17 th Workshop on NEUROTOLOGY and



BPPV

- Most frequent vestibular disease
- Most common cause of vertigo in humans
- Lifetime prevalence: 2.4%
- 1 year incidence: 0.6% (von Brevern, 2007)
- This means that about 1 million adults suffer from BPPV each year in Germany
- Around 60 years of age
- Famale/male ratio: 2/1
- 70% PC-BPPV (right vs left side: 1.5/1)
- 15-20% LC-BPPV
- 50% long-term recurrences
- 5% bilateral-multicanal (90% post-traumatic)

Robert Barany, Acta Otolaryngol (Stockh), 1921

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



Margaret Dix & Charles S. Hallpike (1952)

•



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.





Dix and Hallpike described in detail the characteristics of the syndrome in 100 patients

"Positional nystagmus of the *benign* paroxysmal type"

- "the lesion certainly affects the otoliths and, since it is so often associated with normal caloric responses...it is more likely to be **irritative** in character than destructive"
- Description of the diagnostic maneuver

Cupulolithiasis (Schuknecht HF, 1969)

Histopatologic finding of basophilic deposits in the cupula of the PC of 2 patients with history of BPPV



PN is generated by a gravity-sensitive PC (heavy cupula concept)

The disorder is caused by calcium carbonate deposits on the cupula of the PC that made the cupula sensitive to gravitational forces

Canalolithiasis (Hall, Ruby and Mc Clure 1979)



Suggestion that the pathogenetic mechanism is due to something moving inside the endolymph of the canal, rather than adhering to the cupula of the PC

The concept of canalolithiasis was supported by the intraoperative observation of abundant free-floating debris in the endolymph of the posterior semicircular canal (Parnes & McClure, 1992)

Why do otoconia detach?

- The most logical reason for otoconia detachement is trauma (about 15% of cases)
- head/body trauma (often bilateral)
- Continuous jarring (mountain-biking, skeet shooting)
- High-impact aerobics
- Surgery with the use of a drill (nasal, dental)
- Cochlear implants, stapes surgery

Why do otoconia detach spontaneously?

- BPPV occasionally occurs after assumption of unusual head positions (e.g., prolonged reclining in a dentist's chair, at the hairdresser's, or working underneath a car)
- Following prolonged bed rest and sleeping (Ichijo, 2016)
- Probably everyone has free-floating otoconia in the endolymph, especially in older people (Kveton et al, 1994)
- The syndrome is triggered when the otoconial mass is "critical" and the head is positioned such that the debris can enter the canals

Predisposing factors for BPPV and its recurrences

- Migraine is three times more common in idiopathic than post-traumatic BPPV. Vasospasm of the inner ear can lead to detach of otoconia from the maculae (Ishiyama et al. 2000)
- Possible association between BPPV, osteoporosis and disorders of calcium metabolism (Vibert et al 2003, Yamanaka, 2013)
- Link between otolithic disturbances and vitamin D deficiency is highly probable (Büki et al. 2012, Jeong et al., 2013)
- Vestibular Neuritis: PC BPPV, same side, 9.8 prevalence (Mandalà et al., 2010)

TYPICAL AND ATYPICAL BENIGN PAROXYSMAL POSITIONAL VERTIGO

-	Short arm canalolithiasis	Cupulolithiasis	Long arm canalolithiasis
Horizontal canal	Apogeotropic horizontal (beating laterally up in lateral supine posi- tion when the involved ear is lower- most and no nystagmus in the con- tralateral lateral position)	Apogeotropic horizontal (beating laterally up in both lateral supine positions; stronger on the contralat- eral side, when the involved ear is uppermost).	Geotropic horizontal (beating laterally down in both lateral supine positions) stronger on the ipsilateral side, when the involved ear is low- ermost.
			Peri-cupulo canalolithiasis can cause an apo- geotropic horizontal nystagmus (beating lat- erally up in both lateral supine positions; stronger on the contralateral side, when the in- volved ear is uppermost).
Posterior canal	No nystagmus in Dix-Hallpike po- sition but sitting up vertigo either in the RALP or LARP plane	Depending on anatomy: either no nystagmus in Dix-Hallpike posi- tion, or apogeotropic downbeat	Geotropic-torsional upbeat nystagmus with a short latency, provoked when the affected ear is lowermost in the Dix-Hallpike position
Anterior canal	Sometimes acutely caused by the Dix-Hallpike manoeuver: apogeotropic downbeat	Same as anterior short arm and long arm canalolithiasis, more persistent repeatable	Apogeotropic downbeat nystagmus with a small torsional fast-phase, which beats toward the affected ear (affected ear uppermost in Dix-Hallpike position) (Occurs possibly rarely because the position of the canal)

Buki, Mandalà & Nuti, 2014

PC-BPPV



Vertical upbeating torsional nystagmus (with the upper pole of the eyes beating toward the lower ear – Barany 2014)

PC excitation: ipsilat superior oblique and contralat. Inferior rectus muscles

Treatment of PC BPPV

 Brandt & Daroff (1980): first physiological approach to the treatment of BPPV, which had the purpose of dispersing the debris within the semicircular canals



Brandt-Daroff Exercises

Series of nonspecific repetitive exercises with the aim of obtaining -Dispersion of debris -Dislodgement of otoconia attached to the cupula -To promote habituation

Still used for intractable BPPV patients

Level C of the Classification of Recomendations of the American Academy of Neurology
Possibly effective



Canalith Repositioning Procedure by John Epley

Devised 1979 Published 1992



 Probably the most widely adopted treatment in the world

Designed to allow debris to migrate by gravity out of the PC through the common crus







SÉMONT LIBERATORY MANOEUVRE (1983-1988)



KEY POINTS for success:

- Rapid maneuver (Faldon and Bronstein, 2008)
- Orientation of the canals (Lyu et al., 2016)
- Liberatory nystagmus is nearly always a good prognostic sign
 - Check within 1 hour



PC TREATMENT

- Double blind randomized trials on CRP (Lynn et al, 1995; von Brevern et al, 2006) and Sémont maneuver (Mandalà et al, 2012; Chen et al, 2012) allow to consider both treatments as effective and safe therapy that should be offered to patients of all ages
- They belong to the level A of the Classification of Recomentations of the American Academy of Otolaryngology and of Neurology
- Level A: Treatments with established efficacy
- About 80% of patients symptom-free with the first therapeutic session

Lateral (horizontal) canal BPPV in the geotropic form

• Joseph Mc Clure (1984)

Horizontal Canal BPPV, Am J Otol 1985

Luciano Cipparrone et al (1985)

Lateral Canal BPPV

- Diagnostic Test: Supine head roll test
- Barany position
- Pagnini-McClure test



LC typical Nystagmus features

- Horizontal
- Direction-changing
- Geotropic (towards the lowermost ear)
- Paroxysmal, transitory
- Usually more intense towards one side (affected side)
- No difference between the two sides

Geotropic PPN



- Geotropic, direction-changing, paroxysmal PN, with different intensity between the two sides is always due to canalolithiasis

 No need for further investigations with no additional neuro-otological findings

Persistent geotropic, direction changing PN has been attributed to LIGHT CUPULA (Hiruma, 2004, Bergenius 2006)

Similarities with post-alcoholic positional nystagmus in unilateral labirinthectomy subjects (Tomanovic and Bergenius, 2011)

LATERAL CANAL BPPV MANAGEMENT

- It is first necessary to identify the affected side: wrong identification causes the debris to move towards the ampulla instead of towards the utricle and causes geotropic nystagmus to become apogeotropic
- Barbecue rotation (Lempert-Tiel Wilck, 1994)
- Forced Prolonged Position (FPP) (Vannucchi et al., 1994)
- Liberatory manoeuvre (Gufoni-Mastrosimone (1999)

FORCED PROLONGED POSITION



Very simple method: patient merely has to lie on the healthy side (**with affected side up**) for as long as possible. Outcome treatment: 1-2 days later

Barbecue (Lempert)

- 38 patients
- 24 symptom free (63%)
- 4 PC
- 2 apogeotropic

FPP (Vannucchi)

- 56 patients
- 41 symptom free (73%)
- 2 PC

Nuti D. et al. The management of horizontal-canal paroxysmal positional vertigo. Acta Otolaryngol. (Stockh.), 1998 **118: 445–460**

VS

Treatment of *lateral canal* **BPPV**

Single Liberatory Manoeuvre

Gufoni-Mastrosimone, 1999





Lateral canal treatment

Gufoni's maneuver has been recently validated with randomized double blind trials (Mandalà et al, 2013; Kim JS, 2013)
Level A of the Evidence Based Medicinel



We usually perform a single liberatory manoeuvre and then suggest the patients to lie on the healthy side all the following night.





Lateral Canal BPPV

- Pseudo-spontaneous Nystagmus (sitting position)
- McClure 1985, Bisdorff & Debatisse 2001
- Choung et al 2006, Asprella Libonati 2008
 (so-called pseudospontaneous nystagmus because there is no primary labyrinthine hypofunction)

- Bow and lean test (Choung et al., 2006)
- Head Pitch Test (Asprella Libonati, 2008)

The affected ear is the same direction of bowing nystagmus in canalolithiasis and the same direction of leaning nystagmus in cupulolithiasis.



Sitting position: horizontal, long lasting, not paroxysmal, nystagmus beating away from the affected side.

It disappears when the head is bent 30° forward, with the LC on the horizontal plane



GEO

BOW Inverts its direction with the head 60° forward

LEAN

Increases its intensity by bending the head backwords and/or reaching the supine position

Better seen with videonystagmoscopy

Courtesy of Asprella Libonati

Apogeotropic nystagmus

- Horizontal
- Direction changing
- Beats towards the **Uppermost** ear on the two sides
- Usually more intense towards one side (affected side)
- Persistent and long lasting
- Paroxysmal and transitory (lasting longer than in PC-BPPV)
- Static reversal (secondary nystagmus)







How to manage LC BPPV in the apogeotropic form?

- Many suggestions in recent years
- FPP and/or <u>Gufoni's maneuver</u> on the affected side and then on the healthy side if apogeotropic nystagmus has become geotropic (two steps)
- Gufoni's modified maneuver (one step)





Modified Gufoni (38/52, 73.1%) vs head-shaking (33/53, 62.3%) [p=0.129]

Randomized clinical trial for apogeotropic horizontal canal benign paroxysmal positional vertigo.

KIM et al, 2<u>012</u>

LC-BPPV (APOGEO) DIFFERENTIAL DIAGNOSIS AND SIDE IDENTIFICATION

Look for pseudo-spontaneous nystagmus

Bow and lean test

Try to treat it or transform it (Gufoni maneuver) Supine HST

Unilateral vestibular loss (caloric/HIT)



THANK YOU