FACULDADE DE MEDICINA
UNIVERSIDADE D COIMBRA

MESTRADO INTEGRADO EM MEDICINA – TRABALHO FINAL

SOFIA MONIZ ROCHA

HEARING REHABILITATION BY COCHLEAR IMPLANT IN CONGENITAL COCLEOVESTIBULAR MALFORMATIONS

ARTIGO CIENTÍFICO

ÁREA CIENTÍFICA DE OTORRINOLARINGOLOGIA

Trabalho realizado sob a orientação de:

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MARÇO/2023

HEARING REHABILITATION BY COCHLEAR IMPLANT IN CONGENITAL COCHLEOVESTIBULAR MALFORMATIONS

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Abbreviations

IEM – Inner ear malformation

HL – Hearing loss

CI – Cochlear implant

Hz – Hertz

SPSS – Statistical Package for Social Sciences

Resumo

Introdução: As malformações congénitas do ouvido interno são uma causa rara de surdez congénita. Esta condição afeta o desenvolvimento dos doentes. Com vista a melhorar o desenvolvimento, os implantes cocleares são utilizados como tratamento de escolha e têm sido estudados ao longo dos anos com o intuito de comprovar os benefícios audiológicos do seu uso. O nosso estudo tem como objetivo avaliar o uso de implantes cocleares como reabilitação auditiva em doentes com malformações do ouvido interno no nosso centro hospitalar.

Métodos: Um estudo observacional retrospetivo foi conduzido com 36 pacientes com malformações e 633 sem malformações, após implantação no nosso hospital. Os resultados obtidos nos testes de perceção e descriminação da fala no seguimento após a cirurgia foram recolhidos da base de dados do hospital de forma a comparar ambos os grupos.

Resultados: Foi identificada uma diferença estatisticamente significativa na audiometria aos 4000Hz, no entanto, os restantes resultados foram sobreponíveis em ambos os grupos. Os testes de descriminação demonstraram uma diferença estatisticamente significativa nos testes de monossílabos, números e 100 palavras ao telefone mas não demonstraram diferença nos restantes testes. Houve falha de dados em alguns testes.

Discussão: Os resultados mostraram que doentes com malformações apresentam valores semelhantes aos sem malformações na audiometria após implantação coclear. Não obstante, o grupo de estudo apresentou melhores resultados nos testes de perceção em comparação com o grupo de controlo, o que vai contra alguns artigos já publicados.

Conclusão: Este estudo proporciona uma visão da eficácia da reabilitação auditiva com implantes cocleares em doentes com malformações. Apesar de haver algumas limitações, os resultados comprovam que o uso de implante coclear nestes doentes permite uma melhoria significativa na performance auditiva e perceção do discurso dos nossos doentes.

Palavras-chave: Patologia do ouvido, Malformações do Ouvido Interno, Deformidade de Michel, Hipoplasia Coclear, Partição Incompleta.

Abstract

Introduction: Inner ear malformation is a rare cause of congenital hearing loss. This condition impacts the development of patients. Aiming to improve the development, cochlear implant is used as standard treatment and has been studied since its first use to demonstrate audiological benefits. Our study purpose is to evaluate the use of cochlear implant as hearing rehabilitation for patients with inner ear malformations at our clinic center.

Methods: An observational retrospective study was conducted with 36 patients with and 633 without malformations after cochlear implantation in our clinic center. Data from speech perception and discriminations tests were obtained from the hospital database from post-surgery follow-up and were analyzed comparing both groups in each test.

Results: A statistically significant difference was found in free-field tone audiometry at 4000Hz, but the overall results were comparable between both groups. Discrimination tests showed significant differences in the monosyllabic, number and 100 words on the phone tests, but no significant differences were found in the others. There were some results missing from each test.

Discussion: The results showed that cochlear implantation in patients with malformations had similar outcomes in free-field tone audiometry compared with the control group. However, the study group had better results in speech perception tests than those with normal cochlear anatomy, which contradicts some previously published studies.

Conclusion: This study provides valuable insights into the efficacy of cochlear implant as hearing rehabilitation method for patients with inner ear malformation. While there are some limitations, the findings suggest that cochlear implant can significantly improve auditory performance and speech perception outcomes in our patients.

Keywords: Ear Diseases, Inner Ear Malformation, Michel Deformity, Cochlear Hypoplasia, Incomplete Partition

Introduction

Congenital hearing loss affects approximately 2-3/1000 live births worldwide.[1] Inner ear malformations (IEM) can hinder the development and function of the cochlea and other structures involved in hearing, leading to hearing loss (HL). IEM are responsible for 20% of all cases of HL.[1–4]

HL in children can have a profound impact on their cognitive, social and emotional development. According to Korver *et al.*[5], HL can lead to delays in speech and language acquisition, academic difficulties and social isolation. Early identification and intervention are crucial for minimizing its impact on a child's development.

Due to the rarity of IEM, it's crucial to have a worldwide accepted classification for effective diagnosis and treatment. In 1987, Jackler *et al.*[6] created a classification based on embryological genesis, dividing the malformations in two big categories. Category A consisted of cochlear malformations and B consisted of normal cochlea but abnormal vestibule or semicircular canals. In 2002, Sennaroglu[7] reclassified this system based on computer tomography and magnetic resonance imaging findings, dividing the malformations into six different groups. This classification was updated in 2017 by Sennaroglu[4], becoming the most widely accepted classification system worldwide. The updated classification system divides IEM into eight groups based on similar radiological and audiological findings. The classification includes complete labyrinthine aplasia (Michel deformity), rudimentary otocyst, cochlear aplasia, common cavity, cochlear hypoplasia with subgroups from I to IV, incomplete partition of the cochlea with subgroups from I to III, enlarged vestibular aqueduct and cochlear apertures abnormalities.

With a standardized classification system, clinicians can make informed decisions about treatment options. Although cochlear implants (CI) were initially contraindicated for the hearing rehabilitation of IEM, since Albernaz[8] used a CI as treatment option in 1983, the number of surgeries has increase, and nowadays, the CI is a standard surgical approach for most IEM. CI can be a viable treatment option for children with IEM and HL. According to Naples *et al*.[9], CI can bypass the damaged or underdeveloped structures in the inner ear and stimulate the auditory nerve directly.[1,3,4,10–14]

Numerous studies have demonstrated the audiological benefits from using CI for IEM treatment. Despite changes in surgical approaches and the CI over the years, aiming to improve the outcomes and reduce the complications, treatment decisions still depend on the type of malformation, condition of the cochlear nerve and preoperative audiological findings. Treatment options include CI and/or auditory brainstem implantation, with cerebrospinal fluid gusher, meningitis and facial nerve anomalies being the two main surgical challenges[4,10,13,15,16]. Nevertheless, according to Kocabay *et al.*[1], the individualized fitting parameters required for CI can be more complex in children with IEM and the potential for limited auditory performance outcomes should be considered. Additionally, children with IEM may have more comorbidities that can impact CI outcomes.[17–19]

Since IEM is less common, it is more challenging to study this condition accurately. Therefore, the aim of this study is to evaluate the outcomes of using CI as a treatment option for IEM in patients who underwent surgery at Coimbra Hospital and University Center. The study aims to provide outcomes that reflect our practical reality.

Materials and Methods

An observational, retrospective study was conducted with two groups: one group consisting of 36 patients with IEM who underwent cochlear implantation, the study group, and one with 633 with CI and without IEM as the control group. All patients received treatment at Coimbra Hospital and University Center from January 1992 to December 2022 and were followed up.

The study included 669 individuals with CI, both adults and children were selected. All the data were obtained through consultation of a digital hospital database, and patients were classified according to the Sennaroglu[4] system from 2017. The main inclusion criterion was the existence of recorded results of post-implantation hearing tests. Having normal imaging, total absence of data and refusal to participate on the study were the main exclusion criterion.

To compare the outcomes between the two groups, results were collected from different tests, including free-field tone audiometry ranging from 250 to 6000 hertz (Hz) and discrimination of monosyllables, number, sentences, sentences on the phone, 100 words and 100 words on the phone. The results of the tests were presented in percentage and boxplot graphics. Some patients did not present data from the tests that were used. However, this was not considered a criterion of exclusion in order to enhance the overall statistic power of the sample.

Statistical analysis was performed using the Statistical Package for Social Sciences (SPSS) 23 software. To confirm the normality of distribution and homogeneity of variances, the Levene's test and t-test were used. Statistical significance was determined as p-value of less than 0,05.

Results

Free-field tone Audiometry

The study group consisted of 36 patients. However, only 26 of these results were valid (72.2%) with 10 results missing (27.8%). On the control group, there were 421 valid results (66.5%) and 212 missing (33.5%).

The t-test only showed a statistical significance in the free-field tone audiometry at 4000Hz with a p-value of 0.014 (Table 1). Nevertheless, the results of the free-field tone audiometry were comparable between the group with IEM and the control group (Figure 1). The standard deviation was larger in the control group and there are more outliers.

Table 1 – Free-field tone audiometry statistical analysis comparing the study with the control group.

		Levene's Test for Equality of Variances		T-test for Equality of Means						
									95% Confidence interval of the difference	
		F	Sig.	t	df	Sig (2-tailed)	Mean Difference	St. Error Difference	Lower	Upper
250Hz final	Equal variances assumed	4.004	0.046	1.8990	445	0.058	3.160	1.664	-0.110	6.431
	Equal variances not assumed			1.391	26.542	0.176	3.160	2.271	-1.504	7.824
500 Hz final	Equal variances assumed	0.035	0.852	1.416	445	0.157	1.960	1.384	-0.759	4.679
	Equal variances not assumed			1.374	27.961	0.180	1.960	1.426	-0.962	4.881
1000 Hz final	Equal variances assumed	1.298	0.255	0.913	445	0.362	1.136	1.244	-1.308	3.581
	Equal variances not assumed			1.067	29.566	0.295	1.136	1.065	-1.041	3.313
2000 Hz final	Equal variances assumed	0.001	0.973	1.519	445	0.129	2.021	1.330	-0.593	4.635
	Equal variances not assumed			1.386	27.568	0.177	2.021	1.458	-0.967	5.009
4000 Hz final	Equal variances assumed	1.947	0.164	2.459	445	0.014	3.425	1.393	0.688	6.163
	Equal variances not assumed			1.752	26.444	0.091	3.425	1.955	-0.591	7.441
6000 Hz final	Equal variances assumed	0.283	0.595	1.477	445	0.140	2.165	1.466	-0.716	5.045
	Equal variances not assumed			1.328	27.483	0.195	2.165	1.630	-1.177	5.507

Levene's Test: if Sig<0.05, equal variances are not assumed. The p-value ("Sig (2-tailed)") from T-test should be used according to Levene's Test result.

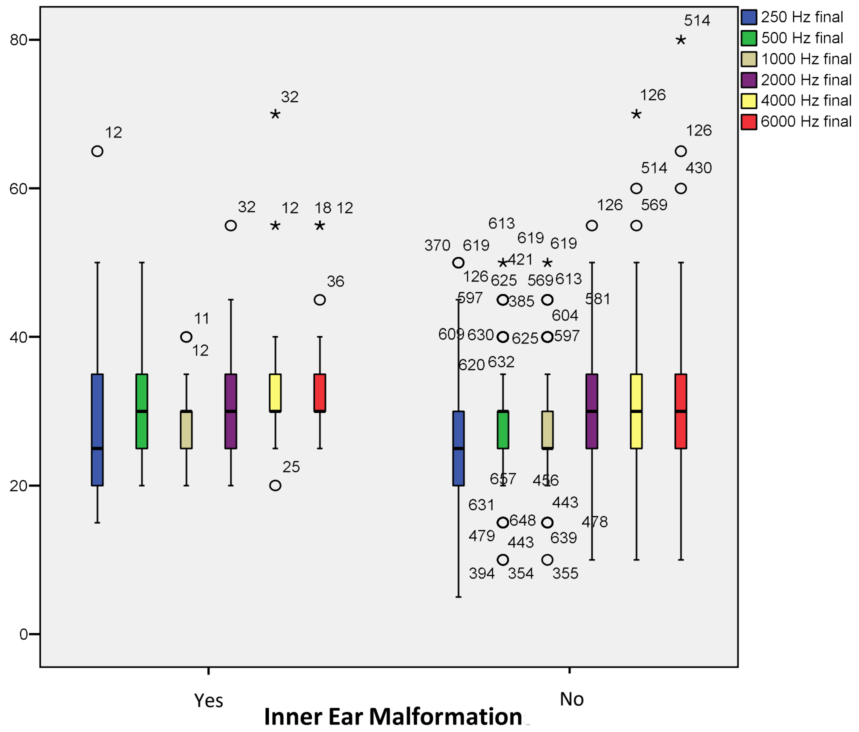

Figure 1 – Boxplot comparing the results from free-field tone audiometry between the study and the control group.

Discrimination tests

In the group with CVM, 21 valid results (58.3%) were obained for monosyllabic, number and setences test with 15 missing results (41.7%). In contrast, for the group without IEM, there were 363 valid results (27.3%) and 270 missing (42.7%).
For the setences on the phone,100 words and 100 words on the phone tests,15 valid results (41.7%) and 21 missing (58.3%) were obtained for the study group, while the control group had 245 valid (38.7%) and 388 missing results (61.3%).

The statistical analysis of the data demonstrated that there was a significant difference in the monosyllabic test, in the number test and in the 100 words on the phone test, with p-value of 0.000, 0.001, 0.041 respectively (Table 2). However, there was no statistical significance found in the other tests: sentences, sentences on the phone and 100 words (Table 2). Although there was no statistical significance, graphical representation suggests that the group with IEM had higher results on these tests (Figure 2 and 3).

Table 2 – Discrimination tests statistical analysis comparing the study with the control group.

		Levene's Test for Equality of Variances		T-test for Equality of Means					95% Confidence interval of the difference	
		F	Sig.	t	df	Sig (2-tailed)	Mean Difference	St. Error Difference	Lower	Upper
Monosyllabic Test	Equal variances assumed	7.308	0.007	2.754	391	0.006	12.0746	4.3843	3.4548	20.6943
	Equal variances not assumed			4.755	28.649	0.000	12.0746	2.5392	6.8786	17.2705
Number Test	Equal variances assumed	6.653	0.010	1.511	390	0.132	6.08392	4.02618	-1.83182	13.99966
	Equal variances not assumed			3.712	45.093	0.001	6.08392	1.63907	2.78285	9.38498
Sentences Test	Equal variances assumed	2.985	0.085	1.826	383	0.069	12.22619	6.69455	-0.93648	25.38886
	Equal variances not assumed			2.305	24.076	0.030	12.22619	5.30339	1.28237	23.17002
Sentences on the phone Test (% of right answers)	Equal variances assumed	0.066	0.798	0.939	273	0.349	7.18895	7.65693	-7.88499	2.26288
	Equal variances not assumed			0.934	16.883	0.364	7.18895	7.69690	-9.06291	23.44080
100 words Test (% of right answers)	Equal variances assumed	3.741	0.054	1.487	309	0.138	5.83894	3.92569	-1.88553	13.56340
	Equal variances not assumed			3.511	34.099	0.001	5.83894	1.66303	2.45961	9.21826
100 words on the phone Test (% of right answers)	Equal variances assumed	3.958	0.048	1.325	268	0.186	7.947	5.996	-3.858	19.751
	Equal variances not assumed			2.171	21.482	0.041	7.947	3.660	0.346	15.547

Levene's Test: if Sig<0.05, equal variances are not assumed. The p-value ("Sig (2-tailed)") from T-test should be used according to Levene's Test result.

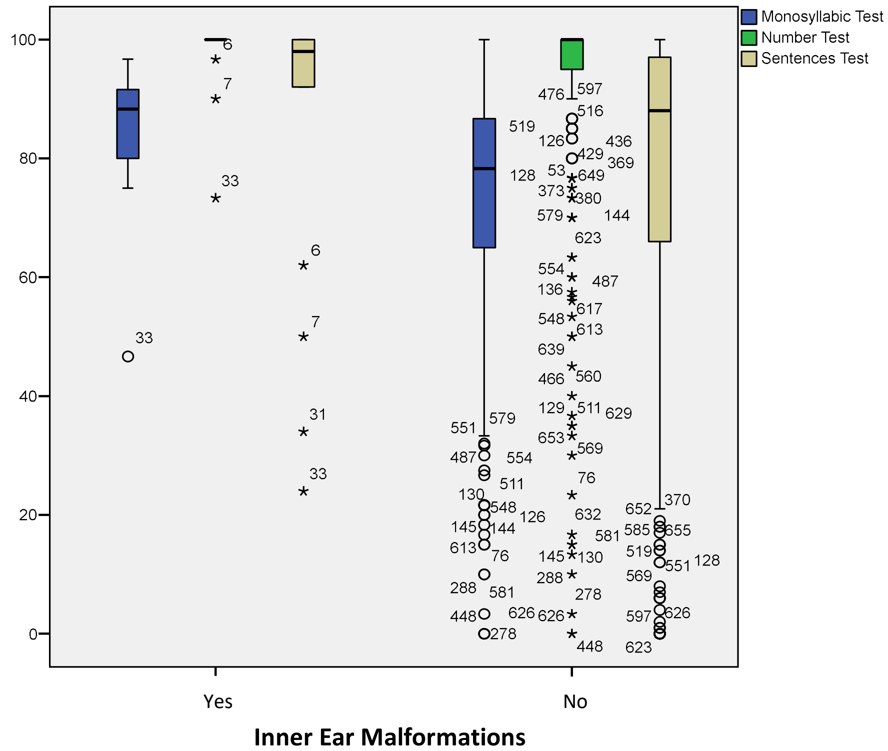

Figure 2 – Boxplot comparing the results from monosyllabic, number and sentences tests between the study and the control group.

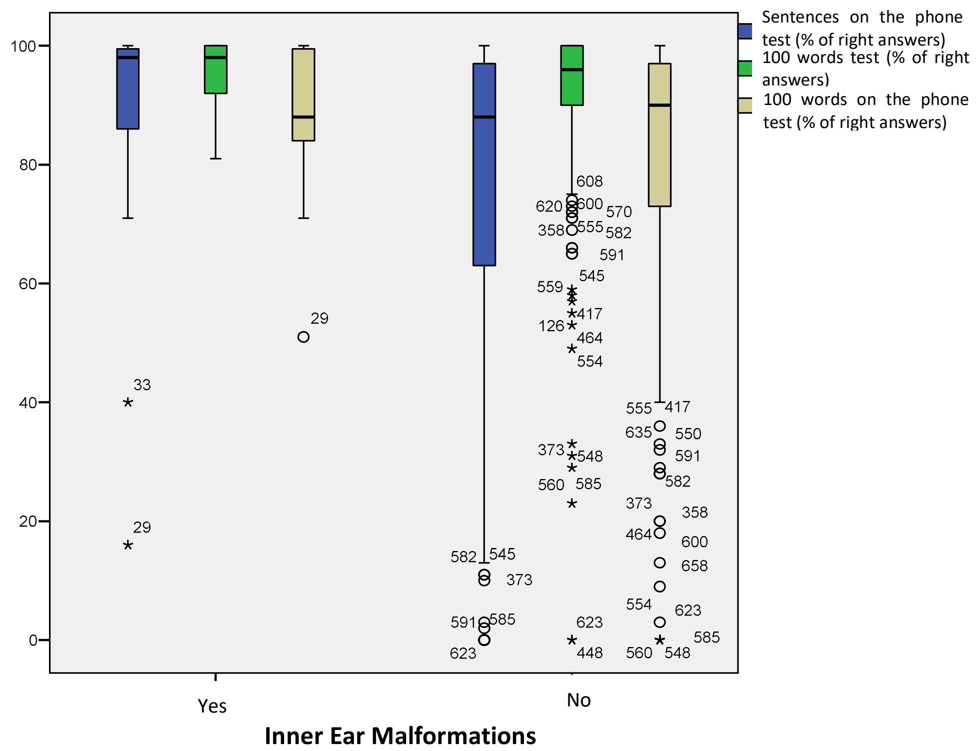

Figure 3 – Boxplot comparing the results from sentences on the phone, 100 word and 100 words on the phone tests between the study and the control group.

Discussion

IEM is a rare condition, accounting for 20% of congenital HL cases. Our study found a prevalence of 5,38% which is comparable to Jackler *et al.*[6] and Melo *et al.*[11] findings of 5 to 15% and 7,9%, respectively. On the other hand, it was lower than Daneshi *et al.*[3] prevalence of 31.5%. The use of CI, through times, has been recognized as the standard surgical approach for most of cases with IEM with numerous studies demonstrating its audiological benefits in hearing reabilitation.

We obtained a sample of 36 patients from the digital hospital database, with the main inclusion criterion being recorded results of post- implatation hearing tests. The data collected was from audiometric performance and speech perception evaluation.

The aim of this study was to evaluate the outcomes of CI as hearing reabiliatiton method in patients with IEM who underwent surgery at our clinical center.Our data showed that the results of the free-field tone audiometry were similar between the IEM group and the control group, as demonstrated by Isaiah *et al.*[13], Onan *et al.*[2] and Celik *et al.*[20]. However, our patients had better results in speech perception tests than those with normal cochlear anatomy, as demonstrated by statistical significance in monosyllabic, numbers and the 100 words on the phone test. These findings contradict some published articles, such as Arnoldner *et al.*[10] and Daneshi *et al.*[3], which showed lower results for the group with IEM. Daneshi *et al.*[3] reported the largest multicenter study of CI in children with IEM and found that although CI could significantly improve auditory performance and speech production outcomes in these children, those with IEM had lower performance and outcomes than those without IEM. One difference between our study and Daneshi *et al.*[3] study is that they had a larger number of participants, including major IEM such as common cavity and cochlear hypoplasia.

Nevertheless, all the studies indicate that IEM benefits from using CI with encouraging results in audiometric and speech perception tests.

We can deduce from speech discriminations tests graphics (Figure 2 and 3) that, although the study group had better results there was greater variability in the control groups, as indicated by the longer standard deviations. This limitation is due to the small sample size of the study group, which had only 36 patients compared to the control group with 633 patients, and may limit the interpretation of the results.

The fact that our patients only presented with minor IEM may have contributed to our better results, as studies have shown that CI outcomes are better for minor IEM than major IEM. Additionally, our clinic center ability to make an early diagnosis of CI, as compared to the large number of possible causes for HL in control group, may have also contributed to the better outcomes in the study group.

It should be noted that patients had speech therapy after the surgery. The number of appointments varied among patients, which may have impacted the results presented on the tests. Also, the patients included in the study underwent implantation at different ages, which could have influenced the outcomes.

Another limitation of our research was missing data due to the retrospective nature of the study, which means we could only use data collected during follow-up. Furthermore, comparing the control group to the study group, we can infer that the study group had a more specific and rigorous treatment due to all diferent possible aetiologies of HL presented on control group.

Despite these limitations, our study has had a significant clinical impact, as treatment for IEM is not standard worldwide and prevalence is low. It has allowed doctors to gain better insight into treatment situation in this hospital compared to data collected worldwide since all patients had surgery at our hospital. This leads to the possibility of a better understanding and comprehension on the treatment option used in our clinic center.

Conclusion

Our study supports that the use of CI is an effective hearing rehabilitation method for patients with IEM. Our study suggest that patients with IEM have benefits from implantation and may even present better results in speech perception and discrimination compared to patients with CI without IEM. However, the small sample size of our study group and the variability on the control group should be taken into account when interpreting our results. Nonetheless, our study provides valuable insights into the treatment of IEM in our hospital.

Aiming to have better results after CI surgery, it could be interesting to analise the cochlear implant fitting parameters necessary to achieving optimal auditory performance outcomes.

Acknowledgements

I thank Dr. João Elói for the guidance, collaboration and all the sugestions given during this journey.

I thank Professor Dra. Sofia Paiva for making possible for this work to happen.

I am grateful to Professor Jorge Humberto Martins for the cooperation and help in providing the necessary data.

I thank all my family and friends for supporting me during this journey.

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