Ototoxicity monitoring as part of risk monitoring in the EHDI system

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JCIH 1990 Position Statement

High risk criteria additions:

» Ototoxic medications
» Prolonged mechanical ventilation
» Physical findings of syndromes
» Parent/caregiver concerns
» Head trauma
» Neurodegenerative disorders
» Infectious diseases associated with hearing loss

Screening recommendation changes:

» Auditory Brainstem Response measurement, not behavioral testing
Recommended ALL infants screened before hospital discharge

- Risk monitoring:
  - Audiological testing every 6 months until age 3 years.
JCIH 2007 Position Statement

- Expanded definition of targeted hearing loss to include:
  - Neural hearing loss (Auditory Neuropathy/Dysynchrony) in infants admitted to the NICU

- Separate protocols for NICU and well baby nurseries:
  - NICU babies (>5 days) are to have ABR screening so that neural hearing loss will not be missed
Re-admissions

- Infant readmitted in the first month of life and present with conditions, which are associated with potential hearing loss, need a repeat hearing screen prior to discharge.

Monitoring of high risk indicators

- “Infants with risk factors for hearing loss should have at least one diagnostic evaluation by 24-30 months of age.”
Caregiver concerns (re: hearing, speech, language, or developmental delay)

Family history of permanent childhood hearing loss

Neonatal Intensive Care (NICU) of more than 5 days or any of the following regardless of length of stay: ECMO, assisted ventilation, exposure to ototoxic medications (gentimycin and tobramycin) or loop diuretics (furosemide, Lasix), and hyperbilirubinemia that requires exchange transfusion.

In-utero infections

Craniofacial anomalies

Known physical findings associated with a syndrome

 Syndromes associated with hearing loss, progressive hearing loss or late-onset hearing loss neurodegenerative disorders

Culture-positive postnatal infections associated with hearing loss

Head trauma, especially basal skull/temporal bone, requiring hospitalization

Chemotherapy
National Perinatal Research Center (NPIC) (Quality Analytic Services (QAS)) made the recommendation regarding NICU stay for JCIH 2007:

- Approximately 25% of NICU infants are considered “LOW” risk and discharged by 5 days old.
- The remaining approximately 75% of NICU infants, who are hospitalized for greater than 5 days, are considered the “TARGET” population to rule out neural hearing loss.

**NICU stay of greater than 5 days and exposure to loop diuretics were not associated with increased risk of hearing loss (Kraft et al, 2014)**
ECMO treatments

- Expracorporeal Membrane Oxygenation (ECMO)- is an aggressive treatment that is used for the life support in infants with respiratory or cardiopulmonary failure.

- Study found receiving aminoglycoside antibiotics cumulative of 14 days or more in the course of ECMO raised the risk of SNHL by 5.56 times.
medications that can damage the ear, resulting in hearing loss, ringing in the ear, or balance disorders.
Ototoxic Medications

- Over 200 known ototoxic medications (prescriptions and OTC)
- Used to treat serious infections, cancer, heart disease
- Damage may be temporary or permanent
  - Aspirin (temporary)
  - Cisplatin (permanent)
Why concern about ototoxicity with infants?
Most frequently occurring risk factors

- Ototoxic Medications (>70%)
- Severe Asphyxia (>50%)
- Mechanical Ventilation less than 5 days (>25%)
- Low birth weight (>20%)
- Parental/Physician concerns (>15%)
- ECMO (>10%)

Least frequently occurring risk factors (<10%)

- Hyperbilirubinemia
- Craniofacial anomalies
- Family history
- Congenital infections
- Bacterial meningitis
- Substance abuse (maternal)
- Neurodegenerative disorders

Frequency of hearing loss among high risk indicators

- Craniofacial anomalies (>50%)
- ECMO treatments (>20%)
- Severe Asphyxia/ Mechanical ventilation (>15%)
- Congenital infections (>15%)
- Family History (>15%)
- Bacterial meningitis (>10%)
- Other risk indicators (<10%)

Aminoglycosides

- Introduced in 1940s
- Used to treat serious infections due to multi-drug resistant Gram negative bacteria
- May remain in hair cells for months after application (Aran et al, 1999)
- “...weekly or biweekly monitoring is recommended ideally.” “...follow-up testing should also be scheduled a few months after drug discontinuation.” (AAA Ototoxicity Monitoring, 2009)
Gentamicin

- Introduced 1963
- Most common aminoglycoside used in NICU
- Low cost
- Effectiveness against most Gram-negative bacteria
ASHA 2010- Evidence Based Systematic Review: Drug-Induced Hearing Loss- Gentamicin

- Systematic literature review (20 studies)
- Reported hearing loss from gentamicin induced cochleototoxicity ranging from 0-58%
- Studies varied in dosing, patient populations, diagnostic testing, diagnostic criteria for hearing loss
ASHA 2010- Evidence Based Systematic Review: Drug-Induced Hearing Loss - Gentamicin

- Trends noted in the studies:
  - Frequency of administration did not influence the likelihood of hearing loss
  - Dosing amount did not influence the likelihood of hearing loss
A1555G genetic mutation

- Prezant et al (1993) reported on the genetic mutation A1555G, associated with aminoglycoside deafness
- Estivill et al (1998) reported profound hearing loss without aminoglycoside treatments
- United Kingdom study (2002) found 1 in 206 newborns expressing the mutation
- Texas study (1999) only 1 in 1,161 newborn with mutation
Ototoxicity in preterm infants (Zimmerman E, Lahav A, 2012)

- Effects of genetics
  - Iowa Children’s Hospital (Ealy et al 2011)
  - N=703 (1.8% with mtDNA variant)
  - No hearing loss

- Loud noise exposure
  - Animal studies have found potentiating effect between noise and aminoglycosides
Recently published aminoglycoside research

- Designer aminoglycosides prevent cochlear hair cell loss and hearing loss (Huth et al, 2015)

- Aminoglycoside ototoxicity and hair cell ablation in the adult gerbil: A simple model to study hair cell loss and regeneration (Abbas et al, 2015)
Risk Monitoring Program
Goals of risk monitoring program

- Identify infants and children at risk for delayed onset or progressive hearing loss
- Timely diagnostic assessments from a pediatric audiologist
- Maintain a monitoring and tracking system in the state EHDI data management system
Risk Monitoring Program

- Birthing hospitals & Birthing centers
- State EHDI program
- Medical home
- Pediatric Audiology center
Birthing hospitals & Birthing centers

State EHDI program

Medical home

Pediatric Audiology center
Birthing Hospitals/Birthing Center roles:

- Identify infants who have 1 or more risk indicators
- Provide family with referral to pediatric audiology clinic
- Provide the family with information about risk indicators
- Provide the medical home information regarding risk indicator referral
- Report the infants with risk indicators to state EHDI program
Provide on-site hospital/birthing center training

- Provide training annually
  - Physicians
  - Nurse Managers
  - Nurses (Screeners)
  - Midwifes
“Your baby has been identified as having a high risk (_____ for a late-onset hearing loss. The recommended protocol for babies with high risk indicators is an audiological evaluation around 9 months of age. We will provide a copy of this referral form to the pediatric audiology center and they will contact you for an appointment.”
Idaho EHDI Referral forms

STEP 3: RISK ASSESSMENT:

RISK INDICATORS FOR LATE-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
___ Head Trauma ___ Other

(monitoring through age 3 is recommended for most risk factors)
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
- Neuromuscular disorders or sensory motor neuropathies

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 A risk indicators = Recommendation for diagnostic ABR evaluation with pediatric audiologists by 3 months 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 (K.IH.2007)

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

References:
Medical home roles:

- Being familiar with risk factors for delayed onset hearing loss
- Explaining screening results and answer questions for the family
- Encourage risk monitoring follow-up
- Providing family with referral to pediatric audiology clinic
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

**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 otoxic 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 A 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 (KIH 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:

450 W. State St. Floor 5, Boise, ID 83702  www.IdahoSoundsBeginnings@dh.idaho.gov  208-334-0829
Birthing hospitals & Birthing centers

State EHDI program

Medical home

Pediatric Audiology center
Pediatric audiology center roles:

- Providing appropriate comprehensive diagnostic testing for children with risk factors
- Knowledge of risk factors that have high prevalence of delayed onset hearing loss and require early and more frequent assessments
- Providing documentation regarding evaluation outcomes to state EHDI program
Monitoring in audiology clinic

- 5 audiology clinics
- Southwest Idaho and eastern Oregon
- 20 audiologists
Increasing # of hospital referring for risk indicators

2007 & 2008: 2 HOSPITALS

2009 & 2010: 3 HOSPITALS

2011: 4 HOSPITALS
# babies referred for high risk monitoring

- 2007: 272
- 2008: 402
- 2009: 922
- 2010: 993
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Tracking outcomes

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# children
2010

- Normal: 30%
- NR-DC: 39%
- Undetermined: 8%
- Refused: 9%
- Conductive: 7%
- Sensory: 1%
- Neural: 0%
- Elsewhere: 6%
- Deceased: 0%
Birthing hospitals & Birthing centers

State EHDI program

Medical home

Pediatric Audiology center
State EHDI program roles:

- Providing training and support for hospitals, birthing center, physicians, and pediatric audiologists on risk factor

- Providing a method for hospitals, birthing centers and pediatric audiologists to report information regarding infants with risk indicators to the state EHDI program

- Tracking and surveillance of infants with risk factors
Idaho EHDI program
Data collected by referral forms
Prevalence of Infants with a Risk Indicator in ISB 2007-2013 Data

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<th>Year</th>
<th>% infants WITH risk factors</th>
<th>% infants WITHOUT risk factors</th>
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Number of Risk Indicators Reported in ISB 2007-2013 Data

- Neurodegenerative disorders: 79.63%
- Head trauma: 8.64%
- Chemotherapy: 5.19%
- Postnatal infection: 3.02%
- Physical findings: 0.91%
- Syndromes: 0.86%
- Mechanical ventilation: 0.28%
- Craniofacial anomalies: 0.10%
- Neonatal indicators: 0.01%
Ototoxic medication

- January 2008-December 2014
- 4701 infants passed newborn hearing screening (Ototoxic medication only)
- 2 with diagnosed with delayed-onset hearing loss
  - 1 unilateral severe to profound
  - 1 bilateral mild to moderately severe
Idaho EHDI: Diagnostic testing recommendations for infants with risk indicators

Audiology Assessment for Risk Factor Follow-up

"The timing and number of hearing evaluations for children with risk factors should be customized and individualized depending on the relative likelihood of a subsequent delayed-onset hearing loss."

(ICH 2007 Position Statement)

Early and more frequent assessment may be indicated for children with: cytomegalovirus (CMV) infection, syndromes associated with progressive hearing loss, neurodegenerative disorders, trauma, culture-positive postnatal infections in association with documented hearing loss, for children who have received ECMO or chemotherapy, and when there is a caregiver concern or a family history of hearing loss (ICH 2008 classification)

Recommended Assessment Standards:

Behavioral testing at 9 months of age**

All testing should be ear-specific.

Tests included in this evaluation are:

- Family/child history
- Otoscopy
- Visual Reinforcement Audiometry for each ear:
  - Minimal Response levels for air conduction: 500, 2000 and 4000 Hz
  - Bone conduction as needed to rule out conductive pathology
- Speech Awareness Thresholds (SAT)
- Limited Octaves Emissions, DPOAE and/or TEOAE
- Immitance battery:
  - 226 Hz probe tone tympanometry each ear.
  - Tympanic acoustic reflexes at 500, 1000 and 2000 Hz (can also use broadband noise reflex - normal in less than 80 dB HL)
- ABR testing is indicated if hearing loss is diagnosed, or if responses to behavioral audiometry are not reliable.


**The recommendation for the initial risk factor evaluation to be done at 9 months of age is based on the following factors:
- The use of Visual Reinforcement Audiometry for the child and family.
- The ability to gather the greatest amount of information quickly with minimal input stimuli, balanced with...
- The ability to identify and address hearing losses and corresponding concerns early enough during the critical language learning period to maximize communication skills and minimize speech and language delays. Testing of a 2 year old can also be difficult, time consuming and delay identification.

Risk Indicators Associated with Permanent Congenital, Delayed onset, or Progressive Hearing Loss in Childhood

1. Caregiver concern reporting hearing, speech, language or developmental delay
2. Notable medical conditions (e.g., meningitis, congenital infections, metabolic disease, etc.)
3. Family history of permanent childhood hearing loss
4. Neural tube defect
5. Traumatic brain injury
6. Physical findings, such as craniofacial anomalies, that are associated with a syndrome known to include a sensorineural or permanent conductive hearing loss.
7. Syndromes associated with hearing loss or progressive or late onset hearing loss such as neurofibromatosis, osteogenesis imperfecta, and Usher syndrome, other frequently identified syndromes including Waardenburg, Alport, Bardet-Biedl, and Pendred and Laga-Gallman
8. Neurodegenerative disorders, such as Wilson syndrome, or sensory motor impairment, such as Friedrich ataxia and Charcot-Marie-Tooth syndrome
9. Culture-positive postnatal infections associated with sensorineural hearing loss, including confirmed bacterial and viral (especially herpes, varicella and cytomegalovirus)
10. Head trauma, especially brain; intrauterine and transport
11. Chromotherapy

Risk factors identified in this document are considered to have a greater concern for delayed onset hearing loss and monitoring of these children should be more frequent once the period following the neonatal period.
Idaho EHDI: Diagnosed hearing loss

- # infants w/hearing loss & HIGH RISK INDICATORS
- # infants w/hearing loss & NO RISK INDICATORS

Year: 2007 - 2010
- 2007: 16 (High Risk), 28 (No Risk)
- 2008: 20 (High Risk), 23 (No Risk)
- 2009: 18 (High Risk), 33 (No Risk)
- 2010: 32 (High Risk), 27 (No Risk)
Idaho data (2007-2011)
Delayed onset hearing loss

- 2.7 infants per 10,000 diagnosed with delayed onset hearing loss with risk indicators
  - Those with hearing loss the most frequently reported risk indicators were NICU stay (15 infants), ototoxic medications (13 infants)
# of risk indicators reported in infants with hearing loss

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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 indicators = 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 (JIHC 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:

450 W. State St. Floor-5, Boise, ID 83702 www.IdahoSound@beginnings@dhw.idaho.gov 208-334-0829
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)
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* 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.
Class A risk indicators (n= 153)

- LTF/LTD 60%
- Normal hearing 24%
- Hearing loss 16%
In utero & postnatal infections (n = 18)

- LTF/LTD 33%
- Normal hearing 45%
- Hearing loss 22%
Syndromes (n=13)

- LTF/LTD: 23%
- Normal hearing: 23%
- Hearing loss: 54%
Craniofacial anomalies (n=114)

- LTF/LTD: 69%
- Normal hearing: 20%
- Hearing loss: 11%
Things to remember

- Risk monitoring programs need participation from hospital, birthing centers, medical homes, audiology centers, and state EHDI programs

- Training from state EHDI programs is important

- No “gold standard” protocol for risk indicator monitoring (ototoxic medications)

- Data collection is an important
Case #1

- 4 yr old female
- Reason for referral: Speech delays
  - Speech therapy twice per week
- Birth history:
  - 32 weeks gestation (2lb 6oz)
  - NICU stay 1 month
  - Ototoxic medication (Gentamicin)
  - Passed AABR hearing screening
    - In 2004, Idaho did not have risk monitoring program
Audiometry

Speech audiometry
SRT at 10 dBHL in each ear

Tympanograms
Type As bilaterally

Ipsilateral MEMR
Absent bilaterally
DPOAE
TEOAE
3 years later...

- 8 years old
- Physician requested audiogram due to previous recommendations
- Mom has no significant hearing or speech concerns, but patient
  - Frequently asks for repetition
  - Listens to TV “very loud”
Audiometry

Speech Audiometry:
Word recognition @ 50 dBHL- 84% right, 100% left
SRT- 10 dBHL right, 0 dBHL left

Tympanograms: Type A bilaterally

MEMR:
- Ipsilateral left/right present
- Contralateral right present
- Contralateral left absent
DPOAE

[Graph showing DPOAE measurements with frequency and dB on the axes.]

Take Care Forward.
TEOAE
Case #2

- Passed AABR hearing screening
- Born at 35 weeks 6/7 days
- NICU stay less than 5 days
- Referred to audiology for risk indicator monitoring (Ototoxic medications)
- No family history of childhood hearing loss
- No history of otitis media
Audiology Evaluation
9 months old
OAE tracing

Right: 22-Jan-13: -: 750-8000 Hz Diagnostic Test - High Noise: 13A22D01.OAE

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Left: 22-Jan-13: -: 750-8000 Hz Diagnostic Test - High Noise: 13A22D00.OAE

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ABR evaluation
10 months old
ABR tracing
ABR tracing
ABR eHL
10 months old, 18 months old
3 years old
Questions and Answers
REFERENCES

- http://www.ncham.org
- The NCHAM book, Chapter 10, Risk monitoring for Delayed-Onset hearing loss.