Babies with Hearing Loss: First Steps for New Practitioners

Early Hearing Detection and Intervention (EHDI)

• Historical Influences
• Principles and Goals
• Systems Perspective
• Current Status
• Challenges and Resources
Early Hearing Detection and Intervention

Historical Influences

- 1960s – Work of Marion Downs
- 1969 – Joint Committee on Infant Hearing
- 1990 – Healthy People 2000
- 1993 – National Institutes of Health Consensus Development Panel
Early Hearing Detection and Intervention
Historical Influences

- 1990s – Rhode Island Hearing Assessment Project, other Universal Newborn Hearing Screening programs
- 1990s – State Legislation
- 1998 – Walsh Act
Early Hearing Detection and Intervention
Historical Influences

- Technology
- Support by Advocacy and Professional Organizations
- Federal Funding
  - Maternal and Child Health Bureau
  - Centers for Disease Control and Prevention
Early Hearing Detection and Intervention

Healthy People 2010

Goal 28-11

Increase the proportion of newborns who are screened for hearing loss by age 1 month, have audiologic evaluation by age 3 months, and are enrolled in appropriate intervention services by age 6 months.
Early Hearing Detection and Intervention
Principles and Goals

• EHDI Principles - Joint Committee on
Infant Hearing Year 2000 Position
Statement
• EHDI National Goals - Centers for
Disease Control and Prevention
Early Hearing Detection and Intervention
Principles and Goals

**JCIH 2000**
Principle 1 – All infants have access to hearing screening using a physiologic measure.

**CDC**
Goal 1 – All newborns will be screened for hearing loss before one month of age, preferably before hospital discharge.
Early Hearing Detection and Intervention
Principles and Goals

JCIH 2000
Principle 2 – All infants who do not pass the birth admission screen...begin appropriate audiologic and medical evaluations...before 3 months of age.

CDC
Goal 2 – All infants who screen positive will have a diagnostic audiologic evaluation before 3 months of age.
Early Hearing Detection and Intervention
Principles and Goals

JCIH 2000

Principle 3 – All infants with confirmed permanent hearing loss receive services before 6 months of age in interdisciplinary intervention programs

CDC

Goal 3 – All infants identified with a hearing loss will receive appropriate early intervention services before 6 months of age.
Early Hearing Detection and Intervention
Principles and Goals

**JCIH 2000**

**Principle 4** – All infants who pass newborn hearing screening but who have risk indicators for other auditory disorders… receive ongoing audiologic and medical surveillance and monitoring for communication development.

**CDC**

**Goal 4** – All infants and children with late onset, progressive or acquired hearing loss will be identified at the earliest possible time.
Early Hearing Detection and Intervention
Principles and Goals

JCIH 2000

Principle 5 – Infant
and family rights are
guaranteed through
informed choice,
decision-making,
and consent.
Early Hearing Detection and Intervention
Principles and Goals

J CIH 2000

Principle 6 – Infant hearing screening and evaluation results are afforded the same protection as all other health care and educational information.

CDC
Early Hearing Detection and Intervention
Principles and Goals

JCIH 2000 CDC

Goal 5 - All infants with hearing loss will have a medical home.
Early Hearing Detection and Intervention Principles and Goals

JCIH 2000

Principle 7 – Information systems are used to measure and report the effectiveness of EHDI services.

CDC

Goal 6 – Every state will have a complete EHDI Tracking and Surveillance System that will minimize loss to follow-up.
Early Hearing Detection and Intervention
Principles and Goals

JCIH 2000
Principle 8 – EHDI programs provide data to monitor quality, demonstrate compliance with legislation and regulations, determine fiscal accountability...and maintain community support

CDC
Goal 7 – Every state will have a comprehensive system that monitors and evaluates the progress towards the EHDI Goals and Objectives.
Early Hearing Detection and Intervention
Systems Perspective

Functions

Screening
Re-screening
Audiological Diagnostic Evaluation
Early Intervention, including amplification
Medical Home
Specialty Evaluations
Parent/Family Support
Early Hearing Detection and Intervention
Systems Perspective

Providers

Birthing Facilities
Audiologists
Primary Care Providers
Medical Specialists (ENT, genetics, ophthalmology, etc.)
Early Intervention (Early Intervention - Part C, Children with Special Health Care Needs, Parent Information Centers, Educators of the Deaf, Speech-Language Pathologists, Early Head Start, etc.)
Parent/Family Support (Hands and Voices, Family Voices, www.babyhearing.org, etc.)
Early Hearing Detection and Intervention
Systems Perspective

**Screening**
before 1 month

**Diagnosis**
before 3 months

**Intervention**
before 6 months

Medical Home

Data Management and Tracking

Program Evaluation and Quality Assurance

Family Support
Early Hearing Detection and Intervention

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

Birth
- Hospital-based Infant Screening
  - Results sent to Medical Home

Before 1 Month
- Outpatient Screening
  - Results sent to Medical Home

Before 3 Months
- Pediatric Audiologic Evaluation
  - Otoscopic Inspection
  - Child & Family History
  - Tympanometry
  - Audiometric Test
- Referral to IDEA Part C

Before 6 Months
- Report to State
- Evaluate
- Name
- Address
- Date
- Signature

Ongoing Care of All Infants From the Medical Home Provider
- Provide parents with information about hearing, speech, and language milestones
- Identify and appropriately treat middle ear disease
- Provide hearing screening assistance and referral as needed
- Provide ongoing clinical surveillance and referral to appropriate resources
- Identify and refer for audiologic monitoring infants who have the following risk indicators for late onset hearing loss
  - Potential or known hearing loss
  - Family history of permanent childhood hearing loss
  - Ototoxic or other findings associated with a syndrome known to include a sensorineural or conductive hearing loss or perinatal or neonatal conditions
  - Perinatal infections associated with sensorineural hearing loss including bacterial meningitis
  - Herpes, rubella, syphilis, and congenital toxoplasmosis
  - Neonatal indicators—specifically hyperbilirubinemia at a serum level requiring exchange transfusion, persistent pulmonary hypertension of the newborn associated with mechanical ventilation, and conditions requiring the use of extracorporeal membrane oxygenation
  - Syndromes associated with progressive hearing loss such as neurofibromatosis, osteogenesis imperfecta, and Usher syndrome
  - Neurodegenerative disorders, such as Hunter syndrome, or sensory motor neuropathies, such as Friedreich ataxia and Charcot-Marie-Tooth disease
  - Head trauma
  - Recurrent or persistent otitis media with effusion for at least 3 months

January 2003
Early Hearing Detection and Intervention

Current Status

- EHDI is part of the public health system in the US, with EHDI coordinators in all 50 states, District of Columbia
- 38 states have EHDI legislation, with 28 passed since 1998
- In 2004, an estimated 91.7% of newborns were screened for hearing loss
- In 2004, 3,568 were identified with hearing loss (incidence = 1.11 per thousand)
- In 2004, 52.0% needing an audiologic evaluation were lost to system
Early Hearing Detection and Intervention Challenges

- Shortage of pediatric audiologists for evaluation and intervention
- Referrals for diagnostic audiologic evaluations not being made consistently
- Inadequate third-party reimbursement
- Lack of adequate early intervention services, especially for mild hearing loss
- Tracking and management of failed screenings due to data systems
- Families don’t understand the advantages of early identification and intervention.
Early Hearing Detection and Intervention
Resources

Maternal and Child Health Bureau, Universal Newborn Hearing Screening -
http://mchb.hrsa.gov/programs/genetics/hearingscreen.htm

Centers for Disease Control and Prevention, Early Hearing Detection and Intervention -
http://www.cdc.gov/ncbddd/ehdi/default.htm

National Institute on Deafness and Other Communication Disorders - http://www.nidcd.nih.gov

National Center for Hearing Assessment and Management -
http://www.infanthearing.org

Joint Committee on Infant Hearing -
http://www.jcih.org
Early Hearing Detection and Intervention

References


National EHDI Goals. www.cdc.gov/ncbddd/ehdi/nationalgoals.htm


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Protocol for Provision of Amplification for Infants & Young Children

**Objectives**

1. Delineate protocol steps
2. Describe the importance of real-ear verification in HI fitting components
3. Initiate development of resource base
Foundation of the Amplification Process: Assessments & Referrals

- Audiological
- Medical
- Educational
What is *Ideal*?

*Guidance from:*

- Joint Committee on Infant Hearing (JCIH)
- Pediatric Working Group (‘96)
- AAA Pediatric Amplification Guidelines (‘04)
- ASHA Guidelines for Audiologic Assessment of Children (‘04)
- ASHA Roles, Knowledge and Skills re: above (‘06)
- American College of Medical Genetics (‘02)
- Data published in peer-reviewed journals
Assessment & Referrals

1. Confirm and quantify hearing status

   • ABR (emerging use of ASSR, cautious application to infants at this time)
   • High-frequency tympanometry
   • Otoacoustic emissions (OAE)
   • Behavioral audiometric tests as developmentally appropriate
Proposed JCIH Recommendation

- Includes screening for neural HL (AN/AD) in infants admitted to NICU for ≥ 5 days
- ABR will be needed for babies in NICU for 5 or more days
Proposed J CIH Recommendation

- At least one ABR for children under 3 years for confirmation of permanent hearing loss
Proposed JCIH Recommendation

- If risk factor for HL, one diagnostic audio by 30 mos of age
- More frequent if risk for late onset or progressive HL
  - CMV
  - ECMO
Proposed J CI H Recommendation

• If permanent HL, fit with amplification in one month of Dx
Assessment & Referrals

2 Refer back to medical home to initiate medical evaluations

- Otolaryngology—medical clearance
- Genetics—etiology evaluation
- Ophthalmology
  - determine vision status
3 Referral for early intervention & support

- Early development network
- IFSP development
  - Baseline communication
  - Baseline developmental assessment
  - Parent to parent support
With this Foundation:
Early & Appropriate Amplification is Possible

- Aim for the ideal
- Anticipate obstacles
- Address challenges
Anticipate challenges:
**Delays** in Fitting Amplification
by Harrison, et al, 2003, *Ear & Hearing*

- Problems with scheduling appointments
- Need for repeat tests to specify hearing loss
- Suspicion of auditory neuropathy
- Money concerns for purchase of devices

*delays of > 1 month after diagnosis of hearing loss*
Additional Challenges:

- Health status of baby
- Cooperation
What the audiologist can do...

- Identify and confirm hearing loss by 3 mos
- Fit amplification within one month of confirmation
- Initiate early intervention before 6 mos of age
- Provide baby and family friendly environment
- Implement pediatric protocols
- Assist in identifying financial resources
Process/Protocol

*SELECTION

VERIFICATION

FOLLOW UP

ORIENTATION

VALIDATION
Selection

Decisions, decisions, decisions

1. Earmolds
2. Devices & Circuitry
3. Electroacoustic targets: based on pediatric prescriptive approach
Earmold Impressions

- Visual inspection
- Estimate length of canal
- **Mark otolight for ~10 mm from canal entrance**
- Insert otoblock
- Double check insertion
- Pediatric sized syringe
Earmold Options—Limited by Size

• Style
  • Shell

• Sound bore
  • Tubing
  • Venting

• Materials (soft)
  • PVC
  • silicone
Feedback Challenges: Size and Growth

- Lubricants
- Feedback *system in HI*
- Frequent remakes needed
- Other options
Selection

- Earmolds
- **Devices & Circuitry**
- Electroacoustic targets based on pediatric prescriptive approach
Selection

Devices & Circuitry

- BTE
- Binaural fitting, in most cases
- Consider fitting unilateral HL
- Flexibility of electroacoustic characteristics
- Tamper-proof features
- FM compatibility
Selection
Circuitry: Review of evidence-based research related to optimal signal processing for children

*Palmer & Grimes, *JAAA 2005*

- WDRC found to be appropriate for
  - Children
  - Mild, moderate and moderately severe
  - To achieve goal of “audibility across a wide frequency bandwidth...across a large range of input levels”
Selection
Circuitry Choices: Evidence-based

• *More Palmer & Grimes, JAAA 2005*

• Use WDRC with these parameters:
  - Low CT (45-55 dB)
  - Moderate CR (1.7 to 2.3)
  - Fast attack time (10 ms)
Selection

Devices & Circuitry

- Anticipate needs over ~ 5 year period
- Other options
  - Color choices
  - FM use at home
  - Multi-memory
  - D-microphone
Selection

- Earmolds
- Devices & circuitry
- Electroacoustic targets based on pediatric prescriptive approach
Selection
Electroacoustic Targets

Data supports need for pediatric prescriptive approach

Desired Sensation Level (DSL) is a method developed for pediatric fittings
Selection

Desired Sensation Level (DSL) targets for children

- Audibility based approach
- Takes into account ear canal acoustics
- Substantiated by research from the UW-Ontario group and others
Selection

• Appropriateness of fitting depends on adequacy and accuracy of frequency-specific threshold data

• Thresholds affect:
  • Targets for amplified speech
  • Targets for RESR
ABR thresholds may over or underestimate behavioral audiometric threshold by ~10-20 dB. Some clinics routinely measure ABR thresholds in 10 dB steps. Trade-off between more frequency-specific info versus more precise thresholds.
Which instruments...?

• Meet targets
  - ✓ Provide audibility
  - ✓ Stay below RESR

• Provide reasonable flexibility for potential hearing changes
DSL m[i/o]v5.0 updates

Not all are implemented in beta version of Verifit

1. RECD predictions by age in one month intervals

2. ABR thresholds can be converted to SPL when entered as nHL or eHL (estimated) or user enters custom correction

3. Corrections for conductive component
DSL updates

4. Multi-stage i/o algorithm for:
   • Expansion portion
   • Linear portion
   • WDRC portion
     • CT varying with hearing loss
   • Output limiting for:
     • broad band (1/3rd octave bands of speech)
     • narrow band signals
DSL updates

4. Multi-channel targets with CR at 1/3 octave band frequencies for single or multi-channel devices

5. Targets for a variety of speech inputs and spectra shapes, and to account for
   - Venting/open fittings
   - Binaural fittings
   - Broadband vs narrow band signals
DSL updates

7. **Prescriptions for noisy environments** (less amplification where less speech importance)

8. **Age-dependent targets/sensation levels** reflecting adult-child preferred listening level differences
   - adults obtain optimal speech intelligibility and loudness at about 6-8 dB below 4.1 targets
Verification

1. Behavioral
2. Electroacoustic
Verification Methods should:
(Per Scollie, 2001)

Show how the instrument processes speech
Show the levels at which output is limited
Be efficient, reliable and valid
Be capable of being used with infants
Amplified Sound Field Thresholds

- Do not tell how the instruments process speech
- Do not tell the levels at which output is limited
- Are not efficient, frequency or level specific
- Cannot be used with infants under 6 months
Aided Sound Field Threshold Uses, when child is developmentally able:

Bone-conduction hearing instruments
Frequency-compression instruments
Parent demonstration
Cochlear implant protocols
As a cross check, not as verification
Electroacoustic Verification: Method of Choice for Infants

*Wiggle Factor:* Most babies, and even some 5 year olds, cannot or will not sit quietly while you perform probe microphone measures!
Therefore, use Simulated real-ear measures.

Real Ear to Coupler Difference (RECD) in combination with coupler measures are a

• valid
• Reliable
• research-supported
• quick and easy to perform

verification option.
RECD is influenced by:

- Ear canal size (esp 0-24 months)
- Impedance characteristics
  - Otitis media w/ effusion
    - larger RECD 200-3000 Hz
  - Tympanostomy tube
    - negative values in low frequencies
RECD factors:

- Probe insertion depth
  - regions of standing waves
  - Bagatto rec insertion depth of ~11 mm from the EC entrance
- Plugged probe tube
- Noise
  - Baby & room

*Tharpe et al data suggests that one ears RECD is a good predictor of the other ear if ears similar.
Electroacoustic Verification is critical:

Noah screen provides estimates of gain and output; can be inaccurate. Hawkins, 2003

simulated values in HI software overestimate gain, esp @ high frequencies.
Orientation: Parent & Child Friendly

- Ask parents what they want to discuss first
- Be sensitive to feelings
- This is a process, NOT a one time event
  - New vocabulary
  - New parental role
- Success of HI use depends on parents
Primary Parental Concerns About Hearing Instruments

Sjolstad, Harrison, Roush, & McWilliam (2001), *AJA*

1. Hearing aid maintenance
2. Appearance of hearing aids
3. Potential benefit
Address

Hearing Aid Maintenance

- Explain, demonstrate, allow practice
- Support with written material
- Provide basic equipment and supplies
- Schedule return visit during first few weeks
Orientation Essentials for Maintenance

• Demo & Practice
  – Earmold Insertion & Hearing Aid Placement
  – Battery Insertion & Ingestion Precautions
  – Daily Listening Check
  – Care and Maintenance Supplies
  – Retention & Loss Prevention

• Teach Troubleshooting
Many Companies have “Care Kits”

- Batteries
- Earmold Blower
- Drying device
- Listening Tube/Steth
- Lubricants
- Retention Device
- Tester for Batteries
- Wax Loop
Child Friendly for Retention

1) Tonehooks

2) Retention
   a) Toupee/double stick tape
   b) Adhesive
   c) Devices
Child Friendly for Loss Prevention

1) “Critter clips”
2) Thin fabric hat
3) “Ear Gear”
When all else fails…
Be sure of the
Loss & damage warranty!
Address Appearance

- Show earmold and hearing instrument examples
- Reassure
- These concerns likely will diminish with time

About 40% of the infants with hearing loss will have other developmental challenges.
Address
Potential Benefit …..Validation
Validation

Purpose

“…to demonstrate the benefits/limitations of a children's aided listening abilities for perceiving the speech of others as well as his or her own speech.” (Pediatric Working Group, 1996)
Validation (PWG, 1996)

When & How?

“...accomplished over time”
“...(as part of) aural rehabilitation process”
“...direct measurement of...auditory performance”
Validation (AAA, 2003):

How?

• Aided speech perception measures
• Functional assessment tools
  – Not validated in infants
Possible Measures

**Objective:** aided speech recognition -- not reliable before 3 years of age

**Subjective**
- COSI-C (Client Oriented Scale of Improvement-Child)
- IT-MAIS (young children w/ profound HL) – Infant Toddler Meaningful auditory integration scale
  [http://www.bionicear.com/printables/it-mais_brochure.pdf#search=%22it-mais%22](http://www.bionicear.com/printables/it-mais_brochure.pdf#search=%22it-mais%22)
- ELF -- Early listening function
  [http://www.phonak.com/com_elf_questionnaire_gb.pdf#search=%22elf%20phonak%22](http://www.phonak.com/com_elf_questionnaire_gb.pdf#search=%22elf%20phonak%22)
- FAPI -- Functional auditory performance indicators
- Phonemic inventory
Planned Follow up:
Typical schedule:
3-month intervals during 1st two years of life
4-6-month intervals after 2 years
Annual thereafter
Data published in peer-reviewed journals

  - 3 DSL articles by Seewald et al, Bagatto et al and Scollie et al
Resources

Joint Committee on Infant Hearing (JCIH)  
jcih.org
Pediatric Working Group (‘96)  
AAA Pediatric Amplification Guidelines (‘04)  
www.audiology.orgASHA
Guidelines for Audiologic Assessment of Children birth to 5 years (‘04)  
asha.org
ASHA Roles, Knowledge and Skills re: above (‘06)  
asha.org
www.infanthearing.org

- The National Center for Hearing Assessment & Management (NCHAM)
  - Extensive information on NHS and EHDI and follow-up
  - Links to pertinent websites
listen-up.org

• Practical tips for parents from parents
  • Feedback control, retention, behavior mod, etc.
• Parent writings
• ADA issues
• IEP issues
• Extensive links to related websites
www.agbell.org

• Information and advocacy for
  • Parents
  • Teens
  • Professionals

• Translates into many languages using worldlingo.com
www.handandvoices.org

—“nationwide non-profit organization dedicated to supporting families and their children who are deaf or hard of hearing, as well as the professionals who serve them.”