17th 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
- Female/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)
First description of the disorder in a 27-year-old woman

Margaret Dix & Charles S. Hallpike (1952)

- 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)

Histopathologic 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 McClure 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 detachment 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

<table>
<thead>
<tr>
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<th>Short arm canalolithiasis</th>
<th>Cupulolithiasis</th>
<th>Long arm canalolithiasis</th>
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<tbody>
<tr>
<td><strong>Horizontal canal</strong></td>
<td>Apogeotropic horizontal (beating laterally up in lateral supine position when the involved ear is lowermost and no nystagmus in the contralateral lateral position)</td>
<td>Apogeotropic horizontal (beating laterally up in both lateral supine positions; stronger on the contralateral side, when the involved ear is uppermost).</td>
<td>Geotropic horizontal (beating laterally down in both lateral supine positions) stronger on the ipsilateral side, when the involved ear is lowermost.</td>
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<td><strong>Posterior canal</strong></td>
<td>No nystagmus in Dix-Hallpike position but sitting up vertigo either in the RALP or LARP plane</td>
<td>Depending on anatomy: either no nystagmus in Dix-Hallpike position, or apogeotropic downbeat</td>
<td>Geotropic-torsional upbeat nystagmus with a short latency, provoked when the affected ear is lowermost in the Dix-Hallpike position</td>
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<td><strong>Anterior canal</strong></td>
<td>Sometimes acutely caused by the Dix-Hallpike manoeuvre: apogeotropic downbeat</td>
<td>Same as anterior short arm and long arm canalolithiasis, more persistent repeatable</td>
<td>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)</td>
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Buki, Mandalà & Nuti, 2014
PC-BPPV

Vertical upbeat 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
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 Recommendations 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 Recommendations 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
Left LC BPPV in the geotropic form
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

A

B

C

ampullopetal

ampullofugal
- 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 labyrinthectomy 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 apo-geotropic

• 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) vs FPP (Vannucchi)

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

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

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 Medicine

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 labyrinthisis 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

**BOW**
Inverts its direction with the head 60° forward

**LEAN**
Increases its intensity by bending the head backwards 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 apogeotrophic form?

- Many suggestions in recent years

- FPP and/or Gufoni’s maneuver on the affected side and then on the healthy side if apogeotrophic nystagmus has become geotropic (two steps)

- Gufoni’s modified maneuver (one step)
Randomized clinical trial for apogeotropic horizontal canal benign paroxysmal positional vertigo.

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

KIM et al, 2012
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