

PERIPHERAL VESTIBULAR DISORDERS

Epidemiology

- ▣ Peripheral vestibular disorders will affect 1 of 13 people in their lifetime
 - 80% of affected persons seek medical consultation
 - Unclear how many of these are for peripheral vs central disorders
 - Generally: pts younger than 50 are more likely to have **Peripheral disease** vs older than 50 generally have **central dysfunction**
 - In the elderly, dizziness is generally a combination of both
 - After age 75: balance dysfunction = #1 reason for visiting primary care offices

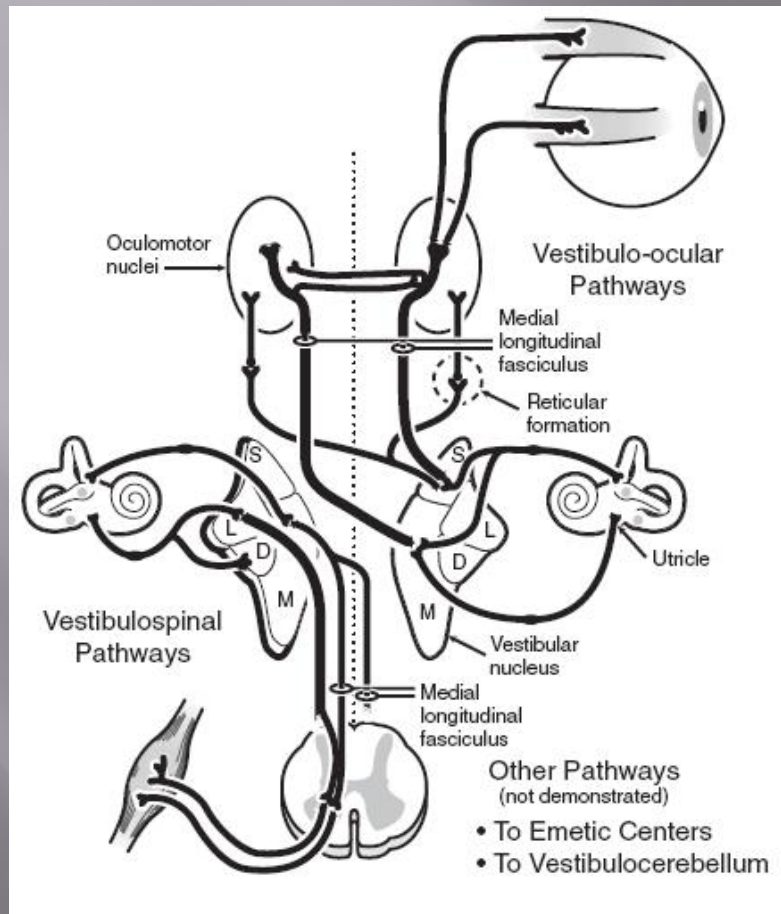
Prosper Meniere

- ▣ 1860s worked at large deaf-mute center in Paris
- ▣ All vertigo thought to be central in origin, he was the first to describe an inner ear origin
 - Published report of girl with sudden hearing loss and acute vertigo after injury → autopsy showed blood in the inner ear
 - Ironic: first pt with Ménière's disease, probably had leukemia, not endolymphatic hydrops
 - Up to 1940s any peripheral vertigo was called Ménière's disease, until true pathophysiology was described

Peripheral Vestibular System

- ▣ Horizontal SCC: horizontal head rotation (yaw)
- ▣ S-SCC & P-SCC: detect pitching of the head front to back as well as roll from side to side
- ▣ Utricle/Saccule: linear acceleration; gravity, and motion in straight trajectory

Normal Function of the Peripheral System



- ▣ Vestibuloocular Reflex (VOR)
 - Physiologic reflex response to a change in head orientation is to move the eyes in an equal and opposite direction
 - → maintain visual stability and focus during head movement

Peripheral Vestibular System

- ▣ All receptors tonically active
 - Resting, spontaneous continual outflow of action potentials
 - With head motion, the spontaneous activity is modulated up or down
 - Ex: turn head to the right → increase R-H-SCC activity and decrease L-H-SCC activity
 - CNS wants balanced input, when input is asymmetric → CNS interprets as head rotation → compensatory eye movements and postural adjustments → sensation of movement

Compensation

- ▣ CNS capable of rebalancing itself
 - Occurs through the ability to compare other sensory, visual, and kinesthetic inputs to vestibular input → readjust when these don't match
 - Will reset the point of balance
 - Typically takes time, on the order of days

Acute Unilateral Disorders

- ▣ Sudden-onset unilateral dysfunction
 - Infectious: viral/bacterial labyrinthitis/ Ramsey-Hunt Syndrome
 - Traumatic: T-bone fracture
 - Iatrogenic: labyrinthectomy/ vestibular nerve section
 - Idiopathic: Meniere's disease
 - Other: ototoxic meds (gentamicin), vascular injury, labyrinthine fistulae

Acute Unilateral Disorders

- ▣ Acute stage: asymmetry interpreted centrally as movement → Vertigo
- ▣ *Oculomotor Nuclei* senses reduced tonic activity → nystagmus
 - If **right** labyrinth is altered: slow eye movement is to the right and the fast phase (nystagmus) is to the **left**
- ▣ *Vestibulospinal* and *cerebellar activity* is altered → sensation of falling
 - Past-pointing, Romberg sign

Acute unilateral dysfunction

▣ Nystagmus

- Immediately after injury it is present in ALL positions of gaze
- ↓↓↓ by visual fixation
- ↑↑↑ when gaze is directed AWAY from injury
- ↓↓↓ when gaze is directed TOWARD injury
- Gradually disappears, however rate of recovery is related to activity level and is decreased with advanced age
- Vestibulo-suppressive meds → ↓↓↓ rate and extent of recovery

Acute Bilateral Vestibular Lesions

- ▣ Common Causes:
 - Ototoxicity, meningitic labyrinthitis, bilateral temporal bone trauma
- ▣ *Symmetric decrease* in tonic activity from each labyrinth to the brainstem
- ▣ If loss is simultaneous and symmetrical → no significant vertigo
 - Instead: pt with poor balance, especially at night; fixed objects jump with any head movement (Oscillopsia)

Acute Bilateral Vestibular Lesions

- ▣ Cerebellar Compensation can adjust the gain of the VOR → typically symptoms resolve within a few days
 - Typically cannot compensate for complete loss → pts with oscillopsia
- ▣ Symptoms: ↓ vestibular sensitivity to head movement and gravitational position → VOR inaccuracy
 - This is necessary for visual acuity during rapid head movement
 - Patients will complain of **visual disturbance** and **lightheadedness** – Blurry vision and disequilibrium

Gradual Unilateral lesions

- ▣ Etiology:
 - CN 8 Neoplasm, degenerative, autoimmune disease
- ▣ PRESENTS WITH:
 - may not produce severe symptoms because of compensation by brainstem
 - Gain of VOR – adjusted by lateral cerebellum
 - With vestibular schwannoma → may be so gradual, that vestibular symptoms imperceptible
 - Pts may not present until lesion is large enough to cause brainstem compression or to compromise blood supply to the peripheral labyrinth → cause sudden unilateral dysfunction
- ▣ TESTING
 - Calorics should still show decreased response on the side of the lesion when tumor affects the superior vestibular nerve

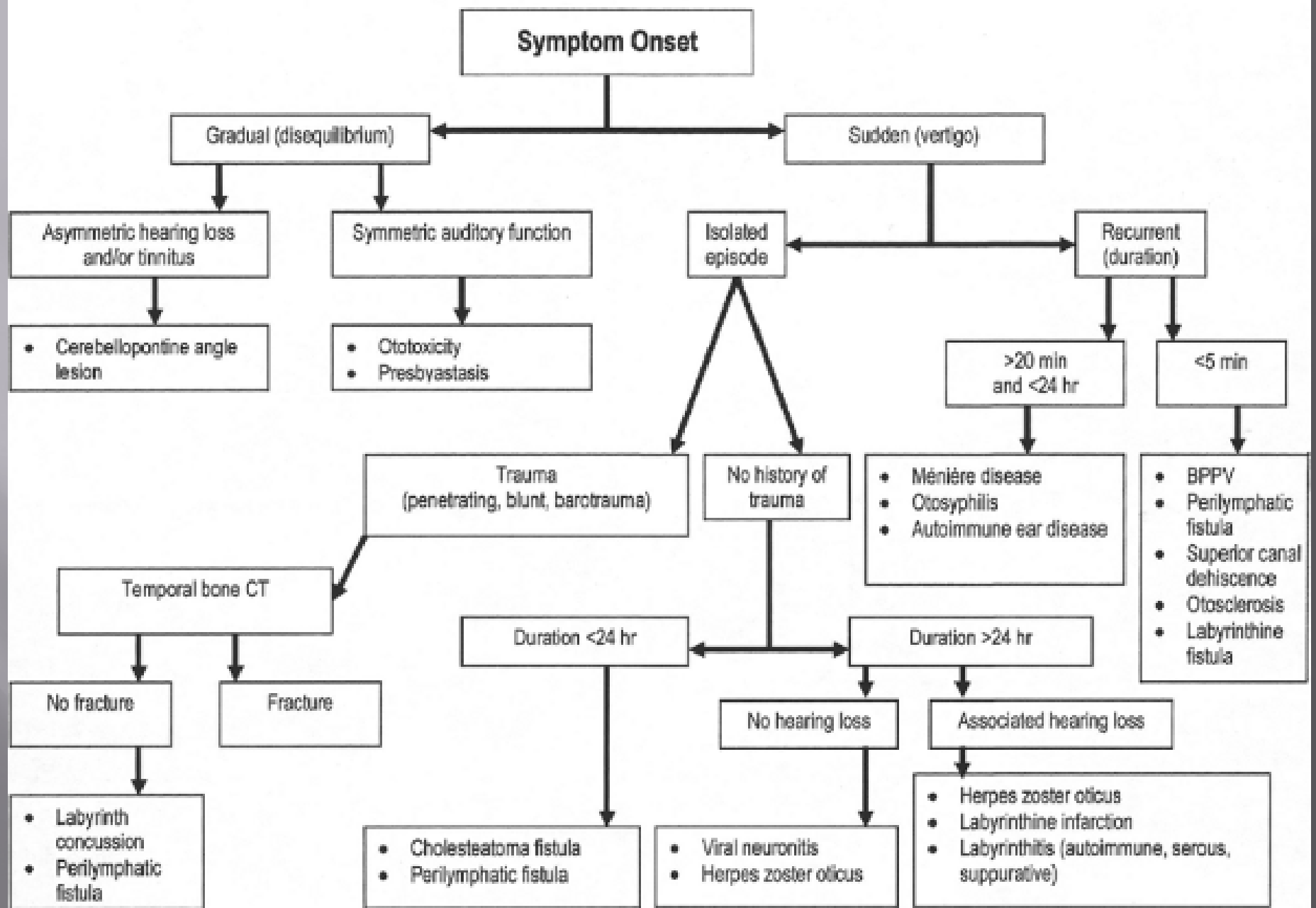
Gradual Bilateral Lesions

▣ ETIOLOGY:

- Aging, ototoxicity, autoimmune disease, syphilis, and degenerative disorders

▣ PRESENTS WITH:

- Cause few symptoms because of cerebellar compensation
- Symptoms when complete or near complete loss of vestibular sensitivity
- Visual disturbance, light-headedness, oscillopsia



Fluctuating or Recurrent Symptoms

- ▣ ETIOLOGY:
 - Hydrops, benign paroxysmal positional vertigo, and dehiscence of the SSC
- ▣ PRESENTS WITH:
 - Intermittent episodes – since they occur episodically, no CNS compensation → each attack is a sudden loss of vestibular function

Diagnosis of peripheral vestibular dysfunction

- ▣ History and physical exam
 - Onset and time course
 - Acute/ progressive/ duration/ frequency
 - Description of symptoms
 - Spinning/ lightheadedness/ imbalance/ blurry vision
 - Associated symptoms
 - Hearing loss/ aural fullness/ otorrhea/ otalgia/ facial paralysis/ headaches/ photophobia/ nausea/ vomiting
 - Precipitating and alleviating factors
 - Head trauma/ Cerebrovascular disease/ autoimmune disease
 - Status of other sensory systems
 - Vision/ proprioception
 - Integrity of CNS
 - Integrity of compensatory mechanisms
 - Medication review

Diagnosis of peripheral vestibular dysfunction

- ▣ Physical Exam
 - H&N exam; CN assessment; Observe gait and posture
 - Oculomotor evaluation w or w/o Frenzel glasses
 - **Tullio phenomenon:** sound-induced vertigo, dizziness, nausea or nystagmus; → SSC dehiscence syndrome
 - Otologic exam: pneumatic otoscopy and audiometry
 - **Hennebert Sign:** vertigo and abn eye movement with positive or negative pressure → SSC DS
 - Spontaneous nystagmus described
 - Type, degree, effect of visual fixation
 - Positional nystagmus and associated vertigo assessed with Dix-Hallpike
 - Head-Thrust test: move head to one side while pt maintains visual fixation → if pt with unilateral vestibular weakness → re-fixation saccade after the head thrust
 - Dynamic Visual Acuity Test:
 - Read Snellen chart with head stationary and then during 2Hz head oscillation → will detect bilateral vestibular loss with loss of 3 lines of acuity

Frenzel Goggles



Combination of magnifying glasses (+20 lenses) and a lighting system – help to detect subtle nystagmus by magnifying the eyes and removes fixation

Diagnosis of peripheral vestibular dysfunction

- ▣ Vestibular Function Tests
- ▣ Electronystagmography (ENG): battery of tests for vestibular function
 - Evaluate spontaneous and gaze-evoked nystagmus with and without fixation
 - Positional testing
 - Caloric response of the lateral SCC
 - Rotational chair testing
- ▣ Eye Movement Studies:
 - Smooth pursuit and optokinetic, saccade and vestibuloocular reflexes
- ▣ VEMP

VEMP

- ▣ Vestibular Evoked Myogenic Potential
 - Assesses function of structures not tested in other tests → the Saccule and Inferior Vestibular Nerve
 - Acoustic signal stimulates the saccule → reflex arc spans the inferior vestibular nerve → vestibular nuclei → vestibulospinal tract with output → SCM Muscle

- ▣ Application:
 - Evaluation of superior canal dehiscence, vestibular Schwannoma, early detection of Menieres disease

IMAGING

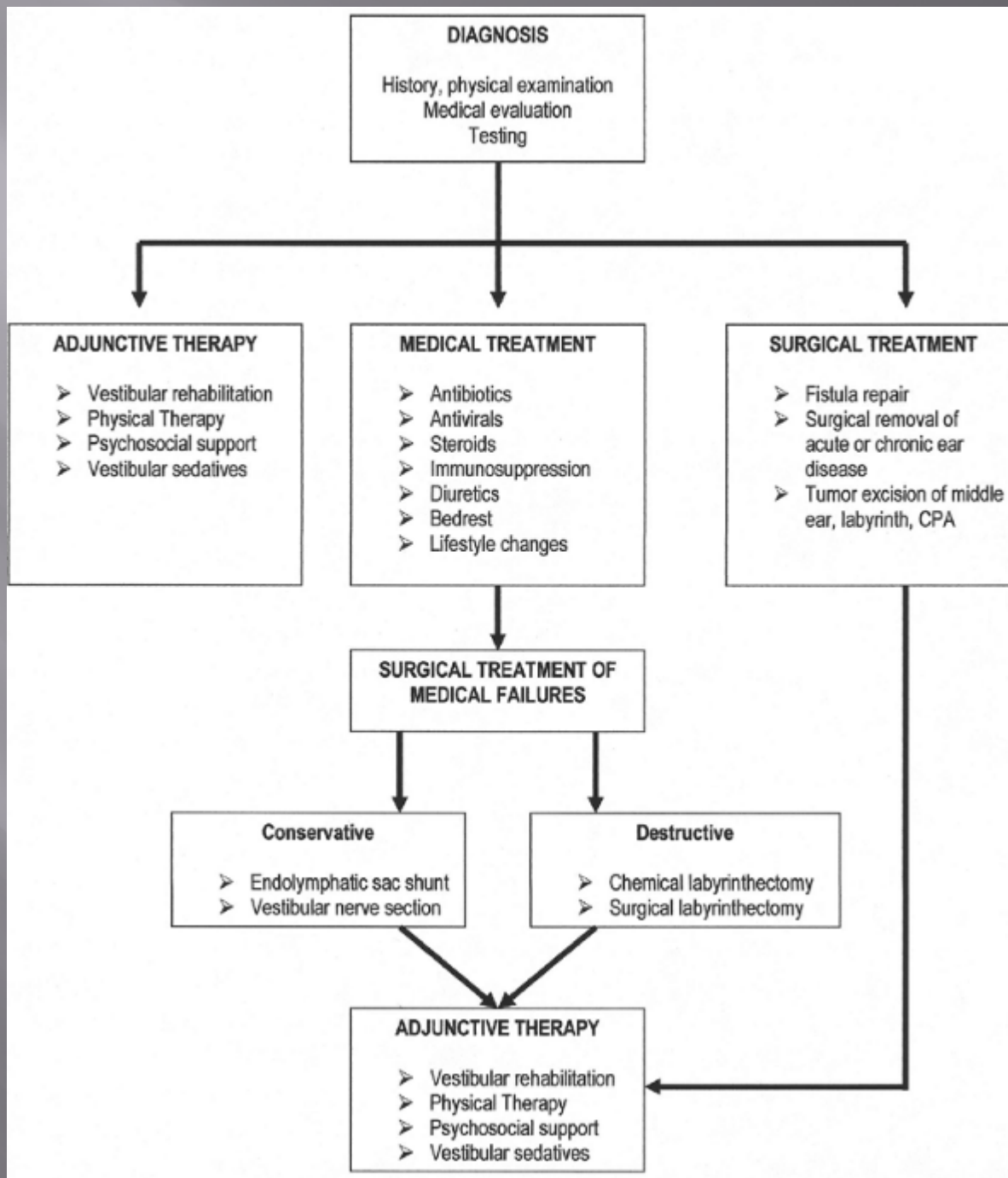
- ▣ If clinical evaluation and ENG suggest middle ear, mastoid, or IAC lesions → consider MRI/CT

TREATMENT

- ▣ GOAL: decrease severity of symptoms and restore function
 - Vestibulo-suppressive medication
 - Ablative surgery
 - Chemical labyrinthectomy
 - Rehabilitation

Sudden unilateral dysfunction

- ▣ Initial management
 - Diazepam 5-10 mg IV over several minutes with reduce severe vertigo 2/2 unilateral loss;
 - USES: vestibular neuronitis, post-traumatic loss, labyrinthitis, severe episodic vertigo
- ▣ After Severe symptoms resolved
 - Movement exercises to rehab remaining function
 - GOAL: promote compensation by altering gain of VOR (Adaptation) and develop substitution strategies to maintain balance
 - Critical Period of 72 hrs: after which rehab is less efficacious and complete recover less likely
 - Reduce doses of vestibulo-suppressive meds to minimize suppression



Ménière's Disease

- ▣ Idiopathic endolymphatic hydrops
- ▣ Episodic vertigo + fluctuating SNHL + Tinnitus + Aural fullness
- ▣ Pathologic Basis: distortion of membranous labyrinth, 2/2 over-accumulation of endolymph from a dysfunction in absorption of the endolymph by the endolymphatic sac
 - Histologic support of this theory

Ménière'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

Ménière's disease - Diagnosis

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

Ménière's Disease

- ▣ Natural history: 60-80% of pts with spontaneous get better — this has made studying treatment difficult
- ▣ Diet Modification and Diuretics
 - Most respond to decrease in dietary salt intake and administration of diuretics
 - Thought to reduce endolymphatic pressure by decrease in volume
 - Never been confirmed by placebo controlled study, but considered first line
- ▣ If still not controlled, or end-stage disease → application of intra-tympanic gentamicin
 - Dexamethasone or gentamicin → Chemical Labyrinthectomy
 - Both carry a risk of hearing loss

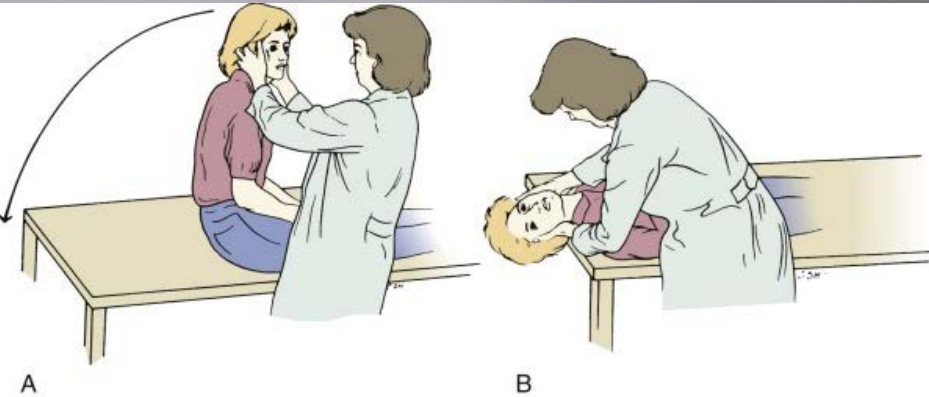
Ménière's Disease - Treatment

- ▣ Surgical Intervention
 - Endolyphatic Sac Surgery:
 - ▣ double-blind placebo controlled study by Thomsen showed mastoidectomy alone has same efficacy; controversial
 - Vestibular neurectomy:
 - ▣ vertigo control in 80-90%, more invasive and technically challenging
 - Labyrinthectomy:
 - ▣ Most destructive procedure, uniform destruction of hearing and vestibular function;
 - ▣ Ideal candidate – no hearing and failed all conservative therapy
 - ▣ High success rate, however used with caution in elderly – poor central compensation
- ▣ Systemic Streptomycin: Chemical Labryinthectomy
 - Used for pts with bilateral Ménière's disease, only hearing ear, or poor surgical candidates
 - Streptomycin given IM BID with daily caloric testing and continued until caloric responses decrease

Benign Paroxysmal Positional Vertigo

- ▣ Due to motion of material in the lumen of the posterior SCC
- ▣ Most common cause of vertigo – 20-40% of cases seen by an otolaryngologist
- ▣ History: severe vertigo with change in head position, especially rolling over or getting out of bed
 - Most cases no etiologic disorder; Baloh published series and half patients had no identified cause, the other half, most commonly had history of head injury or vestibular neuritis
- ▣ Diagnosis: History + Dix-Hallpike maneuver

Dix Hallpike



- 1. nystagmus is combined vertical up-beating and rotary (torsional) toward the downward eye; Pure vertical nystagmus is NOT BPPB
- 2. latency of a few seconds is common
- 3. Duration of nystagmus is < 1min
- 4. Vertiginous symptoms are associated
- 5. disappears with repeated testing (fatigable)
- 6. Symptoms recur with nystagmus in the opposite direction on return of head to upright position

Eliciting nystagmus and vertigo due to posterior canal benign paroxysmal positional vertigo (BPPV).

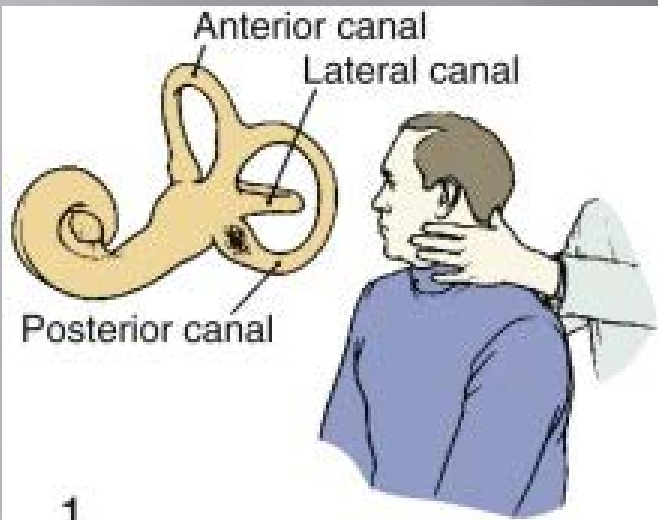
The patient's head is first turned 45 degrees to the right (A).

The patient's neck and shoulders are then brought into the supine position with the neck extended below the level of the examination table (B).

The patient is observed for nystagmus and assessed for symptoms of vertigo. The patient is next returned to the upright position.

Benign Paroxysmal Positional Vertigo

- ▣ signs and symptoms of cupular deflection when otoconial debris either adhere to the crista (cupulolithiasis) or become free-floating within a SSC (canalithiasis)
- ▣ Posterior SCC is most commonly involved
- ▣ Treatment:
 - Particle repositioning maneuvers are highly successful → Epley (next slide)
 - Surgery for incapacitating vertigo →
 - ▣ occlusion of the P-SCC with bone dust (97 cases within the literature → 94 cured)
 - ▣ sectioning the posterior canal ampullary nerve is usually effective → series by Gacek of 340 pts → 40% hearing loss

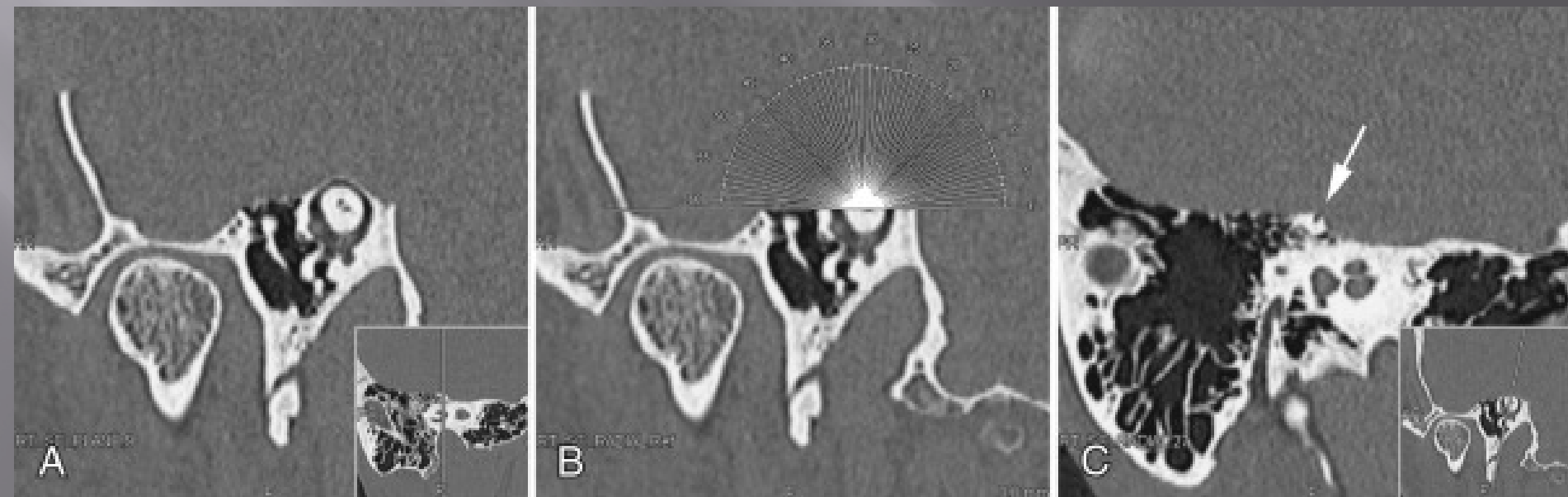


Vestibular Neuritis

- ▣ Dramatic sudden onset of vertigo, lasts for days, with gradual improvement
 - NOT associated with subjective change in hearing or any other focal neurologic complaint
 - Cause is almost never identified, neurotropic viruses, such as herpes or Borellia have been associated
- ▣ Treatment
 - Supportive and symptomatic
 - Recent placebo-controlled, double blind study with Methylprednisolone and Valacyclovir: found significantly improved 1 year symptoms with steroids, no effect of the valacyclovir

Superior Canal Dehiscence Syndrome

- ▣ Absence of bone over the superior canal → 3rd mobile window that allows abn movement of endolymph
- ▣ Expose dehiscent canal via middle cranial fossa approach → occlude with bone dust or resurfaced using fascia and bone



Cogan Syndrome

- ▣ Interstitial keratitis, low-frequency unilateral HL, and vestibular symptoms (nonreactive syphilis testing)
- ▣ Ocular and inner ear changes typically occur within 6 months of each other
 - Vertigo: Simialar to Ménière's
 - Hearing loss: SNHL
- ▣ Etiology: thought to be autoimmune, responds to steroids
- ▣ Clinically: pt will report history of URI within 10 days of symptoms
- ▣ Treatment: systemic corticosteroids, cyclophosphamide and methotrexate

Vestibular Rehab

- ▣ Is highly effective, and can improve symptoms of vertigo of unknown causation and improve general balance conditioning in the elderly
- ▣ Can include therapy at home, or under supervision of a physical therapist
- ▣ Techniques:
 - Vestibular adaptation is responsive for long-term changes in vestibular input
 - Promote altered gain for VOR and VSR by active head and body movement combined with visual input