• A Little Audiology 101
• Normal Auditory Development
• Two Paths: Diagnosed & Undiagnosed
• Syndromes, High Risk Factors and Red Flags
The Ear

Video of how the ear works
(http://www.entnet.org/content/how-ear-works)
Where Hearing Loss Happens

- Semicircular Canals (3)
- Stirrup
- Anvil
- Hammer
- Eardrum
- Outer Ear Canal
- Eustachian Tube
- Cochlea
- Nerves
  - connects to the brain
  - connects to the nose

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Where Hearing Loss Happens
Speech Banana

Audiogram of familiar sounds
Frequency in cycles per second (HZ)
Hearing Loss Simulator

Hearing loss can fool you!

- It’s not as simple as, you hear or you don’t; there is much more to it
- Depending on the hearing loss and the child they may respond to the “bell” during the evaluation……
  *But they may not hear speech clearly
  *If they don’t respond to bell refer for hearing test
Brain Development - We Hear with Our Brain
Joint Committee on Infant Hearing
1-3-6 Goals

National Standards for Early Hearing Detection and Intervention

• 1 Month: All newborns have a hearing screening before 1 month of age.

• 3 Months: Diagnostic audiologic evaluation performed by 3 months of age for babies not passing UNHS.

• 6 Months: Early Intervention services start before 6 months of age for babies with permanent hearing loss.
As We Assess and Plan; No Two Children are Alike

- Age of Onset
- Age of Identification
- Age of Amplification
- Fulltime Use
No Two Children are Alike

- Degree
- Nature
- Configuration
- Listening Environment

- Family Interactions
Outcomes

• Research: Outcomes are better for families of babies/toddlers with hearing loss when they have a specialized provider
Auditory Developmental Milestones:

Birth to 3 years Old

(Adapted from Firstyears.org)
Age 0-3 months:

- Auditory detection/attention: reacts to loud sounds with startle (Moro reflex)
- Reacts initially to sounds that are close by; between 2-4 months begins to develop distance hearing
- Responds to LF sounds (vowels) better than to HF sounds (consonants)
- Is awakened by loud voices and sounds
- By the end of the 3rd month, an infant recognizes his mother's voice; stops crying to listen; listens to his/her own sounds
- Enjoys a few noisemakers

Age 4-6 months:

- Turns his eyes/head to search for sounds
- Enjoys hearing his own sounds (gurgling, laughing, and babbling) - auditory feedback loop develops
- Enjoys sound of musical toys (rattles, bells)
- Responds to voices by babbling
- Begins differentiating between environmental and speech sounds
- After hearing his mother's voice, cries if the face he/she then sees is not his mother's face
- Recognizes familiar sounds for feeding (e.g. spoon in a dish)
Age 7-9 months:
• in sitting position, turns eye/head/body to source of sound (sound localization); has difficulty locating sounds above or behind
• responds to simple requests
• modifies speech to match what was heard
• imitates speech and non-speech (blowing raspberries) sounds
• responds to name
• attends to music/singing
• understands many onomatopoeias (Learning to Listen Sounds)

Age 10-12 months:
• responds physically to music
• responds to questions by searching
• can look for named object that is out of sight
• understands some common phrases
Age 12-18 months:
- shows interest in sounds of radio and television
- listens to simple stories, songs, and rhymes
- demonstrates 2 item memory

Age 18-24 months:
- understands when called from another room
- remembers what was heard in the correct order (e.g. "Put the fish in the water and the turtle on the grass.") (auditory sequencing)
- follows a conversation when the topic is known
- answers questions about a picture or book
Age 2-2 ½ years:
• answers questions about a story

Age 2 ½ - 3 years:
• begins making cognitive judgments about what was heard, e.g. "Tell me about your trip to Disney World." (auditory processing)
• answers questions about an undisclosed but familiar topic
Early Intervention Services

Members of the Team:

• Core Team: Developmental Specialists, SLP, OT, PT and Service Coordinator

• Additional Team Members: Hearing and Vision Services
  *PSP
  *SSP
  *Teaming
A child diagnosed with hearing loss is referred to the county:

- Where Do We Begin and Why
- EI Hearing Service; Family Training and Counseling
- Family Outcomes
Making Informed Decisions

• More than 96% of families who have a child with hearing loss have no history of hearing loss.
• Families have little to no knowledge of hearing loss or its implications.
• At their audiology appointment once the family hears that their child has “hearing loss” they don’t hear anything else and miss important information.
• Families continue to report they do not know what questions to ask.
Digging Deeper

- Why do they have the hearing loss? Understanding the hearing loss? Where is the hearing loss? Where is the damage? Understanding their child’s audiogram....hearing levels? Will it be progressive? Do they need aids in both ears? Will (s)he play sports? Can (s)he learn to talk? Can we wait until preschool? We don’t want to lose the aid and will not use at park, daycare, grandma’s...
Two Paths

- Referred to EI with Diagnosed Hearing Loss
- Referred to EI for Other Needs-Hearing Loss Not Diagnosed
Baby born with profound hearing loss, 3 months old; CI Candidate
*Hearing Loss is what makes her eligible
*Is there a need for service?
  *She did not show any developmental delays
  *Will not be able to get cochlear implant until 1 year old
  *RBI showed no concerns with routines; mom reports she is the best, happiest baby
Question

Is there a need for service?
Why or why not?
Baby Born With Profound Hearing Loss, 3 Months Old

- Bring in hearing professionals to help....
  - Dig Deeper
  - Family Assessment
  - Research Low

- Incidence population.....
Baby Born With Mild Hearing Loss

- Baby failed newborn Hearing Screening
- Baby has hearing aids
- Baby is babbling
- Family sees their clinical audiologist
Question

Is there a need for service?
Why or why not?
But They Already Have ...

• How Are We Different?
• Digging Deeper....
Another Path Into EI

Child Referred to EI

With No Diagnosed Hearing Loss
Baby Passed Newborn Hearing Screening

- Late onset
- Progressive
- Hearing loss in one ear
What to Look for in History and Behavior

- Syndromes
- Risk Factors
- Red Flags
Child Referred to EI for Other Reasons

- Case History: No diagnosed hearing loss
- Child diagnosed with Strickler Syndrome
- Speech and language is delayed
- When parents clap their hands, baby claps too
- Pediatrician said not to worry and that his siblings are talking for him
Question

Is there a need for a hearing evaluation or EIHS?
Why or Why Not?
Common Syndromes Associated with Hearing Loss:

Alport syndrome
Branchio-Oto-Renal syndrome
CHARGE syndrome
Crouzon syndrome
Down's syndrome
Goldenhar syndrome
Jervell and Lange Nielsen syndrome
Pendred syndrome
Stickler syndrome
Treacher Collins syndrome
Usher syndrome Type 1 and Type 2
Waardenburg syndrome
Syndromes

- Roughly 50% of childhood hearing loss is genetic. There are over 400 known genetic causes involving hearing loss. The number of genes known to cause hearing loss is constantly changing as researchers identify them.

- Genetic scientists categorize hearing loss into two general types: Non-Syndromic and Syndromic. By far, the more common is non-syndromic hearing loss which includes 2/3 of all genetic hearing losses. (babyhearing.org)
Case history for 2 year old:
* Child born 4 weeks early
* Stayed in NICU for 4 weeks
* Given antibiotics for 3 weeks
* Given oxygen
* Passed newborn hearing screening
* Language is delayed
Question

Is there a need for a hearing evaluation or EIHS?

Why or Why Not?
High Risk Factors for Hearing Loss

- Family history of permanent hearing loss in childhood
- Maternal infections during pregnancy or delivery (Toxoplasmosis, Syphilis, HIV, Hepatitis B, Rubella, CMV, Herpes simplex, and others)
- Physical problems of the head, face, ears, or neck (cleft lip/palate, ear pits/tags, atresia, and others)
- Ototoxic medications given in the neonatal period
- Syndrome associated with hearing loss (Pendred, Usher, Waardenburg, neurofibromatosis)
- Admission to a neonatal intensive care unit greater than 5 days
- Prematurity (< 37 weeks)
- Hyperbilirubinemia

(Infanthearing.org)
Seven Month Old

- Passed UNHS
- No history of hearing loss in family
- By 6 months, doesn't try to imitate sounds
- Hasn't begun to babble to herself or back at others who speak to her
- Doctor said not to worry there is so much going on in the home with so many other children
Question

Is there a need for a hearing evaluation or for EIHS?
Why or Why Not?
Red Flags

Remember:

* Signs of hearing loss can be different for different babies, and the extent of hearing impairment can vary.

* A baby's ability to hear is in large part the foundation of her/his ability to learn. A hearing test is the most important early way to tell.

Delay in communication development
Red Flags

- **Newborn to 3 months**
- Doesn't startle in response to a sudden loud sound
- Doesn't respond to sounds, music, or voices
- Isn't soothed by soft sounds
- Doesn't move or wake up at the sound of voices or nearby noises when sleeping in a quiet room
- By 2 months, doesn't make vowel sounds like "ohh"
- By 2 months, doesn't become quiet at the sound of familiar voices
Red Flags

- **Warning signs: 4 to 8 months**
- Doesn't turn her head or eyes toward a sound she can't see
- Doesn't change expressions at the sound of a voice or a loud noise when she's in a quiet setting
- Doesn't seem to enjoy shaking a rattle, ringing bells, or squeezing noisemakers
- By 6 months, doesn't try to imitate sounds
- Hasn't begun to babble to herself or back at others who speak to her
- Doesn't respond to "no" and changes in tone of voice
- Seems to hear some sounds but not others
- Seems to pay attention to vibrating noises (those that can be felt) but not those that are only heard
Other Reasons for a Hearing Test:

• Parent/caregiver concern regarding hearing
• Delays in speech/language development
• Recurrent middle ear infections or one episode lasting more than 3 months
• Head trauma associated with loss of consciousness or skull fracture
• Bacterial meningitis and other infections (mumps, encephalitis, viral labyrinthitis)
• Exposure to potentially damaging noise levels
• Ototoxic medications received at any time

(infanthearing.org)
Delays in Development

• The nature of hearing and hearing loss; observed behaviors will be inconsistent

• ASHA- Speech & Language Development
When in Doubt......
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