Update on Newborn Hearing Screening
National Goals for Hearing Screening (1-3-6)\textsuperscript{1, 2}

- All infants will access hearing screening using a physiologic measure
  - no later than 1 month of age

- All infants not passing initial screening and subsequent rescreening should have confirmatory audiological and medical evaluations
  - no later than 3 months of age

- All infants with confirmed permanent hearing loss should receive early intervention as soon as possible
  - no later than 6 months of age
Prerequisites for a Population Screening Program

- Condition sufficiently frequent in screened population
  - YES
- Condition serious or fatal without intervention
  - YES
- Condition must be treatable or preventable
  - YES
- Effective follow-up program possible
  - YES
Why is early identification of hearing loss important?

- Hearing loss is the most common birth condition.
Incidence of Congenital Conditions (Per 10,000)

- Hearing loss: 30
- Cleft lip or palate: ~10
- Down syndrome: ~5
- Limb defects: ~2
- Spina bifida: ~1
- Sickle cell anemia: ~1
- 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\(^3, 4\)
  - 1/1000 from the well baby nursery
  - 10/1000 from the NICU
- Rate increases to 6/1000 by school age\(^4\)
  - Need for surveillance
What does it sound like to have a hearing loss?

Severe hearing loss

Moderate hearing loss

Mild hearing loss

Normal hearing
Why is early identification of hearing loss important?

- Previous methods for detecting hearing loss have been ineffective
  - High risk screening failed to identify ~50% of the infants with hearing loss
  - Large retrospective cohort study\textsuperscript{5, 6}: mean age of diagnosis 21.6 months
  - Similar findings reported in US\textsuperscript{7,8,9}
Newborn hearing screening is effective

- Large, good-quality cohort study conducted in UK\textsuperscript{10}
- 53,781 babies; 25,609 born during NHS era
- 2-step screening (OAE + ABR)
  - Sensitivity = 0.92
  - Specificity = 0.98
- Lower refer rates with qualified examiners\textsuperscript{11}
Why is early identification of hearing loss important?

- Hearing loss is the most common birth condition
- Previous methods for detecting hearing loss have been ineffective
- Undetected hearing loss can delay speech, language, social & academic development
Vocabulary Development in Infants\textsuperscript{12, 13}

Delays in babble also observed\textsuperscript{14, 15}
Reading Comprehension in Children with Mild-Mod Loss
Why is early identification of hearing loss important?

- Early identification and intervention can make a difference.
Effects of Age of Identification on Language Development

- 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
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
Vocabulary at Age Five by Age of Intervention

Significant Predictors:

- Id Age: 8%
- Family Involvement: 37%
Evidence that Early Matters

- 8-year follow up to Wessex (UK) trial\textsuperscript{10}
  - 120 children with permanent HL (from population-based cohort of 157,000 infants)

- Speech-language outcomes at school age (Mean = 7.9 years)

- Children with HL confirmed \(\leq 9\) mos had better receptive and expressive language scores than later identified children
  - Speech scores were equivalent in the 2 groups
American Academy of Pediatrics (AAP)

- Endorsed implementation of universal newborn hearing screening in 1999
- Defined standards for:
  - Screening
  - Tracking & Follow-up
  - Identification & Intervention
  - Program Evaluation
- Encouraged AAP Chapters to provide leadership in physician education and newborn screening in their states
Early Hearing Detection and Intervention (EHDI)

- Endorsed by:
  - AAP, National Institutes of Health, Maternal and Child Health, Centers for Disease Control, Joint Committee on Infant Hearing & in 2008, the USPSTF

- As of 2005, all 50 states implemented statewide EHDI programs

- As of 2006, an average of 95.7% of newborns were screened nationally
Status of Hearing Screening in Nebraska (as of 10/08)

- 99.5% of newborns are being screened
- 68/69 hospitals are screening
- Refer rate is 2.3%
- 54 infants with permanent HL were diagnosed in 2007

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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
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
2007 JCIH Position on Screening

- **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 – rescreen 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
  - Rescreen BOTH ears, even if only one ear fails
Cost effectiveness

- Within each hospital, the optimum approach will depend upon the number of births/year, the availability of trained personnel for testing 365 days/year, follow-up services in the area, and expected loss to follow-up rate.
# Screening Costs

## ABR tests

<table>
<thead>
<tr>
<th>Births/yr.</th>
<th>Cost/Baby (Test only)</th>
<th>Cost/Baby (w/ F/U)</th>
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<tbody>
<tr>
<td>100</td>
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<tr>
<td>8000</td>
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## OAE tests

<table>
<thead>
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<th>Cost/Baby (Test only)</th>
<th>Cost/Baby (w/ F/U)</th>
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<tr>
<td>8000</td>
<td>$ 6.16</td>
<td>$22.16</td>
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</table>

## OAE + ABR

<table>
<thead>
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<th>Births/yr.</th>
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</tr>
</thead>
<tbody>
<tr>
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<tr>
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<td>$15.90</td>
</tr>
<tr>
<td>8000</td>
<td>$ 7.79</td>
<td>$11.79</td>
</tr>
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</table>
Characteristics of a good screening program

- Refer rate of 1.5-5.0% in well baby nursery and slightly lower in the NICU (resulting from 2-stage screening in the hospital)
  - 5.0% = 400 babies per 8000 births
- Ongoing training and monitoring program for personnel
- Structured plan for follow up
- Ability to track program performance (important for quality assurance and for JCAHO requirements)
What if a baby fails UNHS?

- Failure rates range from 1.5-5.0% in good screening programs

- Most babies who fail the initial screening will actually have normal hearing
  - For 10 babies that refer, 1 is expected to have permanent hearing loss
System challenges: Loss to Follow Up

- 8 New York hospitals,
  - 28% infants who did not pass in-hospital screening failed to return
  - Loss to follow up is as high as 50% in some states
- Return rates better for in-hospital fails than in-hospital misses
Medical Home: Strategies to Promote Follow Up

- At prenatal visit, encourage families to identify you as follow-up care location
- Inform hospital to facilitate communication of results
- Provide checkbox on newborn well child form/patient chart for hearing screening results & risk factors
- Set up tracking system for infants who do not pass hearing screening
Counseling Parents

- Effective communication of results to families has an influence on follow up behaviors
- Balance between reassurance and importance of follow up testing
- “Your child may or may not have a hearing loss... but let’s be sure about it. If further testing shows hearing loss, the earlier we get started helping the child, the better.”
Counseling Parents Following Screening
Follow Up Testing

- Referral for follow-up testing
  - Repeat OAE and/or ABR screening
- If a hearing loss is still suspected...
  - Referral to a pediatric audiologist
    - Experienced in testing infants & children
    - Has appropriate equipment to test infants
  - Frequency specific ABR to estimate degree and configuration of hearing loss
    - Early testing can avoid need for sedation
Counseling Parents Following Diagnosis
Components of a Comprehensive Audiological Evaluation

- History
- Assessment of hearing sensitivity (ABR)
- Rule out middle ear pathology; refer to ENT physician if appropriate
- Initiate amplification
- Refer to local early intervention program
- Provide support via other parents of children with hearing loss
- PCP helps to coordinate child’s follow up care in their practice
JCIH 2007 Follow Up Guidelines

- EHDI systems should be family-centered
- Families should have:
  - Access to information on all treatment options
  - Counseling regarding hearing loss
- Child and family should have:
  - Immediate access to hearing technologies
Amplification

- Hearing aids can be fitted as young as 1 month of age
Importance of Intervention in Outcomes

Early Identification needs to be paired with early, appropriate and consistent interventions.
3 year old with moderate-severe loss: Inconsistent Intervention

Child A
3 year old with moderate-severe loss: Consistent early identification

Child B
3 year old with mild-moderate loss: Identified at 3 years, 3 months

Pre-intervention sample

Child C – 3 years

Audio C

FREQUENCY (Hz)

HEARING LEVEL in dB
5 year old with mild-moderate loss:
Identified at 3 years, 3 months
Post- intervention sample

Child C – 5 years
Roles of the Medical Home

- Understand testing results at screening and diagnostic phases & implications for follow up
- Assure follow-up screening; refer for diagnostic and medical specialty evaluations
- Support family in understanding severity & type of hearing loss
- Refer to early intervention
- Offer partnership with parents to identify and develop a plan of health and habilitative care
Optimal Surveillance in the Medical Home (JCIH, 2007)²

- At each visit consistent with AAP periodicity schedule monitor for:
  - Auditory skills, middle ear status
  - Developmental milestones (validated global screening tool)
  - Parental concerns

- If concerns, refer for pediatric audiology and speech-language pathology evaluations
Optimal Surveillance in the Medical Home (JCIH, 2007)²

- If hearing loss is diagnosed, refer siblings of infant for audiological evaluation
- Refer infants with any RISK indicators for audiological 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+
Risk Indicators for permanent congenital, delayed onset or progressive hearing loss

- Caregiver concerns* about hearing, speech, language, development
- Family history* of permanent childhood hearing loss
- NICU stay > 5 days or any of following (regardless of length of stay):
  - ECMO assisted ventilation*
  - Ototoxic medications (gentimycin, tobramycin)
  - Loop diuretics (furosemide, Lasix)
  - Hyperbilirubinemia requiring exchange transfusion
- In Utero infections (cmv*, herpes, rubella, syphillis, toxoplasmosis)

* = greater risk for delayed onset HL
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JCIH, 2007  * = greater risk for delayed onset HL
Risk Indicators for permanent congenital, delayed onset or progressive hearing loss

- In Utero infections
  - CMV*, herpes, rubella, syphilis, toxoplasmosis

- Craniofacial anomalies

- Physical findings (e.g. white forelock)

- Syndromes* involving hearing loss
  - Neurofibromatosis, osteopetrosis, Usher, Waardenburg, Alport, Pendred, Jervell & Lange-Nielson

* = greater risk for delayed onset HL
Risk Indicators for permanent congenital, delayed onset or progressive hearing loss

- Neurodegenerative disorders
  - Hunter syndrome
  - Sensory motor neuropathies (Friedrich ataxia, Charcot-Marie-Tooth)
- Culture positive postnatal infections associated with HL*
  - Herpes, varicella, meningitis
- Head trauma (basal skull, temporal bone)*
- Chemotherapy*

* = greater risk for delayed onset HL
Medical Workup

- Complete prenatal & perinatal history
- Family Hx of onset of HL < age 30
- Physical for stigmata, ear tabs, cleft palate, cardiac, skeletal, microcephaly
- Refer to ENT/CT of temporal bones
- Refer to Genetics and Ophthalmology
- Other: CMV, EKG, Developmental evaluation
CI Candidacy Criteria

- 3-6 month trial with hearing aids; lack of benefit
- Profound loss 90+dB (12 to 18 mos); >18 mos, Severe-to-Profound 70 dB+
- No medical contraindications
- Rehab setting encouraging auditory
- Family factors (motivation, expectations)
Goals of Early Intervention

- Home based services
- Optimally, providers have experience & training with the population and work to:
  - Establish partnerships with families
  - Promote family competence & confidence in parenting child
  - Support family in providing a language-rich environment in everyday routines
  - Support family to become informed decision makers for the child
  - Conduct ongoing assessments of outcomes
    - Adjust interventions as necessary to optimize outcomes
  - Promote family access to formal and informal supports
  - Provide culturally competent services
**Resources:**

- Early Intervention
- Parent-to-Parent
- Physician support

- Contact State EHDI Coordinator – see www.infanthearing.org
- www.nectac.org
- www.handsandvoices.org
- www.beginningssvsc.com
- www.babyhearing.org
- www.aap.org
- www.medicalhomeinfo.org
Physician Resources

http://www.medicalhomeinfo.org/screening/hearing.html

ALSO: hearing loss module on http://www.pedialink.org

http://www.cdc.gov/ncbddd/dd/ddhi.htm
Newborn Screening

Today, many babies will have their hearing tested before leaving the hospital. The tests are simple and painless and take only a few minutes. The following are questions you may have about testing.

What is Universal Newborn Hearing Screening (UNHS)?
Why is Universal Newborn Hearing Screening Important?
How Does Newborn Hearing Screening Testing Work?
What Does it Mean if a Baby Fails the Newborn Hearing Screening Test?
Can a Baby Pass the Test and Still Have Hearing Loss?
What Should I Do If My Baby Fails the Test?
Early Hearing Detection & Intervention Information & Resource Center

In the information & resource center one will find a wealth of information and resources concerning the many dimensions of early hearing detection and intervention. Information and resources include some of the following:

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<th>Resources</th>
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<td>Early Intervention, Legislative Activities, Data Management, Family Support</td>
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<td>EHDI Bulletin Board</td>
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<tr>
<td>Bulletin Board</td>
<td>For Families</td>
</tr>
</tbody>
</table>

[Visit www.infanthearing.org for more information.]
Chapter Champion Contact

For more information...

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  - Phone: 410-329-1995
  - Mobile: 410-746-5084
- Maryland Infant Hearing Screening Program
  http://fha.dhmh.maryland.gov/genetics/SitePages/inf_hrg.aspx
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