BPPV: what we do and do not know

Daniele Nuti

Robert Barany, Acta Otolaryngol (Stockh), 1921

BPPV was described almost 100 years ago

First description of the disorder in a 27-year-old woman

further characterized almost 70 years ago

Charles Hallpike

Margareth Dix

London 1952: Detailed description of the characteristics of the syndrome and the famous diagnostic maneuver

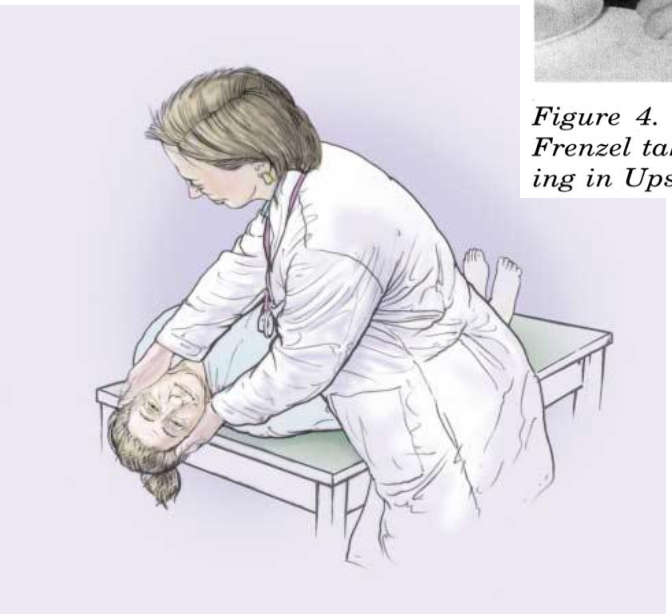
Dix MR, Hallpike CS (1952). The pathology, symptomatology and diagnosis of certain common diseases of vestibular system. Proc R Soc Med 78: 987-1016

Diagnosis and treatment “low cost”

H Frenzel

Figure 4. Photograph of C.S. Hallpike (right) and H. Frenzel taken by C.O. Nylén at the Bárány Society meeting in Upsala, Sweden, May 1963.

C.S Hallpike



- BPPV became widely known in the 1980s, following the therapeutic successes by

- Alain Sémont in Europe (1988)

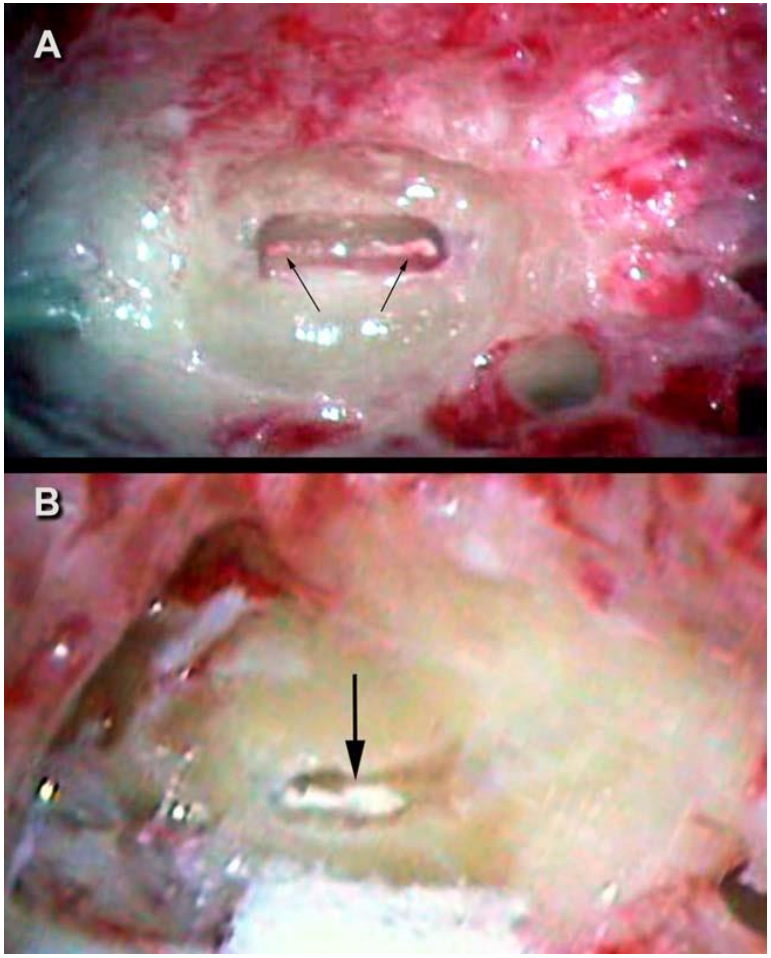
John Epley in USA (1992)

BPPV

- Its pathophysiology is still largely speculative
- Outline of presentation: description of the main features of BPPV, with attention to its more controversial and unexplained aspects.

We know that……

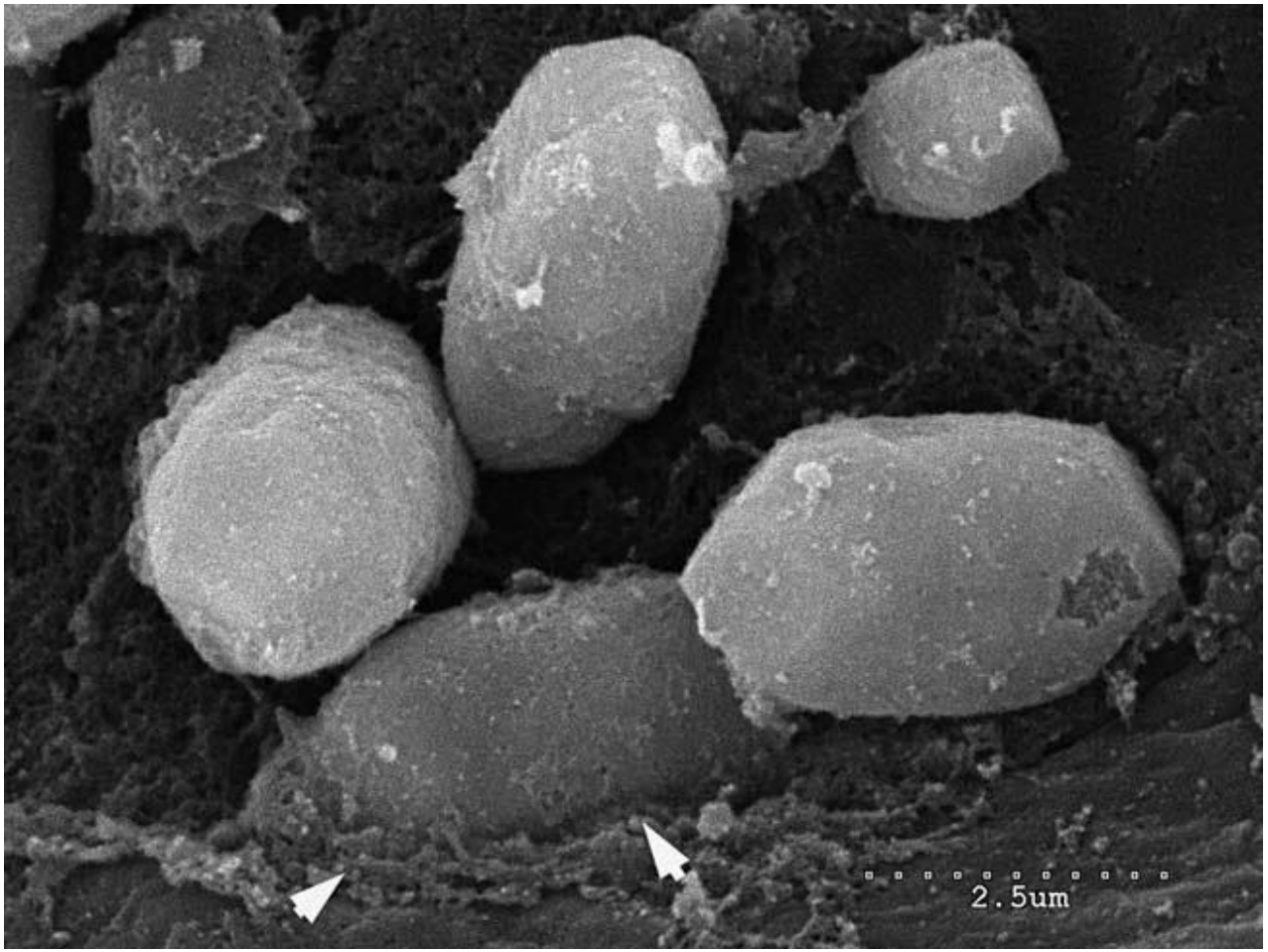
Evidence that the particulate matter found in patients with intractable PC-BPPV is composed of a **conglomeration of typical human otoconia, most of which are attached to a gel matrix** **(Kao et al, 2017)**

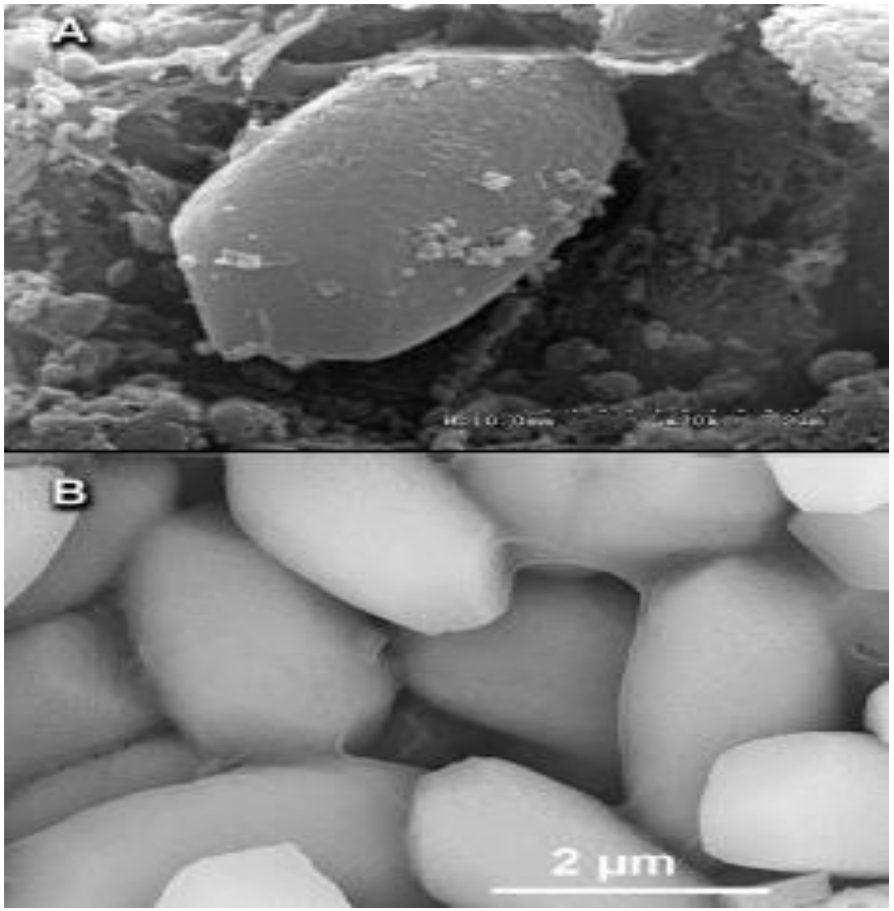


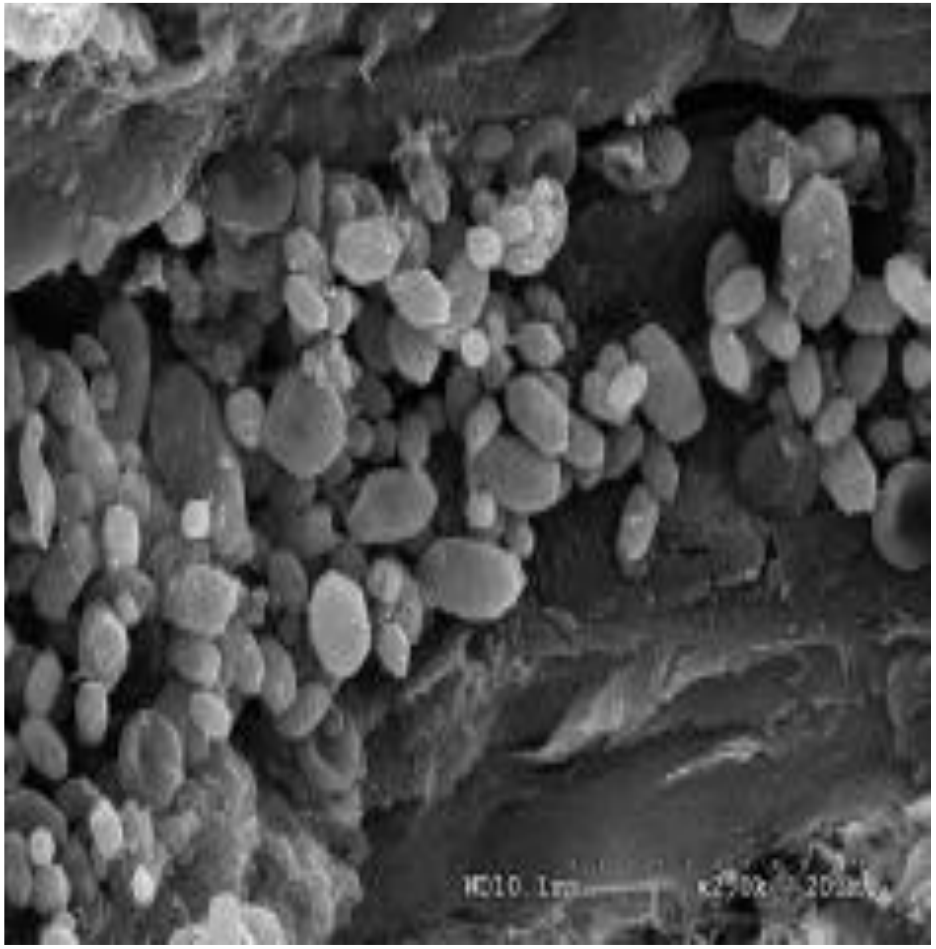
We know that……

Presence of linking filaments between the otoconia and amorphous material

in BPPV, fragments of the otolithic membrane become displaced from the utricle en bloc







Otoconia not always free-floating but also embedded in floating otolithic membranes creating possibilities for peculiar, atypical patterns of benign positional nystagmus (Kao, Parnes 2017)

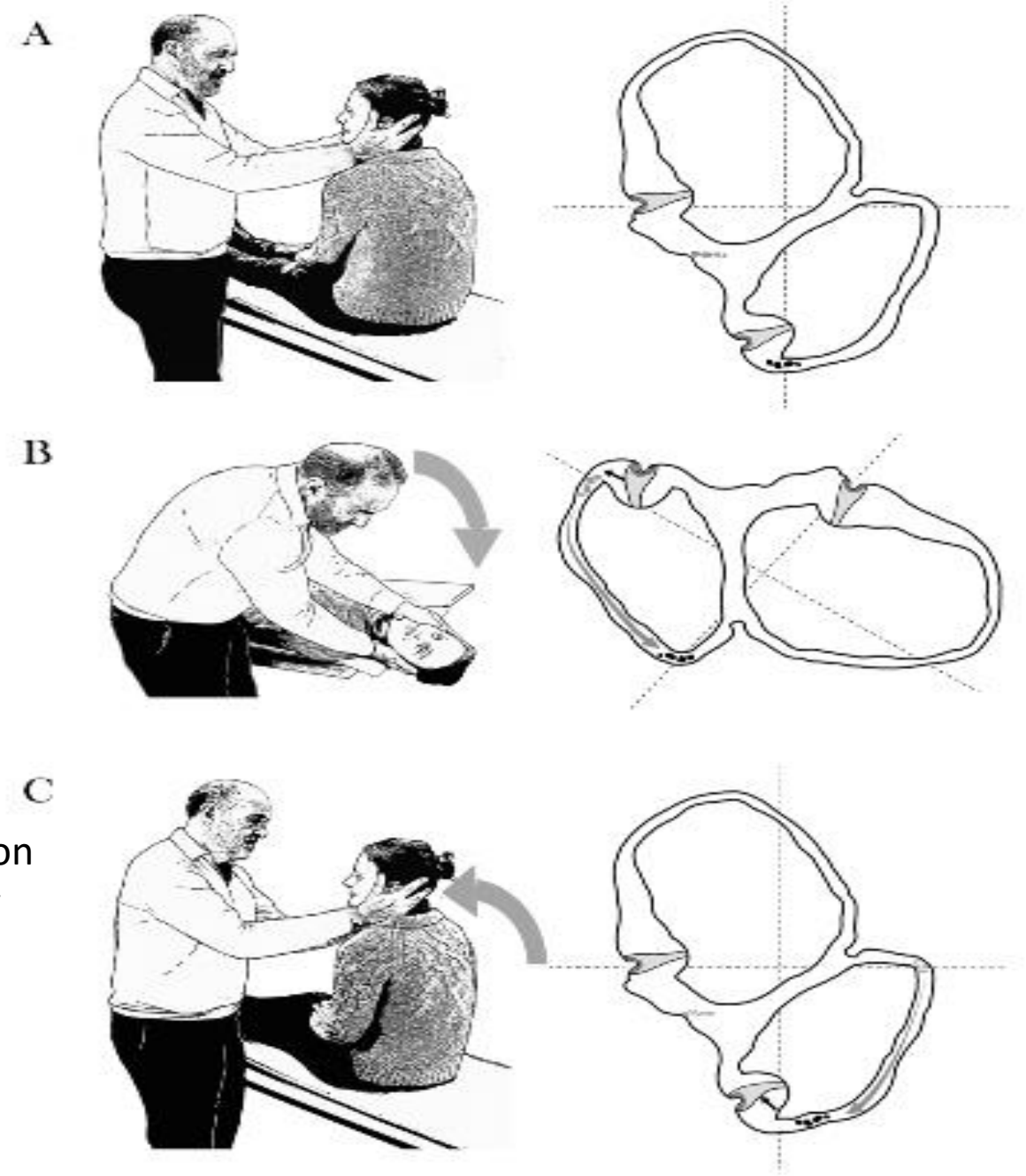
Otoconial debris

- Definitive proof that otoconia are responsible for the disease
- Explanation of some atypical cases
- Gel matrix and filaments: adhesion to the walls of the membranous duct
- abnormal endolymph current with head movements
- Atypical nystagmus patterns (long lasting, not paroxysmal)
- obstacle to the exit of otoconia from the canal (intractable BPPV)

We know that:

PN with D-H maneuver:
- fall of the otoconial mass away from the cupula (excitatory direction)
- Intense torsional PN towards the affected ear

- Sitting up: reverses its direction (less intense torsional PN away from the affected ear)
- Proof that otoconial debris is moving in the canal.

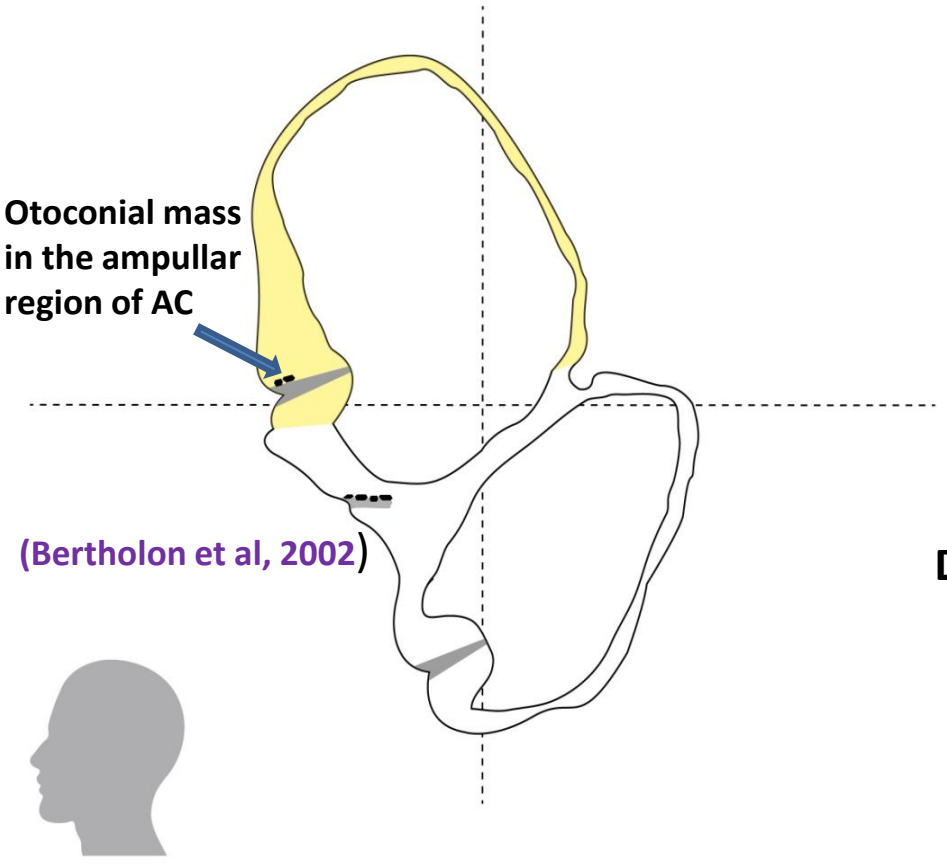


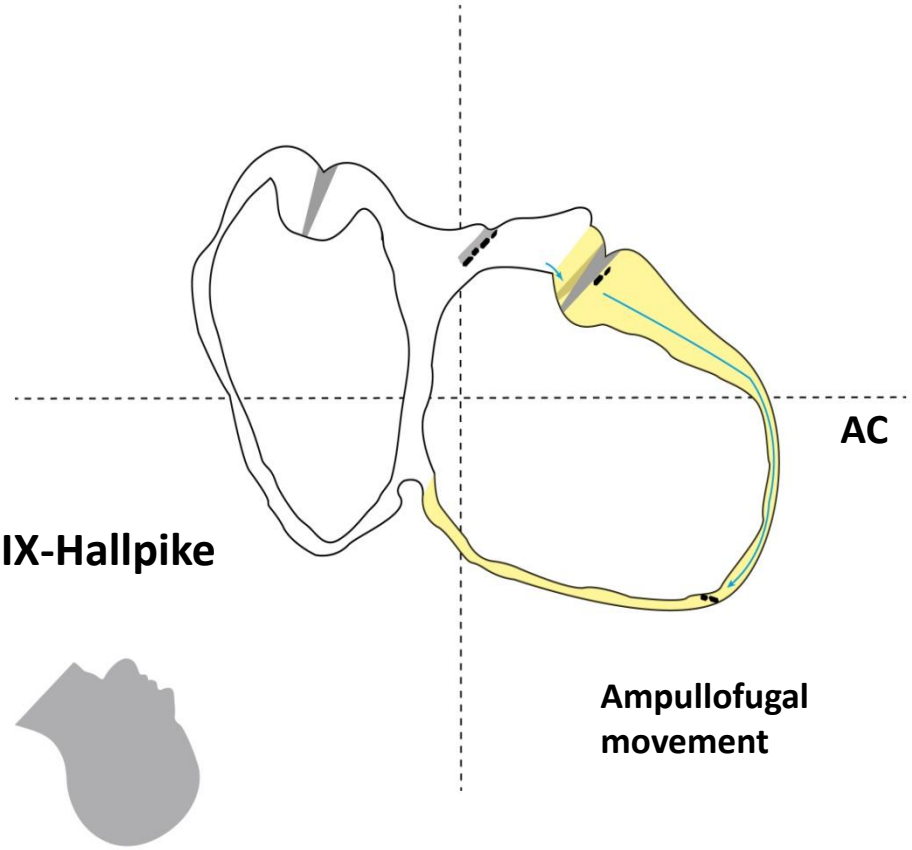
what we don't know

- Positional Downbeating Nystagmus (pDBN)
- Where are the otoconial debris?

AC Canalolithiasis???

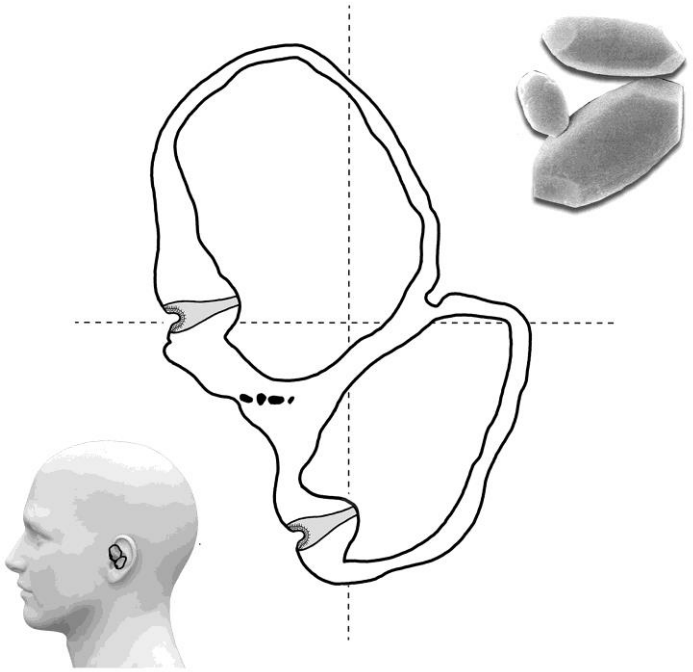
Expected pattern with D-H maneuver:
- positional downbeat nystagmus with a torsional component beating toward the affected ear
- paroxysmal (excitation due to ampullofugal movement)
- when sitting up, nystagmus **should reverse its direction**
- repeatable, with fatigability

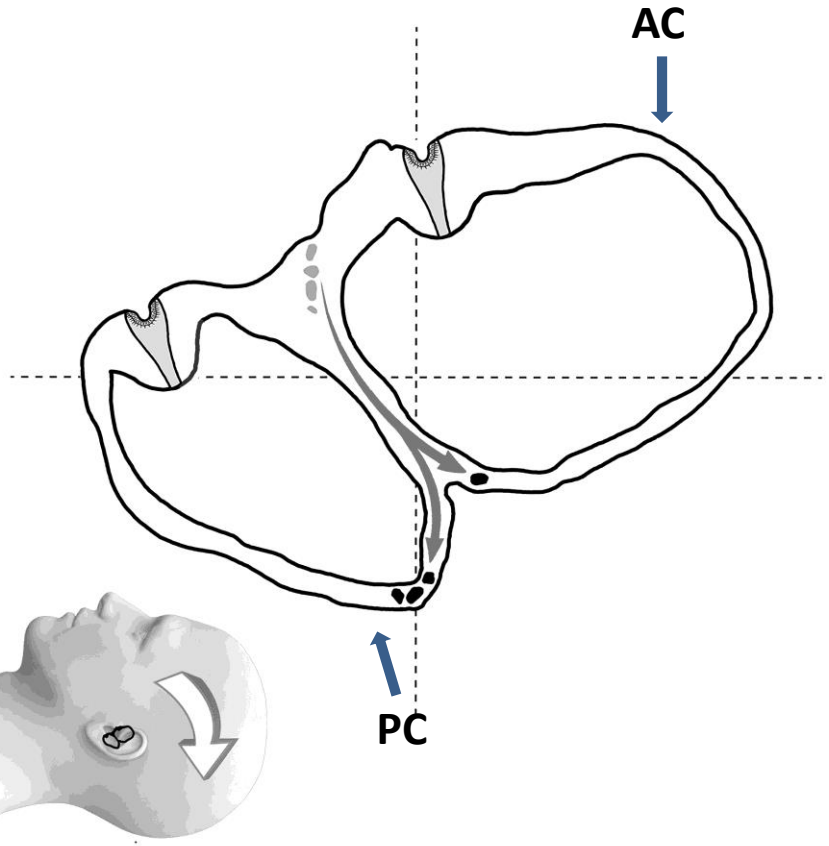




AC canalolithiasis

- Few patients with the expected pattern of AC canalolithiasis
- PN is often not paroxysmal
- No direction reversal when sitting up



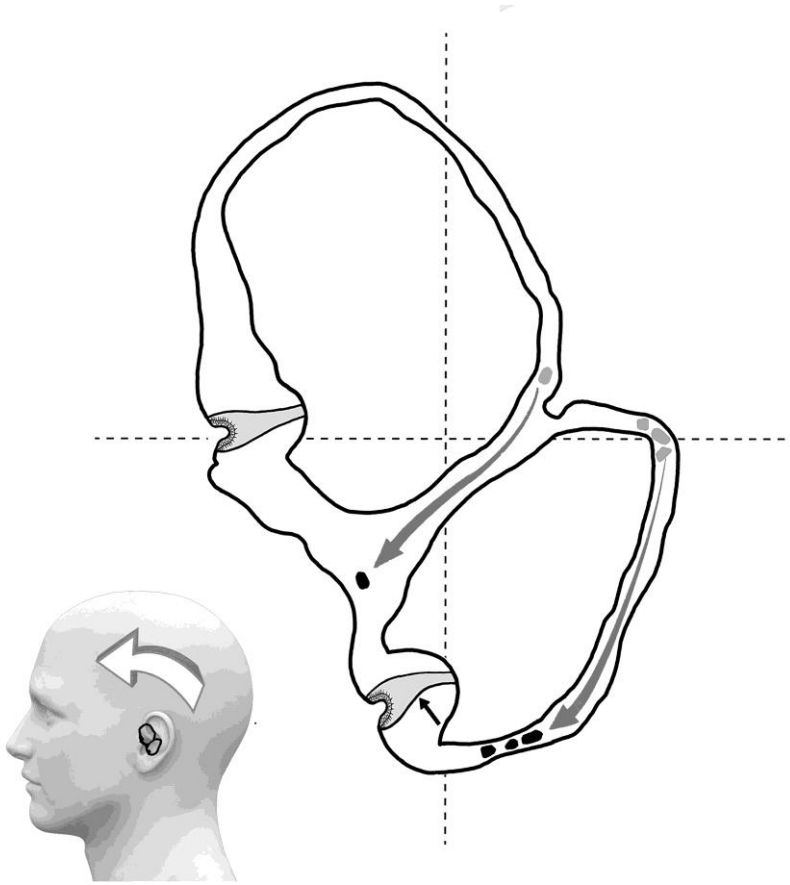


Supine position: common crus immediately below utricular macula

Otoconial mass can approach both PC and AC

Sitting up: the mass should exit due to the orientation of AC

Short-lived syndrome (self-cured)

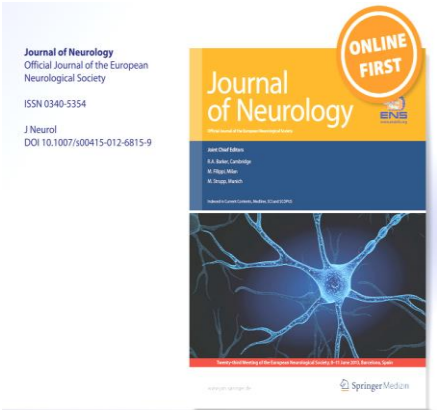


pDBN

- AC BPPV is probably a rare entity
- Very few patients with the expected pattern of AC canalolithiasis
- Positional downbeat nystagmus (pDBN) is not uncommon

Natural course of positional down-beating nystagmus of peripheral origin

Jacopo Cambi · Serena Astore · Marco Mandalà · Franco Trabalzini · Daniele Nuti



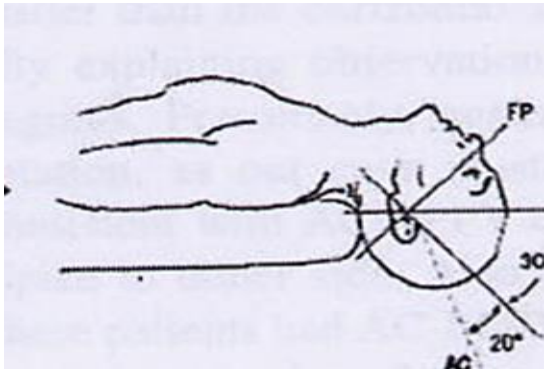
pDBN

- January 2011-April 2012 (16 months): *53 consecutive patients with pDBN*
- 3/53: evidence of CNS involvement (neurological, otoneurological, imaging)
- 50/53: idiopathic (without clinical/imaging evidence of CNS disease)
- Follow up: 1 year
- Age (years): 60 ± 16
- Sex (M/F): 18/32 (ratio 0,6:1)
- Mean time from symptoms onset to examination (days): 10 ± 18

Clinical features summary

- 180 patients (5 years)

- Diagnostic maneuvre:
- The best way to detect pDBN is the SHHP



pDBN features

- pDBN: mostly not paroxysmal, without crescendo-decrescendo pattern
- Detection of torsional component: 35% (Frenzel glasses)
- Pure Vertical: 65%
- Reversal nystagmus, when coming back from D-H or SHHP, detectable in 15% *(in spite of the common presence of vertigo or dizziness)*
- Fatigability: in 80%

pDBN

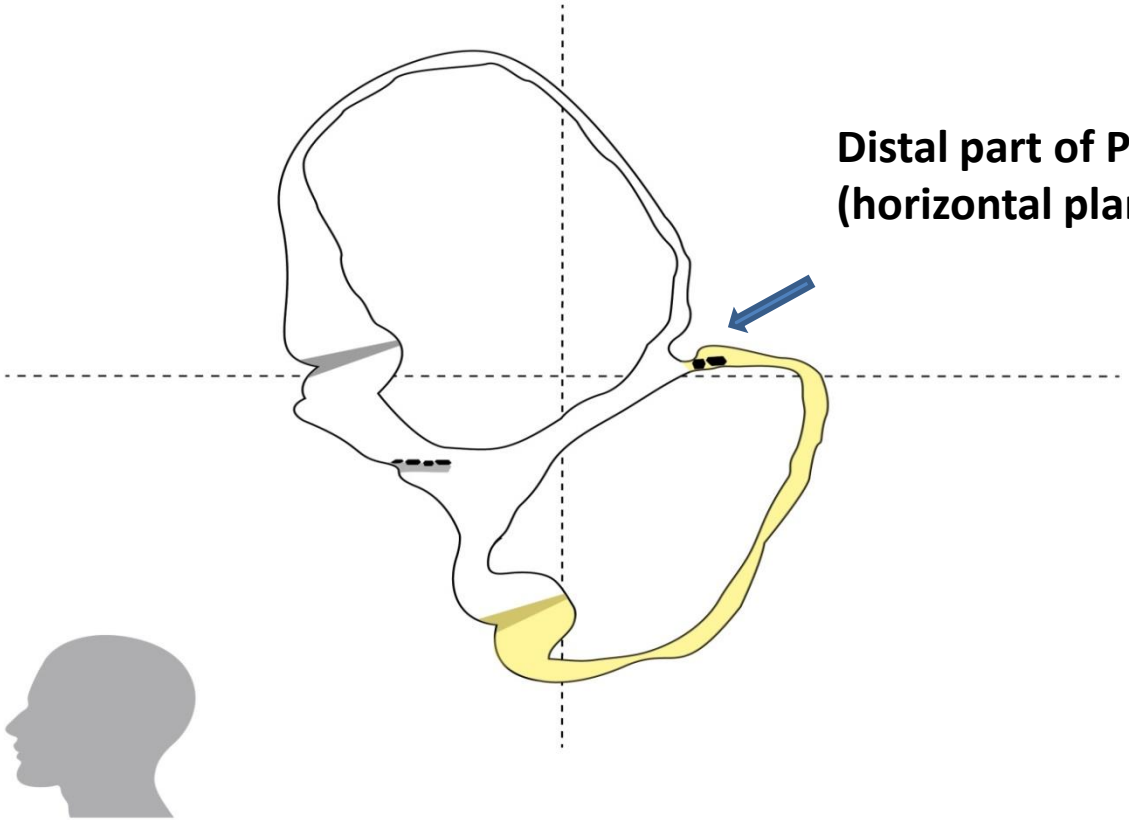
- Around 25% pDBN turns into a typical PC-PPN (also due to therapeutic attempts)
- Around 30%: evidence of previous PC-BPPV

Pathophysiology of pDBN

Excitation due to movement of otoconial mass in *ampullofugal* direction in the anterior canal? (Bertholon et al, 2002)

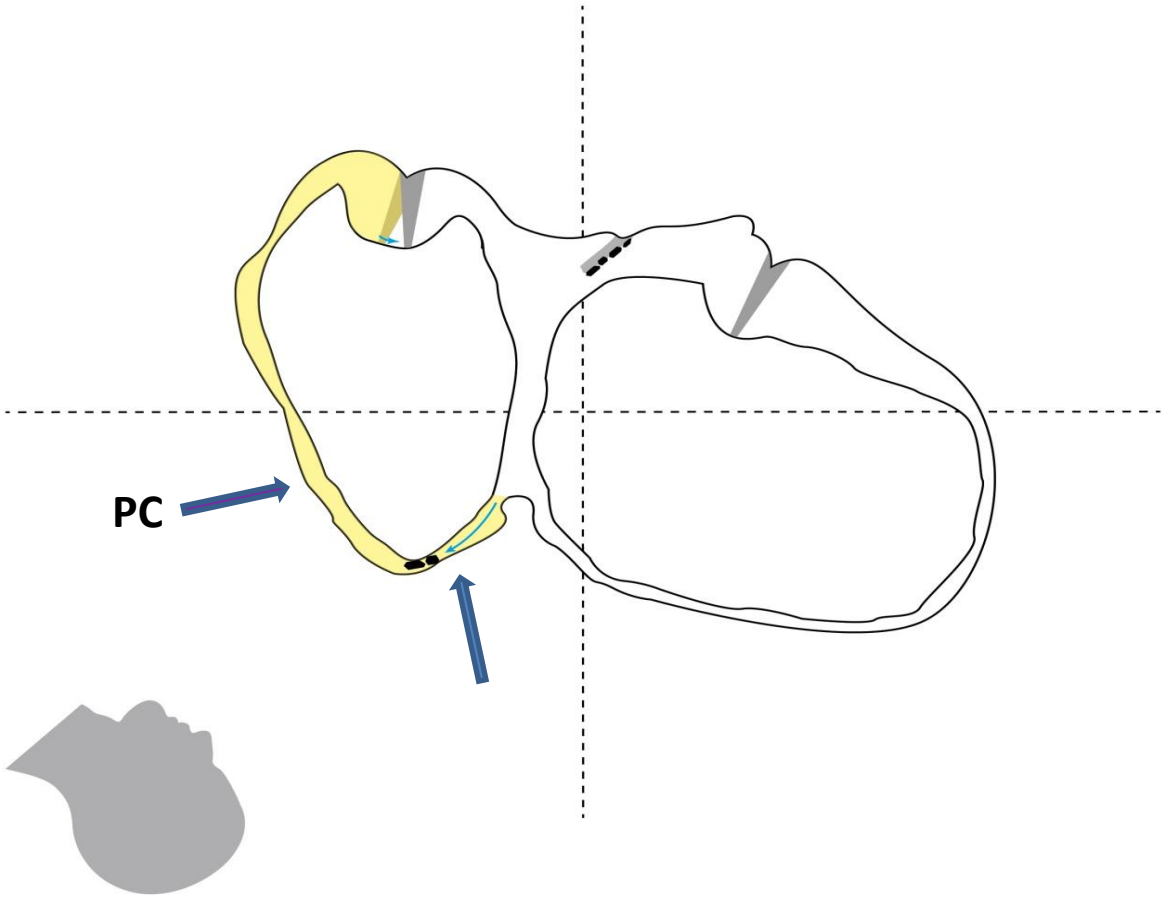
OR

Inhibition due to movement in *ampullopetal* direction in the posterior canal? (Nuti&Yagi, 2010; Vannucchi et al, 2012)

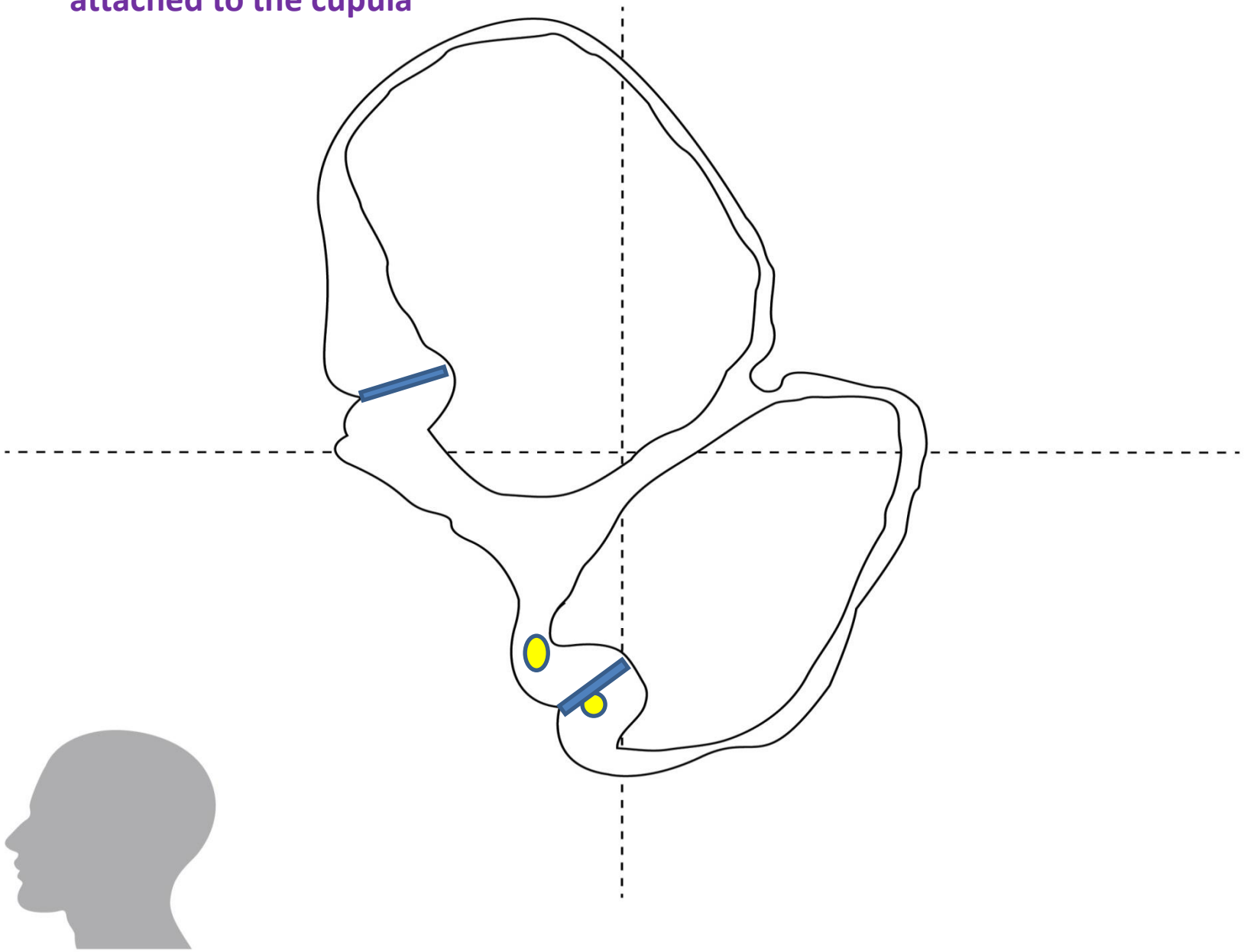


pDBN

-**Dix-Hallpike or SPP**
- **ampullopetal movement**
- **inhibitory pDBN**



Additional theories: otoconia in the short arm of PC or attached to the cupula



Take home messages

- Idiopathic pDBN is considerably more common than has been described in the past
- Its pathophysiology is uncertain
- These patients are reliably affected by a labirinthine disorder because:

- without any evidence of CNS disorder (oculomotor, neurological, images)
- pDBN self-limiting and benign course
- in about 25% pDBN turns into a typical PC-PPN

Take home messages

- Symptoms often different from those of the typical PC or LC BPPV:

- *Unsteadiness when getting up*
- *Prolonged instability (hours-days)*
- *Slight prolonged motion sickness*

- Many patients are missed if not seen in a week
- Spontaneous remission in 50% of patients in the first week and more than 90% within 1 month, without performing any treatment

- AC BPPV??
- Positional downbeat nystagmus of peripheral origin
- “Benign” positional downbeat nystagmus
- Peripheral pDBN

Differential Diagnosis

- Mainly necessary with cerebellar diseases (especially lesions of the **nodulus**)
- pure vertical pDBN: attributed to lesions in the cerebellum or cranio-cervical junction *(Leigh & Zee, 2006)*
- experimental data in animals have linked pDBN with lesions of the nodulus *(Fernandez et al, 1960)*
- family with progressive cerebellar ataxia: pDBN as earlier sign of cerebellar dysfunction- autopsy finding: loss of Purkinje cells primarly in the nodulus *(Kattah & Guirati, Ann. N.Y. Acad. Sci. 2005)*

Differential Diagnosis from nystagmus features?

Peripheral

- With latency in 60%
- Pure vertical in 50%; vertical and torsional in 50%
- Variable duration
- Possible reversal
- Common fatigability
- Associated vertigo/dizziness (almost always)

Central

- No latency
- Pure vertical in 80%

- Variable duration
- Possible reversal
- **Absent fatigability**
- **Uncommon associated vertigo/dizziness**
- **Further neurologic or ocular motor signs and symptoms (gaze SN, headache, dysmetria, severe ataxia)**

Differential diagnosis

pDBN almost always: associated neurologic or ocular motor signs and symptoms of central impairment (gaze nystagmus, saccadic dysmetria, ataxia……)

Otology & Neurotology
35:e204–e205 © 2014, Otology & Neurotology, Inc.

Imaging Case of the Month

Not So Benign Positional Vertigo: Paroxysmal Downbeat Nystagmus From a Superior Cerebellar Peduncle Neoplasm

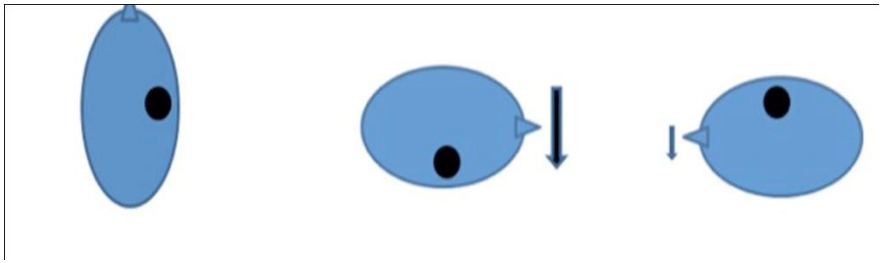
*†Jane Lea, *Corinna Lechner, *G. Michael Halmagyi, and *Miriam S. Welgampola

This report draws attention to the necessity to carefully investigate positional downbeat nystagmus even in the absence of central signs.

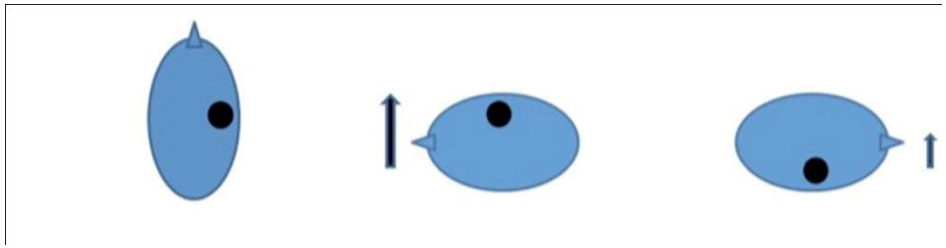
Early Multiple system atrophy

Lateral Canal BPPV: two variants

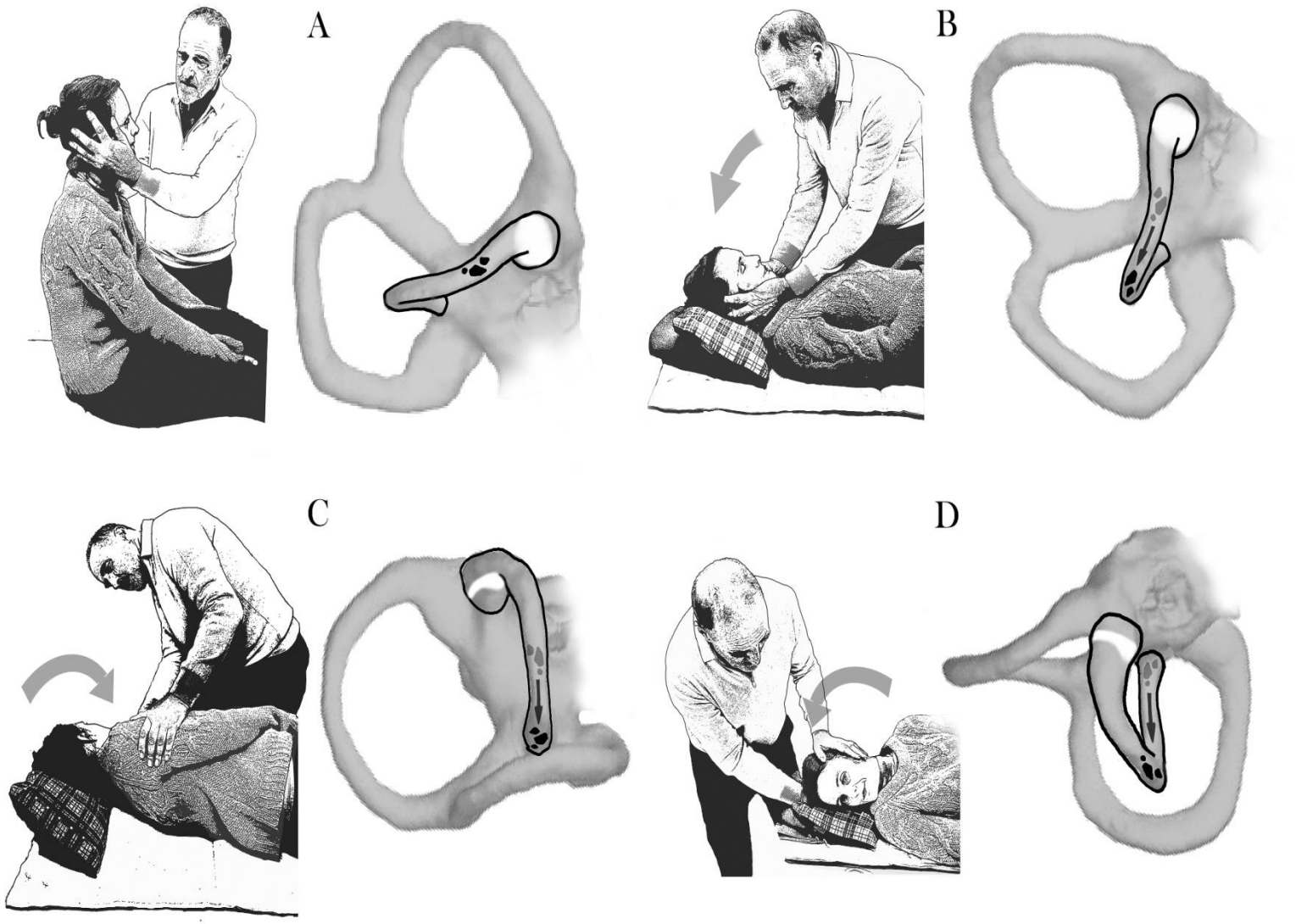
Geotropic Variant (towards the earth): phast phase of PN towards the undermost ear



Apogeotropic variant (away from the earth): phast phase of PN towards the uppermost ear



Supine head-roll test (right LC-BPPV)



Ampullofugal movement – inhibition - PN to the left ear

Ampullopetal movement – excitation - PN to the left ear (more intense)

Left LC-BPPV (canalolithiasis)

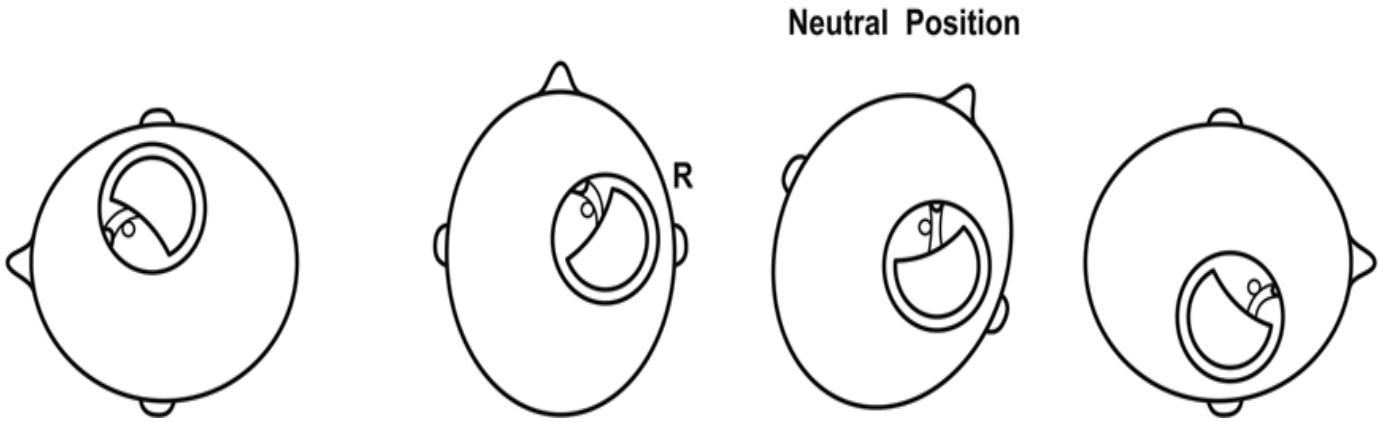
Geotropic variant

- Geotropic, direction-changing, **paroxysmal** PN, with different intensity between the two sides is quite always due to **canalolithiasis**
- No need for further investigations

Geotropic, direction-changing, **persistent** PN has been attributed to **LIGHT CUPULA** **(Hiruma 2004)**

Geotropic variant and light cupula??

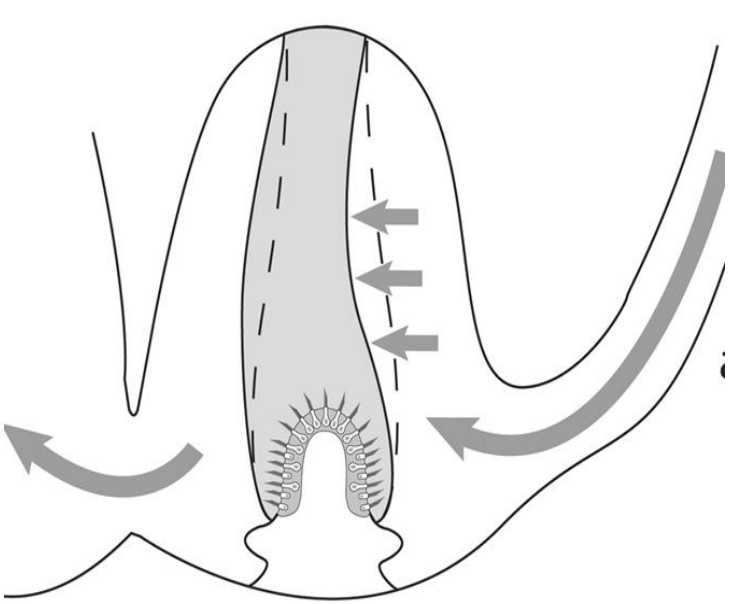
- “lighter cupula” hypothesis (Bergenius and Tomanovic, 2006)
- “heavier endolymph” hypothesis (Kim CH,2014)
- “light cupula due to low density debris” hypothesis (Ichijo H, 2012)

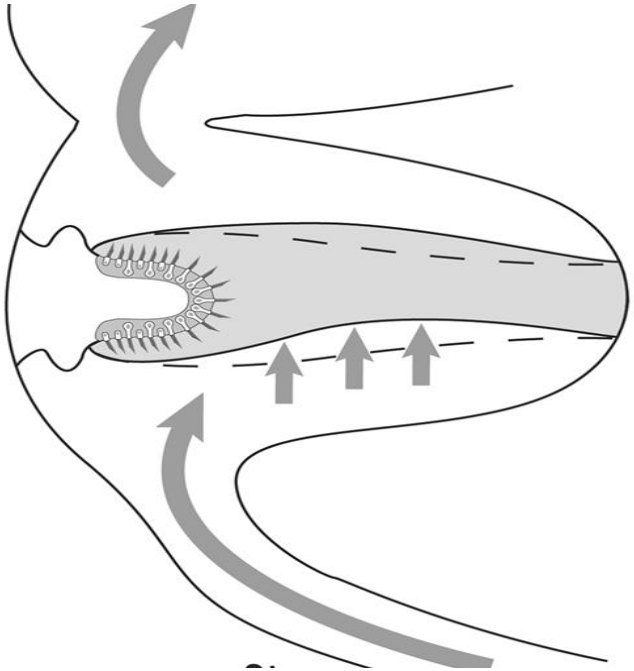


Light cupula concept

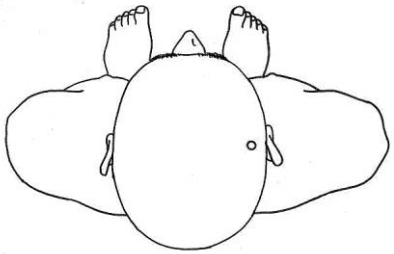
- Positional Alchool Nystagmus (PAN) (Aschan, 1956; Money et al, 1965)
- Diffusion of ethanol from capillaries occurs more rapidly into the cupula than into the surrounding endolymph after alcohol intake, which makes the density of the cupula lower than that of endolymph
- Phase 1 of PAN: lasts for many hours (5-7)

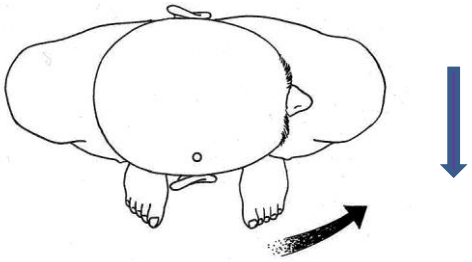
Light cupula (right ear)





Utriculopetal deflection





PN towards the undermost ear

Light cupula

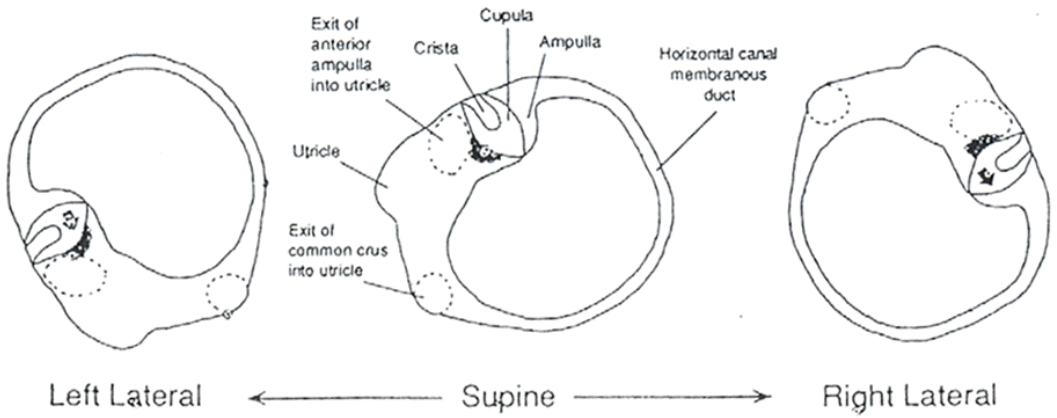
- None of these three hypotheses has been proven yet
- Many inconsistencies (symptoms…)
- Not described for PC
- One or both ears?

- Persistent geotropic PN??
- Light cupula??
- Something similar to a “canal plugging” that provokes a persistent stimulus (lack of adaptation)

Apogeotropic t direction-changing PN

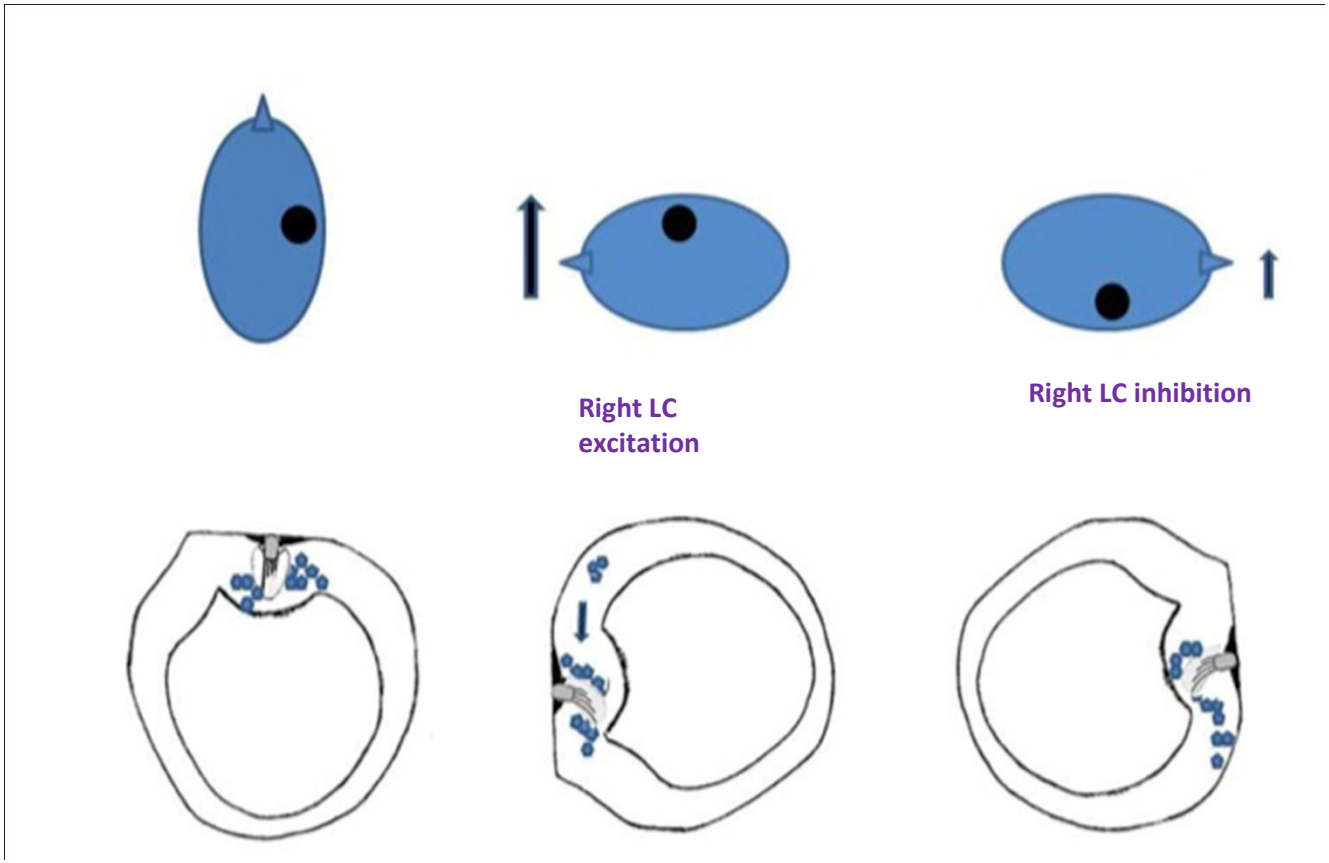
Often considered as a synonymus of cupulolithiasis (heavy cupula)

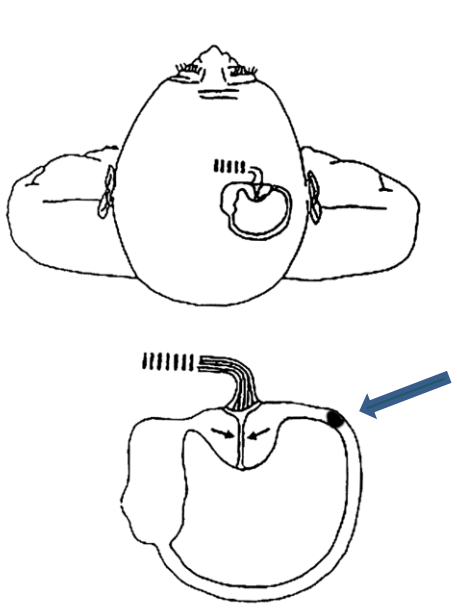
Baloh et al, Neurology 1995



Apogeotropic persistent direction changing positional nystagmus

Heavy cupula concept: sensible to gravity vector ***(Baloh et al, Neurology 1995)***





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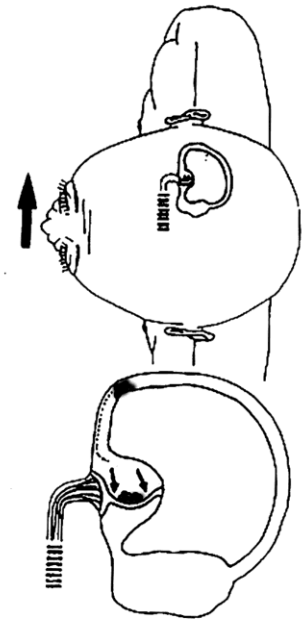

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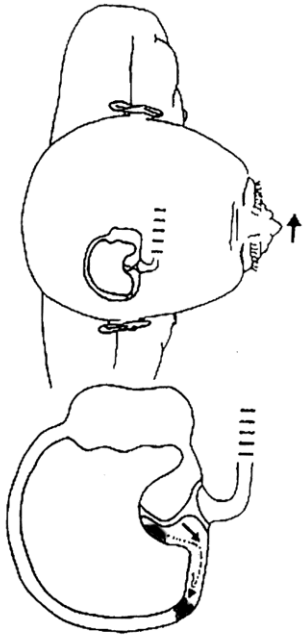
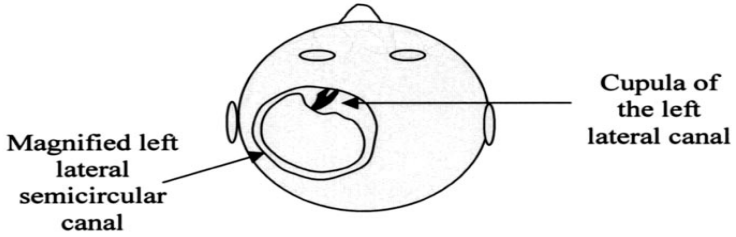


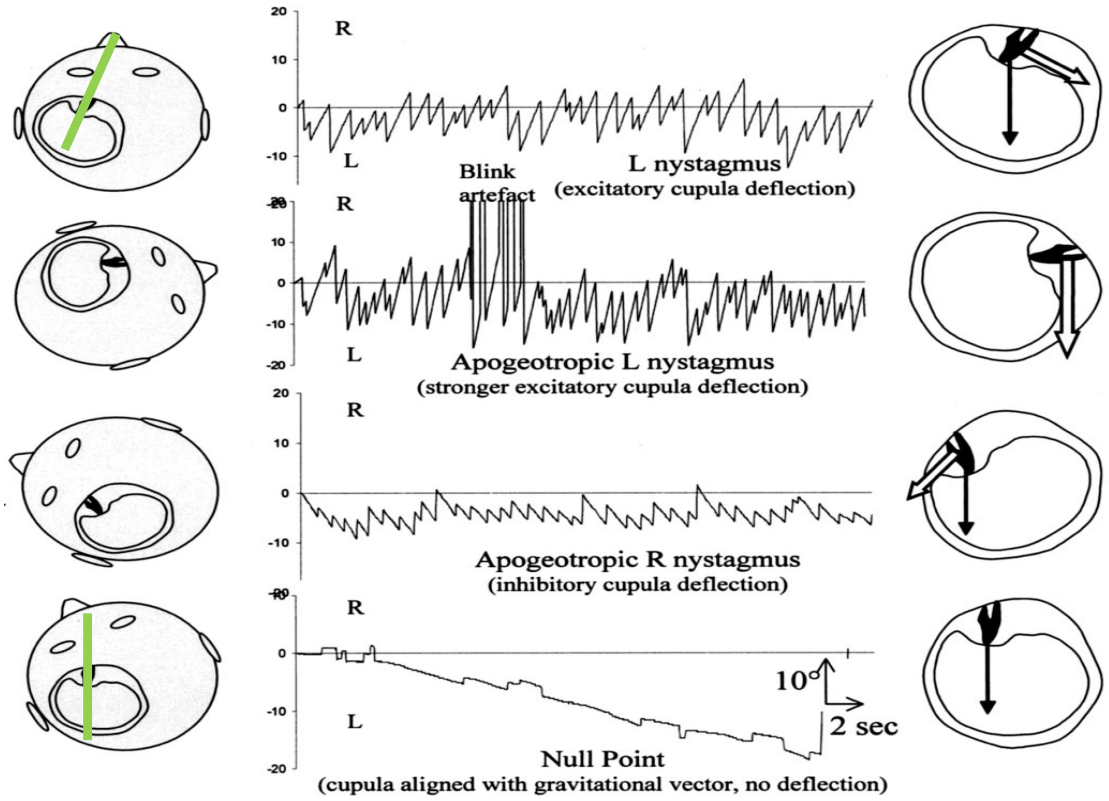
C

Null Point

The “null point” theory predicts that the cupula is not aligned with gravity when the patient is in the supine position with the straight head

“the slight rotation of the head to one side corresponds to a position where the cupula of the ipsilateral lateral canal is aligned with the gravitational vector.
Therefore, no deflection to either side would occur, even if hyperdense debris were attached to it”.

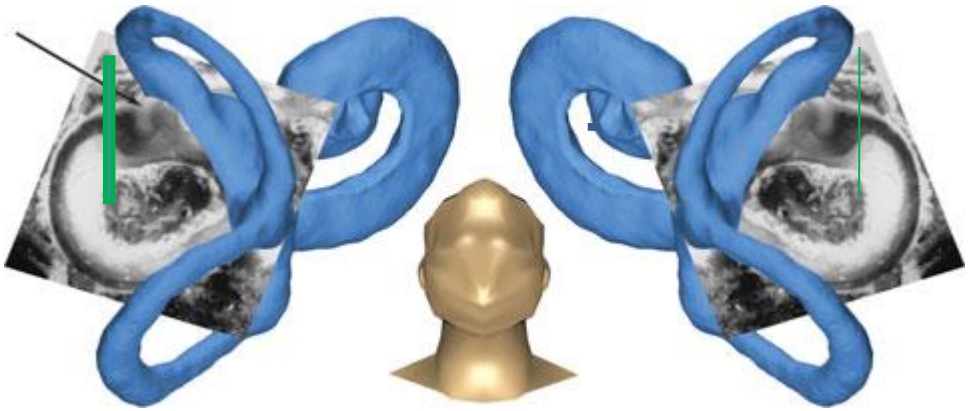


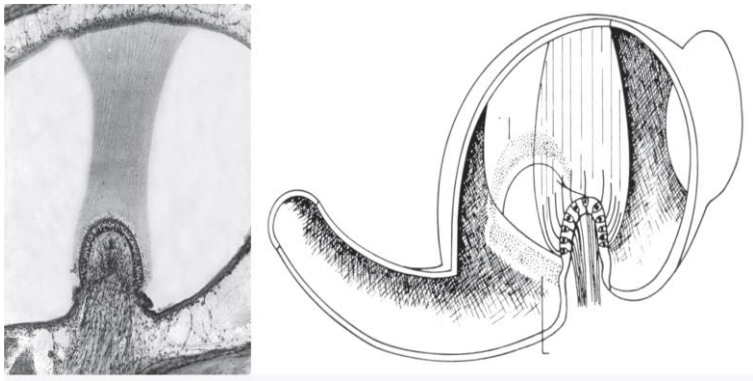


Bisdorff & Debatisse, Neurology 2000

Cupula orientation

- “It is clear…..that the crista and cupula of the horizontal semicircular canals are close to being parallel to the median plane of the head, even allowing for errors…..”(Curthoys et al. 2009)





Microtomography-CT Studies of the Membranous Labyrinth

- Null point theory would be discredited
- Further elements in favour of canalolithiasis

- transformation from apogeotropic to geotropic variant following gentle head movements (bending forward or backward) or with prolonged position with the affected ear up
- Rarity of the cupulolithiasis phenomenon in the PC compared to the high frequency in the LC

Thank you!!