AIED CHEAT SHEET

Autoimmune Inner Ear Disease — Cheat Sheet for Vestibular Physicians

Suspect in any bilateral, progressive, or steroid-responsive SNHL. The steroid trial is both the diagnostic test and the first treatment. Judge it on the audiogram.

► Why AIED matters

One of the few reversible causes of progressive sensorineural hearing loss (SNHL). Under 1% of all SNHL but over-represented in tertiary neuro-otology. Female preponderance, peak third-to-sixth decade. Untreated, severe bilateral loss within ~3 years. ~1/3 of patients have or develop systemic autoimmunity. No pathognomonic test — clinical suspicion drives outcome.

Indications — when this work-up applies

- Bilateral SNHL progressive over weeks to months ± vestibular symptoms.
- Fluctuating hearing, tinnitus, or aural fullness; partial recovery after steroids.
- Steroid-responsive or bilateral hearing loss of unclear cause.
- Distinguish from Ménière's, otosyphilis, schwannoma, ototoxicity, bilateral vestibulopathy.

Mechanism — why AIED happens

Mechanism	Process	Clinical relevance
Molecular mimicry	Viral antigen cross-reacts with inner-ear protein	Post-viral onset; second-ear involvement
Bystander activation	Barrier breach releases IL-1 / TNF-α	Onset after trauma, noise, infection
Hidden-antigen exposure	Cochlin, HSP70, collagen II revealed	Detectable autoantibodies (anti-HSP70)
Cell-mediated injury	CD4+ T cells specific for ear peptides	Transferable disease in models (Solares 2004)
Final common pathway	Hair-cell loss, stria atrophy, microvasculitis	Progressive cochleovestibular failure

Pearl — *The weeks-to-months tempo is the single most discriminating feature — too fast for presbycusis, too slow for sudden SNHL. Conceive AIED as the audiovestibular expression of broader immune dysregulation, not an isolated end-organ accident.*

Diagnostic criteria — Hughes working framework

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

Pearl — *There are no internationally ratified criteria; diagnosis is clinical. Inner-ear serology supports but a negative or unavailable assay NEVER excludes AIED and must not delay treatment.*

Investigations — anchored on serial audiometry

Test	Purpose	When to order
Serial PTA + speech audiometry	Document fluctuation & steroid response; speech loss out of proportion	Mandatory; baseline and through the steroid trial
Otoacoustic emissions	Sensitive early outer-hair-cell index	Adjunct in active disease
Caloric / video head impulse	Detect frequently silent bilateral vestibular loss	Atypical, bilateral, or vestibular symptoms
cVEMP / oVEMP	Otolith-organ involvement	Useful adjunct
Anti-HSP70 + systemic screen	ANA, ANCA, RF, ESR/CRP, complement, TFTs; treponemal serology	Detect secondary AIED & otosyphilis
MRI IAM + gadolinium	Exclude vestibular schwannoma & central pathology	Mandatory at first presentation

Pearl — *The steroid trial is diagnostic AND therapeutic: prednisolone 1 mg/kg/day (max 60–80 mg) ×2–4 wk, judged on serial audiogram BEFORE taper. Responder = ≥10 dB gain at two contiguous frequencies OR ≥12% speech-discrimination gain. Taper slowly over 6–12 wk — rapid taper is the commonest cause of relapse.*

Differential diagnosis — high-yield mimics

Mimic	Key distinguishing features
Ménière's disease	Episodic vertigo 20 min–12 h; unilateral at onset; bilateral 25–45% over time
Otosyphilis	Treatable near-perfect phenocopy — treponemal serology in atypical/bilateral/young
ANCA-associated vasculitis (GPA)	Otological involvement may be presenting feature; screen ANCA
Cogan's syndrome	Interstitial keratitis + audiovestibular failure; large-vessel aortitis risk
Vestibular schwannoma / NF2	Progressive (not fluctuating) unilateral SNHL; bilateral in NF2; MRI

Ototoxicity	Aminoglycoside/platinum/loop-diuretic exposure; symmetric high-frequency loss

► Red flags — refer urgently

Bilateral progressive or steroid-responsive SNHL → urgent vestibular physician / neuro-otology + rheumatology. Sudden bilateral SNHL. Interstitial keratitis with audiovestibular failure (Cogan's, aortitis risk). Systemic vasculitis features. Re-interrogate the diagnosis before labelling disease 'refractory' — it is usually a missed mimic (otosyphilis, schwannoma) or a late presentation with irreversible damage.

Management — stepwise pyramid

Tier	Intervention	Practice principles
First-line induction	Prednisolone 1 mg/kg/day (max 60–80 mg) ×2–4 wk, slow taper 6–12 wk	~2/3 respond; the early audiometric response best predicts outcome
Hearing-targeted	Intratympanic dexamethasone	Steroid-intolerant, asymmetric, or only-hearing-ear disease
Steroid-sparing	Mycophenolate, azathioprine; methotrexate	For dependence/relapse; methotrexate RCT-negative for hearing (Harris 2003)
Biologics	IL-1 blockade — anakinra (biomarker-guided); TNF inhibitors	Anakinra helps steroid-resistant disease (Vambutas 2014); etanercept RCT-negative
Refractory	IV methylprednisolone pulse; IVIG	Reserve; IVIG evidence weak
Rehabilitation (throughout)	Hearing aids; cochlear implant; vestibular rehab	CI outcomes good if nerve spared; image early — fibrosis threatens insertion

Pearl — *Treat early and at full dose, then taper slowly. Escalate early and biomarker-guided rather than repeating steroid courses. Methotrexate and etanercept trials were negative — blanket immunosuppression disappoints; IL-1 blockade is the most promising direction.*

Counselling and follow-up

- Bimodal outlook — early treatment can reverse; delay tends to permanent bilateral loss.
- Relapse on tapering is the rule; monitor by serial audiometry — frequent while active, then at lengthening intervals with prompt re-testing for any change.
- Screen every patient for systemic autoimmunity; co-manage secondary AIED with rheumatology.
- Raise cochlear implantation proactively in bilateral severe-to-profound loss; outcomes are good when the auditory nerve is spared.

Key refs — McCabe 1979 · Harris & Sharp 1990 · Moscicki JAMA 1994 · Solares 2004 · Harris MTX RCT JAMA 2003 · Vambutas anakinra JCI 2014 · Breslin 2020