Australian Dizziness Clinic

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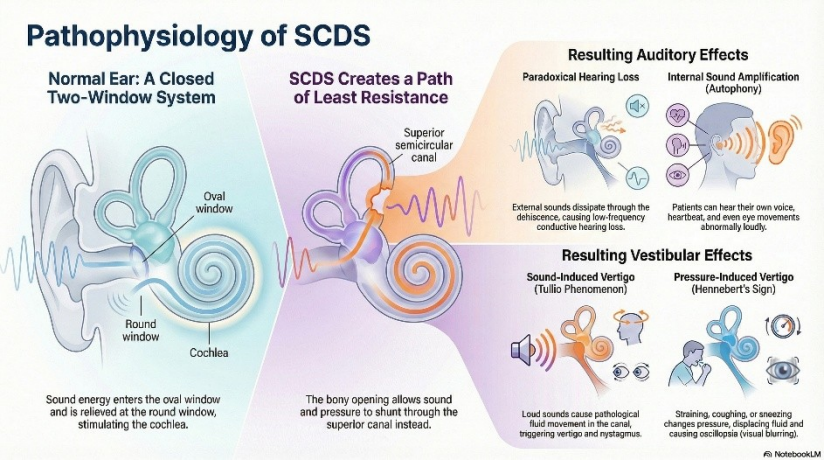
CLINICIAN CHEAT SHEET: Superior Semicircular Canal Dehiscence (SSCD)

1. EPIDEMIOLOGY & DEFINITIONS

- **Incidence/Prevalence:**
  - Anatomic Dehiscence (Radiologic): 0.5–1.5% of adult population. High-res CT may show up to 3%.
  - Symptomatic SCDS: Rare; exact prevalence unknown but significantly lower than anatomic prevalence.
  - Paediatric: High rate of dehiscence in infants (~37% <2 yrs) due to incomplete ossification; mostly asymptomatic and resolves as bone matures.
  - Demographics: Adults: Female > Male (middle-aged bias). Children: Male > Female (1.65:1).
  - Bilateral: Occurs in ~25% of patients.
- **Definitions:**
  - Superior Semicircular Canal Dehiscence (SSCD): Bony defect in the roof (tegmen) of the superior canal.
  - "Third Mobile Window": Pathological shunt of inner ear fluid/pressure through the dehiscence, altering mechanics (dissipates sound energy, increases vestibular sensitivity).
  - Tullio Phenomenon: Vertigo/nystagmus induced by loud sounds.
  - Hennebert's Sign: Vertigo/nystagmus induced by pressure changes (tragal pressure, Valsalva).



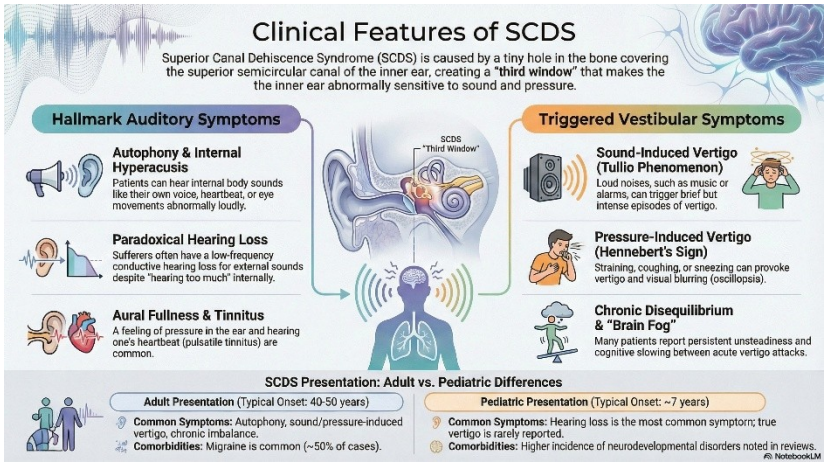
2. RISK FACTOR DOMAINS (Multifactorial)

- **Congenital / Developmental:**
  - Incomplete ossification of canal roof in childhood.
  - Genetic predisposition (e.g., CDH23 variants in Usher syndrome).
- **Anatomic / Structural:**
  - Generalized bone dysplasia or skull base thinning (multiple tegmen defects).
  - Low-lying tegmen or specific temporal bone morphology.
- **Metabolic / Age-Related:**
  - Osteopenia/Osteoporosis (progressive bone thinning in elderly females).
- **Acquired Triggers:**
  - Head Trauma: May precipitate symptoms in asymptomatic dehiscence.
  - ICP: Chronic elevated intracranial pressure (erosion hypothesis).
  - Mechanical: Intense straining/lifting events.

3. CLINICAL ASSESSMENT PROTOCOL

A. History (Key Inquiries)

- **The "Big 3" Symptoms:**
  - Autophony: "Do you hear your own voice loudly/distorted?" (Occurs in >50%).
  - Internal Hyperacusis: "Can you hear your eyes blink, heartbeat (pulsatile tinnitus), or joints creak?".
  - Triggered Vertigo: "Do loud noises (alarms, music) or straining (coughing, heavy lifting) cause dizziness?".
- **Red Flag Symptoms:**
  - "Brain fog" / cognitive slowing.
  - Aural fullness (often confused with Eustachian Tube Dysfunction).
  - Visual blurring (oscillopsia) with loud sounds.



B. Physical Exam

- **Otoscopy:** Typically, NORMAL (Critical to distinguish from middle ear disease).
- **Tuning Fork Tests:**
  - Weber: Lateralizes to the AFFECTED ear.
  - Distal Sensitivity: Patient may hear tuning fork placed on ankle/knee in the affected ear.
- **Vestibular Provocation:**
  - Tragal Pressure: Positive if nystagmus/vertigo induced.
  - Valsalva: Against closed glottis or pinched nose may elicit vertical-torsional nystagmus (aligns with superior canal plane).

C. Diagnostic Workup

- **Diagnosis requires Triad:** Compatible Symptoms + Physiologic Evidence + Anatomic Defect.

Investigation	Findings in SCDS	Clinical Pearl
Audiometry	Low-frequency Air-Bone Gap (20-40 dB) with intact acoustic reflexes.	STOP: Do not diagnose Otosclerosis if reflexes are

	Supranormal bone conduction (<0 dB).	present.
VEMPs	cVEMP: Low threshold (<75-80 dB nHL). oVEMP: High amplitude (>17 µV, 500Hz).	Highly Specific: Raised oVEMP amplitude is diagnostic.
CT Imaging	High-Resolution (≤0.5mm): Bony defect in Pöschl & Stenver views.	1mm cuts or standard axial views commonly miss or over-call dehiscence.
ECochG	Elevated SP:AP ratio (>0.4).	Mimics Meniere's: ratio normalizes post-op.

4. DIFFERENTIAL DIAGNOSIS

Table: SCDS vs. Common Mimics

Feature	SCDS	Otosclerosis	Patulous Eustachian Tube	Ménière's Disease
Vertigo Trigger	Loud sound (Tullio) or Pressure (Hennebert)	Absent (usually)	None (Vestibular symptoms absent)	Spontaneous (not triggered)
Autophony	Voice, Heartbeat, Eye movements, Joints	None	Breath sounds, Voice (echoing)	Roaring tinnitus (not autophony)
Hearing Loss	Pseudo-Conductive (Low freq ABG)	Conductive	None / Fluctuating	Sensorineural (Low freq)
Acoustic Reflexes	PRESENT / INTACT	ABSENT	Normal	Normal / Recruited
Relief Manoeuvre	None (Supine may help 50%)	None	Head down / Supine (Immediate relief)	None specific
VEMP	Low Threshold (Hypersensitive)	Absent or Normal	Normal	Normal or Reduced

5. MANAGEMENT & PREVENTION

Risk Stratification & Management Pathway

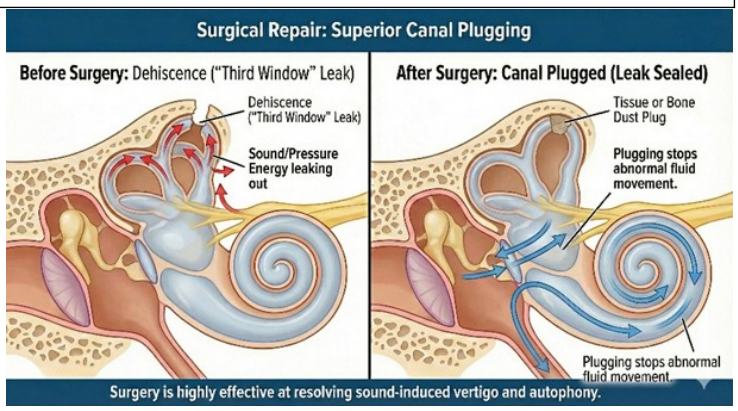
Risk/Severity Level	Criteria	Management Strategy
LOW (Mild)	Incidental finding or mild annoyance. No fall risk.	Conservative: Observation & Reassurance. Educate on benign nature.
MEDIUM (Auditory)	Hearing loss/Autophony dominates. Minimal vertigo.	Non-Surgical: Hearing aids (air conduction). Investigational: Round Window Reinforcement (RWR).
HIGH (Debilitating)	Tullio/Hennebert symptoms limiting life/work. Disequilibrium.	Surgical Repair: MCF Craniotomy or Transmastoid. Method: Plugging + Resurfacing (Standard of Care).

Pharmacological Management (Symptomatic/Adjunctive)

Indication	Drug Class	Pearls / Notes
Migraine (Co-morbid)	Topiramate, TCA, Beta-blockers	Essential: 50% of SCDS patients have migraine. Migraine tx can reduce dizziness significantly.
Straining Prevention	Stool Softeners (e.g., Docusate)	Use to prevent Valsalva/ICP spikes during bowel movements.
Ménière's Overlap	Diuretics / Betahistine	Use if hydrops suspected or confirmed on MRI.
Anxiety	SSRIs / Anxiolytics	For secondary anxiety due to chronic sensory overload.

Non-Pharmacological Interventions

- Trigger Avoidance: Earplugs in loud environments. Avoid heavy lifting/straining.
- Vestibular Rehab: Limited utility for mechanical defect, but useful for general stability/compensation post-op.
- Comorbidity Mgmt: Treat migraine aggressively (diet, lifestyle) prior to surgical consideration.



6. FOLLOW-UP & RED FLAGS

Follow-Up Protocol

- Conservative: Annual audiogram to monitor ABG stability.
- Post-Surgical:
  - Immediate: Watch for BPPV (25% incidence).
  - 3 Months: Repeat Audiogram & VEMP. Success = Closure of ABG & Normalization of VEMP thresholds.
  - Revision: Considered if symptoms persist and imaging confirms residual dehiscence/leak (MRI with gadolinium).