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IMAGING (L MECHTLER, SECTION EDITOR)

Neuroimaging and the Clinical Manifestations of Chiari Malformation Type I (CMI)

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Abstract Chiari malformation type I (CMI) involves the caudal displacement of the cerebellar tonsils through the foramen magnum with resultant brainstem compression in some individuals. Due to pathophysiologic changes, secondary conditions may arise, such as syringohydromyelia (SH) and scoliosis. This disorder is unique, as the diagnosis is confirmed through radiologic findings. At times CMI is discovered incidentally on neuroimaging, but more frequently a patient will present with specific symptoms, the most common being a prototypic occipital headache. Although the true etiology of this complex condition remains speculative, the advent of neuroimaging has allowed for clarification of the enigmatic relationship between cerebrospinal fluid (CSF) dynamics, neuroanatomical compression, and clinical symptoms. Recent advancements in magnetic resonance imaging (MRI) such as diffusion tensor imaging (DTI) and CSF flow studies show promise in clarifying the underlying fluid dynamics in CMI patients and can aid in the prognosis and diagnosis of this complex disorder.

Keywords Chiari malformation type I · Headache · Neuroimaging · Imaging · MRI · Syringomyelia

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Introduction

The general term "Chiari malformation" (CM) refers to caudal displacement of the cerebellar tonsils through the foramen magnum. The most common type is Chiari malformation type I (CMI) which is prevalent in 0.56 to 1 % of the population [1–3]. This condition is unique as the diagnosis is based on well-defined imaging criteria. Magnetic resonance imaging (MRI) remains the gold standard diagnostic modality in CMI patients. Current advanced MRI techniques such as diffusion tensor imaging (DTI) and CSF flow studies have allowed for a better understanding of the complex underlying pathophysiology of CMI [4•]. The clinical signs and symptoms of CMI are believed to be a manifestation of these anatomical changes. Headache is the most common presenting symptom, but the severity of CMI does not always correlate with the degree of head pain [1, 2, 4•]. Gaining a better understanding of the relationship between the pathophysiologic changes as a cause for symptoms such as headache allows for more efficacious treatment decisions. Recently, selection criteria for individuals evaluated for Chiari decompression surgery has been recommended based on these imaging findings to improve outcomes [5].

Definition

CM is a complex disorder that involves many subtypes. Due to the high prevalence, CMI will be the only subtype discussed in this article. CMI is defined by (Table 1) the degree of cerebellar tonsillar extension below the basion-opisthion line on sagittal and coronal MR images [4•] (Fig. 1). In CMI, tonsillar descent extends greater than 6 mm below the line in patients younger than 15 years old and greater than 5 mm below the line in patients older than 15 years of age. The fourth ventricle remains in the normal position, and

Table 1 Definition and classification of CMI [6]

Tonsillar decent (in reference to basion-opistion line)	Classification of CMI
<3 mm	Normal ("cerebellar tonsillarectopia")
3–5 mm	Borderline CMI (can be abnormal if there are co-occurring pathologies, e.g., syringohydromyelia or clinical symptoms)
>5 mm	CMI in patients older than 15 years
>6 mm	CMI in patients younger than 15 years

the disorder can be associated with syringomyelia or syringohydromyelia (SH) in 30 to 70 % of cases [7].

SH is a longitudinally oriented CSF-filled cavity in the spinal cord (Fig. 1). The terminology used when reviewing the MR images defines the specific location of the syrinx. Hydromyelia refers to a dilated central canal, whereas syringomyelia is the cavitation of the cord extending laterally. Syringohydromyelia, involves a combination of both hydromyelia and syringomyelia. In syringobulbia, the cavitation extends to the medulla [8].

In CMI, the presentation and severity of symptoms can change with age and time [9•]. The measure of the cerebellar tonsillar extension has been observed to decrease over time, as a person ages [10]. Cerebellar tonsillar descent can also increase with the presence of intracranial pathological processes such as hydrocephalus, AVM, mass lesion, or hypotension [11•].

CMI is often diagnosed incidentally on MR imaging, after a head injury, accident, infection, or during pregnancy [9•]. Lewis et al. retrospectively reviewed the MRIs of 599 patients with headaches, and 24 (0.04 %) were incidentally found to have CMI [12]. Massimi et al. (2011) acknowledged the high frequency of a temporal association between CMI symptoms and minor head injury [9•]. Remarkably, the severity of the clinical presentation is not always correlated with degree of cerebellar tonsillar ectopia [1, 2]; however, other studies refute this finding [13].

Clinical Manifestations

The clinical symptoms and signs in CMI are diverse. CMI symptoms include head, neck and back pain, cape pain (shoulders), non-radicular limb pain, weakness, paresthesias, vestibular symptoms, diplopia, tinnitus, hearing loss, syncope, slurred speech, dysphagia, urinary incontinence, and sleep disturbance [4•]. Clinical signs include cranial nerve dysfunction (nystagmus, dysphagia, sleep apnea), brainstem compression (syncope, hearing loss, brady or tachycardia), cerebellar signs (ataxia, dysmetria), and spinal cord injury (hyperactive reflexes, spasticity, clonus, incontinence) [4•].

The most frequent symptom in CMI is a very specific posterior occipital or upper cervical headache, described as a sharp or throbbing pain exacerbated by cough, Valsalva maneuver(s), postural changes, or physical exertion [13, 14]. The International Headache Society (IHS) classification ICHD-II code is 7.7 Headache attributed to Chiari malformation type I (Q0.70).

Studies have attempted to clarify headache subtypes in CMI and identify causal relations. Pascual et al. (1992) evaluated 50 patients with CMI and found that 52 % suffered from headaches [13]. Their headaches were evaluated with IHS criteria. Fourteen patients had characteristic CMI posterior occipital headaches, five migraine headaches, six tension type headaches, and one trigeminal neuralgia. The presence of aggravated pain did directly correlate with the degree of cerebellar tonsillar herniation in this study, but not with the degree of bony occipital skull dysplasia. Toldo et al. also showed that in 45 patients <18 years with CMI, headache was the most common presenting symptom with a classic CMI phenotype being the most prevalent headache subtype. Presence of headache with three other clinical signs of CMI was highly predictive of severe tonsillarectopia [15].

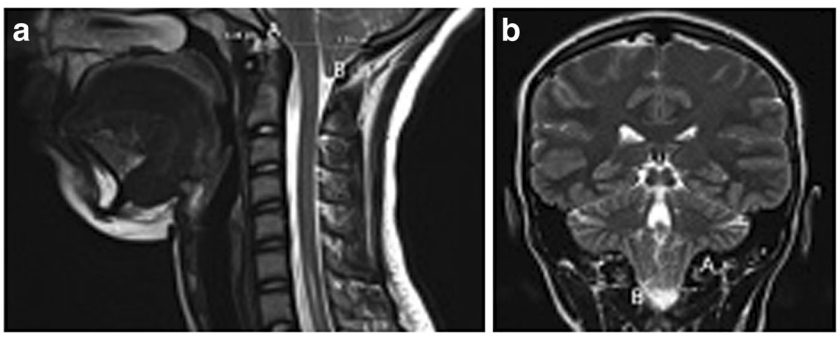

Fig. 1 **a** Sagittal MR measurements of tonsillar descent in a CMI patient. Foraminal line "*A*" spans from clival tip to undersurface of suboccput. Tonsillar line "*B*" is drawn perpendicular from the foraminal line to the caudal tip of the tonsils. Note the associated SH in the caudal cervical cord. **b** Coronal MR measurements of tonsillar descent in a CMI patient. Foraminal line "*A*" is drawn across the foramen magnum, underneath the suboccput. Tonsillar line "*B*" spans perpendicular from the foraminal line to the caudal tonsillar tips

In another study, Wu et al. (1999) evaluated 49 pediatric patients under 14 years old via retrospective chart analysis [16]. The most common symptoms were headache, neck pain, and ataxia. Only three had classic CMI type headaches. The degree of tonsillar herniation varied at 5–23 mm. The severity of the clinical symptoms did not correlate with degree of cerebellar tonsillar herniation. As well, there was no clear association with MRI CSF flow abnormalities and degree of symptoms. However, the sample size was very small.

Pujol et al. (1995) suggested that it is not the size of the cerebellar tonsils but rather the degree of motion of the cerebellar tonsils that determined the presence of cough-strain headache in patients [17]. Six patients presenting with this kind of headache showed a larger tonsillar motion index compared to eight patients showing no cough-strain headache on dynamic CSF flow studies. The study concluded that the "amplitude of the tonsillar pulsation and the severity of the arachnoid space reduction were associated with the symptom of cough-strain headache, but not with the presence of syringomyelia." This theory provokes further questions regarding the etiology of CMI.

Wu et al. [16] observed that "clinical symptoms occur when arachnoidal scarring and adhesions build up at the foramen magnum, possibly because of the cerebellar tonsils rubbing up against bone over many years." It was suggested that arachnoid adhesions increase the compression of the hindbrain, leading to clinical signs and symptoms. They also believe that this may lead to the formation of syringomyelia. This theory is one of many suggested as a cause for CMI.

Pathophysiology

The pathology of CMI involves the cerebellum and how it relates to the posterior fossa. It has been well documented that CMI patients frequently have a small infratentorial to supratentorial space ratio although the volume of brain parenchymal tissue remains the same [9•, 18]. Due to this anatomical discrepancy, patients with CMI who exhibit smaller posterior cranial fossa dimensions have been found to become symptomatic sooner and respond more, favorably to surgical decompression [18]. In addition, the orientation of the clivus may alter the size of the posterior fossa. Platybasia, which can be seen in CMI, is defined as greater than a 143° angle measured from the anterior cranial fossa to the clivus causing abnormal skull base flattening, leading to a decrease in the size of the posterior vault [4•].

CMI can develop due to primary genetic or physiologic causes as well as secondary acquired syndromes leading to physiologic changes and is the only Chiari malformation that can be acquired. There are numerous theories hypothesizing the etiology of CMI. Hydrodynamic theories, describing abnormal CSF pressures in CM, deserve special attention given the posited causal relationship to SH formation. It has been hypothesized that the fourth ventricle and central canal of the spinal cord fail to communicate due to a failure of the fourth ventricle foramen to open in utero. Forces from CSF pulsations create a "water-hammer" effect causing syrinx formation [19].

Other theories propose that an increase in intrathoracic pressure creates a cranial-spinal pressure gradient causing CSF to get "sucked" into the central canal of the cord [20]. It has also been suggested that CSF may enter the canal via perivascular spaces [21] and that cord production of CSF as well as spinal canal stenosis may contribute [22]. Most recently, it was demonstrated intraoperatively that the cerebellar tonsils function like pistons intermittently occluding the subarachnoid space with each beat of the heart [23]. In systole, the cerebellar tonsils descend whereas in diastole they ascend. This finding has been reproduced outside of the operating room with MR CSF flow studies [4•, 24, 25].

SH is commonly seen in adolescents and early adulthood and is believed to be a secondary manifestation of CMI due to the alteration of hydrodynamics as described above. Scoliosis is a common sign observed in patients with CMI and SH. It is questioned whether long standing pressure changes in the spinal cord at different levels may actually create the curvature changes in the spine. According to Godzik et al., in a retrospective review of 92 pediatric patients with CMI and SH, a maximum syrinx diameter (>6 mm) and moderate (5–12 mm) rather than severe (>12 mm) cerebellar tonsillar herniation are significant predictors of spinal deformity, specifically scoliosis [26•]. Secondary causes of CMI must always be excluded, such as hemorrhage, hydrocephalus, AVM, neoplastic processes, and intracranial hypotension, or benign intracranial hypertension [4•, 11•].

Diagnostic Neuroimaging

CMI is traditionally diagnosed by measuring anatomic structures via neuroimaging techniques as described originally by Baker in 1963 [27]. Baker recommended that a line be drawn from the tip of the clivus to the base of the foramen magnum on sagittal MR imaging of the brain (Fig. 1a). A second line is then drawn from the middle of the cerebellar tonsils where they meet line 1, inferiorly at the lowest point of the tonsil. The same method can be used with coronal sequences to evaluate for tonsillar asymmetry (Fig. 1b). The coronal measurement may actually prove to be more informative as to the pathologic changes regarding asymmetries in cerebellar tonsillar compression.

CMI may be suspected initially on a variety of diagnostic imaging modalities. Standard diagnostic imaging protocols and tools are summarized in Table 2.. MR imaging is the gold standard for diagnosis. Revision of diagnostic classification by age has changed over time due to the later discovery that

Table 2 Utility of standard imaging protocols—signs and symptoms chart

Imaging modality	Usefulness
Non-enhanced CT of the head	Identify tonsillar crowding at skull base
Non-enhanced CT of the cervical spine	Identify and measure tonsillar descent
MR imaging of brain with/without contrast	Identify and measure tonsillar descent Exclude presence of intracranial pathology Document other associated anomalies such as platybasia, meningocele, and clival orientation
MR imaging of spinal axis with/without contrast	Identify presence of syringohydromyelia Exclude presence of other skeletal anomalies
Phase-contrast Cine-MR imaging study	Visualize cerebro-spinal fluid flow anterior and posterior to cranial cervical junction as well as flow through the aqueduct
DTI MR imaging	Identify microstructural tissue changes
Plain radiographs of spinal axis	Identify scoliosis and other vertebral anomalies

the cerebellar tonsils ascend with age. This has led to the current accepted diagnostic criteria in Table 1.

The advent of more sophisticated hydrodynamic flow measures, such as phase-contrast MR imaging has allowed for a measure of the severity of compression in the posterior fossa. Studies utilizing this technique have shown that obstruction of the foramen magnum and abnormal CSF flow especially at the C2–3 level leads to impaired systolic and diastolic flow across the craniocervical junction, which may correlate with the presence of SH [24]. Therefore, CSF flow study analysis at this level in particular has proven to be especially informative. Moreover, it is important to evaluate both axial and sagittal views when obtaining CSF flow measurement studies as in CMI, as CSF flow has been shown to increase in the anterolateral subarachnoid space and decrease in the posterolateral subarachnoid space [5, 28, 29, 30•]. MR studies CSF flow are now being used to evaluate CMI patients pre- and post-surgically to predict outcomes, although standardized protocols are not currently available [25, 29, 30•]. More large volume, comprehensive, longitudinal studies are essential to allow for a systematic, standardized approach to evaluating CMI via MRI CSF flow studies [25].

More recently, DTI is being used to evaluate the integrity of the brainstem and cerebellar white matter tracts in patients with CMI. One study, with a pediatric population [31•, 32•], showed microstructural tissue alterations in symptomatic patients, most notably in the middle cerebellar peduncle. This is fascinating as different theories about neuroectodermal abnormalities in the cerebellum in CMI have been proposed in the past. This area of research may prove to be especially informative. In addition, single photon emission computed tomography (SPECT scan) or proton chemical shift imaging (an MR spectroscopy technique) have also been utilized to evaluate similar disorders with pressure-related physiologic changes, such as normal pressure hydrocephalus [33]. Elevated lactate peaks have been found when there is intracranial damage. PET studies may offer another way to look at the cerebellum and brainstem with CMI patients. As we look into the future, these novel imaging techniques could serve as guide in the pre- and post-surgical management of CMI [4•].

Treatment

Both medical and surgical management can be used to treat CMI. After a diagnosis is established, initial medical management may be implemented with the use of pharmacotherapy, physical therapy, and therapeutic injections. Headache is the most common presenting symptom of CMI, but the prototypic CMI type headache is not always present. Typically, patients are treated according to presenting headache phenotype [14].

For individuals who have failed medical management but have a combination of a concerning clinical presentation, neuroimaging and/or physiological studies such as somatosensory evoked potentials, swallow evaluations, and sleep studies, surgical intervention is recommended [34]. The most common surgical intervention is a suboccipital craniectomy with cervical laminectomy with or without duroplasty. Despite surgical intervention, symptoms may persist in some individuals. The most common refractory symptoms include duration of headaches, frontal headaches, and vertigo [35]. In rare cases, revision of decompression surgery is required, which can lead to increased adhesions. These patients may develop intractable headaches that are very challenging to treat. More research is needed in this subpopulation.

Conclusion

CMI remains a complex neurological disorder, associated with a wide array of radiological findings and associated signs and symptoms. The most common symptom is a characteristic CMI type headache. Neuroimaging studies have been an essential tool to diagnose CMI and help to clarify the pathophysiology of this complex disorder. A deeper understanding of the pathophysiology of this condition is necessary to guide in treatment, especially when surgical options are being considered. The utilization of CSF flow MR imaging studies have helped to facilitate a deeper understanding of CSF flow pressure gradients across the cranial cervical junction, thus providing an in vivo way of measuring physiologic processes and defining treatment protocols. Standardized protocols for CSF

flow studies are necessary to allow for more generalized and reliable recommendations. Furthermore, the innovation of DTI as well as MR spectroscopy has shown promise in illustrating the disruption of normal neurological pathways resultant from these compressive forces. These advanced neuroimaging techniques may have future implications in clarification of diagnosis, treatment, and surgical selection and allow for predictions for better treatment outcomes.

More research needs be done to compare pediatric and adult manifestations of CMI, treatment outcomes for CMI headache, treatment of recurrence of headache after decompression surgery and standardization of imaging protocols, such as CSF flow studies.

Compliance with Ethics Guidelines

Conflict of Interest Dr. Jennifer Williams McVige and Dr. Jody Leonardo each declare no potential conflicts of interest.

Human and Animal Rights and Informed Consent This article does not contain any studies with human or animal subjects performed by any of the authors.

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