Update on Pediatric Cochlear Implantation

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Outline

- How does a cochlear implant work?
- Current indications for Cochlear Implantation at BCCH.
- Current indications for Bilateral Cochlear Implantation.
- Evolution of our Cochlear Implant Program
- Surgical Procedure.
- Is bilateral implantation better than unilateral?
Etiology of CI patients at BCCH

- Unknown: 28%
- Genetic non Sd.: 23%
- Genetic Sd.: 11%
- Meningitis: 10%
- Prematurity: 7%
- Inner ear malformation: 9%
- Ototoxic: 3%
- AN/AD: 3%
- Mitocondrial Disease: 1%
- ECMO + CDH: 2%
- CDH: 1%
- CMV: 1%
- Rubella: 1%
- Genetic Sd.: 11%
- Genetic non Sd.: 23%
- Unknown: 28%
Investigations of congenital sensorineural hearing loss

Audiological testing
- Auditory brainstem response (ABR)
- Pure tone audiogram
- Tympanometry
- Vestibular function tests.

Blood tests
- Genetic analysis: DNA sampling of patient for Cx 26 testing.
- Viral serology (rubella, toxoplasmosis, cytomegalovirus, syphilis)
- Thyroxine and thyroid-stimulating hormone (not routine)

Radiological investigations
- Computed tomography of the skull and petrous temporal bone.
- Magnetic resonance imaging of the internal auditory meatus (cochlear nerve)

Cardiological assessment
- Electrocardiogram

Ophthalmological assessment
- Fundal examination

Lab assessment
- Urinanalysis and renal ultrasound. (Not always necessary just if Risk Factors associated).
How does a cochlear implant work?

• Bypassing damaged portions of the inner ear and directly stimulates the auditory nerve.

• Hearing through a cochlear implant is different from normal hearing and takes time to learn or relearn.
Cochlear Implant Components

- A microphone, which picks up sound from the environment.

- A speech processor, which selects and arranges sounds picked up by the microphone.
Cochlear Implant Components

- A transmitter and receiver/stimulator, which receive signals from the speech processor and convert them into electric impulses.
- More recent designs will improve the external profile.
An electrode array, collects the impulses from the stimulator and sends them to different regions of the auditory nerve. Tonotopically arranged.
Pediatric Criteria

- 12 months to 18 years of age plus off label
- Bilateral severe to profound sensorineural hearing impairment (exception: 12 month old bilateral profound sensorineural HL)
- Trial with consistent binaural amplification (3-6 months)
- Receive little or no benefit from hearing aids
- No medical contraindications
- Inner ear capable of accommodating the internal device
- Lack of progress in auditory development
- Realistic expectations
- Enrolment in a habilitation/education program that involves a strong auditory-oral component
- A comprehensive ongoing support system
Evolution of our Cochlear Implant Program
CI Implanted by year and Cumulative Total.

- **CI/year**
  - Year: 88-89, 89-90, 90-91, 91-92, 92-93, 93-94, 94-95, 95-96, 96-97, 97-98, 98-99, 99-00, 00-01, 01-02, 02-03, 03-04, 04-05, 05-06, 06-07, 07-08, 08-09
  - Number of Patients: 3, 2, 1, 1, 0, 1, 7, 4, 2, 2, 4, 10, 10, 11, 18, 19, 25, 20, 19, 21

- **Cumulative**
  - Cumulative Total: 3.0, 5.0, 6.0, 7.0, 7.0, 8.0, 15.0, 19.0, 21.0, 23.0, 27.0, 37.0, 47.0, 57.0, 68.0, 86.0, 105.0, 130.0, 150.0, 169.0, 190.0
Annual Cochlear Implant Performed by Age / Fiscal year
Children with difficulties in addition to their deafness, such as global developmental delay, cognitive impaired, autism, are not routinely excluded in our program;

Out of the 190 patients implanted 31 (16%) have cognitive impairment.

We classified them as mild (6.8%), moderate (6.8%) or severe (2.6%) cognitive impaired according to psychological, neurological, developmental pediatrician or primary pediatrician evaluation.
Implanting the Abnormal Cochlea
Mondini: Absent Interscalar septum + LVA

>2mm: LVA?
Common cavity
Vestibular hypoplasia

Vestibular displasia

VIII nerve agenesis

Facial nerve
Results

CI Pts with Inner Ear Abnormalities

<table>
<thead>
<tr>
<th></th>
<th>Pre - R</th>
<th>Pre - L</th>
<th>Post SAT CI</th>
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</thead>
<tbody>
<tr>
<td>Aided</td>
<td>69.38</td>
<td>70.63</td>
<td>23.33</td>
</tr>
<tr>
<td>Unaided</td>
<td>92.50</td>
<td>92.50</td>
<td>23.46</td>
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Review of Potential Complications With the Patient and Family

- Bleeding
- Infection
- VII Nerve
- Device Extrusion
- Meningitis
- Chorda Tympani Nerve
- Others
SURGICAL
PROCEDURE
Review of Potential Complications With the Patient and Family

- Bleeding
- Infection
- VII Nerve
- Device Extrusion
- Meningitis
- Chorda Tympani Nerve
- Others
PREPARATION
OR SET UP
FACIAL NERVE MONITORING
MARKING THE TEMPLATE, INCISION AND LOCAL ANESTHESIA
“LAZY S” INCISION
SKIN AND SUBCUTANEOUS LAYERS
EXPOSURE
MASTOIDECTOMY
Inspection of the Implant
INSERTION
AUDIOLOGIST IS READY
CHECKING OUT THE CHANNELS
Head X-ray post-op
Postoperative complications

- **AOM:** Less frequent than non implanted patients
- **Facial stimulation**
- **Tinnitus:** 0.05%
- **Vertigo:** 0.08%-39%
- **Device failure:**
  - Malfunction
  - Lack of VIII CN function
  - Damage to electrode
  - Extracochlear placement
- **Head trauma:**
Postoperative complications

In our review the complications were divided as:

- Surgical complications
- Device complications

The surgical complications were considered as:

- Perioperative: for those occurring and up to 24 hours after surgery (6%)
- Early onset (within 2 weeks post-surgery) (5%)
- Late onset (greater than 2 weeks post surgery) (13%)

- A major surgical complication was defined as an adverse event occurring during or after surgery that necessitated a major surgical intervention or a permanent disability

- Minor complications were defined as those managed by medical treatment or by a minor surgical procedure (Bhatia, 2004).
Early Onset Complications (< 2 weeks)

- Facial Palsy:
  - Minor: 2
  - Major: 1
- Wound Acute Infection:
  - Minor: 5
  - Major: 0

Perioperative Complications (24 h)

- Posterior wall canal defect:
  - Minor: 1
  - Major: 0
- TM perforation:
  - Minor: 1
  - Major: 0
- Partial insertion:
  - Minor: 1
  - Major: 3
- Major Bleeding:
  - Minor: 1
  - Major: 0
Late onset Complications (> 2 weeks)

- Minor skin complic.
- Hematoma
- Tinnitus - Vertigo
- Hematoma
- Magnet disloc.
- Cholesteatoma
- Osteomyelitis

Number of Patients

- Minor
- Major
Meningitis following CI

- 4264 children implanted before age 6
- 0.6%
- 18 patients (Nucleus) from ’82 to ’02
- Mostly non device-related
  - Positioner (Clarion device)
- Most of the cases: Mondini+LVA
- Risk factors
  - TB fracture
  - Frontal sinus defect
  - Preoperative episode
  - VP shunt

Meningitis following CI

- *Pneumococcus* more likely to affect <2yo
- *N. meningitides*: 2-60yo → 60% Not reported on implanted patients
- *Staph aureus*: More common after neurosurgery procedures
- *H. influenzae*: Has ↓ since Hib
- 118 cases reported to FDA as of May ‘03

- In 50% of the cases, cochleostomy was not sealed, whilst in 6% it was grafted
Prevention of meningitis

- Vaccination up to date before and after CI
- Meticulous asepsis
- Cochleostomy packing
- Aggressive management AOM
- Avoid or postpone Sx if SOM
- VT 8 weeks before implant
- No VT during first 6 weeks postop
- If TM perforation or COM: 1 year symptoms-free
- “Prepare” chronic ear for implant if indicated
  - CWD, obliteratorive procedures, closure of EAC
We are all responsible

• Please ensure your CI patients are up to date regarding vaccination both before and after surgery
• Many do not understand the difference between the various vaccinations
Vaccination

- Immunization record as part of candidacy evaluation process.
- Complete immunizations at least 2 weeks before implant surgery.
- Minimum interval between doses 8 weeks.
Special cases

Custom-Designed MED-EL Electrode for Common Cavities

- If common cavity:
  - Do not use perimodiolar array.
  - ST electrode (Med-El)
- Congenital dysplasias
  - Mondini+LVA: Higher risk of perilymph gusher and meningitis
- MRI
- If acoustic neuroma:
  - Implantable if VIII CN was preserved.
  - ABI
- Reinsertion
X Linked Gusher -
Is Bilateral Implantation better than unilateral?
Current Indications for Bilateral Cochlear Implantation at BCCH

- **Category 1**: Bilateral Profound sensorineural hearing loss.

- **Category 2**: Post-meningitis (no more than 6 months), bilateral profound sensorineural hearing loss.

- **Category 3**: Dual sensory loss (blindness and hearing loss; can include those children who present with visual deterioration), bilateral profound sensorineural hearing loss.

- **Category 4**: Bilateral profound sensorineural hearing loss (sequential implantation) – first implant done prior to 4 years ago.
Current indications for Bilateral Cochlear Implantation at BCCH

Past history of appropriate and consistent amplification in the second ear

- Use of an auditory-oral mode of communication only.
- Acceptable cochlear anatomy in non-implanted ear
- No other conditions that would preclude the reasonable expectation of optimal binaural benefit with a second implant (e.g., lack of progress in speech-language and auditory development)
- Strong, consistent parental involvement has been demonstrated in therapy and follow-up with the first implant (checklist under development to assess this)
They must have profound hearing loss in the second ear to be implanted (i.e. Pure tone average > 90 db HL @ 500, 1000, 2000).

**Age requirements:**

1. If the hearing loss is congenital/pre-lingual the child must be under 8 years of age.

2. If the hearing loss has been sudden and profound bilaterally from previously normal hearing bilaterally then the child may be any age.

3. If the hearing loss has been acquired and progressive after 8 years of age and contralateral hearing aid use has been appropriate and consistent since initial diagnosis of hearing loss, and there is a record of amplification history that documents audibility of the long-term average speech spectrum from 500 - 2000 Hz within the past 24 months/years for the second ear, then the child may be older than 8 years of age.
Factors

• Head shadow effect (monaural processing)
  Attenuation of up to 7 dB, one ear has better SNR

• Spatial separation of signal and noise, time/ intensity cues

• Binaural summation
  signal redundancy
Factors relating to benefit?

- Relate to pre-op level of aided hearing
- Previous binaural experience
- Performance with unilateral implant prior
- Age at bilateral implantation
- Experience with bilateral implants
Bilateral Cochlear implants - Why?

Binaural Hearing Advantages:
• Localization.

• Hearing in noise

• Binaural advantage (loudness)

• Most noise reduction abilities depend on access to time, level and spectral differences of the sound source between the ears

Do these effects occur in bilateral CI users?
Bilateral implants in children

• Simultaneous

• Sequential (no delay, short delay, long delay).

• Bilateral experience necessary to develop binaural processing. Results depends on time of use.

• Bilateral hearing is compromised by long term unilateral cochlear implant use:
  - Mismatches in brainstem development.
  - Mismatches in cortical responses and speech perception in each ear.
Disadvantages of bilateral implantation

• Limitations of the technology:
  - Lack of fine structure representation - hearing in noise
  - Music appreciation
  - Sound quality

• Poor outcomes for long sequentials
• Poor representation of fine structure
• Interaural timing difficulties
Subjective Benefit

- Sound is reported as “more natural”, “fuller”, “richer”

- Less effort required to understand speech

- No longer dealing with “dead side”
Benefits

• Binaural benefit-able to attend to the ear with the better signal to noise ratio

• Ensures that the better ear has been implanted

• Binaural advantage-makes use of interaural time and intensity differences and spectral cue enhancements.
Integration of acoustic and electrical hearing

The Nucleus Hybrid cochlear implant system.

- Shorter electrode array component than a more traditional cochlear implant.

- Preservation of residual hearing: This type of combination device, or Hybrid, can result in better low frequency hearing preservation than other traditional devices with longer arrays.

(Turner, Reiss, & Gantz, 2007)
Prevention and Treatment of AOM in Children with Cochlear Implants

**AOM – Acute Otitis Media**

- No “watchful waiting” without antibiotic therapy.

- Common pathogens: Streptococcus Pneumoniae; H. Influenza, M. Catarralis. Pathogens of AOM are the same in CI pts.

- First two months after CI:
  Ceftriazone (or cefotaxime) IM,IV. Consider Miringotomy with cultures.

- 2 or more months after CI:
  Treatment with High Dose Amoxicillin or Amoxicillin Clavulanate. (90 mg/Kg/D divide q 12 hrs)
Prevention and Treatment of AOM and Meningitis in Children with Cochlear Implants

Meningitis

- CSF Exam with Culture.
- If AOM: Miringotomy and cultures
- Within first Month after implant
  - Pathogens: Pneumococcus (44%), H.Influenzae type B or non typable, other gram (-) bacillus, enterococcus.
  - Empiric treatment: Vancomycin + meropenen +/- steroids.
- One or more months after implant
  - Pathogens: Pneumococcus (80%), H.Influenzae (type B or non typable), streptococcus pyogenes (group AB), neisseria meningitides.
  - Empiric treatment: Vancomycin + cetriaxone +/- steroids.
Gracias