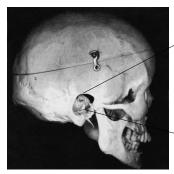
Downbeat Nystagmus Associated with Atypical PC-BPPV:

Differentiating Cupulolithiasis, Short Arm Canalithiasis, and Canal Jam





Blanks RH, Curthovs IS, et.al, 1975

Janet O. Helminski, PT, PhD, Professor, Department of Physical Therapy Rosalind Franklin University of Medicine and Science North Chicago, IL

Recommended Reading

- Bhattacharyya, N., Gubbels, S. P., Schwartz, S. R., Edlow, J. A., El-Kashlan, H., Fife, T., . . . Corrigan, M. D. (2017). Clinical Practice Guideline: Benign Paroxysmal Positional Vertigo (Update). Otolaryngol Head Neck Surg, 156(3 suppl), S1-S47. doi:10.1177/0194599816689667
- Buki, B., Mandala, M., & Nuti, D. (2014). Typical and atypical benign paroxysmal positional vertigo: literature review and new theoretical considerations. J Vestib Res, 24(5-6), 415-423. doi:10.3233/VES-140535
- Nuti D, Zee DS, Mandala M. Benign Paroxysmal Positional Vertigo: What We Do and Do Not Know. Semin Neurol. 2020;40(1):49-58.
- von Brevern M, Bertholon P, Brandt T, et al. Benign paroxysmal positional vertigo: Diagnostic criteria. J Vestib Res. 2015;25(3-4):105-117.
- Epley JM. Human experience with canalith repositioning maneuvers. Ann N Y Acad Sci. 2001 Oct;942:179-91.

Objectives

Upon completion of this session, the participant will be able to:

- 1. Select and implement appropriate **positional tests** to differentially diagnosis PC-BPPV cupulolithiasis, short arm canalithiasis, or obstruction.
- 2. Identify and interpret atypical patterns of benign paroxysmal positional nystagmus found on positional testing and during canalith repositioning maneuvers that suggest PC-BPPV cupulolithiasis, short arm canalithiasis, or obstruction.
- **3. Implement a canalith repositioning maneuver** for the treatment of atypical benign paroxysmal positional vertigo involving the posterior canal based on the anatomy and mechanics of the canal.

Definitions

- (1) Ipsilateral towards involved ear.
- (2) Contralateral away from involved ear.

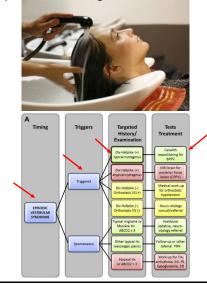
3

Benign Paroxsymal Positional Vertigo

Mechanical disorder of the inner ear caused by abnormal stimulation of 1 or more of the 3 semicircular canals within the ear. The diagnosis of BPPV is determined based on history and findings on positional testing.

TiTrATE (Newman Tolker, Edlow, 2015)

- 1. **Timing** episodic vestibular syndrome
- **2.** Triggered changes in position of head relative to gravity.
- **3. Targeted**history/examination changes in position/DHT



BPPV: Diagnostic Criteria — Consensus Document Bárány Society Indicated translation formed 23 (2003 196-117 per 102(2014) 196-117 p

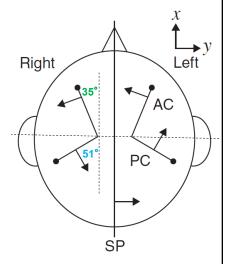
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Biomechanics of Canals

Orientation of Canal Planes:

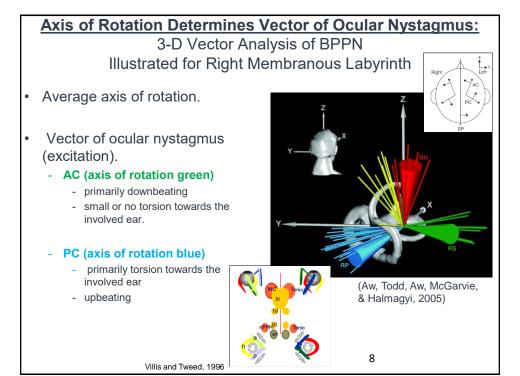
Comparison of Human Canal Planes Using MRI

- Orientation of vertical canal planes from sagittal plane with reference to the frontal plane bisecting the skull.
 - AC 35°
 - PC 51°
- Axes of rotation orthogonal to canal plane.



(Suzuki, Masukawa, Aoki, Arai, & Ueno, 2010)

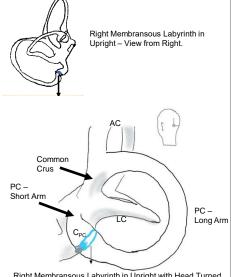
7



Position and Orientation of Initial Ampullary Segment

Illustrated for Right Membranous Labyrinth in the Upright Position

- Position and Orientation of initial ampullary segment (Bertholon, Bronstein, Davies, Rudge, & Thilo, 2002).
 - AC initial ampullary segment is 70° with respect to earth horizontal (almost vertical).
 - PC initial ampullary is 20° below earth horizontal.
- Natural individual variability in location of attachment of PC to the utricle. (Buki, 2014; Tomanovic & Buki, 2016).



Right Membransous Labyrinth in Upright with Head Turned 45° towards Right – View from Left

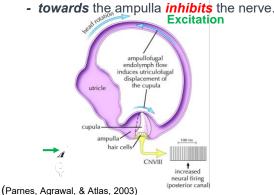
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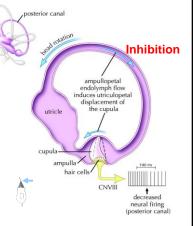
Orientation of Hair Cells of the Cristae Ampullaris:

Position of Kinocilium within Cristae Ampulla and
Direction of Flow of Endolymph Relative to Kinocilium Determine if
Provoked Response of Vestibular Nerve Excitatory or Inhibitory
Deflection of hair cells away from the kinocilium inhibits and
iceal canals towards the kinocilium excites CN VIII.

In the vertical canals towards the kinocilium excites CN VII

- · Kinocilium positioned near long arm of canal.
- Flow of endolymph:
 - away from the ampulla excites the nerve.

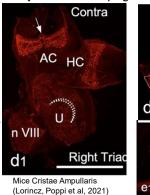


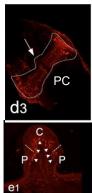


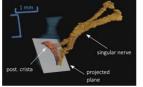
Position & Orientation of Cristae Ampullaris

Illustrated for Right Membranous Labyrinth in the Upright Position

- Position and orientation of cristae ampullaris within ampulla (Lorincz, Poppi et al, 2021). Central enlargement.
 - PC and AC cristae positioned on outer arch of ampulla wall with cupula extending from the crista across the ampulla to the inner arch.
 - LC cristae positioned on inner arch of ampulla wall with cupula extending to the outer arch.
- Topographical orientation of the long axis of the cupula disputed (Buki, 2014; Tomanovic & Buki, 2016).
- Natural individual variability in attachment of cupula to ampulla (Buki, 2014; Tomanovic & Buki, 2016).







Human Cristae Ampullaris and projected plane of PC cupula (Harmat, Tamás, et al 2022)

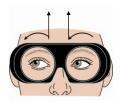
(Buki, Mandala, Nuti, 2014)

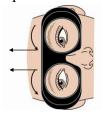
11

Mechanisms of Atypical pc-BPPV Illustrated right ear AC -**Location of Debris** Long Arm Canalithiasis - short arm Cupulolithiasis -Particulate matter adherent or adjacent to cupula. Common Crus Particulate matter located on LC - Long Arm either side of cupula: Utriculus Short arm long arm - Short Arm Canal Jam or Obstruction. Obstruction located: Peri-ampullary segment Non-ampullary segment PC - Long Arm

Positional Nystagmus – Reference Frame

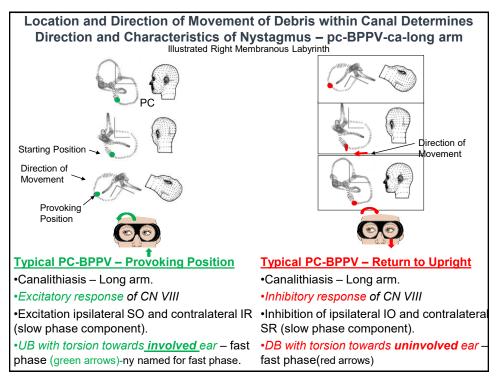
• **Head reference frame**. Name nystagmus relative to patient sitting upright. For example, upbeat towards the forehead and downbeat towards the chin independent of the position.

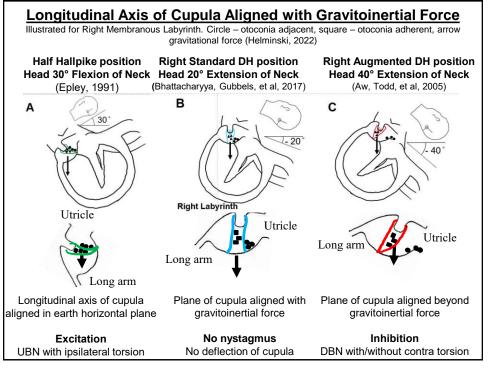




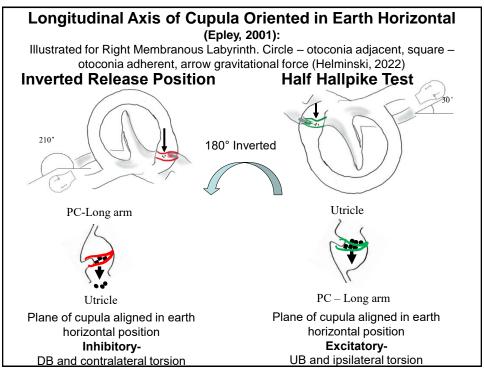
• Earth reference frame. Name nystagmus relative to earth. For example, nystagmus always directed towards the earth - geotropic and away from the earth - apogeotropic independent of the position.

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Factors Contributing to Changes in Cupular Response Dynamics Associated with pc-BPPV

Factors that influence the longitudinal axis of the cupula relative to the gravitoinertial forces that influence the patterns of nystagmus are:

- Position and orientation of the head in DH position:
 - Limited spine mobility or pain
 - Amount of neck extension in DH position less pronounced ~20° or more pronounced ~40°
- Variability in location of attachment of PC to the utricle (Buki, Mandala, et al, 2014)(Harmat, Tamas, et al, 2022)
 - Short arm attached more superiorly no ny is observed – plane of cupula aligned in earth gravitoinertial plane
 - Short arm attached more inferiorly DB ny is observed – plane of cupula aligned just beyond earth gravitoinertial plane.
- Variability in position of the cupula within the ampulla (Buki, Mandala, et al, 2014)(Harmat, Tamas, et al, 2022)
- Location of debris
 - Within ampulla
 - Adherent or adjacent to the cupula





(Gacek . 2008)

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Targeted History and Examination

<u>Targeted History –</u> Critical to Differential Diagnosis Process

Triggers

 Individual 4.3 times more likely to have BPPV if getting out of bed and rolling over in bed causes symptoms of vertigo/dizziness (Whitney, Marchetti, & Morris, 2005).



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Targeted History

- 39% of individuals presenting with atypical pc-BPPV will have been previously treated with a canalith repositioning maneuver for pc-BPPV (Scocco, Barreiro, et al, 2022).
- · Findings that suggest possible atypical pc-BPPV:

	DBN in DH Positions Suggesting Atypical pc- BPPV – Cu (Helminski, 2022) or jam (Vannucchi et al., 2015)	UBN Upon Return to Upright Suggesting pc- BPPV-Short Arm (Helminski, 2022)	
Dizziness/True Rotational Vertigo	Marked sense of dizziness/imbalance. Vertigo transitioning ½ way into recumbent position.	Marked sense of dizziness/imbalance. Truncal retropulsion during/after sitting up.	
Episodic/persistent	Persistent – debris unstable	Persistent – debris unstable	
Vertigo triggered	Symptoms last longer than typical PC-BPPV	Symptoms last longer than typical PC-BPPV	
Symptomatology	Marked Neuro Vegetative	Marked Neuro Vegetative	

Targeted Examination

Step 1: Determine Vertical Canal Involved. Align plane of canal being tested with the gravitational force and rotate canal within plane.

Part 1. Dix-Hallpike Test Part 2. Straight Head Hang

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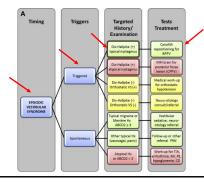
pc-BPPV canalithiasis of long arm

Case 1 – Targeted History

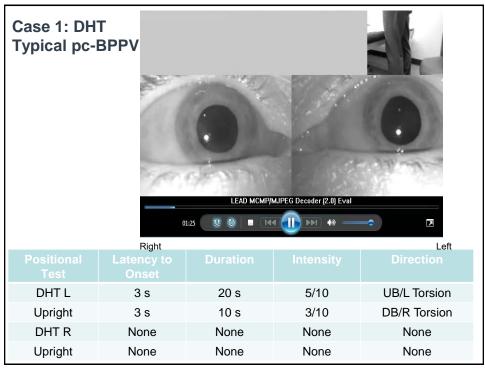
86-year-old female

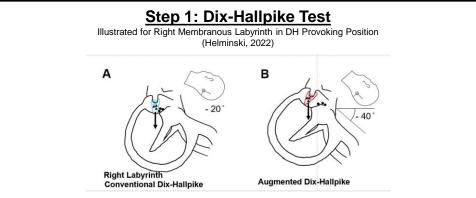
- Targeted History 1 week ago rolled over in bed towards the right side and experienced sudden onset of vertigo.
- Timing Episodic vertigo and nystagmus lasting < 2 minutes.
- Triggers changes in position of head relative to gravity.
 - No history of associated nausea or vomiting.
 - Lightheaded when still.
 - Feels unsteady on her feet.
- Targeted Examination DHT





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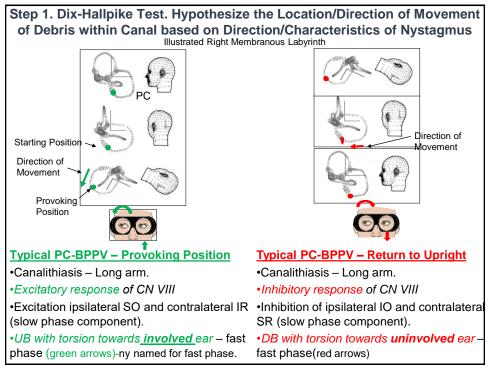




DHT – To evaluate the vertical canals the head needs to be positioned a minimum of 20° below earth horizontal. Each position maintained a minimum of 45 s.

- Conventional DHT head 20° below earth horizontal (Bhattacharyya et al., 2017).
- Augmented DHT head 40° below earth horizontal.(Aw, Todd, Aw, McGarvie, & Halmagyi, 2005). Optimizes canal orientation. Recommend use of Augmented DHT.

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Step 1: Determine Vertical Canal Involved Based on Direction and Characteristics of Nystagmus - DHT

Case 1: Left Posterior Canal BPPV – long arm canalithiasis based on latency of onset and duration.

Findings ipsilateral DH

- Excitation in provoking position upbeat/torsion ny towards involved side.
- Inhibition upon return to sitting downbeat/torsion ny towards uninvolved side.

Intervention – Canalith Repositioning Manuever

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pc-BPPV Ipsilateral Canal Switch

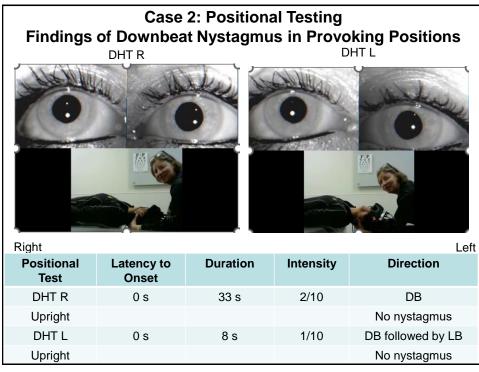
Case 1 – Targeted History

59-year-old female

- Targeted History Last night rolled over in bed and experienced sudden onset of severe vertigo and vomiting.
- **Timing** Episodic vertigo and nystagmus lasting < 2 minutes.
- Triggers changes in position of head relative to gravity.
 - Marked nausea.
 - Vomiting carries emesis basin
 - · Marked imbalance.
- Targeted Examination DHT

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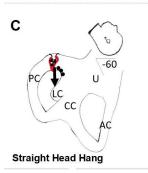


Step 1: Determine Vertical Canal Involved Based on Direction and Characteristics of Nystagmus - SHH

Dix-Hallpike Test – atypical findings:

- In 1 or both positions of DHT. Observe downbeat/with or without torsion.
- Return to sitting No ny.

Straight Head Hang (SHH) - perform if observe atypical downbeat nystagmus, or no nystagmus in provoking positions.



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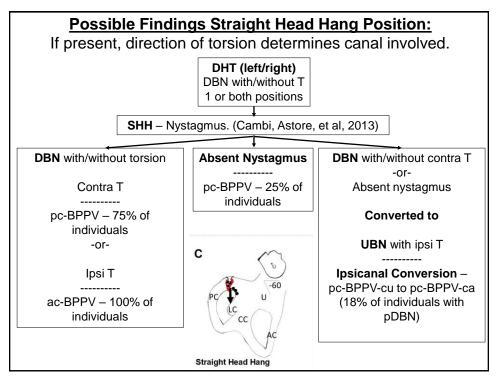
Case 2: Determine Vertical Canal Involved - DHT Findings of Downbeat Nystagmus in Provoking Positions



Right

SHH	Left
	LEIL

Positional Test	Latency to Onset	Duration	Intensity	Direction
DHT R	0 s	33 s	2/10	DB
Upright				No nystagmus
DHT L	0 s	8 s	1/10	DB followed by LB
Upright				No nystagmus
SHHP	6 s	19 s	8/10	UB/L torsion followed by DB
Upright	0 s		8/10	DB/R torsion



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Mechanism - Ipsicanal Switch

- Of individuals with peripheral DBN (n=43), ipsilateral switch occurred from (Cambi, Astore, Mandala, Trabalzini, & Nuti, 2013)
 - Persistent DB to transient UB nystagmus in 18%
 - Transient UB to persistent DB nystagmus in 24%
- Of individuals (n=100) with pc-BPPV canalithiasis of the long arm successfully treated with a particle repositioning maneuver, 19% experienced persistent DBN in the DHT suggesting pc-BPPV cupulolithiasis of the short arm (Harmat, Tamás et al, 2022).





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pc-BPPV cupulolithiasis

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Targeted Examination

Step 1: Determine Vertical Canal Involved – Align plane of canal being tested with the gravitational force and rotate canal within plane.

- Part 1. Dix-Hallpike Test
- Part 2. Straight Head Hang

Step 2: Determine Pathophysiology of Downbeat Nystagmus. Longitudinal axis of affected cupula oriented in earth horizontal position.

- Part 1. Half-Hallpike Test
- Part 2. Inverted Release

Case 3 – Targeted Examination

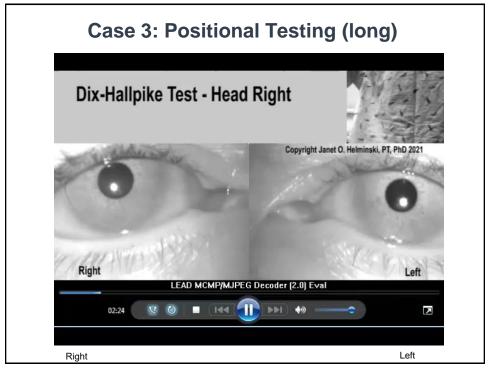
85 year old female

- Targeted History rolled over in bed and became dizzy.
 Vertigo with the following:
 - 1. Roll towards the left
 - Transitions between recumbent position to sitting. Extreme vertigo towards left at position ½ way down.
 - 3. Looking up.
 - 4. Bending forward.
 - 5. Rapid head movements.
- Timing brief periods of episodic vertigo for 3 weeks.
- Triggers changes in position of head relative to gravity. No history of associated nausea or vomiting.
- Targeted Examination

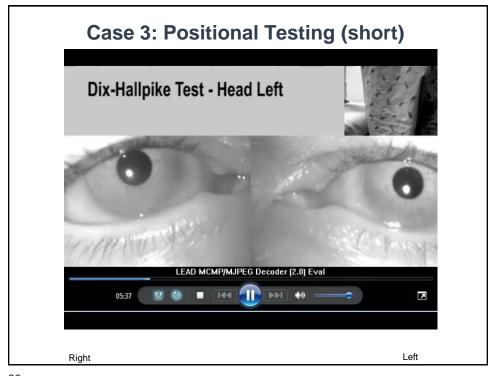




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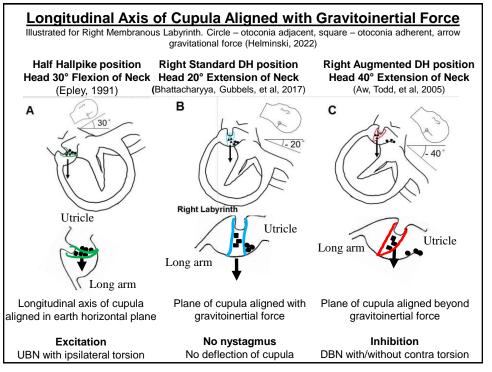
Case 3 – Findings on Targeted Examination

85 year old female

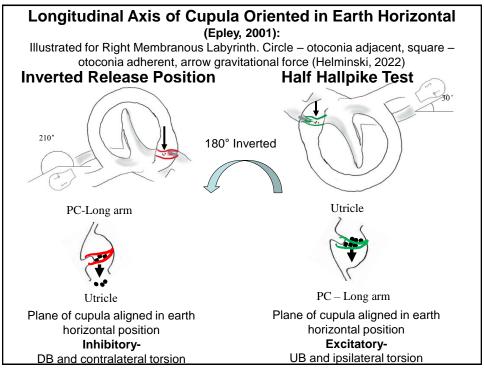
- Direction of nystagmus suggest BPPV involving L PC – cupulolithiasis
 - Half Hallpike Left UB/L Torsion
 - DHT Left DB/R Torsion
 - Inverted R Sidelying DB/R Torsion

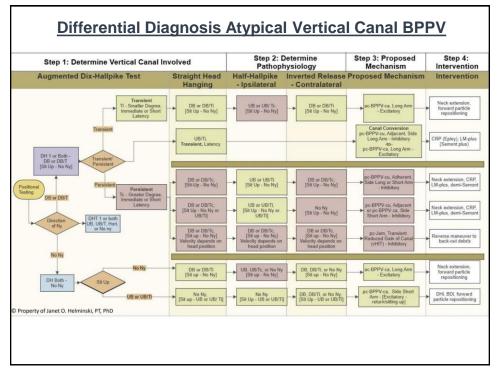
Positional Test	Direction of Ny
Half Hallpike L	UB, L Torsion
DHT L	DB, R Torsion
Upright	LB
Inverted R Sidelying	DB, R Torsion
Half Hallpike R	None
DHT R	DB, R Torsion
Upright	LB
Inverted L Sidelying	RB
SHHP	DB, R Torsion
Upright	LB

40



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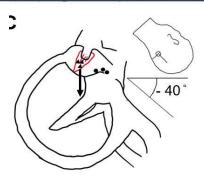
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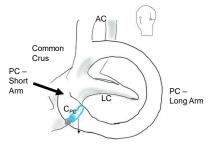
pc-BPPV canalithiasis short arm

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Canalithiasis of Short Arm- Apogeotropic PC-BPPV

- Excitation upbeat nystagmus with torsion towards involved side upon return to upright from provoking position.
- Type 2 BPPV
 - Symptoms suggestive of BPPV
 - · No nystagmus with VOG
 - Vertigo with truncal retropulsion during and immediately after sitting up from the involved side.





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Case 4 – Recurrence of Apogeotropic PC-BPPV - Sitting-up Vertigo

- Targeted History 10 days following initial treatment, BPPV recurred. In bed, rolled towards left and developed intense vertigo.
 - · Intense vertigo when sits up.
 - Extreme imbalance. Fell on risers.
 - Constant "floating" sensation and nausea.
- Targeted Examination Evaluated by PT 3 days after recurrence.
 - · No neurological findings.
 - · No vestibular suppressant medication
 - DHT. Used video oculography to avoid visual fixation.





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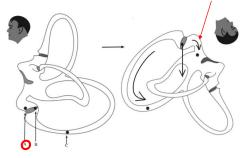
Case 4 – Recurrence Apogeotropic PC – BPPV - Targeted Examination.

- · Head Right, Head Left and SHHP No ny or vertigo.
- Return to sitting UB with Left Torsion associated with vertigo.



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Case 4 – Sitting-up Vertigo - Differential Diagnosis



Proposed mechanism- BPPV involving short arm. Findings on positional testing:

- **UBN** upon return to upright from right/left provoking position of DHT and SHH and no other neurologic signs **suggest vertical canal involvement**.
 - Apogeotropic PC BPPV
- **Left torsion** upon return to upright from right/left provoking position and SHH suggests left PC-BPPV. Torsion component suggests canal involved.
 - · Right torsion excitation of the right PC
 - Left torsion excitation of left PC.

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Canal Jam

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Ic-BPPV Jam

Obstruction to the lumen of the lc due to large fragments of particular matter or a constriction of the lumen and create positive or negative pressure resulting in persistent deflection of cupula on the affected side.

Four observations with VOG:

- Direction fixed spontaneous nystagmus with fixation removed.
- · Direction fixed spontaneous nystagmus during positional testing.
- The velocity of the positional nystagmus and intensity of vertigo depends on supine roll head position.
- Conversion of unidirectional positional nystagmus to geotropic nystagmus.

Case 5 - Positional Vertigo

Targeted History – 63 yo female with no history of BPPV

- Monday evening attended a dinner banquet.
 - Turned her head right to look at the speaker and then straight ahead.
 - Began feeling uncomfortable and stopped moving her head side to side.
 - Then, started experiencing a sensation of dizziness.
 - Left banquet early due to dizziness.
 - Went home. Went to bed. Slept on left side. If lied on right became dizzy.
- Tuesday morning woke up and was feeling fine.
 - Started moving around and became dizzy. Dizziness gradually increased especially with horizontal head movement.
 - Vomited.
 - Rested on left side. Right side experienced dizziness. Within 4 hours was fine.
- Wednesday morning work up and feeling "off".
 - Evaluated by physical therapist

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Case 5 - Positional Vertigo

Subjective

- Working at desk looking between 2 computer screens began experiencing dizziness.
- Horizontal head movements increased intensity of dizziness.
- Lie on right side felt dizziness. Lie on back felt worse. Lie on left side felt best.



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Case 5 - Ic-jam

Spontaneous Nystagmus, GEN, and Positional Testing with VOG.



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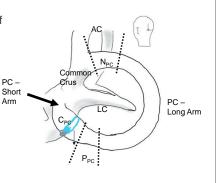
pc-BPPV Jam

Obstruction to the lumen of the pc due to large fragments of particular matter or a constriction of the pc lumen (Vannucchi, Pecci et al, 2012; Vannucchi, Pecci et al, 2015).

Complete obstruction - reduced gain of involved canal (vHIT). (Castellucci A, Malara P et al, 2020).

Hypothesized obstruction to:

- Non-ampullary or distal segment of PC referred to as "apogeotropic PC-BPPV" (Vannucchi, Pecci et al, 2012; Vannucchi, Pecci et al, 2015).
 - DBN with or without contralateral torsion in 1 or both DH position and SHHP
 - No nystagmus upon return to upright.
- Peri-ampullary segment of PC referred to as "sitting-up vertigo" (Scocco et al, 2019).
 - DBN with contralateral torsion or no nystagmus DH position and SHHP
 - UBN with ipsilateral torsion upon return to upright





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Interventions for Atypical pc-BPPV

- Ipsilateral Canal Switch apogeotropic to geotropic nystagmus.
- Intervention for both long and short arm side of pc.
- · Intervention for long arm side of pc.
- Intervention for short arm side of pc.
- Canal jam back out debris

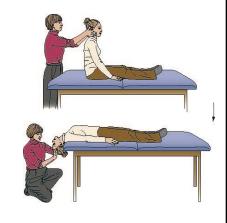
55

Ipsicanal Switch:

Convert Apogeotropic Downbeat to Geotropic Upbeat Nystagmus

Neck Extension (Helminski et al., 2007).

- The patient is brought into the recumbent position with the head extended over the edge of the table. The position is held for 2 minutes to provide adequate time for the debris to settle.
- The patient is returned to the upright position.
- The head is flexed 36° in the chin down forward position to allow debris to settle lowest part of utricle

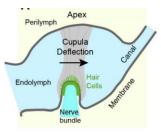


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Interventions for Atypical pc-BPPV

Intervention for both long and short arm side of pc.



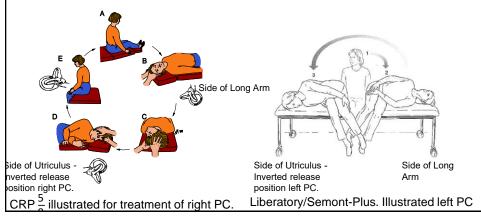
(Rabbitt RD. 2019)

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Intervention for both Long and Short Arm Side

Direction of nystagmus during intervention suggests location os debris.

- Long arm of PC orthotropic nystagmus or excitatory response in dependent positions.
- Short arm of PC excitatory response first position. No response 2nd position and sidelying position of CRP (Ludwig, Schubert 2023).
- Treatment failure excitatory response first position. Inhibitory response 2nd and/or sidelying position of CRP.



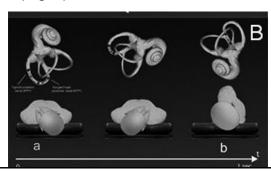
Intervention for both Long and Short Arm Side

Illustrated for Left PC

Quick Liberatory Rotation Maneuver (Califano, Salafia, Mazzone,

Melillo, Califano, 2014).

- From DHT positioning on the affected side to a 45° nose-down position on the uninvolved side.
- Remain in final position for 3 minutes.
- Return to upright position.



Note: Quick Liberatory Rotation Maneuver developed to treat PC jam nonampullary segment.

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Interventions for Atypical pc-BPPV

Intervention for short arm side of pc.

Inverted Release Position – Cupulolithiasis - Side of Utriculus

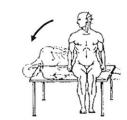
Illustrated for Left PC

Demi Semont maneuver

(Vannucchi, Pecci, Giannoni, et al., 2015)

- Slowly move from sitting to lying on uninvolved side with head turned 45° nose down.
- After 20-30 seconds, quickly move to sitting up.

Note: Demi Semont maneuver developed to treat PC jam non-ampullary segment.





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Inverted Release Position

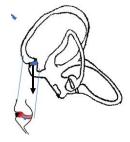
Cupulolithiasis - Side of Utriculus

Illustrated for Left PC

Treated right pc-BPPV cupulolithiasis short arm side or canalithiasis of short arm.

45° Forced Prolonged Position for treatment of PC. Sleep on uninvolved side with head rotated 45° down (Vannucchi et al., 2015).

Note: Developed to treat PC jam non-ampullary segment.





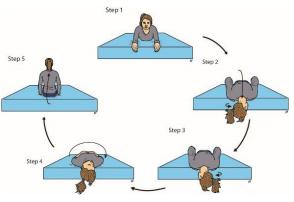
Illustrated Right Side Involvement

62

Intervention pc-BPPV Canalithiasis of Short Arm

Treated pc-BPPV canalithiasis of short arm with:

• Forward Canalith Repositioning Procedure (Faldon and Bronstein, 2008). (Designed to treat AC-BPPV).



Illustrated Right Side Involvement

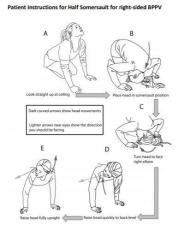
63

Half Somersault (Foster et al, 2012)

Designed to treat long arm canalithiasis.

Suggest treat short arm canalithiasis.

Rx Sessions	Epley	½ Somersault
Two Rx	37%	27%
Six Months	50%	37%

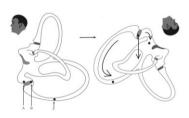


64

Type 2 pc-BPPV:

(Harmat, Tamás, et al, 2022)

- Diagnostic criteria (Buki, Simon, et al, 2011):
 - Symptoms suggestive of BPPV. Episodic vertigo with bending forward, lying down, sitting up, or turning over in bed.
 - No nystagmus during positional testing DHT or SRT.
 - Short episode of vertigo with truncal retropulsion after sitting up from ipsilateral side
- Mechanism pc-BPPV canalithiasis involving short arm.
 Mechanism of Type 2 pc-BPPV supports mechanism of pc-BPPV canalithiasis of short arm.
- Treatment:
 - Repetition of ipsilateral DH provoking position (Harmat, Tamás, et al, 2022).







<u>3</u>5

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Suggested Intervention For Type-2 BPPV

Interventions for pc-BPPV canalithiasis of short arm:

- Ipsilateral DH position (Harmat, Tamás, et al, 2022). Produces nausea sweating.
- Demi-Semont (right sidelying) (Vannucchi et al., 2015).
- Forward Canalith Repositioning Procedure (Faldon and Bronstein, 2008).
- Bow and yaw maneuver (Ping, Yi-Fei, et al, 2020).



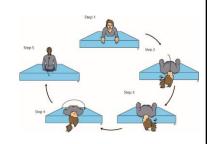


Illustrated Right Side Involvement





Illustrated Right Side Involvement



Illustrated Right Side Involvement

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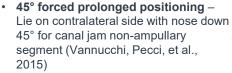
Interventions for Atypical pc-BPPV

Canal jam – back debris out.

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Particle Repositioning Maneuvers for pc-BPPV Jam

- Demi Semont maneuver for canal jam non-ampullary segment (Vannucchi, Pecci, et al., 2015)
 - Slowly move from sitting to lying on uninvolved side with head turned 45° nose down.
 - After 20-30 seconds, quickly move to sitting up.



 Scocco manuever. Treatment of periampullar canal jam involving the PC (Scocco, Garcia, et al, 2019).







Supine - Contralateral Nose Up (Right) side - Nose Turned 45° Down.

Roll onto Back

Nose turned

5° Towards

Ipsilateral (Left)

side.

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Summary: Vertical Canals – Therapeutic Intervention

Goal is to mechanically remove debris out of canal into utricle.

- Canalithiasis Involved canal placed in earth vertical plane and is rotated in the plane of gravity so that the particles gravitate in the opposite direction, out of the canal into the utricle.
- Cupulolithiasis Release particles.
 - Treat both sides of cupula.
 - o Side of long arm SHHP, CRP, or Liberatory Maneuver. The axis of the cupula is aligned with or just beyond gravitoinertial force.
 - o Side of utriculus Inverted position. The axis of cupula is oriented in earth-horizontal.
- **Jam partial or complete.** Back the particles out of the jam, reverse the direction from which the jam was formed.

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