Case Report/Case Series

Head-Jolting Nystagmus
Occlusion of the Horizontal Semicircular Canal Induced by Vigorous Head Shaking

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IMPORTANCE We report a new syndrome, which we are calling *head-jolting nystagmus*, that expands the differential diagnosis of head movement–induced paroxysmal vertigo.

OBSERVATIONS Two male patients (65 and 58 years old) described rotational vertigo after violent and brief (1- to 2-second) oscillations of the head (head jolting) that triggered intense horizontal nystagmus lasting 45 seconds. Accelerations of the head required to induce these episodes could only be achieved by the patients themselves. In case 1, the episodes gradually disappeared over a 6-year period. In case 2, magnetic resonance imaging (3-T) suggested a filling defect within the left horizontal semicircular canal. He underwent surgical canal plugging in March 2013 that resolved the symptoms.

CONCLUSIONS AND RELEVANCE We attribute head-jolting nystagmus to dislodged material within the horizontal semicircular canal and provide a mechanistic model to explain its origin.

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The differential diagnosis of paroxysmal vertigo induced by head movements includes benign paroxysmal positional vertigo (vertigo and nystagmus caused by changes in head position[1]) and head-shaking nystagmus (a jerk nystagmus that follows prolonged sinusoidal head oscillations), often encountered in patients with unilateral vestibular lesions.[2] Vestibular paroxysmia, presumably neurovascular compression of the vestibular nerve,[3] also causes paroxysmal vertigo but is not unequivocally triggered by head movement.

We report 2 cases of high-velocity horizontal nystagmus following a unique maneuver consisting of horizontal high-acceleration head oscillations elicited by the patients. We suggest the term *head-jolting nystagmus* for this phenomenon.

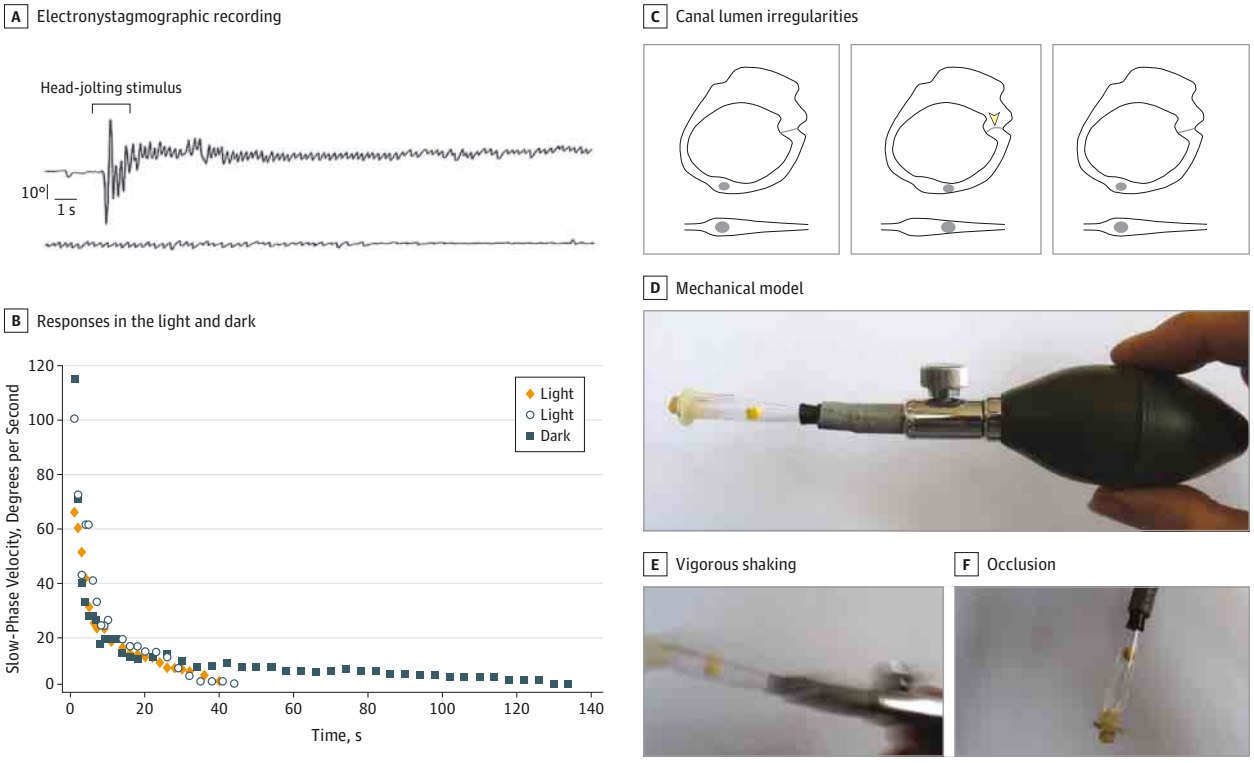
Report of Cases

Case 1 and Case 2

The clinical history and examination of the 2 patients were virtually identical and are summarized together. Case 1 was a 65-year-old man, and case 2 was a 58-year-old man. They both discovered that brief but fast horizontal head shaking triggered horizontal vertigo and oscillopsia. Initially, this was noticed during emphatic shaking of the head in disagreement or during energetic ballroom dancing. No other significant medical history or findings were observed on examination, apart from intense left-beating horizontal nystagmus self-induced by vigorous horizontal head shaking (jolting), shown in **Video 1** and **Video 2**. Two to three head oscillations were sufficient to induce the nystagmus, which lasted 30 to 45 seconds and did not fatigue (the patient in case 1 elicited it 7 times consecutively). This could not be triggered by the examiners (A.M.B., D.K., and N.C.) with vigorous head impulse or head positioning testing for any canal, although video-oculography was not performed during positioning maneuvers. Normal head shaking for eliciting head-shaking nystagmus[2] failed to elicit nystagmus. Indeed, the patient in case 1 was instructed to make progressively brisker head shakes, and the nystagmus was only triggered when the oscillations were of approximately ±10-degree amplitude and reached a mean peak velocity of greater than 700 degrees per second (acceleration, 4500 degrees per second squared). Both cases showed a left canal paresis on caloric testing (additional vestibular testing is described below).

Findings from high-resolution temporal bone computed tomography and 1.5-T magnetic resonance imaging were normal in both cases. The symptom in case 1 remained unchanged for 2 years, but triggering of the nystagmus became progressively more difficult and then impossible over a 2-year period. He has remained free of the symptom for 2 years. The patient in case 2 underwent 3-T magnetic resonance imaging, which showed a filling defect in the posterior limb of the left horizontal semicircular canal that appeared to move with head shaking (eFigure in the Supplement). While filling defects within the semicircular canal have been reported with intrac-

Figure. Electronystagmography and a Putative Mechanistic Model of Head-Jolting Nystagmus in Case 1



A, Electronystagmographic recording of *head-jolting nystagmus*, a new syndrome we have so named and now report on. Following a brief but vigorous head oscillation, the patient develops left-beating horizontal nystagmus lasting 45 seconds. B, Slow-phase nystagmic responses following 2 separate head-jolting stimuli in the light and 1 in the dark. C, On the left, a canalith is freely mobile within the horizontal semicircular canal, which has a tapered distal end. In the middle, a head-jolting stimulus causes the canalith to jam toward the tapered section of the canal, which thus exerts pressure on the endolymph within the canal and induces deflection of the cupula (arrowhead). This induces excitatory nystagmus. On the right, the kinetic energy accumulated after distal endolymphatic pressure and cupular deflection pushes back and unplugs the canal. D, Mechanical model related to **Video 3**. A freely mobile particle or canalith (yellow) is lodged within the horizontal semicircular canal (plastic tubing), which has a tapered end (the hand pump attached only serves the purpose of puffing in air to unjam the canalith from the tapered end of the tubing). E and F, After vigorous shaking of the device (E), the canalith jams up the tapered end (F). See **Video 3**.

table benign paroxysmal positional vertigo,[4] these have not been shown to move within the canal. Numerous attempts at treating the symptoms using repositioning maneuvers and mastoid vibration were unsuccessful. He underwent left-sided surgical plugging of the horizontal semicircular canal as a last resort in March 2013, and the vertigo and nystagmus immediately and permanently disappeared (**Video 2**).

Audiovestibular Testing

In case 1, a raw recording of the head-jolting nystagmus is shown in panel A in the **Figure**. Panel B shows plots of its slow-phase eye velocity, demonstrating longer duration in the dark. The overall time constant of decay of the ocular response was 12 seconds. Bithermal calorics revealed bilateral hypofunction (peak slow-phase velocities of 6.5 degrees per second for the right ear and 3.9 degrees per second for the left ear at 44 degrees), with a pattern of a left canal paresis (23%-88% on 4 different tests). Horizontal chair rotation with velocity steps of 60 degrees per second confirmed bilateral hypofunction (time constants of 4-5 seconds). The horizontal left head impulse test was clinically inconsistently abnormal (video head impulse test gain, 0.57) but was normal for all other canals. Cervical vestibular-evoked myogenic potentials were normal.

In case 2, bithermal caloric testing demonstrated a significant left-sided canal paresis (44%) and normal function on the right side. Cervical vestibular-evoked myogenic potentials were absent on the left. A video head impulse test recording was performed 2 years after surgery, and showed reduced VOR gain on the left, with a mean (SD) of 0.29 (0.05) on the left vs 0.90 (0.11) on the right at 80 milliseconds.

Pure-tone audiography and tympanometry were also performed. The findings were normal for age in both cases.

Discussion

We report 2 cases of horizontal nystagmus following brief, self-imposed head-jolting stimuli. Both cases had left-beating nystagmus and additional evidence of left labyrinthine disease (canal paresis in both cases and absent cervical vestibular-evoked myogenic potentials in case 2), strongly suggesting that the nystagmus was excitatory, originating in the left (paretic) horizontal semicircular canal. This was further supported in case 2 by the presence of a canal-filling defect on 3-T magnetic resonance imaging and was finally confirmed by successful surgical plugging of the canal.

Established vestibular disorders cannot explain this clinical picture. The head oscillations required to induce nystagmus in our cases (4-5 Hz, 700 degrees per second, and acceleration >4500 degrees per second squared) are 2 to 3 times larger than those in conventional head-shaking nystagmus (1-2 Hz and 300 degrees per second).[2] The head-jolting stimulus was also considerably faster than that associated with a positional maneuver (peak angular velocity, 250 degrees per second)[5] or head impulse test (acceleration, 3000-4000 degrees per second squared)[6] that never triggered the nystagmus. While vestibular paroxysmia causes intense but brief paroxysms of nystagmus and oscillopsia, radiological[7] and surgical[8] evidence (as well as the positive response to carbamazepine) suggests pathology within the vestibular nerve and not the semicircular canal. Finally, Merchant and Schuknecht[9] described 8 patients with vestibular atelectasis, defined histopathologically as a collapse of the semicircular canal walls protruding within the canal lumen. While atelectasis remains a possible etiology, vertigo related to head movement was described in only one patient in their series.

The clinical syndrome that most approximates the head-jolting nystagmus described herein is a rare and poorly documented phenomenon called canal jam.[10] Such patients experience benign paroxysmal positional vertigo, but (usually in the course of a repositioning maneuver) the canalith jams in the canal, which leads to long-lasting vertigo and nystagmus. A recently described patient developed a reversible caloric canal paresis during the jam.[11] Although the patients in our cases did not manifest the canalith jam syndrome (the clinical presentation was different, and our patients never had symptoms or findings of benign paroxysmal positional vertigo), similar underlying anatomical mechanisms may be at play. Our cases had objective evidence of unilateral (case 2) and bilateral asymmetric (case 1) vestibulopathy, which would predispose to the appearance of debris that may lie loose in the canal lumen. Equally, the finding that high accelerations were critical in triggering the nystagmus suggests that if a canalith inside the canal was responsible for the symptoms, its size, weight, or adhesiveness would be large. In this light, the head jolting would transiently jam the canalith into the distal (cupular) arm of the canal, thus generating increased distal pressure, cupular deflection, and vestibular excitation. Once the external forces (jolting) settle down, the kinetic energy accumulated as distal endolymphatic pressure and cupular deflection would unplug the canal. Even if this internal plugging of the canal lasted a few seconds, the nystagmus durations observed (30-45 seconds) can easily be explained by the slow return of the cupula to its resting position (time constant, approximately 4-5 seconds)[12] and by the fact that the vestibulo-ocular response would be even further prolonged by the brainstem integrator termed the *velocity storage mechanism* (time constant, approximately 14 seconds).[13]

One intriguing observation is that the nystagmus occurs only in one direction considering that a head-jolting stimulus could also shift the canalith in a proximal direction. However, if the distal end of the canal was tapered as a result of irregularities in the canal lumen (Figure, C), jamming could only occur toward the tapered end. Still photographs (Figure, D-F) and **Video 3** of a putative mechanical model show that shaking the device jams the freely moving object (canalith) inside the device (semicircular canal) only toward the tapered canal end.

Conclusions

We propose a new syndrome of vertigo, nystagmus, and oscillopsia following high-acceleration horizontal head oscillations, with excitatory horizontal nystagmus possibly resulting from a mobile obstructive mass within the horizontal semicircular canal. We call this condition head-jolting nystagmus and suggest that a head-jolting maneuver should be performed in patients with paroxysmal vestibular disorders of unknown origin.

ARTICLE INFORMATION

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REFERENCES

1. Parnes LS, McClure JA. Posterior semicircular canal occlusion for intractable benign paroxysmal positional vertigo. *Ann Otol Rhinol Laryngol*. 1990;99(5, pt 1):330-334.

2. Hain TC, Fetter M, Zee DS. Head-shaking nystagmus in patients with unilateral peripheral vestibular lesions. *Am J Otolaryngol*. 1987;8(1):36-47.

3. Brandt T, Dieterich M. VIIIth nerve vascular compression syndrome: vestibular paroxysmia. *Baillieres Clin Neurol*. 1994;3(3):565-575.

4. Dallan I, Bruschini L, Neri E, et al. The role of high-resolution magnetic resonance in atypical and intractable benign paroxysmal positional vertigo: our preliminary experience. *ORL J Otorhinolaryngol Relat Spec*. 2007;69(4):212-217.

5. Faldon ME, Bronstein AM. Head accelerations during particle repositioning manoeuvres. *Audiol Neurootol*. 2008;13(6):345-356.

6. Aw ST, Halmagyi GM, Haslwanter T, Curthoys IS, Yavor RA, Todd MJ. Three-dimensional vector analysis of the human vestibuloocular reflex in response to high-acceleration head rotations, II: responses in subjects with unilateral vestibular loss and selective semicircular canal occlusion. *J Neurophysiol*. 1996;76(6):4021-4030.

7. Best C, Gawehn J, Krämer HH, et al. MRI and neurophysiology in vestibular paroxysmia: contradiction and correlation. *J Neurol Neurosurg Psychiatry*. 2013;84(12):1349-1356.

8. Reuter W, Fetter M, Albert FK. Vestibular paroxysmia: a rare but important differential diagnosis [in German]. *HNO*. 2008;56(4):421-424.

9. Merchant SN, Schuknecht HF. Vestibular atelectasis. *Ann Otol Rhinol Laryngol*. 1988;97 (6, pt 1):565-576.

10. Fife TD. Benign paroxysmal positional vertigo. *Semin Neurol*. 2009;29(5):500-508.

11. Ko KM, Song MH, Kim JH, Shim DB. Persistent spontaneous nystagmus following a canalith repositioning procedure in horizontal semicircular canal benign paroxysmal positional vertigo. *JAMA Otolaryngol Head Neck Surg*. 2014;140(3):250-252.

12. Dai M, Klein A, Cohen B, Raphan T. Model-based study of the human cupular time constant. *J Vestib Res*. 1999;9(4):293-301.

13. Büttner U, Waespe W. Vestibular nerve activity in the alert monkey during vestibular and optokinetic nystagmus. *Exp Brain Res*. 1981;41(3-4):310-315.

CORRECTION

Misspelled Author Name: In the Original Investigation titled "Percutaneous Ethanol Injection vs Reoperation for Locally Recurrent Papillary Thyroid Cancer: A Systematic Review and Pooled Analysis" published in the June 2015 issue of *JAMA Otolaryngology–Head & Neck Surgery*,[1] an author's name was misspelled. The correct spelling is Ahmed Deniwar, MD. The article has been corrected online.

1. Fontenot TE, Deniwar A, Bhatia P, Al-Qurayshi Z, Randolph GW, Kandil E. Percutaneous ethanol injection vs reoperation for locally recurrent papillary thyroid cancer: a systematic review and pooled analysis. *JAMA Otolaryngol Head Neck Surg*. 2015;141(6):512-518.