Labyrinthitis:

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

Labyrinthitis is an inflammatory disorder of the membranous labyrinth in which both the vestibular and cochlear end-organs are involved simultaneously [1,2]. The defining clinical triad — acute-onset severe vertigo, sensorineural hearing loss (SNHL), and tinnitus — sets it apart from vestibular neuritis, in which hearing is preserved, and from acute isolated SNHL, in which vestibular symptoms are absent or minor [3,4]. For the vestibular physician, making this distinction with precision is not an academic exercise: the aetiology, investigation pathway, management, and prognosis differ substantially between these closely related disorders, and the failure to identify labyrinthitis as the presenting syndrome carries the real risk of missing a bacterial complication with life-threatening potential [5,6].

Population-based incidence estimates for labyrinthitis are difficult to disentangle from the broader literature on sudden SNHL and vestibular neuritis, partly because clinical definitions have been inconsistently applied across studies. Using the strictest criterion — concurrent acute vestibular deafferentation and SNHL in the same ear at the same time — the annual incidence is approximately 3–4 per 100,000 adults, with viral labyrinthitis representing the large majority of cases in ambulatory practice [1,7]. Bacterial labyrinthitis is far less common in absolute terms but carries disproportionate risk and arises in two well-characterised clinical contexts: complication of suppurative otitis media (tympanogenic labyrinthitis) and sequela of bacterial meningitis (meningogenic labyrinthitis) [5,8,9].

Age distribution and sex predominance vary by aetiology. Viral labyrinthitis peaks in the third to fifth decades, with a slight female predominance in most series [7]. Post-meningitic labyrinthitis has a paediatric preponderance, with Haemophilus influenzae type b (Hib) and Streptococcus pneumoniae as the dominant organisms in children; Hib vaccine coverage has dramatically reduced the paediatric burden in high-income countries while pneumococcal disease remains an ongoing cause in both age groups [10,11]. Tympanogenic labyrinthitis complicates cholesteatoma and chronic suppurative otitis media (CSOM), occurring at any age but peaking in adults with longstanding otological disease [5,9].

The economic and functional impact of labyrinthitis is substantial. Acute vestibular deafferentation produces complete work incapacity for a minimum of two to four weeks regardless of aetiology [7]. When significant hearing loss accompanies vestibular symptoms, the combined disability extends further, particularly in patients reliant on binaural hearing for occupational performance. In bacterial labyrinthitis, the consequences extend to permanent profound unilateral deafness in the majority, and to bilateral deafness in the meningitic context — the principal driver of cochlear implantation referral in post-meningitic children [10,12].

□ **Key Point:** Labyrinthitis = cochlear + vestibular involvement. The simultaneous presence of acute SNHL and vestibular deafferentation in the same ear defines the syndrome and distinguishes it from vestibular neuritis and isolated SNHL. This distinction governs investigation and management.

Table 1. Epidemiology and aetiology of labyrinthitis at a glance.

Parameter	Viral	Bacterial
Annual incidence	~3–4 / 100,000 adults [7]	Rare — occurs as complication [5,8]
Peak age	30–50 years [7]	Children (meningogenic); adults (tympanogenic) [10]
Sex ratio	Slight female predominance [7]	No clear sex predominance [5]
Dominant agents	HSV-1 reactivation, mumps, CMV, VZV, COVID-19 [1,13]	S. pneumoniae, N. meningitidis, H. influenzae [10,11]
Route of infection	Haematogenous / neural spread [1,13]	Meningogenic, tympanogenic, haematogenous [5,8,9]
Hearing outcome	60–80% recovery (viral) [7,14]	Usually permanent unilateral or bilateral SNHL [10,12]

II. Anatomy, the Blood-Labyrinth Barrier, and Pathophysiology

A command of labyrinthine microanatomy is prerequisite for understanding how infection and inflammation damage the inner ear with the severity and pattern that characterises labyrinthitis. The membranous labyrinth — suspended within the bony labyrinth of the petrous temporal bone — comprises two functionally distinct compartments: the pars superior (utricle, three semicircular canals, and their cristae ampullares) serving vestibular function, and the pars inferior (saccule and cochlear duct) where sound transduction occurs [15,16]. Both compartments share the same endolymph-filled membrane system and are contiguous via the ductus reuniens, explaining why a single inflammatory insult can devastate both cochlear and vestibular function simultaneously.

The blood-labyrinth barrier

The blood-labyrinth barrier (BLB) is the inner-ear equivalent of the blood-brain barrier, created by tight junctions between the non-fenestrated endothelial cells of the spiral ligament capillaries, the stria vascularis, and the dark cells of the vestibular end-organ [15,17]. This barrier maintains the unique ionic composition of endolymph — high potassium (approximately 150 mEq/L), low sodium — that is essential for hair-cell mechanotransduction. The BLB restricts entry of large molecules, immune cells, and most pathogens into the endolymph space, providing a degree of immune privilege analogous to the central nervous system [17].

BLB disruption is the pivotal pathophysiological event in labyrinthitis of any aetiology. Once inflammatory mediators — including tumour necrosis factor alpha (TNF-alpha), interleukin-1 beta (IL-1B), and reactive oxygen species — breach the tight-junction complex, plasma proteins and immune effectors flood the endolymph space, the potassium gradient collapses, hair-cell resting potential is disrupted, and the stage is set for neuroepithelial injury [1,17,18]. This cascade explains why the clinical onset is acute and severe regardless of whether the inciting agent is a reactivating herpes simplex virus or a bacterium traversing the round window membrane.

Routes of pathogen entry into the labyrinth

Three anatomically distinct routes allow pathogens or inflammatory mediators to access the membranous labyrinth. Understanding them is essential for predicting clinical pattern, guiding imaging, and selecting management.

- **Haematogenous route —** Systemic viraemia or bacteraemia seeds the BLB capillaries. Viral agents (HSV-1, mumps, CMV, VZV, COVID-19) reach the labyrinth via this route, with documented viral DNA present in cochlear and vestibular ganglia at post-mortem [13,19]. The spiral ganglion and vestibular (Scarpa's) ganglion harbour latent herpes simplex virus-1 in approximately 70% of the adult population; reactivation — triggered by physiological stress, immunosuppression, or febrile illness — provides the most plausible mechanism for idiopathic viral labyrinthitis in adults [13,20].
- **Tympanogenic (transtympanic) route —** Infection spreads from the middle-ear space into the labyrinth, principally through the round window membrane and, less commonly, through the oval window or bony labyrinthine dehiscences. This route complicates acute suppurative otitis media (AOM), CSOM with cholesteatoma, and rarely iatrogenic middle-ear procedures. Enzymes and toxins produced by middle-ear bacteria traverse the round window first, producing serous labyrinthitis; subsequent direct bacterial invasion converts this to suppurative labyrinthitis with severe and usually irreversible neuroepithelial destruction [5,8,9,21].
- **Meningogenic route —** Bacteria reach the perilymph via the cochlear aqueduct (connecting the scala tympani to the subarachnoid space) or via the internal acoustic meatus along the eighth-nerve sheath. This route explains labyrinthitis as a recognised complication of bacterial meningitis — most commonly pneumococcal. Labyrinthitis occurs in approximately 25–35% of bacterial meningitis survivors who have any degree of post-meningitic hearing loss, and in virtually all cases of profound bilateral SNHL following meningitis [10,11,22].

Serous versus suppurative labyrinthitis

The anatomopathological distinction between serous and suppurative labyrinthitis has profound prognostic implications. In serous labyrinthitis, the endolymph space is invaded by inflammatory mediators — bacterial toxins, enzymes, cytokines — but not by viable bacteria or leucocytes. Hair cells and spiral ganglion neurons are injured but not universally destroyed, leaving some potential for recovery if the inciting middle-ear infection is controlled swiftly [5,18,21]. Hearing loss is typically partial and may partially recover if the inflammatory source is eliminated in time.

Suppurative labyrinthitis, by contrast, involves actual bacterial invasion of the perilymph and endolymph spaces with leucocyte infiltration, fibrin deposition, and progressive neuroepithelial destruction. The histopathological sequence — first inflammatory exudate, then fibrous organisation, then new bone formation (labyrinthitis ossificans) — is relentless once bacteria gain access [21,23]. Hearing loss is typically profound and permanent. The speed of this progression — sometimes within 24–48 hours of AOM onset — underlines the imperative for early intervention. The round window membrane's selective permeability to large-molecule toxins before viable bacteria can cross defines the serous-to-suppurative transition as the critical therapeutic window [5,18].

□ **Clinical Insight:** The round window membrane is selectively permeable to bacterial toxins and cytokines before it allows viable bacteria to cross. Aggressive middle-ear management during the serous phase — before suppurative conversion — may prevent full labyrinthine destruction and preserve some hearing. This window can close within 24–48 hours of AOM onset [5,21].

III. Aetiology — Viral, Bacterial, Autoimmune, Toxic, and Vascular Causes

Labyrinthitis results from a heterogeneous group of mechanisms, of which viral reactivation is by far the most prevalent in the ambulatory and hospital outpatient setting [1,7]. A systematic aetiological classification informs the investigation approach and underpins rational treatment selection; the clinician who applies a single treatment protocol across all labyrinthitis subtypes will both over-treat and under-treat.

Table 2. Aetiological classification of labyrinthitis with distinguishing features.

Category	Agent / Mechanism	Route	Key distinguishing feature
Viral — HSV-1 reactivation	Herpes simplex virus 1	Haematogenous / neural	Commonest cause in adults; no prodromes in ~50% [13,20]
Viral — Paramyxovirus	Mumps, measles, CMV, VZV	Haematogenous	Childhood / vaccine-preventable; mumps = unilateral profound SNHL [24]
Viral — COVID-19	SARS-CoV-2	Haematogenous / neural	Cochlear and vestibular involvement; evolving evidence [25]
Bacterial — tympanogenic	S. aureus, P. aeruginosa, Gram-negatives	Round window / tympanogenic	Complicates AOM/CSOM/cholesteatoma; serous to suppurative [5,9]
Bacterial — meningogenic	S. pneumoniae, N. meningitidis	Cochlear aqueduct / IAM	Post-meningitic; bilateral risk; ossification within weeks [10,22]
Autoimmune / Cogan's	Immune-mediated BLB injury	Systemic immune activation	Bilateral, progressive; interstitial keratitis, audiovestibular loss [26]
Vascular — labyrinthine infarction	Labyrinthine artery / AICA branch occlusion	Arterial occlusion	Abrupt seconds-onset; no viral prodrome; cardiovascular risk factors; sentinel for AICA stroke

			[55,56]

Viral labyrinthitis — HSV-1 reactivation

Latent herpes simplex virus type 1 is demonstrable by polymerase chain reaction in up to 70–89% of human vestibular ganglia and in a substantial proportion of spiral ganglia at autopsy, establishing the cochlear and vestibular nervous supply as a reservoir for periodic viral reactivation [13,19,20]. The reactivation model — originally proposed for vestibular neuritis on the basis of Arbusow's landmark ganglion studies — extends to labyrinthitis when the reactivating virus spreads into the membranous labyrinth rather than remaining confined to the superior vestibular nerve. This anatomical extension accounts for the simultaneous cochlear involvement that separates labyrinthitis from pure vestibular neuritis [3,20]. In practical terms, a middle-aged patient presenting with a flu-like prodrome followed by acute-onset severe rotatory vertigo, unilateral SNHL, and tinnitus — in the absence of otoscopic abnormality or neurological signs — is most likely experiencing HSV-1 reactivation-associated labyrinthitis. Serology has low diagnostic specificity; diagnosis remains clinical [1,7,27].

Viral labyrinthitis — paramyxovirus and COVID-19

Mumps was historically the dominant cause of sudden unilateral profound SNHL — often permanent — in school-age children and young adults [24]. The MMR vaccination programme has dramatically reduced mumps labyrinthitis in high-income countries, but vaccine hesitancy sustains ongoing cases. Varicella zoster virus (VZV) causes labyrinthitis in the context of Ramsay Hunt syndrome, where ear canal or pinna vesicles, ipsilateral facial nerve palsy, and audiovestibular loss constitute the full clinical picture [28]. SARS-CoV-2 has been associated with audiovestibular symptoms in both acute and post-acute phases. A meta-analysis of COVID-19 audiovestibular involvement reported a pooled prevalence of hearing loss of approximately 7.6% and vestibular symptoms in a smaller proportion meeting criteria for labyrinthitis [25]. Whether this represents direct viral cochlear invasion, immune-mediated BLB disruption, or vascular thromboembolism remains under investigation.

Bacterial labyrinthitis — tympanogenic

Tympanogenic labyrinthitis arises when infection in the middle-ear cleft — from AOM, CSOM, or cholesteatoma — breaches the round window membrane and enters the perilymph. The incidence of tympanogenic labyrinthitis as a complication of AOM has declined markedly with antibiotic availability, but the complication remains clinically relevant in patients who delay medical care, have resistant organisms, or present with cholesteatoma — where enzymatic erosion of the bony labyrinth provides an additional route [5,9,21]. Organisms mirror those causing AOM and CSOM: S. pneumoniae, H. influenzae, M. catarrhalis, S. aureus, P. aeruginosa, and in cholesteatomatous disease, a polymicrobial mix including anaerobes. The clinical presentation — purulent otorrhoea or otalgia, then sudden new-onset vertigo and acute hearing deterioration — is the hallmark, and the ear examination is the most important diagnostic act at presentation [5,8].

Bacterial labyrinthitis — meningogenic

Bacterial meningitis is the most devastating cause of bilateral labyrinthitis. The cochlear aqueduct provides a direct conduit from the subarachnoid space into the scala tympani, while the internal acoustic meatus allows spread along the eighth-nerve sheath. Pneumococcal meningitis carries the highest labyrinthitis risk; approximately 25–35% of survivors develop significant SNHL, with the proportion varying by organism, treatment delay, and adjunctive dexamethasone use [10,11,29]. Cochlear damage in post-meningitic labyrinthitis is characterised by its speed and irreversibility: new bone formation in the cochlea (labyrinthitis ossificans) can begin within days and may be demonstrable on MRI within 2–4 weeks [22,23]. This biological clock defines the urgency of cochlear implantation assessment in post-meningitic deafness — delay beyond 3–6 months may preclude standard implantation [12,22].

Autoimmune inner ear disease and Cogan's syndrome

Autoimmune inner ear disease (AIED) produces fluctuating, often bilateral, rapidly progressive SNHL with variable vestibular involvement — the clinical signature is the multi-week to multi-month time course, bilateral asymmetry, and corticosteroid responsiveness [26,30]. Cogan's syndrome is a rare autoimmune

vasculitis characterised by interstitial keratitis, bilateral audiovestibular loss resembling Ménière's disease, and systemic vasculitis with aortitis as the most feared complication. The vestibular physician must maintain diagnostic vigilance for Cogan's in any young patient with bilateral audiovestibular loss and ocular symptoms; ophthalmological referral is mandatory [26,30].

□ **Important:** Any acute vertigo with ipsilateral hearing loss and ipsilateral facial nerve palsy — especially with periauricular vesicles — is Ramsay Hunt syndrome (VZV) until proven otherwise. Prompt antiviral therapy within 72 hours of vesicle onset significantly improves facial nerve and hearing outcomes [28]. Never attribute the facial palsy to the labyrinthitis itself.

Vascular labyrinthine ischaemia — the sentinel infarct

Ischaemia of the labyrinth from occlusion of the labyrinthine (internal auditory) artery or its branches produces a clinical picture that is anatomically indistinguishable from viral labyrinthitis — simultaneous acute unilateral vestibular deafferentation and SNHL with a peripheral HINTS pattern [55,56,57]. This overlap is the most dangerous diagnostic trap in the acute audiovestibular syndrome, because missing a vascular aetiology means missing a stroke sentinel event.

Labyrinthine artery anatomy and occlusion syndromes

The labyrinthine (internal auditory) artery arises from the AICA in 75–80% of individuals, with direct origin from the basilar artery in the remainder [57,58]. It is an end-artery with no anastomotic reserve. It bifurcates into two principal branches: the anterior vestibular artery (AVA), supplying the superior and lateral semicircular canal cristae and the utricle; and the common cochlear artery (CCA), which further divides into the cochlear branch (apical cochlea) and the vestibulocochlear artery (posterior semicircular canal, saccule, basal cochlea) [57]. Three distinct occlusion syndromes arise from the level of arterial involvement:

- **Full labyrinthine artery occlusion:** complete simultaneous unilateral SNHL and vestibular deafferentation — the presentation most closely mimicking viral labyrinthitis [55,56].
- **Isolated AVA occlusion:** acute vestibular deafferentation without hearing loss — mimics vestibular neuritis; the HINTS exam is peripherally normal yet the cause is vascular [57,58].
- **Isolated CCA / cochlear branch occlusion:** sudden SNHL without vertigo — presents as sudden sensorineural hearing loss requiring vascular exclusion alongside the standard sudden SNHL protocol [59].

AICA stroke risk — the 30-day sentinel window

Labyrinthine infarction is not merely an isolated end-artery event. Lee et al. (2009) demonstrated that 15–25% of patients presenting with isolated audiovestibular loss attributable to labyrinthine infarction experienced an AICA territory stroke within 30 days [55]. The mechanism mirrors transient ischaemic attack preceding major ischaemic stroke: the same atherosclerotic, cardioembolic, or small vessel pathology affecting the labyrinthine artery branch can progress to involve the AICA trunk itself, causing posterior-fossa cerebellar and brainstem infarction. This sequence means that a patient presenting with what appears to be straightforward acute labyrinthitis and harbouring cardiovascular risk factors may be in a high-risk sentinel window. Failing to recognise and investigate this aetiology carries potentially catastrophic consequences [55,56,60].

Clinical flags for vascular aetiology

No single clinical feature pathognomically separates vascular labyrinthine ischaemia from viral labyrinthitis at the bedside. The following constellation should substantially raise clinical suspicion for vascular cause and mandate urgent vascular investigation [55,56,61,62]:

- **Onset in seconds** — viral labyrinthitis typically evolves over hours; vascular onset is instantaneous.
- **No viral prodrome** — absence of coryzal illness, fever, or myalgia in the preceding days.
- **Age over 60 and cardiovascular risk factors** — hypertension, diabetes, dyslipidaemia, AF, prior TIA or stroke, smoking.

- **Associated posterior circulation symptoms** — even transient diplopia, dysphagia, facial numbness, or limb ataxia — any brainstem symptom raises AICA infarct risk substantially.
- **DWI-negative MRI within 24–48 hours** — does NOT exclude labyrinthine or brainstem infarction. DWI sensitivity for small posterior fossa infarcts is approximately 80% in the first 24–48 hours; the labyrinth itself is rarely directly resolved on standard DWI [62].

□ **Important:** Vascular labyrinthine ischaemia is a sentinel event. Any patient with acute audiovestibular syndrome, cardiovascular risk factors, and abrupt-seconds onset — regardless of HINTS peripheral pattern — requires same-day vascular workup: urgent MRI brain with DWI (repeat at 24–48 h if initially negative), MRA posterior circulation, ECG, and cardiology review within 24–48 hours. Do not anchor on 'peripheral pattern' as exclusion of vascular cause [55,56,60].

□ **Key Point:** The HINTS examination distinguishes peripheral from central vestibular disease (brainstem/cerebellar infarct). It does NOT distinguish viral from vascular peripheral labyrinthine disease. A patient with vascular labyrinthine ischaemia will have a positive (abnormal) head impulse test and a HINTS-peripheral pattern — identical to viral labyrinthitis [57,58].

IV. Clinical Features and Natural History

Labyrinthitis presents acutely with a triad of severe rotatory vertigo, sensorineural hearing loss, and tinnitus, with nausea and vomiting proportionate to the acuity of the vestibular deafferentation [1,3]. The onset is typically abrupt — patients recall the exact hour — and the initial hours are dominated by the vestibular component: the room is spinning, the patient is unable to stand without support, and vomiting is often severe. This parallels vestibular neuritis in its vestibular phenomenology but is distinguished by the simultaneous auditory symptoms in the same ear.

The spontaneous nystagmus of acute labyrinthitis reflects loss of peripheral tonic firing from the affected labyrinth [3,31]. In the acute phase, nystagmus is direction-fixed, horizontal-torsional, with the fast phase beating away from the affected ear (the intact ear drives an uncontested slow phase toward the lesioned side). Nystagmus is inhibited by visual fixation in purely peripheral disease — a positive fixation-inhibition test supports a peripheral rather than central generator. The vestibular physician should examine without fixation using video-Frenzel goggles to reveal the full-amplitude spontaneous nystagmus [3,31].

Auditory symptoms and their significance

The hearing loss in viral labyrinthitis is typically unilateral, of sudden onset, and most pronounced in the high frequencies in HSV-associated disease — though all patterns (flat, rising, or pan-cochlear loss) are described [7,14]. Tinnitus accompanies hearing loss in the majority and may persist well beyond vestibular recovery. Aural fullness is less consistent than in Ménière's disease and, when present, likely reflects endolymph compartment oedema from inflammatory BLB disruption rather than true endolymphatic hydrops.

A hearing loss of greater than 30 dB across three contiguous frequencies constitutes sudden SNHL by the standard AAO-HNS definition, and its simultaneous occurrence with vestibular deafferentation satisfies the clinical criteria for labyrinthitis [1,32]. The absence of any hearing change on formal pure tone audiometry in a patient with acute vestibular deafferentation should redirect the clinician to vestibular neuritis, though subclinical cochlear involvement detectable only on formal audiometry is well documented in patients who report subjective hearing change that does not meet threshold criteria [4,7].

Bedside vestibular examination

Bedside examination in acute labyrinthitis mirrors vestibular neuritis: a positive head impulse test (HIT) toward the affected ear reflects canal paresis on vHIT; direction-fixed spontaneous nystagmus with fixation inhibition is present; the HINTS Plus battery is peripheral in pattern. The HIT is the pivotal bedside test — a normal HIT in the context of acute vestibulo-cochlear loss must trigger immediate posterior-fossa imaging to exclude an AICA territory stroke, which can produce identical symptoms and represents the most dangerous mimic of labyrinthitis [33,34].

Skew deviation and gaze-evoked nystagmus are absent in uncomplicated labyrinthitis. Their presence — particularly skew deviation (vertical-ocular misalignment on alternate cover test) and direction-changing or gaze-evoked nystagmus — constitute the central HINTS triad (normal HIT, skew present, direction-changing nystagmus) and demand urgent imaging regardless of how typical the history sounds [33,34]. The HINTS Plus protocol, incorporating finger-rub hearing assessment as a fourth element, increases diagnostic specificity for posterior-fossa stroke when cochlear symptoms accompany a central nystagmus pattern [34].

Natural history by subtype

Viral labyrinthitis follows a broadly predictable natural history. Acute vertigo and nausea peak in the first 24–48 hours and subside over one to two weeks as central vestibular compensation develops. Residual unsteadiness — reflecting incomplete peripheral recovery and imperfect central compensation — persists for six to twelve weeks in most patients and longer in older adults with pre-existing deficits in vision or somatosensation [7,35]. Spontaneous nystagmus resolves within one to three weeks; head-impulse abnormalities may persist indefinitely if canal paresis does not recover, though patients compensate to functional independence [3,35].

Hearing in viral labyrinthitis recovers partially or fully in approximately 60–80% of patients within three months, with most recovery in the first two to six weeks [7,14,27]. Recovery is better for low-frequency and mid-frequency loss than for high-frequency loss. Complete non-recovery of hearing is associated with profound initial loss, delayed corticosteroid initiation beyond two weeks from onset, and advanced age [7,27]. Bacterial labyrinthitis — both tympanogenic and meningogenic — carries a far worse hearing prognosis, with permanent moderate-to-profound SNHL in the large majority, and a significant proportion with bilateral SNHL in the meningogenic context [10,12].

□ **Clinical Pearl:** The HINTS battery (Head Impulse, Nystagmus pattern, Test of Skew) plus hearing assessment is the most sensitive bedside protocol for separating peripheral labyrinthitis from AICA stroke. A normal head impulse test in acute vestibulo-cochlear loss is a stroke until proven otherwise — urgent MRI-DWI before vestibular suppressants [33,34].

V. Investigations — Audiometry, Imaging, and Vestibular Function Testing

The investigation protocol for labyrinthitis is determined by the clinical subtype, the urgency of the presentation, and the presence or absence of red flags suggesting a central mimic or a bacterial aetiology. No investigation should delay MRI where stroke is a credible differential, and no investigation should delay ENT referral where middle-ear disease is visible on otoscopy [33,34].

Pure tone audiometry

Pure tone audiometry (PTA) with air and bone conduction thresholds and speech discrimination scores is the mandatory baseline investigation for all presentations of labyrinthitis. It serves four functions: confirms the sensorineural pattern (ruling out conductive loss from middle-ear pathology); documents the severity and configuration of the hearing loss for medicolegal and prognostic purposes; establishes the baseline for monitoring recovery; and provides eligibility criteria for cochlear implantation if recovery does not occur [1,7,27]. PTA should be performed within 48 hours of presentation where possible, and repeated at four to six weeks and three months to document recovery trajectory.

Speech discrimination scores (word recognition scores) provide functional context beyond the thresholds: a patient may retain near-normal PTA thresholds but have markedly impaired discrimination, reflecting spiral ganglion neuron injury rather than hair-cell damage alone. This pattern has implications for cochlear implant candidacy — poor discrimination with moderate PTA thresholds may qualify for implantation under current guidelines [12]. Auditory brainstem response (ABR) is indicated when PTA cannot be reliably obtained (children, suspected non-organic hearing loss, very young infants post-meningitis) [10,12].

MRI with gadolinium

MRI with gadolinium is the modality of choice for investigating labyrinthitis. In the acute phase, T1-weighted post-gadolinium sequences demonstrate enhancement of the cochlea, vestibule, and semicircular canals, reflecting BLB disruption and inflammatory vasodilation [36,37]. Enhancement confirms an active inflammatory process within the membranous labyrinth and excludes cochlear aplasia or malformation as a cause of hearing loss, but is not pathognomonic of any specific aetiology — viral, bacterial, and autoimmune labyrinthitis all produce labyrinthine enhancement [36,37].

High-resolution T2-weighted sequences (CISS, FIESTA) provide exquisite visualisation of the fluid-filled labyrinthine spaces and are essential in post-meningitic assessment: loss of the normal T2-bright signal within the cochlear turns indicates early labyrinthitis ossificans — a finding of critical urgency marking the beginning of the window for cochlear implantation before irreversible fibrosis occludes the scala tympani [22,36]. Diffusion-weighted imaging (DWI) must be included when clinical examination does not unequivocally exclude a central cause: its sensitivity for posterior-fossa stroke exceeds 90% beyond 24 hours [33,34,38].

Vestibular function testing

Video head impulse testing (vHIT) of all six semicircular canals provides objective quantification of canal paresis in acute labyrinthitis and is the recommended first-line vestibular function test in the acute setting — it is feasible bedside, non-invasive, and does not depend on prolonged water-irrigation stimulation [3,39]. In acute labyrinthitis, vHIT typically demonstrates unilaterally reduced VOR gain across all three canals of the affected ear, reflecting panvescicular involvement. A selective gain reduction in only one or two canals, or a pattern consistent with superior division involvement alone, favours vestibular neuritis over full labyrinthitis [3,4,39].

Vestibular evoked myogenic potentials (cVEMPs for saccular and inferior vestibular nerve function; oVEMPs for utricular and superior vestibular nerve function) complete the functional map of the otolith-vestibular pathways [39,40]. Their pattern complements the semicircular canal vHIT findings: absent or attenuated cVEMPs with preserved horizontal-canal vHIT suggests selective inferior-division involvement, while panvescicular loss on both vHIT and VEMPs confirms global labyrinthine deafferentation. Caloric testing — the gold standard for unilateral canal paresis documentation — adds little to acute management over vHIT and is reserved for the subacute or chronic review phase [39].

Table 3. Investigation protocol for labyrinthitis by clinical scenario.

Clinical scenario	Priority investigations	Rationale
Acute vestibulo-cochlear loss, normal ear exam, no neurological signs	PTA + speech discrimination; MRI gadolinium + DWI; vHIT all canals	Exclude stroke; document hearing baseline; quantify canal paresis [33,36,39]
Acute vestibulo-cochlear loss, abnormal ear exam (purulent AOM / cholesteatoma)	Urgent ENT referral; CT temporal bone; PTA if feasible; blood cultures	Tympanogenic route; need surgical assessment; IV antibiotics immediately [5,9]
Post-meningitic hearing loss in child or adult	Urgent PTA + ABR; MRI T2 FIESTA within 4 weeks; CT temporal bone at 4–6 weeks	Labyrinthitis ossificans timing — cochlear implant window closes quickly [10,22]
Bilateral progressive audiovestibular loss	PTA both ears; cVEMPs; oVEMPs; autoimmune screen (ANA, ANCA, anti-HSP70); ophthalmology	Autoimmune inner ear disease, Cogan's syndrome — steroid-responsive [26,30]

□ **Key Point:** MRI with gadolinium for acute labyrinthitis must include DWI sequences. Labyrinthine enhancement on T1+Gd confirms active BLB disruption — but only DWI detects the posterior-fossa stroke that identically mimics labyrinthitis. Never order one without the other in the acute presentation.

VI. Differential Diagnosis

The differential diagnosis of acute labyrinthitis is structured around the clinical triad: is the hearing loss real, is the vertigo truly peripheral, and are there features pointing to a specific aetiology? Four diagnoses account for the great majority of cases that initially resemble labyrinthitis [3,4,33].

AICA territory stroke — the critical exclusion

The anterior inferior cerebellar artery (AICA) is the dominant arterial supply to the labyrinth via the internal auditory artery. An AICA stroke can produce sudden unilateral SNHL, tinnitus, and vestibular deafferentation — an exact anatomical copy of viral labyrinthitis [33,34]. The incidence of AICA stroke presenting as acute audiovestibular syndrome is estimated at 1–4% of patients presenting with acute vestibulo-cochlear loss in emergency series, but the consequence of a missed stroke is catastrophic. The HINTS Plus examination separates these conditions at the bedside with sensitivity exceeding 95% and specificity approximately 85% in trained hands [34]. Key central features: normal horizontal head impulse test, direction-changing nystagmus, vertical ocular misalignment (skew deviation), ataxia disproportionate to vestibular deafferentation, and any additional brainstem or cerebellar sign [33,34].

Vestibular neuritis

Vestibular neuritis and labyrinthitis share the same acute vestibular deafferentation syndrome but differ in the presence or absence of auditory involvement. The distinction requires formal hearing assessment — bedside screening with voice and finger rub is insensitive to losses under 30–40 dB. A normal PTA within 48 hours of symptom onset, in the context of clinical vestibular deafferentation and intact hearing on formal testing, establishes vestibular neuritis as the diagnosis [3,4,7]. The management differs: labyrinthitis with hearing loss triggers the sudden SNHL corticosteroid protocol with hearing monitoring, while vestibular neuritis management is weighted toward vestibular recovery without audiological follow-up.

Meniere's disease

Meniere's disease produces episodic attacks of vertigo (typically 20 minutes to 12 hours), fluctuating low-frequency SNHL, tinnitus, and aural fullness, caused by endolymphatic hydrops [41]. The differentiation from a first attack of viral labyrinthitis with cochlear involvement can be difficult at initial presentation: both produce acute vertigo and simultaneous SNHL. The key temporal discriminator is attack duration — Meniere's attacks last minutes to hours, never days, and the patient typically returns to a near-normal baseline between attacks. Labyrinthitis produces a sustained, non-episodic deafferentation with persistent spontaneous nystagmus and hearing loss for days to weeks. A fixed, predominantly high-frequency loss without recovery over four to six weeks raises the probability of labyrinthitis; a fluctuating low-frequency loss favours Meniere's [41].

Ramsay Hunt syndrome

VZV reactivation in the geniculate ganglion produces the Ramsay Hunt syndrome: otalgia, periauricular or ear canal vesicles, ipsilateral lower motor neuron facial palsy, and audiovestibular loss [28]. The audiovestibular component is indistinguishable from labyrinthitis at the histological level — VZV spreads within the spiral ganglion and vestibular ganglia, producing direct neuronal injury. Recognition is critical because antiviral therapy with acyclovir or valacyclovir within 72 hours of vesicle appearance demonstrably improves facial nerve and hearing outcomes, and the facial palsy requires neurology or ENT co-management [28]. In the immunocompromised host, VZV can disseminate, making urgent antiviral therapy even more critical.

Table 4. Differential diagnosis of acute labyrinthitis.

Diagnosis	Hearing loss	Vertigo pattern	Ear exam	Key distinguishing feature
Viral labyrinthitis	Acute unilateral SNHL	Sustained, days to weeks	Normal	HINTS-peripheral; corticosteroid-responsive [1,7]
Vestibular neuritis	Absent	Sustained, days to weeks	Normal	Normal PTA; purely vestibular deafferentation [3,4]

AICA stroke	Acute unilateral SNHL	Sustained or brief	Normal	Normal HIT; skew deviation; ataxia; DWI-positive [33,34]
Meniere's disease	Low-frequency, fluctuating	Episodic 20 min to 12 h	Normal	Attacks, not sustained; hydrops on delayed Gd MRI [41]
Ramsay Hunt syndrome	Variable SNHL + tinnitus	Sustained	Vesicles in EAC / pinna	Facial palsy; VZV; antiviral-responsive within 72 h [28]
Vascular labyrinthine ischaemia	Acute unilateral SNHL	Sustained (seconds onset)	Normal	Cardiovascular risk factors; no prodrome; HINTS-peripheral; sentinel for AICA stroke [55,56]

□ **Important:** Cholesteatoma can present as tympanogenic labyrinthitis with little prior warning — the disease may be entirely asymptomatic until it erodes into the labyrinth. Always examine the tympanic membrane under adequate light in any acute audiovestibular presentation. Posterior epitympanic cholesteatoma is easily missed without systematic pneumatoscopy and endoscopic otoscopy.

Vascular labyrinthine ischaemia versus viral labyrinthitis

The critical additional differential the vestibular physician must always hold in mind is that a peripheral HINTS pattern does not distinguish viral from vascular inner ear pathology [55,56,57]. A patient with labyrinthine artery occlusion will have an abnormal (positive) head impulse test, direction-fixed unilateral spontaneous nystagmus, no skew deviation, and no additional brainstem signs — a HINTS-peripheral pattern identical to viral labyrinthitis. The bedside examination cannot make this distinction.

□ **Clinical Pearl:** The HINTS examination differentiates peripheral from brainstem-central vestibular disease — it does NOT differentiate viral from vascular peripheral labyrinthine pathology. A patient with labyrinthine infarction will have a peripherally positive HIT and will HINTS-peripheral. Never use a peripheral HINTS result to exclude a vascular cause in a high-risk patient [55,57,58].

The clinical features that tilt the probability toward vascular labyrinthine ischaemia rather than viral labyrinthitis are listed below. No feature is pathognomonic; the constellation and risk-factor burden drive clinical probability [55,56,61].

Table 5. Viral labyrinthitis versus vascular labyrinthine ischaemia — clinical discrimination.

Feature	Viral labyrinthitis	Vascular labyrinthine ischaemia
Onset speed	Hours (gradual)	Seconds (instantaneous)
Viral prodrome	Present in ~50% [13,20]	Absent
Age	Any; peak 30–60	Typically over 60
Cardiovascular risk factors	Not elevated	Hypertension, DM, AF, dyslipidaemia [55,56]
HINTS pattern	Peripheral	Peripheral — does not differentiate [57,58]
DWI MRI	Normal	May be negative in first 24–48 h even if stroke present [62]
Risk of subsequent stroke	Not elevated	15–25% AICA stroke within 30 days [55]
Urgent investigation	Audiogram, corticosteroids	MRI DWI + MRA + ECG + cardiac workup same day [55,56]

Investigations that help establish vascular aetiology: urgent MRI brain with DWI (repeated at 24–48 h if initially negative), MRA of the posterior circulation, 12-lead ECG and continuous cardiac monitoring (paroxysmal AF), transthoracic or transoesophageal echocardiography, fasting lipid panel, HbA1c, and haematological screen for hypercoagulable state in young patients without traditional risk factors [55,56,60,61]. These investigations are not necessary in the straightforward young patient with a clear

viral prodrome; they are mandatory in the patient with an atypical presentation or significant cardiovascular risk burden.

VII. Medical Management — Acute and Subacute Phase

Management of acute labyrinthitis addresses three parallel objectives: reducing inner-ear inflammation to minimise permanent damage, providing short-term symptomatic relief from acute vestibular deafferentation, and treating the underlying aetiology where a specific pathogen or mechanism is identified. The urgency and content of treatment differ substantially between viral, bacterial-tympanogenic, and bacterial-meningogenic labyrinthitis.

Corticosteroids — the cornerstone of acute treatment

High-dose systemic corticosteroids are the evidence-based first-line treatment for acute labyrinthitis with hearing loss, extrapolated from well-powered randomised trial evidence for sudden SNHL and the vestibular neuritis corticosteroid literature [27,42,43]. The Rauch 2011 New England Journal of Medicine trial comparing oral prednisolone versus intratympanic methylprednisolone for sudden SNHL demonstrated non-inferiority of oral therapy at three months, supporting oral high-dose steroids as the primary route [27]. The recommended regimen is prednisolone 1 mg/kg/day (maximum 60 mg/day) for 7–10 days with a taper over a further 5 days, initiated within two weeks of onset — with evidence suggesting that initiation within 72 hours provides significantly better hearing outcomes [27,42].

Intratympanic steroid injection (ITS) using methylprednisolone 40 mg/mL or dexamethasone 10–24 mg/mL provides high perilymph concentrations without systemic side effects and is the treatment of choice when systemic steroids are contraindicated — poorly controlled diabetes mellitus, active peptic ulceration, psychiatric contraindications [27,44]. ITS can also be offered as salvage treatment for hearing loss that has not fully recovered after a primary systemic steroid course; there is biological rationale for offering this within six weeks of onset, though evidence beyond three months is limited [44]. For the vestibular component, corticosteroid evidence is less robust than for hearing, but the combined audiovestibular disease in labyrinthitis justifies systemic treatment on hearing grounds alone [27,43,45].

Antiviral therapy

The role of antiviral therapy in viral labyrinthitis remains contested. The pivotal Strupp 2004 trial randomised vestibular neuritis patients to methylprednisolone alone, valacyclovir alone, or the combination: the combination offered no benefit over methylprednisolone alone for vestibular recovery, and valacyclovir monotherapy was inferior to steroids [43]. This finding has been widely interpreted as evidence against routine antiviral use in vestibular neuritis. For labyrinthitis with cochlear involvement — where a stronger argument exists for HSV ganglionic reactivation — a definitive equivalent trial has not been completed [43,45].

A pragmatic approach supported by expert consensus is to add valacyclovir 1 g three times daily for 7 days to systemic steroids in patients presenting within 72 hours of onset, on the basis that the risk-benefit profile of valacyclovir is favourable and the pathophysiological rationale is sound, even if the therapeutic increment over steroids alone remains unproven in this specific population [1,43]. The Ramsay Hunt syndrome context is clearly different: antiviral therapy within 72 hours of vesicle onset is supported by multiple controlled trials and is unequivocally indicated [28].

Bacterial labyrinthitis — antibiotics and surgery

Tympanogenic bacterial labyrinthitis is a surgical emergency. Intravenous antibiotics targeting likely causative organisms — ceftriaxone 2 g IV daily is the standard empirical choice, with anti-staphylococcal and anti-pseudomonal cover added for post-tympanostomy or hospital-acquired presentations — must be initiated immediately alongside urgent ENT assessment for myringotomy, drainage, and mastoidectomy where cholesteatoma or extensive middle-ear disease is identified [5,8,21]. The aim of surgery is source control — eliminating the reservoir driving the tympanogenic route — not restoration of labyrinthine function, which is typically already permanently compromised in suppurative disease.

In the meningogenic context, management follows the bacterial meningitis protocol: IV antibiotics guided by organism sensitivity, adjunctive dexamethasone (0.15 mg/kg every 6 hours for 4 days) initiated concurrently with or just before the first antibiotic dose — with evidence from multiple randomised controlled trials showing adjunctive dexamethasone reduces hearing loss in pneumococcal meningitis by approximately 50% [10,11,29]. The vestibular physician's role in this context is typically the subacute assessment of hearing and vestibular function post-meningitis, and the urgent referral pathway for cochlear implantation where ossification is detected [10,12].

Vestibular suppressants — short-term and time-limited

Vestibular suppressants (prochlorperazine, promethazine, diazepam) are indicated for short-term control of acute nausea and vomiting in the first 24–48 hours of acute labyrinthitis, where the vestibular crisis is incapacitating [46]. Beyond 48–72 hours, they must be tapered and discontinued — continued vestibular suppression impairs the development of central vestibular compensation, which depends on cerebellar plasticity in response to the asymmetric tonic input from the deafferented side [35,46]. There is direct animal and human evidence that vestibuloactive drugs prolong postural instability after unilateral peripheral deafferentation. Patients must be counselled explicitly that vertigo will persist for days to weeks, that this represents normal recovery, and that vestibular suppressants beyond the first 48 hours will slow their rehabilitation.

Table 6. Management algorithm by labyrinthitis subtype.

Subtype	First-line treatment	Adjunct	Monitoring
Viral (HSV-1 / idiopathic)	Prednisolone 1 mg/kg/day x 7–10 days, taper [27,42]	Valacyclovir 1 g TDS x 7 days if within 72 h [1,43]	PTA at 4–6 weeks; 3 months; consider ITS if partial recovery [44]
Ramsay Hunt (VZV)	Prednisolone + acyclovir 800 mg 5x daily or valacyclovir 1 g TDS x 7 days [28]	Facial nerve monitoring; neurology co-manage [28]	PTA; facial nerve grading (House-Brackmann) at 4 weeks [28]
Bacterial — tympanogenic	IV ceftriaxone 2 g daily; urgent ENT [5,8,21]	Myringotomy / mastoidectomy; corticosteroids [5]	Post-op CT temporal bone; PTA; CI referral if profound [12]
Bacterial — meningogenic	IV antibiotics + dexamethasone 0.15 mg/kg q6h x 4 days [10,11,29]	Urgent neurology co-manage; post-meningitis CI pathway [12]	ABR within 2 weeks; MRI T2 FIESTA within 4 weeks; CT at 6 weeks [22]

□ **Clinical Pearl:** Start corticosteroids within 72 hours for hearing. Taper vestibular suppressants within 48–72 hours to protect central compensation. In bacterial disease — IV antibiotics and ENT on the same day. These three principles applied consistently will optimise combined audiovestibular outcomes in labyrinthitis of any aetiology.

VIII. Vestibular Rehabilitation and Central Compensation

Central vestibular compensation — the neuroplastic process by which the brainstem and cerebellum adapt to the asymmetric tonic vestibular input produced by a unilateral peripheral deafferentation — is the mechanism underlying clinical recovery from acute labyrinthitis [35,47]. It proceeds through three overlapping processes: static compensation (restoration of resting symmetry, resolution of spontaneous nystagmus and postural instability at rest); dynamic compensation (restoration of VOR during head movement, driven by cerebellar-mediated VOR adaptation); and substitution (use of visual, somatosensory, and cognitive strategies to replace the lost vestibular signal). The vestibular physician's role is to facilitate this process and to identify the minority of patients in whom compensation is impaired or insufficient [35,47].

Mechanisms and prerequisites for compensation

Static compensation is largely driven by changes in resting membrane potential of the central vestibular neurons in the ipsilateral vestibular nuclei, modulated by contralateral commissural inhibition, cerebellar input, and neural sprouting [35,47]. It proceeds rapidly in the first one to two weeks. Dynamic compensation is slower and experience-dependent: repeated activated gaze stabilisation during head movement drives cerebellar adaptive plasticity, which progressively restores VOR gain toward normal. Patients who remain still, rest in darkened rooms, and take vestibular suppressants beyond 48–72 hours experience slower dynamic compensation and prolonged disability. This is the biological basis of the well-established instruction to move early and stop suppressants promptly [46,47].

Vestibular rehabilitation therapy

VRT for acute labyrinthitis and vestibular neuritis is supported by a Cochrane review (McDonnell and Hillier, 2015) demonstrating moderate-quality evidence that VRT is more effective than control conditions for reduction of subjective dizziness, improvement of balance, and return to function in the medium and long term [48]. Early mobilisation should be encouraged from the second day of illness where tolerated, and formal VRT referral to a vestibular physiotherapist with labyrinthitis-specific experience is indicated in all patients who have not returned to normal function by two to three weeks [47,48].

The evidence-based VRT programme for unilateral vestibular deafferentation comprises four pillars: gaze stabilisation exercises (horizontal and vertical VOR x 1 exercises with the head moving while the eye fixates a target, progressively increasing head velocity); habituation exercises (repeated exposure to dizziness-provoking movements to reduce motion sensitivity); balance and gait exercises on progressively challenging surfaces (foam, uneven terrain, tandem walking, eyes closed); and aerobic activity to maintain general conditioning and reduce vestibular anxiety [47,48]. Programmes are individualised by the physiotherapist based on baseline vHIT, posturography, and functional assessment.

Persistent postural-perceptual dizziness

Persistent postural-perceptual dizziness (PPPD) develops as a secondary functional syndrome in approximately 25–30% of patients following vestibular neuritis or labyrinthitis, arising from maladaptive central sensitisation of spatial orientation networks after the initial peripheral injury [49,50]. Clinical features — chronic non-episodic unsteadiness, rocking sensation, visual sensitivity, intolerance of complex visual environments, and exacerbation with visual stimulation — persist after the peripheral deafferentation has resolved and must be distinguished from incomplete vestibular compensation [49].

The distinction is made clinically: a patient six weeks post-labyrinthitis with a negative HIT, resolved spontaneous nystagmus, and improving vHIT who nonetheless reports persistent unsteadiness, visual sensitivity, and rock-on-a-boat sensation is developing PPPD, not failing vestibular compensation. Management shifts to PPPD-specific VRT (visual desensitisation, exposure-based protocols), patient psychoeducation, and SSRI pharmacotherapy — with sertraline or venlafaxine providing the strongest evidence base in PPPD [49,50]. Repeated vestibular investigations at this stage are counterproductive.

□ **Clinical Insight:** Vestibular suppressants beyond 72 hours are rehabilitation inhibitors. The patient must experience residual dizziness during gaze stabilisation exercises for cerebellar adaptation to occur. Prescribing prochlorperazine for weeks after acute labyrinthitis is, in effect, prescribing chronic vestibular disability.

IX. Prognosis, Complications, and Special Populations

Prognosis in labyrinthitis depends critically on aetiology, the presence of cochlear involvement, the timeliness of treatment, and the patient's capacity for central vestibular compensation. Vestibular and auditory recovery must be assessed independently — they may diverge substantially in the same patient.

Vestibular prognosis

The vast majority of patients with viral labyrinthitis achieve functional vestibular recovery within three to six months, driven by central compensation even when peripheral canal paresis persists [7,35]. Objective

vHIT gain may remain reduced in the affected ear indefinitely — partial or complete peripheral recovery occurs in approximately 40–60% at one year — but the functional consequence of persistent canal paresis depends more on compensation adequacy than on the degree of peripheral recovery [35,39]. Age is the strongest predictor of functional vestibular outcome: patients over 65 years compensate more slowly, are more vulnerable to falls, and may require longer supervised VRT. Pre-existing visual or somatosensory impairment limits the substitution strategies available [35].

Auditory prognosis

Hearing recovery in viral labyrinthitis is substantially better than in bacterial disease. In large sudden SNHL cohorts, approximately 45–65% of patients achieve partial or complete recovery with corticosteroid treatment, with the best outcomes in mild-to-moderate losses and low-frequency patterns [7,14,27]. Severe-to-profound initial hearing loss (greater than 70 dB pure tone average) recovers in fewer than 30% of cases [14,27]. The recovery window is largely complete by three months — spontaneous recovery beyond this point is uncommon, and the three-month audiogram is the practical landmark for cochlear implant candidacy decisions.

In bacterial suppurative labyrinthitis — tympanogenic or meningogenic — permanent profound SNHL is the expected outcome in the majority. Bilateral profound SNHL following meningitis occurs in approximately 6–10% of bacterial meningitis survivors in the pre-dexamethasone era, and 2–4% with adjunctive dexamethasone [10,29]. The absolute numbers still represent a significant population requiring cochlear implant services.

Table 7. Prognosis by aetiology — vestibular and auditory outcomes.

Aetiology	Vestibular recovery	Hearing recovery	Key prognostic driver
Viral (HSV-1 / idiopathic)	Functional recovery: 80–90% at 6 months [7,35]	60–80% partial/complete at 3 months [7,14,27]	Initial severity; treatment timing [27]
Ramsay Hunt (VZV)	Functional recovery: 60–75% [28]	50–65% recovery; poorer than HSV-1 [28]	Antiviral initiation within 72 h [28]
Bacterial — tympanogenic	Compensation usual; peripheral loss permanent [5]	Permanent moderate-to-profound in majority [5,8]	Speed of surgical source control [5]
Bacterial — meningogenic	Compensation usual; bilateral risk possible [10]	Permanent bilateral profound in 6–10% without dexamethasone [10,11,29]	Dexamethasone timing; CI referral speed [10,12]

Labyrinthitis ossificans

Labyrinthitis ossificans (LO) is the fibro-osseous obliteration of the membranous labyrinth following suppurative inflammation, representing the end stage of the labyrinthine inflammatory cascade [22,51]. The pathological sequence — exudate, fibrous organisation, new bone formation — proceeds over weeks to months following bacterial meningitis or suppurative labyrinthitis. On MRI, loss of the normal T2-bright signal in the cochlear turns (beginning at the basal turn) is the earliest radiological marker, often apparent within two to four weeks of the acute event. CT demonstrates frank ossification later — typically four to twelve weeks — and assesses the degree of cochlear lumen obliteration for surgical planning [22,36,51].

The cochlear implantation implications of LO are paramount. As new bone progresses to occlude the scala tympani, standard electrode insertion becomes impossible and requires drill-out, split-array electrode, or brainstem implantation. Every month of delay beyond four to six weeks from meningitis onset decreases the likelihood of standard implantation. The vestibular physician who identifies early T2 signal loss on MRI must immediately expedite the cochlear implant pathway — not a routine referral but an urgent surgical queue [12,22,51].

Special populations

In children, post-meningitic labyrinthitis with bilateral profound SNHL is the most consequential special population. Language acquisition depends on auditory input; delays in implantation degrade speech and language outcomes. Early identification (ABR in the NICU), urgent imaging, and streamlined referral to a paediatric cochlear implant centre are the clinical priorities. Sequential bilateral cochlear implantation —

with simultaneous implantation in some centres where ossification is progressing rapidly — is the standard of care [10,12].

Immunocompromised patients — receiving immunosuppression, with HIV, haematological malignancy, or post-solid-organ transplantation — are vulnerable to unusual viral causes (CMV, EBV, VZV dissemination) and may present with more severe, bilateral, or treatment-resistant labyrinthitis. The threshold for empirical antiviral therapy is lower in this group, and viral titres from serum and, where indicated, CSF should be obtained before treatment initiation [28,52]. In HIV-positive patients, labyrinthitis may reflect opportunistic infection (cryptococcal meningitis producing meningogenic labyrinthitis); the CD4 count is an essential contextual investigation.

□ **Important:** Post-meningitic bilateral deafness in a child is an audiological and surgical emergency. Any delay beyond 6–8 weeks increases the risk of labyrinthitis ossificans advancing to occlude the scala tympani. Urgent MRI (within 4 weeks) and referral to a paediatric cochlear implant centre must occur before the T2 signal is fully obliterated [10,12,22].

X. Guidelines, Controversies and Future Directions

Labyrinthitis sits at the intersection of two well-resourced guideline domains — sudden SNHL and vestibular neuritis — but lacks its own dedicated evidence-based guideline from any major vestibular or otological society. The AAO-HNS 2019 Clinical Practice Guideline on Sudden Hearing Loss applies to the cochlear component; the Barany Society framework addresses the vestibular component; but neither specifically governs the combined syndrome [1,32,45]. This creates a guideline gap that practising clinicians must navigate through synthesis of the two domains.

Guideline landscape

The AAO-HNS 2019 guideline on sudden SNHL provides the following key evidence-based recommendations applicable to labyrinthitis: distinguish SNHL from conductive loss at presentation; offer corticosteroids within two weeks of onset; consider salvage intratympanic steroids for incomplete recovery at four to six weeks; do not routinely prescribe antivirals; do not routinely use vasodilators, antioxidants, or anticoagulants [32]. The European Position Statement on vestibular neuritis (2016) includes an expert-level comment that labyrinthitis — defined as combined audiovestibular loss — should receive the same corticosteroid protocol as vestibular neuritis, with additional hearing monitoring [3,45]. No society has yet published a stand-alone labyrinthitis guideline.

Steroid timing and route — ongoing debate

Whether to use systemic versus intratympanic steroids as the primary route remains debated. The Rauch 2011 NEJM trial demonstrated non-inferiority of intratympanic methylprednisolone versus oral prednisolone for sudden SNHL at three months — with the practical implication that intratympanic delivery is a genuine alternative for patients in whom systemic steroids are contraindicated [27]. However, labyrinthitis involves both cochlear and vestibular end-organs, and there is no intratympanic route equivalent for the vestibular component — the anatomy of the perilymph does not guarantee vestibular-ganglia-level drug delivery with transtympanic injection alone. Systemic corticosteroids therefore remain the preferred primary route for labyrinthitis, with intratympanic ITS reserved as salvage or where systemic steroids are contraindicated [27,42,44].

Antiviral therapy — closing the evidence gap

The Strupp 2004 trial remains the definitive study on antivirals for vestibular neuritis, but its findings are not directly transferable to labyrinthitis. The mechanistic rationale for antiviral therapy is stronger in labyrinthitis — cochlear and vestibular ganglionic involvement is a plausible substrate for antiviral benefit, and Ramsay Hunt data clearly support antiviral use for VZV reactivation in the inner ear [28,43]. A well-powered randomised trial of valacyclovir versus placebo added to systemic steroids in acute labyrinthitis with defined audiological and vestibular outcomes at three and twelve months would resolve this question definitively. Such a trial remains unfunded as of 2026.

Cochlear implantation in labyrinthitis ossificans

The cochlear implant surgery literature has substantially extended options for post-meningitic patients with LO. Split-electrode arrays, cochlear drill-out procedures to remove new bone, and auditory brainstem implantation in complete ossification represent the evolving surgical toolkit [12,51]. Outcomes in partially ossified cochleas approach those of non-ossified implantation when electrode coverage exceeds 12–15 mm; when limited to short insertion, speech perception outcomes are inferior but remain far superior to unaided bilateral profound deafness [12,51].

Future directions — regeneration and vestibular implant

Hair-cell regeneration in the mammalian cochlea and vestibular end-organs is an active and rapidly evolving research area [53]. Gene therapy approaches targeting the Atoh1 transcription factor — the master regulator of hair-cell fate — have restored some cochlear function in animal models of ototoxic hair-cell loss and show proof-of-concept in early clinical trials. The application to post-labyrinthitis SNHL is a medium-term clinical horizon rather than current practice [53]. For bilateral vestibular labyrinthitis with complete deafferentation, the vestibular implant — delivering electrical stimulation to the ampullary nerves of the semicircular canals — has demonstrated proof-of-concept VOR restoration in small clinical trials and represents a meaningful future therapeutic option for the most disabled patients who fail VRT [54].

□ **Key Point:** Labyrinthitis lacks a dedicated clinical guideline. Vestibular physicians must synthesise the AAO-HNS sudden SNHL guideline (cochlear component) and the Barany Society vestibular neuritis framework (vestibular component), applying both in parallel. Advocacy for a dedicated labyrinthitis guideline through the Barany Society or AAO-HNS should be a priority for the field.

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