Cochlear Implantation in Children with Multiple Disabilities: Candidacy and Outcomes

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REFERENCES IN SEPARATE HANDOUT

Hearing: “The Connection” Factor

• Hearing allows a person to have access to warning signals, access to a continual source of information about things happening in a person’s immediate physical environment, as well as the acquisition of physical skills (e.g., balance).

• Hearing provides "connectivity" in the link that sound forms between the person and the rest of the world.

• Paraphrased from Pollack, 1970
Acknowledge our Team

- Robin Adkins
- Mary Jane Beaucage
- Marei Bonkowski
- Candy Carrier
- Joanne Guimond
- Carey Pidhayecki-Stephishen
- Bev Hansen
- Allan Ho
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- Trina Uwiera
- Michelle Wiley

Cochlear Implant Team

- Otolaryngology
- Audiology
- Speech - Language Pathology
- Education of the Hearing Impaired
- Listening and Spoken Language Specialist (AVT)
- Social Work
- Psychology
- Developmental Paediatrician

CI in Children with Multiple Disabilities

- 40% of children with hearing loss have additional disabilities
  - Cognition and oral-motor limitations have most impact on CI outcomes
- 2001: Advances in programming technology provide physiological measures that facilitate programming for children who are less able to give consistent behavioural responses
- 2002: First CI at GRH for child with severe multiple disabilities
Cochlear Implant Candidacy

- Candidacy Decision
  - Is candidate an appropriate surgical risk?
  - Will implant be audiologically effective?
  - Will candidate benefit from auditory stimulation?
  - Does candidate participate in communicative acts?
  - Does candidate exhibit needed behaviour for success?
  - Does candidate have needed emotional-social factors for success?

- Examination and investigation must indicate an acceptable level of surgical risk with a good likelihood of an acceptable outcome (medically and functionally)

- Severe to profound bilateral SNHL
- Appropriate & consistent amplification for specified trial period
- Limited/no benefit as defined by speech perception test scores
Cochlear Implant Candidacy

• Will candidate benefit from auditory stimulation?
  • Has the potential to identify the presence of auditory stimuli
  • Appropriate use of residual hearing has been demonstrated
  • Duration of deafness does not significantly limit potential for auditory system development

Cochlear Implant Candidacy

• Does candidate participate in communicative acts?
  • Communicates intentionally
  • Will initiate a communicative act
  • Has potential for symbolic communication
  • Has established joint attention
  • Will participate in verbal or non-verbal turn taking

Cochlear Implant Candidacy

• Does candidate exhibit needed behaviour for success?
  • Not a criteria for children < 3 years
  • Can sustain attention sufficiently to allow for programming of speech processor
  • Demonstrates the ability within a reasonable amount of clinician time and effort to participate in tasks required for assessment, programming and intervention
  • Behaviour allows for assimilation of device into functional daily activities
Cochlear Implant Candidacy

- Does candidate have needed emotional-social factors for success (con’t)?
  - Opportunities or auditory-oral interactions must be present throughout the day
  - Appropriate speech-language and education services are in place
  - In an education setting that consistently uses spoken language
  - If sign is used, it must support spoken language structure

Multi-disciplinary criteria enable the cochlear implant team to identify candidates who are most likely to sustain device use and achieve performance outcomes that reflect significant benefit as a consequence of having access to an auditory signal through a cochlear implant.

How do we know what they hear?
Instruments in use at GRH

- PRISE: Production Infant Scale Evaluation (Kishon- Rabin, et al., 2005)
- Little Ears (Tsiakpini et al., 2004)
- Newly available: Infant Monitor of Vocal Production (Cantle- Moore, 2010)


Instruments in use at GRH

- Rossetti Infant-Toddler Language Scale (1990, Lingui-Systems)
- Champions Evaluation profiles for paediatric cochlear implant users with additional disabilities [www.earfoundation.org.uk](http://www.earfoundation.org.uk)


IT-MAIS Scores

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Pre & Post IT-MAIS scores

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How do we know what they hear?

An example of Ling 6 sound test with a severely motorically involved child

Detection
- Requires increased time in between presentation stimuli
- Extremely important to come out of at least 6-10 seconds of quiet
- Consistent behavioral response
  - May vary considerably but can be highly reliable

Comprehension
- Using behavioral responses to judge if child understood
- Using eye gaze communication to probe comprehension (point with your eyes to one I say)
  - Start with 2 objects,
  - move to symbols,
  - increase number of symbols

Identification of Ling 6 Sounds using eye gaze:
- Using choice of 2 objects
- Using choice of 2 symbols
- Increasing the number of symbols using the e-tran
Expressive Communication Options

- Voice on demand
- Developing control of suprasegmentals (e.g., pitch, loudness, duration) for purposes of meaningful communication
- PECS: Picture Exchange Communication System
- Eye Gaze communication & other Partner Assisted Communication (PAC) Strategies

Some PAC/ Low Tech strategies

See handout on line for details of:
- Choice making with hand selection set
- Eye pointing with E-Tran
- Laser pointing with partner confirmation
- Partner assisted auditory scrolling
- Sliding Scale partner assisted scanning
- Partner assisted row-column scanning

Partner Assisted Communication (PAC)

- ALL Partners MUST MODEL the communication themselves if the child is to expected to learn to use it. Parents & school team need to coordinate!
- Children need adequate TIME
  - To formulate
  - To execute
  - To initiate
- Communication MUST be FUNCTIONAL/MOTIVATING
Why are we using this method?

• To provide for necessary communication
• To achieve better inclusion
• Practice making choices is NOT a reason to use PAC
• PAC/switch use communication should NOT be a physio or OT goal – it must be as physically easy as possible

Easy is not always best

• In choosing vocabulary for PAC the component need to lend themselves to generation of new ideas
  – E.g. For the story Brown Bear, keeping colours and story characters separate is more complex but allows the same vocab to be used for other purposes
  – Children need to know where to find the items on their communication system, so they must stay in the same positions always.
  – Always needs to be a choice to stop or change

Eye Gaze Communication

• Requires intentionality
• Proper seating and physical support required
• Begins in a real communication, meaningful request context
Eye Gaze Communication con’t

• Begin with real objects – one neutral, one highly desired
  – Your face HAS to be neutral when offering choice
  – You must give the child what he gazes at for 2-3 seconds
    even if you think it’s not what he wants
  – Reinforce by saying “You want _____. You picked it with your eyes.”
• Once child is consistently choosing from objects move on to
  pairing objects with pictures
• Once child is consistently using pictures you may increase the
  complexity of the request task

Eye Gaze Communication con’t

• Symbol use – fist lexicon needs to serve relational
  functions that are important for a child’s day to day
  functioning in their environment such as:
  (Tatenhove, 2007)
  – Recurrence (e.g., more, again)
  – Request for assistance (e.g., help, do)
  – Rejection (e.g., no, stop, all done)
  – Requesting Action (e.g., bounce, roll, tickle)
  – Labeling is only a part of first lexicon

Eye Gaze Communication con’t

• Children can generate multiple word
  utterance using eye gaze communication
  using pictures displayed on e-tran
Children with Blindness


- For children with complete loss of vision and severe motoric limitations, the following have been helpful:
  - Db Link
  - Resources from Texas School for the Blind
  - John Tracy Clinic correspondence program
  - Project Ski Hi Deaf – Blind Program
  - (See end of reference list with handout)

Our Questions About Outcomes:

- What CI outcomes do parents report for their children with severe multiple disabilities?
- How do parents rank these outcomes in terms of importance to them?

Why these questions matter:

- In the Canadian socialized medicine system, allocation of scarce resources must be justified
- Desired/expected CI outcomes for typically developing children usually focus on the development of spoken language
- CI outcomes for children with severe multiple disabilities may not include spoken language as primary
Might this study apply to my population?

- Outcome instruments currently available capture neither the full range of outcomes reported by parents nor their priorities
- For the CI population, we need to explore the role that access to sound plays in making the child ‘a part of’ their world (Stewart, 2001)
- We need to count as legitimate constructs that parents identify as making a difference, because these form the basis for hope (see McConkey-Robbins, 2008)

Methods

- Repeated structured questionnaires:
  - Please describe differences you feel are due to cochlear implantation in the areas of:
    - Child’s communication
    - Child’s listening behaviour
    - Family interactions with child
    - Child’s social interactions
    - Child’s behaviour
    - Please describe any difficulties in managing the CI
    - Any additions?

Data Analysis

- Content analysis of repeated structured questionnaires with parents following CI activation
- Parent ranking of outcome categories
- Parent confirmation of analysis
  - Parents review extracted categories and confirm or deny each one as it applies to themselves
All participants’ children have severe multiple dx.

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<th>Subject</th>
<th>Age initial stimulation</th>
<th>Current Age</th>
<th>Type of implant</th>
<th>Additional Dx.</th>
<th>Number of interviews N = 9</th>
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<td>3 – 8</td>
<td>CA Freedom</td>
<td>Chromosome 10 deletion, autism</td>
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<td>Autism, Post CI</td>
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<td>9 – 11</td>
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<td>PDD, CP (quad)</td>
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<td>Autism, seizure, Post CI</td>
<td>1</td>
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<td>3 – 5</td>
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<td>AB Hi Res 90K</td>
<td>PDD</td>
<td>4</td>
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<td>7</td>
<td>3 – 5 (progressive HI)</td>
<td>7 – 2</td>
<td>AB Hi Res 90K</td>
<td>Peroxosomal disorder, blind</td>
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</table>

Results

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<tr>
<th>Category</th>
<th>Number of unique constructs identified</th>
<th>Number of times constructs in category referenced</th>
<th>Parent ranking of importance to them (1=most to 7 = least)</th>
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<tr>
<td>Listening (non-linguistic)</td>
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<td>5</td>
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<td>Expressive Communication</td>
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<td>Connectedness</td>
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<td>Family Systems</td>
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<tr>
<td>Receptive Communication</td>
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<td>20</td>
<td>3</td>
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<td>Child Affect</td>
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<td>19</td>
<td>1</td>
</tr>
<tr>
<td>Challenges</td>
<td>6</td>
<td>9</td>
<td>7</td>
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</tbody>
</table>

Child Affect

• Reduction in self stimulating behaviour
• Less effort for child to participate in family and community interactions
• Better quality attention
• Child happier overall
**Connectedness**

- Others interact more with child simply because they know the child can hear
- Child more able to tolerate new experiences
- Child’s ability to interact with others enhanced
- Child more present, more engaged
- Broadened social sphere, not restricted to signers
- Child more connected to family and peers

**Receptive Communication**

- Child turns consistently to name
- Child continues to progress in auditory comprehension
- Child responds with appropriate actions to familiar songs
- Child follows some routine verbal directions
- Child demonstrates some open set auditory comprehension
- Comprehension is inconsistent

**Family Systems**

- Family feels more ‘normal’ to parent
- Less effort for parent and sibs to include child in family interactions
- CI technology easier to manage than hearing aids
- Ability to interact through sounds brings parent joy
- Family has more fun with child
- Parents now feel that they have done everything possible to support child’s optimal function
Listening (non linguistic)

- Child responds to voice from another room
- Child loves/appreciates music
- Child laughs at funny sounds
- Child reliably communicates when CI signal is bad or off
- Child searches for environmental or voiced sound
- Child removing CI headpiece is a good indicator of engagement
- Child independently replaces headpiece

Expressive Communication

- Child makes non-verbal efforts to engage others
- Child’s vocal volume moderated to socially acceptable level
- Increase in amount and variety of vocalizations post CI
- Child uses inflection with meaning
- Child imitates vocal patterns/melody
- Child imitates c-v combinations
- Expressive communication is inconsistent

Challenges

- Child removes headpiece to get attention or to protest
- Child can’t wear CI when doing some favorite things, e.g. roughhouse or trampoline play
CHILD AFFECT

He seems to be immensely happy to be socially involved in others' lives. It was too hard to get him to focus, and hold focus (before the implant). Sound has definitely changed this.

CONNECTEDNESS / INCLUSION

At church, people will talk to her and she will smile. It takes less specialized effort to engage her, so a wider circle of people can do it successfully. She's more present, more engaged – she's no longer alone in the room.

CONNECTIVITY/ LISTENING

She will turn to look at me when I make one of her favorite sounds. It is the only time she orients toward me. When she looks at me, even if it is really only to see the sound, she looks into my face and into my eyes and I get a chance to love her with my eyes. I use her ears, to get to her eyes, to get to her heart.
RECEPTIVE COMMUNICATION

I said ‘yogurt’ in conversation with my son, and happened to glance across the kitchen at my daughter and she was signing yogurt! She definitely got some, too!

FAMILY SYSTEMS

You used to have to work so hard just to get him to look at you. Now it is effortless on our part, he’s on his own steam.

LISTENING (non-linguistic)

I am happy to see that he can at least hear his environment .. and is able to dance when music comes on. I think that is what matters most, because the CI brought music into my boy’s life that was not there before
EXPRESSIVE COMMUNICATION

He now knows for sure that his voice has power. He has differentiated tone of voice for a long time. But now it seems more deliberate. It's easier now to tell what his mood is from another room.

CHALLENGES

At first she wanted nothing to do with sound and quickly learned to take it (the CI) off. We got a hat for her and my mom attached straps (helmet style) and we tied it under her chin.

Instrument Development Process

First Steps:

- Participants confirmed or denied each proposition identified by the full group for their own child and family

- Propositions that were confirmed by the majority of participants (4/7) will form the basis for items in the instrument development process
The AH HA! Moment

• Parent rankings of importance to them did not coincide with the number of times a theme was cited in the data
• Listening, Expressive Communication and Connectedness were cited most often in the data
• Parents rated Child Affect, Connectedness and Receptive Communication as 1, 2 & 3 in terms of importance to them
• This was an important data confirmation step

Draft Item Development Rationale

• Desire to make parent identified constructs the foundation of the instrument
• The desire to avoid duplication with instruments already in general use: Draft items were generated by review of project data in comparison with IT MAIS, REEL, PRISE, Little Ears, & Rosetti.
• Desire to capture very small changes

Inclusion of Caregiver Well Being

• We contend that caregiver well being should be taken on par with child outcomes in the determination of ‘success’ for this population
• We base this contention not only on our findings in this study and in our practice, but on the 2010 recommendation of a Canadian National Steering Committee on Rehabilitation of Children and Youth that caregiver well being be embedded in every aspect of our practice.
Proposed Rating Scale

Rating Scale for each item:
• (Never) 0 1 2 3 4 5 6 7 8 9 10 (Always)
• A ten point scale allows for sufficient variance to capture small differences
• Research shows that most of the relationship between Likert scale ratings and functional outcomes is seen in the lower and higher extremes of the scale

Joys and Challenges of Unfunded Research

• Joys:
  – Clinically relevant
  – Research incorporated into clinical practice
  – Many families excited about helping to shape practice through research
  – Departmental support
  – The pleasure of drafting more skilled family members to assist in data entry

Child Affect Due to CI Use (4 items)

1. Child shows evidence of happiness and contentment by smiling, lack of agitation, laughing and/or use of happy voice quality.
2. Child demonstrates contentment by lack of agitation and or irritability and/or tone of voice throughout waking hours.
Remainder of Child Affect Items

3. Parent/caregiver can determine child’s emotional state from another room based on the child’s vocalizations.
4. Child reacts to music by smiling, movement associated with pleasure, focused attention and/or demonstration of anticipation.

Child Interaction (12 items)

1. Child enjoys interaction (has fun with) with parent in routine caretaking activities as evidenced by smiles, attentiveness, tone of voice and/or demonstration of anticipation.
2. Child enjoys interaction with siblings in routine activities at home as evidenced by smiles, attentiveness, tone of voice and/or demonstration of anticipation.

Remainder of Child Interaction Items

3. Child enjoys interaction with adults, other than parents in routine caretaking activities as evidenced by smiles, attentiveness, tone of voice and/or demonstration of anticipation.
4. Child enjoys interaction with adults, other than parents, in activities outside the home as evidenced by smiles, attentiveness, tone of voice and/or demonstration of anticipation.
5. Child enjoys interaction with peers, other than siblings, in routine activities at home as evidenced by smiles, attentiveness, tone of voice and/or demonstration of anticipation.

6. Child enjoys interaction with peers, other than siblings, in activities outside of the home as evidenced by smiles, attentiveness, tone of voice and/or demonstration of anticipation.

Child is able to tolerate new experiences as evidenced by lack of agitation and/or behavior indicating pleasure in: Rate each:

7. Family gatherings.
8. Church, Synagogue, Mosque, other
9. Restaurants.
10. New experiences outside the home.

11. The child is “no longer alone in the room” (present but not connected to others in the room as evidenced by focus of attention and appropriate emotional reactivity).

12. People outside of the family’s close social circle interact verbally with child.
Child Device Use (10 items)

1. Child wears device without removing it. Child pulls headpiece off to protest or to get attention. Rate each:
   2. At home.
   3. At school.
   4. In other settings.

Remainder Child Device Use Items

5. Child attempts to put the headpiece back on him/herself.
6. Child removes headpiece if device is not working properly. Child removes headpiece to signal lack engagement or to request a change in activity. Rate each:
   7. At home.
   8. At school.
   9. In other settings.

10. The parent/caregiver feels that a management of equipment requires minimal effort.
Parent/Caregiver Well Being (12 items)

1. Parent feels connected to child through sound in meaningful and mutually enjoyable ways.
2. Parent/caregiver feels confident that they are doing/have done all that is possible to support their child’s ability to be connected to others.

3. Child responds positively to parent/caregiver’s voice as evidenced by calming, smiling, movement associated with pleasure, focused eye gaze and/or taking vocal turns.
4. Parent/caregiver feels able to successfully include their child in routine family interactions.

Remainder Parent/Caregiver Well Being Items

5. Parent/caregiver feels able to successfully include their child in activities outside the home (church, restaurants, shopping, etc.).
6. Parent/caregiver feels well supported by CI team.
7. Parent/caregiver feels able to meet the needs of other family members (siblings, spouse) without feeling overwhelmed.
8. Parent/caregiver feels own personal needs (emotional, intellectual, physical) are being met.
9. Parent/caregiver experiences joy in interacting with child who uses CI.
10. Parent/caregiver feels able to interact with child from a distance (5 feet or more).

11. Child’s behavior can be effectively managed through spoken direction/input.
12. Parent/caregiver feels family life is ‘normal’

Next Steps

- Initial item selection will be vetted with parent participants and clinical experts
- Once the pilot set of items is agreed upon, a pilot study will take place. We would aim for at least 30 families.
- Item analyses will be done (correlational and factor) to explore how they perform
• A second draft incorporating the results of item analyses will be used as the primary trial. This will involve an initial administration as well as re-administration approximately 1 month later to explore reliability and standard error of measure.
• A final form of the instrument will be developed based on these results

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