Clinical Outcomes in Cochlear implantation in young children with unilateral hearing loss: 
Candidacy and post-operative experiences and considerations

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No Disclosures
The impact of diagnosis

UHL is considered significant enough to be diagnosed in UNHS programs

WHO guidelines for screening (Wilson’s Criteria 1968)

1. The condition should be an important health problem.
2. There should be a treatment for the condition.
3. Facilities for diagnosis and treatment should be available.

- Incidence 0.6:1,000 (WHO) 37-58% will have deterioration of hearing
- 10-22% will have a progressive bilateral loss (8% in current study)
- Parental stress at time of diagnosis is not related to level of hearing loss

The impact for children’s listening

- Risk to academic, language, speech and psychosocial development
- Cognitive performance poorer than typical hearing
- Impact on cognitive function and phonological processing
- Difficulty with sound localisation
- Impact on pragmatic skills

(Bess, Klee, & Culbertson, 1986; Culbertson & Gilbert, 1986; Lieu et al., 2012; McKay, Gravel, & Tharpe, 2008), (Martínez-Cruz, Poblano, & Conde-Reyes, 2009), (Bess et al., 1986; Culbertson & Gilbert, 1986; Fischer & Lieu, 2014; Fitzpatrick et al., 2014; Holstrum, Gaffney, Gravel, Oyler, & Ross, 2008; Lieu et al., 2012; Tharpe, 2008), (Ead, Hale, DeAlwis, & Lieu, 2013), (Abel & Lam, 2008; Johnstone, Nabelek, & Robertson, 2010; Tavora-Vieira & Rajan, 2015), (Goberis et al., 2012), (Firszt, Reeder, Dwyer, Burton, & Holden, 2015; Johnstone et al., 2010; Van Deun et al., 2010)

The impact for the brain

- Neuroanatomical and physiological changes in both audition and cognition
- Compromised development of executive functioning
- No compensatory strategies for binaural listening
- Higher proportion of children with UHL with cochlear nerve deficiency (when compared to bilateral HL)

(Yang et al., 2014; Zhang et al., 2015), (Beer et al., 2014; Hutson, Durham, Imig, & Tucci, 2008; A. Kral, Heid, Hubka, & Tillein, 2013; Andrej Kral, Hubka, Heid, & Tillein, 2013; Scheffler, 1998; Schmithorst, 2005; Tibbetts et al., 2011; Zhang et al., 2015), Gordon, Wong, & Papsin, 2013; Hutson et al., 2008; A. Kral et al., 2013), (Clemmens et al., 2013; Kachniarz, Chen, Gilani, & Shin, 2014; Laury, Casey, McKay, & Germiller, 2009; Nakano, Arimoto, & Matsunaga, 2013)
Impact of UNHS on enrolment in EI

Currently 20% of children enrolled in EI (0-6 years) have UHL.
What do we know about children in with UHL in EI? n=94

- 20% of children start early intervention with UHL
- 8% become bilateral
- 17% deteriorating UHL
- 17% ongoing OME

Median age of starting EI higher than for bilateral HL
Long Term Outcomes with UHL

Standardised assessment for children with UHL fit well with normal curve

Shift in auditory comprehension scores
PLUS
Poorer results for auditory memory and use of grammatical markers
Outcomes beyond traditional measures: Functional Listening Index

Number of Observed Listening Skills by Age of Child with no diagnosed additional needs

Functional listening outcomes for children with typical hearing

Functional listening outcomes for children with UHL

The Sydney Children’s Hospitals Network
First Sounds Cochlear Implant Program
The Shepherd Centre
Giving deaf children a voice
Theory of Mind skills with hearing loss

Typical Hearing
Average Age 5.7yrs
(Atypical 8%)

Bilateral HL
(Atypical = 62%
p=0.0004)

Unilateral HL
(Atypical = 45%
p=0.0005)

(Wellman & Liu, 2004)
Amplification choices for UHL in EI

Most likely to be aided if UHL is moderate-severe
Combination of parent choice & professional guidance

Over half are unaided, others have devices based on hearing level, aesthetics or funding availability

Baha 5%
Bone Conductor 16%
Cochlear Implant 12%
Hearing Aid 15%
No Device 52%
Causes of UHL in EI

n=89

- Unknown 48%
- Microtia/Atresia 25%
- LVAS 2%
- Cochlear Fracture 2%
- Cochlear Nerve Deficiency 22%

n=55 with severe SNHL or worse

- Unknown n=19
- MRI n=36

Present Nerve n=16 44%
(13 proceeded to CI & 2 on CI evaluation)

Cochlear Nerve Deficiency (CND)
- n=20 56%
  (5, 25% ANSD)
Cochlear implant and UHL
n = 13 children under 12 years old

<table>
<thead>
<tr>
<th>DEMOGRAPHICS</th>
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<tbody>
<tr>
<td>Median age at implant</td>
<td>6 (3.3-11.1 years)</td>
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<tr>
<td>Ear implanted</td>
<td>62% R, 38% L</td>
</tr>
<tr>
<td>gender</td>
<td>9 M, 4 F</td>
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<tr>
<td>Onset/aetiology of HL</td>
<td>8 = UNHS</td>
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<tr>
<td></td>
<td>2 = head trauma</td>
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<tr>
<td></td>
<td>2 = LVAS</td>
</tr>
<tr>
<td>Early intervention</td>
<td>6 referred &lt;5 years (0.8-4.8 years)</td>
</tr>
<tr>
<td>Previous device use</td>
<td>1 BC then Baha</td>
</tr>
<tr>
<td></td>
<td>1 HA</td>
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Family/individual goals

- 3 children presented to EI before the age of 2
- 10 children presented to CI program specifically to investigate listening/speech/language/academic difficulties or to follow up on optimize listening

I’d really like to be able to imitate individual sounds via direct audio input to one ear

I’d really like to be able to listening easily and accurately in background noise and from a distance
Cochlear implant and UHL: outcomes

n = 13 children under 12 years old

- 12/13 children wearing the device for whole school day
- Gradually increasing use at home as well
- Challenge of a diverse and small group
  - FM system: to better ear? To CI ear?
  - Longer time for improvement: change therapy model?
  - Change programming approaches?
  - Having the tools to measure what children and families are reporting
  - All have individual ear improvement: 8 have discrimination, 5 detection and developing discrimination
  - All report real world improvement for complex listening
Case Study- CI for UHL, 2 years experience

- Congenital unilateral profound hearing loss, diagnosed after UNHS
- BC at 2.5 years, Baha implanted at 4.5 years, CI at 7 years
- Typical speech and language, poor reported function in noise, difficulty in noisy classroom e.g. with spelling tasks, struggling with music lessons

Now 2 years post-implant
- Reported improvements in classroom listening
- Recent piano exam highest score in piano school
- Improvements in language and speech perception

"Now, after one year, things are going very well. I love my CI. My spelling is awesome, I can still hear when my friends talk and I can even hear where sounds are coming from."
How is CI for UHL different?

- **Candidacy**: assessment of monaural listening vs potential binaural listening, individual ear audiometry, MRI

- **Activation**: 9 children reported more of a ‘sensation’ than a sound at activation. Use of intraoperative testing results as the basis of an initial program.

- **Listening development**: how to work on individual ear? Direct audio input via cable/streaming.

- **Therapy goals**: some individual ear work, mostly binaural listening development

- **Outcome measurement**: short-term? long-term?
What supports our work with UHL

• The **good ear** may lose hearing, **both ears** may lose hearing
• Language might be good but consider **other impacts** of listening with one ear:
  – Auditory memory, grammatical markers may show impact
  – Functional listening highly variable
  – Theory of Mind Acquisition more likely to be atypical
• **Best long term outcomes** are children with UHL who enter EI early, early device fitting and good device compliance
• If newborn screening of hearing is going to identify unilateral hearing loss, families need a **clear management plan**
• Extend education in the field: **referral pathways**
The Shepherd Centre team

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