Hearing Loss

What the Health Care Professional Should Know

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HACKENSACK MERIDIAN HEALTH
NJ AAP CHAPTER CHAMPION EHDI PROGRAM

Knowledge, Attitudes, and Practices of physicians in New Jersey

Regarding Newborn Hearing Screening

2012 FINDINGS
What is your level of trust in the newborn hearing screening results?

- Minimal
- Fair Amount
- Complete

2012

28.6% 4.1% 67.3%

What is your best estimate of the earliest age at which:

<table>
<thead>
<tr>
<th>&lt;1 mo</th>
<th>1-3 mos</th>
<th>4-6 mos</th>
<th>7-9 mos</th>
<th>10-12 mo</th>
<th>&gt;12 mo</th>
</tr>
</thead>
<tbody>
<tr>
<td>34.4%</td>
<td>60.4%</td>
<td>4.2%</td>
<td>0.0%</td>
<td>1.0%</td>
<td>0.0%</td>
</tr>
<tr>
<td>20.9%</td>
<td>34.0%</td>
<td>22.0%</td>
<td>4.4%</td>
<td>8.8%</td>
<td>9.9%</td>
</tr>
<tr>
<td>32.6%</td>
<td>28.3%</td>
<td>25.9%</td>
<td>7.8%</td>
<td>16.9%</td>
<td>19.1%</td>
</tr>
</tbody>
</table>

- a. A newborn not passing the hearing screening should receive additional testing
- b. A child can be definitively diagnosed as having a permanent hearing loss
- c. A child can begin wearing hearing aids
- d. A child with permanent hearing loss should be referred to early intervention services

2012 Comparison
30.3%
How confident are you talking to parents about.....

<table>
<thead>
<tr>
<th></th>
<th>Very Confident</th>
<th>Somewhat Confident</th>
<th>Not Confident</th>
</tr>
</thead>
<tbody>
<tr>
<td>Causes of HL</td>
<td>29.6%</td>
<td>59.2%</td>
<td>11.2%</td>
</tr>
<tr>
<td>Sign language, A/O modes</td>
<td>15.6%</td>
<td>38.5%</td>
<td>45.8%</td>
</tr>
<tr>
<td>Unilateral, Mild HL Consequences</td>
<td>32.7%</td>
<td>55.1%</td>
<td>12.2%</td>
</tr>
<tr>
<td>Bilateral Moderate- Profound HL Consequences</td>
<td>22.3%</td>
<td>52.1%</td>
<td>25.5%</td>
</tr>
<tr>
<td>Candidates for cochlear implants</td>
<td>7.2%</td>
<td>41.2%</td>
<td>51.5%</td>
</tr>
<tr>
<td>What to do after diagnosis</td>
<td>33.0%</td>
<td>59.8%</td>
<td>7.2%</td>
</tr>
</tbody>
</table>

National Goals for Hearing Screening (1-3-6 Plan)

All infants will have access to a hearing screen – using a physiologic measure:
- No later than 1 month of age

All infants not passing initial screening and subsequent rescreening should have confirmatory audio-logical and medical evaluations:
- No later than 3 months of age

All infants with a confirmed permanent hearing loss should receive early intervention as soon as possible:
- No later than 6 months of age
Hearing Screening in New Jersey

Regulations

Hearing screening is required by law
All newborns must be screened using an electrophysiological device
All screening results must be reported to the DHSS via the EBC
Hospital must notify the primary care physician of the test results
Inpatient and outpatient results can also be viewed via the New Jersey Immunization Registry (http://njiis.nj.gov)
Premature infants should be screened with an ABR
Hospital must have a mechanism in place for re-evaluation of any abnormal initial evaluation
Rescreening must be done by same methodology as the original evaluation
All infants should have a standardized screening of globalized development with a validated assessment tool at 9, 18 and 24-30 months
Any infant who fails the speech-language portion should be referred for a speech-language evaluation and an audiology assessment

New Jersey Statewide Rates

<table>
<thead>
<tr>
<th>Year</th>
<th>Screened by discharge or by 1 month of age</th>
<th>Year</th>
<th>Refer Rate (final inpatient screen)</th>
</tr>
</thead>
<tbody>
<tr>
<td>2014</td>
<td>99.9%</td>
<td>2014</td>
<td>2.3%</td>
</tr>
<tr>
<td>2015</td>
<td>99.8%</td>
<td>2015</td>
<td>2.3%</td>
</tr>
<tr>
<td>2016</td>
<td>99.8%</td>
<td>2016</td>
<td>2.2%</td>
</tr>
</tbody>
</table>
Includes both children receiving either outpatient rescreening or diagnostic testing. 2016 rates still incomplete.

Documented Repeat Evaluation

Outcome of 2015 Births with Inpatient Refer

data as of 4/13/17
Why is Early Identification of Hearing Loss Important?

Hearing loss is the most common birth condition.

Congenital Defects/Diseases

Incidence per 10,000

- Hearing Loss: 30
- Cleft lip or palate: 12
- Down Syndrome: 11
- Limb defects: 6
- Spina bifida: 5
- Sickle Cell Anemia: 2
- PKU: 1
Prevalence of Hearing Loss

- Prevalence estimates vary across studies
- Estimated that 1 to 3 per 1000 infants will have permanent sensorineural hearing loss*
  - 1/1000 from the well baby nursery
  - 10/1000 from the NICU
- Rate increases to 6/1000 by school age
  - Need for continued surveillance

* NIDCD Outcomes Research in Children with Hearing Loss (2005) available at:

Which of the following can have an impact on speech and language development? (Check all that apply):

- 67.3%
- 80.2%
- 97.0%
- 98.0%
- 0%
- 20%
- 40%
- 60%
- 80%
- 100%

Unilateral HL
Mild Bilateral HL
Moderate Bilateral HL
Severe/Profound Bilateral HL
Why is Early Identification of Hearing Loss Important?

Early identification and intervention can make a difference

Language Development
Effects of Age of Identification

Prospective, longitudinal study of early-identified infants

30 children with mild-profound hearing loss (HL) compared to 96 normal hearing (NH) controls

Children identified < 3 months had stronger language development at 12-16 months than those identified > 3 months

Children with HL were delayed compared to NH infants

Vohr et al. Peds 2008; 122:535-544
Language Development
Effect of Age of Identification


Vocabulary at Age Five
Effect of Age of Intervention

Moeller MP. Pediatrics 2006;118, 1357-1370
Vocabulary

ITS ALL ABOUT THE WORDS!

Early Intervention is about the Family:
   Adult Education

Number of Words heard by the age of 2

- Professional Home - 50 million words
- Working Class Home - 30 million words
- Welfare Home - 11 million words
Hearing Loss
Classification

Genetic (35%)
Non Syndromic (60%)
  - Autosomal recessive (60-70%)
  - Autosomal dominant (30%)
  - X-linked (2%)
  - Other: mitochondrial, multilocus
Syndromic (40%)

Acquired (35%)
  - Prenatal (20%)
  - Perinatal (20%)
  - Later (60%)

Idiopathic (30%)

Inherited Deafness
Non Syndromic
Autosomal Recessive

- Usually have normal hearing parents
- No history of environmental exposure
- Always sensorineural in origin
- Autosomal recessive is more severe than dominant
- Multiple chromosomal locations associated with hearing loss
- Most important gene is connexin 26 (chromosome 13)
  - May be responsible for ½ AR hearing loss
  - Possible to screen for these genes
Inherited Deafness

Non Syndromic

Autosomal Dominant

Accounts for approximately 30% of non syndromic deafness

Six subdivisions
- Congenital severe-profound
- Progressive early onset
- Unilateral
- Low frequency
- Mid frequency
- High frequency progressive

Usually less severe than autosomal recessive

Hearing loss is usually sensorineural

Inherited Deafness

Syndromic

(most common syndromes)

Craniosynostosis (1-2%)
- Crouzon syndrome (50% with conductive loss)
- Apert syndrome
Branchio-Oto-Renal Syndrome (2%)
- preauricular sinus or pits
- autosomal dominant
Branchial Arch Defects (1%)
- Oculo-auriculo-vertebral spectrum (1/5600)
- Treacher Collins Syndrome (hearing loss in 40%)
Alport Syndrome (1%)
- ocular abnl (anterior lenticonus, macular flecks, corneal erosions)
- renal abnl (glomerular sclerosis)
Pendred Syndrome (1-10%)
- associated with a goiter
- defect in sulfate transport protein common in cochlea and thyroid
Usher Syndrome (3-5%) – associated with Retinitis Pigmentosa
Waardenburg Syndrome (2-5%)
Acquired Hearing Loss

Neonatal Intensive Care > 5 days which may include
• Mechanical Ventilation/ECMO
• Hyperbilirubinemia requiring an exchange transfusion
• Exposure to ototoxic medications or loop diuretics

Postnatal infections assoc. w/hearing loss (meningitis)
• Bacterial
• Viral

Intrauterine TORCH infections
• Cytomegalovirus
• Zika
• Herpes
• Rubella
• Syphilis
• Toxoplasmosis

Craniofacial Anomalies involving:
• Pinna
• Ear canal
• Ear tags
• Ear pits
• Temporal bone anomalies

Zika Infections

HEARING LOSS
Birth Defects Among Fetuses and Infants of US Women with Evidence of Possible Zika Virus Infections During Pregnancy

442 Completed Pregnancies (15-50 yrs.) – with lab evidence of Zika Infection
- 271 were asymptomatic (61%)
- 167 were symptomatic (38%)
- 26 had evidence of Birth Defects (6%)

Presence of Birth Defects
- 16/271 asymptomatic pregnant women (6%)
- 10/167 symptomatic pregnant women (6%)
- All defects occurred in women exposed in the first trimester

Types of Defects
- 18/26 had microcephaly (4% of completed pregnancies)
- 4/26 had brain abnormalities w/o microcephaly (i.e. cranial calcifications, absence of the corpus callosum etc.)

Hone et al. JAMA 2017:317;59-68

Hearing Loss in Infants with Microcephaly

Study conducted in Brazil

70 infants with documented Zika Infection and microcephaly
- 5 (7%) had Sensorineural hearing loss
- Similar to the rate seen with other congenital viral infections
- 16 (22.8%) failed the first screening in at least one ear

Leal et al. MMWR Weekly Sept. 2, 2016/65;917-919
Infant positive for Zika Virus Infection

Was Screening by ABR?

Pass?

Yes

No

Symptomatic

Pass?

Yes

No

Monitoring per JCIH guidelines

Yes

No

ABR between 4-6mo

Pass?

Yes

No

ABR between 6-9mo or Behavioral testing At 9mo

Pass?

Yes

No

Hearing Assessment by Audiologist

Yes

No

HEARING LOSS

CMV (Cytomegalovirus)

CDC Guidelines – Zika Infection and Hearing Loss
U.S. Department of Health and Human Services
### Cytomegalovirus

#### Knowledge

<table>
<thead>
<tr>
<th>Condition</th>
<th>2015</th>
<th>2016</th>
<th>Freq in US</th>
</tr>
</thead>
<tbody>
<tr>
<td>Congenital CMV</td>
<td>6.79%</td>
<td>6.70%</td>
<td>6000</td>
</tr>
<tr>
<td>Congenital Toxoplasmosis</td>
<td>8.27%</td>
<td>8.33%</td>
<td>400</td>
</tr>
<tr>
<td>Group B Strep</td>
<td>17.87%</td>
<td>16.91%</td>
<td>380</td>
</tr>
<tr>
<td>Congenital Rubella</td>
<td>16.8%</td>
<td>13.27%</td>
<td>&lt;3</td>
</tr>
<tr>
<td>Parvovirus(fifth dis)</td>
<td>22.52%</td>
<td>19.63%</td>
<td>1045</td>
</tr>
<tr>
<td>Fetal Alcohol Synd</td>
<td>65.56%</td>
<td>64.54%</td>
<td>1200</td>
</tr>
<tr>
<td>Spina Bifida</td>
<td>69.42%</td>
<td>64.54%</td>
<td>1500</td>
</tr>
<tr>
<td>SIDS</td>
<td>83.96%</td>
<td>78.7%</td>
<td>1500</td>
</tr>
<tr>
<td>Down Syndrome</td>
<td>89.57%</td>
<td>85.44%</td>
<td>6000</td>
</tr>
<tr>
<td>HIV/AIDS</td>
<td>91.13%</td>
<td>86.33%</td>
<td>30</td>
</tr>
</tbody>
</table>

HealthStyles survey: questionnaire to over 55000 households with 4000 returned

#### Seroconversion (seronegative to seropositive) in pregnancy

- Low income pregnant women have 3 times the incidence (6.8%)
- Middle income pregnant women – 2.5%
- Women in STD clinics – 37%
- Daycare workers – 7.9-10%

#### CMV Prevention

- CMV can live for up to 15 min on hard plastic
- CMV can live for up to 5 min on crackers
- No virus can be recovered with any intervention
  - Soap
  - Sanitizer
  - Plain water
Cytomegalovirus

Education helps
- 42% of CMV negative mothers with a CMV positive infant converted without education
- Only 3% of CMV seronegative mothers with a CMV positive infant converted after education about hygiene practices
- Only 34% of first year medical students ever heard about CMV (100% of 4th yr knew)
- Only 18.5% of childcare providers heard of CMV

Hearing Loss
- 10% of asymptomatic Congenital CMV infants will develop HL (only 50% detected at birth)
- 0.1-0.4% of general population will develop HL

Guidelines for CMV Detection

Laboratory Confirmation
- Gold Standard has been CMV detection in urine
  - Difficult to obtain urine
  - Poor for universal detection
- Saliva (both liquid and dried)
  - Good sensitivity (99.9%)
  - Good specificity (97%)
- Blood Samples
  - Poor sensitivity (34-100%)
  - Estimated that 2/3 of infected infants would be missed with this method
  - Positive results correlated well with other tests

Guidelines for CMV

Treatment

Treatment (Valganciclovir)
6 weeks vs. 6 months (end point was change in hearing in best ear)

- 6 week group
  - 2 had improvement
  - 36 had no change
  - 5 had worsening
  - Had lower Bayley Scores at 6 months of age
  - Had lower Receptive-communication scale scores

- 6 month group
  - 3 had improvement
  - 37 had no change
  - 3 had worsening

Kimberlin et al. NEJM 2015;372:933-943

Unilateral Hearing Loss
Effects of Unilateral Hearing Loss

Average Results
Math = 30th percentile
Language = 25th percentile

Unilateral Hearing Loss
Prevalence

• In newborns – 0.83-2.7/1000
• In school aged children – 30-56/1000

• In the United States it is estimated that 36% of infants with hearing loss had UHL
  ◦ 10% of UHL will become bilateral
• UHL children cannot localize
Unilateral Hearing Loss

Etiology

Causes can include:

- Heredity
  - 59% had a family history of any hearing loss
  - 26% had a family history of UHL
  - 50% of subjects that had imaging had structural abnormalities (more common than in bilateral (5%)
    - Vestibular aqueducts
    - Mondini dyplasia (dysplasia of the temporal bone)
    - Common cavity deformity
  - 67% of children with UHL and cochlear nerve deficiency had ophthalmologic abnormalities
- Head trauma
- Viral causes: i.e. mumps, rubella, CMV
- Meningitis
- Fetal Alcohol Syndrome

Unilateral Hearing Loss

2/3 of children with UHL reported increased difficulty in challenging environments

- 1/3 had difficulty hearing their teacher
- Children with UHL required a minimum of 2 dB signal-noise ratio to perform as well as their hearing peers

Persist to adulthood

- 95% of UHL adults reported that understanding speech in noise was challenging
- 6.7 of normal hearing reported difficulty in a noisy environment

Academic achievement

- 22-40% of children with UHL repeated a grade compared to 2-4% of hearing children
- 41-45% of children with UHL will require Individual Education Program (IEP) (vs.12%)
- 59% of children with any degree of UHL demonstrated academic and behavioral problems
Unilateral Hearing Loss

Social and Behavioral Development

- 20-33% of UHL have teacher reports of behavioral problems
  - Social withdrawal
  - Inattentiveness
  - Distractibility
  - Lack of co-operation
  - Aggression
- 1/3 were embarrassed by their hearing loss and felt inferior to their peers

Hearing Aids

- Only 26% were recommended for hearing aids
- Does provide some benefit:
  - Improved hearing
  - Better school and social performance
  - Greater ease of hearing in quiet and noisy environments
- Only 26% of children with UHL use their hearing aids

Contralateral routing of signals (CROS) hearing aids

- Works by placing a microphone on the affected ear and delivering the signal to a receiver in the normal ear
- Only work if the user situates themselves with the affected ear away from the noise
- Works best in a quiet environment
- Not an appropriate choice for children
Unilateral Hearing Loss

Intervention

Frequency Modulated (FM) system
- Wireless transmission of the signal of interest via radio waves to the listener
- Teacher wears a microphone and the student has the signal coupled to the hearing aid
- Multiple studies have shown improvement in speech recognition in both noisy and quiet environments

Cochlear Implants (CI)
- Direct electrical stimulation to the auditory nerve (bypasses the cochlea)
- Not FDA approved for UHL (use is increasing – mostly in adults)
- Early indications are that there is no improvement in social interactions and poor compliance

Age of Referral
New Jersey

Year of Birth

- >1 yr
- 6mo-1yr
- 3-6 mo
- <3 mo
Hearing Screening Techniques

- Otoacoustic emissions (OAE)
- Auditory brainstem response (ABR)
- Two stage screening (OAE + ABR)
Otoacoustic Emissions

- Sounds are presented to the ear canal and a small microphone measures the response in the ear canal
- Average test time is 5-15 minutes/baby

Auditory Brainstem Response

- Sounds are presented and surface electrodes measure brainstem activity
- Average test time 20 min/baby
Two Stage Screening
OAE + ABR

- All babies are screened using OAEs
- Those babies who fail the OAE screening receive an ABR screening prior to leaving the hospital
- Average test time/baby (25-35 min)
- Reduces refer rate; useful when follow up is likely to be difficult or costly
- Initial cost of equipment is higher than OAE or ABR screening alone, but follow-up costs are less

JCIH Position on Screening
2007

NICU
- >5 days in NICU
- ABR should be included to screen for neural loss
- Rescreen BOTH ears, even if only one ear fails
- Non pass – refer to Audiologist
- Readmission – re-screen before discharge

Well baby nursery
- Screen with OAE or ABR
- Repeat screen when necessary before discharge
- When using 2 step protocol test order should be OAE then ABR
- Re-screen BOTH ears, even if only one ear fails
Medical Home

ROLE OF THE HEALTH CARE PROVIDER

Optimal Surveillance in the Medical Home (JCIH, 2007)

At each well visit monitor for:

- Auditory skills, middle ear status
- Developmental milestones (validated global screening tool)
- Listen to parental concerns

If concerns, refer for pediatric audiology and speech-language pathology evaluations
Optimal Surveillance in the Medical Home (JCIH, 2007)

If hearing loss is diagnosed, refer siblings of infant for audio-logical evaluation

Refer infants with any RISK indicators for audio-logic assessment by 24-30 months of age

Carefully assess middle ear status at all well child visits; refer for otologic evaluation if persistent middle ear effusion lasts for 3 months+

List specialists to whom you refer a child with confirmed permanent hearing loss (open ended):

<table>
<thead>
<tr>
<th>Specialist</th>
<th>2012</th>
</tr>
</thead>
<tbody>
<tr>
<td>ENT/Otolaryngology*</td>
<td>86.1%</td>
</tr>
<tr>
<td>Geneticist*</td>
<td>10.9%</td>
</tr>
<tr>
<td>Ophthalmologist*</td>
<td>2.0%</td>
</tr>
<tr>
<td>Audiologist</td>
<td>50.5%</td>
</tr>
<tr>
<td>SLP</td>
<td>24.8%</td>
</tr>
<tr>
<td>Occupational Therapist</td>
<td>0.0%</td>
</tr>
<tr>
<td>Child Dev./ EI</td>
<td>9.9%</td>
</tr>
<tr>
<td>Neurologist</td>
<td>5.0%</td>
</tr>
<tr>
<td>Social Worker</td>
<td>0.0%</td>
</tr>
</tbody>
</table>

* = Correct Answer
Follow-up Testing

Referral for follow-up testing

- A repeat OAE and/or ABR screen must be done and if possible completed by 1 month of age
- Any pediatrician conducting re-screening in their office is required to report results to DHSS

If a hearing loss is still suspected…

- Referral to a pediatric audiologist/ENT for diagnostic evaluation to be complete by 3 months of age
- Frequency specific ABR to estimate degree and configuration of hearing loss

Medical Workup

Review the prenatal and perinatal history

Review family history of hearing loss

Look for physical findings associated with HL

- White forelock
- Cardiac anomalies
- External ear anomalies

Refer to ENT Physician

Refer to Genetics, Cardiology, Ophthalmology, Nephrology

Other: CMV, EKG, Developmental evaluation

Amplification

Hearing aids can be fitted as young as 1 month of age

Special Child Health Services County Case Management Units have a hearing aid assistance program for families needing financial aid to pay for hearing aids:

www.nj.gov/heath/fhs/sch/sccase.shtml

Cochlear Implants

• Parents must be involved in the decision making process
• Six month trial of hearing aids is required for Cochlear Implants
• For additional information:
  www.fda.gov/cdrh/cochlear

American Academy of Pediatrics
Early Intervention Referral

**ANY** hearing loss (including mild or unilateral) is presumptive eligibility for EI services

Goal is to begin EI services by 6 months of age

County Case Management Units are single point of entry for EI services in NJ


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Do you ever do hearing screening for infants and young children in your office?

*Yes* – 31.3%

*No* – 68.8%
### If yes, who are you screening? *(n=30)*

<table>
<thead>
<tr>
<th>Screening Category</th>
<th>Percentage</th>
</tr>
</thead>
<tbody>
<tr>
<td>Newborns/infants who <strong>do not pass the newborn hearing screening</strong> in hospital</td>
<td>36.7%</td>
</tr>
<tr>
<td>Newborns/infants for whom you <strong>cannot obtain newborn screening results</strong></td>
<td>26.7%</td>
</tr>
<tr>
<td><strong>All newborns</strong> regardless of previous tests</td>
<td>20.0%</td>
</tr>
<tr>
<td>Children 1-3 years of age as part of their <strong>annual check-ups</strong></td>
<td>53.3%</td>
</tr>
<tr>
<td>Children of <strong>parents who voice concerns</strong> about their child's hearing</td>
<td>80.0%</td>
</tr>
</tbody>
</table>

### How often do you use the following to screen hearing in your office? *(n=30)*

<table>
<thead>
<tr>
<th>Screening Method</th>
<th>Never</th>
<th>Occasionally</th>
<th>Frequently</th>
<th>Always</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>AABR</strong></td>
<td>60.9%</td>
<td>4.3%</td>
<td>21.7%</td>
<td>13.0%</td>
</tr>
<tr>
<td>Response to noisemakers</td>
<td>16.0%</td>
<td>36.0%</td>
<td>28.0%</td>
<td>20.0%</td>
</tr>
<tr>
<td>Caregiver interview/questionnaire</td>
<td>21.7%</td>
<td>21.7%</td>
<td>21.7%</td>
<td>34.8%</td>
</tr>
<tr>
<td><strong>OAE</strong></td>
<td>14.8%</td>
<td>7.4%</td>
<td>25.9%</td>
<td>51.9%</td>
</tr>
<tr>
<td><strong>Tuning Fork</strong></td>
<td>57.9%</td>
<td>31.6%</td>
<td>10.5%</td>
<td>0.0%</td>
</tr>
<tr>
<td><strong>Tympanometry</strong></td>
<td>15.0%</td>
<td>40.0%</td>
<td>30.0%</td>
<td>15.0%</td>
</tr>
<tr>
<td><strong>Other</strong></td>
<td>0.0%</td>
<td>0.0%</td>
<td>66.7%</td>
<td>33.1%</td>
</tr>
</tbody>
</table>
# Resources

## Early Intervention
- Contact State EHDI Coordinator
  - www.infanthearing.org
  - www.nectac.org

## Parent-to-Parent
- www.handsandvoices.org
- www.babyhearing.org

## Physician support
- www.aap.org
- www.medicalhomeinfo.org

---

**www.babyhearing.org**

![babyhearing.org](image)
www.infanthearing.org

www.raisingdeafkids.org

FROM ALD TO ZIKA: NEWBORN SCREENING AND SURVEILLANCE IN NJ

APRIL 25, 2107
NJ-EHDI Project Contacts

phone:  609-292-5676  
TTY:    609-984-1343  
fax:    609-633-7830  
email:  EHDIdoh.state.nj.us  
website: www.nj.gov/health/fhs/ehdi

Other helpful websites:  
AAP EHDI information:  
www.medicalhomeinfo.org/screening/hearing.html  
CDC EHDI program:  
www.cdc.gov/ncbddd/ehdi