Cochlear implant candidacy considerations and outcomes for children with Auditory Neuropathy

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Disclosures

- Dr Jaime Leigh
  - None
- Ms Evelyn Do
  - None
- Prof. Richard Dowell
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  - University of Melbourne department received project-based grant funding
- Ms Dawn Choo
  - None
- Dr Shani Dettman
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- Unlabelled use of commercial product
  - Nucleus CI, use for children younger than 12 months of age
Background

- CI outcomes for patient with Auditory Neuropathy (AN) are variable
- Majority benefit and achieve outcomes comparable to SNHL-CI
- Approx. 25% of children with AN obtain minimal benefit from CI
  - not achieving functionally useful hearing and/or no improvement on pre-op auditory capacity (Gibson & Sanli, 2007; Teagle et al, 2010; Roush et al., 2011)
- 90% of children with 8th nerve hypoplasia get no open set perception with CI (Rance & Star, 2015)
Background - Site of lesion

- AN sites of dysfunction (Rance & Star, 2015)

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<th>Site of lesion</th>
<th>Pathological mechanism</th>
<th>Locus</th>
<th>Aetiology examples</th>
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<td>Presynaptic</td>
<td>Receptor disorder</td>
<td>Inner hair cell</td>
<td>Oxygen deprivation</td>
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<td></td>
<td>Synaptic disorder</td>
<td>Ribbon synapse</td>
<td>Genetic, OTOF</td>
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<tr>
<td>Postsynaptic</td>
<td>Diminished auditory nerve activity</td>
<td>Dendrites &amp; axons</td>
<td>Peripheral neuropathy, FRDA, CMT2</td>
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<td></td>
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<td>Ganglion cells</td>
<td>Kernicterus</td>
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<td>Hypoplasia</td>
<td>Auditory nerve</td>
<td>Congenital malformation</td>
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<td>Conduction disorder</td>
<td>Brainstem</td>
<td>Acoustic neuroma, MS</td>
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- CI benefits for individuals with presynaptic AN similar to those with SNHL
- CI outcomes for those with postsynaptic are variable, and on average poorer than presynaptic cases
Study aims / overview

• Review the range of speech perception and language outcomes achieved by children with AN using cochlear implants
• Compare the outcomes for children with AN to those of children with SNHL using cochlear implants
• Determine if the CI outcomes for children with AN from our cohort are consistent with the findings of Rance & Starr (2015)
Participants

- Retrospective review of the case notes for 76 children with AN findings referred to CIC between 1998 and 2015
- 43 children with AN proceeded with CI (4.5% of children with CI in our centre)

<table>
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<th>Demographic</th>
<th>Children with AN (N=43)</th>
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<tr>
<td>Mean age at HL identification</td>
<td>6.5 months (range 2 wks to 2.8 yrs)</td>
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<td>Mean age at HA fitting</td>
<td>10.7 months (range 7 wks to 3.4 yrs)</td>
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<tr>
<td>Mean age at CI</td>
<td>3.12 years (range 7 mo to 16 yrs)</td>
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<tr>
<td>Mean unaided PTA (best ear)</td>
<td>98 dBHL (range 58 to 125 dBHL)</td>
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<td>Co-morbidity / additional disability</td>
<td>15 children</td>
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</table>
Aetiology / Site of lesion (n=43)

- Presynaptic - 42%
- Unknown - 35%
- Postsynaptic - 23%
- Prematurity/hypoxia (n=18)
- Jaundice/Kernicterus (n=4)
- Cochlear nerve deficiency (n=6)
- Unknown (n=15)
Communication assessments

• Assessment intervals
  • pre implant and where possible 1, 2, 3 years post implant and 5 years age and 8-9 years of age

• Language
  • Preschool Language Scale (PLS)
  • Clinical Evaluation of Language Fundamentals (CELF)

• Speech perception
  • monosyllabic words presented audition alone
  • Categories of auditory performance index (CAPI)

• Results from each child’s most recent assessment are presented
Language outcome for AN-CI group (n=26)

The Royal Victorian Eye and Ear Hospital
Language outcome (n=14)
Children with CI<2.5 years and no additional disabilities

[Leigh, 2016]
Categories of auditory performance index (CAPI)

• Suitable for children of any age
• Does not required child to undergo formal testing
• 8-point categories of auditory performance
  0 = No or limited sound awareness
  1 = Sound awareness
  2 = Limited closed set speech perception (score <75%)
  3 = Established closed set perception (score ≥75%)
  4 = Limited open set perception (score <50%)
  5 = Sound open set perception (score ≥50% but <75%)
  6 = Advanced open set perception (score ≥75%)
  7 = Advanced open set perception in noise (score ≥75% in +20dB SNR)
  8 = Advanced open set perception in noise (score ≥75% in +10dB SNR)
Speech perception for AN-CI - CAPI (n=43)
Speech perception - CAPI (n=20)
Children with CI<2.5 years and no additional disabilities

Significant open set perception

mean SNHL-CI group (n=32) [Leigh, 2016]
Conclusions

• Speech perception performance for children with AN who received their CI under 2.5 years, with no additional disabilities, was similar to peers using CI with SNHL

• Language delayed compare to peers with normal hearing and SNHL-CI
  • additional educational support warranted

• Caution/careful counselling for children with CND due to limited perceptual skills and high rate of non-use
  • no open-set perception
  • 50% non-use

• Outcomes from our cohort consistent with findings reported by Rance & Starr (2015)