RISK MONITORING FOR DELAYED-ONSET HEARING LOSS

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Did U Check?
newborn hearing screening

Services provided by St. Luke’s
Disclaimer

- I have no relevant financial relationships with the manufacturers of any commercial products and/or provider of commercial services discussed in this CME activity.

- I do not intend to discuss an unapproved/investigative use of a commercial product/device in my presentation.
Joint Committee on Infant Hearing (JCIH)

JCIH was established in 1969

Comprised of:
- American Academy of Pediatrics
- American Academy of Ophthalmalogy and Otolaryngology
- American Speech & Hearing Association
JCIH 1972 Position Statement

- High risk criteria
  - Family history of childhood hearing loss
  - Intrauterine fetal infection (Rubella)
  - Defects of ear, nose or throat (atresia, cleft lip/palate)
  - Low birth weight (<1500 grams)
  - High bilirubin levels
JCIH 1982 Position Statement

– High risk criteria
  » Bacterial meningitis, severe asphyxia (i.e. low APGAR) were added

– Screening recommendations
  » Ideally performed by 3 months (no later than 6 months)
  » Preferably under the supervision of an audiologist
  » Observation of behavioral or electrophysiologic response to sound
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
Studies have shown that only 50% of all hearing loss were being identified using the High Risk Register.
JCIH 2000 Position Statement

- Recommended ALL infants screened before hospital discharge

- Risk monitoring:
  - Audiological testing every 6 months until age 3 years.
Joint Committee on Infant Hearing (JCIH)

YEAR 2007 POSITION STATEMENT: Principles and Guidelines for Early Hearing Detection and Intervention Programs
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**
JCIH 2007 Position Statement

- 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.”
JCIH 2007
Appendix 2: RISK INDICATORS FOR HEARING LOSS

- 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
RISK INDICATORS FOR HEARING LOSS
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%)

(Cone-Wesson, et al., 2000; Hall, 2007)
Least frequently occurring risk factors (<10%)

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

(Cone-Wesson, et al., 2000; Hall, 2007)
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%)

(Cone-Wesson, et al., 2000; Fligor, 2008; Hall, 2007)
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)
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
Neonatal Intensive Care (NICU)

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)**
Neonatal indicators

Cone-Wesson et al 2000 - estimates 1/56 children with permanent hearing loss at age 1, had the following risk factors: respiratory distress syndrome, Bronchiopulmonary dysplasia, Mechanical Ventilation >36 days

Beswick et al 2013 – found a correlation between postnatal hearing loss and prolonged ventilation (≥5 days)
ECMO treatments

- Extracorporeal 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 (Fligor, 2008)
Physical findings/Syndromes associated with hearing loss

- Waardenburg syndrome
- Branchio-Oto-Renal (BOR) syndrome
- Stickler syndrome
- CHARGE syndrome
- Neurofibromatosis Type II
- Downs syndrome
- Treacher Collins syndrome
- Usher syndrome
- Pendred syndrome
- Alport syndrome
- Jervell Lange-Nielsen
Infections

**Congenital Infections**
- Cytomegalovirus (CMV)
- Rubella
- Herpes
- Syphilis
- Toxoplasmosis

**Postnatal infections**
- Bacterial or viral meningitis
- Varicella
- Herpes viruses
Craniofacial anomalies

- Head trauma
- Recurrent OME
- Cleft palate
- Abnormal pinna
- Abnormal ear canal
- Ear tags and pits
- Malformed eyes
- Choanal atresia
- Craniosynostosis
- Hemifacial microsomia
Incidence of hearing loss in cleft palate patients

Children’s Hospital of Philadelphia (1972-1976)
n = 70 (2 - 21 year olds) with cleft palate
50% conductive hearing loss

n = 90 infants with cleft palate
82% (74) hearing loss (varying from mild to severe)
18% (16) normal hearing

Australia study (Beswick, et al. 2013) showed that the risk factor of craniofacial anomalies (cleft palate, excluding cleft lip and skins tags) did predict the occurrence of postnatal hearing loss.
Idaho Cleft Palate and Craniofacial Deformities team
(Oct 2007- Feb 2010)

N = 210

104 (Normal hearing) = 50%
   At least 50% of these children have a history of OME and PE tubes
94 (Conductive hearing loss) = 45%
   2 bilateral microtia
4 (Mixed hearing loss) = 1%
8 (Sensorineural hearing loss) = 4%
   3 unilateral – profound left ear
   5 bilateral (1 cochlear implant)
Family History

- Positive family history of congenital hearing loss or hearing loss acquired during childhood

- Family history of hearing loss is the most common risk indicator found in healthy newborns (Hall 2007)

- Australia study (Beswick, et al. 2013) showed that the risk factor of family history did predict the occurrence of postnatal hearing loss
Head trauma

- Involving basal skull/temporal fracture that requires hospitalization
Neurodegenerative disorders/Sensory motor neuropathies

- Hunter syndrome
- Charcot Marie Tooth disease
- Friedreich ataxia
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
Hospitals/Birthing Center roles:

- Identify infants who have 1 or more risk indicators
- Provide family with referral to pediatric audiology clinic
- Provide family with information about risk indicators
- Provide medical home information regarding risk indicator referral
- Report infants with risk indicators to state EHDI program
EHDI program roles:

- Provide training/education
  - Physicians
  - Nurse Managers
  - Nurses (Screeners)
  - Midwifes

- Monitor hospital data and audiology outcomes
  - Give hospitals feedback
“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.”
Provide 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, Osteogenesis, 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 (XH 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-3, Boise, ID 83702  www.istatospundteppings@idaho.istatospund 208-334-0529
Birthing hospitals & Birthing centers

State EHDI program

Pediatric Audiology center

Medical home
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
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
Idaho EHDI: Diagnostic testing recommendations for infants with risk indicators

Recommended Minimum 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 Otoacoustic Emissions, DPOAE and/or TEOAE
- Immittance battery:
  - 226 Hz probe tone tympanometry-each ear.
  - Ipsilateral acoustic reflexes at 500, 1000 and 2000 Hz (can also use broadband noise reflex – normal is less than 80 dB HL)
- ABR testing is indicated, if hearing loss is diagnosed, or if responses to behavioral audiometry are not reliable.

Monitoring risk indicators in audiology clinic

- 5 audiology clinics
- Southwest Idaho and eastern Oregon
- 20 audiologists
# babies referred for high risk monitoring

- 272 in 2007
- 402 in 2008
- 922 in 2009
- 993 in 2010
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<th>NAME</th>
<th>DOB</th>
<th>MR #</th>
<th>1st ATTEMPT</th>
<th>2nd ATTEMPT</th>
<th>3rd ATTEMPT</th>
<th>RESULTS</th>
<th>SENT TO EHD</th>
<th>Au.D.</th>
<th>RFAM</th>
<th>NEONATAL</th>
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<td>OTO</td>
<td>CRANIC</td>
<td>SYND</td>
<td>NEUR</td>
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</table>

- **NAME**: The name of the individual.
- **DOB**: Date of Birth.
- **MR #**: Medical Record Number.
- **1st ATTEMPT**: Date of the first attempt.
- **2nd ATTEMPT**: Date of the second attempt.
- **3rd ATTEMPT**: Date of the third attempt.
- **RESULTS**: Result of the hearing test.
- **SENT TO EHD**: Whether the result was sent to EHDI.
- **Au.D.**: Audiologist's name.
- **RFAM**: Referral to Family Medicine.
- **NEONATAL**: Neonatal conditions.
- **UTERC**: Uterine conditions.
- **OTO**: Otorhinolaryngology conditions.
- **CRANIC**: Craniofacial Syndromes.
- **SYND**: Neurological disorders.
- **NEUR**: Neurological conditions.

The table lists the names and details of patients with hearing test results. Each row represents a patient and their test results, including dates and referrals.
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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

### 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)
Prevalence of Infants with a Risk Indicator in ISB 2007-2013 Data

<table>
<thead>
<tr>
<th>Year</th>
<th>% Infants WITH risk factors</th>
<th>% Infants WITHOUT risk factors</th>
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<tbody>
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<tr>
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<td>2013</td>
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<td>88.6</td>
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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)
CLASS A & CLASS B RISK INDICATOR MONITORING
CLASS A & CLASS B

- Audiologist and NICU physicians began discussion – May 2011
- Developed guidelines – May-October 2011
- Two hospitals implemented – October 2011
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.
DATA 2012-2013

- $N = 10,634$ babies
- $= 1.6\%$ 175 babies with CLASS A risk indicator
- $= 11.04\%$ 1175 babies with any risk indicator (CLASS A and/or CLASS B)
% babies with reported risk indicators (2012-2013)

- Total Babies: 89%
- Risk Babies: 11%
Risk indicator occurrence

N=1175 babies

- CLASS B: OTOTOXIC MEDICATIONS: 734
- CLASS B: REPORTED FAMILY HISTORY OF HL: 175
- CLASS A: 175
Class A risk indicators

N=175 babies

- 50% LOST TO FOLLOW-UP
- 50% TESTED
Class A risk indicators

N=87 babies tested

- NORMAL HEARING: 75%
- SNHL/MIXED: 6%
- CONDUCTIVE: 19%
Class B/Ototoxic Medications

N=743 babies

- 46% LOST TO FOLLOW-UP
- 54% TESTED
Class B/Ototoxic Medications

N=345 babies tested

- NORMAL HEARING: 83%
- SNHL/MIXED: 15%
- CONDUCTIVE: 2%
Class B/Family History

N=175 babies

- 37% LOST TO FOLLOW-UP
- 63% TESTED
Class B/Family History

N = 65 babies tested

- 83% NORMAL HEARING
- 11% SNHL/MIXED
- 6% CONDUCTIVE
Barriers to risk indicator monitoring in EHDI programs
Barriers...

- Accurate reporting by hospital staff
- Accurate reporting by families (i.e. family history)
- Accurate and timely reporting by audiologists
- Lack of pediatric audiologists
- High Lost-To Follow-Up rates
- Lack of support by medical homes
- No “gold standard” protocol for audiological monitoring of risk indicators
Why do we monitor children with risk indicators for delayed-onset hearing loss…
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
Otoacoustic Emissions
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
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
Otoacoustic Emissions
ABR evaluation
10 months old
ABR eHL
3 years old
Points to remember

- EHDI risk monitoring programs need active participation from hospitals/birthing centers, medical homes, audiology centers, and state EHDI programs.

- EHDI programs need to provide training and guidance for all participants (hospitals/birthing centers, medical homes, audiology centers).

- Data collection is an important part of the program.
Questions and Answers
REFERENCES

- http://www.ncham.org