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

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

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

Effects of Unilateral Hearing Loss

- Children with 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

Reading Comprehension Scores of Hearing and Deaf Students

- Deaf
- Hearing

Vocabulary Development in Infants and Toddlers


Effects of Unilateral Hearing Loss

- Keller & Bundy (1993) (n = 20; age = 7.5 yrs)
- Peterson (1993) (n = 30; age = 7.5 yrs)
- Bess & Thorpe (1984) (n = 48; age = 10 yrs)
- Brail, Peterson & Viethong (1985) (n = 50; age = 10 yrs)
- Colburnson & 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.

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.


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.


Boys Town National Research Hospital Study of Earlier vs. Later

129 deaf and hard-of-hearing children assessed bi-annually.

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

Identified <6 mos (n = 25)
Identified >6 mos (n = 104)


Effects of Age of Identification on Language Development

- Language Quotients at Three Years of Age by Age of Identification Category.
- Average range.
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 rescreened
  - ~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* - NICU stay >5 days
  - In utero infection - postnatal infection
  - 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

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%)

So... How’s Newborn Hearing Screening Going?

- In 2010, 97.9% of newborns screened


Source: 2010 CDC EDHI Hearing Screening & Follow-up Survey
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>Age at Which</th>
<th>% of Respondents</th>
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<tbody>
<tr>
<td>1 mo</td>
<td>0.0</td>
</tr>
<tr>
<td>1-3 mo</td>
<td>7.0</td>
</tr>
<tr>
<td>3-6 mo</td>
<td>14.0</td>
</tr>
<tr>
<td>6-9 mo</td>
<td>22.3</td>
</tr>
<tr>
<td>9-12 mo</td>
<td>3.2</td>
</tr>
<tr>
<td>&gt;12 mo</td>
<td>10.8</td>
</tr>
</tbody>
</table>

Specialists to Whom Doctors Refer Infants with Confirmed Sensorineural Hearing Loss

<table>
<thead>
<tr>
<th>Specialist Referral</th>
<th>% of Respondents</th>
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<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!

EHDI and the Medical Home

- Parent Groups
- Mental Health
- Birthing Hospital
- Audiology
- Primary Provider
- ENT
- 3rd Party Payers
- Deaf Community
- Services for Hearing Loss
- Early Intervention Programs
- Genetics

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).
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
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 Screenings.” *Journal of Primary Care & Community Health*, published online March 12, 2012.