HEARING SCREENING
IN THE MEDICAL HOME

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Alabama AAP EHDI Chapter Champion
Disclosures

- I have no relevant financial relationship 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.
Objectives

- Understand rationale behind early hearing loss detection and intervention
- Discuss current hearing screening and follow-up guidelines
- Learn about medical home role in hearing screening
- Ella’s Story
Why is Early Identification of Hearing Loss so Important?

- **Hearing loss is the most frequent birth defect.**
  - Every day, 33 babies are born in the US with permanent hearing loss.
    - 3/1000 newborns
    - In Alabama, 60 to 180 babies are born with hearing loss each year.
  - Incidence increases to 6/1000 by school age
    - Late diagnosis
    - Late onset
    - Progressive hearing loss
  - 10/1000 incidence in NICU patients
Incidence per 10,000 of Congenital Defects/Diseases

- Hearing Loss
- Cleft Lip/Palate
- Down Syndrome
- Spina Bifida
- Sickle Cell Anemia
- PKU
Why is Early Identification of Hearing Loss so Important?

- Hearing loss is the most frequent birth defect.
- Undetected hearing loss has serious negative consequences.
  - Prior to newborn hearing screening, the average age children with more severe hearing losses were diagnosed was ~22 months.
    - Milder and unilateral hearing losses often not diagnosed until school age.
  - Children with hearing loss are at risk for poorer language, academics, social skills, and psychological outcomes.
Understanding What It’s Like to Have a Hearing Loss

Severe hearing loss

Moderate hearing loss

Mild hearing loss

Normal hearing
<table>
<thead>
<tr>
<th>Degree of Hearing Loss</th>
<th>Hearing Level (dB)</th>
<th>Effects</th>
</tr>
</thead>
<tbody>
<tr>
<td>Normal</td>
<td>0 to 15</td>
<td>• Can detect all aspects of speech</td>
</tr>
</tbody>
</table>
| Minimal                | 16 to 25          | • *May miss up to 10% of speech*  
                          |                   | • May respond inappropriately  
                          |                   | • Peer social interaction affected |
| Mild                   | 26 to 40          | • *May miss up to 50% of speech*  
                          |                   | • May be labeled as "behavior problem" and "poor listener" |
| Moderate               | 41 to 55          | • *May miss 50% to 100% of speech*  
                          |                   | • Speech quality likely to be poor  
                          |                   | • Vocabulary is limited  
                          |                   | • Compromised communication ability |
| Moderate/Severe        | 56 to 70          | • *100% of normal volume speech lost*  
                          |                   | • Delayed speech and poor intelligibility |
| Severe                 | 71 to 90          | • *Loud voices only heard within 12in of ear*  
                          |                   | • Delayed speech and language  
                          |                   | • Declining speech abilities and atonal voice |
| Profound               | >90               | • Sound vibrations felt rather than |
                          |                   | heard |


Vocabulary Development in Infants and Toddlers

All Degrees of Hearing Loss Place Children at Risk

- Children with a unilateral hearing loss are 10x more likely to be held back at least one grade

- Children with mild losses:
  - 37% fail one grade
  - 8% don’t have skills at grade level
  - 12-41% receive educational assistance
Effects of Unilateral Hearing Loss

- Keller & Bundy (1980) (n = 26; age = 12 yrs)
- Peterson (1981) (n = 48; age = 7.5 yrs)
- Bess & Thorpe (1984) (n = 50; age = 10 yrs)
- Blair, Peterson & Viehweg (1985) (n = 16; age = 7.5 yrs)
- Culbertson & Gilbert (1986) (n = 50; age = 10 yrs)

Average Results:
- Math = 30th percentile
- Language = 25th percentile
- Social = 32nd percentile
Why is Early Identification of Hearing Loss so Important?

- Hearing loss is the most frequent birth defect.
- Undetected hearing loss has serious negative consequences.
- **There are dramatic benefits associated with early identification of hearing loss.**
Early Intervention prevents or minimizes communication delays

- Moeller examined 112 children with hearing loss enrolled in intervention at various ages
  - Children who were enrolled earliest (by 11 months of age) demonstrated significantly better vocabulary and verbal reasoning skills at 5 years than did later-enrolled children
  - Regardless of degree of hearing loss, early-identified children achieved scores that approximated those of their hearing peers
    - Most successful children also had high levels of family involvement

Boys Town National Research Hospital Study of Earlier vs. Later

129 deaf and hard-of-hearing children assessed 2x each year.

Assessments done by trained diagnostician as normal part of early intervention program.

Moeller, M.P. (1997). Personal communication moeller@boystown.org
Vocabulary at Age 5 by Age of Intervention

![Bar Chart]

- Average range

Age of Enrollment in Services (in Months):
- ≤ 11
- 11.1–23
- 23.1–35
- > 35
Yoshinaga-Itano, et al., 1998

- Compared language abilities of hearing impaired children identified before 6 months of age (n=72) with similar children identified after 6 months of age (n=78)
- All children had bilateral hearing loss ranging from mild to profound and normal hearing parents
- Early identified children had significantly better language scores – this advantage found across all communication modes, degrees of hearing loss, and socioeconomic strata

Effects of Age of Identification on Language Development

Language Quotients at Three Years of Age by Age of Identification Category

Ages of Identification

Language Quotient Score

Average range

0-6 mos  7-12 mos  13-18 mos  19-24 mos  25-34 mos
3 year old newly diagnosed with mild to moderate hearing loss
3 year old with moderate to severe hearing loss who has been in hearing aids and receiving intervention since 4 months of age.
Brief History of Newborn Hearing Screening

- 1994 - Joint Committee on Infant Hearing (JCIH) endorsed goal of universal newborn hearing screening and encouraged further research into screening methods and intervention.
- 1999 - AAP released statement in support of it.
- 2000 - Congress authorized HRSA, CDC, and other agencies to create EHDI (Early Hearing Detection and Intervention) system.
- 2005 – Every state has newborn hearing screening program and ~95% of babies being screened prior to discharge.
- 2007 – JCIH’s latest guidelines
1-3-6 Guidelines

- **Infants should be screened by no later than 1 month of age.**
  - If outpatient rescreening is done, should be completed by 1 month of age.
    - If infant failed ABR screening initially, should be rescreened with ABR and not OAE.
    - If failed in 1 ear, both should be retested
      - ~20% who originally fail screen in only 1 ear have some degree of bilateral hearing loss

- **Infants who do not pass screening, should be referred to audiology by no later than 3 months.**

- **Infants with confirmed hearing loss should receive appropriate intervention by no later than 6 months of age.**
  - Medical evaluation should include genetics consultation, ENT evaluation, ophthalmology evaluation
OAE vs. ABR

- Both methods acceptable for newborn hearing screening in well babies.
- ABR recommended for NICU babies to screen for neural hearing loss.
Auditory Neuropathy/Auditory Dyssynchrony

- Condition that affects the neural processing of auditory stimuli
- Patients usually have normal cochlear function (and will have normal OAE testing)
- Affects 1-3/10,000 children
- Risk factors include:
  - Neonatal history of anoxia, hyperbilirubinemia, mechanical ventilation, or hypoxia
  - Congenital brain abnormalities
  - Low birth weight
  - Prematurity (<28 weeks)
  - Family history of AN/AD
- 1/3 of affected patients have no identifiable risk factor
What about babies who pass initial screen, but have risk factors?

- Should be referred for audiology evaluation by 24 to 30 months; sooner for certain risk factors or if there are concerns
- Risk factors include:
  - Family history*
  - In utero infection (CMV*, herpes, rubella, syphilis, toxo)
  - NICU stay >5 days
  - Postnatal infection (Bacterial meningitis*)
  - Craniofacial abnormality
  - Neurodegenerative d/o*
  - Head trauma
  - Chemotherapy*
  - Findings suggestive of syndrome associated with hearing loss*
  - Baby with h/o ECMO*, mechanically-assisted ventilation, ototoxic meds, loop diuretics, exchange transfusion for jaundice
JCIH Recommendations for Surveillance in the Medical Home

- Do regular surveillance of developmental milestones, auditory skills, parental concerns, and middle ear status at well child exams
- Use objective standardized screening of global development with a validated assessment tool at 9, 18, and 24 to 30 months of age and any time a parent is concerned
- Refer infants who do not pass the speech-language portion of screening or when there is any concern regarding hearing or language for speech-language evaluation and audiology assessment
So…How’s Newborn Hearing Screening Going?

• In 2010, 97.9% of newborns screened

Source: 2010 CDC EDHI Hearing Screening & Follow-up Survey
Documented Intervention Status of Infants with Hearing Loss (U.S., 2010) Total w. Hearing Loss = 4,923

- LFU/LTD, 24.9%
- Died/Declined, 4.9%
- Not Eligible Part C, 1.5%
- Monitoring Only (No EI), 0.4%
- Non-resident / Moved, 1.3%
- Receiving EI, 66.9%

Source: 2010 CDC EDHI Hearing Screening & Follow-up Survey
Alabama’s Story

- Really do great job at universal newborn hearing screening
  - Began tracking screening in 2001; became mandated part of newborn screen in 2008
  - 99.78% of infants screened; 88.9% by 1 month
- ~50% are diagnosed by 3 months
- ~50% are enrolled in Early Intervention by 6 months
PCPs and Newborn Hearing Screening

- Moeller, White and Shisler surveyed PCPs in 21 states and Puerto Rico (n=1,968)

- Demographics:
  - Practice location – metropolitan (57%), small town (22%), rural community (12.1%)
  - Practice setting – community clinic (75.6%), hospital (10.4%), medical school/university (5.8%)
  - Years of experience – 0-10 (40.2%), 11-20 (28.6%), 21-30 (22.5%), 31+ (8.7%)

PCPs and Newborn Hearing Screening

- Majority responded that screening newborns was very important (81.6%) or somewhat important (14.6%)
- Although average cost per infant is $30 or less, 35.6% of respondents estimated cost more than $100
- 77.7% believed it is worth the cost; the remaining 24.3% were unsure or unconvinced
- 12% of pediatricians and 17% of family physicians indicated they receive newborn hearing screening results on <50% of their patients
  - 4% responded their state did not have newborn hearing screening program; 10% were unsure
- 14% believed their training prepared them to meet the needs of infants with permanent hearing loss
Physicians’ Estimates of Ages at Which Various Follow-up Procedures Should be Conducted

<table>
<thead>
<tr>
<th>What Is Your Best Estimate of the Earliest Age at Which:</th>
<th>≤1 mo</th>
<th>1–3 mo</th>
<th>3–6 mo</th>
<th>6–9 mo</th>
<th>9–12 mo</th>
<th>&gt;12 mo</th>
</tr>
</thead>
<tbody>
<tr>
<td>Newborn who does not pass screen should receive additional testing</td>
<td>75.7%</td>
<td>11.8%</td>
<td>7.0%</td>
<td>4.2%</td>
<td>0.1%</td>
<td>1.2%</td>
</tr>
<tr>
<td>Infant can receive a definitive diagnosis of SNHL</td>
<td>51.9%</td>
<td>10.8%</td>
<td>12.4%</td>
<td>15.4%</td>
<td>0.3%</td>
<td>9.3%</td>
</tr>
<tr>
<td>Child can begin wearing hearing aids</td>
<td>38.1%</td>
<td>9.1%</td>
<td>11.2%</td>
<td>22.3%</td>
<td>1.2%</td>
<td>18.1%</td>
</tr>
<tr>
<td>Child with SNHL should be referred to early intervention</td>
<td>61.6%</td>
<td>8.0%</td>
<td>9.8%</td>
<td>13.2%</td>
<td>0.4%</td>
<td>7.0%</td>
</tr>
</tbody>
</table>

* Responses that are consistent with the AAP guidelines for pediatric medical home providers (www.medicalhomeinfo.org).
Specialists to Whom Doctors Refer Infants with Confirmed Sensorineural Hearing Loss

<table>
<thead>
<tr>
<th>Specialist Referral</th>
<th>% of Respondents</th>
</tr>
</thead>
<tbody>
<tr>
<td>ENT/Otolaryngologist</td>
<td>75.8%</td>
</tr>
<tr>
<td>Audiologist</td>
<td>41.3%</td>
</tr>
<tr>
<td>Geneticist</td>
<td>8.9%</td>
</tr>
<tr>
<td>Ophthalmologist</td>
<td>0.9%</td>
</tr>
</tbody>
</table>

So…as a group, we can do better!
Early Hearing Detection and Intervention (EHDI) Guidelines for Pediatric Medical Home Providers

Newborn Screening - Birth
- Identify a Medical Home for every infant
- Hospital-based or Inpatient Screening (only ABHR on NICU)* at 5 days
- All results sent to Medical Home

Screening Completed Before 1 Month
- Pediatric Audiologic Evaluation with Capacity to Perform:
  - OAE
  - AABR
  - Frequency-specific tone burs
  - Air & bone conduction
  - Cochlear implant
  - Audiolist reports on state EHDI
  - Medical & Otologic Evaluations
  - Referral to IDEA Part C

Diagnostic Evaluation Before 3 Months
- Pediatric Audiologyst Reports to State EHDI Program
  - Every child with a permanent hearing loss, as well as all normal follow-up results

Intervention Services Before 6 Months
- Continued enrollment in IDEA Part C 3rd of 4th at 3 years of age
- Referrals to Medical Home for specialty evaluations, to determine eligibility and identify related conditions:
  - Otolaryngologist
  - Ophthalmologist
  - Geneticist (recommended)
  - Developmental pediatrician, neuropsychologist, geneticist (as needed)

Pediatric Audiology
- Behavioral hearing audiometry
- Ongoing monitoring

Ongoing Care of All Infants, Coordinated by the Medical Home Provider
- 
  - Provide care with knowledge about hearing, speech, and language development
  - Identify and aggressively treat middle ear disease
  - Prevent vision screening and referral when indicated in the AAP “Bright Futures Guidelines, 3rd Ed.”
  - Provide ongoing developmental screening (and referral when indicated) per the AAP “Bright Futures Guidelines, 3rd Ed.”
  - Refer promptly for audiological evaluation when there is any parental concern, regarding speech, or language development
  - Refer for audiological evaluation (at least once before age 30 months) infants who have any risk indicators for later-onset hearing loss:
    - Family history of permanent childhood hearing loss
    - Neonatal intensive care unit stay of more than 5 days duration, or any of the following (regardless of length of stay):
      - ECMO, extracorporeal membrane oxygenation
      - Subtleties, subthoracic abnormalities, heart disease, Down syndrome, or other diseases
      - Congenital anomalies, particularly those that involve the ear canal, eustachian tube, and temporal bone anomalies
      - Syndromes associated with progressive or delayed-onset hearing loss (neurofibromatosis, meningomyelomeningocele, Usher Syndrome)
      - Neurodegenerative disorders (such as Hunter Syndrome or sensory motor neuropathies (such as Friedreich’s ataxia and Charcot-Marie-Tooth disease))
      - Head trauma, especially basal skull/temporal bone fracture that requires hospitalization
      - Chondroplasia

*OAE = Otoacoustic Emissions, AABR = Automated Auditory Brainstem Response, EHDI = Early Hearing Detection and Intervention, NICU = Neonatal Intensive Care Unit, IDEA = Individuals with Disabilities Education Act, HCBS = Home and Community-Based Services, AAP = American Academy of Pediatrics

Notes:
- All newborn screening programs that do not meet the American Academy of Pediatrics Environment (AAP) recommendations for early newborn hearing screening will be designated as “not compliant” with EHDI guidelines.
- All children who fail newborn hearing screening should be referred for medical diagnosis, evaluation, and intervention.

February 2010 - American Academy of Pediatrics Task Force for Improving Newborn Hearing Screening, Diagnosis and Intervention (www.medicalhomeinfo.org)
Role of the Medical Home

- Check newborn hearing screening results at first visit
- Know whether your local nursery is using OAE or ABR equipment
- If infant failed initial screen...either refer to audiology or arrange for outpatient rescreening by 1 month of age
- If infant diagnosed with hearing loss...make sure enrolled in EI and remember medical evaluation includes ENT, genetics, and ophthalmology referrals
- Refer children with risk factors who pass initial screen to audiology by 24 to 30 months (earlier for certain risk factors or if concerns arise)
Early Hearing Detection and Intervention (EHDI)
Patient Checklist for Pediatric Medical Home Providers

<table>
<thead>
<tr>
<th>Day(s)</th>
<th>Hospital-based Inpatient Screening Results (OAE/AABR) (also Home Birth)</th>
<th>Ongoing Care of All Infants</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>DKU: <strong>/</strong>/__</td>
<td>Provide parents with information about hearing, speech, and language milestones</td>
</tr>
<tr>
<td></td>
<td>Left ear: Missed</td>
<td>Identity and aggressively treat middle ear disease</td>
</tr>
<tr>
<td></td>
<td>Right ear: Missed</td>
<td>Vision screening and referral as needed</td>
</tr>
<tr>
<td></td>
<td>Incomplete</td>
<td>Ongoing developmental surveillance/referal</td>
</tr>
<tr>
<td></td>
<td>Failed Screen: 1</td>
<td>Risk indicators for delayed-onset hearing loss:</td>
</tr>
<tr>
<td></td>
<td>Failed Screen: 2</td>
<td>(If risk factors are present, refer for audiology evaluation at least once prior to age 30 months)</td>
</tr>
<tr>
<td></td>
<td>Pass</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Outpatient Screening Results (OAE/AABR)</td>
<td></td>
</tr>
<tr>
<td></td>
<td><strong>/</strong>/__</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Left ear: Incomplete</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Right ear: Incomplete</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Failed Re-Screen: 1</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Failed Re-Screen: 2</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Pass</td>
<td></td>
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</tr>
</tbody>
</table>

- **Pediatric Audiology Evaluation**
  - Hearing Loss
  - Normal Hearing
  - Document child and family auricular history
  - Report to State EHDI Program results of diagnostic evaluation
  - Referral to Early Intervention (IDEA, Part C)
  - Advise family about communication options and adaptive listening devices
  - Medical & Otologic Evaluation to recommend treatment and provide clearance for hearing aid fitting and monitoring
  - Pediatric Audiology for hearing aid fitting and monitoring

- **Enrollment in Early Intervention (IDEA, Part C)**
  - Referral to Part B 3 years of age
  - Medical Evaluations to determine etiology and identify related conditions
  - Otolaryngology (required)
  - Ophtalmologist (recommended)
  - Geneticist (recommended)
  - Developmental pediatrician, neurology, cardiology, and nephrology (as needed)

- **Ongoing Pediatric Audiology Services**

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**Patient Name:** __________________________

**Date of Birth:** __/__/___

**Ongoing Care of All Infants**

- Provide parents with information about hearing, speech, and language milestones.
- Identity and aggressively treat middle ear disease.
- Vision screening and referral as needed.
- Ongoing developmental surveillance/referal.
- Risk indicators for delayed-onset hearing loss.

* (If risk factors are present, refer for audiology evaluation at least once prior to age 30 months)

**Service Provider Contact Information**

- **Pediatric Audiologist:**

**Early Intervention Services Coordinators:**

- **Other:**

**Other:**

**Other:**

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*This project is funded by an educational grant from the Maternal and Child Health Bureau, Health Resources and Services Administration, US Department of Health and Human Services.*

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*American Academy of Pediatrics*

**Enrolled in their program for all children.**
If you repeat a failed hearing screening in your office…

- Remember the rescreen needs to be done with ABR if infant initially failed an ABR screen
- Rescreen both ears even if only 1 side didn’t pass initial screen
- Screen must be OAE or ABR (not subjective evaluation)
- Repeat the screen once and by 1 month of age…and then refer
- Report the results of the rescreening to the state EHDI program
Ella’s Story
Behavioral Observation Audiometry (BOA) conducted in sound field and minimal responses noted as follows. Responses consisted of □ localizations □ other (6c):

<table>
<thead>
<tr>
<th>Stimulus</th>
<th>Option</th>
<th>Localization</th>
</tr>
</thead>
<tbody>
<tr>
<td>Maraca</td>
<td>( + )</td>
<td></td>
</tr>
<tr>
<td>Crime Rattle</td>
<td>( + )</td>
<td></td>
</tr>
<tr>
<td>Bean Rattle</td>
<td>( + )</td>
<td></td>
</tr>
<tr>
<td>Jingle Bells</td>
<td>( - )</td>
<td></td>
</tr>
<tr>
<td>Clacker</td>
<td>( + )</td>
<td></td>
</tr>
<tr>
<td>Squeek Toy</td>
<td>( + )</td>
<td></td>
</tr>
</tbody>
</table>

\(0\) 10 20 30 40 50 60 70 80 90 100 110 120

Washington, DC 20004

Test Reliability: 50 Good 50 Fair 50 Poor

Audimeter: 50000 5000 5000 5000 5000 5000 5000

Procedure: 50 BOA 50 COR 50 VRA 50 Play 50 Conventional

Clinician(s): 50 TH 50
Physician Resources

http://www.medicalhomeinfo.org/how/clinical_care/hearing_screening/

Hearing loss module at
http://www.pedialink.org
http://www.cdc.gov/ncbddd/ehdi/

www.infanthearing.org

www.babyhearing.org

http://www.adph.org/newbornscreening/
Any Questions?
Thanks to…

- AAP EHDI program
- NCHAM
- Melissa Richardson
- Jill Smith and Children’s Hospital Audiology Department
- Ella and the Hornsby Family
References


References

- Ross, DR and Visser SN. “Pediatric Primary Care Physicians’ Practices Regarding Newborn Hearing Screening.” *Journal of Primary Care & Community Health*, published online March 12, 2012.