Pediatric Single Sided Deafness
Cochlear Implantation: Outcomes,
Candidacy Considerations and
Research Opportunities

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Disclosures

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  - Cochlear Americas
  - Advanced Bionics
- Research Funding
  - Cochlear Americas
  - Advanced Bionics
- SSD Clinical Pilot Study
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Children with Single-Sided Deafness

- May present with difficulties including
  - Decreased hearing in background noise
  - Poor localization ability
  - Speech and language deficits
  - Decreased educational performance
  - Increased incidence of behavioral issues in the classroom
Current Treatment Options

- Traditional amplification
- CROS (Contralateral Routing of Signal) systems
- Osseo-integrated bone anchored devices
- FM systems in the classroom
- No treatment

These options cannot provide hearing to the affected ear or binaural hearing.
NYU SSD Cohort

• 12 adults
  • 8 off study – off label
    • 2 Meniere’s
    • 6 ISSNHL
      • 1 with AN opposite ear
  • 4 in SSD pilot study
    • All ISSNHL

• 3 Children
  • EVA
    • genetic HL, sibling with bilat CI
    • age 2 with congen. SSD

• Outcomes:
  • all full time users, all adults report tinnitus suppression
  • Overall Adults performing better than children but children have significant benefit
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S3 Results: 3-Month Data
Monosyllabic Words and Sentences
Summary
SSD Pediatric Cohort

- Overall, subjects demonstrated:
  - Open-set speech perception in the implanted ear
  - Bilateral improvement in background noise

- Subjectively, parents and schools report:
  - Increased attention
  - Improvement in grades
  - No longer asking “what” repeatedly
  - No longer fearful of social situations (e.g., school cafeteria, outings with friends)

- Cochlear Implants: Viable treatment option for children with SSD
Trends in SSD

- Counseling for SSD now includes CI routinely
- More adults asking about SSD CI
- More difficult to get insurance precertification and payment
  - Insurers agree to Bone Conduction solutions for SSD
  - Insurers agree to sequential bilat CI
- More children with SSD emerging

  - Why are we not considering CI for young congenital SSD?
  - What is the age cut off for SSD in children?
    - Do we consider SSD CI in an well adapted 7 yo?
Considerations

• SSD Candidacy Indications
  • **Absolute**
    • Adults and children that are likely to lose hearing in the good ear over time
      • EVA and other malformations, genetic conditions, AIED, tumors, inflammatory disease, metabolic disease, ototoxicity etc.
  • **Non absolute**
    • Stable situations (other technology available)
      • ISSNHL, trauma, noise induced, etc
• Financial and QOL issues
Candidate

- 63 yo with Meniere’s AD
  - Treated with diuretics and Na restrict.
  - Treated with transtympanic gentamycin
  - Stable for 2 years, now with vertigo again
  - Options:
    - More gentamycin (steroids)
    - Bone Conduction Options
    - Labyrinthectomy w/ or w/o CI

- CI now at 65% CNC CI only, no vertigo
Candidate

- 64 yo with ISSNHL and vertigo
- Treated with steroids (systemic and IT)
- Hearing never recovered
- MRI- contralateral AN
9 yo with assym SNHL
• Left ear declined 2 years earlier
• EVA
• c.365insT of PDS gene (only one)
• Two other sibs with same genetic issue
• BAHA trial not well received
EVA CASE

• Now 2 years post CI

• 3 mo perception results- CI alone
  • 18% words
  • 47% sentence scores

• 1 year perception results
  • 64% words
  • 72% sentences

• 100% combined condition, BKB-SIN sf/nf +0.5dB, sf/nl -0.5dB, sf/nr +4.5dB

• Very happy user! More engaged in social events, more confident.
SSD CI Candidacy

• Almost everyone with SSD is a candidate for a CI!

• **Except**
  • 5, 10, 15, 20 years SSD: above what duration of deafness is it too late or not a good idea
  • Does a neural auditory gate close?

  • Consider the new AN in an only hearing ear with long duration SSD
  • Consider the NF2 patient with sleeper ABI
Research Opportunities
Clinical Research Considerations

• Factors that affect outcome
  • Duration of hearing loss
  • Bimodal studies
  • Etiology of hearing loss
  • Cognitive issues
  • Age of patient
• Anatomic issues – malformations (is the signal going to be good enough)
• CI technology – new devices/electrodes
• Electrode placement* - depth and position affects pitch allocation
  • Long, short, outer wall, perimodiolar
• Programming methods
  • Pitch matching considerations
A NEW SCIENTIFIC OPPORTUNITY

• Partial hearing patients provide the opportunity to study several phenomena in a much cleaner way than it was ever possible:
  • Pitch matching
    • What is the pitch of a stimulus sent to a given intracochlear electrode?
    • How does the tonotopic frequency map in a normal ear differ from the standard CI map?
    • Does the pitch perception of a given electrode change over time? How much?
  • Adaptation to modified peripheral frequency maps
  • The extent and limitations of auditory plasticity
  • Clinical implications of manipulations/delivery of signal- Are these patients the perfect model? Will lessons learned benefit all CI users?
Pitch matching- Subject 3

Pitch Matching – Subject 1 (Nucleus)

Svirsky et al., 2011
FREQUENCY TABLE SELECTION
“The player lost a shoe”
Svirsky et al., 2011

188-7938
“Sounds very high pitched”

63-3372
“Shoe sounds like sue and the 'p' is not there”

313-5188
“The 'sh' is pretty good here but not the best”

188-18,000
“That’s a REAL sh!”

188-9804
“This may be the clearest, may be the first choice”
“Big dogs can be dangerous.”

500ms of silence

“Big dogs can be dangerous.”

Speech Processor

Acoustic Model,
- Analysis filters fixed
- Synthesis Noise Bands or Tones adjustable by listener

Svirsky et al., 2014

Acoustic Ear

CI
Subject L1

THE PLAYER LOST A SHOE - NOISE

SS MODEL WAS MORE SIMILAR TO THE CI THAN CLASSIC MODEL

THUS, NULL HYPOTHESIS IS FALSE (CLASSIC MODEL NOT VALID) FOR L1

Svirsky et al., 2014
Thanks you for your attention