Newborn Hearing Screening and Screening Beyond the Newborn Period

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Disclosures

HRSA/EHDI Grant compensation as Arizona EHDI program chapter champion and Newborn Screening Partner member

Objectives

1) Interpret and manage Newborn Hearing Screening results
2) Explain how to incorporate EHDI goals into the medical home
3) Describe the importance and procedures for ruling out permanent hearing loss with otitis media
4) Identify and monitor risk factors for late onset and progressive hearing loss
WHAT IS EHDI?

Early Hearing Detection & Intervention (EHDI)

Programs are located in each state, including U.S. territories

EHDI teams consist of a chapter champion, EHDI coordinator and many other components to help with hearing data and tracking, follow-up and family support

What is EHDI?

National EHDI Goals
EHDI Goals

1. All infants will be SCREENED for hearing loss at birth or before 1 month of age.
2. Infants not passing the screening will receive a DIAGNOSTIC AUDIOLOGIC evaluation before 3 months of age.
3. All infants identified with hearing loss will receive EARLY INTERVENTION services before 6 months of age.

National EHDI Goals

- Hearing loss is #1 most common birth defect in the USA yet only 1% of mothers ranked hearing loss as their top concern.
- 33 babies born each day with hearing loss... 12,000 babies born each year.
- 1 to 3 of every 1000 newborns have hearing loss.
- 1 in 50 NICU babies have hearing loss.

Critical Statistics
According to the American Academy of Pediatrics, hearing loss is one of the most frequently occurring birth defects; if hearing loss is not detected and treated early, it can impede speech, language and cognitive development. Over time, such a delay can lead to significant educational costs and learning difficulties.

National Center for Hearing Assessment and Management (NCHAM) reports that detecting and treating hearing loss at birth for one child saves $400,000 in special education costs by the time that child graduates from high school.

An infant who does not pass his/her newborn hearing screening has a potential developmental emergency! Without the active assistance of the medical home, the infant may be considered “lost” in the early hearing detection and intervention (EHDI) system, which undermines the potential benefits of newborn hearing screening. A “wait and see” approach is never appropriate.
3.5y boy presents for possible hearing problems because the daycare provider expressed concerns to the parents after his first day at daycare.

Parents have no concerns. He is their only child.

Examination is unremarkable except for poor eye contact and poor expressive language development.

History is unremarkable except for failed NBHS, inpatient and outpatient.

No risk factors present.

Case study cont.

No follow-up secondary to concerns of “having to be put to sleep”

In office OAE: referred bilaterally, normal tympanograms

Audiology: Bilateral mild sensorineural hearing loss

Bilateral hearing aids, ST, enrolled into ASDB.

Incidence of Children with Hearing Loss

<table>
<thead>
<tr>
<th>Site</th>
<th>Rate</th>
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<tbody>
<tr>
<td>Well Baby Nursery</td>
<td>1 per 1000</td>
</tr>
<tr>
<td>NICU</td>
<td>20 per 1000</td>
</tr>
<tr>
<td>Auditory Neuropathy/Dysynch.</td>
<td>1-2/10,000</td>
</tr>
<tr>
<td>Total population</td>
<td>2-3 per 1000</td>
</tr>
<tr>
<td>Expected # infants identified annually in US (Gravel et al, 2003)</td>
<td>8,000-16,000</td>
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Actual # infants identified (CDC Survey, 2009) 5,064

%age Loss to follow-up/Documentation (2009 CDC Survey) 44.6%
Loss to Follow-up
How can you help?

History of Universal Newborn Hearing Screening Program

- Increase in State Compliance over the last 20 years. More than 90% of the 4 million babies born in the United States each year receive a hearing screening.
- Problem now is follow-up. Infants are diagnosed by the UNHS; however, the JCIH follow-up goals are not consistently being met here in Arizona and throughout the country.
- Your role as Medical Home is critical.

Arizona Newborn Hearing Screening Program

- Arizona hospitals are doing a phenomenal job screening 99% of the 87,000 babies born each year.
- However, we are struggling with follow-up for those infants that do not get a screen or have not passed a screen.
- An inability to contact families was the number one reason for not getting the babies back for follow-up. This is where you as a provider play a critical role in making a referral to a diagnostic facility for follow-up testing.
Infants suspected of a hearing loss:

- Really listen to parents concerns
- 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 document hearing results
- Flag patient chart that requires follow-up
- Flag charts that are at risk for late-onset hearing loss

Types of Screening Devices:

- A-ABR-Automated Brainstem Auditory Evoked Response
  - Measurements are obtained from surface electrodes that record neural activity generated in the cochlea, auditory nerve, and brainstem in response to acoustic stimuli delivered via an earphone. Automated ABR measurements reflect the status of the peripheral auditory system, the eighth nerve, and the brainstem auditory pathway.

- OAE-Otoacoustic Emissions
  - OAE measurements are obtained from the ear canal by using a sensitive microphone within a probe assembly that records cochlear responses to acoustic stimuli. Thus, OAEs reflect the status of the peripheral auditory system extending to the cochlear outer hair cells.

Rescreen:

- >95% of babies born in Arizona are screened either inpatient or outpatient with an Automated ABR system.
- Thus, rescreens should be completed using an Automated ABR not an OAE screening system.
- Exception, if you as the primary care physician feel that the family will not be seen again (i.e. live in a rural area, or are non-compliant) then any follow-up test is better than no follow-up test.
Meeting the Universal Newborn Hearing Screening Goals

- Hearing Screen completed by
- Diagnostic Audiology by 3 months if the baby did not pass the screening test.
- Early Intervention by 6 months if baby is diagnosed with hearing loss.

Arizona Statistics

Total Number of Infants born vs. Total Number of Infants Screened

Diagnosed by 3 months

Diagnosed

Targeted Group
Diagnosed but out of compliance with Recommended Guidelines

Babies that still need assessment at 12 months

Diagnosed but out of compliance with Recommended Guidelines

Enrolled in Early Intervention by 6 months

Infants Enrolled in EI Before 6 Months

Your Responsibility

"The primary care physician must assume responsibility to ensure that audiological assessment is conducted on infants who do not pass screening and must initiate referrals for medical specialty evaluations necessary to determine the etiology of the hearing loss."

Joint Committee on Infant Hearing, 2007

http://www.jcih.org/
When is it appropriate to refer for Diagnostic Audiology

- At 1st well-baby visit—Check hearing screening
- Refer Well Babies that have not passed inpatient/outpatient screen for diagnostic audiology
- Refer NICU babies with a >5 day stay or with risk factors that have not passed
- Primary Referral should be to a pediatric audiologist prior to a referral ENT.

Case study
- Newborn seen in the office for the first visit.
- Parents forgot to bring in blue vaccine card, but claims his hearing screen was normal
- Parents forget to bring in blue vaccine card on multiple visits.
- Mother calls (in a panic) to report hearing screen results found on the blue vaccine card at 5 months of age.

Diagnostic Audiology
Referrals

Where should I refer my babies and young patients?

Early Hearing Detection & Intervention – Pediatric Audiology Links to Services (EHDI-PALS)

EHDI-PALS is a web-based link to information, resources, and services for children with hearing loss.

At the heart of EHDI-PALS is a national web-based directory of facilities that offer pediatric audiology services to young children who are younger than five years of age.

Visit http://www.ehdipals.org/ to start using this online tool.

Diagnostic Audiology Evaluation

A Diagnostic Evaluation should be a battery of tests.

- Toneburst (Frequency specific responses)
- Bone Conduction (if HL is present)
- Clicks
- Otoacoustic Emissions
- Tympanometry
- with 1000Hz probe tone
Test Preparation

- BAER testing typically takes 1 ½ hours to 2 hours.
- Baby needs to be asleep for the test.
- We ask that the parents bring the baby tired and hungry but not asleep.
- Once they have arrived we prep the babies for testing. They then eat and hopefully sleep.
- Following the preparation guidelines are critical in facilitating a test environment most conducive of obtaining comprehensive test results.

Meeting the Universal Newborn Hearing Screening Goals

- Hearing Screen completed by 1 month.
- Diagnostic Audiology by 3 months if the baby did not pass the screening test.
- Early Intervention by 6 months if baby is diagnosed with hearing loss.

Case Studies

- Baby Boy: born full term without complications
  - First screen at 12 hours, referred both ears.
  - Second screen at 47 hours, referred both ears.
  - Outpatient screen at birth facility: refer, bilaterally.
  - Non-sedated BAER study at 2 months of age.
  - Diagnosed with bilateral moderate sensorineural hearing loss.
  - He was referred for Early Intervention.
  - Three weeks later, he was fit with binaural amplification.
  - Infant meets the 1/3/6 guidelines.
Baby Boy: 35 weeks gestation with a brief stay in a Level II nursery for 4 days.
Initial screen: Referred bilaterally. Did not receive a follow-up inpatient screen.
Outpatient screen: Referred, bilaterally. He was referred to an ENT for follow-up.
The ENT completed a follow-up screen (OAE) which he did not pass. Serous fluid was visualized, bilaterally. Follow-up was recommended in 3 months.
Baby Boy returned in 3 months and had another screening and did not pass, but again fluid was visualized. It was recommended that he return in 3 months.
At nine months of age, the baby returns and still has fluid and is still not passing his hearing screen. He is sent for a diagnostic BAER which he now has to be sedated for because he is over 6 months. He is diagnosed with a severe/profound sensorineural hearing loss.
He returns to ENT at which time they decide to place PE tubes and follow-up with another BAER. Results from the BAER indicate a severe sensorineural hearing loss.
He is fit with hearing aids 1 month later. Unfortunately, he is almost 1 year. The hearing screening goals 1/3/6 have not been met and he has lost 1 year of speech/language development.

Case Study

Otitis Media is not a barrier to an early diagnosis.
Our current equipment and training allows us to looking beyond the middle ear status and gives us the capability to make a diagnosis even when fluid is present.

Ruling Out Hearing Loss with Otitis Media

An otology referral should be made if:
- Sensorineural hearing loss is diagnosed.
- Mixed hearing loss is diagnosed.
- Conductive hearing loss associated with syndromes, craniofacial abnormalities, or disorders associated with hearing loss.

Otolaryngology Referral
Newborn Screening → Diagnosis → Intervention

- Babies identified with a hearing loss should be referred to ENT/Otology and Early Intervention (AzEIP)
- Coordinate referrals to specialty providers based on etiology of hearing loss (Otology, Genetics, Ophthalmologist)
- Ongoing Monitoring—results, treatment, education and family support

System Coordination

Late Onset and Progressive Hearing Loss

- Caregiver concern* regarding hearing, speech, language
- Family history* of permanent childhood HL
- NICU Care of >5 days, or any of the following regardless of length of stay: assisted ventilation, ototoxic meds (gentamycin, tobramycin, loop diuretics), hyperbilirubinemia with exchange transfusion and ECMO*
- Craniofacial anomalies, especially those involving the pinna, ear canal, ear tags, ear pits, and temporal bone anomalies. Physical findings associated with a syndrome known to include permanent HL

Late Onset and Progressive HL

Continued...

- Syndromes associated with progressive HL such as NF, osteopetrosis, Usher’s syndrome
- Neurodegenerative disorders*, such as Hunter syndrome
- Postnatal infections associated with SNHL especially bacterial meningitis*
- Head trauma requiring hospitalization
- Intra-uterine TORCH infections, particularly CMV*
- Chemotherapy*
Case study

- 2yr WCC visit.
- Normal development including receptive and expressive language.
- Parental concerns that child recently is sitting closer to the TV and likes it loud.
- PMH significant for multiple admissions and IV antibiotics for VUR through 1st year of life.
- Bilateral OAE in the office: referred
- Audiology: moderate sensorineural HL, bilateral

Early Hearing Detection and Intervention (EHDI) Guidelines for Pediatric Medical Home Providers

- At 1st well-baby visit—Check hearing screening results on back of immunization card or hospital discharge summary for referred or missed/incomplete screens
- Refer well babies that have not passed inpatient/outpatient screen for diagnostic audiology
- Refer NICU babies that have not passed any screen (inpatient or outpatient)
- Brainstem Auditory Evoked Response (BAER)/Auditory Brainstem Response (ABR) test by an audiologist specializing in pediatrics (prior to 3mos of age to avoid sedation.)

Recommendations for Pediatricians
Important Resources and Websites

- Arizona EHDI program  www.AZNewborn.com
- The EAR Foundation of Arizona  www.earfoundationaz.com
- National Center on Hearing Assessment and Management (NCHAM)  www.infanthearing.org
- Guide By Your Side/Hands & Voices  www.gbys.azhv.org
- EHDI-PALS  www.ehdipals.org

Thank You

- For further questions or inquiries, please contact
- Brad Golner M.D.  bgolnermd@gmail.com
Interpreting the Results

- The report should clearly state the diagnosis and recommendations.
  - Right Ear: Mild sensorineural hearing loss
  - Left Ear: Mild/moderate sensorineural hearing loss

- Results are most consistent with a communicatively significant permanent sensorineural hearing loss. This patient is a candidate for binaural amplification. Return for a hearing aid consultation in 1-2 weeks. An appointment has been scheduled. Consider a referral to otolaryngology for an otologic workup.

Interpreting Results

- The report should clearly state the diagnosis and recommendations.
  - Right Ear: Severe mixed hearing loss.
  - Left Ear: Moderate/severe mixed hearing loss.

- Results are most consistent with abnormal middle ear function and a moderate/severe mixed hearing loss. Consider a referral to otolaryngology and return for confirmation BAER testing. An appointment has been scheduled. Once hearing loss is confirmed, a hearing aid consultation will be scheduled.

Types of Screening Devices

- **ABR-Automated Brainstem Auditory Evoked Response**
  - Measurements are obtained from surface electrodes that record neural activity generated in the cochlea, auditory nerve, and brainstem in response to acoustic stimuli delivered via an earphone. Automated ABR measurements reflect the status of the peripheral auditory system, the eighth nerve, and the brainstem auditory pathway.

- **OAE-Otoacoustic Emissions**
  - Measurements are obtained from the ear canal by using a sensitive microphone within a probe assembly that stimulates cochlear responses to acoustic stimuli. Thus, OAEs reflect the status of the peripheral auditory system extending to the cochlear outer hair cells.
OAE Practices and Follow-up Recommendations

- Well babies that pass the hospital screen with normal development:
  - Conduct OAE annually
  - Report to ADHS up to 2 years
- Well Babies and babies in the NICU for < 5 days with a risk indicator that pass the hospital screen:
  - OAE monitoring more often; approximately every 6 months
  - However, if OAE is normal but concerns about abnormal development, refer to audiologist
  - Audiological evaluation between 24-30 months
  - Report to ADHS up to 2 years

Required reporting of all screens ages birth to three

MANDATORY REPORTING:

Per A.R.S. 36-694 (c)
- Hearing test results must be reported to the Arizona Department of Health Services

Per A.A.C. R9-13-207(E)
- All screening and diagnostic hearing evaluations shall be submitted within one week following the hearing test (note: even if the results are normal)

For reporting forms
www.aznewborn.com

Recommendations for Pediatricians

- At 1st well-baby visit—Check hearing screening results on back of immunization card or hospital discharge summary for referred or missed/incomplete screens
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Case Study

- Newborn with history of NICU stay >5 days
- Did not pass inpatient hearing screen
- Refer for Diagnostic Audiology

NICU                           Missed/Referred >5 days and/or risk factors
 missed/referred                Diagnostic Audiology

Case Study

- Baby Girl: Born at 38 3/7 weeks gestation without complications.
  - First screen at 16 hours, passed the right but referred on the left ear.
  - Second screen the next day, referred both right and left ear.
  - Outpatient screen 2 weeks later. (As per parent report)
  - Left ear screened 3 times (passed once)
  - Right ear (referred twice)
  - Given that the baby had passed the right ear one time and the left ear another time, she was not initially referred for a diagnostic evaluation.
  - The parents were concerned, and thus a referral for a diagnostic BAER was made following the baby’s 4 month checkup.
  - Results from the BAER found a severe rising to mild sensorineural hearing loss, bilaterally.
  - Infant does not meet the 1/3/6 guidelines.

Case Study

- Newborn seen in office for the first time.
- Referred inpatient bilaterally.
- Referred outpatient right ear.
- Refer for Diagnostic Audiology

Well Baby               Outpatient Screen               Diagnostic Audiology
 missed/referred         (Refer)                      (Refer)
Interpreting Results

- Report should clearly state diagnosis and recommendations.
  - Right Ear: Moderate conductive hearing loss.
  - Left Ear: Mild conductive hearing loss.

- Results are most consistent with abnormal middle ear function and a mild/moderate conductive hearing loss. Consider referral to otolaryngology. Return for repeat BAER to further assess auditory status.

Resources

- [http://www.ehdipals.org/](http://www.ehdipals.org/)
  - Audiology Facilities
  - Parent Resources
  - Professional Resources
  - Other Helpful Websites
JCIH 2007 Position Statement with an update in 2013

www.jcih.org


Page 906

Check back of immunization records.

When screening programs do not provide Outpatient Screening, infants will be referred directly to Pediatric Audiology for an Evaluation.

NICU babies that “fail” should be referred immediately to Pediatric Audiology for an Evaluation.

Infants with high risk factors (CMV, Syndromes assoc. with H.L., neurodegenerative disorders, trauma, ECMO, chemotherapy, caregiver concern, or family history of hearing loss) should have a diagnostic evaluation.

Even infants who fail screening in only one ear should be referred for further testing of both ears.

Important Information

- Check back of immunization records.
- When screening programs do not provide Outpatient Screening, infants will be referred directly to Pediatric Audiology for an Evaluation.
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Recommendations for Pediatricians

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Referral Path

- Well Baby
  Missed/Referred
  Outpatient Screen (if refer) Diagnostic Audiology (BAER)

- NICU
  Missed/Referred
  Diagnostic Audiology (BAER)