Vestibular Schwannoma

Vestibular Physician Literature Review

A comprehensive evidence-based review for vestibular physicians, neuro-otologists, advanced ENT trainees, and senior vestibular physiotherapists.

Vestibular Medicine for Vestibular Physicians

Peripheral Vestibular Pathology — Module 2.6

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How to Use This Document

This literature review is designed for vestibular physicians, neuro-otologists, advanced ENT trainees, and senior vestibular physiotherapists. It provides a deep-dive evidence synthesis spanning pathophysiology, diagnostic evaluation, grading, and multidisciplinary management of vestibular schwannoma (VS). Each section is referenced using Vancouver style with inline citations [N]. All recommendations are graded by evidence level where applicable.

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 and an audio podcast series available via the Australian Dizziness Clinics education hub at www.AustralianDizzinessClinics.com.

Callout Box Guide

□ **Key Point:** Highlights a core clinical fact or principal finding central to understanding this condition.

□ **Clinical Insight:** Provides deeper mechanistic or investigational context to enrich clinical reasoning.

□ **Clinical Pearl:** A practical tip or pattern-recognition point that has direct value at the bedside or in clinic.

□ **Important:** Flags a high-stakes pitfall, red flag, or common error that can lead to patient harm if missed.

□ **Key Point:** Vestibular schwannoma is the most common tumour of the cerebellopontine angle (CPA), accounting for approximately 80–90% of CPA masses. Timely diagnosis and individualised management are essential to preserve function and quality of life [1].

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I. Epidemiology and Aetiology

Vestibular schwannoma (VS), historically termed acoustic neuroma, is a benign Schwann-cell-derived neoplasm arising from the vestibular division of the eighth cranial nerve (vestibulocochlear nerve). It is the most common tumour of the internal auditory canal (IAC) and cerebellopontine angle (CPA), accounting for approximately 80–90% of CPA masses [1,2]. The annual incidence is approximately 1–2 per 100,000 population, though population-based MRI studies suggest true incidence may be higher due to asymptomatic tumours [3].

The median age at diagnosis is 50–55 years, with a slight female predominance [4]. Sporadic unilateral VS comprises approximately 95% of cases. The remaining 5% are associated with neurofibromatosis type 2 (NF2), an autosomal dominant tumour suppressor syndrome characterised by bilateral VS, meningiomas, ependymomas, and ocular abnormalities [5]. Sporadic VS has no established environmental aetiology; although early mobile phone studies raised concern about radiofrequency exposure, current evidence does not support a causal relationship [6,7].

Approximately 25–30% of VS demonstrate no growth on surveillance, 50–60% grow slowly (median 1–2 mm/year), and 10–15% exhibit accelerated growth requiring intervention [8]. This biological heterogeneity underpins the individualised management paradigm — observation, radiosurgery, or microsurgery — that governs contemporary vestibular schwannoma care [9].

□ **Clinical Insight:** The term 'acoustic neuroma' is a misnomer — these tumours arise from vestibular Schwann cells, not cochlear neurons. 'Vestibular schwannoma' is the preferred terminology in contemporary practice [1].

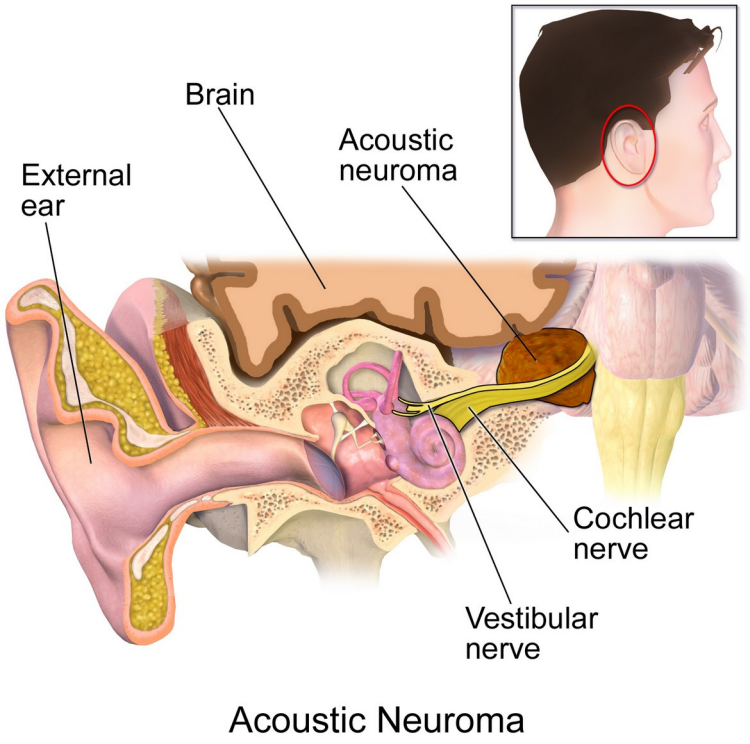

Figure 1A. Three-dimensional anatomical illustration showing a vestibular schwannoma (acoustic neuroma) arising from the vestibular nerve within the internal auditory canal and expanding into the cerebellopontine angle.
Source: BruceBlaus. Blausen Medical, 2013. Wikimedia Commons. CC BY 3.0.

II. Pathophysiology and Molecular Biology

VS arises from Schwann cells of the vestibular nerve, most commonly the superior division. The tumour is invariably benign (WHO Grade I) and does not undergo malignant transformation in sporadic cases [10]. Histologically, VS exhibits two classical tissue patterns: Antoni A tissue (densely packed bipolar

spindle cells with nuclear palisading into Verocay bodies) and Antoni B tissue (loose, disorganised hypocellular myxoid stroma with lipid-laden cells) [11]. Larger tumours typically display a mixture of both patterns.

The NF2 gene on chromosome 22q12 encodes merlin (schwannomin), a tumour suppressor protein that regulates cell proliferation through interaction with the mTOR, Ras/MAPK, PI3K/Akt, and Hippo signalling pathways [12]. Loss-of-function mutations in NF2 underlie virtually all VS — both NF2-associated (germline mutation + loss of heterozygosity) and sporadic (two somatic hits, consistent with Knudson's two-hit hypothesis) [13]. The degree of merlin dysfunction correlates with tumour growth behaviour, with lower merlin expression associated with more aggressive growth [14].

VS induces progressive vestibular deafferentation via two mechanisms: compressive ischaemia of the internal auditory artery (leading to sudden-onset hearing loss in up to 10% of patients) and chronic secretion of glutamate, S100 protein, and other neurotoxic factors that damage spiral ganglion neurons and hair cells [15]. Central vestibular compensation partially corrects the unilateral peripheral vestibular deficit through cerebellar-mediated plasticity, which explains the relatively mild vestibular symptoms in many patients despite significant canal paresis on caloric testing [16].

□ **Clinical Pearl:** Merlin regulates contact-dependent inhibition of proliferation. Loss of merlin function activates mTOR signalling — a therapeutic target currently under investigation with everolimus in NF2-associated schwannomas [12,13].

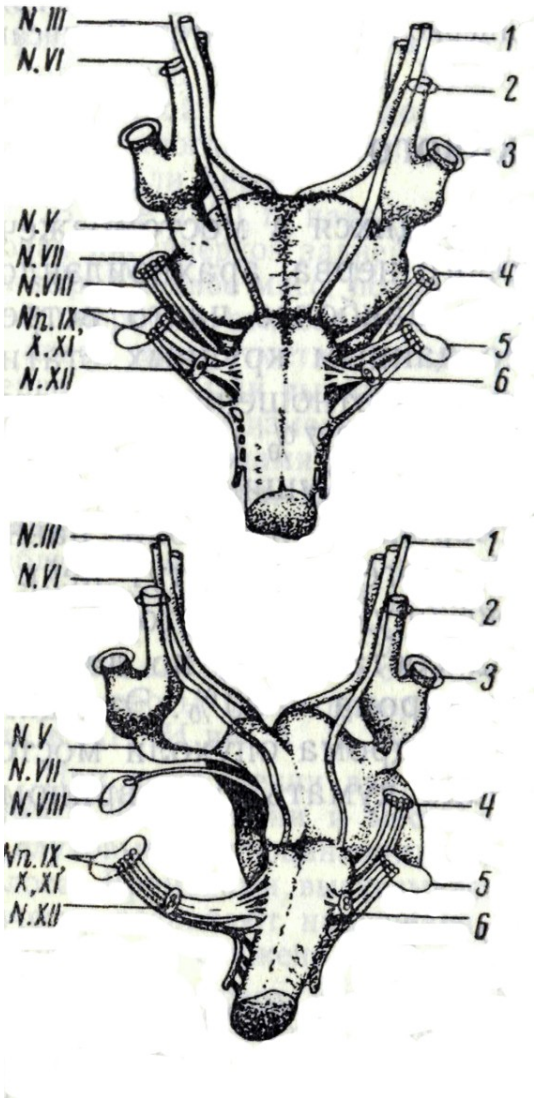

Figure 1B. Historical surgical illustration of vestibular schwannoma topography, demonstrating the tumour's relationship to the cerebellopontine angle, posterior cranial fossa, and adjacent cranial nerves. Note the displacement of the facial nerve and brainstem in larger tumours.
Source: Harvey Cushing (1869–1939). Published in Razdolsky IA, 'Neurinoma of the Auditory Nerve', 1936. Public domain (author died 1939).

III. Clinical Presentation

The clinical presentation of VS is characterised by insidious onset of unilateral audiovestibular symptoms, often with a prolonged pre-diagnostic interval of 3–7 years [17]. The classic triad of unilateral sensorineural hearing loss (SNHL), tinnitus, and disequilibrium reflects compression of cranial nerve VIII and its vascular supply within the IAC.

Auditory Features

Asymmetric SNHL is the most common presenting feature, occurring in approximately 95% of patients with symptomatic VS [18]. The hearing loss is typically high-frequency and slowly progressive, though sudden SNHL (occurring over 72 hours) is the presenting event in 10–20% of cases and may mislead clinicians into diagnosing idiopathic sudden SNHL without MRI evaluation [19]. Rollover on speech discrimination testing — a paradoxical decrease in speech recognition at high sound pressure levels — is a distinctive, though not universal, feature of retrocochlear pathology [20]. Unilateral tinnitus, typically high-pitched, is present in 70–80% of patients and may precede hearing loss by months to years [18].

Vestibular Features

Acute vertigo is uncommon as the primary presentation of VS, occurring in fewer than 20% of patients [21]. The more typical vestibular presentation is chronic unsteadiness and disequilibrium — a reflection of the slow, partial central compensation that occurs in response to progressive unilateral deafferentation. Paroxysmal positional vertigo may coexist in approximately 25% of VS patients as a secondary phenomenon, likely reflecting utricular dysfunction causing canalith displacement [22]. Oscillopsia during head movement is present when canal paresis is severe and compensation incomplete.

Neurological Features

Facial nerve involvement, though anatomically adjacent, is clinically rare in sporadic VS — occurring in fewer than 10% of untreated patients — owing to the nerve's resilience to stretch rather than compression [23]. In contrast, trigeminal (V3) sensory changes, particularly facial numbness or hypoaesthesia, indicate tumours with significant CPA extension compressing the trigeminal nerve. Large Koos IV tumours may produce hydrocephalus, cerebellar ataxia, and raised intracranial pressure symptoms (headache, papilloedema), reflecting brainstem compression [24].

□ **Important:** Any patient presenting with unilateral sudden SNHL should be offered MRI with gadolinium to exclude VS. Studies show VS underlies 3–11% of sudden SNHL presentations — a proportion frequently missed when MRI is deferred [19]. Early diagnosis significantly expands the management options available.

IV. Diagnostic Investigations

The diagnostic workup for VS integrates audiological, vestibular, and neuroimaging assessments. MRI with gadolinium is the gold standard for diagnosis; the functional tests inform anatomical localisation, deafferentation severity, and vestibular compensation status, all of which guide management selection and rehabilitation planning [25].

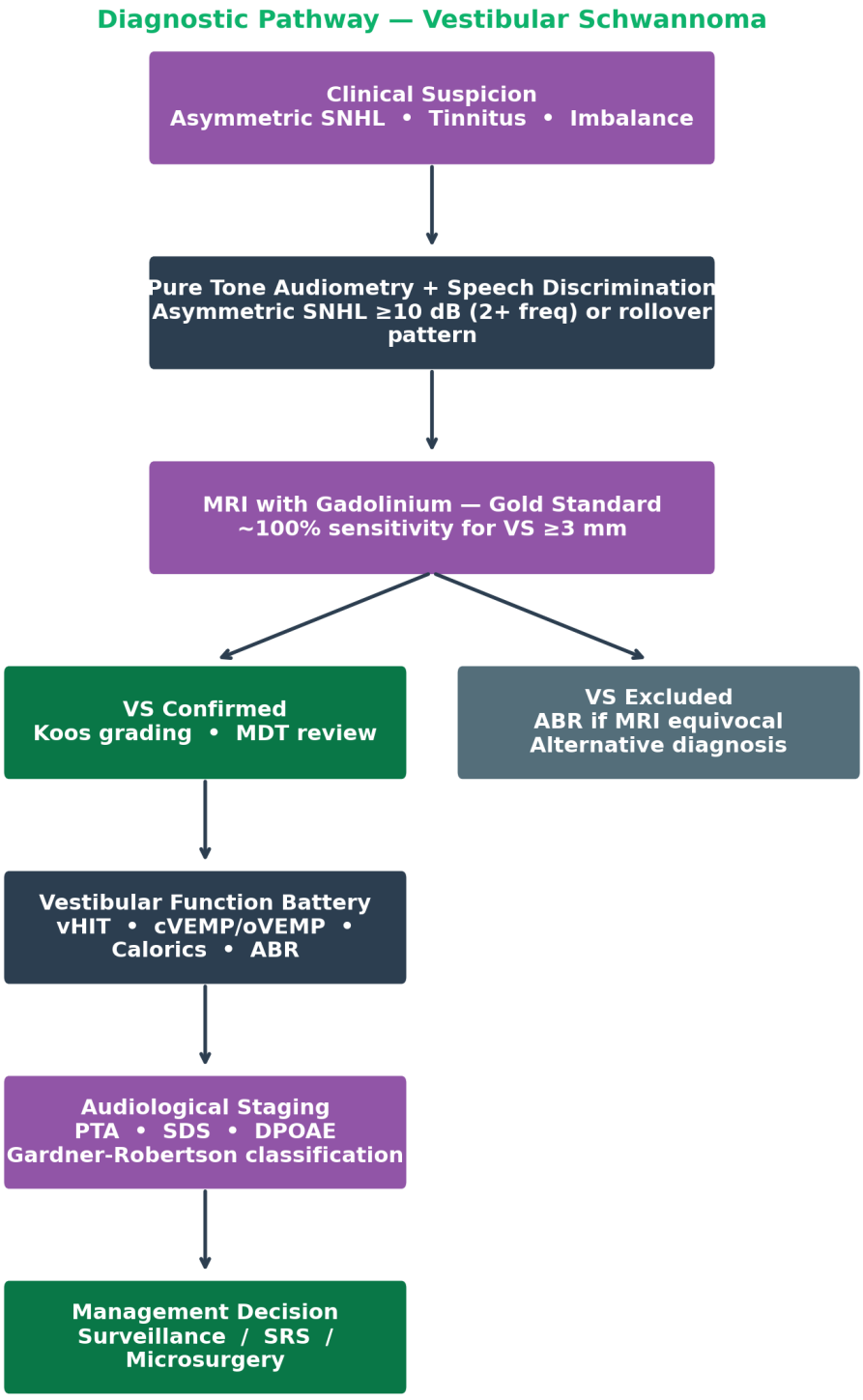

Figure 2. Diagnostic evaluation pathway for suspected vestibular schwannoma. The pathway integrates audiological screening (PTA, SDS), targeted vestibular testing (vHIT, VEMP, calorics), ABR, and gadolinium-enhanced MRI.

Source: Australian Dizziness Clinics, 2026.

Audiological Assessment

Pure tone audiometry (PTA) with speech discrimination testing (SDS) is the essential first-line evaluation. Asymmetric SNHL — defined as a difference of ≥10 dB at two or more frequencies, or ≥15 dB at any single frequency between ears — is the principal audiological trigger for further investigation [26]. The

rollover index (RI = [SDS_max - SDS_high] / SDS_max) is positive when RI >0.35, suggesting retrocochlear pathology [20]. Distortion product otoacoustic emissions (DPOAE) may be preserved early in VS, reflecting intact outer hair cell function with isolated neural involvement [27].

Auditory brainstem response (ABR) demonstrates prolonged wave III and V latencies and increased interpeak latency (I–V >4.4 ms) in the majority of VS patients, reflecting demyelination and axonal compression of eighth nerve fibres [28]. ABR sensitivity for VS detection ranges from 70–90% for tumours >1.5 cm, but falls to 40–60% for small intracanalicular tumours, limiting its utility as a screening tool when MRI is available [29].

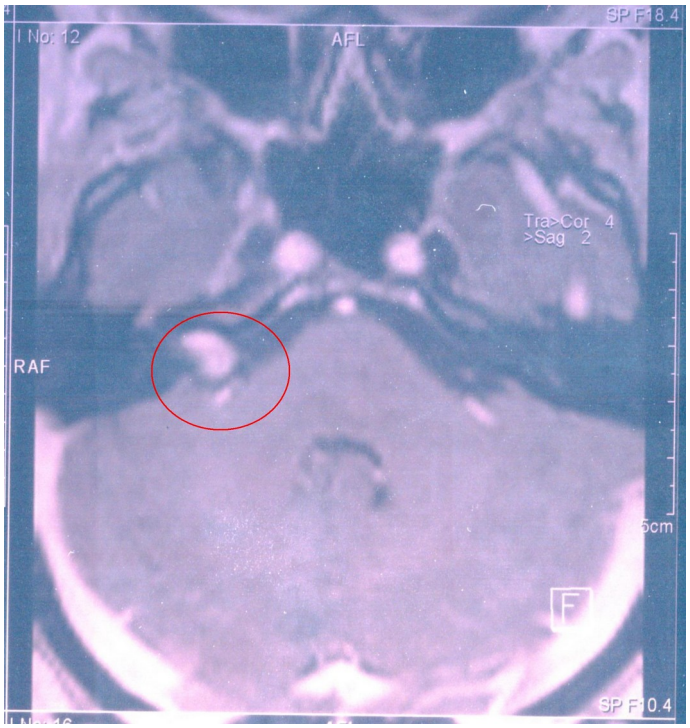

Figure 3. Axial MRI (T1-weighted with gadolinium) demonstrating an enhancing cerebellopontine angle mass consistent with a vestibular schwannoma (Koos Grade II, approximately 2 cm). Note the characteristic 'ice cream cone' appearance with the narrow IAC component and the larger CPA component. The brainstem is not yet displaced.

Source: Slingerjansen, Wikimedia Commons. Public domain.

Vestibular Function Testing

Video head impulse testing (vHIT) quantifies horizontal VOR gain in response to high-frequency (1–6 Hz) head impulses. In VS, the ipsilesional gain is typically reduced (normal >0.8) with compensatory overt and covert saccades, reflecting superior vestibular nerve dysfunction [30]. vHIT has a sensitivity of 85–95% for detecting significant unilateral canal paresis and serves as a rapid, non-sedating alternative to caloric testing [31].

Cervical VEMP (cVEMP) and ocular VEMP (oVEMP) test saccular and utricular function respectively, complementing the horizontal semicircular canal information from vHIT. In VS, cVEMP amplitude is reduced or absent on the affected side in 60–80% of cases, and abnormalities correlate with inferior vestibular nerve involvement [32]. The combination of vHIT and VEMP provides a comprehensive peripheral vestibular profile and can help predict which patients will have difficulty compensating post-treatment [33].

Caloric testing provides gold-standard measurement of low-frequency (0.003 Hz) horizontal semicircular canal function. Unilateral canal paresis (UCP) >25% on bithermal caloric testing is present in approximately 70–85% of VS patients; the degree of UCP does not reliably correlate with tumour size, as central compensation may mask severe peripheral deficit [34].

MRI with Gadolinium

Gadolinium-enhanced MRI is the gold standard for VS diagnosis with sensitivity approaching 100% for tumours >3 mm [35]. The characteristic MRI appearance is a well-circumscribed, homogeneously enhancing mass in the IAC with or without CPA extension — the 'ice cream cone' sign. T2 FIESTA/CISS sequences (3D heavily T2-weighted) can identify small intracanalicular tumours <5 mm as filling defects within the fluid-filled IAC [36]. The minimum sequence requirement for adequate VS evaluation is thin-slice (≤1 mm) T2 FIESTA plus post-gadolinium T1 fat-saturated sequences through the IAC and posterior fossa [37].

□ **Clinical Insight:** High-resolution T2 (FIESTA/CISS) sequences can detect intracanalicular VS <5 mm without gadolinium in many centres, reducing contrast load in elderly patients with renal impairment. Gadolinium remains essential when T2 sequences are equivocal or when CPA meningioma must be excluded [36].

Table 1: Sensitivity and Specificity of Diagnostic Tests for Vestibular Schwannoma

Investigation	Sensitivity (%)	Specificity (%)	Clinical Role
Gadolinium MRI	~100	~100	Gold standard — diagnosis and sizing
ABR (wave I–V latency)	70–90	70–80	Useful if MRI unavailable; poor for small VS
vHIT (ipsilesional gain)	85–95	60–75	Quantifies canal paresis; guides rehab
Caloric testing (UCP)	70–85	65–75	Low-frequency canal function; treatment monitoring
cVEMP amplitude	60–80	70–80	Saccular/inferior vestibular nerve function

V. Grading and Classification

The Koos grading system is the most widely applied clinical classification for VS, providing a reproducible framework for treatment decision-making based on tumour size and anatomical extension [38]. It stratifies tumours into four grades using MRI measurements, with the maximum extrameatal diameter as the primary metric.

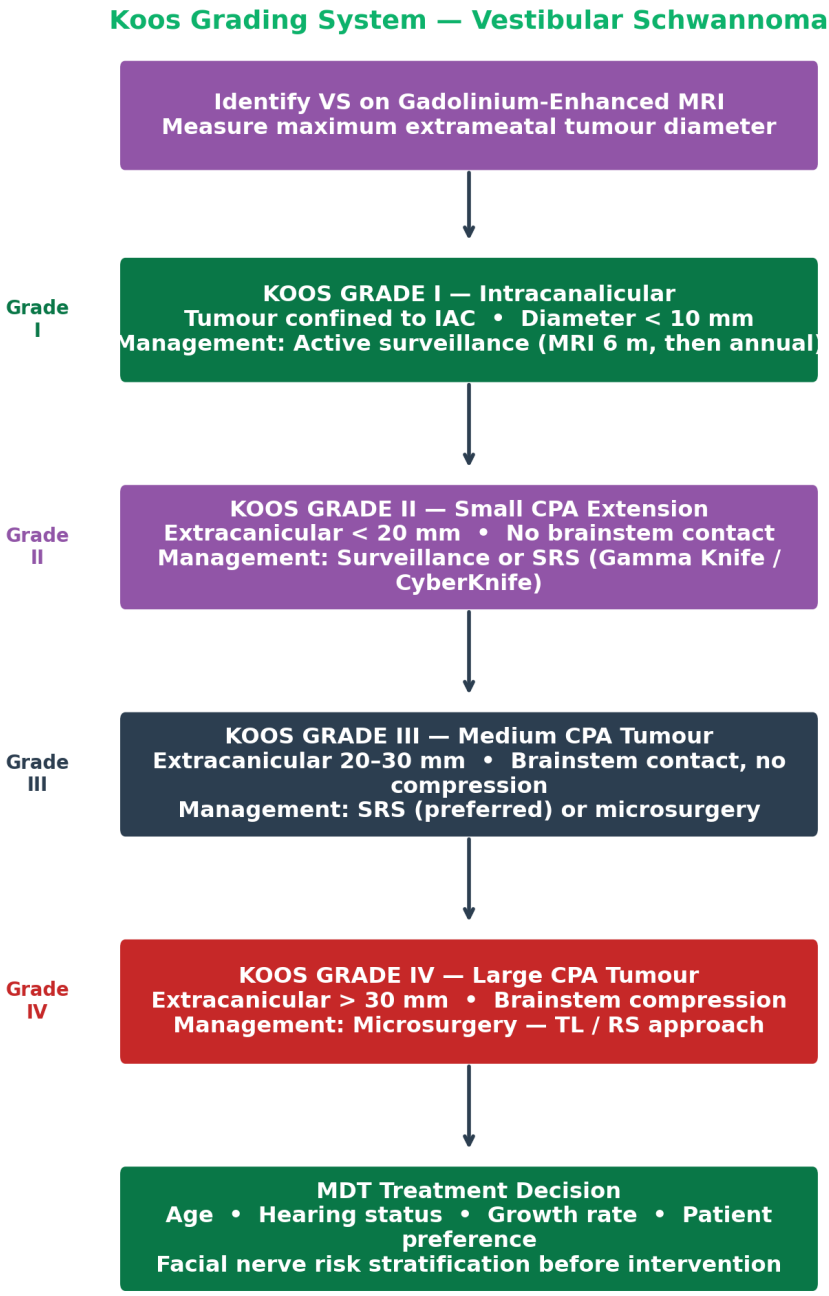

Figure 4. Koos grading classification of vestibular schwannoma. Grade I: intracanalicular, confined to IAC. Grade II: CPA extension <2 cm, no brainstem contact. Grade III: 2–3 cm, brainstem contact without compression. Grade IV: >3 cm, brainstem compression.

Source: Australian Dizziness Clinics, 2026.

Table 2: Koos Grading System for Vestibular Schwannoma

Koos Grade	Size / Location	Brainstem	Typical Management
Grade I (Intracanalicular)	Confined to IAC	No contact	Surveillance or SRS
Grade II	CPA extension, <2 cm	No contact	Surveillance or SRS

	extrameatal		
Grade III	2–3 cm extrameatal	Contact, no compression	SRS or microsurgery
Grade IV	>3 cm extrameatal	Compression / distortion	Microsurgery (preferred)

The House-Brackmann facial nerve grading scale (I–VI) and the American Academy of Otolaryngology hearing classification (Classes A–D based on PTA and SDS) are the standard outcome measures for pre- and post-treatment functional assessment [39]. Gardner-Robertson hearing classification is also used in surgical outcome reporting. Serviceable hearing — typically defined as PTA ≤50 dB and SDS ≥50% (Gardner-Robertson Class I–II) — is the threshold for considering hearing-preserving surgical approaches [40].

□ **Key Point:** Koos grading guides the initial management discussion but should be interpreted alongside patient age, contralateral hearing, growth rate on serial MRI, and patient preference. No single parameter should drive management in isolation [38].

VI. Active Surveillance

Active surveillance (AS) — also termed 'wait and scan' — is now the initial management strategy for the majority of newly diagnosed VS, particularly for Koos I–II tumours in patients over 65, those with serviceable hearing, and those with no or minimal symptoms [41]. Population-based registries demonstrate that approximately 50–60% of VS under surveillance require no active intervention over 10 years [42].

The standard MRI surveillance protocol comprises gadolinium-enhanced MRI at 6 months following initial diagnosis, then annually for 5 years, transitioning to biennial if no growth is demonstrated [43]. Growth is defined as an increase in maximum extrameatal diameter of ≥2 mm on sequential imaging (accounting for measurement variability). Clinically relevant growth — particularly tumours approaching Koos III — typically triggers transition to active treatment [44].

Audiological monitoring (annual PTA and SDS) runs in parallel with imaging surveillance. Significant audiological decline — defined as a shift of ≥20 dB in PTA or ≥20% in SDS — may prompt intervention even in the absence of tumour growth, as the window for hearing-preserving treatment may close with progressive cochlear nerve damage [45].

□ **Clinical Insight:** Patient anxiety associated with 'living with an untreated tumour' is a documented driver of management escalation in AS cohorts. Structured patient education and shared decision-making — including clear communication of the high likelihood of tumour stability — are essential to minimise anxiety-driven treatment choices [41].

Table 3: Active Surveillance Protocol for Vestibular Schwannoma

Timepoint	MRI Protocol	Audiological Assessment
Baseline	Gadolinium MRI — IAC and posterior fossa, thin-slice	PTA, SDS, tympanometry
6 months	Gadolinium MRI (growth baseline)	PTA, SDS
12 months (Year 1)	Gadolinium MRI	PTA, SDS, ABR if indicated
Annual (Years 2–5)	Gadolinium MRI	PTA, SDS annually
Biennial (Year 5+)	Gadolinium MRI if stable	PTA annually

VII. Stereotactic Radiosurgery

Stereotactic radiosurgery (SRS) delivers a precisely targeted high dose of radiation to the tumour in a single fraction, exploiting the radiobiological sensitivity of Schwann cells while minimising dose to adjacent structures — in particular the facial (VII) and cochlear (VIII) nerves and the brainstem [46]. Gamma Knife radiosurgery (GKRS) is the most extensively studied SRS platform, with cohort data

spanning more than three decades. Linear accelerator (LINAC)-based SRS and CyberKnife provide alternative delivery platforms with equivalent efficacy in comparative studies [47].

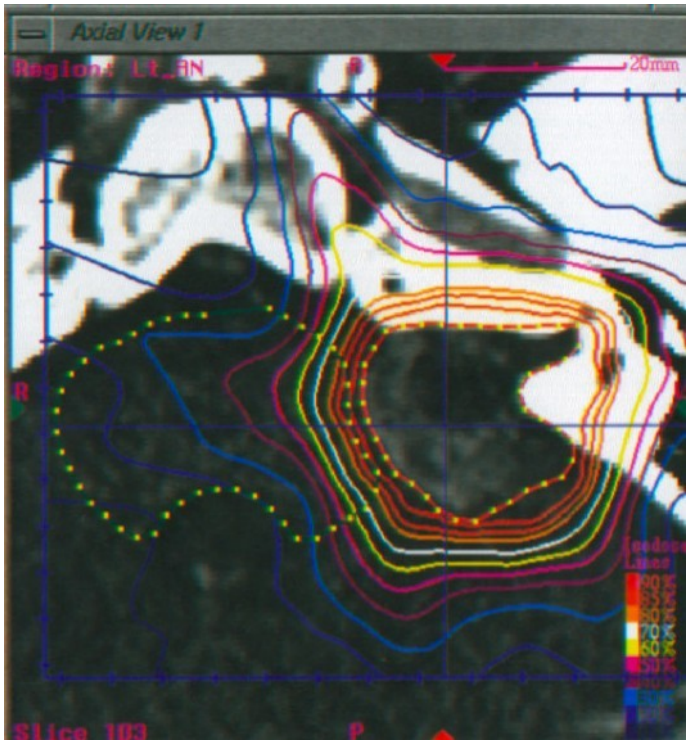

Figure 5. Axial MRI demonstrating CyberKnife/stereotactic radiosurgery treatment planning for a right-sided vestibular schwannoma. Isodose lines show the conformal radiation dose distribution targeted to the tumour with sharp dose falloff protecting the brainstem and cochlea.

Source: Collins SP et al. 'CyberKnife Radiosurgery for Acoustic Neuromas.' Neurosurg Focus 2007. Wikimedia Commons. CC BY 2.0.

Dose and Technique

The standard GKRS marginal dose for VS is 12–13 Gy prescribed to the 50% isodose line, following evidence from the Pittsburgh series that dose reduction from 16 to 12–13 Gy dramatically reduced facial nerve morbidity while maintaining equivalent tumour control [48]. At 12–13 Gy, 5-year tumour control rates (defined as absence of radiological growth requiring additional intervention) exceed 95% in contemporary series [49]. Ten-year control rates of 85–93% are reported in large single-institution cohorts [50].

Cochlea dose constraints — typically aiming for a mean cochlear dose <4–5 Gy — are associated with improved hearing preservation at 3 years [51]. Hearing preservation rates of 60–75% at 5 years are reported in patients with serviceable hearing (Gardner-Robertson Class I–II) pre-treatment, compared to <40% with historical higher doses [52]. Facial nerve preservation (House-Brackmann Grade I–II) is achieved in >97% of patients at contemporary dose levels [49,53].

Fractionated stereotactic radiotherapy (FSRT) — delivering 25 Gy in 5 fractions or 50 Gy in 25 fractions — is employed for larger tumours (Koos III), where single-fraction SRS would deliver unacceptably high integral brainstem dose [54]. FSRT exploits normal tissue repair between fractions to improve the therapeutic ratio for larger volumes.

□ **Key Point:** Post-SRS tumour swelling (transient enlargement) occurs in 20–30% of cases in the first 12–24 months and should not be interpreted as treatment failure. True progression — defined as growth beyond 24 months requiring re-treatment — occurs in <10% of cases at 12–13 Gy [49].

Table 4: Stereotactic Radiosurgery Outcomes — Key Evidence

Study / Series	n	Key Outcome	Notes
Kondziolka et al. 2003 [49]	162	95% tumour control at 5 years	12–13 Gy, GKRS

Hasegawa et al. 2013 [50]	440	89% control at 10 years	Long-term GKRS cohort
Kano et al. 2009 [51]	117	61% serviceable hearing preservation	Cochlear dose <4 Gy
Chopra et al. 2007 [53]	216	97.5% facial preservation	Contemporary dose series
Foote et al. 2012 [54]	149 (FSRT)	94% control, 82% hearing	Large VS, fractionated

VIII. Microsurgical Management

Microsurgery remains the definitive treatment for large VS (Koos IV), tumours causing brainstem compression or hydrocephalus, cystic VS (which respond poorly to SRS), and in patients with NF2 requiring maximal hearing preservation in their better-hearing ear [55]. The three principal approaches — translabyrinthine (TL), retrosigmoid (RS), and middle fossa (MF) — differ in their access to the IAC, hearing preservation potential, and surgical morbidity profile.

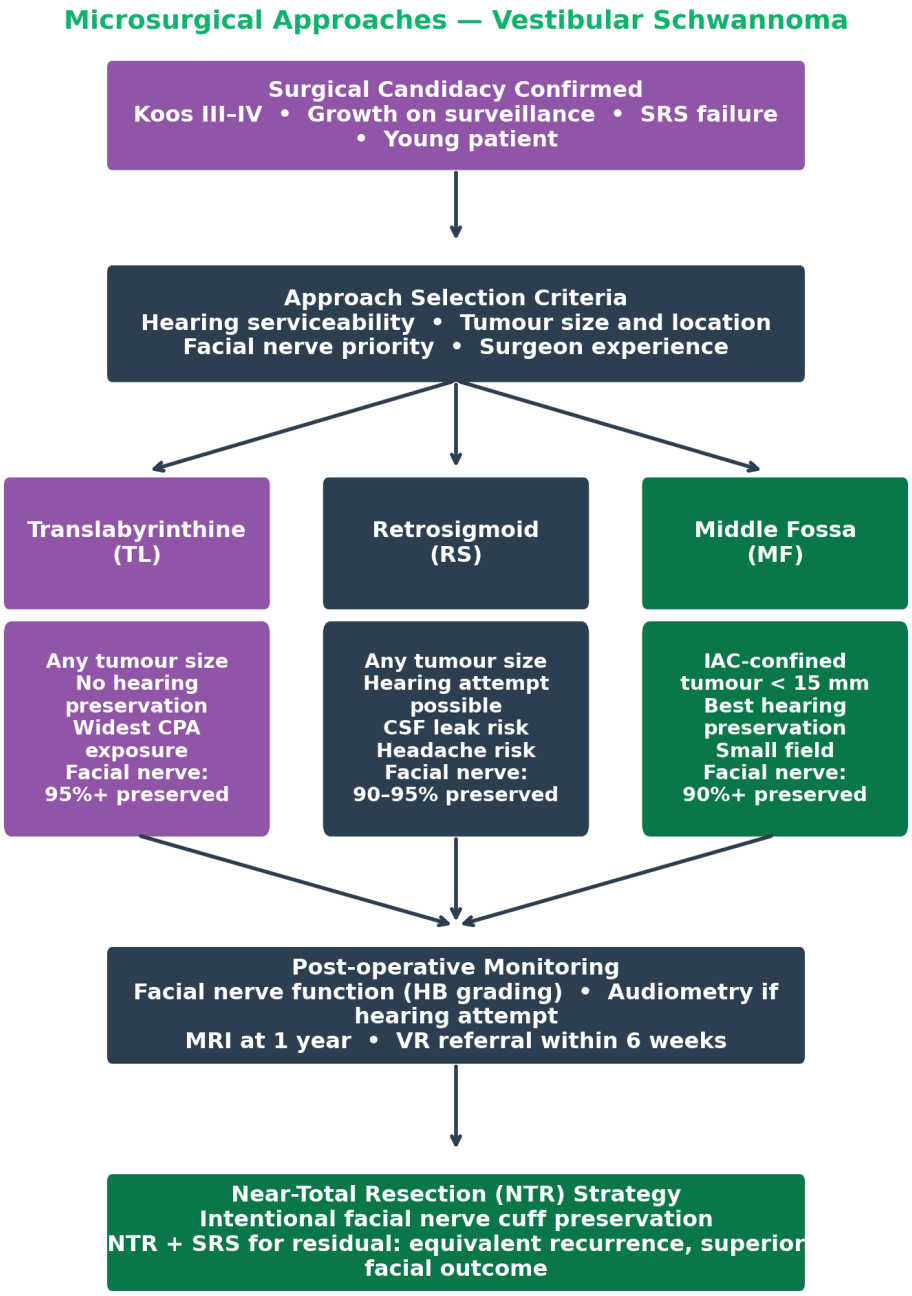

Figure 6. Comparison of the three microsurgical approaches to vestibular schwannoma. The translabyrinthine approach sacrifices hearing to provide optimal facial nerve exposure. The retrosigmoid approach allows hearing preservation attempts for tumours of any size. The middle fossa approach offers the best hearing preservation but is limited to small intracanalicular tumours.

Source: Australian Dizziness Clinics, 2026.

Surgical Approaches

The translabyrinthine (TL) approach provides the most direct access to the IAC from a posterior to anterior direction, allowing identification of the facial nerve from the stylomastoid foramen to the IAC fundus before

tumour dissection. It is the preferred approach for large tumours and patients with non-serviceable hearing. Post-operative CSF leak risk is managed with fat graft obliteration of the mastoid cavity [56].

The retrosigmoid (RS) approach via posterior craniotomy provides excellent CPA exposure and is applicable to tumours of any size. It allows attempted hearing preservation through preservation of the posterior lip of the IAC and posterior semicircular canal. Headache — attributable to bone dust in the posterior fossa and dural adhesions — affects 30–40% of patients post-operatively and may persist for years [57]. The RS approach is limited by restricted IAC fundus access, increasing the risk of residual tumour at the fundal extent.

The middle fossa (MF) approach offers the highest hearing preservation rates (up to 65–75% for Gardner-Robertson Class I–II) but is technically demanding due to brain retraction, temporal lobe exposure risk, and the limited working space for tumours >1.5 cm [58]. It is best reserved for small (<1.5 cm) intracanalicular tumours in young patients with excellent pre-operative hearing and no significant CPA component.

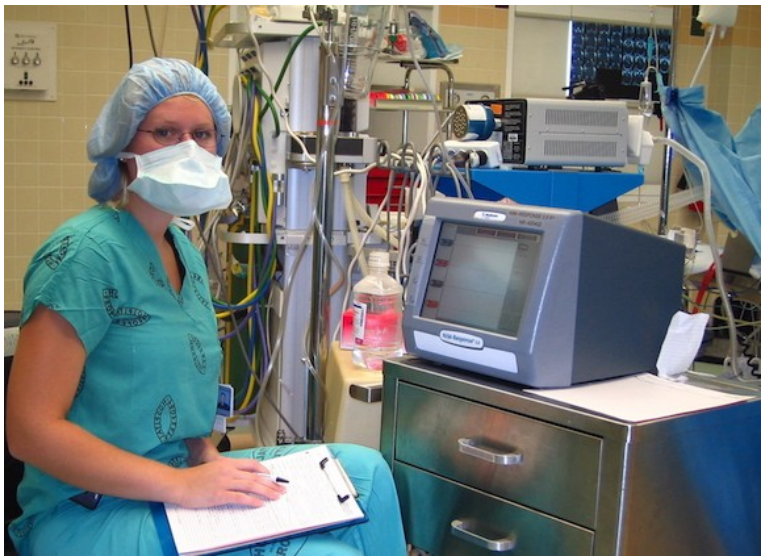

Figure 7. Intraoperative facial nerve electromyography (EMG) monitoring setup during microsurgical vestibular schwannoma resection. Continuous facial nerve monitoring is mandatory in all VS surgery to guide dissection and predict post-operative facial function. Spontaneous activity, triggered EMG, and A-train patterns provide real-time facial nerve integrity information.

Source: Student30, Wikimedia Commons. CC BY-SA 4.0.

Facial Nerve Outcomes and Complications

Facial nerve anatomical preservation is achieved in >90% of cases by experienced surgeons in high-volume centres [59]. However, anatomical preservation does not guarantee functional preservation — immediate post-operative House-Brackmann Grade I–II is reported in 50–80% of cases for large tumours, improving to 70–90% at 12 months with nerve recovery [60]. Intraoperative facial nerve monitoring (continuous EMG and evoked responses) is mandatory in all VS surgery and is the strongest predictor of post-operative facial function [61].

Hearing preservation (serviceable hearing, Gardner-Robertson Class I–II) is achieved in 40–65% of RS approaches and 45–75% of MF approaches for small tumours, with rates falling significantly for tumours >2 cm due to cochlear nerve stretch and internal auditory artery compromise [62]. CSF leak requiring lumbar drain or revision occurs in 5–15% of posterior fossa approaches and is the most common surgical complication requiring return to theatre [57].

□ **Clinical Pearl:** Near-total resection (NTR) — intentional preservation of a small tumour cuff adherent to the facial nerve — has equivalent recurrence rates to gross total resection in most series and significantly better facial nerve outcomes. NTR followed by SRS for any measurable residual is an increasingly adopted strategy for large tumours [59].

IX. Special Considerations: NF2 and Bilateral VS

Neurofibromatosis type 2 (NF2) is an autosomal dominant tumour suppressor syndrome caused by germline mutations in the NF2 gene on chromosome 22q12. Bilateral VS is the hallmark feature and the diagnostic criterion sufficient to establish NF2 in isolation [63]. NF2 affects approximately 1 in 25,000–33,000 live births, with ~50% of cases arising from de novo mutations. The Manchester diagnostic criteria (revised 2019) allow diagnosis of NF2 on the basis of bilateral VS, unilateral VS plus first-degree relative with NF2, or a combination of VS with meningioma, schwannoma, ependymoma, or glioma [64].

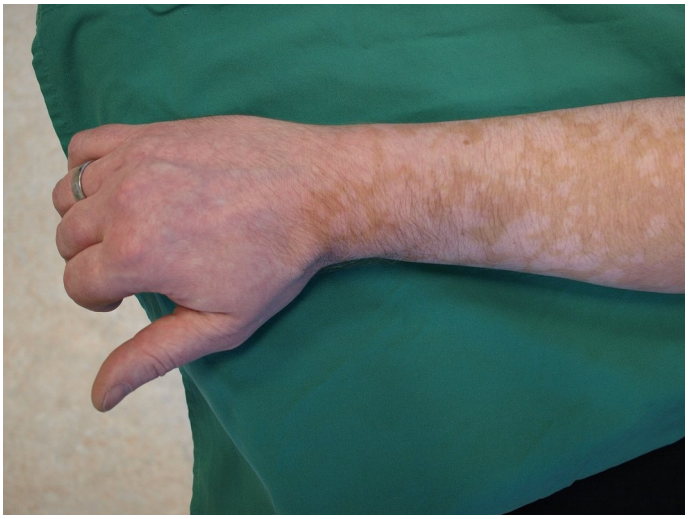

Figure 8. Café-au-lait macules — hyperpigmented skin lesions that may be present in NF2, though they are more numerous and diagnostic in NF1. In NF2, fewer than 6 café-au-lait spots are typical. Any patient presenting with bilateral vestibular schwannoma or VS under 30 years of age should undergo formal NF2 genetic screening and full cutaneous examination.

Source: Klaus D. Peter, Wiehl, Germany. Wikimedia Commons. CC BY 3.0 DE.

The management of NF2-associated bilateral VS is considerably more complex than sporadic VS due to the imperative to preserve hearing in at least one ear. Treatment decisions must account for the differential tumour burden in each ear, the status of contralateral hearing, tumour growth trajectory, age of onset (younger patients have more aggressive phenotypes), and the concurrent burden of non-VS schwannomas and meningiomas [65]. Bevacizumab — an anti-VEGF monoclonal antibody — has demonstrated radiological and audiological response in NF2 patients with progressive VS, with a hearing improvement or stabilisation rate of approximately 60% in reported series [66].

Auditory brainstem implants (ABI) are indicated in NF2 patients who have lost serviceable hearing bilaterally following surgery or progressive deafferentation. ABI delivers electrical stimulation directly to the cochlear nucleus, bypassing the absent or non-functional auditory nerve. Performance is variable — most users achieve awareness of environmental sounds and improved lip-reading, with a minority achieving open-set speech understanding — but ABI significantly improves quality of life and communication for profoundly deaf NF2 patients [67].

□ **Important:** All patients presenting with VS under 30 years of age, bilateral VS, or VS with additional intracranial or spinal tumours should be referred for formal NF2 genetic counselling and testing. A diagnosis of NF2 has profound implications for surveillance, treatment planning, and family screening [63,64].

The mTOR inhibitor everolimus has been investigated in NF2-associated schwannomas based on the rationale that merlin loss activates mTOR signalling [12]. Early phase trials have shown modest radiological responses. Lapatinib (ErbB inhibitor) showed partial responses in approximately 23% of NF2 patients in a phase II study [68]. No systemic therapy is currently approved for NF2 in Australia, but patients with progressive NF2-related VS should be considered for clinical trials where available.

X. Vestibular Rehabilitation and Quality of Life

Vestibular rehabilitation (VR) is indicated at all stages of VS management: pre-treatment (to optimise compensation prior to intervention), post-SRS (to address transitional decompensation during the 3–18-month treatment effect latency), and post-microsurgery (to manage acute unilateral deafferentation) [69]. The evidence base for VR in VS is smaller than for idiopathic vestibular neuritis but consistently demonstrates improvement in DHI scores, balance performance, and falls risk [70].

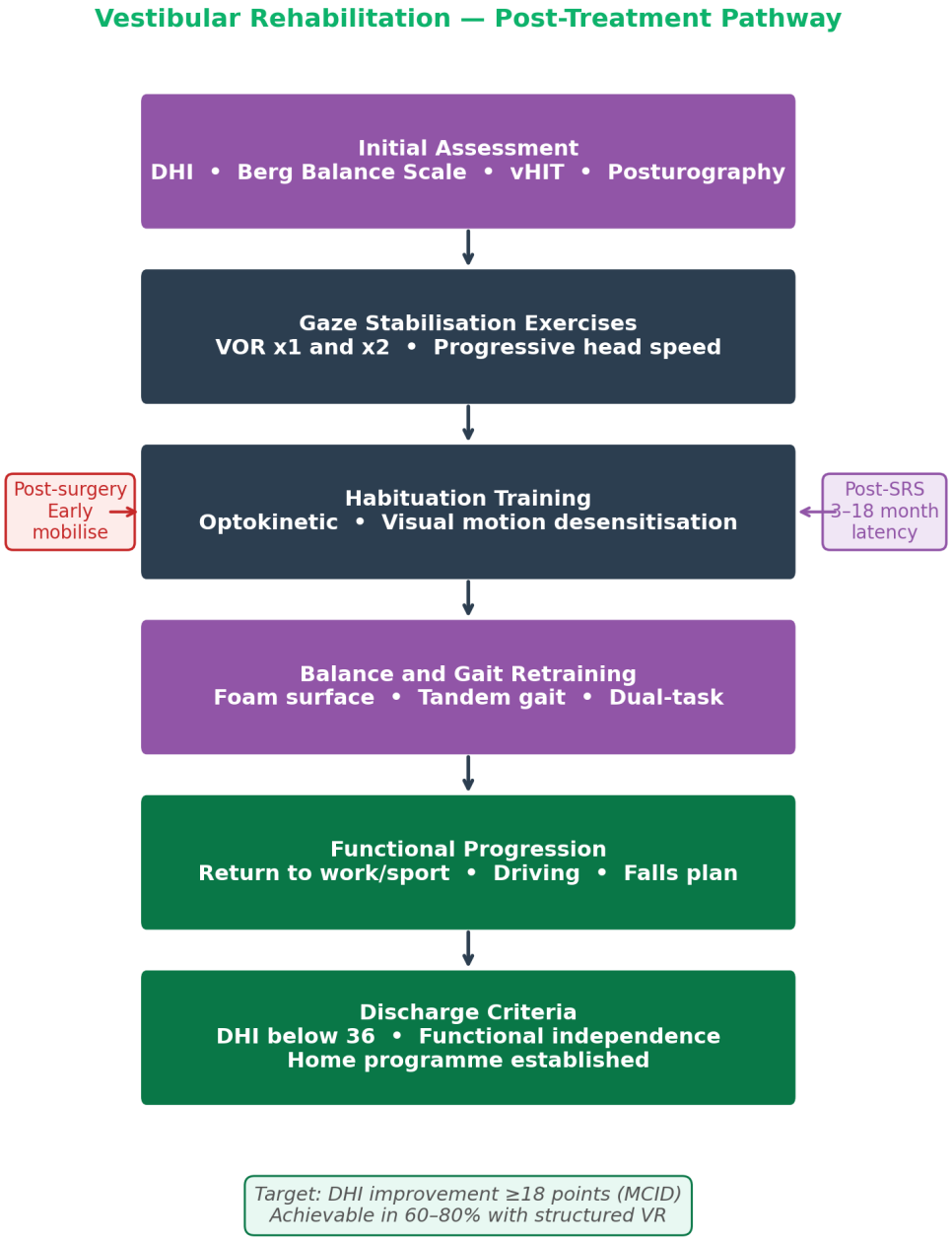

Figure 9. Vestibular rehabilitation pathway following VS treatment. The programme integrates gaze stabilisation, habituation, and balance retraining, with differentiated post-SRS and post-surgery protocols. Discharge criteria include DHI <36 and functional independence.

Source: Australian Dizziness Clinics, 2026.

Gaze stabilisation exercises (GSE) — VOR x1 and x2 exercises at progressively higher frequencies and amplitudes — are the cornerstone of VR for unilateral vestibular hypofunction [71]. In the post-surgical context, GSE may begin within days of surgery; in post-SRS patients, the optimal timing depends on the clinical trajectory of the treatment effect, which typically lags by 3–18 months [72]. Habituation exercises targeting visually-induced dizziness (VID) are particularly important in VS patients, as VID is a common and disabling sequela of unilateral vestibular deafferentation [73].

The Dizziness Handicap Inventory (DHI) is the primary patient-reported outcome measure. Pre-treatment DHI scores in VS patients correlate poorly with objective vestibular test results, reflecting the dominant role of central compensation and psychological factors (anxiety, catastrophising) in determining functional disability [74]. Post-treatment DHI improvement of ≥18 points (minimum clinically important difference) is achievable with structured VR in 60–80% of patients [75].

Long-term quality of life (QoL) data from large prospective cohorts show no significant difference in QoL outcomes between active surveillance, SRS, and surgery for small VS managed in specialised centres — provided complications are avoided [76]. Patient-specific factors — particularly pre-treatment anxiety, hearing loss severity, and tinnitus burden — are stronger predictors of long-term QoL than the treatment modality chosen [77]. Tinnitus management with structured tinnitus counselling and sound enrichment should be incorporated into all VS care pathways [78].

□ **Key Point:** Vestibular rehabilitation delivered by a trained physiotherapist experienced in vestibular disorders significantly improves outcomes compared to generic physiotherapy or no rehabilitation in VS patients post-treatment. Referral should be proactive, not reactive, and ideally initiated before intervention [69,70].

Table 5: Quality of Life Outcome Domains in Vestibular Schwannoma

Domain	Assessment Tool	Clinical Significance
Vestibular handicap	Dizziness Handicap Inventory (DHI)	Primary PRO; >18pt change = MCID
Hearing function	PTA, SDS, AzBio in noise	Direct treatment outcome
Tinnitus severity	Tinnitus Handicap Inventory (THI)	Present in 70–80%; needs specific Rx
Psychological wellbeing	HADS / GAD-7	Anxiety common; drives treatment choice
Balance and falls	Berg Balance Scale, TUG	Functional VR outcome measure

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