

Chapter 10

Risk Monitoring for Delayed-Onset Hearing Loss

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Even if hearing appears to be normal, infants falling in this category should receive regular hearing evaluations thereafter at the physician's office, well-baby clinic, or audiologic center.

A child who is found to have normal hearing as an infant and begins to lose hearing by 3 years of age is considered to have delayed-onset hearing loss. Delayed-onset hearing loss happens more frequently to children with specific risk factors. These risk factors have been identified by the Joint Committee on Infant Hearing (JCIH). This chapter will discuss the importance of knowing these risk factors, as well as the importance of monitoring the hearing of children who possess these risk factors.

The Committee recognized the need for early identification of hearing loss in newborns. The original charges were to make recommendations concerning early identification of children with hearing loss and newborn hearing screening procedures. Their first published statement recommended research efforts on newborn hearing screening to improve techniques and test procedures (JCIH, 1971).

JCIH published the first high-risk criteria in 1973, which included:

History of High-Risk Register

In 1969, the JCIH was established and was composed of representatives from professional organizations interested in children with hearing loss. The committee included:

- American Academy of Pediatrics
- American Academy of Ophthalmology and Otolaryngology
- American Speech & Hearing Association

- History of genetically determined childhood hearing impairment.
- Rubella or other nonbacterial intrauterine fetal infection.
- Defects of ear, nose, or throat.
- Birth weight less than 1500 grams.
- Any indirect or free bilirubin concentration that is potentially toxic.

Newborns would be identified as "at risk" by medical history and physical examination. Continued surveillance of infants with risk factors was also recommended in this

In 1990, the JCIH position statement risk criterion was redefined into two categories: neonates and infants.

position statement. The JCIH concluded that: “Even if hearing appears to be normal, infants falling in this category should receive regular hearing evaluations thereafter at the physician’s office, well-baby clinic, or audiologic center.”

The JCIH 1982 position statement expanded the risk criteria to include bacterial meningitis and severe asphyxia. The statement recommended that any infant with an identified risk indicator “should be screened, preferably under the supervision of an audiologist, optimally by 3 months of age but not later than 6 months of age.” JCIH also recommended that screening for at-risk infants “should

include observation of behavioral or electrophysiological response to sound.”

In 1990, JCIH risk criterion was redefined into two categories: neonates and infants (see *Table 1*). The Committee statement recognized the need for further research in the area of risk criteria.

By 1994, studies revealed that only 50% of infants with significant hearing loss have a risk indicator (Pappas, 1983; Elssmann, Matkin, & Sabo, 1987; Mauk, White, Mortensen, & Behrens, 1991). In light of these findings, JCIH recommended that **ALL** infants should have a newborn hearing screening. Although this document

Table 1
 JCIH 1990 Position Statement Risk Criterion

Risk Criteria/Indicators

JCIH recommended that neonates with one or more risk indicators should be screened using auditory brainstem response (ABR) measurement.

JCIH recommended that infants who had one or more risk indicators “should be screened as soon as possible, but no later than 3 months after the child has been identified as at risk.”

Neonates (birth to 28 days)	Infants (29 days to 2 years)
<ul style="list-style-type: none"> • Family history of congenital or delayed-onset childhood sensorineural impairment. • Congenital infection. • Craniofacial anomalies. • Birthweight less than 1500 grams. • Hyperbilirubinemia requiring exchange transfusion. • Ototoxic medications used for more than 5 days. • Bacterial meningitis. • Severe asphyxia. • Prolonged mechanical ventilation equal to or greater than 10 days. • Physical findings associated with syndromes known to include sensorineural hearing loss. 	<ul style="list-style-type: none"> • Parental/caregiver concerns regarding hearing, speech/language, and/or developmental delay. • Bacterial meningitis. • Neonatal risk factors associated with progressive sensorineural hearing loss. • Head trauma. • Physical findings associated with syndromes known to include sensorineural hearing loss. • Ototoxic medications used for more than 5 days. • Children with neurodegenerative disorders. • Childhood infectious diseases known to be associated with sensorineural hearing loss.

The JCIH 2000 position statement revised the risk criterion based on recently published research from a large National Institute of Health (NIH) sponsored multicenter study.

was a pivotal statement that endorsed universal detection of all infants with hearing loss before 3 months of age and intervention by 6 months of age, the risk indicators remained relatively unchanged.

The JCIH 2000 position statement revised the risk criterion based on recently published research from a large National Institute of Health (NIH) sponsored multicenter study (Cone-Wesson et al., 2000). This study provided data on hearing loss prevalence and incidence for different risk indicators. Of the risk indicators analyzed, prevalence of hearing loss was highest among infants with syndromes (11.7%). Data revealed that

only a small percentage (1.5%) of infants receiving ototoxic medications have hearing loss. Cone-Wesson et al. (2000) confirmed previous findings that only a small percentage of infants with a conventional risk indicator for hearing loss actually had a hearing loss, while a significantly larger number of infants with hearing loss did not have a risk indicator. The findings continued to support the importance of universal neonatal hearing screening for all infants, not just those newborns with risk indicators. Cone-Wesson et al. (2000) provided evidence of delayed-onset permanent hearing loss in young children with certain risk indicators (see *Table 2*).

Table 2
JCIH 2000 Position Statement Risk Criterion

Risk Criteria/Indicators

The JCIH 2000 position statement recommended the following risk criteria for neonates (birth to 28 days):

The JCIH 2000 position statement recommended “audiological monitoring every 6 months until age 3 years” for these risk indicators:

Neonates (birth to 28 days)	Infants (29 days to 3 years)
<ul style="list-style-type: none"> • An illness or condition requiring admission of 48 hours or greater to a neonatal intensive care unit (NICU). • Stigmata or other findings associated with a syndrome known to include a sensorineural and/or conductive hearing loss. • Family history of permanent childhood sensorineural hearing loss. • Craniofacial anomalies, including those with morphologic abnormalities of the pinna and ear canal. • In utero infection, such as cytomegalovirus, herpes, toxoplasmosis, or rubella. 	<ul style="list-style-type: none"> • Parental or caregiver concern regarding hearing, speech language, and/or developmental delay. • Family history of permanent childhood hearing loss. • Stigmata or other findings associated with a syndrome known to include a sensorineural or conductive hearing loss or Eustachian tube dysfunction. • Postnatal infections associated with sensorineural hearing loss, including bacterial meningitis. • In utero infections, such as cytomegalovirus, herpes, rubella, syphilis, and toxoplasmosis. • Neonatal indicators, specifically hyperbilirubinemia at a serum level requiring exchange transfusion, persistent pulmonary hypertension of the newborn associate with mechanical ventilation, and conditions requiring the use of extracorporeal membrane oxygenation. • Syndromes associated with progressive hearing loss, such as neurofibromatosis, osteopetrosis, and Usher’s syndrome. • Neurodegenerative disorders, such as Hunter syndrome, or sensory motor neuropathies, such as Friedreich’s ataxia and Charcot-Marie-Tooth syndrome. • Head trauma. • Recurrent or persistent otitis media for at least 3 months.

In 2007, JCIH published the most recent position statement. This statement highlighted the need for customized reevaluation schedules for children with risk factors based on the subsequent delayed-onset hearing loss. It was recommended that infants who pass the neonatal screening but have a risk factor should have at least one diagnostic audiology assessment by 24 to 30 months of age. For some risk factors with higher prevalence of delayed-onset hearing loss, earlier and more frequent assessments may be indicated. Those “high-risk” factors are listed in *Table 3*. An additional

recommendation indicated if significant family concern was present at any time regarding hearing or communication status, the infant should be referred promptly for an audiological and speech-language assessment.

The risk indicators in the 2007 statement were combined into a single list of congenital and acquired hearing loss risk factors given the “significant overlap among those indicators associated with congenital/neonatal hearing loss and those associated with delayed-onset/acquired or progressive hearing loss.”

Table 3 JCIH 2007 Position Statement Risk Indicators

Risk Criteria/Indicators

The risk factors in the 2007 position statement listed as being associated with permanent congenital, delayed-onset, or progressive childhood hearing loss include:

The 2007 position statement specified that several of the risk factors are of greater concern for delayed-onset hearing loss, including:

Congenital and Acquired Hearing Loss Risk Factors

- **Caregiver concern** regarding hearing, speech, language, or developmental 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 Jervell 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 (especially herpes viruses and varicella), including confirmed bacterial and viral meningitis.
- **Head trauma**, especially basal skull/temporal bone fracture that requires hospitalization.
- **Chemotherapy**

Greater Concern for Delayed-Onset Hearing Loss Risk Factors

- Caregiver concern
- Family history of permanent childhood hearing loss
- ECMO
- CMV
- Syndromes associated with progressive hearing loss
- Neurodegenerative disorders
- Cultural-positive postnatal infections associated with sensorineural hearing loss
- Head trauma
- Chemotherapy

The JCIH 2007 position statement encouraged further research for the success of the early hearing detection and intervention (EHDI) system.

The JCIH 2007 position statement encouraged further research for the success of the early hearing detection and intervention (EHDI) system. Specific research recommendations that affect risk factors related to delayed-onset or progressive hearing loss are to:

- Develop state data management systems with the capacity for accurate determination of the prevalence for delayed-onset or progressive hearing loss.
- Develop state data tracking systems to follow infants with suspected and confirmed hearing loss through individual state EHDI programs.
- Track genetic, environmental, and pharmacologic factors that contribute to hearing loss, thus allowing for tailored prevention and intervention strategies.

To assist with understanding the current risk indicators, each factor identified in the JCIH 2007 position statement is highlighted below:

Caregiver concern. JCIH 2007 recommends that if there are significant hearing, speech, language, or developmental delay concerns, the medical home will provide families with referrals for audiological and speech-language evaluations. These referrals should occur whenever concerns are expressed or noted.

Family history of permanent childhood hearing loss. Infants (well-baby or NICU) with a positive family history of permanent childhood hearing loss should receive early and ongoing audiological monitoring.

Neonatal indicators. NICU stay—approximately 400,000 infants are admitted annually to NICUs in the U.S. National Perinatal Research Center (NPIC) data from 2005 indicated infants in the NICU for less than 5 days were at low risk for hearing loss. Infants in the NICU for greater than 5 days, however, were identified as a population for which it is extremely important to rule out neural hearing loss.

The prevalence of hearing loss found in infants who were admitted to NICU for greater than 5 days with no additional risk factors has not been reported.

ECMO. ECMO is life support used for individuals in cardiopulmonary or respiratory failure. The machine circulates the child's blood through an artificial lung, while also providing oxygen to the child to give the lungs and heart adequate time to heal. Fligor (2008) reported on a retrospective chart review of ECMO patients at Children's Hospital in Boston. Of 111 neonates, 26% had sensorineural hearing loss—nearly half of those having a delayed onset. Seventy-five percent of the infants with hearing loss had progressive hearing loss. JCIH 2007 recommended that children receiving ECMO treatments should have “early and more frequent assessments.”

Cone-Wesson et al. (2000) estimated that 1 in 56 children with permanent hearing loss by age 1 had the following risk factors:

- Low birth weight
- Respiratory distress syndrome
- Bronchiopulmonary dysplasia
- Mechanical ventilation greater than 36 days. When an infant requires mechanical ventilation, there is oxygen deprivation that may result in destruction of hair cells in the cochlea.

Ototoxic medications. Cone-Wesson et al. (2000) documented the most prevalent risk factor was ototoxic medications—occurring in more than 70% of infants studied. Interestingly, it was reported that ototoxic exposure resulted in hearing loss for only 1.5% of infants. So why is there concern about ototoxicity in infants? In 1993, Prezant et al. reported on the genetic mutation A1555G, which has been associated with aminoglycoside deafness. For individuals with the A1555G mutation, it was found that even a single dose may result in sensorineural hearing loss. Estivill et al. (1998) reported profound hearing loss in individuals with A1555G mutation with no aminoglycoside treatments. Two studies reported on the prevalence of the

mutation in newborns (Tang et al., 2002; Scrimshaw, Faed, Tate, & Yun, 1999). The study from the United Kingdom found 1 in 206 newborns expressing the mutation; while in Texas, only 1 in 1,161 newborns were found with the mutation. Unfortunately, there is currently a lack of sufficient data regarding prevalence of hearing loss in newborns receiving ototoxic medications with no additional risk factors. The JCIH 2007 position statement recommendation for ototoxic monitoring is at least one audiology evaluation by 24-30 months of age.

Hyperbilirubinemia or jaundice that requires exchange transfusion. In severe cases, damage can occur in the central nervous system that may result in neural hearing loss. Nickisch, Massinger, Ertl-Wagner, and vonVoss (2009) compared auditory test results (ABR, transient-evoked otoacoustic emissions, and behavioral audiometry) of 15 children with high bilirubin levels (above 20 mg/dL) to 15 children with low bilirubin levels. Hearing function disorders were reported in 87% of the high bilirubin level group, with auditory neuropathy spectrum disorder found in 8 of these children. In the low bilirubin group, only two children were found to have cochlear hearing dysfunction. A literature search completed by Ip et al. (2004), reported in a technical report for the American Academy of

Pediatrics, found 14 studies that examined the effect of bilirubin on hearing impairment. Eight of these were deemed high-quality studies that showed significant relationship between abnormal ABRs and high bilirubin levels.

In utero infections. JCIH 2007 recommends risk monitoring for infants with CMV, herpes, rubella, syphilis, or toxoplasmosis. In the U.S., CMV is the most common congenital infection. It has been associated with progressive and delayed-onset hearing loss, which requires more frequent audiological monitoring. (For further information on CMV, see the CMV section of this publication.)

Herpes is a sexually transmitted infection and is a risk if:

- The infant has neonatal herpes.
- The mother had an active lesion during a vaginal delivery.
- The mother had an active lesion during a c-section delivery with a premature rupture of membranes.

Rubella, if contracted during pregnancy, can result in serious birth defects, including hearing loss. Syphilis infection without immediate antibiotic treatment can result in hearing loss. Toxoplasmosis is a parasite infection that, depending on the severity of the infected infant, may cause hearing loss.

Craniofacial anomalies. Craniofacial anomalies include those that involve the pinna, ear canal, ear tags, ear pits, and temporal bone anomalies. Cone-Wesson et al. (2000) reported greater than 50% of children with craniofacial anomalies present with hearing loss. Infants with cleft palate are at greater risk for educationally significant hearing loss. Studies have shown a prevalence of hearing loss (conductive, mixed, or sensorineural) greater than 50% in infants, children, and adults who have cleft palates (Helias, Chobaut, Mourat, & Lafon, 1988; Paradise 1975; Viswanathan, Vidler, & Richard, 2008; Yules, 1970).

Physical findings associated with a syndrome known to include a sensorineural or permanent conductive hearing loss. The most commonly reported physical finding is a white forelock, which is associated with Waardenburg syndrome.



Photo courtesy of Advanced Bionics

Hall III (2007) highlighted the importance of audiologists who work with the infant population being intimately familiar with all the risk factors for hearing loss, whether peripheral, auditory dysfunction, or delayed onset.

Syndromes associated with hearing loss or progressive or delayed-onset hearing loss.

JCIH 2007 recommends risk monitoring of children with:

- Neurofibromatosis
- Osteopetrosis
- Usher syndrome
- Waardenburg syndrome
- Alport syndrome
- Pendred syndrome
- Lange-Nielson syndrome

Any child with a syndrome associated with progressive hearing loss should receive frequent audiological monitoring.

Neurodegenerative disorders. Hunter syndrome, Friedreich ataxia, and Charcot-Marie-Tooth syndrome are of greater concerns for delayed-onset hearing loss and require more frequent audiological monitoring.

Culture-positive postnatal infections associated with sensorineural hearing loss. Bacterial meningitis and sepsis are culture-positive postnatal infections associated with hearing loss. Bacterial meningitis occurring at any age may result in damage to the cochlea. An early and more frequent audiological monitoring schedule is recommended.

Head trauma. Head trauma, especially basal skull and temporal bone fractures that require hospitalization, are of concern regarding potential hearing loss. Trauma is a risk factor that requires an early and more frequent monitoring schedule. The use of vacuum suction during delivery has not been associated with hearing loss.

Chemotherapy. Similar to ECMO treatments, infants who receive chemotherapy treatments should receive early and frequent assessments. In 2009, the *American Academy of Audiology Position Statement and Clinical Practice Guidelines: Ototoxicity Monitoring* was published, which provides recommendations for audiological assessments and scheduling.

Current Risk Factors for Monitoring

Hall III (2007) provided a summary of the relationship of risk factors with subsequently diagnosed hearing loss from the most current and frequently cited studies that estimated prevalence of risk factors for delayed-onset or progressive hearing loss (Cone-Wessen et al., 2000; Van Riper & Kileny, 2002). In a series of 2,103 infants, Hall III identified six risk factors that occurred most frequently in the entire population. When considering only those infants in this population who were diagnosed with hearing loss, Hall III (2007) identified six risk factors associated with hearing loss that occurred most frequently (see *Table 4*).

Hall III (2007) highlighted the importance of audiologists who work with the infant population being intimately familiar with all the risk factors for hearing loss, whether peripheral, auditory dysfunction, or delayed onset. The risk factor prevalence data provided in *Table 4* outlines this importance. Although administration of ototoxic medications occurs frequently in the infant population, it is one of the least frequent factors in those infants actually diagnosed with hearing loss. However, the presence of ototoxic medications is a risk for progressive hearing loss. Risk factors that occur most frequently in both conditions, including severe asphyxia and mechanical ventilation, require earlier and more frequent hearing evaluations. Specifically for delayed-onset hearing loss, which occurs in 1 per 56 children with permanent hearing loss by 1 year of age, the risk factors that need to be monitored closely are:

- Low birth weight
- Respiratory distress syndrome
- Bronchiopulmonary dysplasia
- Mechanical ventilation

These four factors are not the most frequently occurring factors in all infants or those infants diagnosed with hearing loss. Knowing all the risk factors is extremely important when working with this population.

Table 4 Risk Factors with Subsequently Diagnosed Hearing Loss

Risk Factors in Entire Infant Population

Risk factors that occurred most frequently in the entire population:	Risk factors that occurred least frequently (<10%) in this population:
Risk Factors	Risk Factors <10%
<ol style="list-style-type: none"> Ototoxic medications (>70%) Severe asphyxia (>50%) Mechanical ventilation less than 5 days (>25%) Low birthweight (>20%) Parental/physician concerns (>15%) ECMO (>10%) 	<ol style="list-style-type: none"> Hyperbilirubinemia Craniofacial anomalies Family history Congenital infections Bacterial meningitis Maternal substance abuse Neurodegenerative disorders

Risk Factors in Infants Diagnosed with Hearing Loss

Risk factors associated with hearing loss that occurred most frequently:	Risk factors that occurred least frequently (<10%) in this population:
Risk Factors	Risk Factors <10%
<ol style="list-style-type: none"> Craniofacial anomalies (>50%) Family history of childhood hearing loss (>15%) Severe asphyxia (>15%) Congenital infections (>15%) Mechanical ventilation (>10%) Bacterial meningitis (>10%) 	<ol style="list-style-type: none"> Low birth weight Hyperbilirubinemia Ototoxic medications ECMO Substance abuse 6-7. Parent or physician concern and neurodegenerative disorders—two factors found to occur with negligible frequency.

Risk Monitoring Program

The first goal of a risk monitoring program is to identify infants and children at risk for delayed-onset or progressive hearing loss. Hospital staff, birthing center staff, and medical homes play a critical role in the identification of these infants and children. The second goal is timely diagnostic assessments from a pediatric audiologist. As

previously mentioned, JCIH 2007 recommends early and more frequent monitoring schedules for certain risk indicators. The final goal of a risk monitoring program is to maintain a monitoring and tracking system in the state EHDI data management system. Monitoring and tracking will improve the data on hearing loss prevalence among the risk indicators, allowing for improvements to be made in prevention and intervention methods.

The first goal of a risk monitoring program is to identify infants and children at risk for delayed-onset or progressive hearing loss.

An explanation of the screening results alone is not sufficient. Special attention should be given to delayed-onset risk indicators for those infants who pass their screen with risk indicators present. Educating the family on all aspects of the screening process can be expected to significantly improve follow up.

Key components of a successful risk monitoring program include:

- Team of stakeholders
- Data management system
- Annual program evaluation

These components are reviewed below.

Team of Stakeholders for a Risk Monitoring Program

A successful risk monitoring program requires collaboration from a team of stakeholders:

- Hospital/birthing center
- Medical home
- Pediatric audiology center
- State EHDI program

Hospital/birthing center roles:

- Identify infants who have one or more risk indicators.
- Provide the family with a referral to a pediatric audiology center.
- Provide the family with information about risk indicators.
- Provide the medical home with information regarding the risk indicator referral.
- Report the infants with risk indicators to state EHDI program.

Using a referral form helps hospitals and birthing centers to be successful with these roles. *Figure 1* shows the referral form created by Idaho Sound Beginnings (ISB) that is used by Idaho hospitals and birthing centers.

To assist with improving the percentage of babies screened, hospitals and birthing centers should also identify infants who are discharged early, transferred to another hospital, or not screened for some reason. Parents of these infants can be contacted by the monitoring program representative and informed of outpatient hearing screening options and education on the screening process, including risk indicators for delayed-onset hearing loss. Details of the screening process, along with the purpose of the screening, need to be communicated to the family through verbal communication as well as written

information. An explanation of the screening results alone is not sufficient. Special attention should be given to delayed-onset risk indicators for those infants who pass their screen with risk indicators present. Educating the family on all aspects of the screening process can be expected to significantly improve follow up.

Medical home roles:

- Being familiar with risk factors for delayed-onset hearing loss.
- Explaining screening results and answer questions for the family.
- Highlighting existing risk indicators.
- Encouraging risk monitoring follow up.
- Providing the family with a referral to a pediatric audiology center.

Medical homes are expected to have knowledge of the risk factors and to support and encourage the families to follow up with pediatric audiology centers. When it comes to hearing loss, the “wait and see” approach is not appropriate. The window of opportunity to learn speech and language is far too narrow to delay identification and, if indicated, intervention. Primary care physicians (PCP) have a unique responsibility to review infants’ medical records, discuss the results with parents, and recommend the necessary steps in the monitoring process. The medical home’s active and deliberate involvement in the risk factor monitoring process should significantly reduce the number of children lost to follow up in the EHDI system.

Pediatric audiology center roles:

- Providing appropriate comprehensive diagnostic testing for children with risk factors.
- Knowing the risk factors that have higher prevalence of delayed-onset hearing loss and requiring earlier and more frequent assessments.
- Providing documentation regarding evaluation outcomes to state EHDI program.

Audiologists at pediatric audiology centers are expected to provide appropriate diagnostic evaluations for all children, particularly those with risk factors for

delayed-onset hearing loss. Protocols for monitoring this population should be in place to ensure follow up is completed in a timely manner, allowing for the earliest possible identification of hearing loss and implementation of appropriate intervention. Dividing the risk factors into Class A—monitoring required by 3 months of age—and Class B—monitoring required by 12 months of age— is an example of a protocol that would highlight those risk factors that have a higher prevalence of delayed-onset hearing loss. Audiologists must be diligent in their documentation and ensure that infant toddler services (ITS) and state EHDI programs are informed of all diagnostic evaluation outcomes. Informing ITS will ensure speech and language intervention and other early intervention services are initiated in a timely manner. Keeping the EHDI programs informed will assist with tracking and reduce loss to follow up.

State EHDI program roles:

- Providing training and support for hospitals, birthing centers, physicians, and pediatric audiologists on risk factors.
- Providing a method for hospitals, birthing centers, and pediatric audiologists to report information on infants at risk to the state EHDI program.
- Tracking and surveillance of infants with risk factors.

State EHDI programs have an important role in monitoring children with risk factors for delayed-onset hearing loss. These programs provide essential training and lend support to all entities involved with the monitoring and identification of hearing loss. They create and disseminate tools designed to improve the system and reduce loss of children to follow up. One such tool is a letter to the medical homes to inform providers of screening and diagnostic results, as well as note risk factors for delayed-onset hearing loss. This is intended to encourage providers to remind families to schedule and keep monitoring appointments. This information can also be a starting point

for discussion of appropriate intervention for these children who are at risk.

Data Management System for a Risk Monitoring Program

National EHDI Goal 6 states: “Every state will have an EHDI tracking and surveillance system that minimizes loss to follow-up” (CDC, 2010). State EHDI programs need a data management system to track infants with risk indicators. These systems will also need to track infants identified with delayed-onset hearing loss.

Annual Program Evaluation of a Risk Monitoring Program

National EHDI Goal 7 states: “Every state will have a system that monitors and evaluates the progress towards the EHDI goals and objectives” (CDC, 2010). The monitoring and evaluation of risk indicators for delayed-onset hearing loss is a continual process. Annually, state EHDI programs should evaluate:

- Education and training of medical staff (nurses, physicians, midwives) regarding risk monitoring for delayed-onset hearing loss.
- Data collection of risk indicators from hospitals and birthing centers.
- Effectiveness of the referral process for infants with risk indicators.
- Information given to families of infants with risk indicators.
- Education for pediatric audiologists on risk monitoring for delayed-onset hearing loss.
- Data collection of audiological test results from pediatric audiologists.
- Education for medical homes on risk monitoring for delayed-onset hearing loss.
- Performance of the data management system used to track risk indicators.
- Information and support for families of infants diagnosed with delayed-onset hearing loss.
- The plan for monitoring and evaluation of opportunities for infants and children to receive appropriate intervention services.

State EHDI programs have an important role in monitoring children with risk factors for delayed-onset hearing loss. These programs provide essential training and lend support to all entities involved with the monitoring and identification of hearing loss.

National EHDl Goals:

“Every state will have an EHDl tracking and surveillance system that minimizes loss to follow-up”

“Every state will have a system that monitors and evaluates the progress towards the EHDl goals and objectives”

Current Data and Future Research on Risk Monitoring

As previously stated, JCIH recommends research in the following areas:

- Development of state data management systems with the capacity for the accurate determination of the prevalence for delayed-onset or progressive hearing loss.
- Development of state data tracking systems to follow infants with suspected and confirmed hearing loss through the EHDl program.
- Track genetic, environmental, and pharmacologic factors that contribute to hearing loss, allowing for tailored prevention and intervention strategies.

Idaho Data on Risk Indicators and Delayed-Onset Hearing Loss

ISB is the EHDl program for the state of Idaho. In 2007, ISB program began collecting data on risk indicators and delayed-onset hearing loss using the Hi*Track data collection system.

After the publication of the JCIH 2007 position statement, Idaho Sound Beginnings initiated efforts to train staff at birthing centers and hospitals, as well as audiologists across the state. Training included education on risk factors, monitoring protocols, and role of stakeholders in the process. As a result of these training efforts, the number of infants with documented risk factors gradually increased over the next 6 years. Idaho data showed the prevalence of infants with a risk indicator ranging from 3.1-11.4%, increasing from 660 infants in 2007 to 2,102 infants in 2013 (see *Figure 2*). The prevalence information includes all infants born in Idaho from January 2007 to December 2013 (n=160,833).

The most frequently reported risk indicators were within the neonatal indicator category. Neonatal indicators—combined together for the purpose of this data analysis—included:

- NICU stay of greater than 5 days
- ECMO
- Exposure to ototoxic medications
- Low APGAR scores
- Low birth weight
- Hyperbilirubinemia requiring exchange transfusion

Additional risk indicators reported with significant frequency were family history and craniofacial abnormalities (see *Figure 3*). This data coincides with the findings of Hall III (2007), discussed earlier in this chapter. This data does not account for infants with one or more risk factors.

When looking specifically at delayed-onset permanent hearing loss, data revealed 3.5 infants per 10,000 were diagnosed from 2007 to 2011. Of those 35 infants, 7 had no reported risk indicators, which resulted in 2.7 per 10,000 infants with risk indicators diagnosed with delayed-onset permanent hearing loss. Of the 28 infants with risk indicators, 10 risk indicators were reported (see *Figure 4*). The most frequently occurring risk indicators were a NICU stay longer than 5 days (15 infants) and ototoxic medication treatment (13). It should also be noted that 75% of those 28 infants had multiple risk indicators reported.

In October 2011, Idaho began collecting data on early and more frequently monitored risk indicators—classified as Class A. All other risk indicators were classified as Class B (see *Figure 5*). Infants with Class A risk indicators were recommended for evaluation by a pediatric audiologist by 3 months of age.

From October 2011 to May 2014, data was collected from all reporting birthing hospitals in Idaho. Data indicated 145 infants with risk factors for delayed-onset hearing loss as defined by JCIH passed their newborn hearing screening (see *Figure 6*).


Of those infants, 57 returned for monitoring diagnostic audiology evaluations. Present factors—denoted as Class A risk factors—were craniofacial abnormalities, postnatal infections, and syndromes associated with hearing loss.

Of the infants with Class A risk indicators that returned for comprehensive diagnostic audiology evaluations, 40% (23 of 57 infants) were found to have an educationally significant hearing loss by 3 months of age. These findings support early and more frequent monitoring of certain risk indicators. It also indicates

over 60% of the infants with risk factors for delayed-onset hearing loss are being lost to follow up. If the trend noted for those infants that did follow up is true for those lost to follow up, an estimated 35 infants with delayed-onset hearing loss were missed during this time period in Idaho. Dividing the risk indicators into Class A and B has improved Idaho's early diagnosis of infants who have hearing loss associated with craniofacial abnormalities, postnatal infections, and syndromes. However, with a loss to follow up rate of greater than 60%, much work remains to be completed.

Figure 1

ISB Referral Form 2014



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

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

Newborn Hearing Screening Referral Form (*Transfers only) **Birth Hospital:** _____ [Please Press Firmly]

Receiving Hospital: _____

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, PO Box 83720, Boise, ID 83720 **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 NICU/Special Care

Baby's Primary Physician/Clinic: _____

Mother's name: _____

3. HEARING SCREEN RESULTS:

First Screen: **R** Pass Refer No Result
Date: _____ **L** Pass Refer No Result

Second Screen: **R** Pass Refer No Result
Date: _____ **L** Pass Refer No Result

2. CONTACT INFORMATION:

Parent/Guardian: _____
Last First

Address: _____

City: _____ State: _____ Zip: _____

Main Phone: _____ Text? _____

Alternate Phone/Contact: _____

Email/other contact: _____

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)

Nursing/screening staff will inform you of the final results of the baby's newborn hearing screen and give you a copy of these results. If your baby needs testing or follow-up for risks, you will be given an appointment and/or follow-up information. 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.

Audiologist: _____

Phone: _____

Address: _____

(For a listing of *Pediatric Audiologists* call Idaho Sound Beginnings at: (208) 334-0829)

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 and diagnostic audiology evaluations 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, as 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 :** _____ 5/14

Figure 1
(continued)



IDAHO SOUND BEGINNINGS
Early Hearing Detection and Intervention (EHDI)
Department of Health and Welfare, Infant Toddler Program

AUDIOLOGY RESULTS FORM
BIRTH TO 3 YEARS

Please enter details regarding patient's hearing status, testing and recommendations. Complete (or verify) contact and risk information on side one.
FAX completed form to 208-332-7331 within 5 days of evaluation.

BABY'S INFORMATION:
Baby's Name: _____
Mothers Name: _____
Last First
DOB: ____/____/____ **Gender:** M F
Name of Birth Hospital: _____
Baby's Primary Care Provider: _____

SIDE 1 OF FORM SHOULD BE USED TO ENTER RISK FACTOR AND CONTACT INFORMATION OR ATTACH THE HOSPITAL REFERRAL FORM IF AVAILABLE.

DIAGNOSTIC TEST BATTERY:

ABR	Click – Wave V threshold	(dBeHL)			
	<u>Air</u> - RIGHT	_____	LEFT	_____	
	<u>Bone</u> - RIGHT	_____	LEFT	_____	
	Tone – (kHz)	.5	1	2	4
	<u>Air</u> - RIGHT	_____	_____	_____	_____
	LEFT	_____	_____	_____	_____

OAE TEOAE or DPOAE

<u>RIGHT</u>	<u>LEFT</u>
Pass _____	Pass _____
Refer _____	Refer _____
Could Not Test _____	Could Not Test _____

ACOUSTIC IMMITTANCE

TYMPANOMETRY: Hz- _____

Type RIGHT: _____ LEFT: _____

BEHAVIORAL- threshold	VRA _____	CPA _____
(kHz) - .5 1 2 4 8	- Speech	
RIGHT _____	_____ (dB HL)	
LEFT _____	_____ (dB HL)	
Sound Field _____	_____ (dB HL)	

Reason for Testing:
Hearing Screening Refer -
Risk Indicators or Concerns -

DATE OF EVALUATION: _____
This is baby's first visit to audiologist -
This is **Follow-up** testing after initial visit -

DIAGNOSIS: (STATUS OF HEARING AT THIS VISIT)

Hearing Loss-	RIGHT EAR	LEFT EAR
	<input type="checkbox"/> No	<input type="checkbox"/> No
	<input type="checkbox"/> Yes	<input type="checkbox"/> Yes

Degree of Loss-	RIGHT EAR	LEFT EAR
	<input type="checkbox"/> Mild	<input type="checkbox"/> Mild
	<input type="checkbox"/> Moderate	<input type="checkbox"/> Moderate
	<input type="checkbox"/> Mod-Severe	<input type="checkbox"/> Mod-Severe
	<input type="checkbox"/> Severe	<input type="checkbox"/> Severe
	<input type="checkbox"/> Profound	<input type="checkbox"/> Profound

Type of Loss-	RIGHT EAR	LEFT EAR
	<input type="checkbox"/> Conductive-fluctuating	<input type="checkbox"/> Conductive-fluctuating
	<input type="checkbox"/> Conductive-permanent	<input type="checkbox"/> Conductive-permanent
	<input type="checkbox"/> Sensorineural	<input type="checkbox"/> Sensorineural
	<input type="checkbox"/> Mixed	<input type="checkbox"/> Mixed
	<input type="checkbox"/> Central/Neural	<input type="checkbox"/> Central/Neural
	<input type="checkbox"/> Undetermined	<input type="checkbox"/> Undetermined

FOLLOW-UP CHECKLIST:

REPORT ALL RESULTS TO IDAHO SOUND BEGINNINGS (Birth-3)

Audiologic Re-evaluation and/or Monitoring needed
When/How often? _____
Return Appointment Pending: yes no

Referred for Medical Follow-up/ENT Consult-Clearance

No Follow-up is needed -Referred to Medical Home

Lost to Follow-up-Discharged after no response/no show

Amplification is Recommended

Ophthalmology Exam is Recommended

Genetic Counseling is Recommended

IF A HEARING LOSS HAS BEEN IDENTIFIED -

Referral has also been made to Infant Toddler Program

COMMENTS/NOTES:

(Audiologist Signature)

Clinic Name: _____

Mail to: Idaho Sound Beginnings-ITP
450 W. State St. Fl-5 (208) 334-0829
Boise, ID 83720

Fax to: (208) 332-7331

5/14

Figure 2
Prevalence of Infants with a Risk Indicator in ISB 2007-2013 Data

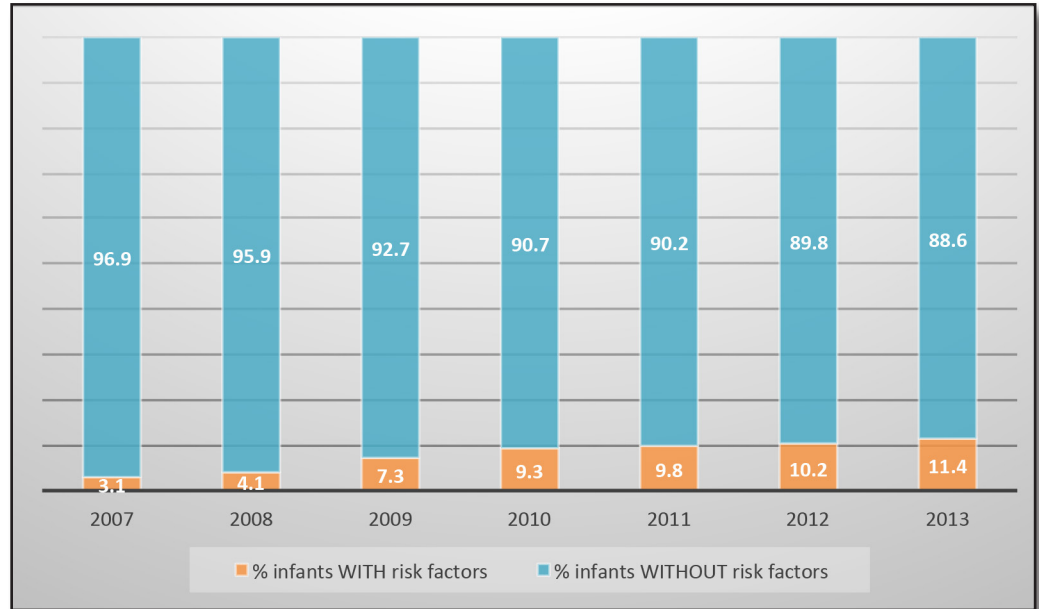


Figure 3
Number of Risk Indicators Reported in ISB 2007-2013 Data

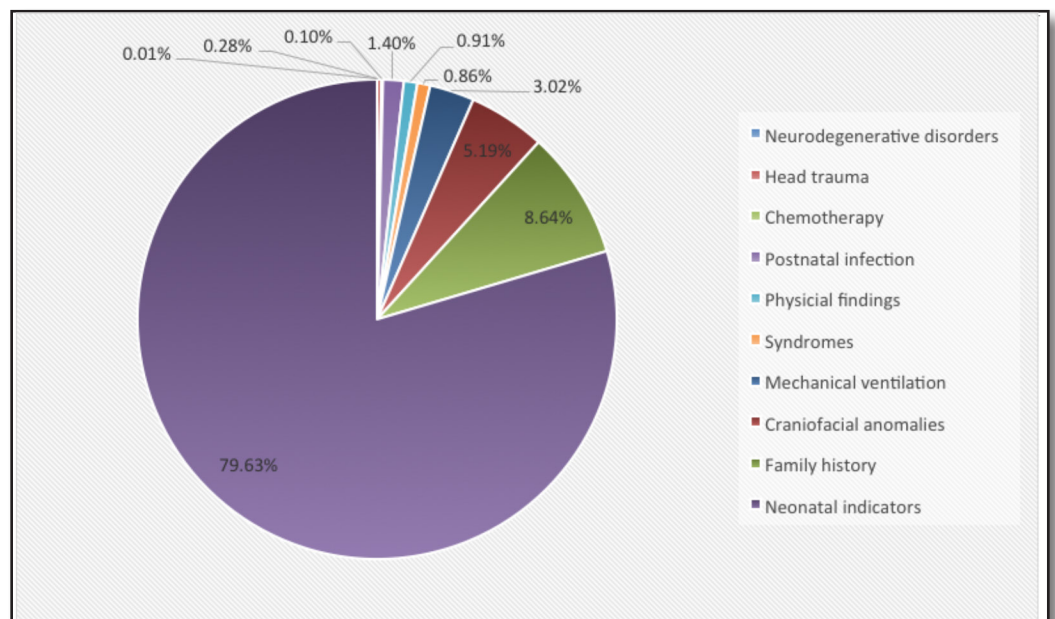


Figure 4

Risk Indicators Reported for the 28 Infants Diagnosed with Delayed-Onset Permanent Hearing Loss with Risk Indicators Present Reported in ISB 2007-2011 Data

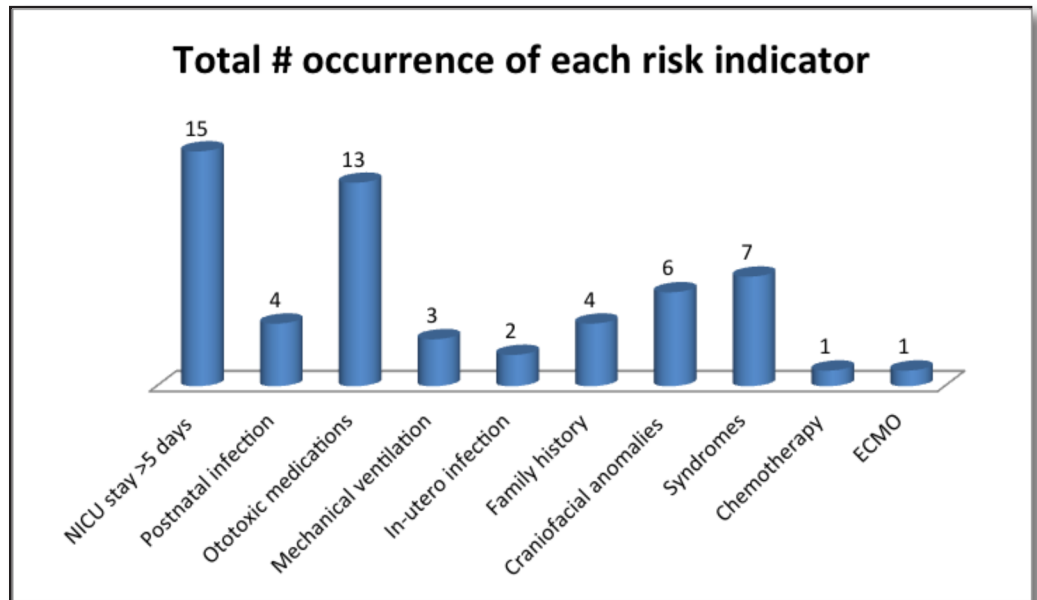


Figure 5

Idaho Risk Monitoring Algorithm

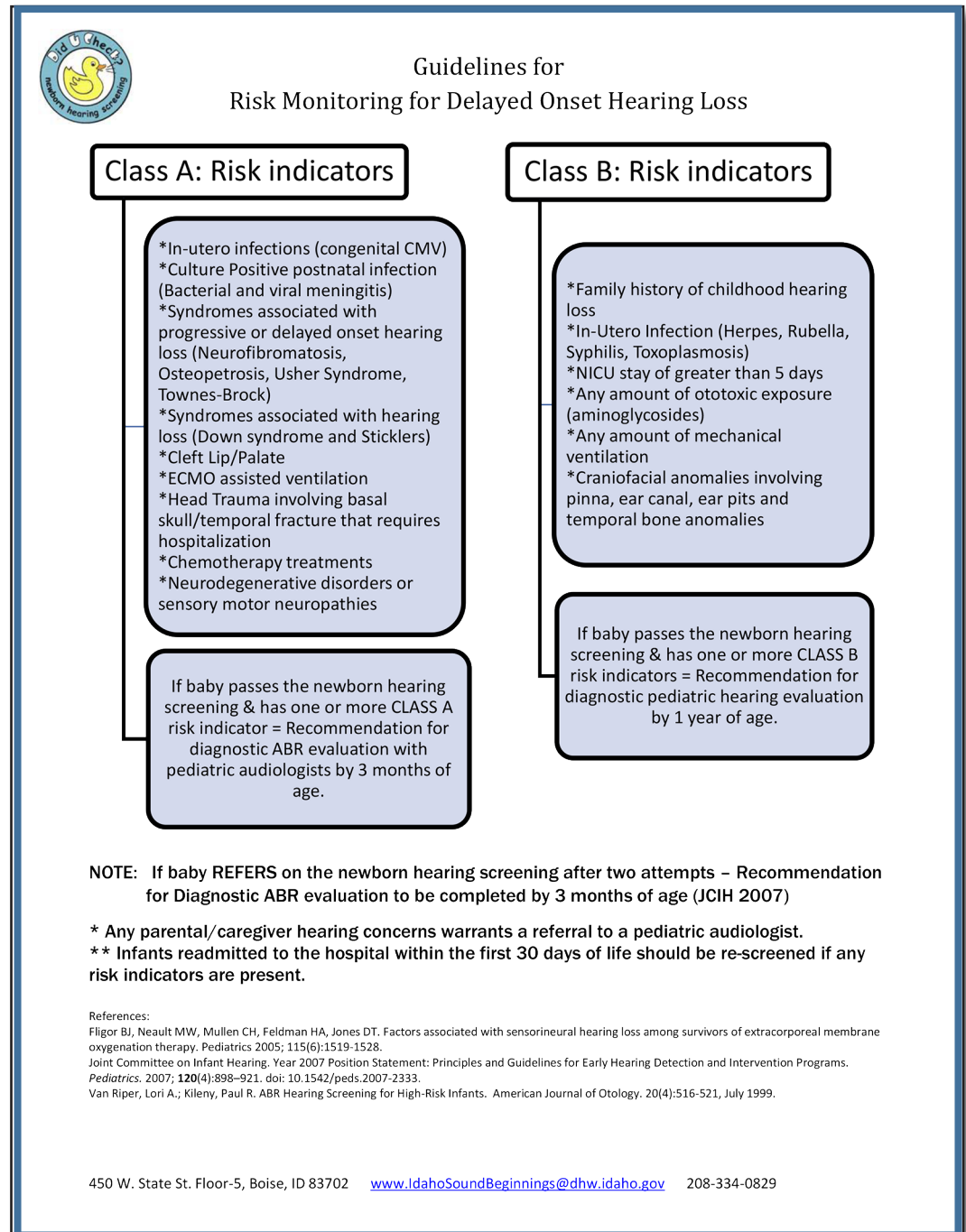
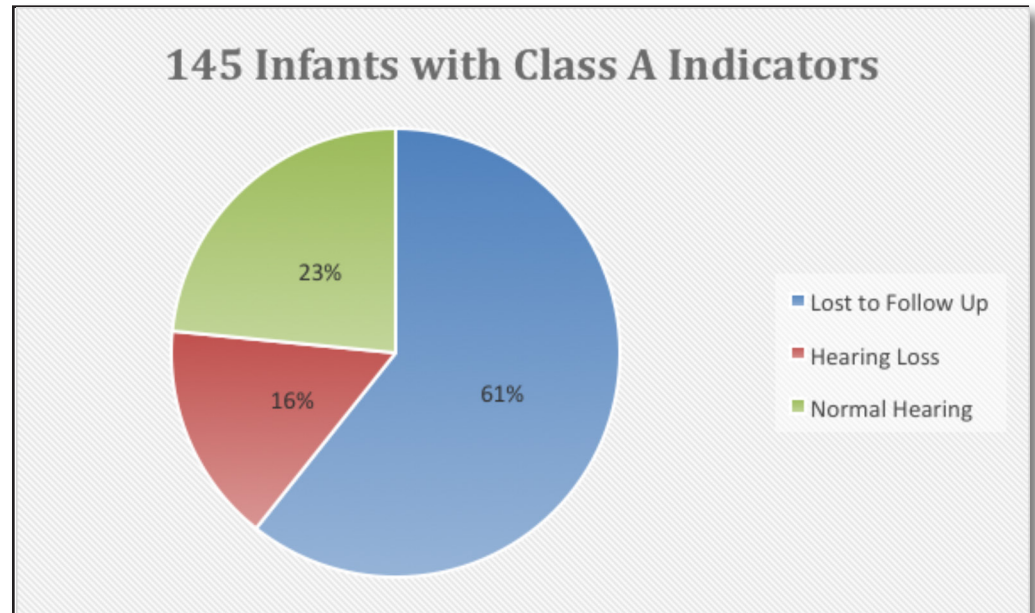


Figure 6
Class A Risk Factors in Idaho Birthing Hospitals



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