Autoimmune Inner Ear Disease (AIED):

A Vestibular Physician's Deep Review of Immune-Mediated Audiovestibular Dysfunction

Vestibular Medicine for Vestibular Physicians

Peripheral Vestibular Pathology — Module 2.9

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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, History and Epidemiology

Autoimmune inner ear disease (AIED) is an uncommon but distinctive cause of audiovestibular dysfunction defined by rapidly progressive, frequently fluctuating, and characteristically bilateral sensorineural hearing loss (SNHL) that responds, at least initially, to immunosuppression [1,2,13,14]. For the vestibular physician it occupies a strategically important niche: it is one of the few causes of progressive cochleovestibular failure that is genuinely treatable, yet it has no pathognomonic test, no validated diagnostic criteria, and a natural history that is easily mistaken for atypical Ménière's disease, bilateral vestibulopathy, or idiopathic sudden deafness [8,12,14]. Maintaining a low threshold of suspicion is therefore the single most important determinant of outcome [12,35].

The concept of immune-mediated hearing loss was first articulated by Lehnhardt in 1958, who proposed that anti-cochlear antibodies might explain sequential bilateral progressive deafness [2,7]. The field was transformed in 1979 when McCabe reported eighteen patients with idiopathic progressive bilateral SNHL whose hearing improved on corticosteroids and cyclophosphamide, coining the term "autoimmune sensorineural hearing loss" and establishing that some apparently irreversible losses were medically reversible [1]. Hughes and colleagues subsequently proposed the first working clinical criteria, emphasising the bilateral, progressive, steroid-responsive phenotype [3].

Immunological confirmation followed in the 1980s and 1990s. The inner ear, long regarded as immune-privileged behind the blood-labyrinth barrier, was shown to mount competent immune responses, with the endolymphatic sac identified as the principal site of antigen processing [7,18]. Harris and Sharp detected antibodies against inner-ear proteins in patients with rapidly progressive SNHL [2], and Moscicki's landmark JAMA study correlated a 68-kDa inner-ear antigen — later identified as heat-shock protein 70 (HSP70) — with disease activity and corticosteroid responsiveness [4,24]. These observations consolidated AIED as a discrete clinical entity rather than a diagnosis of exclusion alone [4,12,16].

Epidemiologically, AIED is rare. It is generally estimated to account for under 1% of all cases of hearing loss and dizziness, with a frequently quoted point prevalence near 15 per 100 000, although this figure is poorly validated and almost certainly conflates primary and secondary forms [13,14]. It predominantly affects adults between the third and sixth decades, with a consistent female preponderance, mirroring the demographic skew of systemic autoimmunity [10,12,15]. Because the disease progresses over weeks to months, untreated patients commonly reach severe bilateral impairment within around three years of onset [12,14].

Table 1. Autoimmune inner ear disease at a glance.

Feature	Typical value / pattern	Notes
Share of all SNHL	Under 1%	Higher in tertiary neuro-otology referral cohorts [13,14]
Estimated prevalence	~15 / 100 000 (poorly validated)	Conflates primary and secondary forms [13,14]
Peak age of onset	20–60 years	Mirrors systemic autoimmunity demographics [10,12]
Sex ratio	Female predominance	Consistent across cohorts [12,15]
Laterality	Bilateral (may begin unilateral)	Bilaterality is a defining clinical clue [1,12]
Secondary (systemic) form	~15–30% of cases	Associated systemic autoimmune disease [22,23,24]

□ **Key Point:** AIED is rare but treatable. Because no single test confirms it and its phenotype overlaps Ménière's disease and bilateral vestibulopathy, outcome depends almost entirely on the clinician maintaining active suspicion in any bilateral, progressive, or steroid-responsive audiovestibular presentation [12,14,35].

A clinically vital distinction is drawn between primary AIED, in which the immune process is confined to the labyrinth, and secondary AIED, in which audiovestibular involvement is one manifestation of a

systemic autoimmune disorder [22,23,24]. Roughly 15–30% of cases are secondary, and the proportion rises in cohorts screened systematically for connective-tissue disease [22,23]. Vestibular symptoms are common across systemic autoimmunity: in systemic lupus erythematosus, dizziness or vertigo is reported in a large minority of patients over the disease course, and abnormal vestibular testing is found in the majority of patients with active connective-tissue disease when sought [22,25]. Cogan's syndrome — non-syphilitic interstitial keratitis with audiovestibular failure, first described in 1945 — is the prototypical organ-overlap syndrome and a not-to-be-missed cause of severe deafness and vestibulopathy in young adults [43,44].

□ **Clinical Insight:** AIED accounts for a small fraction of hearing loss in general otology but is over-represented in tertiary dizziness and hearing-loss clinics because of referral bias. The vestibular physician will therefore see it more often than population estimates suggest, and should treat bilateral progressive cochleovestibular failure as AIED until proven otherwise [13,14].

II. Pathophysiology — Immune Mechanisms and Loss of Inner-Ear Tolerance

The inner ear is not, as once thought, absolutely immune-privileged. It is sequestered behind the blood-labyrinth barrier — a tight-junctioned analogue of the blood-brain barrier that ordinarily excludes large molecules and circulating leucocytes — yet it retains the machinery for a competent local immune response [7,18,25]. The endolymphatic sac is the immunological hub of the labyrinth, capable of antigen uptake, processing and presentation, and of recruiting systemic immune effectors into perilymph [7,18]. Resident perivascular and scala-tympani macrophages, identified by Iba1 expression, populate the cochlea constitutively and proliferate after insult, providing the antigen-presenting and cytokine-releasing substrate for an autoimmune reaction [10,11,26].

The blood–labyrinth barrier and inner-ear immune anatomy

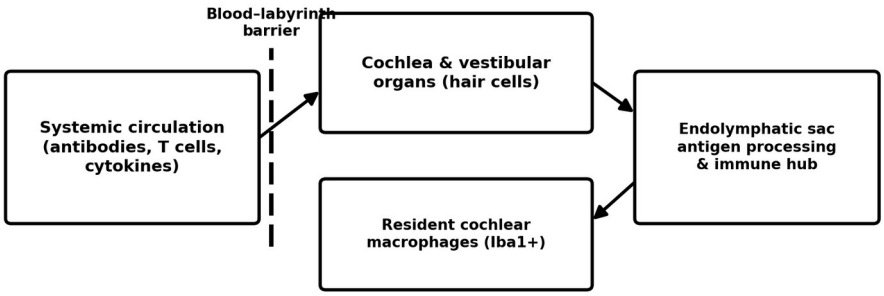

Figure 1. The blood-labyrinth barrier and the immunological anatomy of the inner ear.

Loss of immunological tolerance to inner-ear antigens is the central event. Several non-exclusive mechanisms are invoked [7,12,13]. In molecular mimicry, an environmental trigger — most often a viral infection — generates antigens structurally similar to inner-ear proteins, so that cross-reactive antibodies or T cells subsequently attack the labyrinth [7,12]. In the bystander mechanism, a primary insult (infection, trauma, ischaemia or noise) breaches the blood-labyrinth barrier and releases pro-inflammatory cytokines such as interleukin-1 (IL-1) and tumour necrosis factor-alpha (TNF-α), non-specifically activating local immunity [10,21,33]. Hidden- or sequestered-antigen exposure follows barrier disruption, presenting normally concealed proteins — cochlin, type II collagen, β-tectorin and HSP70 — to a naïve immune system [4,8,12,23]. Overlaid on these is a genetic and systemic propensity, with HLA-DR associations and a high background rate of coexisting autoimmunity [10,15,24].

□ **Clinical Insight:** The blood-labyrinth barrier is not a static wall. Infection, trauma, noise and inflammation transiently disrupt it, allowing antigen exposure and leucocyte entry. This explains

why AIED so often follows an apparently trivial precipitant, and why a breach in one ear can prime a cross-reactive attack on the contralateral, as-yet-asymptomatic ear [7,10,11].

Both humoral and cell-mediated arms contribute to injury [12,27]. Autoantibodies against HSP70, cochlin, type II collagen and S-100β have been detected in patient sera, and may form immune complexes within labyrinthine fluids that fix complement and damage the stria vascularis and hair cells [4,5,8,23,24]. The murine work of Solares and colleagues was pivotal in demonstrating that CD4+ T cells specific for inner-ear peptides can transfer autoimmune hearing loss, establishing a directly pathogenic cell-mediated component rather than a purely antibody-driven process [27]. Cytokine networks — particularly IL-1β and TNF-α — are increasingly seen as the effector common pathway, a view supported by the therapeutic response of corticosteroid-resistant disease to IL-1 blockade [21,22,33].

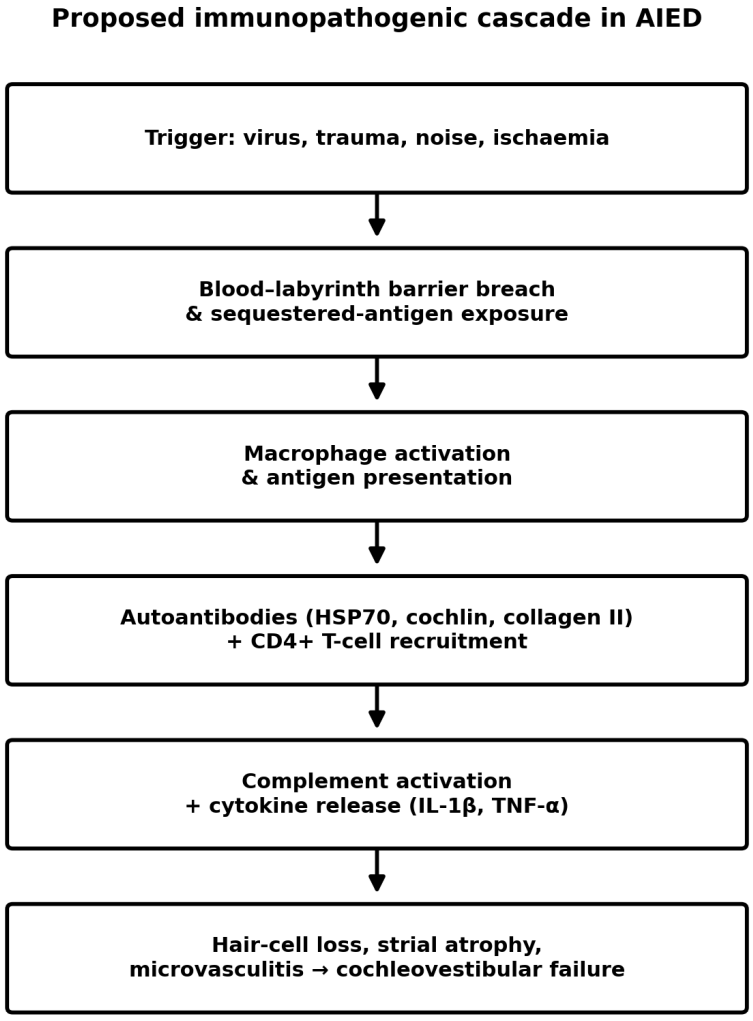

Figure 2. Proposed immunopathogenic cascade in autoimmune inner ear disease.

Histopathology, though scarce because temporal bones are rarely obtained during active disease, supports these mechanisms. Reported findings include lymphocytic infiltration of the endolymphatic sac and cochlea, fibrosis and new bone formation within the cochlear duct, atrophy of the stria vascularis, hair-cell and neuronal loss, and obliterative vasculitis of the labyrinthine microvasculature [7,12,18]. The cochleovestibular distribution of these changes parallels the clinical observation that hearing loss dominates the picture while vestibular end-organ involvement, when present, produces bilateral hypofunction and chronic disequilibrium rather than discrete attacks [12,23,25].

□ **Key Point:** The final common pathway in all forms of AIED is immune-mediated cochleovestibular injury: barrier breach and antigen exposure, macrophage activation and antigen presentation, autoantibody production and CD4+ T-cell recruitment, complement

activation and cytokine release, culminating in hair-cell loss, strial atrophy and microvasculitis [7,10,21,27].

Table 2. Proposed immunopathogenic mechanisms and their clinical correlates.

Mechanism	Putative process	Clinical correlate
Molecular mimicry	Viral antigen cross-reacts with inner-ear protein	Post-viral onset; second-ear involvement [7,12]
Bystander activation	Barrier breach releases IL-1/TNF-α	Onset after trauma, noise, infection [10,21]
Hidden-antigen exposure	Sequestered cochlin/HSP70/collagen revealed	Detectable autoantibodies [4,8,23,24]
Cell-mediated attack	CD4+ T cells specific for ear peptides	Transferable disease in models [27]
Genetic/systemic propensity	HLA-DR; coexisting autoimmunity	Female skew; secondary AIED [10,15,24]

Considerable effort has gone into identifying the responsible autoantigen, with cochlin, beta-tectorin, type II collagen, the 68-kDa/HSP70 protein and S-100beta all proposed, yet none has emerged as a single dominant target — a heterogeneity that mirrors the clinical variability of the disease and frustrates development of a specific assay [4,7,8,12]. Experimental models have nonetheless been informative: immunisation of animals with crude inner-ear antigen or defined peptides reproducibly induces hearing loss with histology resembling human disease, and adoptive-transfer experiments confirmed that antigen-specific CD4+ T cells are sufficient to cause it [9,27]. The contemporary synthesis frames AIED less as a single antibody-mediated disease and more as a cytokine-driven inflammatory process in which innate immunity — resident macrophages, the inflammasome and IL-1beta — amplifies an adaptive trigger, a model that directly motivated therapeutic targeting of IL-1 in corticosteroid-resistant disease [11,16,21,37].

This innate-adaptive interplay also helps explain two clinically familiar observations. The frequent post-viral or post-traumatic onset fits a bystander mechanism in which barrier breach and cytokine release precede any antigen-specific response, while the female preponderance and the high background rate of coexisting autoimmunity point to a systemic loss of self-tolerance on which the labyrinth-specific attack is superimposed [10,12,15,24]. The practical corollary is that AIED should be conceived as the audiovestibular expression of a broader immune dysregulation rather than an isolated end-organ accident [16,22,24].

III. Clinical Features — Audiovestibular Presentation and Systemic Associations

The cardinal presentation of AIED is bilateral, progressive sensorineural hearing loss evolving over weeks to a few months [1,12,14]. This tempo is the most useful single discriminator: it is too fast for presbycusis or most genetic losses, yet too slow for the abrupt deficit of idiopathic sudden SNHL or vestibular neuritis [1,14,29]. Disease may begin unilaterally before involving the second ear, but established bilaterality — frequently asymmetric — is the rule and a key diagnostic clue [1,12]. A unilateral, non-progressive loss should prompt re-evaluation for retrocochlear pathology rather than a default AIED label [12,14].

□ **Clinical Pearl:** The constellation of bilateral progressive SNHL, vestibular symptoms and fluctuation in hearing, particularly with documented partial recovery after a corticosteroid course, is highly suggestive of AIED. The tempo — weeks to months — is the discriminator: faster suggests sudden SNHL, slower suggests presbycusis or genetic loss [1,12,14].

Vestibular involvement is reported in roughly a quarter to a half of patients at presentation and ranges from discrete episodic vertigo through to chronic disequilibrium and oscillopsia from bilateral vestibular

hypofunction [12,20,23]. Because the immune injury tends to be bilateral and indolent, the vestibular phenotype more often resembles progressive bilateral vestibulopathy than the discrete attacks of Ménière's disease, although a Ménière-like fluctuating picture ("autoimmune hydrops") is well recognised [20,23,46]. Tinnitus and aural fullness are common, and roughly half of patients describe fluctuation in hearing that may transiently respond to steroids [12,14].

On audiometry the typical finding is bilateral SNHL that is often asymmetric; high-frequency involvement is common but any configuration may occur, and early thresholds characteristically fluctuate [12,30]. Speech-discrimination scores frequently deteriorate out of proportion to pure-tone thresholds, a pattern that signals retro- or intra-cochlear neural involvement and carries prognostic weight [12,30]. Vestibular laboratory testing reveals abnormality in a high proportion of patients with active systemic autoimmunity — most often bilateral caloric weakness — underscoring that subclinical vestibular involvement is more common than the symptom history suggests [22,25].

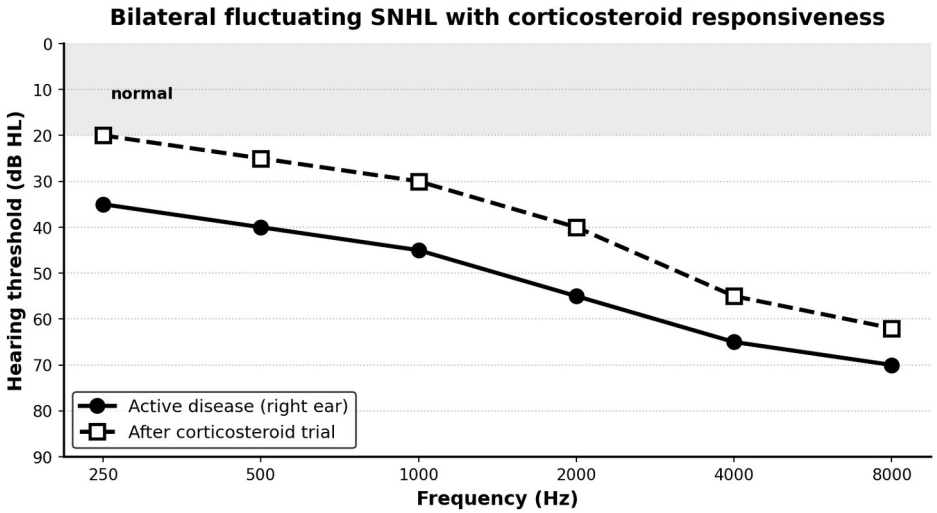

Figure 3. Bilateral fluctuating sensorineural hearing loss with corticosteroid responsiveness.

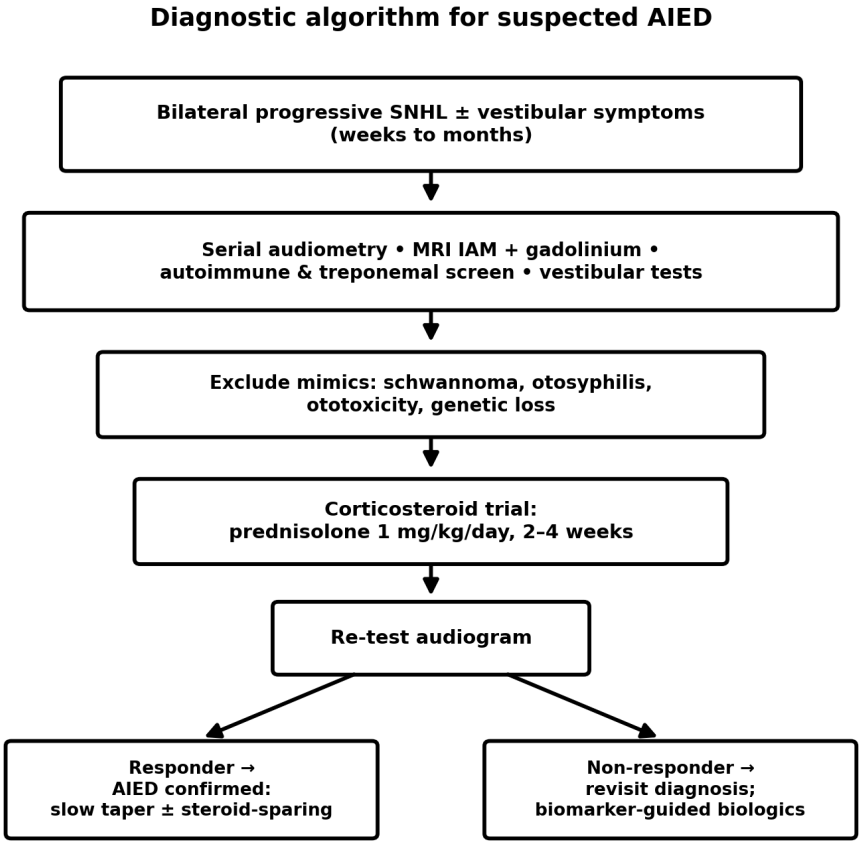

Figure 4. A practical diagnostic algorithm for suspected autoimmune inner ear disease.

Systemic associations

Because up to a third of patients have, or will develop, a systemic autoimmune disorder, a directed review of systems is mandatory at every AIED assessment [22,23,24]. In primary disease the patient is typically otherwise well, though fatigue, low-grade fever and arthralgia may appear during active phases [14,22]. In secondary disease the features of the underlying condition usually dominate or precede the audiovestibular symptoms [22,23,43].

- **Cogan's syndrome:** interstitial keratitis with audiovestibular failure; large-vessel vasculitis may supervene and demands urgent systemic immunosuppression [43,44].
- **Systemic lupus erythematosus:** arthralgia, photosensitive rash, serositis; coexisting antiphospholipid antibodies raise audiovestibular risk through microthrombosis [22,25,47].
- **Granulomatosis with polyangiitis and other ANCA vasculitides:** otological involvement is common and may be the presenting feature; ANCA screening is essential [22,47].
- **Rheumatoid arthritis and other connective-tissue disease:** joint and systemic symptoms usually precede or accompany hearing loss [22,25,47].

□ **Key Point:** Approximately one-third of AIED patients have or will develop an identifiable systemic autoimmune disease. Screening for systemic autoimmunity is not optional — the audiovestibular system can be the sentinel presentation of lupus, vasculitis or Cogan's syndrome [22,23,24].

IV. Diagnostic Criteria and the Place of the Steroid Trial

There is no internationally ratified diagnostic standard for AIED, and this remains the field's central weakness [8,12,14]. In practice diagnosis rests on integrating a compatible clinical picture, objective audiovestibular dysfunction, documented response to immunosuppression, and rigorous exclusion of mimics [1,3,12]. The Hughes criteria, derived from McCabe's original observations, remain the most widely used working framework and emphasise the bilateral, progressive, steroid-responsive phenotype [1,3].

Table 3. Working diagnostic criteria for autoimmune inner ear disease.

Domain	Requirement
Hearing pattern	Bilateral SNHL, usually progressive over weeks to months [1,3]
Tempo	Rapid progression — excludes presbycusis and congenital loss [1,14]
Objective dysfunction	Audiometric and/or vestibular abnormality documented [3,12]
Treatment response	Improvement or stabilisation on corticosteroids/immunosuppression [1,4,30]
Exclusion	Infective, genetic, neoplastic, metabolic and structural causes excluded [12,14]
Supportive (not required)	Positive inner-ear serology or recognised systemic autoimmune disease [4,22,24]

Two caveats must be held firmly. First, positive inner-ear antibody serology supports the diagnosis but negative serology never excludes it; AIED remains a clinical diagnosis [4,8,24]. Second, the presence of a defined systemic autoimmune disease strengthens a diagnosis of secondary AIED but its absence does not refute primary disease [22,23]. A subset of patients also satisfy criteria for Ménière's disease — episodic vertigo, aural fullness and hydrops on delayed-contrast MRI — the so-called autoimmune hydrops, which sits at the diagnostic boundary between the two conditions [23,46].

□ **Clinical Insight:** The corticosteroid trial is simultaneously diagnostic and therapeutic. A measurable improvement or stabilisation of hearing within four to six weeks of adequate oral corticosteroid therapy is strongly suggestive of an immune aetiology — but it is not specific, because sudden SNHL and Ménière's may also respond [1,4,30].

The steroid trial is the practical cornerstone of diagnosis. A typical regimen is oral prednisolone 1 mg/kg/day (to a maximum of 60–80 mg) for two to four weeks, with the response judged on serial audiometry before any taper [1,4,16,30]. An audiometric improvement — conventionally a gain of 10 dB or more at two contiguous frequencies, or a 12% or greater improvement in speech-discrimination score — defines a responder [4,30]. Crucially, the trial must be assessed objectively and serially; subjective impressions of hearing are unreliable in fluctuating disease, and a hasty taper before the response is documented invites both diagnostic error and relapse [16,30].

□ **Clinical Pearl:** Judge the steroid trial on the audiogram, not the patient's impression. Document baseline thresholds and speech discrimination, treat at full dose for two to four weeks, and re-test before tapering. A 10 dB gain at two frequencies or a 12% speech-discrimination improvement defines a responder [4,30].

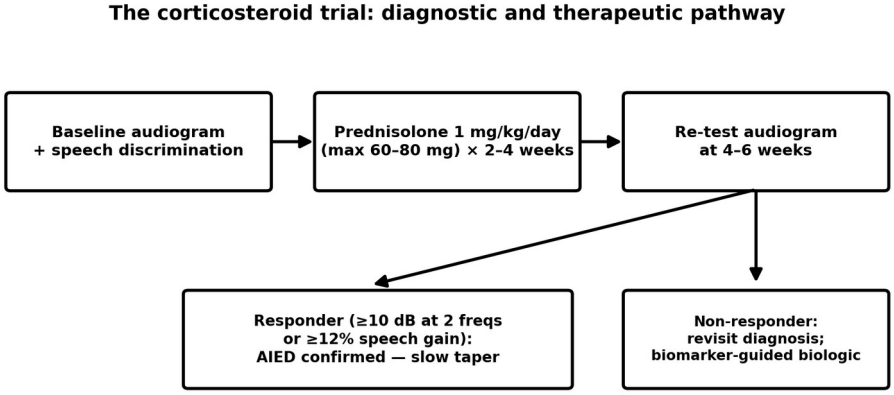

Figure 5. The corticosteroid trial as a combined diagnostic and therapeutic pathway.

V. Investigations: Audiometry, Vestibular Testing, Serology and Imaging

Investigation in suspected AIED serves three purposes: to document and quantify cochleovestibular dysfunction, to track the response to treatment, and to exclude the broad differential of bilateral progressive SNHL [12,14]. No single test is confirmatory, and over-investigation rarely changes management; tests should be ordered against a specific question [8,14].

Audiological assessment

Pure-tone and speech audiometry are mandatory at baseline and must be repeated serially — fluctuation and steroid responsiveness are demonstrated, not assumed [4,30]. Speech-discrimination loss disproportionate to pure-tone thresholds is characteristic and prognostically adverse [12,30]. Otoacoustic emissions provide a sensitive early index of outer-hair-cell function and may change rapidly during active disease, while wideband acoustic immittance confirms normal middle-ear mechanics and excludes a conductive overlay [12,14].

Vestibular testing

Caloric testing and the video head impulse test quantify canal function and, importantly, document bilateral involvement that may be clinically silent [22,25]. Bilateral caloric weakness is the most frequently reported abnormality in patients with active connective-tissue disease, and serial testing helps distinguish progressive immune vestibulopathy from a stable deficit [22,25]. Cervical and ocular vestibular-evoked myogenic potentials add otolith-organ information and may reveal asymmetries supporting labyrinthine immune injury [20,25].

Table 4. Investigation panel in suspected autoimmune inner ear disease.

Investigation	Purpose	Comment
Serial pure-tone & speech audiometry	Document SNHL, fluctuation, steroid response	Mandatory; the diagnostic backbone [4,30]
Otoacoustic emissions	Sensitive outer-hair-cell index	May change rapidly in active disease [12]
Caloric / video head impulse test	Quantify and lateralise vestibular loss	Detects subclinical bilateral involvement [22,25]
cVEMP / oVEMP	Assess otolith-organ function	Adjunct in atypical disease [20,25]
Inner-ear serology (anti-HSP70 etc.)	Support diagnosis	Sensitivity ~50%; negative does not exclude [4,24]
Systemic autoimmune screen	Detect secondary AIED	ANA, ANCA, RF, ESR/CRP, complement, TFTs [22,23,47]
MRI internal auditory meati +	Exclude retrocochlear/structural	Mandatory at first presentation

gadolinium	mimics	[12,14]

Serological testing has two tiers. Inner-ear-specific antibodies — historically the Western-blot anti-68-kDa/HSP70 assay — are positive in around half of clinically active patients and were associated in early work with corticosteroid responsiveness, but their specificity is limited and the assay is not widely available [4,24]. The second tier is a systemic autoimmune screen — antinuclear antibody, ANCA, rheumatoid factor, ESR and CRP, complement levels, antiphospholipid antibodies and thyroid function with thyroid antibodies — directed at identifying secondary AIED, which materially changes management [22,23,47].

□ **Important:** Negative inner-ear antibody serology does not exclude AIED, and a positive result does not confirm it. Diagnosis remains clinical, anchored on the bilateral progressive phenotype and the documented response to immunosuppression. Do not withhold a steroid trial on the basis of negative serology [4,8,24].

Imaging centres on MRI of the internal auditory meati with gadolinium, mandatory at first presentation to exclude vestibular schwannoma, endolymphatic-sac tumour, demyelination and other retrocochlear or central pathology [12,14]. High-resolution CT of the temporal bones is reserved for suspected structural disease such as third-window lesions or cochlear ossification, the latter being relevant to cochlear-implant planning [12,41]. Delayed post-gadolinium 3D-FLAIR can demonstrate endolymphatic hydrops in the subset with an autoimmune-hydrops phenotype and is emerging as a means of documenting labyrinthine inflammation, though it remains a research-adjacent tool [33,46].

Two practical pitfalls deserve emphasis. First, a single audiogram cannot establish or exclude AIED: the diagnosis depends on demonstrating change over time, so the temptation to act on one study should be resisted in favour of short-interval serial testing during the steroid trial [4,30]. Second, the inner-ear antibody assay is neither standardised across laboratories nor uniformly available, and a negative or unobtainable result must never delay treatment in a compatible clinical picture [4,8,24]. Electrocochleography and delayed-contrast MRI may show an autoimmune-hydrops pattern in the Meniere-overlap subgroup, but neither is diagnostic in isolation and both are best reserved for atypical cases where the differential genuinely includes Meniere disease, in whom raised anti-HSP70 titres have themselves been reported [6,33,46].

□ **Clinical Pearl:** Investigate to answer a question, not to reassure. The only test mandatory in every case is serial audiometry; MRI is mandatory once to exclude structural mimics; everything else — VEMPs, ECochG, inner-ear serology, delayed-FLAIR hydrops imaging — is selective and rarely changes the decision to trial corticosteroids [4,8,14].

VI. Differential Diagnosis

The differential diagnosis of bilateral progressive SNHL with vestibular symptoms is broad, and systematic exclusion is part of the diagnosis of AIED rather than a separate exercise [12,14]. The conditions that most often masquerade as AIED — or are masqueraded by it — are Ménière's disease, idiopathic sudden SNHL, bilateral vestibulopathy and, critically, the treatable infective and neoplastic mimics [12,14,29].

Table 5. Differential diagnosis of bilateral progressive sensorineural hearing loss with vestibular symptoms.

Condition	Distinguishing features
Ménière's disease	Episodic vertigo (20 min–12 h), fluctuating low-frequency SNHL, aural fullness; usually unilateral at onset; bilateral in 25–45% over time [23,46]
Idiopathic sudden SNHL	Abrupt (under 72 h) loss, more often unilateral; bilateral sudden loss should raise AIED [14,29]
Bilateral vestibulopathy	Oscillopsia and gait ataxia; absent caloric/vHIT responses; may itself be immune-mediated

	[20,25]
Otosyphilis	Can phenocopy AIED or Ménière's; treatable — screen with treponemal serology [12,14]
Vestibular schwannoma / NF2	Progressive unilateral (or bilateral in NF2) loss; confirmed on gadolinium MRI [12]
Ototoxicity	Aminoglycoside, platinum, loop-diuretic exposure; bilateral symmetrical high-frequency loss [12]
Genetic / mitochondrial	Family history, syndromic features; slower tempo, less fluctuation [12]
Susac syndrome	Encephalopathy, branch retinal artery occlusion, SNHL; MRI corpus-callosum lesions [9,20]

Ménière's disease is the most important and most frequent point of overlap. Classical Ménière's is unilateral at onset, with discrete vertigo attacks of twenty minutes to twelve hours and fluctuating low-frequency SNHL; AIED is bilateral and progressive with an indolent or continuous vestibular phenotype [23,46]. The distinction blurs in bilateral Ménière's and in autoimmune hydrops, and a meaningful minority of bilateral Ménière's patients carry markers of systemic autoimmunity, which is why steroid responsiveness and a systemic screen are informative when the picture is atypical [23,46].

□ **Important:** Three treatable mimics must never be missed in bilateral progressive audiovestibular failure: otosyphilis (screen with treponemal serology), a systemic vasculitis such as granulomatosis with polyangiitis (screen with ANCA), and Cogan's syndrome with impending large-vessel involvement. Each requires specific, time-critical therapy distinct from a simple steroid trial [9,43,44,47].

Cogan's syndrome warrants separate emphasis. The combination of interstitial keratitis with rapidly progressive audiovestibular failure in a young adult is characteristic, and the systemic vasculitic form carries a risk of life-threatening aortitis [43,44]. Susac syndrome — the triad of encephalopathy, branch retinal artery occlusions and SNHL — is a further immune-mediated mimic distinguished by characteristic corpus-callosum lesions on MRI [9,20]. Idiopathic bilateral vestibulopathy, increasingly recognised as immune-mediated in a subset, may present with oscillopsia and gait ataxia and absent caloric responses, and should prompt consideration of an immune aetiology when no ototoxic or genetic cause is found [20,25].

VII. Medical Management — Corticosteroids, Steroid-Sparing Agents and Biologics

Management of AIED is staged: prompt high-dose corticosteroid induction, assessment of response on serial audiometry, and — for steroid-dependent or steroid-resistant disease — escalation to steroid-sparing immunosuppression or biologic therapy, all underpinned by hearing and vestibular rehabilitation [15,34,35]. The therapeutic window is narrow; irreversible hair-cell and neuronal loss accrues with delay, so treatment should begin on clinical suspicion rather than await serological confirmation [12,30,35].

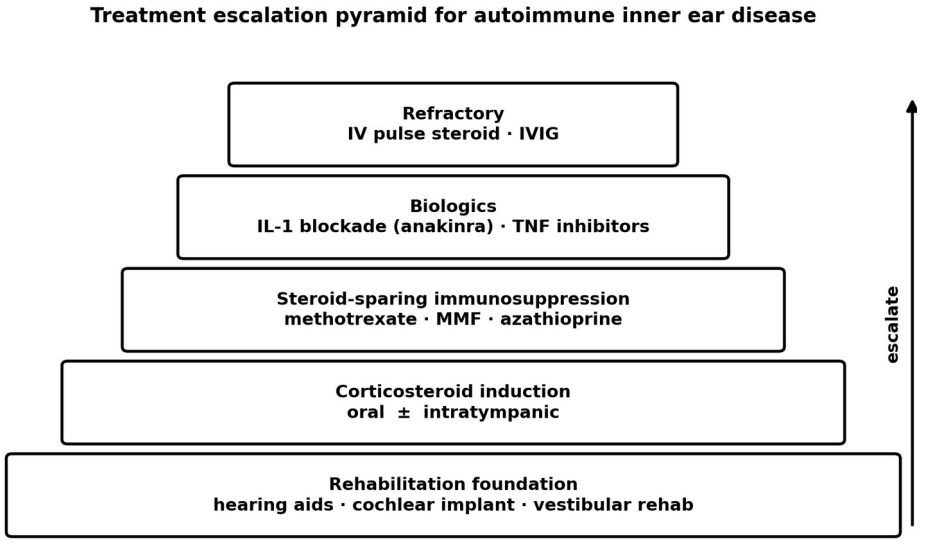

Figure 6. Treatment escalation pyramid for autoimmune inner ear disease.

First-line: corticosteroids

Oral corticosteroids remain the cornerstone of induction. A typical regimen is prednisolone 1 mg/kg/day (maximum 60–80 mg) for two to four weeks at full dose, with the response judged audiometrically before a slow taper over a further six to twelve weeks [1,4,16,30]. Approximately two-thirds of patients achieve partial hearing recovery or stabilisation with initial corticosteroid therapy, and the magnitude of the early response is the strongest single predictor of longer-term outcome [4,30,36]. The mechanism extends beyond immunosuppression: glucocorticoids also act on inner-ear ion and fluid homeostasis through mineralocorticoid as well as glucocorticoid receptors, which may explain responses in disease that is not purely inflammatory [17,38].

□ **Clinical Pearl:** Treat early and at full dose, then taper slowly. Two-thirds of patients respond initially, and the size of that early audiometric response is the best predictor of long-term hearing. A rapid taper is the commonest avoidable cause of relapse [4,30,36].

Intratympanic corticosteroid delivery — dexamethasone or methylprednisolone given by transtympanic injection — achieves high perilymph concentrations while avoiding systemic toxicity, and is valuable when oral steroids are contraindicated or poorly tolerated, in asymmetric disease, or as an adjunct to systemic therapy [40]. Its evidence base in AIED specifically is limited and largely extrapolated from sudden SNHL, but it is a reasonable hearing-targeted option, particularly in only-hearing-ear or steroid-intolerant patients [40].

Steroid-sparing immunosuppression

Because long-term high-dose corticosteroids are toxic and many patients relapse on tapering, steroid-sparing agents are used in steroid-dependent or chronically relapsing disease [15,34]. The pivotal evidence is sobering: the multicentre randomised trial of methotrexate by Harris and colleagues found that methotrexate did not maintain the hearing improvement achieved with prednisone, tempering earlier enthusiasm for its routine use [19]. Methotrexate, azathioprine, mycophenolate mofetil and cyclophosphamide are nonetheless used pragmatically, with cyclophosphamide reserved for severe, rapidly progressive or vasculitic disease given its toxicity [15,29,34].

Table 6. Immunosuppressive and biologic agents in autoimmune inner ear disease.

Agent	Typical regimen	Role / evidence	Monitoring
Prednisolone	1 mg/kg/day, 2–4 wk then taper	First-line induction; ~2/3 respond [1,4,30]	Glucose, BP, bone, mood [39]
Intratympanic steroid	Dexamethasone, weekly x4	Hearing-targeted; steroid-intolerant ears [40]	Otoscopy; TM perforation risk
Methotrexate	10–20 mg weekly	Steroid-sparing; RCT-	FBC, LFTs

		negative for hearing [19]	
Mycophenolate mofetil	0.5–1.5 g twice daily	Steroid-sparing; better tolerated [34]	FBC
Azathioprine	1–2 mg/kg/day	Steroid-sparing alternative [15,34]	FBC, TPMT
Cyclophosphamide	Pulsed IV / oral	Severe, vasculitic or refractory disease [29,34]	FBC; bladder; fertility
TNF-α inhibitors	Infliximab / etanercept	Cogan's and refractory disease; etanercept RCT-negative [20,31]	Infection, TB screen
Anakinra (IL-1 blockade)	100 mg/day SC	Corticosteroid-resistant disease [21,32]	FBC, injection-site, infection

Biologic agents

Biologic therapy is the most active area of AIED research. TNF-α inhibitors showed early promise in case series, particularly in Cogan's syndrome, but the placebo-controlled pilot of etanercept by Cohen and colleagues failed to demonstrate benefit in primary AIED, again cautioning against uncontrolled enthusiasm [20,31]. The most important recent advance is the work on IL-1 blockade: Vambutas and colleagues showed in corticosteroid-resistant AIED that elevated IL-1β predicted response to the IL-1 receptor antagonist anakinra, with measurable audiometric improvement — the first mechanism-targeted, biomarker-guided therapy in the field [21,32]. B-cell depletion with rituximab has a theoretical rationale in antibody-driven disease but lacks robust data [34].

□ **Clinical Insight:** The therapeutic trajectory of AIED mirrors its pathophysiology. Corticosteroids treat the whole inflammatory cascade; the negative methotrexate and etanercept trials show that broad or mistargeted immunosuppression fails; and the positive anakinra signal shows that biomarker-guided, cytokine-specific therapy — here IL-1β blockade in corticosteroid-resistant disease — is the most promising direction [19,20,21,32].

Corticosteroid toxicity is the principal limiting factor of first-line therapy and must be actively managed: hyperglycaemia, hypertension, mood disturbance, gastric irritation, weight gain and, with prolonged courses, osteoporosis and avascular necrosis all accrue with cumulative dose, so the risk-benefit balance shifts steadily against continued high-dose steroid as weeks pass [29,39]. Gastroprotection, bone protection in at-risk patients, glucose monitoring and a defined exit strategy should be planned from the outset rather than improvised at relapse [29,39]. This toxicity profile is precisely why durable disease demands a steroid-sparing partner agent rather than repeated steroid courses [15,34,39].

The optimal duration of induction and the tapering schedule are not standardised, but the principle is consistent: treat at full dose until the audiometric response plateaus, then taper slowly enough to detect early relapse, re-testing hearing at each dose reduction [16,30,36]. Patients who relapse predictably at a given threshold dose are, by definition, steroid-dependent and are the clearest candidates for early introduction of a steroid-sparing or biologic agent rather than indefinite steroid exposure [21,34,36].

VIII. Refractory Disease, Adjuncts and Rehabilitation

A patient who fails to respond to an adequate steroid trial over four to six weeks, or who relapses repeatedly on tapering, has refractory or steroid-dependent disease [15,34]. Before escalating, the clinician should revisit the diagnosis: apparent steroid resistance is frequently mislabelled disease — otosyphilis, schwannoma, genetic or ototoxic loss, or simply too-late presentation with established irreversible damage [12,29]. Where the diagnosis holds, escalation options include pulsed intravenous methylprednisolone, addition of a steroid-sparing agent, biomarker-guided IL-1 blockade, and — in selected refractory cases — intravenous immunoglobulin, although the evidence for IVIG is weak [21,34,42].

□ **Important:** Re-interrogate the diagnosis before treating "refractory AIED". The commonest reasons for apparent steroid failure are an alternative diagnosis (otosyphilis, schwannoma, ototoxicity, genetic loss) and irreversible cochleovestibular damage from delayed presentation. Escalating immunosuppression in a misdiagnosed patient exposes them to toxicity without benefit [12,29,34].

Supportive pharmacotherapy has a defined but limited place. Vestibular suppressants such as prochlorperazine may be used for short periods during acute vertigo but must not be continued, since they impair central compensation and worsen chronic disequilibrium [20]. Bothersome tinnitus is managed with sound enrichment, tinnitus-retraining and cognitive behavioural approaches rather than pharmacotherapy [14].

Hearing rehabilitation

Hearing rehabilitation should run in parallel with disease-modifying treatment rather than waiting for medical failure [35,41]. Appropriately fitted hearing aids, programmable for fluctuating thresholds, transform communication for residual hearing. Cochlear implantation is the definitive option for bilateral severe-to-profound loss and yields good outcomes in AIED, provided the auditory nerve is spared and cochlear patency is preserved — labyrinthine fibrosis and new bone formation from chronic inflammation can complicate electrode insertion, so timely imaging and referral matter [41]. Implantation should be raised proactively in progressive bilateral disease, not held back as a last resort [41].

Vestibular rehabilitation

For patients with established bilateral vestibular hypofunction, customised vestibular rehabilitation promotes central adaptation, substitution and gaze stabilisation, and reduces fall risk and oscillopsia-related handicap [20,25]. Realistic counselling is essential: vestibular hair cells do not regenerate, so late-stage autoimmune vestibulopathy leaves a permanent deficit that rehabilitation improves but does not abolish [20,25]. Fall-risk reduction, workplace accommodation and driving assessment complete the functional package, particularly where vestibular and hearing loss combine [14,25].

The threat of cochlear fibrosis and labyrinthitis ossificans deserves specific mention in implant planning. Chronic labyrinthine inflammation can progressively obliterate the scala tympani, so a patient who is a straightforward implant candidate during active disease may become a far more difficult one if referral is delayed until hearing is lost; serial MRI and timely audiological-implant referral therefore protect future surgical options [41]. Where implantation proceeds, outcomes in AIED are generally good provided the spiral ganglion and auditory nerve are spared, which is usual unless the immune process has extended to the nerve itself [41].

IX. Prognosis, Relapse and Special Populations

The prognosis of AIED is bimodal: early, prompt treatment offers a real chance of recovery, while delayed or refractory disease tends toward progressive, permanent loss [12,30,36]. Approximately two-thirds of patients improve or stabilise with initial corticosteroids, and patients treated within weeks of onset fare substantially better than those with long-standing untreated disease [4,30,36]. The early audiometric response remains the most useful prognostic marker [4,30].

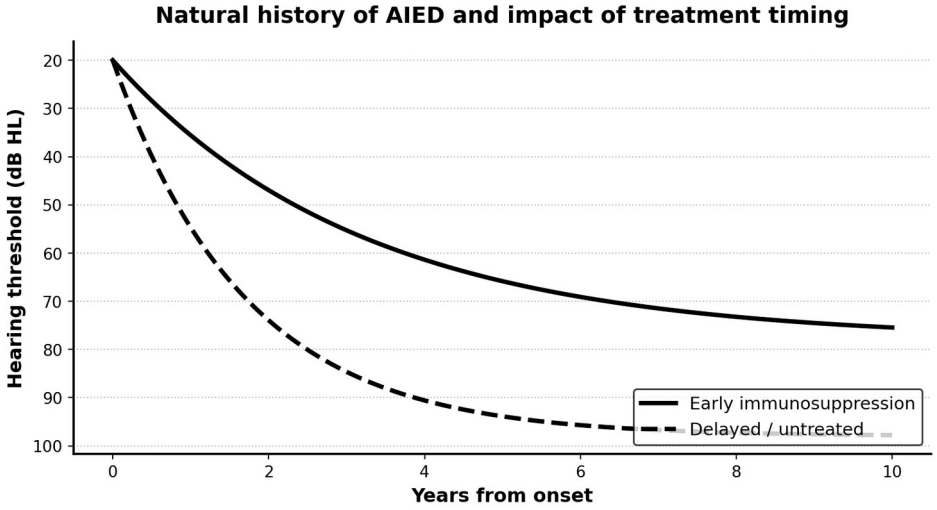

Figure 7. Natural history of autoimmune inner ear disease and the impact of treatment timing.

The longer-term course is less encouraging and underlines the chronicity of the disease. Although most patients respond initially, the proportion maintaining that improvement on continued therapy falls markedly over the following years, and many relapse or become refractory; longitudinal cohorts describe repeated immunosuppressive courses and progression to severe bilateral loss in a significant subset despite treatment [28,31,36]. Relapse on tapering is the rule rather than the exception, and a too-rapid taper frequently precipitates audiometric decline within weeks [16,30,36].

Table 7. Prognostic phases and management emphasis in autoimmune inner ear disease.

Phase	Typical course	Management emphasis
Early / active	Rapid bilateral progression; ~2/3 steroid-responsive	Prompt high-dose steroids; document response [4,30]
Relapsing / dependent	Decline on taper; repeated courses needed	Slow taper; steroid-sparing or IL-1 blockade [21,34,36]
Refractory	Poor or no steroid response	Revisit diagnosis; biomarker-guided biologics [21,34]
Burnt-out / late	Stable severe bilateral loss	Hearing aids, cochlear implant, vestibular rehab [25,41]

□ **Clinical Pearl:** About a third of AIED patients eventually achieve durable remission and can come off treatment; the remainder need long-term therapy and monitoring. Better outcomes track with later onset age, isolated cochlear (without vestibular) involvement, and a strong initial steroid response [31,34,36].

Special populations

Paediatric AIED is rare but important: it is more often bilateral and acute at presentation, may carry a stronger systemic-autoimmune association, and — when treated promptly — children sometimes recover a remarkable amount of hearing, making early recognition especially worthwhile [45]. In secondary AIED the audiovestibular disease is managed within the systemic treatment plan, in partnership with rheumatology, because controlling the underlying disorder is integral to controlling the ear [10,24,47]. Psychological comorbidity — anxiety and low mood driven by unpredictable hearing loss and dizziness — is common and warrants explicit support [14].

Monitoring in established disease is structured around the audiogram. During active treatment hearing is re-tested every two to four weeks; once stable the interval lengthens, but patients are counselled to present promptly with any subjective change, because early re-treatment of relapse offers the best chance of recovering lost thresholds [30,36]. Factors associated with a poorer trajectory include delayed presentation, vestibular as well as cochlear involvement, profound loss at diagnosis, and a weak initial steroid response; recognising these early helps calibrate how aggressively to pursue steroid-sparing escalation [12,28,36].

X. Controversies, the Guidelines Gap and Future Directions

AIED remains one of the least standardised conditions in neuro-otology, and several controversies follow directly from that [8,12,14]. The most fundamental is the absence of agreed diagnostic criteria: some centres require positive inner-ear serology, others diagnose on clinical grounds and steroid response alone, and the resulting heterogeneity has crippled multicentre research and delayed individual diagnoses [8,12]. There is, as yet, no clinical practice guideline analogous to those that exist for Ménière's disease or sudden SNHL [12,35].

□ **Key Point:** The defining problem of AIED in 2026 is not a lack of treatments but a lack of standards. With no agreed diagnostic criteria, no validated biomarker, and no practice guideline, diagnosis remains clinical and management individualised. Progress depends on biomarker-defined cohorts and multicentre trials [8,12,35].

The clinical utility of inner-ear antibodies is the second enduring controversy. Anti-HSP70 is detectable in roughly half of active patients but also in some Ménière's and sudden-SNHL patients, so its specificity and predictive value are uncertain, and whether it forecasts steroid responsiveness remains contested [4,5,6,24]. The third controversy concerns steroid-refractory disease: whether non-response reflects a distinct immunopathology, inadequate treatment, or misdiagnosis is unresolved, and the optimal escalation pathway is not established [19,21,34].

The most promising future directions are mechanistic and biomarker-driven. The anakinra experience demonstrated that an inflammatory biomarker — IL-1β — can both identify a treatable subgroup and predict response, offering a template for precision immunotherapy of the inner ear [21,32]. Cochlear macrophage biology, perilymph cytokine profiling obtained at cochlear-implant surgery, HLA and autoinflammatory-gene studies, and contrast-enhanced MRI of labyrinthine inflammation are each converging on a more granular, subgroup-stratified understanding of immune-mediated inner-ear disease [10,11,16,18,33,37]. Inner-ear-targeted drug delivery, tolerance induction, and gene-directed protection of cochlear cells are plausible longer-term therapeutic avenues [16,18].

□ **Clinical Insight:** AIED is best conceived not as a single disease but as a final common pathway — immune-mediated cochleovestibular injury — reached by molecular mimicry, bystander activation, systemic autoimmunity and genetic susceptibility. The future likely belongs to biomarker-defined subgroups treated with cytokine-specific therapy rather than blanket immunosuppression [16,21,32].

For the practising vestibular physician the take-home is pragmatic. AIED is rare, but it is one of the few progressive cochleovestibular disorders that can be arrested or reversed, and the cost of missing it — permanent bilateral deafness and vestibulopathy — is high. A bilateral, progressive or steroid-responsive audiovestibular presentation warrants serial audiometry, exclusion of treatable mimics, a directed autoimmune screen, MRI, and a properly conducted corticosteroid trial, with early escalation to steroid-sparing or biomarker-guided biologic therapy in dependent or resistant disease, all wrapped in proactive hearing and vestibular rehabilitation [12,21,34,35].

References

[1] McCabe BF. Autoimmune sensorineural hearing loss. Ann Otol Rhinol Laryngol. 1979;88(5):585-9.

[2] Harris JP, Sharp PA. Inner ear autoantibodies in patients with rapidly progressive sensorineural hearing loss. Laryngoscope. 1990;100(5):516-24.

[3] Hughes GB, Barna BP, Kinney SE, Calabrese LH, Nalepa NJ. Clinical diagnosis of immune inner-ear disease. Laryngoscope. 1988;98(3):251-3.

[4] Moscicki RA, San Martin JE, Quintero CH, Rauch SD, Nadol JB Jr, Bloch KJ. Serum antibody to inner ear proteins in patients with progressive hearing loss: correlation with disease activity and response to corticosteroid treatment. JAMA. 1994;272(8):611-6.

[5] Bloch DB, San Martin JE, Rauch SD, Moscicki RA, Bloch KJ. Serum antibodies to heat shock protein 70 in sensorineural hearing loss. Arch Otolaryngol Head Neck Surg. 1995;121(10):1167-71.

[6] Rauch SD, Jenison EL, Poor VM, San Martin JE, Bloch KJ. Elevated serum antibody titer to heat shock protein 70 in Ménière's disease. Am J Otol. 1995;16(3):345-50.

[7] Tomiyama S, Harris JP. The endolymphatic sac: its importance in inner ear immune responses. Laryngoscope. 1986;96(6):685-91.

[8] Gloddek B, Lassmann S, Gloddek J, Arnold W. Role of S-100beta as potential autoantigen in an autoimmune disease of the inner ear. J Neuroimmunol. 1999;101(1):39-46.

[9] Solares CA, Edling AE, Johnson JM, Baek MJ, Hirose K, Hughes GB, et al. Murine autoimmune hearing loss mediated by CD4+ T cells specific for inner ear peptides. J Clin Invest. 2004;113(8):1210-7.

[10] Hirose K, Discolo CM, Keasler JR, Ransohoff R. Mononuclear phagocytes migrate into the murine cochlea after acoustic trauma. J Comp Neurol. 2005;489(2):180-94.

[11] Okano T, Nakagawa T, Kita T, Kada S, Yoshimoto M, Nakahata T, et al. Bone marrow-derived cells expressing Iba1 are constitutively present as resident tissue macrophages in the mouse cochlea. J Neurosci Res. 2008;86(8):1758-67.

[12] Goodall AF, Siddiq MA. Current understanding of the pathogenesis of autoimmune inner ear disease: a review. Clin Otolaryngol. 2015;40(5):412-9.

[13] Bovo R, Aimoni C, Martini A. Immune-mediated inner ear disease. Acta Otolaryngol. 2006;126(10):1012-21.

[14] Ruckenstein MJ. Autoimmune inner ear disease. Curr Opin Otolaryngol Head Neck Surg. 2004;12(5):426-30.

[15] Mijovic T, Zeitouni A, Colmegna I. Autoimmune sensorineural hearing loss: the otology-rheumatology interface. Rheumatology (Oxford). 2013;52(5):780-9.

[16] Vambutas A, Pathak S. AAO: autoimmune and autoinflammatory (disease) in otology: what is new in immune-mediated hearing loss. Laryngoscope Investig Otolaryngol. 2016;1(5):110-5.

[17] Trune DR, Kempton JB, Gross ND. Mineralocorticoid receptor mediates glucocorticoid treatment effects in the autoimmune mouse ear. Hear Res. 2006;212(1-2):22-32.

[18] Tomiyama S. Development of endolymphatic hydrops following immune response in the endolymphatic sac of the guinea pig. Acta Otolaryngol. 1992;112(3):470-8.

[19] Harris JP, Weisman MH, Derebery JM, Espeland MA, Gantz BJ, Gulya AJ, et al. Treatment of corticosteroid-responsive autoimmune inner ear disease with methotrexate: a randomized controlled trial. JAMA. 2003;290(14):1875-83.

[20] Girasoli L, Cazzador D, Padoan R, Nardello E, Felicetti M, Zanoletti E, et al. Update on vertigo in autoimmune disorders, from diagnosis to treatment. J Immunol Res. 2018;2018:5072582.

[21] Vambutas A, Lesser M, Mullooly V, Pathak S, Zahtz G, Rosen L, et al. Early efficacy trial of anakinra in corticosteroid-resistant autoimmune inner ear disease. J Clin Invest. 2014;124(9):4115-22.

[22] Ralli M, D'Aguanno V, Di Stadio A, De Virgilio A, Croce A, Longo L, et al. Audiovestibular symptoms in systemic autoimmune diseases. J Immunol Res. 2018;2018:5798103.

[23] Shah S, Chidarala S, Jeong S, Bhatt N, Nguyen SA, McRackan TR, et al. Secondary autoimmune inner ear disease: a systematic review and meta-analysis of vestibular manifestations of systemic autoimmune disorders. Clin Rheumatol. 2023;42(10):2747-59.

[24] Pathak S, Vambutas A. Autoimmune inner ear disease: current understanding and future directions. Clin Exp Otorhinolaryngol. 2016;9(4):271-80.

[25] Gabr TA, Kotait M, Abdel Noor R. Audiovestibular findings in autoimmune diseases. Egypt J Otolaryngol. 2019;35(1):71-8.

[26] Matsuoka AJ, Harris JP. Autoimmune inner ear disease: a retrospective review of forty-seven patients. Audiol Neurootol. 2013;18(4):228-39.

[27] Broughton SS, Meyerhoff WE, Cohen SB. Immune-mediated inner ear disease: 10-year experience. Semin Arthritis Rheum. 2004;34(2):544-8.

[28] Loveman DM, de Comarmond C, Cepero R, Baldwin DM. Autoimmune sensorineural hearing loss: clinical course and treatment outcome. Semin Arthritis Rheum. 2004;34(2):538-43.

[29] Garcia-Berrocal JR, Ramirez-Camacho R, Lobo D, Trinidad A, Verdaguer JM. Adverse effects of glucocorticoid therapy for inner ear disorders. ORL J Otorhinolaryngol Relat Spec. 2008;70(4):271-4.

[30] Niparko JK, Wang NY, Rauch SD, Russell GB, Espeland MA, Pierce JJ, et al. Serial audiometry in a clinical trial of AIED treatment. Otol Neurotol. 2005;26(5):908-17.

[31] Cohen S, Shoup A, Weisman MH, Harris J. Etanercept treatment for autoimmune inner ear disease: results of a pilot placebo-controlled study. Otol Neurotol. 2005;26(5):903-7.

[32] Svrakic M, Pathak S, Goldofsky E, Hoffman R, Chandrasekhar SS, Sperling N, et al. Diagnostic and prognostic utility of measuring tumor necrosis factor in the peripheral circulation of patients with immune-mediated sensorineural hearing loss. Arch Otolaryngol Head Neck Surg. 2012;138(11):1052-8.

[33] Sugiura M, Naganawa S, Teranishi M, Nakashima T. Three-dimensional fluid-attenuated inversion recovery MRI findings in patients with sudden sensorineural hearing loss. Laryngoscope. 2006;116(8):1451-4.

[34] Buniel MC, Geelan-Hansen K, Weber PC, Tuohy VK. Immunosuppressive therapy for autoimmune inner ear disease. Immunotherapy. 2009;1(3):425-34.

[35] Breslin NK, Varadarajan VV, Sobel ES, Haberman RS. Autoimmune inner ear disease: a systematic review of management. Laryngoscope Investig Otolaryngol. 2020;5(6):1217-26.

[36] Cohen S, Roland P, Shoup A, Lowenstein M, Silverstein H, Kavanaugh A, et al. A pilot study of rituximab in immune-mediated inner ear disease. Audiol Neurootol. 2011;16(4):214-21.

[37] Kullar P, Santa Maria PL. Advances in immune-mediated inner ear disease. Front Audiol Otol. 2025;3:1624303.

[38] Garcia-Berrocal JR, Ramirez-Camacho R, Millan I, Gorriz C, Trinidad A, Arellano B, et al. Sudden presentation of immune-mediated inner ear disease: characterization and acceptance of a cochleovestibular dysfunction. J Laryngol Otol. 2003;117(10):775-9.

[39] Lobo D, Garcia-Berrocal JR, Ramirez-Camacho R. New prospects in the diagnosis and treatment of immune-mediated inner ear disease. Acta Otorrinolaringol Esp. 2009;60(1):59-64.

[40] Cohen S, Haynes D. Autoimmune inner ear disease: steroid responsiveness and long-term follow-up. J Am Acad Audiol. 2004;15(6):545-53.

[41] Licameli GR, Kenna MA, Folkman J. Cochlear implantation in autoimmune inner ear disease. Laryngoscope. 2009;119(10):2136-41.

[42] Karosi T, Konya J, Szabo LZ, Sziklai I. Intravenous immunoglobulin therapy in autoimmune inner ear disease: does it work? Audiol Neurootol. 2011;16(4):199-206.

[43] Ozbek VG, Kaplama ME, Kara CO. Cogan's syndrome: an autoimmune inner ear disease. Auris Nasus Larynx. 2012;39(4):388-91.

[44] Kessel A, Vadasz Z, Toubi E. Cogan syndrome: pathogenesis, clinical variants and treatment approaches. Autoimmun Rev. 2014;13(4-5):420-3.

[45] Semo Oz R, Gluth MB, Tesher MS. Pediatric autoimmune inner ear disease: a rare but treatable condition. Pediatr Ann. 2019;48(10):e391-4.

[46] Escamez JA, Vela J, Lopez-Escamez JA. High prevalence of systemic autoimmune diseases in patients with Ménière's disease. PLoS One. 2011;6(10):e26759.

[47] Rahne T, Clauss F, Plontke SK, Keilmann A. Prevalence of hearing impairment in patients with rheumatoid arthritis, granulomatosis with polyangiitis, or systemic lupus erythematosus. Clin Rheumatol. 2017;36(7):1501-10.

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