

Hearing Loss in Children



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Disclosures

- Med El corporation (implant company)
 - Pediatric Advisory Board (consultant)
- Much of current standard clinical practice in pediatric cochlear implantation is “off-label”



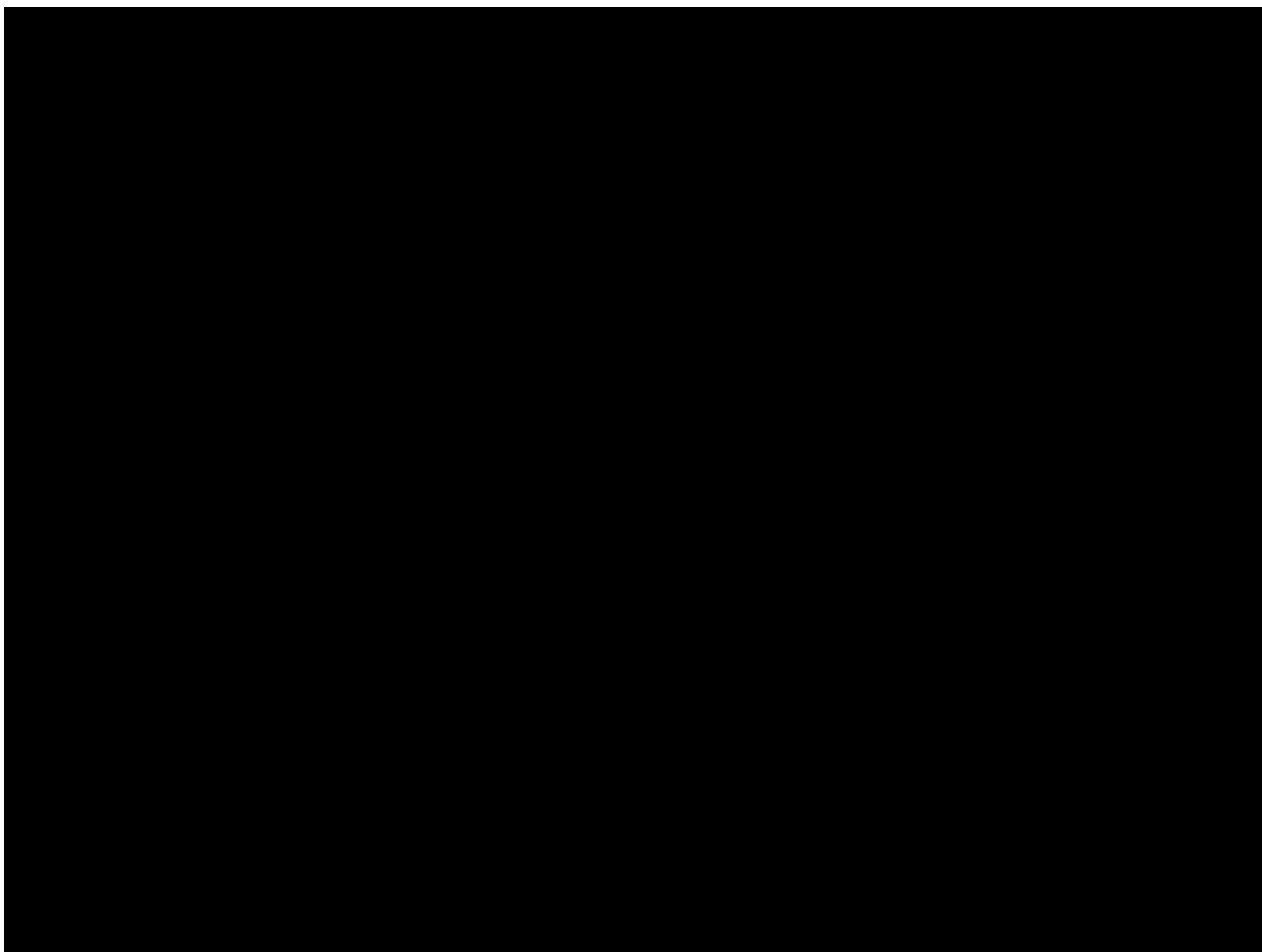
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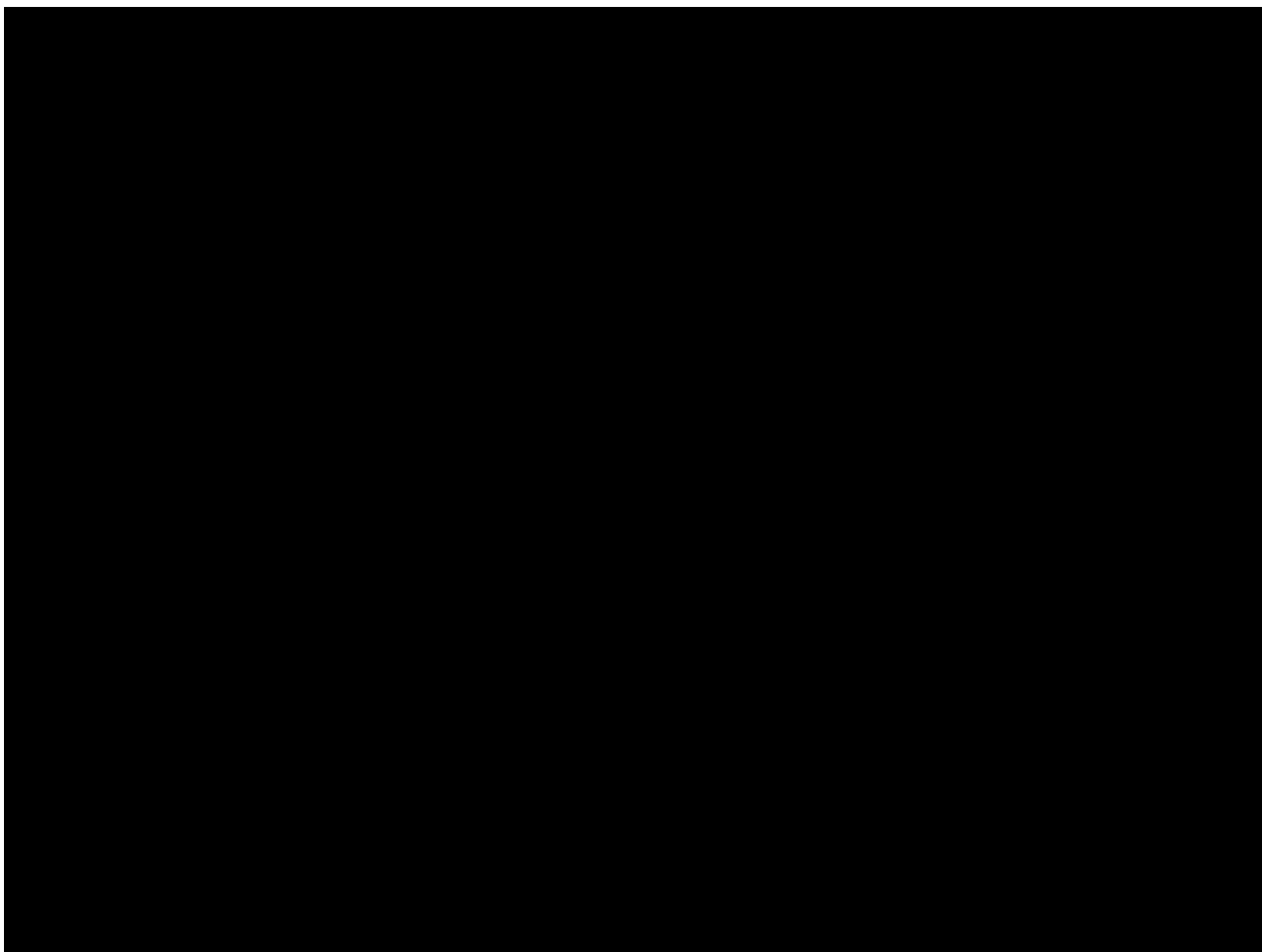


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YouTube

<https://youtu.be/BJez6xxGeMs>







Why Hearing Loss Matters – *Especially* in Children



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Hearing Loss and Language

Moeller et al. (2007)

- Children with (even) mild to severe hearing loss have difficulties with:
 - Semantics
 - Vocabulary
 - Novel word learning
 - Conceptual categories
 - Morphologic development
 - Syntactic development



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Impact of Deafness (historical)

- Unhabilitated HOH/deaf children
 - attain median reading scores at 4th grade level by 18 yrs
 - Of those accepted into college, 70% withdrew before earning degree (1980s data)
 - Less than 1/3 find a job
 - Less wages earned compared to national avg

Literacy in Deaf Children...

- Reading comprehension in **D**eaf teens 15-17 yrs old is low (1st-4th grade level)
 - Holt (1993), Traxler (2000), Wauters (**2006**), Qi (**2011**)
- Louisiana 8th grade reading proficiency:
 - 23.8% LSD vs. 58.9% state-wide
- 35%-65% of children (D/HH) did not meet proficiency for the reading requirements in their state assessment, Grades 3–8
 - Easterbrooks (**2012**)



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Why? Possibly...

- Phonemic awareness is low, leading to poor word identification (decoding)
 - Mixed studies (some low, some equal to NH)
- Poorer understanding of grammatical rules
 - Quigley et al (1977)
- Lower vocabulary levels
 - Kelly (1996), Marschark et al (2002), Paul (2003)
- Lack of access to language may cause difficulties with the learning of word meanings acquired via linguistic explanation (rather than perceptual)
 - Wauters et al (2003)



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Reading and Spoken Language

Perfetti and Sendak (2000)

- Writing systems evolved to encode spoken language
- Hearing children successful in reading learn to do so by connecting spoken language to print
- Hearing adults demonstrate implicit speech components are activated by reading words
- **Speech processes are foundational in skilled reading**
- **Phonologic awareness important**

Hearing Loss and School Behavior

- Has difficulty attending to spoken or other auditory information
- Frequently requests repetition
- Fatigues easily when listening
- Gives inappropriate answers to simple questions
- Appears isolated from peers
- Has difficulty with reading skills
- Has difficulty with spoken and/or written language
- Is easily frustrated

Johnson & Seaton, 2011

Beyond “Speech Delay”

- 30-40% of children have at least 1 additional disability
 - Global Developmental Delay
 - Specific Learning Disabilities
 - Intellectual Disability
 - Autism
 - Attention and executive dysfunction
 - Adjustment problems

Chilosì et al. (2010, Gallaudet Research Institute (2008), Wiley, Arjmand, Meinzen-Derr, Dixon (2011)



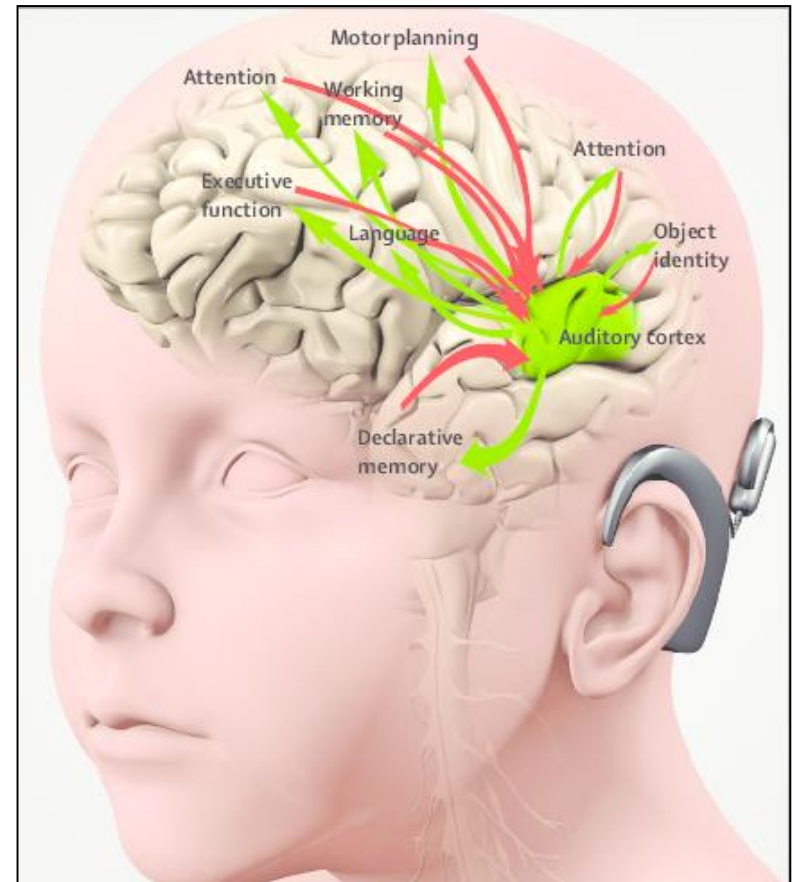
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Neurocognitive Impact

- Executive Functioning
 - Working memory
 - Controlled attention
 - Self-monitoring
 - Organization
 - Inhibition
 - Flexibility
 - Goal Direction



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Unilateral Hearing Loss

- In the past... "One good ear is all you need"
- More accurately understood now:
 - Language and intelligence likely same as normal hearing children, **yet**
 - Higher incidence of behavioral problems
 - 37% more likely to repeat a grade



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The Good News

- These negative outcomes can largely be prevented or mitigated...



<https://www.google.com/url?sa=i&rct=j&q=&esrc=s&source=images&cd=&cad=rja&uact=8&ved=0ahUKEwiDzIzN2Y7VAhUn34MKHc0HDZ4QjRwIBw&url=http%3A%2F%2Fwww.cikguhailmi.com%2F2014%2F10%2F&psig=AFQjCNE9l7iCzDjOu7-wVFFaOPosLPmsXw&ust=1500325303820538>

Baby Boy XG

- Born FT
 - Good prenatal care, no perinatal events, etc
 - Parents in your office for first well baby visit
 - Referred UNHS on Left ear and Right ear
-
- What next?
 - What do you tell family?



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A close-up photograph of a man with a beard and a blue shirt holding a baby. The baby is wearing a blue and green plaid shirt and a green hearing aid in their right ear. The man is looking down at the baby with a gentle expression. The background is blurred.

Early Identification



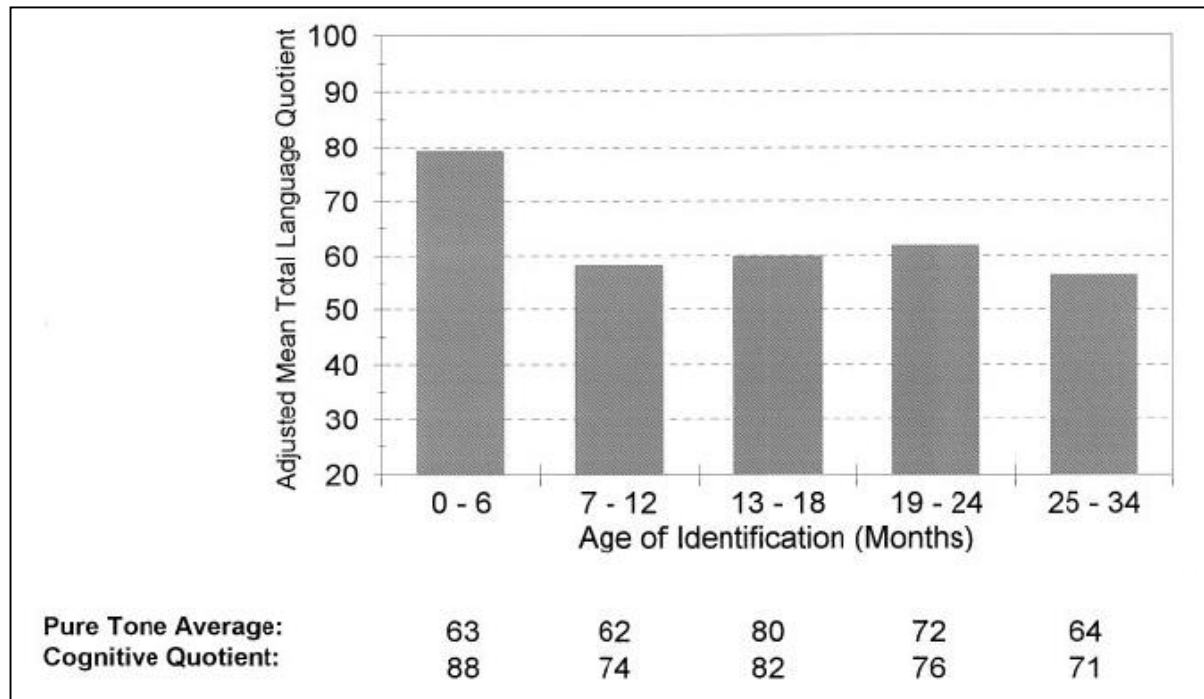
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Timing of Identification

- Critical period of identification (by 6 months) and quality intervention services



Yoshinaga-Itano (1995, 1998, 2000, 2004)

Early Hearing Detection and Intervention (EHDI) - Components

- Universal Newborn Hearing Screening
- Confirmation of Hearing Loss in Infants Referred from UNHS
- Early Intervention
- Continued Surveillance, Screening, and Referral of Infants and Toddlers

Joint Commission on Infant Hearing (2007) - Principles

- Lays timeline for screening/intervention: “1-3-6” rule
- EHDI should be family centered
- Child/family should have access to high level technology (hearing aids, cochlear implants)
- Continued assessment and monitoring in the medical home
- Interdisciplinary intervention by knowledgeable professionals
- Measured outcomes and reported effectiveness



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“1-3-6 Rule”

- By 1 month (preferably before discharge)
 - Universal Newborn Hearing Screen
 - “Refer” - Re-screen with confirmatory AABR
- By 3 months
 - Confirmatory audiologic and medical diagnostic assessment
- By 6 months
 - Begin early intervention/treatment

Joint Committee on Infant Hearing (2007)



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If refers Universal Newborn Hearing Screen...



Next Step: Diagnostic ABR. Period.



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Screen vs. Diagnostic

Screen: Pass/Refer (no information)

ABR:

Corrected Response Thresholds (dB HL)
for Assumed Behavioral Thresholds in dB HL (Stapells, 2000)

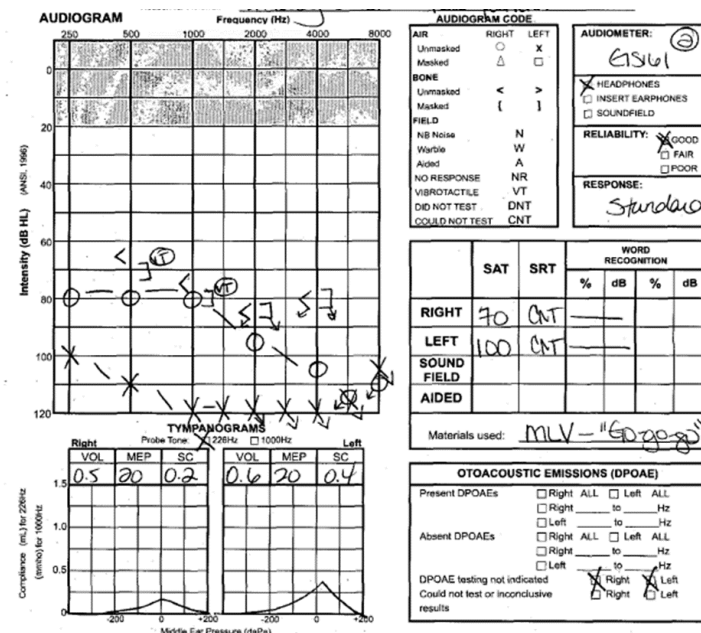
	Air Conduction				Masked Bone Conduction 500 Hz	Masked Bone Conduction 2000 Hz	Masked Bone Conduction 4000 Hz
	500 Hz	1000 Hz	2000 Hz	4000 Hz			
Right Ear	50	DNT	50	50	20	50	50
Left Ear	20	20	20	20	-	-	-

Today's results are consistent with:

Right Ear: Moderate conductive hearing loss for the 500 Hz region and a sensorineural hearing loss for the 2000-4000 Hz regions.

Left Ear: Normal hearing for the 500-4000 Hz region.

Behavioral:



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Outcomes after UNHS and EHDI

- Yoshinaga-Itano (2003)
 - UNHS results in **earlier identification** of hearing loss
 - UNHS results in **earlier intervention**
 - Early identified children more likely to have **normal language** development
- Harrison et al (2003)
 - the average age at which hearing loss is confirmed has dropped **from 24-30 months to 2 to 3 months**
- Moeller et al (2000)
 - Early enrollment in intervention and high family involvement resulted in **higher language scores** by age 5



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Implementation of UNHS

- Ohio (*N=138,704 births in 2016*)
 - 97.69% newborns screened
 - At least 2.31% not screened
 - 71.7% of the “Not Pass” get diagnostic testing
 - At least 23.36% Lost to F/U of Diagnosis
 - ~63.9% of those with HL receive Early Intervention
 - At least 19.8% Lost to F/U for Intervention

N=202 total cases of identified hearing loss



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CDC Hearing Screening and Follow Up Survey, 2018



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Don't Drop the Ball

DIAGNOSIS SUMMARY ^d	2012	2013	2014	2015	2016
Percent non-pass	2.90%	2.80%	2.90%	2.70%	2.78%
Percent non-pass with normal hearing	59.20%	65.60%	65.80%	60.30%	64.87%
Percent non-pass with hearing loss	5.40%	5.80%	5.10%	5.50%	5.24%
Percent with no diagnosis	35.40%	28.60%	29.20%	34.30%	29.89%

CDC Hearing Screening and Follow Up Survey, 2018

- Yes, only 5%, **BUT...**
- Don't let OM and “fluid” distract
- It **IS** worth sedation to know early
- Delays and gaps in the system



Baby XG

- Family did **NOT** get a diagnostic ABR
 - “probably fluid” by PCP
 - Family didn’t see importance
 - Speech and language delay, seen by SLP
 - Diagnosed with Autism Spectrum Disorder
 - Diagnostic hearing testing done at age 4 shows bilateral profound SNHL
 - CI done...***but very late...***
-



Congenital Hearing Loss



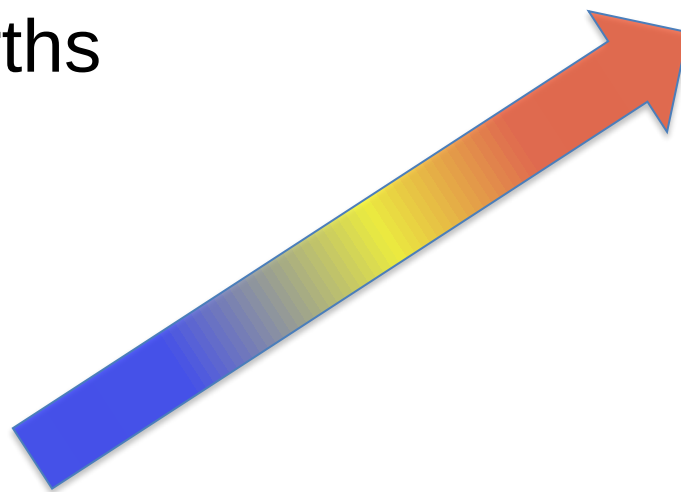
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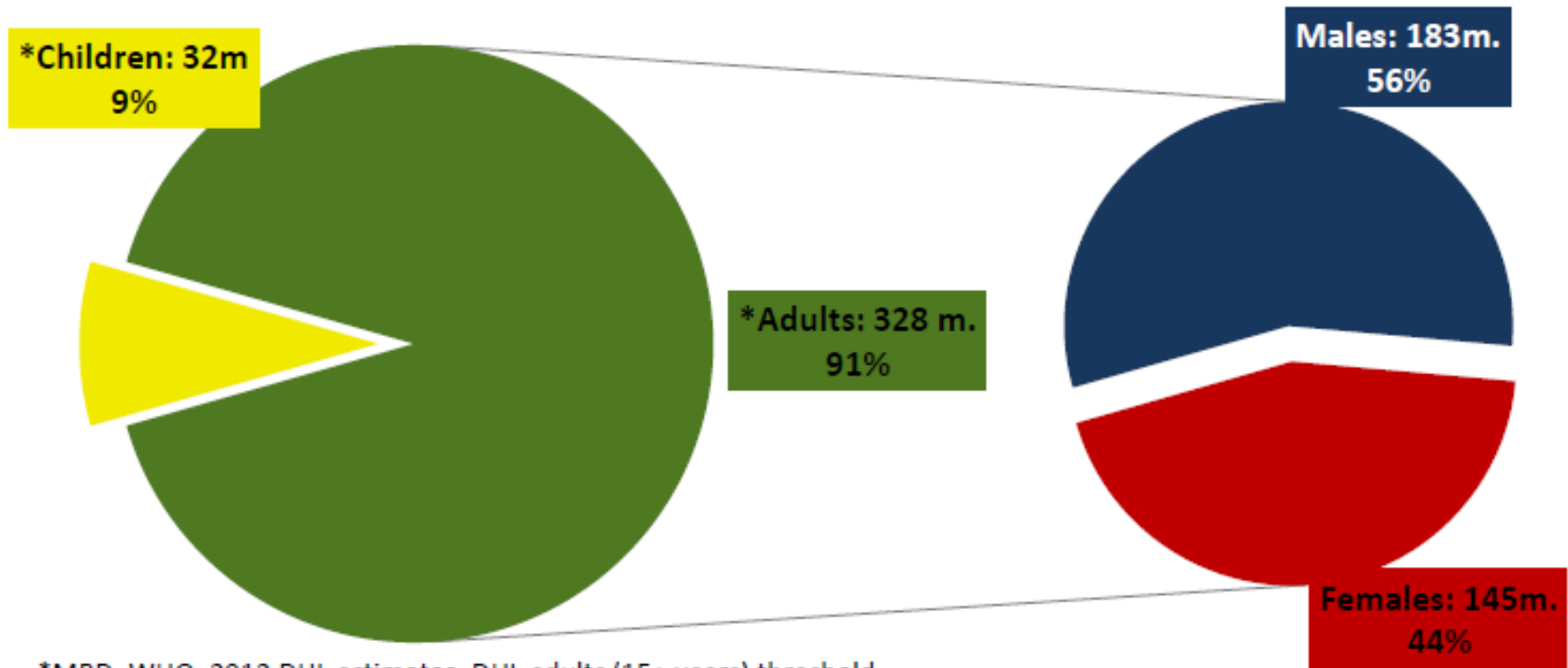
Permanent Childhood Hearing Loss Prevalence

- Congenital
 - 1.86 per 1000 live births
- By age 5
 - 2.7 per 1000
- By adolescence
 - 3.5 per 1000



Morton and Nance, NEJM, 2010

World Health Organization, 2012: Global estimates



*MBD, WHO, 2012 DHL estimates; DHL adults (15+ years) threshold is ≥ 41 dB and children threshold is ≥ 31 dB.



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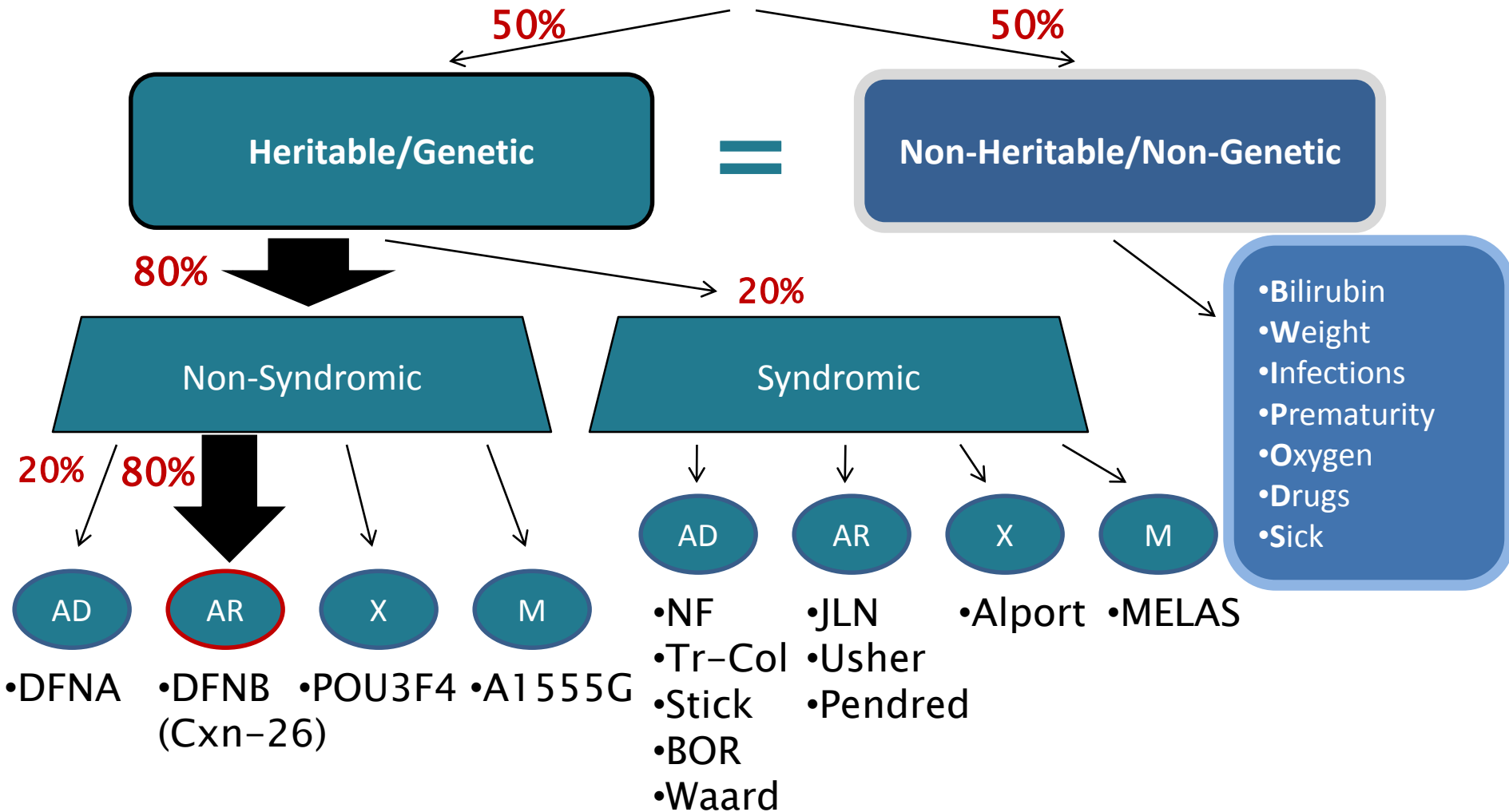
http://www.who.int/pbd/deafness/WHO_GE_HL.pdf



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Congenital Hearing Loss

2-3:1000 births > 50 dB



Congenital Hearing Loss

1:1000 births > 50 dB

50%

Non-Heritable/Non-Genetic

- Bilirubin
- Weight
- Infections
- Prematurity
- Oxygen
- Drugs
- Sick

Mnemonic:

Babies Want IPODS



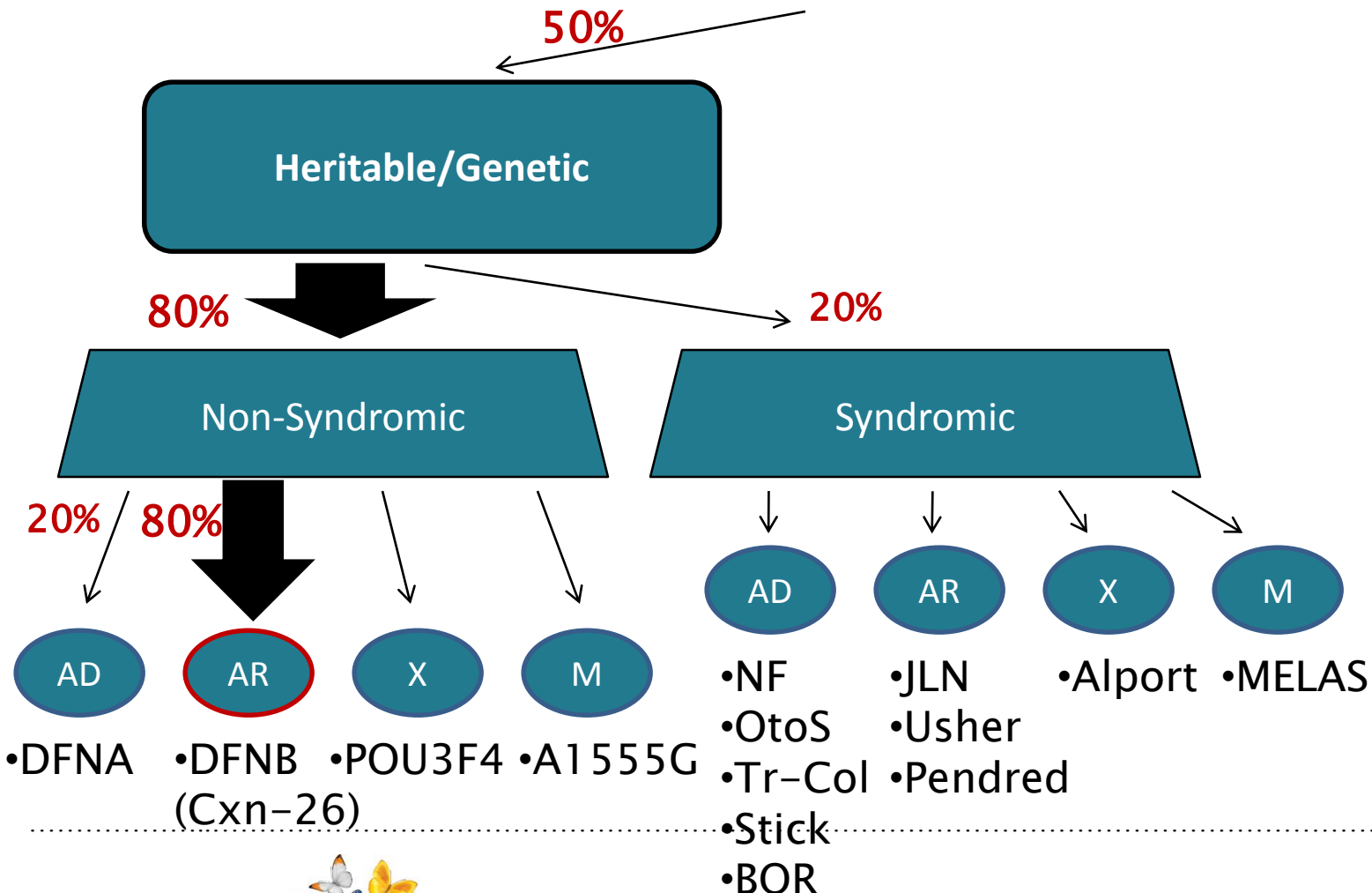
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Congenital Hearing Loss

1:1000 births > 50 dB



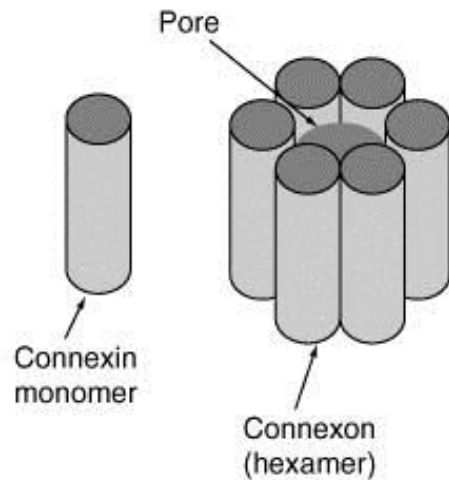
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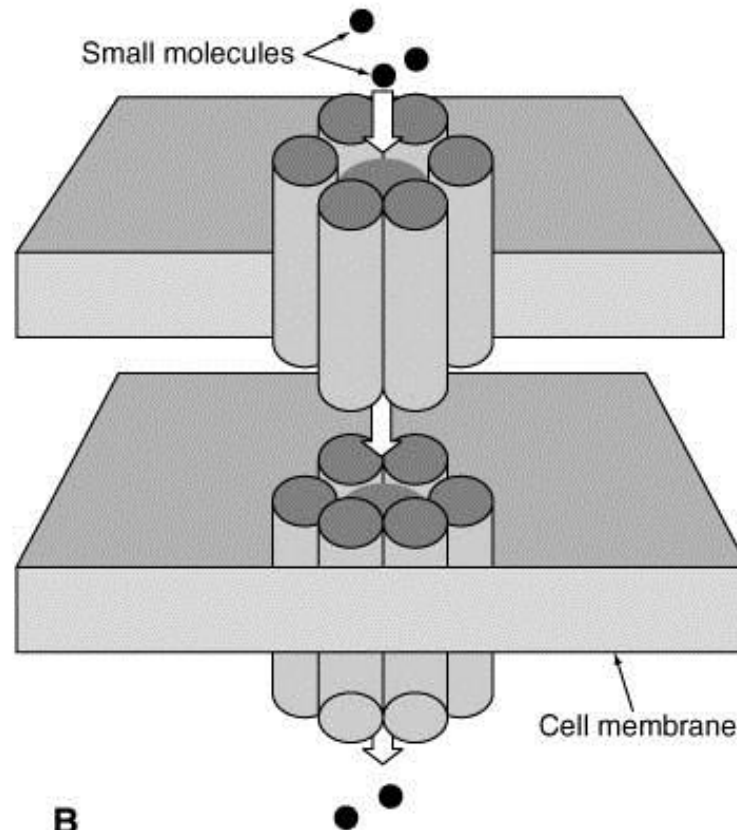
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Connexin-26

- Gap Junction protein (6 connexins make up a connexon)
- Are in supporting cells of the hair cells and in the stria vascularis **potassium** ionic exchangers.



A



B

Workup of Congenital SNHL

- History...
- Physical Exam...
- Stepwise Diagnostic Evaluation...
- **Audiology, Ophthalmology, Speech, EIS for all**



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Workup of Congenital SNHL

- Labs
 - Routine lab battery is **NOT** recommended
 - History and Exam based testing (UA, renal ultrasound, etc)
 - CMV IgG
 - ECG if bilateral severe to profound
 - Unilateral SNHL
 - **Imaging first**
 - Connexin testing if negative
 - Bilateral SNHL
 - **Connexin testing first**
 - Imaging if negative
 - Genetics if dysmorphic/AD pattern, or if above workup negative
-



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Ophthalmology

- Sharma et al. (2009)
 - N=226 children with SNHL
 - 22% had abnormal ophthalmologic findings
 - (~2.5-14% in general population)
 - Refractive and non-refractive errors (only ~2% syndromic)
 - Impact of 2nd sensory loss?



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Hearing Loss Gene Panels

- Differences in cost, genes covered, and turnaround time
- May or may not be covered by insurance

Table 2. Comprehensive genetic test panels, commercially available in United States

Test	Laboratory	Method	CNV	NSHL	SHL	Time	Cost
Otoscope	University of Iowa Molecular Otolaryngology and Renal Research Laboratory	NGS (TGE + MPS)	Yes	90	28	12 weeks	\$1500
OtoSeq	Cincinnati Children's Molecular Genetics Laboratory	NGS (TGE + MPS)	No	13	10	12 weeks	\$3602
OtoGenome	Laboratory for Molecular Medicine, Partners HealthCare Personalized Medicine	NGS	Yes	66	21	8–12 weeks	\$3800
OtoGenetics Deafness Test	OtoGenetics Corporation	NGS	No	74	55	5–6 weeks	\$596
Emory Hearing Loss Expanded Panel	Emory Genetics Laboratory	NGS	No	61	31	12 weeks	\$3200

CNV, copy number variations; MPS, massively parallel sequencing; NGS, next generation sequencing; NSHL, nonsyndromic hearing loss; SHL, nonsyndromic hearing loss; TGE, targeted genomic enrichment.



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Review of Basics



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Classifying Hearing Loss

- **Type:**
 - Conductive Hearing Loss
 - Sensorineural Hearing Loss
 - Mixed Hearing Loss
- **Unilateral / Bilateral**
- **Severity**
 - Mild, Moderate, Severe, Profound

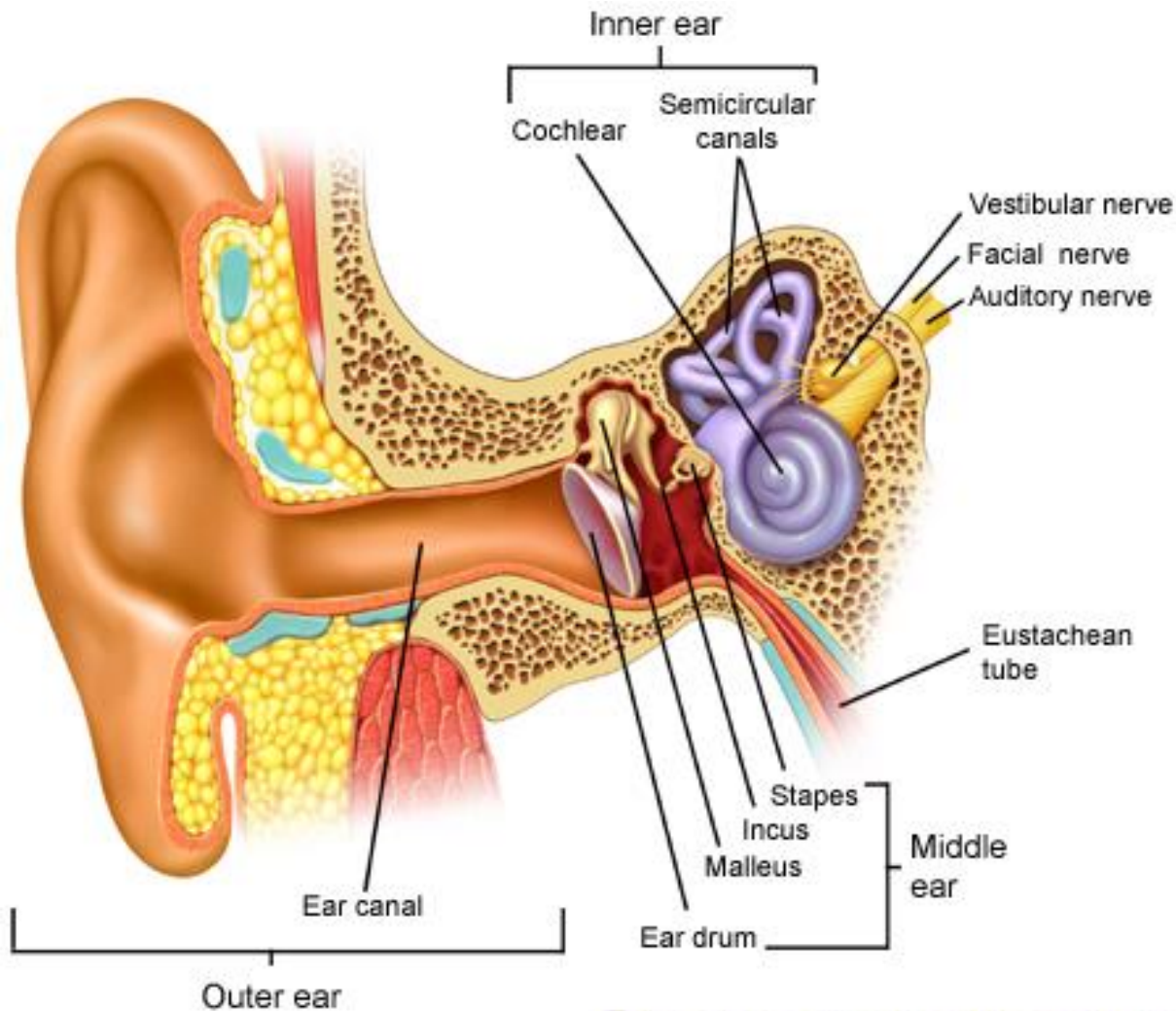


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The “Ear”



The Tympanic Membrane

- Annulus
- Malleus
- Pars Tensa
- Pars Flaccida
- Attic/Epitympanum
- Mesotympanum
- Incus



TM – Pulling back the curtain

- Imagine what is **BEHIND** the eardrum (middle ear)
- “attic” area
- Anterosuperior vs. posterosuperior quadrant



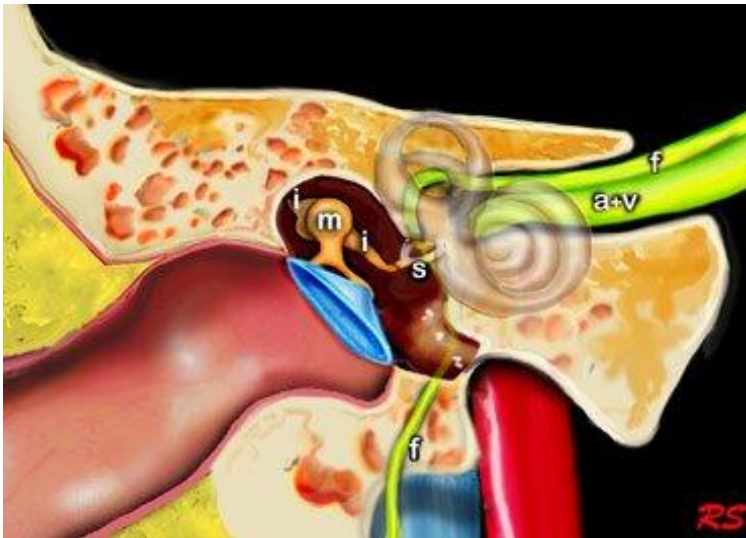
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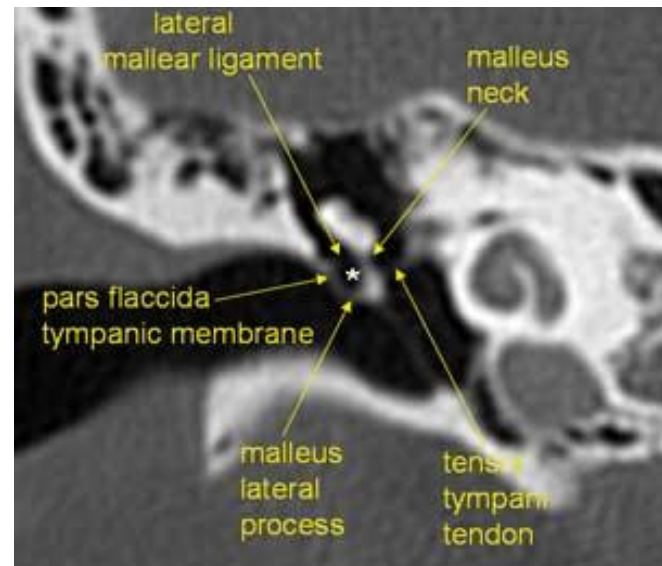
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Middle Ear

- Epitympanum = Above TM
- Mesotympanum = Behind TM
- Hypotympanum = Below TM



<http://www.radiologyassistant.nl/en/43facba0911f5>



<http://www.radsmd.com/ossiclesPrussaksSpace.htm>

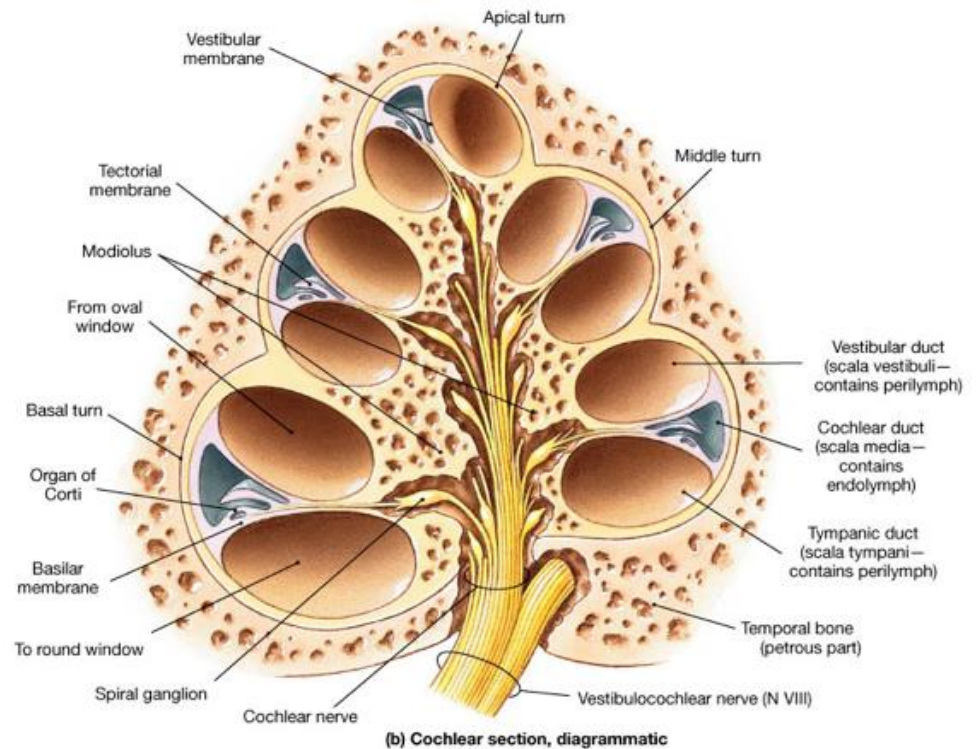
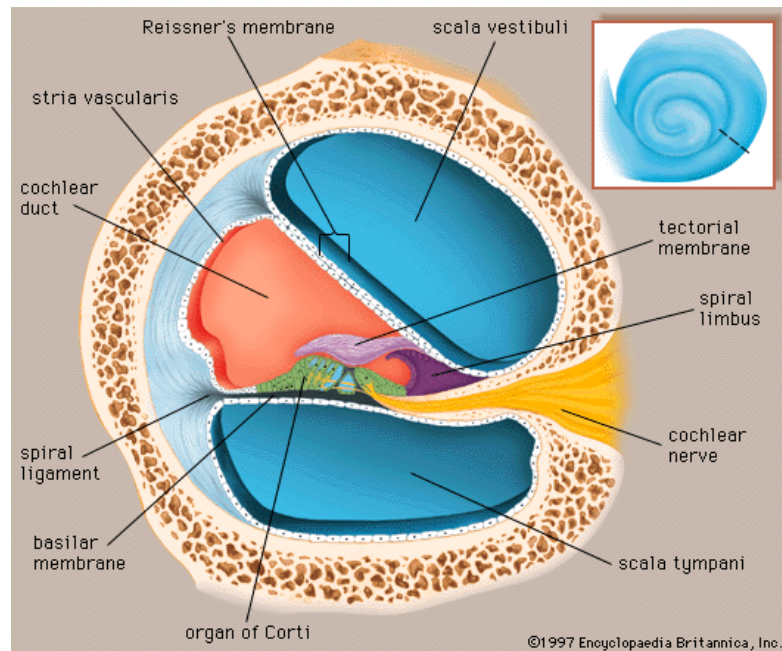


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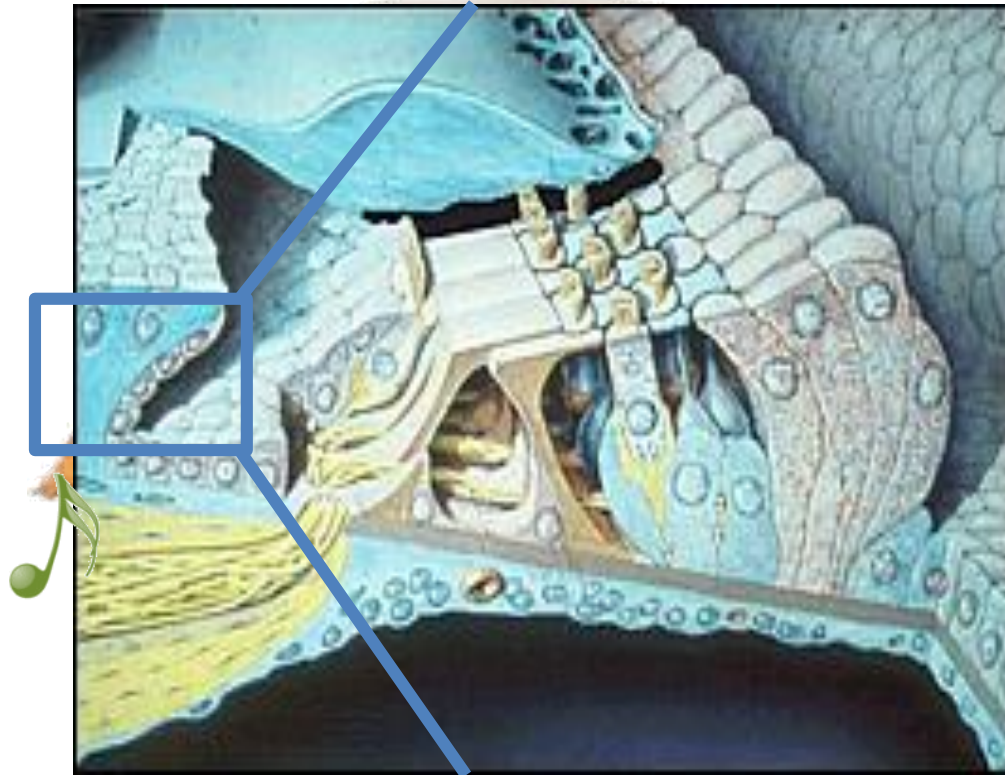
Bony and Membranous Labyrinth



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Let's look at the cochlea...

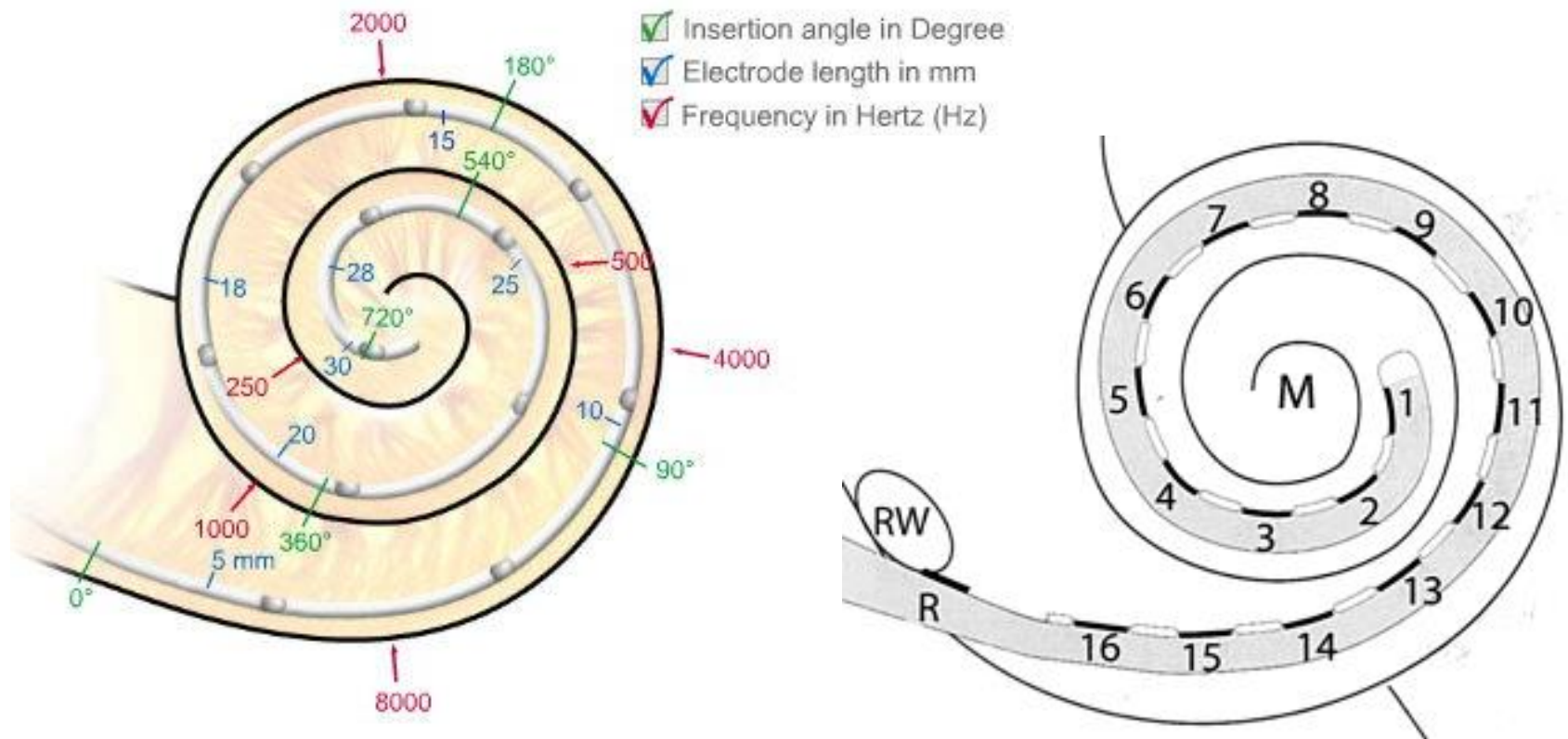


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Cochlea is Tonotopic



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Spiral Ganglia



<http://bmc.erin.utoronto.ca/~andreaaz/newMediaSite/>

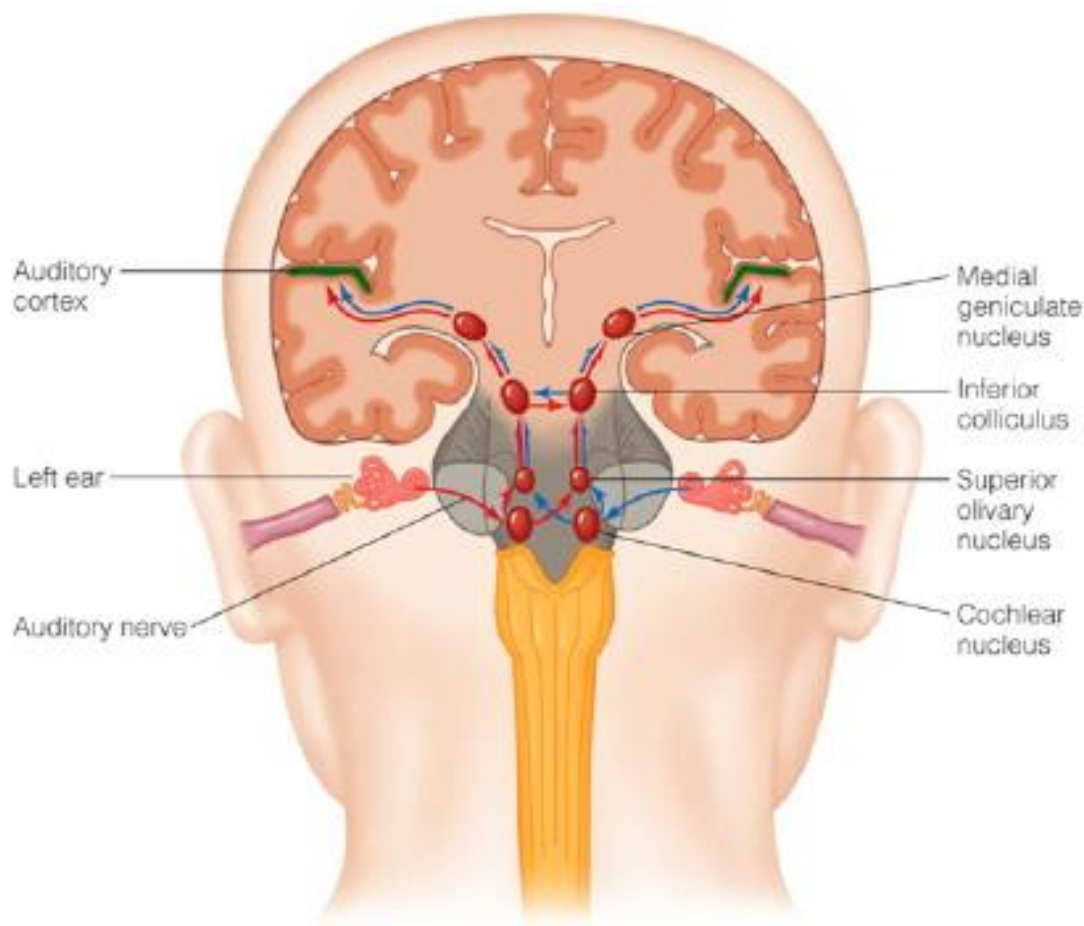


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Intracranial Pathway



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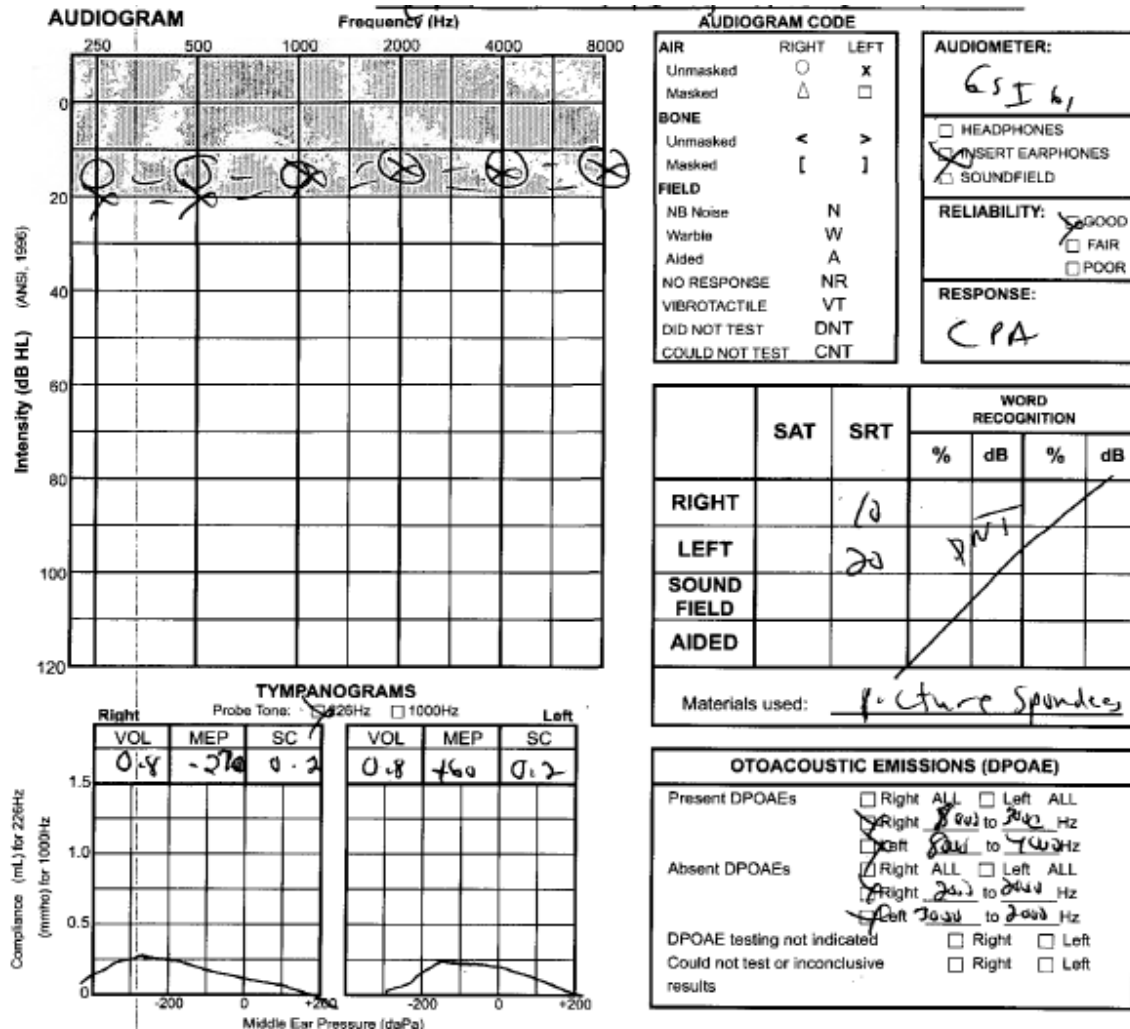


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<http://www.ssc.education.ed.ac.uk/courses/deaf/dnov10i.html>

Normal Audiogram



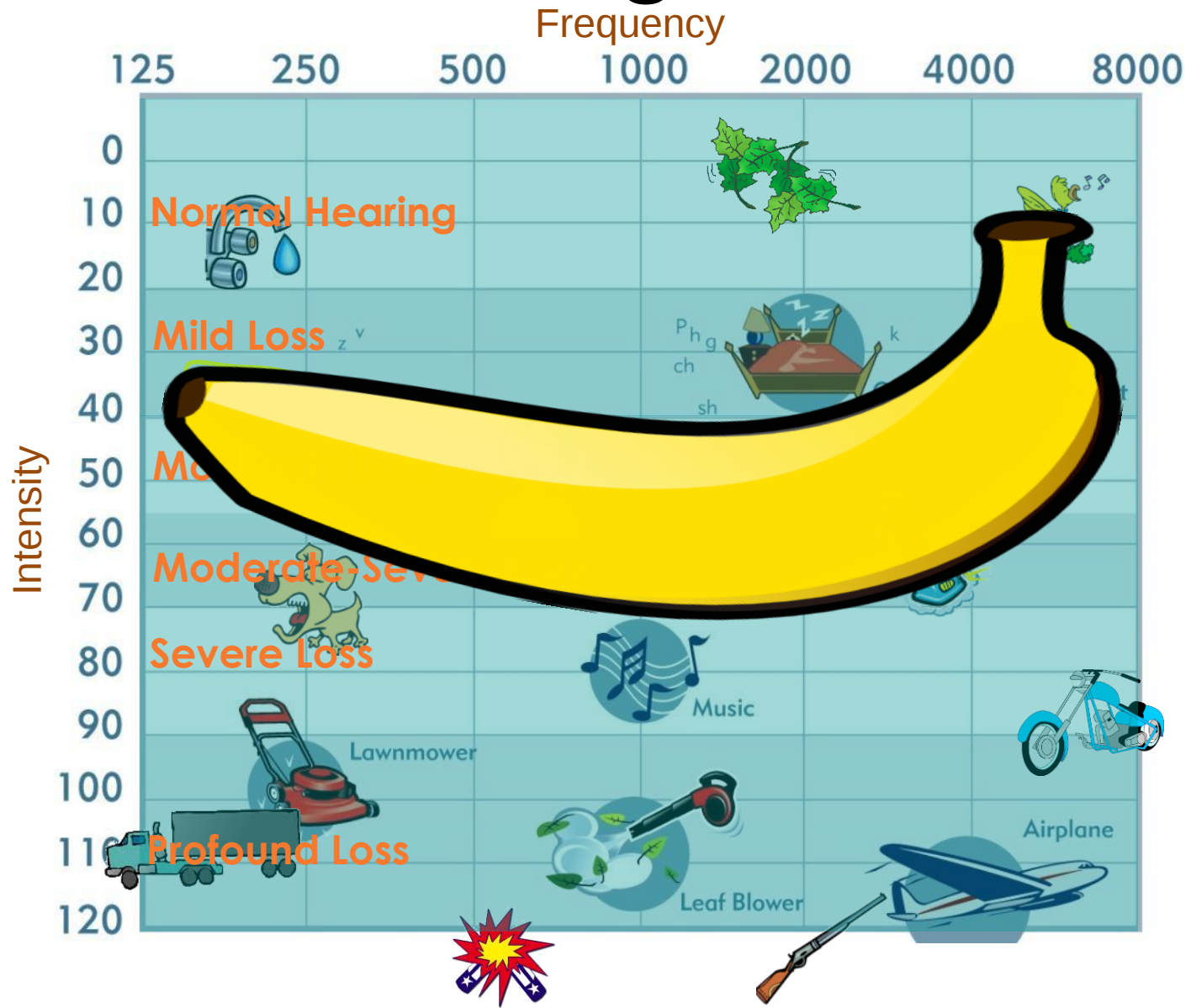
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The Audiogram



Management



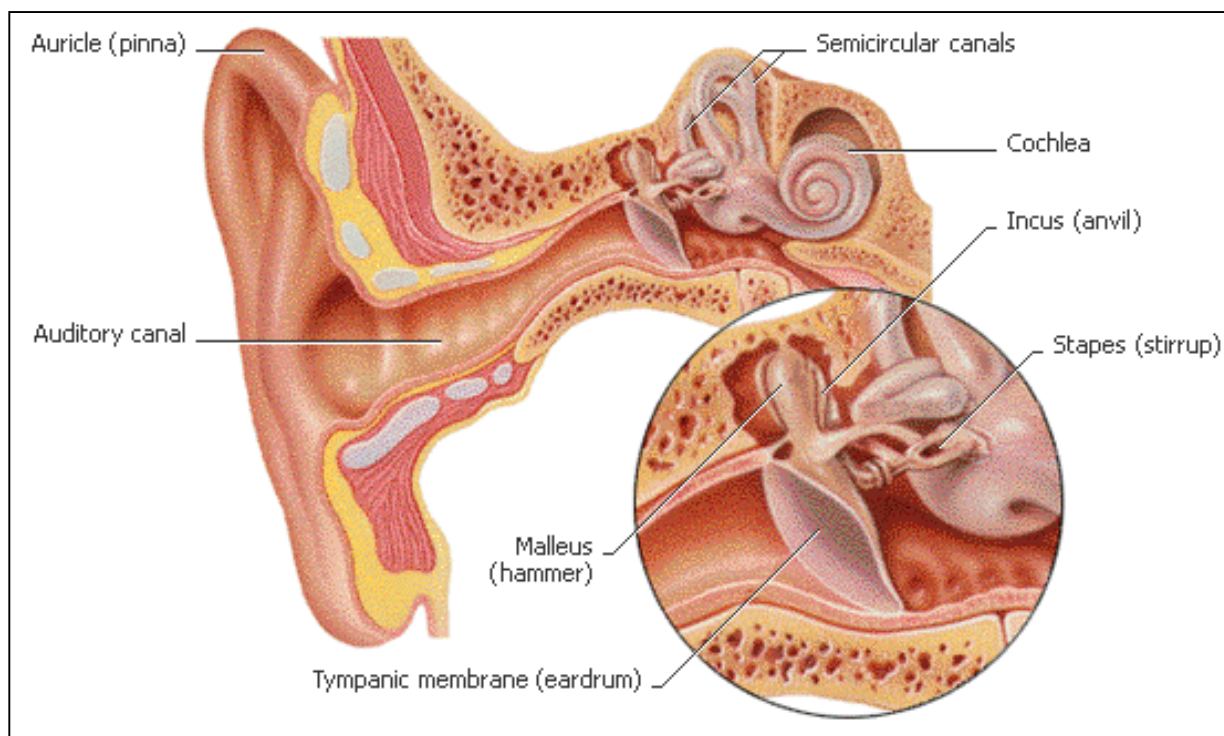
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Conductive Hearing Loss

- Interference with reception and transmission of sound through the outer and middle ears



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Conductive Hearing Loss

- Aural Atresia
- Ear wax
- Foreign body in ear
- Perforated ear drum
- Middle ear infection or fluid
- Ossicular erosion, discontinuity, fixation or malformations
- Middle ear masses



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Surgery for Conductive Hearing Loss

- Repair of aural atresia
- Tympanostomy tubes (667,000/year)
- Myringoplasty/Tympanoplasty
- Removal of masses (i.e cholesteatoma)
- Management of ossicular fixation/discontinuity
 - Ossicular Chain Reconstruction
 - Stapedectomy
- Bone Anchored Auditory Implant

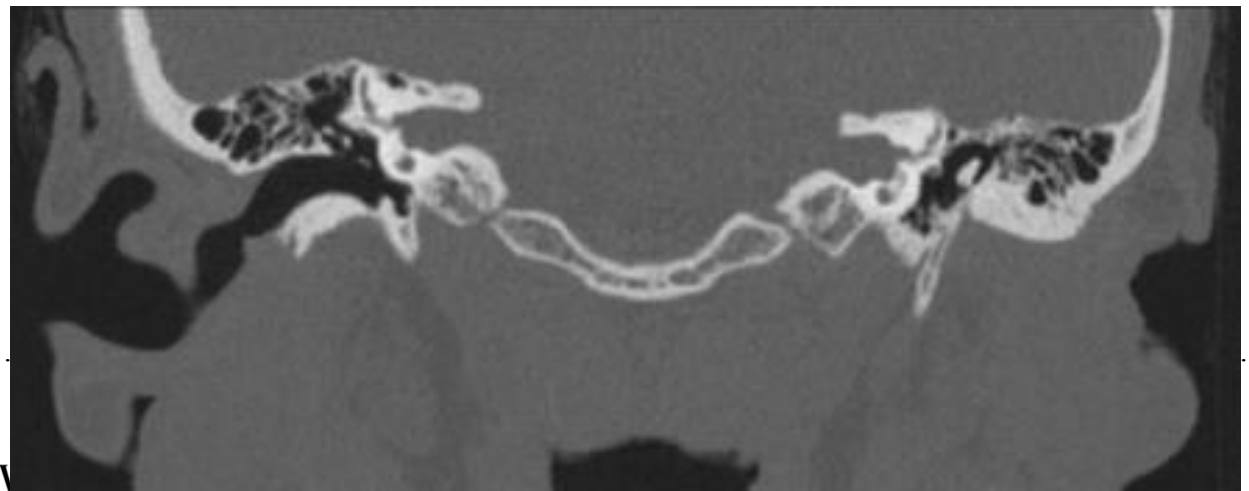
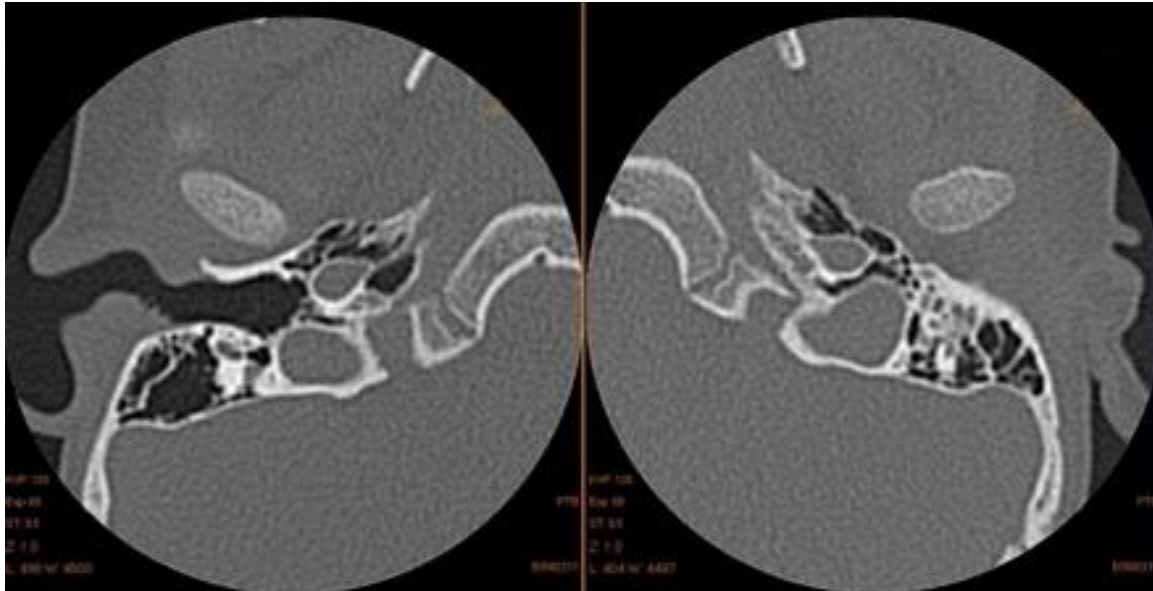


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Abnormal EAC – Aural Atresia



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Eustachian Tube

- Connects middle ear to nasopharynx
- Opens by muscular action
- Pressure equalization across TM
- **Eustachian Tube Dysfunction (ETD)**



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Tympanostomy Tubes – Otitis Media with Effusion (OME)

- Recurrent acute otitis media (3/6 months, 4/12 months)
- Bilateral OME > 3 months with:
 - Hearing loss or symptoms from ETD
- Unilateral or Bilateral OME any duration in “at risk” kids:
 - Speech and language disorders
 - Coexistent permanent hearing loss
 - Autism spectrum, ADHD...
 - Developmental delay
 - Syndromes (Down) or craniofacial disorders that include cognitive, speech or language delays
 - Cleft palate, with or without associated syndrome
 - Blindness or uncorrectable visual impairment

AAO-HNS Guidelines 2013

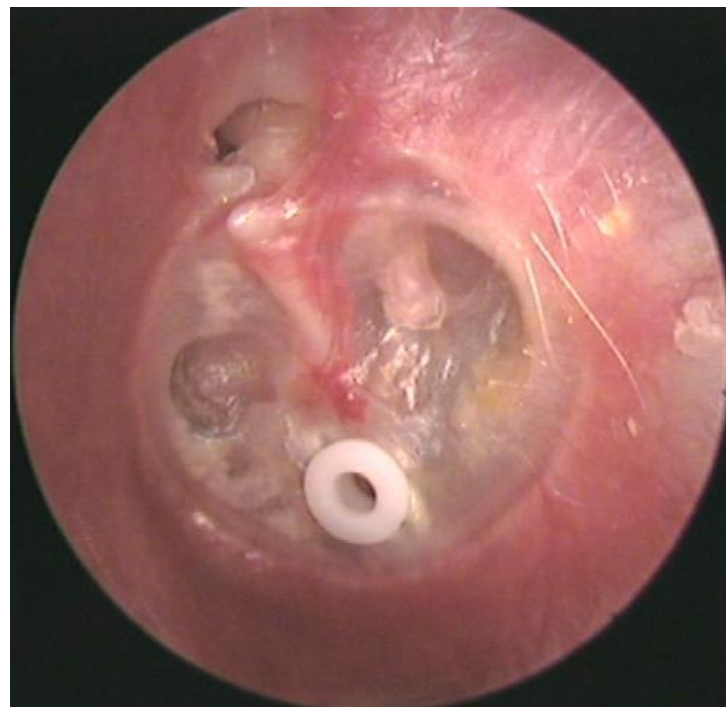
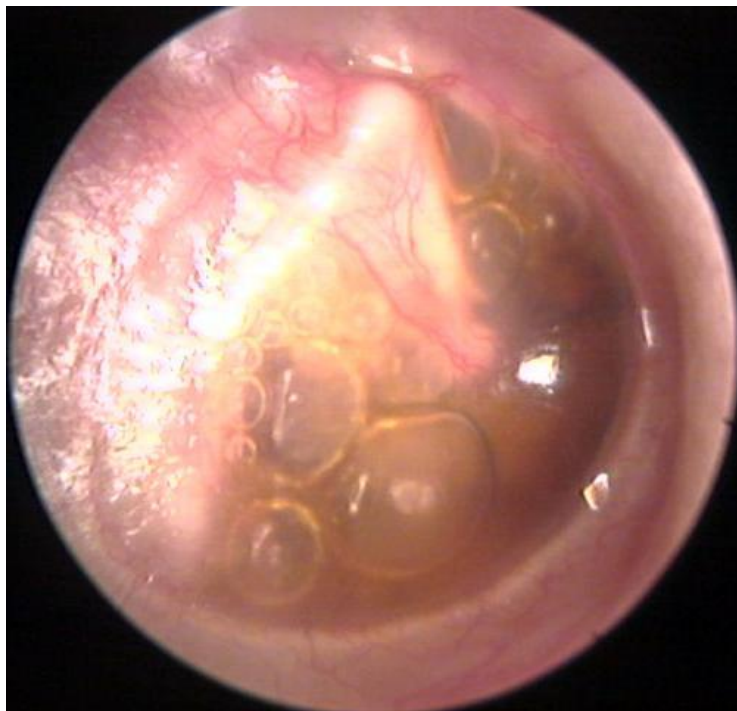


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Conductive Hearing Loss



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ETD and Chronic Otitis Media

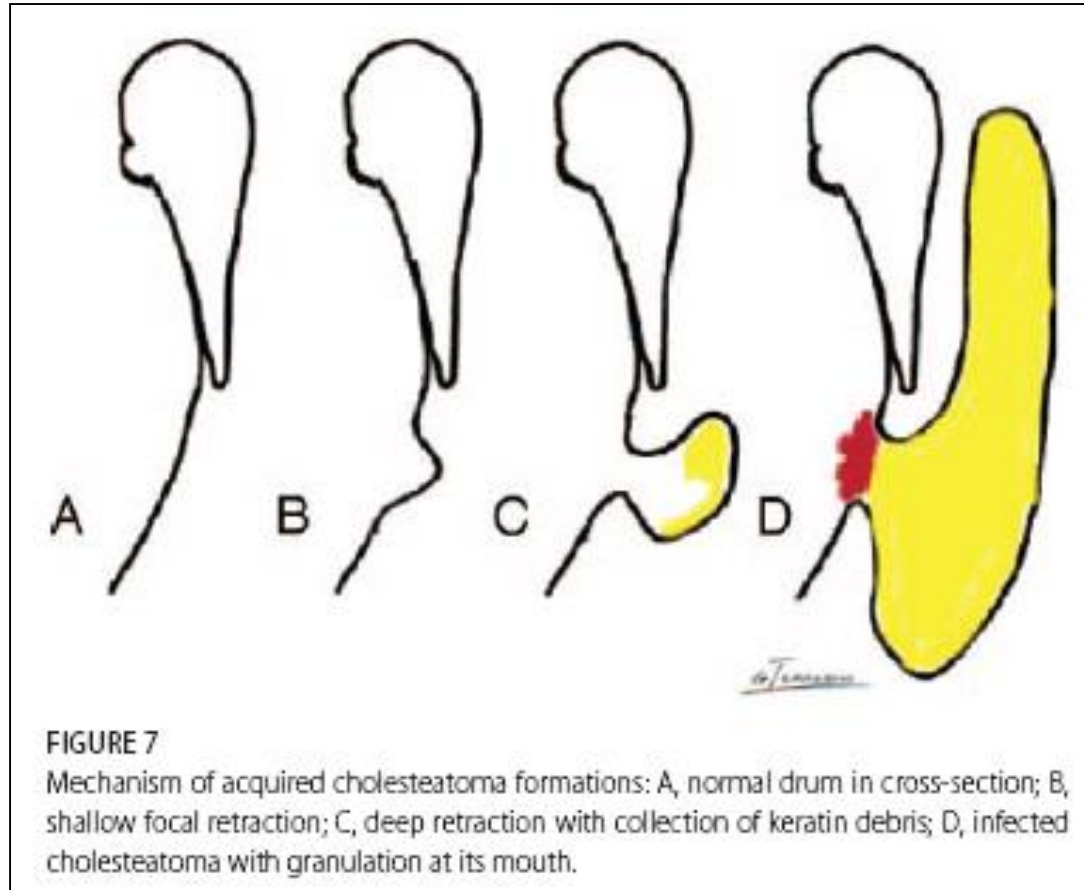


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Formation of Cholesteatoma (Acquired)



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Cholesteatoma



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Cholesteatoma



Other Whitish Things



Acute Otitis Media



Myringosclerosis



Myringosclerosis



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Audiogram

- **CONDUCTIVE HEARING LOSS**
 - If no fluid, be concerned!!
 - If **>40 dB**, be concerned!!
- Mass effect on middle ear transformer
- Erosion of ossicles
- TM perforation
- Altered TM mobility



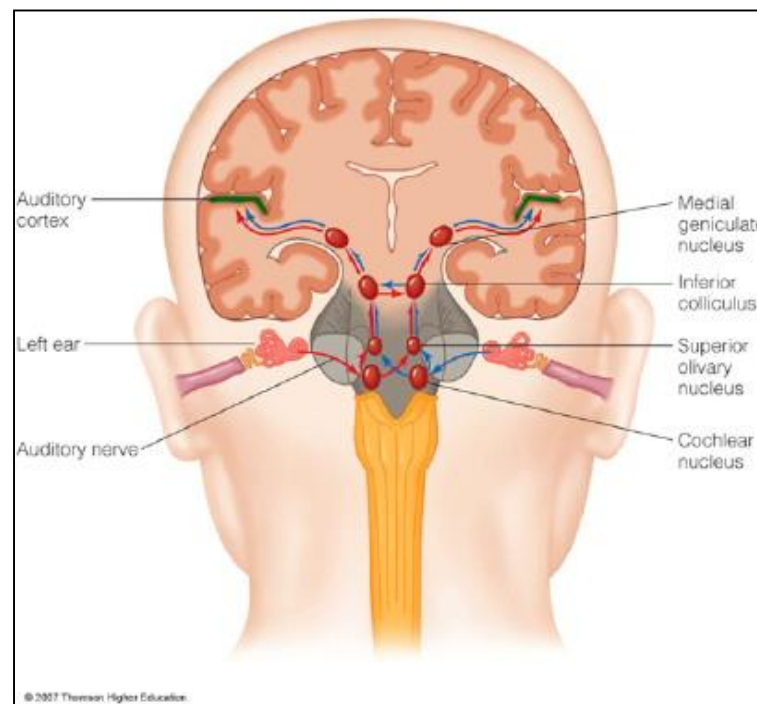
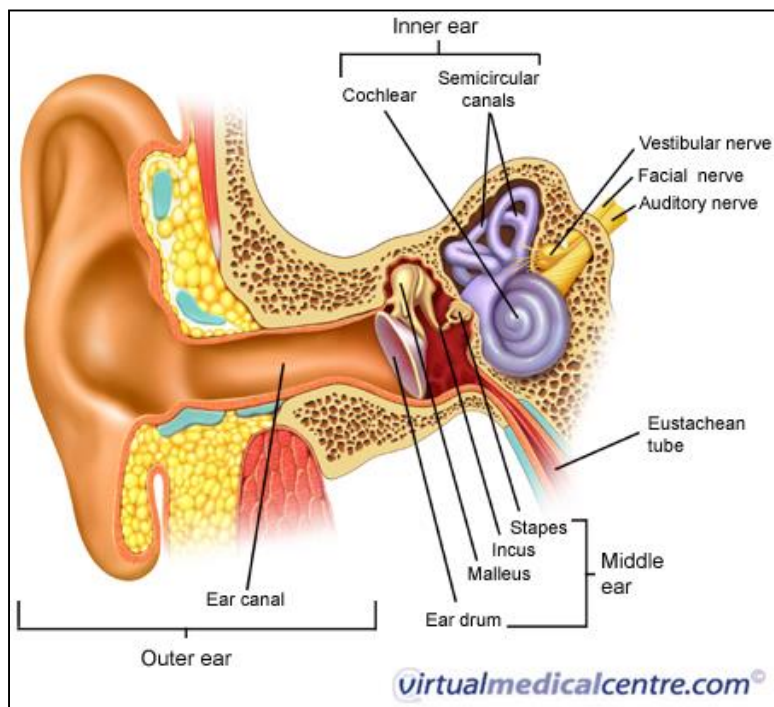
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Sensorineural Hearing Loss

- Due to damage or deficit in the inner ear or auditory nerve



Management of Hearing Loss

- Non-surgical habilitation
 - Amplification (Hearing Aids)
- **Surgical habilitation**
 -
- Speech and language therapy
- Early Intervention Services
- Regular audiologic follow up



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Hearing Aids



Cochlear Implant – Audiologic Candidacy

FDA approval: 1 yr and older

- 1 yr to 2 yrs: BILATERAL **profound** sensorineural hearing loss
- >2 yrs old: BILATERAL **severe to profound** sensorineural hearing loss
- Limited auditory development with best fit hearing aids
 - IT-MAIS for younger kids
 - Open-set word recognition scores for older kids
 - e.g. MLNT/LNT $\leq 30\%$

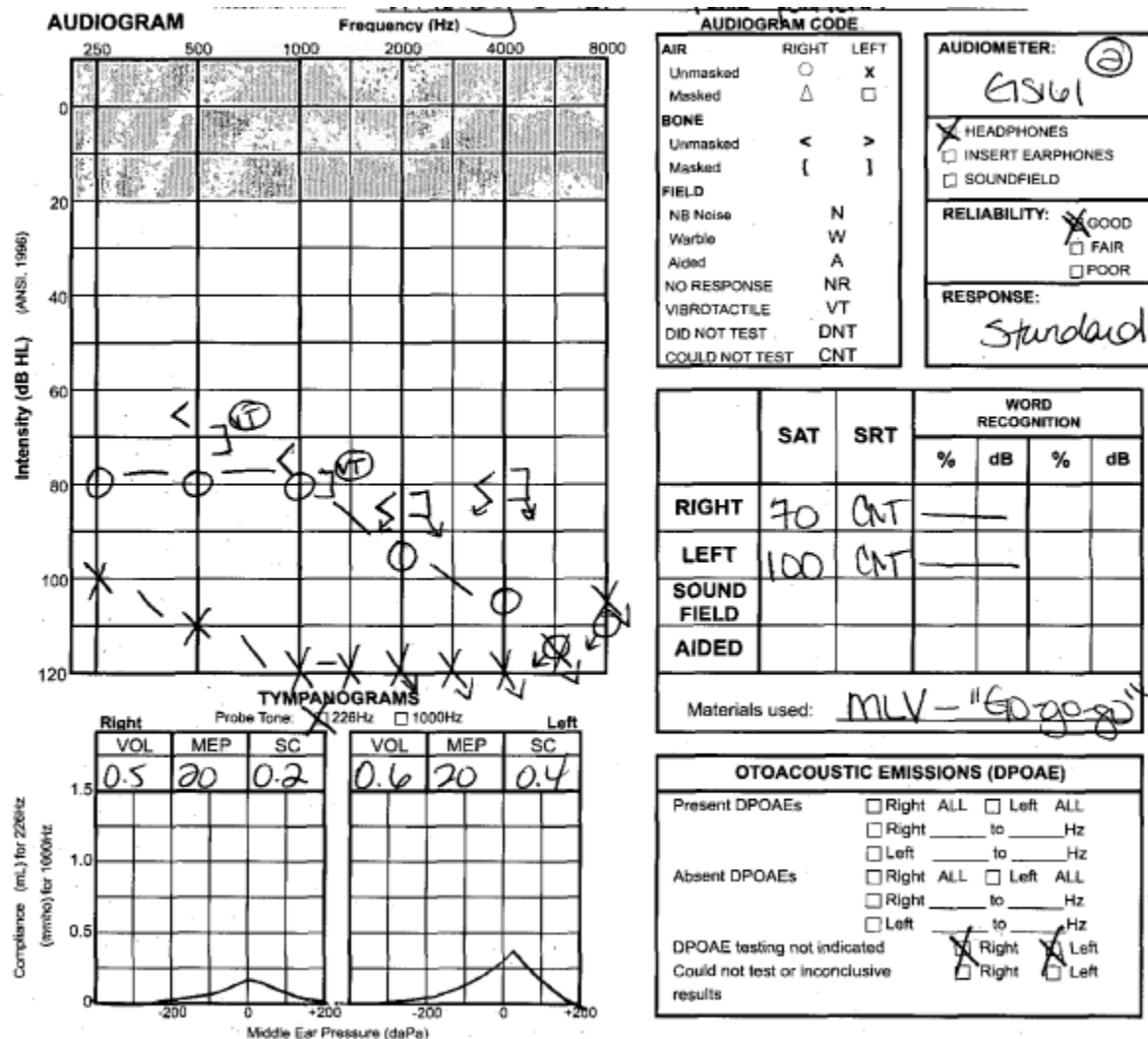


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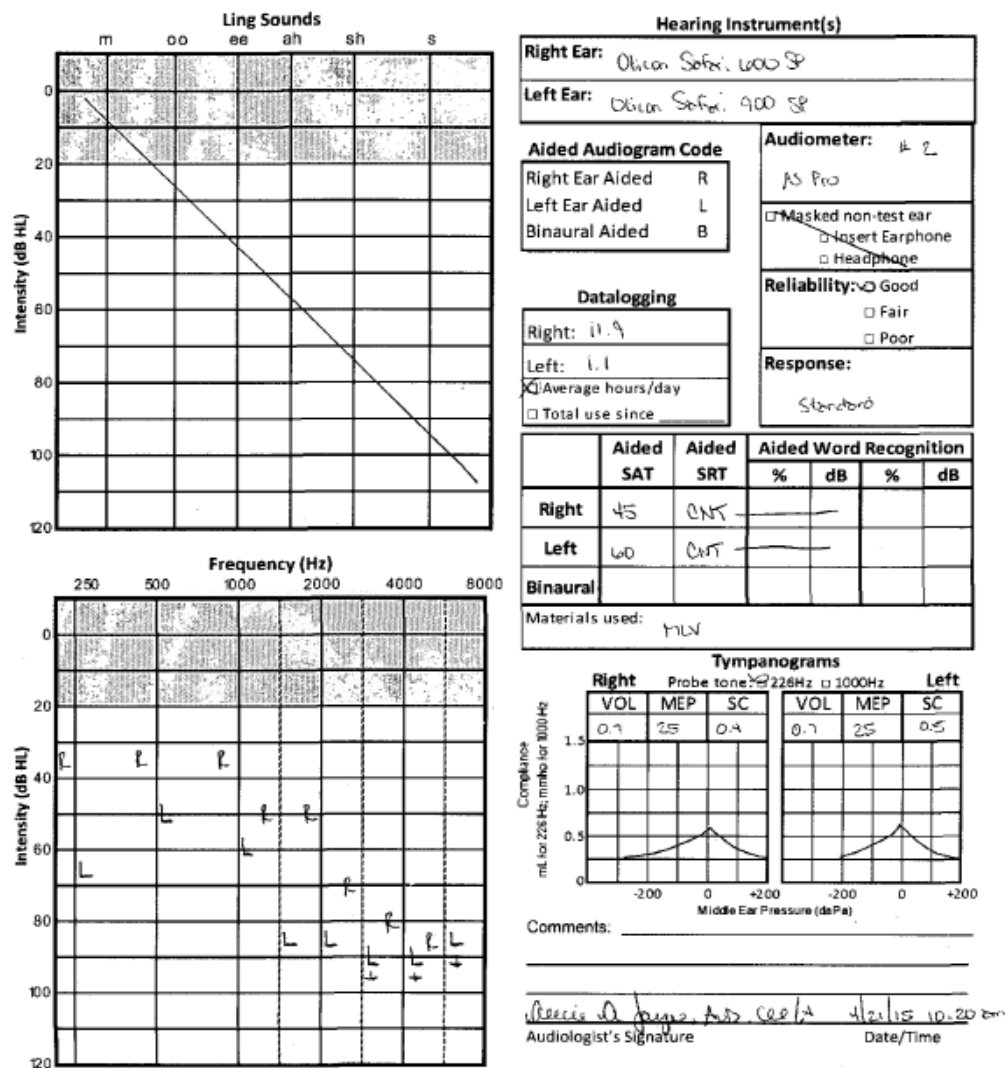


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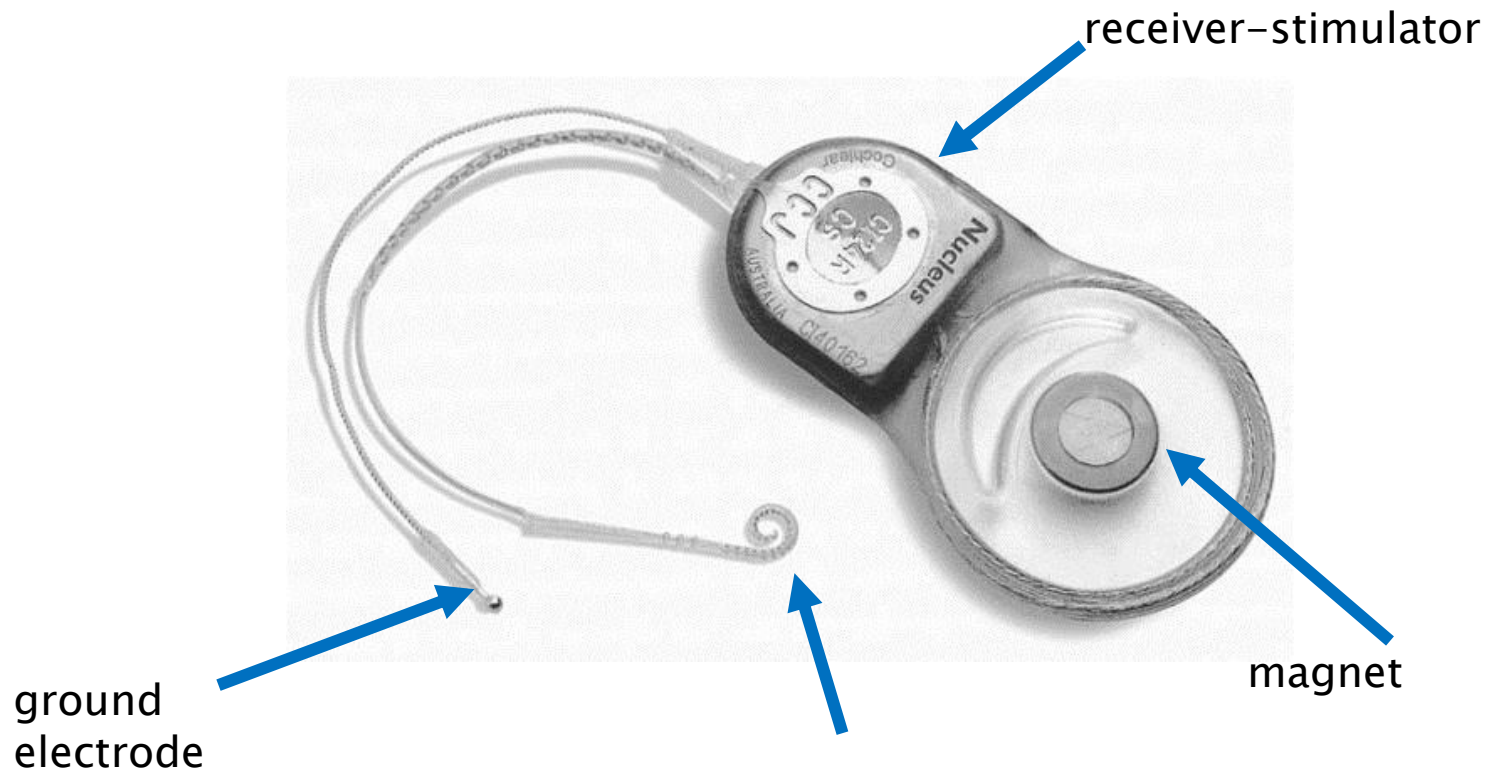
Bilateral Severe to Profound Sensorineural Hearing Loss



Insufficient Hearing Aid Benefit



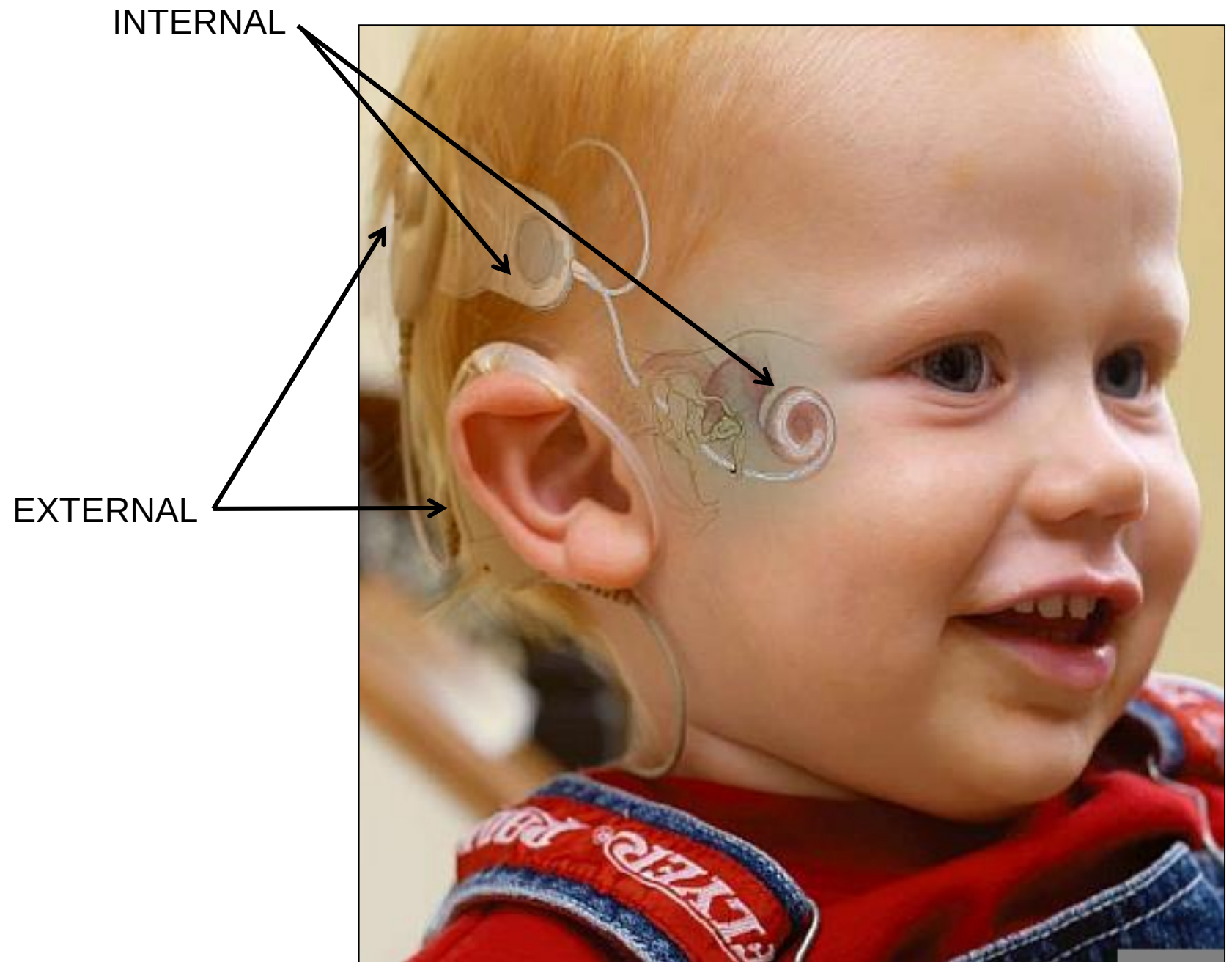
The Cochlear Implant

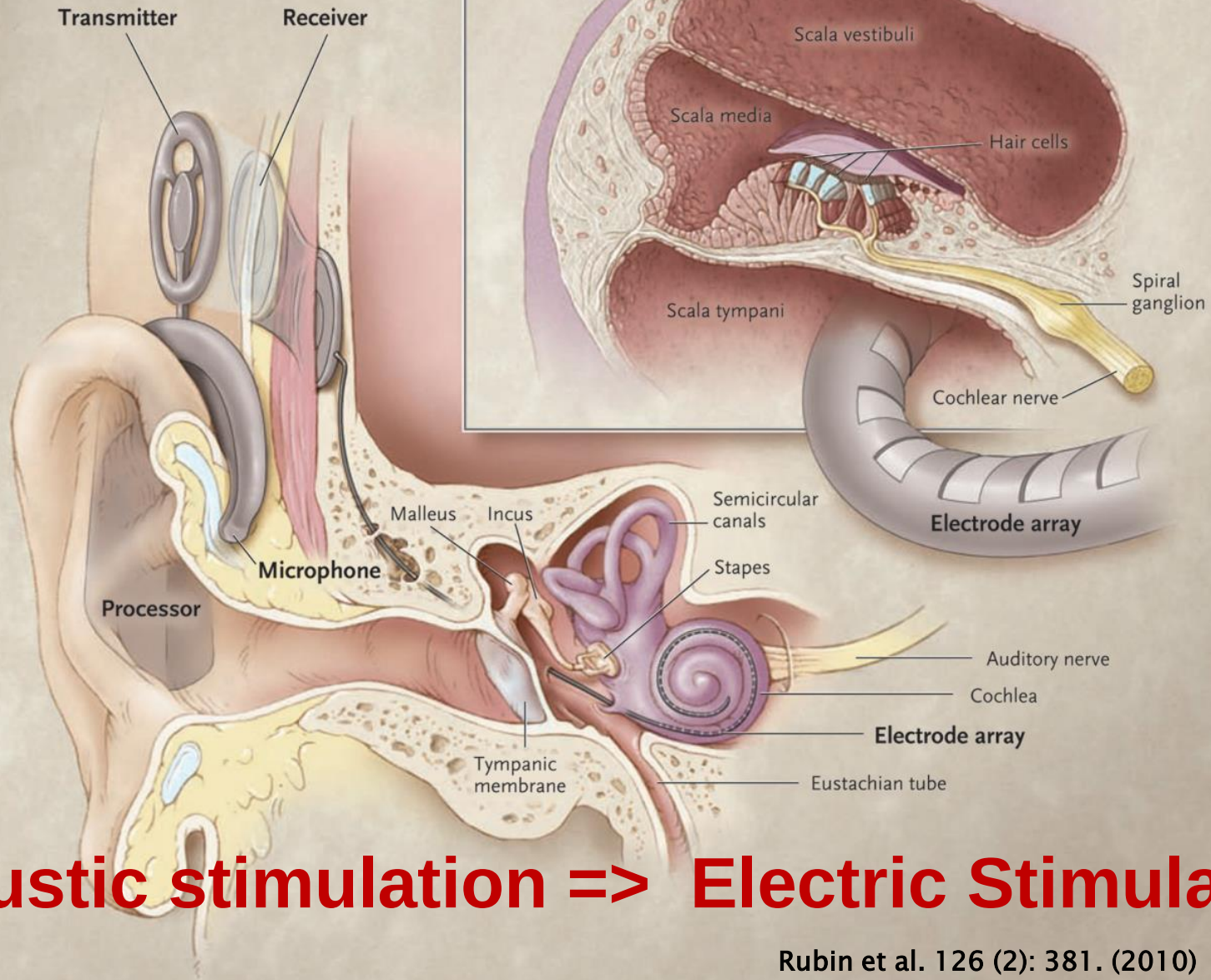


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Acoustic stimulation => Electric Stimulation

Cochlear Implant Evaluation

- Audiologic diagnosis of SNHL (electrophysiologic)
- Medical evaluation and MRI (ENT)
- Audiologic confirmation of SNHL (behavioral)
- Best hearing aided evaluation
- Speech/Language evaluation
- Vaccination
- Financial clearance
- Implantation surgery (**by 1 year of age**)



Every kid is counseled uniquely on THEIR expectations

Post-operative Habilitation !!! Sound =>Speech.

Social work evaluation, Neuropsychological evaluation, Educational assessment,
.....Infectious Diseases (CMV).....

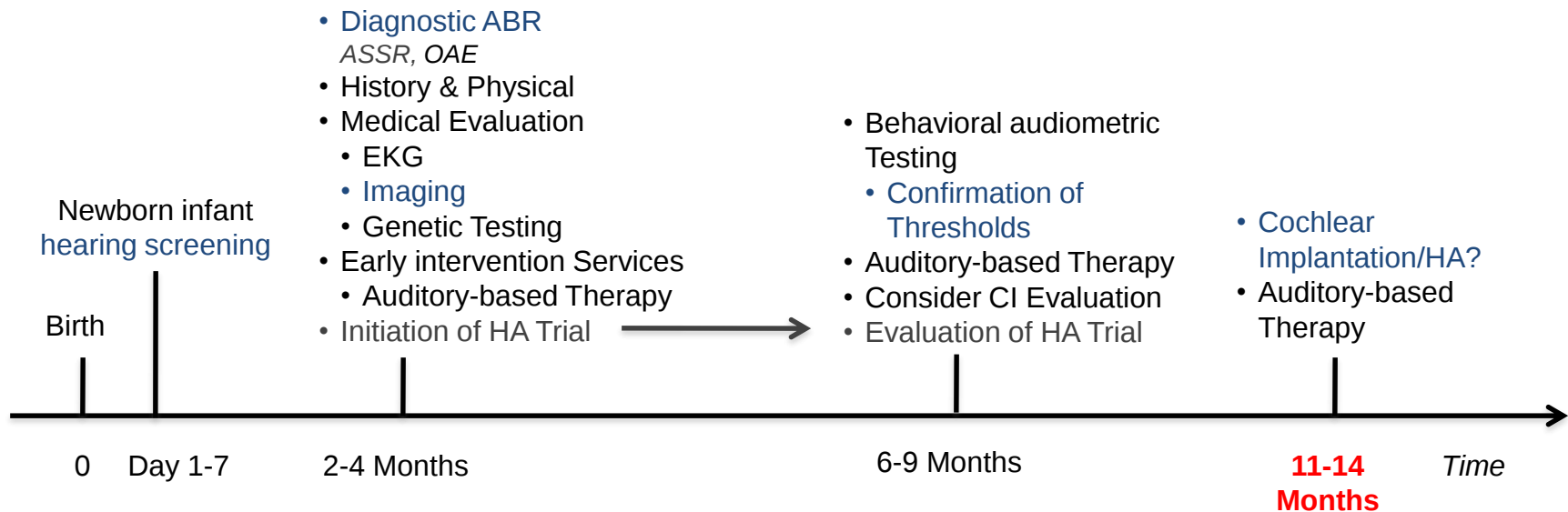


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Timeline



Courtesy, Dr. Oliver Adunka

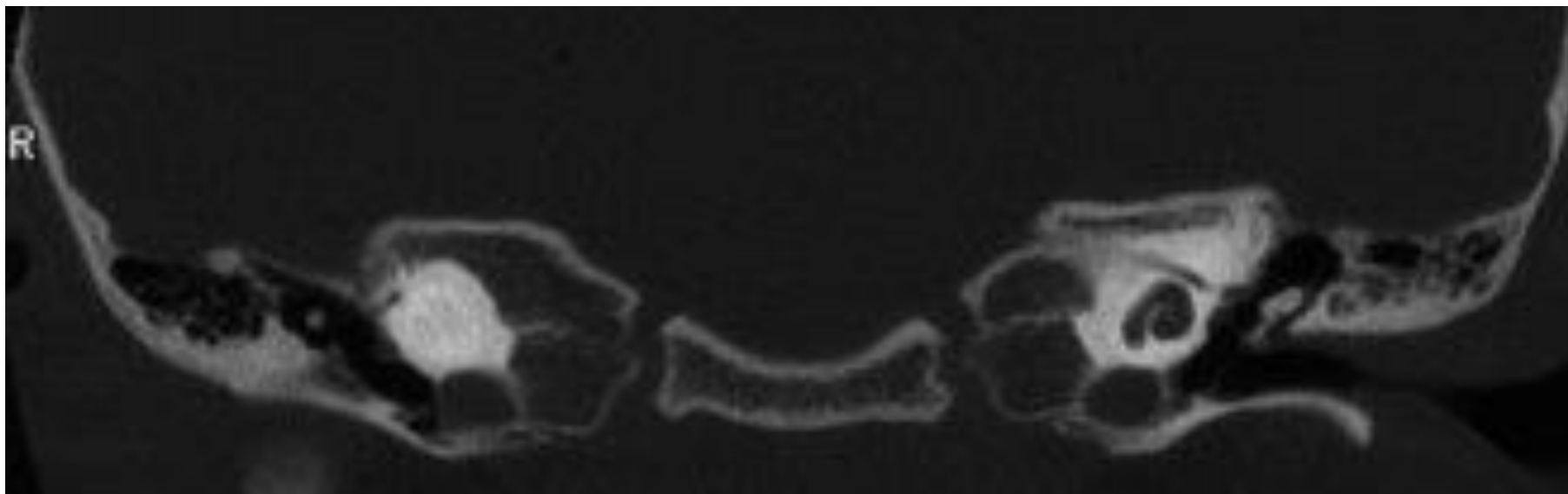


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Imaging

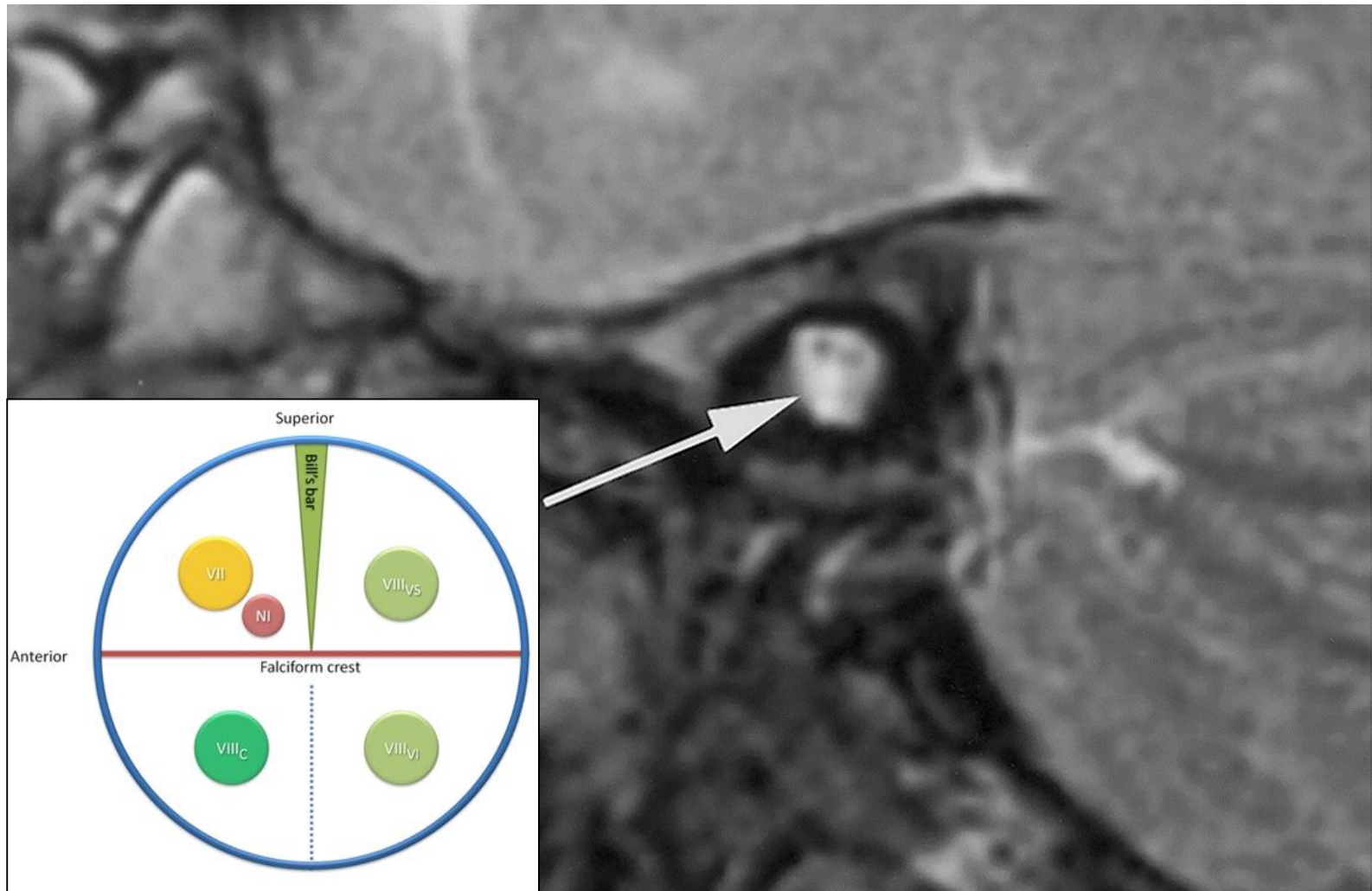


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MRI (FSE) - RIGHT



Witte R J et al. Radiographics 2003;23:1185-1200

RadioGraphics

Vaccination and CI

- Bacterial meningitis is more common in children with cochlear implants than in controls
- Increased risk of meningitis **(4-30x)** due to **S. Pneumoniae**

Recommendations:

- All children should get PCV-13 and Hib (as per routine)
- At age 2, PPSV23 completed at least 2 weeks before implant surgery
- Influenza vaccine to reduce otitis media

Critical, and **REQUIRED** step before implantation, *Pediatrics* Volume 126, Number 2, August 2010 385



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Surgery

- 2-3 hours (in OR to out of OR)
- Outpatient



(Right Ear)

↑ Anterior

(Ear Canal)

→ Inferior

Acc 1 - Still

IMAGE₁
HD[®]
KARL STORZ — ENDOSKOPE

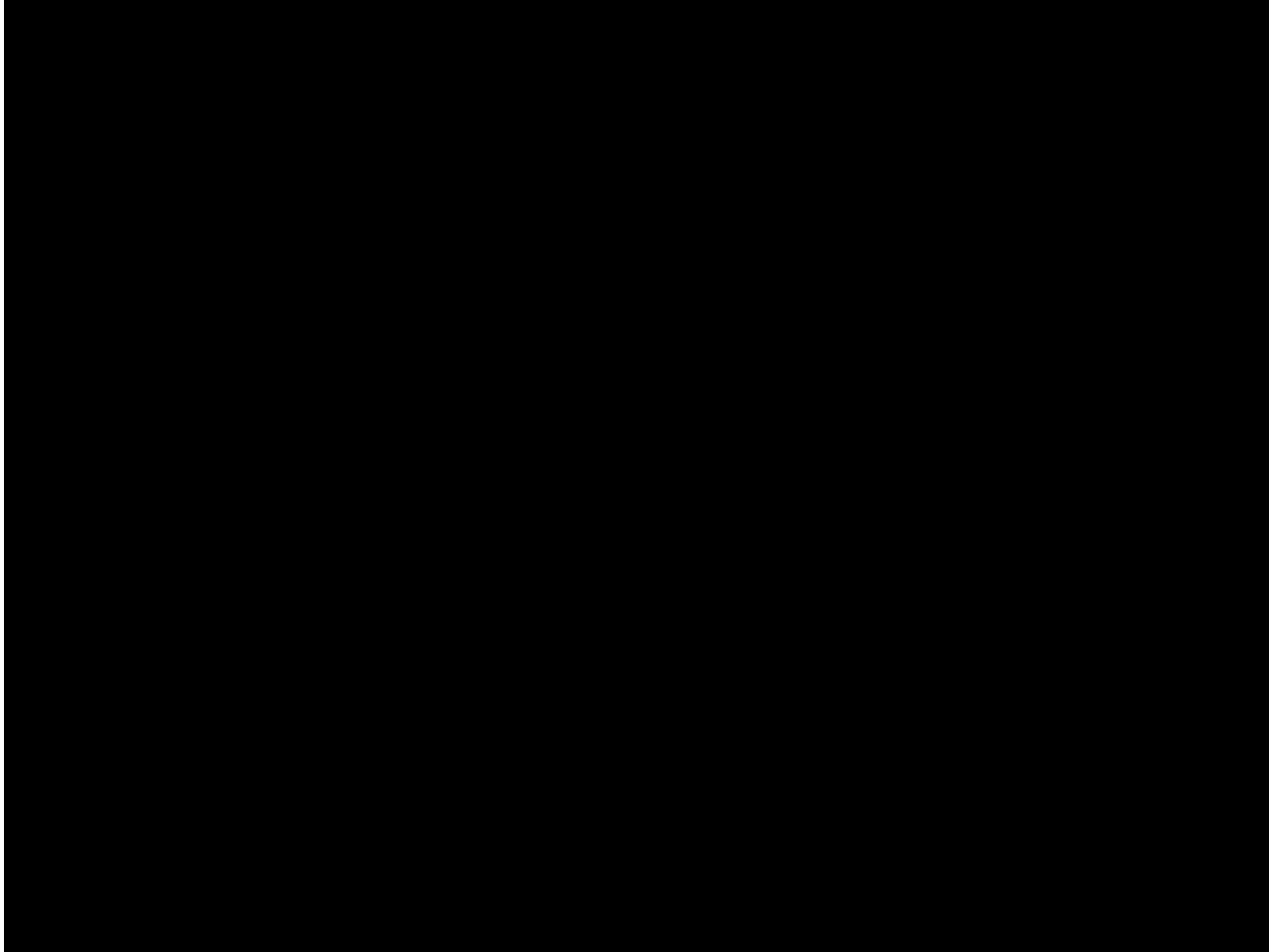


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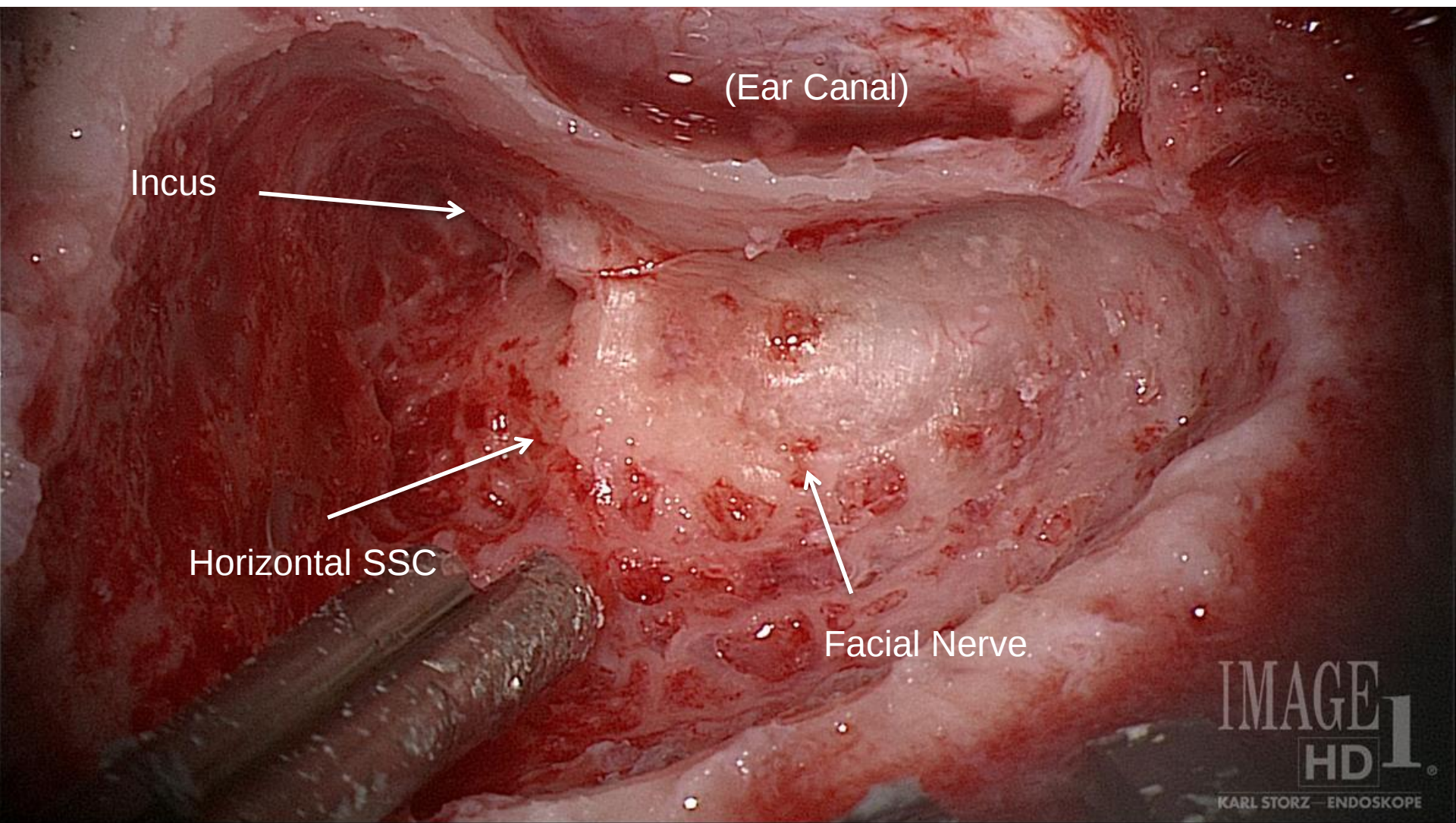
Surgery



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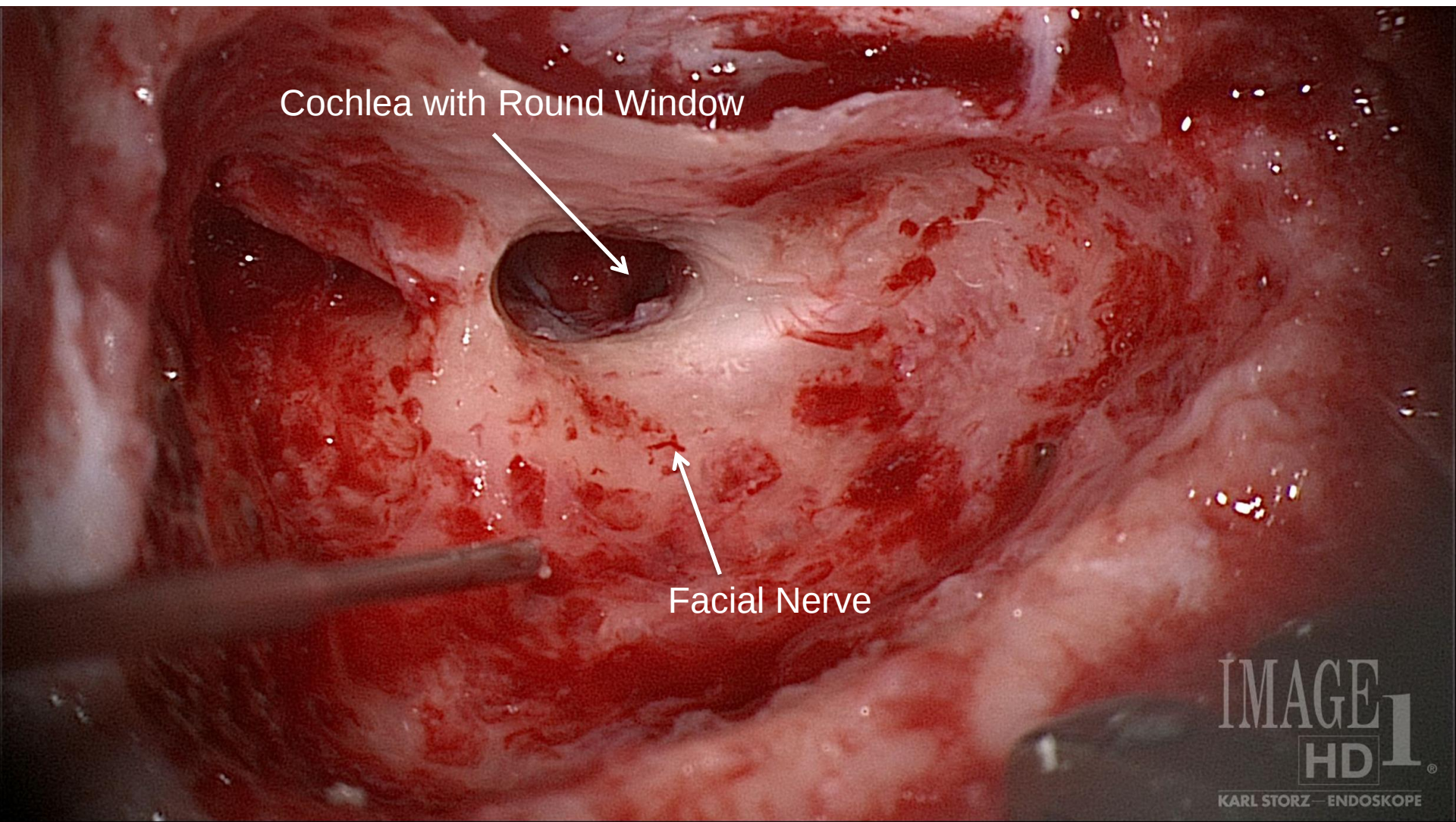
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Cochlea with Round Window

Facial Nerve

IMAGE¹
HD
KARL STORZ — ENDOSKOPE



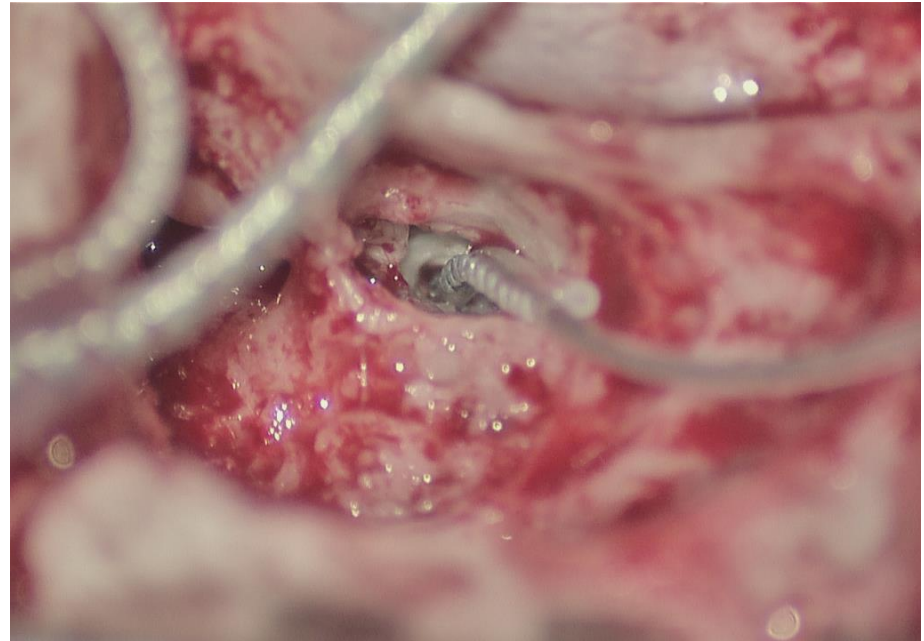
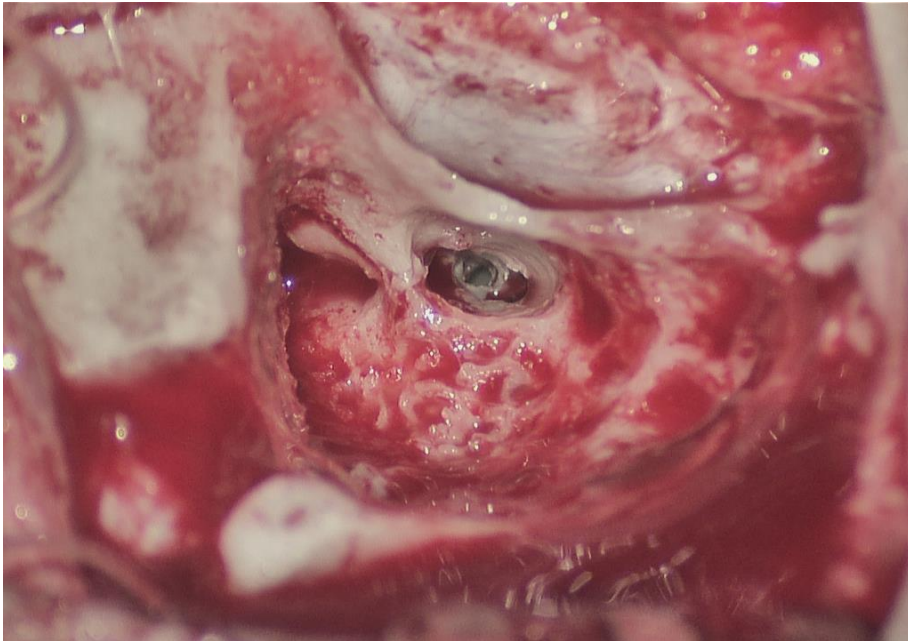
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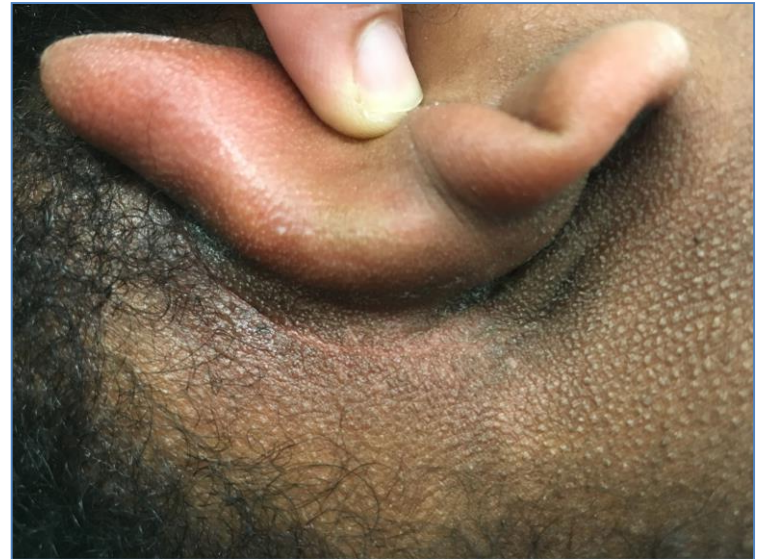
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Surgery

- Electrode Insertion



Surgery



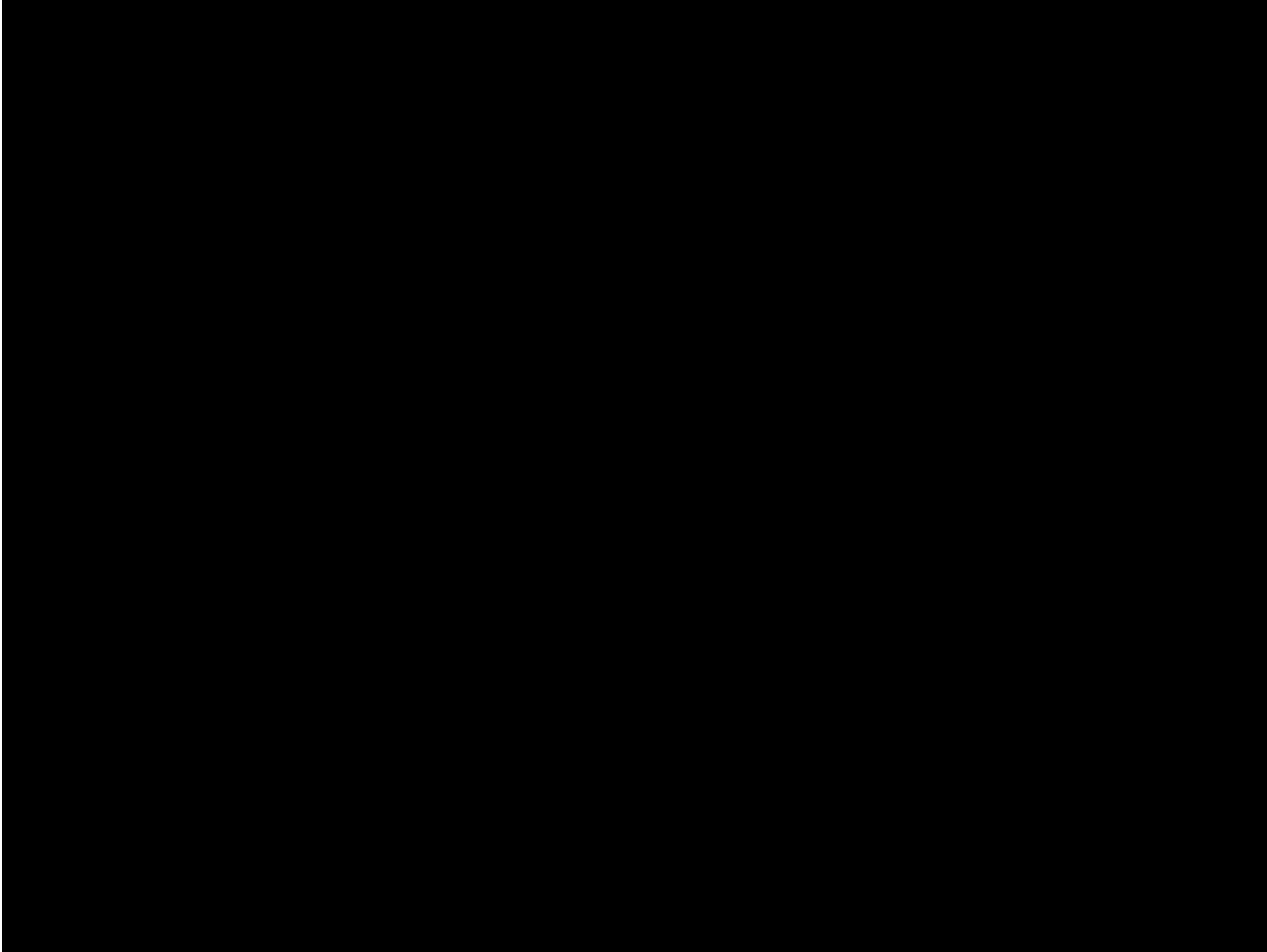
Activation

- 2-3 weeks, after incision has healed and pain and edema gone



Go to YouTube and see these...

Activation

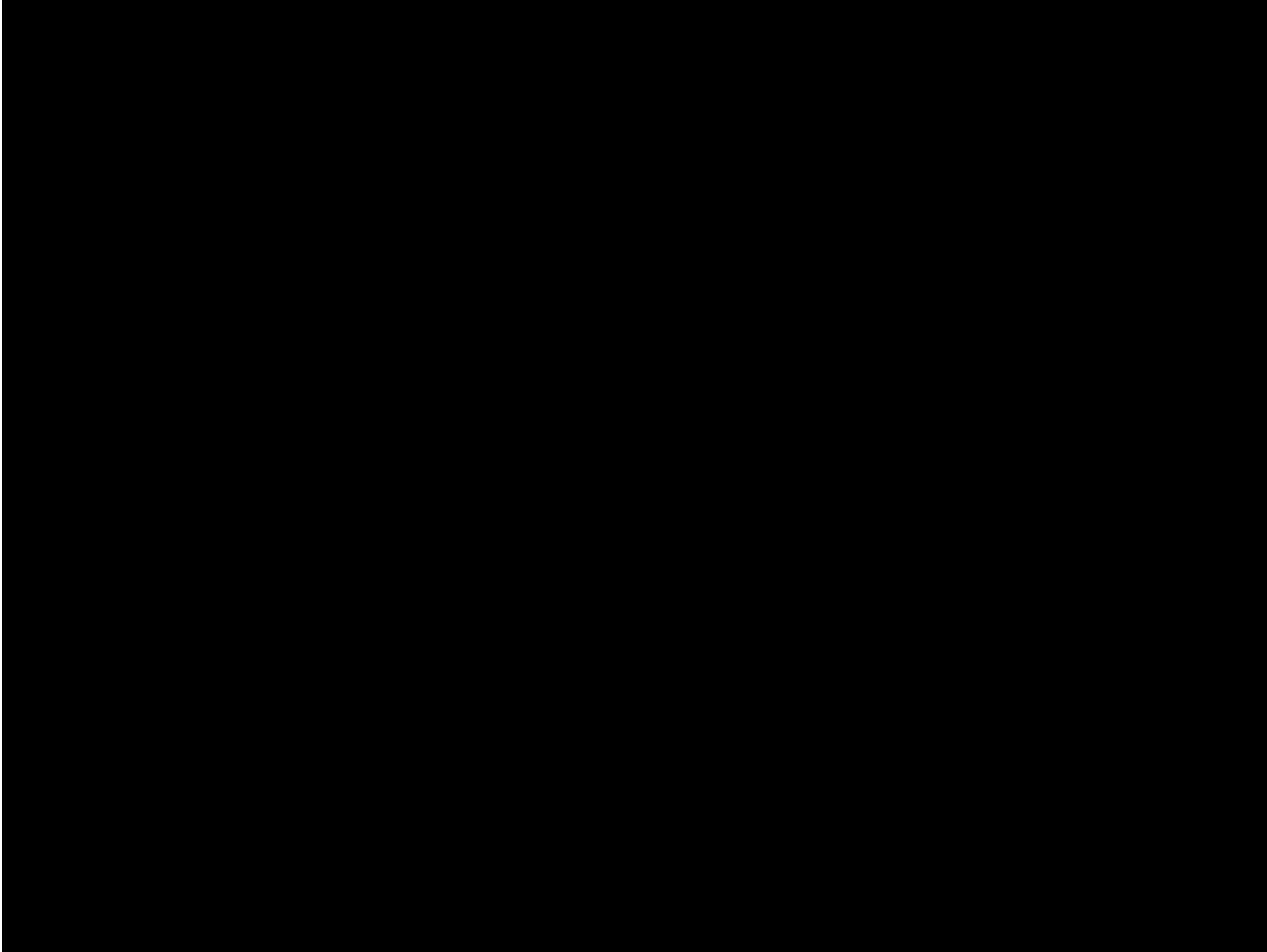


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Activation



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Outcomes – Patient Factors

- **Age at implantation**
- **Auditory Foundation (neuronal activity?)**
 - Duration of deafness
 - Preoperative hearing/speech understanding
- **Etiology (*e.g. Connexin*)**
- **Cognitive level/intelligence**



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Speech Perception

- If no context (monosyllabic words), can understand 55-60%
 - (*Dorman et al, 2008*)
- With context (sentence recognition), can achieve 90% in quiet settings
 - (*Davidson et al, 2011*)



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General Outcomes

- 60-80% can use telephone
- 40-70% have intelligible speech
- 80-90% do not require lip reading
- Can achieve similar academic levels as normal hearing peers
- Can achieve similar occupational levels



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CDaCI Studies

- Multicenter trial 2002-2004, NIDCD funded
 - N=188 CI vs. 97 normal hearing

RDLS

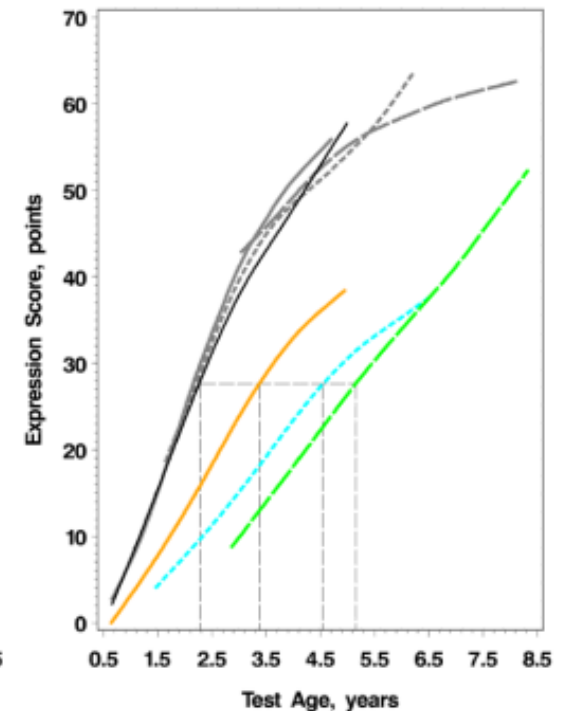
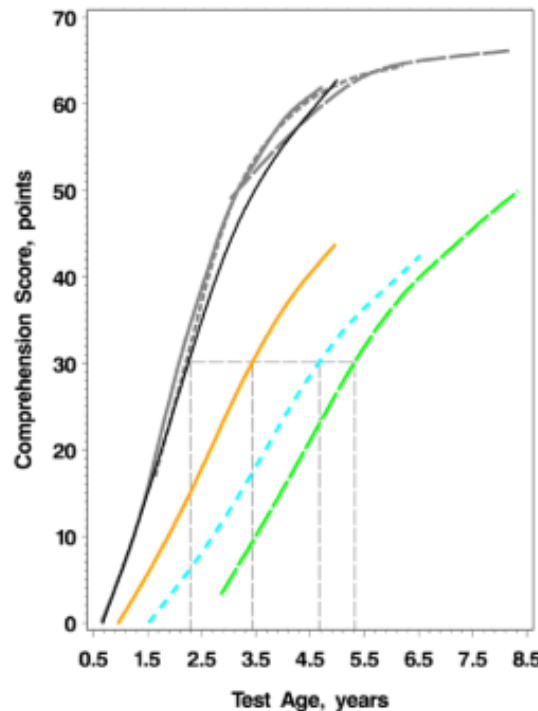
- Receptive Language
- Expressive Language

NH = 2.3/2.3 yrs

CI <18 months = 3.4/3.4 yrs

CI 18-36 months = 4.7/4.5 yrs

CI >36 months = 5.3 yrs/5.2 yrs



JAMA 2010



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CDaCI Studies

- Mainstream classroom integration:
 - After 6 years, 81% vs 57% vs 63%
 - For non-CI (severe to profound SNHL) = 12%

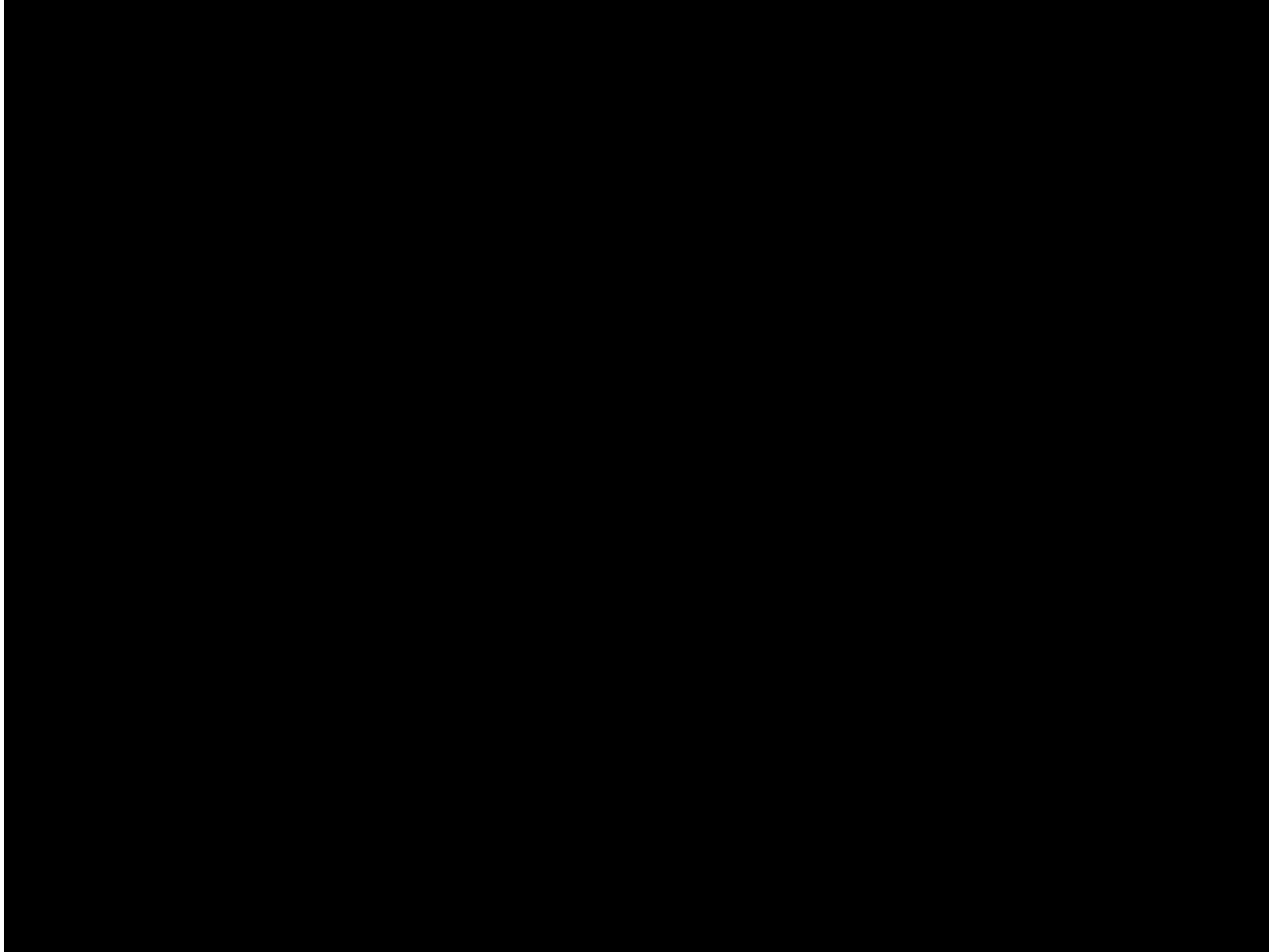


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Bilateral CI at 10 months



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Screening Beyond Infancy - Rationale

- UNHS designed to target hearing losses 30-40 dB or more
- All newborns “referring” on UNHS do not get follow up
- UNHS cannot identify delayed onset or progressive hearing loss
- Only 50% of children with hearing loss are identified with risk factor questionnaires



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Risk Factors – After Newborn

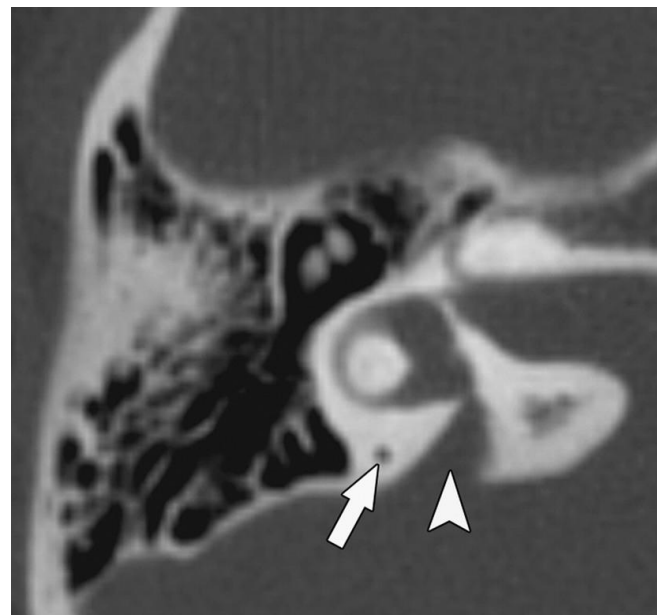
Table 1. Risk Indicators for Audiologic Monitoring for Progressive or Delayed-Onset Sensorineural Hearing Loss, Conductive Hearing Loss, or Both, in Infants (29 Days through 2 Years of Age) with Normal Hearing on Newborn Screening.*

Parental or caregiver concern regarding child's hearing, speech, language, or developmental delay
Family history of permanent hearing loss in childhood
Stigmata or other findings associated with a syndrome known to include a sensorineural or conductive hearing loss or eustachian-tube dysfunction
Postnatal infections associated with sensorineural hearing loss, including bacterial meningitis
In utero infections such as cytomegalovirus infection, herpes, rubella, syphilis, and toxoplasmosis
Neonatal indicators such as 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, osteopetrosis, and some forms of Usher's syndrome
Neurodegenerative disorders such as Hunter's syndrome or sensory neuropathies such as Friedreich's ataxia and Charcot-Marie-Tooth syndrome
Head trauma
Recurrent or persistent otitis media with effusion for at least 3 months

* Information is from the Joint Committee on Infant Hearing.²¹

Etiology of Delayed Onset HL

- Middle ear disorders (ETD, COME, cholesteatoma)
- Actually a congenital loss...
- Noise Induced
- Otosclerosis
- Meniere's
- Autoimmune Hearing Loss
- Enlarged Vestibular Aqueduct
- Idiopathic Sudden Sensorineural Hearing Loss



Large vestibular aqueduct syndrome in a 9-year-old girl with progressive sensorineural hearing loss.



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Conclusions

- Follow up on every newborn hearing screen that refers with a diagnostic ABR
- Have a low threshold to get a diagnostic audiogram
- Identifying and managing a permanent hearing loss in a child **early** is life changing
- Cochlear implantation is a safe, reliable and a standard option
- We should set high expectations for these kids

What YOU say and do changes these kids' lives



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Support Children with Hearing Loss!

Join us in supporting our chosen Children's Champion, the Hearing Program and **ALL** children with hearing loss!

- **When:** Sunday, October 21, 2018
- Fun for kids and adults!
- <http://nchf.convio.net/goto/HearingProgram2018>

