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Does Waardenburg Syndrome Interfere with Cochlear Implant Rehabilitation?

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

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 CA and mean SI in the two groups was not significant (Table 1).

The correlation between age at operation and CA as well as SI 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. ur cases were evaluated 3 years after intervention and rehabilitation with no correlation found between age at operation, CA , and SI (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 imperton [5] studies.

As mean CA and SI 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.