Cochlear Implantation in Patients with Cochlear-Facial Dehiscence
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ABSTRACT

Objective: To describe patients with cochlear facial dehiscence (CFD) who develop facial nerve stimulation (FNS) after cochlear implantation (CI).

Study Design: Case series

Methods: The medical charts and imaging of three patients with bilateral sensorineural hearing loss (SNHL) who presented for CI evaluation were reviewed.

Results: Case 1 (C1) had a history of Meniere’s disease. Case 2 (C2) had a history of medulloloblastoma treated with surgery and chemoradiation. Case 3 (C3) had a history of progressive SNHL. Audiometry showed moderate-to-severe SNHL in P1, severe-to-profound SNHL in P2 and profound SNHL in P3. All had poor speech discrimination ability. Temporal bone CT coronal views were suspicious for bilateral dehiscence between the superior basal turn of the cochlea and labyrinthine segment of the facial nerve in C2 and C3, with maximum dehiscence lengths of 2.0 mm on the left in C2, and 1.8 mm on the right in C3. A thin bony partition was visualized bilaterally in C1. The left ear of C1, left ear of C2 and right ear of C3 were implanted. FNS occurred immediately upon activation in C1 and C2, which resolved by decreasing the dynamic range of the offending electrodes. No FNS was observed in C3.

Conclusion: CFD can predispose patients to post-implant FNS. Prior temporal bone irradiation may carry a higher risk of FNS. We recommend scrutiny in CTs of CI candidates and appropriate risk counseling for FNS if CFD is discovered.

INTRODUCTION

Cochlear facial dehiscence (CFD) is a recently described but uncommon dehiscence of the otic capsule between the basal turn of the cochlea and the labyrinthine segment of the facial nerve. Similar to other otic capsule dehiscences, this defect is thought to create a mobile third window. Dissipation of energy through this window results in a pseudo-conductive hearing loss.1 Patients with CFD can also present with vestibular and auditory symptoms, such as vertigo and autophony, similar to those with superior semicircular canal dehiscence (SSCD). A well-known complication of cochlear implantation is FNS which can lead to significant patient discomfort causing the need to limit electrical current on one or more electrodes.1

Dissection of 8 temporal bones showed that electrodes 8-11 on a Nucleus-22 electrode array were found to be closest to the labyrinthine segment of the facial nerve, further suggesting the importance of this segment in the pathogenesis of FNS.2 Electrodes 9-13 and electrodes 2-8 were responsible for FNS in Case 1 and 2 respectively, which is consistent with the previous findings.

Case 2 may have been at higher risk for post-CI FNS given his history of radiotherapy. We suspect that radiation-induced thinning or necrosis of the bony partition resulted in decreased impedance and hence electrical shunting.

Based on the increased risk for FNS in patients with CFD undergoing CI, we recommend careful review of all pre-operative CT scans for the presence of this dehiscence. Given the audiologic outcomes of these two patients, the presence of CFD and the risk of FNS should not exclude patients from CI; however, they should be counseled about the added risk of this complication.

REFERENCES


DISCUSSION

• The audiograms of all three patients showed profound SNHL narrating CI. Close examination of the CT scans revealed a thin bony partition between the basal turn of the cochlea and facial canal in Case 1, and apparent complete CDF in Cases 2 and 3.

• We demonstrate that patients with CFD can undergo CI with excellent audiologic outcomes, with the exception of a predisposition to FNS. In the setting of CFD, thinning or dehiscence of the bone may allow spread of electrical current or the electrode to come into close contact with the facial nerve causing direct stimulation.2

• Electrode position has also been shown to contribute to CI outcomes. Dissection of 8 temporal bones showed that electrodes 8-11 on a Nucleus-22 electrode array were found to be closest to the labyrinthine segment of the facial nerve, further suggesting the importance of this segment in the pathogenesis of FNS.2 Electrodes 9-13 and electrodes 2-8 were responsible for FNS in Case 1 and 2 respectively, which is consistent with the previous findings.

Case 1: An 81-year-old male with left-sided Meniere’s disease who sustained SNHL in the right ear from the sound of airbag deployment in a motor vehicle accident presented for CI evaluation. He had limited benefit from an in-the-ear hearing aid use. He denied other otologic complaints. Pure tone audiometry revealed moderately-severe SNHL in the right ear and severe SNHL in the left ear. Morurally aided speech discrimination ability was very poor (0% and 2%) in the right and left ears, respectively when assessed using the Hearing In Noise Test in Quiet (HINT-Q) at 47dBHL. Temporal bone CT shown in Figure 1A, B. A Cochlear Nucleus (S12) device was implanted on the left side. All electrodes were in compliance at one month follow-up. No FNS was noted at that time. The patient was seen for subsequent mapping 7 months post-activation, at which time the patient reported FNS (eye twitching) on the fourth progressive program (dynamic range of 36 current units) which was determined to be caused by electrodes 9-13. These electrodes were disabled. Electodes 9, 11-13 were later reintroduced without FNS by globally decreasing dynamic range and changing the pulse width. Electrode 10 continued to cause FNS (lip twitching) and remained disabled. The patient continued to report some intermittent, mild, and non-painful FNS in response to loud sounds. Three years post-implantation, the patient is able to hear within normal limits from 250-6000 Hz with excellent open set speech discrimination ability.

Case 2: A 34-year-old male with a history of medulloblastoma treated with surgical resection, cisplatin chemotherapy and radiation; meningitis; intracranial hemangioma, and bilateral progressive SNHL was referred to us for CI. The hearing loss was greater in the left ear with no ability of speech discrimination. He had been using a right In-the-Ear hearing aid. The patient denied other otologic complaints. Pure tone audiometry showed severe sloping to profound SNHL in the right ear and profound SNHL in the left ear. Speech discrimination ability was very poor (52% and 0%) in the right and left ears. Coronal views of his temporal bone CTs were suspicious for dehiscence (Figure 1C). A Cochlear Freedom Contour Advance (CI24RE) device was implanted on the left side. At initial stimulation of the Nucleus 6 speech processor one month later, all electrodes were in compliance. FNS was noted immediately during Auto Neural Response Telemetry (Auto-NRT) on electrodes: 1, 3, 6, 8, 10, 13 and 19. The patient was given progressive maps created based on psychophysics threshold levels. Maps did not reveal FNS until 6 months post-initial stimulation when the patient was noted to have significant left FNS (twisting of his left eye, nostril, upper and lower lip). Electrodes 2 and 11 were identical as the offending electrodes and were deactivated. The patient was seen again at 7 months post activation, at that time, his speech discrimination ability was noted to be slightly poorer (86% to 92%). The patient also reported increased hearing difficulty in noise. All offending electrodes were successfully reintroduced with smaller dynamic ranges determined by establishing comfort levels based on observed FNS. The patient currently receives excellent benefit from his cochlear implant: he is able to hear within normal limits and has excellent speech discrimination ability.

Case 3: A 59-year-old male with severe obstructive sleep apnea was referred to us for CI evaluation for a long-standing history of bilateral SNHL. His hearing loss resulted from exposure to loud noise. The hearing loss was worse on the right side and progressively worsened since his mid-30s. He had been using a left-sided hearing aid. Three months prior, he experienced a “popping” sensation and sudden loss of hearing in his left ear after blowing his nose. He denied any other otologic complaints. Audiometry revealed bilateral profound SNHL with no speech discrimination ability in both the aided and unaided conditions. Review of his temporal bone CT revealed CFD (Figure 1D). Prior to CI, the patient underwent uvealshapalopatroypharyngoplasty, lingual tonsillectomy and adenoidectomy for daytime somnolence and severe obstructive sleep apnea. Two months later, the patient underwent right CI with a Cochlear Freedom Contour Advance (CI24RE) device. One month following implantation, the patient returned to the clinic for initial stimulation of the Nucleus 6 speech processor. At initial stimulation, the implant test revealed all electrodes were in compliance and impedances were high yet flat. To date the patient has not experienced FNS. He receives excellent benefit from the CI; he is able to hear within normal limits from 250-6000Hz and has excellent open-set sentence discrimination ability in quiet and competing background noise.

CASE SERIES

Figure 1. High-resolution temporal bone CTs were performed on a GE Lightspeed Pro 16 (GE Healthcare, Milwaukee, WI) in three cases. (A) In Case 1, review of the CT showed very thin bony partitions between the basal turn of the cochlea and the labyrinthine segment of the facial nerve on the right. (B) Similar findings were seen on the left. (C) In Case 2, coronal views were suspicious for dehiscence between the cochlea and facial nerve and had a maximum length of 2.0 mm on the left side. (D) In Case 3, there was a maximum dehiscence length of 1.8 mm on the right side in the coronal view.

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