Association Between Craniofacial Anomalies and Newborn Hearing Screening Fail Rate

Kaitlyn F. Sheapp, AuD
Ruth S. Marin, PhD
Larry Medwetsky, PhD

1Department of Rehabilitation Medicine, Adventist HealthCare Shady Grove Medical Center, Rockville, MD
2Department of Rehabilitation Medicine, Adventist HealthCare White Oak Medical Center, Silver Spring, MD
3Department of Hearing and Speech Sciences, Gallaudet University, Washington, DC

Abstract

Purpose: Increased knowledge of the prevalence of various craniofacial anomalies and their associated risks for hearing loss can help (a) guide the development of evidence-based practice regarding detection and documentation of risk factors at birth, and (b) health care professionals make appropriate recommendations for follow-up testing and monitoring.

Method: Records were reviewed for 39,813 infants born at two hospitals between January 1, 2014 and December 31, 2019 to determine the association between the presence of craniofacial anomalies and newborn hearing screening fail rates. Prevalence of confirmed hearing loss for infants born with and without risk factors were also examined. Additionally, surveys were sent to state EHDI programs and newborn hearing screening program coordinators across the United States to determine how facilities document risk factors for hearing loss, specifically craniofacial anomalies.

Conclusions: Study outcomes revealed four primary conclusions: (a) Infants with craniofacial anomalies are at a greater risk for failing their newborn hearing screening; (b) There is a need to better delineate craniofacial anomaly risk factors into subgroups; (c) Follow-up audiolgic evaluations are not warranted for infants with preauricular sinuses/tags and; (d) A universal protocol needs to be developed for recording risk factors for all infants and for training newborn hearing screening (NBHS) staff to identify such risk factors.

Keywords: newborn hearing screening, risk factors, craniofacial anomalies, hearing loss, Early Hearing Detection and Intervention, Neonatal Intensive Care Unit

Acronyms: AABR = automated auditory brainstem response; CFA = craniofacial anomalies; DPOAE = distortion product otoacoustic emissions ECMO = extracorporeal membrane oxygenation; EHDI = early hearing detection and intervention; NBHS = newborn hearing screening; OAE = otoacoustic emissions; TEOAE = transient-evoked otoacoustic emissions; WBN = Well-Baby Nursery

Correspondence concerning this article should be addressed to: Kaitlyn F. Sheapp, AuD, Department of Rehabilitation Medicine, Adventist HealthCare, Shady Grove Medical Center, 9901 Medical Center Dr., Rockville, MD 20850. Email: audiologysheapp@gmail.com

Universal newborn hearing screening (NBHS) programs have been established to provide the early detection of, as well as guide intervention for, hearing loss in newborns (Joint Committee on Infant Hearing [JCIH], 2019). One of the primary goals of NBHS is to “maximize linguistic competence and literacy development for children who are deaf or hard of hearing” since they are more likely to fall behind their hearing peers in communication, cognition, reading, and social-emotional development (p. 898, JCIH, 2007) without early intervention. The 1-3-6 Early Hearing Detection and Intervention (EHDI) model recommends that all infants be screened by one month of age, identified with hearing loss by three months of age, and receive intervention by six months of age (JCIH, 2019) so that they have the best chance to reach their potential.

The Joint Committee on Infant Hearing (JCIH) currently identifies 12 risk indicators that are associated with congenital, late onset, or progressive hearing loss in newborns: family history of childhood hearing loss, neonatal intensive care unit (NICU) stay of greater than five days, hyperbilirubinemia with exchange transfusion, treatment with ototoxic medications for greater than five days, asphyxia or hypoxic ischemic encephalopathy, extracorporeal membrane oxygenation (ECMO), in-utero infections, craniofacial conditions and physical conditions associated with hearing loss, syndromes associated with hearing loss, perinatal or postnatal bacterial and/or viral meningitis or encephalitis, events associated with hearing loss, and family/caregiver concern. It is imperative that infants identified as having one or more of these risk factors be closely monitored and re-evaluated routinely to rule out later onset or progressive sensorineural, mixed, or conductive hearing loss, regardless of NBHS screening results. The specific timing and number of these evaluations vary for each individual based on the identified
risk factor(s) and clinical judgment of the audiologist and/or primary care provider. It is recommended, however, that infants who have a craniofacial anomaly (CFA), regardless of the type, be re-evaluated by nine months of age (JCIH, 2019). It is the responsibility of the pediatrician or primary care provider (also known as the medical home) to monitor these risk indicators to ensure that audiological evaluations are completed as recommended (JCIH, 2019).

Research suggests that there is a lack of knowledge among healthcare professionals as to which of the aforementioned risk factors are “discoverable, predictive, and useful” (Karace et al., 2014, p. 262). This lack of knowledge lessens the effectiveness of initial screening and the impact of the JCIH guidelines, as well as the occurrence of follow-up testing, because medical professionals and newborn hearing staff are often unable to recognize the need for follow-up hearing testing when a specified risk factor is present (Hutt & Rhodes, 2008). Increasing knowledge about the various risk factors and their associated risk for hearing loss will help guide evidence-based practice and policy development regarding the detection and intervention of hearing loss in infants (Hutt & Rhodes, 2008).

The present study addresses NBHS outcomes for infants who have a documented CFA. CFAs are defined as those that include microtia/atrophia, ear dysplasia, oral facial clefting, white forelock, microphthalmia, congenital microcephaly, congenital or acquired hydrocephalus and/or temporal bone abnormalities, and skull malformations (JCIH, 2019). CFAs are also found within certain syndromes such as: Trisomy 21, Treacher Collins syndrome, Waardenburg syndrome, CHARGE Association, Crouzon disease, Klippel-Feil syndrome, DiGeorge syndrome, Goldenhar syndrome and Pierre Robin syndrome (Greydanus et al., 2007). There is substantial evidence that the CFA risk factor group yields the highest prevalence of hearing loss in infants and children, however, the prevalence of hearing loss associated with each specific craniofacial disorder has not been consistently reported in the literature (Appelbaum et al., 2018; Cone-Wesson et al., 2000; Duman et al., 2017; Yelverton et al., 2013). For example, the published prevalence of hearing loss in cleft lip and cleft palate ranges from 26% to 82% (Chen et al., 2008; Viswanathan, Vidler, & Richard, 2008) and .3% to 18% for preauricular sinuses and tags (Firat et al., 2008; Kankkunen & Thiringer, 1987; Roth et al., 2008). These variations appear to be due to different methods for ascertaining the presence of hearing loss and risk factors including newborn hearing screenings, retrospective review of medical charts, and auditory brainstem response threshold assessments. There is also insufficient data on prevalence figures for other CFAs such as malformed ears, microtia, and skull malformations. Consequently, the exact association between each specific CFA and hearing loss risk at birth is unknown. Risk factors are only as “useful as their predictive power” (Karace et al., 2014, p. 262); therefore, it is imperative to determine the associated risk for hearing loss at birth for each of these disorders. In turn, this could lead to the development of effective follow-up guidelines and recommendations appropriate to each CFA. Since not every CFA has the same incidence/prevalence of congenital, progressive, or late-onset hearing loss, this clarification is crucial.

Aside from a lack of knowledge concerning the prevalence of each specific CFA and their respective contributions to NBHS fail rates, there is also a lack of documentation in state databases regarding risk factor information from hospitals in the country (Hutt & Rhodes, 2008; White, 2014). It is a guideline, not a requirement, to record risk factors in the NBHS databases (JCIH, 2019). Current risk factor registers are designed to ensure that newborn infants who need evaluation and follow-up are identified, however, these registers often lack specific/universal criteria, are under-utilized, and NBHS programs likely underreport various risk factors associated with hearing loss in their databases (Hutt & Rhodes, 2008). Without documentation of risk factors, healthcare providers are unable to ensure efficient, effective, timely and appropriate follow-up recommendations. For example, in a study examining birth certificate records, Purcell and colleagues (2018) “found that only 39% of children with cleft palates were correctly identified as having a craniofacial risk factor [for hearing loss] at the time of hearing screening”, which ultimately led to a delayed diagnosis of hearing loss for many of these children (p. 26). That is, many of these children may have initially passed their newborn hearing screening but developed progressive/late onset hearing loss that was not caught. This delayed diagnosis of hearing loss may have been due to a lack of follow-up and monitoring for these children. Determining which CFAs result in a child having a greater risk for childhood hearing loss, in turn, ensures adequate and appropriate follow-up and intervention.

The following were the specific research questions examined in this study:

1. What is the association between the presence of CFAs (as a general category) and NBHS fail rates for infants born at two hospitals, Adventist HealthCare Shady Grove Medical Center (AHC SGMC) and Adventist HealthCare White Oak Medical Center (AHC WOMC) in the Greater DC area?

2. What is the association between specific CFAs and NBHS fail rates for infants born at AHC SGMC and AHC WOMC?

3. What is the prevalence of confirmed hearing loss (conductive, mixed, or, sensorineural) for infants born at AHC SGMC and AHC WOMC who failed the NBHS, with or without risk factors?

4. What information does each state’s EHDI program require from individual screening programs as it pertains to the documentation of CFAs? How successful are the EHDI programs in obtaining such information?

5. How well are NBHS programs across the country recording and documenting information about the presence of risk factors for hearing loss and CFAs?
Method

This study was approved by the Institutional Review Boards at Adventist HealthCare (2019-29) and Gallaudet University (Legacy-IRB-FY20-04). Descriptive statistics were used to report the outcomes obtained from this study.

Participation

To answer the first three research questions, records were reviewed for 39,813 infants born at AHC SGMC and AHC WOMC over a six-year period between January 1, 2014 and December 31, 2019; reporting requirements remained consistent during this time frame. Both hospitals provide newborn hearing screening services, but not diagnostic evaluations.

The following data were retrieved from the Maryland Early Hearing Detection and Intervention (MD EHDI) program database, also known as OZ: (a) risk factor(s) present for hearing loss, (b) NBHS outcomes, (c) outpatient screening outcomes (if applicable and if available), and (d) diagnostic outcomes (if applicable and if available). If specific CFA information was missing, or if confirmation of OZ was needed, the hospitals’ electronic medical records were subsequently reviewed. A review of both hospitals’ NBHS program department records was also conducted to obtain information about specific CFAs for each infant, and when available, to clarify any discrepancies in OZ, as well as identify any risk factors for hearing loss that were incorrectly documented in OZ. Incorrect documentation could include omissions of risk factors in the state database and inaccurately recorded results in the hospital records.

Both hospitals in this study used a two-step screening protocol. All babies in the Well-Baby Nursery (WBN) without a risk factor for hearing loss were tested using either transient otoacoustic emissions (TEOAE) or distortion product otoacoustic emissions (DPOAE). If an infant failed the initial screening, a second OAE test was performed the next day. If the infant failed the second OAE, then an automated auditory brainstem response (AABR) screening was performed. All babies born with a risk factor for hearing loss (with the exception of preauricular pits and preauricular tags) or treated in the NICU were screened using AABR. If the infant failed the initial AABR, a subsequent and final AABR was performed, time permitting. All OAE and AABR equipment were calibrated annually according to manufacturers’ guidelines. AHC WOMC uses the Otodynamics Otoport for portable, bedside TEOAE screening, while AHC SGMC uses the Maico EroScan DPOAE for bedside DPOAE screening. For AABR and additional TEOAE screening, both hospitals use the Intelligent Hearing Systems (IHS) Smart Screener Plus.

To answer research questions 4 and 5, two separate surveys were distributed to: (a) State EHDI leaders and (b) NBHS program coordinators throughout the United States. The EHDI state leader survey can be found in Appendix A, while the NBHS Coordinator Survey can be found in Appendix B. Participants for both surveys signed an informed consent form and remain anonymous. The surveys were designed to be completed easily and quickly by participants using multiple choice, multi-answer, yes/no, and open-ended questions. Sample surveys were piloted with two independent audiologists (one state leader and one NBHS program coordinator) to ensure ease of completion, address any ambiguities, and determine the time needed to complete the surveys.

Surveys were designed and posted on a secure online platform (REDCap), where participants were able to access and complete the survey anonymously. Requests for participation for each survey were distributed electronically in three ways. The first was through two professional audiology groups (American Speech-Language-Hearing Association and the American Academy of Audiology). The second was through posting on two audiology Facebook pages (Audiology Antics and Anecdotes- for professionals only, and Audiology Happy Hour). The last was by distributing two different emails to all state EHDI leaders. The first email requested their participation in the State EHDI Leader Survey; this email included a link to the REDCap survey. The second email requested that they forward via email a description of, and a link to, the NBHS Coordinator Survey to NBHS coordinators in their state. To complete the NBHS Coordinator Survey, the participants had to currently be in charge of a NBHS program at a hospital, birthing center, NICU or other facility providing NBHS. A follow-up email was distributed to EHDI state leaders four weeks after the first email was sent if they had not yet completed the survey.

For the EHDI state leader survey, data from 13 states was received. Participating states are not identified in this paper to protect the privacy of those who responded. A total of 90 participants responded to the NBHS program coordinator study with a total of 18 states being represented across various regions of the United States; one participant did not report in which state they practiced. Descriptive statistics were used to analyze responses from both surveys.

Results

Presence of Any CFA and NBHS Fail Rates

There were 39,813 infants born at AHC SGMC and AHC WOMC between January 1, 2014 and December 31, 2019. A total of 2.05% \( n = 817 \) of all infants born during this period had a CFA that was documented in the department’s paper records and/or hospital electronic medical records. The hearing screening fail rate for those identified with any CFA was 4.41% \( n = 36 \), compared to the overall fail rate of 0.74% \( n = 293 \) for all newborns, with or without a risk factor for hearing loss. The majority of the 293 infants who failed the NBHS had no risk factors for hearing loss (64.51%, \( n = 189 \)), while 12.29% \( n = 36 \) had a CFA, and 23.20% \( n = 68 \) had other risk factors for hearing loss.

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Association Between Specific CFAs and NBHS Fail Rates

As described earlier, a total of 817 of the 39,813 infants seen for a newborn hearing screening were identified as having a CFA. This group of infants was further categorized based on the specific anomalies present. The syndromes (associated with hearing loss) identified in our population were: Trisomy 13, Trisomy 18, 13q syndrome, Osteogenesis Imperfecta, Achondroplasia, Waardenburg syndrome, Goldenhaar syndrome, Pallister Killian syndrome, Beckwith Wiedemann syndrome, Turner syndrome, Albinism, and Cornelia de Lange syndrome.

Other CFAs present included preauricular sinuses/tags, atresia/microtia, malformed ears, skull malformations, and cleft lip/palate. The prevalence for each of these specific CFAs and their associated NBHS fail rates are reported in Table 1. It is important to also note that two infants in the cleft lip/palate category were counted in two categories: once in the skull malformation category and once in the syndrome category.

Table 1
Prevalence of Specific Craniofacial Anomalies (CFAs) at Birth and the Associated Newborn Hearing Screening (NBHS) Fail Rate

<table>
<thead>
<tr>
<th>Category of CFA</th>
<th>Total Percentage of CFA (n = 819)</th>
<th>NBHS Fail Rate (n = 36)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Syndrome associated with hearing loss</td>
<td>9.4% (77)</td>
<td>18.18% (14)</td>
</tr>
<tr>
<td>Ear Sinuses/Tags</td>
<td>82.25% (672)</td>
<td>0.74% (5)</td>
</tr>
<tr>
<td>Atresia/Microtia</td>
<td>1.47% (12)</td>
<td>100% (12)</td>
</tr>
<tr>
<td>Malformed Ears, other</td>
<td>1.35% (11)</td>
<td>36.36% (4)</td>
</tr>
<tr>
<td>Skull Malformations</td>
<td>0.61% (5)</td>
<td>20% (1)</td>
</tr>
<tr>
<td>Cleft Lip/Palate</td>
<td>4.16% (34)</td>
<td>5.8% (2)</td>
</tr>
<tr>
<td>Other</td>
<td>0.98% (8)</td>
<td>0.00% (0)</td>
</tr>
</tbody>
</table>

Note: Includes two babies with other CFAs: Syndrome & Skull Malformations

Considering that 82.25% of babies with a CFA had preauricular sinuses and tags, this category was further analyzed. Table 2 shows the prevalence of, and the fail rate for, each sub-category of preauricular sinuses and tags. Regardless of whether a sinus and/or tag was unilateral or bilateral, the NBHS fail rate was less than 1%.

Table 2
Prevalence of Unilateral vs Bilateral Preauricular Sinuses/Tags and the Associated Fail Rate

<table>
<thead>
<tr>
<th>Category of CFA</th>
<th>Prevalence (n)</th>
<th>NBHS Fail Rate (n)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Unilateral Preauricular Tags</td>
<td>30.95% (208)</td>
<td>0.96% (2)</td>
</tr>
<tr>
<td>Unilateral Preauricular Sinuses</td>
<td>51.34% (345)</td>
<td>0.29% (1)</td>
</tr>
<tr>
<td>Bilateral Preauricular Tags</td>
<td>2.68% (18)</td>
<td>0.00% (0)</td>
</tr>
<tr>
<td>Bilateral Preauricular Sinuses</td>
<td>14.43% (97)</td>
<td>0.21% (2)</td>
</tr>
<tr>
<td>Preauricular Sinuses and Preauricular Tags</td>
<td>0.61% (4)</td>
<td>0.00% (0)</td>
</tr>
</tbody>
</table>

Note: NBHS = Newborn Hearing Screening; CFA = Craniofacial Anomaly.

Prevalence of Confirmed Hearing Loss for Infants Who Failed the Newborn Hearing Screening, With or Without Risk Factors

Data from the outpatient hearing screenings, as well as diagnostic outcomes for infants who had failed their newborn hearing screening, were accessed through OZ. For the 293 infants who failed the newborn hearing screening, 70.99% (n = 208) were referred for a follow up screening, and were seen either internally or at an outside clinic. Of these infants, 69.23% (n = 144) passed the rescreening, 14.90% (n = 31) failed the rescreening, and 15.87% (n = 33) were lost to follow-up. The 31 infants who failed their rescreening were referred to a local children’s hospital for diagnostic testing. Those results revealed that 43.75% (n = 13) infants had normal hearing acuity, 46.88% (n = 15) were diagnosed with hearing loss, and 9.38% (n = 3) were lost to follow-up. Most of the infants who failed their initial screening and had a risk factor for hearing loss (n = 85) were referred directly to a pediatric audiologist for diagnostic evaluation. Hearing loss was identified in 48.23% (n = 41) of these infants, while normal hearing acuity was found in 30% (n = 26), and 21.12% (n = 18) were lost to follow-up. Note that the definition of normal hearing and hearing loss, as well as the degree and type of hearing loss, were not provided in OZ, so parameters are unknown.

A risk factor was present in 76.79% (n = 43) of the 56 infants who were diagnosed with hearing loss, while no risk factors were present in 23.21% (n = 13). A CFA was identified in 33.93% (n = 19) of the infants diagnosed with hearing loss; that is, 19 of the total number of infants identified with a risk factor and subsequently identified with a hearing loss had a CFA (i.e., 19/43 = 44.2%). Of these infants, unilateral hearing loss was found in 63.16% (n = 12), while 36.84% (n = 7) were diagnosed with bilateral hearing loss. For those infants who did not have a CFA (66.07%; n = 37), a unilateral hearing loss was diagnosed in 18.92% (n = 7) and a bilateral hearing loss was diagnosed in 81.08% (n = 30).
State EHDI Program Survey Results

As previously described, a total of 13 EHDI state leaders (22.03%) responded to the EHDI State Leader Survey. Results from this survey are presented in Table 3; the numbering beside each question corresponds to the specific question found in Appendix A.

### Table 3

**Results from the Early Hearing Detection and Intervention State Leader Survey**

<table>
<thead>
<tr>
<th>Survey Questions from EHDI State Leaders</th>
<th>Yes</th>
<th>No</th>
</tr>
</thead>
<tbody>
<tr>
<td>3) Does your state mandate newborn hearing screenings in all, or most, birthing facilities?</td>
<td>84.62% (n = 11)</td>
<td>15.38% (n = 2)</td>
</tr>
<tr>
<td>5) Are facilities required to document specific risk factors in the state EHDI database?</td>
<td>46.15% (n = 6)</td>
<td>53.85% (n = 7)</td>
</tr>
<tr>
<td>8) If a CFA is reported, do you request additional information about the specific CFA from program coordinators?</td>
<td>23.08% (n = 3)</td>
<td>76.92% (n = 10)</td>
</tr>
<tr>
<td>9) Are you satisfied with the documentation completed by the facilities in your state regarding the risk factors for hearing loss?</td>
<td>38.36% (n = 5)</td>
<td>61.54% (n = 8)</td>
</tr>
</tbody>
</table>

Note. EHDI = Early Hearing Detection and Intervention; CFA = Craniofacial Anomaly.

Multiple state leaders had suggestions for NBHS programs in response to question #10 “Do you have any suggestions for how to improve the recording, reporting or follow-up process for infants with risk factors for hearing loss, including infants with craniofacial anomalies?” Top responses included (a) involve primary care physicians, (b) increase the education and training for staff involved in NBHS programs, (c) include a system that automatically links data from the hospital/facility’s electronic health records system to the state database, and (d) utilize the Centers for Disease Control and Prevention Birth Defects Registry.

### NBHS Program Coordinator Survey Results

A total of 90 newborn hearing screening program coordinators responded to our survey. Appendix C1 demonstrates a breakdown of the professions of the program coordinators, Appendix C2 shows the professions of those conducting the newborn hearing screenings, and the breakdown of states in which the respondents practice is shown in Appendix C3.

When surveyed, 75.56% (n = 68) of the program coordinators reported that information regarding risk factors for hearing loss were collected at their facility. This information was collected in multiple ways (Question 9 from survey): by asking hospital staff, 28.89% (n = 26); checking infant medical records, 62.22% (n = 56); asking the infant’s mother case history questions, 60.00% (n = 54); and collecting risk factor information by another method, 3.90% (n = 3).

In some facilities, information concerning risk factor information was obtained from multiple sources, resulting in the total percentage exceeding 100%.

Table 4 displays responses to other questions (corresponding to the numbered questions in Appendix B) that were posed to the NBHS program coordinators. In addition to the information contained in Table 4, nine of the 90 NBHS coordinators (10%) also reported their newborn hearing screening fail rate for infants with CFAs.

### Table 4

**Results from the Newborn Hearing Screening Program Coordinator Survey**

<table>
<thead>
<tr>
<th>Survey Questions from Program Coordinator Survey</th>
<th>Yes</th>
<th>No</th>
</tr>
</thead>
<tbody>
<tr>
<td>6) Does your facility employ audiologists to oversee the program?</td>
<td>24.44% (n = 22)</td>
<td>75.56% (n = 68)</td>
</tr>
<tr>
<td>7) Is your staff trained to identify the different risk factors associated with childhood hearing loss?</td>
<td>33.33% (n = 36)</td>
<td>66.67% (n = 74)</td>
</tr>
<tr>
<td>8) Is information regarding risk factors for childhood hearing loss collected prior to or following each screening? (Questions 10 through 13 were recorded only if answered ‘yes’ for question 8)</td>
<td>75.56% (n = 68)</td>
<td>24.44% (n = 22)</td>
</tr>
<tr>
<td>10) Does your program record the type of risk factor(s) in the hospital medical records?</td>
<td>77.94% (n = 53)</td>
<td>22.06% (n = 15)</td>
</tr>
<tr>
<td>11) If an infant is identified as having a risk factor, does your program report the information to the state EHDI program?</td>
<td>76.47% (n = 52)</td>
<td>23.53% (n = 16)</td>
</tr>
<tr>
<td>12) If an infant is identified with a CFA, does your program record the specific type of CFA present?</td>
<td>76.47% (n = 52)</td>
<td>23.53% (n = 16)</td>
</tr>
<tr>
<td>13) Do you report the specific CFA to the state EHDI program?</td>
<td>69.23% (n = 36)</td>
<td>30.77% (n = 16)</td>
</tr>
<tr>
<td>14) Do you feel as though you are getting enough guidance from your state EHDI program on how to document and report risk factors?</td>
<td>58.89% (n = 53)</td>
<td>41.11% (n = 37)</td>
</tr>
</tbody>
</table>

Note. EHDI = Early Hearing Detection and Intervention; CFA = Craniofacial Anomaly.
Discussion

The overall newborn hearing screening fail rate measured at the two study hospitals (0.74%) was lower than the national fail rate of 4% (ASHA, n.d.). This lower fail rate may be attributed to the two-step screening protocol used at these two hospitals, as described in the Method section. A failed OAE followed by an AABR may reduce false positive rates since the AABR is less sensitive to vernix or debris in the ear canal which is a significant cause of failed screenings (American Speech-Language-Hearing Association [AHSA], n.d.). Without the follow-up AABR, there would be a much higher false-positive rate. Reducing false-positive rates (a) allows infants at risk to be more accurately targeted for follow-up testing, and (b) reduces the wait time and workload for pediatric audiologists by reducing the demand for diagnostic testing.

The hearing screening fail rate for those identified with a CFA (4.4%) is six times higher than the overall fail rate of 0.74% for all newborns. This supports the current JCIH inclusion of CFA on the list of risk factors for hearing loss. In examining the NBHS fail rates for the seven CFA subgroups identified in this study, the fail rates vary greatly, from 0% to 100%. This suggests the need to further delineate the craniofacial risk factor category into subgroups, with follow-up evaluation recommendations based on the specific CFA, rather than CFA category as a whole. For example, an infant with only a preauricular sinus (.25% fail rate) should not receive the same follow-up recommendations as an infant with microtia/ atresia (100% fail rate). Currently, the JCIH recommends audiological follow-up/re-evaluation by nine months of age, regardless of the type of CFA present (JCIH, 2019). However, appropriate follow-up evaluations should be recommended based on the specific needs of an infant to ensure that the infant has the best opportunity for early detection and intervention or plan for monitoring in case of a possible progressive or late-onset hearing loss. These findings suggest that JCIH should consider refining their list of risk factors for hearing loss to include separate recommendations for the specific CFAs.

Our screening fail rate of 0.74% for infants with preauricular sinuses and/or tags, in the absence of other physical findings, indicates that routine audiological reevaluation is not warranted for this population. After subdividing the preauricular sinuses/tag group into more specific sub-groups, we determined that infants with either a preauricular sinus or tag, whether unilateral or bilateral, exhibited similar NBHS fail rates (i.e., all less than 1%). A progressive hearing loss would not be expected if these were the only anomalies, however, it is recommended that these anomalies continue to be documented since they can be associated with various syndromes that do have a higher likelihood of hearing loss, such as Trisomy 21, Treacher Collins syndrome, CHARGE Association, Waardenburg Syndrome, Crouzon disease, and so on (Greydanus et al., 2007). Accurate documentation of preauricular sinuses and preauricular tags will assist the medical home in monitoring for any additional signs and/or symptoms associated with such syndromes. Research suggests that skin tags can also be associated with maternal diabetes and may not be related to any ear issues at all; therefore, it is important to obtain a mother’s medical history to determine if maternal diabetes is a possible underlying cause for the preauricular tags (Grix et al., 1982; Johnson, Fineman & Opitz, 1982; Sait et al., 2019).

Based on the findings from the two hospitals, of the 56 infants subsequently diagnosed with a hearing loss following their birth screening, infants with a risk factor for hearing loss (43/56) were 3.3 times more likely than infants without a risk factor (13/56) to be diagnosed with a hearing loss following their birth screening. And, of the infants with a risk factor(s) that were identified with a hearing loss, 19 of 43 (44.2%) of them had a CFA. This data provides strong evidence of the need for follow-up evaluations for all infants with a CFA, except for infants with preauricular sinuses and tags due to the low prevalence of NBHS fail rate.

Multiple errors were discovered when comparing the hospital records to OZ. Information pertaining to preauricular sinuses and tags that had been documented appropriately in the hospital records was omitted 154 times in the state database. In addition, one infant’s NBHS result was entered incorrectly into the state database as having passed the NBHS when, in fact, the infant had actually failed in both ears. When errors were discovered, they were corrected in the database. The two NBHS programs in this study are managed by audiologists and have a well-developed protocol for documenting and recording risk factors, yet, errors still occurred. Programs without such audiology oversight and thorough protocols could potentially have even more documentation errors.

Based on the survey findings, it appears that a majority of the NBHS program coordinators are documenting and reporting the presence of risk factors in their hospital medical records (see Table 4), however, the majority of the EDHI State Leaders indicated that newborn hearing screening programs were failing to document this information in their state EHDI records (see Table 3). In addition, responses suggest that close to 2/3 of the EDHI NBHS state leaders who responded (61.54%; n = 8) were not satisfied with how facilities in their state document and record risk factors for hearing loss. Currently, JCIH has a list of guidelines that each newborn hearing screening program should follow. Because the guidelines are suggestions, not policy, and because each program documents and reports risk factors differently, measuring prevalence figures for hearing loss risk is a challenge. If the findings from this study are indeed reflective of most NBHS programs, then these guidelines should become protocol. Having a universal protocol (including training of staff) would reduce the chances of NBHS programs overlooking risk factors, which would increase appropriate referrals for diagnostic testing and early intervention.

Results from the NBHS Coordinator Survey also indicate that some programs (16.67%; n = 15) do not train their newborn hearing screeners to identify the different risk factors for hearing loss. While conducting
the hospital records review for this study, it was noted that pediatricians at the hospitals often neglected to document the presence of preauricular sinuses and tags as well as other ear malformations during the newborn’s physical exam, and that our trained hearing screeners were often the ones to identify the presence of such abnormalities. Correctly and accurately identifying infants with risk factors for hearing loss, such as CFAs, helps guide referrals for follow up care. In addition to detecting physical anomalies associated with possible hearing loss, newborn hearing screening staff should also be trained to review hospital records and question parents to identify other possible risk factors for hearing loss, including a family history of childhood hearing loss. The NBHS Program Coordinator survey suggests that roughly a quarter of newborn hearing screening programs do not collect general risk factor information, as recommended by JCIH guidelines. Additionally, 31% of the newborn hearing screening programs do not report CFAs to their state database (see Table 4). This information is vital because without such information, infants with these risk factors may not receive the appropriate follow-up recommendations and referrals.

Responses from this survey also revealed that 75% \((n = 68/90)\) of facilities do not employ an audiologist to oversee the newborn hearing screening program. Having audiologist oversight, as recommended in the new JCIH 2019 guidelines, would increase the probability that staff are trained properly to identify risk factors for hearing loss, and ensure appropriate referrals and recommendations. In addition to having audiologist oversight, it is important for NBHS programs to identify the newborn’s medical home, and communicate the hearing screening results with them, if the infant failed their screening (ASHA, n.d.).

Approximately 40% \((n = 37)\) of the NBHS Program Coordinators reported that they did not feel they had received enough guidance from their state EHDI program as to how they should document and record risk factors for hearing loss. EHDI programs are designed to maintain a coordinated, statewide screening and referral system for those infants who do not pass the newborn hearing screening. Without strong guidance from their state EHDI program, infants are at risk for not receiving adequate and appropriate follow-up care (EHDI, 2020). Thus, based on the findings in this study, JCIH should consider developing a universal newborn hearing screening program protocol that all birthing facilities must follow.

**Study Limitations**

The first limitation of this study concerns the number of responses from NBHS program coordinators and state leaders of EDHI programs. It is hard to generalize our conclusions with responses from only 13 state leaders and 90 program coordinators from 18 different states. Increased participation would have allowed for a better representation of how NBHS programs across the country are recording and documenting risk factors for hearing loss.

A second limitation of the study was that the population from the two hospitals lacked geographic diversity. Both hospitals are located in suburban neighborhoods outside of Washington, DC. It is possible that the incidence of CFAs and the screening/documentation protocols may differ from birthing centers in other regions. Therefore, additional data are needed from birthing centers throughout the United States of America before our findings can be generalized.

A third limitation is that the technicians who performed the newborn hearing screenings could have omitted or incorrectly documented risk factors for hearing loss, particularly preauricular sinuses and preauricular tags, as well as other ear malformations. If these risk factors were missed or entered incorrectly, then the hearing screening fail rate relative to the possible risk factors and CFA type could be inaccurately represented.

A fourth limitation of this study was that some of the department paper records for a period of a few months were lost when one of the two study hospitals moved to a new location, while the other was not in possession of some of their records from one of the six years being studied. Therefore, the ability to cross-check information from the department records with OZ was impacted, leading to possible missed identification of infants with risk factors for hearing loss, and potentially influencing the screening fail rate that was obtained.

**Future Directions**

In this study, seven CFA sub-groups were described based on the anomalies present in the infants at the two study hospitals. Development of a standardized list of specific CFAs and their associated NBHS fail rates is needed to maintain consistency across and, in turn, guide NBHS programs in the United States. Because the findings in this study were obtained from only two hospitals, a larger scale study would provide information that could facilitate the development of policies to address the specific hearing needs of different sub-groups of the CFA group.

Having described NBHS fail rates in infants with different CFAs in this paper, future research should focus on describing the degree and type of hearing loss associated with this population. Because specific diagnostic audiology outcomes are not reported in OZ, we were unable to obtain and analyze this information. Collaboration with diagnostic testing facilities would allow access to this data. In addition, it would be beneficial for NBHS programs to document the specific diagnostic outcomes for infants who are diagnosed with a hearing loss in their state database. Such information would be beneficial for the medical home, as well as audiologists, as it would help guide appropriate monitoring and intervention.

Further research should aim to gather information about how risk factor information is being entered and stored into the various databases from a larger proportion of states. In addition, future studies should also examine how NBHS sites are conducting their screening protocols (such as a one-step versus two-step screening approach as used in
the two hospitals in this study), as that may influence the pass/fail rate. The protocol that NBHS sites use as a pass/fail criteria should be looked at as well. Getting all of this information would help determine areas of weakness in NBHS policy. This would help result in solutions that could be universally applied to guide policy making, such as ensuring accurate entry of information (e.g., for CFA and its specific subtypes).

In the current study, 17.41% \( (n = 51) \) of infants who failed the NBHS were lost to follow-up. This rate is less than the national average of 31.3% (Subbiah et. al., 2018), presumably due to audiology oversight of the NBHS programs described in this paper. There have been studies that explore reasons for loss to follow-up in newborn hearing screening programs (ASHA, 2008), however, there are no specific protocols in place for guiding these programs. It is important to better understand the underlying causes of loss to follow-up rates so that JCIH can make recommendations and guidelines for improving service to infants and families.

**Conclusions**

Results from this study revealed that children with CFAs were six times more likely to fail their NBHS when compared to the fail rate for all infants at the two participating hospitals.

Audiological follow-up and monitoring is not warranted for infants with preauricular sinuses and tags unless the infant exhibits other features associated with a syndrome that has an associated risk for hearing loss. It is important to document these anomalies in the state EHDI database so that the medical home can monitor for any additional signs and/or symptoms associated with such syndromes and make appropriate referrals to other medical professionals.

Because the NBHS fail rates in this study varied greatly for the different CFAs, further research should be completed to determine if these findings can be replicated. If so, JCIH should consider updating the list of risk factors for hearing loss to delineate the current CFA category into different subgroups. Along with updating the list of risk factors. JCIH should also consider updating the recommendations for each specific CFA, as infants in this risk factor group should be followed based on their specific anomaly rather than the group as a whole.

Results from the two surveys in this study demonstrate that NBHS programs are not recording and documenting risk factor information adequately and consistently. If our findings are representative of the other state EDHI programs that did not respond to the survey, this would suggest a need for changes to the existing NBHS protocol to include programs that train staff to identify and document risk factors for hearing loss. This training would improve the chances that risk factors for hearing loss are being identified and properly documented. Correct documentation of risk factors would also provide medical professionals the information they need to appropriately refer infants for follow-up evaluations, monitoring, and early intervention services so that they have the best opportunity to maximize their potential. This is imperative because, without appropriate and timely referrals, children are more likely to fall behind their hearing peers in communication, cognition, reading, and social-emotional development (JCIH, 2007).

Lastly, it would be advantageous to change the NBHS guidelines to protocols to ensure consistency across all programs. It is crucial that all NBHS programs throughout the country follow the same procedures to improve recommendations for follow-up care in a timely manner.

**References**

http://www.asha.org/Practice-Portal/Professional-Issues/Newborn-Hearing-Screening


Appendix A

EHDI State Leader Survey

1) If you agree with the informed consent above, please add a signature.

2) Which state do you represent?

3) Does your state mandate newborn hearing screenings in all, or most, birthing facilities?
   a) Yes
   b) No

4) Do facilities in your state submit newborn hearing screening data for infants born at that facility?
   a) Yes, most facilities do
   b) Some facilities do, some facilities do not
   c) No, most facilities do not

5) If an infant is identified with a specific risk factor associated with childhood hearing loss including: family history of hearing loss, NICU stay of greater than 5 days, hyperbilirubinemia with exchange transfusion, ototoxic medications for greater than 5 days, asphyxia or hypoxic ischemic encephalopathy, ECMO, in-utero infections, craniofacial anomalies, syndromes associated with hearing loss, and significant head trauma (re: JCIH 2019 Risk Factor Indicators), are facilities in your state required to document this information in the state EHDI database?
   a) Yes
   b) No

6) Do facilities in your state submit this documentation, as required (re: question 5)?
   a) Yes, most facilities do
   b) Some facilities do, some facilities do not
   c) No, most facilities do not

7) If an infant is identified as having a craniofacial anomaly, do facilities in your state record the specific anomaly present (e.g., cleft lip and palate, preauricular sinus and tags, microtia, atresia, and/or syndromes such as Trisomy 21, Treacher Collins syndrome, CHARGE Association, Crouzon disease, Klippel-Feil syndrome, Goldenhar syndrome, Pierre Robin syndrome, etc.)?
   a) Yes, most facilities do
   b) Some facilities do, some facilities do not
   c) No, most facilities do not

8) If a craniofacial anomaly risk factor is reported, but the specific anomaly is not recorded, do you request additional information from the program coordinator about the specific craniofacial anomaly?
   a) Yes
   b) No

9) Are you satisfied with the documentation completed by the facilities in your state regarding the risk factors for hearing loss?
   a) Yes
   b) No

10) Do you have any suggestions for how to improve the recording, reporting or follow-up process for infants with risk factors for hearing loss, including infants with craniofacial anomalies?

11) Would you like the final results of the study sent to you?
    a) Yes, electronically
    b) Yes, via mail
    c) No

12) If ‘yes’, where should the results be sent? (email address or mail address)
Appendix B
Newborn Hearing Screening Program Coordinator Survey

1) If you agree with the informed consent above, please add a signature.

2) What is your profession?

3) In which state is your facility located?

4) Who performs the newborn hearing screenings? (Mark all that apply)
   a) Audiologists
   b) Nurses/Nurse Techs
   c) Technicians hired specifically to perform the screenings
   d) Other; please specify in the next question

5) If your answer was ‘other’ from the previous question, please specify.

6) If an audiologist does not perform the hearing screenings, does your facility employ an audiologist to oversee the program?
   a) Yes
   b) No
   c) N/A

7) Is your staff trained to identify the different risk factors associated with childhood hearing loss including: family history of hearing loss, NICU stay of greater than 5 days, hyperbilirubinemia with exchange transfusion, ototoxic medications for greater than 5 days, asphyxia or hypoxic ischemic encephalopathy, ECMO, in-utero infections, craniofacial anomalies, syndromes associated with hearing loss, and significant head trauma (re: JCIH 2019 Risk Factor Indicators)?
   a) Yes
   b) No

8) Is information regarding risk factors for childhood hearing loss collected either prior to or following each newborn hearing screening? (if no skip to question 14).
   a) Yes
   b) No

9) If yes (re: question 8), how is this information collected? (Mark all that apply)
   a) Ask hospital staff about risk factors
   b) Check infant’s medical records (e.g., admission reports, lab reports, physician, nurse and/or social worker assessments, etc.)
   c) Ask the mother case history questions at the time of the hearing screening
   d) Other

10) Does your program record the type of risk factor(s) in the hospital medical records (re: question 8)?
    a) Yes
    b) No

11) If an infant is identified as having one of the risk factors (re: question 7), does your program report this information to your state Early Hearing Detection and Identification (EHDI) program?
    a) Yes
    b) No

12) If an infant is identified as having a craniofacial anomaly, does your program record the specific type of craniofacial anomaly present (e.g., cleft lip and palate, preauricular sinus and tags, microtia, atresia, and/or syndromes such as Trisomy 21, Treacher Collins syndrome, CHARGE Association, Crouzon disease, Klippel-Feil syndrome, Goldenhar syndrome, Pierre Robin syndrome, etc.)?
    a) Yes
    b) No
Appendix B (cont.)

13) If you answered ‘yes’ for the previous question, do you report the specific craniofacial anomaly to the state EHDI program?
   a) Yes
   b) No

14) Do you feel that you are getting enough guidance from your state EHDI program on how to document and report risk factors, including craniofacial anomalies?
   a) Yes
   b) No

15) Are you able to access the newborn hearing screening pass/fail data for infants with craniofacial anomalies tested at your facility?
   a) Yes
   b) No

16) If you answered ‘yes’ for the previous question, what is the refer rate for these infants for the period of January 2017 through December 2019?

17) Would you like the final results of the study sent to you?
   a) Yes, electronically
   b) Yes, via mail
   c) No

18) If ‘yes’, where should the results be sent? (email address or mail address)
### Table C1

**Profession of Participating Newborn Hearing Screening (NBHS) Program Coordinators**

<table>
<thead>
<tr>
<th>Profession</th>
<th>Total Percentage (n)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Nurse</td>
<td>54.44% (49)</td>
</tr>
<tr>
<td>Audiologist</td>
<td>37.66% (29)</td>
</tr>
<tr>
<td>NBHS Coordinator</td>
<td>4.44% (4)</td>
</tr>
<tr>
<td>Nurse Practitioner</td>
<td>1.11% (1)</td>
</tr>
<tr>
<td>Operations Coordinator</td>
<td>1.11% (1)</td>
</tr>
<tr>
<td>Perinatal Safety Specialist</td>
<td>1.11% (1)</td>
</tr>
<tr>
<td>Unit Secretary</td>
<td>1.11% (1)</td>
</tr>
<tr>
<td>Practice Manager</td>
<td>1.11% (1)</td>
</tr>
<tr>
<td>Administrative Assistant</td>
<td>1.11% (1)</td>
</tr>
<tr>
<td>Hearing Technician</td>
<td>1.11% (1)</td>
</tr>
<tr>
<td>Did Not Answer</td>
<td>1.11% (1)</td>
</tr>
</tbody>
</table>

### Table C2

**Profession of Those Conducting NBHS**

<table>
<thead>
<tr>
<th>Profession</th>
<th>Total Facilities</th>
</tr>
</thead>
<tbody>
<tr>
<td>Audiologists</td>
<td>20/90 facilities</td>
</tr>
<tr>
<td>Nurses</td>
<td>61/90 facilities</td>
</tr>
<tr>
<td>Technicians (hired specifically for the hearing screening)</td>
<td>25/90 facilities</td>
</tr>
<tr>
<td>Others: Physician assistants, nurse practitioners, trained volunteers and student interns</td>
<td>4/90 facilities</td>
</tr>
</tbody>
</table>

*Note. NBHS= Newborn Hearing Screening*

*aSome facilities employ multiple professionals to perform the screenings.*

### Table C3

**States Represented in the Newborn Hearing Screening Program Coordinator Survey**

<table>
<thead>
<tr>
<th>Profession</th>
<th>Total Percentage (n)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Arkansas</td>
<td>2.22% (2)</td>
</tr>
<tr>
<td>California</td>
<td>1.11% (1)</td>
</tr>
<tr>
<td>District of Columbia</td>
<td>2.22% (2)</td>
</tr>
<tr>
<td>Illinois</td>
<td>1.11% (1)</td>
</tr>
<tr>
<td>Louisiana</td>
<td>1.11% (1)</td>
</tr>
<tr>
<td>Maryland</td>
<td>12.22% (11)</td>
</tr>
<tr>
<td>Michigan</td>
<td>36.67% (33)</td>
</tr>
<tr>
<td>Missouri</td>
<td>1.11% (1)</td>
</tr>
<tr>
<td>North Carolina</td>
<td>1.11% (1)</td>
</tr>
<tr>
<td>Nebraska</td>
<td>14.44% (13)</td>
</tr>
<tr>
<td>New Jersey</td>
<td>6.67% (6)</td>
</tr>
<tr>
<td>New York</td>
<td>3.33% (3)</td>
</tr>
<tr>
<td>Nevada</td>
<td>1.11% (1)</td>
</tr>
<tr>
<td>Ohio</td>
<td>5.55% (5)</td>
</tr>
<tr>
<td>Oregon</td>
<td>1.11% (1)</td>
</tr>
<tr>
<td>South Carolina</td>
<td>1.11% (1)</td>
</tr>
<tr>
<td>Tennessee</td>
<td>1.11% (1)</td>
</tr>
<tr>
<td>Virginia</td>
<td>5.55% (5)</td>
</tr>
<tr>
<td>Blank Responses</td>
<td>1.11% (1)</td>
</tr>
</tbody>
</table>