Rev. Otorrinolaringol. Cir. Cabeza Cuello 2024; 84: 68-71

CLINICAL CASE

doi: 10.4067/S0718-48162024000100110

# Cochleo-carotid dehiscence as a third window syndrome: a review of an unusual presentation and its clinical-radiological approach

## Dehiscencia cócleo-carotídea como síndrome de la tercera ventana: una revisión de una presentación inusual y su enfoque clínico-radiológico

*Joan Lorente-Piera[1], Raquel Manrique-Huarte[1], Natalia Díaz-Zufiaurre[1], Pablo Domínguez-Echávarri[2], Manuel Manrique-Rodríguez[1]*

[1]Department of Otorhinolaryngology, University Clinic of Navarra, Pamplona, Spain.
[2]Department of Radiology, University Clinic of Navarra, Pamplona, Spain.

The authors declare no conflicts of interest.

Received 05 September 2023.
Accepted 20 December 2023.

Corresponding author:
Joan Lorente-Piera
Department of Otorhinolaryngology, Clínica Universidad de Navarra, Pamplona, Spain.
Email: jlorentep@unav.es

### Abstract

When examining patients who present both fluctuating auditory symptoms and vestibular symptoms, which can be mistaken for an ear disease, it is important to consider cochlear-carotid fistula as a condition that could generate these clinical manifestations. In this case, additional imaging tests should be performed to demonstrate the lack of continuity at that level and rule out other entities. The objective is to describe the cochleo-carotid dehiscence in a 39-year-old woman and propose it as a possible etiology of a third window syndrome, in order to discuss the relationship between clinical findings, neurotologic examination, and magnetic resonance imaging.
**Keywords**: Cochleo-carotid dehiscence, vestibular, third window, cochlear implant.

### Resumen

*Al examinar a pacientes que presentan síntomas auditivos fluctuantes y síntomas vestibulares, que pueden ser confundidos con una enfermedad del oído, es importante considerar la fístula coclear-carotídea como una condición que podría generar estas manifestaciones clínicas. En este caso, se deben realizar pruebas de imagen adicionales para demostrar la falta de continuidad en ese nivel y descartar otras entidades. El objetivo es describir la dehiscencia cocleo-carotídea en una mujer de 39 años y proponerla como una posible etiología del síndrome de la tercera ventana, con el fin de discutir la relación entre los hallazgos clínicos, el examen neurotológico y la resonancia magnética.*
***Palabras clave:*** *Dehiscencia cocleo-carotídea, vestibular, tercera ventana, implante coclear.*

### Introduction

Cochleo-carotid dehiscence is a condition with a virtually residual incidence[1], but of high importance due to the wide range of symptoms it can present, necessitating consideration of various entities in the differential diagnosis. Additionally, it is relevant to consider this anomaly when planning cochlear implant surgery[2], as it is a critical anatomical factor for the success of the surgery or its possible intraoperative complications.

The objective of this study was to describe the cochleo-carotid dehiscence in a 39-year-old woman and propose it as a possible etiology of a third window syndrome, in order to discuss the relationship between clinical findings, neurotologic examination, and magnetic resonance imaging.



ISSN 0718-4816 versión en línea - ISSN 0716-4084 versión impresa

## Clinical Case

A 39-year-old woman presented to the specialist reporting an exacerbation of her sensation of instability, lasting for hours, without associated vegetative symptoms or a sensation of objects spinning with head movements over the past year. She also experienced a recently onset tinnitus in the left ear, with a high-pitched, fluctuating, non-pulsatile character, predominantly in the evenings, and with moderate limitations in basic activities of daily life. Additionally, she described progressive hearing loss in the left ear lasting over ten years, which did not worsen or coincide with her episodes of instability and was not compatible with *Tullio phenomenon*. Otoscopic examination revealed relevant findings only in the left ear, with an anteroinferior monomeric area and type II atelectasis in the flaccid pars. In terms of otoneurological examination, the only notable finding was the presence of right horizontal nystagmus that appeared during cervical flexion and right lateral decubitus, occurring synchronously with the cardiac cycle. As for complementary studies, pure-tone audiometry (**Figure 1A**) confirmed the presence of moderate-grade conductive hearing loss, more pronounced in low frequencies, with a type C2 tympanogram in that ear (**Figure AB**) and absent stapedial reflex in both ears. Vestibular tests (vestibulo-ocular reflex) did not reveal pathological results, showing normal functioning gains in all planes.

Given these findings, a CT evaluation of the petrous bones is requested (Siemens

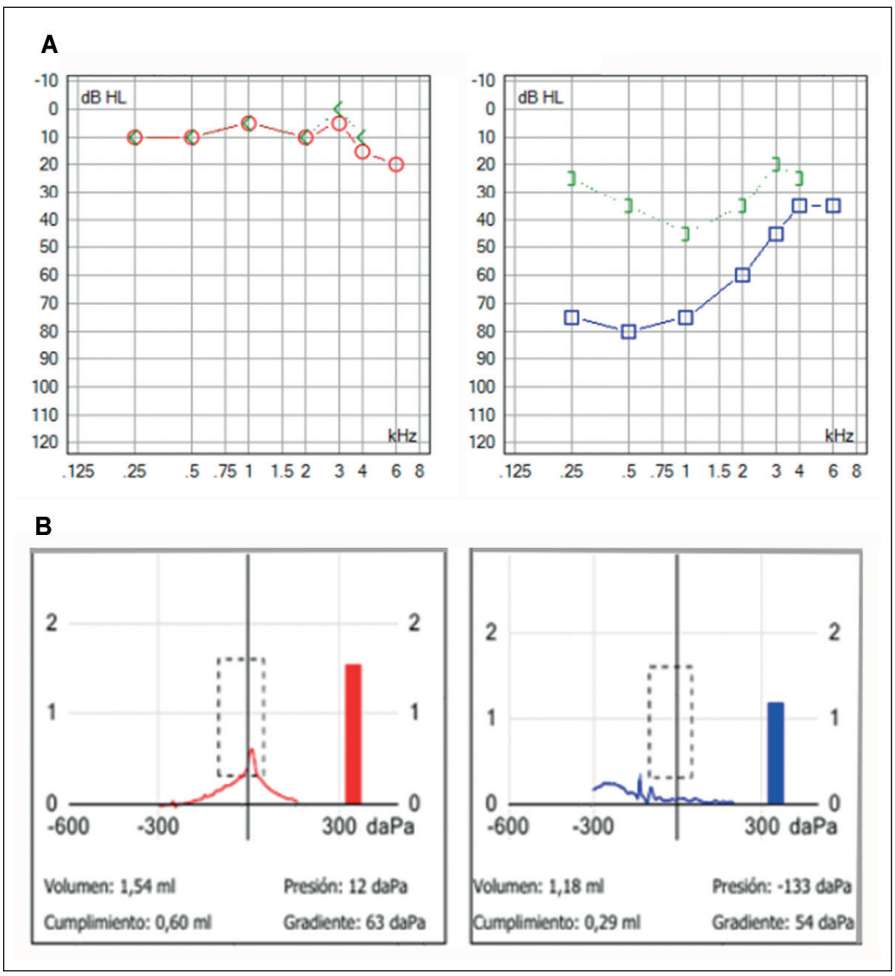


**Figure 1. A:** Tonal audiometry showing moderate-grade conductive hearing loss in the left ear according to *ASHA* criteria. **B:** Type A tympanogram in the right ear and type C2 tympanogram in the left ear.

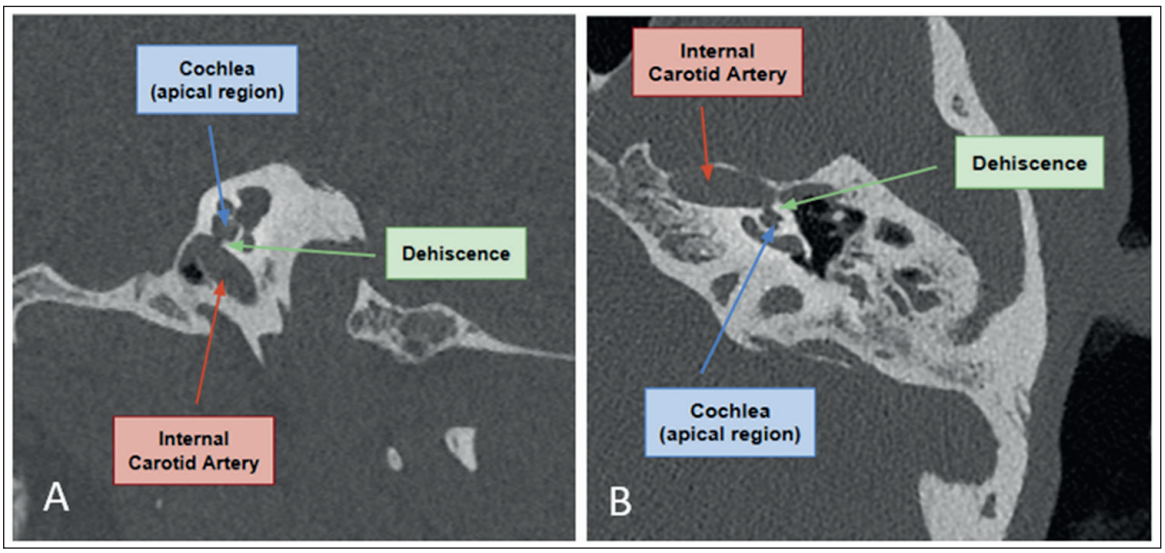


**Figure 2.** Computed tomography image of the left petrous bone in the sagittal plane **(A)** and axial plane **(B)**. Cochleo-carotid dehiscence is observed (green), with a lack of bony coverage between the apical region of the cochlea (blue) and the carotid canal (red).

Healthineers Somatom Force equipment, with main parameters of 120 kV, 230 mAs max, with a slice thickness of 0.4 mm every 0.2 mm, resolution of 512x512 pixels for a field of view of 70x70 mm, and a final voxel size of 0.13x0.13x0.4 mm). The evaluation of the right ear is normal. In the left ear, there are no signs of otosclerosis, but there is a notable absence of bony coverage between the cochlear apical region and the carotid canal, consistent with cochleo-carotid dehiscence (**Figure 2**). No dehiscence is observed in other locations, nor is there vestibular aqueduct dilatation. Minimal opacification of the middle ear is identified without suggestive images of cholesteatoma.

## Discussion

In the initial suspicion of a possible case of otosclerosis, based on the clinical concordance with this condition and, especially, the obtained audiometric pattern, a petrous bone CT scan was performed. The imaging test not only ruled out signs of otosclerosis in the otic capsule or other temporal bone otodystrophies but also led to the diagnosis of a left apical cochleo-carotid dehiscence, without other associated labyrinth malformations.

Cochleo-carotid dehiscence is defined[1] as the thinning or absence of the bony lamina separating the intratemporal portion of the internal carotid artery from the cochlea, with the normal[2] measurement being between 0.2-0.5 mm. Studies such as Moreano et al.[3], which analyzed up to 1000 temporal bones and found dehiscence at this level in only 7 cases, or others like Gunbey et al., which studied up to 2100 temporal bones and found this finding in only 10 of them, with 80% of cases being unilateral and the remaining 20% involving both ears, reaffirm the low incidence of this condition, which often goes unnoticed due to its usually mild clinical presentation. However, it is important to consider it as a possible differential diagnosis[3] in patients who present with tinnitus, which may or may not be pulsatile, along with typically conductive or mixed hearing loss. At the vestibular level, it can cause a sensation of fluctuating instability, which often intensifies with noise *(Tullio phenomenon)* or changes in pressure in the external auditory canal *(Hennebert sign)*[4].

In the presence of a presentation like the attached case, it is important to conduct an exhaustive differential diagnosis with the various pathologies that form the hypothesis of the third window (oval, round, and anomalous windows) proposed to explain the

decrease in sound conduction through the air pathway due to hypersensitivity of the bone pathway. This results in an air-bone gap or difference in hearing conduction of 20 to 60 dB, as seen in our case. Examples of pathologies associated with the third window phenomenon include semicircular canal dehiscences, vestibular aqueduct dilatation, incomplete partitions of the cochlea, bone dyscrasias such as Paget's disease, and even perilymphatic fistulas. There is no clear evidence as to why these alterations manifest with such varied clinical presentations. In some cases, they may be asymptomatic, and there are even cases of bilateral involvement that only produce symptoms in one ear.

Initial treatment in these cases is usually conservative. In cases where the symptoms become incapacitating[5], consideration may be given to surgical procedures to seal the underlying communication causing the symptoms. The proposed approaches for closure are transmastoid or via the middle cranial fossa, aiming to cover the discontinuity or dehiscence with a bone graft, temporal muscle fascia, or bone wax. In our case, a decision was made to adopt an active surveillance approach. Firstly, due to the high risk of carotid exposure, and later, due to the lack of evidence suggesting that sealing the round window to alleviate vestibular symptoms would not worsen the patient's audiometric results. This decision was based on the fact that the patient already had moderate-to-severe hearing loss in the affected ear.

## Conclusion

Cochleo-carotid dehiscence is an extremely rare condition but should be considered in the differential diagnosis of audiovestibular clinical entities. Diagnostic confirmation through imaging tests is essential. Although there are multiple therapeutic options available for symptomatic cases, expectant or conservative management is often pursued.

### Informed consent

Written informed consent was obtained from the patient involved in this case.

## Bibliography

1. Koochakzadeh S, Dornhoffer JR, Horton JD, Meyer TA. Carotid-cochlear dehiscence: a dangerous mimicker of inner ear pathologies. *BMJ Case Rep*. 2019;12(7):229773.
2. Wackym PA, Mackay-Promitas HT, Demirel S, Gianoli GJ, Gizzi MS, Carter DM, et al. Comorbidities confounding the outcomes of surgery for third window syndrome: Outlier analysis: Third Window Syndrome Outlier Analysis. *Laryngoscope Investig Otolaryngol.*
3. Iversen MM, Rabbitt RD. Biomechanics of third window syndrome. *Front Neurol* 2020; 11
4. Wackym PA, Agrawal Y, Ikezono T, Balaban CD. Editorial: Third window syndrome. *Front Neurol.* 2021;12:704095.
5. Wackym PA, Balaban CD, Zhang P, Siker DA, Hundal JS. Third window syndrome: Surgical management of cochlea-facial nerve dehiscence. *Front Neurol.* 2019;10:1281