Cochlear Implantation of Children with malformations of the Inner Ear

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The aim of the study was to prove, that it is possible to implant children with congenital malformations of the inner ear with good results. Under certain conditions, these children can perform on the same level as their implanted peers with a physiological cochlear development.

Questions, which are important when assessing possible cochlear implantation in children with inner ear anomalies:

- Are cochlear structures patent?
- Where is the hearing nerve ganglion (ganglion spirale cells)?
- Is the hearing nerve present?

Our group
During a period 06/2014 till 12/2017, there were implanted 122 children, out of this number the inner ear anomaly was diagnosed by CT and MRI in 5 cases (4%). Two examples are described in details.

1. Bilateral common cavity. Preoperative MRI showing common cavities bilaterally with an identifiable hearing nerve:

The boy was implanted at the age of 13 months, the right side first. Due to good results, he was implanted in the left side after in 10 months period. The implant used was Nucleus CI24RE(ST)™, the array was inserted into the cavity by a “banana” cochleostomy.

Pictures showing a slit banana cochleostomy for an insertion of a straight electrode array. After insertion, the cochleostomy opening is packed with muscles. No gusher was observed on either side.

Postoperative native fluoroscopy showing correct position of both the arrays in cavities.

Currently, the boy is 3 years after the surgeries has a good speech, uses about 400 words and simple sentences. The profit is comparable with his implanted peers with physiological cochleas.

2. Incomplete partition I, CHARGE syndrom.
Preoperative MRI:

Patient was implanted at the age of 5 years 2 months, right ear was used. The electrode was Nucleus CIS22™ (“slim straight”). The array was inserted by a round window, there was a small gusher (“oozer”) peroperatively and postoperatively for a short time.

Postoperative native fluoroscopy showing a position of the electrode in the cavity.

During the beginning of a rehabilitation, the patient had a vestibular reaction with a head tilting. Maximum stimulating level is limited by a parasitic stimulation of the facial nerve. Currently, she uses about 10 words, needs to lip read and take advantage of gestures during communication.

Conclusion: children with inner ear malformations should not be excluded from an implantation programme. Some of them can be very good, comparable with children with physiological cochleas. In other cases, the progress is rather slower, especially in children with additional handicaps and a later indication for surgery. In cases of a good implantation outcome, a sequential bilateral implantation is a method of choice like in standard cases.