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Prevalence of Cochlear-Facial and Other Non-Superior Semicircular Canal Third Window Dehiscence on High-Resolution Temporal Bone CT

Vladislav Razskazovskiy, Andrew A. McCall, and Barton F. Branstetter



ABSTRACT

BACKGROUND AND PURPOSE: The radiologic prevalence of superior semicircular canal dehiscence in the asymptomatic population has been widely studied, but less is known about the rates of other forms of third window dehiscence. Per the existing literature, the radiologic prevalence of cochlear-facial nerve dehiscence, for example, exceeds that seen in histologic studies, suggesting that conventional CT is unreliable for cochlear-facial dehiscence. These studies relied on nonisometric CT acquisitions, however, and underused multiplanar reformatting techniques, leading to false-positive findings. Our purpose was to determine the rate of cochlear-facial dehiscence and other non-superior semicircular canal third window dehiscences on optimized CT in asymptomatic patients.

MATERIALS AND METHODS: Sixty-four-channel temporal bone CT scans from 602 patients in emergency departments were assessed for cochlear-facial and other non-superior semicircular canal third window dehiscences by using high-resolution, multiplanar oblique reformats. Confidence intervals for dehiscence prevalence were calculated using the Newcombe 95% interval confidence method.

RESULTS: Of 602 patients, 500 were asymptomatic, while 102 had an imaging indication consistent with possible third window syndrome (symptomatic). Eight asymptomatic patients (1.6%) had cochlear-facial dehiscence, while 43 (8.4%) had jugular bulb-vestibular aqueduct dehiscence. There was no statistically significant difference between the prevalence of cochlear-facial dehiscence or jugular bulb-vestibular aqueduct dehiscence in asymptomatic patients compared with symptomatic patients. Cochlear-carotid canal, cochlear-internal auditory canal, and cochlear-petrosal sinus dehiscences were not observed.

CONCLUSIONS: Sixty-four-channel CT with multioblique reformatting is sensitive and specific for identifying cochlear-facial dehiscence, with rates similar to those in postmortem series. Jugular bulb-vestibular aqueduct dehiscence is a common incidental finding and is unlikely to produce third window physiology. Other non-superior semicircular canal third window dehiscences are rare in asymptomatic patients.

ABBREVIATIONS: CFD = cochlear-facial nerve dehiscence; JVD = jugular bulb-vestibular aqueduct dehiscence; OCD = otic capsule dehiscence; SSCD = superior semicircular canal dehiscence

There are 2 anatomic locations where pressure is normally transmitted between the middle and inner ear, the oval and round windows. Third window syndromes occur when an additional communication forms between the inner ear and surrounding spaces, resulting in altered perilymph hydrodynamics and symptoms including hearing loss, tinnitus, autophony, and sound- or pressure-induced vertigo.¹⁻³ These most frequently

occur as a result of bony otic capsule dehiscence (OCD) as is seen with superior semicircular canal dehiscence (SSCD). Since the discovery of CT-positive SSCD in humans, other sites of OCD have been identified, including cochlear-facial nerve canal dehiscence (CFD), cochlear-carotid canal dehiscence, cochlear-internal auditory canal dehiscence, superior semicircular canal-superior petrosal sinus dehiscence, posterior semicircular canal dehiscence, lateral semicircular canal dehiscence, and vestibular dehiscence.^{1,4,5} Although not an OCD, jugular bulb-vestibular aqueduct dehiscence (JVD) has also been reported to occasionally produce third window symptoms.⁶ Additional examples of anatomic variants that can result in third window syndromes include an enlarged vestibular aqueduct, an X-linked stapes gusher, and bone dyscrasias.⁵ While much research has been

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devoted to SSCD, the other forms of OCD have received relatively little attention. Importantly, diagnosis of OCD in patients experiencing auditory/vestibular symptoms allows surgical intervention, which has been shown to reduce often debilitating third window symptoms and improve quality of life.^{7,8}

In the original literature on the CT diagnosis of OCD, CT was performed on scanners without the capability of submillimeter section thickness, resulting in increased rates of false-positive findings.⁹ Modern 64-channel helical scanners are much more specific in detecting SSCD so that the rate of CT findings of SSCD in asymptomatic patients now approaches the rate of histologic findings in cadavers, and otolaryngology evaluation can be recommended when SSCD is incidentally discovered on CT.¹⁰ It remains unclear whether this rate is true for CFD (and other non-SCC forms of OTC), however, because there is currently a discrepancy between the published prevalence of radiologic CFD (6.3%–9.2%) and the prevalence of histologic CFD seen on temporal bone analysis (0.59%–1.6%).^{11–14}

The purpose of this study was to measure the rate of non-SSCD on high-resolution CT in asymptomatic individuals and compare that with the rate in patients with audiologic or vestibular symptoms.

MATERIALS AND METHODS

This study was approved by University of Pittsburgh Medical Center institutional review board as an exempt study of existing data.

Patients

Pre-hoc power analysis indicated that for a 95% CI spanning 4% and a finding with an underlying rate of 5%, 500 patients would be required. Adult patients who had undergone high-resolution temporal bone CT in the emergency department of a large tertiary care academic medical center between February 2012 and December 2022 were retrospectively identified. Only patients scanned on a 64-channel CT scanner were eligible. Although we enrolled patients presenting to the emergency department at an adult hospital, we included the small number of adolescent patients (14–17 years of age) who met inclusion because existing evidence suggests that otic capsule development is generally complete by adolescence.¹⁵ Patients with pathology within the otic capsule or fractures that prevented otic capsule evaluation were not included. Patients were divided into 2 groups based on the indication for imaging: patients with audiologic or vestibular symptoms that might be consistent with a third window syndrome (symptomatic patients) and those with all other imaging indications (asymptomatic patients). For this study, symptoms consistent with third window syndrome were vertigo, hearing loss, dizziness, and tinnitus. Symptoms were based on review of systems obtained in the emergency department; for those with hearing loss, audiometric data were not available to differentiate between conductive and sensorineural hearing loss. Patients with auditory or vestibular symptoms were excluded from the asymptomatic patient group. We concluded enrollment when we had accumulated 500 asymptomatic patients, in keeping with our power analysis.

Imaging Protocol and Techniques

CT was acquired using LightSpeed 64-channel CT scanners (GE Healthcare) with a section thickness of 0.63 mm, spacing of

0.375 mm, 120 kV(peak), 195 mA, pitch of 0.53°, a bone kernel, and a matrix of 512 × 512. Reformatted images in sagittal and coronal planes at 1-mm section thickness and 1-mm interslice spacing were routinely obtained on the CT scanners.

Image Interpretation

The patient's indication for imaging, date of birth, date of imaging, and sex were recorded. Patient CT scans were reviewed by a Certificate of Added Qualification–certified neuroradiologist with fellowship training in head and neck radiology and 20 years of experience who was blinded to the indication for the examination. The presence of OCD was recorded and categorized as CFD, JVD, cochlear–internal auditory canal, cochlear–carotid canal, or other dehiscence (eg, posterior semicircular canal dehiscence, lateral semicircular canal dehiscence, and vestibular aqueduct dehiscence). Screening for dehiscence was in the axial plane, with 1-mm sagittal reformatted images serving to corroborate findings in the axial plane. In patients who demonstrated dehiscence on both axial and sagittal images, final confirmation of CFD was assessed using postprocessing software (Vitrea; Canon Medical Systems) to produce high-definition multiplanar oblique reformats. These additional images were 0.625-mm thick with interslice spacing of 0.625 mm. The plane of reformat was a modified plane of Stenvers, along the basal turn of the cochlea and orthogonal to the labyrinthine portion of the facial nerve. The reformats were angled in both the coronal and sagittal plane to ensure that the entire basal turn of the cochlea was included in a single image. Interpretation was performed with a high threshold for positivity (ie, a barely perceptible bony covering was still considered intact) to avoid false-positive results.¹⁶

Data Analysis

The prevalence of CFD and JVD was calculated, and 95% CIs were generated using the Newcombe method for binomial proportions.¹⁷ If no events were recorded, 95% CIs were calculated using exact techniques. All statistical calculations were performed using SPSS software, Version 28 (IBM).

RESULTS

Six hundred two patients were included in the study. For every patient, images from both temporal bones were interpreted, for a total of 1204 temporal bones. On the basis of their clinical indication for temporal bone imaging, 500 patients were asymptomatic and 102 were symptomatic. The median age at imaging for asymptomatic patients was 47 years with an age range of 15–93 years. The median age at imaging for symptomatic patients was 50.5 years with a range of 21–87 years. Reasons for examinations in asymptomatic and symptomatic patients are summarized in Tables 1 and 2.

In asymptomatic patients, 100 of 1000 temporal bones (10%) exhibited sufficient evidence of CFD on axial imaging to prompt evaluation in sagittal reformatted images (Fig 1). Of these 100 temporal bones, 14 (1.4% of the asymptomatic population) demonstrated apparent CFD dehiscence on the automated 1-mm-thick sagittal reformatted images. Following evaluation of these 14 temporal bones in high-definition multioblique reformatted images, only 8 temporal bones (0.8%) demonstrated CFD (Fig 2).

Table 1: Asymptomatic patient characteristics and indications for temporal bone CT imaging

Asymptomatic Patients	
Median age (range) (yr)	47 (15–93)
No. Male (%)	325 (65.0%)
Imaging indication	
Trauma	368 (73.6%)
Mass	45 (9.0%)
Infection	37 (7.4%)
Pain	20 (4.0%)
CSF leak/cephalocele	15 (3.0%)
Cranial nerve palsy	5 (1.0%)
Otosclerosis	4 (0.8%)
Surgical planning	2 (0.4%)
Other	4 (0.8%)

Table 2: Symptomatic patient characteristics and indications for temporal bone CT imaging^a

Symptomatic Patients	
Median age (range) (yr)	50 (14–87)
No. of males (%)	43 (42.2%)
Imaging indication	
Dizziness	52 (51.0%)
Hearing loss	16 (15.7%)
Suspected SSCD	14 (13.7%)
Tinnitus	7 (6.9%)
Vertigo	6 (5.9%)
Otosclerosis	3 (2.9%)
Multiple auditory/vestibular symptoms	3 (2.9%)
Mass involving otic capsule	1 (1.0%)

^a Symptomatic patients with multiple symptoms were classified on the basis of their primary symptom, so that there is no overlap between imaging indications.

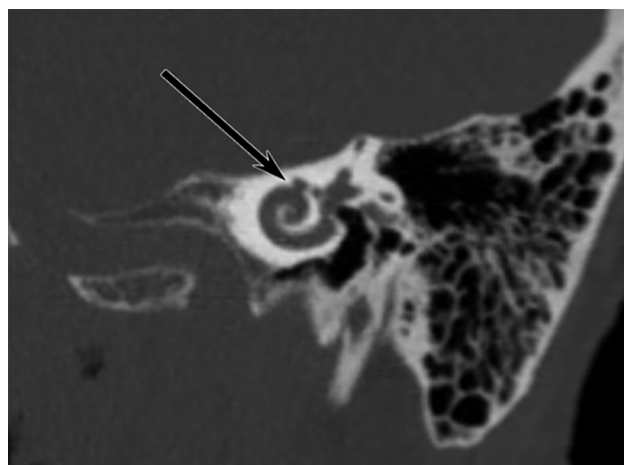


FIG 1. Cochlear-facial dehiscence. Oblique reformatted CT scan along the basal turn of the cochlea (modified Stenvers reformat) shows a dehiscence (arrow) between the middle turn of the cochlea and the labyrinthine segment of the facial nerve canal.

On a patient-level analysis, these represented 1.6% of the asymptomatic patient population (95% CI, 0.8%–3.1%). There were no cases of bilateral CFD.

In asymptomatic patients, 41 of the 1000 temporal bones had JVD (Fig 3), with 1 patient exhibiting bilateral JVD, yielding a dehiscence prevalence of 4.1% per ear and a patient-level prevalence of 8.0% (95% CI, 5.9%–10.7%).

Of the 102 symptomatic patients, 3 demonstrated CFD and 8 demonstrated JVD, resulting in a CFD and JVD patient prevalence of 2.9% (95% CI, 1.0%–8.3%) and 7.8% (95% CI, 4.0%–14.7%), respectively. One symptomatic patient presenting with hearing loss exhibited left-sided CFD with concomitant, contralateral JVD.

No statistically significant difference existed between the rates of CFD and JVD in symptomatic-versus-asymptomatic patients. No instances of cochlea-carotid, cochlea-internal auditory canal, or other sites of dehiscence were identified in the 1204 temporal bones studied (95% CI, 0.0%–0.2%; Fig 4).

DISCUSSION

In the current study of 500 patients without third window symptoms and 102 patients with possible third window symptoms, 1.6% of asymptomatic patients exhibited CFD and 8.0% exhibited JVD. The values in patients with symptoms that might be attributable to third window dehiscence are similar (2.9% CFD and 7.8% JVD). This finding contrasts with the those in SSCD, in which symptomatic patients have a substantially higher rate of dehiscence.¹⁰ This presumably reflects the rarity of symptomatic CFD and JVD, making it harder to discern any differences, even with a relatively large patient population.

Two previous radiologic studies using temporal bone CTs have reported a CFD prevalence ranging from 6.3% to 9.2%.^{11,12} Both of these studies have limitations that likely contributed to the discrepancy between radiologic and histologic CFD prevalence, however. The first study examined only patients presenting to highly specialized, academic neurotology clinics with auditory and/or vestibular symptoms, making it difficult to extrapolate their findings to the general population. Additionally, this study included both conventional temporal bone CTs with section thickness up to 1 mm as well as images produced by conebeam CT, making it difficult to draw conclusions on the specificity of conventional, thin-section temporal bone CT in diagnosing CFD. The second study used a 16-section CT scanner for image acquisition and exclusively relied on image interpretation in conventional CT planes without using optimized oblique plane reformats to evaluate the labyrinthine section of the facial nerve in an orthogonal plane. Despite these design and technological limitations, authors in both studies concluded that CT overcalled CFD and that limitations in CT section thickness and volume-averaging protocol effects obfuscated specific radiologic detection of CFD. Our results contradict the assertion that modern CT scan techniques inherently overcall CFD.

The low prevalence rates of CFD that we identified were highly dependent on our use of multioblique images along the basal turn of the cochlea. When only axial slices were analyzed, the apparent prevalence was 12 times higher than the true prevalence. The use of routine sagittal reformats improved the false-positive rate, but it was still twice the true value. Thus, we recommend that high-resolution multiplanar oblique reformats be used before making a diagnosis of CFD on CT. This method contrasts with radiologic SSCD detection, in which conventional coronal reformatted images are sufficient for diagnosis.¹⁶

The radiologic CFD prevalence in the current study (0.9% of all temporal bones) is congruent with the rate in 2 prior histologic studies that reported rates of temporal bone dehiscence instead of

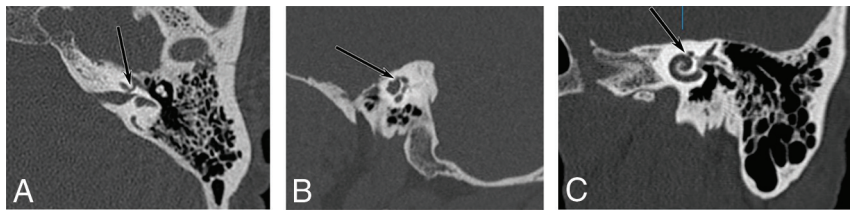


FIG 2. The importance of multiplanar oblique reformats for the diagnosis of cochlear-facial dehiscence. Axial CT image (A) shows an apparent dehiscence (arrow) between the middle turn of the cochlea and the labyrinthine segment of the facial nerve. Sagittal reformatted image (B) appears to confirm the dehiscence (arrow). However, multiple oblique reformats along the basal turn of the cochlea (C) reveal that the bone between the cochlea and the facial canal (arrow) is thin but intact.

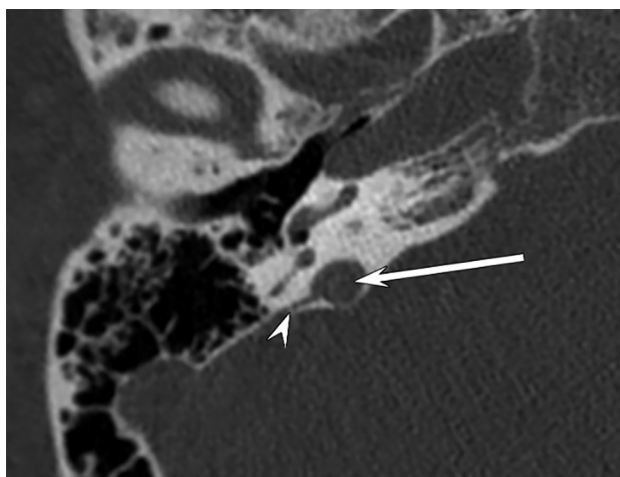


FIG 3. JVD. Axial CT image shows dehiscence between a diverticulum of the jugular bulb (arrow) and the vestibular aqueduct (arrowhead).

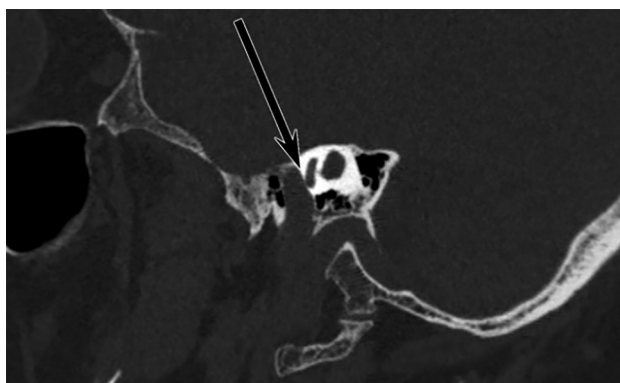


FIG 4. Cochlear-carotid plate. Sagittal reformatted CT image shows thin-but-intact bone (arrow) between the petrous segment of the ICA and the basal turn of the cochlea. None of the 1204 temporal bones in this study demonstrated a true dehiscence at this location.

patient-level prevalence. Fang et al¹³ reported a CFD prevalence of 0.59% in a histologic survey of 1020 temporal bones at Johns Hopkins, while Schart-Morén et al¹⁴ reported a slightly higher CFD prevalence of 1.6% in molds from 282 temporal bones at Uppsala University in Sweden. Differences between these 2 studies can be explained by small differences in CFD prevalence

rates among different ethnic groups as well as incomplete coverage of the otic capsule during the molding process used in the latter study. Unfortunately, neither of the histologic studies reported patients' clinical characteristics, so it is unknown whether patients that exhibited confirmed CFD dehiscence demonstrated third window symptoms, and conclusions cannot be drawn on the prevalence of histologic dehiscence in asymptomatic-versus-symptomatic patients.

Our observations on JVD, cochlear-carotid dehiscence, and cochlear-internal auditory canal dehiscence prevalence are consistent with those in prior studies.^{6,11,18,19} While JVD was the most frequently observed OCD here, the lack of a difference in prevalence between symptomatic and asymptomatic patients suggests that it represents a benign anatomic variant in most individuals and is an infrequent cause of third window symptoms. This suggestion supports the conclusions of a prior study examining the association between JVD and auditory/vestibular symptoms in a cohort of 8325 patients undergoing temporal bone CT imaging for all causes.²⁰ The lack of observed cochlear-carotid, and cochlear-internal auditory canal dehiscences in this study suggests that very few individuals in the general population have these anatomic variants.

In patients with incidentally discovered SSCD, the rate of sub-clinical hearing and vestibular abnormalities is sufficiently high to recommend clinical evaluation, even in the absence of patient-reported symptoms.¹⁰ In JVD and CFD, this relationship is less clear. Our study found no statistically significant difference in the rates of radiologic findings between patients with reported symptoms suggestive of a possible third window syndrome and those without auditory or vestibular symptoms; a prior report on JVD had similar findings as did a report of CFD in which only 1 patient had hearing loss unattributable to any other cause.^{12,18} Surgical intervention has been proposed as an option for patients with symptomatic CFD, as has endovascular intervention for JVD.^{7,21} We, thus, advise caution before ascribing symptomatology to these radiographic findings. In particular, our data show that JVD is a very common incidental finding among asymptomatic patients. Also, our data show that CFD is likely to be over-called, even with modern, multidetector CT scanners, unless particular attention is given to reconstructing in multiple planes, most notably the modified Stenvers plane along the basal turn of the cochlea orthogonal to the labyrinthine facial nerve.

Our study has several limitations. We included patients from a single institution, and interpretations were performed by a single radiologist. There was no attempt to assess intra- or inter-reader variability. Our asymptomatic patient population had a male predominance, likely due to the high proportion of trauma cases in the emergency department. Because our results are dependent on image generation using 64-section CT scanners with a section thickness of 0.63 mm and overlap of 0.375 mm, our results may not be generalizable to imaging protocols using different specifications.

CONCLUSIONS

Modern 64-channel CT scanning techniques can demonstrate third window dehiscence such as CFD with rates similar to those in postmortem series. In asymptomatic patients, the radiologic prevalences of CFD and JVD are 1.6% and 8.0%, respectively. Excluding SSCD, no other third window dehiscences were observed in our study population. Diagnosis of non-SSCD requires that images be obtained with submillimeter section thickness and overlap, that images be reformatted in multiplanar oblique planes, and that images be interpreted with a high degree of specificity. JVD is a common incidental radiologic finding and is not necessarily indicative of third window pathophysiology, and neither CFD nor JVD, when seen incidentally in asymptomatic individuals, requires further testing. Cochlear-carotid and cochlear-internal auditory canal dehiscences are extremely rare in the asymptomatic population and were not observed in our study.

Disclosure forms provided by the authors are available with the full text and PDF of this article at www.ajnr.org.

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