Changes in Cochlear Implant Candidacy and Future Innovations

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AND COMMUNICATION DISORDERS

Introductions

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Professor

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Session Outline

- A brief history of CIs
- Expanded CI candidacy criteria:
  - Age at implant
  - Degree of hearing loss
- What is on the Horizon?
- Conclusions and Questions/Discussion

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Part 1

A Brief History of Cochlear Implants – The "Firsts"

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Early 1800's

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1972

First successful single-channel CI (FDA approval 1984)

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1977

Electrical Stimulation with Multichannel Electrodes in Deaf Patients 131

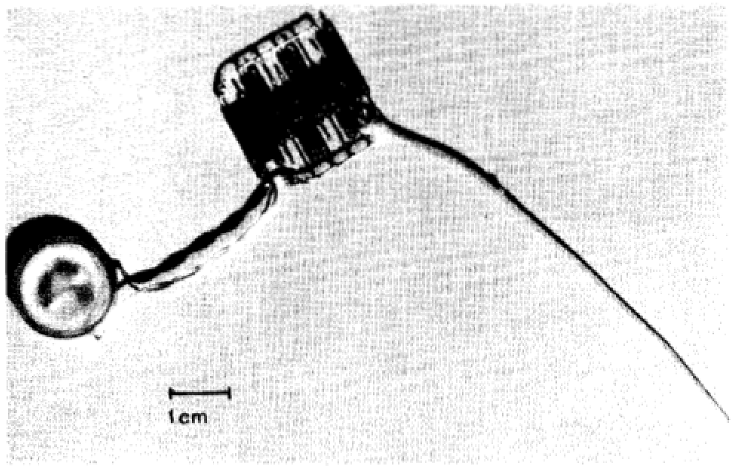

Fig. 3. Bipolar reed contact system before encapsulation.

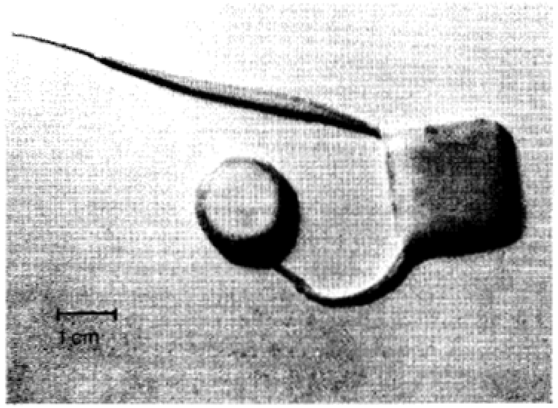

Fig. 4. Bipolar reed contact system after encapsulation.

First multichannel CI implanted in Vienna

(Burian et al. 1979)

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Outcomes of single-channel vs. multi-channel CIs

TABLE 2. *Open-set sentences > 6 months postimplant/reimplant*

	Single-channel		Multichannel		
	%	Time	%	Time	Strategy*
CID or Iowa sentences					
Patient 1	0	7 mos	45	6 mos	F0F1F2
Patient 2	4	5 yrs	78	6 mos	SPEAK
Patient 3	0	7 mos	9	6 mos	F0F1F2
Lindeman et al[16]	1	21 mos	31	18 mos	F0F1F2
Japanese sentences					
1, Gyo et al[6]	4	5 yrs	23	6 mos	SPEAK
2, Gyo et al[6]	0	7 yrs	92	6 mos	SPEAK

*F0F1F2 and SPEAK refer to the speak processing strategy used by the multichannel device.

(Rubinstein et al. 1998)

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Outcomes of single-channel vs. multi-channel CIs

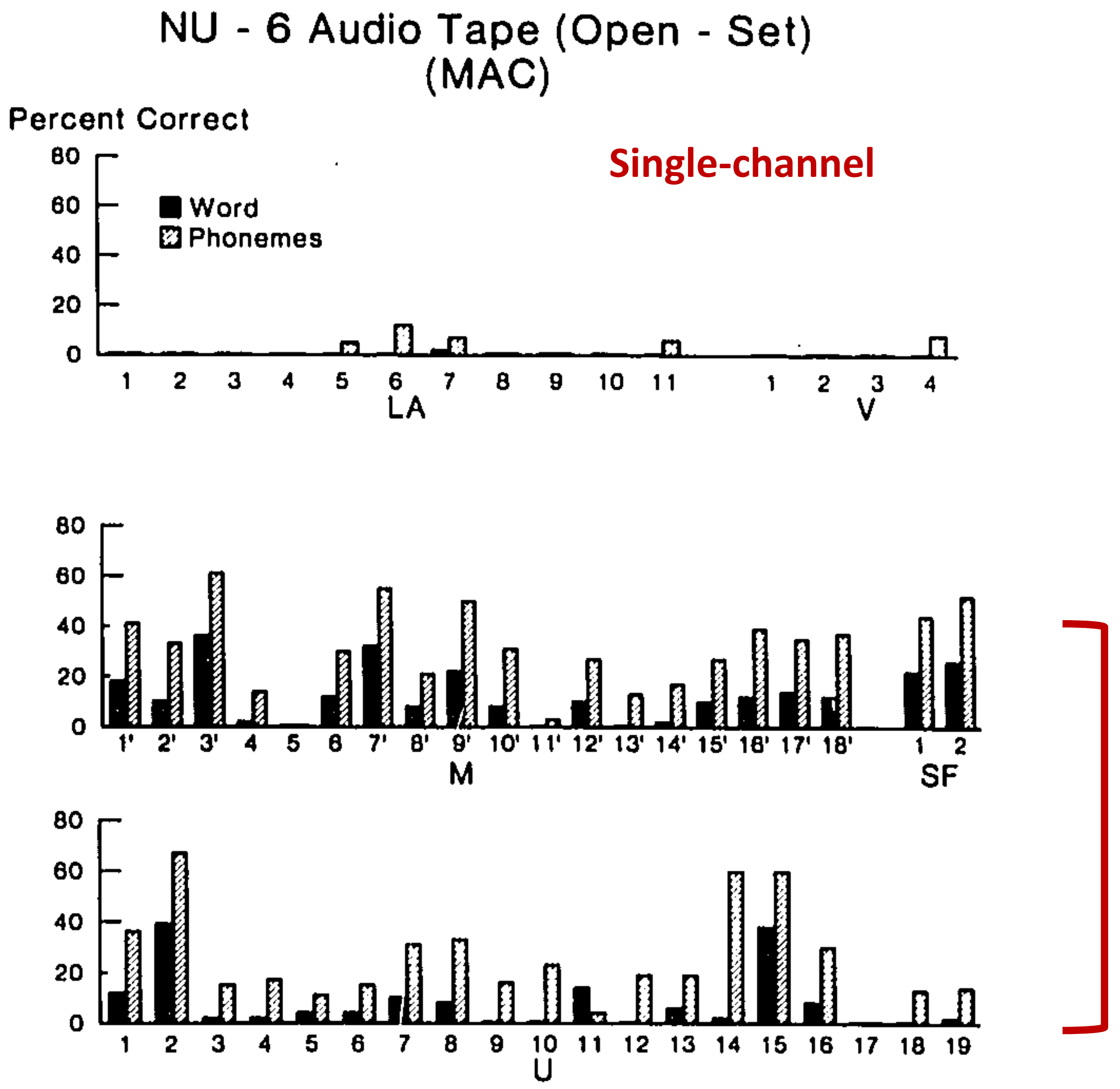

Fig. 4. Performance scores—NU-6 (MAC Battery) Sound-Only. LA = Los Angeles implant, V = Vienna implant, M = Melbourne implant, (prime indicates F_1F_2 speech processor), U = Utah implant, SF = San Francisco implant.

(Tyler et al. 1988)

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1996

First bilateral CIs implanted in Vienna

(Helms et al. 1997)

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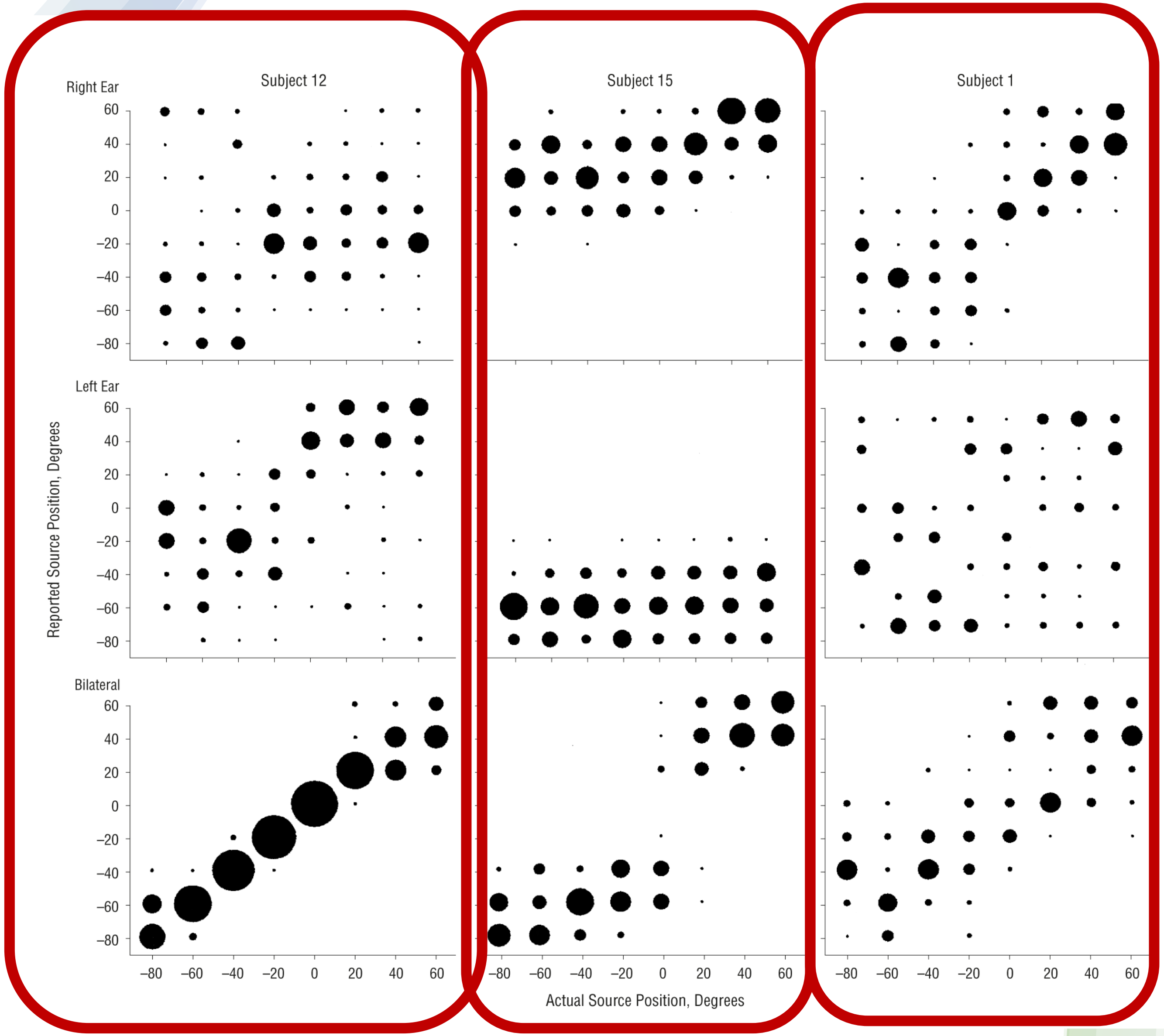
Bilateral CI Outcomes: Localization Patterns

(Litovsky et al. 2004, Fig. 1)



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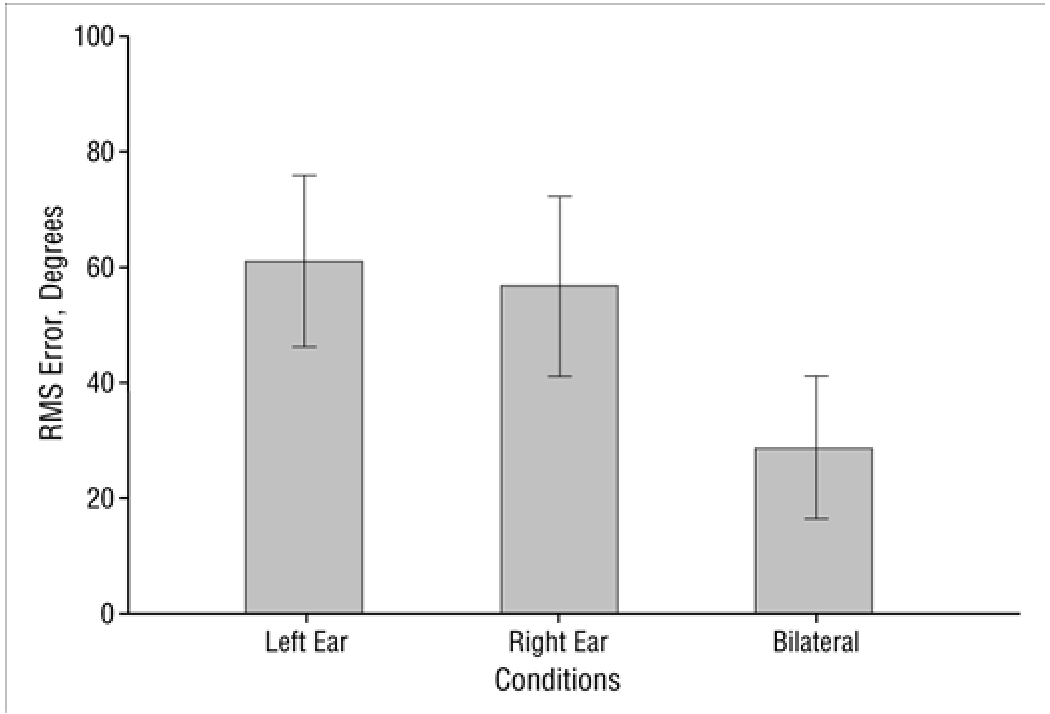
Bilateral CI Outcomes: Localization Patterns

(Litovsky et al. 2004, Fig. 2)



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Bilateral CI Outcomes: Speech Recognition in Noise

(Wolfe et al. 2007, Fig. 5)

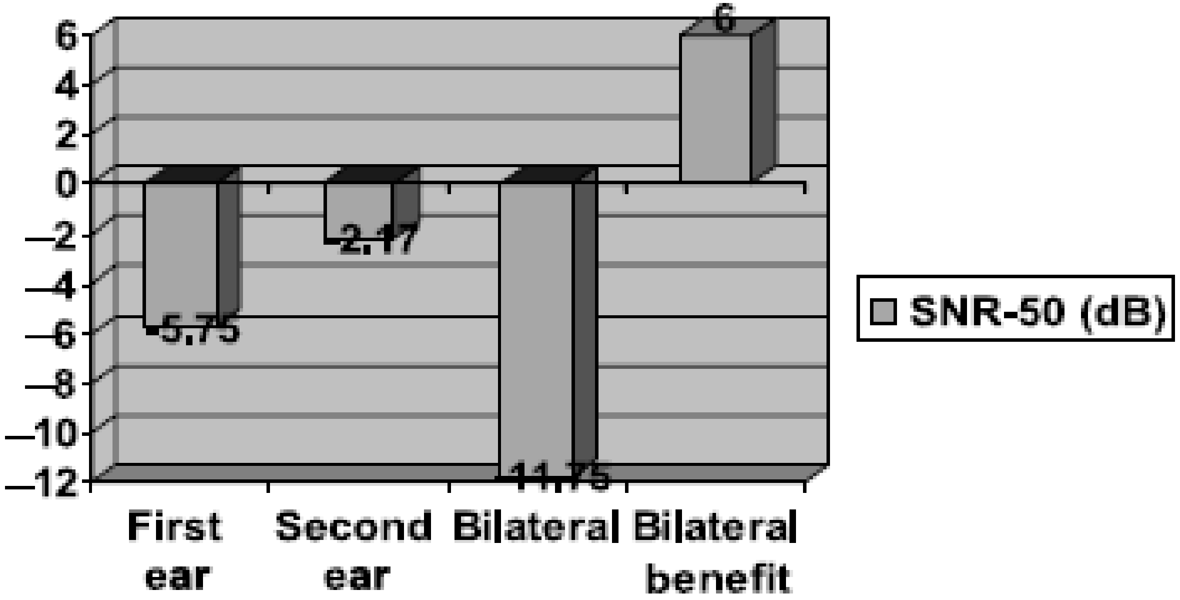


FIG. 5. Mean results for speech recognition in noise assessment. The SNR_50 is provided for three conditions: (A) when only the first cochlear implant is used; (B) when only the second cochlear implant is used, and (C) when both cochlear implants are used. Bilateral benefit is defined as the difference in SNR-50 between the first ear condition and the bilateral condition.

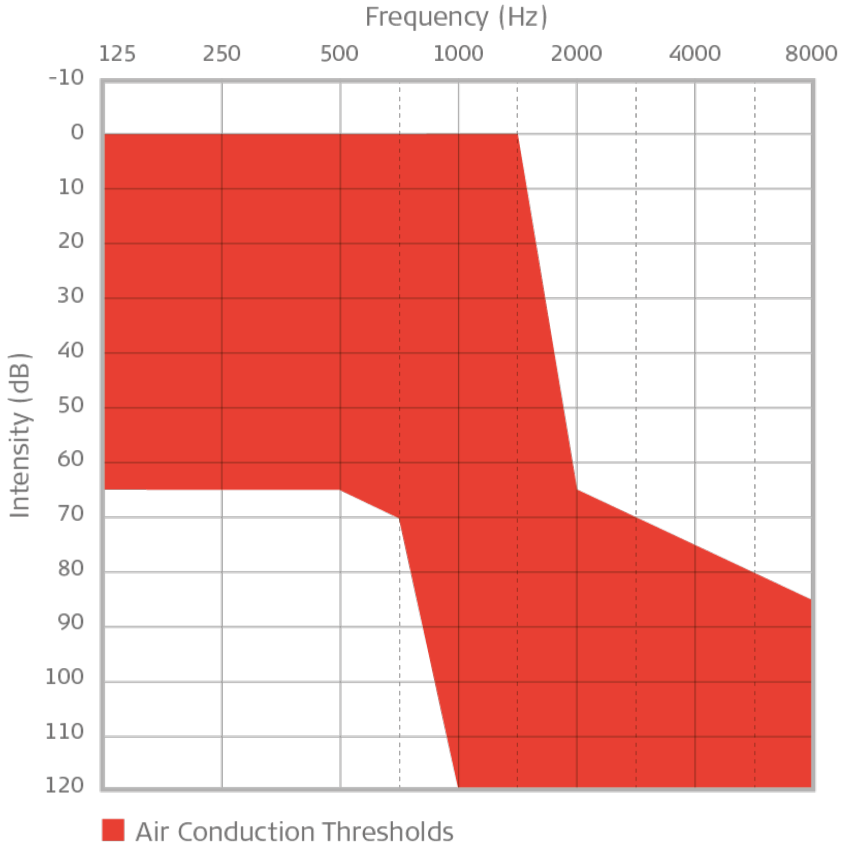
1999

Electric-Acoustic Stimulation of the Auditory System

New Technology for Severe Hearing Loss

C. von Ilberg[a] J. Kiefer[a] J. Tillein[b] T. Pfenningdorff[a] R. Hartmann[b]
E. Stürzebecher[a] R. Klinke[b]

Departments of [a]Otorhinolaryngology and [b]Physiology, Johann Wolfgang Goethe University, Frankfurt/Main, Germany

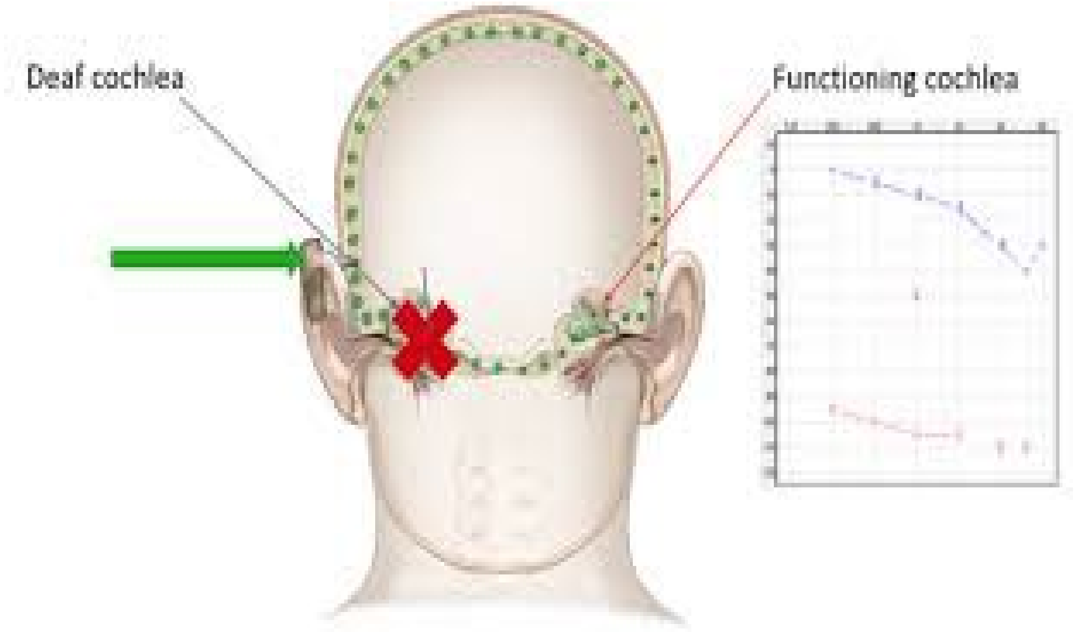


First electroacoustic stimulation (EAS) device implanted

(von Ilberg et al. 1999)

First CI surgery for tinnitus in single-sided deafness

(van de Heyning et al. 2008)



2003

Annals of Otology, Rhinology & Laryngology 117(9):645-652.

Incapacitating Unilateral Tinnitus in Single-Sided Deafness Treated by Cochlear Implantation

Paul Van de Heyning, MD, PhD; Katrien Vermeire, PhD; Martina Diebl, MS; Peter Nopp, PhD; Ilona Anderson, BA; Dirk De Ridder, MD, PhD

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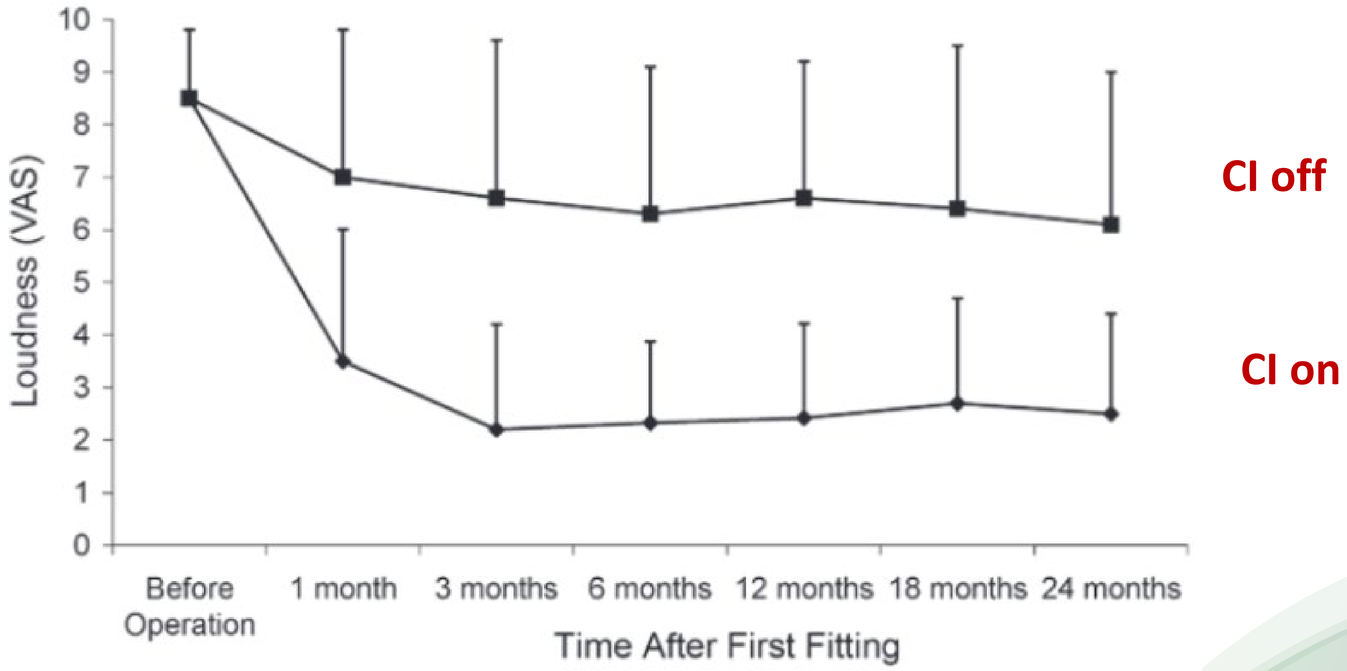
First CI surgery for single-sided deafness

(van de Heyning et al. 2008)

2003



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First totally implantable CI

(Briggs et al. 2008)

2005

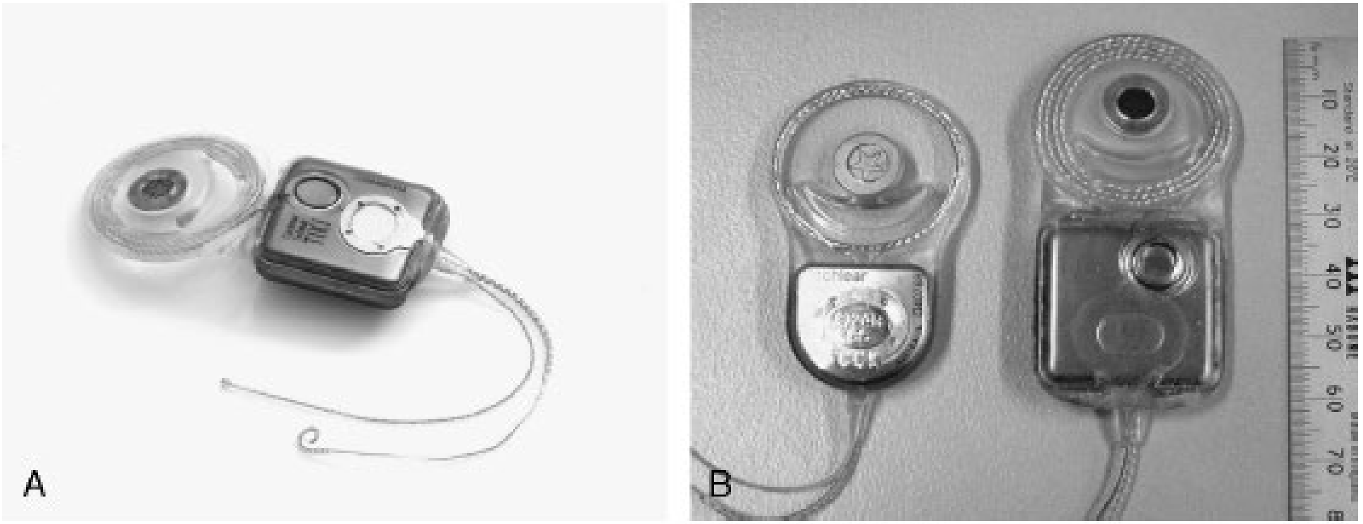

FIG. 1. *A*, The TIKI implant showing electrode array, microphone, receiver coil, magnet, and extracochlear plate electrode and ball electrode. *B*, With the CI24RE Freedom for comparison.

Otology & Neurotology, Vol. 29, No. 2, 2008

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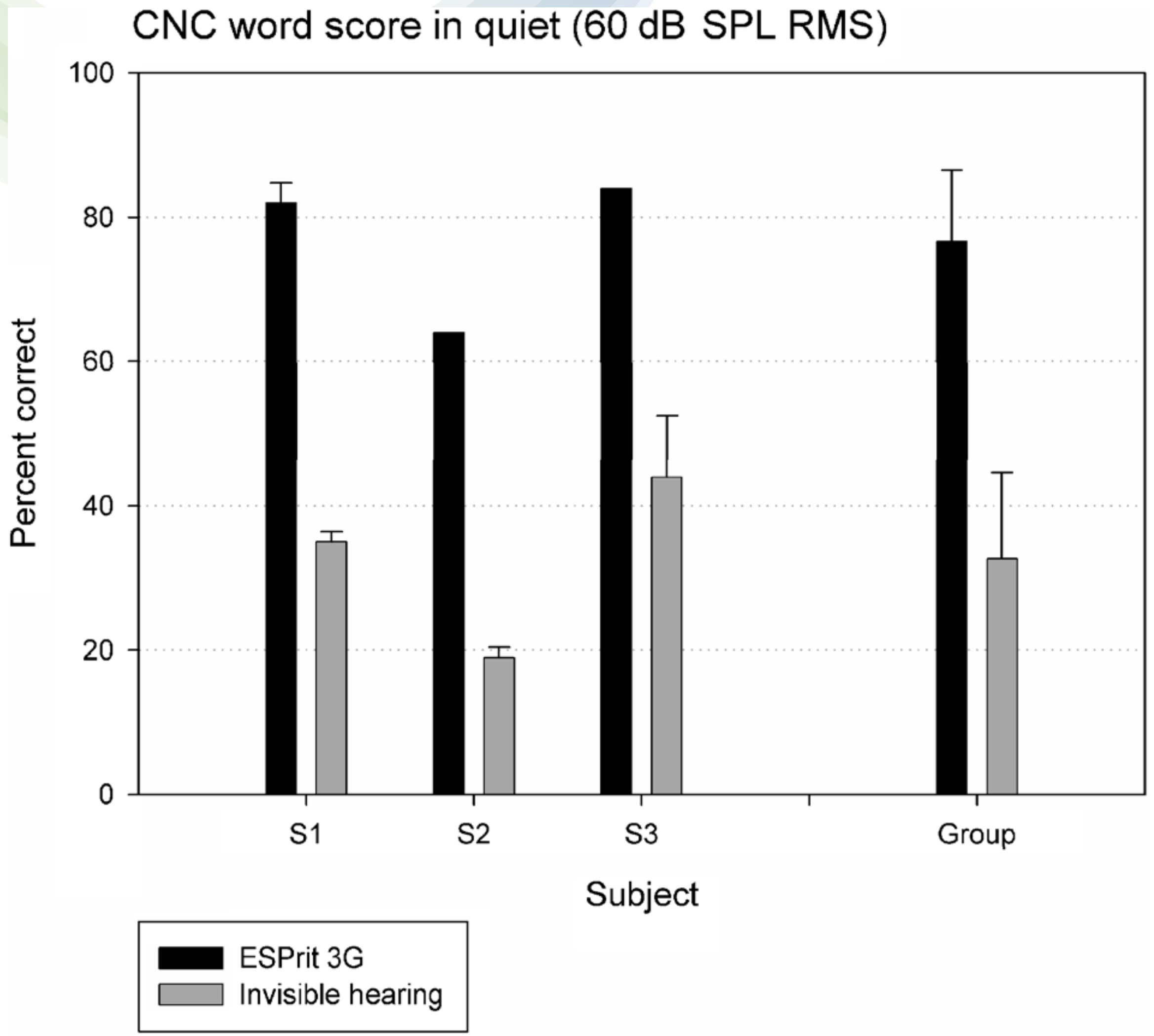
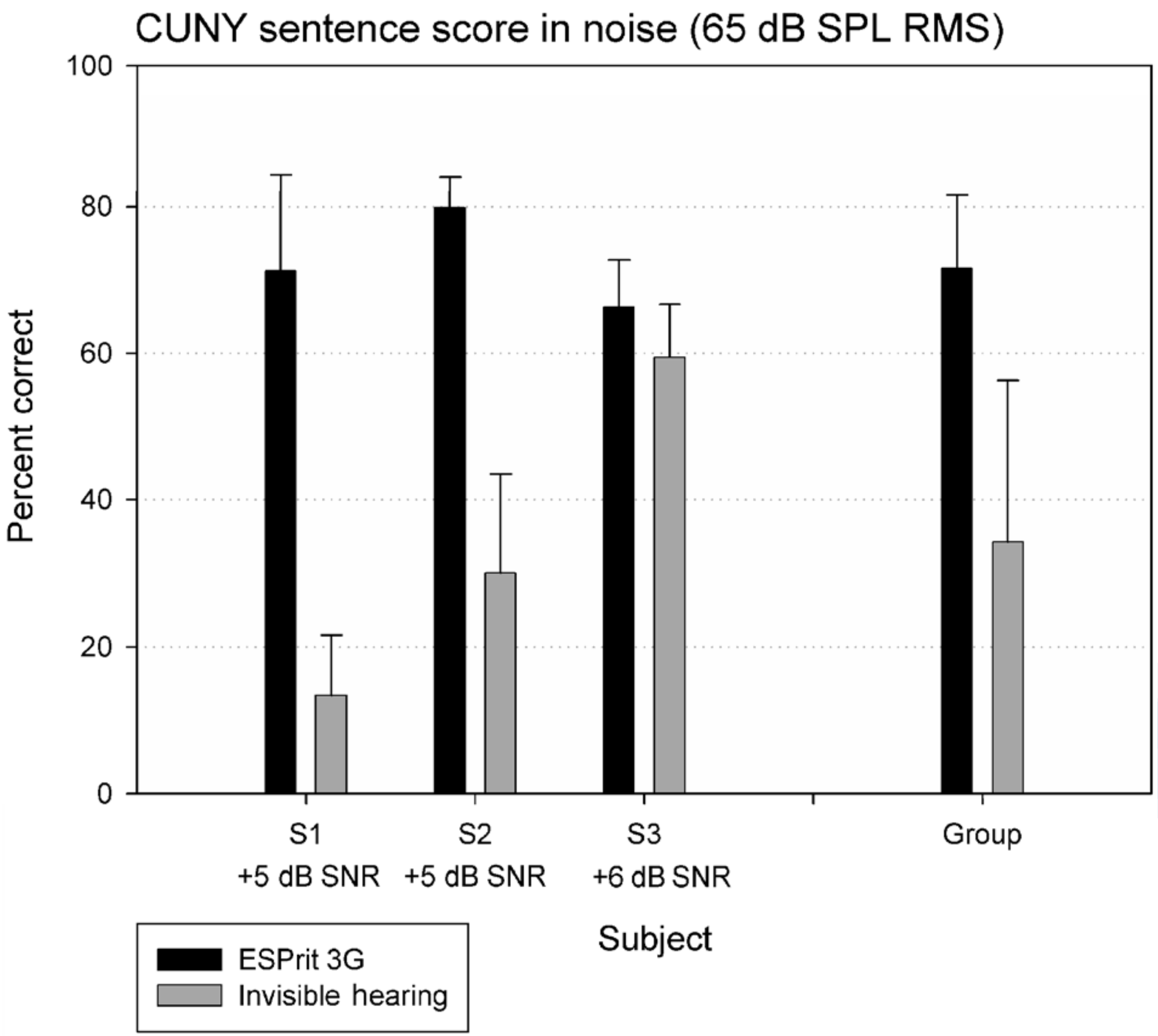
2005

First totally implantable CI

(Briggs et al. 2008)





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Part 2A

Expanded Cochlear Implant Candidacy Criteria: Age at Implant

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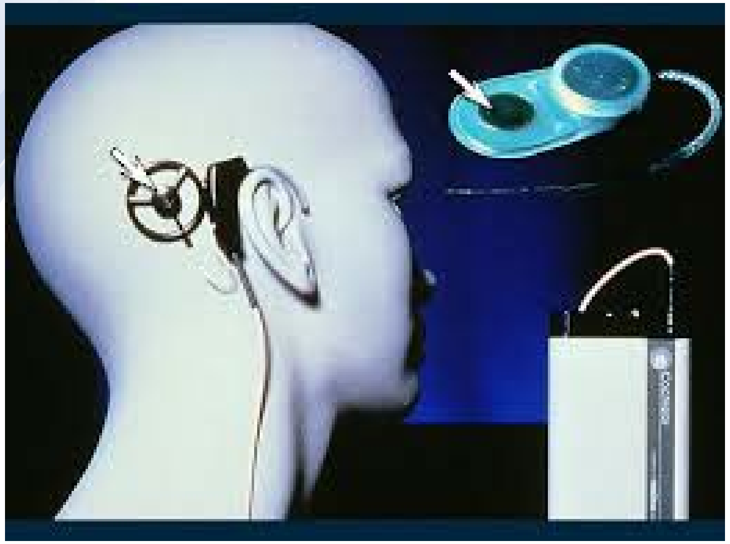


Nucleus 22 with WSP

1985

FDA approval of first multichannel CI for post-lingually deafened adults

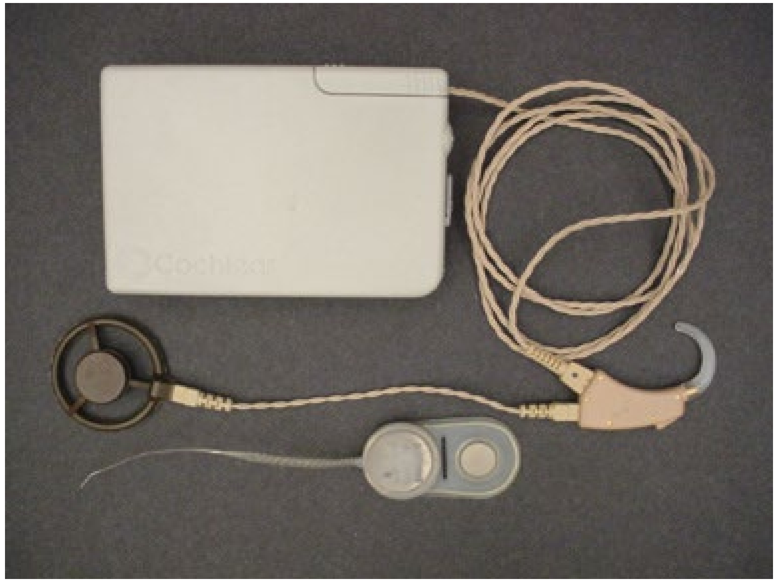
TABLE 1. CENTRAL INSTITUTE FOR THE DEAF EVERYDAY SENTENCE TEST

	Percent of Key Words Correct		
Patient	*F0/F2*	*F0/F1/F2*	*Difference*
1	54	90	+36
2	38	70	+32
3	31	54	+23
4	16	62	+46
5	10	26	+16
6	20	38	+18
7	44	100	+56
Mean	30.4	62.9	+32.4

$t = 5.79$, $df = 6$, p 0.01.

Dowell et al. (1987)

1990



Nucleus 22 with MSP

FDA approval for children aged 2 years and up

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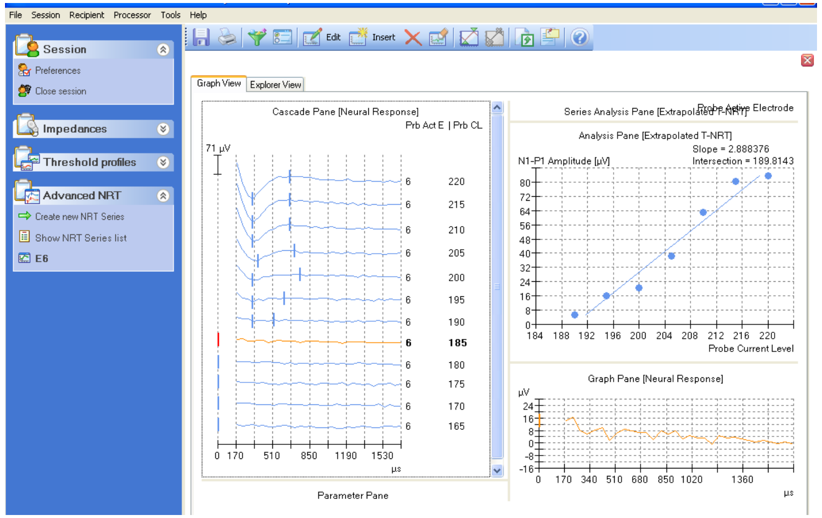
1998

Nucleus Esprit



Nucleus Neural Response Telemetry

- First BTE processor
- First CI with eCAP telemetry
- Implant age reduced to 18 months

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2000

Implant age for CI reduced to 12 months

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Cochlear®

Diagnosis and treatment | Products and accessories | Ongoing care and support | Your Cochlear stories

Cochlear receives FDA approval to lower the age of pediatric cochlear implantation to 9 months

Approval underscores necessity of earlier cochlear implantation for better hearing, speech and language outcomes in children born deaf

Centennial, Colo. **(March 18, 2020)** — Cochlear Limited (ASX: COH), the global leader in implantable hearing solutions, obtained U.S. Food and Drug Administration (FDA) approval to lower the age of cochlear implantation from 12 months to 9 months for children with bilateral, profound sensorineural hearing loss. This approval ensures children born deaf have earlier access to a cochlear implant which can provide them with the hearing capabilities to develop speech and language at a trajectory similar to their hearing peers.

2020

Implant age reduced to 9 months

https://www.cochlear.com/us/en/corporate/media-center/media-releases/2020/fda-ped-9-months#:~:text=(March%2018%2C%202020)%20%E2%80%94,bilateral%2C%20profound%20sensorineural%20hearing%20loss.

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Expanded CI Candidacy

- **1985**: Adults with bilateral profound SNHL
- **1990**: Addition of children 2 years and up (profound HL only)
- **1998**: Age lowered to 18 months (profound only)
- **2000**: Age lowered to 12 months (profound); allowed for severe-profound for 2+ years
- **2013**: First EAS device approved for adults with residual low frequency hearing
- **2019**: Inclusion of SSD/AHL for 5 years and up
- **2020**: Age lowered to 9 months (traditional CI)

Expanded CI Candidacy: 9 months

- 10 years of research to lower age from 2 years to 1 year
- 20 years to lower age from 1 year to 9 months
- Primary concerns:
  - Accurate estimates of hearing thresholds
  - Accurate estimates of functional outcomes with amplification
  - Safety (anesthetic/surgical risks)
  - Post-operative CI programming

Accurate Estimates of Acoustic Thresholds

- Tymps and reflexes
- ABR (exercise caution with ANSD!)
- OAE
- VRA starting ~6 months
- Etiology, imaging, genetic testing

Bottom line: Ensure we do not implant children who are not deaf!

Accurate Estimates of Functional Outcomes

- Can't use standard benchmarks of aided word recognition to assess HA benefit
- Can use IT-MAIS, LittlEars parent inventories

Surgical/ Anesthetic Risks

- No difference in the incidence of perioperative complications between children aged 1-12 months and those aged 1-5 years across various types of surgeries (Cohen et al. 1990)
- Similar findings for studies specific to CI surgery for <12 months vs. >12 months (e.g., Lesinski-Schiedat et al. 2004; Roland et al. 2009; Cosetti & Roland 2010; Chweya et al. 2021)
- Use of pediatric anesthesiologist significantly decreases risk of perioperative complications (Keenan et al. 1991)
- Challenges associated with anatomical differences require knowledgeable and experienced surgeons.

Cosetti & Roland (2010)

Post-Operative Programming

- Combined use of behavioral (VRA) and objective measures
  - Telemetry
  - Electrode impedance
  - eCAP
  - eSRT
  - eABR

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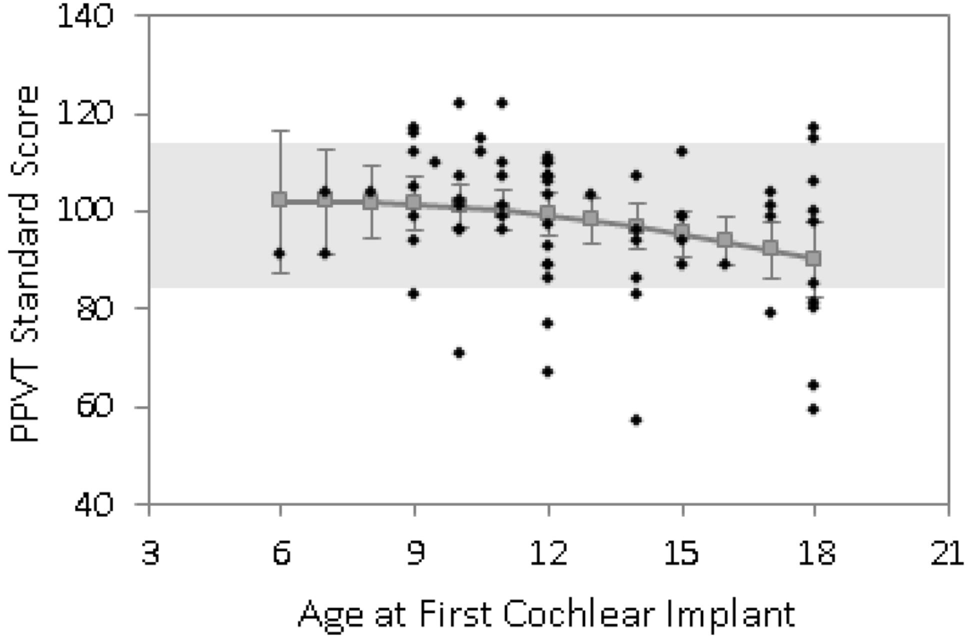
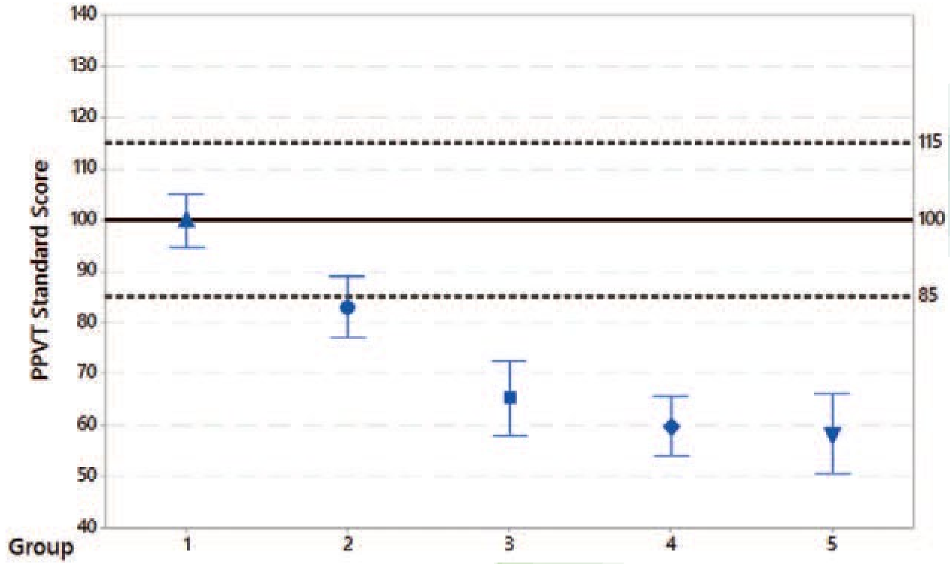
Evidence Supporting Younger Age of Implantation

- More cost effective for CI <12 months than for 12-23 months (Colletti et al. 2011).
- Higher scores on receptive and expressive spoken language outcomes for children implanted:
  - Between 12-24 months versus those implanted one year later (Miyamoto et al. 2008)
  - Between 6-11 months and those implanted between 12-18 months (Nicholas & Geers 2013; Dettman et al. 2016) or later.

Evidence Supporting Younger Age of Implantation

Vocabulary





Top: Nicholas & Geers (2013), Figure 1

Bottom: Dettman et al. (2016), Figure 4

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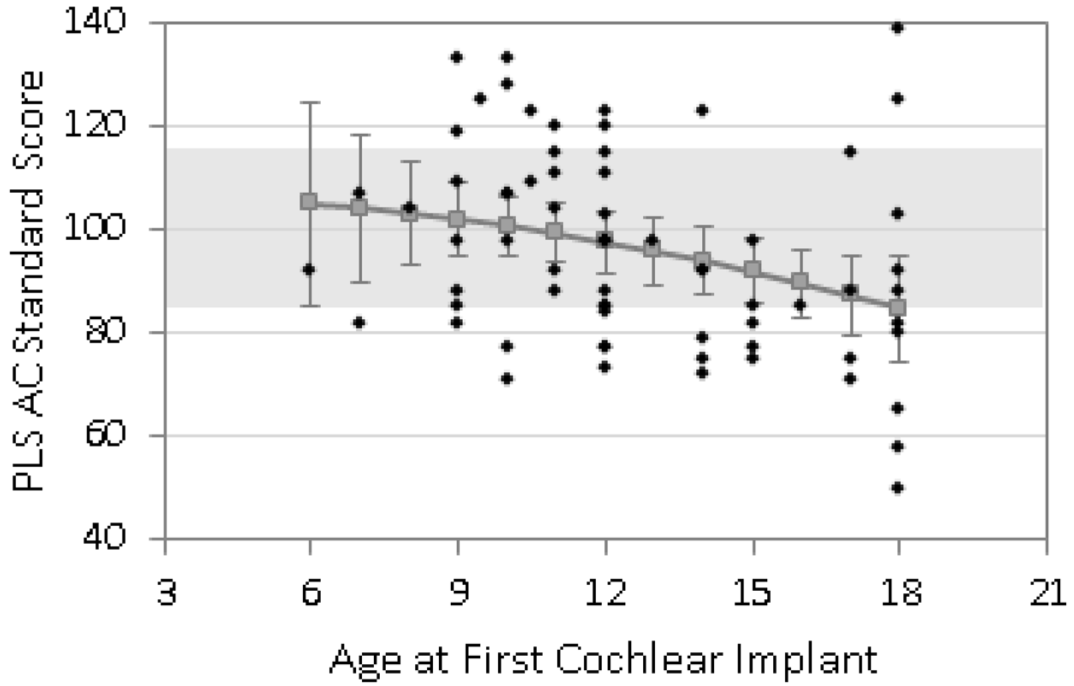
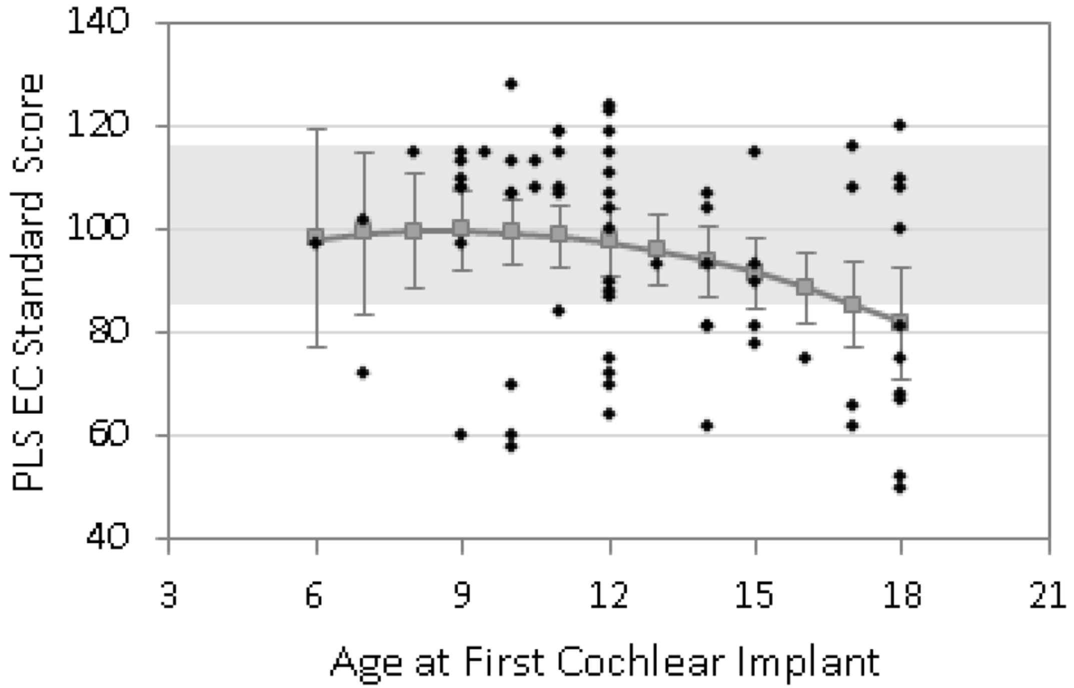
Evidence Supporting Younger Age of Implantation

Auditory Comprehension



Expressive Language

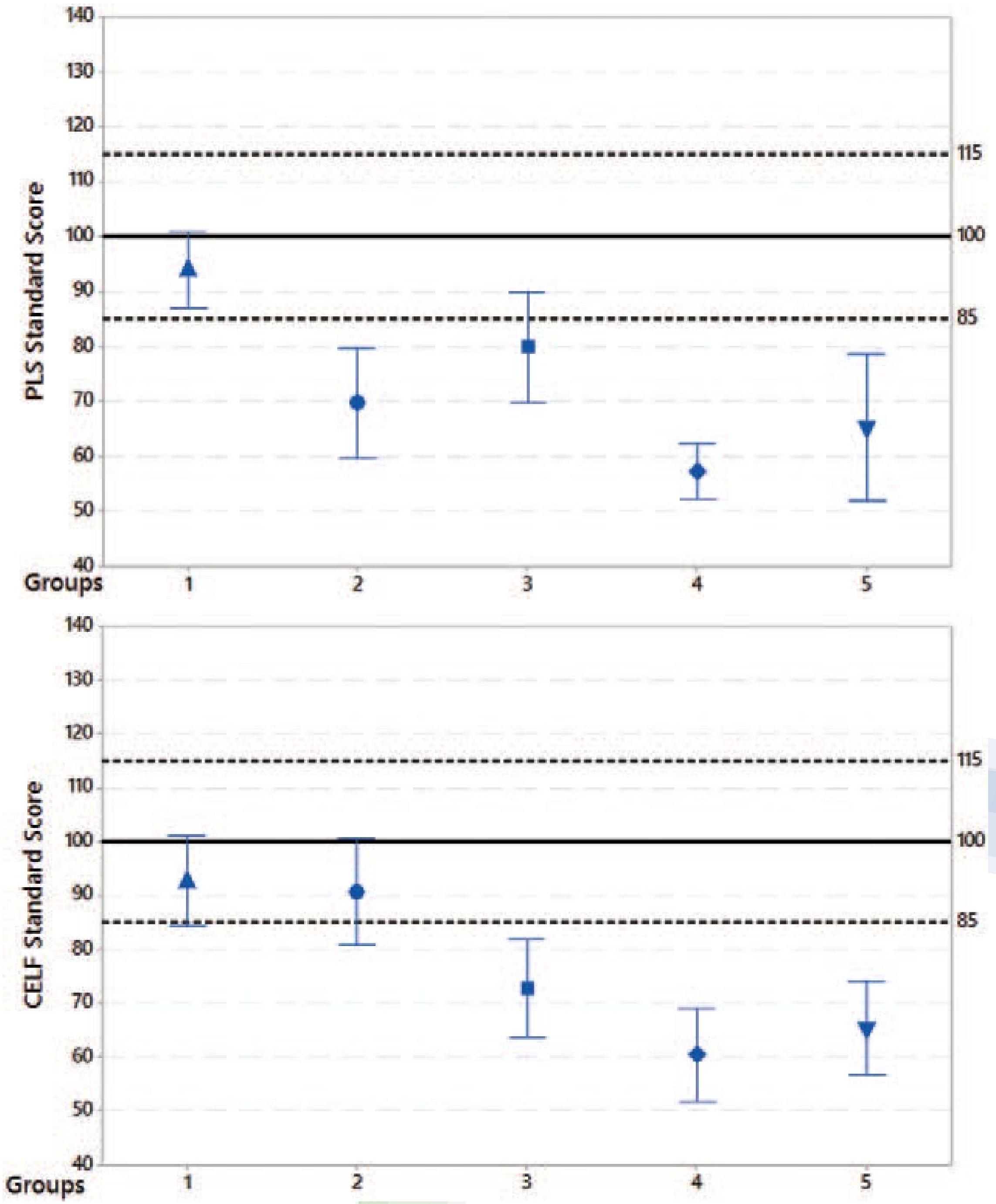


Nicholas & Geers (2013), Fig. 2 (top) & Fig. 3 (bottom)

Evidence Supporting Younger Age of Implantation

Preschool Language Scales

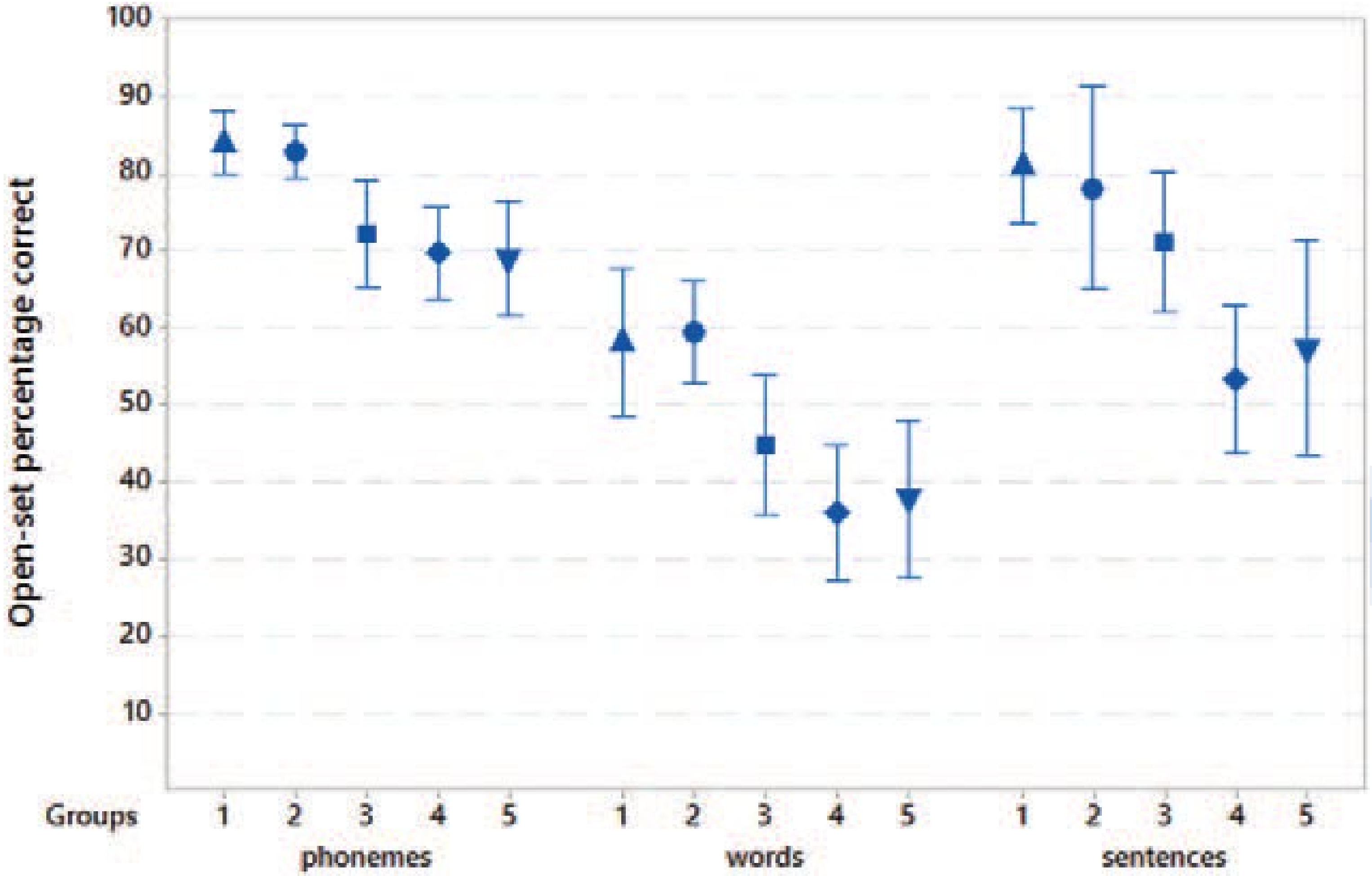
CELF-4



Dettman (2016), Fig. 3 (top) & Fig. 6 (bottom)

Evidence Supporting Younger Age of Implantation

Speech Recognition



Dettman (2016), Fig. 1

Part 2B

Expanded Cochlear Implant Candidacy Criteria: Degree of Hearing Loss

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Future Directions: Expanding Degree of HL

- Current Criteria for Adults:
  - Moderate-profound SNHL
  - ≤50% sentences ear to implant, ≤60% best aided

- Current Criteria for Children:
  - Profound SNHL <2 yrs, severe-profound SNHL >2 yrs
  - ≤30% words

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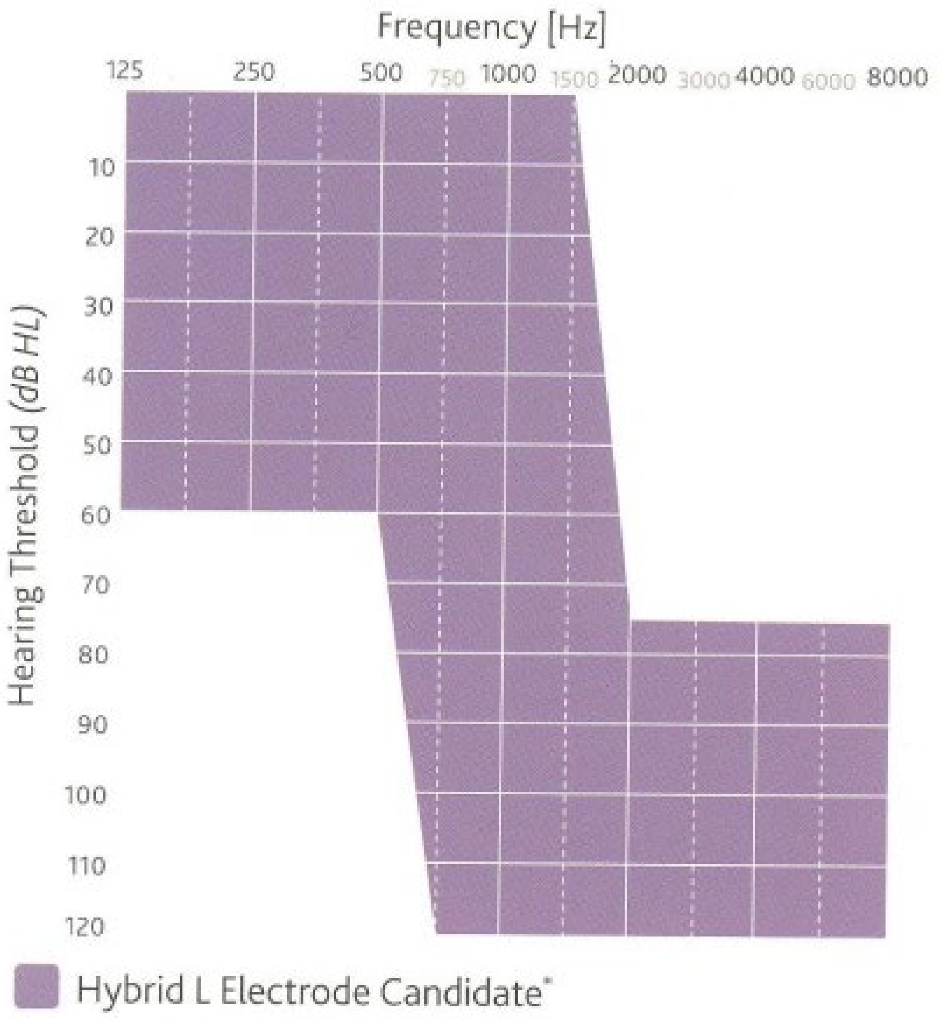
2013



FDA approval of EAS

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EAS Hearing Preservation

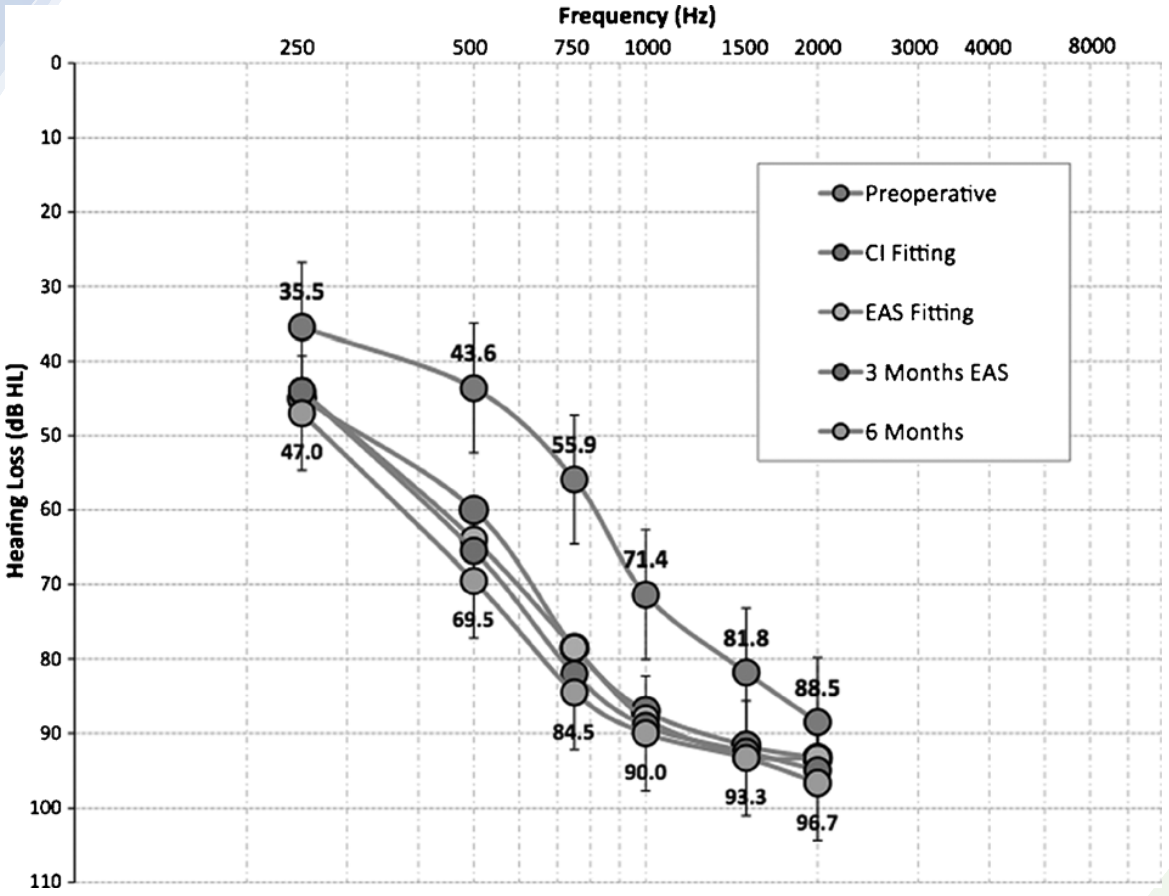

FIG. 1. Ipsilateral pure-tone audiometric data of the EAS group (n = 10) for various intervals before and after surgery. One subject lost hearing completely, and data from this subject have not been included in this graph.

Adunka et al. (2010)

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EAS Hearing Preservation

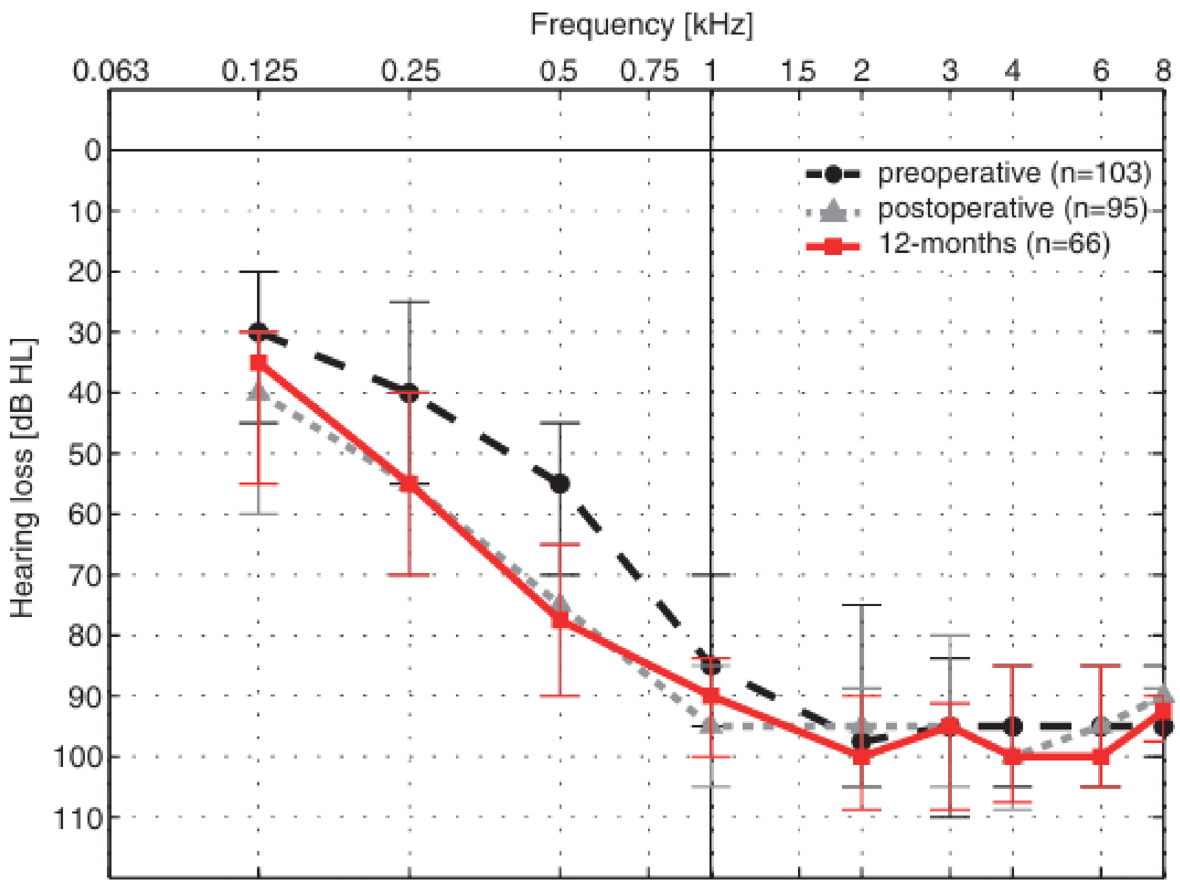


FIG. 1. Hearing threshold medians and interquartile ranges for the time intervals: preoperative (*black dashed line, circles*), postoperative (*gray dotted line, triangles*), and 12 months (*red solid line, squares*); n denotes number of subjects for each condition.

Helbig et al. (2016), Fig. 1

EAS Hearing Preservation

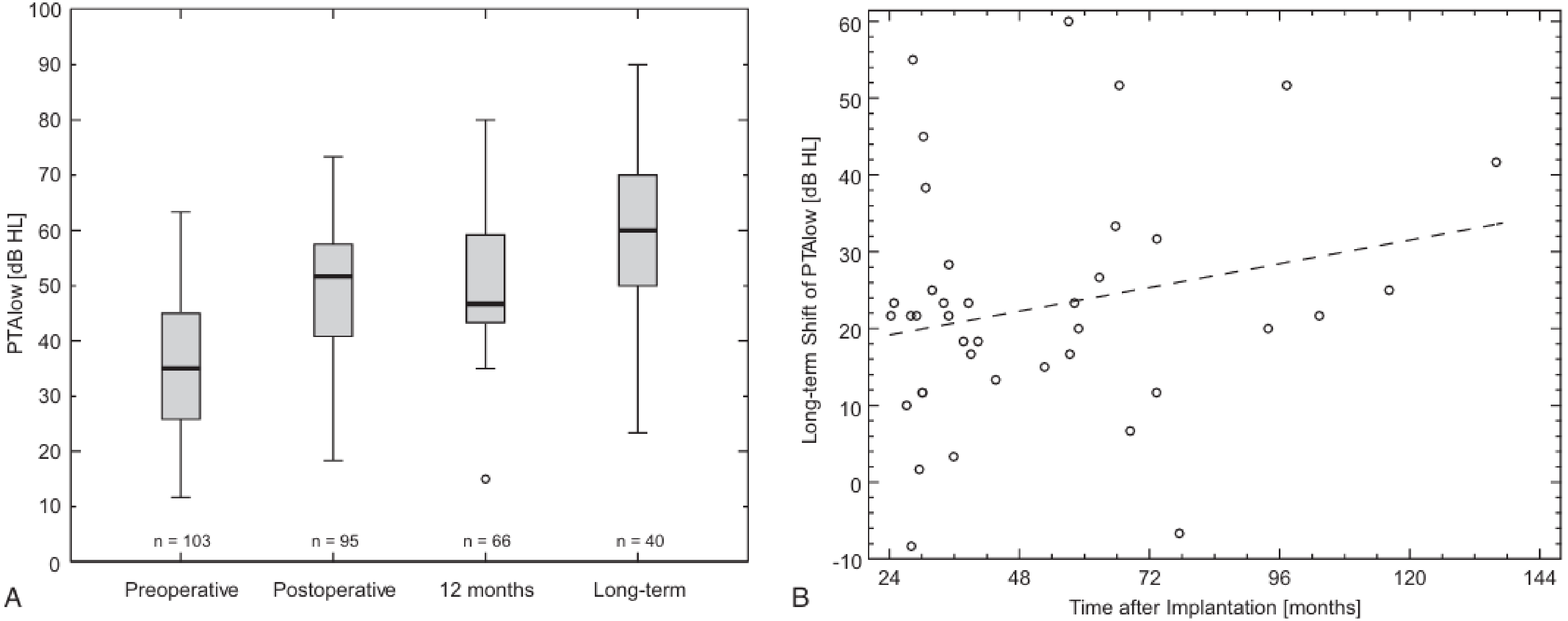


FIG. 2. Panel *A*: Pure-tone averages for low frequencies (PTAlow; mean of air conduction thresholds for 125 Hz, 250 Hz, and 500 Hz) shown as box plots (median, 1st and 3rd quartiles, minimum and maximum values, *circles* indicate outliers, n denotes number of subjects for each condition). Long-term is >24 months, mean, 51.4 months, range, 2–11 years. Panel *B*: Scatter plot of long-term (>24 months) shifts of pure-tone averages for low frequencies (PTAlow; mean of air conduction thresholds for 125 Hz, 250 Hz, and 500 Hz) with regard to preoperative PTAlow. Linear regression (*dashed line*, $y = 16.02 + 0.13x$, $R^2 = 0.053$) shows a trend of residual hearing deterioration for this population sample (n = 40).

Helbig et al. (2016), Fig. 2

EAS Performance

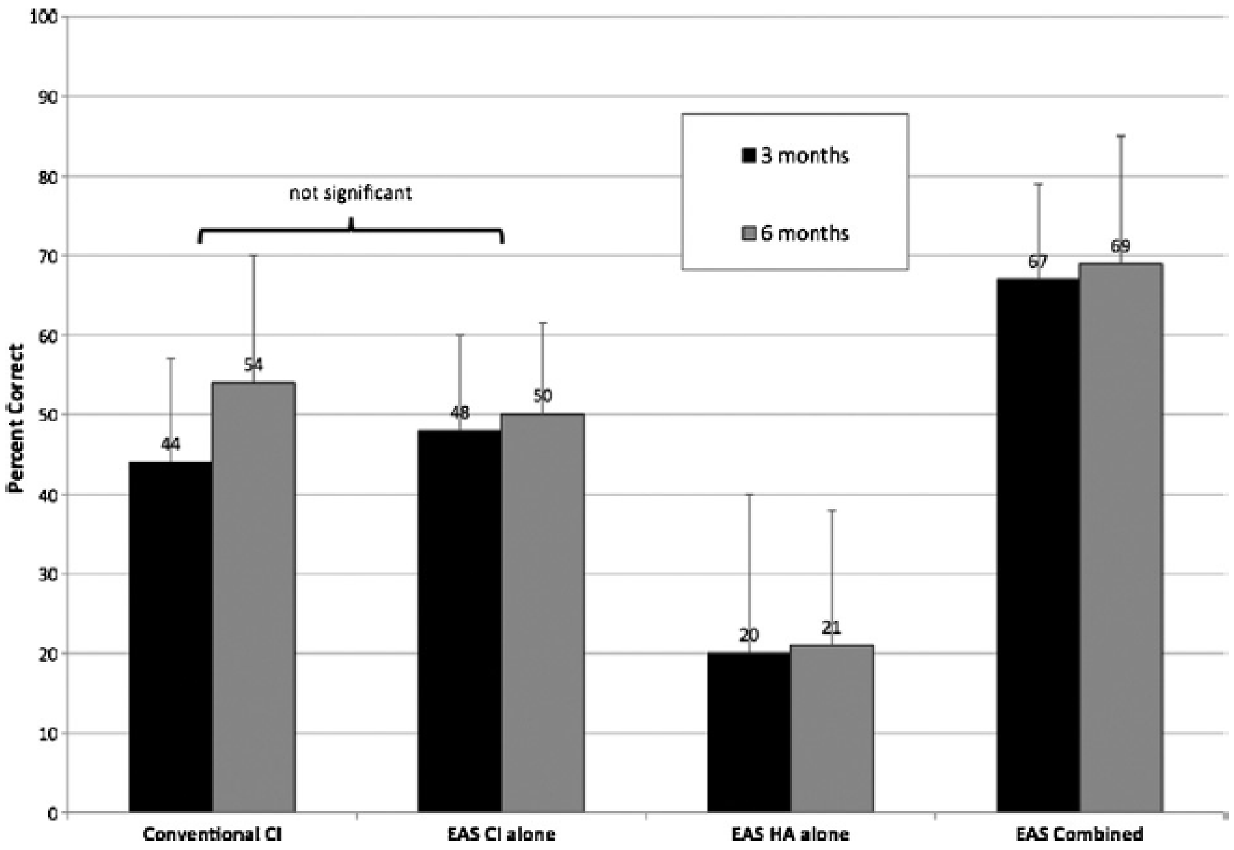
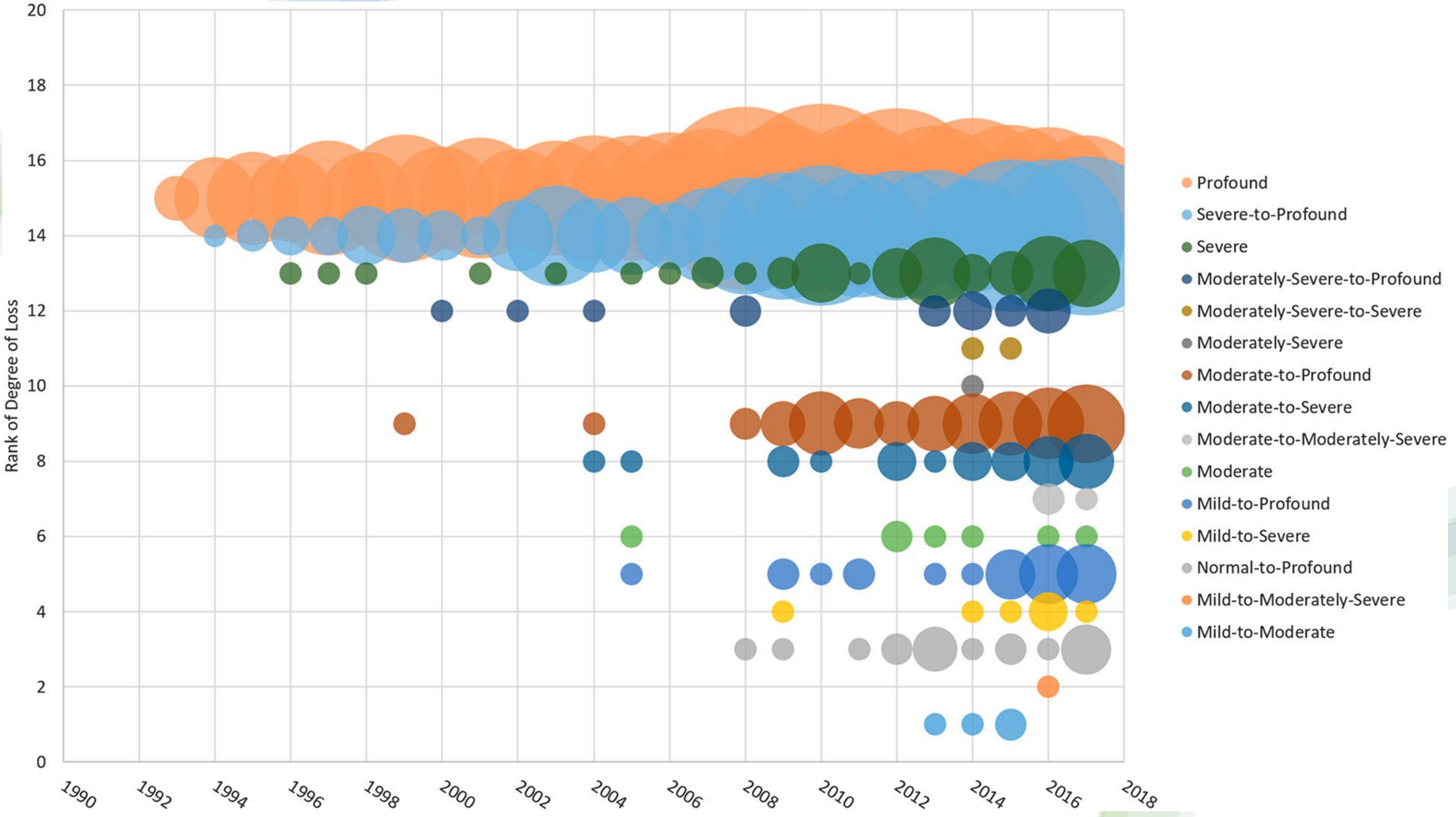


FIG. 3. CNC word scores 3 and 6 months after fitting for both groups. Differences with a *p* value of less than .05 are considered statistically significant.

Adunka et al. (2010)

Expanding Degree of HL: Children



Teagle et al. (2019), Fig. 5, N = 1,570 children

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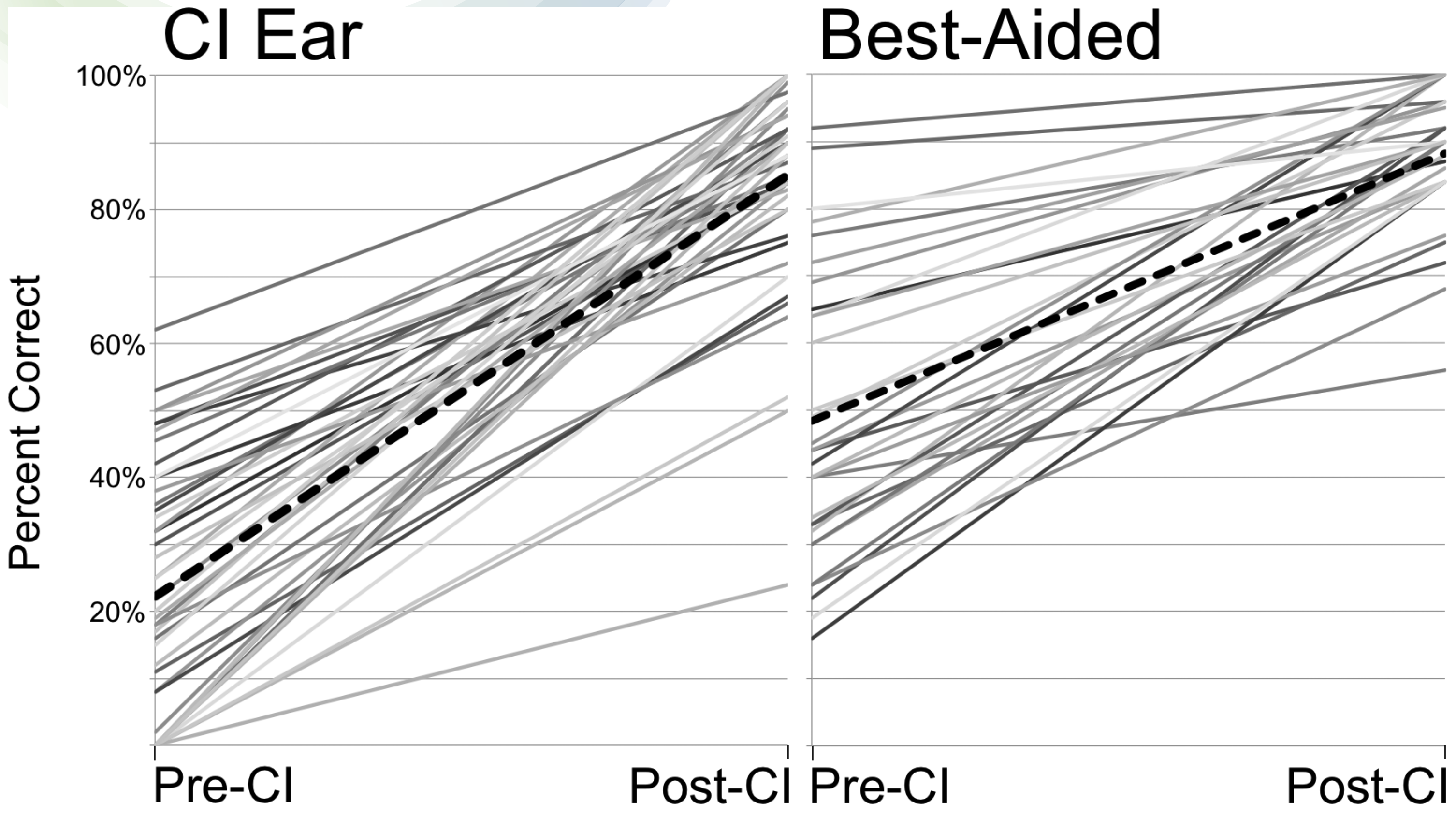
Expanding Degree of HL: Children



- Carlson et al. (2015):
  - Examined performance for 51 children implanted outside standard criteria
  - 63% improvement in speech recognition in CI ear and 40% improvement bimodal (mean 17 months post-CI)
- Teagle et al. (2019): 76% average word score post CI

Carlson et al. (2015), Fig. 6

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Single-Sided Deafness

Outline:

- Review of binaural hearing mechanisms
- How SSD impacts auditory development
- Considerations for candidacy and fitting
- Outcomes

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2019

Med-El Synchrony

FDA approval of CI for single-sided deafness

Binaural Hearing Mechanisms

- Spatial hearing abilities developed by 4 – 5 years in NH
- Primary sequela of SSD: lack of binaural hearing development
  - Sound localization
  - Head-shadow (ITD, ILD)
  - Binaural squelch
  - Binaural summation

squelch summation

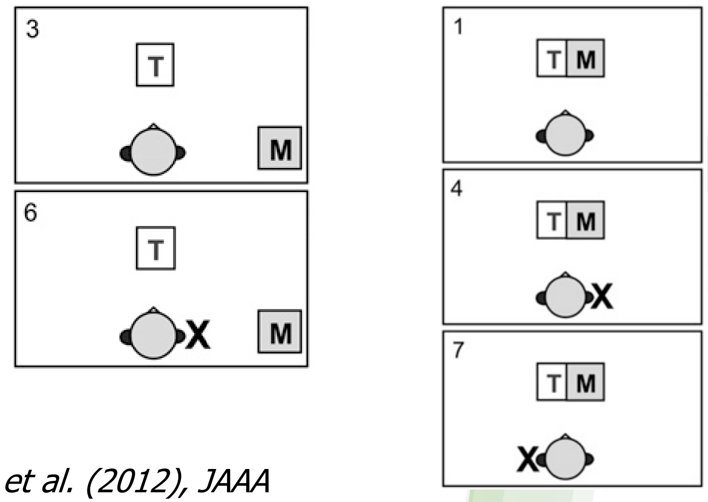

Fig. 1, Litovsky et al. (2012), JAAA

Binaural Hearing Mechanisms

- Functional sequelae:
  - Sound source segregation/listening in noise
  - Incidental learning
  - Cognitive load
  - Listening fatigue
  - Educational progress

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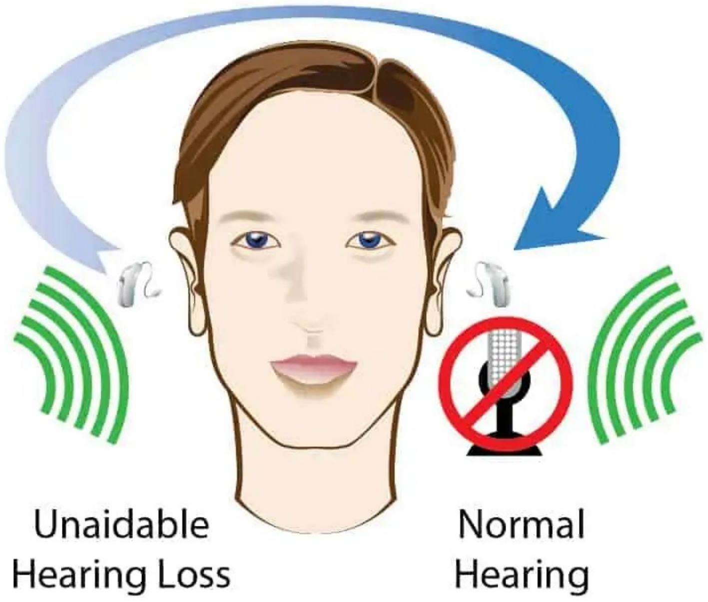
Effects of SSD on Auditory Development

- Cortical reorganization with early SSD
- Lack of development of binaural processing
- Cortical EEG studies show preference for NH ear early on (Lee 2020)
- Goal for implantation: development of binaural hearing mechanisms

- Treatment options: CROS via air or bone (BC HA or OIAI), CI

CROS



BCHA

CI

BAHA/ OIAI

Considerations for SSD CI Candidacy

- Treatment options: CROS via air or bone (BC HA or OIAI), CI

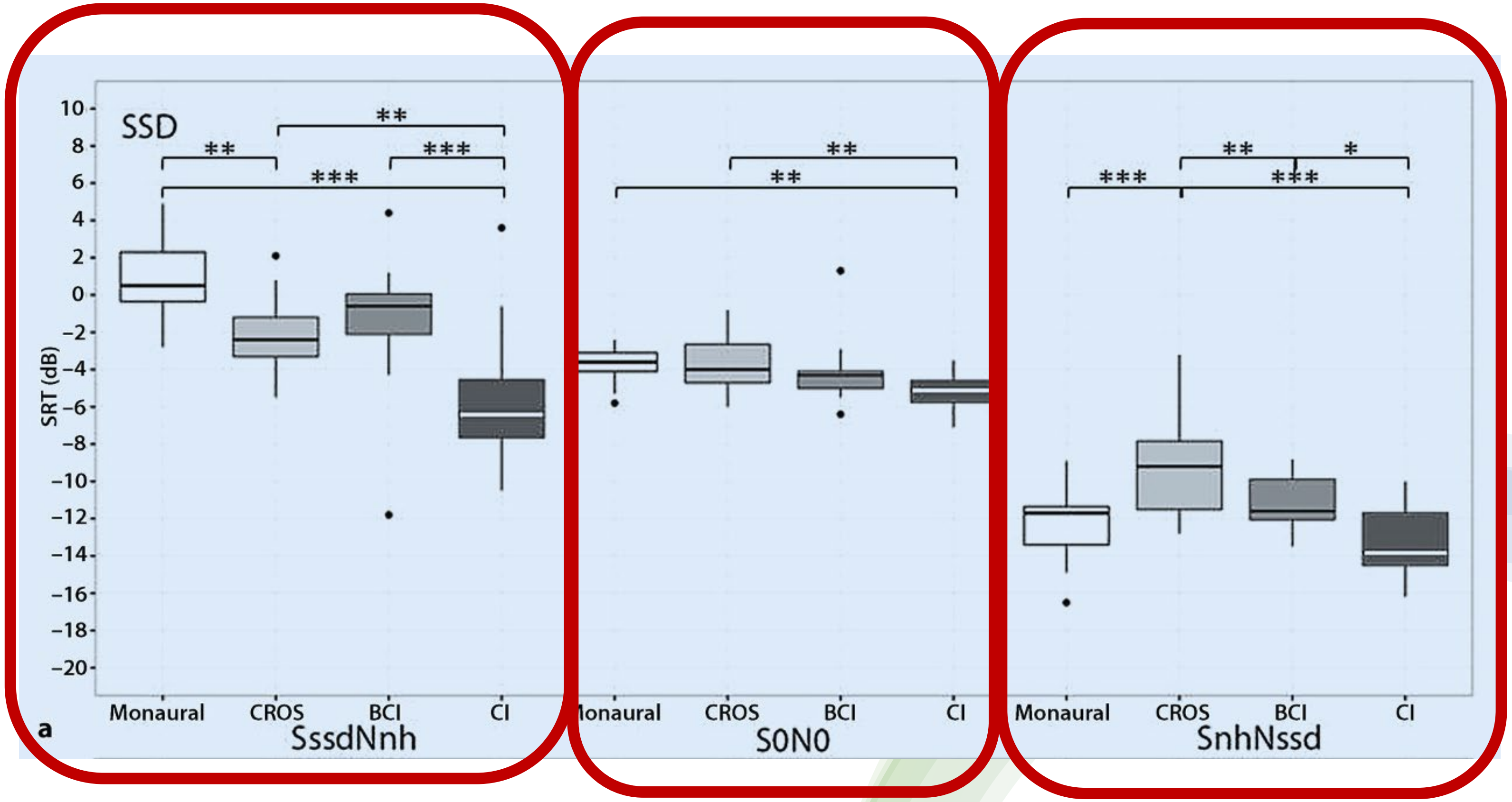


Fig. 3a, Arndt et al. (2017) HNO

N=45 adults with SSD

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Considerations for SSD CI Candidacy

- Localization abilities with different tx options:

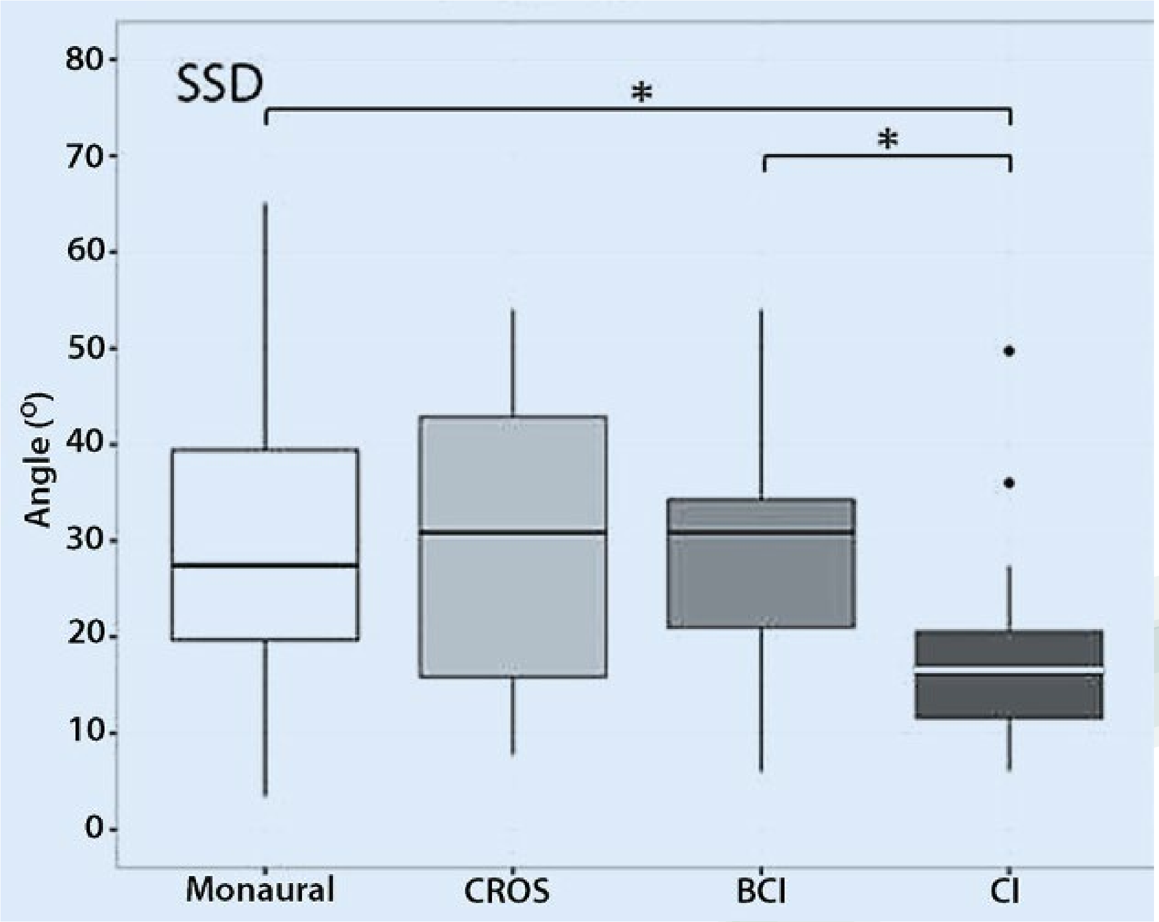

Fig. 3c, Arndt et al. (2017) HNO

N=45 adults with SSD

N
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Considerations for SSD CI Candidacy

- Etiology
  - CND (~25% cases, Dewyer et al. 2021; 58%, Arndt et al. 2015)
  - CCMV (67% cases; Lee et al. 2021)
  - ~2/3 of SSD cases are potential CI candidates
- Age (development of binaural hearing)
- Test conditions must exploit binaural mechanisms
- Med-El Synchrony 2 approved for age 5+ years, 4FPTA (500-4k Hz) ≥ 90 dB HL, ≤5% open-set word rec

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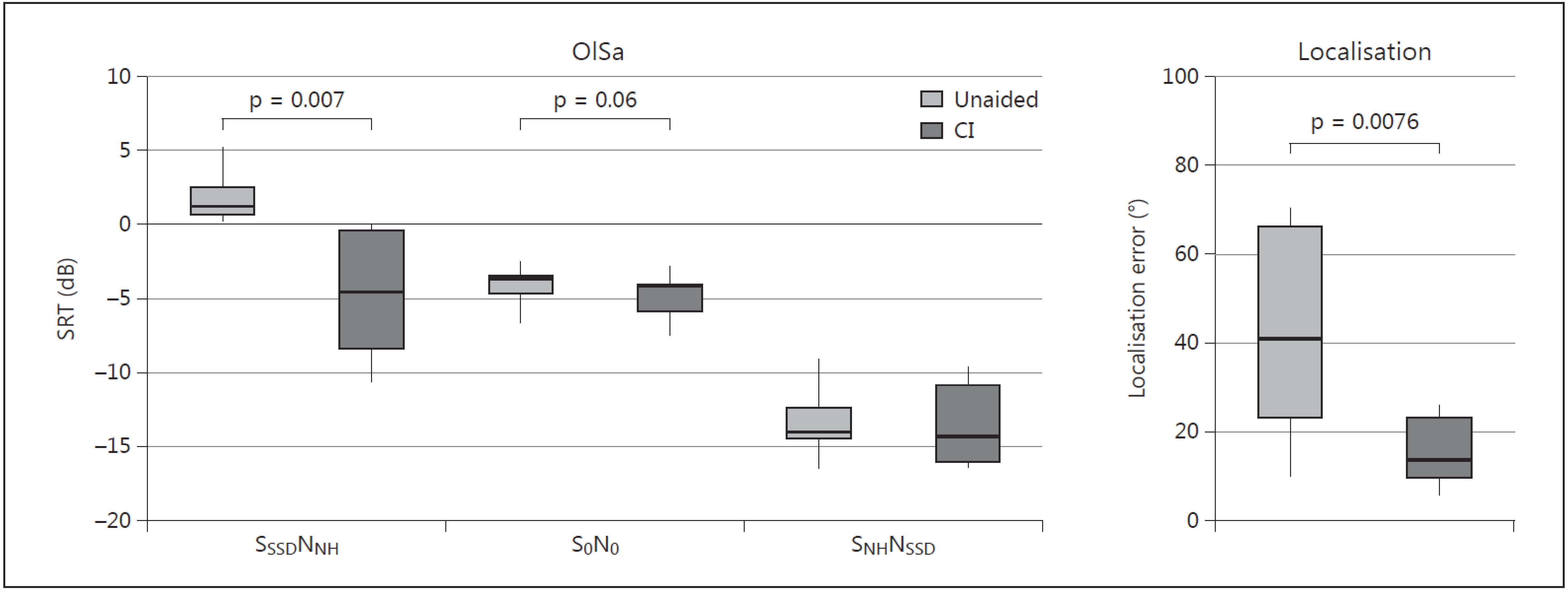
SSD CI Outcomes: Spatial Hearing



Arndt et al. (2015), Fig. 3, N=9 children

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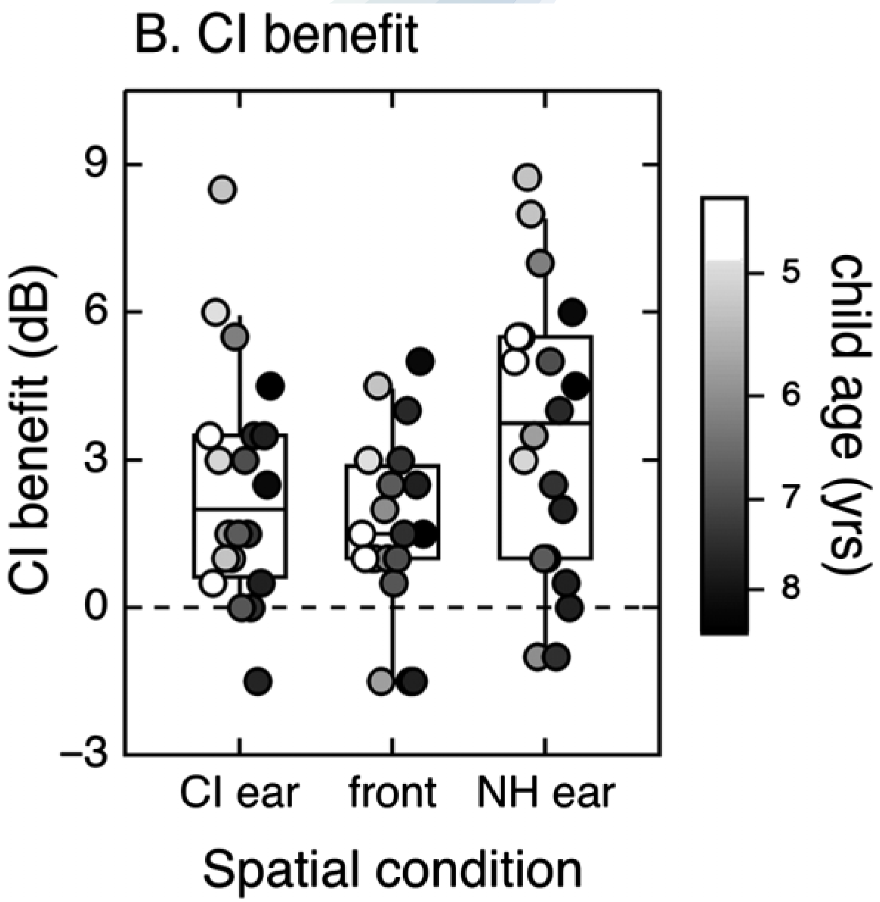
SSD CI Outcomes: Spatial Hearing



- 1.6 dB CI benefit with co-located target/masker (front)
- 2.5 dB CI benefit with masker on CI side
- 3.5 dB CI benefit with masker on NH side

→ Benefit of CI in diffuse noise, no interference of CI, exhibits binaural effects

Park et al. (2021), Fig. 1, N=20 children

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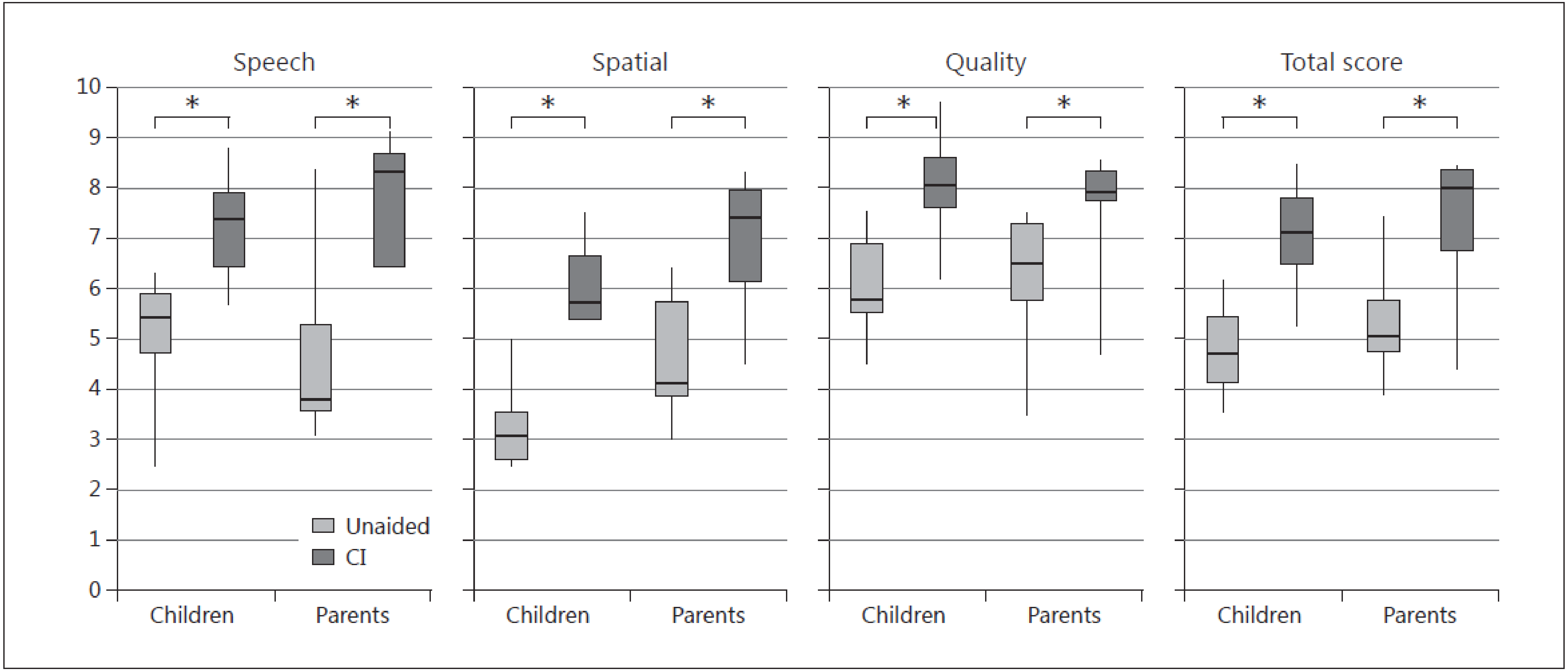
SSD CI Outcomes: SSQ



Arndt et al. (2015), Fig. 5, N=9 children

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Summary

- CI candidacy reduced to 9 months
  - Evidence supports better outcomes in vocabulary, expressive and receptive language, and speech recognition for earlier implantation
  - Future directions – address gap between maximum aided % for candidacy vs. typical CI performance
- CI for SSD is the only intervention that provides stimulation of deaf ear
  - Evidence of CI benefit and no interference in complex listening environments
  - Future directions – address gap in candidacy age vs. standard candidacy age and critical period for binaural development

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Part 3

What is on the Horizon?

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Where Are We Headed?

- Improved sound fidelity (signal processing, microphones, electrode array designs, pulse designs)
- Robotic-assisted surgery
- Continued work on totally implantable CIs (battery, microphone)
- Gene therapy

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Improved Sound Fidelity

<u>Cochlear</u>

- Nucleus 8 (waterproof)
  - Bluetooth LE Audio Technology --> Auracast compatible
  - SCAN 2 (SCAN was industry's first automatic scene classifier)
  - SmartSound iQ
- Nucleus 8 Hybrid
- Kanso 3 (waterproof)
  - Release mid 2025
- Nexa Internals
  - CI 1012 (contour advance), CI1022 (slim straight), CI1032 (slim modiolar)

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Improved Sound Fidelity

Med-El

- Sonnet 3 (waterproof)
  - Integrated direct streaming
  - Automatic Sound Management (ASM 3.0)
    - Adaptive intelligence
    - Fine hearing
    - Compatible with any hearing aid
    - Enhanced noise reduction
- Synchrony 2
  - 720-degree insertion depth
  - Anatomy-based fitting (OTOPLAN)
  - MRI conditional at 3.0 Tesla w/o magnet removal (rotating, self-aligning)
  - Flex soft (26.4 mm stimulation range)

Minimize Insertion Trauma

Fig 4, Adunka & Kiefer (2006) OHNS, 135, p. 380

Med-El FLEX; BM rupture basal turn

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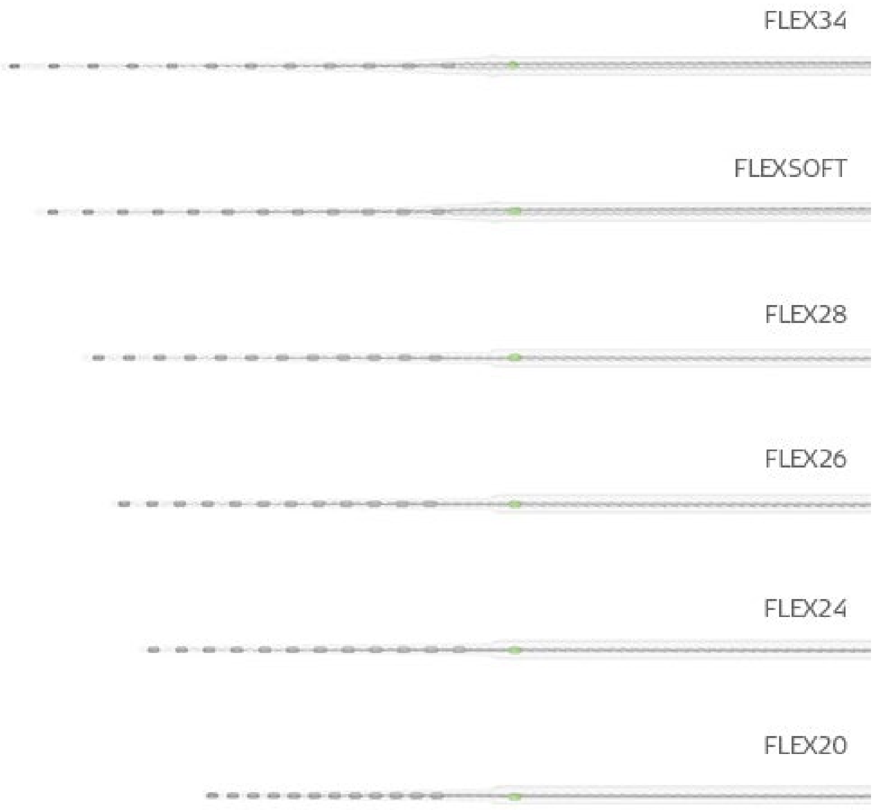
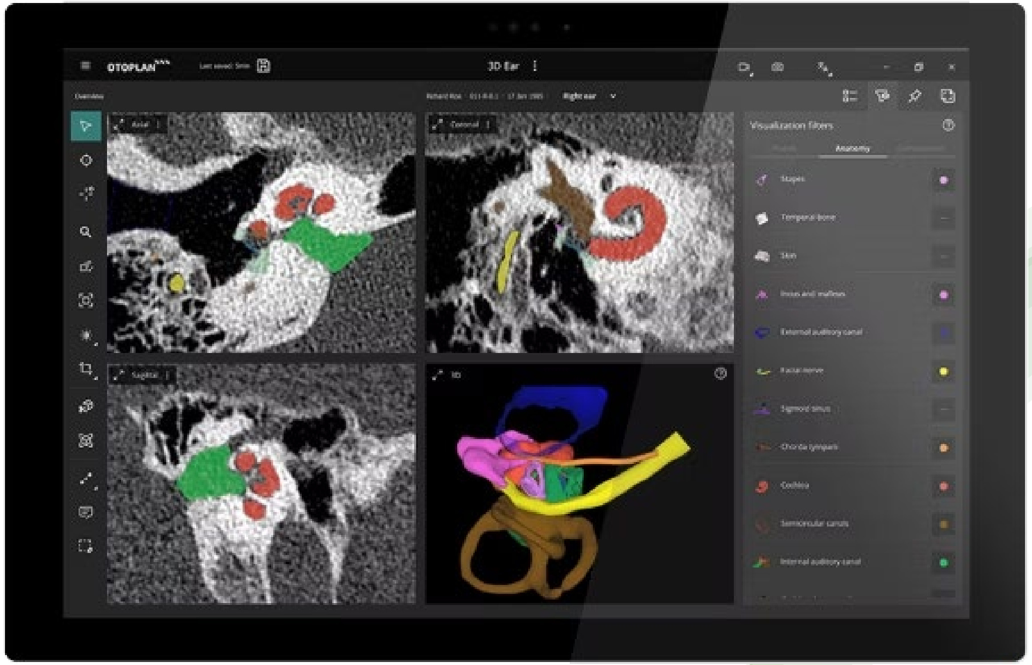
OTOPLAN

https://www.medel.pro/products/otoplan

- Cochlear duct length ranges from 25-36 mm
- Patient-specific 3D reconstruction
- Better selection of electrode array length
- Prediction of angular insertion depth and tonotopic frequency
- Simulated 3D electrode insertion
- Optimize insertion angle





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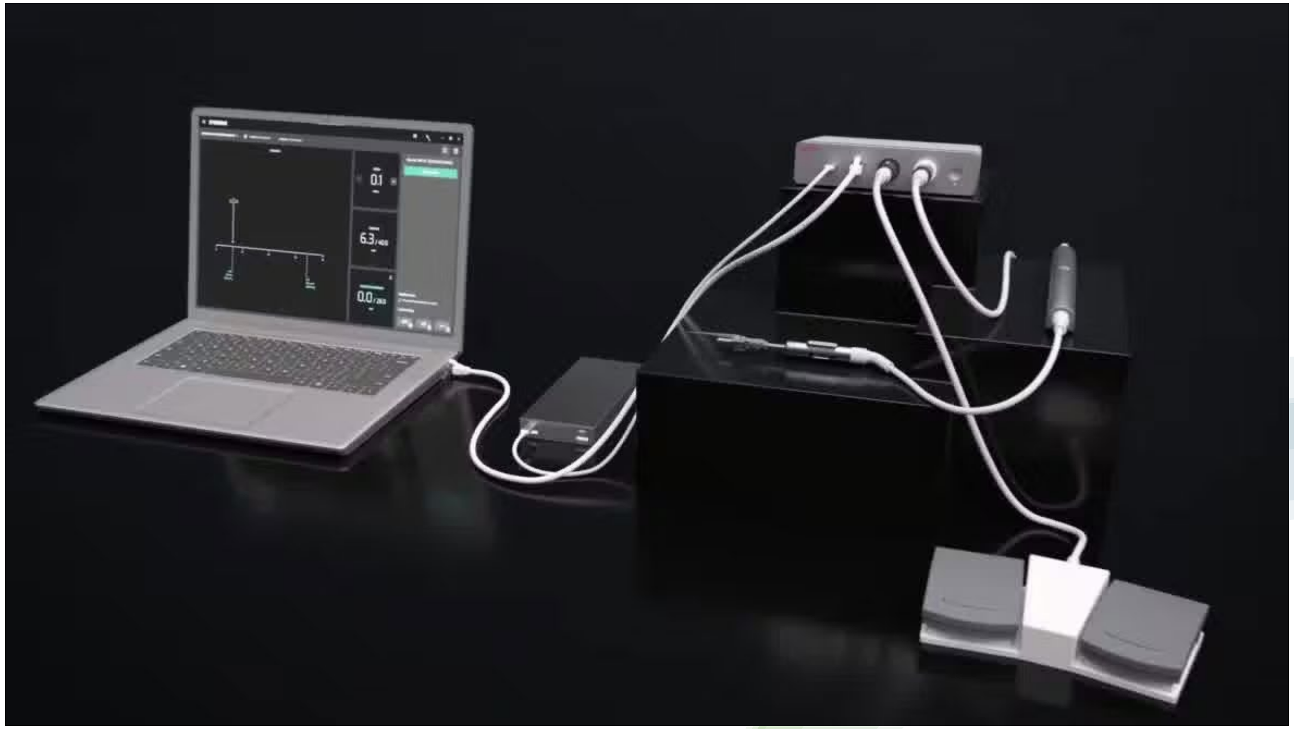
Robotic-Assisted CI Surgery

- OTODRIVE
  - Developed by Med-EL and CASCINATION
  - Connects with OTOPLAN
  - Slow, controlled electrode insertion



https://www.medel.pro/products/surgical-systems

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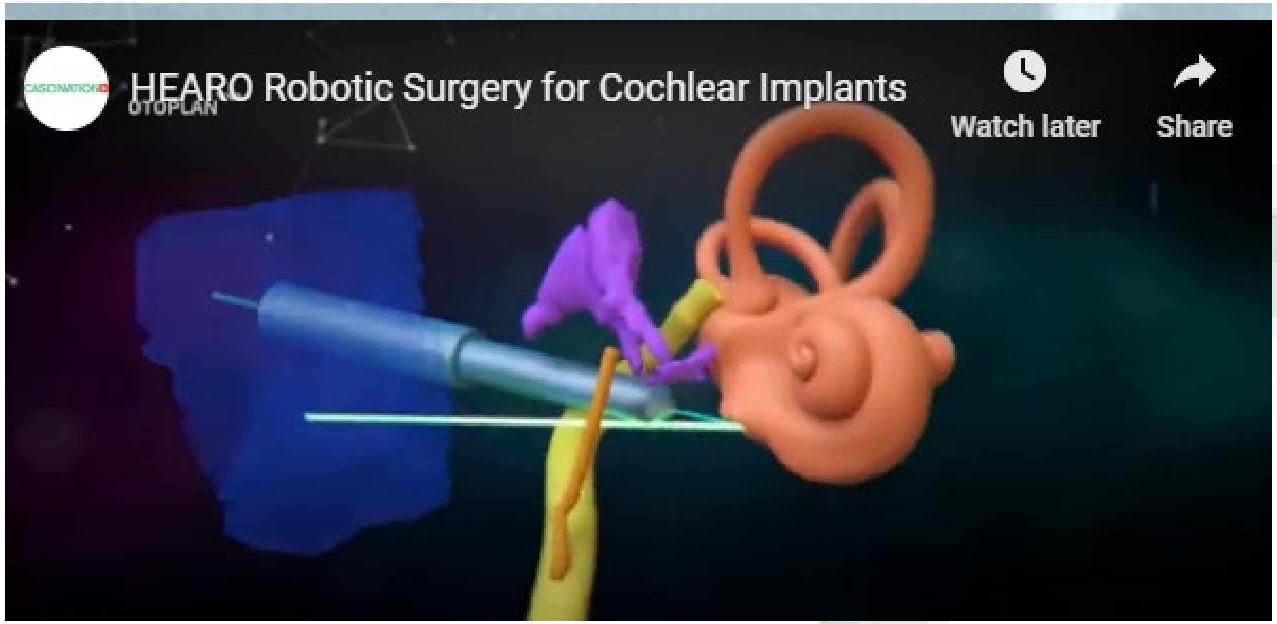
Robotic-Assisted CI Surgery

- HEARO
  - Minimally invasive access to inner ear
  - Minimize damage to facial nerve and chorda tympani
  - Used in conjunction with OTOPLAN
  - Real-time neural monitoring, imaging, and torque drilling



https://www.cascination.com/en/hearo#:~:text=Cochlear%20implantation%20is%20one%20of,to%20the%20middle%20ear%20cavity

Robotic-Assisted CI Surgery

- IotaSoft Insertion System
  - FDA cleared in 2021
  - Thumb-sized robot to precisely guide placement of cochlear implants
  - Temporarily attaches to mastoid during surgery
  - 10x slower insertion than surgeon, which has shown to better preserve the inner ear
  - Prospective, single arm, open label study (Gantz et al., 2023)

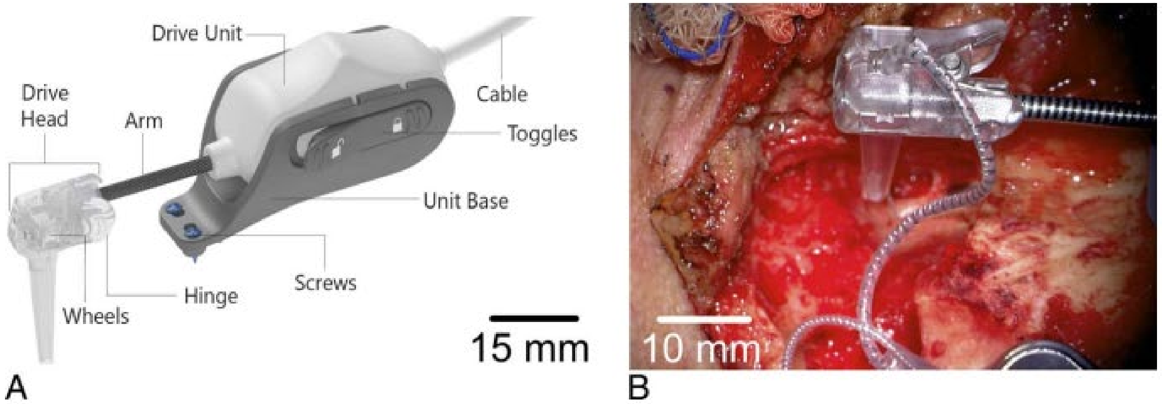


FIG. 1. *A*, Labeled diagram of the single-use robotic-assisted cochlear implant electrode array insertion device. *B*, Intraoperative photomicrograph of the electrode array insertion device loaded with a cochlear implant electrode array during a left-sided transmastoid facial recess approach, ready for insertion.

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Totally Implantable CIs

- Envoy Medical
  - FDA approval in November 2024 for staged clinical study of the Acclaim fully implantable CI (currently an investigational device)
  - Received the Breakthrough Device Designation from the U.S. Food and Drug Administration (FDA) in 2019

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Totally Implantable CIs

- Envoy Medical
  - Piezoelectric sensor on incus designed to leverage the natural anatomy of the ear to capture sound (no external microphone)
  - ME sensor → implanted processor → intracochlear electrode array

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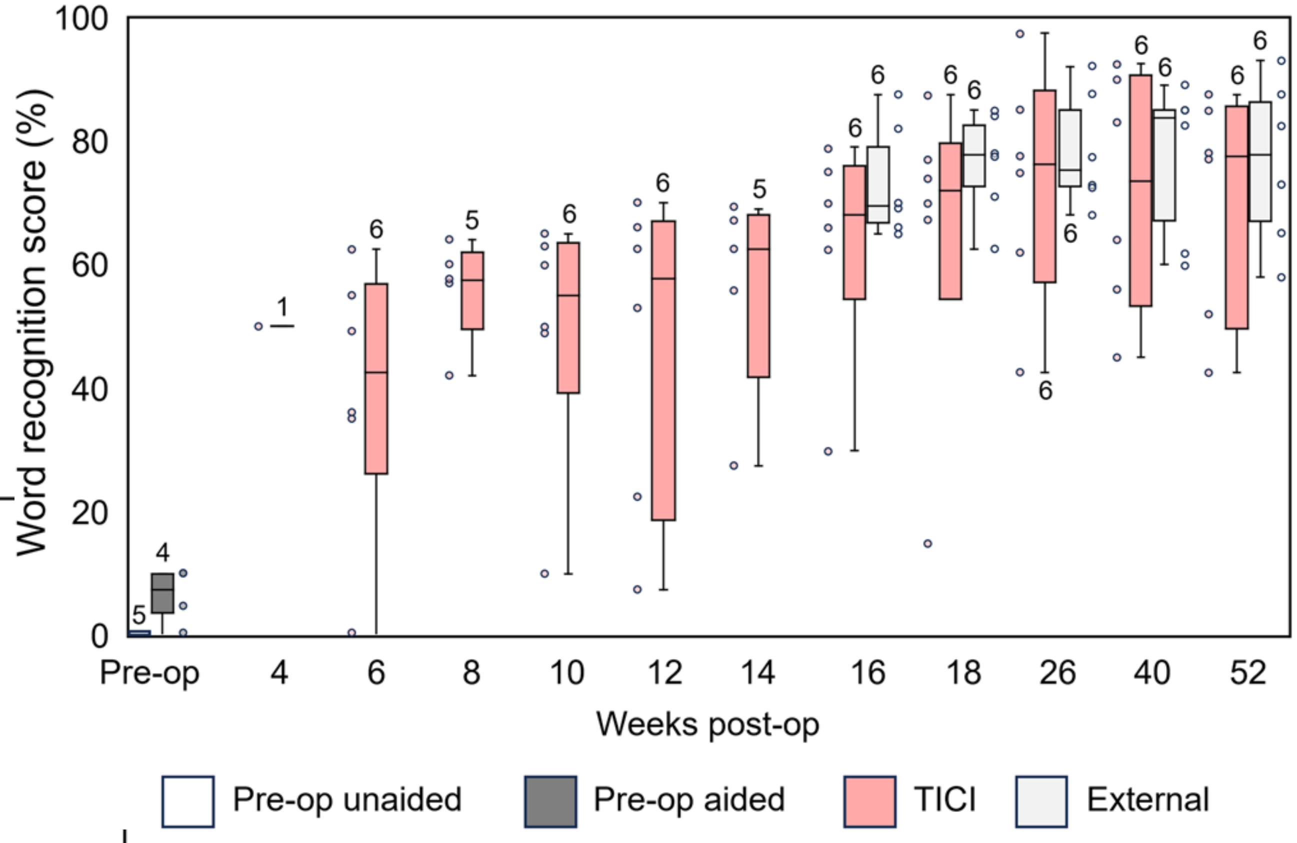
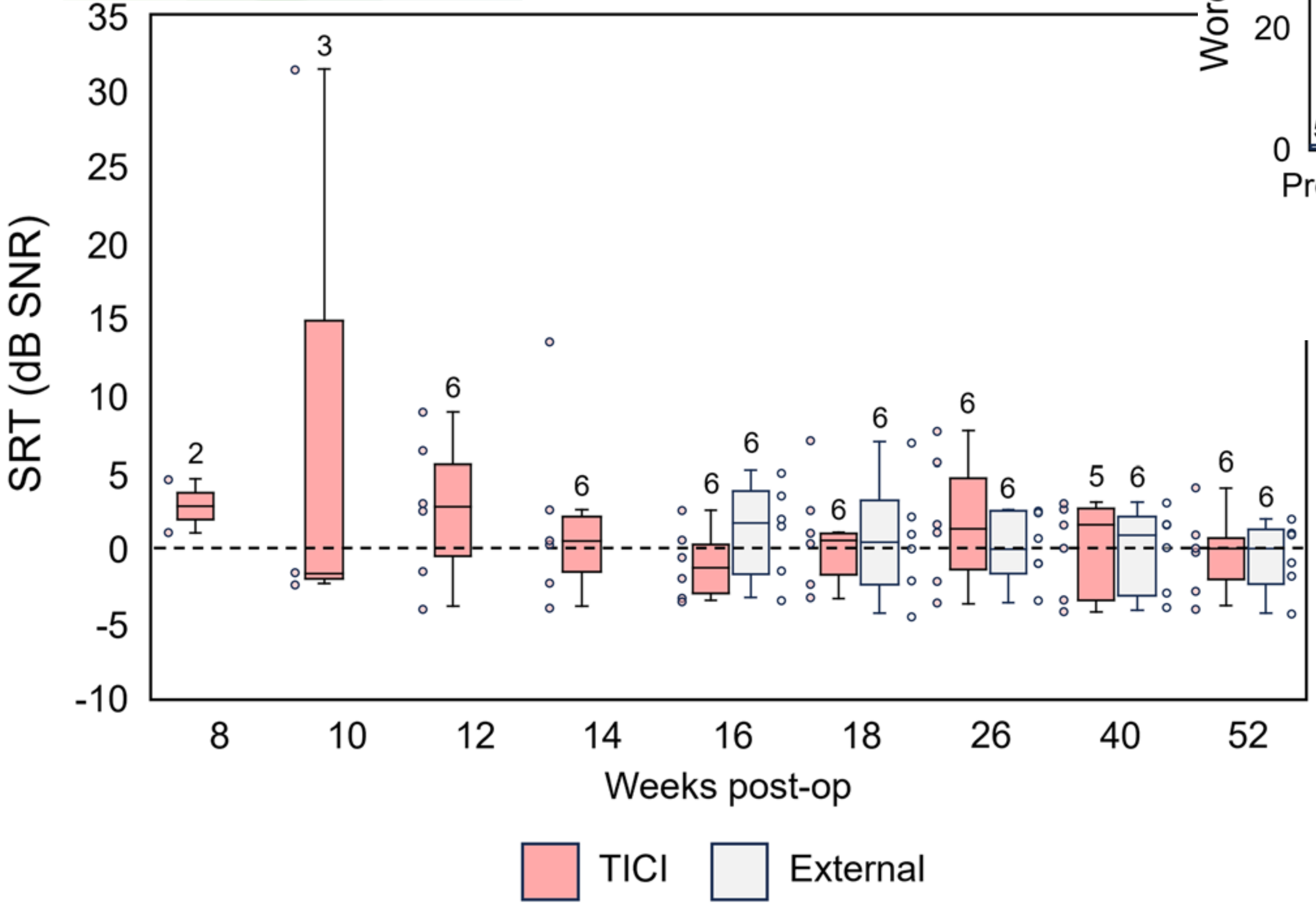
Totally Implantable CIs

- Med-El
    - Device is Mi2000
    - Similar to Cochlear's TICI (subdermal mic, option of external processor wear)
    - Comparable outcomes to traditional CI users
    - Improved comfort, satisfaction, and quality of life

Lefebvre et al., 2025

Lefebvre et al., 2025
Fig 1 (right)
Fig 2 (left)





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Gene Therapy

- ~80% of prelingual deafness is recessive genetic
- What is otoferlin?
  - Protein expressed in IHCs that mediates synaptic vesicle fusion
  - Recessive, DFNB9
  - Results in congenital severe-profound SNHL
- Otoferlin-based gene therapy with four clinical trials:
  - Akouos (n=2)
  - Fudan University (n=11)
  - Regeneron (n=1)
  - Southeast University/Otovia Therapeutics (n=3)
- Dual rAAV approach
- No dosage-related toxicity or severe adverse events
- Variability in outcomes from near-normal thresholds to moderate SNHL (from severe-profound)

Gene Therapy

- Single local injection of adeno-associated virus (AAV) vectors
  - Round window injection

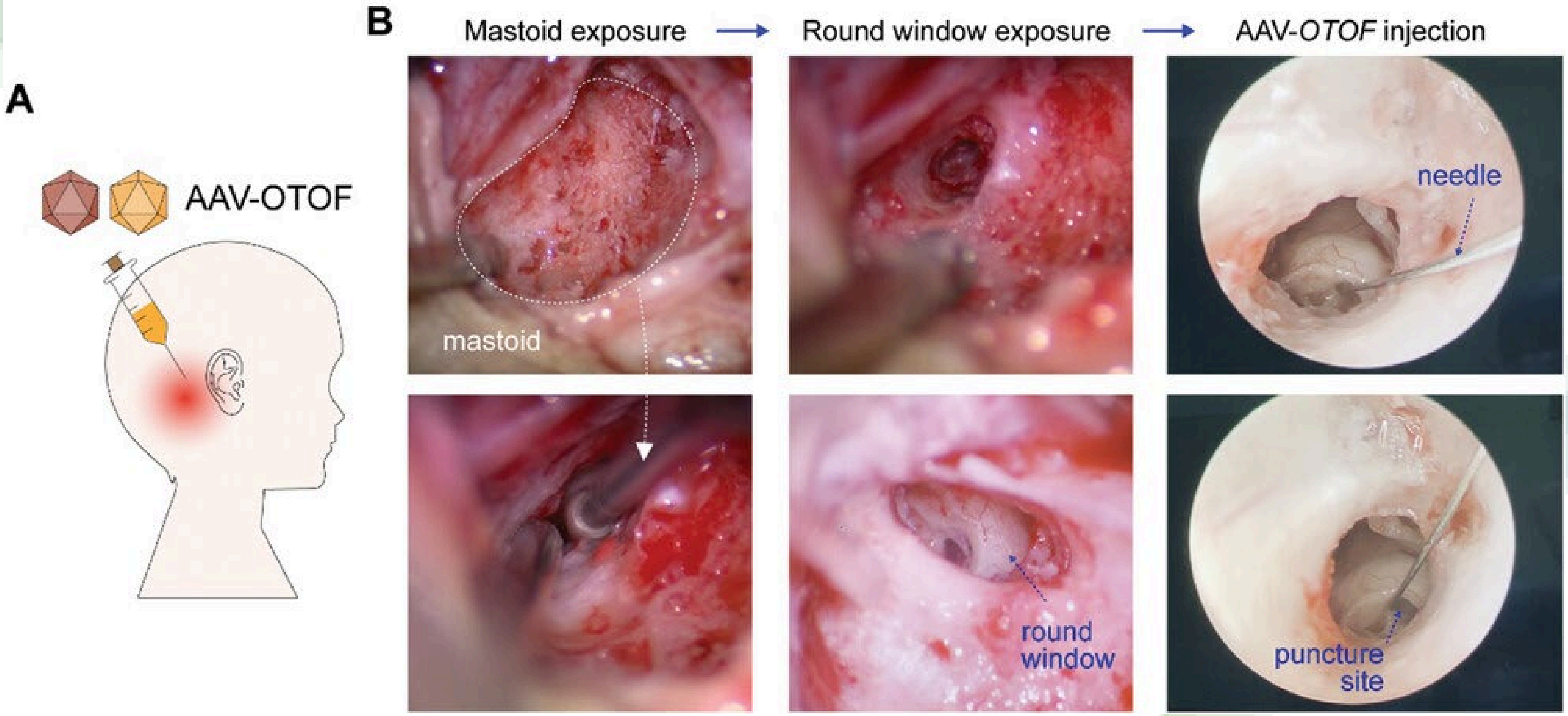

Fig 4, Qi et al. (2024) Advanced Science

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5 y/o patient

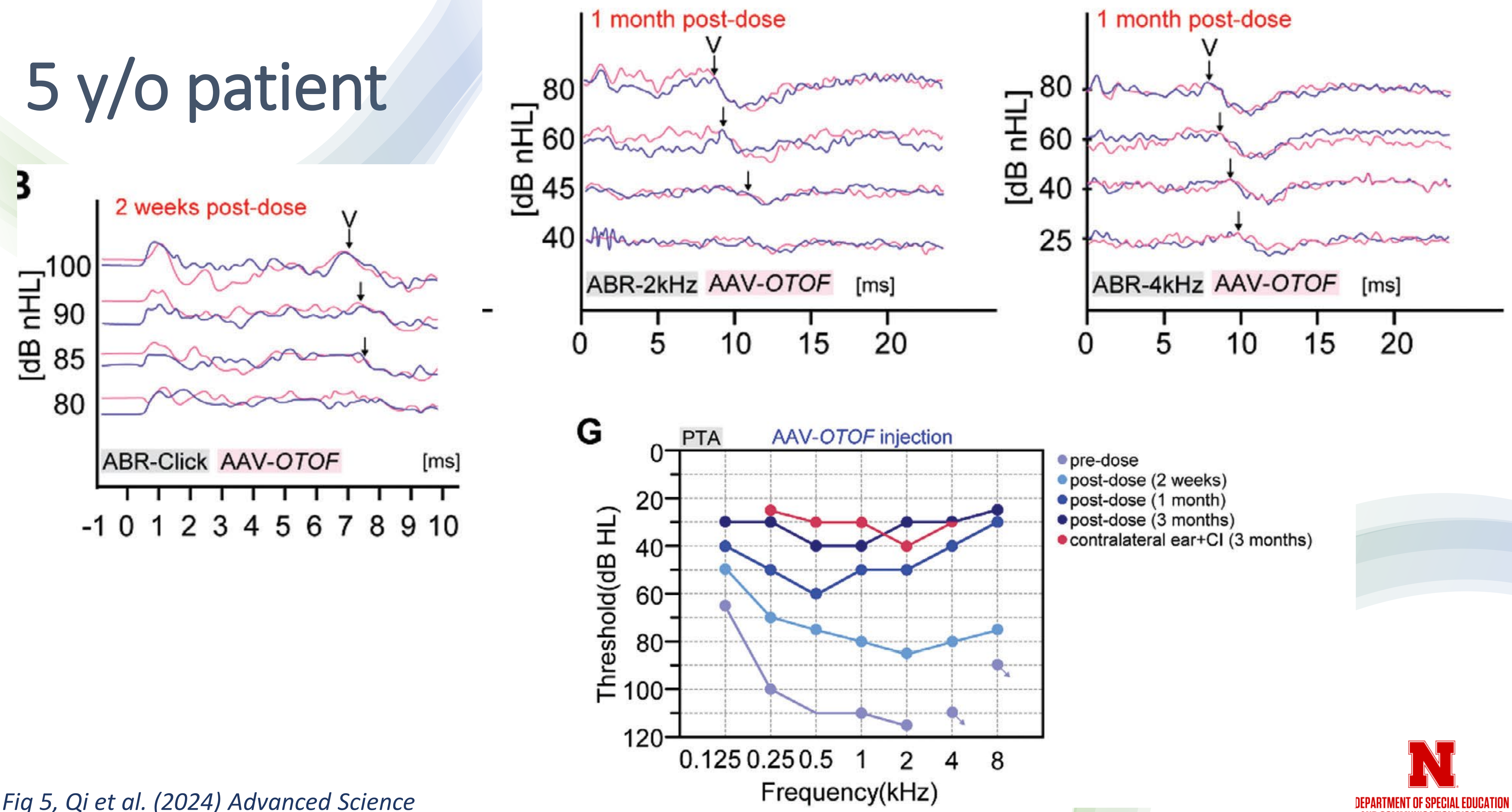
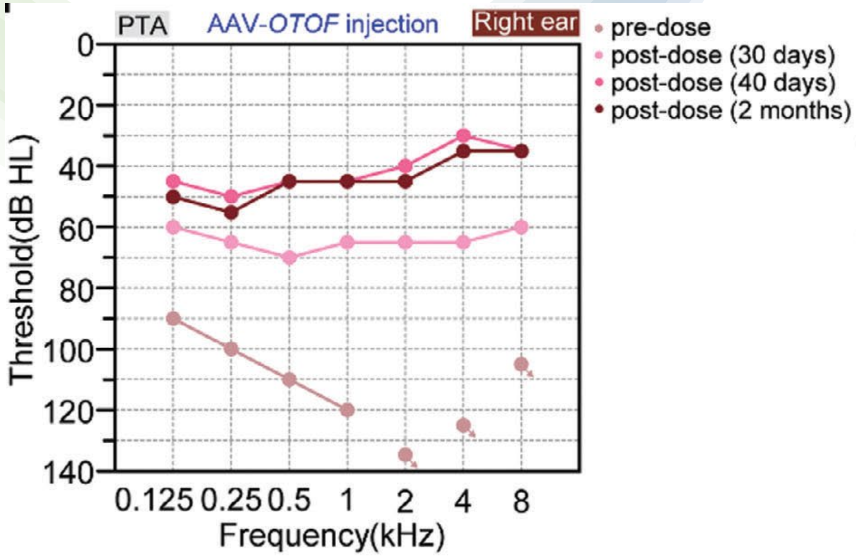
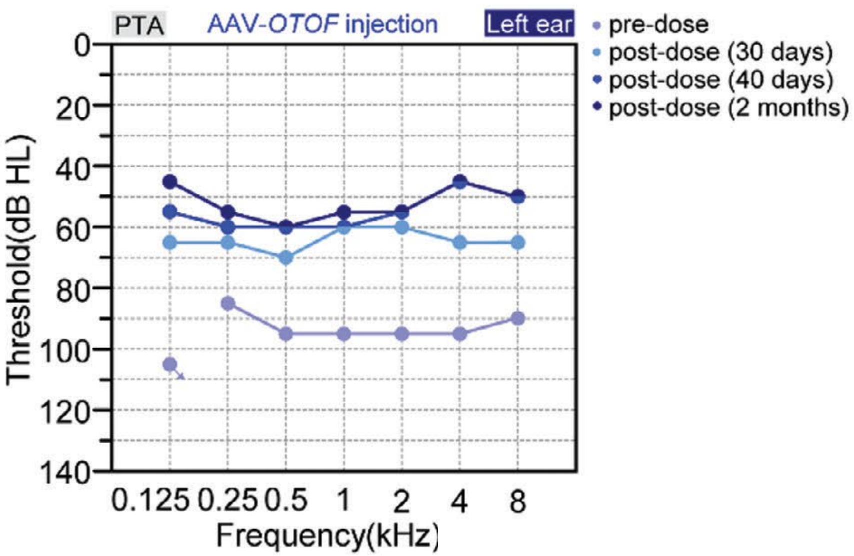
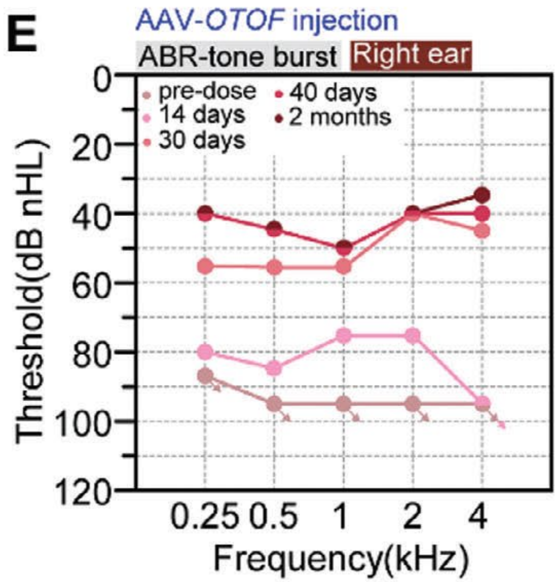


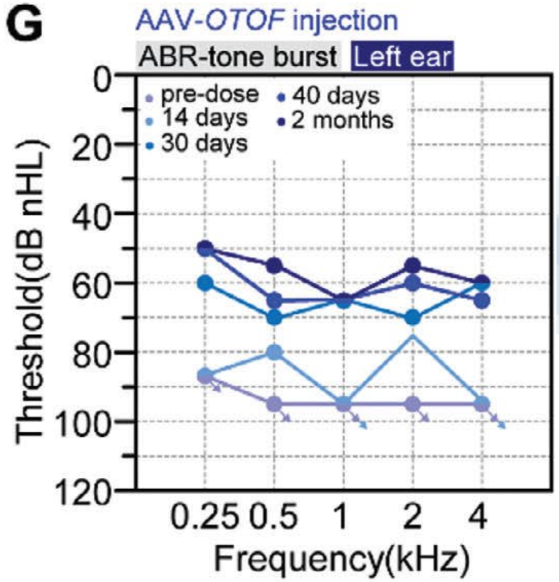
Fig 5, Qi et al. (2024) Advanced Science

8 y/o patient









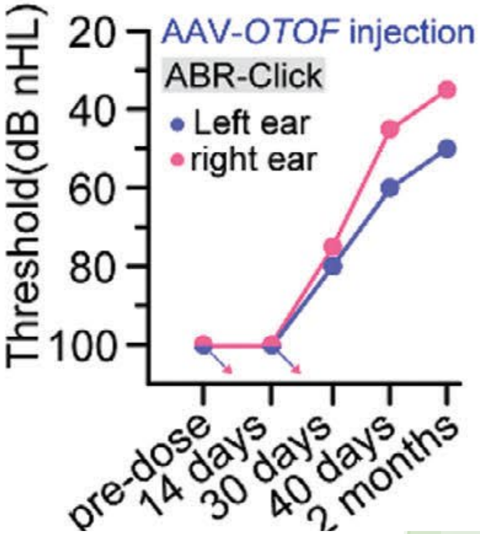


Fig 6, Qi et al. (2024) Advanced Science

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Gene Therapy: Remaining Questions

- Is a single dose sufficient for the lifetime?
- If only one cochlea is treated, can the other be treated later?
- Would redosing work if hearing declines?
- Will the treated ear be more (or less) susceptible to NIHL, aging, or ototoxicity?

Brigande 2024

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Questions?

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