Paediatric cochlear implants: challenges and future directions

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Current Developments and New Directions
In
Pediatric Audiology, Istambul 2011
The best of times ...

- Late identification
- Delayed diagnosis
- Poor amplification
- Education: low expectation
- No cochlear implants
- Public Health Policy
Prostheses - implantable
Overview

- Neurobiology of deafness
- Plasticity into Practice
- Early cochlear implantation
- Bilateral CI
- Medical and surgical aspects
- Electro-acoustic hearing / ABI etc
- Turkish Temple
Lost around time of birth
Normal morphogenesis
Critical gene product *(Steel 2009)*
50-90% loss of CN neurones
Quality and number of synapses
Hyperexcitability (Eggermont 2008)

Ryugo et al 1997
- No sensory system exists in isolation
- Non-auditory effects of deprivation – attention, working memory, executive functioning etc
Development Human Cortex

Conel, 1939-1967
Synaptic Pruning

-3. M. Birth
1 y. 2 y. 4 y. 6 y. 15 y. Adult

Synaptic Maturation

Kral and O’Donoghue, New Eng J Med 2010
Cochlear Implants and Plasticity
Cochleotopic Organisation of Cortex - CI
Cochleotopic Organisation of Cortex - CI

(Fallon et al., 2009)
Cross modal reorganisation after CI

Current Density Reconstructions for the “P1” CAEP

Normal Hearing  
n=10

Early Implanted  
n=8*

Late Implanted  
n=8*

* Corrected for ear of stimulation

Dorman et al 2009
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Not about ears: its all about the brain
Prostheses: *brain access tools*
‘Its not what we hear, its what we do with what we hear’
Plasticity in Practice

- 46 million words heard by age 4 \textit{(Hart and Risley)}
- 20,000 hours listening for reading \textit{(Dehaene)}
- Hearing loss: 3 times exposure to learn new words / concepts \textit{(Pittman)}
- Acoustically favourable conditions
Synaptic Pruning

Synaptic Pruning

Synaptic Pruning

- Auditory cortex
- Visual cortex
- Prefrontal cortex

Reading

20,000 hours listening
(normally hearing)

Early intervention

Receptive and Expressive standardised language scores as a function of age of identification for children with 'normal cognition' (Yoshinago-Itano, 1999)

<table>
<thead>
<tr>
<th>Age of identification</th>
<th>Language quotient</th>
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<tbody>
<tr>
<td>&lt;6m</td>
<td>40 60 80 100 120</td>
</tr>
<tr>
<td>&gt;6m</td>
<td></td>
</tr>
</tbody>
</table>

Receptive

Expressive
Early intervention

Receptive and Expressive standardised language scores as a function of age of identification for children with 'low cognition' (Yoshinago-Itano, 1999)

- **Receptive**
  - <6m
  - >6m

- **Expressive**
  - <6m
  - >6m

Age of identification

40 60 80 100 120

Language quotient
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Early Cochlear Implantation
Age at implant 0-4 years 2000 to 2010

Source: Cochlear Ltd database
Age at implant 0-4 years 2000 to 2010

Source: Cochlear Ltd database
Audiological Certainty

- What age?
- What measures?
- What validity?
- Who decides?
- Parameters of hearing aid trial
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European Data - Bilaterals

Number of bilateral Nucleus recipients

Source: Cochlear Ltd database
Bilateral implants children 2005:2010

Source: Cochlear Ltd database
Bilateral implants children 2005:2010

Source: Cochlear Ltd database
Bilateral implantation recommended for children & some adults

- Simultaneous
- Additional contralateral
National Audit Bilaterals: UK

- Localisation
- Speech recognition in background noise
- Speech
- Language
- Listening
- Parental perception
- Quality of life
- Surgical data including complications
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Surgical challenges - BICI

- Minimal Access Surgery
- Operating time
- Anesthesia
- Facial nerve injury
- Co-morbidity – known or unknown
- Electrocautery
- Loss vestibular function / residual hearing
- Meningitis risk
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Hearing aids + Implants

Electrical Stimulation

&

Acoustical Stimulation
Electro-acoustic hearing
Single Side Deafness and CI

Assessed for Eligibility

- Excluded

Randomized

- CI
- CROS Aid

Allocated to Intervention

- Did Not Receive Intervention

Received Intervention

Follow-Up

- Discontinued Intervention
- Lost to Follow-Up

Followed Up

Analysis

- Not Analyzed
- Analyzed
Advances in Implant Technology

Drug and Stem Cell delivery systems
Auditory Brainstem Implant

(Lenarz et al., Colletti et al. 2006)
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Temple of Artemis
Visioning the Future: the Sound Foundation

Children with Hearing Loss

- Identification
- Assessment
- Intervention
- Education
- Families
- Personnel

- Quality Services
- Information Systems
- Public Health Policy
- Access
- Evidence Based Practice
- Technological Advances

J. Gravel 2007
Information and systems give meaning to everything we do; there is no point in going through the motions of providing a service if there are no systems on which to support, record or monitor the process and outcomes.

Sir Muir Gray 2007
Summary

- Deafness - not just about hearing loss
- Prostheses as brain access tools
- Earlier is best - *but not a cure!*
- Bilateral CI - timely, safe, efficient
- Combined acoustical and electrical hearing
- Audiological certainty - central
- Paediatric audiology ‘Temple’