Surgical Management of 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

Impact of Unidentified Hearing Loss on Children

- Significant effects on language, speech, academic, and social-emotional development
  - Carney AE, Moeller MP, 1998
- Less educational achievement, limited choices for higher education, decreased vocational options
  - Holden-Pitt and Diaz, 1998

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
  - More likely to repeat a grade (24-35% vs 3%)

Management of Hearing Loss

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

At the end of this session, participants will be able to:
- feel more familiar with surgical ear anatomy
- identify at least two surgical interventions for conductive hearing loss
- identify at least two surgical interventions for sensorineural hearing loss

“Ear” = Temporal Bone

- External Ear
- External Auditory Canal
- Middle Ear
- Otic Capsule Structures
- Internal Auditory Canal
- Intracranial

External Ear

- Pinna
  - Cosmesis
  - Sound localization
  - Focuses or “funnels” sound
    - ~20dB gain
  - Meatus is air/sound entrance to rest of ear

External Auditory Canal

- Cartilaginous (lateral) and Bony (medial)
  - About ½ each
  - No cerumen at bony portion
  - Anterior: TMJ
  - Posterior: Mastoid

The Tympanic Membrane

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

The TM - Orientation

- 4 Quadrants
- Clock face
- Malleus lateral process “knee”
- Anterior canal wall bulge
TM – Pulling back the curtain

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

Middle Ear Contents

• Ossicles
• Muscles
  – Stapedius
  – Tensor Tympani
• Nerves
  – Facial Nerve
  – Chorda Tympani
  – CN IX (Jacobson’s)
  – CN X (Arnold’s)
• Cochlea
  – Promontory
  – Round Window
  – Oval Window

Middle Ear

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

Middle Ear - Pathways

• Aditus ad Antrum (Mastoid)
• Oval Window (Scala Vestibuli)
• Round Window (Scala Tympani)
• Eustachian Tube orifice (Nasopharynx)

Otic Capsule Structures = “Inner Ear”

› Cochlea
  • 2 ½ turns
  • Scala Vestibuli and Scala Tympani
  • Modiolus
  • Cochlear nerve (CN VIII)
› Vestibule
  • Saccule
  • Utricle
  • Inferior/Superior vestibular Nerves (CN VIII)
› Semicircular Canals

Bony and Membranous Labyrinth

• Perilymph and Endolymph (battery)
### Classifying Hearing Loss

- **Type:**
  - Conductive Hearing Loss
  - Sensorineural Hearing Loss
  - Mixed Hearing Loss

- **Unilateral/Bilateral**

- **Severity**
  - Mild, Moderate, Severe, Profound

### Conductive Hearing Loss

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

- 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
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

Abnormal EAC – Aural Atresia

Eustachian Tube

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

Conductive Hearing Loss

- 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

Tympanostomy Tubes – Procedure

- 10-15 minutes
  - Clean wax, incision into ear drum, suction out fluid, place tube, put in drops
- General anesthesia (mask)
- Outpatient
ETD and **Chronic Otitis Media**

**Formation of Cholesteatoma (Acquired)**

*FIGURE 7*
Mechanisms of acquired cholesteatoma formations: A, normal tympanic membrane; B, cholesterol granuloma; C, deep retraction with collection of keratin debris; D, infected cholesteatoma with granulation in the middle ear.

**Cholesteatoma**

**Cholesteatoma**

**Other Whitish Things**

**Cholesteatoma**

*Acute Otitis Media*  *Myringosclerosis*  *Myringosclerosis*
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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Goals of Surgery - Cholesteatoma

1. Eradicate Cholesteatoma
   - Make ear “safe” – prevent complications
2. Hearing Reconstruction
   - Ossicular Chain Reconstruction

Can’t always do both at same time...

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Tympanoplasty, Mastoidectomy, Ossicular Chain Reconstruction

- “Tympanoplasty”
  - Surgery on the TM or middle ear structures
- “Mastoidectomy”
  - Drilling out mastoid air cells, to clear disease or provide improved access to middle ear
- “OCR” – Ossicular Chain Reconstruction
  - Restoration of function of ossicular chain with or without prostheses

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Transcanal – Congenital Cholesteatoma
Mastoidectomy

Surgical Anatomy
Surgical Options – Restoring Hearing

- Total Ossicular Reconstruction Prosthesis (TORP)
  - Titanium, Hydroxyapatite, Fluoroplastic
  - TM to stapes footplate
- Partial Ossicular Reconstruction Prosthesis (PORP)
  - Some ossicles intact
- No reconstruction

Ossicular Reconstruction

Photos courtesy of wilsonear.com
Hearing Outcomes at 1 year

- Intact Ossicular Chain:
  - >90-95% <20 dB
- Canal Wall Up (CWU):
  - 60%-90% <20dB
  - 85%-98% <30dB
- Canal Wall Down (CWD):
  - 45%-80% <20dB
  - 75%-95% <30dB

Bone Anchored Auditory Implant – Conductive Hearing Loss

- Indications include:
  - congenital aural atresia
  - chronic suppurative otitis media
  - chronic otitis externa
  - Ossicular dislocation/trauma
  - Conductive losses not manageable surgically
  - Inability to tolerate traditional hearing aids
- Cannot have bone thresholds (SNHL) > 65dB

Bone Anchored Auditory Implant – Conductive Hearing Loss

- OAV/Goldenhaar
- Treacher Collins

Bone Anchored Auditory Implant

- Sound processor
- Osseointegrated implant
- Abutment
Bone Anchored Auditory Implant

- FDA approved for 5 years old and older
  - Skull thickness
  - Bone conduction head band until skull thick enough

Bone Anchored Auditory Implant

- Approximately 30dB gain over unaided thresholds
- Can close air bone gap well
  - 80% to within 10 dB air bone gap
  - 60% to within 5 dB air bone gap
  (Lustig et al 2001)
- Possibly better speech discrimination compared with conventional HA
- Thresholds comparable to (some ?better) to conventional HA

New Transcutaneous System
(FDA approved December 2013)
Sensorineural Hearing Loss

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

Sensorineural Hearing Loss

- Congenital
  - Genetic
  - Environmental/Neonatal exposures
- Inner ear malformations
- Noise Induced
- Labyrinthitis
- CNS pathology/masses
- Ototoxicity

Surgery for Sensorineural Hearing Loss

- **UNILATERAL** profound = Single Sided Deafness
  - Bone Anchored Auditory Implant
  - Other Bone Conduction Devices (Soundbite)
  - (Cochlear Implantation - future)

- **BILATERAL** severe-profound SNHL
  - Cochlear Implantation

Bone Anchored Auditory Implant – Single Sided Deafness

Cochlear Implants – Epidemiology

- As of December 2010:
  - ~ 219,000 people worldwide have CIs
  - In the US: ~42,600 adults and ~28,400 children

- ~55% of all candidates (N~12,800) for cochlear implants aged 1 to 6 years received cochlear implants in year 2000 (Bradham and Jones, 2008)

- At least 30 kids/year from NCH catchment area
Cochlear Implant – Audiologic Candidacy

FDA approval: 1 yr and older

- 1 yr to 2 yrs: BILATERAL profound hearing loss
- >2 yrs old: BILATERAL severe to profound hearing loss

- Limited auditory development with best fit hearing aids
  - IT-MAIS for younger kids
  - Open-set word recognition scores for older kids

Cochlear Implant Evaluation

- Audiologic evaluation (Audiology)
- Medical evaluation and imaging (ENT)
- Speech/Communications evaluation
- Social support evaluation (Speech)
- Hearing Aid Trial
- Financial Clearance
- Auditory therapy
- Vaccination
- Implantation
- Then, REHAB, REHAB, REHAB!!!!

The Cochlear Implant

- receiver-stimulator
- magnet
- ground electrode


Spiral Ganglia

http://bmc.erin.utoronto.ca/~andreaz/newMediaSite/

Surgery

- Cochlear Implant procedure:
Surgery

• Cochlear Implant procedure:

Cochlear Implant Outcomes

• Depends on age of implantation!
  – Critical point around 3-4 yrs old
• 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

Bilateral Cochlear Implantation

In kids:
• Improved sound localization
• Probably better speech perception and understanding in noise

Complications

• Wound infection
• Extrusion of device
• Device failure (hard or soft)
• Meningitis
• Failure to use implant
• CSF leak
• Vertigo/Disequilibrium
• Facial nerve paralysis
• Malpositioned implant

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 surgery
• Influenza vaccine to reduce otitis media

Critical, and REQUIRED step for implantation
**Current Frontier of CI**

- Bimodal habilitation (one ear HA, one ear CI)
- Younger implantation
- Severe to Profound criteria for youngest kids
- Auditory Neuropathy
- **Unilateral** Profound Sensorineural Hearing Loss
- Complex medical and developmental comorbidities
- Hearing preservation surgery and “electroacoustic stimulation” (CI and HA in same ear)

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