

Semicircular Canal Dehiscence Syndrome (SCDS): What to look for and why?

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Lakeland Regional **Health**[®]

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SCDS Epidemiology

- Well known condition first described in 1998
- Cadaveric and in vivo studies demonstrate dehiscence in 0.5% and near-dehiscence in 1.4%
- Superior variation is more common, but Posterior variation is clinically similar
 - Evaluation of both is essential as they can occur simultaneously or independently

SCDS Clinical Presentation

- Vestibular dysfunction:
 - Phenomena of Tullio (vertigo and/or nystagmus induced by loud noise)
 - Hennebert (vertigo and/or nystagmus induced by pressure in the external auditory canal)
 - Disequilibrium
 - Oscillopsia
- Auditory dysfunction:
 - Low-frequency hearing loss
 - Aural fullness
 - Tinnitus
 - Autophony, bone conduction hyperacusis (hearing eyeballs move, chewing, belching, hearing own footsteps)

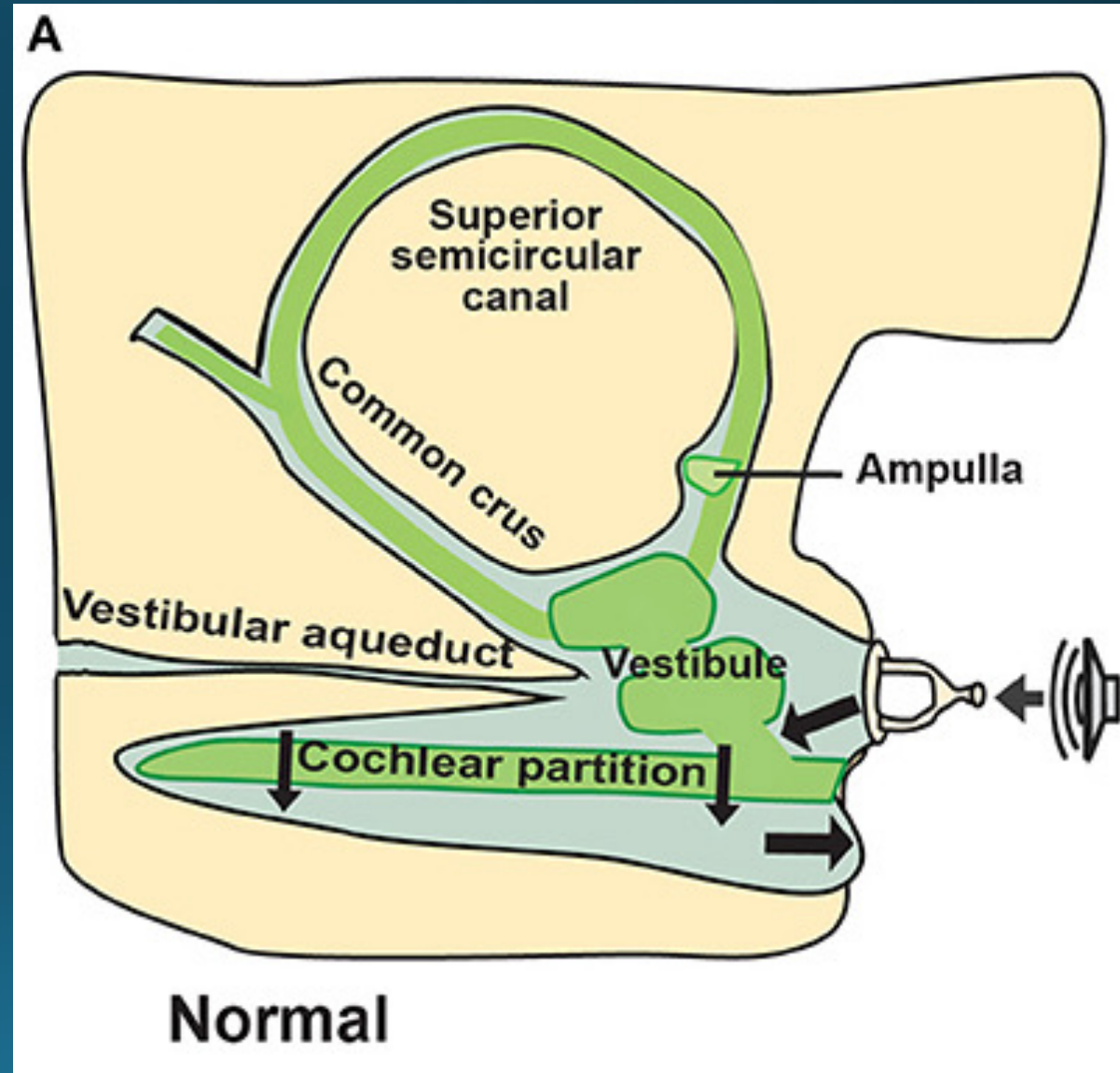
SCDS Pathophysiology

- Complete loss of bone structure between semicircular canal and intracranial fossa
- Semicircular canal dehisces to:
 - Superior plane (most common)
 - Posterior plane (less common)
- Forming a pathological “third window” to the labyrinth
- Shunting energy away from cochlea (hearing loss) and into labyrinth (vestibular symptoms)

- In air conduction sound:

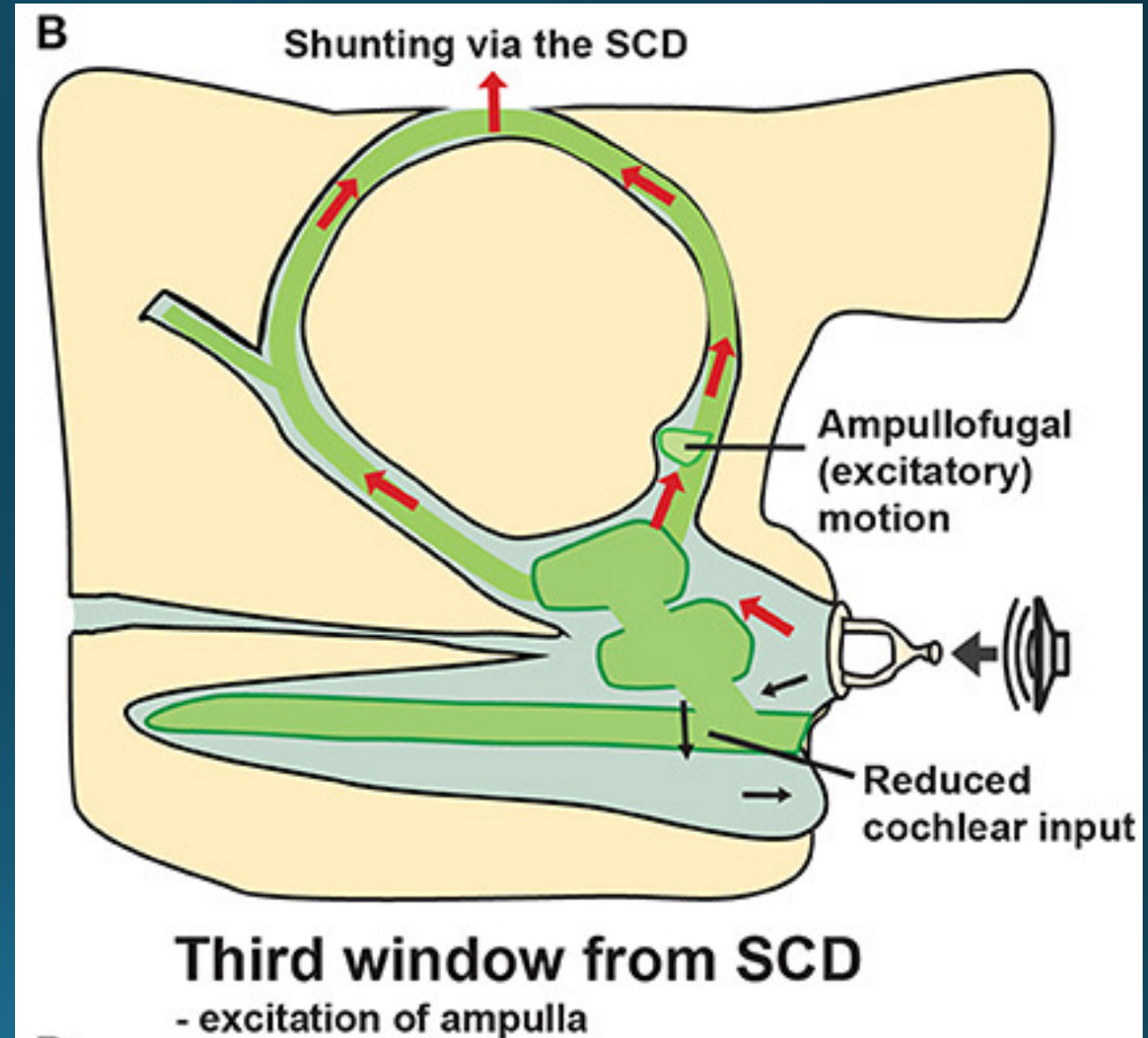
Normal hearing: sound wave > ossicles stimulate oval window > dissipated through round window (below ossicles on picture)

Normal anatomy allows volume velocity across the cochlear partition from the oval to the round window (two windows)



- Air conducted sound: Stimulation from the stapes is shunted toward the dehiscence (third window), away from the cochlea

Resulting in increased air-conduction thresholds (hearing loss at low frequencies) and vestibular symptoms from overstimulation of the labyrinth



SCDS Pathophysiology

- 2 main types: Superior and Posterior dehiscence
- Superior: communication between semicircular canal with the middle cranial fossa and/or superior petrosal sinus
- Posterior: communication between semicircular canal with the posterior cranial fossa and/or jugular bulb

SCDS Diagnosis

Clinical presentation

Diagnostic Imaging

Supportive evidence: VEMP, audiometry

SCDS: Imaging

CT

- Sensitivity: 100%
- Specificity: 95%
- Positive Predictive Value: 93%
- Negative Predictive Value: 100%

MRI

- Sensitivity: 100%
- Specificity: 99.1%
- Positive Predictive Value: 33.3%
- Negative Predictive Value: 100%

SCDS: Imaging Considerations

- For MRI, there is preference for T2-weighted acquisition (FIESTA, b-FFE, others).
 - Enhances contrast between fluid and bone
- MRI findings:
 - Normal bright signal within semicircular canal is lost as it moves out of the semicircular canal into the CSF
 - Absence of normal low-signal bone margins between semicircular canal and the cerebellopontine angle cistern (posterior SCDS) or cranial fossa

SCDS: Imaging Considerations

- For CT, there is preference for:
 - Section thickness of 0.625mm is optimal
 - Lower collimation increases PPV
 - Poschl and Stenvers slice views increases PPV
- CT findings: absence of high attenuation bone coverage between the semicircular canal and
 - Superior SCDS: middle cranial fossa and/or superior petrosal sinus
 - Posterior SCDS: posterior cranial fossa and/or jugular bulb

Specific Imaging Variations

- Beyond Axial, Coronal, and Sagittal:
 - Stenver: perpendicular to Superior semicircular canal
 - Poschl: parallel to Superior semicircular canal
- Utilizing these views increases PPV

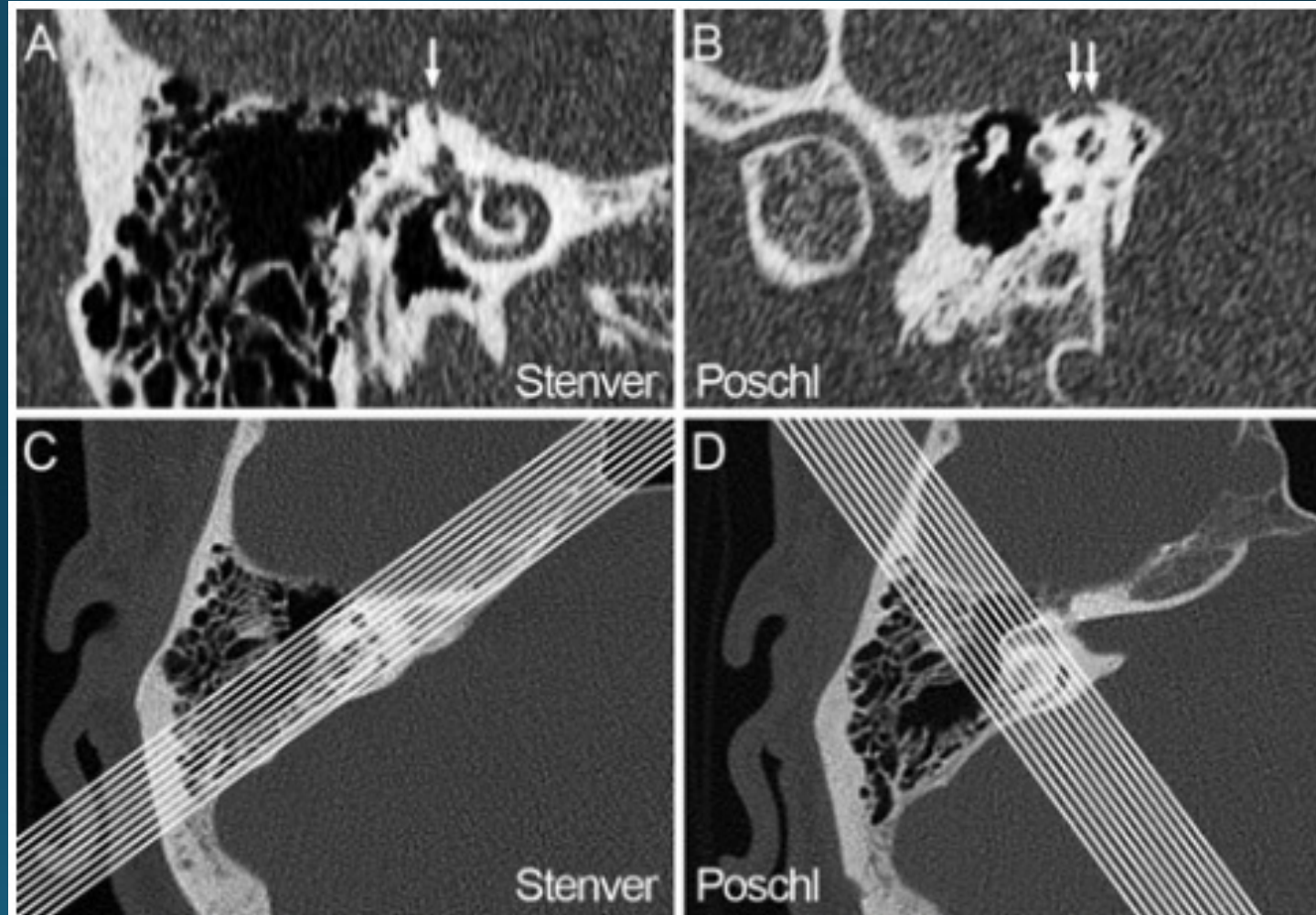
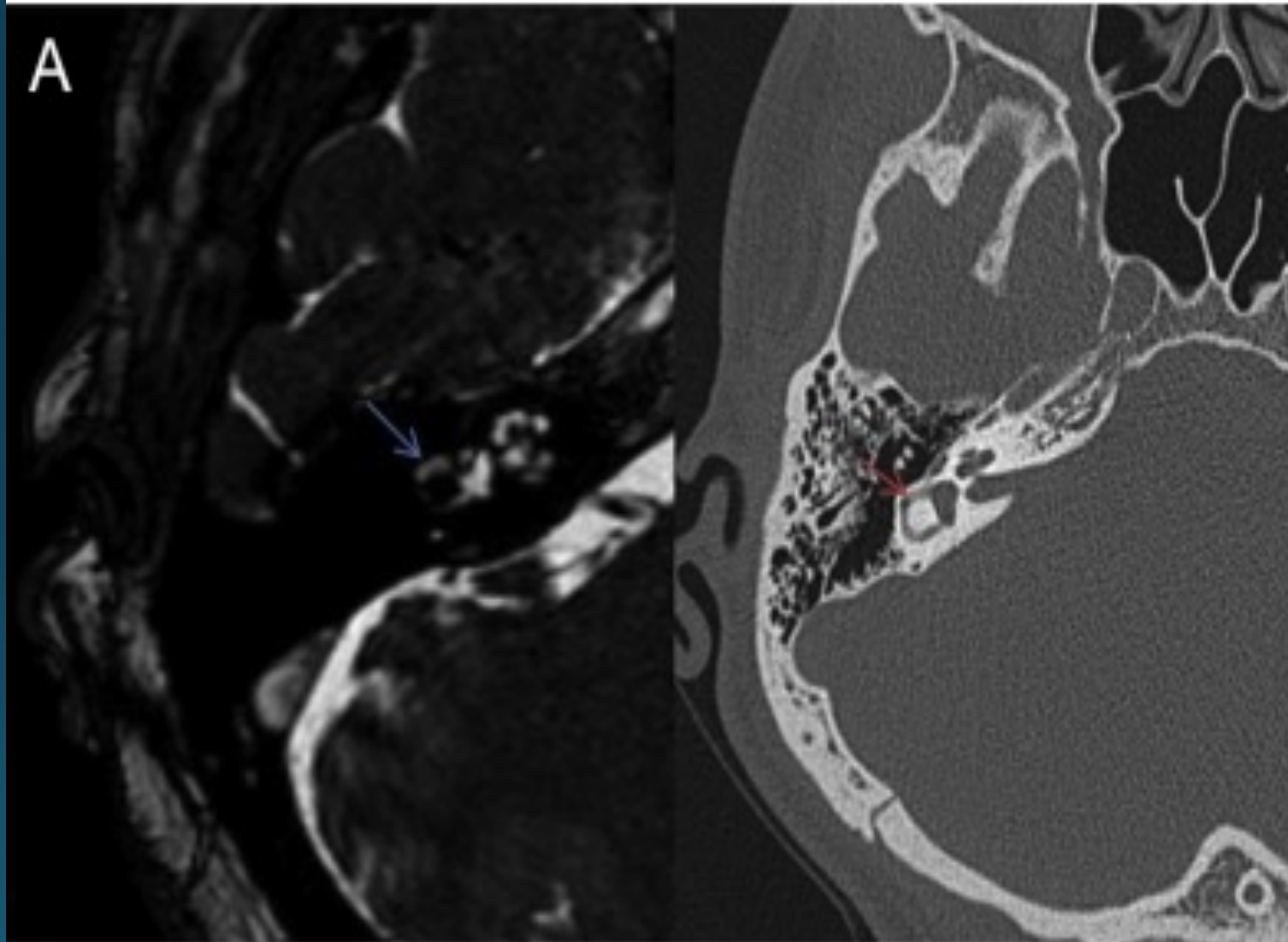


FIG. 3. Reformatted CT images perpendicular (A, Stenver's plane) or parallel (B, Pöschl's plane) to the SSC may aid visualization of a bony dehiscence (arrows). Lines superimposed on an axial temporal bone CT image indicate the plane of reconstruction for (C) Stenver and (D) Pöschl views.

Lateral SC Anatomy: Imaging Findings

T-2 weighted MRI Axial

Temporal CT Axial



BLUE ARROW:

- Endolymph (hyperintense fluid) from lateral semicircular canal within bone tissue (low intensity)
- Presence of hypointense bone tissue between canal and posterior cranial fossa

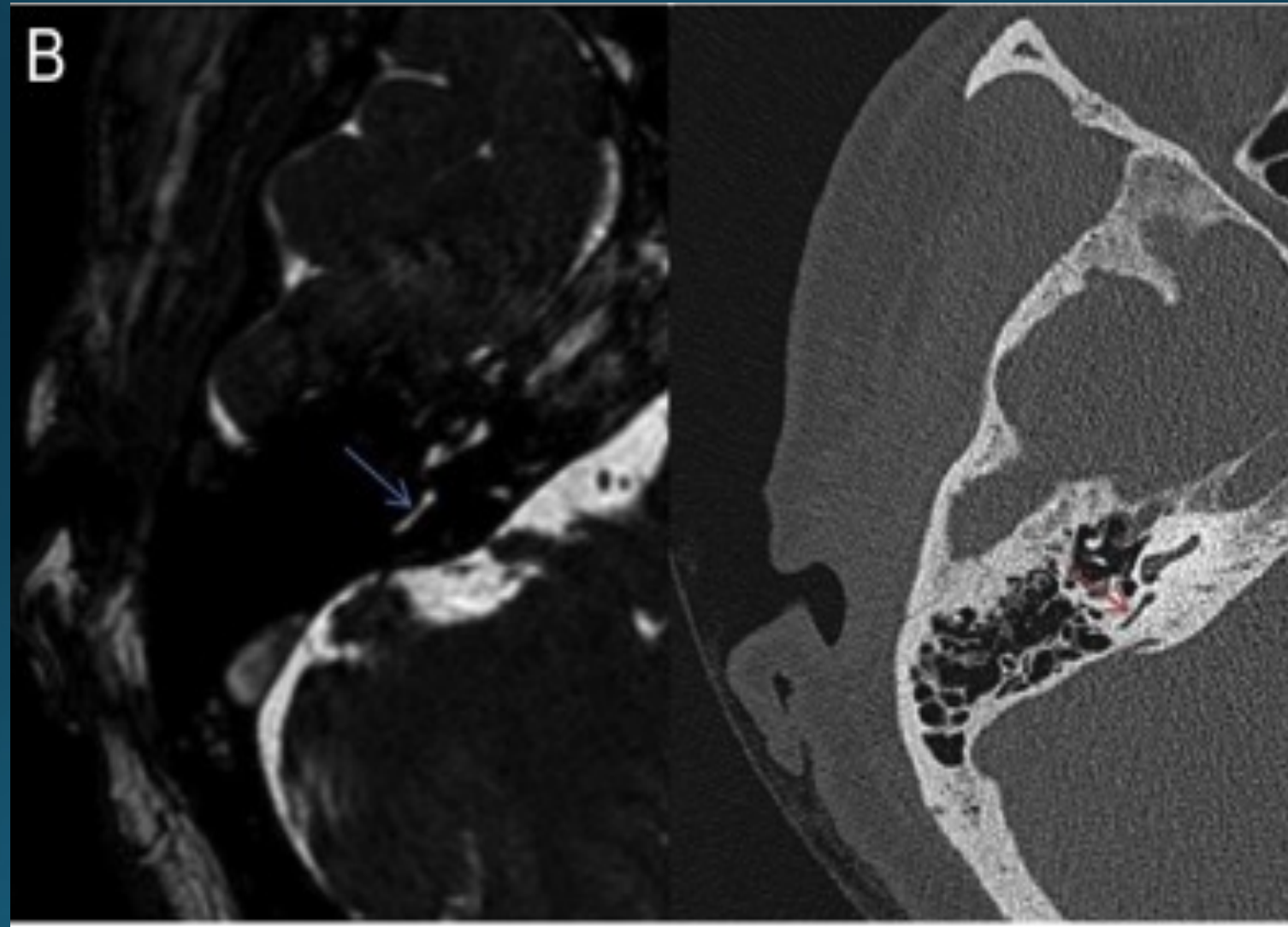
RED ARROW:

No bone defect can be seen in the lateral semicircular canal

Posterior SC Anatomy: Imaging Findings

T-2 weighted MRI Axial

Temporal CT Axial



B

BLUE ARROW:

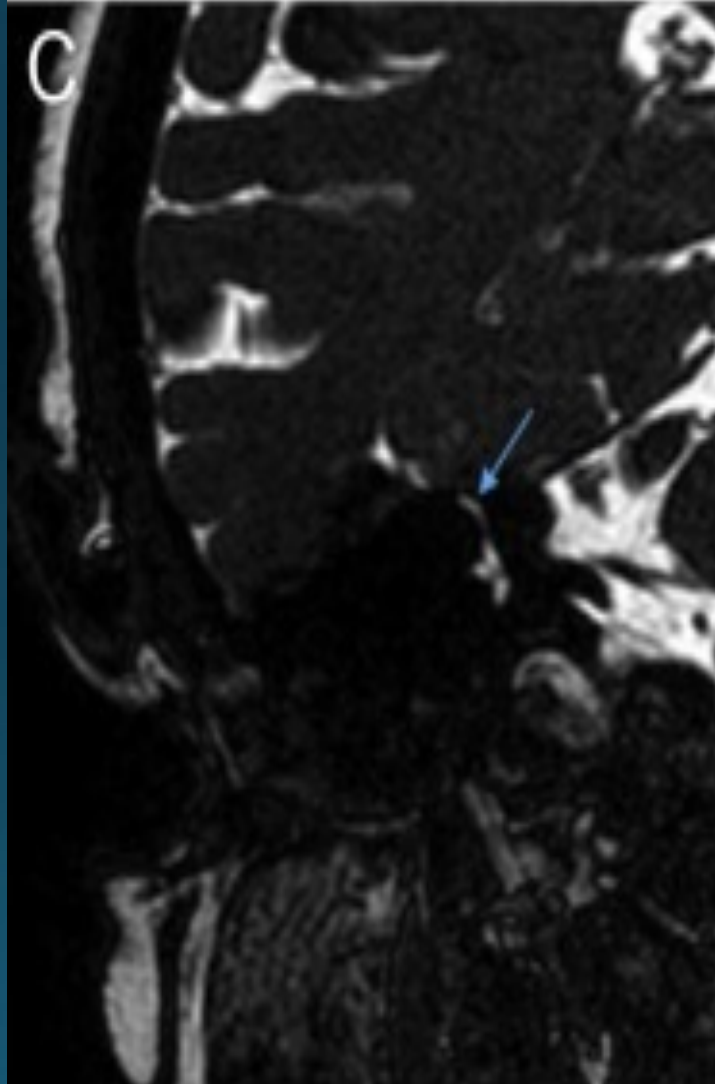
- Endolymph (hyperintense fluid) from posterior semicircular canal within bone tissue (low intensity)
- Presence of hypointense bone tissue between canal and posterior cranial fossa

RED ARROW:

No bone defect can be seen in the posterior semicircular canal

Superior SC Anatomy: Imaging Findings

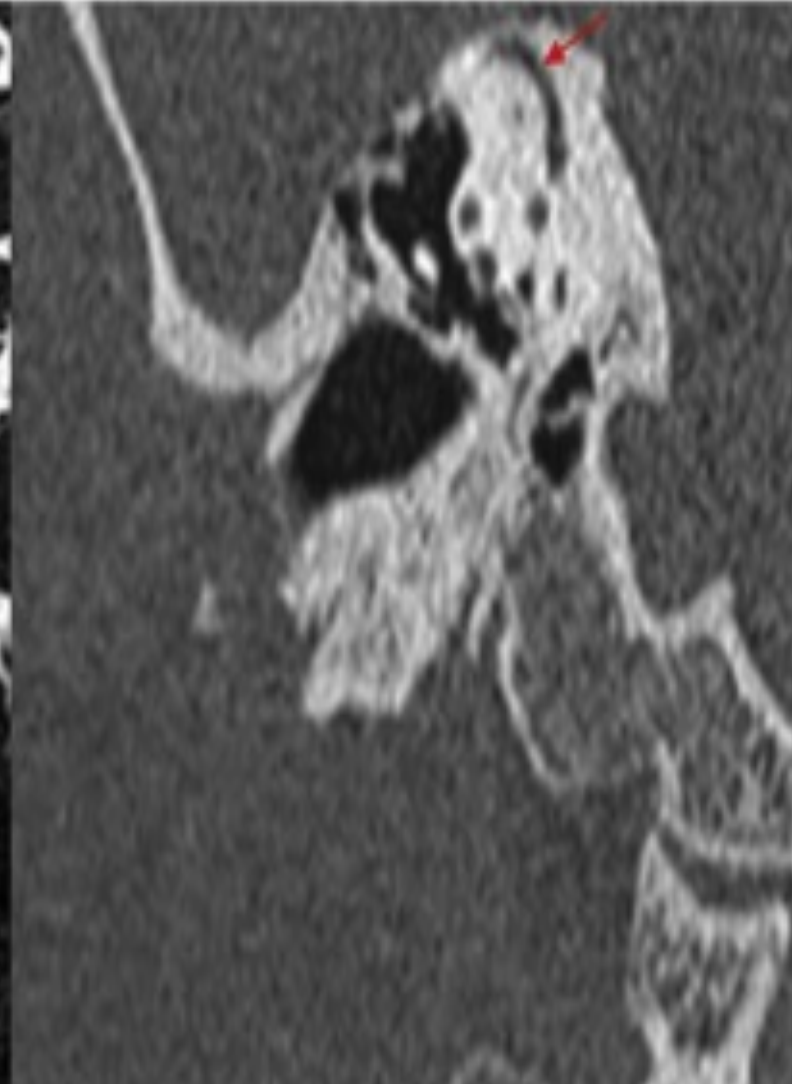
T-2 weighted MRI Axial



BLUE ARROW:

- Endolymph (hyperintense fluid) from superior semicircular canal within bone tissue (low intensity)
- Presence of hypointense bone tissue between canal and posterior cranial fossa

Temporal CT Axial



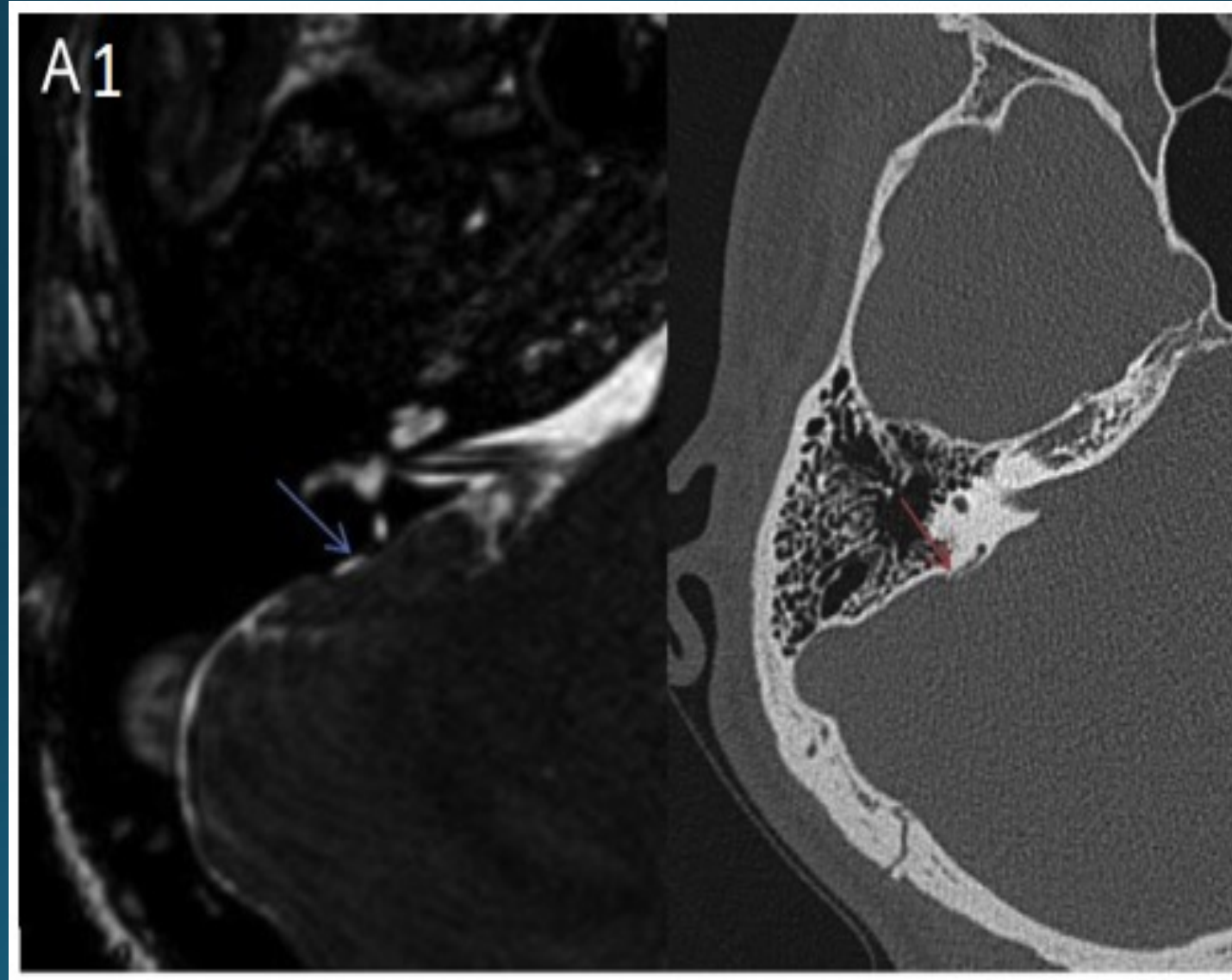
RED ARROW:

- No bone defect can be seen in the superior semicircular canal

Posterior SCDS: Imaging Findings

T-2 weighted MRI Axial

Temporal CT Axial



BLUE ARROW:

-Endolymph (hyperintense fluid) from posterior semicircular canal moves out of the bony structure (low intensity) to the CSF (hyperintense fluid)

-No hypointense bone tissue between canal and posterior cranial fossa

RED ARROW:

Bone defect at the top of the posterior semicircular canal communicating with the posterior cranial fossa

Superior SCDS: Imaging Findings

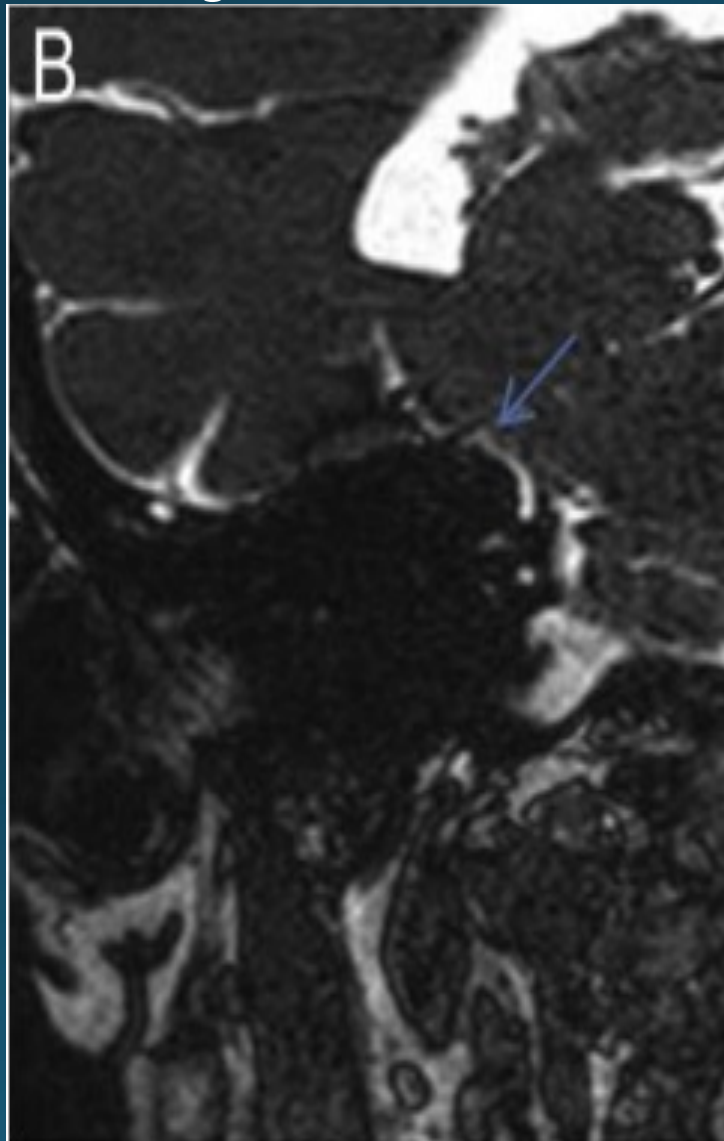
T-2 weighted MRI Coronal

Temporal CT Coronal

BLUE ARROW:

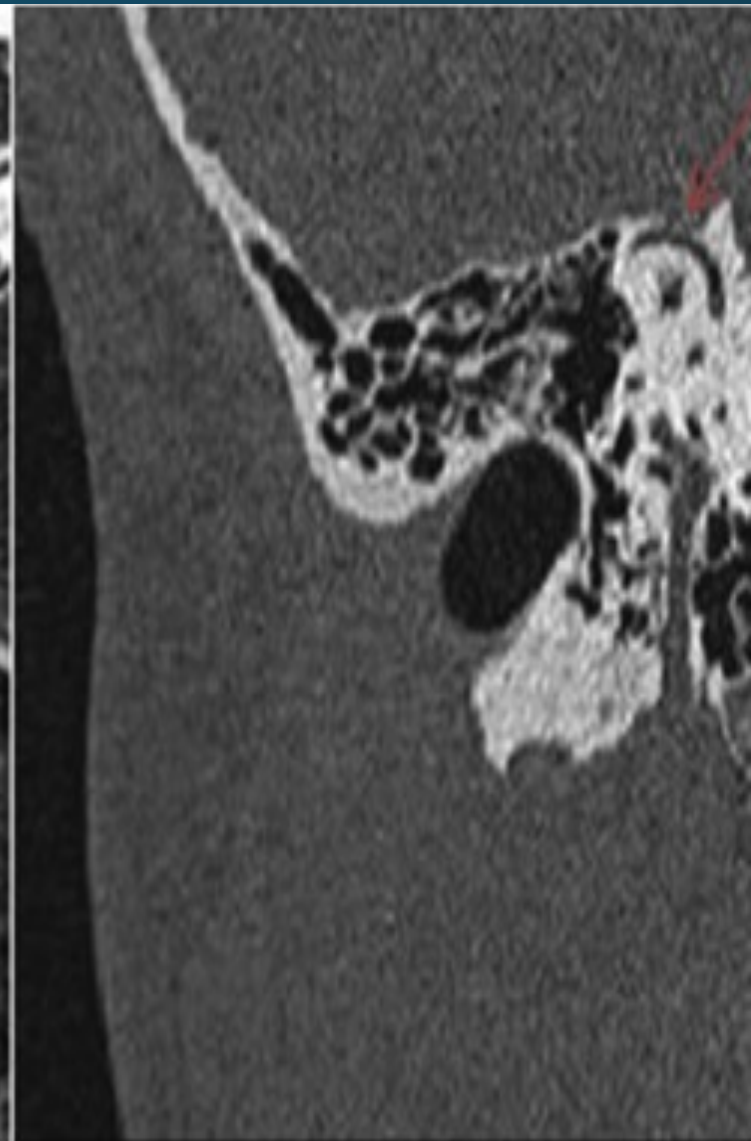
-Endolymph (hyperintense fluid) from superior semicircular canal moves out of the bony structure (low intensity) to the CSF (hyperintense fluid)

-No hypointense bone tissue covering the canal and middle cranial fossa



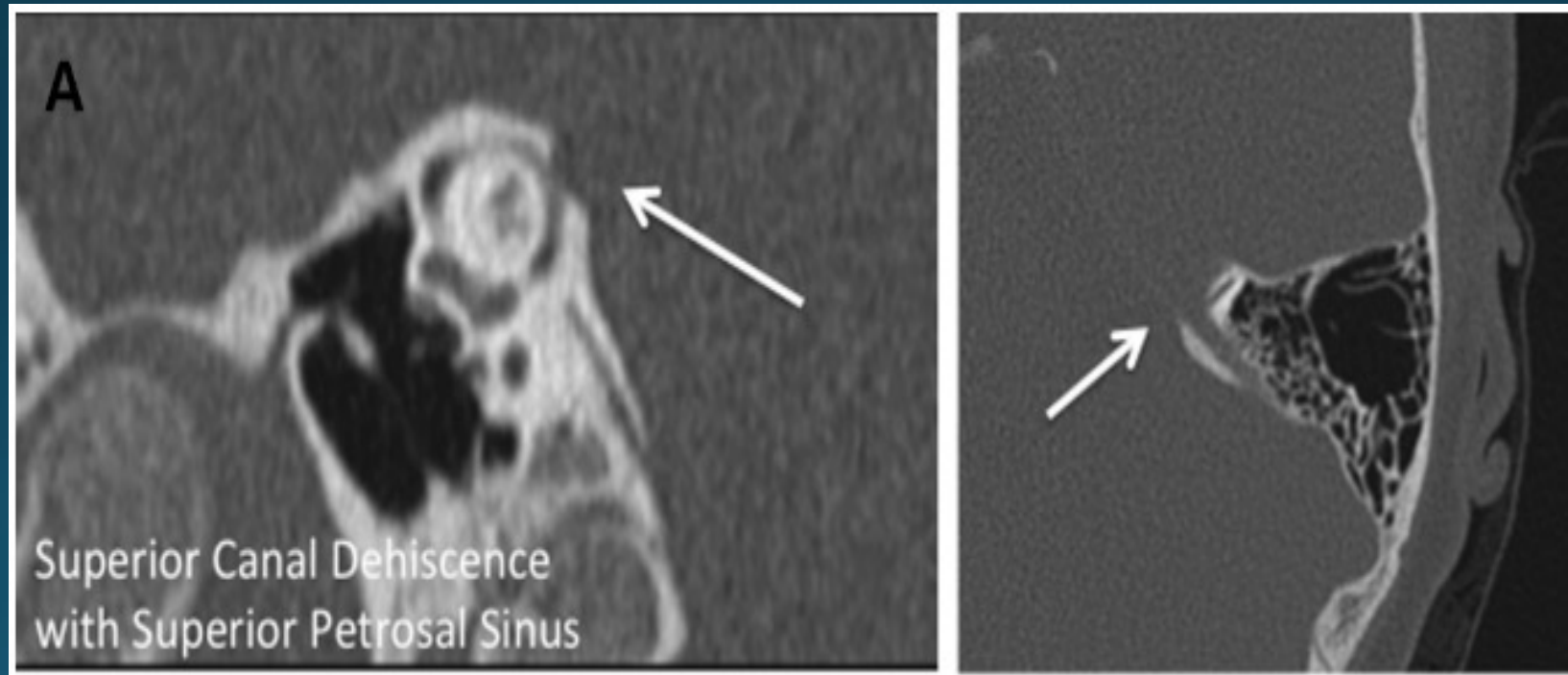
RED ARROW:

Bone defect at the top of the superior semicircular canal communicating with the posterior cranial fossa



Vascular Associations of SCDS

- Superior SCDS:
 - Superior Petrosal Sinus may be in close proximity
 - Communication between semicircular canal with the middle cranial fossa and/or superior petrosal sinus



Superior Canal Dehiscence
with Superior Petrosal Sinus

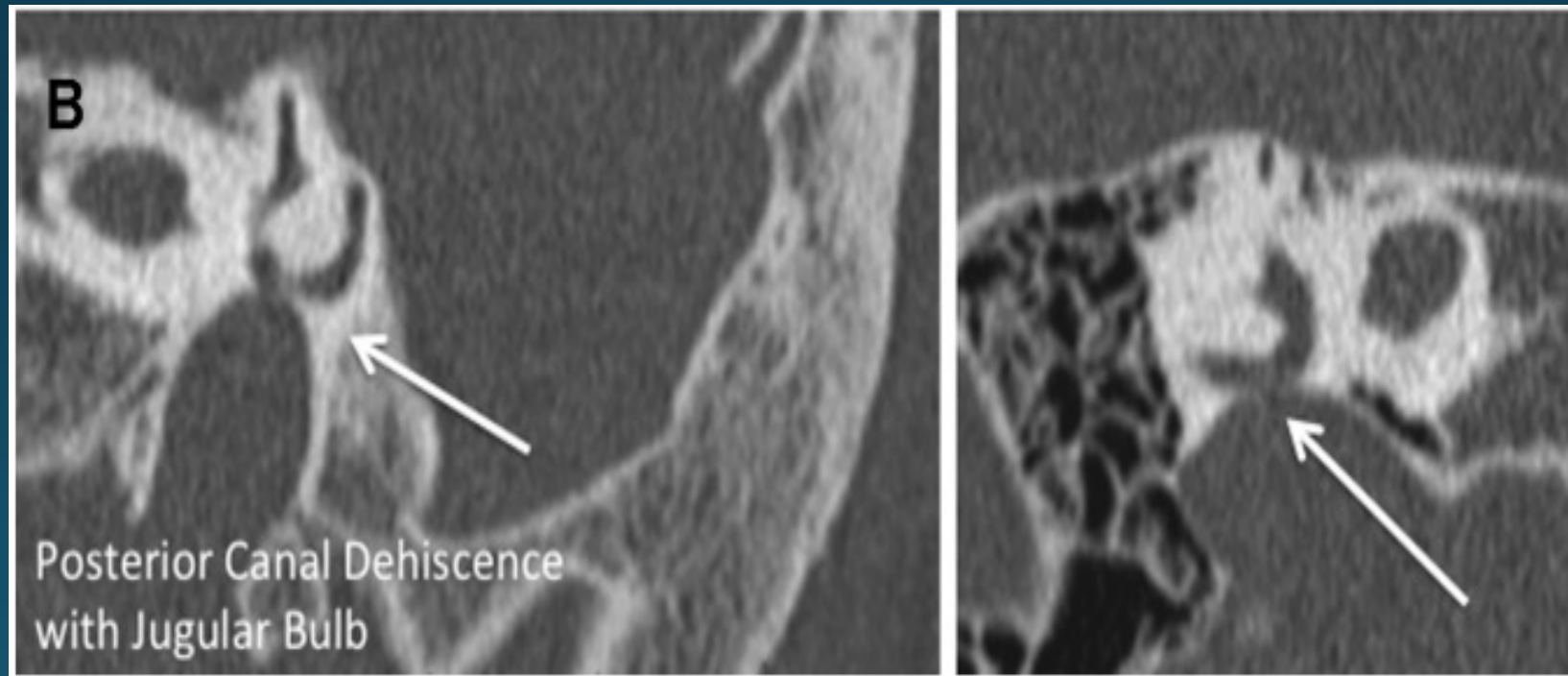
Poschl plane

Axial plane

Saxby et al., 2015

Vascular Associations of SCDS

- Posterior SCDS:
 - Jugular bulb may be in close proximity
 - Communication between semicircular canal with the posterior cranial fossa and/or jugular bulb



Poschl plane

Axial plane

Saxby et al., 2015

Management of SCDS

- Mainly surgical
 - Multiple techniques, such as canal plugging and resurfacing
- Multiple surgical approaches can be used and choice takes into consideration where the dehiscence is located
- For radiologists, it is important to be familiar with all types of dehiscence and accurately describe anatomic correlations and all possible sites of dehiscence, as not every surgical approach is able to access all semicircular canals

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