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Horizontal semicircular canal jam: Two new cases and possible mechanisms

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Abstract

Introduction: Benign paroxysmal positional vertigo (BPPV) of the horizontal semicircular canal (hSCC) can present with otoconia blocking its lumen (canalith jam), with signs and symptoms that make it difficult to distinguish from central nervous system pathology.

Objective: Here we report two cases of canalith jam affecting the hSCC and offer a theoretical mechanism based on known vestibular neurophysiology.

Methods: We use video-oculography to document the canalith jam and show the moment the otoconia loosen.

Results: Canalith jam is a rare form of BPPV remedied with repositioning maneuvers.

Conclusion: Clinicians should consider canalith jam as a mechanism for BPPV when the nystagmus is (a) Direction fixed with fixation removed and during positional testing; (b) Velocity dependent on supine head position; (c) Converts to geotropic directional changing nystagmus.

1 | INTRODUCTION

Horizontal semicircular canal (hSCC) benign paroxysmal positional vertigo (BPPV) accounts for 22% of all BPPV, with paroxysmal nystagmus during positional testing being geotropic in 70%, apogeotropic in 28%, and direction-fixed in 1.8%.[1] The diagnosis of hSCC BPPV is based on history of episodic positional vertigo triggered with positional changes of the head relative to gravity, typically bi-directional changing horizontal positional nystagmus during the supine roll test (SRT or Pagnini-McClure Test) and often during the bow and lean test.[2] Four

types of hSCC BPPV have been described based on the pattern of positional nystagmus during the SRT. Canalithiasis presents with an asymmetric decaying geotropic nystagmus with an increased slow-phase velocity when positioned on the affected side; cupulolithiasis presents with a sustained asymmetric apogeotropic nystagmus with a decreased slow-phase velocity when positioned on the affected side; anterior segment or "short-arm" canalithiasis presents with a short duration, asymmetric apogeotropic nystagmus with a decrease in intensity when positioned on the affected side; and canalith jam presents with a horizontal, direction-fixed nystagmus irrespective of head position.[3-5] Of these subtypes of hSCC BPPV, canalith jam with its direction-fixed nystagmus is most difficult to distinguish from central nervous system pathology.

The mechanism of direction-fixed paroxysmal nystagmus hSCC-BPPV has been attributed to a canalith jam creating a partial or complete obstruction within the lumen of the hSCC. It is hypothesized that as the debris moves from a larger to a narrower portion of the hSCC, the otoconia occlude the lumen (thereby becoming "jammed"). An obstruction within the lumen has also been suggested to occur spontaneously or as a complication following a canalith repositioning maneuver.[6] Hypothetically, the canalith jam may induce a prolonged utriculofugal deviation of the cupula from the horizontal semicircular (deflection away from the ampulla for an inhibitory stimulus) or a prolonged utriculopetal deviation of the cupula (deflection toward the ampulla for an excitatory stimulus).[7] Both would cause a nonfatiguing, spontaneous, horizontal nystagmus regardless of head position.

Geotropization is a term coined by Vannucchi & Pecci (2011) and implies the liberation of the occluding otoconia with an immediate change of the nystagmus from direction-fixed to geotropic.[4] Once converted to geotropic nystagmus, the BPPV may be treated using the barbeque roll (log roll) or Gufoni's liberatory maneuver.[8,9]

Persistent direction-fixed, spontaneous nystagmus consistent with canalith jam of the hSCC can be associated with ipsilateral canal paresis based on caloric testing[7,10]; the video head impulse test[11,12]; and asymmetrical cervical and ocular vestibular evoked myogenic potentials.[12] In each case in which vestibular function was reduced there was recovery within 30 days.[7,11,12]

2 | CASE DESCRIPTIONS

Here we present video evidence and description of two patients presenting with positional symptoms consistent with BPPV who also showed a peripheral, horizontal, unidirectional-fixed nystagmus irrespective of the head position suggesting canalith jam of the hSCC. In both cases, the canalith jam occurred spontaneously, presumably within the lumen of the hSCC. In patient 1, the cupula was presumably deflected in the inhibitory direction and a video shows the moment of geotropization. In patient 2 the cupula was presumably deflected in the excitatory direction. At time of follow-up, the patient showed a geotropic LC BPPV, presumably following the geotropization.

2.1 | Case 1

A 56-year old woman was evaluated for an inability to lie on her left side for the preceding 4 days due to a "strong vertigo." Prior to the onset of vertigo, she was in good health and she had no history of migraine or other balance disorders.

Clinical oculomotor exam revealed a spontaneous left beating nystagmus both with fixation and with fixation removed. She showed no change in direction or intensity of nystagmus with eccentric gaze. Her smooth pursuit and saccade testing was normal. The head impulse test was not performed at this time given the clinician's suspicion of BPPV. The patient underwent positional testing with the Dix-Hallpike Test (DHT). In both DHT, a left beat nystagmus was persistent though more intense in the left DHT. The SRT was then performed. In supine, she developed a left beat nystagmus with velocity similar to the nystagmus in upright position. In right roll, the left beat nystagmus initially reduced in intensity, however, after ~ 10 second mildly increased (15.2 deg/sec) and then settled to a velocity of ~ 6 deg/sec persisting beyond 100 seconds (Figure 1). In left roll, the left beating nystagmus increased robustly to 24 deg/sec before settling to 13 deg/sec persisting beyond 120 seconds (Video S1).

The supine roll test was repeated once more. In right roll, geotropization occurred. The left beating nystagmus changed to right beating nystagmus thus becoming geotropic in form. This geotropic

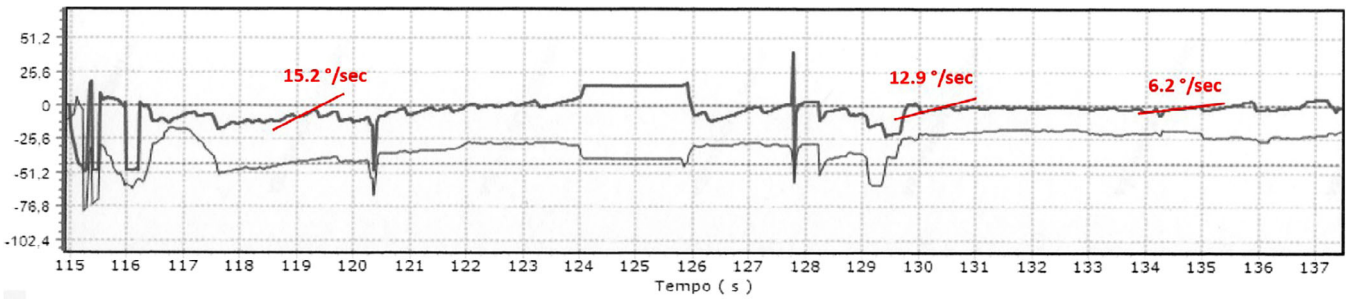

FIGURE 1 Apogeotropic nystagmus (left beat) in right side lie position. The slow-phase velocity gradually decreases before settling to a velocity of ~ 6 deg/sec that persists beyond 100 seconds. The different slopes of the red lines show the decaying slow eye velocity. The upper dark trace is the horizontal eye channel, the lower and lighter colored trace the vertical eye channel

nystagmus was confirmed in repeating the left roll position. The velocity/intensity of her nystagmus was greater in right roll (Video S1). Brief exposure to a fixation target suppressed the nystagmus in each roll position. The recently converted presentation of right hSCC canalithiasis was treated using modified Gufoni maneuver that resolved the BPPV.[9]

The patient went home and remained symptom free for 9 days. On the tenth day, she returned to the clinic reporting a return of the positional vertigo when laying on her right side. The intensity of the vertigo was less than her prior experience. Repeat oculomotor exam was normal, except for a mild left beat nystagmus following horizontal head shaking. She had no spontaneous nystagmus in upright with or without fixation. Positional testing revealed a slight left beat nystagmus in supine. Findings on the right DHT revealed an upbeat nystagmus with right torsion accompanied with a sense of vertigo. Findings suggested a right posterior semicircular canal canalithiasis. The patient was treated successfully with a liberatory maneuver for the posterior semicircular canal. The patient was seen 15 days later and was symptom free with no further recurrence.

2.2 | Case 2

A 63-year-old woman with no history of inner ear dysfunction was referred to outpatient physical therapy with complaints of intermittent dizziness and vomiting. Over 3 days, she experienced dizziness when she would rotate her head from right to left. As the dizziness became more intense, she would stop her activity and lie down. She noted the intensity of her dizziness varied while lying down—if she lay on her right side, she experienced dizziness, yet on her left side she did not. When supine, her dizziness was most severe. If the dizziness became very intense, she would vomit. This pattern repeated once per day for three consecutive days.

Upon clinical examination, she had no neurologic findings. An oculomotor examination conducted in room light was normal. When vision was denied with video-oculography, a strong, fixed, unidirectional, horizontal, right beating spontaneous nystagmus was observed in all positions of gaze and increased in right gaze. Oculomotor control was normal. The clinical head impulse test was not performed to avoid increasing symptoms. Positional testing using the Dix Hallpike, SRT, and the bow and lean test was performed with video-oculography. Regardless of positional testing, she had a strong unidirectional horizontal right beat nystagmus. In supine and head positions below the horizon, her right beat nystagmus included a right torsional component. She experienced the most intense dizziness in the neutral position of the SRT and the greatest relief with her head rotated ~ 70° toward the left. Upon return to upright from each position a right beat nystagmus was observed. Following positional testing, the individual became extremely nauseous and required Ondansetron (Zofran). She refused further treatment, went home and slept on her left side with her head slightly elevated. She phoned the therapist and reported her symptoms resolved within 4 hours.

Three days after the initial evaluation, the patient returned to clinic and reported she felt 90% better. She experienced a sensation of mild dizziness. On examination, she still had direction-fixed, right beat nystagmus with fixation removed. However, once she lay supine, her nystagmus changed to left beating, suggesting utriculofugal (inhibition) movement of the otoconia and endolymph within the right hSCC. SRT confirmed her positional nystagmus was low amplitude, geotropic, lasting 1 minute. The patient was treated with three cycles of the BBQ roll maneuver for the right hSCC and provided with activity restrictions (sleep on the left side and avoid horizontal head motion). Via phone, the patient reported that her vertigo resolved.

3 | DISCUSSION

We report here two cases of BPPV affecting the hSCC, documented with VOG that support the idea of canalith jam, in which a mass of otoconia block the lumen and create a persistently deflected cupula on the affected side. The deflection of the cupula is inhibitory for case 1 and excitatory for case 2. John Epley first suggested this possibility when he wrote, "*An interesting phenomenon that I have occasionally observed while undertaking the canalith repositioning procedure (CRP), CRP is a sudden conversion of transient nystagmus to a rapid form that persists irrespective of head position. Simultaneously, the patient usually complains of intense vertigo. I believe the mechanism to be a jamming of the canaliths…*".[6] Since John Epley first hypothesized canalith jam, we have found three articles describing a similar pattern of nystagmus.[1,4,11] We show here videographic evidence of geotropization (Video S1). The video-graphic evidence for canalith jam of the hSCC includes:

1. Direction fixed spontaneous nystagmus with fixation removed.
2. Direction fixed spontaneous nystagmus during positional testing.
3. The velocity of the positional nystagmus and intensity of vertigo depend on supine roll head position.
4. Conversion of unidirectional positional nystagmus to geotropic nystagmus.

We suggest these four observations help identify otoconia physically plugging (ie, jamming) the lumen of the hSCC, which are different from the more common types of hSCC BPPV in which otoconia are freely floating (canalithiasis) or adherent to the cupula (cupulolithiasis). In these more common types of hSCC BPPV, it remains possible that patients may have spontaneous nystagmus in an upright position that will change when moving from sit to supine and in the SRT, all of which can help determine the affected ear.[13,14] For example, an initial left beating spontaneous nystagmus in sitting position that becomes geotropic in the SRT suggests an affected right hSCC due to canalithiasis. In contrast, a left beating nystagmus in sitting that becomes apogeotropic in the SRT suggests the left hSCC as the affected ear due to cupulolithiasis. Additionally, the velocity of the induced direction-changing nystagmus can help identify the side of BPPV within the hSCC: the nystagmus will always be more intense (higher velocity) when it beats toward the affected labyrinth.

In both of our cases of right hSCC canalith jam, we offer two possibilities for the direction-fixed but velocity-dependent positional

nystagmus. One explanation is that the otoconial jam acts as a "false" cupula, which causes a modification in velocity but not direction of the nystagmus that depends on head position. In video case 1, nystagmus velocity increases when positioned on the unaffected (left) side because the jam (false cupula) exerts a greater magnitude of cupular deflection in the inhibitory direction (Figure 2). However, in video case 2, the canalith jam in the right hSCC deflects the cupula in the excitatory direction, which also changes velocity (not direction) depending on head position. In both cases, we propose that the excitation-inhibition asymmetry principle is followed, though not with a large enough magnitude to cause nystagmus reversal expected in the SRT. Instead, the velocity of the nystagmus changes. Regardless of direction of deflection, this evidence suggests the otolith jam is acting like a "false" cupula. It is interesting to note that like another syndrome in which the cupula is maintained in a sustained deviated position—magnetic vestibular stimulation, the nystagmus in canal jam does not completely adapt to zero.[15]

A second possibility involves a "double" pathology of jam and free-floating otoconia. Applying this hypothesis to our first case, the jam caused an initial inhibitory deflection of the cupula, but free-floating otoconia between the jam and the cupula cause a variable velocity though still direction-fixed nystagmus (Figure 3). Califano et al suggested two masses of otoconia, one within the ampullary and one within the non-ampullary arms of the lumen of the hSCC as an explanation for the unidirectional positional nystagmus that eventually converted to the geotropic form of BPPV (canalithiasis).[1] In that case series, the five subjects each had pseudo-spontaneous nystagmus as our subjects did, but the duration of their positional nystagmus was not reported thereby making it difficult to confirm the otoconia were truly jammed.

Based on the presentation of these two cases, there remain few differential diagnoses that would cause the sudden reversal of nystagmus during positional testing, and thus this finding is critical to be confident in diagnosing canalith jam. Vestibular migraine can cause a persistent nystagmus that is position dependent, though the velocities of the nystagmus tend to be less than we report here.[16] One final thought. It has recently been visually verified that there is more than just free-floating otoconia in patients with BPPV. At the time of surgery for intractable BPPV, two patients were found to have fragments of otolithic membrane and otoconia encased in their

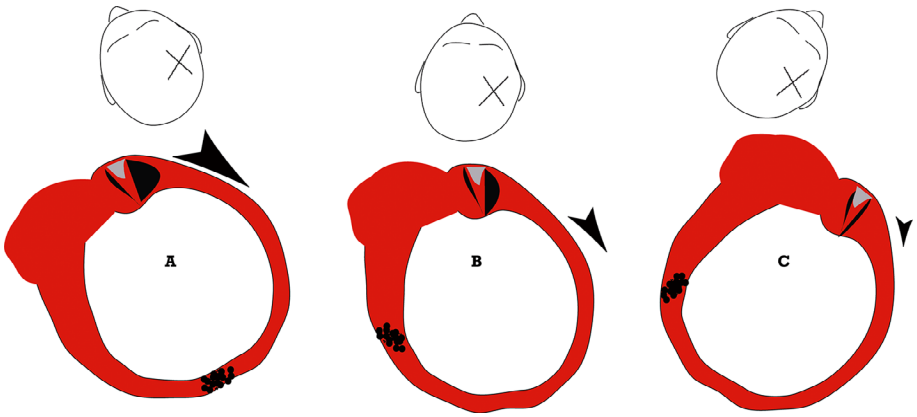

FIGURE 2 Example of otolith debris plugging/blocking the lumen within the right horizontal semicircular canal and deflecting the cupula in inhibition that changes magnitude depending on head position. The largest cupular deflection, A, occurs during head position rolled toward the unaffected side, which progressively reduces though does not stop as the subject moves from supine, B, to lying on their affected side, C. Arrows denotes magnitude of inhibitory stimulation

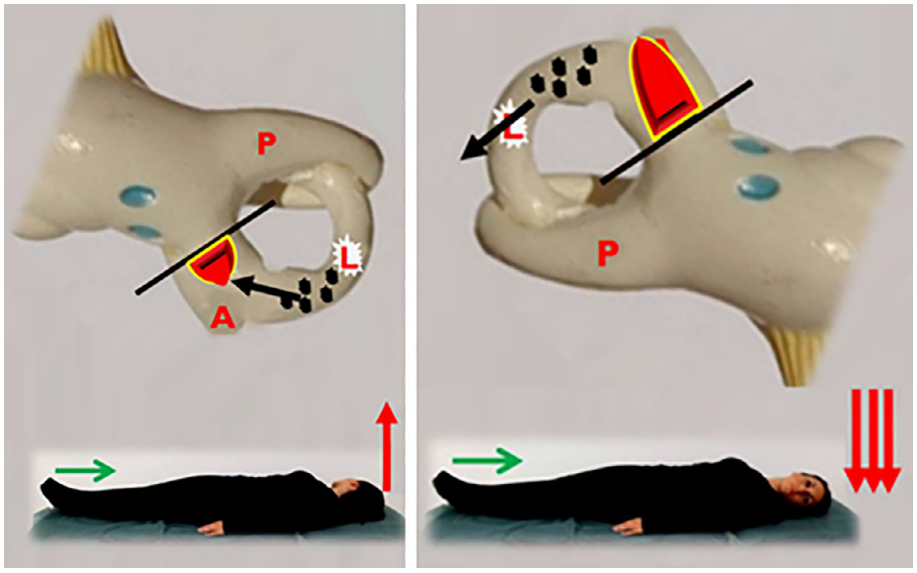

FIGURE 3 Example of otolith plug and freely floating otoconia within the lumen of the right hSCC. The cupula is initially deflected in inhibition. Next, in left roll position the free-floating otoconia move in an inhibitory direction, which increases the utriculofugal deflection causing a resultant increased left beat nystagmus. A, anterior semicircular canal; P, posterior semicircular canal; L, lateral semicircular canal and site of the jam

gelatinous matrix, putatively sluffed from the otolith organ. It is not hard to imagine that these relatively large chunks of membrane and otoconia could be held up or trapped in various locations inside the membranous portion of the semicircular canals, which themselves almost certainly have some irregularities in their wall, and fluctuate in their circumference.[17] Semicircular canal membrane plugs (jams), even perhaps more than one in a single canal, may be more common than appreciated and might be responsible for other unusual patterns of nystagmus such as persistent geotropic nystagmus with vertigo that currently is attributed to the "light cupula" phenomenon.

4 | CONCLUSION

Although rare, there is clinical evidence that otoconia can plug the lumen of the semicircular canals. Clinicians treating patients with vertigo should be aware of this possibility. Being mindful of four unique clinical presentations can help distinguish this confusing cause for positional vertigo:

1. Direction fixed spontaneous nystagmus (seated or supine) with fixation removed.
2. Direction fixed spontaneous nystagmus that does not change direction with positional testing
3. Velocity of the positional nystagmus and intensity of vertigo depends on head position
4. Conversion of unidirectional positional nystagmus to geotropic nystagmus

DECLARATION OF INTEREST

None of the authors has any financial interest to disclose.

CONFLICT OF INTEREST

None of the authors has any conflict of interest with the paper or its result.

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SUPPORTING INFORMATION

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