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Expanding Indications for Cochlear Implants to Children with Unilateral Hearing Loss

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Disclosures

- Dr. Park receives research grant support from MED-EL Corporation
- This study is being completed under an Investigational Device Exemption (IDE). The MED-EL SYNCHRONY Cochlear Implant System is not FDA approved for single sided deafness in children under 5 years of age.
Learning Outcomes

After this course, participants will be able to:

- Describe the characteristics of children with severe-to-profound unilateral sensorineural hearing loss.
- Discuss challenges in testing and programming children with cochlear implants and normal hearing in the contralateral ear.
- Identify potential benefits and outcomes children with unilateral hearing loss may experience when using a cochlear implant.
Characteristics of Children with Unilateral Hearing Loss

- **Educational Challenges**
  - Approximately 1/3 of children with UHL repeat a grade (Bess & Tharpe, 1984)
  - 12-41% require additional educational support (Lieu et al, 2004; Lieu, 2018)
  - Fare more poorly on cognitive measures (Fischer & Lieu, 2014; Lieu, 2013; Niedzielski, 2006; Purcell et al, 2016)

- **Behavior concerns** (Bess & Tharpe, 1984; Lieu, 2004; Tharpe, 2008)
Characteristics of Children with Unilateral Hearing Loss

- Communication Challenges
  - Poorer language outcomes (Lieu, 2013; Lieu et al, 2010; Sangen, 2017)
  - Difficulty understanding speech in noise (Griffin, Poissant, & Freyman, 2018)
  - Difficulty with localization (Reeder, Cadieux, & Firszt, 2015)

- Quality of Life Challenges
  - Greater difficulties reported on QoL measures (Griffin, Poissant, & Freyman, 2018; Reeder et al 2015)
  - Higher levels of fatigue (Hornsby et al, 2013).
  - May impact higher level functions such as auditory attention, executive function, and sensory-motor control (Conway, Pisoni, & Kronenberger, 2009; Ead et al, 2013; Lieu, 2013; Polonenko, Papsin, & Gordon, 2018; Propst et al, 2010; Sharma, Dorman, & Spahr, 2002; Tibbets et al, 2011)
    - Cortical changes in children have been noted as a result of imbalanced auditory input (Kral et al, 2013; Propst et al, 2010; Schmithorst et al, 2014; Zhang et al 2016)
Etiologies

- Cochlear Nerve Deficiency
  - Absent Nerve
  - Hypoplastic Nerve
- Cytomegalovirus (CMV)
- Malformation
- Meningitis or infection
- Trauma
- Idiopathic

Current Treatment Options

- Limitations:
  - Not providing neural input to the affected side.
  - Variable outcomes for using binaural cues to aid in listening in noise (Hol et al, 2010; Kunst et al, 2008)
  - Localization abilities no better than chance (Rosman et al, 2003; Wazen 2005; Hol et al, 2004; 2010)
Cochlear Implantation In Pediatric Cases of Unilateral Hearing Loss

PUHL Clinical Trial
Aim: To investigate the effectiveness of cochlear implantation in children with moderate-to-profound unilateral hearing loss.

Protocol Development
- Inclusion Criteria
- Patient Characteristics
- Test Battery
Inclusion Criteria

- Children age 3.5-6.5 years
- Typically developing
- PTA of $\geq 70 \text{ dB HL}$ in one ear and normal hearing in the contralateral ear
- Aided CNC word score of $\leq 30\%$ in the ear to be implanted
- No evidence of cochlear nerve deficiency (CND)
- No evidence of ossification
- No significant malformations
- English is the primary language

Device

- MED-EL SYNCHRONY Flex28 or Flex24
  - Array choice at surgeon’s discretion
- SONNET Speech Processor
  - All programmed in FS4
  - Omni-directional mode with wind noise reduction disabled
  - No use of RONDO processors
Participant Characteristics

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<th>Years Profound</th>
<th>Etiology</th>
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Test Battery

- Monitoring of unaided thresholds
- Sound field CI thresholds
- Word Recognition
  - CNC
- Hearing in Noise
  - BKB-SIN, spatially separated
- Localization
- Questionnaires
Detection Thresholds
Detection Thresholds
Detection Thresholds

SL with comment: Plug and muff thresholds, without CI processor

CR: Plug and muff thresholds, with CI processor
Outcomes
The Good Stuff

QoL and Inventory Outcomes
- Just 12-month data
- Peds-QL Fatigue Scales
  - Parent and Child ratings
- Pediatric SSQ
  - Listening Effort Subscale
Word Rec Setup

[Image of earphones and a device]

Word Rec Setup

[Image of earphones and a device]

[Continued]
Word Rec Setup

Masking Concerns

- Children are more susceptible to masking than adults. (Corbin, Bonino, Buss, & Leibold, 2016).
- Children with unilateral hearing loss may not develop auditory attention networks as robust as children with normal hearing in both ears. (Propst, Greinwald, & Schmithorst, 2010)
Plug and muff concerns
Localization

Localization

[Diagram of localization setup]

[Image of localization equipment]

[Image of localization setup with markers]
Localization

![Image of localization experiment]

Localization

![Graph showing localization response over 3 months]

3-Months
Localization

9-Months

Localization

18-Months
Localization

Speech Perception in Spatially Separated Noise
Speech Perception in Spatially Separated Noise

BKB-SIN: Normal Hearing Controls

SNR-50 (dB)
Children with CIs and UHL need to set their own targets!
Programming Considerations

- Unilateral malformation cases may benefit from advanced mapping.
  - Pitch ranking and comparisons
- Plug ear when measuring Ts
  - They use all kinds of cues
  - They can hear you click a mouse.
- Unique methods for scaling
  - ESRTs are your friend
  - Create your own charts
Summary

Clinical Consideration: Threshold Testing
- Be aware of what the normal hearing ear is contributing

Clinical Consideration: Word Recognition Testing
- Isolate the CI ear, preferably with a direct connect system.

Clinical Consideration: Programming
- Block the normal hearing ear when measuring Ts.
- Think outside the box, and don’t forget about ESRTs
Summary

Clinical Consideration: Spatial Hearing
- Incorporate some form of testing that takes spatial hearing into account.
- The children have unique goals.

Outcomes: Word Recognition
- Isolated speech understanding is improved with a CI

Outcomes: Localization
- Localization improves over time with CI use
- Many children are approximating skills of kids with normal hearing by 18 months

Outcomes: Hearing in noise
- Hearing in noise is generally better CI on vs CI off
- Spatial release from masking is becoming evident when noise is directed to either side by 12-months post op

Outcomes: Listening effort
- Parents report that their children are exuding less effort to listen over time with the CI.

Outcomes: Speech, Spatial, and Qualities data
- Parents report improvement over time in all subscales with CI use.
Contact Info

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