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Management of Chiari I malformations: a paradigm in evolution

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Abstract

Purpose Despite decades of experience and research, the etiology and management of Chiari I malformations (CM-I) continue to raise more questions than answers. Controversy abounds in every aspect of management, including the indications, timing, and type of surgery, as well as clinical and radiographic outcomes. This review aims to outline past experiences, consolidate current evidence, and recommend directions for the future management of the Chiari I malformation.

Methods A review of recent literature on the management of CM-I in pediatric patients is presented, along with our experience in managing 1073 patients who were diagnosed with CM-I over the past two decades (1998–2018) at Children's National Medical Center (CNMC) in Washington DC.

Results The general trend reveals an increase in the diagnosis of CM-I at younger ages with a significant proportion of these being incidental findings (0.5–3.6%) in asymptomatic patients as well as a rise in the number of patients undergoing Chiari posterior fossa decompression surgery (PFD). The type of surgical intervention varies widely. At our institution, 104 (37%) Chiari surgeries were bone-only PFD with/without outer leaf durectomy, whereas 177 (63%) were PFD with duraplasty. We did not find a significant difference in outcomes between the PFD and PFDD groups ($p = 0.59$). An analysis of failures revealed a significant difference between patients who underwent tonsillar coagulation versus those whose tonsils were not manipulated ($p = 0.02$).

Conclusion While the optimal surgical intervention continues to remain elusive, there is a shift away from intradural techniques in favor of a simple, extradural approach (including dural delamination) in pediatric patients due to high rates of clinical and radiographic success, along with a lower complication rate. The efficacy, safety, and necessity of tonsillar manipulation continue to be heavily contested, as evidence increasingly supports the efficacy and safety of less tonsillar manipulation, including our own experience.

Keywords Chiari malformation · Syringomyelia · Long-term outcomes · Decompression · Duraplasty · Tonsillar coagulation · Shunt · Pediatric

Introduction

Despite considerable attention and effort given over the past decades to understanding the clinical continuum of the Chiari type I malformation (CM-I), the etiology and ideal therapeutic approach continue to raise more questions than answers. *Not surprisingly, despite innumerable publications and an abundance of clinical recommendations, there is a paucity of widely believed or acknowledged "well-proven" therapeutic guidelines*. While there may be an infinite number of proposed approaches, there are, however, widely accepted concepts and therapeutic strategies to this problem. It is only a matter of time before we have meaningful answers to the myriad issues now facing the clinician with the patient who presents with tonsillar herniation in the setting of multiple objective as well as subjective complaints. *The purpose of this chapter is to outline where we have come from, what we know today, and ultimately a final destination*.

History

Chiari malformations, a heterogenous group of hindbrain disorders, represent one of the oldest, yet still controversial,

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subjects in neurosurgery. Recognized in 1896 by Hans Chiari, an Austrian pathologist, they are characterized by varying degrees of malformations in the rhombencephalic derivatives. Chiari malformation type I–III involves herniation of hindbrain structures through the foramen magnum while type IV is characterized by cerebellar hypoplasia or aplasia [68]. Historically, these disorders were thought to represent a continuous spectrum of a single disorder; however, they are now known to be discrete anatomic entities with distinct clinicoanatomic features [105]. This may also be true for the more common presentation known as CM-I, and as newer variants of the disorder are identified due to improvements in imaging techniques, the need for an expansion of the traditional classification is becoming increasingly recognized [85]. Additionally, the increased utilization of magnetic resonance imaging (MRI) in the young child has led to the identification of Chiari malformations at a younger age as well as incidental findings, raising further questions about the adequacy of standard conventions and nomenclature [2, 25, 83, 99].

The classic definition of CM-I, as defined by Hans Chiari is "elongation of the tonsils and medial parts of the inferior lobes of the cerebellum into cone-shaped projections, which accompany the medulla oblongata into the spinal canal" [103]. While the pathogenesis is not clearly elucidated, it is thought to arise from an insufficiency of the paraxial mesoderm characterized by an underdevelopment of the posterior fossa resulting in a mismatch between neural "contents" and the bony "container" [71, 74, 109]. The original definition by Chiari based on autopsy findings still holds true; however, CM-I has since evolved into a *radiologic diagnosis* defined as a greater than 5-mm caudal descent of the cerebellar tonsils below the level of the foramen magnum. This cutoff was established in 1986 by Barkovich et al., who compared brain MRI between 200 normal individuals and 25 patients with classic symptoms of CM-I, demonstrating 5 mm as the cutoff seen at 2 standard deviations [1, 12, 70]. However, recent findings of neurologic symptoms and even syringomyelia in lesser degrees of herniation collectively termed "tonsillar ectopia" highlight the subjective nature of this definition [33, 74, 90]. Additionally, the observation that cerebellar tonsils ascend with age challenges the application of this diagnostic criteria in the pediatric population [93, 72]. *Ultimately, it is critical to determine the true etiology of the radiographic finding (tonsillar herniation) if we are to arrive at an appropriate understanding of this condition and potential therapeutic measures*.

Presentation

The clinical presentation for a CM-I is universally variable. It is not uncommon to find a radiographic presentation in an incidental fashion, especially in the setting of a nearly 1% incidence of CM-I in the general population and up to 3.6% in children [2, 25, 70, 99, 115]. Over the past two decades at Children's National Medical Center (CNMC) in Washington DC, 1073 patients were diagnosed with CM-I between 1998 and 2018; 620 cases with radiographic findings of tonsillar herniation greater than 5 mm below the foramen magnum were identified *incidentally*. The average age at diagnosis was 7.6 (0.36–23.6) years, and there were 55% males vs 45% females (Table 1). Initial indications for magnetic resonance imaging (MRI) involved seizures, trauma, nonspecific headache, frontal headache, developmental delay, neurobehavioral, infection, endocrine, micro/macrocephaly, surveillance, research study, and other (Table 2). While these children were followed with serial MRI scans over a number of years, only a small proportion of them (~9%) progressed to develop intractable symptoms, neurological deterioration, or manifestation of a syrinx. Many of these patients had significant radiographic findings of severe crowding of cerebrospinal fluid (CSF) spaces at the craniocervical junction as well as tonsillar descent well below 10 mm and yet remained asymptomatic over decades. The true long-term clinical significance remains unclear, and it may be many years before we have a final answer. It is also known that adults present differently from children and thus raises the possibility of an ongoing clinical continuum versus a completely different etiology (and presumably a different therapeutic regime). We have found that younger patients more often present with neurological (brain stem) symptoms with/without syringomyelia whereas the adult patients most often present with pain (headaches) and generally are comprised of women (*90% in adults* vs *55% in children*) [18]. Symptomatic patients with CM-I often presented with occipital headache, neurological findings, or scoliosis (with and without syringomyelia). Scoliosis, gait abnormality, exertional headache, syrinx, and basilar invagination were significantly different between younger (0–6 years) and older children (11–14 years).

While CM-I is primarily discussed in the context of tonsillar herniation, the predominant concern is any associated syringomyelia, which is seen in 30–70% of pediatric patients [55, 64, 98, 102, 106]. Historically, the pathogenesis of this intramedullary cavitation was explained using Gardner's hydrodynamic theory and William's theory of craniospinal pressure dissociation, in which a patent vestigial canal between the fourth ventricle and syrinx was suspected [35, 112]. However, over time, MRI studies have failed to completely confirm this hypothesis in a large number of patients with syringomyelia, leading to the more recent hypothesis that the formation and progression of syringomyelia results from a "piston-like" mechanism in which obstruction of CSF flow at the level of the foramen magnum during systole results in subsequent compression of the spinal subarachnoid space and inferior propelling of the syrinx [79]. Early identification of syringomyelia is critical since surgery has been shown to offer benefit

Table 1 Demographic and clinical profile of CNMC CM-I patients, diagnosed 1998–2018

Demographic information	
Sex	550 (51%) M, 523 (49%) F
Age at diagnosis	8.74 (0.2–36.7) years
Presentation	
Symptomatic	453/1073 (42%)
Incidental	620/1073 (58%)
Treatment summary	
Observation cohort	792/1073 (74%)
Surgical cohort	281/1073 (62%)
Type of surgery	
PFD	104/281 (37%)
PFD with duraplasty	177/281 (63%)
PFD with duraplasty and fourth ventricular stent	43/177 (24%)
Indications for surgery	
Worsening neurological condition	161/281 (57%)
Syrinx	149/281 (53%)
Scoliosis	55/281 (20%)
Radiographic deterioration	6/281 (2%)
Other	12/281 (4%)

to the patient [4, 26, 30, 42, 53, 59, 60, 77, 76, 78, 102, 106, 108], whereas delay in diagnosis may contribute to irreversible neurological compromise. While surgery may offer significant improvement or stabilization in symptomatic patients, it is less clear when, if ever, surgery is indicated in the asymptomatic syringomyelia patient who remains neurologically intact [46, 87]. However, many surgeons believe that the benefits of surgery outweigh the risks when dealing with a significant hydromyelic cavity [15, 45, 46, 66, 67, 82, 87, 89, 92, 102, 106], and there are countless reports of patients suffering from irreversible neurological deficits in the setting of syringomyelia despite successful although delayed surgery [7, 38, 75, 86, 88, 94]. Nevertheless, other contingents of neurosurgeons recommend waiting until the patient develops symptoms [46, 87, 89, 92]. Teenage patients may present with scoliosis, often in the setting of a large syrinx accompanied by a CM-I, and one may also see a prominent Chiari malformation in the absence of a syrinx but in the presence of a significant spinal deformity [5, 39, 52, 98]. A majority of neurosurgeons will offer a Chiari decompression in an attempt to reduce the size of the syrinx, which in turn has been shown to help improve the spinal deformity [3, 7, 13, 14, 26, 36, 42, 46–48, 51, 58, 87, 89, 92, 114].

Table 2 Reasons for MR imaging in patients with incidental CM-I, diagnosed 1998–2018

Reason for scan	Number (%)
Seizures	99 (16%)
Trauma	83 (13)
Non-specific headache	55 (9%)
Frontal headache	27 (4%)
Developmental delay	49 (8%)
Neurobehavioral	27 (4%)
Infection	21 (3%)
Endocrine	24 (4%)
Macrocephaly/microcephaly	29 (5%)
Surveillance (sickle cell/neurofibromatosis)	38 (6%)
Research study	3 (< 1%)
Other	165 (26%)
Total	620

Therapeutic options

To date, there is no consensus on the *management* of patients with incidental CM-I, which is seen in 0.5–3.6% of all pediatric patients undergoing brain MRI [2, 6, 25, 44, 70, 99]. Controversy abounds in every aspect of management, including indications, timing, and type of surgery as well as outcome [8]. The lack of uniform treatment guidelines can be attributed to several factors: (1) the clinical presentation is highly variable and often accompanied by less than clear subjective somatic complaints, (2) the pathogenesis of CM-I remains elusive and treatment algorithms are often made based on radiographic features, (3) the nomenclature and diagnostic criteria continue to be in flux, (4) the underlying etiology is often confounded by coexisting pathology (e.g., Ehlers-Danlos complex (EDS), migraines, etc.), (5) the paucity of literature on the natural history of the disorder, (6) the predictive factors

of neurologic progression are uncertain, (7) the long-term outcomes of surgical intervention have not been analyzed prospectively, and (8) comparative analyses of various surgical options remain less than complete. Over time, it is anticipated that a number of large, multi-institutional prospective studies, such as the Park-Reeves Syringomyelia Consortium and the University of Wisconsin surgical outcomes study, will help to address some of these concerns [96].

In the setting of tonsillar herniation 5 mm or more below the foramen magnum, clear and less controversial indications for surgery include (1) *intractable* occipital ≫ frontal headaches often exacerbated by exertion or Valsalva maneuvers, (2) development of neurological deficit(s) attributed to brain stem compression, and (3) the presence of a meaningful syrinx with or without scoliosis. Considerable debate has centered on the meaning of headaches with an associated CM-I, and this is challenging due to the subjective nature of headaches, as well as difficulty ascribing frontal or other non-occipital headaches to a hindbrain compression. Greater occipital pain than frontal/vertex discomfort with an exertional component offers successful symptomatic relief from a surgical decompression [3, 6, 60, 67, 84]. When there is connective tissue hypermobility or EDS comorbidity, it may be difficult to distinguish symptoms related to the ligamentous hypermobility/autonomic dysfunction from those directly related to cerebellar ectopia. Although it is not uncommon for this cohort of patients to have associated tonsillar herniation, the failure to recognize the etiology may result in non-therapeutic surgical intervention and potentially new issues for these complex patients. It is best to remain conservative in their approach unless there are clear-cut neurological changes or the development of a prominent syrinx. Although this cohort of patients usually benefits from standard Chiari decompression techniques, their recovery is often prolonged and it is not uncommon to have CSF hydrodynamic issues in this group [16, 49, 73].

Although there is an inordinate amount of data/publications on how to "correctly" approach a surgical patient with CM-I [4, 6, 8, 37, 81, 83], the long-term results are less transparent. Techniques have evolved that require minimal invasiveness such as non-bone resection with endoscopic intradural resection of tonsils [60], to bony decompressions of the arch of C1 and posterior ring of the foramen magnum/occiput, as well as classical bone decompression of the posterior vault/C1 in addition to subsequent intradural exploration for tonsillar manipulation and direct examination of CSF flow at the fourth ventricular outflow. This may also be done in the setting of intraoperative ultrasound or MRI visualization of the tonsils, and various adjuncts such as fourth ventricular stents or cranial cervical fusion may add to the complexity. The various approaches and lack of uniformity not only during the surgical intervention but also with respect to the long-term outcomes most likely results from an incomplete understanding of the underlying etiology for the radiographic appearance of tonsillar herniation and its subsequent effect on surrounding tissues. It is likely that there are a variety of reasons leading to the diagnostic radiographic appearance of the cerebellar tonsils. Unless the surgeon truly appreciates the underlying cause for the MRI appearance, it is likely that they will be unable to effectively treat the true cause of the patient's current condition. With respect to potential etiological determinants for tonsillar herniation, it is possile that the tonsils are low due to (1) inadequate volume in the posterior fossa [32, 57, 101, 118], (2) lack of egress for CSF outflow at the foramen Magendi/Lushka [21, 23, 69, 74, 102, 104, 106, 107], (3) elevation in supratentorial pressure contributing to a pressure gradient across the foramen magnum [9, 19, 22, 27, 28, 91, 97, 100], and (4) connective tissue hypermobility as seen in various conditions such as EDS, leading to cranial-cervical instability and often seen with concomitant pseudo-basilar invagination [41, 40, 49, 73] in addition to a tight posterior aspect of the cranial cervical junction.

Surgical techniques

If one is to offer a surgical approach to address the patient with Chiari symptomatology, it is imperative to weigh the known risks (unknown risks not withstanding) with the likelihood of successfully treating the underling indication for surgery (risk versus benefit). *It is also critical to attempt to understand the probable cause for the tonsillar herniation and address this with the most appropriate surgical option, if indicated*. Therefore, the presumed etiology should help to direct the ensuing surgical approach. Patients with a likely small posterior fossa (often younger patients) will benefit from a simple expansion of their posterior vault (Fig. 1), which may also be combined with an outer leaf durectomy. We previously demonstrated that increasing the volumetric expansion resulted in correlative improvement of the patient's symptoms [57]. By focusing on a bone-only decompression ± partial durectomy, one is able to maintain a low risk of CSF leak while offering an adequate expansion at the craniocervical junction. The use of intraoperative ultrasound (iUS) may facilitate direct observation of increased space at this location with direct, real-time changes at the tonsils and their interface with the brainstem. We have used iUS for the past decade to help guide whether there has been an adequate decompression or if a duraplasty and direct exploration of the CSF outflow at the obex needs to be undertaken (Fig. 2). When reviewing our past experience with Chiari surgical failures (Table 3), patients with large, holocord syrinxes frequently failed with non-duraplasty procedures and often required subsequent repeat surgery at which time it was necessary to open the dura for exploration of the fourth ventricular outflow and perform a duraplasty (irrespective of the intraoperative US findings) [29]. At the time of the second Chiari surgery, after the dura was opened, one could appreciate in > 90% of the cases

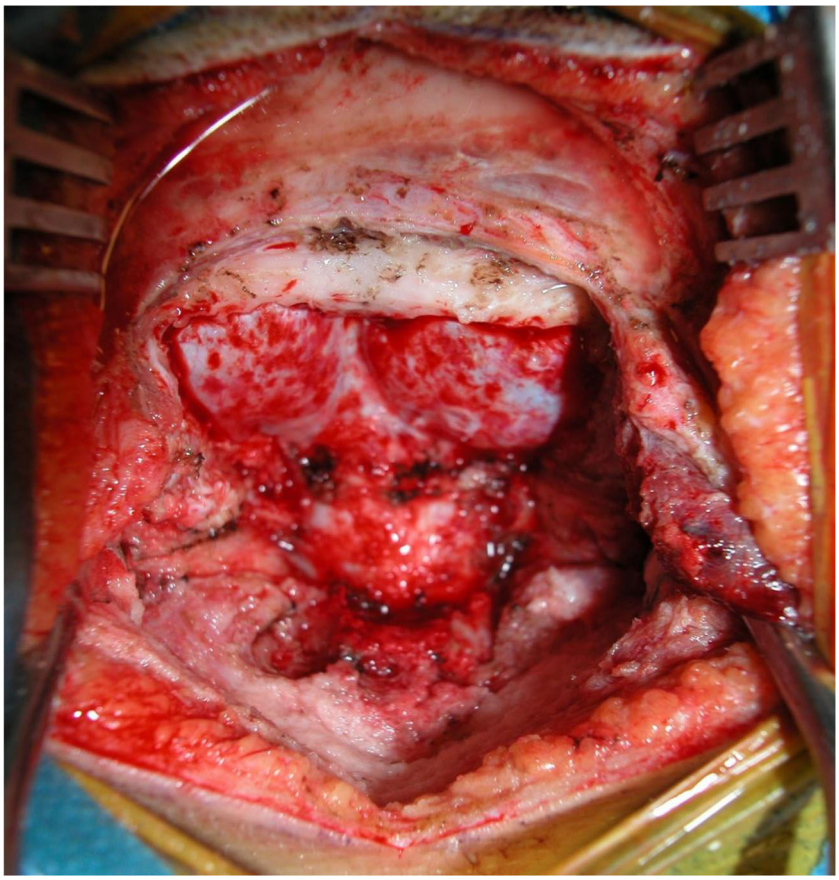

Fig. 1 Posterior fossa decompression (bone only) may be sufficient in patients with a small posterior fossa (often younger patients)

significant scarring at the obex, thus explaining the lack of success by not opening the dura. This significant scarring was also seen even in patients who had undergone previous open explorations complete with duraplasty and often required placement of a fourth ventricular stent in this area.

In the failed patients with the prior intradural decompression, it is unclear whether the scarring resulted from lack of prior exposure or was the result of the healing process, especially in the setting of tonsillar manipulation. We have nevertheless frequently utilized bone-only ± outer leaf durectomy

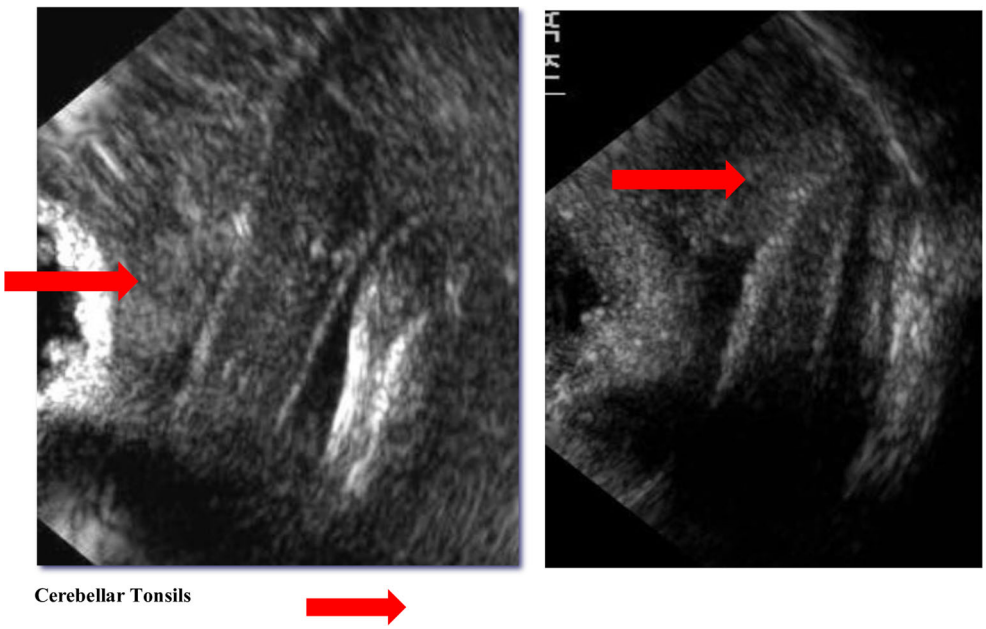

Fig. 2 Intraoperative ultrasound is used to determine whether adequate been an adequate decompression has been achieved and evaluate the need for a duraplasty and/or direct exploration of the CSF outflow at the obex. Cerebellar tonsils are marked with the red arrow

procedures for patients presenting with only pain or modest syrinxes and have seen excellent outcomes. However, patients with large holocord syrinxes or those with neurological deficits usually require a more involved surgical approach, requiring both bone and dural decompressions, complete with exploration of the fourth ventricular outflow (+/- 4th ventricular stent) and a dural graft.

In the setting of severe scarring at the obex or problematic location of either posterior inferior cerebellar artery (PICA), it is often safer and efficacious to place a fourth ventricular stent. This is illustrated by the clinical course of a 13-year-old female with a prior history of CM-I in the setting of EDS who has status post an expansile posterior fossa decompression (PFD) with duraplasty (bovine dural graft) from another institution. She presented to the clinic with intractable occipital and neck pain that had initially improved following the first surgery but returned later. An MRI of the spine revealed a large, significant holocord syrinx extending from the obex to the lumbar spine (Fig. 4a). Given the clinical and radiologic findings, a surgical re-exploration was undertaken. Intraoperatively, a significant bony defect (4 cm × 4 cm) extending from the suboccipuT to C2 was noted. Careful dissection of the dura under the microscope resulted in a jet of spinal fluid under moderately high pressure. Significant scarring and adhesions around what appeared to be previously amputated tonsils were noted, and the outflow of the fourth ventricle was covered with a horizontal band of dense, fibrotic tissue (Fig. 3a). Release of the bands of scar tissue restored the normal, pulsatile motion of the cerebellum and relieved the obstruction at the foramen of Magendie. Given the significant scarring and adhesions, a fourth ventricular stent was placed, with the top portion of the T-tube passed into the fourth ventricle and the caudal end laterally into the left paracervical border (Fig. 3b). This resulted in visible relaxation of the spinal cord and the release of a significant amount of CSF. The dura was then re-approximated as well as the previous bovine graft. Her postoperative course was uneventful. Follow-up imaging at 2 months revealed a near disappearance of the previous holocord syrinx with a very small residual component and significant symptom improvement with resolution of neck pain and reduction in occipital pain (Fig. 3a,b).

The general surgical approach is relatively consistent among surgeons but with occasional variation in technique. Patients at least 3 years old are placed in a prone position on

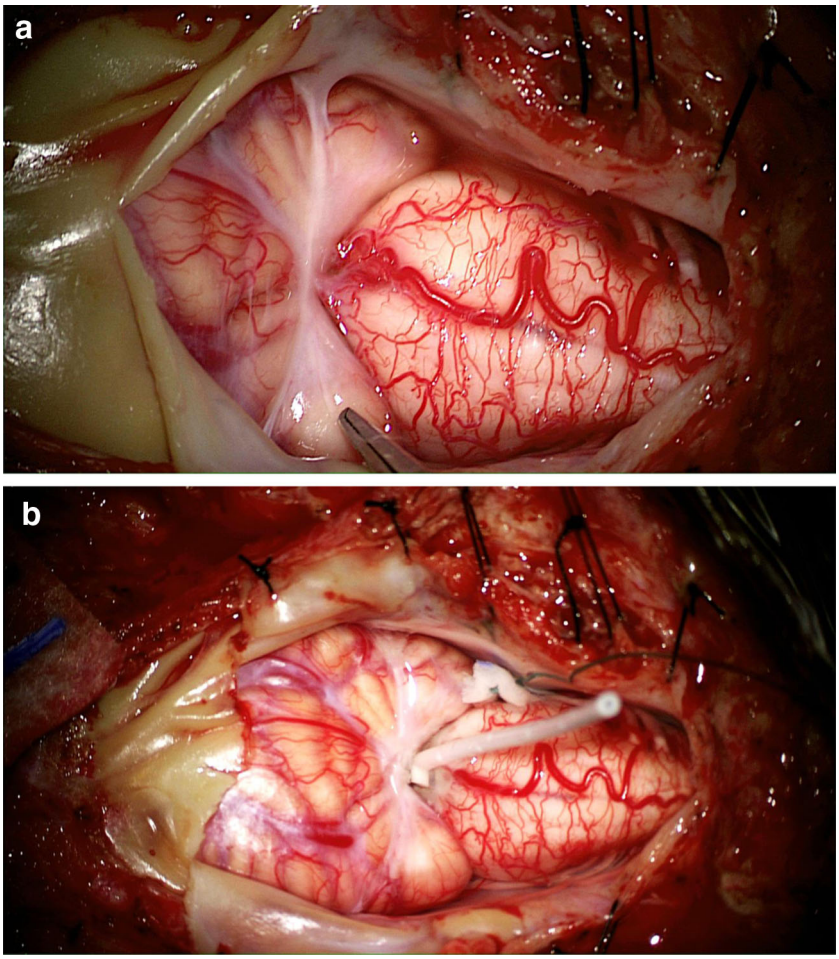

Fig. 3 **a** Significant adhesions and scarring from prior posterior fossa decompression with duraplasty and tonsilar manipulation are seen. The fourth ventricular outflow is obstructed by a horizontal band of dense, fibrous tissue. **b** Stent placement into the fourth ventricle is shown. The top portion of the T-tube is passed into the fourth ventricle and subsequently laterally into the left paracervical border

Table 3 Characteristics of surgical failures in CM-I, diagnosed 1998–2015

Failure type	Number (%)
Overall	19/195 (9.74%)
Posterior fossa decompression only	10/70 (14%)
Posterior fossa decompression with duraplasty	9/125 (7%)
Scarring at fourth ventricular outflow	17/19 (89%)
Required fourth ventricular stent	10/19 (53%)

This data is based on our previous study on surgical failures that was presented at the Congress of Neurological Surgeons, 2015, entitled *Bone-Only Chiari Decompression Failure Rate Is No Different Than That of Open Duraplasty*

well-padded bolsters with their head supported by pins or by a Mayfield head holder with judicious padding, if younger. Great care should be taken around the eyes and forehead to avoid any potential pressure points or intrusion into the orbital region (Fig. 5). The neck should be flexed adequately to allow two fingers to be placed between the chin and chest. Inadequate flexion fails to open the craniocervical junction sufficiently, whereas too extreme flexion can place the patient at risk for airway compromise or congestion at the orophyarngeal compartment with potential postoperative macroglossia. A narrow, 3-cm-wide microshave is employed from the inion to the C2 spinous process in the midline (Fig. 6) and prepped with standard chloroprep solution while ensuring that the patient has undergone "universal protocol" (confirmation of proper ID, planned procedure, perioperative supplies, and disposition) prior to the institution of prophylactic antibiotics and steroids if intradural exploration is anticipated. An arterial line, multiple intravenous lines, and a Foley catheter are inserted before the start of surgery. Injection of 5–10 cc of 0.5% lidocaine with 1/200,000 parts of epinephrine into the planned incision will help to minimize bleeding at the outset. A Colorado microcoagulation needle is used to make the skin incision (again to minimize bleeding), although many surgeons will use a scalpel. The incision is taken down to the occipital bone (leaving the periosteum intact to minimize bleeding) as well as the spinous processes of C1 and C2.

The iUS is brought in at this early point to see if the tonsils and brainstem can be visualized for a baseline study. This will be dependent upon the degree of neck flexion and the space between the foramen magnum and posterior arch of C1. Once the posterior arch of C1 is removed (to the lateral portion of the spinal canal), one frequently encounters a significant amount of fibrous bands across the dura beneath the prior bony arch of C1. This is removed under magnification and often affords greater visualization of the underlying tonsils on subsequent iUS images. On rare occasions, removal of this stricture will open up the craniocervical area of compression and demonstrate a

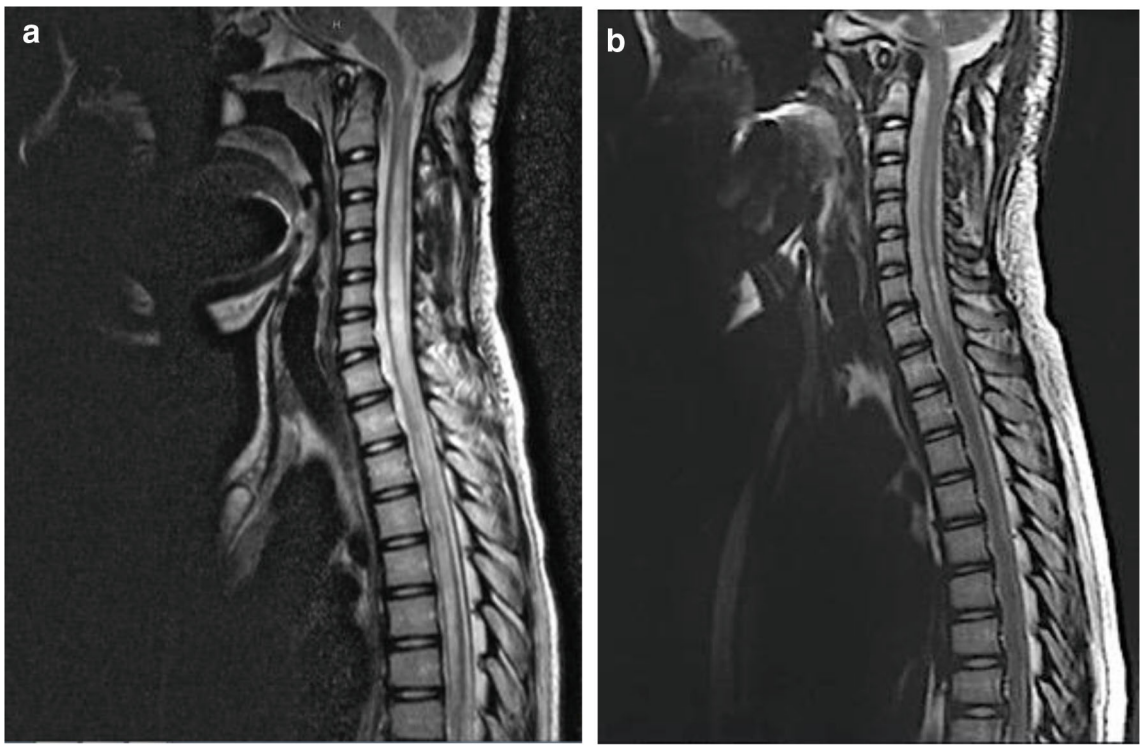

Fig. 4 **a** Preoperative MRI of the spine revealed a significant holocord syrinx (extending from the obex to the lumbar spine). **b** Follow-up imaging 2 months after a posterior fossa decompressive craniectomy, intradural release of adhesions, and placement of a fourth ventricular stent demonstrates near disappearance of the previous holocord syrinx

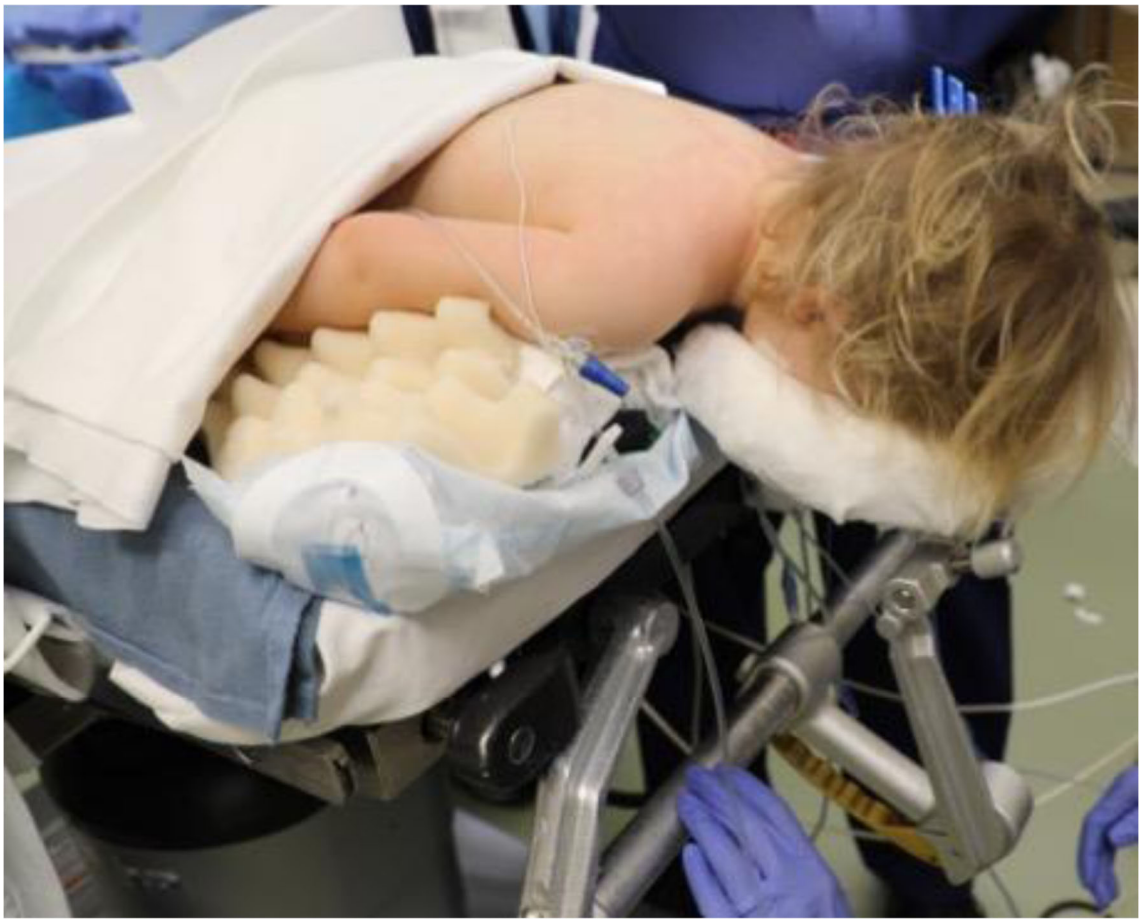

Fig. 5 The patient is placed in a prone position with the head supported by a Mayfield head holder with judicious padding around the eyes and forehead to avoid any pressure points or intrusion into the orbital region. Pins may be used in the older and larger patients for better support of the head, however with the added risk of intracranial injury from the pins

significant improvement to the tonsillar herniation (more space, less pointed tonsillar tips, less brainstem compression, less pistoning of the brainstem), as in Fig. 2. However, the majority of time it will be necessary to continue with additional bony decompression at the foramen magnum. Historically, large areas of the occipital bone were removed to provide adequate decompression, but this technique has grown out of favor due to the uncommon but extremely challenging condition of "brain sag," otherwise known as cerebellar ptosis. Our dissection of the occipital shelf at the foramen magnum usually involves a limited superior-inferior amount (2–3 cm) but a more involved extension to the lateral borders of the occipital bone. If one is to adequately address constriction at the craniocervical junction, it is imperative to extend the decompression to the lateral region of the spinal canal and offer more than 60°–90° of circumferential decompression. After the occipital shelf is removed, one can appreciate better visualization on the iUS as well as a more detailed view of the area of likely constriction at the junction between the occiput and spinal canal. Removing the occipital shelf on occasion will improve the iUS picture, especially in the younger patient with hypoplasia of the posterior fossa.

If the iUS shows considerable improvement at this point, the operation is finished. However, if there is little change manifested by the iUS, it will be necessary to remove the outer leaf of the dura from the craniocervical junction down to C2. This is done under the microscope and is facilitated by using a rounded microsurgical knife (such as a Beavor blade #69). When removing the outer leaf of dura, one often encounters modest bleeding from the vascular compartment between the two layers of dura. This low-pressure venous bleeding is easily controlled by hemostatic agents or gentle bipolar cautery (Fig. 7). After removal of the outer leaf of dura, the inner leaf is relatively thin and often transparent with the cerebellar tonsils pulsating below. Again, the iUS is brought in to assess whether the previous surgical maneuvers have helped change the position of the tonsils or offer additional space at this chokepoint. If imaging is less than satisfactory and there are clinical concerns that warrant an effective decompression at this junction (neurological deficits, syrinx), it will be

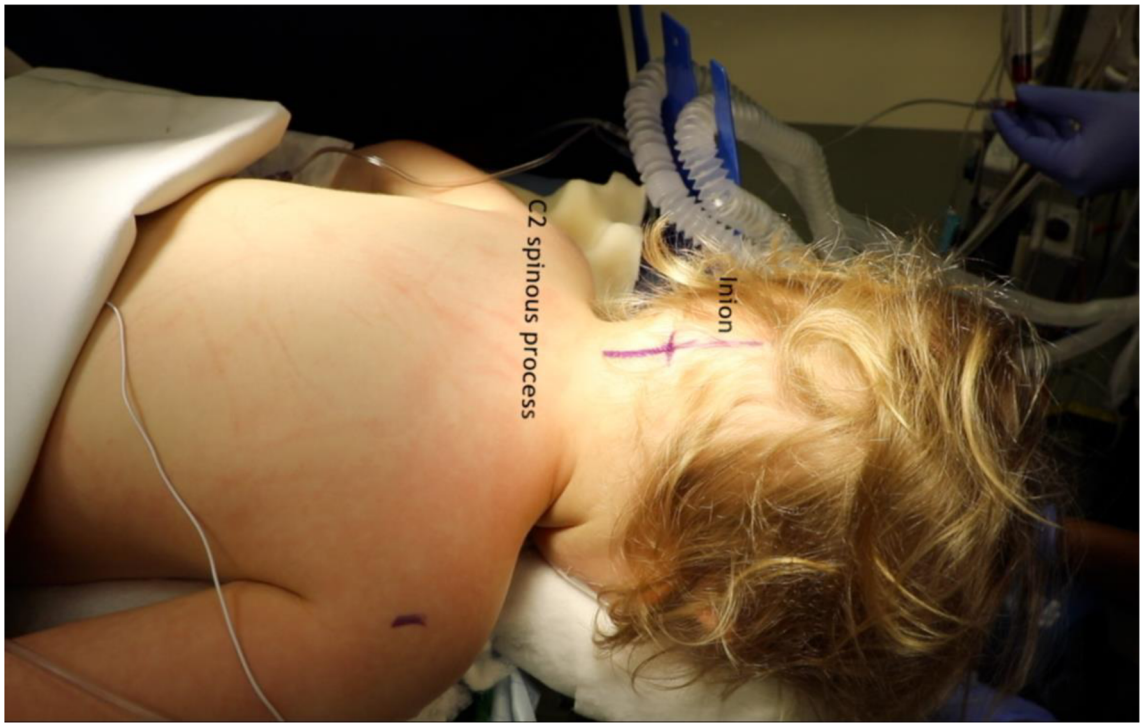

Fig. 6 A 3-cm-wide microshave is employed from the inion to the C2 spinous process in the midline

necessary to open the dura. Opening the remaining inner layer of dura as well as arachnoid is easily accomplished at this point and affords a direct view of the obex and surrounding tonsils as well as vasculature. If there are no visible adhesions at the CSF outflow, the decompression is considered complete.

A number of surgeons will coagulate the tonsils or even resect them to offer additional space for egress of CSF from

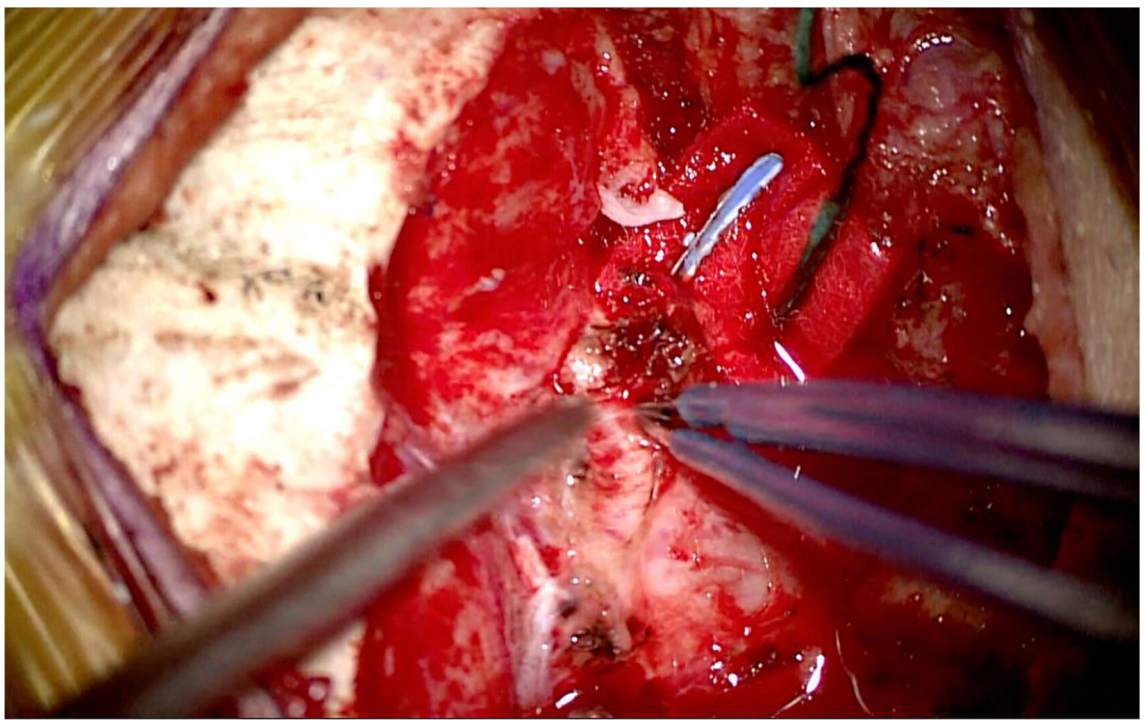

Fig. 7 Removal of the outer leaflet of the dura can result in low-pressure venous bleeding that can be easily controlled by hemostatic agents or gentle bipolar cautery

the foramen of Magendie. However, if possible, we attempt to limit direct manipulation of the tonsils if it is not necessary to ensure an unrestricted pathway for CSF from the fourth ventricle. Our experience at CNMC has demonstrated less surgical failures with less tonsillar manipulation, presumably from minimizing subsequent scarring at this critical choke point. In the event that there are considerable bands of adhesions at this outflow or large and relatively immobile blood vessels such as PICA, it is often easier and presumably safer to place a fourth ventricular stent across this area of CSF egress. One can use the top T portion of a syringoperitoneal shunt and cut this to an appropriate length to accommodate placement in the fourth ventricle (not the aqueduct or third ventricle) and have the distal end terminate below the C1/2 junction along the lateral portion of the spinal canal. The holes in the catheter allow placement of a 5-0 or 6-0 nylon suture to tack this down to the superficial arachnoid/dura to prevent potential migration of the catheter over time (Fig. 8). After the surgeon is confident that there is no restriction to CSF draining from the fourth ventricular outflow, attention is then made in performing a relatively water-tight duraplasty. Choice of duraplasty material is variable although the senior author prefers using pericranium due to its local proximity and lack of potential complications while also being cost effective. Neurolon™ (4-0) sutures are used in an interrupted or continuous fashion, and the dural closure may be facilitated by using Tisseal™ or another sealant material (fibrin glue) to help minimize any potential CSF leak. The remaining closure is undertaken with 2-0 or 3-0 absorbable sutures at the muscle and subcutaneous layers. Skin is closed with 3-0 Monocryl™ sutures, which is a monofilament that resorbs over time. A sterile dressing is applied although one can also use a postsurgical sealant.

Postoperative care includes steroids (Decadron) for 48 h, as well as antibiotic coverage for staphylococcus/streptococcus. Generous analgesics (Morphine, Toradol) are offered as well as liberal benzodiazepines (Valium) to ensure that the patient is comfortable and allows for effective healing of the CSF spaces. The patients are mobilized by the second postoperative day and often discharged to home on the third or fourth day depending upon the type of procedure and whether there are any concerns regarding a possible CSF leak. They are followed up at 1 month postoperatively and receive a new MRI 1–3 months later to assess any changes in the tonsils, craniocervical junction, or syrinx.

Surgical outcomes

The outcomes of PFD, both long and short term, are murky due to the myriad surgical approaches/techniques, paucity of randomized prospective studies, and significant variability in reported outcome measures; however, some general trends can be appreciated. The rate of surgical treatment for CM-I in pediatric patients has increased significantly in recent years reflecting a rise in CM-I diagnosis at earlier ages and renewing the age-old debate on the best surgical approach (intradural versus extradural) for this complex disorder [80, 111]. A review of the surgical series across different countries showed the use of PFD in 97%, dural opening in 81%, arachnoid opening and dissection in 18%, arachnoid opening and tonsillar resection in 43% (higher than adult series), and shunting in 4% of the pediatric series [68]. At our institution, 104 (37%) Chiari surgeries were bone-only PFD with/without outer leaf durectomy, whereas 177 (63%) were PFD with duraplasty. Among the duraplasty group, 43 (24%) also included a fourth ventricular stent (Table 1). Although the "classic" intradural approach involving a posterior fossa decompression with duraplasty (PFDD) ± tonsillar manipulation is still widely utilized, the literature portrays a shift toward a more simplistic extradural approach (PFD ± dural delamination) in pediatric patients [49]. Proponents of the extradural approach cite comparable outcomes to duraplasty/tonsillar manipulation with an enhanced safety profile owing to a lower risk of complications, particularly CSF leaks [15, 36, 54, 56, 61, 62, 77]. Critics argue that a bone-only decompression, despite the low complication rate, is insufficient in the long term, particularly in patients with syringomyelia, as evidenced by the high rate of repeat surgeries in this population. These authors recommend routine dural opening to facilitate decompression of neural structures and enhance CSF flow [34, 59, 63, 102, 106, 119].

Evidence for the success of the traditional intradural approach was demonstrated by the landmark study by Tubbs et al. on a large cohort of 500 pediatric patients who underwent surgical treatment with PFD, duraplasty (99.8%), and unilateral tonsillar coagulation (9.8%) for CM-I (57% with syrinx). Relief of preoperative signs and symptoms was observed in 83% of patients and syrinx resolution occurred in 80% after initial surgery (95% after second surgery). Repeat surgery was needed in a small group (3%) of patients with the prevalent indication being persistent syringomyelia, which resolved in all but two patients with second surgery when tonsillar coagulation was performed. Restriction of CSF egress from the foramen of Magendie secondary to arachnoid veil was seen in 12% of patients. Despite the well-documented risks of dural opening, the complication rate in this cohort was minimal (2.4%), leading the authors to recommend posterior fossa decompression with routine exploration for the patency of the foramen of Magendie [22]. While several subsequent studies have confirmed these findings in favor of an intradural approach in patients with syrinx [34, 59, 63, 82], this notion has been challenged by other authors that have demonstrated similar rates of syrinx resolution or improvement with a simple extradural approach [36, 67, 77].

A recent study by Massimi et al. evaluated long-term outcomes (mean follow-up 11.3 years) of bone-only PFD

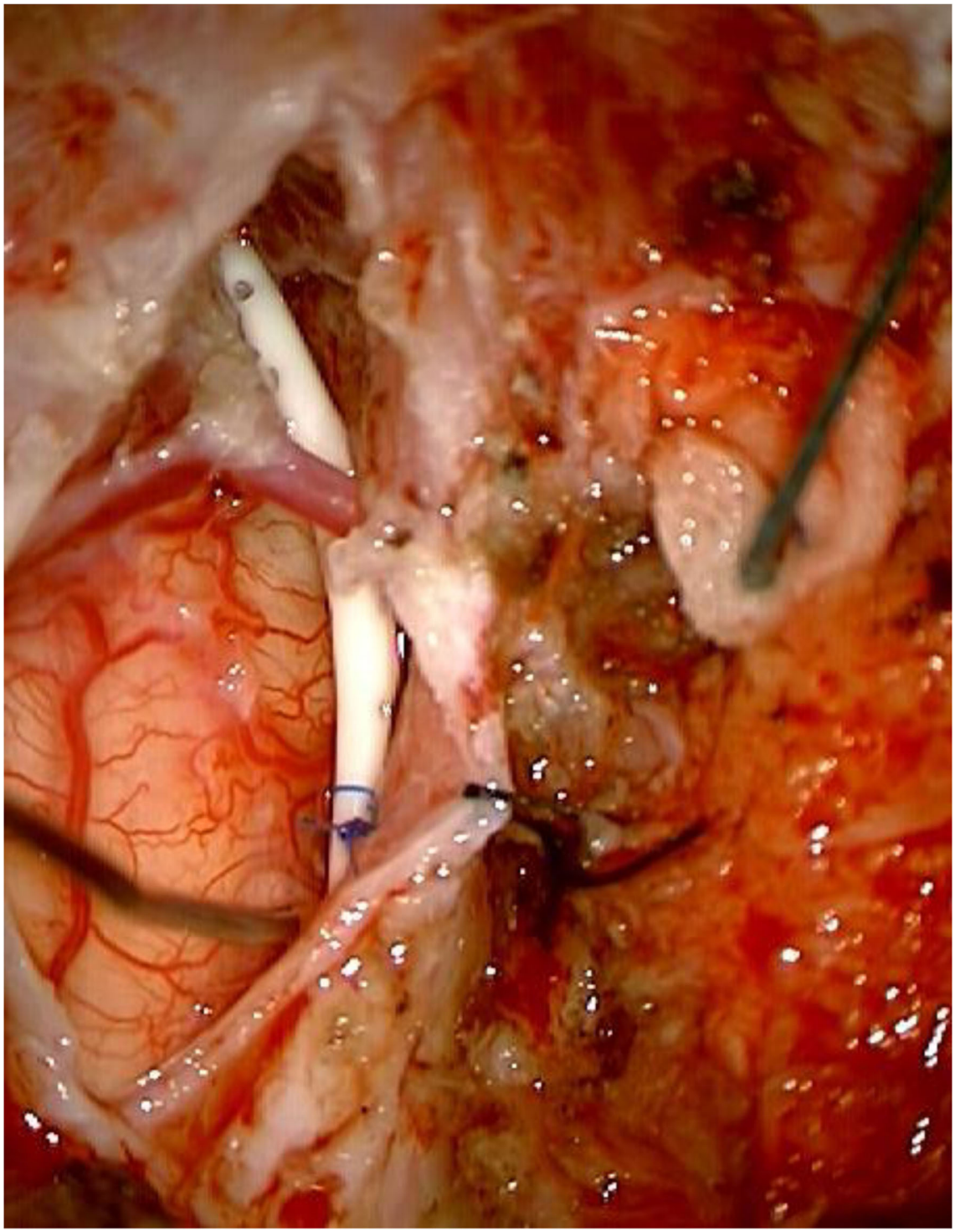

Fig. 8 The fourth ventricular stent is tacked down superficial arachnoid/dura to prevent potential migration of the catheter over time by placing 5-0 or 6-0 nylon sutures

(with/without C1 laminectomy and dural delamination) in children with CM-I (45% had an associated syrinx), and they observed disappearance or significant reduction in syrinx size in 31% and 48%, respectively. Clinical and functional improvement, however, were seen in 76.5% and 70%, respectively [67]. Other studies have cited syrinx resolution rates ranging from 50 to 94% and clinical improvement rates up to 100% with a bone-only decompression [15, 24, 36, 50, 54, 56, 61, 77]. Although the radiographic improvement with extradural decompression is lower when compared with an

intradural approach, it is important to note that radiologic improvement is often delayed (in both intradural and extradural approaches) and does not necessarily correlate with clinical outcomes [56, 110]. The advantage of PFD compared with PFDD is the minimal risk of complications. Studies have repeatedly shown a safer profile in favor of PFD. CSF leak, aseptic meningitis, and pseudomeningocele have demonstrated a higher incidence in the PFDD group [17, 24, 54, 61, 63, 65, 77]. Likewise, length of stay and operative time were also consistently higher in PFDD compared with PFD [61, 63, 65].

The primary concern with a simple bony decompression is symptom or syrinx recurrence requiring repeat surgery. Massimi et al. noted a 7% recurrence of symptoms occurring 1–7 years after initial surgery. One patient experienced newly formed suboccipital bone while another developed a thick fibrous scar, both of which resolved after a second extradural (bone-only) decompression. The third patient required two surgeries, a fibrous scar removal followed by duraplasty and tonsillar coagulation with minimal symptom improvement. The authors concluded that *dural opening may not always be a necessity in children* due to ongoing calvarial growth. These findings echo our own experience that *bone-only decompression is an effective initial treatment strategy for pediatric patients with CM-I, particularly those with a less severe clinical presentation* [67]. Our 20-year experience has failed to demonstrate any significant difference in symptom or syrinx resolution/improvement between the PFD and PFDD groups for those patients without a holocord syrinx (Table 4). However, complications, length of stay, and duration of follow-up were significantly higher in the PFDD group. While our rate of reoperation in the PFD group (14%) was double that of the PFDD group (7%), this did not reach statistical significance (Table 4), but we did note that patients with large, holocord syringes were the cohort who most often failed with a non-intradural surgery. Durham et al., however, found significantly lower reoperation rates in the PFDD group [24], while Lu et al. did not find any difference in revision rates between the two groups [65].

Despite the proven efficacy of extradural approaches, duraplasty with augmented intradural manipulation (tonsillar manipulation, obex plugging, stent) is still considered necessary in a subset of patients. In addition, there continues to be a debate in the setting of intradural exploration for the necessity of tonsillar manipulation, particularly in pediatric patients. The advantage of tonsillar manipulation is the elimination of adhesions obstructing CSF egress at the foramen of Magendie, which is particularly preferred in patients with syringomyelia. However, tonsillar manipulation itself may lead to the development of postsurgical adhesions resulting in surgical failures requiring further reoperations. In order to decrease this risk, tonsillar resection without a craniectomy or laminectomy has been proposed [60]. An analysis of complications associated with tonsillar manipulation by Navarro et al. in 96 patients who underwent one of three procedures: PFD with dural scoring, PFD with duraplasty, PFD with duraplasty, and tonsillar manipulation found a higher incidence of CSF-related complications in PFD with duraplasty (42.1%) and PFD with duraplasty and tonsillar manipulation groups (21.74%). By contrast, PFD with dural scoring group (and no intradural nor tonsillar exposure) only had a 5.6% complication rate. Patients with tonsillar manipulation had higher reoperation rates of as well as associated higher risk of morbidity. Interestingly, clinical outcomes were similar between the three groups. In light of these findings, the authors recommend leaving *the arachnoid intact* [77]. Others have highlighted similar concerns and recommended against tonsillar manipulation [42, 64, 119]. Galarza et al., on the other hand, did not notice any significant differences in complication rates between patients undergoing PFD with tonsillar resection compared with those who had PFD with or without duraplasty. Additionally, they noted a significant clinical and syrinx improvement in patients who underwent PFD with tonsillar resection [34]. Similar findings in favor of tonsillar resection were noted by Guyotat et al. with better clinical outcomes, no increased operative risk, and no additional surgery [45]. The importance of intradural exploration with tonsillar coagulation was similarly highlighted by Tubbs et al. who noticed obstruction of the fourth ventricular outlet by arachnoid veil in almost 8% of patients in the initial series and up to 12% in the subsequent larger series [102, 106].

The notion that arachnoid opening (and tonsillar manipulation) is necessary in patients with syringohydromyelia was challenged by Feldstein et al. who treated seven children with CM-I and holocord syrinx with PFDD but without intradural manipulation. Marked reduction in syrinx was seen in six of seven (85.7%) of patients at 2–4 months, which remained stable at 1 year. The single child without early improvement demonstrated a 50% reduction in syrinx size at 1-year follow-up imaging, suggesting that presence of a syrinx is not always an indicator for intradural manipulation [30]. Xie et al. also demonstrated significant resolution of the syrinx in 90.8% and acquisition of rounded tonsil shape in 91.8% of patients undergoing duraplasty but without intradural manipulation [116]. Our own experience with tonsillar coagulation was similar to Navarro et al., resulting in a higher rate of complications and surgical failures. The failure rate with and without tonsillar coagulation were 23.81% and 9.09%, respectively ($p = 0.02$) (Fig. 7). We found a significant reduction in failure rates from 13.3 to 4.9% following more judicious use of tonsillar coagulation. We therefore recommend against universal tonsillar manipulation in the pediatric population. Careful selection of patients based on intraoperative ultrasound is necessary to limit unnecessary complications and reoperations [20].

Overall, there is an increasing trend toward a simple extradural approach in pediatric patients with CM-I regardless

Table 4 Surgical outcomes in the PFD versus PFDD groups, diagnosed 1998–2015

	PFD	PFDD	*p* value
Total number of patients	70	125	
Mean age at diagnosis	9.1 years	9.3 years	
Mean age at surgery	10.7 years	9.7 years	
Mean time to surgery	12.5 months	6.4 months	0.01
Mean length of follow-up	42 months	47 months	0.50
Clinical improvement	62%	63%	0.59
Syrinx improvement	62%	63%	
Reoperation	14%	7%	0.11
Complications	3%	26%	0.0001
Length of stay	2.9 days	4.6 days	0.007

This data is based on our previous study on surgical failures that was presented at the Congress of Neurological Surgeons, 2015, entitled *Bone-Only Chiari Decompression Failure Rate Is No Different Than That of Open Duraplasty*

of the presence of syringomyelia. Favorable or comparable clinical, radiographic, and functional outcomes have been demonstrated along with an enhanced safety profile. The enhanced surgical outcomes in the pediatric cohort compared with adults highlights the importance of early surgical intervention in the child with CM-I.

Failures

The concept of failure for Chiari surgery may be as challenging to define as it is for the surgical indications. Whereas patients may present in a variable capacity, all predicated by the radiographic manifestation of hindbrain herniation, the objective criteria to base success upon may be more difficult to quantify. If one's surgical indications involve "intractable" occipital pain, a neurological deficit, or the presence of a meaningful syrinx, then it should be a simple matter to assess whether the patient is better, the same, or worse. The inherent difficulty comes from the challenge of assigning a quantitative value to a qualitative symptom. Most clinicians, for the sake of simplicity, utilize a binary system when it comes to pain and neurological compromise. Assessment of syrinx volume can be much more problematic, and most surgeons will assess whether the syrinx is different at its widest diameter as well as the extent of the syrinx relative to the longitudinal spine. This applies a modest degree of quantification in this setting, but in the end, it often comes down to whether the syrinx is smaller, the same size, or getting larger. The question of recurrence versus failure to improve may also be difficult to separate, especially if there has been minimal radiographic follow-up. For this reason alone, most clinicians will obtain a postoperative study to assess surgical outcome. However, it remains unanswered as to how long one should perform surveillance radiographic studies, even after the syrinx has completely resolved. While uncommon, it is not unheard of to have Chiari patients successfully treated by surgery return to clinic many years later (after a normal MRI) with new or unrelated symptoms and a new MRI demonstrating a large recurrent syrinx, despite prior resolution [7, 38, 86, 88, 94].

Another inherent challenge in assessing success or failure of prior surgical endeavors has been the relative paucity of such reports in the literature. While reported failure rates of a posterior fossa decompression with or without duraplasty range from 3 to 15% in large series [50, 65, 117, 119], there are a number of patients in whom the end-result is ambiguous when evaluating persistence of a syrinx or chronic pain. It is not uncommon for syrinxes to take time to improve (up to 30 months) [56, 110], and it may be difficult to meaningfully determine if a syrinx is better with respect to accurate volumetric measurement or when there are multiple compartments to the syrinx. This is especially challenging when the patient is asymptomatic except for the presence of scoliosis. Nevertheless, when assessing postsurgical failure in the setting of a persistent syrinx, it is important to rule out an underlying component of intracranial hypertension or occult hydrocephalus which may be contributing to the presence of the "fifth ventricle." A number of studies have documented the not uncommon comorbidity of hydrocephalus [21, 43, 95, 102, 106, 120] and when dealing with a large syrinx, especially of the holocord variety, it is important to observe closely for any evidence of supratentorial pressure and the subsequent complication(s) of a CSF leak, pseudomeningocele, or persistent holocord syrinx. In this setting, it may be necessary to perform a CSF diversion before the pseudomeningocele, CSF leak, or holocord syrinx improves. It may also be possible to temporize a CSF collection/leak with steroids or acetazolamide (carbonic anhydrase inhibitor), but ultimately, one must consider overall hydrodynamic forces and the CSF compartment.

When a supratentorial etiology (occult hydrocephalus) has been satisfactorily excluded, one must evaluate whether the egress of CSF from the fourth ventricular outflow spaces is satisfactory. There are a number of reasons why this may still be impeded, and was the most common factor in our own series of failures (17/19, 90%) among both extradural and intradural surgical approaches. In the event that a bony decompression ± outer leaf durectomy was performed, the lack of exploration at the obex most likely implicates an inadequate exposure of CSF egress pathways and in turn will necessitate an open exploration of the foramen of Magendie for lysis of adhesions and the possible need of placing a fourth ventricular stent. In the series of 100 consecutive Chiari decompressions in Boston, there were 16 failures, with the majority of these due to scarring of CSF outflow [88]. In this series, it was necessary to place a fourth ventricular stent in 44% of these patients to ensure that CSF could escape the fourth ventricle

and not be diverted down the central canal. In our own cohort over 20 years, we had 16% of our patients require a fourth ventricular stent for extensive fibrosis at this CSF chokepoint. In the Boston series as well as our own experience, there were no failures after stent placement.

Ultimately, when addressing a surgical failure, it is imperative to gain an insight into possible factors that may be contributing to the persistence of symptoms, neurological deficits, or volume of the syrinx. Thus, the importance of elucidating the most likely etiology for the Chiari in the first place if one is to successfully implement a therapeutic plan. If the patient appears to have a small posterior fossa, especially in a young child, and they remain symptomatic, it is critical to assess whether an adequate bony decompression was originally made or whether there is re-ossification of the area of prior bone removal (often seen in the patient less than 2 years of age). Not only is re-ossification common in this patient cohort (also seen in the achondroplastic patients), but one can also see a reconstitution of fibrous bands across the craniocervical junction at the foramen magnum as well as at the region of the previously removed posterior element of C1.

For patients who have an obstruction at the foramen of Magendie, and especially those with a syrinx, attention should be drawn to the obex to scrutinize for any unappreciated arachnoid bands at the outflow or the development of new fibrous adhesions leading to new or additional obstruction at this juncture. This is frequently observed after extradural decompressions in addition to patients who have already undergone an open exploration at this very location but did not undergo an adequate fenestration of their CSF outflow pathways. It is also possible that there may be delayed fibrosis as part of an iatrogenic event or excessive healing resulting in localized fibrosis. Ultimately, the value and long-term outcomes of tonsillar manipulation must be addressed in this matter. Does it make sense to potentially create more scarring or adhesions at the craniocervical chokepoint for CSF flow? There have been innumerable studies pointing out the safety and efficacy of manipulating the cerebellar tonsils via by coagulation or direct resection [10, 11, 31, 45, 60, 113]. On the other hand, there has also been increasing evidence that patients who undergo tonsillar manipulation are at a higher risk of requiring additional surgery as well as increased morbidity [42, 59, 64, 77, 119].

Our own experience at CNMC reviewed 281 surgical patients over 20 years (1996–2017) and looked at the first decade (1996–2007) at which surgical manipulation of the cerebellar tonsils was common and done in 74% of the patients undergoing an intradural procedure. The second decade brought forth a more conservative approach with respect to tonsillar anatomy, and in this cohort, only 42% of open procedures had any tonsillar manipulation. The reoperation rate for the earlier time frame (74% receiving tonsillar manipulation) was 23.8% vs 9% ($p = 0.02$) for the intradural patients who had no documented tonsillar involvement. We attributed the decrease in our reoperation from 13.3 to 4.95% ($p = 0.0125$) in part to this conservative approach and our avoidance of extradural decompressions for patients with holocord syrinxes [20].

Patients with a likely supratentorial contribution to their tonsillar herniation (idiopathic intracranial hypertension/pseudotumor, pansynostosis, arachnoid cyst, arrested hydrocephalus, or unappreciated shunt failure) should have the source of their tonsillar herniation addressed in an adequate fashion before expectation of a radiographic or subsequent clinical improvement. Ideally, this should have been addressed at the outset, but frequently, it may be difficult to determine the causative factor, especially in cases where there may be an underlying contribution from occult hydrocephalus. In cases with pre-existing ventriculomegaly or concerns about the function of a shunt, it is imperative to address this first before potentially opening the dura and risking the patient to a CSF leak or pseudomeningocele. It is safer and simpler to shunt the patient in question before proceeding with a Chiari decompression and not uncommonly, the tonsillar herniation or volume of the syrinx, go on to improve. Again, the suspected etiology should direct the appropriate therapeutic choice.

Finally, for those individuals who have contributing comorbidities such as EDS, fibromyalgia, chronic fatigue, or any other variety of somatic illness, it is critical to assess whether the tonsillar herniation is truly responsible for the patient's current symptoms. In the current environment whereby the incidence of tonsillar herniation is seen in 0.8–3.6% of the population [2, 25, 70, 99, 115], it is not uncommon to see associated tonsillar herniation in a patient with symptoms that mimic those found in Chiari patients. If the patient has surgery for a non-contributing Chiari malformation, it should not be a surprise if their symptoms fail to improve. Should their symptoms be attributed to their Chiari malformation, the underlying condition should help guide the surgeon's operative decisions.

Future considerations

Although the topic of Chiari I malformations continues to provide for lively debate on the academic platform, we anticipate that confusion surrounding the ongoing controversies in nomenclature, diagnosis, etiology, and management will decrease in the coming decade. A number of prospective, multi-institutional studies with robust statistical architecture are currently underway and are now beginning to answer a number of long-held questions about the management and outcome of this clinico-radiographic condition. It is unlikely that future data mining will offer much assistance in this matter due to the relative infrequency of this condition, but this too may change dramatically over the next decade.

Ultimately, it is important to recognize the etiology of the descent of the hindbrain/cerebellar tonsils (below the foramen magnum) if one is to ever truly understand the significance of the radiographic findings and offer a meaningful and efficacious approach to patients' symptoms. Possibilities today for tonsillar herniation include an inadequate posterior fossa volume, obstruction to CSF outflow at the obex, elevated intracranial pressure due to supratentorial influences, or disruption of craniocervical stability at the skull base from a connective tissue disorder. These are all diagnostic arenas that must be investigated if one is to arrive at the correct therapeutic approach. It is also critical to investigate any associated comorbidities as well as reviewing the genetic underpinnings of the overall process. In time, investigators will have more answers than questions, and it is at this point, the clinician will be able to better treat and care for this challenged population.

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Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

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