

Implementing a Two-Class System for Monitoring Risk Factors for Delayed-Onset Hearing Loss

Jessica Stich-Hennen, AuD, PASC¹
Gabriel Anne Bargaen, PhD, CCC-A/SLP²

¹Idaho Elks Hearing and Balance Centers
²Idaho State University

Abstract

Purpose: This manuscript discusses the importance of establishing risk indicator monitoring guidelines for state Early Hearing Detection and Intervention programs.

Method: Idaho Sound Beginnings (ISB) implemented a guideline which divided risk indicators associated with delayed-onset hearing loss into two classes (Class A and Class B). From 2012–2013, the incidence of delayed-onset hearing loss in the presence of Class A and Class B risk indicators were evaluated. For Class B risk indicators, ototoxic medication exposure and family history were analyzed.

Results: Of the 10,634 infants born, 1,175 were found to have passed the newborn hearing screening and have at least one risk indicator. Of the infants evaluated with Class A risk indicators, 21 children had an educationally significant hearing loss. Of the 345 children who received ototoxic medications, 55 children were diagnosed with educationally significant hearing loss. An educationally significant hearing loss was found in 10 children who returned for diagnostic evaluation who had family history of childhood hearing loss.

Conclusion: ISB's risk monitoring classification system has enhanced Idaho's EHD program by early identification of children who are at higher risk for delayed-onset hearing loss. Early identification has ultimately led to early intervention.

Key Words: JCIH, risk indicators, hearing loss, infant, Idaho Sound Beginnings

Acronyms: AABR = automatic auditory brainstem response; ABR = auditory brainstem response; CMV = cytomegalovirus; ECMO = extracorporeal membrane oxygenation; EDHI = Early Hearing Detection and Intervention; ISB = Idaho Sound Beginnings; JCIH = Joint Committee on Infant Hearing; NICU = neonatal intensive care unit; OAE = otoacoustic emissions

Correspondence concerning this article should be addressed to: Gabriel Anne Bargaen, Communication Sciences & Disorders, Idaho State University, Meridian Health Science Center, 1311 E Central Drive, Meridian, ID 83642.
Email: barggabr@isu.edu

The Joint Committee on Infant Hearing (JCIH) was established in 1969 to investigate the need for mass screening programs in an effort to identify children with hearing loss earlier in life. In 1973, the JCIH recommended using criteria to identify newborns at risk for hearing loss. Over the next five position statements, JCIH modified the criteria based on research and clinical findings. The suggested audiological monitoring schedule ranged from strict (e.g., monitor hearing every six months until the age of 3; JCIH, 2000) to lax (e.g., at least one diagnostic evaluation by 24–30 months of age; JCIH, 2007). The variability in the monitoring schedules has the potential to create confusion for physicians and audiologists. The JCIH 2007 Position Statement recommended earlier and more frequent monitoring for some risk indicators with higher prevalence of delayed-onset hearing loss. For others, the JCIH 2007 Position Statement indicates one monitoring appointment by at least 24–30 months of age may be sufficient. To efficiently implement these recommendations, the use of a systematic approach may be appropriate.

In 2011, Idaho Sound Beginnings (ISB) developed a classification system of the 2007 JCIH risk indicators to provide guidance to those involved with newborn hearing screening programs regarding when to refer infants to pediatric audiologists for risk monitoring of delayed-onset hearing loss. This article will discuss the development of this classification system for the JCIH (2007) risk indicators and initial findings after implementation within two hospitals.

Introduction

Monitoring JCIH 2007 Risk Indicators

JCIH published the most recent position statement in 2007. As shown in Table 1 the statement listed risk indicators associated with permanent congenital, delayed-onset, or progressive hearing loss in childhood. The neonatal risk indicators were redefined to specifically include neonatal intensive care of more than five days or any of the following regardless of length of stay: Extracorporeal membrane

oxygenation (ECMO), assisted ventilation, exposure to ototoxic medications (gentamycin and tobramycin) or loop diuretics (furosemide/Lasix), and hyperbilirubinemia that requires exchange transfusion (JCIH, 2007). The monitoring schedule was also redefined to include at least one diagnostic audiology assessment by 24 to 30 months of age. Additionally, the time and number of hearing evaluations for children with risk indicators should be customized and individualized (JCIH, 2007), giving the clinical judgement back to the audiologists and the medical home. Earlier and more frequent assessments may be indicated for some risk indicators with higher prevalence of delayed-onset hearing loss. For others, one diagnostic appointment may be sufficient.

The following risk indicators were listed in the JCIH 2007 Position Statement as being of greater concern for delayed-onset hearing loss: caregiver concern, family history of hearing loss, ECMO, cytomegalovirus (CMV), syndromes associated with hearing loss, neurodegenerative disorders, culture-positive postnatal infections associated with sensorineural hearing loss, head trauma, and chemotherapy. When considering only those infants in the at-risk population who were diagnosed with hearing loss, Hall (2007) reviewed the frequency of occurrence of individual risk indicators associated with hearing loss and identified six risk indicators that occurred most frequently (as shown in Table 2). Hall (2007) highlighted the importance of audiological professionals who work with

Table 1
JCIH 2007 Position Statement Risk Indicators

Caregiver concern regarding hearing, speech, language, or development delay
Family history of permanent childhood hearing loss
Neonatal intensive care of more than 5 days or any of the following regardless of length of stay: extracorporeal membrane oxygenation (ECMO), assisted ventilation, exposure to ototoxic medications (gentimycin and tobramycin) or loop diuretics (furosemide/Lasix), and hyperbilirubinemia that requires exchange transfusion
In utero infections, such as cytomegalovirus (CMV), herpes, rubella, syphilis, and toxoplasmosis
Craniofacial anomalies, including those that involve the pinna, ear canal, ear tags, ear pits, and temporal bone anomalies
Physical findings, such as white forelock, that are associated with a syndrome known to include a sensorineural or permanent conductive hearing loss
Syndromes associated with hearing loss or progressive or delayed-onset hearing loss, such as neurofibromatosis, osteopetrosis, and Usher syndrome; other frequently identified syndromes including Waardenburg, Alport, Pendred, and Jervel and Lange-Nielson
Neurodegenerative disorders, such as Hunter syndrome, or sensory motor neuropathies, such as Friedreich ataxia and Charcot-Marie-Tooth syndrome
Culture-positive postnatal infections associated with sensorineural hearing loss, including confirmed bacterial and viral (especially herpes viruses and varicella) meningitis
Head trauma, especially basal skull/temporal bone fracture that requires hospitalization
Chemotherapy

Table 2
Individual Risk Indicators Associated with Hearing Loss Occurring Most and Least Frequently (Hall, 2007)

Most frequent	Least frequent (< 10%)
Craniofacial anomalies (> 50%)	Low birth weight
Family history of childhood hearing loss (> 15%)	Hyperbilirubinemia
Severe asphyxia (> 15%)	Ototoxic medications
Congenital infections (> 15%)	ECMO
Mechanical ventilation (> 10%)	Substance abuse (maternal)
Bacterial meningitis (> 10%)	

Note. ECMO = extracorporeal membrane oxygenation.

the infant population being intimately familiar with all risk indicators for hearing loss whether peripheral, auditory dysfunction, or delayed onset.

In 2012, Beswick, Driscoll, and Kei systematically identified 753 publications from 1973 to March 2011 and reviewed 40 of those publications to draw evidence-based conclusions on risk indicators and risk monitoring programs that detect postnatal hearing loss. They found the most common risk indicators reported were “gestational-age, low-birth weight, toxoplasmosis, other infections, rubella, CMV, herpes simplex virus infections, craniofacial anomalies, respirator support, and the administration of aminoglycosides” (p. 745). Based on two of the publications reviewed, 3 to 3.5% of infants were referred for follow-up testing due to the presence of risk indicators defined by each study. Authors found a strong relationship between postnatal hearing loss and CMV, congenital diaphragmatic hernia, ECMO, and persistent pulmonary hypertension. Conversely, a weak link was found between postnatal hearing loss and toxoplasmosis, pre-auricular skin tags and ear pits, and low birth weight.

A retrospective study by Beswick, Driscoll, Kei, Khan, and Glennon (2013) evaluated audiological findings for 2,107 children who were identified with one or more risk indicators for hearing loss. Of children who initially passed the newborn hearing screening but had risk indicator(s), 2.7% were diagnosed with hearing loss. A statistical analysis identified family history and craniofacial anomalies to be high predictors for postnatal hearing loss, whereas, low birth weight was a low predictor.

Wood, Davis, and Sutton (2013) retrospectively examined the effectiveness of targeted surveillance to identify moderate-profound permanent childhood hearing impairment in babies who passed the newborn hearing screening in the presence of risk indicators for delayed-onset hearing loss in England. England newborn hearing screening program data ($n = 2,307,880$ children) was reviewed from 2006 to 2009. Based on follow-up evaluation of more than 38,000 infants who passed newborn hearing screening with risk factor for delayed-onset hearing loss, five factors were identified as most often associated with permanent childhood hearing impairment: syndrome (other than Down's) associated with a hearing loss, neonatal intensive care unit (NICU) with refer in both ears at otoacoustic emissions (OAE) and pass in both ears at automatic auditory brainstem response (AABR), craniofacial anomaly, Down's syndrome, and congenital infection. Monitoring only these five criteria was estimated to reduce the percentage of the birth population that require targeted surveillance from 3% to 0.25% (Wood et al., 2013). It was also noted that neonatal bacterial meningitis and aminoglycoside antibiotics were not considered in this review. Bacterial meningitis occurring before the hearing screen is considered a contraindication to screening and patients are referred directly for a full audiological assessment. The protocol of England's newborn hearing screening program states that babies who

receive aminoglycoside and have blood levels exceeding the therapeutic range should be referred for audiological assessment by the prescribing pediatrician. Otherwise, screening programs in England no longer record aminoglycoside as a risk factor.

Kraft, Malhotra, Boerst, and Thorne (2014) evaluated the economic impact of monitoring children with risk indicator for delayed-onset hearing loss. University of Michigan newborn hearing screening program data was reviewed from 2001 to 2007. Ninety children were diagnosed with hearing loss, including 16 children with delayed-onset hearing loss. They concluded that a “NICU stay of greater than 5 days and exposure to loop diuretics were not associated with an increased risk of either congenital or delayed-onset hearing loss” (p. 1842). Monitoring children with these risk indicators, NICU length of stay greater than five days, or exposure to potentially ototoxic medications, in the absence of other risk indicators was reported to have “increased the monitoring burden” nearly five times which “contributes to the high cost of screening per case identified” (p. 1842).

Vos, Senterre, Lagasse, SurdiScreen Group, and Levêque (2015) retrospectively evaluated the clinical management and follow-up of newborns with neonatal risk indicators of hearing loss for the newborn screening program in Belgium to systematically update the monitor recommendations. The group completed a literature review of 15 years of publications and graded the quality of evidence found in regard to the risk indicators for delayed-onset hearing loss as defined by the 2000 JCIH Position Statement and the clinical experience of professionals from the Fédération Wallonie-Bruselles. The study found congenital infections (i.e., cytomegalovirus, toxoplasmosis, and syphilis), a family history of hearing loss, consanguinity, malformation syndromes, and fetal alcohol syndrome to have a high level of evidence quality as neonatal risk indicators for hearing loss. Additionally, hyperbilirubinemia had a moderate level of evidence quality while very low birth weight, low Apgar score, ototoxic drugs, and hospitalization in the NICU had a very low or low level of evidence quality. Vos et al. recommended monitoring all risk indicators for hearing loss, even those with weak evidence, in order to avoid “unidentified neonatal hearing loss” (p. 6). The authors also recommended completing the initial hearing evaluation for those newborns with any of these risk indicators prior to discharge from the hospital using at least an auditory brainstem response (ABR) to assess the entire auditory brainstem pathway.

Review of current literature on risk indicators for delayed-onset hearing loss revealed variability in which risk indicators should be monitored, which risk indicators have increased risk for delayed-onset hearing loss, and variability on how and when to monitor individual risk indicators. The JCIH 2007 Position Statement provided guidance for Early Hearing Detection and Intervention (EDHI) programs on which risk indicators to monitor and which risk indicators have increased risk for delayed-

onset hearing loss. Unfortunately, the JCIH 2007 Position Statement did not provide concrete guidance on when to begin monitoring (i.e., what age), how often to monitor (i.e., months vs. years), and how long to continue monitoring (i.e., until what age). The purpose of the ISB risk monitoring classification system was to provide guidance to stakeholders in Idaho newborn hearing screening programs regarding when to refer infants for risk monitoring of delayed-onset hearing loss.

Idaho Monitoring Risk Indicators

In October 2011, with guidance from the ISB pediatric audiology consultant, hospitals with NICU programs in Idaho began collecting data on early and more frequently monitored risk indicators. Indicators with higher incidence and earlier onset of hearing loss were classified as Class A and all other risk indicators were classified as Class B. This classification system was based on the JCIH 2007 Position Statement and evidence-based research on craniofacial anomalies. The JCIH 2007 Position Statement and additional studies (Vos et al., 2015; Beswick et al., 2013; Wood et al., 2013) identified early and more frequent assessment may be indicated for children with CMV infection; syndromes associated with progressive hearing loss, neurodegenerative disorders, trauma, or culture-positive postnatal infections associated with sensorineural hearing loss; and for children who have received ECMO or chemotherapy. Those risk indicators were designated Class A. Cleft palate was also included in the Class A category based on evidence-based research on craniofacial anomalies from multiple publications (Beswick et al., 2013; Helias, Chobaut, Mourot, & Lafon, 1988; Paradise, 1975; Potsic, Cohen, Randall, & Winchester, 1979; Viswanathan, Vidler, & Richard, 2008; Yules, 1970). All other risk indicators identified by the JCIH 2007 Position Statement were categorized in Class B including family history of childhood hearing loss, other in-utero infections (not CMV), NICU stay of greater than five days, any amount of ototoxic exposure, any amount of mechanical ventilation, and other craniofacial anomalies excluding cleft palate (Kraft et al., 2014; Wood et al., 2013). See Figure 1 for Class A and Class B lists.

The terminology of Class A and B were defined based on a collaborative effort between a neonatologist and a pediatric audiology consultant. The Class terms are commonly used within the NICU environment and readily identified by the medical community. Infants with Class A risk indicators were recommended for evaluations by a pediatric audiologist by 3 months of age. At a minimum, the evaluation should include diagnostic ABR. Infants with Class B risk indicators were recommended for a behavioral hearing evaluation by a pediatric audiologist by 1 year of age. Guidelines provided to Idaho pediatric audiologists indicate, at a minimum, the evaluation should include ear specific measurements at multiple frequencies as recommended by the JCIH 2007 Position Statement when evaluating a child 6 to 36 months of age

The purpose of the risk indicator classification system is to allow for early identification of children with delayed-onset hearing loss. In Idaho, when a child is identified with hearing loss the diagnosing audiologist completes the ISB audiology results form (Figure 2) and submits it to ISB. This form also serves as a release of information to early intervention programs within the state of Idaho including Infant Toddler Program, Idaho Education Services for the Deaf and the Blind, and Idaho Hands and Voices. Therefore, the risk indicator classification system, subsequent early diagnosis of hearing loss, and the ISB reporting process should lead to timely enrollment in early intervention.

Method

ISB, Idaho's EHDI program, has been collecting data on risk indicators since the implementation of the Idaho EHDI program using Hi*Track data collection system. Implementation of the Class A and Class B classification system did not alter how data within Hi*Track was collected or maintained. Hi*Track allows for retrospective analysis of risk indicators based on the two-class classification system. Idaho birthing hospitals report information regarding risk indicators with results of each newborn hearing screening. A positive family history of childhood hearing loss is self-reported by families. The presence of other risk indicators is identified from a review of the infant's medical chart. Diagnostic audiological findings are reported to ISB by Idaho audiologists using the ISB audiology results form (Figure 2).

Data of infants born from January 2012 through December 2013 for two of the larger hospitals with NICU programs in the state of Idaho were reviewed. Data on infants who passed the newborn hearing screening and were identified as having one or more risk indicators were included in the review. Infants who referred on the newborn hearing screening and had present risk indicators were excluded from the study. Data was collected for analysis in November 2015, all diagnostic audiological follow-up information reported to ISB at that time was available for review.

Results

According to ISB Hi*Track, 10,634 infants were born at the two selected hospitals in Idaho during this time frame. Of the 10,634 infants reviewed, 1,175 (11.04%) infants were found to have passed the newborn hearing screening and have at least one risk indicator in either Class A or Class B. From these infants, 175 (1.6%) infants were found to have passed the newborn hearing screening and have at least one Class A risk indicator. Infants within the first group of Class A risk indicators could also be represented in the second group of either Class A or Class B risk indicators. Infants with Class A risk indicators frequently have at least one risk indicator from the Class B list which accounts



Guidelines for Risk Monitoring for Delayed Onset Hearing Loss

Class A: Risk indicators

- *In-utero infections (congenital CMV)
- *Culture Positive postnatal infection (Bacterial and viral meningitis)
- *Syndromes associated with progressive or delayed onset hearing loss (Neurofibromatosis, Osteopetrosis, Usher Syndrome, Townes-Brock)
- *Syndromes associated with hearing loss (Down syndrome and Sticklers)
- *Cleft Lip/Palate
- *ECMO assisted ventilation
- *Head Trauma involving basal skull/temporal fracture that requires hospitalization
- *Chemotherapy treatments
- *Neurodegenerative disorders or sensory motor neuropathies

If baby passes the newborn hearing screening & has one or more CLASS A risk indicator =
Recommendation for diagnostic ABR evaluation with pediatric audiologists by 3 months of age.

Class B: Risk indicators

- *Family history of childhood hearing loss
- *In-Utero Infection (Herpes, Rubella, Syphilis, Toxoplasmosis)
- *NICU stay of greater than 5 days
- *Any amount of ototoxic exposure (aminoglycosides)
- *Any amount of mechanical ventilation
- *Craniofacial anomalies involving pinna, ear canal, ear pits and temporal bone anomalies

If baby passes the newborn hearing screening & has one or more CLASS B risk indicators =
Recommendation for diagnostic pediatric hearing evaluation by 1 year of age.

NOTE: If baby REFERS on the newborn hearing screening after two attempts – Recommendation for Diagnostic ABR evaluation to be completed by 3 months of age (JCIH 2007)

- * Any parental/caregiver hearing concerns warrants a referral to a pediatric audiologist.
- ** Infants readmitted to the hospital within the first 30 days of life should be re-screened if any risk indicators are present.

References:

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Joint Committee on Infant Hearing. Year 2007 Position Statement: Principles and Guidelines for Early Hearing Detection and Intervention Programs. *Pediatrics*. 2007; 120(4):898–921. doi: 10.1542/peds.2007-2333.
Van Riper, Lori A.; Kileny, Paul R. ABR Hearing Screening for High-Risk Infants. *American Journal of Otology*. 20(4):516-521, July 1999.



IDAHO SOUND BEGINNINGS (ISB)

Early Hearing Detection and Intervention
Department of Health and Welfare, Infant Toddler Program

FAX TO (208) 332-7331

Within 5 days

Early
Hearing
Screening
Referral Form

Complete Form for All: Refers ☐ Risks ☐ Transfers* ☐ Missed ☐ or Incomplete ☐

Birth Hospital: _____

(*Transfers only) Receiving Hospital: _____ [Please Press Firmly]

Within **5 days** of screening or discharge—Distribute copies to: Audiologist - White ISB - Gold Hospital - Pink Parent - Green Physician - Yellow

Send to: Idaho Sound Beginnings-EHDI, 450 W State St 5th Fl Boise, ID 83702 or Fax: (208) 332-7331

1. BABY'S INFORMATION:

Baby's Med Record #: _____

Baby's Name: _____
Last First

DOB: ____/____/____ Gender: ☐ M ☐ F

Nursery: ☐ Well Baby ____ Number of days in NICU/PICU

Baby's Primary Physician/Clinic: _____

Mother's name: _____

2. CONTACT INFORMATION:

Parent/Guardian: _____
Last First

Address: _____

City: _____ State: _____ Zip: _____

Main Phone: _____ Text? _____

Alternate Phone/Contact: _____

Email/other contact: _____

3. HEARING SCREEN RESULTS:

First Screen: R ☐ Pass ☐ Refer ☐ No Result

L ☐ Pass ☐ Refer ☐ No Result

Date

Second Screen: R ☐ Pass ☐ Refer ☐ No Result

L ☐ Pass ☐ Refer ☐ No Result

4. RISK ASSESSMENT (check all that apply)

FOR LATER-ONSET CHILDHOOD HEARING LOSS:

____ Family History of Permanent Hearing Loss <18 yrs of age

____ NICU stay >5 days

____ Syndrome Associated with HL (e.g. Downs)

____ Congenital Infection (e.g. T-O-R-C-H)

____ Postnatal Infection (e.g. Meningitis)

____ Craniofacial Anomalies- _____

____ Ototoxic Medications - any amount

____ Mechanical Ventilation - any amount

____ Parent or Physician Concern

____ Head Trauma ____ Other _____

(monitoring through age 3 is recommended for most risk factors)

If you have any questions about testing, or need information on financial assistance,
please contact Idaho's Early Hearing Program, Idaho Sound Beginnings, at (208) 334-0829.

☐ Your baby **REFERRED** on the hearing screen. Diagnostic testing needs to be completed before baby is **3 months** old. If baby is not hearing **all** the sounds necessary for speech and language development, early identification can minimize communication delays.

☐ Your baby is **AT RISK for later-onset childhood hearing loss**. Diagnostic testing at approximately **9 -12 months** of age is recommended for most risk factors. A Pediatric Audiologist can advise on the appropriate monitoring schedule for your baby.

Your Follow-Up Appointment:

Clinic: _____

Phone: _____

Appt Date: _____ Time: _____

(For a listing of Pediatric Audiologists visit
www.EHDI-PALS.org)

I have been informed of my baby's hearing screen results and of the need for diagnostic audiology (hearing) testing before the age of 3 months (if baby did not pass) to determine if a hearing loss is present. If baby passed the hearing screen, but risk factors are present (see above), hearing testing is recommended at approximately 9 months of age. (American Academy of Pediatrics (AAP) Guidelines)

I hereby give permission to the staff of the above-named hospital/screening site to release medical information necessary to complete an audiology evaluation for my child to the listed audiologist/clinic (or the audiologist of my choice) and physician. I also give permission to the hospital and audiologist/clinic, and Idaho Sound Beginnings to share the results of the hearing screening, diagnostic audiology evaluations, and early intervention choices (if any) with the above-named physician, the Idaho Infant-Toddler Program, Idaho School for the Deaf and Blind, Idaho Hands & Voices, and other states' EHDI Coordinators, if needed.

I understand that the information will only be used to ensure that appropriate and timely medical, educational, and audiologic services are made available to my child.

Hearing screening results are reported to Idaho Sound Beginnings -Idaho's Early Hearing Detection & Intervention Program and are not shared with the above listed entities or any other outside entities without parent/guardian consent.

I have had the opportunity to read this clinic's Notice of Privacy Practices. I understand that this information will not be shared with unauthorized individuals. This authorization expires 36 months from the date signed.

PARENT/GUARDIAN : _____ DATE : _____

7/15

Figure 2. Idaho Sound Beginnings audiology results form.

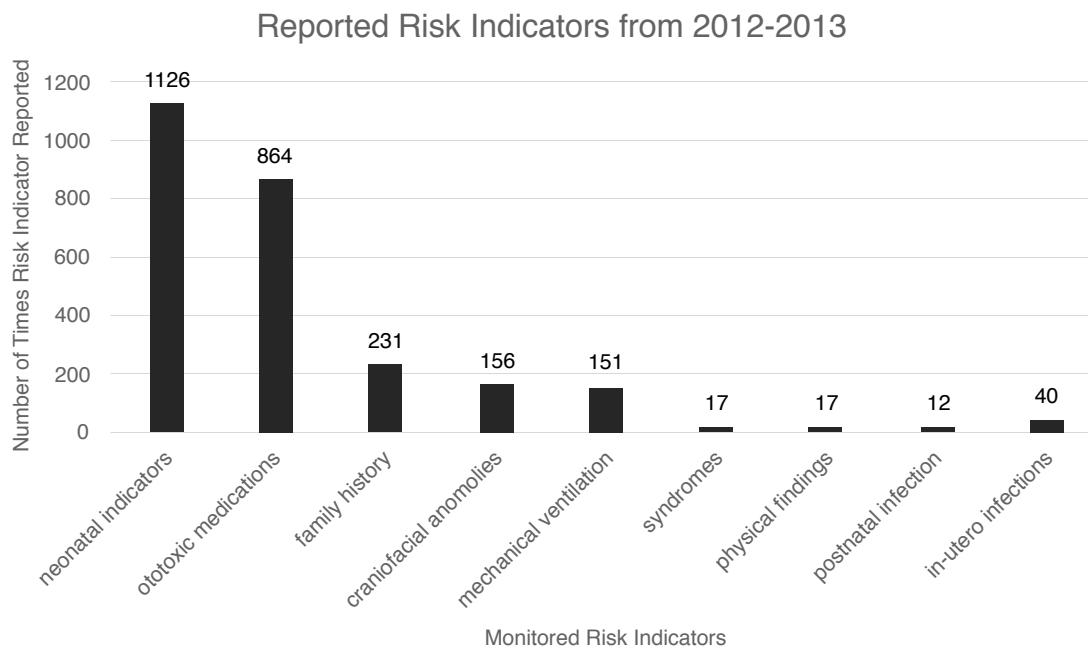


Figure 3. Reported risk indicators from Idaho Sound Beginnings Hi*Track database in 2012 - 2013

for the crossover of the two groups. The number of risk indicators reported from this population totaled 2,614 (Figure 3).

Of the 175 infants with Class A risk indicators, 87 returned for comprehensive diagnostic audiology evaluations. Of those, 25% (21 of 87 infants) were found to have an educationally significant hearing loss. Educationally significant hearing loss is defined as any type of hearing loss (chronic conductive, sensorineural, or mixed hearing loss), unilateral or bilateral, at 25 dB or greater (worse) at one or more frequencies. Of the 25% that were diagnosed with hearing loss, five (5.7%) were found to have sensorineural or mixed hearing loss. The Class A risk indicators present in these five children included two children with cleft palate, one child with Townes Brock syndrome, one child with Acrofacial Dysostosis, and one child with congenital CMV. The most common Class A indicators present in this population were syndromes (e.g., Down's Syndrome) and cleft palate.

Of the 1,175 infants who passed the newborn hearing screening and were identified as having at least one risk indicator within Class A and Class B categories, 743 infants received ototoxic medication, most commonly gentamicin. Of those infants who received ototoxic medication, 345 (46.4%) returned for follow-up diagnostic audiological evaluation and 55 (15.9%) were diagnosed with educationally significant hearing loss, five (1.4%) of which were sensorineural or mixed hearing loss and 50 (14.4%) were chronic conductive hearing loss. Of the five infants diagnosed with sensorineural or mixed hearing loss, three presented with Class A risk indicator along with the Class B ototoxic medication exposure. The remaining two infants presented with additional Class B indicators along with ototoxic medication exposure, specifically extended NICU stay and prematurity.

Over half of the infants ($n = 398$; 53.6%) who were identified as having the Class B risk indicator of ototoxic medication exposure were lost to follow-up. If the trend found for this population can be generalized to the children who did not receive follow-up evaluations, then approximately 63 infants have educationally significant hearing losses and did not benefit from early diagnosis and intervention.

Of the 1,175 infants who passed the newborn hearing screening and were identified as having at least one risk indicator, 175 infants were identified as having present family history of permanent childhood hearing loss which is a Class B risk indicator. Of the 175 infants, 65 (37.1%) returned for diagnostic audiology evaluation for a lost to follow-up rate of 62.8%. This was the highest lost to follow-up rate of all the risk indicators present within this population. An educationally significant hearing loss was found in 10 (16%) of those infants that returned for diagnostic evaluation, three of which were sensorineural or mixed hearing loss and seven were chronic conductive hearing loss. Additional risk indicators were present in only one of the three infants diagnosed with sensorineural or mixed hearing loss, indicating that family history of permanent childhood hearing loss was the only indicator to assist with early identification of hearing loss in these two infants.

Age of diagnosis was reviewed in all children identified with educationally significant hearing loss in the population. Children with educationally significant hearing loss were identified prior to 24 months of age. Children identified with Class A risk indicators returned for initial audiological evaluation at an average of 4 months of age (range = 2 to 9 months). Children identified with Class B risk indicators returned for initial audiological evaluation at an average

of 11.3 months of age (range = 9 to 13 months). The nine children found to have sensorineural or mixed hearing loss were diagnosed at an average of 12.6 months of age (range = 4 to 24 months).

Discussion

ISB's goal of using the risk monitoring classification system was to identify infants with the higher risk for delayed-onset hearing loss (i.e., Class A) and refer them to audiology for earlier and more frequent monitoring per JCIH (2007) recommendation. The infants with Class B indicators who have lower incidence of delayed-onset hearing loss would warrant less frequent monitoring. In this study, children with a Class A risk indicator and delayed-onset hearing loss were seen for initial evaluation by 10 months old (range = 2 to 9 months) and diagnosed by 25 months of age (range = 4 to 24 months). One infant who was diagnosed at age 24 months was monitored for delay-onset hearing loss beginning at 3 months of age. Due to the Class A risk factor of cleft palate, the child was monitored every 6 months on the recommendation of the managing audiologist. At 24 months of age a sensorineural hearing loss was diagnosed in this child. Because of the JCIH (2007) position statement recommendation to monitor earlier and more frequent for some risk indicators, this hearing loss was identified. The Class A and Class B classification system was designed to refer children for audiological evaluation at appropriate times based on the presence of risk indicators for delayed-onset hearing loss. Once a child is initially referred for risk indicator monitoring, it is at the discretion of the managing audiologist to set the future monitoring schedule.

The findings of the Class B ototoxic medication exposure in the study population align with Cone-Wesson et al. (2000) and Van Riper and Kileny (1999; 2002), identifying a high occurrence of the risk factor with a low prevalence of associated hearing loss. Although a low incidence, early identification is critical for those infants and their families impacted by hearing loss related to ototoxic medication exposure. Based on previous research, including Prezant et al. (1993), damage from ototoxicity typically occurs within the cochlea. This suggests an evaluation of cochlear outer hair cell function is the most appropriate tool to triage this population to determine necessity of further audiological evaluation. OAE testing has been reported as a non-invasive, cost-effective physiologic measure of cochlear outer hair cell function (JCIH, 2007; Kezirian, White, Yueh, & Sullivan, 2001). Therefore, an OAE test using an ototoxic protocol alone could suffice as a triage protocol for this risk indicator to determine if further diagnostic evaluation is necessary. Implementing OAE triage evaluation to optimize the audiology diagnostic test protocols should be considered to decrease economic impact and improve program efficiency.

The Class B risk indicator of family history of permanent childhood hearing loss was the third most reported risk indicator in this population. It is also the most frequently reported risk indicator from the well-baby population (Hall,

2007; ISB, 2007–2013). Beswick et al. (2013) reported that children with a family history of permanent childhood hearing loss were nearly two times more likely to develop a postnatal hearing loss than those without such family history. Unfortunately, given the lost to follow-up rate of 63% within the current study population, we are potentially missing early diagnosis of more than 17 children with educationally significant hearing loss during this time frame. Potential factors for lost to follow-up which have been cited include maternal race/ethnicity, maternal smoking during pregnancy, public insurance coverage, and area of residence within the state (Liu, Farrell, MacNeil, Stone, & Barfield, 2008).

A question to consider is if family history is reported by the parents, then why is it the highest lost to follow-up rate? To address this question, risk indicator monitoring programs may want to consider improvement in the following two areas: 1) explanation of criteria for family history of hearing loss, and 2) scripts for screeners to inquire about family history of hearing loss. If programs rely on families to interpret family history of hearing loss then reports will more than likely include middle ear dysfunction, presbycusis, and noise induced hearing loss or other acquired hearing loss not due to congenital or genetic factors. To improve the family reported presence of family history of childhood hearing loss and subsequently reduce the lost to follow-up rate for this population, scripts for screeners should be provided which detail the criteria for family history of hearing loss. Additionally, when the risk factor is present, the family should be provided with an explanation of the risk factor and why it is important to receive follow-up services. If the high rate of diagnosing educationally significant hearing loss in the presence of family history risk indicator is accurate, consideration should be given to placing the risk indicator of family history in the Class A category.

During this data review, families were provided with information regarding the risk indicator present and the need for future follow-up based on the risk indicator classification system. Additionally, audiology clinics who received the ISB referral forms attempted to contact the families to schedule appropriate follow-up diagnostic appointments based on the risk indicator classification system. Attempts to contact infants listed in the Class A classification were made by 3 months of age, while attempts to contact infants listed in the Class B classification were made by approximately 9 months of age. Recently, additional steps have been implemented by ISB to reduce the lost to follow-up rate in Idaho. A letter is mailed to the child's primary care physician immediately following identification of an infant who passes their newborn hearing screening with present risk indicators. A letter is also mailed to the child's parents at approximately 6 months of age reminding the parents to schedule an appointment with a pediatric audiologist. Further research is necessary to determine the impact, if any, on the lost to follow-up rate with the implementation of reminder letters from ISB to physicians and parents.

The JCIH (2007) recommendation of completing at least one diagnostic evaluation by 24 to 30 months of age is fairly broad compared to the JCIH 2000 guidelines which recommended monitoring all risk indicators every 6 months until 3 years of age. During 2012–2013, the ISB program used the Class A and Class B monitoring schedules and by doing so children with sensorineural hearing loss were identified prior to 24 months of age. Using the two-class system schedule to provide ongoing monitoring for the Class A risk indicators and one-time monitoring for the Class B risk indicators reduces burden to families and all stakeholders while maintaining an appropriate level for those indicators that pose a higher level of risk. On average children were diagnosed with sensorineural hearing loss by 12 months of age. Having risk monitoring guidelines for state EHDI programs provides structure to the JCIH (2007) recommendations and appears to decrease the diagnosis age for children with risk indicators for delayed-onset hearing loss. As previously mentioned, earlier diagnosis of delayed-onset hearing loss should lead to timely enrollment into early intervention. Further research investigating the impact of the risk indicator classification system on timely enrollment into early intervention is indicated.

Summary

Use of the risk monitoring classification system has enhanced Idaho's EHDI program by providing access to early identification of children who are at higher risk for delayed-onset hearing loss. Subsequently, the early identification of children with delayed-onset hearing loss should lead to timely early intervention. Unfortunately, the high lost to follow-up rate (e.g., over 60% in Idaho) for infants with risk indicators indicates a need for ongoing program improvement.

Monitoring for the risk indicator of ototoxic medication exposure continues to be warranted as indicated by previous and current research. Further research on the potential risk of hearing loss from ototoxic medication exposure is required. With regards to infants with only ototoxic medication exposure, effort should be focused on optimizing the audiology diagnostic test protocols while considering program efficiency and economic impact.

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