Hearing Loss in Infants
Please cite this module using the following:

Faculty Developers:

Sarah Jane Dodd, PhD, Silverman School of Social Work, Hunter College, CUNY, NY, NY
Jeanne Finch, DSW, School of Social Work, Stony Brook University, Stony Brook, NY
Mary Ann Jones, PhD, Silver School of Social Work, New York University, NY, NY
Susan Mason, PhD, Wurzweiler School of Social Work, Yeshiva University, NY, NY
Lynn Spivak, PhD, Adelphi University and Hofstra Universities, Long Island, NY
Brenda Williams-Gray, DSW, Department of Social Work, Lehman College, CUNY, Bronx, NY
Wendy Zeitlin, PhD, Wurzweiler School of Social Work, Yeshiva University, NY, NY
Three Parts of the Ear

• Outer Ear
  o Pinna
    • Shell like structure collects sound waves
  o Ear canal
    • Channels sound waves to the ear drum

• Middle Ear
  o Ear Drum
    • At the end of the ear canal
    • Connected to the ossicles
  o Ossicles
    • 3 tiny bones connected in a chain
    • The first bone (malleus) is attached to the ear drum
    • The last bone (stapes) is in contact with the fluids in the inner ear
Three Parts of the Ear (cont.)

• Inner Ear
  o Cochlea
    • Contains the sense organ for hearing
  o Vestibular Apparatus
    • Contains the sense organ for balance
How we hear
How we Hear

- Sound = vibration of air molecules
- Sound enters the outer ear (1)
- Vibrations are transmitted through the ear canal to the ear drum
- The ear drum transmits vibrations to the ossicles (2)
- The stapes (last ossicle in the chain) transmits vibrations to the inner ear fluids (3)
- Wave like motion is set up in the cochlea which causes sensory cells (hair cells) to be stimulated. This causes the mechanical energy (vibration) to be transformed into electrical (neural) energy.
- Neural impulses are carried through the central auditory pathways to the brain (4)
TYPES OF HEARING LOSS

Categorized by Location of Impairment
Conductive

Sensorineural
Types of Hearing Loss

• CONDUCTIVE
  o Outer or middle ear
  o Impedes transmission of sound to the cochlea
  o Can cause hearing loss ranging from mild to moderate
  o Often is temporary. Many causes of conductive hearing loss are medically treatable
Conductive Hearing Loss

- Outer Ear
  - Wax
  - Infection
  - Congenital Malformation
  - Foreign Body

- Middle Ear
  - Ossicular Chain Problem
  - Cyst
  - Perforated Ear Drum
  - Fluid (otitis media)
Types of Hearing Loss

• SENSORINEURAL
  o Inner ear (cochlea)
  o Impedes transduction of mechanical to neural energy
  o Can cause mild to profound hearing loss
  o Permanent. Damage to the cochlea is not medically treatable
Sensorineural Hearing Loss

• Inner Ear (cochlea)

• Hereditary
• Congenital
• Medication
• Disease
• Noise Exposure
• Age
Hearing Loss in Infants & Children
Congenital Hearing Loss

• **50% Genetic**
  o Inherited from parents
    • Autosomal Recessive or Dominant
  o New mutation
  o May or may not be part of a syndrome

• **50% Acquired**
  o Pre- or perinatal period
  o Maternal infections
  o Prenatal drugs
  o Infection in newborn
  o Ototoxic drugs given to a newborn
  o Trauma to fetus
Risk Factors for Hearing Loss: JCIH, 2007

- Caregiver concern regarding hearing, speech, language
- Family history of permanent childhood HL.
- NICU care of >5 days, or any of following regardless of length of stay: assisted ventilation, ototoxic medications, exchange transfusion, and ECMO
- Intra-uterine TORCH infections, particularly CMV
- Craniofacial anomalies, especially those involving the pinna, ear canal, ear tags, ear pits, and temporal bone anomalies
Risk Factors for Hearing Loss, cont.

- Physical findings associated with a syndrome known to include permanent HL
- Syndromes associated with progressive HL such as NF, osteopetrosis, Usher’s syndrome
- Neurodegenerative disorders, such as Hunter syndrome
- Postnatal infections associated with SNHL especially bacterial meningitis
- Head trauma requiring hospitalization
- Chemotherapy
50% of all Hearing Impaired Infants will have NO risk factors
Therefore, **ALL** newborns must be screened to ensure 100% detection of hearing loss in newborns.
Why is Early Identification & Diagnosis so Important?

- Normal hearing is required for speech & language development
- Babies with intervention by 6 months develop normal speech and language
- Even mild and unilateral hearing loss can negatively impact development
Early Intervention for Hearing Loss

- Infants diagnosed by 6 months of age have significantly better speech and language outcomes than infants diagnosed later.
Expressive Language Scores for Hearing Impaired Children Identified Before and After 6 Months of Age

Chronological Age in Months

Language Age in Months

Identified BEFORE 6 Months

Identified AFTER 6 Months

13-18 mos (n = 15/8)

19-24 mos (n = 12/16)

25-30 mos (n = 11/20)

31-36 mos (n = 8/19)
Early Intervention Reduces Cost to Society

• Early identified infants receiving early intervention are likely to attain oral speech and language skills that will permit them to be educated in a mainstream setting.
How can we ensure early intervention & optimal outcomes?

1-3-6 Model
EHDI 1, 3, 6 Model

• EHDI = Early Hearing Detection and Intervention
• Each State has an EHDI program designed to ensure that all newborns are screened and Early Intervention is provided for babies with hearing loss
• The CDC guidelines for early detection and intervention of hearing loss is known as the 1, 3, 6 model
  o Screening by 1 month of age (preferably before hospital discharge)
  o Diagnosis by 3 months of age
  o Begin intervention by 6 months of age
• This model has been adopted universally
Screening Method

• **Physiologic Screen**
  o Newborns unable to respond behaviorally
  o Hearing is measured in natural sleep by recording a physiological response to sound stimulation
  o Diagnostic tests have been adapted and automated for use in newborn hearing screening
    • Automated administration and scoring of test allows use by non-professional personnel
Physiological Tests used in NBHS

- Auditory Brainstem Response (ABR)
  - Measures brain waves in response to sounds

- Otoacoustic Emissions (OAE)
  - Transient (TEOAE)
  - Distortion Product (DPOAE)
  - Measures response of sensory cells (hair cells) in the cochlea to sound stimulation
Before One Month of Age

- Outpatient re-screening for all failed, missed, or incomplete screenings
  - Early re-screening allows earlier diagnosis
  - Early re-screening is technically easier
  - Early re-screening minimizes parental anxiety
  - Unilateral (one ear) or bilateral (two ear) failure must proceed to full audiology evaluation
Follow-up Model

- Referred from Nursery
  - Re-screen after discharge
    - Ideally within 4 wks.
      - Pass
        - Discharge
      - Fail
        - Schedule Complete Audiologic Eval
Follow-up Truths

- Screening will not accomplish the goal of early identification and early intervention without effective follow-up

- Follow-up is the most difficult part of a screening program
94% of infants in USA are screened for hearing loss
  - CDC, 2007; NCHAM, 2007
54% return for follow-up
  - CDC, 2007
Follow-up is essential for achieving early intervention goals
CDC 2007 Follow-up Statistics

- LTF/LTD, 44.80%
- Hearing Loss, 6.30%
- Normal Hearing, 37%
- In Process, 7.50%
- Died/Refused, 2.70%
- Moved, 1.60%
Points of Loss

Refer from In-Patient Screen

- Out Patient Visit: Rescreen/Diagnosis
- Lost or Late to FU

- Intervention/
  Hearing Aid Fitting
- Lost or Late to FU
Obstacles to Follow-up

• Tracking
  o incorrect phone number, address, name

• Economics
  o fees, transportation, time off from work

• Location
  o distance to follow-up facility

• Scheduling
  o work obligations, older siblings, other appts.
Obstacles to Follow-up

• **Importance**  
  o Parents not convinced of the need  
  o “My baby is fine.”

• **Reassurance**  
  o signs that baby “hears”  
  o pediatrician recommends waiting

• **Anxiety**  
  o fear of procedure, denial, anger
Minimizing Parental Anxiety

• **Timing:** post natal period is an emotional time
• **Personnel:** must be well trained, sensitive, & knowledgeable.
• **Script:** avoid saying *fail*, communicate importance w/o alarming.
Guidelines for the Audiologic Assessment of Children from Birth to 5 years of Age

2004

www.asha.org/members/deskref-journals/deskref/default
“Success of UNHS has resulted in a rapidly growing need for audiologic care for the pediatric population.”

Recognition of increasing number of infants with multiple developmental disabilities who need hearing assessment.

Practice patterns must bridge UNHS program and early intervention programs.
Objectives of Assessment

- Determine status of auditory system
- Identify type, degree, configuration of hearing loss for each ear
- Characterize associated disability
- Assess ability to use auditory information
- Risk factors for late onset hearing loss
Objectives of Assessment

• Assess candidacy for amplification
• Provide counseling
• Communicate findings to other professionals
• Consider the need further evaluation or screening

- Appropriately credentialed audiologists
- Test environment meets ANSI standards for background noise levels
- Equipment calibrated to ANSI specs
  - Daily listening checks
  - Periodic electroacoustic calibration
ASHA Guidelines 2004

Principles

• Evidence based practice
• Cultural competence
• Family-centered service provision
• Documentation
  o For each visit
  o Contain background information
• Informed consent and privacy
Before **Three** Months of Age: 
Audiological Evaluation (Diagnostic Test)

- **Physiological testing is required.**
  - Babies cannot cooperate with traditional hearing test (behavioral) techniques
  - Babies are tested in natural or sedated sleep
  - Three physiological tests:
    - Otoacoustic Emissions (OAE)
      - Tests the sense organ of hearing
    - Auditory Brainstem Response (ABR)
      - Tests transmission of neural impulses from the cochlea through the central auditory pathways in the brainstem
    - Tympanometry
      - Tests the middle ear
Behavioral Testing
0 - 6 months

“It is now known that unconditioned behavioral observation techniques with infants are confounded by poor test-retest reliability, and high inter- and intrasubject variability.”

ASHA 2004 Guidelines
Diagnostic Evaluation: 0 - 6 months

- Diagnostic ABR
  - Click Thresholds (broad spectrum)
  - Tone Thresholds (frequency specific)
Tympanometry
Middle Ear Function
Otoacoustic Emissions
Inner Ear Function

DPOAE
TEOAE
By Six Months of Age
If Hearing Loss is Confirmed

• Continuing medical evaluation including pediatric otologist evaluation and clearance for hearing aid fitting
• Hearing aid fitting by pediatric audiologist
• Parental information and choices concerning amplification and communication options
Diagnostic Evaluation After 6 Months of Age

- By 6 months of age, most babies can be tested using behavioral techniques
- These techniques are based on operant conditioning
- The baby is taught a behavior which is used as the response to sound.
- Behavior is reinforced
  - Behaviors:
    - Turning in direction of sound
    - Serial play activity, i.e. drop a block in a box, put a peg in the board, etc. every time sound is heard
Diagnostic Evaluation: 6 Months - 2 Years

- Visual Reinforcement Audiometry (VRA)
  - Natural reflex to turn in the direction of sound is reinforced and used as the response to sound
Diagnostic Evaluation
2 – 4 years

• Conditioned Play
  Audiometry
  o Child is taught to perform a play task each time she hears a sound
  o Correct responses are reinforced
  o Incorrect responses are corrected
Diagnostic Evaluation after 6 months

- In addition to pure tone audiometry:
  - **Tympanometry**
    - Middle ear test
  - **Otoacoustic Emissions**
    - Test of sensory cells in the inner ear
  - **Speech Audiometry**
    - Test of threshold for speech sounds
    - Test of ability to recognize specific sounds of speech (2 yrs +)
- **If for some reason behavioral audiometry cannot be done:**
  - ABR under sedation may be recommended
Pure Tone Audiogram

- The audiogram is a chart of hearing.
- Threshold for different frequency (pitch) sounds are plotted on the chart.
  - Threshold = the softest sound that a person can hear
- Frequency is measured in Hertz (Hz)
- Intensity is measured in decibels (dB)
Hearing Loss

- Normal hearing for children is defined as thresholds from 0 dB to 15 dB.
- The shaded area indicates the intensity for most speech sounds. As long as hearing is better (above) the shaded area, hearing is adequate for hearing speech.
The intensity or common sounds
Degree of hearing loss is based on the intensity range into which thresholds fall.
Two ways in which threshold is measured

- **Air Conduction**
  - Through earphones
  - Measures hearing through outer, middle and inner ear

- **Bone Conduction**
  - Vibrator is placed behind the ear
  - Sound is transmitted directly to cochlea
    - Outer and middle ear are bypassed
  - Measures hearing through the cochlea
### Audiogram Symbols

<table>
<thead>
<tr>
<th>Condition</th>
<th>Symbols</th>
</tr>
</thead>
<tbody>
<tr>
<td>Air Conduction</td>
<td><img src="image1" alt="Symbol" /> <img src="image2" alt="Symbol" /></td>
</tr>
<tr>
<td>Unmasked</td>
<td><img src="image3" alt="Symbol" /> <img src="image4" alt="Symbol" /></td>
</tr>
<tr>
<td>Unmasked (no response)</td>
<td><img src="image5" alt="Symbol" /> <img src="image6" alt="Symbol" /></td>
</tr>
<tr>
<td>Masked</td>
<td><img src="image7" alt="Symbol" /> <img src="image8" alt="Symbol" /></td>
</tr>
<tr>
<td>Masked (no response)</td>
<td><img src="image9" alt="Symbol" /> <img src="image10" alt="Symbol" /></td>
</tr>
</tbody>
</table>
### Audiogram Symbols, cont.

<table>
<thead>
<tr>
<th>Symbol</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>Bone Conduction</td>
<td>( R ) or ( L )</td>
</tr>
<tr>
<td>Unmasked</td>
<td>( \text{Unmasked} )</td>
</tr>
<tr>
<td>Unmasked (no response)</td>
<td>( \text{Unmasked (no response)} )</td>
</tr>
<tr>
<td>Masked</td>
<td>( \text{Masked} )</td>
</tr>
<tr>
<td>Masked (no response)</td>
<td>( \text{Masked (no response)} )</td>
</tr>
</tbody>
</table>
This is a sensorineural hearing loss. Air and bone conduction thresholds are equal and below normal.
In conductive hearing loss, bone conduction is normal and air conduction shows a loss.
It is possible for a sensorineural and conductive hearing loss to co-exist. Bone conduction is better than air conduction BUT there is a loss of hearing by bone conduction.
AUDITORY HABILITATION

What happens after hearing loss is confirmed?
Early Intervention

- Part C of IDEA is an important resource for infants with hearing loss and their families
  - Audiology services are included in Early Intervention Program regulations
    (10 NYCRR Section 69-4.1(k)(2)(iii)(a-f))
JCIH Proposed 2007

• All families offered Early Intervention

• Point of entry to EI linked to EHDI

• Home based & center based options
Considerations for Early Intervention

• For children with hearing loss and their families...
  o Family Support
    • Counseling
    • Education
  o Technology Decisions
    • Hearing Aids
    • FM Systems
    • Cochlear Implants
  o Communication Choices
    • Oral speech & language
    • Sign Language
    • Total communication
Family-Centered Model

• Focus on involvement of family in therapy
• Empower families with knowledge & skills
• Caregivers taught to optimize daily interactions with child
• Primary goal is to support family
  o Family needs
  o Equal team members
  o Respect family’s decisions
Auditory Habilitation Model

• Family determines communication system to be used by child

• Auditory/oral
  o Emphasis on development of auditory mode
  o Incorporate all modalities
    • Speech reading
    • Auditory

• Auditory Verbal
  o Focus exclusively on auditory
  o No visual cues

• Manual communication
• Total communication
Key Components of Rehabilitative Audiology

- Hearing Aid
- Cochlear Implant
- Auditory Training
Assistive Technology Devices

CI

FM

Hearing Aid
Computerized Fitting Techniques
Auditory Training

A program of intensive training is needed to maximize the child’s residual hearing and facilitate speech and language development.

Child with a Cochlear Implant works with a Speech-Language Pathologist
Other Resources

• National Center for Hearing Assessment and Management (NCHAM)
  o Web site: www.infanthearing.org

• American Speech-Language-Hearing Association (ASHA)
  o Web site: www.asha.org

• New York State Department of Health
  o Web site: http://www.health.state.ny.us
    • Family & Community Health, Infants & Children, Early Intervention