

Endolymphatic disease From hydrops to atelectasis

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 **VERTICO**
CENTRE DE R  F  RENCE DES MALADIES VESTIBULAIRES
ET COCHL  O-VESTIBULAIRES RARES

Vestibular atelectasis.

Merchant SN, Schuknecht HF.

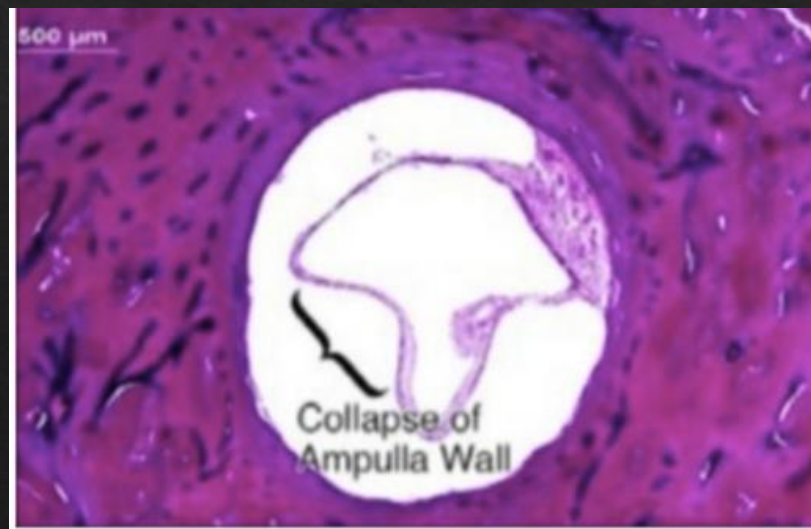
Ann Otol Rhinol Laryngol. 1988 Nov-Dec;97(6 Pt 1):565-76. doi: 10.1177/000348948809700601.

PMID: 3264487

1988

Merchant and Schuknecht described 8 cases out of 213 subjects with a **collapse of the walls of the ampullas and the utricle.**

The endolymphatic collapse was considered responsible for the history of vertigo of these cases



For 20 years, we were unable to demonstrate this endolymphatic atelectasis in clinical practice.

Vestibular atelectasis: Myth or reality?

Eliezer M, Attyé A, Guichard JP, Vitaux H, Guillonnet A, Toupet M, Herman P, Kania R, Houdart E, Hautefort C.

Laryngoscope. 2019 Jul;129(7):1689-1695. doi: 10.1002/lary.27793. Epub 2019 Jan 10.

PMID: 30632154

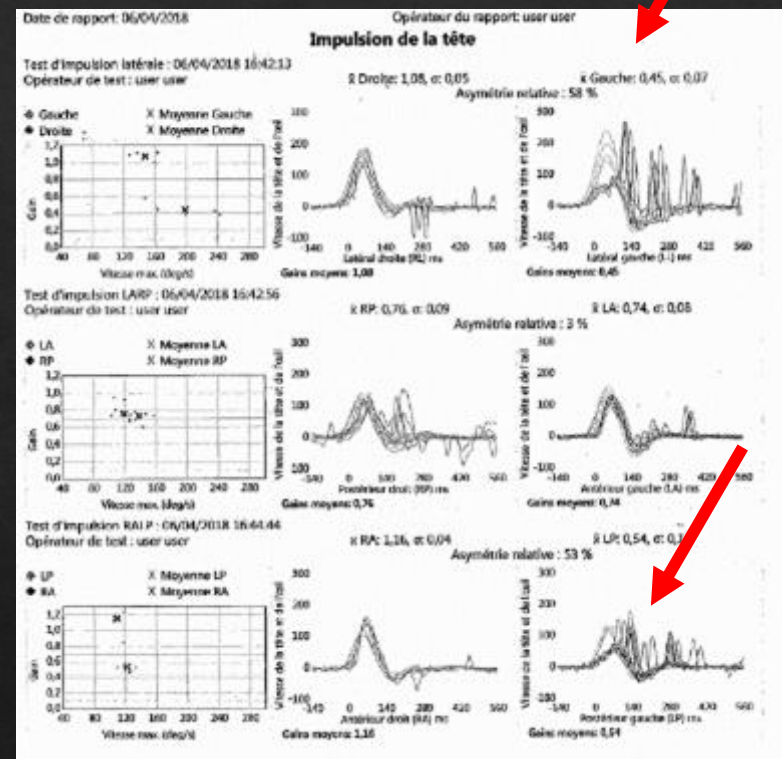
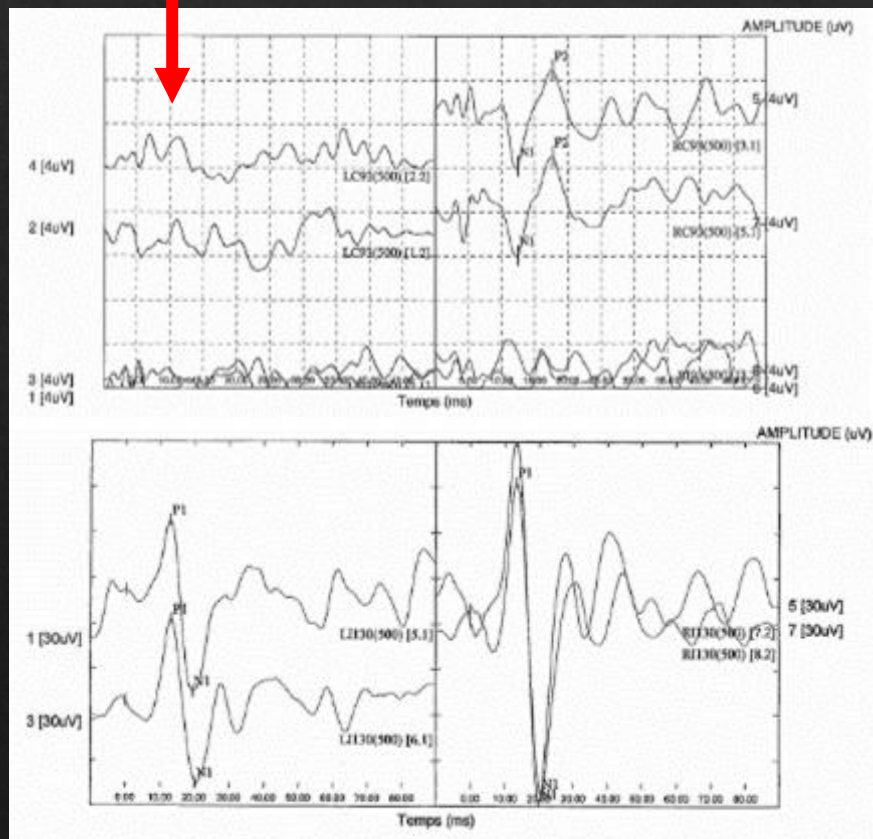
- ✓ Since, 2005, Mrs E, 40 years old, had every 2 years, an history of 2 days of acute rotatory vertigo with spontaneous nystagmus
 - ✓ At each crisis: caloric tests and clinical examination confirmed a left canal deficit.
 - ✓ No history of hearing loss
 - ✓ No history of migraine...

At this step, two diagnosis were proposed by the medical team:

- recurrent left "neuritis"
- or fluctuating hydrops....

- ✓ In 2018 => **ROOM TILT ILLUSION** during few seconds then **dizziness** for 15 days

- What we found at the vestibular tests:
A Left lateral and posterior canal deficit
A Left utricular deficit
Normal audiometric and saccular tests



In 2017, 3D FLAIR delayed MRI allowed us
to identify endolymphatic hydrops in clinical routine
and, finally
to identify vestibular utricular collapse!!

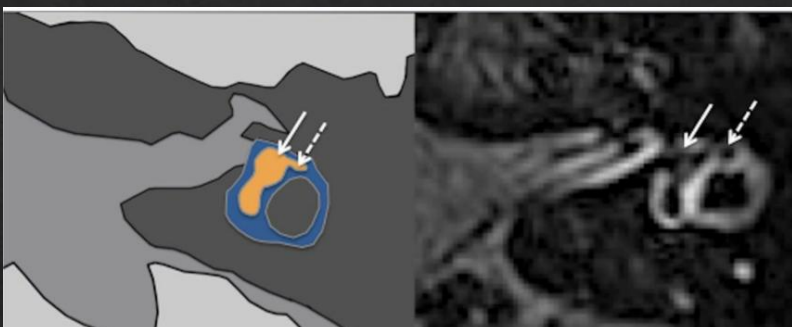
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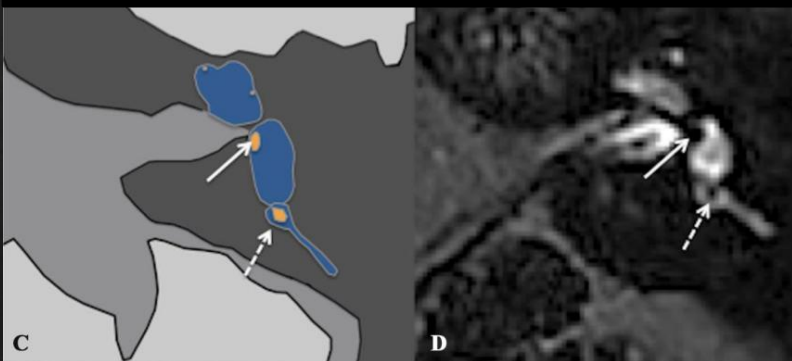
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Normal endolymphatic 3D flair MRI.

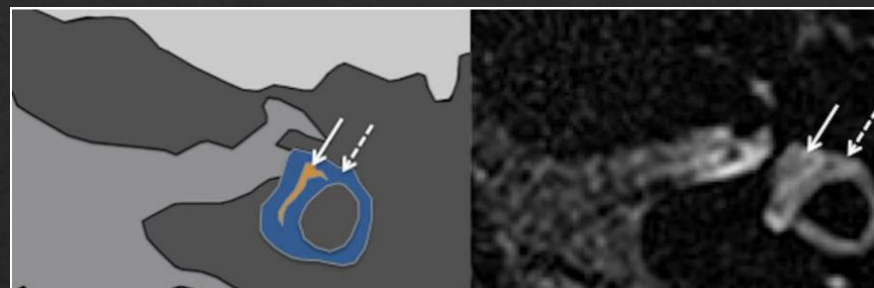


Level of the lateral SCC

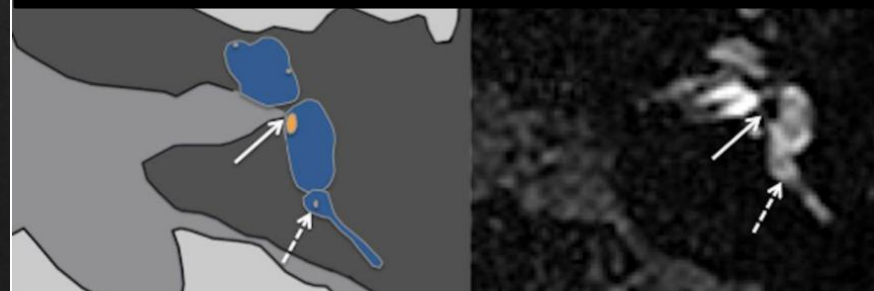


Level of the saccule and the posterior ampullae.

Typical aspect of vestibular atelectasis



On the level of the lateral canal:
the utricular space disappear, as the
lateral ampullae.



On the level of the saccule, the
collapse of the posterior canal while
the saccule space is normal.

We realized that many patients

=> were likely misdiagnosed as having endolymphatic hydrops or vestibular migraine,

=> when in fact,

they may have had vestibular utricular atelectasis

This led us to ask: which patients are at risk?

Clinical characteristics in **unilateral vestibular** atelectasis.

Marc M, Hautefort C, Guichard JP, Herman P, Houdart E, Toupet M, Eliezer M.

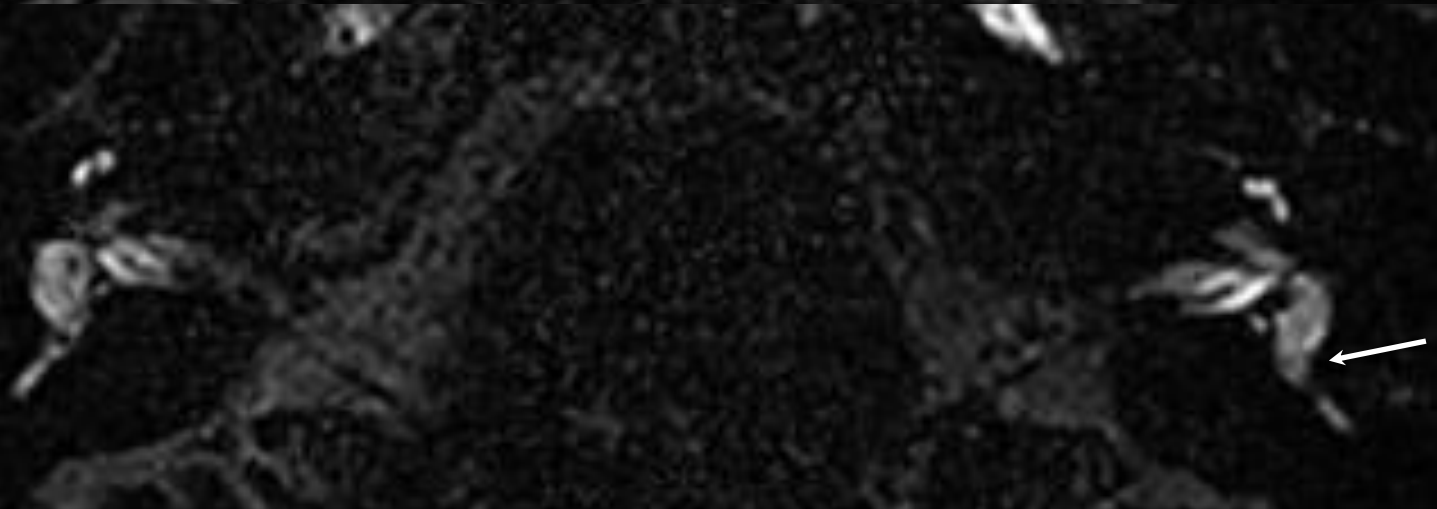
J Neurol. 2021 Feb;268(2):689-700. doi: 10.1007/s00415-020-10220-y. Epub 2020 Sep 9.

PMID: 32909094

Population

- ♦ 22 patients diagnosed with Unilateral Vestibular Atelectasis on MRI (SR=1,44)
- ♦ Mean age of 58.6 ± 13.7 years
- ♦ Four patients reported a history of fluctuating dizziness in a close family member.

MRI Criteria

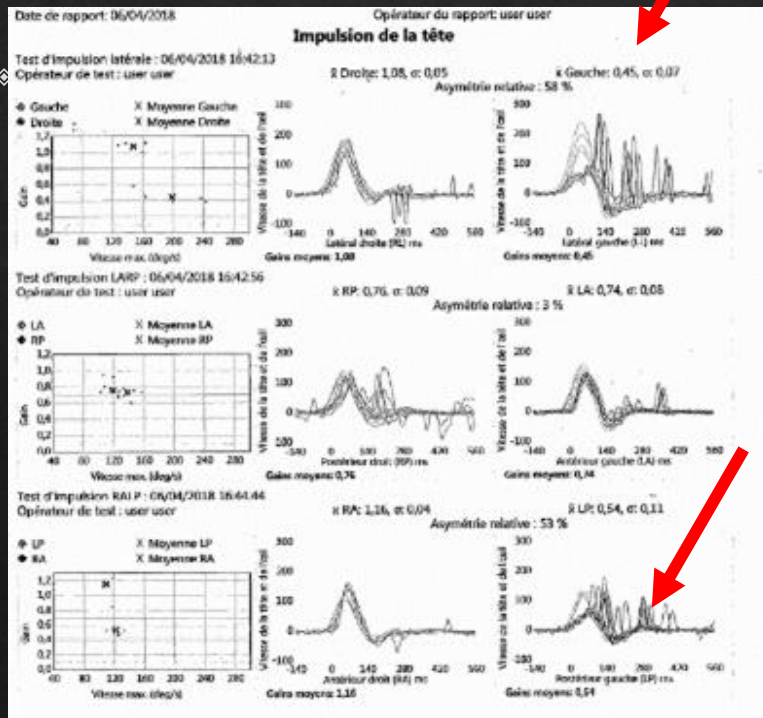


- ◆ According to Schucknecht observation and histopathological description, diagnostic criteria to assess vestibular atelectasis were defined as:
 - endolymphatic collapse,
 - including the utricle and at least two ampullae

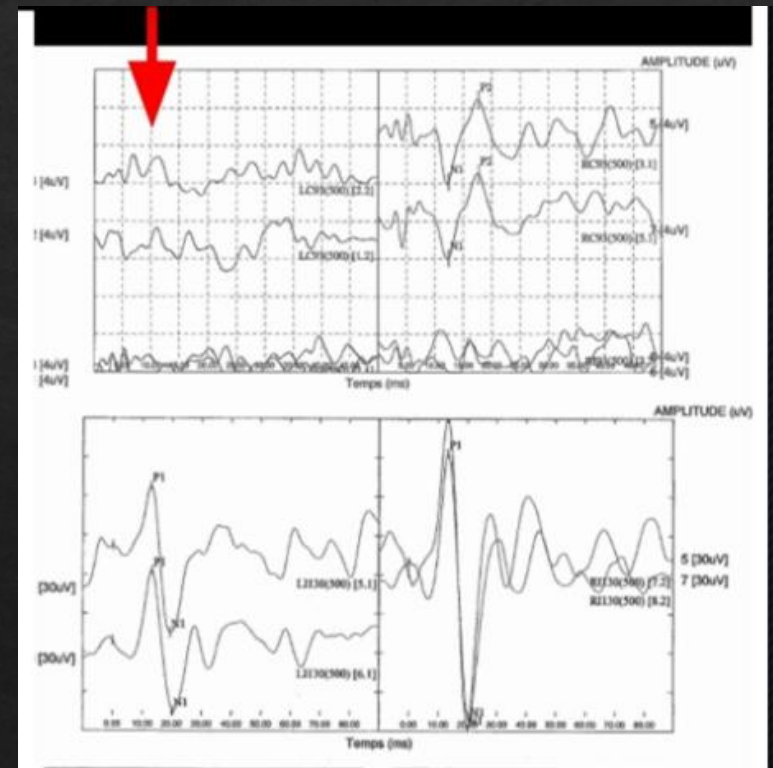
Otoneurological testing

- **Utricular involvement:** Present in 82% of subjects
- **Video head impulse test deficits:** Consistent with imaging findings
- **Saccular and audiometric tests:** Preserved

VHIT => all patients had a canal deficit



oVEMP deficit => 82%



Clinical presentation

VESTIBULAR SYMPTOMS 100%

Fluctuating vertigo <20min =86%

Dizziness =14%

Sudden onset = 73%

Tullio 9%

NO HEARING LOSS

Fullness/Tinnitus 54%

Mean pure-tone audiometry (PTA)
level in the affected ears = 22 ± 19 dB,

Mean PTA level in the contralateral
ears was 23 ± 26 dB, $p=0.69$.

From a unilateral disease to a bilateral vestibular disease

Thanks to our clinical and radiological experience we suspected vestibular atelectasis to be one of the causes of Idiopathic Bilateral Vestibulopathy.

To assess our hypothesis we proposed to the patients of the french bilateral vestibulopathy association to perform the 3D flair delayed MRI.

Electrophysiological and inner ear MRI findings in patients with **bilateral vestibulopathy**.

Eliezer M, Hautefort C, Van Nechel C, Duquesne U, Guichard JP, Herman P, Kania R, Houdart E, Attyé A, Toupet M.

Eur Arch Otorhinolaryngol. 2020 May;277(5):1305-1314. doi: 10.1007/s00405-020-05829-8. Epub 2020 Feb 8.

PMID: 32036409

Population

- ◆ 42 patients were included
- ◆ Bilateral vestibulopathy according to the Barany criteria

RESULTS

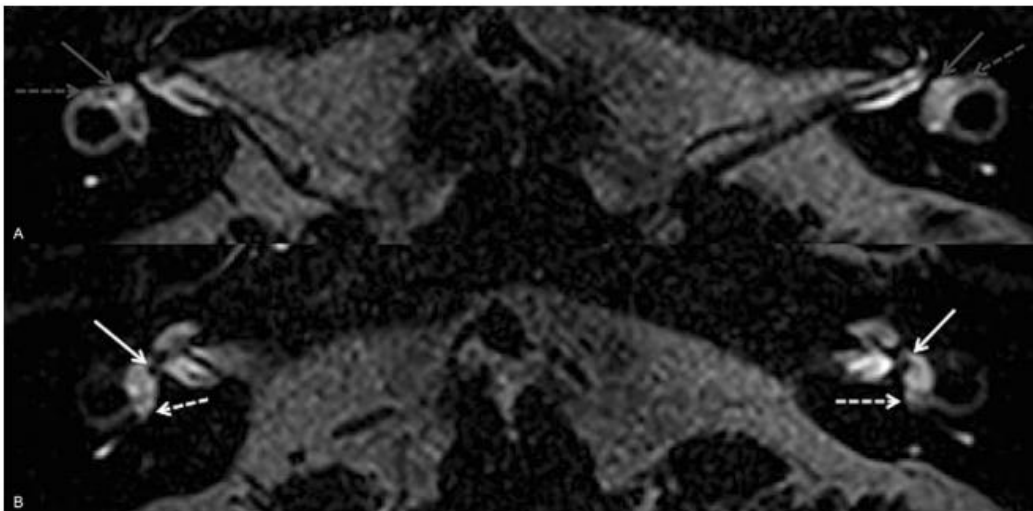
◆ MRI 3D FLAIR delayed .

Three MRI abnormalities were found for 59,6% of the patients:

- Endolymphatic abnormalities
- Impairment of the blood labyrinthine barrier
- Morphological disorder

Endolymphatic space abnormalities

- ◆ **Bilateral vestibular atelectasis (utricular and ampullae compartment) was found for 21 patients (50%) associated with oVEMP and VHIT deficits**
 - ▢ Saccular space and cVEMP responses were still normal for all of them
 - ▢ no hearing complains..



Etiology

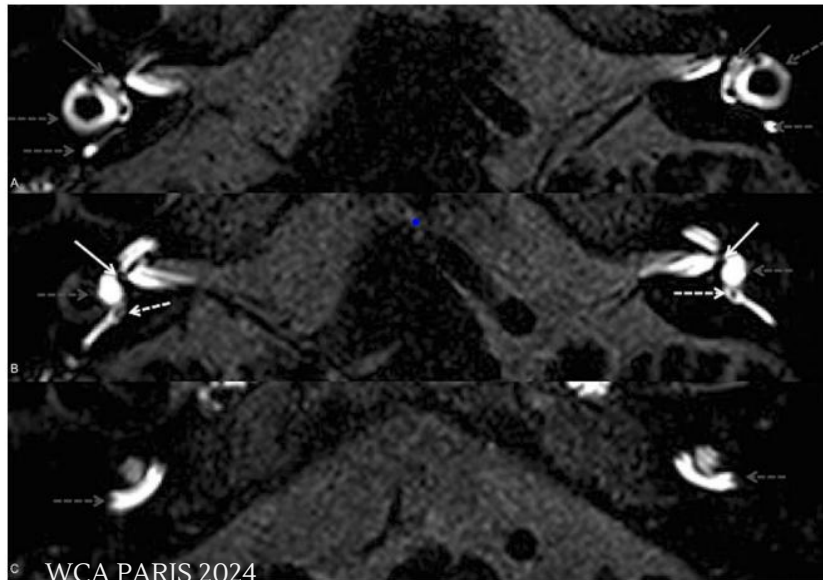
- ▢ Idiopathic (n = 19)
- ▢ Post aminoglycoside (n = 1)
- ▢ Post abdominal surgery (n = 1)

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Blood–labyrinthine barrier impairment

- ❖ Five patients had an impairment of the blood labyrinthine barrier
- ❖ They had both hearing and vestibular loss



Etiology:

- ❖ lymphome (n = 1),
- ❖ auto-immun (n = 3)

Take home message

- ◆ Recent 3D-FLAIR delayed MRI highlights the pars superior atelectasis described by Schuknecht in 1988.
- ◆ we must first recognize and identify vestibular utricular atelectasis.
- ◆ This may be a pathophysiological explanation for fluctuating vestibular disorders leading, after a few years, to unilateral or bilateral utricular and canal deficits.
- ◆ We do not know yet why patients develop vestibular atelectasis.
- ◆ Management of this condition probably differs from other endolymphatic pressure-related disorders.

Merci beaucoup pour votre
attention!

