Blindness separates us from things but deafness separates us from people.

~Helen Keller

the evaluation and treatment of Hearing Loss

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Clinical Situations

- 45 YO female who has noted hearing loss in the right ear for 2 to 3 years now. Initially this was mild but it seems to have progressed. She does note occasional ringing in the right ear only. There has been no drainage or dizziness
- 6 YO male brought to your office because he failed a hearing screening at school. Parents state that he seems to have selective hearing at home. He had one or two ear infections when he was younger but never required tubes or surgery. He has not had ear pain or fevers in the past 6 months.
- 50 YO male on vacation in Florida 1 week ago. He was drinking from a fountain when he noted the hearing in his right ear was severely diminished. He now hears a loud roaring in his ear. No pain, fever or dizziness.
- 30 YO female who has noted bilateral hearing loss which seems to fluctuate. This has progressively become worse in the last 2 years. She is now having difficulty using the phone at work. There has been no pain, drainage or dizziness

Anatomy of Hearing Eustachian tube Cochlea Eardrum Middle ear © Mayo Foundation for Medical Education and Research. All rights reserved.

Central Processing



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Weber and Rinne





Middle Ear



- Congenital
 - Atresia, Ossicular Fixation, Syndromic: Treacher Collins
- Infectious
 - AOM, OME, COM
- Tumor
 - Cholesteatoma, Otosclerosis, Squamous Cell
- Trauma
 - TM Perforation, Ossicular Disruption
- Endocrine
- Neurologic
- Systemic
 - OI, Pagets

Inner Ear

- Congenitial
 - Dysplasia, Aplasia, Familial
- Infectious
 - TORCH, Syphilis
 - Meningitis
 - Viral Cochleitis
- Tumor
 - Acoustic Neuroma, Squamous Cell CA
- Trauma
 - Noise induced, Acoustic Trauma, Barotrauma
- Endocrine
 - Diabetes, Hyper / Hypothyroid, Anemia,
- Neurologic
 - Presbycusis, MS, Arnold-Chiari
- Systemic
 - Autoimmune IED, Cogan Syndrome, Wegeners, Ototoxic Medications



50 YO male on vacation in Florida 1 week ago. He was drinking from a fountain when he noted the hearing in his right ear was severely diminished. He now hears a loud roaring in his ear. No pain, fever or dizziness.

- Weber: Lateralizes to the Left
- Rinne: Air > Bone Bilaterally but right is barely perceptible

Sudden Sensorineural Hearing Loss

• Presentation

 - 50 YO male on vacation in Florida 1 week ago. He was drinking from a fountain when he noted the hearing in his right ear was severely diminished. He now hears a loud roaring in his ear. No pain, fever or dizziness.

Incidence

- 5 to 20 per 100,000 per year
- Etiology
 - Idiopathic
 - Multiple theories, Viral is most prevalent
- Diagnosis
 - Complete audiometric evaluation
 - MRI
 - Laboratory; CBC, Sed Rate, FTA, TSH, Lipid profile

Sudden Sensorineural Hearing Loss

Treatment

Prednisone 1mg/Kg

- Wilson 1980 treated 119 pt with in 10 days of onset
- Prospective, double blind, multi center
- Subjects with moderate loss showed resolution
 - 78% treatment group
 - 38% placebo
- Antivirals
 - No benefit as mono therapy
- Intra Tympanic Steroids

Sudden Sensorineural Hearing Loss

- Results
 - Predictors of Poor Prognosis
 - Profound loss
 - Down sloping loss
 - Associated vetigo
 - Elevated ESR
 - DELAY IN INITIATION OF TREATMENT
 - 50% Resolve spontaneously
 - Treatment can improve resolution to 70%

45 YO female who has noted hearing loss in the right ear for 2 to 3 years now. Initially this was mild but it seems to have progressed. She does note occasional ringing in the right ear only. There has been no drainage or dizziness

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Inner Ear

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Single Sided Deafness

- Examples of SSD
 - Acoustic Neuroma
 - 45 YO female who has noted hearing loss in the right ear for 2 to 3 years now. Initially this was mild but it seems to have progressed. She does note occasional ringing in the right ear only. There has been no drainage or dizziness
 - Sudden Sensorineural Hearing Loss
 - Trauma
 - Meniere's Disease

Hearing Rehabilitation

BAHA Indications

- Single sided deafness
- Unilateral conductive hearing loss
- Bilateral conductive hearing loss
- Mixed hearing loss

The BAHA System for SSD

Conductive Hearing Loss

- Examples of CHL
 - Congenital Anomalies
 - Atresia
 - Otosclerosis
 - COM after surgery
 - Trauma

The BAHA System Mixed and Conductive Hearing Loss

The BAHA System

- The BAHA system is composed of three parts:
 - a detachable sound processor
 - a titanium implant
 - an external abutment

Post Operative

6 YO male brought to your office because he failed a hearing screening at school. Parents state that he seems to have selective hearing at home. He had one or two ear infections when he was younger but never required tubes or surgery. He has not had ear pain or fevers in the past 6 months.

- Weber: Lateralizes to the Left
- Rinne: Left Bone > Air

Right - Air > Bone Bilaterally

30 YO female who has noted bilateral hearing loss which seems to fluctuate. This has progressively become worse in the last 2 years. She is now having difficulty using the phone at work. There has been no pain, drainage or dizziness

Cochlear Implants

Cochlear Implants

History

- 1957 Electrical stimulation *Djourno & Eyries*
- 1972 First single-channel implant *William House*
- 1984 Release of multi-channel implants
- 1985 Nucleus 22 gains FDA approval for adults
- 1990 Nucleus 22 gets FDA approval for children
- 1991 Clarion Multi-Strategy implant released
 - 1996 FDA approval for adults
 - 1997 FDA approval for children
- Bilateral implants
- Hybrid implants

Prevalence

Prelingual profound SNHL 1 – 4 per 1000 births

- 40% genetic
 - 90% will have 2 hearing parents
 - 97% will have 1 hearing parent
- 30% environmental
- 9% meningitis
- By age 75
 - 360 per 1000 adults have profound SNHL

Candidacy Criteria Adults

- Moderate hearing loss in low frequencies
- Profound hearing loss
 - >70 dB mid to high frequencies
 - < 40% correct in HINT
- Little or no benefit from hearing aids
 - < 60% correct in best aided listening condition
 - < 50% correct in ear to be aided
- No medical or radiologic contraindication
- Motivated patient

Candidacy Criteria Children

- Greater than 12 months old
- Profound hearing loss >90 dB SRT / PTA
- <20% correct in HINT or similar
 - LNT/ MLNT \leq 30%
- Little or no benefit from hearing aids
 - Lack of progress in auditory skills
 - Parental responses to questionnaires MAIS
- No medical or radiological contraindication
- Placement in educational setting for concentrated auditory skills development
- Motivated family

Changing Criteria for Implantation

	1985	1990	1998	2000
			(Nucleus 24)	(Nucleus 24 Contour)
AGE at Implantation	Adults (18 yrs)	Adults & Children (2 yrs)	Adults & Children (18 mo)	Adults & Children (12 mo)
ONSET of	Postlinguistic	Postlinguistic	Adults & Children	Adults & Children
Hearing Loss		Adults Pre & Postlinguistic Children	Pre & Postlinguistic	Pre & Postlinguistic
DEGREE	Profound	Profound	Severe-Profound	Severe-Profound
of SNHL			Profound Children	– 2 yrs & older Profound Children – younger than 2 yrs
ADULT Speech Scores (open-set)	0%	0%	40% or less (CID)	 <u>< 50% (HINT) in ear to</u> be implanted with <u><</u> 60% in the best-aided condition
CHILD Speech Scores	Not candidates	0% open-set	Less than 20%	Lack of auditory progress (MAIS, <u><</u> 30% (MLNT/LNT) (depending on age)

Cochlear Implant Evaluation

- Audiologic Evaluation
- CT Scan
- Meningitis Risk
- Realistic Expectations
- Commitment
- Manual Dexterity

Predictors of Poor Performance

- Age at onset of deafness
- Prolonged duration of deafness
- Severe cochlear dysmorphia
- Associated mental handicaps
- Lack of auditory based intervention
- Diminished spiral ganglion cell count

Surgical Planning

Incisions

Mastoidectomy

Right Ear

Cochleostomy

Contour Design

Occupies $< \frac{1}{2}$ cross section of scala tympani

Securing the Implant

Results

 Prelingually deafened children and Postlingually deafened adults

Duration of Deafness

- Friedland 2003
- 58 postlingually deafened adults

Results

- Earlier implantation appears to provide better results
- Open Set Prelingual Deafness

Clark 1999

Age at Implant

- Wooi Teoh 2004
- Prelingually deafened children
- PB-K test scores
 - Open set word recognition
- Significant differences after age 5
- Only minimal results after age 8

Adult Implants

- Chatelin 2004
 - 65 adults >70
 - 101 adults < 70</p>
- Significant differences in performance
- Reasons for difference
 - Age related loss of spiral ganglion cell
 - "Central Presbyacusis"
 - General cognitive deficits impede rehabilitation

Non Hearing Benefits

