Arnold–Chiari Malformation:

A Vestibular Physician's Deep Review of Hindbrain Herniation, Central Vestibular Features, and Management

Vestibular Medicine for Vestibular Physicians

Central Vestibular Pathology — Module 2.5

Australian Dizziness Clinics | *www.AustralianDizzinessClinics.com*
Version 1.0 | June 2026

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 central 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.

Table of Contents

I. Introduction, History and Epidemiology

II. Pathophysiology — Hindbrain Herniation and CSF Dynamics

III. Clinical Features and the Central Vestibular Syndrome

IV. Classification and Diagnostic Criteria

V. Investigations: MRI, Cine CSF Flow and Audiovestibular Testing

VI. Differential Diagnosis

VII. Management — Conservative Care and Surgical Decompression

VIII. Pharmacological Adjuncts, Refractory Disease and Associated Conditions

IX. Prognosis, Recurrence and Special Populations

X. Controversies and Future Directions

References

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I. Introduction, History and Epidemiology

Arnold–Chiari malformation refers to a family of congenital hindbrain herniation disorders defined by caudal displacement of the cerebellum and, in the more severe forms, the brainstem through the foramen magnum. The Austrian pathologist Hans Chiari produced the first systematic descriptions in 1891, classifying several patterns of hindbrain herniation [1]. Julius Arnold's 1894 account of an infant with spina bifida prompted his students Schwalbe and Gredig to introduce the eponym 'Arnold–Chiari malformation' in 1907 for the severe myelomeningocele-associated form now designated Chiari type II [2]. Modern usage favours 'Chiari malformation' as the umbrella term and reserves 'Arnold–Chiari' for the type II variant, while recognising that the original four-type taxonomy has been progressively refined as imaging and surgical experience have accumulated [21,30].

For the vestibular physician the relevant entity is overwhelmingly Chiari I malformation (CM-I): tonsillar ectopia without an open neural-tube defect, presenting in adolescence or adulthood with headache, central vestibular symptoms, and — in a substantial minority — syringomyelia [3,15]. CM-I is at once one of the commonest incidental findings on brain MRI and one of the more treacherous diagnoses in a dizziness clinic, because the radiological abnormality is frequently silent and its mere presence does not establish causation [4,8].

Population imaging studies place radiological CM-I — usually defined by tonsillar descent beyond an agreed threshold — in roughly 0.5–1% of unselected subjects, with cerebellar tonsillar ectopia detectable in up to about 3% of scans when lenient criteria are applied [4,18,35]. The great majority of these individuals are asymptomatic [4]. Symptomatic CM-I is far rarer, with prevalence estimates near 0.1% (about 1 per 1,000) [8]. There is no consistent ethnic or geographical predilection, most series show an equal or slightly female-predominant sex distribution, and a positive family history is present in roughly 3–12% of cases, pointing to a polygenic genetic contribution [5,7,9].

Table 1. Epidemiology of Chiari malformation at a glance.

Measure	Value	Notes
Radiological CM-I (general population)	~0.5–1%	Tonsillar ectopia up to ~3% with lenient criteria [4,18,35]
Symptomatic CM-I prevalence	~0.1% (1 / 1,000)	Most radiological CM-I is silent [4,8]
CM-I among dizziness referrals	~0.2% (≈1 / 500)	Mean herniation ~6 mm; often no classic headache [10]
Syringomyelia in CM-I	~20–30%	Higher when herniation is large [6,11]
Chiari II birth prevalence	~3.4–4.8 / 10,000 live births	Tracks open spina bifida; falls with folate [27,28]

In the specific setting of a neurotology service the diagnosis is uncommon but not negligible. A retrospective review of more than 6,400 patients presenting with dizziness identified MRI-confirmed CM-I in roughly 0.2% — about 1 in 500 — with a mean tonsillar herniation of approximately 6 mm, and many of these patients lacked the textbook occipital cough headache [10]. This is the central epidemiological message for vestibular practice: Chiari can present predominantly, or even exclusively, with vestibular complaints, so it belongs on the differential for unexplained central vertigo, downbeat nystagmus, or progressive disequilibrium [10,32].

□ **Clinical Insight:** Roughly one dizziness-clinic patient in five hundred harbours an occult Chiari I malformation, and a meaningful fraction of these never report the classical Valsalva headache. Treat unexplained central vestibular signs as an indication to scrutinise the craniocervical junction on MRI [10,32].

The functional burden of symptomatic CM-I is easily underestimated because each cough headache is brief. In practice patients describe months of restricted activity, anticipatory avoidance of straining, exertion and even laughter, and a slow erosion of confidence in balance that drives them from work and exercise long before a diagnosis is reached [3,32]. Disease-specific handicap is concentrated in the vestibular domain: chronic disequilibrium and oscillopsia, rather than the headache, are what most often disable the neurotology referral, and these are precisely the features that respond least predictably to surgery [10,43]. The incidental-finding problem is the mirror image of this burden — a tonsillar

measurement reported on a scan ordered for unrelated reasons generates anxiety, repeat imaging, and pressure to operate on anatomy that may never have caused a symptom [4,8].

Chiari II is essentially a condition of open spina bifida and shares the epidemiology of neural-tube defects, with a historical birth prevalence of about 3.4–4.8 per 10,000 live births that has declined markedly in folate-supplemented populations [27,28]. Types III and IV — occipital encephalocele with herniated cerebellar tissue, and cerebellar hypoplasia or aplasia respectively — are vanishingly rare and lie outside routine vestibular practice [30]. The remainder of this review concentrates on CM-I, with Chiari II addressed where its mechanisms and management illuminate the spectrum [15,16].

II. Pathophysiology — Hindbrain Herniation and CSF Dynamics

The dominant model of CM-I is a volumetric mismatch between a congenitally small posterior fossa and a normally developed hindbrain [13]. Morphometric MRI studies consistently demonstrate reduced posterior fossa dimensions — a shortened clivus and basiocciput, often with paraxial mesodermal underdevelopment — that crowd the cerebellum and force the tonsils caudally through the foramen magnum [13,17]. The herniated tonsils assume the characteristic 'peg-like' configuration on sagittal imaging [17,19].

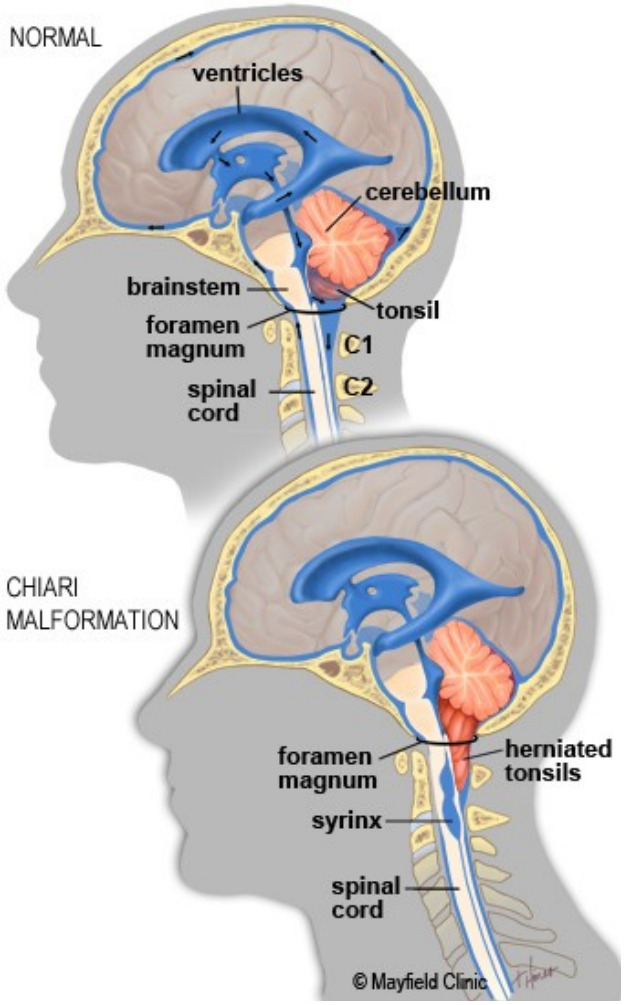

Figure 1. Normal hindbrain anatomy compared with Chiari I malformation. In Chiari I the cerebellar tonsils herniate through the foramen magnum below C1, obstructing CSF flow and producing a cervical syrinx.
Source: Mayfield Clinic.

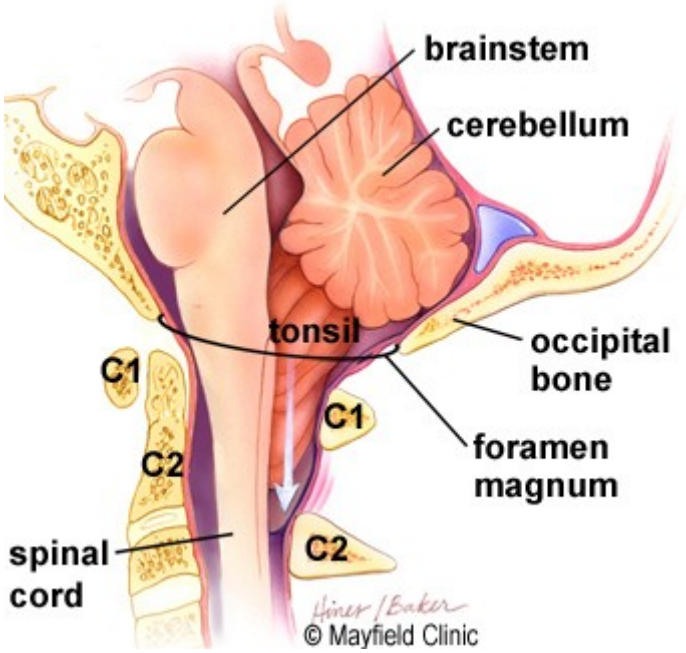

Figure 2. Sagittal close-up of the craniocervical junction. The cerebellar tonsil is impacted against the occipital bone at the foramen magnum, compressing the brainstem and upper cervical cord at C1–C2.
Source: Mayfield Clinic.

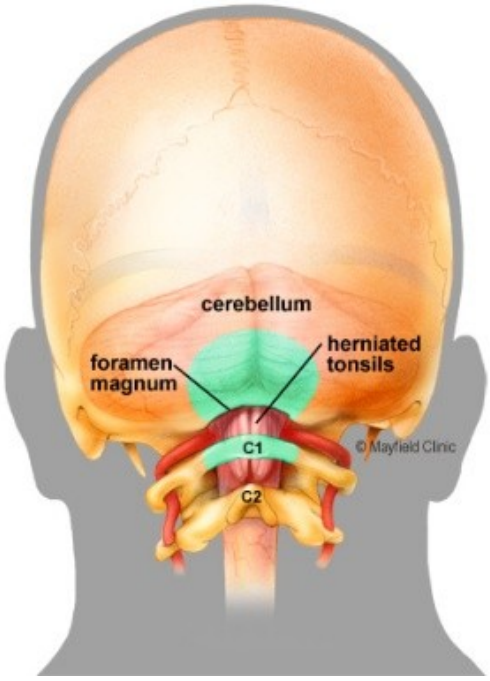

Figure 3. Posterior view of the craniocervical junction showing the cerebellum and herniated tonsils descending through the foramen magnum past C1 and C2.
Source: Mayfield Clinic.

Genetic work has nuanced the 'small skull' paradigm. Familial clustering, twin concordance, and linkage and candidate-gene studies indicate a heritable, polygenic basis, with developmental genes governing skull-base and hindbrain growth implicated [7,9]. Variants in chromodomain-helicase genes have been linked to familial CM-I, and children carrying such variants frequently show macrocephaly — a larger head circumference raising CM-I risk several-fold [9]. This suggests that disproportionate brain growth relative to skull-base capacity can produce the same foramen-magnum crowding as a primarily small posterior fossa, arriving at tonsillar herniation from the opposite direction [9,13].

Although usually congenital, CM-I can be acquired. Any process that lowers intracranial pressure or allows the brain to settle may reproduce tonsillar descent: chronic CSF leakage from lumbar punctures,

over-draining shunts, or spontaneous intracranial hypotension generates a 'pseudo-Chiari' that reverses when the leak is sealed [42]. Heritable connective-tissue disorders — particularly hypermobile Ehlers–Danlos syndrome — produce craniocervical instability and progressive cervicomedullary settling with secondary herniation, a mechanism distinct from primary bony hypoplasia and with different surgical implications [14].

Two consequences flow from herniation: mechanical compression and disordered CSF circulation [11,41]. The descended tonsils compress the medulla and upper cervical cord, disturbing cranial-nerve nuclei, central vestibular pathways, and long tracts; simultaneously, impaction at the foramen magnum obstructs the pulsatile to-and-fro of CSF between cranial and spinal compartments [20,41]. During systole, brain expansion drives CSF through a narrowed outlet as a pressure jet; diastolic return is impaired, and the tonsils may 'piston' with each cardiac cycle [20,41]. Reduced or absent CSF flow at the craniocervical junction on phase-contrast imaging correlates with symptomatic disease and with Valsalva-triggered headache [20].

These hydrodynamic disturbances drive syringomyelia, the most consequential secondary pathology. The prevailing mechanism holds that obstructed subarachnoid flow and altered craniospinal pressure dissociation propel fluid into the central cord, producing a syrinx in roughly one-quarter to one-third of CM-I patients [6,11,12]. The cavity preferentially injures decussating spinothalamic fibres — yielding dissociated, cape-distribution sensory loss — and anterior horn cells, and may elongate over time to cause hand-muscle wasting and neurogenic scoliosis [12,40].

Chiari II has a distinct pathogenesis tied to the open neural-tube defect. The 'unified theory' proposes that fetal CSF leakage through the dysraphic spinal canal collapses the rhombencephalic vesicle, preventing normal posterior-fossa expansion and drawing the hindbrain caudally [29]. The result is a small posterior fossa with herniation of the vermis, an elongated medulla, tectal beaking, a fourth ventricle drawn into the cervical canal, and near-universal hydrocephalus from outflow obstruction [28,29]. The MOMS trial's demonstration that in-utero closure reduces hindbrain herniation and shunt dependence is the strongest evidence that arresting the CSF leak alters the malformation itself [27].

□ **Key Point:** Chiari I is fundamentally a disorder of CSF dynamics at the foramen magnum, not merely a measurement of tonsillar descent. Mechanical compression and obstructed pulsatile flow together generate the central vestibular syndrome and drive syrinx formation in 20–30% of patients [11,12,20].

III. Clinical Features and the Central Vestibular Syndrome

CM-I has a broad and frequently atypical clinical spectrum [3,32]. The single most characteristic symptom is the Chiari headache: a short-lived suboccipital or occipital pressure pain provoked by Valsalva manoeuvres — coughing, straining, sneezing, or laughing — and often accompanied by neck pain [33]. Its tussive, transient quality reflects the cardiac-cycle pressure dissociation across an obstructed foramen magnum, and it is the symptom most reliably relieved by decompression [20,33].

Vestibular and oculomotor features dominate the remainder of the picture and are the reason CM-I reaches a dizziness clinic. More than three-quarters of symptomatic patients report otoneurological symptoms — vertigo, disequilibrium, oscillopsia, or blurred vision — and gait ataxia from cerebellar involvement is common [3,10,32]. Nystagmus is frequent and characteristically central in type, most classically downbeat but also gaze-evoked [3]. Lower-cranial-nerve dysfunction (dysphagia, dysarthria, hoarseness) and long-tract signs appear when herniation is marked or a syrinx is present; neck and arm pain with motor-sensory deficits affect roughly half of symptomatic patients [3,32]. Auditory symptoms — tinnitus, aural fullness, mild hearing loss — occur but are less frequent than vestibular complaints [10].

Downbeat nystagmus deserves particular emphasis because CM-I is one of its classical causes [3]. It reflects failure of the flocculus and the vestibulocerebellar circuitry that normally supply a downward bias to vertical gaze-holding; loss of this inhibition leaves an unopposed upward drift corrected by downward saccades, producing nystagmus that intensifies on downgaze and lateral gaze and is accompanied by impaired vertical smooth pursuit and defective visual suppression of the vestibulo-ocular reflex [3]. For

the vestibular physician, fixation-present vertical nystagmus is a central sign that should prompt craniocervical imaging.

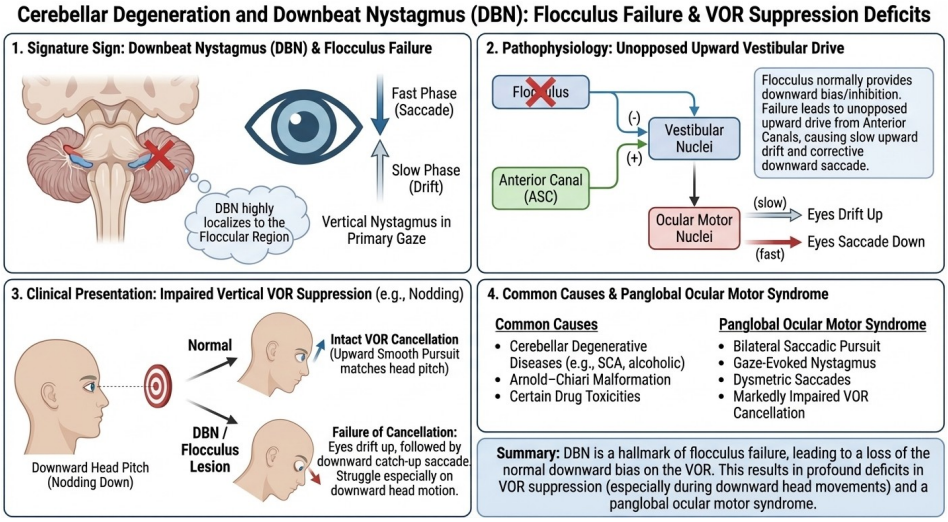

Figure 4. Downbeat nystagmus and flocculus failure. Arnold–Chiari malformation is a classical cause of downbeat nystagmus, with impaired VOR suppression and a panglobal ocular-motor syndrome.

Source: Downbeat nystagmus and VOR-suppression deficits, Australian Dizziness Clinics Education Hub.

On examination CM-I produces signs of brainstem and cerebellar compression: downbeat or gaze-evoked nystagmus, impaired tandem gait, limb dysmetria, and hyperreflexia [3,32]. Where a syrinx has formed, the classical dissociated sensory loss (pain and temperature lost in a cape distribution with preserved light touch) and small-hand-muscle wasting appear, and scoliosis is an important paediatric clue — present in up to a fifth of CM-I children and over half of those with a syrinx [40]. Presentation is age-dependent: infants may show stridor, feeding difficulty, or central apnoea from lower-cranial-nerve compression; school-age children more often present with scoliosis or recurrent headache than with the adult cough headache [15,39]. An infant with unexplained central apnoea or stridor warrants consideration of Chiari, particularly type II in the context of spina bifida [15,28].

□ **Important:** Persistent downbeat nystagmus, fixation-present vertical or direction-changing nystagmus, or any focal brainstem or lower-cranial-nerve sign in a dizzy patient is central until proven otherwise and mandates MRI of the craniocervical junction [3,10].

The crucial clinical caveat is variability. A meaningful subset of patients present primarily with episodic vertigo or chronic disequilibrium and little or no headache, and it is precisely these patients who are misclassified as having a peripheral or functional disorder [10,32]. Anchoring on the cough headache as a prerequisite will miss the vestibular-predominant presentations that are common in neurotology referrals.

A practical examination point for this audience concerns positional testing. Because CM-I can generate central positional nystagmus, a Dix–Hallpike or supine roll test may provoke nystagmus that superficially resembles benign paroxysmal positional vertigo but lacks its latency, torsional-upbeat signature, and fatigability; downbeat positional nystagmus in particular should never be dismissed as a posterior-canal variant [3,10]. Equally, the head-impulse–nystagmus–test-of-skew framework calibrated for the acute vestibular syndrome is built to detect stroke, not the chronic, fluctuating presentation of Chiari, and a normal bedside head-impulse response in a patient with central nystagmus reinforces rather than excludes a central localisation [3,32].

IV. Classification and Diagnostic Criteria

Chiari malformation is fundamentally a radiological diagnosis, and CM-I in particular is defined by the position of the cerebellar tonsils relative to the foramen magnum on MRI [16,17]. The widely accepted benchmark is tonsillar descent greater than 5 mm below the McRae (basion–opisthion) line in adults, with a 6 mm threshold in children to allow for the higher-lying tonsils of early life and their physiological ascent

through adolescence [17,18]. Descent of 3–5 mm is regarded as borderline and demands clinical correlation rather than automatic diagnosis [16,19].

Table 2. Classification of Chiari malformations.

Type	Defining features	Clinical relevance
Chiari 0	Syringomyelia with minimal/no tonsillar descent; impaired CSF flow at foramen magnum	Controversial; confirmed only if syrinx resolves after decompression [22]
Chiari I	Cerebellar tonsillar herniation over 5 mm (adult); no open neural-tube defect	The entity relevant to vestibular practice [3,16]
Chiari 1.5	Tonsillar plus brainstem (obex) descent; no spina bifida	Greater syrinx risk; may need more extensive decompression [23]
Chiari II	Vermis + brainstem herniation; tectal beaking; myelomeningocele; hydrocephalus	Presents in infancy; the classic Arnold–Chiari [28,29]
Chiari III	Occipital/high-cervical encephalocele containing cerebellar tissue	Very rare; high morbidity and mortality [30]
Chiari IV	Cerebellar hypoplasia/aplasia with enlarged posterior fossa CSF space	A developmental malformation rather than a herniation [30]

On MRI, CM-I shows a crowded posterior fossa with effaced cisterna magna and peg-shaped tonsils protruding into the upper cervical canal; syringomyelia, when present, appears as an elongated intramedullary fluid signal on spinal sequences [17,19]. Radiological thresholds are guides, not absolutes — some individuals with 6–7 mm of descent are wholly asymptomatic while others with 3–4 mm and a tight posterior fossa are symptomatic [4,19]. Contemporary assessment therefore weighs morphology and CSF dynamics alongside the millimetre measurement: an effaced cisterna magna, tonsils impacting the medulla, and reduced CSF flow argue for a pathological Chiari even when descent is borderline [16,20].

The conceptual key is the distinction between cerebellar tonsillar ectopia (an anatomical finding) and Chiari malformation syndrome (anatomy plus concordant clinical features) [4,21]. Because incidental mild ectopia is common, radiology alone cannot establish the diagnosis; guidelines increasingly frame CM-I as a clinico-radiological syndrome to avoid over-calling silent descent as disease [4,21,45]. Severity grading instruments such as the Chiari Severity Index, which combine headache type, myelopathy, and syrinx status, attempt to formalise this anatomy–symptom integration and to predict surgical benefit [36].

Chiari II is a clinico-radiological diagnosis usually evident prenatally or at birth, with the ultrasound 'lemon' (skull) and 'banana' (curved cerebellum) signs flagging the hindbrain herniation of an open neural-tube defect, and fetal or postnatal MRI confirming vermian and brainstem descent, tectal beaking, and hydrocephalus [27,28]. Types III and IV are defined by their gross structural anomalies on imaging and rarely present a diagnostic question [30].

V. Investigations: MRI, Cine CSF Flow and Audiovestibular Testing

MRI is the investigation of choice once Chiari is suspected on symptoms or examination [16]. A dedicated brain and craniocervical-junction study visualises the tonsils, brainstem, and posterior-fossa dimensions, confirms the degree of herniation, and detects secondary features such as hydrocephalus or a low-lying, kinked medulla [16,17]. Sagittal T1 and T2 sequences through the foramen magnum are the workhorses; flexion–extension imaging is added when craniocervical instability is suspected, as in hypermobile connective-tissue disorders [14].

Phase-contrast cine MRI quantifies pulsatile CSF flow at the craniocervical junction and helps separate anatomically borderline but physiologically obstructive herniation from incidental ectopia [20]. Diminished or absent flow correlates with symptomatic disease and with Valsalva headache, and a normal flow study in a patient with borderline descent argues against attributing symptoms to the tonsillar position [20,41].

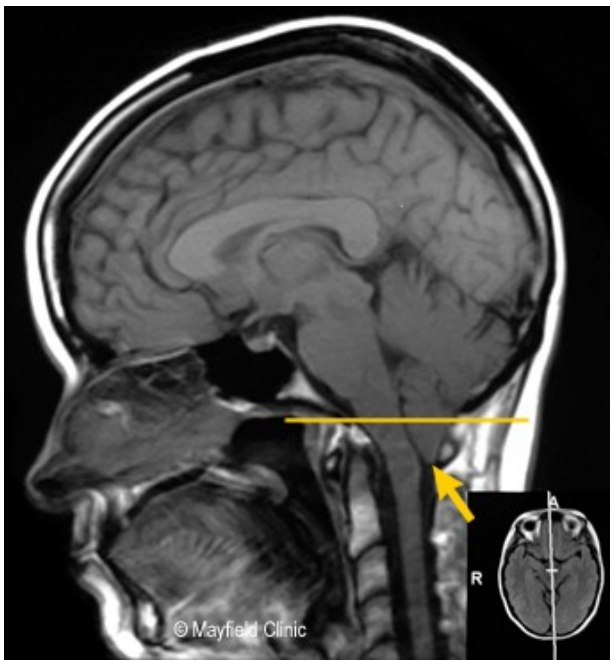

Figure 5. Sagittal T1 MRI of the brain in Chiari I malformation. The cerebellar tonsils descend below the foramen magnum (yellow line, arrow) — the defining radiological feature.
Source: Mayfield Clinic.

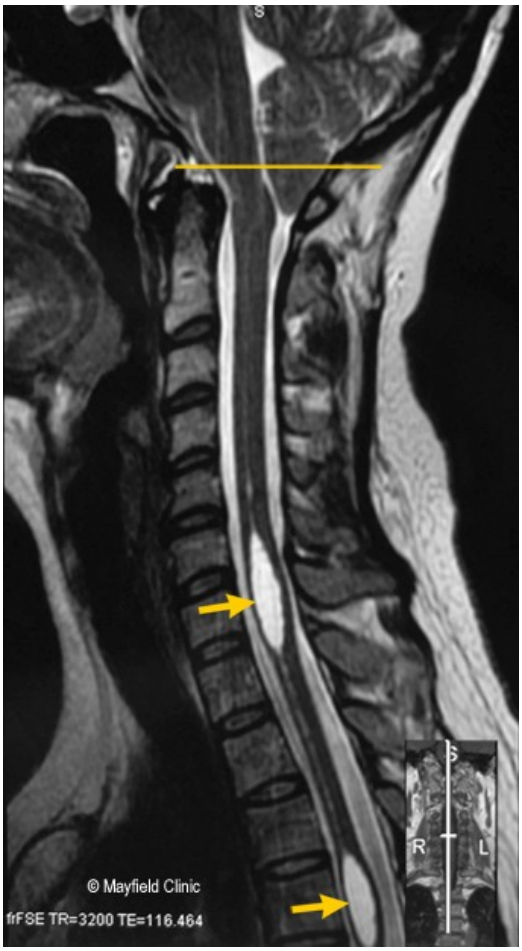

Figure 6. Sagittal T2 MRI of the cervical spine showing a syrinx — an elongated intramedullary fluid cavity (arrows) — associated with the tonsillar herniation above.
Source: Mayfield Clinic.

Whole-spine MRI is strongly recommended in any confirmed CM-I to screen for syringomyelia and associated anomalies such as a tethered cord, because a syrinx — present in about a quarter of cases

and sometimes low in the thoracic cord — materially changes management and is a recognised indication for decompression [6,11,40]. Imaging also defines the extent of any scoliosis and excludes occult spinal lesions that might themselves cause a syrinx [40,42].

Audiovestibular testing is adjunctive but valuable in vestibular-predominant presentations. Videonystagmography commonly reveals central oculomotor signs in CM-I — downbeat nystagmus, impaired smooth pursuit, and defective VOR suppression — while caloric responses may be normal or mildly reduced if brainstem involvement affects vestibular-nucleus output [10]. In the published neurotology series a majority of Chiari patients who underwent VNG had abnormal, centrally patterned results, and it was often the vestibular testing that raised suspicion and prompted the MRI [10]. Figure 7 contrasts the central pattern with peripheral nystagmus.

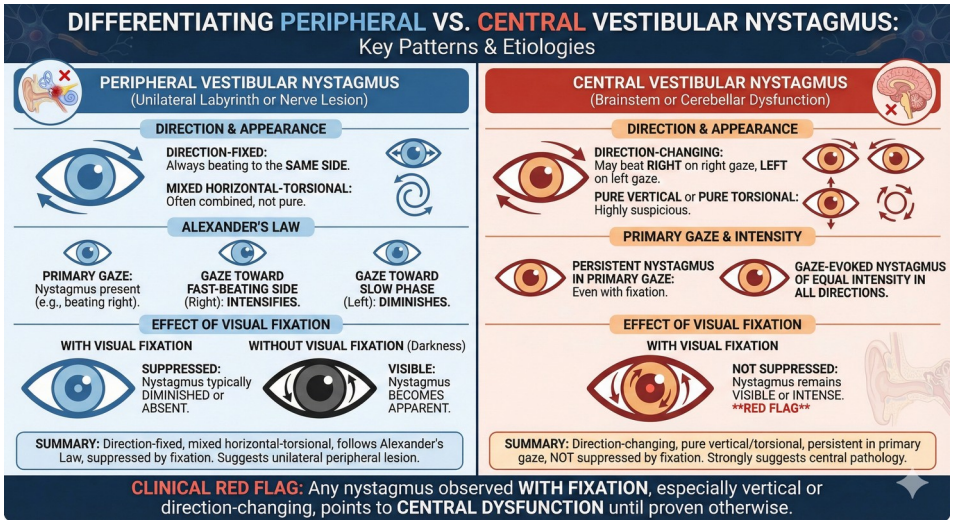

Figure 7. Differentiating peripheral from central vestibular nystagmus. Chiari I produces the central pattern — direction-changing or pure vertical nystagmus, persistent in primary gaze and not suppressed by fixation.
Source: Peripheral versus central vestibular nystagmus, Australian Dizziness Clinics Education Hub.

Supplementary investigations are directed by the presentation. Fundoscopy screens for papilloedema, which is not a feature of uncomplicated CM-I and should redirect attention to raised intracranial pressure or coexisting idiopathic intracranial hypertension [19]. Polysomnography quantifies central or obstructive sleep apnoea, particularly in children, and may accelerate a decision to decompress [15]. Prenatal ultrasound and fetal MRI establish Chiari II in the context of spina bifida [27]. No blood test is diagnostic, though assessment for a heritable connective-tissue disorder is reasonable when hypermobility is evident, as it influences the surgical plan [14].

□ **Clinical Pearl:** Pair the millimetres with the physiology. A cine CSF flow study and a whole-spine screen convert a bare tonsillar measurement into a defensible decision: obstructed flow plus a syrinx supports intervention; preserved flow with borderline descent argues for restraint and a search for another cause [20,41].

VI. Differential Diagnosis

The differential operates on two levels: the causes of a Chiari-like clinical syndrome, and the causes of tonsillar herniation once it is seen on imaging [16,42]. Both matter, because the treatments diverge sharply — a congenital small posterior fossa is decompressed, whereas an acquired herniation from a CSF leak is treated by sealing the leak [14,42].

Among clinical mimics, primary cough headache reproduces the tussive occipital pain of CM-I but with normal imaging and a characteristic response to indomethacin, so all exertional headache warrants imaging to exclude a structural cause [33]. Spontaneous intracranial hypotension produces acquired tonsillar descent with orthostatic (upright-worse) headache, diffuse pachymeningeal enhancement, and brain sagging, and reverses with an epidural blood patch — a 'pseudo-Chiari' that must not be decompressed [42]. Demyelinating disease, especially multiple sclerosis, causes brainstem and

cerebellar vertigo, ataxia, and nystagmus, but is distinguished by white-matter lesions and the absence of Valsalva-triggered headache [16].

Table 3. Differential diagnosis of Chiari I malformation — key distinguishing features.

Condition	Overlapping features	Distinguishing features
Primary cough headache	Valsalva-triggered occipital headache	Normal MRI; indomethacin-responsive [33]
Spontaneous intracranial hypotension	Acquired tonsillar descent	Orthostatic headache; pachymeningeal enhancement; reverses with blood patch [42]
Basilar invagination / CVJ anomaly	Brainstem compression, lower-CN signs	Odontoid above foramen magnum; needs stabilisation, not posterior fossa decompression alone [14]
Foramen magnum / cervicomedullary tumour	Vertigo, lower-CN palsies, long-tract signs	Enhancing mass on MRI; treated by resection [16]
Multiple sclerosis	Vertigo, ataxia, nystagmus	White-matter lesions; no Valsalva headache [16]
Vertebrobasilar insufficiency	Vertigo, drop attacks, occipital pain	Position-dependent; vascular risk; MRA abnormal [16]
Syringomyelia of other cause	Cape sensory loss, hand wasting	Cord tumour or post-traumatic syrinx without herniation [12,40]

Bony and structural lesions of the craniovertebral junction round out the differential. Basilar invagination and atlantoaxial instability compress the cervicomedullary junction and may coexist with or secondarily produce tonsillar herniation, but require stabilisation rather than — or in addition to — posterior fossa decompression, a distinction made on CT and dynamic imaging [14]. Foramen-magnum meningiomas and other cervicomedullary tumours mimic Chiari and are excluded by identifying the enhancing mass [16]. When a syrinx is found without significant herniation, the clinician must actively search for a small overlooked tonsillar descent (Chiari 0) or an alternative cause such as a cord tumour or tethered cord before attributing it to Chiari [12,22].

VII. Management — Conservative Care and Surgical Decompression

Management is stratified by type, symptom burden, and the presence of a syrinx or progressive deficit [16,34]. The guiding principle in CM-I is to avoid unnecessary surgery in mild or incidental disease while intervening decisively where neurological function is threatened [34,43].

Conservative management

Asymptomatic and mildly symptomatic CM-I without a syrinx is generally managed conservatively, and prospective natural-history data support this restraint: most non-operatively managed patients remain stable over years, and asymptomatic tonsillar ectopia rarely progresses [34,44,45]. Care centres on activity and Valsalva modification, vestibular rehabilitation for dizziness and imbalance, cervical physiotherapy for neck pain, and periodic clinical and MRI surveillance for the emergence of a syrinx or new deficit [34,45]. Vestibular rehabilitation merits emphasis in this audience: targeted gaze-stabilisation and balance training reduce oscillopsia and disequilibrium and are valuable both as primary therapy in mild disease and for residual central deficits after surgery [10,43].

Patient education frames the conservative pathway. Patients are counselled to report new limb weakness, numbness, sphincter disturbance, or worsening headache promptly, as these may herald syrinx progression and a shift to operative management [34,45]. A trial of conservative therapy is justified for moderate, non-progressive symptoms provided neurological monitoring is maintained [43,45].

The evidence base for conservative care has matured. Prospective natural-history cohorts of nonoperatively managed CM-I report that the large majority of patients neither deteriorate neurologically nor develop a new syrinx over years of follow-up, and that asymptomatic tonsillar ectopia in particular

behaves benignly [44,45]. This underwrites a confident policy of watchful waiting in the mild and incidental patient, provided the safety-netting is explicit and imaging is repeated should symptoms or signs evolve [34,45].

Surgical decompression

Surgery is indicated for disabling or progressive symptoms, a syrinx, or objective neurological deficit [16,24]. The standard operation is posterior fossa (foramen magnum) decompression by suboccipital craniectomy and C1 laminectomy, which enlarges the posterior fossa and re-establishes CSF flow [24,38]. Most patients improve: contemporary series report meaningful symptom improvement in roughly 70–85%, with headache responding particularly well and syrinxes shrinking after adequate decompression [24,37,38].

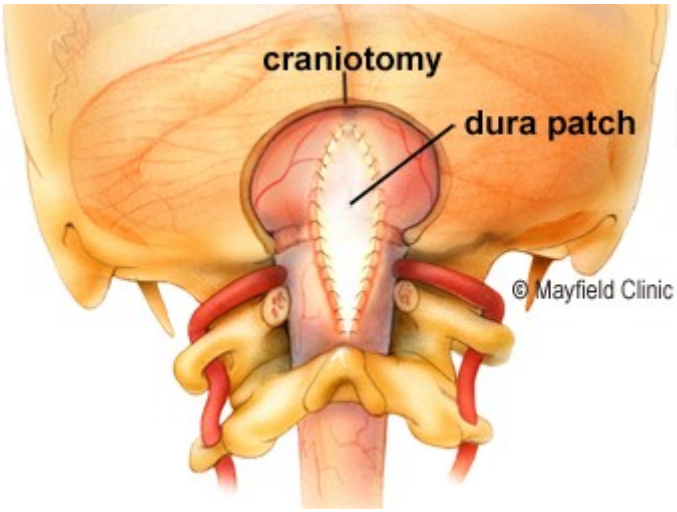

Figure 8. Posterior fossa decompression. Suboccipital craniotomy enlarges the posterior fossa and a dural patch (duraplasty) expands the space for the cerebellum and restores CSF flow.
Source: Mayfield Clinic.

The central technical controversy is whether to open the dura. Bone-only decompression carries a lower rate of CSF leak and aseptic meningitis but a higher rate of persistent syrinx and reoperation, whereas opening the dura and adding a duraplasty achieves more complete decompression and better syrinx resolution at the cost of greater morbidity [24,25,26]. Meta-analyses in children show duraplasty reduces reoperation but increases CSF-related complications, so practice is individualised — bone-only decompression is often reserved for children without a syrinx, with duraplasty favoured where a syrinx is present or bony decompression proves inadequate [25,26]. Adjunctive tonsillar coagulation to enlarge the CSF space remains debated given the theoretical risk of neural injury [24].

Where hydrocephalus accompanies the malformation — the rule in Chiari II and occasional in CM-I — CSF diversion by ventriculoperitoneal shunt or endoscopic third ventriculostomy is undertaken first, and may itself relieve hindbrain pressure [16,28]. In Chiari II the transformative advance is prenatal myelomeningocele repair: the MOMS trial showed in-utero closure reduces hindbrain herniation and the need for shunting, with later reports describing substantial reversal of the herniation by birth [27].

VIII. Pharmacological Adjuncts, Refractory Disease and Associated Conditions

No drug reverses the structural malformation, so pharmacotherapy is directed at symptoms and complications [31]. A systematic review of pain management in CM-I found that standard analgesics, NSAIDs, and muscle relaxants predominate, with limited evidence for any disease-specific agent and a clear movement toward non-opioid, neuropathic-pain strategies such as gabapentinoids and tricyclics for syrinx-related and chronic pain [31]. Table 4 summarises symptom-targeted options.

Table 4. Symptom-based pharmacological management in Chiari malformation.

Symptom	Options	Notes

Suboccipital / cough headache	Simple analgesics, NSAIDs; trial of indomethacin	Indomethacin response suggests primary cough headache rather than Chiari [31,33]
Neuropathic / syrinx pain	Gabapentinoids, tricyclics, SNRIs	Preferred over long-term opioids [31]
Downbeat nystagmus / oscillopsia	4-aminopyridine, clonazepam	Reduces nystagmus intensity when decompression is incomplete [3]
Vertigo / nausea	Short-course vestibular sedatives, antiemetics	Symptomatic only; avoid chronic use [10]
Raised ICP with borderline Chiari	Acetazolamide	Selected acquired/high-pressure cases; not standard [42]

A specific caveat governs pharmacotherapy: medication must never substitute for decompression where a deficit is progressing, because analgesia or vestibular sedation will mask advancing syrinx-related injury while the underlying compression continues [16,31]. Symptomatic drugs are adjuncts within a multidisciplinary plan that includes physiotherapy and lifestyle measures [31,43].

Refractory and recurrent disease is usually mechanical. Persistent or recurrent symptoms after decompression most often reflect an inadequate initial decompression, scarring that re-obstructs CSF flow, or a syrinx that fails to collapse, and reoperation — frequently with dural opening or duraplasty where the first procedure was bone-only — is effective when appropriately targeted [24,25,37]. In patients with heritable connective-tissue disorders, ongoing craniocervical instability can undermine an anatomically adequate decompression, and occipitocervical fusion is considered where instability is demonstrated [14].

Several associated conditions shape vestibular management. Comorbid migraine is common and may persist after the Valsalva headache is cured by surgery, so it should be treated on its own merits rather than attributed wholesale to the malformation [33]. A tethered cord may coexist and contribute to caudal traction on the hindbrain, raising the question of which to address first [12,40]. Idiopathic intracranial hypertension can masquerade as, or coexist with, CM-I, and its identification redirects treatment toward pressure control rather than posterior fossa decompression [42].

IX. Prognosis, Recurrence and Special Populations

CM-I carries a favourable prognosis when managed appropriately [34,38]. Many patients with mild disease remain stable for years on conservative care, and among those decompressed, about 70–85% achieve meaningful or complete relief of their primary symptoms, with headache the most reliably improved and syrinxes shrinking in the majority after successful surgery [24,37,38]. Roughly three-quarters of operated patients report satisfaction and return to near-normal activity [38].

Table 5. Outcomes and recurrence in Chiari I malformation.

Outcome measure	Approximate figure	Notes
Symptom improvement after decompression	~70–85%	Headache responds best; syrinx usually shrinks [24,37,38]
Symptom recurrence / progression (long-term)	~10–30%	Scarring, syrinx re-expansion, ongoing instability [37,38]
Reoperation rate	~5–10%	~7% needing second surgery in a recent cohort [37]
Scoliosis needing orthopaedic care	~20% of syrinx-associated curves	Often stabilises if decompressed pre-puberty [40]

A minority relapse. Long-term follow-up shows symptom recurrence or progression in roughly 10–30% of patients, driven by scarring at the decompression site, slow syrinx re-expansion, or — in connective-tissue disorders — continued cervicomedullary settling [37,38]. Reoperation is required in about 5–10% of modern series; an Eastern Finland cohort reported approximately 7% needing a second posterior fossa

procedure, with targeted revision generally effective [37]. Overall mortality from CM-I is very low, confined to rare severe brainstem compression or intractable central apnoea [38].

Chiari II carries a more guarded outlook tied to the associated spina bifida and hydrocephalus. Myelomeningocele closure and shunting greatly improve survival, but lifelong deficits — lower-limb paralysis, neurogenic bladder and bowel, scoliosis — are common, and severe neonatal brainstem dysfunction predicts higher morbidity [27,28]. Modern multidisciplinary care, including fetal surgery, allows most affected children to reach adulthood, albeit with ongoing medical needs [27,28].

Special populations require tailored counselling. Scoliosis associated with a syrinx often stabilises or improves after early decompression, though about a fifth of advanced curves still need bracing or fusion [40]. Significant preoperative central sleep apnoea, particularly in children, frequently improves with decompression but portends a more serious course if it persists [15]. Uncomplicated CM-I is not a contraindication to pregnancy; most pregnancies are uneventful, and where the malformation is symptomatic, many clinicians favour epidural analgesia to blunt the haemodynamic and CSF-pressure spikes of bearing down, individualising the mode of delivery rather than mandating caesarean section [14,16]. Some residual disequilibrium may persist despite anatomically successful surgery, reflecting longstanding central vestibular change, and is the proper target of ongoing vestibular rehabilitation [10,43].

Patient-reported outcome research adds nuance to the headline figures. Headache and general quality-of-life scores improve in most operated patients, yet a consistent minority report persistent fatigue, neck pain, or imbalance that anatomically successful decompression does not abolish, underlining that some central vestibular change becomes fixed once established [38,43]. Setting this expectation before surgery — that decompression reliably treats the cough headache and arrests syrinx progression but may leave residual disequilibrium for rehabilitation to address — is central to honest counselling [16,43].

X. Controversies and Future Directions

Several debates remain unresolved and bear directly on vestibular practice [16,21]. The first is the threshold and timing of surgery. There is consensus to operate on clear progressive deficit or symptomatic syrinx, but the management of borderline herniation with compatible symptoms divides opinion — whether to 'treat the patient, not the millimetres' or to demand firm anatomical criteria — and this is sharpest in children with headache alone, some of whom outgrow symptoms [34,36,43]. Severity indices and CSF-flow metrics are attempts to bring objectivity to a decision that remains partly judgement [20,36].

The duraplasty question — bone-only versus dural opening — is the most active surgical controversy, balancing lower complication rates against higher reoperation, and is the subject of ongoing comparative study [24,25,26]. The status of Chiari 0, in which a syrinx coexists with minimal herniation, is similarly contested: some recognise it and offer decompression with good results, while others insist on excluding alternative causes of the syrinx first, and the entity is confirmed only retrospectively if the syrinx resolves after surgery [22]. Proposed refinements such as Chiari 1.5 likewise prompt debate over whether finer subclassification is clinically useful [23].

The relationship between CM-I, connective-tissue disorders, and intracranial pressure is an expanding frontier. Hypermobile patients are over-represented in Chiari clinics, raising unsettled questions about routine screening and about whether craniocervical fusion should accompany decompression [14]. Conversely, evidence that some apparent Chiari reflects raised intracranial pressure — reversible with acetazolamide or CSF diversion — blurs the boundary with idiopathic intracranial hypertension and cautions against reflexive decompression of borderline herniation with pressure signs [42]. Incidental CM-I discovered on scans performed for other reasons poses the recurring dilemma of when, if ever, to intervene in the asymptomatic patient; the prevailing answer is observation unless a syrinx or symptoms appear [4,34,45].

Future progress is likely to come from physiology and genetics rather than from millimetres. Phase-contrast and computational CSF-flow modelling promise quantitative biomarkers of which incidental Chiari will become symptomatic and which patients will benefit from surgery [20,41]. Genome-wide and candidate-gene studies continue to define the polygenic architecture of idiopathic CM-I and may eventually stratify patients by mechanism — bony hypoplasia versus cranial settling — with divergent optimal management [7,9]. For the vestibular physician, the enduring lesson is that Chiari sits at the

intersection of neurology, neurosurgery, radiology, and vestibular medicine, and that the central vestibular features — downbeat nystagmus, central positional patterns, and progressive disequilibrium — are often the earliest and most actionable clues to the diagnosis [3,10].

□ **Clinical Pearl:** Five practice points consolidate this review:

- **Keep Chiari on the central differential —** about 1 dizziness-clinic patient in 500 harbours an occult CM-I, often without the cough headache [10].
- **Trust the central oculomotor signs —** fixation-present downbeat or direction-changing nystagmus warrants craniocervical MRI [3].
- **Read physiology, not just millimetres —** cine CSF flow and a whole-spine syrinx screen turn a bare measurement into a defensible decision [20,40].
- **Exclude the reversible mimics —** intracranial hypotension and raised ICP produce acquired herniation that must not be decompressed [42].
- **Rehabilitate the residue —** vestibular rehabilitation helps both mild disease and the disequilibrium that persists after surgery [43].

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