

## Case report

## Bilateral implantation in children with cochleovestibular nerve hypoplasia

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## ABSTRACT

To report on the outcomes of sequential bilateral cochlear implantation (CI) in children with inner ear malformation. The study design is a retrospective case study. The setting is a tertiary reference center. Two children presenting a profound bilateral congenital hearing loss with bilateral hypoplasia of the cochleovestibular nerves and hypoplastic external semicircular canal had a cochlear implant at respectively 16 months and 33 months. A second implant was proposed at respectively 17 and 20 months after the first implant. The main outcome measures are audiometry, perceptive results in closed and open set words (CSW and OSW) and oral production at follow-up. The first cochlear implant gave respectively mean thresholds at 60 dB and 70 dB. Bilateral CI showed mean threshold at respectively 40 dB and 55 dB. In case 1, perceptive assessment was 83% and 70% in respectively CSW and OSW with oral production and comprehension of sentences after 1 year follow-up. In case 2, the perceptive assessment showed no perceptive or linguistic evolution at 6 months follow-up. In cochleovestibular nerve hypoplasia, bilateral implantation could be discussed in cases of limited result after unilateral implant.

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## 1. Introduction

Since the advent of cochlear implantation, the indications for surgery have been more precisely defined and broadened. The main indicator remains in children with profound deafness showing limited gain with a powerful hearing aid [1–3].

Results of unilateral implants are variable. There are known factors such as the age at implantation, additional disabilities, cochlear malformation and/or ossification, and neuropathies.

Absence or limited results can be caused by device failure, tuning defaults, insufficient stimulation or neurological problems [4]. Revision implantation appears to be a safe and effective procedure [5,6]. After the mechanical problems have been eliminated and the tuning checked, other causes for those limited results must be researched [7]. In these cases of limited or absent response of the first implantation, the interest of sequential bilateral implantation should be discussed.

We report the case of two children with hypotrophic auditory nerve and inner ear malformation who had poor results with a unilateral implant. In those cases, sequential bilateral implantation was realized and their results are discussed in the following case report.

## 2. Cases reports

## 2.1. Child 1: girl, born November 2005

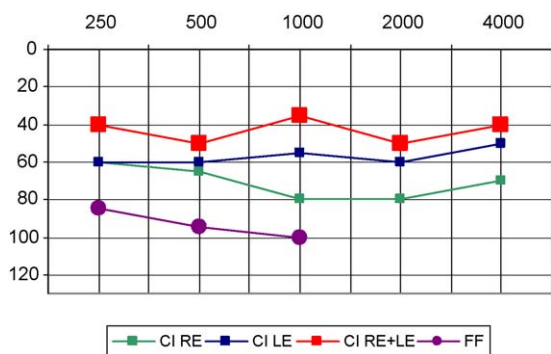
Profound deafness was confirmed with the auditory brainstem response (ABR) at 12 months of age, the evoked otoacoustic emissions (OAE) were present bilaterally. The mean audiometric thresholds in free field testing were at 70 dB with hearing aids (HA).

CT scan revealed normal cochlea and hypoplastic external semicircular canals (SCC) (Fig. 2). The magnetic resonance imaging (MRI) visualized normal cochleovestibular nerves (Fig. 3a–c). Genetic research remained negative (Connexine 26/30, mutation Q829X Otoferline, mitochondrial mutations). Suspicion of Waardenburg type II and minor CHARGE syndrome have been eliminated (respectively *PAXS*, *MITF*, *SOX10* and *CHD7*).

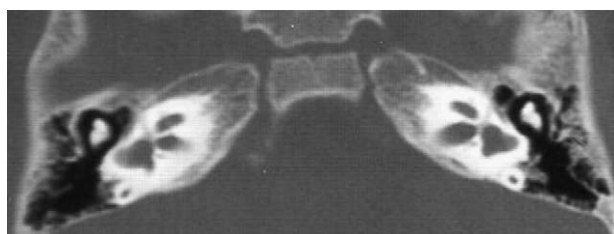
A right cochlear implantation (CI) was performed at 16 months of age (Nucleus<sup>®</sup> 24 RECA). There was no response with the pre-operative neural response telemetry (NRT).

At 6 months post-implant mean hearing responses with CI were at 50 dB. She showed some noise recognition capacities but 0% recognition in closed set words (CSW). There was no response to the neural telemetry (NRT) at this time. At 11 months post-implant, the parents described a drop in the auditory performance. The hearing level with CI was above 70 dB. Re-mapping with wider stimulation was not clinically efficient and did not improve NRT. Integrity test did not show device dysfunction; however;

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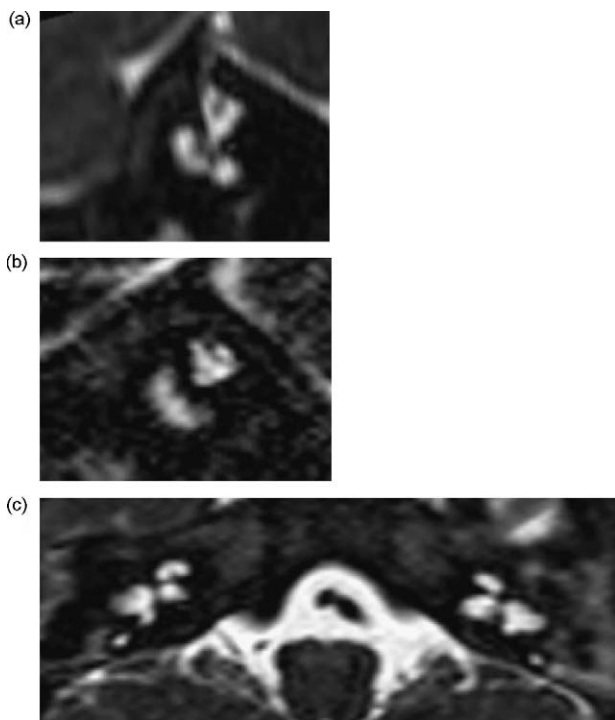
**Fig. 1.** Case 1: Hearing thresholds. FF: free field without HA/CI; CI RE: with cochlear implant right ear; CI LE: with cochlear implant left ear; CI RE + LE: with cochlear implant right + left ears, 12 months after bilateral implantation.



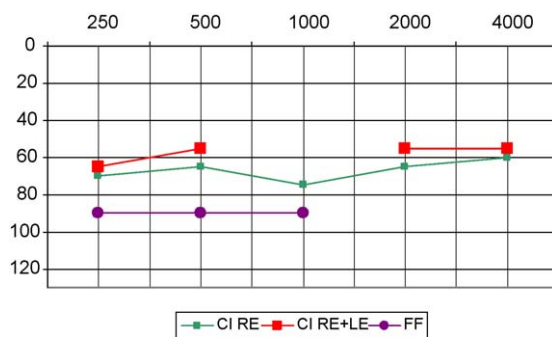
**Fig. 2.** Case 1: CT scanner (axial): bilateral abnormal external semicircular canal.

reimplantation was programmed (same device). Pre-operative NRT test was negative.

Three months after reimplantation, hearing threshold with CI remained at 70 dB, in spite of several tuning adjustments. She showed some noise detection but 0% identification in CSW. Speech production was limited to a few words.



**Fig. 3.** Case 1: (a) MRI (parasagittal); internal auditory canal; left ear. (b) MRI (parasagittal); internal auditory canal; right ear. (c) MRI axial; inferior vestibular nerve branch only, no cochlear branch visible.



**Fig. 4.** Case 2: Hearing thresholds. FF: free field without HA/CI; CI RE: with cochlear implant right ear; CI LE: with cochlear implant left ear; CI RE + LE: with cochlear implant right + left ears, 6 months after bilateral implantation.

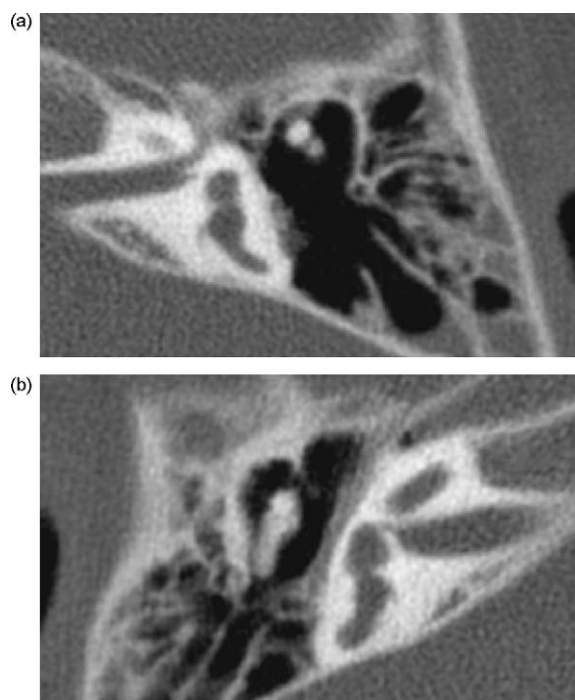
MRI re-lectures suggested hypotrophic auditory nerves on both sides.

A second implant was performed at 33 months of age (Nucleus<sup>®</sup> 24 RECA), no NRT response at surgery.

One year after bilateral implantation, the audiogram showed thresholds at 70 dB when switching on the right CI (+30 months post-operative), 60 dB when switching on the left CI (+12 months). With both CI, thresholds were at 40 dB (Fig. 1). She showed 83% in CSW, 70% in OSW, and had acquired oral production (short sentences) and oral comprehension.

2.2. Child 2: boy, born January 2004

Profound deafness was confirmed at 16 months with the ABR, the OAE were absent. The mean audiometric thresholds with HA were at 70 dB. He had persistent arterial canal, cryptorchidism and syndactyly. The CT scan described bilateral dilation of the vestibule and of the superior SCC, agenesis of the lateral SCC, normal cochlea (Fig. 5). The MRI showed hypotrophic cochleovestibular nerves on



**Fig. 5.** Case 2: (a) CT scanner (axial) left ear: dilation of the vestibule and of the superior semicircular canal. (b) CT scanner (axial) right ear: dilation of the vestibule and of the superior semicircular canal.

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