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Advanced Vestibular Topics: Examination, Differential Diagnosis and Interventions

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Disclosures

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Learning Outcomes

After this course, participants will be able to:

- Identify at least two tests that would help with differential diagnoses of vestibular conditions.
- Describe at least two measures/tests to help manage vestibular disorders.
- Identify at least three key anatomical structures involved in the central or peripheral vestibular disease process.
- Identify at least three history taking strategies for central and peripheral vestibular disorders.

Vestibular Anatomy and Physiology

- Vestibular Apparatus
- The vestibular system detects angular and linear acceleration through five end organs in membranous labyrinth on each side:
- Otolith organs: saccule, utricle (linear acceleration)
- Semi circular canals: anterior, posterior, and horizontal or lateral canal (angular acceleration)
- Three semicircular ducts are oriented at right angles to one another.

Introduction

- The vestibular system is a complex set of structures and neural pathways that contributes to our sense of proprioception and equilibrium
- Vestibular end organs after sensing angular and linear acceleration transduce these forces to electrochemical signals to be used by the central nervous system
- Central nervous system integrates the information from the vestibular system to stabilize gaze during head motion by means of the vestibular ocular reflex (VOR) and to modulate muscle tone by the vestibulocollic and vestibulospinal reflexes.

  - Casale J. et al. 2020

Physiology

- Sensory neuroepithelium in the utricle and saccule is the macula, and the sensory neuroepithelium in the semicircular ducts is the crista ampullaris.
- Neuroepithelial structures contain specialized mechanoreceptor cells called "hair cells."
- Hair cells contain a vast number of cross-linked actin filaments called stereocilia that are connected at the tips by “tip links”
- Acceleration of endolymph results in the movement of stereocilia, leading to either depolarization or hyperpolarization depending on the direction of the inertial drag.

Casale J. et al. 2020

Physiology

- **Movement towards the kinocilium** causes the interconnected tip links to pull open cation channels resulting in an influx of potassium ions and **depolarization.**
- **Movement in the opposite direction to the kinocilium** causes stereocilia to converge resulting in tip links closing the cation channels leading to hyperpolarization.

- Casale J. et al. 2020

Physiology

- The utricle and the saccule are responsible for sensing linear acceleration, gravitational forces, and tilting of the head.
- macula provides neural feedback about horizontal motion from the utricle and vertical motion from the saccule.
- Angular acceleration and rotation of the head in various planes are sensed by the three semicircular ducts that are oriented at right angles to one another

- Casale J. et al. 2020

Semi Circular Canals and Otolith Organ

- Dilation near the opening to the utricle is called the ampulla which contains a neuroepithelial structure called the "crista ampullaris."
- It is coated by a gelatinous protein-polysaccharide substance known as the cupula which holds the hair cells in place
- The mechanism involved with the function of the peripheral vestibular system involves the acceleration of endolymph within the various structures of the vestibular apparatus
- Utricular excitation occurs during horizontal linear acceleration or static head tilt, and saccular excitation occurs during vertical linear acceleration.
    - Schubert and Minor, 2004

Vestibular and Visual system

- Vestibular system is responsible for sensing motion of the head and maintains stability of images on the fovea of the retina and postural control during that motion.
- Stabilization of a visual target on the fovea can be achieved by various systems, including the vestibular and smooth pursuit oculomotor sys- tems.
- Brain uses target velocity and distance as well as head motion velocity and frequency to determine which oculomotor system is recruited for gaze stability

  - Schubert and Minor 2004

Vestibular and Visual System

- Vestibular system can detect head velocities of up to 550°/s ; head acceleration of 6000 deg/ sec – noted during normal activities like running.
- Ocular system : Mechanisms such as smooth pursuit, generate slower eye velocities (< 60°/s) and have relatively long latencies (up to 100 milliseconds
- VOR latency is 5- 7 milliseconds.
    - Schubert and Minor, 2004

Vestibular and Oculomotor terminology

- **Gaze pursuit/ Smooth pursuit** : the eye follows a moving target
- **Saccadic pursuit**—that is, the gaze rapidly jumps from one fixation point to another
- **Vergence eye movements**—movements during which the eyes do not move in parallel but relative to one another
- **Optokinetic reflex** (consists of smooth pursuit and saccades).
- All these eye movements serve to keep the visual target on the macula stable and thus avoid illusory movements and blurred vision.

  - Strupp et al. 2011

Vestibular and Oculomotor Tests terminology

- **VOR :** Rapid head rotation and fixation of a stationary point
- **VOR Cancellation:** Fixation of a target while rotating the head and moving the target at the same angular velocity
- **Rapid Head Thrust:** The head thrust test is a widely accepted clinical tool that is used to assess semicircular canal function. head is flexed 30 degrees (to ensure cupular stimulation primarily in the tested lateral SCC
    - Strupp et al. 2011

Vestibular and Oculomotor Tests

- **Positional Testing:**
  - Dix Hall Pike / Side lying Test : Test for BPPV (PC – BPPV)
  - Bow and Lean test (HC – BPPV , especially to assess lateralization when unable with supine roll test)
  - Supine Roll Test (HC – BPPV)
- **Gaze evoked Nystagmus:** gaze is assessed at 0- 40 deg, horizontal and vertical
- **Subjective Visual Vertical** : Perception of visual Verticality is altered in Brainstem leasions and also found in cerebellar strokes (Lee et al. 2009)
  - Strupp et al. 2011

Cerebellar regions for oculomotor control

- Flocculus/Paraflocculus
- Nodulus
- Vermis/fastigial nucleus
- **Damage to these structures - usual oculomotor signs seen:**
- Flocculos/ Paraflocculus: abnormal smooth pursuit, downbeat nystagmus, rebound nystagmus, positive VOR cancellation
- Nodulus: Central positional nystagmus, periodic alternating nystagmus
- Vermis/ Fastigial Nucleus : Hypermetric or hypometric saccades

- Strupp et al. 2011

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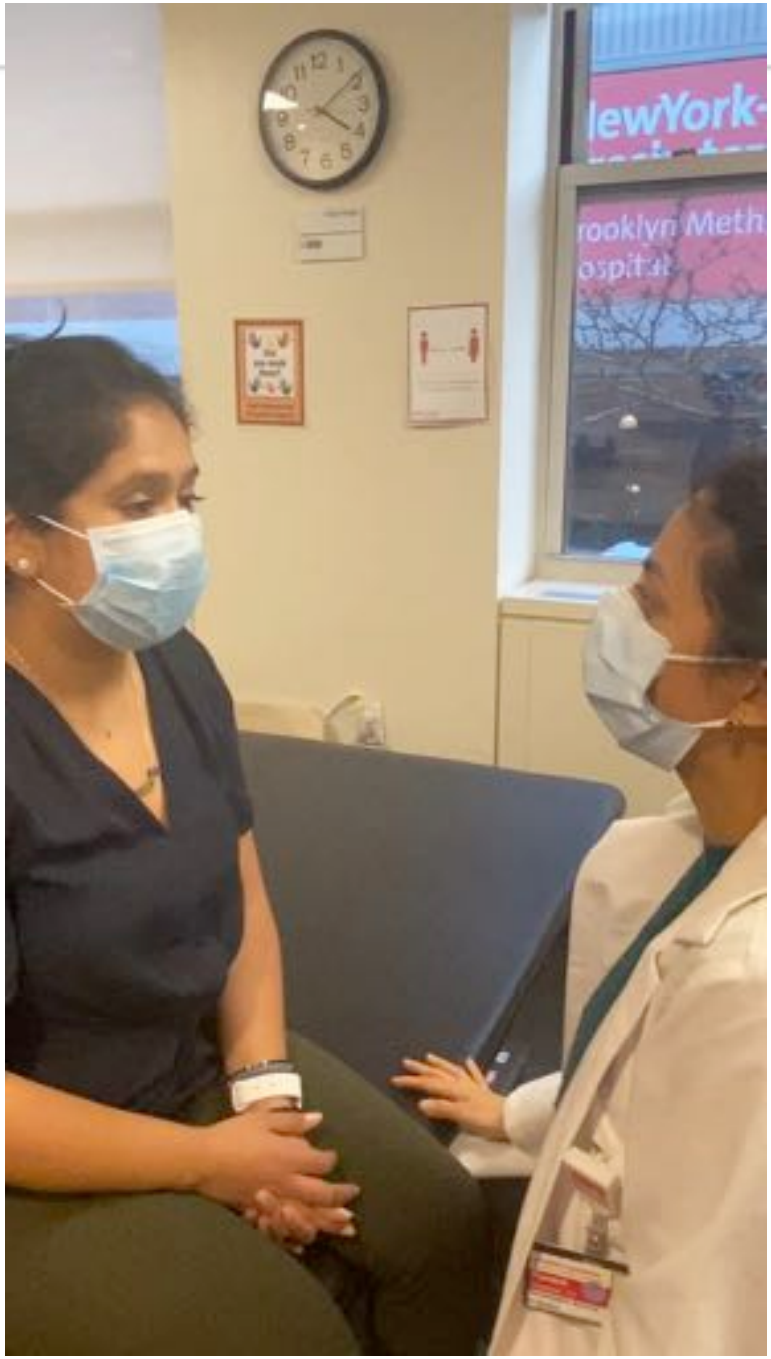
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Acute Vestibular Syndrome: A clinical Challenge

Introduction

- Acute vestibular Syndrome term was introduced by Hotson and Baloh (1998)
- Rapid, unilateral injury to either peripheral or central vestibular structures produces the acute vestibular syndrome, which consists of severe vertigo (prolonged vertigo > 24 hours), nausea and vomiting, spontaneous nystagmus, and postural instability.
- Semicircular canals or their central connections usually produce a rotational vertigo
    - Hotson and Baloh, 1998

Clinical Symptoms

- When otolithic organs are affected a feeling of tilting or linear displacement may occur in disorders affecting the otolithic organs or their projections.
- feeling of imbalance during standing or walking.
- Patients want to lie still and avoid movement.
- Acute vertigo is accompanied by nausea, vomiting, and autonomic distress of varying degrees of severity.
    - Kim and Lee , 2012

Causes

- Peripheral (i.e., inner and vestibular nerve)
  - acute vestibular neuritis (VN), Meniere's disease, and migraine.
- Central (i.e., brainstem and cerebellum)
  - Lateral medullary, lateral pontine, and inferior cerebellar infarctions mimic APV (Acute peripheral vestibulopathy) very closely (Neuman- Toker et al. 2009)

- The peripheral causes of AVS included **Vertebrobasilar ischemic stroke (VBIS) can also cause isolated prolonged vertigo mimicking peripheral AVS

Kim and Lee, 2012

Misdiagnosis is common

- Misdiagnosis in 37% of patients with posterior circulation strokes had an when compared to 16% of anterior strokes
- 9% of cerebrovascular events [ischemic strokes, transient ischemic attacks (TIAs), and subarachnoid hemorrhages] were missed at initial presentation
- 1 –in 5 strokes causing AVS affects a patient under age 50 and 1- in 10 under age 40.
- Typical neurologic signs are absent in about 50% cases
- Only half have severe truncal ataxia without other obvious neurologic or oculomotor signs.
    - Newman- Toker et al. 2009; Tsang B. K.T, et al. 2017

Easy to Misdiagnose

- Usual diagnosis in the emergency is acute peripheral vestibulopathy (APV) and more dangerous brainstem lesions can be missed.
- 25% patients with AVS may have posterior cerebellar infarction
- CT scans have low sensitivity (~16%) for acute infarction, particularly in the posterior fossa . Studies also suggest that false negative MRI can occur with acute vertebrobasilar central vestibulopathies. (upto 12%)
- Higher sensitivity and specificity of identifying central causes of the AVS with HINTS compared to diffusion-weighted (DW)-MRI in the first 24- 48 hours
    - Newman Toker et al. 2009

Central or Peripheral Dysfunction

- Examination
- HINTS
- HINTS PLUS
- STANDING

HINTS: 3 step bedside oculomotor exam

- The mnemonic HINTS stands for
- **Head Impulse**
- **Nystagmus** (observation for nystagmus in primary, right, and left gaze)
- **Test of skew** (alternate cover test for skew deviation)

- Newman- Toker, et al. 2009; Kattah J. 2009

HINTS

- Test of skew
  - vertical ocular misalignment that results from a right-left imbalance of vestibular tone (i.e., neural firing particularly otolithic inputs to oculomotor system)
  - Detected by alternate cover test

- Newman-Toker, et al. 2009

Central vs Peripheral

- Normal HIT
  - Central Localization

  **abnormal HIT (implying APV) can also be present in lateral pontine strokes.
- Nystagmus
- Peripheral :
  - dominantly-horizontal nystagmus that beats only in one direction and increases in intensity when the patient looks in the direction of fast phase (Alexander's Law)
- Central : Horizontal Nystagmus that changes direction on eccentric gaze
  - Vertical or torsional nystagmus in this clinical context is a clear sign of central pathology
- Test of Skew
  - Skew deviation is an insensitive marker of central pathology but ***fairly specific predictor of brainstem involvement among AVS patients***

Newman- Toker, et al. 2009

Central vs Peripheral

- **Skew deviation** in AVS is strongly linked to the presence of brainstem lesions, most often ischemic strokes in the lateral medulla or pons
- **INFARCT:**
- **I**mpulse **N**ormal **F**ast-phase **A**lternating (direction changing) **R**efixation on **C**over **T**est.

- **HINTS should not be solely relied on outside the 72-hour time window or validated clinical context (including age)
- ** HINTS also cannot be relied on in patients with episodic vestibular syndromes -such etiologies as vestibular migraine, Meniere disease or transient ischemic attacks
  - ** Nathan K. et al. 2018; Kattah, J. 2018

HINTS PLUS

- Head Impulse
- Nystagmus (observation for nystagmus in primary, right, and left gaze)
- Test of skew (alternate cover test for skew deviation)
- Sensorineural hearing loss (hearing loss will usually depict a central sign)

- ** The presence of new hearing loss is generally unilateral and on the side of the abnormal head impulse test
- ** Hearing loss more often indicates a vascular rather than viral cause of the acute vestibular syndrome presentation.
  - Newman – Toer et al. 2013

STAnDing - A 4 step algorithm for differentiating acute vertigo in emergency

- SponTAneous nystagmus, Direction, head Impulse test, standiNG – A 4 step algorithm for differentiating acute vertigo in emergency
- They propose a diagnostic algorithm, which includes nystagmus examination performed by emergency physicians as compared to neuro- ophthalmologists (in HINTS study)
- STAnDing algorithm provides the essential tools to recognize the most frequent peripheral vestibular diseases (BPPV and VN) and can help emergency physicians to identify the population of patients with central disease.
    - Vanni et al. 2014

When to recommend imaging

Kim and Lee, 2012

- Older patients presenting with isolated spontaneous prolonged vertigo
- any patient with vascular risk factors and isolated spontaneous prolonged vertigo who has a normal HIT result
- Patients with isolated spontaneous prolonged vertigo who had di- rection changing gaze-evoked nystagmus or severe gait ataxia with falling at upright posture
- Patients presenting with acute spontaneous vertigo and new onset headache, especially occipital,
- Patients with vascular risk factors and acute onset of vertigo and hearing loss without Meniere's history

BPPV

Introduction

- Definition: Benign paroxysmal positional vertigo (BPPV) is a common form of vertigo, accounting for nearly one-half of patients with peripheral vestibular dysfunction.
- commonly attributed to calcium debris within the posterior semicircular canal, known as canalithiasis.
- Epidemiology
  - the lifetime prevalence of BPPV was 2.4 percent
  - The one-year prevalence of BPPV increased with age and was seven times higher in those older than 60 years
  - BPPV was more common in women than men in all age groups, with a reported ratio of 2:1 to 3:1
    - Kim and Zee, 2014

Independent Risk factors associated with BPPV

- BPPV has been associated with
  - Age
  - migraine
  - Head Injury
  - hypertension
  - hyperlipidemia
  - stroke – possibly due to vascular issues. a potential vascular mechanism for at least some cases of BPPV
  - Osteopenia and Osteoporosis (Altered Calcium Metabolosim)*
    - Brevern, V.N. et al. 2007; * Jeong et al. 2009

Pathophysiology

- BPPV is commonly attributed to canalithiasis that is calcium debris within the semicircular canal originating from utricular sac
- Most common site : Posterior semicircular Canal
  - Less common are horizontal canal and anterior canal.
- Usual Causes of PC- BPPV include :
  - Idiopathic (in about 35 % cases)
  - Previous head injury /Whiplash injuries (15%)
  - Association with vestibular pathologies
    - Meniere's
    - Vestibular Neuronitis
    - Maxillary sinus surgery (possibly due to percussive sources)
    - Vestibular Migraine

• Hughes and Proctor, 1997

Clinical Presentation

- Recurrent episodes of vertigo that last one minute or less.
- Vertigo is provoked with position changes.
  - Getting out of bed
  - Bending activities
  - Looking overhead
  - Bed mobility
- Balance dysfunction maybe present in some patients.
- Episodes may occur for weeks to months.
- May self resolve

Differential Diagnosis

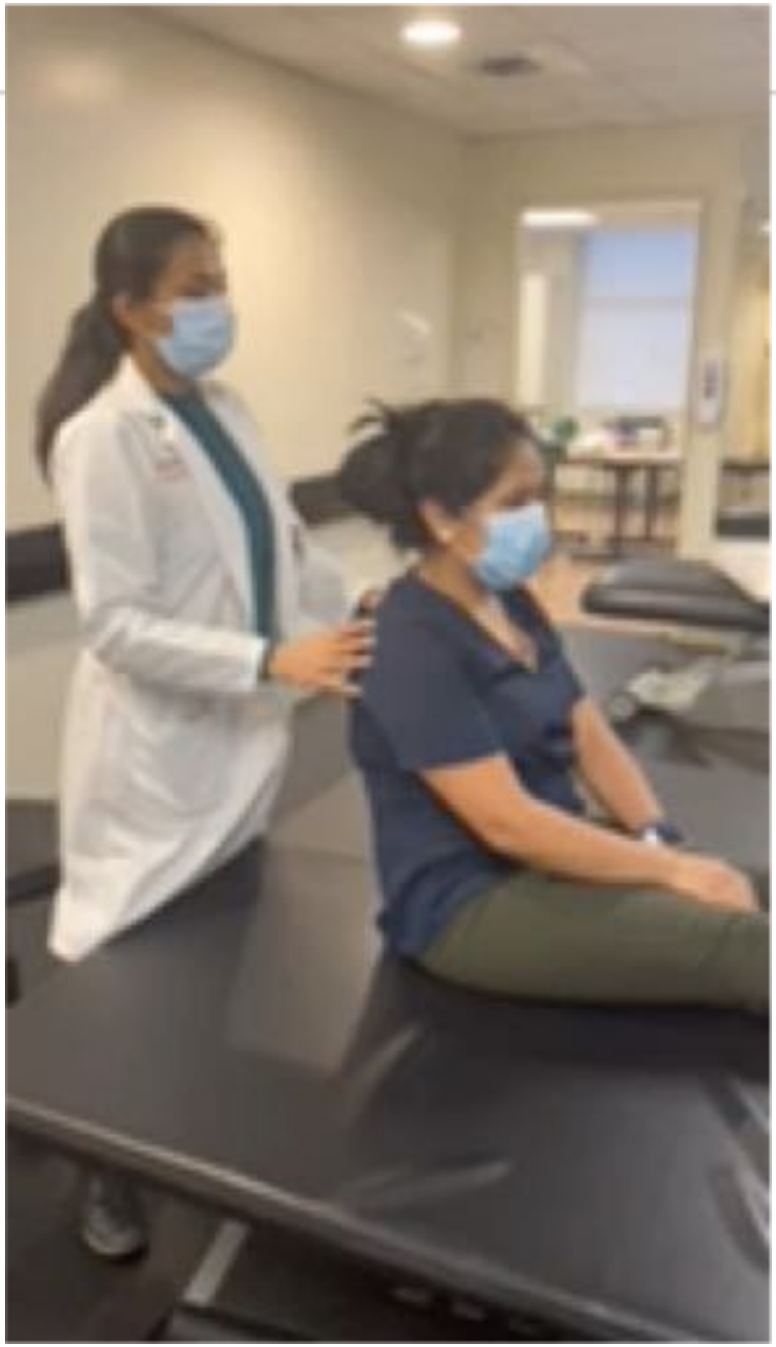
- Postural hypotension
- Chronic unilateral vestibular hypofunction
- Vestibular paroxysmia
- Vestibular migraine
  - Episodes are shorter in duration
  - More frequent subsequent periods of recurrent dizziness than BPPV
  - Positional nystagmus atypical for BPPV
- Central positional vertigo
  - Differentiate downbeat nystagmus from Anterior canal nystagmus.
  - a lack of latency
  - lack of fatigability
  - inability to suppress nystagmus with fixation
    - Barton et al. 2020

Physical Examination

- Dix Hall Pike
- Sidelying Test
- Bow and Lean Test
- Supine head hanging test
- Supine roll test

Dix Hall Pike

- For assessing Posterior canal and Anterior Canal – BPPV (** for AC – BPPV – best test is supine head hanging – Bertholon, et al. 2002)
- With the patient sitting, the neck is turned to one side.
- The patient is then brought into supine position rapidly with the head hanging over the edge of the bed.
- The patient is kept in this position atleast 30 seconds has passed if no nystagmus occurs the patient is then returned to upright



:L Side lying Test

- To test Posterior or Anterior SC BPPV
- For R side lying Test:
- Patient is seated with head turned to R side
- Patient is then brought into sidelying towards the L side.
- Can be used for patients who have limited ROM

** no significant differences were found between Dix Hall Pike Test and Sidelying test (Cohen, H. 2004)

Supine Roll Test/ Horizontal Roll Test

- The nystagmus is elicited by a lateral head turn in the supine position
- This may induce geotropic or apogeotropic nystagmus:
- Geotropic HC-BPPV
  - horizontal nystagmus beating toward the floor.

- Apogeotropic HC-BPPV
  - the induced nystagmus beats toward the uppermost ear
- Affected side is determined by the strength of nystagmus
  - Geotropic: Stronger nystagmus is the side of affected ear
  - Ageotropic: Weaker nystagmus is the side of affected ear

- Herdman S. J. (3rd Edition)

Bow and Lean test

- Used to test for HC BPPV if one cannot lateralize it
- The patient first bends his or her head forward, aligning the horizontal canal with the gravity vector, and then leans his or her head backward
- If it is a R horizontal canal canalithiasis : R beating nystagmus during the bow test and a L beating nystagmus with the lean
- Bow and Lean test may have superiority over Supine Roll Test

Choung, Y.H. et al. 2006


EXIT

Supine Head- Hanging Test

- strict rotation in the canal plane is of relatively less importance than a final low head down position.
- additional 20° when compared to Dix Hall Pike may be crucial for provoking anterior canal BPPV
- Patient is seated and then brought down supine with head hanging off the table (~50 degrees)

- Betholon et al. 2002

Test Results

- PC – BPPV
  - Upbeating and torsional nystagmus
- AC –BPPV
  - Downbeating and torsional nystagmus (torsional nystagmus is limited) (Bertholon et al. 2002)

- Patient reports of sensation of vertigo
- Vertigo will last usually less than a minute
- Latency to the onset of vertigo and nystagmus
- AC BPPV (fatigues with repeated positioning)
  - Bertholon et al. 2002

Difficulty in clearing of debris

- About 10% PC – BPPV
- % of AC – small proportion (% not expressed)
- due to the conglomerate of debris being too large
- continuous production of debris
- failure of disaggregation
- a narrow common crus.

- Bertholon et al. 2002

Subjective BPPV

- Symptoms of Vertigo without nystagmus
- Other diagnosis should be considered in these patients.
- Treatment failures were only marginally more likely in the remaining 45 patients compared with those with BPPV whose nystagmus was evident on exam.
    - Balatsouras et al. 2012

Treatment for PSCC

- Canalith repositioning Maneuvers
- Epley's Maneuver
- Semont Maneuver

**Epley and Semont's maneuver are both safe and better than sham treatment for BPPV with a similar recovery rate at 1 week (Liu Y et al. 2015)

*** In study by Anagnostou E. et al. 2014 it was noted that no patients had conversion to HC BPPV with Semont's maneuver

**** Repeated Epley and switch to Semont maneuver following a failed initial Epley's shows a similar efficacy in treating PC-BPPV (Oh S.Y etal. 2017)

Could it be central positional nystagmus

- Central positional vertigo and nystagmus may occur with lesions of the cerebellum, particularly the cerebellar vermis.
- The classic sign of central positional vertigo is downbeat nystagmus. In contrast with BPPV, the nystagmus is static and persists as long as the provocative position is maintained.
- n some patients, the downbeat nystagmus is present or increased only when lying down, more so when prone than supine
- Central positional vertigo and nystagmus has been described with both cerebrovascular and demyelinating diseases
- Other possible causes include Chiari malformation, idiopathic cerebellar degeneration, and spinocerebellar ataxia type 6
    - Bertholon, P et al. 2002

Anterior canal BPPV

- It is rare, accounting for only approximately 1 to 2 percent of patients with BPPV
- 75% of cases with downbeat nystagmus are central (Bertholon et al. 2002)
- Song et al (2015) reported that treatment responses is less than for PC BPPV.
- The straight head-hanging maneuver should be carried out in all patients with a history of positional vertigo and a negative Dix-Hallpike manoeuvre (Bertholon et al. 2002)

Deep head hanging maneuver

- Sequential head positioning beginning supine with head hanging 30° dependent with respect to the body
- then supine with head inclined 30° forward
- Bringing patient to sitting with head 30° forward.
  - Bertholon, P. et al. 2002

Treatment for HC-BPPV

- Barbeque Roll
- Gufoni Maneuver
- For Geotropic:
  - Barbeque Roll (69% effective)
  - Gufoni (61%)
  - No difference in outcome between the two after 1 month. ((Kim Jo etal. 2011)
- For Ageotropic/ Apogeotropic
  - Gufoni for Ageotropic
  - Cupolith repositioning maneuver (Kim Jo etal. 2011)
- Forced Prolonged Positioning (Vannuchhi 1997)
  - Used in conjunction with other maneuvers

Vestibular Migraine

Previously used terminology

- migraine-associated vertigo/ dizziness
- migraine-related vestibulopathy
- migrainous vertigo.

- Vestibular Migraine terminology given by Dietrich M. and Brandt T. (1999)

Introduction

- Vestibular migraine (VM) is the most common cause of spontaneous episodic vestibular syndrome with a lifetime prevalence of about 1%
- VM is likely the most common vestibular disorder
- It afflicts predominantly females, with a 5-fold increased risk
- Has a mean age of onset of 38 years in women and 42 in men
- VM accounts for about 10% of patients seen for migraine and about 10% for dizziness
    - (Liu and Xu,2016)

Presentation in Childhood

- Migraine related syndromes are the most common cause of dizziness and vertigo
- ‘‘Benign Paroxysmal Vertigo in Childhood’’ - represents VM with aura but without headache.
- Children : VM is the most frequent form of vertigo (39 %) followed by psychogenic/functional dizziness (21%)

  - Dietrich et al. 2016

BPVC - Benign Paroxysmal Vertigo in Childhood

- Diagnosis*
  - five episodes of severe vertigo
  - occurring without warning
  - Resolves spontaneously
- Brief attacks of vertigo associated with nystagmus
- Age of presentation : first and fourth year of life
- Vertigo last only seconds to minutes
- Symptoms disappear spontaneously within a few years
- It is benign and treatable.
- There are frequent transitions to other forms of migraine with and without aura.

- (Dietrich et al. 2016 ; *LempertTh. 2012)

Symptom Presentation

- Vertigo
- Headaches
- Auditory symptoms
- Dizziness maybe reported as Lightheadedness , Swimming sensation, Giddiness, Heavy-headedness, Rising or sinking , Rocking or swaying
- Motion sickness

- Dieterich and Brandt, 1999

Symptom Presentation

- Duration
- Variable (minutes to hours usually)
- Less than few minutes (<15 percent)
- > 24 hours (in about 25%)
- Motion intolerance and unsteadiness may follow the acute attacks.
- Symptoms rarely persist more than 72 hours.
- Frequency
- Variable presentation
- Few to several times a year
    - (Johnson GD, 1998; Dieterich and Brandt, 1999)

Vertigo:

- *Type*
- internal a false sense of self motion
- External a false sensation that the visual surround is spinning or flowing
- *Character of vertigo* can be
- Rotational
- Swaying
- Tilting
- Falling
- Floating sensation

- (Lempert Th. 2012)

Vertigo

- *Aggravating Factors*:
  - Positional Changes (at times the vertigo is provoked by head position)
  - In the latter case, persistence of vertigo as long as the precipitating head position is maintained BPPV (sensitivity to position change lasting days/ weeks) VM (sensitivity lasting hours)
  - Menstural Cycle
  - visually-induced vertigo: It is usually triggered by a complex or large moving visual stimulus
  - Head motion induced dizziness with nausea.

(Lempart, Th et al. 2012, Dieterich& Brandt, 1999)

Headache

- Headache does not have a temporal relationship.
- Vertigo may occur during or immediately precede the headache.
- No temporal relationship between vertigo and headache.
- Few patients experience vertigo consistently as a typical aura lasting 5 to 60 minutes before headache onset.
- One or more migraine symptoms (photophobia, phonophobia, visual aura) often occur during the episodes of vertigo
    - (Brantberg et al. 2005)

Headache

- Headache:
  - Migraine-type headache
  - Episodic vestibular symptoms without other neurologic symptoms.
- *History of migraine* headaches may precede the development of vestibular migraine, often by several years
- Patient may or may not have headache with their vertigo.
- Very variable presentation (54-94%)
  - Johnson, G.D. 1998

Auditory symptoms

- **Auditory symptoms** – Sensitivity to sound, or phonophobia, occurs in two-thirds of patients.
- Minor symptoms of bilateral or unilateral tinnitus, aural fullness, and subjective hearing impairment (usually during attack)
- visual aura is usually absent
- Nausea and vomiting are frequent, but nonspecific, and may simply be secondary to vertigo.

- (Baloh, RW, 1997)

Interictal Features

- 25 percent prevalence of peripheral vestibular abnormalities is reported
- Non-localizing vestibular signs and signs suggestive of central vestibular dysfunction are present commonly
- Presence of motion sickness in Migraineurs and prominence is higher with vestibular migraine

(Furman etal. 2003)

Vestibular and Oculomotor signs

- Nystagmus
  - Spontaneous
  - Positional
  - gaze-evoked nystagmus
- VOR: abnormal vestibulo-ocular reflex
- Smooth Pursuit impaired pursuit are seen frequently

53 to 66 % of patients have the symptoms

(Furman et al. 2003, Furman et al. 2013)

Examination and Diagnosis

- Diagnostic Criteria established by Bárány Society and ICHD
  - 5 or more vestibular symptoms of moderate or severe intensity
  - Duration of the symptoms usually lasting from 5 min to 72 hoursc
  - Any previous or current history of migraine with or without aura as established by ICHD
  - One or more migranious features with more than 50% vestibular symptoms. (Please (refer to full diagnostic criteria - Headache Classification Committee of the International Head- ache Society. The International Classification of Headache Disorders, 3rd edition. Cephalalgia. 2018;38(1):1-211)

- Criteria for probable vestibular migraine that can be applied when the patient has either a history of migraine or migrainous features
  - Lempert et al. 2012

Differential diagnosis

- Meniere disease
- Migraine with brainstem aura
- Brainstem ischemia
- Benign paroxysmal positional vertigo
- Vestibular paroxysmia
  - refers to very brief (one to several seconds)
  - frequent symptoms (up to several times a day)
  - Imicrovascular compression of the eighth cranial nerve.

Persistent postural-perceptual dizziness

Evaluation

- Assess for BPPV very brief attacks of vertigo – especially positionally provoked
- Audiogram to help differentiate between Meniere's
- Imaging :
  - MRI
  - MRA
- Vestibular laboratory is usually performed by physicians. Role of Vestibular testing is more for differentiating between peripheral vs central condition.

- Robertson, C.E. 2020

Treatment for Vestibular Migraine

- Management of Acute Attacks
- Preventative treatment
- Management for interictal symptoms

Treatment – Acute Attacks

- The efficacy of treatments for vestibular migraine is not well studied (Furman et al. 2013)
- Vestibular suppressants can be used to treat acute attacks of vestibular migraine that last more than 20 to 30 minutes
  - Diazapam
  - Lorazepam
  - Promethazine (Antiemetic)
  - Antihistamines (meclizine, dimenhydrinate)
  - Acute antivertiginous and antiemetic drugs are considered useful for suppressing vestibular symptoms

- Bisdorff, A, 2011

Treatment- Acute Attacks

- Antihistaminic drugs such as dimenhydrinate and meclizine are useful for treating milder episodes of vertigo and for controlling motion sickness.
- Drugs that are effective in treating migraine headache (triptans, NSAIDs) do not work as well for vertigo. They might even be hazardous according to the warnings for aura treatments (ergots).
- Triptans such as sumatriptan retrospective study based on patient records, sumatriptan was found to be efficient when the vestibular symptoms were linked or not linked to the headache
    - Bisdorff, A. 2011

Prophylactic Management

- Watching for Diet:
  - Eating habits,
  - food that may trigger the migraine
- Sleep hygiene
- Exercise

- Bisdorff, A. 2011;

Prophalactic Management-Pharmacological

- Tricyclic antidepressants
- Beta blockers like propranolol, atenolol, metoprolol
- Calcium channel blockers verapamil, diltiazem
- Antiepileptic drugs like gabapentin, topiramate
- Clonazepam (0.25-0.5 mg/day)
- Hormonal treatment for menstrual associated headches (in some isolated cases)
    - Bisdorff A., 2011; Johnson GD, 1998; Reploeg MD et al. 2002

Interictal symptoms and management

- Pharmacotherapy
  - clonazepam
- Non-pharmacologic
  - Physical Therapy

- Furman et al. 2003

Physical therapy and Vestibular Migraine

- *Vestibular rehabilitation for Vestibular Migraine is not so well established
- Physiotherapy seems to be useful for
  - anxiety
  - visual dependence
  - Triggers (visual)
  - loss of confidence in the balance system (not for episodic vertigo) (* Alghadir and Anwer, 2018)
- In study by Whitney et al. 2000. It was noted that there was improvement in physical performance measures and self-perceived abilities after vestibular physical therapy (DHI, ABC and DGI)

Headache and PT intervention

Sugaya etal. 2017

- Measures Used:
  - Headache Impact Test (15) measures the impact of headaches on a patient's life and consists of 6 items.
  - DHI
  - Frequency of Headaches
- Results
- Vestibular rehabilitation contributed to improvement of headache both in patients with VM and patients with dizziness and tension-type headache

PT intervention

(Vitkovic etal. 2013)

- The same degree of improvement was observed in the migraine group regardless of medication regime. (vestibular migraine and vestibular impairment)

- Anxiety and depression is common
  - Anxiety may present as a consequence
  - Presence of anxiety may also exacerbate symptoms (Furman etal. 2013)
- BPPV
- PPPD

Managing Comorbid Conditions

- Managing Comorbid conditions is Important
- BPPV
- Motion Sickness
- PPPD
- Refer patient out for any psychiatric conditions such as Anxiety, depression.

Meniere's Disease

CONTINUED

Introduction

- Initially described by Prosper Meniere in 1861 as a condition with typical symptom triad of hearing loss, tinnitus/ aural pressure and vertigo.
- Meniere disease (MD) occurrence is most common between the ages of 40 and 60 years, symptoms may occur at 20- 40 years.
- MD affects 50- 200 individuals /100,000 adults

- Basura et al. 2020

Diagnosis for MD

- Meniere's Disease is a clinical diagnosis with unilateral ear symptoms lasting for years.
- Further, disease can be remission from months to years
- Diagnosis may not be made at one point of time but may take multiple years until a definite diagnosis is made
- Meniere's Disease has been strictly clinically classified by American Academy of Otolaryngology (1995). With revision in 2 categories by Barany Society in 2015.
    - Basura et al. 2020

CONTINUED

Definite MD

- Two or more 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 vertigo
- Fluctuating aural symptoms (hearing, tinnitus or fullness) in the affected ear
- Not better accounted for by another vestibular diagnosis

Lopez-Escamez et al. 2015

Probable MD

- Two or more episodes of vertigo or dizziness, each lasting 20 minutes to 24 hours.
- Fluctuating aural symptoms (hearing, tinnitus or fullness) in the affected ear
- Not better accounted for by another vestibular diagnosis .

Variability in clinical presentation in patients with definite and probable MD, a full and accurate diagnosis may take many months to attain

Lopez-Escamez et al. 2015

Pathophysiology of MD

- MD is a consequence of overaccumulation of endolymph in the inner ear

- Secondary to inadequate absorption of endolymph by the endolymphatic sac results in filling of the perilymphatic space with endolymph.

- Tabet and Salima, 2017

Clinical Presentation

- Vertigo
  - Usually described as a rotatory spinning or rocking sensation.
  - Spontaneous spells of vertigo, some patients identify dietary triggers, such as excessive consumption of sodium or caffeine
  - Duration of the episodes may last less than 20 minutes or more than 12 hours, but neither is a common finding, and other disorders should be considered when such durations are noted.
    - Lopez-Escamez et al. 2015

Clinical Presentation

- Tinnitus
  - Tinnitus may be fluctuating or constant with variable pitch and intensity.
  - Tinnitus may occur concurrent or independent of auditory symptoms or vertigo
- Aural fullness is reported in most patients and usually pre- cedes the attack of vertigo. (Syed and Aldren, 2012)
- Auditory symptoms are unilateral
  - (Syed and Aldren, 2012)

Differential Diagnosis

- Autoimmune (ie, multiple sclerosis)
- Benign paroxysmal positional vertigo
- Infectious (ie, Lyme
- disease)
- Otosyphilis
- Stroke/TIA/ischemia
- Vestibular migraine
- Vestibular schwannoma
- Labyrinthitis
- Vestibular neuritis

- Basura et al. 2020

Vestibular Migraine and MD

- Clinical features of MD and VM can overlap
- When doing a differential: VM diagnostic criteria should be determined as established by Barany Society.
- Especially in early disease the differential diagnosis may be difficult
- Diagnosis of MD should be made when the characteristic audiometric hearing loss is identified on audiograms, even when migraine features are present.
    - Basura etal. 2020

VM and MD

- These two conditions can occur concurrently ~40% (Shepard 2006)
- Migraine is more common in patients with Menie`re's disease than in healthy controls*
  - (Lempart, T, 2016*)
- MD patients have been found to be twice as likely to have migraine compared to those without MD (Liu and Xu 2016)
- Migraine is the common thread connecting VM and MD.

Decision making

- When *there is uncertainty* about VM or MD, treatment decisions can be difficult but should proceed through noninvasive therapeutic trials prior to any surgical or inner ear ablative interventions.

(Basura, etal. 2020)

Prognosis of MD

- Natural course of MD is typically progressive and fluctuates unpredictably
- Frequency of acute vertigo attacks increases during the first few years and may eventually decline to near complete cessation of vertigo
- Hearing loss may worsen or persist with some patients showing stabilization of hearing over time

- Basura et al. 2020

Clinical Tests

- Audiogram
- Vestibular function
  - VNG with caloric testing
  - video head impulse testing (vHIT)
  - rotary chair
  - cervical and ocular vestibular evoked myogenic potentials (cVEMP and oVEMP)

Clinical Testing

- **Audiometry:** Necessary part of evaluation of MD
  - Low-frequency or combined low- and high-frequency sensorineural hearing loss
  - Over time, the hearing loss "flattens out" with hearing loss in all frequency ranges
  - As a subset of patients will demonstrate MD bilaterally it is important to document hearing loss in both ears to identify a potential onset of the disorder in the contralateral ear
- Vestibular Testing: May be normal early in the course of the disease
  - Over time, abnormal on the affected side.
  - VNG: 65% of patients have unilateral weakness noted on caloric testing (Cordero- Yanza etal. 2017)
    - Basura et al. 2020

Clinical Tests

- vHIT is another vestibular test that uses high-frequency stimulation to assess function of all 6 semicircular canals independently
- Discordant results between vHIT and caloric testing have been observed in multiple studies of patients with MD.
- Can be used to identify a unilateral peripheral hypofunction to help guide further management especially in uncompensated cases
    - Basura et al. 2020

Imaging

- Imaging:
  - According to CPG for MD (Basura et al. 2020):
  - MRI may be offered of the internal auditory canal and posterior fossa in patients with possible Me´nie`re's disease and audiometrically verified asymmetric sensorineural hearing loss.
  - MRI may detect distention of the endolymphatic space in the cochlea and vestibule.
    - Endolymphatic Hydrops is commonly identified in patients with definite MD however this finding is not present in all patients with MD
  - Primary purpose of MRI is to exclude an inner ear or retrocochlear lesion such as
    - vestibular schwannoma
    - other internal auditory canal
    - CPA mass (eg, meningioma)
    - abnormal brain finding (eg, multiple sclerosis, vascular lesion)

CONTINUED

Endolymphatic Hydrops

- Tests for endolymphatic hydrops
  - eCog
  - Sorbitol Test
- Electrocochleography measures the electrical responses of the cochlea and auditory nerve to electrical stimulation.
  - Previously popular technique
  - Have low sensitivity and specificity

- Basura et al. 2020

Endolymphatic Hydrops

- The meta-analysis of all worldwide published temporal bone studies has shown that every single case with definite MD also shows signs of ELH (Gürkov, et al. 2017)
- Endolymphatic hydrops was present in 137 (91.9%) of the definite Ménière disease ears and in 9 (7.0%) of the ears with other vertigo-associated inner ear pathology (Steekelenberg, et al. 2020)
- Hydrops alone is insufficient to cause Ménière's disease, indicating that there must be one or more additional cofactors that cause asymptomatic hydrops to become symptomatic Ménière's disease (Foster, C.A and Breeze, R.E 2013)

VEMP

- Used to assess the function of the otolith organs and their afferent vestibular pathways.
- cVEMPs provide information from the saccule and inferior vestibular nerve
- oVEMPs provide information from the utricle and the superior vestibular nerve
- In some studies cVEMP has been associated as an adjunct measure for vestibular dysfunction in MD
    - Basura et al. 2020

VEMP

- VEMP
  - Cervical VEMP: Cervical VEMP (cVEMP) is an inhibitory sacculocolic reflex test that shows characteristic changes in symptomatic ears of patients with MD
  - May detect early saccular hydrops before the onset of classic Meniere symptoms
  - Ocular VEMP (oVEMP) engages both utricular and saccular afferent nerve fibers and may also be useful in assessment of Meniere patients
  - In addition to diagnosis, VEMP may be useful for monitoring patients for disease progression and to identify the active ear in patients with bilateral disease.
    - Rauch, SD et al. 2004

Treatment Intervention

- Lifestyle changes
- Physical Therapy
- Pharmacological Intervention

Lifestyle Management

- Patients with MD are more vulnerable to dietary and environmental factors
- triggers
- high salt intake
- Caffeine
- alcohol
- Nicotine
- Stress
- monosodium glutamate (MSG)
- allergies (food and environmental)
    - Basura et al. 2020

Vestibular Rehabilitation and MD

- Patients with bilateral MD have a limited ability to compensate for the peripheral vestibular loss and are at an even higher risk of falls
- Primary goal is recovery of function and mitigation of symptoms related to balance disorders.
- unilateral peripheral vestibular hypofunction due to MD with incomplete central vestibular compensation usually experience chronic
- imbalance symptoms, subjective dizziness, postural instability
    - Whitney et al. 2020

VR and Meniere's Disease

- Acute vertiginous Attacks :
- According to Basura et al. 2020 CPG recommends stopping VR for patients with acute vertigo and fluctuating vestibular function from active MD.
- Lack of evidence to use VR to help mitigate the severity or frequency of acute vertigo episodes in patients with MD (Basura et al. 2020)

Vestibular Rehabilitation

Chronic Cases

- Vestibular Rehabilitation should be recommended for interictal symptoms of MD
- VR for patients after ablative surgery
- Chronic imbalance in bilateral MD may benefit from Vestibular Rehabilitation
- virtual reality–based VR combined with diet and medical management improved subjective symptoms based on the DHI and Dizziness Analogue Scale as compared with those treated with diet and medical management alone (Garcia et al. 2013
    - Basura et al. 2020

Pharmacological Intervention

- Vestibular Suppresants
  - Clonazepam
  - Promethazine
  - Betahistine (also for patients with recurrent vertigo)
- Non Ablative :
  - Intratympanic Steroids
  - Positive Pressure therapy
  - Endolymphatic sac surgery
- Ablative
  - Intratympanic gentamycin
  - Vestibular nerve section
  - Labrynthectomy

Basura et al. 2020

Intratympanic Steroids

- For patients who are not responsive to pharmacological intervention intratympanic steroids is suggested.
- It shows greater management of vertigo symptoms than sham/ placebo treatment
- Patient preference over intratympanic gentamycin
  - (Basura et al. 2020)

Endolymphatic surgery

- expose the endolymphatic sac and duct, with the aim to improve drainage of endolymph
- endolymphatic shunt surgery is superior to sham surgery for control of vertigo, nausea, vomiting, and tinnitus
- Intratympanic Gentamycin
  - Aminoglycosides are toxic to the sensory neuroepithelium of the inner ear
  - gentamicin is more vestibulotoxic than cochleotoxic and is the preferred aminoglycoside for intratympanic administration

• Basura et al. 2020

Intratympanic Gentamycin

- Significant reduction in vertigo
- Considered safe and effective treatment intervention
- Caution for bilateral MD due to risk of bilateral vestibular hypofunction development
- No specific dosing protocol , it is on as needed basis
- Hearing loss
- Those receiving Intratympanic gentamycin injection should be recommended VR
    - Basura et al. 2020

Labyrinthectomy

- Recommended for patients with non usable hearing (hearing that is not communicative)
- Used for patients who are resistant to pharmacological intervention and also IT medications
- Hearing Loss and vestibular loss
- Improvement in QOL is reported by patients
    - Basura et al. 2020

Vestibular Nerve Section

- Selective transection of vestibular nerve and not the cochlear nerve.
- Used as opposed to endolymphatic sac surgery for treatment of drop attacks
- Complications from procedure include hearing loss, facial nerve injury, postoperative headache, and risks of craniotomy, such as bleeding, meningitis, and CSF leak
    - Basura et al. 2020

PPPD

- Barany Society and WHO defined a new functional vestibular disorder termed ‘persistent postural-perceptual dizziness’ (PPPD) based *on 3 decades of research on phobic postural vertigo (PPV), space-motion discomfort, visual vertigo, and chronic subjective dizziness*.
- Persistent postural- perceptual dizziness (PPPD), a condition based on the common features of PPV and CSD

Epidemiology

The prevalence of functional dizziness as a primary cause of vestibular symptoms amounts to 10% in neuro-otology centers.

- 50 % rate of psychiatric comorbidity in patients with structural vestibular syndromes
  - Especially higher association with VM, Meniere's Disease , Vestibular Paroxysmia
    - Popkirov, et al. 2017

Definition

- PPPD is a long term functional, and non-vertiginous dizziness with no current organic vestibular lesion as revealed by history, clinical testing, and vestibular investigations. Exacerbation of symptoms can follow upright posture, motion stimuli, or visual stimuli.
    - (Nada et al. 2019)

Clinical Presentation

- Clinical presentation can be dominated with
- Primary Symptoms
  - Unsteadiness
  - Dizziness
  - Hypersensitivity to self motion/ complex visual Stimuli
- Secondary Complications
  - phobic avoidance of provocative situations(stairs roads)
  - functional gait abnormalities
- In typical cases, patients do not experience symptom-free intervals, but rather a transition from acute to chronic symptoms
  - Popkirov etal. 2017

- Precipitating triggers
  - Vestibular Crisis
  - Acute anxiety attack
  - Medical event (such as HI, panic attack, syncope)
- Normal physiologic and behavioral adaptations
  - a shift in sensory integration to favor visual or somatosensory inputs (increased dependency)
  - increased attention to head and body motion (stiffened posture due to dizziness)
  - more cautious ambulation
- Predisposing factors such as anxiety-related personality traits / introverted temperament can delay a return to normal postural and oculomotor control
- Continued use of high-risk strategies to manage routine movements (postural control) and responses to low-demand space and motion stimuli.

- Dieterich et al. 2017

- The diagnosis of PPPD requires all Bárány Society criteria to be fulfilled.
- Unsteadiness and non-spinning vertigo tend to dominate the clinical picture
- Diagnostic Criteria for PPPD has been established by One or more symptoms of dizziness, unsteadiness or non-spinning vertigo on most days for at least 3 months.
  - Symptoms last for prolonged (hours-long) periods
  - of time,but may wax and wane in severity.
  - Symptoms need not be present continuously
  - throughout the entire day.
- Persistent symptoms occur without specific provocation, but are exacerbated by three factors: upright posture, active or passive motion without regard to direction or position, and exposure to moving visual stimuli or complex visual patterns.
  - Staab etal. 2017

Diagnostic Criteria

- The disorder is triggered by events that cause vertigo, unsteadiness, dizziness, or problems with balance, including acute, episodic or chronic vestibular syndromes, other neurological or medical illnesses, and psychological distress.
  - When triggered by an acute or episodic precipi- tant, symptoms settle into the pattern of crite- rion A as the precipitant resolves, but may occur intermittently at first, and then consolidate into a persistent course.
  - When triggered by a chronic precipitant, symptoms may develop slowly at first and worsen gradually.
- Symptoms cause significant distress or functional impairment.
- Symptoms are not better accounted for by another disease or disorder.

- Staab etal. 2017

Differential Diagnosis

- Vestibular Migraine
  - important to distinguish symptoms from the chronic dizziness and unsteadiness due to PPPD, as management is different.
- PPPD can occur with structural, metabolic or psychiatric disorders.
- PPPD has an acute onset – with acute symptoms turning chronic. For insidious illnesses like Degenerative diseases of central nervous system / vestibular labyrinth, patients maybe needed to be monitored over time to complete diagnosis.
  - Papkirov etal. 2017

Differential Diagnosis

- Co-occurrence of comorbid disorders such as presence of structural, metabolic or psychiatric disorders does not preclude the diagnosis of PPPD.
- Historical enquiry should be extended to symptoms of migraine, panic attacks and generalised anxiety, the three conditions that most commonly coexist with PPPD.
- Medications can Consider temporal associations between symptoms of PPPD and initiation or changes in doses of prescription and non-prescription medications.

  - Popkirov, et al. 2017

Differential Diagnosis

- Additional diagnosis of a specific phobia such as fear of dizziness, fear of falling or agoraphobia is warranted in patients with disabling
- PPPD does not include recurrent falls or gait impairment. Appropriate history should be taken to account for falls/ gait impairment to differentially diagnose
    - Popkirov et al. 2017

Posture Control and PPPD

- *functional gait disorders (Popkirov, et al. 2018)
  - Patient education
  - distraction (dual task) techniques
  - Alternative/ exaggerated gaits (e.g., backwards, running, sliding) to help return to a normal gait
- ** Morisod et al.(2018) noted an improvement in posturography following VR in CSD (Abnormal baseline posturography : 79%, Reduced to 33 % after intervention)
- It is noted that patients with functional dizziness (PPV) have increased postural sway during normal stance by co-contracting the flexor and extensor leg muscles (Brandt et. al. 2012)

Visual Dependence and PPPD

- Cousins et al. 2014 investigated a possible link between visual dependence and long-term symptomatic outcomes specifically in patients with well established VN

Treatments for PPPD

- SSRI
- Physical Therapy
- Psychotherapy (Cognitive Behavioral Therapy)

** Treatment starts with education of the patient about the diagnosis.

- *Physiotherapy* to desensitize the vestibular and balance system,
- *Medications* (which may alter the tone of interactions among vestibular, visual and threat systems in the brain)
- *Psychological therapy* (behavioral ‘reprogramming’ to reduce heightened vigi- lance about dizziness
    - Poprikov et al. 2017.

SSRI

- Selective serotonin reuptake inhibitors (SSRI) and serotonin norepinephrine reuptake inhibitors (SNRI) help manage symptoms of dizziness and unsteadiness in majority patients with Chronic Subjective Dizziness (Staab, 2004)
- The success of intervention does not depend upon presence of psychiatric comorbidity (Poprikov et al. 2017)
- Clinical change is usually seen after 8–12 weeks and if effective management is noted medication is continued for atleast a year (Poprikov et al. 2017)

Physical Therapy and PPPD

Habituation training through vestibular Rehabilitation is most successful with people with PPPD (Whitney et al. 2016; Staab 2011)

Paprikov et al. 2017 have recommended supervision of Habituation training from vestibular therapist to ensure success with intervention.

Thompson et al 2015

- Gentle start with exercises and slow increase in intensity is recommended for a long-term clinical benefit
- Aim is to fatigue abnormal reflexive responses to movement tasks and reducing sensitivity to visual stimuli

Psychotherapy and PPPD

- In a study on 39 patients with phobic postural vertigo, CBT along with vestibular rehabilitation exercises had a positive short-term effect that was not sustained at 1-year (Holmberg et al. 2006)
- Combined approach using psychoeducation, cognitive-behavioral therapy, physical therapy, and antidepressant drugs, when needed, may reduce dysfunctional illness behavior and dizziness (Dieterich and Staab, 2017)

Thank you

References and reading List

- See attached PDF