Sound Beginnings:
Changing the World for Children with Hearing Loss

Disclosures:
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I have no relevant financial relationships with the manufacturer/s of any commercial product/s 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.
Disclosures:
Susan Wiley, MD

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Learning Objectives

• Explain how to screen for hearing loss, describe the different degrees of hearing loss, and identify the risk factors of hearing loss

• Apply the strategies identified to screen by 1 month, identify by 3 months, and enroll into early intervention by 6 months children who are deaf or hard of hearing

• Identify follow-up strategies and procedures for children who didn't 'pass' their NBHS
Key Points

1. Most parents with a newly identified deaf child are completely surprised

2. Many of the professionals from whom parents seek help are not up-to-date

3. The most important thing to parents is to be able to COMMUNICATE with their child
Incidence per 10,000 of Congenital Defects/Diseases

• Permanent hearing loss is the most frequent birth defect in the United States.

• If hearing loss is not identified and treated early, deaf children will require an additional $400,000 per child in educational costs.

Little Known Facts About Congenital Hearing Loss

<table>
<thead>
<tr>
<th>Condition</th>
<th>Incidence Rate</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hearing Loss</td>
<td>30</td>
</tr>
<tr>
<td>Other Birth Defects</td>
<td>12</td>
</tr>
<tr>
<td>Down Syndrome</td>
<td>11</td>
</tr>
<tr>
<td>Spina Bifida</td>
<td>6</td>
</tr>
<tr>
<td>Spina Nova</td>
<td>5</td>
</tr>
<tr>
<td>Middle Ear Anomaly</td>
<td>2</td>
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<tr>
<td>Other</td>
<td>1</td>
</tr>
</tbody>
</table>

Total Communication

- Visual Language
- Sign Language
- Listening & Spoken Language

Cued Speech

- Spoken Language
CAN YOU HEAR DIFFERENT PEOPLE TALKING AT SCHOOL?
Spring is my favorite season. The sun shines bright. The flowers begin to grow. I like spring.
Earlier Identification of Hearing Loss

High Quality Early Intervention Programs that focus on teaching LANGUAGE

Availability of Better Hearing Technology

Advocacy and Public Policy Initiatives

Components of an Effective Early Hearing Detection and Intervention (EHDI) Program

- Diagnosis before 3 months
- Screening before 1 month
- Intervention before 6 months
- Medical Home
- Data Management and Tracking
- Program Evaluation and Quality Assurance
- Family Support!!

What enabled us to move from ….
In March 1993, a Consensus Development Panel convened by the National Institutes of Health concluded that:

- “The average diagnosis of hearing impairment remains constant at about 2 ½ years of age.
- All infants should be screened for hearing impairment...this will be accomplished most efficiently by screening prior to discharge from the well-baby nursery.
- Identification of hearing impairment must be seen as imperative for all infants.”
Why is it important?

- It meets the prerequisite for screening
  - Condition sufficiently frequent in screened population
  - Condition serious or fatal without intervention
  - Condition must be treatable or preventable
  - Effective follow-up program possible
- Early intervention for hearing loss improves language outcomes

Rate Per 1,000 of Permanent Childhood Hearing Loss in EHDI Programs

<table>
<thead>
<tr>
<th>Site</th>
<th>Sample Size</th>
<th>Prevalence Per 1000</th>
</tr>
</thead>
<tbody>
<tr>
<td>Texas (Finitzo et al 1998) (1/94 to 6/97)</td>
<td>54,228</td>
<td>2.15</td>
</tr>
<tr>
<td>Colorado (Mehl &amp; Thomson, 1998) (1/92 - 12/96)</td>
<td>41,976</td>
<td>2.56</td>
</tr>
<tr>
<td>New Jersey (Barsky-Firs, 1993-12/95)</td>
<td>15,749</td>
<td>3.30</td>
</tr>
<tr>
<td>Hawaii (Johnson et al 1997) 1/96 - 12/96</td>
<td>9,605</td>
<td>4.15</td>
</tr>
<tr>
<td>Massachusetts (2004) (1/04 – 12/04)</td>
<td>78,515</td>
<td>2.87</td>
</tr>
</tbody>
</table>
Age in Months at Which Permanent Hearing Loss Was Diagnosed

<table>
<thead>
<tr>
<th>Study</th>
<th>Age at Diagnosis</th>
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</thead>
<tbody>
<tr>
<td>Coplan (1987)</td>
<td>35</td>
</tr>
<tr>
<td>Elssman et al. (1987)</td>
<td>30</td>
</tr>
<tr>
<td>Gustason (1987)</td>
<td>30</td>
</tr>
<tr>
<td>Meadow-Orlans (1987)</td>
<td>30</td>
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<tr>
<td>Stein et al. (1990)</td>
<td>31</td>
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<tr>
<td>Mace et al. (1991)</td>
<td>25</td>
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<tr>
<td>Johnson et al. (1997)</td>
<td>20</td>
</tr>
<tr>
<td>Vohr et al. (1998)</td>
<td>19</td>
</tr>
<tr>
<td>Harrison and Roush (2003)</td>
<td>10</td>
</tr>
<tr>
<td>Massachusetts (2004)</td>
<td>0</td>
</tr>
</tbody>
</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)
Hearing Screen Protocols

• Screening results should be conveyed immediately to families so they understand the outcome and the importance of follow-up when indicated.

• For rescreening, a complete evaluation of both ears is recommended, even if only 1 ear failed the initial screen.

Hearing Screen Protocols

• Separate protocols are recommended for NICU and well baby nurseries.

• NICU babies >5 days are to have ABR included as part of their screen so that neural HL will not be missed.
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

Audiograms

Bilateral Profound Sensorineural Hearing Loss
3 year old with moderate-severe loss: Inconsistent Intervention
3 year old with moderate-severe loss: Consistent early intervention

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

Pre-intervention sample
5 year old with mild-moderate loss: Identified at 3 years, 3 months

Post-intervention sample

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

*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**
  - 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** (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
What Contributes to “Loss to Follow-up”? 

- **Referral rates in the hospital are too high** (because of poorly trained screeners, poorly maintained equipment, lack of commitment, etc)
- **Ineffective information for parents** (about initial results, need for follow-up, what to do next, etc)
- **Accurate data isn’t shared quickly with the right stakeholders** (hospitals, state EHDI program, medical home, audiologists, early interventionists, etc)
- **Shortage of pediatric audiologists** (because of not enough training programs, poor reimbursement rates, rural/remote residences, etc)
- **Lack of knowledge about current “effective practices”** (among program managers, health care providers, early interventionists, etc).
- **Not enough public awareness about importance of issue** (taxpayers, administrators, extended family, etc)
- **Lack of resources** (for screening, follow-up diagnosis, early intervention, case management, etc)

The Hearing Head Start Project

- Feasibility study from 2001-2004
- 69 programs in 3 states with 3,000+ children screened
- Identified 2 per 1,000 with permanent hearing loss and 20 per 1,000 with unidentified transient losses
- Currently in 21 of 50 states—expanding to others by 2015
Assume a newborn for whom you are caring is diagnosed with a moderate to profound bilateral hearing loss. If no other indications are present, to which specialists would you refer the baby?

When can an infant be fit with hearing aids?

American Academy of Pediatrics

Universal Newborn Hearing Screening, Diagnosis, and Intervention
Guidelines for Pediatric Medical Home Providers
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 (or set up system to receive birth information)
- Add checkbox on newborn well child form/patient chart/EMR for hearing screening results and risk factors
- At 1 week visit, make sure you have the screening results
- In children who did not pass their hearing screen, Confirm audiologist appointment with parents at time of PCP visit

National Initiative for Children's Healthcare Quality
Medical Home: Strategies to Promote Follow Up

- Set up tracking system for infants who do not pass hearing screening
- Set up tracking system for children with identified risk factors for HL
- Encourage communication between providers (audiology, ENT, speech pathology, Early Intervention, etc)

National Initiative for Children's Healthcare Quality

AAP: online module on Hearing Loss

www.pedialink.org

- Web-based learning:
  - medical home in screening, follow-up, management, referral, and family support of children with hearing loss
  - the rates of hearing loss and understand the importance of early recognition and the ongoing screening/surveillance process
  - the differences between screening methods available for hearing loss and the audiologic evaluation process in infants and children
  - different types of hearing loss including the medical work-up of a child with an identified hearing loss
  - understand the risk factors of hearing loss, including the differences between medical and genetic risk factors
Educational Materials:
- What to ask your.... series
- A parent’s guide to HL
- Guide to Early Intervention
- Guide to Genetics and HL

www.cdc.gov/ncbddd/ehdi/
What to ask your...

Audiologist, ENT, Medical Provider, SLP, Geneticist

Mild and Unilateral Loss

www.cdc.gov/ncbddd/ehdi/unilateralhi.htm

Here’s the research
Questions?

Communicating with your child

Once your child is a bit older, there are 5 main ways he can learn to communicate:

- Learn to use his hearing and speak.
  - This is called the auditory-verbal method.
  - This method teaches children to listen, read lips and speak.
- Learn to use the hearing he has left.
  - This is called the auditory-verbal therapy.
  - This method helps children use whatever hearing they have left.
  - This method does not encourage lipreading.
- Learn American Sign Language
  - This is called BiModality.
  - With this method, children learn American Sign Language and become part of Deaf culture.
- Learn to use handshapes that stand for sounds.
  - This is called cued speech.
  - This method gives people a way to "see" English.
  - It uses handshapes to "show" the sounds you can't see by reading lips.
- Learn to use some of these methods together.
  - This is called total communication.
  - This approach uses different ways of communicating together, like signing and talking at the same time.

How to choose a way to communicate
Question #1

What are specific, evidence-based strategies for strengthening the communication and reporting between the state newborn hearing screening program and the medical home?

Components of an Effective Early Hearing Detection and Intervention (EHDI) Program

- **Diagnosis** before 3 months
- **Screening** before 1 month
- **Intervention** before 6 months
- Medical Home
- Data Management and Tracking
- Program Evaluation and Quality Assurance
- Family Support!!
Answer #1

• State EHDI programs recognize the importance of and do their best to notify the medical home of a child’s hearing screening results.
• This works best when the primary care pediatrician is identified prior to a child’s hearing screen, or prior to the child’s discharge from the hospital.
• Primary care pediatricians can help by verifying that their practice contact information is in the birthing hospital’s/center’s record, and that they have been identified as the primary health care provider for that child.
• With various policies and programs across the country, there isn’t a “one-size fits all” system at this time.
• We have a long way to go in improving the link between the newborn hearing screening programs and the medical home, and we need YOUR help!

Question #2

What early intervention services are recommended for children with hearing loss, and when is the optimal time to begin utilizing these services?
Answer #2

• As soon as hearing loss is identified, children should be enrolled in early intervention (EI). Even very young children (infants at 2 months of age) can benefit from early intervention. This is usually based primarily on parent training in early infancy on what to expect and how to communicate with your child.

• States and counties’ EI programs vary greatly from region to region. Preferably, early intervention providers would be knowledgeable in educational strategies for children who are deaf or hard of hearing. The Ski*Hi curriculum has been used by many states to evaluate their programs:
  – Who are the providers?
  – How often are the service visits?
  – Are families feeling supported?
  – Are the children making progress?

• Additional services (such as occupational therapists, physical therapists, and/or behavioral specialists) should be provided when concerns arise. Having a screening system in place for children enrolled in Early Intervention for additional needs is appropriate.

Question #3

What are the best data tracking systems used by hospital systems and/or state programs?
Answer #3

• The Centers for Disease Control and Prevention has given grants to almost all states to assist in developing data tracking systems.
• Types of tracking systems:
  – Commercial products
  – State system by state IT departments
  – Built-in component of the medical record
• Screening, Diagnosis, and Intervention referral records need to be transmitted to the entire care team: the medical home, audiologist, and the early intervention program.
• The most progressive systems have now automated this process. The primary care provider will receive an email or letter with the records/results and the next steps that must be taken.

Question #4

How do we screen preschoolers for hearing loss in a pediatric practice?

What equipment is needed?
Answer #4

- The age (1 – 5 years old) of the preschooler is a primary factor in determining the best type of screen to provide.

- The Administration on Children and Families funded a national project to provide early intervention programs with auto-acoustic emission equipment to screen children’s hearing.
  - 2 / 1,000 children were found to have permanent hearing loss that would not have otherwise been identified
  - 20 / 1,000 children were found to have fluctuating conductive hearing loss that would not have otherwise been identified

- Some pediatricians offer in-office screening as part of the well-child visit.

- Day care and other early childhood programs often use behavioral screening (and sometimes auto-acoustic emission).

- All caretakers in a child’s life need to consider hearing screening if a child’s language development is lagging behind norms.