NORTH AMERICAN CONFERENCE ON DEAFNESS SCREENING AND INTERVENTION IN EARLY CHILDHOOD

ABSTRACTS AND POWERPOINTS

LENDING AN EAR
EXTENDING A HAND
ENCOURAGING SUCCESS

JANUARY
23 • 24 • 25, 2003

QUEBEC CITY, CANADA
HOTEL HILTON QUEBEC

www.inspq.qc.ca/ConferenceDeafnessScreening
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EARLY HEARING DETECTION AND INTERVENTION SYSTEMS (EHDI) IN THE NEW MILLENNIUM

Dr. Betty Vohr, Department of Pediatrics, Women and Infants Hospital, Brown University, Providence, Rhode Island, United States
Thursday, January 23, 2003, 9:00 to 10:00 AM

Newborn hearing screening for permanent hearing loss fulfills the recommended criteria for universal screening. The incidence of permanent hearing loss is between 2 and 3 per 1000 which is higher than any of the metabolic disorders; there are reliable methods available (automated auditory evoked response and transient evoked otoacoustic emissions); late identification has serious negative effects on outcome, and treatments and interventions are available to modify the outcome. In 1999, the American Academy of Pediatrics Task Force on Newborn and Infant Hearing endorsed universal newborn hearing screening programs to provide screening, early assessment and intervention. Thirty-eight states in the U.S. now recommend or mandate such screening.

Three protocols are commonly used for universal newborn hearing screening programs:
(1) transient evoked otoacoustic emissions (TEOAE) or distortion product otoacoustic emissions (DPOAE),
(2) automated auditory brainstem response (AABR), and
(3) a “two-step” program in which infants are first screened with OAE followed by an AABR screen for those infants who fail the OAE screen.

Access to audiologists experienced with infants and young children, working in appropriately equipped facilities sensitive to the impact of diagnostic news on parents, undergirds successful universal newborn hearing screening. Timely follow-up, empathetic counselling, reasonable reimbursement for services, and a system-wide collaboration that supports families are also needed. For most infants with hearing loss, a key component of intervention will be the use of amplification and other assistive listening devices. The goal of providing auditory input, specifically the best amplified speech signal possible through amplification by no later than six months of age, is recommended.

Children ages birth to 3 years identified with hearing loss are eligible and should receive early intervention services. Early identification of hearing loss in conjunction with early intervention holds with it the promise of early and beneficial experiences that enable parents to help their child develop language and communication.
Early Hearing Detection and Intervention (EHDI) Systems in the New Millennium

Betty Vohr, M.D., FAAP
Brown Medical School

Background

Universal newborn screening in the US is a public health program for identification of conditions for which early intervention or treatment lead to a reduction of mortality, morbidity or disabilities associated with the condition.

Criteria for Eligibility for Universal Newborn Screening

The disorder must have the following:

- a reasonable incidence among newborns
- reliable methods available for screening
- there is a negative impact if identified late
- there are interventions available to improve outcome

Does Newborn Hearing Screening Fit These Criteria?

- incidence of permanent hearing loss 2-3/1000
- reliable methods for screening AABR/TEOAE
- late identification has serious effects yes
- Rx and interventions are available to improve outcomes yes

Characteristics of Children with Hearing Loss

<table>
<thead>
<tr>
<th>Site</th>
<th>Rate</th>
</tr>
</thead>
<tbody>
<tr>
<td>Well Baby Nursery</td>
<td>1 per 1000</td>
</tr>
<tr>
<td>NICU</td>
<td>10 per 1000</td>
</tr>
<tr>
<td>Total population</td>
<td>2-4 per 1000</td>
</tr>
<tr>
<td># infants ident annually US</td>
<td>8,000-16,000</td>
</tr>
<tr>
<td>Average career pediatrician</td>
<td>12 patients</td>
</tr>
</tbody>
</table>
How Does Permanent HL Compare to Other Screenable Newborn Disorders?

Sickle cell disease* 47 per 100,000
Hypothyroidism* 26 per 100,000
PKU* 2 per 100,000
Adrenal hyperplasia* 2 per 100,000
Permanent HL 220 per 100,000

*Texas Dept. of Health

Current Methods Available

- Transient Evoked Otoacoustic Emissions (TEOAE or DPOAE)
- Automated Auditory Brainstem Response (AABR)
- Standard Auditory Brainstem Response (ABR)
- 2 Step: OAE + AABR

OAE

AABR
Rate per 1000 of Permanent Childhood Hearing Loss in UNHS Programs

<table>
<thead>
<tr>
<th>Site</th>
<th>Sample Size</th>
<th>Prevalence per 1000</th>
</tr>
</thead>
<tbody>
<tr>
<td>Rhode Island 1993-94</td>
<td>16,395</td>
<td>1.71</td>
</tr>
<tr>
<td>Colorado 1992-96</td>
<td>41,976</td>
<td>2.56</td>
</tr>
<tr>
<td>New York 1995-1997</td>
<td>69,761</td>
<td>1.95</td>
</tr>
<tr>
<td>Texas 1994-97</td>
<td>52,508</td>
<td>2.15</td>
</tr>
<tr>
<td>Hawaii 1996-96</td>
<td>9,605</td>
<td>4.15</td>
</tr>
<tr>
<td>New Jersey 1993-95</td>
<td>15,749</td>
<td>3.30</td>
</tr>
</tbody>
</table>

Why is Early Identification of Hearing Loss so Important?

- Hearing loss is the most frequent birth defect.
- Undetected hearing loss has serious negative consequences.
- There are dramatic benefits associated with early identification of hearing loss.

Better Language Outcomes After Early Identification of HL

- White: ↑ language scores of severe to profound (14 vs 26 m)
- Apuzzo: better language scores at age 4 if ident ≤2 m
- Moeller: 100 DHH children with early ident = better outcomes
- Yoshinagatozawa: better scores at 36 m if ident <6 m

What is Happening With Universal NB Hearing Screening in US?

- It is developing as a public health program
- 37 states currently have legislation mandating universal screening
- All states are taking steps to develop screen programs
- RI was the 1st state to successfully implement Universal Newborn Hearing Screening for 99% of newborns in 1993.
AAP & JCIH Recommendations
Components of EHDI Programs in the US
- Universal Newborn Hearing Screenings < 1 m
- Effective Tracking and Follow-up as a part of the Public Health System
- Appropriate and Timely Diagnosis of the HL < 3 m
- Prompt Enrollment in Appropriate EI < 6 m
- All infants will have a medical home

Components of A Medical Home for EHDI
includes Family Care that is:
- accessible
- family centered
- comprehensive
- continuous
- coordinated
- compassionate
- culturally competent

How Does the Concept at the Medical Home Become An Effective Working Model
- Educational programs are needed for all stakeholders.
- The PCP must be well informed about:
  - The components of EHDI systems
  - Local, regional and national resources available
  - Standards of care for each component of the system
  - Skills to facilitate access to care for all families
  - Team with parents to advocate for needed services

EHDI and the Medical Home

How Can These Linkages Be Made Seamless?
- Development of Assessment Protocols
- Ongoing Research
The Medical Workup

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

Causes of Permanent Hearing Loss in 100 Infants

- 50% Environmental
- 50% Genetic
  - 30% syndromes (>300)
  - 20% >75 genes identified
  - ½ are GJB2 - Connexin 26

Genetic Causes

- Single gene
- Gene + environment
- Gene + gene

Connexin 26
Mitochondrial + ototoxic
Gene + other gene

Most Common Environmental Causes

- CMV
- Meningitis
- Otoxic meds
- Rubella
- Asphyxia
- Prematurity

Early Intervention Birth to 3

- Services in the US are good for children with severe to profound hearing loss
- Services are weak for children with unilateral, mild, and moderate hearing loss.

Rhode Island - 7 Birthing Hospitals

- 1 Tertiary Care Center (WRI) - 60 bed NICU
- Convalescent nursery
- 3 well baby nurseries
- Birth rate of 8,000, 9th largest birthing hospital in US
- 2 level 2 nurseries, 4 level 1 nurseries
- 14,000 births
Hearing Advisory Committee - est. 1993
- Multidisciplinary - monitors all program components
- Identified strengths and weaknesses of system
- Reports on a quarterly basis to the DOH
- Makes recommendations to the Hearing Follow-up Committee

Hearing Follow-up Committee - est. 1994
- Multiple constituency working committee
- Mission: provide a forum from which to address systems issues related to the delivery of services including evaluation, intervention, habilitation, equipment, access, barriers, and costs

Hearing Follow-up Committee Achievements
- Developed a Parent Resource Manual
- Developed a Parent Guide on Amplification
- Dissemination of information on HL to pediatricians
- Participation in Strategic Planning for EI
- Formal presentations to managed care insurers to obtain reimbursements for amplification

Hearing Follow-up Committee - Example
- Area of need identified: mental health resources for deaf and HOH and their families
- Conducted a survey of RI Mental Health providers
- Invited those interested to an in-service screening, dx, and intervention
- Developed a resource list of providers interested in working with deaf or HOH or their families

Benchmarking a Hearing Screen Program
- Quality indicators:
  - >69% screened
  - 1st stage (TECAE/ABR) fail rate ≤4%
  - Rescreen return rate >90%
  - Dx procedures on referrals >90%
  - Referrals for intervention services <6m

“KIDSNET”
A Data Management System for Universal Screening Vital Records
- Level 1 - Metabolic - Hearing
- Lead - Immunizations - WIC
- VNA - EI - Primary Provider
Medical Home "First Connections" Task Force

Dr. E. Kutter White RNMPH

- Multidisciplinary – pediatricians, audiologists, parents, health department
- 
- Developing manuals and a website for EHDI system protocols and resources called “Navigating the System”

RIHAP Quality Improvement

Rhode Island State-Wide Initial Screen Fail Rates: Impact of Hearing Screen Protocol

Mean Age of Identification (Months)

Example of Medical Home EHDI System Evolving
Hailey D.O.B. 6-21-93
Full term, well baby nursery
Age RT LT
1 day OAE F T
3 days ABR 85 dB 85 dB
Fam Hx - grandfather - Meniere’s; father - lifelong mild HL
Pediatrician referred to pediatric neurology
2m neurologist report - hearing N in at least 1 ear, FU not needed

Hailey Follow-up
6 m Pedi calls RINAP, requests stat testing on HL, says family not convinced Hailey is hearing
Given referral to an Audiologist
7 m Diagnostic ABR bilat sev prof
9 m 2nd diagnostic ABR bilat sev prof → E1
11 m Aids
24m Cochlear implant

Hailey’s brother Jake 1-4-97
Full term - sister deaf, paternal cousin deaf, maternal uncle with cleft palate & lip
Age RT LT
1 da OAE F F
1 da & 3 days OAE 85 dB 85 dB Refer E1
3 wks ABR 100 dB 100 dB → molds poured for hearing aids, referred to early intervention

Conclusions
• Successful implementation of EHDI systems within the Medical Home Framework will allow us to:
  • celebrate the resiliency and the
  • successes of children with congenital HL
WHAT ARE THE ADVANTAGES AND DISADVANTAGES FOR EARLY DETECTION AND INTERVENTION OF CHILDREN WITH HEARING LOSS

Dr. Christine Yoshinaga-Itano, Speech, Language and Hearing Sciences Department, University of Colorado, Boulder, Colorado, United States

Thursday, January 23, 2003, 10:30 AM to 12:00 PM

This session will present research data that led to the support for establishing early hearing detection and intervention (EHDI) programs begun through universal newborn hearing screening programs (UNHS) in the United States. Evidence that there are sensitive periods in the development of children with significant hearing loss and that the first six months of life are critical to age-appropriate language development will be presented. Both cross-sectional and longitudinal information will be presented. Once a language path, rate of language growth, is established, the path is resistant to change. The sensitive periods for different aspects of language development differ. The development of vocabulary, phonology, syntax and morphology appear to have different sensitive periods. Vocabulary development is very sensitive to early identification and the rate of growth is established early in life. Vocabulary can serve as a gateway to other aspects of language development. Parental involvement and other parent variables can impact changes in the rate of development. Language, speech, and social-emotional developmental outcomes as a result of early-identification and earlier intervention of children with hearing loss will be discussed. Universal newborn hearing screening programs result in earlier identification (an average of 2 to 3 months of age), earlier initiation of intervention (within 2 months of identification of the hearing loss), and better developmental outcomes. UNHS/EHDI programs have identified new populations of children for which intervention protocols need to be developed: unilateral hearing loss, auditory neuropathy, and infant/toddler cochlear implant candidates. Some children with unilateral hearing loss progress to bilateral hearing loss. About 34% of the unilaterals followed longitudinally have significant language delays. These unilateral hearing losses were identified but did not have treatment or use of amplification.
What are the advantages of detection and intervention for children with congenital hearing loss?
Christine Yoshinaga-Itano
University of Colorado, Boulder
Marion Downs National Center

Efficacy of Universal Newborn Hearing Screening
• Do children with hearing loss, born in hospitals that screen, have a higher probability of earlier identification and better developmental outcomes?

Opportunity to be screened
• Child must be born in a hospital after the establishment of universal newborn hearing screening.
• Child is in the Screen category whether or not the child was actually screened, i.e. the screening program had no testing on weekends and the child was born and discharged over the weekend

Screen category
• Child was in the screen category if child was referred from UNHS but did not return for follow-through
• Child was in the screen category if child was referred from second level screen to diagnostic audiology but family did not return for audiological testing
• Child was diagnosed with a hearing loss but did not enroll in intervention until much later

No Screen category
• Children born in hospitals prior to establishment of universal newborn hearing screening programs in Colorado
• Children born in hospitals after screening was begun but in non-participating hospitals

Participants
• N=25 Screened
• No secondary disabilities
• Hearing loss from mild to profound
• Cognitive quotients from 75 to 132 (Mean=97.1)
• Age: 9 to 59 months (M=29.9)
Participants
- Matched pair design
- CA= 9 to 61 months (Mean=30.5)
- CQ=98.9
- Matched by gender, ethnicity, presence of additional disabilities, educational level of the caretaker, and mode of communication

Screen vs. No Screen
- 25% identified by 2.5 weeks, 50% by 5 weeks and 75% by 3 months – Screen
- Four children in the “screen” group were identified after 8 months of age
- No Screen group: 25th %ile identified by 13.5 months, median by 23 months and 75th %ile by 30 months

Screen vs. No Screen
- Children in the Screen group had higher expressive and receptive language
- Sig (p<.001) Expressive Language, Receptive Language and Total Language
- Screen group: 56% Normal >80, 20% LQ=70-79, 24% LQ< 70
- No Screen group: 24% LQ>80, 8% LQ=70-79, 68% LQ<70

Non-verbal cognitive and language development
- Children who are early-identified whether born in hospitals that screen or those that did not had non-verbal cognitive development commensurate with their verbal language development
- Later-identified children had large discrepancies, 30+ points with non-verbal cognitive development higher than verbal language development
Screen vs. No Screen

- Vocabulary development
- Children in the Screen group had significantly higher vocabulary levels
- A child at the 75th percentile of the No Screen group had fewer words than a child at the 25th percentile of the Screen Group
- Significant differences (p<.001)

Speech results: Screen/No screen

- Children in the screen group had significantly more vowels (p<.001)
- Children in the screen group had significantly more consonants (p<.001)
- Children in the screen group had significantly more initial and final blends. No children in the “no screen group had blends

Mean Length of Utterance

- Children in the Screen group had longer sentence structure than children in the Non-screen group.
**Speech Intelligibility: Screen vs. no screen**
- 25th %ile of both groups: always or almost always unintelligible
- 50th %ile of “no screen” group was still always or almost always unintelligible
- 50th %ile of “screen” group was speech was hard to understand
- 75th %ile of no-screen group was speech was hard to understand
- 75th %ile of “screen” group was always or almost always understandable

**Parental Harm**
- False Positives
  - Abdala de Uzcategui and Yoshinaga-Itano, 1996
    - Few families report negative responses
    - < 8 of 100 families
    - Majority report benefits of screening
    - Increased attention to speech, language development

**Parental Harm**
- Tharpe, 1999
  - Study compared false positive emotional response to control group of true positives
  - No significant difference in percent of families reporting negative responses

**Parental Harm**
- True negatives
  - Early-identified population have higher language levels which are highly related to:
    - Better Emotional availability (Figgie-Siegel, Pressman, Yoshinaga-Itano, 1999)
    - Lower Parental stress (Figgie-Siegel & Yoshinaga-Itano, 1999)
    - Lower Parental depression (Siegel, 1999)
    - Lower Parental Needs/Hassles (Siegel, 1999)
    - Faster Resolution of Grief (Siegel, 1999)

**Parental Stress**
- Clemens, Davis & Bailey (Pediatrics-2000), Watkin, Baldwin, Dixon, Beckman (1996-British Journal of Audiology)
  - Barringer & Mauk (1997-Audiology Today)
  - Indicate that 3-13% of false positive group have residual negative impact
  - Approximately 10-13% of the population at large have clinical levels of stress
What is the role of the characteristics of intervention programs?
- There are three cohorts of children identified early (within the first six months) or with initiation of intervention within the first 11-12 months of life.
  - Colorado (N=72 to 150) dependent upon study
  - Nebraska (N=25)
  - Washington (N=5)

All three programs are home-intervention programs with similar intervention philosophies and whose intervention providers have professional training in education for children who are deaf or hard of hearing.

Can the results from the study be generalized
- Regression analyses: Accounting for variance within the sample studied.
  - Predictor variables change as characteristics of the sample changes.
  - Predictors for samples of early-identified children only differ for predictors of later-identified children.
    - The proportion of early to late-identified children alters significant predictor variables
  - Accounting for critical variables: degree of hearing loss, age of identification/intervention initiation, age at testing, socio-economic status.

- Time from diagnosis of hearing loss to follow-through may differ in other states or countries.
- Type of follow-through may differ dramatically and may result in differences.
- Colorado has a Co-Hear system of referral of identified coordinators who do first contact with the families. They are highly knowledgeable about hearing loss and infant diagnostic evaluations, intervention and counseling strategies.

Random assignment does not guarantee generalization
- Random assignment to treatment vs. no treatment will not occur as a result of federal legislation mandating the initiation of intervention services through referral within 48 hours of identification of the disability.
- Random assignment is designed to insure a normal distribution but won’t insure this outcome when starting with skewed distributions.

Does universal newborn hearing screening (UNHS)/early hearing detection and intervention (EHDI) result in better outcomes than early identification through other methods?
- The question may be pertinent to health authorities because of financial considerations.
- Children identified through the high-risk registry within the first six months of life show the same language advantage (Apuzzo & Yoshinaga-Itano, 1998).
- A more pertinent question: Not will children born in screening hospitals do better, but will children born in non-screening hospitals be identified.
Is there a critical age? Is there a sensitive period of language development?

- In the first three years of life, the age of identification (within first 6 months) and earlier initiation of intervention (before 12 months) are associated with significantly better language development (Yoshinaga-Itano, Sedey, Coulter & Mehli, 1998, Moeller, 2000).

- When a broad age range is used, i.e., 6 months to 72 months, age of initiation of intervention/age of identification play a significant role in predicting language outcome (Pipp-Siegel, Sedey, Mayne & Yoshinaga-Itano, 2003).

Sensitive Periods of Development

- Identification of hearing loss by six months of age was found to be a sensitive period for maintaining language development within the normal range in the first three years of life (Yoshinaga-Itano, Sedey, Coulter & Mehli, 1998 Pediatrics).

Later Identification of Hearing Loss results in significant delays in development

- Children with hearing loss only who were later-identified had language development similar to children with multiple disabilities who had hearing loss which was early-identified.

Early Identification Effect

- The language impact of early identification of hearing loss is not an auditory/speech phenomenon only.
- Among EID children, there were no significant differences by degree of hearing loss or by mode of communication. Children without residual hearing or amplification benefit who used sign language as the primary mode of communication had similar language outcomes when early-identified.
The EID/Early RX effect was present
- For both genders
- For children whose mothers had less than a high school education and those with greater than a high school education.
- For children from families on Medicaid and those who did not quality for Medicaid.

The EID/ERX effect
- Children from ethnic minority families and from ethnic majority families
- Children who used sign language
- Children who used speech
- Children with hearing loss only
- Children with multiple disabilities
- Children at all age levels (birth – 3 yrs)

Sensitive Period: Speech Development
- The sensitive period for the development of speech is longer than for the development of language.
- Children with minimal or no speech development in the first three years of life could develop intelligible speech.
Sensitive Period differs according to aspect of language
- The sensitive period differs according to the aspect of language
  - Semantics (lexicon)
  - Syntax/morphology
  - Phonology
- Both phonology and syntax/morphology have longer sensitive periods than the development of vocabulary
- Vocabulary development can serve as a gateway to support development of phonology and syntax/morphology
- Rate of vocabulary development appears to be the most resistant to change

Predictors differ according to the characteristics of the population
- Early identification/earlier intervention
- Degree of hearing loss
- Cognitive non-verbal abilities
- Maternal level of education
- Presence of additional disabilities
- Gender
- Age of the child

Predictors differ according to the language variable
- Lexical knowledge
- Syntax and morphology
- Pragmatic language
- Phonology

Predictors of successful language outcomes at 15 months-2/3 EID
- Vocabulary-Lexical knowledge: MacArthur Communicative Development Inventory
  - Symbolic Play: 73% predicted by NV cognitive status
  - MACI Comprehension/Conceptual: 88.7% predicted by NV cognitive
  - MCDI Expressive Language: 56.4% - NV cognitive, parent (min) unit
  - Mac CDI Words produced: 44.3%, maternal education, child gesture/minute
  - One Word: Produced in language sample: 55.4% - maternal education, child gesture/minute, parent gesture

Predictors of successful language outcome at 21 months-2/3 EID
- Lexical knowledge: MacArthur Communicative Development Inventory
  - 70% of variance
  - Situation Comprehension
  - Parental imitation of the Child

Predictors of successful language outcome at 27 months: 2/3 EID
- Lexical knowledge: MacArthur Communicative Development Inventory
  - 42%
  - Additional Disabilities
  - Parental use of pragmatics no longer significantly predicts

- General language: Minnesota MCDI Expressive Language
  - 65% of variance
  - Situation Comprehension
  - Parental use of pragmatics no longer significantly predicts
Predictors of successful language outcome at 27 mo.: ½ EID, ½ LID

- Age of ID is a significant predictor at 27 months
- Lexical knowledge (CA= 24-36 mo.)
  - 57% variance
  - CA & Age of ID (24%)
  - Situation Comprehension (CQ) (30%)
  - Additional medical issues (5%)
- General language (CA=24-30 mo.)
  - 57.2%
  - Age of ID
  - Situation Comprehension (CQ)

Predictors of successful language outcome at 36 months

- General language: Minnesota CDI
  - (N=144) 59% variance (43% EID) (Yoshinaga-Itano & Gavin)
  - Situation Comprehension
  - Degree of hearing loss
  - 43% of variance (Stevens, 2002) (44% EID)
  - N=55 (longitudinal, 36 & 48 months)
  - Age of ID, Additional Disability, Degree of hearing loss

36 months (Yoshinaga-Itano & Gavin, 2003)

- At 36 months, a significant amount of variance can be accounted for when groups are separated by age of identification
- General language: MCDI
  - (N=85) 70% variance (EID only) (Yoshinaga-Itano & Gavin)
  - Degree of hearing loss
  - Additional Disabilities
  - (N=50) 46% variance (LID)
  - Degree of hearing loss
  - Situation Comprehension

Predictors of successful language outcome at 48 months

- Age of ID is highly significantly as a predictor
- General language: Minnesota CDI
  - Yoshinaga-Itano & Gavin, 2003 (N=107)
  - Cross-sectional
  - 56% variance
  - Age of ID
  - Situation Comprehension

Longitudinal: 36 & 48 months

- Stevens, 2002 (N=55) Longitudinal
- 41% of variance: presence of additional disabilities
- 74% of variance if Language at 36 mo. added
- LQ at 36 mo. (70% of 74%) – all other variables added 4%
- Once the language pathway is established, rate of language growth is resistant to change

Age of identification/age at intervention

- 27 months – age of ID highly significant
- 36 months – age of ID non-significant
- 48 months – age of ID highly significant
- Instrument change – item analysis
- Emphasis from lexicon to syntax at 36 months
- English Syntax development is highly related to phonology development and hearing level possibly more than early-identification of hearing loss
Predictors of successful language outcome at 60 months

- Moeller, 2000
- N=112
- 25 Rx begun before 12 mo.
- 87 Rx begun after 12 mo.
- Reynell Expressive Language
- 55% of variance accounted for by:
  - Parental involvement
  - Age of intervention initiation
  - Performance intelligence

Longitudinal: 48 mo. & 60 mo.

- Once the language pathway is set, it is resistant to change. (38% profound, 55% severe, 90% early-identified)
- 87% of the variance accounted for by:
  - Language Quotient at 48 months
  - Education of the mother

Unilateral Hearing Loss

- Unilateral Hearing Loss
- Earlier Cochlear Implantation
- Auditory Neuropathy

Unilateral to Bilateral Loss

- 30 children initially identified with unilateral loss
  - 2 (7%) progressed to bilateral within first year of life
  - 2 (7%) later diagnosed with bilateral losses that apparently were present from birth
    - One mild (30dB) in poorer ear
    - One moderate low frequency loss with normal high frequency hearing

Types of Hearing Loss

- Progression of Hearing Loss
- Developmental Outcomes
- Implications for follow-through

- Sedey, A., Carpenter, K., Stredler-Brown, A.
Participants
- 26 children with unilateral hearing loss
- N=24 (CO), N=1 (NM), N=1 (VA)
- N=14 (54%)-Male, N=12 (46%)-Female
- 69% Caucasian, 31% (Minority)

Participants
- Hearing Loss only: 85%
- Additional disability: 15%
- Median level of education – 16 yrs.
- Median income: $60,000
- 65% oral, 27% some sign, 8% frequent
- 89% screened, 8% not screened or unknown
- 92% ID <6 mo., 4%-14 mo., 4%-18 mo.

Participants
- 88% - congenital, 8% - acquired, 4% - unknown
- Etiology: 77% unknown, 12% genetic, 8% Waardenburg, 4% meningitis, 4% seizures
- Malformation of the ear structures:
  - 69% none, 23% atresia, 8% Mondini
- 54% RE, 46% LE

Degree of Loss
<table>
<thead>
<tr>
<th>Level</th>
<th>n</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mild</td>
<td>2</td>
<td>8</td>
</tr>
<tr>
<td>Moderate</td>
<td>6</td>
<td>24</td>
</tr>
<tr>
<td>Moderate-severe</td>
<td>6</td>
<td>24</td>
</tr>
<tr>
<td>Severe</td>
<td>4</td>
<td>16</td>
</tr>
<tr>
<td>Severe or profound</td>
<td>7</td>
<td>28</td>
</tr>
</tbody>
</table>

N = 26

Language Ability
- Assessments:
  - Minnesota Child Development Inventory
  - MacArthur Communicative Development Inventories
  - Spontaneous language sample

Minneapolis Inventory
- Participant Description:
  - 18 children
  - No additional disabilities
  - Selected oldest age available
  - Chronological age:
    - Range = 7 to 69 months
    - Mean = 25 months
**Minnesota Inventory**

<table>
<thead>
<tr>
<th>Expressive</th>
<th>n</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td>Borderline (70 – 79)</td>
<td>3</td>
<td>17</td>
</tr>
<tr>
<td>Average (80+)</td>
<td>15</td>
<td>83</td>
</tr>
<tr>
<td>Receptive</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Below average (&lt; 70)</td>
<td>1</td>
<td>6</td>
</tr>
<tr>
<td>Borderline (70 – 79)</td>
<td>3</td>
<td>17</td>
</tr>
<tr>
<td>Average (80+)</td>
<td>14</td>
<td>78</td>
</tr>
</tbody>
</table>

**MacArthur Inventory: Expressive**

- Participant Description:
  - 12 children
  - No additional disabilities
  - Selected closest assessment to 21 months
  - Chronological age:
    - Range = 14 to 23 months
    - Mean = 21 months

**MacArthur Inventory: Receptive**

- Participant Description:
  - 11 children
  - No additional disabilities
  - Selected all children who were the appropriate age for the test
  - Chronological age:
    - Range = 12 to 16 months
    - Mean = 14.5 months

**MacArthur Inventories**

<table>
<thead>
<tr>
<th>Expressive</th>
<th>n</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td>&lt; 10th percentile</td>
<td>2</td>
<td>17</td>
</tr>
<tr>
<td>&gt; 10th percentile</td>
<td>10</td>
<td>83</td>
</tr>
<tr>
<td>Receptive</td>
<td></td>
<td></td>
</tr>
<tr>
<td>&lt; 10th percentile</td>
<td>2</td>
<td>17</td>
</tr>
<tr>
<td>&gt; 10th percentile</td>
<td>9</td>
<td>82</td>
</tr>
</tbody>
</table>

**MacArthur Inventories**

- Median Percentile
  - Expressive = 27th
  - Receptive = 15th

**Spontaneous Language Sample**

- Participant Description:
  - 15 children
  - No additional disabilities
  - Selected oldest age available for each child
  - Chronological age:
    - Range = 15 to 62 months
    - Mean = 29 months
**Spontaneous Language Sample**
- **Procedure**
  - 25-minute parent-child interaction
  - Videotaped at home
  - All child utterances transcribed
  - Mean length of utterance in morphemes (MLU) calculated

**Mean Length of Utterance (MLU)**
- 10 (67%) children within age expectations
- 5 (33%) children below age expectations

**Summary of Language Results**
- 15 children examined across measures and time
  - Considered assessments after 12 mos. of age
  - No additional disabilities
  - Number of children with language delays
    - Delayed = 4 (27%)
    - Borderline = 1 (7%)

**Profile of 4 Children with Delays**
- Caucasian
- Identified by 2 months of age
- Congenital
- Etiology unknown
- 3 used oral communication only, 1 used oral plus some signs
- Parental education 16 years +
- Annual income > $80,000

**Profile of Children with Delays**
- No outer or middle ear malformation
- Affected ear: 50% right, 50% left
- Degree of loss
  - All “severe or profound” (i.e., no response on ABR) or profound

**Needs Assessment**
- 2000-01 summary of students with hearing loss in CO
  - Incidence in school-age population = 1.5/1000
  - 36% of children identified with UHL in the schools are on IEPs
  - Research from 1980s suggests academic failure – 1/3
**Needs Assessment**

- Implications of current study
  - Is amplification helpful? (1:26 used amplification)
  - Audiological management:
    - Possible progression in SN population
    - “Missed” identification of bilateral hearing loss
    - Fluctuating loss in good ear due to middle ear fluid
  - Consistent or borderline language delay evidenced in 34% of the children

**RECOMMENDATIONS**

- Click ABRs, OAEs may pass mild and borderline mild hearing loss on children with asymmetrical bilateral losses. Tone ABRs for both ears in diagnostic evaluations
- Frequent diagnostic evaluations – every 3 months
- Verify with behavioral thresholds, both ears – looking for asymmetrical, borderline mild, mild or rising losses in the contralateral passed ear

**UNHS and lower age of implantation**

- Surgical considerations for early implantation – Sinninger, 2001
- Early implantation and developmental outcomes
- Relationship between speech and language outcomes

**Children with cochlear implants**

- Universal newborn hearing screening programs (UNHS)/early hearing detection and intervention programs (EHDI)

- At what age should children who are early-identified.
- Should children have cochlear implants at 6 months of age?

**Relationship between speech and language**

- Ettmer, Young, Grohne, Mellon, Johnson, Corbett & Saindon (2002) – followed two children every month for 12 months
- Case 1 - CI 10 mo. No language scores available
- Case 1: 12 mo. Post CI 2 consonants, 9 vowels (CA=22 mo.)
Two Colorado Case Studies: Case Study 1
- Case 1: CI 22 mo. (CA 15 mo. pre-implant, 10 vowels, 7 consonants)
- Case 1: 6 mo. Post CI (CA=28 mo.), 15 vowels, 12 consonants
- Case 1: 14 mo. Post CI (CA=36 mo.) 15 vowels, 21 consonants/consonant blends
- Vocabulary used as gateway to speech
- Intelligible speaker, Vocabulary pre and post implant at low average level, Speech predominant pre-implant, Speech predominant post-implant

Colorado Case Study 2
- Case 2: CI at 30 months
- Case 2: Pre CI at 27 mo. 7 vowels, 5 consonants
- Case 2: 7 mo. Post CI (CA=34 mo.) 10 vowels, 11 consonants
- Case 2: 14 mo. Post CI (CA=48), 14 vowels, 22 consonants
- Intelligible speaker, Vocabulary pre and post implant at low average level
- Vocabulary used as gateway to speech
- Signs pre-implant, Speech predominant post-implant

Geers, 2002
- Implant characteristics:
  - Duration of Speak
  - # of active electrodes
  - Dynamic range
  - Loudness Growth
- Accounted for 28% of the variance in speech perception, 22% in speech production, 23% spoken language, 21% total language, 17% reading

Geers, 2002
- Type of class and Communication mode accounted for
- 16% of the variance in speech perception, 18% in speech production, 10% in spoken language, NS total language, 7% reading
- Total Variance accounted for: 54%

Method of communication and syntax development
- Geers (2002)
- Receptive and expressive English syntax development is significantly better for children taught OC as compared to TC
- 47% were at age level or within 2 years

Age of implantation did not predict outcome

- N= 147
- Used implant for 6 months to 10 years
- 70 children implanted by 5 years showed no difference between OC and TC groups in consonant production difference
- Greater vocabulary was reported for the TC group
- Not many followed had implant for over 5 years

Does age impact the role of method of communication after cochlear implantation?

- Children implanted before 18 months were able to successfully transition from manual to oral communication
- 50% of children implanted between 18-30 months successfully transition
- After 30 months, transition probability is significantly reduced

Method of communication and syntax development

- Geers (2002)
- Receptive and expressive English syntax development is significantly better for children taught OC as compared to TC
- 47% were at age level or within 2 years

IMPLICATIONS

- No evidence yet that earlier implantation (first year of life) results in better outcomes
- For early implantation, monitor language development closely; higher language levels appear related to faster auditory/speech development
- Implantation prior to 18 months results in ease of transition from sign to speech
- Mode of communication effects on vocabulary development differs from effect on syntax development

Auditory Neuropathy

- Children diagnosed with auditory neuropathy
- Well-baby vs. NICU

Newborns screened with AABR first (excluding OAE first) – 1999-2001 CO Births: 194,551

- Newborns screened, AABR first only: 144,318 (74 % of population)
- Newborns in NICU screened (estimate): 14,432 (10 % estimate at AABR hospitals)
Prevalence confirmed hearing loss

- Confirmed hearing loss (CHL): 176 (1.22 per thousand screened)
- CHL, bilateral only: 145 (1.00 per thousand screened)
- CHL, bilateral only, from NICU: 39 (27% of all bilateral CHL)

Prevalence of auditory neuropathy in Colorado 1999-2001

- Auditory neuropathy (bilateral) from NICU: 8 of 39 (21% of all CHL detected in the NICU)
- Auditory neuropathy from well baby nursery: 0 of 106 (0% of all CHL detected in the WBN)

Auditory Neuropathy WBN 1993-2001

Primary screening method: AABR

Confirmed congenital hearing loss: 421
Confirmed bilateral hearing loss: 304
Bilateral hearing loss from well baby nursery (estimated): 222

Auditory Neuropathy WBN (1993-2001)

- Confirmed auditory neuropathy from well baby nursery: 0
- Confirmed auditory neuropathy, well baby, never screened: 1 of 528,874 births

Developmental Profile Characteristics: AN

- Poor and inconsistent auditory skills development
- Speech quality generally poor
- Inconsistency in speech production, wide variety of use of vowels and consonants, but not developmental increases
- Strong visual learners

Developmental Profile Characteristics

- Some auditory neuropathy infants have recovered from absent/abnormal ABRs to normal ABR - typically premature, hyperbilirubinemia
- Wide variation of behavioral audiograms
- Some children with profound HL (behavioral thresholds) and AN have been successful CI users – must be certain that thresholds are not improving
Website

- http://www.colorado.edu/slhs/mdnc/research/publications.html
- Abstracts of published research and figures
- Some complete articles of non-copyrighted articles

Developmental Norms

- http://www.colorado.edu/slhs/mdnc/research.html
- Minnesota Child Development Inventory
- MacArthur Words and Gestures
- MacArthur Words and Sentences
- MacArthur III
- Play Assessment Questionnaire
- Speech(# vowels, consonants, blends, speech intelligibility)
- Expressive One Word Picture Vocabulary

- Norms are provided by:
  - age at testing
  - Degree of hearing loss (if significant)
  - Language level (if significant)
  - Age of identification (if significant)
  - With hearing loss only and separately for children with additional disabilities
  - By non-verbal cognitive levels
COST EVALUATION OF EARLY HEARING DETECTION AND INTERVENTION (EHDI) SYSTEMS

Dr. Betty Vohr, Department of Pediatrics, Women and Infants Hospital, Brown University, Providence, Rhode Island, United States

Thursday, January 23, 1:30 to 3:00 PM

In the current economy program cost containment is a high priority. Hearing screening, audiology services, otolaryngology services, genetic counselling and early intervention for infants identified early with hearing loss all have associated costs for the hospital, the family and the service provider. These costs, however, must be viewed with the knowledge that undetected hearing loss significantly affects language, speech, cognitive and behavioural development resulting in higher family and societal costs.

Factors affecting newborn hearing screening costs including hospital costs, operating expenses, screening skills, follow-up costs and number of babies screened. Studies evaluating the costs of TEOAE (transient evoked otoacoustic emissions), AABR (automated auditory brainstem response) and a two-step protocol using TEOAE and AABR will be presented. Post discharge costs will be evaluated.
Cost Evaluation of Early Hearing Detection and Intervention (EHDI) Systems

Betty Vohr, M.D., FAAP
Brown Medical School

Justification for Newborn Universal Screening (AAP 1999)

- An easy-to-use test / high sensitivity & specificity
- Condition not otherwise detectable clinically
- Interventions available to remediate the condition
- Early screening & detection results in improved outcome
- Screening process is in an acceptable cost effective range.

Incidence


How Many Infants Have Permanent HL at Birth?

<table>
<thead>
<tr>
<th>Site</th>
<th>Rate</th>
</tr>
</thead>
<tbody>
<tr>
<td>Well Baby Nursery</td>
<td>1 per 1000</td>
</tr>
<tr>
<td>NICU</td>
<td>10 per 1000</td>
</tr>
<tr>
<td>Total population</td>
<td>2-3 per 1000</td>
</tr>
<tr>
<td># infants ident annually US</td>
<td>8,000-12,000</td>
</tr>
<tr>
<td>Average career pediatrician</td>
<td>12 patients</td>
</tr>
</tbody>
</table>

HL is the most common congenital abnormality

Current Screen Methods Available

- Transient Evoked Otoacoustic Emissions TEOAE
- Automated Auditory Brainstem Response AABR
- Standard Auditory Brainstem Response ABR
- Two Step TEOAE + AABR
Impact of Hearing Loss

Undetected congenital hearing loss significantly affects language, speech, cognitive and behavioral development resulting in high family and societal costs.

Severe to Profound Hearing Impairment in the US

- Population estimate 464,000 to 738,000
- Adults with S/P HL (prior to age of Early Identification) are more likely to be
  - less educated
  - publicly insured
  - have lower family income
  - be unemployed

Why is Early Identification of Hearing Loss so Important?

- Not just because Undetected hearing loss has serious negative consequences.
- But, because there are significant benefits associated with early identification of hearing loss.

Costs

- In the current economy, everyone (hospitals, providers, insurers, families) are concerned about containing costs.
EHDI System Costs

- Hearing Screening
- Audiology Diagnostic
- Medical and Otolaryngology workup
- Genetics Consultation
- Hearing Aids/FM systems/Cochlear Implants
- Speech/Language Therapy
- Early Intervention and Special Education

Who pays the Costs of a Newborn Hearing Screening And Detection System (EHDI)?

Family  
Hospital  
Society  
What are the factors which affect cost?

Factors Affecting Screen Costs for the Hospital

- Personnel: volunteers, techs, audiologists
- Equipment, supplies
- Protocol (rescreen)
- # of infants, high or low risk
- Fail rate
- Tracking - Information System

Newborn Screening- Operating Expenses

- Costs which continue daily
- Personnel - vary by geographic area, may be volunteers, students, nurses, techs, audiologists
- Disposables - ear tips, couplers $1-2 per infant
  - electrodes for ABR $4-8 per infant

Costs of Screen Personnel

- Volunteers training/supervision costs - short term
- Technicians intermediate cost - long term
- Audiologists most expensive, minimal training

Newborn Screening – Capital Expenses

- Equipment: range $50,000 to $250,000
- A one-time expense
- Capital Equipment is Amortized
  - $25,000 - 10,000 babies screened = $2.50 per baby
  - $25,000 - 1000 babies screened = $25 per baby
- Therefore smaller hospitals have greater capital expenses.
Newborn Screen Protocols Affect Costs

<table>
<thead>
<tr>
<th>Method</th>
<th>In Hospital</th>
<th>Post Discharge</th>
</tr>
</thead>
<tbody>
<tr>
<td>TEOAE</td>
<td>1 Rescreen</td>
<td>1 Rescreen</td>
</tr>
<tr>
<td>AABR</td>
<td>1 Rescreen</td>
<td>1 Rescreen</td>
</tr>
<tr>
<td>Two Step</td>
<td>1 Rescreen</td>
<td>1 Rescreen</td>
</tr>
<tr>
<td>ABR</td>
<td>1 Rescreen</td>
<td>1 Rescreen</td>
</tr>
</tbody>
</table>

The Screen Population Affects Costs

- High risk NICU infants vs. well babies
  - more difficult to test, older at time of testing
  - higher fail rate
- Small birthing hospital vs. large census hospital
  - less experience screening
  - higher fail rate secondary to inexperience
  - higher cost per infant screened

Fail Rates Affect Costs

- High hospital fail rates result in higher costs for tracking, more re-screenings after discharge and more referrals for diagnostics.
- Fail rates tend to be higher for OAE screening than AABR secondary to early transient, external and middle ear conditions which impact on the reverse transmission OAE signal.
- One approach is a second screen prior to discharge

Rhode Island State-Wide Initial Screen Fail Rates: Impact of Hearing Screen Protocol

Data Management System Requirements

- Electronically store core demographic and hearing assessment data elements
- Modifiable data model
- Multi-user ability to access & retrieve data
- Protect and ensure security of confidential health care information
- Enhanced tracking and follow up capability**

Data Management System Requirements

- Exchange demographic and hearing assessment data via a variety of mechanisms.
- Capability to produce set of standard individual and group reports and letters and generate ad-hoc reports.
- Promote accuracy and completeness of hearing assessment data via data quality tools and merging/de-duplication tools.
- **Maintainable, upgradeable - technical support**
Data Management System
- Saves money by
- Facilitating quality management
- benchmarking

Newborn Screen Data Management Costs
- Paper Trail - least expensive - least effective
data management issues
- Data base - developed at site - individualized
  requires designer
  start up time
- Purchased system - single fee, fee per screen, or annual fee
  protested at other sites.

Benchmarking a Hearing Screen Program
- Quality indicators
- >90% screened
- 1st stage (TEOAE/AABR) fail rate
  ≤4%
- Rescreen return rate
  >90%
- Diagnostic procedures on referrals
  >90%
- Referrals for intervention services
  <6m
  100%

Who pays for newborn screening?
- Private insurance
- Public insurance
- The family
- The hospital absorbs the cost

Background
Currently 2 of the most commonly used validated
and proven methods for newborn hearing screening
in the United States are:
  - Transient Evoked Otoacoustic Emissions (TEOAE)
  - Automated Auditory Brainstem Response (AABR)

Purpose of Cost Study
The purpose of this study was to investigate the
hospital costs, post discharge costs and cost per
child identified with hearing loss of 3 newborn
hearing screen protocols
  TEOAE
  AABR
  Two step protocol TEOAE

Vohr et al., J Pediatr., 2001
Hypothesis

- **Primary**
  It was hypothesized that costs would be similar for the 3 protocols.

- **Secondary**
  Environmental factors (24 vs 48 hr discharge and screener type) would affect costs and refer rates.

Sites

- **Two step**
  TEOAE + ABR
  Women & Infants’ Hospital
  University of New Mexico

- **One step TEOAE:**
  Memorial Hospital of RI

- **One step ABR:**
  St. Elizabeth’s Hospital
  Boulder Children’s Hospital

Site Characteristics

<table>
<thead>
<tr>
<th>Protocol</th>
<th>A</th>
<th>B</th>
<th>C</th>
<th>D</th>
<th>E</th>
</tr>
</thead>
<tbody>
<tr>
<td>Stay</td>
<td>48</td>
<td>24</td>
<td>48</td>
<td>48</td>
<td>24</td>
</tr>
<tr>
<td>Screener</td>
<td>tech</td>
<td>student</td>
<td>tech</td>
<td>student</td>
<td>vol</td>
</tr>
<tr>
<td>Annual births</td>
<td>8,034</td>
<td>2,068</td>
<td>694</td>
<td>1,530</td>
<td>1,422</td>
</tr>
</tbody>
</table>

Methods - Two Components

- **Retrospective**
  Hearing screen data were abstracted from records of 12,081 well baby nursery infants from the 5 sites. (>1500 /site).
  1500 infants - average U.S. hospital.

- **Prospective**
  Activity based cost techniques were used. Resource utilization data of at least 100 infants at each site were collected between 7/98 and 9/98 using case report forms.

Retrospective Data Points

- Refer rates for TEOAE
- ABR
- two-step

Prospective Data Points

- Gender
- Screening materials
- Age of screen
- Equipment costs
- C-section delivery
- Personnel used
- # of times screened
Methods

- Economic and clinical outcomes were calculated.
- Post-discharge screening was modeled using a published protocol.


Study Sample Size

<table>
<thead>
<tr>
<th></th>
<th>A</th>
<th>B</th>
<th>C</th>
<th>D</th>
<th>E</th>
</tr>
</thead>
<tbody>
<tr>
<td>2 Step</td>
<td>4,684</td>
<td>1,551</td>
<td>2,777</td>
<td>1,540</td>
<td>1,529</td>
</tr>
<tr>
<td>Retrospective</td>
<td>2 Step TEOAE AABR AABR</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Prospective</td>
<td>346</td>
<td>300</td>
<td>190</td>
<td>150</td>
<td>160</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

Post Discharge Protocol Within 1st 2 months

Rescreen (A) ABR at 70 or 75 dB nHL
30 or 35 dB nHL

(B) Otoacoustic emissions

(C) Discuss results & recommendations with family

Diagnostic referral

Audiologic Diagnostic Components

A. ABR including bone conduction
B. Otoscopic evaluation
C. Acoustic emittance
D. Evoked emissions
E. Behavioral assessment > 6 months
F. Recommendations to the family

Site Specific Refer Rates at discharge

<table>
<thead>
<tr>
<th></th>
<th>A</th>
<th>B</th>
<th>C</th>
<th>D</th>
<th>E</th>
</tr>
</thead>
<tbody>
<tr>
<td>2 Step</td>
<td>48</td>
<td>24</td>
<td>48</td>
<td>48</td>
<td>24</td>
</tr>
<tr>
<td>Stay (hr)</td>
<td>FTT St FTT St Vol</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Pers</td>
<td>3.7%</td>
<td>7.4%</td>
<td>6.5%</td>
<td>1.7%</td>
<td>3.9%</td>
</tr>
</tbody>
</table>

Screen refer rates

TEOAR 2 step AABR

65.9% 47.9% 3.2%

*P < 0.001 vs AABR
Cost Assumptions: Audiologist Time

- Time of program management proportional to # technologies & # people supervised
- Time of FU/scheduling: to refer rates
- Time of training: # technologies, # screeners, & complexity of technology based on 1500 births

Other Assumptions

- All programs had >3 y screening experience.
- Training and start up costs not included.
- 100% of infants referred, return for follow-up.
- Although the AABR screens were done by volunteers, in the base case their time was valued at the same hourly rate as technicians.
- Equipment costs were amortized over 5 yrs.

Pre-discharge Costs/ 1500 infants

<table>
<thead>
<tr>
<th></th>
<th>2-Step</th>
<th>TEOAE</th>
<th>AABR</th>
</tr>
</thead>
<tbody>
<tr>
<td>Variable Screening Costs</td>
<td>$13,343</td>
<td>$8,786</td>
<td>$21,517</td>
</tr>
<tr>
<td>Personnel</td>
<td>13,356</td>
<td>11,331</td>
<td>8,270</td>
</tr>
<tr>
<td>Equipment</td>
<td>8,455</td>
<td>4,506</td>
<td>8,081</td>
</tr>
<tr>
<td>Supplies</td>
<td>102</td>
<td>45</td>
<td>270</td>
</tr>
<tr>
<td>Total Overhead</td>
<td>21,914</td>
<td>15,882</td>
<td>16,820</td>
</tr>
<tr>
<td>Total Pre-discharge Costs</td>
<td>35,247</td>
<td>24,668</td>
<td>38,137</td>
</tr>
<tr>
<td>Total Costs per birth</td>
<td>$23.50</td>
<td>$16.45</td>
<td>$25.42</td>
</tr>
<tr>
<td>Refer Rate at Discharge</td>
<td>4.7%</td>
<td>6.5%</td>
<td>3.2%</td>
</tr>
</tbody>
</table>

Post-discharge Costs/ 1500 infants

<table>
<thead>
<tr>
<th></th>
<th>2-Step</th>
<th>TEOAE</th>
<th>AABR</th>
</tr>
</thead>
<tbody>
<tr>
<td>Follow-up Screen Costs</td>
<td>$10,388</td>
<td>$14,436</td>
<td>$7,140</td>
</tr>
<tr>
<td>Diagnostic Evaluation Costs</td>
<td>3,037</td>
<td>3,937</td>
<td>3,937</td>
</tr>
<tr>
<td>Total Post-discharge Costs</td>
<td>14,925</td>
<td>18,373</td>
<td>11,077</td>
</tr>
<tr>
<td>Total Pre- and Post-discharge Costs</td>
<td>$49,582</td>
<td>$43,041</td>
<td>$47,216</td>
</tr>
<tr>
<td>Total cost per birth</td>
<td>$33.05</td>
<td>$26.69</td>
<td>$32.81</td>
</tr>
<tr>
<td>Cost per identified child</td>
<td>$16,527</td>
<td>$14,347</td>
<td>$16,405</td>
</tr>
</tbody>
</table>

Factors Affecting Refer Rates and Costs

<table>
<thead>
<tr>
<th></th>
<th>2-Step</th>
<th>TEOAE</th>
<th>AABR</th>
</tr>
</thead>
<tbody>
<tr>
<td>PT vs FT personnel</td>
<td>++</td>
<td>+++</td>
<td>+</td>
</tr>
<tr>
<td>48 hrs vs 24 hr discharge</td>
<td>+++</td>
<td>++</td>
<td>+</td>
</tr>
</tbody>
</table>

Number of Times an Infant is Screened

<table>
<thead>
<tr>
<th></th>
<th>2-Step</th>
<th>TEOAE</th>
<th>AABR</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>1.97</td>
<td>1.23</td>
<td>1.15</td>
</tr>
</tbody>
</table>

- Discharge time affects the time available to obtain a definitive result (pass/refer).
- Therefore, 24 h discharge has a negative impact on pass rates for protocols with a # of screens needed.
Conclusions

- Total cost of hospital screen and post discharge screen per infant remains economical at $28 to $33 per infant relative to high costs of undetected loss.

Conclusions

- Refer rates at discharge differed among protocols from a low of 3.3% for AABR to a high of 6.5% for TEOAE.
- Total cost per child identified, however, did not differ among programs.
- Program characteristics including personnel type and length of stay impact on both costs and refer rates.

Other Screening Cost Studies

<table>
<thead>
<tr>
<th>Sample Time</th>
<th>Method</th>
<th>Screen Cost/Infant</th>
</tr>
</thead>
<tbody>
<tr>
<td>4,253 1993</td>
<td>2 Step screen</td>
<td>$26.05 (Mason et al 95)</td>
</tr>
<tr>
<td>10,372 92-97</td>
<td>AABR</td>
<td>$17.00 (Mason et al 98)</td>
</tr>
<tr>
<td>41,796 92-96</td>
<td>OAE, AABR or ABR</td>
<td>$25.00 (Mehl et al 99)</td>
</tr>
</tbody>
</table>

Cost of Identification

<table>
<thead>
<tr>
<th>Screen Method</th>
<th>Cost to Identify Infant</th>
</tr>
</thead>
<tbody>
<tr>
<td>2 Step TEOAE</td>
<td>$13,032 (Mason et al 95)</td>
</tr>
<tr>
<td>AABR</td>
<td>$17,705 (Mason et al 98)</td>
</tr>
<tr>
<td>OAE, AABR or ABR</td>
<td>$9,600 (Mehl et al 99)</td>
</tr>
</tbody>
</table>

Screening Costs per Confirmed Diagnosis

Who pays for amplification and medical evaluation costs?

- Private or Public Insurance
- Early Intervention
- The family
- The Department of Education
- Private Foundations
- Partial reimbursements
North American Conference on Deafness Screening and Intervention in Early Childhood

Who pays for Cochlear Implants?

- Private or Public Insurance
- The family
- The Hospital

Cochlear Implant Cost Coverage

- Medicaid and Medicare fail to cover
  - Aural rehabilitation
  - Surgical costs
- Private Insurance does not cover
  - Hospital costs
- Costs are not covered by Medicaid in 18 states

Societal Costs of Severe to Profound HL in US

- $297,000 per person costs over lifetime
- 21% - special education costs
- 67% - reduced work productivity
- 1 million-pre-lingual asset
- 4.6 billion-lifetime costs for all persons identified in a single year 1998

EHD, the Medical Home, and Costs, Costs, Costs

Baby Girl MB

- Initial Screen – completed at 2 days of age
  - OAE: Refer bilaterally
  - AABR: Refer bilaterally
- Rescreen – completed at 1 month
  - OAE: Refer bilaterally
  - AABR: Refer at 40 dB bilaterally
  - Commercial Insurance paid

Diagnostic ABR

- Completed at 5 weeks of age
  - Right Ear: Moderate SN HL
  - Left Ear: Moderately-severe SN HL
- Commercial insurance: no coverage for hearing aids
- Referred to Shriners Foundation for payment

Canadian Academy of Audiology
Institut national de santé publique du Québec
Ordre des orthophonistes et audiologistes du Québec
Hearing Aid Fitting

- Completed at 2 months of age
- **Shrine provided:**
  - One digital hearing aid. **Parents** purchased the second
  - Earmolds as needed
  - Hearing aid repair, loss and damage coverage
  - Continues services through age 18

FM System

- Commercial insurance and Shrine provided **no** coverage for an FM system
- Phonak, MicroLink FM system was purchased through the **Safety Net Program**
  - Covered cost of system and warranty for 2 years
- **Fit at 2 years, 2 months of age**

Speech Language Therapy

- **Shrine** provided speech language therapy in parents chosen communication method
- **Shrine** SLP left the state when MB was 4 years old
- **School system** provided group speech therapy once per week
- Parents preferred a private SLP therapist
  - Was not a Shrine provider
  - The school system would not cover cost
  - **Parents** currently pay out-of-pocket

Baby Boy T.R.

- Born at 27 weeks GA
  - 905 grams
  - Multiple courses ototoxic medications
  - Mechanical vent 3 days
- Initial screen completed at 9 weeks of age
  - OAE: Refer bilaterally
  - AABR: Refer at 40 dB bilaterally
  - **Public Insurance paid**

Diagnostic ABR

- Completed at 12 weeks of age
- Right ear: Severe SN hearing loss
- Left ear: Severe SN hearing loss
- **State Ritecare (Public) insurance:** covered the cost of binaural hearing aids

Ritecare

- Provided:
  - Hearing aids every 5 years
  - Earmolds every 6 months
  - 1 year loss and damage warranty
  - Repairs as needed
  - Hearing tests
- Parent counseling and ear impressions not covered
FM System

- Fit at one year of age
- T.R.'s current public insurance, Medicaid, covered the cost in full
  - One year loss and damage warranty
  - Repair coverage as needed

Speech Language Therapy

- Received through Early Intervention
  - Birth to 3 years
- Currently receives services through the school system
  - Group therapy twice per week
  - Individual therapy once per week

Multiple Coverage Sources of EHDl System Costs

- Private Insurance
- Public Insurance
- Hospital absorbs cost
- Health Department
- Private Foundations
- Parents
- Audiologist/SLT absorbs cost

Costs of Identification For the Family and For Society

Early
- \(\uparrow\) costs amplification, habilitation birth to 3 years
- \(\uparrow\) communication skills, mainstreaming
- \(\uparrow\) success in work, private insurance

Late
- \(\downarrow\) costs birth to 3 years
- \(\downarrow\) success school language, work force
- \(\uparrow\) costs of education, habilitation, public insurance

Can We: afford universal newborn screening? not afford to do universal newborn screening?

Savings for Families and for Society

- Increased numbers of happy and productive children and adults who are active and effective communicators.
AUDITORY SYSTEM DEVELOPMENT AND PLASTICITY: IS EARLY INTERVENTION NECESSARY?

Gaspard Montandon, Laboratoire de physiologie de l’audition de Bordeaux, Bordeaux, France
Thursday, January 23, 2003, 1:30 to 3:00 PM

From the moment of birth, the newborn child is plunged into a world that is rich in auditory stimulation. The child progressively learns to receive and decrypt this information, and to integrate and relate it to other sensory information. This sonic information, by way of several episodes of greater sensitivity, will gradually foster the development of the auditory system and the maturation of the auditory areas of the brain, and eventually enable the child to acquire a language. The role of audition highlights the possible consequences hearing impairment can have on a child’s cognitive development.

The importance of sensory activity on cerebral development can be understood by observing the auditory development of children with normal hearing, children with deafness and children with hearing aids. Recent behavioural and neurobiological data for these three types of listeners will be presented along with the critical stages of cerebral development. These data will be completed by data from animal models. This comparison will demonstrate the consequences of hearing impairment on maturation and will help in understanding the possible benefits of hearing aids such as cochlear implants. Even though the auditory system is highly plastic, certain stages are irreversible. Defining these stages of development and understanding why they are fundamental to the normal development of the auditory system will follow.
Développement du système auditif et plasticité. Doit-on nécessairement intervenir précocement ?

Development of the hearing system and plasticity: Is early action required?

« Colloque nord-américain sur le dépistage et l'intervention de la surdité à la petite enfance »

Gisèle Montandon, Université Victor Segalen Bordeaux 2, France

Questions importantes...

- Quels sont les étapes critiques du développement auditif ?
- Quels sont les mécanismes neurobiologiques responsables de ces étapes ?
- A partir de quel âge devient-il difficile de réhabiliter un enfant malentendant ?
- Combien de temps est-il nécessaire à un enfant pour réapprendre un langage ?

A. Déficits comportementaux liés à une privation auditive

- Périodes critiques pour l'acquisition d'un deuxième langage (Johnson et Newport, 1989)

La compétence linguistique d'individus ayant appris un deuxième langage à des âges différents est comparée. Les immigrants ayant appris l'anglais entre 3 et 7 ans ont eu des performances plus élevées que les anglophones de langue maternelle. Les anglophones qui ont appris l'anglais entre 5 et 7 ans ont performé aussi bien que les immigrants.

Mais les immigrants arrivés à des âges plus avancés ont des performances diminuant progressivement avec l'âge. Les groupes ont été évalués et leurs résultats ont été représentés par des courbes de performance, indiquant une diminution progressive des performances avec l'âge.

B. Périodes critiques de l'apprentissage d'un langage

- Phascept 2-10 mois
- Capacités phonologiques 2-4 ans
- Capacités syntaxiques 4-15 ans

C. Neurophysiologie du développement du cortex auditif

Neurophysiology of sensitive periods in the auditory cortex
A. Déficits comportementaux liés à une privation auditive

Behavioral deficits following auditory deprivation

1. Le cas des enfants sauvages
   The case of the wild children
   - Un enfant a été enfermé dans une chambre sombre pendant 6 ans avec sa mère sourde (Mason et al., 1942)
   - A child is isolated in a dark room for 6 years with deaf mother (Mason and
     et al., 1942)
   - Après 10 mois d'apprentissage, l'enfant sauvage a appris à lire, à écrire et à composait des phrases.
   - After 10 months of exposure, the wild child could read, write, and compose sentences.
   - Génie a été découvert à 15 ans. Il a été isolé depuis l'âge de 20 mois (Cartier, 1971)
   - Genius was discovered at 15 years of age. He was isolated since the age of 20 months (Cartier, 1971)
   - Après 7 ans de rééducation, Génie a acquis une bonne maîtrise de la langue. Il a développé une bonne compétence en langage (2)
   - After 7 years of rehabilitation, Genius achieved good language acquisition.

2. Périodes critiques lors de l'acquisition d'un langage chez un militantmendant
   Critical periods for language acquisition for deaf people
   - Learning American Sign Language at different ages (Newport et al., 1985)
   - Learning of a second language (Unpublished Newport, 1987)
   - Phonological development (Menn et al., 1990)

   Temps (années) / Time (years)
   0 2 4 6 8 10 12

3. Facteurs nécessaires à la récupération du langage grâce à une implantation cochléaire
   Important factors in language recovery following cochlear implantation
   - 1. Ages critiques pour l'implantation cochléaire
      Critical ages for cochlear implantation
   - 2. Ages du début de la surdité
      Ages of the beginning of auditory deprivation
   - 3. Durée de la privation auditive
      Duration of auditory deprivation
A. Déficits comportementaux liés à une privation auditive

- **Recouvrement du langage à l'âge bas** (Manrique et coll., 1999)
  - Test de perception de la parole
  - Étude croisée dans un contexte homogène
  - Comportement de langage
  - Étude de suivi et de contrôle
  - Étude de suivi et de contrôle

B. **Compétences de langage**

- Étude de suivi et de contrôle
- Étude de suivi et de contrôle
- Étude de suivi et de contrôle

**Implantation séquençée chez les enfants sourds prématurés** (Frye et coll., 1997)
- Cocéhan implant use by prématurément déblindés enfants (Frye et coll., 1997)
- Étude de suivi et de contrôle

**Conclusion**
- Périodes critiques du développement du langage et de l’audition
- Pau d’effet de la durée de privation sur l’acquisition du langage
- Acquisition du langage possible chez l’adulte (Gill et coll., 1997)
B. Périodes critiques du développement auditif

Critical periods for auditory development

- Introduction

1. Maturation des aires du langage
2. Maturation des aires auditives ( cortex auditif primaire et cortex auditif associatif)

- Efficacité des implants cochléaires chez des sourds pré- et postlinguaires (Okazawa et coll., 1996)

- Plasticité subérente à une implantation (Giraud et coll., 2001)

- Visuo-spatial localization and its relation to deviance detection in adult congenitally deaf listeners (Amaral et al., 2003)

- Integrating Mechanisms: A1 and A2 in the Human Auditory Cortex (Halgren et al., 1982)

- Activations chez des sourds pré- et postlinguaires (Okazawa et coll., 1996)

- Plasticité associée à une sordité pré-linguale

- Analyse des performances visuelles associées à une augmentation de l'activation des aires visuelles

- Usage des zires auditives et de cellules du langage par d'autres modalités sensorielles

- Disaccord entre les données fonctionnelles et la sensation reportée par les patients stimulés électriquement
B. Périodes critiques du développement auditif

- Une amélioration des performances visuelles associée à une augmentation de l'activation des aires visuelles (Bavelier et coll., 2000)
- L'activité du cortex auditif associée induite par un implant cocchléaire décroît avec l'augmentation de l'âge d'implantation (Lee et coll., 2001)
- Une longue privation auditivement d'activités du cortex auditif induit une baisse d'activité cérébrale (Tao et coll., 1993)
- Les paramètres fonctionnels du cortex auditif primaire se développent pendant les 12-15 premières années (Eggermont et coll., 1988)
- Les aires du langage semblent épargnées par une privation auditive
- Le cortex auditif primaire doit être stimulé pour se développer (Baines et coll., 1995)

C. Neurophysiologie du développement du cortex auditif

- Introduction
- Comme les aires du langage, le cortex auditif primaire se développe durant la vie postnatale (Ponton et coll., 1996)
- Les aires du langage, les aires auditives et les aires visuelles sont également stimulées au cours de la grossesse (Abbott et coll., 1997)
- Les aires auditives et les aires visuelles possèdent une activité neuronale même avant la naissance (Duffy et coll., 1996)
- Les aires auditives et les aires visuelles possèdent une activité neuronale même avant la naissance (Duffy et coll., 1996)
ABSTRACTS AND POWERPOINTS

C. Neurophysiologie du développement du cortex auditif
Rechromatography of sensitive periods in the Auditory Cortex

1. Mécanismes neuronaux du développement du cortex auditif
Neural development of auditory cortex

1. Rappel synaptogénèse génétiquement déterminée
Reap synaptogénèse génétiquement déterminée

2. M éturation synaptique lente et stabilisation ou élimination de 50% des synapses (pruning)
Slow synaptogénie et stabilisation ou élimination de 50% des synapses (pruning)

Développement du cortex auditif chez l’animal sourd
Development of auditory cortex in deaf animals

Activation synaptique décroissante au niveau du cortex auditif primaire des animaux sourds congenitaux
Decrease of synaptic activity in deaf animals

Faible réorganisation céphalique
Low cortical reorganization

Les projections du cortex auditif vers les structures sous-corticales et vers les parties supérieures sont diminuées
Projections of the primary auditory cortex towards subcortical structures and towards higher-order cortices are diminished

Conclusion

A. Défis comportementaux : Période critique vers l’âge de 3-5 ans pour la perception de la parole chez les enfants sourds congenitaux

Critical period between 3 and 5 years of age for speech perception for deaf children

B. Plasticité du cortex auditif :

- Utilisation des aires auditives par d’autres modalités sensorielles chez les sourds prédéterminés
- Recrutement et acquisitions de nouvelles modalités sensorielles prédéterminées

- Dépendance de l’activité extrémale des aires auditives avec l’augmentation de la durée de la privation

- Dépendance de l’activité extrémale des aires auditives avec l’augmentation de la durée de la privation

C. Développement du cortex auditif

Mise en place de la synaptogénèse pariétale à partir de la première année de la vie, puis élimination des synapses

- Postnatal synaptogenesis starts in the first year of life, then synapse elimination

Activité électrique du cortex auditif primaire plus faible chez les sourds implantés tardivement

Low electrical activity of the primary auditory cortex for late-implanted deaf

Merci pour votre attention
Thank you for your attention
EVALUATING A PROGRAM: INFRASTRUCTURE AND QUALITY STANDARDS

Dr. David K. Brown, Auditory Research Program, University of Calgary, Alberta, Canada
Thursday, January 23, 2003, 3:30 to 5:30 PM

Screening a newborn’s hearing has become easier as technology has evolved. Numerous automated devices are now available, which provide an objective physiologic screening and can be performed by non-audiologists. However, the act of hearing screening is in reality a small part of the overall objective in a screening program. Screening a newborn may in fact be the easiest part of the program. Once the newborn has been screened, their results need to be tracked. This is especially true in the case of a “refer” result, where it is the underlying objective of the program for that child to receive follow-up evaluations to determine their hearing status. Therefore tracking infants as they flow through Early Hearing Detection and Intervention (EDHI) Programs is vital so that they are not lost somewhere in the system. The tracking system is also an integral part of the evaluation of the Program.

Program evaluation and quality improvement are essential components of any high-quality EDHI Program. They must exist as sub-programs of the overall EHDI program and should address structure, process and outcome elements of the entire Program. Goals and objectives must be developed, to not only guide a program but also to measure its success. Quality improvement is a conceptual approach that incorporates ongoing and continuous re-examination of the program components in order to determine proactively whether the program is functioning as effectively and efficiently as possible. It also can determine when and where problems or concerns arise, so that the components can be maintained, repaired and wherever feasible, enhanced. Program evaluation is a formal method to ascertain if the overall goals and specific objectives are actually being achieved. Both of these components must be present not only to determine the success of an EDHI program but to effect change to ensure its success.
The PowerPoint is not available.
THRESHOLD ESTIMATION USING THE TONE-EVOKED AUDITORY BRAINSTEM RESPONSE: FUNDAMENTALS AND RESULTS

Dr. David R. Stapells, School of Audiology & Speech Sciences, University of British Columbia, Vancouver, Canada
Thursday, January 23, 2003, 3:30 to 5:30 PM

The auditory brainstem response (ABR) is an essential tool for the audiology clinician. With the advent of universal newborn hearing screening and subsequent need for timely and appropriate diagnostic evaluation for young infants, ABRs have actually increased in their importance for audiologists. It is now essential to obtain the information necessary to fit amplification in a 3-month-old infant. Currently, only the tone-evoked ABR can adequately provide the frequency-specific air- and bone-conduction threshold information required. This presentation cover:

(i) what are the current problems with ABR audiometry by clinicians,
(ii) why tone-ABRs are required,
(iii) results with tone-ABRs,
(iv) bone-conduction tone-ABR assessment, and
(v) test protocols, procedures and sequence.
The PowerPoint is not available.
THE CANADIAN WORKING GROUP ON CHILDHOOD HEARING

Sharon Bartholomew, Health Canada, Ottawa, Ontario, Canada
Dr. Andrée Durieux-Smith, Vice-dean, Faculty of Health Sciences, University of Ottawa, Ontario, Canada
for the Canadian Working Group on Childhood Hearing
Friday, January 24, 2003, 8:30 to 10:00 AM

Introduction
The Canadian Working Group on Childhood Hearing (CWGCH) was established in September 2000 by Health Canada. It was formed in response to recent growing interest in the identification and management of newborn hearing loss by various groups of health professionals, educators and consumers and at different levels of government. Membership in the CWGCH includes professional associations; consumers/parents; and experts in otolaryngology, audiology, speech-language pathology, nursing, child health and public health.

Objectives
The mandate of the CWGCH is to develop an evidence-based resource manual for early hearing detection and intervention to address the needs of children with hearing loss and their families in Canada. The resource manual will address the following: target group, screening, audiological and medical assessment, interventions, and infrastructure. The Working Group is committed to four guiding principles: National Role, Evidence-Based Approach, Family-centred Approach, and Partnership and Collaboration.

Methodology
The resource manual is being developed by a process which includes: systematic and critical reviews of the evidence; drafting of the manual by Working Group members with various content expertise; and a process of consultation with various stakeholders in Canada. The consultations will provide an opportunity for provincial and territorial governments and healthcare professionals, educators and consumers to contribute towards the finalization of the manual.
Conclusion
The Working Group aims to foster collaboration among all stakeholders to build on experiences, create linkages and provide opportunities for the promotion of best practices in early hearing detection and intervention across Canada. The Working Group will contribute to the field by producing a science-based report which will be a useful reference document for jurisdictions in Canada for their policy and programming decisions in early hearing detection and intervention.
Canadian Working Group on Childhood Hearing (CWGCH)

Québec
January 24, 2003

Andrée Durleux-Smith and Sharon Bartholomew, Co-chairs, CWGCH

CWGCH@hc-sc.gc.ca

Outline
- Introduction
- Guiding Principles
- Evidence-based Approach
- Resource Manual
- Next Steps

History of Working Group
- Group of researchers approached HSED to support the Canadian Newborn Hearing Screening Survey
  - Survey found that there was no unified approach in Canada
  - Little being done in terms of evidence-based approach

History of Working Group (2)
- Other activities beginning:
  - Canadian Association of Speech-Language Pathologists and Audiologists case statement
  - Canadian Pediatric Society (CPS)
  - Alberta and Ontario newborn hearing screening programs
  - Need for a national leadership identified

Canadian Working Group on Childhood Hearing established

Mandate
To develop an evidence-based resource manual on early hearing detection and intervention (EHDI) which will assist those wishing to develop EHDI policies and programs.
Guiding Principles (1)
The CWGCH is committed to four guiding principles:

- **National Role**: providing leadership in the development and dissemination of a resource manual on early hearing detection and intervention in Canada.

- **Evidence-based Approach**: to the development of the resource manual for early hearing detection and intervention, through the systematic review of currently available research and program information.

Guiding Principles (2)

- **Family-centred Approach**: an integrated approach to families, reflecting an understanding of the physical, emotional, mental and psychosocial aspects of hearing detection and intervention for children with hearing impairment and their families.

- **Partnership and Collaboration**: working in partnership with various stakeholders, including federal, provincial and territorial governments; professional associations; consumers/parents; and national and international experts in otolaryngology, audiology, speech-language pathology, nursing, child health and public health.

WHO Principles and Practices of Screening for Disease

1. The condition sought should be an important health problem.
2. There should be an accepted treatment for patients with recognized disease.
3. Facilities for diagnosis and treatment should be available.
4. There should be a recognizable latent or early symptomatic stage.
5. There should be a suitable test or examination.
6. The test should be acceptable to the population.
7. The natural history of the condition (including development from latent to declared disease) should be adequately understood.
8. There should be an agreed upon policy on whom to treat as patients.
9. The cost of case finding (including diagnosis and treatment of patients diagnosed) should be economically balanced in relation to possible expenditure on medical care as a whole.
10. Case finding should be a continuing process and not a ‘once and for all’ project.

Multidisciplinarity

- **Increased relevance to all those who are part of the system**

- **Membership**: national professional associations; consumers/parents; and experts in otolaryngology, audiology, speech-language pathology, nursing, child health and public health from across Canada.

CWGCH: Multidisciplinary representation.

- Health Surveillance and Epidemiology Division and Childhood and Youth Division, Centre for Healthy Human Development, Health Canada.

- Canadian Academy of Audiology (CAA)

- Canadian Association of Speech-Language Pathologists and Audiologists (CASLPA)

- Childhood Hearing Network of Canada

CWGCH representation (cont.)

- Canadian Paediatric Society.

- Canadian Society of Otolaryngology- Head and Neck Surgery.

- College of Family Physicians

- Canadian Association of Educators of the Deaf and Hard of Hearing.

- Institut National de Sante publique du Quebec (INSPP)

- Consumer representative (parent)

- Other selected researchers and health practitioners.
**CWGCH : geographic representation**
- British Columbia
- Alberta
- Ontario
- Quebec
- New Brunswick
- Nova Scotia
- Newfoundland

**Why Health Canada?**
- **Mission**
  - To help the people of Canada maintain and improve their health
- **Children**
  - Children are a priority to Health Canada
  - Health Surveillance and Epidemiology Division
- **Leadership**
  - Ability and role to bring different disciplines and jurisdictions together at a National Level

**Why Health Canada? (2)**

**Knowledge Development**
- Information gap identified
- Health Canada has the mandate and a proven track record
- Important Health Canada role to support effective public health programs through knowledge synthesis
- Surveillance and Evidence building work to facilitate knowledge development

**Health Canada Experience**
- Expert working group on travel medicine – Committee to Advise on Tropical Medicine and Travel (CATMAT)
- National Advisory Committee on Immunization
- Sudden Infant Death Syndrome Joint Statement
- Folic acid and prevention of neural tube defects

**EVIDENCE-BASED APPROACH**
  - Screening tests for hearing impairment can improve identification of newborns with PHL, but the efficacy of UNHS to improve long-term language outcomes remains uncertain.
  - *JAMA* Vol.286, No.16, 2000-2010
The evidence (cont.)

- Miller and Zwaigenbaum (2001)
  - New provincial initiatives for childhood disabilities: the imperative for research.
  - UNHS is endorsed by a broad consensus of professional opinion, but it has never been subjected to a clinical trial of the kind demanded by contemporary standards for evidence-based health care.
  - JAMC Vol. 164, No. 12, 1704-1705

The U.S. Preventive Services Task Force (USPSTF): available scientific evidence is insufficient to recommend for or against routine screening of newborns for hearing loss. (based on report by the Evidence-based Practice Centre at Oregon Health and Science University) (October 2001)

The evidence (cont.)

- Canadian Task Force on Preventive Health: Enhancing the Health of Canadians through Evidence-based Prevention:
  - Conclusion regarding the evidence for benefit for speech and language: INSUFFICIENT EVIDENCE.
  - March 2002

Development of a Resource Manual

1) Identify through a consultative process, the issues to be addressed
2) Draft Table of Contents of Manual
3) Undertake systematic and critical reviews of the evidence
4) Draft resource manual
5) Consult stakeholders
6) Finalize, publish, present and distribute manual

Draft Table of Contents

- Forward
- Introduction
- Guiding Principles
- Background
- Burden of Target Disorder
- Screening
- Assessment
- Audiological
- Medical
- Intervention
- Amplification
- Habilitation
- Outcomes
- Program Evaluation
- Infrastructure and Systems
- Conclusions and Future Directions
- Glossary of Terms
- Bibliography

BACKGROUND: HISTORICAL PERSPECTIVE OF EHDI IN CANADA

Conferences

- 1964: Conference on the Young Deaf Child
  - Objective: to find ways to alleviate handicap to auditory communication imposed by early hearing loss.
  - Neonatal hearing testing seen as a goal but technology not available.
BACKGROUND: Conferences (cont.)

  - Focal point: approval of methods for screening newborns for hearing loss
  - Use of High Risk Register
  - Behavioural screening

- 1978: Saskatoon Conference on Early Diagnosis of Hearing loss.
  - Focal point: methods for the confirmation of the presence and degree of hearing loss within the first 6 months of life.
  - 4 papers on electrophysiological methods for the assessment of auditory function in infants
  - ABR: valuable method for accurately identifying hearing loss in graduates of NICUs
  - IT IS POSSIBLE TO IDENTIFY HEARING LOSS IN NEWBORNS.

BACKGROUND: Conferences (cont.)

- 1983: Canadian Experience in Neonatal hearing assessment by ABR.
- Biennial meeting of the Electric Response Audiometry Study Group (Ottawa)

- ABR POWERFUL TOOL IN THE IDENTIFICATION OF HEARING LOSS IN INFANTS

BACKGROUND: Committees and Task Forces.

- 1977: Meeting of the Canadian Otolaryngological Society’s Committee on Childhood Hearing Impairment (Montebello)
- 1979: Canadian Advisory Coalition on Childhood Hearing Impairment (CACCHI)

Task Force on Childhood Hearing Impairment

- Recommendations:
  1) Screening of high risk children in infancy
  2) Increased awareness of primary care physicians and other health care professionals on normal speech and language milestones and danger signals for hearing loss.

Task Force Recommendations (cont.)

- 3) Provincial and territorial centralized computerized record keeping system of children with a permanent hearing loss
- 4) Comprehensive services in health care and education.
- 5) STANDING INTERDISCIPLINARY JOINT COMMITTEE TO MONITOR NEW KNOWLEDGE ABOUT HEARING LOSS IN CHILDREN.
BACKGROUND (cont.)

- Brown, Dort and Sauve (2000)
  - Canadian Newborn Hearing Screening Survey, sent to all birthing centres in Canada (n=467)
  - 384 replies (82%)
  - 10% (n=35) reported some kind of screening activity of newborns
  - LITTLE PROGRESS SINCE 1984 REPORT OF CHI TASK FORCE.

- In Canada, up till recently, there has been no systematic approach to early identification, diagnosis and management of hearing loss in children.
- There has been a sparse uncoordinated patchwork of ad hoc local initiatives covering only a fraction of newborns considered at high risk for hearing loss.

BACKGROUND (cont.)

- Durieux-Smith and Whittingham (2000):
  - Study of the age of diagnosis of screened (NICU) and non-screened children.

Risk Status for the 613 children, wearing hearing aids, born between 1974 and 1995

- NICU premature (14.2% 91)
- In Utero Infections 1.9% (7)
- Congenital Anomalies 1.9% (1)
- Family History 13.4% (82)
- Unknown 57.1% (350)

Age of diagnosis of hearing loss

Degree of hearing loss and age of diagnosis
BURDEN OF THE TARGET DISORDER

Prevalence
- Ascertainment studies: Fortnum et al. (2001)
  - 15 year birth cohort in the UK (1980-85): 17160 cases ascertained
  - PCHI: 40 dB HL avg in better ear
  - Adjusted prevalence: 1.07 out of 1000 live births
    - at 3 years and 2.05 at 9 plus years

BURDEN OF TARGET DISORDER: prevalence (cont.)
- UNHS programs:
  - Prieve et al. 2000: adjusted prevalence of hearing loss greater than 20 dB HL in either ear: 2.8 out of 1000
  - Differences in estimates of congenital prevalence of PCHI due to differences in definitions of the target disorder, sensitivity
    of screening methods used, accuracy and timing of hearing assessments and incomplete follow-up
    - 1.06 out of 1000 to 3.2 out of 1000

BURDEN OF TARGET DISORDER: prevalence (cont.)
- Risk Groups
  - Proportion of infants at risk: 3-15%
  - Ascertainment studies:
    - 1.12 out of 1000
    - 3.2 out of 1000 (NICU)
  - UNHS: 4.8 out of 1000 (bilateral)

BURDEN OF THE TARGET DISORDER
- Temporal pattern of PCHI detection in the absence of UNHS
  - Studies on the age of diagnosis in the absence of any screening activities,
  - Studies on the age of diagnosis in the presence of some screening activities.

BURDEN OF THE TARGET DISORDER: Temporal pattern (cont.)
- Summary:
  - In the absence of UNHS, the average age of diagnosis of children exceeds 12 months
  - Children with risk factors or additional medical conditions are diagnosed sooner
  - There is an inverse relationship between the age of diagnosis and the degree of hearing loss.

BURDEN OF THE TARGET DISORDER: Temporal pattern (cont.)
- Summary (cont.)
  - The ages of diagnosis of children born in the absence of any screening activities or in the presence of some screening activities are very similar
  - In studies which have investigated different birth cohorts, the ages of diagnosis have improved over time
OUTCOMES: Evidence supporting UNHS

- Assumptions:
  - Screening tests are accurate
  - Screening reduces delays in diagnosis and intervention
  - Earlier treatment results in:
    - Better language function within the pre-school period
    - Improvement in early language which will improve educational, occupational and social functioning in later life

OUTCOMES (Cont.)

- Thompson et al. 2001: critically assessed the evidence supporting UNHS

- Results:
  - Modern hearing screening tests can improve identification of newborns with PHL
  - Effectiveness of UNHS to improve long-term language outcomes remain uncertain

SCREENING

Importance of defining target disorder and target population

- Target disorder: severity, frequency range, laterality, permanence of site of lesion, time of onset
- Target population: at risk or all children

SCREENING (Cont.)

Automated ABR (AABR)

- Binary pass-fail outcome
- Factors: behavioural state of infant, electrode placement, earphone application
- Click stimulus
- Accuracy of AABR: depends on definition of the target disorder

SCREENING (Cont.)

Measures of screening test performance

- Common measures of test accuracy:
  - Sensitivity
  - Specificity
  - Positive Predictive Value (PPV)
  - Negative Predictive Value (NPV)
  - Referral rate
  - Number needed to screen (NNS) in order to identify a single case.

Automated Otoacoustic Emissions (AOAE)

- TEAOs, DPOAEs
  - Factors: external canal free of debris and verno
  - MULTISTAGE TESTING WITH AOAEs FOLLOWED BY AABR LOWERS REFERRAL RATE.
SCREENING (Cont.)
- Other factors:
  - Screening coverage and follow-up compliance
  - Key differences between high risk and universal screening
  - Potential harms associated with screening outcomes
  - Surveillance and referral components

AUDIOLOGICAL ASSESSMENT
- Electrophysiological frequency-specific threshold estimation:
  - Tonepip ABRs
    - Subjective interpretation of results
    - Careful stimulus calibration
  - Auditory Steady State Responses (ASSR)
    - Emerging technology

AUDIOLOGICAL ASSESSMENT (Cont.)
- OAEs: useful contributors to the audiological assessment by providing a limited cross-check of other test results
- Middle Ear Analysis
  - Importance of high frequency probe tone
  - VRA: ear specific and frequency specific (6-10 months of age)
  - Special consideration: Auditory Neuropathy

OTOLOGICAL ASSESSMENT AND MANAGEMENT
- Assessment and management of middle ear disease in children less than two years of age
  - Diagnosis
  - Use of pneumatic otoscopy
  - Treatment
  - Ventilation
    - Bilateral myringotomy with tubes
  - Challenges: little published literature on children with OME who are under 1 year of age

INTERVENTION: AMPLIFICATION
- Evidence that hearing aids improve auditory performance: Use of binaural amplification in infants with measurable hearing is recommended.
- For infants, frequency-specific ABR threshold estimates should be used to fit amplification prior to 6 months of age.

INTERVENTION: AMPLIFICATION (Cont.)
- Acoustic properties of ear canals of infants and young children show high between subject variability. These differences need to be applied to the amplification process.
- "Rear Ear Coupler Difference (RECD): reliable and valid measure for the purpose of fitting amplification in infants."
INTRODUCTION: AMPLIFICATION (Cont.)
- A pediatric-specific selection strategy that is evidence-based should be utilized whenever possible.
- Couple based verification in conjunction with individual RECD measures is a valid procedure for the electroacoustic verification of hearing instruments in infants.

INTRODUCTION: AMPLIFICATION (Cont.)
- What are the adverse effects (if any) from hearing aid fitting?
- Poorly controlled retrospective studies and use of group data contribute to these discrepancies.
- Importance of monitoring auditory thresholds and function of hearing aids.

INTRODUCTION: HABILITATION
- Systematic Review
  - Are early habilitation programs effective in improving communication development?
  - What is the evidence for the efficacy and effectiveness of specific intervention strategies (AV, Oral, ASL, TC)

- Review carried out by the Thomas Chalmers Centre for Systematic Reviews (Ottawa)

INTRODUCTION: HABILITATION (Cont.)
- Historically there have been 2 major philosophies aimed at developing communication skills.
- Regardless of the approach, the belief is that early intervention is key to the development of communication and social skills, and academic functioning.
- The choice of an intervention option for parents must be an informed choice based on scientific evidence.

INTRODUCTION: HABILITATION (Cont.)
- Systematic Review: Schachter et al. 2002
  - 625 citations
  - 194 unique studies entered into data abstraction
  - Evidence tables were derived with variables highlighted by clinical content experts
  - PREPONDERANCE OF MISSING DATA

- Observations obtained in systematic review do not permit to confirm or disconfirm the absolute or comparative effectiveness of any of the four types of intervention programs reviewed.
- Evaluation of effectiveness of habilitation programs is complex
- NEED FOR WELL CONTROLLED AND WELL DESIGNED STUDIES
Program Evaluation and Quality Improvement

- Essential Components of EHDI programs
- Require explicit and precise *a priori* definitions of objectives
- Address program as a whole and each major component
- Should address structure, process and outcome elements
- Include clear mechanisms for responding to deficiencies

Program Evaluation (PE)

- Formal method to ascertain if goals and objectives are being achieved
- Process can and must effect change
- Requires explicitly defined and quantifiable outcomes

Quality Improvement (QI)

- Ongoing and continuous re-examination of program components
- Used to determine proactively whether program is functioning as effectively and efficiently as possible
- Determines when and where problems arise
- Allows program to be revised and enhanced

Components of PE and QI

- Structural
  - Examples: personnel, information systems
- Process
  - Examples: screening and followup, audiological assessment

Measuring Performance

- Realistic indicators and benchmarks must be defined
  - These are used to determine if the program is meeting its performance targets
- Benchmarks are levels to which the program aspires
  - Important for proving the program is beneficial and cost effective

Outcome Measures

- Measures range from screening proportions to communication development outcomes
- Examples:
  - Number (N) and proportion (P) of overall target population screened by one month of age and one month post-discharge
  - N and P of infants with refer (by site, screening personnel and risk status)
  - N and P of referrals for whom audiological assessment is initiated by three months of age and within two months of the screening result
Performance benchmarks

Benchmarks range from percent of UNHS coverage to intervention uptake.

Examples:
- UNHS coverage: at least 95% of all infants successfully screened by 3 months of age.
- Referral Rate at discharge: less than 4% of infants should be referred for second stage screening or follow-up evaluations.
- Infant Follow-up: at least 90% of referred infants should return for second stage screening.
- Audiological Assessment: no more than 2% of infants screened should require detailed audiological assessment.

Long term Outcomes

- Sub-program outcomes vs ultimate measure of success.
- Ultimate measures of success include:
  - Family satisfaction with services.
  - Improved hearing ability.
  - Many factors impact on outcome.

Economic Evaluations

- Routinely required for accountability and sustainability.
- Capital and operating costs of all program structures and process.
- Basic measures: overall cost per infant screened, cost per infant identified with PCHI.
- Further investigation is needed to develop a more comprehensive methodology to approach true cost-benefit analysis.

PE and QI Conclusion

- PE and QI is an indispensable tool:
  - for measuring achievement of program’s objectives.
  - for justification of a program’s continued existence.

Essential that a comprehensive and effective PE and QI program be a core part of any high quality EHDI initiative.

Infrastructure and Context

- The scope of an EHDI program is very broad.
- A public health approach most likely to succeed.
- Well designed infrastructure is essential for success.
- All components of an EHDI program must be given equal attention.

Infrastructure

- Those elements that support, sustain and link all program components to achieve the program goal.
- Main components:
  - Human Resources.
  - Information Services.
  - Administrative Systems.
  - Communication Systems.
Human Resources
- Non-traditional staff recruitment and retention considerations
- Consider who among staff will find work challenging and interesting
- Clerical support, information systems staff, a co-ordinator, as well as professional staff are key to program success

Information Systems
- Critical for tracking, follow-up, seamless transition from one program stage to the next, and program evaluation
- Standard software packages are available
- In-depth understanding of the goals of the program and use of the information is essential when custom designing a program
- Consent and confidentiality are important considerations

Administrative Systems
- Includes
  - Financial, staff, and clinical records
  - Budgeting and fund raising
  - Accountability/reporting
  - Forms development
  - Program evaluation and quality improvement

Communication Systems
- A broad-based communications/public relations program is invaluable
- Parents, professional groups and consumers should receive information by multiple means
- Different groups prefer different information formats
- Should result in improving awareness, consent, compliance and ultimately long-term sustainability of program

Infrastructure: Conclusion
- EHDI programs exist within social, cultural, and political values and contexts
- Being prepared with evidence such as the resource manual

Resource Manual Conclusion
- The goal of the resource manual is to provide the most up-to-date evidence
- Uses:
  - Programs and Policy
  - Professional practice
  - Everyday decision-making

What has been presented here is a draft
CWGCH Status
Current Status
- Drafting of resource manual
- Consultations with key stakeholders

Consultations
- Provincial/Territorial or Regional
- Key stakeholders
- Comment on Draft resource manual
- Comments used to revise resource manual so that it is applicable to all jurisdictions in Canada

Next Steps
- Revision of manual
- Resource Manual and Critical/Systematic Reviews released next Summer
- Presentation of results

Discussion Items
- Is this the type of information which would be useful to you
- Can you suggest any areas we have not covered

CWGCH Membership
Expert Representatives of:
- Canadian Pediatric Society
- Canadian Association of Educators of the Deaf and Hard of Hearing
- Institut national de santé publique du Québec (INSPEQ)
- The Colleges of Family Physicians of Canada
- Canadian Association of Speech-Language Pathologists and Audiologists
- Childhood Hearing Network of Canada
- Canadian Academy of Audiology
- Canadian Society of Otorhinolaryngology-Head & Neck Surgery
- Health Canada Representatives
- Individuals Researchers and Practitioners
- Parent of a deaf child

Department of Health Act, 1996
The Minister’s powers, duties and functions relating to health include the following matters:
c) Investigation and research into public health, including the monitoring of diseases
EXAMPLES OF EARLY DETECTION AND INTERVENTION OF DEAFNESS PROGRAMS AT DIFFERENT STAGE OF IMPLANTATION: NEW YORK STATE, ONTARIO, ALBERTA AND NEW BRUNSWICK

Dr. David K. Brown, Auditory Research Program, University of Calgary, Alberta, Canada

Dr. Judith S. Gravel, Hunter College of the City University of New York, New York, New York State, United States

Dr. Martyn Hyde, Hearing and Balance Unit, Mount Sinai Hospital, University of Toronto, Ontario, Canada

Dr. Johanne Roussel-Maltais, Hôpital Dr G.-L. Dumont, Moncton, Nouveau-Brunswick, Canada

Friday, January 24, 2003, 10:30 AM to 12:00 PM

The abstracts are not available.
The PowerPoint is not available.
SCREENING TEST PERFORMANCE

Dr. Martyn Hyde, Hearing and Balance Unit, Mount Sinai Hospital, University of Toronto, Ontario, Canada
Friday, January 24, 2003, 1:30 to 3:00 PM

A review is presented of hearing screening tests in common use in programs for Universal Newborn Hearing Screening (UNHS)/Early Hearing Detection and Intervention (EHDI). The focus is upon the operating characteristics, strengths and limitations of Otoacoustic Emissions (OAE) and Automated Auditory Brainstem Response (AABR) screening.

The review begins with principles of test evaluation, including definition and explanation of sensitivity and specificity, and associated false-negative and false-positive error rates. Sensitivity and specificity are not fixed, but vary inversely as the pass-refer criteria change; their relationship is summarized by the relative (receiver) operating characteristic (ROC).

It is common to repeat a screening test of a given type or to combine tests of different types into a serial, multi-stage protocol. The usual aim is to increase the protocol specificity while maintaining the highest possible overall sensitivity. The principles of test combination and the resulting effects on sensitivity and specificity are discussed.

When screening tests or multi-test protocols are used, they assign infants to categories of high or low risk of impairment. The outcomes are quantified by measures such as positive and negative predictive values, referral rates and yield. These are defined and explained.

The definition and prevalence of the target disorder can affect sensitivity and specificity, and have strong effects on the screening outcome measures. These interactions are explained. Recent evidence on prevalence of hearing impairment is outlined, including the effects of hearing loss severity, frequency profile, laterality and time of expression. Variations in the definition of the target disorder are discussed.

Characteristics of typical, commercial OAE and AABR screening devices are outlined. Some current evidence about their performance is summarized. Difficulties in obtaining accurate and consistent values for sensitivity include extreme sample size demands, the requirement for complete follow-up of the screened cohort, limitations of "gold standard" tests, and changes in true hearing status over time.
The review concludes with comments on operational issues in screening programs. These include choice of screening test(s) for different sub-populations, the timing and exact conduct of screening, environmental and personnel factors. These variables contribute to performance variations across programs and among centres, and also to trends over time in test, protocol and program performance.
Screening Test Performance

Martyn Hyde, PhD
Mount Sinai Hospital, Toronto

Context & goal of screening
Universal Newborn Hearing Screening (UNHS)
Early Hearing Detection and Intervention (EHDI)
Early Hearing & Communication Development Program (EHCD)

A good screening program is essential BUT it is only the first step in EHCD

The goal is to provide effective services for hearing & communication development to all affected families who need and choose them.

What does screening do?
Screening DOES NOT identify infants with hearing impairment.

Screening divides infants into two categories:
HIGH & LOW RISK for significant, permanent hearing loss (PHL = SNHL + "permanent" CML)

'Fail' (+ve screen) $\Rightarrow$ high risk (eg. 1/20)
'Pass' (-ve screen) $\Rightarrow$ low risk (eg. 1/5000)

An Ideal Screening Test is:
- Safe
- Accurate
- Objective
- Quick and easy
- Not expensive

Screening Test Types
- There is NO accurate, behavioural screening test for neonates & young infants
- Automated Otoacoustic Emissions (AOAE)
  - Transient Evoked (AITEA)
  - Distortion Product (ADPOAE)
- Automated Auditory Brainstem Response (AABR)

Automated OAE Screeners
Can discriminate thresholds $\geq 35-40$ dBHL
No standard refer criteria: default + options
No standard parameters, some consensus
Similar performance for TEA, DPOAE
Insensitive to low frequency losses
Affected by environmental noise
Affected by minor middle ear disorders
Insensitive to auditory neuropathy
Hand-held, battery powered, automated, $36-10$ k
Typical test duration 2-5 minutes
Automated ABR Screeners

- ABR adjustable for target minimum loss
- Stimuli vary, usually 35 dB nHL clicks
- Must account for neonatal ear, ABR bias
- Clicks insensitive to frequency-specific loss
- No standards for ABR detection algorithms
- Less affected by minor, middle-ear disorders
- Less affected by environmental noise
- Affected by electromagnetic artifact
- Sensitive to auditory neuropathy
- Laptop (palm), mains power, $25-30k
- Typical test time 6-12 minutes
- Recent: UAE+ABB & frequency-specific AABR

‘Accurate’ means high sensitivity and high specificity

- **Sensitivity**
  - The probability that an infant with the target disorder will screen positive (refer)

- **Specificity**
  - The probability that an infant without the target disorder will screen negative (pass)

Sensitivity example

- **Sensitivity**, true positive rate, TPR

  - Example
  - Screen 1000 babies who definitely have PHL
  - Suppose 900 screen positive, 100 negative
  - Sensitivity = 0.9 (90%)
  - False negative rate (FNR) = 0.1 (10%)
  - Sensitivity + FNR = 1.0 (100%)

Specificity example

- **Specificity**, true negative rate, TNR

  - Example
  - Screen 1000 babies who DO NOT have PHL
  - Suppose 850 screen negative, 150 positive
  - Specificity = 0.85 (85%)
  - False positive rate (FPR) = 0.15 (15%)
  - Specificity + FPR = 1.0 (100%)

Relationships between sensitivity and specificity

- Sensitivity and specificity are NOT fixed, but depend on the screening pass-fail criterion

  - **Strict criterion** - positives less likely
    - Sensitivity down, false positive rate down
    - Specificity up, false negative rate up

  - **Lenient criterion** - positives more likely
    - Sensitivity up, false positive rate up
    - Specificity down, false negative rate down

Relationships between sensitivity and specificity

- Sensitivity and specificity are NOT fixed, but vary inversely as the criterion is changed. Almost any value is possible, so...

- Must have BOTH values to know test accuracy
- but how to quantify accuracy for various criteria?

- Complete picture given by Relative (Receiver) Operating Characteristic (ROC) curve, plotting sensitivity (y) against false-positive rate (x)
How to compare test accuracy?

Sensitivity or specificity alone are not useful

Sensitivity AND specificity may be insufficient, eg (0.9, 0.95) vs (0.88, 0.97)?

ROC area (A) a useful, global measure but we are usually interested in specific regions, such as high sensitivity, or low false positive rate

Sensitivity at given specificity or vice versa

Sensitivity is VERY difficult to measure, evidence limited

Cohort studies
Definitive audiometry in EVERY infant screened
Need >500 PHL cases for accurate estimate
Must screen & validate 100-500,000 babies!

UNHS programs
Only screen refers followed = sensitivity ??
Lower bound on sensitivity, if prevalence known: sensitivity >= # true positives/# screened*prev

Specificity easy to estimate, but data limited for individual tests

Prevalence of PHL is low, so a screened group will numerically approximate a normal group

Pass rate in >1000 babies screened is close approximation to actual specificity

However, most UNHS programs use multi-test screening protocols, and may not report data for infall screens only

Best sensitivity/specificity data


2,995 at-risk screened with OAE (T & DP), AABR
Reliable frequency=ear-specific VRA 9-12 months

Sensitivity ~ > 0.85 at specificity 0.9, all tests,
for PHL >= 30 HL average at 2.4 kHz
ROC A-values > 0.9

ABR better than OAE if 1 kHz losses included
**Limitations of NIH study data**

- Small sample size (56 infants with hearing loss)
- Longitudinal validation will lower sensitivity estimates (neg bias, eg by late-onset PHL)
- May have better performance due to high-quality test procedures, trained & experienced testers
- May not have optimized all technical parameters
- ? validity for low-risk population (eg more false positives in the well-baby nursery)

**Screening outcome measures**

- **Positive predictive value**
  \[ PPV = \frac{\text{true positives}}{\text{true positives + false positives}} \]
- **Negative predictive value**
  \[ NPV = \frac{\text{true negatives}}{\text{true negatives + false negatives}} \]
- **Number needed to screen (NNS)**
  \[ NNS = \frac{1}{\text{false positives}} \]

**Effect of sensitivity & specificity on screening outcomes**

**Example**

- Sensitivity = 0.9, specificity = 0.95, prevalence = 5/1000
- PPV = 450/450 = 100%
- NPV = 95,615/95,615 = 100%

**Effect of specificity on PPV**

- Sensitivity = 0.9, specificity = 0.95, prevalence = 5/1000
- PPV = 450/450 = 100%
- NPV = 95,615/95,615 = 100%

**Effects of low PPV**

- Low credibility of positive screen, eg only 1 in 12 or less actually have the target disorder
- Low follow-up compliance
- Needless family anxiety - effects?
- Expense/effort for needless diagnostic follow-up
- Possible diagnostic and intervention errors

**High specificity (low false-positive rate) VERY IMPORTANT**

- In UNHS, single-test PPVs are generally low
- Specificity has a very strong effect on PPV
- Change in specificity from 98% to 99% almost doubles the PPV
- Major effort is necessary to achieve very low false-positive rate (very high specificity)
Effect of prevalence on screening outcome measures

Sensitivity: .9, specificity: .95, prevalence 5/1000
PPV 450/5,325 = 8.5%, 1 baby in 12 has PHL
Yield 450 cases

Sensitivity: .9, specificity: .95, prevalence 1/1000
PPV 90/5,085 = 1.8%, 1 baby in 55 has PHL
Yield 90 cases

Prevalence of the target PHL has a strong effect on screening PPV and yield.

Prevalence of PHL in Infants

Is not known accurately, because:

Statistical sampling errors large

Need >1000 true cases = definitive audiometry in ~ 500,000 infants, for accurate estimate

Prevalence is affected by audiometric criteria for target disorder, age at expression, population genetics, perinatal care quality, etc.

Effects of audiometric criteria on prevalence

Lower criterion dBHL, higher prevalence

Unilaterals included, higher prevalence

Broader frequency range, lower prevalence

Effects of age at assessment on prevalence of PHL

As age increases, prevalence increases, due to

Late-onset impairment

Progressive impairment

Acquired impairment

Prevalence is a moving target.

Prevalence - sources of evidence

Prospective cohort studies (none)

Very expensive, very large samples needed, all subjects need gold standard at specific age(s)

UNHS programs (several large studies)

Give lower bound for prevalence (negative bias), imperfect sensitivity, incomplete follow-up

Ascertainment studies (1 definitive, in UK)

Health/education service records. Disorder MUST be served. Adjusted for -ve bias.

Best prevalence data

Portnaim H et al, BMJ 2001;323:336

UK national ascertainment study, all live births from 1980 to 1995

Target PHL >40 dBHL average, better ear, 17,160 children with target disorder

Prevalence estimate 1.07/1000 at 3 years increasing to 2.05 at age 9 years
Non-congenital PHL

Very complex problem, little little reliable data
NO useful data before UNHS
('congenital' should be 'presumed congenital')
Estimates of non-congenital proportion of PHL expressed at age 3 years range 5-16%
UNHS cannot detect what is not yet present,
NOT a sensitivity deficit, but a UNHS limitation
if re-screening/surveillance not included

Protocol sensitivity, specificity for 2-stage series screening
Prevalence 2/1000
Tests A & B, sensitivity .9, specificity .9
PPV 180/160 = 1.125%, 1 baby in 29 has PHL
Series protocol: Test A, if refer then Test B.
Protocol sensitivity 0.9, specificity 0.99
PPV 15.3%, > 1 baby in 7 has PHL
Moderate error correlations yield slight loss of sensitivity, still much improved specificity

Other best prevalence data
Wexner UNHS trial, Lancet 320:1957-64
prevalence (>40 ave, biat) 0.94/1000
Preve B, NY State UNHS, Ear Hear 00;21:85-91
prevalence (>20 any freq, unbl/b) 2.8/1000
Best overall estimates of congenital prevalence:
>40 dB, average, bilateral 0.8-1/1000
>20 dB, any frequency, any ear 2-3/1000

Must decrease false-positives, increase positive predictive value
Sensitivity .9, specificity .95, prevalence 2/1000
PPV 180/160 = 3.5%, 1 baby in 29 has PHL
Multi-stage screening: Test A, if refer => Test B
Series protocol sensitivity (PHL present)
~ sensitivity A, if FN errors strongly correlated
Series protocol specificity (PHL absent)
~ 1 - FPR A,FPR B, if FP errors poorly correlated

Best UNHS Outcome Data
Preve et al, Ear Hear 00;21:85 (NY State)
Typical protocol: two-stage, two technology
S1 (TEDAE->ABR) refer => S2 (TEDAE->ABR)
Refer rate: Year 1 3.7% Year 2 2.8%
Specificity: >96% >97%
PPV for PHL S1 4.5% S2 22.1%
Prevalence PHL 2.0/1000 adjusted 2.0/1000
**Practical issues underlying test performance**

*Choice of tests, e.g. missing neuropathies*

*Timing of tests, e.g. high FPR in first 24 hrs*

*Tester skills (e.g. access timing, baby state)*

*Actual test practices (e.g. multiple tests, turning baby, etc.)*

*Much variation possible. Skill, dedication critical.*

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**Effective program sensitivity**

*Program is a chain*

*Access for pre-discharge screen, Access for re-screen, Access for audiologic assessment, Access for follow-up services...*

*The net effective sensitivity is the product of all test sensitivities and all access deficits.*

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**Bottom lines**

*Test performance depends strongly on many factors, especially target disorder definition*

*Knowledge about sensitivity is limited. No definitive data, but convergent validity*

*Specificity is known and high-quality series protocols give high PPV & FPRs <2%, ? ~ 1%*

*Field screening performance depends on many small details of approach and procedure*

*Good screening is only the first step...*
POTENTIAL PITFALLS IN DETERMINING AUDITORY STATUS IN BEHAVIORAL AUDIOLOGIC ASSESSMENT

Dr. Judith S. Gravel, Hunter College of the City University of New York, New York, New York State, United States
Friday, January 24, 2003, 1:30 to 3:00 PM

Remarkable progress made in North America over the last several years towards the early identification, assessment and management of permanent childhood hearing loss. Recently, the Joint Committee on Infant Hearing (2000) recommended that medical and audiologic assessment of hearing loss be completed by 3-months of age in an infant identified as risk in the newborn period. The time line of identification by one month, confirmation by three months and intervention by six months has become the goal of early hearing detection and intervention (EHDI) programs.

Along with this long-desired opportunity for EHDI services delivery comes the challenge for audiologists of accurately determining the type, degree and configuration of the infant’s hearing loss. This allows the counseling of parents, medical intervention when appropriate, amplification selection and fitting, management and follow-up to proceed in a timely manner. A key in this process is the provision of comprehensive and on-going audiologic assessment. During the audiologic assessment, the auditory status of the infant is determined: information regarding threshold sensitivity is obtained across the speech frequency range and monitoring for change in auditory status (due to temporary or permanent changes in hearing sensitivity) is undertaken. As more is learned about the infant’s hearing loss, personal amplification devices, initially fit on somewhat limited data, are individualized.

As clinicians are learning rapidly, there are numerous ‘audiologic pitfalls’ that must be anticipated when evaluating infants and young children. These pitfalls can influence the accuracy of the diagnosis of the hearing loss, delay the initiation of appropriate management, and create stress and uncertainty in parents and caregivers. This presentation will address when to anticipate the unique audiologic pitfalls that arise in the audiologic evaluation of infants, particularly those relevant to the behavioral audiologic assessment. Where and why these pitfalls arise will be reviewed and ways to avoid them will be suggested. A comprehensive test battery approach will be encouraged. This approach encourages the audiologist to continually examine the accord among physiologic and behavioral measures of auditory status.
Potential Pitfalls in Determining Auditory Status in Behavioral Audiologic Assessment

Judith S. Gravel, Ph.D.
Hunter College of the City University of New York

Joint Committee on Infant Hearing
Year 2000 Position Statement:
Principles & Guidelines

- JCIIH Principles state that all infants who are referred from UNHS:
  - Begin appropriate audiologic and medical confirmation of the presence of hearing loss before 3 months of age
  - Those with confirmed permanent hearing loss receive intervention services before 6 months of age

Audiological Assessment of Infants

**Characterize:**
- Degree
- Type
- Configuration
  of any existing Hearing Loss

**Purpose:**
- Initiate appropriate intervention services
  - Selection of initial communication mode
  - HA fitting requires minimal low- to high-frequency information
  - Medical management
- Establish baseline for monitoring hearing over time

Audiological Assessment of Infants

- Two primary procedures for use in evaluating threshold sensitivity of infants
  - Electrophysiologic: ABR
  - Behavioral: Visual Reinforcement Audiometry (VRA)

Test Battery Approach

Numerous Pitfalls in Determining Auditory Status of Infants

Can seriously influence provision of timely & appropriate intervention

- 3rd USPSTF recommendation on NHS
- Recommended only targeted screening in 1995
- Revisited 1995 recommendation; asked:
  - Is widespread support for UNHS in the US currently justified?


- USPSTF undertook a review of the evidence on UNHS:
  - Published summary article simultaneously with the USPSTF Recommendation (Thompson et al., JAMA October 2001)
- USPSTF concluded:
  - “Insufficient evidence to recommend for or against routine screening of newborns for HL before hospital discharge”

Thompson et al., JAMA 2001
“UNHS: Summary of Evidence”

- Relative to audiological assessment
Thompson et al. state:
  - “Decisions about diagnosis and treatment [of HL] are made on the basis of a diagnostic ABR performed when the infant is 1 to 6 months of age.”
  - “Use of this diagnostic standard facilitates early intervention, but may overestimate the number of cases of permanent hearing loss (PHL).”

Thompson et al., JAMA 2001
“UNHS: Summary of Evidence”

- Cited the Wessex trial (Kennedy et al., 1999)
  - 1st audiological exam completed when infants were 6-12 weeks
  - Of 158 infants referred from screening, 27 diagnosed with PHL
  - Noted: “In 2 cases (7.4%), the diagnosis of PHL was wrong, and infants had normal hearing when examined at 4 or 10 months of age”

Thompson et al., JAMA 2001
“UNHS: Summary of Evidence”

- Also reviewed results of the RIHAP
  - Veur et al., 1998, J Pediatr 113:353-7
  - reported on outcomes of infants screened between 1993-1995.
  - USPSTF noted: “5 of 17 infants (29%) initially diagnosed with moderate SNHL later found to have mild SNHL.”

- Others have also reported that in some infants detected through UNHS, the initial diagnosis of degree of HL was different than final determination
  - Mason & Hermann (1998) and Watkin (1988) reported several cases of moderate & greater hearing loss diagnosed at initial ABR changed over follow-up tests to a definitive diagnosis of mild-moderate or mild
Thompson et al., JAMA 2001
“UNHS: Summary of Evidence”

- Report concluded:
  - “In expert hands, as many as 7% of infants diagnosed as having PHL may eventually prove to have normal hearing.”
  - “Frequency of misdiagnosis in everyday practice settings has not been studied.”

Thompson et al., JAMA 2001
“UNHS: Summary of Evidence”

- NYS NHS Demonstration Project (Daizell et al. 2000)
- Reasons for delay in audiological diagnosis
  - Mild hearing loss
  - Parental non-compliance with follow-up
  - Illness or developmental delay
  - “Audiological uncertainty”

NYS NHS Demonstration Project (Daizell et al. 2000)

- Potential Avoidable Pitfalls in Behavioral Audiological Assessment that could lead to Misdiagnosis of Hearing Loss
  - When to anticipate them
  - Why they happen
  - How to avoid them

Potentially Avoidable Pitfalls in Behavioral Audiological Assessment that could lead to Misdiagnosis of Hearing Loss

- Why should we worry about audiological misdiagnosis?
  - Delays in:
    - confirmation of true hearing status
    - medical referrals
    - referral for intervention
  - Inappropriate intervention
  - Parent anxiety, confusion, and lack of confidence in any subsequent recommendations
  - Unnecessary expenditure of resources: time, money, personnel & services
  - Reduced confidence in our professional capabilities by others (e.g., policy makers, administrators, medical practitioners, etc.)

Types of Audiological Assessment Pitfalls

- False-positive diagnosis
  - normal ears are incorrectly labeled as impaired

- False-negative diagnosis
  - impaired ears are incorrectly labeled as normal

- Misdiagnosis:
  - Correct diagnosis of auditory disorder; incorrect diagnosis of type or degree
False-negative diagnosis (hearing loss diagnosed as normal hearing)

- Behavioral Assessment PITFALLS:
  - Infant conditioned with inaudible stimulus
  - Failure to:
    - Use one examiner
    - Include control silent test trials
    - Follow a specific test protocol
    - Fail to confirm results with physiologic measures (AMEMR, OAE, ABR)

False-negative diagnosis

Berger & Hood, 1993
"False central HL" J Am Acad Audiol 4: 215-223

- Infants referred 7 months and diagnosed having severe-to-profound hearing loss
  - Little progress made when fit with amplification
- Behavioral observation audiometry and SF VRA consistent responses at 25-35 dB HL
  - Aided responses were higher than non-aided
  - Determined to have central processing disorder

False-negative diagnosis

Berger & Hood, 1993
"False central HL" J Am Acad Audiol 4: 215-223

- 6-mo-old infant demonstrated severe SNHL using VRA, referred for ABR
- ABR interpreted as "normal"
- Following ABR (interpreted as "normal"), every subsequent behavioral test result was also normal
- Conclusion: Normal hearing; not aided; lack of speech and language development diagnosed as verbal auditory agnosia (VAA) "central deafness"

False-negative diagnosis (SNHL diagnosed as normal hearing)

- Reality: At 4 years, severe-to-profound SNHL (same as at 6 months)
  - Repeat ABR consistent with SNHL of same degree

- PITFALL: Inappropriate interpretation of ABR waveforms
  - Failure to replicate responses
  - Failure to complete full ABR protocol (threshold and supra-threshold assessment)

- PITFALL: Symptom only; neither AMEMR or OAE completed

False-negative diagnosis

SNHL diagnosed as normal hearing

- Reality: At 4 years, severe-to-profound SNHL (same as at 6 months)
  - Repeat ABR consistent with SNHL of same degree

- PITFALL: Inappropriate interpretation of ABR waveforms
  - Failure to replicate responses
  - Failure to complete full ABR protocol (threshold and supra-threshold assessment)

- PITFALL: Symptom only; neither AMEMR or OAE completed

Misdiagnosis:

(degree of HL diagnosed as different than actual)

- Behavioral Assessment Pitfalls
  - Use of sound field testing only
    - Failure to diagnose unilateral HL
    - Failure to diagnose asymmetrical HL
    - Failure to diagnose conductive or mixed HL
  - Failure to diagnose high-frequency HL when only speech awareness levels are determined
  - Misdiagnosis of profound HL when significant residual hearing exists
Misdiagnosis:
(degree of SNHL diagnosed as greater than actual)
- Reality: moderate HL sloping to severe-profound SNHL; residual hearing across speech-frequency range
- Testing PITFALLS: Failure to obtain:
  - FS-ABR
  - Ear-specific behavioral responses
Critical considerations as the age of cochlear implantation decreases

False-positive diagnosis
(normal hearing diagnosed as HL)
- Behavioral Assessment PITFALLS:
  - Elevated Minimal Response Levels (MRLs) due to unreliable behavioral responses resulting from:
    - Inattention (poor task orientation),
    - Response habituation,
    - General malaise,
    - Developmental disorder, or a
    - Developmental level inappropriate for the test procedure

False-negative diagnosis
(Normal hearing based on physiological tests alone)
- Air-conduction threshold ABR completed – normal bilaterally
- Tympanograms normal
- Conclusion: Normal hearing
- Reality: Cortical damage secondary to perinatal asphyxia (revealed with CAEP)
- Behavioral PITFALL: Failure to observe behavioral responses to sound; reliance only on physiological tests

False-positive diagnosis
(Conductive HL diagnosed as SNHL)
- Infant 6 months of age: ongoing audiological testing following referral from UNHS
  - AC-ABR thresholds elevated: moderate-severe range
  - OAEs absent:
    - Conclusion: SNHL
  - Reality: Conductive HL associated with MEE
- PITFALL:
  - Reliance on only ABR
  - Failure to record BC-ABR
- Behavioral PITFALL: Failure to assess behavioral thresholds (AC & BC)
**Behavioral Test Methods**

- **Unconditioned response techniques**
  - Behavioral Observation Audiometry (BOA)
  - Observation of auditory behaviors
- **Conditioned response procedures**
  - Visual Reinforcement Audiometry (VRA)
  - Conditioned Orienting Response (COR)
  - Conditioned Play

**Auditory Response Index**

<table>
<thead>
<tr>
<th>NM</th>
<th>FM Tone</th>
<th>Speech</th>
<th>Resp</th>
<th>Startle</th>
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<tr>
<td>0-6 wk</td>
<td>50-70</td>
<td>75</td>
<td>40-60</td>
<td>Eye-widen, blink, startle, etc.</td>
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<td>4-7 mos</td>
<td>40-50</td>
<td>50</td>
<td>20</td>
<td>Head turn, listens</td>
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**Limitations of Behavioral Observation Audiometry**

- Biased observers often used.
- Numerous behaviors accepted as response indicators.
- Age and developmental level of the infant/child strongly influence test results.
- Response probability dependent on infant state, the nature of the stimulus, the ambient noise level, and the agreement among two or more examiners.
- Infants with normal hearing show wide variability in responsiveness.

Wilson & Thompson, 1984

**Observation of Auditory Behaviors**

- Behavioral tests may have limited value for quantifying hearing sensitivity.
- Behavioral observations are useful in gaining insight into auditory function - the quality of the infant's or child's auditory responses.
- Behavioral orienting responses from young infants may provide insight into neuro-development.
- Observations should always be used regardless of outcome of electrophysiologic tests.

Diefendorf & Gravel, 1985

**Visual Reinforcement Audiometry**

- VRA is an operant discrimination procedure.
- Stimulus (test signal) cues infant that a response will result in (visual) reinforcement.
- Operant behavior (head turn) is increased by the application of reinforcement.
- Audible signals have little or no reinforcing value; therefore, best to use a procedure in which the signal & reinforcer are separate.

Diefendorf & Gravel, 1985

BOA should **not** be used to:
- Screen hearing
- Estimate hearing thresholds
- Define the settings of hearing aids
**Problem with Classical Conditioning:**
VRA in the clinical setting

- Assume infant can detect the stimulus used during the conditioning phase
- Numerous trials (signal and control) to reach criterion for test initiation

**Factors in VRA**

**Visual Reinforcement**

- Use of Animation—Novelty
  - Primus, JSHR—1987
- Reinforcement Duration
  - Culloppe & Thompson, Ear & Hearing—1984
- Reinforcement Schedule
  - Primus JSHR—1985

**Visual Reinforcers**

- Dark smoked-Plexiglas compartments:
  - Completely out of sight except during periods of reinforcement
- Animated, colorful, brightly illuminated
- Multiple reinforcers: increase novelty, support responding over repeated trials

**Factors in VRA**

**Use of Distraction**

- Decreases the number of false positive responses
- Reduces the duration of inter-trial intervals
- Prolongs the period of time during which the infant’s behavior is appropriate for testing

**Factors in VRA**

**Test Arrangement**

- Infant seated on parent’s lap
  - Audiologist located in test booth, sits opposite (audiometer located within test booth)
  - Audiologist controls distraction, presentation of test signals & delivery of reinforcement
**North American Conference on Deafness Screening and Intervention in Early Childhood**

**ABSTRACTS AND POWERPOINTS**

---

**VRA Protocol**
- Two audiologists; one controls test from outside test booth; second inside booth with infant and serves as 'distractor'
- Infant seated in highchair

**VRA and Developmental Age**
Widen, 1990

- Percent Successful VRA Tests

<table>
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<tr>
<th>Developmental Age (BSID-MA) in Months</th>
<th>0</th>
<th>20</th>
<th>53</th>
<th>79</th>
<th>80</th>
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**VRA and Age (Corrected for Prematurity)**
Widen, 1990

- Percent Successful VRA Tests

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<th>Corrected Age in Months</th>
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*Gravel & Wallace, 2000*

"Effects of OME on hearing in the first 3 years of life"* JSHR, 2000*

- Prospective follow-up: 114 children through first 3 years of life
- Success rate for computer-assisted VRA
- 7-, 10-, and 12-month-olds
  - 82% of infants provided 4-frequency audiogram using computer-assisted method
- 5-month-olds
  - 49% tested successfully tested with computer-assisted VRA
Widen et al., 2000
“Hearing status at 8-12 months CA using VRA protocol” Ear Hear 21: 471-477

- Specified a stringent VRA protocol incorporating signal and control trials
- 3134 infants (NICU or WBBN with risk factors)
- 95.6% of infants (N = 2995) successfully conditioned
- 50% completed full VRA protocol; 8 minimal response levels; 4 in each ear

- Of 139 (4.4%) babies CNT:
  - 20% developmental delays/visual impairments;
  - 80% consistently unreliable or failed to return after unsuccessful 1st test.

Avoiding VRA Pitfalls

- Obtain ear-specific responses
- Obtain BC-VRA responses
- Use frequency-specific stimuli (be aware of the potential pitfall of NBN)

Day, Bamford et al., 2000
“Evidence of the efficacy of insert earphone and SF VRA with young infants” Ear

- VRA performed on 41 typically-developing infants aged 5 months to 10.5 months
- Infants tested in SF (n=22) and with insert earphones (n=19)
- Results: significantly more minimal response levels (MRLs) obtained for SF testing and with older infants
- Insert earphone:
  - 36% aged ≥ 6 months ≥ 2 MRL
  - 25% aged ≤ 6 months ≥ 1 MRL

Earphones

- Younger infants retain earphones better than older (Nozza & Wilson, 1994; Gravel & Traquina, 1992).
- Most difficult age range is 18 - 24 months
- Insert earphones better than TDH-series; e.g., comfort, movement... and importantly for hearing aid selection.
- Thresholds must be corrected for ER-3A versus TDH-series differences

Identification of Neonatal Hearing Impairment:
Hearing Status at 8 to 12 months CA using VRA

Widen et al. 2000

- 95.6% of infants (n = 2095) successfully conditioned;
  - many (not all) successfully completed full protocol:
  - thresholds; 4 MRLs in each ear.
- Of 139 (4.4%) babies CNT:
  - 20% developmental delays/visual impairments;
  - 80% consistently unreliable or failed to return after unsuccessful 1st test.
Normal Hearing in Infants (6 to 12 months)
Visual Reinforcement Audiometry: ER-3A Insert Earphones
- Thresholds for infants with normal hearing within 10-15 dB of adults (Nozza & Wilson, 1984)
- Non-sensory factors (intensity-generated noise and attention/motivation) rather than sensory factors account for most of the difference (Nozza & Henson, 1999).
- Likely thresholds at 500 Hz = 20 dB HL, 1000 Hz = 15 dB HL (Nozza, 2002)

Intelligent VRA: IVRA (IHS, Miami, FL)
- Computer-assisted, single examiner, bias-free procedure
- 4-button hand-held response box interfaces with computer.
- 4 complex visual reinforcers randomly activated for correct responses.
- Quantifies false-positive rate and attention & motivation during test.
- Standard, optimized algorithm, screening

"Teaching a baby the head-turn response for VRA is easy.
The hard part is teaching the baby not to turn" (Nozza, 1999)
- Critical to evaluate (quantify) the false-positive response rate in order to determine the validity of the behavioral assessment.
- Control trials (silent test intervals) must be included in the behavioral assessment.

Conditioned Orienting Response or 'Reflex' (COR) Audiometry
- Two loudspeakers located to the right and left of child.
- Correct response behavior is a head turn towards signal side.
- Symmetrical hearing is 'assumed' if the infant performs task correctly.
- Infant must detect and then localize (orient) to the correct side.
- COR is not the same as VRA.

PITFALL: Undiagnosed OME
Percent of Infants (n = 2268) with MEE by Residence Location and Age

Middle Ear Effusion and Audiological Assessment
- In some infants, ABR may overestimate the degree of hearing loss when MEE present.
- Delaying ABR because of MEE can result in late ID of SNHL (Davis et al.)
- BC-ABR critical early in diagnostic assessment when AC-ABR elevated.
Recommendations of the UK Working Group on Follow-up Assessment
(Sutton et al. 2000)
- High frequency tympanometry (660 Hz & 1000 Hz) appears to be valid (i.e. reasonably sensitive and specific) in detecting MEE below 4 months (Shanks et al. 1977, Marchant et al. 1984, McKinley et al. 1997, SalzWet 2000)
- Tympanometry (single-component admittance) should be included in the audiological test battery regardless of the age of the infant.
- 220 Hz for < 4 months should not be used
- AMEMR – use probe frequencies > 220 Hz

2-Year-Olds
- Primus & Thompson (1985)
  - No difference in rate of conditioning or response reliability between 1- and 2-year olds
  - BUT… 1-year olds provided 50% MORE responses than 2-year-olds using same response protocols

2-year-olds
Reinforcement | # of Responses
--------------|---------------
Initial       | Post-break    
1 yr - 1 VR   | 26.9          | 8.8         
1 yr - 2 VR   | 33.7          | 16.7        
2 yr - 2 VR   | 14.6          | 2.3         

Thompson, Thompson & McCall, 1992

2-Year-Olds
- 31 toddlers 24-months (+/- mo.) tested with both VRA and Play
  - Provided at least one threshold:
    - VRA 45%
    - Play 84%

Olsen, 1994

2-Year-Olds
- Play Audiometry (provided at least one threshold)
  - 16-23 months 45%
  - 24-29 months 63%
  - 30-35 months 83%

Nielsen & Olsen, 2001

TROCA/VROCA
- Tangible/Visual Reinforcement Operant Conditioning Audiometry
- Technique not often used clinically but may be very useful for infants ‘too old for VRA and too young for conditioned play audiometry’
- Classic TROCA/VROCA, the auditory stimulus is paired with a light on the response box located in front of the child
TROCA/VROCA
- Child conditioned to push response button when the light/signal are present.
- Visual reinforcer activated or tangible reinforcer is dispensed.
- Light is faded on successive trials until only the auditory signal is presented. If the child responds to the auditory signal, the response is reinforced. If not signal level is raised (e.g., 20 dB) and light and signal are paired again.

2-year-olds

<table>
<thead>
<tr>
<th>Test Type</th>
<th># Resp</th>
<th>Failed conditioning</th>
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</thead>
<tbody>
<tr>
<td>VRA</td>
<td>11.4</td>
<td>0% (0/15)</td>
</tr>
<tr>
<td>PLAY</td>
<td>28.3</td>
<td>32% (7/23)</td>
</tr>
<tr>
<td>VROCA</td>
<td>21.4</td>
<td>17% (3/18)</td>
</tr>
</tbody>
</table>

Thompson, Thompson & Vetivelu, 1989

Avoiding VRA Pitfalls
- Specify a VRA protocol & follow it.
- Obtain ear-specific & frequency-specific responses.
- Include control (silent) trials.
- Evaluate (quantify) the false-positive response rate to determine the reliability of assessment.
  - Add more ‘objectivity’ to VRA (Maden, 1993).
  - Specify signal & control trials.
  - Computer-assisted VRA procedures.

Avoiding Audiological Pitfalls in the Assessment of Infants & Young Children
- Follow test protocols that are evidence-based and time efficient.
- Always provide comprehensive audiological evaluation at the initial referral.
- Repeat measures wherever there is question of diagnosis.
- Complete full battery of behavioral & physiologic test procedures (cross-check).
  - No test is redundant; all provide important information regarding auditory integrity.
  - ACCORD among test results must be evident.

“Audiologists should have experience with the assessment of infants & children with HL and the knowledge and equipment necessary for use with current pediatric assessment methods”

“Facilities that lack the expertise or equipment for assessing infants & children should establish consultative arrangements with those that do”.

Pediatric Working Group, 1996
ACQUIRED HEARING LOSS IN CHILDREN

Dr Michel Picard, École d’orthophonie et d’audiologie, Université de Montréal, Québec, Canada

Friday, January 24, 2003, 4:00 to 4:30 PM

The prevalence of acquired irreversible bilateral hearing loss to a degree greater than 40 dB increases with age, especially between three and nine years of age. From 1.06 cases per 1000 births by age three, this increases to 2.05 cases per 1000 births by age nine. This trend appears to have two leading causes. First, the effects of progressive hearing loss of genetic origin seem to delay the diagnosis of the problem because of a history of natural sensory loss occurring at a later age. The second cause seems to be related to the fact that lesser degrees of hearing loss are detected at later ages: in Germany, light deafness is diagnosed at age six, moderate deafness at age four, severe deafness at age two and a half, and profound deafness at just under two years of age. Advances in neonatal medicine have contributed to a decrease in newborn mortality (including the severely premature), but have been linked to an increased prevalence of acquired deafness. On another level, certain ethnic groups that have emigrated to heavily industrialized western countries are being singled out because they seem more prone to manifestations of significant deafness during childhood. These groups include people from Pakistan, Asia, Cuba, Puerto Rico, and Mexico.

It is estimated that in the United-States, at least one school-aged child out of six (16.6%) has irreversible hearing loss either from birth or from endogenous causes. Added to this is occupational hearing loss, which has been documented in children in the US since the late Sixties, and which is on the rise because of a universal increase in noisy leisure activities. Loud music in all its forms is increasingly well documented as a propagation vector. In the US, exposure to firearm and firework detonations appears to be a more important vector than loud music. Participation in noisy sport activities, such as watching car races or driving race cars, is frequently part of the history of exposure. Adolescents engaged in farm work face a special risk. Because of cultural factors, boys are affected by deafness in greater numbers than girls (9:1). Occupational deafness is more prevalent in areas with high levels of poverty, in rural areas, and in the southern and western parts of the US.
The majority of new cases of deafness are found in adolescents (12-19 years), but 26% of diagnoses concern children aged ten years or less. Boys represent the majority of these cases, owing to the fact that they are more inclined to participate in noisy activities with a parent (motorcycle and snowmobile riding, use of noisy tools, exposure to firearm detonations, etc.). Occupational deafness in the US is estimated to affect 12.5% of children and adolescents aged from 6 to 19 years. There is no reason to believe the situation is any different in Canada. All things considered, acquired hearing loss would seem to affect one out of three children.
Acquired hearing loss in children

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Université de Montréal
michel.picard@umontreal.ca

Introduction

How relevant is the epidemiology of acquired hearing loss in children to screening and intervention?

Epidemiological studies estimating the prevalence of acquired hearing loss in children at various ages are of paramount importance to set priorities for prevention and treatment (Farbey, 1992).

Changing epidemiology of acquired hearing loss in children

- Acquired hearing loss in children is on the rise between ages 3 and 9 years
- A conservative estimate of the prevalence of permanent bilateral hearing loss in excess of 40 dB is as follows:
  - 0.81% of 1000 children (age 0-3 years),
  - 1.22% of 1000 at age 5,
  - 1.32% of 1000 at age 6,
  - 1.66% of 1000 at age 7,
  - 1.55% of 1000 at age 8,
  - 1.55% of 1000 from 3 years on.
- (from Porrum et al., 2001)

Changing epidemiology of acquired hearing loss in children

- Current estimate of hearing loss (mono- or bilateral)
  1. Adams & Kresen (1992) estimate acquired hearing loss in children to be as high as 16.4% in the United States
  2. Niskar et al. (1995) found 14.9% of hearing loss breaking down as follows:
    - 12.7% with only high-tone loss (3, 4, 6 kHz),
    - 7.1% with only low-tone loss (3, 4, 6 kHz),
    - 4.9% with both high- and low-frequency loss

Changing epidemiology of acquired hearing loss in children

Population characteristics:

- High-tone loss was more frequent in boys
- Socio-economic variables:
  - Hearing loss more frequent in children from low socio-economic groups
- Ethnic origin:
  - Greater risk for children of Mexican origin living in the United States

Known causes

1. Delayed identification as a result of progressive hearing loss of genetic origin
2. The pervasive problem of early identification, especially hearing losses of a milder degree:
   Reporting on the Boston Registry for hearing loss in children for period 1996-98, Fishkin-Kramer et al. (2004) determined diagnoses of mild hearing loss at 6/2 years on average, moderate ones at 4/2, severe and profound ones at 2/3 and 1/9 years, respectively
Changing epidemiology of acquired hearing loss in children

Known causes (cont.)

- Improved perinatal medical care results in more surviving infants with perinatal complications (Strollo, et al., 1999). In particular, reduced mortality of preemies with very low birthweight. Similar age results in more infants with hearing loss (Cano, et al., 2001).

Epidemiology of acquired hearing loss in children: future trends

- Prevalence is high worldwide with industrial status of the country a hearing-related covariate.
- Prevalence of permanent bilateral hearing loss worldwide (Davis & Hind, 1995):
  - More developed countries: 4.81%
  - Less developed countries: 32.71%
  - Africa: 2.33%
  - North America: 8.8%
  - Latin America: 2.0%
  - Oceania: 0.11%
  - Asia: 47.1%

Epidemiology of acquired hearing loss in children: future trends

- Point prevalence is likely to increase: immigration as a hearing-related covariate (cont.)
- Immigration to highly industrialized countries brings a greater risk:
  - In Pakistan and Egypt, a greater risk of severe to profound hearing loss of genetic origin.
  - In Asia children are 2.4 to 3.6 times at greater risk of having permanent hearing loss.

Epidemiology of acquired hearing loss in children: an unwittingly newcomer

- Noise-induced hearing loss in children:
  - Since the early 70’s, 3-6 dB hearing loss for high tones most sensitive to noise damage has been reported in boys starting at age 12 (Roberts & Atunga, 1975).
  - Noise-induced hearing loss in children was acknowledged in 1959 by NIH.
  - Blackman (1992) found 5% of children with noise-induced hearing loss in a group of 2284 consecutive patients under age 20 attending an ENT clinic; there were more boys than girls (85%) and most importantly, 25% of cohort were under 11 y.o.

Epidemiology of acquired hearing loss in children: an unwittingly newcomer

- Noise-induced hearing loss in children (cont.):
  - Broste, et al. (1989) have found doubling the incidence of noise-induced hearing loss in teens actively involved in farm work (most of them were boys) than other activities.
  - Prevalence of noise-induced in 15-19 year olds is currently estimated at 12.5% in the United States (Niskar, et al., 2001).
Epidemiology of acquired hearing loss in children: an unwittingly newcomer

- Noise-induced hearing loss would result from noisy leisure activities...
  - Exposure to loud music (ear-level listening devices and rock concerts) is a pervasive cause; damage risk criteria for these two activities is estimated respectively, at 7 and 2 b/rack (Meyer-Bisch, 1996); still, at least 22% of college and university students use ear-level personal devices at a pace of 5 b/rack and 5% of cohorts report attending rock concerts for 3 b/rack; this adds up to the 7 b/rack of loud music delivered through speakers acknowledged by 83% of students (Chossian et coll., 2001)

- In the Northwest of the United States – and possibly Canada, exposure to gun fire and firecrackers is the prevailing cause of noise-induced hearing loss in children being responsible for 45% of reported cases compared to 12% for those listening to loud music (Brookhauzer, 1992)
- Among children 18 years and under, participating to noisy leisure activities with a parent is the leading cause of exposure (Brookhauzer, 1992)

Conclusions

1. Without considering causes, acquired permanent hearing loss in children has a prevalence of 27.4% and is more a concern for boys; it is more prevalent in low socio-economic groups
2. 12.5% of these losses are attributable to significant exposure resulting from participating in noisy leisure activities

Conclusions

3. Identification of acquired permanent hearing loss is delayed because of current limitations in identifying mild to moderate degree of impairment
4. Improvement of medical practices including neonatal care contributes to the increase in congenital and acquired hearing loss in children
5. Socio-economic and industrial factors increase the prevalence of congenital and acquired hearing loss in children
6. Immigration brings increased risk

Severity of consequences associated with acquired hearing loss in school-age children

- Studying academic achievement of 1230 3rd, 6th and 9th graders, Boss et al. (1998) found that:
  - 5.4% presented undetected permanent hearing loss including monaural and just noticeable bilateral loss
  - Among those ‘unnoticed’ cases, 37% repeated at least one year of school

Canadian Academy of Audiology
Institut national de santé publique du Québec
Ordre des orthophonistes et audiologistes du Québec
North American Conference on Deafness Screening and Intervention in Early Childhood

References on « prevalence »


References on « prevalence »


References on « prevalence »


Reference on "academic failure"

Bess, F.H., Dodd-Murphy, J., Parker, R.A.  

5.4% of just noticeable hearing losses in the school population go undetected although associated with a high risk of failing a degree.
THE USER’S PERSPECTIVE

Marc Choquette, Quebec’s association for children with hearing problems (AQEPA)
Nicole De Rouin, Quebec’s association for children with hearing problems (AQEPA)

Friday, January 24, 2003, 4:30 to 5:00 PM

The Quebec Association for Children with Hearing Impairment is a provincial non-governmental organization that was established in 1969. It has almost 600 members, most of whom are parents of hearing-impaired children. The Association’s mission is to promote and develop all the services needed for the social integration of young people with hearing impairment.

The Association works to meet the following objectives:

- To promote the dissemination of information and to support the parents of hearing-impaired children.
- To promote early screening, early childhood intervention and use of appropriate amplification devices.
- To promote parents’ and children’s rights to take full part in decisions relating to them.
- To act as advocate for parents and children in their dealings with authorities in order to foster the development of resources that better meet the needs of hearing-impaired children.

In December 1999, a committee for “adaptation-readaptation” was created in order to identify strategies to help prepare hearing-impaired children for school. Why are our children not ready for their first year in school? A final report, tabled in May 2002, contains several recommendations. Among other things, it calls for universal neonatal hearing screening, early diagnosis and fitting of appropriate amplification devices and intensive early childhood readaptation.

We understand that the family plays a primordial role in children’s development. We wish to learn how to communicate with them as soon as possible. Why not give hearing-impaired children the tools they need to become fully functioning members of society, and to use their differences and talents to help make it better?
The PowerPoint is not available.