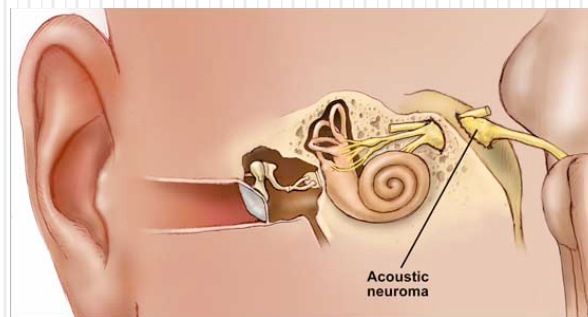


Vestibular Schwannomas Made Ridiculously Simple

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January 2010



What are we talking about?

- Nerve sheath tumor
- Superior and inferior vestibular nerves (hence 'vestibular' and not 'acoustic')
- The most common tumor of the CPA



Conjoined fetus lady. South Park, episode 18, 1998.

Other facts...

- 6 % of all intracranial tumors
- Incidence in US: 10 per million / year
 - 2000-3000 individuals diagnosed per year (US)
- Vast majority in adulthood
- 95% Sporadic (unilateral)
- 5% Neurofibromatosis type 2 (bilateral)
- No known race or gender predilection
- Average growth rate is 1.8 mm/year

Pathogenesis

- Neither Neuroma or Acoustic (auditory)
- Schwannoma arising from vestibular nerve
Schwann cells at transition zone of the peripheral and central myelin (at the lateral CPA / medial IAC)
 - *For otologists only:* **Obersteiner-Redlich zone**
 - Rarely arise from cochlear nerve
- Malignant degeneration exceedingly rare
- Majority originate within the IAC
- Equal frequency on Superior and Inferior vestibular nerves (*controversial*)



Possible In-Service Material

- VS occurs as a result of mutations in a tumor suppressor gene
 - **Merlin**
 - Located on chromosome 22q12
- VS requires both copies to be mutated
- People with NF2 inherit one mutated gene





Jackler Staging System

For Trac, Mia, Audrey, and Ashley

Stage	Tumor Size
Intracanalicular	Tumor confined to IAC
I (small)	< 10 mm
II (medium)	11-25 mm
III (Large)	25-40 mm
IV (Giant)	> 40 mm

What do patients complain of?

- Hearing loss
 - 95% of patients
 - Most have slowly progressive loss with noise distortion
 - 20% have sudden HL
 - Level of hearing loss is NOT a predictor of size
- Tinnitus
 - 65% of patients
 - Usually constant with a high buzzing pitch
- Disequilibrium
 - 60% of patients
 - Usually well-compensated



More complaints....

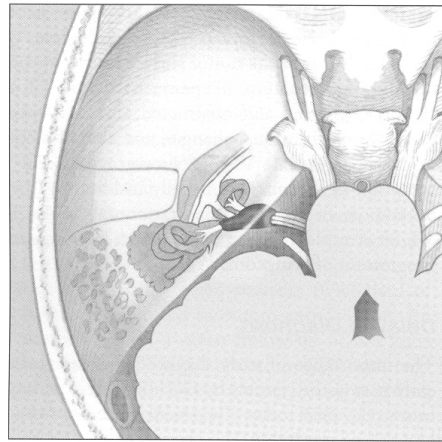
- Facial and trigeminal nerve dysfunction
 - Usually V2 numbness
 - Sensory component of CN VII is usually involved first
 - Hitselberger sign – numbness of the posterior EAC
 - Facial weakness or spasm occurs in 17% of patients
- Rare symptoms
 - Decreased visual acuity and diplopia
 - May occur from rapid expansion and compression of II, IV, or VI
 - Headache, altered MS, nausea, papilledema
 - Hydrocephalus
 - Compression of CN's IX and X

Phases of Tumor Growth

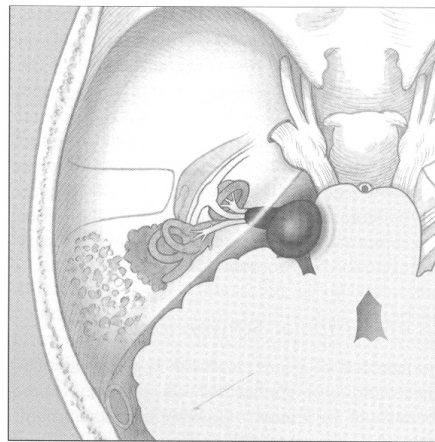
- **Intracanalicular:**
 - Hearing loss, tinnitus, vertigo
- **Cisternal:**
 - Worsened hearing and dysequilibrium
- **Compressive:**
 - Occasional occipital headache
 - CN V: Midface, corneal hypesthesia
- **Hydrocephalic:**
 - Fourth ventricle compressed and obstructed
 - Headache, visual changes, altered mental status

Phases of Tumor Growth

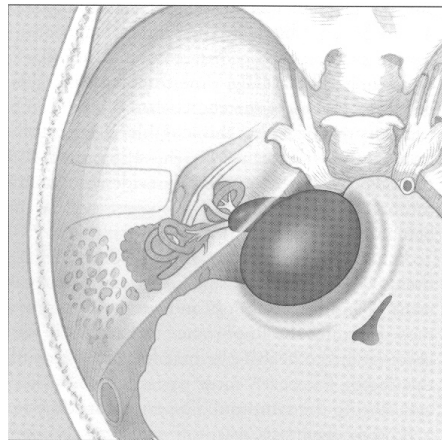
Intracanalicular



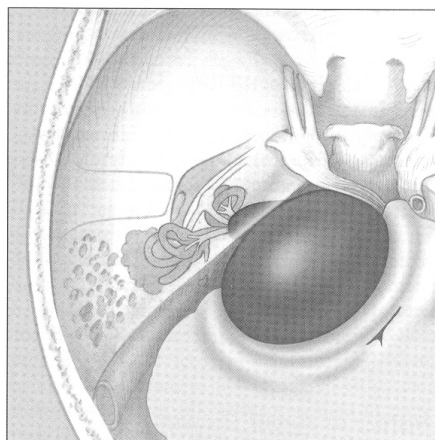
Cisternal



Compressive



Hydrocephalic

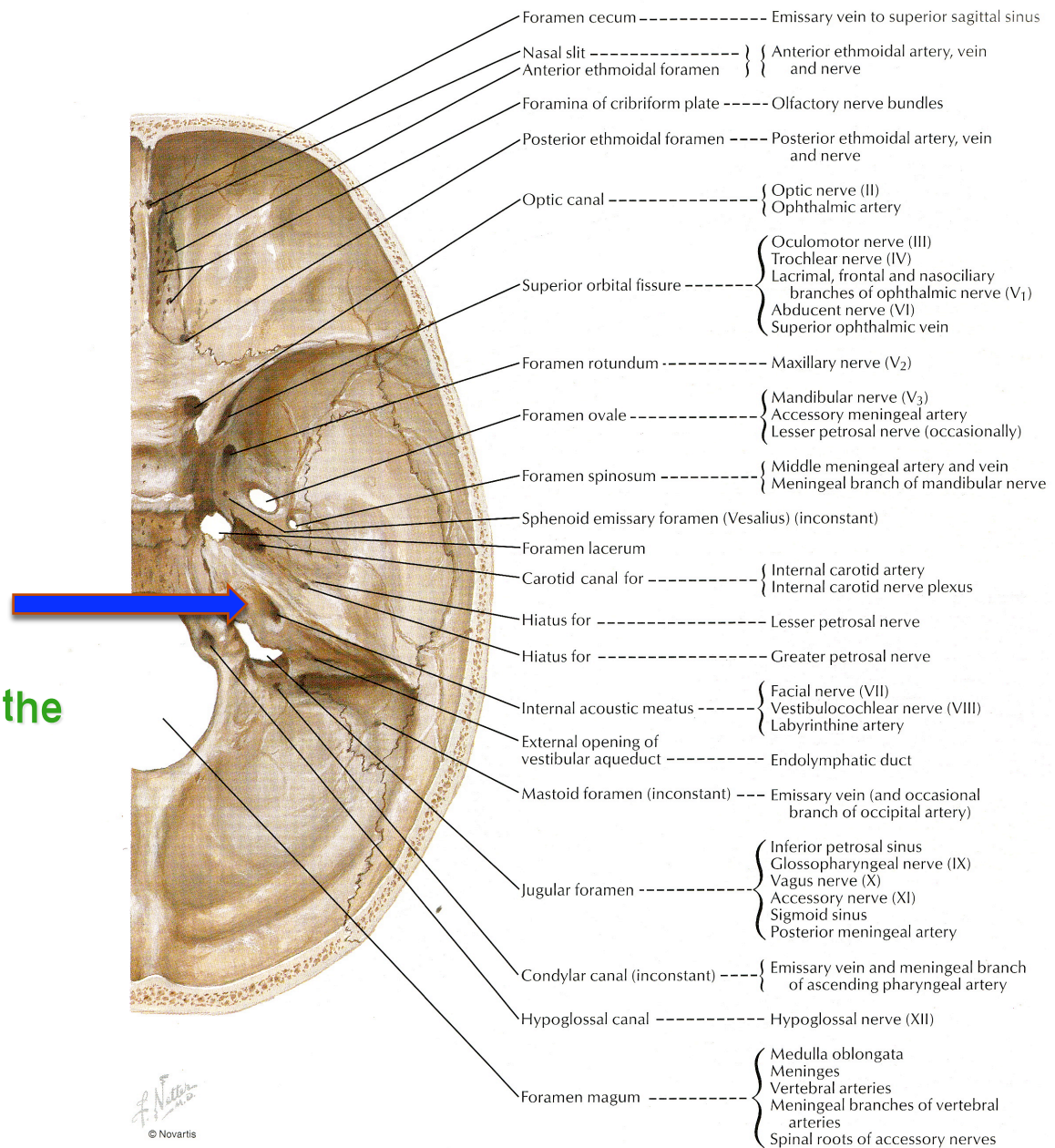


Jackler RK. 2000, p. 180: Tumors of the Ear and Temporal Bone

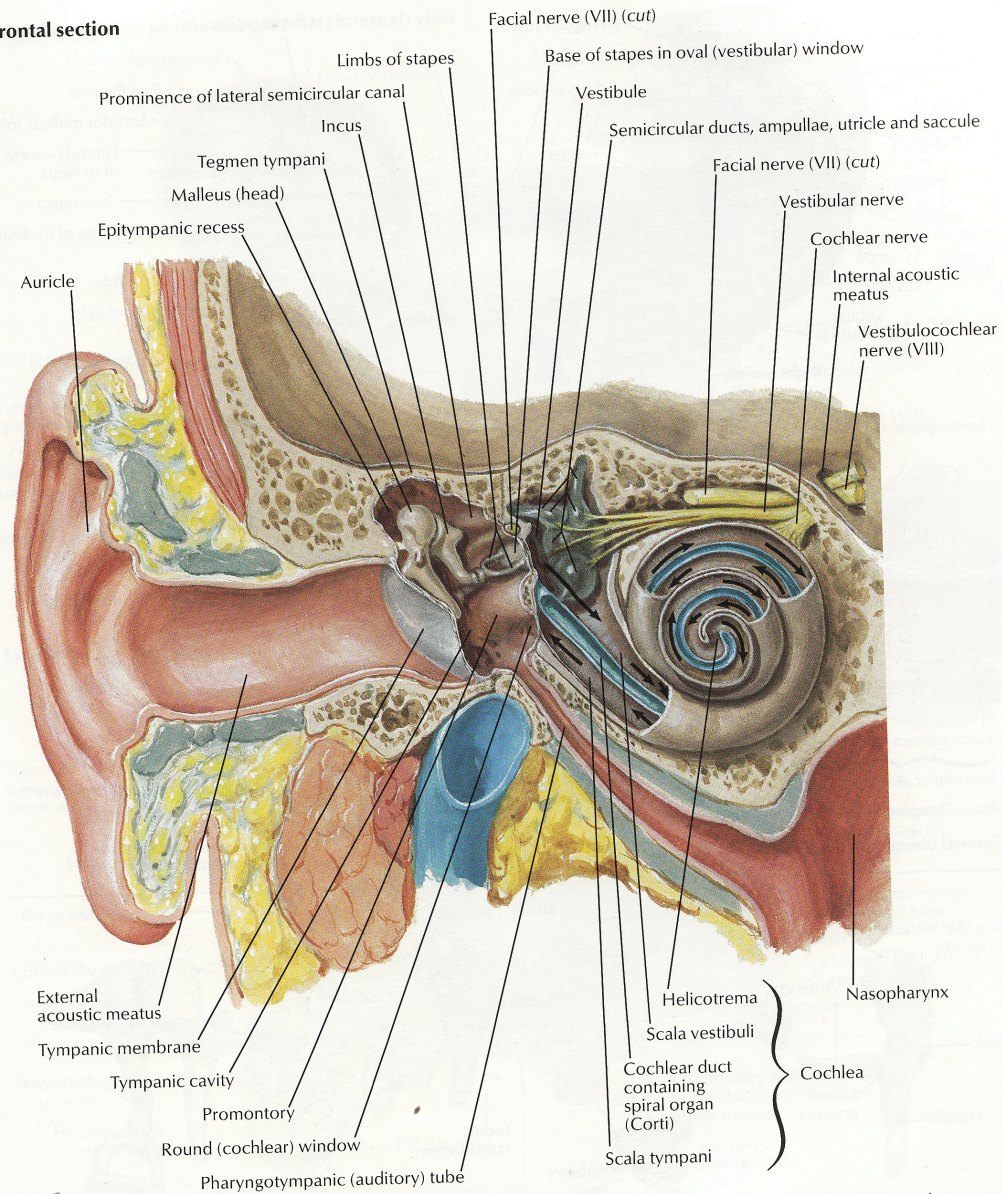
Anatomy

What bone is this?

Petrous portion of the temporal bone

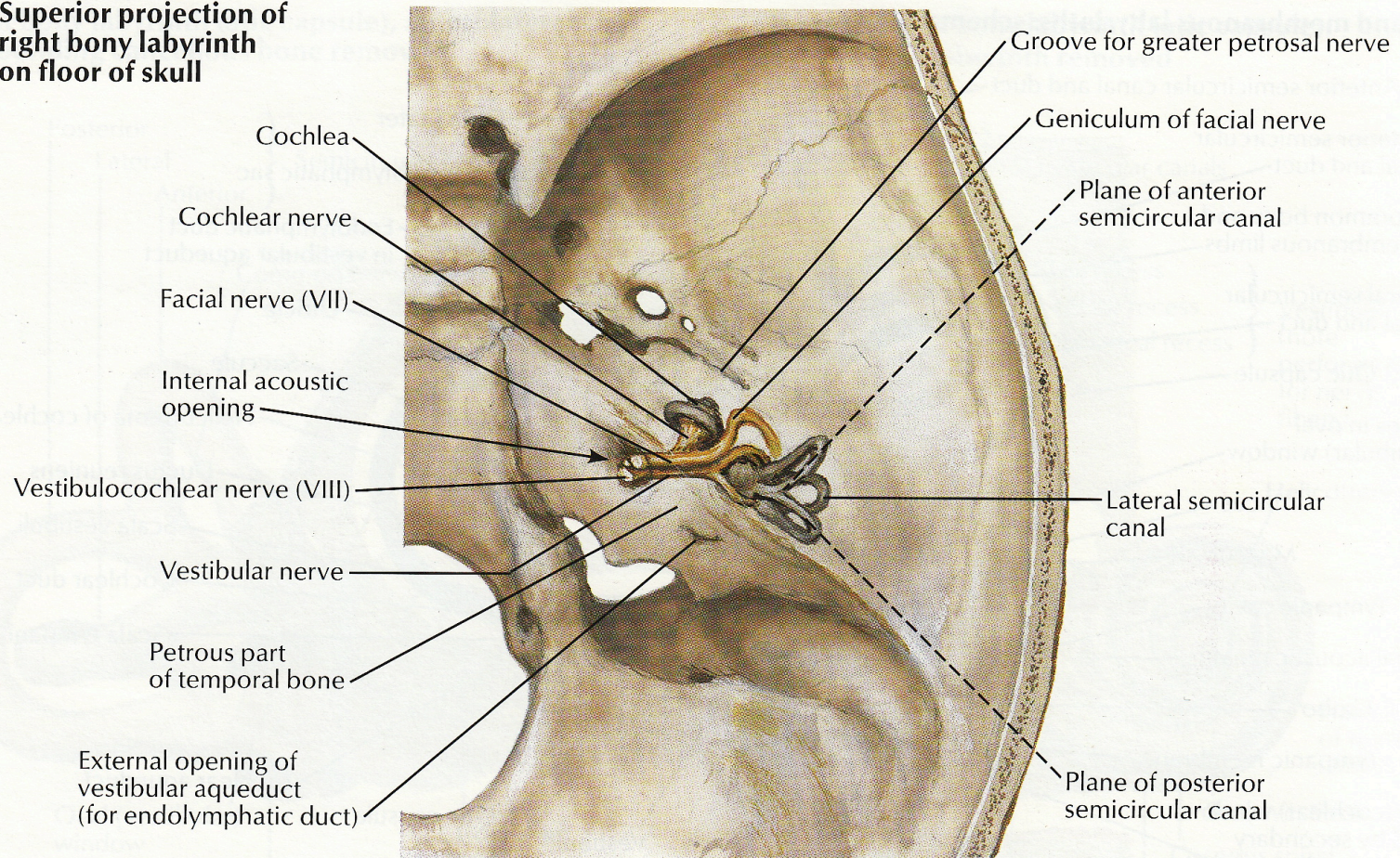


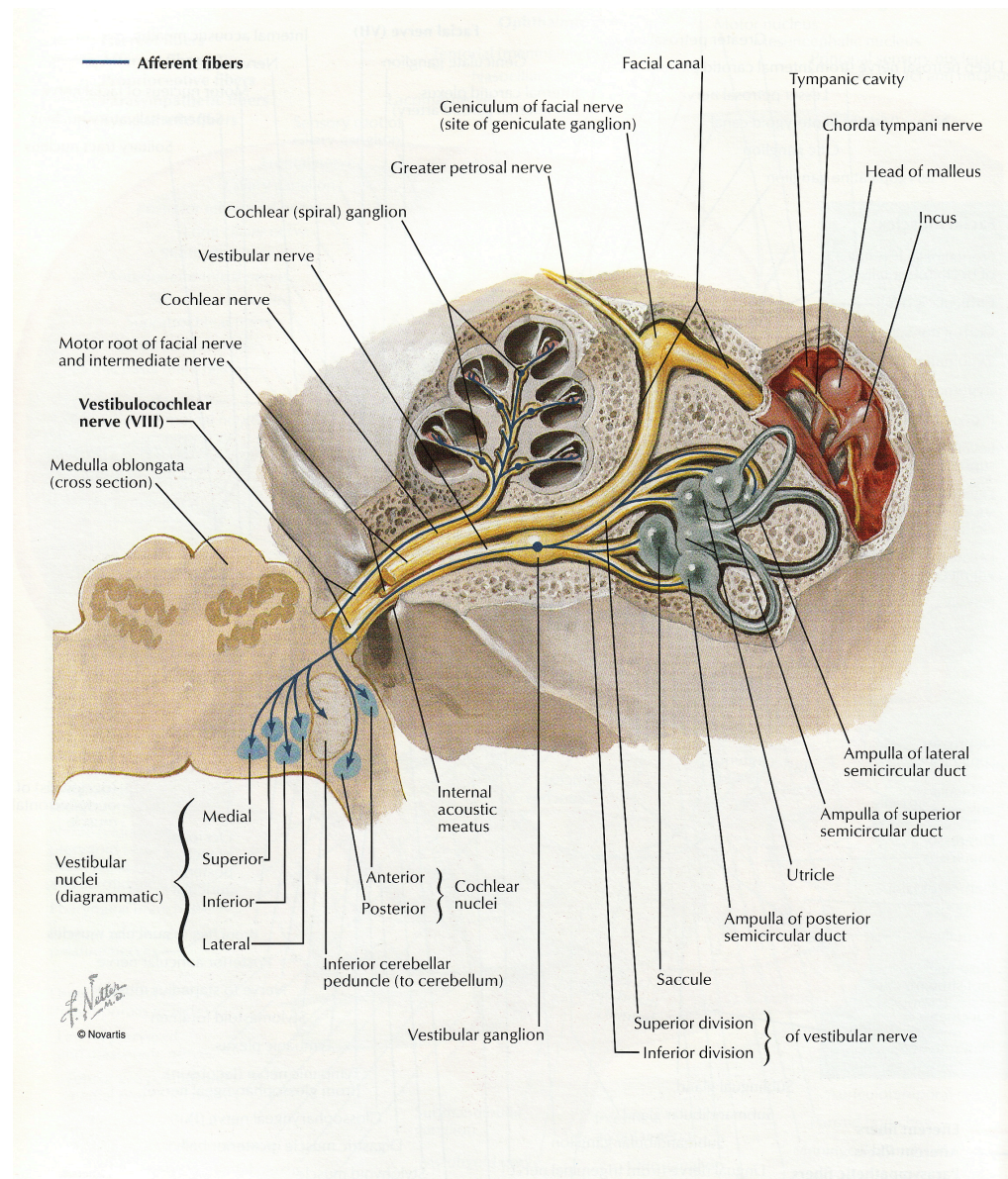
Frontal section

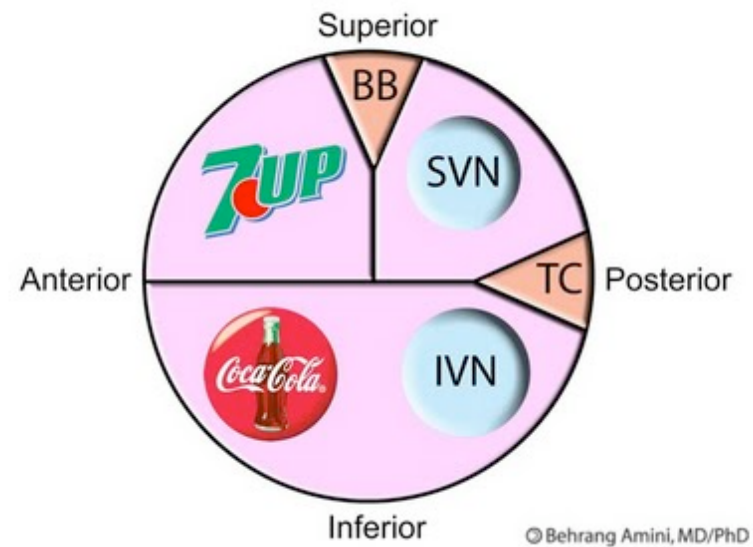
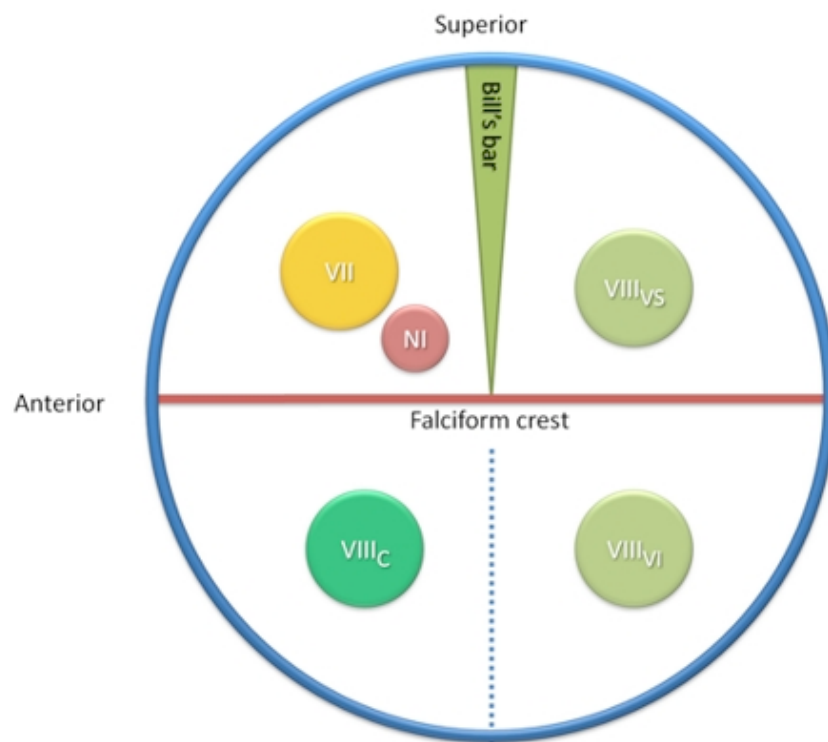


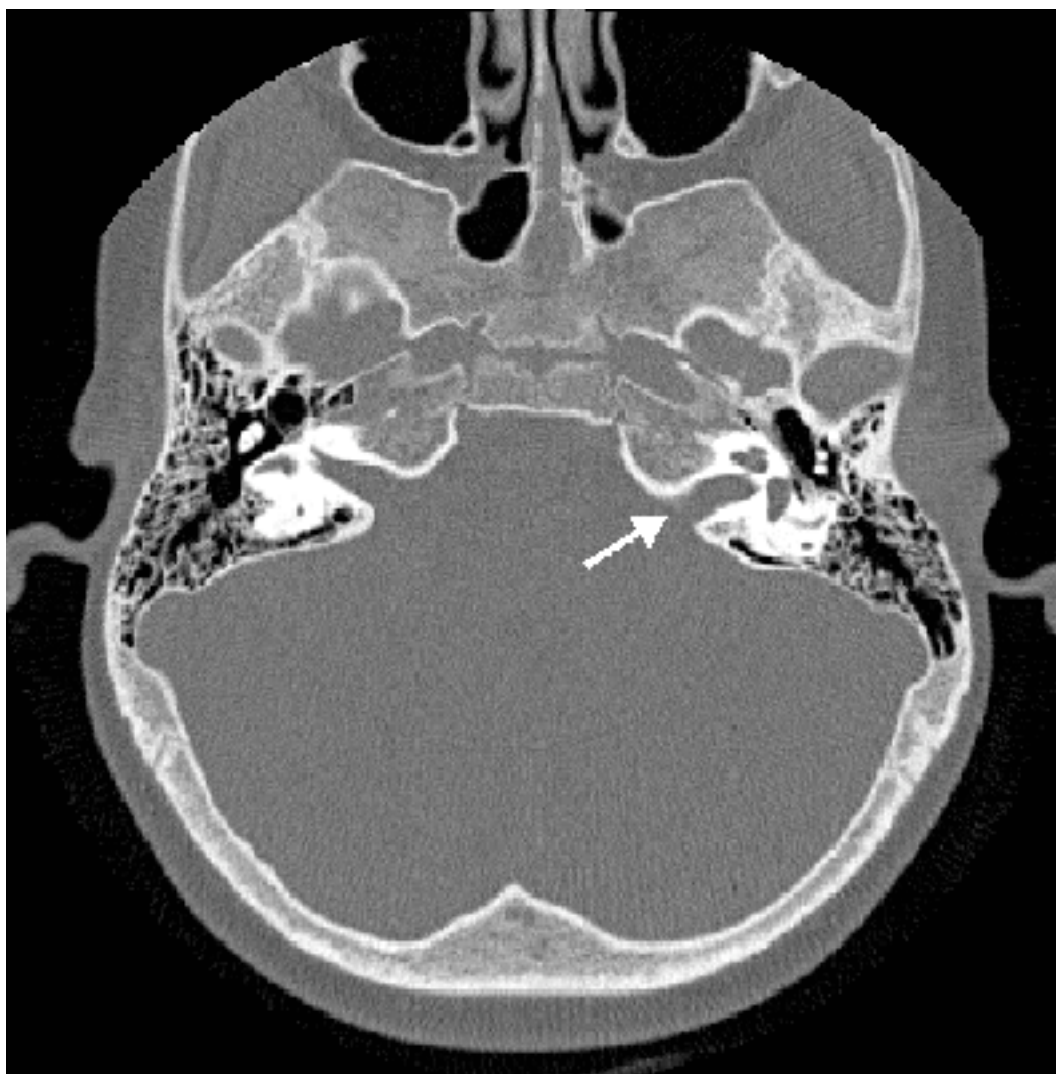
Note: Arrows indicate course of sound waves

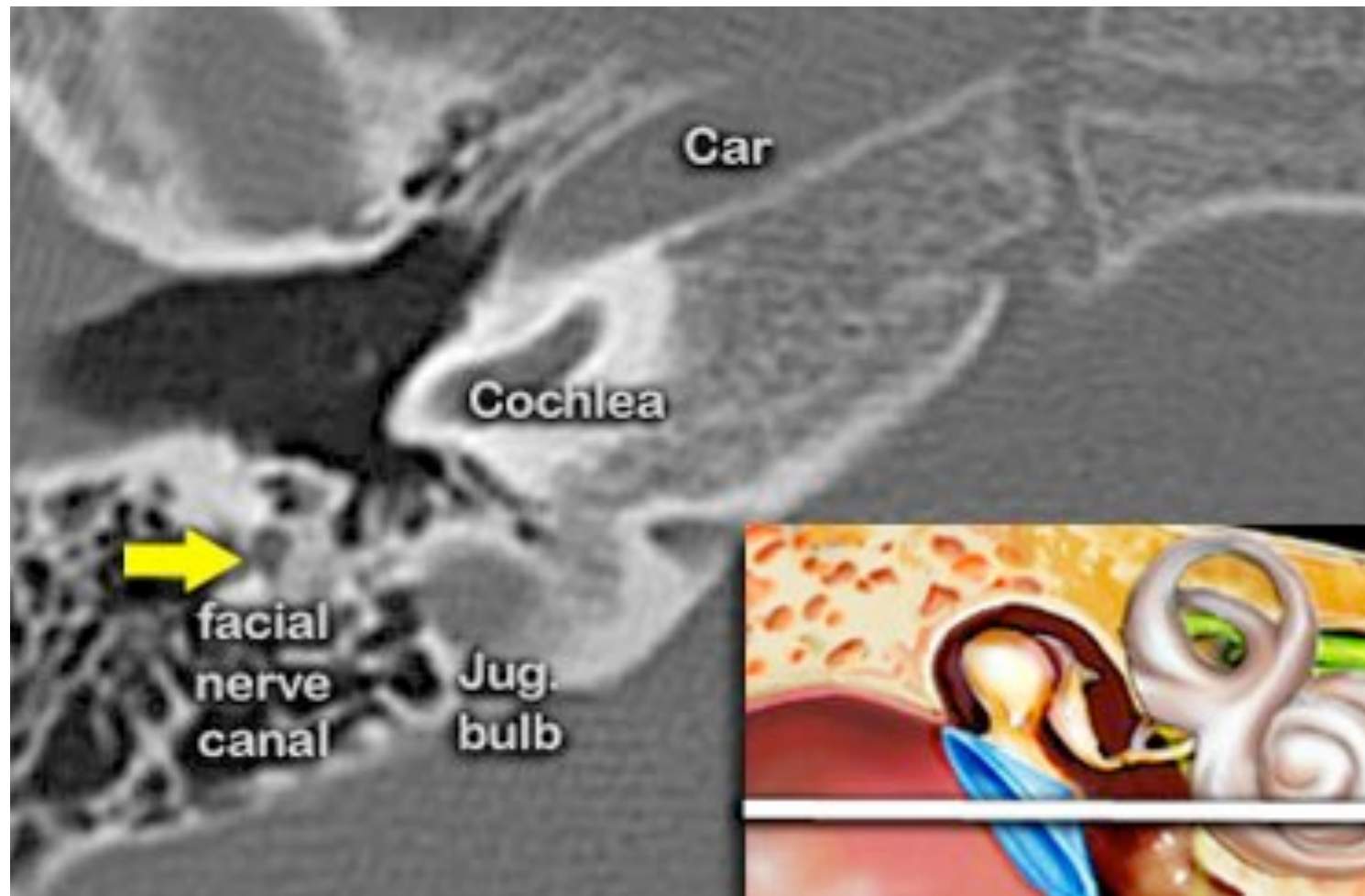
**Superior projection of
right bony labyrinth
on floor of skull**

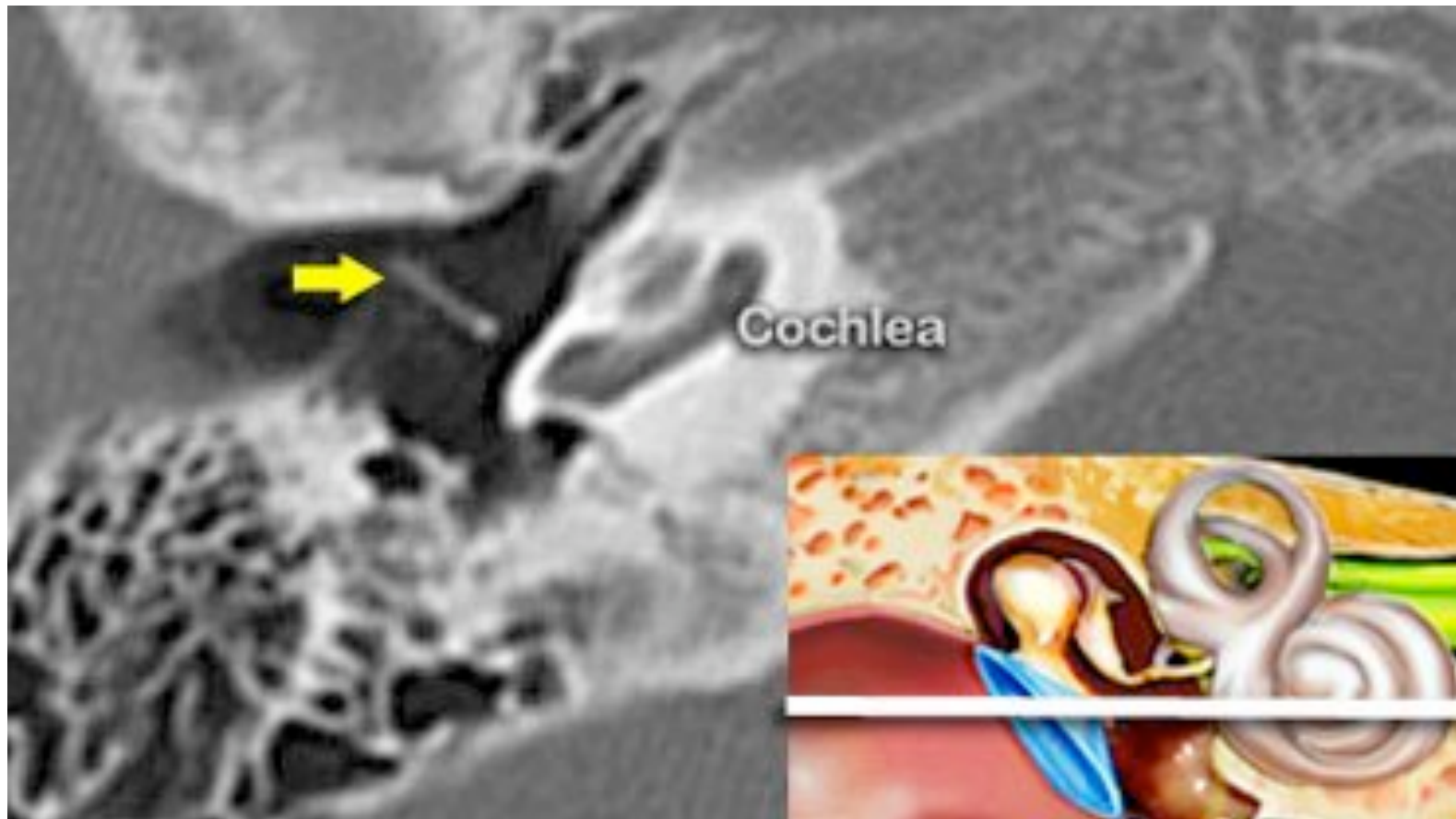


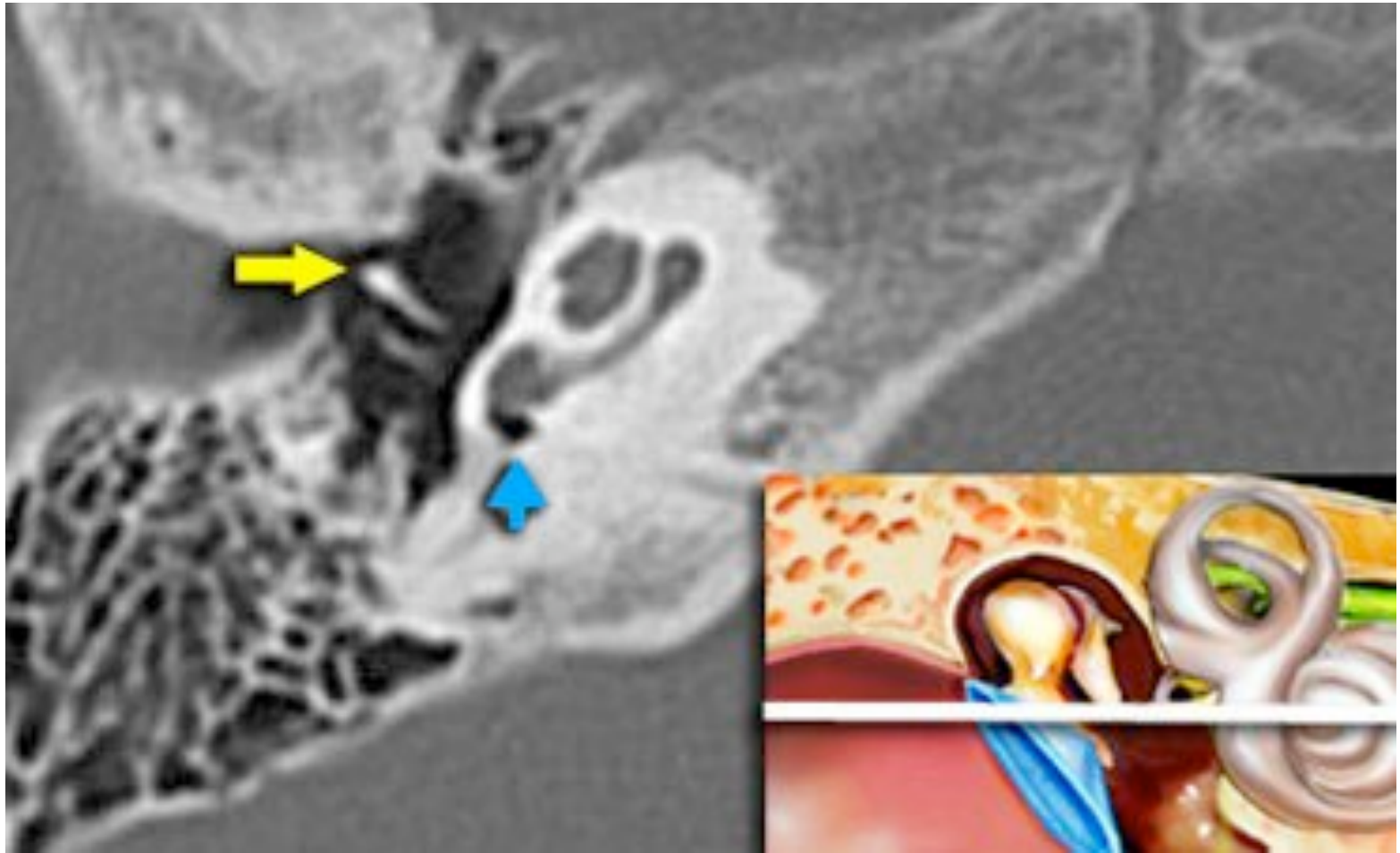


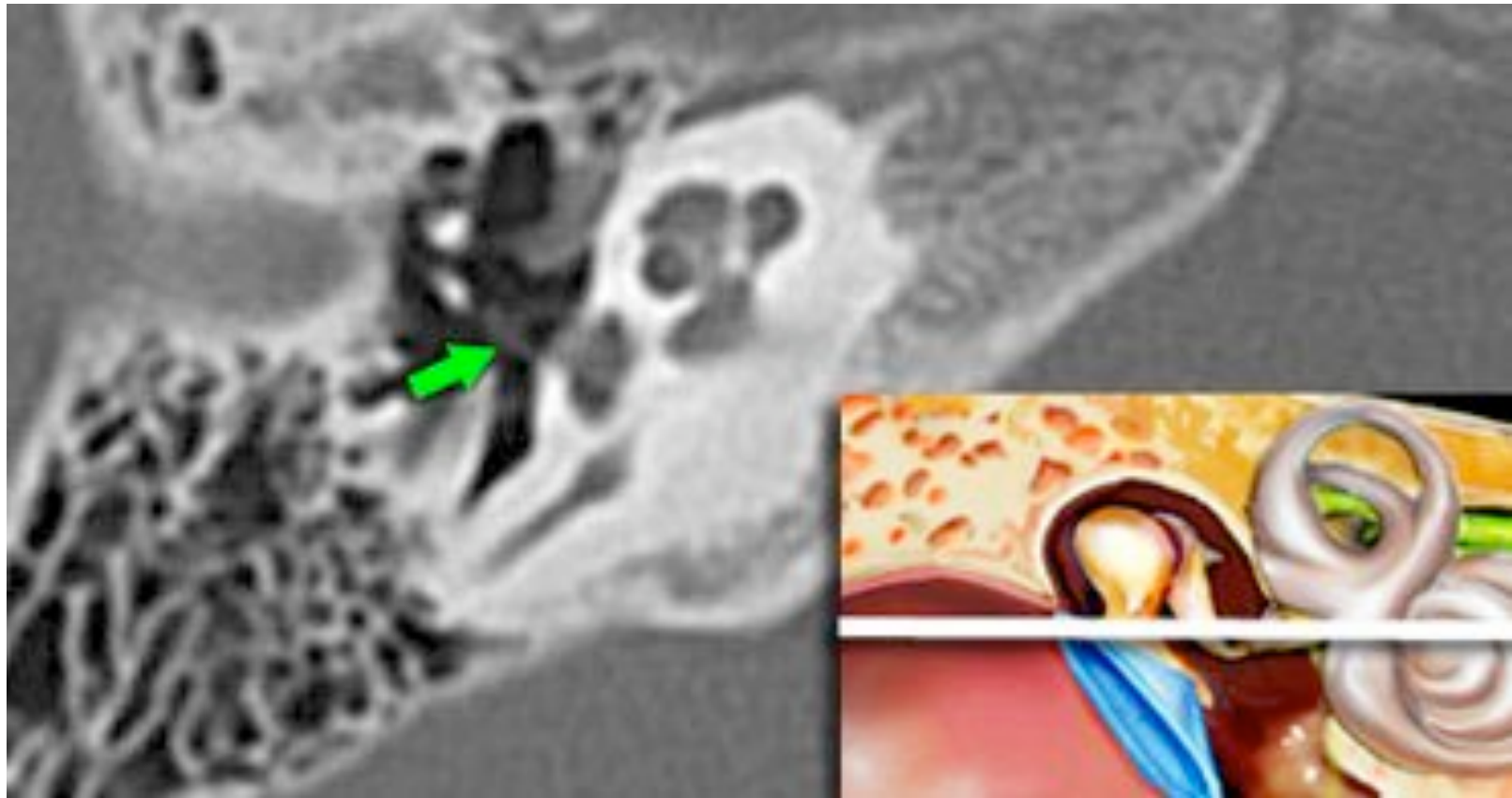


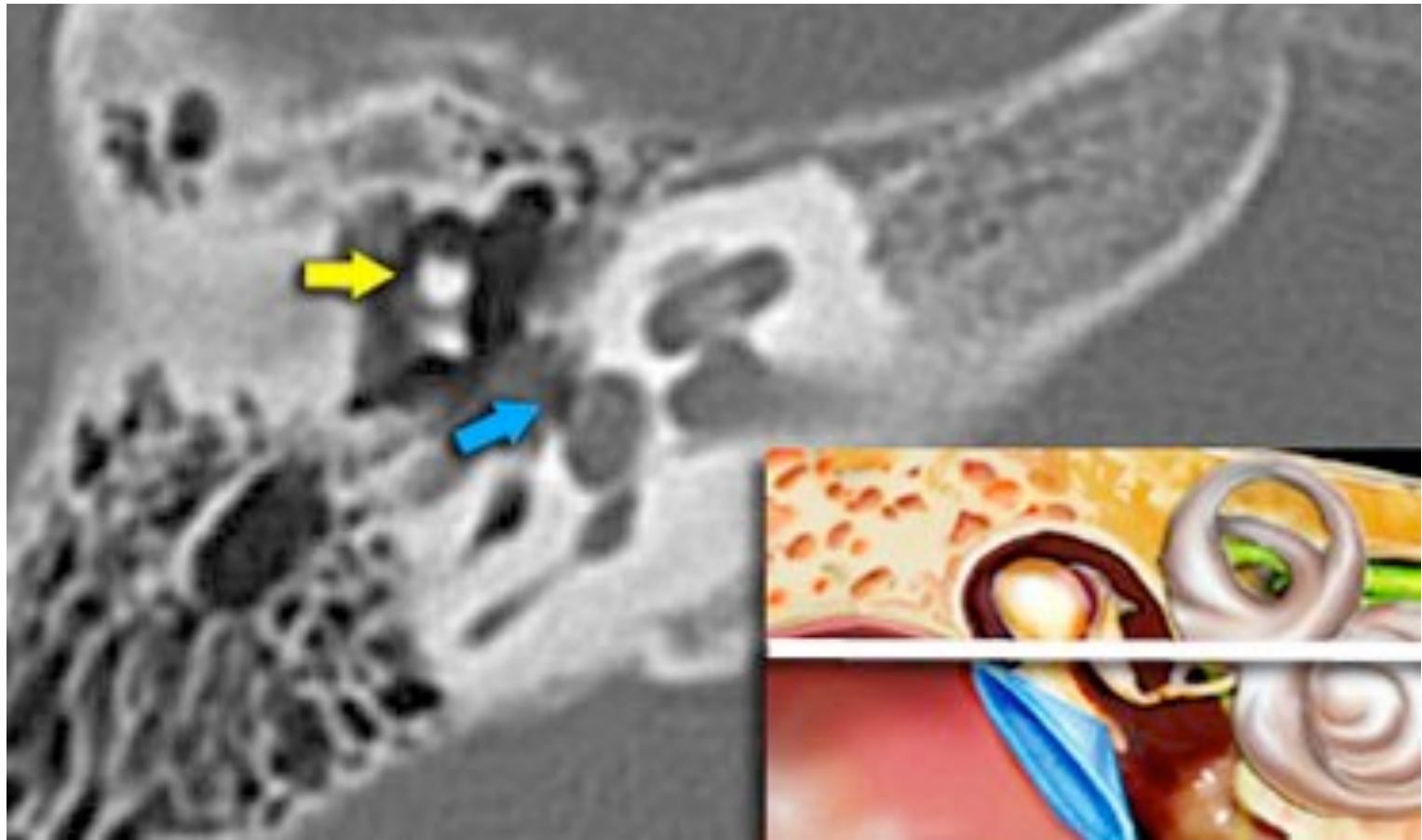


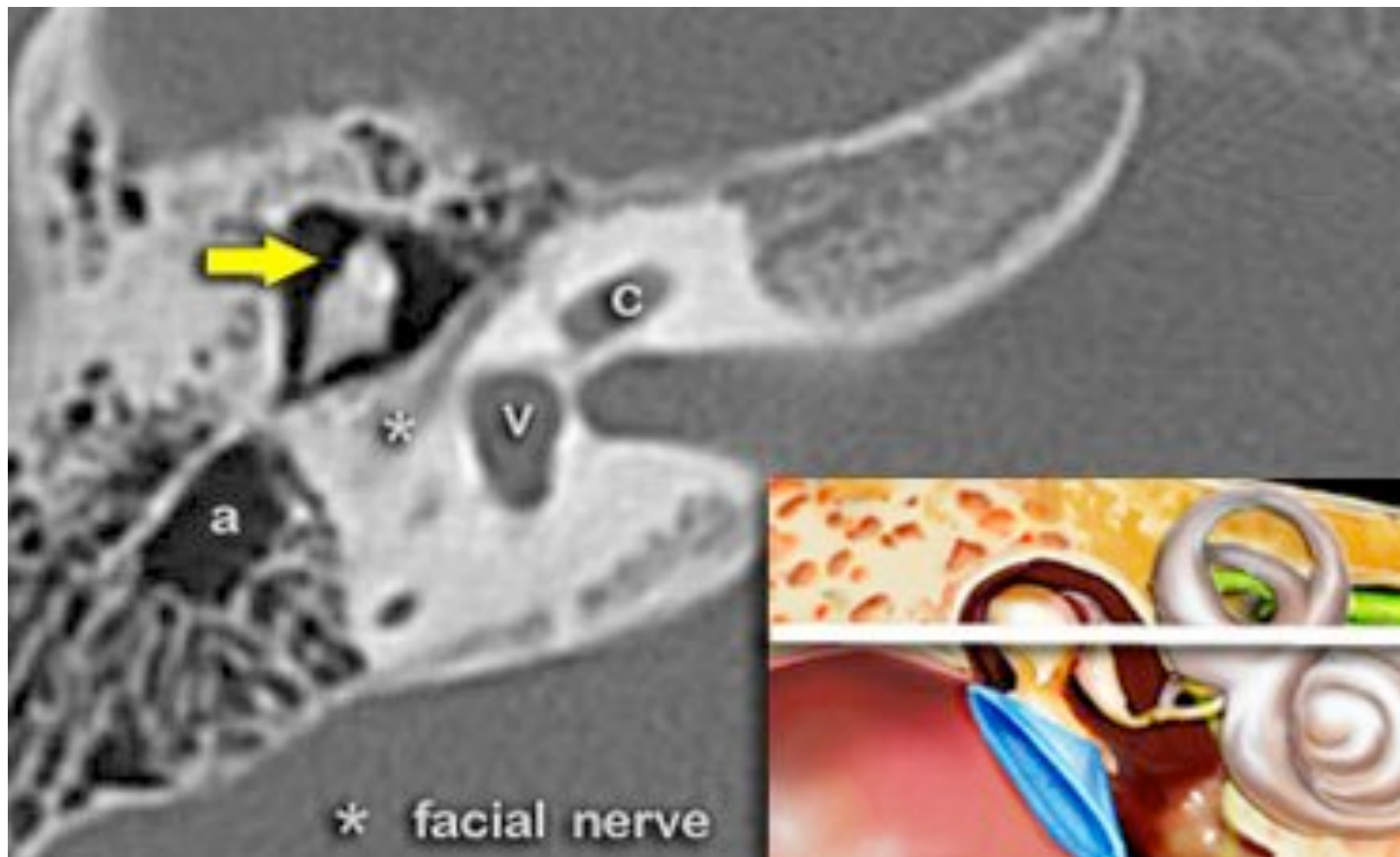


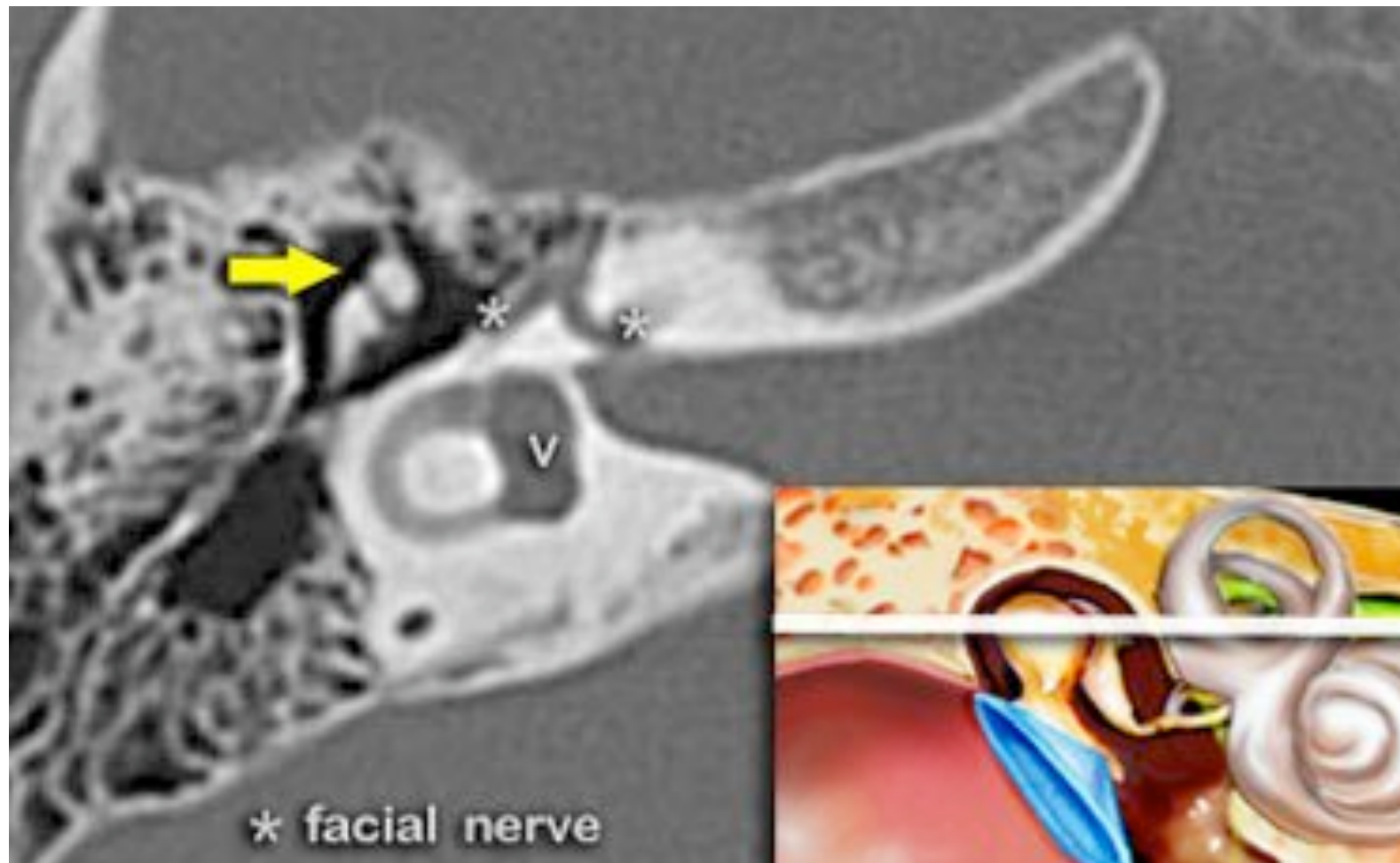


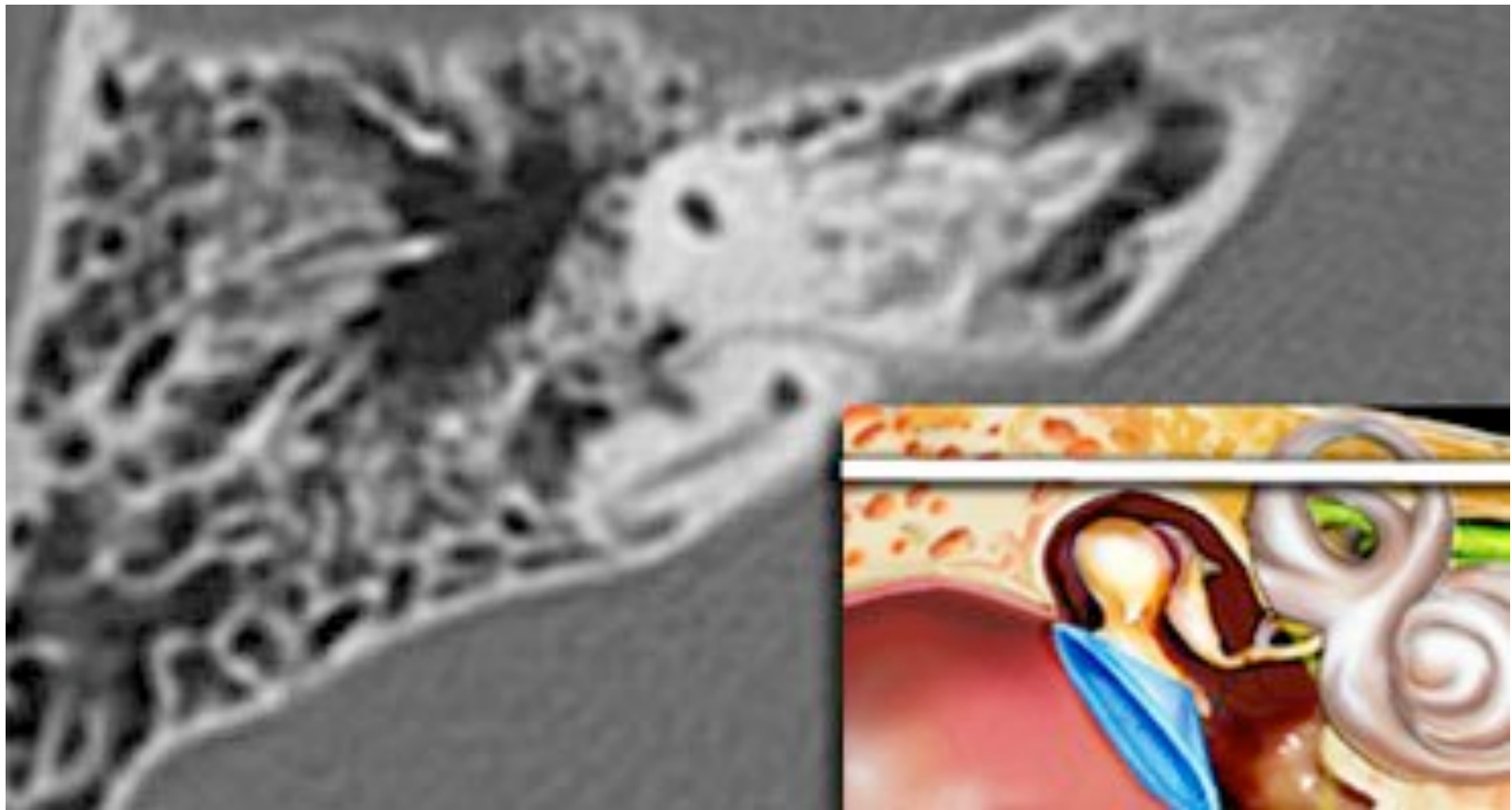


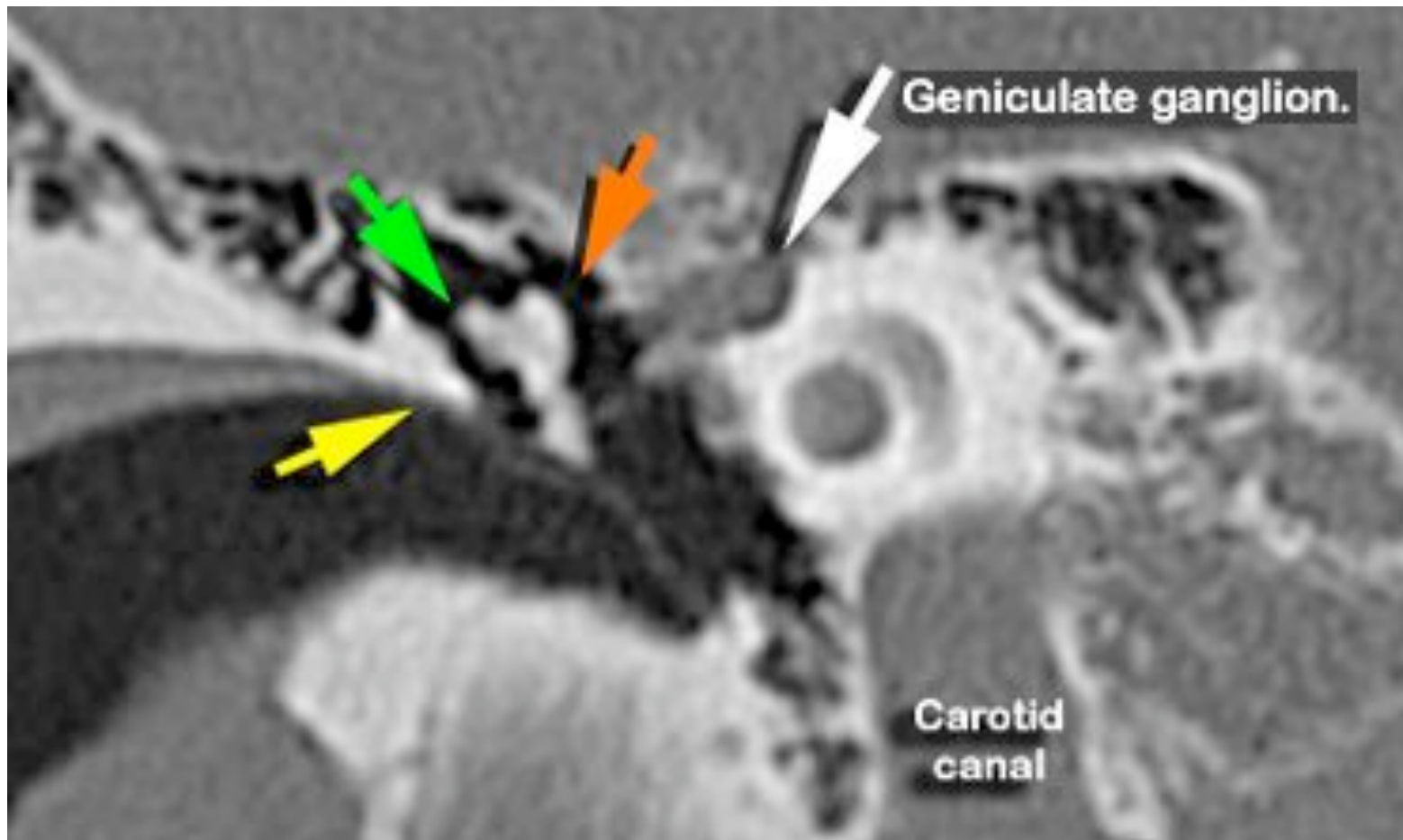


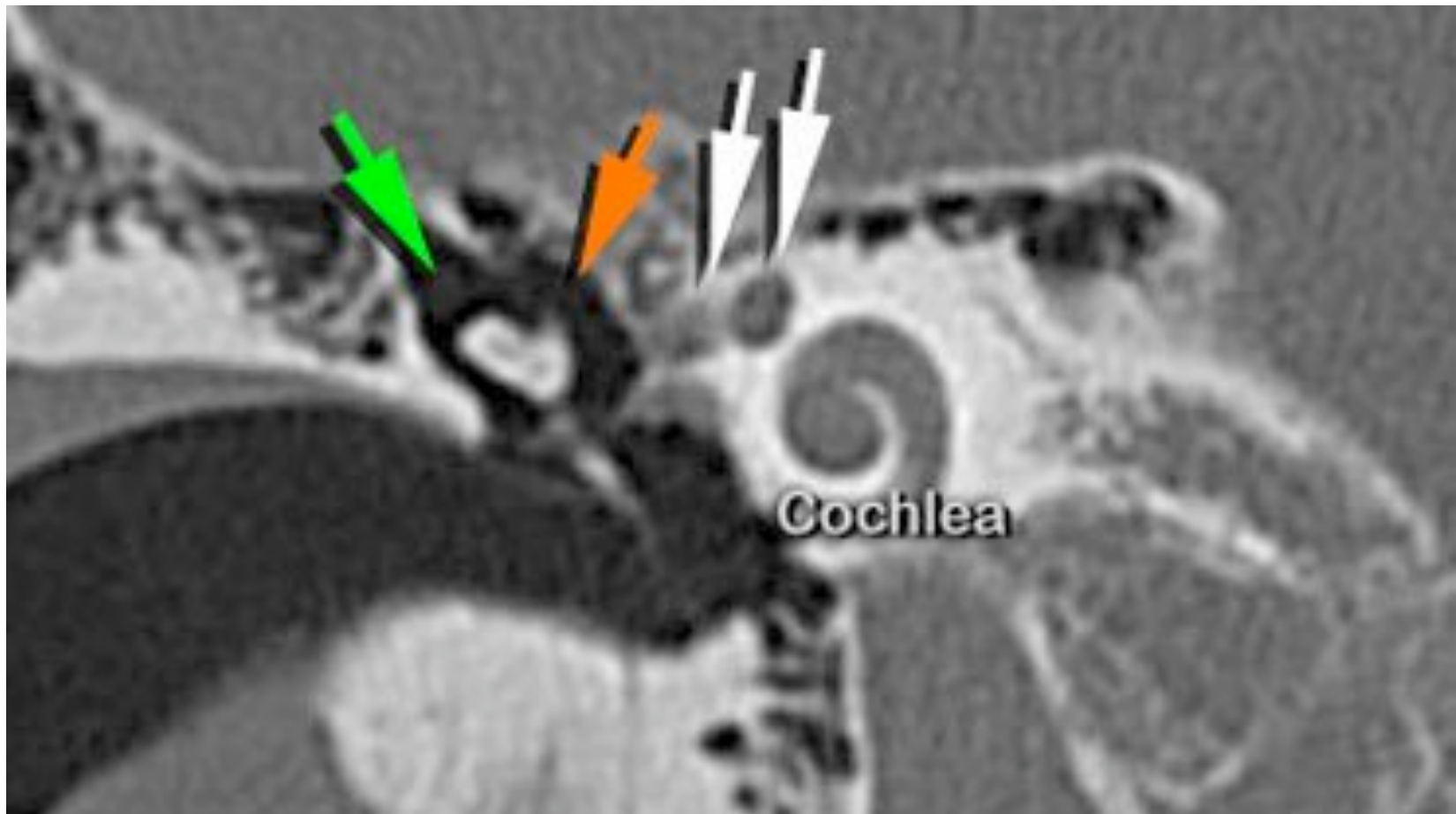


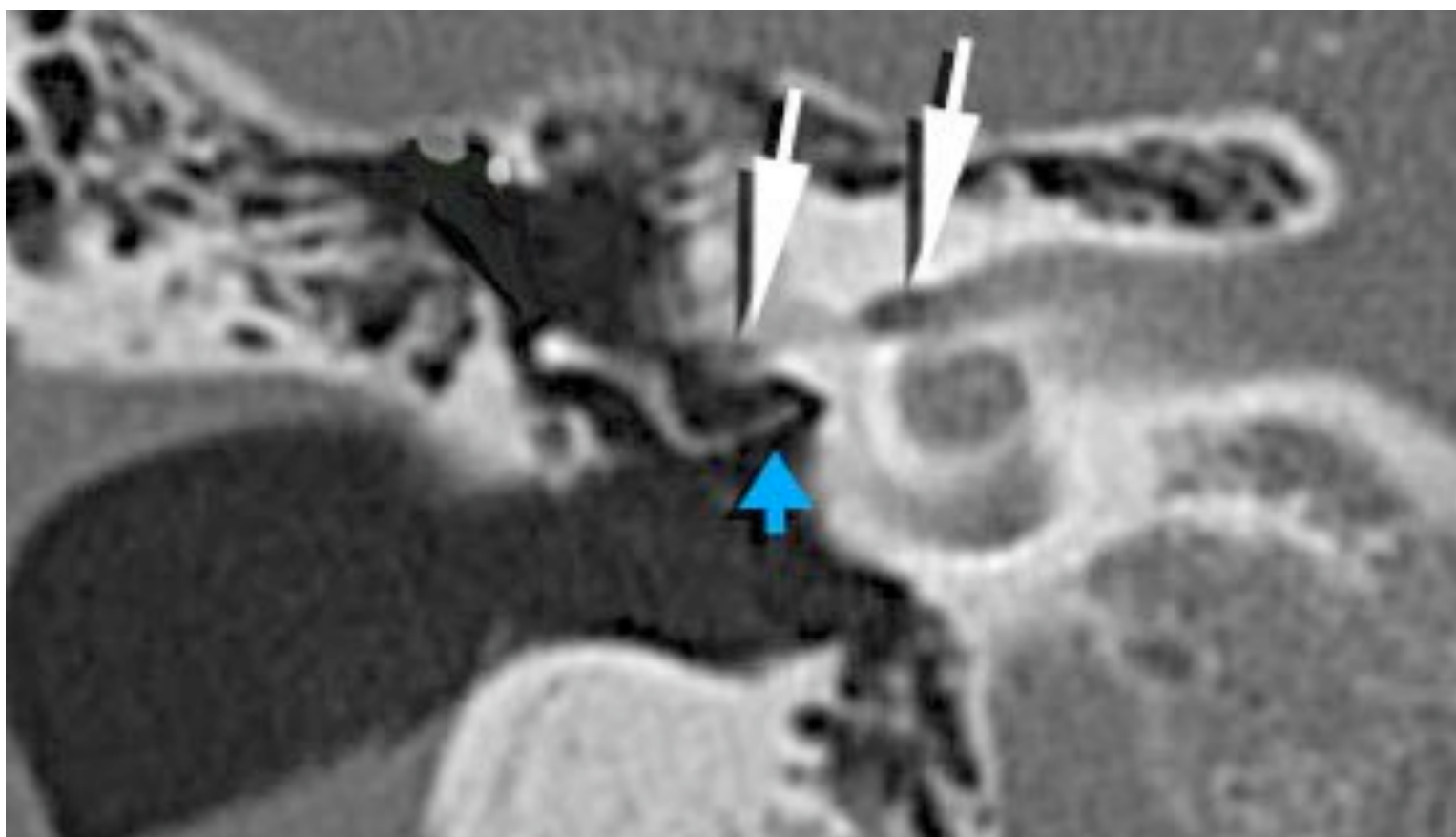


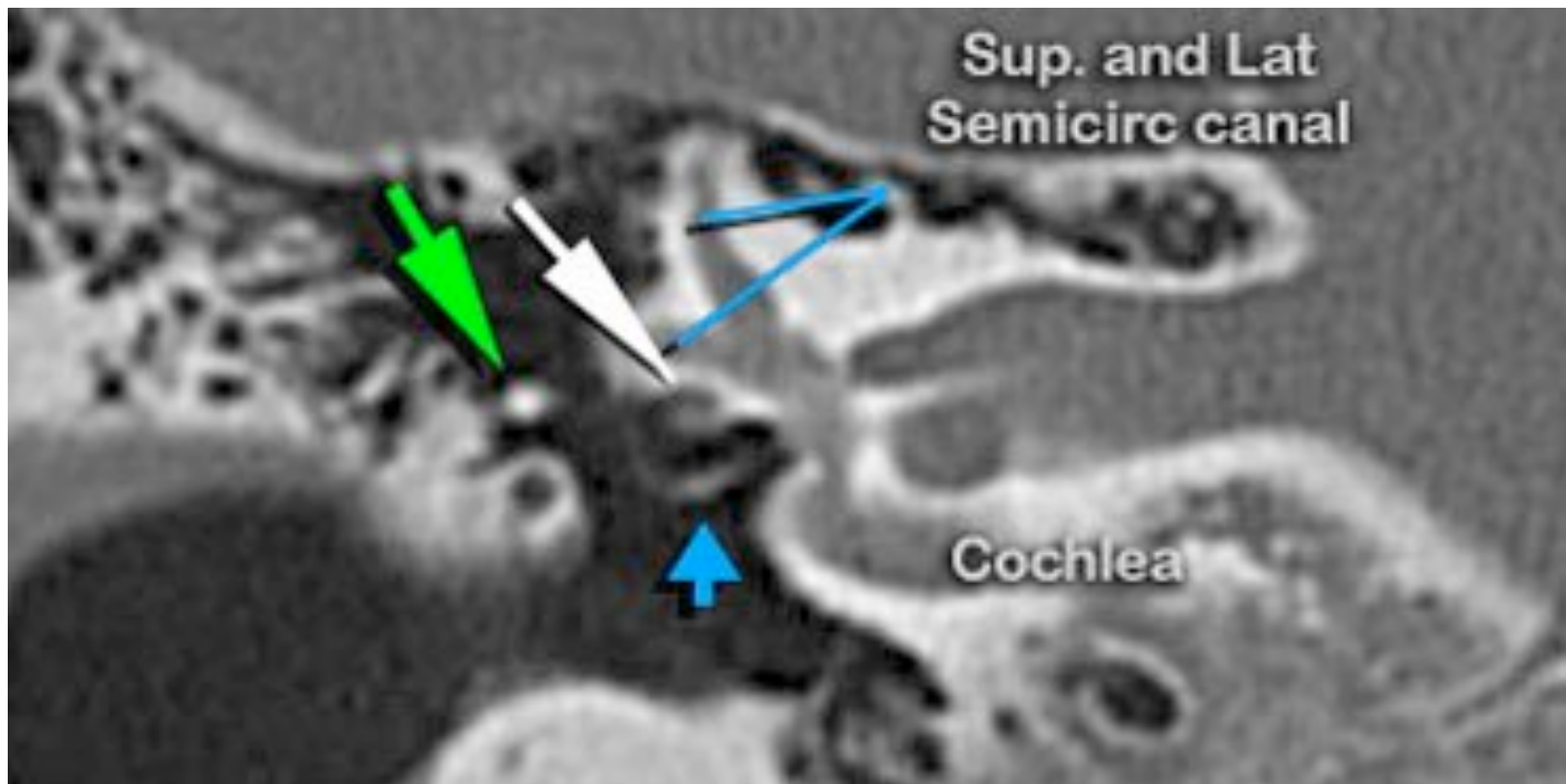


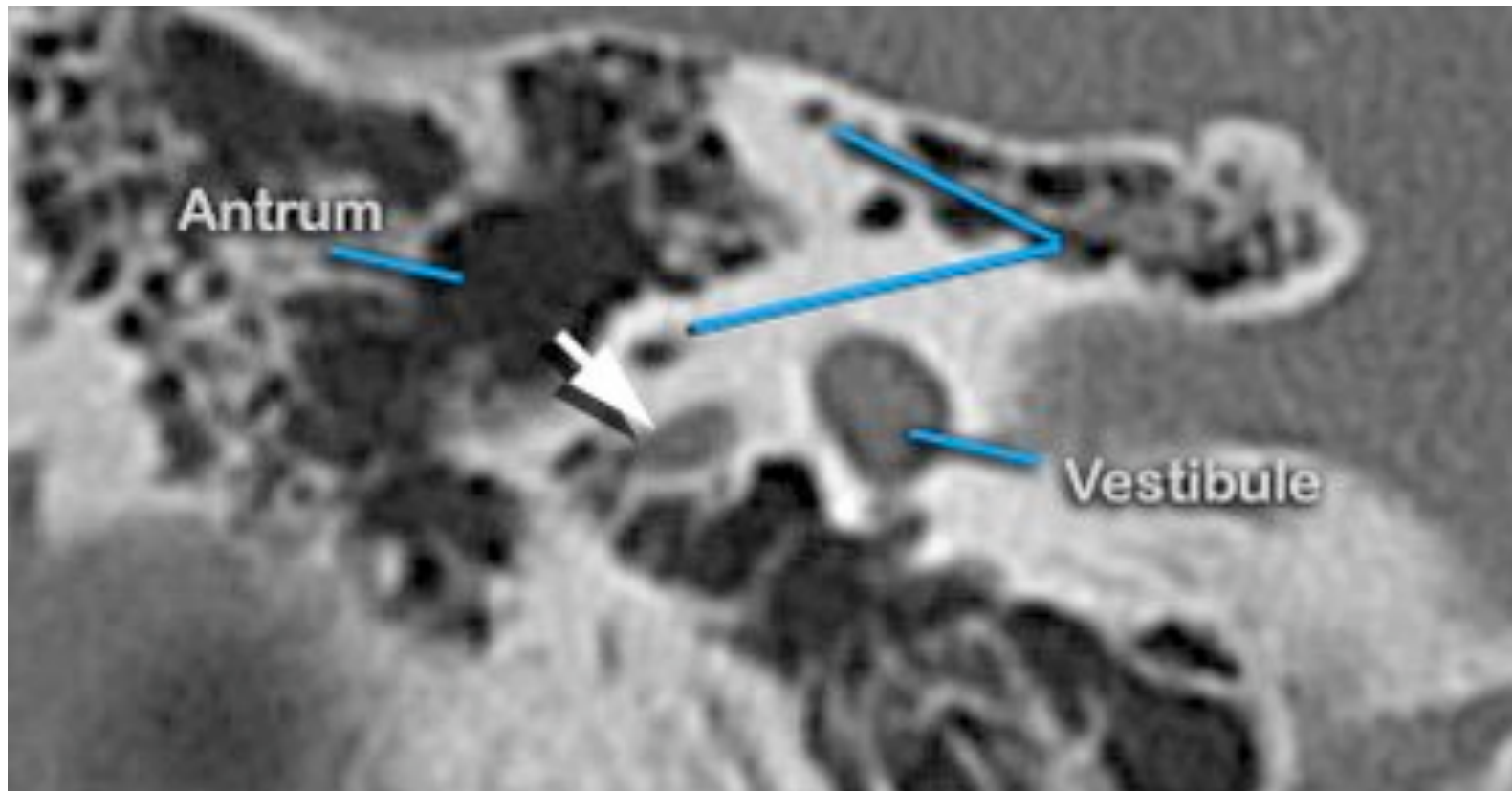




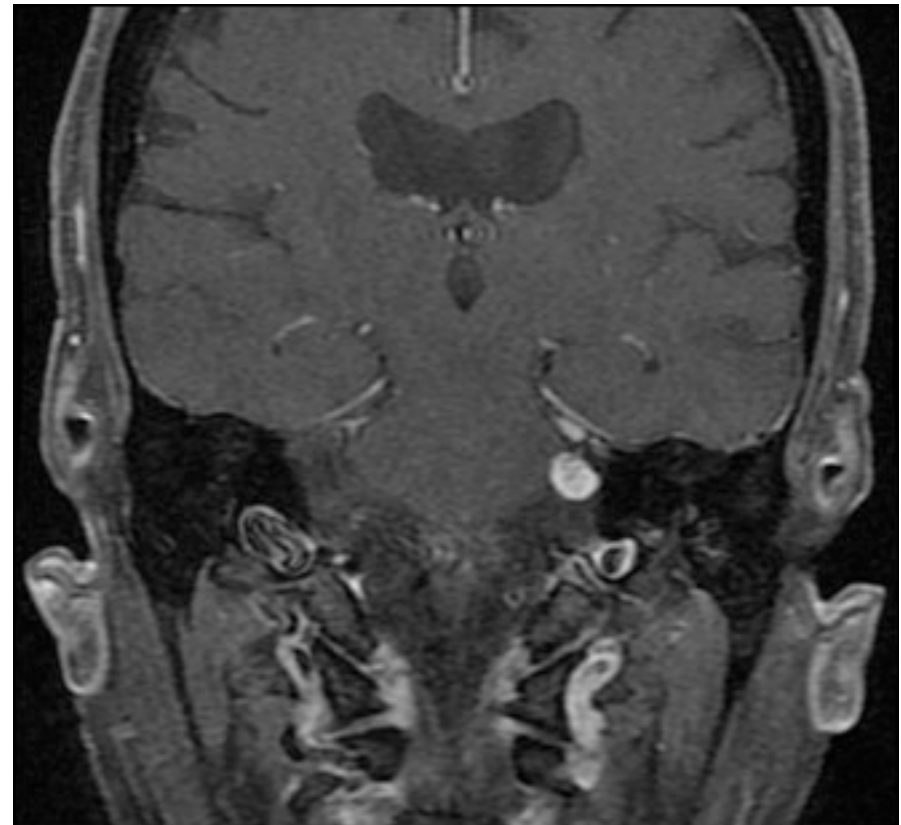
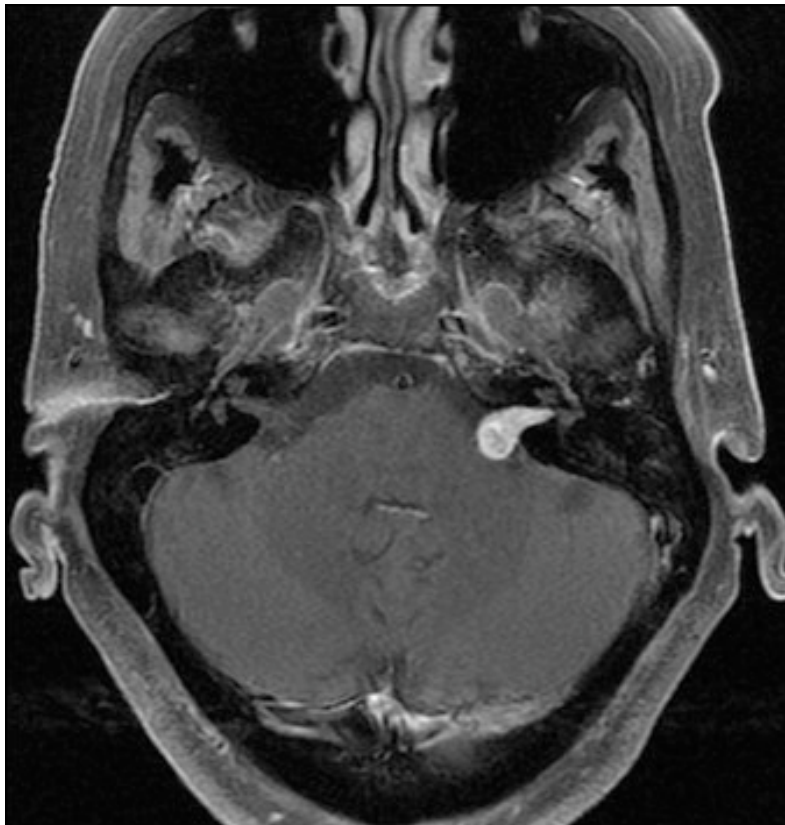








MRI is the Gold Standard



Let's play... Name that lesion!

Tumor	T1	T1-Gad	T2
Vestibular schwannoma	Iso or hypo	Oh yeah!	Hyperintense
Meningioma	Iso or hypo	Oh yeah!	Variable
Cholesteatoma	Hypointense	No	Hyperintense
Arachnoid cyst	Hypointense	No	Hyperintense
Lipoma	Hyperintense	No	Intermediate
Cholesterol granuloma	Hyperintense	No	Hyperintense

Other tests

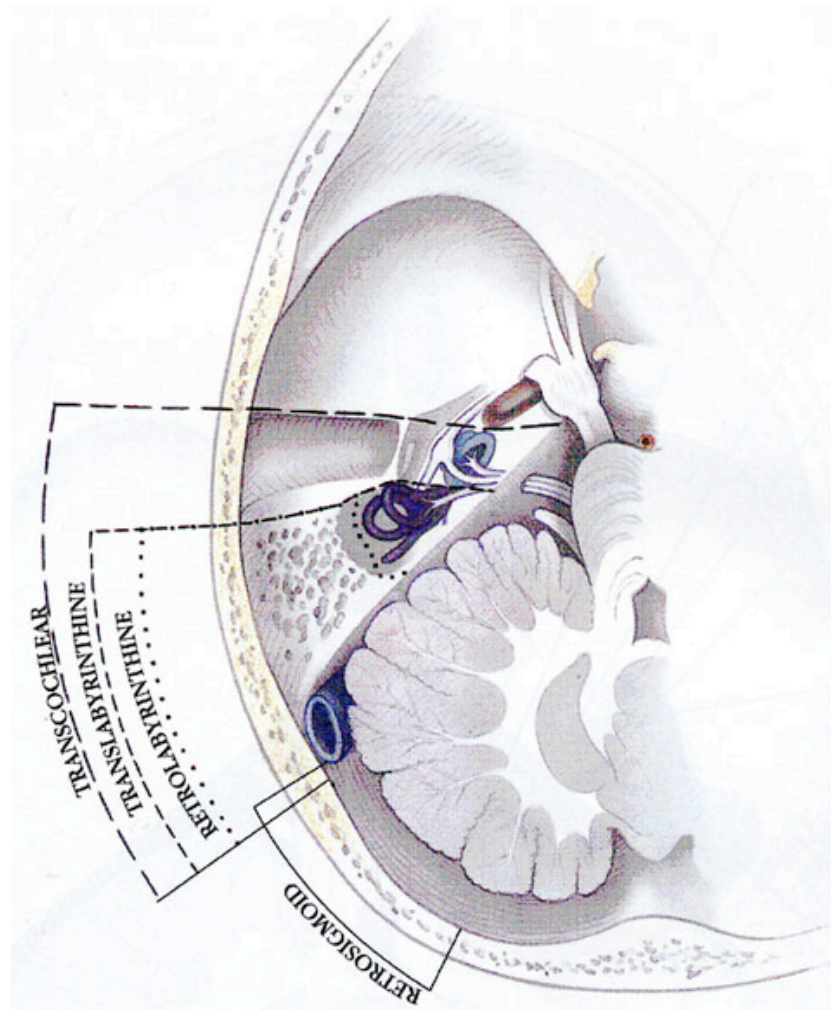
- Audiology exam
 - Pure-tone audiometry
 - Speech discrimination score (lower than predicted by PTA)
 - Rollover – as speech intensity increases, performance rises to a maximum level, then “rolls over” sharply as intensity continues to increase
 - Acoustic reflexes
- ENG
 - Reduced caloric response in problematic ear
- ABR
 - Fully or partially absent
 - Delay in latency of Wave V
 - An interaural delay of Wave V latency >0.2 ms is abnormal
 - Sensitivity and specificity of $>90\%$
 - Not great for small intracanalicular tumors

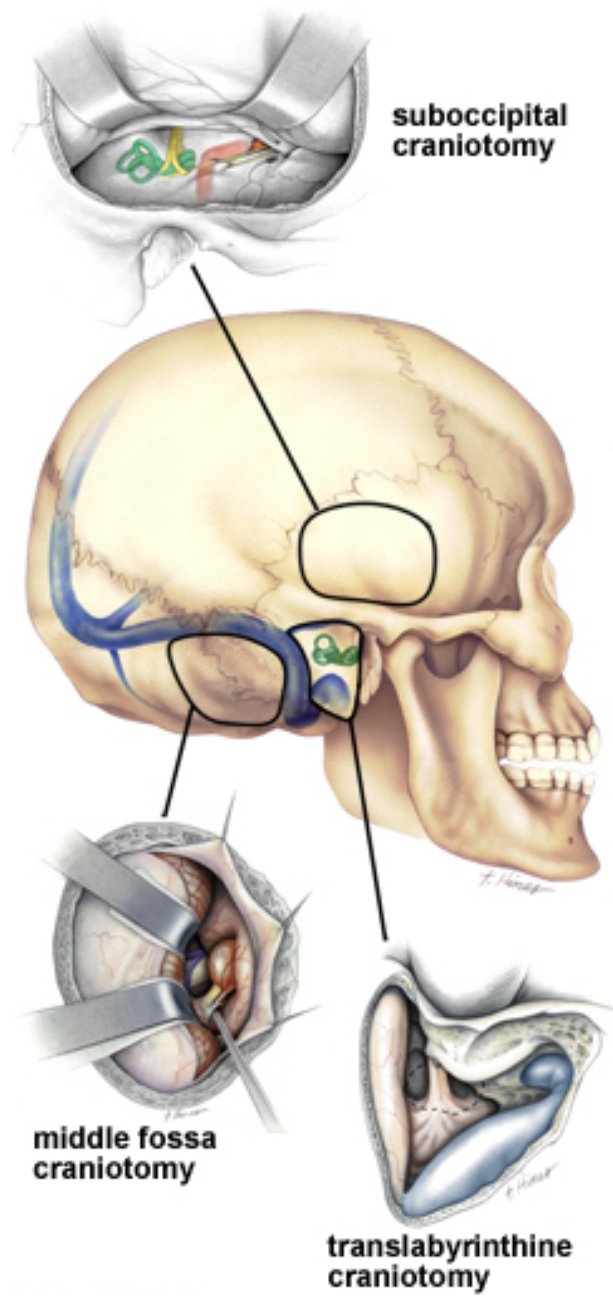
Treatment - Nonsurgical

- Observation
 - 2nd radiologic exam in 6 months, then yearly
 - If the tumor grows 2-3 mm in the first year, then they will likely need treatment
- Stereotactic radiation
 - Transient swelling in the first two years
 - Control rates of >85%
 - Hearing preservation rate decreases each year after radiation and stabilizes after 3 years in 50% of patients
 - Facial nerve dysfunction ranges from 3-50%
 - Trigeminal neuropathy in 20% of patients



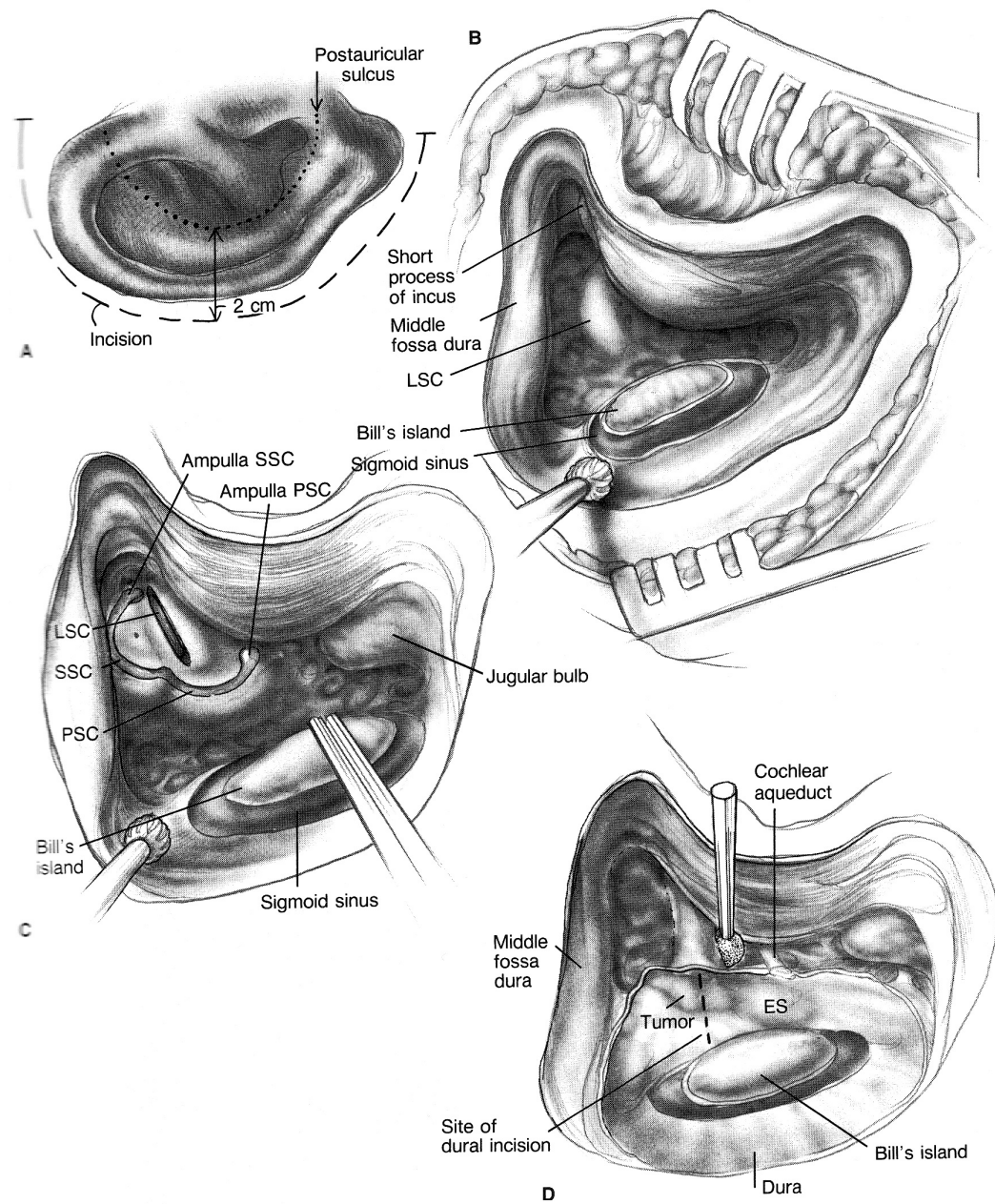
Surgical Approaches





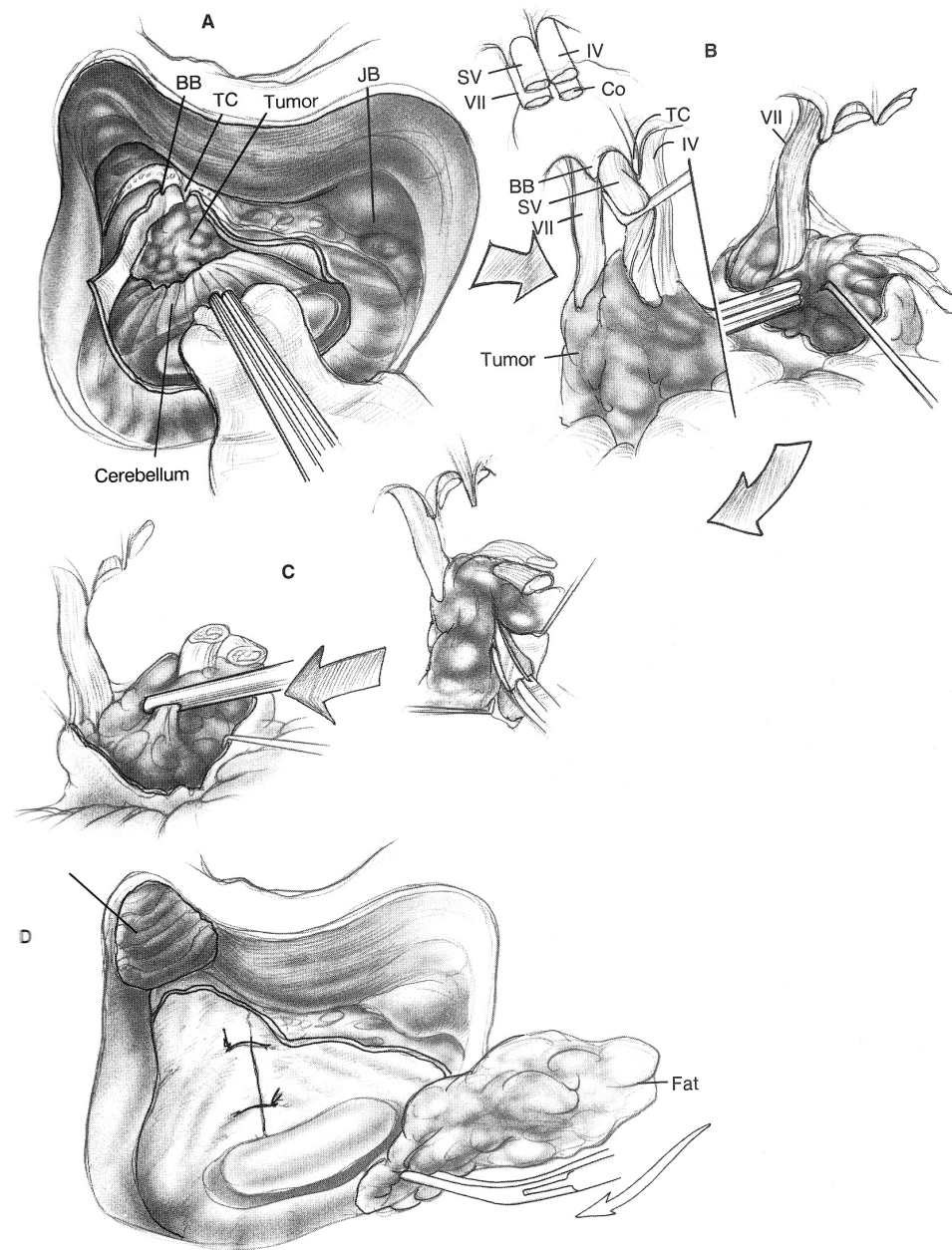
Translabyrinthine – step by step

- Postauricular incision 2 cm behind sulcus
- Complete mastoidectomy
- Sigmoid sinus is decompressed with preservation of an island of bone over the sinus (“Bill island”)
- Labyrinthectomy is then performed
- Ampulla of PSCC is identified
 - Jugular bulb is always inferior (inferior extent of dissection)
- Bone is removed around inferior aspect of IAC until the cochlear aqueduct is identified
 - CA is an important landmark for avoidance of CN IX, X, XI (anterior and inferior to CA)

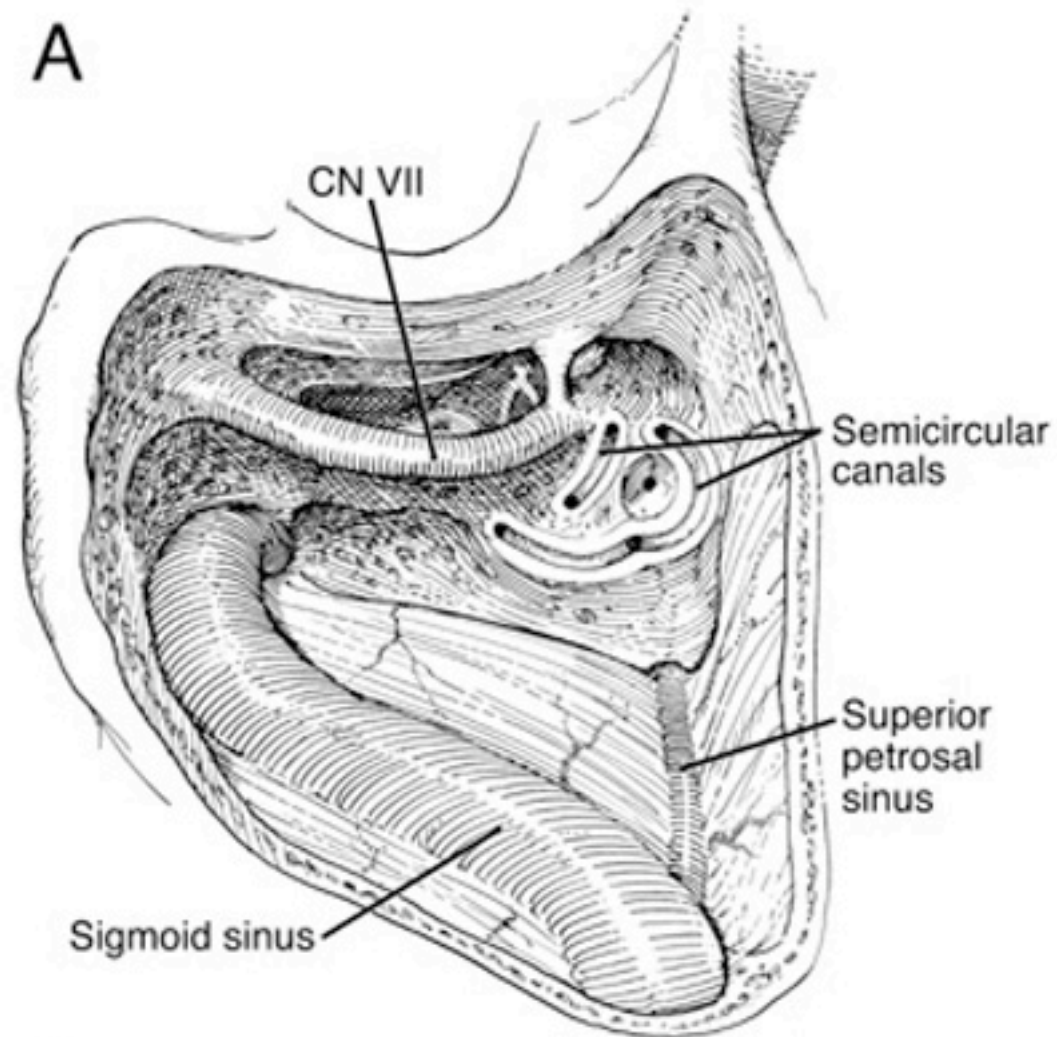


Translabrynthine – step by step

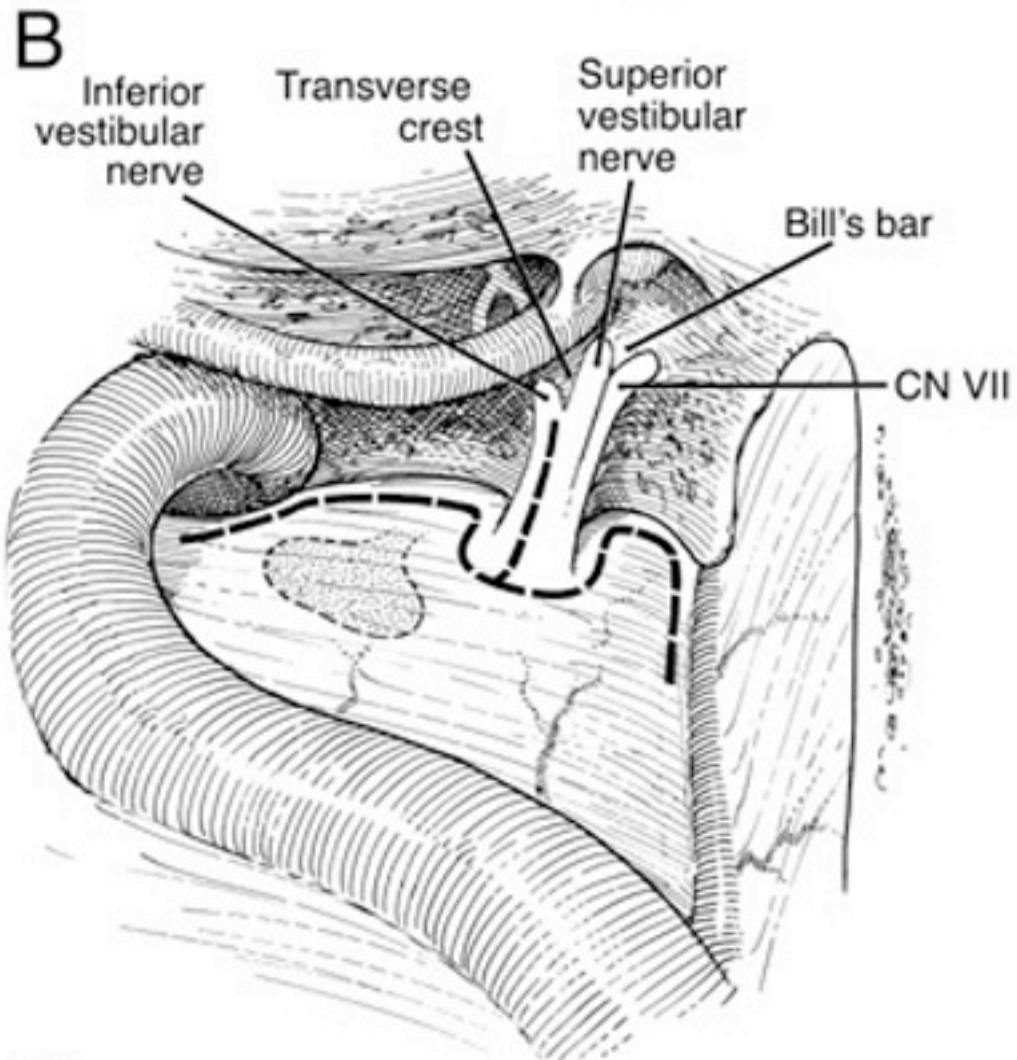
- IAC is skeletonized
- Transverse crest and Bill bar are identified
- At the conclusion of the dissection, the bone covering the middle and posterior cranial fossa dura is removed along with the bone covering the entire posterior two-thirds of the IAC
- Dura is opened
- Tumor capsule is incised
- Tumor is gutted
- Remaining tumor and capsule are separated from facial nerve



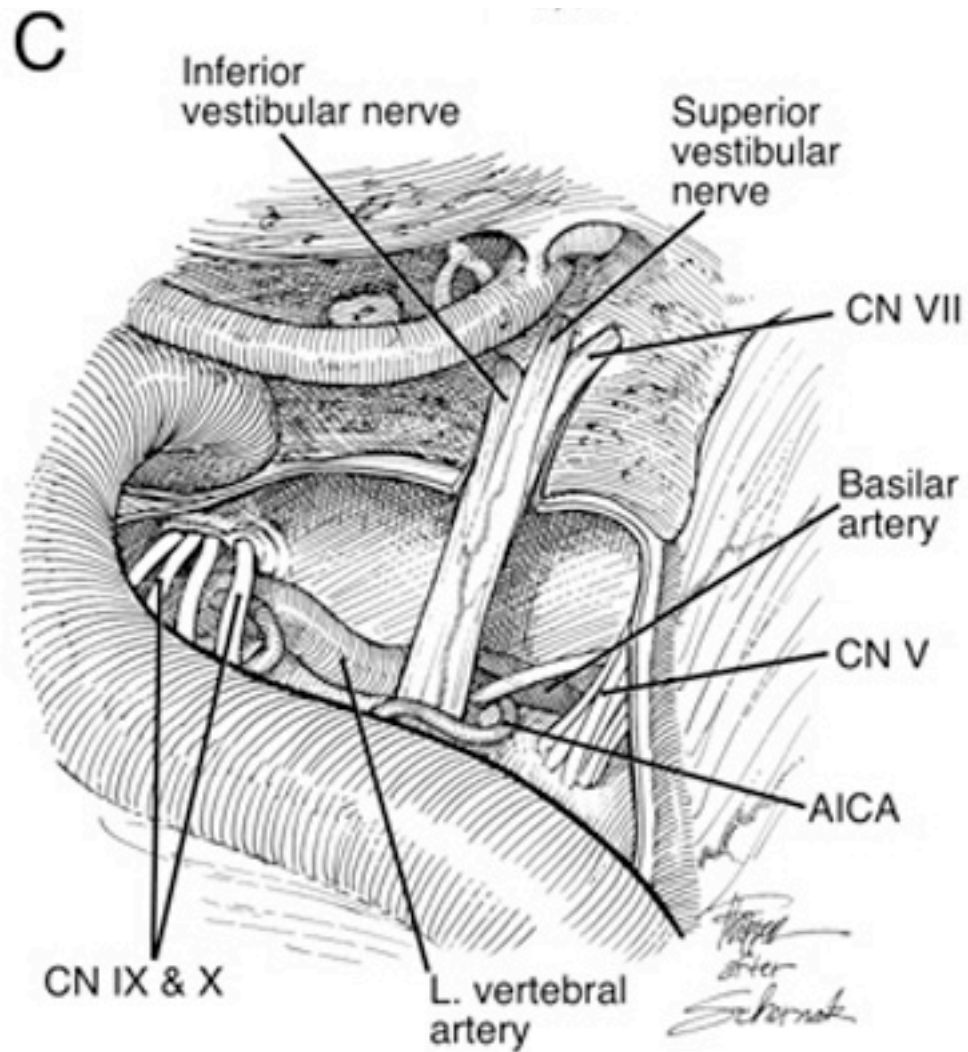
Translabyrinthine



Translabyrinthine



Translabyrinthine



Source: Neurosurg Focus © 2005 American Association of Neurological Surgeons

Translabrynthine

- Advantages
 - Ability to remove tumors of all sizes
 - Minimal brain retraction
 - Directly visualize and preserve facial nerve (97% preservation)
 - Less postop headaches
- Disadvantages
 - Complete hearing loss
 - Up to 21% CSF leak (most common complication)
 - Limited in patients with anterior sigmoid sinuses and high-riding jugular bulbs

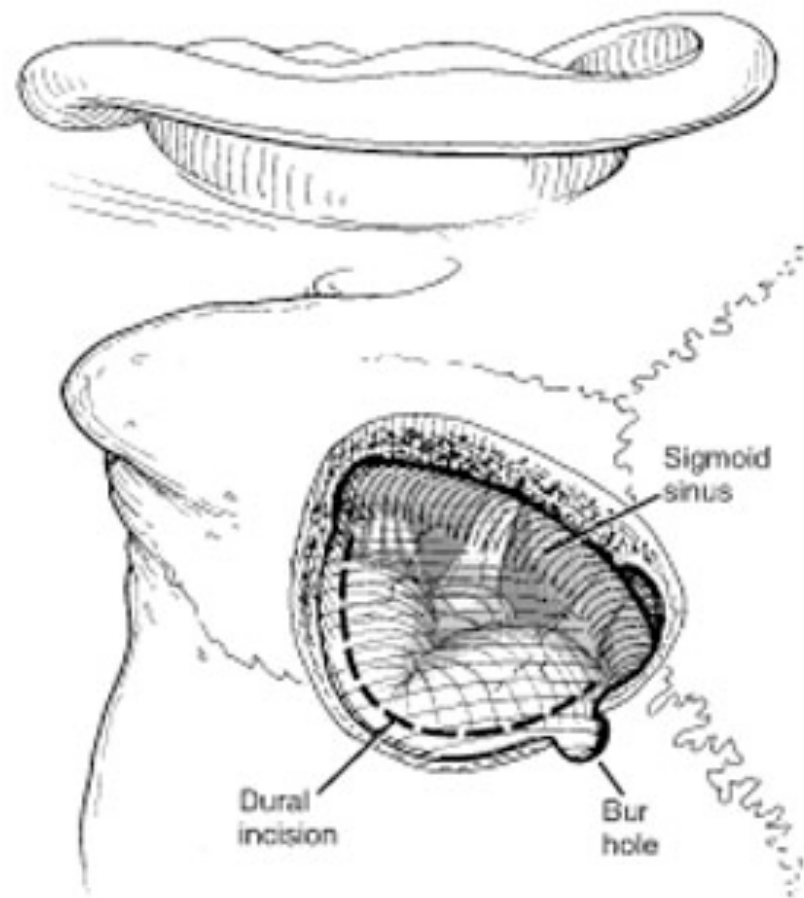
Case question

- A patient who recently underwent a translabyrinthine resection of a VS now has clear rhinorrhea. What is your management plan?
 - CSF leak
 - Mastoid pressure dressing and lumbar drain for 3-4 days
 - If leak persists, return to OR for re-oblitration of ET and blind sac closure of EAC

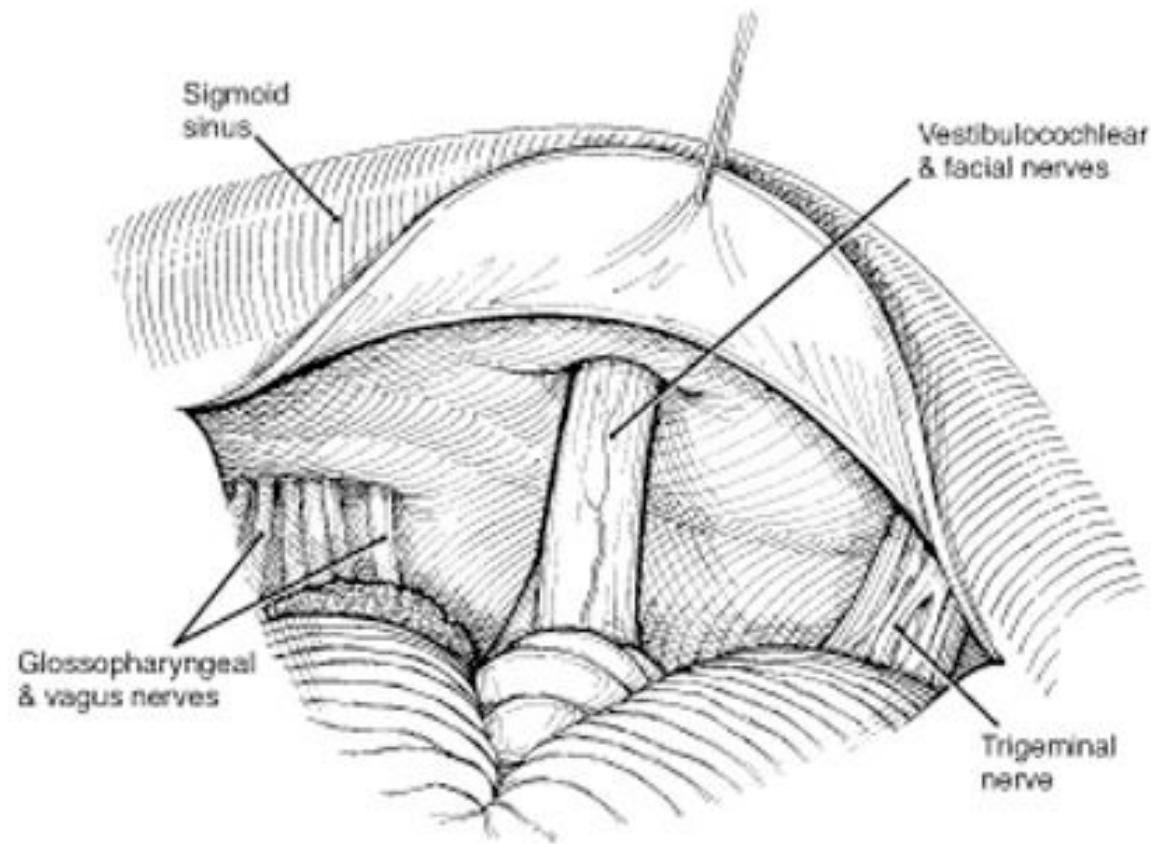
Retrosigmoid – step by step

- Incision is more posterior (3 cm behind sulcus)
- Limited mastoidectomy is performed to identify sigmoid sinus and jugular bulb
- 4 cm craniotomy is made over subocciput
- Dura is incised
- Sigmoid is reflected anteriorly and cerebellum is retracted posteriorly
- IAC is skeletonized

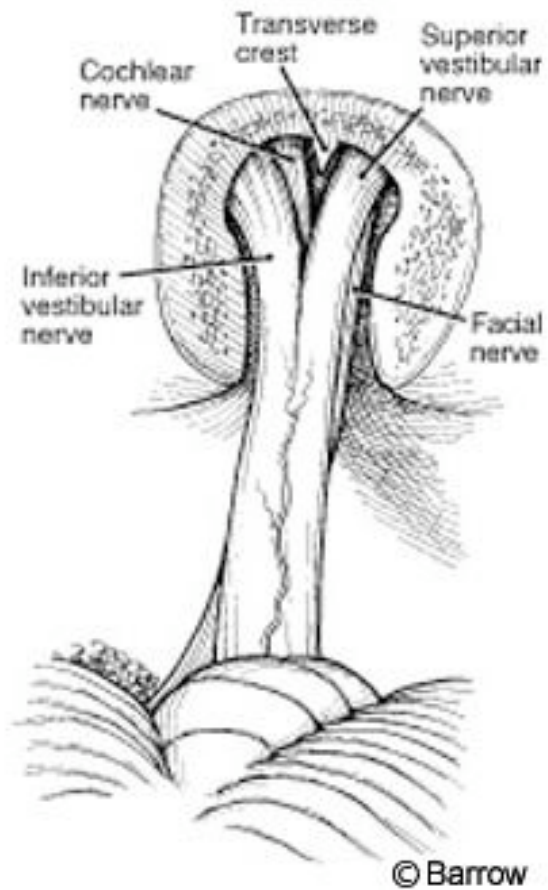
Retrosigmoid



Retrosigmoid



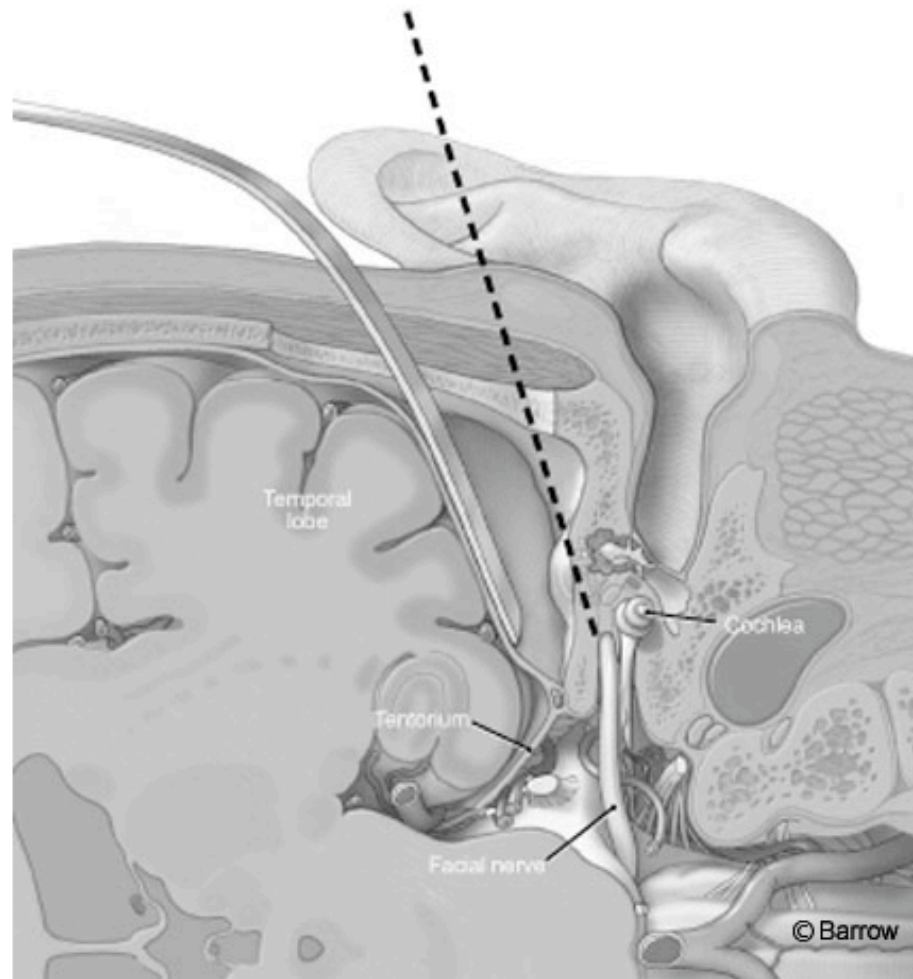
Retrosigmoid

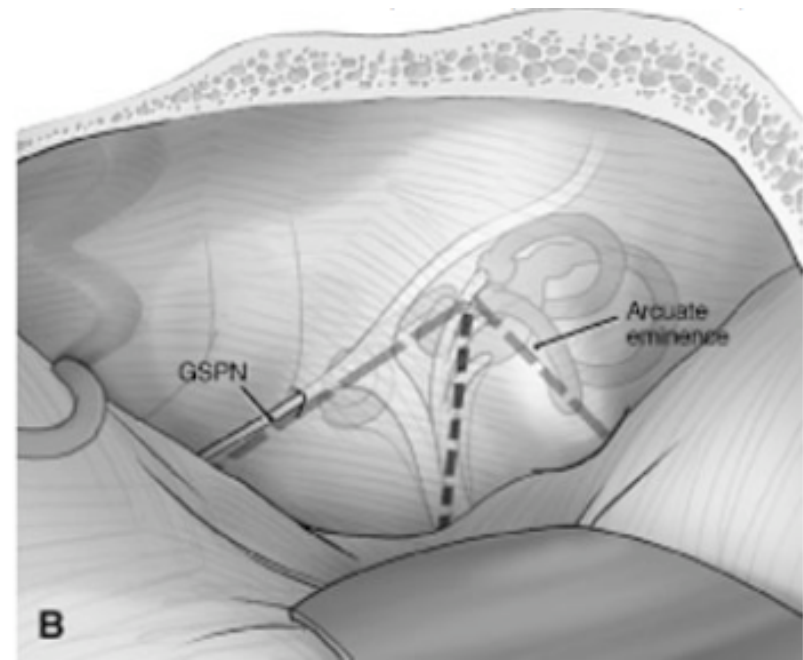
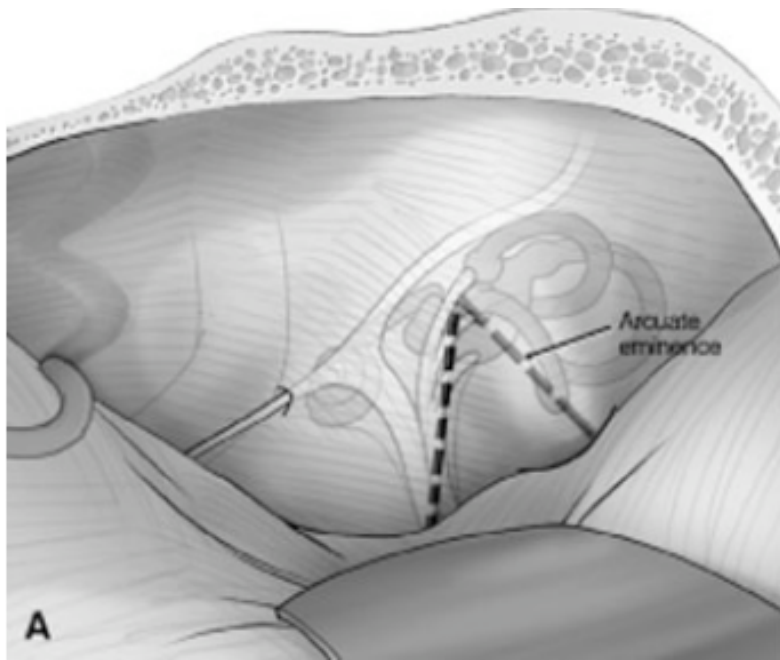


Retrosigmoid

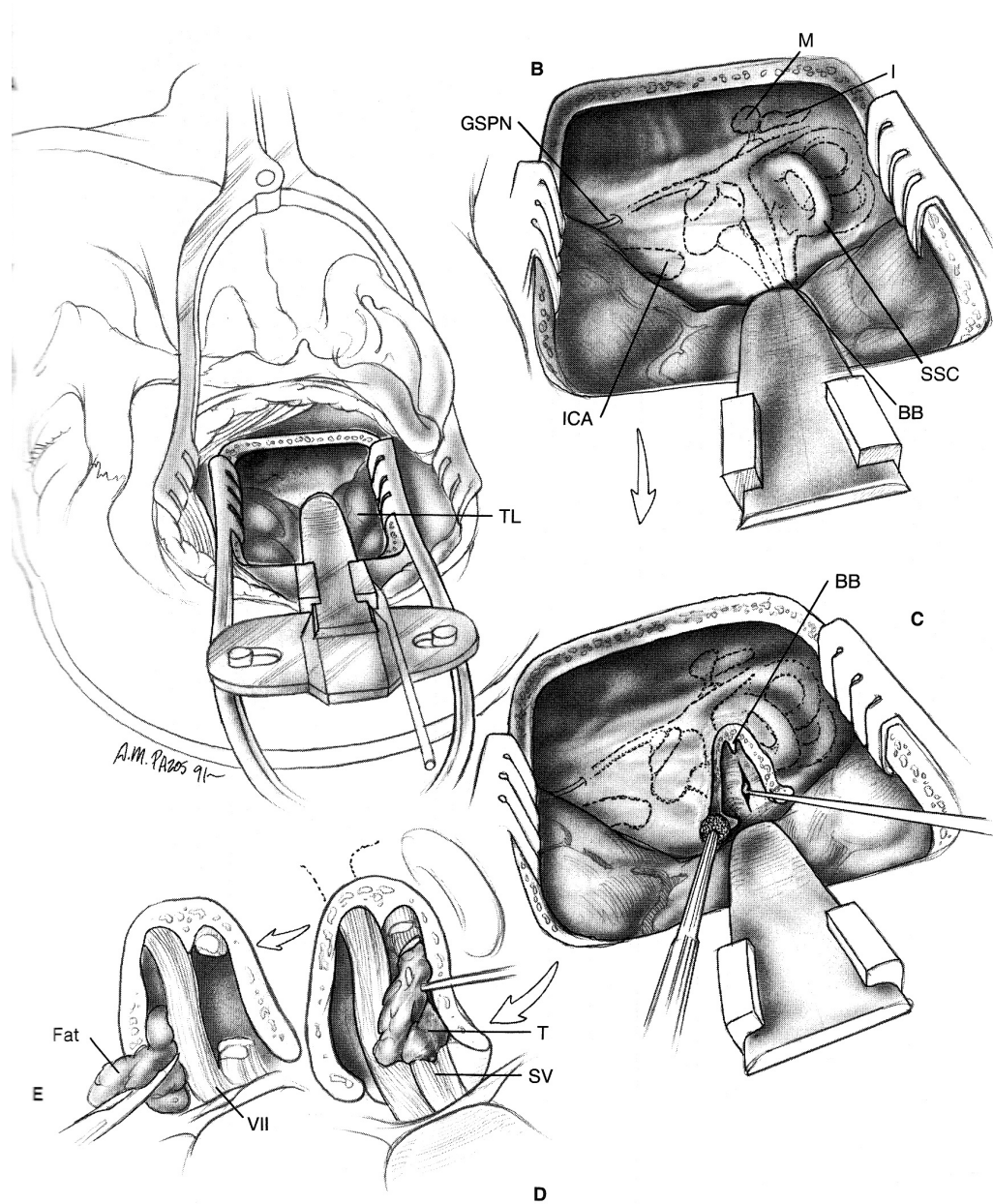
- Advantages
 - Quicker
 - Hearing preservation (50% in tumors $<2\text{cm}$)
 - Good for medial tumors
- Disadvantages
 - Persistent postop headache
 - CSF leak (7-21%)
 - Cerebellar retraction
 - In large tumors, dissection precedes identification of facial nerve

Middle Fossa





1. Garcia-Ibanez – arcuate eminence and the GSPN form a 120 degree angle; IAC is located where the two lines bisect
2. Fisch – a line 60 degrees anterior to the arcuate eminence shows the anterior lip of the IAC
3. House – once the GSPN is identified, drill medially towards IAC



Middle Fossa

- Advantages
 - Good for small intracanalicular tumors (lateral)
 - Hearing preservation (50-75%)
 - Minimal risk of CSF leak
- Disadvantages
 - Temporal lobe retraction
 - Increased risk of facial nerve paresis

Postop Complications

- Bleeding
- Stroke
- SIADH
- CSF leak
- Meningitis