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
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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

Site	Rate
Well Baby Nursery	1 per 1000
NICU	10 per 1000
Total population	2-4 per 1000
# infants ident annually US	8,000-16,000
Average career pediatrician	12 patients

How Does Permanent HL Compare to Other Screenable Newborn Disorders ?

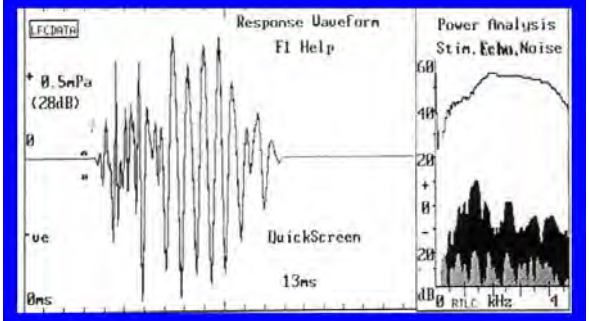
Sickle cell disease*	47 per 100,000
Hypothyroidism*	28 per 100,000
PKU*	2 per 100,000
Adrenal hyperplasia*	2 per 100,000
Permanent HL	220 per 100,000

*Texas Dept. of Health

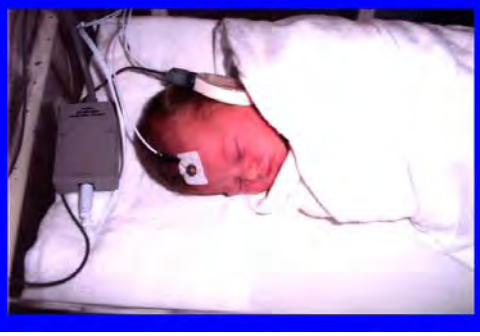
Current Methods Available

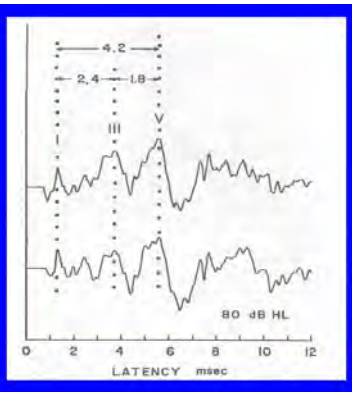
Transient Evoked Otoacoustic Emissions	TEOAE or DPOAE
Automated Auditory Brainstem Response	AABR
Standard Auditory Brainstem Response	ABR
2 Step	OAE + AABR

OAE



AABR





Rate per 1000 of Permanent Childhood Hearing Loss in UNHS Programs

Site	Sample Size	Prevalence Per 1000
Rhode Island 3/93-6/94	16,395	1.71
Colorado 1/92 – 12/96	41,976	2.56
New York (1/95-12/97)	69,761	1.95
Texas (1/94 – 6/97)	52,508	2.15
Hawaii (1/96 – 12/96)	9,605	4.15
New Jersey (1/93-12/95)	15,749	3.30

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 sev to profound (14 vs 26 m)

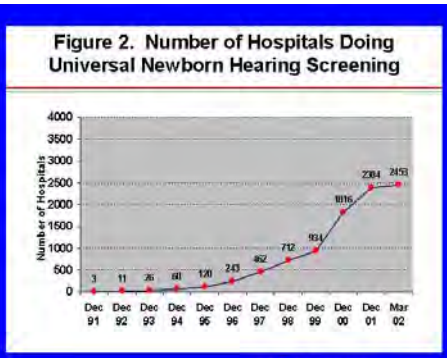
Apizzo - better language scores at age 4 if ident ≤2 m

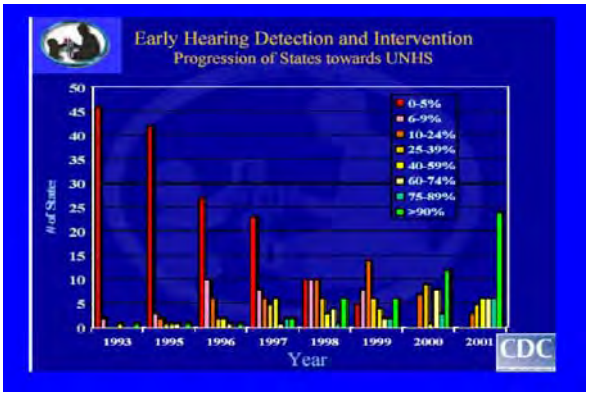
Moeller - 100 D/HH children with early ident – better outcomes

Yoshinago-Itano - 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

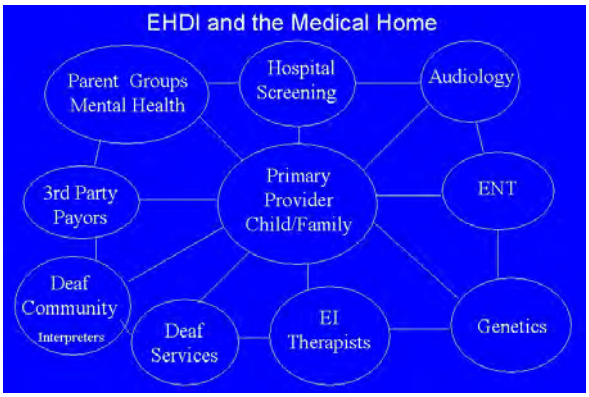
- Universal Newborn Hearing Screening< 1 m
- Effective Tracking and Follow-up as a part of the Public Health System
- Appropriate and Timely Diagnosis of the HL < 3m
- Prompt Enrollment in Appropriate EI < 6m
- All infants will have a medical home

Components of A Medical Home for EHDI includes Family Care that is:

accessible
family centered
comprehensive
continuous
coordinated
compassionate
culturally competent

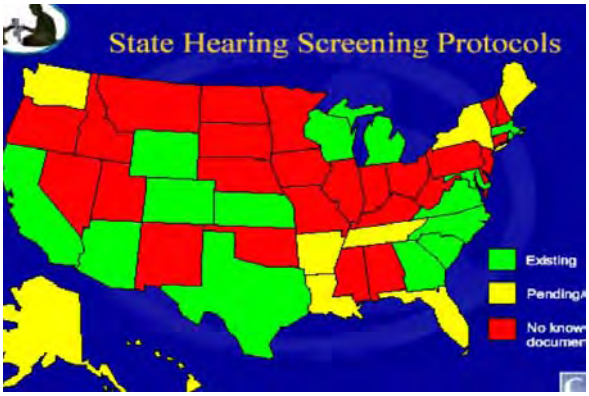
How Does the Concept at the Medical Home Become An Effective Working Model

- 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



How Can These Linkages Be Made Seamless?

- Development of Assessment Protocols
- On going Research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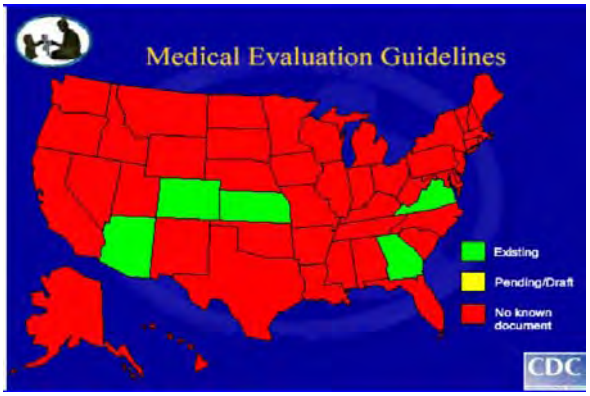
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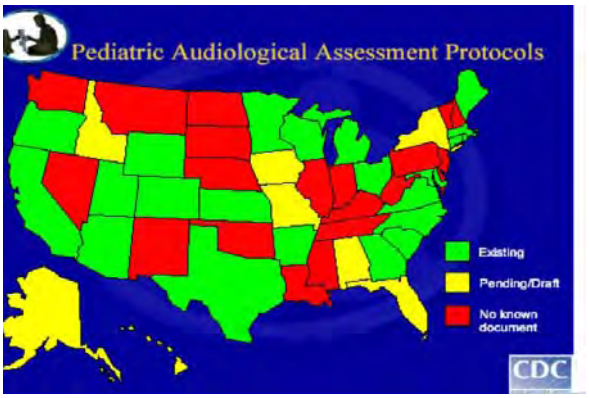
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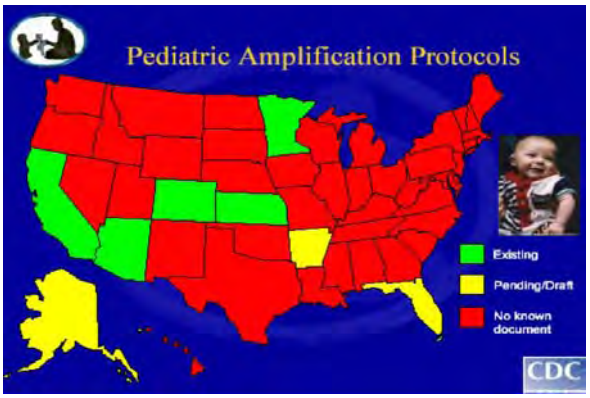
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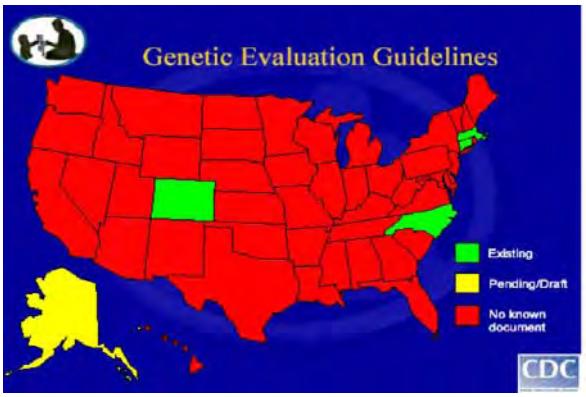
Genetic Evaluation Guidelines
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Alaska
Arizona
Arkansas
Colorado
Delaware
Florida
Georgia
Hawaii
Illinois
Iowa
Kentucky
Louisiana
Maine
Massachusetts
Michigan
Minnesota
Missouri
New Hampshire
New Jersey
North Carolina
Oregon
Rhode Island
South Dakota
Tennessee
Utah
Vermont
Wisconsin
Wyoming

The Medical Workup

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

Causes of Permanent Hearing Loss in 100 Infants

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

Genetic Causes

Single gene	Connexin 26
Gene + environment	Mitochondrial + ototoxic
Gene + gene	Gene + other gene

Most Common Environmental Causes

CMV	Rubella
Meningitis	asphyxia
Ototoxic meds	prematurity

Early Intervention Birth to 3

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

Rhode Island - 7 Birthing Hospitals

1 Tertiary Care Center (WIH) - 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
14,00 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 inc: 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	>99% screened
1st stage (TEOAE/AABR) fail rate	≤4%
Rescreen return rate	>90%
Dx procedures on refers	>90%
Referrals for intervention services	<6m

"KIDSNET"

A Data Management System for Universal Screening Vital Records

Level 1 - Metabolic - Hearing

Lead - Immunizations - WIC

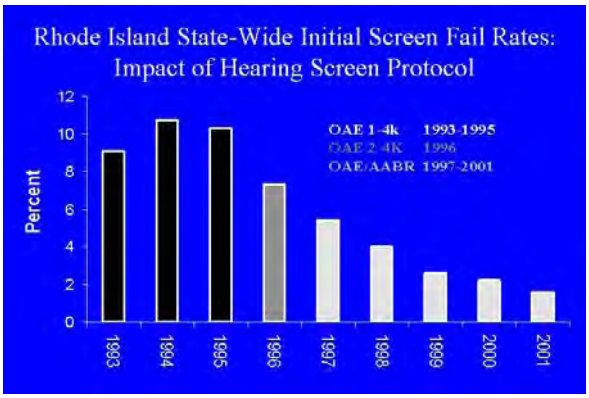
VNA - EI - Primary Provider

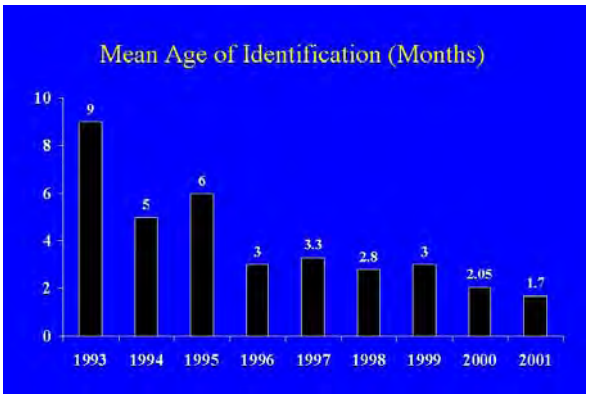
Medical Home "First Connections" Task Force

Dr. E. Kurtzer White HRSA#

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

RIHAP Quality Improvement





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	F
3 days	ABR	85 dB	85 dB

Fam Hx - grandfather - Menieres, father - lifelong mild HL

Pediatrician refers to to ped neurology

2m neurologist report - hearing N in at least 1 ear, FU not needed

Hailey Follow-up

6 m	Ped calls RIHAP, requests statistics on HL, says family not convinced Hailey is hearing	
	Given a referral to an Audiologist	
7 m	Diagnostic ABR	bilat sev prof
9 m	2nd diagnostic ABR	bilat sev prof → EI
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 EI
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

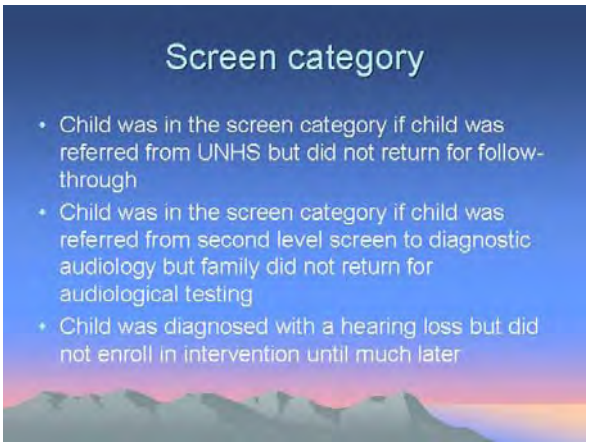
Thursday, January 23, 2003, 10:30 AM to 12:00 PM

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

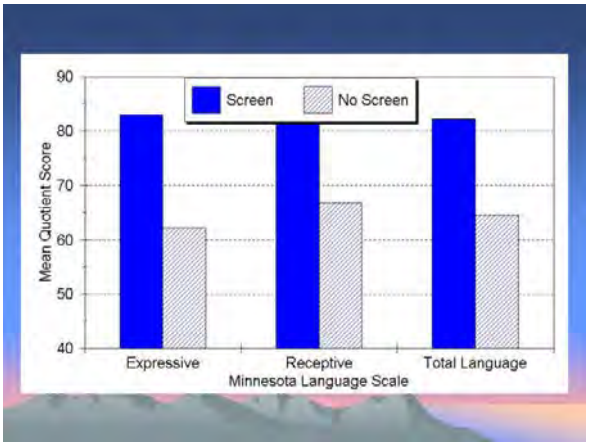
- Matched pair design
- CA= 9 to 61 months (Mean=30.5)
- CQ=98.9
- Matched by gender, ethnicity, presence of additional disabilities, educational level of the caretaker, and mode of communication

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 6 months of age
- No Screen group: 25th %ile identified by 13.5 months, median by 23 months and 75th %ile by 30 months

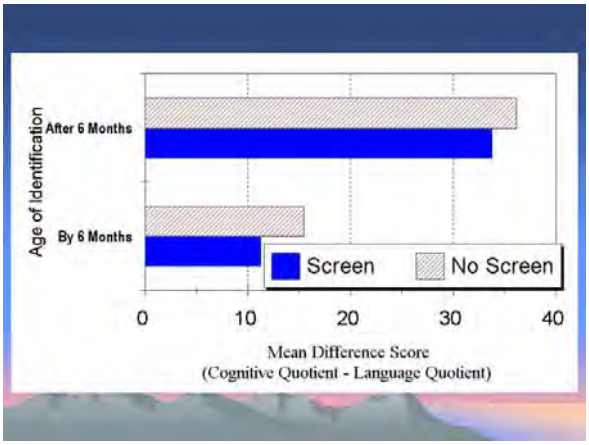
Screen vs. No Screen

- 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



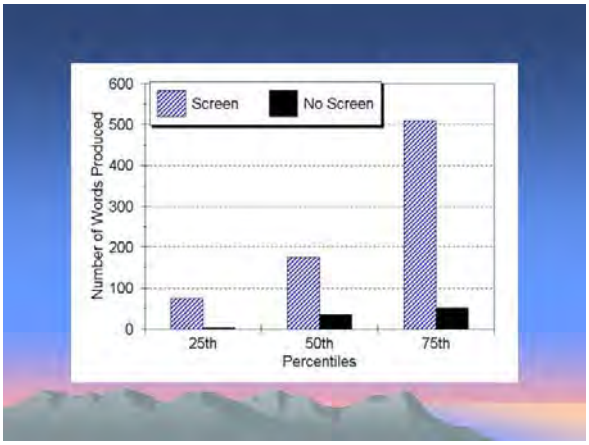
Non-verbal cognitive and language development

- 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



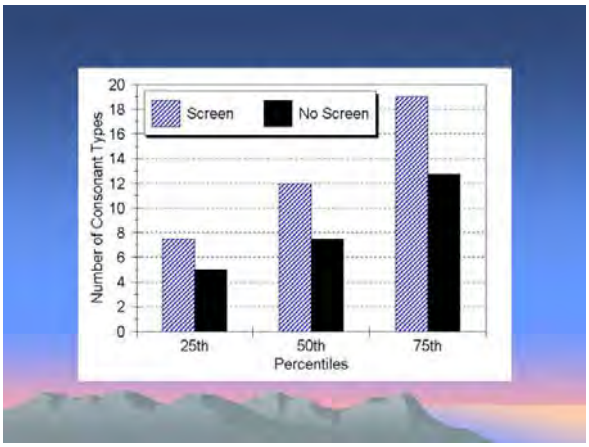
Screen vs. No Screen

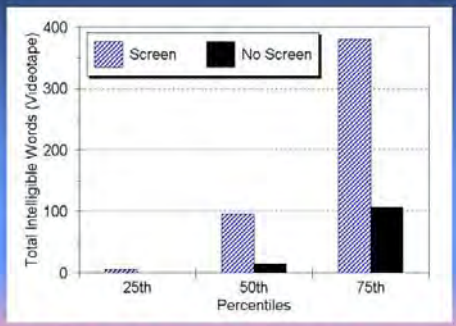
- Vocabulary development
- Children in the Screen group had significantly higher vocabulary levels
- A child at the 75th %ile of the No Screen group had fewer words than a child at the 25th %ile of the Screen Group
- Significant differences (p<.001)



Speech results: Screen/No screen

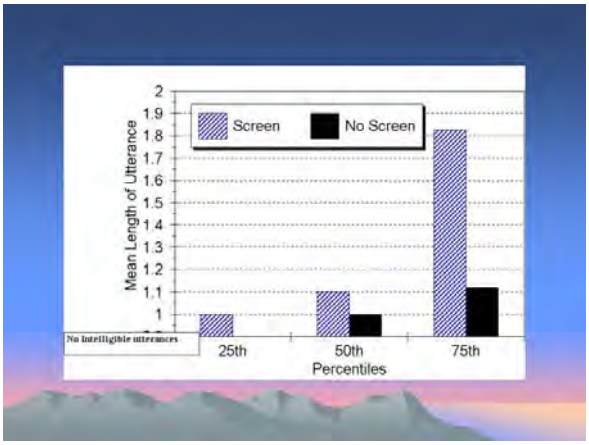
- Children in the screen group had significantly more vowels (p<.001)
- Children in the screen group had significantly more consonants (p<.001)
- Children in the screen group had significantly more initial and final blends. No children in the "no screen group had blends





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

- 25%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
- 75%ile of "screen" group was always or almost always understand

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

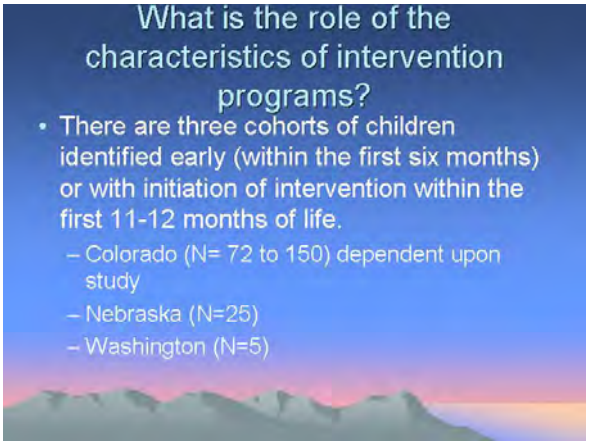
- Tharpe, 1999
- Study compared false positive emotional response to control group of true positives
- No significant difference in percent of families reporting negative responses

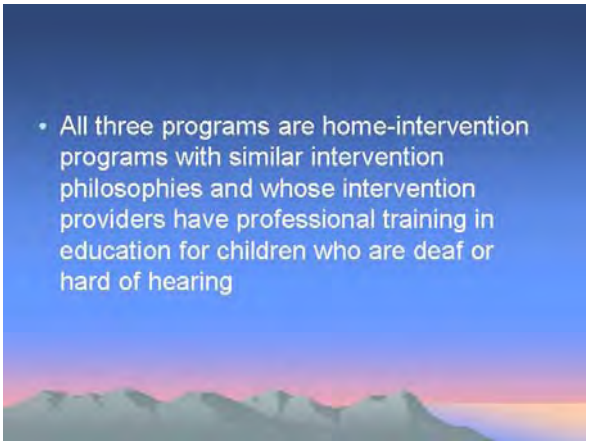
Parental Harm

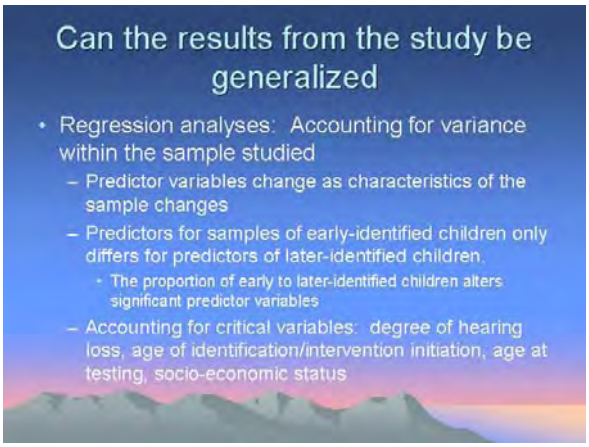
- True negatives
- Early-identified population have higher language levels which are highly related to:
  - Better Emotional availability (Pipp-Siegel, Pressman, Yoshinaga-Itano, 1999)
  - Lower Parental stress (Pipp-Siegel & Yoshinaga-Itano, 1999)
  - Lower Parental depression (Siegel, 1999) Lower Parental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

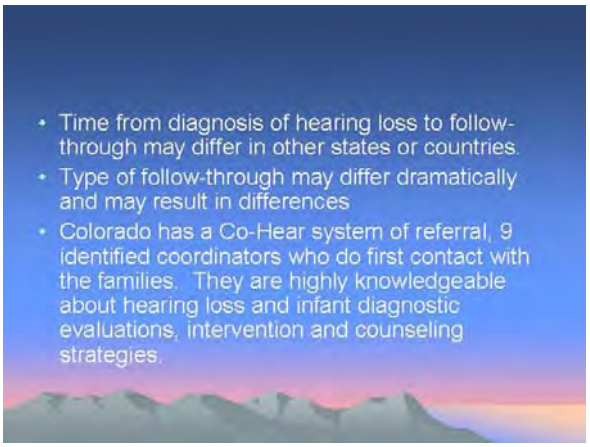
Parental Stress

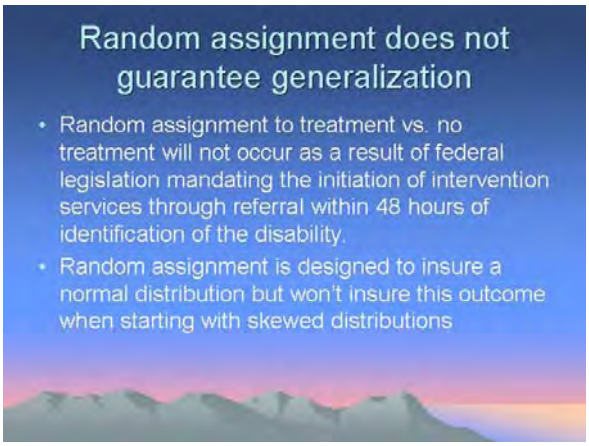
- Clemens, Davis & Bailey (Pediatrics-2000), Watkin, Baldwin, Dixon, Beckman (1998-British Journal of Audiology) Barringer & Mauk (1997-Audiology Today) indicate that 3-13% of false positive group have residual negative impact
- Approximately 10-13% of the population at large have clinical levels of stress

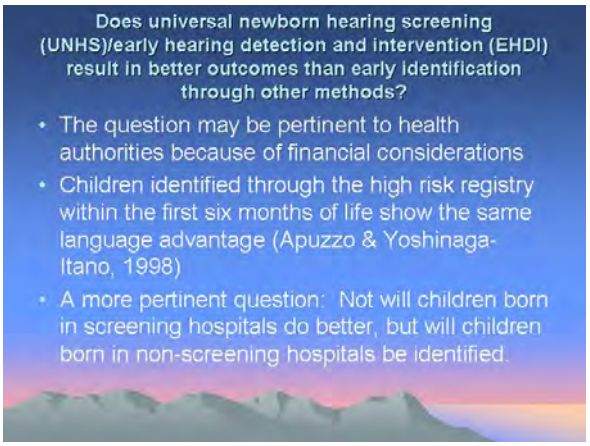

What is the role of the characteristics of intervention programs?
• There are three cohorts of children identified early (within the first six months) or with initiation of intervention within the first 11-12 months of life.
– Colorado (N= 72 to 150) dependent upon study
– Nebraska (N=25)
– Washington (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 differs for predictors of later-identified children.
• The proportion of early to later-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, 9 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

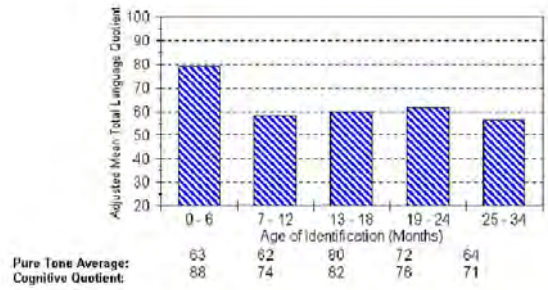

Does universal newborn hearing screening (UNHS)/early hearing detection and intervention (EHDI) result in better outcomes than early identification through other methods?
• The question may be pertinent to health authorities because of financial considerations
• Children identified through the high risk 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 & Mehl, 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 & Mehl, 1998 Pediatrics)



Adjusted mean total language quotients for groups based on age of identification of hearing loss.

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

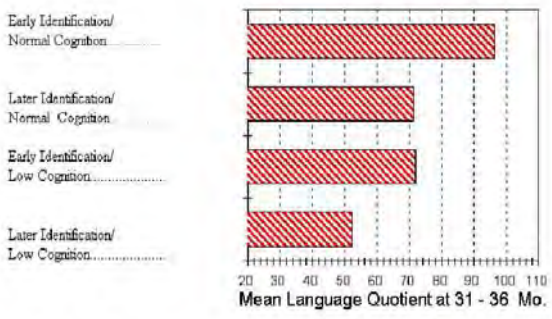
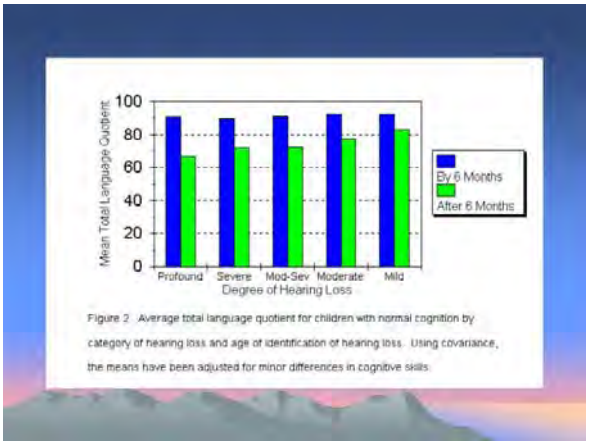


Figure 9. Mean total language quotient scores at 31 to 36 months by age of identification of hearing loss and cognition.

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.

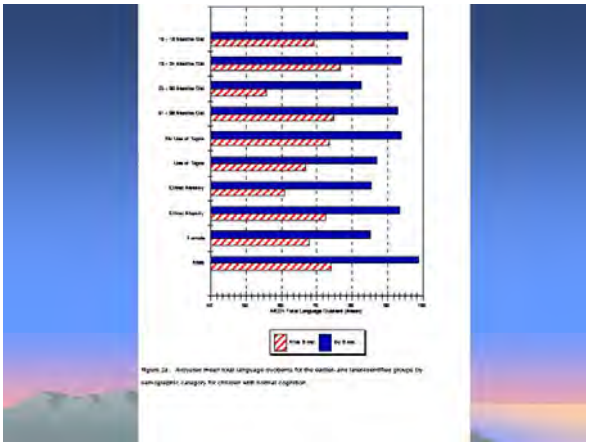


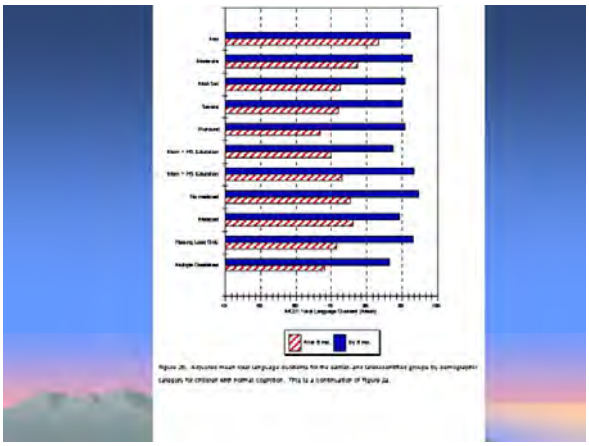
The EID/Early RX effect was present

- 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

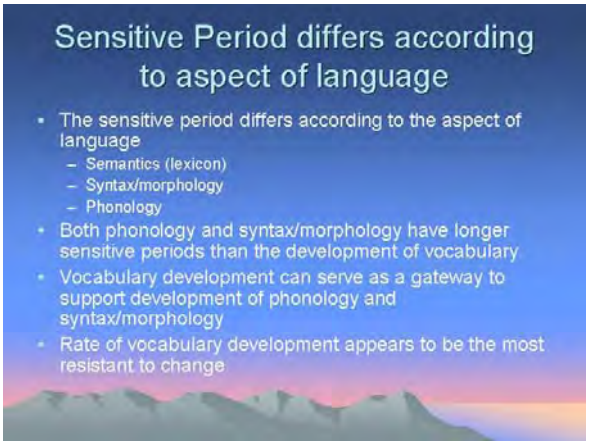
- Children from ethnic minority families and from ethnic majority families
- Children who used sign language
- Children who used speech
- Children with hearing loss only
- Children with multiple disabilities
- Children at all age levels (birth – 3 yrs)

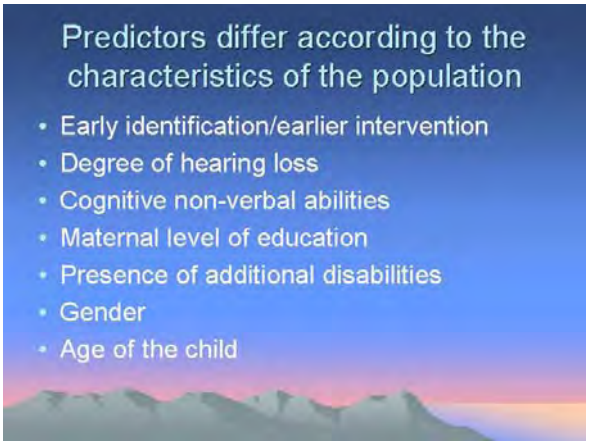


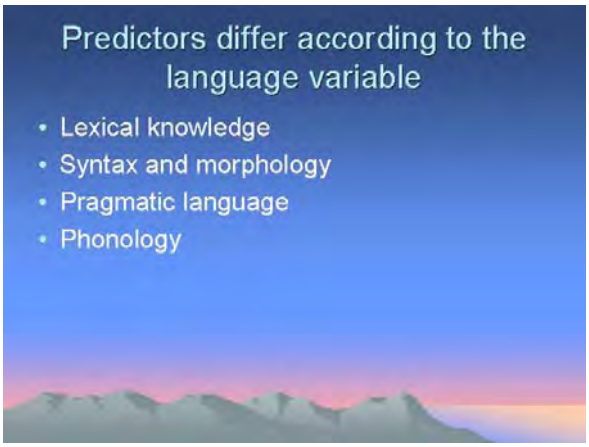


Sensitive Period: Speech Development

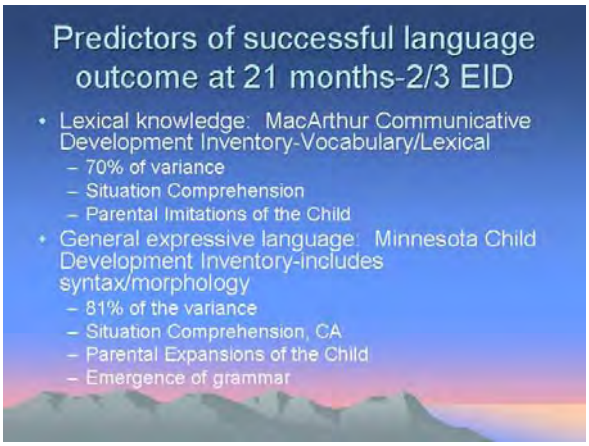
- The sensitive period for the development of speech is longer than for the development of language.
- Children with minimal or no speech development in the first three years of life could develop intelligible speech.


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
Gestures produced: 73% predicted by NV cognitive status
MCDI Comprehension/Conceptual: 68.7% predicted by NV cognitive
MCDI Expressive Language: 58.4% -NV cognitive,parent point/minute)
Mac CDI Words produced: 44.3%, maternal education, child gesture/minute
Diff Words Produced in language sample: 55.4% - maternal education, child gesture/minute, parent point/minute, parent symbolic gesture


Predictors of successful language outcome at 21 months-2/3 EID
Lexical knowledge: MacArthur Communicative Development Inventory-Vocabulary/Lexical
70% of variance
Situation Comprehension
Parental Imitations of the Child
General expressive language: Minnesota Child Development Inventory-includes syntax/morphology
81% of the variance
Situation Comprehension, CA
Parental Expansions of the Child
Emergence of grammar


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
69% of the 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 (3%)
- 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=59) 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

- Vocabulary development has the shortest sensitive period of development. Rate of vocabulary growth is also the most resistant to change.
- Vocabulary development predicts speech intelligibility
- Vocabulary is a gateway to the other aspects of speech development
- Vocabulary develops exponentially while phonology and syntax/morphology are a finite set of knowledge.

- **Unilateral Hearing Loss**
- **Earlier Cochlear Implantation**
- **Auditory Neuropathy**

Unilateral Hearing Loss

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

- **Sedey, A., Carpenter, K., Stredler-Brown, A.**

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**

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

	n	%
Mild	2	8
Moderate	6	24
Moderate-severe	6	24
Severe	4	16
Severe or profound	7	28

N = 25

Language Ability

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

Minnesota Inventory

- Participant Description:
  - 18 children
  - No additional disabilities
  - Selected oldest age available
  - Chronological age:
    - Range = 7 to 59 months
    - Mean = 25 months

Minnesota Inventory

	n	%
Expressive		
Borderline (70 – 79)	3	17
Average (80+)	15	83
Receptive		
Below average (< 70)	1	6
Borderline (70 – 79)	3	17
Average (80+)	14	78

MacArthur Inventory: Expressive

- Participant Description:
  - 12 children
  - No additional disabilities
  - Selected closest assessment to 21 months
  - Chronological age:
    - Range = 14 to 28 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

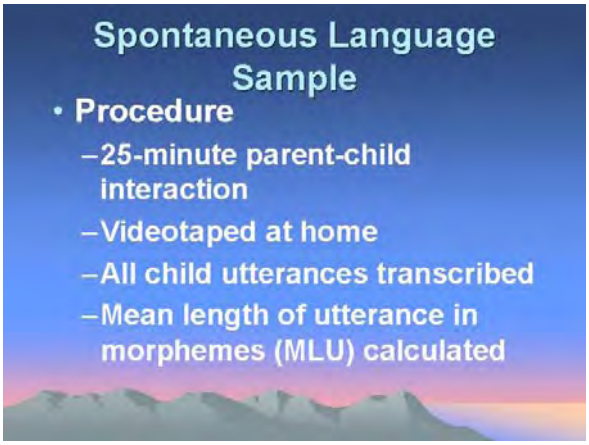
- Median Percentile
  - Expressive = 27th
  - Receptive = 15th

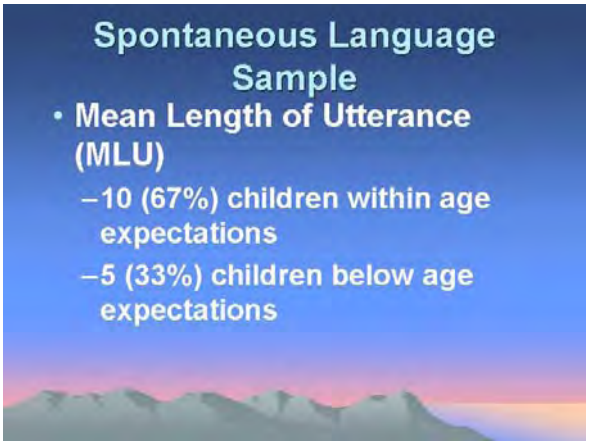
MacArthur Inventories

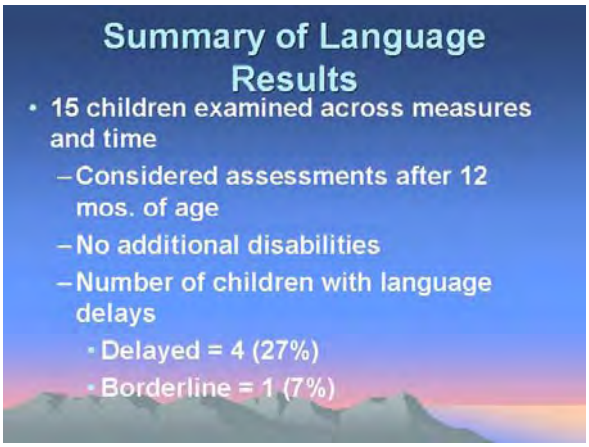
	n	%
Expressive		
< 10th percentile	2	17
> 10th percentile	10	83
Receptive		
< 10th percentile	2	18
> 10th percentile	9	82

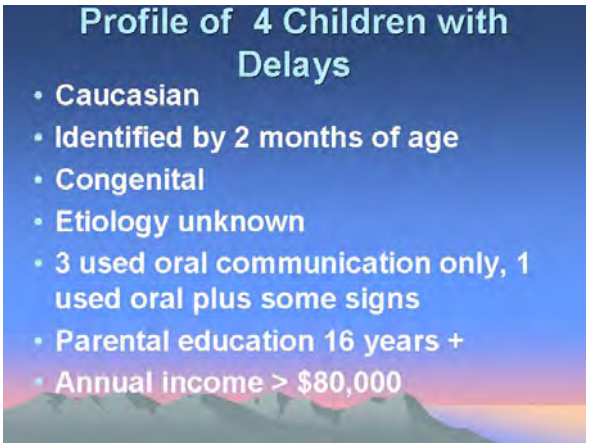
Spontaneous Language Sample

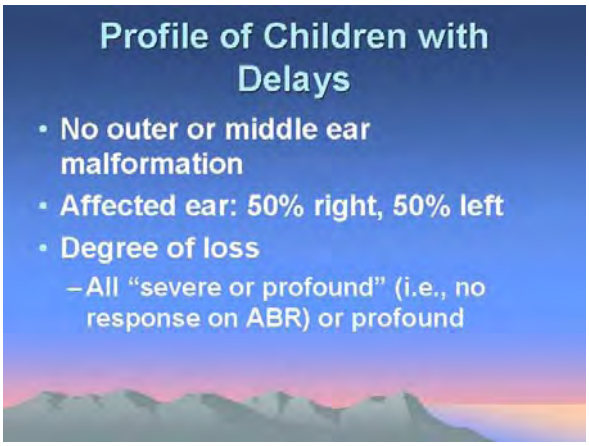
- Participant Description:
  - 15 children
  - No additional disabilities
  - Selected oldest age available for each child
  - Chronological age:
    - Range = 15 to 62 months
    - Mean = 29 months


Spontaneous Language Sample
• Procedure
– 25-minute parent-child interaction
– Videotaped at home
– All child utterances transcribed
– Mean length of utterance in morphemes (MLU) calculated


Spontaneous Language Sample
• 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

- Monitor language development on all unilaterals with severe-profound/profound losses-receptive/expressive vocabulary, general language and syntax delays-(27-34% - borderline or delayed)

UNHS and lower age of implantation

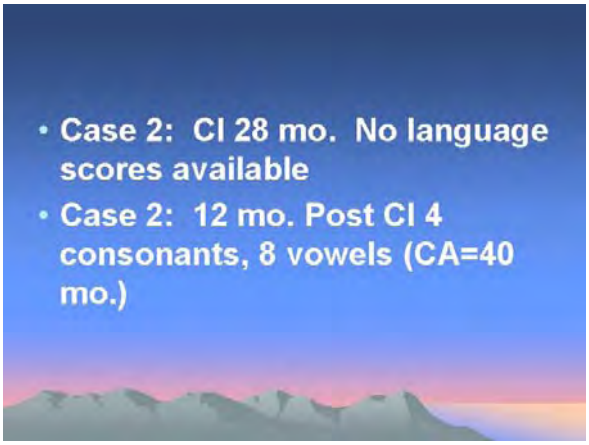
- Surgical considerations for early implantation – Sinninger, 2001
- Early implantation and developmental outcomes
- Relationship between speech and language outcomes

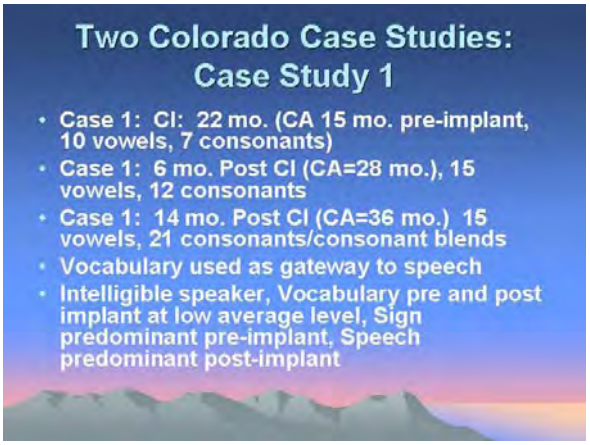
Children with cochlear implants

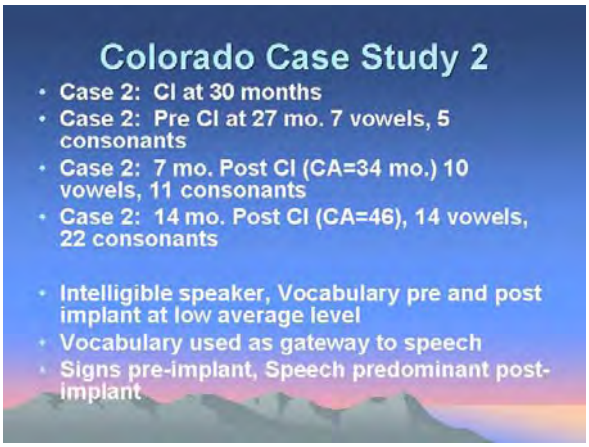
- Universal newborn hearing screening programs (UNHS) /early hearing detection and intervention programs (EHDI)
- At what age should children who are early-identified.
- Should children have cochlear implants at 6 months of age?

Relationship between speech and language

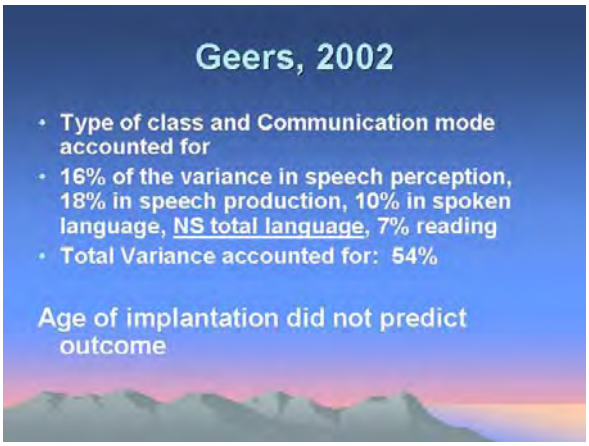
- Ertmer, Young, Grohne, Mellon, Johnson, Corbett & Saindon (2002) – followed two children every month for 12 months
- Case 1 -CI 10 mo. No language scores available
- Case 1: 12 mo. Post CI 2 consonants, 9 vowels (CA=22 mo.)


• Case 2: CI 28 mo. No language scores available
• Case 2: 12 mo. Post CI 4 consonants, 8 vowels (CA=40 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, Sign 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=46), 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 26% 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%
Age of implantation did not predict outcome


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

Connor, Hieber, Arts & Zwolan (2000)

- 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?

- Hammes, Novak, Rotz, Willis & Edmondson, 2002, Novak et al., 2000
- 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

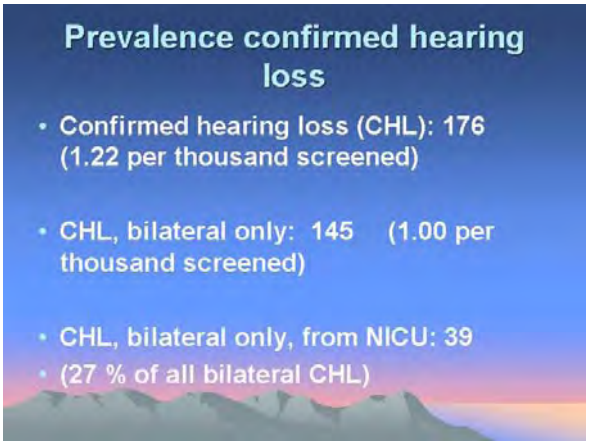
Auditory Neuropathy

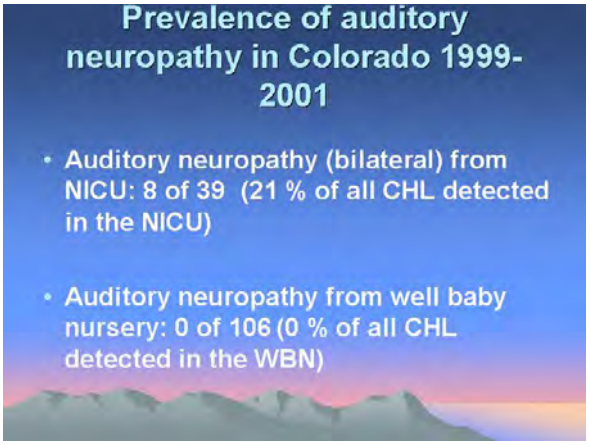
- Colorado statistics analyzed by Mehl, A. (2002)
- Children diagnosed with auditory neuropathy
- Well-baby vs. NICU

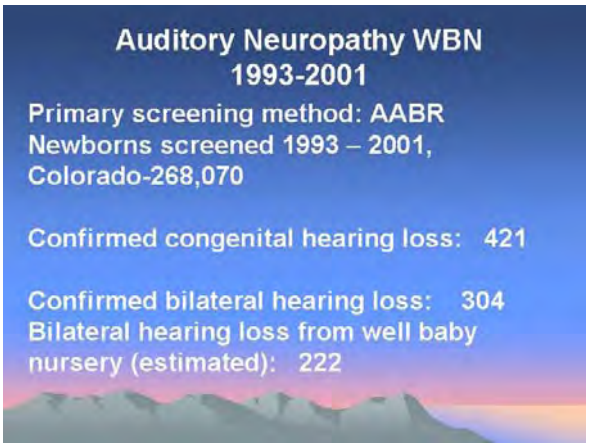
Newborns screened with AABR first (excluding OAE first) – 1999-2001 CO

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)

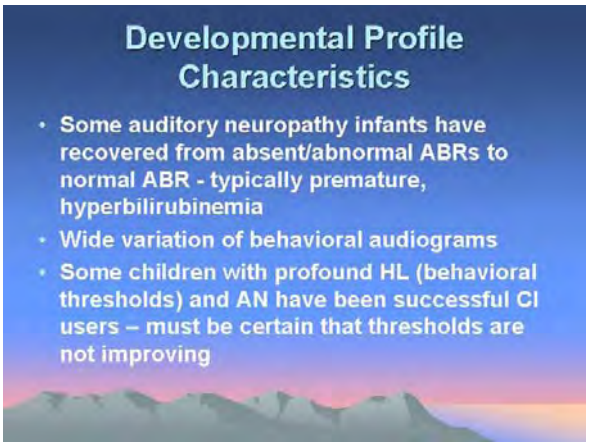

Prevalence confirmed hearing loss
• Confirmed hearing loss (CHL): 176 (1.22 per thousand screened)
• CHL, bilateral only: 145 (1.00 per thousand screened)
• CHL, bilateral only, from NICU: 39
• (27 % of all bilateral CHL)


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
Newborns screened 1993 – 2001, Colorado-268,070
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

Current data estimate 1 to 3 per thousand infants are born with significant hearing loss. The American Academy of Pediatrics Task Force on Newborn and Infant Hearing 1999 Position Statement and the JCIH 2000 Statement endorse universal newborn hearing screening.

How Many Infants Have Permanent HL at Birth?

Site	Rate
Well Baby Nursery	1 per 1000
NICU	10 per 1000
Total population	2-3 per 1000
# infants ident annually US	8,000-12,000
Average career pediatrician	12 patients

HL is the most common congenital abnormality

Current Screen Methods Available

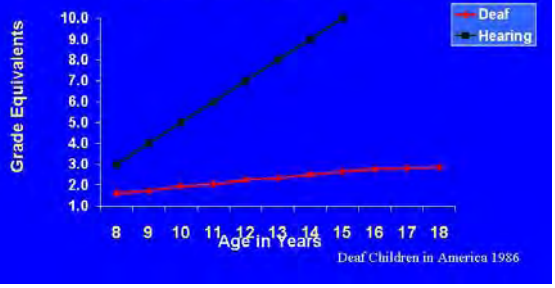
- Transient Evoked Otoacoustic Emissions — TEOAE
- Automated Auditory Brainstem Response — AABR
- Standard Auditory Brainstem Response — ABR
- Two Step — TEOAE + AABR

OAE

Impact of Hearing Loss

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

Reading Comprehension Scores of Hearing and Deaf Students



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 are, 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 $5000 to $25,000
- A one time expense
- Capital Equipment is Amoritized
  $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

Method	In Hospital	Post Discharge
TEOAE	± Rescreen	±Rescreen
AABR	± Rescreen	± Rescreen
Two Step	± Rescreen	± Rescreen
ABR	± Rescreen	± Rescreen

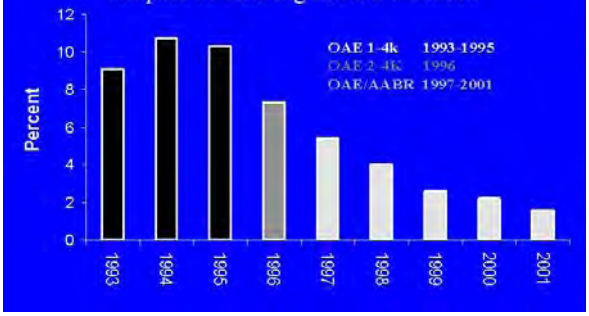
The Screen Population Affects Costs

- High Risk NICU infants vs Well babies
  - more difficult to test, older at time of testing
  - higher fail rate
- Small birthing hospital vs the high 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 rescreens 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 pretested at other sites.

Benchmarking a Hearing Screen Program

Quality indicators	>99% screened
1st stage (TEOAE/AABR) fail rate	≤4%
Rescreen return rate	>90%
Diagnostic procedures on refers	>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 + AABR	Women & Infants' Hospital University of New Mexico
One step TEOAE:	Memorial Hospital of RI
One step AABR:	St. Elizabeth's Hospital Boulder Children's Hospital

Site Characteristics

	A	B	C	D	E
Protocol	2 Step	2 Step	TEOAE	AABR	AABR
Stay	48	24	48	48	24
Screener	tech	student	tech	student	vol
Annual births	8,034	2,068	694	1,530	1,422

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
AABR
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.

Gabbard et al. Aud Today Dec. 1998

Study Sample Size

	A 2 Step	B 2 Step	C TEOAE	D AABR	E AABR
Retrospective	4,684	1,551	2,777	1,540	1,529
Prospective	346	300	100	150	160

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

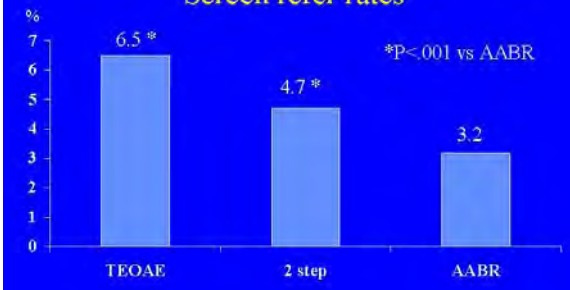
Audiologic Diagnostic Components

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

	A 2 Step	B 2 Step	C TEOAE	D AABR	E AABR
Stay (hr)	48	24	48	48	24
Pers	FTT	St	FTT	St	Vol
	3.7%	7.4%	6.5%	1.7%	3.9%

Screen refer rates



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

	2-Step	TEOAE	AABR
Variable Screening Costs	$13,343	$8,786	$21,517
Personnel	13,356	11,331	8,270
Equipment	8,456	4,506	8,081
Supplies	102	45	270
Total Overhead	21,914	15,882	16,620
Total Pre-discharge Costs	**35,247**	**24,668**	**38,137**
Total Costs per birth	**$23.50**	**$16.45**	**$25.42**
Refer Rate at Discharge	**4.7%**	**6.5%**	**3.2%**

Post-discharge Costs/ 1500 infants

	2-Step	TEOAE	AABR
Follow-up Screen Costs	**$10,388**	**$14,436**	**$7,140**
Diagnostic Evaluation Costs	3,937	3,937	3,937
Total Post-discharge Costs (F/U and Dx)	14,925	18,373	11,077
Total Pre- and Post-discharge Costs	**$49,582**	**$43,041**	**$47,215**
Total cost per birth	$33.05	$28.69	$32.81
Cost per identified child	**$16,527**	**$14,347**	**$16,405**

Factors Affecting Refer Rates and Costs

	2 Step	TEOAE	AABR
PT vs FT personnel	++	+++	+
48 hrs vs 24 hr discharge	+++	++	+

Number of Times an Infant is Screened

2-Step	TEOAE	AABR
1.37	1.23	1.15

- Discharge time affects the time available to obtain a definitive result (pass/refer).
- Therefore, 24 h discharge has ↑ negative impact on pass rates for protocols with ↑ # 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 3.2% 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

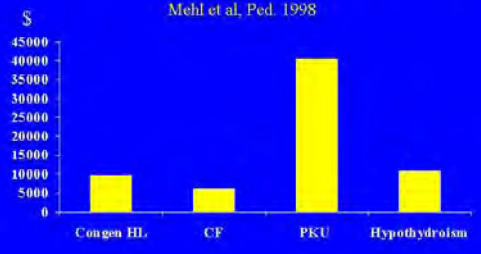
Sample	Time	Method	Screen Cost/infant
4,253	1993	2 Step screen	$26.05 Mason et al 95
10,372	92-97	AABR	$17.00 Mason et al 98
41,796	92-96	OAE, AABR or ABR	$25.00 Mehl et al 98

Cost of Identification

Screen Method	Cost to Identify Infant
2 Step TEOAE	$13,032 Mason et al 95
AABR	$17,705 Mason et al 98
OAE, AABR or ABR	$ 9,600 Mehl et al 98

Screening Costs per Confirmed Diagnosis

Mehl et al, Ped. 1998



Who pays for amplification and medical evaluation costs ?

- Private or Public Insurance
- Early Intervention
- The family
- The Department of Education
- Private Foundations
- ? Partial reimbursements

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

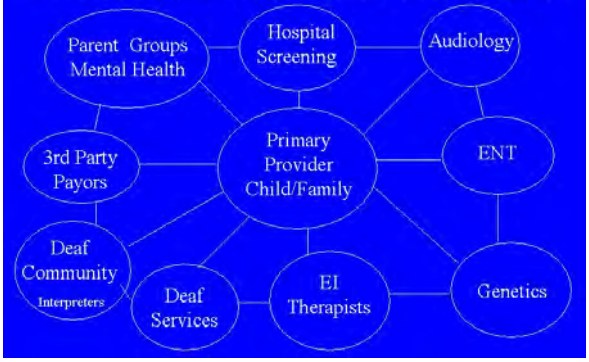
Garber S. Arch Otolaryn Head Neck Surg, 2002

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 onset
- 4.6 billion- lifetime costs for all persons identified in a single year 1998

Int J Technol Asses Health Care, 2000

EHDI, 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

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 AV 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 >5 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 EHDI 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	↑ costs amplification, habilitation birth to 3 years
	↑ communication skills, mainstreaming
	↑ success in work, private insurance
Late	↓ costs birth to 3 years
	↓ success school,language, work force
	↑ 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 »

Gaspard Montandon, Université Victor Segalen Bordeaux 2, France

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Introduction

- A. Déficits comportementaux liés à une privation auditive
  Behavioral deficits following auditory deprivation
- B. Périodes critiques du développement auditif
  Critical periods of auditory development
- C. Neurophysiologie du développement du cortex auditif
  Neurophysiology of sensitive periods in the auditory cortex

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Questions importantes…

- Quels sont les étapes critiques du développement auditif ?
  Which are the critical periods of the auditory development ?
- Quels sont les mécanismes neurobiologiques responsable de ces étapes?
  Which are the neurobiological mecanisms underlying these steps ?
- A partir de quel âge devient-il difficile de réhabiliter un enfant malentendant ?
  From when is it difficult to rehabilit a deaf child ?
- Combien de temps est-il nécessaire à un enfant pour réapprendre un langage ?
  How much time is necessary for a deaf child to re-learn a language ?

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A. Déficits comportementaux liés à une privation auditive
Behavioral deficits following auditory deprivation

- Périodes clés de l'acquisition du langage chez les enfants normo-entendants (Werker et coll., 1981)
  Key periods for language acquisition in normal child (Werker and al., 1981)
  - Perception phonologique 8-10 mois
    Phonologic perception 8-10 months
  - Capacités sémantiques 2-4 ans
    Semantic capacities 2-4 years
  - Capacités syntaxiques → 15 ans
    Syntactic capacities →15 years

⇨Plusieurs aspects du langage sont affectés irréversiblement s'il y a privation auditive durant ces périodes (Ruben et coll., 1997)
Several aspects of language are irreversibly affected if hearing is impaired during these periods (Ruben and al., 1997)

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---

A. Déficits comportementaux liés à une privation auditive
Behavioral deficits following auditory deprivation

- Périodes critiques pour l'acquisition d'un deuxième langage (Johnson et Newport, 1989)
  Critical periods for acquisition of a second langage in normal child (Johnson and Newport, 1989)

Comparaison entre des immigrants chinois arrivant à des âges différents
Comparison between Chineses immigrants at different ages

- Les immigrants ayant appris l'anglais entre 3 et 7 ans ont d'aussi bonnes performances que les anglophones de langue maternelle
  Immigrants who had learned English between 3 and 7 years old performed as well as native speakers
- Mais les immigrants étant arrivés à des âges plus avancés ont des performances diminuant progressivement avec l'âge
  But those who arrived at later ages performed progressively worse with increasing age at arrival

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A. Déficits comportementaux liés à une privation auditive
Behavioral deficits following auditory deprivation

- Périodes critiques lors de l'acquisition d'un langage
  Critical periods for language acquisition

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A. Déficits comportementaux liés à une privation auditive
Behavioral deficits following auditory deprivation

- Le cas des enfants sauvages *The case of the wild childs*
  - Un enfant a été enfermé dans une chambre sombre pendant 6 ans avec sa mère sourde (Mason et coll., 1942)
    A child isolated in a dark room for 6 years with her deaf mother (Mason and al., 1942)
  - Après 18 mois d'apprentissage, l'enfant savait lire, écrire et composer des histoires
    Within 18 months of discovery, the child could read, write and compose stories
  - Genie a été découverte à 13 ans, elle a été isolée depuis l'âge de 20 mois (Curtiss, 1977)
    Genie was discovered at 13 years of age, she had been isolated since the age of 20 months (Curtiss, 1977)
  - Après 7 ans de réhabilitation, Genie n'a jamais achevé l'acquisition d'un langage
    Even after 7 years of rehabilitation, Genie never achieved good language acquisition

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A. Déficits comportementaux liés à une privation auditive
Behavioral deficits following auditory deprivation

- Apprentissage du langage des signes (ASL) par des sourds congénitaux à des âges différents (Newport, 1990)
  American Sign Language acquisition shown by congenitally deafened adults (Newport, 1990)

Comparaison de trois groupes de sourds ayant appris le langage des signes à des âges différents
Comparison between three groups of adults

Depuis la naissance	*From birth*
Entre 4 et 6 ans	*Between 4 and 6 years*
Après l'adolescence	*Adolescent learners*

Les groupes les plus précoces ont de meilleures performances que les autres
The earlier the learning, the better the performances

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A. Déficits comportementaux liés à une privation auditive
Behavioral deficits following auditory deprivation

- Périodes critiques lors de l'acquisition d'un langage chez un malentendant
  Critical periods for language acquisition for deaf people

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A. Déficits comportementaux liés à une privation auditive
Behavioral deficits following auditory deprivation

- Récupération du langage grâce aux implants cochléaires
  Reversing auditory language deprivation through cochlear implants
  - Récupération d'une partie des informations cochléotopiques (Hartmann et coll., 1984)
    Recovering cochleotopic informations (Hartmann et al., 1984)
  - Récupération de la compréhension de la parole
    Recovering speech perception
  - Récupération de la production de la parole
    Recovering speech production

⇨ Selon certains facteurs ...
Some factors are required...

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A. Déficits comportementaux liés à une privation auditive
Behavioral deficits following auditory deprivation

- 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 implantations
  - 2. Ages du début de la surdité
    Ages of the beginning of auditory deprivation
  - 3. Durée de la privation auditive
    Duration of auditory deprivation

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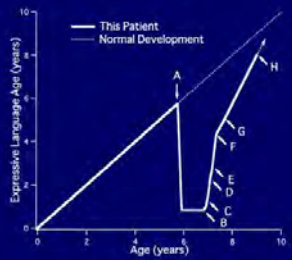
A. Déficits comportementaux liés à une privation auditive
Behavioral deficits following auditory deprivation

- Perte et récupération du langage après implantation cochléaire : étude de cas (Ito et coll., 2002)
  Postlingual collapse of language and its recovery after cochlear implantation : Case report (Ito and al., 1990)



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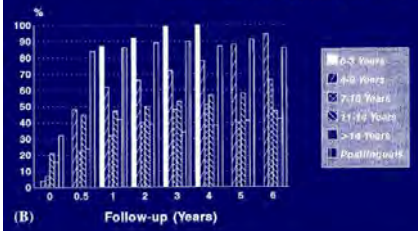
A. Déficits comportementaux liés à une privation auditive
Behavioral deficits following auditory deprivation

- Récupération du langage si implantation à bas âge (Manrique et coll., 1999)

Recovering language if early implantation (Manrique et al., 1999)



Test de perception de la parole : Mots quotidiens dans un contexte fermé

Speech test perception : Daily words in a Closed-set context

98 enfants sourds pré-linguaux

98 prelingually deaf children

Implants: Nucleus 22

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A. Déficits comportementaux liés à une privation auditive
Behavioral deficits following auditory deprivation

- Implantations cochléaires chez des enfants sourds pré-linguaux (Fryauf-Bertschy et coll., 1997)

Cochlear implant use by prelingually deafened children (Fryauf-Bertschy and al., 1997)

Comparaison entre 34 enfants implantés avant et après 5 ans

Comparison between 34 children implanted before and after 5 years of age

- Pas de différences significatives entre les performances à des tests fermés de perception de la parole

No significant differences in performance on closed-set tests of speech perception ability.

- Meilleures performances à des tests ouverts de reconnaissance de mots pour les implantés avant 5 ans

Open-set word recognition performance is significantly better for children implanted before 5 years of age

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A. Déficits comportementaux liés à une privation auditive
Behavioral deficits following auditory deprivation

- Effet de la durée de la privation auditive (Staller et coll., 1991)

Effect of duration of deafness on speech perception (Staller and al., 1991)

- Les enfants ayant eu une longue privation montrent de moins bonnes performances après 12 mois d'implantation

Children who had longer duration of deafness tended to demonstrate poorer performances on the more difficult perceptual tasks after 12 months of implant experience.

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A. Déficits comportementaux liés à une privation auditive
Behavioral deficits following auditory deprivation

- Facteurs potentiels contribuant à de pauvres performances dans le langage (Gordon et coll., 2000 ; O'Donoghue et coll., 2000)

Potentials factors contributing to poor language performances (Gordon and al., 2000 ; O'Donoghue and al., 2000)

- Age lors de l'implantation

Age at implantation

- Mode de communication pré-implantation

Pre-implant language deafness duration

- Suivi post-implantation

Post-implant follow-up

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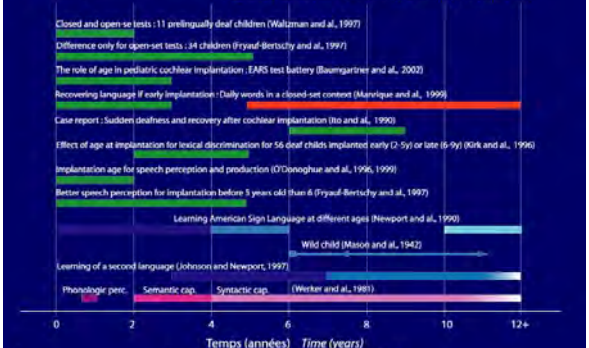
A. Déficits comportementaux liés à une privation auditive
Behavioral deficits following auditory deprivation



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A. Déficits comportementaux liés à une privation auditive
Behavioral deficits following auditory deprivation

- Conclusion

Conclusion

- Périodes critiques du développement du langage et de l'audition

Critical periods in development of language and audition

- Peu d'effet de la durée de privation sur l'acquisition du langage

No important effect of deprivation duration on language acquisition

- Développement du cortex auditif versus aires du langage

Development of auditory cortex versus language areas

- Acquisition du langage possible chez l'adulte (Staller ou Dawson)

Possible language acquisition by adults

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B. Périodes critiques du développement auditif
Critical periods for auditory development

- Introduction *Introduction*
  - L'étude neurobiologique est nécessaire à la compréhension du comportement (Lenneberg, 1967)
    Biological considerations were necessary for the understanding of behavior (Lennerberg, 1967)
  - Le langage est composé de plusieurs sous-systèmes avec des périodes de développement différentes (Neville et coll., 1992)
    Language consists of several subsystems with different developmental periods (Neville and al., 1992)
    - 1. Maturation des aires du langage
      Language areas maturation
    - 2. Maturation des aires auditives (cortex auditif primaire et cortex auditif associatif
      Auditory areas maturation (primary auditory cortex and associative auditory cortex)

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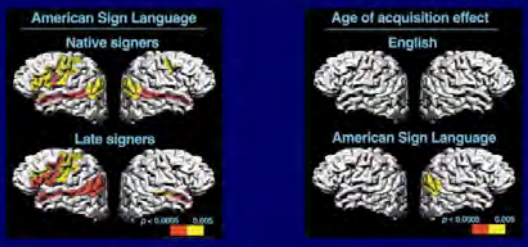
B. Périodes critiques du développement auditif
Critical periods for auditory development

- Période critique dans l'utilisation de l'hémisphère droit lors de production de signes ASL (Newman et coll., 2002)
  Critical period for right hemisphere recruitment in American Sign Language processing (Newman and al., 2002)



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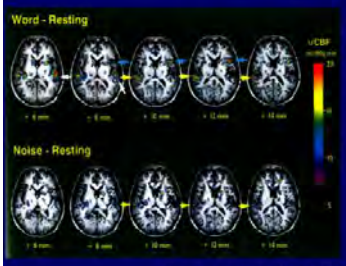
B. Périodes critiques du développement auditif
Critical periods for auditory development

- Efficacité des implants cochléaires chez des sourds pré- et post-linguaux (Okazawa et coll., 1996)
  Cochlear implant efficiency in pre- and postlingually deaf subjects (Okazawa and al., 1996)



5 sourds pré-linguaux
versus
5 sourds post-linguaux
5 prelingually deafs versus 5 postlingually

Stimuli verbaux vs bruit blanc
Verbal stimuli versus white noise

Mesure de débit sanguin cérébral (rCBF) par H_2O radiomarqué
Regional cerebral blood flow (rCBF) radiolabelled

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B. Périodes critiques du développement auditif
Critical periods for auditory development

- Activations chez des sourds pré- et post-linguaux (Okazawa et coll., 1996)
  Activations of prelingually and postlingually deaf (Okazawa and al., 1996)

Region of insterest	Prelingually implanted deafs		Postlingually implanted deafs	
	White noise	Words	White noise	Words
Primary auditory cortex	-	-	x	X
Wernicke's area	-	X	-	X
Broca's area	-	X	-	X

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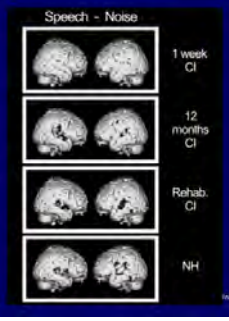
B. Périodes critiques du développement auditif
Critical periods for auditory development

- Plasticité subséquente à une implantation (Giraud et coll., 2001)
  Plasticity subsequent to implantation (Giraud and al., 2001)



Réorganisation du système de la perception de la parole chez des sourds post-linguaux
Reorganisation of the speech perception system in postlingual cochlear implanted patients

Activité cérébrale mesurée par PET
Cerebral activity with PET

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B. Périodes critiques du développement auditif
Critical periods for auditory development

- Plasticité associée à une surdité pré-linguale
  Plasticity associated with prelingual deafness
  - Amélioration des performances visuelles associée à une augmentation de l'activation des aires visuelles?
    Enhancement of visual performance associated with increased activation of areas related to vision
  - Utilisation des aires auditives et de celles du langage par d'autres modalités sensorielles
    Takeover of the regions normally dedicated to auditory and language processing by other sensory modalities
  - Désaccord entre les données fonctionnelles et la sensation reportée par les patients stimulés électriquement
    Discrepancy between functional neuro-anatomical data and the sensation reported by patients in response to electrical stimulation

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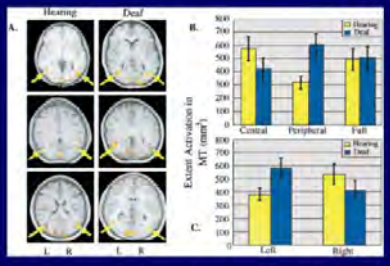
B. Périodes critiques du développement auditif
Critical periods for auditory development

- Une amélioration des performances visuelles associée à une augmentation de l'activation des aires visuelles (Bavelier et coll., 2000)
  An enhancement of visual performance associated with increase activation of areas related to vision (Bavelier and al., 2000)



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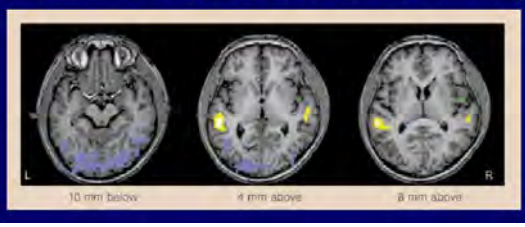
B. Périodes critiques du développement auditif
Critical periods for auditory development

- Le langage des signes utilisé par les sourds congénitaux n'active pas seulement les aires visuelles (Nishimura et coll., 1999)
  Sign language used by congenitally deaf adults activates not only visual areas (Nishimura and al., 1999)

Activités cérébrales mesurées par IRMf pour le langage des signes (jaune), l'audition (vert) et la vision (bleu)
Activation of areas of the brain by fMRI for sign language (yellow), audition (green) and vision (blue)



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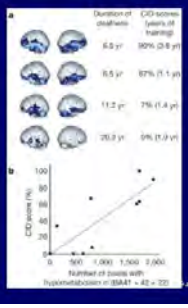
B. Périodes critiques du développement auditif
Critical periods for auditory development

- L'activité du cortex auditif associatif induite par un implant cochléaire décroît avec l'augmentation de l'âge d'implantation (Lee et coll., 2001)
  The amount of activation of the high-order auditory cortices by a cochlear implant decreases with increasing implant age (Lee and al., 2001)



Activité cérébrale de 15 patients pré-linguaux au niveau des aires auditives
Cerebral auditory activity of 15 prelingually patients
Performances à des tests de perception de la parole
Speech perception performances

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B. Périodes critiques du développement auditif
Critical periods for auditory development

- Une longue privation entraîne une baisse d'activité cérébrale
  Long duration of deafness lead to low auditory cortex activity
  - Plus longue est la période de surdité, plus basse sera l'activité cérébrale du cortex auditif mesurée par PET (Ito et coll., 1993)
    The longer the duration of deafness, the lower the brain activity in the auditory cortex measured by PET (Ito and al., 1993)
  - Potentiels évoqués auditifs chez les pré- et post-linguaux implantés (Jordan et coll., 1997)
    Auditory event-related potentials in post- and prelingually deaf cochlear implant recipients (Jordan and al., 1997)

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B. Périodes critiques du développement auditif
Critical periods for auditory development

- Périodes sensibles des aires auditives
  Critical periods for auditory cortices
  - Les sourds pré-linguaux montrent une réorganisation de leurs aires auditives.
    Prelingually deafs show a reorganisation of the auditory areas
  - Une longue privation auditive diminue l'activité des aires auditives
    A long auditory deprivation decrease auditory areas activity
  - L'absence de stimulations auditives durant les premières années de la vie d'un enfant malentendant est responsable du développement particulier du cortex auditif
    Auditory deprivation in the first years of the child life lead to special auditory cortices development
  - Les aires du langage semblent épargnées par une privation auditive
    Language areas are not disturbed by auditory deprivation, but stop their development

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

- Introduction *Introduction*
  - Comme les aires du langage, le cortex auditif primaire se développe durant la vie postnatal (Ponton et coll., 1999)
    As the language areas, the primary auditory cortex is evolving during postnatal life (Ponton and al., 1999)
  - Les paramètres fonctionnelles du cortex auditif primaire se développent durant les 12-15 premières années (Eggermont et coll., 1988)
    Primary auditory cortex developping during the 12th-15th first years of life (Eggermont and al., 1988)
  - Le cortex auditif primaire doit être stimulé pour se développer (Binns et coll., 1995)
    Primary auditory cortex must be stimulated to mature (Binns and al., 1995)

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

- Mécanismes neuronaux du développement du cortex auditif
  Neuronal development of auditory cortex
  - *1. Rapide synaptogenèse génétiquement déterminée*
    Fast synaptogenesis genetically determined
  - *2. Maturation synaptique lente et stabilisation ou élimination de 50% des synapses (« pruning »)*
    Slow synaptic maturation et stabilization or elimination of 50% of the synaps (« pruning »)

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

- Développement du cortex auditif chez l'animal sourd
  Development of auditory cortex in deaf animals
  - Activité synaptique décroissante au niveau du cortex auditif primaire des animaux sourds congénitaux
    Decrease of synaptic activity in deaf animals
  - Faible réorganisation cochléotopique
    Low cochleotopic 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 cortical areas are diminished

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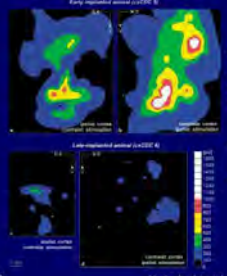
C. Neurophysiologie du développement du cortex auditif
Neurophysiology of sensitive periods in the Auditory Cortex

- Activation du cortex auditif de chats sourds par électrostimulation cochléaire (Klinke et coll., 1999)
  Recruitment of the auditory cortex in Congenitally deaf cats by long-term cochlear electrostimulation (Klinke and al., 1999)



Potentiels électriques mesurés au niveau du cortex auditif primaire de chats sourds lors d'une stimulation cochléaire de plusieurs mois
Electrical potentials recorded from the exposed primary auditory cortex of cats after several months of auditory experience

- Précoce: implantation à 3,5 mois et stimulation durant 5 mois
  Early : implantation at 3,5 months and stimulation during 5 months
- Tardif: implantation à 6 mois et stimulation durant 5 mois
  Late: implantation at 6 months and stimulation during 5 months

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Conclusion *Conclusion*

A. Déficits comportementaux : Période critique vers l'âge de 2-5 ans pour la perception de la parole chez les enfants sourds congénitaux
Critical period between 2 and 5 years of age for speech perception for deaf child

B. Plasticité du cortex auditif : *Auditory cortex plasticity*

- Utilisation des aires auditives par d'autres modalités sensorielles chez les sourds pré-linguaux
  Recruitment of auditory areas by other sensory modalities for prelingually deaf
- Décroissance de l'activité des aires auditives avec l'augmentation de la durée de la privation
  Decreasing of auditory areas activity with increasing of deprivation duration

C. Développement du cortex auditif *Auditory cortex development*

- Rapide synaptogenèse durant la première année de la vie, puis élimination des synapses
  Fast synaptogenesis during the first year of life, then synaps 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 dead

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Merci pour votre attention

Thank you for your attention

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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 preformed 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.

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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.

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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 Durieux-Smith and Sharon Bartholomew,
Co-chairs, CWGCH

CWGCH@hc-sc.gc.ca

Outline

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

Canadian Working Group on Childhood Hearing (CWGCH)

Introduction

- Established in September 2000 by the Health Surveillance and Epidemiology Division (HSED), Health Canada
- Formed in response to growing interest in the identification and management of newborn hearing loss
- Membership is multidisciplinary

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

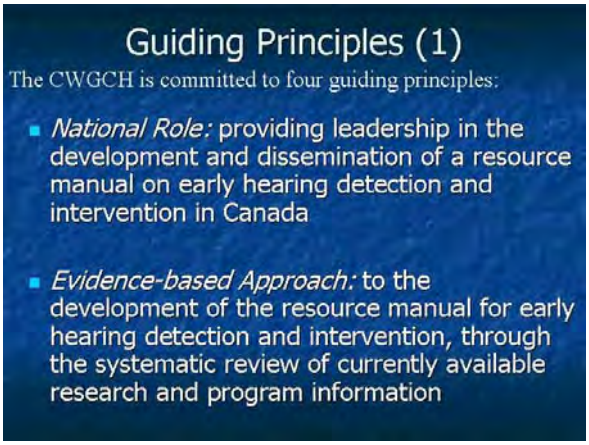
History of Working Group (2)

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

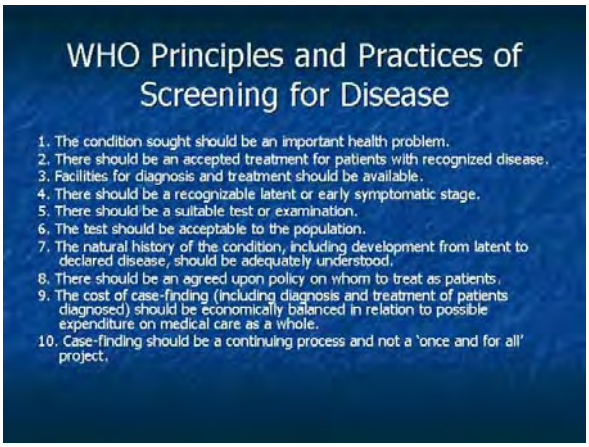
Canadian Working Group on Childhood Hearing established

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

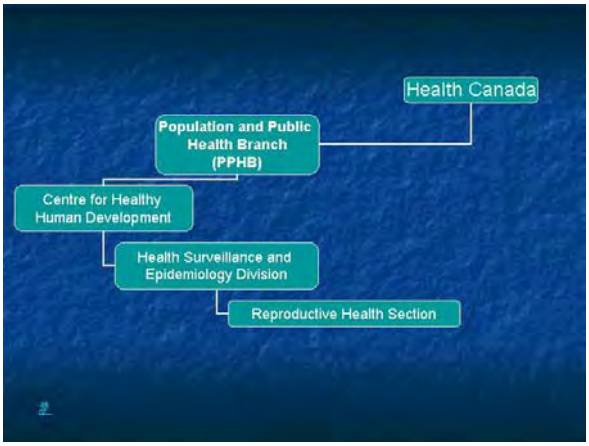

CWGCH representation (cont.)
Canadian Paediatric Society.
Canadian Society of Otolaryngology- Head and Neck Surgery.
College of Family Physicians
Canadian Association of Educators of the Deaf and Hard of Hearing.
Institut National de Sante publique du Quebec (INSPQ).
Consumer representative (parent)
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

- Thompson et al. (2001) Universal Hearing Screening : Summary of Evidence.
  - 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 evidence (cont.)

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
- Interventions
  - 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.)

- 1974 : Nova Scotia Conference on the Early Identification of Hearing loss
  - Focal point: approval of methods for screening newborns for hearing loss
  - Use of High Risk Register
  - Behavioural screening

BACKGROUND : Conferences (cont.)

- 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 : viable 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)
- 1981 : Task Force on Childhood Hearing Impairment (Department of Health and Welfare)

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.

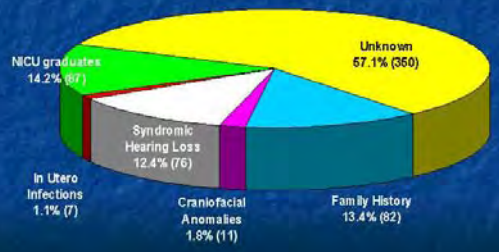
BACKGROUND (cont.)

- 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.

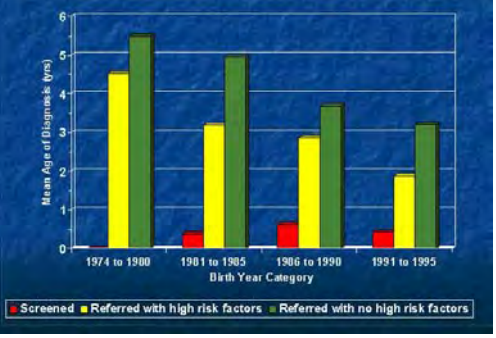
BACKGROUND (cont.)

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

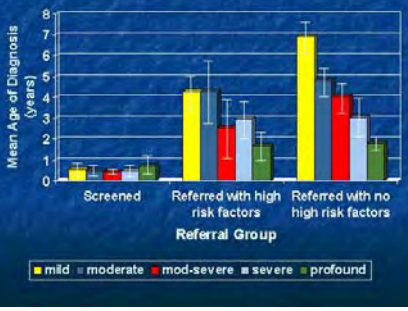
Risk Status for the 613 children, wearing hearing aids, born between 1974 and 1995



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 (1980-95): 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

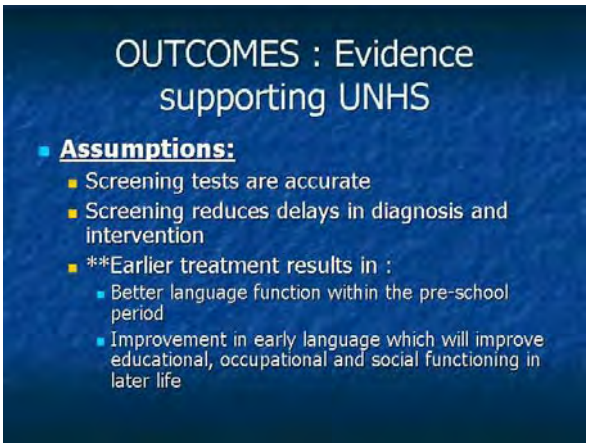
- **Temporal pattern of PCHI detection in the absence of UNHS**
  - Studies on the age of diagnosis in the absence of any screening activities,
  - Studies on the age of diagnosis in the presence of some screening activities.

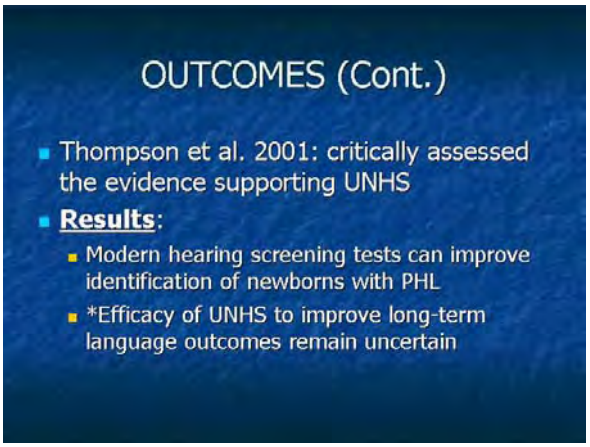
BURDEN OF THE TARGET DISORDER : Temporal pattern (cont.)

- 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.

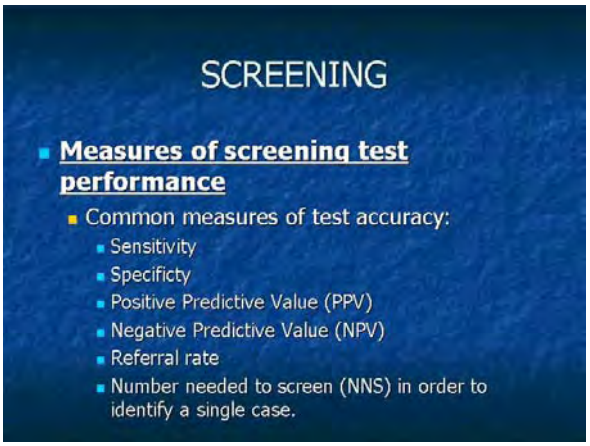
BURDEN OF THE TARGET DISORDER : Temporal pattern (cont.)

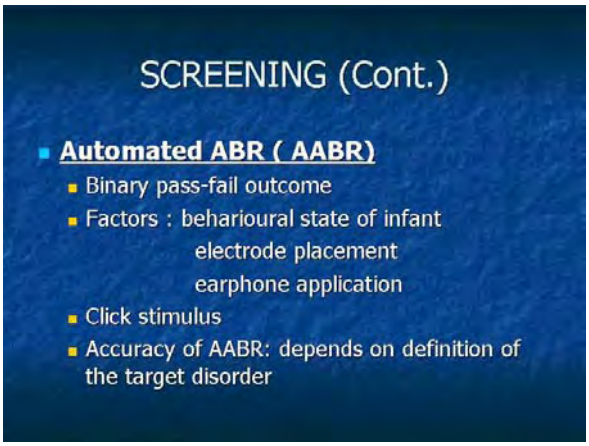
- Summary (cont.)
  - The ages of diagnosis of children born in the absence of any screening activities or in the presence of some screening activities are very similar
  - In studies which have investigated different birth cohorts, the ages of diagnosis have improved over time

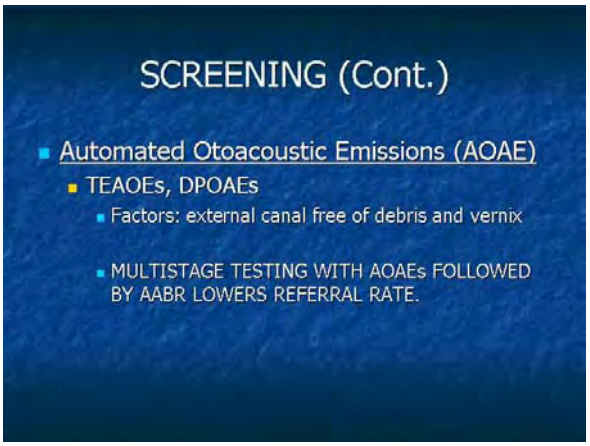

OUTCOMES : Evidence supporting UNHS
Assumptions:
Screening tests are accurate
Screening reduces delays in diagnosis and intervention
**Earlier treatment results in :
Better language function within the pre-school period
Improvement in early language which will improve educational, occupational and social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

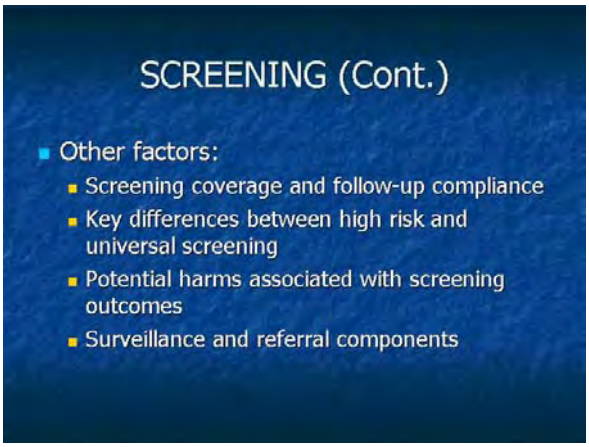

OUTCOMES (Cont.)
Thompson et al. 2001: critically assessed the evidence supporting UNHS
Results:
Modern hearing screening tests can improve identification of newborns with PHL
*Efficacy of UNHS to improve long-term language outcomes remain uncertain

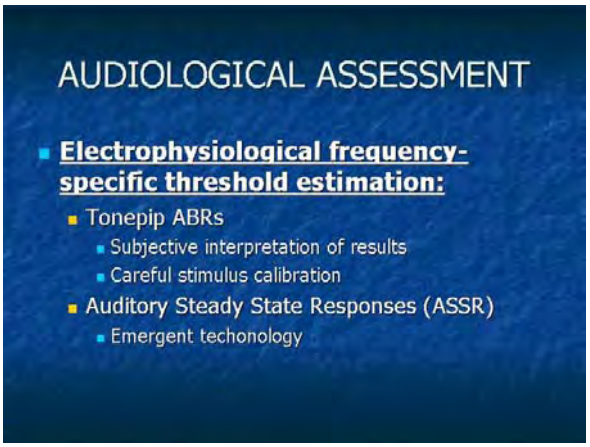

SCREENING
Importance of defining target disorder and target population
Target disorder : severity, frequency range, laterality, permanence of site of lesion, time of onset
Target population : at risk or all children

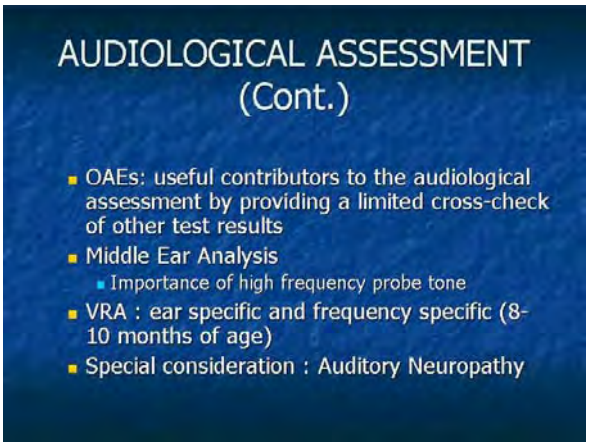

SCREENING
Measures of screening test performance
Common measures of test accuracy:
Sensitivity
Specificty
Positive Predictive Value (PPV)
Negative Predictive Value (NPV)
Referral rate
Number needed to screen (NNS) in order to identify a single case.

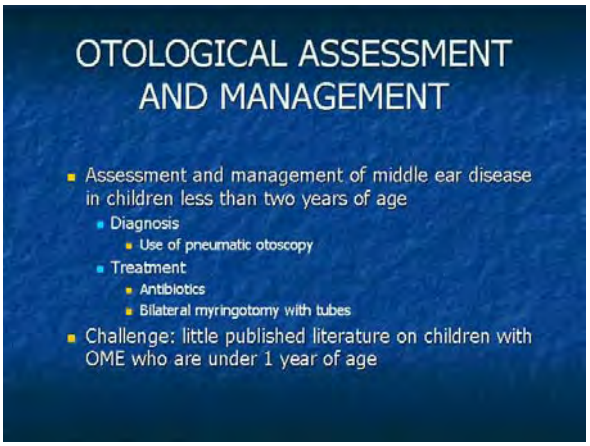

SCREENING (Cont.)
Automated ABR (AABR)
Binary pass-fail outcome
Factors : behavioural state of infant
electrode placement
earphone application
Click stimulus
Accuracy of AABR: depends on definition of the target disorder

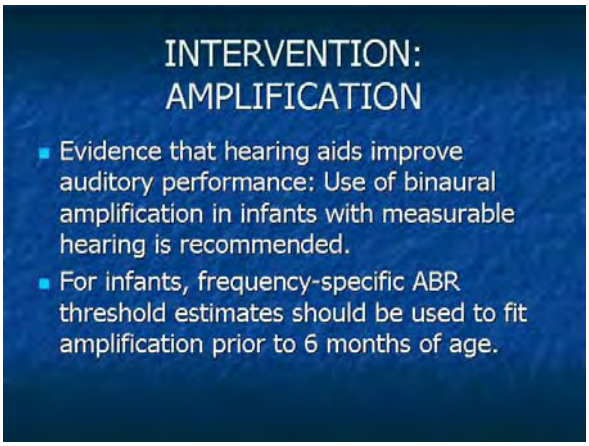

SCREENING (Cont.)
Automated Otoacoustic Emissions (AOAE)
TEAOEs, DPOAEs
Factors: external canal free of debris and vernix
MULTISTAGE TESTING WITH AOAEs FOLLOWED BY AABR LOWERS REFERRAL RATE.


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

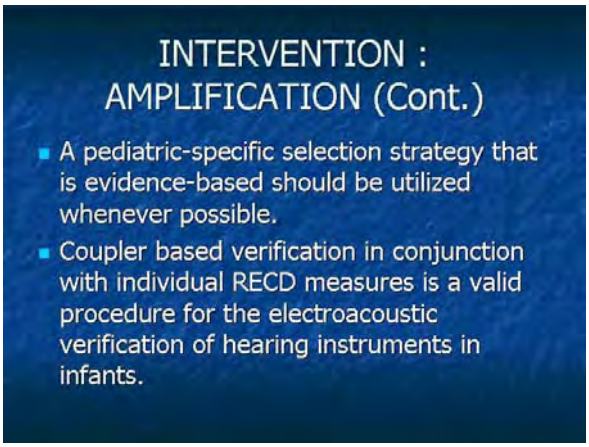

AUDIOLOGICAL ASSESSMENT
Electrophysiological frequency-specific threshold estimation:
Tonepip ABRs
Subjective interpretation of results
Careful stimulus calibration
Auditory Steady State Responses (ASSR)
Emergent techonology

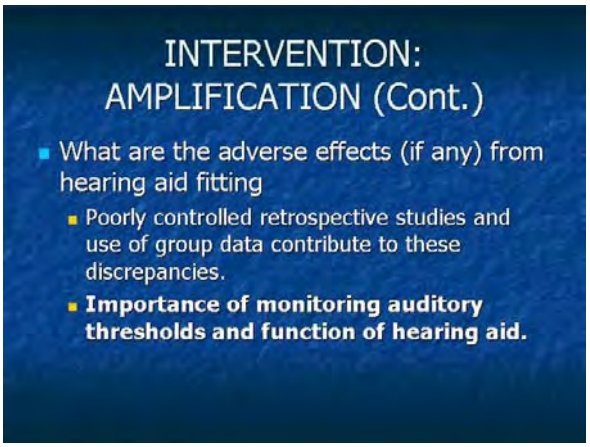

AUDIOLOGICAL ASSESSMENT (Cont.)
OAEs: useful contributors to the audiological assessment by providing a limited cross-check of other test results
Middle Ear Analysis
Importance of high frequency probe tone
VRA : ear specific and frequency specific (8-10 months of age)
Special consideration : Auditory Neuropathy

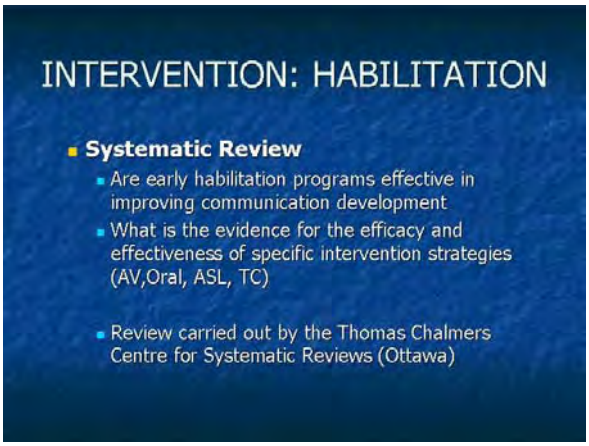

OTOLOGICAL ASSESSMENT AND MANAGEMENT
Assessment and management of middle ear disease in children less than two years of age
Diagnosis
Use of pneumatic otoscopy
Treatment
Antibiotics
Bilateral myringotomy with tubes
Challenge: little published literature on children with OME who are under 1 year of age

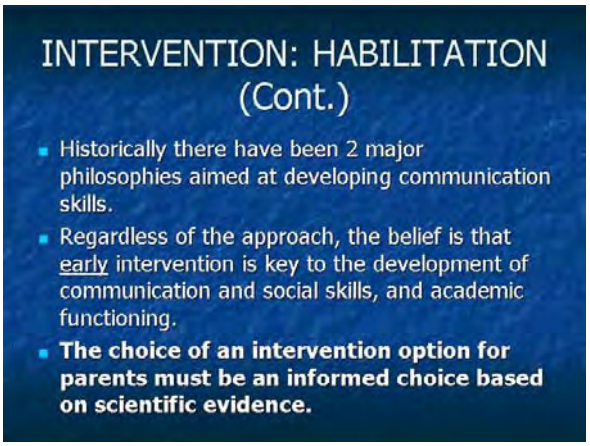

INTERVENTION: AMPLIFICATION
Evidence that hearing aids improve auditory performance: Use of binaural amplification in infants with measurable hearing is recommended.
For infants, frequency-specific ABR threshold estimates should be used to fit amplification prior to 6 months of age.

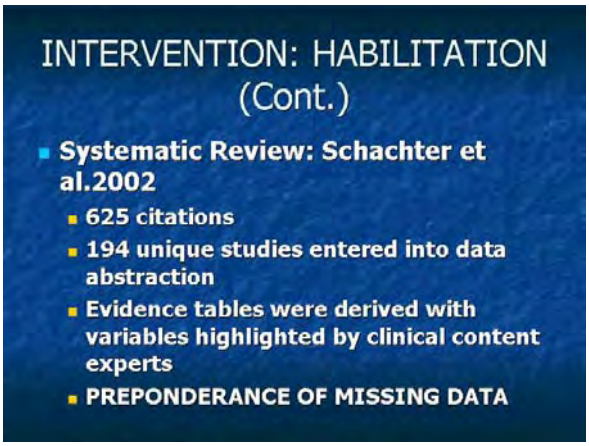

INTERVENTION : AMPLIFICATION (Cont.)
Acoustic properties of ear canals of infants and young children show high between subject variability. These differences need to be applied to the amplification process.
*Rear Ear Coupler Difference (RECD): reliable and valid measure for the purpose of fitting amplification in infants.

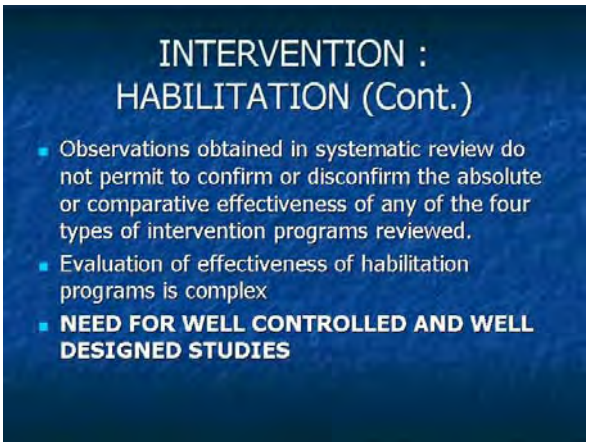

INTERVENTION : AMPLIFICATION (Cont.)
A pediatric-specific selection strategy that is evidence-based should be utilized whenever possible.
Coupler based verification in conjunction with individual RECD measures is a valid procedure for the electroacoustic verification of hearing instruments in infants.


INTERVENTION: 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 aid.


INTERVENTION: 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)


INTERVENTION: 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.


INTERVENTION: 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


INTERVENTION : HABILITATION (Cont.)
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

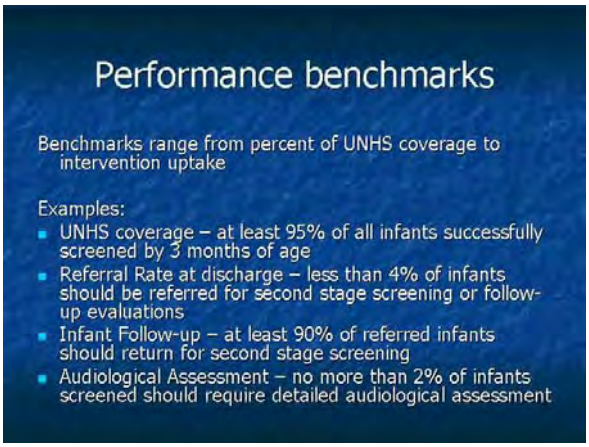
- Realistic indicators and benchmarks must be defined
  - These are used to determine if the program is meeting its performance targets
- Benchmarks are levels to which the program aspires
  - Important for proving the program is beneficial and cost effective

Outcome Measures

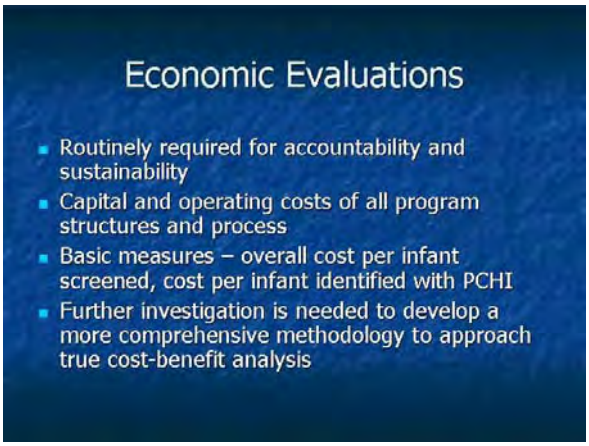
Measures range from screening proportions to communication development outcomes

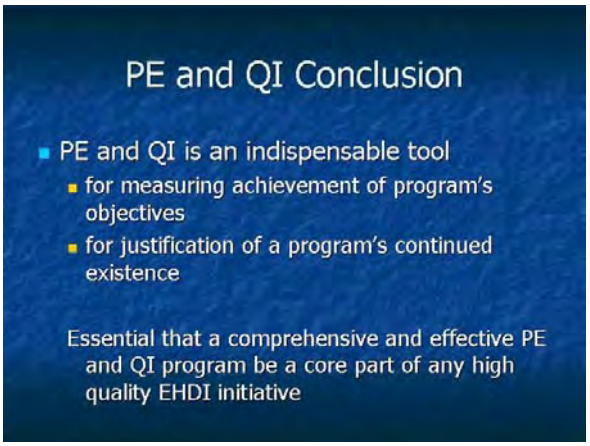
Examples:

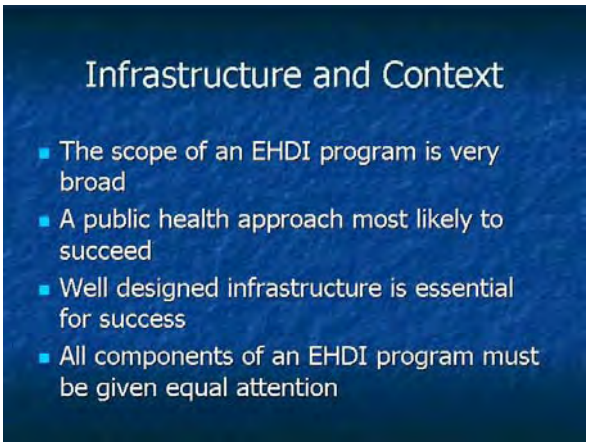
- Number (N) and proportion (P) of overall target population screened by one month of age and one month post-discharge
- N and P infants with refer (by site, screening personnel and risk status)
- N and P of referrals for whom audiological assessment is initiated by three months of age and within two 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

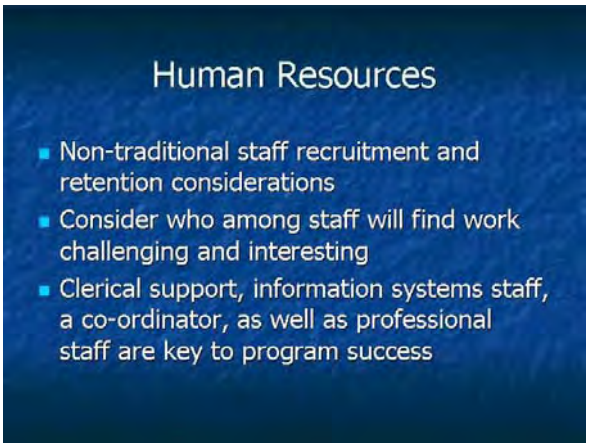

Long term Outcomes
Sub-program outcomes vs ultimate measure of success
Ultimate measures of success include:
family satisfaction with services
Improved hearing ability
Many factors impact on outcome

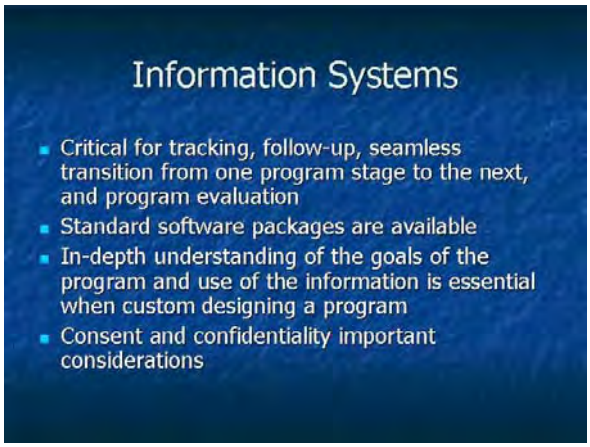

Economic Evaluations
Routinely required for accountability and sustainability
Capital and operating costs of all program structures and process
Basic measures – overall cost per infant screened, cost per infant identified with PCHI
Further investigation is needed to develop a more comprehensive methodology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

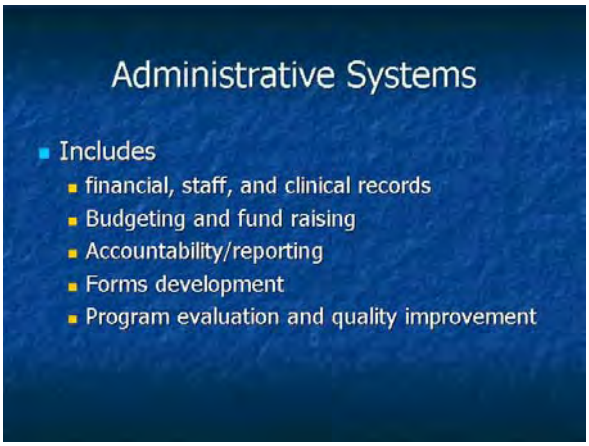

PE and QI Conclusion
PE and QI is an indispensable tool
for measuring achievement of program's objectives
for justification of a program's continued existence
Essential that a comprehensive and effective PE and QI program be a core part of any high quality EHDI initiative


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

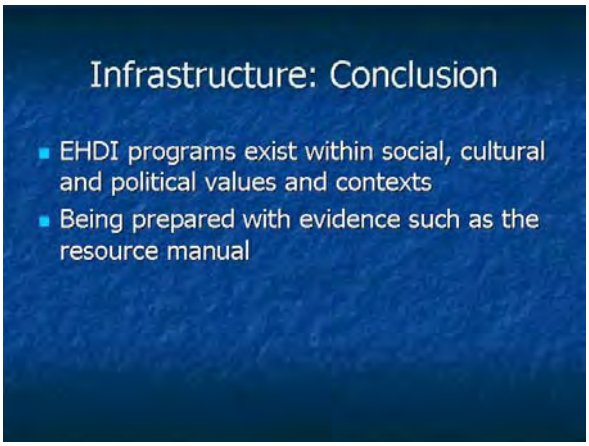

Infrastructure
Those elements that support, sustain and link all program components to achieve the program goal
Main components
Human Resources
Information Services
Administrative Systems
Communication Systems

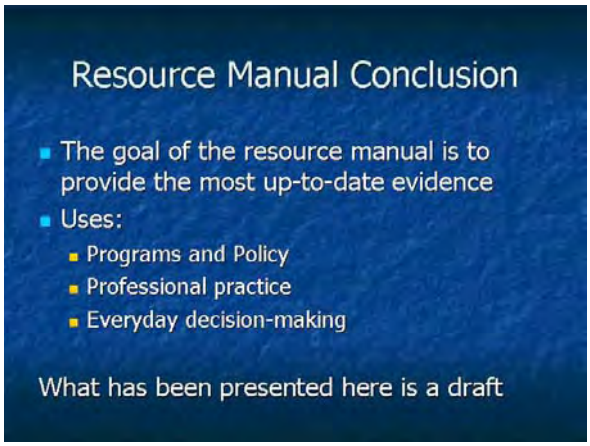

Human Resources
Non-traditional staff recruitment and retention considerations
Consider who among staff will find work challenging and interesting
Clerical support, information systems staff, a co-ordinator, as well as professional staff are key to program success

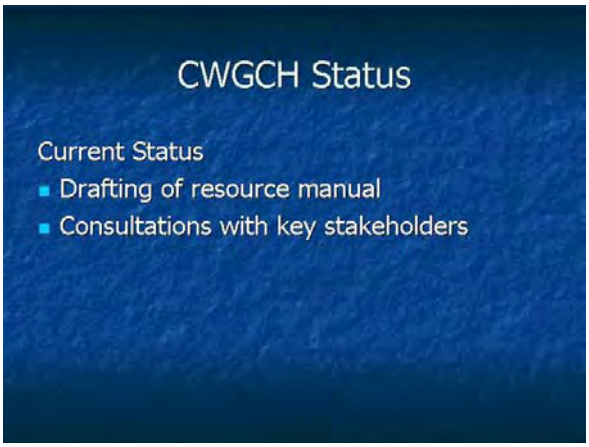

Information Systems
Critical for tracking, follow-up, seamless transition from one program stage to the next, and program evaluation
Standard software packages are available
In-depth understanding of the goals of the program and use of the information is essential when custom designing a program
Consent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


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

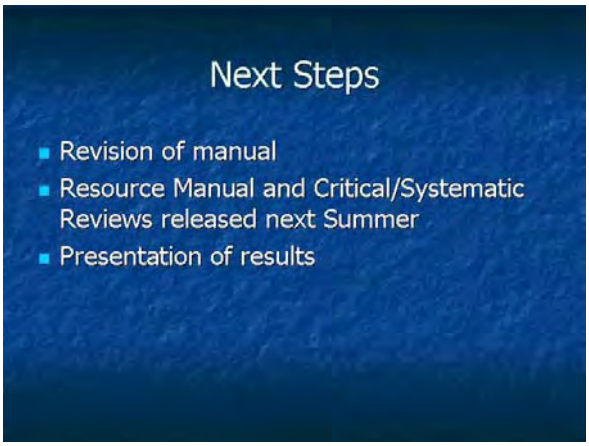

Communication Systems
A broad-based communications/public relations program is invaluable
Parents, professional groups and consumers should receive information by multiple means
Different groups prefer different information formats
Should result in improving awareness, consent, compliance and ultimately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

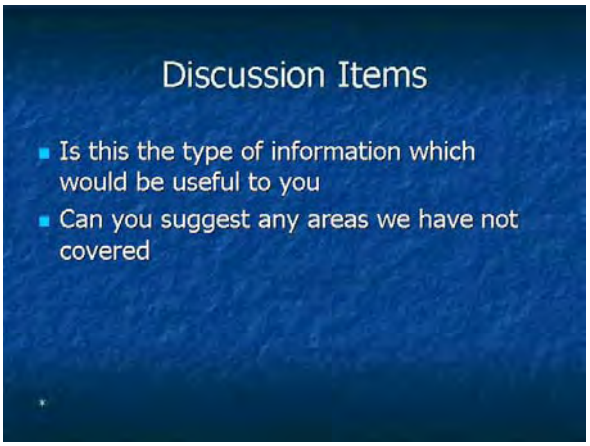

Infrastructure: Conclusion
EHDI programs exist within social, cultural and political values and contexts
Being prepared with evidence such as the resource manual


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

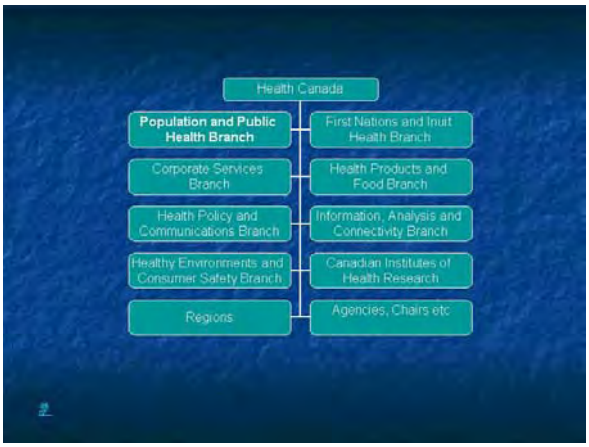

Consultations
Provincial/Territorial or Regional
Key stakeholders
Comment on Draft resource manual
Comments used to revise resource manual so that it is applicable to all jurisdictions in Canada

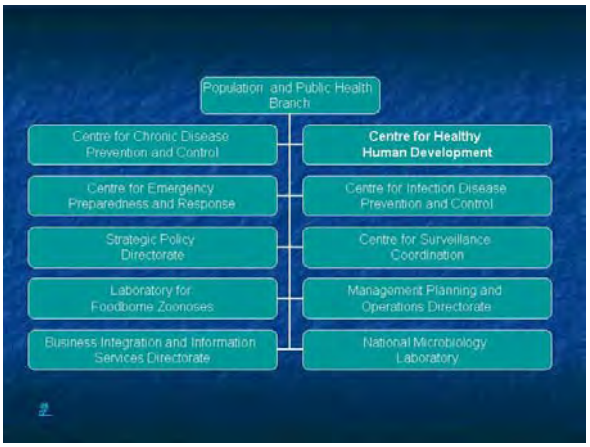

Next Steps
Revision of manual
Resource Manual and Critical/Systematic Reviews released next Summer
Presentation of results


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 Paediatric Society
Canadian Association of Educators of the Deaf and Hard of Hearing
Institut national de santé publique du Québec (INSPQ)
The College 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 Otolaryngology-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


Health Canada
Population and Public Health Branch
First Nations and Inuit Health Branch
Corporate Services Branch
Health Products and Food Branch
Health Policy and Communications Branch
Information, Analysis and Connectivity Branch
Healthy Environments and Consumer Safety Branch
Canadian Institutes of Health Research
Regions
Agencies, Chairs etc


Population and Public Health Branch
Centre for Chronic Disease Prevention and Control
Centre for Healthy Human Development
Centre for Emergency Preparedness and Response
Centre for Infection Disease Prevention and Control
Strategic Policy Directorate
Centre for Surveillance Coordination
Laboratory for Foodborne Zoonoses
Management Planning and Operations Directorate
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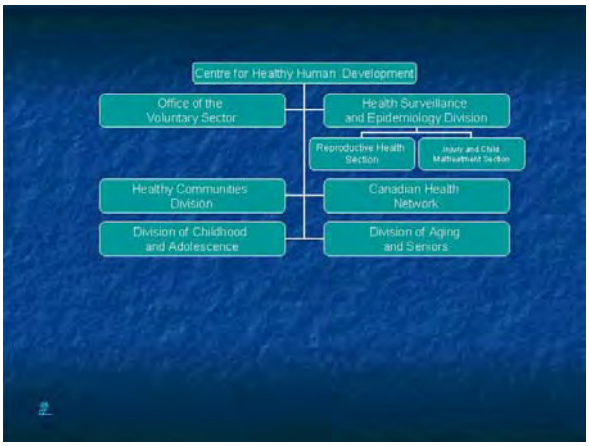
Centre for Healthy Human Development
Office of the Voluntary Sector
Health Surveillance and Epidemiology Division
Reproductive Health Section
Healthy Communities Division
Canadian Health Network
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Division of Aging and Seniors


CWGCH@hc-sc.gc.ca

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

1

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

2

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' CHL)

'Refer' (+ ve screen) => high risk (eg ~1/20)
'Pass' (- ve screen) => low risk (eg ~1/5000)

3

An Ideal Screening Test is:

- *Safe*
- *Accurate*
- *Objective*
- *Quick and easy*
- *Not expensive*

4

Screening Test Types

- *There is NO accurate, behavioural screening test for neonates & young infants*
- *Automated Otoacoustic Emissions (AOAE)*
  - *Transient Evoked, (A)TEOAE*
  - *Distortion Product, (A)DPOAE*
- *Automated Auditory Brainstem Response (AABR)*

5

Automated OAE Screeners

Can discriminate thresholds > < 35-40 dBHL
No standard refer criteria: default + options
No standard parameters, some consensus
Similar performance for TEOAE, 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, $6-10k
Typical test duration 2-5 minutes

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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: OAE+AABR & frequency-specific AABR

7

'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)

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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%)

9

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%)

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Relationships between sensitivity and specificity

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

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

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

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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)

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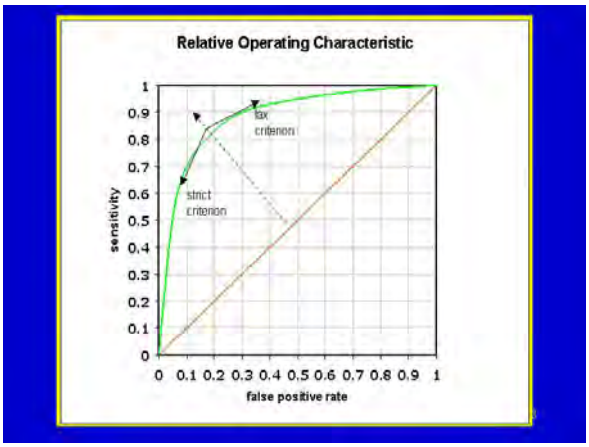

How to compare test accuracy?

Sensitivity or specificity alone are not useful

Sensitivity AND specificity may be insufficient, eg (0.9,0.85) vs (0.88,0.87)?

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

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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*

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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 initial screens only

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Best sensitivity/specificity data

US NIH: Norton et al 00, Ear Hear 21(5):508

2,995 at-risk screened with OAE (T & DP), AABR
Reliable frequency-ear-specific VRA 8-12 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

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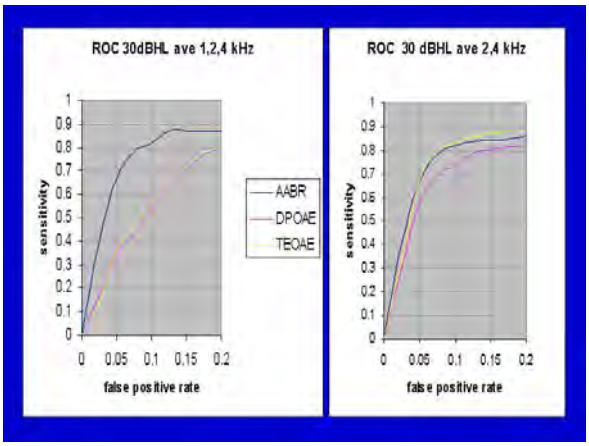

Limitations of NIH study data

Small sample size (56 infants with hearing loss)

Longitudinal validation will lower sensitivity estimates (-ve 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)

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Screening outcome measures

Positive predictive value

PPV = probability that a baby with +ve screen will have the target disorder,
= # true +ve/total # +ve

Negative predictive value

NPV = probability that a baby with -ve screen will NOT have the target disorder,
= # true -ve/total # -ve

Number needed to screen (NNS)
= # screened/ yield (# true positives)

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Effect of sensitivity & specificity on screening outcomes

Example

Sensitivity .9, specificity .95, prevalence 5/1000

	+	450	PPV 450/5,325
PHI	500	>S	NPV 92,625/92,675
100,000	-	50	
	+	4,875	Refer rate 5,325/100,000
N	95,500	>S	FP Rate 4,875/95,500
	-92,625		Yield 450 per 100,000
			NNS 100,000/450

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Effect of specificity on PPV

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

Sensitivity .9, specificity .98, prevalence 5/1000
PPV 450/2,440 = 18%, 1 baby in 6 has PHL

Sensitivity .9, specificity .99, prevalence 5/1000
PPV 450/1,445 = 31%, 1 baby in 3 has PHL

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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

23

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)

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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

25

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

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Effects of audiometric criteria on prevalence

Lower criterion dBHL, higher prevalence

Unilaterals included, higher prevalence

Broader frequency range, lower prevalence

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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

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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 serviced. Adjusted for -ve bias.

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Best prevalence data

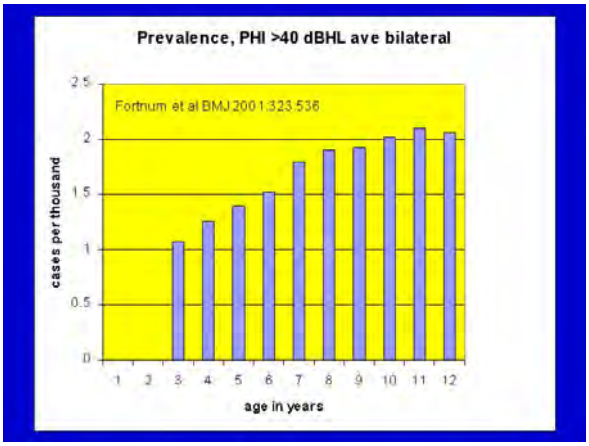
Fortnum H et al, BMJ 2001;323:536

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

30



Other best prevalence data

Wessex UNHS trial, Lancet 98;352:1957-64
prevalence (>40 ave, bilat) 0.94/1000

Prieve B, NY State UNHS, Ear Hear 00;21:85-91
prevalence (>20 any freq, uni/bi) 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

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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

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Must decrease false-positives, increase positive predictive value

Sensitivity .9, specificity .95, prevalence 2/1000
PPV 180/5,170 = 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

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Protocol sensitivity, specificity for 2-stage series screening

Prevalence 2/1000
Tests A & B, sensitivity .9, specificity .9
PPV 180/10,160 = 1.8%, 1 baby in 56 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

35

Best UNHS Outcome Data

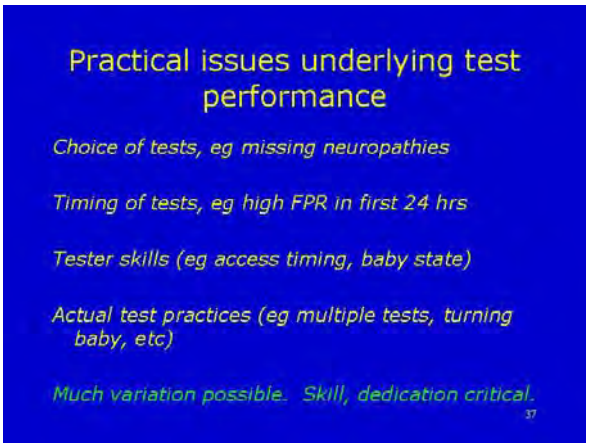
Prieve et al, Ear Hear 00;21:85 (NY State)

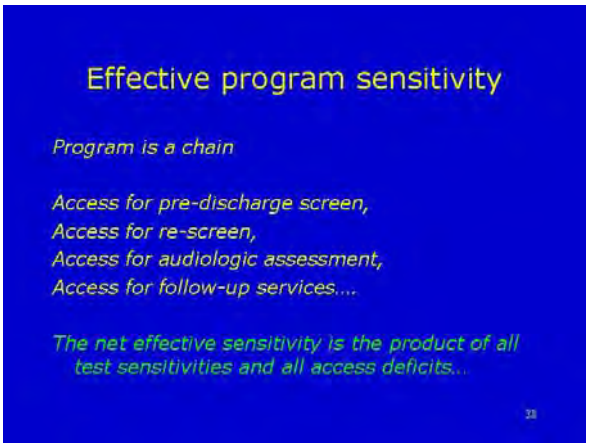
Typical protocol: two-stage, two technology
S1 (TEOAE=>ABR) refer => S2 (TEOAE=>ABR)

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 2.8/1000

36


Practical issues underlying test performance
Choice of tests, eg missing neuropathies
Timing of tests, eg high FPR in first 24 hrs
Tester skills (eg access timing, baby state)
Actual test practices (eg multiple tests, turning baby, etc)
Much variation possible. Skill, dedication critical.
37


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...


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
39
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

- JCIH 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 minimally low- and 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

Gravel & Hood, 1999

Numerous Pitfalls in Determining Auditory Status of Infants

Can seriously influence provision of timely & appropriate intervention

US Preventative Services Task Force (USPSTF) (Agency for Health Care Research & Quality) issued "*Recommendation on Newborn Hearing Screening*" – Oct 2001

- 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?

US Preventative Services Task Force (USPSTF) (Agency for Health Care Research & Quality) "*Recommendation on Newborn Hearing Screening*" – Oct 2001

- 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 8-12 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 (Vohr et al., 1998. *J Pediatr* 113:353-7)
  - reported on outcomes of infants screened between 1993-1996.
  - 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 (1996) 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 (Dalzell et al. 2000)
- Age at diagnosis of hearing loss in NYSDP:
  - 81% (29 of 36 infants) with bilateral PHL (who were aided) confirmed by 5 months (median)
  - NICU infants diagnosed later than well-babies
  - Infants with mild/moderate HL diagnosed later than severe/profound HL (median = 3.5 mo versus 2.0 mo, respectively)
  - Some definitive confirmations took months longer

NYS NHS Demonstration Project
(Dalzell et al. 2000)

- Reasons for delay in audiological diagnosis
  - Mild hearing loss
  - Parental non-compliance with follow-up
  - Illness or developmental delay
  - "Audiological uncertainty"

Potentially 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

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

Why should we worry about audiological misdiagnoses?

- 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)

False-negative diagnosis
(hearing loss diagnosed as normal hearing)

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

False-Negative Diagnosis
Berlin & Hood, 1993 "Pseudo central HL" *Sem Hear*; 14:215-223

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

False-negative diagnosis
Berlin & Hood, 1993
"Pseudo central HL" *Sem Hear*; 14:215-223

- Aids removed
- Repeated behavioral testing consistent with normal hearing
- Eventual ABR (at 3.5 years) consistent with peripheral SNHL;
  - behavioral results attributed to random head turning and biased examiners.

False-negative diagnosis
(SNHL diagnosed as normal hearing)

- 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"

Gravel, 2000

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: tympanograms only; neither AMEMR or OAE completed

Gravel, 2000

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 diagnosis 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-positive diagnosis
(normal hearing diagnosed as HL)

- Behavioral Assessment PITFALLS:
  - Failure to consider maturational/developmental level of infant when performing behavioral testing
  - Failure to consider attention & motivation during test
  - Failure to use physiologic tests to confirm results
    - AMEMR
    - OAE
    - ABR

Misdiagnosis
(AN diagnosed as SNHL)

- AC-ABR
  - Atypical waveforms
  - Use of single polarity clicks (or alternating clicks)
- Tympanograms normal
- Conclusion: SNHL
- Reality: auditory neuropathy/dysynchrony
- PITFALLS: Failure to examine-
  - Behavioral Responses
  - ABR with separate runs of opposite single polarity clicks
  - OAE
  - AMEMR

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

Gravel, Kurtzberg, Stapells et al., 1989
Sem Hear:10:272-287

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
(Northern & Downs)

	NM	FM Tone	Speech	Resp	Startle
0-6 wk	50-70	75	40-60	Eye-widen blink, startle, etc.	65
4-7 mos	40-50	50	20	Head turn, listens	65

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

BOA should not be used to:

- ✓screen hearing
- ✓estimate hearing thresholds
- ✓define the settings of hearing aids

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, 1996

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, 1996

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; *JSHD*--1987
- Reinforcement Duration
  Culpepper & Thompson; *Ear & Hearing* --1984
- Reinforcement Schedule
  Primus *JSHR*--1985

Visual Reinforcers

- Dark smoked-Plexiglas comparments:
  - 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 Visual Reinforcement

- displays located 90º to ONE side of the infant
- toys housed in dark smoked Plexiglas cabinets
- toy cabinets located adjacent to loudspeaker

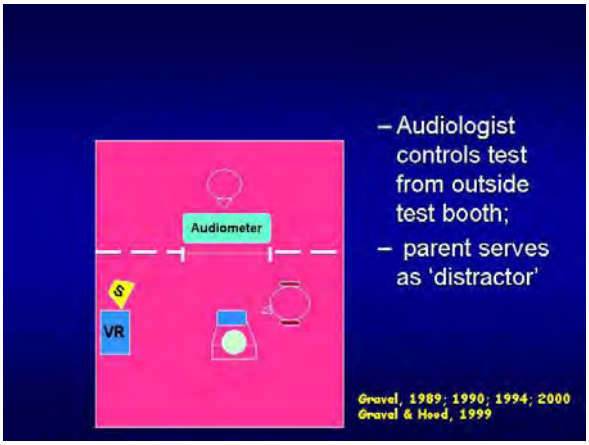
Gravel, 1989; 1990; 1994; 2000
Gravel & Hood, 1999

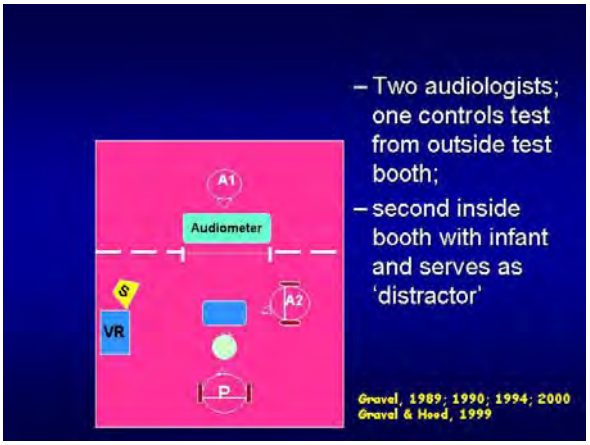
Factors in VRA Use of Distraction

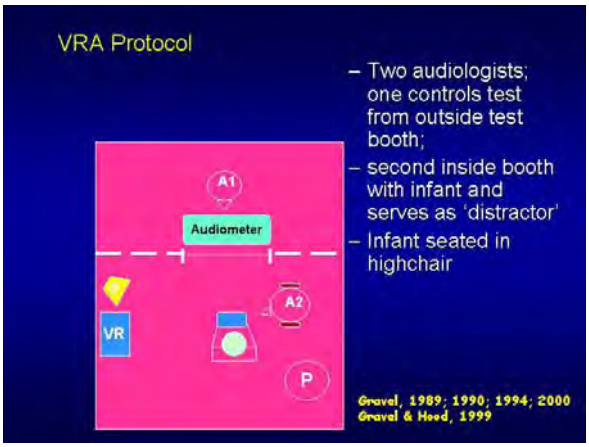
- Decreases the number of false-positive responses
- Reduces the duration of inter-trial intervals
- Prolongs the period of time during which the infant's behavior is appropriate for testing

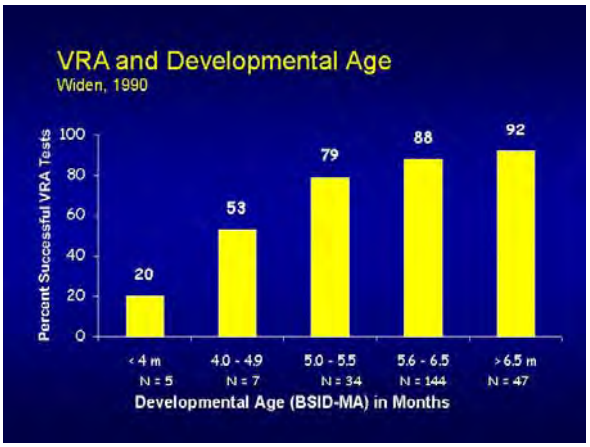
Factors in VRA Test Arrangement

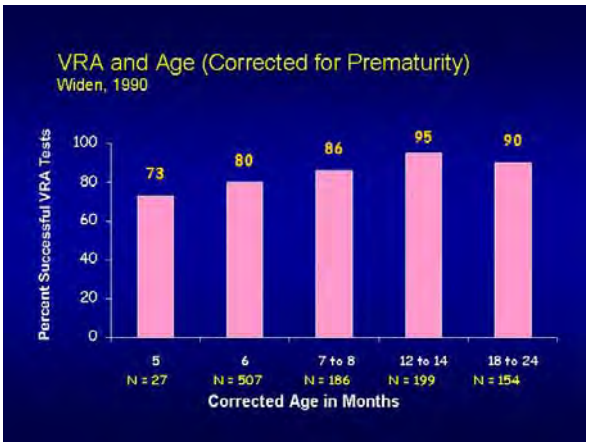
- Infant seated on parent's lap:
  - Audiologist located in test booth, sits opposite (audiometer located within test booth
  - Audiologist controls distraction, presentation of test signals & delivery of reinforcement);

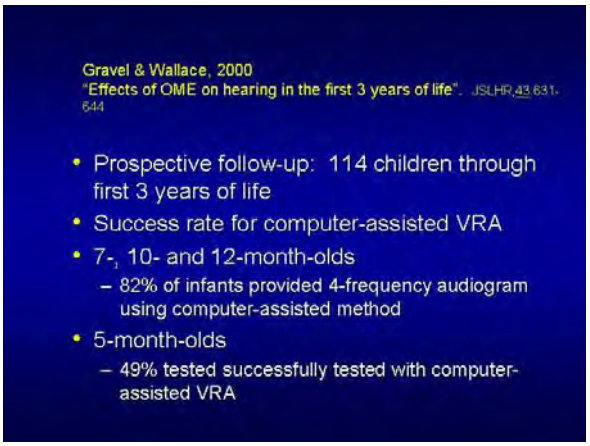

– Audiologist controls test from outside test booth;
– parent serves as 'distractor'
Audiometer
S
VR
Gravel, 1989; 1990; 1994; 2000
Gravel & Hood, 1999


– Two audiologists; one controls test from outside test booth;
– second inside booth with infant and serves as 'distractor'
A1
Audiometer
S
VR
A2
P
Gravel, 1989; 1990; 1994; 2000
Gravel & Hood, 1999


VRA Protocol
– Two audiologists; one controls test from outside test booth;
– second inside booth with infant and serves as 'distractor'
– Infant seated in highchair
A1
Audiometer
A2
VR
P
Gravel, 1989; 1990; 1994; 2000
Gravel & Hood, 1999


VRA and Developmental Age
Widen, 1990
Percent Successful VRA Tests
100
80
60
40
20
0
20
53
79
88
92
< 4 m
N = 5
4.0 - 4.9
N = 7
5.0 - 5.5
N = 34
5.6 - 6.5
N = 144
> 6.5 m
N = 47
Developmental Age (BSID-MA) in Months


VRA and Age (Corrected for Prematurity)
Widen, 1990
Percent Successful VRA Tests
100
80
60
40
20
0
73
80
86
95
90
5
N = 27
6
N = 507
7 to 8
N = 186
12 to 14
N = 199
18 to 24
N = 154
Corrected Age in Months


Gravel & Wallace, 2000
"Effects of OME on hearing in the first 3 years of life". JSLHR 43 631-644
• 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*;21 471-487

- Specified a stringent VRA protocol incorporating signal and control trials
- 3134 infants (NICU or WBN with risk factors)
- 95.6% of infants; (N = 2995) successfully conditioned
- 90% completed full VRA protocol: 8 minimal response levels; 4 in each ear

Widen et al., 2000
"Hearing status at 8-12 months corrected age using VRA protocol" *Ear Hear*;21: 471-487

- 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" *BJA*

- 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 ≥ 8 months ≥ 2 MRL
  - 25% aged ≤ 6 months ≥ 1 MRL.

Earphones

- Younger infants retain earphones better than older (Nozza & Wilson, 1984; 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 = 2995) successfully conditioned;
  - many (not all) successfully completed full protocol: 8 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 (internally-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)

"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

Intelligent VRA: IVRA (IHS, Miami, FL)

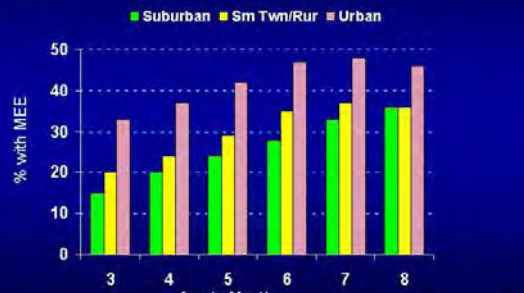
- Computer-assisted, single examiner, bias-free procedure
- 4-button hand-held response box interfaces with computer.
- 4 complex visual reinforcers randomly activated for correct responses.
- Quantifies false-positive rate and attention & motivation during test
- Standard, optimized algorithm, screening

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 = 2253) 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 (Shurin et al 1977, Marchant et al 1984, McKinley et al 1997, Baldwin 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 (eg., 20 dB) and light and signal are paired again

2-year-olds

Test Type

	# Resp	Failed conditioning
• VRA	11.4	0% (0/15)
• PLAY	28.3	32% (7/23)
• VROCA	21.4	17% (3/18)

Thompson, Thompson & Vethivelu, 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 (Widen, 1993)
    - Specify signal & control trials
    - Computer-assisted VRA procedures

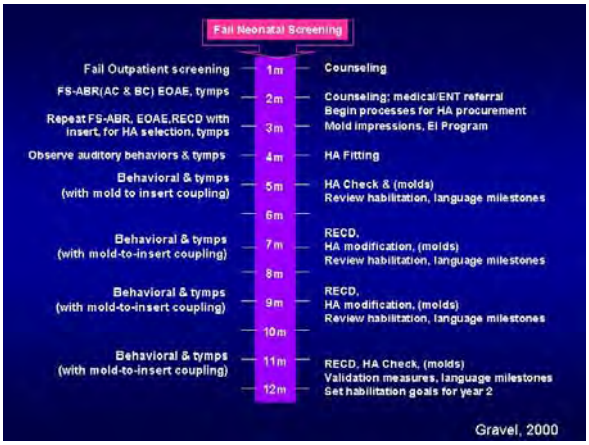
Avoiding Audiological Pitfalls in the Assessment of Infants & Young Children

- Follow test protocols that are evidence-based and time efficient
- Always provide comprehensive audiological evaluation at the initial referral.
- Repeat measures whenever 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 consortial 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 case 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 seems 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.


North American Conference on Deafness Screening and Intervention in Early Childhood. Quebec city, January 2003.
Acquired hearing loss in children
Michel Picard, B.Ps., Ph.D.
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Université de Montréal
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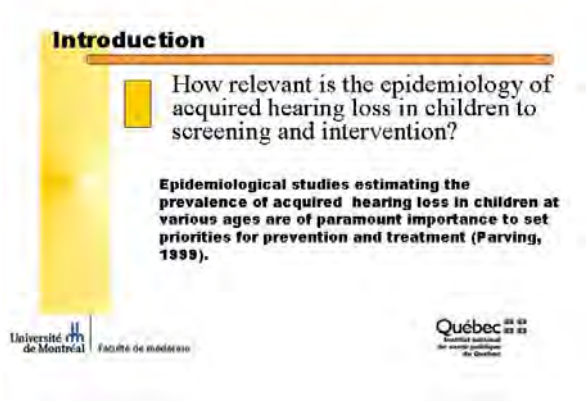
Faculté de médecine
Québec


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 (Parving, 1999).
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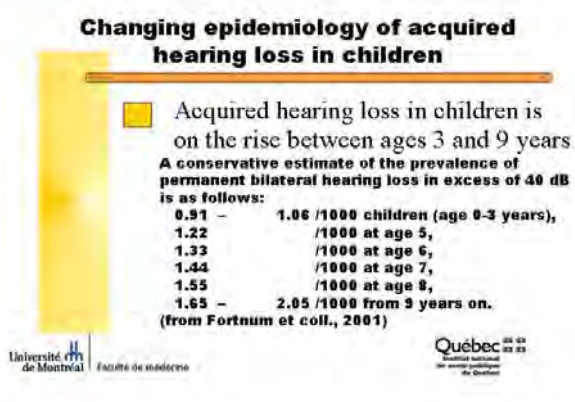
Changing epidemiology of acquired hearing loss in children
Acquired hearing loss in children is on the rise between ages 3 and 9 years
A conservative estimate of the prevalence of permanent bilateral hearing loss in excess of 40 dB is as follows:
0.91 – 1.06 /1000 children (age 0-3 years),
1.22 /1000 at age 5,
1.33 /1000 at age 6,
1.44 /1000 at age 7,
1.55 /1000 at age 8,
1.65 – 2.05 /1000 from 9 years on.
(from Fortnum et coll., 2001)
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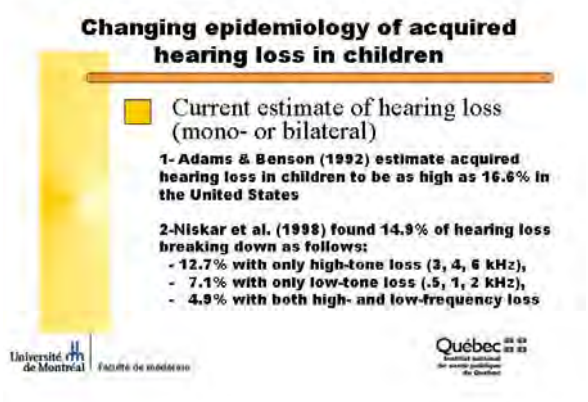
Changing epidemiology of acquired hearing loss in children
Current estimate of hearing loss (mono- or bilateral)
1- Adams & Benson (1992) estimate acquired hearing loss in children to be as high as 16.6% in the United States
2-Niskar et al. (1998) found 14.9% of hearing loss breaking down as follows:
- 12.7% with only high-tone loss (3, 4, 6 kHz),
- 7.1% with only low-tone loss (.5, 1, 2 kHz),
- 4.9% with both high- and low-frequency loss
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Changing epidemiology of acquired hearing loss in children
Population characteristics:
Niskar et al. (1998) studying 6189 children aged 6-19 years
Sex
High-tone loss was more frequent in boys
Socio-economic variables
Hearing loss was more frequent in children from low socio-economic groups
Ethnic origin
Greater risk for children of Mexican origin living in the United States
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Changing epidemiology of acquired hearing loss in children
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 German Registry for hearing loss in children for period 1996-00, Finckh-Kraemer et al. (2000) determined diagnoses of mild hearing loss at 6;2 years on average, moderate one at 4;4, severe and profound ones at 2;5 and 1;9 years, respectively
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Changing epidemiology of acquired hearing loss in children
Known causes (cont.)
3- Improved perinatal medical care results in more surviving infants with perinatal complications (Streppel et al.,1998). In particular, reduced mortality of prematures with very low birth/gestational age results in more infants with hearing loss (Cano et al., 2001)
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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 et Hind, 1999):
More developed countries 4.01%
Less developed countries 24.71%
Africa 5.39%
North America 0.0 %
Latin America 2.5 %
Oceania 0.11%
Asia 17.1 %
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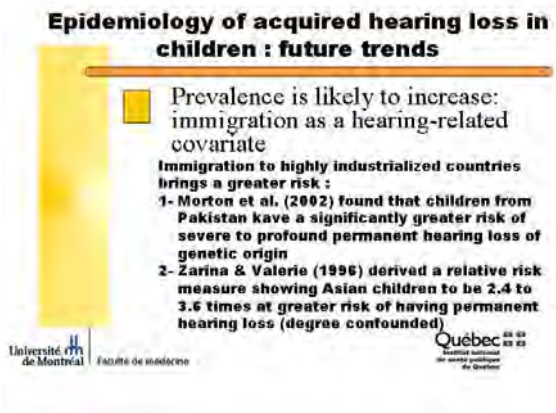
Epidemiology of acquired hearing loss in children : future trends
Prevalence is likely to increase: immigration as a hearing-related covariate
Immigration to highly industrialized countries brings a greater risk :
1- Morton et al. (2002) found that children from Pakistan kave a significantly greater risk of severe to profound permanent hearing loss of genetic origin
2- Zarina & Valerie (1996) derived a relative risk measure showing Asian children to be 2.4 to 3.6 times at greater risk of having permanent hearing loss (degree confounded)
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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 :
3- Compared toreference white caucasians, David et al. (1996) attributed a four times greater risk factor of having hearing lossin children from Hispanic communities living in the United States (Cuban and Puerto Rican, in particular)
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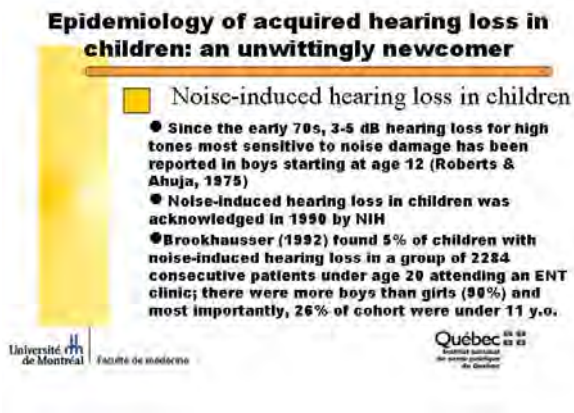
Epidemiology of acquired hearing loss in children: an unwittingly newcomer
Noise-induced hearing loss in children
Since the early 70s, 3-5 dB hearing loss for high tones most sensitive to noise damage has been reported in boys starting at age 12 (Roberts & Ahuja, 1975)
Noise-induced hearing loss in children was acknowledged in 1990 by NIH
Brookhausser (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 (90%) and most importantly, 26% of cohort were under 11 y.o.
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Epidemiology of acquired hearing loss in children: an unwittingly newcomer
Noise-induced hearing loss in children (cont.)
Broste et al. (1989) have found double 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 6-19 year olds is currently estimated at 12.5% in the United States (Niskar et al., 2001)
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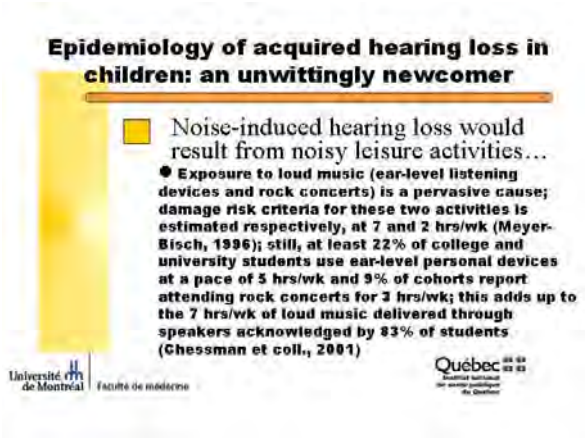
Epidemiology of acquired hearing loss in children: an unwittingly newcomer
Noise-induced hearing loss would result from noisy leisure activities…
Exposure to loud music (ear-level listening devices and rock concerts) is a pervasive cause; damage risk criteria for these two activities is estimated respectively, at 7 and 2 hrs/wk (Meyer-Bisch, 1996); still, at least 22% of college and university students use ear-level personal devices at a pace of 5 hrs/wk and 9% of cohorts report attending rock concerts for 3 hrs/wk; this adds up to the 7 hrs/wk of loud music delivered through speakers acknowledged by 83% of students (Chessman et coll., 2001)
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Epidemiology of acquired hearing loss in children: an unwittingly newcomer
Noise-induced hearing loss would result from noisy leisure activities… (cont.)
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 46% of reported cases compared to 12% for those listening to loud music (Brookhausser, 1992)
Among children 10 years and under, participating to noisy leisure activities with a parent is the leading cause of exposure (Brookhausser, 1992)
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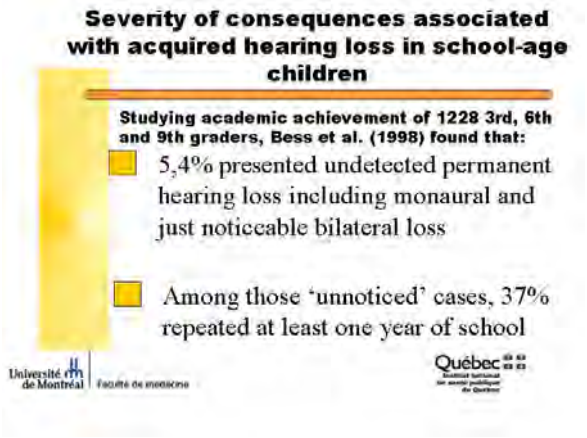
Severity of consequences associated with acquired hearing loss in school-age children
Studying academic achievement of 1228 3rd, 6th and 9th graders, Bess et al. (1998) found that:
5,4% presented undetected permanent hearing loss including monaural and just noticeable bilateral loss
Among those 'unnoticed' cases, 37% repeated at least one year of school
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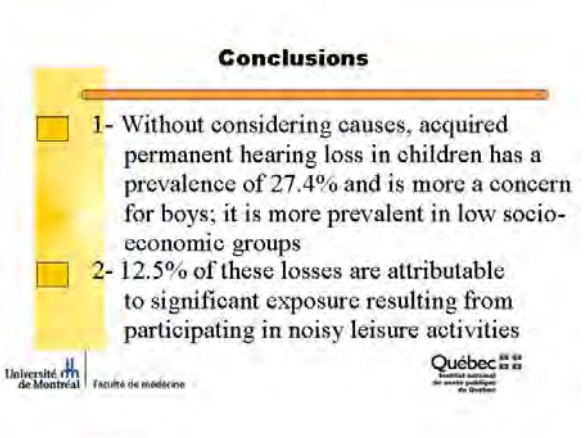
Conclusions
1- Without considering causes, acquired permanent hearing loss in children has a prevalence of 27.4% and is more a concern for boys; it is more prevalent in low socio-economic groups
2- 12.5% of these losses are attributable to significant exposure resulting from participating in noisy leisure activities
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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
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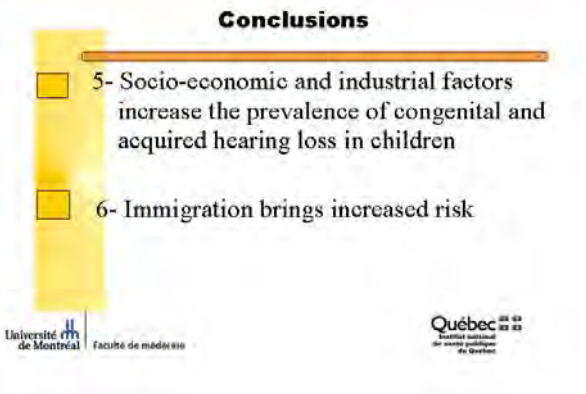
Conclusions
5- Socio-economic and industrial factors increase the prevalence of congenital and acquired hearing loss in children
6- Immigration brings increased risk
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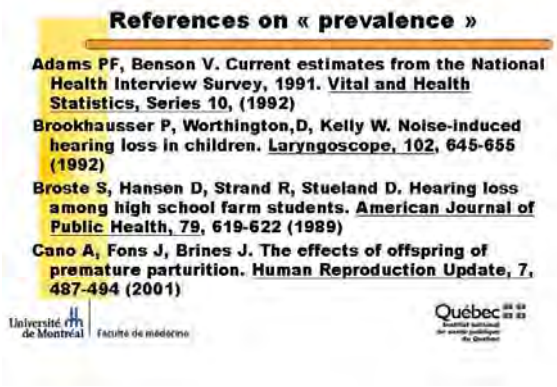
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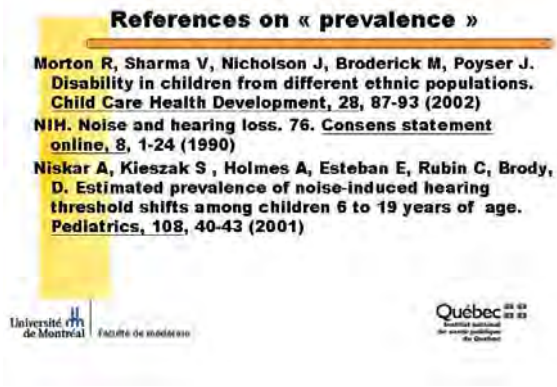
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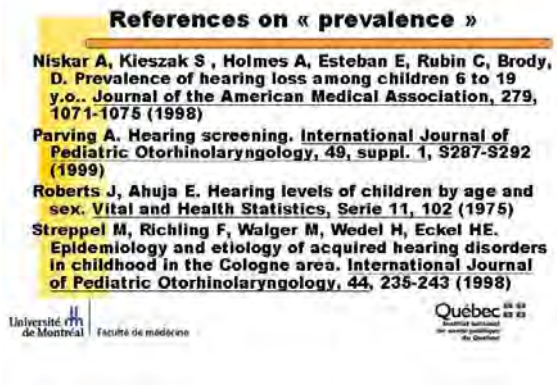
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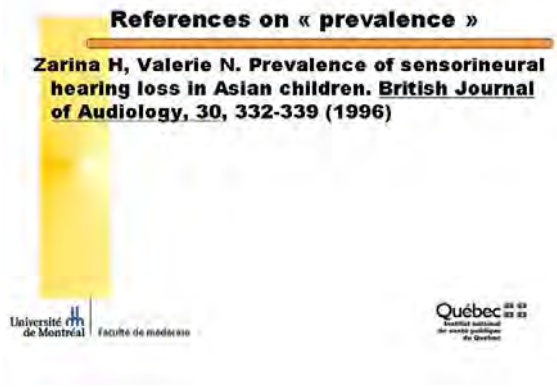
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5.4% of just noticeable hearing losses in the school population go undetected although associated with a high risk of failing a degree.
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THE USER'S PERSPECTIVE

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.