

## Case Report

# Transient Vertigo with Horizontal Nystagmus to Loud Noise and Pressure: Utricular Hydrops or Vestibular Atelectasis?

Fatemeh Hassannia , Simon D. Carr , John A. Rutka 

Department of Otolaryngology, Head and Neck Surgery, Toronto General Hospital, University Health Network, University of Toronto, Toronto, Canada

ORCID iDs of the authors: F.H. 0000-0003-2358-9738; S.C. 0000-0001-8863-8022; J.A.R. 0000-0002-4318-8467.

Cite this article as: Hassannia F, Carr SD, Rutka JA. Transient Vertigo with Horizontal Nystagmus to Loud Noise and Pressure: Utricular Hydrops or Vestibular Atelectasis? J Int Adv Otol 2020; 16(1): 127-9.

We present an unusual case of a patient with a positive Tullio phenomenon, brief Valsalva-induced transient horizontal nystagmus, reduced left caloric response, and bilateral vestibulo-ocular reflex loss. This study discusses the pathophysiology and differential diagnosis concerning the suspected pathology for the phenomenon of utricular hydrops or vestibular atelectasis and presents a literature review.

**KEYWORDS:** Horizontal nystagmus, hydrops, utricle

## INTRODUCTION

The Tullio phenomenon consistent with sound-induced vertigo and the corresponding evoked eye movement was first described in 1929 by the Italian biologist Pietro Tullio <sup>[1]</sup>. Since then, clinical studies have identified this phenomenon in several diseases, such as congenital deafness, Meniere's disease, perilymphatic fistula, head trauma, and superior canal dehiscence syndrome.

Different theories have been proposed to explain the mechanism underlying this phenomenon. The close anatomical relationship between stapes footplate and saccule seems to play a great role. This case report presents a patient with a positive Tullio phenomenon, Valsalva induced horizontal nystagmus, bilateral vestibulo-ocular reflex (VOR) loss and a reduced caloric response. The pathophysiological theory has been discussed and the relevant literature has been reviewed.

## CASE PRESENTATION

A 49-year-old male patient presented at our multidisciplinary neurotology clinic in October 2014 with a 1-year history of disequilibrium exacerbated by a forced Valsalva maneuver and loud noise, which resulted in a transient horizontal pulsion toward his right side. He also described experiencing episodic attacks of vertigo lasting from minutes to hours, which was associated with a sensation of left aural fullness. There were no complaints of tinnitus or hearing loss. He denied any history of headache, including migraine. Five months prior to his presentation at clinic, he had been diagnosed with right posterior canal benign paroxysmal positional vertigo (BPPV) and had responded well to the Epley maneuver. His past otologic history had been unremarkable otherwise.

Neuro-otologic examination demonstrated volleys of rapid transient horizontal nystagmus (Video 1) during the Valsalva maneuver. Dynamic visual acuity testing using a logMAR chart demonstrated a 4-line loss in the horizontal plane. No spontaneous or gaze-evoked nystagmus was detected. Smooth pursuit and saccades were normal. Fundoscopic examination of the eyes was normal. He was able to suppress the VOR bilaterally. There was no post-headshake nystagmus. Gait and hemispheric cerebellar testing were normal. The fistula test was negative.

Pure tone audiometry revealed normal hearing with normal stapedial reflexes. Electrocochleography (Biologic Navigator-Pro, San Carlos, CA USA) with and without straining was normal. Videonystagmography (I-Portal VOG by Neurokinetics, Pittsburgh, US) revealed an absent caloric response on the left ear. Video head impulse test (ICS Impulse by Otometrics) demonstrated bilateral VOR

loss with consistent covert saccades (more so on the left side). There were reduced gains throughout the semicircular canals (SCCs), particularly in the left lateral and posterior canals. Gains were measured as follows: lateral SCC's-left 0.45, right 0.57; posterior SCC's-left 0.51, right 0.68; and superior SCC's-left 0.66, right 0.74. Vestibular-evoked myogenic potential (VEMP) testing (ChartR-EP by Otometrics) revealed reduced amplitude and increased threshold responses on the left cervical VEMP testing compared with those on the right side. Ocular VEMP was absent on the left but present on the right side.

High-resolution computed tomography (CT) of the temporal bones revealed some thinning of the bone covering the superior SCC's, with no evidence of a dehiscence. The findings of high-resolution intracranial magnetic resonance imaging (MRI) were unremarkable. Serological investigations, including syphilis serology and anti-neutrophil cytoplasmic antibody tests, were normal.

## DISCUSSION

The phenomenon of vestibular responses to sound transmitted to the inner ear was initially described by Tullio<sup>[1]</sup>. Different theories have been proposed and post-mortem temporal bone studies have been performed to explain this phenomenon<sup>[2]</sup>. Physiologic- and histopathologic-based theories for this phenomenon have included stapes hypermobility<sup>[3]</sup>, fibrous attachments between the saccule/utricle and undersurface of the stapes footplate (vestibulofibrosis)<sup>[2,4]</sup>, hydropic changes involving the saccule, a third window phenomenon (from otic capsule softening or a canal dehiscence)<sup>[5]</sup>, and vestibular atelectasis<sup>[6]</sup>. Theories promoting saccular hydrops as a cause of Tullio phenomenon are mainly based on the close natural proximity between the saccule and stapes footplate. Hydropic changes within the saccule might be all that is necessary for more direct physical transmission of mechanical energy from stapes footplate excursion<sup>[4]</sup>.

Clinical differential diagnoses in patients with a positive Tullio phenomenon include the consideration of a perilymphatic fistula<sup>[7]</sup>, lateral SCC erosion from cholesteatoma<sup>[8]</sup>, congenital syphilis<sup>[9]</sup>, Meniere's disease, and superior SCC dehiscence. The differential diagnoses that were refuted in this patient included a fistula involving the horizontal SCC and superior SCC dehiscence, neither of which was evident on the CT of temporal bones. Normal syphilis serology ruled out a luetic infection. There was no history of a previous otologic surgery, barotrauma, or head trauma suggestive of a perilymphatic fistula. The differential diagnosis would have also included vestibular migraine, hindbrain abnormalities (i.e., Chiari malformation), and raised intracranial pressure (i.e., idiopathic intracranial hypertension). However, there was no history of headache or symptoms suggestive of raised intracranial pressure. Anatomical hindbrain pathologies were excluded based on normal intracranial MRI. Fundoscopic examination did not reveal features of increased intracranial pressure or papilledema.

We speculate that the constellation of a positive Tullio phenomenon and other symptoms (recurrent attacks of episodic vertigo lasting from minutes to hours associated with left aural fullness) in this patient are reasonably explained by isolated utricular hydrops resulting in relative fixation of the cupula of the lateral SCC. This could explain the reduced caloric response on the left side. Movement of the stapes footplate associated with the Valsalva maneuver or loud noise would be expected to cause transient ampullofugal movement of

the lateral canal cupula juxtaposed to an enlarged utricle, resulting in transient horizontal nystagmus. Alternatively, the response might have been generated within the utricle. The underlying condition of suspected hydrops might also explain the presence of a bilateral VOR impairment. Bilateral VOR involvement is not infrequently observed in Meniere's disease over time. Concomitant BPPV is also not infrequently observed in Meniere's disease (although it was present on the right and not on the left side in our patient). Whether there might be an ultimate evolution to classic Meniere's disease over time remains to be determined.

The other relevant consideration for this presentation might include the phenomenon of vestibular atelectasis, which results from collapse of the ampulla and utricle. Due to the proximity of vestibular end organs to the center-point of the stapes footplate, any condition causing changes in the soft tissue connection between the stapes footplate and vestibular end organ can produce a vestibular reaction to sound. It has been hypothesized that in the event of collapse of the membranous labyrinth, the utricle and saccule are brought in contact with the footplate. Upon increased excursion of the stapes to loud noise, both end organs might be stimulated<sup>[6]</sup>. While the histopathology of vestibular atelectasis has been well documented in a series of temporal bones by Merchant et al.<sup>[10]</sup>, unfortunately no documentation of vertigo being produced by sound or pressure changes upon clinical correlation prior to necropsy is available.

On vestibular testing, vestibular atelectasis would be anticipated to result in an absent caloric response and reduced VOR gain, similar to that present in this patient. An abnormal VEMP study in the absence of a conductive hearing loss suggests an otolithic abnormality possibly due to a hydropic or atelectatic cause.

Currently, there is no definitive means to prove the diagnosis for either vestibular atelectasis or utricular hydrops. Both these conditions might be diagnosed during a post-mortem histopathologic study of the temporal bone. Another possible consideration is whether MRI following intratympanic contrast enhancement reveals findings suggestive of hydrops. Variable contrast enhancement of labyrinthine fluids has already been used to visualize hydrops in patients with Meniere's disease<sup>[11]</sup> and may also be a potential way to diagnose vestibular atelectasis additionally<sup>[6]</sup>.

## CONCLUSION

We presented the case of transient sound- and/or pressure-induced horizontal eye movement with bilateral vestibular hypofunction and normal hearing. We suggest that this presentation is explained either by the phenomenon of utricular hydrops or vestibular atelectasis.

**Informed Consent:** Informed consent was obtained from the patient who participated in this study.

**Peer-review:** Externally peer-reviewed.

**Author Contributions:** Concept - J.R.; Design - J.R.; Supervision - J.R.; Resource - J.R.; Materials - J.R.; Data Collection and/or Processing - S.C., F.H.; Analysis and/or Interpretation - J.R.; Literature Search - F.H.; Writing - F.H., S.C.; Critical Reviews - J.R.

**Acknowledgements:** The authors would like to thank Ms. Sulway for her wonderful collaboration in recording eye movements.

**Conflict of Interest:** The authors have no conflict of interest to declare.

**Financial Disclosure:** The authors declared that this study has received no financial support.

**Video 1.** Neuro-otologic examination demonstrated volleys of rapid transient horizontal nystagmus (attached video) during the Valsalva maneuver.

## REFERENCES

1. Tullio P. Das Ohr und die Entstehung der Sprache und Schrift. Berlin, Germany: Urban & Schwarzenberg, 1929.
2. Nadol JB Jr. Positive “fistula sign” with intact tympanic membrane. *Arch Otolaryngol* 1974; 100: 273-8. [\[Crossref\]](#)
3. Dieterich M, Brandt TH, Fries W. Otolith function in man. *Brain* 1989; 112: 1377-92. [\[Crossref\]](#)
4. Backous DD, Aboujaoude SE, Minor BL, Nager TG. Relationship of the utricle and saccule to the stapes footplate: anatomic implications for sound-and/or pressure-induced otolith activation. *Ann Otol Rhinol Laryngol* 1999; 108: 548-53. [\[Crossref\]](#)
5. Kacker SK, Hinchcliffe R. Unusual Tullio phenomena. *J Laryngol Otol* 1970; 84: 155-66. [\[Crossref\]](#)
6. Wenzel A, Ward BK, Schubert MC, Kheradmand A, Zee DS, Mantokoudis G, et al. Patients with vestibular loss, Tullio phenomenon, and pressure-induced nystagmus: Vestibular Atelectasis? *Otol Neurotol* 2014; 35: 866-72. [\[Crossref\]](#)
7. Fox EJ, Balkany TJ, Arenberg IK. The Tullio phenomenon and perilymph fistula. *Otolaryngo Head Neck Surg* 1988; 98: 88-9. [\[Crossref\]](#)
8. Ishizaki H, Pyrkko I, Aalto H, Strack J. Tullio phenomenon and postural stability: experimental study in normal subjects and patients with vertigo. *Ann Otol Rhinol Laryngol* 1991; 100: 976: 83. [\[Crossref\]](#)
9. Karmody CS, Schuknecht HF. Deafness in congenital syphilis. *Arch Otolaryngol* 1966; 83: 18-27. [\[Crossref\]](#)
10. Merchant S, Schuknecht HF. Vestibular atelectasis. *Ann Otol Rhinol Laryngol* 1988; 97: 565-76. [\[Crossref\]](#)
11. Naganawa S, Yamazaki M, Kawai H, Bokura K, Sone M, Nakashima T. Visualization of endolymphatic hydrops in Meniere’s disease after single-dose intravenous gadolinium-based contrast medium: timing of optimal enhancement. *Magn Resonance Med* 2012; 11: 43-51. [\[Crossref\]](#)