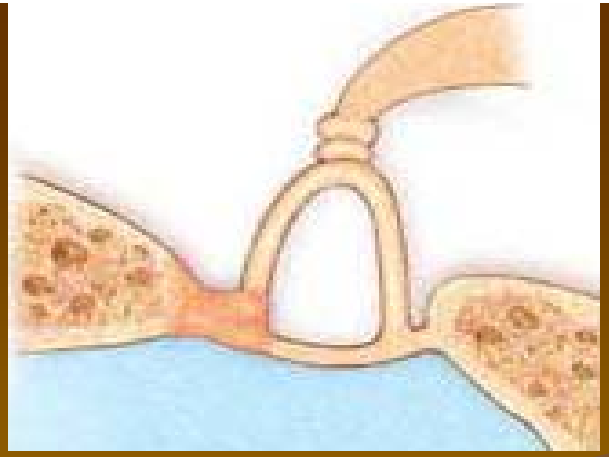


Otosclerosis

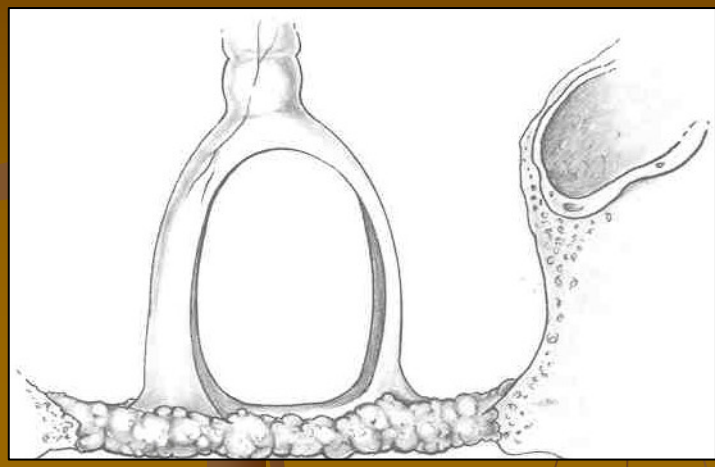
Abdul-Rahman Hagr

MBBS FRCSc

Feb 27, 2005

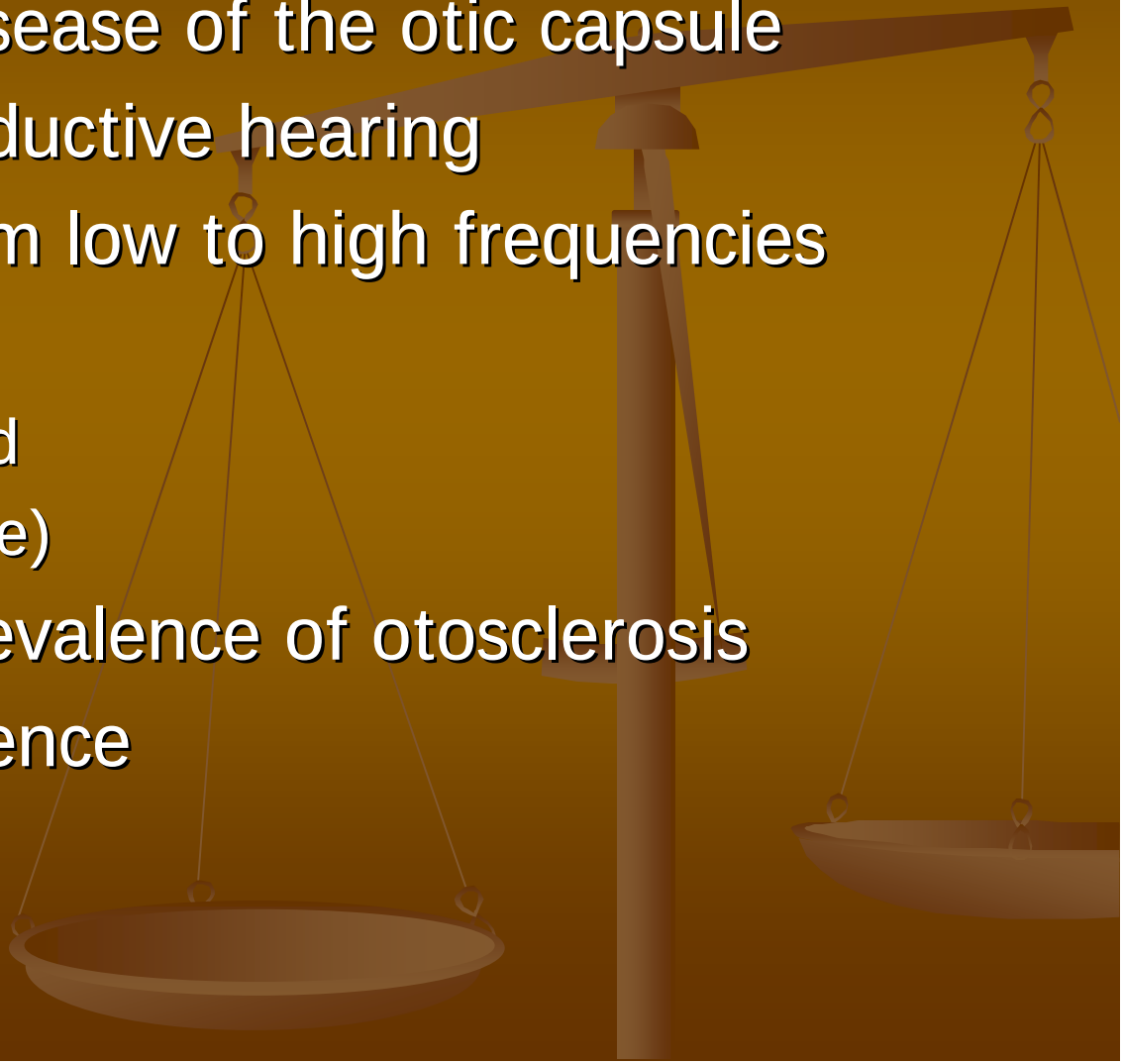
Objectives

- Introduction
- Pathology
- Diagnosis
- Treatment



Introduction

- Metabolic bone disease of the otic capsule
- FP fixation → conductive hearing
- HL progresses form low to high frequencies
- Sensorineural
  - Cochlea is involved
  - False (Over-closure)
- 10% histologic prevalence of otosclerosis
- 1% clinical prevalence



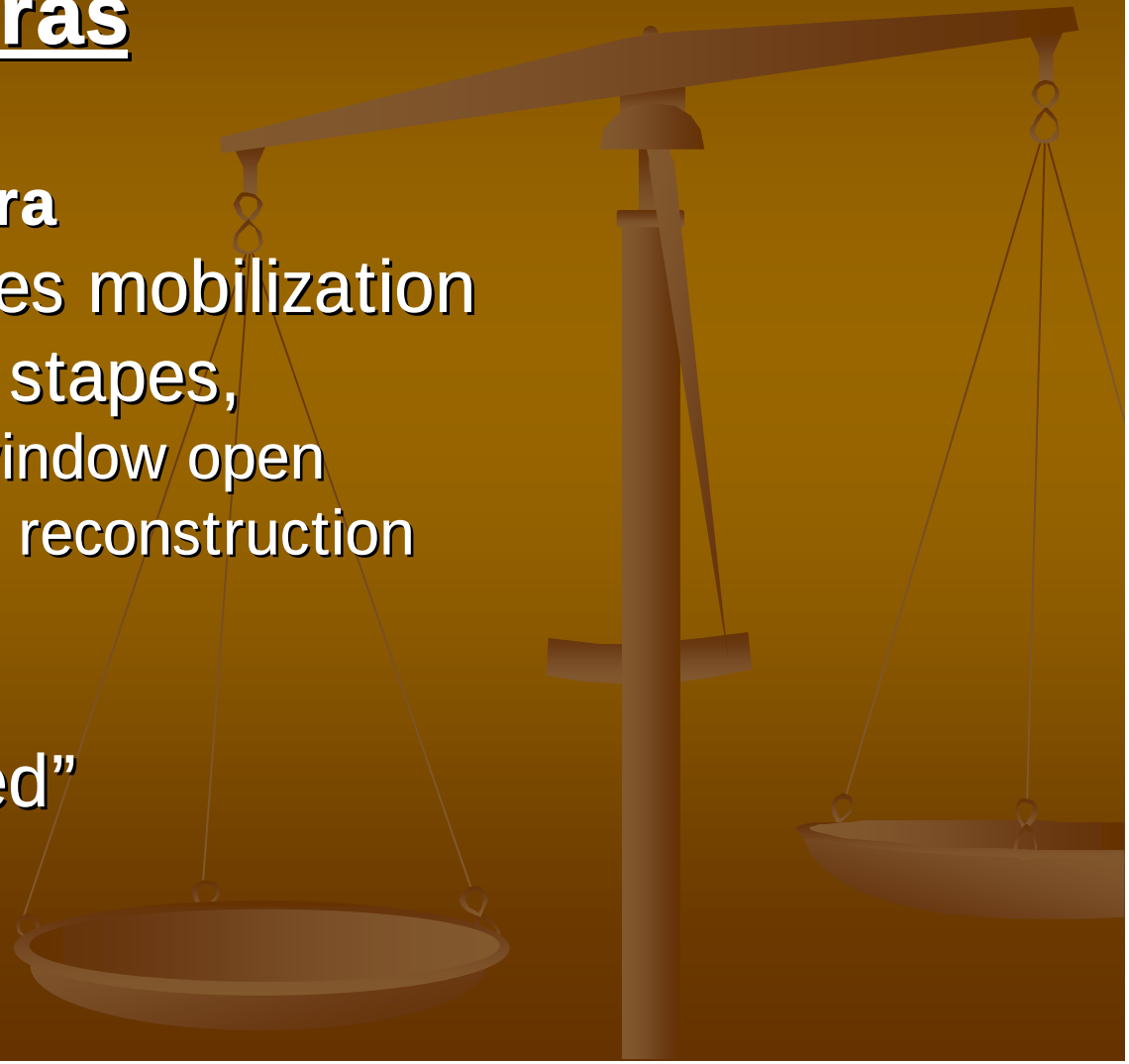
History of the Procedure

Three distinct eras

1-The mobilization era

- Kessel 1800s stapes mobilization
- Jack removed the stapes,
  - leaving the oval window open
  - No ossicular chain reconstruction

→fatal meningitis
→temporary “re-fixed”



History of the Procedure

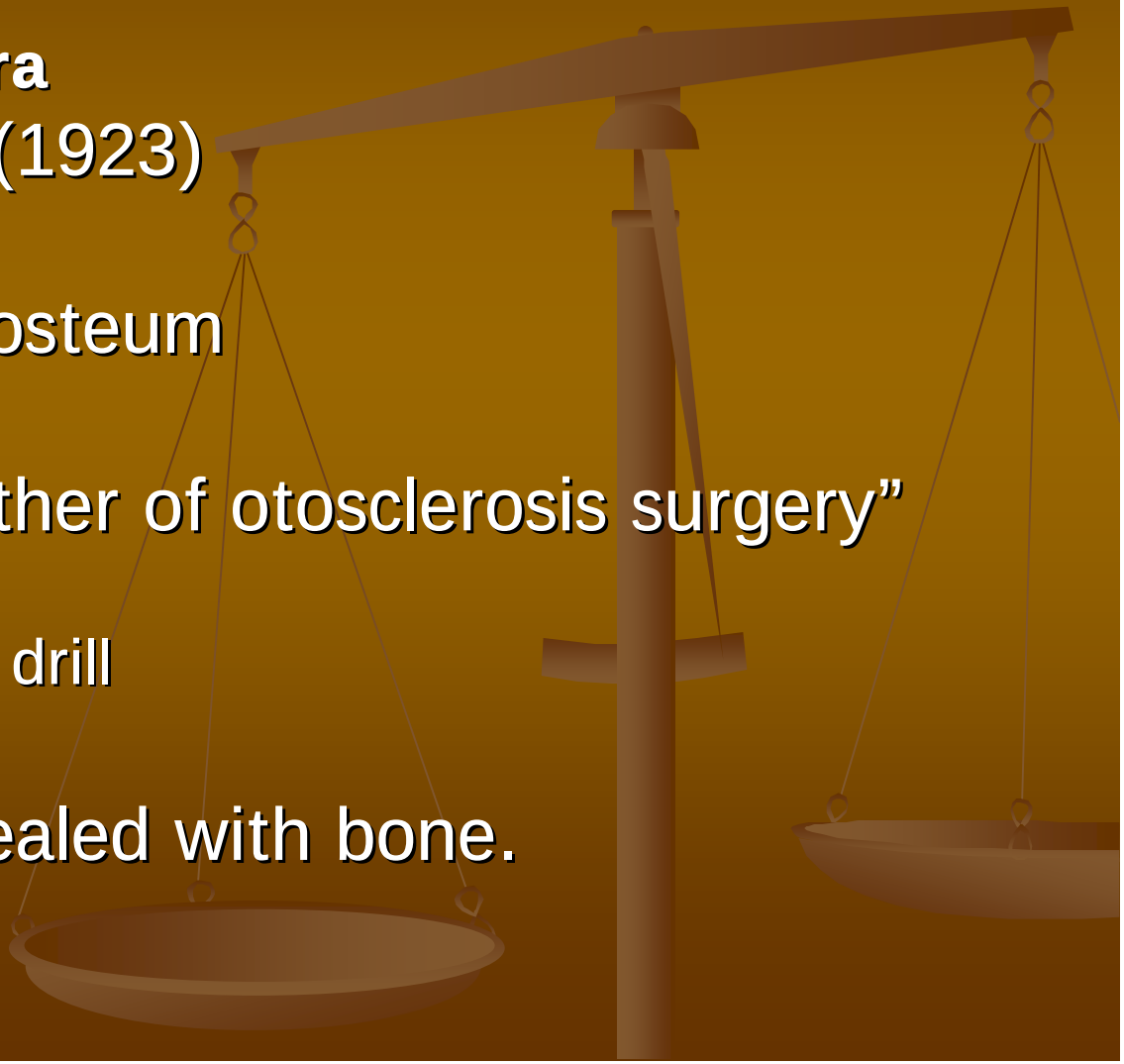
2-The fenestration era

Holmgren (1923)

- fistula in HSCC
- sealed it with periosteum

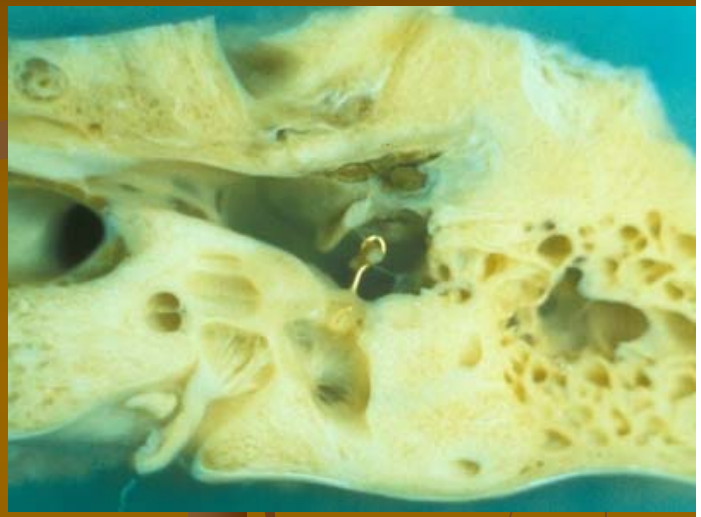
- Lempert 1938 “Father of otosclerosis surgery”
  - One stage Sx
  - Endaural + dental drill

- Temporary → resealed with bone.



History of the Procedure

3- The stapedectomy era

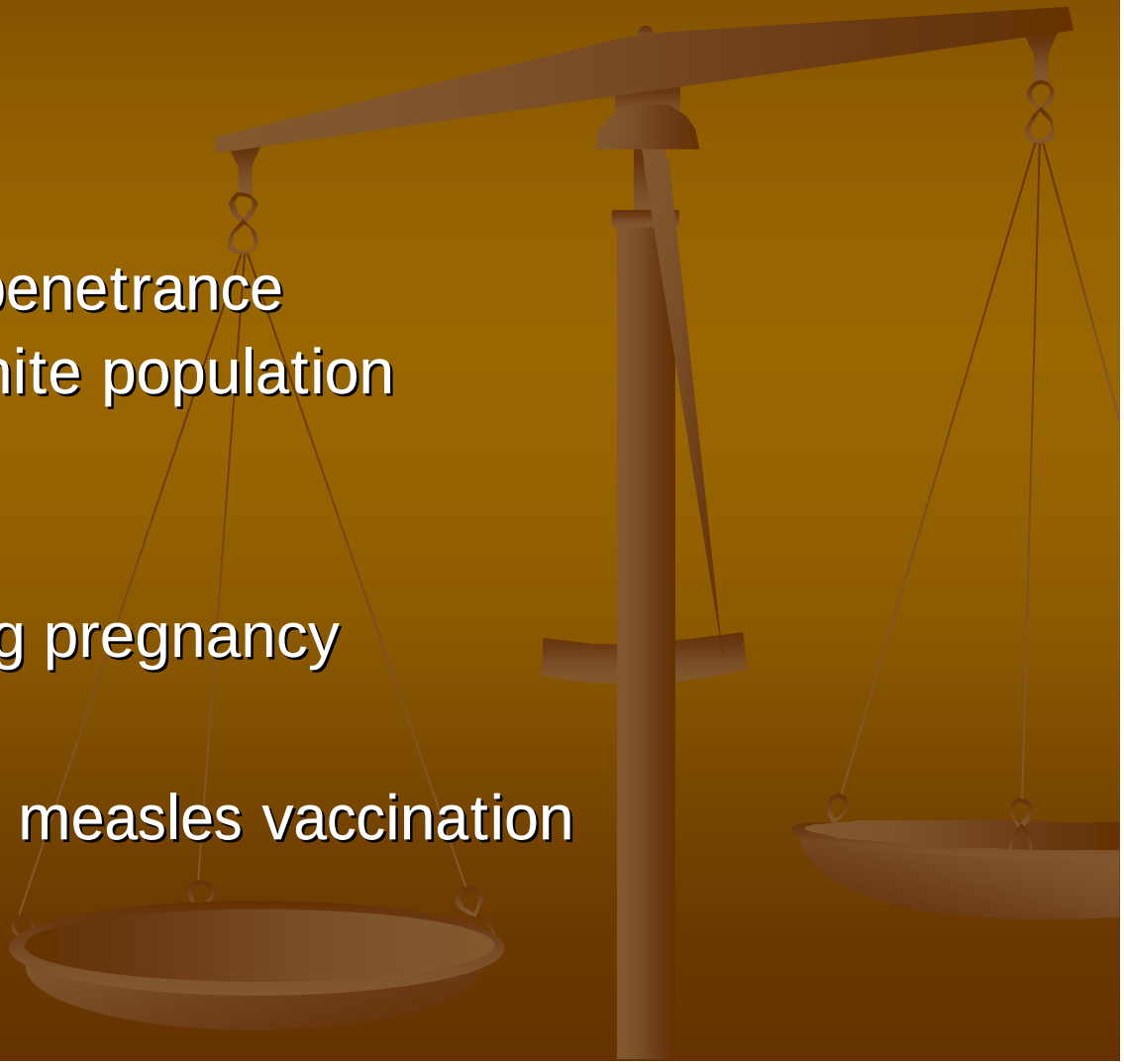


- Shea 1958
  - removed the stapes
  - sealed the oval window with an autograft vein wall
  - Reconstructed with an artificial prosthesis.

- Myers → stapedotomy
- Perkins→ Laser for stapedotomy

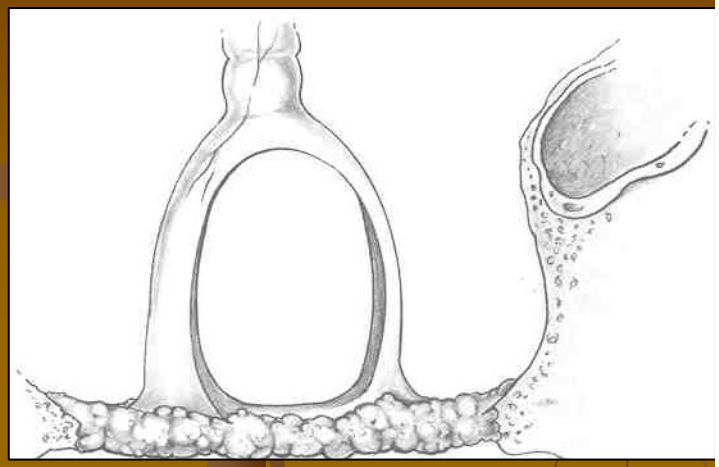
Etiology of Otosclerosis

- Unknown
- Genetic
  - +ve FH 50-70%
  - AD incomplete penetrance
  - limited to the white population
- Hormonal
  - F>M
  - accelerate during pregnancy
- Viral
  - Decreasing after measles vaccination



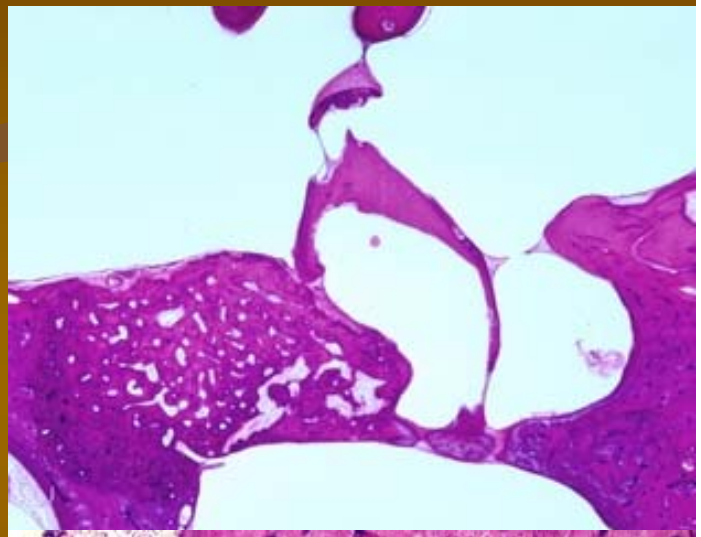
Objectives

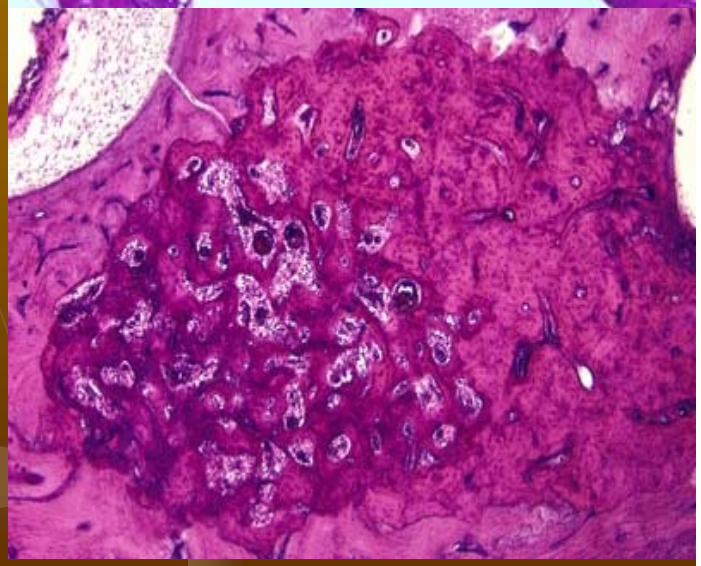
- Introduction
- ***Pathology***
- Diagnosis
- Treatment

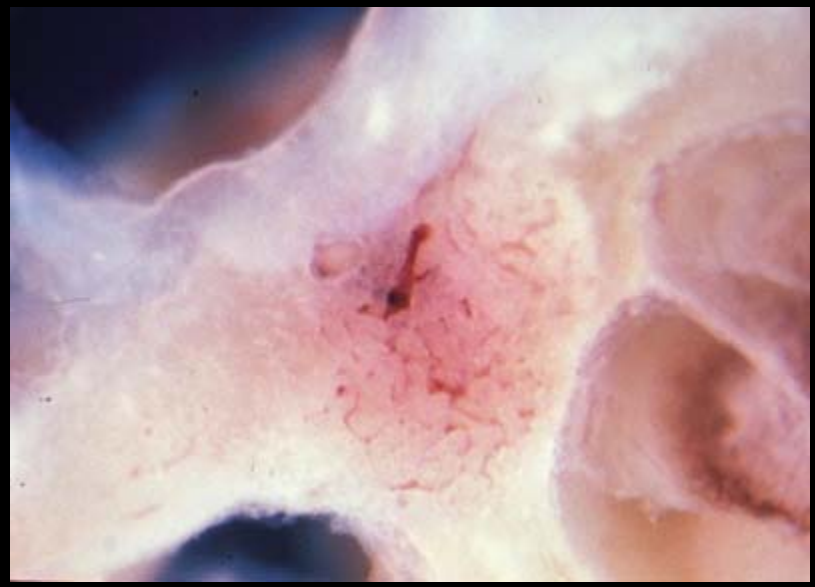


Pathology

- Two phases of disease
  - Active (otospongiosis)
    - Osteocytes, histiocytes,
    - Active resorption of bone
    - Schwartze's sign
  - Mature (sclerotic phase)
    - Deposition of new bone
    - osteoblasts



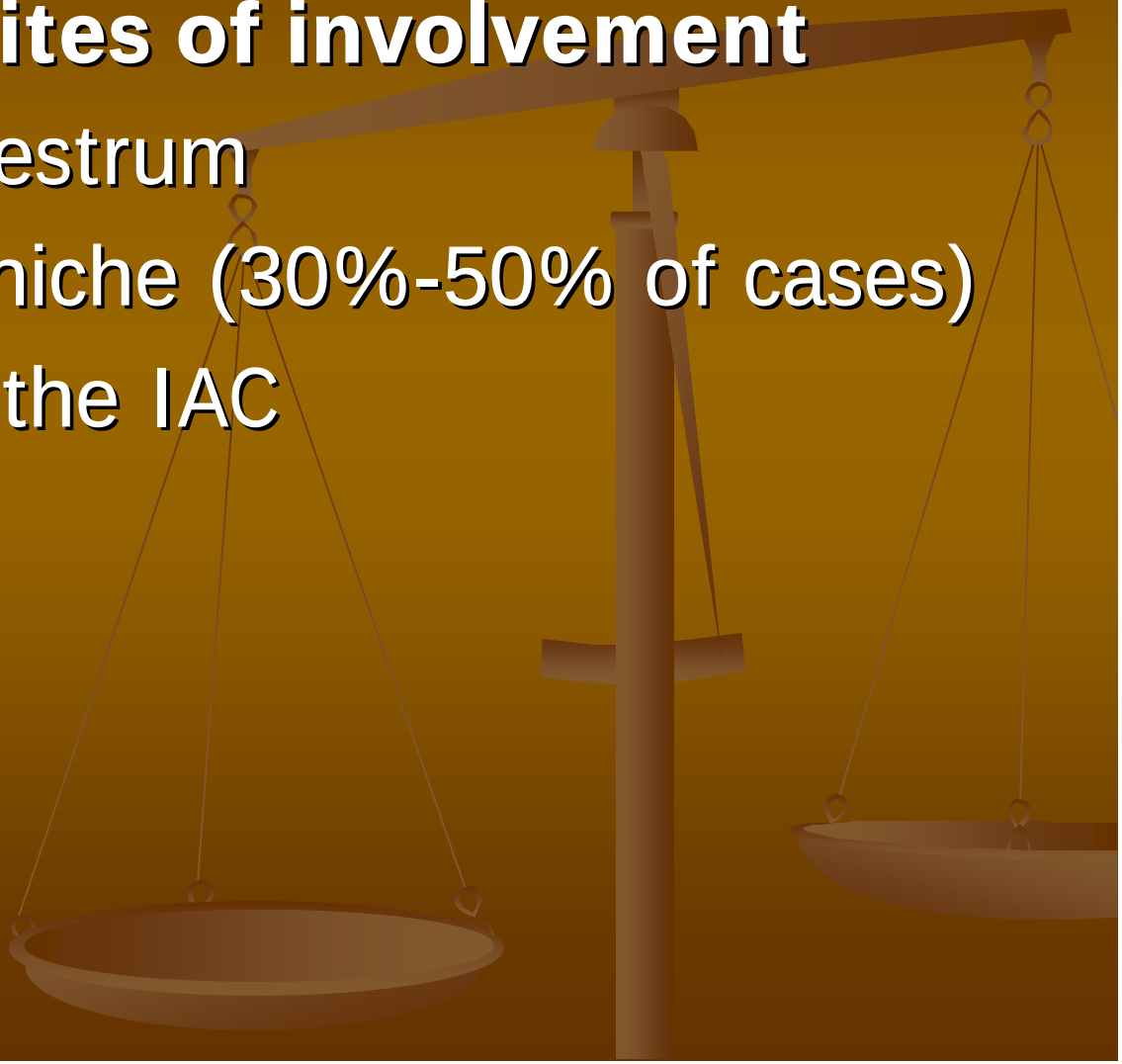




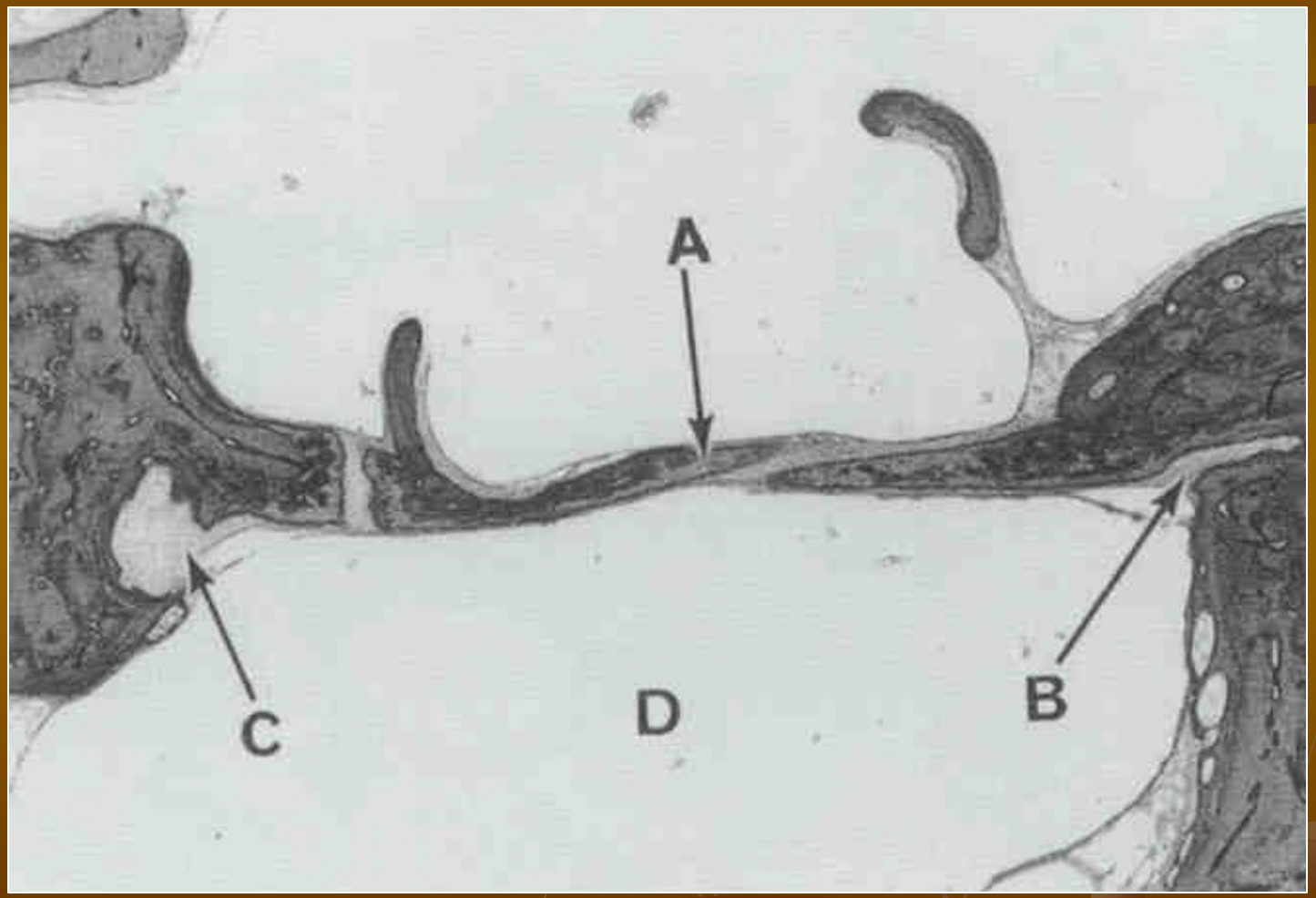
Pathology

Most common sites of involvement

- Fissula ante fenestrum
- Round window niche (30%-50% of cases)
- Anterior wall of the IAC

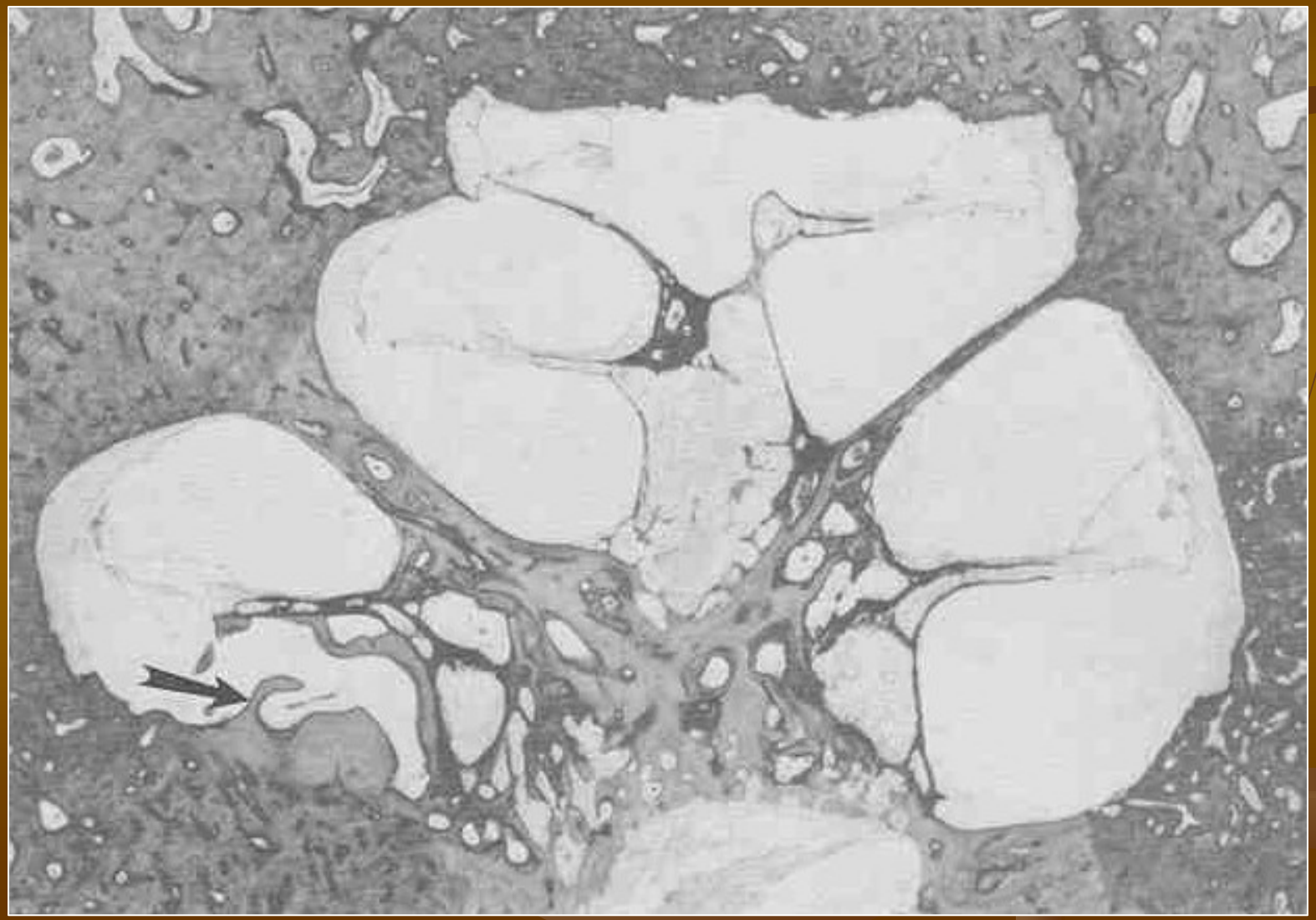


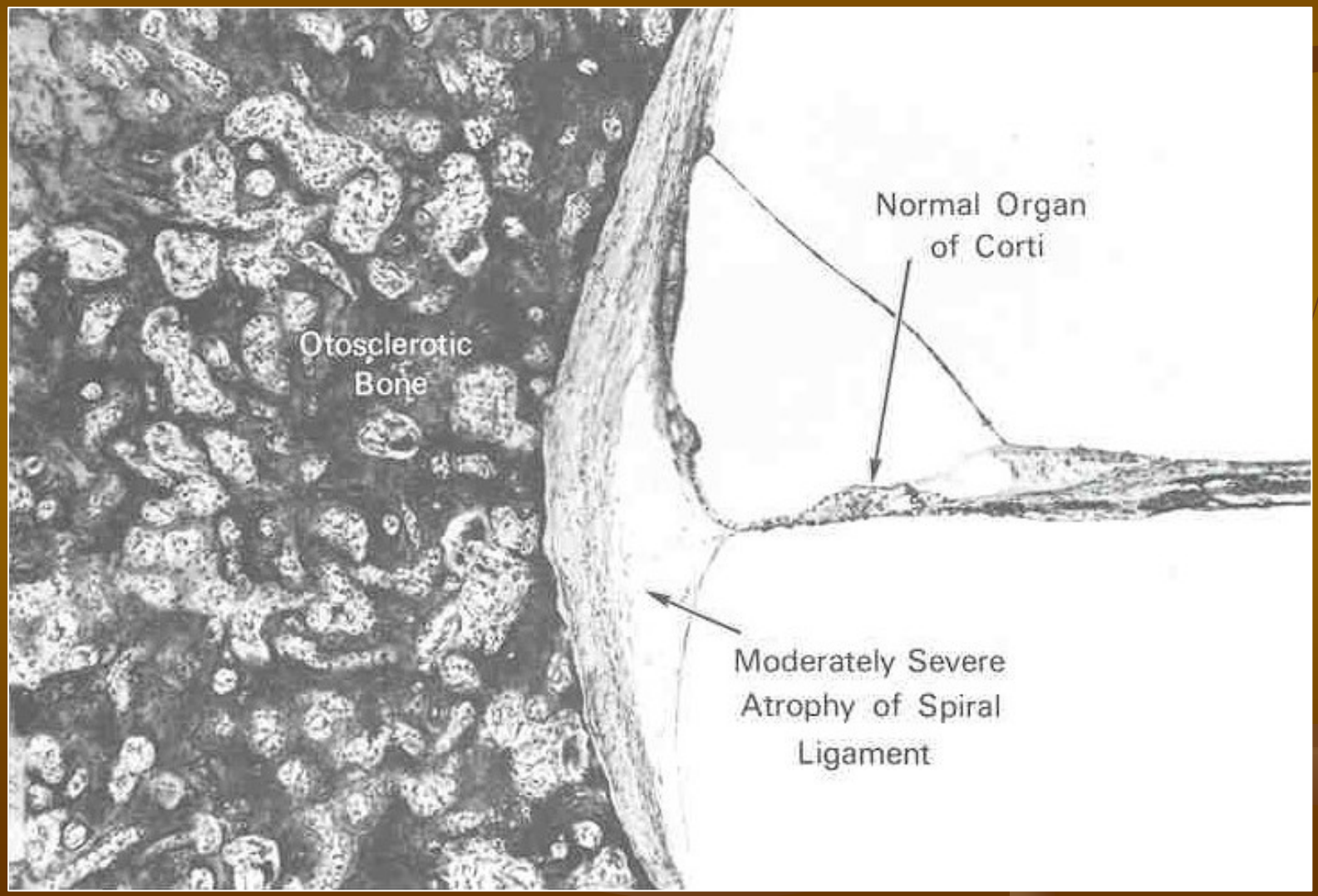
Fissula ante and post fenestrum



Post Footplate Involvement

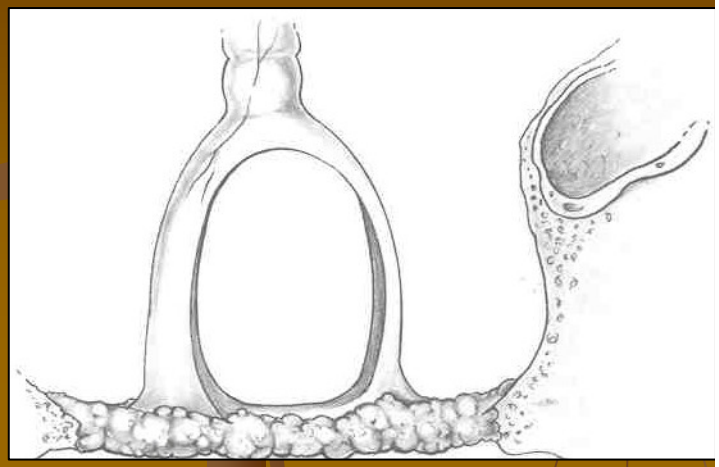
Erosion into inner ear




Otosclerotic
Bone
Normal Organ
of Corti
Moderately Severe
Atrophy of Spiral
Ligament

Objectives

- Introduction
- Pathology
- ***Diagnosis***
- Treatment

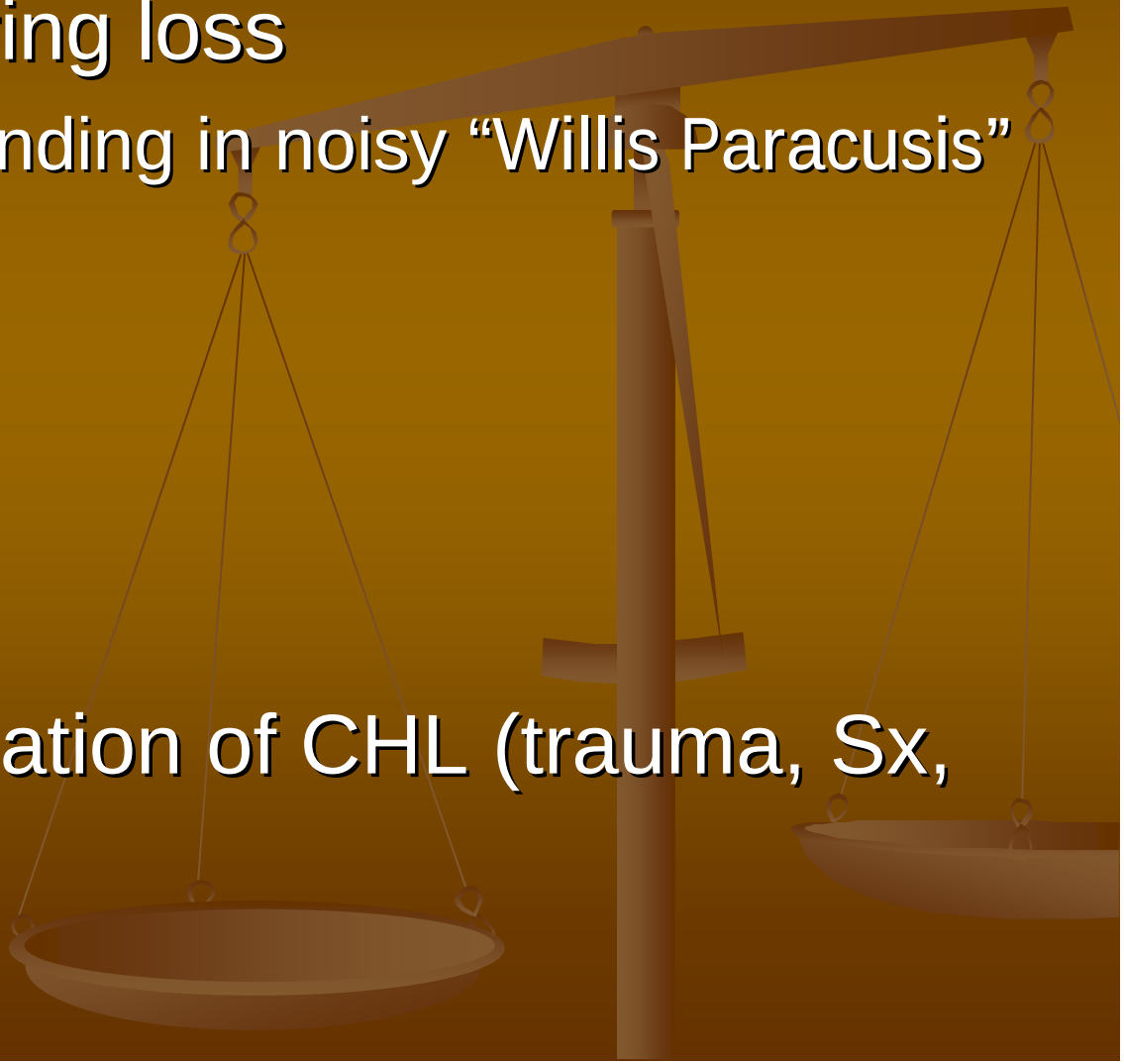


Diagnosis

- Symptoms
- Sings
- Audiometric Findings
- Differential Diagnosis

Symptoms

- conductive hearing loss
  - Better understanding in noisy “Willis Paracusis”
- Tinnitus
- 70% bilateral
- +ve FHx
- Pregnancy
- No other explanation of CHL (trauma, Sx, infection)

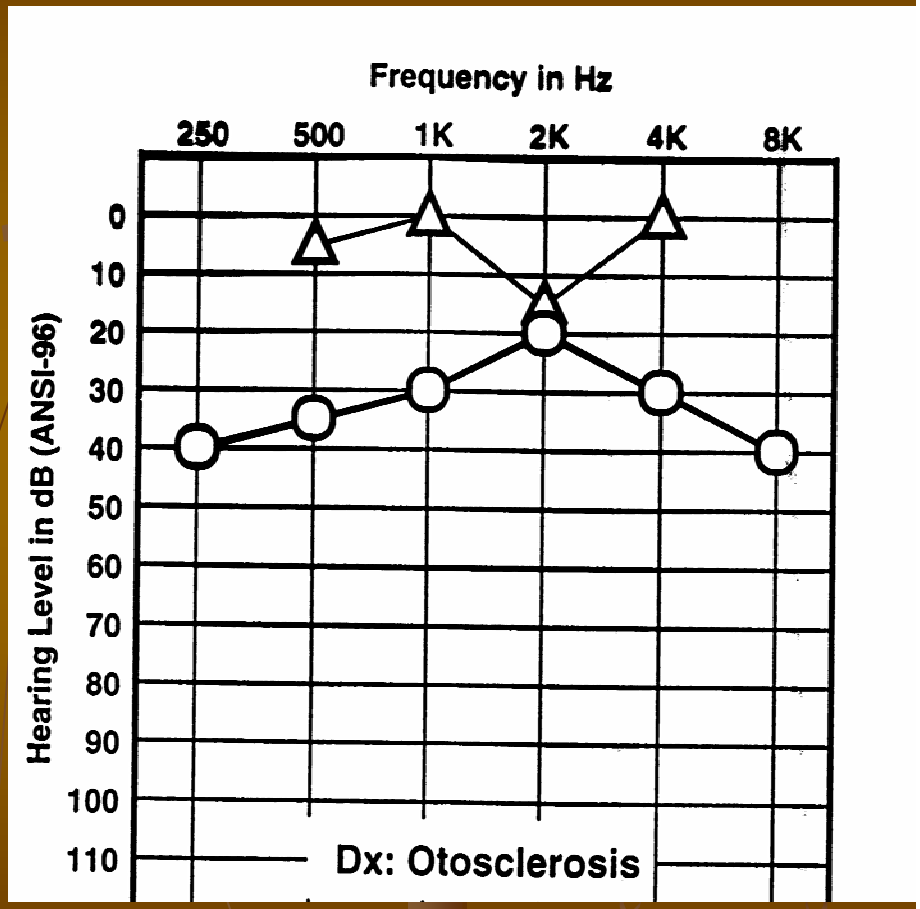


Sings

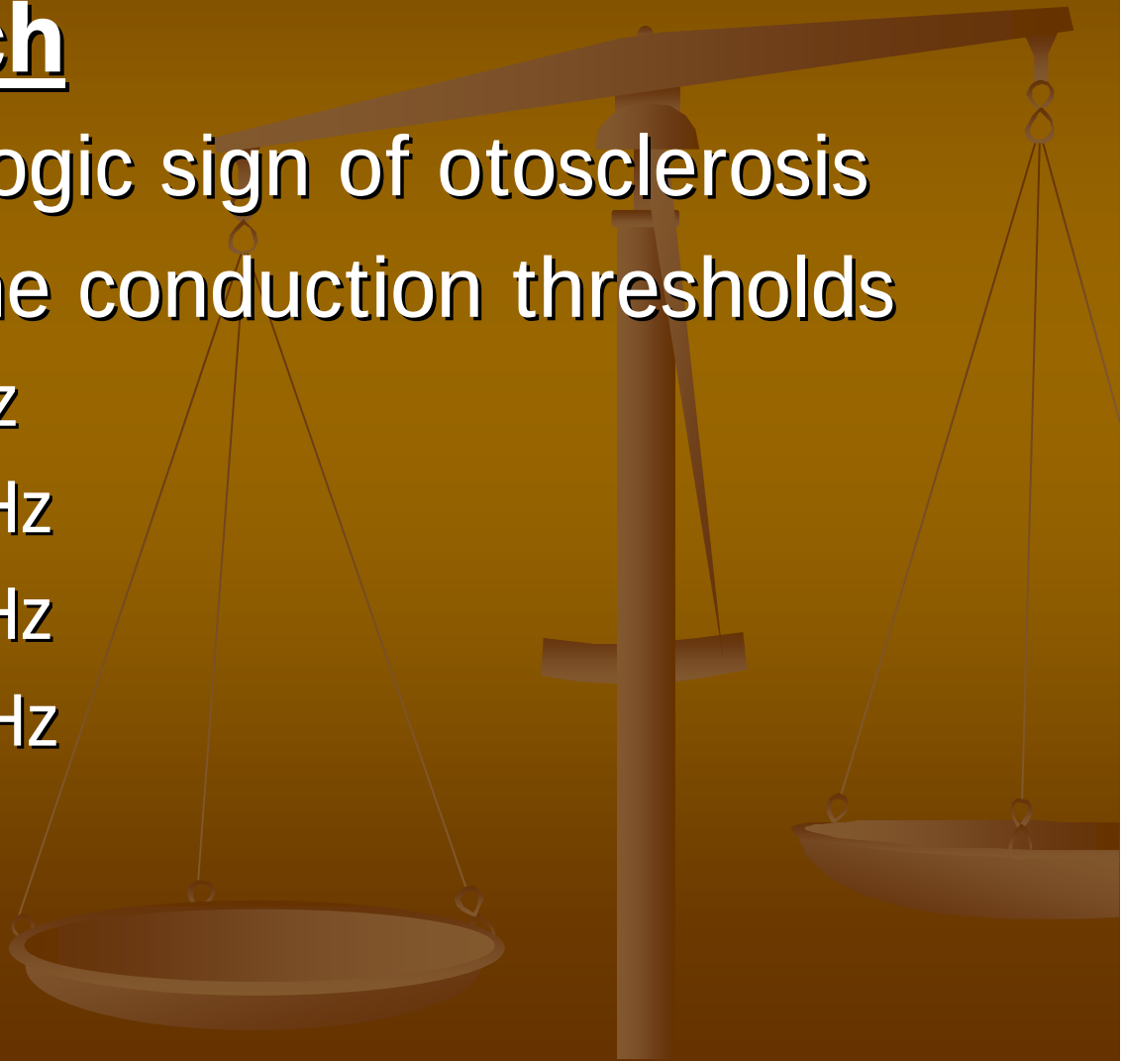
- CHL (T-Fork)
- Schwartze sign

Audiometric Findings

1. Conductive HL
2. Rising configuration
3. Carhart's notch
4. Tympanograms
   - Normal
   - As
   - On and off
5. SR -ve



Pure Tone Audiometry



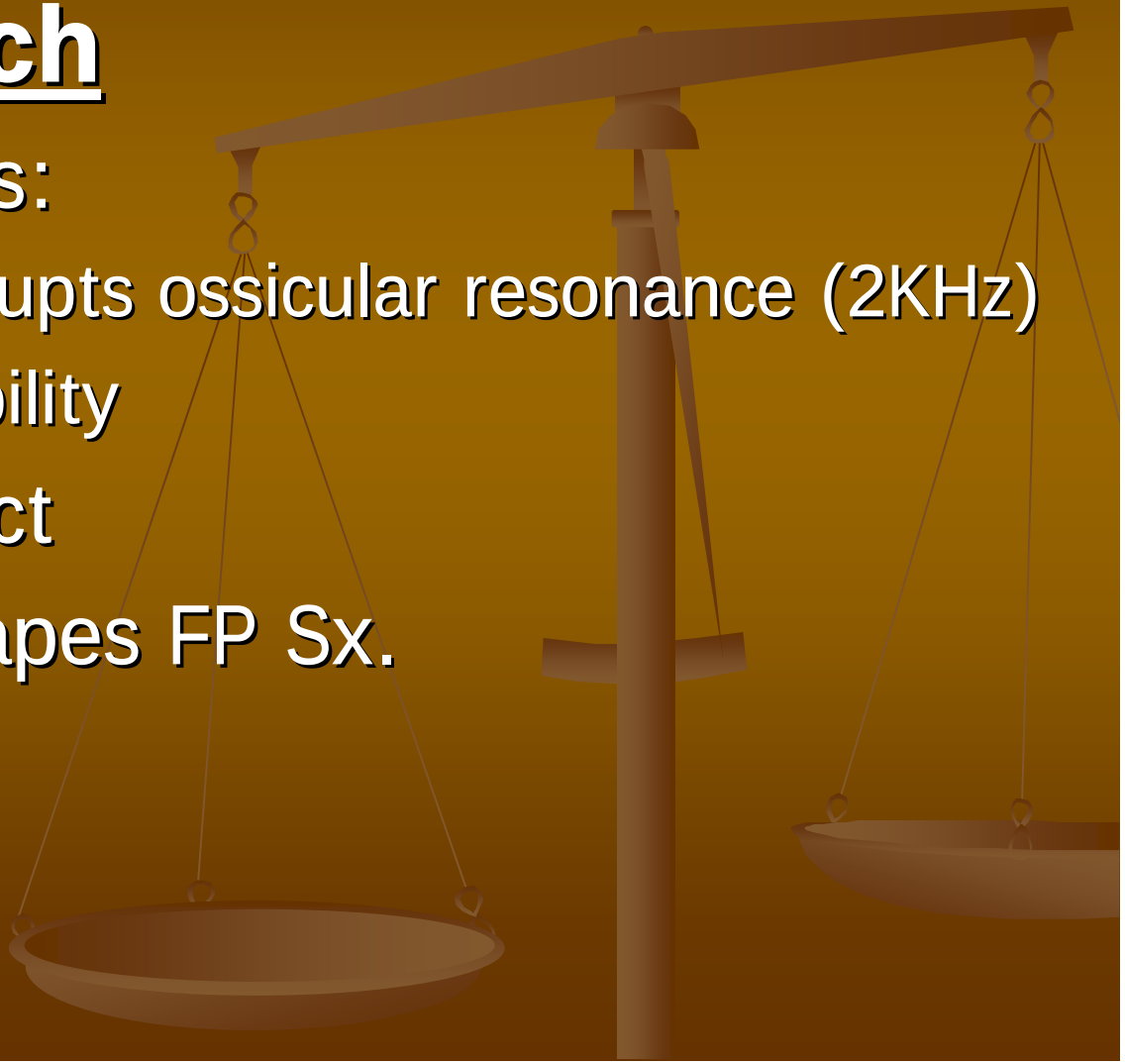
Carhart's notch

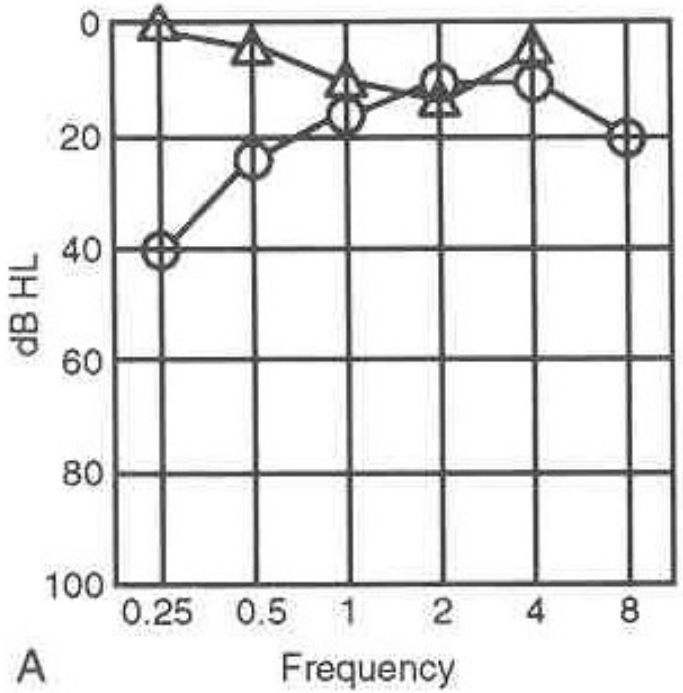
- Hallmark audiologic sign of otosclerosis
- Decrease in bone conduction thresholds
  - 5 dB at 500 Hz
  - 10 dB at 1000 Hz
  - 15 dB at 2000 Hz
  - 5 dB at 4000 Hz

Pure Tone Audiometry

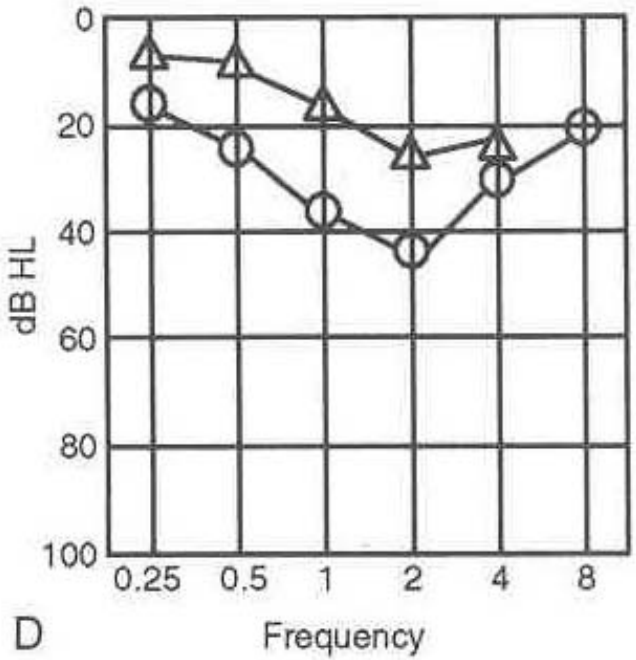
Carhart's notch

- Proposed theories:
  - FP fixation→ disrupts ossicular resonance (2KHz)
  - Perilymph immobility
- Mechanical artifact
- Reverses with stapes FP Sx.
- Over-closure




0
20
40
60
80
100
dB HL
0.25
0.5
1
2
4
8
Frequency

A


0
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40
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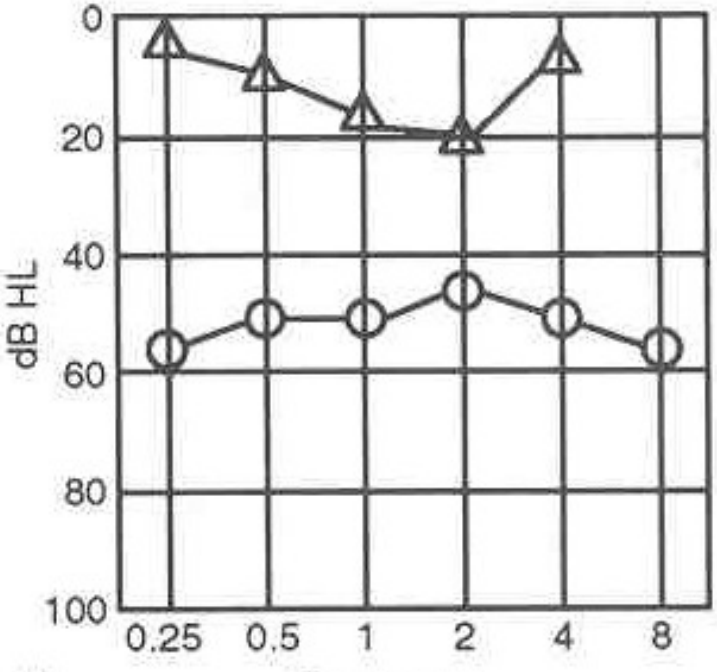
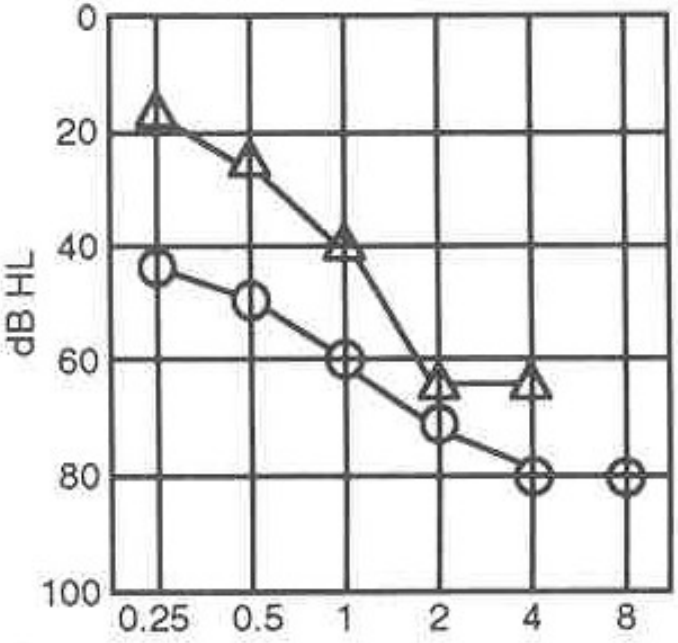
80
100
dB HL
0.25
0.5
1
2
4
8
Frequency

D

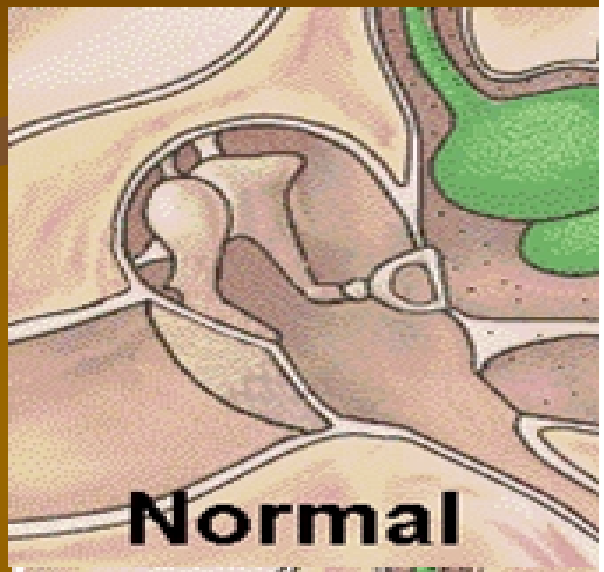

0
20
40
60
80
100
dB HL
0.25
0.5
1
2
4
8

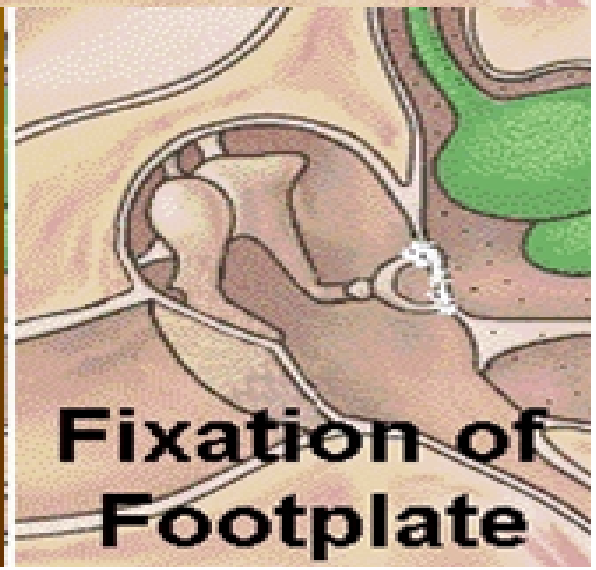

0
20
40
60
80
100
dB HL
0.25
0.5
1
2
4
8

Differential Diagnosis

- Any CHL “Intra-operative Dx”
- Ossicular discontinuity
- Malleus head fixation (0.5%)
- Paget’s disease
- Osteogenesis imperfecta
- Osteopetrosis
- SSCCDS
- Congenital FP fixation*

*Apert





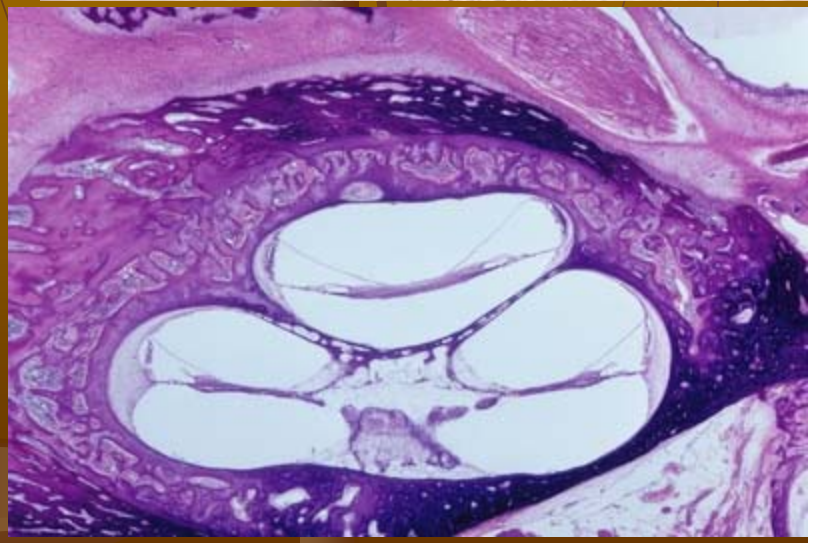
Osteogenesis imperfecta

- 30 Y
- Bilateral HL
- -ve FHx
- Fractures

Osteogenesis imperfecta

- Translucent sclera → choroid membrane →The blue sclera

- Endochondral layer contains abnormally large rests of cartilage.



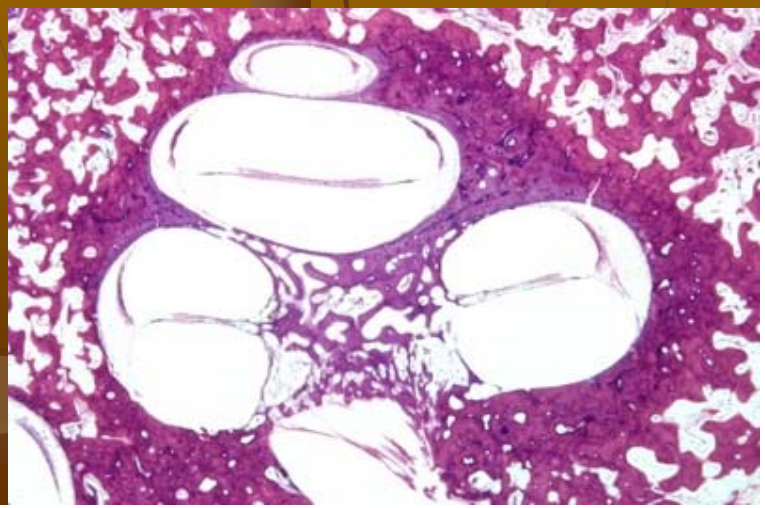
Paget's disease

- 80 Y
- Bil mix-HL

Paget's disease.

- SNHL is not caused by compression of VII nerve fibers
- CHL is not caused by ossicular fixation
- ? bone mineral density
- Sx correction of CHL are generally not considered worthwhile

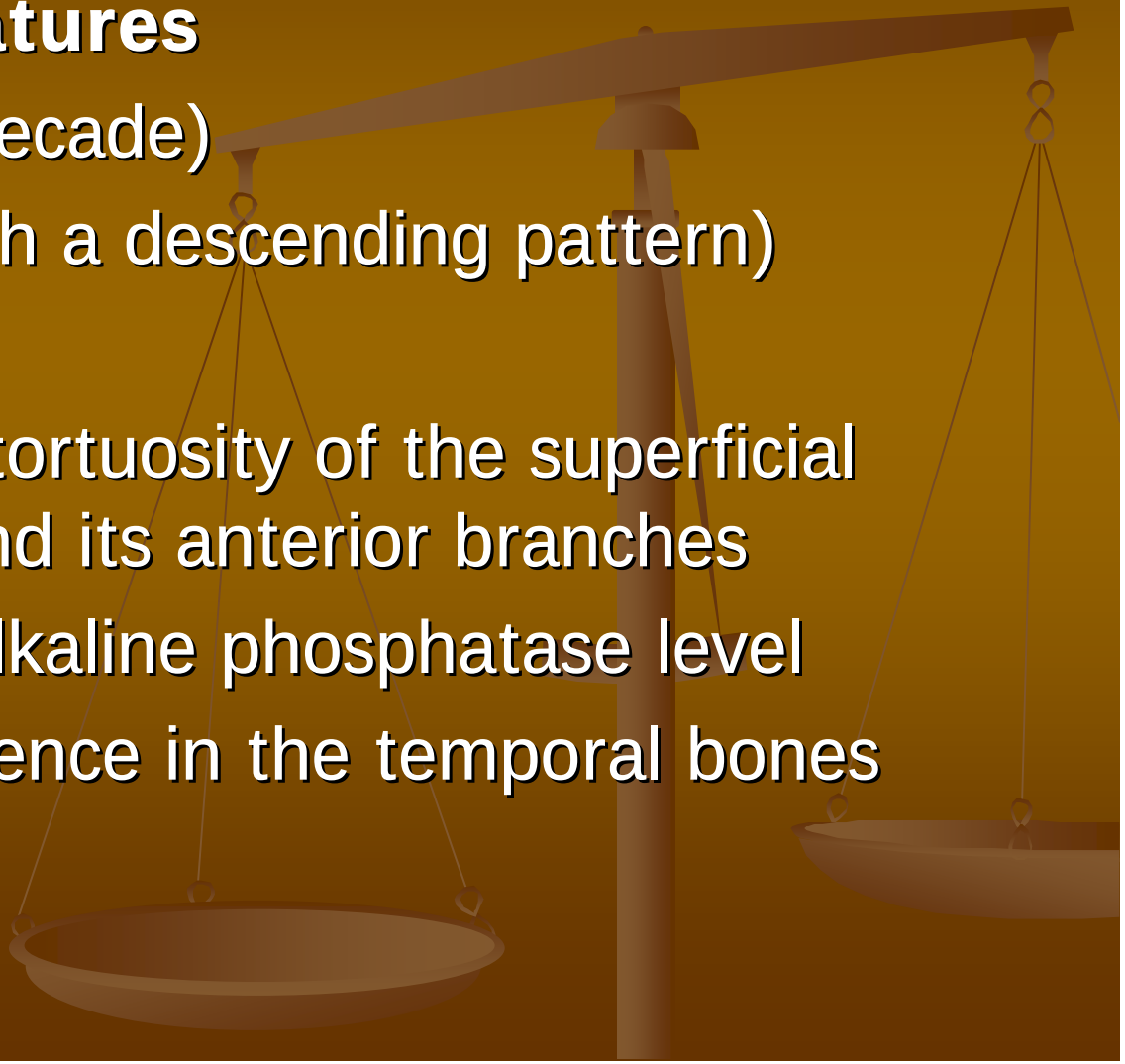
- **Otic capsule**
  - extensively eroded
  - replaced by pagetic bone
  - Normal FP



Paget's disease vs otosclerosis

Distinguishing features

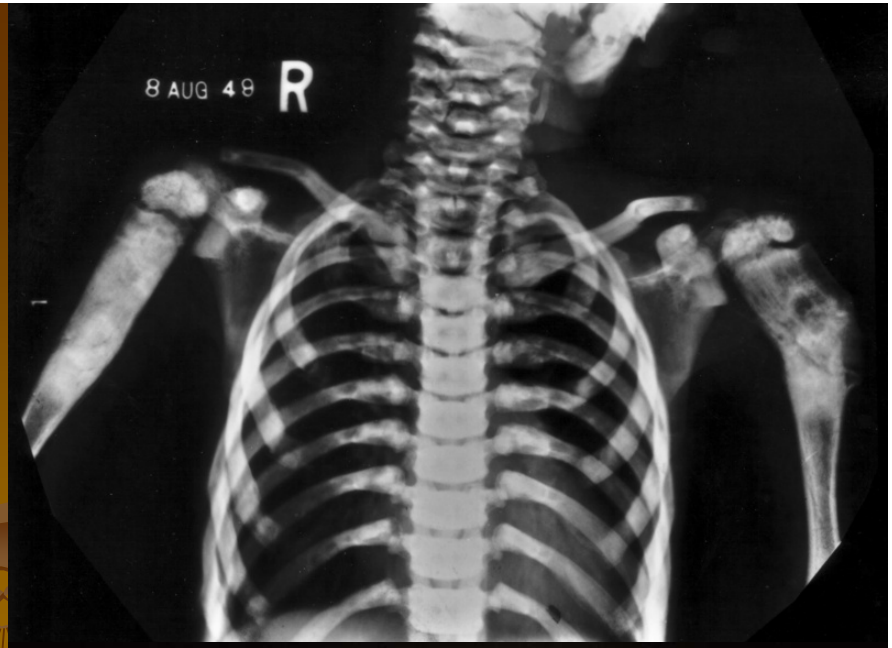
- late onset (sixth decade)
- Greater SNHL (with a descending pattern)
- enlarged calvaria
- enlargement and tortuosity of the superficial temporal artery and its anterior branches
- elevated serum alkaline phosphatase level
- radiographic evidence in the temporal bones



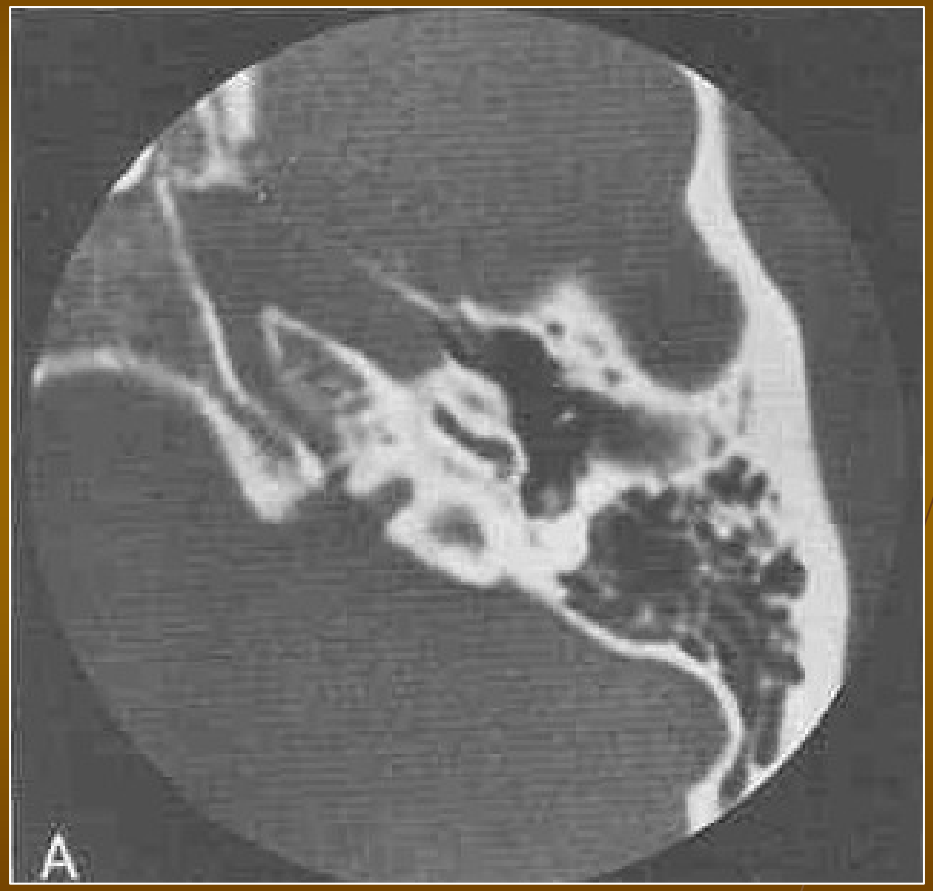
Osteopetrosis

- Uniformly increased density of all the bones and the lack of any cortical medullary differentiation

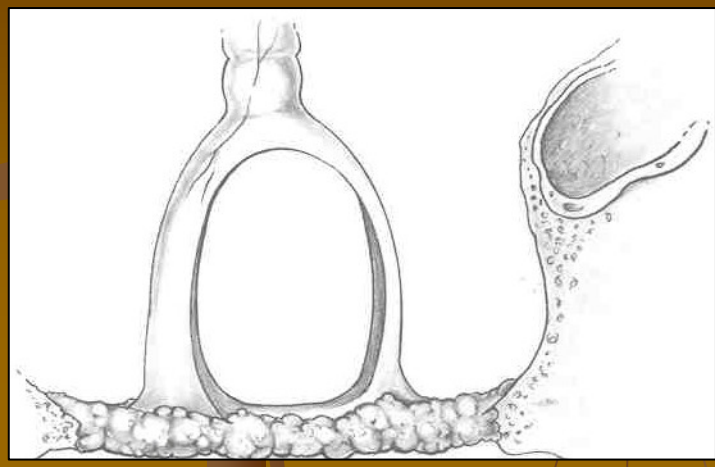
- Thickening of the calvarium with obliteration of the diploic layer



“Halo sign”



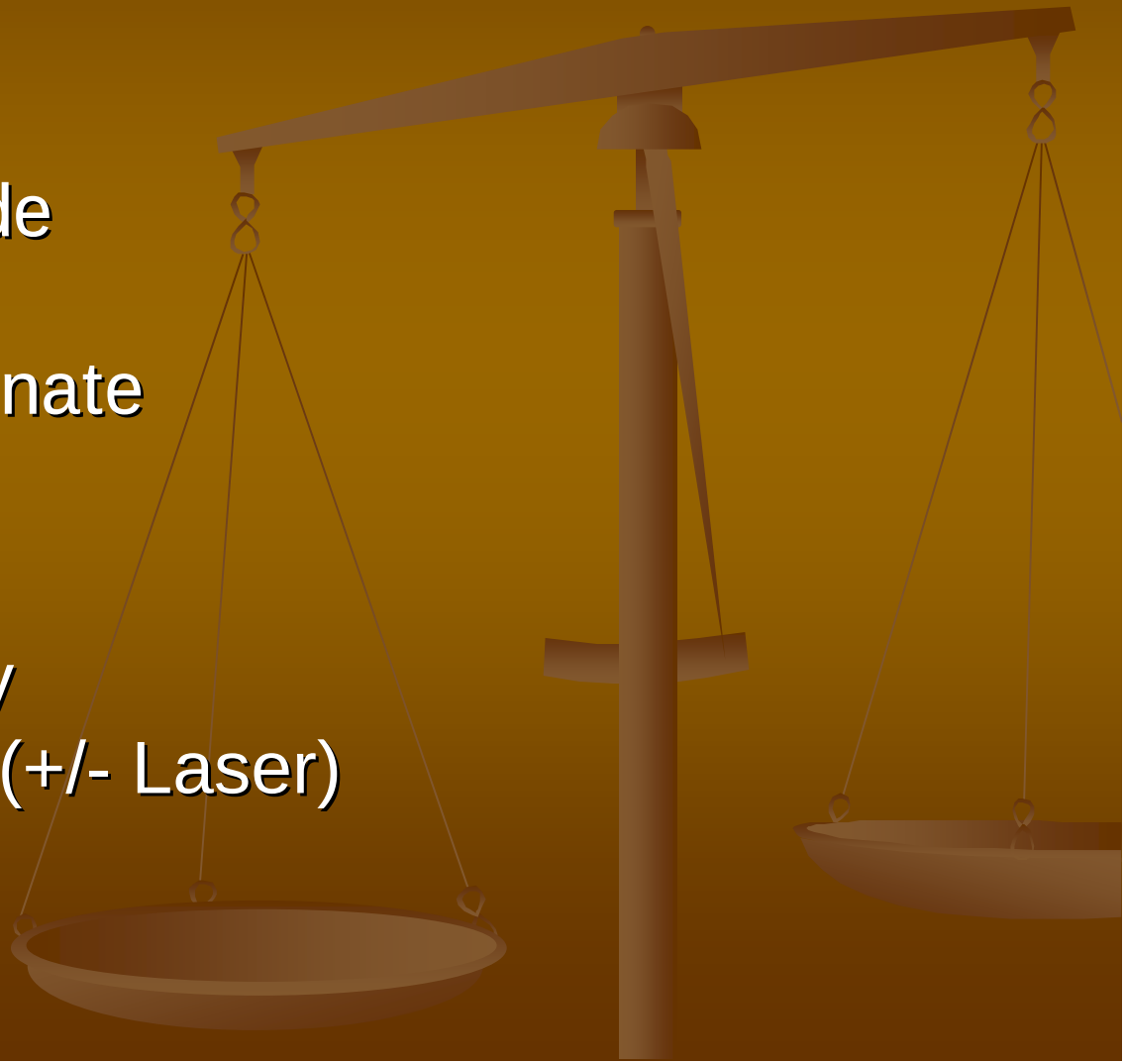
Objectives



- Introduction
- Pathology
- Diagnosis
- ***Treatment***

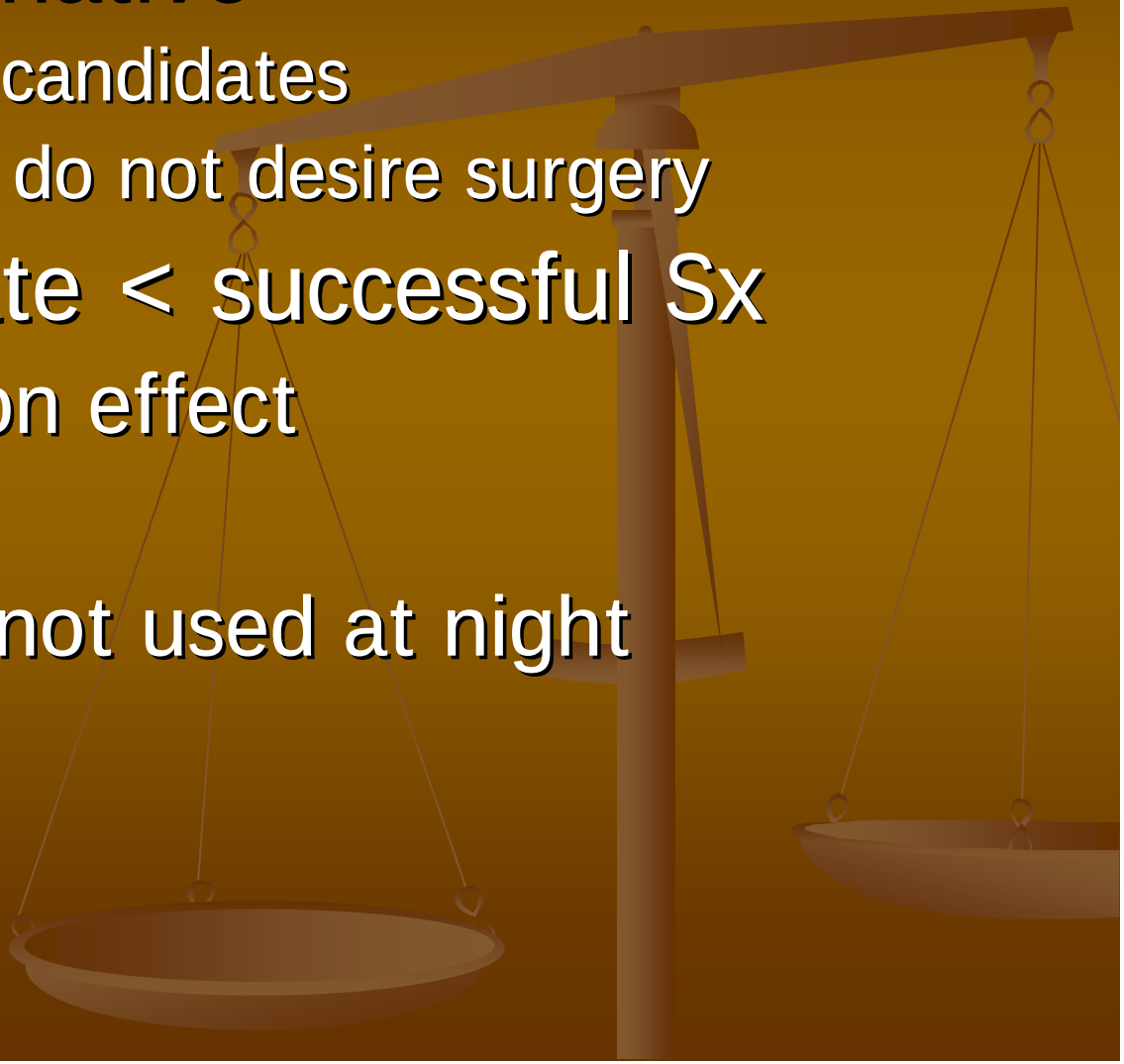
Treatment

1. Do nothing
2. Medication
   - Sodium fluoride
   - Vitamin D
   - Calcium carbonate
3. Amplification
4. Surgery
   - Stapedectomy
   - Stepedotomy (+/- Laser)



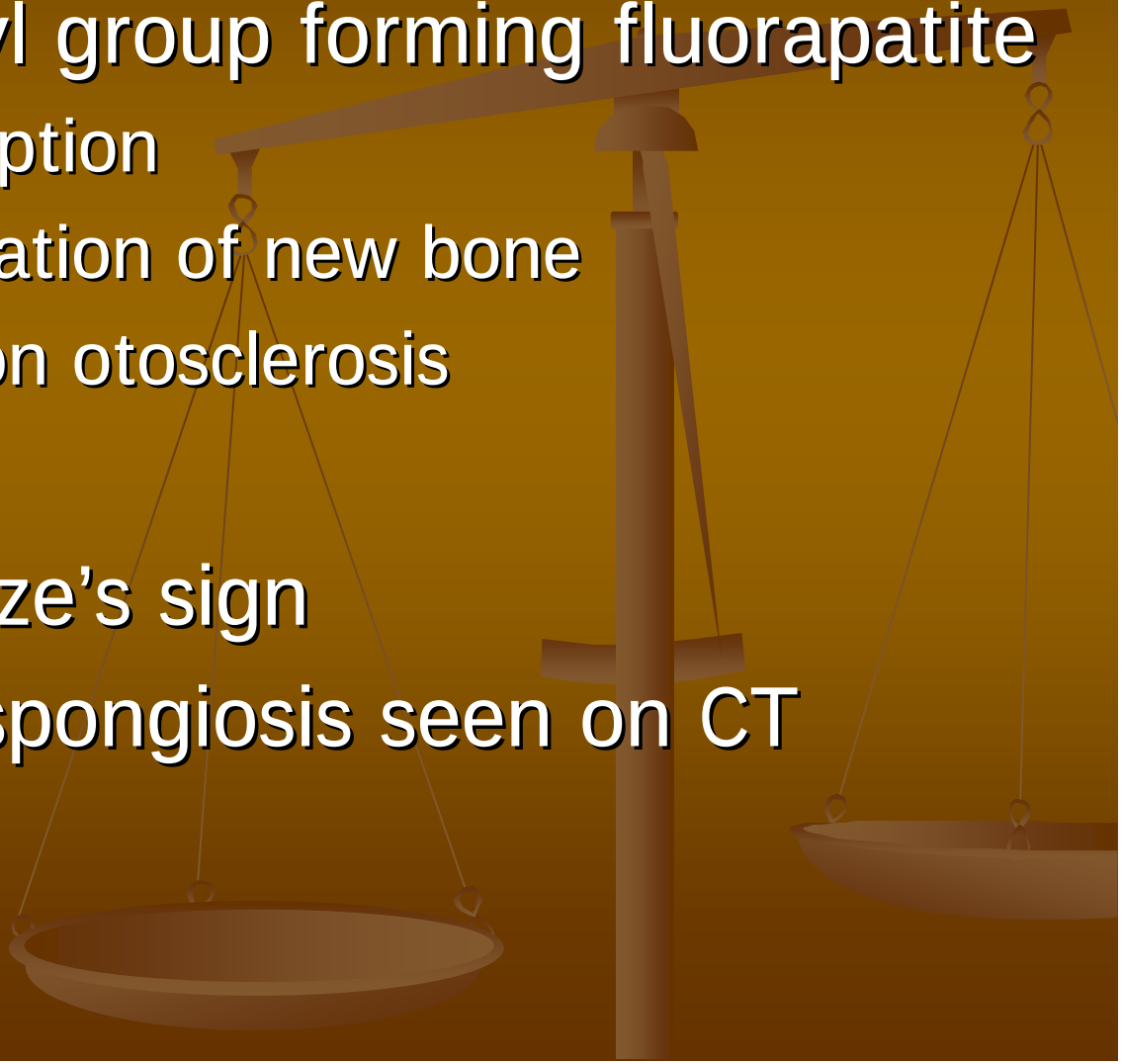
Amplification

- Excellent alternative
  - Non-surgical candidates
  - Patients who do not desire surgery
- Satisfaction rate < successful Sx
  - Canal occlusion effect
  - Quality
  - Amplification not used at night
  - Expensive
  - Cosmetic
- BAHA



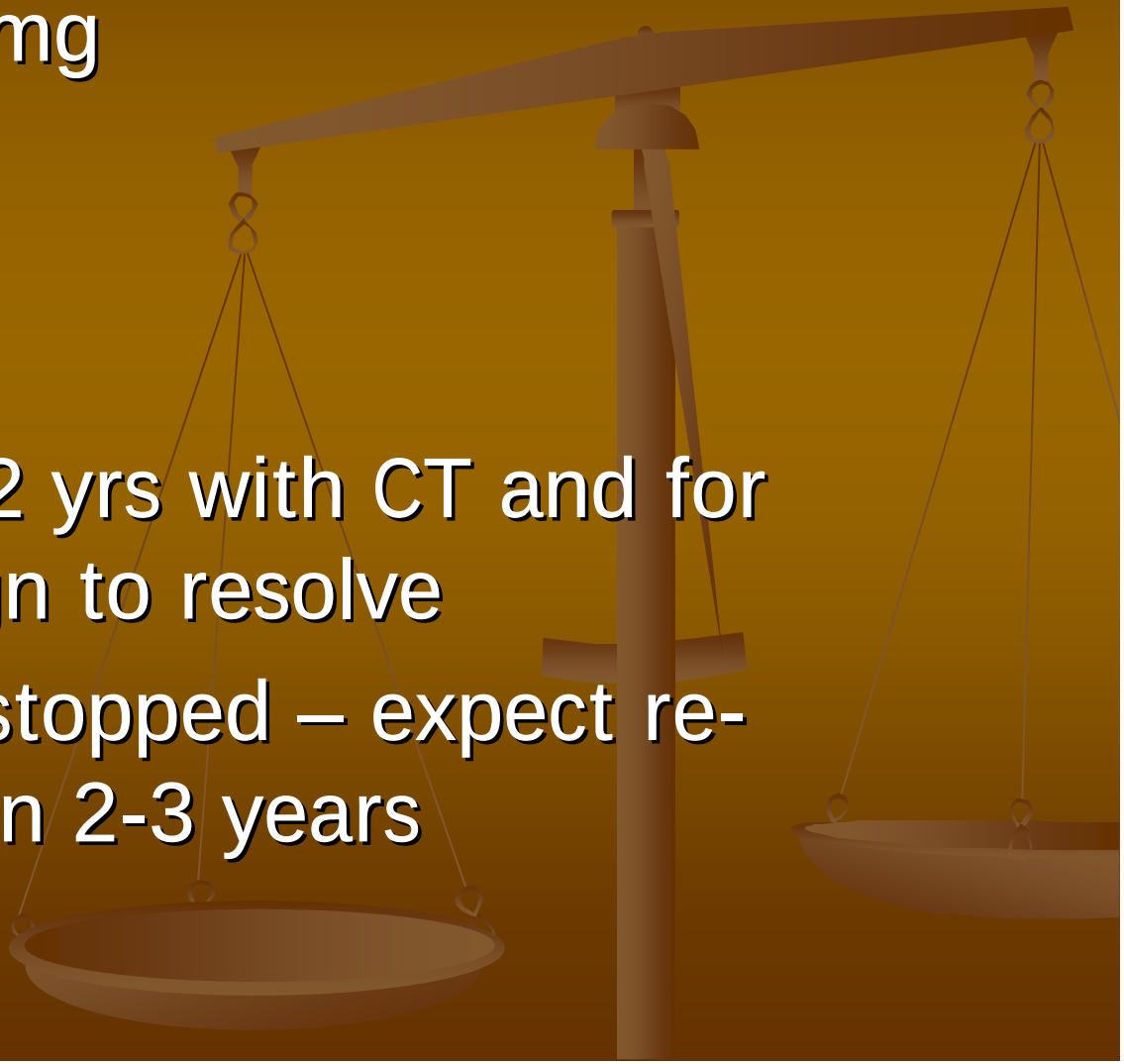
Sodium Fluoride

- Replaces hydroxyl group forming fluorapatite
  - resistant to resorption
  - Increases calcification of new bone
  - Causes maturation otosclerosis
- Reduces tinnitus
- reverses Schwartze's sign
- resolution of otospongiosis seen on CT



Sodium Fluoride

- Dose – 20-120mg
- Hearing results
  - 50% stabilize
  - 30% improve
- Re-evaluate q 2 yrs with CT and for Schwartze's sign to resolve
- If fluoride are stopped – expect re-activation within 2-3 years



Surgery

- Best surgical candidate
- Contraindications
- Procedure
- Complications
- Outcome & Prognosis

Surgery

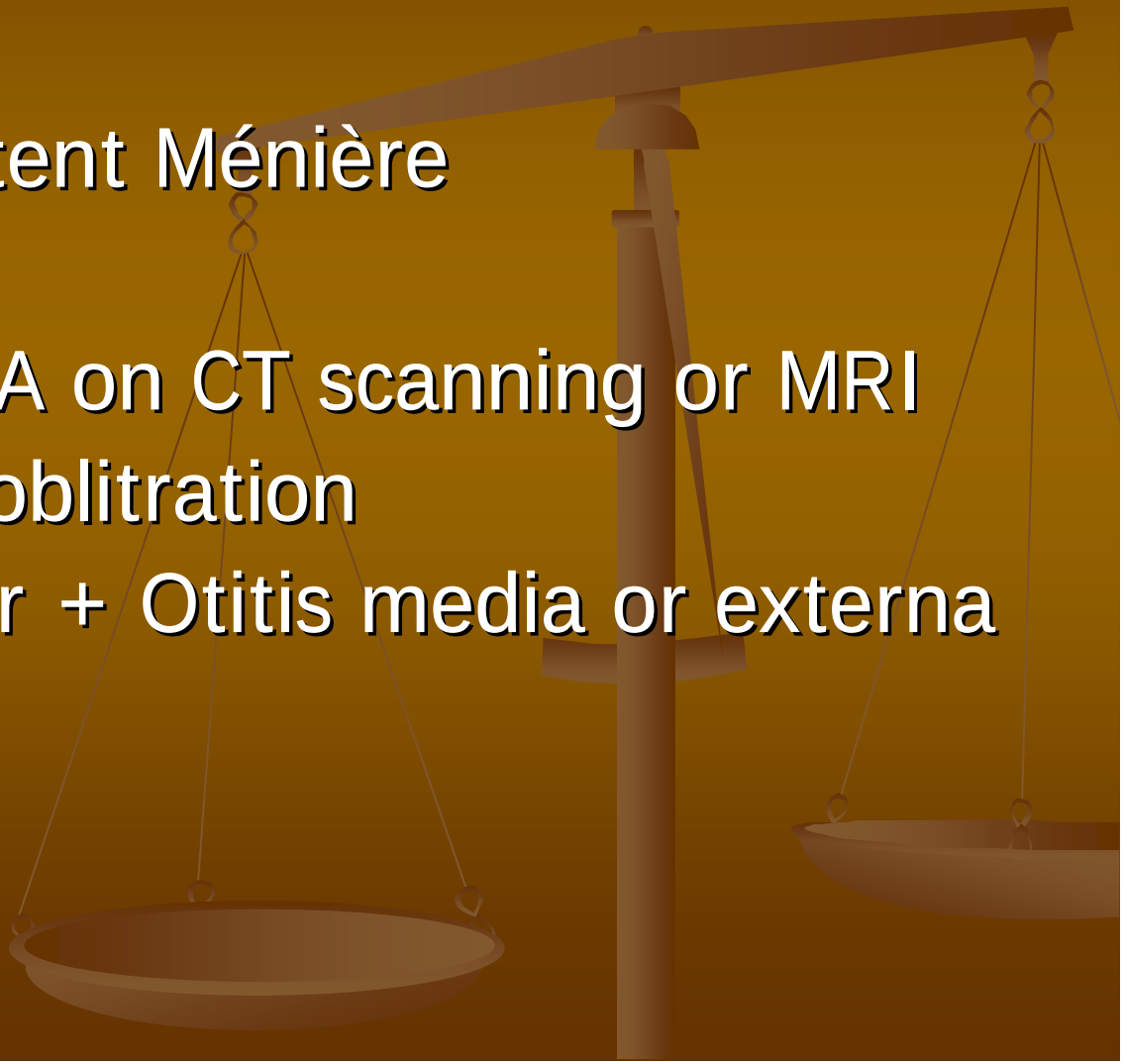
Best surgical candidate

- Previously un-operated ear
- Good health
- Negative Rinne test
- Excellent discrimination
- Desire for surgery

Contraindications

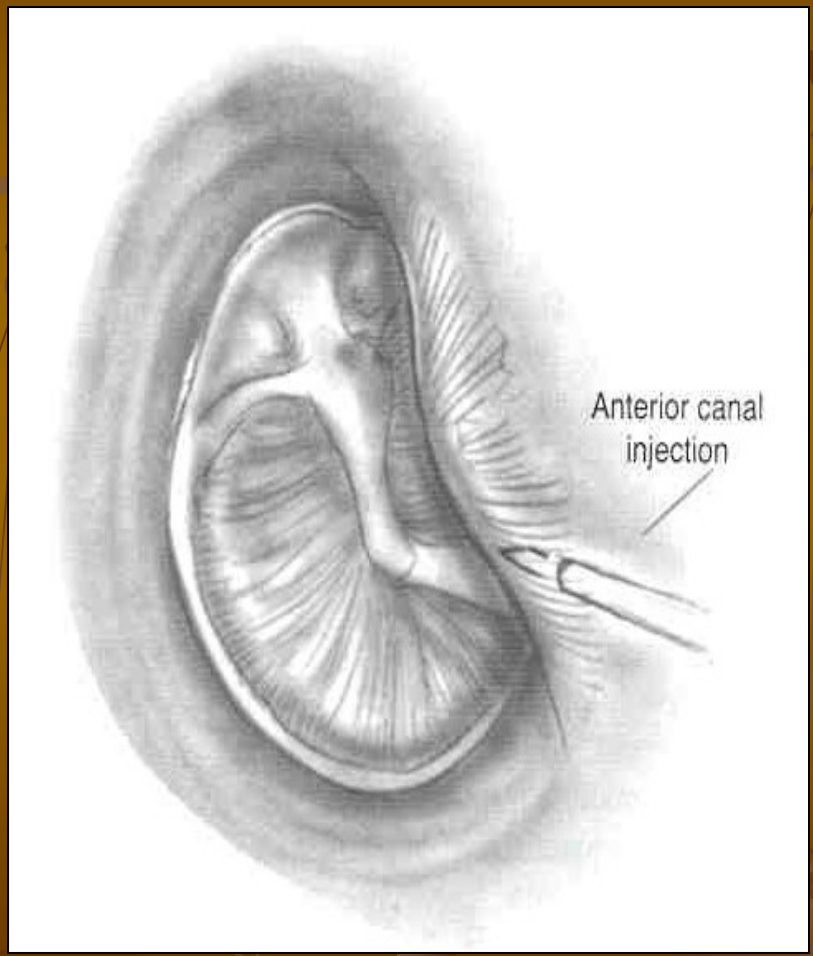
HYDROPS

- Hydrops Coexistent Ménière
- Young
- Dilation CA or VA on CT scanning or MRI
- Round window oblitration
- Only hearing ear + Otitis media or externa
- Perforation
- Silent < 20db



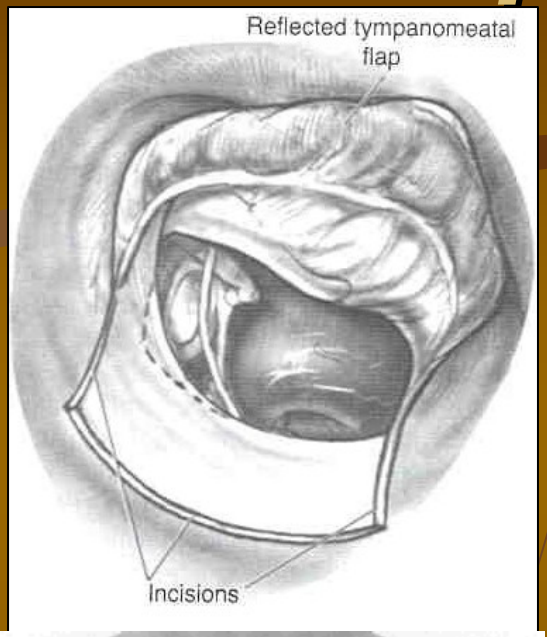
Canal Injection

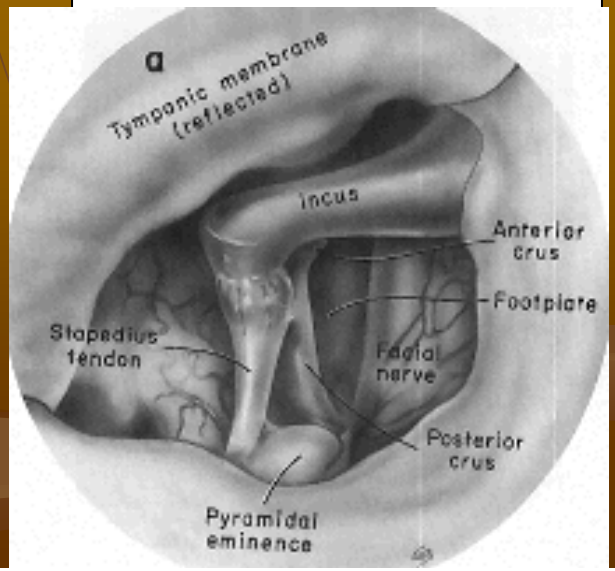
- 1% lidocaine with 1:100,000 epi
- 4 quadrants
- Bony cartilaginous junction



Raise Tympanomeatal Flap

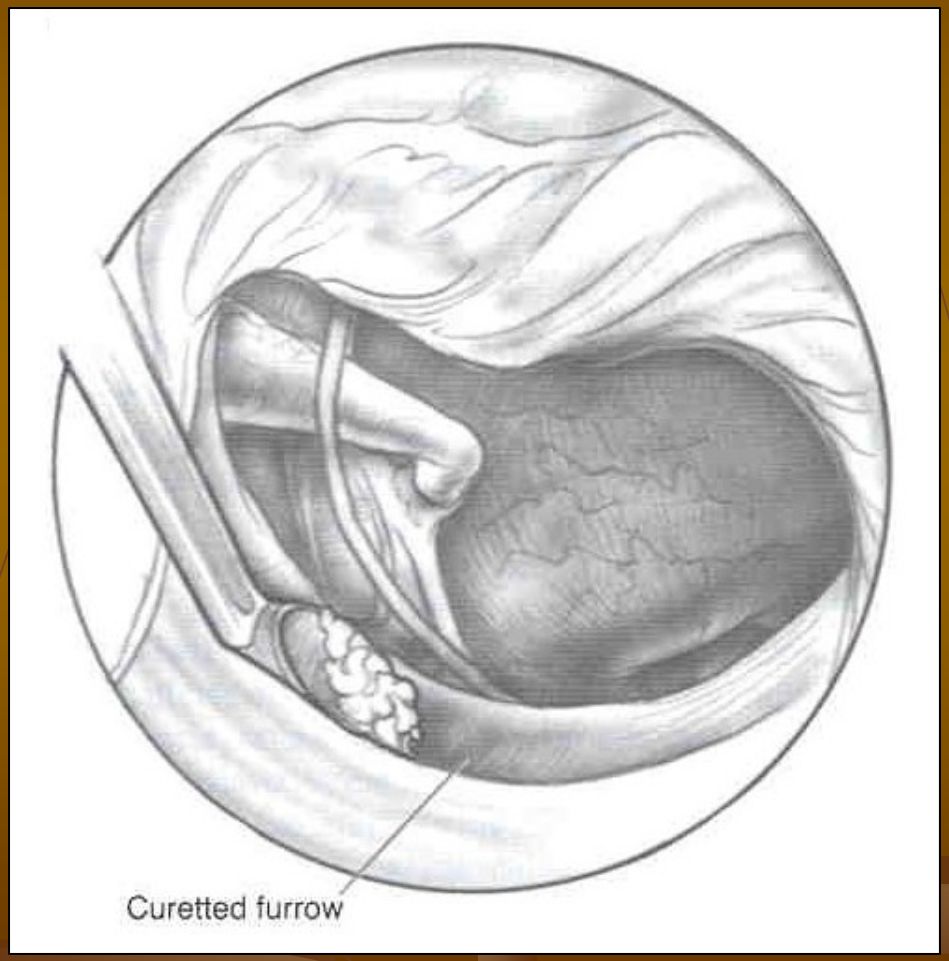
- 6 and 12 o'clock positions
- 6-8 mm lateral to the annulus
- Curettage of the scutum
- Exposure → Lt ear





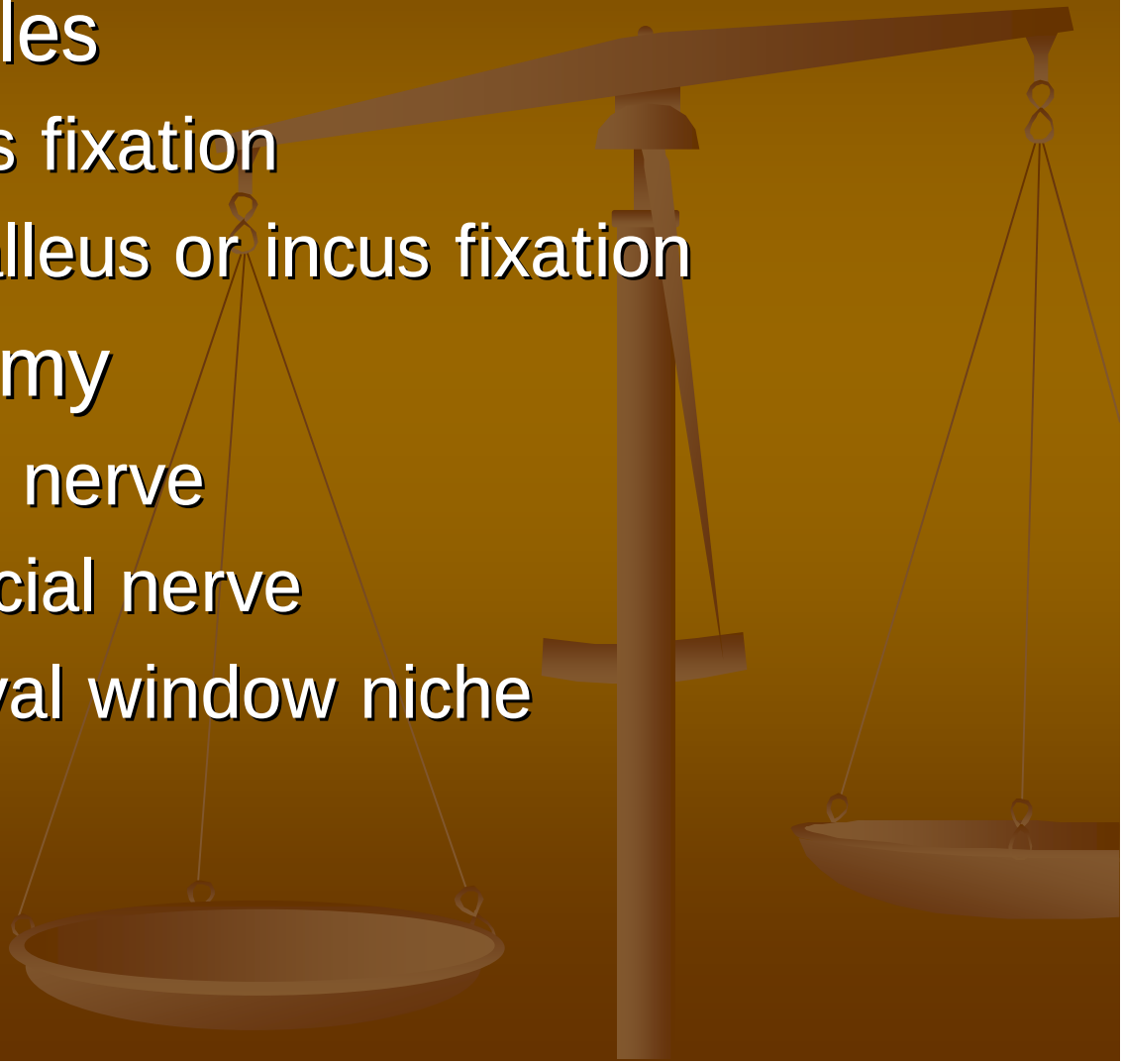
Curettage of Scutum

- Curettage a trough lateral to the scutum, thinning it
- Then remove the scutum (incus to the round window)
- Visualize the pyramidal process and facial n.



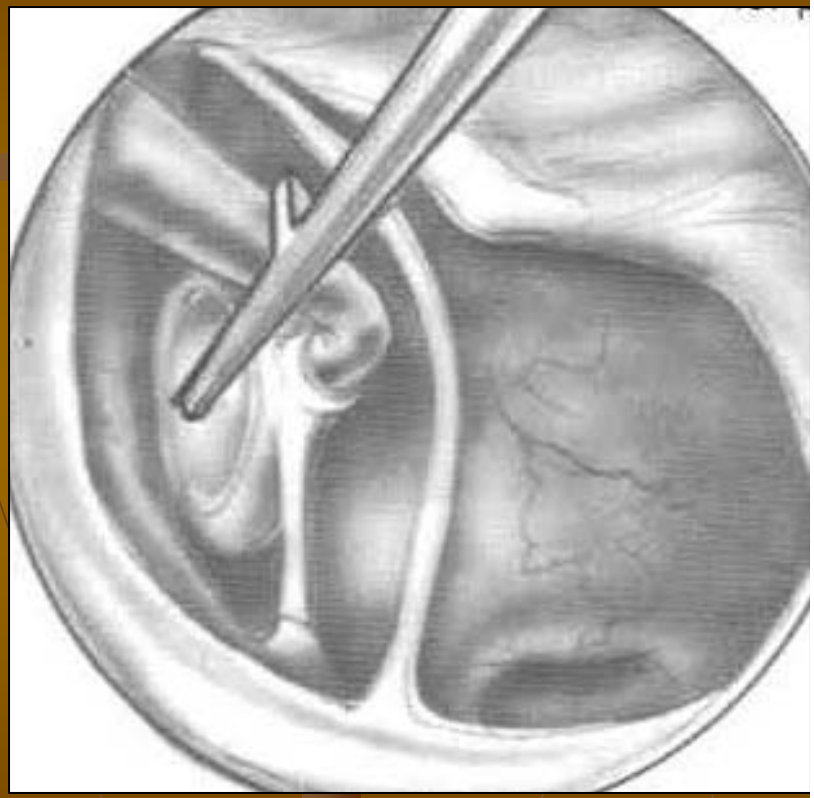
Middle ear examination

- Mobility of ossicles
  - **Confirm** stapes fixation
  - Evaluate for malleus or incus fixation
- Abnormal anatomy
  - Dehiscent facial nerve
  - Overhanging facial nerve
  - Deep narrow oval window niche



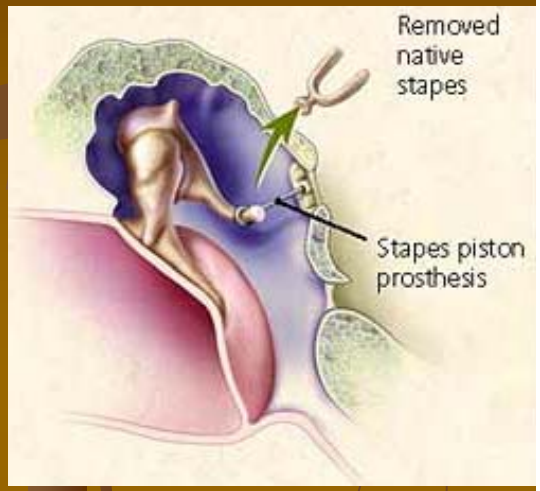
Measurement for prosthesis

- Lateral aspect of the long process of the incus to the footplate
- Add 0.25 mm
- Average 4.5 mm
- Diameter 0.6 / 0.8 mm

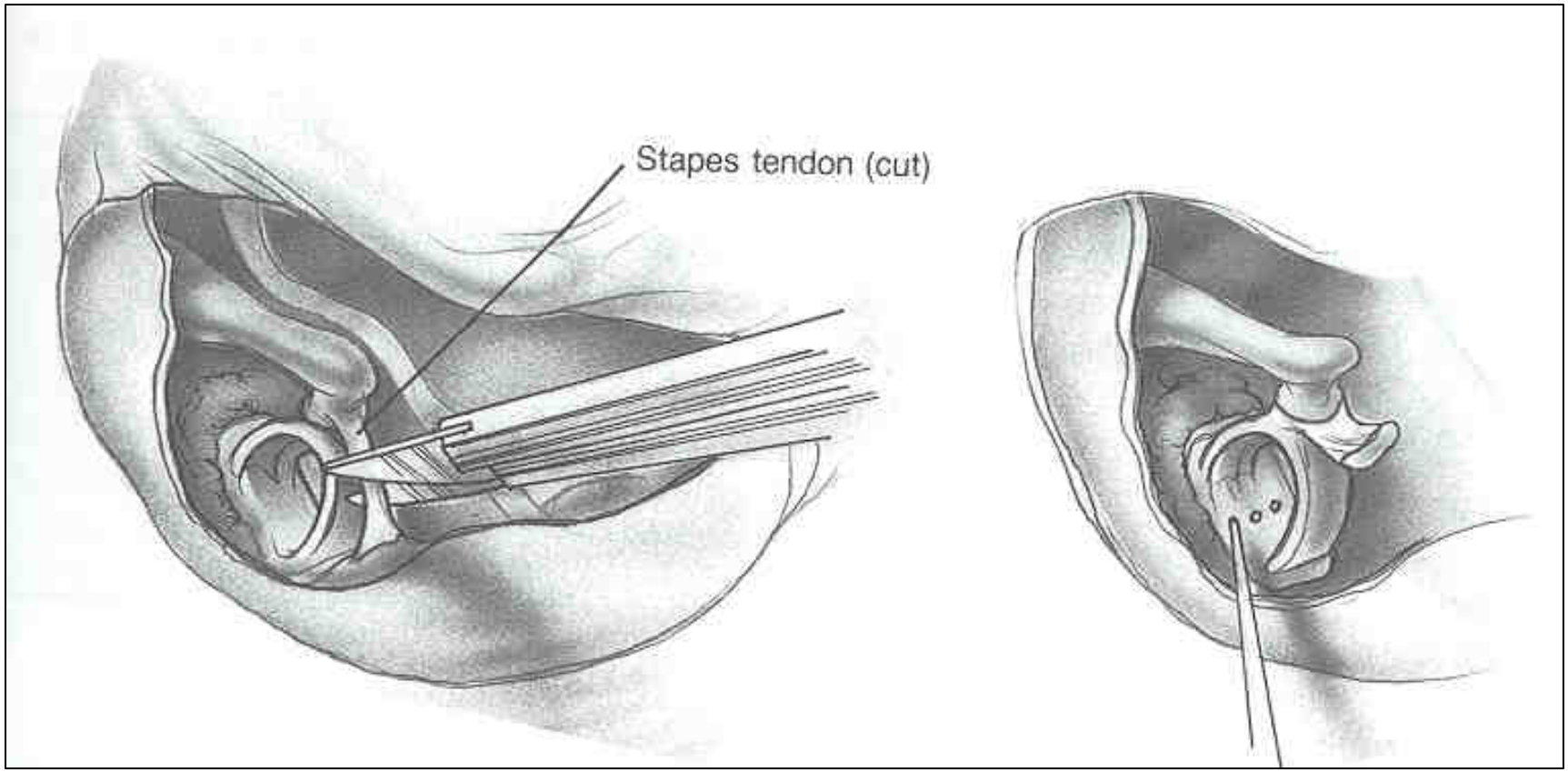


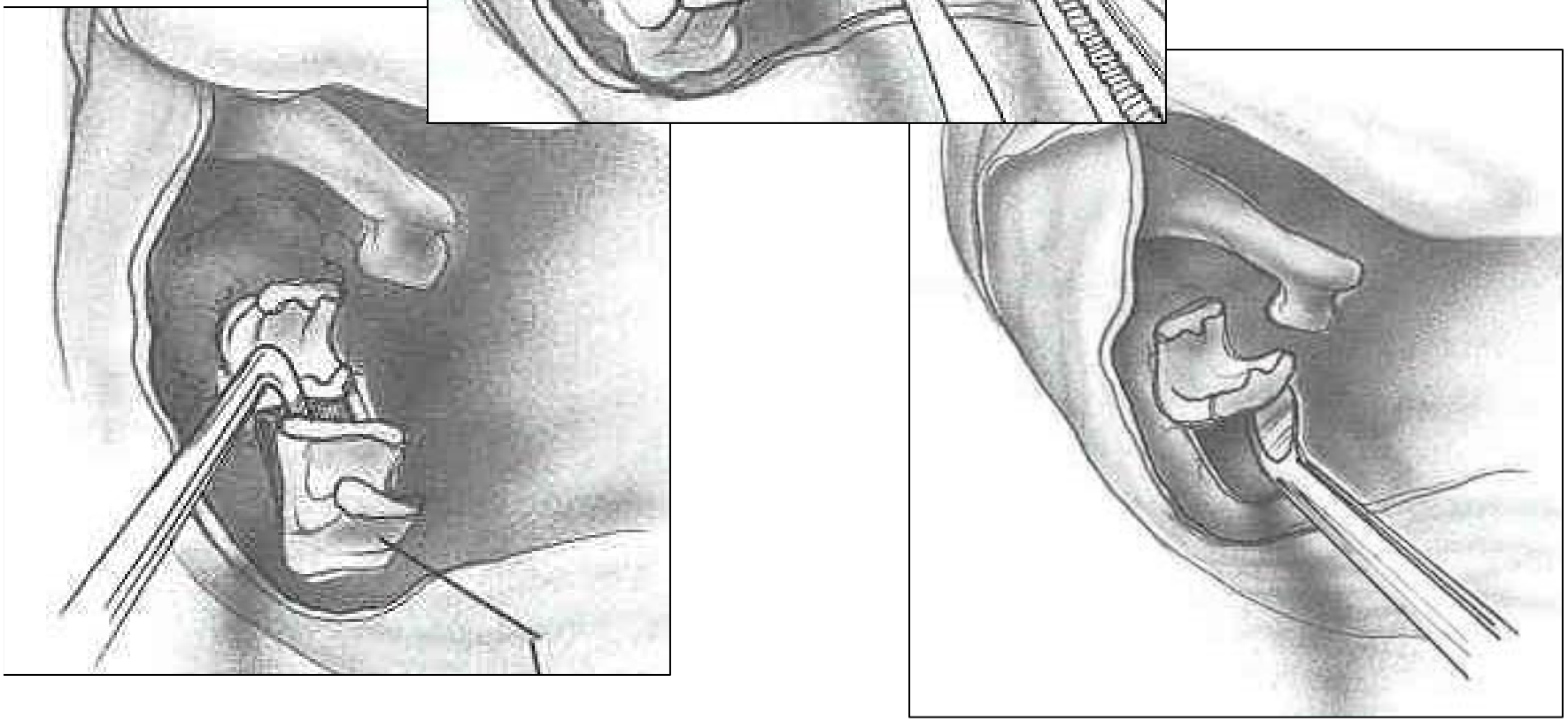
Total Stapedectomy

- Uses
  - Surgeon preference
  - Floating footplate
  - Same result



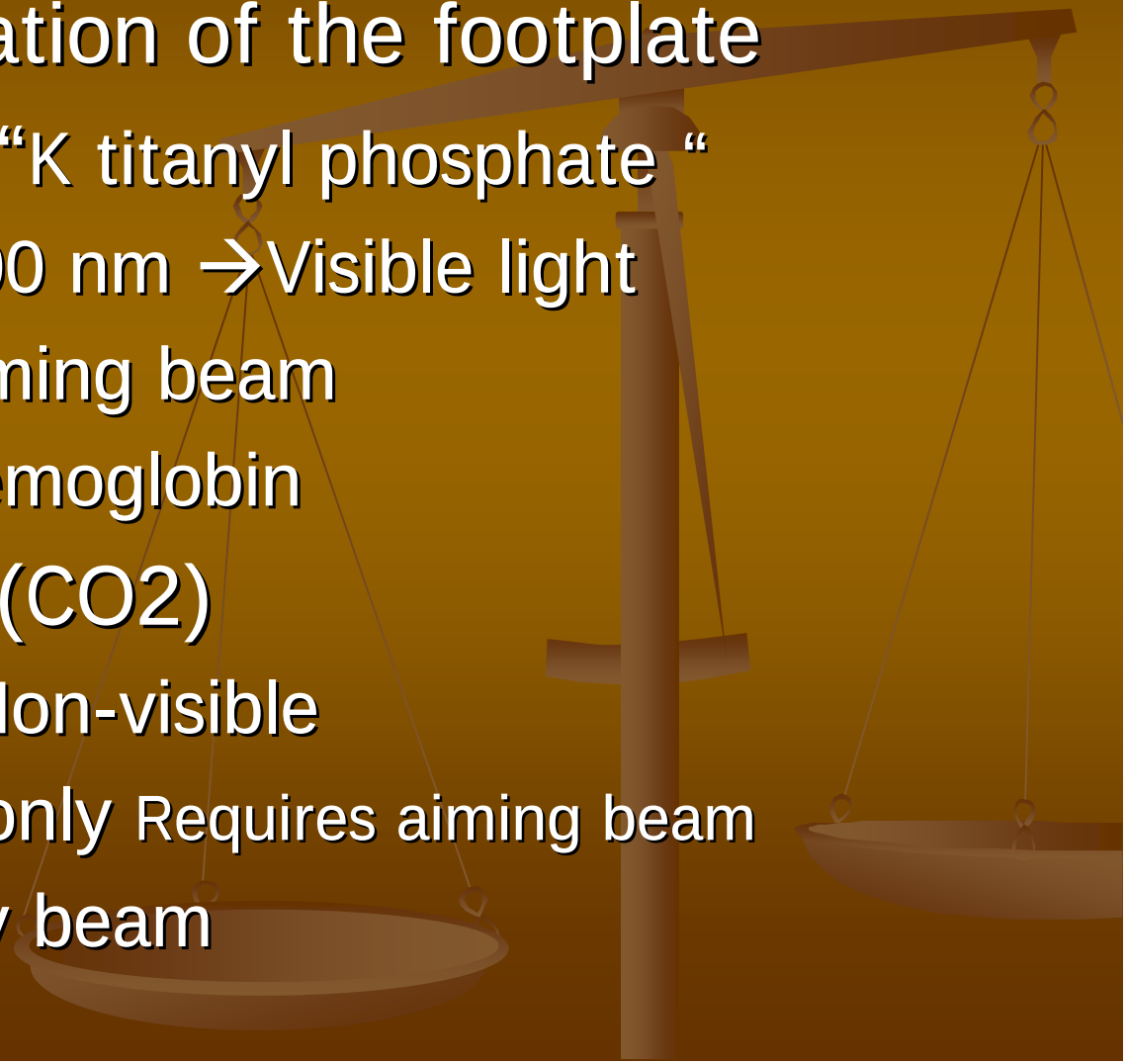
- Disadvantages
  - Increased post-op vestibular symptoms
  - More technically difficult
  - Increased potential for prosthesis migration


Stapes tendon (cut)

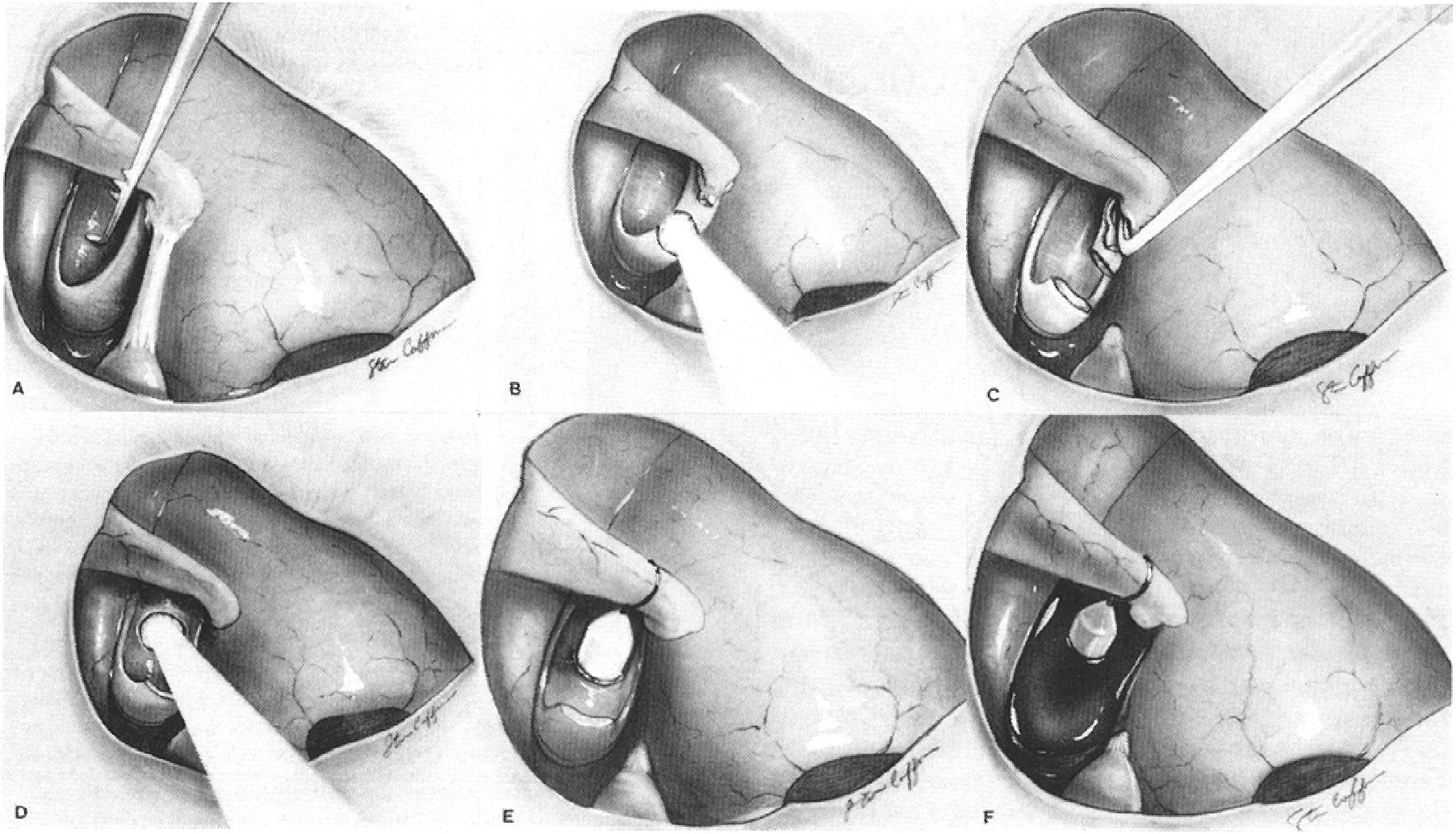


Laser Fenestration

- Avoids manipulation of the footplate
- Argon and KTP “K titanyl phosphate “
  - Wave length 500 nm →Visible light
  - Surgical and aiming beam
  - Absorbed by hemoglobin
- Carbon dioxide (CO2)
  - 10,000 nm → Non-visible
  - Surgical beam only Requires aiming beam
  - Ill defined fuzzy beam



Laser Stapedotomy

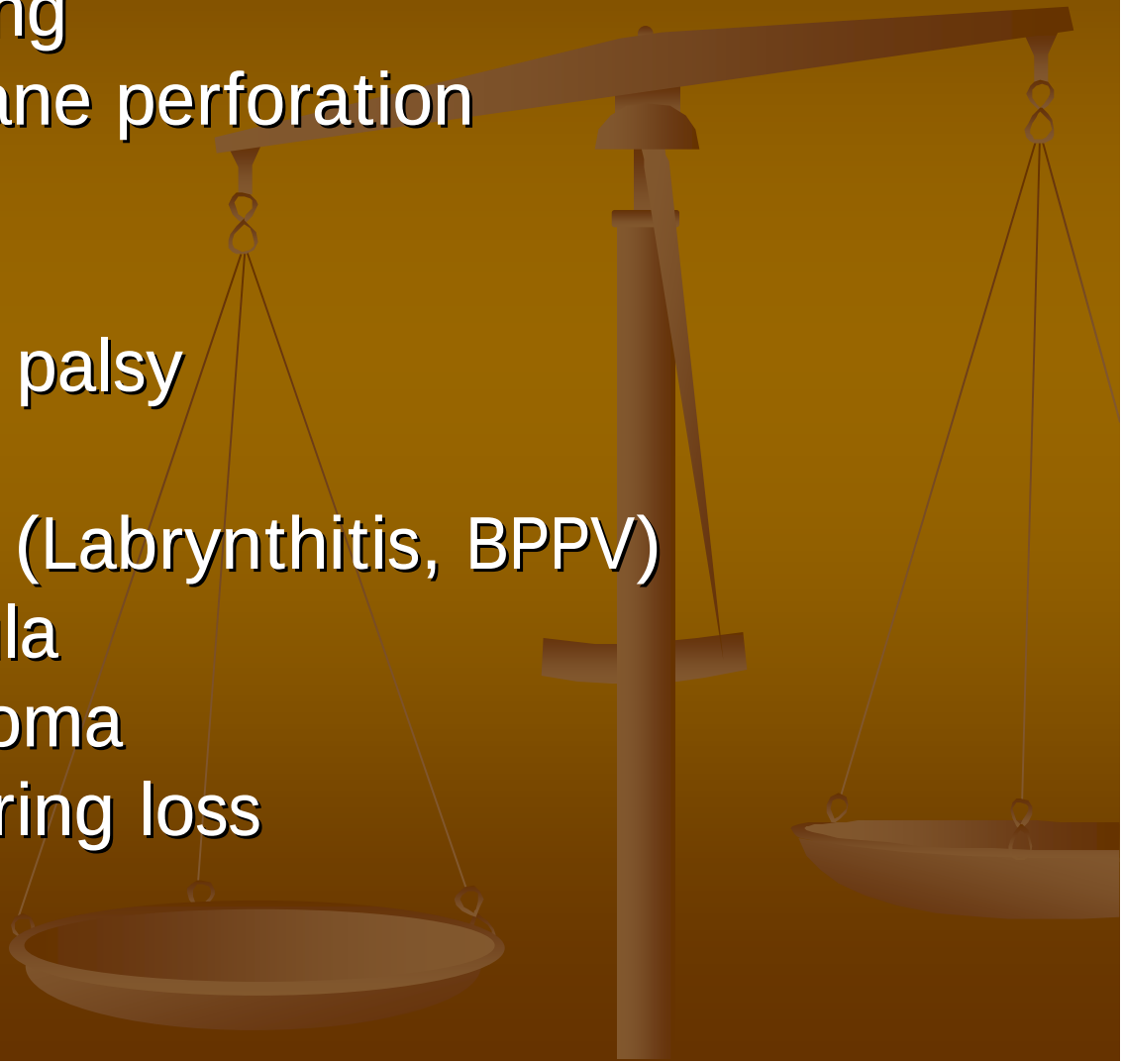


Surgery

- Best surgical candidate
- Contraindications
- Procedure
- ***Complications***
- Outcome & Prognosis

Complications

- Infection & Bleeding
- Tympanic membrane perforation
- Taste disturbance
- Ossicle trauma
- 0.5% Facial nerve palsy
- Tinnitus
- Significant vertigo (Labrynthitis, BPPV)
- Perilymphatic fistula
- Reparative granuloma
- Sensorineural hearing loss
- Delayed failure



Perilymphatic fistula

- 3-10% of
- Fluctuating SNHL
- Vertigo.
- Total stapedectomies.
- Absorbable gelatin sponge

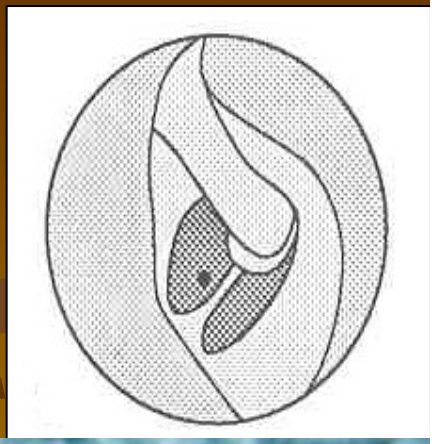
Overhanging Facial Nerve

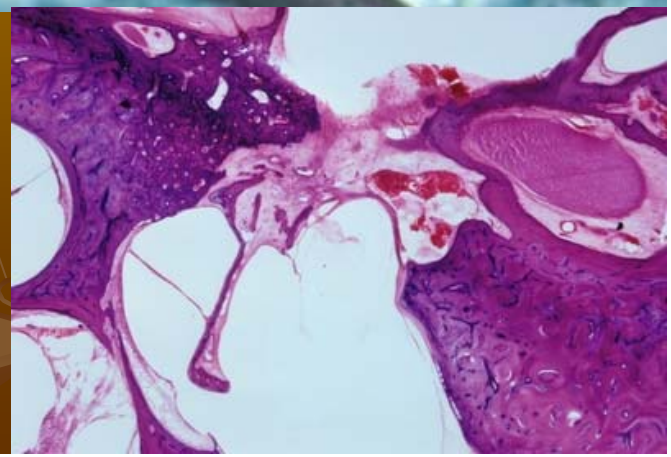
- Usually dehiscent
- Consider aborting the procedure
- Facial nerve displacement (Perkins, 2001)
  - Facial nerve is compressed superiorly with No. 24 suction (5 second periods)
  - 10-15 sec delay between compressions
- Wire piston used
  - Add 0.5 to 0.75 mm to accommodate curve around the nerve

Floating Footplate

- Prevention
  - ABG > 20db (-ve R)
  - Laser
  - Footplate control hole →
  - “biscuit” footplate →
- Management
  - Abort → re-fix → re-operat
  - promontory fenestration
  - laser fenestration

H House→ worse enemy





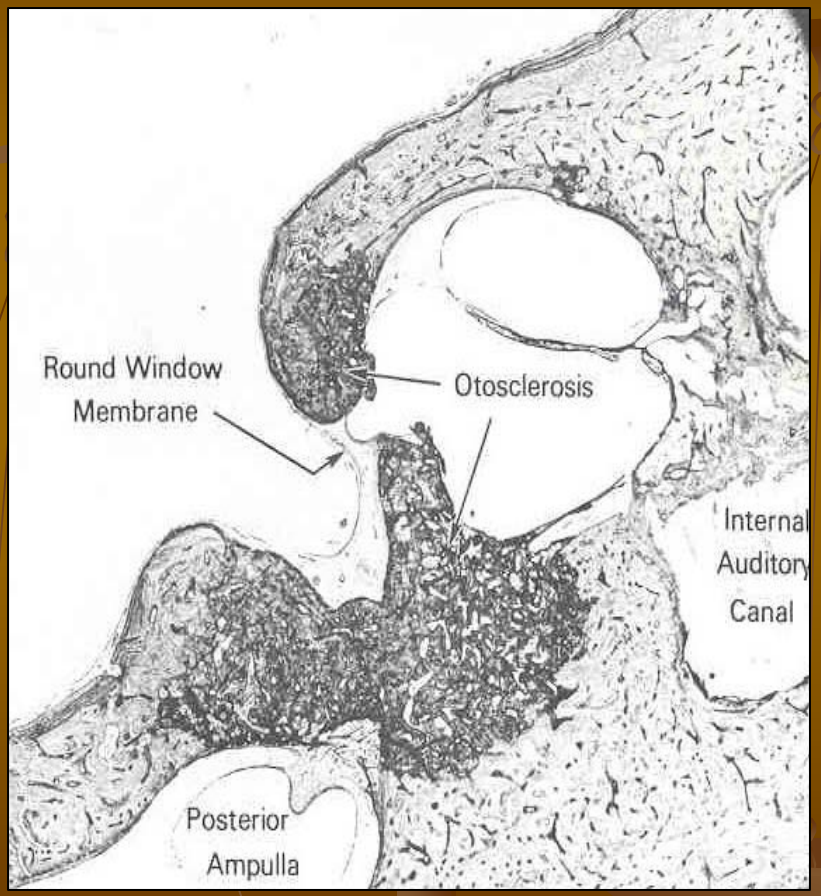
Perilymphatic Gusher

- Associated with patent cochlear aqueduct*
- More common on the left
- Increased with congenital* stapes fixation
- Increases risk of SNHL
- Management
  - Ruff up the footplate
  - Rapid placement of the OW seal then the prosthesis
  - HOB elevated, stool softeners, bed rest, avoid Valsalva, +/- lumbar drain

*+OM Sx is C/I in children

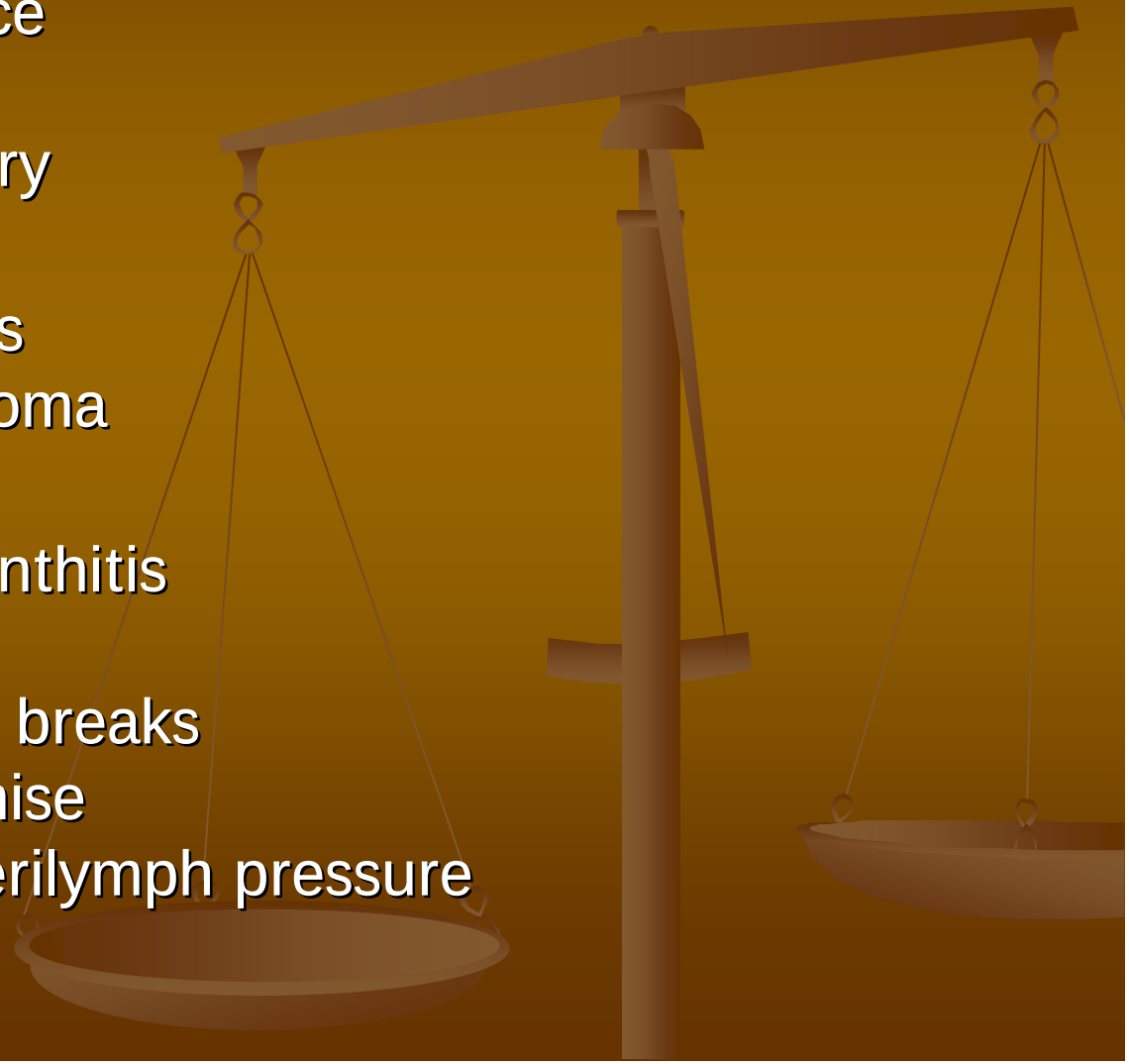
Round Window Closure

- 20%-50% of cases
- No effect on hearing unless 100% closed
- 1% completely closed
- Opening has a high rate of SNHL

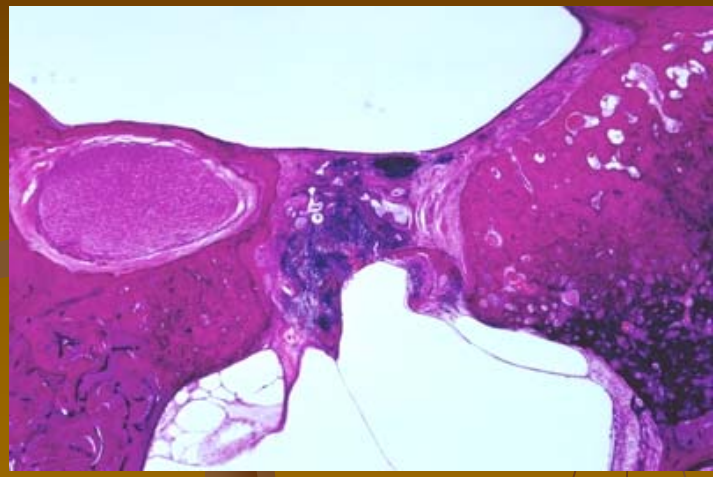


SNHL

- 1%-3% incidence of profound permanent SNHL
  - Surgeon experience
  - Extent of disease
  - Prior stapes surgery
- Temporary
  - Serous labyrinthitis
  - Reparative granuloma
- Permanent
  - Suppurative labyrinthitis
  - Extensive drilling
  - Basilar membrane breaks
  - Vascular compromise
  - Sudden drop in perilymph pressure



Reparative Granuloma



- 2 -3 weeks postop
- ? Gelfome
- Initial good hearing results
- → increase in the HF-SNHL
- Associated tinnitus and vertigo
- Exam – reddish discoloration of the posterior TM
- Treatment
  - ME exploration
  - Removal of granuloma
- Prognosis – return of hearing with early excision

Vertigo

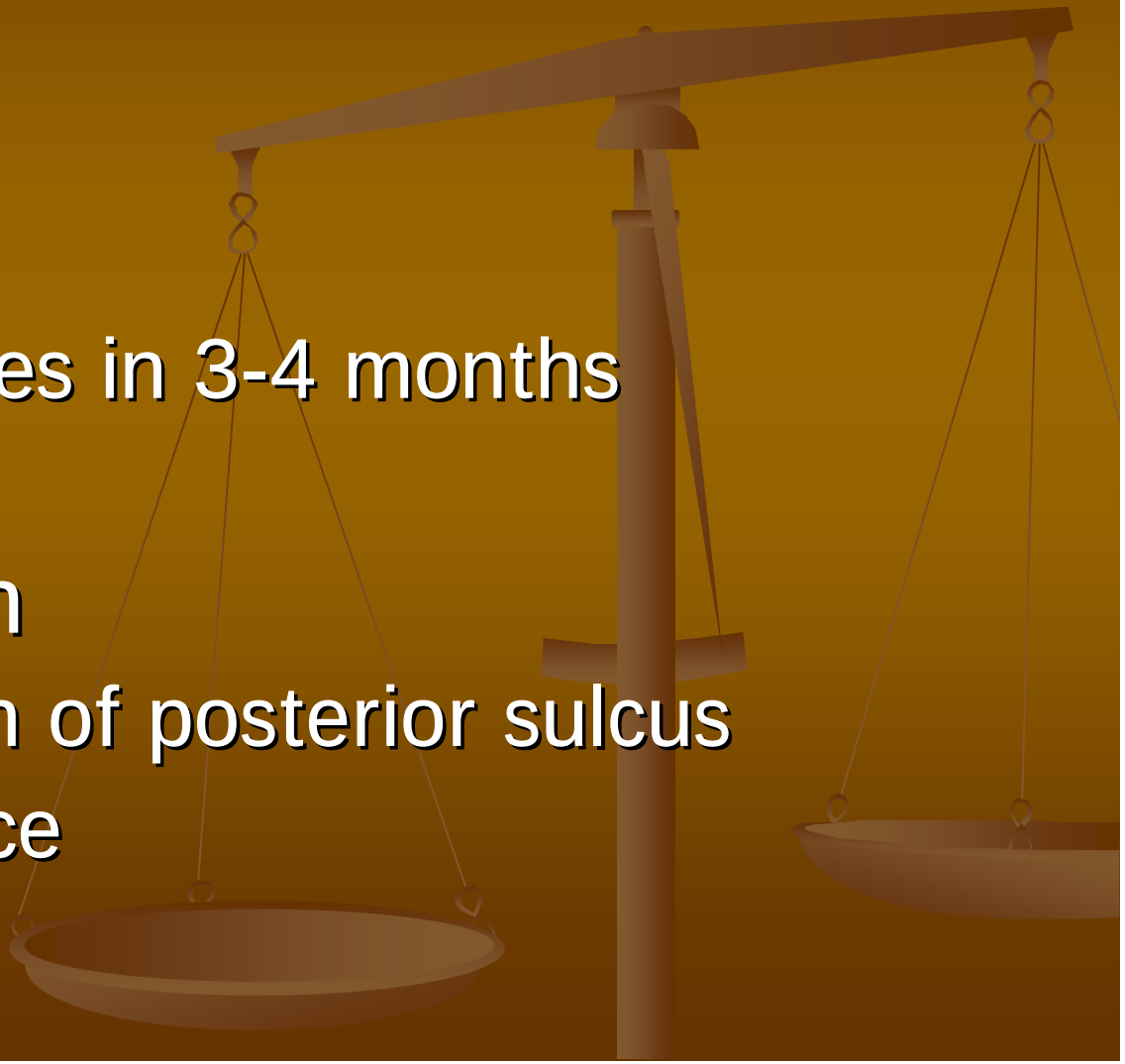
- Most commonly short lived (2-3 days)
- Stapedectomy > stapedotomy > Laser
  - Due to serous labyrinthitis
- Medialization of the prosthesis into the vestibule
  - With or without perilymphatic fistula
- Reparative granuloma
- *Other side Sx

Recurrent Conductive Hearing Loss

- Slippage from incus/FP
- Incus erosion
- Adherence to edge of OW niche
- Re-obliteration of OW
- Malleus or incus ankylosis
- Slippage or displacement of the prosthesis

Other Cx

- Chorda injury
  - 30% of cases
  - metallic taste
  - usually resolves in 3-4 months
  - Stretching
- TM perforation
  - With elevation of posterior sulcus
  - 1.9% incidence
  - repair



Outcome & Prognosis

Commonly quoted statistics indicate that

- 90% significant hearing improvement.
- 8% no significant hearing improvement.
- 2% additional HL
- Revision stapedectomy
  - Successful results in 65%
  - SNHL in 3 to 20%

- Stapedectomy, in experienced hands, is generally considered a safe procedure.

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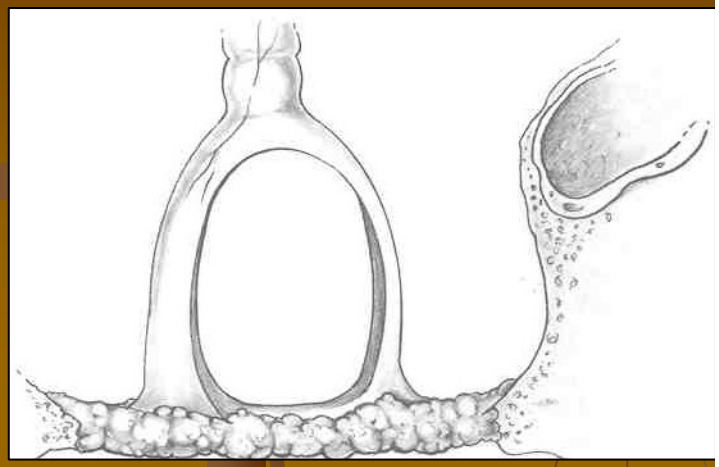
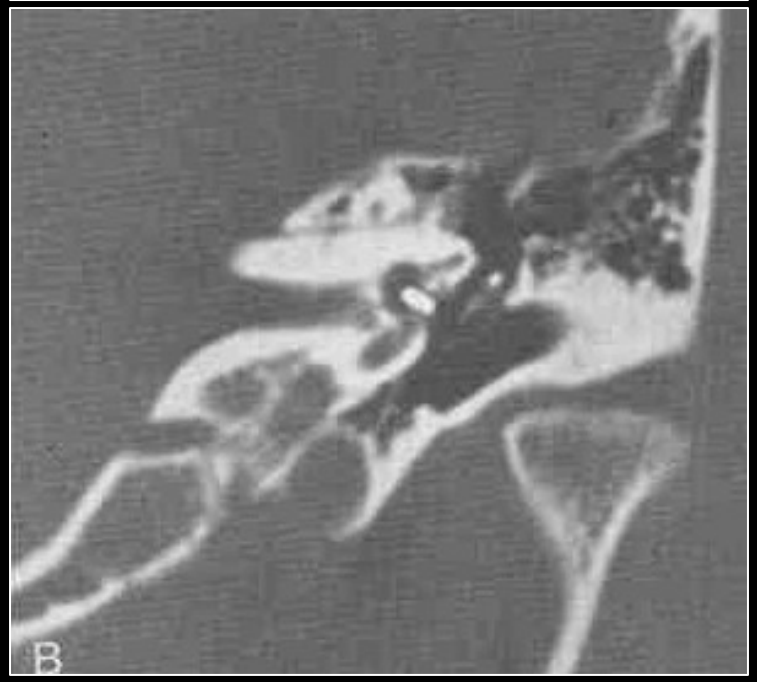


What do you think?


A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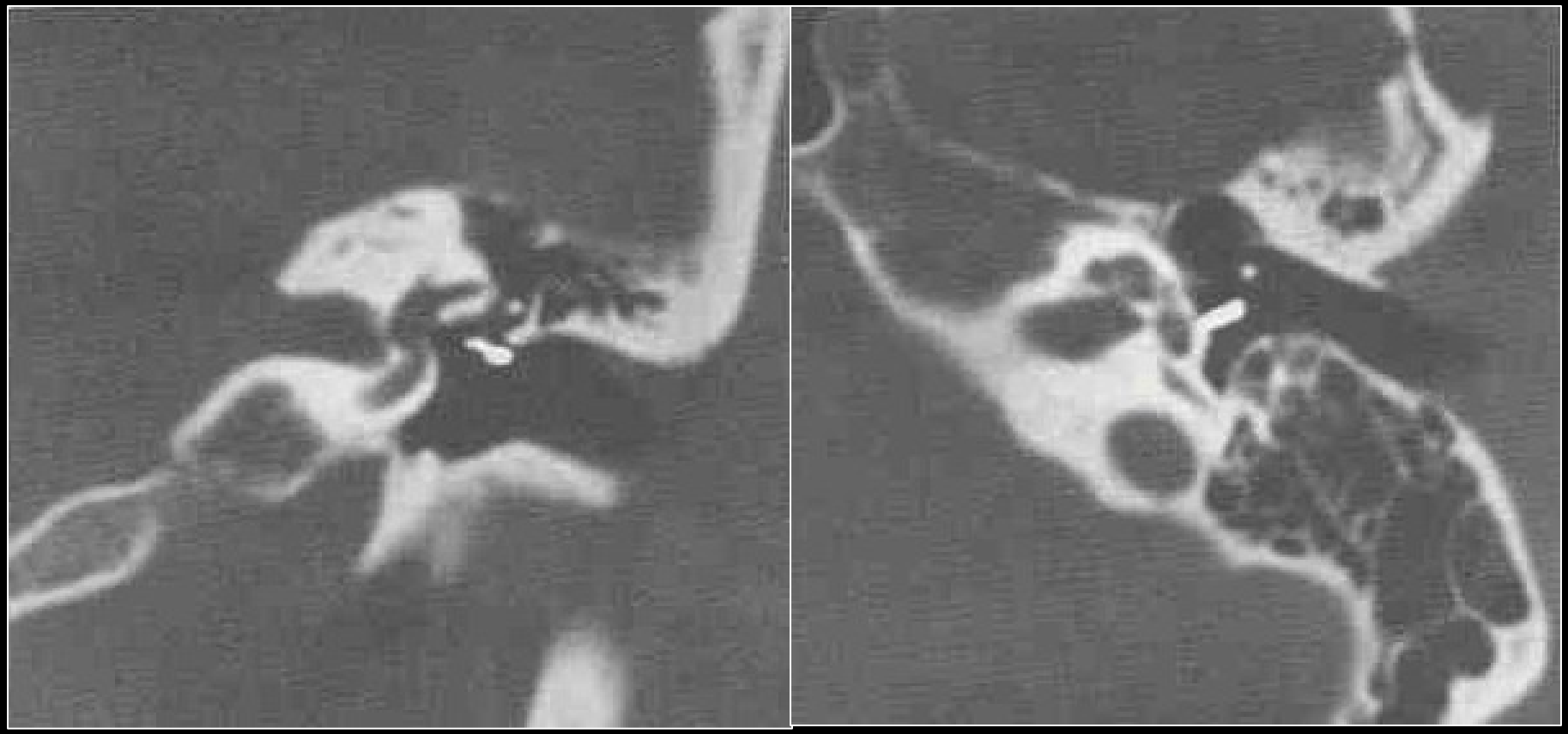

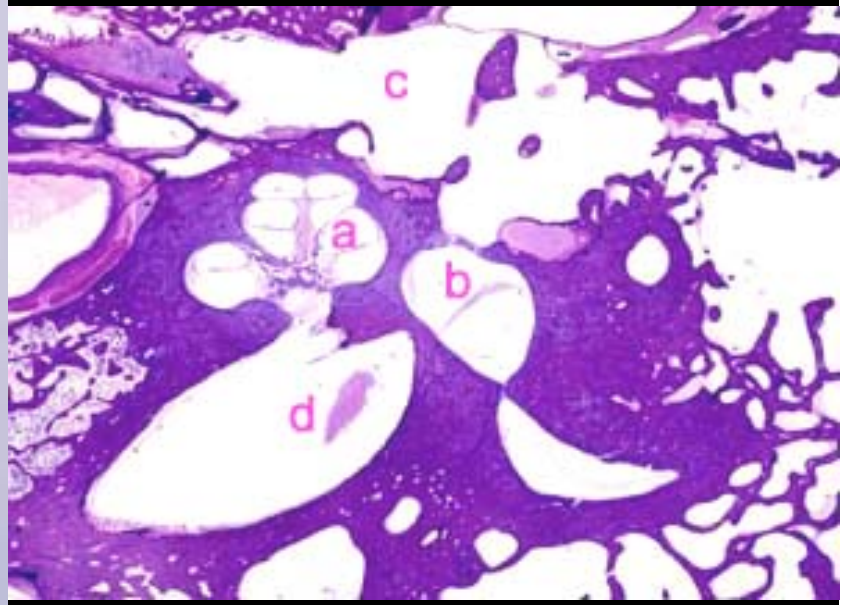
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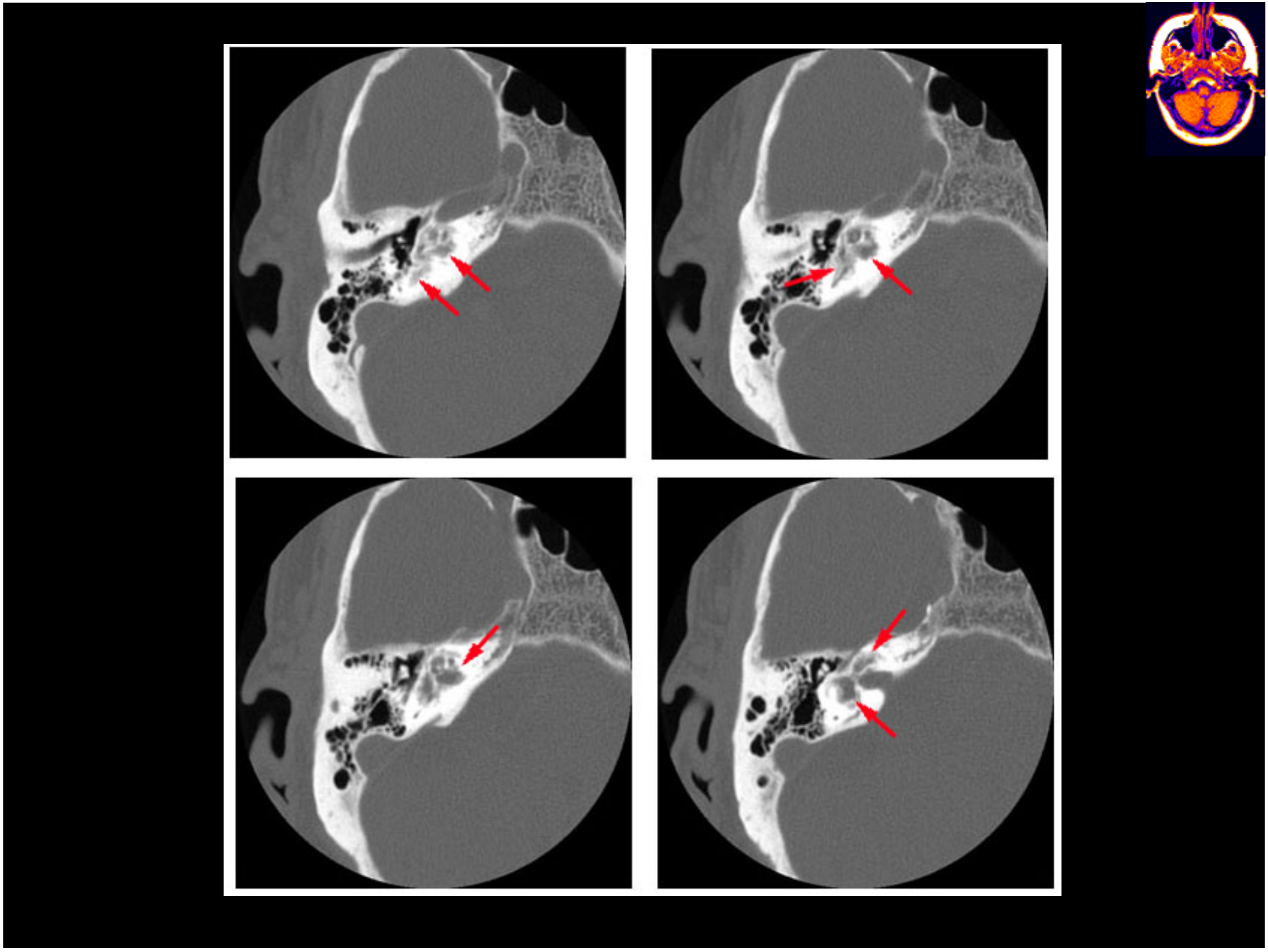


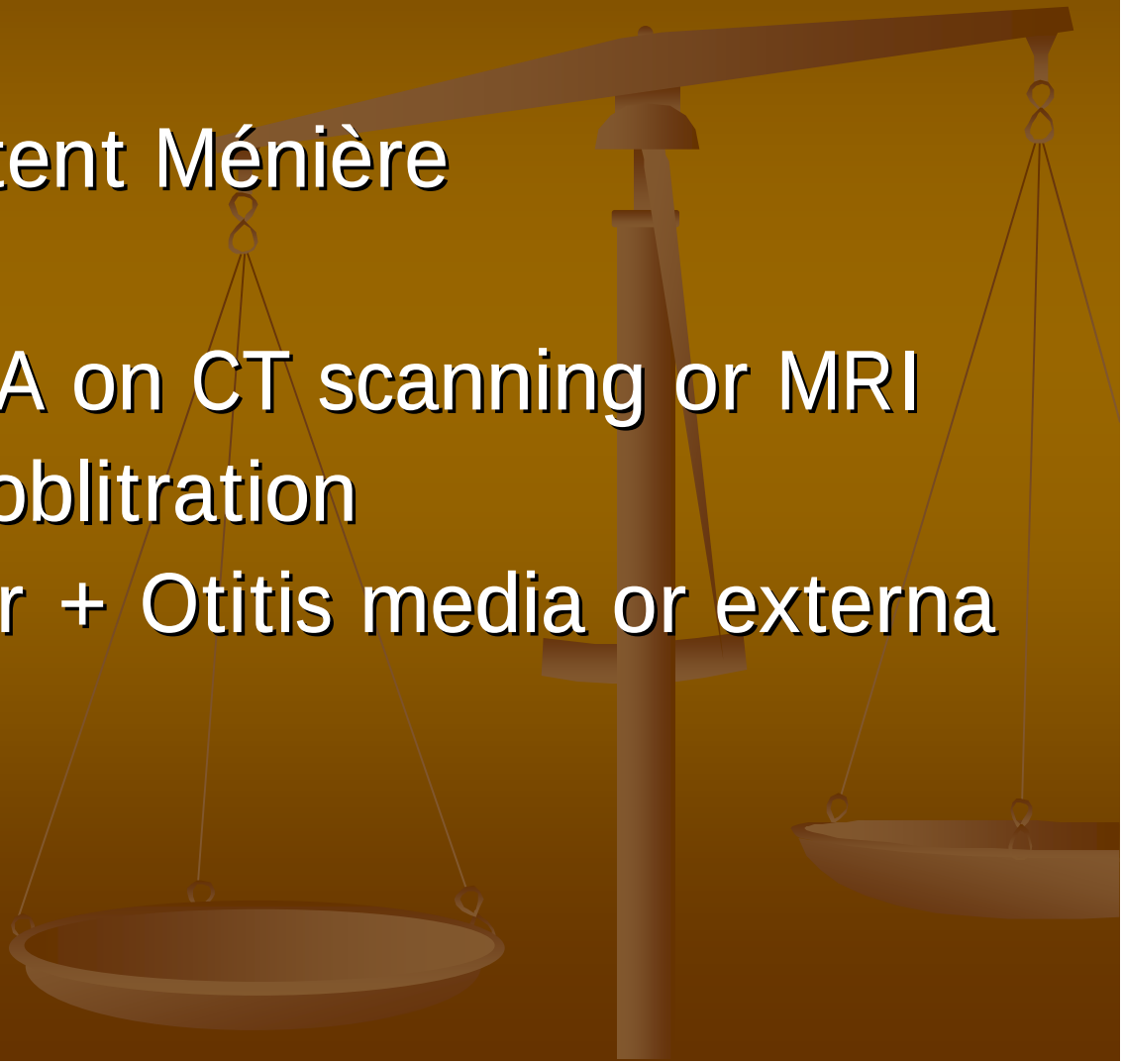
Osteogenesis imperfecta

- 30 Y
- Bilateral HL
- -ve FHx
- Fractures

Contraindications

HYDROPS

- Hydrops Coexistent Ménière
- Young
- Dilation CA or VA on CT scanning or MRI
- Round window oblitration
- Only hearing ear + Otitis media or externa
- Perforation
- Silent < 20db



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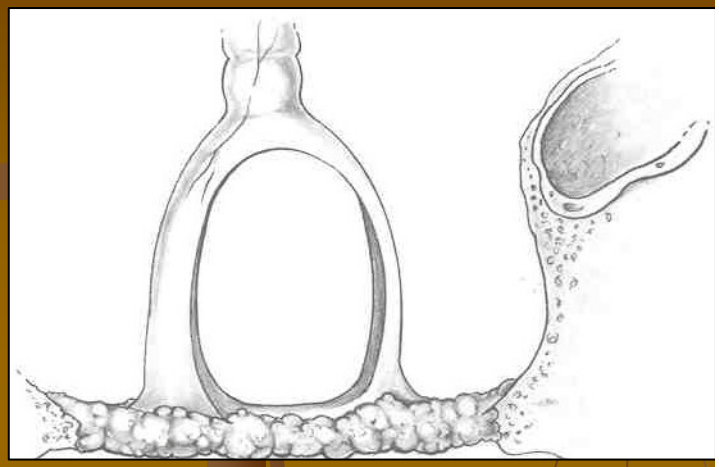


Thank
You