Complex Cases

Jan Larky, AuD
Stanford Ear Institute

Melissa Hall, AuD
University of Florida Health
Disclosures

- M. Hall - Board of the American Cochlear Implant Alliance
- J. Larky - Board of the American Cochlear Implant Alliance and Envoy Medical Audiology Advisory Board
Patient Background

- **Uncomplicated pregnancy and birth**
  - Born full term, no complications, no infections, no prolonged hospital stay.
- No family hx of hearing loss.
- REFERed on Newborn hearing screening x 3 (outside report)
- Multiple ABRs confirmed bilateral severe-profound SN hearing loss
- Absent DPOAE’s
- Parent reported no auditory responses with or without amplification
- Meeting developmental milestones/typically developing
- Referred for CI evaluation
Comprehensive CI Evaluation

- **Audiology** (8 months)
- **Speech-language** (10 months)
- **Pediatric ophthalmology** (10 months) - unremarkable exam
- **Genetics**: Negative for Connexin 26 and 30 and Pendred. No disease causing mutations detected in the GJB2 gene
- **MRI**: very enlarged bilateral endolymphatic sacs
  - Cranial nerves VII and VIII present and intact in caliber.
  - Other anatomy - normal findings.
- **Bilateral CI recommended** (11 months)
Bilateral simultaneous implantation, 14 months of age
Advanced Bionics HiRes 90K Advantage with HiFocus Mid-Scala Electrode

Left ear
- Perilymph gusher upon entering round window - resolved with muscle packing
- Full insertion of the Advanced Bionics device
- No NRI responses, good impedances of all channels (AB rep)
- No abnormal activity of facial nerve monitor

Right ear
- Perilymph gusher upon entering round window - packed with muscle but continued to leak through incision
- Full insertion of the Advanced Bionics device.
- No NRI responses, good impedances of all channels (AB rep)
- No abnormal activity of facial nerve monitor

post surgery x-ray \(\rightarrow\) full insertion for both ears
Key observations over the next 15 months

- Consistent CI use
- Appropriate S-L (AVT), DHH BabyTalk therapy and services
- Good parental follow-through; never missed a visit
- Typically developing though walked late (>17 months); good eye contact and communicative intent
- Inconsistent responses to external sound; no responses to name noted by therapists & parent.
  - On occasion responses noted to stimulation during mapping
- No variability in sound productions
- Learning sign language suggests he can learn language
- He reportedly replaces the coils when they fall off (ear specific)
- Makes sounds when wearing right processor (reported and observed)
Additional testing

- Low impedances 1-2 kilohm range
- Various Programs created including CIS, Optima-P, Optima-S with no response up to 345 cu for either ear.
- 4+ processors tried to r/o external device involvement
- Integrity Test WNL AU (4 1/2 months post-op)
- CT Scan to confirm electrode placement – electrodes in cochlea AU, EVA noted AU (new)
- NRI attempted up to 847 cu – NR AU
15 months post activation

- Offered left ear reimplantation

- PLAN – **revise LEFT EAR** with Cochlear Nucleus Profile with Contour Advance CI512 implant
Left Ear Revision – 20 mos post activation (CA ~34 months)

- Cochlear CI 512 inserted
- Impedance testing - normal
- Intra-op NRT – no responses obtained
- X-ray shows excellent placement of the new cochlear implant.
Left Ear Cochlear Activation

- No response noted with a pulse width of 37 or 50. Eye blinks with PW 100. Reduced Cs, and created map.
- Live Voice listening with both AB and CA – cried.
- AB program lowered until he could tolerate both processors
- He began to vocalize consistently and loudly which mother and grandmother stated was different than previous behavior.
- At no point did he attempt to remove his processors.
1 Month post left ear revision

- Mother reported:
  1. Full time left ear Cochlear device use
  2. Increase in the consistency of his vocalizations
  3. Increase in the variety of his vocalizations

- Pulse Width was reduced from PW 100 to PW 50 usec.

- C-levels were set using Conditioned Play Audiometry.
  - Consistent and reliable; different from right AB ear
  - He readily conditioned to the task.

- Left Ear Detection of the Ling Six Sounds from 5' away
4 Month Post Left Ear Reimplantation

- Now in local TC program + private AVT weekly
- Per AVT - Good responses with Cochlear device; no responses with AB device
- Soundfield Threshold Testing
  - Conditioned Play Audiometry with a second audiologist
  - Reliability: Good left; Fair-poor right (device issue, not child issue)

Mother requested right ear revision
Right Ear Reimplantation & Activation

2 years; 2 months following initial implantation (age 3 now)

- **Reimplantation**
  - Full insertion of CI512 electrode array
  - Impedances WNL on all electrodes, partial neural response on one channel.

- **Activation**
  - Conditioned play audiometry employed with ease.
  - Comfort levels set where he responded consistently and quickly to all presentations of all the Ling Sounds.
  - No adverse reactions.
Last Audiogram from earlier this year. Age 4

1 Yr post left ear revision
6 Mo post right ear revision
• Mother thrilled but feels guilt about time lost
• Much better hearing
• AB device analysis WNL
• Time lost
  • 14 months first CI
  • Not hearing until 37 mos
  • Delayed speech-language
Recent Programming

Left Ear

Processor: CP1000 sound processor
Implant: CI512 cochlear implant
Strategy: ACE
Mode: MP1+2
Rate: 500
Maxima: 8
Pulse Width: 50
Parent MAP: 30 50 PW, 61 CL DR
Last Modified By: Custom Sound
Recent Programming

Right Ear

Processor: CP1000 sound processor
Implant: CI512 cochlear implant
Strategy: ACE
Mode: MP1+2
Rate: 900
Maxima: 8
Pulse Width: 25
Parent MAP: 24E16 142-193
Last Modified By: Custom Sound
Final thoughts?

- Longer time line than desired to achieve consistent sound awareness
- When to recommend reimplantation
- Still unresolved answers
  - What was he responding to with the AB devices? Internal sound/noise? Intermittent signal?
  - No response from both ears?! Explant device analysis – WNL AU
- Purpose & value of integrity tests
- Clinical experience and clinical expectations for progress given patient factors, intervention, parental involvement, amount of device use
- Collaboration with CI companies
- Approvals from insurance companies
- Collaboration with surgeon
- Counseling parents