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Cochlear implantation in patients suffering from Cogan's syndrome

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Abstract
Among patients who receive cochlear implants, those with Cogan's syndrome make a unique group. On one hand they are part of the post-lingual patients and good results can be anticipated. On the other hand, their basic illness is thought to have an autoimmune aetiology and for that reason more susceptible to complications, especially flap problems. In a series of 60 patients who were implanted at the Sheba Medical Center, three had Cogan's syndrome. No post-operative complications, including flap problems, were observed (followed-up for at least 18 months). Subjective and objective hearing results were very good.

Key words: Cogan's syndrome; Cochlear implant

Introduction

Cogan's syndrome is a rare clinical entity considered to be an autoimmune disease. It was first described by David Cogan in 1945 and characterized as nonsyphilitic interstitial keratitis with audiological and vestibular symptoms. Some patients may develop uveitis, scleritis, retinal haemorrhages without interstitial keratitis (Allen *et al.*, 1990). Seventy per cent of the patients have systemic manifestations which include; adenopathy, gastrointestinal bleeding, aortic regurgitation, musculoskeletal complaints, splenomegaly and neurological symptoms. Systemic necrotizing vasculitis has been reported in about 10 per cent of the patients (Haynes *et al.*, 1980; Allen *et al.*, 1990; Bulory *et al.*, 1990; Schuknecht, 1991; Hammer *et al.*, 1994).

The audiological and vestibular manifestations usually fluctuate but are progressive over a period of months rather than sudden. The inner ear pathology shows severe degeneration of the organ of Corti, macula and crista with total loss of hair cells, fibrous tissue and bone proliferation that involves the cochlea and vestibular labyrinths. Endolymphatic hydrops can also be evident (Schuknecht, 1991). Since these patients become deaf after years of hearing, employing a cochlear implant can offer an excellent outcome.

Complications associated with cochlear implant surgery are at an overall range of 6.8-12 per cent most of which are flap complications (Cohen *et al.*, 1988; Cohen and Hoffman, 1991; Webb *et al.*, 1991). Those can be major, such as wound breakdown, necrosis or infection that require removal of implant. Other can be minor, such as seroma or an uncomplicated infection (Harris and Cueva, 1987; Wang *et al.*, 1990).

As Cogan's syndrome patients suffer from vasculitis one could anticipate they would have more flap complications. This paper presents three patients suffering from Cogan's syndrome, in a series of 60 implantations, with whom we had no flap complications and remarkable hearing results.

Case reports

Case 1

A 53-year-old woman known to have rheumatoid arthritis, first presented with left sensorineural hearing loss and vertigo. She deteriorated to binaural profound, sensorineural hearing loss. Anterior uveitis and interstitial keratitis occurred within a period of three months. High resolution computed tomography (CT) of the brain, cerebellopontine angle and temporal bones were normal. Electronystagmography (ENG) showed bilateral areflexia. Audiometry showed bilateral profound sensorineural deafness. The rheumatoid-factor titre as 1:40. VDRL testing was negative. Echocardiogram was normal. The diagnosis of Cogan's syndrome was ultimately made.

A year and a half after complete deafness the patient underwent a left cochlear implantation with a Nucleus 22 channel device. A wide inferiorly based, inverted 'J'-shaped flap was carried out. Twenty-two active channels were inserted through the facial recess into the scala tympani via a cochleotomy.

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FIG. 1
Skin markings of the inverted 'J' flap and implant placement.

Through a separate skin incision a soft silicone drain was left in the mastoid, kept without vacuum and removed at the third post-operative day. The skin was closed with metal clips (Figures 1, 2 and 3). Antibiotic prophylaxis with cephalosporins were given by the iv route perioperatively and continued orally for one week. The post-operative period was without complications. There was no flap breakdown, necrosis, extrusion, seroma or infection and the skin incision healed well. After 18 months no flap problems have developed.

One month after the operation, the process of implant programming and auditory training began. Speech perception evaluation conducted one year post-implantation using the Spectra Speech Processor demonstrated excellent open speech perception ability. On the two-syllabic word recognition test, Hebrew monosyllabic, phonetically-balanced (PB) word discrimination, and everyday sentence recognition test, the patient scored 100 per cent, 40 per cent and 86 per cent respectively. The patient uses the telephone to communicate.

Case 2

A 36-year-old male, first presented with left sensorineural hearing loss, vertigo and keratoconjunctivitis at age of 15. In a one month period he progressed to have binaural complete deafness and Cogan's syndrome was diagnosed. Prior to implantation, ENG demonstrated bilateral areflexia, computed tomography (CT) of the brain, cerebellopontine angle and temporal bones were normal and audiometric tests showed bilateral deafness. VDRL testing was negative. Nineteen years after complete deafness the patient underwent a right cochlear implantation with a Nucleus 22 multichannel device. A wide inferiorly based 'U' shaped flap was elevated. Twenty-two active channels plus five were inserted through the facial recess to the scala tympani by a cochleotomy. A soft silicone drain was left open for three days. Metal clips were used for skin closure. Prophylactic antibiotics (cephalosporins) were given for 24 hours iv, and for another week po. The post-operative period was without local or systemic complications, and within a three years' follow-up there were no flap problems.

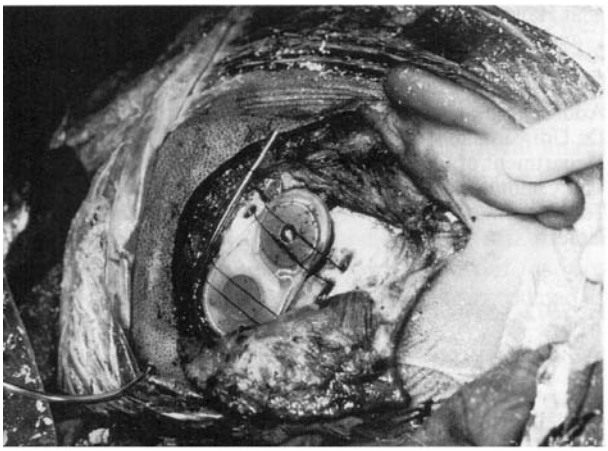

FIG. 2
Skin flap elevated and implant secured in place with Nylon stitches. A silicone drain above the implant.

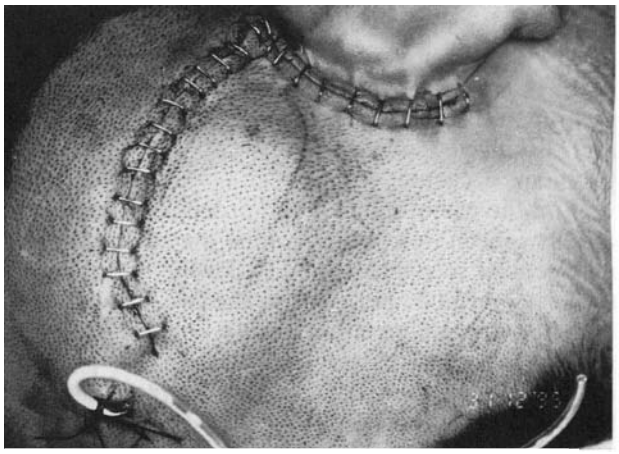

FIG. 3
Skin closed with metal clips and the drain left 'open', with no vacuum.

This young adult obtains remarkable benefit from his Spectra Speech Processor. He works for a large company and receives telephone calls unhindered, using the telephone fluently and frequently. He also obtains excellent results on open set recognition speech tests: for recognition of two-syllable words, PB words and every day sentences he attains 90 per cent, 50 per cent and 96 per cent respectively.

Case 3

A 38-year-old male first complained of tinnitus and vertigo at age of 23. Three weeks later he developed a profound, bilateral hearing loss and interstitial keratitis. Diagnosis of Cogan's syndrome was made. Before implantation a CT of the brain, cerebellopontine angle and temporal bones were carried out and found to be normal. ENG demonstrated bilateral areflexia and audiometric tests showed bilateral deafness. VDRL testing was negative. He was implanted with a Nucleus 22 channel mini system device, 15 years after diagnosis. At surgery, an inferiorly based inverted 'J' flap was elevated and 22 active channels were inserted to the scala tympani through a cochleotomy. A soft silicone

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drain was left in the mastoid open for three days. Antibiotic prophylaxis with cephalosporins were given iv for 24 hours and po for one week. In the post-operative period no complications were seen and during a follow-up for two years no flap problems occurred.

With the use of mini-speech processor, the patient attains speech perception results that demonstrated no open-set recognition ability. However, in closed-set he achieved 55 per cent for the two-syllable word test. On vowels and consonants recognition subtests (of the Iowa test battery) he achieved 70 per cent and 55 per cent respectively. The implant supports the speech-reading ability of this patient, demonstrated by an increase from a 55 per cent score, visual only, to 82 per cent on auditory visual CID sentences.

Discussion

Sixty implantations (including three reimplantations) were performed at the Sheba Medical Center between June 1989 and June 1996. Three of those patients suffered from Cogan's disease. In the first 22 implantations an inverted 'U' flap was employed, and with the following 38 patients an inverted 'J' flap was used. We found this flap to have a wide axial blood supply from the posterior auricular and occipital arteries. After elevating the flap, a cortical mastoidectomy was performed, the facial recess opened and the electrode inserted to the cochlea through a cochleotomy. As a routine, perioperative antibiotic prophylaxis was administrated (cephalosporins, if not contraindicated), a mastoid silicone drain was left open (with no vacuum) for three days and the skin incision closed with metal clips.

The literature reports most of the complications to be flap problems including infection, breakdown, necrosis and extrusion (Harris and Cueva, 1987; Cohen *et al.*, 1988; Wang *et al.*, 1990; Cohen and Hoffman, 1991: Webb *et al.*, 1991). In our series we had a surgical complication rate of 10 per cent (including two cases of device failure, three patients had temporary facial paresis that resolved completely and one case of perilymphatic fistula). The non-surgical complication rate was 6.6 per cent (including two cases that had seroma and two suffering post-operative otitis media that were treated successfully with antibiotics). The three patients with Cogan's syndrome had no post-operative complications at all.

Except for a description by Wang (Wang *et al.*, 1990) we have not found in the literature a specific mention of cochlear implanted patients with Cogan's syndrome, their post-operative complications and results of hearing. Wang reports a patient who was implanted for the second time on the same side and suffered from superficial sloughing at the flap site. He attributes this complication to a deprived blood supply due to the prior surgical scarring.

It is of interest to mention that in all three patients the cochleae were patent as demonstrated on CT and proved during surgery, as all 22 active channels were inserted easily (with one case – plus five).

The three patients were adults when they became deaf (post lingual). As expected, they gained a great benefit from the implant, a fact that is revealed from their excellent test achievements, by regaining social abilities and their own statements.

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