Controversies and Trends in Pediatric Cochlear Implantation

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Disclaimer

- I have no financial interest or partnership with any company that manufactures cochlear implants
- I use Cochlear Corporation and Advanced Bionics devices
  - No good evidence exists that one implant is superior to another
Overview

- Recurrent otitis / otitis media with effusion
- Anatomic abnormalities
- Unilateral vs bilateral
- Age at implantation (< 12 months)
- Implanting better or poorer hearing ear
- Drilling a well vs no well
- Location of cochleostomy
Acute Otitis Media and CI

- Acute otitis media is obviously common in children, but CI should not be delayed
  - Long term speech and language developmental delays

- Some evidence that the cortical mastoidectomy performed at implantation decreases incidence of AOM (Luntz 2004)
  - 34 AOM prone children, no complications
  - 4 yr follow up
Acute Otitis Media and CI

- When AOM occurs post-implant, it must be treated aggressively
- Of utmost importance to pack cochleostomy tightly in these children to decrease risk of meningitis (Nadol 2004)
- Vaccination is critical (Hib, Prevnar < 2 y/o, Prevnar / Pneumovax 2-5 y/o, Pneumovax > 5 y/o)
Chronic Otitis Media with Effusion

- More controversial than AOM – no good evidence which approach is superior

- Options / Preferences (Kennedy 2005):
  - Ignore OME and proceed with CI (22%)
  - Place vent tube and perform CI at 2\textsuperscript{nd} operation (53%)
  - Vent tube and CI at one operation (6%)
  - CI with “extra maneuvers” – i.e. antibiotic irrigation/gelfoam (12%)
  - Avoid CI altogether (6%)
Chronic Otitis Media with Effusion

- **What if a vent tube is in place already?**
  - Proceed with CI with tube in place (38%)
  - Remove tube, let drum heal, CI at 2\textsuperscript{nd} surgery (43%)
  - Remove tube at CI and patch drum (18%)

  *Kennedy 2005*

- **Must weigh risk of noninfected effusion vs creating “nonsterile” middle ear with vent tube by exposure to external environment**

- **Role of adenoidectomy?**
  - Known to improve OME in older children (>3-4 y/o), but not in younger children (Casselbrant 2009)
  - May delay CI surgery
Inner Ear Malformations

- It is generally accepted that some congenital inner ear malformations are NOT contraindications for CI
  - Mondini deformity
  - Enlarged vestibular aqueduct
  - SCC hypoplasia / aplasia
  - Cochlear hypoplasia
Inner Ear Malformations

- Medical / Surgical considerations:
  - Higher risk of CSF leaks (21%)
  - Aberrant facial nerve (32%)
  - May have incomplete insertion
  - Consider straight array in common cavity / severe Mondini as neural tissue likely lies on the walls of the cavity
  - Higher risk of electrode insertion into IAC (consider intraop skull film)
  - Slower to develop speech perception, but typically achieve mean performance levels

Buchman 2004, Graham 2000
IAC Insertion
Preop Evaluation: CT or MRI?

- **CT traditionally study of choice:**
  - Better definition of bony anatomy
    - Superior to MRI in identifying vestibulocochlear / modiolar abnormalities (Parry 2005, Trimble 2007)
  - Less need for sedation
  - Lower cost
  - Most ENT more comfortable with CT, particularly with surgical anatomy
    - Marrowed mastoid bone
    - Facial recess pneumatization
    - Course of facial nerve
    - Tegmen

Woolley 1997, Trimble 2007
Preop Evaluation: CT or MRI?

- **Limitations of CT**
  - Cannot identify cochlear n. aplasia
    - Narrow IAC does not correlate 100% (Trimble 2007) – 8/92 patients had narrow IAC on CT, all were shown to have cochlear nerves on MRI
    - Reverse is also true – absence / attenuation of the cochlear nerve has been shown in normal caliber IAC’s
  - Radiation exposure
Preop Evaluation: CT or MRI?

- **Advantages of MRI:**
  - Assess caliber of cochlear nerve
  - Argument can be made that most relevant surgical anatomy can be obtained from MRI (obviating need for second study in most cases)
  - “Abnormalities on MRI are more likely to influence the implant process” (Parry 2005)
Narrow IAC

- Historically considered an absolute contraindication to CI (absent or attenuated cochlear n.) when seen on CT

- MRI (FIESTA imaging) can reveal an intact nerve even in the presence of a narrow IAC
  - These patients can be considered implant candidates
  - Kim 2006 – 6/6 patients showed increased awareness / discrimination of environmental sounds and pure tone thresholds at 30 dB or better
    - Only 2/6 achieved open set word recognition
Narrow IAC

- **Other considerations – functional hearing**
  - Presence of functional hearing prior to implantation may improve outcomes
    - 2/2 children with PTA < 100 achieved open set word recognition (Kim 2006)
  - Question moving forward:
    - Who has a better prognosis:
      - Better functional hearing with absent/attenuated nerve
      - Worse functional hearing with intact cochlear nerve
Labyrinthitis Ossificans

- Critical to identify post-meningitic profound deafness early
  - Ossificans can begin as early as 12 days (Philippon 2009)
  - Implant early and bilaterally - LO associated with worse hearing result post implant

- MRI superior in assessing cochlear obstruction
  - CT only 50% sensitive (Young 2000)
  - MRI 94% sensitive (Isaacson 2009)
    - Can identify early fibrous ossificans
    - Will help in preoperative planning
Case - JR

- 4 y/o male - 6 weeks s/p pneumococcal meningitis from an AOM
- Bilateral profound sensorineural hearing loss
- CT shows normal cochlea
Case - JR

- MRI reveals decreased signal intensity in the right cochlea, early ossificans
- Bilateral implantation is planned for 1 week later
- MRI information allows preoperative planning:
  - Address left ear first – “easier” ear
  - Be prepared for right ear – possible failed insertion and discussion with parents
  - Be prepared for possible scala vestibuli insertion
  - Have straight array available and plan on no “advance off stylet” technique if usual electrode can be placed
  - Have split electrode available if drillout is necessary (Bredberg 1997)
Case - JR
Which Ear to Implant?

- Long standing and continuing argument about whether to implant better or worse hearing ear
- Favorable vs unfavorable anatomy also considered
- Traditionally, the better hearing ear was chosen:
  - Pathophysiologically based on correlation between level of residual hearing and spiral ganglion cell counts (Incesulu 1998)
Which Ear to Implant?

- Preoperative residual hearing is associated with superior speech perception (Rubinstein 1999, Cowan 1997) BUT:
  - This may have a central auditory pathway origin and not be related to the implant ear (Francis 2004)

- Chen 2001
  - 38 matched patients with B/L profound SNHL
    - 19 had better hearing ear implanted, 19 had worse hearing ear implanted
      - No difference in open set word or sentence recognition
Which Ear to Implant?

- **Francis 2004**
  - 3 groups: B/L severe, B/L profound, severe/profound (one ear each)
    - Worse hearing ear implanted
      - No difference in speech perception b/t B/L severe and severe/profound groups
      - 3 severe/profound patients chose to have the better hearing ear implanted – these were matched against 3 who had the worse hearing ear implanted
        » No difference in open set word rec
    - B/L profound SNHL did worse overall, but no difference b/t better/worse ear
    - Suggests that preoperative residual hearing is an important predictor of success, but NOT related to which ear is implanted
Which Ear to Implant?

- Implanting the worse hearing ear may allow for continued use of a hearing aid in the other ear
  - Acoustic stimulation + implant broadens auditory inputs to the brain, allowing improved hearing (Ching 2004, Gifford 2007)
  - But it may be difficult or impossible for some patients to fuse the electrical (CI) and acoustic (aid) signals centrally (Tyler 2002)
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- Implant the worse hearing ear, particularly if the patient is having success with a hearing aid on the contralateral ear
  - Good results with adaptation to electrical + acoustic stimulation may be due to plasticity of children
- In bilateral symmetric profound SNHL, particularly in infants, strongly consider bilateral implantation
  - If unilateral, consider ideal anatomic ear
  - If anatomy is not a concern, implant right ear
    - Left is dominant hemisphere for speech processing in 98% of right handed individuals and most left handers as well
    - No strong data supports improved outcomes by implanting one side over the other in symmetric profound ears
Age at Implantation

- 1990 – FDA approves CI in children > 2 y/o
- 1998 – FDA drops age to 18 months
- 2002 – FDA drops age to 12 months
- Traditionally, most implants performed less than age 12 months in the US have been in post-meningitic patients
< 12 Months: The Future?

- Earlier age being considered because:
  - Universal newborn hearing screen diagnosing most children prior to 3 months of age
  - Hearing aid data suggests optimal timing for amplification is 6 months, so why not CI if there is no benefit with aids?
  - Technically feasible as cochlea reaches adult size by birth
< 12 Months: The Future?

- **Challenges:**
  - Thinner skin flap / subcutaneous tissue
  - Smaller blood volume (80 ml/kg in children <12 mos)
    - Hypovolemic shock can occur with 10% total blood volume loss
  - Underdeveloped mastoid tip (higher risk to CN VII)
  - ?Role of skull growth related to implant migration
    - Consider well + fixation sutures
    - Roland 2009 – 50 implants < 12 months, no migration seen (mean f/u 6.8 yrs)
< 12 months: The Future?

- Roland 2009 – 50 patients, 6.8 yr mean f/u
  - No anesthesia complications
  - Total of 8 complications:
    - 3 major:
      - CSF leak
      - Implant failure (9 months post op)
      - Infection / exposed implant (10 months post op)
    - 5 minor:
      - Hematoma, skin flap erythema, cellulitis
  - Conclusion: Safe and efficacious in children <12 months
< 12 months: The Future?

- **Colletti 2005**
  - 10 children
    - Increased CAP scores compared to infants implanted after 1 y/o
      - No difference until 6 months after implant
      - Approached normal for age by 12 months post implant
    - No complications
  - Conclusion: Encouraging preliminary results that implantation < 12 months has significant auditory/speech benefit compared to those > 12 months
Colletti 2009
- 4-9 year follow up to previous study
- Compared with those implanted 12-23 mos and 24-36 mos
- Children implanted early reached normal CAP more quickly and performed better on other receptive/speech tests
- No early implants in schools for the deaf
  - 30% implanted 12-23 mos
  - 60% implanted 24-36 mos
<12 months: The Future?

- Vlastarakos 2010
  - Meta-analysis
  - Compared 10 patients who had open/closed set testing with 10 controls implanted b/t 12-23 mos
    - Only 4 had better outcomes
  - Conclusion: Possible benefit but data is too limited, short term and subjective to draw a firm conclusion
Well – To Drill or not to Drill?

- **Potential advantages:**
  - Prevents implant migration
  - Reduces device profile

- **Infant considerations:**
  - Thin skull often requires drilling to dura
Well – To Drill or not to Drill?

- **Potential complications:**
  - CSF leak
  - Subdural hematoma
  - Epidural hematoma
  - Lateral sinus thrombosis

- **Balkany 2009**
  - 227 implants
    - 56 with well, 171 without well
    - No cases of migration (minimum follow up 12 months)
The CHP Experience

- Small periosteal pocket
- Fixation sutures VS small well / ledge
  - Lower profile Cochlear Corp device design
  - No cases of migration
  - One case of screw extrusion into subQ tissue that required repeat surgery
Well – To Drill or not to Drill?

- Secondary advantages of no well:
  - Smaller incision
  - Shorter operative time
Location of Cochleostomy

- Scala tympani (2) insertion is ideal
  - Associated with better speech outcome compared to scala vestibuli insertion (Aschendorff 2007)
  - Less traumatic
  - Decreased vestibular symptoms (Todt 2008)
Unilateral vs Bilateral Implantation

- Advantages of binaural hearing:
  - Hearing in Noise:
    - Binaural summation effect
    - Binaural squelch effect
    - Head shadow effect
  - Sound localization
    - Head shadow effect
Binaural Summation Effect

- **Redundancy:**
  - The auditory system can combine information derived from 2 ears versus monaural

- **Improved hearing thresholds:**
  - Increased perceptual loudness
  - 3 dB difference between binaural and monaural
  - Improves speech intelligibility in noise
  - 21% benefit binaural vs monaural

Schleich 2004, Murphy 2007
Squelch Effect

- Auditory cortex is able to use differences in signal to noise ratio between the 2 ears and identify what’s not wanted and suppress it
  - i.e. how loud the voice you’re listening to is compared to all other noise
- Improves speech perception in noise
- 3 dB difference between binaural and monaural
- Does not develop until 1 year after implantation and continues to improve for at least 4 years
  - Eapen 2009 – 9 patients with simultaneous bilateral implants
Head Shadow Effect

- Improves signal to noise ratio:
  - Acts as a baffle to shadow competing noise from signal
  - Can attenuate high frequencies up to 20 dB and low frequencies 4-7 dB
  - Immediate improvement after bilateral CI and continues to improve to at least 12 months (Nava 2009)
  - Also demonstrated with second side CI (Galvin 2007)
Sound Localization

- Head shadow effect also contributes in localizing sound
  - Interaural difference

- Safety issue:
  - Crossing roads
  - Bicycles
  - Particularly important in children – more prone to dangerous behaviors
Other Benefits of Bilateral Implants

- Ensures that the ear with the best postoperative performance is implanted
- If different speech cues are encoded by the two ears, they act in a complimentary way to improve performance
- Back up device (dead battery, hardware malfunction)
Concerns with Bilateral Implantation

- Precludes usage of future, “better” technology for hearing amplification
  - Hair cell regeneration
  - Stem cells
  - Drug delivery systems
  - Better implant device

- Longer procedure, ?increased meningitis risk
Concerns with Bilateral Implantation

- **Potential risk of vestibulopathy**
  - Clinical and vestibular findings often do not correlate, particularly in children (Basura 2009)
  - Make patient aware of potential risk
  - Scala tympani insertion important (Todt 2008)

- **Cost**
  - $25,000-$50,000
  - Favorable cost-utility and cost-differential when quality of life measures are included
Auditory Plasticity – Critical Time?

- **Sharma 2002, 2005**
  - Measured cortical auditory evoked potentials in implanted children compared with normal controls
    - EEG activity in response to sound stimulation
    - P1 wave latency tracks maturation of auditory thalamic and cortical sources
    - Latencies improved (decreased) after implantation
Auditory Plasticity – Critical Time?

- Key time appears to be 3.5 years

- Children implanted after 7 years of age never achieve normal P1 latency
  - Poorer speech and language outcomes
Hybrid Implant

- FDA approved 2014 ages 18+
- Thinner, smaller electrode designed to maintain low frequency acoustic hearing
- Combines acoustic and electrical amplification/stimulation
Single Sided Deafness

- **Potential advantages**
  - Improved sound localization, quality of life and reduced tinnitus

- **Questions:**
  - Cost! – especially vs expected benefit
  - Length of duration of profound HL
    - Auditory plasticity limitation
  - Benefit vs other options (i.e. BAHA, CROS)
  - Congenital hearing loss
  - Insurance coverage
Conclusions

- Cochlear implants remain the treatment of choice for children with severe to profound SNHL
- Ideal surgical approach in OME still not defined
- Anatomic malformations, particularly narrow IAC, becoming less of a contraindication to CI
- Lower profile implant devices obviating need to drill a well, even in infants
- Scala tympani insertion improves outcomes
- Trend away from always implanting better hearing ear
- Changing indications: many centers routinely implanting at less than 12 months of age
- Bilateral implants are the binaural option in bilateral severe to profound SNHL
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