management of vestibular disorders

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problems in management of dizziness

- complex history
  which complaints relate to vestibular deficits?

- vestibular tests
  low sensitivity - low specificity: new tests?

- vestibular diseases
  pathophysiological mechanisms: new insights?

- therapy
  causal versus symptomatic
  medication: indication area? mode of action?
  ablation
  labyrinthine substitution systems - vestibular implants
vestibular pathology (traditional view)

- initial symptoms: vertigo, nausea and nystagmus
- central compensation
- recovery in a few weeks to months
- unilateral: the other labyrinth takes over
- sensory substitution
acute but **transient** symptoms

acute **unilateral** loss or fluctuating function (neuritis, Ménière…)

- acute severe vertigo, severe nausea, falling and imbalance
  (the classical leading symptoms for diagnosis)
Topical prilocaine (EMLA) on eardrums

Intratympanal injection of lidocaine (spinal tap needle)
1. Right side
2. Left side
3. Wait and see
one hour later
returning from the toilet after severe vomiting
Duration 8 hours
acute bilateral vestibular loss

no vertigo

no spontaneous nystagmus

no auditory symptoms

severe unsteadiness / ataxia

hypersensitivity to voluntary head movements

clear neuro-vegetative symptoms
acute but **transient** symptoms

acute **unilateral** loss or fluctuating function (neuritis, Ménière…)

- acute severe vertigo, severe nausea, falling and imbalance
  (the classical leading symptoms for diagnosis)

acute **bilateral** loss

- acute severe intolerance to head movements, nausea and imbalance (no vertigo: so the diagnosis is often missed)
which complaints are related to vestibular deficits?

which complaints are related to natural limitations?
which complaints are related to vestibular deficits?

which complaints are related to natural limitations?
acute but transient symptoms

acute unilateral loss or fluctuating function (neuritis, Ménière…)

- acute severe vertigo, severe nausea, falling and imbalance (the classical leading symptoms for diagnosis)

acute bilateral loss

- acute severe intolerance to head movements, nausea and imbalance (no vertigo: so the diagnosis is often missed)
poor dynamic compensation: sustained

- impact on various autonomic functions
- reduced automatisation of balance
- reduced dynamic visual acuity
- reduced perception of self motion
- hypersensitivity for optokinetic stimuli
- reduced ability to discriminate between self-motion and environmental motion
- secondary: fear and fatigue (cognitive load)
**CNS**

**interpretation**

**learning**

**adaptation**

**compensation**

**somatosensory**

e.g. foot sole pressure

**vision**

**hearing**

**labyrinths**

**gravitoreceptors**

blood pressure sensors in large blood vessels

**autonomic processes**

fast blood pressure regulation

heart beat frequency

nausea / vomiting

**image stabilisation**

**balance control**

**spatial orientation**

**e.g. foot sole pressure**
permanent complaints / symptoms in patients with centrally compensated vestibular function loss

- increased neuro-vegetative sensitivity
**CNS**

- **Interpretation**
- **Learning**
- **Adaptation**
- **Compensation**

**Sensory Inputs:**
- **Gravitoreceptors**
  - Blood pressure sensors in large blood vessels
- **Labyrinths**
- **Vision**
- **Hearing**
- **Somatosensory**
  - E.g. foot sole pressure

**Autonomic Processes:**
- Fast blood pressure regulation
- Heartbeat frequency
- Nausea / vomiting

**Outputs:**
- Image stabilisation
- Spatial orientation
- Balance control

**Image Stabilisation**

**Spatial Orientation**
permanent complaints / symptoms in patients with centrally compensated vestibular function loss

- increased neuro-vegetative sensitivity
- reduced automatisation of spatial orientation
- hypersensitivity for optokinetic stimuli
vestibular impact upon postural control

- regulation of muscle tone relative to gravity

- regulation of COM relative to base of support balancing correction steps

- labyrinths important for balance at low speed learning motor activities → automatisation
• Frontal cortex: initiation, dual tasks
• Cerebellum: rhythm and velocity
• Basal: ganglia modulation
• Brainstem: start and stop
• Spinal cord: automatic spinal patterns (running)
• Labyrinth: fast detection and correction of imbalance (VSR)
otolith function especially relevant for:

- motor learning (retardation in congenital areflexia)
- maintaining complex postures
- standing or slow walking
  - on a soft surface (wind-surfing)
  - in darkness
  - in presence of misleading visual stimuli

labyrinths less relevant for:

- walking at normal speed or running (visual anticipation)

bilateral areflexia leads to degeneration of
“head direction” and head “place” cells in the hippocampus
patient with severe bilateral vestibular hyporeflexia

slow tandem walk

fast tandem walk
permanent complaints / symptoms in patients with centrally compensated vestibular function loss

- increased neuro-vegetative sensitivity
- reduced automatisation of orientation
- hypersensitivity for optokinetic stimuli
- reduced automatisation of balance
- **image stabilisation**
- **balance control**
- **labyrinths**
- **CNS**
  - interpretation
  - learning
  - adaptation
  - compensation
- **somatosensory**
  - e.g. foot sole pressure
- **vision**
- **hearing**
- **gravitoreceptors**
  - blood pressure sensors in large blood vessels
- **autonomic processes**
  - fast blood pressure regulation
  - heart beat frequency
  - nausea / vomiting
- **somatosensory**
- **spatial orientation**
- **balance control**
VOR: 8 msec
OKR and Smooth pursuit: >75 msec
head impulse test in unilateral loss
standard video (50 Hz)
pathology: central compensation

the other labyrinth does NOT take over
simulation of oscillopsia \( \approx \) reduced dynamic visual acuity in case of bilateral vestibular areflexia
Dynamic Visual Acuity (VA) measurement

treadmill: 2, 4 and 6 km/h
decrease of VA during walking

Normalized VA difference

normal values

Velocity [km/h]

- 0.21
- 0.20
- 0.30

bilateral loss: visual acuity drops below 0.4
which complaints are related to vestibular deficits?

which complaints are related to natural limitations?
- acute vertigo in case of a sudden change of function of **one** labyrinth (harmonie vestibulaire)

- severe nausea, imbalance and intolerance to head movements in case of sudden bilateral loss (no nystagmus)

- permanent complaints / symptoms in patients with centrally compensated unilateral or bilateral vestibular function loss
  
  - increased neuro-vegetative sensitivity
  - orthostatic hypotension
  - reduced automatisation of orientation in space
  - increased sensitivity for optokinetic stimuli
  - reduced automatisation of balance
  - reduced automatisation of gaze stabilisation
  - reduced dynamic visual acuity
  - oscillopsia
  - secondary: fear/anxiety and fatigue
many vestibular syndromes where vertigo is the leading symptom

- Benign Paroxysmal Positioning Vertigo and Nystagmus
- vestibular neuritis or labyrinthitis / peripheral vestibular ischemia
- pseudo vestibular neuritis: vestibular TIA or infarction
- motion sickness / mal de debarquement
- Meniere’s disease (MD)
- recurrent vestibulopathy (vestibular Meniere? no early stage of MD?)
- vestibular migraine (benign paroxysmal vertigo of childhood?)
- vestibular paroxysms (neuro-vascular compression vestibular nerve, analogon trigeminus neuralgia)
- vestibular epilepsy
- fistula / superior canal dehiscence syndrome (SCDS)
-- central vestibular vertigo
which complaints are related to vestibular deficits?

which complaints are related to natural limitations?
vestibular labyrinth senses **low** frequency motions: movement
cochlear labyrinth senses **high** frequency motions: sound
auditory: sounds = high frequency vibrations
vestibular: movements = low frequency vibrations
canals: rotations
statoliths: translations + tilt
myosine filaments

action potentials

sensitive

less sensitive

80 mV

60 mV

120 mV
Ewald’s 2nd Law
acceleration / inertia of mass

elasticity

viscosity (friction)

latency SP = 0.8 ms

max. deflection

cupula = 2 ms

maximum deflection ≈ 1°

latency VOR = 8 ms
canals are insensitive for translations or gravity
(specific mass endolymph = specific mass cupula)

exceptions: alcohol, cupulolithiasis etc
canals are insensitive for translations or gravity (specific mass endolymphhe = specific mass cupula)

 exceptions: alcohol, cupulolithiasis etc
canals are insensitive for constant rotations

viscosity (friction)

mass → elasticity

back_{cupula} = 20 \text{ s}
Ewald's 1st Law
we almost always stimulate 2 labyrinths
asymmetries only for fast head movements
VOR 3D: nystagmus 3D

direction  =  fast phase
magnitude  =  slow phase

*horizontal (left – right)*
*vertical (up – down)*
*torsional (in- and extorsion)*
in-torsion

ex-torsion

up

down

right

left

nose
frequency dependence of semicircular canals?
frequency dependence canals: gain

sensitivity

0.1 Hz  10 Hz

K

B

B

I

frequency (Hz)

calorics  chair  head impulses

I ~ mass
B ~ friction (visc)
K ~ elasticity
ageing (>60) frequency dependence canals
presbyo-vertigo

sensitivity

general population

elderly > 65 yo

frequency (Hz)
labyrinth
• rotations: canal system
• translations + tilt: statolith systems

utriculus + sacculus
accelerometers
• function based on inertia of statoconia mass
• multi-directional symmetrical sensitivity
• frequency dependence
velocity

constant velocity

acceleration

deceleration

no discrimination between translation and tilt possible
frequency dependence
semicircular statolith systems?
frequency dependence statolith systems (tilt or translation)
limitations labyrinth

- canals: no difference between constant rotation and stand still
- statololiths: no difference between constant translation and stand still
  no difference between tilt and translation
The diagram illustrates the relationship between sensitivity and frequency for the statolith in response to tilt or translation. The x-axis represents the frequency in Hz (0.2 Hz, 2 Hz, 20 Hz), and the y-axis represents sensitivity. The statolith sensitivity decreases as the frequency increases from 0.2 Hz to 20 Hz, indicating diminishing response to higher frequencies.
sensitivity

vision and/or propriocepsis

statolith

canals

frequency (Hz)

0.2 Hz

2 Hz

20 Hz

correct

...... tilt or translation
some facts and findings that need to be explained

- divers under water can’t orient themselves without vision!
  submersion in water:
  principle of inertia of mass in labyrinth remains
  → normal detection of accelerations should be possible

- no detection of orientation when covered by an avalanche

so: the brain needs multi-sensory input or pre-knowledge
otherwise statolith input is neglected:

…….falling asleep
which complaints are related to natural limitations?

motion sickness!
The graph illustrates the relationship between sensitivity and age, showing a peak in motion sickness around the age of 20 years and a decrease as age increases.
VOR
Vestibulo-Collic, Cervico-Collic
Vestibulo-Spinal, Perception
thalamus

vestibular most active hemisphere located in the non-dominant hemisphere

labyrinth stimulation activities ipsi-laterally

PIVC activation: parallel deactivation of occipital and parietal visual areas and vv

perception: cortical network
temporo-insular and temporo-parietal cortex
parieto-insular vestibular cortex (PIVC)
retro-insular cortex
superior temporal gyrus (STG)
inferior parietal lobule (IPL)
precuneus
anterior cingulum
hippocampus
central compensation

joint processes set at work
to achieve fast and optimal recovery
central compensation: neuroplasticity

- increase of visual sensitivity
- usage of commisural input
- formation of new neurons stimulated by movement and high dosage betahistine (Lacour 2004)
- reprogramming connections to balance spontaneous activity of both vestibular nuclei
No cerebellar shut down

Harmonie vestibulaire directed towards lesion side:
- slow phase
- falling
- finger pointing

Sedation impairs central compensation
Methylprednisolon (100 mg every 2-days ▼) + Betahistine 2-3 dd. 48 mg
250 mg/ml endolymphe
50 mg/kg in cats

New neurons and connections
localisation of labyrinth dysfunction in detail is now more often possible, but requires complex equipment
- Benign Paroxysmal Positioning Vertigo and Nystagmus
- vestibular neuritis or labyrinthitis / peripheral vestibular ischemia
- pseudo vestibular neuritis: vestibular TIA or infarction
- motion sickness / mal de debarquement
- phobic postural vertigo / visual vertigo / anxiety / psychogenic vertigo / conversion
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ethiology  paroxysmal positioning dizziness or vertigo

- canalolithiasis or cupulolithiasis
- orthostatic hypotension
- panic and anxiety disorders
- hyperventilation
- peripheral vestibular hyporeflexia
- post-alcoholic vertigo
- central vestibular pathology (vermis / Arnold Chiari)
Benign Paroxysmal Positioning Vertigo in general:

- induced by a change of head position relative to gravity when laying down!
  - after turning from one side to another in bed
  - washing hair under the shower
  - reaching for something placed high

- vertigo is induced after a latency (1 – 60 secs)
- nystagmus follows vertigo within a few seconds
- vertigo and nystagmus last about 5-20 secs
- vertigo and nystagmus are less upon repetition
- nystagmus reversible (when coming up again)

Differences between HC, PC and AC canalolithiasis
cupulolithiasis hypothesis (Schuknecht)

calcium carbonate crystals attach to cupula

cupulolithiasis:
head position change leads to long lasting vertigo and nystagmus without a clear latency or decrease upon repetition (no fatigue) but with slow central adaptation
canalolithiasis hypothesis (Epley)

calcium crystals sink into canal and clod together

canalolithiasis:
fast position change leads after a latency to vertigo and nystagmus
that decreases in time and upon repetition (fatigue)
possible causes of canalolithiasis and cupulolithiasis

- disturbance in statolith metabolism
  (vascular, ageing, Ca^{2+} metabolism/osteo-porosis?)

- head trauma (statolith detachment)

- bed rest (clod formation in canals)

- neuritis vestibularis, labyrinthitis

- ear surgery

- idiopathic
AVOR: free APP for Iphone/Ipad

PC: rotatory-upbeat nystagmus
HC: horizontal nystagmus
AC: downbeat-rotatory nystagmus

VOG recording is the best way to analyse the nystagmus precisely

(fatigue prevents us to see the same type of eye movement when we repeat the diagnostic positioning manoeuvre)
- mostly in the posterior canal
- less in the horizontal canal
- seldom in the anterior canal

why?
due to their orientation of the canals in the head:

- mostly in the posterior canal
- less in the horizontal canal
- seldom in the anterior canal

AD

nee

utriculus

canaloliths

supine position
posterior canalolithiasis AD
Dix-Hallpike: diagnostic manoeuvre for PC-BPPV
Halpike: posterior canalolithiasis AD
right PC-canalolithiasis
or cupulolithiasis

to the right
Hallpike

PC-AD

to the left
left PC-canalolithiasis
or cupulolithiasis

to the right
Hallpike
to the left
PC-AS
Therapy

hope and pray
natural remission 100 hrs ?
action!

• Epley
Hallpike is a good begin!

but Epley finishes the job
Epley manoeuvre PC-AD
wait minimal 30 secs after cessation of nystagmus
most common vestibular syndromes

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Meniere’s Disease (ethiology ?)
spontaneous vertigo attacks lasting 20 minutes to many hours
nausea, vomiting
hearing loss, tinnitus and/or fullness
with or without dropatacks

Recurrent Vestibulopathy (ethiology ?)
similar as Meniere’s disease but no hearing loss or tinnitus

Vestibular Migraine / BPPV of childhood (ethiology ?)
spontaneous vertigo attacks lasting seconds to days
nausea, vomiting
not obligatory: history or present headaches, and/or aura’s
specific triggers: light, food, fatigue, sleep, hormonal cycli ….
history: migraine or BPPV of childhood
Meniere’s Disease and Recurrent Vestibulopathy
  counseling
  3 dd 48-120 mg betahistine > 2 months
  3 dd 25-50 mg cinnerazine
  25 mg promethazine / 20 mg primperan at an attack
  transtympanic dexamethason (5 mg)
  transtympanic gentamycin (20 mg in buffer)
  selective neurectomy or labyrinthectomy

Vestibular Migraine
  prophylaxe:
  propanolol 1-3 dd 40 mg or metoprolol 1-2 dd 50-100 mg
  valproic acid 1-3 dd 500 mg
  topiramaat 1 dd 100 mg
  amitriptyline 1-3 dd 25-50 mg

BPPV of childhood
  1 dd 12.5 mg metoprolol
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acquired bilateral vestibular areflexia

- gentamycine intoxication (aminoglycosides)
- ageing
- auto-immune, vascular
• prevalence of severe bilateral loss (US): 80/100,000  
  (Della Santina et al. 2009)

• patients with acquired severely reduced bilateral vestibular function do suffer and need treatment  
  (Guinand et al. 2012)

• role for a substitution system or implant  
miniature 6-DOF detector in the belt
4x30x40 mm (gyroscopes + accelerometers)
vibration belt on the trunk
12 vibrators

battery + processor
with zero-posture reset
sensor random

sensor on

placebo effect?

double blind placebo controlled study
vibrotactile vest and belt: supports body tilt and body rotation perception

• in 64 out of 83 patients the effect is significant more than placebo:
  vibrotactile feedback improves automatisation of balance and gait in many patients

• 1 system is now used by a patient for about 3 years: he reports impressive increase in his quality of life

• the new adjustable system allows easy testing in patients whether they feel any benefit

• 5 patients are now provided with an individually customized belt for long term ambulatory evaluation study sponsored by health insurance companies
vestibular implants: state of the art knowledge (Guyot, Merfeld, Kingma, Stokroos)
vestibular implants: state of the art knowledge  Geneve – Maastricht
(vd Berg, Guinand, Guyot, Merfeld,, Stokroos, Kingma)

- surgical routes in humans explored for stimulation of PC, HC and AC (vdBerg, Stokroos)
- technology developed (Kingma, Guyot, Merfeld, CLONS)

Geneve: extra ampular nerve stimulation (PC and HC)
Maastricht: intra-ampular stimulation (PC, HC and AC)

- per-operative and chronic stimulation studies in humans (Geneva / Maastricht)
- Maastricht: 2 patients in Octobre 2012: full vestibular implant (1st in the world):
  stimulation of all 3 canals possible leading to correct 3D eye movements
  VOR increases during rotation
  DVA improves
I hope this was useful to you and help you with the management of your patients.

thank you

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