

Temporal Bone 2: Inner Ear and Petrous Apex

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Chair

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ACR Head and Neck



@bpoliceni



**American College
of Radiology**

Focused. Forward. Together.



Financial Disclosure

No conflict of interest relevant to
this topic

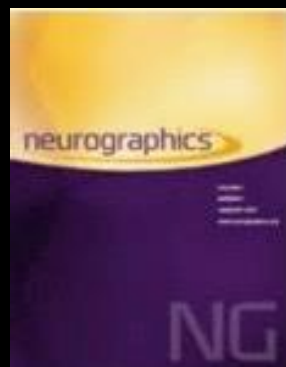
Acknowledgments

Dr Samir Noujaim – Oakland University William
Beaumont School of Medicine

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Dr Wendy Smoker - University of Iowa

Dr Achint Singh – University of Texas - SA



Imaging Algorithm of Petrous Apex Lesions

Achint K. Singh, Wendy R.K. Smoker, and Bruno A. Policeni

Learning Objectives

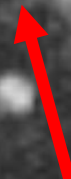
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- Inner ear anatomy
- Congenital inner ear malformations
- Acquired etiologies of SNHL
- Petrous temporal bone

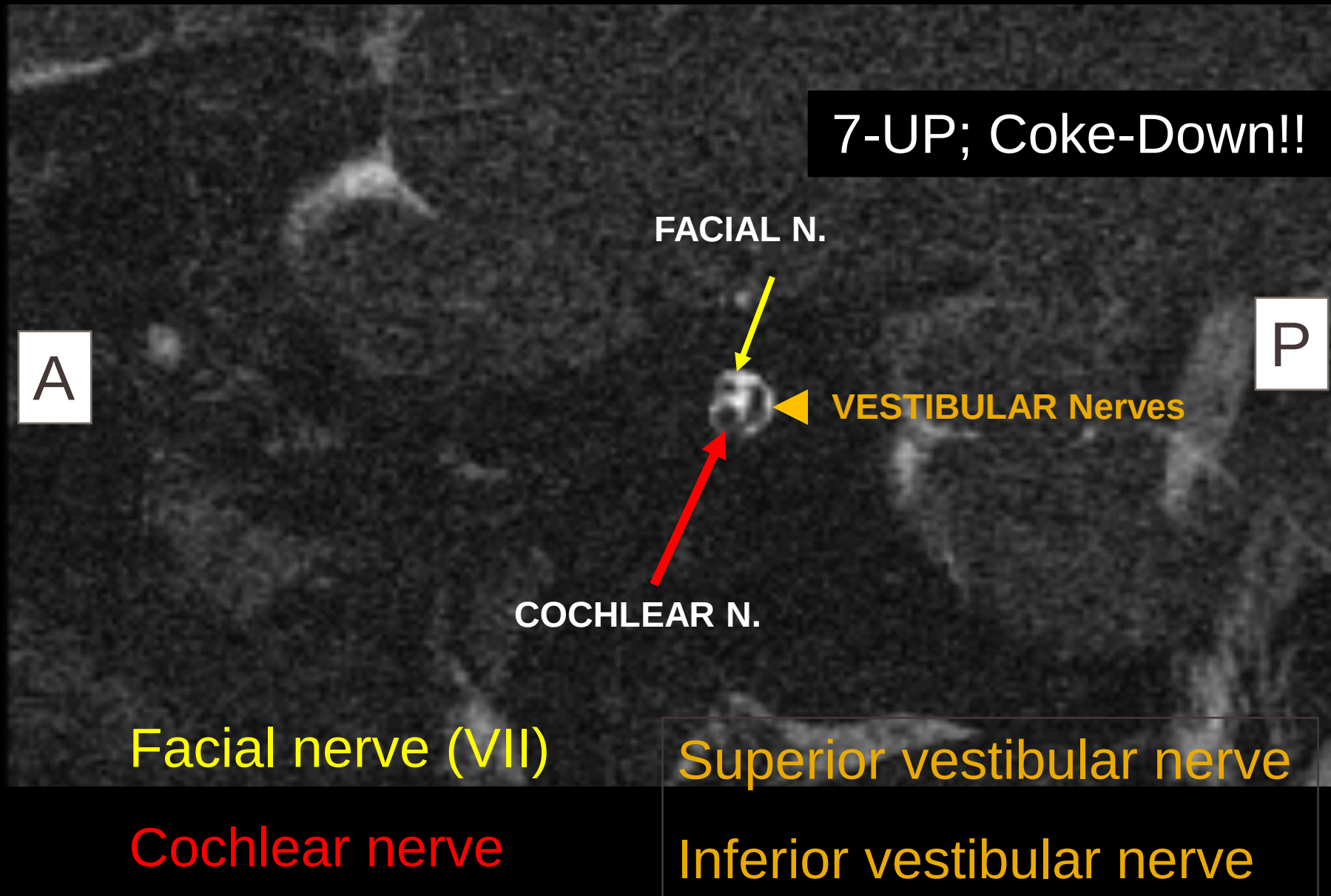
SAGITAL

FACIAL VII

VESTIBULOCOCHLEAR VIII

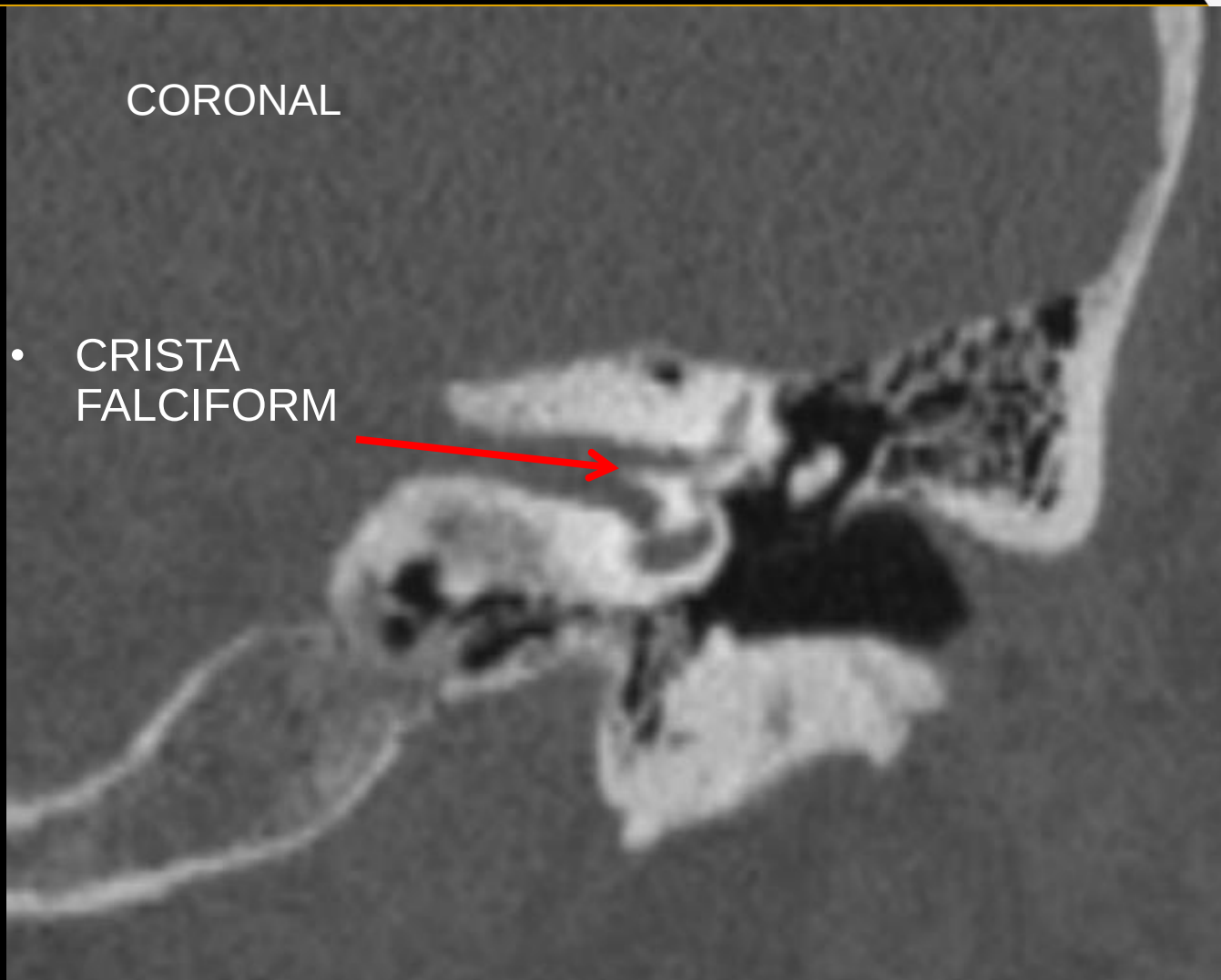


Oblique sagittal left lateral IAC



CRISTA FALCIFORM

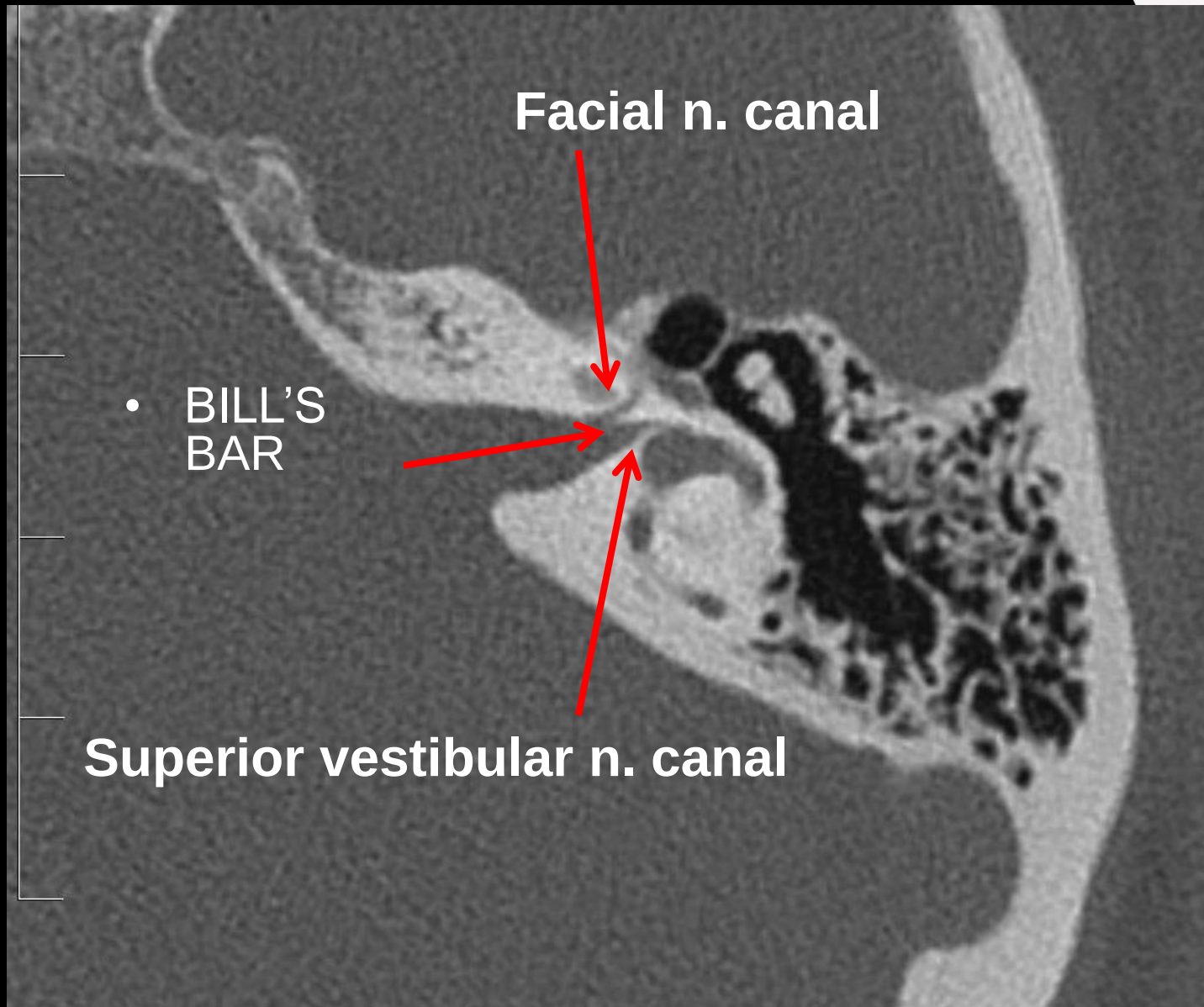
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SUPERIOR and INFERIOR COMPARTMENTS at IAC FUNDUS

Compartment Above Crista Falciform:

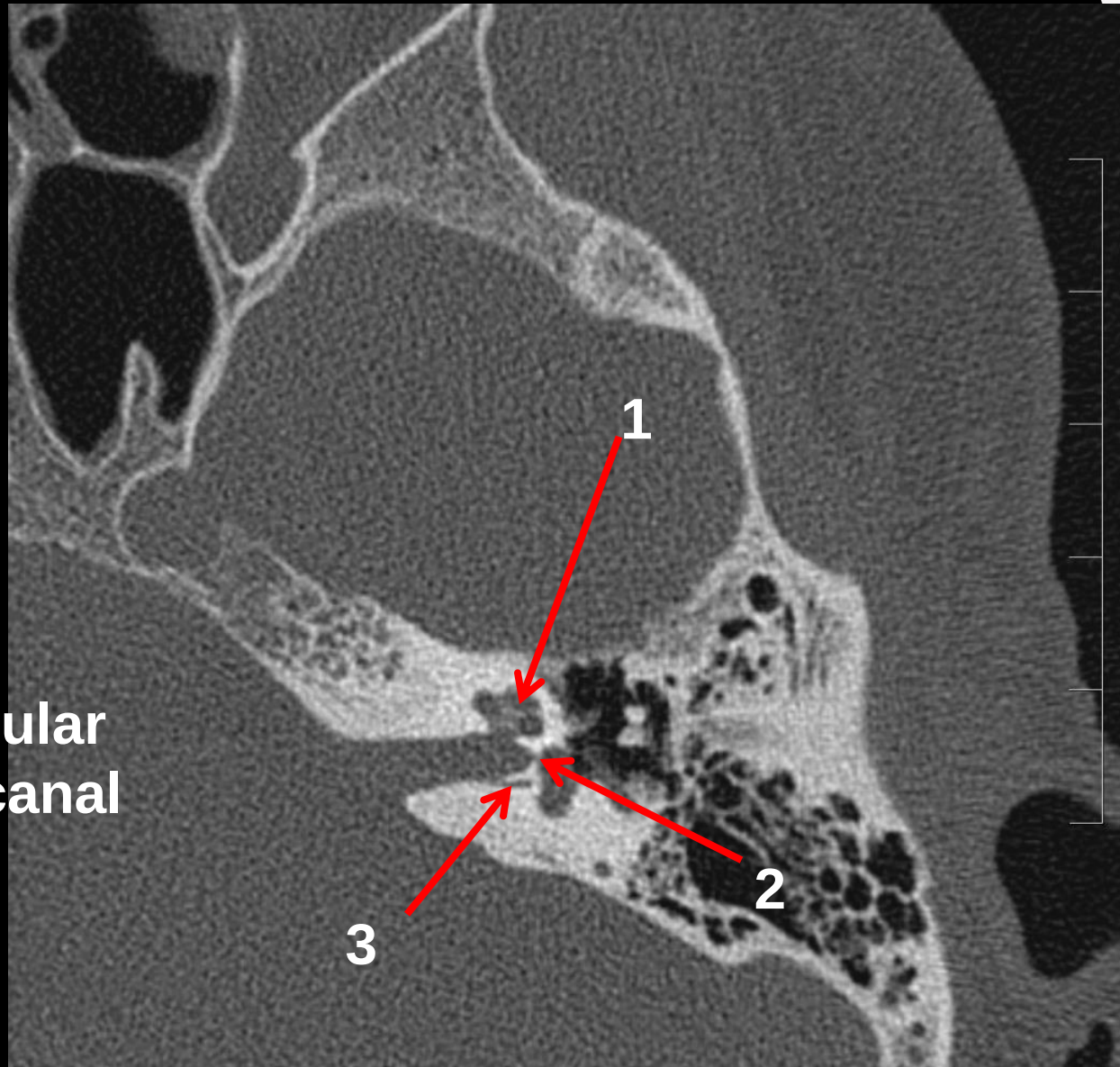
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Compartment Below Crista Falciform:

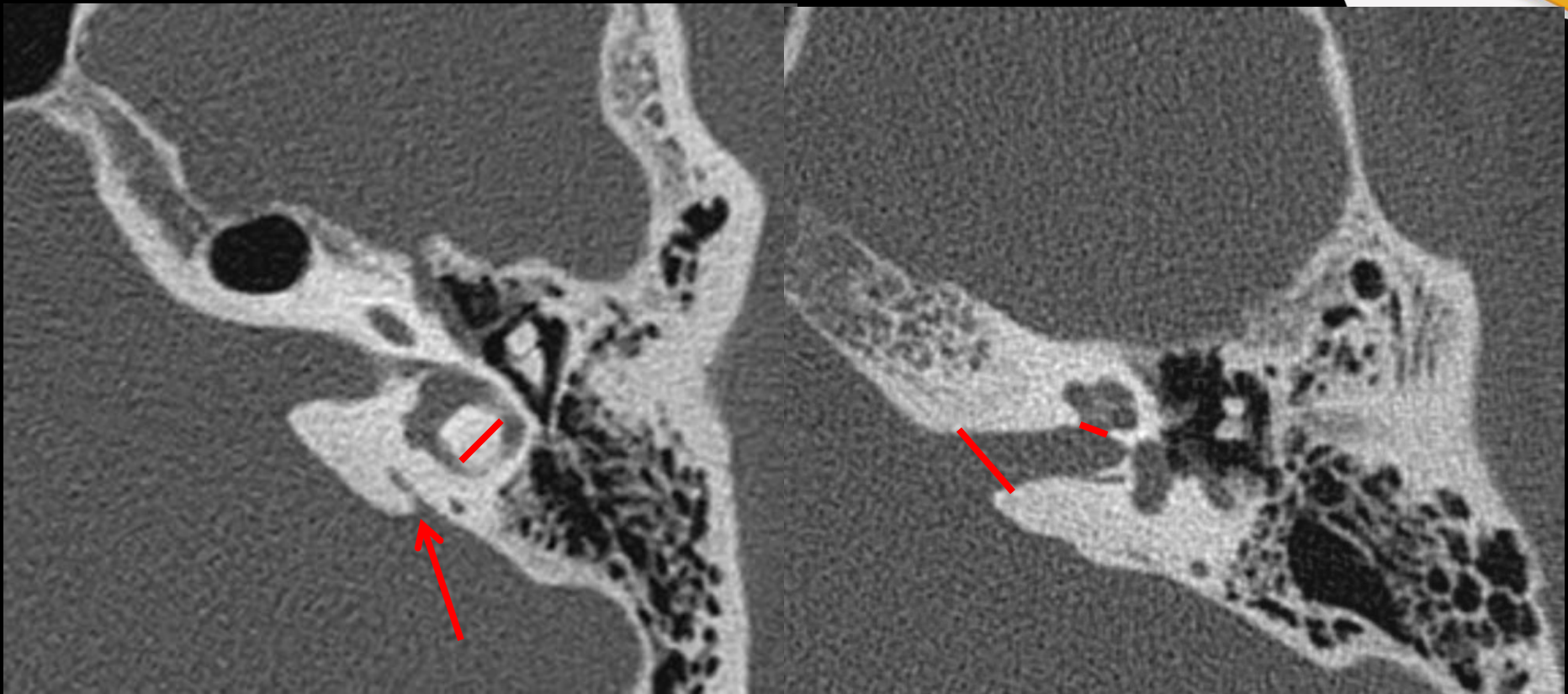
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- 1-Cochlear
- 2-Inf. Vestibular
- 3-Singular canal



FOUR MEASUREMENTS YOU WILL NEED TO KNOW !

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Transverse diameter bone island LSCC:
2.5mm-5mm

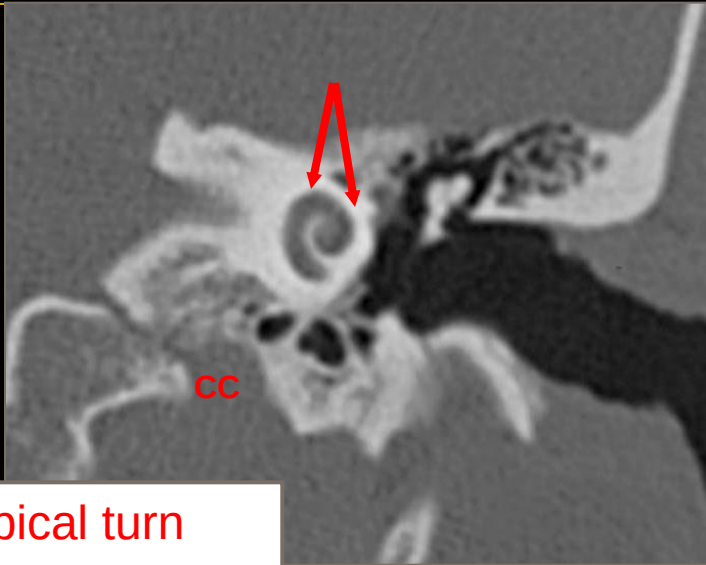
Vestibular aqueduct: 1.0mm – 1.5mm

Base cochlear n. canal: 1.5mm-3mm

IAC: 2.5mm-8mm Average: 4mm

Coronal Cochlear Plane

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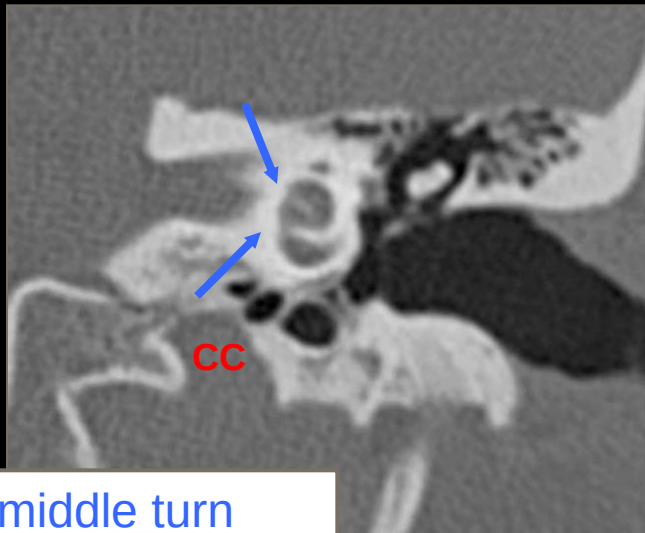


apical turn

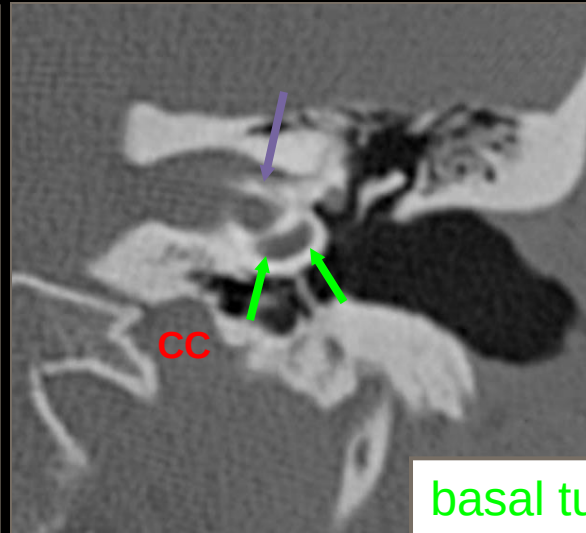
Cochlea: **apical**, **middle**,
and **basal** turns

Falciform crest

Carotid canal (CC)
“Cochlear-Carotid”



middle turn

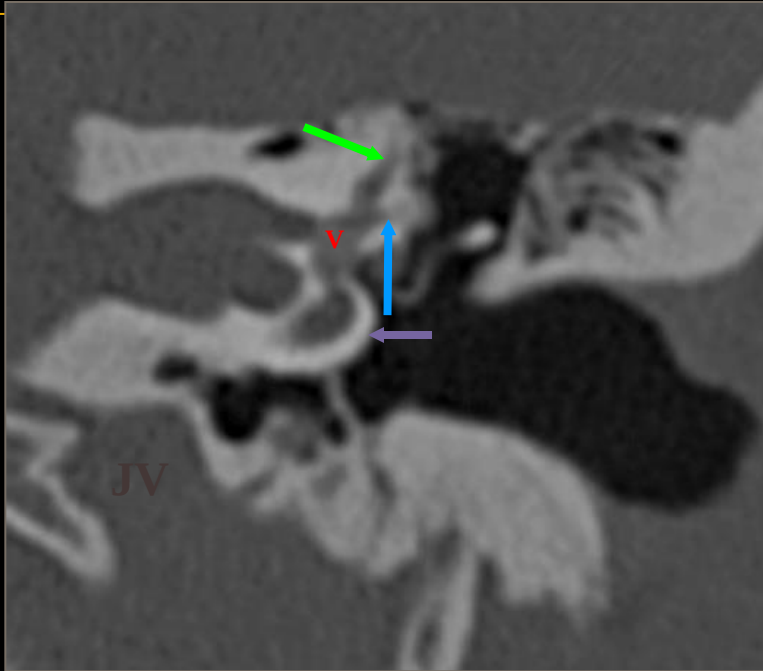


basal turn



Coronal Vestibule Plane (“Vestibule-Vein”)

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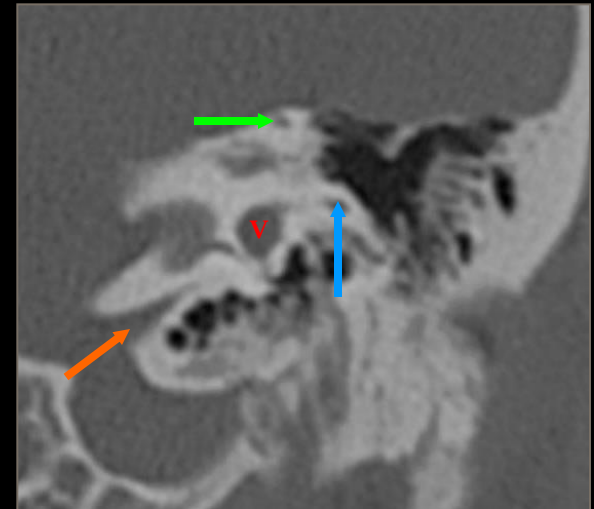
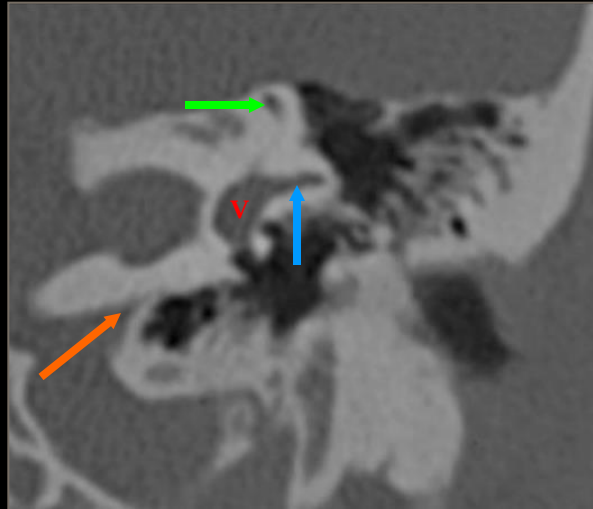
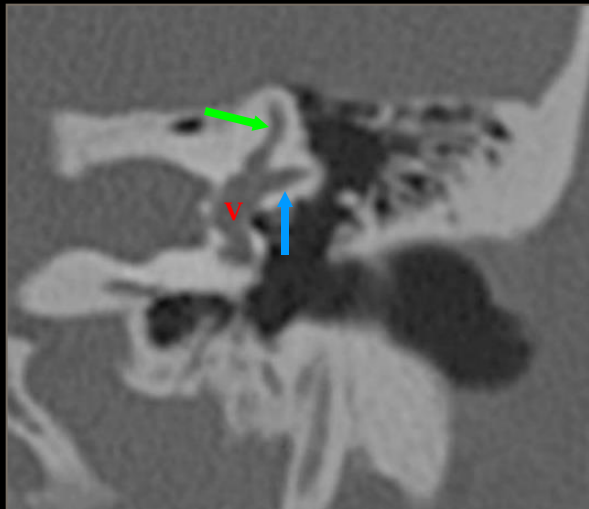


Superior semicircular canal

Lateral semicircular canal

Cochlear promontory

Cochlear aqueduct

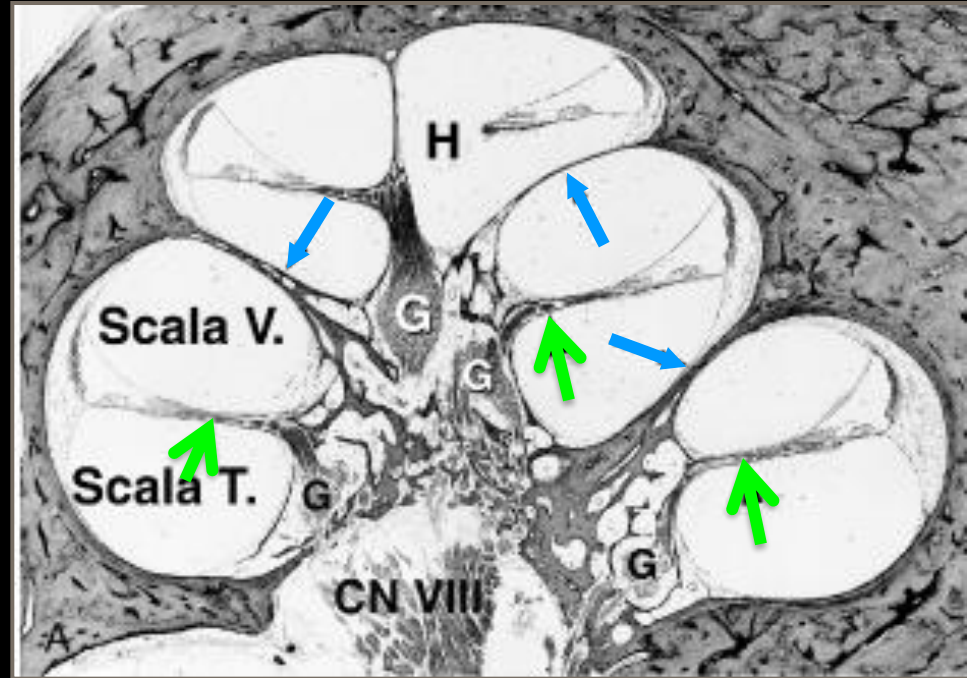


Cochlear

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3D CISS MR image

modiolus



Spiral lamina from modiolus into cochlear turns partially separate scala vestibuli from scala tympani. Interscalar septae

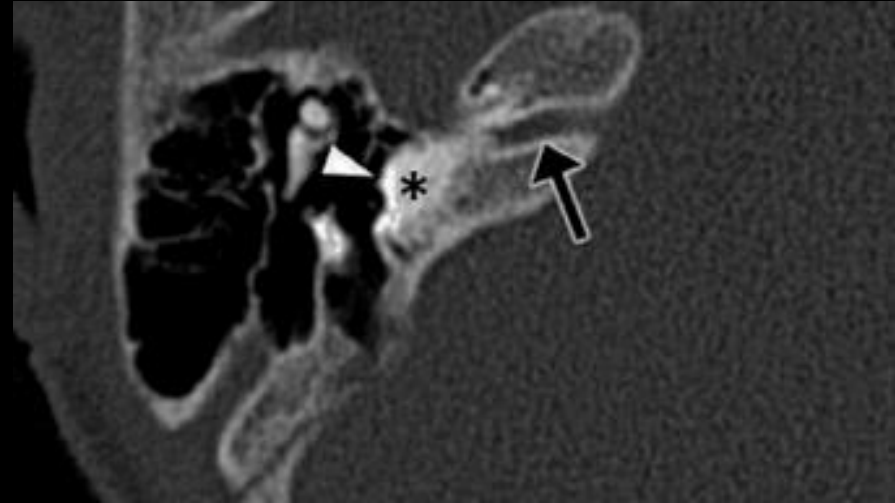
Inner Ear embryology

- The inner ear stems from the otic placode that starts developing during the 3rd week of pregnancy.
- The development of the cochlea is completed in the 8th week of pregnancy
- The vestibule in the 11th week
- The semicircular ducts between the 19th and 22nd weeks of pregnancy.
- The first semicircular duct to develop is the superior one followed by the posterior one and the last one to develop is the lateral one
- After 24 week only the vestibular aqueduct and sac continue to grow.

- Complete labyrinthine aplasia (Michel aplasia) (before the 3rd week)
- Cochlear aplasia (differential with labyrinthitis ossificans) (3rd week)
- Common cavity (4th week)
- Cochlear hypoplasia (6th week)
- Type I incomplete partition (5th week)
- Type II incomplete partition (7th week)
- Type III incomplete partition (CN VIII present) (syndromic)

Michel aplasia

- Interruption of the otic placode development before the 3rd week of pregnancy.
- Absent:
 - Cochlea
 - Vestibule
 - Semicircular canals (SCCs)
 - Vestibular and cochlear aqueducts
- Development of middle ear ossicles are usually normal.
- It amounts to 1% of IE malformations only
- Cochlear implant is not a good treatment option (brainstem implant)
- IAC aplasia or narrowing.
- Aplasia of the cochleovestibular nerve and the facial nerve trajectory is aberrant.
- Associated anomalies:
 - Petrous apex hypoplasia
 - Absence of round and oval windows
 - Flattening of the middle ear medial wall (absent promotorium)

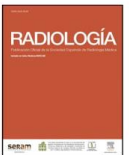


Radiología. 2017;59(4):297–305

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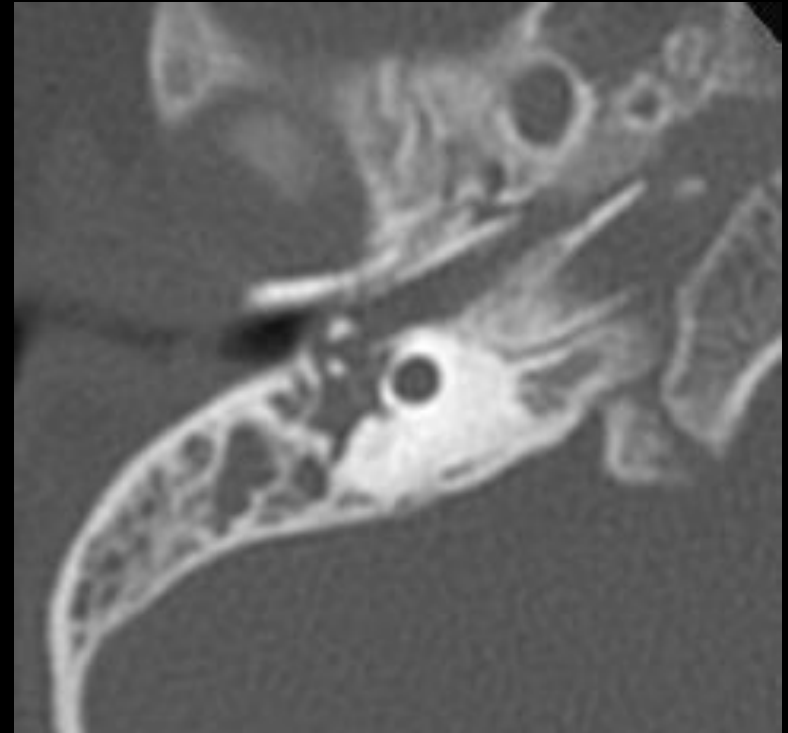
RADIOLOGY THROUGH IMAGES

Inner ear malformations: A practical diagnostic approach☆



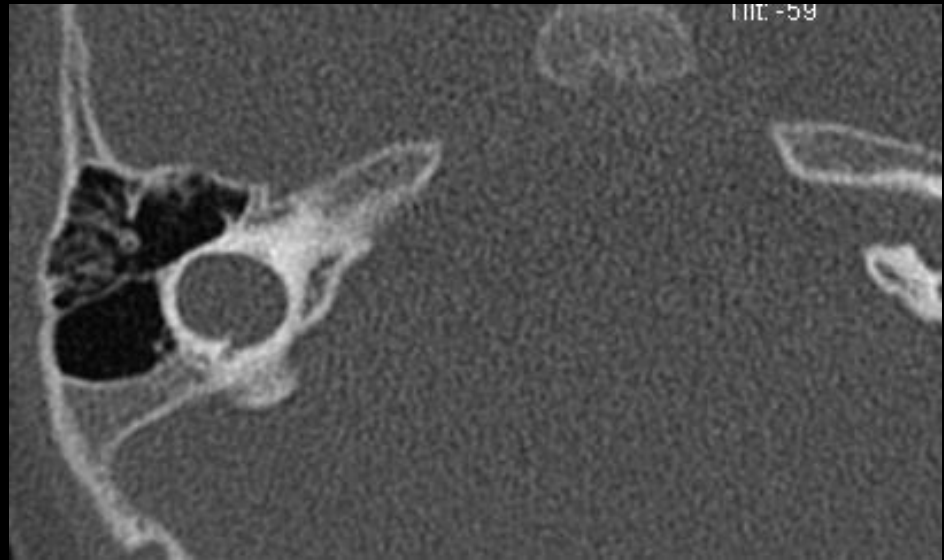
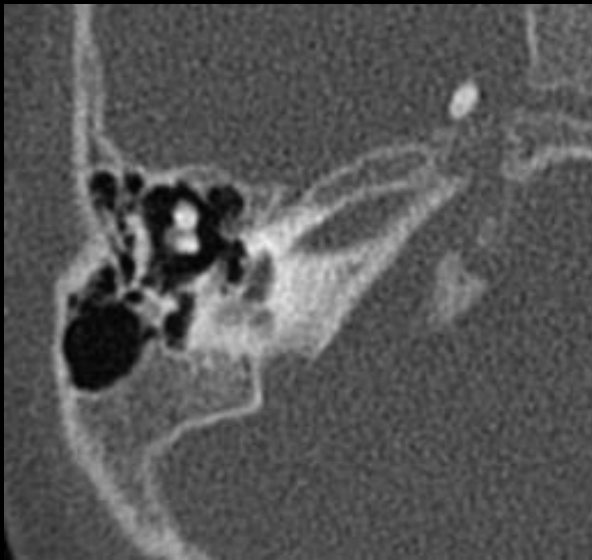
M. Mazón^{a,*}, E. Pont^b, A. Montoya-Filardi^a, J. Carreres-Polo^a, F. Más-Estellés^a

- Ovoid or round cavity within the otic capsule
- IAC absent or rudimentary
- The embryological insult is around the third week



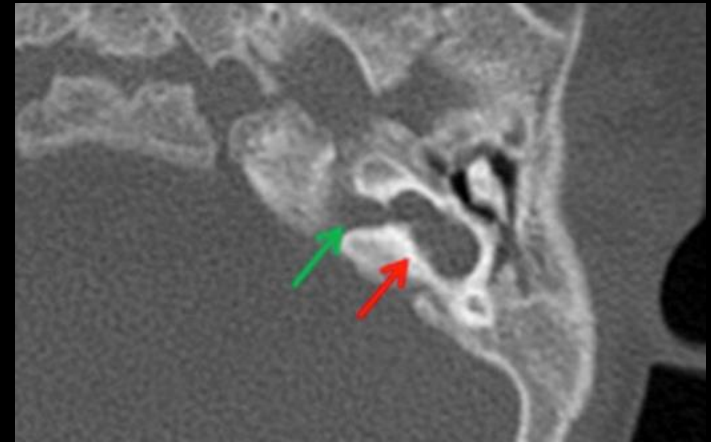
Courtesy: Dr Juliano and
Dr Robson

- It is due to developmental interruption at the end of the 3rd week of pregnancy and it amounts to 3% of all IE malformations.
- **Cochlear aplasia with normal labyrinth**
- **Cochlear aplasia with a dilated vestibule (CADV)**



Courtesy: Dr Juliano

- Result of developmental interruption during the 4th week of pregnancy and amounts to 25% of all malformations.
- Cochlear implant is possible though hard to accomplish and the rate of complications is higher
- Single, ovoid or round chamber, representing cochlea and vestibule
- There may be accompanying SCC or their rudimentary parts
- The IAC usually enters the cavity at its center.



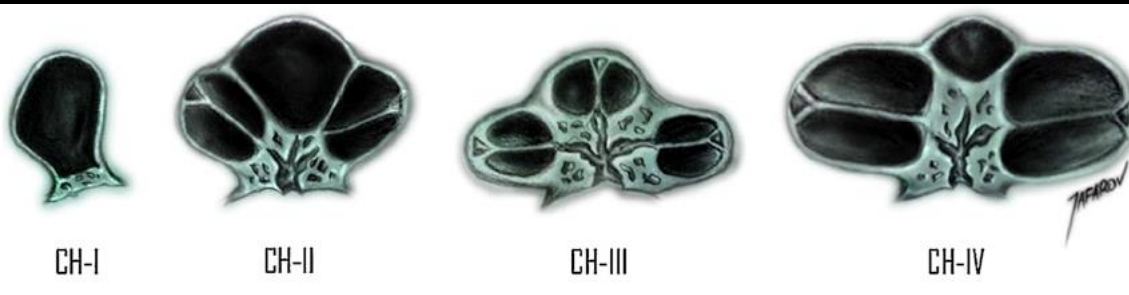
Radiological diagnosis of the inner ear malformations in children with sensorineural hearing loss

¹BERNADINE QUIRK, MBBS, ¹ADAM YOUSSEF, MBBS, ²MARIO GANAU, MD and ¹FELICE D'ARCO, MD

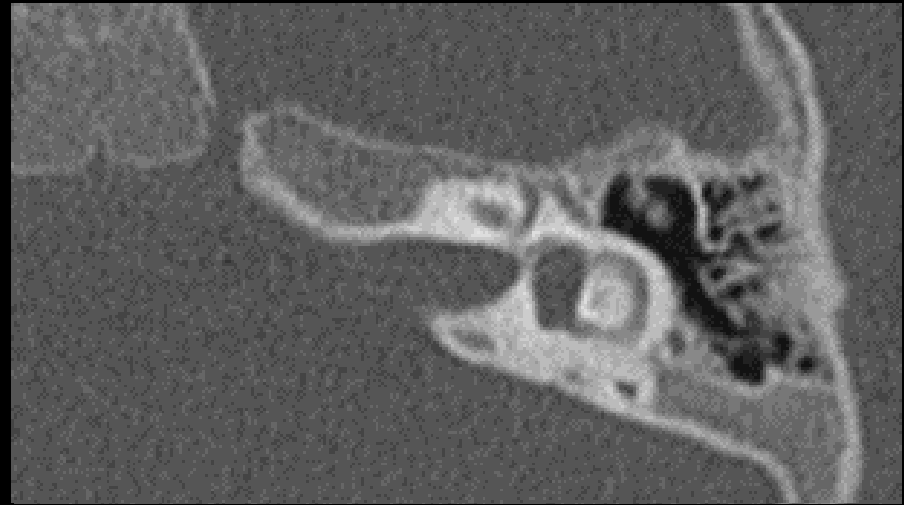
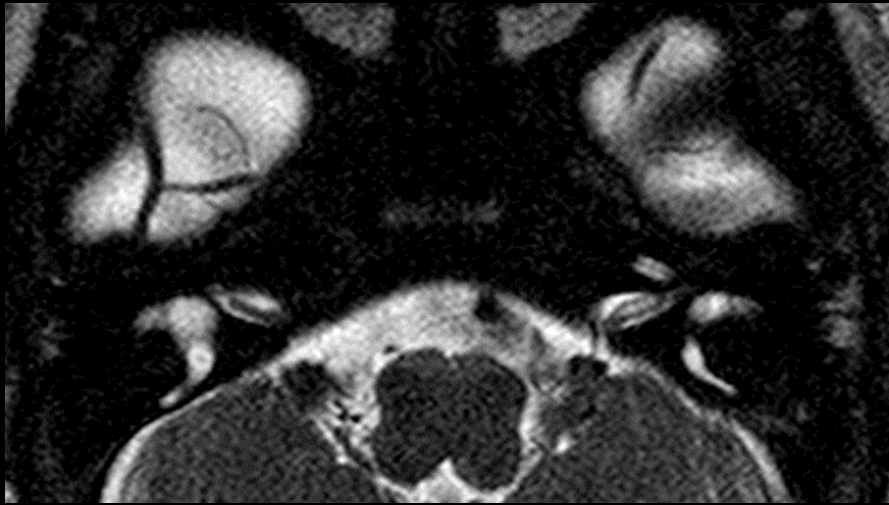
¹Great Ormond Street Hospital for Children NHS Foundation Trust. ²Department of Neurosurgery, Oxford University Hospitals NHS Foundation Trust.

Cochlear hypoplasia

- Cochlear hypoplasia type I (CH-I) is defined as a bud like cochlea (round or ovoid) arising from the internal acoustic canal.
- Cochlear hypoplasia type II (CH-II) is defined as cochlea with smaller dimensions and without modiolus and ISS. But external architecture is similar to that of a normal cochlea and makes a wide connection with the existing IAC.
- Cochlear hypoplasia type III (CH-III) has a shorter modiolus than normal and the cochlea has a reduced number of turns (<2 turns)
- Cochlear hypoplasia type IV (CH-IV) has a cochlea with approximately a normal sized basal turn and severely hypoplastic middle and apical turns



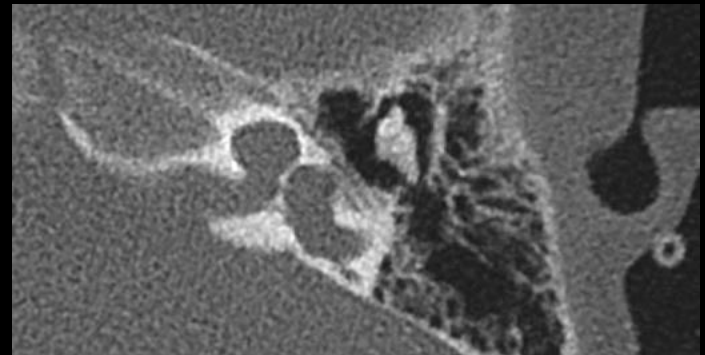
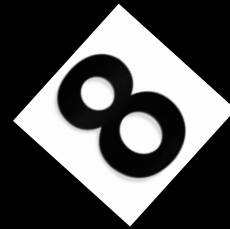
Cochlear hypoplasia



Courtesy: Dr Juliano

Incomplete partition type I

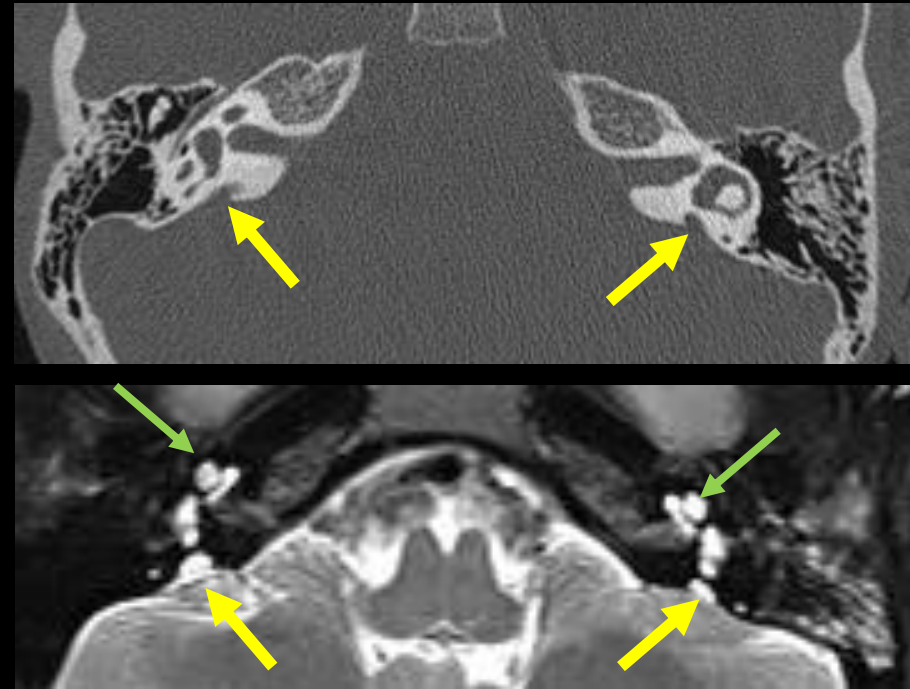
- Result of developmental interruption during the 5th week of pregnancy
- AKA: cystic cochleovestibular malformation
- The cochlea shows a total absence of the modiolus and cystic appearance without internal architecture; the vestibule is dilated
- Appearance of the figure eight



Incomplete partition type II mondini

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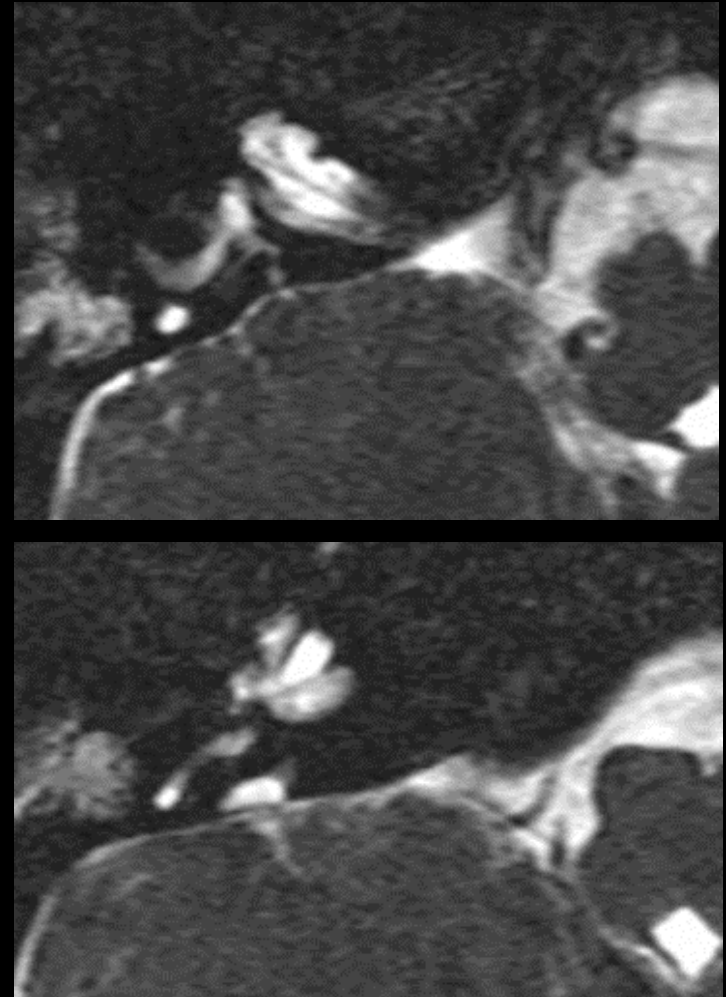
- Developmental interruption during the 7th week of pregnancy.
- Most common Inner Ear dysplasia (present in 50% of all malformations)
- Fusion of the middle and apical turns
- Cystic appearance due to a defect in the apical segment of the modiolus, the interscalar septum and the osseus spiral lamina
- The modiolus basal coil and the basal segment look normal
- Usually associated with Mondini deformity triad
 - Enlarged vestibular aqueduct (due to duct and endolymphatic sac dilation)
 - Enlarged vestibule
 - Cochlear deformity



Courtesy: Dr Noujaim

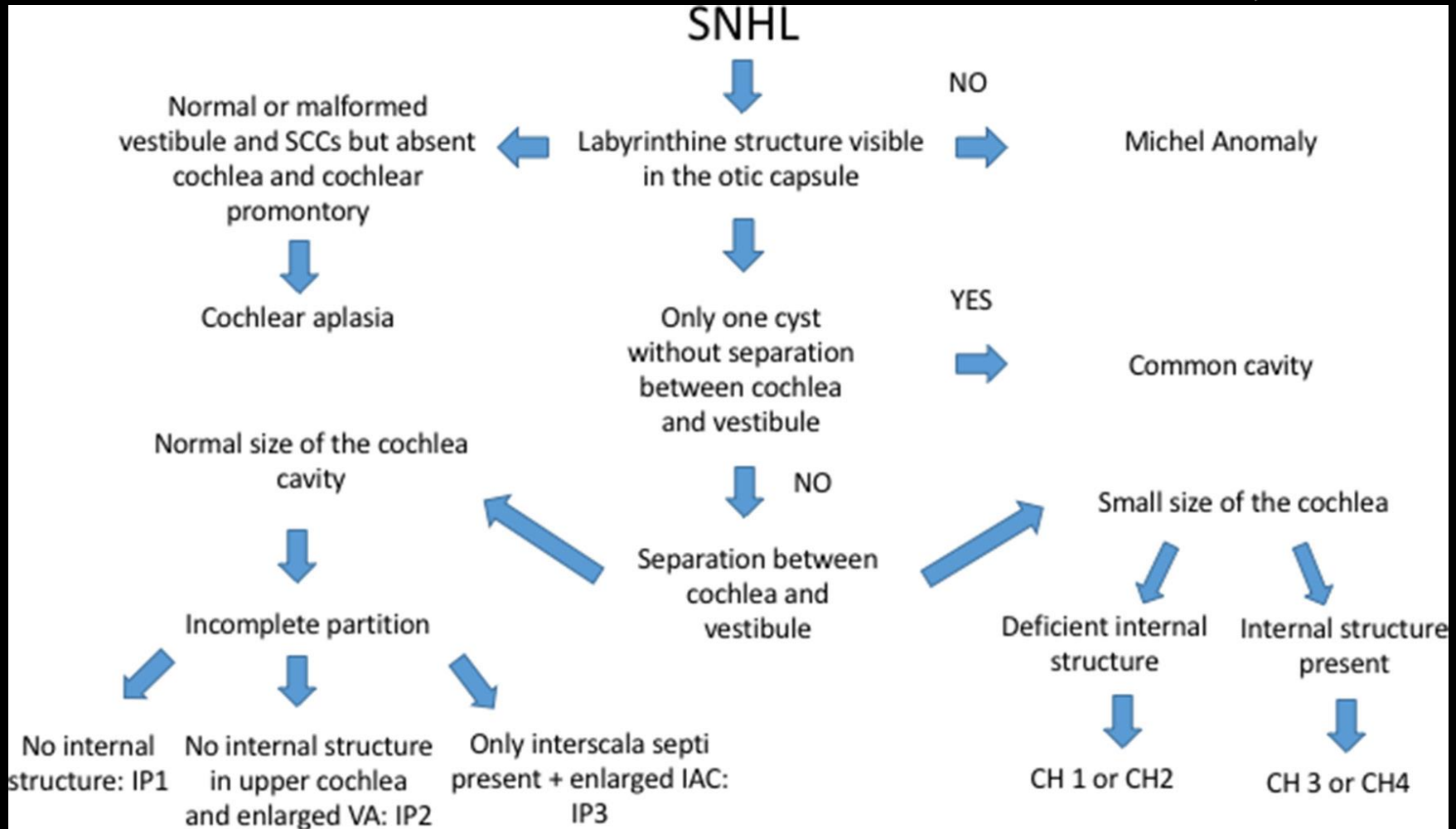
Incomplete partition type III

- Recessive genetically inherited disease linked to chromosome X and it is not due to the developmental interruption of the otic placode
- POU3F
- It usually affects males and mixed hearing loss of rapid progression
- Total absence of the modiolus though the inter- scalar septum is present
- The cochlea is located laterally to the IAC that is in turn widened and both widely communicated due to a complete deficit of the cribose layer
- “X-linked stapes gusher”



Courtesy: Dr Noujaim

Classification



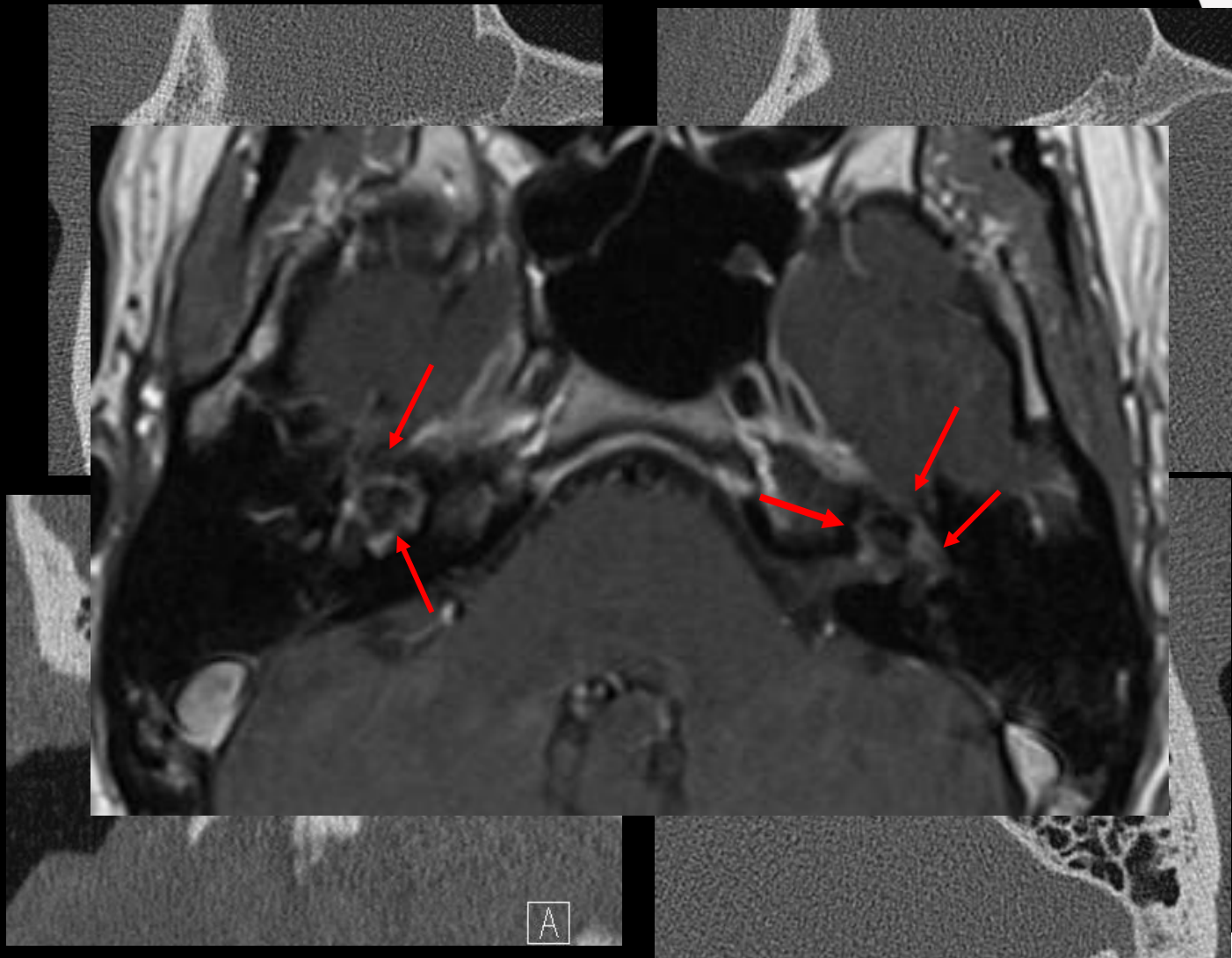
Quirk B, Youssef A, Ganau M, D'Arco F. Radiological diagnosis of the inner ear malformations in children with sensorineural hearing loss. BJR Open 2019; 1: 20180050.

Otosclerosis

- Otosclerosis is a primary osteodystrophy of the otic capsule
- Commonly presents with hearing loss, most commonly conductive, but it can also be sensorineural.
- Conductive, sensorineural, or mixed hearing loss.
- Subtypes:
 - Fenestral:
 - Involves the oval window.
 - Most common
 - Conductive hearing loss
 - Retrofenestral:
 - Involves the cochlear
 - SNHL

31 y.o. male. Bilateral progressive sensorineural hearing loss

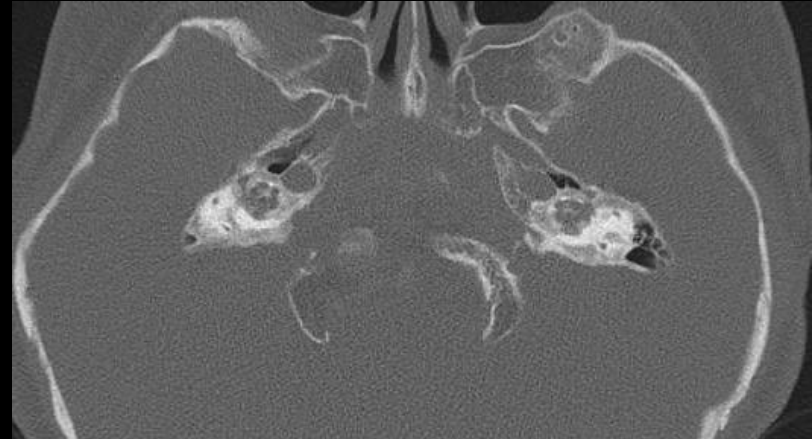
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loujaim

Osteogenesis imperfecta

- Rare inherited disorder with a prevalence of 1/10,000–30,000
- Inborn error in collagen type 1 formation
- OI is histopathologically characterized in general by undermineralized thickened bone.
- Hearing loss is an important clinical feature in patients with OI. The prevalence in several surveys has been reported between 48% to 72%
- Reduced density in the bony regions around the semicircular canals, the distal internal auditory canal and oval window.
- Differential diagnosis: Otosclerosis.(The extension of the involvement of the bony labyrinth is wider in OI than in otosclerosis)



Paget's disease

- Pathophysiology is not completely understood, but genetic factors play an important role and 15–40% of patients have at least one affected first-degree relative.
- Indirect evidence based on mortality rates suggests a trend towards a reduction in disease severity.



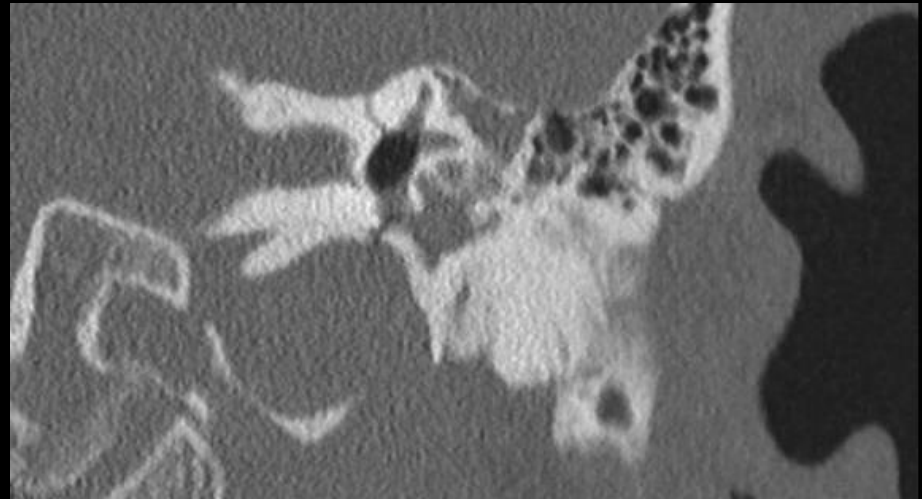
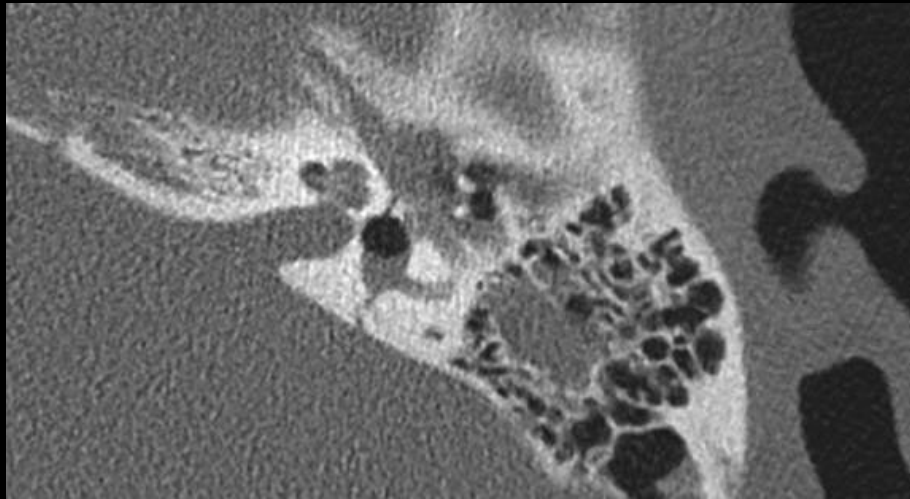
Acquired SNHL

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- Traumatic
- Neoplastic
- Infectious

Traumatic (skull-base fracture)

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Superficial siderosis

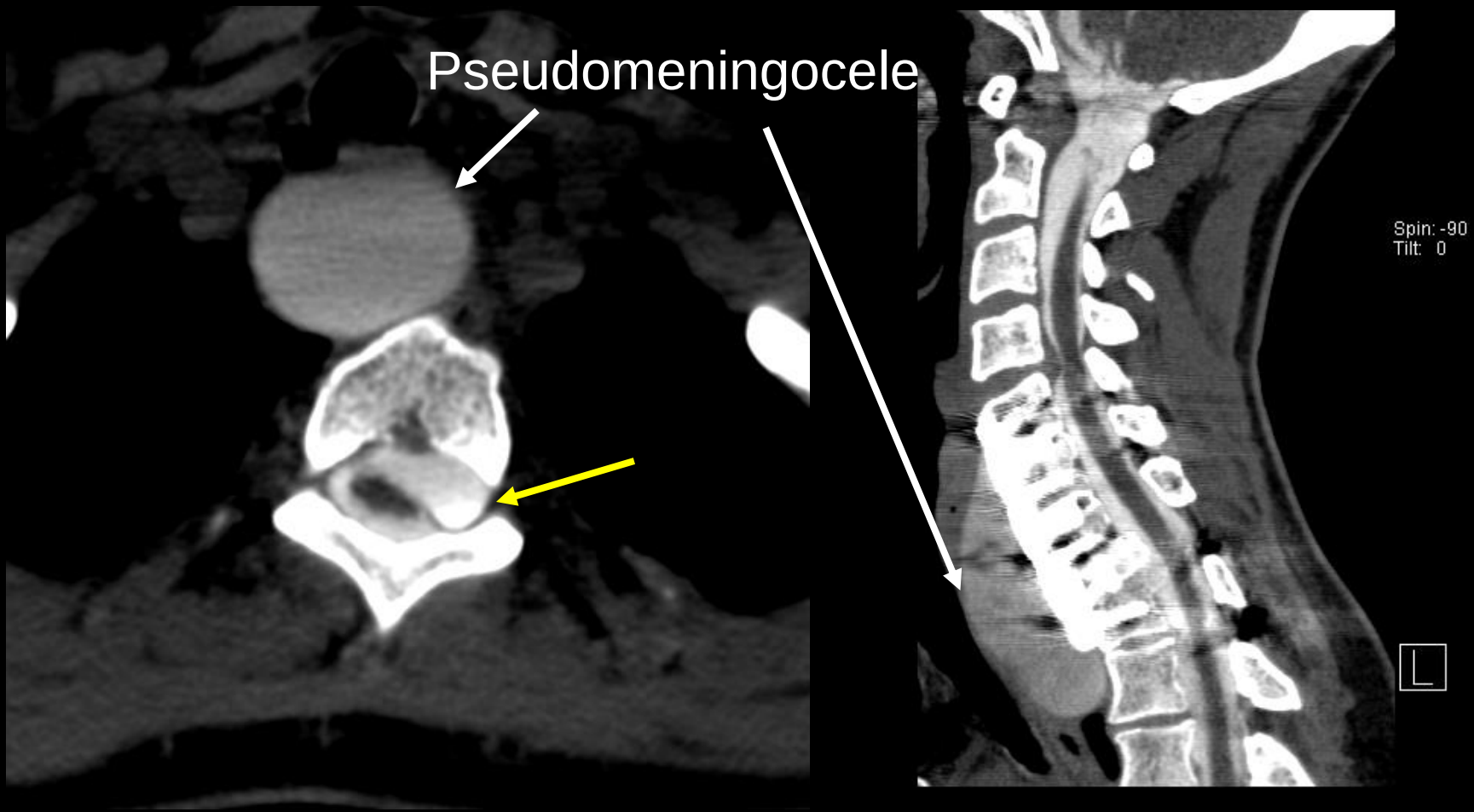
- The classic clinical presentation of SS includes adult-onset slowly progressive gait ataxia with cerebellar dysarthria and sensorineural hearing impairment.
- Imaging the entire neuroaxis
- A fluid-filled collection of variable dimensions can be seen on spine MR images or CT myelograms in patients with or without a prior history of injury or surgery.

41yo female left sided hearing loss



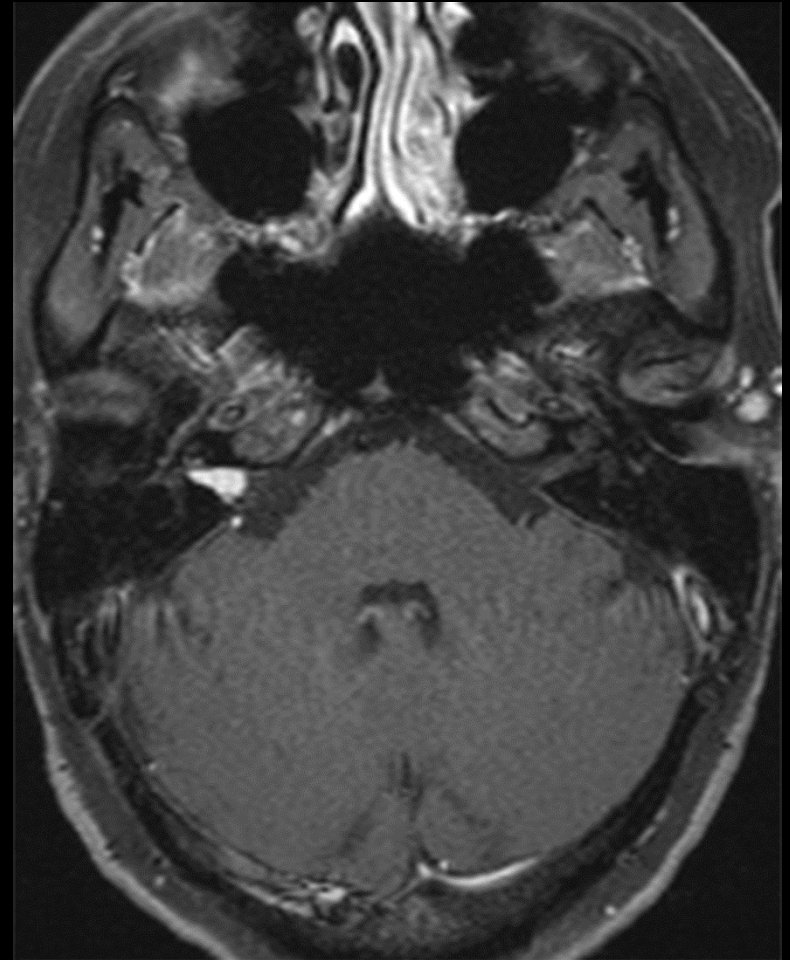
Pseudo meningocele Myelogram

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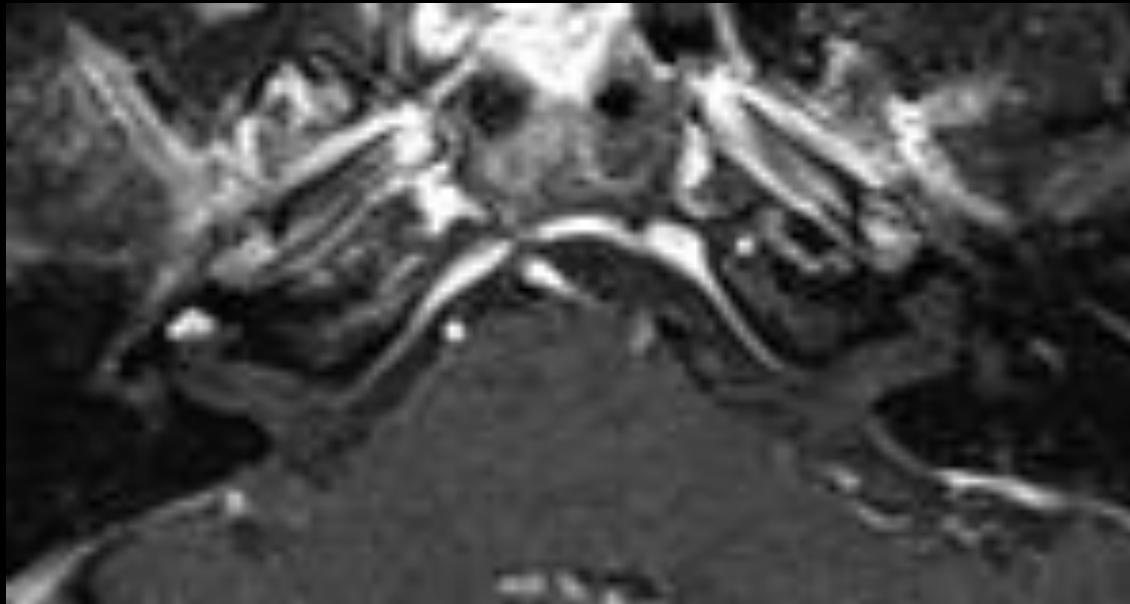
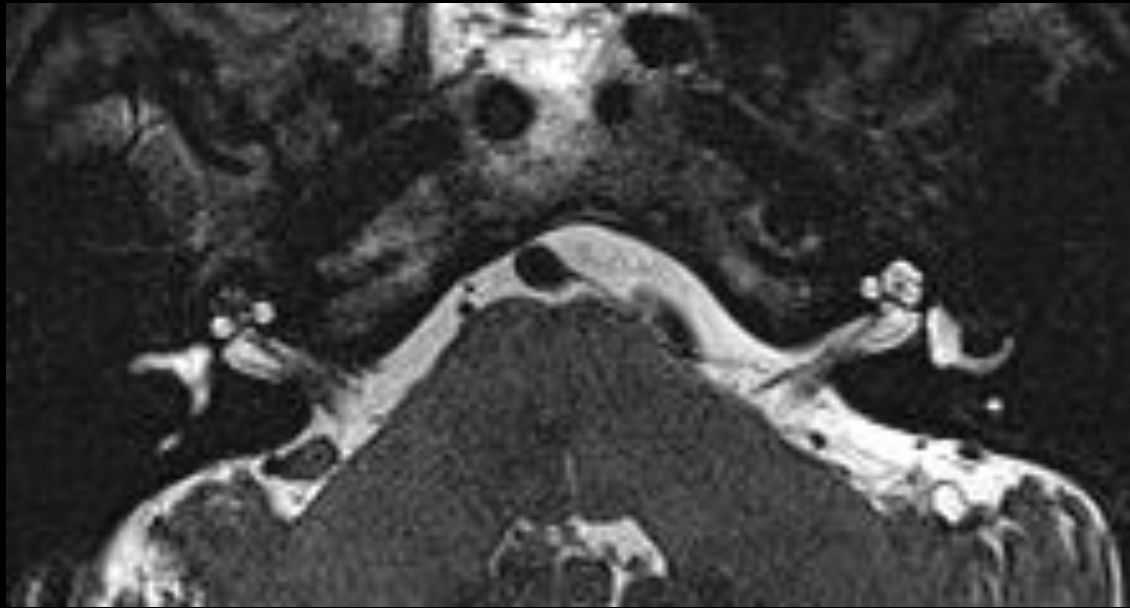
Neoplastic

- Vestibular schwannoma: most common cause of neural hearing loss, with an incidence of 1.74 per 100000 persons per year.
- Patient with unilateral loss of bone conduction on pure-tone audiometry, without any immediately recognizable cause, should undergo brainstem potential recording (BERA) and an MRI scan.
- Treatment: surgery, radiosurgery, observation



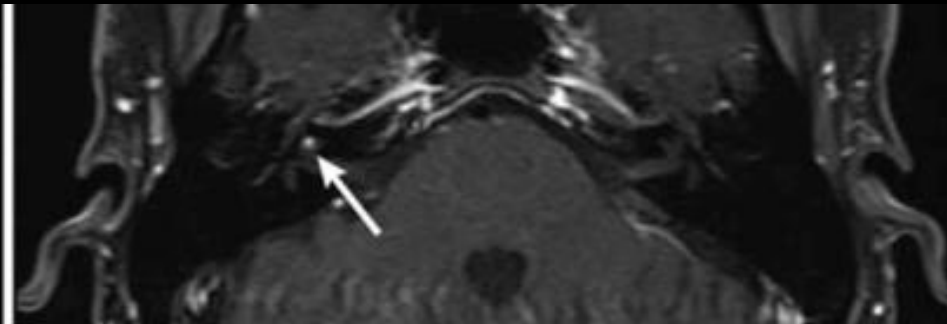
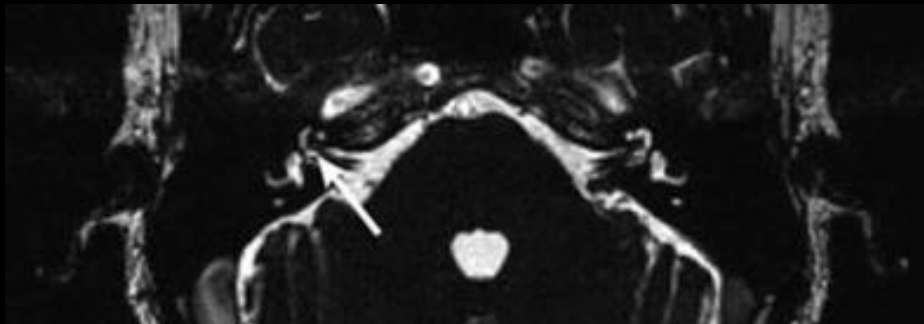
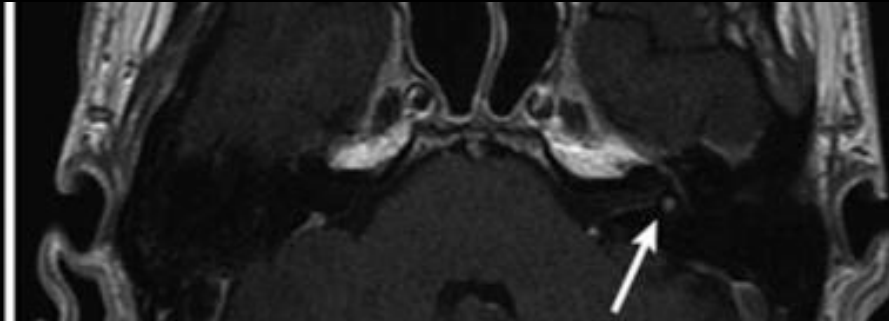
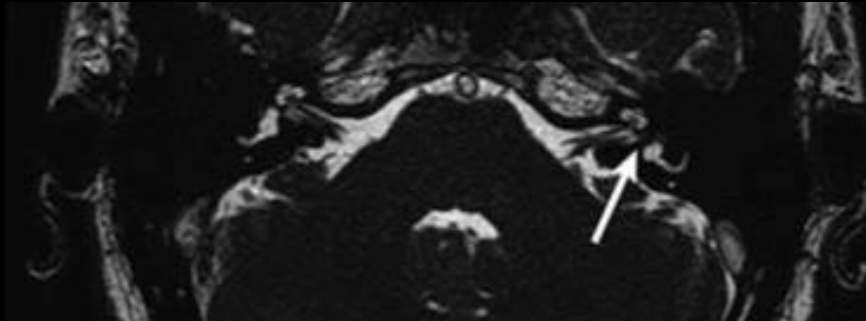
Intracochlear

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CISS accuracy

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Diagnosis of Small Vestibular Schwannomas Using Constructive Interference Steady State Sequence

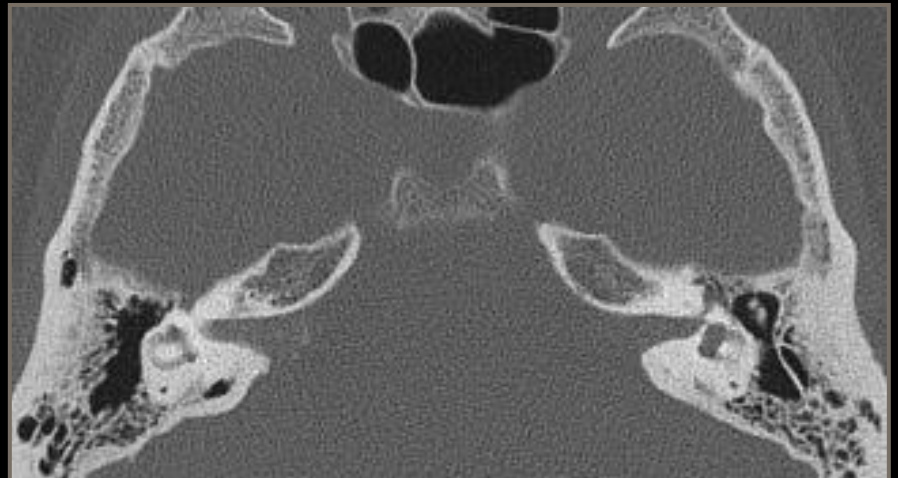
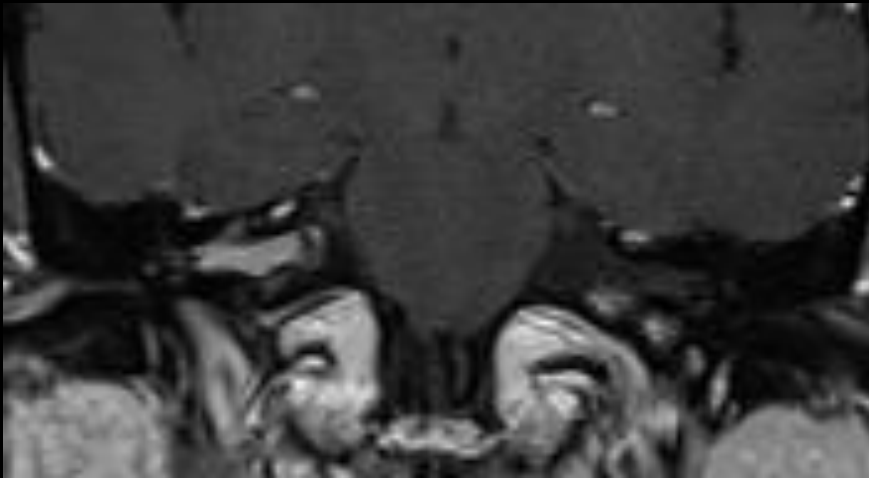
Adam A. Liudahl, MD; Andrew B. Davis, MD ; Danielle S. Liudahl, MD; Joan Maley, MD;
Bruno Policeni, MD; Marlan R. Hansen, MD 

Constructive Interference Steady State Sensitivity and Specificity
Based on Tumor Location.

Location	No. of Cases	Sensitivity, %	Specificity, %
Intracanalicular without fundal or CPA extension	48 (35%)	92.5	100
Intracanalicular with fundal extension	68 (49%)	96.3	100
Intracanalicular with CPA extension	15 (11%)	100	100
Intralabyrinthine	8 (6%)	62.3	100

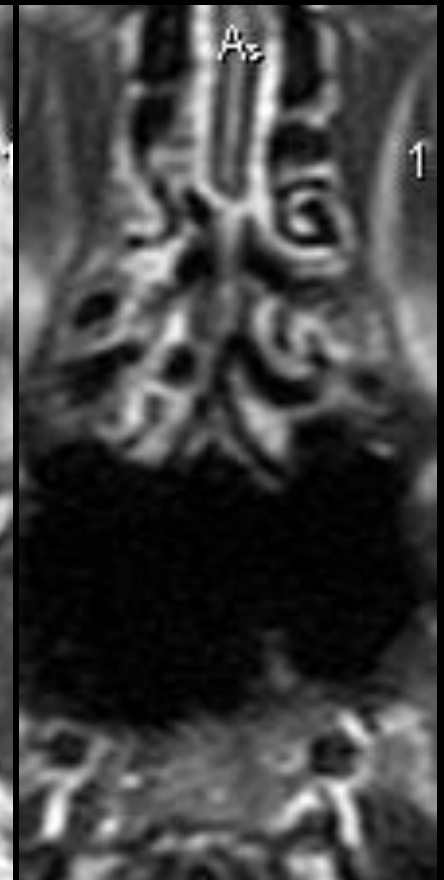
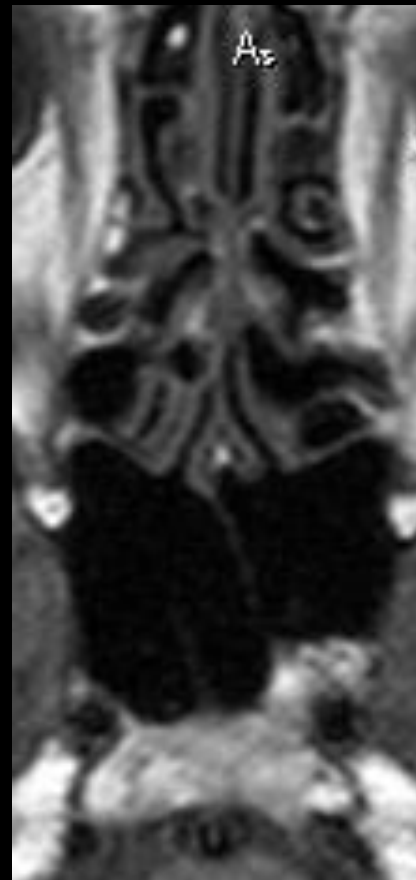
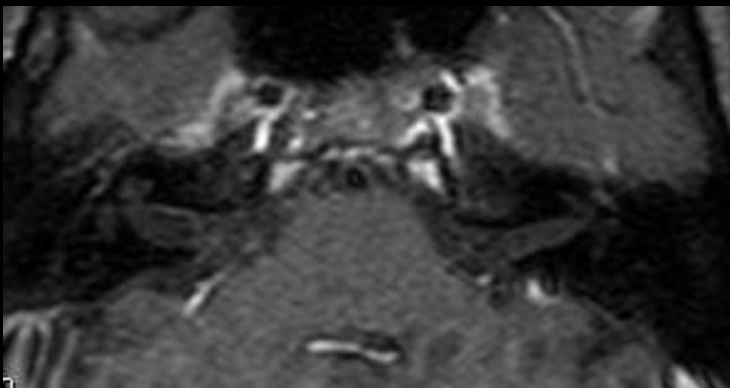
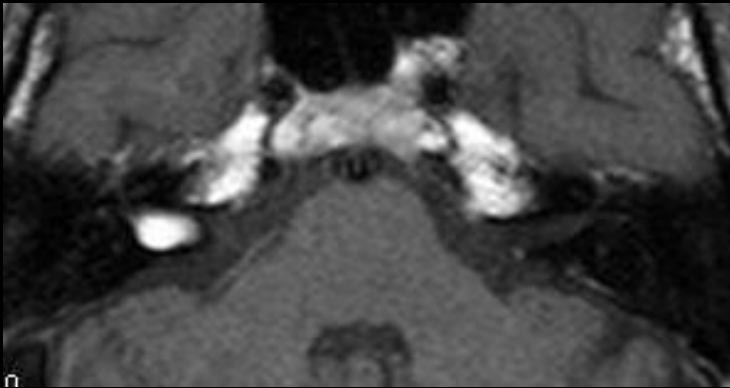
IAC Meningioma

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IAC Lipoma

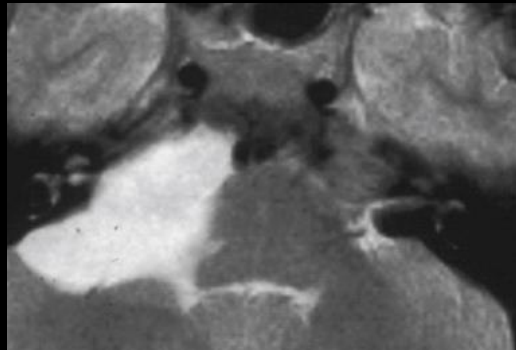
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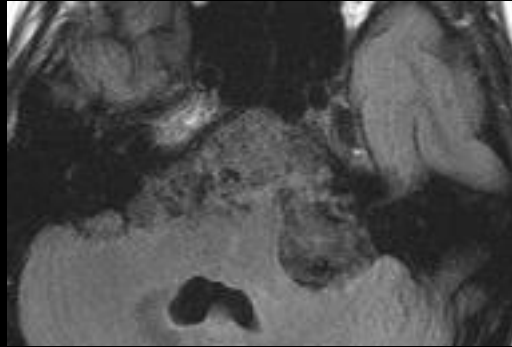
Cisternal lesions

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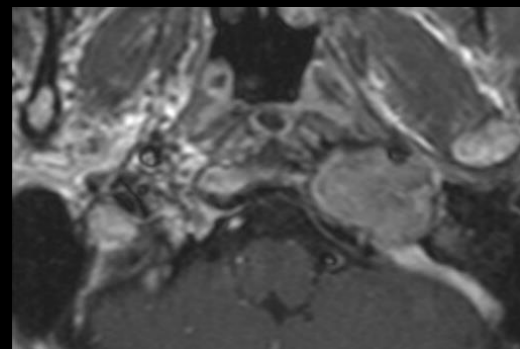
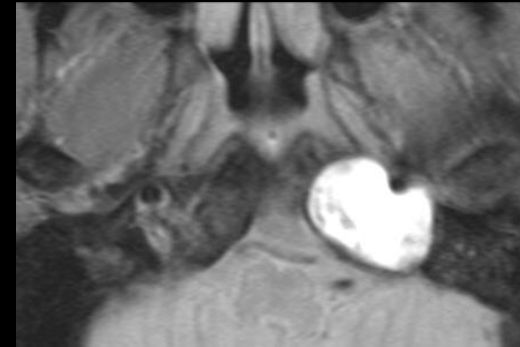
Arachnoid Cyst



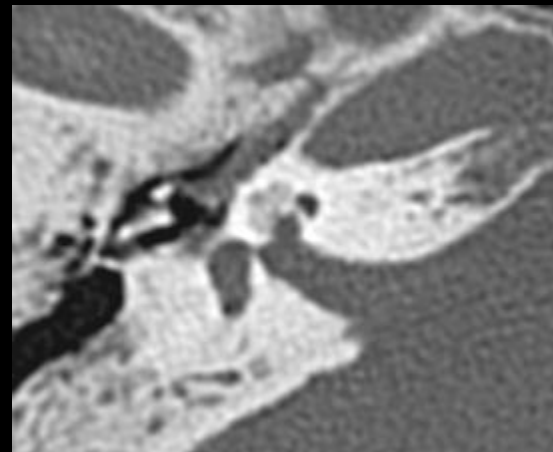
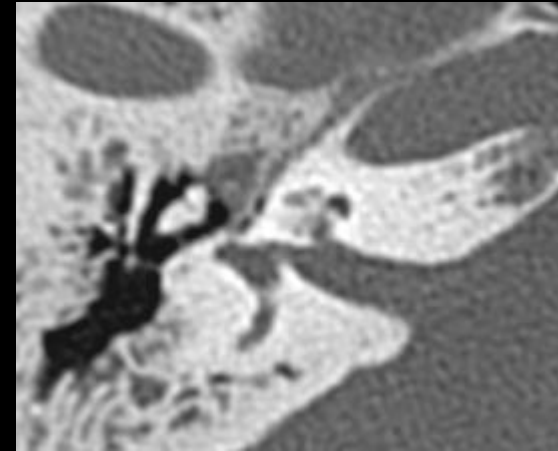
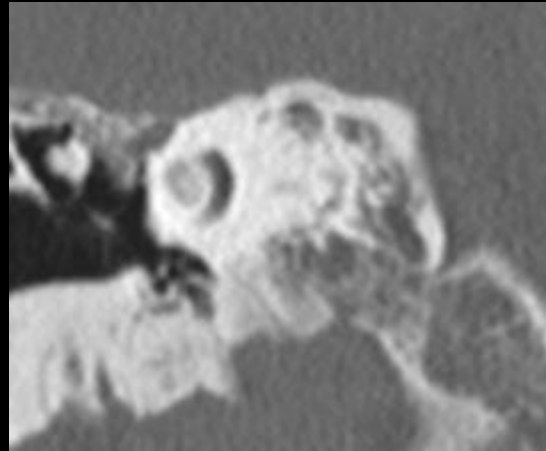
Epidermoid Cyst



Cholesterol Granuloma

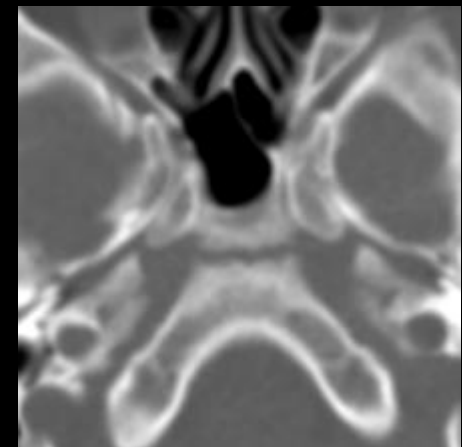


- SNHL is a known complication of otologic or central nervous system infections such as acute labyrinthitis, meningitis, or as a sequela of chronic suppurative otitis media.
- Pediatric SNHL has been linked to multiple congenital infections such as toxoplasmosis and syphilis.
- Labyrinthitis ossificans and permanent SNHL is a known sequela of bacterial meningitis.

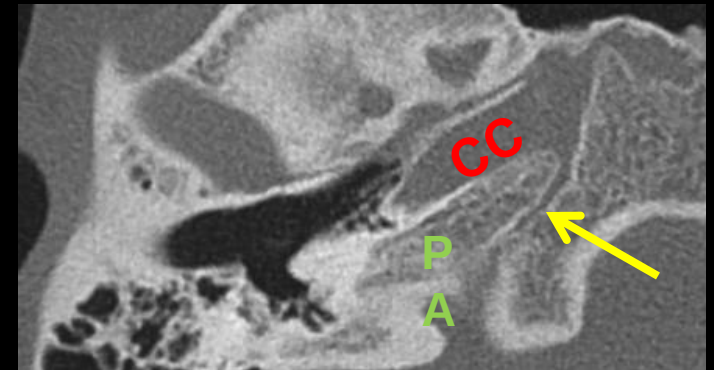
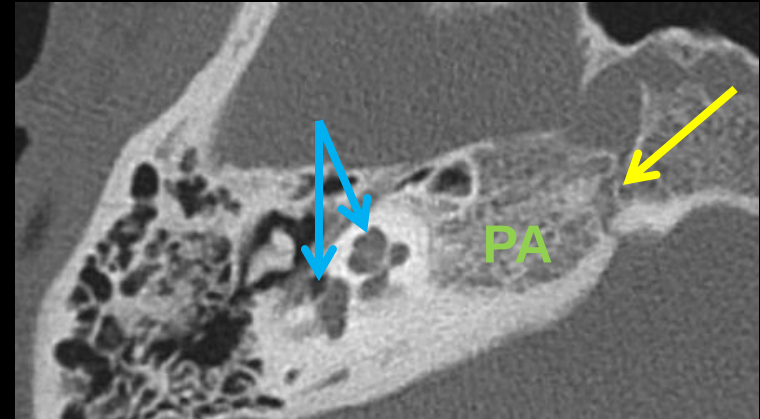


Petrous Apex

- Occipital bone
- Sphenoid bone
- Temporal bone
- Petrooccipital fissure (synchondrosis)

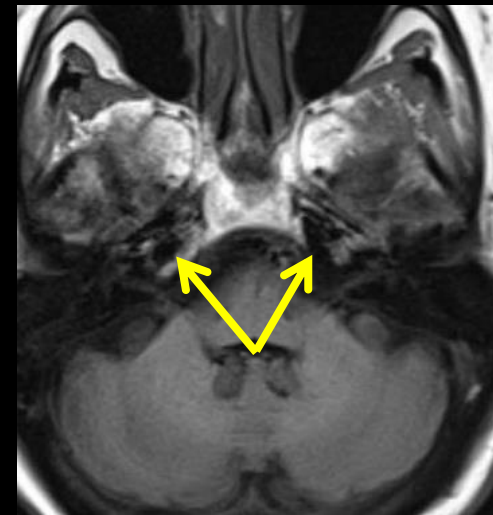
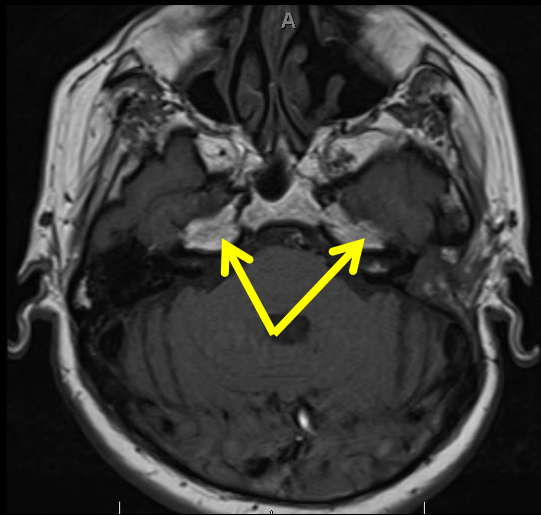


- **Petrous apex (PA)** is a pyramidal shaped part of the temporal bone lying anteromedial to the **inner ear** and lateral to the **petro-occipital fissure**
- **Carotid canal (CC)** lies anterolateral to the petrous apex



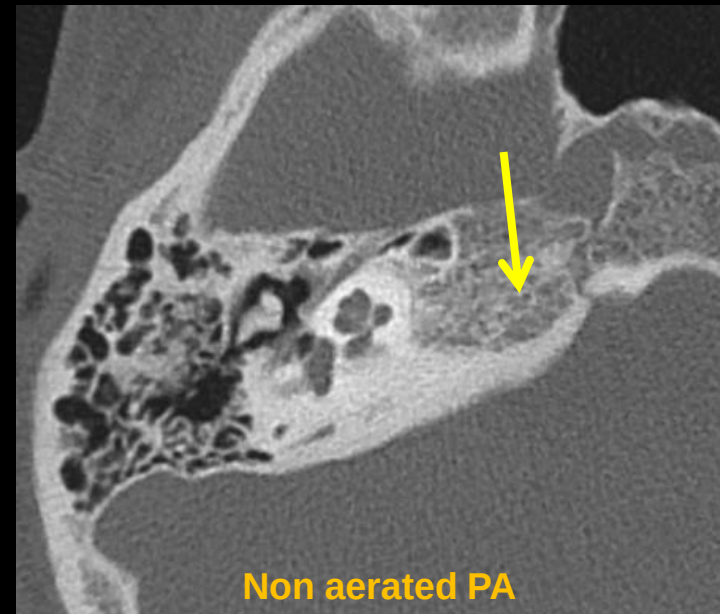
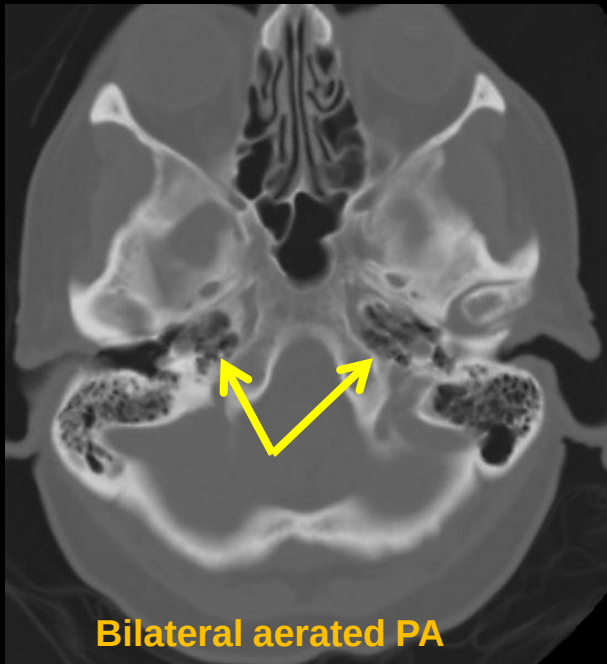
Normal MR Appearance

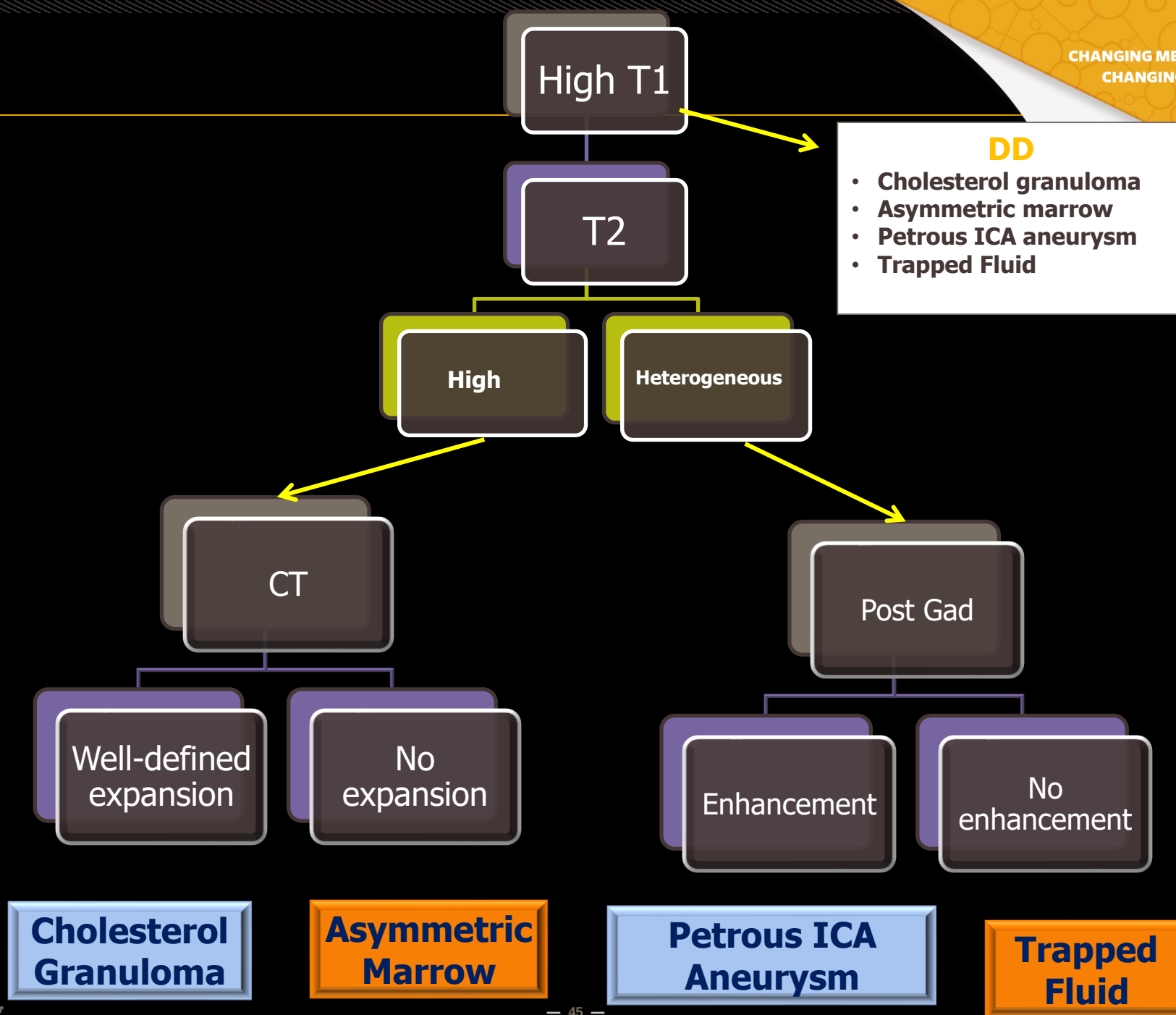
- Petrous apex most commonly is non-pneumatized and appears hyperintense on T1 due to fatty bone marrow
- The aerated petrous apex demonstrates low signal on all MR sequences



Normal CT appearance

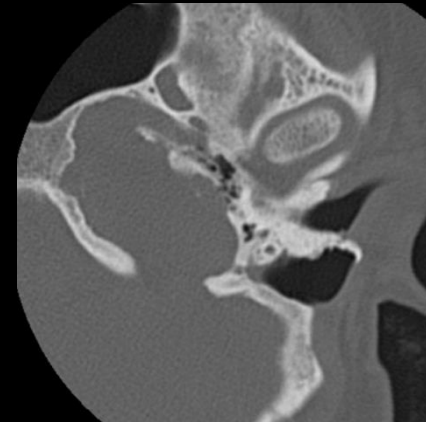
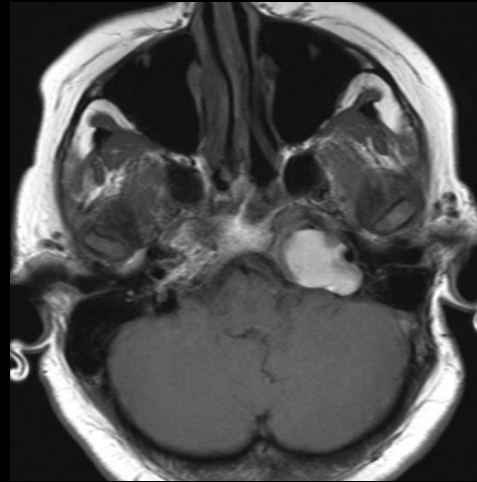
CHANGING MEDICINE.
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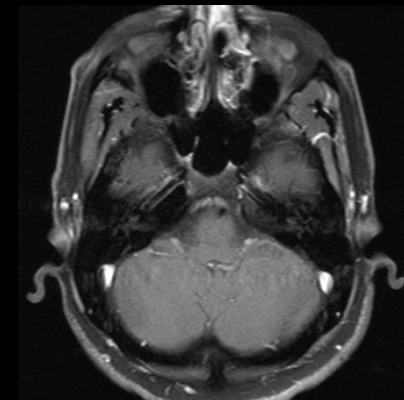
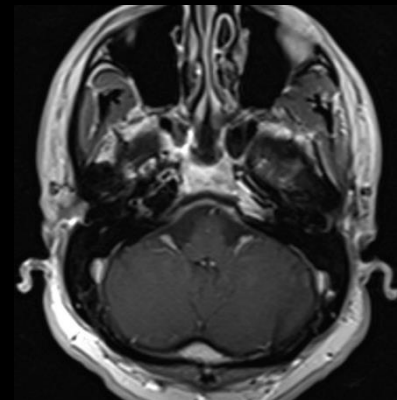
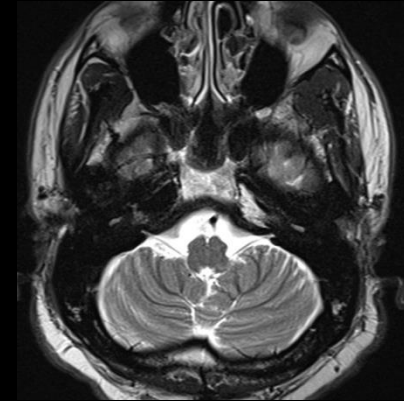
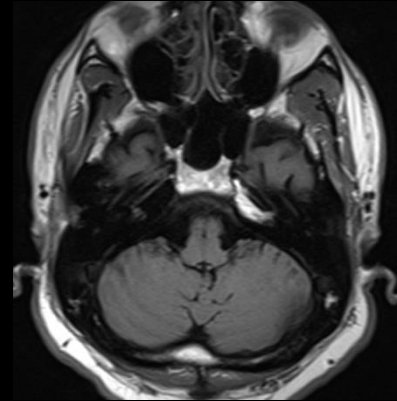
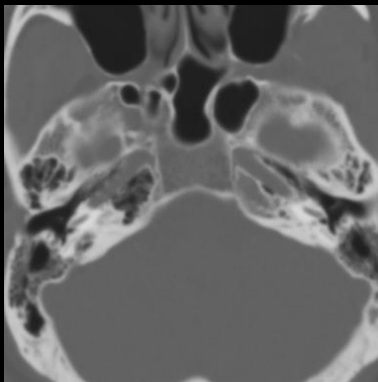
Cholesterol Granuloma

- Most common surgical lesion of the petrous apex
- Etiology is obstruction of air cells in a pneumatized petrous bone
- Contralateral side is usually pneumatized
- **CT**
 - Smooth expansile lesion
 - Large lesions can cause trabecular destruction
- **MRI**
 - T1: High signal on T1 due to blood breakdown products
 - T2: High signal on T2
 - Post Gad: No enhancement



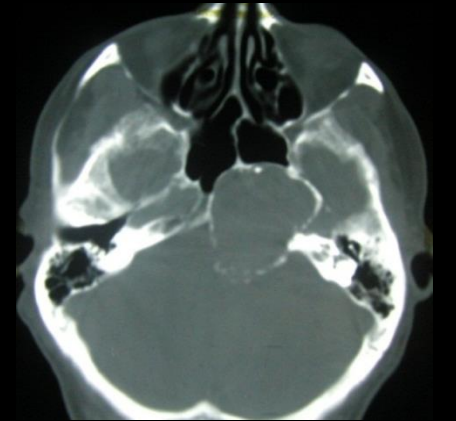
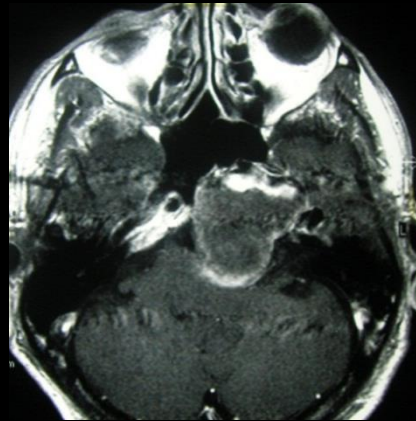
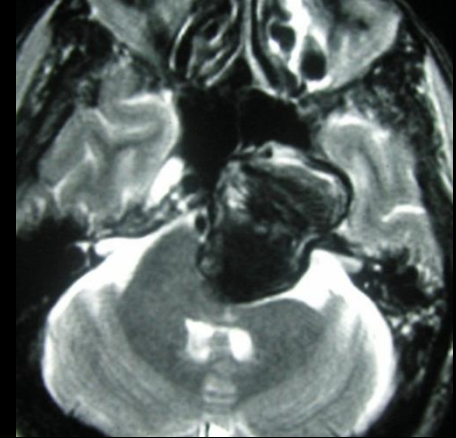
Asymmetric PA aeration

- Pseudolesion i.e. normal variant which mimics as a pathological lesion
- If marrow is asymmetric, either of two sides can be mistaken for pathology, however typically the non-pneumatized marrow is confused with pathology
- Pneumatized PA seen in 33 %
- < 5 % of these are asymmetric.
- **CT**
 - Pneumatized: PA with air cells
 - Non-pneumatized : normal marrow space
- **MRI**
 - Pneumatized: Low T1 and T2 signal without enhancement
 - Non-pneumatized: High T1 and T2 signal due to fatty marrow with suppression on fat sat sequences. On post gad sequence, signal will be high if done without fat saturation. With fat saturation, there will be no enhancement



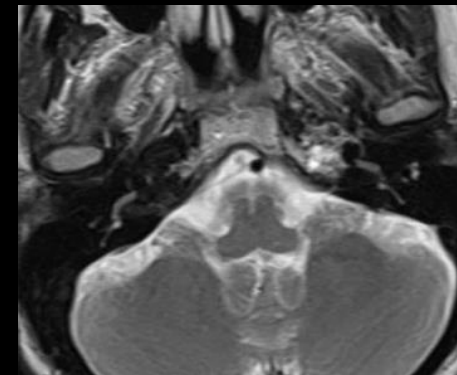
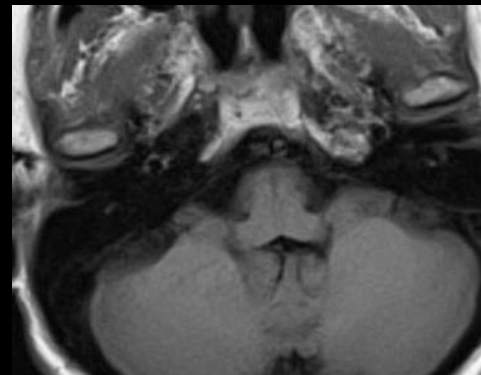
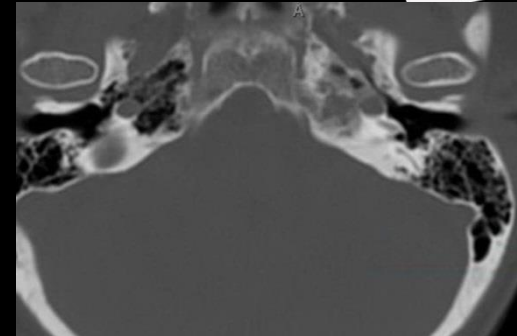
Petrous ICA Aneurysm

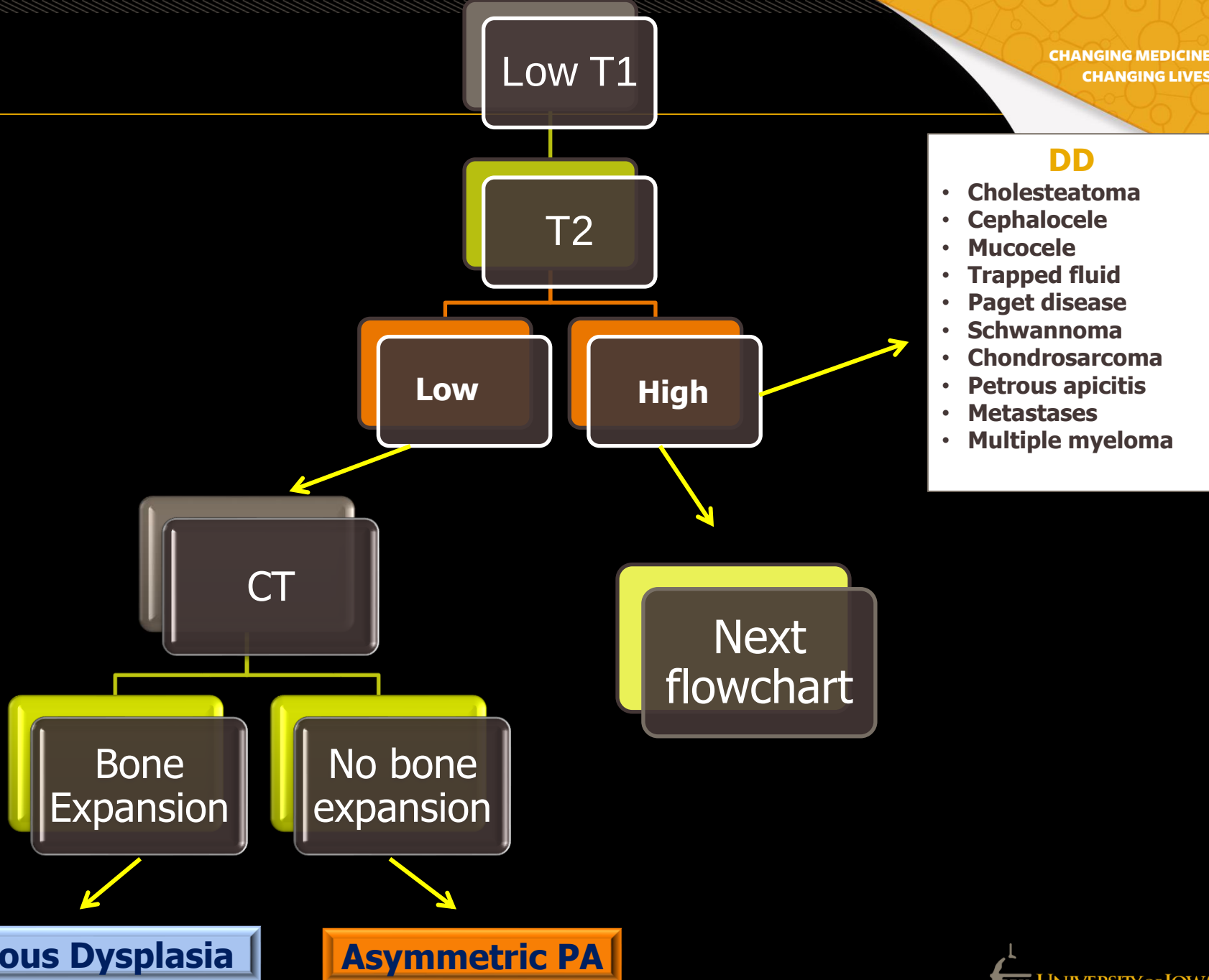
- Aneurysms of the petrous segment of the ICA are rare
- **CT**
 - Expansile mass with or without calcifications
- **MRI**
 - Signal intensity depends on the multiple clots of different ages
 - T1: Complex signal mass usually hyperintense
 - T2: Heterogeneous with internal flow voids
 - Post Gad: Diffuse enhancement. Thrombosed portion doesn't enhance



Trapped fluid

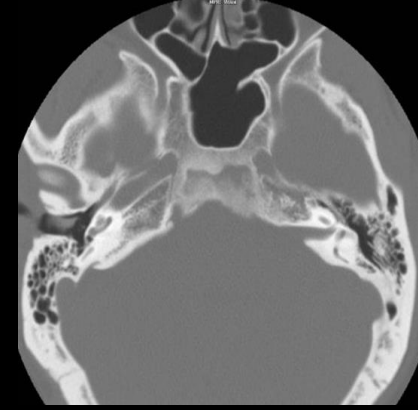
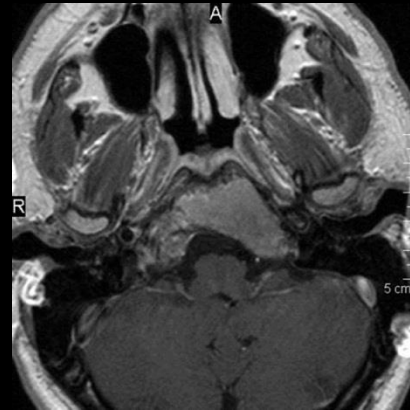
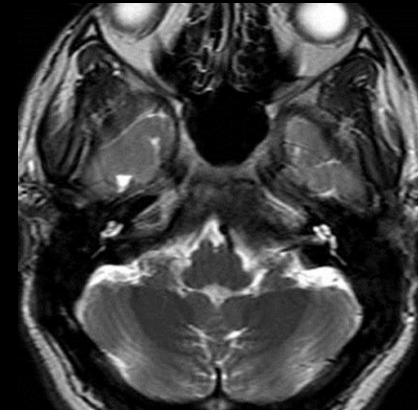
- Most common lesion of the PA
- Fluid collection in pneumatized PA after otomastoiditis
- “Do not touch lesion”
- **CT**
 - Opacified PA air cells without bony expansion or destruction
- **MRI**
 - T1: Low T1 signal is most common, but can be high or intermediate depending on the protein content
 - T2: High T2 signal
 - Post Gad: No enhancement





Fibrous dysplasia

- Fibrous dysplasia is characterized by progressive replacement of normal bone by dysplastic fibro-osseous tissue
- Skull and facial bones are involved in 25 % with monostotic FD and 50 % with polyostotic FD.
- Polyostotic form is usually unilateral
- **CT**
 - Bone expansion with increased density and ground glass pattern. Otic capsule is rarely involved
- **MRI**
 - T1: Low signal
 - T2: Low signal
 - Post Gad: Avid enhancement



Courtesy:
Dr. Ilona M. Schmalfuss

Low T1

High T2

Post Gad

Enhancement

No
enhancement

DD

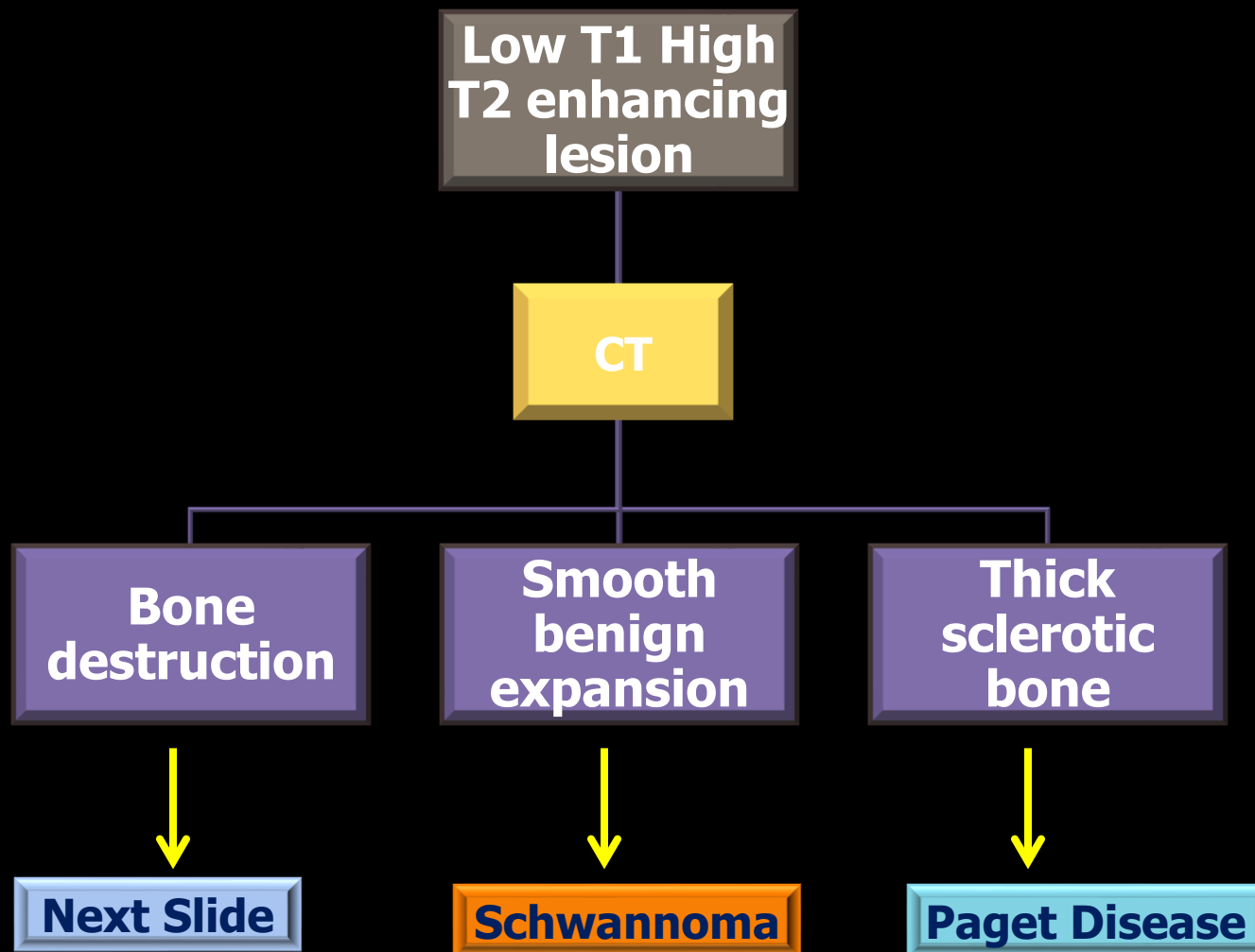
- Schwannoma
- Paget disease
- Petrous apicitis
- Chondrosarcoma
- Metastases
- Multiple myeloma

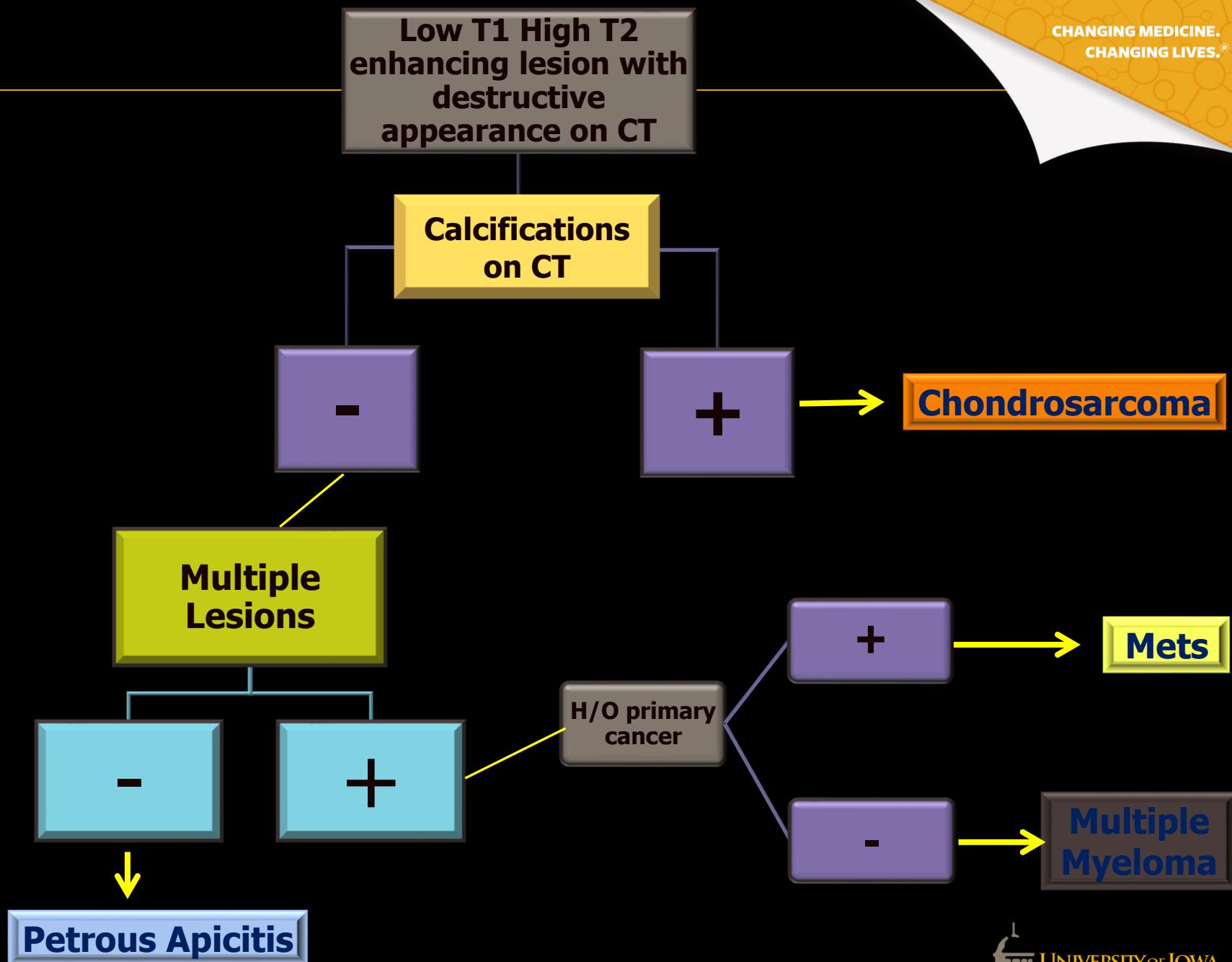
DD

- Cholesteatoma
- Mucocele
- Cephalocele
- Trapped fluid

Next slide

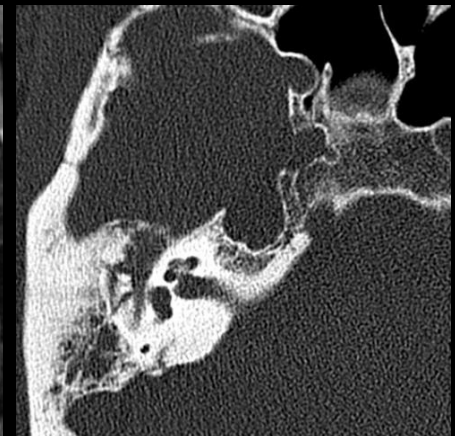
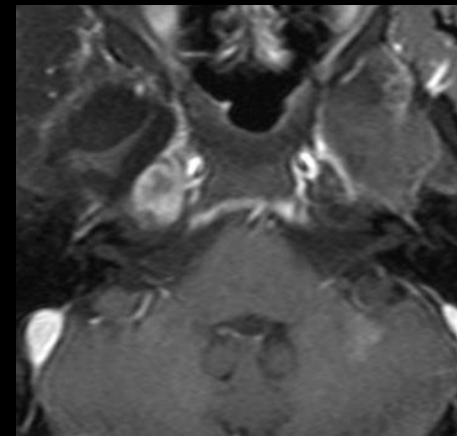
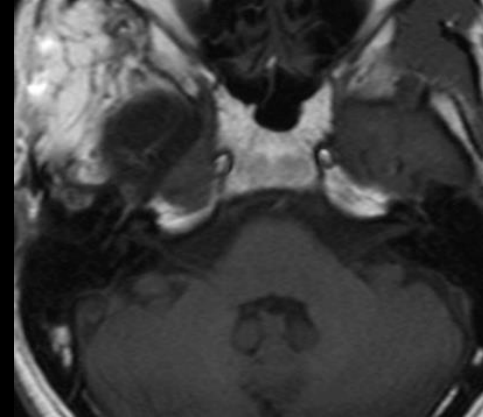
Other
flowchart





Schwannoma

- Trigeminal nerve schwannoma can involve the petrous apex
- They usually arise in the Meckel cave or cisternal segment of the trigeminal nerve
- **CT**
 - Smooth bony remodelling
- **MRI**
 - T1: Low/Isointense
 - T2: High signal intensity, cystic components can be seen causing very high signal on T2
 - Post Gad: Strong heterogeneous enhancement



Paget Disease

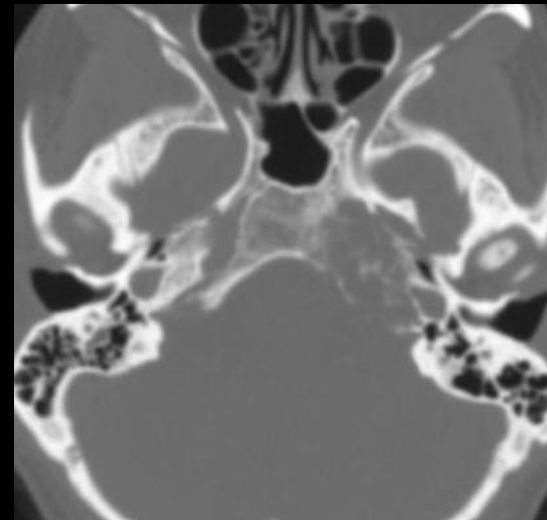
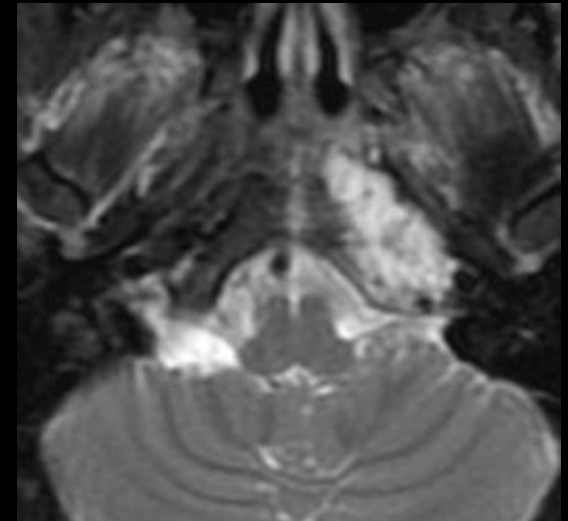
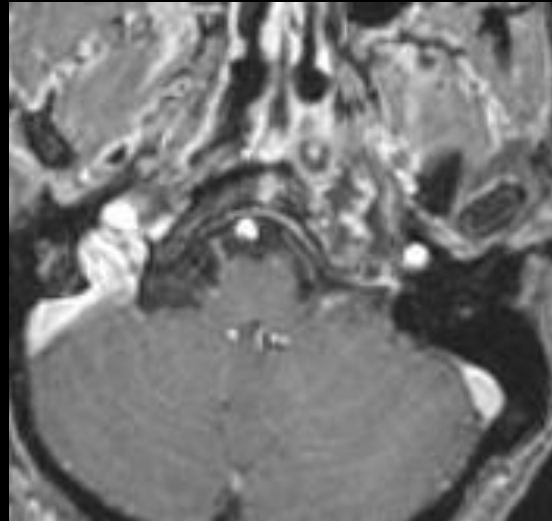
- Disorder of bone remodelling
- PA is the earliest part of temporal bone to be involved
- Otic capsule involvement occurs late
- **CT**
 - Early phase: Demineralization
 - Intermediate phase: Mixed lytic and blastic activity
 - Late phase: Thick and expanded bone
- **MRI**
 - T1: Usually low T1 with bone expansion
 - T2: High signal
 - Post Gad: Heterogeneous enhancement



Courtesy:
Dr. Deborah L. Reede

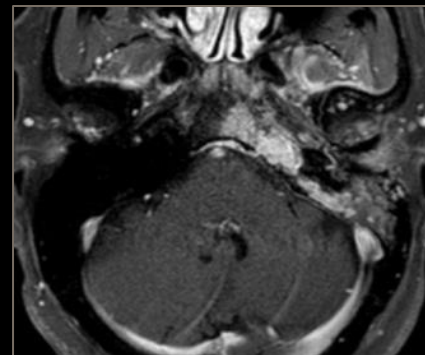
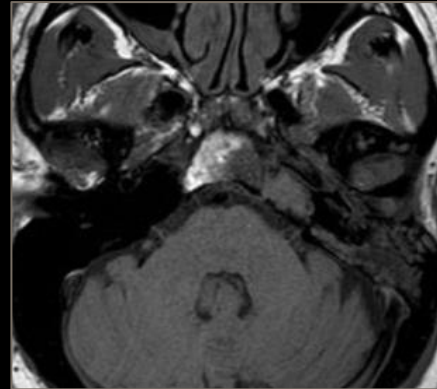
Chondrosarcoma

- Malignant cartilaginous tumor arising from the petro-occipital fissure
- 6% of all skull base tumors
- **CT**
 - Characteristic chondroid (arc or ring like) calcifications. Bone destruction
- **MRI**
 - T1: Low signal
 - T2: High signal. Low signal can be seen due to calcifications
 - Post Gad: Heterogeneous enhancing lesion



Petrous Apicitis

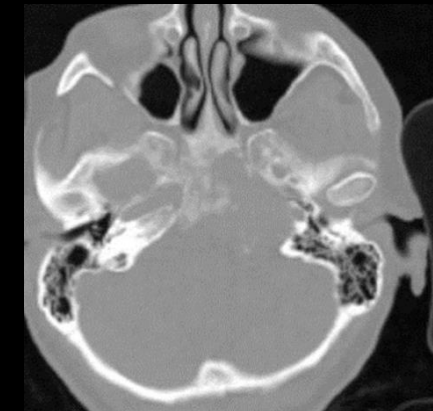
- Rare complication of otomastoiditis where infection extends from mastoid air cells to pneumatized petrous apex air cells
- **CT**
 - Opacified PA air cells with trabecular destruction
 - Fluid in the middle ear and mastoid air cells is usually seen
- **MRI**
 - T1: Low/Intermediate signal
 - T2: High fluid signal
 - Post gad: Peripheral enhancing fluid-filled PA
 - Infection can extend to the cavernous sinus or Meckel cave
- Skull base osteomyelitis can involve the PA



Metastases

- Hematogenous metastases can involve the PA
- Breast, lung, prostate, melanoma and renal cancer are the common primary sites
- **CT**
 - Lytic lesion, large lesions cause bone destruction
- **MRI**
 - T1: Low-Intermediate
 - T2: Depends on the primary site, usually high
 - Post Gad: Usually enhancement is present

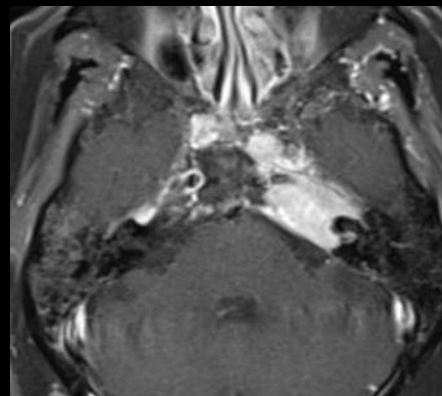
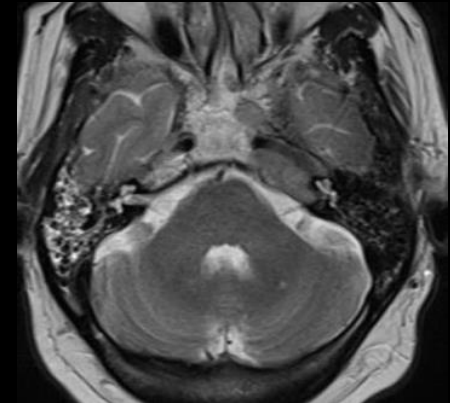
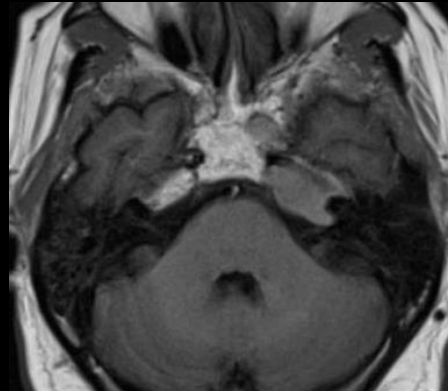
H/O thyroid cancer

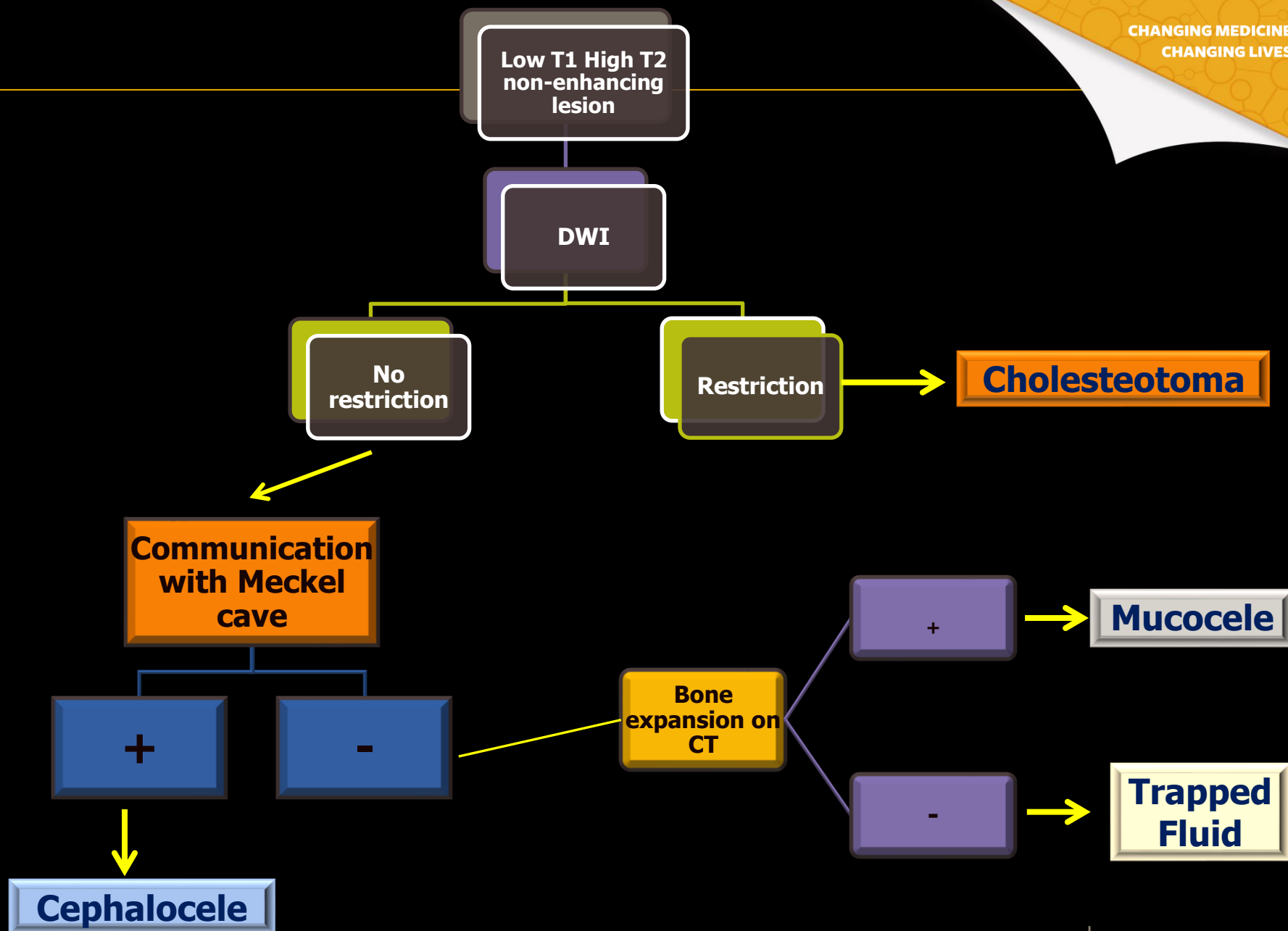


Courtesy:
Dr. Lindell R. Gentry

Multiple Myeloma

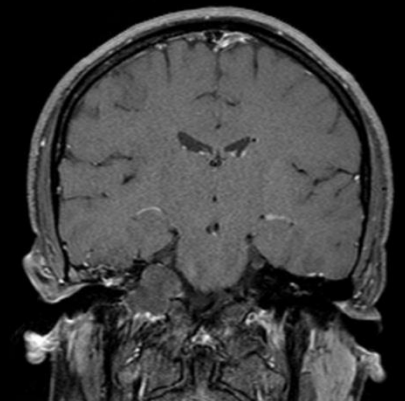
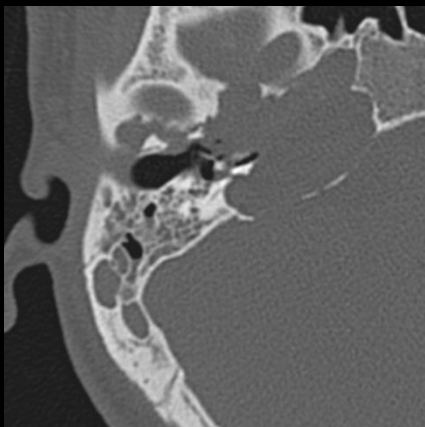
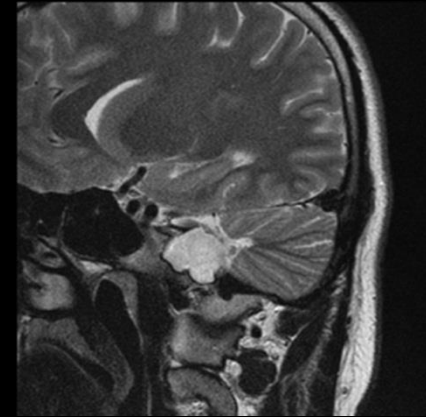
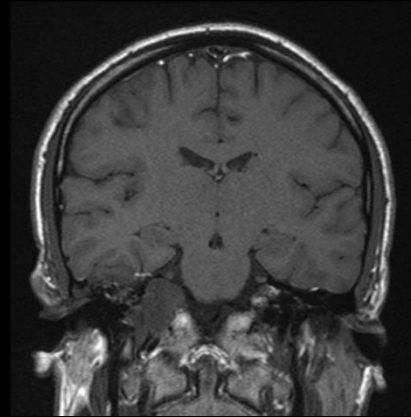
- Multifocal proliferation of plasma cells
- **CT**
 - Multiple lesions
 - Expansile lesion with lytic destruction
- **MRI**
 - Multiple lesions
 - T1: Low signal
 - T2: High signal
 - Post Gad: Homogenous enhancement





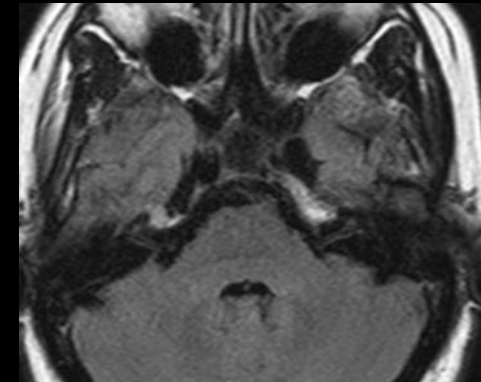
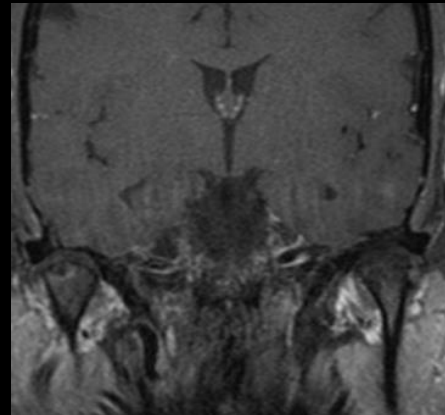
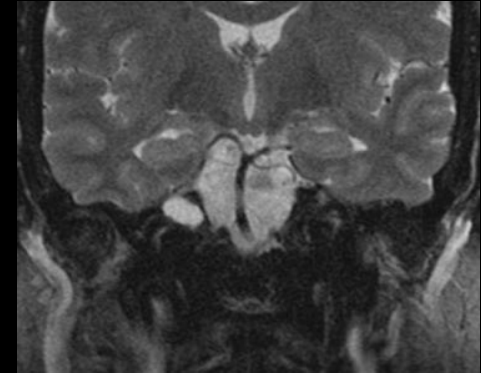
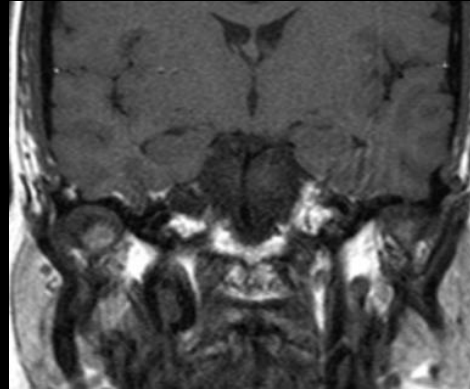
Cholesteatoma

- Petrous apex cholesteatoma is rare
- It can be congenital (more common) or acquired
- **CT**
 - Smooth expansile lesion. Large lesions may show bone erosions
- **MRI**
 - T1: Low
 - T2: High
 - FLAIR: No or partial attenuation
 - DWI: Restricted diffusion
 - Post Gad: No enhancement



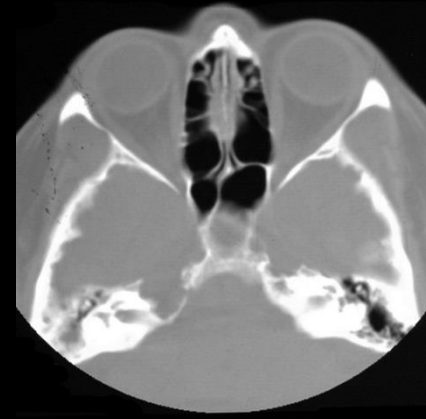
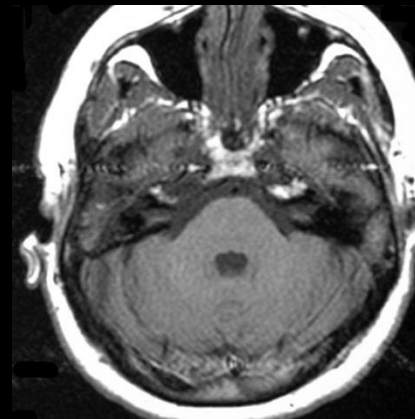
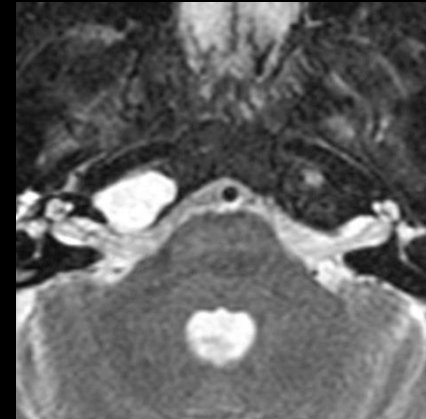
Cephalocele

- Herniation of posterolateral wall of the Meckel cave into PA
- 30% are bilateral
- **CT**
 - Smooth expansion
- **MRI**
 - T1: Low signal isointense to CSF
 - T2: High signal isointense to CSF, communicates with the Meckel cave
 - FLAIR: Complete suppression
 - Post Gad: No enhancement



Mucocele

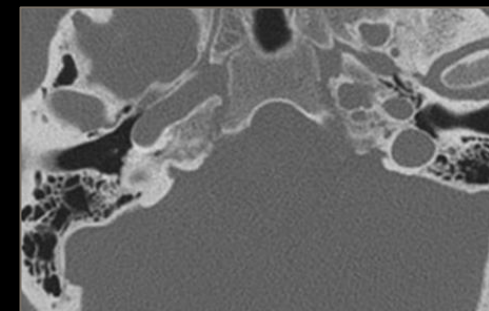
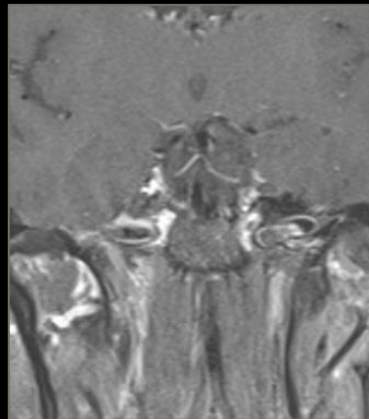
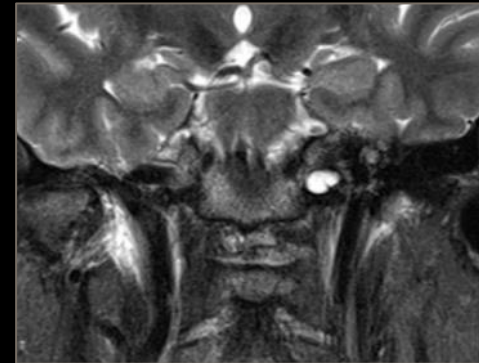
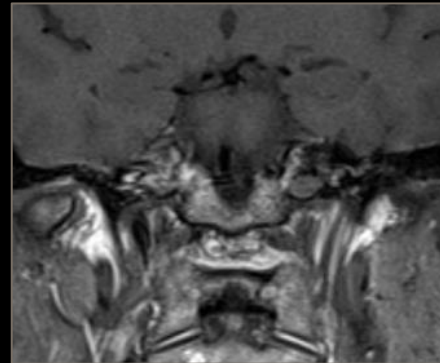
- Rare lesion caused by post-inflammatory obstruction
- **CT**
 - Smooth expansile lesion
- **MRI**
 - T1: Low SI
 - T2: High SI
 - Post Gad: No enhancement
 - DWI (Diffusion): No restriction

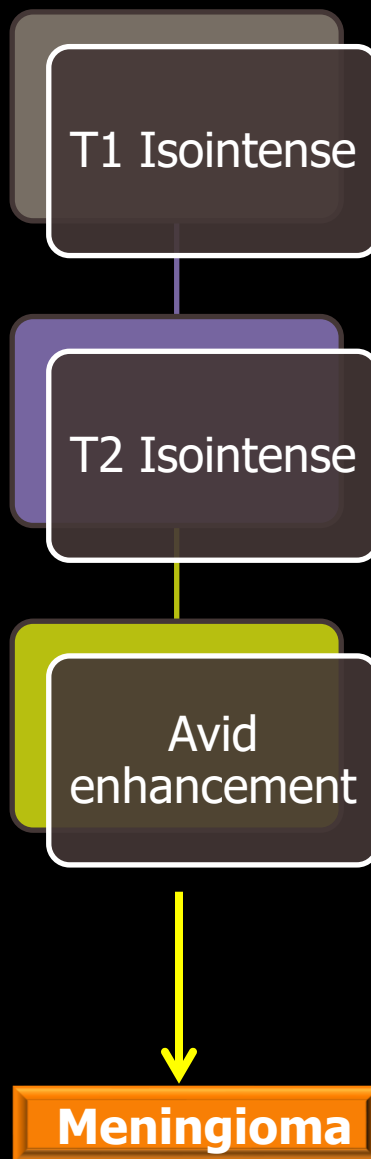


Courtesy:
Dr. Deborah L. Reede

Trapped fluid

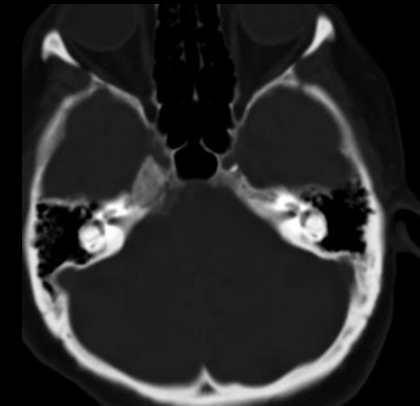
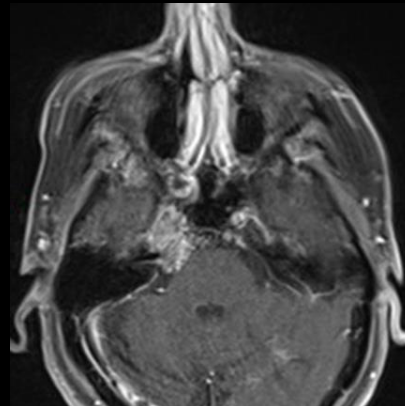
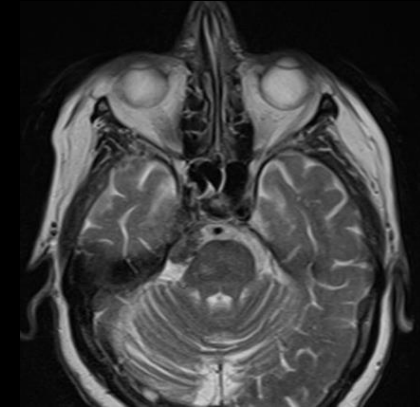
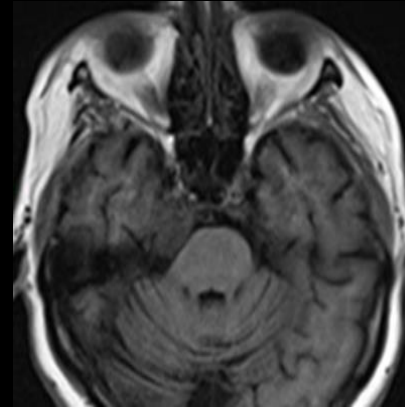
- Most common lesion of the PA
- Fluid collection in pneumatized PA after otomastoiditis
- “Do not touch lesion”
- **CT**
 - Opacified PA air cells without bony expansion or destruction
- **MRI**
 - T1: Low T1 signal is most common, but can be high or intermediate depending on the protein content
 - T2: High T2 signal
 - Post Gad: No enhancement





Meningioma

- Both CP angle and petroclival meningiomas can involve the petrous apex
- **CT**
 - Permeative and sclerotic bone changes
- **MRI**
 - T1: Isointense
 - T2: Iso to slightly hyperintense
 - Post Gad: Avid homogenous enhancement, dural tail can be seen



Thank you