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Vestibular Atelectasis: Myth or Reality?

Michael Eliezer, MD; Arnaud Attyé, MD; Jean-Pierre Guichard, MD; Hélène Vieux, MD; Antoine Guilloulet, MD; Michel Toupet, MD; Philippe Herman, MD, PhD; Romain Kania, MD, PhD; Emmanuel Houdart, MD, PhD; Charlotte Hautefort, MD

Objectives/Hypothesis: Because delayed post-contrast three-dimensional fluid-attenuated inversion recovery imaging sequences enable the distinction between the utricle and the saccule, we raised the hypothesis that patients with vestibular atelectasis (VA) could show unilateral collapse of the utricle and the ampullas on imaging.

Study Design: Retrospective case series.

Methods: We retrospectively reviewed 200 patients who underwent 3 T magnetic resonance imaging (MRI) after intravenous administration of gadolinium. MRI scans were assessed for the presence of VA. The endolymphatic space was considered as collapsed when the utricle and at least two ampullas were not visible or were barely visible.

Results: We reported four patients with VA on MRI responsible for atypical clinical presentations of acute vestibular deficit. All patients presented a specific involvement of the pars superior sensory captors (utricle, ampullas), preserving the pars inferior sensory captors (cochlea and saccule). This was confirmed both clinically and on MRI.

Conclusions: Our study is the first to describe in vivo unilateral collapse of the pars superior on delayed postcontrast MRI in patients with a clinical unilateral vestibular loss.

Key Words: Magnetic resonance imaging, vestibular atelectasis, vestibular neuritis.

Level of Evidence: 4

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INTRODUCTION

Merchant and Schuknecht were the first to describe, in histopathological studies in patients presenting with vertigo, the collapse of the walls of the ampullas and the utricle, also known as vestibular atelectasis (VA).1 The initial symptoms of VA can be paroxysmal or insidious. The paroxysmal form is similar to vestibular neuritis; patients present with a sudden vertigo during several days without cochlear or neurological symptoms, followed by chronic disequilibrium. Making the clinical distinction between these two entities was only possible in histopathological studies. By contrast, the insidious form is characterized by a subtle onset of vertigo, followed by chronic unsteadiness or transient vertigo.

Recently, Wenzel et al. have described patients with bilateral vestibular loss associated with pressure-induced nystagmus and normal hearing.2 They have suggested that these patients presented with a collapsed membranous labyrinth that might come in contact with the stapes, causing abnormal vestibular stimulation with loud sound and/or pressure (Tullio or Hennebert signs). Yet, there was no direct evidence of this disorder. Furthermore, in the study of Merchant and Schuknecht, all pathological cases of VA were unilateral, and loud sound-induced vertigo was never reported.1

Delayed postcontrast three-dimensional fluid-attenuated inversion recovery (3D-FLAIR) sequences enable the distinction between the utricle and the saccule, which is crucial for the diagnosis of endolymphatic hydrops in patients with Menière’s disease.3–6 Based on the pathophysiology of VA, we raised the hypothesis that patients with VA could show unilateral collapse of the utricle and the ampullas on delayed postcontrast 3D-FLAIR sequences.

MATERIALS AND METHODS

This retrospective imaging study included 200 patients with unilateral cochleovestibular symptoms who underwent magnetic resonance imaging (MRI) examination with delayed acquisition at least 4 hours after intravenous administration of gadolinium chelate. Patients were recruited consecutively between May 2017 and June 2018. Patients with a history of exposure to aminoglycosides or chemotherapy, head trauma, stapes surgery, and family history of vestibular loss were excluded.

Imaging

MRI examinations were carried out on a 3 T scanner (3 T Siemens Skrya; Siemens Healthineers, Erlangen, Germany) with a 64-channel head-neck-spine coil. All patients underwent an MRI scan 4 hours after a single intravenous dose of gadobutrol (Gd-DOTA-butrol Gadovist 0.1 mmol/kg, 1 mmol/mL) that provided a
high contrast in the labyrinth.\textsuperscript{7} We performed the 3D-FLAIR with the following parameters: field of view: 160 × 160 mm, repetition time: 10,000 ms, echo time: 323 ms, inversion time: 2500 ms, 320 × 280, flip angle: 180°, number of excitations: 2, generalized autocalibrating partially parallel acquisitions: 2, and scan time of 7 minutes 50 seconds.

The presence of the following structures was verified on the 3D-FLAIR sequence:

- The saccule: The saccule appears as an area of low signal. It is located on the medial and anterior wall of the vestibule underneath the level of the lateral semicircular canal.\textsuperscript{8,9}
- The utricle: The utricle appears as an elliptical zone of low contrast in the labyrinth.\textsuperscript{7} We performed the 3D-FLAIR with ocular re

470 F Bilateral Not performed Normal Reduced Reduced Absent Normal Barely visible Not visible Normal
343 M Normal 100 Normal Reduced Reduced Absent Normal Barely visible Barely visible Normal
269 F Normal Not performed Reduced Reduced Reduced Absent Normal Not visible Not visible Normal
130 F Normal 92 Normal Reduced Reduced Absent Normal Not visible Not visible Normal

Cervical Vestibular Evoked Myogenic Potentials

During the cervical vestibular evoked myogenic potentials (cVEMP) test recordings, all subjects were asked to rotate their head away from the stimulated side. Surface electromyogram activity was recorded with superficial electrodes. Tone bursts (90 dB nHL, 500 Hz) were presented through headphones. The amplitude of the first positive–negative peak (P13–N23) was recorded. Absence of a meaningful wave form with p13 and n23 was defined as no response.

Ocular Vestibular Evoked Myogenic Potentials

During the ocular vestibular evoked myogenic potentials (oVEMP) test recordings, all subjects were asked to look straight up at a small fixed target above them. The active electrodes were placed on the face, below the center of the lower eyelid. The reference electrode was placed about 1 cm below the active electrode on the cheek, and the ground electrode was placed on the forehead. Tone bursts (95 dB nHL, 500 Hz) were presented through headphones. The initial negative–positive biphasic waveform comprised peaks N1 and P1. An absence of a meaningful waveform with N1 and P1 was defined as no response.

Video Head Impulse Test

For the video head impulse test (VHIT), at least five head impulses were performed in the axis of each semicircular canal. The threshold for normal vestibulo-ocular reflex gain (VOR) was 0.64 for vertical canals and 0.78 for horizontal canals.\textsuperscript{10}

**RESULTS**

Clinical findings are summarized in Table I. We have identified four patients with unilateral collapse of the utricle and the ampullas. None of these patients had a family history of vestibular deficit or a history of exposure to aminoglycosides or chemotherapy that could explain the vestibular loss.

Case 1

A 30-year-old female presented, for 3 years, recurrent spells of acute rotatory vertigo lasting at least 3 hours. She complained about intermittent bilateral tinnitus and aural fullness lasting less than a minute that was not concomitant with the onset of vertigo. She also described spells of vertigo provoked by loud sound. She had a history of Raynaud's disease. The ocular examination did not find a spontaneous or provoked nystagmus. There was no gaze-evoked nystagmus or no skew deviation. The Dix-Hallpike test found no evidence of positional nystagmus.

The audiogram revealed normal threshold on both sides (Fig. 1). VHIT revealed a decreased right lateral (0.74) and right posterior (0.66) VOR gain. The oVEMP was not found on the right side, suggesting a right utricular hypofunction, whereas the cVEMP was found and confirmed a normal saccular function. Caloric tests found a profound right canal paresis (92%).

<table>
<thead>
<tr>
<th>Patient</th>
<th>Age, yr</th>
<th>Sex</th>
<th>Hearing</th>
<th>Canal Paresis (%)</th>
<th>VOR (Gain)</th>
<th>Imaging</th>
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<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Superior</td>
<td>Lateral</td>
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<tr>
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<td>F</td>
<td>Normal</td>
<td>92</td>
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<td>Not performed</td>
<td>Reduced</td>
<td>Reduced</td>
</tr>
<tr>
<td>3</td>
<td>43</td>
<td>M</td>
<td>Normal</td>
<td>100</td>
<td>Normal</td>
<td>Reduced</td>
</tr>
<tr>
<td>4</td>
<td>70</td>
<td>F</td>
<td>Bilateral</td>
<td>Not performed</td>
<td>Normal</td>
<td>Reduced</td>
</tr>
</tbody>
</table>

\textsuperscript{cVEMP} = cervical vestibular evoked myogenic potential; F = female; M = male; \textsuperscript{oVEMP} = ocular vestibular evoked myogenic potential; VOR = vestibulo-ocular reflex.
The utricle and all three ampullas were not displayed on the right side with delayed postcontrast 3D-FLAIR sequences (Fig. 1). There was no radiological sign of endolymphatic hydrops or blood-labyrinthine barrier impairment.

Case 2

A 69-year-old female presented 10 years ago with an acute rotatory vertigo during 48 hours without cochlear symptoms suggesting vestibular neuritis. She had a history of dyslipidemia and migraine. Two years later, she
experienced two spells of unsteadiness associated with oscillopsia and then complained of recurrent episodes of disequilibrium without rotatory vertigo. The ocular examination did not find a spontaneous nystagmus; however, a right nystagmus with horizontal and torsional components was found during the head-shaking test. The nystagmus decreased during eye fixation. There was no gaze-evoked nystagmus or no skew deviation. The Dix-Hallpike test found no evidence of positional nystagmus. Based on the history of migraine, vestibular migraine was also suspected. Yet, the patient did not meet the criteria of vestibular migraine, because she had no migraine features during vestibular symptoms.

The audiogram revealed normal threshold on both sides. VHIT showed a decreased VOR gain of the superior (0.44), lateral (0.51), and posterior (0.49) semicircular canals on the left side. The oVEMP was absent on the left side, whereas the cVEMP was present. The left utricle and ampullas were barely visible on the left side with delayed postcontrast 3D-FLAIR sequences (Fig. 2). There was no radiological sign of endolymphatic hydrops or blood-labyrinthine barrier impairment.

Fig. 3. (A) Axial three-dimensional fluid-attenuated inversion recovery (3D-FLAIR) imaging at the level of the utricle (white arrow) and the lateral ampulla (white dotted arrow). The utricle and the lateral ampulla were barely visible on the left side. (B) Axial 3D-FLAIR at the level of the saccule (grey arrow) and the posterior ampulla (grey dotted arrow) demonstrated a left partial collapsed posterior ampulla in comparison to the contralateral asymptomatic side. The saccule appeared normal on both sides.

Fig. 4. (A) Axial three-dimensional fluid-attenuated inversion recovery (3D-FLAIR) imaging at the level of the utricle (white arrow) and the lateral ampulla (white dotted arrow). The utricle and the lateral ampulla were barely visible on the left side. (B) Axial 3D-FLAIR at the level of the saccule (grey arrow) and the posterior ampulla (grey dotted arrow) demonstrated a left total collapsed posterior ampulla in comparison to the right side. The saccule appeared normal on both sides.
Case 3
A 43-year-old male was referred to our tertiary neuro-otological center because he first developed an acute rotatory vertigo followed by unsteadiness lasting at least 2 days. Then, he experienced recurrent spells of rotatory vertigo (20 to 30 seconds). He denied having any auditory symptoms, such as tinnitus, hearing loss, or aural fullness. On examination, a right spontaneous nystagmus with horizontal and torsional components was noticed. The nystagmus decreased during eye fixation. There was no gaze-evoked nystagmus, no skew deviation, or loud sound and/or pressure-induced nystagmus. The Dix-Hallpike test found no evidence of positional nystagmus.

The audiogram revealed normal hearing on both sides, but VHIT showed a decreased VOR gain of the left lateral (0.67) and posterior (0.65) canals, whereas the anterior VOR gain was preserved (0.85). The oVEMP was absent on the left side, whereas the cVEMP was present, suggesting that only the left utricular was involved. Moreover, a profound left canal paresis (100%) was observed with caloric tests.

The left utricle and ampullas were barely visible on the left side with delayed postcontrast 3D-FLAIR sequences, suggesting a collapsed membranous labyrinth (Fig. 3). There was no radiological sign of endolymphatic hydrops or blood-labyrinthine barrier impairment.

Case 4
A 70-year-old female was referred to our tertiary neuro-otological center. She presented an acute unsteadiness and nausea but did not complain of cochlear symptoms such as tinnitus and hearing loss. She had a history of dyslipidemia and hypertension. The ocular examination performed with 3D infrared video-oculography (VNG Framiral, Grasse, France) did not find a spontaneous nystagmus; however, a right nystagmus with horizontal and torsional components was found during the head-shaking test and decreased during eye fixation. There were no Tullio or Hennebert signs. The Dix-Hallpike test found no evidence of positional nystagmus.

The audiogram revealed bilateral, mild low-tone sensorineural hearing loss. PTA was 26.5 dB on the right side and 25.25 dB on the left side. VHIT revealed a decreased VOR gain in the superior (0.74), lateral (0.45), and posterior (0.54) semicircular canals on the left side. The oVEMP was absent on the left side, whereas the cVEMP was present. This association suggests a left vestibular hypofunction focus on utricular and semicircular canal captors.

Delayed postcontrast 3D-FLAIR sequences (Fig. 4) showed a collapsed utricle involving also all the ampullas on the left side in comparison to the right side where endolymphatic structures appeared normal. There was no radiological sign of endolymphatic hydrops or blood-labyrinthine barrier impairment.

DISCUSSION
We reported four patients with VA on MRI responsible for atypical clinical presentations of acute vestibular deficit. All patients presented a specific involvement of the pars superior sensory captors (utricle, ampullas), preserving the pars inferior sensory captors (cochlea and saccule). This was confirmed both clinically and on MRI. VA has been already suggested on histological reports and clinical observations, but no radiological images have confirmed it.

In 1988, Merchant and Schuknecht described in a histopathological study, eight of 213 subjects who presented with a collapse of the walls of the ampullas and utricle, which was the primary pathologic event responsible for the vertigo. They have carefully ruled out the possibility that these histopathological findings were caused by preparation artifacts and have excluded Menière’s disease, perilymphatic fistula, vestibular schwannoma, labyrinthitis, temporal bone fractures, and surgical trauma to the inner ear to assess only primary VA. Collapse was considered as significant when at least two ampullas were involved with a partial or total atelectasis. The utricle and at least two ampullas were always involved, whereas the saccule and the cochlea were always preserved. Here, we have carefully excluded secondary VA based on clinical and imaging findings. Furthermore, to our knowledge, collapse of the utricle in Menière’s disease patients or in healthy subjects has never been reported in case-controlled imaging studies.

Merchant and Schuknecht have described in patients with VA two initial presentation forms. First, in the paroxysmal form, patients experienced a sudden spell of vertigo without auditory or neurologic symptoms. Second, in the insidious form, the onset is subtle. In both cases, the initial event is followed by chronic unsteadiness or transient recurrent spells of vertigo. Here, two of our four patients (patients 2 and 3) presented, at the initial stage of the disease, an acute rotatory vertigo during several days without any cochlear symptoms, as were observed in the paroxysmal form of VA by Merchant and Schuknecht. After the initial event, patient 2 presented recurrent spells of unsteadiness associated with oscillopsia, whereas patient 3 experienced recurrent spells of rotatory vertigo lasting for 20 to 30 seconds. Surprisingly, patient 1 also described fluctuant cochlear symptoms such as tinnitus and aural fullness as could be observed in Menière’s disease. However, these cochlear symptoms were not concomitant with the onset of vertigo. Furthermore, it has been reported lately that outside the episodes of crisis, patients with advanced Menière’s disease had normal VOR gain on VHIT, in contrast to abnormal caloric tests. Here, patient 1 had both abnormal VOR gain and caloric tests. Moreover, MRI did not show any evidence of endolymphatic hydrops.

In our study, all patients presented with a unilateral hypofunction of the utricle on the affected side, as evaluated with oVEMP, whereas the saccule function was still normal as confirmed with cVEMP. Regarding semicircular canal function, both high and low frequencies, as evaluated by VHIT and caloric tests, were abnormal. However, responses to caloric stimulation showed a profound and complete unilateral areflexia on the affected side, whereas VHIT suggested a partial deficit. As Wenzel et al. have claimed, we also suggest that a collapse of the
endolymphatic space of the semicircular canal could stop low-velocity stimulation, whereas high-velocity stimulation can work.\textsuperscript{2}

Recently, two studies have described patients with bilateral vestibular hypofunction associated with Tullio phenomenon and pressure-induced nystagmus.\textsuperscript{2,11} They have raised the hypothesis that these patients had VA, but unfortunately, they could not prove this diagnosis. As Kaski and Bronstein have stated, we also believe that there is insufficient evidence to claim a correlation between bilateral vestibular hypofunction and VA.\textsuperscript{12} In the study of Merchant and Schuknecht, all pathologic cases of VA were unilateral, and no Tullio sign was reported. We should mention that our study focused only on unilateral disease, and none of our four patients had bilateral impairment on MRI. Wenzel et al.\textsuperscript{2} raised the hypothesis that the collapsed membranous labyrinth might come in contact with the stapes, causing abnormal vestibular stimulation with loud sound and/or pressure. Interestingly, one of our four patients (patient 1) had loud sound-induced vertigo, but MRI did not show any evidence of contact between the membranous labyrinth and the stapes. Yet, we could not exclude the possibility that this contact was too subtle to be detected by MRI.

3D-FLAIR sequences with variable flip angle refocusing have the advantage of increasing echo train length, enabling a shorter acquisition time without blurring. They have also the advantage of being more sensitive than conventional sequences to detect T1-shortening.\textsuperscript{13,14} Hydrops exploration with MRI consists in a delayed 3D-FLAIR acquisition 4 hours after contrast media administration, allowing the distinction between the utricle, the saccule, and the ampullas from the perilymphatic space. Delayed 3D-FLAIR sequences also evaluate the permeability of the blood-labyrinth barrier in various otological diseases.\textsuperscript{15–17} It has been demonstrated that patients with acute vestibular syndrome, also known as vestibular neuritis, presented with a strong enhancement of the semicircular canals.\textsuperscript{18}

**Potential Therapeutic Implications**

VA becomes one of the differential diagnoses of acute vestibular deficit, Menière’s disease, perilymphatic fistula, and multiple sclerosis. MRI could enable a better management of those patients, because the distinction between these clinical entities could be challenging. Faced with a unilateral acute vestibular deficit, the paraxysmal form of VA could be missed by the clinician. Yet, on one hand, MRI could demonstrate unilateral utricular and ampullar collapse in cases of VA, and on the other hand, an increased enhancement of the semicircular canals as has been reported in patients with acute unilateral vestibular deficit.\textsuperscript{18} Furthermore, MRI could show saccular hydrops in patients with Menière’s disease, and also highlight the presence of a perilymphatic fistula, when round and/or oval window recesses are filled with a fluid leak.\textsuperscript{4}

To date, we do not know the mechanism causing VA. However, we expect that local treatment using intratympanic administration might be adapted to target this intralabyrinthine disease.\textsuperscript{19} As Wenzel et al.\textsuperscript{2} have suggested, VA could involve both labyrinths and could be one mechanism of bilateral vestibular deficit, but this idea could not be confirmed by MRI or histology. Postcontrast 3D-FLAIR sequences should bring us more information concerning this bilateral vestibular disease and confirm what Wenzel et al. have suggested. Moreover this patient population could be eligible for vestibular implants, because the lesions affected only the labyrinthine sensory receptors and not the vestibular nerve. In this case, we could imagine that the direct vestibularafferent nerve stimulation could bypass the collapsed vestibular sensory structures.\textsuperscript{20}

The main problem we encountered was the impossibility of confirming the volume of the vestibular endolymphatic space with pathological analysis in vivo. Although these results are encouraging in a small number of patients, further multicenter studies are needed in a larger population to assess these patients. Based on the design of our study, we cannot exclude the possibility of bilateral VA in patients with bilateral vestibular loss, as it has been reported in the study of Viana et al.\textsuperscript{21} Opening the study to idiopathic bilateral vestibular deficit cases would improve doctors’ clinical etiologic diagnoses and our knowledge of vestibular disorders.

Another limitation is that MRI was obtained without precontrast or immediate postcontrast 3D-FLAIR sequences. However, precontrast or immediate postcontrast FLAIR hyper-intensities have never been described in VA.

We considered the possibility that the endolymphatic space of the pars superior was barely visible due to blood-endolymphatic barrier impairment, as has been observed in acoustic trauma.\textsuperscript{22} However, the intensity of the signal of the endolymphatic space of the pars superior structures did not appear as contrast media effect, so we can eliminate this mechanism.

**CONCLUSION**

To our knowledge, our study is the first to describe in vivo unilateral collapse of the pars superior on delayed postcontrast MRI in patients with a clinical unilateral vestibular loss. We have suggested that the combination of these clinical and MRI findings should alert the clinician and consider VA as one cause of the vestibular deficit lesion.

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**BIBLIOGRAPHY**


