CHIARI CHEAT SHEET	**Arnold–Chiari Malformation — Cheat Sheet for Vestibular Physicians** *Keep it on the central differential: about one dizziness-clinic patient in 500 has an occult Chiari I. Read the central oculomotor signs and the craniocervical junction on MRI.*

► Why Arnold–Chiari matters

A congenital hindbrain herniation in which the cerebellar tonsils descend through the foramen magnum. Radiological Chiari I is found in ~0.5–1% of scans but is usually silent; symptomatic disease is ~0.1%. It reaches the dizziness clinic in about 1 in 500 referrals — often without the classical cough headache — so vestibular-predominant presentations are easily missed.

Chiari I — cerebellar tonsils herniate through the foramen magnum with a cervical syrinx (left); sagittal MRI shows tonsillar descent below the foramen magnum (right). Source: Mayfield Clinic.

► Indications — when to suspect Chiari

- ► **Cough headache —** suboccipital / occipital headache provoked by cough, strain or Valsalva, often with neck pain.
- ► **Central oculomotor signs —** downbeat or gaze-evoked nystagmus, oscillopsia, gait ataxia, central positional nystagmus.
- ► **Long-tract / syrinx signs —** cape sensory loss, hand wasting, scoliosis — suggest an associated syrinx.
- ► **Context —** hypermobility (hEDS), prior CSF leak, or unexplained progressive disequilibrium.

► Mechanism — why Chiari happens

Step	Mechanism	Clinical relevance
Posterior fossa crowding	Small/underdeveloped posterior fossa or cranial settling (hEDS).	Tonsils forced caudally through the foramen magnum.
Foramen magnum obstruction	Tonsils 'piston' with each heartbeat; pulsatile CSF flow impaired.	Reduced cine CSF flow correlates with Valsalva headache.
Neural compression	Brainstem, lower cranial nerves and central vestibular pathways compressed.	Vertigo, downbeat nystagmus, ataxia, dysphagia.
Syrinx formation	Altered CSF dynamics drive fluid into the central cord (~20–30%).	Cape sensory loss, hand wasting, scoliosis.

Pearl — *Read physiology, not just millimetres. Tonsillar descent is a measurement; obstructed pulsatile CSF flow at the craniocervical junction is the disease.*

► Diagnostic & radiological criteria

Tier	Defining features
Chiari I	Tonsillar herniation over 5 mm (adult) / over 6 mm (child); no open neural-tube defect.
Borderline	3–5 mm descent — diagnose only with crowded cisterna magna, effaced CSF, and concordant symptoms.
Chiari 0 / 1.5	Syrinx with minimal descent (Chiari 0); tonsils + brainstem/obex descent (Chiari 1.5).
Chiari II	Vermis + brainstem herniation, tectal beaking, myelomeningocele, hydrocephalus — the classic Arnold–Chiari.

***Pearl** — Diagnose the syndrome, not the scan. Incidental tonsillar ectopia is common — anatomy must be matched to compatible clinical and vestibular findings.*

► Investigations — imaging-led work-up

Test	Purpose	When to order
MRI brain + CVJ	Confirm tonsillar herniation, posterior-fossa crowding, hydrocephalus.	First-line in every suspected case.
MRI whole spine	Screen for syringomyelia and tethered cord.	Mandatory once Chiari I confirmed.
Cine CSF flow	Quantify obstruction at the craniocervical junction.	Borderline descent; pre-surgical assessment.
VNG / audiovestibular	Central oculomotor signs; downbeat nystagmus, poor VOR suppression.	Vestibular-predominant presentations.
Polysomnography	Central / obstructive sleep apnoea.	Children; brainstem symptoms.

***Pearl** — Fixation-present vertical or direction-changing nystagmus is central until proven otherwise — image the craniocervical junction.*

► Differential diagnosis — high-yield mimics

Mimic	Key distinguishing features
Primary cough headache	Valsalva headache but normal MRI; indomethacin-responsive.
Spontaneous intracranial hypotension	Acquired tonsillar descent; orthostatic headache; pachymeningeal enhancement; reverses with blood patch — DO NOT decompress.
Basilar invagination / CVJ instability	Odontoid above foramen magnum; needs stabilisation, not decompression alone.
Foramen magnum tumour	Enhancing mass; treated by resection.
Multiple sclerosis	White-matter lesions; vertigo/ataxia without Valsalva headache.

► **Red flags —** Acquired (not congenital) herniation from a CSF leak or raised ICP — it must not be decompressed · progressive myelopathy or enlarging syrinx · lower-cranial-nerve dysfunction (dysphagia, stridor, apnoea) · infant central apnoea or stridor. Each warrants MRI and escalation before symptomatic therapy.

► Management — conservative to surgical

Tier	Intervention	Practice principles
Conservative	Observe, Valsalva/activity modification, vestibular rehab, periodic MRI.	Mild/incidental disease without syrinx; natural history is usually benign.
Surgical decompression	Suboccipital craniectomy + C1 laminectomy.	For disabling symptoms, syrinx, or progressive deficit; ~70–85% improve.
Duraplasty	Open dura + patch when bone-only inadequate.	Better syrinx resolution; higher CSF-leak / meningitis risk.
Hydrocephalus / fetal	VP shunt or ETV; in-utero myelomeningocele repair (Chiari II).	Treat hydrocephalus first; MOMS trial reduces herniation and shunting.

***Pearl** — Match the operation to the mechanism. Bone-only decompression lowers complications; add duraplasty where a syrinx is present or bony decompression is inadequate.*

► Counselling and follow-up

- Most mild Chiari I is stable on conservative care; decompression reliably treats the cough headache and arrests syrinx progression.
- Residual disequilibrium may persist after anatomically successful surgery — vestibular rehabilitation is the proper target.
- Recurrence/progression in ~10–30% long-term; reoperation ~5–10%. Maintain neurological and MRI surveillance.
- Comorbid migraine is common and persists after surgery — treat on its own merits; safety-net for new weakness, numbness or sphincter change.

► Associated conditions — management shift

Connective-tissue disorders (hEDS) cause craniocervical instability and may need occipitocervical fusion in addition to decompression. Spontaneous intracranial hypotension and idiopathic intracranial hypertension both mimic Chiari and demand the opposite treatment — sealing a leak or lowering pressure — so exclude them before operating on borderline herniation. A coexisting tethered cord may need release; decisions are best made in a multidisciplinary setting.