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OUTCOMES OF COCHLEAR IMPLANTATION IN INNER EAR MALFORMATION

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The cochlear implants are the most successful neural prothesis to date but in patients with internal ear anomalies there are several difficulties. Inner ear malformations (IEM) are the cause of congenital sensorioneural hearing loss in approximately 20% of hearing impaired children.

Using the most commonly accepted classification of Jackler et al. Sennaroğlu and Saatçi reclassified the most commonly accepted inner ear anomalies based on state-of-art computerized tomography (CT) and magnetic resonance imaging (MRI) findings. In their study, cochlear, vestibular, semicircular canal (SCC), internal acoustic canal (IAC), and vestibularcochlear aqueduct malformations were classified into subgroups. As a result of this examination, cochlear malformations were divided into 7 groups as Michel deformity, common cavity, cochlear aplasia, hypoplasic cochlea, incomplete partition type I (IP-I), incomplete partition type II (IP-II/Mondini deformity), and incomplete partition type III (IP-III); vestibular malformations were divided into 3 groups as vestibular dilatation, SCC malformations, and IAC anomalies.

In this study it includes incomplete partition of cochlea type I, II and III (IP I, IP II and IP III), cochlea hypoplasia type II, III and IV (CH-2, CH-III and CH-IV), large vestibular aqueduct syndrome (LVAS) and isolated semicircular canal malformation (ISCC).

OBJECTIVES

To evaluate the outcomes of cochlear implantation in patients with severe to profound sensorioneural hearing loss due to IEM when compared to patients without IEMs.

METHODS

In this retrospective case control study, 220 patients who underwent cochlear implantation between May 2007 and September 2024 were analyzed at Pediatric Otorhinolaryngology Department of Unidade Local de Saúde de São José, a Tertiary Center in Lisbon. Twenty eight out of 220 patients who were diagnosed with inner ear malformations were included in the study. The patients were divided into control group I (normal inner ear), group II (mild IEM: IPII, LVAS, ISCC and CH-4) and group III (severe IEM: CH-2, CH-3, IPI, IPIII).
Audiological outcomes were compared by Speech perception test (open or closed testing) and by measuring Categories of auditory performance (CAP) and Speech intelligibility rating (SIR). In addition to the auditory performances, we also analyze intraoperative findings and complications.

RESULTS AND DISCUSSION

In this study were included, 28 patients with IEM, 44 ears in total, and 28 patients in control group.

Numbers of patients/ears and type of IEM

	Nº Patients	Nº ears
IP1	1	2

IP2	10	19
IP3	1	1
HC 2	2*	3
HC 3	1**	1
HC 4	6	9
LVA	4	5
ISCC	3	4
TOTAL	28	44

*One patient has CHARGE
** A CHARGE patient

Figure 1. Gender distribution

Figure 2. Type of cochlear implantation

Operation Findings	Radiological Findings
Oozing	IP2 (10 out of 19 ears) LVA (1 out of 5 ears)
Gusher	IP1 (2 out of 2 ears) IP3 (1 out of 1 ear)
Facial nerve pathway anomaly	IP1 (2 out of 2 ears) CH 2 and CH 3 in CHARGE patients
Round window agenesia	Isolated SCC malformations (1 out of 4 ears

Figure 3. Distribution of operation findings among inner ear malformation

Figure 4. Pure tone average after one or two months after CI compared to 2 years after CI. There is a statistically significant difference between group 3 and 1 ($p=0,033 < p= 0.05$) and between group 3 and 2 ($p=0,041 < p=0.05$)

Both control group patients and in IEM groups showed significant improvements in PTA but these improvements were less evident in severe IME group (group III). Evaluation of Auditory Responses to Speech test battery was composed of 2-syllable open-ended words and 2-syllable closed-ended words in 9 patients of group II, in 3 patients of group III and in the control group. In all 3 groups, 1 to 2 years after cochlear implantation, the voice discrimination and recognition increased steadily. The improvement reached 89% in the group II, 76% in the group III and 95 in the group I. The poorest speech perception was seen in IP1 and HC-2, CH-3 patients.

	Mean CAP score (1 to 2 year)	Mean SIR score (1 to 2 years)
Group I	6,91	4,34
Group II	6,71	4,18

Group III	4,41	3,15

Figure 5. Comparative CAP and SIR scores among groups. There was a statistically significant difference in the CAP as well as SIR scores between group III and group I ($p<0,001$) and between group III and II ($p<0,001$).

CONCLUSION

Cochlear implants have the potential to provide auditory rehabilitation to individuals with IEMs. Despite severe IEM have the poorest audiological and speech perception outcomes, patients like to use their implants. However, it is important to manage family expectations, particularly when considering patients with more severe inner ear malformation.