causes of dizziness and vestibular dysfunction

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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 unilateral loss or fluctuating function (neuritis, Ménière…)

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

slow unilateral loss (vestibular schwannoma, aging):

no vertigo or nystagmus

what happens in case of acute bilateral loss?
acute bilateral vestibular loss

no vertigo, no nystagmus

severe unsteadiness / ataxia

intolerance for voluntary head movements

clear neuro-vegetative symptoms
in case of acute asymmetries
- vertigo and nystagmus

slow unilateral, slow or acute bilateral loss
- no vertigo or nystagmus

don’t miss the BVL diagnosis by focusing on vertigo alone
take home message:

vertigo is only 1 aspect of vestibular loss
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 detection of gravity-vector for maintaining balance with fast vestibulo-spinal corrections
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 in case of: motor learning (retardation in congenital areflexia) maintaining postures that need fast feedback when other senses are compromised;

- soft surface (wind-surfing)
- in darkness
- in presence of confusing optokinetic stimuli

bilateral loss: constant visual anticipation or support is necessary

bilateral areflexia leads to degeneration of “head direction” and head “place” cells in the hippocampus
patient with severe bilateral vestibular hyporeflexia
stop walking when talking: predictor of falls

slow tandem walk
needs fast feedback ..... but is not there!
imbalance as corrections are too slow

fast tandem walk
uses visual anticipation and mass inertia
falls in case of unexpected imbalance
thalamus

visual cortex

midbrain

pons

cerebellum

visual image stabilisation: >75 msec

oculomotor nuclei

vestibular nuclei

vestibular image stabilisation: 8 msec
head impulse test in unilateral loss
standard video (50 Hz)
pathology: central compensation

the other labyrinth does NOT take over
loss of gaze stabilisation (towards bad-side) especially for fast head movements
head impulse test in bilateral loss

CASIO Exilem: high speed recording (300 Hz)
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 (maximum VA decrease)

- 0.2  - 0.2  - 0.3

Velocity [km/h]

- BV Patients
- Healthy Subjects
acute vestibular asymmetries (attacks)
- vertigo, spont. nystagmus, fall-tendency, nausea

consequence of slow or acute bilateral vestibular loss:
- often persisting neuro-vegetative symptoms
- reduced ability for fast balance correction
  (need for visual anticipation and support: fear to fall)
- reduced dynamic visual acuity
- reduced perception of self motion and orientation
- intolerance for strong optokinetic stimuli
- imbalance and poor DVA: high cognitive load: 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
causes of 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)
cupulolithiasis hypothesis
on both sides of the cupula

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
calcium crystals sink into canal and clod together
canalolithiasis hypothesis on both sides of the cupula
canalolithiasis:
fast position change leads after a latency to vertigo and nystagmus that decreases in time and upon repetition (fatigue)
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

(Nuti et al: natural remission)
relevant physical and pathophysiological considerations

- limited presence of small canaloliths in endolymph might be quite normal and asymptomatic

- small canaloliths induce less flow and cupula deflection than large canaloliths

- vigorous head movement and vibrations:
  1. big canaloliths may fall into smaller canaloliths (decrease of impact = fatigue)
  2. distribution of canaloliths over the whole labyrinth (multi-canal BPPV)

- increase of presence of canaloliths:
  1. cause BPPV (recidives)
  2. make canals sensitive for translations and gravity
  3. might obstruct endolymphatic flow in ductus reuniens/endolymphaticus and lead to pathophysiological conditions (MM ?)
KEY QUESTION

what is the cause of canalolithiasis and cupulolithiasis
..... or density changes endolymphe or cupula ???

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

- head trauma (statolith detachment)

- bed rest (clod formation in canals)

- neuritis vestibularis, labyrinthitis

- ear surgery

- idiopathic
AVOR: free APP for Iphone/Ipad
CDS, Phobic Postural Vertigo, Visual Vertigo

subjective, fluctuating instability, fear to fall and vegetative symptoms, hours,
induced by crowds, visual stimulation, improves by alcohol and during physical activity,
increases during the day, avoidance behaviour, can be secondary to vestibular deficits

anxiety / psychiatric disorder ? (primary or secondary)
motion sickness
canals are insensitive for constant rotations
labyrinth
  • rotations: canal system
  • translations + tilt: statolith systems

utriculus + sacculus
accelerometers
  • function based on inertia of statoconia mass
  • multi-directional symmetrical sensitivity
  • frequency dependence
no discrimination between translation and tilt possible
<table>
<thead>
<tr>
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<th>Orientation in space: constant rotation or stand still?</th>
<th>Orientation in space: constant translation or stand still?</th>
<th>Orientation relative to gravity: tilt or translation?</th>
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<tbody>
<tr>
<td>canals:</td>
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<td>statoliths:</td>
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When correct interpretation fails (gravity / selfmotion)

**motion sickness**
sensitivity

vision and/or propriocepsis

statolith

canals

0.2 Hz

2 Hz

20 Hz

frequency (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
vestibular perception needs to be confirmed by vision or propriocepsis

otherwise motion sickness might occur

- if we are AWARE we lose orientation and when correct perception of gravity vector fails when correct perception of self motion fails

- almost all subjects are susceptible with correct stimulus unless a low neuro-vegetative sensitivity

- a (partly) working labyrinth is prerequisite for Motion Sickness
Meniere’s Disease (ethiology ? genetically determined ?)
spontaneous vertigo attacks lasting 20 minutes to many hours
nausea, vomiting
hearing loss, tinnitus and/or fullness
with or without dropatacks

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

Vestibular Migraine / BPV 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 BPV of childhood
Meniere’s disease (MD):

- pathophysiology?
- hydrops
- metabolic, genetic
- obstruction ductus reuniens / endolympathicus by statoconia debris?
- bilateral - unilateral

Diagnosis: 3D Flair MRI + transtympanic gadolinium?
fistula / superior canal dehiscence syndrome (SCDS)
superior canal dehiscence syndrome (SCDS) or fistula
WHY

fistula / superior canal dehiscence syndrome (SCDS)

study of skulls: SCDS common feature, often asymptomatic
I hope this was useful to you and help you with the management of your patients.

thank you

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