

Evaluation and management of the Chiari malformation type 1 for the primary care pediatrician

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The Chiari malformation type 1 (CM1) has been subject to a recent re-examination anatomically and clinically [1,2]. Although the hallmark of the clinical syndrome has been tussive head pain, recent studies have begun to list a much longer collection of potential symptoms and signs [1,3–6]. These symptoms relate to pain or other sensory changes in the head, neck, extremities, and body as well as a variety of changes in brain stem function [6–18]. Similarly, the classic neuroradiologic description of CM1 has been the descent of the cerebellar tonsils to a level more than 5 mm below the foramen magnum. This definition is also in the process of being revised [2]. There are a variety of reasons for this re-examination of the clinical Chiari syndrome (encompassing the potential presenting symptoms) and the anatomic malformation in CM1, most important of which is a heightened awareness of the potential value of decompression in selected cases of Chiari malformation where symptoms are not entirely classic in nature [19]. For the purposes of this discussion, the clinical Chiari syndrome and the anatomic Chiari malformation are considered as two separate entities.

In children, particularly young children, definitions based on millimeters of herniation are not particularly useful, given the variability of the distance between the foramen magnum and the arches of C1 and C2 and the variable size of the foramen magnum [20,21]. Therefore, herniation has generally been referred to in terms of segments (ie, between the foramen magnum and C1 or beyond C1). The clinical Chiari syndrome can be divided into symptoms that are cardinal to the disease and generally referred to as classic in presentation (such as tussive headaches and syncope) and symptoms that have recently been recognized as

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secondary manifestations of the syndrome (such as tinnitus or cognitive changes) [3,7,22–26]. The latter can often be debilitating and potentially alleviated by surgical decompression.

The emphasis of this article is on the diagnosis of the anatomic Chiari malformation and the clinical Chiari syndrome and the significance of the presence of herniation in the evaluation of symptomatic and asymptomatic children. Referral to a neurologist or a neurosurgeon should occur early in the process of evaluation, because many entities present with some sort of head pain. In addition, medical treatment of the Chiari syndrome and surgical decompression and potential postoperative complications are discussed briefly.

The anatomic Chiari malformation type 1

Classically, neuroradiologic texts define CM1 as herniation of the cerebellar tonsils 5 mm or more below the level of the foramen magnum without the necessity for additional or associated brain and posterior fossa anatomic abnormalities (Fig. 1) [27,28]. This definition is based on contrast-enhanced radio-

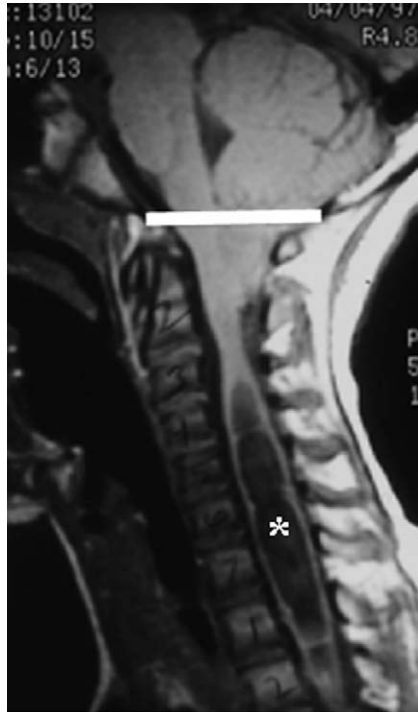


Fig. 1. Sagittal MR scan of a patient with Chiari malformation type 1. Cerebellar tonsils are displaced through the foramen magnum (*white bar*) to the lower aspect of C2 with clear crowding at the foramen. A syrinx (*white asterisk*) is visible extending from C3 to T2.

graphic studies of the craniocervical junction. With the advent of MRI, the ability to image the posterior fossa and the craniocervical junction has increased remarkably in quality and sensitivity [21,29–31]. This technology has necessitated a re-examination of the definition of anatomic CM1, because herniation of less than 5 mm can be accompanied by classic Chiari symptomatology [1]. In addition, because of the potential screening nature of MRI, herniation far beyond the 5 mm of the classically described CM1 can be seen without symptoms [2]. A number of studies have begun to address issues of variations in the anatomy of the posterior fossa, which either accompany the anatomic CM1 or are seen with herniation of cerebellar tonsils below the level of the foramen magnum without the presence of an anatomic CM1 [21,32–34]. Some investigators have begun to define the degree of herniation by anatomic levels (eg, herniation from the foramen to C1, C1 to C2, or beyond C2). Such categorization is more applicable in the pediatric population because of the variable distances in those structures [20]. Because intervention on the Chiari malformation is based more on symptomatology than on structural characteristics, the exact anatomic definition beyond the requirement of tonsillar herniation is not always germane to the specific choice of treatment.

Other anatomic associations with CM1 are syrinx and myelodysplasia, which manifests as tethered spinal cord [35,36]. These entities are also recognized by MRI and can anatomically define reasons for intervention. CM1, often associated with the syrinx, does not necessarily require the presence of syrinx to have symptomatic presentation [1,37]. Conversely syringomyelia can also occur in the absence of CM1 [38,39]. Some studies have used posterior fossa decompression as a treatment for syrinx in the absence of Chiari malformation but only under specialized circumstances [40,41]. There is an association of myelodysplasia with CM1, but this association has not been well defined and generally does not affect the treatment [42].

In summarizing the current anatomic re-examination of the Chiari malformation, it is necessary to remember that lack of tonsillar herniation below the foramen magnum almost always rules out presence of an anatomic Chiari malformation even in the presence of a compelling clinical Chiari syndrome. Without the anatomic malformation a diagnosis of Chiari syndrome cannot be made, and referral to a neurologist or a neurosurgeon is warranted. In these cases, surgical decompression is rarely performed except in unusual circumstances at pediatric neurosurgical centers experienced in the care of posterior fossa and hindbrain anomalies.

Clinical Chiari syndrome

Classically, the Chiari syndrome consisted of tussive occipital head pain that was relatively short lived, generally lasting from a few seconds to a few minutes [4,8,13,43–48]. This pain could also be accompanied by posterior neck pain running down the shoulders. Generalized pain from the occiput to the frontal area

is often associated with the headaches as well as pain or paresthesia that runs down the upper extremities [49,50]. A number of other symptoms can be associated with the Chiari syndrome, including dropping attacks and apneic spells. Typically a short exertional or tussive fainting spell is described in which the child initially feels faint and actually loses consciousness for a few seconds before awakening in a relaxed state on the ground [9,51]. Apneic spells and aspiration pneumonia have also been associated with CM1 and are generally described as occurring at times of exertion [52–58]. When a syrinx is present, CM1 patients complain of pain or numbness in a bandlike distribution on the back as well as leg weakness [59–62]. Syrinx is sometimes associated with scoliosis, but cases of CM1 with scoliosis and absent syrinx have also been reported [61,63–68].

These symptoms make up the general list of cardinal Chiari symptoms and signs. In the recent past a list of secondary Chiari symptoms and signs has been constructed [1]. Patients with an anatomic Chiari malformation can present with a combination of classic and secondary symptoms or with only secondary symptoms [3–5,11,14,24,26,69–80]. Secondary symptoms include almost any symptom that can be ascribed to the brain stem or craniocervical junction area along with pain or paresthesia anywhere from the crown of the head to the feet. They include tinnitus, facial pain and numbness, neck pain or numbness, shoulder pain or spasm, numbness and tingling in the hands, a feeling of whole-body weakness or heaviness, dysphagia or irritation in the throat, chest pain in a bandlike fashion around the chest, poor balance or tripping, speech delay in a young child, constipation, balance difficulties including a positive Romberg's test, weakness or hemiparesis to face or arm, or leg, and other symptoms, as listed in Table 1.

Because the list of symptoms included in the clinical Chiari syndrome or found in association with Chiari malformation is quite long, almost any constellation of symptomatology could potentially be included in a Chiari syndrome. Often in discussing potential surgical outcome with patients, the

Table 1
Primary and secondary symptoms and signs in Chiari malformation type 1

Primary symptoms and signs	Secondary symptoms and signs
Tussive headache [5]	Papilledema [78,114]
Visual changes [115,116]	Absent gag reflex (cranial nerve IX) [5]
Syncope [9,10,51,117]	Hoarseness (cranial nerve X) [79]
Muscle weakness	Tinnitus [14,22,24]
Sensory deficits	Hyperreflexia [5]
Dysphagia [71,118–120]	Hyporeflexia [5]
Dysarthria	Babinski's sign
Ataxia [22,58]	Positive Romberg's test
	Nystagmus (vertical and downbeating) [121–123]
	Speech delay [25]
	Seizures [26,124,125]
	Tremor [126]

neurosurgeon can offer that after surgical decompression the primary symptoms are likely to improve and secondary symptoms may improve if they are indeed related or may persist if they are unrelated. Because there are no specific diagnostic tests that can connect the anatomic Chiari malformation to a specific Chiari syndrome symptom, the surgical decompression can be as much diagnostic as therapeutic. In the absence of any of the primary Chiari symptoms, a constellation of secondary symptoms can only be ascribed to the Chiari syndrome in a cautious fashion and generally by a neurosurgeon or neurologist who is familiar and comfortable with treating patients who have symptomatic Chiari malformation.

Evaluation of patients with Chiari malformation

Generally, the presenting symptom of Chiari malformation should include head pain of some kind [47]. In addition, some sort of neurologic deficit is common. Occasionally, scoliosis or other symptoms related to spinal cord dysfunction can be the presenting complaint of a Chiari patient [62,64,65,68, 81–83]. There is, however, a long list of much more common potential causes for headache in children, including migraine, headache pain from sinus disease, posttraumatic headaches, and a variety of other common agents [4]. More worrisome entities, such as brain tumor, can also cause head pain and are a source of concern for patients, family members, and primary physicians. The more common causes of headache should be investigated before evaluation for Chiari malformation is entertained. Today, consistent headache without a clear cause has become a relative indication for brain imaging with the MR scanner. Thus a complaint of consistent headache or any other neurologic deficit will lead to evaluation by a subspecialist such as a neurologist or to an imaging study before a referral is made [84]. The diagnostic method of choice for Chiari malformation is, of course, the MR scan in which the tonsillar herniation is best seen on sagittal imaging [27,29,30]. Once an abnormality is recognized on the MR scan, referral to a neurologist or a neurosurgeon is obligatory. Even for an asymptomatic patient in whom the Chiari malformation is found incidentally (after trauma or for the diagnosis of headaches that have spontaneously resolved), family reassurance is probably best given by a neurologist or neurosurgeon familiar in treating this disease [2,85–87]. Unfortunately, other than the MR scan, there is no noninvasive diagnostic imaging study that can connect a clinical syndrome with the anatomic Chiari malformation.

In addition to brain imaging, once a Chiari malformation is noted anatomically, the authors recommend a spine survey MR scan [65]. This scan is performed to evaluate for syrinx, which occurs most commonly in the cervical spinal cord in association with Chiari but can occur in the thoracic or lumbar spinal cord as well [68,88–90]. CM1 has been associated with occult lumbar myelodysplastic lesions [42]. This association would justify imaging of the lumbosacral spine. In addition to MR imaging of the entire neural axis, careful

neurologic examination and history are required to elicit evidence for any other secondary Chiari symptoms. Hyperreflexia, loss of pinprick sensation on the back of the neck or on the shoulders, and loss of brain stem reflexes such as the gag reflex are common in the presence of anatomic Chiari malformation but are seldom noticed by the patient or the patient's family [91]. Once the clinical and radiologic evaluation is complete and has provided proof of anatomic Chiari malformation, a therapeutic plan is necessary and should be formulated by a neurologist or neurosurgeon familiar with treating the disorder.

Treatment of patients with Chiari malformation type 1

As in all neurosurgical disease, the first line of treatment, if possible, is generally medical. In the authors' practice, absolute indications for surgical intervention on a patient with Chiari malformation include the presence of a syrinx (particularly if the syrinx causes widening of the spinal cord), the presence of syncope that are documented and are tussive or exertional in nature, the presence of apnea or documentation of apneic spells, and the presence of a swallowing disorder causing aspiration or a history of aspiration pneumonia [19,28,57,58,92–96]. These symptoms of the disease carry potential life-threatening consequences, and the authors strongly recommend surgical decompression without delay. Presentations with these absolute indications are relatively rare, however, and patients generally present with head pain and other sensory symptoms of varying intensities. Thus the treatment decision depends on the subjective impact of the symptoms on the life or life-style of the patient.

Before embarking on surgical treatment of the Chiari pain syndrome, the authors have generally recommended a trial of medical therapy through an interdisciplinary pain service or a neurologist comfortable with treating head and body pain. Such a regimen might include neuropathic pain medicines in the family of anticonvulsants or in the family of the heterocyclic antidepressant agents. Often medical management of the head pain can reduce the pain adequately to avert surgery. Alternatively, medical management can put off surgery for some time, allowing ongoing discussion and better understanding of the impact of the symptoms on life and lifestyle. In children, there is certainly the argument that if years of chronic medical treatment are going to be required and if surgical therapy might achieve a condition that would require minimal or no medicines, considering surgical intervention might be reasonable [97].

The decision to proceed with surgical intervention on the Chiari malformation then relies on the presence of one of the symptoms or signs that are considered an absolute indication, or on a careful and reasoned decision based on the impact of head pain on life and lifestyle, or on sensory symptoms that are refractory to medical management [92–94]. In such circumstances it is reasonable to entertain surgical intervention.

The specifics of surgical intervention for Chiari malformation are currently controversial in the neurosurgical literature because the pathophysiology of

Chiari syndrome is still vigorously debated. The surgical approach varies depending on the theoretic approach to that pathophysiology. For surgeons who believe that Chiari symptomatology is brought on by a lack of cerebrospinal fluid (CSF) flow through the foramen magnum because of crowding of the tonsils, restoration of foramen magnum CSF flow becomes the goal of surgical intervention [98–103]. This intervention might include bony removal at the foramen magnum by limited suboccipital craniectomy and removal of the laminar arch of C1 [95]. Many surgeons continue the approach of enlarging the CSF space at the craniocervical junction by opening the dura and constructing a duraplasty from autologous pericranium, another autologous material, or a foreign body material [104]. For surgeons who believe that the pathophysiologic mechanism of Chiari malformation stems from a congenital inadequate size of the posterior fossa, the approach entails enlarging the posterior fossa in an operation that may remove much or all of the bone in the suboccipital region up to and potentially including the transverse sinus and torcular area [1,33,34]. This approach has not been widely used. A matter of ongoing debate Chiari surgery is whether removing bony compression only, without dural reconstruction, is as effective as performing duraplasty [94,105,106]. Some data suggest a more reliable outcome in size reduction and even disappearance of an existing syrinx is achieved when duroplasty is performed [105,107,108]. In addition, the choice of duraplasty material has also been debated. Artificial materials have the potential for causing foreign body reactions that are a concern in the context of Chiari decompression [109,110]. Despite these controversies, the authors' general recommendation is that surgery be performed by a neurosurgeon familiar with the nuances of the procedure.

One additional subset of Chiari patients deserves special consideration: patients presenting with a clear anatomic Chiari malformation but no demonstrable clinical syndrome [2]. The authors do not advocate prophylactic decompression for these patients, because neither the natural history of the disease nor the prevalence of asymptomatic CM1 is known. Asymptomatic patients should be followed expectantly, and in the authors' practice very few of these patients have progressed to significant symptomatology. The reasons for this lack of progression remain elusive, but in practical terms the authors recommended a yearly follow-up that includes questions regarding historical changes consistent with Chiari symptomatology and a neurologic examination. In patients without syrinx the authors do not advocate repeating the MR scan unless symptoms develop.

Postoperative complications after Chiari surgery and general results

Chiari decompression remains a low-risk surgical procedure. The general risks are those of any neurosurgical procedure, including problems with wound healing and infection as well as muscle spasm and pain in the suboccipital region. If the dura is opened, there are the risks of CSF leakage and meningitis, which can be either infectious or chemical. The biggest risk associated with Chiari surgery, of

course, is that the surgery may not resolve the symptoms. Symptoms may remain unchanged, or relief of symptoms may be transient. Results of Chiari decompression using a variety of neurosurgical approaches have described with resolution of Chiari symptoms ranging from 60% to 100% [19,41,58,59, 94,108,111–113]. In the adult population, the percentage of patients experiencing complete symptomatic resolution after surgery may be somewhat lower [1,28].

The group of children for whom Chiari surgery fails to relieve symptoms is relatively small but is very difficult to manage. Symptoms of pain and neurologic deficits can persist or worsen despite MR scans showing significant decompression with intact or even capacious subarachnoid spaces. At that point the authors have recommended ongoing medical management for several months. If symptoms persist beyond 3 months after the surgery, the authors have repeated a craniocervical MR scan with cinematic CSF flow at the foramen magnum to ascertain whether an occlusive or scarring process has again blocked CSF flow at the foramen magnum. If CSF flow is compromised, the authors have recommended surgical re-exploration, because they have usually found inadequate initial decompression or a scarring process. If flow is present at the foramen magnum, the authors have examined the CSF by lumbar puncture, measuring pressure and examining the CSF for pleocytosis or other signs of inflammation. The authors have encountered several pediatric patients with this scarring process who have responded to enlarged decompression or resection of the tonsils to allow free access of fluid in and out of the fourth ventricle. They have also encountered a few patients who have developed postoperative elevated CSF pressures consistent with a post-Chiari pseudotumor syndrome. Some of these patients have progressed to requiring CSF shunting devices from the ventricular or lumbar space. The treatment of the patients for whom Chiari surgery has failed is challenging in many ways.

General instructions for the postoperative Chiari patient include restrictions on maximum weight to be lifted for up to 3 months postoperatively and a graduated return to activity. The authors restrict children from any strenuous activity for 2 to 4 weeks and recommend restriction from any contact sports for 3 months. Beyond that time they slowly release all restrictions, depending on relief of symptoms and patient tolerance of activity. Beyond the 3-month mark, the questions most frequently asked by parents involve sports activities and riding on roller coasters or other similar activities. The authors are unaware of any data suggesting that these activities pose a greater risk of injury to a child Chiari decompression than to the general population. On first principles, the operation is not destabilizing to the bony structures of the cranial–cervical junction and should not appreciably weaken the soft tissue structures. That being said, there is anecdotal experience with head and neck injury in the postoperative Chiari patient that has led to recapitulation of nearly all the initial presenting symptoms. The reason for this phenomenon is not understood, because, in the authors' experience, the radiographic evaluation and the physical examination are usually unrevealing. Perhaps one reasonable approach to these questions is to present to

both parent and child the paucity of data regarding risk of injury after Chiari surgery as well as the caution of the anecdotal experience. In this fashion, physician and family can construct a joint recommendation guided by available data and the patient's and family's needs and concerns.

Overall, 90% to 95% of children under age 16 years who require surgical intervention for Chiari are discharged from follow-up because of complete resolution or near resolution of symptoms by 1 year after surgery.

Summary

Diagnosis and treatment of CM1 is undergoing reexamination that includes redefinition of the anatomic Chiari malformation and refinement and redefinition of the clinical syndrome. Children with SM1 present with head pain of some kind, a neurologic deficit, or with signs of spinal cord dysfunction from syrinx. Some will present with no clinical syndrome at all. Presence of anatomic Chiari malformation or compelling clinical Chiari syndrome should lead to evaluation by a neurologist or neurosurgeon experienced with the syndromes and their treatment. Treatment options are varied but usually result in resolution of symptoms. When symptoms persist after surgery, management is complex and not uniformly successful, even in the most experienced hands.

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