REVIEW ARTICLE

Allan H. Ropper, M.D., *Editor*

# Congenital and Acquired Chiari Syndrome

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CME

Chiari malformation, an anomaly of the posterior cranial fossa, also known as Arnold–Chiari malformation, was described in autopsies by Hans Chiari, an Austrian pathologist, in 1891. He defined four distinct anatomical configurations in which the cerebellar tonsils (the most caudal part of the cerebellum) protruded below the lower margin of the foramen magnum.[1] Several subtypes have subsequently been described, often in association with other anomalies, but the essential feature remains the descent of the cerebellar tonsils below their normal position just above the rim of the posterior fossa.

## CLASSIFICATION

Although many characteristics of Chiari malformation are similar in children and adults, this review focuses on the most common type, adult Chiari malformation type 1 (CM1), in which only the cerebellar tonsils descend below the foramen magnum. This malformation can be due to a congenitally small posterior fossa, or it can be acquired as a result of high pressure from above the cerebellum or low pressure below the cerebellum that displaces the tonsils. The next most common Chiari malformation is type 2, which is characterized by descent of the cerebellar vermis and fourth ventricle below the foramen magnum, usually accompanied by an open myelomeningocele. This malformation was recently reviewed in the *Journal*.[2]

Chiari malformation types 3 (occipital encephalocele), 4 (cerebellar hypoplasia), and 5 (cerebellar hypoplasia and occipital-lobe ectopia into the posterior fossa) are seen in children and newborns and are more rare and more severe than CM1. Types 2, 3, 4, and 5, as well as a type that is associated with craniosynostosis (premature closure of cranial sutures), are congenital syndromes in children.[3-5]

Congenital CM1 occurs in patients with a constitutionally small posterior fossa.[6,7] In adults, symptoms from CM1 most commonly develop before the age of 45 years (mean age, 40 years), with an approximate female:male ratio of 3:1, whereas in pediatric CM1, the mean age at presentation is 8 years, with an approximately equal female:male ratio.[3,8-12]

As the cerebellar tonsils protrude below the foramen magnum, obstruction of the foramina of Luschka and Magendie of the lower medulla and compression of the upper cervical spine may occur, impeding the flow of cerebrospinal fluid (CSF) from the fourth ventricle to the upper spinal subarachnoid space. In some instances, this is accompanied by a syrinx (cystic cavity), which is most commonly located in the cervical cord but can involve any part of the spinal cord and rarely the brainstem, as described below. Herniation of the cerebellar tonsils to at least 5 mm below the foramen magnum is the usual definition of CM1 (Fig. 1). However, some symptomatic patients have smaller herniations, and some asymptomatic patients have larger herniations.[9,13] A study involving 2000 asymptomatic adults showed a prevalence of 0.9% for tonsillar descent that was at least 5 mm below the foramen magnum on magnetic resonance imaging (MRI), a finding that is consis-

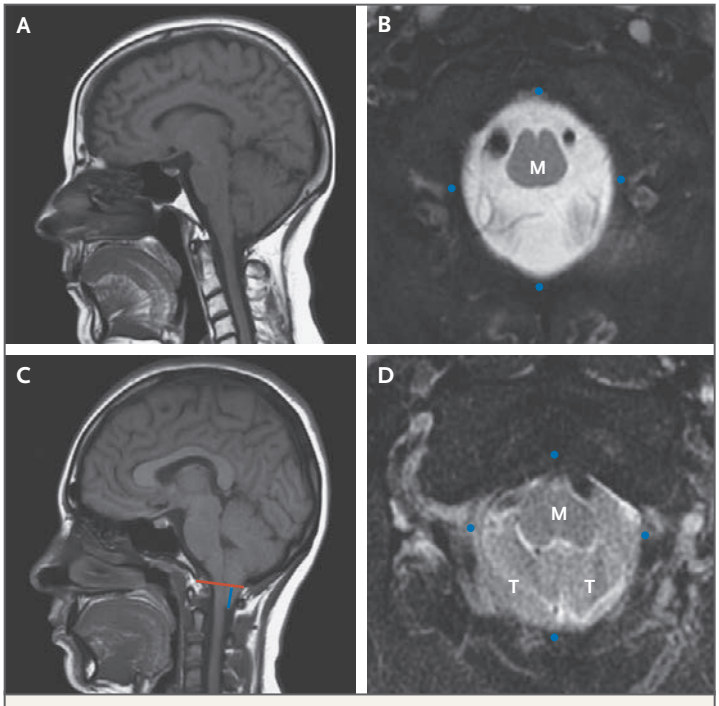


**Figure 1. Features of CM1 on Magnetic Resonance Imaging.**

A normal sagittal T1 image (Panel A) shows that the cerebellar tonsils are above the plane of the foramen magnum. A normal axial T2-weighted image (Panel B) shows the foramen magnum (outlined by blue dots) and the lower medulla (M). Ample space for cerebrospinal fluid (CSF; white signal surrounding the medulla) is present. A sagittal T1-weighted image of a typical patient with Chiari malformation type 1 (CM1) (Panel C) shows cerebellar tonsillar herniation below the foramen magnum, as indicated by the red line (McRae's line). The blue line, perpendicular to McRae's line and extending to the tip of the tonsil, is used to determine the magnitude of tonsillar herniation. An axial T2-weighted image of a patient with CM1 (Panel D) shows the tonsillar tissue (T) within the foramen magnum (outlined by blue dots) and compression of the lower medulla (M). Obliterated CSF space at the foramen magnum is indicated by the virtual absence of white signal.

tent with the anatomical definition of CM1.[14] The term Chiari syndrome refers to the manifestation of symptoms attributable to the malformation (Table 1).[9,16,17]

Although most often CM1 occurs as a sporadic condition, with a low familial prevalence, tentative evidence of a genetic cause has been reported in a few cases, with many genes implicated — none strongly — and no candidate gene offering a unifying explanation for the malformation.[16,18] An association has also been found between CM1 and connective-tissue disorders such as the Ehlers–Danlos syndrome.[19] In one study, the prevalence of connective-tissue disorders with CM1 was 12.7%. However, surgical treatment of the malformation (see below) had failed in many of the patients in this series, creating a potential referral bias that may have led to overestimation of the prevalence because the rate of surgical failure is increased among patients with connective-tissue disorders.[20]

## PATHOPHYSIOLOGY

As mentioned above, low-lying cerebellar tonsils can result in obstruction of normal CSF outflow of the fourth ventricle.[21,22] It has been suggested that this obstruction leads to dissociation of normal CSF pulsatile flow between the cranial and spinal compartments, which is likely to play a role in the headache that often accompanies the malformation and in the formation of a CSF-filled cavity (syrinx). The other main symptoms of CM1 are due more directly to downward tonsillar herniation through the foramen magnum and pressure on the lower medulla and upper cervical spinal cord, as described below. In some cases, particularly those with less than 5 mm of tonsillar descent, partial obstruction of CSF flow occurs posteriorly at the fourth ventricular CSF outlets as a result of arachnoid webs, tumors, or an ectatic posterior inferior cerebellar artery.

Although a congenitally small posterior fossa is the most common cause of cerebellar tonsillar herniation, acquired cerebellar descent can result from pressure gradients created by a pulling effect from below or a pushing effect from above. Pull from below the foramen magnum is the result of spinal CSF leakage, a CSF–venous fistula, or a tethered cord.[23,24] Push from above the foramen magnum can result from hydrocephalus, subdural collections, a brain tumor, or an arachnoid cyst, with a tumor or cyst particularly implicated when it is large.[9,13,17,25,26]

## SIGNS AND SYMPTOMS OF CHIARI MALFORMATION

Symptoms attributable to descent of the cerebellar tonsils into the foramen magnum vary. The most common symptom is headache, which is most likely due to transiently raised intracranial pressure from partial blockage of CSF flow. The headache is usually suboccipital and dull or throbbing, but it can be located anywhere in the cranium and have other characteristics, including migraine and tensionlike discomfort. Chiari

headaches are characteristically exacerbated by Valsalva-like activities such as coughing, laughing, sneezing, straining, lifting heavy objects, and changing body or head positions.

An additional symptom, which is independent of but often associated with headaches, is paresthesia in one or both hands, often involving all the fingers (without a radicular distribution). This symptom is probably related to compression of the dorsal medulla or dorsal columns of the upper cervical cord. Similar symptoms can occur in the feet, although this location is less common. Other, less common but characteristic signs and symptoms are sleep apnea; tinnitus, which may be pulsatile; hypoacusis or hyperacusis; difficulty swallowing; several forms of nystagmus, including downbeat nystagmus (caused by only a few other disorders, such as lithium toxicity and tumors at the cervicomedullary junction); upgaze paresis; oscillopsia; blurry or double vision; and dizziness.[8,9,27] Neurocognitive and psychiatric symptoms have been detected by means of formal testing in patients with CM1, although a direct cause and effect have not been identified.[28,29] It is unclear, for example, whether cognitive and psychiatric symptoms result from chronic pain, but some investigators consider the symptoms to be due to impaired cerebellar circuitry involved in cognition.[30] Chronic fatigue syndrome has been proposed as a CM1 symptom, although the association has not been convincingly shown.[31]

The spinal cord syrinx may be symptomatic or asymptomatic, with a large syrinx more likely to be symptomatic. Patients with a syrinx may present with complex symptoms that are independent of those directly caused by the tonsillar herniation — typically numbness or decreased pain or temperature sensitivity in the hands and over the shoulders, hand weakness and atrophy, and arm, leg, neck, or back pain, as well as bladder or bowel incontinence. In patients with positional headaches that are more severe when the patients are standing and less severe when they are lying down, the diagnosis of spinal CSF leak and intracranial hypotension as an acquired cause of tonsillar herniation should be considered.

**Table 1. Imaging Features of CM1 and Signs and Symptoms of the Chiari Syndrome (Symptomatic CM1).***

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| **Imaging features of CM1** |
| Cerebellar tonsillar herniation below the foramen magnum; ≥5 mm, or 3–5 mm in the presence of a syrinx or peglike tonsillar shape (<3 mm considered normal) |
| **Signs and symptoms of the Chiari syndrome** |
| Headache (typically occipital or suboccipital), Valsalva-associated headache |
| Numbness or weakness in hands, feet, or both |
| Oscillopsia (oscillating objects or blurry vision) |
| Hearing problems (ear pressure, hyperacusia, or hypoacusia), tinnitus |
| Balance problems (ataxia or dizziness) |
| Nystagmus, particularly downbeating |
| Sleep apnea |
| Swallowing difficulty and various cranial-nerve palsies |

* Information adapted from Massimi et al.,[5] Ciaramitaro et al.,[9] and Kumar et al.[15] CM1 denotes Chiari malformation type 1.

## IMAGING

Although tonsillar descent can be identified by computed tomographic (CT) scanning, Chiari malformations are better visualized with brain MRI, particularly sagittal images, and with cervical spine MRI to detect a syrinx. As mentioned above, a cerebellar tonsillar descent of at least 5 mm below the foramen magnum is required to meet imaging criteria for CM1.[13,17,27] The tonsils are often also pointed (i.e., peglike), instead of normally rounded. Although this finding is not required for the diagnosis, it indicates tonsillar compression and shaping by the dorsal bony structures of the posterior fossa. In one consensus report that broadly reflects clinical practice, cerebellar tonsillar herniation to between 3 and 5 mm below the foramen magnum has also been classified as CM1 in the presence of a syrinx or peglike shape of the tonsils.[9] This measurement is obtained on a midsagittal MRI scan by drawing a line from basion to opisthion (the anterior and posterior bone limits, respectively, of the foramen magnum, McRea's line) and drawing a perpendicular line down to the lowest aspect of the cerebellar tonsils, the length of the latter representing the magnitude of descent of the tonsils. An additional consideration in affirming the diagnosis is the degree of "crowding" in the posterior fossa (reduction in the space for CSF to flow) at the level of the foramen magnum on a T2-weighted axial image (Fig. 1).

Cardiac-gated CSF flow imaging (cine flow MRI), a technique used to evaluate CSF flow at the level of the fourth ventricle and the foramen magnum, normally shows bidirectional CSF flow

with systole and diastole. Compromised CSF dynamics on cine flow MRI is characteristic of symptomatic CM1 and has been predictive of abatement of symptoms after surgery.[22] An abnormal cine flow study is not required for diagnosis but provides supportive evidence of symptomatic CM1, with 76% sensitivity and 62% specificity in one series.[32] Increased cardiac cycle–associated motion of the tonsils has also been shown on MRI imaging in patients with symptomatic CM1, but its role in diagnosis has not been established. This abnormal type of tonsillar motion is reduced after surgical decompression of the posterior fossa.[33]

An imaging feature often found in patients with CM1 is compression of the pituitary gland from above, resulting in CSF within the sella turcica, referred to as empty sella. This finding is consistent with increased intracranial pressure. It is more commonly associated with idiopathic intracranial hypertension (pseudotumor cerebri), and its presence can aid in determining that the Chiari malformation is likely to be acquired from raised intracranial pressure rather than congenital.[34]

A syrinx develops within the upper spinal cord in 27 to 66% of patients with CM1. The syrinx is located in the middle of the spinal cord within the central canal (hydromyelia) and is lined by normal ependymal cells or is located eccentrically in the spinal cord (syringomyelia) and is not lined by ependyma.[10,35] A syrinx in the cervical spinal cord can extend caudally throughout the cord or rostrally into the brain stem.

Some patients have compression of the ventral lower brain stem or kinking of the brain stem as a result of the downward displacement of the cerebellum (classified as Chiari malformation type 1.5), which is addressed with a complex diagnostic approach (dynamic plain films and a CT scan) and special surgical procedures. Depending on the degree of ventral brain-stem compression and occipital–cervical mechanical instability, ventral odontoid decompression and occipital–cervical fusion may be performed in addition to standard decompression of the posterior fossa.[9,36,37] The proximate causes of ventral compression include basilar invagination (superior translation of the top of the C2 vertebra into the foramen magnum), a retroflexed odontoid process (posterior angulation of the odontoid process causing brain-stem compression), platybasia (abnormal angulation of the skull base and spine), and craniocervical instability.

As noted above, acquired cerebellar tonsillar herniation can also result from a spinal CSF leak as the contents of the posterior fossa are pulled down into the upper spinal canal because of the pressure gradient created by the leak. In such cases, cranial and spinal MRI often shows dural enhancement after the administration of gadolinium, a finding consistent with intracranial hypotension.[24,38] A spinal MRI is performed to identify the presence and site of the CSF leak, but both are challenging to detect, and more elaborate studies, such as injection of nonionic iodinated contrast medium into the CSF space, may be needed. Another cause of reduced CSF pressure is a direct CSF–venous fistula (drainage of spinal CSF into a vein), which results in secondary downward tonsillar herniation.[23,24]

## TREATMENT

The twofold goal of treatment is to relieve compression of the cervicomedullary junction and normalize CSF flow at the foramen magnum. Validated medical treatments that will accomplish this are lacking. Asymptomatic patients, in whom CM1 may have been detected incidentally on imaging performed for various reasons, do not require surgery if there is no syrinx. The Congress of Neurological Surgeons has issued guidelines generated from a systematic review that concluded that decompression of the posterior fossa leads to a decrease in symptoms of Chiari malformation.[39] However, the consensus guidelines also indicated that evidence to support any specific surgical approach was insufficient.[9]

The management of asymptomatic CM1 in patients with a syrinx is controversial. Guidelines suggest close follow-up of such patients, with surgery recommended if the syrinx expands or associated symptoms appear.[5,9]

Surgical procedures to decompress the posterior fossa vary in complexity and extent and include removing the lower aspect of the occipital bone in the posterior fossa; widening the opening of the foramen magnum and removing the posterior arch of C1; opening the dura and then incising the arachnoid, which can tether the tonsils inferiorly, to release the tissue; resecting

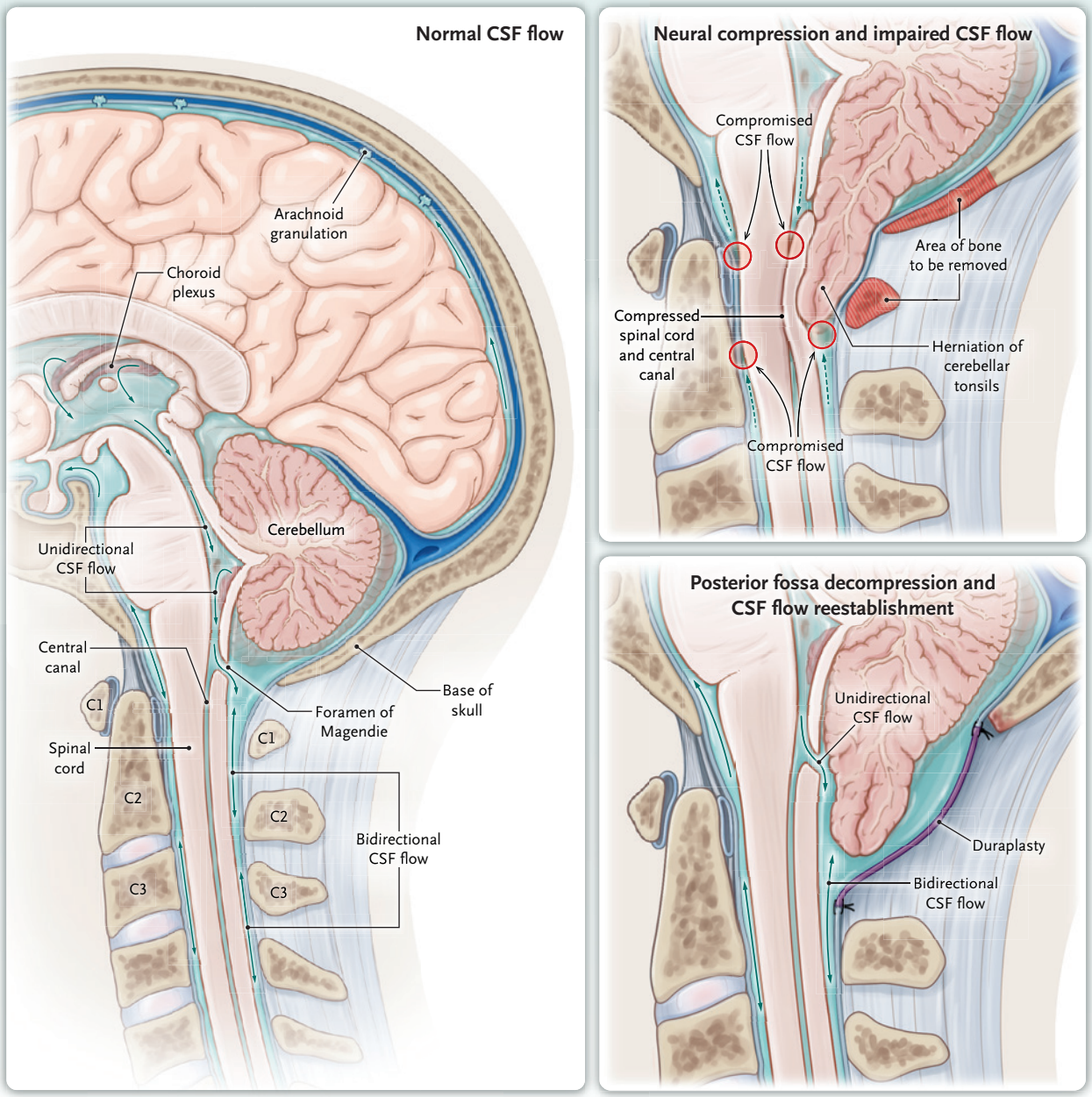


**Figure 2. Surgical Goal of Posterior Fossa Decompression and Reestablishment of CSF Flow.**

In normal CSF flow (left panel), CSF is produced by the choroid plexus (located in the lateral, third, and fourth ventricles) and flows essentially unidirectionally through the ventricles. In the caudal aspect of the fourth ventricle, CSF exits from the foramen of Magendie (midline) into the cisterna magna and from the foramina of Luschka (lateral) into the pontine cistern, where through bidirectional flow, CSF distributes down the thecal sac, and back up around the cerebral hemispheres and is absorbed by the arachnoid granulations. In CM1, the cerebellar tonsils herniate through the foramen magnum (upper right panel), which compromises CSF flow out from the fourth ventricle and rostral flow through the cisterna magna. In addition, tonsillar herniation results in pressure on the lower medulla and upper cervical cord. The lower aspect of the occipital bone and medial aspect of the posterior arch of C1 (in red) are removed during decompression of the posterior fossa. Following bone decompression and expansile duraplasty, normal CSF flow is reestablished (lower right panel), and pressure on the medulla and cervical cord is eliminated.

arachnoid membranes at the level of the fourth ventricular outlet (which can obstruct CSF flow); and reducing the volume of the tonsils (by electrocautery) or performing a complete tonsillar resection. When the dura is opened, expansion of the subarachnoid space is achieved with a duraplasty graft and closure of the dura in a watertight fashion (Fig. 2). As mentioned above, no surgical procedure has been validated as superior to others, and practice varies across institutions and surgeons. The use of duraplasty is supported by some systematic reviews, especially for patients with CM1 and syringomyelia.[35,40] Preliminary results from a cluster randomized trial comparing posterior fossa decompression with and without duraplasty in children and young adults (ClinicalTrials.gov number, NCT02669836) suggest similar rates of surgical complications and similar clinical outcomes.[41] The overall extent of the intradural procedure is also controversial.[9,40]

Acquired tonsillar herniation due to a CSF leak or a mass is treated by addressing the underlying leak or lesion.[24] Lumbar punctures are relatively contraindicated in patients with congenital CM1 who have not undergone surgery, given the risk that lowered spinal CSF pressure may cause further tonsillar herniation.

## SURGICAL OUTCOMES

Patients often experience immediate relief of limb paresthesia and tinnitus after surgical decompression of CM1. Most of the other symptoms are diminished or eliminated within several weeks to months. For example, abatement of headaches and other symptoms after surgery has been reported in 80% or more of patients in several series.[3,8,29,35,42] The two most common postoperative complications with posterior fossa decompression are foreign-body (chemical) aseptic meningitis from a dural graft and CSF leak, which occur, respectively, in up to 32% and 21% of patients who have undergone surgery.[12,35,42] The risk of chemical meningitis may be associated with the graft type. Rates of chemical meningitis are lower with autografts or allografts than with artificial or bovine grafts.[35,43] CSF leak results from CSF seeping around the graft suture line and is often exaggerated by a coexisting increase in intracranial pressure. After surgery, some patients have a transient or permanent increase in intracranial pressure, including idiopathic intracranial hypertension.[44] In patients with persistent or recurrent symptoms after surgery, a lumbar puncture can help distinguish chemical meningitis from infection and increased intracranial pressure.

Chemical meningitis can be treated with glucocorticoids; infection can be treated with appropriate antibiotic agents and, when indicated, surgical irrigation and débridement; and elevated CSF pressures can be treated with a diuretic agent (i.e., acetazolamide) or may require a ventriculoperitoneal shunt. Patients with a syrinx who have undergone decompression may have abatement of associated symptoms, and regression of the syrinx occurs in approximately 78% of patients.[3] The consensus guidelines indicate that alleviation of symptoms does not correlate with resolution of the syrinx, and it is not clear why some patients, despite having undergone proper decompression, do not have decreases in syrinx-related symptoms or improvements in the findings on imaging studies.

A meta-analysis of surgical outcomes has provided support for greater relief from symptoms after bone decompression and duraplasty (with or without tonsillar shrinkage or resection) than after bone-only decompression. The differential improvement is more evident for patients with syringomyelia. More extensive surgery is associated with higher complication rates.[35,40] A meta-analysis comparing bone decompression and duraplasty with the more extensive procedure of bone decompression, duraplasty and subarachnoid dissection, and tonsillar shrinkage showed similar alleviation of symptoms with the two procedures, but complication rates were higher with the latter procedure.[45]

According to one study involving an international database,[12] 6.8% of patients underwent reoperation because of symptoms that had relapsed or had not been alleviated. These circumstances occur for two main reasons: insufficient decompression from the initial procedure or the development of intradural scarring. Cine flow MRI can be useful in evaluating these problems. Abnormal cine flow in patients who never had improvement suggests insufficient decompression. Abnormal cine flow in patients who initially had improvement and thereafter had symptomatic relapse suggests subarachnoid scarring from the surgery. Patients who undergo bone-only decompression without duraplasty appear to have a

higher rate of symptomatic relapse or lack of improvement leading to reoperation.[46]

## CONCLUSIONS

With the wide availability of cerebral imaging, recognition of Chiari malformations has become common, and determining which patients have symptoms associated with this anatomical abnormality can be challenging. Many patients have atypical symptoms or borderline imaging findings. For these patients, cine flow MRI can be informative.[22] Research is required in order to identify patients who would benefit from surgical decompression, delineate appropriate surgical techniques, and improve our understanding of the association of CM1 with cognitive and psychiatric symptoms.

Disclosure forms provided by the author are available with the full text of this article at NEJM.org.

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