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Cochlear Nerve Stimulation in the Internal Auditory Canal in Ossified Cochlea: A Case Study.

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ABSTRACT

Objective: To present the first reported case of intraneural direct cochlear nerve stimulation in a human being.

Study design: case report.

Results: A 23-year-old patient with bilateral progressive hearing loss associated with bilateral complete semicircular canal aplasia and ossified cochleas underwent cochlear implantation. During surgery, a patent cochlear lumen could not be found, and the array was positioned in the internal auditory canal adjacent to the cochlear nerve. Against our expectations, an assiduous rehabilitation and frequent fitting adjustments have led to a word recognition score, in open set speech with lip reading, of 18/25 and acceptable frequency discrimination.

Conclusions: We are aware that this was an anomalous use of the cochlear implant, and it is not our aim to suggest a new indication for cochlear array positioning. However, this case shows that auditory perception, to some degree, can be obtained with intraneural direct cochlear nerve stimulation.

Keywords: cochlear implant, cochlear nerve, otology.

INTRODUCTION

Although technology evolution has been providing surgeons with new devices capable of overcoming many difficulties, even in severe inner ear malformations, the ossified cochlea still represents a challenge in cochlear implantation. In such cases, electrode array misplacement is not rare. We report a case of cochlear implantation in the internal auditory canal in a patient with complete semicircular canal aplasia and small ossified cochleas.

Computed tomography (CT) can reveal developmental anomalies of the inner ear during the preoperative evaluation of cochlear implant candidates (1). Among inner ear malformations, the prevalence of semicircular canal (SCC) anomalies varies with the overall severity of the anomaly: mild anomalies of the lateral SCC are the most common, while isolated aplasia of the SCC is very rare.

In general, syndromic and non-syndromic cases of isolated semicircular canals aplasia are eligible for cochlear implantation (2). In slightly permeable cochlea, the insertion of a conventional electrode array is impossible, and in many cases, cochlear implantation with conventional electrodes is contraindicated (3).

Currently, various types of electrodes (short or split) are designed for neural stimulation and are useful alternatives to traditional cochlear implants in treating deaf patients with cochlear ossification (4, 5).

To the best of our knowledge, the electrical stimulation of cranial nerve VIII with the multielectrode array in the internal auditory canal has not been reported in the literature to date.

CASE REPORT

In February 2011, a 23-year-old native Rumanian male presented at our outpatient clinic with bilateral anacusis. His hearing had been normal until the age of nine years, when bilateral progressive hearing loss began. At the age of 12 years, he was fitted with bilateral hearing aids. At 16 years old, he moved to Italy and, despite his moderate hearing loss, he was able to learn Italian. He completed high school and was employed. He then experienced further hearing loss until complete bilateral deafness; at first consultation he was no longer wearing his hearing aids. He had no history of meningitis and was otherwise healthy.

We used tests extracted from the Italian version of the “Rehabilitation Manual” (6) and CAP (Categories of Auditory Performance) to assess his auditory perception; his lip reading skills in Italian were initially very poor and barely improved after speech therapy (from seven to nine words out of 25).

Computed tomography (CT) and magnetic resonance imaging (MRI) revealed a bilateral absence of the semicircular canals and small, severely ossified cochleas; a very thin cochlear lumen could be appreciated on scrupulous analysis of the CT scan (Fig. 1a-b).

Pure tone audiometry confirmed bilateral deafness.

In March 2011, cochlear implant surgery with a Med-EL CONCERTO compressed electrode array (Med-EL, Innsbruck, Austria) was performed under general anesthesia.

After mastoidectomy, the middle ear was reached through a posterior tympanotomy and the landmark structures (oval window, round window and promontory) were visualized. The cochlea was drilled to reach the lumen seen on the preoperative CT scan, which we could not find. However, having inadvertently punctured the internal auditory canal (IAC), we decided to insert the array into the IAC adjacent to the cochlear nerve, fixing it and sealing the hole with pieces of muscle and fibrin glue to prevent the leakage of CSF (Fig. 2). The electrode impedances were low for all 12 electrodes and the neural response telemetry (NRT) was adequate for electrodes 4 to 12. Stimulation of the apical electrodes 1, 2 and 3 elicited facial contraction. No facial weakness was observed after awakening.

In April 2011, the implant was activated. Initially a continuous interleaved sampling (CIS) strategy was used. In a CIS strategy, the frequency information of the incoming sound is processed on a tonotopicity basis. We decided to assess his improvements both with ordinary verbal tests and frequency discrimination tests. Figure 3 shows his scores over time.

At first, auditory perception was possible only with electrodes 7 and 8 because the patient reported discomfort with the others. The apical electrodes (1, 2 and 3) were disabled because of facial nerve stimulation. Frequency discrimination was very poor, with only two different tones distinguished within an interval of four octaves.

In July 2011, following three months of speech therapy and CI mapping, all electrodes from 4 to 12 evoked auditory perception; however, he was still unable to distinguish sounds from speech and could only detect acoustic stimuli.

He then started to slowly improve and in December 2011 he could identify words and sentence lengths (10/10 at both tasks) and differentiate minimal pairs depending on their second formants (8/10).

In August 2012, he was able to discriminate between male and female voices. During word recognition tests using uncommon words with lip reading support, he performed well with words both with deep and high acoustic components (7/10 at both tasks), but he still had difficulties recognizing words with a medium - deep acoustic component (2/10).

In October 2012, the CIS strategy was replaced with the temporal fine structure strategy (FS4); with this strategy, the electrodes stimulate the nerve in sync with the frequency of the incoming sound. Electrodes 1, 2 and 3 remained switched off, and we assigned FS4 to electrodes 4, 5 and 6. The frequency range coded by these electrodes was from 150 to 600 Hz. Electrodes 7 to 12 had constant stimulation rates.

In May 2013, seven months after the initiation of the FS4 strategy, on the uncommon words recognition test, he improved for words with deep and medium-deep acoustic components (9/10 and 7/10, respectively), whereas he performed slightly worse for words with high acoustic components (5/10).

On isolated word recognition in open set speech with lip reading, the patient recognized 18 words out of 25. His frequency discrimination improved, and he could distinguish two different tones within an interval of approximately five semitones. Free field pure tone audiometry with CI showed a pure tone average (0.5-4 kHz) of 40 dB HL.

The patient has done very well so far, and although he is a potential candidate for ABI, he prefers to continue using his CI.

DISCUSSION

We report a case of a cochlear implant multielectrode array positioned in the internal auditory canal. The first report of direct acoustic nerve stimulation dates back to 1957, when Djourno and Eyries implanted a single-electrode device in a deaf 50-year-old patient (7). Although their patient developed some word recognition abilities, it is still debated whether their device elicited acoustic responses by stimulating the nerve or the cochlear nuclei (8). There is only one recent report of cochlear electrode misplacement: a four-year-old male with a right cochlear implant in the internal auditory canal positioned two years earlier (9). Audiologic testing demonstrated normal impedance in the child but irregular telemetry mapping and no significant improvement in his auditory perception or behavioral response despite several device map adjustments.

In our patient, positioning of the electrode array in the internal auditory canal had not been preoperatively planned. Because imaging revealed severely ossified and small cochleas, we discussed with the patient the high risk of failure, in which case, an auditory brainstem implant (ABI) would have been indicated, as suggested by the literature. In their case report, Sanna and colleagues (10) conclude that cochleostomy should first be attempted to evaluate the option of CI, reserving ABI for patients unlikely to benefit from CI.

The electrode array position remained stable (follow-up of 27 months) and the impedances remained unchanged. The distal electrodes (1-3) remained switched off because they stimulated the facial nerve. A likely explanation is the bending of the array tip (Fig. 2B). Badi and colleagues demonstrated in a cat model that narrow-band frequency perception could be selectively evoked with direct cochlear nerve stimulation via intraneural electrodes, which selectively excites small subpopulations of cochlear nerve fibers. Our case provides in vivo confirmation of this observation (11).

We observed that after switching from a CIS to an FS4 strategy, the patient auditory perception and CAP scores improved.

This result was probably due to the differences in frequency analysis of the incoming sound between the cochlea and the cochlear nerve. The former mainly uses place coding, whereas the latter uses the stimulation rate.

In an FS4 strategy, the apical electrodes have a stimulation rate in sync with the frequency of the incoming sound; in our patient, this strategy provided physiological stimulation of the cochlear nerve in the frequency range of 150 – 600 Hz.

Despite his poor lip reading skills, comparison of the word recognition test scores obtained with the CIS strategy and the FS4 strategy shows an improvement from 7/10 to 9/10 for words with a deep acoustic component, but a slight decrease from 7/10 to 5/10 words with a high acoustic component, confirming that frequency analysis is more accurate at low frequencies. The patient can discriminate a frequency interval of five semitones in the central octaves of the piano (130 Hz – 520 Hz). Satisfied with his daily auditory performance, he can now detect environmental sounds and discriminate them. Some concerns exist in case of future revision surgery. Should this occur, we would not replace the CI but opt directly for ABI surgery. We believe that the main issue could be removing the CI array from the IAC without damaging the facial nerve; cutting the array and leaving its distal part inside the IAC could solve this problem and prevent facial nerve lesions.

In conclusion, while this case does not suggest a new indication for cochlear array positioning, it does show that auditory perception, to some degree, can be obtained with direct intraneural cochlear nerve stimulation in a patient with profound deafness due to small ossified cochleas.

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Fig. 1 Axial (A) and coronal (B) temporal bone CT scan.

Fig. 2 Postoperative CT scan in axial (A) and coronal (B) planes.

Fig. 3 Auditory performance over time. Pre-op SLP: preoperative after speech therapy; WR: word recognition with lip reading; WL: word length identification; SL: sentence length identification; 2ndF: minimal pair discrimination by the second formant; D_W: deep acoustic component word recognition; MD_W: medium-deep acoustic component word recognition; H_W: high acoustic component word recognition.