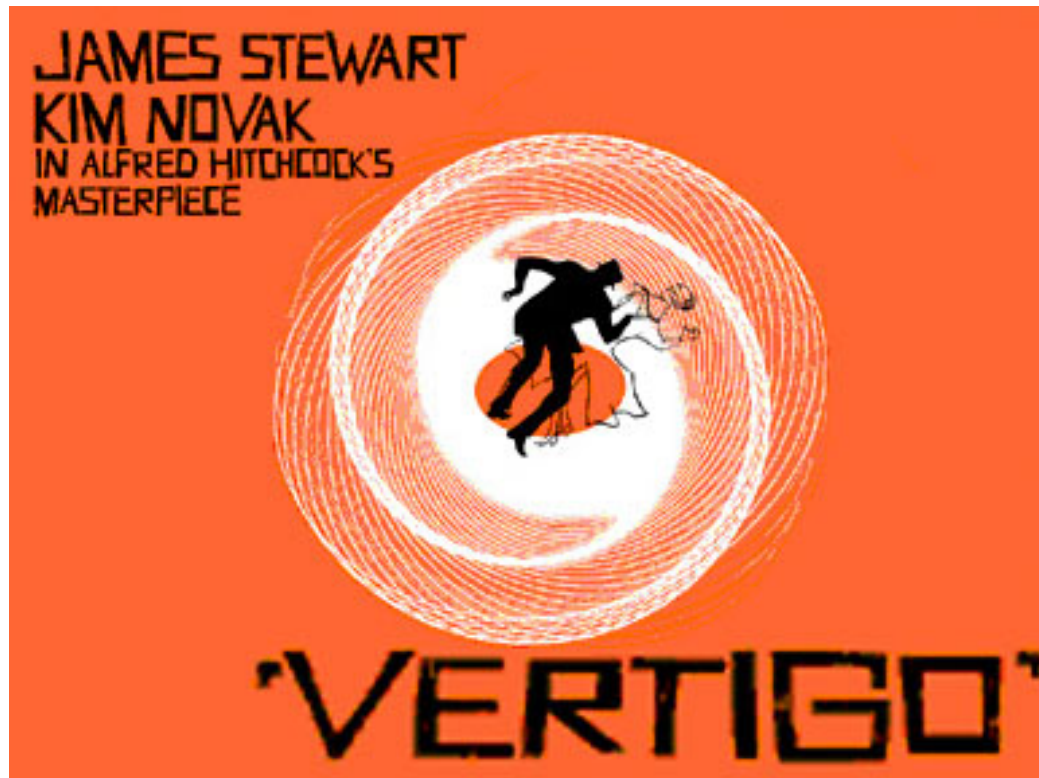


# Neurotology for dummies

Mikael Karlberg

ÖNH Lund -Trelleborg



# Aims:

1. Karlberg's basic clinical 5 minutes exam (+ video-Frenzel and a tuning fork 256 Hz)
2. Some tests for specific questions (fistula test, vestibulo-spinal tests, cerebellar tests...)
3. Acute vestibular syndrome
4. Some cases
5. Ask questions!!! All the time (>30 years experience!!)

# Vertigo is common

- 1,3% of all visits to emergency departments in USA
- 2,3% of all visits to emergency departments in Skåne 2010 (= 7500, 50% admitted)
- 2,1% of all visits to Umeå E.D. 2012-2013  
750/year      65% admitted      45% "medical diagnosis"

# Acute vertigo/dizziness i northern Sweden NUS Umeå 2012-2013 (population 150.000)

1437 pat > 18 ys *dizziness* = 2.1%

65% admitted

39% CT / MR

54% symptom diagnosis only R42.9

14% oto-vestibular (BPPV 6.5%)

9% cardio-vascular

9% neurologic

5.4% stroke / TIA

CT brain: 4.8% pathology

MR brain 13.5% pathology

Acute vestibular syndrome (AVS): 11% stroke

Focal neurological findings / or ataxia:  
53% stroke!!!

0.56% will have a stroke within 3 months

*Ljunggren & Salzer pers rep 2015*

# 1/3

of patients in Umeå admitted with vertigo/dizziness had undiagnosed

## BPPV!

They could easily be treated!

The treatment results were good!

Doctors in emergency clinics must learn how to diagnose and treat BPPV!

*Ljunggren & Barrenäs pers rep 2016*

# CT vs MR vid stroke *Chalela et al,* *Lancet 2007*

- “in 190 patients (3-12h) with final clinical diagnosis of ischaemic stroke, independent, blinded assessment with MRI diagnosed ischaemic stroke in 83% of patients, and in **16% with CT**”
- Acute CT does NOT rule out stroke!
- but can find bleedings/tumors?

# How common is "vertigo" in intracranial bleeding?

595 patients with intracerebral bleedings (8 år)

2.2% (13 of 595): vertigo + NIHSS < 2

Only 1 patient had "only vertigo" but obvious  
dysmetria in status

The rest had headache / confusion / hemiparesis /  
dysarthria mm

Do we have to rule out bleedings in "only vertigo"?

Kerber et al. *Emerg Med J*. Publ on line Jan 18, 2011

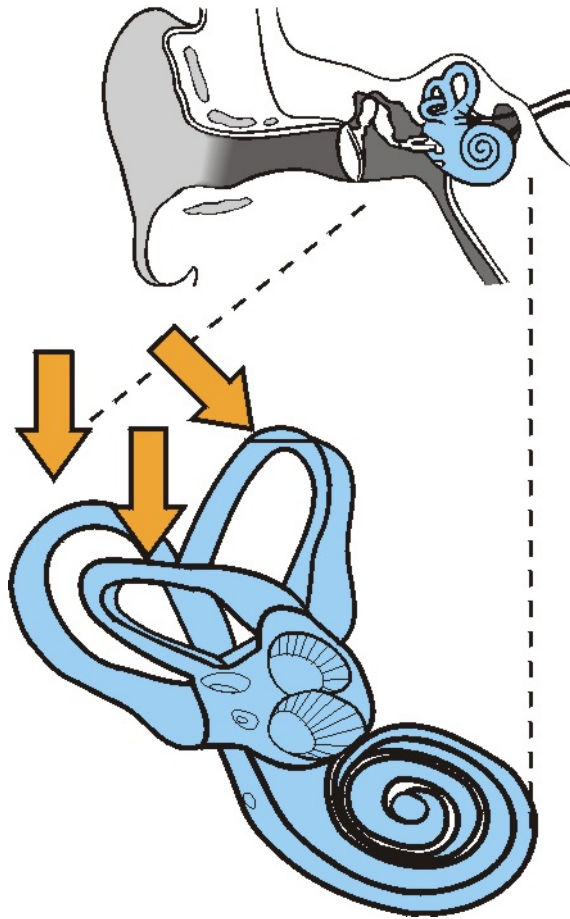


# What is our mission as ENTs for acute vertigo?

- Identify the most common disorders!!  
posterior and lateral canal BPPV 30% of all!  
vestibular neuritis 90 of acute vestibular syndrome
- Identify otitis / labyrinthitis!!
- Identify "what it's not!" (any of the above)
- Use the clinical tests that best differ between peripheral and central vertigo (and there are recent studies on this!)

# The organs of balance

(vestibular organs)



Semicircular canals= "gyros"

Head rotations in all planes

stabilises the eyes in space  
(vestibulo-ocular reflex)

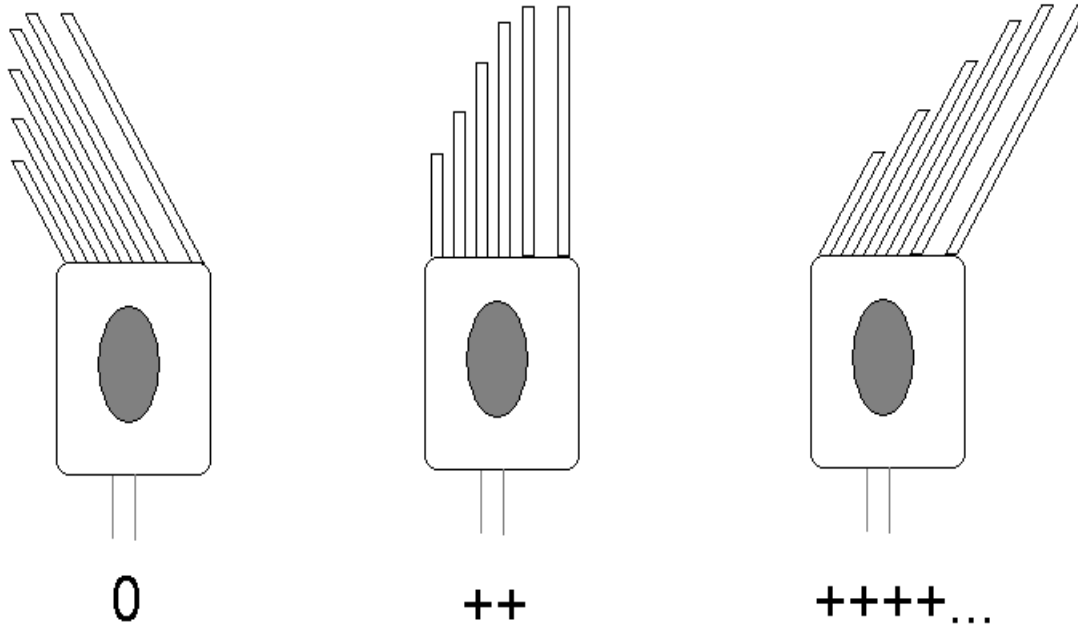
Otolithic organs = "accelerometers"

gravitation  
head position relative the ground

linear acceleration

stabilises body (and eyes) muscle  
tension in neck, trunk and legs

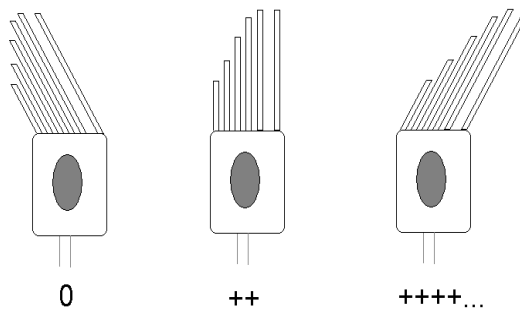
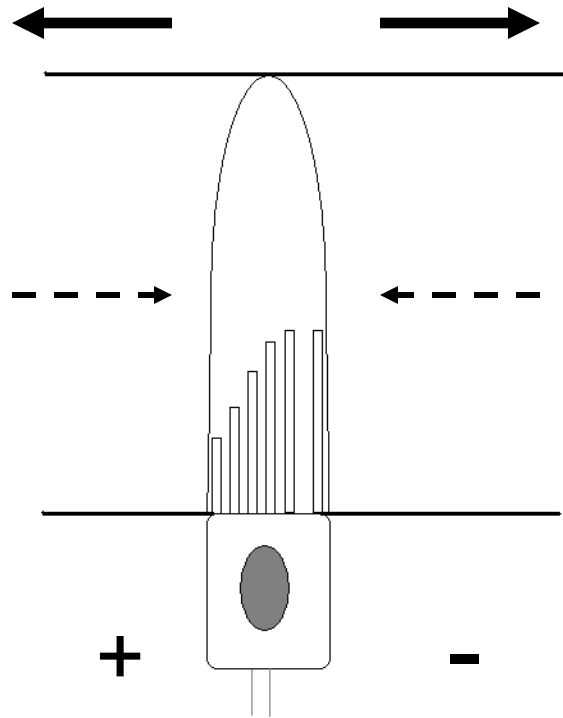
# The hair cell: common nominator



hearing  
"rotation"  
"acceleration"

Cortic organ  
semicircular canals  
otolithic organs

# Semicircular canals

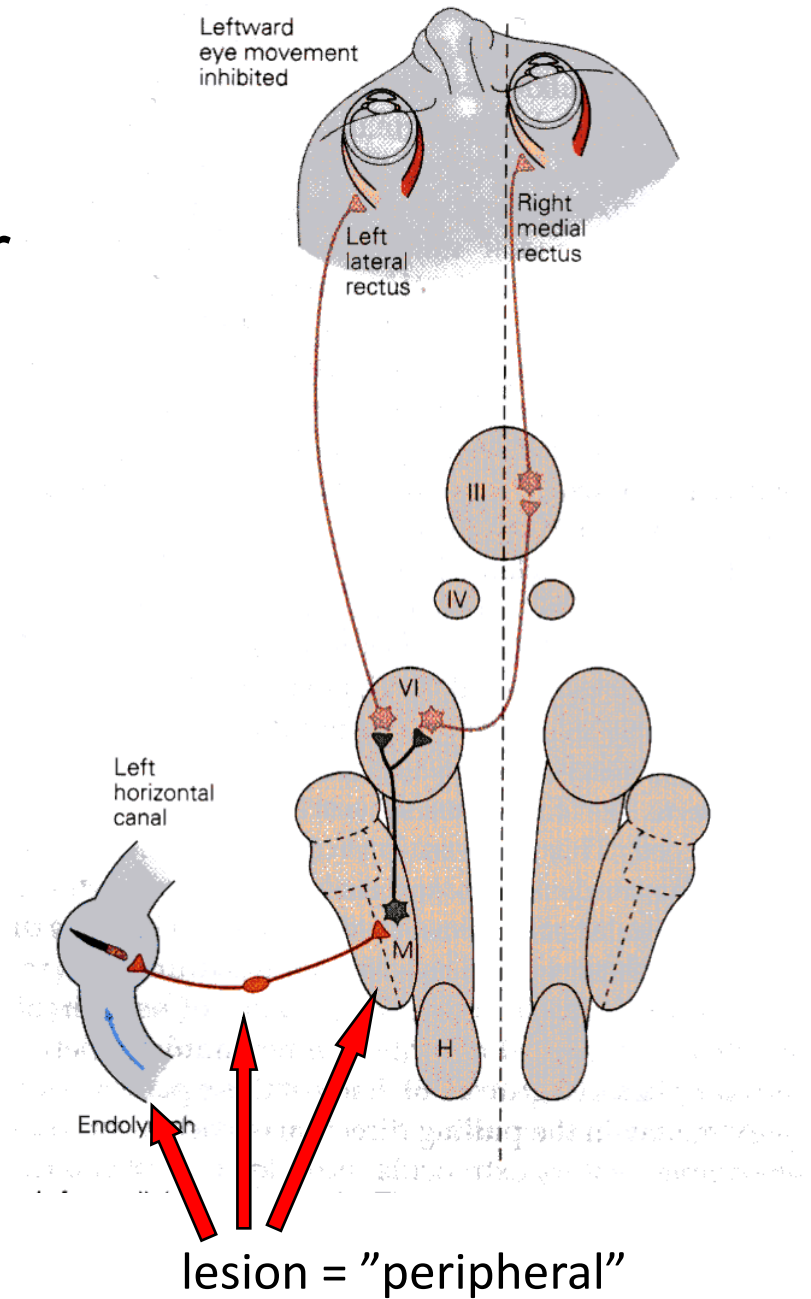


Hårcellen: minsta  
gemensamma nämnare

Semicircular canals  
connected to extra-ocular  
muscles

3-4 neurons

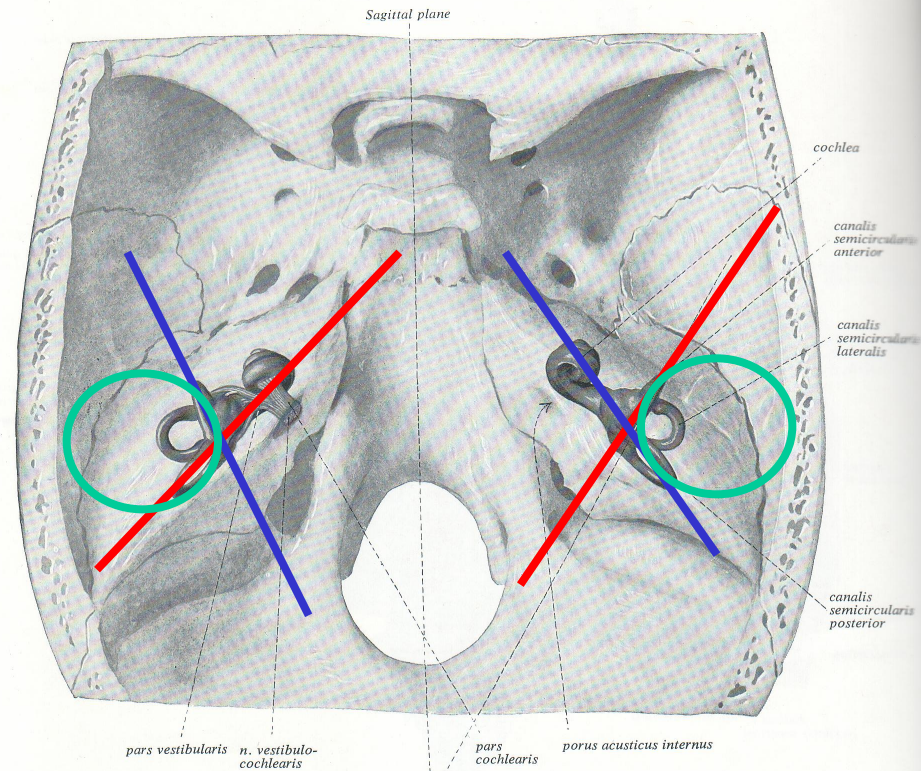
Fast reflex 9 ms latency



LARP =  
Left Anterior  
Right Posterior

RALP =  
Right Anterior  
Left Posterior

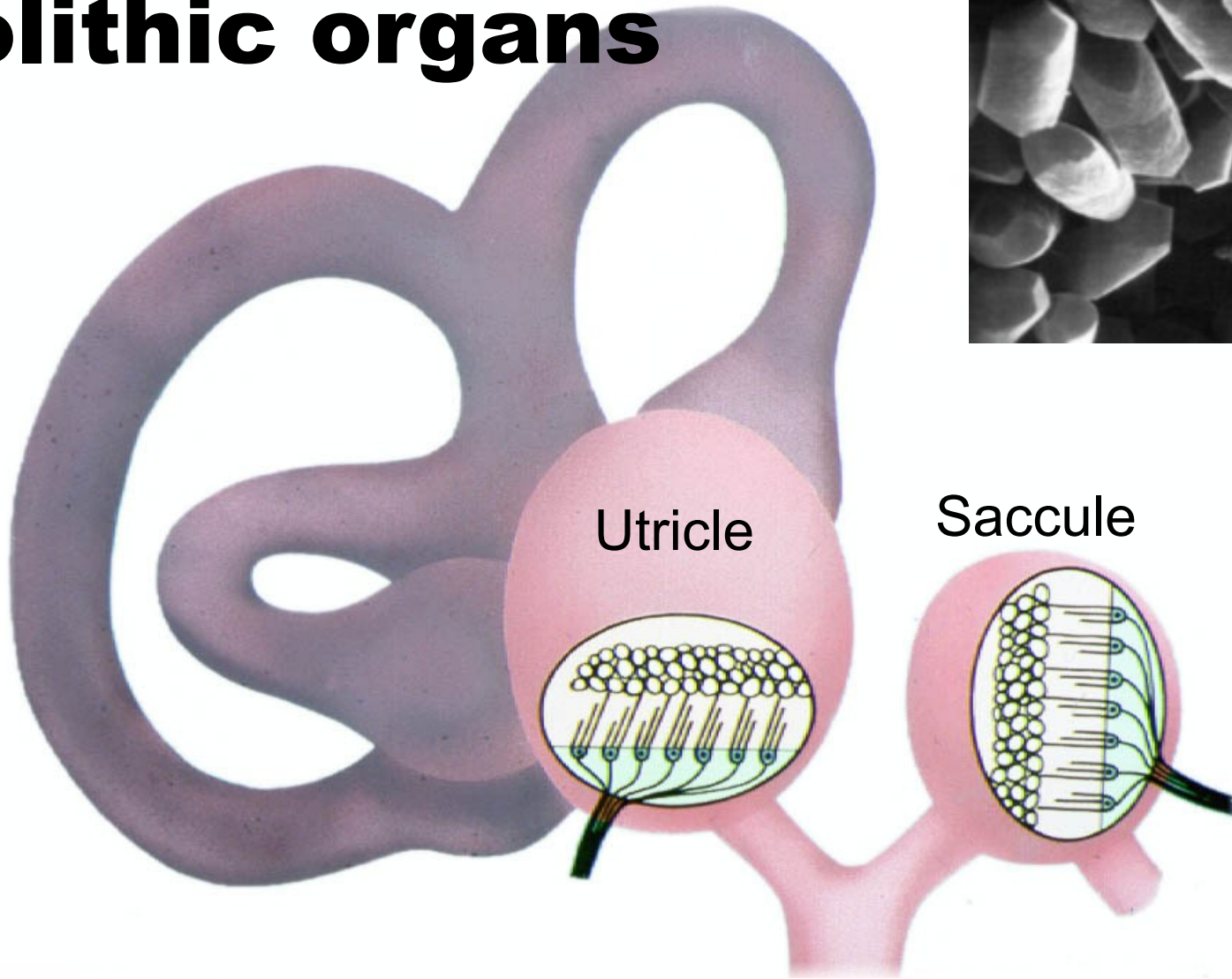
=  
Right Lateral  
Left Lateral



**A differences in signalling  
between canal pairs gives eye  
movements as if a head movement  
in that plane takes place!**

The anatomy of the canal planes  
can vary between individuals!!

# Otolithic organs



No clinical tests for otolith function!

# Neuroanatomy

after Balaban & co-workers

anxiety / conditioning

vestibulo-parabrachial nc network

coeruleo-vestibular network

Raphe nc – vestibular network

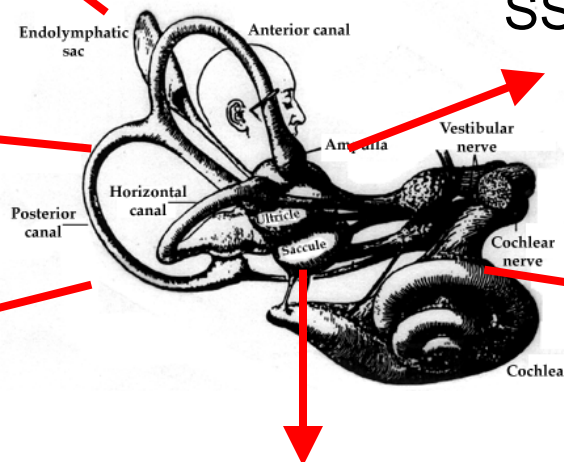
Limbic system (hippocampus)

adrenergic system  
arousal

serotonergic system?  
SSRI discontinuation syndrome

cortex:  
orientation

oculo-motor



postural muscles

autonomic reflexes:

cardio-vascular/respiratory/gastro-intestinal



# 1. Spontaneous nystagmus

Direction? Describe with words!!!

Quick phase = direction

Grade 1, 2, 3?

Grade 1 = gaze evoked nystagmus in one direction

Grade 2 = nystagmus in primary position + in one gaze direction

Grade 3 = nystagmus in all horizontal gaze directions

Vestibular spontaneous nystagmus stronger with gaze in quick phase

Visual suppression = "vestibular" nystagmus only visible with "tools"

Always use Video-Nystagmoscopy or Frenzel glasses!

## 2. Gaze-evoked nystagmus



Cerebellum

Toxic: alcohol, lithium, anti-epileptics.....

## Test also vertical gaze

**OBS! Pure vertical or torsional spontaneous nystagmus implies a CNS lesion!**



Down-beating spontaneous nystagmus = cerebellum (lower) "No emergency!"  
Up-beating spontaneous nystagmus = upper pons/mid brain "More emergent!"

# Lots of different strange & rare nystagmus!!

Congenital nystagmus: can look as anything!

Rebound nystagmus: changes direction with gaze direction = cerebellum

Ocular flutter: eyes jump with horizontal saccades

Opsoclonus: eyes jump with saccades in all directions

Ocular flutter and opsoclonus = brain stem

If you see something you don't understand, search the net (YouTube / Neuroocular)!!!

### 3. Smooth pursuit eye movements



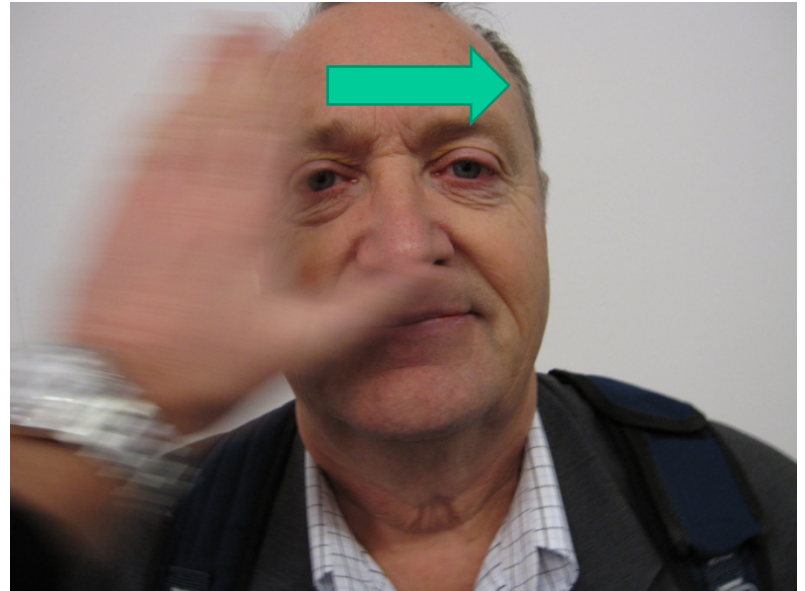
Or use patientent's own fingers as fixation point!

Saccadic smooth pursuit usually togehter with gaze-evoked nystagmus and impaired visual suppression!! = pathology in medial cerebellum! (flocculus/para-flocculus)

# 4. Test skew = alternernating cover test



5-6 s



Skew deviation = "vertical squint"

Cover one eye 5-6 seconds  
Cover the other eye  
Vertical saccade?

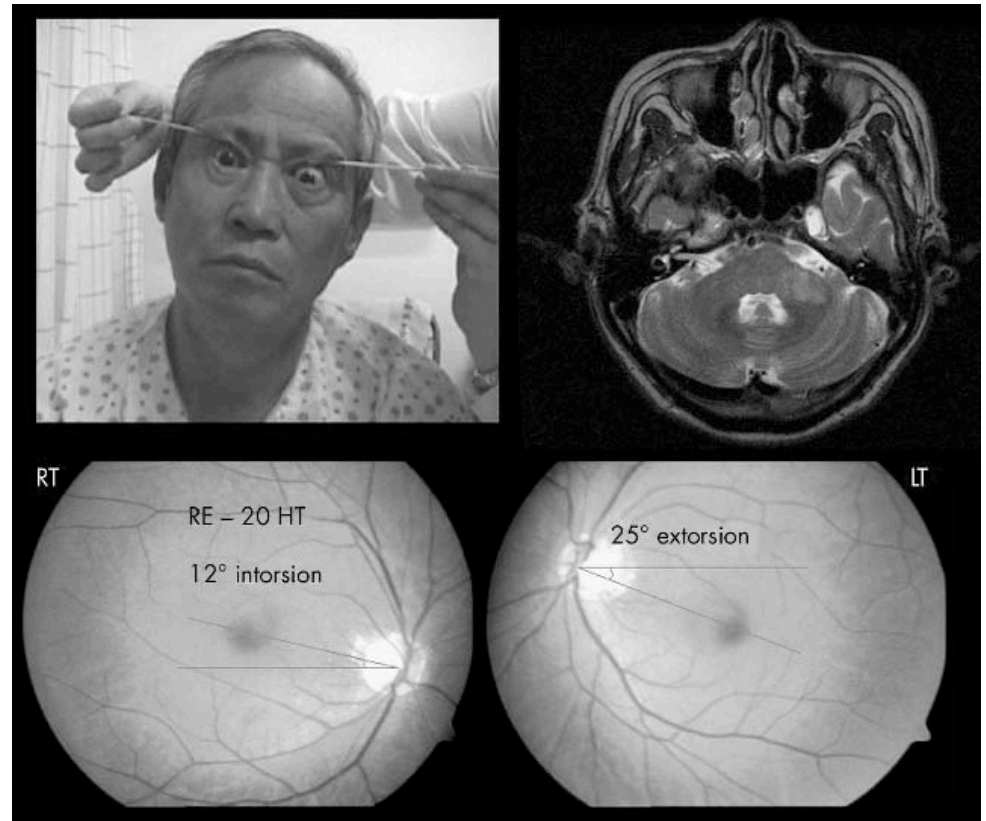


# Ocular tilt reaction

1. Ocular torsion
2. Head tilt
3. Skew deviation
4. Tilt of subjective visual vertical

tonic vestibular imbalance  
In the frontal plane

unilateral brain stem lesion  
(Wallenberg syndrome)



## 5. Visual suppression (of vestibular nystagmus)

Ask patient to fixate on own thumb  
Rotate back and forth

Obvious nystagmus = pathology!

Medial cerebellum?

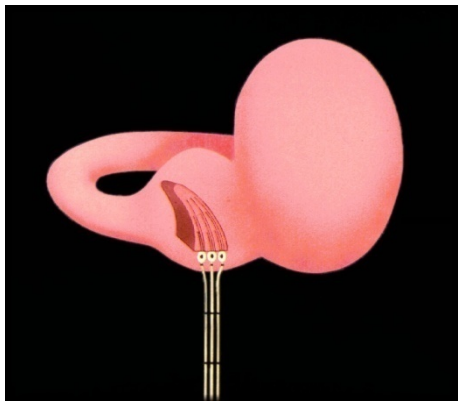




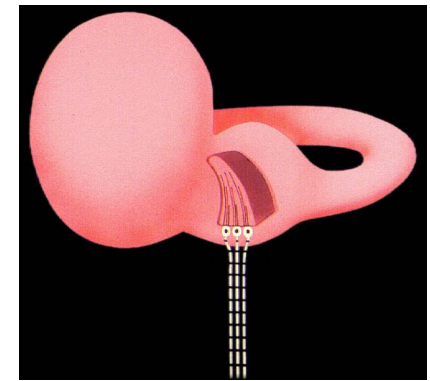
# 6. Head Impulse Test

(Halmagyi & Curthoys 1988)

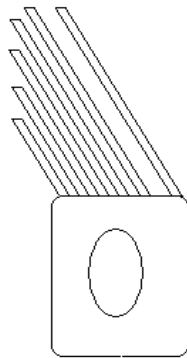
Head rotation left



Right lateral canal

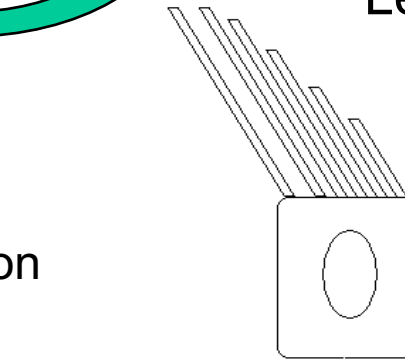


Left lateral canal



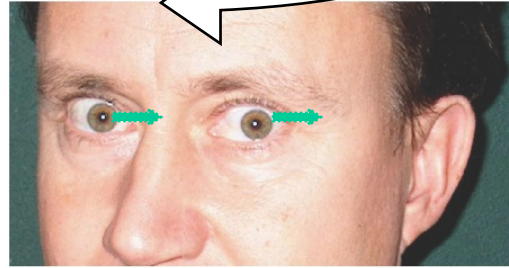
+ →

VOR  
Eye rotation  
right

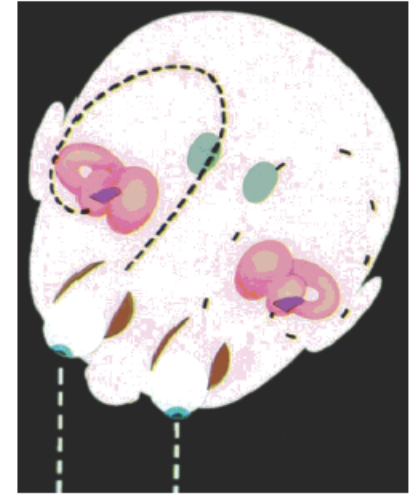


← +++++

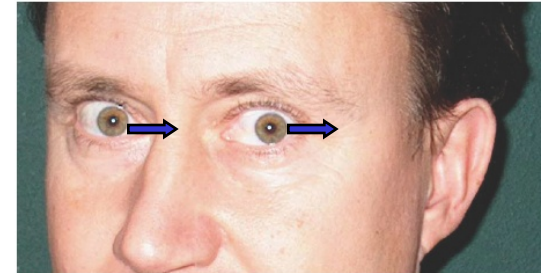
# Normal Head Impulse Test



VOR



# Pathologic Head Impulse Test



0 → ← 0

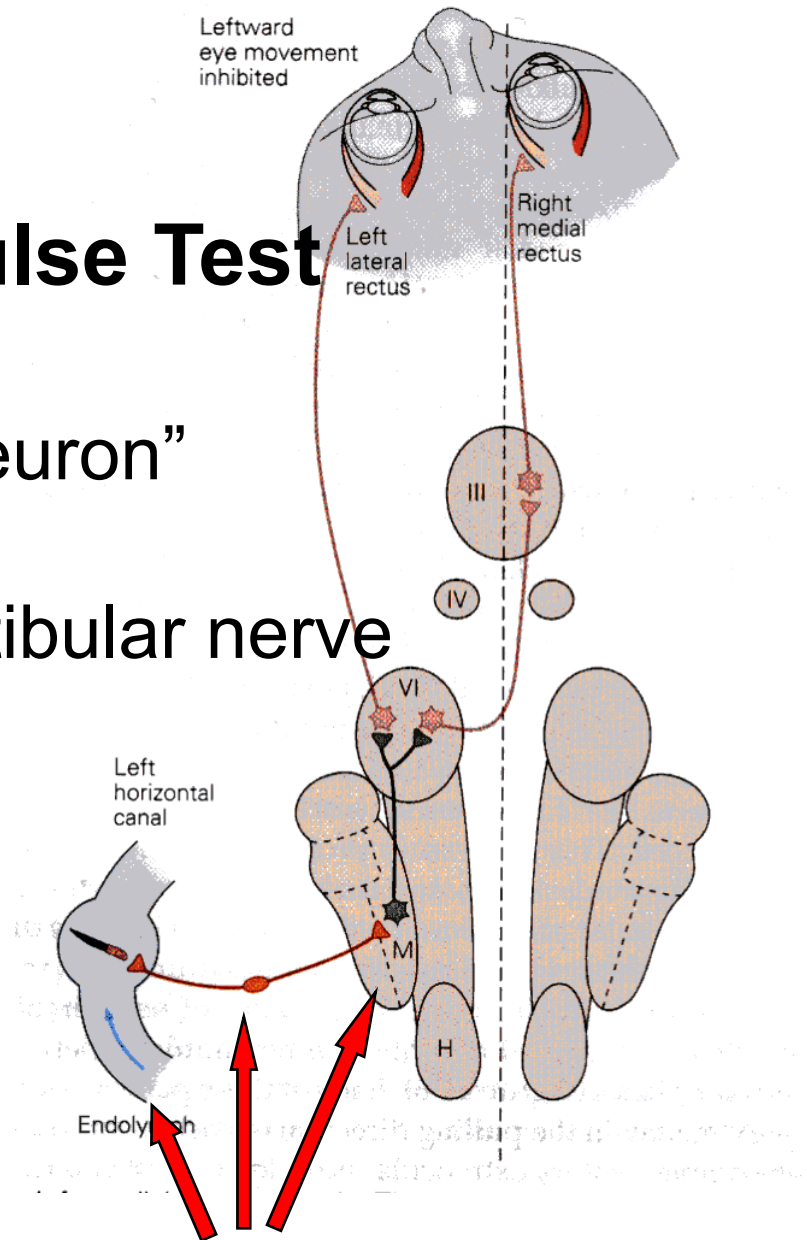
Pathologic is  
"peripheral"!

Overt saccade !

# "Positive" Head Impulse Test

Only with lesion in "1:st neuron"

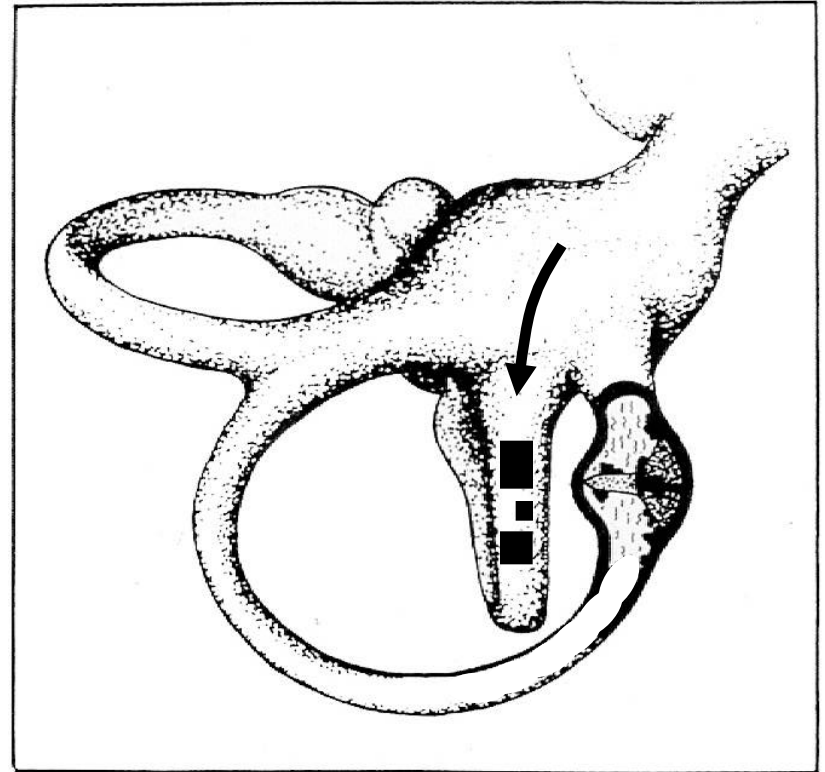
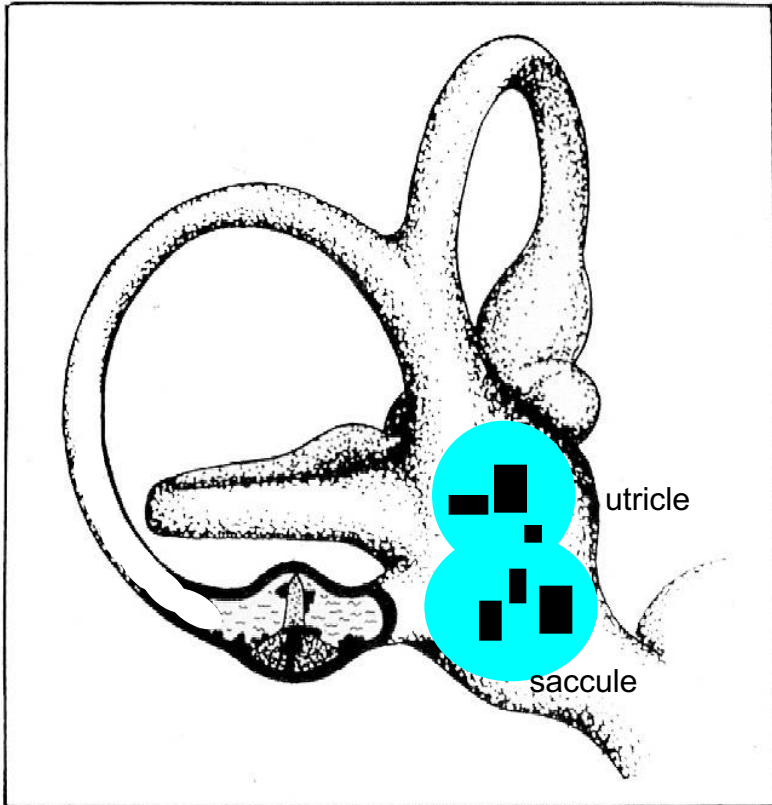
= semicircular canal / vestibular nerve  
and (Sorry!) brainstem!



Lesions give same symptoms / findings

# Video-nystagmoscopy:

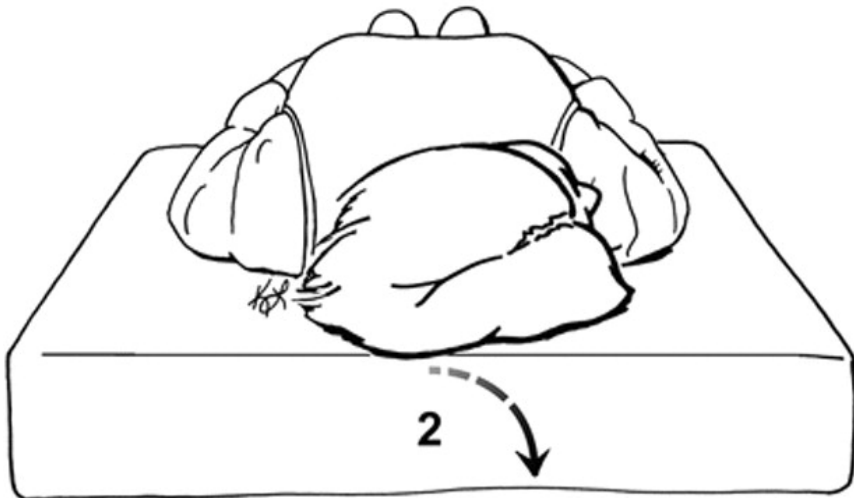
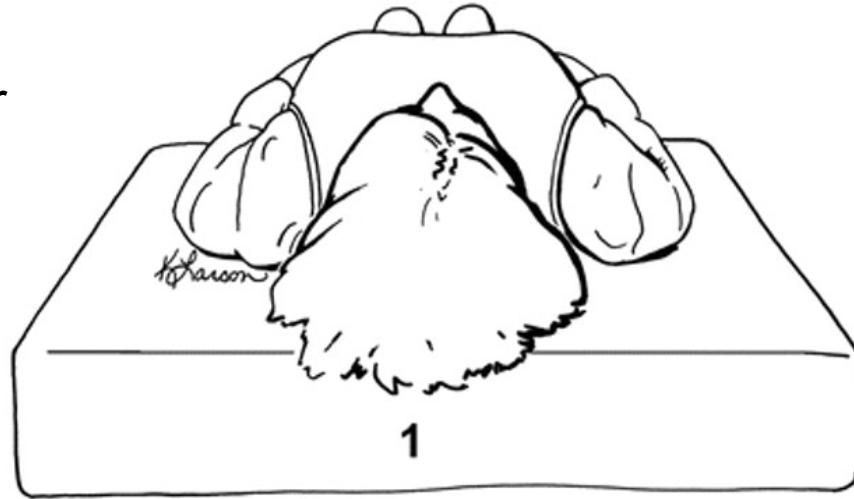
1. Spontaneous nystagmus
2. (ev.blickriktning)
3. Lean forward (lateral canal-BPPV?)
4. Lateral BPPV-test  
(pillow– head 30° nose up – left / right)
5. Dix-Hallpike test (head hanging over pillow)
6. Head-shake test (sitting 30 head rotations 2 Hz)



Right inner ear

# Lateral canal-BPPV test

"Sicker" than posterior canal BPPV!

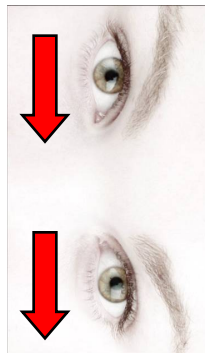
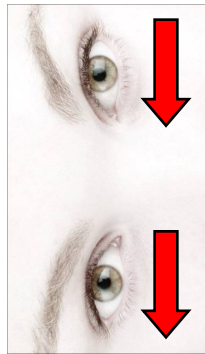
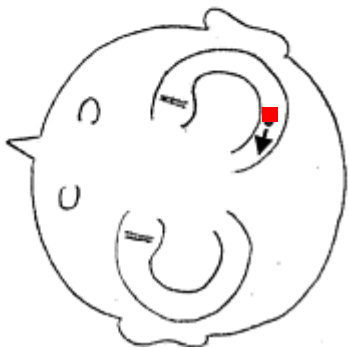
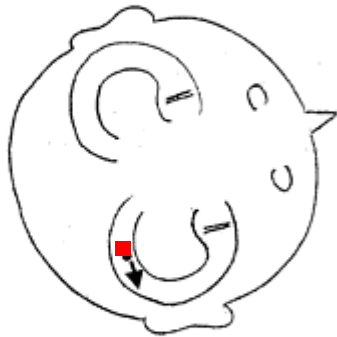


# BPPV - lateral canal

(20-40%)

## canalolithiasis

Loose otoliths in lateral canal  
intense vertigo (Ambulance! Emergency!)



nystagmus towards the floor (canal) in  
both right and left side!

Long duration but decreasing

Intense vertigo/nystagmus=sick side

### Treatment:

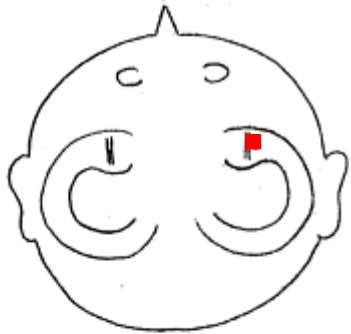
Lie on "best" side 10 minutes, then  
prone 10 minutes

Gufoni/Appiani-manoeuvre

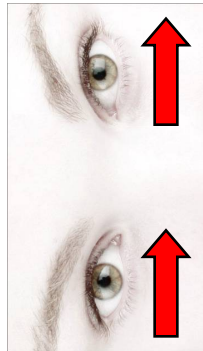
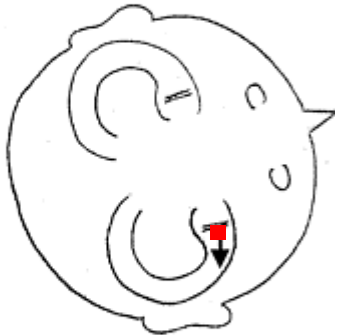
Forced prolonged positioning = lie  
on best side 24 hours

# BPPV - lateral canal

## cupulolithiasis



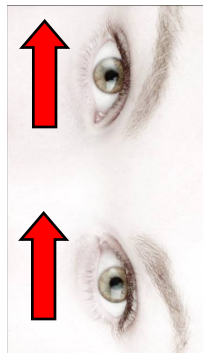
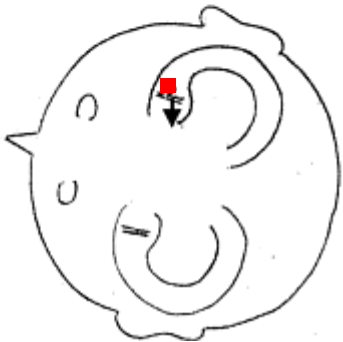
otoliths stuck on lateral canal cupula  
Less intense vertigo



nystagmus towards heaven (cupola) in  
both right and left side!

Not decreasing!!

Intense vertigo/nystagmus=healthy side



### Treatment:

Turn into canalolithiasis

Head-shaking/vibration?

Shake head several times an hour

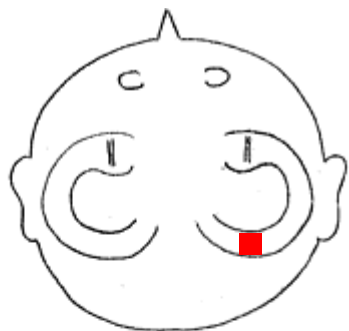
Lie on "best" side

Brandt-Daroff excersises

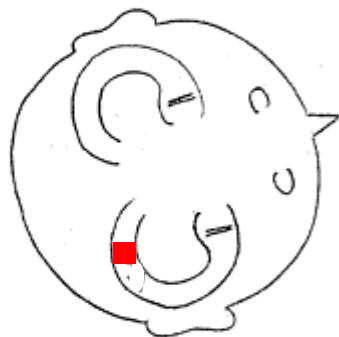


# BPPV - lateral canal

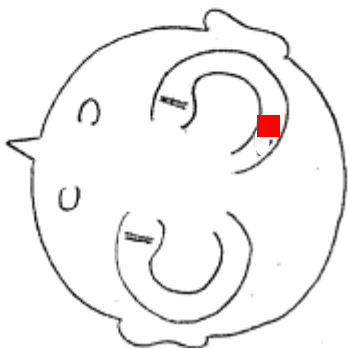
## "canalith jam"



loose otoconia in lateral canal "get stuck"  
continuous vertigo / nystagmus

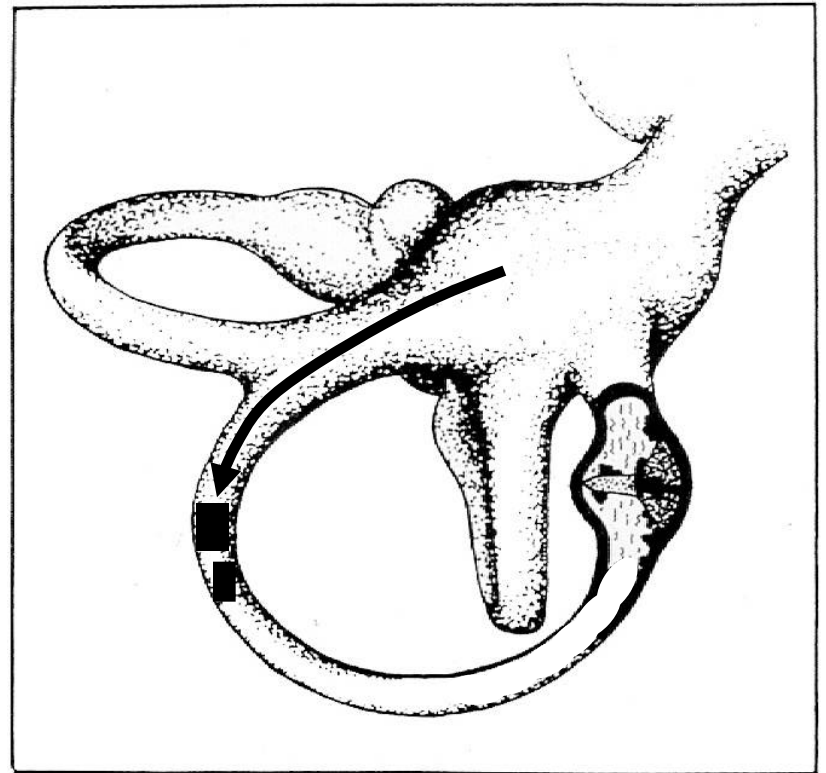
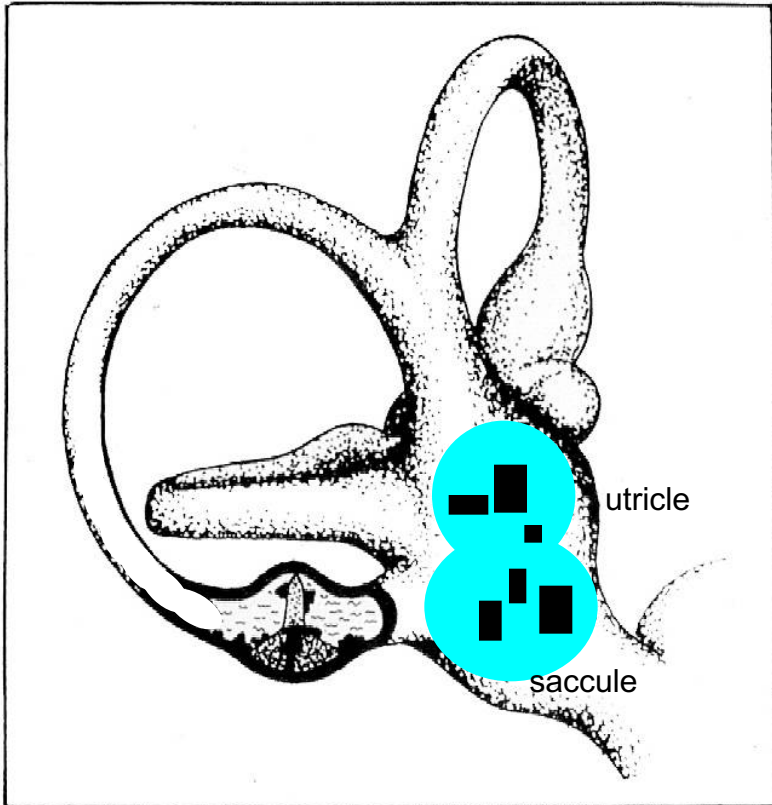
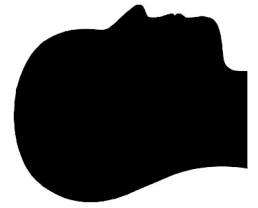


spontaneous nystagmus left / right  
not affected by position changes



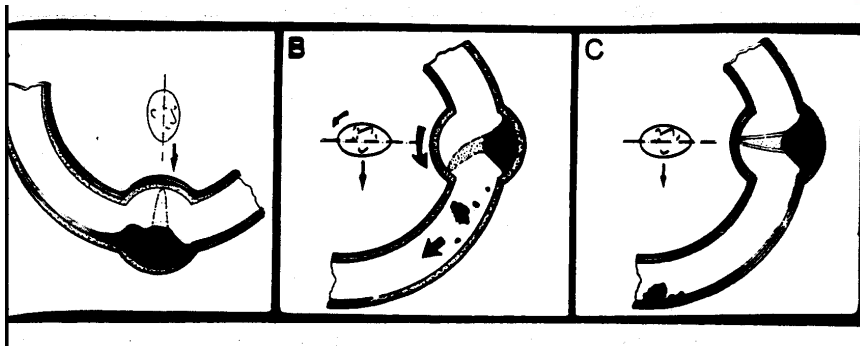
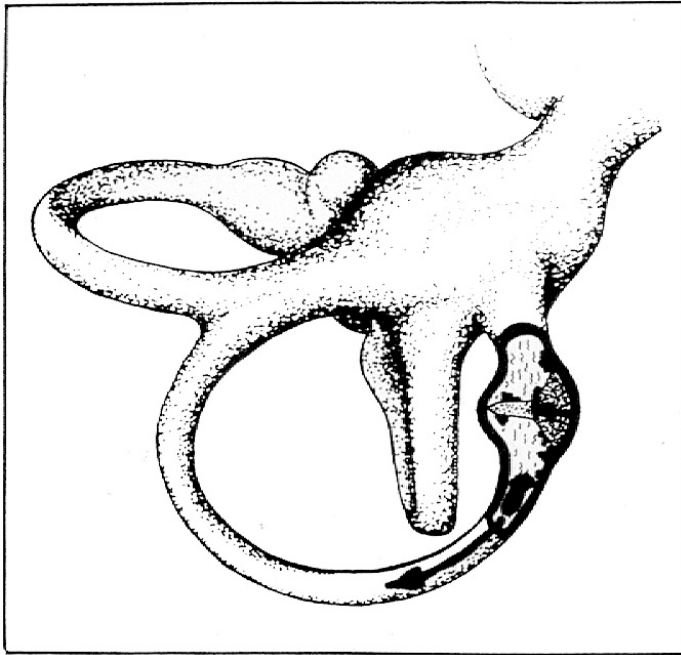
### Treatment:

Turn into canalolithiasis with "loose"  
otoconia  
vibration / head shaking  
then positional treatment

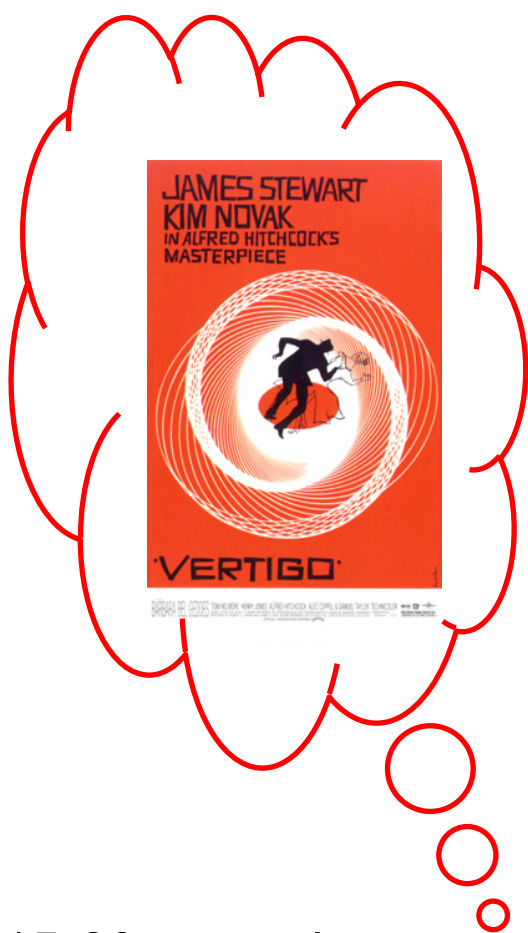


Right inner ear

# BPPV – posterior canal (60-80%)



Dix-Hallpike test



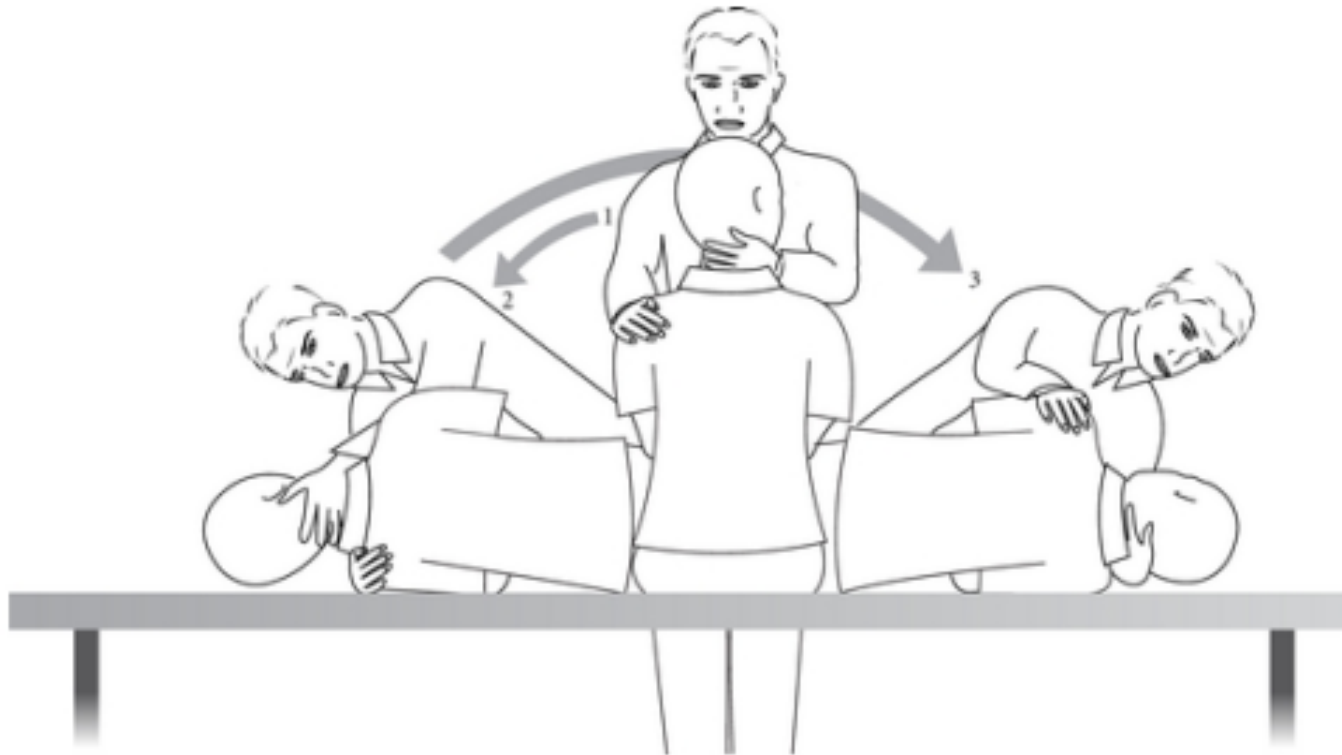
Latency

Duration 15-60 seconds

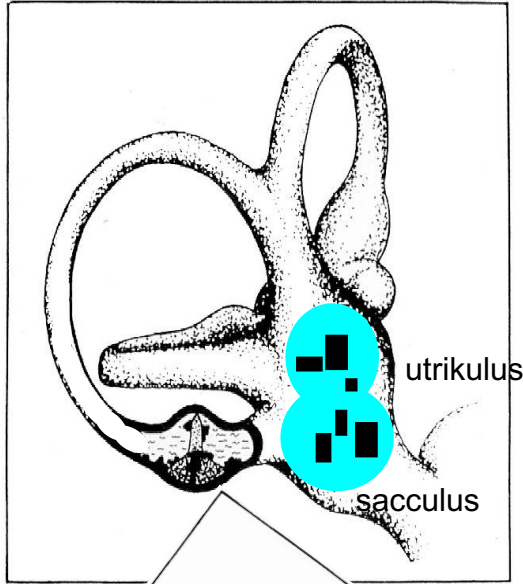
Vertical-torsional  
nystagmus



# Alternative, "kinder", way to test (older, overweight..)



# BPPV - superior canal (0-10%)



RARE!

Dix-Hallpike: torsional-vertical nystagmus  
Down-beating vertical nystagmus!!

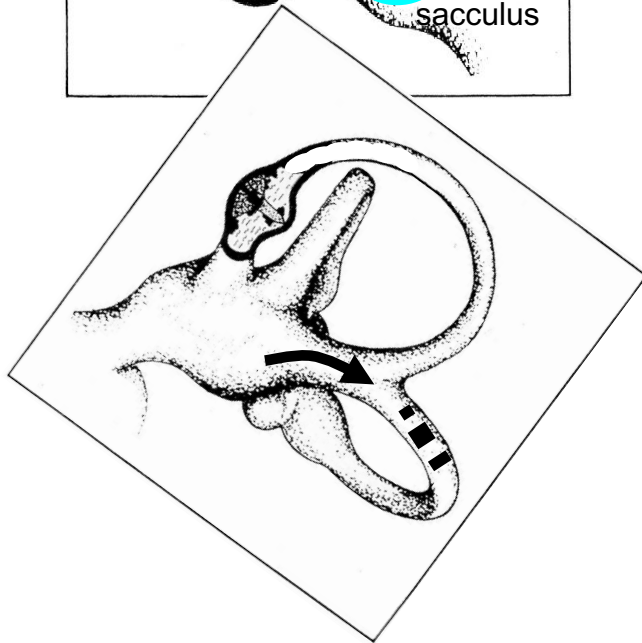
Disappears = canalolithiasis  
Continuous = cupulolithiasis

**Treatment:**

Epley manouvre

Brandt-Daroff excersises

”Mecka excersises”



# Head-shaking nystagmus

sensitive but unspecific

30 shakes                      ca 2 Hz

>3 nystagmus beats = "not normal"

Beats towards "most functioning side"

Might change direction

Can be peripheral or central

Vertical nystagmus after horizontal head-shaking  
is always central!

# Neurologic tests

1. Vibratory sensation ankle (128 Hz)
2. Romberg's test
3. "Balance walk"

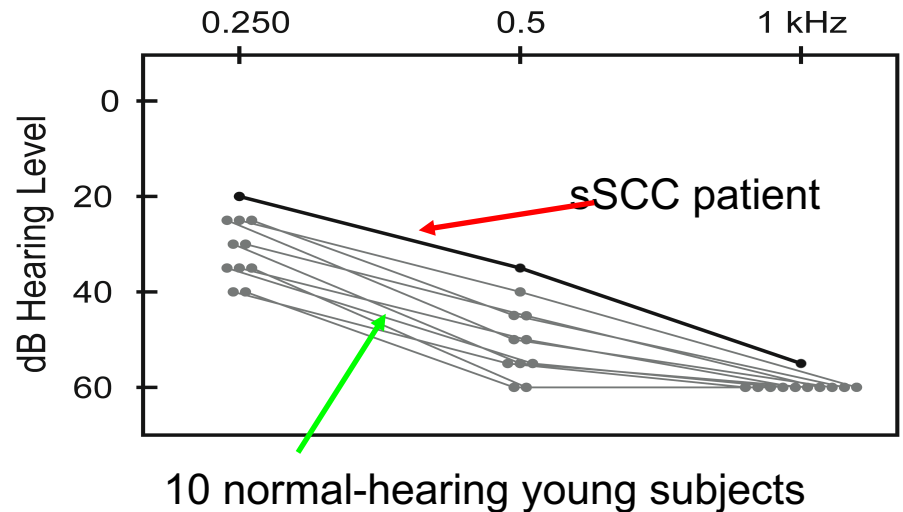
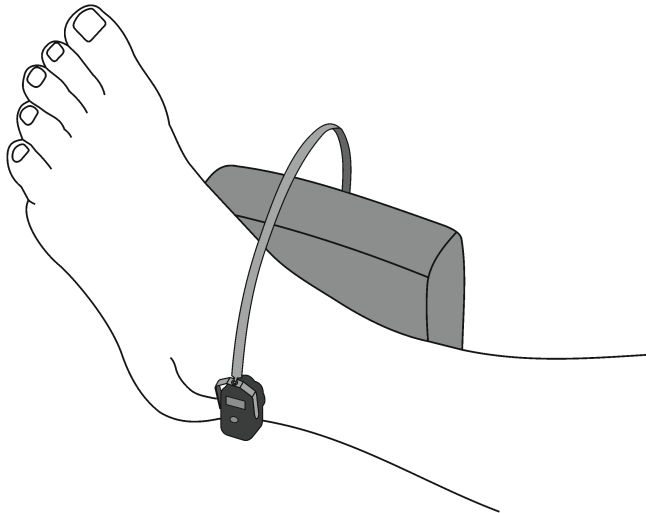


# ”Ankle-Weber”

In patients that lateralises Weber

Put tuning fork (128 Hz) to lateral malleolus

Hears it clearly in one ear = ”3rd window”



# Fistula test

Politzer balloon in ear

Over pressure/under pressure

Horizontal nystagmus =  
lateral canal fistula

Vertical/torsional nystagmus =  
vertical canal fistula ("sSCC dehiscence")

ALBERT MUDRY



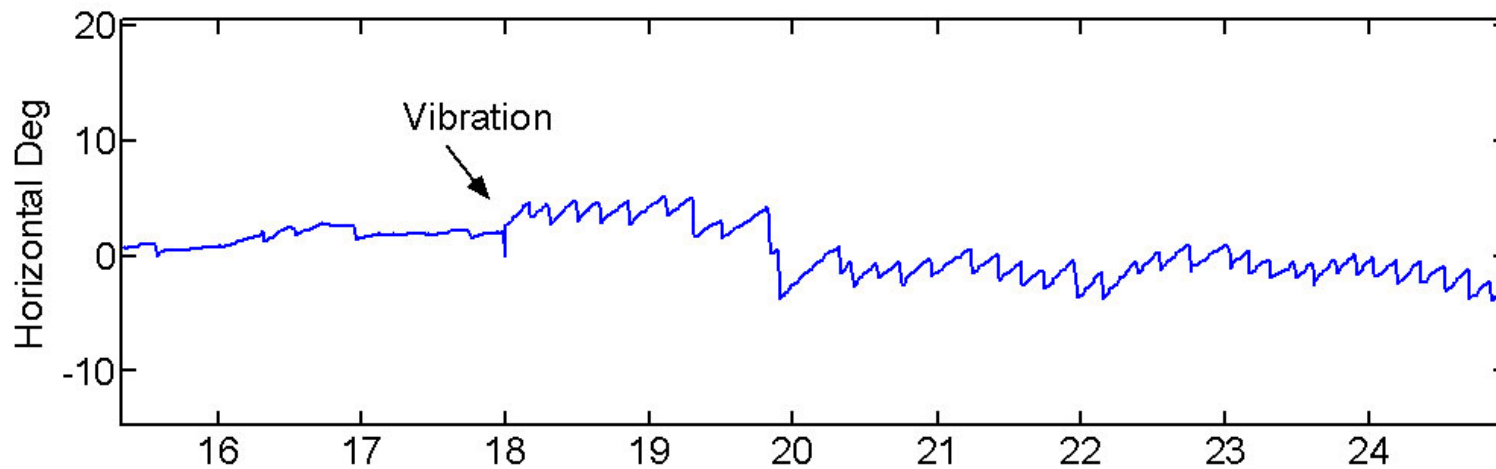
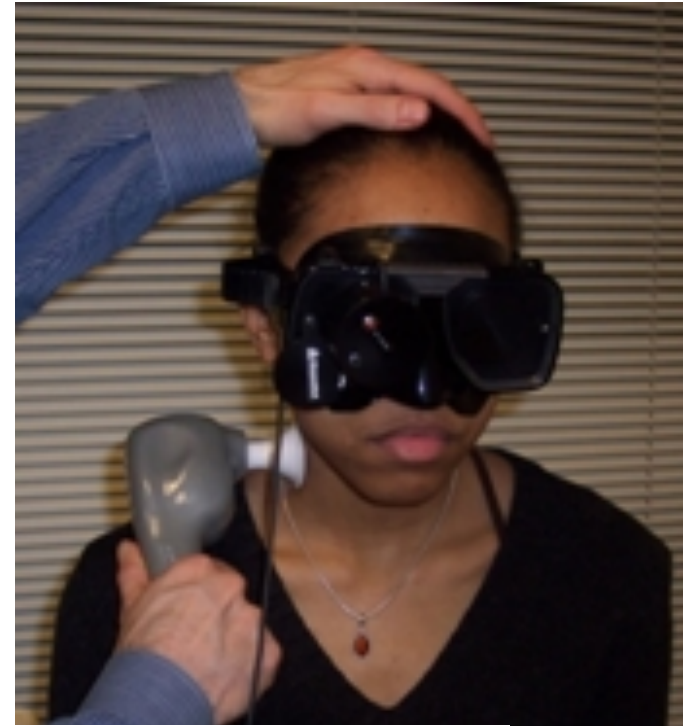
ADAM POLITZER  
*A LIFE FOR OTOLGY*

WATSONVILLE, CA  
2000

# Vibration-evoked nystagmus

Vibrator ca 100 Hz to  
mastoid / below mastoid

= head-shaking nystagmus



# Hyper-ventilation evoked nystagmus

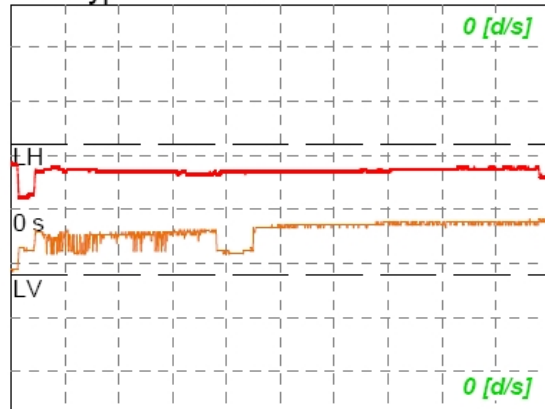
Ca 1 minute hyperventilation 1 Hz

Nystagmus?

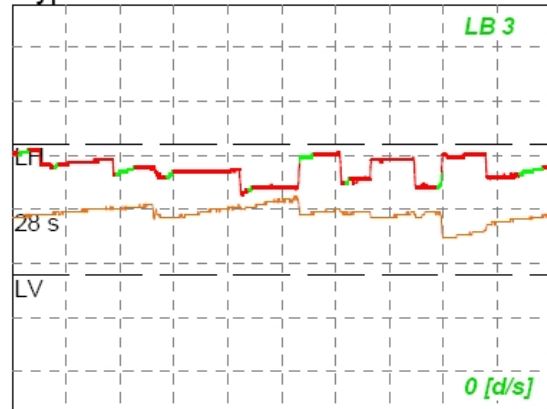
= head-shaking nystagmus

## Hyperventilation Video

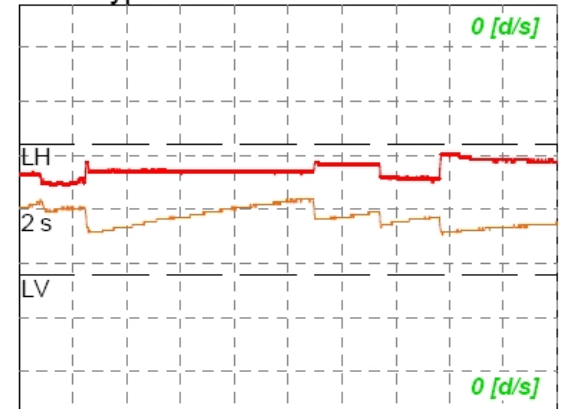
Pre-hyperventilation



Hyperventilation



Post-hyperventilation



# Steppingtest Unterberger / Fukuda

Closed eyes

Step in place 60 steps / 1 Hz

Obvious rotation = vestibulo-spinal  
"skew"

Hard to standardise

Interpretation?



# **”Acute vestibular syndrome”**

## vestibular neuritis or stroke?

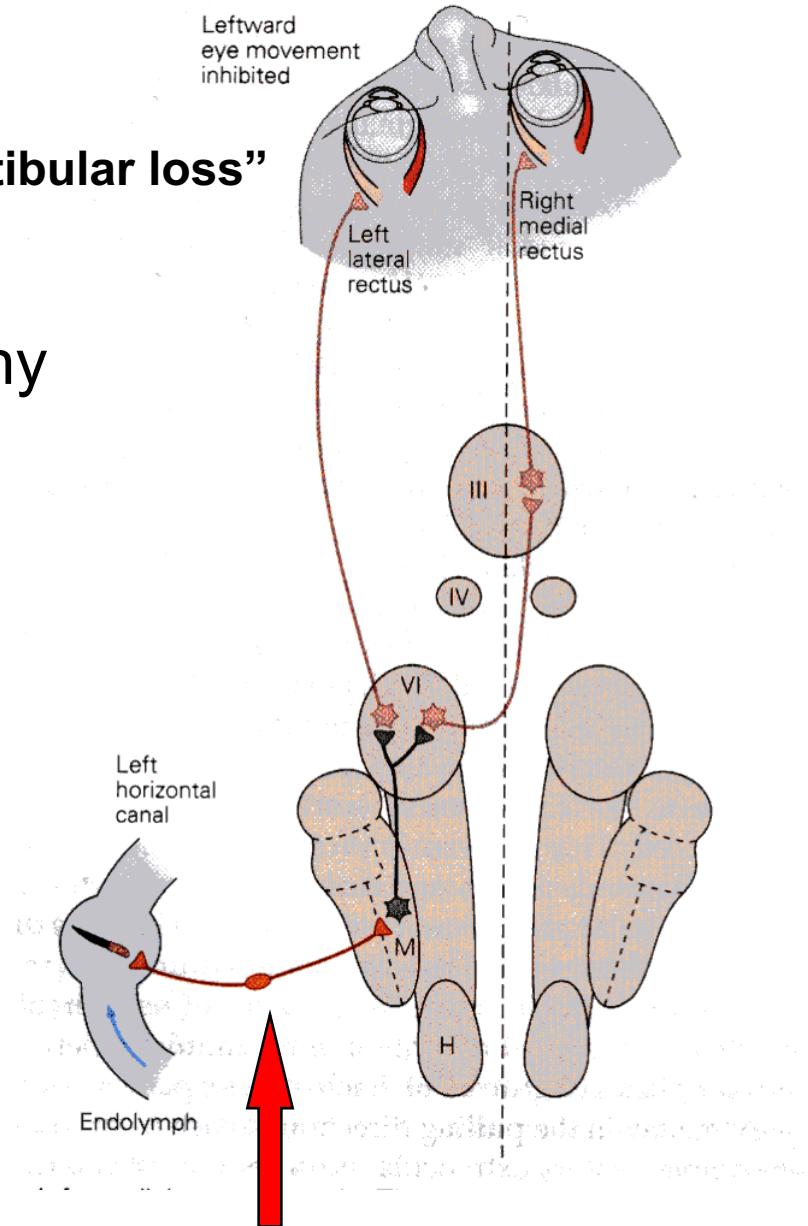
- Sudden onset of continuous vertigo (>24 h)
  - Nausea and/or vomiting (usually)
  - Spontaneous nystagmus
  - Worsening by head movements
  - Unsteadiness / ”lateropulsion”
- 
- No other obvious neurological symptoms
  - No hearing loss

# Acute vestibular neuritis

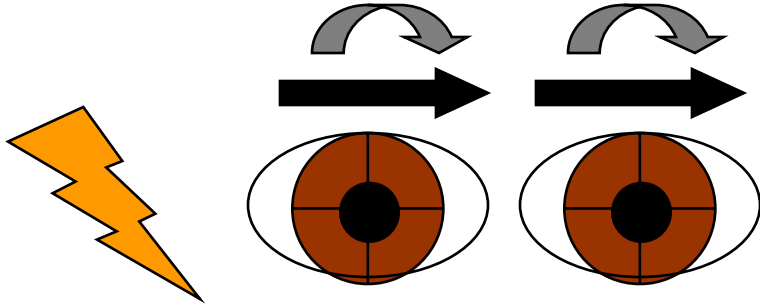
”sudden peripheral idiopathic unilateral vestibular loss”

probably an inflammatory neuropathy  
(as Bell’s palsy?)

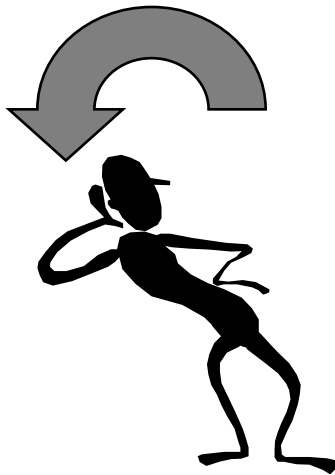
90 % of acute vestibular syndrome



## 'Tonic' findings



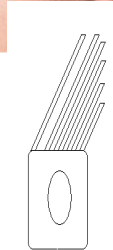
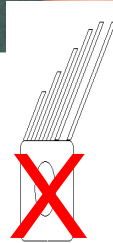
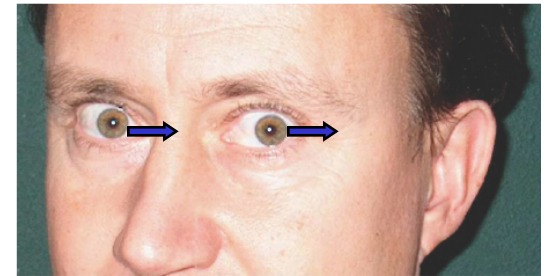
horizontal-torsional nystagmus  
fast phase towards intact side



Tendency to fall - 'lateropulsion'  
towards lesioned side



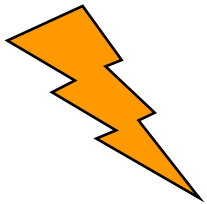
**”Dynamic” finding**



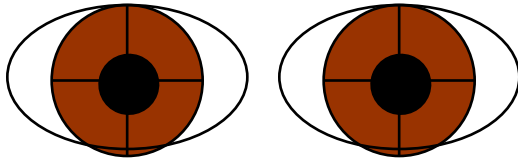
0 → ← 0

**Saccade !**

Pathologic head-impulse test to lesioned side



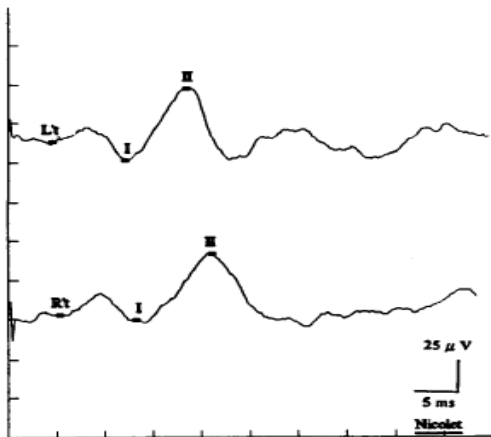
44°  
H<sub>2</sub>O



**Caloric test**  
canal paresis



**Subjective Visual Horizontal**  
tilts towards lesioned side  
5-15°



**VEMPs**  
usually intact (2/3)

*Fig. 2.* VEMP results in a 38-year-old male with vestibular neuritis on the left side. VEMP responses are positive on both sides (I: p13; II: n23).

# Why is the inferior vestibular nerve spared?



Temporal bone studies

inferior vestibular nerve : double innervation

2 nerves in separate channels      ***Arbusow et al -99***

superior vestibular nerve

narrower nerve channel

longer channel

narrower arterioli

***Goebel & Gianoli -00***

Inflammatory entrapment of the superior vestibular nerve???

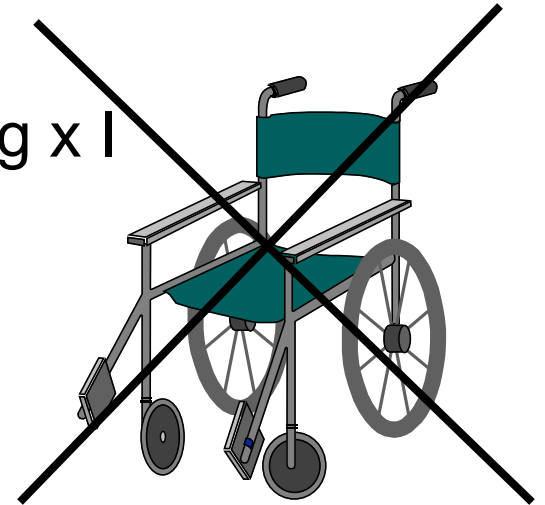


Since 2004 we have offered steroids to patients with acute vestibular neuritis

Betapred (betamethasone) 8 mg i.v.  
followed by:

T. Prednisolone 60mg x 1 x V  
50mg x I, 40mg x I, 30mg x I, 20mg x I, 10mg x I

Caloric test after 12 months





# Vestibular rehabilitation

Performing movements that exacerbate dizziness and vertigo lead to CNS adaptation

”The dizzier you are,  
the quicker you get well!”



# Stroke (“pseudoneuritis”) in acute vestibular syndrome (AVS)

## Epidemiology

- In USA 250,000 - 500,000 patients with AVS at emergency departments yearly (Newman-Toker)
- Most have vestibular neuritis (VN), but some have an ischemic stroke i brain stem or cerebellum ??“~25% ±15%”??
- In Umeå 2012-13: 11% of AVS = stroke

# Clinical examination

## Acute vertigo

Acute history

Emergency

Acute exam  
'vital signs'

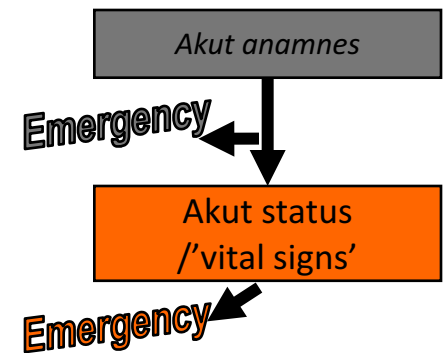
Emergency

### Acute history:

- When?
- (How does it feel?)
- Pain?? Head? Chest? Ear? Neck?
- **Hearing?** Changed?/ Hearing loss?  
Side difference?
- **Other?** Vision? Nausea?
- **Loss of consciousness?**
- Other neurological symptom?  
Double vision, swallowing,  
dysphonia, sensibility/ motor  
OBS! Also transient symptoms?!

# Acute exam

- **Level of consciousness**
- **BP/pulse/Heart/Lungs** Arrhythmia?, chock?
- **Neck stiffness??** Meningitis, SAH
- **Motor-/sensory: extremities/face/ finger-nose**  
CNS-lesion, brain stem/cerebellum
- **Eye movements – nystagmus?** spontaneous  
nystagmus =  
peripheral vestibular lesion or CNS lesion
- **HINTS: Head Impulse test – Nystagmus – Test Skew**
- **Ear** otitis - (cholesteatoma) –labyrinthitis – zoster oticus
- **Positional vertigo tests:** Dix-Hallpike, test of lateral sSCC BPPV





# **HINTS** – discriminates central from peripheral lesions in acute vestibular syndrome

**H** – **H**ead

**I** – **I**mpulse test

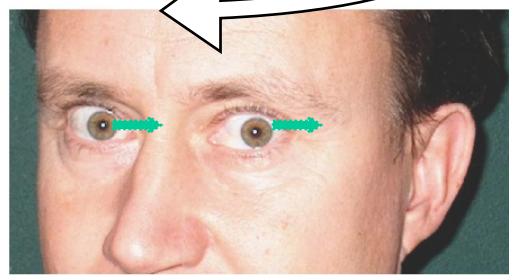
**N** – (gaze) **N**ystagmus

**T** – **T**est

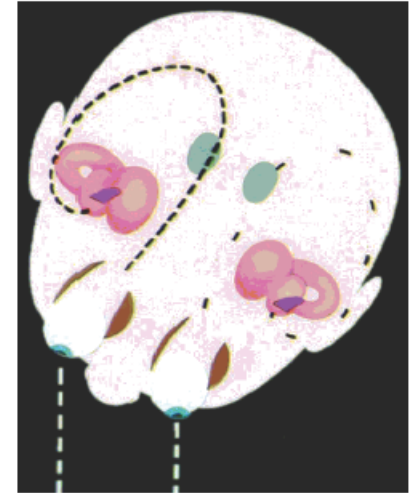
**S** – **S**kew

Better than MRI-DWI to find or rule out posterior fossa stroke in AVS!

# Normal Head Impulse test



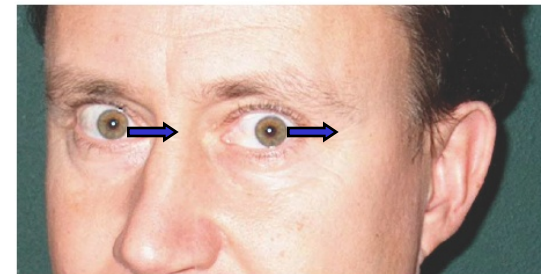
VOR



# Pathologic Head Impulse test



0 → ← 0



Saccade !

Film!

“Pathologic is peripheral!”

# Head impulse test differs PICA-infarction from "vestibular neuritis"

	n[subjects]		Sensitivity (95% CI*)	Specificity (95% CI*)	NLR (95% CI*)	PLR (95% CI*)
	P-AVS	C-AVS				
<i>Bedside diagnostic tests†</i>						
<b>Normal h-HIT</b> (all C-AVS)	65	152‡	0.85 (0.79-0.91)	0.95 (0.90-1.00)	0.16 (0.11-0.23)	18.39 (6.08-55.64)
PICA-stroke vs. P-AVS only	25	72	0.99 (0.96-1.00)	1.00§	0.01 (0.00-0.10)	NC§
AICA-stroke vs. P-AVS only	25	13	0.62 (0.35-0.88)	1.00§	0.40 (0.20-0.80)	NC§

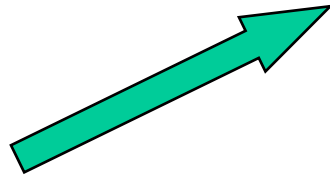
**PICA-infarction: Acute Vestibular Syndrome with normal head impulse test THE MOST IMPORTANT of HINTS!!**

# N = Nystagmus

spontaneous nystagmus  
= either CNS or "ear"

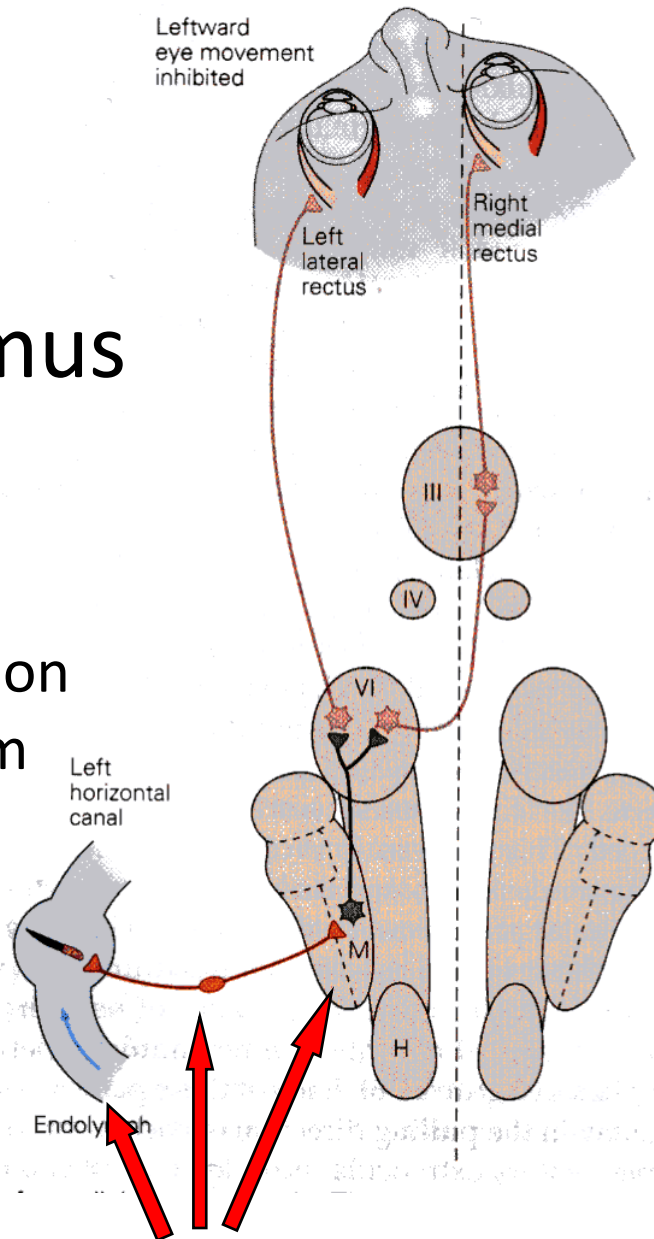
"ear" = unilateral vestibular lesion  
CNS = cerebellum OR brain stem

Horizontal-(torsional) nystagmus  
fast phase towards intact side  
Only "benign" nystagmus!!!

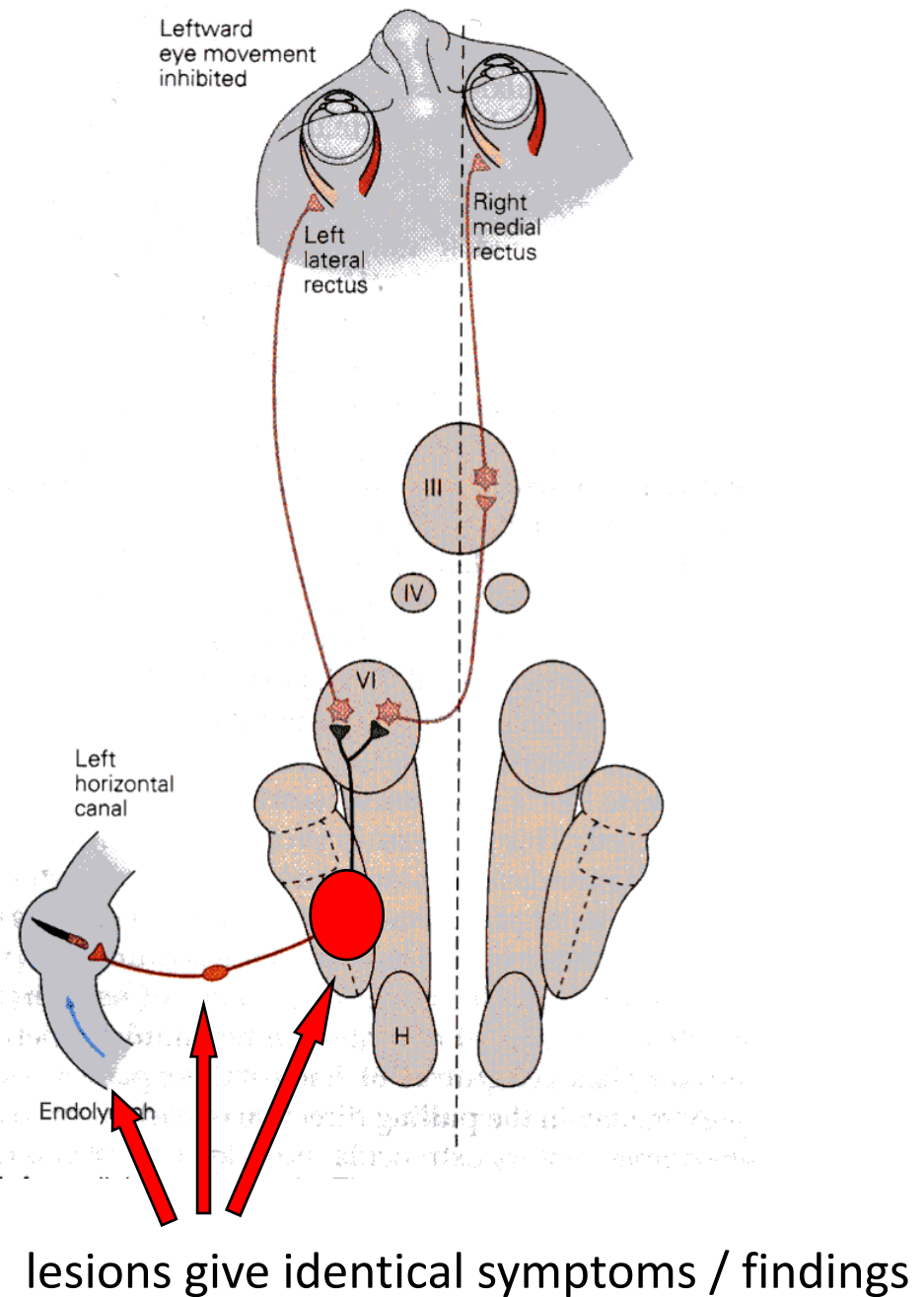


peripheral or central?

lesions give identical symptoms / findings



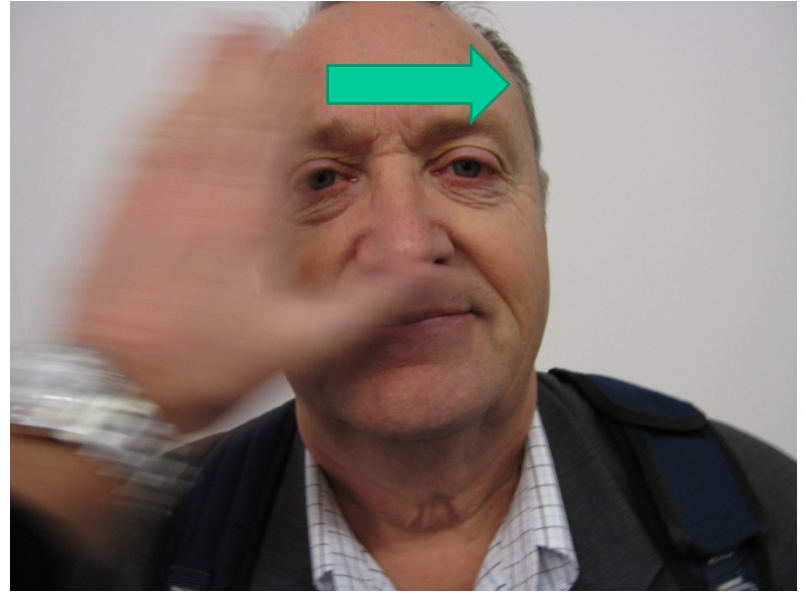
Vertigo due to  
lesion in the ear  
or in the brain  
stem?  
peripheral or central?



# Test Skew Alternating cover test



5-6 s



Skew deviation = "vertical squint"

Cover one eye 5-6 seconds  
Cover the other eye  
Vertical saccade?



# HINTS

## 3 tests in HINTS

- Head Impulse
- Nystagmus
- Test of Skew = Cover test

## **Dangerous H.I.N.T.S.** (in patient with acute vestibular syndrome)

- **Normal head impulse test**
- **Gaze nystagmus / purely vertical or torsional spontaneous nystagmus**
- Vertical refixation saccade in cover test

**Any of these in patient with AVS -  
suspect posterior fossa stroke!**

- Acute vestibular syndrome is caused either by vestibular neuritis (90%) or by stroke (10%)
- High dose steroids + vestibular rehab improves outcome after vestibular neuritis
- **HINTS** – differ between stroke and VN
- **Pain + vertigo** is "dangerous"!
- **Acute unilat hearing loss + vertigo?**  
Think stroke!!!!
- **Other neurological symptoms?** Think stroke!
- **Impossible to stand up?** Think stroke!
- **Stop ordering acute CT brain for vertigo!**