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Waardenburg syndrome as a challenging experience in pediatric cochlear implantation

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Abstract

Background

Waardenburg syndrome (WS) is a major cause of symptomatic sensorineural hearing loss, which accounts for 2–5% of patients with congenital hearing loss. Cochlear implantation (CI) has shown improvement in auditory perception and language skills of syndromic sensorineural hearing-impaired children and is now accepted as a gold standard treatment.

Aim

The aim of the present study was to evaluate the outcome of CI in cases with WS and to compare the results with nonsyndromic CI cases.

Participants and methods

A total of 268 children of less than 7 years of age underwent CI in the National Hearing and Speech Institute, and of these, six children, as group A, had WS, whereas the control group, as group B, consisted of 16 congenitally deaf children without any other comorbidities or inner ear anomalies. The following assessments were done: intraoperative impedance and auditory response threshold, and then at 3 and at 12 months, impedance, aided average pure tone threshold, Meaningful Auditory Integration Scale questionnaire, LittlEARS Auditory Questionnaire, and language assessment using modified Arabic preschool language scale-4.

Results

Intraoperative impedance, auditory response threshold, and neural response impedance were measured. Aided free field response at 500–4000 kHz and speech detection threshold were measured for both groups at 3 and 12 months after initiation. There was no statistically significant difference between both groups at all measured parameters. Meaningful Auditory Integration Scale questionnaire, LittlEARS Auditory Questionnaire, and language assessment using modified Arabic preschool language scale-4 revealed improvements in speech perception and production; however, comparison of the results between the WS group and the control group was insignificant.

Conclusion

CI is a good rehabilitation option for children with WS. The study indicates that children with WS benefit from CI similar to typically developing CI children.

Keywords: Cochlear implantation, speech perception, Waardenburg syndrome

Introduction

Waardenburg syndrome (WS) is a major cause of symptomatic sensorineural hearing loss (SNHL), which accounts for 2–5% of patients with congenital hearing loss. It is an autosomal dominant disease characterized by dystopia canthorum, hyperplasia of the eyebrows, heterochromia iridis, white forelock, and congenital SNHL. Clinically, WS is divided into four types according to the following clinical criteria: type 1 includes the presence of dystopia canthorum, type 2 excludes it, type 3 has additional musculoskeletal anomalies and coarser facial characteristics, and type 4 includes Hirschsprung's

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disease. The SNHL in patients with WS is owing to the result of atrophy of stria vascularis in the cochlea and caused by a lack of or a decreased number of melanocytes [1]. These cells produce endolymph which is necessary to build up a positive potential in the cochlea which is needed for the excitation of the inner hair cells, which forward an electrical impulse to the optic nerves. The atrophied stria vascularis will lead to the collapse of Reissner's membrane followed by the destruction of the organ of Corti [2].

Yoshinaga-Itano [3] has shown that children diagnosed early with hearing impairment, and who received adequate interventions by the age of 6 months, develop language capabilities at the same rate as normal-hearing children of the same age, regardless of the degree of hearing loss. Cochlear implantation (CI) has shown considerable improvement in speech and language of syndromic sensorineural hearing-impaired children and now accepted as a gold standard treatment. However, benefits of CI vary, and many determinants such as etiology, implantation age, duration of deafness, and additional comorbidities may play a role on its results. Syndromic cases are not rare among patients with CI, and either multiple handicaps or abnormalities of the bony labyrinth may have detrimental effects on satisfactory outcomes [4]. According to Miyamoto *et al.* [5], the gap between linguistic and chronological age should be minimized, and auditory information should be introduced during critical language development periods to attain the benefits of early implants. They point out that early CI placement minimizes delayed language acquisition and helps the development of appropriate language abilities according to the child's age.

This study is conducted to assess the audiological and language outcomes in children with WS and to compare it with the nonsyndromic SNHL control group.

Aim

The aim of the present study was to evaluate the outcome of CI in cases with WS and to compare the results with CI nonsyndromic cases (control group).

Participants and methods

A total of 268 children of less than 7 years of age underwent CI surgeries from December 2016 to December 2019 at The National Hearing and Speech Institute Giza, Egypt. Six children had congenital, prelingual severe to profound SNHL with WS, which accounts for 2.23% of the total number of children. According to general examination, we found five cases of type 1 WS and one case of type 2 WS.

The data obtained from 22 children in this study were divided into the following two groups: group A consisted of six congenitally severe to profound hearing loss children labeled as WS, where the control group B consisted of 16 congenitally severe to profound hearing loss children patients without any other comorbidities or inner ear anomalies.

Group A (six cases)

A total of six patients with WS were implanted. Three case were implanted with HiFocus 1 J electrode (50%) (AB, Sylmar, California, USA), one case was implanted with Sonata TI100 + titanium implant footprint with standard electrode (16.6%), one case was implanted with Flex 28 (16.6%), and one (16.6%) case was implanted with Form 24 (SA; Med-El, Innsbruck, Austria). The mean age of patients was 4 years (range: 2 years, 6 months to 7 years). Of the six cases, one (16.6%) was a male, and five (83.33%) were females. Approaches included a cochleostomy in three (50%) cases and round window approach in three (50%) cases.

Group B (16 control cases)

A total of 16 patients with normal inner ear anatomy were implanted. Eight cases received HiFocus Mid-Scala Electrode (50%) (AB). Two of them received HiFocus 1 J electrode (12.5%). Sonata TI100 + titanium implant footprint with standard electrode was used in five (31.25%) cases, and Sonata TI100 + titanium implant footprint with Flex 28 electrode was used in one (6.25%) case (SA; Med-El).

The mean age of patients was 4 years (range: 2 years, 1 month to 5 years and 9 months). Of the 16 cases, four (25%) were males and 12 (75%) were females. Approaches included a cochleostomy approach in seven (43.75%) cases and a round window approach in nine (56.25%) cases.

The approval of the local ethics CI committee of the National Hearing and Speech Institute was obtained, and a written informed consent was signed by the parents before participating in the study.

Preoperative evaluation

All young children underwent a thorough otorhinolaryngological examination and audiometric tests using behavioral audiometry, aided free-field audiometry, tympanometry, and electrophysiological tests including auditory brainstem response and otoacoustic emission.

Each child with a diagnosis of prelingual severe to profound hearing loss received high-resolution computed tomography examination and inner ear MRI. Inner ear anomalies included isolated enlarged vestibular aqueduct detected in one case of the WS cases.

Their other developmental milestones were average. There was a negative history of visual disturbances, hypothyroidism, syncopal spells, or renal problems. General examination showed average intelligence quotient with no neurological dysfunction.

Intraoperative protocol

All children received vaccinations for Pneumococcal (PREVNAR 13 vaccines), meningococci, and *Haemophilus influenza* 2 weeks before operation.

One senior surgeon (I.S.) performed CI surgeries. Cochleostomy positioned inferior and anterior to the round window was done in 10 patients (three in group A and seven in group B) and

round window approach in 12 patients (three in group A and nine in group B). The round window membrane was exposed via transmastoid facial recess approach. The scala tympani was accessed directly through the round window.

CSF pulsatile ooze or leak started in one case of group A with enlarged vestibular aqueduct syndrome. Intravenous 20% mannitol drip (1.5 g/kg body weight for >20 min) was started, and the head end of the table was raised. The leak was significantly reduced within 10 min, and the electrode array form 24 of the implant was inserted via the round window.

Intraoperative parenteral antibiotic was given. Complete insertion of the electrode array up to the mark was achieved. Temporalis muscle or fascia was used to seal tightly around the electrode array. The intraoperative telemetry showed satisfactory impedance and neural response in all the selected electrodes.

Postoperatively

A petrous bone computed tomography was carried out 2 days after the surgery that showed a correct position and no kinking of the array electrode. At 4 weeks after surgery, the implant switch-on programming session was performed by an audiologist. Regular programming was done to the cases, and all patients were referred postoperatively for auditory and language rehabilitation.

Assessment

The first assessment was done at 3 months for both groups. Then the second assessment was done after 9 months of auditory and language therapy sessions at 12 months. The following data were collected:

(1) Neural response telemetry results intraoperatively and 3 and 12 months after initiation.
(2) Aided average pure tone threshold in sound field before and after surgery and at 3 and 12 months after initiation.
(3) Postoperative speech perception and production test results: Speech perception tests for children using arabic monosyllabic and phonetically balanced words were tested at 3 months and at 12 months. Loud speakers were placed in a quite room at a fixed distance of 1 meter from children. Words were administered at normal conversational level of 65 dB sound pressure level. Speech perception was expressed as a percentage of correct phonemes perceived.
(4) Meaningful Auditory Integration Scale (MAIS) questionnaire was given to the parents at 3 and 12 months after initiation. The MAIS is designed to identify the meaning of hearing loss for a child who uses sound in daily life [6]. It gathers auditory behavioral information through 10 auditory areas to be probed. The MAIS are to be scored on the basis of parent report and clinician observation.
(5) The LittlEARS Auditory Questionnaire (LEAQ) has been used in both groups postoperatively at 3 months and at 1 year. The LEAQ is a quick and useful tool for the assessment of auditory skills in infants and toddlers [7]. The LEAQ was created to assess the auditory skills of children who receive hearing devices either hearing aids or CIs. The LEAQ relies on a parent questionnaire, which has been shown to be a reliable way of assessing auditory skills development in children. By combined information from the parent and the examiner, the examiner assigns a rating regarding the child's skill: 0, absence of skill; 1, inconsistent or emerging skill development; and 2, consistently demonstrates the skill.
(6) Language assessment was done using modified preschool language scale-4 (Arabic edition). This test measures receptive, expressive, and total language age [8]. The language assessment was done in both groups postoperatively at 3 months and at 1 year. The language improvement quotient was used to compare between the rates of progress in language and was determined by calculating the difference between the language age in a period divided by the period of time.

Statistical analysis

Statistical calculations were done using computer program IBM SPSS (Statistical Package for the Social Science; IBM Corp., Armonk, New York, USA) release 22 for Microsoft Windows. Data were statistically described in terms of mean ± SD. Comparison between the study groups was done using Mann–Whitney *U* test for independent samples. Within group comparison between 3 and 12 months data was done using Wilcoxon signed rank test for paired (matched) samples. Two-sided *P* value less than 0.05 was considered statistically significant.

Results

A total of six children in group A were diagnosed with WS and implanted at our department. They consisted of five females and one male. Of the six children, three received Medel CI, and three received AB CIs. All cases had congenital hearing loss since birth, autosomal dominant AD type of inheritance, with positive consanguinity in three cases and positive family history of similar condition in four cases. Radiological testing revealed normal inner ear by MRI and computed tomography scan except one case with enlarged vestibular aqueduct. The control cases consisted of 16 congenitally deaf children without any other comorbidities or inner ear anomalies who had CI. The group B consisted of 12 females and four males.

The mean of the aided free field response at 500–4000 kHz and speech detection threshold was measured at 3 months (Table 1), whereas the aided free field response at 500–4000 kHz, the aided speech recognition therapy (SRT), and discrimination were detected at 12 months (Table 2). By statistical analysis, a high statistically significant difference was found in both the WS cases and the control cases between 3 and 12 months after initiation when testing, which indicates that the response with time usage of CI showed improvement in both control and study groups. Meanwhile, there was no statistically significant difference between both groups at all measured parameters except at 1000 Hz (Fig. 1).

Intraoperative impedance, auditory response threshold, neural response impedance and then impedance at 3 and 12 months after initiation were measured for both groups. There was no statistically significant difference between both groups at all measured parameters (Table 3 and Fig. 2).

The mean and SD of the MAIS questionnaire scores were measured at 3 and 12 months (Table 4), which shows no statistical difference between both groups (WS cases and control cases).

The scores of the LEAQ were compared between the two groups. Both groups showed clinical improvements in speech perception and production. The mean performance of LittlEARS questionnaire in the Waardenburg group was 10.3 (SD 2.06) at 3 months and 31.6 (SD 2.25) at 12 months. However, in the controls group was 10.5 (SD 2.16) at 3 months and 31.81 (SD 2.07) at 12 months which is considered to be insignificant, as shown in Table 5 and Fig. 3. Meanwhile, there is a significant difference between the results of the LEAQ at 3 and at 12 months in each group ($P < 0.05$) (Table 6). This indicates the improvement of the auditory skills during the period of rehabilitation.

The results of the language assessment using the preschool language scale-4 were compared between the two groups. The mean of the receptive language quotient was 1.25 (SD 0.09) for group A and 1.21 (SD 0.18) for group B ($P = 0.54$). The mean of the expressive language quotient was 0.61 (SD 0.35) for group A and 0.66 (SD 0.41) for group B ($P = 0.9$). The mean of the total language quotient was 0.88 (SD 0.22) for group A and 0.92 (SD 0.378) for group B ($P = 0.85$). A nonsignificant difference was found between the both groups, as shown in Table 7 and Fig. 4.

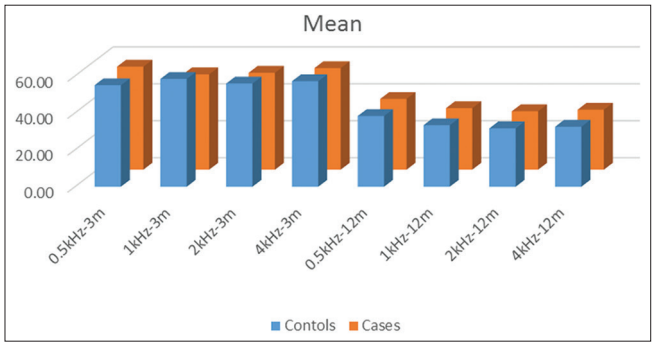

Figure 1: Comparison between control and cases for measured aided free field 3 and 12 months after fitting (0.5, 1, 2, and 4 kHz).

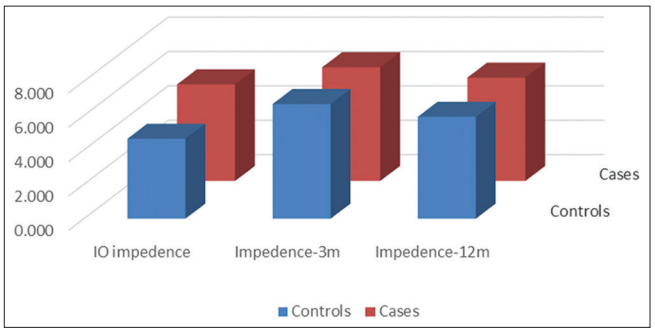

Figure 2: Comparison of mean between control and cases for measured intraoperative impedance (Imp-IO), 1 and 3 months postoperative impedances (Imp-1m and Imp-3m, respectively).

Table 1: Mean and SD of aided free field (0.5, 1, 2, and 4 kHz) and aided speech detection threshold 3 months after fitting in both groups

Groups	0.5 kHz-3 months	1 kHz-3 months	2 kHz-3 months	4 kHz-3 months	SDT 3 months
Controls					
Mean	55.00	58.44	55.94	57.19	55.63
SD	5.164	7.004	5.234	5.154	6.021
Cases					
Mean	55.83	51.67	52.50	55.00	52.50
SD	3.764	4.082	2.739	4.472	2.739
P	0.698	0.028*	0.159	0.398	0.288

*There is statistically significant difference between both groups at 1 HZ) ($p<0.05$), SDT, speech detection threshold. There is no statistically significant difference between both groups at all measured parameters except at 1 kHz.

Table 2: Mean and SD of aided free field (0.5, 1, 2, and 4 kHz), aided speech recognition threshold, and discrimination 12 months after fitting in both groups

Groups	0.5 kHz-12 months	1 kHz-12 months	2 kHz-12 months	4 kHz-12 months	SRT-12 months	Discrimination (%)
Controls						
Mean	38.33	33.33	31.67	32.50	33.33	34.000
SD	5.323	6.115	5.836	4.082	3.096	13.3766
Cases						
Mean	38.33	33.33	31.67	32.50	33.33	34.000
SD	5.164	5.164	7.528	4.183	4.082	23.6981
P	0.969	0.346	0.456	0.964	0.386	0.708

There was no statistically significant difference between both groups at all measured parameters.

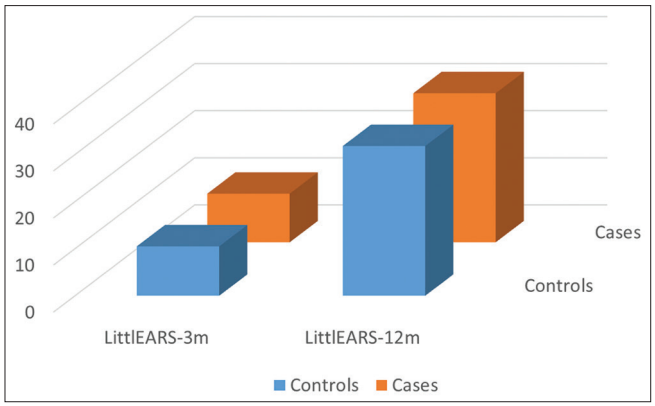

Figure 3: Comparison of mean between controls group and cases group for measured. LittlEARS Auditory Questionnaire (LEAQ) (3 and 12 months postoperatively).

Table 3: Intraoperative impedance and IO auditory response threshold, 3 and 12 months postoperative impedances in both groups

Groups	Imp-IO	Imp-3 months	Imp-12 months	ART IO
Controls				
Mean	4.654	6.674	5.930	492.14
SD	3.0475	0.8844	1.4266	398.529
Cases				
Mean	5.622	6.617	6.016	436.88
SD	2.6971	2.0116	1.4407	290.604
P	0.269	0.941	0.883	0.417

ART, auditory response threshold; Imp, impedance; IO, intraoperative impedance. No statistically significant difference between both groups at all measured parameters.

Table 4: The mean and SD of Meaningful Auditory Integration Scale Questionnaire (3, 12 months postfitting) in both groups

Group	MAIS 3 months	MAIS 12 months	*P*
Control			
Mean	8.375	17.5	0.881002
SD	2.526526	6.870226	
Cases			
Mean	8.5	20.33333	0.551501
SD	3.271085	9.330952	

MAIS, Meaningful Auditory Integration Scale. There is no statistically significant difference between both groups regarding the scores of the MAIS questionnaire.

Discussion

All Waardenburg cases were diagnosed with severe to profound congenital hearing loss, which is in agreement with Barzotto and Folador [9], who showed that the most common form of hearing loss in WS is profound sensorineural. During the pre-implant evaluation, patients were found to have no benefit from conventional hearing aids. Thus, CI was indicated.

All patients had complete insertion of electrodes and showed an intraoperative neural response, which means that the auditory nerve responded to the first electrical stimulation of the CI. Guedes *et al.* [10] showed that adult patients who showed intraoperative telemetry responses had better results in speech perception tests, but this relationship was not statistically significant among children.

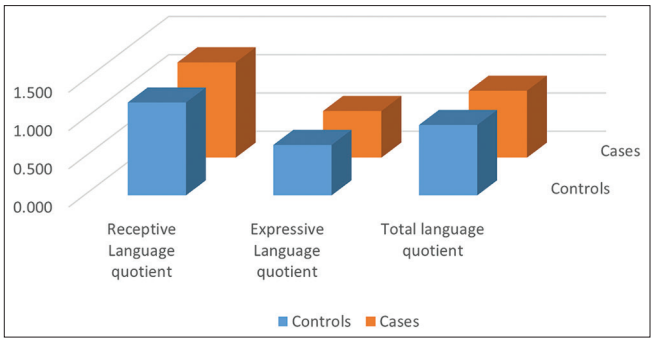

Figure 4: Comparison of mean between controls group and cases group for measured receptive, expressive and total language quotient.

Table 5: Mean and SD results of LittlEARS Auditory Questionnaire (3 and 12 months postoperatively) in both groups using Mann-Whitney *U* test

Groups	LittlEARS-3m	LittlEARS-12m	*P*
Controls			
Mean	10.5	31.8125	0.93934
SD	2.160247	2.072639	
Cases			
Mean	10.33333	31.66667	0.754513
SD	2.065591	2.250926	

LittlEARS Auditory Questionnaire. No statistically significant difference between both groups at all measured parameters.

Table 6: Comparison between the results of the LittlEARS Auditory Questionnaire at 3 months and results of the LittlEARS Auditory Questionnaire at 12 months in both groups

Groups	LittlEARS-3 months	LittlEARS-12 months	*P*
Controls			
Mean	10.50	31.81	0.00*
SD	2.160	2.073	
Cases			
Mean	10.33	31.67	0.02*
SD	2.066	2.251	

Using Mann-Whitney *U* test. *There is statistically significant difference between results at 3 months and at 12 months in both groups($p<0.05$).

The MAIS questionnaire results confirmed what had been demonstrated in the test battery used for assessing speech perception. It has been used in many studies to assess CI use in daily activities. This study showed significant clinical improvements in most cases with increasing time of CI use, reflecting improvements in listening skills, not only for detection but also for the recognition of some sounds. Meanwhile, the comparisons of the results of the MAIS scores between both the WS cases and the control cases were

Table 7: Comparison between the controls group and the cases group as regards receptive, expressive, and total language quotient

Groups	Receptive language quotient	Expressive language quotient	Total language quotient
Control			
Mean	1.214	0.66	0.9200
SD	0.1794	0.405	0.37870
Cases			
Mean	1.245	0.61	0.8750
SD	0.0931	0.347	0.21742
P	0.544	0.909	0.852

No statistically significant difference between both groups at all measured parameters.

insignificant. Kubo and Sasaki [11] showed that after 6–12 months of use of a CI, children were able to distinguish and recognize sounds. One case of the present study showed a poor clinical improvement as detected by MAIS because this case had the CI operation at an older age (6 years old), and he did not use the CI effectively, as there were maintenance problems with poor family support as well as infrequent use of the implant during the rehabilitation process. Miyamoto *et al.* [5] used the MAIS successfully to check the progression of auditory behavior in children aged below 3 years who had implants. These findings indicate that CIs provide access to speech sounds, but that the development of auditory and language skills is dependent on systematic rehabilitation and family involvement. Another study done by Barzotto and Folador [9] 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 preoperation radiological evaluation, all the cases have had normal inner ear.

Many studies found improved auditory and language performance after CI in cases with WS. The LittlEars questionnaire results in this study showed significant clinical improvements in both groups, reflecting improvements in the listening skills. All of the children with WS demonstrated significant improvement in auditory perception from 3 to 12 months. However, there was no significant difference in the scores between the WS group and the control group, which indicates that WS cases benefit from CI. The results are similar to the studies done by Daneshi *et al.* [12], Amirsalari *et al.* [13], and Kontorinis *et al.* [14].

Amirsalari *et al.* [13] compared six young deaf children with WS (mean age 26 ± 15.8 months) and 75 deaf children without WS (mean age 54.5 ± 14.8 months) 1 year following implantation. All of the children with WS demonstrated significant improvement in auditory perception and speech intelligibility from preimplantation to postimplantation. The study done by Deka *et al.* [15] showed that WS group achieved postimplant scores comparable with children without WS using category auditory performance and MAIS. All children with WS significantly benefitted from CI.

Conclusion

In the group of patients with WS who received a CI, hearing thresholds that allow access to speech sounds were achieved. They showed good development of auditory perception and language skills. CI is a good rehabilitation option for children with WS. The study indicates that CI children with WS benefit from CI similarly to nonsyndromic CI children. However, further research studies on a larger scale of Waardenburg cases and for longer duration of follow-up of cases are recommended.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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