Superior Canal Dehiscence Syndrome (SCDS):

A Vestibular Physician's Deep Review of Pathophysiology, Diagnosis, and Management

Vestibular Medicine for Vestibular Physicians

Peripheral Vestibular Pathology — Module 2.5

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How to Use This Review

This literature review forms part of the Vestibular Medicine for Vestibular Physicians series published by the Australian Dizziness Clinics Education Hub. It is written for vestibular physicians, neuro-otologists, advanced ENT trainees, and vestibular physiotherapists working at the deep end of peripheral vestibular practice, where a working command of mechanism, criteria, and atypical presentations is expected rather than optional.

The review is dense by design — intended as a 30–40 minute deep read or a desktop reference. It is supported by an A4 clinician cheat sheet, short-form clinician videos, audio episodes, and a patient information leaflet within the same Education Hub module.

Callout Box Guide

□ **Key Point:** Foundational concepts and summary statements that anchor the core clinical content of each section.

□ **Clinical Insight:** Clinically relevant observations for direct application in assessment and management.

□ **Clinical Pearl:** High-yield memorable clinical points — the take-home messages most likely to change practice.

□ **Important:** Red flags, atypical presentations, and critical safety points requiring escalation or imaging.

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I. Introduction and Epidemiology

Historical Background

Superior semicircular canal dehiscence syndrome (SCDS) was first formally characterised by Dr Lloyd B. Minor and colleagues in 1998, in a landmark case series published in the Archives of Otolaryngology — Head and Neck Surgery [1]. Minor described a group of patients experiencing chronic disequilibrium and vertigo precipitated by loud sounds (Tullio phenomenon) or by pressure changes (Hennebert's sign), accompanied by torsional-downbeat nystagmus in the plane of the superior semicircular canal. High-resolution computed tomography revealed a focal bony opening — a dehiscence — in the arcuate eminence of the temporal bone overlying the superior canal [1,2]. This anatomical defect created an abnormal communication between the inner ear and the middle cranial fossa, providing a structural basis for what would become known as the "third mobile window" syndrome.

The clinical phenomena described by Minor had conceptual precursors spanning several decades. In 1929, Pietro Tullio demonstrated that creating a fenestration in the semicircular canal of pigeons produced sound-evoked vestibular responses — a finding named the Tullio phenomenon [1,45]. Concurrently, Heinrich Hennebert observed that patients with congenital syphilis could experience vertigo and compensatory eye movements when external ear canal pressure was applied — the Hennebert sign [1]. In the mid-twentieth century, surgical fenestration of the horizontal semicircular canal for otosclerosis, as described by Cawthorne, produced similar auditory–vestibular symptoms, demonstrating that any iatrogenic opening in the otic capsule could mimic the clinical picture [3]. Minor's seminal contribution was to localise the defect anatomically to the superior canal and to demonstrate reversal of symptoms after surgical repair — establishing SCDS as a surgically treatable condition with a definable pathological substrate [1,4].

Since the initial 1998 report, awareness of SCDS has grown substantially. The condition is now recognised globally, with over 600 documented cases in the literature by 2010 and many thousands subsequently diagnosed as high-resolution temporal bone CT became widely available [1,5]. SCDS is increasingly understood not as a rare curiosity but as an under-recognised cause of chronic auditory–vestibular morbidity — one that for years was frequently misdiagnosed as otosclerosis, Ménière's disease, or a functional disorder [2,5].

□ **Key Point:** SCDS was first described by Minor et al. in 1998. The syndrome results from a focal bony defect overlying the superior semicircular canal, creating a pathological "third window" that disrupts both auditory and vestibular mechanics. It is a surgically treatable condition with excellent outcomes when correctly diagnosed [1,4].

Incidence and Prevalence

The prevalence of anatomical SCD visible on imaging is considerably higher than the prevalence of symptomatic SCDS. Large temporal bone autopsy studies report dehiscences in approximately 0.5% of specimens [6], while prospective high-resolution CT series in otologically normal populations identify dehiscences in approximately 1–3% of individuals [7]. Earlier CT studies using 1 mm slice thickness systematically overestimated prevalence at up to 9% due to partial volume averaging artefact — an important source of false positives that led to over-diagnosis before the adoption of sub-millimetre acquisitions [7,8]. The critical implication is that radiological dehiscence alone is not sufficient for diagnosis: the majority of individuals with an anatomical opening are asymptomatic, and clinical, physiological, and imaging criteria must all be satisfied before the diagnosis can be made [9].

Bilateral dehiscence is identified in approximately 25% of patients with symptomatic SCDS [1,7]. The high bilateral rate, combined with developmental data, strongly supports a constitutional predisposition — either congenitally thin bone or incomplete ossification of the superior canal roof — rather than a purely acquired mechanism [10,21].

Paediatric Epidemiology

The bony roof of the superior semicircular canal (the tegmen superior canal) normally undergoes progressive ossification through infancy and early childhood. This developmental timeline explains the striking age-dependent pattern of radiological dehiscence in children. Multicentre paediatric CT review studies report dehiscence rates of approximately 36.7% in children under two years of age, falling to 5.6%

at ages 2–8, and 3% by ages 9–18 — approaching adult rates [11]. This ontogenetic trajectory strongly implicates delayed or incomplete ossification as the dominant mechanism in most cases, and provides the mechanistic framework for understanding why symptomatic SCDS is overwhelmingly an adult diagnosis despite the high paediatric radiological prevalence [10,11].

Symptomatic SCDS in children is rare. A systematic review in 2017 identified approximately 122 paediatric cases in the published literature, with a mean age of presentation of approximately seven years [12]. Importantly, the paediatric phenotype differs from the adult presentation: hearing loss — including conductive, sensorineural, and mixed patterns — predominates, while the classic vestibular symptoms (Tullio, Hennebert, chronic disequilibrium) are less prominent [12]. Paediatric SCDS also shows a male predominance of approximately 1.65:1, in contrast to the adult population where a slight female preponderance has been noted in some series [12]. Management in children requires particular caution given the risk of spontaneous ossification with maturation, the potential for unnecessary intervention, and the age-specific risks of surgical approaches [12,13].

Table 1. Epidemiology of Superior Canal Dehiscence by Age Group

Parameter	Paediatric (<18 years)	Adult (>18 years)
Radiological prevalence	Up to 36.7% in infants; <3% by adolescence [11]	0.5–3% on HRCT [6,7]
Symptomatic prevalence	Rare; ~122 cases in literature [12]	Uncommon; exact prevalence unknown [5]
Bilateral involvement	Not well characterised	~25% of symptomatic cases [7]
Sex predominance	Male (~1.65:1) [12]	Slight female predominance in some series [1]
Typical age at diagnosis	Mean ~7 years for symptomatic cases [12]	Mid-life (40s–50s) most common [1]
Dominant symptoms	Hearing loss (conductive/mixed) [12]	Mixed auditory–vestibular syndrome [1,2]
Aetiology	Developmental ossification failure [10,11]	Congenital + possible acquired progression [21]

II. Pathophysiology — The Third Mobile Window Mechanism

The Two-Window Model and its Disruption

Under normal conditions, the inner ear communicates with the middle ear through two compliant windows: the oval window (into which the stapes footplate transmits sound vibration) and the round window (which acts as a pressure-release valve for the incompressible perilymph fluid column). This “two-window” model ensures that acoustic energy drives the basilar membrane efficiently and produces the cochlear travelling wave responsible for hearing [14,15]. The vestibular apparatus — the semicircular canals, utricle, and saccule — is embedded within the same perilymph and endolymph compartments but is physically isolated from sound energy in a normal ear because the otic capsule bone surrounding it is rigid [14].

A dehiscence in the superior canal roof creates a third compliant interface between the inner ear and the middle cranial fossa [14,15,42]. This third window fundamentally disrupts the fluid mechanics established by the two-window model: acoustic energy and pressure changes can now shunt through the dehiscence, diverting fluid displacement away from the normal cochlear partition pathway and instead driving endolymph movement within the superior semicircular canal [15,42]. The net effect is a set of paradoxical auditory and vestibular consequences that are mechanistically explained by this single anatomical defect [14,15].

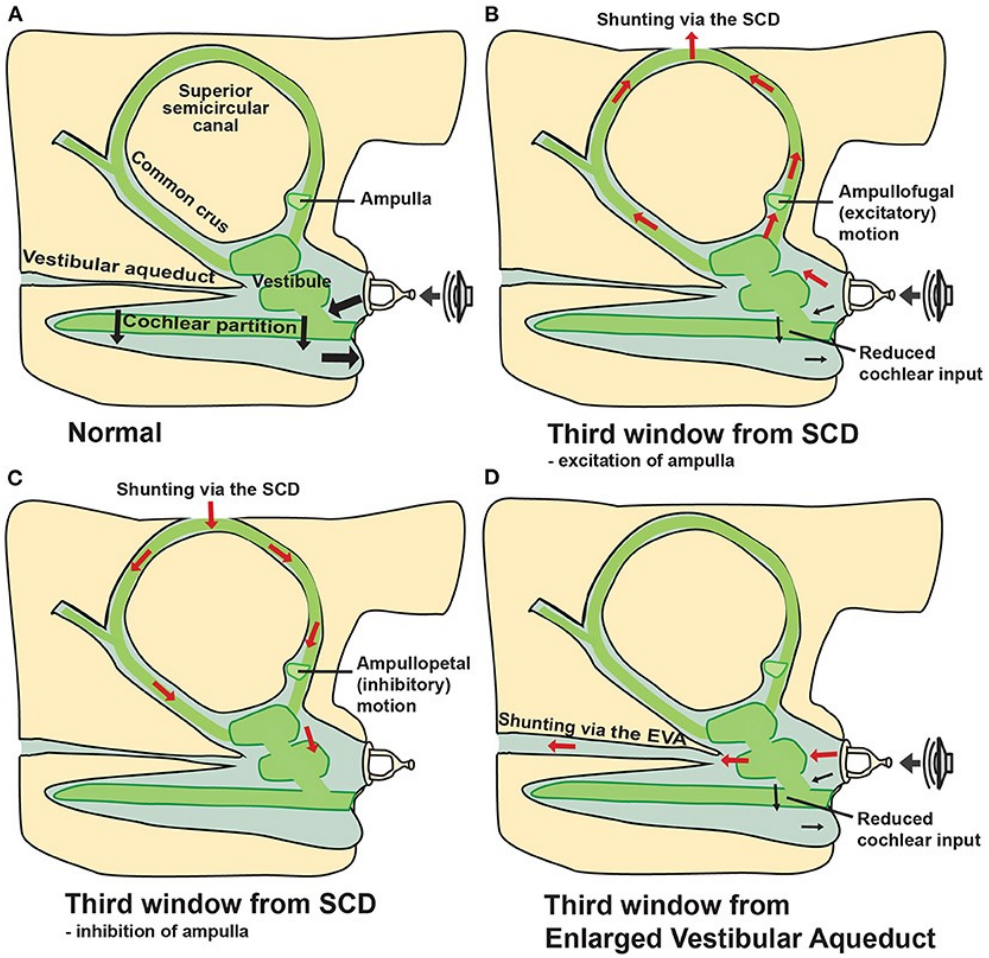

Figure 1. "Third window" mechanism due to SCD and enlarged vestibular aqueduct. (A) Normal two-window mechanics. (B) Air-conducted sound shunted toward the dehiscence, raising air-conduction thresholds and producing Tullio phenomenon. (C) Elevated intracranial pressure transmitted via dehiscence causing vestibular symptoms (Hennebert sign). (D) Enlarged vestibular aqueduct as an alternative third window syndrome.
Source: Eberhard KE, Chari DA, Nakajima HH, Kozin ED, Lee DJ. Front Neurol. 2021;12:638574. doi:10.3389/fneur.2021.638574.

Auditory Consequences of the Third Window

The auditory consequences of the third window operate through two distinct mechanisms that produce seemingly paradoxical effects — conductive-type hearing loss coexisting with hypersensitivity to internally generated sounds [14,16,42]. First, air-conducted acoustic energy entering through the stapes is partially diverted through the dehiscence rather than being directed entirely across the cochlear partition from oval window to round window. This reduces the effective energy available for basilar membrane displacement, raising air-conduction thresholds preferentially at low frequencies (250–1000 Hz) where the dehiscence impedance is lowest — producing the characteristic low-frequency air–bone gap on audiometry [16,17,42].

Second, the third window increases the efficiency of bone-conducted sound transmission to the inner ear by providing an additional pathway for perilymph displacement. Bone-conducted sounds bypass the middle ear impedance and drive fluid movement more effectively in the SCDS ear than in a normal ear, producing supranormal bone conduction thresholds (values below 0 dB HL) and explaining the phenomenon of internal hyperacusis [16,17]. Patients perceive their own voice, heartbeat, eye movements, and even bowel sounds with distracting clarity because bone-conducted internal vibrations are amplified by the additional fluid pathway created by the dehiscence [1,2]. Pulsatile tinnitus synchronous with the heartbeat arises because dural pulsations from the middle cranial fossa are transmitted directly to the perilymph through the bony defect [18].

Vestibular Consequences of the Third Window

In addition to cochlear effects, the third window permits pathological coupling of intracranial pressure changes and middle ear pressure to the vestibular labyrinth [1,15,19]. Sound-induced pressure waves can directly drive endolymph in the superior semicircular canal, deflecting the cupula and producing vertigo with nystagmus in the plane of the superior canal — the Tullio phenomenon [1,19]. The nystagmus is characteristically torsional-downbeat (excitatory, superior canal stimulation), and its onset is immediate and time-locked to the triggering sound [1,45].

Pressure changes transmitted via the third window explain the Hennebert sign [15,19]. Positive pressure in the middle ear (as with pneumatic otoscopy or Valsalva against closed nostrils) drives fluid ampullofugally in the superior canal; conversely, raised intracranial pressure (Valsalva against closed glottis, heavy lifting, coughing) is transmitted from the middle cranial fossa through the dehiscence, pushing fluid ampullopetally [15,19]. The precise direction of nystagmus with each pressure manoeuvre depends on whether the cupula is deflected excitatorily or inhibitorily — a point of diagnostic significance when examining the patient at the bedside [15,19].

□ **Clinical Pearl:** The torsional-downbeat nystagmus of Tullio phenomenon reflects excitation of the ipsilateral superior canal ampullary neurons. The key examination finding is nystagmus beating in the plane of the superior canal (torsional component toward the affected ear, vertical component downward). When reproducibly triggered by loud sound or pressure, this is pathognomonic of a superior canal dehiscence on the stimulated side [1,15].

Aetiology — Congenital vs Acquired Mechanisms

The weight of epidemiological and histological evidence supports a predominantly congenital aetiology for most SCDS cases. The superior canal roof normally achieves full ossification during the first decade of life; failure to complete this developmental process leaves a focal area of thin or absent bone vulnerable throughout adulthood [10,11]. The high bilateral prevalence (~25%), combined with documented familial clustering and associations with gene variants affecting bone mineralisation, supports a constitutional predisposition [1,7,20].

An acquired or progressive component may account for a subset of cases. Progressive thinning of the tegmen with ageing, skull base trauma, chronic elevated intracranial pressure, and rare inflammatory or neoplastic processes have all been proposed as precipitants that convert subclinical thin bone into frank dehiscence [1,21]. Serial CT imaging in a small number of patients has documented progression from near-dehiscence to dehiscence over time. Coexisting tegmen defects (multiple superior canal thinning, dehiscent facial nerve canal) in some patients hint at a more generalised bone dysplasia phenotype [21]. The morphological diversity of otic capsule defects, including gushers and oozers, reflects the spectrum of bony fragility in this region [48].

SCDS is one member of the broader family of "third window syndromes." Other conditions that create pathological inner ear windows — enlarged vestibular aqueduct (EVA), posterior or lateral canal dehiscence, cochlea-jugular bulb dehiscence — produce overlapping auditory–vestibular phenotypes through the same fundamental mechanism [1,14,22,46]. Recognising this mechanistic kinship is essential for the differential diagnosis and for interpreting physiological test results in the context of the full clinical picture.

□ **Key Point:** SCDS belongs to the broader "third window syndrome" family, which includes enlarged vestibular aqueduct, posterior canal dehiscence, and cochlea-jugular bulb dehiscence. All share the fundamental mechanism of a pathological shunt that bypasses the normal two-window model. CT imaging must evaluate for all potential dehiscence sites — not only the superior canal — before assigning a diagnosis.

III. Clinical Features: Auditory and Vestibular Presentations

Auditory Symptoms

Autophony — the abnormal perception of one's own voice as excessively loud, resonant, or distorted in the affected ear — is one of the most characteristic and diagnostically useful symptoms of SCDS, present in more than 50% of patients [1,2]. Autophony in SCDS results from enhanced bone-conduction sensitivity: the patient's own vocal vibrations, transmitted through the skull, are amplified by the third window effect. Unlike autophony from a patulous Eustachian tube (which typically involves breathing sounds and improves supine), SCDS autophony involves voice and body sounds and does not reliably change with posture [2].

Internal hyperacusis encompasses autophony as well as perception of heartbeat (pulsatile tinnitus, which in SCDS is a true bone-conducted vascular sound amplified by the third window [18]), eye movements (described as "clicking" or "swooshing"), footfall while walking, joint creaking, and even bowel sounds [1,2]. These symptoms are highly specific to SCDS and third window syndromes and are rarely described with other vestibular disorders. Patients commonly describe profound distraction and social disability from these symptoms. A perceived low-frequency conductive hearing loss for external sounds coexists paradoxically — patients hear their own internal sounds too loudly but perceive external sounds as muffled [1,2,16].

Vestibular Symptoms

Sound-induced vertigo (Tullio phenomenon) and pressure-induced vertigo (Hennebert sign) are the pathognomonic vestibular features of SCDS [1,2,45]. Tullio phenomenon presents as brief episodes of vertigo, oscillopsia, or disequilibrium triggered by loud sounds — musical instruments, alarms, shouting, or the patient's own voice. The duration mirrors the stimulus and is measured in seconds [1]. Patients become avoidant of noisy environments, concerts, and conversation, with significant quality of life consequences [2]. Oscillopsia during triggered episodes — visible blur or image jump — reflects the strong VOR driven by pathological superior canal activation and can be debilitating for those working in visually demanding occupations [1].

Hennebert sign — pressure-induced vestibular symptoms — is elicited by any manoeuvre that raises intracranial or middle ear pressure: Valsalva against a closed glottis (straining, heavy lifting, defecation), pneumatic otoscopy, coughing, sneezing, or bearing down [1,19]. Careful bedside examination using a pneumatic otoscope or Valsalva manoeuvre, while the patient fixates on a target and the examiner watches for nystagmus, is a high-yield diagnostic examination procedure. Observation of torsional-downbeat nystagmus during the manoeuvre confirms the diagnosis with high specificity [1,15].

Chronic disequilibrium — a background sense of unsteadiness or tilting that persists between acute episodes — is present in the majority of SCDS patients and is mechanistically distinct from the episodic triggered symptoms [2]. It likely reflects a chronic low-level perturbation of superior canal signalling from minor inertial head movements. This chronic component is frequently mistaken for central pathology, functional vestibular disorder, or psychiatric anxiety, particularly when episodic triggered symptoms are mild or absent, contributing to the mean diagnostic delay of 3–5 years in published series [2,5].

Associated Symptoms and Comorbidities

Cognitive symptoms — difficulty concentrating, mental fatigue, and "brain fog" — are reported by a substantial proportion of SCDS patients and likely result from the cognitive load imposed by continuous, unpredictable vestibular disturbance and internal sound hyperacusis [2]. These symptoms are frequently attributed to anxiety or psychiatric disorders before the diagnosis of SCDS is established. The health-related quality of life in SCDS patients is measurably lower than population norms (mean utility value approximately 0.68 vs. 0.80), and productivity losses are significant [2].

A clinically important comorbidity is vestibular migraine, which co-occurs in approximately 50% of SCDS patients in some series [2,23]. Migraine amplifies both sound sensitivity and vestibular symptoms, creating a complex and often confusing picture. Identification and treatment of comorbid migraine is essential before and after surgical intervention, as uncontrolled migraine predicts suboptimal post-surgical outcomes and may perpetuate symptoms via central sensitisation after structural repair [23].

Table 2. Comparative Clinical Presentation: Adults vs Children with SCDS

Feature	Adult Presentation	Paediatric Presentation
Dominant complaint	Mixed auditory–vestibular syndrome [1,2]	Hearing loss (conductive/mixed) [12]
Autophony	>50% present [1]	Less commonly reported [12]

Tullio phenomenon	Common; often presenting symptom [1]	Occasionally reported [12]
Hennebert sign	Common; elicitable on examination [1,15]	Less frequently documented [12]
Chronic disequilibrium	Frequent; significant morbidity [2]	Rare in young children [12]
Pulsatile tinnitus	Present in subset [18]	Occasionally reported [12]
Audiogram pattern	Low-frequency ABG; supranormal BC; reflexes intact [16,17]	Conductive, sensorineural, or mixed [12]
Surgical candidacy	Common when symptoms severe [4,35]	Reserved; observation preferred [13]

IV. Diagnostic Criteria

The Three-Pillar Diagnostic Framework

SCDS diagnosis requires the convergence of three independent lines of evidence: (1) characteristic clinical symptoms consistent with a third-window syndrome, (2) physiological evidence of an inner ear third window on audiometric and electrophysiological testing, and (3) anatomical confirmation of a superior canal dehiscence on high-resolution computed tomography [9]. All three elements should be present before a definitive diagnosis is assigned. Satisfying two out of three pillars warrants a provisional diagnosis and further investigation; a single positive element alone is insufficient [9].

This framework is critical because each pillar independently lacks perfect specificity. CT imaging reveals anatomical dehiscences in asymptomatic individuals (incidental findings, near-dehiscence artefacts). Physiological testing — particularly elevated VEMP amplitudes and low VEMP thresholds — can be seen in other third window disorders and in some normal variants. Clinical symptoms overlap substantially with Ménière's disease, perilymph fistula, and patulous Eustachian tube. Requiring all three pillars dramatically increases diagnostic specificity and reduces unnecessary surgical intervention [9,24].

Formal classification criteria for SCDS have not yet been published by the Bárány Society's International Classification of Vestibular Disorders (ICVD), in contrast to the detailed diagnostic criteria established for Ménière's disease, BPPV, and vestibular migraine under that framework [41]. This represents a significant gap in the international nosology of vestibular disease. The three-pillar framework described above represents the operative consensus used in expert centres worldwide, and fulfils the requirements of the diagnostic elements most commonly cited in the surgical literature [9,41].

□ **Key Point:** The three-pillar diagnostic requirement for SCDS — symptoms + physiology + imaging — is essential. CT alone is insufficient; anatomical dehiscence is present in 1–3% of the population, the majority asymptomatic. Physiological testing confirms the functional consequence of the dehiscence before surgical intervention is contemplated [9].

Near-Dehiscence and the Diagnostic Threshold

A diagnostically important concept is "near-dehiscence" — a condition where the superior canal roof is extremely thin (sub-millimetre) but not completely absent. Near-dehiscence may produce a partial third-window effect, explaining cases where symptoms and physiology are consistent with SCDS but CT appears equivocal [7,24]. High-resolution CT with Pöschl plane reformats at 0.5 mm or finer is required to reliably distinguish true dehiscence from near-dehiscence; cone-beam CT may further reduce partial volume averaging artefact [8]. Clinical decision-making in the near-dehiscence scenario requires particularly compelling physiological and symptomatic evidence before surgical intervention is recommended [24].

V. Investigations: Imaging, Audiometry, and Electrophysiology

High-Resolution CT of the Temporal Bone

Thin-slice (≤0.5 mm collimation) multidetector CT of the temporal bone is the gold standard imaging investigation for SCDS [9]. Images must be reformatted in two planes relative to the superior canal: the Pöschl plane (parallel to the superior canal, showing the canal en face) and the Stenver plane (orthogonal to the superior canal, showing the canal in cross-section) [9]. Imaging in only the axial plane substantially increases the probability of partial volume averaging artefact, either misrepresenting canal dehiscence or, conversely, missing a true defect [7,8]. The Massachusetts Eye and Ear classification system grades superior canal dehiscences by size and location along the arcuate eminence, with implications for surgical planning and surgical approach selection [4,8]. [43]

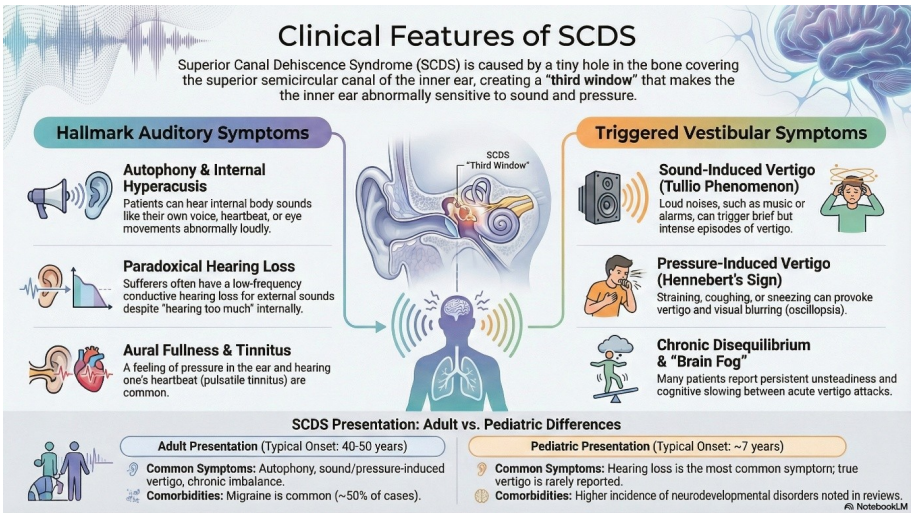

Figure 2. Massachusetts Eye and Ear CT classification of SCD. Left column (A–F): schematic location of superior canal defect. Right column: corresponding CT images in the Pöschl plane. Dehiscence size and location guide surgical approach selection.

Source: Eberhard KE, Chari DA, Nakajima HH, Kozin ED, Lee DJ. Front Neurol. 2021;12:638574. doi:10.3389/fneur.2021.638574.

Audiometry

Pure-tone audiometry in SCDS typically reveals a low-frequency air–bone gap (ABG) in the 250–1000 Hz range, commonly 10–40 dB [16,17]. This pattern mimics conductive hearing loss from middle ear pathology (otosclerosis, ossicular disruption), but the middle ear examination is normal in SCDS — intact tympanic membrane, type A tympanogram, and present acoustic reflexes [17]. The presence of an air–bone gap with normal acoustic reflexes is a key red flag for a third-window lesion and must prompt VEMP testing rather than proceeding directly to stapes surgery [9,17].

Bone conduction thresholds in SCDS may be genuinely supranormal — falling below 0 dB HL — particularly at 250 and 500 Hz. Standard clinical audiometry often does not test below 0 dB HL, and audiologists unfamiliar with SCDS may not recognise these values [16]. The Weber test with a 512 Hz fork placed on remote bones (the ankle, knee, or patella) lateralising to the affected ear is a useful and specific clinical finding in SCDS [2]. Any patient presenting with a low-frequency conductive hearing loss should have acoustic reflex testing before being directed to stapes surgery; if reflexes are normal, VEMP testing should follow before any middle ear intervention [9,17].

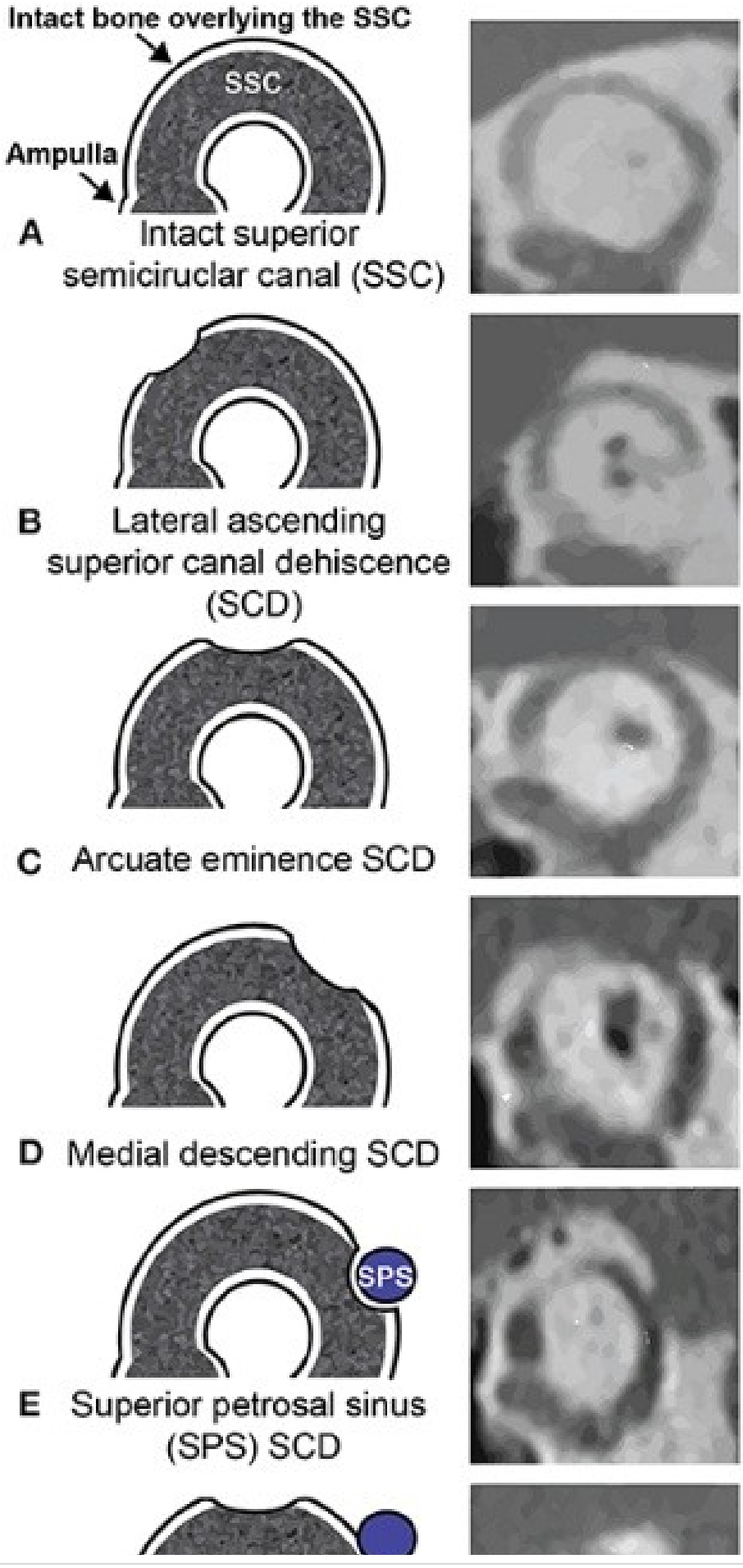
Intact bone overlying the SSC
SSC
Ampulla
A Intact superior semiciruclar canal (SSC)
B Lateral ascending superior canal dehiscence (SCD)
C Arcuate eminence SCD
D Medial descending SCD
SPS
E Superior petrosal sinus (SPS) SCD

Figure 3. Audiogram from a patient with SSCD before and after superior canal plugging surgery. (A) Preoperative: low-frequency air–bone gap up to 25 dB with supranormal bone conduction. (B) Postoperative: resolution of the air–bone gap; residual high-frequency sensorineural hearing loss at 8 kHz.

Source: Iversen MM, Rabbitt RD. Front Neurol. 2020;11:891. doi:10.3389/fneur.2020.00891.

Vestibular Evoked Myogenic Potentials (VEMPs)

VEMP testing has become central to the diagnosis of SCDS and is recommended in consensus guidelines as part of the standard workup for suspected third-window syndrome [9,26]. Two VEMP types are complementary in SCDS: the cervical VEMP (cVEMP), measuring saccule-mediated inhibition of the sternocleidomastoid muscle, and the ocular VEMP (oVEMP), measuring utricle-mediated excitation of the inferior oblique muscle. In SCDS, both are characteristically abnormal due to pathologically lowered vestibular thresholds produced by the third window [26,27].

Cervical VEMP thresholds in normal individuals require sound levels of 80–95 dB nHL to elicit a reproducible response. In SCDS, cVEMP thresholds are shifted to levels of 65–75 dB nHL or lower on the affected side [26]. A threshold of 70 dB nHL or less is highly suggestive of a third-window lesion [26,27]. Additionally, cVEMP amplitudes may be elevated, and an interpeak amplitude asymmetry ratio exceeding 50% is another useful metric. [44]

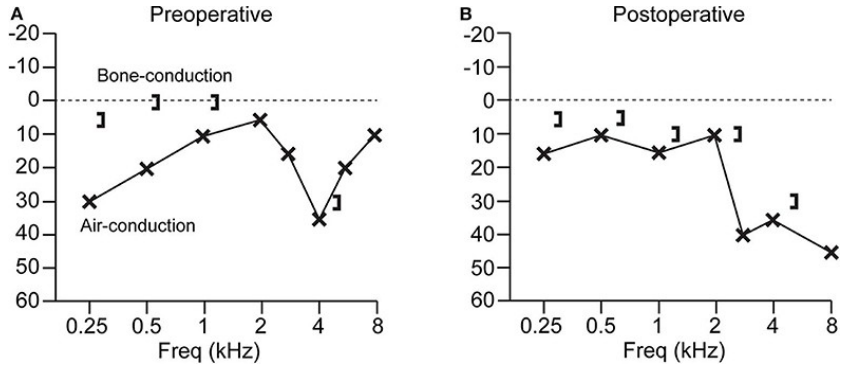

Figure 4. Cervical VEMP responses illustrating vestibular acoustic hypersensitivity in SSCD. Left ear: normal amplitude and threshold. Right ear: cVEMP threshold significantly reduced to 65 dB nHL, indicating pathological inner ear third-window hypersensitivity.

Source: Sauter TB. GN Otometrics; February 2009.

Ocular VEMP amplitude to 500 Hz tone burst or bone-conducted vibration is substantially elevated in SCDS on the affected side [27,28]. An oVEMP amplitude exceeding 17 µV (for a 500 Hz tone burst stimulus) or a significant asymmetry ratio favouring the affected ear is considered supportive of the diagnosis [27]. Bone-conducted vibration at the Fz midline forehead typically elicits asymmetric oVEMPs in SCDS, with the contralesional eye (receiving excitatory projection from the affected utricle via the third window) showing an enlarged n10 component [27,28].

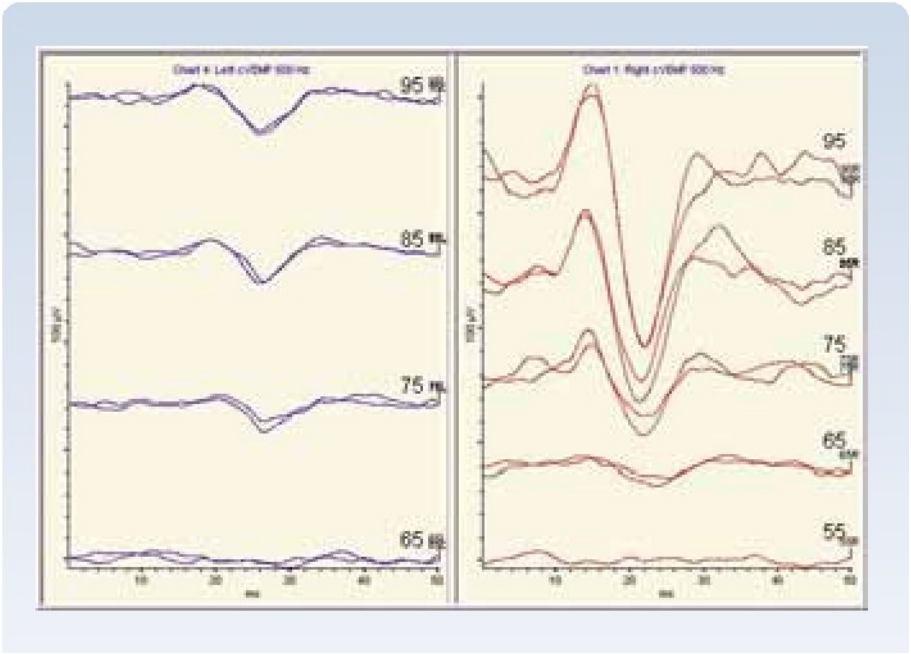

Figure 5. Ocular VEMP recordings to 500 Hz bone-conducted vibration. (a) Healthy subject: symmetric oVEMPs from bilateral infraorbital electrodes. (b) SCD patient: markedly asymmetric oVEMPs with enlarged contralesional n10, indicating exaggerated ipsilateral utricular response through the third window.
Source: Pastras CJ, Curthoys IS. Audiol Res. 2023;13(6):910–928. doi:10.3390/audiolres13060079.

Electrocochleography (ECochG)

Electrocochleography measures the ratio of the summating potential to the action potential (SP/AP ratio) as a marker of cochlear hydrodynamic abnormality. SCDS patients frequently exhibit elevated SP/AP ratios — classically associated with endolymphatic hydrops in Ménière's disease — because the altered perilymph mechanics created by the third window distort cochlear mechanics [29]. Importantly, SP/AP abnormalities in SCDS normalise after successful canal plugging surgery, confirming a direct link between the dehiscence and the electrocochleographic finding [29]. ECochG has been used intraoperatively as a real-time monitor of effective canal plugging [29].

Wideband Acoustic Immittance and Emerging Indices

Wideband acoustic immittance (WAI) measures acoustic energy absorbance across a broad frequency range. In ears with a third window, WAI demonstrates characteristic increased low-frequency absorbance (particularly around 0.5–1 kHz), reflecting shunting of low-frequency acoustic energy through the dehiscence rather than into the cochlea [30]. The "third window index" — a composite measure derived from the combination of low-frequency air–bone gap and cVEMP threshold — has been proposed as a quantitative tool to distinguish SCD ears from other third-window conditions [30]. These emerging metrics remain research tools at present but are increasingly available in specialist centres.

Table 3. Investigations in Suspected SCDS: Characteristic Findings and Clinical Significance

Investigation	Characteristic SCDS Finding	Clinical Significance
HRCT temporal bone (≤0.5 mm)	Absent bone over superior canal in Pöschl and Stenver views [9]	Anatomical pillar; mandatory for diagnosis
Pure-tone audiometry	Low-frequency ABG (250–1000 Hz); intact acoustic reflexes; supranormal BC [16,17]	Physiological pillar; ABG + reflexes = third window red flag
Tympanometry	Normal type A; no type As/B [17]	Excludes middle ear pathology
Acoustic reflexes	Present at normal levels [17]	Distinguishes from otosclerosis (key differentiator)
cVEMP threshold	Lowered (<70–75 dB nHL) on affected side [26]	High sensitivity for third window; supports diagnosis
oVEMP amplitude	Elevated (>17 µV) with asymmetry to affected side	High sensitivity and specificity in surgical series

	[27,28]	
Electrocochleography	Elevated SP/AP ratio; normalises post-plugging [29]	Supports diagnosis; intraoperative monitoring tool
Wideband acoustic immittance	Increased low-frequency absorbance [30]	Emerging; not yet standard of care
MRI temporal bone	Normal (cannot image cortical bone); excludes retrocochlear pathology [25]	Role in differential diagnosis workup

VI. Differential Diagnosis

SCDS has been described as a "great otologic mimicker" because its symptom complex overlaps substantially with several common and uncommon disorders of the ear and balance system [5,47]. Systematic application of the three-pillar diagnostic framework, combined with thorough understanding of each diagnostic mimicker, is essential to avoid the misdiagnosis that historically delayed treatment by years in many patients [5,9]. A structured multimodal diagnostic approach is essential to achieve the correct diagnosis before surgical planning [49,50].

Otosclerosis

Otosclerosis is the most clinically dangerous differential because both conditions produce a low-frequency conductive hearing loss on audiometry and may present in middle-aged individuals. The critical distinction is the acoustic reflex: otosclerosis abolishes or reduces stapedial reflexes because the stapes footplate is fixed, while SCDS preserves normal acoustic reflexes because the middle ear ossicular chain is intact [17]. Any patient presenting with a low-frequency conductive hearing loss should have acoustic reflex testing before being directed to stapes surgery; if reflexes are normal, VEMP testing must follow before any middle ear intervention is contemplated [9,17]. Patients with SCDS who were misdiagnosed as otosclerosis and underwent stapes surgery without relief are well described in the surgical literature [5].

Ménière's Disease

Ménière's disease produces episodic vertigo, fluctuating low-frequency sensorineural hearing loss, tinnitus, and aural fullness. Key distinguishing features from SCDS: Ménière's attacks typically last 20 minutes to 12 hours (SCDS episodes are seconds to minutes, time-locked to triggers); Ménière's does not produce sound-induced nystagmus; Ménière's audiogram shows a sensorineural rather than conductive pattern; VEMP findings may be abnormal in both but in opposite patterns [9,31]. Elevated SP/AP ratio is seen in both conditions, which can create diagnostic confusion, particularly in SCDS patients who have coincidental hydropic changes [29,31].

Patulous Eustachian Tube

Patulous Eustachian tube (PET) produces autophony and is the most commonly misdiagnosed alternative for the autophony symptom of SCDS. PET patients characteristically hear their own breathing (respiratory cycle) loudly, while SCDS patients do not [2]. PET autophony dramatically improves in the supine position (venous engorgement closes the tube), while SCDS autophony shows no consistent postural change. PET typically does not produce vertigo, audiometric air–bone gaps, or VEMP abnormalities [2].

Perilymph Fistula

Perilymph fistula (PLF) — abnormal communication between the perilymph space and the middle ear, typically through the oval or round window membranes — can produce pressure-induced vertigo and conductive hearing loss resembling SCDS [32]. Key distinguishing features: PLF does not produce the characteristic VEMP pattern of SCDS, audiometric air–bone gaps are not typical, CT is normal (no bony dehiscence), and the history usually includes an acute precipitating event (barotrauma, straining) [32].

Table 4. Differential Diagnosis: SCDS vs Key Mimickers

Condition	Key Differentiating Features	Definitive Test
Otosclerosis	Low-frequency ABG but ABSENT acoustic reflexes [17]; no Tullio/Hennebert	Acoustic reflex absent; CT: stapes footplate changes
Ménière's disease	Sensorineural HL; attacks 20	Glycerol test; MRI hydrops

	min–12 h; no sound-induced nystagmus [31]	protocol; VEMPs
Patulous Eustachian tube	Autophony of respiration; improves supine; no vertigo or ABG [2]	Tympanometry during forced respiration; nasopharyngoscopy
Perilymph fistula	Usually post-traumatic; no VEMP pattern; no bony defect on CT [32]	Exploratory tympanotomy (gold standard)
Enlarged vestibular aqueduct	Predominantly sensorineural HL in children; CT shows wide aqueduct [22]	HRCT/MRI: aqueduct diameter >1.5 mm
Vestibular migraine	Vertigo without sound/pressure trigger; phonophobia not autophony [23]	Clinical: ICHD-3 criteria; VEMP/CT normal
Anterior canal BPPV	Positional, not sound/pressure-triggered; resolves in seconds [33]	Dix-Hallpike: downbeating nystagmus, fatigable
Lateral canal dehiscence	Can mimic SCDS; check lateral canal on CT [46]	HRCT with multiplanar reformats; VEMP pattern

□ **Important:** Misdiagnosis of SCDS as otosclerosis, with subsequent stapes surgery, is a well-documented and avoidable harm. Any low-frequency conductive hearing loss with intact acoustic reflexes mandates VEMP testing and temporal bone CT before any middle ear surgical intervention is scheduled [9,17].

VII. Conservative and Medical Management

Management of SCDS is individualised and follows a stepwise approach that begins with conservative measures for all patients, escalating to surgery only in those with significant functional impairment and a compelling risk–benefit ratio. A proportion of patients will elect to manage conservatively for extended periods — or indefinitely [1,2,34]. Co-existing conditions (migraine, anxiety, Eustachian tube dysfunction) should be optimised in parallel, as they substantially modulate the patient's experience of SCDS [2,23].

Patient Education and Trigger Avoidance

Education is the cornerstone of initial management. Patients benefit from understanding the third-window mechanism — that their symptoms are caused by a structural defect with a known physiological explanation rather than a psychological or functional disorder. This framing can substantially reduce health anxiety and improve coping [2]. Trigger identification and avoidance provides direct symptom reduction: patients are guided to identify specific sounds, physical manoeuvres, and environments that precipitate symptoms, and to modify their exposure. Custom hearing protection is frequently useful in noisy environments [2,34].

Activity modification — avoiding Valsalva manoeuvres, heavy lifting, straining, and activities that raise intracranial pressure — reduces the frequency and severity of pressure-induced vestibular episodes. Patients should be counselled about open-mouth sneezing (reduces intracranial pressure spike), toilet posture, and exercise modification. High-impact exercise may need to be replaced with lower-impact alternatives during periods of symptom severity [34].

Vestibular Rehabilitation

Vestibular rehabilitation therapy (VRT) does not address the underlying anatomical defect and does not eliminate sound- or pressure-triggered symptoms. However, it has a meaningful role in managing the chronic disequilibrium and functional impairment that characterise many SCDS patients outside of their acute episodes [34]. VRT addresses gaze stabilisation, postural control, and central vestibular compensation through structured exercise programmes. It is also critically important in the post-surgical rehabilitation phase, supporting recovery of vestibular compensation after intentional canal occlusion [34,39].

Management of Comorbidities

Vestibular migraine co-occurs in approximately 50% of SCDS patients in some series and requires active identification and treatment [2,23]. Standard migraine preventive and acute therapies should be initiated or optimised before surgical SCDS intervention is considered: dietary modification, sleep hygiene, adequate hydration, and pharmacological prophylaxis (amitriptyline, topiramate, propranolol, nortriptyline, or CGRP pathway agents) [23]. Uncontrolled migraine is a predictor of suboptimal post-surgical symptom resolution, as migraine-related central sensitisation may perpetuate vestibular symptoms independently of the structural defect [23].

Anxiety and depression are common in SCDS patients after years of chronic symptoms and diagnostic uncertainty. Cognitive behavioural therapy for vestibular symptom-related anxiety, combined with appropriate pharmacological support when indicated, should be offered as part of a holistic management plan [2]. Addressing psychological comorbidity both improves quality of life and increases the probability of a satisfactory response to surgical intervention when that step is taken.

Hearing Aids and Amplification

The low-frequency conductive hearing loss of SCDS may respond to conventional hearing amplification in patients who are not surgical candidates or who decline surgery. However, amplification of external sounds in SCDS patients already sensitised to internal sounds requires careful fitting to avoid aggravating autophony or hyperacusis [2]. Some patients find that partially occluding the ear canal (with an earmould or earplug) reduces autophony by increasing impedance in the external canal — an inexpensive test of conservative symptomatic management that can be performed in clinic before formal audiological review [2,34].

VIII. Surgical Management and Emerging Techniques

Indications for Surgery

Surgical intervention is appropriate for patients with: (1) confirmed SCDS by all three diagnostic pillars, (2) significant functional impairment from vestibular symptoms incompletely controlled by conservative measures, (3) acceptable anaesthetic and surgical risk, and (4) realistic expectations about surgical outcomes [4,35]. Vestibular predominance — particularly debilitating Tullio phenomenon and Hennebert sign — is generally a stronger indication than isolated auditory symptoms. The patient's individual anatomy, bilateral disease status, and the experience of the operating surgeon with each approach must all inform the choice of technique [35].

Bilateral SCDS is present in approximately 25% of patients. Surgical management of bilateral disease is staged: the more symptomatic side is operated upon first and an adequate recovery period (typically 6–12 months) is observed before considering contralateral surgery. The symptomatic side is determined by lateralisation of VEMP findings, direction of sound/pressure-induced nystagmus, and the patient's own symptom attribution [4,35].

Middle Cranial Fossa Approach (MCF)

The middle cranial fossa craniotomy approach is the original surgical technique described by Minor and remains the approach with the highest reported rates of complete vestibular symptom resolution and the most extensive published follow-up data [1,4,35]. The procedure involves a temporal craniotomy above the ear, gentle temporal lobe elevation to expose the floor of the middle cranial fossa and the arcuate eminence, and direct visualisation of the superior canal dehiscence under an operating microscope [4,35].

Two operative techniques can be applied through the MCF approach: canal plugging and canal resurfacing (capping). Canal plugging introduces autologous material (temporalis fascia, bone wax, hydroxyapatite cement, or bone dust with fascia) into the lumen of the superior canal at the dehiscence, eliminating endolymph movement in the canal [4,35,36]. Resurfacing covers the dehiscence from above with fascia, bone chips, or bone cement without entering the canal lumen [36]. Published series suggest plugging produces higher rates of vestibular symptom resolution, while resurfacing may be preferable when preservation of residual superior canal function is a priority — though both approaches significantly impair superior canal function to some degree [36].

Transmastoid Approach

The transmastoid approach performs SCDS repair through the mastoid cavity without intracranial entry. The superior canal is identified within the mastoid, and the dehiscent segment is plugged or resurfaced via the posterior route [35,37]. The primary advantage is avoidance of temporal lobe retraction and intracranial risk (meningitis, temporal lobe oedema, seizure) [37]. The transmastoid approach is particularly suited to laterally located dehiscences. A practical disadvantage is that the dehiscence cannot be directly visualised (as it opens toward the cranial fossa, not the mastoid), requiring intraoperative navigation to confirm adequate repair [37]. Outcomes are generally equivalent for vestibular symptoms in experienced hands.

Round Window Reinforcement (RWR)

Round window reinforcement — the application of a fascia or fat graft over the round window membrane via a transcanal approach — has been proposed as a less invasive alternative for SCDS [38]. The theoretical mechanism is that reinforcing the round window reduces its compliance, partially restoring the two-window balance and reducing the functional impact of the third window without directly addressing the dehiscence [38]. Early reports showed some benefit in vestibular symptoms, but longer-term data are inconsistent. Current evidence suggests it may be most appropriate as a first surgical option for patients with mild-to-moderate vestibular symptoms who are reluctant to undergo craniotomy, with MCF or transmastoid approaches reserved if RWR fails [38].

Table 5. Summary of Surgical Approaches and Techniques for SCDS Repair

Approach	Technique	Key Advantages	Key Disadvantages	Best Indication
Middle cranial fossa (MCF)	Plugging or resurfacing via temporal craniotomy	Direct dehiscence visualisation; highest vestibular symptom resolution rates [4,35]	Intracranial risks (temporal lobe retraction, meningitis, seizure); longer recovery	Medially located defects; standard first approach for most centres
Transmastoid	Plugging via mastoid; no intracranial entry	Avoids intracranial exposure; lower risk; shorter hospital stay [37]	No direct visualisation; requires intraoperative imaging; less hearing data	Laterally located defects; high anaesthetic risk patients
Round window reinforcement	Fascia/fat over round window membrane; middle ear only	No intracranial entry; lower risk; reversible [38]	Lower vestibular symptom resolution; unclear mechanism; limited evidence [38]	Mild-moderate symptoms; poor surgical candidates; trial before definitive surgery

□ **Clinical Insight:** Canal plugging via either MCF or transmastoid approaches effectively eliminates superior canal function on the operated side. Patients should be counselled that they will lose superior canal VOR responses on that side and that vestibular rehabilitation is standard and essential for optimal recovery. The contralateral vestibular system and visual/somatosensory systems compensate adequately in the great majority of patients within 6–12 months [35,39].

IX. Prognosis, Recurrence, and Special Populations

Surgical Outcomes for Vestibular Symptoms

Published series consistently demonstrate good overall outcomes after canal plugging via MCF or transmastoid approaches. Vestibular symptoms — particularly Tullio phenomenon and Hennebert sign — respond most reliably, with complete or substantial resolution in 75–95% of patients in the larger case series [4,35,36,39]. Chronic disequilibrium is more variable: it resolves completely in approximately 60–70% of patients, with some residual unsteadiness attributable to the loss of ipsilateral superior canal

function requiring central compensation [35,39]. Autophony and internal hyperacusis improve in 70–85% of surgically treated patients [35,36].

Surgical Outcomes for Hearing

Hearing outcomes after surgery are generally positive for the conductive component: the audiometric air–bone gap resolves or substantially reduces in most patients following successful canal plugging, consistent with elimination of the third-window shunting effect [4,35,36]. However, a small but meaningful risk of sensorineural hearing loss exists as a surgical complication: high-frequency SNHL — most commonly at 8 kHz — is reported in approximately 5–10% of surgeries, reflecting proximity of the cochlear duct to the operative field and potential for acoustic trauma during drilling [35,36]. Profound deafness or dead ear is a rare but described complication (<2% in experienced hands) and must be explicitly discussed during informed consent [35].

Formal quality of life assessments using validated instruments (DHI, SF-36) document significant improvements post-operatively in the domains most affected by SCDS — vestibular function, hearing, and daily activity limitations [39,40]. Patient satisfaction scores are high in the majority of published series, with most patients indicating they would choose surgery again given the same circumstances. Outcomes are most favourable when pre-surgical symptom severity was high, when all three diagnostic pillars were confirmed, and when comorbid migraine was appropriately managed perioperatively [35,39].

Recurrence and Graft Failure

Graft failure or recurrence after surgical repair is uncommon but documented. Displacement or absorption of plug material can allow re-canalisation of the superior canal, producing return of symptoms. Reports suggest recurrence rates of approximately 3–8% over medium-term follow-up [35,36]. Revision surgery via the same or alternative approach is technically feasible but involves greater risk due to scarring and altered anatomy. Intraoperative ECochG monitoring may reduce the risk of incomplete plugging by providing real-time confirmation of functional canal occlusion [29].

Bilateral Disease

Patients with bilateral SCDS managed surgically for the more symptomatic side generally have favourable outcomes, with improvement in overall vestibular function despite functional loss of one superior canal [35]. Decision-making regarding contralateral surgery must weigh residual symptoms attributable to the unoperated side (confirmed by physiological testing and symptom lateralisation), the risk of losing bilateral superior canal function, and the patient's overall health status [35,4]. Contralateral surgery is not routinely performed prophylactically; it is reserved for cases where physiological testing confirms significant contralateral third-window effect and symptoms are clearly attributable to the unoperated ear [4].

Paediatric Outcomes

Surgical outcomes in children with symptomatic SCDS are limited by small case numbers. Conservative management is preferred in most paediatric cases given the risk of spontaneous ossification with skeletal maturation and the higher relative risk of craniotomy in young children [12,13]. When surgery is performed in children with significant functional impairment that fails conservative management, reported outcomes are generally positive and comparable to adult series. Post-operative rehabilitation is particularly important in children to support vestibular compensation and minimise academic and developmental impact [13].

X. Guidelines, Controversies, and Future Directions

Current Guideline Status

There are no internationally adopted clinical practice guidelines specifically for SCDS from the major vestibular and otolaryngological societies (AAO-HNSF, Bárány Society) as of 2026 [9,41]. The closest framework is the consensus guidance on VEMP indications, which includes recommendations for VEMP testing in suspected third-window lesions [26]. Most clinical guidance is derived from expert opinion, single-centre case series, and systematic reviews rather than randomised controlled trials — a significant evidence gap given the surgical nature of definitive treatment. The International Classification of Vestibular Disorders (ICVD) under the Bárány Society has not yet published formal diagnostic classification criteria for SCDS, in contrast to the detailed criteria established for BPPV, Ménière's disease, and vestibular migraine [41].

Near-Dehiscence Controversy

One of the most clinically contested areas is the management of "near-dehiscence" — cases where the superior canal roof is extremely thin but not demonstrably absent on CT, yet the patient has symptoms and physiological findings consistent with SCDS [7,24]. A subset of these patients appears to have a genuine partial third-window effect from the hyperthin bone, and some respond to surgical plugging. However, the diagnostic threshold is not standardised, the proportion who will develop complete dehiscence is unknown, and operating on the basis of thin bone alone risks unnecessary intervention [7,24]. Cone-beam CT with sub-millimetre resolution and 3D rendering is being evaluated as a more precise characterisation tool [8].

Plugging vs Resurfacing

The relative merits of canal plugging vs resurfacing (capping) remain debated. Plugging provides more reliable vestibular symptom resolution by definitively eliminating endolymph movement in the operated canal but also guarantees permanent loss of superior canal VOR function on that side [36]. Resurfacing theoretically preserves some canal function but shows lower rates of complete vestibular symptom resolution in comparative series [36]. Some surgeons advocate a resurfacing-first approach to minimise functional loss, with plugging reserved for failure. No randomised trial has directly compared the two approaches.

Round Window Reinforcement: Legitimate Alternative or Temporising Measure?

Whether round window reinforcement is a legitimate alternative to canal plugging or merely a temporising measure is contested [38]. Proponents note its significantly lower risk profile and reversibility; critics argue its mechanism is unclear, its outcomes are inferior, and that it may delay definitive treatment. The optimal patient selection criteria for RWR — if any — remain to be defined prospectively [38].

Future Directions

Several research directions hold promise for improving SCDS diagnosis, non-invasive management, and surgical outcomes. Ultra-high-resolution photon-counting CT is being evaluated to better characterise canal wall thickness and distinguish true dehiscence from near-dehiscence [8]. Expanded VEMP normative datasets across age groups and stimulus parameters will refine the sensitivity and specificity of electrophysiological diagnosis [27,28]. Machine learning applied to temporal bone CT imaging is being developed to detect sub-threshold thinning and predict which near-dehiscences are physiologically active [8].

Pharmacological approaches to SCDS are not yet established. Genetic studies are advancing toward identifying susceptibility variants affecting bone mineralisation and otic capsule formation, which could eventually yield targets for prevention in predisposed individuals [20]. Intraoperative monitoring with ECochG and electrophysiological feedback is advancing toward real-time confirmation of adequate canal plugging, potentially reducing the reoperation rate from incomplete plugging [29]. The most impactful near-term advance for SCDS management, however, is likely the systematic adoption of the three-pillar diagnostic framework in all clinicians who assess patients with unexplained conductive hearing loss or pressure/sound-induced vestibular symptoms, eliminating the diagnostic delay that currently averages 3–5 years in many published series [5,9].

□ **Clinical Pearl:** The most impactful near-term advance for SCDS management is the systematic adoption of the three-pillar diagnostic framework (symptoms + physiology + imaging) in all clinicians who assess patients with unexplained conductive hearing loss or pressure/sound-induced vestibular symptoms. Eliminating the diagnostic delay — currently averaging 3–5 years in many published series — will have a larger impact on patient outcomes than any single technical advance in surgery or imaging [5,9].

References

[1] Minor LB, Solomon D, Zinreich JS, Zee DS. Sound- and/or pressure-induced vertigo due to bone dehiscence of the superior semicircular canal. Arch Otolaryngol Head Neck Surg. 1998;124(3):249–258.

[2] Minor LB. Superior canal dehiscence syndrome. Am J Otol. 2000;21(1):9–19.

[3] Cawthorne T. Fenestration of the horizontal semicircular canal. Proc R Soc Med. 1947;40(4):193–195.

[4] Minor LB, Carey JP, Cremer PD, Lustig LR, Streubel SO, Ruckenstein MJ. Dehiscence of bone overlying the superior canal as a cause of apparent conductive hearing loss. Otol Neurotol. 2003;24(2):270–278.

[5] Ward BK, Carey JP, Minor LB. Superior canal dehiscence syndrome: lessons from the first 20 years. Front Neurol. 2017;8:177.

[6] Carey JP, Minor LB, Nager GT. Dehiscence or thinning of bone overlying the superior semicircular canal in a temporal bone survey. Arch Otolaryngol Head Neck Surg. 2000;126(2):137–147.

[7] Williamson RA, Vrabec JT, Coker NJ, Sanderson A. Coronal computed tomography prevalence of superior semicircular canal dehiscence. Otolaryngol Head Neck Surg. 2003;129(5):481–489.

[8] Lookabaugh S, Kelly HR, Carter MS, et al. Radiologic classification of superior canal dehiscence: implications for surgical repair. Otol Neurotol. 2015;36(1):118–125.

[9] Ward BK, Agrawal Y, Nguyen E, et al. Hearing outcomes after surgical treatment of superior semicircular canal dehiscence. Otol Neurotol. 2012;33(2):264–270.

[10] Hirvonen TP, Weg N, Zinreich SJ, Minor LB. High-resolution CT findings suggest a developmental abnormality underlying superior canal dehiscence syndrome. Acta Otolaryngol. 2003;123(4):477–481.

[11] Mong A, Bhati MT, Booth TN, et al. Prevalence of superior semicircular canal dehiscence on CT in pediatric patients. Pediatr Radiol. 2019;49(2):218–225.

[12] Kim HH, Chung MK, Wiet RJ. Superior semicircular canal dehiscence syndrome: review and cases in pediatric patients. Otol Neurotol. 2011;32(7):1179–1182.

[13] Teixido M, Manoukian J, Haber J. Pediatric SSCD — indications for surgical repair. Int J Pediatr Otorhinolaryngol. 2015;79(4):498–502.

[14] Merchant SN, Rosowski JJ. Conductive hearing loss caused by third-window lesions of the inner ear. Otol Neurotol. 2008;29(3):282–289.

[15] Rosowski JJ, Songer JE, Nakajima HH, Brinsko KM, Merchant SN. Clinical, experimental, and theoretical investigations of the effect of superior semicircular canal dehiscence on hearing mechanisms. Otol Neurotol. 2004;25(3):323–332.

[16] Halmagyi GM, McGarvie LA, Aw ST, et al. The click-evoked vestibulo-ocular reflex in superior semicircular canal dehiscence. Neurology. 2003;60(7):1172–1175.

[17] Belden CJ, Weg N, Minor LB, Zinreich J. CT evaluation of bone dehiscence of the superior semicircular canal as a cause of sound- and/or pressure-induced vertigo. Radiology. 2003;226(2):337–343.

[18] Sismanis A, Stamm MA, Sobel M. Pulsatile tinnitus as a symptom of superior semicircular canal dehiscence. Otolaryngol Head Neck Surg. 2009;141(3):403–406.

[19] Songer JE, Rosowski JJ. A mechano-acoustic model of the effect of superior canal dehiscence on hearing in chinchilla. J Acoust Soc Am. 2007;122(2):943–951.

[20] Castellucci A, Brandolini C, Ferri GG, Ghidini A, Presutti L. CDH23 gene mutation in superior canal dehiscence syndrome. Audiol Neurootol. 2015;20(4):246–254.

[21] Nadgir RN, Ozonoff A, Devaiah AK, Sakai O. Superior semicircular canal dehiscence: congenital or acquired condition? AJNR Am J Neuroradiol. 2011;32(5):947–949.

[22] Friedland DR, Runge-Samuelson C. Softening of the otic capsule: implications for the pathogenesis of enlarged vestibular aqueduct. Ann Otol Rhinol Laryngol. 2009;118(7):519–524.

[23] Boleas-Aguirre MS, Lin FR, Carey JP, Minor LB. Vestibular-evoked myogenic potentials with air-conducted tones in patients with superior semicircular canal dehiscence. Acta Otolaryngol. 2007;127(12):1218–224.

[24] Zuniga MG, Janky KL, Nguyen KD, Carey JP, Zuniga MG. Ocular versus cervical VEMPs in the diagnosis of superior semicircular canal dehiscence syndrome. Otol Neurotol. 2013;34(1):121–126.

[25] Brantberg K, Bagger-Sjöbäck D, Mathiesen T, et al. Posterior canal dehiscence syndrome caused by an apex cholesteatoma. Otol Neurotol. 2006;27(4):531–534.

[26] Rosengren SM, Welgampola MS, Colebatch JG. Vestibular evoked myogenic potentials: past, present and future. Clin Neurophysiol. 2010;121(5):636–651.

[27] Zuniga MG, Janky KL, Nguyen KD, Carey JP. Can vestibular-evoked myogenic potentials help differentiate Ménière disease from superior semicircular canal dehiscence? Otolaryngol Head Neck Surg. 2012;146(5):788–796.

[28] Pastras CJ, Curthoys IS. Vestibular Testing — New Physiological Results for the Optimisation of Clinical VEMP Stimuli. Audiol Res. 2023;13(6):910–928.

[29] Mikulec AA, Poe DS, McKenna MJ. Operative management of superior semicircular canal dehiscence. Laryngoscope. 2005;115(3):501–507.

[30] Merchant GR, Roosli C, Niesten ME, et al. Power reflectance as a screening tool for the diagnosis of superior semicircular canal dehiscence. Otol Neurotol. 2015;36(1):172–177.

[31] Crane BT, Minor LB, Carey JP. Superior canal dehiscence plugging reduces dizziness handicap. Laryngoscope. 2008;118(10):1809–814.

[32] Hornibrook J. Perilymph fistula: fifty years of controversy. ISRN Otolaryngol. 2012;2012:281248.

[33] Büttner U, Helmchen C, Brandt T. Diagnostic criteria for central versus peripheral positioning nystagmus and vertigo: a review. Acta Otolaryngol. 1999;119(1):1–5.

[34] Cloutier JF, Belair M, Saliba I. Superior semicircular canal dehiscence: positive predictive value of high-resolution CT scanning. Eur Arch Otorhinolaryngol. 2008;265(12):1455–1460.

[35] Agrawal Y, Minor LB, Schubert MC, Janky K, Davalos-Bichara M, Carey JP. Second-side surgery in superior canal dehiscence syndrome. Otol Neurotol. 2012;33(6):1013–1018.

[36] Cheng YS, Kozin ED, Lee DJ. Transmastoid repair of superior semicircular canal dehiscence. Otolaryngol Clin North Am. 2018;51(2):407–421.

[37] Carter MS, Lookabaugh S, Lee DJ. Endoscopic-assisted repair of superior canal dehiscence syndrome. Laryngoscope. 2014;124(6):1464–1468.

[38] Silverstein H, Van Ess MJ. Complete round window niche occlusion for superior semicircular canal dehiscence syndrome: a relative indication. Ear Nose Throat J. 2009;88(8):1042–1056.

[39] Agrawal Y, Migliaccio AA, Minor LB, Carey JP. Vestibular hypofunction in the initial postoperative period after surgical treatment of superior semicircular canal dehiscence. Otol Neurotol. 2009;30(4):502–506.

[40] Goddard JC, Schwartz MS, Friedman RA. Bone island or superior semicircular canal dehiscence? Otol Neurotol. 2006;27(6):862–866.

[41] Lempert T, von Brevern M. The International Classification of Vestibular Disorders. Neurol Clin. 2015;33(3):541–550.

[42] Iversen MM, Rabbitt RD. Biomechanics of third window syndrome. Front Neurol. 2020;11:891.

[43] Eberhard KE, Chari DA, Nakajima HH, Kozin ED, Lee DJ. Current trends, controversies, and future directions in the evaluation and management of superior canal dehiscence syndrome. Front Neurol. 2021;12:638574.

[44] Sauter TB. Cervical and Ocular Vestibular Evoked Myogenic Potentials (VEMPs) in Vestibular Hypersensitivity. Insights in Practice. GN Otometrics; 2009.

[45] Kaski D, Davies R, Luxon L, Bronstein AM, Rudge P. The Tullio phenomenon: a neurologically neglected presentation. J Neurol. 2012;259(1):4–21.

[46] Hullar TE, Bhatt KA, Janky KL, et al. Lateral canal dehiscence as a cause of hyperacusis, pulsatile tinnitus, and autophony. Otol Neurotol. 2012;33(7):1235–1238.

[47] Banerjee A, Whyte A, Atlas MD. Superior canal dehiscence: review of a new condition. Clin Otolaryngol. 2005;30(1):9–15.

[48] Schuknecht HF, Reisser C. The morphological basis for perilymphatic gushers and oozers. Adv Otorhinolaryngol. 1988;39:1–12.

[49] Nikles S, Grossmann W, Almeida C, Pedrosa Carrasco A, Heyse Freiherr von Twickel J. Superior canal dehiscence syndrome: from diagnosis to therapy. HNO. 2018;66(S1):71–77.

[50] Brookes GB, Maw AR, Coleman MJ. Superior canal dehiscence syndrome as an otological mimic. J Laryngol Otol. 2011;125(10):999–1004.

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