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Case Report

A case of cochlear-facial dehiscence revealed after bilateral cochlear implants ☆

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ABSTRACT

The cochlear implant is an implanted auditory prosthesis that can restore severe and profound hearing loss. About 20% of patients with congenital sensorineural hearing loss have a malformation of the inner ear. These abnormalities must be investigated before a cochlear implant because they can lead to intra and postoperative complications and/or anomalies. Most labyrinthine malformations are well known; some are less frequent and can be underdiagnosed at the preoperative computed tomography. This report presents the case of bilateral cochlear-facial dehiscence, bony dehiscence between the facial nerve labyrinthine segment, and cochlear basal turn. In our 56-year-old patient, this malformation was misdiagnosed before the cochlear implant and revealed afterward because of abnormal facial nerve stimulation during intraoperative electrophysiological checking.

Introduction

The Cochlear implant (CI) is an auditory prosthesis that can restore severe and profound hearing loss, and has been used for several decades. About 20% of patients with congenital perceptive hearing loss have a malformation of the inner ear [1]. These malformations were described and classified [1,2] in the literature. Some of them can lead to clinical consequences in the case of CI, such as the cochlear-facial dehiscence (CFD): the bony dehiscence between the facial nerve labyrinthine segment and cochlear basal turn. Bigelow et al. [3] first suspected the anomaly of the wall in between the labyrinthine segment of the facial nerve and the cochlea to be the cause of stimulation of the facial nerve after CI. There is a paucity of studies outlining the detailed imaging appearance of CFD in contemporary literature, usually described as the absence or discontinuity of bony covering [4], and some CFD can be misdiagnosed at the preoperative CT. The major limit of CT is that

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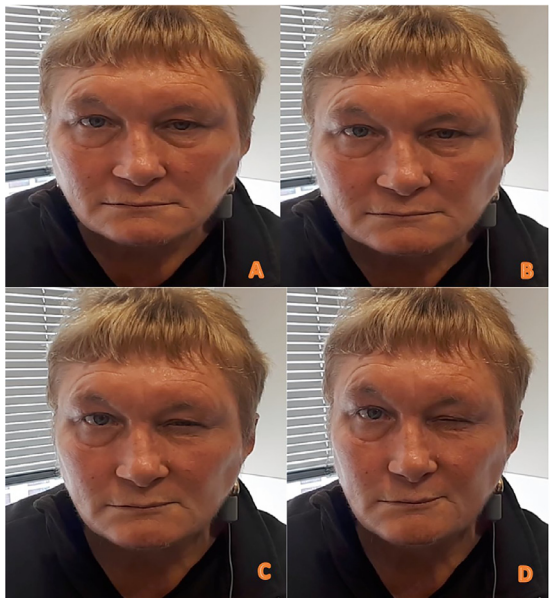


Fig. 1a – Stimulation of left facial nerve during stimulation of electrode 13. (A) at rest. (B) Stimulation at 15 dB HL. Slight contraction of the lower part of the orbicularis muscle of the left eye and the lower part of the orbicularis muscle of the mouth. (C) Stimulation at 17 dB HL. Almost complete left eyelid occlusion and movement of the left labial commissure. (D) Stimulation at 19 dB HL. Complete left eyelid occlusion and marked movement of the left labial commissure. This problem was solved by the deactivation of 2 electrodes.

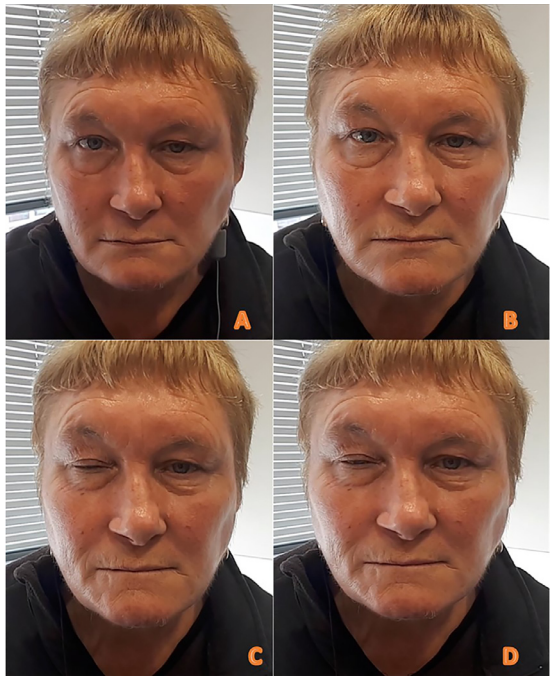


Fig. 1b – Stimulation of right facial nerve during stimulation of electrode 13. (A) At rest. (B) Stimulation at 20 dB HL. Movement of the corrugator muscle of the right eyebrow. (C) Stimulation at 25 dB HL. Almost complete palpebral occlusion and movement of the right labial commissure. (D) Stimulation at 30 dB HL. Complete lid occlusion and clear movement of the right labial commissure.

the bony coating of the facial canal is thin, often surpassing the spatial resolution of multislices CT.

This report presents the case of a bilateral CFD diagnosed after bilateral CI in a 56-year-old female patient and discusses the aetiological, physiological, and electrical mechanisms.

Case report

A 56-year-old patient with a history of polycythemia, respiratory and renal failure, was referred some years ago to our department for left CI. She had been wearing bilateral external air conduction hearing aids for 13 years, however, she had never developed a good oral language and had only acquired a limited lexical stock. She had a progressive sensorineural hearing loss of unknown etiology. The patient did not have pulsatile tinnitus, autophony, otalgia, otorrhea, vertigo, or imbalance. The otoscopic examination was normal bilaterally and she had a normal facial motility. Pure tone audiometry revealed bilateral profound sensorineural hearing loss with complete loss of higher frequencies above 500 Hz. Only poor discrimination was possible in between monosyllabic and disyllabic lists in the free field, under the best possible amplification conditions, with hearing aids on. The patient's auditory performance, assessed using the category of auditory performance (CAP) scale, was 4/7 (possible discrimination of a few speech sounds without lip reading). The vestibular evoked myogenic potentials (VEMPs) were present on each side at 100 dB. The computed tomography (CT) of the temporal bones (Discovery CT750 HD, 0.625mm slices) was interpreted as normal. The DIGISONIC SP CI (Oticon Medical) was placed on the left side under facial nerve monitoring (NIM Response, Medtronic, Minneapolis, MN). A surgical procedure was performed in the angio-room (Artis Zeego, by Siemens) in the interventional radiology department, in order to have real-time 2D fluoroscopy guidance to improve the quality of electrode-array insertion [5]. Immediately after the procedure, facial nerve monitoring was switched off, and then the electrode-array impedances were checked. Electrical evoked compound action potentials (eCAP) on electrodes 7, 13, and 17 were recorded with a coherent shape. At the end of the procedure, a 3D cone-beam CT acquisition was performed with a standard dedicated neuroprotocol that showed good winding of the electrode-array. Nevertheless, during CI activation, the technician objectified a sign of stimulation of the left facial nerve (left eye shut) during the stimulation of electrode 13 (Fig. 1A); this electrode was disabled without affecting the patient's quality of life. Three years later the patient developed a degradation of hearing on the right side. The patient's auditory performance, assessed using the CAP scale, was 5/7 (understanding common sentences possible without lip reading), slightly better than before CI, without the benefit of contralateral hearing aid. A NeuroZTI-CLA CI (Oticon Medical, Val-

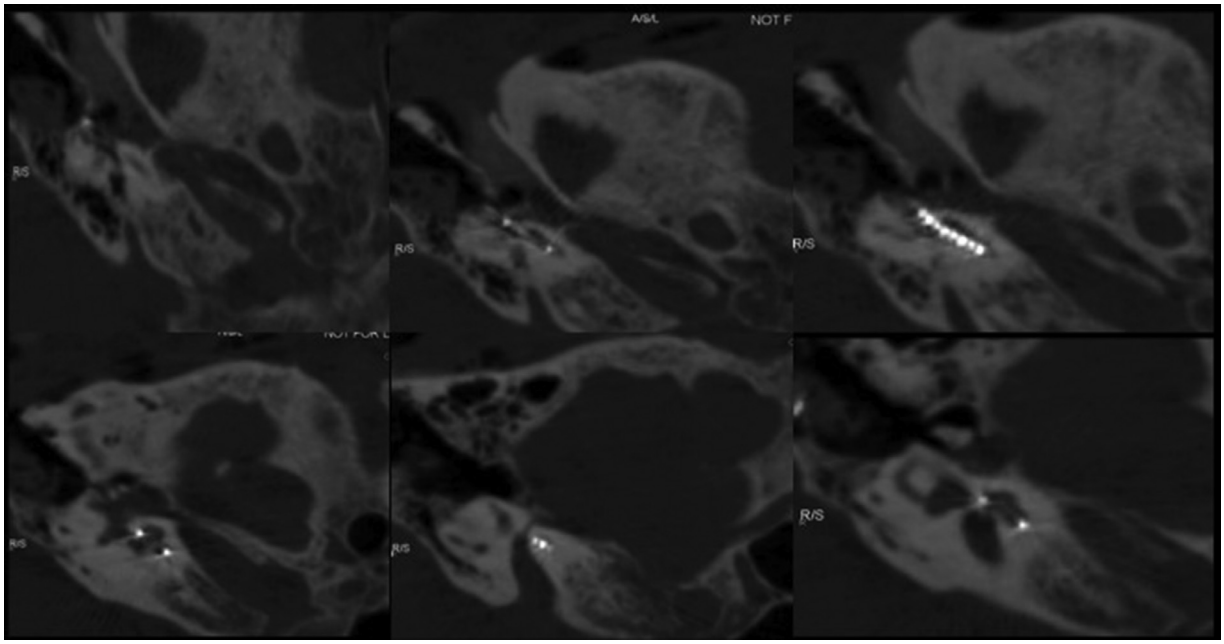


Fig. 2 – Intraoperative fluoroscopy images. Intraoperative fluoroscopy images during CI positioning.

Table 1 – Summary of intensities (comfort intensities = maximum intensities) at the origin of contraction of the skin muscles of the face, by stimulated electrode on the left.

Left ear	Electrode 13	Electrode 12	Electrode 14
Light movement of the ipsilateral skin muscles	15 dB HL (lower part of the orbicularis muscle of the left eye)	45 dB HL (left orbicularis muscle of the mouth)	27 dB HL (muscles of the left angle of the mouth)
Marked palpebral movements and labial rictus	19 dB HL	55 dB HL	33 dB HL

Table 2 – Summary of intensities (comfort intensities = maximum intensities) at the origin of contraction of the skin muscles of the face, by stimulated electrode on the right.

Right ear	Electrode 13	Electrode 14
Light movement of the ipsilateral skin muscles	20 dB HL (movement of the corrugator muscle of the right eyebrow)	20 dB HL (right mouth angle depressor movement)
Marked palpebral movements and labial rictus	30 dB HL	35 dB HL

lauris, France) was placed on the right side, following the same procedure performed on the left one, under facial nerve monitoring and 2D-fluoroscopic control. Intraoperative fluoroscopy images and postoperative 3D cone-beam CT showed a satisfactory positioning of the CI (Figs. 2 and 3). The impedances were tested immediately after the surgery and showed normal responses while, the facial nerve monitoring was still active. In the immediate postoperative, the stimulation of all electrodes one by one, to check the electrode impedance and the nervous response of the implant by eCAPs, provoked clinical and electrical right facial nerve response.

Retrospectively a review of the preoperative CT images revealed the Fallopian aqueduct to be very close to the cochlea bilaterally, without any bony separation between the cochlea and labyrinth segment of the facial nerve (Figs. 4A and B, Fig. 5). This bilateral dehiscence of the cochlear wall is responsible for the stimulation of the left facial nerve (left eye shut) revealed by the technician after the left CI and for the right facial nerve response after right CI. There was no cochlear hypoplasia or associated semicircular canal abnormalities. During the activation of the right CI, at the first postoperative CI fitting, a response of the facial nerve was detected during

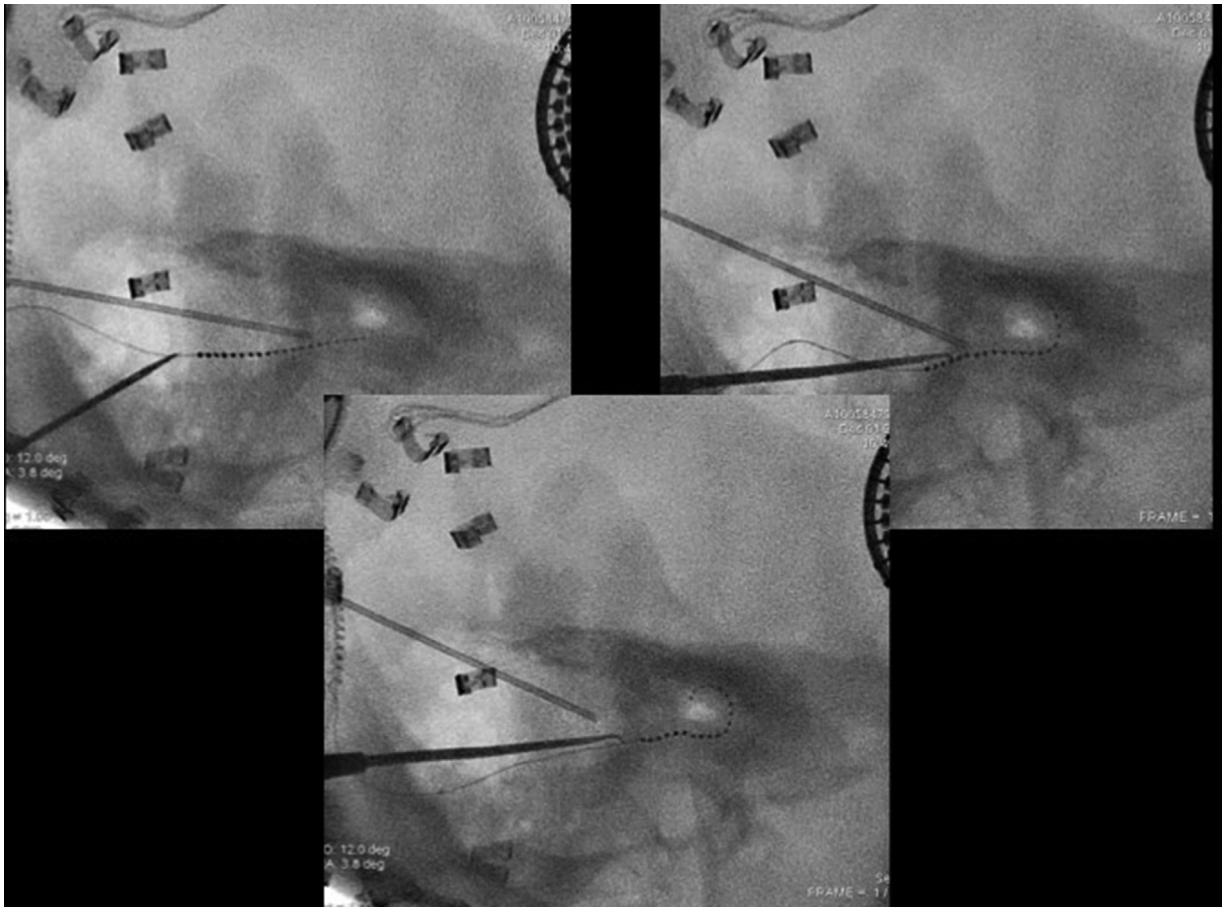


Fig. 3 – 3D cone beam CT. Images show satisfactory positioning of the implant on the right side.

stimulation of electrode 13, leading to its successful deactivation (Fig. 1B). Eventually, electrodes 12 and 14 on the left (Table 1) and electrode 14 on the right (Table 2) were deactivated because of undesirable facial nerve stimulation. At the last follow-up, 3.5 years after the left CI and 4 months after the right one, the patient was satisfied; she had a mean hearing threshold on pure tone audiometry at 30 dB, with a 50% speech discrimination score without lip-reading (SDS), at 60 dB (dissyllabic-word list) versus 20% dissyllabic-word list (SDS) on preoperative period. The monosyllabic-world-list SDS remained at 10% (Fig. 6).

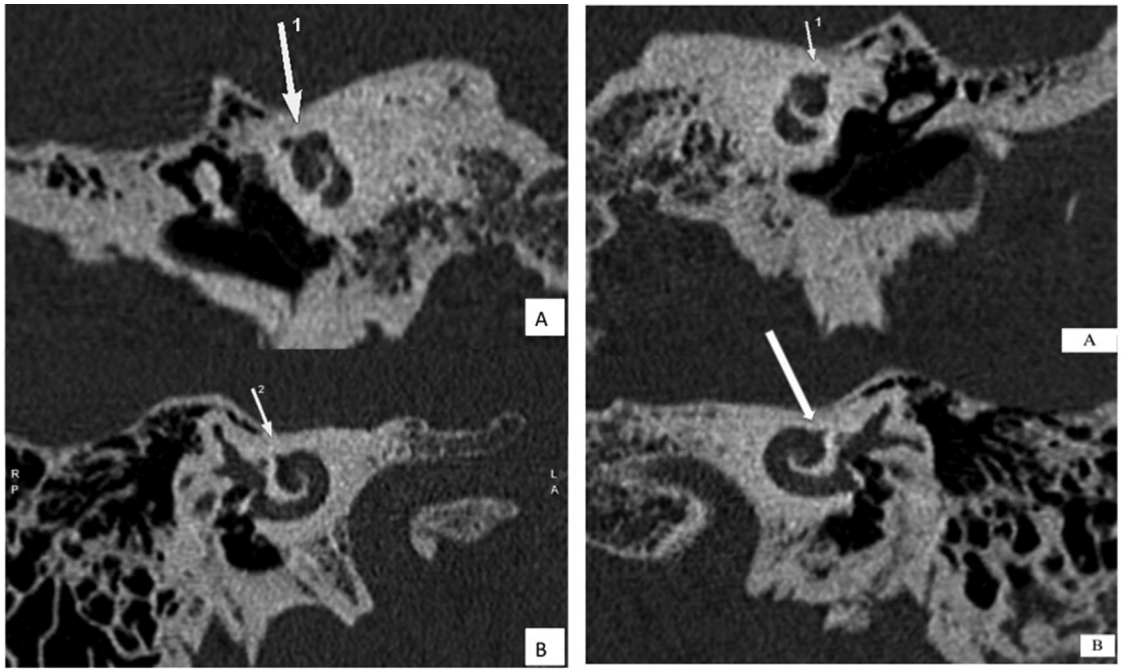
Discussion

A broad spectrum of malformation of the inner ear has been described in the literature [1,2]. Some of them can lead to clinical consequences in the case of CI, such as CFD. In our patient, the electrodes implicated in the stimulation of the facial nerve are, bilaterally, those located in the middle of the electrode array that is closest to the labyrinthine segment of the facial canal and therefore more likely to be involved in facial nerve stimulation by transmitting electrical stimuli. A significant proximity, on both sides, of the electrodes 8 to 11 of the electrode-array with the labyrinthine segment of the facial nerve has been described in the literature [3]. Although we found no studies on OTICON Medical CIs the architecture is likely to be similar, since in our case, facial nerve stimulation appears later for electrodes 12 and 14 on the left and for electrode 14 on the right (for electrode 12 a facial stimulation takes place with higher intensities).

In 2014, Blake described the CFD as a bony dehiscence between the facial nerve and cochlea at the CT imaging [4].

Fang et al. [6] conducted a descriptive study of 1,020 archived temporal bone specimens examined for CFD. Cochlear-facial partition width (CFPW) and otic capsule area (OCA), a marker of bone thickness, were measured using an image analysis software. The mean CFPW was 0.23 mm (range 0-0.92 mm; SD 0.15 mm). Six patients were completely dehiscent (0.59%). The mean OCA for dehiscent specimens (mean, 9.48 mm; range, 6.65-11.58 mm; SD 3.21 mm) was significantly smaller than the mean OCA for nondehiscent specimens, (mean 12.88 mm; range 6.63-21.92 mm; SD 2.47 mm) ($P < .01$). CFD occurred in nearly 0.6% of specimens in this temporal bone collection.

Schart-Moren et al. [7], by analyzing microdissection of 282 temporal bones, described the same year the presence of a dehiscence in 1.4% of cases. On the other hand, in 2017, a revision of CT images revealed 5.4% dehiscence in 406 ears; this rate



Figs. 4 – a and b: CT images revealing a bilateral CFD.
a: right side. (A) Coronal sections of the right petrous bone: visualization of the dehiscence of the first portion of the canal of the facial nerve, next to the cochlea (arrow). (B) Oblique view of the right petrous bone: visualization of the dehiscence of the first portion of the canal of the facial nerve, next to the cochlea (arrow).
b: Left side. (A) Coronal sections of the left petrous bone: visualization of the dehiscence of the first portion of the canal of the facial nerve, next to the cochlea (arrow). (B) Oblique view of the left petrous bone: visualization of the dehiscence of the first portion of the canal of the facial nerve, next to the cochlea (arrow).

of radiographic cochlea-facial bony wall dehiscence is higher than that reported in histological studies and may overestimate the prevalence of real dehiscence [8].

Taking intraoperative microscopic findings as a gold standard, in 2017 some authors assessed the diagnostic accuracy of preoperative CT and intraoperative neurophysiological monitoring for the detection of CFD. They found a high concordance between neurophysiological findings and microscopic observations during microsurgery. Regarding preoperative CT, they found low concordance with intraoperative findings, unlike other authors who reported high sensitivity and good concordance. In their series, the intraoperative neurophysiological test showed very high sensitivity (99%) for the detection of CFD in primary surgeries. CT showed a sensitivity of 64.7%, and specificity of 78.4% [9].

CT appearance of CFD and the diagnostic accuracy still remain undetermined. To date, several attempts have been made to describe CT findings. Tanrivermi et al. [10] defined CFD as the discontinuity of the bony structure, presenting as a direct connection between the nerve and the middle ear space. Arias-Marzán et al. [9] considered CFD as an interruption of the bony coating in both coronal and axial planes. The major limit of CT is that the bony coating of the facial canal is thin, often surpassing the spatial resolution of multislices CT. With a slice thickness of 1 mm, the concordance between imaging diagnosis and surgical findings ranged from 42% to 88.2%. The discrepancy in diagnostic values can be explained by different settings of CT devices or undetermined CT appearance, but is more likely attributable to the thinness of the bony covering. In the future CT devices with higher spatial resolution may provide a more in-depth characterization of the radiological aspect of CFD. A recently developed ultra-high-resolution computed tomography (U-HRCT), with a spatial resolution of 0.1 mm, may be helpful in detecting the presence of CFD. Studies have demonstrated the capability of U-HRCT in delineating fine structures of the temporal bone, both in cadavers and in patients with otologic diseases. Based on observation from U-HRCT images, recent studies introduced 2 new different entities: discontinuous bony covering with linear deficiency versus discontinuous bony covering with dotted deficiency (instead of the binary classification of imaging: lack of bony coating versus continuous bony covering) [11]. In the future, the U-HRCT would improve the preoperative CT evaluation that plays a key role in preventing iatrogenic injury or complication in CI.

In consideration of the actual limits of the imaging, some authors suggest an interest in the intraoperative recording of electrical evoked auditory brainstem responses (e-ABRs). In their case report, the authors identified in 2 patients with CFD, a late myogenic potential at low levels of stimulation during e-ABRs [7]. In our case, facial nerve stimulation was immediately detected, during impedance checking, thanks to the facial nerve monitoring that had not been turned off. We did not record e-ABRs, but facial nerve stimulation was nevertheless

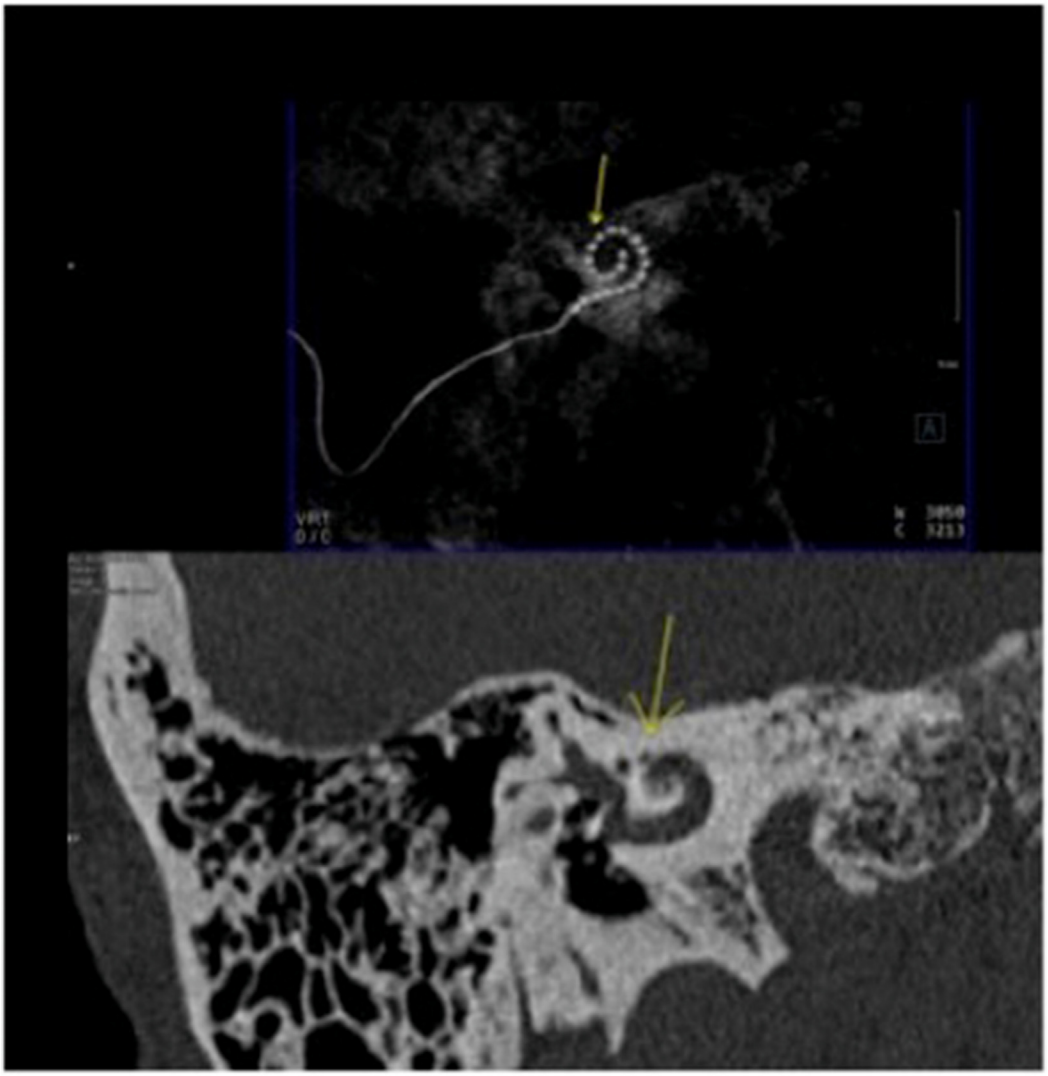


Fig. 5 – CT image and 3D cone beam CT postprocedure. Comparison between the pre-operative CT, coronal view, of the right petrous bone and the postoperative 3DCT reconstruction image: visualization of the dehiscence of the first portion of the facial nerve canal, facing the electrode 13 (arrows).

detected. We thus recommend keeping facial nerve monitor turned on at the end of surgery during impedance checking, and eCAPs recording.

One aspect remains unclear: why did the stimulation of almost all the electrodes of the right CI cause facial stimulation during intraoperative e-CAP recordings, but only electrodes 12 and 13 during the initial activation of the device? It is likely that electrical energy used in clinical practice for CI activation is far less important than that used for collecting e-CAP intraoperatively.

Smullen et al. [12] found that in 11 out of 44 patients (35%), the onset of facial stimulation occurred more than 12 months after initial activation. Other patients experienced a gradual increase in the number of electrodes causing facial nerve stimulation. Several hypotheses have been put forward to explain this delayed phenomenon: erosion of the bony wall between the facial nerve and the scala tympani under the pressure of electrode [3], change in current path, tissue impedance, or facial nerve tenderness.

Song et al. [13] suggest that CFD may be due to acquired causes such as trauma, infection, or bone erosion induced by intracranial pressure. These mechanisms are not completely comparable because the otic capsule separating the facial canal from the cochlea is embedded in the temporal bone and is not contiguous to the intracranial fossae where dural pulsations or hypertension could contribute to progressive bone erosion. Furthermore, bone resorption has been mentioned secondary to aging [14]. On the other hand, this entity can be congenital, caused by impaired development of the otic capsule [15]. In our patient, the CFD was bilateral, suggesting a possible congenital component in the etiopathogenesis; the fact that the cochlea and the vestibule show no malforma-

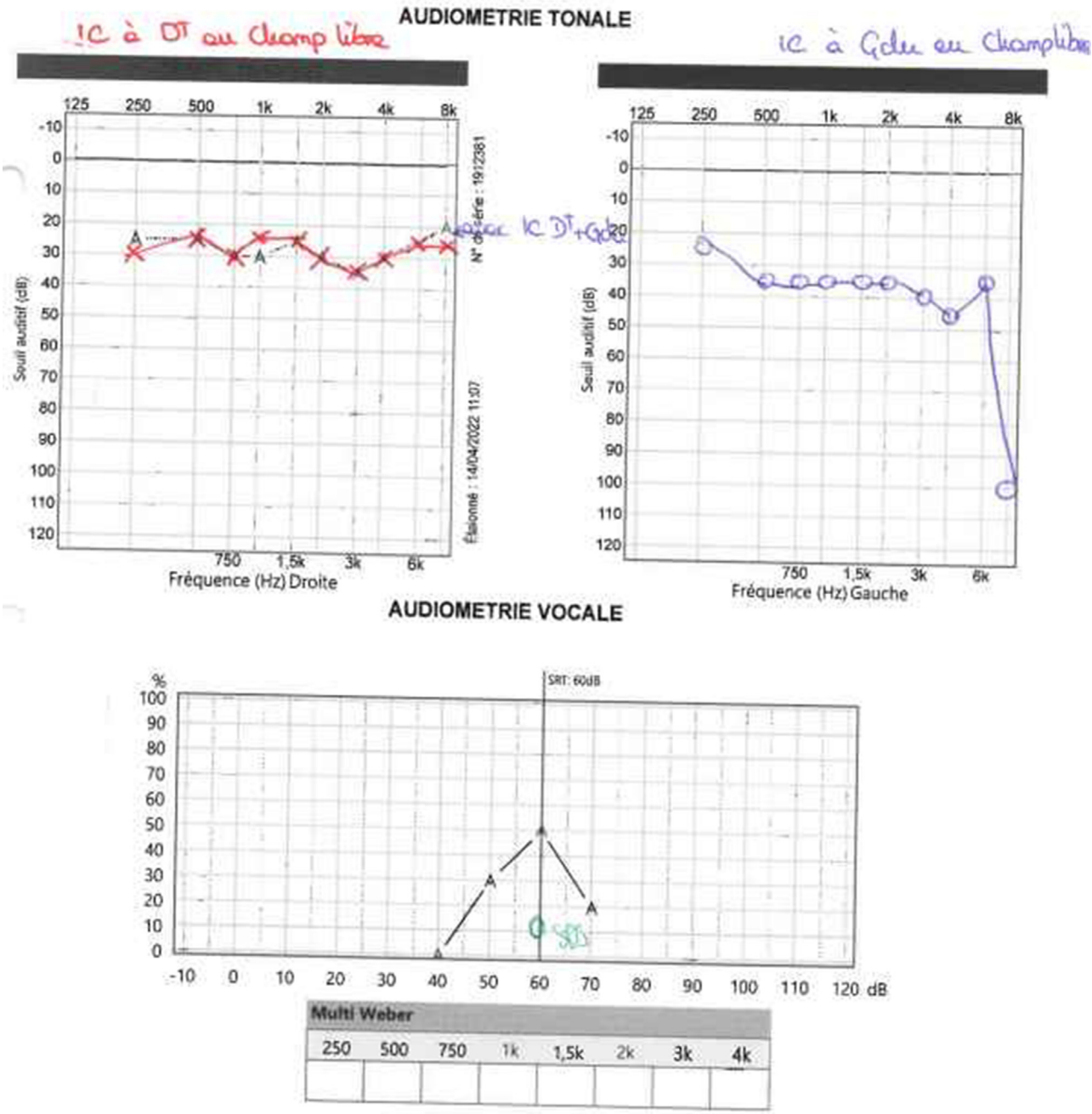


Fig. 6 – Last follow-up audiometry. As described in the text, despite of facial nerve stimulation by the CI, it was possible to adapt the strategy and intensity of the stimulation to provide good results at the follow-up with no more facial nerve stimulation.

tions suggests that the defect develops at a later stage than the otocyst stage [1] and the partial absence of bone suggests failure of ossification of the otic capsule [16].

The problem of this undesirable facial nerve stimulation can be solved in different ways: by reducing the intensities up to the level of sound perception that does not trigger facial stimulation, or by switching off some electrodes. In our case, the second option was applied with very satisfactory hearing results.

Conclusion

The CFD is an entity to look for before CI, like any other malformation. The diagnosis of CFD on preoperative CT is often a challenge even for a knowledgeable radiologist. A suspected misdiagnosed CFD can be confirmed by keeping the facial nerve monitoring during CI electrophysiological checking at the end of the surgical procedure.

Patient consent

Appropriate consents, permissions, and releases have been obtained from the patient.

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