# Headache in Chiari Malformation

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**KEYWORDS**

• Chiari malformation • Chiari 1 malformation • Headache • Cough-associated headache

**KEY POINTS**

- The majority of Chiari I malformation (CMI) patients experience headaches, but not all such headaches can be directly attributable to the diagnosis.
- Transient cough-associated headache is the most distinctive symptom of CMI.
- Certain routine imaging findings may suggest that the headache in a CMI patient is likely associated with the diagnosis, but their absence does not preclude an association.
- A normal cine phase-contrast study suggests that the headache is not caused by CMI. However, an abnormal cerebrospinal fluid (CSF) flow study is only moderately predictive of an association between the headache and CMI.
- Real-time CSF flow imaging with coughing as a physiologic challenge holds great promise for an objective evaluation of CMI patients with headache, but further studies are needed.

**Video content accompanies this article at http://www.neuroimaging.theclinics.com.**

## INTRODUCTION

Chiari malformations are a collection of hindbrain and craniocervical junction abnormalities (**Table 1**), of which Chiari I malformation (CMI) is the most commonly seen type in clinical practice, with a reported prevalence of 0.56% to 0.75% on MRI.[1,2] The main imaging feature of CMI is cerebellar tonsillar herniation of 5 mm or greater below the level of foramen magnum (**Fig. 1**),[3] with headache being the most common symptom.[4] Recently, many have questioned this anatomic criteria for the diagnosis of CMI given that many fitting this criteria are asymptomatic or have no specific symptoms related to tonsillar herniation.[5] Nevertheless, until a better criterion emerges, the degree of tonsillar herniation has remained the only accepted definition of CMI. Many patients with primary or congenital CMI are believed to have a skull base bony abnormality,[6] which can vary from a shallow posterior fossa, retroflexed odontoid, or short basilar process, to fusion between the occiput and C1. The term secondary or acquired CMI is often used when tonsillar herniation meeting the diagnostic criteria for CMI is caused by downward pressure by a posterior fossa mass or results from intracranial hypotension or intracranial hypertension.[6] For the purpose of this discussion, CMI will be used to refer to primary or congenital CMI, and when referring to secondary or acquired CMI, it will be specifically stated.

Disclosure Statement: Nothing to disclose.
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Neuroimag Clin N Am 29 (2019) 243–253
https://doi.org/10.1016/j.nic.2019.01.005

**Table 1**
**Types of chiari malformations**

| Type of Chiari Malformation | Pathology and Imaging Findings |
|---|---|
| Chiari I<br>• Chiari 0<br>• Chiari 1.5 | 5-mm ≥ Cerebellar tonsillar herniation below foramen magnum<br>Small posterior fossa and syringomyelia without tonsillar herniation below foramen magnum<br>Cerebellar tonsillar plus obex herniation below foramen magnum |
| Chiari II | Herniation of cerebellum and hindbrain below foramen magnum plus lumbar myelomeningocele |
| Chiari III | Cranio-cervical meningo-encephalocele |
| Chiari IV | Cerebellar hypoplasia |

Chiari II, III, and IV usually present with severe neurologic abnormalities in the neonatal period or early childhood,[7] and headache is typically not a presenting or important symptom. Given the preponderance of data available related to headache in CMI regarding types of headache, pathophysiology, and imaging features, discussion of headache in this article will be limited to CMI.


**Fig. 1.** The diagnosis of CMI is typically made when there is 5 mm or greater downward displacement of the cerebellar tonsils below the foramen magnum as measured on sagittal MRI. In this patient, a 10 mm tonsillar herniation is seen.

## TYPES AND LOCATION OF HEADACHE IN CHIARI I MALFORMATION

Prevalence of headache of any type in CMI is believed to be around 81% of patients at presentation.[8] The headaches experienced by CMI patients are transient activity-associated (related to activities such as coughing, Valsalva, sneezing, laughing, or exercise, but are collectively known as cough-associated), migraine, tension-type, or of the cluster variety.[9] Of all headache types experienced by CMI patients, transient cough-associated (seconds to <5 min duration) is the most distinctive and is seen in approximately one-third of patients.[9,10] In a study of 30 CMI patients, the authors found 9 patients (30%) presented with typical cough-associated headache.[11] Recently, Curone and colleagues[10] found that 87% of CMI patients with headache complained of some increase in intensity with cough or other activities. However, only 34% of their patients had short-lasting cough-associated headache. Thus, it is essential to distinguish the change in intensity of long-lasting headaches with cough or other activities from typical cough-associated headaches of CMI by their temporal relationship and short duration. The criteria for the diagnosis for headache attributed to CMI has been established by the International Headache Society (IHS), and it is important for a radiologist interpreting CMI imaging studies to be aware of these criteria to aid in clinical decision making (**Box 1**).[12]

Although CMI headaches are characteristically cough-associated, they need to be differentiated from primary cough headache.[13,14] In clinical practice, cough-associated headaches are reported by about 1% of patients getting consultation for headaches.[15] It is reported that nearly two-thirds of patients with cough-associated headache have an MRI-demonstrable abnormality in the posterior fossa, and 90% of them have CMI.[9] The remaining one-third of headaches is classified as primary or benign cough headache. The major clinical distinction between primary and secondary cough-associated headache is that the primary cough-associated headaches are generally seen in older patients (>60-year in age).[9] They are characterized by bilateral posterior headaches that are temporally related to cough and last for a few seconds to minutes but can last up to 2 hours, show a male preponderance, and respond well to treatment with pain medications.[15]

As opposed to primary cough-headache, the secondary cough-associated headaches are

**Box 1**
**International classification of headache disorders criteria for the diagnosis of headache attributed to Chiari I malformation**

A. Headache fulfilling criterion C
B. CM1 has been demonstrated
   1. 5 mm or greater cerebellar tonsillar herniation below foramen magnum
   2. 3 mm cerebellar tonsillar herniation plus crowding of the subarachnoid space at the craniocervical junction as evidenced by compression of the CSF spaces posterior and lateral to the cerebellum
   3. Reduced height of the supraocciput, or increased slope of the tentorium, or kinking of the medulla
C. Evidence of causation demonstrated by at least 2 of the following
   1. Either or both of the following
      a. Headache has developed in temporal relation to the CM1
      b. Headache has resolved within 3 months after successful treatment of the CM1
   2. Headache has at least one of the following 3 characteristics
      a. Precipitated by cough or other Valsalva-like maneuver
      b. Occipital or suboccipital location
      c. Lasting less than 5 minutes
   3. Headache is associated with other symptoms and/or clinical signs of brainstem, cerebellar, lower cranial nerve, and/or cervical spinal cord dysfunction
D. Not better accounted for by another ICHD-3 diagnosis

*From* Headache Classification Committee of the International Headache Society (IHS). The International Classification of Headache Disorders, 3rd edition (beta version). Cephalalgia 2013;33(9):713; with permission.

seen in younger (<40 years of age) patients; they are predominantly sub-occipital and have female preponderance. In a recent study by Alperin and colleagues,[16] 85% of patients with cough-associated headache and CMI were female. Given the high prevalence of posterior fossa abnormalities in young patients with cough-associated headache, it is recommended that all these patients should undergo an MRI examination to exclude a posterior fossa abnormality such as CMI.[9] In addition to the brief episodes of cough-associated headaches, many CMI patients can have continuous or long-lasting headaches.[17] Some CMI patients can also have continuous daily headache (CDH) and in some, CDH can be exaggerated by cough. A hypothetical relationship between CDH and CMI has been suggested but not proven by controlled clinical studies.[9]

IHS defines Chiari attributed cough-associated headache to be typically suboccipital in location. However, many variations of that pattern exist. These variations are in their association with cough and also in their location. For example, some CMI patients have brief cough-associated headaches that are not suboccipital but could be frontal or on the side of head. Alperin and colleagues[18] divided CMI-associated headaches in 3 categories. In 63 CMI patients with headache, 40 patients (63%) had suboccipital Valsalva-related headaches; 15 patients (24%) had suboccipital headache not related to Valsalva, and 8 patients (13%) had nonoccipital non-Valsalva-related headaches (it should be noted that the terms cough- and Valsalva-related headache are often used interchangeably). For the authors' ongoing research endeavors in CMI patients with headache, different classifications and location of headaches have been determined, as demonstrated in **Table 2**.

**Table 2**
**Authors' proposed classification of headaches in Chiari I malformation**

| Type of Headache | Characteristics of Headache |
|---|---|
| Type 1 | • Transient localized suboccipital cough-related headaches (headache triggered by cough, Valsalva, sneezing, exercise, bending forward, or laughing) |
| Type 2 | • Transient localized nonoccipital or generalized cough-related headaches |
| Type 3 | • Constant localized (occipital or nonoccipital) or generalized headache (constant daily headache) that may be exacerbated by cough |
| Type 4 | • Transient or constant suboccipital headaches that are not cough related |
| Type 5 | • Transient or constant localized nonoccipital or generalized headaches that are not cough related |

## PATHOPHYSIOLOGY OF HEADACHE IN CHIARI MALFORMATION

It is generally agreed that the underlying pathophysiology of cough-associated headache in CMI is the result of impaired CSF flow between the head and spine secondary to tonsillar herniation and neural crowding at the foramen magnum.[19] However, consensus has not been reached as to how the CSF flow obstruction produces cough-associated headache.

Previously published invasive pressure studies by Williams and Sansur and colleagues described pressure changes in the head and spine in CMI patients with cough-associated headache.

Williams assessed pressure changes at rest and after coughing in CMI patients and showed that physiologic challenges such as coughing or Valsalva maneuver produce pressure dissociation between the head and spine.[20] The development of pressure dissociation is explained by an initial elevation of spinal pressure (from increased intrathoracic pressure and consequent distension of the epidural veins) during coughing, displacing CSF to the head (**Fig. 2**). However, immediately after coughing, this displaced CSF returns with relative ease to the spinal canal in a healthy patient but not in a patient with CMI because of impaction of the cerebellar tonsils, thereby creating a pressure dissociation between the head and spine, where intracranial pressure is higher than that of the spine (see **Fig. 2**B, D).The transiently increased intracranial pressure results in a brief headache by stimulation of pressure receptors of the dura. It was also shown by Williams that there was minimal to no pressure dissociation in these CMI patients at baseline or rest (in absence of cough).

Sansur and colleagues[21] measured spinal intrathecal pressures and observed increased pressures at baseline and during coughing in patients with CMI and cough-associated headache compared with CMI patients without cough-associated headache and normal subjects. They suggested that headache in CMI may be caused by the absolute value of spinal CSF pressure reached during a cough and concluded that elevated spinal CSF pressure in CMI patients with cough-associated headache is associated with significant foramen magnum obstruction to CSF flow, which increases intracranial pressure and results in the headache.

## IMAGING APPROACH AND IMAGING FINDINGS

### *Anatomic Imaging*

It is the authors' experience that in most patients with CMI, diagnosis is usually made on a routine brain or cervical spine MRI obtained to evaluate headache, neck pain, and paresthesia or for other unrelated reasons such as trauma evaluation. The diagnosis of CMI is made when there is 5 mm or greater downward displacement of the cerebellar tonsils below the foramen magnum (see **Fig. 1**). Tonsillar displacement of 3 to 5 mm is often referred to as tonsillar ectopia, and less than 3 mm tonsillar herniation is considered to be within normal limits.[3] From here, the imaging approach depends on what information is already available and the clinical circumstances of the initial imaging. The main goal for any additional imaging is to assess the brain, spinal cord, and the craniocervical junction.

#### *Brain*

Routine brain imaging is required to exclude hydrocephalus, mass, or manifestations of intracranial hypotension, often referred to as acquired CMI. Use of gadolinium is warranted when a posterior fossa mass is suspected, which can displace the cerebellar tonsils downwards and mimic primary CMI. Gadolinium is also useful when intracranial hypotension is suspected (such as with positional headache elicited in the upright position that is relieved when lying down), which is characterized in imaging by diffuse pachymeningeal enhancement.

#### *Spinal cord*

In patients with CMI, imaging of the entire spinal cord is required to exclude a spinal cord syrinx (syringohydromyelia). A syrinx is seen in up to 34% to 40% of patients with CMI.[2,22,23] Syrinx is generally defined as linear/cylindrical CSF intensity collection in the spinal cord of 2 mm or greater width.[22] A linear CSF collection in the central aspect of the spinal cord less than 2-mm is generally considered a prominent central canal. In patients with an unequivocal diagnosis of CMI, the authors do not recommend use of gadolinium for evaluation of spinal cord syrinx. However, many recommend the use of contrast at the initial evaluation but not for the follow-up. Many CMI patients with syrinx also complain of headache, but the exact number has not been previously reported.

#### *Cranio-cervical junction imaging*

The authors use high-resolution 3-dimensional T2-weighted images like FIESTA or CISS for dedicated imaging of the craniocervical junction. These images provide exquisite detail of CSF spaces in the region and help assess some of the findings that will be described.

Anatomic imaging is generally considered to be less reliable in predicting which patients have CMI-associated headache.[11,18] However,

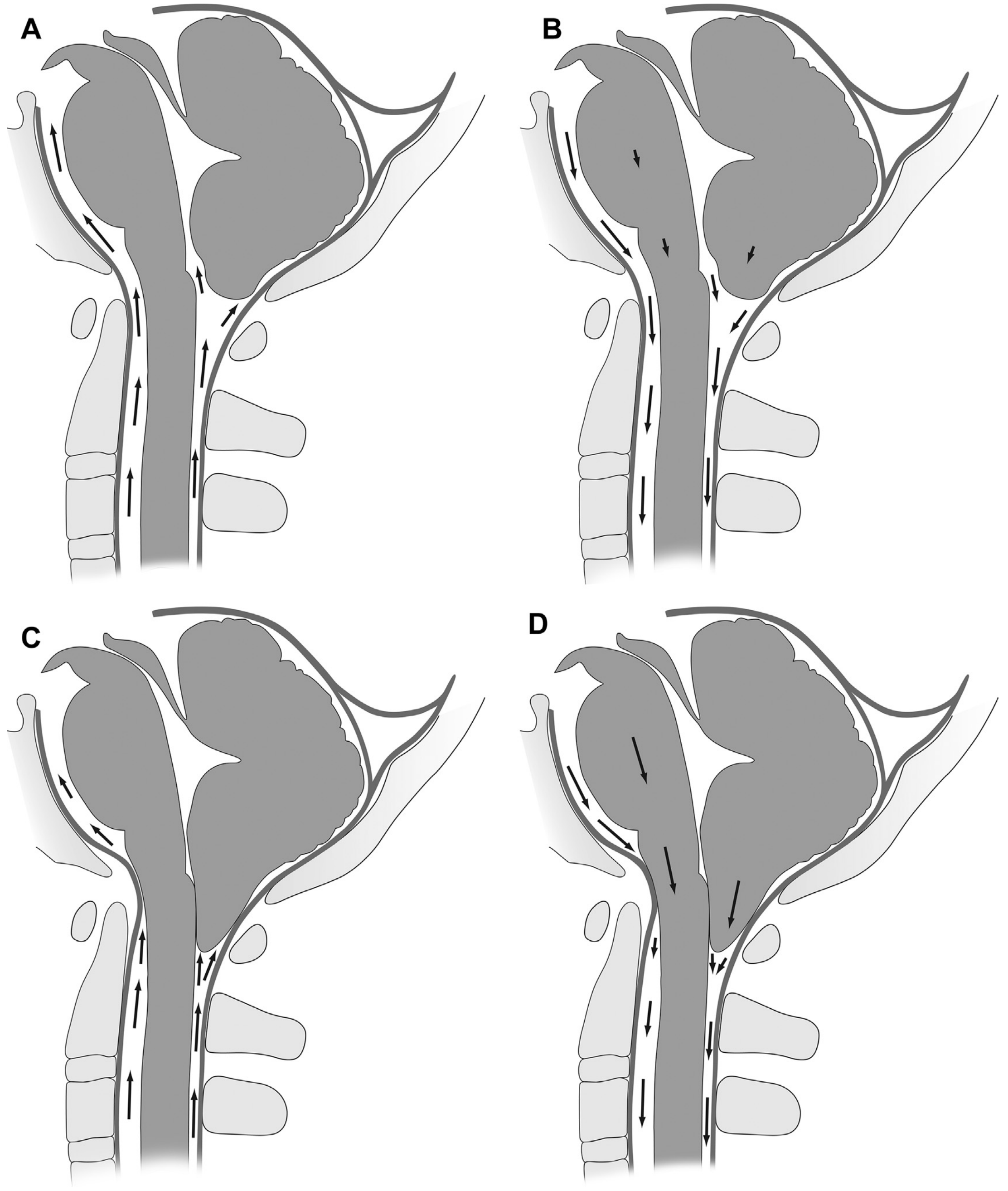


**Fig. 2.** (*A*, *B*) Control patient; (*C*, *D*) CMI patient. During coughing there is an elevation of spinal pressure (from increased intrathoracic pressure and consequent distension of the epidural veins), which displaces CSF to the head (*A*, *C*). However, immediately after coughing, this displaced CSF returns with relative ease to the spinal canal in a healthy patient (*B*), but not in a patient with CMI because of further impaction of the cerebellar tonsils. This creates pressure dissociation between the head and spine, where intracranial pressure is higher than spinal pressure (*D*).

there are certain anatomic imaging findings that suggest the patient's headache may be related to CMI. Degree of tonsillar decent of greater than 12 mm, deformity of the tonsils with a pointed shape, crowding of the neural structures, and effacement of the retro-cerebellar cisterns are findings that suggest that the headache is more likely to be associated with CMI[2] (**Fig. 3**A). In contrast, tonsillar herniation without obliteration of fourth ventricular recess and rounded tonsils

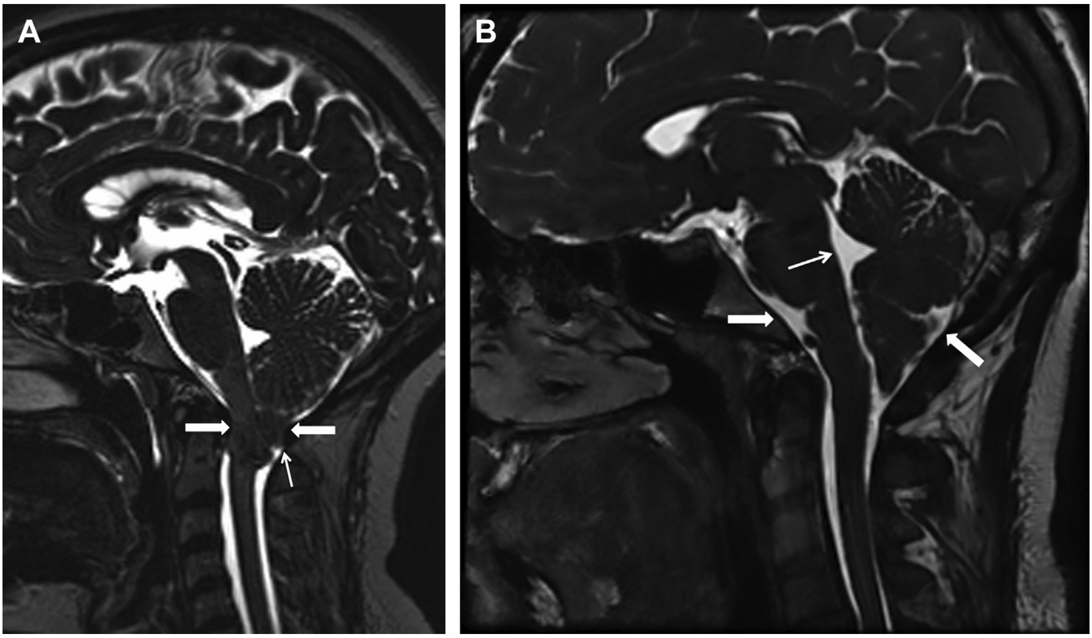


Fig. 3. (*A*) Anatomic imaging findings suggesting the patient's headache symptoms are related to CMI include tonsillar herniation of 14 mm (>12 mm), pointed shape of the tonsils (*skinny arrow*), crowding of the neural structures, and effacement of the retrocerebellar cisterns (*block arrows*). (*B*) Anatomic imaging findings suggesting that headache symptoms are likely not associated with CMI include tonsillar herniation of 10 mm (<12 mm) without obliteration of fourth ventricular recess (*skinny arrow*) and rounded tonsils with open subarachnoid space at the cervicomedullary junction/foramen magnum (*block arrows*).

with open subarachnoid space at the cervicomedullary junction/foramen magnum are less likely to be suggestive of the headache associated with CMI (**Fig. 3**B).

Milhorat and colleagues[8] also measured dimensions of the posterior cranial fossa with MRI and suggested that in CMI patients with cough-associated headache, the posterior fossa is smaller than those without it, leading to reduced compliance and therefore headache from transiently elevated intracranial pressure. Alperin and colleagues[18] (2015) measured several anatomic and physiologic (CSF and blood flow) parameters and demonstrated that in CMI patients with cough associated suboccipital headache, intracranial volume (and thereby compliance) is lower than those without cough-associated suboccipital headache.

## *Physiologic Imaging*

Although anatomic MRI provides detailed information about foramen magnum narrowing through assessment of the degree of tonsillar herniation and crowding of the neural structures, these findings are poorly correlated with the type and severity of headaches experienced by CMI patients.[11,18] Because it is generally agreed that CMI-associated headache is caused by impairment of CSF flow at the level of the foramen magnum, motion-sensitive MRI techniques (mainly cine phase-contrast [cine-PC]) have been used to help guide patient management. Cine-PC is available on most scanners and can be utilized in patients with CMI. Sagittal cine-PC provides an excellent means for qualitative visual inspection of CSF spaces at the craniocervical junction, and the authors use them extensively. Axial cine-PC is important for quantitative evaluation of velocity profile and calculation of flow rates.[24–26] The authors generally use a velocity encoding (VENC) of 5 to 7 cm/s with a superior to inferior directional encoding in scanning adult patients with CMI. The VENC can be increased to 10 to 15 cm/s in children to eliminate the potential for velocity aliasing. Quantitative evaluation requires dedicated software for analysis. Hofkes and colleagues[27] used sagittal and axial cine-PC to correlate different CSF flow patterns with CMI symptomatology. The authors use sagittal qualitative assessment of cine-PC to grade severity of CSF flow obstruction in CMI (**Fig. 4**, **Table 3**, Videos 1–4).

Work correlating CSF flow abnormalities at the cranio-cervical junction with CMI patient head type and anatomic abnormalities at the foramen magnum has been performed by McGirt and colleagues,[28] used cine-PC imaging in 33 patients

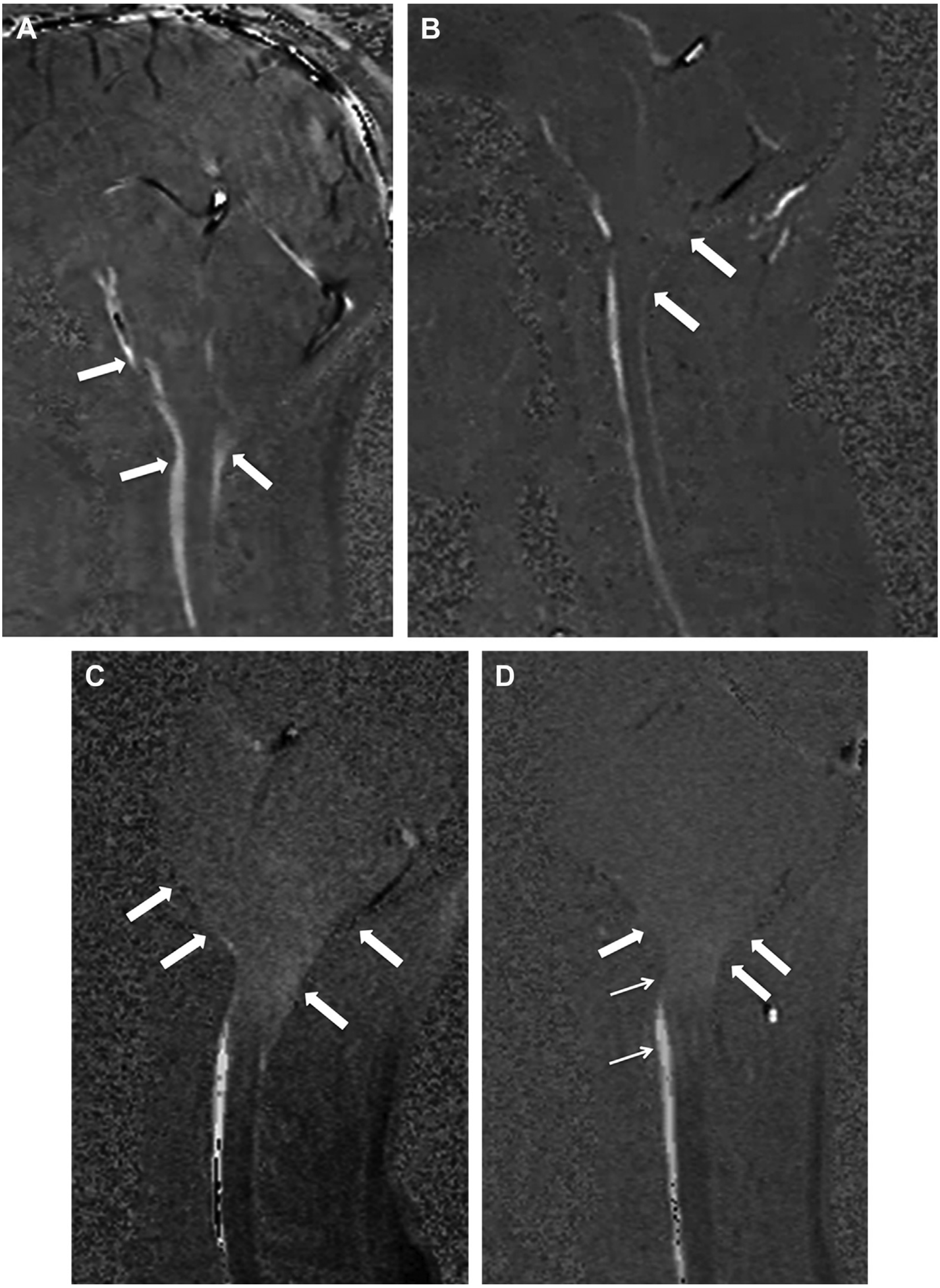


Fig. 4. (*A–D*) Different severities (qualitative) of craniocervical junction CSF flow obstruction on cine-PC images in a patient with CMI. (*A*) Demonstrates a normal CSF flow pattern as evidenced by continuous uninterrupted anterior CSF systolic flow with flow also noted in the posterior fossa as well as posterior to the upper cervical cord (*block arrows*; CSF diastolic phase not shown). (*B*) Demonstrates mild obstruction as evidenced by normal anterior flow, but decreased/absent flow in the posterior fossa and posterior to the upper cervical cord (*block arrows*). (*C*) Demonstrates moderate obstruction as evidenced by partially interrupted anterior systolic flow at the level of the foramen magnum as well as absent flow in the posterior fossa and posterior to the upper cervical cord (*block arrows*). (*D*) Demonstrates severe obstruction as evidenced by interrupted CSF systolic flow at the foramen magnum (*block arrow*), asynchronous flow between the level just below the foramen magnum and the C2-C4 level (*skinny arrow*), as well as absent flow in the posterior fossa and posterior to the upper cervical cord (*block arrows*).

**Table 3**
**Qualitative interpretation of craniocervical junction cerebrospinal fluid flow on cine-phase contest images in patient with Chiari I malformation**

| CSF Flow Pattern | Anterior CSF Flow | Posterior CSF Flow |
|---|---|---|
| Normal (see Fig. 4A, Video 1) | • Continuous uninterrupted bidirectional systolic and diastolic flow present<br>• Flow immediately below foramen magnum is synchronous with flow at C2-3 and C3-4 | • Foramen Magendie/fourth ventricular flow present<br>• Posterior upper cervical flow present |
| Mild obstruction (see Fig. 4B, Video 2) | • Normal | • Foramen Magendie/4th ventricular flow: decreased or absent<br>• Posterior to upper cervical cord (below tonsils) flow decreased but present |
| Moderate obstruction (see Fig. 4C, Video 3) | • Partially interrupted bidirectional systolic and diastolic flow (flow interruption seen in a couple to several phase) | • Foramen Magendie/4th ventricular flow absent.<br>• Posterior to upper cord (below tonsils) flow absent |
| Severe obstruction (see Fig. 4D, Video 4) | • Interrupted bidirectional systolic and diastolic flow at foramen magnum<br>• Flow immediately below foramen magnum asynchronous with flow at C2-3 and C3-4 | • Foramen Magendie/fourth ventricular flow absent<br>• Posterior to upper cord (below tonsils) flow absent |

with CMI. In this study, CSF flow abnormalities on cine-PC were defined as absence of normal biphasic CSF flow within any of the aqueduct, fourth ventricle, foramen of Magendie, foramen magnum, around the cerebellar tonsils, prepontine cistern, and ventral or dorsal to the upper cervical spinal cord. Headaches were divided in to frontal/generalized or occipital. They observed that CMI patients with frontal and generalized headache were 10 times less likely to demonstrate CSF flow abnormalities at the craniocervical junction and 8 times less likely to have tonsillar herniation greater than 7 mm compared with CMI patients with occipital headache. The most important conclusion of this study was that a frontal or generalized headache in the setting of normal cine-PC study (see Fig. 4A) indicates that the headache is not related to CMI and will not benefit from surgery.

Bhadelia and colleagues[11] (2011) studied 30 CMI patients with cine-PC and evaluated differences in the CSF flow pattern between CMI patients with and without cough-related headache. Nine of the 30 patients had cough-associated headache, which was defined as short-lasting occipital headache initiated or aggravated by cough, exertion, or a Valsalva-like maneuver. It was observed that in CMI patients with cough-associated headache, prolonged CSF diastolic flow duration was seen compared to those without cough-associated headache. This can be seen by observing the presence of persistent CSF diastolic flow below foramen magnum on cine-PC images (Fig. 5).

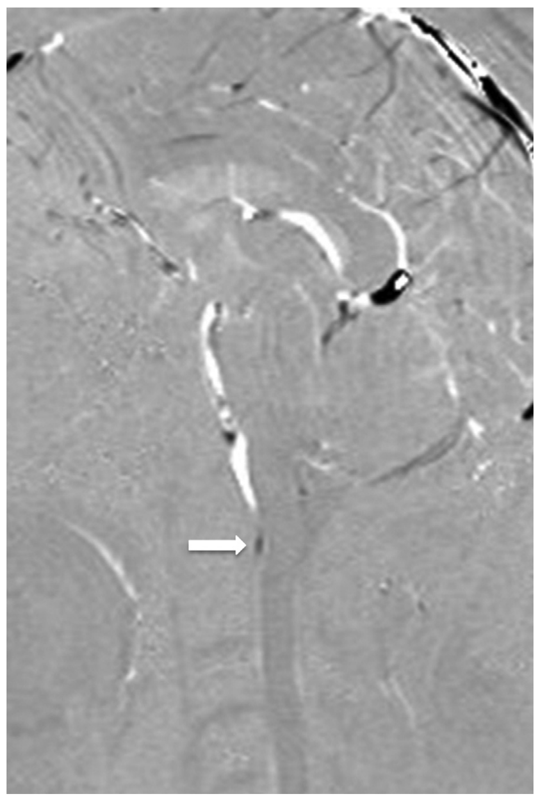

Fig. 5. Prolonged/persistent diastolic flow as evidenced by black signal (*block arrow*) just below the foramen magnum compared with systolic flow (white signal) at other levels above and below which more frequently seen in CMI patients with cough-associated headaches.

By assessing CSF and blood flow with cine-PC, Alperin measured cardiac cycle-related intracranial volume changes, intracranial compliance, and MRI-derived intracranial pressure.[18] The results indicated that CMI patients with cough-associated suboccipital headache had lower intracranial volume change, lower intracranial compliance index, and higher MRI-derived intracranial pressure compared with CMI patients with other types of headaches.

Bhadelia and colleagues evaluated cough-associated changes in CSF flow by utilizing a novel real-time pencil-beam MRI technique.[19] Those interested in the detailed description of the technique are referred to previous publications.[19,29] Each real-time scan was acquired for approximately 90 seconds, during which time the patient was asked to: breathe quietly for the first 15 to 20 seconds, then cough as forcefully as possible consecutively 6 times, and breathe quietly again after the end of coughing period until the end of the acquisition time. Results provided the first objective demonstration of CSF flow changes in CMI patients with cough-associated headache.[19,29] In CMI patients with cough-associated headache, a brief 10- to 15-second decrease in CSF flow was observed below the foramen magnum (**Fig. 6**A). However, such a decrease in CSF flow after coughing was not observed in CMI patients without typical transient cough-associated headache (**Fig. 6**B). These findings are thus congruent with Williams' initial proposal of the underlying pathophysiology of cough headache in CMI patients, which hypothesized that there is decreased CSF flow after coughing below foramen magnum (see **Fig. 2**).[20]

## PEARLS AND PITFALLS

Transient cough-associated headaches are the most distinctive symptom of Chiari I malformation. It is important to assess CSF pathways at the craniocervical junction with high-resolution T2-weighted images as well as with cine-PC to help in the management of a CMI patient with headache. Conditions such as intracranial hypotension need to be excluded before making a diagnosis of CMI.

## WHAT THE REFERRING PHYSICIAN NEEDS TO KNOW

It is not sufficient for the radiologist to simply describe the extent of cerebellar tonsil herniation in CMI. In order to provide imaging information that may be used by the clinician to determine if a patient's headache is likely attributable to CMI, the report should include

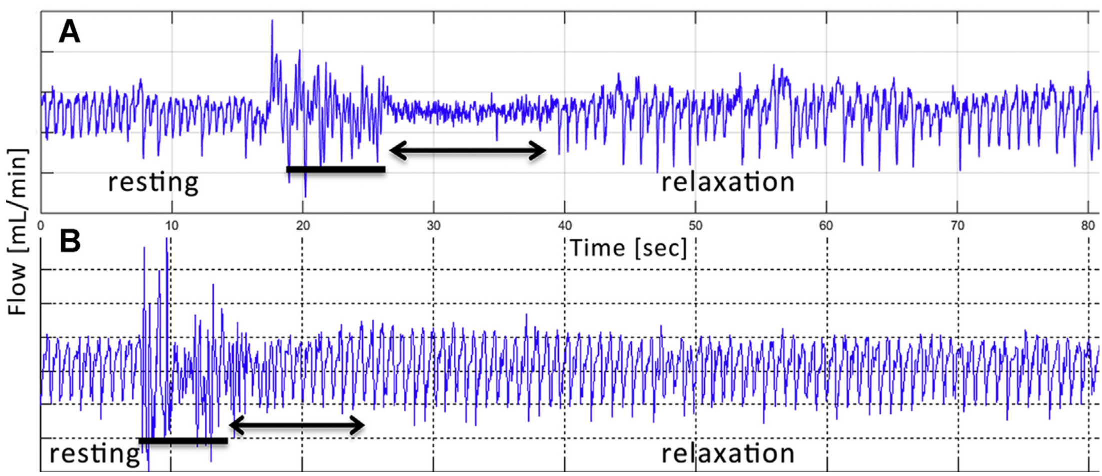


**Fig. 6.** Real-time MRI in 2 different CMI patients with headache demonstrating the effect of coughing on cardiac cycle–related CSF flow waveforms. The x-axis indicates time in seconds; the y-axis, CSF flow rate in milliliters per minute. Left-to-right: resting, coughing (*underlined*), after coughing (*underlined with double-headed arrows*), and relaxation waveforms are seen. CSF stroke volume is the average of the absolute flow from craniocaudal and caudocranial CSF flow. (*A*) (*upper panel*) From a 39-year-old CMI female patient with transient cough-associated headache lasting for about 10 to 15 seconds. Immediately after coughing, there is more than 50% decrease in CSF flow seen lasting for about 12 seconds (*underlined with double headed arrows*), which then returns to normal during the relaxation period (compare resting and relaxation phases with postcoughing phase). (*B*) (*lower panel* From a 34-year old female CMI patient with continuous daily headache. No decrease in CSF flow is seen after coughing, which remains at similar magnitude as resting and relaxation phases (compare resting and relaxation phases with postcoughing phase).

- Whether the tonsils are deformed and pointed versus in a rounded configuration
- If there is obliteration of the retrocerebellar CSF space and foramen of Magendie
- If the CSF pathways of the anterior and posterior cervicomedullary junction are patent or obliterated
- If cine-PC imaging is available, then CSF flow along these pathways should be assessed and described as normal or obstructed (mild, moderate, or severe)
- If there are bony abnormalities such as basilar invagination, retroflexed odontoid, or craniocervical fusion.
- If a syrinx is visualized, evaluation of the entire spinal cord should be recommended
- If brain imaging has not been performed, this should be recommended to exclude secondary causes of tonsillar herniation

## SUMMARY

Headache is a common symptom in patients with CMI, characterized by 5 mm or greater cerebellar tonsillar herniation below foramen magnum. It is important for the radiologist to be aware of the different types of headaches reported by a CMI patient and which headache patterns are distinctive features of the diagnosis. A methodical imaging strategy is required to fully assess a CMI patient to exclude secondary causes of tonsillar herniation such as intracranial hypotension or associated conditions such as syrinx. Both anatomic and physiologic imaging can help determine if headaches are CMI associated, and assist clinicians in therapeutic decision making.

## SUPPLEMENTARY DATA

Supplementary data related to this article can be found online at https://doi.org/10.1016/j.nic.2019.01.005.

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