

# ***Meniere's***



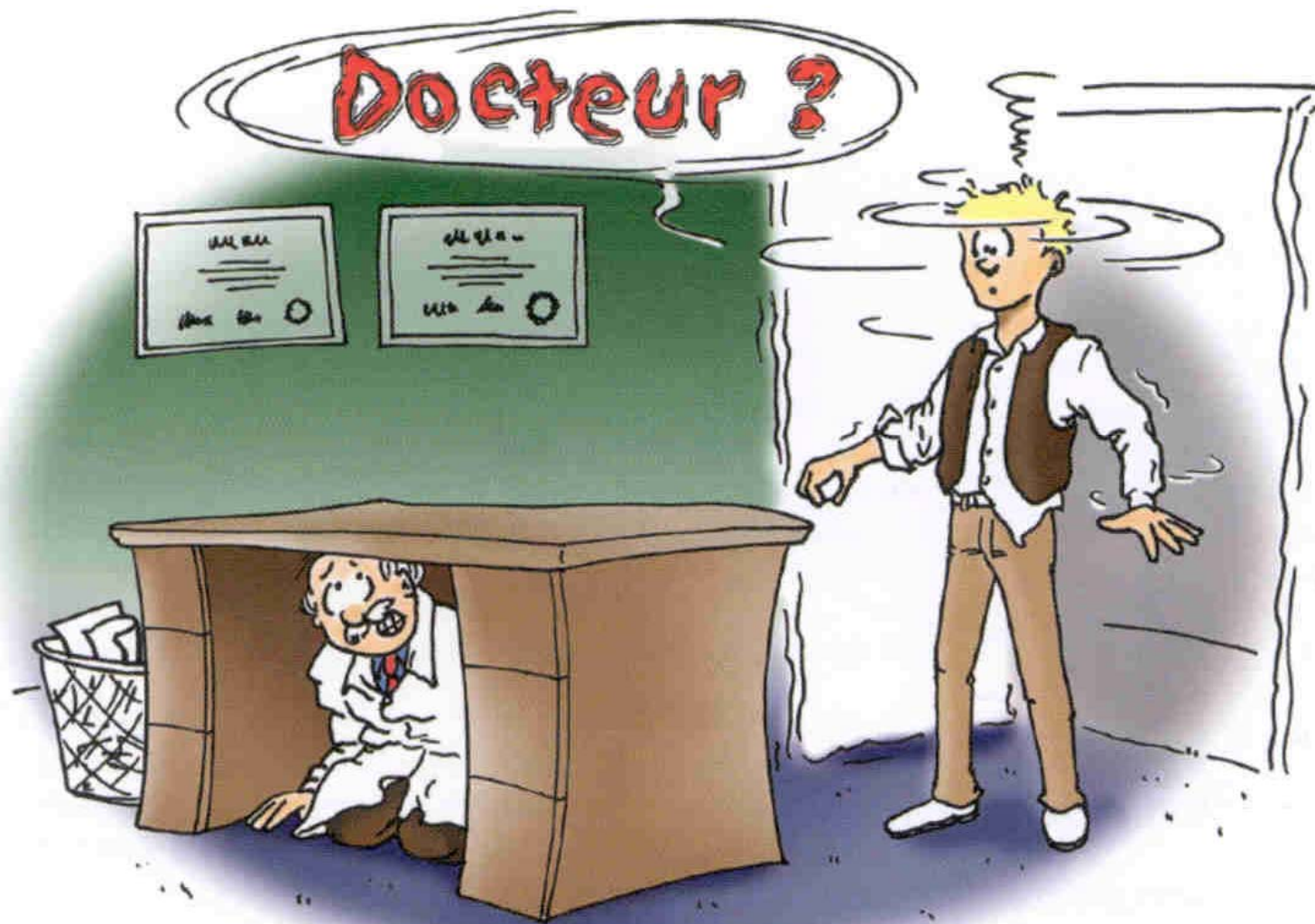
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***MBBS FRCSc***

***Assistant Professor***

***KAUH***

***Apr, 18 2005***



# *Objectives*

- *History*
- *Incidences*
- Pathology
- Diagnosis
- Treatment

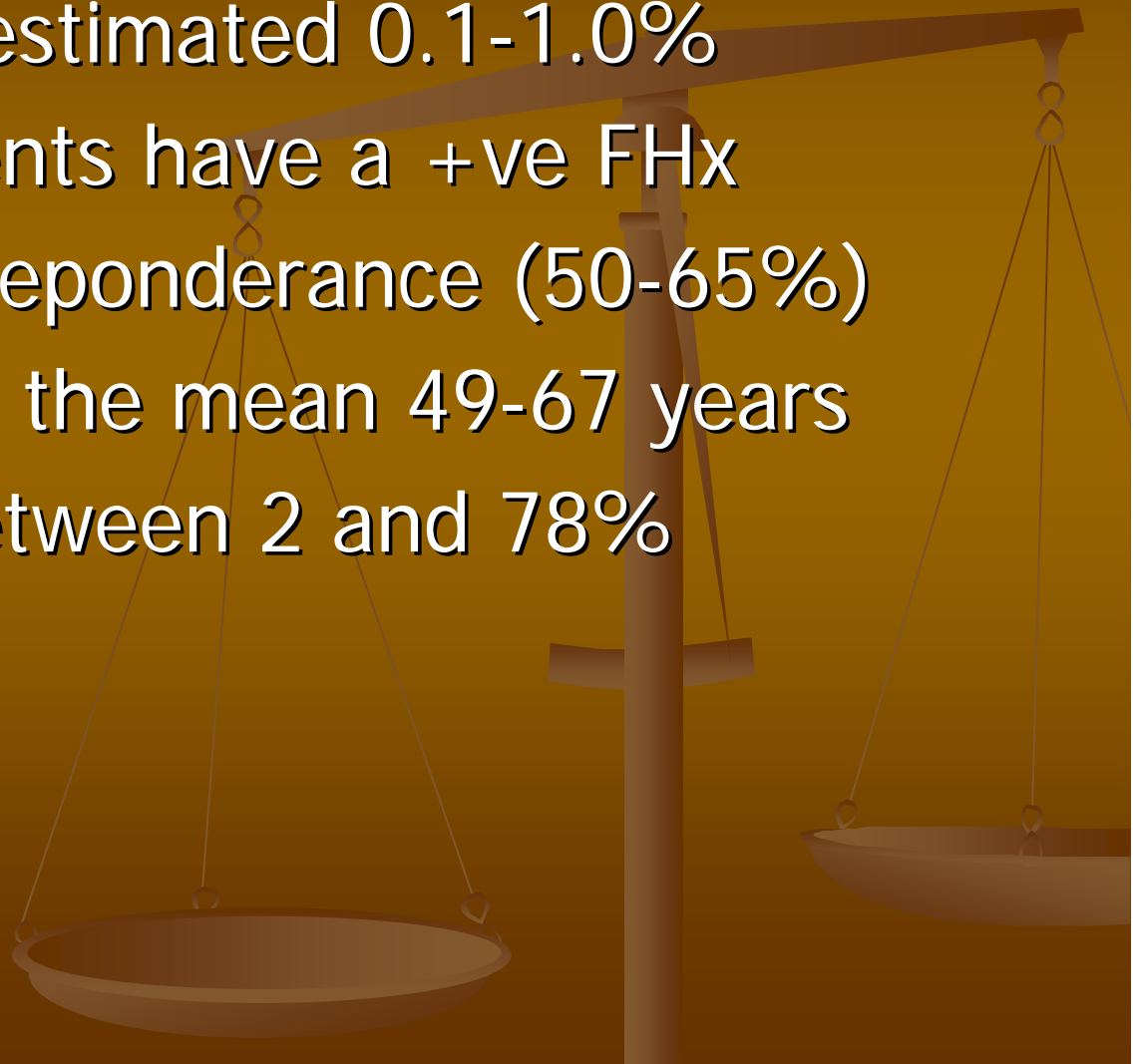


# *History*

- Apoplectic cerebral congestion
- 1861 Prosper Meniere "IE disturbance"
- 1927 Guild ELS site of endolymph outflow in pigs
- 1927 Portmann described ELS surgery as Rx
- Dandy proposed sectioning of cranial nerve VIII
- 1938, Hallpike and Cairns described endolymphatic hydrops in two patients who died from complications of surgery to section the eighth nerve providing, in this manner
- 1965 Schuknecht produced hydrops in pigs
- 1965 Dohlman infused artificial endolymph into perilymph → Menieres

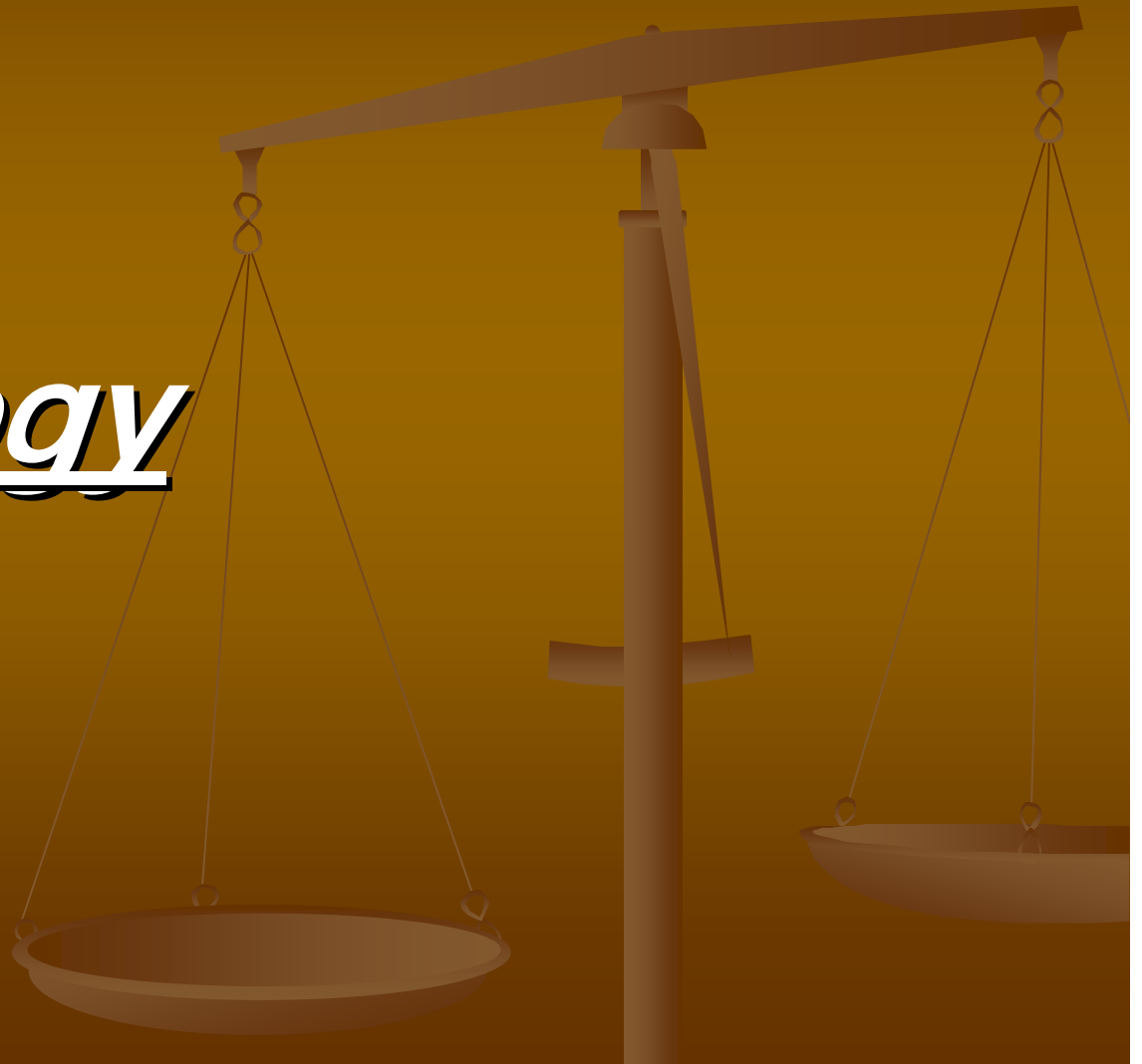
# ***Incidences***

- probably underestimated 0.1-1.0%
- half of the patients have a +ve FHx
- Slight female preponderance (50-65%)
- Almost all ages. the mean 49-67 years
- Contralateral between 2 and 78%  
(Dx+F/U)



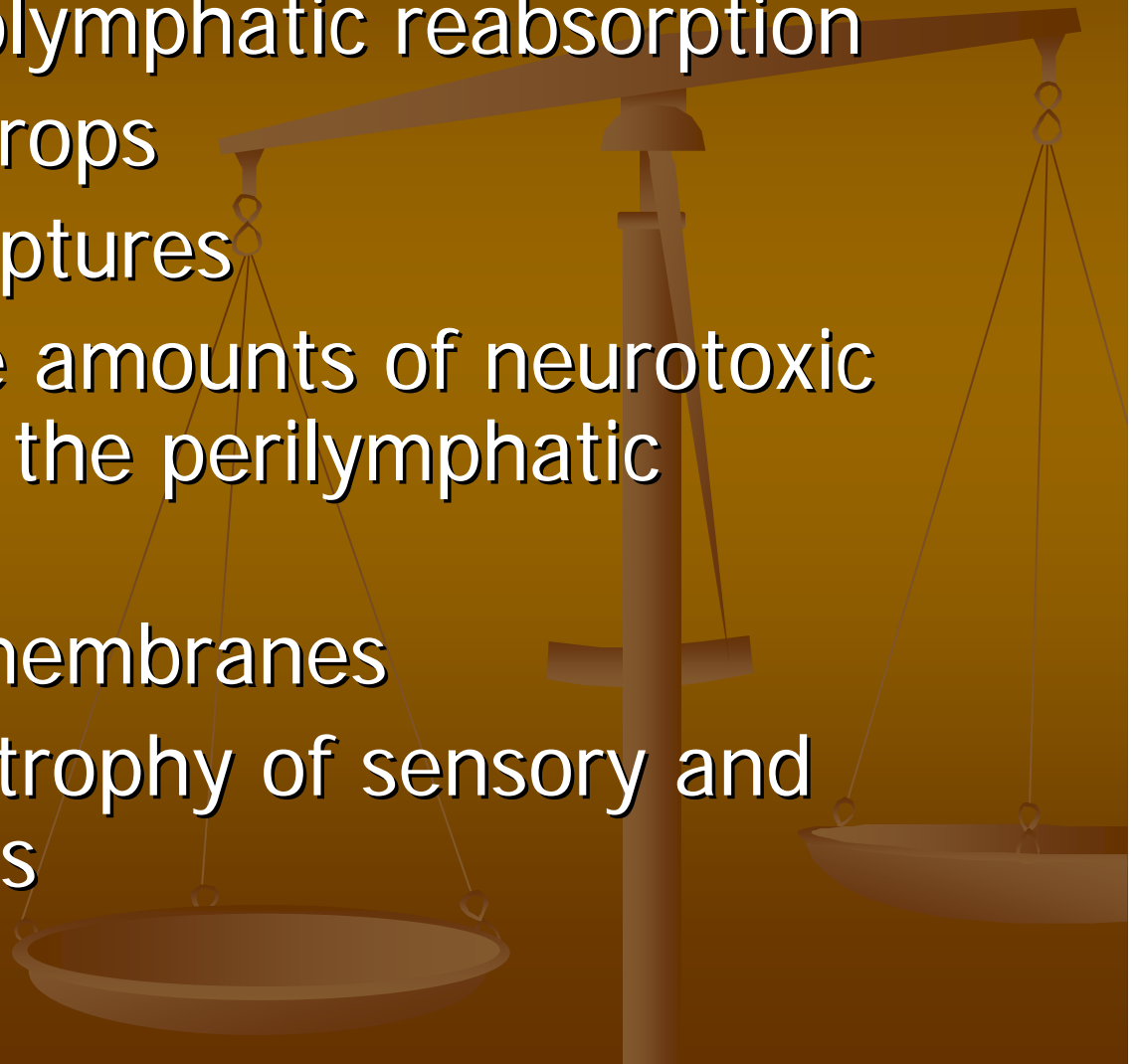
# *Objectives*

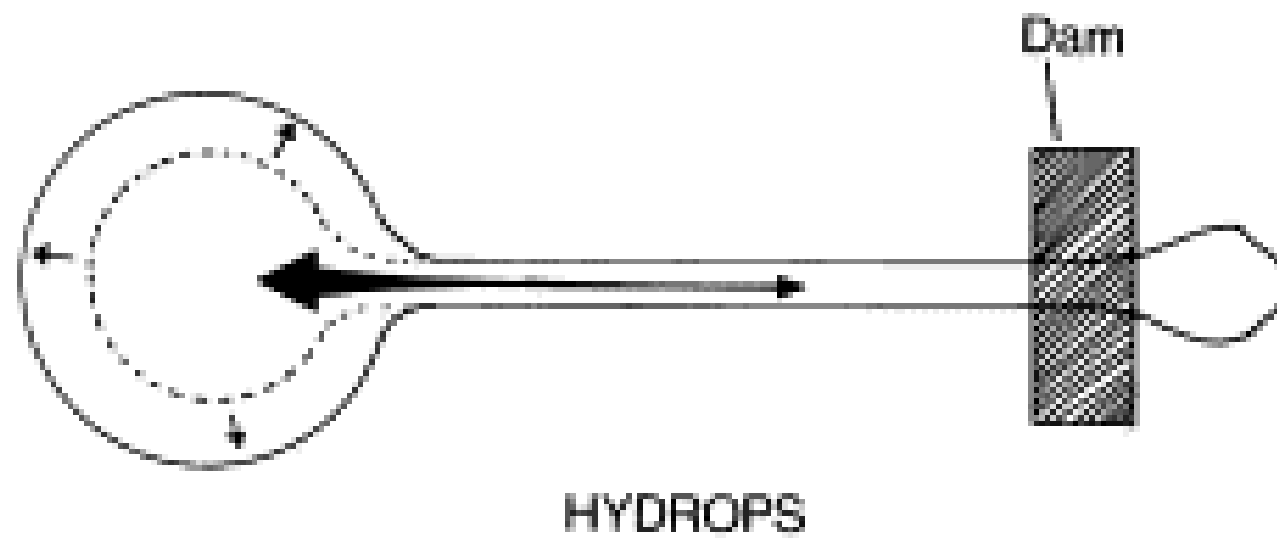
- History
- Incidences
- *Pathology*
- Diagnosis
- Treatment



# *Pathology*

- Decreased endolymphatic reabsorption
- progressive hydrops
- membranous ruptures
- Spillage of large amounts of neurotoxic endolymph into the perilymphatic compartment
- healing of the membranes
- distortion and atrophy of sensory and neural structures



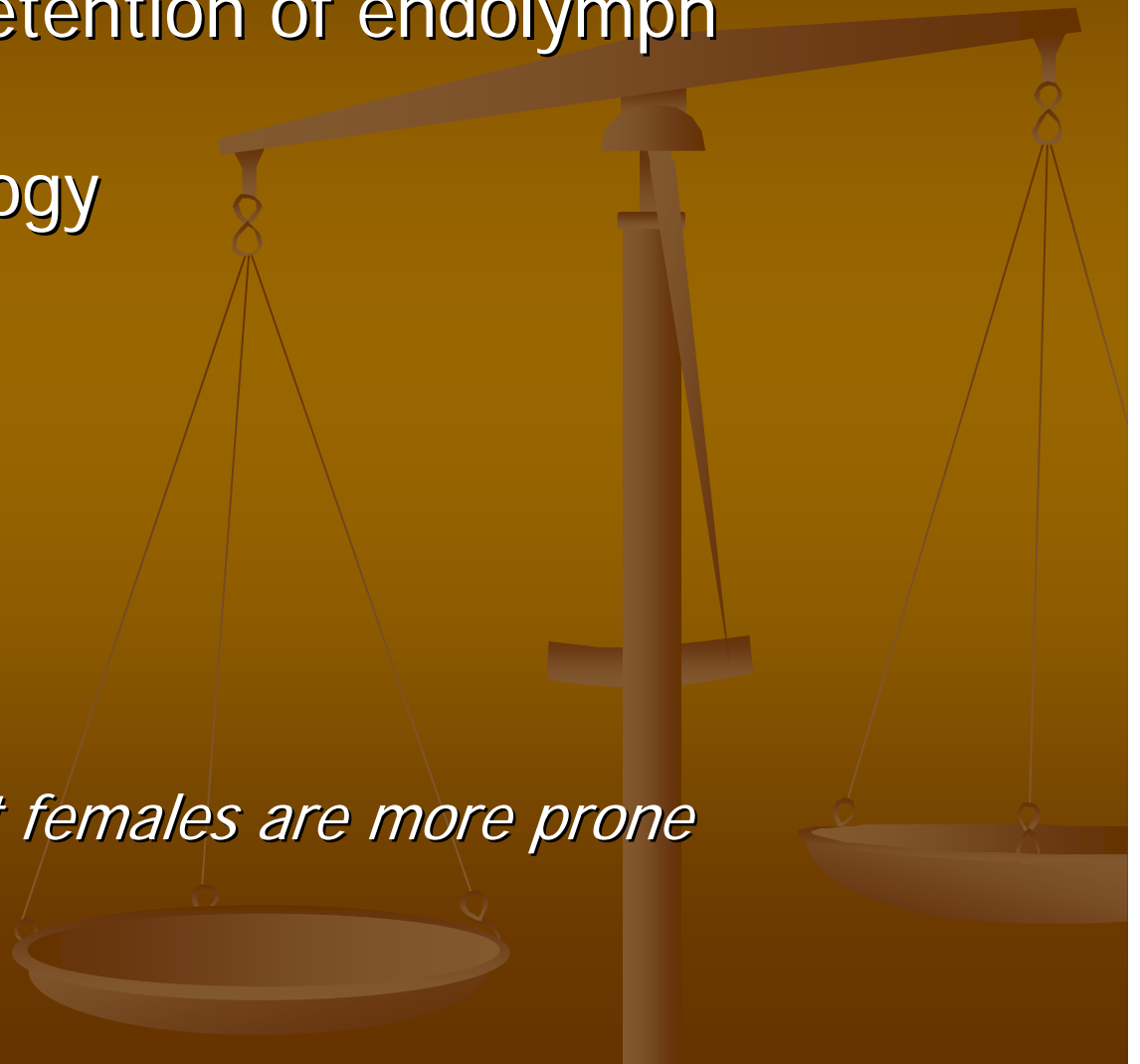




# ***Cause of Meniere's***

Overproduction or retention of endolymph

- Unknown
- Autoimmune etiology
- Ischemia
- Mumps
- Syphilis
- Hypothyroidism
- Head trauma
- Previous infection
- Hormonal *Pregnant females are more prone*



# *Meniere's Disease*

- Newer theories
  - Multifactorial inheritance
  - Immune-mediated phenomena
  - Association of allergies
- Study by Gottschlich\* et al.
  - 50% meeting criteria have antibodies to 70-kD heat-shock protein

Laryngoscope. 1995 Dec;105(12 Pt 1):1347-52

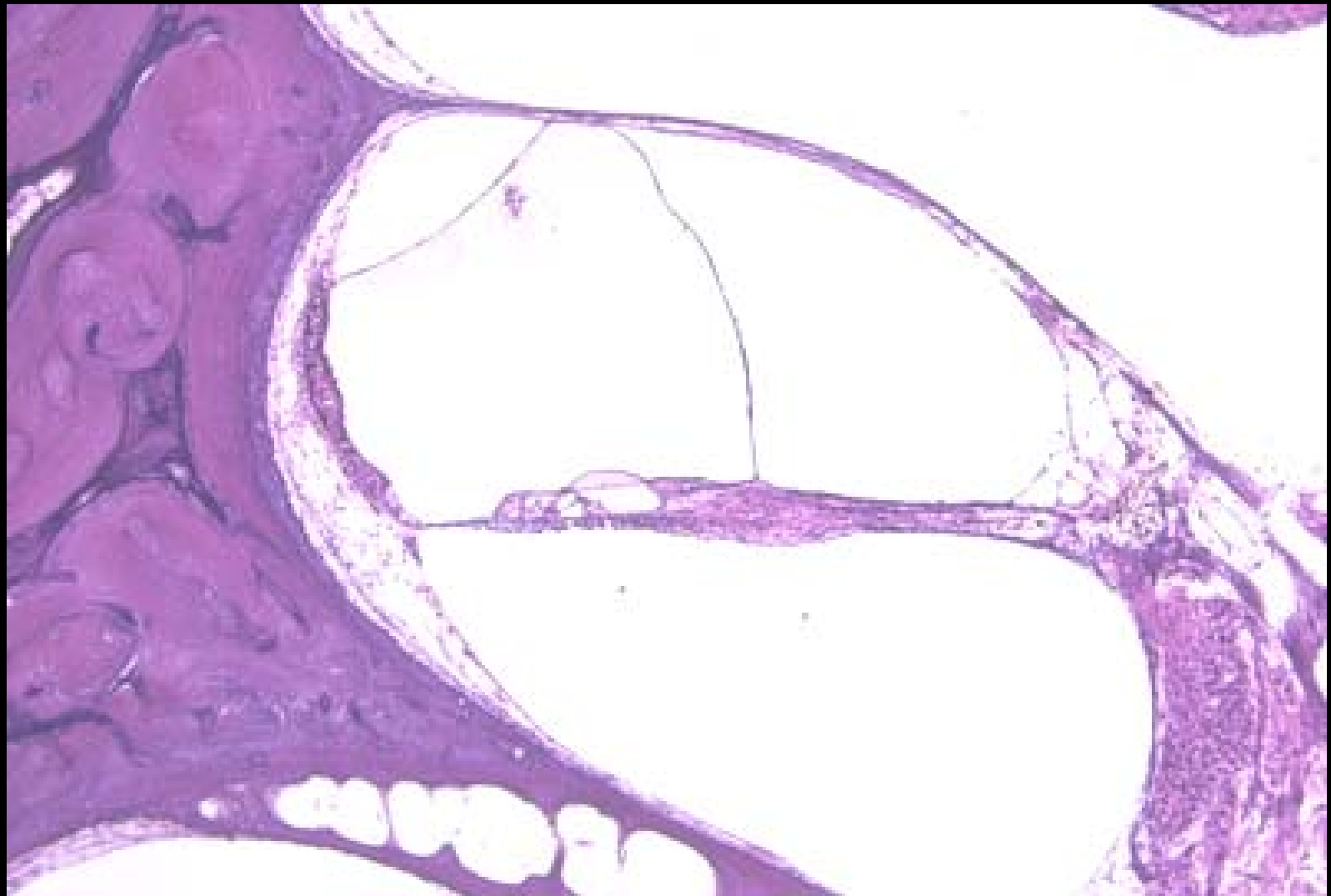
# ***Pathophysiology***

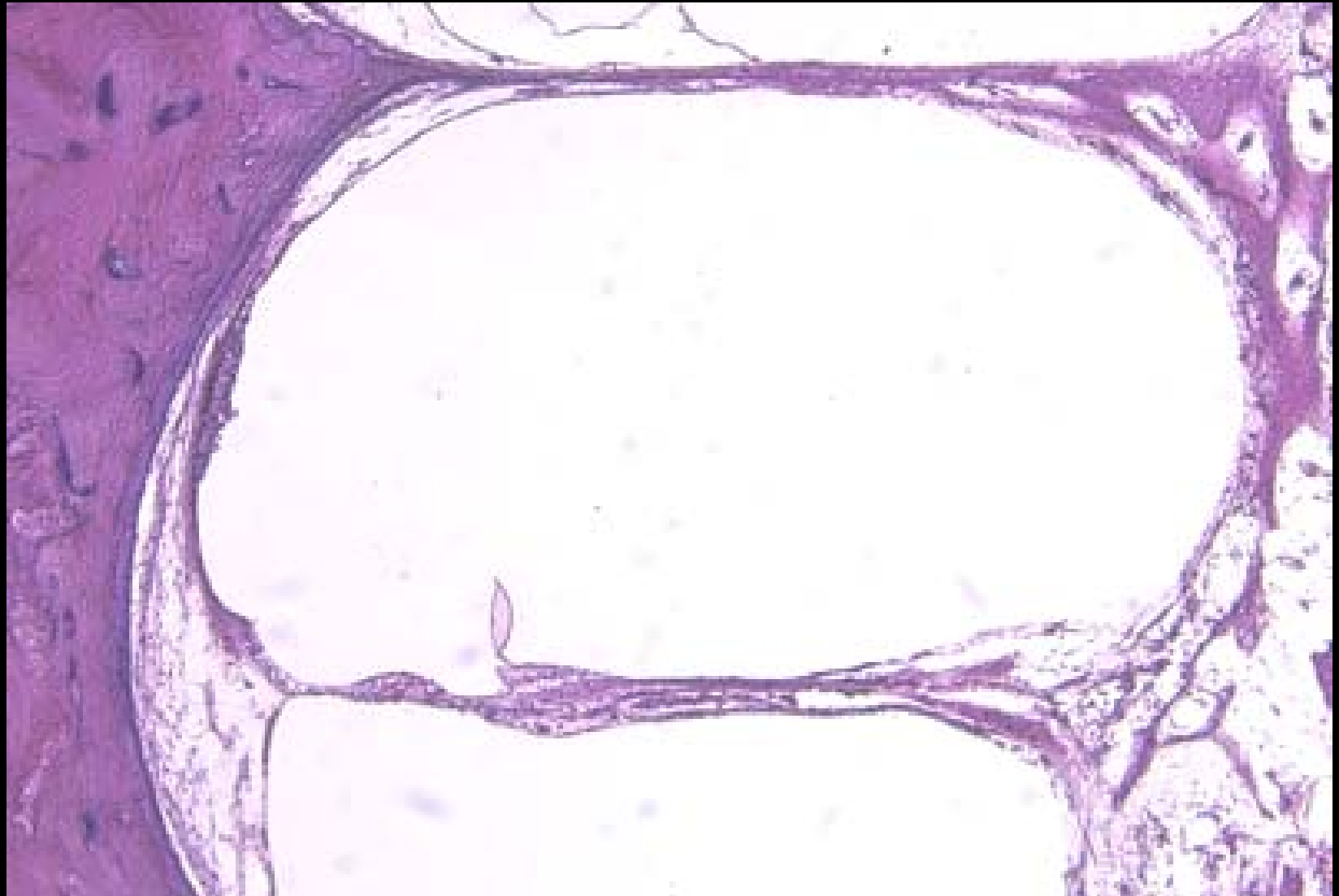
## Histology

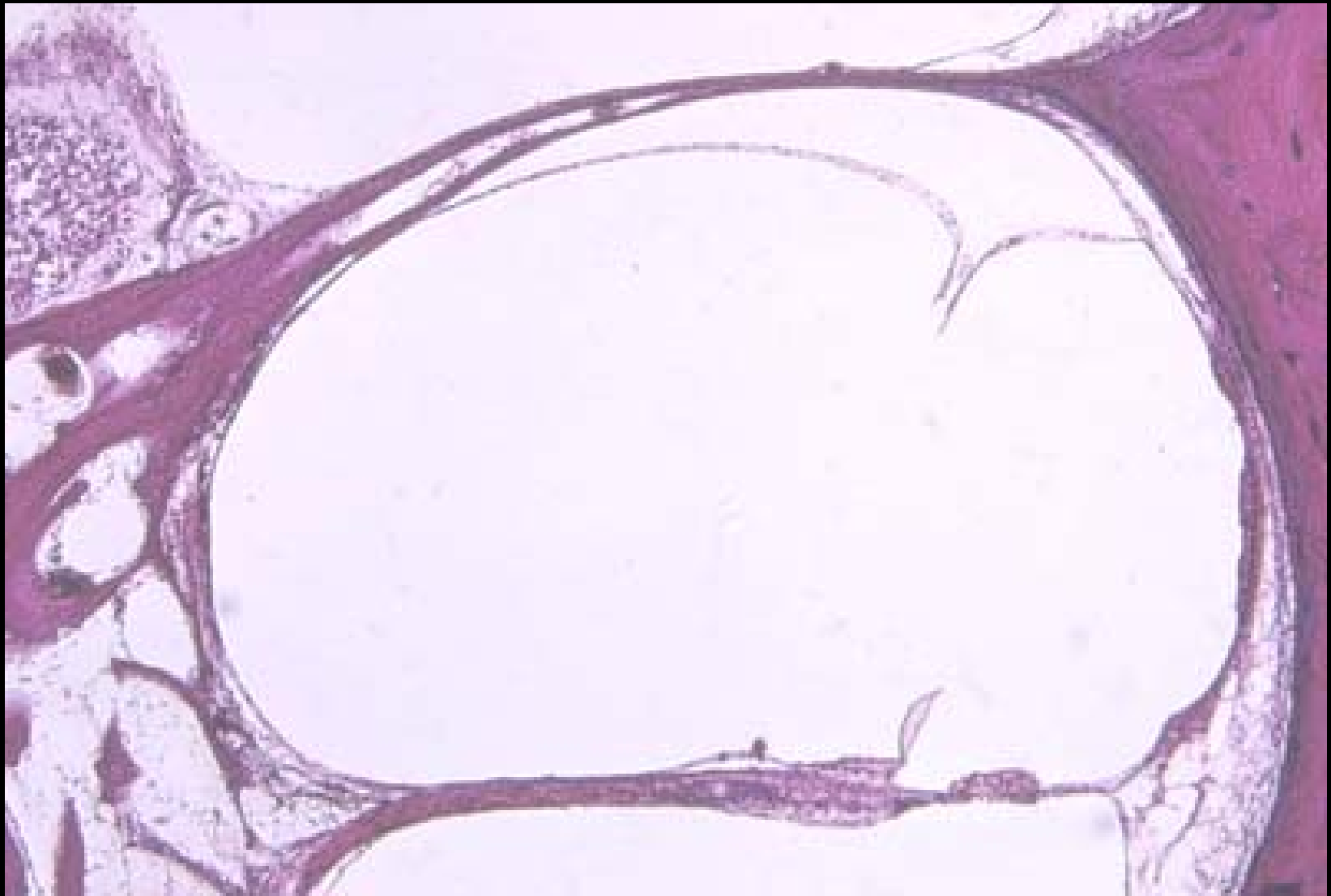
- Raising of Reissner membrane
- Dilation of endolymphatic spaces
- perisaccular ischemia and fibrosis
- smaller diameters of the ducts
- severely affected cases
  - decreases in hair cells
  - loss of the neuronal population
- accumulations of excess glycoprotein
- Membranous ruptures + Healed scars
- shortening of stereocilia



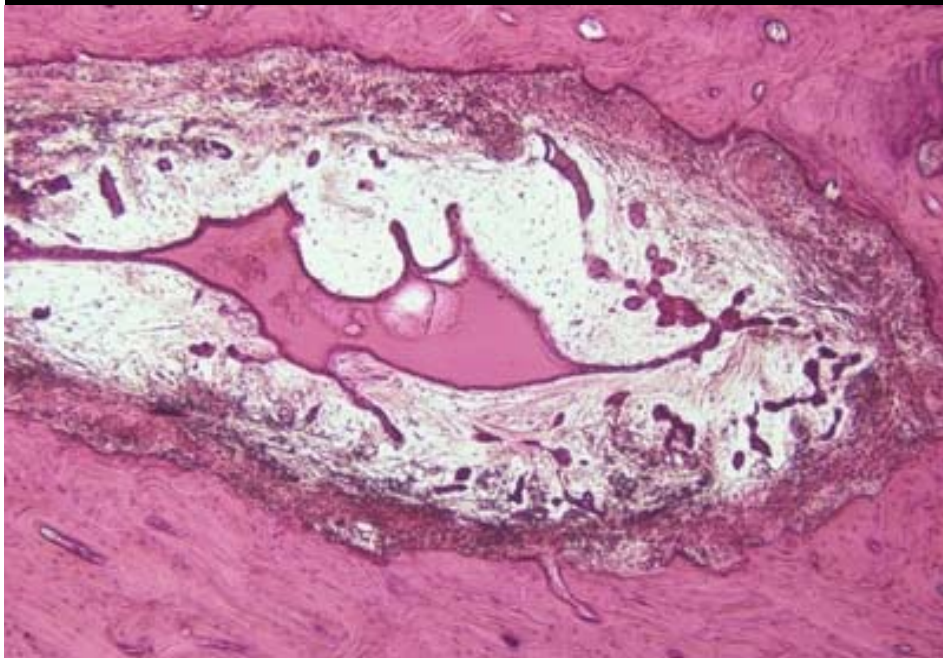








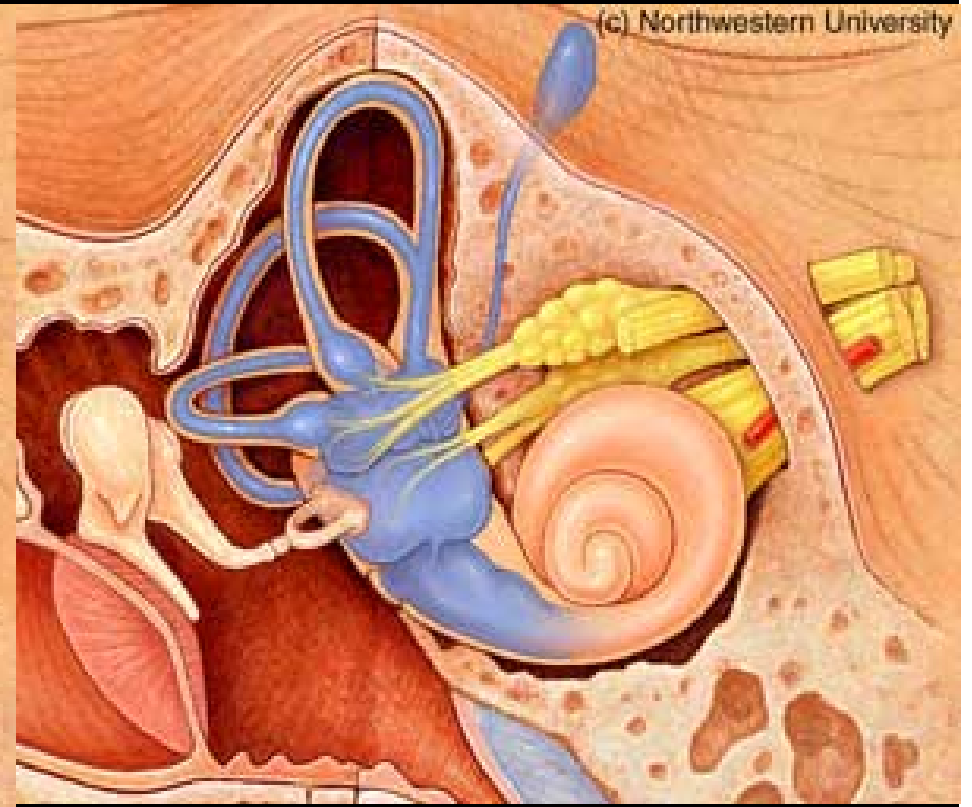
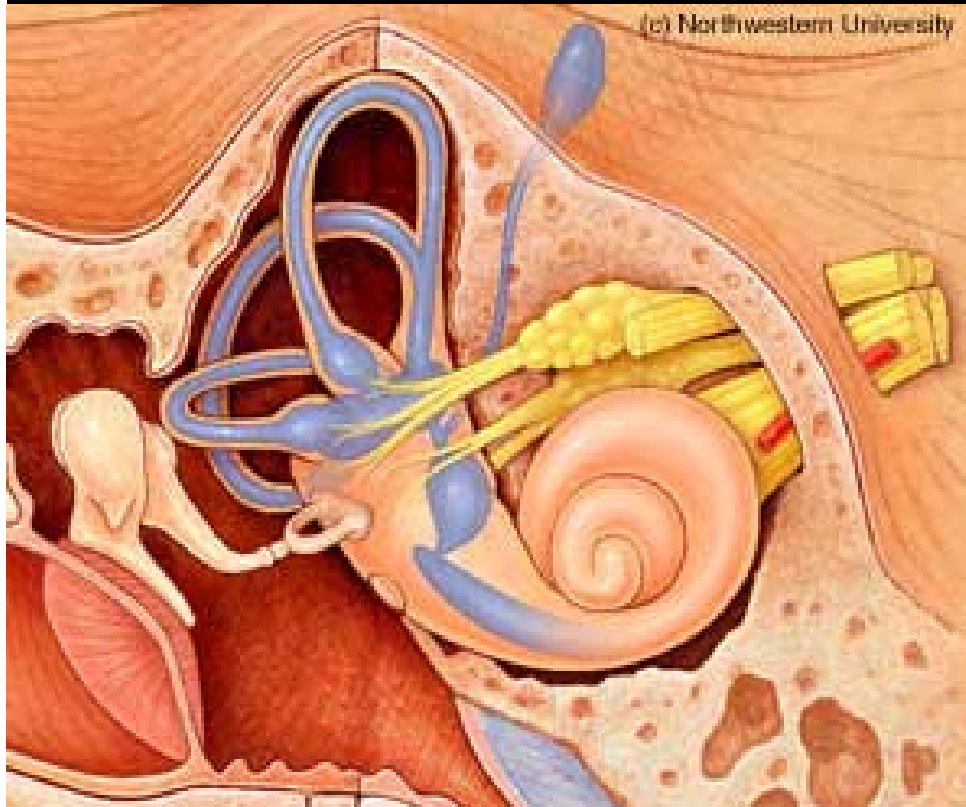
**NORMAL ENDOLYMPHATIC SAC**



**MENIERE'S DISEASE**







**Endolymphatic hydrops is most consistently found in the pars inferior (cochlea and saccule)**

# ***Endolymphatic hydrops***

Rauch et al

- double-blind temporal bone study demonstrated histologic evidence of endolymphatic hydrops in 13/13 cases of clinical Ménière's disease.
- Review of medical records associated with six of 19 temporal bones with endolymphatic hydrops, however, did not reveal symptoms or signs of Ménière's disease.

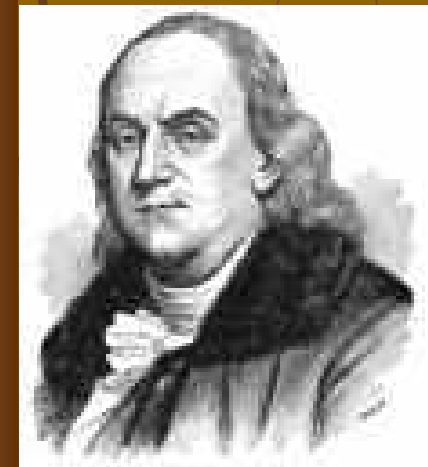
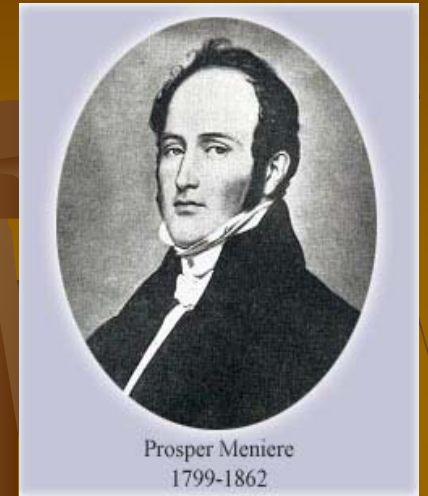
# *Menieres - Course*

## ■ Early

- Predominant Vertigo
- Deafness
- Normal hearing between

## ■ Later

- Hearing loss stops fluctuating
- Progressively worse (50db)



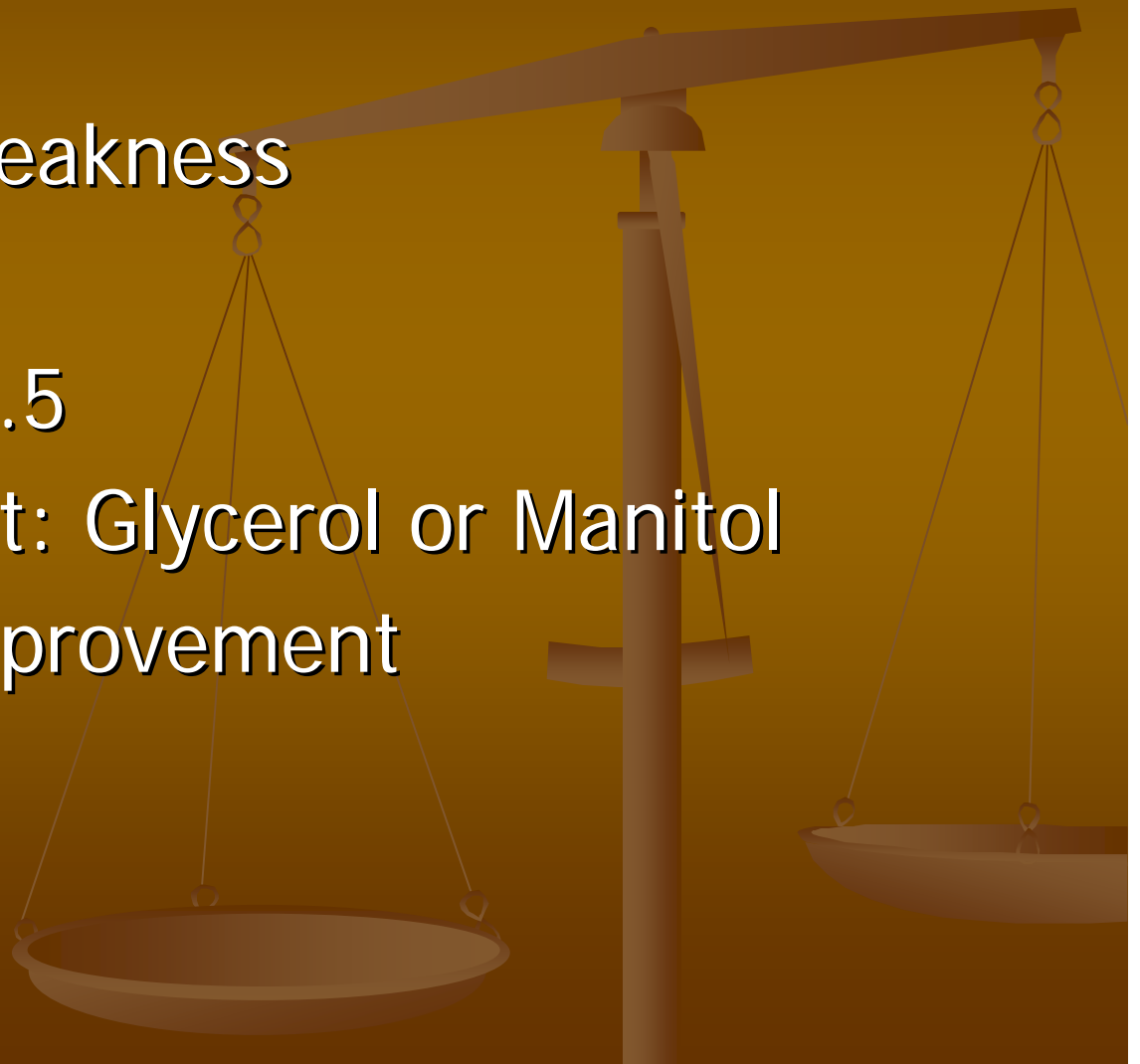
# *Objectives*

- History
- Incidences
- Pathology
- *Diagnosis*
- Treatment



# *Diagnosis*

- Hx
- PE: unilateral weakness
- PTA: LF-SNHL
- ECoG: SP/AP > 0.5
- Dehydration test: Glycerol or Mannitol  
→ temporary improvement
- R/O other Dx

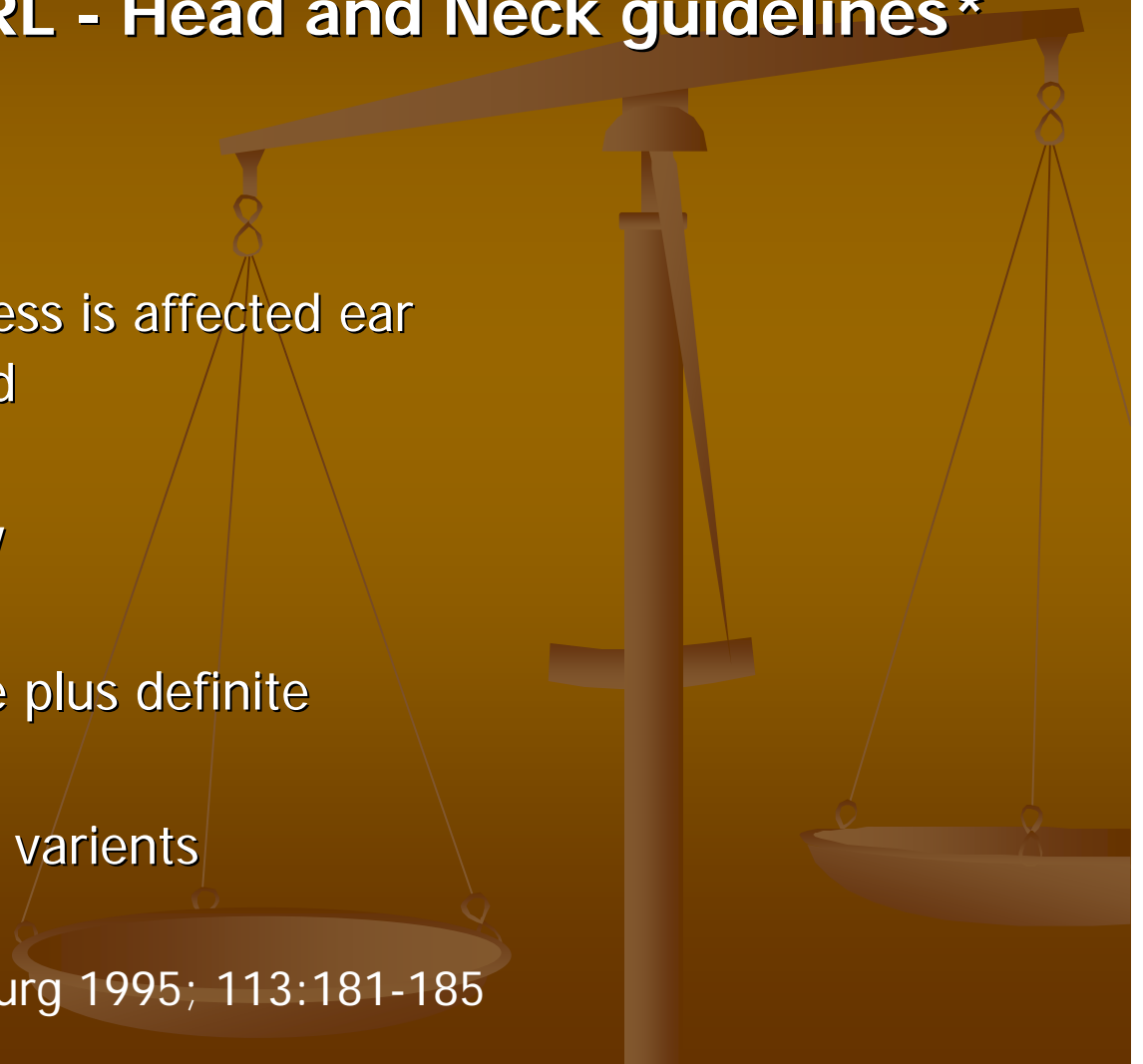


# *Meniere's Categories*

## The A. Academy of ORL - Head and Neck guidelines\*

- Definite
  - Two episodes 20 min
  - Documented SNHL
  - Tinnitus or aural fullness is affected ear
  - Other causes excluded
- Certain
  - Definite plus histology
- Probable
  - One definitive episode plus definite
- Possible
  - Cochlear or vestibular variants

\*Otolaryngol Head Neck Surg 1995; 113:181-185

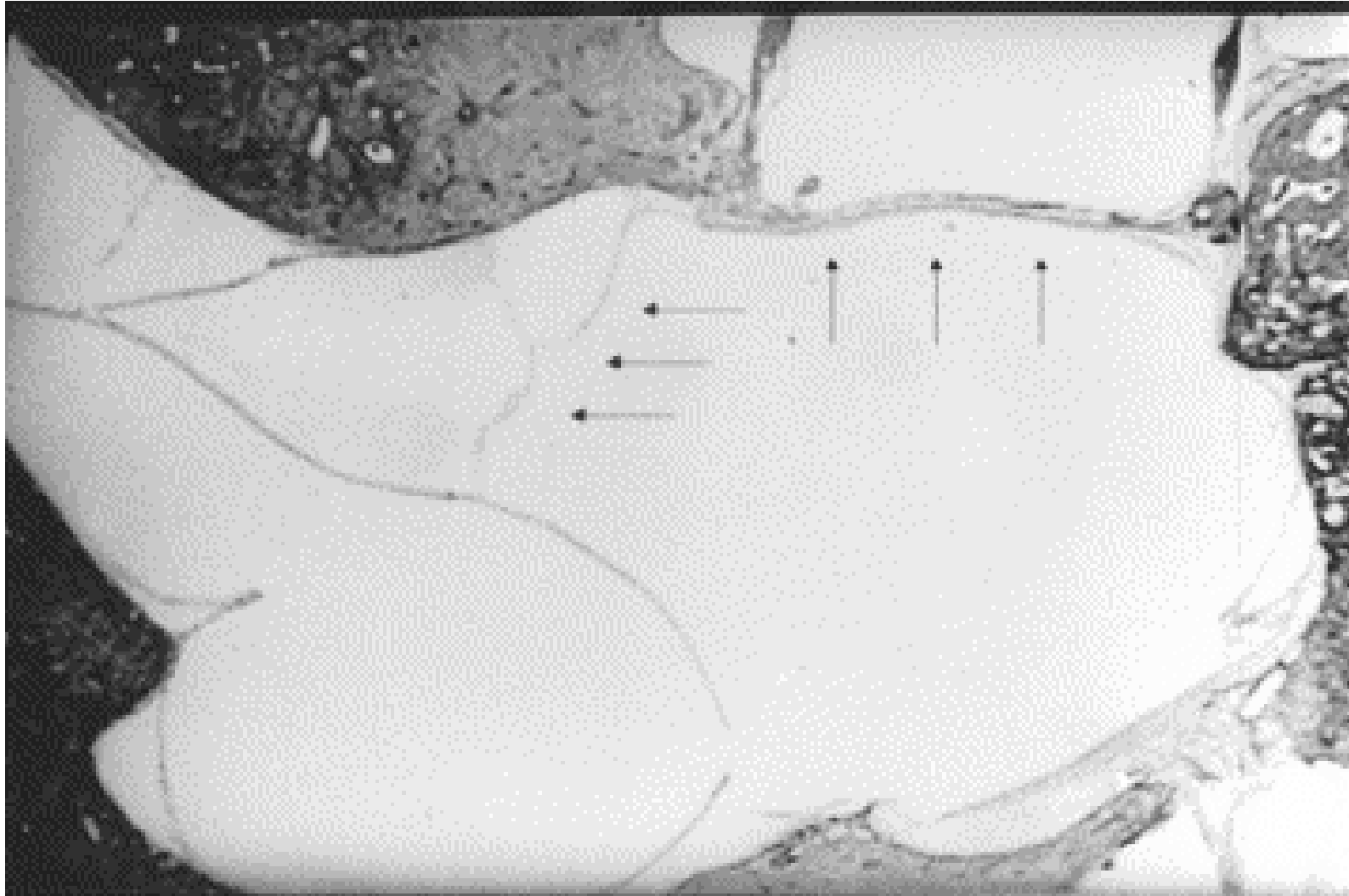


# *Meniere's syndrome / disease*

- 50 Y Female
- Lt stapedectomy 2 y ago → Dead ear
- Rt Ear
  - HL
  - Fullness & Tinnitus
  - Vertigo X3 for 2 hours
  - +ve Fistula test
  - +ve Dix Hal-
- PTA
  - Peak audiogram\*



\*Paparella Arch Otolaryngol 1982;108:555

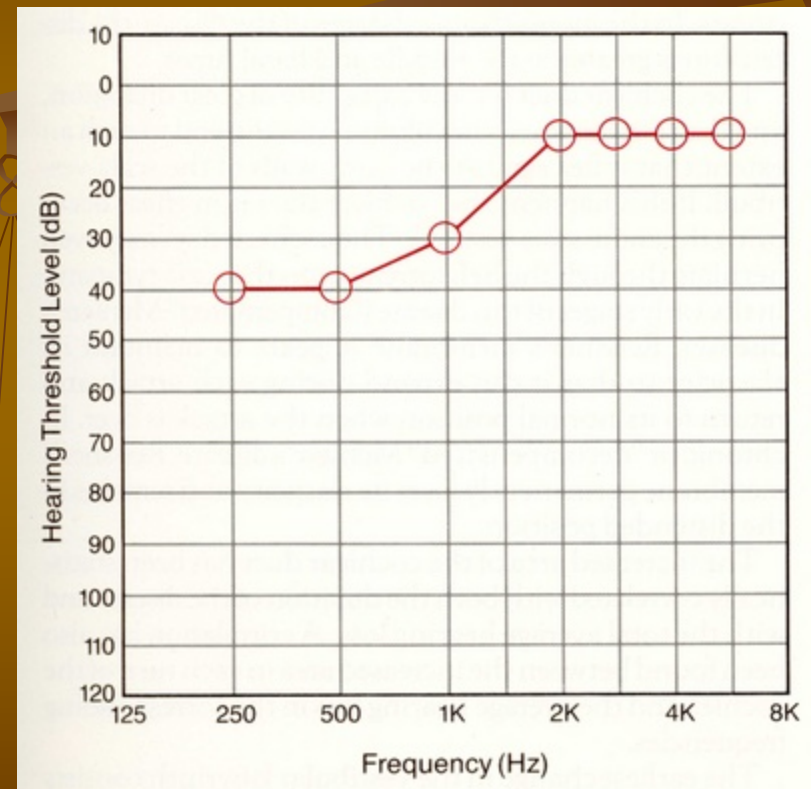


Stapedectomy



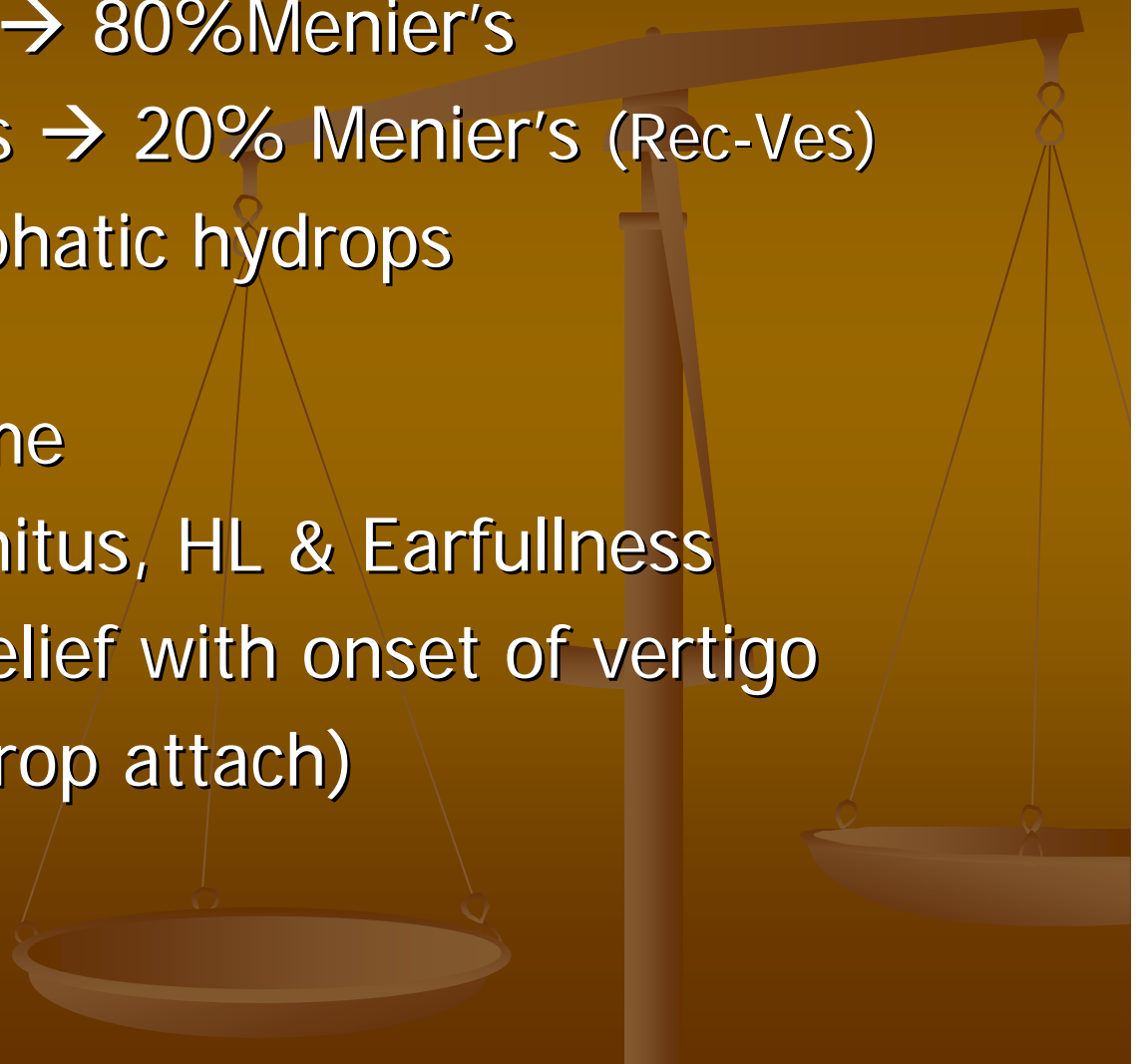
# ***Tumarkin crisis***

- 55 Y Female
- Rt Ear
  - HL
  - Sudden fall
  - Vertigo X5 for 5 hours



# ***Variants***

- Cochlear hydrops → 80% Menier's
- Vestibular hydrops → 20% Menier's (Rec-Ves)
- Delayed endolymphatic hydrops  
HL → Menier's
- Lermoyez syndrome  
Increasing tinnitus, HL & Earfullness  
→ Sudden relief with onset of vertigo
- Tumarkin crisis (drop attack)



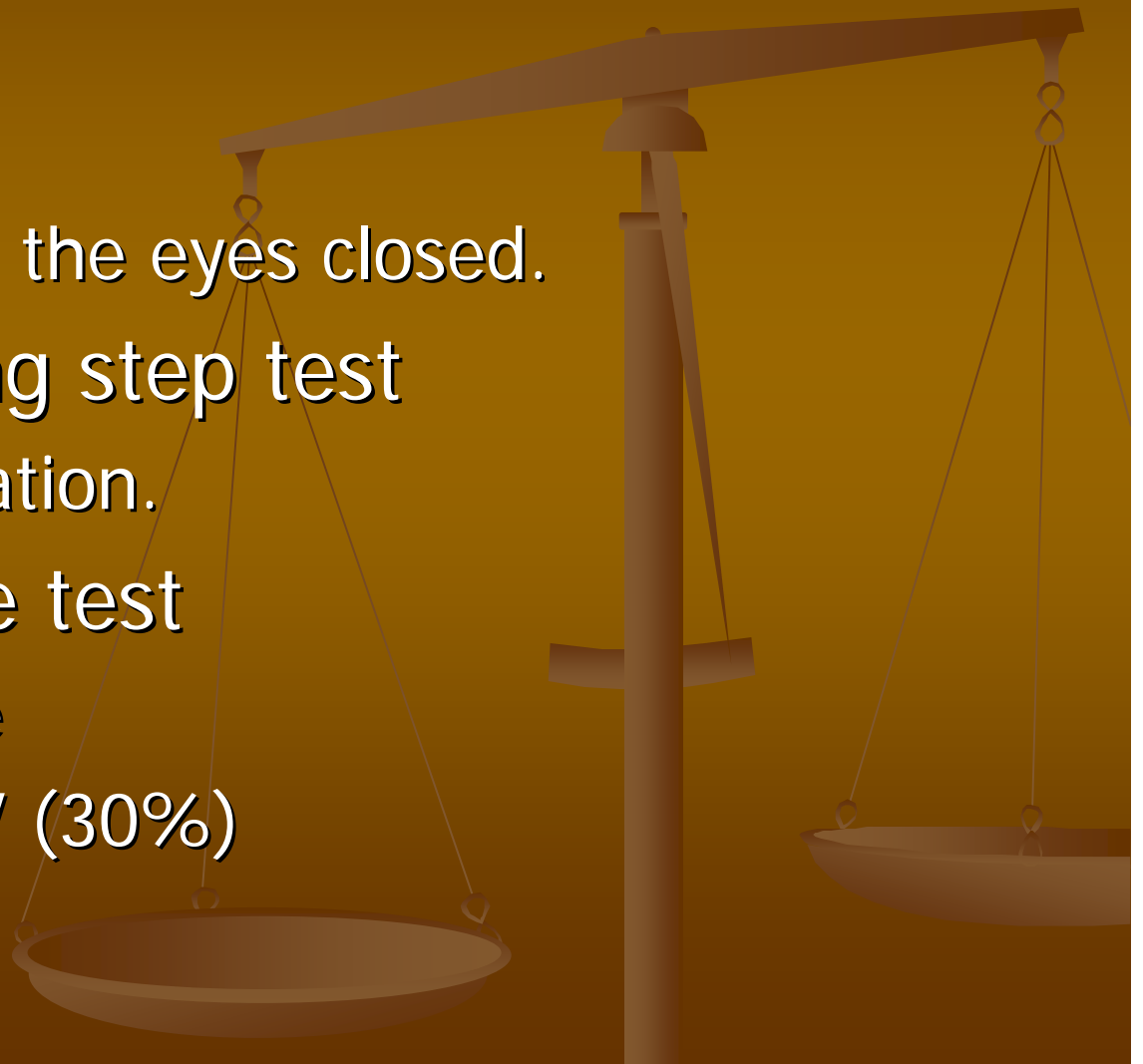
# *Otolithic crises of Tumarkin*

- Tumarkin\*
- drop attacks
- acute utriculosaccular dysfunction
- Abrupt otolith input → Loss of extensor tone
- NO loss of consciousness
- occur in 2-6%
- D/Dx cardiogenic vertebral basilar insufficiency  
epilepsy

\*BMJ (Clinical Research) 1936; 2:175-177

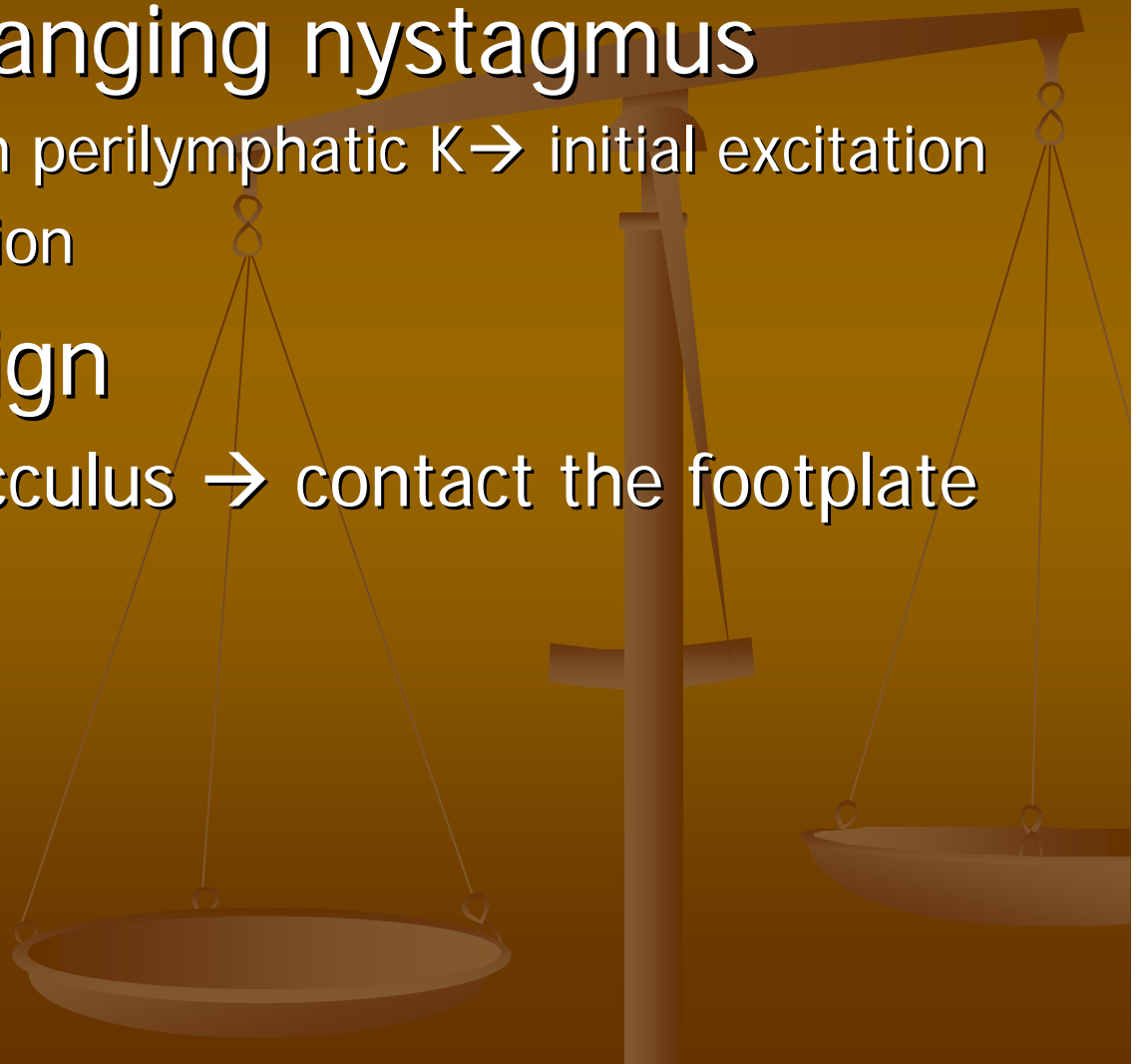
# PEX

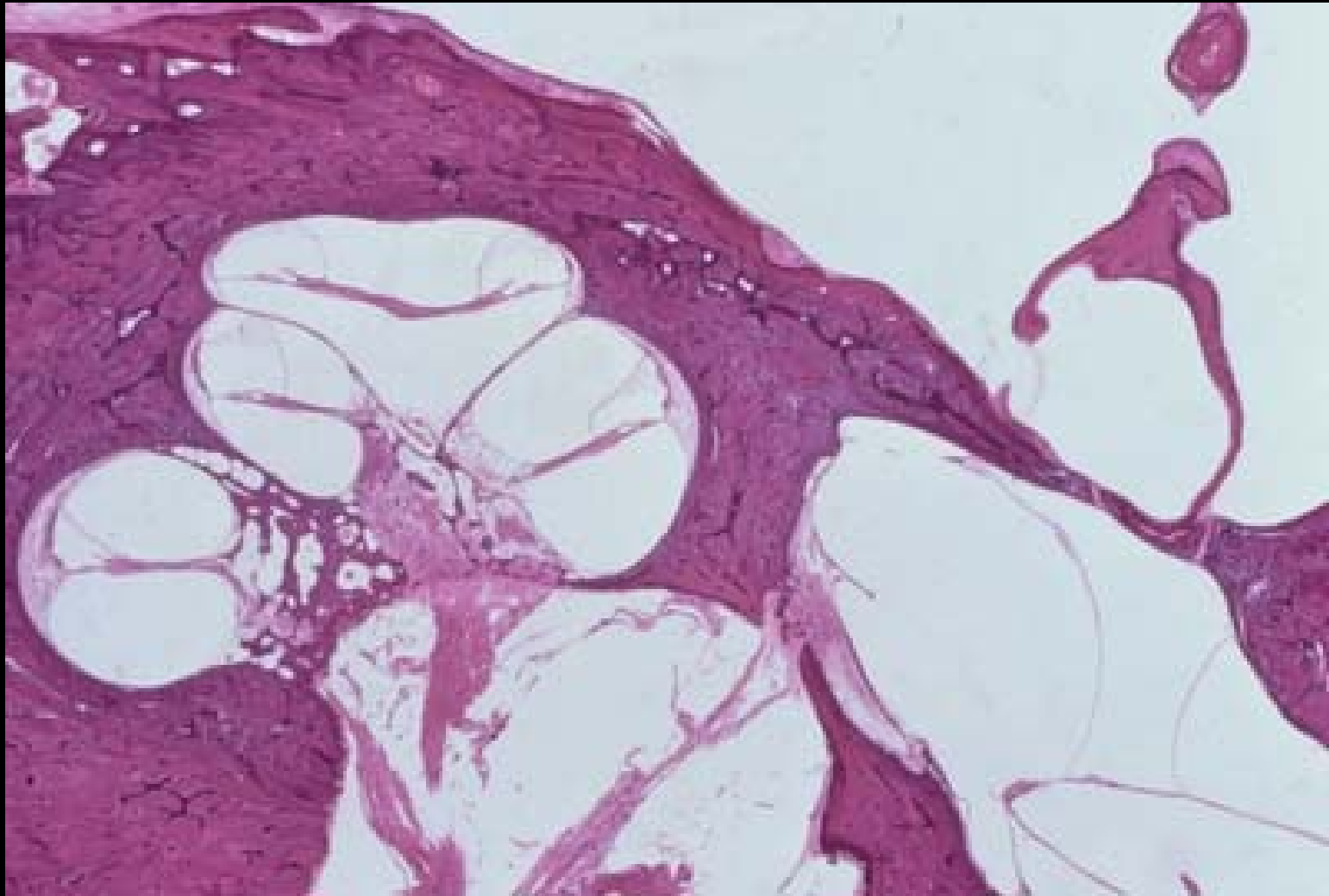
- Romberg test
  - Instability
  - Worsening with the eyes closed.
- Fukuda marching step test
  - significant deviation.
- The Dix-Hallpike test
  - may be positive
  - coexisting BPPV (30%)



# PEX

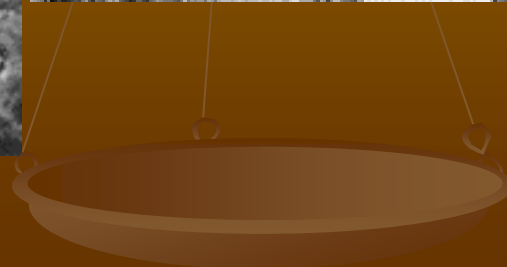
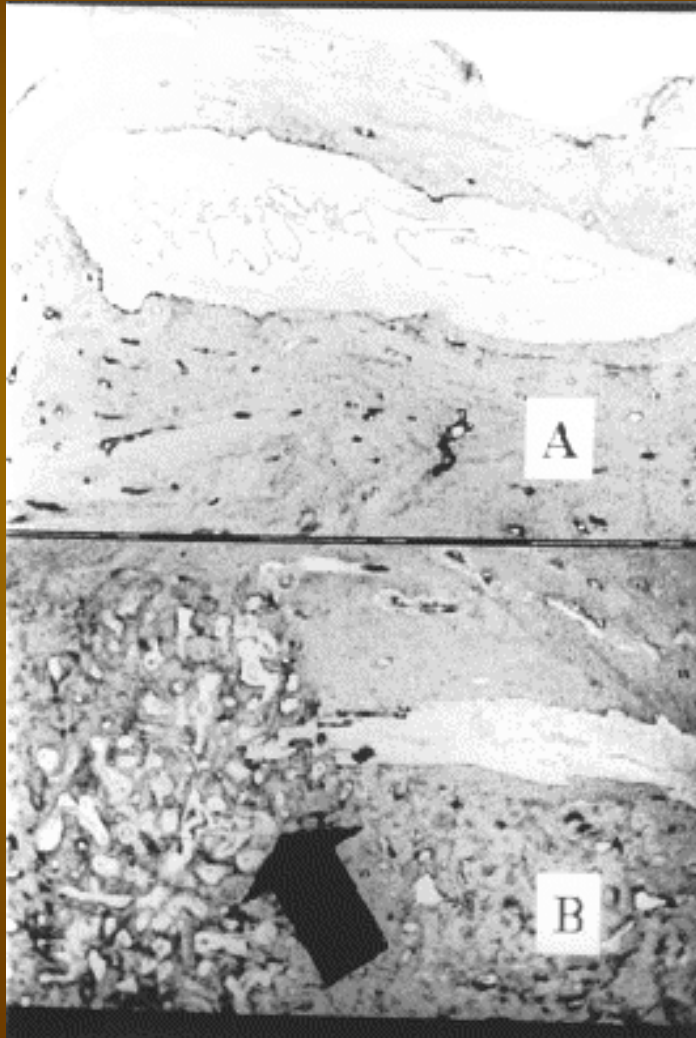
- A direction-changing nystagmus
  - Ruptures → rise in perilymphatic K<sup>+</sup> → initial excitation
  - blockade → inhibition
- Hennebert's sign  
expansion of the sacculus → contact the footplate directly
- Recruitment





# *D/Dx*

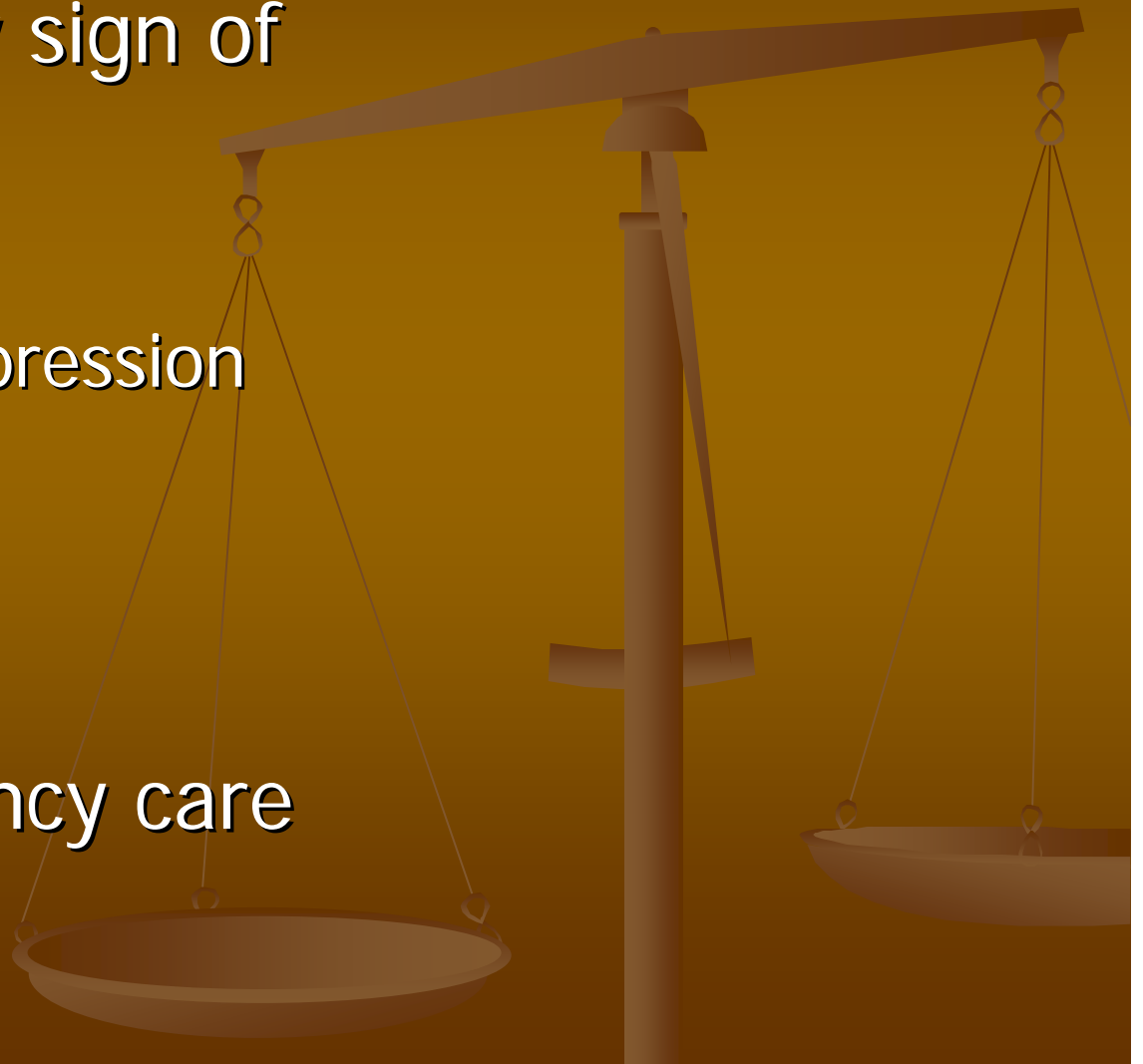
- I. Vertigo lasting seconds (BPPV)
- II. Vertigo lasting minutes to hours
  - A. Idiopathic endolymphatic hydrops (Ménière's)
  - B. Secondary endolymphatic hydrops
    - 1. Otic syphilis
    - 2. Delayed endolymphatic hydrops
    - 3. Cogan's disease
    - 4. Recurrent vestibulopathy
- III. Vertigo lasting days (vestibular neuritis)
- IV. Vertigo of variable duration
  - A. Inner ear fistula
  - C. Familial vestibulopathy





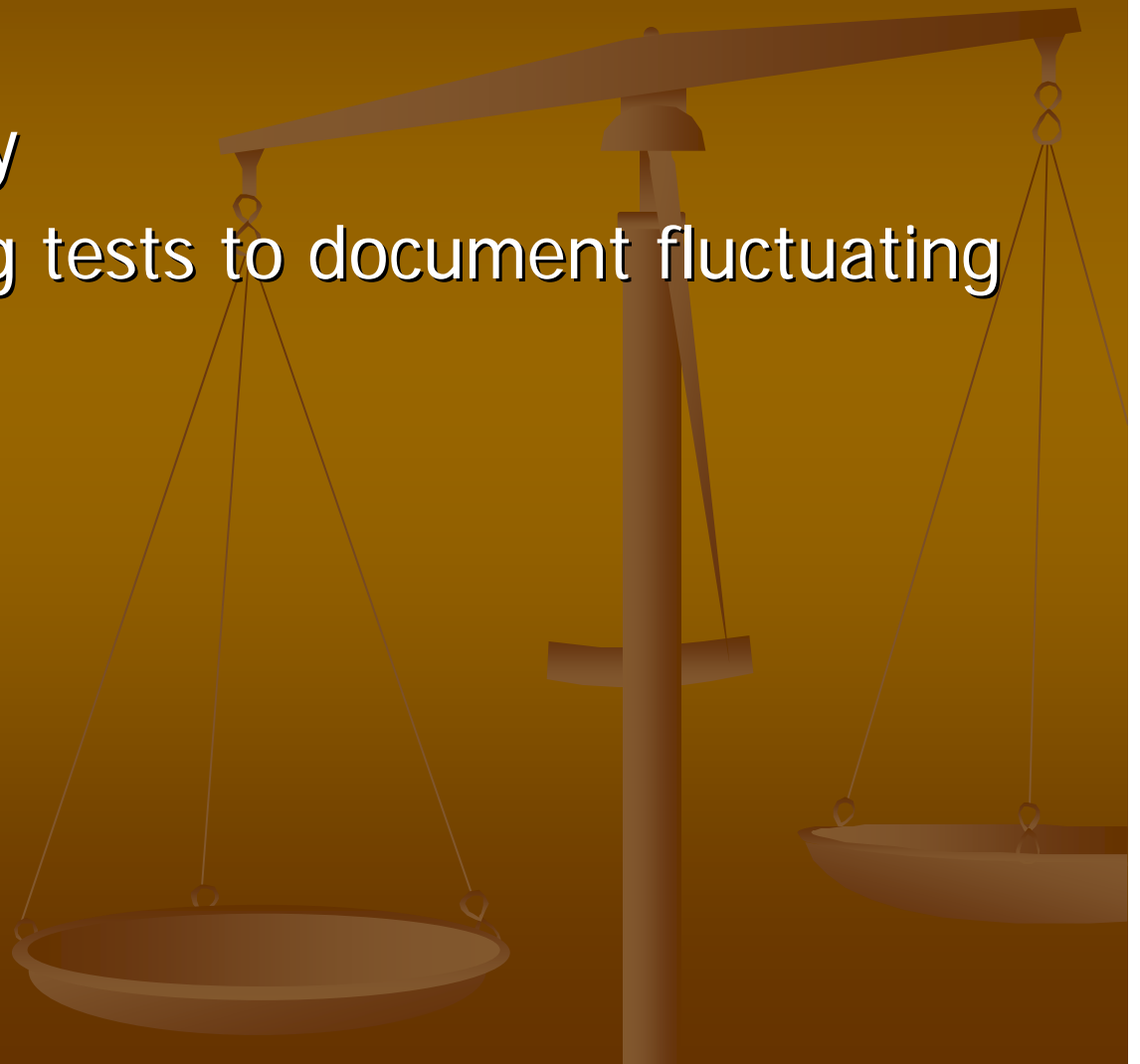
# ***New-onset vertigo***

- May be an early sign of
  - Stroke
  - Migraine
  - brainstem compression
  - Labrynthitis
  - Fistula
  - SSNHL
- require emergency care



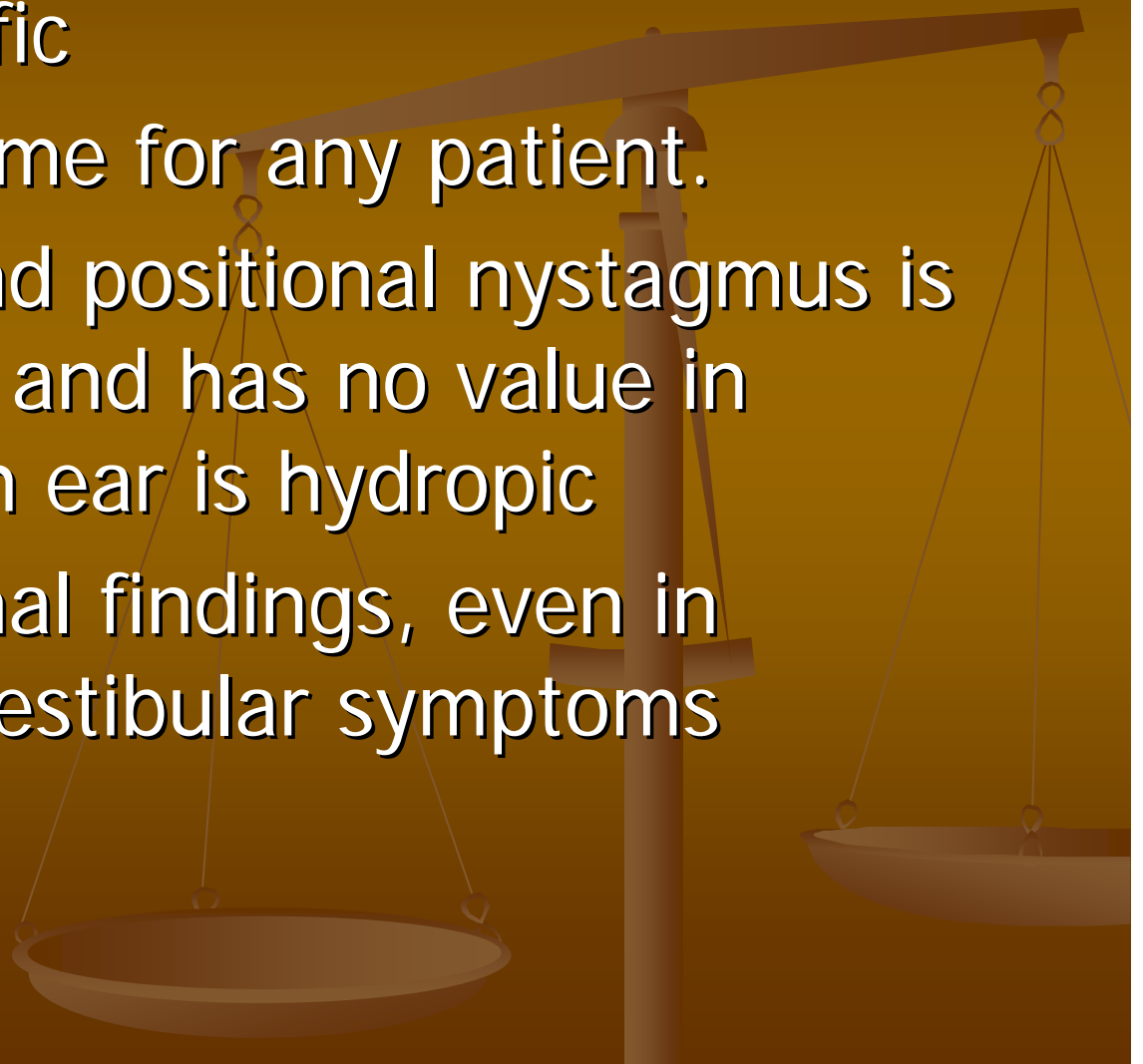
# *Investigation*

- PTA
  - A low-frequency
  - multiple hearing tests to document fluctuating hearing
  - Recruitment



# ***Electronystagmography***

- highly nonspecific
- fluctuate with time for any patient.
- Spontaneous and positional nystagmus is frequently seen and has no value in predicting which ear is hydropic
- 50% have normal findings, even in incapacitating vestibular symptoms



# ***ECOG***

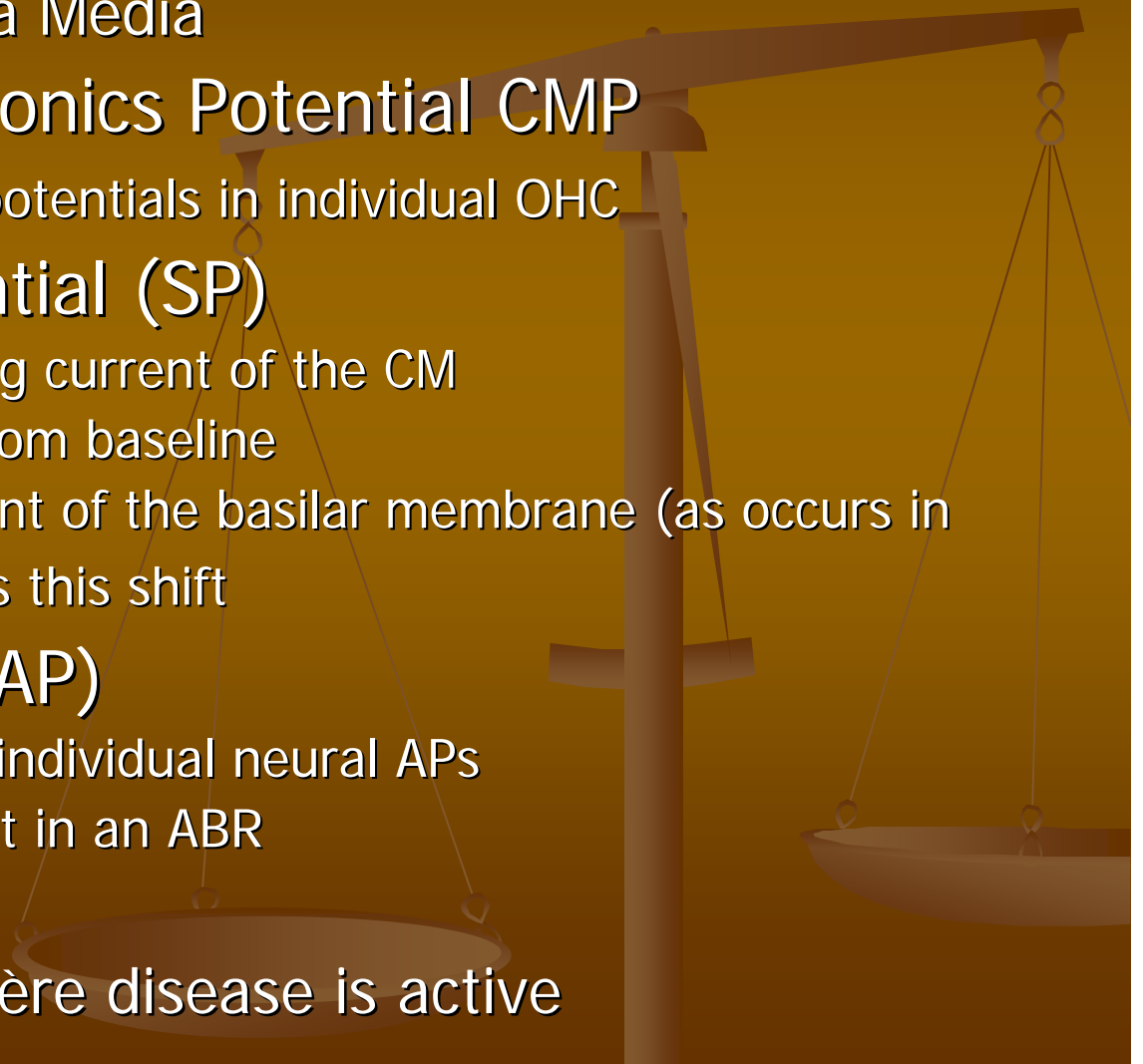
## **Cochlear Potentials**

1. Endolymphatic Potential EP
2. Cochlear microphonics CM
3. Summating potential (SP)
4. Action potential (AP)



# ECOG

- Endolymphatic Potential EP
  - Stria Vasi → Scala Media
- Cochlear microphonics Potential CMP
  - sum of intracellular potentials in individual OHC
- Summating potential (SP)
  - sum of the alternating current of the CM
  - direct-current shift from baseline
  - Asymmetric movement of the basilar membrane (as occurs in hydrops) exacerbates this shift
- Action potential (AP)
  - sum of synchronous individual neural APs
  - as the wave I present in an ABR
- $SP/AP > 50\%$
- Accurate when Ménière disease is active



Lilly TM electrode

Alternating clicks

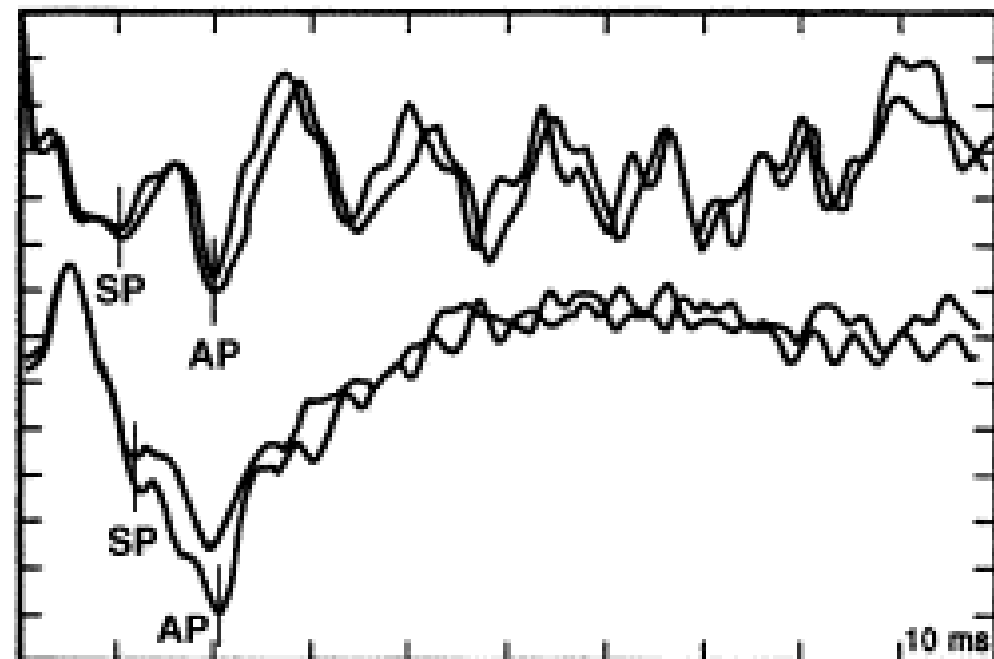
90 dBnHL

ER-3 insert phones

Amplaid MK 12

Upper trace: normal ear

Lower trace: hydropic ear



Nickel electrode

Alternating clicks

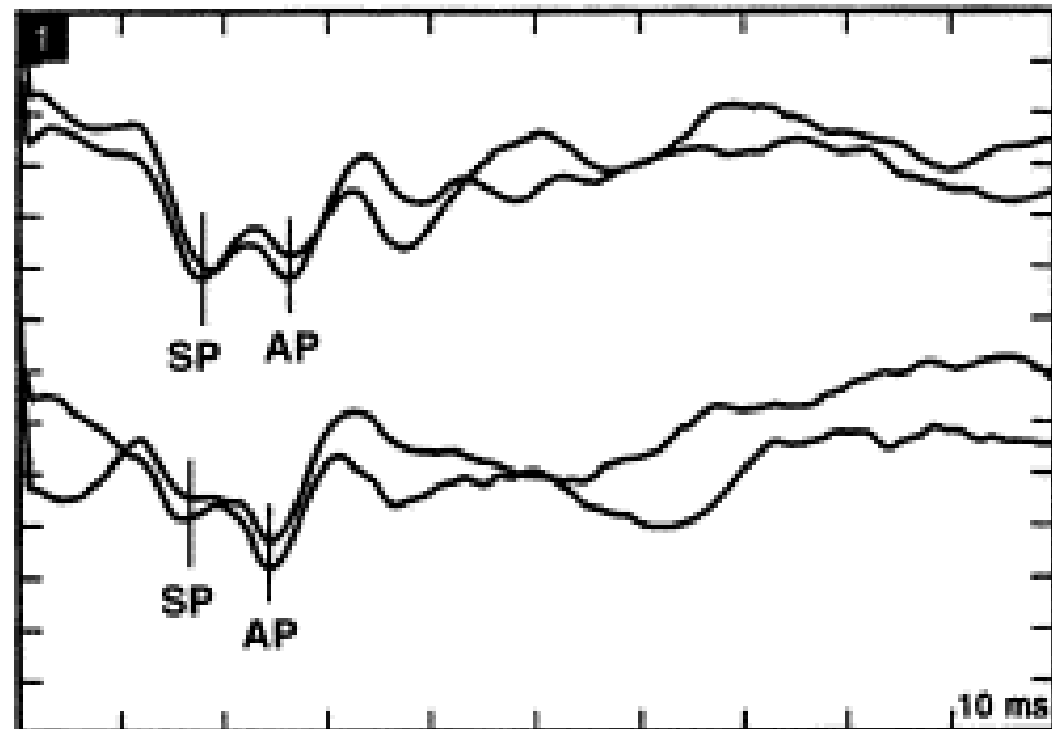
90 dBnHL

ER-3 insert phones

Amplaid MK 22

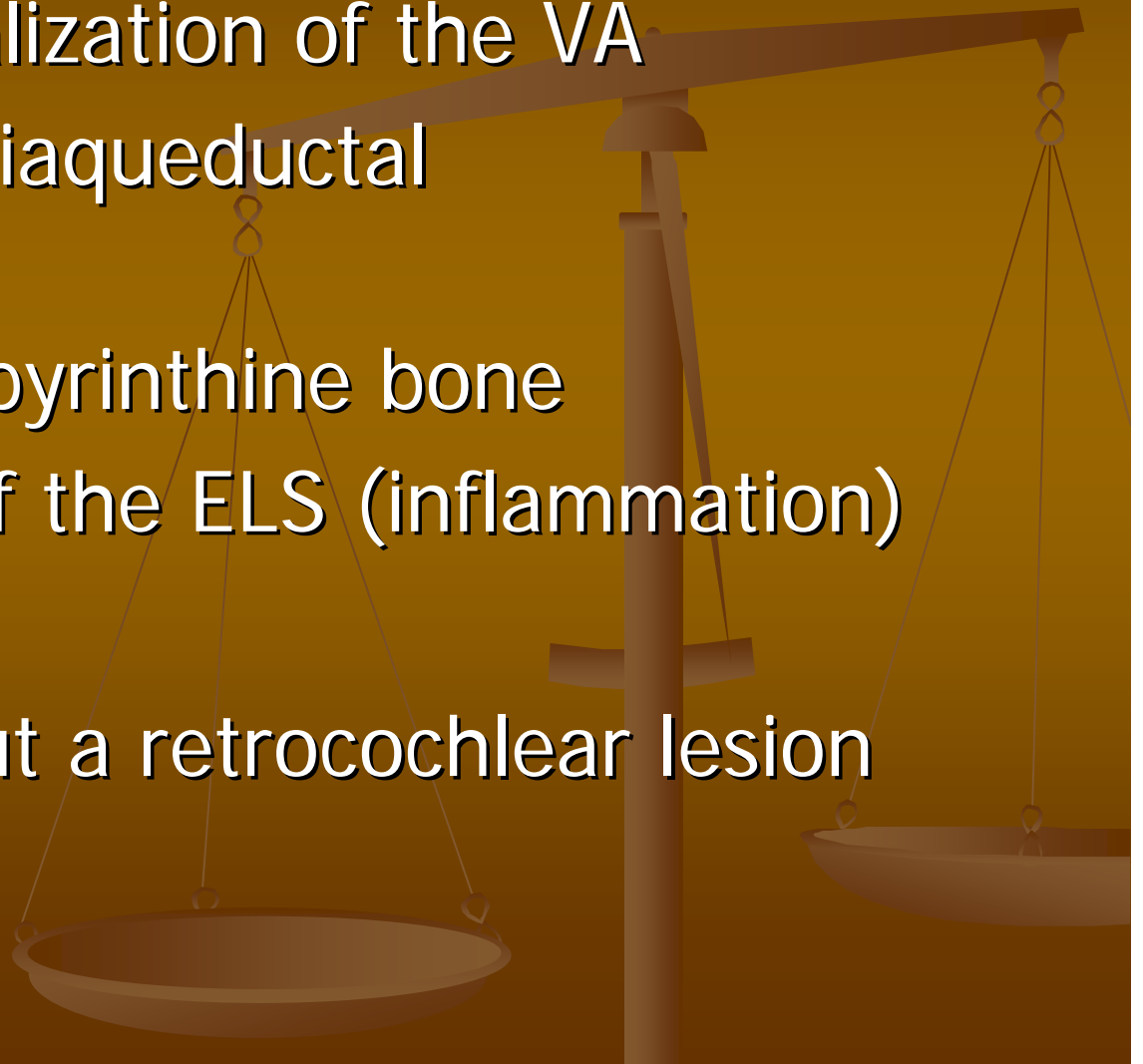
Both traces show hydropic ears.

Upper trace: more affected ear



# ***Imaging of the temporal bone***

- decreased visualization of the VA
- reduction in periaqueductal pneumatization
- reduced retrolabyrinthine bone
- Enhancement of the ELS (inflammation)
- imaging Rule out a retrocochlear lesion







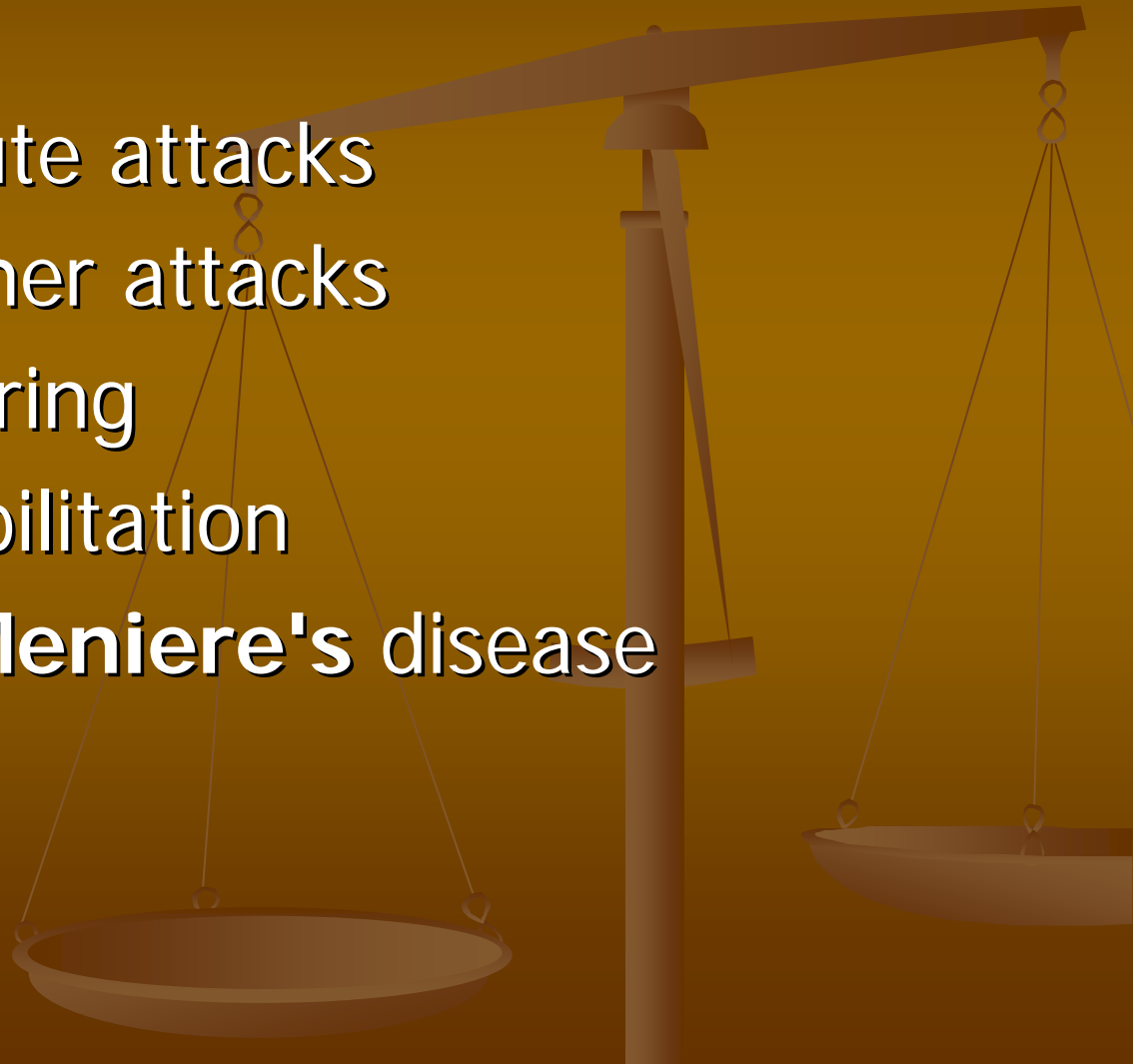
# *Objectives*

- History
- Incidences
- Pathology
- Diagnosis
- *Treatment*



# ***Treatments***

- Education
- To treat the acute attacks
- To prevent further attacks
- To improve hearing
- Vestibular rehabilitation
- F/U.. bilateral **Meniere's** disease



# ***Acute attacks***

- prevent falls
- head should be restricted
- Anticholinergics
- Antihistamines
- Phenothiazine
- Benzodiazepines



# Meniere's Treatment

## Diet control

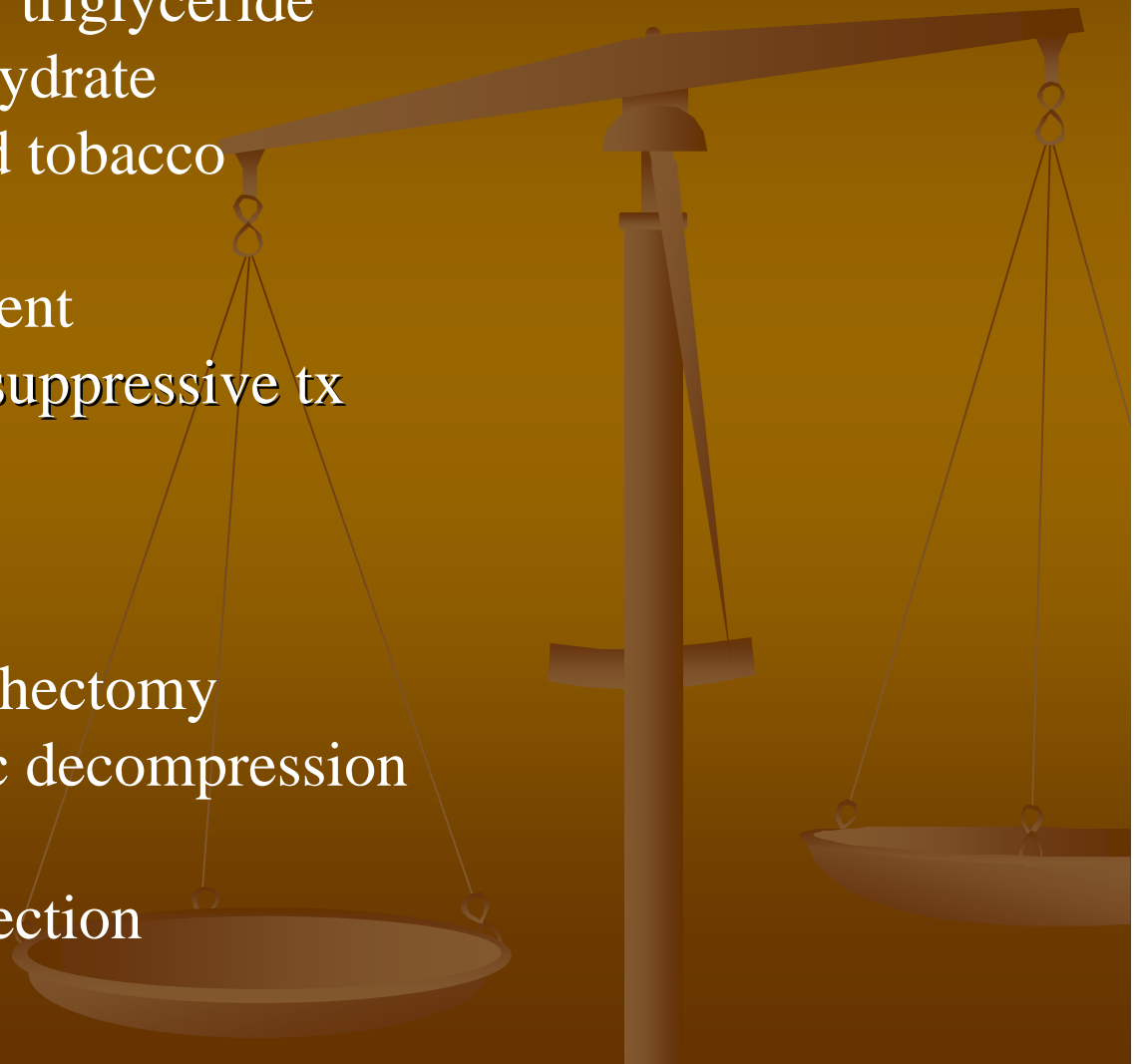
- a. Low salt diet
- b. Low cholesterol or triglyceride
- c. Avoid high carbohydrate
- d. Avoid caffeine and tobacco

## Medical treatment

- a. Antivertiginous agent
- b. Steroids/ Immunosuppressive tx
- c. Diuretics
- d. Vasodilators

## Surgical treatment

- a. Chemical Labyrinthectomy
- b. Endolymphatic sac decompression
- c. Labyrinthectomy
- d. Vestibular nerve section

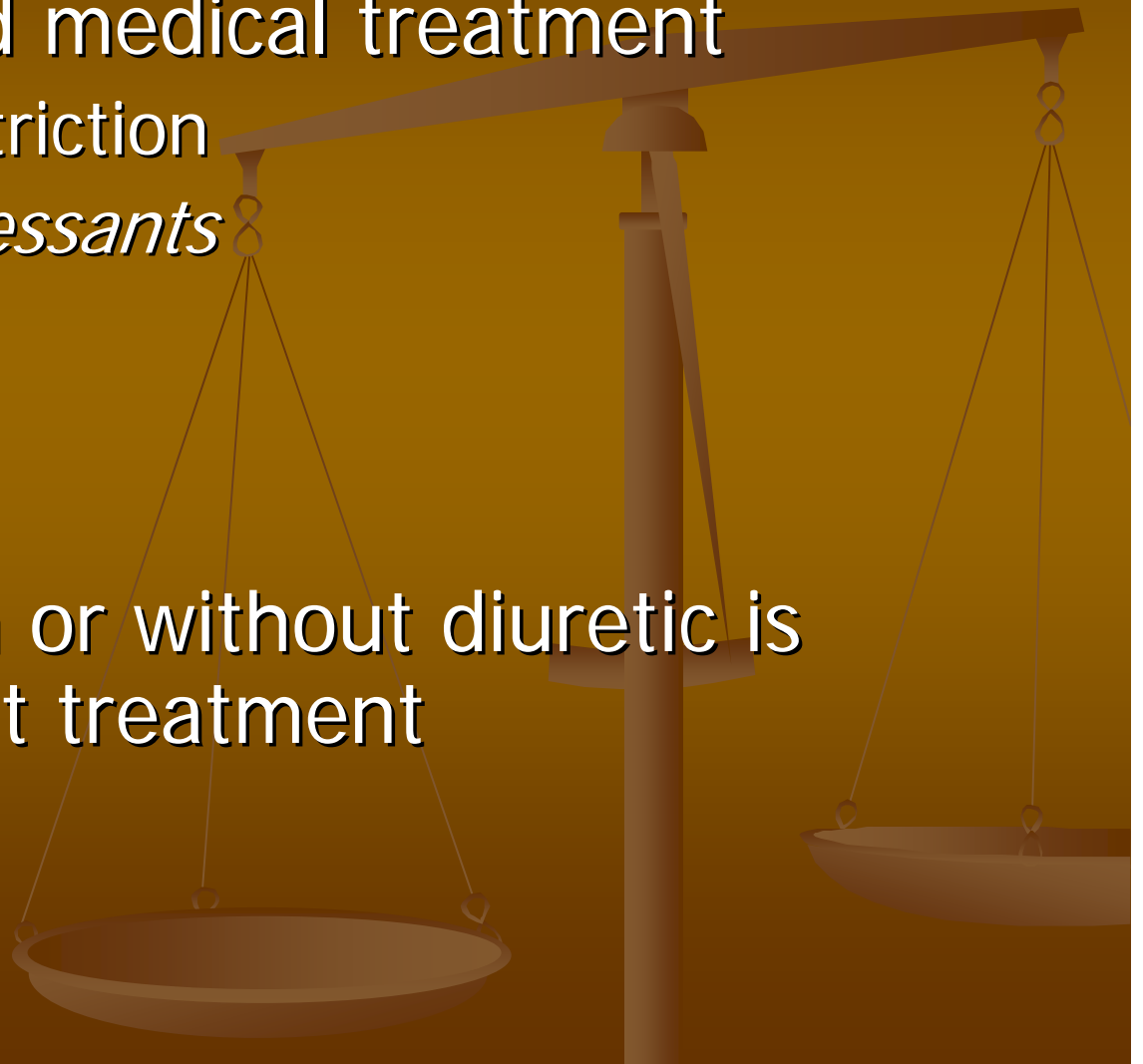




# ***Medical TREATMENT***

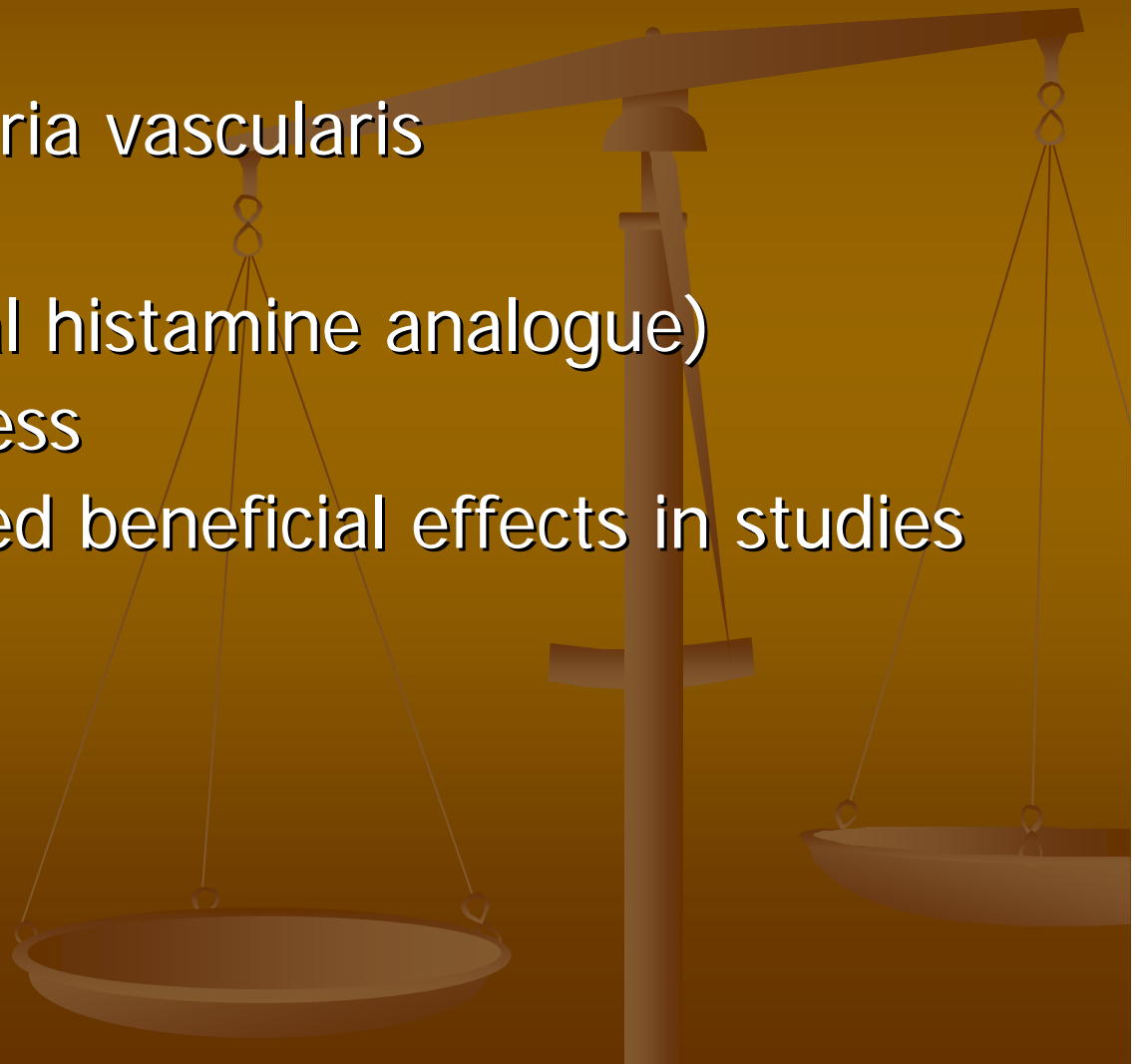
# *Meniere's Disease*

- Widely accepted medical treatment
  - Dietary salt restriction
  - *Vestibulosuppressants*
  - Vasodilators
  - Diuretics
- Betahistine with or without diuretic is favoured current treatment



# *Meniere's Disease*

- Vasodilators
  - ? ischemia of stria vascularis
  - IV histamine
  - betahistine (oral histamine analogue)
  - Anecdotal success
  - No demonstrated beneficial effects in studies





# *Vestibulosuppressants*

- Meclizine (Antivert, Marezine, Meni-D)
  - Decreases excitability of labyrinth
  - blocks conduction in vestibular-cerebellar pathways.
  
- *Benzodiazepines*
  - *Effective in panic attacks*
  - *depresses all levels of CNS, including limbic and reticular formation*

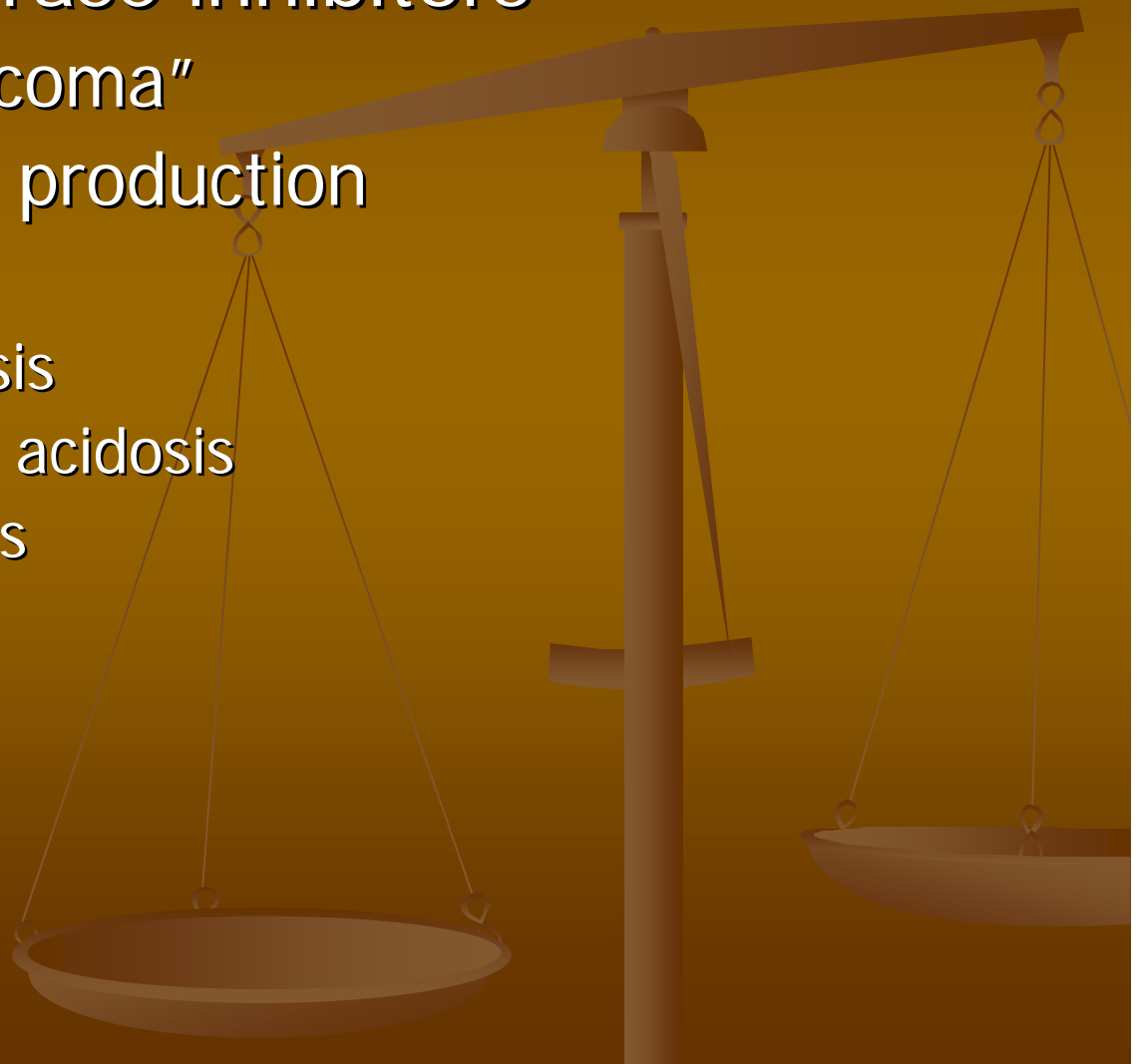
# *Meniere's Disease*

- Thiazide diuretics
  - Side effects - hypokalemia, hypotension, hyperuricemia, hyperlipoproteinemia
  - conducted using double-blind methodology, have shown no effect of diuretics\*

\*ORL J Otorhinolaryngol Relat Spec 1986; 48:287-292

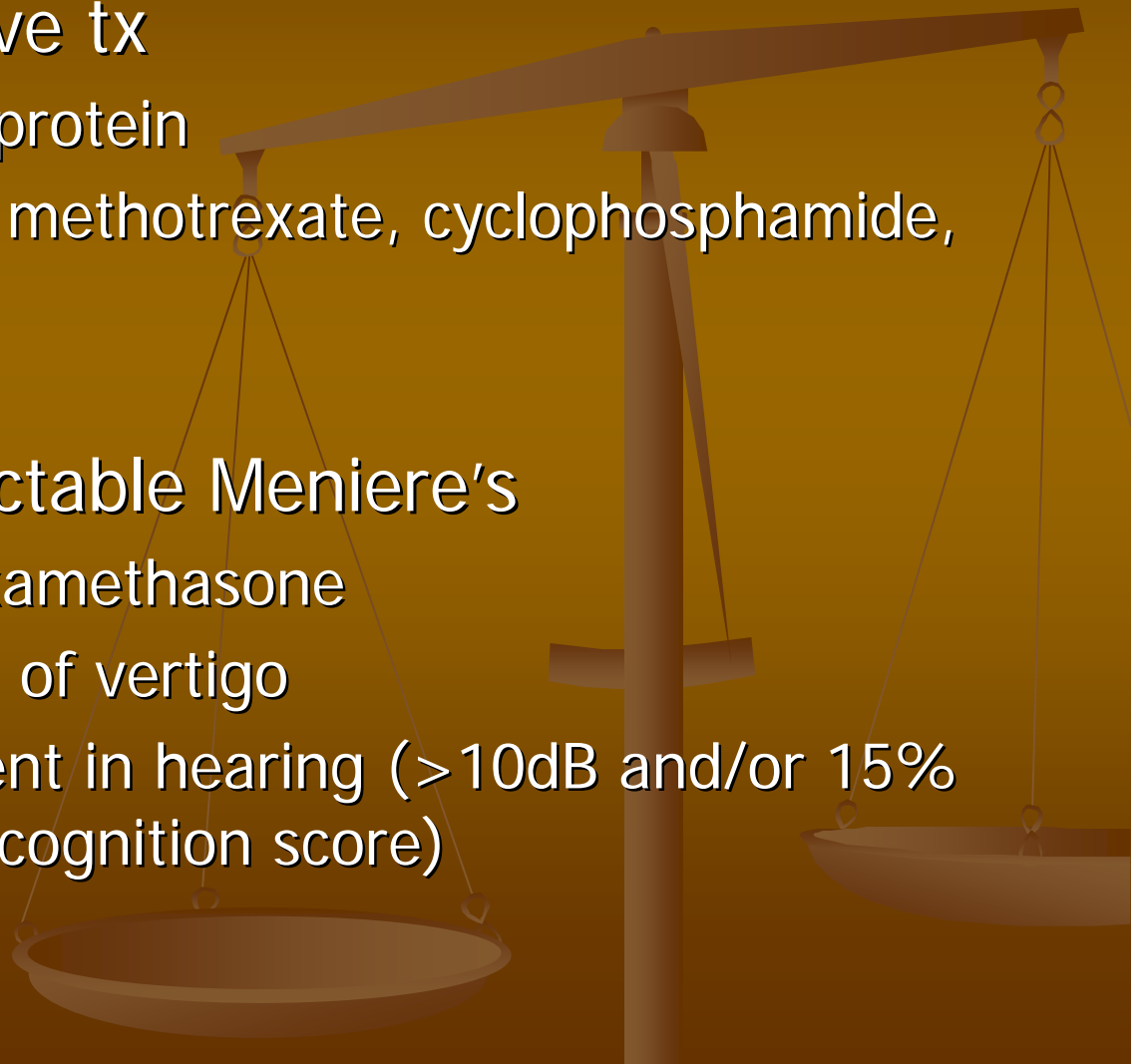
# *Meniere's Disease*

- Carbonic anhydrase inhibitors
  - "inner ear glaucoma"
  - Decreased CSF production
  - Side effects
    - Nephrocalcinosis
    - mild metabolic acidosis
    - GI disturbances



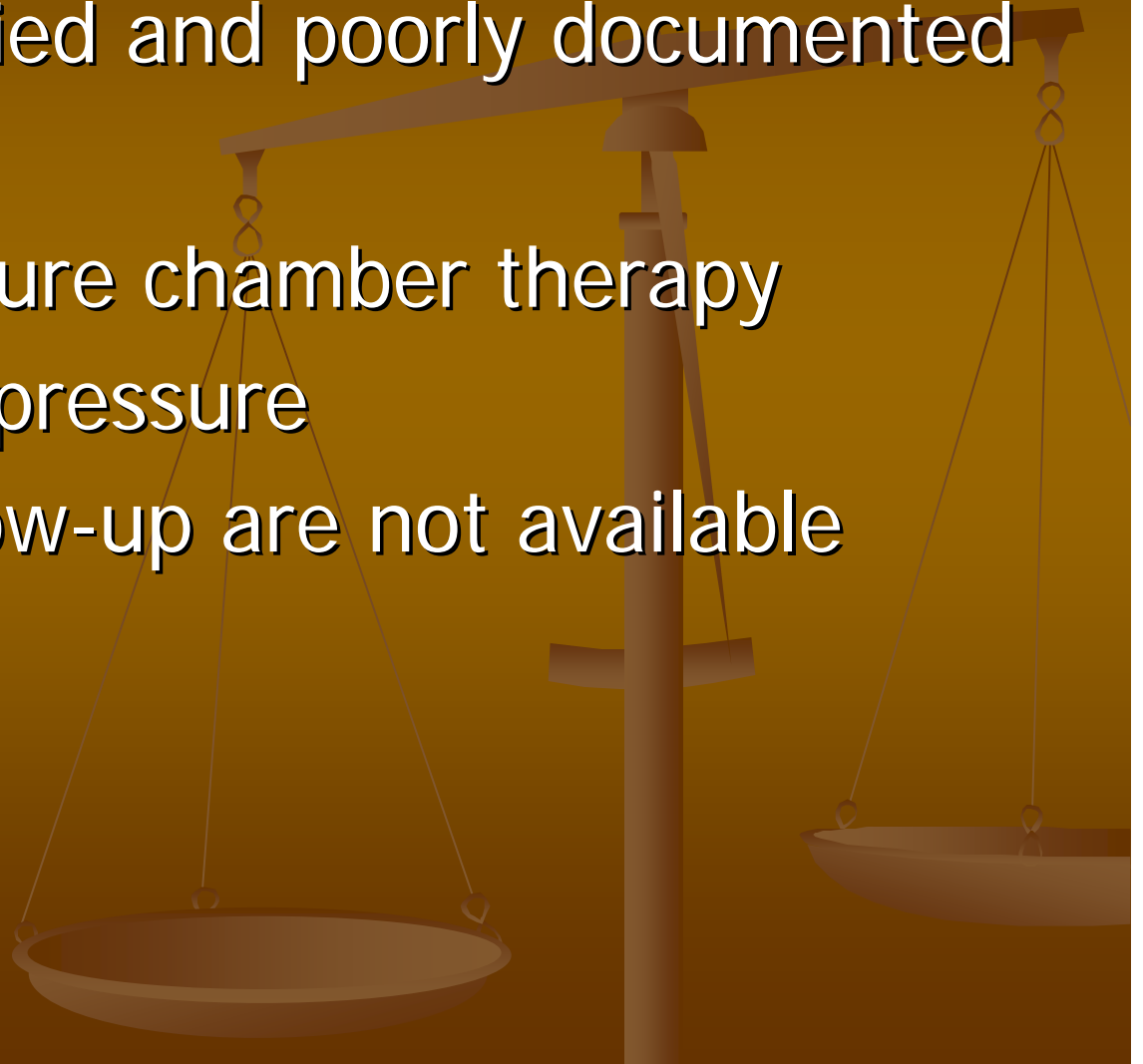
# *Meniere's Disease*

- Immunosuppressive tx
  - 70 kd heat-shock protein
  - systemic steroids, methotrexate, cyclophosphamide, IT steroids
- Shea study - intractable Meniere's
  - 48 patients IT dexamethasone
  - 66.7% elimination of vertigo
  - 35.4% improvement in hearing (>10dB and/or 15% change in word recognition score)



# Other therapies

- empirically applied and poorly documented
- acupuncture
- hypobaric pressure chamber therapy
- pulsed positive pressure
- Long-term follow-up are not available

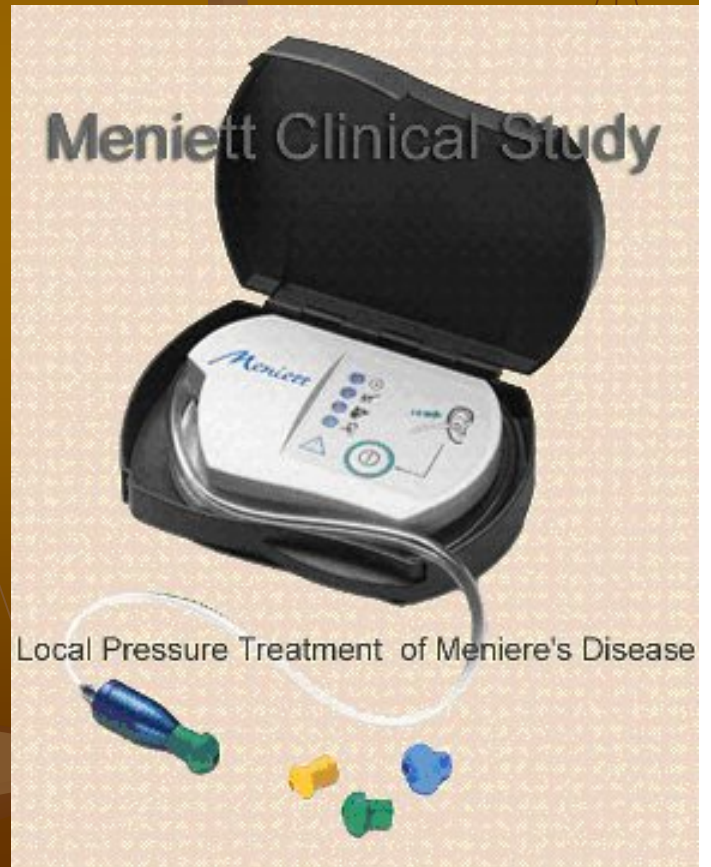


# Meniett

- 51 % free from vertigo spells
- 41 % significant decrease in frequency of  
→ No medications.
- 8 % not respond
- No side effects

<http://www.meniett.com/>

<http://www.midwestear.com/meniett.htm>



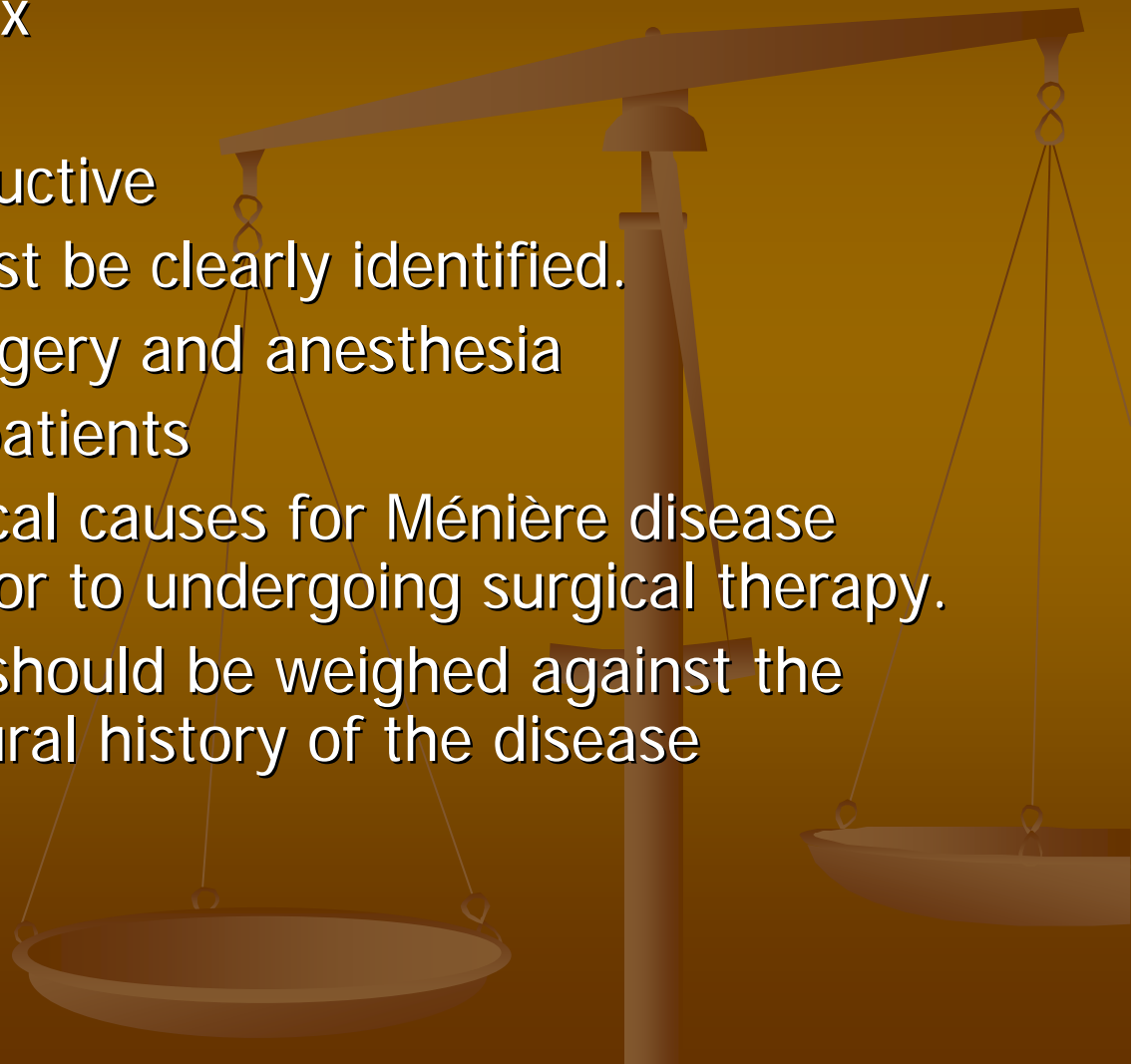


**Sx**

***TREATMENT***

# *Sx TREATMENT*

- Failed conservative Rx
- ENG –other side-
- Start with least destructive
- The diseased ear must be clearly identified.
- able to withstand surgery and anesthesia
- severely debilitated patients
- Any underlying medical causes for Ménière disease should be treated prior to undergoing surgical therapy.
- The risks of surgery should be weighed against the benefits and the natural history of the disease



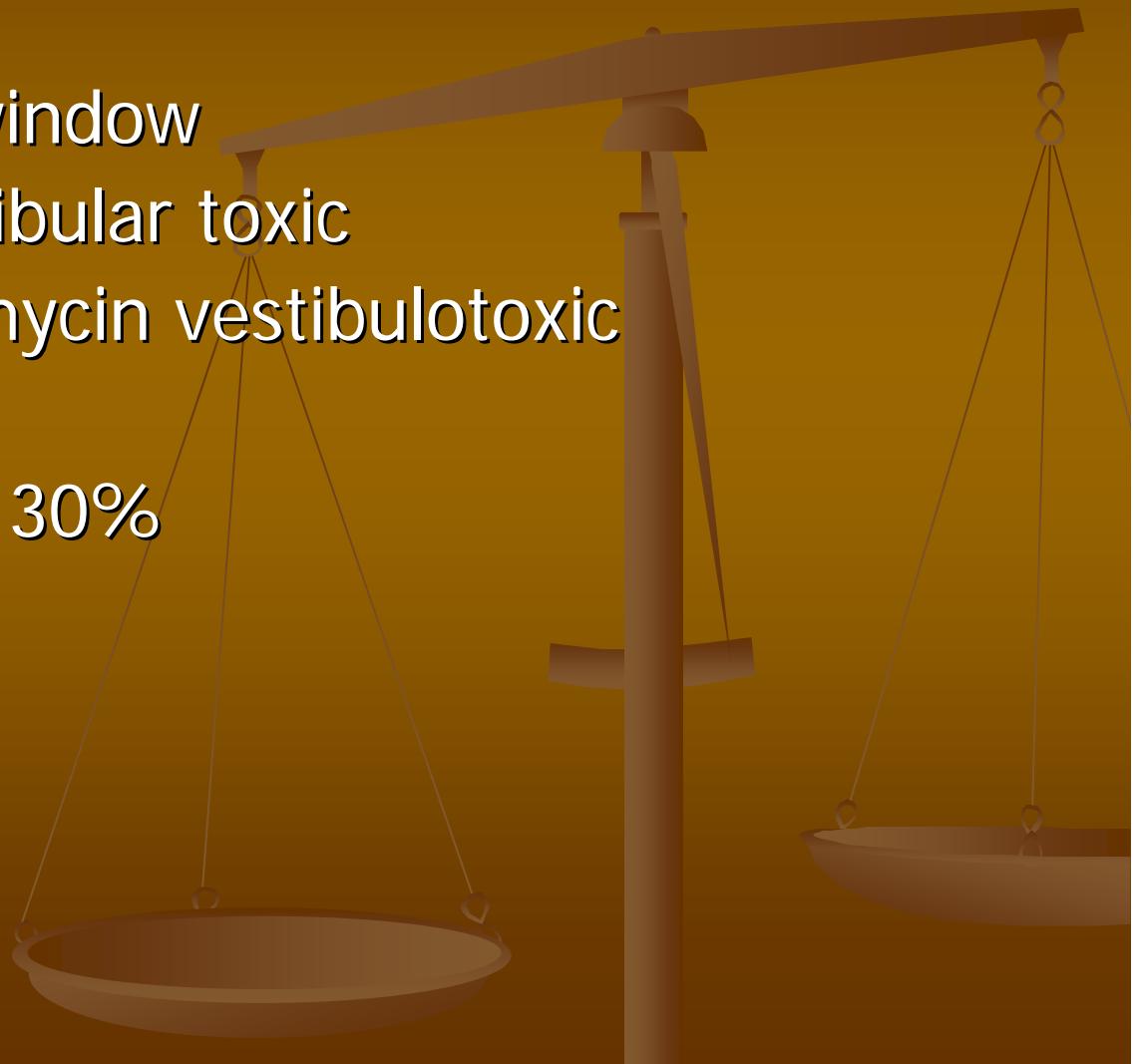


# ***Chemical Labrynthectomy***



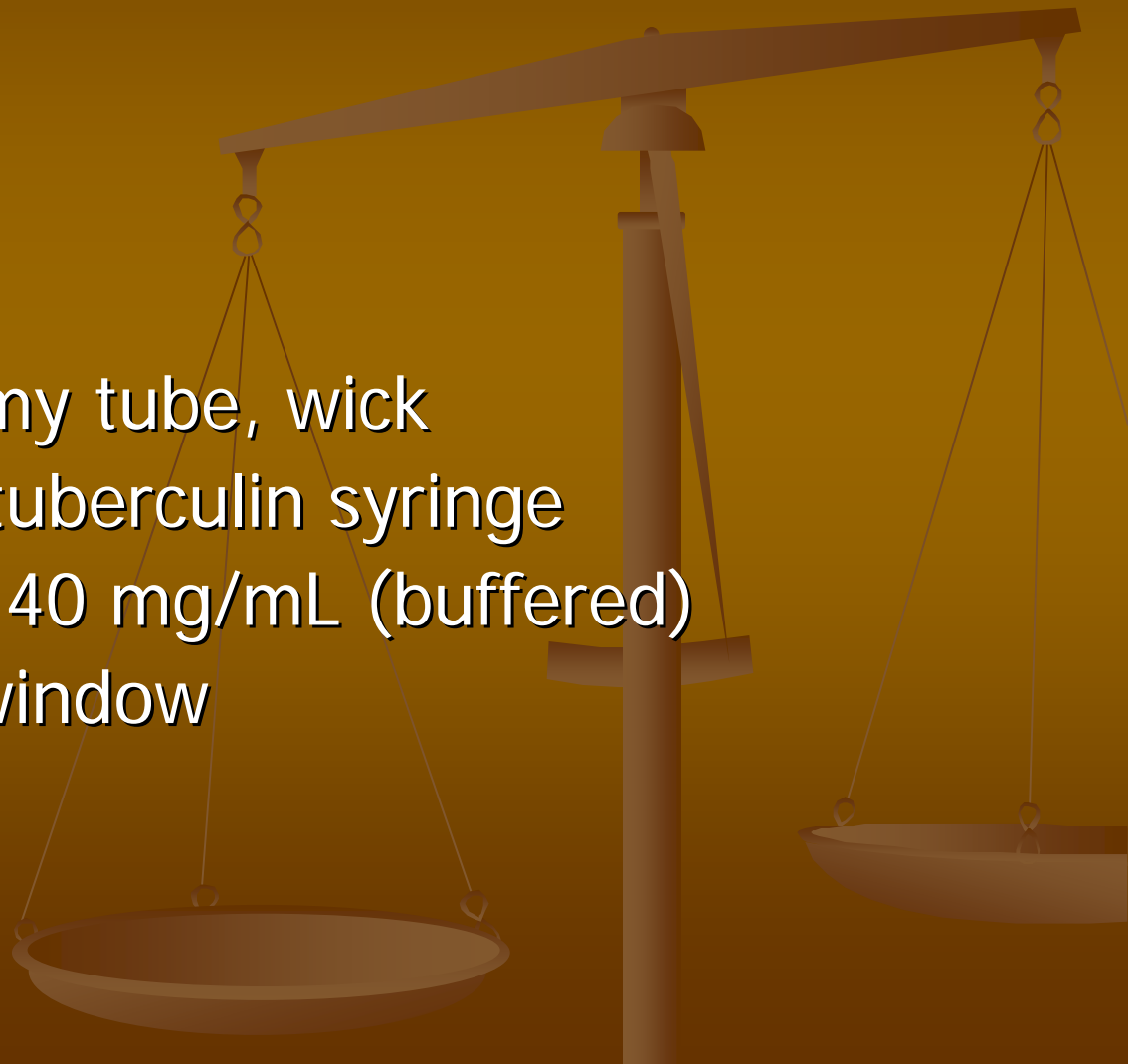
# ***Chemical Labyrinthectomy***

- Schuknecht 1956
- Absorbed round window
- Cochlear and vestibular toxic
- Gent and streptomycin vestibulotoxic
- Many regimens
- hearing loss risk - 30%



# Chemical Labyrinthectomy

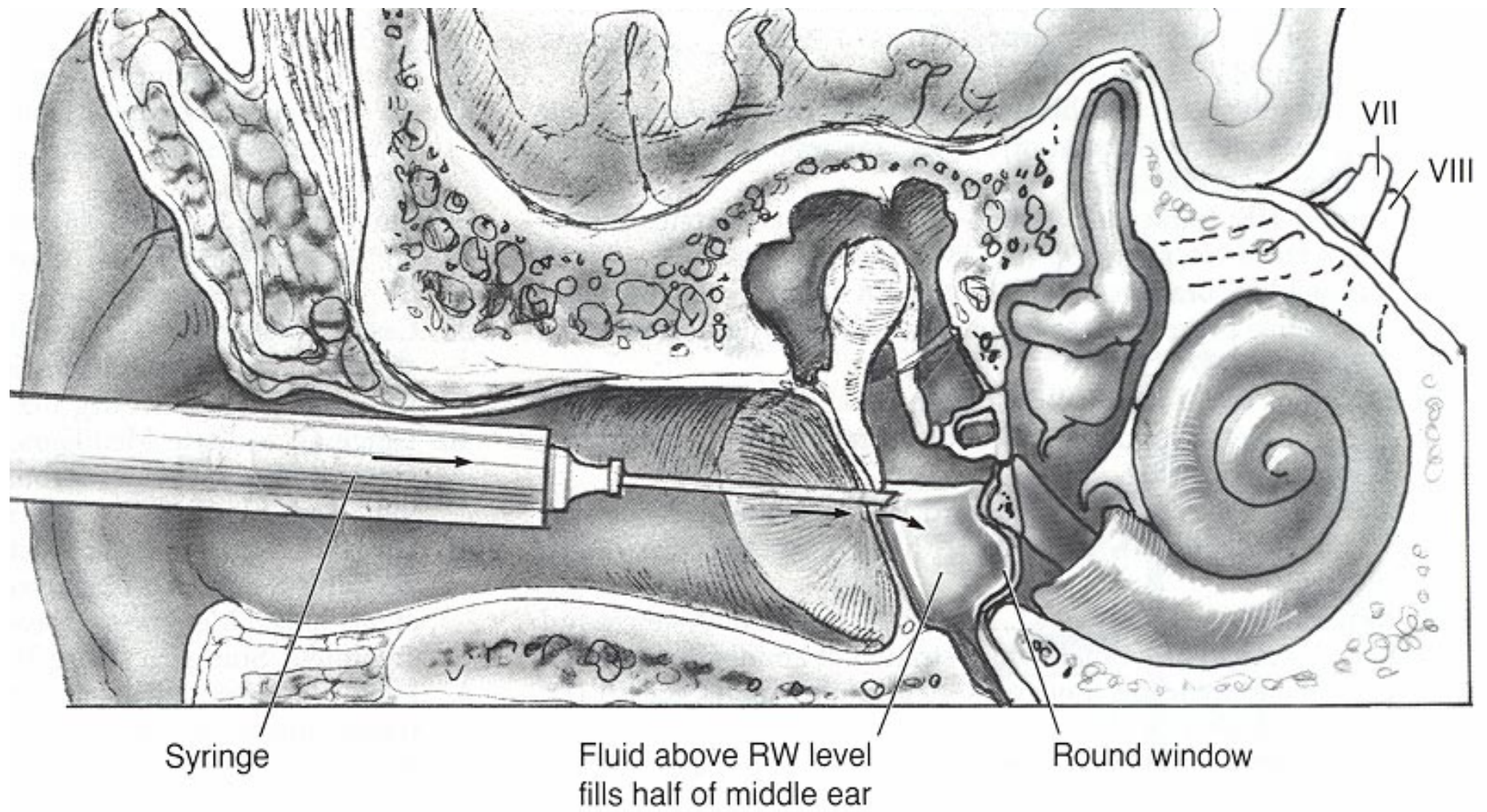
- Office procedure
- Anesthesia
  - Injectable local
  - Emla
  - Phenol
- + / - Tympanostomy tube, wick
- 25 guage needle, tuberculin syringe
- 0.5 - 0.75 ml gent 40 mg/mL (buffered)
- Submerge round window
- 30-45 min
- No swallowing



# ITAG

- IV solution - 40mg/mL gentamicin
- 10 to 20 mg injected over round window
- ***Instructed not to swallow***
- Bolus injections - weekly or bi-weekly
- End point variable - vestibular hypofunction
- Audiometry monitoring between injections
- Total vestibular ablation not necessary

# ITAG



# ITAG

TABLE 42-1. Review of Literature on Intratympanic Aminoglycoside Therapy

AUTHOR AND REFERENCE	NUMBER OF PATIENTS TREATED	AMINO-GLYCOSIDE	DOSAGE	TREATMENT END POINT	CONTROL OF VERTIGO (%)	LOSS OF CALORIC RESPONSE (%)	HEARING PRESERVED (%)	TINNITUS DISAPPEARED OR IMPROVED (%)	AURAL FULLNESS IMPROVED (%)
Schuknecht <sup>14</sup>	8	Streptomycin	50-300mg/dose 350-600mg total dose	Vestibular ablation	63	63	37	NR	NR
Beck and Schmidt <sup>43</sup>	43	Gentamicin	30mg/day	Vestibular ablation	91	NR	42	86	95
	40	Gentamicin, 40mg/ml	"6 doses planned"	First ototoxic reaction	92	0	85	95	100
Lange <sup>42</sup>	Total 83 92	Streptomycin Tobramycin Gentamicin, 40mg/ml	60mg/day, "typically several days"	First ototoxic reaction	90	NR	76	35	43
Moller et al. <sup>46</sup>	15	Gentamicin 30mg/ml	15-30mg/dose 1-11 doses, mean = 5	First ototoxic reaction	93	100	66	82	78
Sala <sup>47</sup>	62	Gentamicin 30mg/ml	Up to 30mg/day 1-8 doses, mean = 3.5	First ototoxic reaction	86	51	70	76	78
Blessing and Schlenter <sup>48</sup>	82	Gentamicin 40mg/ml	1 or 2/day 5-40mg/dose × 7 days	Ablative nystagmus or hearing loss	89	28	67	NR	NR
Laitakari <sup>49</sup>	20	Gentamicin 40mg/ml	0.2ml/day × 3 days, then 0.2ml qod 3-12 doses, mean = 5.3	Ablative nystagmus	90	70	55	NR	NR
Nedzelski et al. <sup>45</sup>	20	Gentamicin 26.7mg/ml pH 6.4	0.65ml tid × 4 days 52mg/dose × 4 = 208 mg	12 doses or first ototoxic reaction	90	85	80	NR	NR
Magnusson and Padoan <sup>44</sup>	5	Gentamicin 30mg/ml pH 6.4	30mg/ml bid for 1 day	2 doses	100	100	100	0	NR

bid = twice a day; NR = not reported; qod = every other day; tid = three times a day.



# Intratympanic gentamicin for Meniere's disease: a meta-analysis

Rutka JA et al\*

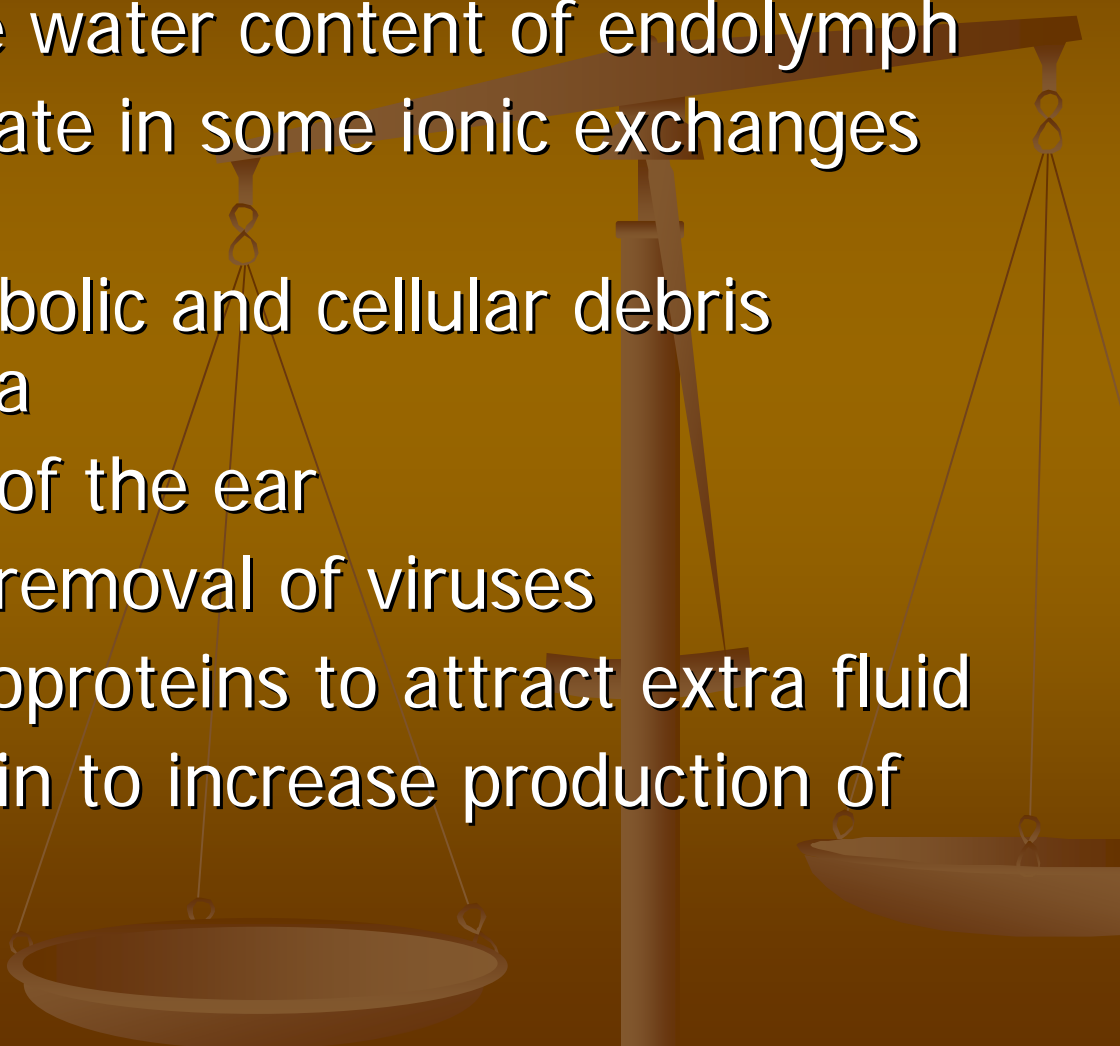
- Appears to be effective in the relief of vertigo.
- Cochleotoxicity and ototoxicity is unlikely to be a major side effect.
- Insufficient evidence from the eligible articles
- titrated with low-dose gentamicin.
- Further investigation with this treatment modality with control subjects is warranted



# ***ELS TREATMENT***



# ***Endolymphatic sac functions***

1. Resorption of the water content of endolymph
  2. Ability to participate in some ionic exchanges with endolymph
  3. Removal of metabolic and cellular debris including otoconia
  4. Immunodefense of the ear
  5. Inactivation and removal of viruses
  6. Secretion of glycoproteins to attract extra fluid
  7. Secretion of sacsin to increase production of endolymph
- 

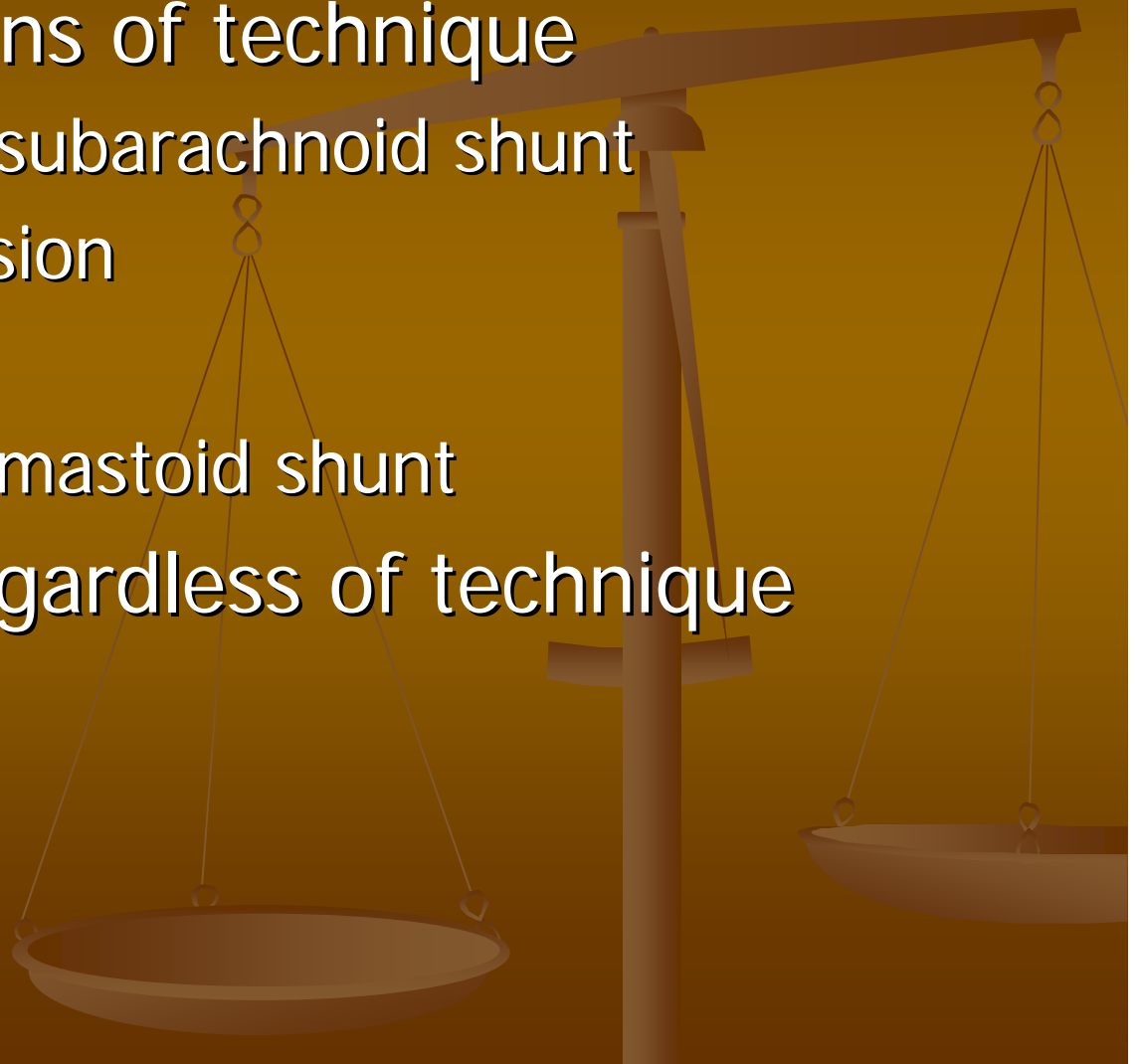
# *Choice*

- Young
- Bilateral
- Normal hearing
- Only hearing ear



# *Endolymphatic Sac Procedures*

- Multiple variations of technique
  - Endolymphatic-subarachnoid shunt
  - Sac decompression
  - Sac excision
  - Endolymphatic-mastoid shunt
- 75% success regardless of technique



# *Endolymphatic Sac Procedures*

- No controlled studies
  - Difficulty in finding control group
  - Unpredictability of natural course
- Bretlau, Thomsen\* et. al. 1981
  - Prospective, blinded
  - Simple mastoid vs. active mastoid shunt
  - no difference in vertigo control yearly for up to 9 y
- \*\*Thomsen
  - Shunt vs tympanostomy tubes
  - No difference

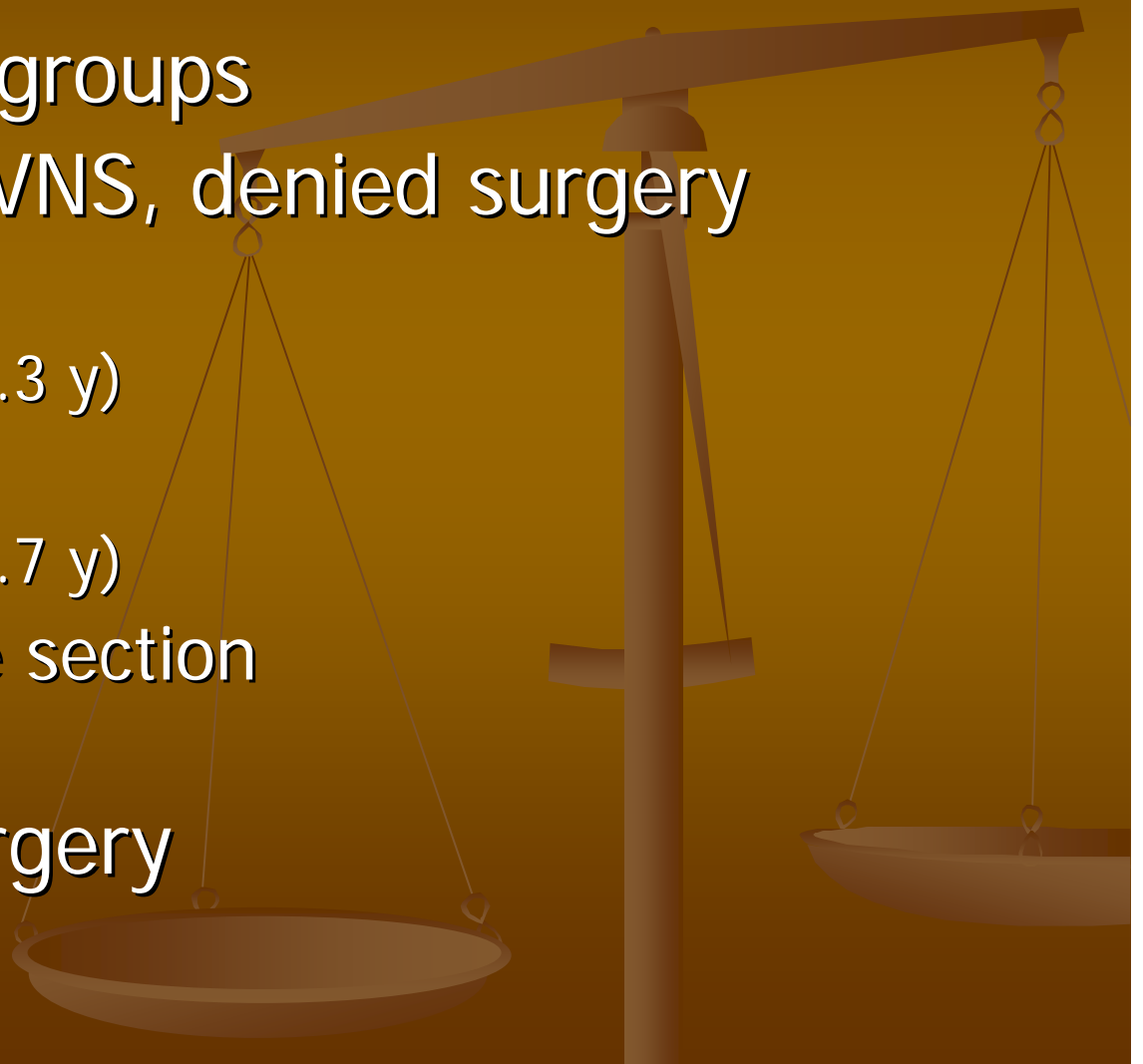
\*Ann N Y Acad Sci. 1981;374:820-30

\*\*Acta Otolaryngol. 1998 Nov;118(6):769-73

# *Endolymphatic Sac Procedures*

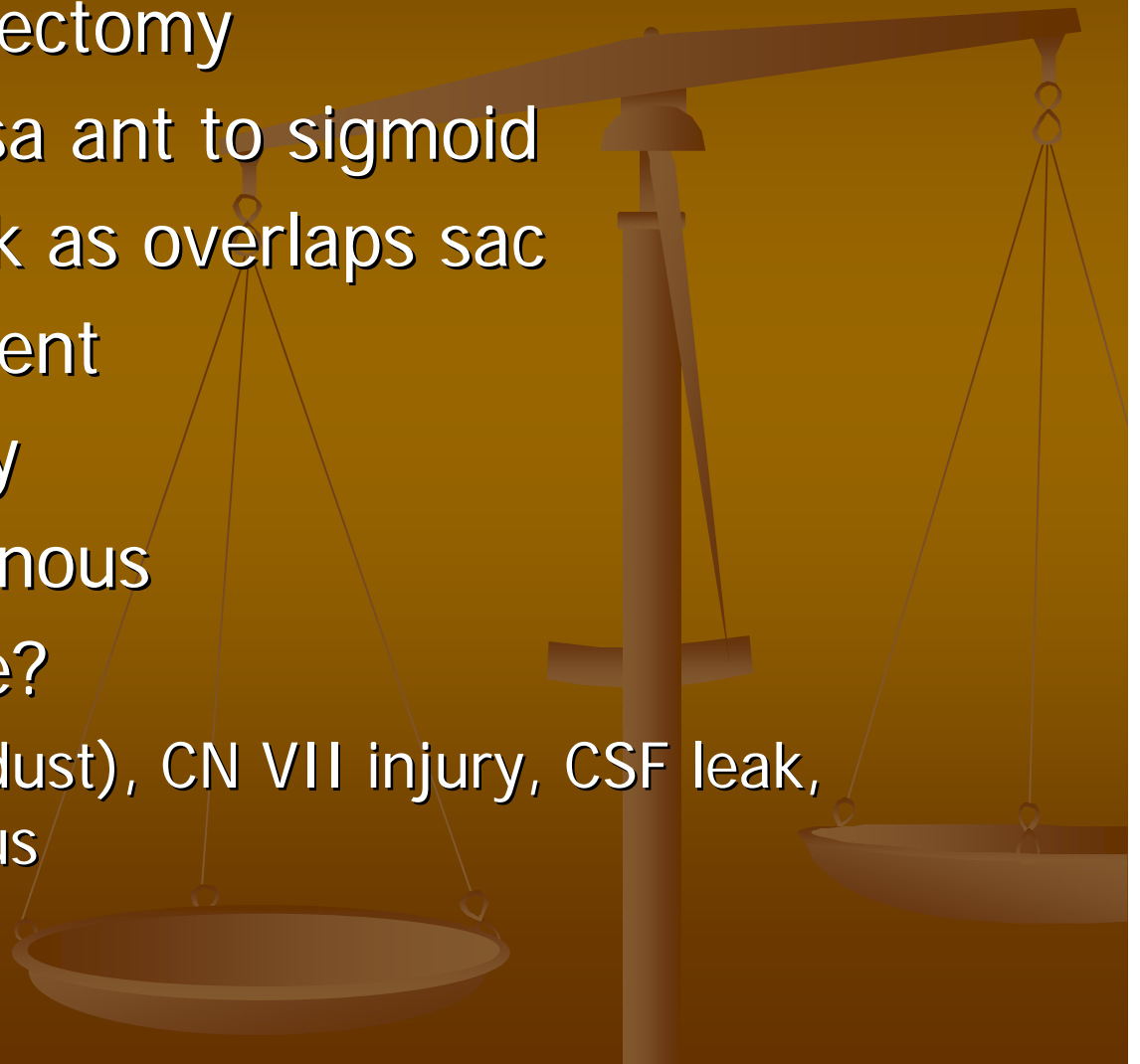
Silverstein et. al.

- Retrospective 3 groups
- 2 years : ESSx, VNS, denied surgery
  - Controls
    - 57% (71% at 8.3 y)
  - Sac surgery
    - 40% (70% at 8.7 y)
  - Vestibular nerve section
    - 93%
- ? Benefit sac surgery

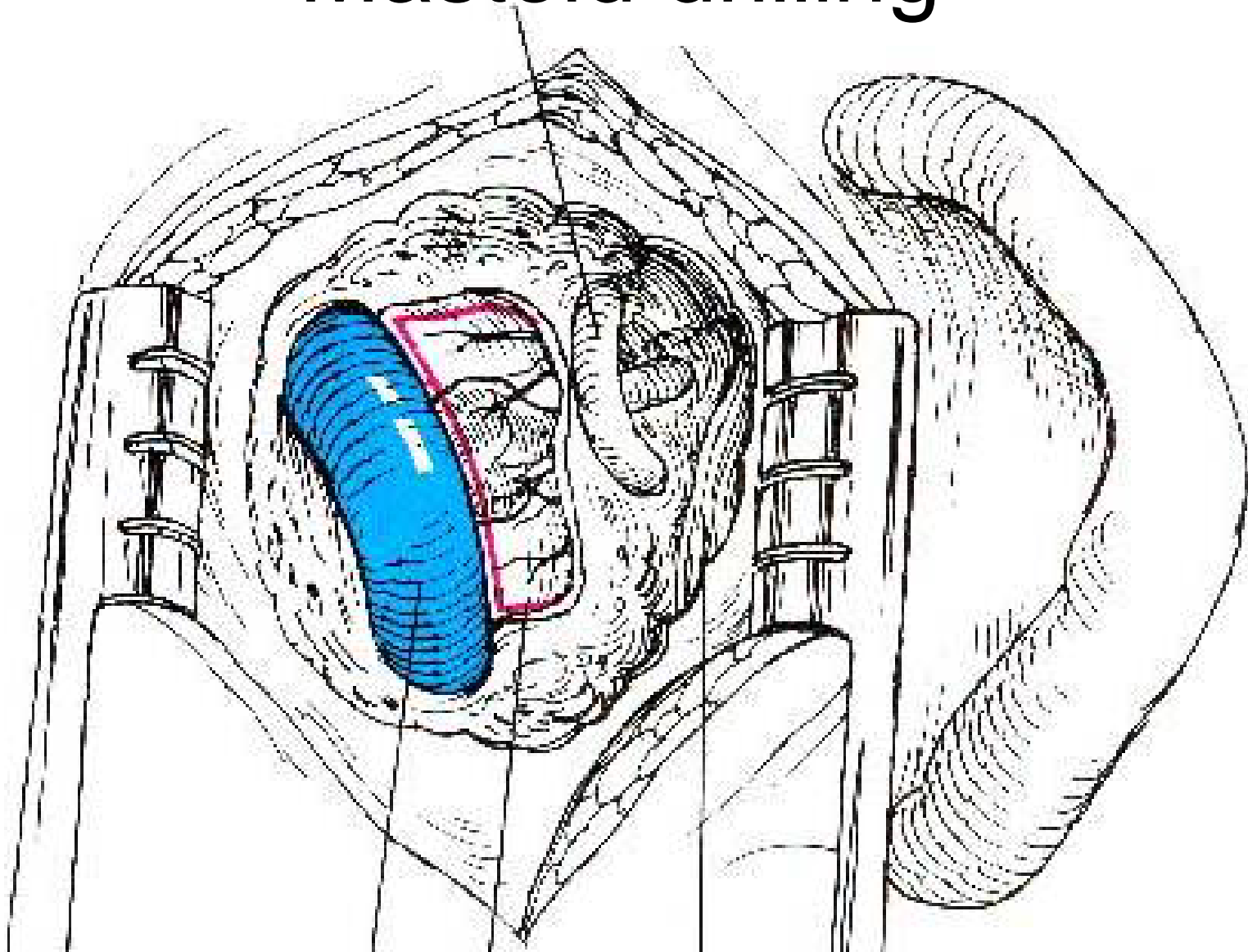


# *Endolymphatic Sac Procedure*

- Complete mastoidectomy
- All bone post. fossa ant to sigmoid
- Dura appears thick as overlaps sac
- Open, excise or stent
- Outpatient surgery
- Usually not vertiginous
- Complications rare?
  - SNHL, CHL(bone dust), CN VII injury, CSF leak, bleeding from sinus



# Mastoid drilling



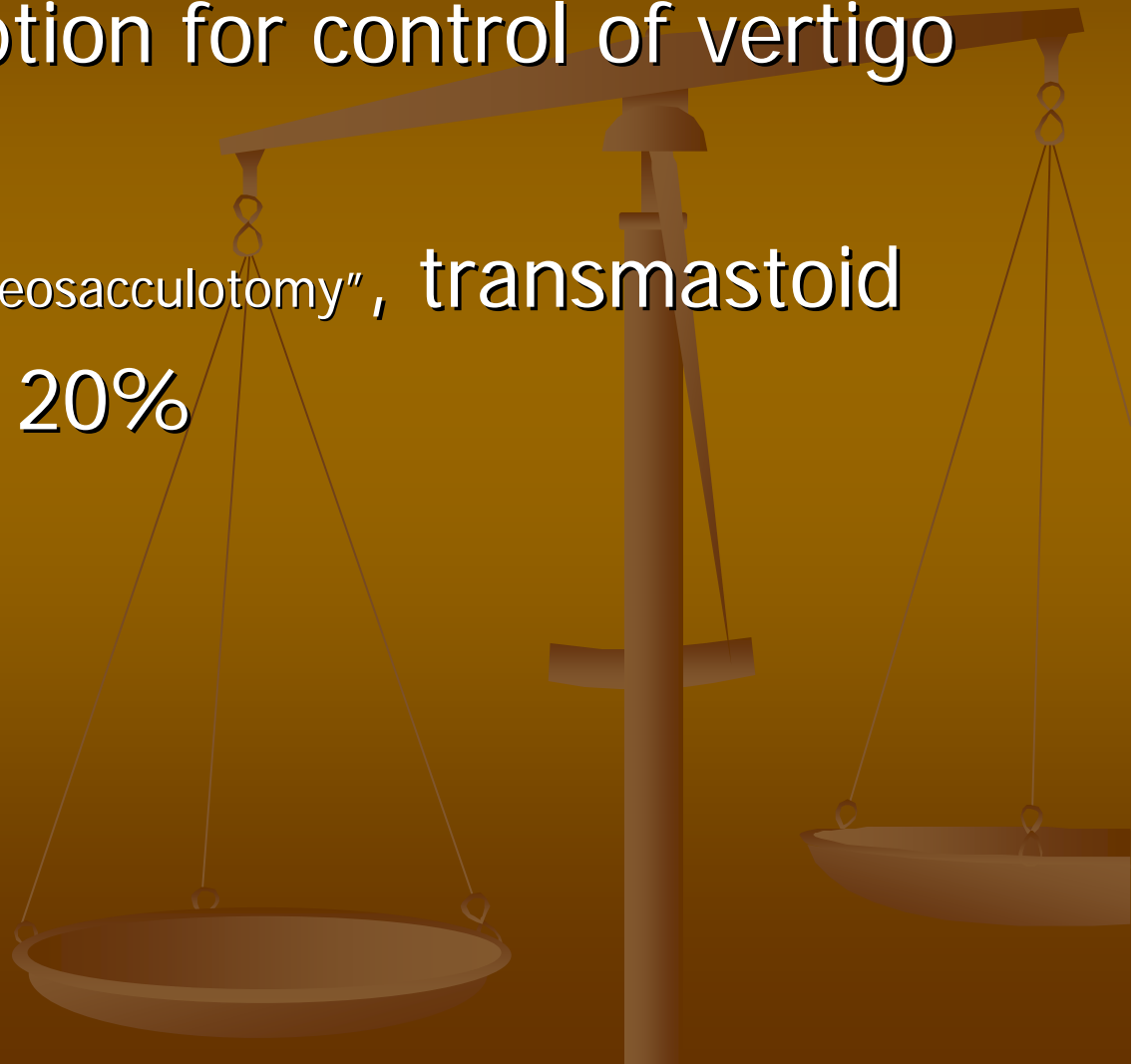
# ***Labrynthectomy***





# ***Labyrinthectomy***

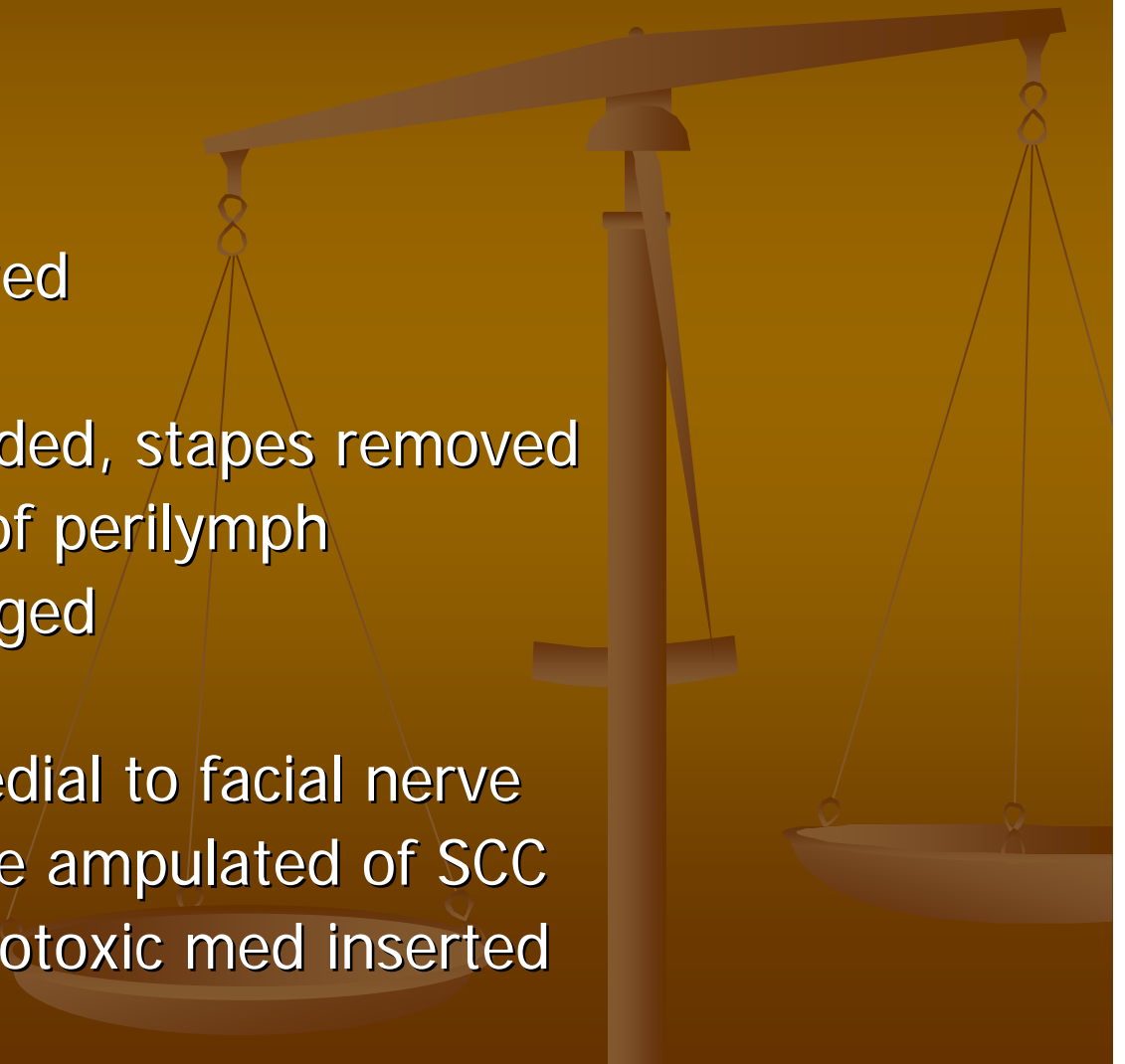
- Final surgical option for control of vertigo
- 1904 described
- Transcanal "Cochleosacculotomy", transmastoid
- PTA 70, discrim 20%



# ***Labyrinthectomy***

## ■ Transcanal

- Local or general
- Tympanomeatal flap
- IS joint disarticulated
- Incus removed
- Stapes tendon divided, stapes removed
- Vestibule drained of perilymph
- Oval window enlarged
- Sacculle removed
- Utricle superior medial to facial nerve
- Hook used to probe amputated of SCC
- Gelfoam soaked ototoxic med inserted

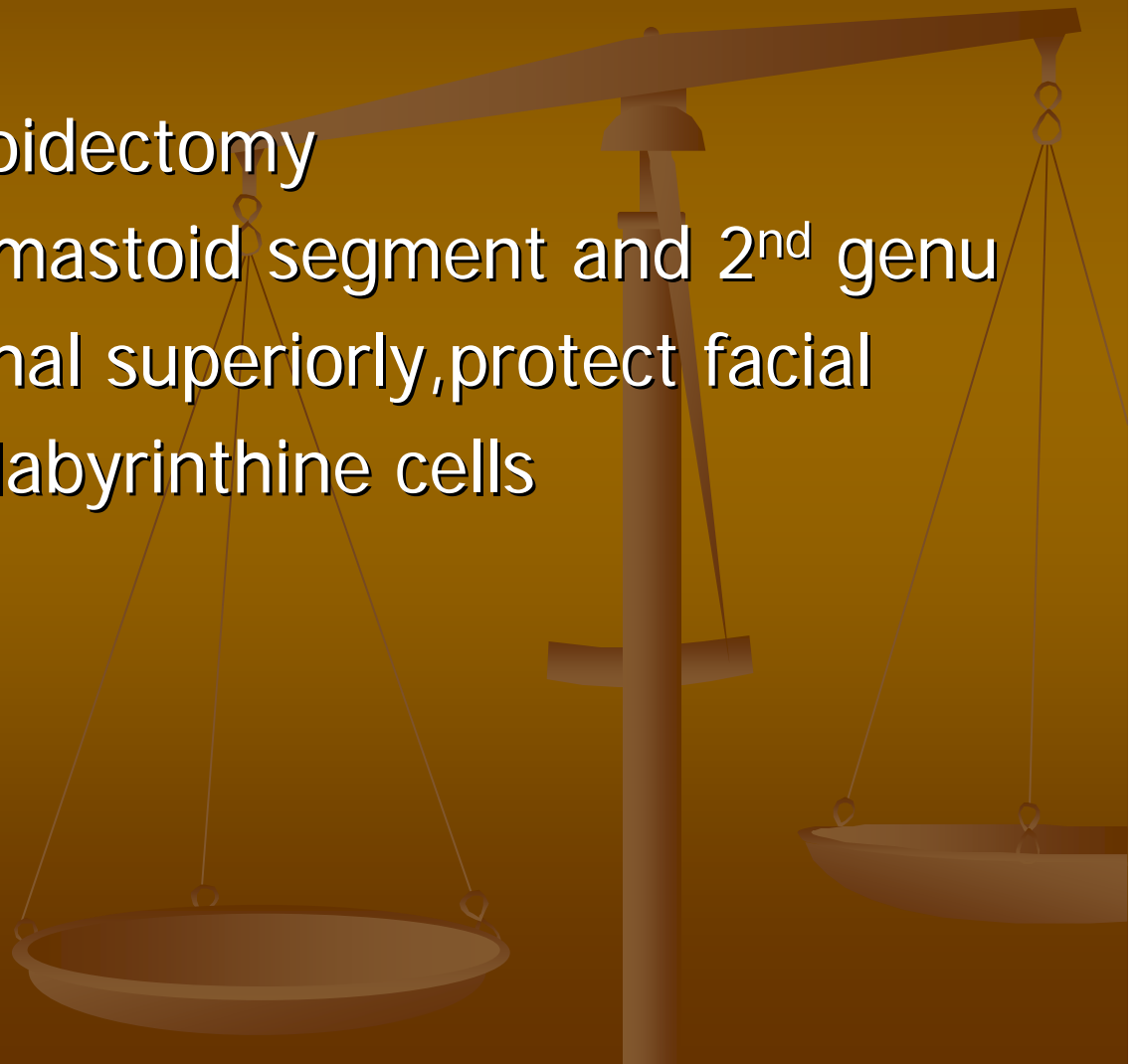


# Cochleosacculotomy



# ***Labyrinthectomy***

- Transmastoid
  - Complete mastoidectomy
  - Visualize facial mastoid segment and 2<sup>nd</sup> genu
  - Enter lateral canal superiorly, protect facial
  - Exenterate perilabyrinthine cells

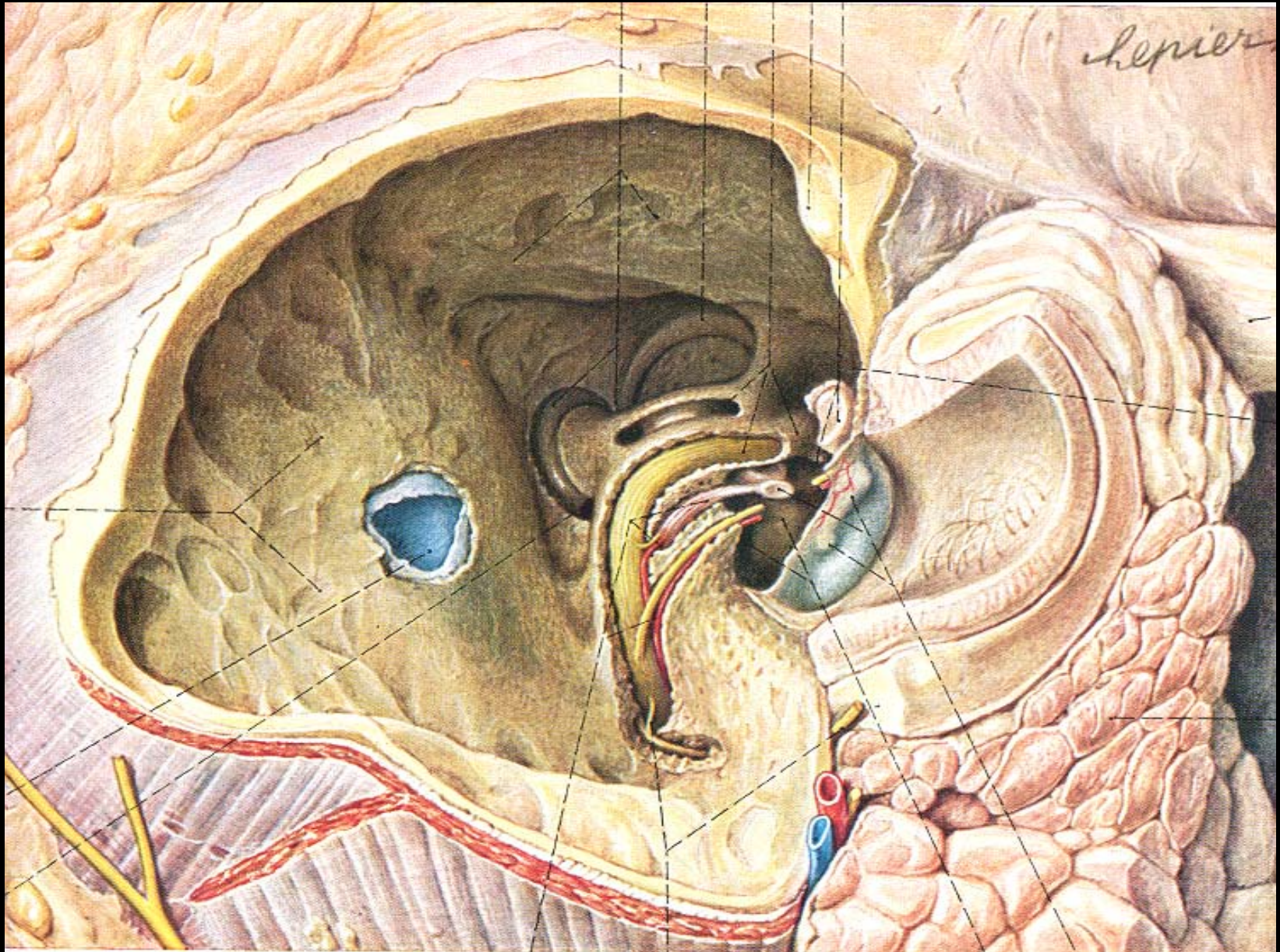


# ***Labyrinthectomy***

- Follow to ampulla located superior to vestibule, and avulse
- Enlarge vestibule and remove utricle and saccule
- Respect lateral wall
- Carry posteriorly medial to second genu to locate PSCC ampulla and remove
- Closed in layer and mastoid dressing




# Labyrinthectomy



# ***Labyrinthectomy***

- Post-operative course
  - Anti-emetics
  - Ambulation
- Results
  - 85% relief of vertigo
- Complications
  - Facial nerve injury
  - CSF leak
  - Vertigo 30%



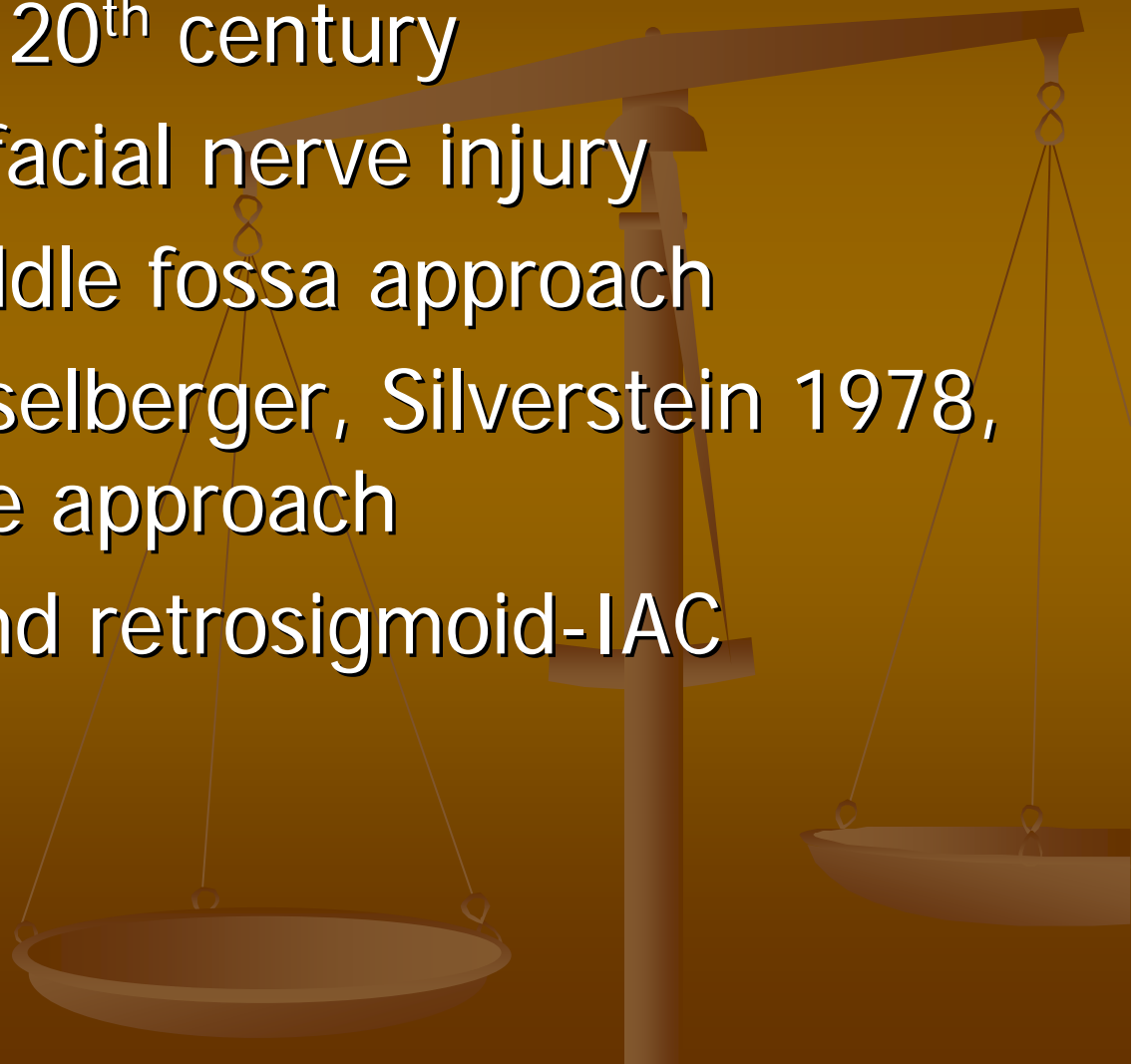


# ***VNS*** ***TREATMENT***



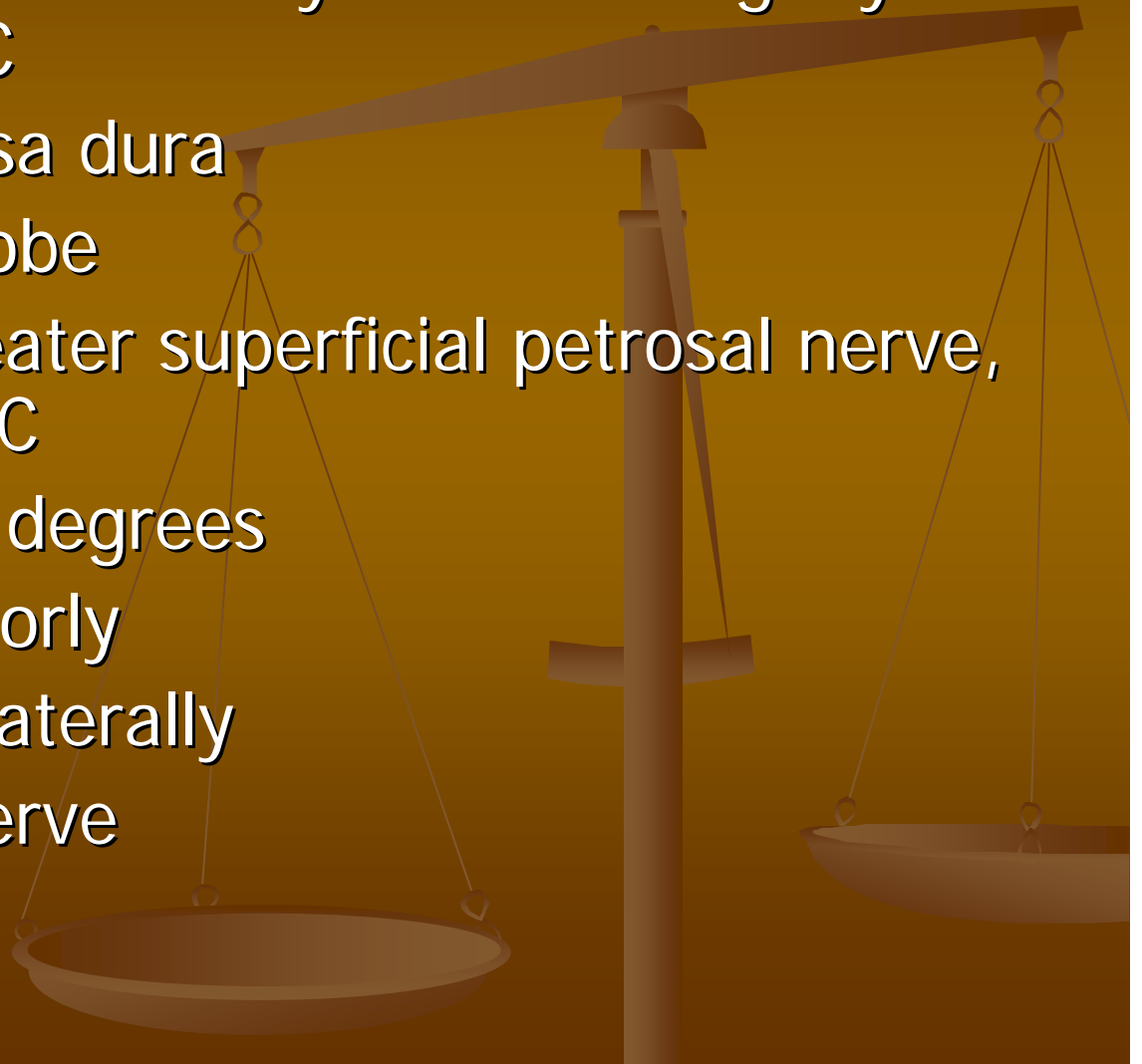
# ***Selective Vestibular Nerve Section***

- Described early 20<sup>th</sup> century
- High incidence facial nerve injury
- House 1961 Middle fossa approach
- Brackmann, Hitselberger, Silverstein 1978, retrolabyrinthine approach
- Retrosigmoid and retrosigmoid-IAC

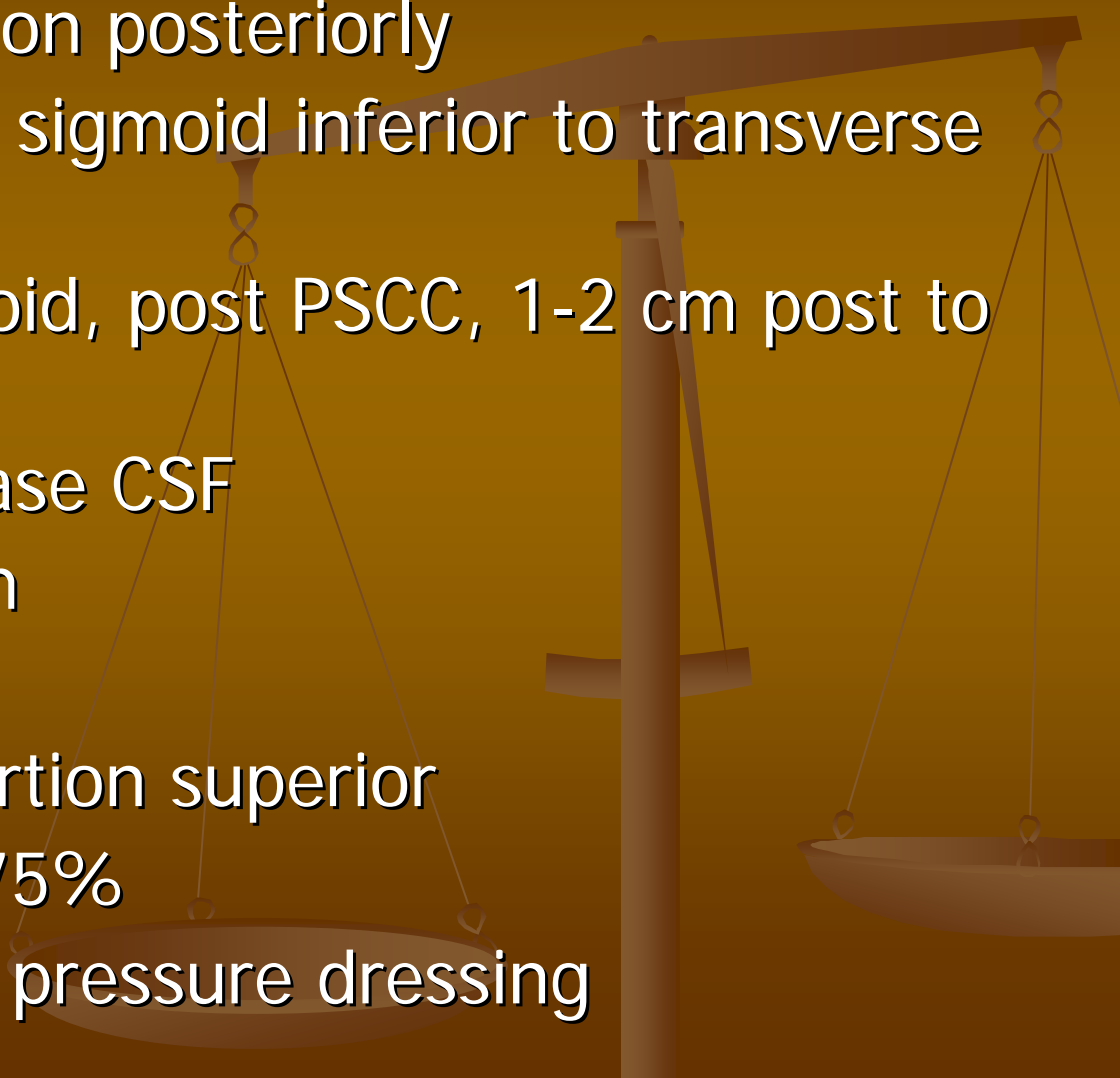


# ***Middle Fossa Approach***

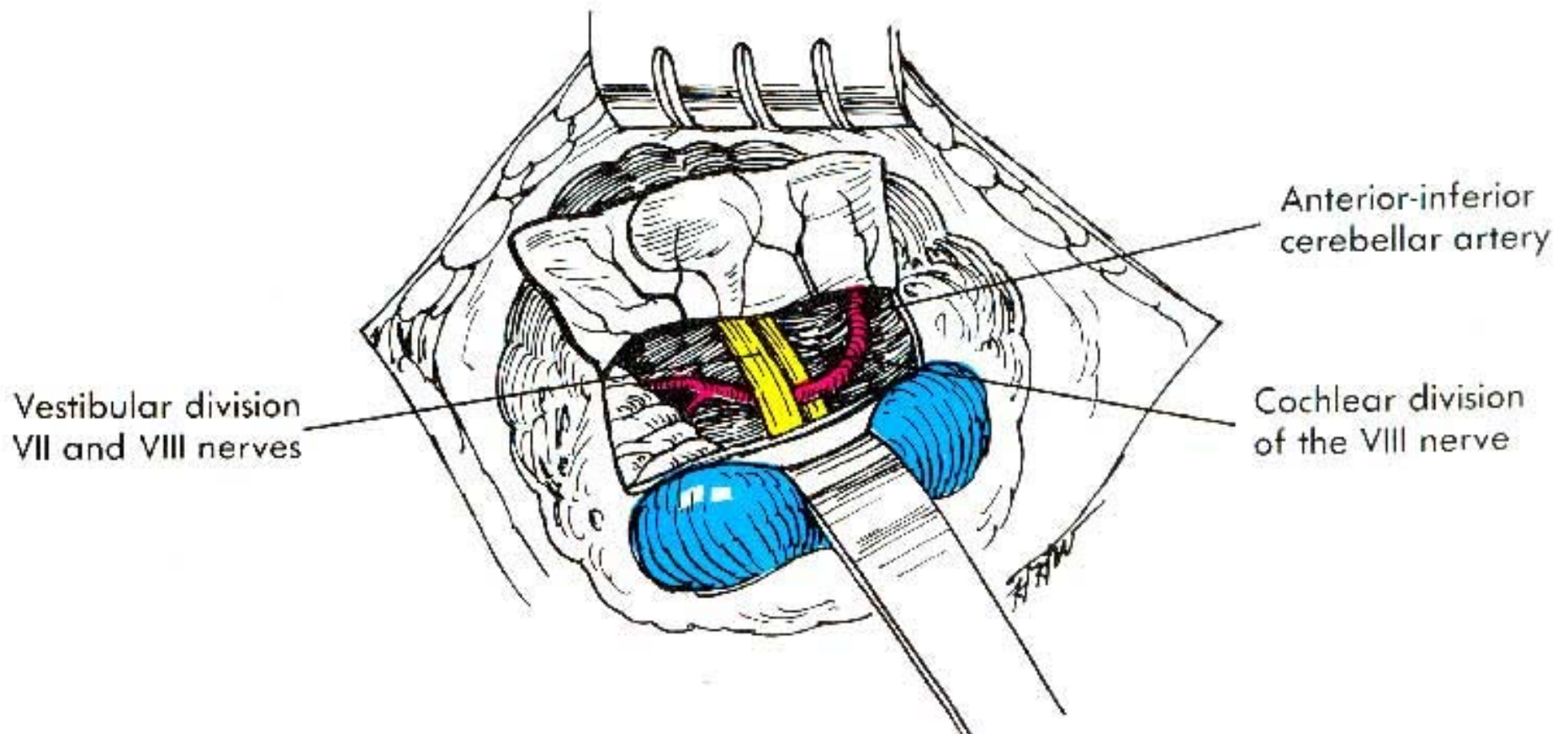
- 4X4 cm temporal craniotomy centered slightly anterior to the EAC
- Elevate Middle fossa dura
- Retract temporal lobe
- landmarks IAC Greater superficial petrosal nerve, malleus head, SSCC
- Remove bone 180 degrees
- Incise dura posteriorly
- Section SVN, IVN laterally
- Include singular nerve
- Muscle or fat plug



# ***Retrolabyrinthine/retrosigmoid***

- Post-auricular incision posteriorly
  - Craniotomy post to sigmoid inferior to transverse sinus 4x5 cm
  - RL- complete mastoid, post PSSC, 1-2 cm post to sigmoid
  - Dural incision, release CSF
  - Displace cerebellum
  - Sigmoid retracted
  - Porus vestibular portion superior
  - Cleavage plane in 75%
  - Abd fat in retrolab, pressure dressing
- 

# Retrolab. Posterior fossa Surgery



# VNS

- Approach success varies by author
- Overall 90% elimination of vertigo MFA
- 95% substantial improvement
- Complications
  - headache, hearing loss, CSF leak
  - Dysequilibrium 30%
  - Hearing loss uncommon
  - CN VII injury less than 5%
  - Meningitis, hemorrhage, stroke more rare

# VNS

## ■ MFA

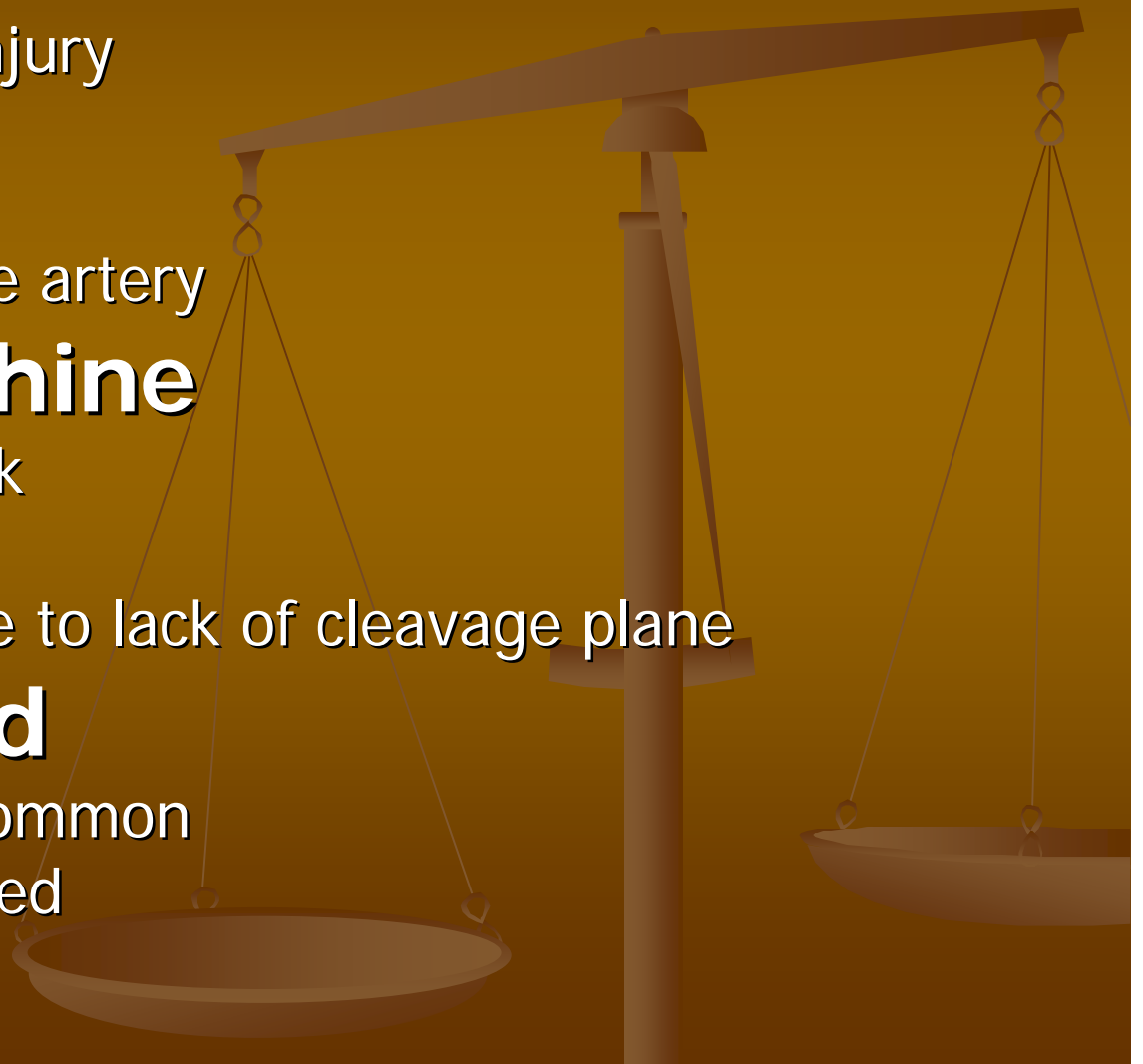
- Increase CN VII injury
- Memory loss
- Convulsion
- SNHL ?labyrinthine artery

## ■ Retrolabyrinthine

- Increased CSF leak
- CHL
- Lower success due to lack of cleavage plane

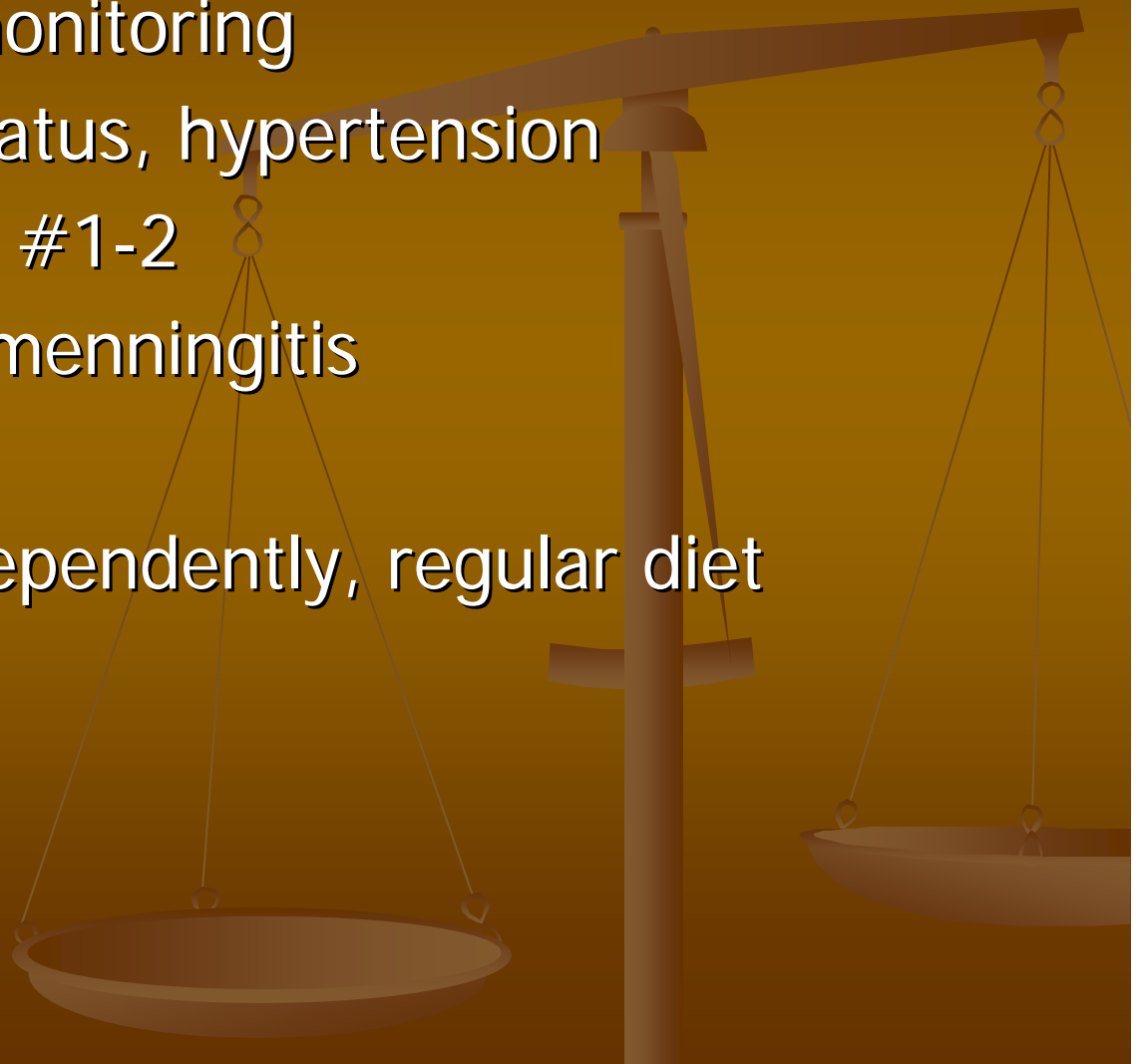
## ■ Retrosigmoid

- Headache more common
- Greater if IAC drilled



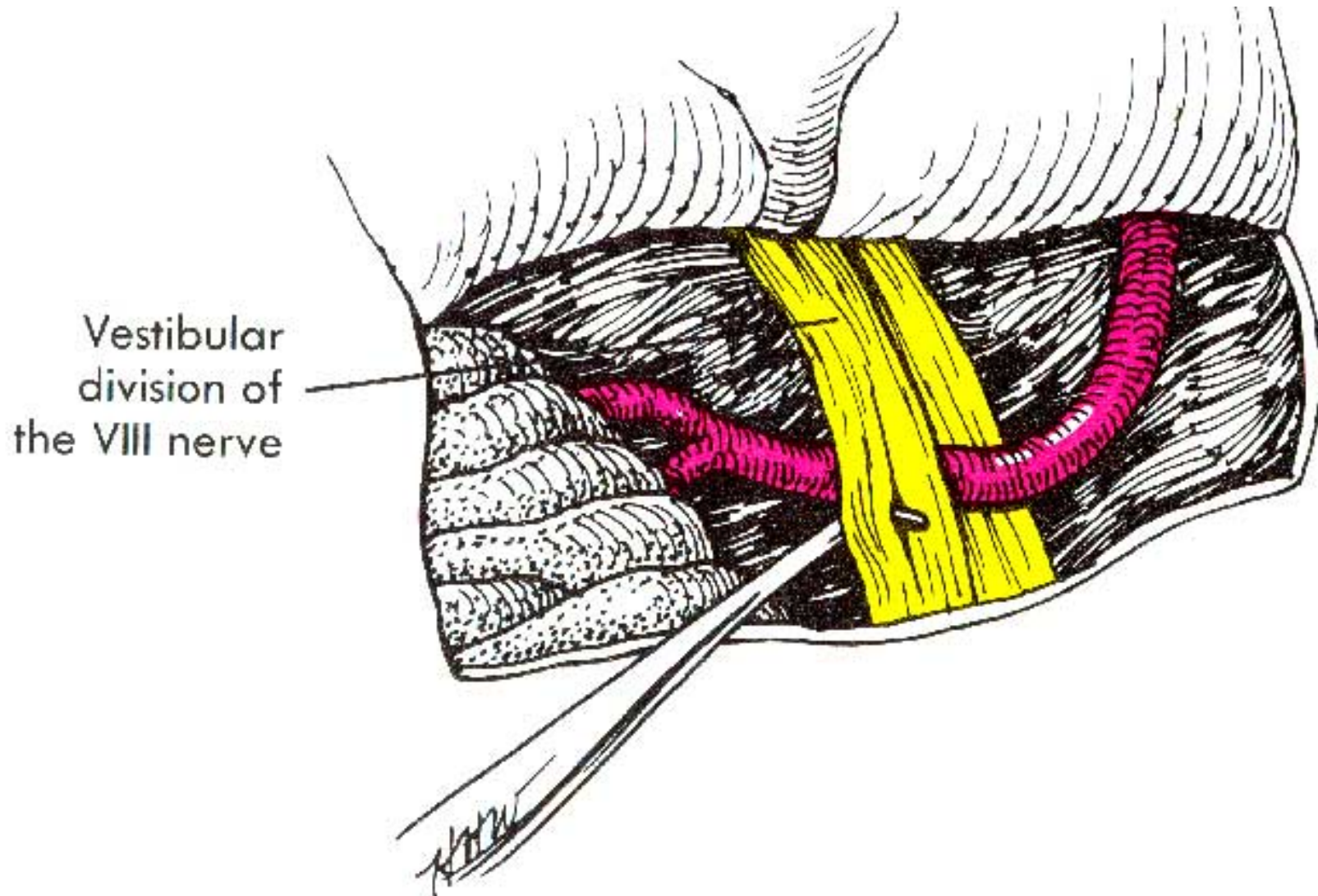
# Selective VNS

- CN VII and VIII monitoring
- ICU, neurologic status, hypertension
- Regular floor POD #1-2
- Observe for CSF, meningitis
- Early ambulation
- D/C ambulate independently, regular diet





# Vestibular nerve section





# ***Bilateral***

- Serc
- Steroid (RIBS)
- Sac Sx
- Streptomycin IM



## ***Functional scale for Meniere's***

- 1 No effect on activities at all.
- 2 I have to stop what I am doing during an attack but may resume activities when it has passed. I continue to work, drive, and engage in most activities I choose without restriction. I have made no changes in my activities in order to accommodate the dizziness.
- 3 I have to stop what I am doing during an attack but may resume activities when it has passed. I continue to work, drive, and engage in most activities I choose, but I have had to make changes in my activities in order to allow for the dizziness.
- 4 I am able to work, drive, travel, take care of my family, or engage in most essential activities, but I must exert a great deal of effort to do so. I must constantly make adjustments in my activities and budget my energy. I am barely making it.
- 5 I am unable to work, drive, or take care of my family. I am unable to do most of the active things I used to do. Even essential activities must be limited. I am disabled.
- 6 I have been disabled for 1 year or longer, and/or I receive compensation because of dizziness or balance problems.

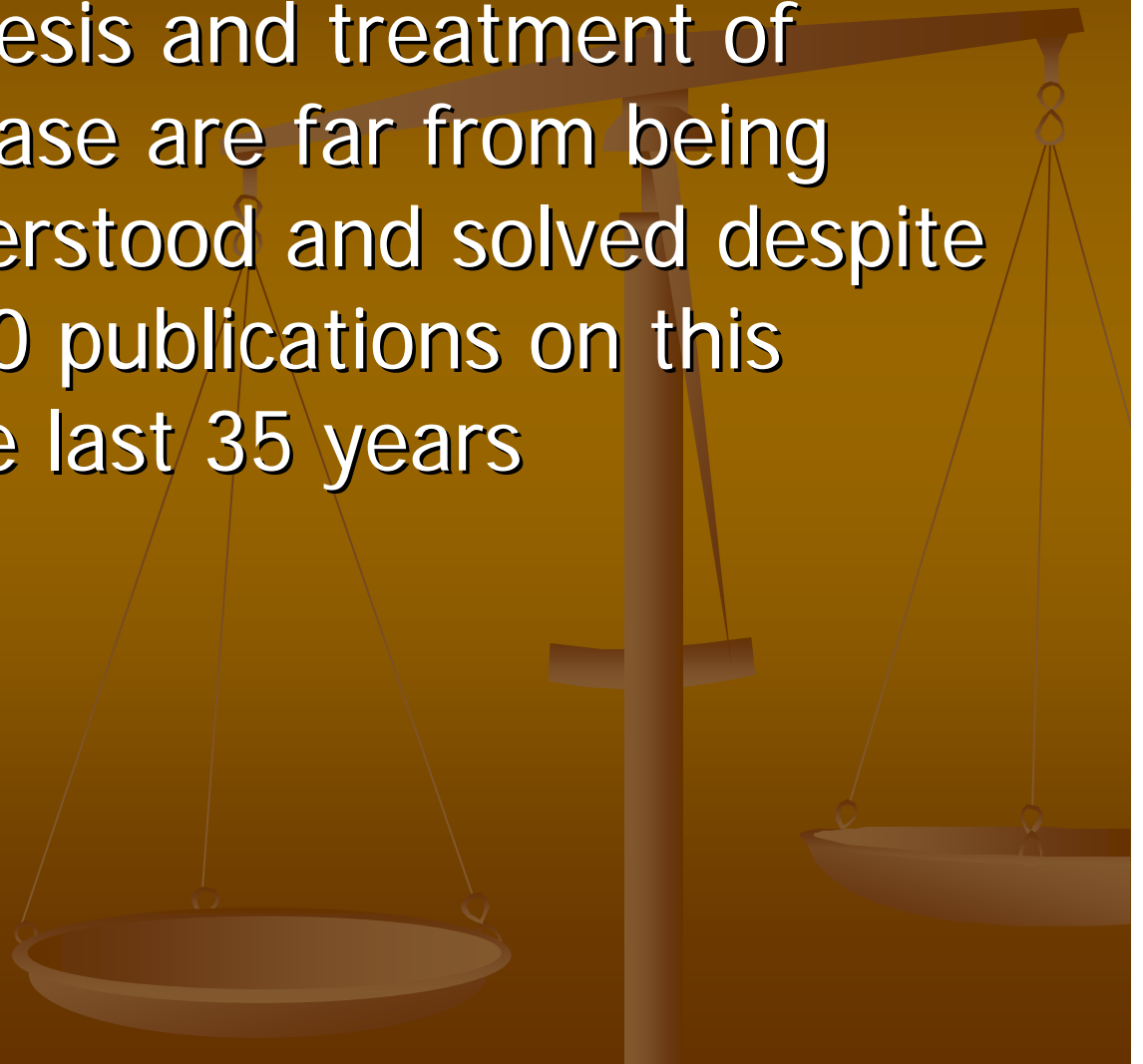
## *Staging of hearing in Meniere's*

Stage	Four-tone pure-tone average <sup>a</sup>
1	<26
2	26–40
3	41–70
4	>70

<sup>a</sup>Calculated as the mean of thresholds at 0.5, 1, 2, and 3 kHz from the patient's worst audiogram in the 6 months preceding treatment. These guidelines were designed only for certain and definite cases of Meniere's disease.

# ***Conclusion***

The etiopathogenesis and treatment of **Meniere's** disease are far from being completely understood and solved despite more than 4,000 publications on this disease over the last 35 years



# 10%

- Dizziness
- Bilateral 1<sup>st</sup> year
- FHx
- Fail Gent



# 50

- 50 Y Age
- 50% Duration 1 to 2 hours
- 50 dB pure tone average
- 50% mean speech discrimination score
- 50% Normal ENG
- 50% average caloric response reduction
- 50% bilateral in 5Y
- 50% vertigo ceased spontaneously in 2 Y
- 50% complete remission

Otolaryngol Head Neck Surg 1989; 100:6-16

Acta Otolaryngol Suppl (Stockh) 1984; 406:72-77

***Thanks***

