Differential diagnosis of Vertigo and Meneier’s Disease
Terminologies..

Vertigo: (latin ‘verto’: Spinning or whirling movement)

`An illusionary sense that either the environment or one’s own body is moving’

- Light headeness:
  - Blackout
  - Fainting attacks
- Instability/imbalance
  - Unstadiness with stumbling/falling while walking
- Oscillopsia
  - Oscillating vision, objects seem to move back/forth, to jerk
Approach..

✓ History taking
  ▶ Whether balance disorder??
  ▶ If yes: central/peripheral
  ▶ Etiology

✓ Thorough clinical neuro otological examination

✓ Investigations
History taking..

- Does the pt have vertigo?
- What happened the first time?

<table>
<thead>
<tr>
<th>Association</th>
<th>Disease</th>
</tr>
</thead>
<tbody>
<tr>
<td>After head trauma</td>
<td>BPPV</td>
</tr>
<tr>
<td>Straining</td>
<td>Semicircular canal dehiscence, perilymphatic fistula, presyncope</td>
</tr>
<tr>
<td>Salt load</td>
<td>Meniere’s disease</td>
</tr>
<tr>
<td>With menstrual periods in women</td>
<td>Migraine</td>
</tr>
<tr>
<td>Preceding URTI, mumps, herpes zoster oticus</td>
<td>Vestibular neuritis, labyrinthitis</td>
</tr>
<tr>
<td>Duration</td>
<td>Disease</td>
</tr>
<tr>
<td>--------------------------</td>
<td>-------------------------------------------------------------------------</td>
</tr>
<tr>
<td>Seconds</td>
<td>BPPV&lt;br&gt;Vascular compression of VIIIth nerve</td>
</tr>
<tr>
<td>Minutes to hours</td>
<td>Meniere’s disease&lt;br&gt;Migraine associated&lt;br&gt;Acoustic neuroma&lt;br&gt;Otic syphilis&lt;br&gt;Cogan’s disease</td>
</tr>
<tr>
<td>Days to weeks</td>
<td>Vestibular neuritis</td>
</tr>
<tr>
<td>Continuous symptoms</td>
<td>Migraine&lt;br&gt;Psychogenic dizziness&lt;br&gt;Mal de debarquement</td>
</tr>
<tr>
<td>Variable duration</td>
<td>Inner ear fistula&lt;br&gt;Labyrinthine concussion&lt;br&gt;Blast trauma&lt;br&gt;Barotrauma&lt;br&gt;Familial vestibulopathy&lt;br&gt;Superior semicircular canal dehiscence syndrome</td>
</tr>
<tr>
<td>Precipitating factors</td>
<td>Disease</td>
</tr>
<tr>
<td>-------------------------------------------------------------------------------------</td>
<td>---------------------------------------------------</td>
</tr>
<tr>
<td>Head movement</td>
<td>BPPV, vascular loop</td>
</tr>
<tr>
<td>Foods—caffeine, cheese, wine</td>
<td>Migraine</td>
</tr>
<tr>
<td>Stress /lack of sleep</td>
<td></td>
</tr>
<tr>
<td>Fluoroscent lights</td>
<td></td>
</tr>
<tr>
<td>Loud sound, Pressure changes; valsalva, sneezing, coughing</td>
<td>SSC dehiscence</td>
</tr>
<tr>
<td>Alcohol, exercise</td>
<td>Perilymphatic fistula</td>
</tr>
<tr>
<td>Immunosupression (advanced age, stress)</td>
<td>Enlarged vestibular aqueduct</td>
</tr>
<tr>
<td></td>
<td>Episodic ataxias</td>
</tr>
<tr>
<td></td>
<td>Herpes zoster oticus</td>
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<tr>
<td>Associated ear findings</td>
<td>Disease</td>
</tr>
<tr>
<td>-----------------------------------------</td>
<td>-------------------------------------------------------------------------</td>
</tr>
<tr>
<td>Hearing loss</td>
<td>Ménière's disease; perilymphatic fistula; acoustic neuroma; cholesteatoma; otosclerosis; transient ischemic attack or stroke involving anterior inferior cerebellar artery, herpes zoster oticus</td>
</tr>
<tr>
<td>Tinnitus</td>
<td>Meniere’s disease, acute labyrinthitis</td>
</tr>
<tr>
<td>Earache</td>
<td>Acute middle ear diseases, herpes zoster oticus</td>
</tr>
<tr>
<td>Facial weakness</td>
<td>Acoustic neuroma, herpes zoster oticus</td>
</tr>
<tr>
<td>Sweating, dyspnea, palpitations</td>
<td>Panic attacks</td>
</tr>
<tr>
<td></td>
<td>Cardiogenic cause</td>
</tr>
<tr>
<td>Aura, Headache</td>
<td>Migraine</td>
</tr>
<tr>
<td>Neurological signs—limb weakness, numbness</td>
<td>Central mass effect, CVA, multiple sclerosis, CP angle tumour</td>
</tr>
</tbody>
</table>
History taking..

Comorbid illness

- Diabetes mellitus
- Thyroid disease
- Cardiac arrhythmias

Medications:

- Aminoglycosides, cisplatin
- Tranquilisers
- Antiepileptics
- Antihypertensives, diuretics
- Alcohol
- Methotrexate
- Anticoagulants
Clinical examination

Eye movements:

- **Spontaneous nystagmus test**
- **Unilateral vestibular hypofunction**
- **Jerky nystagmus**
- Still head, 1 min with eyes closed room darkened and then again for 1 min with eyes open and looking straight ahead; Frenzel glasses
- Suppressed by visual fixation

Clinical interpretation:

- **Horizontal torsional nystagmus in acute u/l loss of vestibular function**
Positioning testing..

Head movements elicit nystagmus
For posterior semicircular canal BPPV:
- Geotropic torsional nystagmus with the affected ear down
- Delayed onset 2–20 secs, transient 45 secs, a/w vertigo, fatigable
Figure 165-2. Canalith repositioning maneuver for treatment of benign paroxysmal positional vertigo (BPPV) affecting the posterior canal of the right ear. 1. The patient's head is turned to the right at the beginning of the canalith repositioning maneuver. The inset shows the location of the debris near the ampulla of the posterior canal. The diagram of the head in each inset shows the orientation from which the labyrinth is viewed. 2. The patient is brought into the supine position with the head extended below the level of the gurney. The debris falls toward the common crus as the head is moved backward. 3. The head is moved approximately 180 degrees to the left while keeping the neck extended with the head below the level of the gurney. Debris enters the common crus as the head is turned toward the contralateral side. 4. The patient's head is further rotated to the left by rolling onto the left side until the patient is face down. Debris begins to enter the vestibule. 5. The patient is brought back to the upright position. Debris collects in the vestibule.

Log roll over exercises for lateral semicircular canal
Positioning testing

- Lateral semicircular canal BPPV: m.c atypical variant (3–9%)
- Pt lying supine with head inclined 30° forward
- Nystagmus:
  - Geotropic
  - Stronger with the diseased ear undermost
  - Intense vertigo
  - Not delayed onset
  - Not fatigable
## Positional testing.

<table>
<thead>
<tr>
<th></th>
<th>Benign positional nystagmus</th>
<th>Central positional nystagmus</th>
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</thead>
<tbody>
<tr>
<td>Latent period</td>
<td>2–10 secs</td>
<td>None</td>
</tr>
<tr>
<td>Adaptation</td>
<td>Within 30 secs</td>
<td>Persists</td>
</tr>
<tr>
<td>Fatigability</td>
<td>Yes</td>
<td>None</td>
</tr>
<tr>
<td>Vertigo</td>
<td>Yes and severe</td>
<td>Absent / very mild</td>
</tr>
<tr>
<td>Direction</td>
<td>Towards undermost ear</td>
<td>Variable</td>
</tr>
<tr>
<td>Visual fixation</td>
<td>Supression</td>
<td>None</td>
</tr>
</tbody>
</table>
Other bedside tests..

- Changes in the pressure in the inner ear:
  - Valsalva manoeuvre
  - Pneumatic otoscopy
  - Tragal compression
- Head Impulse test
- Head shake Nystagmus
- Untenberger test
- Romberg's test
- Cerebellar signs
Vestibulospinal functions:

- CRANIOCORPOROGRAPHY:
  - Romberg’s test
  - Unterberger/Fukuda stepping test
  - Tandem walking/WOFEC
Romberg’s test:

- Blindfolded, stand erect for 1 minute
- Sway >10cm – abnormal
Kukuda stepping test..

Only input — vestibular system

3 parameters:

- Displacement
- Angular deviation
- Angle of rotation:
  - >30° />45° towards the lesioned side
- Distance of displacement:
  - 0.5m / 1 m

Also positive in asymptomatic pts
**ukuda writing test**

- Sit erect not touching chair back, elbow and shoulder extended, non-used hand on lap
- Only pen touching the paper
- "A B C... top to bottom"
- Once with eyes open and 3 times blindfolded
- A line drawn from the middle of the 1st letter to the last letter and angle compared to the original
- 6 to 9° imbalance in vestibular dysfunction
- >9° diagnosis of vestibular dysfunction
Investigations

Electronystagmography (ENG)

A basic investigation in the management of all patients suffering from vertigo and equilibrium disorder allows to calculate various Nystagmus parameters like slow & fast phase velocity, amplitude, frequency, duration, total number of beats, latency, etc.

Documentary evidence for medico legal purpose, teaching, publication & patient follow up

It only evaluates function of vestibulo-ocular reflex (VOR) (vestibulo-spinal & vestibulo-colic reflex –Craniocorpography & computerized Dynamic posturography )
Principle

- Cornea-retinal potential difference.
- Voltage differences can be recorded for eye movements.
- Electro-occulography to objectively measure eye movements.
Essentially ENG consists of 3 parts:

**Visual–oculomotor evaluation:**
- Three eye movements assessed as part of the ENG: saccades, smooth pursuit, and optokinetic nystagmus.

**Positional-positional testing,**
- Gaze stability, ocular flutter, spontaneous nystagmus, latent nystagmus.

**Vestibulo-oculomotor function,**
- The bithermal caloric test.
Advantages of Electronystagmography

The results of the test are quantified, and there are well-defined normal limits;

Because ENG provides accurate documentation of results, it can be used to follow up the patient with known vestibular disease;

Standardized documentation is helpful in medical-legal and workers' compensation cases;

It is the only test that assesses each ear separately and can provide lesion localizing information.
Limitations

ENG tests only the lateral semicircular canal and provides little information about the status of the posterior or superior semicircular canals, utricle, or saccule.

Relatively insensitive to torsional nystagmus. However, this limitation is easily overcome using VNG.
technique determines eye position by locating the pupil and tracking its center; the internal
iter program plots, measures, and analyzes the eye movement similar to traditional ENG.
ally important in evaluating patients with benign paroxysmal positional vertigo (BPPV).
ystamographic Advantage

- No patient medication only one calibration eliminates the need for any
- Eliminating the cost of accessories
- No special equipment
- Easier to perform technique
- Torsional nystagmus

ystamographic Disadvantage

- More expensive, some patients with significant claustrophobia may not tolerate the
- sensation of confinement
- Patients with ptosis, overlapping eyelashes, or other eye abnormalities may be difficult
Patient in a cage controlling a magnetic field

The patient wears a soft contact lens in which a wire coil is embedded.

Eye movement effects a change in the magnetic field, which is recorded.

Advantages:
- Very high-resolution data for all types of eye movements, including torsional nystagmus.

Disadvantages:
- Slight discomfort to the patient (owing to the lens).
- Very high cost of the equipment.

This procedure has yet to gain widespread acceptance and is rarely used.
The rotational tests...

Test of vestibulo-ocular reflex
Carried out by BARANY
Passive and active
Vestibular autorotational testing (VAT)
computerized dynamic posturography

Developed by Nashner and Black

Potential mechanism for all sensory system evaluation

Planning and monitoring course of vestibular rehabilitation

Suspected malingering, conversion disorder
<table>
<thead>
<tr>
<th></th>
<th>1</th>
<th>2</th>
<th>3</th>
<th>4</th>
<th>5</th>
<th>6</th>
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<tr>
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<td>fixed</td>
<td>fixed</td>
<td>fixed</td>
<td>swayed</td>
<td>swayed</td>
<td>swayed</td>
</tr>
<tr>
<td>2</td>
<td>open,</td>
<td>closed</td>
<td>open,</td>
<td>open,</td>
<td>closed</td>
<td>open,</td>
</tr>
<tr>
<td>3</td>
<td>fixed</td>
<td>fixed</td>
<td>sway-referenced</td>
<td>fixed</td>
<td>fixed</td>
<td>sway-referenced</td>
</tr>
</tbody>
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<thead>
<tr>
<th></th>
<th>2</th>
<th>3</th>
<th>4</th>
<th>5</th>
<th>6</th>
<th>Interpretation</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>N</td>
<td>N</td>
<td>N</td>
<td>Ab</td>
<td>Ab</td>
<td>vestibular dysfunction</td>
</tr>
<tr>
<td>2</td>
<td>N</td>
<td>Ab</td>
<td>N</td>
<td>N</td>
<td>Ab</td>
<td>visual preference</td>
</tr>
<tr>
<td>3</td>
<td>Ab</td>
<td>Ab</td>
<td>N</td>
<td>Ab</td>
<td>Ab</td>
<td>somatosensory and vestibular dysfunction</td>
</tr>
</tbody>
</table>
VEMP test

Testing the vestibulo-collic reflex

Pure tone sounds of 500 Hz at 95–105dB
3–5 stimuli/second

 SCM-tonically contracted

Absent VEMP

- Failure of activation of SCM
- Saccular disorder
- Menier’s disease

Lower threshold in VEMP with CHL–SSC dehiscence
Meniere’s disease
Prosper Meniere -1861 first described the symptom complex
Before 1938, it was used as a generic term for peripheral vertigo

Whites

M:F-1:14

4th-5th decade

B/L-50% within 5 yrs–if second ear involved rapidly

Familial occurrence-10%-20% cases–a/w migraine

Autosomal dominant

a/w specific MHC’s- HLA B8/DR3 Cw7—autoimmune etiology
Pathogenesis..

Hallmark: endolymphatic hydrops

Overaccumulation of endolymph at the expense of perilymphatic space

Inadequate absorption of endolymph by endolymphatic sac—present theory

HPE: perisaccular fibrosis and decreased duct size

Imaging: shorter endolymphatic drainage systems—distance between the posterior SSC and posterior fossa
Rupture of membranous labyrinth
Leakage of K+ rich endolymph in perilymph
High concentration of extracellular K+

Inactivation
Neural conduction in auditory and vestibular neuronal outflow-s/s
Resolution of symptoms
Reinstitution of the membranous restoration of chemical milieu-resolution of s/s
Distortion in inner ear function
multifactorial, common endpoint of variety of injuries/anatomic variables

Unknown cause: Meniere’s disease

Secondary endolymphatic hydrops

- Viral infection: mumps, measles—delayed endolymphatic hydrops
- Ischemia of the endolyphatic sac/inner ear
- Autoimmune: association with HLA-Ab to HSP70
- a/w development of hydrops—acute otitis media, labyrinthitis, congenital inner ear
Clinical presentation..

Typical triad

- Recurring attacks of vertigo (96.2%)
- Tinnitus (91.1%)
- I/L hearing loss (87.7%)

- Cochlear Meniere’s disease
- Vestibular Meniere’s disease
- Recurrent vestibulopathy and atypical Meniere’s disease
Clinical course—highly variable
Cluster of attacks separated by long remissions
Vertigo Ceases spontaneously in 57%–2 yrs, 71%–8.3 yrs
Tumarkin crisis / drop attacks: “feeling of being pushed”/
- Sudden unexplained falls without LOC/associated vertigo
- Acute utriculosaccular dysfunction
- 2–6%
- Clusters and then remits
Lermoyez:
- Tinnitus and hearing loss precede and worsen with the onset of vertigo
Table 165-1 — AAO-HNS Criteria for Meniere’s Disease Diagnosis

**Major Symptoms**

**Vertigo**
- Recurrent, well-defined episodes of spinning or rotation
- Duration from 20 minutes to 24 hours.
- Nystagmus associated with attacks
- Nausea and vomiting during vertigo spells common
- No neurologic symptoms with vertigo

**Deafness**
- Hearing deficits fluctuate
- Sensorineural hearing loss
- Hearing loss progressive, usually unilateral

**Tinnitus**
- Variable, often low-pitched and louder during attacks
- Usually unilateral
- Subjective

**Diagnosis**

**Possible Meniere’s disease**
- Episodic vertigo without hearing loss or
- Sensorineural hearing loss, fluctuating or fixed, with dysequilibrium, but without definite episodes
- Other causes excluded

**Probable Meniere’s disease**
- One definitive episode of vertigo
- Hearing loss documented by audiogram at least once
- Tinnitus or sense of aural fullness in the presumed affected ear
- Other causes excluded

**Definite Meniere’s disease**
- Two or more definitive spontaneous episodes of vertigo lasting at least 20 minutes
- Audiometrically documented hearing loss on at least one occasion
- Tinnitus or sense of aural fullness in the presumed affected ear
- Other causes excluded

**Certain Meniere’s disease**
- Definite Meniere’s disease, plus histopathologic confirmation

Investigations..

ENG

- Reduction in caloric response – 48%–73.5%
- Complete absence – 6–11%

Head thrusting test

- Asymmetry is subtle – 29% pts

Electrocochleography

- SP/AP ratio increases
- 62% pts have elevated ratios
Dehydrating agents

- Urea, glycerol, furosemide
- 10dB or more improvement in at least 2 frequencies
- 12% improvement in speech discrimination scores

VEMP

- Elevated VEMP threshold with flattened tuning
Electrocochleography

Recording of 3 parameters:

- Cochlear microphonics
- Summating potential-complex
- Action potential—auditory nerve

Increase of SP attributed to the basilar membrane
# Differential diagnosis of episodic vertigo

<table>
<thead>
<tr>
<th>Disease</th>
<th>History and examination</th>
<th>Investigations</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>1 Auto Immune Inner Ear disease AIED</strong></td>
<td>B/L rapidly progressive SNHL (monthly intervals), Recurrent vertigo, Ocular inflammation, Depigmentation (VKH), URTI, LRTI (WG), Recurrent thrombosis, spontaneous abortions (APLA)</td>
<td>CBC, DLC, ESR, RF, ANA, Anti dsDNA, APLA Ab, C3, C4, Western Blot Assay for anti HSP-70, RESPONSIVENESS TO CORTICOSTEROIDS</td>
</tr>
<tr>
<td><strong>2 Perilymphatic fistula</strong></td>
<td>Dysequilibrium with nose blowing/lifting heavy wt, Atecedent H/o trauma, ear surgery, HENNBERT’S SIGN TULLIO’S PHENOMENON</td>
<td>Intra-op--- Fluid, B2 transferrin, HRCT</td>
</tr>
<tr>
<td>Disease</td>
<td>History and clinical features</td>
<td>Investigations</td>
</tr>
<tr>
<td>------------------------------</td>
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</tr>
<tr>
<td><strong>3. Migraine</strong></td>
<td>Recurrent characteristic headache—throbbing, pulsatile, unilateral, nausea, vomiting, photophobia, phonophobia, aura, paraesthesias</td>
<td></td>
</tr>
<tr>
<td><strong>4. Otosyphilis</strong></td>
<td>Late otosyphilis—HUTCHINSON’s TRIAD—EXCLUSIVE FEATURE, Hennbert’s and tullio’s phenomenon</td>
<td>VDRL, FTA-AB, MHA-TP</td>
</tr>
<tr>
<td><strong>5. Labyrinthine concussion</strong></td>
<td>H/O trauma, Vegetative symptoms—nausea, vomiting, Vertigo—subsides over days to weeks</td>
<td>PTA—NIHL with 4 KHZ loss MR=imaging</td>
</tr>
<tr>
<td><strong>6. EVAS</strong></td>
<td>Hearing loss since childhood, Progresses with minor head trauma, Vertigo—late onset—adulthood</td>
<td>CT--&gt;1.5mm at midpoint</td>
</tr>
</tbody>
</table>
Treatment options..

Aim

- Stop vertigo, abolish tinnitus and reverse hearing loss
- Spontaneous improvement in 60%-80% cases
- Placebo effect

3 broad options

- Dietary
- Medications
- Surgery
Dietary modifications..

- Salt restriction
- Diuretics
  - Neither has its efficacy confirmed by double-blind placebo controlled studies*
- Carbonic anhydrase inhibitors–Acetazolamide
  - Not more effective than diuretics
Medications..

- **Vasodilators**: strial ischemia
  - Betahistine
- **Symptomatic treatment**:
  - Antihistaminics
  - Anti-emetics
  - Sedatives
  - Anti-depressants
  - Psychiatric treatment
Local overpressure therapy

Rationale use

Energy of the pressure pulses displaces the perilymphatic fluid and stimulates the flow of endolymphatic fluid --- results in a reduction of endolymphatic fluid.

- Meniett device: FDA approval since 2000
  - Complex pulses 20 cm water over 5 min period
  - 3 times daily
  - Ventilation tube placed thru tympanic membrane

Significant decrease in vertigo for the first 3 mths, later similar to placebo.
Intratympanic Injection

- Gentamicin
  - Through tympanostomy tube/through tympanic membrane
  - Vestibulotoxicity high relative to cochleotoxicity
  - 90% complete control of vertigo, 3% SNHL*
- Current regime:
  - Low-dose (16 mg/mL) gentamicin buffered with HCO₃--
  - Injected intratympanically by a 22-gauge fine needle--
  - Through posteroinferior quadrant of tympanic membrane—
  - Total amount 1ml
- Lie in supine position with the effected ear up for 30 minutes
- Encouraged not to swallow
Intratympanic injection..

- Dexamethasone:
  - Addresses the autoimmune component
  - Intractable vertigo
  - Functional hearing left
  - Concentrations ranging from 2-24 mg/ml
  - Repeat every 3 mths
  - Complete resolution in 82% v/s 57% receiving saline*
Surgical treatment..

Indications

- “Intractable vertigo in whom medical therapy has failed”
- Cocleosacculotomy
- Endolymphatic sac surgery
- Ablative surgery
Cochleosacculotomy

- Create a permanent communication to equilibrate endolymphatic and perilymphatic pressures
- Alternative to labyrinthectomy in elderly patients with preexisting hearing loss
Endolymphatic sac surgery
Ablative vestibular surgery

Labyrinthectomy

Recurrent/persisting vertigo with severe to profound SNHL

1. Transcanal
2. Transmastoid – gold standard
Vestibular nerve section:

Selective vestibular nerve section
Take home message

- Identify vertigo!!!
- History taking
- Clinical neurootological examination
- Diagnostic investigations—last resort
Thank you..............