Early cochlear implantation in babies with congenital unilateral deafness. Results and revalidation/assessment methods.

A. Vermeiren, L. Theuwis, A. Zarowski, E. Offeciers

14th International Conference on Cochlear Implants and Other Implantable Technologies
Toronto, May 2016
I have no relevant financial relationships nor conflicts of interest pertaining to this presentation.
CI and unilateral deafness

• A number of publications show benefits of CI’s in postlingual adults and children with unilateral deafness
  ▪ Improved speech in noise results
  ▪ Better localisation skills
  ▪ Reduced listening effort
  ▪ Suppression of tinnitus

• What about CI in congenital SSD?
  ▪ No reports available on the results of CI in unilaterally deaf pre- / perilingual babies
Rationale for CI in congenital SSD


- Congenital SSD causes important diminishment of significant language input.

- As such, SSD often has the same impact as too little language input in bilaterally hearing children.

- Implanting the deaf ear can at least partially remediate the impact of SSD on cognitive development.
Case 1 (LB)

- Girl, born March 11, 2011
- Unilateral refer on neonatal hearing screening (April 2011)
- Diagnosis based on ABR/OAE (May - June 2011)
  - Right ear: deaf
  - Left ear: normal at initial testing, progressive hearing loss
- Cause: congenital CMV
- MRI: hyperintensity in the anterior part of the basal turn of the right cochlea – fear for fibrosis & ossification
- CI June 12, 2012 at the RIGHT side (age 15m)
Case 1 (LB) - Rehabilitation

- Intermittent **masking** of the normal ear
  
  - Explicitly challenge the brain to deal with the sound of the CI
  - Reach the maximum benefit of the CI in a faster way
  - Training protocol consolidated in adult patients
  - 3-5 hours daily during the 1st year post-implantation
  - White noise via BTE noise generator at 60dBSPL
Adequate masking of the better ear is needed to evaluate results with CI

- Mere plugging of the ear canal or covering of the better ear is not sufficient (experience in adult patients)

- **Active masking is necessary!**
  - Headphone or insert phone
  - Masking level = intensity of the stimulus
Case 1 (LB) – Results Tonal Audiometry and phoneme tests

- Tonal audiometry

- Phoneme detection and discrimination:
  - Stimulus level 70 dB HL in free field
  - Masking of the better ear
  - Results: 100% detection and discrimination with CI
Case 1 (LB) – Results Speech Audiometry in quiet

Due to the progressive hearing loss in the left ear, we recently advised the parents to use a hearing aid in the better ear.

- Even in quiet, improvement is shown!
Case (LB) – Results Speech in Noise Tests

**Diotic condition (Göttinger)**

- TR = CI right switched on
- X = CI right switched off

**Dichotic condition (NVA)**

- TB = CI right and HA left switched on
- TL = CI right switched off, hearing aid left switched on

Speech and noise 0°

Speech at CI side, noise HA side
Case 1 (LB) – Results Lateralisation / Localisation

- Perfect lateralisation (+90° versus -90°)
- Localisation (-90°, -45°, 0°, +45°, +90°)

HA switched on, CI switched off

HA and CI switched on

x attempt 1, + attempt 2, o attempt 3
Case 2 (EB)

- Girl, born December 16, 2012
- SSD due to CMV
- CI 09/09/2014 at the LEFT side (age 20 m)
- Rehabilitation:
  - Intermittent masking of the normal ear
  - Multidisciplinary therapy due to developmental speech language, mental and motor delay
- Good audiogram with CI, 100% phoneme detection and discrimination
- Due to developmental delay, not possible to test speech understanding and localisation until now
What did we learn from these cases?

- Cochlear implantation in a baby with unilateral deafness seems to offer a clear benefit in speech understanding.
- Improvement offered by the implant is especially important in difficult listening environments.
- The children wear the implant spontaneously every day and seem to integrate both inputs very well.
- There is evidence for at least limited binaural processing enabled by implantation of the deaf ear in these children.
- Specific revalidation and testing protocols might be required in such patients.

- Further cases are scheduled in the near future
Thank you for listening!