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An online presentation of a case and moderated discussion between the experts held on the last Friday of every month

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## 1. Issue in Focus:

 i) Classical benign paroxysmal positional vertigo BPPV of Posterior Semicircular canal (PSCC) is charachterised by a geotropic nystagmus on provocative manoeuvres, the presence of a crescendo-decrescendo nystagmus, appearing after a lag of less than ten seconds, lasting about 10 to 30 seconds with the direction being upwards and torsional towards the affected ear. The nystagmus is fatiguable on repeated manoeuvres.<sup>1</sup> It is due to free floating otoconial debris near the ampullary end of the canal, long arm canalolithiasis of the posterior canal

The direction of the nystagmus is explained by the endolymphatic flow induced in the affected semi-circular canal as per Ewald's laws.



[Fig 1: Parts of posterior semi-circular canal 1. Long arm 2. Non ampullary arm 3. Cupula 4. Short arm (utricular arm) 5. Periampullary region in long arm.]



[Fig 2: Typical form of PSCC BPPV nystagmus due to long arm canalolithiasis.]

#### 1.1 Variants of PSCC BPPV

The variant clinical presentation of benign paroxysmal positional vertigo (BPPV) is determined by the position of otoconia in the semi-circular canals due to anatomic variations or structural changes such as stenosis and fibrosis etc.

ii) **Apogeotropic type PSCC BPPV** is the most common variant characterised by downbeating and torsional nystagmus beating towards the upward ear (fig 3). Compared to typical PSCC BPPV, nystagmus may be elicited in any of the head hanging positions or even in lateral position. The latency of nystagmus is lower and the nystagmus is similarly-paroxysmal though of a lesser amplitude and longer duration (usually more than two minutes). This type of nystagmus is also poorly fatigueable. Vanucci et al. had explained this type of nystagmus as caused by otoconia lodged in the non-ampullary arm with partial canal jam.<sup>2</sup>

Fig 3 shows otoconia in non-ampullary arm of posterior semi-circular canal because of which invoked endolymph flow(ampullopetal) which is reverse as in typical form (left picture).

Treatment of this type of BPPV is with the demi-Semont Manoeuvre, Epleys' reverted procedure or 45 degrees forced prolonged positioning.



[Fig 3 Apogeotropic type PSCC BPPV characterised by down-beating and torsional nystagmus beating towards the upward ear]

It should be differentiated from anterior canal BPPV which has a similar type of nystagmus. by the absence of reversal of direction on sitting up.

- iii)Short arm canalolithiasis is a condition in which otoconia are in the utricular arm of the posterior semi-circular canal. After the paroxysmal phase of the positional nystagmus has ended, the short arm canalolithiasis provokes a slight and persistent ipsidirectional torsional nystagmus. In the typical form, a transient reversed torsional nystagmus occurs.<sup>3</sup>
- iv) Sitting up vertigo is characterised by positional vertigo and a nystagmus typical of PSCC BPPV
   - but is evoked when the patient sits up from the Dix-Hallpike position on the affected side (Sitting
   Up Vertigo). A narrowing of the periampullary arm of the canal or large-sized otoconial debris
   move the fluid distally in the Dix-Hallpike positioning. When the patient suddenly sits up, it
   invokes a "counter-positioning" nystagmus analogous nystagmus usually evoked in the ipsilateral
   Dix-Hallpike position in typical PSCC BPPV.<sup>4</sup>
- v) **Posterior canal cupulolithiasis** is caused by otoconia attached to the cupula. The quick phase direction of nystagmus is similarly geotropic (typical), upbeat and rotatory towards the down-most ea. This nystagmus has short or no latency, is less paroxysmal, longer-lasting and less fatigable. Liberatory manoeuvres may not be effective in heavy cupula from light endolymph, instead, physical therapy works better in cupulolithiasis, and mastoid vibration can help to detach otoconia from the cupula.<sup>5</sup>
- vi)**Biphasic variant** also known as Yetiser's variant is characterised by a biphasic paroxysmal nystagmus, whose first phase presents a torsional component opposite to that expected to the side of stimulation. It has been explained by otoliths which are located in the long arm of the posterior canal close to the common crus. It may result in ampullopetal flow initially. When the head is brought to left hanging position, debris will fall under the influence of gravity creating an endolymphatic flow toward the ampulla. Fig 4 <sup>6</sup>



[Fig 4 Biphasic variant]

vii) **Subjective posterior canal BPPV** : also known as "Type 2 BPPV", caused by the presence of free-floating otoconia in the utricle near the posterior canal cupula. Otoconial mass inertia produces neither significant cupular displacement nor nystagmus in the Dix-Hallpike positioning. When patients returned to the sitting position, otoconia will be floated in the short arm causing dizziness, sometimes with complaint of retropulsion.<sup>7</sup>

Epley stated that - "the more different a nystagmus is from the most common nystagmus observed in that disease, the more likely other diseases could be its cause"<sup>8</sup> Hence, the less common variants of posterior canal BPPV require in-depth evaluation to rule out cerebellar disease.

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## **Contributed by**



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# VERTICIO Grand Rounds

## 2.1 Apogeotropic Variant of Posterior Semi Circular Canal BPPV **Case Presentation**

i) A 38-year-old female patient presented on January 23, 2020, complaining of vertigo symptoms. Initial diagnosis identified left anterior canal benign paroxysmal positional vertigo (BPPV) based on down-beating torsional nystagmus. The treatment involved a daily regimen of 5 reverse Epley manoeuvres for 7 days. Upon her return on January 30th, the patient experienced a recurrence of vertigo. The Dix Hallpike test revealed classical up-beating and right torsional nystagmus, leading to a diagnosis of right posterior canal BPPV. The patient underwent a right Epley manoeuvre, resulting in symptomatic improvement. Therefore, it is postulated that the initial diagnosis of BPPV might have been complicated by apogeotropic BPPV of the right side.



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ii)Two additional cases were also presented suggestive of apogeotropic posterior canal benign paroxysmal positional vertigo (BPPV). The therapeutic approach involved application of the Demi Semont manoeuvre, quick liberatory manoeuvre, and forced prolonged positioning at a 45-degree angle.

#### 2.2 Discussion

This case of posterior canal BPPV lacked the usual presentation of up-beating ipsilateral nystagmus making it atypical. The mimicking of anterior canal BPPV could have been due to the presence of Canalolithiasis involving the non-ampullary arm of the posterior semicircular canal (PSCC), accompanied by partial canal jam due to otoconia. This manifests as a specific nystagmus pattern. During the Dix-Hallpike manoeuvre, this is characterized by the presence of down-beating nystagmus with a torsional component directed towards the contralateral side, denoting an apogeotropic response. ENT Consultant, Manipal Hospital, This diagnosis can be further explained by Califano's classification as described in Table 1:



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Califano's Classification of apo-PSC-BPPV	
Grade of Classification	Effect of Physical Therapy
Definite	Implies intracanalar transformation to typical
	PSC-BPPV within 2 days of physical therapy
Probable	When it resolves directly without
	transforming to typical PSC-BPPV after
	physical therapy
Possible	When symptoms persist despite five cycles
	physical therapy/ manoeuvres and a normal
	MRI or when the patient is lost to follow up
	despite ongoing symptoms



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## VERTIC<sup>G</sup>Grand Rounds

Patients exhibiting apogeotropic posterior semicircular canal benign paroxysmal positional vertigo (apo-PSC-BPPV) typically present with episodic dizziness triggered by positional changes, accompanied by persistent non-positional disequilibrium.

Procedures such as the Dix-Hallpike Test (performed bilaterally) in conjunction with the Enhanced Straight Head Hanging Test may induce down-beating torsional positional nystagmus directed towards the non-involved side. The repositioning manuvers for treating this kind of variant of posterior canal BPPV are enlisted below:

- 1. Demi semont manuvers
- 2. Quick liberatory rotation manuvers
- 3. 45 degree-forced prolonged positioning

## 2.3 Take Home Pearls

- i) Differentiating the canal involved: Cupulolithiasis of the posterior and horizontal semicircular canals involves observing persisting up-beating ipsilateral tortional nystagmus or down-beating contra-tortional nystagmus, dependent on the angulation of the cupula. Inversion testing is crucial for identifying cupulolithiasis.
- ii) Prevalence: Cupulolithiasis of the horizontal semicircular canal is more prevalent than that of the posterior canal.
- iii) Incidence of Recurrence: occurs in approximately 50% of patients even after re-positional manoeuvres.
- iv) VNG Features: Positional nystagmus is also observed in vestibular migraine patients, but they exhibit normal oculomotor patterns on videonystagmography (VNG) with non-fatiguing nystagmus possible in any head positioning manoeuvres.
- v) Treatment: Vibration on the mastoid is ineffective in dislodging cupulolithiasis; head positional maneuvers are the primary intervention.
- vi) Duration Between Reposition Manoeuvres: The duration between reposition manoeuvres depends on the patient's tolerating capacity, with a recommended minimum of two manoeuvres 24 hours apart.
- vii) Intervention: Zuma's manoeuvres, performed with patients in the sitting position, involves lying down on the affected side for 3 minutes (step I) and rotating the head 90° toward the ceiling for another 3 minutes (step II), providing relief for horizontal canal cupulolithiasis.
- viii) Challenges in intervention: Cupulolithiasis of the posterior canal does not improve with Semont manoeuvres, necessitating further research. Performing a Half Hallpike manoeuvres should be followed by an inversion test; a negative inversion test suggests posterior canal cupulolithiasis.
- ix) Diagnostic Value of vHIT: Video Head Impulse Test (vHIT) is a valuable diagnostic tool for patients with down-beating nystagmus.
- x) Subsequent Studies: There is a pressing need for further research on the diagnosis and treatment of cupulolithiasis.



## 2.4 Online VGR Video

## Scan the below QR Code



## to watch the complete Vertigo Grand Round Session



Postura - DR comes with dozens of calibrated exercise protocols as well as interactive games to rehabilitate every aspect of balance. Patient's progress can be measured from one visit to another.



## 3. Journal Scan

i) Agus G, Puxeddu R, Demontis GP et al

Atypical "reversed" paroxysmal positioning nystagmus in benign paroxysmal positional vertigo. Acta Otolaryngol Suppl. 1995;520 Pt 1:143-7. doi: 10.3109/00016489509125213. Abstract

atypical paroxysmal positioning nystagmus, An "reversed" in its directional components, and took on the typical paroxysmal positioning nystagmus (PPNy) pattern after repeated diagnostic manoeuvres. This pattern was observed in 7 patients.. They proposed that canalolithiasis, the presence of free-floating debris in the canal, explains this atypical PPNy .The debris can induce a gravity-driven movement of the cupula, leading to the atypical presentation. Additionally, repeated diagnostic maneuvers may influence the debris position and consequently alter the PPNy pattern, highlighting the potential benefit of repeating maneuvers in diagnosing BPPV even when PPNy appears atypical.

#### Editorial Comment:

This was the first description of atypical nystagmus in BPPV. The authors have suggested an elegant explanation for these findings and the method of treatment based on this vizthe presence of "free endolymph aggregates" (canalolithiasis) within the posterior semicircular canal (p.s.c).

#### ii) Ichimura A, Otsuka K.

Persistent Down-Beating Torsional Positional Nystagmus: Posterior Semicircular Canal Light Cupula? Case Rep Otolaryngol. 2016;2016:1249325. doi: 10.1155/2016/1249325.

#### Abstract

A case report of 16-year-old boy with rotatory positional vertigo and nausea, particularly when lying down. there vertical/torsional *Editorial Comment:* fatiguing was а non (downward/leftward) nystagmus in the supine position. Neurological examinations were normal. Authors speculated that persistent torsional down-beating nystagmus was caused by the light cupula of the posterior semicircular canal.

the patient's persistent torsional down beating nystagmus originated from a light cupula affecting the posterior semicircular canal. Due to a decreased density of the cupula within the inner ear crucial for equilibrioception, relative to the surrounding endolymph. Consequently, the cupula experiences gravitational deflection independent of head movements, resulting in persistent nystagmus.

The case presented explains atypical nystagmus using a novel light cupula hypothesis. Authors explained the importance of ruling out other possibilities of down beating positioning nystagmus like anterior canal **BPPV** and iii) Vannucchi P, Pecci R, Giannoni B.

Posterior semicircular canal benign paroxysmal positional vertigo presenting with torsional downbeating nystagmus: an apogeotropic variant.

Int J Otolaryngol. 2012;2012:413603. doi: 10.1155/2012/413603. Abstract

Among 45 patients with vestibular signs suggesting anterior semicircular canal paroxysmal positional vertigo (PPV), 6 subjects had clinical findings of a singular behavior during follow-up. At the first check-up, all patients were submitted to different types of physical manoeuvres for ASC canalolithiasis. At a next follow up, after having performed some physical therapies, all patients had a typical PSC PPNy of the opposite side, with respect to that of the ASC initially diagnosed. Basing on these observations the authors conclude that PSC PPV, could manifests in a apogeotropic variant.

the author hypothesised that free-floating particles could sometimes localize into the distal portion of the non-ampullary arm of the posterior semicircular canal (PSC) so that assuming the Dix-Hallpike's positions, the clot could move towards the ampulla eliciting a inhibitory torsional-down beating paroxysmal positional nystagmus (PPNy), instead of typical excitatory torsional-up beating PPNy.

#### Editorial Comment:

The study aims to verify the canalolithiasis hypothesis. This studu shows that the apogeotropic posterior canal BPPV mimics anterior canal BPPV of opposite side. It also additional proposes an mechanism of obstruction of endolymphatic flow due to a jam of otoconia. The authors using the vHITpropose maneveur to release this "jam" and aid recovery.

They propose that vHIT may play a key role in the diagnosis of the affected canal in BPPV involving vertical SC presenting with atypical nystagmus.

#### iv) Yetiser S. A

New Variant of Posterior Canal Benign Paroxysmal Positional Vertigo:

A Nonampullary or Common Crus Canalolithiasis.

Case Rep Otolaryngol. 2015;2015:816081. doi: 10.1155/2015/816081.

#### Abstract

This is a case report involving biphasic variety of positioning nystagmus i.e rotational nystagmus in opposite *A further* direction to the ear tested and reversal of initial positioning variant of rotational nystagmus after some time. Reversal of nystagmus *presented* is a rare condition and is basically due to endolymphatic flow *nystagmus* and clot movement in the opposite direction away from the rotatory ng cupula which is characteristic feature of canalolithiasis. *the scena* Authors had indicated that the reversal of positioning of the hypothesis nystagmus in this patient with PC-BPPV is related to unusual endolymph location of the otoliths in the long arm or the non-ampullary such cases noteworthy.

Atypical counterclockwise torsional up-beating nystagmus on the left head-hanging position is followed by true clockwise nystagmus by inversion of the direction of clot movement due to spontaneous reflux of the endolymph.

#### Editorial Comment:

A further example of atypical variant of BPPV which was presented with biphasic nystagmus with inversion of rotatory nystagmus. Explained the scenario well with the hypothesis of reflux of endolymph. The occurrence of such cases is infrequent but noteworthy.

#### 4. History of BPPV



Magnus Gustaf Retzius a Swedish anatomist dissected and documented the inner ear's labyrinth and reported the findings in 1846 paper, "On the Structure of the Cochlea and Labyrinth in Humans and Higher Mammals". He detailed the structure of the semicircular canals, utricle, and saccule, illustrating his findings with his own drawings. His work significantly contributed to the understanding of inner ear's anatomy, particularly its role in maintaining balance, and paved the way for future research like the study of the vestibular system.

Flourens observed pigeons circling after their horizontal JP semicircular canals were destroyed, pioneering vestibular research in 1824. He linked these canals to balance and posture, proposing their role in detecting head rotation (horizontal, vertical planes) and contributing to balance alongside the visual system. His findings, published in 1824 and laid the groundwork for understanding the vestibular system's role in balance.





Robert Barany is credited with the initial description of Benign Paroxysmal Positional Vertigo (BPPV) in 1921. His paper, "Diagnosis of Disease Symptoms in the Area of the Otolith Apparatus" outlined a method of diagnosing inner ear disorders by positioning the patient's head and observing eye movements. His work was crucial in recognizing and understanding BPPV, a condition causing brief vertigo episodes. While Barany didn't identify the specific cause of BPPV (later found to be displaced crystals), his description facilitated ongoing research and treatment development.

Barany's groundbreaking work on the inner ear earned him the 1914 Nobel Prize in Physiology/Medicine

Dix and Hallpike built upon the initial description of Benign Paroxysmal Positional Vertigo (BPPV) by providing a more comprehensive account in 1952. Using a positional test, they showed that the under-most ear caused the characteristic rotational nystagmus. Their findings were documented in two publications: "The investigation of vestibular function" (Br Med Bull 1956) and "Pathology, symptomatology and diagnosis of certain disorders of the vestibular system" (Proc R Soc Med 1952).



Margeret Ruth Dix and Charles Skinner Hallpike



JM Epley

JM Epley proposed the "Canalith Repositioning Procedure" (CRP) as a successful treatment for BPPV, achieving high resolution of symptoms in the initial study in 1992. He argued that the procedure dislodges free-moving particles in the inner ear responsible for BPPV symptoms. This offered an alternative explanation for BPPV and suggests CRP as a promising treatment option in his paper "The canalith repositioning procedure: for treatment of benign paroxysmal positional vertigo. Otolaryngol Head Neck Surg. 1992.

## 5. Gallery - Art work of the month



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