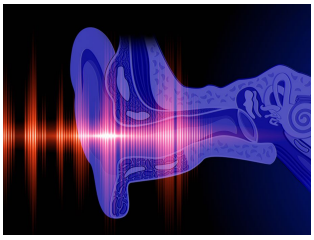

Deafness/Hearing Loss and Cochlear Implants in the Pediatric Population

2019 International Academy of Life Care Planning Symposium

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Objectives

- Review the auditory system and hearing
- Understand how hearing loss impacts communication, learning and development
- Learn about options for hearing intervention and standards of care

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Hearing loss

Definition: Reduction in sensitivity to sound.

- Results from abnormal structure or function within the auditory pathway.

- Deaf: Profound hearing loss, culturally Deaf
- Hard of hearing: mild to severe hearing loss

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Aspects of sound

- Loudness (intensity)
  - Measured in decibels (dB), logarithmic scale
- Pitch (frequency)
  - Ear resolves ~1500 pitches
  - Frequencies of human speech: 250-8000 Hz
- Duration (timing)
  - Length of sound: onset/offset
  - Voice onset time: /ba/ vs /pa/ 20 ms space between /p/ and /a/

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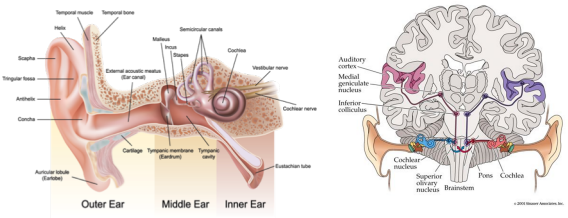
Anatomy: auditory system



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Types of hearing loss

1. Conductive
2. Sensorineural
3. Mixed
4. Neural or central

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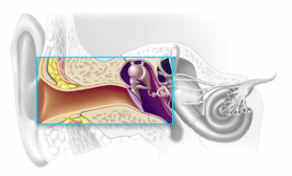
Conductive hearing loss



- Anomaly or pathology in outer ear and/or middle ear
  - Eardrum perforation, middle ear fluid, ear infection, absent ear canal

Results in
- loss of audibility (loudness)

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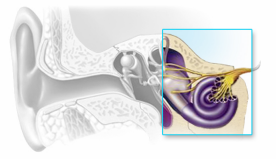
Sensory hearing loss



- Anomaly or pathology in the inner ear (usually cochlear hair cells)

Results in:
- loss of audibility
- loss of frequency resolution

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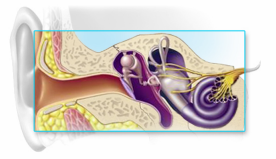
Mixed hearing loss



- Anomaly/pathology in the conductive mechanism and the inner ear

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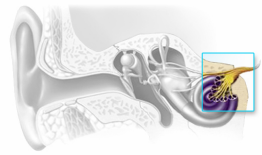
Neural/central hearing loss



- Anomaly or pathology of the auditory nerve, auditory brainstem pathways, or auditory cortex.

Results in:

- loss of audibility,
- loss frequency resolution
- loss of timing cues – poor speech recognition

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Audiometry

Measures hearing thresholds for each ear at sound frequencies within the speech range

Threshold: Softest level of sound that can be perceived

Information provided:

- Type of hearing loss (conductive, sensory, mixed, neural)
- Degree/severity of hearing loss
- Audibility of the speech spectrum

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Audiometry

- Standard testing: Button press, hand raise (6 years+)
- Conditioned play audiometry: Physical response with toys (2-5 years)
- Visual reinforcement audiometry: Conditioned head-turn to a visual reinforcer: animated toy, video (beginning at 6 months-2 years)

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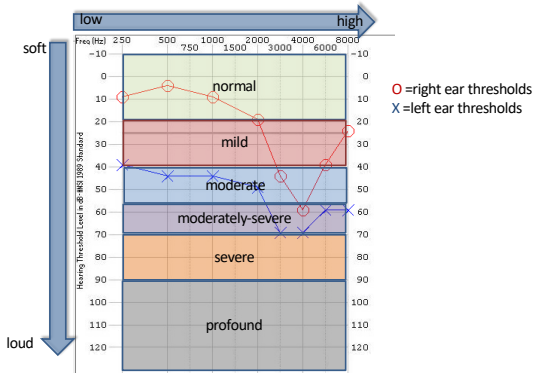
Audiogram



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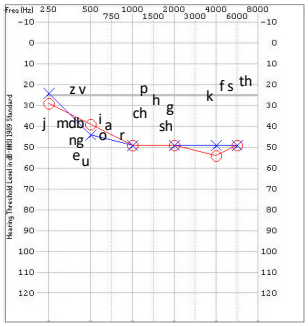
Predicting audibility for speech



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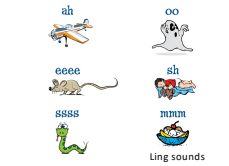
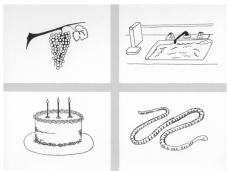
Other behavioral assessments

- Speech audiometry
  - Identification of speech sounds (m, a, i, u, s, sh)
  - Word and sentence recognition
- Obtained for
  - average conversational loudness
  - soft speech
  - speech in noise
  - with/without hearing technology





Northwestern University Children's Perception of Speech Test (NU-Chips)

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Objective testing: Auditory brainstem response (ABR or BAER)

Measurement of electrical responses produced by the auditory nerves and auditory neural pathways in response to sound.

Used to determine hearing thresholds for:

- Infants and toddlers
- Those who can't reliably provide behavioral responses

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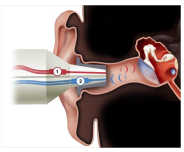
Other objective measures

- Tympanometry: middle ear function
- Otoacoustic emissions: function of the cochlear sensory cells



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Newborn hearing screening

- Objective, automated interpretation
  - OAEs (cochlear function)
  - AABR (auditory nerve and brainstem pathways)

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Before universal newborn hearing screening

- Average age at diagnosis of hearing loss:
  - 12-24 months for severe-profound hearing loss
  - 3 years + for mild-moderate, high frequency or unilateral

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Prevalence of childhood hearing loss

Congenital hearing loss: 1/1000 births

Congenital deafness: 4-11/10,000

Children 6-19 years: 3/1000

Korver et al., Nat Rev Dis Primers. 2017; 3: 16094. doi:10.1038/nrdp.2016.94.

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Etiology of congenital hearing loss

- Genetic: 50-60%
  - 70% autosomal recessive
  - 15% autosomal dominant
  - 15% other (x-linked, mitochondrial)
- Non-genetic: 25%
  - Maternal infection
  - Perinatal complications
- Unknown: 25%

Centers for Disease Control Annual Data http://www.cdc.gov/ncbddd/hearingloss/data.html

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Etiology of congenital hearing loss

- Non-genetic: 25%
  Maternal infection (TORCH)
  Toxoplasmosis
  Other
  Rubella
  Cytomegalovirus (30-40% non-genetic congenital hearing loss)
  Herpes simplex virus

- Perinatal complications
  NICU: Hearing loss prevalence 20-50 times that of babies in well-baby nursery

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Syndromic hearing loss

- 25-30% of genetic hearing loss is associated with a syndrome
- Over 400 syndromes with hearing loss as a feature
- Can be congenital or delayed onset/progressive

Usher syndrome
CHARGE syndrome
Branchial-oto-renal syndrome
Pendred syndrome
Waardenburg syndrome
Jervell and Lange-Nielsen syndrome
Stickler syndrome
Treacher-Collins syndrome
Down syndrome

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Hearing loss and other conditions (%)

Hearing loss with no other disability	60%
Learning disability	7.2
Intellectual disability	8.8
Attention deficit disorder (ADD/ADHD)	5.4
Autism	2.2
Blindness or vision impairment	4.1
Emotional disturbance	2.1
TBI	0.4
Cerebral palsy	3.4

Gallaudet Research Institute, August 2013

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Children identified with hearing loss after passing NBHS

923 children diagnosed with hearing loss 2001-2011

78 (11.8%) passed NBHS

Etiology

- Unknown: 54%
- Genetic 17%
- Anatomic abnormality 14%
- Acquired perinatal 12%

Dedhia et al., *Otolaryngol Head Neck Surg* 2013, Jan 17:1-5

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Acquired childhood sensorineural hearing loss

- Infection (meningitis)
- Trauma (temporal bone fracture)
- Noise-induced
- Ototoxicity (cisplatin, aminoglycoside antibiotics)
- Neurodegenerative disease
- Idiopathic sudden hearing loss

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Untreated childhood hearing loss

1. Delays in development of auditory skills, receptive and expressive communication skills

2. Language deficit may cause learning problems and reduced academic achievement

3. Communication difficulties can lead to social isolation and poor self-concept

4. May impact educational opportunities and vocational choices, and lifetime earnings

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Acoustic access

- Brain needs access to sound for auditory brain development
- Auditory cortex is directly involved in speech perception and language processing
- Central auditory pathways develop quickly in infants and is a precondition for preverbal and verbal language development
- Delayed diagnosis: auditory deprivation
  - Auditory centers reorganize to receive input from other senses

Kretzmer et al, Arch Otolaryngol Head Neck Surg 2004;130(5):499-508
Sharma, et al, Int J Audiol 2007*46*(9), 494–499.

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Hearing, speech perception and language

- Hearing speech leads to spoken language, reading and learning
- Infants become specific language listeners between 6-12 months
  - Brain develops phonetic categories: recognizing phonetic differences that have meaning
  - Children use phonetic categories to learn new words
- Children must repeatedly hear the details of speech sounds in order to understand the subtle aspects of language

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Unilateral hearing loss

- Hearing loss in one ear, normal hearing in the other ear
  - At-risk for speech and language delays
  - 10 times the risk for academic difficulties
  - Teachers more likely to describe as having attention and behavior problems
  - Hearing technology is now recommended

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Standards of care for children with hearing loss

- Audiologic evaluations at least every 6 months
  - Monitor hearing loss for progression
  - Monitor for middle ear fluid/infections
  - Check hearing technology, adjust as child grows or if hearing changes, replace hearing devices every 3-5 years

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Intervention options for children with hearing loss

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- No current methods to correct abnormal structure or function of the inner ear (or extensive anomalies of the outer and middle ear)
- Management options include
  - Hearing technology
  - ASL/Signed English/Visual language
  - Aural rehabilitation
  - Early intervention
  - Speech and language intervention
  - Educational support services

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ASL and signed language

92% of infants with hearing loss are born to parents who are hearing and communicate with spoken language

Deaf children born to Deaf families have access to visual language from birth

- Bimodal bilingualism for hearing families:
  - Entire family needs to become fluent in ASL or signed English and sign all of the time
  - Provide a complete language model
  - Demands of fluency may be difficult for many

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Hearing technology

- Bone anchored hearing system (baha)
- Hearing aids
- Cochlear implants
- FM system/remote microphone technology

- Goal: to make the entire speech spectrum audible
  - Average speech
  - Quiet speech
  - Distant speech
- Get sound to the brain to develop the auditory system

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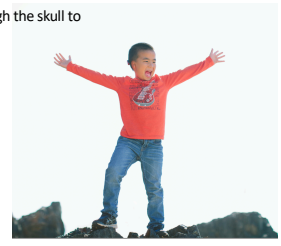
Bone anchored hearing device (baha)

Used for:
Conductive and mixed hearing loss, microtia, unilateral severe-profound sensorineural hearing loss

Device oscillates to send sound waves through the skull to reach the inner ear
Softband or surgical (age 5+)



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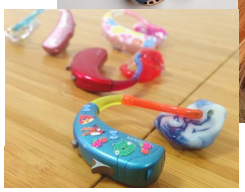
Hearing aids

Digital signal processing

Programmed specifically for child's hearing loss and ear canal acoustics

Prescriptive targets for each frequency band



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Cochlear Implants

For severe to profound sensorineural hearing loss hearing aids can't provide enough audibility.

A cochlear implant (CI) converts sound into electrical impulses which are transmitted to electrode array implanted in the cochlea which directly stimulates the auditory nerve.

Does not sound like "normal hearing" -- 24 channel electrode doing the job of thousands of inner ear sensory cells.


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FM system/remote microphone

- Improves the ability to hear and understand speech in noise and distant speech
- Wireless microphone worn by teacher or person speaking and the signal is transmitted to hearing aid, cochlear implant, or a receiver.
- Often used at school but can be used in any setting (work, community).

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What is needed?

- Early identification of hearing loss
- Immediate auditory access through the use of technology
- Full-time use of hearing technology
- Language-rich environment with daily listening experience and practice
- Motivated family
- Close audiologic management
- Appropriate early intervention and school support

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Costs: hearing technology

- Average cost a mid-level pair of hearing aids is $4,800 (not including batteries, earmolds and repairs)
- Expected life span: 5 years
- Most private U.S. health insurance plans do not cover hearing aids
  - 21 states require that private health insurance provide some coverage for children's hearing aids[2]
  - Average benefit: $3025.00
  - Only 4 states mandate any hearing aid benefits for adults[2]

[1]Cropp I. http://www.aarp.org/health/conditions-treatments/info-05-2011/hearing-aids-cost.html
[2]American Speech-Language Hearing Association: http://www.asha.org/advocacy/state/issues/ha_reimbursement/

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Costs: hearing technology

- Cochlear implant: $30,000-50,000
  - Surgery, device, audiologic services
  - Most major insurance companies, Medicaid, Medicare include cochlear implant benefits
  - Medicaid required to cover cochlear implant costs for children under 21
- Baha: $5000
  - Processor + audiologic services

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Costs: healthcare

Healthcare costs
- Audiologic services to monitor hearing loss and maintain/manage hearing technology
- Speech-language therapy services

Mean annual health-care cost of hearing loss: $9412* per child

*international dollars

Global costs of unaddressed hearing loss and cost-effectiveness of interventions: a WHO report, 2017. Geneva: World Health Organization; 2017. License: CC BY-NC-SA 3.0 IGO.

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Costs: education

Educational costs
- Specialized education and support services
- Hearing services, speech-language services
- Classroom hearing technology (FM systems, loop systems)

- Annual average additional educational cost per child with hearing loss: $5,075-11,909*[1] (Australia and UK, year 2015)
- Lifetime educational cost per child with moderate or greater hearing loss without other disabilities $115,600[2] (U.S., year 2007)

*international dollars

[1]Global costs of unaddressed hearing loss and cost-effectiveness of interventions: a WHO report, 2017. Geneva: World Health Organization; 2017. Licence: CC BY-NC-SA 3.0 IGO.
[2]Grosse SD. *Volta Voices* 2007; 14(6):38-40.

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Costs: productivity

- Difference in annual earnings for U.S. adults with hearing loss versus those without hearing loss: average $12,000 less[1]
- After controlling for education, age, sex, and race, individuals with hearing loss had 1.58 times higher odds of low income (95% CI: 1.16-2.15) and 1.98 times higher odds of being unemployed or underemployed (95% CI: 1.38-2.85) compared to individuals without hearing loss[1]
- Employment rate for adults with hearing loss 13% lower compared to general population

[1]Emmett & Francis, *Otol Neurotol*,2015; 36(3):545-550
[2]McNeil, *Employment, earnings and disability*, US Bureau of the Census, 2000.

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Thank you and questions

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