

# Severe and persistent facial nerve stimulation after cochlear implantation in a patient with cochlear–facial dehiscence: a case report

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## Abstract

Generally, cochlear implants (CIs) are effective in helping patients improve their hearing performance; however, some patients have poor hearing performance owing to facial nerve stimulation (FNS), which is often associated with cochlear anomalies. We report a case with a normal cochlea and severe and persistent FNS owing to cochlear–facial dehiscence (CFD) that affected the CI outcomes. Preoperatively, a careful review of the computed tomography images before CI surgery is necessary not only for patients with otosclerosis and inner ear malformations but also for patients with normal cochlear structures because facial nerve anomalies could be present.

## Keywords

Cochlear–facial dehiscence, cochlear implantation, facial nerve stimulation, case report, hearing impairment, computed tomography, facial nerve malformation

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## Introduction

Cochlear implants (CIs) are the most effective way to restore hearing in patients with severe or profound hearing loss. Initially, CIs were used only for patients with sensorineural hearing loss, but with technological and research advancements, patients with vestibular schwannoma are now also

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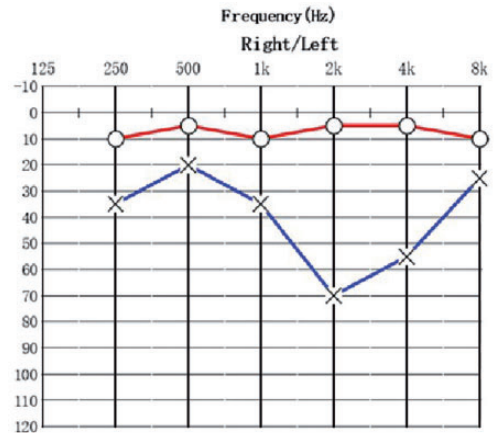
candidates for CIs. Arriaga and Marks<sup>1</sup> first reported a case of simultaneous cochlear implantation and vestibular schwannoma resection, in 1995.

Although most patients with CIs achieve good hearing rehabilitation after surgery, some postoperative negative effects, such as facial nerve stimulation (FNS) and vestibular dysfunction, may be seen. FNS often occurs in patients with otosclerosis and inner ear malformations,<sup>2</sup> and FNS may be resolved by decreasing the stimulation levels, widening the pulse width, and deactivating electrodes.<sup>3</sup> Sometimes, patients show poor hearing improvement owing to severe FNS.<sup>4</sup> Proper treatment of FNS helps reduce discomfort from wearing the CI and can also improve hearing quality.

As FNS can usually be alleviated by remapping the coding strategy and because the incidence of cochlear–facial dehiscence (CFD) is low, surgeons do not recognize the severity of FNS occurring because of CFD. We present a case of severe and persistent FNS triggered by CFD that resulted in poor auditory experience. To the best of our knowledge, this is the first report of this interaction. This case report is presented in accordance with the CARE guidelines.<sup>5</sup>

## Case report

A 36-year-old man complained of hearing loss and tinnitus for 2 months. Magnetic resonance imaging revealed a mass (11.3 mm × 4.6 mm) in the left auditory canal. Pure tone audiometry, shown in Figure 1, indicated mild hearing loss in his left ear and normal hearing in his right ear. The speech discrimination score was 32% for his left ear. To restore bilateral hearing, the patient underwent vestibular schwannoma resection using presigmoid retrolabyrinthine and partial translabyrinthine surgical approaches and simultaneous cochlear implantation (MED-EL, Innsbruck,



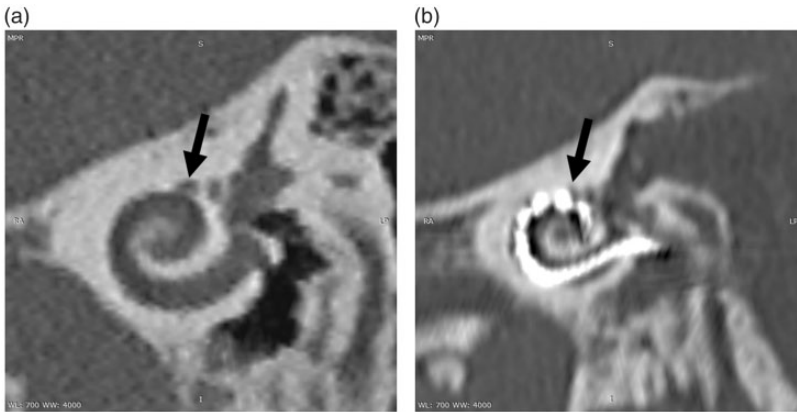
**Figure 1.** Preoperative pure tone audiogram. The blue line represents the left ear, and the red line represents the right ear.

Austria; Flex, 28 mm) in June 2020. Intraoperatively, we found that the tumor originated from the inferior vestibular nerve and that the auditory and facial nerves were completely preserved. The electrode implantation went smoothly, and neural response telemetry (NRT) was successfully recorded. No severe postoperative complications occurred. The preoperative and postoperative images are shown in Figure 2a–d.

One month after the surgery, we activated the CI, and the patient presented with auditory responses at most of the electrodes, but with severe FNS (muscle spasms around the mouth, lips, and eyes) and with no auditory responses in three electrodes (8th, 9th, and 10th) with low stimulation. We changed the stimulus conditions from biphasic to triphasic pulse patterns, increased the pulse width from 50  $\mu$ s to a maximum of 150  $\mu$ s, and adjusted the speech encoding strategy from fine structure processing to high-definition continuous interleaved sampling; however, none of these changes alleviated the patient's FNS symptoms. Finally, we deactivated the electrodes triggering the severe FNS.



**Figure 2.** Pre- and postoperative images. Preoperative computed tomography (CT) images (a and b), preoperative magnetic resonance images (MRI) (c), and postoperative CT images (d). An auditory neuroma is visible in the left internal auditory canal on MRI (white arrow, panel c).



**Figure 3.** Cochlear-facial dehiscence in computed tomography images. Cochlear-facial dehiscence is seen in preoperative (a) and postoperative (b) multiplanar computed tomography reconstruction images. The black arrow represents the labyrinth segment of the facial nerve, without bony separation between the cochlea and labyrinth segment of the facial nerve.

In the following 9 months, the CI programming was remapped three times, as described above; each time, the patient experienced slight relief from FNS and was able to receive slightly higher stimulation compared with the last CI mapping without developing FNS. However, several electrodes (8th, 9th, and 10th) were still not activated because they triggered severe FNS at very low levels of stimulation. In addition, the patient continued to complain of low sound levels on the CI side, making it difficult to establish binaural hearing. The patient believes that wearing the CI helps reduce tinnitus, and the visual analog scale score changed from 8 preoperatively to 6

with the CI working. Therefore, even though there is no improvement in the hearing performance, he is willing to wear the CI all day.

Because the patient's FNS was severe, we reviewed the surgical video and images to elucidate the cause. We eventually found CFD on computed tomography (CT) images (Figure 3a,b).

## Discussion

CFD refers to dehiscence between the cochlea and labyrinth segment of the facial nerve, which was first described in two patients with symptoms of third-window

lesions by Blake et al. in 2014.<sup>6</sup> CFD has been reported in very few cases.<sup>6–10</sup> Fang et al.<sup>9</sup> reported three cases of CFD and cochlear implantation, two of whom presented with FNS and one did not. Both patients with FNS obtained good postoperative hearing rehabilitation results by adjusting the coding strategy. Garaycochea et al.<sup>8</sup> reported a case with two dehiscences in an otic capsule (cochlear–internal canal and CFD). No FNS was found in this patient, who underwent cochlear implantation. A child with bilateral severe to profound sensorineural hearing loss was reported to have bilateral CFD and underwent bilateral cochlear implantation without developing FNS.<sup>7</sup> Camerin et al.<sup>10</sup> reported a case of CFD causing facial nerve palsy in a patient with CIs.

The overall incidence rate of FNS is 5.6%.<sup>2</sup> While the exact mechanism of FNS is yet to be fully elucidated, patients with cochlear anomalies, such as otosclerosis and inner ear malformations, are more likely to have FNS.<sup>2</sup> Patients with otosclerosis may more easily experience FNS, mainly because the impedance between the lateral wall of the cochlea and the facial nerve is reduced owing to changes in the dysplastic otosclerotic bone, and when the CI is working, the electricity released from the electrodes is easily transmitted through the low-impedance area, stimulating the facial nerve and triggering FNS.

In the present case, the mechanism of FNS was very similar to that of otosclerosis. CFD was observed on CT, without the common bony separation between the cochlea and labyrinth segment of the facial nerve; thus, the power released from the CI could be transmitted directly to the facial nerve to trigger FNS. Furthermore, CT suggested that CFD approximately corresponded to the 8th and 9th electrodes, which induced FNS at a lower stimulation, indicating that FNS in this patient was associated with CFD. However, the patient

had a history of vestibular schwannoma, which may have led to auditory nerve dysfunction and usually requires higher stimulation to obtain a better auditory experience. NRT was successfully recorded, implying that the auditory nerve was intact. Therefore, we believe that the poor outcome in this patient was mainly owing to low stimulation because of FNS.

To date, six patients with CI and CFD have been reported; two presented with FNS, which resolved after adjusting the coding strategy, one developed facial nerve palsy 7 days after surgery, and the remaining three did not experience FNS or facial nerve palsy. None of these patients experienced persistent and unremitting FNS. However, in our case, we remapped the coding strategy by decreasing stimulation levels, widening the pulse width, and deactivating electrodes (8th, 9th, 10th electrodes), which were ineffective. We hypothesize that FNS is related not only to the level of stimulations but also to the width of cochlear–facial nerve partition, which could explain to some extent why this patient experienced such severe and persistent FNS.

Our patient experienced slight relief from FNS after each adjustment and was able to receive slightly higher stimulation compared with the previous adjustment, without developing FNS. We hypothesized that the CFD area undergoes organization and wrapping over time, which leads to an increase in impedance at the corresponding location such that the FNS is slightly relieved. Although the patient's FNS symptoms were slightly relieved in this case, longer follow-up and careful mapping are required to verify whether the patient can successfully achieve binaural hearing.

The prevalence of CFD was 0.59% in 1020 temporal bone specimens;<sup>11</sup> however, the prevalence of radiographic CFD was 1.4% to 5.4%.<sup>12,13</sup> Because of the difficulty detecting CFD in the horizontal plane, the

low incidence of CFD, and its unknown occurrence in patients with CI, the clinical surgeon did not consider CFD during the preoperative evaluation, in this case. However, our experience tells us that during the preoperative evaluation of patients with CI, surgeons must consider that patients with normal cochlear structures may still have facial nerve aberrations or CFD and may develop severe and persistent FNS that could affect the CI outcomes.

Although CIs may cause discomfort, such as FNS, vertigo, and headaches, post-operatively, CIs effectively restore hearing in patients with severe and profound sensorineural hearing loss. The incidence of FNS after cochlear implantation is 5.6%.<sup>2</sup> However, the incidence of a serious impact on CI outcomes owing to FNS is extremely low, and in most cases, FNS can be alleviated by adjusting the CI mapping strategy. For patients with bilateral severe and profound sensorineural hearing loss, we should choose the side without CFD for cochlear implantation. If both sides have CFD and if cochlear implantation is necessary, we should try to choose perimodiolar/mid-scalar electrodes, as suggested by Kaufman et al.<sup>14</sup> Then, depending on the patient's clinical presentation, different mapping strategies should be adopted to relieve FNS, such as decreasing the stimulation levels, widening the pulse width, and deactivating the electrodes.

## Conclusion

Our experience suggests that a careful review of CT images before CI surgery is necessary, especially for patients with otosclerosis, inner ear malformations, and even patients with normal cochlear structures, because facial nerve anomalies may be present. CFD may lead to severe and persistent FNS, which can affect the CI outcomes. In preoperative evaluations for patients with

CI, emphasis should be placed on examinations for the presence of CFD.

## Ethics statement

Ethical approval was provided by the Medical Committee of Beijing Tongren Hospital, Capital Medical University (TRECKY-2019-055-XZ-1). Written informed consent was obtained from the patient and his family.

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## Declaration of conflicting interest

The authors declare that there is no conflict of interest.

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