Newborn Hearing Screening Programs and Their Impact on Early Usher Diagnosis

Presented at
First International Symposium on Usher Syndrome and Related Disorders
Omaha, Nebraska

by
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National Center for Hearing Assessment and Management
www.infanthearing.org
Universal Newborn Hearing Screening is not a new idea…

“[There is] an urgent need to study further and more critically methods of testing hearing in young children . . . during this first year the existence of deafness needs to be ascertained . . . training needs to be begun at the earliest age that the diagnosis of deafness can be established.”

What I Hope You Will Remember From This Presentation

1. A rising tide lifts all ships
2. Education is important, but expensive
3. Science and advocacy can and should be partners
4. Lessons learned from newborn hearing screening
Montreal School
For the Deaf
Group 4 1977-78
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 Assistive Listening Devices

Advocacy and Public Policy Initiatives

What enabled us to move from ....

There to Here?
Why is Early Identification of Hearing Loss so Important?

• Hearing loss occurs more frequently than any other condition for which population-based screening is done.
Frequency of Congenital Hearing Loss?

- 1 per 1,000
- 2 per 1,000
- 3 per 1,000
- 6 per 1000
## Rate Per 1000 of Permanent Childhood Hearing Loss in EHDI Programs

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Incidence per 10,000 of Congenital Conditions

Hearing Loss
Cleft lip or palate
Down Syndrome
Limb defects
Spina bifida
Hypothyroidism
Sickle Cell Anemia
PKU
Why is Early Identification of Hearing Loss so Important?

- Hearing occurs more frequently than any other birth defect.

- Undetected hearing loss has serious negative consequences.
Reading Comprehension Scores of Hearing and Deaf Students

**Effects of Unilateral Hearing Loss**

Keller & Bundy (1980)  
(n = 26; age = 12 yrs)

Peterson (1981)  
(n = 48; age = 7.5 yrs)

(n = 50; age = 10 yrs)

By 3rd grade, the average child is ~24 months behind his or her peers in math, language and social skills.

**Average Results**  
Math = 30th percentile  
Language = 25th percentile  
Social = 32nd percentile
Why is Early Identification of Hearing Loss so Important?

• Hearing loss occurs more frequently than any other birth defect.

• Undetected hearing loss has serious negative consequences.

• There are dramatic benefits associated with early identification of hearing loss.
Boys Town National Research Hospital Study of Earlier vs. Later

129 deaf and hard-of-hearing children assessed 2x each year.

Assessments done by trained diagnostician as normal part of early intervention program.

Moeller, M.P. (1997). Personal communication moeller@boystown.org
Newborn Hearing Screening
Prior to 1990

- **Conventional Auditory Brainstem Response**
  - Accurate, but too expensive

- **High Risk indicators**
  - Only about 50% of children with congenital hearing loss exhibit one or more of these high risk indicators
What Percentage of Hearing Impaired Children were High Risk as Infants?

- Feinmesser et al. (1982): 49%
- Pappas & Schaibly (1984): 54%
- Elssmann et al. (1987): 48%
- Watkin et al. (1991): 43%
- Mauk et al. (1991): 50%
- Mehl & Thomson (1998): 50%
**Accuracy of High Risk Based UNHS Programs**

Mahoney and Eichwald (1987)

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JCIH indicators incorporated into legally required birth certificate.

Computerized mailing and follow-up, and free diagnostic assessments at regional offices and/or mobile van.

Program now discontinued because:

- parents only made appointments for about 1/2 the children who had a risk indicator.

- only about 1/2 of the children with an appointment showed up.

- difficulty obtaining accurate information from hospitals for some risk indicators.

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Newborn Hearing Screening Prior to 1990

• **Auditory Brainstem Response**
  – Accurate, but too expensive

• **High Risk indicators**
  – Only about 50% of children with congenital hearing loss exhibit high risk indicators
  – Only about ½ of those with high risk indicators make an appointment for further testing and only about ½ of those are ever tested

• **Behaviorally-based hearing screening**
  – Expensive
  – Inaccurate
Percentage of Children with Permanent Hearing Loss Identified by the Infant Distraction Test Performed at 8 Months of Age

Severe/Profound Bilateral (n = 39)
Mild/Moderate Bilateral (n = 72)
Unilateral (n = 60)

From 1988-1993 the first large-scale clinical trial of universal newborn hearing screening was conducted -- the Rhode Island Hearing Assessment Project --

UNIVERSAL NEWBORN HEARING SCREENING USING TRANSIENT EVOKED OTOACOUSTIC EMISSIONS: RESULTS OF THE RHODE ISLAND HEARING ASSESSMENT PROJECT

Karl R. White, Ph.D., Betty R. Vohr, M.D., and Thomas R. Behrens, Ph.D.

The earlier that hearing loss can be identified and intervention begun, the better the prognosis for the child in areas ranging from language development to academic success, social interactions, and successful participation in society. Indeed, early identification of significant hearing loss is so important that the U.S. Department of Health and Human Services (HHS) recently set a goal to reduce to 12 months the average age at which significant hearing loss can be identified using auditory brainstem response (ABR) to identify hearing loss among infants and toddlers.\textsuperscript{1,2} Such research certainly contributed substantially to the American Speech-Hearing Language Association's (ASHA) recommendation of ABR as the preferred method for screening the hearing of newborns.\textsuperscript{3} However, the expense of doing ABR testing of newborns was very likely what led to ASHA's recommendation that it be done only with
In March, 1993 an NIH Consensus Panel concluded that:

- The average age of diagnosis of hearing loss remains constant at about 2 ½ years of age.
- All infants should be screened for hearing loss…this will be accomplished most efficiently by screening prior to discharge from the well-baby nursery.
- Identification of hearing loss must be seen as imperative for all infants.
Percentage of Newborns Screened for Hearing in the United States
Then a miracle occurs

Good work, but I think we might need a little more detail right here
The Impact of Early Hearing Detection and Intervention (EHDI) Programs on Early Usher Diagnosis

- Screening before 1 month
- Diagnosis before 3 months
- Intervention before 6 months

Medical Home

Data Management and Tracking

Program Evaluation and Quality Assurance

Family Support!!
The Impact of Early Hearing Detection and Intervention (EHDI) Programs on Early Usher Diagnosis

1. A Rising Tide Lifts All Ships

2. Education and Public Awareness

3. Advocacy and Policy Initiatives.
Status of EHDI Programs in the US: Universal Newborn Hearing Screening

• With ~95% of infants screened, newborn hearing screening has become the “standard of care”

• There are hundreds of excellent programs - - - regardless of the type of equipment or protocol used

• Some programs are still struggling with high refer rates and poor follow-up
Does a 2-stage (OAE/AABR) newborn hearing screening protocol miss babies with mild hearing loss?

**Study Sample**
Comprehensive Audiological Assessment at 8-12 months of age

**Comparison Group**
Comprehensive Hearing Evaluation Before 6 Months of Age
How Many Additional Babies with Permanent Hearing Loss were Identified?

<table>
<thead>
<tr>
<th></th>
<th>Comparison Group (Fail OAE/ Fail AABR)</th>
<th>Study Group (Fail OAE/ Pass AABR)</th>
<th>Total</th>
</tr>
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<tbody>
<tr>
<td>Number of Babies</td>
<td>158</td>
<td>21</td>
<td>179</td>
</tr>
<tr>
<td>Prevalence per 1,000</td>
<td>1.82</td>
<td>.55*</td>
<td>2.37</td>
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*Adjusted for proportion of OAE fails that enrolled

Represents 23% of all babies with PHL in birth cohort

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
- Programs now being replicated in 12 additional states

Hearing Screening During Well Child Visits to Health Care Providers

- Pilot studies and materials development 2005-2006
- Worked with American Academy of Pediatrics to develop recommended policy changes
- Training and implementation materials available from www.HearAndNow.org
Status of EHDI Programs in the United States

- Universal Newborn Hearing Screening
- Effective Tracking and Follow-up as a part of the Public Health System
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89% of states have created a statewide tracking system
  
  - information submitted for 80% of the births in 2003
  
  - 72% have individual identifying data --- up from 32% in 2001

57% track babies until at least 3 years of age

Linkages with other Public Health Information systems are expanding (eg, Vital Statistics, heelstick, EI, Immunizations)
Status of EHDI Programs in the United States

- Universal Newborn Hearing Screening
- Effective Tracking and Follow-up as a part of the Public Health System
- Appropriate and Timely Diagnosis of the Hearing Loss
Confirmation of Permanent Hearing Loss

- Coplan (1987) - 35 months
- Eissman et al. (1987) - 19 months
- Gustason (1987) - 30 months
- Meadow-Orlans (1987) - 30 months
- Yoshinago-Itano (1995) - 24 months
- Stein et al. (1990) - 25 months
- Mace et al. (1991) - 31 months
- O’Neil (1996) - 56 months
- Johnson et al. (1997)* - 3 months
- Vohr et al. (1998)* - 3 months

Average Age in Months
Status of EHDI Programs in the US: Audiological Diagnosis

- Equipment and techniques for diagnosis of hearing loss in infants continues to improve.
- Severe shortages in experienced pediatric audiologists delays confirmation of hearing loss.
- State coordinators estimate only 56.1% “receive diagnostic evaluations by 3 months of age.”
Status of EHDI Programs in the United States

- Universal Newborn Hearing Screening
- Effective Tracking and Follow-up as a part of the Public Health System
- Appropriate and Timely Diagnosis of the Hearing Loss
- Prompt Enrollment in Appropriate Early Intervention
Status of EHDI Programs in the US: Early Intervention

- Current system designed to serve infants with bilateral severe/profound losses— but, majority of those identified have mild, moderate, and unilateral losses

- State EHDI Coordinators estimate that only 53% of infants with hearing loss are enrolled in EI programs before 6 months of age

- Public or insurance funding is seldom available for high quality hearing aids
Most Early Intervention Programs for Children with Hearing Loss are “Missing the Mark”

- 95% of all newborns with hearing loss have parents with normal hearing.
- When parents in North Carolina were given a choice
  
  **In 1995:** 60% chose sign-language options; 40% chose auditory-oral
  
  **In 2005:** 15% chose sign-language options; 85% chose auditory-oral

- The number of cochlear implant for children under age 5 has quadrupled in the last 4 years (to 2000+ implants per year)


Primary Emphasis of Personnel Preparation Programs for Teachers of Deaf and Hard of Hearing

Primary Emphasis

*Sign Language-based*

*Spoken Language-based*

LEGEND

Graduates per Year:

- 1-5
- 6-15
- 16+

Note: Although many programs describe themselves as providing “comprehensive” services, most have a primary emphasis on a specific approach as indicated by the curriculum offerings, the placement of graduates, the type of practicum available, etc. Classification of programs on this map considered those factors in conjunction with annual self-report survey data from the 2004 and 2005 issues of the *American Annals of the Deaf*. 
Status of EHDI Programs in the United States

• Universal Newborn Hearing Screening
• Effective Tracking and Follow-up as a part of the Public Health System
• Appropriate and Timely Diagnosis of the Hearing Loss
• Prompt Enrollment in Appropriate Early Intervention
• A Medical Home for all Newborns
EHDI and the Medical Home

Parent Groups
Mental Health

Birthing Hospital

Audiology

3rd Party Payers

Deaf Community

Services for Hearing Loss

Primary Health Care Provider
Child/Family

Early Intervention Programs

ENT

Genetics

Community Services for Hearing Loss
### Educating Primary Health Care Providers About Early Identification of Hearing Loss

Assume a newborn for whom you are caring is diagnosed with a moderate to profound bilateral hearing loss. If no other indications are present, would you refer the baby for a(n):

<table>
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<th>Evaluation</th>
<th>Always or Often</th>
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<tr>
<td>Ophthalmological evaluation</td>
<td>0.6%</td>
</tr>
<tr>
<td>Genetic evaluation</td>
<td>8.9%</td>
</tr>
<tr>
<td>Otolaryngological evaluation</td>
<td>75.6%</td>
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Responses of 1975 physicians in 21 states

American Academy of Pediatrics

Universal Newborn Hearing Screening, Diagnosis, and Intervention Guidelines for Pediatric Medical Home Providers

Medical Evaluations
To determine etiology and identify related conditions

- Ophthalmologic (annually)
- Genetic
- Developmental pediatrics, neurology, cardiology, and nephrology (as needed)

Pediatric Audiologic Services
- Behavioral response audiometry
- Ongoing monitoring
When can an infant be fit with hearing aids?
<table>
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<tr>
<th>Type of Physician</th>
<th>&lt;=1 mo</th>
<th>2-3 mos</th>
<th>4-6 mos</th>
<th>7-11 mos</th>
<th>12+ mos</th>
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<td>Pediatrician (n=1145)</td>
<td>36.3%</td>
<td>16.9%</td>
<td>29.0%</td>
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- A Medical Home for all Newborns
- Culturally Competent Family Support
Information Wanted vs. Received by Parents at Hearing Loss Confirmation

Martin, George, O'Neal, & Daly (1987); *Sweetow & Barrager (1980)

- Degree of loss
- Auditory system
- Amplification
- Educational options
- Speech/Lang dev
- Etiology
- Home activities
- *Written Information
- *Financial Support
- *Emotional Support
- *Parent Contacts
- *Referral Sources

Wanted vs. Received

Martin, George, O'Neal, & Daly (1987); *Sweetow & Barrager (1980)
Are current EHDI materials effective?
Brochure Readability

Gold Standard Readability: ≤6th Grade

- Initial Screening / Retest
- Intervention

- 7th
- 8th-9th
- 10th-12th
- College+
Policy and Legislative Initiatives with Local, State and Federal Partners
Take Home Messages

- The world has changed for infants and young children with permanent hearing loss
- Screening is only the first (and the easiest!) step
- Just as scientific and technological advances have made the revolutionary changes of the last 15 years possible --- more are coming
- Education and advocacy are the foundation on which future progress will be built
- Usher Syndrome is one of many specific conditions (but an important one) that will benefit from effective comprehensive screening for permanent hearing loss
To ensure that all infants and toddlers with hearing loss are identified as early as possible and provided with timely and appropriate audiological, educational, and medical intervention, an early hearing detection and intervention (EHDI) program should comprise three basic components—newborn hearing screening, audiological diagnosis, and early intervention. Threaded throughout these components should also be some key elements—culturally-competent family support, medical home, data management, legislative mandates, and program evaluation tools. Follow the links below to find information about these basic components and key elements, and about other related EHDI resources and information.
Appropriate management of all persons identified with congenital hearing loss, as defined above, requires a comprehensive genetic evaluation.
... families should be offered the option of genetic evaluation and counseling by a medical geneticist