Beyond BPPV and Meniere's Disease

What vertiginous diseases ENT doctors may encounter

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Most Frequently Encountered Vertiginous Diseases

- BPPV
- Meniere's disease
- Vestibular migraine
- Vestibular neuritis

……and…..

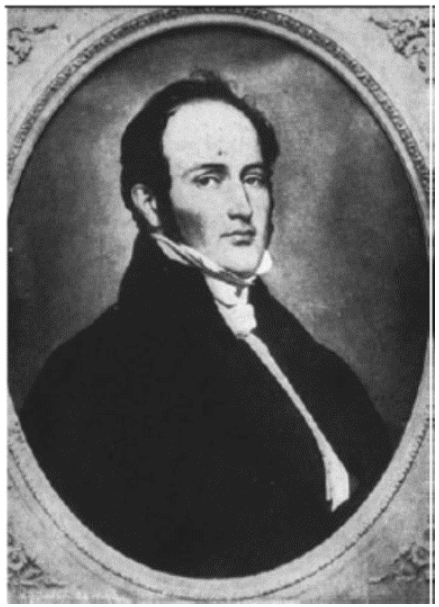
……. something else

Benign Paroxysmal Positional Vertigo (BPPV)

- Natural history：
  - Prevalence：2.4%
  - 17~42% of all vertigo patients
  - Age：>50 y/o；F:M≈2:1
  - May be recovered spontaneously after a period
    - average 1.5 months
  - Recurrence (in a few weeks to years)
    - Annual recurrence rate: 12%
    - Accumulation recurrence rate: 50%

Ménière's Disease (MD)

- History
  - Proposed by French doctor, Prosper Menière, in 1800s
  - Meniere's triad: tinnitus, recurrent vertigo, fluctuating hearing impairment
    - A. Vestibular symptoms (Recurrent attacks of vertigo)
    - B. Auditory symptoms (Tinnitus and Fluctuant hearing loss - mainly unilateral low tone SNHL)
    - C. Aural fullness
  - Pathogenesis: endolymphatic hydrops in the cochlea and vestibula
  - Called as "耳水不平衡"



Prosper Ménière (1799 – 1862)

Ménière's Disease

- Epidemiology
  - Average age of MD onset: forth decade (20~60↑ y/o)
  - Prevalence: > 43/100,000
    - ~0.2% (may be over estimated)
  - Incidence: 4.3/100,000 per year
  - 3 ~11% of all vertigo patients
  - Female : Male = 1.3 : 1

Ménière's Disease - Diagnosis

Diagnosis of Meniere's Disease (AAO-HNS 1995)

1. Recurrent spontaneous and episodic vertigo. A definitive spell of vertigo lasting at least 20 min, often prostrating, accompanied by disequilibrium that can last several days; usually nausea or vomiting, or both; no loss of consciousness. Horizontal rotatory nystagmus is always present.
2. Hearing loss (not necessarily fluctuating)
3. Either aural fullness of tinnitus, or both

Certain Ménière's disease

- Definite disease with histopathological confirmation

Definite Ménière's disease

- Two or more definitive spontaneous episodes of vertigo 20 minutes or longer
- Audiometrically documented hearing loss on at least one occasion
- Tinnitus of aural fullness in the treated ear

Probable Ménière's disease

- One definitive episode of vertigo
- Audiometrically documented hearing loss on at least one occasion
- Tinnitus of aural fullness in the treated ear

Possible Ménière's disease

- Episodic vertigo of the Meniere type without documented hearing loss, or sensorineural hearing loss, fluctuating or fixed, with disequilibrium but without definitive episodes

**In all cases, other causes must be excluded*

Diagnosis of Meniere's Disease (AAO-HNS 2015)

Definite Ménière's disease

- ≥ 2 definitive spontaneous episodes of vertigo, each lasting 20 minutes to 12 hours
- Audiometrically documented low- to medium-frequency sensorineural hearing loss in one ear, defining the affected ear on at least one occasion before, during or after one of the episodes of vertigo
- Fluctuating aural symptoms (hearing, tinnitus or aural fullness) in the affected ear

Probable Ménière's disease

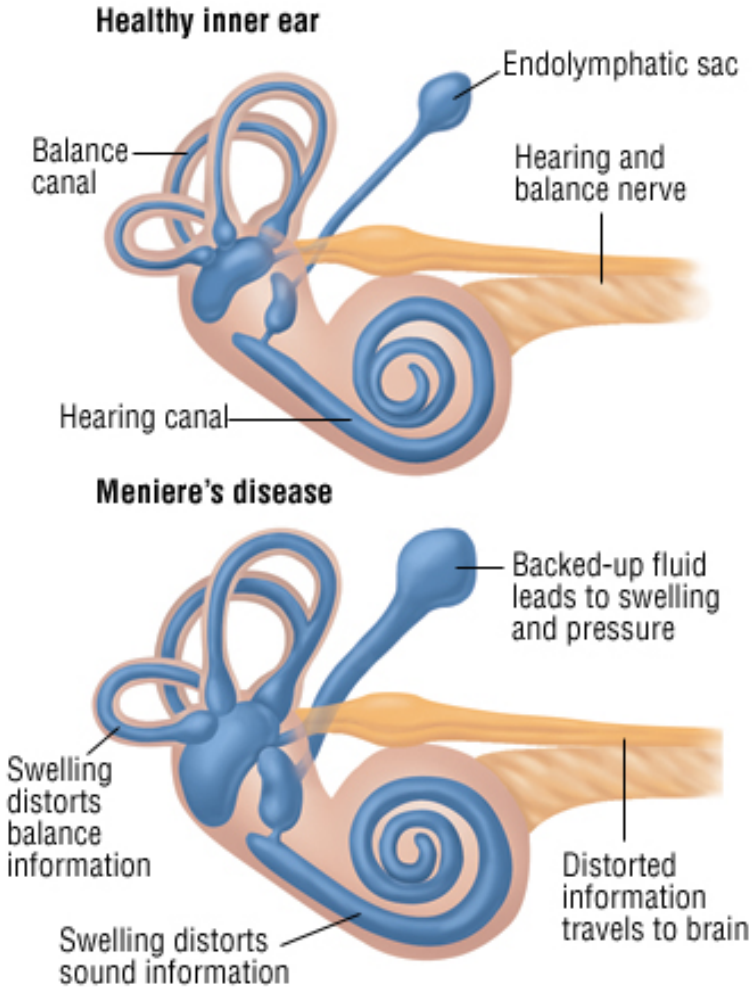
- Two or more episodes of vertigo or dizziness, each lasting 20 minutes to 24 hours
- ~~Audiometrically documented hearing loss on at least one occasion~~
- Fluctuating aural symptoms (hearing, tinnitus or aural fullness) in the affected ear

**In all cases, Not better accounted for by another vestibular diagnosis*

Ménière's Disease - Staging

Staging of Meniere's Disease (AAO-HNS 1994)	
Stage	Four-Tone Average (dB)
1	≤25
2	26 – 40
3	41 – 70
4	>70

Staging is based on four tone average (arithmetic mean rounded to nearest whole number) of pure-tone thresholds 0.5, 1, 2, and 3 kHz of worst audiogram during interval 6 months before treatment. This is same audiogram that is used as baseline evaluation to determine hearing outcome from treatment. Staging should be applied only to cases of definite or certain of Meniere's disease .



https://www.health.harvard.edu/a_to_z/menieres-disease-a-to-z

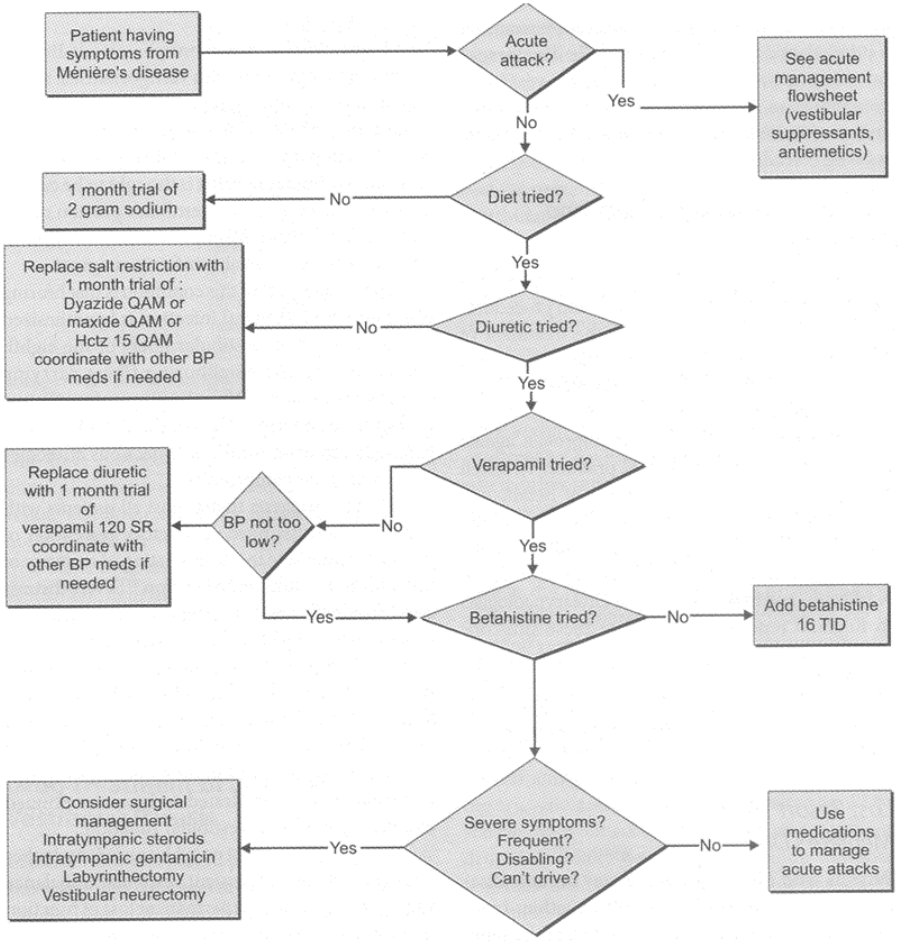
Ménière's Disease – Management (I)

- Nonsurgical treatment
  - Acute treatment of MD

Vestibular suppressants, arranged in order of preference			
Drug (brand name)	***Dose***	***Pharmacologic class***	***Adverse reactions***
Meclizine (Bonamine 保耐暈片….)	12.5-50 mg q 4-6h	Antihistamine, Anticholinergic	Sedating, precaution in prostatic enlargement
Clonazapam (Clonopam 克癇平錠, Rivotril 利福全….)	0.5 mg BID	Benzodiazepine	Mild sedating, drug dependency
Dimenhydrinate (Trimin 克暈片, ….)	50 mg q 4-6h	Antihistamine, Anticholinergic	Same as Meclizine
Diphenhydramine (Diphenhydramine 得拉敏膠囊, Vena 柏那錠 ….)	25-50 mg (1 dose) given acutely orally, IM or IV	Antihistamine, Anticholinergic	Same as Meclizine with more sedating effect
Diazepam (Valium煩靜錠, Bayu 拔憂錠, …..)	2-10 mg (1 dose) given acutely orally, IM or IV 2 mg BID	Benzodiazepine	Sedating, respiratory depressant, drug dependency, precaution in glaucoma
Lorazepam (Ativan安定文錠 , Lowen 樂穩, …..)	0.5 mg BID	Benzodiazepine	Mildly sedating, drug dependency

Ménière's Disease – Management (II)

- Nonsurgical treatment
  - Prophylactic Treatment of MD
    - Diet
    - Diuretics
    - Verapamil
    - Betahistine
  - Chemical ablation
    - Aminoglycoside IT
      - Gentamicin
      - May cause SNHL



Ménière's Disease – Management (II)

- Surgical treatment for MD (Rarely performed)
  - Nondestructive:
    - Endolymphatic sac surgery
    - Vestibular implants (Under survey on human application)
  - Destructive:
    - Cochleosacculotomy
    - Labyrinthectomy
    - Vestibular neurectomy

Vestibular Migraine

- Epidemiology
  - Prevalence: 0.98%
  - 3.2~9% of all vertigo patients
  - Female : Male = 3 : 1
  - 70% patients may present anxiety of depression symptoms
  - 10% of general population has migraine headaches, and 1/3 of persons with migraine experience dizziness

Vestibular Migraine – clinical features

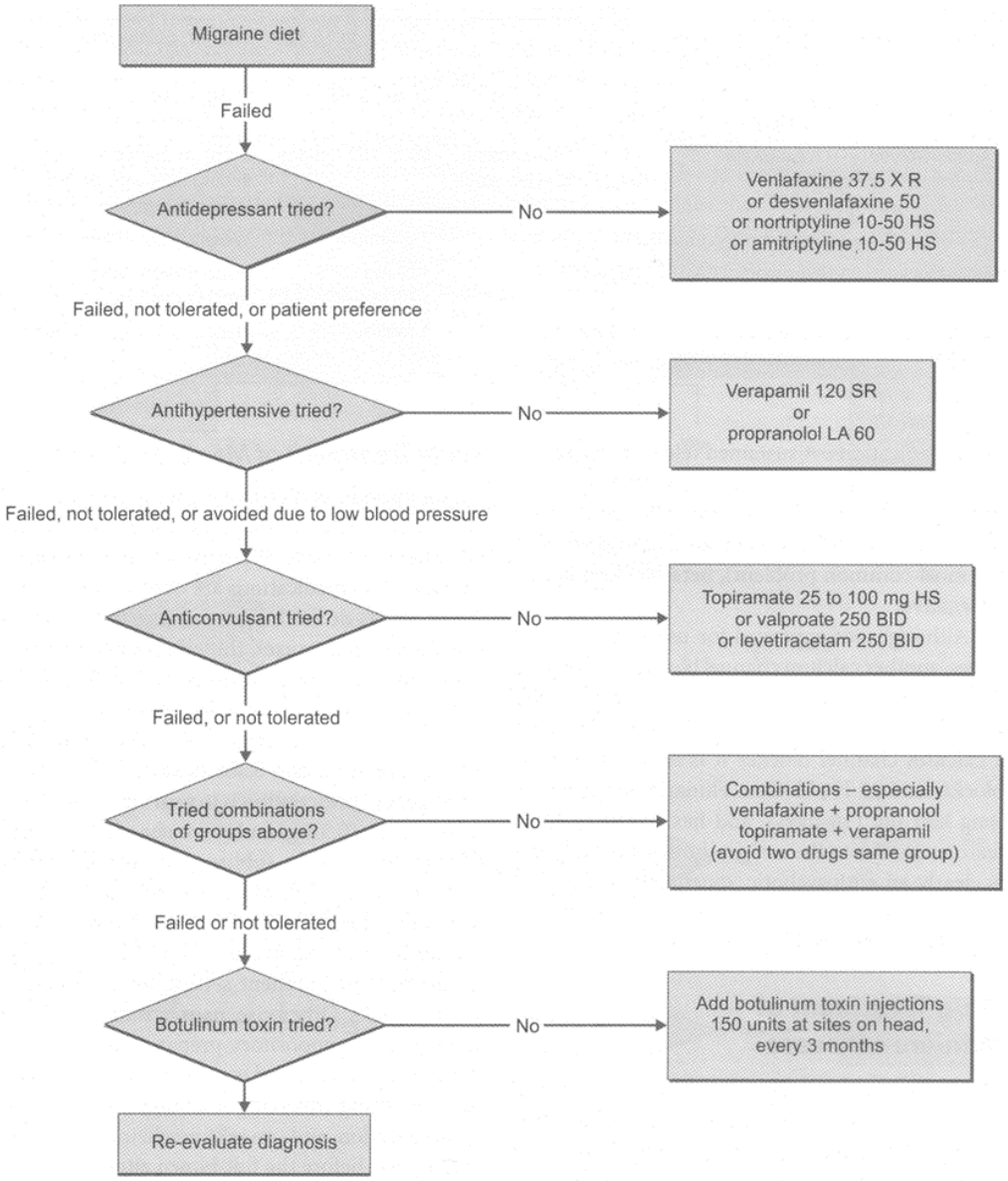
- Common symptoms:
  - Rotational vertigo (70%)
  - Intolerance of head motion (48%)
  - Positional vertigo (42%)
  - Sensation of motion sickness, floating, rocking, tilting and walking on an uneven surface and lightheadedness (less common)
- Duration of symptoms:
  - 5-60 minutes (33%)
  - 1-24 hours (21%)
  - Second to 5 minutes (18%)
  - More than 24 hours (2%)

Vestibular Migraine - Criteria

ICHD-3 criteria for vestibular migraine (International Headache Society/Barany Society) 2018
A current or past history of migraine (with or without aura)
At least **five episods** fulfilling the following **two** criteria:
• Vestibular symptoms of moderate or severe intensity, lasting between 5 minutes and 72 hours, **and**
• At least 50 percent of episodes are associated with at least one of the following three migrainous criteria:
• Headache with at least two of the following characteristics (unilateral, pulsating, moderate or severe intensity, aggravation by routine physical activity)
• Photophobia and phonophobia
• Visual aura
Symptoms not better accounted for by another diagnosis

Vestibular Migraine - Management

- VM is treated in the same way as migraine headache is treated
- Migraine diet: avoidance of triggers
  - Avoidance of:
    - Excessive caffeine (> 2 cups of coffee/day)
    - Monosodium glutamate
    - Chocolate
    - Red wine
    - Aged cheese
    - (kimchi, pizza, etc.)



Vestibular Migraine - Prophylaxis

Common prophylactic medication for Vestibular Migraine				
Medication	Initial dosing	Final dose	Common side effects	Contraindications
Antidepressant				
Venlafaxine in an extended release	37.5 mg XL (1/3 capsule QAM, increased by 1/3 per week)	37.5 XL to 75 XL	Tremor, palpitation	Glaucoma, pregnant women
Amitriptyline or Nortriptylne	10 mg HS	25-50 mg HS	Weight gain, somnolescence	Hypersensitivity, MAOI, clinically significant cardiovascular diseases
Anticonvulsant				
Topiramate	25 mg HS, increase by 25 mg every week	50 to 100 mg/day	Acral paresthesias, word finding difficulty, weight loss, teratogen	Hypersensitivity, pregnant women
Sodium Valproate	500 mg BID	1 gm/day	Weight gain, tremor	Significant hepatic dysfunction, hypersensitivity, urea cycle disorders, porphyria (紫質症)
Beta-Blocker				
Propranolol LA	60 mg QHS	120 mg QHS	Hypotension, fatigue, erectile dysfunction	Cardiogenic shock, sinus bradycardia and greater than first-degree block, bronchial asthma, hypersensitivity
Calcium Channel blocker				
Verapamil in an extended release	120 mg HS	1mg/lb	Hypotension, constipation	Severe left ventricular dysfunction, Hypotension, Sick sinus syndrome, 2nd or 3rd degree AV block, AF or atrial fibrillation and an accessory bypass tract
Flunarizine	5 mg HS	10 mg HS	Tiredness, dizziness, constipation, mouth dryness, body weight gain	Depressive illness, pre-existing symptoms of Parkinson's disease or other EPS

Vestibular neuritis

- Epidemiology
  - Prevalence: 3.5~24/100,000
  - 3~10% of all vertigo patients
  - F:M=1:1
  - Recurrence rate 1.9%

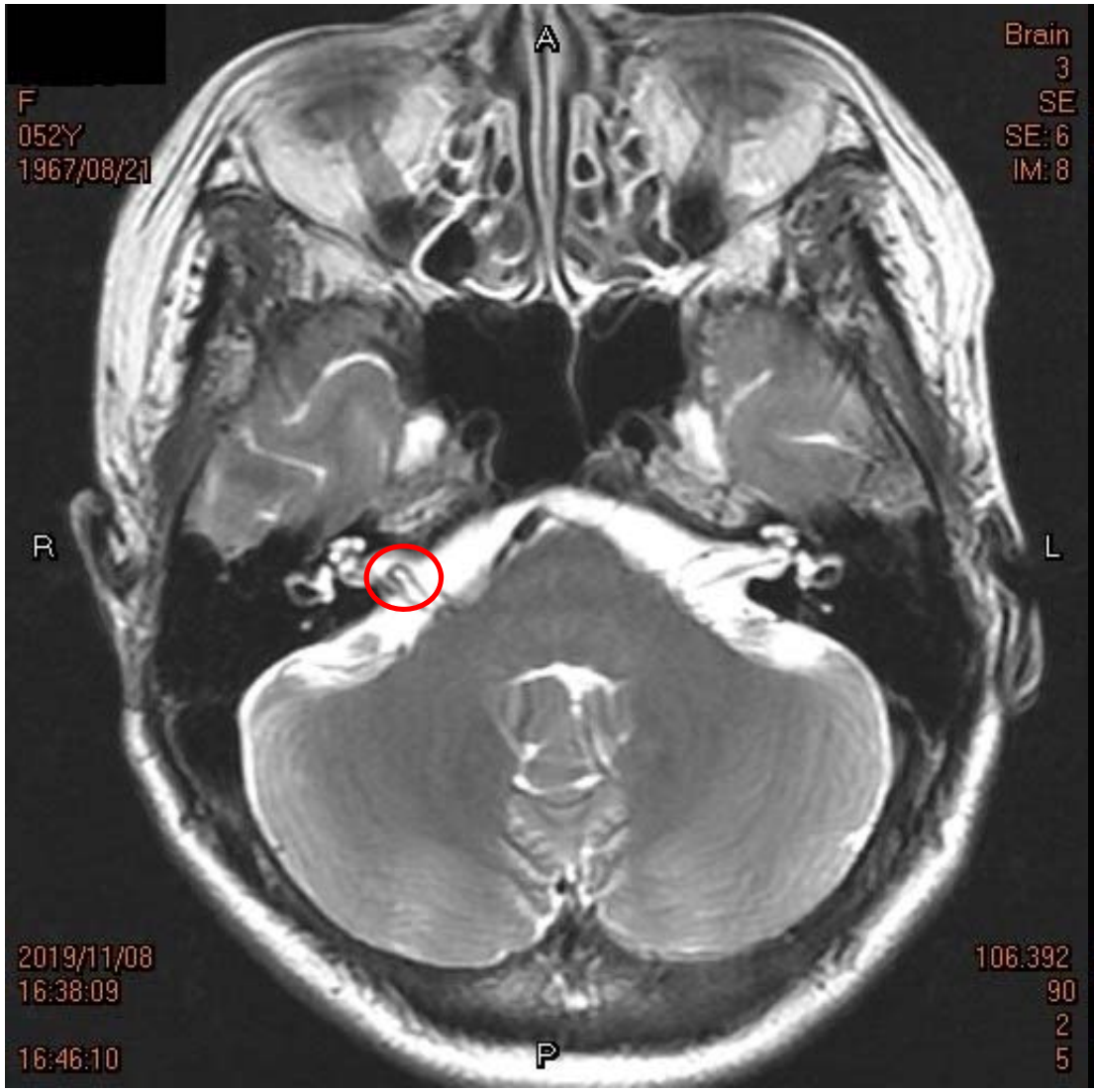
Vestibular neuritis – Diagnosis and Treatment

- Diagnosis
  - History
    - Sudden onset
    - Persistent vertigo
    - Peak of symptoms: 12 – 48 hours after onset
    - May have URI symptoms within 1 month previous to episode
  - Physical examination
    - Spontaneous nystagmus
    - Head impulse test
    - vHIT (if feasible)
- Treatment
  - Steroid
    - Prednisolone 1mg/kg
  - Self-limited
  - Vestibular rehabilitation

Vestibular Paroxysmia

- Epidemiology
  - Prevalence: < 0.5‰ (rare, estimated)
  - ~4% of persistent vertigo/dizziness patients (in medical center)
  - F:M=1:1
  - Mean age: around 50 y/o
- Pathogenesis
  - “vestibular neuralgia” (analog to trigeminal neuralgia)
  - Some kind of vascular compression syndrome
    - Mostly AICA (75%)
    - Seldom PICA
- Treatment
  - Medical:
    - carbamazepine (200–800 mg/day)
    - oxcarbazepine (300–900 mg/day)
  - Surgical
    - reserved

(Strupp, Lopez-Escamez et al. 2016)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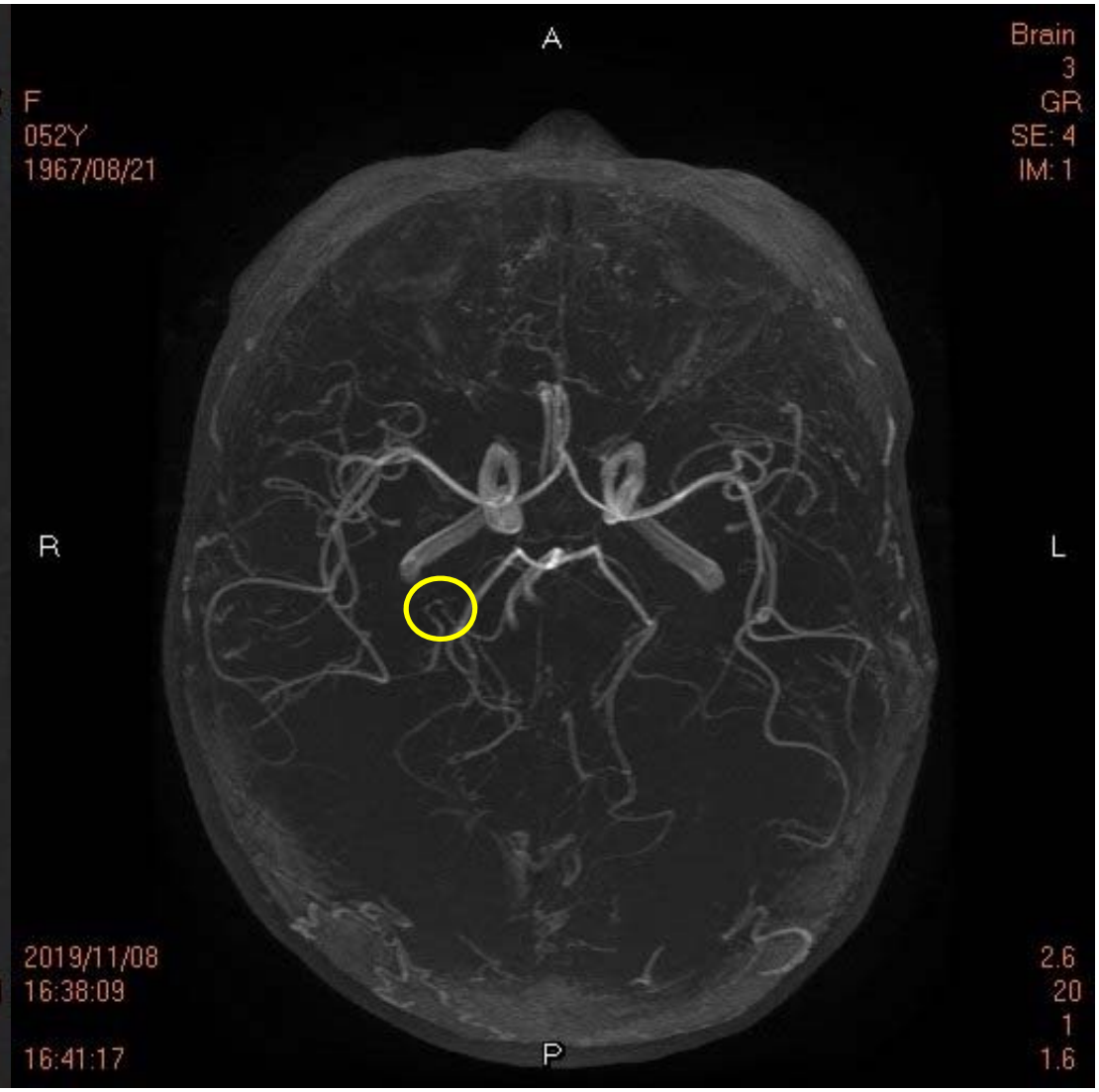
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Vestibular Paroxysmia – Diagnostic Criteria

Definite Vestibular Paroxysmia (***each point needs to be fulfilled***)	**Probable Vestibular Paroxysmia** (***each point needs to be fulfilled***)
A) At least ten attacks of spontaneous spinning or non-spinning vertigo	A) At least five attacks of spinning or non-spinning vertigo
B) Duration less than 1 minute	B) Duration less than 5 minutes
C) Stereotyped phenomenology in a particular patient	C) Spontaneous occurrence or provoked by certain head-movements
D) Response to a treatment with carbamazepine/ oxcarbazepine	D) Stereotyped phenomenology in a particular patient
E) Not better accounted for by another diagnosis	E) Not better accounted for by another diagnosis

Postural Orthostatic Tachycardia Syndrome (POTS)

- Epidemiology
  - Prevalence: ~170/100,000 (1% in American)
  - 5 – 10 times as common as postural hypotension
  - F:M=4 - 5:1
  - Mean age: 15 – 50 y/o
    - 2nd common vertiginous/dizziness disease among children and adolescents
- Etiology
  - Primary
    - Neuropathic
      - Most Common
      - Norepinephrine (NE): high normal or slightly elevated
    - Hyperadrenergic
      - Elevated standing NE level: >600 pg/mL (frequently >1000 pg/mL)
  - Secondary
    - Relates to medical illness

(Thanavaro and Thanavaro 2011)
(Zadourian, Doherty et al. 2018)

POTS - diagnosis

POTS - Definition
1) symptoms that occur with standing[a]
2) an increase in HR of≥30 bpm when moving from a recumbent to a upright posture within 10 min of standing (or≥40 bpm in individuals 12–19 years of age)
3) the absence of orthostatic hypotension[b]
a: symptoms such as lightheadedness, palpitations, tremor, generalized weakness, blurred vision, exercise intolerance, and fatigue b: >20 mmHg drop in systolic blood pressure (BP)

- Presyncope symptoms chronically (≥6 months)
- The standing HR is often≥120 bpm with greater increases in the morning than in the evening

POTS - Treatment

- Drink at least 2 L of water daily
- Intake >200mEq of sodium daily (3-5 g per day)
- Low dose Beta-blocker
  - Propranolol 10 – 20 mg, PO, bid – qid
- Alpha-1 agonist
  - Midodrine 5 -10 mg, PO, tid

(Taylor and Goodkin 2011)
(Thanavaro and Thanavaro 2011)

Persistent postural-perceptual dizziness (PPPD)

- A New Nomenclature (since 2014)
  - phobic postural vertigo (PPV)/chronic subjective dizziness =>PPPD
- Epidemiology
  - Prevalence: invalid, reported to be 2%
  - 15 – 20% diagnosis in dizziness center
  - F:M=2 - 6:1
  - Anxiety-related

(Bittar and Lins 2015, Staab, Eckhardt-Henn et al. 2017, Popkirov, Staab et al. 2018)

PPPD - Diagnosis

Criteria for the diagnosis of Persistent Postural-Perceptual Dizziness (PPPD) – Barany Society 2017 (*All five criteria must be fulfilled to make the diagnosis*)
A) One or more symptoms of dizziness, unsteadiness, or non-spinning vertigo are present on most days for 3 months or more 1. Symptoms last for prolonged (hours-long) periods of time, but may wax and wane in severity. 2. Symptoms need not be present continuously throughout the entire day.
B) Persistent symptoms occur without specific provocation, but are exacerbated by three factors: 1. Upright posture. 2. Active or passive motion without regard to direction or position, and 3. Exposure to moving visual stimuli or complex visual patterns
C) The disorder is precipitated by conditions that cause vertigo, unsteadiness, dizziness, or problems with balance including acute, episodic, or chronic vestibular syndromes, other neurologic or medical illnesses, or psychological distress. 1. When the precipitant is an acute or episodic condition, symptoms settle into the pattern of criterion A as the precipitant resolves, but they may occur intermittently at first, and then consolidate into a persistent course. 2. When the precipitant is a chronic syndrome, symptoms may develop slowly at first and worsen gradually.
D) Symptoms cause significant distress or functional impairment.
E) Symptoms are not better accounted for by another disease or disorder

PPPD - Treatment

- Communicating the diagnosis
- Individualized treatment
  - Vestibular Rehabilitation
  - Medication
    - Selective serotonin reuptake inhibitors (SSRI)
    - Serotonin norepinephrine reuptake inhibitors (SNRI)
    - Responsive in 8-12 weeks, continuous use for at least 1 year

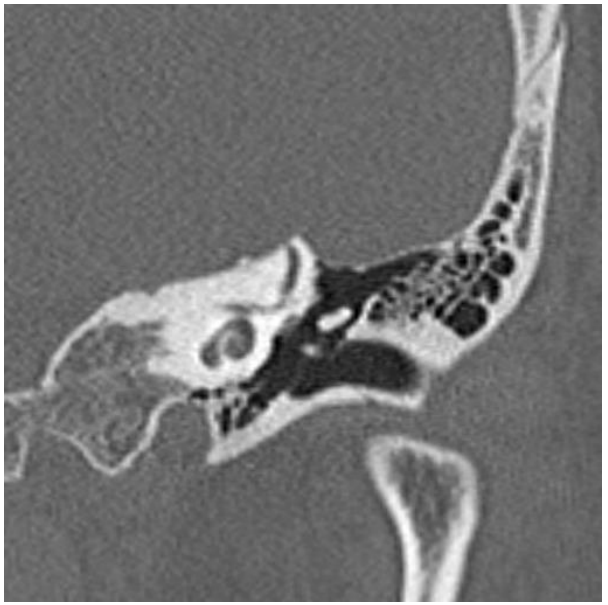
(Popkirov, Staab et al. 2018)

Other vertiginous diseases (I)

- Chronic cervical dizziness
- Vertebrobasilar insufficiency (VBI)
  - Mostly in elder patients
  - May have C-spine osteophytes at C2-C4
  - Related to head movement, similar to BPPV but vertigo persists with postural sustains
  - Diagnosis:
    - Carotid/transcranial Doppler
    - CTA/MRA
  - Treatment:
    - acetylsalicylic acid (ASA) : 30 - 325 mg/day
    - Angioplasty

Other vertiginous diseases (II)

- Superior Semicircular Canal Dehiscence (SSCD)
- Perilymph Fistula (PF)
- Symptoms:
  - vertigo, conductive hearing loss, autophony, tinnitus, sound-induced vertigo (Tullio phenomenon)
- Sings:
  - Positive fistula sign
- Diagnosis
  - Audiometry
    - Conductive hyperacusis
  - cVEMP
    - Decreased VEMP threshold
  - High resolution CT
- Treatment
  - Symptomatic control
  - Surgical treatment
    - Rarely done for SSCD
    - Performs in PF of Horizontal canal/round window if ineffective conservative treatments



Other Remarks

- 1/3 patients of migraine has vertigo/dizziness complaints; 1/2 patients with MD has migraine
- BPPV incidence increases in vestibular neuritis and MD
- Carefully DDx vestibular neuritis from posterior circulation stroke (PCS)
  - HINTS
  - vHIT

Thanks for your attention

Uppsala, Sweden 2018. 6. 13. PM: 10:59

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