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USE OF A COCHLEAR IMPLANT IN A PATIENT WITH COGAN'S SYNDROME AND PROFOUND SENSORINEURAL HEARING LOSS: A CASE STUDY

Contributions:
A Study design/planning
B Data collection/entry
C Data analysis/statistics
D Data interpretation
E Preparation of manuscript
F Literature analysis/search
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Abstract

Background: Cogan's syndrome is a very rare autoimmune disease characterized by the coexistence of inflammatory lesions in the eyeball and inner ear dysfunction. The symptoms of Cogan's syndrome within the inner ear appear suddenly and resemble Meniere's disease symptoms: severe vertigo, nausea, vomiting, and usually bilateral, fluctuating sensorineural hearing loss with concomitant tinnitus.

Case report: A 59-year-old woman was admitted to the Otolaryngology Head and Neck Surgery Clinic to undergo cochlear implantation. At 57 years of age, the patient had been diagnosed with scleritis. About a year later, vertigo and nausea arose, followed by rapidly progressing hearing loss – at first in the left ear and subsequently in the right, accompanied by tinnitus. At the time of implantation, the patient's speech discrimination score was 0% for both ears. Taking into consideration the audiometric tests results and imaging scans, promontorial cochleostomy was carried out during the cochlear implantation procedure. In the postoperative period, no complications were observed. After 24 months of using the speech processor, the subjective assessment of speech intelligibility given by the patient on a 0–10 scale was 8.5 in quiet and 5.0 in a noisy environment. The patient could identify 95% of monosyllabic words in silence and 35% in noise (in conditions of SNR +10 dB, speech level at 70 dB HL, and noise level at 60 dB HL). The benefits obtained were confirmed by results of an Abbreviated Profile of Hearing Aid Benefit (APHAB) questionnaire. The patient is presently eligible for cochlear implantation in the other ear.

Conclusions: Cochlear implantation was effective in improving hearing in a patient with Cogan's syndrome.

Key words: cochlear implants • Cogan's syndrome • hearing loss correction • hearing

USO DEL IMPLANTE COCLEAR EN UNA PACIENTE CON SÍNDROME DE COGAN Y PÉRDIDA AUDITIVA RECEPTIVA PROFUNDA: ESTUDIO DEL CASO

Resumen

Introducción: El síndrome de Cogan es una enfermedad auto-inmune muy rara, que se caracteriza por la coexistencia de procesos inflamatorios en el globo acular y trastornos del oído interno. Los síntomas del síndrome de Cogan aparecen en el oído interno de forma repentina y son similares a los síntomas del síndrome de Ménière: vértigo severo, nauseas, vomito, así como pérdida auditiva receptiva progresiva, normalmente bilateral, acompañada de tinnitus.

Estudio del caso: Mujer de 59 años fue ingresada en el Departamento de Otorrinolaringología y Cirugía de Cabeza y Cuello para implantación coclear. A los 57 años, la paciente fue diagnosticada con escleritis. Un año más tarde aparecieron el vértigo y nauseas, y a continuación pérdida auditiva de progreso rápido: primero en el oído izquierdo, después en el derecho, acompañada de tinnitus. A la hora de la implantación, el resultado de la prueba de reconocimiento del habla por la paciente fue 0% en ambos oídos. Tomando en cuenta los resultados de las pruebas audiométricas y de las pruebas de imagen, durante la implantación coclear se realizó una cocleostomía. En el periodo post-operativo no se observaron complicaciones. Pasados los 24 meses de usar el procesador del habla, la evaluación subjetiva de la compresión del habla determinada por la paciente en una escala 0-10 fue de 8,5 en silencio y de 5,0 en ruido. La paciente podía identificar un 95% de palabras monosilábicas en silencio y 35% en presencia de ruido (en condiciones de la relación señal/ruido +10 dB, nivel del habla 70 dB HL y nivel del ruido 60 dB HL). Los beneficios obtenidos se confirmaron mediante los resultados del cuestionario que evaluaba la calidad de vida con dispositivos que refuerzan la audición y los beneficios provenientes de su uso (APHAB). Actualmente la paciente es apta para la implantación coclear en el otro oído.

Conclusiones: La implantación coclear ha sido eficaz y ha causado una mejora de la audición en la paciente con síndrome de Cogan.

Palabras clave: implantes cocleares • síndrome de Cogan • corrección de pérdidas auditivas • audición

ИСПОЛЬЗОВАНИЕ КОХЛЕАРНОГО ИМПЛАНТАТА У ПАЦИЕНТА С СИНДРОМОМ КОГАНА И ГЛУБОКОЙ ПЕРЦЕПТИВНОЙ ТУГОУХОСТЬЮ: РАЗБОР СЛУЧАЯ

Изложение

Введение: Синдром Когана – это очень редкое аутоимунное заболевание, характеризующееся воспалительными изменениями в глазном яблоке и нарушениями во внутреннем ухе. Симптомы синдрома Когана во внутреннем ухе появляются резко и напоминают симптомы болезни Меньера: тяжёлые головокружения, тошнота, рвота и обычно двусторонняя прогрессирующая нейросенсорная тугоухость с сопутствующим шумом в ушах.

Описание случая: 59-летняя женщина была принята в Клинику отоларингологии и хирургии головы и шеи с целью вживления кохлеарного имплантата. В возрасте 57 лет у пациентки был обнаружен склерит. Чуть более года спустя появились головокружения и тошнота, а потом быстро прогрессирующая тугоухость – сначала в левом, а потом в правом ухе, которой сопутствовал шум в ушах. В моменте имплантации результат теста различения речи пациенткой составил 0% в обоих ушах. Принимая во внимание результаты аудиометрических исследований и визуальной диагностики, во время кохлеарной имплантации была проведена кохлеостомия. В постоперационный период не наблюдалось осложнений. Через 24 месяца использования процессора речи субъективная оценка понятности речи оценивалась пациенткой по шкале 0–10 баллов на уровне 8,5 в тихой среде и 5,0 в шумной среде. Пациентка смогла идентифицировать 95% односложных слов в тишине и 35% в шуме (в условиях отношения сигнала к шуму + 10дБ, при уровне речи 70 дБ HL и уровне шума 60 дБ HL). Полученная польза была подтверждена результатами опросника, оценивающего качество жизни с устройствами, улучшающими слух, и пользу, исходящую из их использования (APHAB). В настоящее время пациентка квалифицируется на кохлеарную имплантацию во втором ухе.

Выводы: Кохлеарная имплантация эффективным образом повлияла на улучшение слуха у пациентки с синдромом Когана.

Ключевые слова: кохлеарные имплантаты • синдром Когана • корректирование тугоухости • слух

ZASTOSOWANIE IMPLANTU ŚLIMAKOWEGO U PACJENTA Z ZESPOŁEM COGANA I GŁĘBOKIM ODBIORCZYM UBYTKIEM SŁUCHU: STUDIUM PRZYPADKU

Streszczenie

Wprowadzenie: Zespół Cogana jest bardzo rzadką chorobą autoimmunologiczną charakteryzującą się współistnieniem zmian zapalnych w gałce ocznej i zaburzeniami ucha wewnętrznego. Objawy zespołu Cogana w uchu wewnętrznym pojawiają się nagle i przypominają objawy choroby Meniere'a: ciężkie zawroty głowy, nudności, wymioty i zwykle obustronne, postępujące odbiorcze ubytki słuchu wraz z towarzyszącymi szumami usznymi.

Opis przypadku: 59-letnia kobieta została przyjęta do Kliniki Otolaryngologii i Chirurgii Głowy i Szyi w celu wszczepienia implantu ślimakowego. W wieku 57 lat u pacjentki stwierdzono zapalenie twardówki. Około roku później pojawiły się zawroty głowy i nudności, a następnie szybko postępujące ubytki słuchu - najpierw w uchu lewym, później w prawym, wraz z szumami usznymi. W chwili implantacji wynik testu rozróżniania mowy przez pacjentkę wynosił 0% w obu uszach. Biorąc pod uwagę wyniki badań audiometrycznych i diagnostyki obrazowej, wykonano kochleostomię podczas implantacji ślimakowej. W okresie pooperacyjnym nie zaobserwowano powikłań. Po 24 miesiącach używania procesora mowy subiektywna ocena zrozumiałości mowy określana przez pacjentkę w skali 0-10 była na poziomie 8,5 w cichym i 5,0 w hałaśliwym otoczeniu. Pacjentka mogła zidentyfikować 95% wyrazów jednosylabowych w ciszy i 35% w szumie (w warunkach stosunku sygnału do szumu +10 dB, poziomie mowy 70 dB HL i poziomie szumu 60 dB HL). Uzyskane korzyści zostały potwierdzone wynikami kwestionariusza oceniającego jakość życia z urządzeniami wspomagającymi słyszenie oraz korzyści płynące z jego użytkowania (APHAB). Pacjentka obecnie kwalifikuje się do implantacji ślimakowej w drugim uchu.

Wnioski: Implantacja ślimakowa skutecznie wpłynęła na poprawę słuchu u pacjentki z zespołem Cogana.

Słowa kluczowe: implanty ślimakowe • zespół Cogana • korygowanie ubytków słuchu • słuch

Background

Cogan's syndrome is a very rare autoimmune disease characterized by the coexistence of inflammatory lesions in the eyeball and inner ear dysfunction. Even though several hundred cases of Cogan's syndrome have been described in the literature, its prevalence rate is still not established [1], but in the majority of cases it affects adults. It seems that the prevalence is similar in men and women.

A typical patient with Cogan's syndrome presents with nonsyphilitic interstitial keratitis and disorders of the labyrinth comparable to those observed in Meniere's disease. The time-lag between the onset of ophthalmic and auricular symptoms is less than 2 years. Cogan's syndrome

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was first described in 1945 by ophthalmologist David Cogan [1–4]. In contrast to the typical presentation, atypical Cogan's syndrome is characterized by delayed-onset inner ear disorders, again similar to those reported in Meniere's disease, which coexist with the ophthalmitis (i.e., episcleritis, scleritis, retinal vasculitis, subconjunctival haemorrhage, or macular oedema, with or without keratitis), but here the time lag between the occurrence of the ophthalmic symptoms and the auricular symptoms is longer than 2 years [3].

In some patients (especially during the initial period of the disease or in the case of atypical Cogan's syndrome), nonspecific systemic symptoms occur and the inflammation affects other organs also. In such cases, vasculitis often develops and affects the cardiovascular, digestive, nervous, or osteoarticular system. Progression to the cardiovascular system is most commonly manifested by symptoms of aortitis, although some patients develop systemic vasculitis, which can be fatal in the cases of coronary arteritis or subarachnoid hemorrhage [5,6]. When smaller blood vessels are affected, the syndrome may appear as acute limb ischaemia, foot or hand necrosis, or abdominal pain caused by intestinal ischemia. The symptoms of the osteoauricular system affection mainly include arthralgia or myalgia, rarely arthritis. Patients can also report headaches in cases of nervous system infection. Sometimes it may also result in hemiparesis, hemiplegia, or pyramidal or cerebellar syndromes [5,7–9].

The symptoms of Cogan's syndrome within the inner ear are abrupt and resemble Meniere's disease: severe vertigo, nausea, vomiting, and tinnitus, which persist until sensorineural hearing loss develops, usually bilaterally. At that point, the previous symptoms disappear or become milder. Hearing loss quickly worsens, and after 1–3 months it almost always develops into complete deafness [3].

To the authors' best knowledge, only a few articles have so far been published describing cochlear implantation in patients with Cogan's syndrome [10–19]. There are at least two reasons. First, Cogan's syndrome is quite a rare disorder in itself, and its prevalence is currently not well known. Second, cochlear implantation in Cogan's syndrome patients could reflect the difficulty associated with properly placing electrodes into scala tympani because of the latter's obliteration [10,11,13]. In addition, the pathology of the disease adversely impacts the rehabilitation process, since atrophy of the cochlear nerve and spiral ganglion [3,20] may reduce the number of active electrodes and impair the cochlear implant's effectiveness. For example, Bovo et al. reported major complications in 3 cases of patients with Cogan's syndrome [10]. In the first patient, cochlear ossification was found, which caused a deterioration in speech ability from 90% (3 months after implantation) to 80% at the 1-year follow-up. In the second patient, progressive atrophy of the auditory nerve or decline in spiral ganglion cells was suspected, manifesting in a decrease in reported loudness with no change in electrode impedance. In the third patient, interestingly, the authors hypothesized that poor understanding of speech was due to auditory dyssynchrony.

Taking into account the above-mentioned difficulties, together with the extreme rarity of reports of cochlear implantation in patients with Cogan's syndrome (less than 30 cases in total for the studies cited 10–19), the aim of this study was to present our long-term clinical experience with a 59-year-old woman who successfully underwent cochlear implantation.

Case report

In July 2014, a 59-year-old woman was admitted to the Otolaryngology Head and Neck Surgery Clinic of the Institute of Physiology and Pathology of Hearing to undergo cochlear implantation. Her medical history was as follows. At 57 years of age, the patient was diagnosed with scleritis, which was initially treated with Dexamethasone 0.1% instilled in the conjunctival sac. Subsequently, as no remission occurred, Prednisolone 80 mg in oral dosage was introduced. After about a year, vertigo and nausea arose, followed by rapidly progressing hearing loss – at first in the left, and then in the right ear. Finally, concomitant tinnitus appeared. Despite an intensified therapy of systemic Methylprednisolone and Methotrexate 15 mg, the patient's hearing loss progressed and tinnitus increased, although vertigo and nausea were reduced and became rare. The patient began using hearing aids in May 2013, at first bilaterally and then in her right ear only. Since December 2013 she did not wear any aid due to lack of observable benefits. Up to the present day, the patient continues to receive Prednisone 5 mg in oral dosage.

The patient, diagnosed with Cogan's syndrome, was referred to our department after a differential diagnosis from the Rheumatology Department outside our hospital. The coexistence of concomitant hearing loss was then confirmed in our Department. Due to the preliminary diagnosis of profound sensorineural hearing loss, the patient was referred to further audiological evaluation aimed at determining her candidacy for an implantable device. Subsequently, in April 2014, the verification commission stated that the woman was eligible for cochlear implantation, and the patient was referred for this procedure. At the time of implantation, speech discrimination was 0% in both ears. Pure tone audiometry results according to ISO 8253-1: 2010 are shown in Figure 1. Impedance audiometry revealed bilateral type A tympanograms, but acoustic reflexes were not present.

Magnetic resonance angiography (MRA) of the patient's brain (T2, 3D TOF, and Art. 3D sequences) showed no vascular lesions. A head CT revealed no focal lesions in the central nervous system. However, a high-resolution CT scan (HRCT) of the temporal bones attracted special attention, as it indicated bilateral increased sclerosis of the modioli and thickening of the spiral lamellae at the basal turns of both cochleae (Figure 2). These lesions imply at least partial atrophy of the spiral ganglia and cochlear nerves. Additionally, bilateral atresia of the scala tympani in the area next to the round window was displayed, as well as a subtle narrowing of the scala vestibuli in the area next to the oval window. Moreover, bilateral narrowing of the round windows was observed. The middle ear appeared normal, subsequently confirmed by video-otoscopic and microscopic images. Furthermore, a CT displayed typical topography of the facial nerve.

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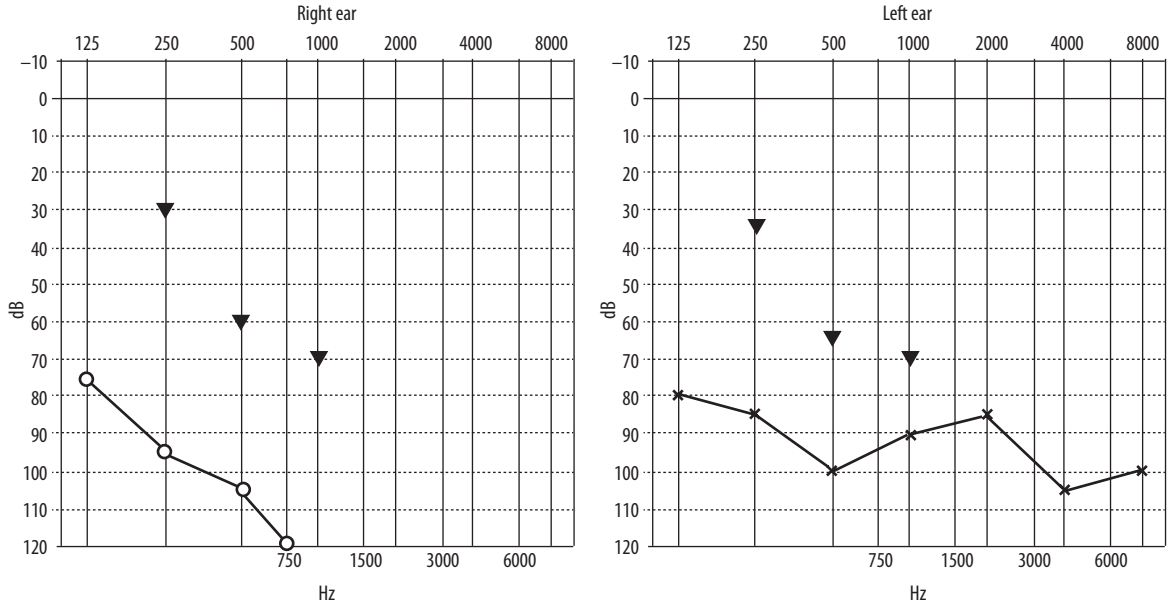

Figure 1. Results of preoperative pure tone audiometry (July 2014) for the right and left ears

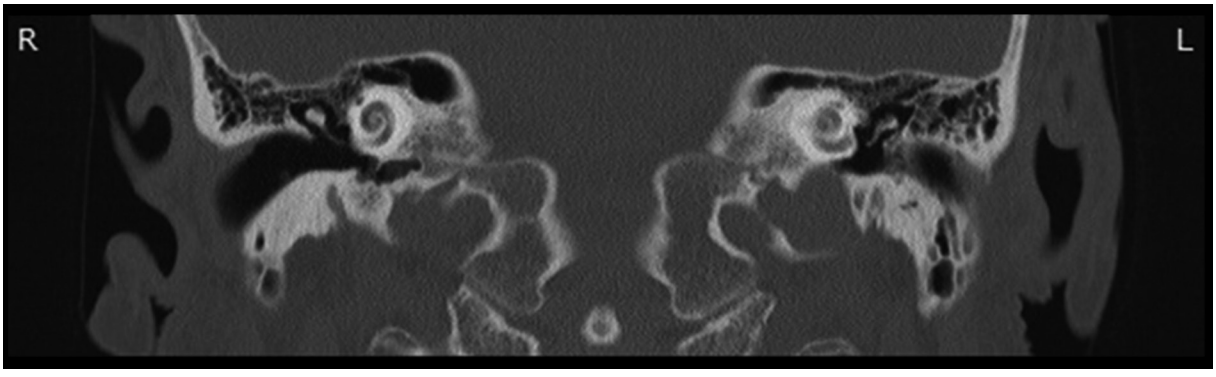

Figure 2. Preoperative HRCT scan of temporal bones showing bilateral sclerosis of modioli and thickening of the spiral lamella at the basal turns

In view of the above, promontorial cochleostomy [21,22] was carried out during the cochlear implantation. Cochlear implantation was conducted unilaterally and, based on the audiogram, the right (worse-hearing) ear was chosen. An active electrode array was inserted, although several resistances were noted during the process. Although we routinely prefer round window insertion, in this case the complicated anatomy (e.g. facial nerve canal position and challenging conditions of the round window niche) justified a very delicate cochleostomy [23,24]. No complications were observed, either during surgery or in the post-surgical period.

Results

Since the operation, the patient has been using her cochlear implant for 28 months and has remained under regular medical supervision of the Rehabilitation Clinic. Monosyllabic speech tests in quiet were done 4 months after CI activation, and discrimination using the CI was only 5%. Some 9 months after activation of the speech processor, free-field audiometry was done. The patient could hear overr a wide spectrum of frequencies: for frequencies of 0.25–0.5 kHz, responses were about 50–70dB HL, and for 1–6 kHz levels were 30–40 dB HL (Figure 3).

Some 9 and 24 months after activation of her speech processor the patient completed an APHAB questionnaire, and the results are presented in Figure 4.

As shown in Figure 4, the patient initially had difficulties with communication in silence at a level of 43%, which then decreased to 16.3%. Moreover, the patient first reported serious problems in communicating in noisy (93%) and in reverberant (95%) conditions, which were alleviated after 2 years of using the implantable device: problems with the first aspect were reduced to 76.5% and problems with the second were reduced to 82.8%. However, hyperacusis increased from 74% to 93%, probably because of the broad spectrum of frequencies perceived with the cochlear implant.

The APHAB questionnaires revealed that her initial problems and difficulties in communicating had been partially alleviated. In addition, during the verification of electrical stimulation parameters in the 24-month follow-up,

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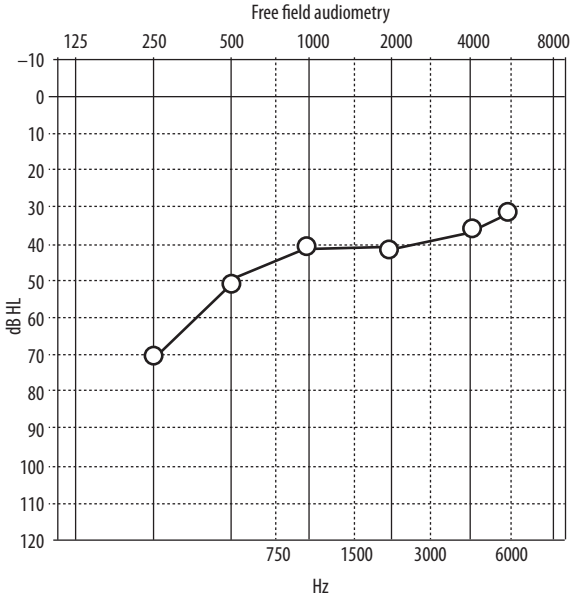

Figure 3. Pure-tone audiogram (free-field) of the patient after 9 months of CI use

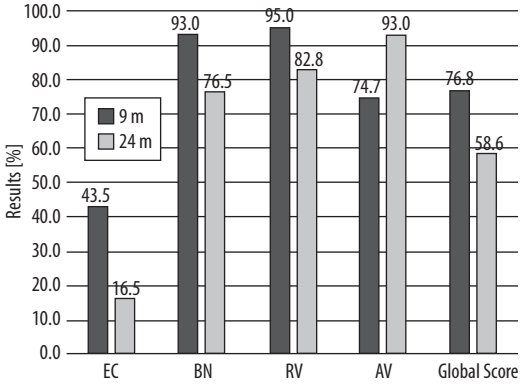

Figure 4. Results of the APHAB questionnaire 9 and 24 months after speech processor activation. EC – ease of communication; BN – background noise; RV – reverberation; AV – aversiveness; right column shows global score

the subjective assessment of speech intelligibility given by the patient (on a 0–10 scale) was 8.5 in quiet and 5.0 in noise. Cochlear implant satisfaction level given by the patient was 10. After using the implant for 2 years, the patient made enormous progress as reflected by the monosyllabic words in silence discrimination, which was 95% in quiet and 35% in noise (SNR +10 dB, with the speech level at 70 dB HL, and noise level at 60 dB HL). Unilateral implantation generally provides good speech understanding under quiet conditions, however patients frequently report difficulty understanding speech when exposed to background noise and with sound localization [25]. Currently, the patient is qualified for a CI in the left ear to improve her hearing ability by providing bilateral hearing.

Discussion

Cogan's syndrome is a rare disease which can be treated with immunosuppressive therapy if diagnosed early. Drug-based treatment uses topical glycocorticosteroids application directly to the eye, or systemically. The following medications are also used: methotrexate, cyclophosphamide, azathioprine, cyclosporine, mycophenolate, and mofetil. In recent years, the disease has also been treated with drugs such as TNF-alpha blockers (etanercept and infliximab) or rituximab [6,26,27], which can stop damage to the labyrinth in the early phases of the disease.

However, in most cases (up to 80%) irreversible bilateral hearing loss occurs, despite any introduced drug therapy [10,28–30]. Hence, the only way to help such patients in returning to the world of sounds is cochlear implantation. Because the syndrome is rare, there are very few reports of how the patient might benefit from cochlear implants; altogether less than 30 cases have been described [10–19]. Overall results of such treatment are good, but only if all the channels of the electrode array are properly placed in the cochlea [11–14]. For instance, Kawamura et al. [28] stated that in this group of patients, speech identification after one year of implant use was 80%. Bacciu et al. [19], in the largest group of implanted patients with Cogan's syndrome, presented results with levels of 75–100%. Kontorinis et al. stated that among 3000 patients implanted with a CI in their center between 1992–2007, only 4 were diagnosed with Cogan's syndrome [16]. In a follow-up period of almost 10 years, the authors presented long-term results of auditory rehabilitation (2 of 4 patients were implanted bilaterally) giving 82.5% for the monosyllabic word test. Wang et al. compared the results obtained by patients with autoimmune inner ear disease (including 7 Cogan's syndrome cases) with age- and sex-matched controls and stated that, surprisingly, patients with a CI and autoimmune inner ear disease achieved significantly higher results ($p<0.05$) than a control group at all follow-ups. However, it is worth stressing that cochlear ossification, noted in some other studies, was not observed in these patients.

In most of the patients who suffer hearing loss from Cogan's syndrome, imaging scans may reveal inner ear lesions, such as atresia of scala tympani. By contrast, some other patients with image scans showing no abnormalities were recognised to have such lesions intraoperatively [1,11,18,19]. In post-mortem histopathological examinations of the temporal bones performed in Cogan's syndrome patients, the following lesions have been reported: sclerosis of the modiolus, atrophy of the elements of the spiral ganglion (also with lymphocytic and plasmocytic infiltration), atresia of the scala tympani (connective tissue, bone), or atrophy of the cochlear nerve. Furthermore, stria vascularis degeneration or endolymphatic hydrops may be seen [3,20]. These lesions can be challenging for an otosurgeon [19], and can additionally lead to misplacement of the electrode array [10], diminishing the benefits of cochlear implantation.

In the presented case study, the entire electrode array was successfully placed in the cochlea, even though a CT scan of the temporal bones revealed bilateral atresia of the scala tympani in the area next to the round window. Moreover,

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despite partial atrophy of the elements of the spiral ganglia and the facial nerves suggested by the CT scans (i.e. increased sclerosis of the modiolus and thickening of the spiral lamellae at the basal turns), the rehabilitation outcomes are satisfactory. There are also no changes in electrode impedance in the implant. The patient has been deemed eligible for cochlear implantation of the other ear.

Currently, there are no self-report questionnaires assessing hearing ability in patients with comorbidities, as happened in our patient's case. In such patients, there are additional expectations than in otherwise healthy adults with SNHL only. For such cases our aim is first to provide the best communication with the world. Better understanding in noise, or improved hearing in demanding conditions such as reverberation, is really a secondary benefit. We believe that the subjective opinion of the patients – who is expert in her own hearing and can compare both conditions (before the CI and 2 years after the operation) – is the best indicator of the procedure's success or failure. The benefits from the CI are clearly reflected in the patient's subjective satisfaction from the device – declared as 10/10 possible points. So as not to bias our results, we have also presented APHAB scores which show that there were observable benefits (a general improvement of about 20%), although not as high as in a healthy patient with SNHL.

Conclusions

Cochlear implantation in patients with Cogan's syndrome is in most cases the only way to restore hearing ability and communication with the surrounding world. Due to anatomical disorders, the CI procedure is very challenging, and this alone might contribute to the extremely low number of publications dealing with the issue. The presented case study proves that substantial benefits can be obtained, and the patient is currently awaiting cochlear implantation of the other ear.

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