Early Hearing Detection and Intervention (EHDI): A Primer for Residents

A PRESENTATION FROM THE AMERICAN ACADEMY OF PEDIATRICS
Early identification and intervention of a child who is Deaf or Hard of Hearing (D/HH) will support the development of good communication, language, and social skills.

Delayed Early Intervention can be associated with speech and language delays and inability to reach each child’s full potential.
Comparison of Select Congenital Conditions

Incidence per 10,000 of Congenital Conditions

Newborns

<table>
<thead>
<tr>
<th>Condition</th>
<th>Incidence</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hearing loss</td>
<td>30</td>
</tr>
<tr>
<td>Cleft lip or palate</td>
<td>17</td>
</tr>
<tr>
<td>Down Syndrome</td>
<td>14</td>
</tr>
<tr>
<td>Limb defects</td>
<td>5</td>
</tr>
<tr>
<td>Sickle Cell Anemia</td>
<td>5</td>
</tr>
<tr>
<td>Spina bifida</td>
<td>4</td>
</tr>
<tr>
<td>PKU</td>
<td>1</td>
</tr>
</tbody>
</table>
Early Hearing Detection & Intervention (EHDI) Program

1-3-6

National EHDI Goals

- All infants will receive a hearing screening before 1 month of age
- Infants not passing the screening will receive appropriate audiologic and medical evaluation before 3 months of age
- All infants identified as D/HH will begin receiving early intervention services before 6 months of age
Spoken Language Development in Late vs Early Identification of Child who is Deaf or Hard of Hearing (D/HH)

- Late diagnosis of hearing status
- Early diagnosis of hearing status
Consider inserting a slide with information about the data and laws in your state.
Early Hearing Detection and Intervention (EHDI) Guidelines for Pediatric Medical Home Providers

The Role of the Medical Home

If there is any suspicion that an infant is D/HH

- Do listen to parents concerns and refer immediately for full audiology evaluation for any parental worries about hearing or language development
- Encourage prompt follow-up with rescreens and diagnostic evaluations
- Make sure diagnostic evaluations are done by an audiologist who has experience with infants
- Set up electronic medical record (EMR) system to include results of auditory screening
- Flag all patient charts for children that require follow-up for hearing screens
- Flag all patient charts for children that are at risk for late onset hearing loss
The Role of the Medical Home

Infants identified as D/HH

- Address the family’s concerns
- Ensure the family is seeing an experienced pediatric audiologist
- Refer the family to appropriate specialists
- Otolaryngology, Genetics, Ophthalmology
- Help the family obtain early intervention services
- Monitor developmental milestones and ear infections
**Otolaryngology**
- Assess integrity of ear canal and middle ear
- Order appropriate diagnostic screening such as temporal bone CT, MRI, etc.
- Discuss possible surgical interventions
- Counsel family and follow for success of intervention

**Genetics**
- Evaluate for possible genetic causes of hearing change
- Counsel family and patient

**Ophthalmology**
- Assess integrity of visual system
- Evaluate for visual changes known to be associated with hearing changes
Case Study 1: Baby James

- Baby James comes to your office for a well child visit at 2 weeks of age
- Full term, 3300 g, uncomplicated pregnancy and delivery
- Parents are concerned because he did not pass his newborn hearing screen in the hospital and they wonder what to do now
- How do you counsel them?
OAE: Otoacoustic Emissions


AABR: Automated Auditory Brainstem Response


[Diagram] Retrieved from https://www.medicalhomeportal.org/image/65
## Newborn Hearing Screening

### Otoacoustic Emissions (OAE) vs Automated Auditory Brainstem Response (AABR)

<table>
<thead>
<tr>
<th></th>
<th>Otoacoustic Emissions (OAE)</th>
<th>Automated Auditory Brainstem Response (AABR)</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Technique</strong></td>
<td>Probe with microphone placed in the ear canal. Acoustic stimuli presented.</td>
<td>Earphone placed in the ear canal, electrodes placed on baby’s scalp. Acoustic stimuli presented.</td>
</tr>
<tr>
<td><strong>Measurement</strong></td>
<td>OAEs are measured in the ear canal. With outer/middle ear and/or cochlear problems, no OAEs are detected.</td>
<td>Neural activity of cochlea, auditory nerve and brainstem is measured. Problems with peripheral auditory and/or auditory nerve and/or brainstem result in abnormal or absent measurements of auditory neural activity.</td>
</tr>
<tr>
<td><strong>Advantage</strong></td>
<td>Is easier and quicker.</td>
<td>Can indicate auditory nerve or auditory brainstem pathway dysfunction.</td>
</tr>
<tr>
<td><strong>Disadvantage</strong></td>
<td>Will NOT identify auditory nerve or auditory brainstem dysfunction.</td>
<td>May require sedation after 4 months of age.</td>
</tr>
</tbody>
</table>
Audiogram

Normal: 0-15 dB
Mild: 15-40 dB
Moderate: 40-55 dB
Moderate to Severe: 55-70 dB
Severe: 70-90 dB
Profound: 90-110 dB

Case Study 1: James at 2 months

- James returns for his 2 month well child visit
- Parents report that after the 2 week visit, he returned to the birth hospital for a repeat OAE, and because he did not pass this, had an ABR with an audiologist
- ABR revealed that he had bilateral moderate to severe sensorineural hearing deficit. The audiologist recommended return visit for hearing aid fitting
- Parents are doubtful of the results because James is cooing and startles to loud noises
- What other referrals need to be placed at this time?
- What needs to happen by 6 months?
Case Study 2: James’ Brother

- The same mother now presents with the child’s two and a half year old brother, Nathan, at his well child check up
Case Study 2: James’ Brother

- She is concerned that he is not responding to her consistently, and doesn’t appear to have as much spoken language as the other children in his daycare.
- She confides to you that she thinks she is just being paranoid since the recent identification of his brother’s hearing status.
Case Study 2: James’ Brother

What is the appropriate next step?

A. Use developmental assessment tool (eg, ASQ, PEDs) to evaluate language development – if normal, reassure mother and re-evaluate child in 6 months

B. Stand behind child and whisper “what is your name?” to see if they answer correctly

C. Use pure tone hearing screening in office

D. Refer directly to audiology
What is the appropriate next step?

A. Use developmental assessment tool (eg, ASQ, PEDs) to evaluate language development – if normal, reassure mother and re-evaluate child in 6 months

B. Stand behind child and whisper “what is your name?” to see if they answer correctly

C. Use pure tone hearing screening in office

D. Refer directly to audiology
# Early Hearing Detection and Intervention (EHDI) Guidelines for Pediatric Medical Home Providers

## Newborn Screening
- Identify a Medical Home for every infant
- Hospital-based Inpatient Screening (0-12 days; October ABR or APR if NICU <6 days, then ABR sent to Medical Home)

## Screening Completed Before 1 Month
- Outpatient Re-Screening (0-12 months): All results sent to Medical Home if State EHDI Program
- Ongoing Care of All Infants: Coordinated by the Medical Home Provider
  - Provide parents with information about hearing, speech, and language milestones
  - Identify and aggressively treat middle ear disease
  - Provide vision screening (and referral when indicated) as recommended 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 hearing, 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

## Diagnostic Evaluation Before 3 Months
- Pediatric Audiologic Evaluation with Capacity to Perform:
  - OAE (outer auditory membrane emission)
  - AABR (automated auditory brainstem response)
  - Assessment, evaluation, and intervention for those with hearing loss

## Audiologist Reports to State EHDI Program
- Every child with a permanent hearing loss, as well as all normal follow-up results
- Referral to IDEA* Part C
- Team Advises Family About:
  - All communication options, different communication modes, assistive listening devices (hearing aids, cochlear implants, etc.), parent support programs

## Medical & Otologic Evaluations
- To recommend treatment and provide clearance for hearing aid fitting

## Intervention Services Before 6 Months
- Continued enrollment in IDEA* Part C (Transition to Part B at 3 years of age)
- Referrals by Medical Home for developmental evaluations, to determine eligibility and identify related conditions:
  - Otolaryngologist
  - Ophthalmologist
  - Geneticist (recommended)
  - Developmental pediatrician, neurologist, cardiologist, psychologist, audiologist
- Pediatric audiology
  - Ongoing monitoring

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### Notes:

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February 2010 - American Academy of Pediatrics Task Force for Improving Newborn Hearing Screening, Diagnosis and Intervention (www.medicalhomeinfo.org)
Ongoing Care of All Infants\textsuperscript{d}; Coordinated by the Medical Home Provider

- Refer promptly for audiology evaluation when there is any parental concern\textsuperscript{f} regarding hearing, speech, or language development
- 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‡, mechanically-assisted ventilation, ototoxic medications or loop diuretics, exchange transfusion for hyperbiliruinenia
- In utero infections such as cytomegalovirus‡, herpes, rubella, syphilis, and toxoplasmosis
- Postnatal infections associated with hearing loss‡, including bacterial and viral meningitis
- Craniofacial anomalies, particularly those that involve the pinna, ear canal, ear tags, ear pits, and temporal bone anomalies
- Findings suggestive of a syndrome associated with hearing loss (Waardenburg, Alport, Jervell and Lange-Nielsen, Pendred)
- Syndromes associated with progressive or delayed-onset hearing loss‡ (neurofibromatosis, osteopetrosis, 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
- Chemotherapy‡

‡Denotes risk indicators of greater concern. Earlier and/or more frequent referral should be considered.
Take Home Points

- When an infant does not pass the initial hearing screen, the repeat screen should occur before 1 month of age.
- Diagnostic ABR should be completed by 3 months of age.
- Appropriate hearing testing is required to identify hearing status.
- Early intervention referral should be completed by 6 months of age, along with subspecialty referrals.
Parent concern is a very sensitive indicator of hearing changes, and warrants an immediate referral for audiology evaluation.

It is important to develop practice parameters for tracking high risk registry infants/children for late onset hearing changes.

For example, include “at risk for late-onset hearing loss” in the running problem list (ICD-10 code Z91.89, “Other specified personal risk factors, not elsewhere specified”)

Take Home Points

- Any child with risk factors for late-onset hearing changes should be evaluated by audiology once before age 30 months.
- Even if language development is on track and there are no concerns, any child with risk factors should still be evaluated.
Useful Web sites

- American Academy of Pediatrics (AAP) EHDI page

- Joint Committee on Infant Hearing (JCIH)
  http://www.jcih.org/

- Boys Town National Research Hospital
  http://www.boystownhospital.org/
Acknowledgements

This presentation was developed as part of cooperative agreements between the American Academy of Pediatrics and the Maternal and Child Health Bureau of the Health Resources and Services Administration (HRSA) and the Centers for Disease Control and Prevention.