

## COCHLEAR IMPLANTATION IN A CHILD WITH COMPLEX BILATERAL INNER EAR AND COCHLEO-VESTIBULAR NERVE MALFORMATIONS

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### ABSTRACT

**Introduction:** The cochlear implantation in patients with inner ear malformation has always been a challenge even to the most experienced clinicians. We present the case of a child of 8 years old with profound bilateral sensorineural hearing loss and the absence of language development with indication of cochlear implantation as single solution for hearing and speech rehabilitation. **Methods:** The audiological assessment indicates the cochlear implantation. Preoperative CT scan revealed a bilateral inner ear malformation (cochlear common cavity deformity on the right side, cochlear aplasia on the left ear and bilateral vestibular malformation). Brain magnetic resonance showed the presence of auditory nerve only to the right side. The girl was implanted on the right ear in the common cavity with an Advanced Bionics device, HiRes90K with HiFocus1j electrode. **Results:** The insertion of the portelectrode inside the malformed inner ear was confirmed by X-ray. Audiological evaluation had shown an average of hearing thresholds at 36,25 dB HL in free field pure tone audiometry at 22 months post-implantation. The speech perception was evaluated according to CAP (categories of auditory performance) index at 5 and to the speech intelligibility rating scale (SIR) to 3. **Conclusions:** The children with inner ear malformations can safely benefit of cochlear implantation. The results for tonal hearing were close to that of cochlear implanted children with normal ear anatomy, but were limited for speech understanding and speech production.

**Keywords:** cochlear implant, inner ear malformations, cochleo-vestibular nerve malformation

### INTRODUCTION

The medical evaluation and management of children with profound hearing loss (HL) with associated inner ear developmental malformations presents a significant challenge even for the most experienced ear surgeons and audiologists. In recent years, the number of cochlear implantations in children has increased worldwide, as well as cochlear

implantation in congenital malformations. First results related to cochlear implantation in children with inner ear malformations have been appearing since 1988 and encouraging results have also been reported [1]. The results are better with the technological developing of cochlear devices, nowadays existing different electrode options for malformed cochlea including the customized

electrodes.

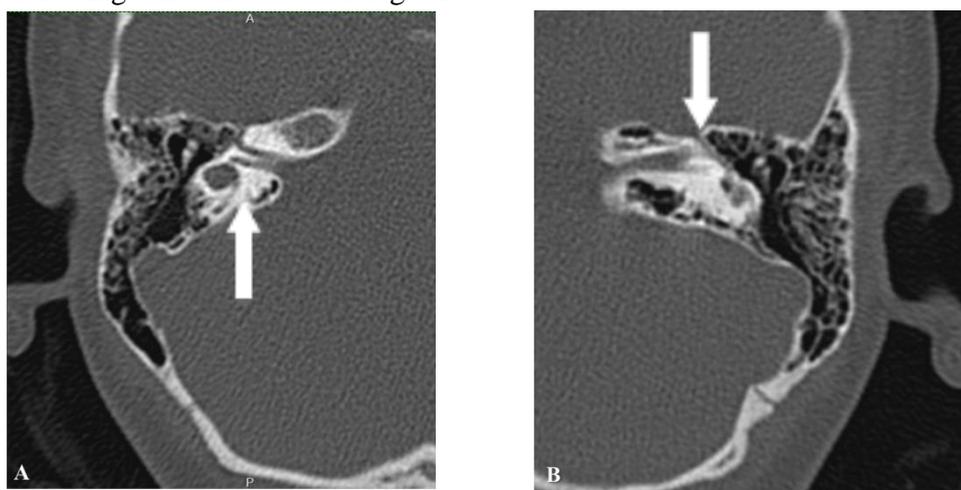
We present a case of a 8,6 years old girl, without family antecedents of hypoacusia, full-term born without any neonatal intercurrence, not submitted to newborn hearing screening, who addressed to us for bilateral prelingually hearing loss. The patient was identified as having bilateral profound hearing loss and no language development at the age of 3 year old in another hospital. Immediately after, she was fitted bilaterally with conventional hearing aids. Because the lack of the results, she changed three times the conventional hearing aids in the next 5 years, but the only one result was very limited responses at high levels of intensity for low frequencies and no benefit in speech perception and speech production.

Considering the new audiological

reevaluation at the age of 8 years old and the child's previous evolution (absence of language acquisition) we decided that the only one possibility for the auditory rehabilitation is the cochlear implantation.

## MATERIAL AND METHODS

According to the age of the child, the audiological assessment was made either by subjective tests as pure tone audiometry (PTA) and free field audiometry without/with hearing aids as well as objective tests: tympanometry and acoustic reflex, otoacoustic emissions, auditory brainstem responses (ABR) and auditory steady state responses (ASSR). All audiological tests were performed on calibrated Interacoustics equipment in soundproof rooms.



**Figure 1. Preoperative CT scan of the temporal bone (axial plane)**  
**A. Right ear: cochlear common cavity and vestibule malformation;**  
**B. Left ear: cochlear aplasia and vestibule malformation.**

The PTA revealed a profound hearing loss with residual hearing only at low frequencies on the right side and the free field audiometry with hearing aids showed a very poor gain. No auditory brainstem response could be evoked in any ear at a stimulation level of 100dB normal hearing level (nHL), otoacoustic emissions were absent bilaterally. No auditory steady-state responses were identified. Tympanometry was normal and

acoustic reflex was bilaterally absent.

We therefore decided to carry out cochlear implantation. Preoperative CT scan revealed a bilateral congenital inner ear malformation (cochlear common cavity deformity on the right side, cochlear aplasia on the left ear and bilateral vestibular malformation) (fig. 1). Brain magnetic resonance showed the presence of some very thin nerve fibers corresponding to the single cavity of the inner

ear on the right side, which seems to be just a branch of the vestibular right nerve (fig. 2).

The child was submitted to right ear cochlear implantation procedure at 8,6 years old. The electrode-array was fully inserted through the cochleostomy, which was accurately packed with a piece of muscle and temporalis fascia shaped around it. Continuous facial nerve monitoring was used during the surgery and the course of the facial nerve seemed to be normal.

High impedances were observed

intraoperative, as indication of open circuits for 14 of 16 electrodes. At the activation time we found normal values of impedances for all electrodes (fig. 3). As a special feature that can be noticed is a huge difference between the intraoperative and the successive values of impedances from 67,5 kΩ to values around 4 - 5 kΩ. In terms of cochlear malformations, intraoperative impedances value profile is different from the one we encounter in patients implanted with normal cochlea.

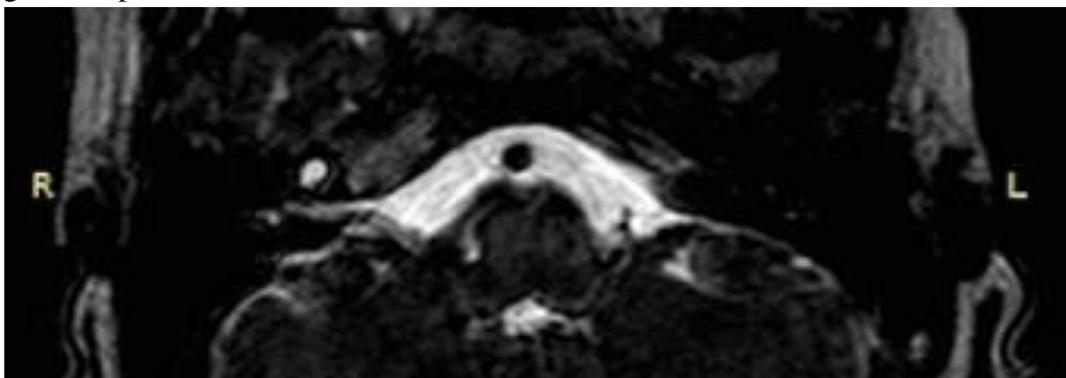


Figure 2. Preoperative MRI - very thin nerve fibers corresponding to the single cavity of the inner ear on the right side (branch of the vestibular right nerve)

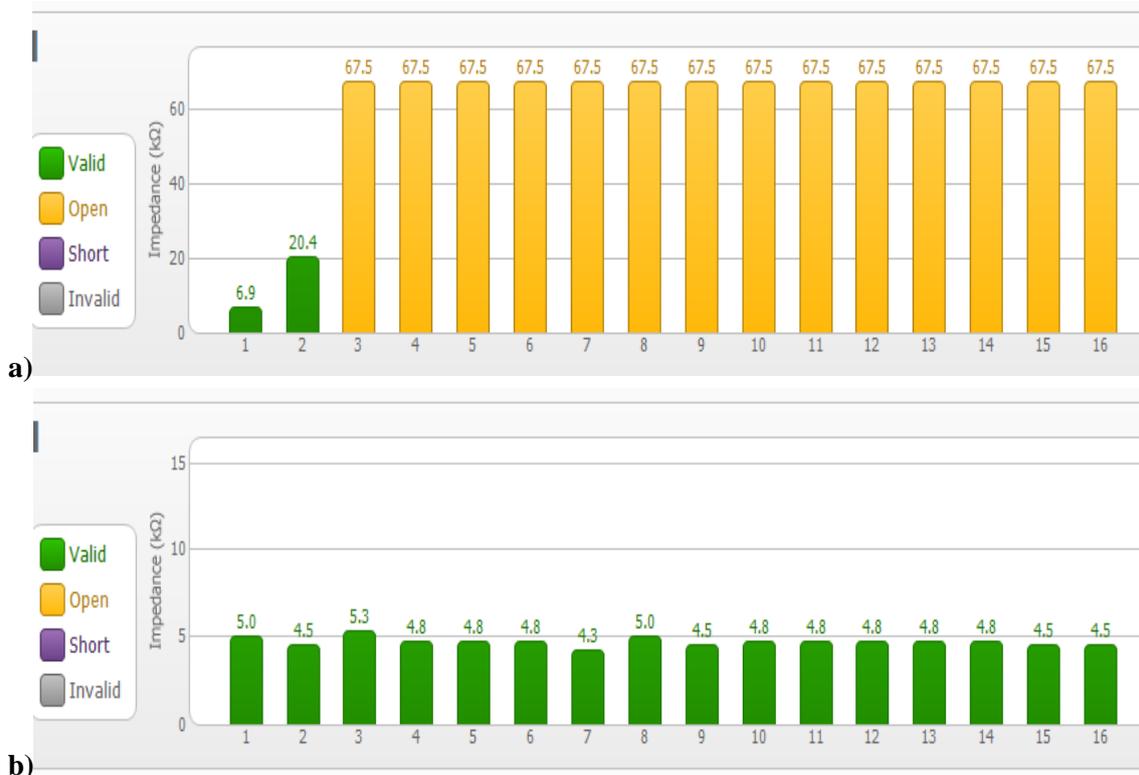


Figure 3. Impedances values measured intraoperative (a), respectively at the activation moment (b)

We have to note also that we did not obtain electrically compound action potentials after electric stimulation by neural response imaging software.

We verified the position of the cochlear implant's electrode array in the single cavity of the malformed right inner ear by post-surgery skull X-ray with two incidences (antero-posterior and lateral), which shown the portelectrode placed circularly near the inner wall, without any contacts between electrodes (fig 4). No complications were observed either during or after the surgery. The cochlear implant activation was performed one month after the implantation, when the patient had her first subjective sound perceptions.

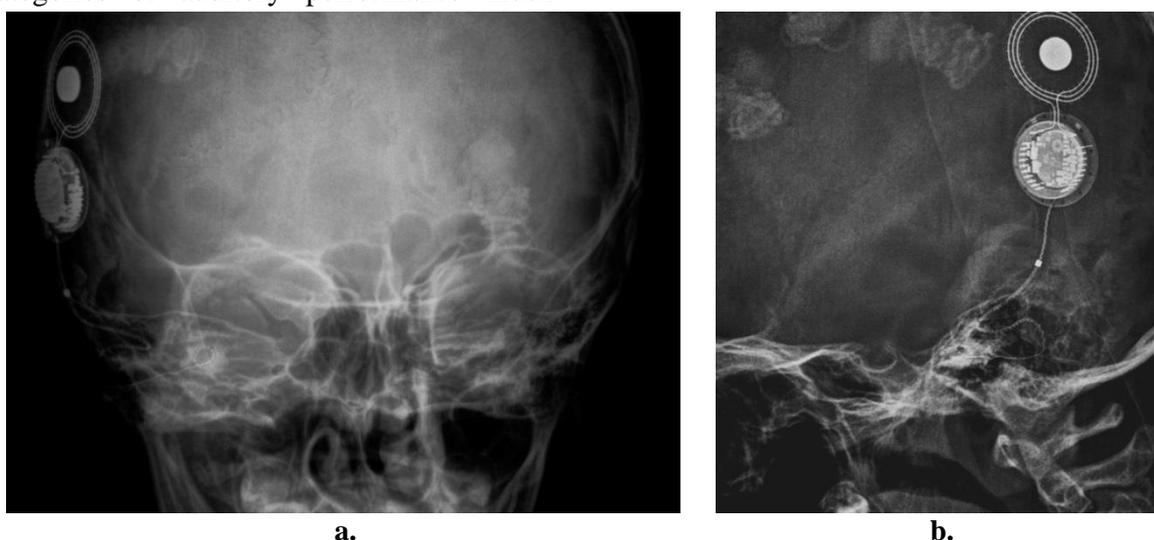
Postimplantation tonal auditory performance was measured using free field pure tone thresholds audiometry (PTA) in soundproof room. After the activation of the implant the patient was submitted to intensive speech therapy. We analysed the specialist reports and the family questionnaires to evaluate the speech performance by categories of auditory performance index

(CAP) described by Archbold S. (2) and by speech intelligibility rating scale (SIR) [3].

## RESULTS

The postoperative audiological performance for our patient was good; the average PTA threshold changed from 56,25 dB HL at first audiological evaluation (5 months postactivation) to 33,75 dB HL at 8 months. We noticed a small increase of PTA thresholds in the next year with an average of 36,25 dB HL at last evaluation (22 months postactivation).

According to PTA evolution, the patient achieved a maximum performance characterized by an index of 5 for categories of auditory performance (CAP): the patient understand common phrases without lip-reading/ understands conversation without lip-reading with a familiar talker; on the speech intelligibility rating scale (SIR) our patient was placed on category 3 (connected speech is intelligible to a listener who concentrates and lip-reads within a known context).



**Figure 4. Postoperative skull X-ray: cochlear implant electrode array position inside the common cochlea cavity (a – antero-posterior view; b – lateral view)**

## DISCUSSIONS

Inner ear malformations are found in 15% - 20% of patients with severe or profound

sensorineural hearing loss [4, 5].

Congenital malformations of inner ear may be considered in two broad categories

[6]:

a) malformation of membranous labyrinth (complete membranous labyrinth dysplasia, cochleosaccular dysplasia and cochlear basal turn dysplasia);

b) malformations of both the osseous and the membranous labyrinth [4] - classification system developed by Jackler et al. [7] and Sennauroglu and Saatci [8].

Recently, X-linked deafness has been recognized as a third type of incomplete partition [9]: the interscalar septa are present, but the modiolus is completely absent.

The common cavity has a frequency of around 16-25% of all ear malformations. In the 4th week of intrauterine life, development of inner ear stops [6, 10, 11]. Complete aplasia of the labyrinth and cochlear aplasia are very rare (1-3 % of all ear malformations), usually associated with other malformations that affect the temporal bone and results from interruption of otic development at the 3rd gestational week [11].

Previous studies about malformed inner ear, have documented some problems with surgical approach, cerebrospinal fluid gusher, electrode migration (fixation and stabilization of the electrodes), aberrant facial nerve. The type of malformation generally dictates the surgical approach for implantation. Ramos et al. [12] classified inner ear malformations into three groups with respect to the surgical possibilities of implantation: (a) gross malformation constituting surgical contraindications; (b) major malformations with high risk surgery; (c) minor malformations - the surgical risks is lesser than in the previous group. With the exception of cochlear aplasia, labyrinthine aplasia and agenesis of cochlear nerve, all cochleovestibular malformations can be implanted. Major malformations include common cavity and severe hypoplasia are often associated with cerebrospinal fluid leakage, meningitis. The development of

facial nerve has a complete interaction with otic capsule that is why it is important to look for an anomalous course of the facial nerve in inner ear dysplasia. Minor malformations are considered to be light hypoplasia, abnormalities of the aqueduct and abnormalities of the vestibule.

In our case, we had two different inner ear anomalies: cochlear aplasia on the left ear constituting a surgical contraindication for cochlear implantation and common cavity with vestibule malformation on the right ear which is an implantable type of anomaly and for this patient the only auditory rehabilitation possibility. There were no intraoperative surgical difficulties. The postoperative results are mainly related to the surgical placement of the electrode and residual neural nerve fibers [13]. For our patient we had an intracavity placement of the electrode, close to the inner wall, so close to the vestibular nerve fibers.

The results of cochlear implantation in cases with inner ear anomalies are generally good and appear to be related to the history of hearing loss, the degree of malformation and residual neural function [13].

Research on children with cochlear implants suggests that there is a "critical period" (a rather fixed time window of opportunity for change), a "sensitive period" and "age-related plasticity" (less abrupt transitions for the plasticity of the system) for the optimal auditory stimulation for normal hearing and speech development [14]. The central auditory pathways are maximally plastic for a period of about 3,5 - 4 years of life being the optimal time to implant a young congenitally deaf; the reduced plasticity of the central auditory system in congenitally deaf children implanted after 7 years is correlated with relatively poor development of speech and language skills [14, 15].

The studies on outcome of cochlear implantation in children with cochlear

malformations showed that the patients with different cochlear anomalies had the same or poorer results comparatively with normal cochlear implanted cases [16]. In terms of audiologic performance, children with inner ear malformations may perform very well. Predictors of good performance include the constellation of incomplete partition, enlarged vestibular aqueduct, and a dilated vestibule, isolated enlarged vestibular aqueduct and partial semicircular canal aplasia. Conversely, children with total semicircular canal aplasia, isolated incomplete partition, cochlear hypoplasia or common cavity appear to perform at a lower level. Children with common cavity tend to steadily and significantly improve their audiological skills over time and this development may, however, be highly individual [16, 17].

The particularity of our case consist in association of a complex inner ear malformation with a late cochlear implantation after 7 years of age, due to different factor: no newborn hearing screening program at his birth time, wrong hearing loss diagnosis and inadequate auditory rehabilitation for a long period of time including the critical and sensitive period for auditory system plasticity, lack of etiological diagnosis with no imaging exploration. The postimplantation speech and language performance was good considering all these factors with poorer results than in no

malformed implanted ears but with comparable postrehabilitation auditory skills reported in cases with common cavity cochlear implantation [17].

Another aspect to be discussed in this case is the bioethics and medical/legal considerations on the delay in right diagnosis and reahabilitation for our patient [18]. The early imagistic diagnosis for this patient could offer the benefits of an early cochlear implantation before the age of 4 years with better results.

## CONCLUSIONS

Children with inner ear malformations can safely benefit of cochlear implantation, a successful way of rehabilitation although complications should be expected and auditory responses may be highly variable and relatively moderate.

Cochlear implantation is a safe and effective treatment option in children with common cavity. The majority of children with common cavity derive significant audiological benefit from implantation. It is considered very important, in patients with congenital profound hearing loss, to follow a strict evaluation in order to adapt the treatment. It is mandatory to provide information about the inner ear and the vestibulocochlear nerve, high resolution CT and magnetic resonance imaging playing an important role in evaluation.

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