

VESTIBULAR
SCHWANNOMA
CHEAT SHEET

Vestibular Schwannoma — Cheat Sheet for Vestibular Physicians

Anchor on gadolinium MRI for any asymmetric SNHL. Koos grade drives the management pathway.

► Why Vestibular Schwannoma matters

Annual incidence 1–2/100,000; most common CPA tumour (80–90% of CPA masses). 95% sporadic, 5% NF2-associated (bilateral). Peak fifth decade. Diagnosis typically delayed 3–7 years.

- Asymmetric SNHL (95%) — the single most sensitive symptom. >10 dB at ≥ 2 frequencies: gadolinium MRI mandatory.
- Unilateral tinnitus (70%); imbalance / unsteadiness (50–70%); true rotatory vertigo uncommon.
- Sudden SNHL in 1–2%: gadolinium MRI within 2 weeks mandatory, even after spontaneous recovery.
- Growth unpredictable: 50% stable, 30% slow (<2 mm/yr), 20% rapid. Year-1 MRI = strongest predictor.

Pearl — Facial weakness or numbness in the context of VS = large tumour or rapid growth. Escalate same day.

Koos classification — anatomical grade and management pathway

| Koos Grade | Tumour extent | Management pathway |
|------------|--|---|
| Grade I | Intracanalicular; IAC only; <10 mm. | Active surveillance first-line. |
| Grade II | CPA extension <20 mm; no brainstem contact. | Surveillance or SRS; shared decision. |
| Grade III | 20–30 mm; brainstem contact, no compression. | SRS preferred; microsurgery if SRS fails. |
| Grade IV | >30 mm; brainstem compression. | Microsurgery recommended; NTR preferred over GTR. |

Pearl — Age, hearing serviceability, and patient preference modify pathway at every Koos grade. Year-1 MRI growth is the strongest predictor of continued progression.

Diagnostic workup — investigation hierarchy

| Test | Purpose | When to order |
|-----------------------------|--|--|
| PTA + speech discrimination | Asymmetric SNHL; rollover on speech discrimination suggests retrocochlear pathology. | Mandatory at presentation; repeat if asymmetry progresses or symptoms change. |
| Gadolinium MRI (IAM) | Gold standard; ~100% sensitivity for VS >3 mm. Enhancing mass in IAC/CPA. | SNHL >10 dB at ≥ 2 frequencies; sudden SNHL; unilateral tinnitus without alternative; any VS suspicion. |
| vHIT | Ipsilesional gain reduction (60–75%); corrective saccades confirm deafferentation. | Baseline; stage nerve involvement; guide vestibular rehabilitation programme. |
| Caloric testing | Unilateral canal paresis (70–85%); lateralise and quantify hypofunction. | Baseline or when vHIT equivocal; pre-treatment; serial monitoring. |
| cVEMP / ABR | cVEMP: saccular/inferior nerve staging. ABR: retrocochlear; limited sensitivity. | cVEMP: inferior nerve involvement. ABR: when MRI unavailable — 30% false-negative rate for small VS. |

Pearl — ABR has a 30% false-negative rate for small VS — never use as MRI substitute. vHIT and calorics stage nerve function for both prognosis and rehabilitation planning, independent of treatment pathway.

- **Red flags** — Sudden SNHL: MRI within 2 weeks regardless of recovery. Facial weakness/numbness: escalate same day. Rapid growth (>2.5 mm/yr) or new brainstem contact: urgent MDT.

Active surveillance — evidence-based MRI protocol

| Timepoint | Investigation | Escalation trigger |
|-----------|---------------|--------------------|
|-----------|---------------|--------------------|

| | | |
|------------------|---|---|
| 6 months | Gadolinium MRI + audiometry — establishes critical growth baseline. | Growth >2–3 mm OR new brainstem contact: step up to MDT review. |
| Annual × 5 years | Gadolinium MRI + audiometry. | Significant SNHL progression: treatment discussion. Year-1 growth strongest predictor of ongoing progression. |
| Biennial year 5+ | Gadolinium MRI (audiometry if symptomatic). | Continued growth in previously stable tumour: MDT review; consider SRS. |
| Escalate if | Any: rapid symptom change; new facial involvement; NF2 diagnosis confirmed; patient changes preference. | Step to appropriate treatment tier without delay. |

Pearl — 50% of VS show no measurable growth at 5 years and never require active treatment. Year-1 MRI is the single most diagnostically important surveillance scan.

Treatment — evidence-based pathways

| Option | Indications | Key outcomes |
|--------------------------------|--|---|
| Active surveillance | Koos I–II; elderly; patient preference; minimal symptoms. Serial MRI monitoring. | 50% never need active treatment. Safe long-term strategy with structured protocol. |
| SRS (Gamma Knife / CyberKnife) | Koos I–III; 12–13 Gy single fraction; serviceable hearing; elderly; medical comorbidities. | Tumour control >90% at 10 yr. CN VII preserved 97–99%. Pseudoprogession 20% at 6–18 months — do not escalate. |
| Microsurgery | Koos III–IV with brainstem compression; SRS failure; younger patient with long horizon. | NTR preferred over GTR for CN VII preservation. TL / RS / MF approach by tumour size + hearing. CN VII >95%. |
| Bevacizumab (NF2 only) | NF2-associated bilateral VS; growing bilateral tumours; hearing-preservation goal. | Hearing stabilisation + tumour shrinkage 40–60%. Anti-VEGF 5–10 mg/kg IV 3-weekly. MDT mandatory. |
| Vestibular rehabilitation | All pathways post-treatment; especially post-microsurgery (within 6 weeks). | DHI improvement >18 pts (MCID) in 60–80%. Gaze stabilisation → balance retraining → dual-task gait. |

Pearl — NTR (near-total resection) with intentional facial nerve cuff preservation achieves equivalent recurrence control with superior CN VII outcomes vs GTR. Post-operative SRS for measurable residual is the preferred strategy at high-volume centres.

NF2 and bilateral VS

Autosomal dominant; NF2 gene germline mutation; penetrance ~100% by age 60. Manchester criteria: bilateral VS, or FDR with NF2 + unilateral VS or two other NF2 tumours.

Hearing preservation is the paramount goal — bilateral deafness risk mandates ABI planning from outset. Avoid aggressive surgery in either ear until MDT review.

SRS control rates lower (70–80%) than sporadic VS. Re-treatment SRS feasible for rare failures. Bevacizumab preferred for growing bilateral tumours.

Counselling and follow-up

- ▶ 50% of VS never require active treatment — reassure patients that surveillance is safe and effective.
- ▶ Post-SRS pseudoprogession (20% at 6–18 months): MRI enlargement ≠ failure. Resolves spontaneously — do not escalate prematurely.
- ▶ QoL monitoring across all four domains: DHI, Tinnitus Handicap Inventory, HADS, Berg Balance Scale.
- ▶ Psychological comorbidity (anxiety/depression) in 30–40% — raise CBT and SSRI/SNRI early.
- ▶ Driving: defer while vestibular symptoms affect safety. Jurisdiction-specific re-assessment when stable.
- ▶ Audiometry 6-monthly in first 2 years → annually; more frequent if SNHL is progressing.

Key references — Koos WT et al. Acta Neurochir 1998 · Reznitsky M et al. Eur Arch Otorhinolaryngol 2019 · Combs SE et al. Int J Radiat Oncol 2010 · Plotkin SR et al. N Engl J Med 2009 · Macielak RJ et al. Otol Neurotol 2023 (NTR vs GTR)