LABYRINTHITIS
CHEAT SHEET

Labyrinthitis (Acute Vestibulo-cochlear Syndrome) - Cheat Sheet for Vestibular Physicians

Vertigo + ipsilateral SNHL = labyrinthitis until stroke excluded. HINTS Plus first. Corticosteroids within 72 h maximise hearing recovery. VRT from day 2-3.

► Why Labyrinthitis matters

Labyrinthitis (Acute Vestibulo-cochlear Syndrome, AVCS) denotes simultaneous inflammation of the cochlea and vestibular labyrinth, producing both acute vertigo and ipsilateral sensorineural hearing loss (SNHL). This distinguishes it from vestibular neuritis, in which cochlear function is spared. Estimated incidence 3-4/100,000/year; viral aetiology predominates. The critical emergency is AICA stroke - the labyrinthine artery is an AICA branch and the sole blood supply to the inner ear. HINTS Plus examination is the primary stroke exclusion tool; MRI-DWI has a false-negative rate up to 50% within 48 h. Corticosteroids within 72 h optimise both hearing and vestibular recovery.

► Indications - when this diagnosis fits

- Acute-onset sustained (>24 h) severe vertigo WITH simultaneous ipsilateral SNHL and tinnitus - same ear, same onset.
- Spontaneous horizontal-torsional nystagmus, direction-fixed, fast phase beating toward the healthy ear.
- Positive head impulse test (corrective saccade) toward the affected side - reassuring peripheral pattern.
- No central HINTS features: no direction-changing nystagmus, no vertical skew deviation, no normal HIT in AVS context.
- Audiogram confirms SNHL ipsilateral to the pathological HIT side (threshold shift >/= 30 dB across >/= 3 contiguous frequencies).
- May follow URTI or viral prodrome. Preceding otitis media or mastoiditis suggests tympanogenic bacterial route.

► Aetiology - four pathways to labyrinthine injury

Type	Agent / Mechanism	Key clinical distinction
Viral (most common)	HSV-1 reactivation, VZV, mumps, CMV, COVID-19 - haematogenous or direct neural spread	Outpatient; antivirals debated; spontaneous recovery possible
Bacterial - tympanogenic	S. aureus, Strep. via round/oval window from AOM, CSOM or cholesteatoma	Surgical emergency; source control essential; often permanent SNHL
Bacterial - meningogenic	S. pneumoniae, N. meningitidis via cochlear aqueduct or IAC sheath	Bilateral SNHL risk; cochlear implant pathway; IV antibiotics urgent
Autoimmune / Vascular	AIED pattern; or labyrinthine ischaemia in AICA territory stroke	MRI mandatory; bilateral or rapidly progressive; steroid-responsive (AIED)

► HINTS Plus - bedside stroke exclusion in Acute Vestibular Syndrome

Test	Peripheral - reassuring	Central - HIGH RISK (posterior fossa)
Head Impulse	Abnormal - corrective saccade toward the lesion side	Normal HIT - no corrective saccade - STROKE until MRI
Nystagmus	Direction-FIXED horizontal-torsional; Alexander law present	Direction-CHANGING or purely vertical / torsional
Test of Skew	Absent - no vertical skew deviation	Vertical skew present - brainstem crossed pathway involvement
PLUS Audiometry	SNHL present - supports labyrinthitis; fits peripheral AVS	New SNHL with normal HIT = AICA infarct until MRI proven

Pearl - *HINTS Plus sensitivity >95% for posterior fossa stroke in true AVS, outperforming MRI-DWI within 24-48 h. A single central element mandates urgent MRI regardless of clinical gestalt. Valid ONLY in Acute Vestibular Syndrome - not episodic vertigo.*

► Mechanism - disruption of the blood-labyrinth barrier

Compartment	Pathophysiological process	Clinical relevance
Blood-labyrinth barrier	Pro-inflammatory cytokines (TNF-alpha, IL-1b) + ROS trigger tight junction disruption and endolymph ionic collapse	Simultaneous cochlear + vestibular failure in single event
Cochlear hair cells	K+ gradient collapses, mechanotransduction fails; outer then inner hair cells affected	SNHL + tinnitus; outer hair cells most vulnerable; may be permanent
Scarpa ganglion (viral)	HSV-1 latency reactivation in ~70% adults causes acute neural inflammation and VIIIth nerve dysfunction	Viral aetiology predominant; antivirals added empirically
Round window (bacterial)	Bacterial toxins then organisms cross from middle ear into perilymph via round/oval window membranes	Tympanogenic suppurative labyrinthitis - surgical emergency; AOM/cholesteatoma must be identified

► Differential diagnosis - high-yield mimics

Diagnosis	Key distinguishing features

AICA territory stroke	Normal HIT; direction-changing nystagmus; vertical skew; ipsilateral facial palsy; Horners; truncal ataxia; AICA territory = SNHL + vertigo + cerebellar + brainstem signs
Vestibular neuritis	Identical vestibular syndrome - direction-fixed nystagmus + positive HIT - but NO hearing loss on formal PTA; cochlear function entirely spared
Meniere disease	Episodic attacks 20 min-12 h; fluctuating low-frequency SNHL; aural fullness; tinnitus; recurrent - NOT sustained AVS
Ramsay Hunt syndrome	Facial nerve palsy + auricular vesicles (zoster sine herpete possible) + severe otalgia; VZV - antivirals within 72 h reduce palsy severity and progression
Autoimmune inner ear disease	Bilateral or rapidly progressive SNHL developing over weeks to months; systemic autoimmune history; typically steroid-responsive; AIED panel may support
Bacterial labyrinthitis	Fever, otalgia, otoscopy abnormal (AOM / perforation / cholesteatoma); meningism if meningogenic; CT petrous bone and urgent ENT essential

Pearl - *Red flag combination: fever + meningism + SNHL = meningogenic labyrinthitis - IV antibiotics + urgent ENT. AOM or cholesteatoma + labyrinthitis signs = tympanogenic labyrinthitis - surgical emergency. Do NOT observe.*

► Management - evidence-based stepwise approach

Intervention	Regimen	Evidence / principles
Vestibular suppressants (acute)	Prochlorperazine or promethazine PRN; strictly limit to 72 h maximum	Reduces acute distress; prolonged use blocks central compensation and delays recovery
Corticosteroids	Prednisolone 1 mg/kg/day (max 60 mg) x 10-14 days then taper; intratympanic dexamethasone if systemic CI	Rauch 2011 NEJM: IT non-inferior to oral; Strupp 2004: improves caloric recovery; earlier = better hearing outcomes
Antivirals (empirical, viral)	Valacyclovir 1 g TDS x 7 days alongside corticosteroids for likely viral cases	Expert consensus; AUVP trial: no significant additive benefit over steroids alone; used empirically in herpes zoster context
Bacterial labyrinthitis	IV ceftriaxone 2 g daily + urgent ENT referral + mastoidectomy for tympanogenic source	Surgical emergency - source control mandatory; IV antibiotics for meningogenic route; liaise ENT + ID
Vestibular rehabilitation	Gaze stabilisation + balance retraining from day 2-3; mobilise early; avoid bed rest	Cochrane 2015: earlier onset = better functional recovery; PPPD risk reduced with early VRT programme
Audiology follow-up	Formal PTA at day 0, 4 weeks, 3 months; cochlear implant referral if profound permanent SNHL	CI candidacy assessed at 3-month audiogram; do not delay beyond 3 months for CI referral

► Prognosis and counselling

- Vestibular recovery: 70-80% functional compensation at 3-6 months via central reweighting. Persistent canal paresis on vHIT is common and does not preclude functional recovery. Age >65, bilateral involvement, or brainstem comorbidity: supervised VRT programme and formal falls assessment mandatory.
- Hearing recovery (viral): 45-65% partial recovery with early corticosteroids; severe-to-profound loss recovers in fewer than 30%. Bacterial labyrinthitis: permanent profound SNHL likely - initiate cochlear implant pathway promptly at 3-month audiogram.
- PPPD risk: 15-20% develop persistent postural-perceptual dizziness; screen at 3-month review. Early VRT and mobilisation are protective. Cochlear and vestibular recovery timelines differ - improvement continues 6-12 months.

► Key references

[1] Strupp M et al. Methylprednisolone, valacyclovir, or combination for vestibular neuritis. N Engl J Med 2004;351:354-361.

[2] Kattah JC et al. HINTS to diagnose stroke in the acute vestibular syndrome. Stroke 2009;40:3504-3510.

[3] Rauch SD et al. Oral vs intratympanic corticosteroid therapy for idiopathic sudden sensorineural hearing loss. JAMA 2011;305:2071-2079.

[4] Arbusow V et al. Distribution of HSV-1 in human geniculate and vestibular ganglia. Neurology 1999;52:1872-1875.