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- Call 800-753-2160 (M-F, 8 AM-8 PM ET)
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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.

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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.

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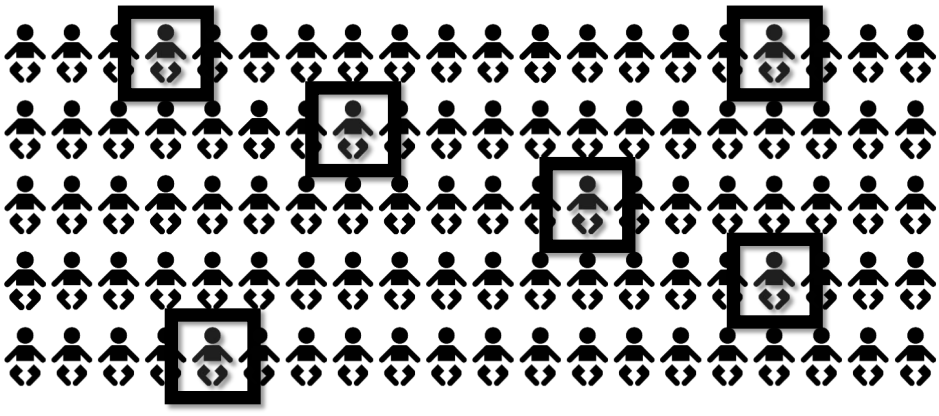

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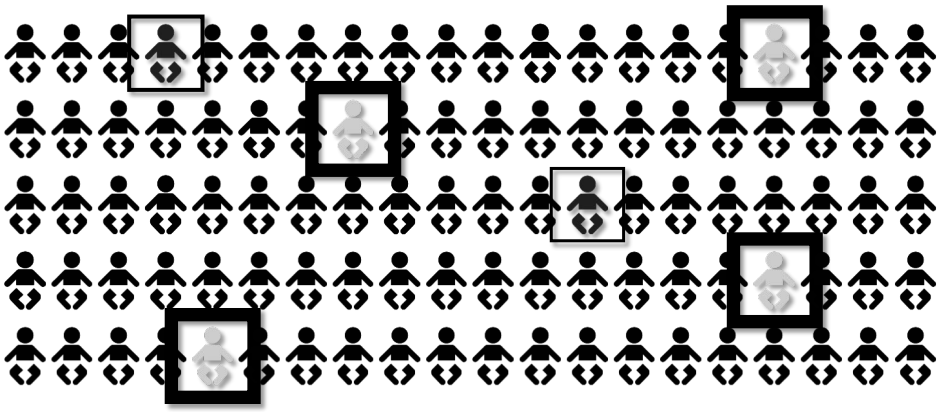


Characteristics of Children with Unilateral Hearing Loss

- 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)

Characteristics of Children with Unilateral Hearing Loss

- 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 (Bosman 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 ≥ 70 dB HL in one ear and normal hearing in the contralateral ear
- Aided CNC word score of ≤ 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

ID	Age at CI	Years Profound	Etiology	ID	Age at CI	Years Profound	Etiology
01	6.50	1.87	Infection	11	6.17	3.55	Unknown
02	6.42	1.37	Unknown	12	3.97	1.79	CMV
03	4.59	1.41	Trauma	13	4.90	1.26	Unknown
04	6.13	6.18	Mondini	14	5.46	4.99	Unknown
05	4.74	4.79	Waardenburg	15	5.58	0.79	Unknown
06	12.85*	2.78	Unknown	16	3.79	3.83	Unknown
07	4.00	4.04	Mondini	17	5.45	5.49	Unknown
08	6.50	4.62	Unknown	18	3.58	3.63	Unknown
09	6.50	6.62	Unknown	19	3.93	3.96	Unknown
10	7.09*	2.38	CMV	20	3.61	3.66	CMV

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

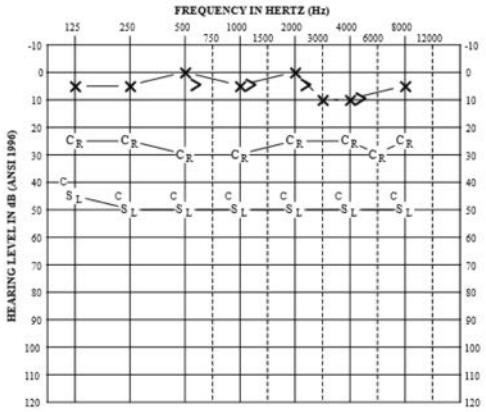
Detection Thresholds

Detection Thresholds

Detection Thresholds

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

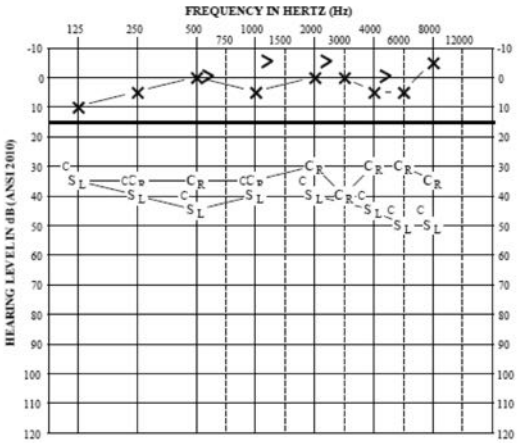
CR: Plug and muff thresholds, with CI processor



Detection Thresholds

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

CR: Plug and muff thresholds, with CI processor



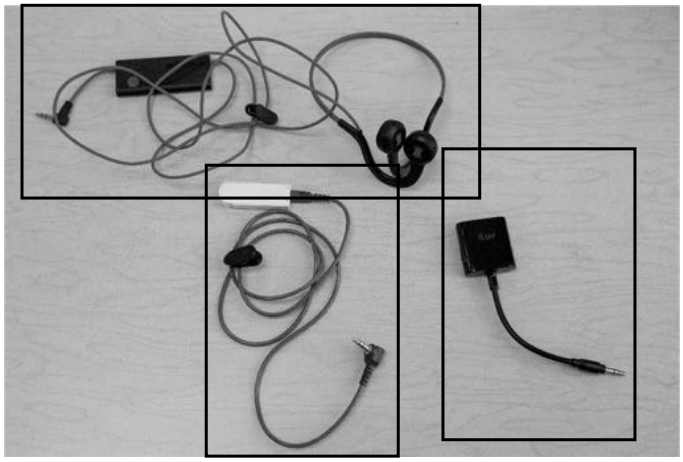
Outcomes

The Good Stuff

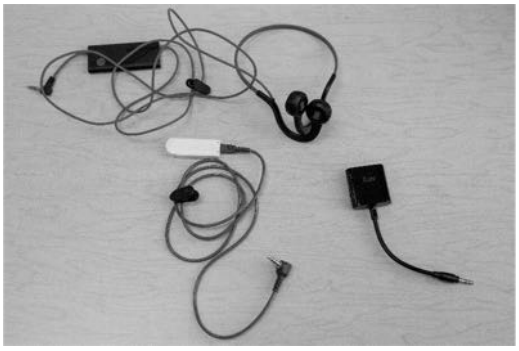
QoL and Inventory Outcomes

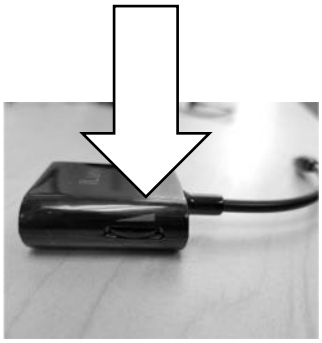
- Just 12-month data
- Peds-QL Fatigue Scales
  - Parent and Child ratings
- Pediatric SSQ
  - Listening Effort Subscale

Word Rec Setup



Word Rec Setup





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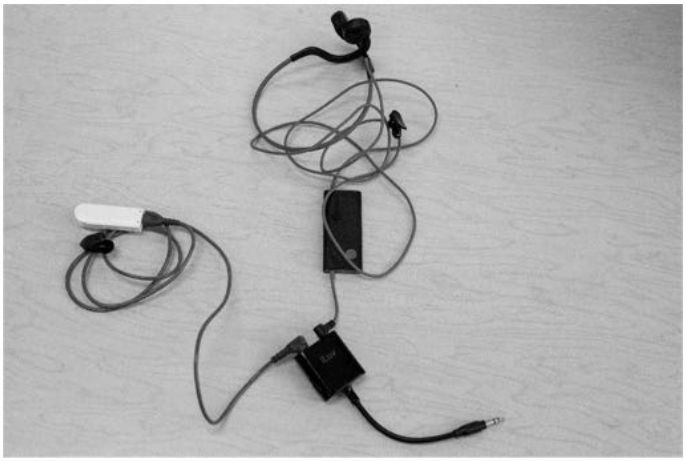
Word Rec Setup



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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)

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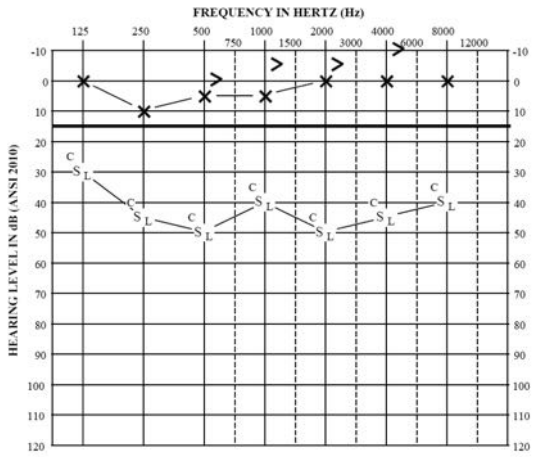
Plug and muff concerns



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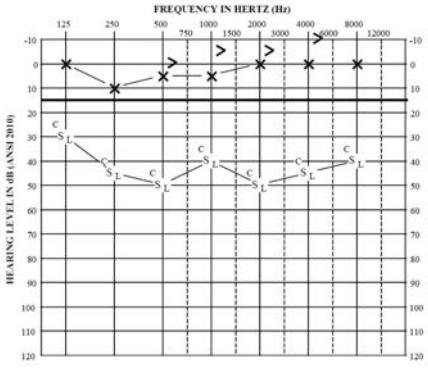
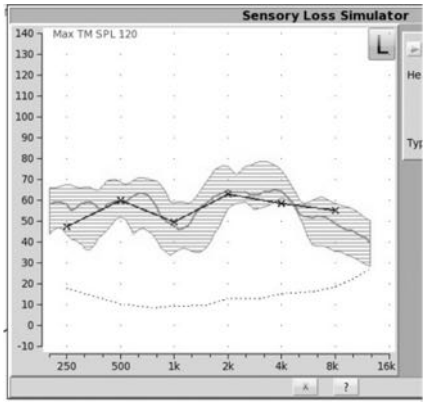
Plug and muff concerns





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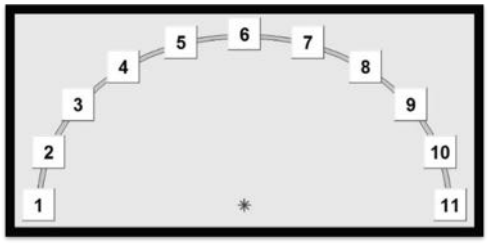
Localization

Localization



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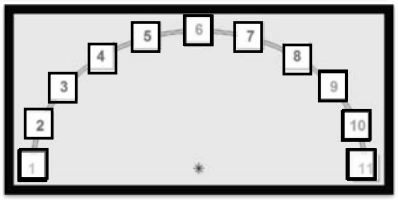

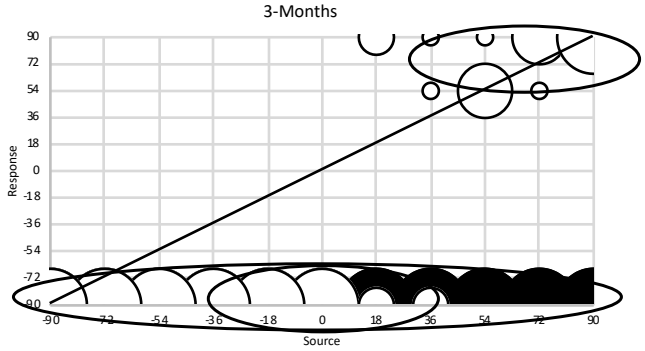

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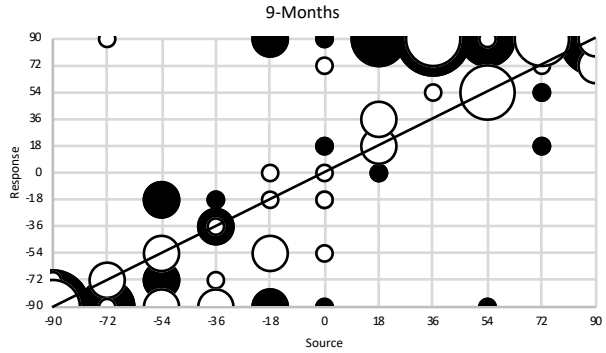

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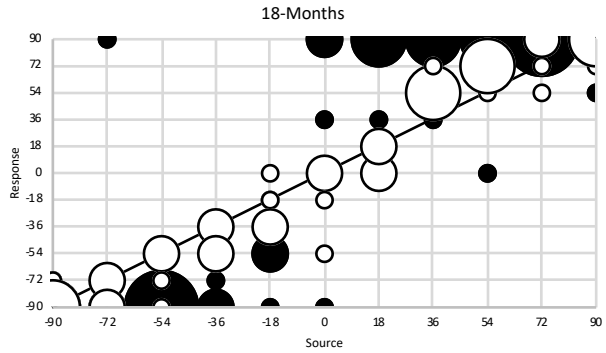
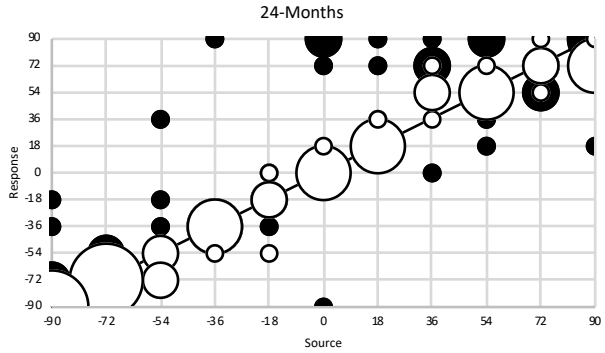
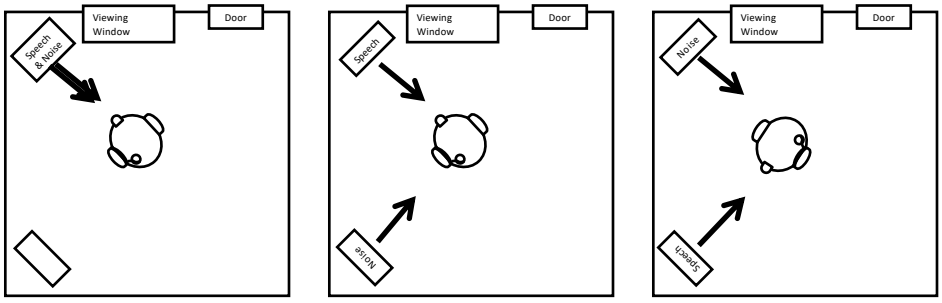


Localization



Speech Perception in Spatially Separated Noise



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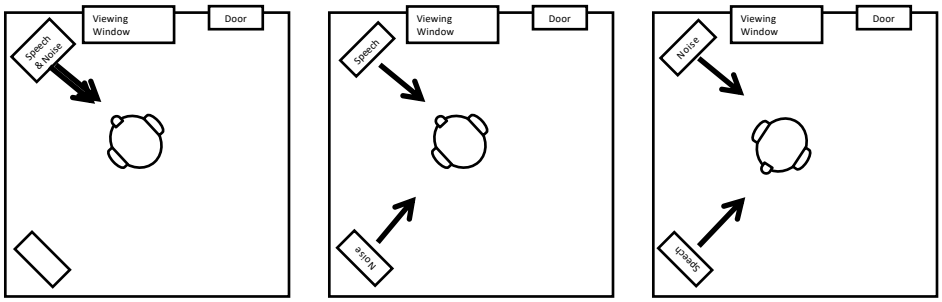

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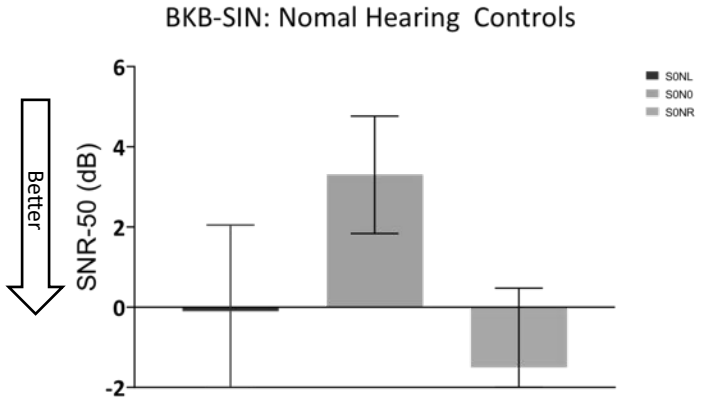
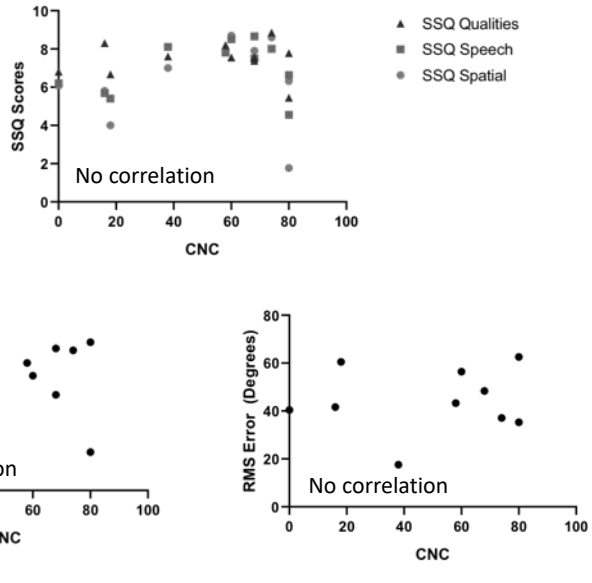
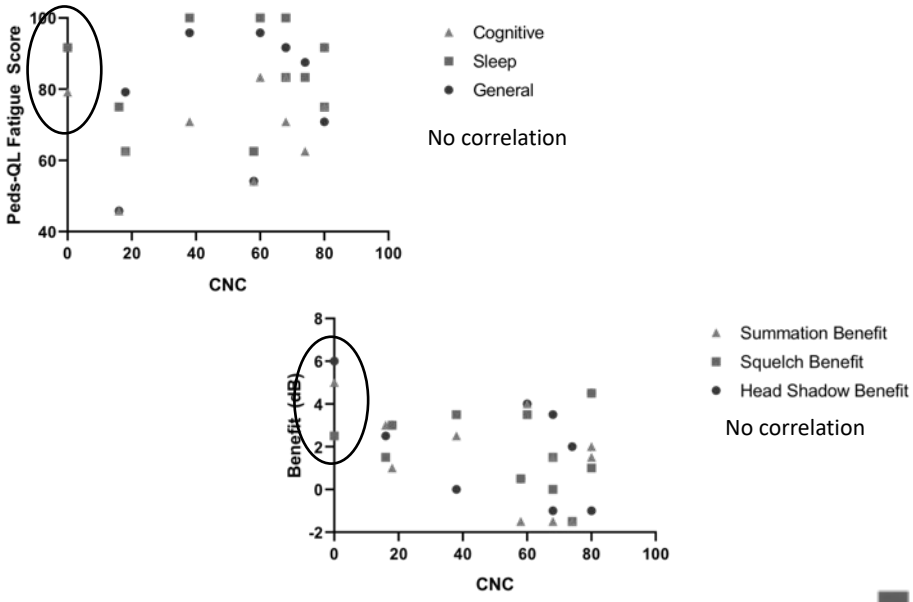


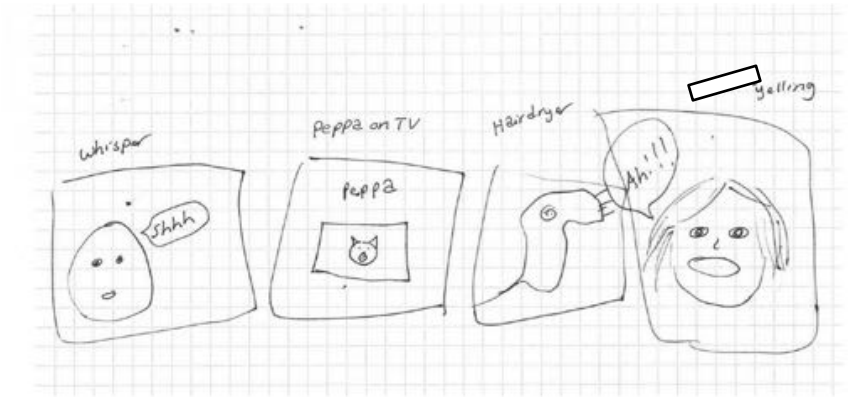
Children with CIs and UHL need to set their own targets!





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

Summary

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.

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Contact Info

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