Risk Factors for Late-Onset Hearing Loss in Children

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In collaboration with Washington State DOH EHDDI Program

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Specific Objectives

1. Evaluate the efficacy of the JCIH 2000 recommended neonatal risk indicators for progressive and/or late onset hearing loss.

2. Ensure the accuracy of reporting of the JCIH 2000 neonatal risk indicators for progressive and/or late onset hearing loss by hospitals by implementing quality control measures.

3. Evaluate the compliance with the JCIH 2000 recommendations for monitoring and assessment by the child’s primary care physician and parents.
Why Track Risk Factors?

<table>
<thead>
<tr>
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</thead>
<tbody>
<tr>
<td>Birth</td>
<td>Age 3 Years</td>
<td>Age 10 Years</td>
</tr>
<tr>
<td>0.3%</td>
<td>0.67%</td>
<td>1.38 %</td>
</tr>
<tr>
<td>3/1000</td>
<td>7/1000</td>
<td>14/1000</td>
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</table>

Normal hearing at birth does not rule out a delayed onset hearing loss later.
Neonatal (birth–28 days) risk indicators for late-onset hearing loss

1. An illness or condition requiring admission of 48 hours or greater to a NICU.
2. Stigmata or other findings associated with a syndrome known to include a sensorineural or conductive hearing loss.
3. Family history of permanent childhood sensorineural hearing loss.
4. Craniofacial anomalies, including those with morphologic abnormalities of the pinna and ear canal.
5. In-utero infections such as cytomegalovirus, herpes, toxoplasmosis, or rubella.

Joint Committee on Infant Hearing, 2000
Data Collection and Analysis
EHDDI Tracking & Surveillance Database

- Hospitals report screening & risk factor information to Department of Health.
- DOH follows up with PCP for babies referred, missed, and babies who passed but are reported to have 1 or more of 4 specified risk factors for late onset hearing loss.
- DOH does not follow infants who pass newborn hearing screening whose only risk factor is NICU stay >48 hours due to volume.
RISK FACTORS FOR HEARING LOSS

If an infant meets one or more of the following risk factors listed below, check the corresponding box(es) on the front of the card. If unknown, leave boxes blank.

1. Illness or condition requiring admission of 48 hours or greater to a neonatal intensive care unit (NICU).
2. Stigmata or other findings associated with a syndrome known to include a sensorineural and/or conductive hearing loss.
3. Family history of permanent childhood sensorineural hearing loss.
4. Craniofacial anomalies, including those with morphological abnormalities of the pinna and ear canal.
5. In-utero infections such as cytomegalovirus, herpes, toxoplasmosis, or rubella.
PATIENT

Child: @CHILD_NAME @MOTHER_LAST_NAME
Date of Birth: @DOB
Mother: @MOTHER_FIRST_NAME @MOTHER_LAST_NAME
Birth Hospital: @SUBMITTER_DESCRIPTION
EHDDI #: @UNIQID

EHDDI PROGRAM COMMENTS
This patient was born at a hospital currently reporting their hearing screen results to the Washington State Department of Health Early Hearing Loss Detection, Diagnosis, and Intervention Program.

1. SCREEN RESULTS: This patient passed the hearing screen.

2. RISK FACTORS: This patient was reported to have one or more of the following risk factors associated with late-onset hearing loss as outlined by the Joint Committee on Infant Hearing 2000:
   - Spent 48 hours or longer in NICU
   - Syndromic Syndromes
   - Family History of Hearing Loss
   - Maternal Illness During Pregnancy
   - Craniofacial Anomalies

3. RECOMMENDATIONS: Based on the above information it is recommended that this patient receive routine hearing screening with an audiologist every six months until the age of three.

A list of pediatric audiology clinics has been included for your convenience.

PROVIDER COMMENTS - Please indicate that you have seen and fax a copy of this report to the EHDDI Program at (206) 418-5415. Thank you.

- [ ] I am not currently seeing this patient.
- [ ] I have shared the above information and screening recommendations with the patient's parent or legal guardian.
- [ ] OTHER.

For questions or concerns please call the EHDDI program at (206) 418-5613. Please visit our website at www.doh.wa.gov/ehddi for more information about the EHDDI program and the importance of hearing screening.

Report Code: RTYP
Phase II
Data Collection and Analysis

- Audiologists report detailed hearing health information and history for 0-3 year olds seen for diagnostic hearing evaluations via web-based application.
Specific Aim 1

Evaluate the efficacy of the Joint Committee on Infant Hearing 2000 recommended neonatal risk indicators for progressive and/or late-onset hearing loss.
Risk Factors for Newborns in EHDDI Database 2005-2006
Total screened = 147,431
Infants with one or more risk factors = 13,251 (9%)

DOH Sends Risk Factor Letter to PCP

- NICU > 48 Hours: 74%
- Syndrome: 2%
- Family History: 20%
- Craniofacial Anomalies: 4%
- In-Utero Infections: 3%
NICU Stay as a Surrogate

Incidence of Hearing Loss by Risk Factor

- Syndromes: 26%
- Mechanical Vent > 5 days: 7.6%
- Low apgar at 1 or 5 min: 6.4%
- Meningitis: 6.2%
- Aminoglycoside >5 days: 6%
- Hyperbilirubinemia (XT): 8.9%
- BW < 1500g: 7.8%
- Craniofacial Anomaly: 17.6%
- Congenital Infection: 6.9%
- Family History: 11%

Cone-Wesson et al., 2000
Risk Factor Status for all Infants with Hearing Loss (N=482)

- Total infants with one or more risk factors: 77
- NICU >48 hours: 46
- Syndrome: 11
- Family History: 20
- Craniofacial Anomalies: 21
- In-Utero Infections: 2

(16%)
Risk Factor Status of Infants who Passed Newborn Hearing Screen & were Later Diagnosed with Permanent Hearing Loss (N=31)

52% had one or more risk factor
48% had no risk factors

- NICU > 48 Hours: 13
- Syndrome: 8
- Family History: 3
- Craniofacial Anomalies: 5
- In-Utero Infections: 1
- No Risk Factors: 1
- Unknown: 3

Number of Infants
Misses or Late-Onset/Progressive Hearing Loss?

Possible Misses?

- **Screening until a pass is obtained.**
- Screening tool insensitive to degree and/or configuration of hearing loss.
- Recording error by screener.
Misses or Late-Onset/Progressive Hearing Loss?

At least five of the babies failed their initial hearing screening and then “passed” a re-screening

- Multiple inpatient screens until a pass
- Failed AABR as an inpatient and passed DPOAE as an outpatient (2 cases)
- Failed TEOAE passed outpatient AABR re-screen
- Failed RE DPOAE, passed LE. Then passed RE, failed LE. Counted as a pass bilaterally.
Infants with Hearing Loss who Passed Newborn Hearing Screening (N=31)

- ABR: 42%
- DPOAE: 35%
- TEOAE: 19%
- Unknown: 3%

Final Screening
Test Type for All Infants

- ABR: 35%
- DPOAE: 25%
- DPOAE & ABR: 28%
- TEOAE: 12%

Number of Births

Screening Protocol
Degree of Hearing Loss as a Function of Screening Protocol

<table>
<thead>
<tr>
<th></th>
<th>ABR</th>
<th>DPOAE</th>
<th>TEOAE</th>
<th>Unknown</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mild</td>
<td>6</td>
<td>6</td>
<td>3</td>
<td></td>
</tr>
<tr>
<td>Moderate</td>
<td>4</td>
<td>2</td>
<td>1</td>
<td></td>
</tr>
<tr>
<td>Severe</td>
<td>2</td>
<td></td>
<td>1</td>
<td></td>
</tr>
<tr>
<td>Profound</td>
<td></td>
<td>1</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Unknown</td>
<td>1</td>
<td>2</td>
<td>1</td>
<td>1</td>
</tr>
</tbody>
</table>
Specific Aim 2

Ensure the accuracy of reporting of the Joint Committee on Infant Hearing 2000 neonatal risk indicators for progressive and/or late onset hearing loss by hospitals by implementing quality control measures.
Hospital Quality Control & Education

- Site visits to all screening hospitals.
- Review risk factors with screeners.
- Review overall screening program.
- Conduct training for programs with high refer rates.
- Re-train screeners when needed.
Inconsistencies in Risk Factors Reported for Children with Hearing Loss (N=482)

- Screener Reported RF
- Audiologist Reported Any RF in Phase II
- Audiologist Reported Same RF as Screener in Phase II

NICU
- Syndrome
- Family History
- Craniofacial Anomalies
- In-Utero Infections
### Inconsistencies in Risk Factors Reported for Children with Hearing Loss (N=482)

<table>
<thead>
<tr>
<th>Risk Factor</th>
<th>Screener Reported</th>
<th>Audiolgist Reported Any RF in Phase II</th>
<th>Audiolgist Reported Specific RF in Phase II</th>
</tr>
</thead>
<tbody>
<tr>
<td>NICU &gt; 48 hours</td>
<td>19.7%</td>
<td>9.5%</td>
<td>7.5%</td>
</tr>
<tr>
<td>Syndrome</td>
<td>4.1%</td>
<td>2.3%</td>
<td>0%</td>
</tr>
<tr>
<td>Family History</td>
<td>8.1%</td>
<td>4.1%</td>
<td>3.7%</td>
</tr>
<tr>
<td>Craniofacial Anomalies</td>
<td>7.7%</td>
<td>4.4%</td>
<td>4.1%</td>
</tr>
<tr>
<td>In-Utero Infections</td>
<td>0.8%</td>
<td>0.4%</td>
<td>0.2%</td>
</tr>
<tr>
<td><strong>Total</strong></td>
<td><strong>40.5%</strong></td>
<td><strong>20.7%</strong></td>
<td><strong>15.6%</strong></td>
</tr>
</tbody>
</table>
Risk Factor Reporting Issues: Hospitals & Screeners

- Disconnect between person completing DOH card & person knowledgeable about risk factors.
- Unclear when to check NICU.
- Checking in-utero infection if mother was ill anytime during pregnancy.
- Indicating family history of hearing loss if history of otitis media.
Risk Factor Reporting by Hospitals

Percentage of Time Risk Factors are Completed by Hospitals

- 2006: 93%
- Pre-2006: 78%
Risk Factor Reporting Issues: Audiologists

- Time required to enter data into Phase II
- Inaccurate parent report of medical history
Missing Information in Phase II

Blank Data Elements in Phase II for Confirmed Hearing Loss
(includes multiple visits for ~130 patients)
Missing Information in Phase II

Phase II Risk Factors Reported

- Risk factor reported: 123
- Risk factor not reported: 175
- Patient reported as having no risk factors: 184
- Total: 482
Specific Aim 3

Evaluate the compliance with the JCIH 2000 recommendations for monitoring and assessment by the child’s primary care physician and parents.
Number of Patients born in 2005-2006 for whom a Risk Factor Letter was sent to PCP = 2984

- Normal: 375
- Info Shared with Parent: 177
- Lost: 692
- Moved: 25
- Refused: 8
- Expired: 1
- Recorded Audiologic Evaluation: 39
Infants With RF 2-5 Who Passed UNHS
PCP Sent RF Letter by DOH

PCP Shared Information with Parents

Referred to Audiology

Hearing Eval Recorded

Pre 2005

2005-2006

57%

68%

6%

1.3%

6%

1.3%
Questions - - -

- Is all the data being reported to DOH?
- Are physicians referring?
- Are families following through?
Survey of Physicians

- Sent to 650 physicians with 1 or more children in their practice who have risk factors and passed NHS.
- Purpose: To evaluate attitudes and experiences towards follow-up.
- 190 surveys were returned (29%).
- Majority indicated they share information with the parent, monitor hearing at well-child checks, and refer to audiology as indicated.
Physician Perspective on JCIH (2000) Recommendations for Follow-up of Infants with Risk Factors

The recommendations are critical to identifying hearing loss early in children. I strongly encourage families to follow-up accordingly.  

<table>
<thead>
<tr>
<th>Response</th>
<th>Count</th>
<th>Percentage</th>
</tr>
</thead>
<tbody>
<tr>
<td>The recommendations are critical to identifying hearing loss early in children. I strongly encourage families to follow-up accordingly.</td>
<td>108</td>
<td>57%</td>
</tr>
<tr>
<td>The recommendations are of unproven benefit. I inform families about recommended follow-up and leave it up to them to pursue.</td>
<td>50</td>
<td>26%</td>
</tr>
<tr>
<td>The recommendations are too burdensome and a poor use of healthcare resources.</td>
<td>11</td>
<td>6%</td>
</tr>
</tbody>
</table>

Comments include depends on risk factor (1), all of the above (1), none of the above (2), don’t know (1), don’t know about these recommendations (1)
Accuracy in Reporting Risk Factors by Birth Hospital as Judged by PCP

- Almost Always Accurate: 32%
- Sometimes Accurate: 43%
- Rarely Accurate: 15%
- Never Accurate: 3%
Importance of Each Risk Factor in Identifying Hearing Loss as Judged by PCP

- NICU Stay: > 48 hours
- Syndrome Family History
- Craniofacial Anomalies
- In-Utero Infections

Legend:
- Not Important
- Moderately Important
- Very Important
Action(s) taken by PCP upon receiving a letter from DOH about infant with risk factors for late onset and progressive hearing loss

- 77% Verify risk factor
- 86% Share with parent
- 63% Monitor Hearing

Timeframe of referral by PCPs who indicated they refer to an Audiologist (N=139)
Survey of Physicians
Most Common Barriers to Follow-up

- Family compliance (33%)
- Cost of follow-up/insurance coverage (18%)
- Physician compliance (14%)
- Availability of local pediatric audiology (13%)
- Lack of stable medical home (10%)
- Accuracy in reporting risk factors (7%)
- Frequency of Audiology visits too high (5%)
Survey of Physicians
Reasons for Poor Family Compliance

- Unspecified - 40%
- No concern about baby’s hearing – 33%
- Too time consuming – 25%
- Awareness/Understanding of importance – 10%
- Other (≤ 4% each) – logistics of making/keeping appointments; transportation, language & socioeconomic barriers
Survey of Physicians
Reasons for Poor Physician Compliance as Judged by Physicians

- Unspecified - 14%
- Awareness/Understanding – 38%
- Time/forget – 38%
- Other (≤ 4%) – not enough hearing loss to warrant; lack of accuracy in reporting risk factors; lack of evidence to support; family history is a poor indicator
Other Activities
Audiology Workshops

- **2005**
  - Project Overview
  - Data Update from DOH
  - Linda Ramsdell - Genetics
  - Ann Melvin - CMV
  - Kathy Sie - Medical Work-up of SNHL
  - Jeff Stolz - NICU Babies

- **2006**
  - Judy Widen – JCIH
  - Karen Fowler – CMV
  - Kathy Sie – EVAS
  - Data Update from DOH
2007 Audiology Workshop

- Friday, August 10, 2007
  - Betty Vohr – NICU babies and hearing loss
  - Henry Ou – Ototoxicity
  - Kathy Sie – Sensorineural Hearing Loss
  - National Speaker – Genetics (to be confirmed)
  - Data Update from DOH

- Graham Visitor Center in Washington Park Arboretum (UW)
- Travel stipends will be available
Physician Education

- Presentations to pediatric practices
- WCAAP Annual CME Meeting
- Developing brochure about risk factors
- DOH developed risk factor info sheets
JCIH 2007 Statement
JCIH 2007 Position Statement

- In final stages of approval by member organizations

- Changes affecting risk factors
  - NICU stay >5 days.
  - For infants with risk factors, at least one diagnostic evaluation before 30 months of age.
  - Repeat hearing screen recommended for re-admissions with risk factors.
Summary

• ~50% of infants who pass NHS and are later identified with hearing loss have one or more JCIH 2000 risk factors. More work to be done in evaluating specifics of risk factors, and whether there are other factors involved. (i.e. CMV, EVA, false passes, genetics)

• Improvements can be made in accurate identification of risk factors by hospital screening staff, as well as data reporting by audiologists.

• Physicians see importance of follow-up for infants with risk factors. However, there are compliance issues for both parents and physicians primarily around time, awareness, understanding, cost, and availability of services.