Atypical BPPV An Often Confusing Entity: Review of Literature

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ABSTRACT

Benign Paroxysmal Positional Vertigo (BPPV) is not an uncommon disorder involving the vestibular system, which is associated with recurrent attacks of vertigo and nystagmus and aggravated by positional change of the head. The "Barany Society diagnostic criteria for BPPV" includes vertigo or dizziness associated with a change in position along with positional nystagmus, and if these features are absent, we classify them as atypical BPPV. Lately, various forms of atypical BPPV have emerged, including cases with no nystagmus in typical positions, intense short-lasting nystagmus in different head positions. The phenomenon of "canal switch," where otolith from one semicircular canal shifts into another, has been proposed to explain the occurrence of paradoxical nystagmus. This review aims to provide a comprehensive overview of the possible origins of eye movements in all three semicircular canals and explore the different forms of atypical BPPV, contributing to a deeper understanding of this enigmatic condition.

INTRODUCTION

Benign paroxysmal positional vertigo (BPPV) is typically characterized by vertigo and nystagmus, which is triggered by changes in head position. It often occurs when otoconia (calcium carbonate crystals) become dislodged from the otolith macula and go into one of the semicircular canals. These otoconia move with changes in head position, causing abnormal fluid movement (endolymph flow) in the canal. This results in the deflection of the cupula, which alters the activity within the affected canal, producing vertigo and visible nystagmus. This process is known as canalolithiasis.¹ In rare cases, BPPV can occur when otoconia are displaced and adhere to the cupula of a semicircular canal rather than floating freely. In this situation, the otoconia respond to gravity, leading to vertigo and nystagmus. This phenomenon is referred to as cupulolithiasis.²

Currently, as per the Barany's society the diagnostic criteria of benign paroxysmal positional vertigo (BPPV) consist of recurrent attacks of positional vertigo or dizziness which is induced by change in head position, there will be associated positional nystagmus in the direction of the canal stimulated with various maneuvers, each attack lasting for few minutes,³ anything not fitting in this criteria were considered atypical BPPV.

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In the past years, many atypical BPPVs have been found, wherein either no nystagmus was found in otherwise normalprovoking head positions or instances of paradoxical, intense transient nystagmus were found in different head positions. Atypical BPPV was an unexplained phenomenon until recently, when theories of "canal switch" have been put forward, which says that paradoxical nystagmus happens when debris from one canal shifts into another during head positioning.⁴ Our goal in this review article is to provide a complete framework for the possible origins of eye movements across all three semicircular canals by summarizing these possibilities and various forms of atypical BPPV.

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METHODS FOR EXAMINATION

During the examination of patients with BPPV, based on the symptoms and the most common predictable canal involved, the examiner should perform positional maneuvers to confirm the canal involved.

For posterior canal, Dix-Hallpike maneuver is the diagnostic maneuver wherein, the patient is made to sit, and head is turned to 45 degrees to the side which is to be examined, the patient is made supine with head hanging below the edge of the bed, if posterior canal is stimulated an up-beating torsional nystagmus is elicited on the side which is examined. For anterior canal, head hanging maneuver is done wherein the patient has to lie supine with the head hanging below the edge of the bed, if anterior canal is stimulated a down-beating torsional nystagmus will be elicited, anterior canals can also be checked using Dix-Hallpike maneuver, we only need to see the beat pattern of the nystagmus whether it is down-beating or not. For the lateral canals, a supine roll test is done wherein the patient will be made to lie supine, and head is flexed, and the head is turned 90 degrees to the side tested; a horizontal nystagmus is elicited with the fast beating component towards the side of the canal stimulated.³

TYPICAL BPPV NYSTAGMUS PATTERNS

Posterior semicircular canal canalolithiasis

Positional nystagmus, which is triggered by the Dix-Hallpike maneuver or Semont diagnostic maneuver-side lying maneuver, after a delay of few seconds typically consists of a combination of torsional upbeating nystagmus. This nystagmus usually lasts for less than a minute.⁵

Posterior semicircular canal cupulolithiasis

Here, the nystagmus elicited from Dix-Hallpike maneuver or semi Dix-Hallpike maneuver is same with that of posterior canal canalolithiasis, except that the duration of the nystagmus lasts more than a minute in cupulolithiasis⁶ there is yet another hypothesis to this which we shall see below.

The horizontal semicircular canal canalolithiasis

Positional nystagmus, which is triggered by the supine roll test after a transient delay or with the absence of latency, typically beats horizontally⁷ toward the side on which the head is turned. This nystagmus generally lasts for approximately less than one minute.⁸

The horizontal semicircular canal cupulolithiasis

Positional nystagmus, which is triggered by the supine roll test after a transient delay or with the absence of latency, typically beats horizontally⁷ in the direction of the upper ear when the head is turned to the left or right side. This nystagmus generally lasts less than one minute.

The anterior semicircular canal canalolithiasis and cupulolithiasis

This is seen very rarely, and various hypothesis has been brought forward, it is hypothesised that during sleep, debris in the common crus may shift into the ampullary end of the long arm, another possibility is that during the Dix-Hallpike maneuver, dislocated debris could immediately fall onto the cupula, particularly in cases of anterior cupulolithiasis, causing an apogeotropic down-beating nystagmus with a minimal torsional component.⁹ In cases of anterior semicircular canal (SCC) involvement, a minimal torsional fast-phase nystagmus may occur, with the fast phase beating toward the affected ear when the patient is in the Dix-Hallpike position. Therefore, the direction of the torsional component, rather than the side of the Dix-Hallpike test, indicates which side is affected.

ATYPICAL NYSTAGMUS ARISING IN THE ANTE-RIOR AND POSTERIOR SEMICIRCULAR (VERTI-CAL CANALS)

BPPV without nystagmus and posterior semicircular canal positional downbeating nystagmus

The usual typical BPPV is associated with nystagmus and acute vertigo within few seconds after positioning. It has been found that in some cases, patients with symptoms of vertigo will get aggravated vertigo during positional maneuvers like Dix-Hallpike and supine head roll test, but they will not elicit nystagmus, it has earlier been named as subjective BPPV and it is not yet characterised under typical BPPV. Various authors have come forward with their hypotheses to explain this phenomenon. Oas et al. considered it as the "short arm canalolithiasis", wherein the debris freely moves in the short arm, causing vertigo while getting up during Dix-Hallpike maneuver only on the side where the canal is affected, but no nystagmus is elicited.10 Cambi and Vannucchi brought forward a variant of atypical BPPV wherein they suggested that positional down-beating nystagmus can also be seen in cases of posterior canal; earlier it was known that down-beating nystagmus was only associated with anterior canal.¹¹⁻¹² The hypothesis put forward by them was that the debris lies in the highest part of the long arm of the canal at the start of the Dix-Hallpike maneuver. It is theoretically possible for otoconia to become dislodged without migrating into the common crus of the superior and posterior semicircular canals or horizontal canal. Rather, the otoconia can shift in the direction of the posterior utricle, specifically the posterior canal ampulla.

Depending upon their action—whether they go freely or cling to the cupula—and the exact location of the posterior semicircular ampulla (this can be varied), the effect might be that there is neither nystagmus nor there is a reduction in the nystagmus when the patient is moved from sitting to Dix-Hallpike position. This situation could explain clinical conditions like "subjective BPPV" or peripheral positional down-beating nystagmus cases.⁴ If this theory is embraced, as difficult to verify or negate as it is, it may lead to the discovery of two more variations of the classical forms of BPPV.

Debris, when stuck to the cupula, deflects it. This will result in either no. nystagmus in Dix-Hallpike position or an apogeotropic downbeat nystagmus might. Develop in this position, which can be provoked by a bilateral Dix-Hallpike maneuver (and is possibly increased when the diseased ear is in the downward position). If there is a torsional element within the nystagmus, it will beat towards the opposite ear.

Posterior semicircular canal cupulolithiasis

Debris, when attached to the cupula, causes it to deflect. This will lead to either no nystagmus in the Dix-Hallpike position or an apogeotropic downbeat nystagmus that may develop in this position, which can be triggered by a bilateral Dix-Hallpike maneuver (and may be more pronounced when the affected ear is in the lowest position). If there is a torsional component in the nystagmus, it will beat toward the opposite ear.

Debris, when it binds to the cupula, deflects it. This will result in no. Nystagmus on the Dix-Hallpike position, or an apogeotropic downbeat nystagmus, can occur in this position and is provoked by a bilateral Dix-Hallpike maneuver (and perhaps more intense with the affected ear in the most inferior position). If the nystagmus has a torsional component, it will beat toward the other ear.

The Posterior semicircular canal short arm canalolithiasis

Since the debris is moving away from the short arm, no nystagmus is evoked when the patient is in the Dix-Hallpike position. However, on sitting up, the patient will suffer from vertigo or a sensation of body sway, usually in the diagonal vertical planes corresponding to the right anterior-posterior (RALP) or left anterior-right posterior (LARP) semicircular canals. In both posterior canal cupulolithiasis and posterior short arm canalolithiasis, an intense vegetative reaction, like nausea and perspiration, has been reported following repeated position maneuvering. The reaction is remarkably disproportionate because there was no nystagmus.⁴

ATYPICAL NYSTAGMUS ARISING IN THE HORI-ZONTAL SEMICIRCULAR CANALS

The Horizontal semicircular canal short arm canalolithiasis

Hypothetically, suppose debris were to fall into the short arm of the horizontal semicircular canal. In that case, it is expected to enter into the cupula, which will lead to nystagmus in the horizontal plane, which will be apogeotropic (that means, eye beating in the direction opposite to the side of head turn). While doing a supine roll in the opposite direction, the debris may move out, so no nystagmus would be expected during the contralateral supine position. While this scenario is commonly seen after Epley or Semont maneuvers, no prior studies have suggested this mechanism involving the short arm of the horizontal semicircular canal. Other researchers have explained this nystagmus pattern and have attributed it to an unusual initial positioning of the debris in the long arm of the horizontal semicircular canal.¹³

ATYPICAL NYSTAGMUS ASSOCIATED WITH SWITCHING OF CANAL

The Posterior semicircular-horizontal semicircular canal switch

Following Epley or Semont maneuvers, canal switch is not an uncommon phenomenon. Here, debris from the horizontal semicircular canal can move into the posterior semicircular canal and vice versa, which will result in characteristic nystagmus, which is characterised by atypical nystagmus initially originating from the affected canal and then shifting to the secondary canal. The most frequent pattern seen is the transition from posterior semicircular canal canalolithiasis to horizontal semicircular canal canalolithiasis. In these cases, the debris appears to be large enough that it doesn't dissolve during its journey through one canal, out of it, and into the next.¹⁴

The Horizontal semicircular ipsicanal switch

Hypothetically, in the horizontal canal, short arm canalolithiasis can switch over or transition to canalolithiasis of the long arm while doing a supine roll test if the debris moves from the short arm to enter into the vestibulum and finally enter the long arm, or vice versa fro long arm to short arm is also possible. This would cause the direction of nystagmus to reverse when the patient repeatedly rotates their head in the supine position to the lateral side with the affected ear facing downward.¹⁴

Previously, Nuti et al. proposed an alternative theory for this unusual nystagmus,¹⁵ suggesting that it involves debris initially located in an atypical position. Previous researchers explained this phenomenon as a conversion of canalolithiasis to cupulolithiasis.¹⁶ However, horizontal ipsicanal switch as a potential explanation has been first described by Büki et al.¹⁴

Atypical nystagmus associated with atypical positioning

For completeness sake, manifestations of two common atypical nystagmus have been outlined, providing clues for diagnosis. Firstly, the nystagmus that occurs during the lying-down position, from sitting position to supine during examination. This is because of the sinking of the debris, which was initially located in and around the ampulla or sometimes in the cupula of the horizontal canal leading to horizontal nystagmus, which can sometimes be confused with spontaneous nystagmus. Second, down-beating nystagmus that occurs during sitting up from lying position in patients with acute posterior canalolithiasis, which might confuse posterior canalolithiasis, usually has an up-beating type of nystagmus.¹⁴

OCCURRENCE OF VARIANTS

Studies have shown that the right labyrinth is more commonly involved in cases of canalolithiasis.¹⁷ However, it remains unclear whether this observation applies solely to canalolithiasis or also to cupulolithiasis. This preferential involvement still needs to be confirmed through new epidemiological studies that incorporate the updated criteria (such as no nystagmus seen during Dix-Hallpike position in hypothetical posterior cupulolithiasis, if the scientific community accepts these criteria). This issue highlights a broader point about epidemiological studies: if new hypothetical mechanisms are to be generalized, additional research should be conducted. Vestibular migraine is one such example wherein it can overlap with BPPV even in the absence of nystagmus. A study by Jeong et al.¹⁸ is yet another good example where 100 patients diagnosed with idiopathic BPPV were recruited, but it was found that 84 patients had typical symptoms of BPPV, but no nystagmus was seen; they were found either to have a posterior short arm canalolithiasis or cupulolithiasis.

In regards to unilateral end organ involvement, the posterior semicircular canal and ampulla are more prone for debris getting trapped in comparison to the anterior semicircular canal and ampulla. Typical BPPV, where debris must reach the openings of the long arms, may be less common than vestibulolithiasis, where debris simply sinks into the posterior short arm and moves there. The higher frequency of classical BPPV could be due to the utricular macula being nearly horizontal in an upright position, making detachment less likely. However, when the patient is lying down in the supine position (with the utricular surface vertical or upside down), the orifices are positioned below the utricular macula, facilitating debris dislodgement.

CONCLUSION

In conclusion, atypical BPPV is a common and often challenging condition to diagnose, especially when atypical presentations arise. The hypotheses presented here suggest that atypical BPPV may be more frequent than currently recognized, with certain cases displaying either no nystagmus or atypical nystagmus patterns. While these theories are not directly provable at this stage, they are in line with the evolving understanding of the various forms of BPPV that have been observed over time. If accepted in the future, these insights could significantly ease the diagnostic process, preventing cases without nystagmus or with unusual nystagmus patterns from puzzling clinicians. This paper aims to encourage further discussion and research into these atypical forms of BPPV, which, despite their complexity, can be easily treated once correctly identified, offering relief to patients who suffer from this often distressing condition.

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