Audiology 101

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Waaaaaaaah!!!

- Baby’s cry (115 dB) is louder than a car horn!!
Rock On!

- Sitting in front of a speaker at a rock concert can begin to damage hearing in 7 ½ minutes.
Polly warns of an airplane

- During WWI parrots were kept on the Eiffel Tower in Paris.
- Because of their keen sense of hearing, they warned people of enemy aircraft long before humans could hear it.
Why is hearing important?

- Development of spoken language is dependent on hearing.
- The basis for cognitive and social development is language.
- Learning in mainstreamed classroom is dependent on hearing and listening.
How much hearing loss is significant?

- **Any** type and/or degree of hearing loss that reduces the intelligibility of speech to a level inadequate for accurate:
  - Perception of sounds
  - Interpretation of sounds
  - Learning
Prevalence of Hearing Loss

- Well Baby: 1-3 per 1000
- NICU: 2-4 per 100
- Additional 2 per 1000 will acquire hearing loss in childhood
- Estimated 1.2 million children have hearing loss
Why screen for hearing loss at birth?

After heart defect, hearing loss is the most common congenital condition.
National Early Hearing Detection and Intervention (EHDI) Guidelines

1-3-6

1 – Screen by **one month** of age

3 – Diagnose by **three months** of age

6 – Fit with amplification and enrolled in early intervention by **six months** of age
Oregon EHDI

- Each state has its own EHDI program
- Oregon EHDI program has one of the strongest reporting and tracking programs in the nation (thanks to its awesome staff!)
Anatomy of The Ear!!
How We Hear (video)

- http://www.youtube.com/watch?v=stiPMLtjYAw
Process of Hearing

- Sound travels in waves
- Pinna “catches sound” and directs it into the ear canal
- Sound waves cause eardrum to vibrate
- Vibration of eardrum makes ossicles move
- Last ossicle “pumps” in and out of the cochlea
- Inside cochlea are “sensors”/hair cells that respond to the pumping action and allow us to hear different sounds
- At bottom of sensor’s are neurons/”fibers” that make the auditory nerve
- Nerve takes sound to brain where it is processed.
Types of Hearing Loss

- **Conductive**
  - Outer or middle ear

- **Sensory**
  - Cochlea

- **Neural**
  - Auditory nerve

- **Mixed**
  - Combination of conductive and sensory/neural
Degrees of Hearing Loss

- Unilateral or bilateral
  - One ear or both ears
- Slight, mild, moderate, moderately severe, severe, profound degree
  - Listed in order of severity from low to high
- Symmetrical or asymmetrical
  - Same or different degree in each ear
Impact of loss to Speech & Language

Example:
- Mild to moderate hearing loss
- Sounds occurring in the gray/shaded area are INAUDIBLE to this child.

- “I’s__ime_ o_o o le’ _ _u_ on our_oa__”
- It’s time to go, so let’s put on our coats.

“o_e your_oe_”

“Go get your shoes”
Hearing Loss Simulations

- Better Hearing Institute
- Phonak
- Starkey
- University of WI
Etiology of Hearing Loss

- Congenital – present at birth
- Acquired – occurred after birth
Hearing Loss

- 50% environmental
  - Congenital rubella syndrome
  - Cytomegalovirus infection
    - Ototoxicity
    - Prematurity
    - Asphyxiation
    - Others

- 30% syndromic
  - Pendred’s
  - Usher’s
  - Waardenburg’s
  - Branchio-oto-renal
  - Others

- 50% genetic
  - 80% recessive
  - 20% dominant
  - 1% X-linked and mitochondrial

- DFNB1
- DFNB4
- Others
Before Newborn Hearing Screening

- Average age of identification 18+ months of age
- Unilateral and mild losses often undetected
  - (My brother)
Oregon Newborn Hearing Screening Legislation

- July 2000 and Jan 2004 (amendment)
  - All Oregon hospitals and birthing centers with 200+ live births per year must provide hearing screen within one month of birth
  - Babies are screened with OAEs and/or AABR
  - Report results to parents
  - Report results to EHDI
  - Refer for additional testing if baby “Refers” or has risk factors
  - Mandatory reporting from diagnostic centers
Newborn Hearing Screening

Otoacoustic Emission (OAE)
Newborn Hearing Screening

Automated Auditory Brainstem Response (AABR)
Why screen for hearing loss at birth?

- **Development!!**
  - Study (Yoshinaga Itano et. Al 1998) looked at receptive and expressive language development in 150 deaf/hh children in same EI program
  
  - With diagnosis/intervention by 6 months – significantly better rec/exp language, social, vocabulary, speech, literacy compared to children diagnosed after 6 months of age
  - No difference intervention occurred at 7-12 months vs. 25 months or later

- Language input within the first 6 months critical
2011 - Oregon

Total Births: 45,363

Screened: 43,779 (96.5%)

- Pass: 42,556 (97.2%)
- Refer: 1,223 (2.8%)

Missed: 113 (7.1%)
Deceased: 45 (2.8%)
Declined: 70 (4.4%)
Unknown: 1,356 (85.6%)

Not Screened: 1,584 (3.5%)

Normal Hearing: 493 (40%)
Lost: 639 (52%)

Pending: 17 (1.4%)
Other: 42 (3.4%)
Hearing Loss: 82 (6.7%)

Enrolled in EI: 59 (72%)
Not Enrolled - Other: 10 (12.2%)
Not Enrolled - Lost: 13 (15.6%)
Loss to Follow Up

- About 50% of babies that “Refer” do not return for testing
- Obstacles:
  - Parents don’t “see” it
  - Primary care provider does not see need
  - Transportation
  - No insurance
  - Distance to diagnostic center
  - Finances
  - Child Care for other children
  - Other
Additional Screening During Childhood

- Medical home (well-child visits)
- Speech/language milestone monitored in medical home, home, education system
- School-based hearing screening

**There may be big gaps in time from newborn hearing screening till next screening**
Monitoring for Children with Risk Factors for Hearing Loss

- Audiologic evaluation by 24-30 months (JCIH, 2007)
  - NICU >5 days
  - Syndrome
  - Family History
  - Craniofacial
  - In utero infections
What about the NICU makes it a risk factor?

- A stay of 5 or more days in the NICU is a risk factor for hearing loss due to conditions that result in babies needing to be admitted to NICU and for treatments they must receive while they are there.
Common conditions resulting in NICU stay 5+ days

- **Prematurity:**
  - While the cochlea is fully developed by 20 weeks gestation, the neural pathways for hearing are still forming until 34 weeks, which is the soonest an AABR should be performed. Sometimes these pathways never reach normal development, resulting in the 'neural' type of hearing loss.

- **Underdeveloped lungs/kidneys/heart:**
  - Oxygenated blood flow is essential to the cochlea, and the structures are very sensitive to toxicity. If the infant isn't able to maintain appropriate air and blood circulation, and is not able to filter out toxic elements, the cochlea can be affected.

- **In utero infections:**
  - Many of these infections have severe implications for the life of the newborn, as well as for the hearing system. Examples: CMV, toxoplasmosis, rubella, herpes, syphilis.
Some treatments that occur in the NICU are linked to hearing loss.

- Often these treatments are viewed as an acceptable risk to/result of hearing loss, because the treatments are to address life-essential functions.

- Extracorporeal membrane oxygenation (ECMO)
- Blood transfusion for hyperbilirubinemia
- Ototoxic medications
  - Chemotherapy
  - High doses of antibiotics
- Loop diuretics (furosemide/Lasix)
Diagnosis of Hearing Loss
Who should be referred for an audiologic evaluation?

- Infants and children who do not pass a hearing screening
- Infants and children at risk for late-onset hearing loss
- Infants and children whose speech, language, or auditory development is a concern
Diagnosing Childhood Hearing Loss Requires

- Appropriate equipment
- Clinician trained and experienced in use of current test methods, procedures and interpretation
- Clinician experienced with testing infants and young children
- Enough time for evaluation and discussion with family
- Test-battery approach
Goals of Audiologic Evaluation

- Rule-out hearing loss
- If hearing loss present, to determine:
  - Degree
  - Type
  - Configuration

**FOR EACH EAR, SEPARATELY**
0 to 6 months

Otoacoustic Emission (OAE)

Auditory Brainstem Response (ABR)
Otoacoustic Emissions (OAE)

- Quick
- Not invasive
- Completed at any age (even adults)
- Infant/child can be awake
- Gives good indication of how cochlea is working
- If normal, vast majority of children have normal hearing
Normal vs. Abnormal OAE

O = Emission   \( \Delta = \) Noise Floor
Auditory Brainstem Response (ABR)

- Test for infants and children who can’t provide reliable information through behavioral testing

- Natural sleep, sedation
ABR

- Electrodes placed on the head
- Insert earphones in ears
- Different sounds are played at different volumes
- Electrodes pick up responses from auditory nerve
- A series of unique waveforms are viewed and compared with normative data (marked by I, III, and V)
Normal vs. Abnormal ABR
Fun Fact

- ABRs completed on animals to make sure they have normal hearing (pure bred)
- Cat ears have 32 muscles in each ear and they can turn each ear independently
6 months to 2.5/3 years

- Visual Reinforcement Audiometry (VRA)
Visual Reinforcement Audiometry (VRA)

- Child sits on lap or in highchair
- Requires good head control
- Sounds presented via speakers or headphones
- Baby turns head when hears a sound
- Toy in the direction of head turn will light up and/or move
  - Responses must be time-locked to stimulus presentation and consistent
  - Ambiguous or delayed responses are not reinforced
VRA (video)

- https://www.youtube.com/watch?v=9Zjtx8GF8Ac
3 to 5 years

- Conditioned Play Audiometry (CPA)
Conditioned Play Audiometry

- Teach the child to listen for a sound and perform an activity in response.
- Child performs the activity each time the sound is heard.
CPA (video)

- https://www.youtube.com/watch?v=1RuELimgLk
Hearing Evaluation: 5 years to adult

- Conventional audiometry
  - Patient raises hand or presses button to indicate they heard the sound
Examination of External Ear

- Microtia/Atresia
- Malformed Pinna
Otoscopy
Common Sightings

Ear infection

Normal
Common Sightings

Normal

P.E. Tube
Common Sightings

Wax

Perforated eardrum
Ears are self-cleaning

- Pores in the ear canal produce cerumen/wax and tiny hairs/cilia, push the wax out of the ear.

- No Q-tips!!
Tympa nometry
Tympanometry

- Probe in ear
- Pressure change ("puff")
- Assesses middle ear function

Can tell us:
- Presence/absence of middle ear fluid
- Typanostomy tubes (PE tubes) working or not
Tympaanometry
Normal vs. Middle Ear Fluid

[Graphs showing tympanometry data for normal and middle ear fluid conditions]
Audiologist’s Reality
Tricks
After Diagnosis

- Referrals to:
  - Early Intervention (Part C)
  - Pediatric Otolaryngologist
  - Other Specialists (Genetics, Ophthalmologist)
  - Hands and Voices (Guide By Your Side)
  - Community Health Nurses
  - Return audiology appointments (testing, hearing aids, cochlear implant team)
Pediatric Otolaryngology

- Provide medical clearance for hearing aids
- Order CT or MRI to identify ear malformations/integrity
- Provide Connexin testing
Early Intervention

- Supported by Part C of IDEA
- Family-centered
- Home-based
- Communication mode choices
Communication Spectrum

Auditory communication

Auditory and visual communication

Visual communication

*All* choices can lead to successful language and communication for children and families
Amplification & Intervention

- **EHDI goal:** Amplification and enrolled in early intervention by **6 months**

- **Reality:** Amplification and intervention by **6 weeks** of age!
  - Hearing aids can be fitted as soon as diagnosed – **we do not need to wait!**
  - Families can start teaching their baby as soon as he/she is born:
    - Foster good infant-parent relationship by encouraging families to love, support, and interact with the new baby
    - Family involvement is the best predictor for a child’s success
Amplification

- Access to sound for ALL children with hearing loss
- Make all of speech spectrum audible
Hearing Aids

- Can be fit at **any** age
- Appropriate for most degrees of hearing loss
- **Behind-the-Ear (BTE)** style
- Typically last 5-7 years, and can easily be re-programmed if hearing changes
- Earmolds replaced frequently for very young (as the ears grow)
- A fraction of the cost of the hearing aid
Making an earmold impression
Amplification

- With a hearing aid, this child has access to the sounds that he/she was unable to detect before.

  - “It’s time to go, so let’s put on our coats.”
  - It’s time to go, so let’s put on our coats.
Bone Conduction Hearing Aid (BAHA)

Designed primarily for those with conductive hearing loss, especially those without an ear or ear canal. Uses vibration to get sound to the normal inner ear.
Cochlear Implant

- Implanted
- Provides access to sound via electric stimulation
Cochlear Implant Candidacy

- FDA - **12 months or older**
- Severe to Profound hearing loss
- Indicated when oral communication is desired and hearing aids do not provide enough benefit for the child to acquire oral/aural language
Needs After Diagnosis

- Ongoing audiologic assessments to:
  - Confirm ABR results
  - Monitor for progression
  - Evaluate benefit of hearing technology
  - Monitor for middle ear disease
Intervention/Educational Options

- State school for the Deaf
  - ASL, Deaf Culture focus
- Regional Programs for the Deaf and Hearing Impaired
  - Public programs, work with county EI and neighborhood schools
  - Auditory/oral, total communication, or ASL focus
- Private auditory/oral or auditory/verbal school
  - Tucker Maxon Oral School
EHDI Improves Outcomes

Moeller et al, 2000  (Boys Town National Research Hospital)
Impact of Hearing Loss

- Depends on:
  - Age of child (when loss occurred and when diagnosed)
  - Severity
  - Configuration
  - Type of loss
  - Age of intervention/amplification, quality of intervention
  - Development, cognition, health
  - Family attributes
Summary

- Early diagnosis, amplification with advanced technology, and intervention leads to improved outcomes

- Education of children with hearing loss has switched from a remedial model to a developmental model......
...we’ve come a long way!
FAQs

What % hearing loss does my baby have?

- We do not categorize hearing loss in percentages. It is misleading because of the variability in amount of hearing loss for each frequency/sound that is tested.
  - “50% hearing loss” could mean...
    - A child has excellent low frequency hearing, but very poor high frequency hearing
    - A child hears medium-loud and loud sounds
    - Not very clear or consistent!

- An audiologist can help parents translate technical terms into language that is easier to understand and will help parents when they are talking with their family and friends.
  - Ex: My child has moderate hearing loss, that means he can’t hear average conversation without his hearing aids.
  - Ex: My child has severe, high-frequency hearing loss. That means that without his hearing aids, he knows you’re talking, but can’t understand what you’re saying. With his hearing aids, he understands better, but will not hear you perfectly.
FAQs:

How do you cure hearing loss?

- Most sensorineural hearing loss is permanent, and can be “treated” with hearing aids or a cochlear implant.

- Scientists are trying to figure out how to regenerate damaged cochlea (and the structures inside), but are still a long, long way from this being possible.

- That said, some conductive hearing loss (ex: microtia/atresia) can be improved (not cured) by surgery.
FAQs:

What are ear infections and why do kids get them all the time?

- Typically, in children, this term refers to middle ear infections, also known as otitis media, and indicates that there is infected fluid (or mucous) behind the eardrum, in the space shared by the middle ear bones.

Children are especially prone to ear infections because the tube that helps keep the space behind the eardrum pressurized and clear is not angled as steep as it is in adults. This means fluid does not drain as easily.
Ear drums

Ear infection

Normal
FAQs: What else is important about ear infections?

- Children with “flat tympanograms” are often said to have “fluid behind their eardrum.” This fluid may or may not be infected, but can block sound from getting to the cochlea, causing temporary hearing loss.

- Some parents misunderstand the phrase “fluid in the ear” and think it is a result of getting water in the ear canal (i.e. bath, swimming)

- Treatments include: watchful waiting, antibiotics, or Pressure Equalization (PE) tubes that are surgically placed into the eardrum.

- Children with sensorineural hearing loss should be watched carefully for ear infections. The fluid in the middle ear will cause an additional hearing loss and puts them at a greater disadvantage if it is not treated.

- Studies have shown that exposure to second-hand smoke increases the likelihood of ear infections in children.
FAQs:

How can I help guide families through the process after not passing a screening? What comes next?

- This process can be confusing, especially with inconsistent information between providers. In an effort to streamline the process for everyone, a Roadmap for Families has been developed: [http://www.infanthearing.org/documents/ParentRoadmap.pdf](http://www.infanthearing.org/documents/ParentRoadmap.pdf)

- Even after the infant/child has been diagnosed with hearing loss, they will continue to have their hearing tested regularly (about every 3 months, initially).
Resources

- **Helpful Websites**
  - Oregon EHDI: [healthoregon.org/ehdi](http://healthoregon.org/ehdi)
  - National Center on Hearing Assessment and Management (NCHAM): [infanthearing.org](http://infanthearing.org)
  - Centers for Disease Control and Prevention (CDC) - Hearing Loss: [cdc.gov/ncbddd/hearingloss/index.html](http://cdc.gov/ncbddd/hearingloss/index.html)

- **Videos**
  - Hearing: [http://www.youtube.com/watch?v=stiPMLtjYAw](http://www.youtube.com/watch?v=stiPMLtjYAw)
  - Hearing tests: [https://www.youtube.com/watch?v=KmlXwHCCQRE](https://www.youtube.com/watch?v=KmlXwHCCQRE)
  - VRA: [https://www.youtube.com/watch?v=9Ztx8GF8Ac](https://www.youtube.com/watch?v=9Ztx8GF8Ac)
  - CPA: [https://www.youtube.com/watch?v=1RuE1LimgLk](https://www.youtube.com/watch?v=1RuE1LimgLk)