

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 headedness:
  - ▶ Blackout
  - ▶ Fainting attacks
- Instability/imbalance
  - ▶ Unsteadiness 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 ?

Association	Disease
After head trauma	BPPV
Straining	Semicircular canal dehiscence, perilymphatic fistula, presyncope
Salt load	Meniere's disease
With menstrual periods in women	Migraine
Preceding URTI, mumps, herpes zoster oticus	Vestibular neuritis , labyrinthitis



Duration	Disease
Seconds	BPPV Vascular compression of VIIIth nerve
Minutes to hours	Meniere's disease Migraine associated Acoustic neuroma Otic syphilis Cogan's disease
Days to weeks	Vestibular neuritis
Continuous symptoms	Migraine Psychogenic dizziness Mal de debarquement
Variable duration	Inner ear fistula Labyrinthine concussion Blast trauma Barotrauma Familial vestibulopathy Superior semicircular canal dehiscence syndrome

Precipitating factors	Disease
Head movement	BPPV,vascular loop
Foods–caffeine,cheese,wine Stress /lack of sleep Fluoroscent lights	Migraine
Loud sound, Pressure changes:Valsalva,sneezing,coughing	SSC dehiscence Perilymphatic fistula Enlarged vestibular aqueduct
Alcohol,exercise	Episodic ataxias
Immunosupression (advanced age,stress)	Herpes zoster oticus

Associated ear findings	Disease
Hearing loss	Ménière's disease; perilymphatic fistula; acoustic neuroma; cholesteatoma; otosclerosis; transient ischemic attack or stroke involving anterior inferior cerebellar artery, herpes zoster oticus
Tinnitus	Meniere's disease, acute labyrinthitis
Earache	Acute middle ear diseases, herpes zoster oticus
Facial weakness	Acoustic neuroma, herpes zoster oticus
Sweating, dyspnea, palpitations	Panic attacks Cardiogenic cause
Aura, Headache	Migraine
Neurological signs- limb weakness, numbness	Central mass effect, CVA, multiple sclerosis, CP angle tumour

# History taking..

## Comorbid illness

- ❑ Diabetes mellitus
- ❑ Thyroid disease
- ❑ Cardiac arrhythmias

## Medications:

- ▶ Aminoglycosides ,cisplatin
- ▶ Tranquilisers
- ▶ Antiepileptics
- ▶ Antihypertensives,diuretics
- ▶ Alcohol
- ▶ Methotrexate
- ▶ Anticoagulants



# Clinical examination

## Eye movements:

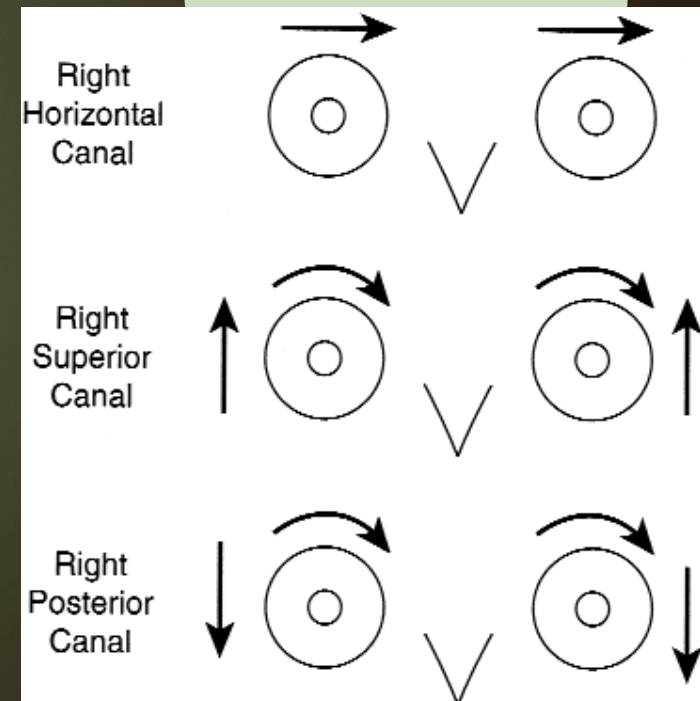
- ▶ **Spontaneous nystagmus test**
  - ▶ Unilateral vestibular hypofunction
    - ▶ Jerky nystagmus
    - ▶ Still head, 1 min with eyes closed room darkened and then again for 1 min with eyes open and looking straight ahead; Frenzel glasses
    - ▶ Supressed by visual fixation
    - ▶ Clinical interpretation:
      - ▶ Horizontal torsional nystagmus in acute u/l loss of vestibular function

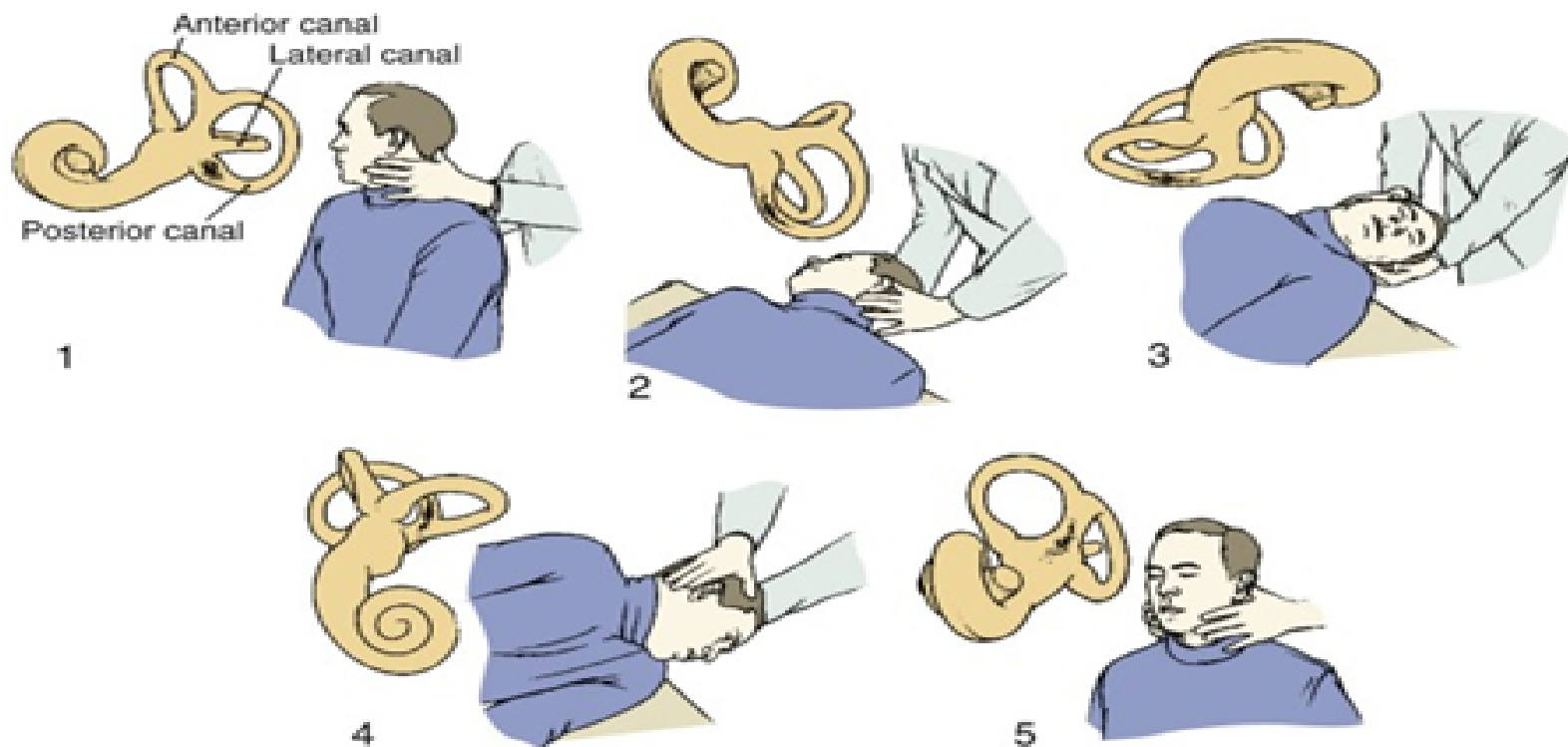
# Positioning testing..

Head movements elicit nystagmus

For posterior semicircular canal BPPV:

- ▶ Geotropic torsional nystagmus with the affected ear down
- ▶ Delayed onset 2-20secs, transient 45 secs, a/w vertigo, fatigable

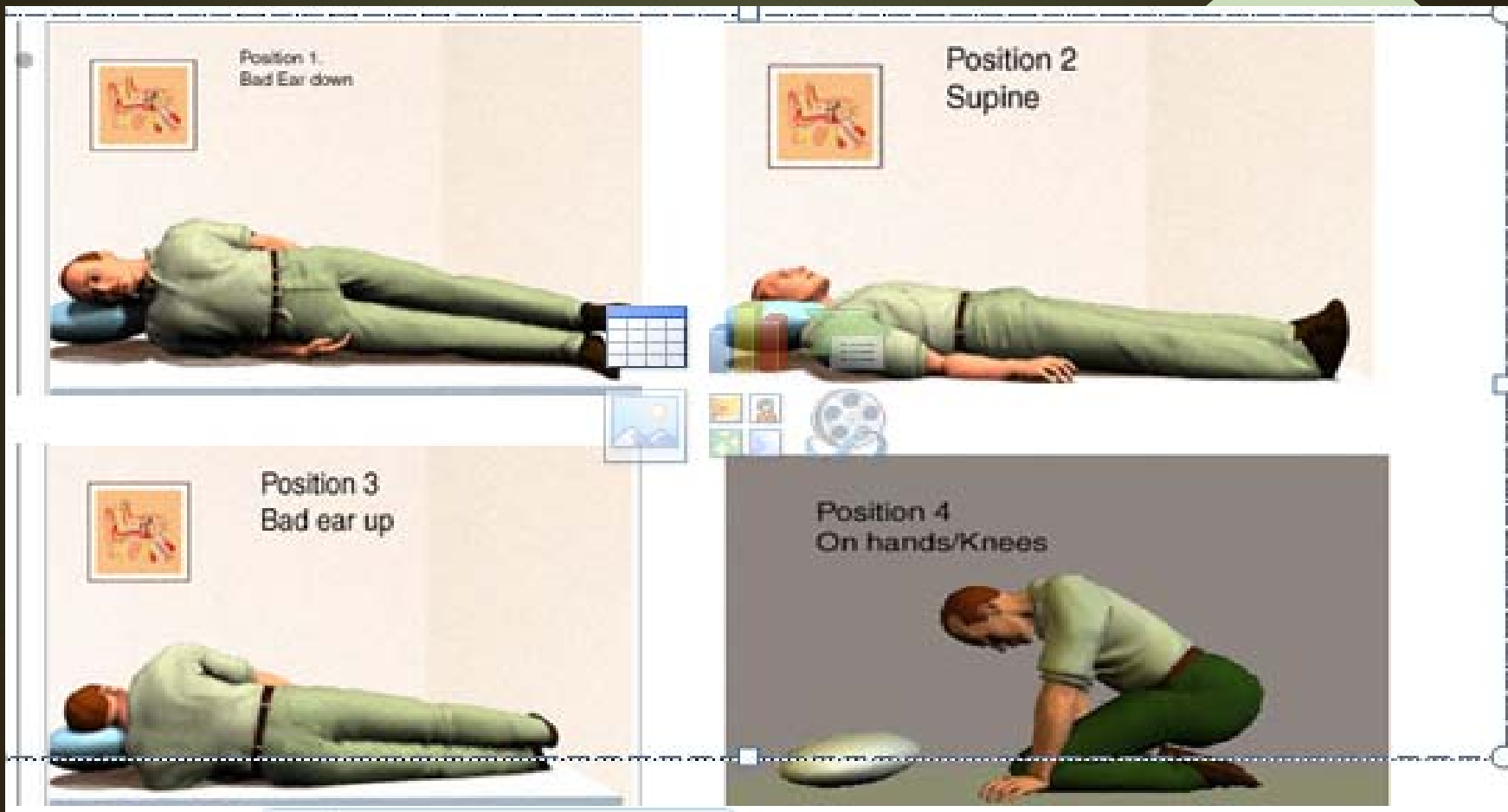




**Figure 165-2.** Canalith repositioning maneuver for treatment of benign paroxysmal positional vertigo (BPPV) affecting the posterior canal of the right ear. **1,** The patient's head is turned to the right at the beginning of the canalith repositioning maneuver. The inset shows the location of the 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.

*(Illustration by David Rini. From Hullar TE, Minor LB: Vestibular physiology and disorders of the labyrinth. In: Glasscock ME, Gulya AJ, eds. Surgery of the Ear. 5th ed. Hamilton, Ontario: B.C. Decker; 2003.)*

# 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..

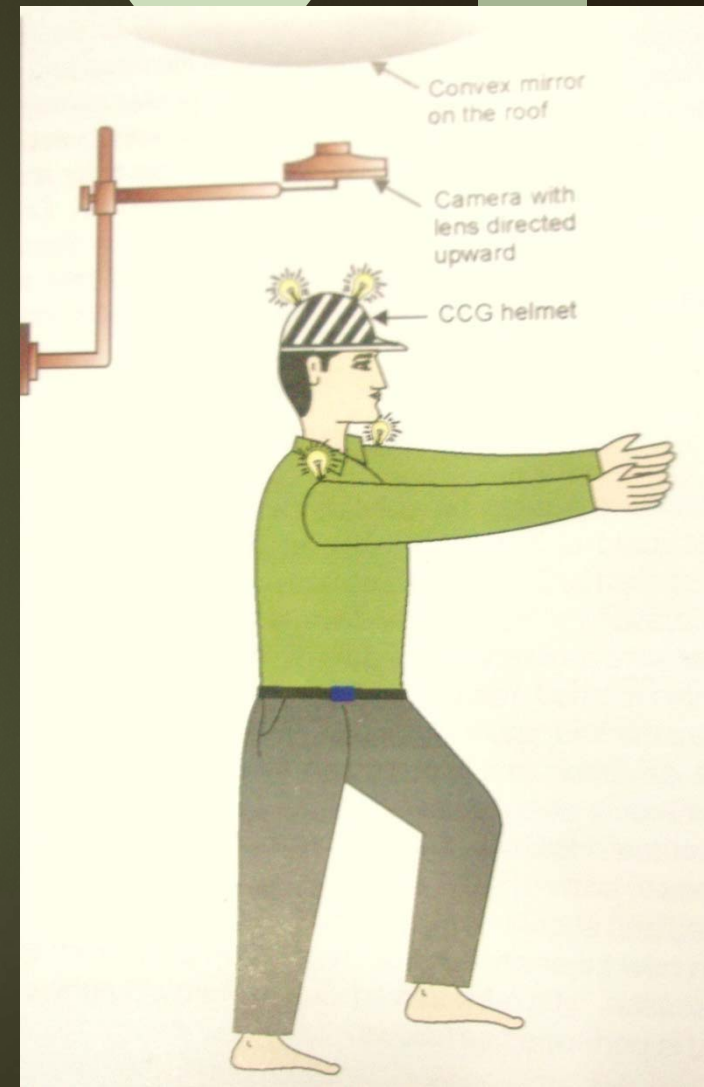
	Benign positional nystagmus	Central positional nystagmus
Latent period	2-10 secs	None
Adaptation	Within 30 secs	Persists
Fatigability	Yes	None
Vertigo	Yes and severe	Absent / very mild
Direction	Towards undermost ear	Variable
Visual fixation	Supression	None

## Other bedside tests..

- Changes in the pressure in the inner ear:
  - ▶ Valsalva manouvere
  - ▶ Pneumatic otoscopy
  - ▶ Tragal compression
- Head Impulse test
- Head shake Nystagmus
- Untenberger test
- Rombergs 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



Figure 7-5 Romberg's test.



Figure 7-6 Sharpened Romberg's test.



# Yokuda stepping test..

Only input –vestibular system

3 parameters:

▶ Displacement

▶ Angular deviation

▶ Step length

▶ Step / 100 step cycle

▶ Feet closed and hands stretched 90 °

▶ >45° hip flexion

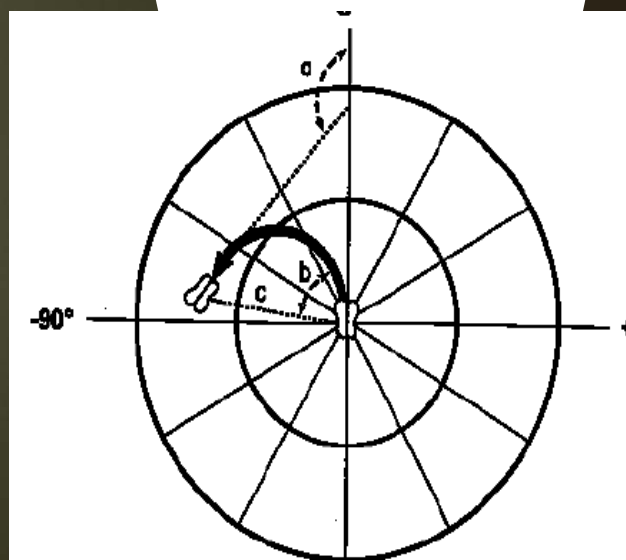
▶ Angle of rotation:

▶ <30° / >45° towards the left / right side

▶ Distance of displacements:

▶ 0.5m / 1 m

▶ Also positive in automatic pts



# Okuda writing test

- ▶ Sit erect not touching chair back , elbow and shoulder extended, non dominant hand on lap
- ▶ Write the word "ABC" 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 1<sup>st</sup> letter to the last letter and angle compared to the original
- ▶ 6 to 9 °- imbalance or vestibular dysfunction
- ▶ >9 °-diagnosis of vestibular dysfunction

# Investigations

## Electronystagmography (ENG)

basic investigation in the management of all patients suffering from vertigo and equilibrium disorder

allows to calculate various Nystagmus parameters

direction, intensity, amplitude, frequency, duration, total duration, etc.

documentary evidence for medico legal purpose, publication & patient follow up

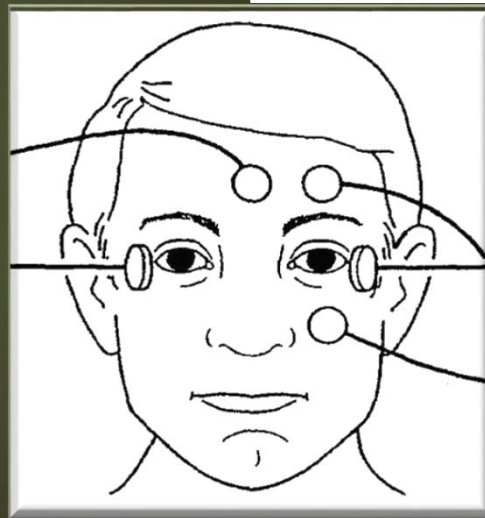
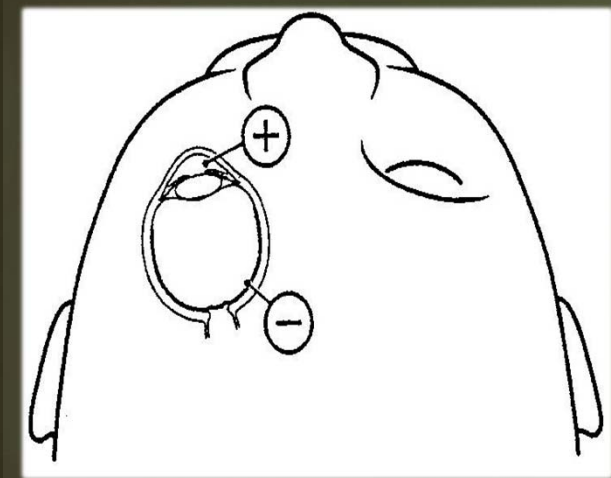
only evaluates function of vestibulo-ocular reflex (VOR)

(vestibulo-spinal & vestibulo-colic reflex -Cranio-corpography & computerized dynamic posturography )



## Principle

- ▶ Cornea-retinal potential difference.
- ▶ Voltage differences can be recorded for eye movements.
- ▶ Electro-oculography to objectively measure eye movements.



essentially ENG consists of 3 parts:

Visual-oculomotor evaluation:

- ▶ Three eye movements assessed as part of the visual-oculomotor test: saccades, smooth pursuit, and optokinetic nystagmus.

Visual-vestibular positional testing,

- ▶ Gaze stability, ocular flutter, spontaneous nystagmus, and 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 is useful to follow up the patient with known vestibular dysfunction;

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

It is the only test that assesses each ear separately and provides 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.

Video-Oculography

Magnetic Search Coil Technique



technique determines eye position by locating the pupil and tracking its center; the internal computer program plots, measures, and analyzes the eye movement similar to traditional ENG.

Especially important in evaluating patients with benign paroxysmal positional vertigo (BPPV)

#### Gonioscopic Advantage

• No contact with no drift

• No skin irritation, no messier than using electrodes

• Only one calibration is necessary

• Reducing the cost of accessories

• No positional nystagmus

#### Gonioscopic Disadvantage

• More expensive, some patients with significant claustrophobia may not tolerate the sensation of confinement

• Patients with prosthesis, protruding 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

► Provides accurate data for all types of eye movements, including torsional nystagmus

► Advantages

► 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

Caloric tests

Vestibular rotational  
testing (VRT)



# Computerized dynamic posturography

Developed by Nashner and Black

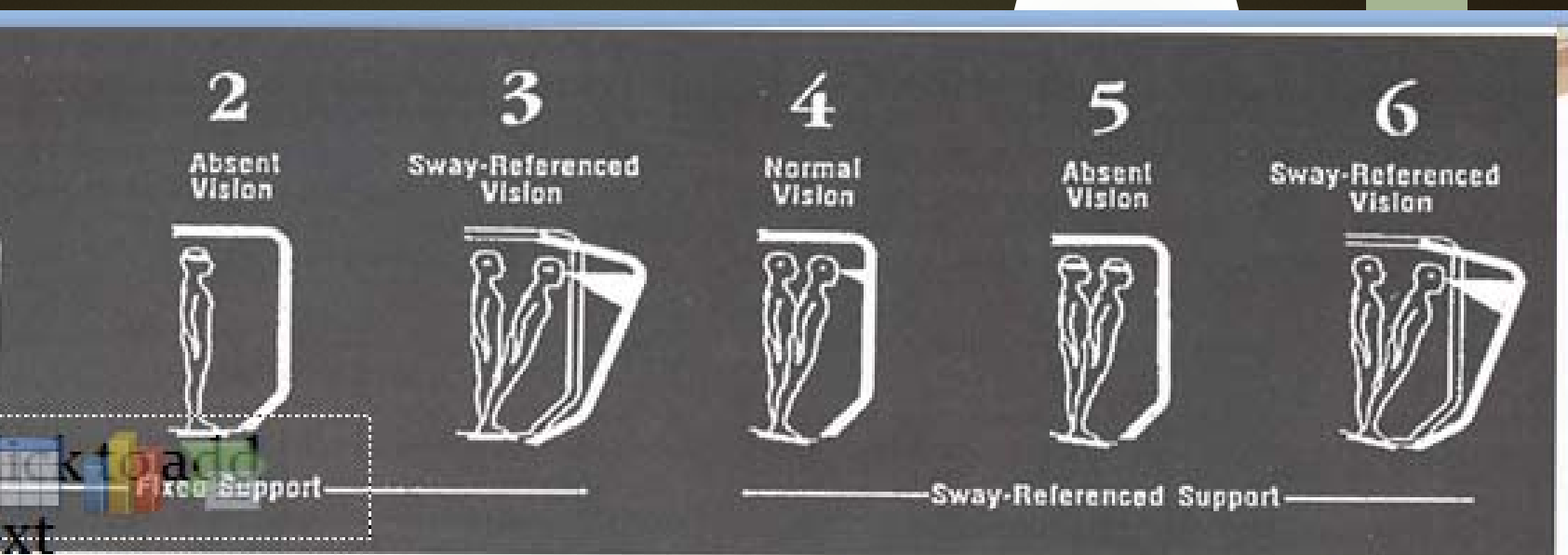
Potential mechanism for all sensory system evaluation

Planning and monitoring course of vestibular rehabilitation

Prevention of lingering, conversion disorder







	1	2	3	4	5	6
Support	fixed	fixed	fixed	swayed	swayed	swayed
Vision	open,	closed	open,	open,	closed	open,
Support Reference	fixed	fixed	sway-referenced	fixed	fixed	sway-referenced

2	3	4	5	6	Interpretation
N	N	N	<u>Ab</u>	<u>Ab</u>	vestibular dysfunction
N	<u>Ab</u>	N	N	<u>Ab</u>	visual preference
Ab	Ab	N	Ab	Ab	somatosensory and vestibular dysfunction

## VEMP test

Testing the vestibulo-collic reflex

Pure tone sounds of 500 Hz at 95–105dB

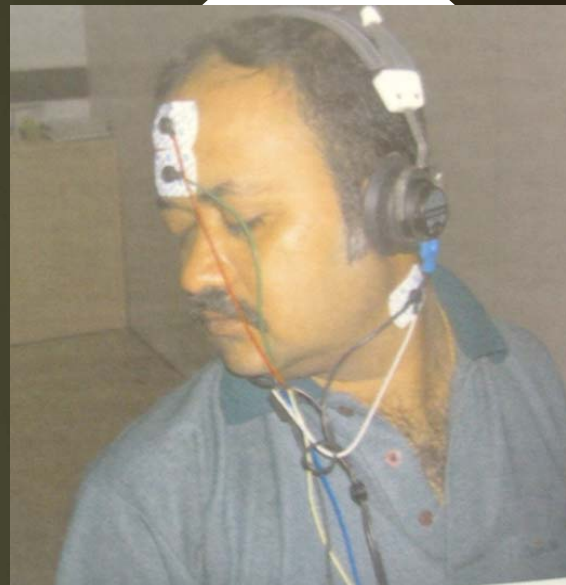
3–5 stimuli/second

Neck muscles voluntarily 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

4<sup>th</sup>-5<sup>th</sup> decade

HL-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 resorption of endolymph by endolymphatic sac theory

PE : perisaccular fibrosis and decreased duct size

Imaging:shorter endolymphatic drainage systems–distance between the posterior SSC and posterior fossa

# Pathophysiology..

Rupture of membranous labyrinth

Leakage of  $K^+$  rich endolymph in perilymph

High concentration of extracellular  $K^+$

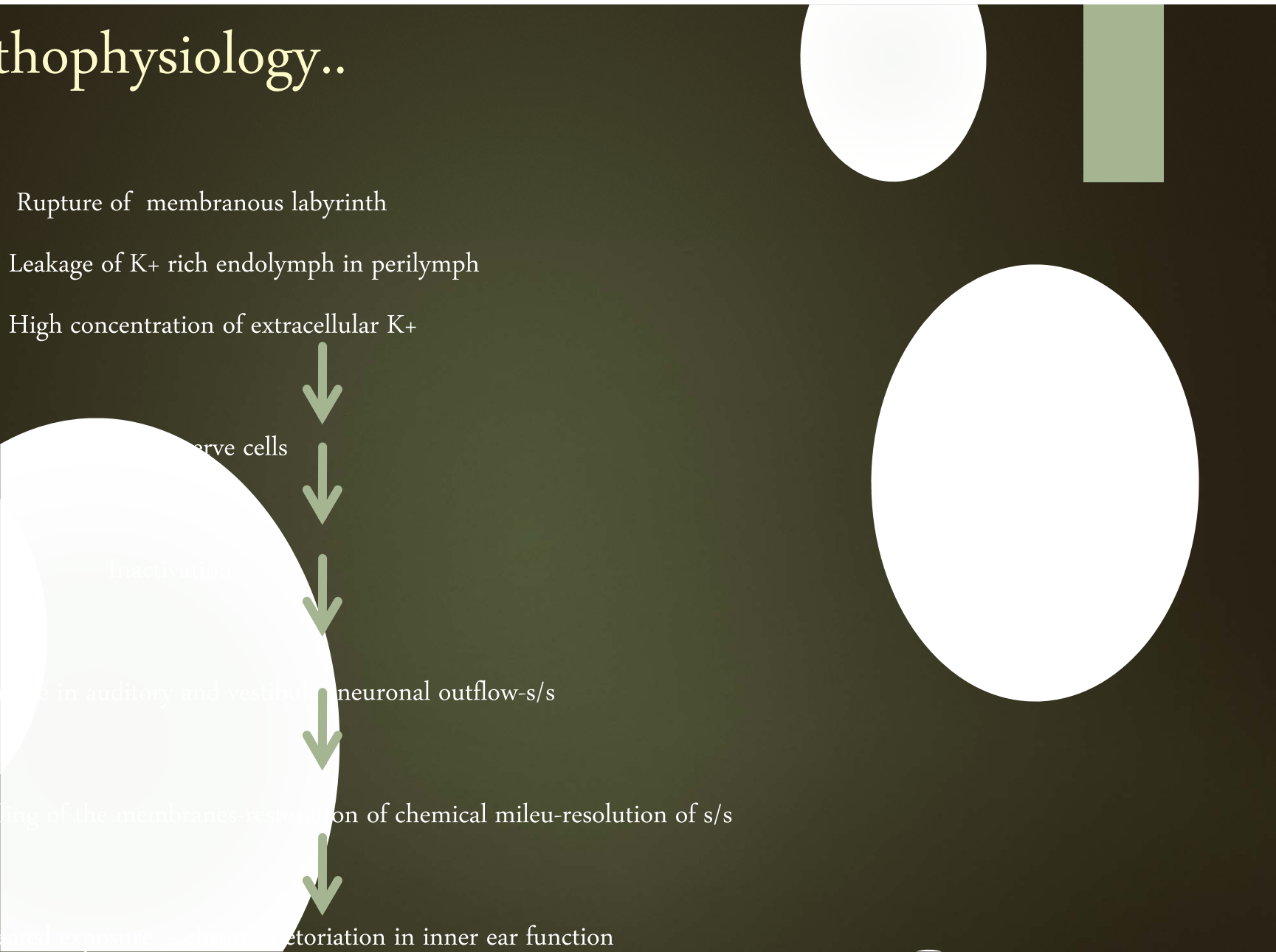
nerve cells

Inactivation

decrease in auditory and vestibular neuronal outflow-s/s

healing of the membranes-restoration of chemical milieu-resolution of s/s

prolonged exposure to elevated  $K^+$  concentration in inner ear function



# etiology..

multifactorial,

common endpoint of variety of injuries/anatomic variations

Primary cause: Meniere's disease

secondary endolymphatic hydrops

- ▶ Viral infection (mumps, measles)-delayed endolymphatic sac
- ▶ Ischemia of the endolymphatic 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%)
- ▶ 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

Tumarkin crisis / drop attacks: “feeling of being pushed” /

▶ Unexplained falls without LOC/associated

▶ Acute bilateral vestibular 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

*From Committee on Hearing and Equilibrium guidelines for the diagnosis and evaluation of therapy in Meniere disease. American Academy of Otolaryngology-Head and Neck Foundation, Inc. Otolaryngol Head Neck Surg*

# Investigations..

## ENG

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

## Lead through test

- ▶ Asymmetry is subtle-29%pts

## Electrocochleography

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

## Dehydrating agents

- ▶ Urea, glycerol, furosemide
- ▶ 10% improvement in hearing
- ▶ 12% improvement in speech discrimination score

## VEMP

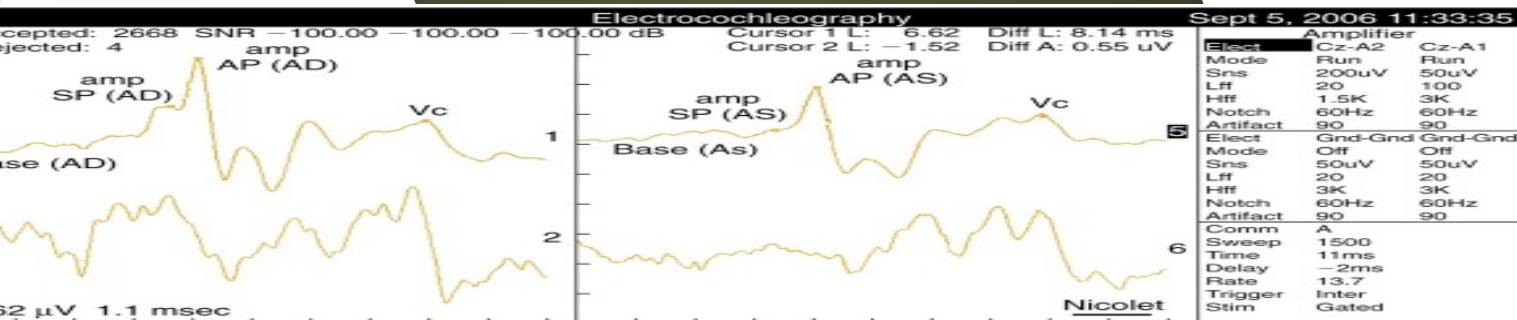
- ▶ Elevated VEMP threshold with flattened tuning

# Electrocochleography

Recording of 3 parameters:

- ▶ Cochlear microphonics
- ▶ Summating potential-complex
- ▶ Compound action potential— auditory nerve

Stimulus-related increase of SP attributed to the basilar membrane



Electrocochleography			
Base (AD)	-1.52ms	Base (AS)	-1.52ms
SP (AD)	1.96ms	SP (AS)	1.70ms
amp	1.01uV	amp	0.24uV
AP (AD)	2.49ms	AP (AS)	2.44ms
amp	2.00uV	amp	1.12uV
Vc	6.71ms	Vc	6.62ms
SP/AP (AD)	0.50 A	SP/AP (As)	0.22 A
Vc-AP-AD	4.22ms	Vc-AP (As)	4.18ms



# Differential diagnosis of episodic vertigo

Disease	History and examination	Investigations
<p><u>1 Auto Immune Inner Ear disease AIED</u></p>	<p>B/L rapidly progressive SNHL,(monthly intervals),                      Recurrent vertigo ,                      Ocular inflammation,                      Depigmentation (VKH),                      URTI,LRTI(WG),                      Recurrent thrombosis,spontaneous abortions(APLA)</p>	<p>CBC,DLC,ESR,RF,ANA,                      Anti dsDNA,APLA                      Ab,C3,C4,Western Blot Assay for anti HSP-70,                      RESPONSIVENESS TO CORTICOSTEROIDS</p>
<p><u>2.Perilymphatic fistula</u></p>	<p>Dysequilibrium with nose blowing/lifting heavy wt,                      Atecedent H/o trauma,ear surgery,                      HENNBERT'S SIGN                      TULLIO'S PHENOMENON</p>	<p>Intra-op--- Fluid,                      B2 transferrin,                      HRCT</p>

Disease	History and clinical features	Investigations
<u>3.Migraine</u>	Recurrent characteristic headache- throbbing,pulsatile,unilateral, nausea,vomiting,photophobia,pho nophobia,aura,parasthesias	
<u>4.Otosyphilis</u>	Late otosyphilis- HUTCHINSON's TRIAD- EXCLUSIVE FEATURE, Hennbert's and tullio's phenomenon	VDRL,FTA-AB,MHA-TP
<u>5.Labyrinthine concussion</u>	H/O trauma Vegetative symptoms- nausea,vomiting Vertigo-subsidies over days to weeks	PTA-NIHL with 4 KHZ loss MR=imaging
<u>6.EVAS</u>	Hearing loss since childhood, Progresses with minor head trauma, Vertigo-late onset-adulthood	CT->1.5mm at midpoint

# 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 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  $\text{HCO}_3^-$  --  
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..

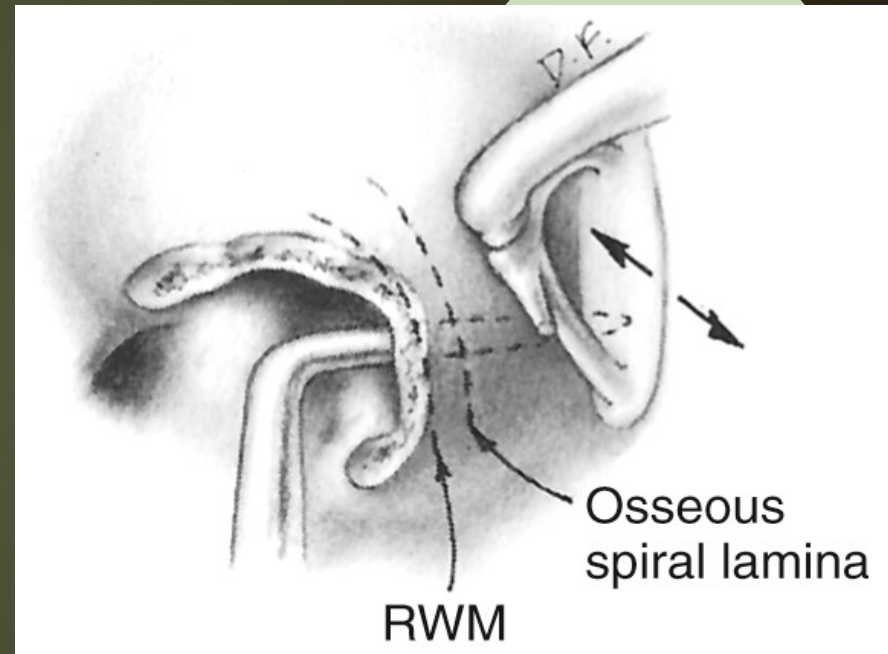
## Indications

▶ “Intractable vertigo in whom medical therapy has failed”

- Cocleosacculotomy
- Endolymphatic sac surgery
- Ablative surgery

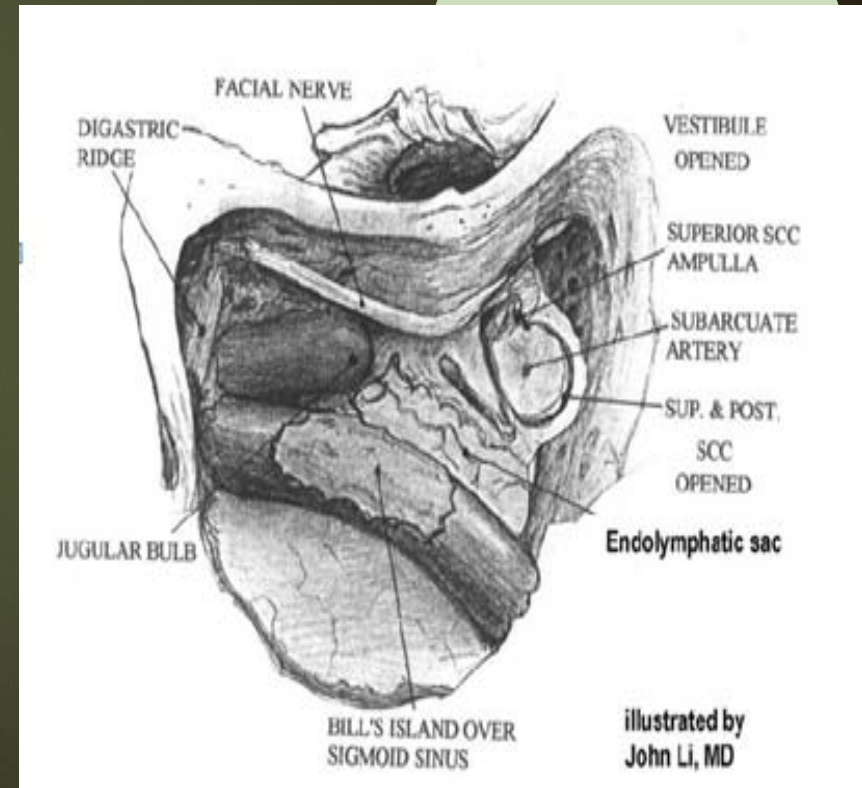
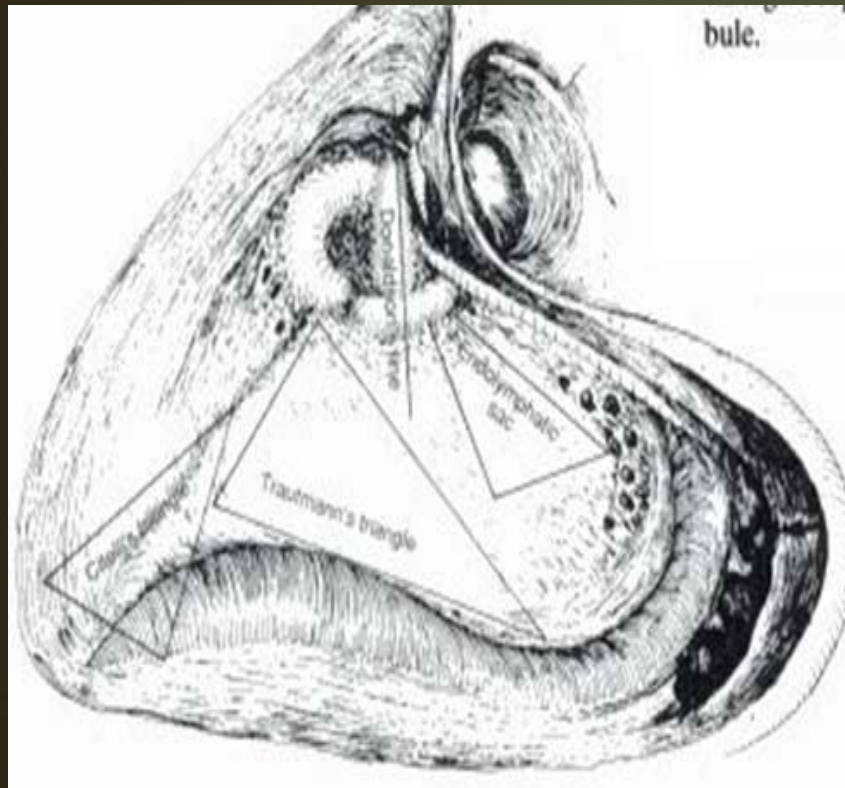
# Cochleosacculotomy

- ▶ 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

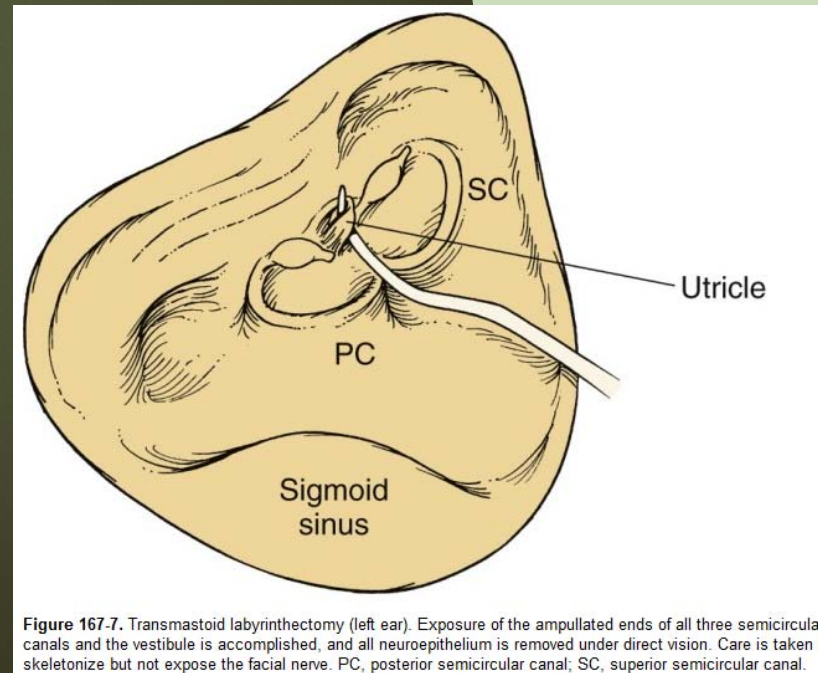
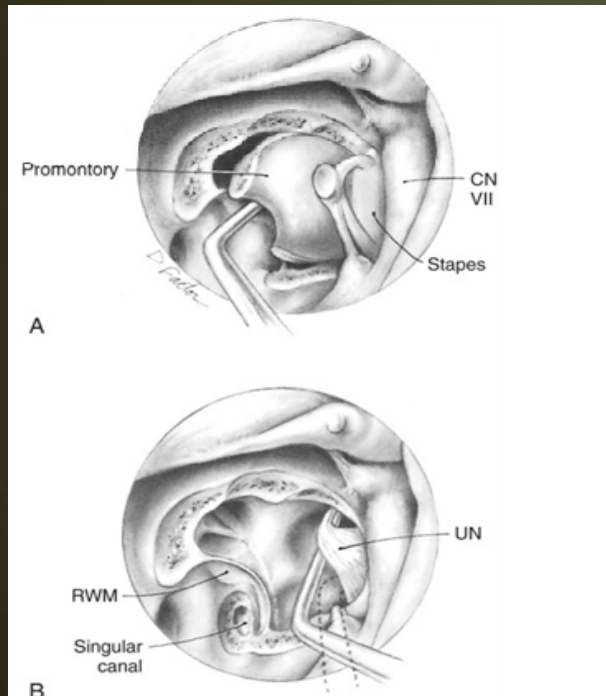
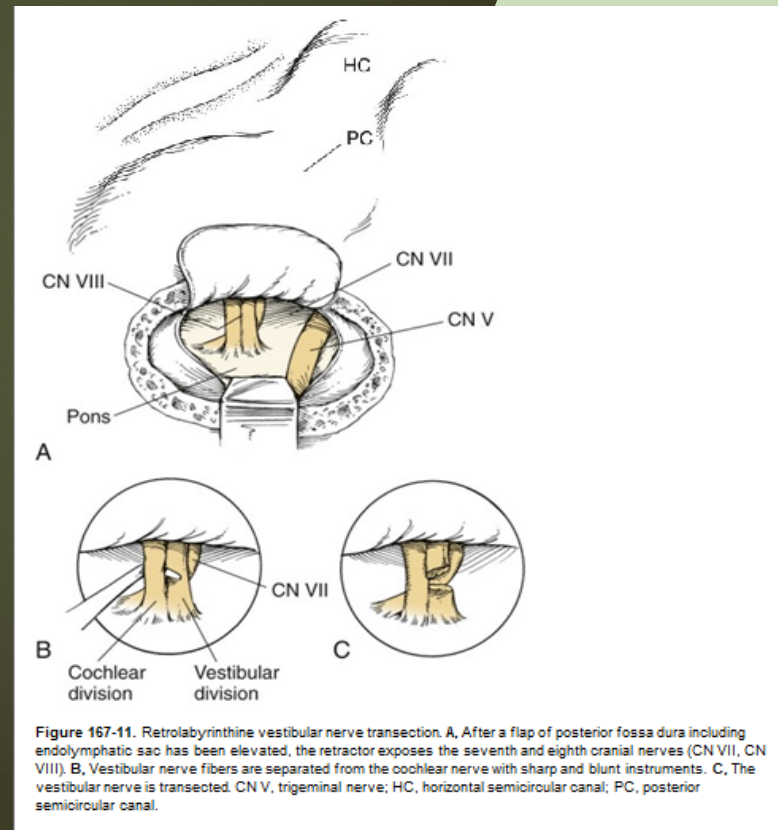
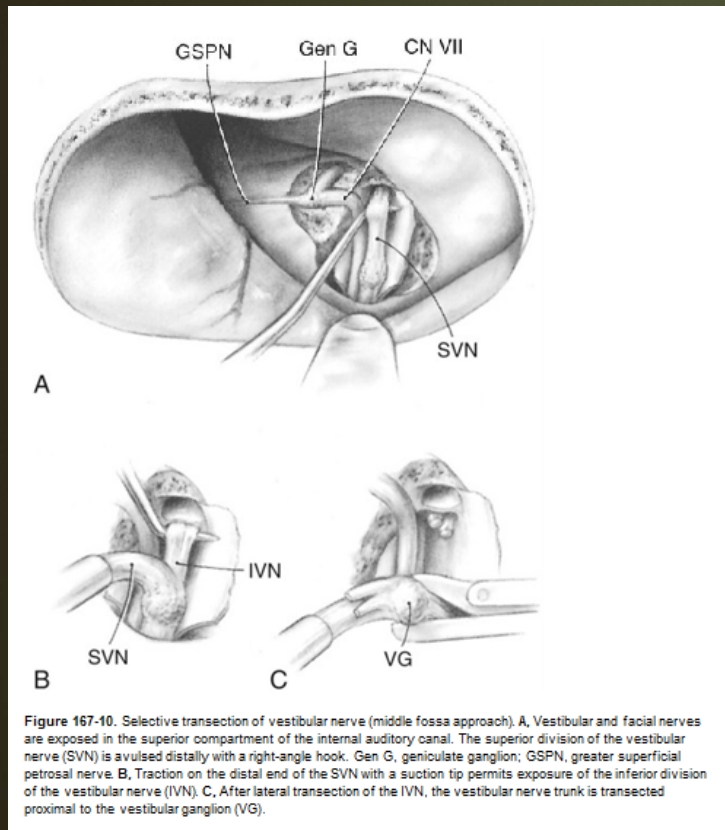


Figure 167.7. Transmastoid labyrinthectomy (left ear). Exposure of the ampullated ends of all three semicircular canals and the vestibule is accomplished, and all neuroepithelium is removed under direct vision. Care is taken to skeletonize but not expose the facial nerve. PC, posterior semicircular canal; SC, superior semicircular canal.



# Vestibular nerve section:

## Selective vestibular nerve section



# Take home message

- Identify vertigo!!!
- History taking
- Clinical neurootological examination
- Diagnostic investigations–last resort



A photograph of a sunset sky with scattered clouds. The sun is low on the horizon, creating a warm orange and yellow glow. In the foreground, there are dark silhouettes of trees on the left and a tall, thin tower or antenna structure on the right. The overall mood is peaceful and contemplative.

*Thank  
you.....*