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# Cochlear implantation in children with anomalous cochleovestibular anatomy

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ABSTRACT

*Objective:* The aim of the study is to assess the audiological and surgical outcome after cochlear implantation in children with inner ear malformation and to compare them with a group of congenitally deaf children implantees without inner ear malformation.
*Introduction:* Children with profound sensorineural deafness with malformations of the inner ear represent a challenge, accounting to 5–15% of congenital sensorineural deafness. These cases were originally regarded as a contraindication for cochlear implant surgery.
*Methods:* Retrospective study of 26 patients with congenital inner ear malformation, from a total population of 329 cochlear implant patients. Radiological evaluation with high resolution computed tomography and magnet resonance was performed to all patients in order to evaluate all the preoperative conditions. All patients were tested using European Portuguese word discrimination tests (monosyllabic test, number test and sentences test), capacity of auditory performance (CAP) and speech intelligibility rating (SIR).
*Results:* In all 7.9% of deaf children in our center study have inner ear abnormalities. All children underwent successful implantation. CAP yielded an average 7.1 (+/−1.7), SIR 4.3 (+/−1.0). The children without inner ear abnormalities did not achieve statistically significant better scores. Two children had a perilymph gusher, and there were no other complications.
*Conclusion:* Cochlear implantation can be successfully performed in children with inner ear malformation. Audiological performance after cochlear implantation in malformed inner ears is comparable to that found in other congenitally deaf patients. The risk of CSF leak is associated with inner ear abnormalities and should be anticipated during surgery.


## 1. Introduction

Cochlear implantation in children with congenital and acquired deafness is an accepted auditory rehabilitation treatment for more than 20 years [1].

The diagnosis and management of children with profound sensorineural deafness with malformations of the inner ear represent a challenge, and these cases were originally regarded as a contraindication for cochlear implant (CI) surgery in severe inner ear abnormalities [2,3,4]. The first case of inner ear malformation to receive a CI was reported by Mangaberia-Albernaz [5] in a Mondini deformity, showing that CI surgery was possible in inner ear malformations with a successful outcome.

A classification system of inner ear malformations based on embryological genesis was presented in 1987 by Jackler et al., where inner ear malformations were divided into two categories—category A with cochlear malformation and

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category B with normal cochlea but abnormal vestibule/semicircular canals. Category A consists of Michel aplasia, cochlear aplasia, cochlear hypoplasia (CH), incomplete partition (IP) and common cavity (CC) [2]. In 2002, Sennaroglu suggested an extension where IP was subdivided into two categories, reviewing this classification in 2010 [4,6]. According to Sennaroglu and Saatci the Michel deformity was defined as a full labyrinthine aplasia; the cochlear aplasia was defined as the absence of cochlea on the presence of a dilated or hypoplastic vestibule; the CC when the cochlea and vestibule form a unique cavity, representing both undifferentiated structures; the CH is described as a cochlea with smaller dimensions and can be divided into three types, CH type I (bud-like cochlea) the cochlea resembles a small bud arising from the Internal Auditory Canal (IAC), the internal architecture is severely deformed; no modiolus or interscalar septa can be identified; the CH type II (cystic hypoplastic cochlea) cochlea is smaller in its dimensions with no modiolus and interscalar septa, but its external architecture is normal; the CH type III (cochlea with less than two turns) the cochlea has a shorter modiolus and the overall length of the interscalar septa is smaller, resulting in a reduced number of turns (less than two turns), the internal and external architecture (modiolus, interscalar septa) is similar to that of a normal cochlea, but the dimensions are smaller hence the lesser number of turns, the vestibule and the SCCs are hypoplastic. Three different types of IP can be diagnosed according to the defect in the modiolus and the interscalar septa, IP type I is represented by a dilated cystic vestibule accompanied by an empty cystic cochlea; the IP type II (Mondini deformity) is represented by a cochlea where only the basal part of the modiolus is present, the apical part of the modiolus and the corresponding interscalar septa are defective and a dilated vestibule and an enlarged vestibule aqueduct (EVA) are present; IP type III is the type of cochlea observed and reported in X-linked deafness, the interscalar septa are present but the modiolus is completely absent, placed directly at the lateral end of the IAC instead of its usual anterolateral position For the evaluation of the enlarged vestibule aqueduct, the midpoint between the posterior labyrinth and operculum is larger than 1.5 mm [2,3]. Labyrinthine malformation includes total or partial aplasia of the semi-circular channels.

Jackler et al. found that bone malformations of the inner ear are relatively frequent anomalies that represent 5–15% of congenital sensorineural deafness [2]. Recent studies, that used high resolutions computed tomography, reported an incidence as high as 30–35% [7,8]. The other 70–80% are due to membrane malformations in which the inner ear bony architecture is preserved and the lesion is located at a cellular level [2].

Theoretically, the outcome of CI might be impeded in patients with cochlear malformations due to possible disorganization or misplacement of the remaining neural structures and due to more complex anatomy that may result in complicated surgery [3,9–11]. Nevertheless, several studies have shown successful results of cochlear implantation regarding both surgical technique and audiological outcomes – comparable to that of other profoundly deaf children – except for more severe inner ear malformations like cochlear hypoplasia (CH) or common cavity (CC) [3,7,12–17].

Overall, the functional outcome of CI depends on the duration of deafness before CI, age at implantation, cause of deafness, co-morbidity like mental retardation and CHARGE syndrome [15–22].

The purpose of the study was to present our experience about the surgical and audiological outcome of cochlear implants in children with inner ear malformations, and compare them with a group of children with profound congenital sensorineural deafness without inner ear abnormalities.

## 2. Materials and methods

A retrospective analysis was made involving 329 children, which have had cochlear implants performed in the Cochlear Implant Unit Department of Otorhinolaryngology at Centro Hospitalar Universitário de Coimbra, between 1992 and 2014. Twenty-six of these children were diagnosed with congenital malformations of the inner ear. The gathered data consisted on the subject's age at the time of surgery, clinical and radiological characteristics, surgical and perioperative complications.

The classification of inner ear malformations was performed by two independent otologists and one radiologist, reviewing high-resolution computerized tomography and magnetic resonance imaging of each patient, according to the classification system of Sennaroglu [6] CH type III and IP type II were regarded as major inner ear malformations where the other abnormalities encountered in the study were classified as minor inner ear malformations.

Different electrodes arrays configurations were used on our patients. After cochlear implantation and its activation and programming, the number of active electrodes was assessed and recorded.

The postoperative performance was evaluated and compared with the results collected from the children with profound sensorineural deafness without inner ear changes of our department.

This second group, which acted as a control group, consists of 303 (92.1%) children, with an average implantation age of 43 months, ranging from 12 to 94 months, and minimum follow-up of 36 months with an average 9,14 ± 3,78 years.

The postoperative audiological results were obtained using European Portuguese word discrimination tests (monosyllabic word test, number word test and words in sentence test), considering that the lists were adapted to the children's age, at a 65 dB SPL, with the cochlear implant activated, in a soundproof cabin without visual cues. The results were registered taking into account the percentage of correctly repeated items. For the monosyllabically and number test, the percentage of correct phonemes was also taken into account. Categories of Auditory Performance (CAP—eight performance categories arranged in order of increasing difficulty with very high inter-user agreement making it a reliable tool in measuring the auditory capacity after CI) and Speech Intelligibility Rating (SIR—index 1–5 with increasingly better language, and is a valid tool for testing children's speech intelligibility after CI) were also measured.

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