RAMSAY HUNT CHEAT SHEET

Ramsay Hunt Syndrome — Cheat Sheet for Vestibular Physicians

Anchor on the 72-hour antiviral-steroid window and the audiovestibular limb. Test for VZV in any painful or vertiginous facial palsy.

► Why Ramsay Hunt matters

Ramsay Hunt syndrome (herpes zoster oticus) is the second most common cause of atraumatic facial palsy after Bell's palsy (incidence ~5 per 100,000/year), but it is clinically louder: severe otalgia, vertigo (~31% vs ~9% in Bell's) and high-frequency SNHL are intrinsic. It is time-critical — combined antiviral and corticosteroid within 72 hours is the dominant modifiable determinant of facial recovery — and up to a third of cases present without a rash (zoster sine herpete).

Indications — when this work-up fits

► When to apply this pathway

- **► Acute lower motor neuron facial palsy with severe otalgia, vesicles (auricle, canal, drum or palate), or audiovestibular symptoms.**
- **► Apparent Bell's palsy with disproportionate pain, vertigo or hearing loss — test for VZV rather than reassure.**
- **► Distinguish from Bell's palsy, vestibular schwannoma, AOM/mastoiditis, Lyme disease and central AICA/pontine infarct.**

Mechanism — why Ramsay Hunt happens

Site	Mechanism	Clinical relevance
Geniculate ganglion	VZV reactivation → haemorrhagic ganglionitis; anterograde spread to Hunt's zone	Otalgia + auricular vesicles; the reactivation site
Fallopian canal	Inflammatory oedema raises endoneurial pressure; ischaemic demyelination + axonal loss	Compartment palsy; worse recovery than Bell's
Spiral / vestibular ganglia	Primary VZV reactivation; viral load tracks symptom severity	SNHL, tinnitus, vertigo — may precede the palsy

Pearl — *Cochleovestibular symptoms are direct ganglion reactivation, not spillover — which is why vertigo and hearing loss can arrive before the face droops.*

Diagnosis — clinical, virological and audiovestibular

Setting	Diagnostic approach
Rash + palsy	Clinical diagnosis; no confirmatory testing required
Rashless (zoster sine herpete)	Saliva, vesicle or CSF VZV PCR (test of choice); paired serology as backup
Audiovestibular work-up	Audiometry, caloric/VNG, vHIT (all six canals), cVEMP/oVEMP

Pearl — *A painful facial palsy with vertigo or hearing loss but no rash: send a saliva VZV PCR. A positive result mandates antiviral therapy that pure Bell's palsy would not receive.*

Investigations — function over imaging

Test	Purpose	When to order
VZV PCR (saliva/vesicle/CSF)	Confirm aetiology, especially zoster sine herpete	Rashless or atypical case
Pure-tone / speech audiometry	Quantify high-frequency SNHL; baseline	Every case
Video head impulse test	VOR gain across all six semicircular canals	Characterise multi-canal loss
cVEMP / oVEMP	Saccular / utricular afferents; inferior vs superior nerve	Map otolith involvement
MRI + gadolinium	Exclude schwannoma, AICA/pontine infarct, infiltration	Atypical/stalled; central signs
ENoG / EMG	Degree of axonal degeneration (prognostic)	Complete palsy; decompression decision

Pearl — *vHIT and VEMP reveal multi-canal and otolith involvement calorics miss, and deficits can persist despite a normal-appearing eighth nerve on MRI — test function, do not rely on imaging.*

Differential diagnosis — high-yield mimics

Mimic	Key distinguishing features
Bell's palsy	No vesicles; mild pain; ~85% reach HB I; steroids help but aciclovir did not in the Bell's palsy trial
Zoster sine herpete	RHS features without rash; VZV PCR / serology positive
Vestibular schwannoma	Progressive SNHL; facial weakness late; MRI mass
AOM / mastoiditis	Bulging or perforated drum, fever, conductive loss
Lyme disease	Erythema migrans; tick exposure; serology
Central (AICA/pontine)	Forehead may be spared; brainstem signs; MRI DWI

► ***Red flags —*** *Immunocompromised + focal deficits (VZV vasculopathy/stroke) · Depressed consciousness or meningism (MRI + LP) · Lower cranial nerve (IX/X) palsy with aspiration risk · Stalled or atypical recovery. Each warrants escalation and imaging.*

Management — the 72-hour window

Component	Regimen	Principle

Antiviral (first-line)	Valaciclovir 1 g PO tds 7–10 d (famciclovir 500 mg tds alt.)	Start within 72 h — the dominant modifiable factor
Antiviral (severe)	IV aciclovir 10 mg/kg every 8 h	Disseminated / immunocompromised / vomiting
Corticosteroid	Prednisolone ~1 mg/kg/day, taper 7–14 d	Adjuvant; reduces canal oedema
Eye protection	Lubricants by day; ointment + taping/moisture chamber overnight	Prevent exposure keratitis; ophthalmology if cornea involved
Pain	Simple analgesia → gabapentinoids / tricyclics	Severe otalgia and postherpetic neuralgia
Rehabilitation	Vestibular suppressants acutely only; then vestibular rehab + facial retraining	Suppressants retard compensation; retraining limits synkinesis

Pearl — *Treat empirically on suspicion; do not wait for virology. The cost of treating a Bell's palsy as RHS is low, and the 72-hour window is decisive.*

Prognosis and counselling

- **Complete recovery ~75% if treated within 72 h, ~30% if delayed beyond a week, ~20% untreated.**
- **Two-year House-Brackmann grade I in ~58% of RHS versus ~86% of Bell's palsy.**
- **Hearing recovers poorly (~11%); vertigo compensates over weeks, assisted by rehabilitation.**
- **Adverse signs: complete paralysis, advanced age, diabetes, dense ENoG degeneration, cranial polyneuropathy.**
- **Recurrence <5% in the immunocompetent; counsel the recombinant zoster vaccine after recovery.**

Special populations and prevention

The immunocompromised are prone to severe, disseminated or polyneuropathic disease and VZV vasculopathy — give intravenous antivirals, longer courses and surveillance. In pregnancy, corticosteroids and aciclovir/valaciclovir are reasonable with obstetric and infectious-diseases input. Children recover better than adults; the elderly carry the highest incidence and postherpetic neuralgia risk. Prevention now matters as much as treatment: the recombinant zoster vaccine exceeds 90% efficacy (ZOE-50/ZOE-70) and is reasonable in eligible recovered patients.

Key references — Sweeney CJ, Gilden DH. J Neurol Neurosurg Psychiatry 2001;71:149–154 · Murakami S et al. Ann Neurol 1997;41:353–357 · Coulson S et al. Otol Neurotol 2011;32:1025–1030 · Furuta Y et al. J Clin Microbiol 2001;39:2856–2859 · Uscategui T et al. Cochrane 2008;CD006851 · Cunningham AL et al. N Engl J Med 2016;375:1019–1032.