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Cochlear Nerve Deficiency: Cochlear Implant or Auditory Brainstem Implant?

Holly FB Teagle, AuD

Lillian Henderson, MSP, Shuman He, PhD, Lisa Park, AuD, Jennifer Woodard, AuD, Erika Gagnon, AuD,

Matt Ewend, MD, Carlton Zdanski, MD, H. Pillsbury, MD,

Craig A. Buchman, MD

the little place for big miracles of sound & speech

THE CHILDREN'S COCHLEAR IMPLANT CENTER AT UNC

Cochlear Nerve Deficiency

- Small or absent auditory nerve on high resolution imaging
- Identified among patients with normal inner ear morphology, severe inner ear malformations, narrow internal auditory canals or completely absent cochlea
- Surgical options:
  - Cochlear Implant
  - Auditory Brainstem Implant

Children's Cochlear Implant Center at UNC

1014 children, 1567 ears since 1991

Cochlear Nerve Deficiency & CI

50 children with imaging and results

- Age at implant 1-15 years
- Years of use <1 – 18 years

2012 Pediatric ABI Feasibility Study

5 children

- Age at implant 2.5 – 5.5 years
- Years of use 1 - 2.5 years

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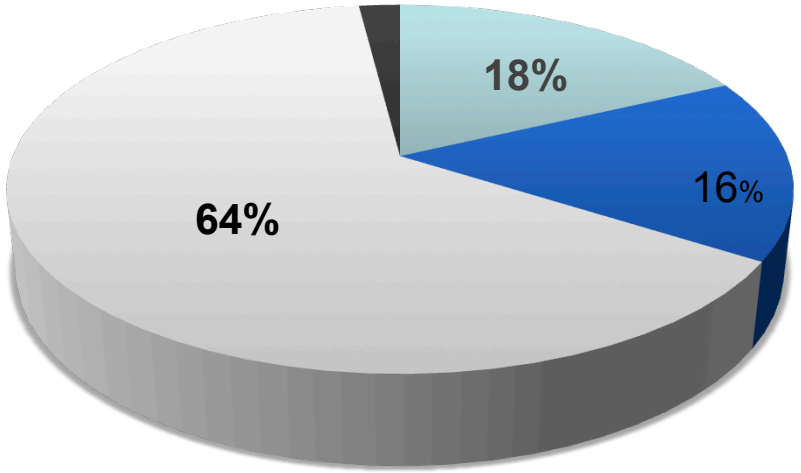
Children's Cochlear Implant Center at UNC
CI with CND N=50



- No Malformation 18%
- Cochlear Malformation Only 16%
- Cochlear/Vestibular Malformation 64%
- Vestibular Malformation Only 2%

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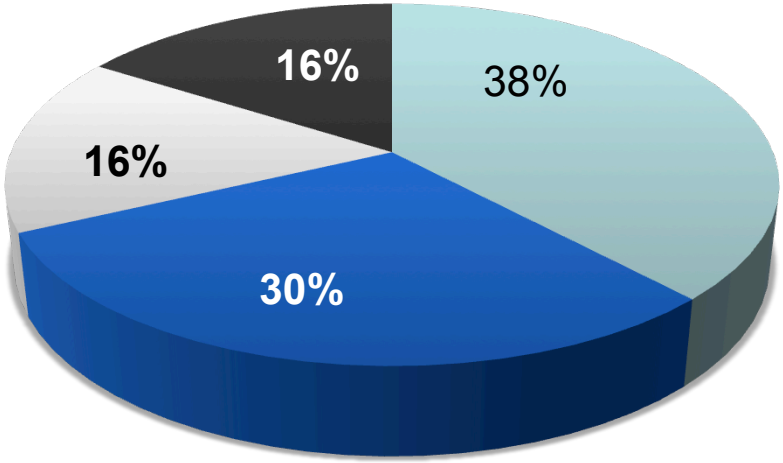
CI with CND N=50



- Non User 38%
- Sound Detection only 30%
- Pattern and Duration 16%
- Limited Open Set 16%

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Limited Open Set Performers CI-CND

	Age at CI	Years use	Imaging/Syndrome		Open Set	Com Mode	Device/
S1	5.5	18.5	C & V	no	36%	TC	Cochlear
S2	2.3	16	C & V	no	24%	TC	Med EL
S3	3.1	12	C & V	no	20%	TC	Cochlear
S4	1.7	6.5	C only	no	28%	TC	Cochlear
S5	1	2.5	C & V	VACTERL	52%	LSL	Med El
S6	4.7	8	C & V	no	36%	CS	Cochlear
S7	15	7.5	None	Yes /unknown	36%	TC	Cochlear
S8	2.3	8	C & V	Yes/unknown	45%	CS	Cochlear

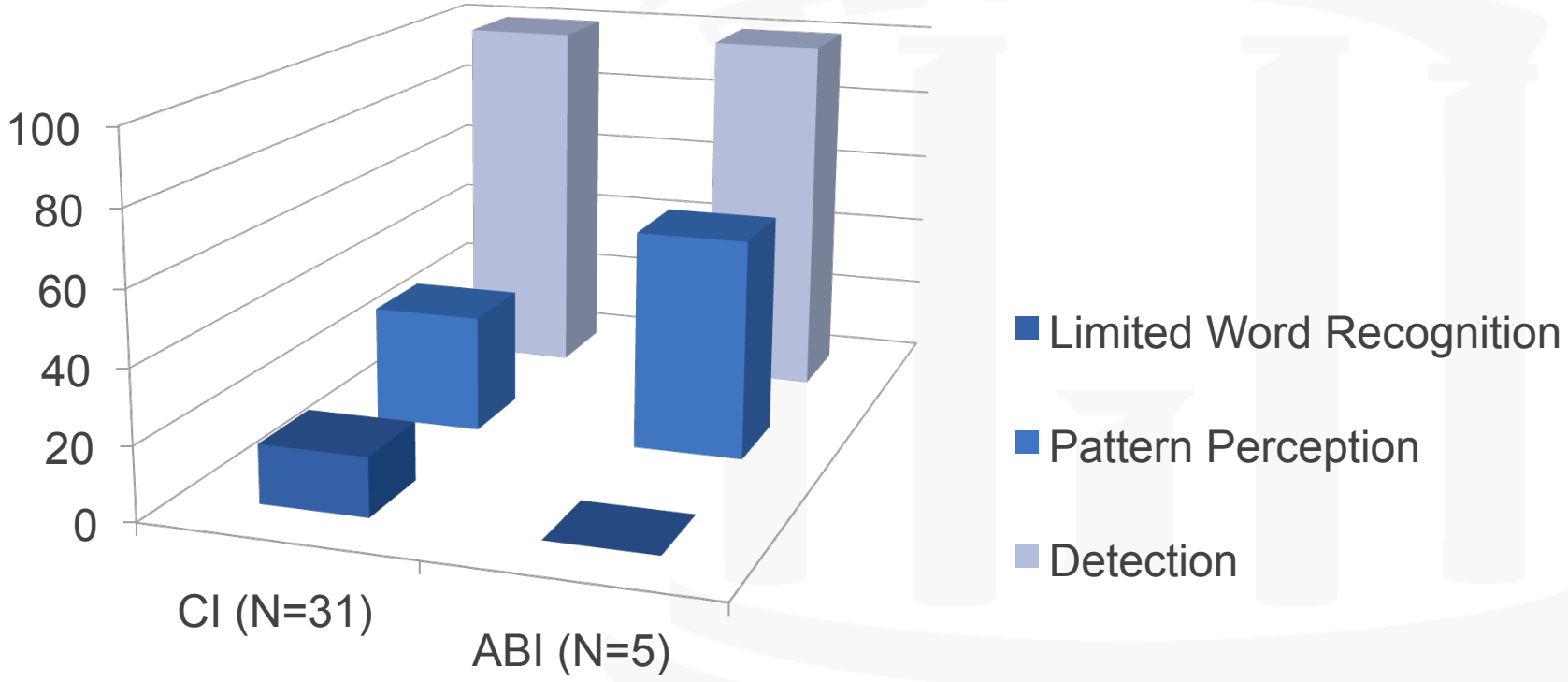
Demographics ABI Recipients

	UNC1	UNC2	UNC3	UNC4	UNC5
Previous CI	Yes	No	No	Yes	Yes
Age at ABI	3.3	2.5	3.5	5.5	2.1
Gender	M	F	M	F	F
Side	L	L	R	R	R
Etiology	CHARGE	Michele Aplasia	CHARGE	Unknown Normal	Unknown Normal
Com Mode	Cued Speech	ASL	SEE	SEE	ASL

Outcomes ABI Recipients

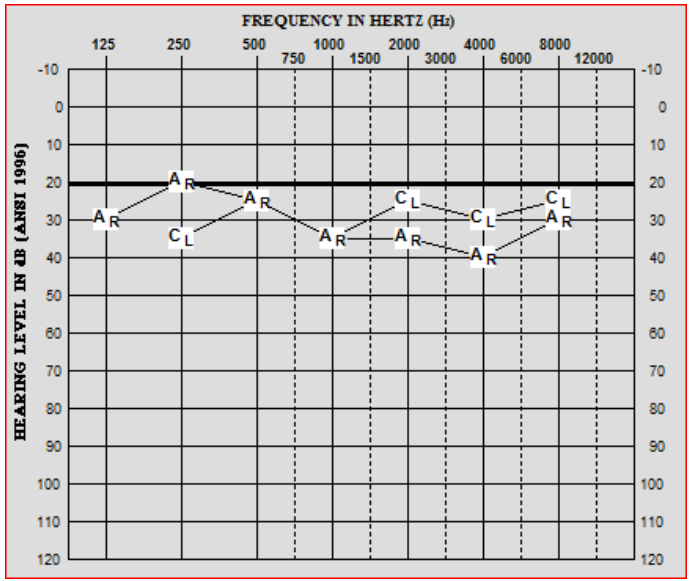
	UNC1	UNC2	UNC3	UNC4	UNC5
Duration of use (yrs)	2.5	2	1.5	1	1
PTA	30	48	30	35	35
SDT	15	45	20	20	30
IT-MAIS	65%	12%	53%	70%	5%
ESP Pattern	66% Standard	CNT	92% Low Verbal	50% Standard	CNT
ESP Word	66% Standard	CNT	25% Low Verbal	45% Standard	CNT

CI v ABI Auditory Skills

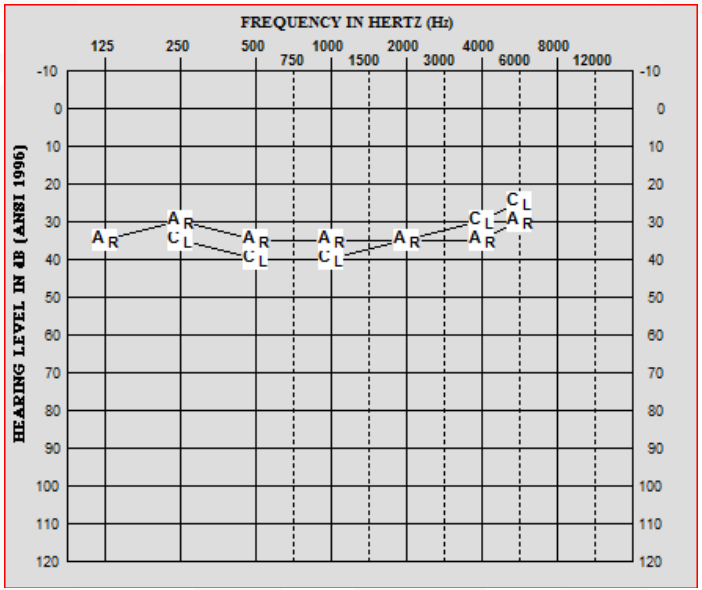


ABI and CI

1 year post ABI



CI LE @ 12 months/ABI @ 5.5 years



CI LE @ 18 months/ABI @ 2.1 years

Summary

- CND diagnosis requires a combination of imaging modalities
  - MRI +/- CT
- Benefit from a CI can not be predicted by imaging
  - ~1/3 discontinue use
  - ~2/3 continue to wear with consistency
    - All have sound detection
    - ~1/3 use suprasegmental cues to supplement communication
    - ~1/6 realize limited open set word understanding
- ABI (after 2.5yrs)
  - All have sound awareness
  - suprasegmental cues in some
  - open set word understanding in none

Conclusions

- A CI should be explored before ABI
- For children with CND using CI or ABI, auditory skills develop to a lesser extent and over a longer period of time compared to traditional CI recipients
- Visual support of language strongly recommended for all, along with auditory therapy

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