mostly communicates with her in Spanish or with gestures but has recently begun to learn American Sign Language (ASL). In a preschool program at a specialized school for the deaf, Sonia is learning both English and ASL. Sonia seems to prefer to use ASL to communicate. Sonia receives speech and language therapy (SLT) 3 times per week (90 minutes total) individually in school and once per week within a group. She is also receiving outpatient SLT once per week. Therapy sessions are completed in English, with the aid of an ASL interpreter. Sonia's language scores remain low, with her receptive skills in the first percentile, and her expressive skills in the fifth percentile. During her evaluation in Developmental and Behavioral Pediatrics, an ASL interpreter

was present, and the examiner is a fluent Spanish speaker. Testing was completed through a combination of English, Spanish, and ASL. Sonia seemed to prefer ASL to communicate, although she used some English words with errors of pronunciation. On the Beery Visual-Motor Integration Test, she obtained a standard score of 95. Parent and teacher rating scales were not significant for symptoms of attention-deficit/hyperactivity disorder. What factors are contributing to her slow language acquisition and how would you modify her treatment plan?

**Wang J, Sun J, Sun J, Chen J.**

[Variations in electrode impedance during and after cochlear implantation: Round window versus extended round window insertions.](#)

*Int J Pediatr Otorhinolaryngol.* 2017 Nov;102:44-48. doi: 10.1016/j.ijporl.2017.08.032. Epub 2017 Aug 31.

**OBJECTIVES:** To assess differences in intra- and postoperative electrode impedances following cochlear implantation between round window insertions (RWI) and extended round window insertions (ERWI).

**METHODS:** Fifty patients with congenital hearing loss received unilateral hearing implants (Sonata Ti100, Med-El GmbH, Innsbruck, Austria) with standard electrode arrays. The patients were divided into two groups according to the surgical technique used. Thirty-five procedures were performed with RWI (group A) and 15 with ERWI (group B). Electrode impedance was measured and analysed during the operation, and one week and one month postoperatively.

**RESULTS:** There were no statistically significant differences (i.e.,  $P > 0.05$ ) in electrode impedance between groups A and B intraoperatively, or at one week or one month postoperatively. Electrode impedance at one month postoperatively was higher than the intraoperative and postoperative one week values in group A ( $P < 0.05$ ), with similar results in group B.

**CONCLUSION:** There was no significant difference between RWI and ERWI in operative duration or complications of cochlear implantation. Moreover, no significant differences in postoperative electrode impedance values were found between the two surgical routes.

**Wang X, Wu D, Zhao Y, Li D, He D.**

[Knowledge and attitude of mothers regarding infant hearing loss in Changsha, Hunan province, China.](#)

*Int J Audiol.* 2017 Dec;56(12):997-1002. doi: 10.1080/14992027.2017.1366671. Epub 2017 Oct 16.

**OBJECTIVE:** The objective of this study was to explore the knowledge and attitude among mothers of newborns regarding infant hearing loss (HL) in Changsha, Hunan province, China.

**DESIGN:** A questionnaire including 18 items was given to mothers. **STUDY SAMPLE:** A total of 115 mothers participated in the study.

**RESULTS:** Seven risk factors for hearing loss were identified correctly by above 60% of respondents and the top three were prolonged noise (88.7%), high fever (82.6%) and ear discharge (82.6%). Poor knowledge was demonstrated on risk factors jaundice (20.0%), measles (22.6%), convulsion (33.0%) and traditional Chinese medicine (39.1%). Maternal knowledge scores in identification and intervention ( $2.68 \pm 0.31$ ) was slightly higher than the score in risk factors ( $2.47 \pm 0.34$ ). Ninety-nine per cent of the mothers expressed the willingness to test baby's hearing soon after birth and concern about hearing.

**CONCLUSIONS:** Mothers were concerned about baby's hearing and the attitude was positive. However, the correct recognition rate towards some risk factors for HL was low. Action needs to be taken to raise awareness about ear and hearing care, prevent HL caused by preventable causes and prompt early identification, early diagnosis and intervention of HL.

**Wang Y, Fan X, Wang P, Fan Y, Chen X.**

[Hearing improvement with softband and implanted bone-anchored hearing devices and modified implantation surgery in patients with bilateral microtia-atresia.](#)

*Int J Pediatr Otorhinolaryngol.* 2018 Jan;104:120-125. doi: 10.1016/j.ijporl.2017.11.010. Epub 2017 Nov 14.

**OBJECTIVE:** To evaluate auditory development and hearing improvement in patients with bilateral microtia-atresia using softband and implanted bone-anchored hearing devices and to modify the implantation surgery.

**METHODS:** The subjects were divided into two groups: the softband group (40 infants, 3 months to 2 years old, Ponto softband) and the implanted group (6 patients, 6-28 years old, Ponto). The Infant-Toddler Meaning Auditory Integration Scale was used conducted to evaluate auditory development at baseline and after 3, 6, 12, and 24 months, and visual reinforcement audiometry was used to assess the auditory threshold in the softband group. In the implanted group, bone-anchored hearing devices were implanted combined with the auricular reconstruction surgery, and high-resolution CT was used to assess the deformity preoperatively. Auditory threshold and speech discrimination scores of the patients with implants were measured under the unaided, softband, and implanted conditions.

**RESULTS:** Total Infant-Toddler Meaning Auditory Integration Scale scores in the softband group improved significantly and approached normal levels. The average visual reinforcement audiometry values under the unaided and softband conditions were  $76.75 \pm 6.05$  dB HL and  $32.25 \pm 6.20$  dB HL ( $P < 0.01$ ), respectively. In the implanted group, the auditory thresholds under the unaided, softband, and implanted conditions were  $59.17 \pm 3.76$  dB HL,  $32.5 \pm 2.74$  dB HL, and  $17.5 \pm 5.24$  dB HL ( $P < 0.01$ ), respectively. The respective speech discrimination scores were  $23.33 \pm 14.72\%$ ,  $77.17 \pm 6.46\%$ , and  $96.50 \pm 2.66\%$  ( $P < 0.01$ ).



**CONCLUSIONS:** Using softband bone-anchored hearing devices is effective for auditory development and hearing improvement in infants with bilateral microtia-atresia. Wearing softband bone-anchored hearing devices before auricle reconstruction and combining bone-anchored hearing device implantation with auricular reconstruction surgery may be the optimal clinical choice for these patients, and results in more significant hearing improvement and minimal surgical and anesthetic injury.

**Wang Y, Bergeson TR, Houston DM.**

[Infant-Directed Speech Enhances Attention to Speech in Deaf Infants With Cochlear Implants.](#)

*J Speech Lang Hear Res.* 2017 Nov 9;60(11):3321-3333. doi: 10.1044/2017\_JSLHR-H-17-0149.

**Purpose:** Both theoretical models of infant language acquisition and empirical studies posit important roles for attention to speech in early language development. However, deaf infants with cochlear implants (CIs) show reduced attention to speech as compared with their peers with normal hearing (NH; Horn, Davis, Pisoni, & Miyamoto, 2005; Houston, Pisoni, Kirk, Ying, & Miyamoto, 2003), which may affect their acquisition of spoken language. The main purpose of this study was to determine (a) whether infant-directed speech (IDS) enhances attention to speech in infants with CIs, as compared with adult-directed speech (ADS), and (b) whether the degree to which infants with CIs pay attention to IDS is associated with later language outcomes.

**Method:** We tested 46 infants-12 prelingually deaf infants who received CIs before 24 months of age and had 12 months of hearing experience (CI group), 22 hearing experience-matched infants with NH (NH-HEM group), and 12 chronological age-matched infants with NH (NH-CAM group)-on their listening preference in 3 randomized blocks: IDS versus silence, ADS versus silence, and IDS versus ADS. We administered the Preschool Language Scale-Fourth Edition (PLS-4; Zimmerman, Steiner, & Pond, 2002) approximately 18 months after implantation to assess receptive and expressive language skills of infants with CIs.

**Results:** In the IDS versus silence block, all 3 groups looked significantly longer to IDS than to silence. In the ADS versus silence block, both the NH-HEM and NH-CAM groups looked significantly longer to ADS relative to silence; however, the CI group did not show any preference. In the IDS versus ADS block, whereas both the CI and NH-HEM groups preferred IDS over ADS, the NH-CAM group looked equally long to IDS and ADS. IDS preference quotient among infants with CIs in the IDS versus ADS block was associated with PLS-4 Auditory Comprehension and PLS-4 Expressive Communication measures.

**Conclusions:** Two major findings emerge: (a) IDS enhances attention to speech in deaf infants with CIs; (b) the degree of IDS preference over ADS relates to language development in infants with CIs. These results support a focus on input in developing intervention strategies to mitigate the effects of hearing loss on language development in infants with hearing loss.

**Wang Z, Liu Y, Wang L, Shen X, Han S, Wang W, Gao F, Liang W, Peng KA.**

[Characteristics of electrically evoked auditory brainstem responses in patients with cochlear nerve canal stenosis receiving cochlear implants.](#)

*Int J Pediatr Otorhinolaryngol.* 2018 Jan;104:98-103. doi: 10.1016/j.ijporl.2017.10.029. Epub 2017 Oct 20.

**OBJECTIVE:** To explore the characteristics of the electrically evoked auditory brainstem responses (EABR) in children with cochlear nerve canal stenosis (CNCs) following cochlear implantation (CI), and the EABR thresholds in children with stenotic versus normal cochlear nerve canals.

**METHOD:** Sixteen children with profound sensorineural hearing loss were included in this study: 8 with CNCs (CNCs group) and 8 with normal cochlear nerve canals (control group). All children underwent cochlear implantation with full insertion of all electrodes. EABR was performed 6 months postoperatively in both groups.

**RESULTS:** The EABR extraction rate was 100% in children with normal cochlear nerve canals and only 50% in children with CNCs. EABR thresholds were significantly higher in children with CNCs of electrodes No. 11 and 22 than in children with normal cochlear nerve canals ( $P < 0.05$  for both comparisons). There was no significant difference in EABR thresholds among electrode No. 1, 11 and 22 in CNCs group ( $P > 0.05$  for all comparisons); while in the control group, the EABR threshold at electrode No 22 was lower than those at both electrodes No. 11 and 1 ( $P < 0.05$  for both comparisons), and the EABR threshold at electrode No. 11 was also lower than that at electrode No. 1 ( $P < 0.05$ ).

**CONCLUSION:** The EABR thresholds in children with normal cochlear nerve canals vary according to the different locations of electrodes in the cochlea; while in children with CNCs, there was no significant difference among different electrode locations. The EABR thresholds in CNCs children were higher than those of children with normal cochlear nerve canals at electrode 11 and 22.

**Wenjin W, Xiangrong T, Yun L, Jingrong L, Jianyong C, Xueling W, Zhiwu H, Hao W.**

[Neonatal hearing screening in remote areas of China: a comparison between rural and urban populations.](#)

*J Int Med Res.* 2018 Feb;46(2):637-651. doi: 10.1177/0300060517706643. Epub 2017 Jun 12.

**Objectives** Universal neonatal hearing screening (UNHS) started late in some underdeveloped areas in China, with relatively scarce screening resources and a wide regional distribution. This study aimed to compare the screening performance between rural and urban populations, and to examine the characteristics and problems of UNHS in underdeveloped regions in China.



**Methods** A two-step hearing screening program was used in neonates born in Liuzhou Maternal and Child Health Hospital and in patients who were born in other hospitals, but admitted to the neonatal intensive care unit. This program involved distortion product otoacoustic emission and automated auditory brainstem response. Characteristics of each newborn, as well as the screening outcomes and performance were compared between rural and urban populations.

**Results** A total of 19,098 newborns were screened with a referral rate of 17.9% at the first step. Sixty-three (0.33%) newborns had hearing loss. The prevalence of permanent hearing loss was 2.25%. The average screening age was significantly older in the rural population than in the urban population in the first ( $P < 0.01$ ) and second steps of screening ( $P < 0.05$ ). The rural population had a higher referral rate in both steps than the urban population ( $P < 0.01$ ). The follow-up rate was much lower in the rural population than in the urban population ( $P < 0.05$ ), but dramatically increased in 2014 compared with the previous 2 years.

**Conclusions** A low follow-up rate is a critical issue when carrying out UNHS in developing countries, such as China, especially for rural populations. The government should establish more hearing referral centres to increase service coverage and supply financial assistance for low-income populations.

**Wroblewska-Seniuk K, Dabrowski P, Greczka G, Szabatowska K, Glowacka A, Szyfter W, Mazela J.**

[Sensorineural and conductive hearing loss in infants diagnosed in the program of universal newborn hearing screening.](#)

*Int J Pediatr Otorhinolaryngol.* 2018 Feb;105:181-186. doi: 10.1016/j.ijporl.2017.12.007. Epub 2017 Dec 8.

**OBJECTIVE:** The aim of this study was to analyze infants diagnosed with sensorineural or conductive hearing deficit and to identify risk factors associated with these defects.

**MATERIAL AND METHODS:** A retrospective analysis of infants diagnosed with hearing deficit based on the database of the universal newborn hearing screening program and medical records of the patients.

**RESULTS:** 27 935 infants were covered by the universal neonatal hearing screening program. 109 (0.39%) were diagnosed with hearing deficit and referred for treatment and rehabilitation. 56 (51.4%) children were diagnosed with conductive, 38 (34.9%) with sensorineural and 15 (13.8%) with mixed type of hearing deficit. Children with sensorineural hearing deficit more frequently suffered from hyperbilirubinemia ( $p < 0.05$ ), while infants with conductive hearing loss were more frequently diagnosed with isolated craniofacial anomalies ( $p < 0.05$ ). The prevalence of other risk factors did not differ between the groups. Sensorineural hearing deficit occurred almost 3 times more often bilaterally than unilaterally ( $p < 0.05$ ). In other types of hearing deficit, the difference was not significant. In children with conductive and mixed type of hearing loss the impairment was mainly mild while among those with sensorineural hearing deficit in almost 45% it was severe and profound ( $p < 0.05$ ). When analyzing the consistency between hearing screening test by means of otoacoustic emissions and the final diagnosis of hearing deficit we found that the highest agreement rate was observed in children with sensorineural hearing loss ( $p < 0.01$ ).

**CONCLUSIONS:** The prevalence of most risk factors of hearing deficit was similar in children with sensorineural, conductive and mixed type of hearing loss, only hyperbilirubinemia seemed to predispose to sensorineural hearing deficit and isolated craniofacial malformations seemed to be associated with conductive hearing loss. Sensorineural hearing deficit usually occurred bilaterally and was severe or profound, while conductive and mixed type of hearing deficit were most often of mild degree. Most children with the final diagnosis of sensorineural hearing deficit had positive result of hearing screening by means of otoacoustic emissions.

**Yamamoto R, Naito Y, Tona R, Moroto S, Tamaya R, Fujiwara K, Shinohara S, Takebayashi S, Kikuchi M, Michida T.**

[Audio-visual speech perception in prelingually deafened Japanese children following sequential bilateral cochlear implantation.](#)

*Int J Pediatr Otorhinolaryngol.* 2017 Nov;102:160-168. doi: 10.1016/j.ijporl.2017.09.022. Epub 2017 Sep 23.

**OBJECTIVES:** An effect of audio-visual (AV) integration is observed when the auditory and visual stimuli are incongruent (the McGurk effect). In general, AV integration is helpful especially in subjects wearing hearing aids or cochlear implants (CIs). However, the influence of AV integration on spoken word recognition in individuals with bilateral CIs (Bi-CIs) has not been fully investigated so far. In this study, we investigated AV integration in children with Bi-CIs.

**METHODS:** The study sample included thirty one prelingually deafened children who underwent sequential bilateral cochlear implantation. We assessed their responses to congruent and incongruent AV stimuli with three CI-listening modes: only the 1st CI, only the 2nd CI, and Bi-CIs. The responses were assessed in the whole group as well as in two sub-groups: a proficient group (syllable intelligibility  $\geq 80\%$  with the 1st CI) and a non-proficient group (syllable intelligibility  $< 80\%$  with the 1st CI).

**RESULTS:** We found evidence of the McGurk effect in each of the three CI-listening modes. AV integration responses were observed in a subset of incongruent AV stimuli, and the patterns observed with the 1st CI and with Bi-CIs were similar. In the proficient group, the responses with the 2nd CI were not significantly different from those with the 1st CI whereas in the non-proficient group the responses with the 2nd CI were driven by visual stimuli more than those with the 1st CI.

**CONCLUSION:** Our results suggested that prelingually deafened Japanese children who underwent sequential bilateral cochlear implantation exhibit AV integration abilities, both in monaural listening as well as in binaural listening. We also observed a higher influence of visual stimuli on speech perception with the 2nd CI in the non-proficient group, sug-

gesting that Bi-CIs listeners with poorer speech recognition rely on visual information more compared to the proficient subjects to compensate for poorer auditory input. Nevertheless, poorer quality auditory input with the 2nd CI did not interfere with AV integration with binaural listening (with Bi-CIs). Overall, the findings of this study might be used to inform future research to identify the best strategies for speech training using AV integration effectively in prelingually deafened children.

**Zych M, Greczka G, Dąbrowski P, Wróbel M, Szyfter-Harris J, Szyfter W.**

[The report of the Polish Universal Neonatal Hearing Screening Program in 2016.](#)

*Otolaryngol Pol.* 2018 Feb 28;72(1):1-4. doi: 10.5604/01.3001.0011.5913.

The Polish Universal Neonatal Hearing Screening Program (PUNHSP) has been carried out in Poland for 14 years. The main aim of this Program is to organize hearing screening tests and to gather the information about risk factors of hearing loss in almost all newborns in Poland. It consists of 496 centers at 3 referral levels. A total of 5 458 114 children had been registered in the Central Database (CDB) of PUNHSP by the 22nd of August 2017. Bilateral sensorineural hearing loss was the most frequently appearing hearing impairment in children. It was diagnosed in 260 cases in 2016. This report presents the most important results and conclusions concerning the running of the PUNHSP in 2016.