Cochlear Implantation in Inner Ear Anomalies

Mehmet Ziya Özüer MD. Professor
Department of Otolaryngology Head and Surgery
Ahmet Hançer, MD. Ass. Professor
Çif. University Medical Faculty, Izmir, Turkey
Cochlear Implantation in Inner Ear Anomalies

Mehmet Ziya Özüer MD.
Professor
Department of Otolaryngology Head and Surgery
Ahmet Hançer, MD. Ass. Professor
Şifa University Medical Faculty, Izmir, Turkey
CI in inner ear anomalies
- Large vestibular aqueduct syndrome (LVAS)
- Incomplete partition type II (IP-II)
- Cochlear nerve deficiency
- Cochlear hypoplasia
Large vestibular aqueduct syndrome
Large vestibular aqueduct syndrome

- He is an 8 year old boy with LVAS, who had **progressive hearing loss**.
- He had CI surgery with an standard array through RW. Perilenf oozing was observed during surgery,
- He has been using his CI for 9 months.
Large vestibular aqueduct syndrome

- He has ESRT so his MCL’s can be set objectively.
- He hears well with his CI - Soundfield THR are as follows 250 – 6KHZ 35,35,25,30,35,30 dBHL
- Since receiving his CI he has started to use more words and join them together. He can understand simple commands without lip reading e.g. close the door
- He has 55% monosyllabic and 88% bisyllabic closed set word perception
Incomplete partition type II (IP-II)
Incomplete partition type II (IP-II)
Is a 19 year old girl with IP11 inner ear deformity. She has worn 2 powerful BTE’s since early childhood and has been in an auditory aural training program.

She was implanted with a special compressed electrode with a cork-like stopper. (FORM 24, from Medel company)

The electrode’s total length is 24 mm, which is ideal for IP-II anomalies.
FORM 24 electrode

It was designed and developed by Professor Levent Sennaroglu from Hacettepe, Turkey.

Electrode’s silicon ring is in the shape of a cork, in order to firmly close the cochleostomy, thereby to avoid perilennf oozing or gusher in inner ear anomalies.

Total length 24 mm
She has had her CI for 1 month. Her MCL’s are set objectively at ESRT level. She reports that she hears better with her CI and uses it all waking hours. Her sound field implant THR’s are 60-50-45-30-45-25 dBHL 250 -6KHZ and she scores 45% on closed set monosyllables.

Taking into consideration her very late age at implant these first results look promising.
Incomplete partition type II (IP-II)

Cystic appearance in the apex of the cochlea

Components of IP-II
- Enlarged vestibular aqueduct
- Dilated vestibule
- Defective apical part of the modiolus and the corresponding interscalar septa.

Laryngoscope, 112:2230–2241, 2002
Incomplete partition type II (IP-II)
Incomplete partition type II (IP-II)
This child is 2y 3 months old. She has IP II inner ear deformity.

She, also received Sonata implant with Form 24 electrode.

She has been using her CI for 5-6 months.
Her parents report that she wants to wear her AP, she hears soft sounds and can recognise many sounds. She has started to use words. She scores on LittlEARS Auditory Questionnaire like a hearing child of 19 - 20 months of age. She has **cortical P1 responses to /G/and/T/ at 55 dBSPL with latencies in the reference range.**

**This child is making excellent progress similar to a good CI performer with a normal cochlea**
Cochlear nerve deficiency

- This child is 3y 7m old. He was aided at 9 months but has not been able to use hearing aids consistently because of chronic middle ear discharge.
Cochlear nerve deficiency
Experienced audiologist detected a slight response with insert earphones, on the right side.

This information was very valuable in the side selection of CI.

The parents refused to receive an ABI and also counseled about insufficient progress with CI.
CI to right side 13/11/2014

ESRT seen on E 1,4,7,11 at 128,112,115, and 55 qu respectively during operation.
Cortical response

One week after the operation
At one week after CI to right ear his parents report that he wants to wear CI, that he vocalizes when it is first switched on and that he has been experimenting with loud sounds e.g. knocking. He can respond to 2 and 4KHZ at 70 dBSPL on VRA. He also has a **cortical P1 response to speech token /T/presented at 55 dBSPL** this demonstrates access to quiet high frequency sound. During fitting aeropalpebral reflexes were found on E 7-12 but not on other electrodes.

Yusuf has only had his CI for 1 week, these early responses are quite promising.
Conclusion

- Surgeons should have acquaintance to accurately recognize inner ear anomalies on HRCT and MRI images.

- Imaging studies may not precisely demonstrate or preclude the presence of the cochlear nerve (CN).

- Form 24 electrode could be an alternative in IP-II patients.

- While decision making in side selection in CI with CND; clinical audiologist observations and recommendations have the priority to imaging findings.