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Universal Newborn Hearing Screening: The Evolution of a Public Health Revolution

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Disclosure

• Christine Yoshinaga-Itano, Ph.D.
• Professor
• Speaker Disclosures
  – Relevant financial relationships to disclose: AAS (expenses, honorarium)
  – Relevant non-financial relationships: Scientific advisory board LENA foundation
Learning Objectives

• After this course learners will be able to explain what evidence was required to establish universal newborn hearing screening and EHDI systems.
• After this course learners will be able to identify characteristics/variables that can negatively impact developmental outcomes of children who are deaf or hard of hearing even when early-identified.
• After this course learners will be able to identify what interventions could positively impact developmental outcomes of at-risk populations.

Marion Downs Center
established 1996 with MCH grant

• Under the leadership of the Marion Downs Center, the US went from 2 states with legislation in 1996 to 30 states with legislation by the end of 2000
UNHS legislation

- 1990: Hawaii
- 1992: Rhode Island
- 1997: Colorado, Connecticut, Mississippi
- 1998: Utah, Virginia, West Virginia
- 1999: Arkansas, Georgia, Illinois, Indiana, Kansas, Louisiana, Maine, Maryland, Missouri, North Carolina, New York, Oregon, Texas, Wisconsin, Wyoming
- 2000: Florida, Kentucky, Nebraska, New Hampshire, Nevada, Oklahoma, South Carolina
- 2001: District of Colombia, Montana, New Mexico, Pennsylvania

Formula for success

*Met with Chair of the Colorado State Genetic/Metabolic Screen Committee-legislation to establish the only mandated newborn screening*

*She provided the formula for gaining support from Public Health and the State legislature for establishing a universal newborn screen.*
Anatomy of an universal newborn screening program

Frequency of the Disorder Merits Screening

Ability to detect accurately in mass screening

Acceptable ratio of false positive screening

Screening programs do not cause parental harm

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Anatomy of a Screening Program

Effective treatment is available and assured

Early intervention improves outcome

Costs are reasonable and justified
Who had to be convinced

The general public
Genetic newborn screen committee
Pediatricians/Family Practice Physicians
US Preventative Task Force
Insurance companies/Medicaid
Hospital Associations
Ethicists
Audiologists
Early Intervention providers
Consumers who were deaf or hard of hearing and their families

Criteria for securing support from AAP

Met with Fetus and Newborn committee of the AAP (American Academy of Pediatrics)

Criteria for support: Demonstrate that the impact of deafness is as serious as PKU and the intervention is as successful.

Genetic/metabolic: That social-emotional harm is not like cystic fibrosis
Theoretical foundation for NIH 6 year contract – 1994-2000

- Investigation of connectionist theories of language development including cognition & social-emotional variables
- Different predictors for different aspects of language development: semantics, syntax/morphology, pragmatics, phonology
- Different weighting of predictors at different developmental stages
- Age of identification was not an identified variable

FREQUENCY OF THE DISORDER MERITS SCREENING
### Frequency of disorder in every 100,000 births

<table>
<thead>
<tr>
<th>Disorder</th>
<th>Frequency</th>
</tr>
</thead>
<tbody>
<tr>
<td>Congenital hearing loss</td>
<td>260</td>
</tr>
<tr>
<td>Bilateral hearing loss</td>
<td>200</td>
</tr>
<tr>
<td>Hypothyroidism</td>
<td>25</td>
</tr>
<tr>
<td>Phenylketonouria</td>
<td>7</td>
</tr>
<tr>
<td>Cystic Fibrosis</td>
<td>15</td>
</tr>
<tr>
<td>Hemoglobinopathy</td>
<td>30</td>
</tr>
</tbody>
</table>

### ABILITY TO DETECT DISORDER IN MASS SCREENING

Acceptable ratio of false positive rate

Mehl & Thompson, 1998
Screening Rates at Hospital Discharge


Most states report screening rates of 2 to 7% at hospital discharge

Positive Predictive Value

The probability that a positive screening result represents a true positive detection of a disorder

Number of infants with a positive test and a hearing loss divided by the number of infants with a positive test

Number referred at 1st screen:
1100/54000 births

Number referred at 2nd screen for audiological diagnostic evaluation:
550/54000 births

140 children identified

26% Positive Predictive Value from 2nd outpatient screen

Does the screening detect HL Positive Predictive Value with a low false positive rate

The probability that a positive screening result represents a true positive detection of a disorder

Hypothyroidism 3%
Cystic Fibrosis 4%
PKU 80%
Colorado Hearing Loss 26%
Screening programs do not cause parental harm

To parents whose babies have normal hearing and are screened
No sig difference between parents whose babies were screened versus those who were not screened (Abdala de Uzcategui & Yoshinaga-Itano, 2000)

To parents whose babies are deaf or hard of hearing and are screened
No sig difference with hearing: maternal bonding-emotional availability, parental stress (Pipp-Siegel et al., 2003; Pearman et al., 2000)

Cost savings: Total Screening Costs per Confirmed Diagnosis

- Congenital Hearing Loss: $9,600
- Bilateral Hearing Loss: $12,300
- Hypothyroidism: $10,800
- Phenylketonuria: $40,500
- Cystic Fibrosis: $6,000
- Hemoglobinopathy: $23,100
NECESSARY TO PROVE: DIAGNOSIS HAS TO OCCUR CLOSE TO NEWBORN PERIOD OR OPTIMAL OUTCOMES MAY NOT BE ACTUALIZED

EARLY INTERVENTION IMPROVES OUTCOMES – POSITIVE MESSAGE – PREVENT COMMUNICATION DISORDER

Variables predictive of successful language outcome

• Yoshinaga-Itano, Sedey, Coulter & Mehl, 1998 in Pediatrics

• Early Identified Group I: 72 children (34 males, 38 females) had hearing loss identified before six months

• Later Identified Group II: 78 children (41 males; 37 females) had hearing loss identified after six months of age
Adjusted mean total language quotients for groups based on age of identification of hearing loss.

Figure 9. Mean total language quotient scores at 31 to 36 months by age of identification of hearing loss and cognition.
Variables predictive of successful language outcome

• Not only did EID children have significantly better language outcomes than LID children. Language outcomes were within the normal range.

• Children with normal cognitive status have expressive and receptive language DQs between 85 and 102 (within the normal range of development) when their hearing losses are identified by 6 months of age.

CONCLUSIONS BY AAP

• Screening had to be done in the newborn period in order to be able to diagnose, fit with amplification and enroll into early intervention by 6 months.

• Failing to diagnose and enroll results in performance for children with hearing loss and normal cognition that is equivalent to children with hearing loss and cognitive disabilities – creating an environmentally produced cognitive disability.
CONCLUSIONS BY AAP

• It is possible for children with hearing loss to enter kindergarten with developmental skills comparable to their hearing typically developing peers. (Yoshinaga-Itano, Coulter & Thompson, 2000, 2001)

Is there an effect?

• Kennedy et al., 2006
• N=57 confirmed HL < 9 months
• N=63 confirmed HL > 9 months
• N=63 normal hearing
• ID < 9 month significantly higher scores
• Receptive language
• Expressive language
IS THE EFFECT LASTING?

MODEL PREDICTION OF VARIANCE OF LANGUAGE AT 84 MONTHS AND RATE OF LANGUAGE DEVELOPMENT

- **68%** of variance in expressive vocabulary (EOWPVT) outcome at 84 mo.
- **71%** of the variance in receptive syntax (TACL III) outcome at 84 mo.
- **46%** of rate of language growth for EOWPVT
- **81%** of rate of language growth for TACL III (receptive syntax)
PARENT WORD FREQUENCY: EOWPVT III
Lower (984) Inter (1245) Upper (1515) ½ hr videotape
**EOWPVT differences by Maternal Level of Educational Level (Baca, 2009) - emerges after 3 years**

- 35 month language age difference at 84 months of age between group with mean age level for mothers with educational level less than 12 years (HS grad) as compared to group for mothers with educational level 16 years or greater (college)

55.75 months versus 91.33 months
Data Collection and Processing

• Digital recorder children wear
• Records continuously for 16 hours
• Audio transferred to computer
• Speech recognition software processes file, automatically analyzing audio stream

LENAs technology data: Aragon & Yoshinaga-Itano, 2012
Effect is lasting

- Pemperton et al., 2014 – Long term literacy outcomes – not screening but identification of HL by 9 months
- Teen outcomes for N=35 ID before 9 months
- .6 SD from mean of typically developing

TREATMENT IS ASSURED
Colorado controlled the quality of early intervention

- Early first contact is knowledgeable and immediate
- Early amplification
- All early intervention providers have specialized training in early childhood deafness and hearing loss
- Intensity or dose of intervention is consistent across the population
- Parent partnerships and Deaf/Hard of Hearing partnerships are infused into the system

- Early intervention providers participate in regular in-service training
- Developmental assessment is monitored every 6 months
- Mentors – Colorado Hearing Coordinators and consultants monitor the fidelity of the intervention
- Screening, early ID and early amplification are not enough (Ching et al., 2014)
THE PRESENT: HOW ARE THE CHILDREN DOING ACROSS THE UNITED STATES?

NECAP: National Early Childhood Assessment Project: Deaf/Hard of Hearing

- 15+ states have committed to collecting state-wide data.
- 4/15 states other than Colorado now have population data of developmental outcomes.
- 1077 assessments (English)
- 142 (Spanish)
States Represented in Results

- ENGLISH & SPANISH
- Arizona
- California
- Colorado
- Idaho
- Indiana
- Texas
- Wyoming
- ENGLISH DATA ONLY
- Wisconsin
- Maine
- Minnesota
- New Mexico
- Oregon
- Utah

Participant Characteristics

<table>
<thead>
<tr>
<th>Age at…</th>
<th>Median Eng (mos)</th>
<th>Median Span (mos)</th>
<th>Range (mos)</th>
<th>Range</th>
</tr>
</thead>
<tbody>
<tr>
<td>Identification</td>
<td>2.3*</td>
<td>2*</td>
<td>.25 to 48</td>
<td>.25 to 30</td>
</tr>
<tr>
<td>Amplification</td>
<td>5</td>
<td>6</td>
<td>1 to 48</td>
<td>1.5 to 32</td>
</tr>
<tr>
<td>Intervention</td>
<td>5*</td>
<td>6*</td>
<td>.25 to 44</td>
<td>1 to 31</td>
</tr>
</tbody>
</table>

*67% (Eng) 59% (Span) of children were identified by 3 mos.
*66% (Eng) 57% (Span) of children were in intervention by 6 mos.
### Participant Characteristics

<table>
<thead>
<tr>
<th>Highest degree completed</th>
<th>% of primary caregivers English</th>
<th>% of primary Caregivers Spanish</th>
</tr>
</thead>
<tbody>
<tr>
<td>Less than HS</td>
<td>8%</td>
<td>47%</td>
</tr>
<tr>
<td>High school diploma</td>
<td>46%</td>
<td>33%</td>
</tr>
<tr>
<td>Vocational or Associates</td>
<td>16%</td>
<td>9%</td>
</tr>
<tr>
<td>Bachelor’s degree</td>
<td>18%</td>
<td>11%</td>
</tr>
<tr>
<td>Graduate degree</td>
<td>12%</td>
<td>0%</td>
</tr>
</tbody>
</table>

### Median Language Quotients: English vs. Spanish

<table>
<thead>
<tr>
<th>Assessment</th>
<th>Language Quotient English</th>
<th>Language Quotient Spanish</th>
</tr>
</thead>
<tbody>
<tr>
<td>Minn Exp</td>
<td>92</td>
<td>78</td>
</tr>
<tr>
<td>Minn Concept</td>
<td>84</td>
<td>67</td>
</tr>
<tr>
<td>Mac Vocab</td>
<td>81</td>
<td>74</td>
</tr>
</tbody>
</table>
Percent of Scores in the Average Range (LQ = 80+)

Minnesota CDI: Median Language Quotients
MacArthur-Bates: Median Vocabulary Production Quotients

Deaf/hard of hearing parents versus hearing parents
THE PAST

Intervention by 6 months versus after 6 months
Implementation of Universal Newborn Hearing Screening resulted in

- Implementation of ABR both tone and click
- Implementation of ASSR
- Implementation of TEOAE and DPOAE in both screening and diagnostic evaluations
- Implementation of high frequency tympanometry
- Implementation of diagnostic procedures for auditory neuropathy – cochlear microphonic
- Earlier cochlear implantation

THE FUTURE:
What we are not yet doing

• VALIDATING AMPLIFICATION FIT IN THE NEWBORN:
  – Cortical auditory evoked potentials
  – Speech perception for infants and toddlers-VRISD, CAEP
• Demonstrating efficacy or fidelity of intervention strategies for infants and toddlers
• Cochlear implantation below the age of 1 year
• Measuring quantity of adult words, conversational turns and child vocalizations occur in an average day-LENA

What if….

• You could measure ability of child to detect/discriminate change from one phoneme to another at 2 months of age
• Clinical protocol – ½ hour
• Automated calculation
• Affordable equipment
• Physiological validation of amplification fitting
Validating amplification fitting: Cortical auditory evoked potentials – 2 month olds

- Cone, B. (2014) CAEP amplitude and latency – vowel discrimination
- Gilley & Uhler (2014) CAEP MMN for white noise, vowels (NH/DHH) and consonants (NH – not yet run DHH)

MMR/N (using PCA) in HOH infant and toddler while wearing hearing aids (Uhler & Gilley, 2014)
Uhler & Gilley, 2015 pilot data

- To address Signal to Noise Ratio and inter-trial variability issues, Gilley developed a method that employs a Principal Component Analysis-based reduction of time-frequency transformed and frequency normalized EEG. Results from this algorithm revealed a significant MMFR in all 12 infants in the theta, alpha, and beta bands (Figure 1). The consistent MMFRs at both the individual and group levels of analysis suggest that the MMFR is a valid tool for translational investigation.
What if…

- We could behaviorally test speech discrimination with a stopping criteria using an automated clinical protocol from 6 to 15 months of age.
- We could assure by 12 months of age that children with amplification fitting- hearing aids and cochlear implants could discriminate all combinations of English phonemes

Neurodynamics Laboratory

Behavioral testing (VRA / VRISD) – Intelligent Hearing Systems

VRISD: 12 Month Old Bilateral Hearing Aid User
Neurodynamics Laboratory
Behavioral testing (VRA / VRISD)

VRISD: 16 Month Old C.I. User post IA

Uhler, 2010 Subject 3: M.G. Vowel Discrim

<table>
<thead>
<tr>
<th></th>
<th>/a/-/i/</th>
<th>/a/-/u/</th>
<th>/u/-/i/</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pre CI</td>
<td>62%</td>
<td>0%</td>
<td>0%</td>
</tr>
<tr>
<td>24 hrs</td>
<td>69%</td>
<td>55%</td>
<td>85%</td>
</tr>
<tr>
<td>1 mo.</td>
<td>92%</td>
<td>91%</td>
<td>85%</td>
</tr>
<tr>
<td>2 mo.</td>
<td>92%</td>
<td>92%</td>
<td>91%</td>
</tr>
<tr>
<td>3 mo.</td>
<td>91%</td>
<td>92%</td>
<td>91%</td>
</tr>
</tbody>
</table>
### Subject 3: Voicing

<table>
<thead>
<tr>
<th></th>
<th>ta-da</th>
<th>ka-ga</th>
<th>pa-ba</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pre CI</td>
<td>0%</td>
<td>DNT</td>
<td>DNT</td>
</tr>
<tr>
<td>24 hr.</td>
<td>92%</td>
<td>DNT</td>
<td>DNT</td>
</tr>
<tr>
<td>1 mo.</td>
<td>91%</td>
<td>DNT</td>
<td>DNT</td>
</tr>
<tr>
<td>2 mo.</td>
<td>93%</td>
<td>DNT</td>
<td>DNT</td>
</tr>
<tr>
<td>3 mo.</td>
<td>94%</td>
<td>92%</td>
<td>90%</td>
</tr>
</tbody>
</table>

### Case 3: Place of Articulation

<table>
<thead>
<tr>
<th></th>
<th>pa-ka</th>
<th>ta-ka</th>
<th>pa-ta</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pre CI</td>
<td>51%</td>
<td>DNT</td>
<td></td>
</tr>
<tr>
<td>24 hr.</td>
<td>81%</td>
<td>DNT</td>
<td></td>
</tr>
<tr>
<td>1 mo.</td>
<td>92%</td>
<td>66%</td>
<td></td>
</tr>
<tr>
<td>2 mo.</td>
<td>90%</td>
<td>90%</td>
<td>90%</td>
</tr>
<tr>
<td>3 mo.</td>
<td>92%</td>
<td>DNT</td>
<td>91%</td>
</tr>
</tbody>
</table>
What if…

• We designed auditory habilitation/treatment for infants based on evidence-based practice
• DHH infants were exposed to phoneme contrasts and could demonstrate discrimination for all contrasts by 12 months of age
• Many children with 5 to 10 minute exposure can learn to master discrimination of phoneme pairs

What if…

• We could evaluate the effectiveness of auditory therapies/interventions by the change in cortical auditory pathway organizations
Beyond sensori-neural hearing loss

- Mason, 2014 (epidemiologist, EHDI, Maine)
- Analyzed the literacy outcomes 3rd-12th grade of:
  - Children who failed the initial screen but passed the second screen
  - Children who failed both screens
  - Children who passed the initial screen
- Children who failed the initial screen but eventually passed had significantly lower literacy rates

ACOUSTIC FEATURES OF SPEECH MEASURED BY LENA TECHNOLOGY ACCURATELY PREDICT AUTISM DIAGNOSIS

Oller, et al 2010, Xu et al., 2012
Frequency of Consonant-like Sound

**t-test**
(Welch 2-sample 2-side)

- **TD** versus **ASD:**
  \[ t(90) = 7.95^{***} \]

- **TD** versus **LD:**
  \[ t(68) = 5.52^{***} \]

- **LD** versus **ASD:**
  \[ t(118) = 2.62^{**} \]

Correlation with age:
- **TD:** 0.67^{***}
- **LD:** 0.42^{**}
- **ASD:** 0.32^{**}

Probability of Sound Collision

**t-test**
(Welch 2-sample 2-side)

- **ASD** versus **TD:**
  \[ t(132) = 3.66^{***} \]

- **ASD** versus **LD:**
  \[ t(111) = 2.94^{**} \]

- **TD** versus **LD:**
  \[ t(90) = 0.13 \]

\*p<0.05
\**p<0.01
\***p<0.001
Child Vowel Volume (dB)

Mean and Standard-Error

- TD
- LD
- ASD

**t-test** (Welch 2-sample 2-side)

- ASD versus TD: t(125) = 5.84***
- ASD versus LD: t(117) = 4.78***
- TD versus LD: t(97) = 0.45

*p<0.05
**p<0.01
***p<0.001

Result of C-MLU: Trajectories & Correlation with Chronological Age

Correlation with chronological-age:

- HH: 0.51 ***
- TD: 0.63 ***
- LD: 0.32 *
- ASD: 0.32 *

*: p < 0.05
**: p < 0.01
***: p < 0.001
Conclusion

- Unique Characteristics of Children with Autism:
  - Less Frequent Consonant-like Sounds
  - Higher Chance of Sound Collision
  - Louder Vowel-like Sounds
  - Lower Spectrum Entropy of Unvoiced Consonant Sounds
  - (how noise-like versus tone-like a sound is)
  - Discriminant Analysis: 94% (6% Equal-Error-Rate)

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- University of Colorado