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OTOLOGY

Cochlear implantation in patients with Cogan syndrome: long-term results

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Abstract The objective of this study was to evaluate the long-term outcomes of patients with Cogan syndrome (CS) who have undergone cochlear implantation. Subjects consisted of 12 cochlear implant users with a typical form of CS. Measures included word and sentence recognition scores. The speech recognition performance was rated before cochlear implantation and at 1 and 5 years after implantation. The speech materials were presented in quiet only condition. The mean 12-month post-operative word and sentence recognition scores were 91.4 and 93.1 %, respectively. Five years after implantation, the group means for word and sentence recognition tests were 94 and 96.3 %, respectively. No patients in this series experienced flap complication or other local or systemic complications. This long-term study on 12 subjects with CS over 5 years of cochlear implant use reveals that cochlear implantation is safe in the long term and provides excellent and stable hearing results.

Keywords Cochlear implant · Cogan syndrome · Deafness · Ossified cochlea · Results

Introduction

Cogan Syndrome (CS) is a rare autoimmune vasculitis characterized by recurrent ocular inflammation associated with vestibulo auditory dysfunction [1, 2]. Cogan syndrome has been divided into classic and atypical forms on the basis of the ocular findings. The classic form, first described by Dr. David G. Cogan in 1945 [3], is characterized by the presence of bilateral non-syphilitic interstitial keratitis and vestibulo auditory dysfunction which is usually bilateral and mimic the more common Ménière disease with tinnitus, vertigo and sensorineural hearing loss (SNHL) [4, 5]. The audiovestibular manifestations may develop coincidentally or within 1–2 years after the onset of ocular signs [6, 7]. The atypical form may involve every ocular structure, leading to conjunctivitis, scleritis, uveitis, retinitis, and optic neuritis. In atypical CS, audiovestibular symptoms are different from Meniere-like episodes and appear more than 2 years after ocular manifestations [2]. Systemic disease arises in about 70 % of cases and includes fever, headache, myalgia, arthralgia, lymphadenopathy, hepatosplenomegaly, aortitis, coronary arteritis, pulmonary nodules and pleuritis [4, 5]. Systemic vasculitis develops in up to 10 % of patients and may involve any sized vessels [4, 8].

The ocular inflammation generally responds to topical atropine and ocular corticosteroids [9, 10]. Audiovestibular dysfunction requires systemic corticosteroids and, in selected cases, immunosuppressive drugs such as methotrexate and cyclophosphamide [1, 10, 11]. Despite this aggressive treatment, approximately 50–85 % of patients with CS experience severe to profound irreversible SNHL [4, 7, 12, 13]. In deafened patients in whom no benefits are obtained from conventional hearing aids, cochlear implantation is an highly effective technique in restoring hearing abilities.

Due to the rarity of the disease, few reports have specifically studied the use of cochlear implants in patients suffering from CS [4, 5, 8, 10, 14–18]. These studies have already demonstrated the efficacy of cochlear implantation

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in patients with CS. However, the majority of the articles report small series or case reports with a limited follow-up.

This study aimed at evaluating the long-term outcome performance in a series of 12 patients with CS who received a cochlear implant. To the best of our knowledge, this is the largest series of cochlear implantation in patients with Cogan syndrome.

Materials and methods

After institutional review board approval, a retrospective analysis of all patients entering the cochlear implant program at the Cochlear Implant Center of the University-Hospital of Parma was conducted to identify the patients who were diagnosed pre-operatively with CS. Twelve patients with a confirmed diagnosis of CS who received cochlear implants were identified and participated in the study. This patient population accounted for 4.4 % of all implanted patients in our center. This high percentage may be explained by the presence in our Hospital of a reference Center for the treatment of autoimmune diseases. All the 12 patients were affected by the classic form of CS. The diagnosis of the classic form of CS was made on the basis of the observation of recurrent interstitial keratitis, acute-onset SNHL, Ménière's-like audiovestibular dysfunction, and after exclusion of all other causes of interstitial keratitis (i.e., syphilis, sarcoidosis, leprosy, Lyme disease, viruses, hypersensitivity to drugs). Fluorescent treponemal antibody absorption test and tuberculosis skin test were performed in all patients. Laboratory and immunological tests completed the diagnostic work-up. One month before surgery, all patients were pre-operatively investigated with high-resolution computed tomography (HRCT) of the petrous bones and magnetic resonance imaging (MRI) scans of the brain focusing on the inner ear.

Data collected from the patients' medical records included demographic information, operative procedures, radiological features, intra- and post-operative complications, and performance outcomes.

Speech perception was evaluated using word and everyday sentence speech recognition tests. Italian versions of the North-western University Phonetically Balanced Word Lists and the Central Institute for the Deaf Everyday Sentence Lists were used to measure speech perception benefits. The speech materials were presented in hearing-only conditions using a monitored live voice through the sound field at a level of 70 dB sound pressure level. For the present study, results were collected for three specific time intervals: before surgery, at 12 months post-implantation, and after a minimum of 5 years of follow-up. Follow-up was defined as the period of time from surgery to the most recent office visit. The follow-up of the series ranged from 64 to 158 months (mean, 94.7 ± 29.3 months).

Results

A summary of the characteristics of the 12 subjects included in this study is shown in Table 1. Five patients reported previously are included in this study with longer follow-up [18]. There were 8 (66.6 %) women and 4 (33.4 %) men, providing a female–male ratio of 2:1. Pre-operatively, all patients exhibited either complete deafness or a bilateral profound SNHL (unaided pure tone average thresholds greater than 90 dB). All patients had been fitted with hearing aids immediately after diagnosis of hearing loss. In all cases cochlear implantation was considered when any progress in speech perception abilities from amplification and rehabilitation was noted. Age at implantation ranged from 16 to 52 years (mean 34.1 ± 10 years). The length of the time from initial deafness diagnosis and cochlear implantation ranged from 6 to 48 months (mean 19 ± 13 months). All patients but one received pre-operative steroid and immunosuppressive therapy.

Four patients received the Nucleus 24M device (Cochlear Ltd., Melbourne, Australia); one, the Nucleus 22M device; and two, the Nucleus Contour model; and five patients were implanted with the MXM Digisonic device (MXM Corporation, Antibes, France). On pre-operative HRCT scan, 10 subjects (83.3 %) had normal cochleas and 2 (16.7 %) had some degree of ossification of the inferior segment of the basal turn of the cochlea; MRI confirmed the two cases of ossification detected by HRCT and revealed another case of ossification. At surgery, the three cases with abnormal radiologic findings were proven to have cochlear ossification; cochlear ossification was an intraoperative surprise in other three cases. In total, six patients (50 %) were found to have some degree of ossification. In four cases, there was bone obliteration confined to the round window niche and the first millimeters (2–3 mm) of the inferior segment of the basal turn; however, these were successfully managed by tunneling through the ossified portion of the basal turn. In the other two cases, the inferior segment of the basal turn was completely filled by fibrous and bony tissue, and the electrode was inserted into the scala vestibuli. We did not have cases of ossification of the ascending segment of the scala tympani or of the scala vestibuli. In the remaining six subjects, neither ossification nor fibrosis was found, and a standard scala tympani insertion was performed. Full insertion of the electrode array was accomplished in all the cases. Ten of the 12 patients had an uneventful post-operative recovery. Two patients developed a recurrence of

Table 1 Demographic data of the 12 Cogan Syndrome patients who received cochlear implants

Patient	Sex	Age at surgery (years)	Hearing loss	Duration of deafness (months)	HRCT	MRI	Intraoperative findings	Surgical technique	CI model	Follow-up (months)
1	F	27	Profound	6	Abnormal	Abnormal	ST ossification	SV	Contour	158
2	F	27	Total	12	Normal	Abnormal	ST ossification	SV	N24	96
3	F	41	Total	36	Normal	Normal	Patent cochlea	ST	N22	134
4	F	37	Profound	12	Normal	Normal	Patent Cochlea	ST	N24	124
5	M	52	Profound	10	Normal	Normal	ST ossification	ST	N24	72
6	F	44	Profound	28	Normal	Normal	Patent cochlea	ST	Contour	78
7	M	33	Total	24	Normal	Normal	ST ossification	ST	MXM	92
8	F	40	Profound	48	Normal	Normal	Patent cochlea	ST	MXM	94
9	M	23	Total	8	Normal	Normal	ST ossification	ST	MXM	64
10	M	16	Profound	9	Normal	Normal	Patent cochlea	ST	MXM	66
11	F	30	Total	12	Abnormal	Abnormal	ST ossification	ST	MXM	74
12	F	40	Profound	24	Normal	Normal	Patent cochlea	ST	N24	85

F female, *M* male, *HRCT* high-resolution computed tomography, *MRI* magnetic resonance imaging, *ST* scala tympani, *SV* scala vestibuli

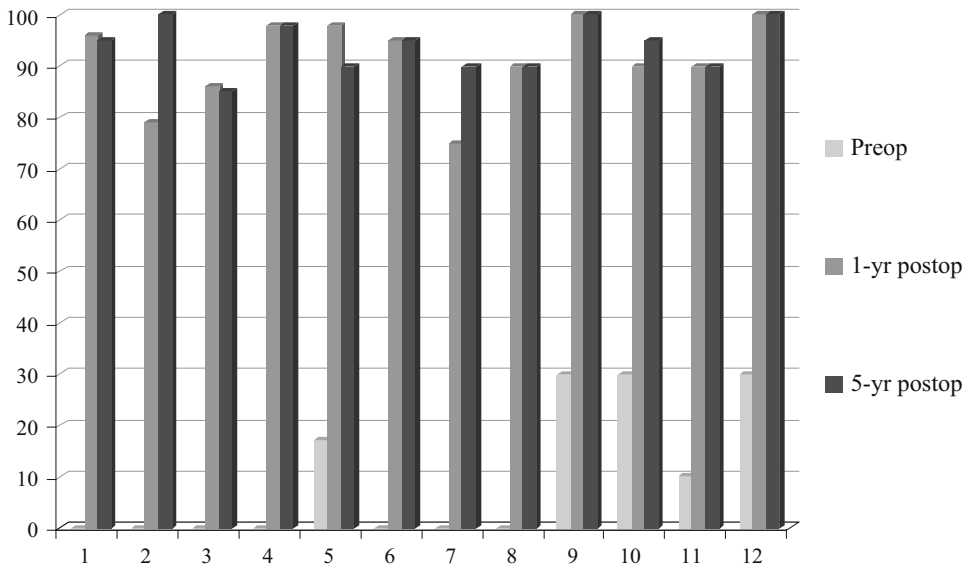

Fig. 1 Pre- and post-implantation individual word recognition scores

keratitis the day after surgery that was successfully treated with local atropine and corticosteroid therapy. No patients in this series experienced flap complications or other local or systemic complications. Before implantation, the mean correct score for the 12 patients was 9.7 % (range 0–30 %) and 10.9 % +16 (range 0–48 %), on the word and sentence tests. At 12 months after implantation, the group means for word and sentence recognition tests were 91.4 % (range 75–100 %) and 93.1 % (range 76–100 %). After 5 years of implant use, the mean percentage of correct word and sentence recognition scores was 94 % (range 85–100 %) and 96.3 % (range 90–100 %). Figures 1 and 2 show hearing test scores for the 12 patients before and at 1 and 5 years after implantation.

Discussion

Hearing loss in patients with CS is generally bilateral and often progresses to irreversible bilateral profound SNHL despite high-dose corticosteroids and immunosuppressive treatment [5, 8, 10, 14–18]. Temporal bone histological analysis in patients with CS has shown (1) atrophy of the organ of Corti [19, 20], (2) fibrous tissue and bone

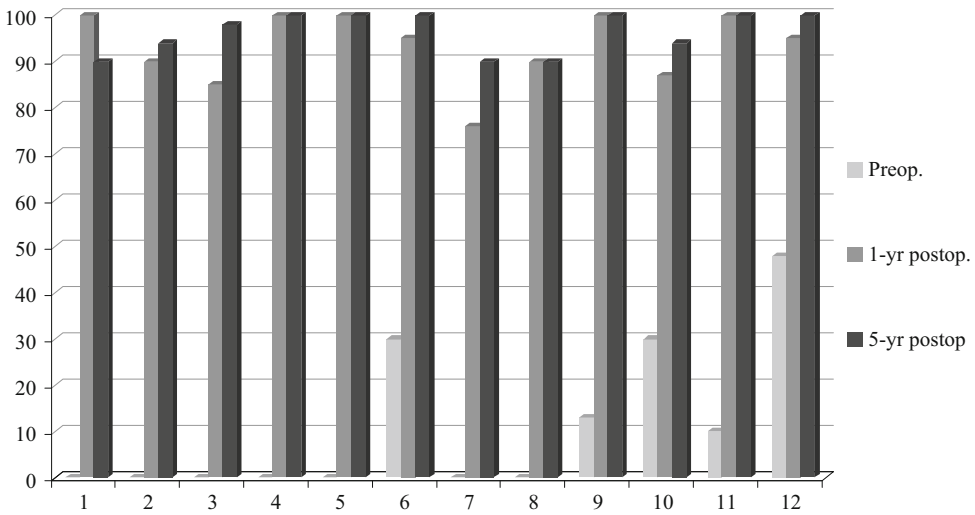

Fig. 2 Pre- and post-implantation individual sentence recognition scores

proliferation that involves the cochlea and vestibular labyrinths [19, 21], (3) demyelinization of the eighth cranial nerve [20], (4) degeneration of the sensory receptors and supporting structures of the cochlea and vestibule [19] and, (5) endolymphatic hydrops [19–21]. These histopathologic findings suggest that the pathogenesis of SNHL may be related to an immunologic mechanism secondary to an inflammatory attack on the membranous labyrinth [7].

Cochlear implantation in CS may be technically difficult due to the tendency for inflammatory disease to induce endosteal reaction leading to obliteration of the intracochlear ducts by soft-tissue and bone [21–23]. Since management of ossified cochlea still represents a surgical difficulty, early cochlear implantation should be recommended. In our population of 12 patients, 6 presented an intracochlear obliteration, demanding modifications of the surgical technique. In four cases, bone obliteration, which was confined to the first millimeters of the inferior segment of the basal turn, was successfully managed by tunneling through the ossified portion of the basal turn. In the other two patients, the inferior segment of the basal turn was completely filled by bone tissue, and scala vestibuli insertion was performed using the technique described by Steenerson et al. [24, 25]. Pre-operative imaging was normal in 22 of the 24 patients reported in the English literature and abnormal in 2 patients. Intraoperatively, other four patients were found to have some degree of ossification for a total of six patients with ossification. Five of these patients had scala tympani electrode insertion by drilling out the inferior segment of the basal turn and one patient had scala vestibule insertion. All patients but one had full electrode insertion.

It has been reported that the basic illness renders patients affected by CS potentially more susceptible to complications. Among the potential post-operative complications, those related to the flap are reported as the more common [18]. Flap ischemia in a young adult with CS who had undergone implantation for the second time on the same side was reported by Kontorinis et al. [5]. Wang et al. [26] have reported a wound healing disorder and recurrent episodes of skin infections in one of their four patients during the post-operative period. The patient reported by Low et al. [15] experienced a pressure sore of the occiput from the post-operative head bandage. Skin atrophy from long-term corticosteroid and immunosuppressant therapy and ischemia caused by vasculitis may be considered to be adverse factors contributing to the described flap complications (Table 2).

Diligent planning of a flap with a wide blood supply is fundamental to avoid ischemia-related complications. In our 12 patients, we used an inverted "J" shaped flap, which provided a wide blood supply from the posterior auricular and occipital arteries. No patient in our series has experienced flap problems. Other potential complications are related to the worsening of CS symptoms due to the stress caused by the surgical procedure [17]. Two patients of our series developed a recurrence of keratitis the day after surgery. Also, Kontorinis et al. [5] reported of a patient who suffered from an episode of interstitial keratitis post-operatively which was successfully treated with atropine and corticosteroids. It is probable that in these patients, an acute phase of the disease with recurrence of the ocular inflammation occurred as a result of the stress consequent to the surgical procedure. To date, very few reports have specifically focused on the post-implantation outcomes in patients who become deaf due to CS [4, 5, 8, 10, 14–17]. However, most authors agree to the fact that prognosis of cochlear implantation with regards to hearing results is excellent in this population [5, 10, 16, 17]. This can be explained by the fact that these patients became deaf post-

Table 2 Review of the Literature

Author	Age/sex	Pre-operative imaging	Device	Surgical technique	Ossification	Open-set word recognition (%)	Open-set sentence recognition (%)	Complications	Follow-up (months)
Cinamon et al. [14]									
Case 1	53/F	Normal	Nucleus	ST	No	40	86	–	12
Case 2	36/M	Normal	Nucleus	ST	No	50	96	–	36
Case 3	38/M	Normal	Nucleus	ST	No	–	–	–	24
Minet et al. [10]									
Case 1	56/M	Normal	Nucleus	ST	No	42.5	–	–	–
Case 2	47/F	Normal	Nucleus	ST	No	92.5	95.5	–	–
Case 3	40/F	Normal	Nucleus	ST	No	92.5	97	–	–
Case 4	32/F	Normal	Nucleus	ST	No	100	100	–	–
Low et al. [15]									
Case 1	39/M	Normal	Nucleus	ST	Yes	31*	72*	Pressure sore of the occiput from dressing	3
Vishwakarma et al. [8]									
Case 1	58/F	Normal	MED-EL	ST	No	90	–	–	–
Wang et al. [16]									
Case 1	23/F	Normal	–	ST	No	–	–	–	–
Case 2	30/F	Normal	–	ST	No	–	–	–	–
Case 3	65/F	Normal	–	ST	No	–	–	–	–
Case 4	46/M	Normal	–	ST	No	–	–	–	–
Case 5	58/M	Normal	–	ST	No	–	–	–	–
Case 6	34/F	Normal	–	ST	No	–	–	–	–
Case 7	40/F	Normal	–	ST	No	–	–	–	–
Kawamura et al. [17]									
Case 1	55/F	Abnormal	Nucleus	ST	Yes	78	79	–	12
Kontorinis et al. [5]									
Case 1	21/F	Normal	–	ST	No	90	100	–	192
Case 2	35/F	Normal	–	ST	No	95	100	–	144
Case 3	9/F	Normal	–	ST	Yes	65	87.7	–	96
Case 4	30F	Normal	–	ST	No	70	99.1	One episode of interstitial keratitis post-operatively. Skin atrophy and infection above the implant	12
Bovo et al. [4]									
Case 1	48/M	Normal	Nucleus	ST	Yes	90	–	Tactile and painful stimulation in the hemiface which required a disconnection of all the basal electrodes. Speech perception worsened to closed-set word identification	132

Table 2 continued

Author	Age/sex	Pre-operative imaging	Device	Surgical technique	Ossification	Open-set word recognition (%)	Open-set sentence recognition (%)	Complications	Follow-up (months)
Case 2	31/F	Abnormal	MED-EL	ST (bilateral)	Yes	90	–	–	28
Case 3	18/F	Normal	Nucleus	ST (RS) SV (LS)	Yes (LS)	–	–	The patient only reached closed-set word identification	42

F female, *M* male, *ST* scala tympani insertion, *SV* scala vestibule insertion, *RS* right side, *LS* left side

* Closed-set auditory alone testing

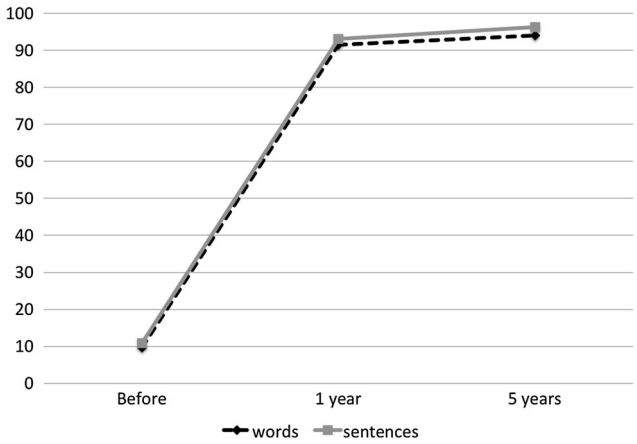

Fig. 3 Pre- and post-implantation (at 1 and 5 years) mean word and sentence recognition scores for the 12 patients

lingually and are generally implanted after a short period from the onset of profound SNHL. Minet et al. [10] described the hearing results obtained in a series of four patients with CS. At the post-operative evaluation, the mean open-set word and sentence recognition scores were about 82 and 97.5 %, respectively. In data on seven implanted patients with CS provided by Wang et al. [16], the mean 2-year post-operative open-set sentence recognition score was 98.7 %. However, Bovo et al. [4] reported a different experience with two of their three CS implanted patients reaching non-satisfactory outcome. In one patient, cochlear ossification advanced after implantation with four electrodes becoming progressively non-functioning and six electrodes being switched off due to painful sensations. The remaining patient reached only identification performances: the authors ascribed the limited benefit to an intervening nerve dyssynchrony or to cognitive/psychological disorders which interfere with auditory processing [4]. In addition, a post-implant deterioration of speech perception was observed in one patient with CS by Quaranta et al. [27]. The authors speculated that the deterioration of hearing after cochlear implantation could be a consequence of apposition or progression of new bone formation, which in turn increases the distance of the electrodes from neural structures.

Although several studies have assessed CS patients' progress after implantation, there is only one study concerning the long-term performance of cochlear implant recipients with CS [5]. Kontorinis et al. [5] reported the long-term outcomes of four patients with CS (average follow-up of 9.25 years) providing evidence of hearing outcome's persistence. These four patients achieved mean scores of 78.7 and 92.4 % on word and sentence recognition tests, respectively. At their last evaluation, the mean word score was 80 %, whereas the mean sentence score was 96.6 %. These results are in agreement with the results

of the present study. In the current study, the mean 12-month post-operative open-set word recognition score for our 12 patients was 91.4 % and the mean sentence recognition score was 93.1 %. At the 5-year post-operative evaluation, the mean word and sentence recognition scores were 94 and 96.3 %, respectively. These excellent results can be explained by the fact that all patients were young adults deafened postlingually and progressively who had been using hearing aids before implantation; in addition, the length of the profound hearing loss was relatively brief in all cases (6–48 months). It is interesting to mention that hearing performances remained stable over time (Fig. 3).

Conclusion

The data of this study further confirm the results obtained in previous studies and demonstrate that patients with CS receive significant open-set speech recognition benefits from a cochlear implant that remain stable in the long term.

Conflict of interest None.

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