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Does Waardenburg Syndrome Interfere with Cochlear Implant Rehabilitation?

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Abstract

Objectives: Waardenburg syndrome is known as a collagen and pigment disease which causes different phenotypes. One of the most important phenotypes is severe bilateral congenital hearing loss. It may be possible to improve their hearing and speech abilities with cochlear implantation and rehabilitation. Nevertheless, long-term outcome after surgery in these patients is obscured. In this study, we evaluated both auditory and speech abilities in order to examine to what extent they had improved three years after surgery.

Methods: We retrospectively gathered 51 deaf Waardenburg children without any other ear anomalies along with 210 congenitally deaf and age matched children without any other anomalies as control group. All children were at least 3 years post-operation. All of them underwent speech and auditory examinations. "Categories of Auditory Performance" for was used auditory and "Speech Intelligibility Rating" was employed for speech evaluations.

Results: Male-to-female ratio was 50% in the control group and 47% in Waardenburg. The difference between hearing and speech outcomes in both genders was insignificant. Speech Intelligibility Rating in the control group was 3.49 (S.D. ± 0.07) and in Waardenburg group was 3.09 (S.D. ± 0.09), which was insignificant. Similarly, Categories of Auditory Performance in the control and Waardenburg groups were 4.4 (S.D. ± 0.69) and 4.1 (S.D. ± 2.10) respectively, which is insignificant either. There was no significant correlation between age at operation and hearing and speech results. Finally, the correlation between hearing and speech was not significant either ($p \leq 0.81$).

Conclusion: This study confirmed that there is not correlation between age at operation in cochlear implantation in Waardenburg patients and there would be no difference in hearing and speech results between them and other deaf implanted populations. The results were not sex dependent.

Keywords: Cochlear implantation; Waardenburg; Categories of auditory performance

Introduction

Waardenburg syndrome (Ws) is a rare syndrome involving auditory-pigmented disorders passed onto the children as autosomal dominant inheritance. The mutation affects the *MITF* and *Pax3* genes. Their general phenotype consists of white forelock, telecanthus, heterochromia iridis, and deafness. Four different types of Ws have been diagnosed. Type 1 contains dystopia canthorum. Type 2 excludes it. Type 3 involves musculoskeletal abnormalities, and Type 4 is characterized by aganglionic mega colon. Prevalence of this syndrome is 2-5% in deaf newborns [1]. Cochlear implantation is the accepted modality for rehabilitation of deaf population. The age at implantation, training methods, the device, and the presence of co-morbidities may affect the results.

Concerning the effect of Cochlear implantation (CI) in patients with Ws, very limited studies with different findings have been reported. This study was conducted to evaluate speech and hearing outcomes long after CI in Ws in comparison to control group.

Materials and Methods

We designed a retrospective case series study on all 21 congenitally deaf children labeled as Ws in two Iranian CI centers in 2014, where the control group consisted of 89 age and sex-matched autosomal dominant deaf children without any other co-morbidities. All of the cases were Persian speakers and examined three years after implantation. Cases with inner ear anomalies were excluded. Their age at operation varied from one to four years, and their preoperative exact intelligence quotient could not be evaluated at this age. We used Categories of auditory performance (CAP) for auditory and Speech intelligibility rating (SIR) for speech evaluations [2]. As the cases had been prelingually deaf, their CAP and SIR score were zero pre-operatively. CI devices of three companies (Med-El 25%, Nucleus 50% and AB 25%) were used. The Auditory verbal training (AVT) method was utilized for all of the cases [3]. Pre operation protocol consisted of Auditory brainstem response (ABR), temporal bone Computed tomography (CT), CAP and SIR evaluation. Six weeks post operation the device was fitted and then adjusted monthly for one year. CAP and SIR were evaluated and recorded yearly. Medical status in these patients were uneventful.

As the study was retrospective chart review without any intervention, our institutional ethical board approved the study without need for signed consent of parents.

Independent t-test was used to compare the results. However, according to Shapiro's normality test, the data were not normally distributed. Although transformation is an acceptable approach to

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	CAP		SIR		m/f
	SD	mean	SD	mean	
Ws	2.12	4.14	0.99	3.09	9/12
Control	0.69	4.48	0.7	3.49	44/45
p-sig 2 tailed	p<0.34		p<0.71		

Table 1: CAP and SIR 3 years post operation (Nonparametric independent T-test).

	Mean age at operation (month)	Correlation with CAP	Correlation with SIR
Ws	28.47	p<0.23	p<0.87
Control	21.89	p<0.35	p<0.13
Total	23.14	p<0.81	p<0.87

Table 2: Correlation between age at operation and CAP and SIR (Non-parametric T-test).

handle abnormal data, but transformation can sometimes decrease clinical comprehensiveness and applicability of results. So, we used non-parametric t-test to compare the results.

Results

A total of 1369 children were implanted in 2005–2013 in these CI centers. There were 21 Ws children among them (1.5%). Further, 89 children were chosen as the control group. There were no significant differences in sex and age between two groups. The difference between mean CAP and mean SIR in the two groups was not significant (Table 1).

The correlation between age at operation and CAP as well as SIR in both groups was evaluated. Finally, this evaluation was performed in all 110 cases. Correlations were insignificant at all. Non-parametric correlation test was used for analysis (Table 2).

Discussion

In this study, Ws prevalence was 1.5% which was less than the global prevalence (2-5%) [1]. This may be a race effect. Our cases were evaluated 3 years after intervention and rehabilitation with no correlation found between age at operation, CAP, and SIR (Table 2). This means that age at operation will not affect the results in this age group. This finding is compatible with Tinnemore [4] and Pimperton [5] studies.

As mean CAP and SIR in both groups had no significant differences, we may conclude that there would be no difference in hearing and speech results between Ws children and other deaf children, i.e. they benefit as much as they do. This result is compatible with Kontorinis et al. [6], Daneshi et al. [7], and Deka et al. [8] studies. Amirsalari, et al [9], however, reported poor speech results in Ws in comparison with control group. This difference may be the result of their early study (only 1 year after surgery) and few Ws cases. In pre-operation radiological evaluation, all the cases have had normal inner ear, which obviates influencer effect on the results [9].

Conclusion

In this study, which was performed at least 3 years after cochlear implantation, we concluded that cochlear implantation surgery would be beneficial for Waardenburg patients and there would be no difference between Waardenburg and other non-syndromic deaf children. Gender and age at operation date would not affect the results. However, speech abilities will be affected by auditory abilities.

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Conflict of Interest

This study was not granted by any company and authors have not any interest.

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