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Review Article

Updates on Surgical Strategies for Adult Chiari Malformation Type I: A Review

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Abstract

Chiari malformation type I (CMI) is a congenital neurological disorder characterized by the herniation of the cerebellar tonsils through the foramen magnum, which impairs cerebrospinal fluid circulation at the craniocervical junction. The primary hypothesis regarding its pathogenesis involves a mismatch between the posterior cranial fossa volume and the developing nervous tissue, leading to crowding and subsequent herniation. CMI presents a wide range of clinical manifestations, including cerebrospinal fluid-related symptoms, brainstem and cranial nerve compression, and spinal cord dysfunction, typically diagnosed through magnetic resonance imaging. The surgical treatment of adult CMI remains controversial due to its heterogeneous manifestations and the lack of standardized surgical protocols. Posterior fossa decompression (PFD), with or without duraplasty (hereinafter referred to as PFDD), remains the most common intervention. In this review, we focus on the following aspects to provide an overview of the current surgical strategies: 1. Surgical indications; 2. The extent of bony decompression in PFD; 3. Choosing between PFD, PFDD, and the dura-splitting technique; 4. Atlantoaxial fixation; 5. Techniques for intradural procedures; 6. Timing and approach for syrinx shunting. Additionally, emerging surgical innovations, such as endoscopic techniques, offer promising avenues for treatment.

Introduction

Chiari malformation type I (CMI) is a heterogeneous condition characterized by the herniation of the cerebellar tonsils through the foramen magnum (usually more than 5 mm) at the craniocervical junction, which is accompanied by a range of nervous and bony malformations, including basilar invagination, atlanto-occipital fusion, and syringomyelia, etc.[1–3] As our understanding of Chiari malformation has improved, differences in how these conditions are defined have become more apparent.[4] The classic CMI definition typically involves the caudal displacement of the cerebellar tonsils, distinguishing it from Chiari malformation type II, where the cerebellum, fourth ventricle, and brainstem are displaced downward with a high incidence of myelomeningocele.[2,4] A variant known as Chiari Malformation Type 1.5 is characterized by the downward displacement of both the cerebellar tonsils and brainstem, without the presence of myelomeningocele.[5] CMI discussed in this review includes the classic type and type 1.5 and excludes secondary causes such as intracranial tumors, hydrocephalus, intracranial hematomas, cranial trauma, or iatrogenic factors. Moreover, due to differences in etiology, symptoms, surgical decision-making, and operative techniques between adult and pediatric CMI, this review focuses specifically on adult CMI.

The prevalence of CMI in the general population is estimated to be between 0.24% and 3.5%, with a slightly higher prevalence in women.[3,6,7] The main pathological characteristic of CMI is the disruption of cerebrospinal fluid (CSF) circulation around the craniocervical junction or out of the fourth ventricle. The failure of CSF pulsations to effectively dissipate into the spinal subarachnoid space is believed to play a critical role in the development of syrinx formation.[8,9] Although the pathogenesis of CMI remains unclear, the dominant theory indicated that the discrepancy between the occipital bone and nervous tissue leads to crowding of the posterior cranial fossa, pushing the cerebellar tonsils downward into the spinal canal and obstructing normal CSF flow.[10,11] Recent research has suggested that variants in collagen genes, including COL7A1, COL6A5, COL1A2, and COL5A2, might also contribute to CMI.[12]

Surgery is the main treatment for CMI (the indications are discussed later). However, due to the diverse clinical manifestations

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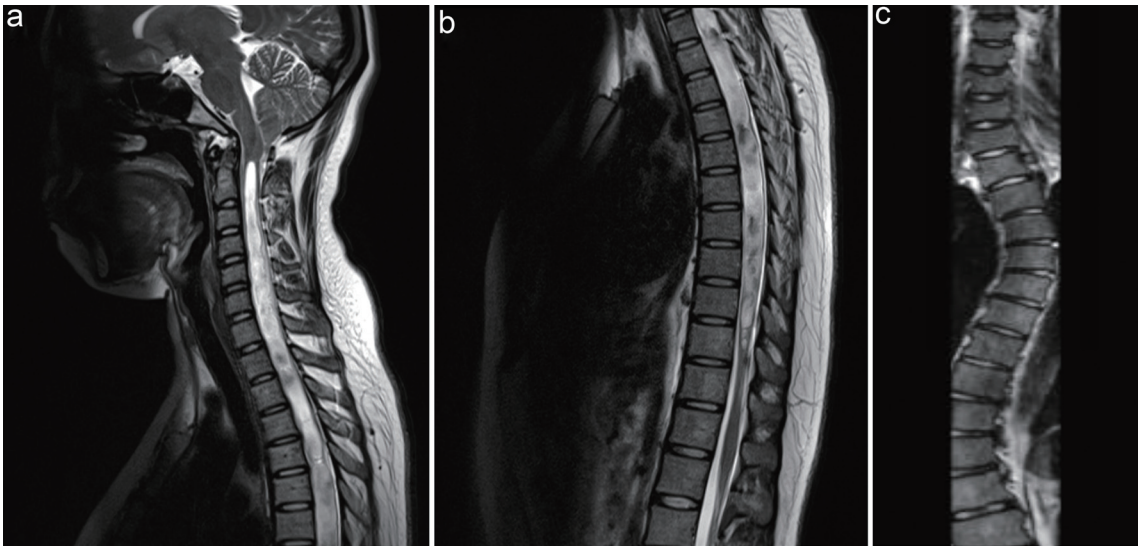

Fig. 1. Typical magnetic resonance imaging (MRI) findings of Chiari malformation type I. (a) Cerebellar tonsillar herniation accompanied by a syrinx at the high cervical spinal cord level. (b) A long-segment syrinx in the cervicothoracic spinal cord. (c) Associated scoliosis.

and poorly understood pathogenesis, the surgical approaches for CMI vary significantly, with different neurosurgeons adhering to their preferences. The choice of surgical method in different medical institutions often leans towards their habitual practices or experiences.[2] In this review, we will summarize our current understanding of CMI and focus on the clinical presentation, natural history, treatment methods, surgical indications, diverse surgical techniques, and prognosis.

Clinical presentation and imaging in CMI

The majority of CM-I patients are asymptomatic. In patients who do exhibit symptoms, the onset is often insidious, with an array of initial symptoms that can vary significantly between individuals.[13] In adults, the typical presenting symptom is pain or headache in the occipital and cervical regions, which can be exacerbated by Valsalva maneuvers, coughing, sneezing, or laughing.[14,15] Overall, the symptoms of CMI can generally be categorized into three groups:

1. CSF-related symptoms: Typical symptoms associated with impaired CSF flow include Valsalva-induced occipital or upper cervical pain/headache;
2. Compression of the brainstem, cerebellum, or cranial nerves: These symptoms include swallowing difficulty, choking, aspiration, dysphagia, central sleep apnea, nystagmus, tinnitus, and others. Neurological symptoms also include vertigo, autonomic dysfunction, and cranial nerve impairments such as trigeminal neuralgia or palatal weakness;
3. Spinal cord dysfunction (Syringomyelia): These symptoms primarily relate to dysfunction in the spinal cord, including motor and sensory disturbances. Common manifestations include sensory loss, motor weakness, scoliosis, spasticity, and upper or lower motor neuron signs.

The gold standard for CMI evaluation is magnetic resonance imaging (MRI), which helps in identifying the descent of cerebellar tonsils and associated conditions like syringomyelia.[16,17] Standard MRI focuses on assessing the distance between the cerebellar tonsils and the foramen magnum. Generally, a descent greater than 5 mm is considered indicative of CMI (Typical MRI scans refer to Fig. 1). The PB-C2 line is another important imaging measure in CMI, offering insights into ventral compression of the brainstem.[18–20] The PB-C2 line assesses the maximum distance from the odontoid process to the line from the basion to the posterior and inferior C2 vertebral body. According to published data, patients with a PB-C2 distance of 3 mm or greater had better outcomes in terms of symptom resolution and syringomyelia reduction after decompression surgery compared to those with smaller measurements.[21] Grabb *et al*.[20] proposed that patients with a PB-C2 measurement greater than 9 mm may require additional ventral decompression, as standard posterior decompression alone might not suffice.

Beyond conventional MRI, advanced imaging techniques like cine flow MRI (cfMRI) have gained popularity for their ability to assess CSF dynamics in CMI patients.[22,23] cfMRI is used to visualize and assess CSF flow and potential obstructions in the craniocervical junction.[22,23] Research by Wang and colleagues showed that preoperative cfMRI findings, such as CSF peak velocity greater than 2.63 cm/s, were predictive of favorable surgical outcomes, including symptom relief following posterior fossa decompression.[23] This suggests that cfMRI may be a useful non-invasive tool to evaluate the necessity of surgery for CMI, although care must be taken with its interpretation, particularly for patients with moderate CSF dysfunction, given variability in data across studies.

In addition, diffusion tensor imaging has been used to assess the integrity of white matter tracts in the brainstem of CMI patients.[24,25] Increased fractional anisotropy in CMI patients compared to non-CMI patients points to white matter abnormalities, which may improve after decompression surgery.[24,25] Diffusion tensor imaging, alongside cfMRI and standard MRI, provides valuable information on the structural and functional disturbances seen in CMI, thereby guiding both diagnosis and surgical planning.

Moreover, computed tomography and X-ray are valuable for assessing cranial base and spinal abnormalities in CMI. These imaging modalities help identify features like basilar invagination, posterior fossa volume measurement, and bony abnormalities at the craniovertebral junction, which are important for evaluating

the craniocervical junction stability and anterior compression.[26,27] Taken together, these various imaging modalities offer a combination of anatomical, functional, and physiological information, which is essential for understanding the complex pathophysiology of CMI. Their appropriate application, considering both clinical context and individual characteristics, guides surgical decision-making.

The natural history and conservative treatment of CMI

With the increasing prevalence of MRI examinations, the incidence of CMI has been on the rise, leading to more diagnoses in patients who are either asymptomatic or have nonspecific symptoms. Strahle *et al*.[28] reported on 147 CMI patients (mean follow-up of 4.6 years), where 133 patients had mild symptoms and only 14 underwent surgery due to symptom worsening. Novegno *et al*.[13] followed up with 22 CMI patients at their medical center who were temporarily treated conservatively (average follow-up of 5.9 years). Of these, 17 patients (77.3%) showed symptom improvement, while five patients (22.7%) experienced worsening symptoms, and three eventually required surgery.[13] In 2017, Langridge *et al*.[29] performed a systematic review to evaluate the natural history and conservative management of CMI. The authors revealed that 93.3% of asymptomatic CMI individuals remained asymptomatic, while a significant portion of symptomatic patients improved without surgery, especially those with headaches or nausea.[29]

According to the systematic review and evidence-based guidelines, it is recommended not to perform prophylactic surgery on patients with asymptomatic CMI without syrinx (recommendation strength grade C, level III evidence).[30] A small percentage of patients develop new or worsening symptoms during follow-up.[30] Thus, for patients with mild or minimal symptoms, conservative treatment with follow-up observation may be more appropriate. However, if patients experience progressive worsening of symptoms or present with a large syrinx during follow-up, surgical intervention should be considered.

Surgery indications

Posterior fossa decompression (PFD) alone or with duraplasty (PFDD) is frequently performed as a first-line surgical intervention to restore cerebrospinal fluid flow and alleviate symptoms.[31–33] Early surgical intervention in children with clear indications is recommended to minimize irreversible spinal cord damage and optimize surgical prognosis.

Based on the recent review of surgical guidelines for CMI, surgical intervention is generally indicated in the following circumstances[31–33]:

1. Patients with symptomatic CMI who fail conservative management or whose symptoms are worsening. This includes persistent headaches, neurological deficits, or other debilitating symptoms that do not respond to conservative treatment;
2. The presence of syringomyelia, significant cerebellar tonsillar descent, scoliosis, or other comorbidities like spinal cord compression, which suggest the risk of progressive neurological deterioration. In such cases, decompression surgery, with or without duraplasty, is often considered beneficial;
3. Craniocervical instability (e.g., basilar invagination, etc.) may require atlantoaxial or occipitocervical fixation with or without decompression to stabilize the region and prevent further neurological damage.

Surgical strategies for CMI

Due to the inherent structural abnormalities of CMI, surgery remains the primary intervention in patients with clear indications, as discussed above. Although various surgical techniques and their modifications are applied to CMI patients, most neurosurgical centers choose PFD alone or PFDD as the first surgery for CMI patients.[31–33] The primary aim of surgery is to relieve bony compression and effectively restore CSF flow at the craniocervical junction.[34] Arnautovic *et al*.[35] reviewed 145 English studies on CMI surgery published between 1963 and 2013, showing that 134 studies (92%) used PFD or PFDD for treatment. Studies report that 94–97% of CMI patients experienced symptom relief after PFD or PFDD surgery.[35,36] Long-term follow-up studies of CMI patients who underwent PFD or PFDD showed that over 90% of patients had stable or improved imaging and clinical symptoms after surgery.[37,38] Dr. Williams also proposed that PFDD with a sutureless dural graft could achieve a comparable clinical outcome to traditional PFDD with watertight suturing.[39] Other literature points out that precise removal of the C1 posterior arch can prevent sharp angulation between the posterior dura mater of the cervical spine and the decompressed dura mater, which is crucial for improving CSF circulation in the foramen magnum area.[40]

Due to the complexity of the pathogenesis and varied presentation of CMI, the standard surgical treatment remains controversial. In the following sections, we will focus on the following aspects for a comprehensive view of its surgical strategy: 1. The extent of bony decompression in PFD; 2. Choosing between PFD and PFDD; 3. The need for atlantoaxial fixation; 4. Techniques for intradural procedures; 5. Timing and approach for syrinx shunting; 6. Innovative advancements in surgical techniques.

The extent of bony decompression in PFD

The extent of bony decompression in PFD for CMI is a key aspect of the surgical strategy and remains a topic of considerable discussion. PFDD involves two major decompression components: decompression of the lower occipital bone and the removal of the C1 lamina. Currently, there is relative consensus regarding C1 decompression, with the standard practice being the removal of approximately 1.5–2 cm of the C1 lamina, depending on the extent of cerebellar tonsil herniation. However, opinions differ on the ideal amount of occipital bone that should be removed. In earlier research, some scholars proposed a more aggressive approach with extensive resection, extending superiorly to the transverse sinuses and laterally to the sigmoid sinuses, which theoretically creates greater space for the posterior fossa and alleviates crowding. However, aggressive decompression can increase the risk of complications. Klekamp *et al*.[41] reported that extensive posterior fossa decompression led to complications such as cerebellar sagging, while standard posterior fossa decompression provided better clinical outcomes. Later, Sindou *et al*.[42] described an expanded occipital foramen decompression technique, extending from one occipital condyle to the other, allowing for both posterior and lateral decompression. This method offers equivalent therapeutic effects to standard PFD but does not show significant advantages, and due to the need to expose both vertebral arteries, there is an increased risk of damaging major arteries during surgery, limiting its application.[42]

In recent studies, opinion favors small-bone-window decompression (around 3 cm), which aims to minimize bone removal to only what is necessary for adequate CSF flow restoration, thus reducing the potential for complications. The existing literature has shown that limited decompression, involving around 2–3 cm

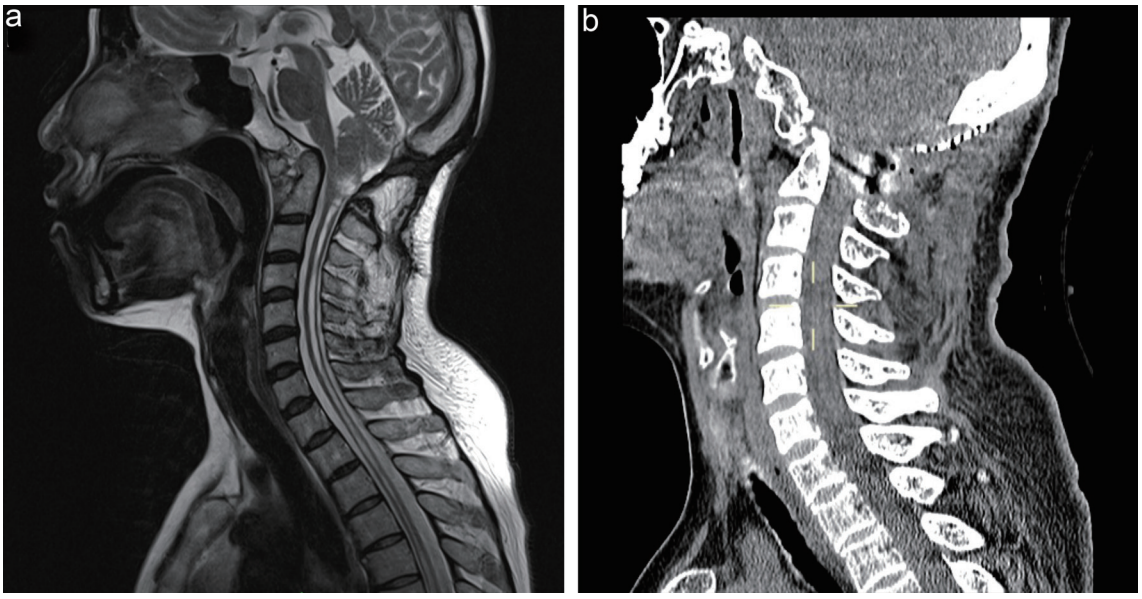

Fig. 2. Revision surgery for extended bony removal of posterior fossa decompression. A 50-year-old female Chiari malformation type I (CMI) patient underwent extensive decompression and experienced symptom recurrence one year after surgery. (a) Preoperative magnetic resonance imaging (MRI) showed cerebellar descent with compression at the foramen magnum and recurrence of syrinx. (b) Postoperative computed tomography (CT) scans demonstrated cranial reconstruction with occipital bone reshaping to support the cerebellum.

of occipital bone along with C1 laminectomy, provides effective symptom relief while minimizing surgical risks.[34,43] According to our center's experience, we prefer performing limited bone window decompression, which helps to prevent symptom recurrence caused by postoperative cerebellar sagging.[34] The presented case involved revision surgery for a CMI patient who had undergone extensive decompression before and experienced symptom recurrence one year post-surgery. Preoperative MRI showed cerebellar descent with compression at the foramen magnum. To elevate the position of the cerebellum, we performed cranial reconstruction with occipital bone reshaping to support the cerebellum, resulting in symptom relief for the patient (Fig. 2).

The choice between PFD, PFDD and dura-splitting techniques

PFD or PFDD is a crucial decision for treating CMI. PFD involves removing bone from the posterior fossa, enlarging the space and improving CSF flow at the craniovertebral junction.[44,45] However, PFDD goes a step further by opening the dura and adding a dural graft to create additional space and reduce pressure.[44,45] The choice between these two procedures depends on various clinical factors, such as symptom severity, the presence of syringomyelia, and the surgeon's specific expertise. Current studies illustrate the benefits and limitations of both approaches. Research has shown that PFDD is more effective than PFD in reducing symptoms such as headaches and improving syringomyelia, primarily due to better decompression of neural structures and restoration of CSF flow.[46] However, PFDD carries increased risks, such as cerebrospinal fluid leaks, infections, and pseudo-meningocele formation.[46] In 2018, Chai *et al*.[47] conducted a meta-analysis and systematic review involving 3,666 CMI patients, comparing PFDD vs. PFD. It found that PFDD was more effective than PFD in reducing syringomyelia, with a relative risk of 1.57. However, it also carries higher risks of complications, such as CSF leaks and aseptic meningitis.[47] Another systematic review and meta-analysis emphasized that the decision to use PFD or PFDD often depends on the surgeon's experience with each procedure and the specific clinical scenario. PFDD is generally recommended for patients with syringomyelia, as it offers a higher rate of syrinx resolution. In contrast, PFD may suffice for cases without a syrinx, as it is less invasive and carries lower complication risks.[48] Moreover, a large population study using the Park-Reeves Syringomyelia Research Consortium database showed that PFDD provides better clinical outcomes for CMI patients with syringomyelia, particularly in reducing syrinx size and improving symptoms like headaches. However, it also noted higher incidences of CSF-related complications, such as CSF leaks and pseudo-meningocele formation.[49]

In 1993, Isu *et al*.[50] proposed a novel dura-splitting technique to overcome the insufficient decompression seen with PFD, particularly in CMI with syringomyelia. The dura-splitting technique involves splitting the inner and outer dura at the foramen magnum, removing the outer dural layer while maintaining the inner layer intact.[50–53] By preserving the integrity of the inner dura layer, this technique lowers the risk of CSF leakage and related complications. In comparison with PFDD, which involves duraplasty, arachnoid dissection, and manipulation of the herniated tonsils, the dura-splitting technique is less invasive.[53] Studies have shown that while PFDD provides more thorough decompression and significantly improves long-term symptom relief and syrinx reduction, it is also associated with a higher complication rate.[53] In a study by Geng *et al*.,[52] PFDD demonstrated superior outcomes in terms of long-term syrinx reduction (8.09 ± 3.46 vs 5.73 ± 3.02 mm in diameter) and symptom improvement (75% vs. 47%) compared to dura-splitting. However, complication rates were significantly higher in the PFDD group, with CSF leakage and meningitis reported in 31.25% of cases, whereas no complications were reported in the dura-splitting group.[52] A recent meta-analysis of PFDD and dura splitting, which included ten studies with 370 patients, found fewer CSF-related complications, shorter hospital stays, and less blood loss in the dura-splitting group, with comparable clinical and radiologic outcomes to PFDD.[53] Thus, the dura-splitting tech-

Table 1. Summary of the systematic review and meta-analysis regarding the surgical treatment of CMI

Ref.	Year	Studies	Patients	Key findings
PFDD vs PFD				
Förander *et al*.[54]	2014	12	432	There is a higher re-operation rate in PFD, but clinical improvement is not higher than PFDD
Xu *et al*.[45]	2017	12	841	There is a significant difference in the operative time in favor of PFD compared with PFDD. There is a significant difference in overall complication rates and CSF leak rates in favor of PFD groups. However, there is a significantly higher improvement rate in favor of the PFDD
Lin *et al*.[48]	2018	13	3,481	Compared with PFD, PFDD led to a greater increase in operative time, a higher likelihood of clinical improvement in patients with syringomyelia, no increase of clinical improvement in patients without syringomyelia, but an increased rate of CSF-related complication, and a decreased likelihood of recurrence rate
de Oliveira Sousa *et al*.[55]	2018	27	1,352	All posterior fossa decompression techniques were very successful. The success rate with PFD techniques was 0.76 versus 0.81 in PFDD technique and 0.83 in PFDD with intra-arachnoidal techniques. The main complication was CSF leak, most common in patients with PFDD with intra-arachnoidal techniques. The overall mortality rate was 1%
Chai *et al*.[47]	2018	14	3,666	This study showed that the decrease in syringomyelia was better for patients with PFDD than PFD alone. The risk of CSF-related complications was higher in PFDD. No significant difference was found in the clinical improvement and the reoperation rate between PFDD and PFD
Tam *et al*.[56]	2021	17	3,618	PFDD is associated with better clinical outcomes but has a higher complication rate. In adults, clinical outcomes differences between PFDD and PFD did not reach statistical significance, but re-operation rates were higher with PFD. Patients with a syrinx did better with PFDD
Chenghua *et al*.[57]	2021	9	497	PFDD was related to a lower revision rate but a higher complication rate. No significant difference was noted between PFD and PFDD in terms of overall symptom improvement or syringomyelia reduction
PFDD vs Dura-splitting				
Tavallaii *et al*.[53]	2021	10	370	The study revealed significantly shorter operation duration and less intraoperative blood loss in the dura-splitting technique compared to PFDD. There was no significant difference between these two techniques in terms of clinical and radiological outcomes. Overall complication rate and incidence of CSF-related complications were significantly in favor of the dura-splitting technique
Tavallaii *et al*.[51]	2023	8	615	The findings revealed the significant advantage of the dura-splitting technique in terms of shorter operation duration and hospital stay. The recurrence rate and clinical and radiological outcomes were almost similar between the two surgical techniques. Complication rates were significantly lower in the dura-splitting technique

CMI, Chiari malformation type I; CSF, cerebrospinal fluid; PFD, posterior fossa decompression; PFDD, posterior fossa decompression with duraplasty.

nique may serve as an attractive alternative for selected patients, particularly those at higher risk of complications from duraplasty and syringomyelia. However, these studies should be interpreted with caution, as they are relatively limited and lack high-quality randomized controlled trials.

Taken together, controversies remain regarding PFD, PFDD, and dura-splitting techniques. To compare the three surgical techniques, we summarized recent systematic reviews and meta-analyses with their key findings in Table 1. In general, regarding clinical relief, radiological improvement, and reoperation rates, PFDD is superior to dura-splitting, and dura-splitting is superior to PFD. In terms of complications, CSF-related complications are significantly higher with PFDD compared to dura-splitting and PFD. In terms of hospital stay, surgery duration, and intraoperative blood loss, PFDD exceeds dura-splitting and PFD significantly.[45,47,48,51,53–57] Thus, the decision among these three techniques may depend on individual pathology, surgeon experience, and potential complication risks. In our clinical practice, for CMI with syringomyelia, we perform PFDD as the first-line treatment. This preference is primarily due to the complex etiology of CMI, which may include factors such as previous intracranial hemorrhage or meningitis. Even with advanced imaging techniques like cfMRI, it remains challenging to accurately assess arachnoid adhesions involving the cerebellar tonsils and the fourth ventricular outlets, making intradural manipulation crucial for restoring CSF circulation. Figure 3 illustrates intraoperative images from a revision surgery of a CMI patient with syringomyelia, who did not achieve symptom relief after an initial PFD. The adhesions found at the fourth ventricular outlet were identified as the cause of syrinx recurrence and the need for reoperation. Moreover, with proficiency in dura repair materials and microsurgical suturing techniques, the rate of CSF leakage following duraplasty using autologous fascia or suturable dural substitutes is not particularly high, and most CSF leaks can be managed conservatively.

Atlantoaxial fixation—Dr. Goel's approach

Based on the hypothesis that atlantoaxial instability is the primary initiating factor of Chiari malformation, Dr. Goel performed at-

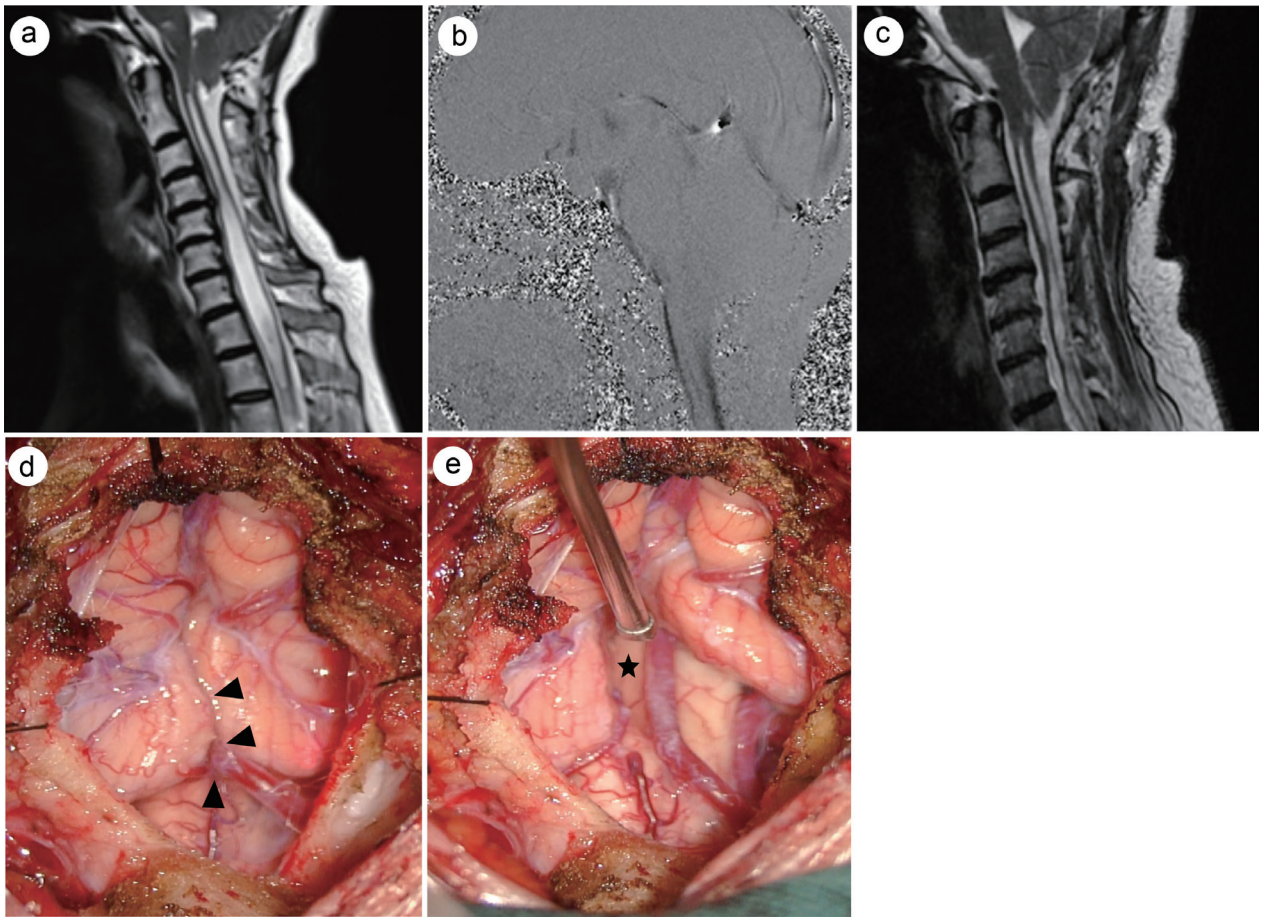

Fig. 3. Representative case for posterior fossa decompression with duraplasty. A 61-year-old female Chiari malformation type I (CMI) patient underwent posterior fossa bony decompression surgery 14 years ago. Postoperatively, the syringomyelia showed no significant improvement and repeated rehabilitation treatments were ineffective. Over the past year, symptoms worsened, leading to an inability to walk. (a) Before revision surgery, magnetic resonance imaging (MRI) showed cerebellar tonsillar herniation with recurrence of a large syrinx at the cervical level. (b) Cine flow MRI indicated cerebrospinal fluid (CSF) obstruction around the posterior foramen magnum. (c) Postoperative MRI demonstrated relief of the syrinx at the cervical level. (d) The intraoperative image showed extensive adhesions (black arrows) around the cerebellar tonsils and the outlet of the fourth ventricle. (e) The intraoperative image showed arachnoid dissection and opening of the outlet of the fourth ventricle (black star).

lantoaxial fixation to treat CMI.[58,59] Unlike the conventional view that Chiari malformation results from abnormal posterior cranial development, Dr. Goel's hypothesis emphasizes CMI as a condition stemming from instability at the craniocervical junction.[58,59] He argues that this instability causes herniation of the cerebellar tonsils, and that stabilization of the atlantoaxial joint can reverse this pathology.[60]

In a recent study published in 2023, Dr. Goel reviewed his publications and updated clinical material obtained over the last 12 years, involving 393 patients with central or axial atlantoaxial dislocation, 367 of whom presented with Chiari malformation, many of whom also had syringomyelia.[60] He applied atlantoaxial fixation in all cases, and significant improvements were observed in symptoms such as neck pain, paresthesia, and motor weakness. Furthermore, Dr. Goel found that atlantoaxial stabilization was effective in resolving syringomyelia in CMI patients (85%). His approach challenges the conventional need for decompression and suggests that stabilization of the craniovertebral junction could resolve symptoms more effectively and with fewer surgical complications.[60]

However, Dr. Goel's approach remains highly controversial globally.[61–63] After reviewing Dr. Goel's publications, it is evident that many of the cases he described might involve concomitant craniovertebral junction bony anomalies, such as basilar invagination, while some cases feature cerebellar tonsillar herniation of less than 5 mm, which does not meet the criteria for diagnosing CMI. Concerns raised by other colleagues focus on the validity of atlantoaxial instability as the initiating factor for Chiari malformation and the appropriateness of fixation in the absence of basilar invagination.[61,63] Additionally, critics question whether there is sufficient evidence to justify the use of atlantoaxial fixation over traditional posterior fossa decompression, especially considering the reported cases of spontaneous syringomyelia regression following decompression surgery.[61,63] There have also been arguments regarding the uniform application of atlantoaxial fixation for all congenital craniovertebral malformations, given the wide variation in clinical presentations and anatomical structures among patients.[61,63]

Techniques for intradural procedures

There is still debate over whether arachnoid dissection should be

performed in PFDD. Some studies suggest that arachnoid dissection is beneficial for removing local arachnoid scarring, especially around the fourth ventricle, thereby improving CSF circulation in revision surgeries. MRI findings showing segmentation or thickening of the arachnoid space at the craniocervical junction suggest that these patients may benefit more from arachnoid dissection. However, some opponents argue that manipulating the arachnoid membrane during first-time surgeries may promote the formation of scar tissue, and the dissection itself increases the risk of CSF-related complications. Chotai *et al*.[64] reported a study of 30 CMI patients who underwent PFDD combined with arachnoid dissection, with more than 90% of patients experiencing symptom improvement postoperatively. However, it should be noted that the CSF-related complication rate was high, with a 30% incidence of pseudomeningocele (23%) and CSF leakage (7%).[64] Recent applications of intraoperative neurophysiological monitoring (such as somatosensory evoked potentials and brainstem-evoked potentials) in PFDD have shown additional value in deciding whether to perform arachnoid dissection.[65] Although large-scale case studies are lacking, Grossauer *et al*.[66] reported a case of a CMI patient whose somatosensory evoked potentials significantly improved only after arachnoid dissection.

Currently, there remains debate among scholars regarding whether surgical manipulation of the cerebellar tonsils is necessary. Histopathological examination of the resected cerebellar tonsil in CMI patients has revealed varying degrees of gliosis and Purkinje cell loss in the herniated portion of the cerebellar tonsils. In a small number of cases, the distal part presented with cystic changes, which are believed to result from compression or trauma to the cerebellar tonsils, providing a histological basis for managing the cerebellar tonsils.[67] Moreover, according to the current understanding of CMI pathogenesis, management of the cerebellar tonsils serves two purposes: reducing the conflict between brain parenchyma and the volume of the posterior fossa and enhancing cerebrospinal fluid flow at the craniocervical junction to mitigate syrinx formation.

The main approach to managing the cerebellar tonsils involves coagulation, partial resection, and suspension.[68–70] No comparative randomized controlled studies have yet been conducted to determine whether different treatments of the cerebellar tonsils affect the outcome of PFDD. Lou *et al*.[71] reported a study of 130 CMI patients with syringomyelia who underwent partial resection of the herniated cerebellar tonsils (even without bony decompression), and postoperative symptoms improved to varying degrees in all patients. In seven cases with concurrent syringomyelia, the syrinx reduced in size.[68] Galarza *et al*.[69] reported that the "3R" approach (repositioning, coagulation shrinkage, and partial resection of the cerebellar tonsils) could improve outcomes for some CMI patients. In 2023, Braga *et al*.[70] retrospectively reviewed 437 CMI children to assess the efficacy and safety of different surgical techniques for cerebellar tonsils. Patients underwent various procedures, including PFDD, PFDD with arachnoid dissection, PFDD with cerebellar tonsil coagulation, and PFDD with subpial resection. Their findings showed that tonsil reduction techniques resulted in better syringomyelia reduction compared to arachnoid dissection alone, without significant differences in complications or reoperation rates.[70]

In our institute, Prof. Zhao proposed the application of cerebellar tonsil suspension (CTS), which involves suspending the cerebellar tonsils to the margin of the reconstructed dura via sutures on the basis of cerebellar tonsil coagulation (a step-by-step demonstration is shown in Fig. 4).[34] This method, combined with PFDD, can alleviate the relative anatomical narrowing of the posterior fossa while mitigating symptoms associated with tonsillar herniation and reducing the size of associated syringomyelia. Compared to cerebellar tonsil resection and coagulation alone, this approach minimizes structural brain damage, reduces injury to surrounding vessels through direct visualization, and provides more solid support to alleviate herniated cerebellar tonsils.[34] In our study, patients undergoing PFDD with CTS showed significantly greater cerebellar tonsil elevation (mean 7.06 ± 2.42 mm) compared to those with PFDD alone or PFDD with CTC (2.85 ± 1.45 mm and 1.57 ± 1.35 mm, respectively), indicating a more definite and stable elevation of the cerebellar tonsils that contributed to symptom relief.[34] Moreover, this method led to a notable improvement in syringomyelia, with a significant reduction in syrinx width and an increased percentage of reduction compared to other surgical regimens, making it an effective approach to improving patient outcomes while maintaining a less invasive strategy compared to cerebellar tonsillar resection.[34]

Timing and approach for syrinx shunting

The timing and approach for syrinx shunting in CMI is another critical area of debate. Syringomyelia is commonly associated with CMI and can cause progressive neurological deficits if left untreated. The first-line treatment for a syrinx is typically PFDD, which aims to restore normal CSF flow and promote syrinx resolution. In most cases, PFDD alone is sufficient, and the syrinx decreases in size or even resolves without further intervention.[48] However, in some cases, where the syrinx persists or continues to enlarge despite successful decompression, syrinx shunting may be considered.[72] Shunting involves placing a catheter to drain the syrinx into the subarachnoid space, pleural space, abdominal cavity, or other areas, reducing pressure and preventing neurological deterioration. The timing of shunting is crucial; prophylactic shunting may lead to complications such as catheter obstruction, infection, or injury to the spinal cord, while delayed intervention can result in irreversible neurological damage. Some surgeons recommend close monitoring with serial MRIs following decompression, reserving shunting for cases with persistent or symptomatic syringomyelia that do not respond to PFDD.[49] Overall, the decision to perform a shunting procedure should be individualized, based on the progression of symptoms and the response to decompression, to optimize outcomes while minimizing risks.[49]

Innovative advancements in surgical techniques

Recent literature suggests that endoscopy in CMI has made significant progress, particularly in PFD. Endoscope-assisted PFD, as detailed by Mobbs and Teo in 2001, offers advantages over traditional microsurgery by improving visualization and reducing tissue damage, leading to fewer complications and faster recovery.[73] Fully endoscopic PFD, proposed by Staribacher *et al*.,[74] has proven effective for certain patients by enhancing safety through surgical navigation and intraoperative monitoring, minimizing trauma, and shortening hospital stays. Dolaş *et al*. also demonstrated the successful application of fully endoscopic techniques in CMI, showing promising outcomes in symptom relief and decreased complication rates.[75,76] These endoscopic advancements provide a minimally invasive approach to treating CMI, underscoring their potential to improve clinical outcomes.

Limitations

Although this review summarizes the surgical strategies for CMI, it is also subject to several limitations. Firstly, as a narrative re-

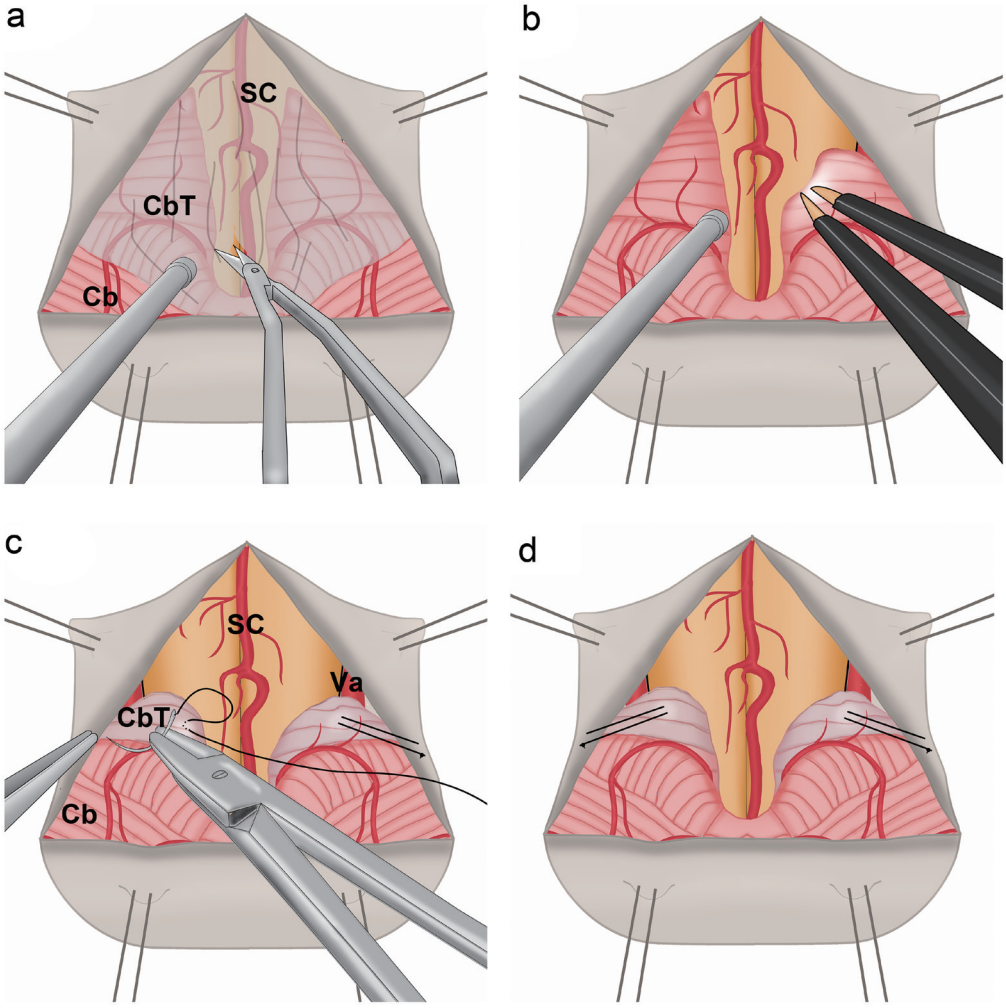

Fig. 4. Illustration of surgical procedures for cerebellar tonsil suspension (CTS) during posterior fossa decompression with duraplasty (PFDD). (a) Sharp dissection of the arachnoid membrane after a Y-shaped opening of the dura. (b) Coagulation of the left cerebellar tonsil (CbT). (c) Suspension of the right CbT with sutures. (d) CTS of the bilateral CbTs. Cb, cerebellum; SC, spinal cord; Va, vertebral artery.

view, it lacks a systematic review of the existing literature across different databases, which may lead to biases due to the potential omission of relevant research. The original and secondary statistical analyses are limited and cannot draw solid conclusions about CMI surgical strategies. Secondly, the review does not involve a thorough discussion of complicated CMI associated with craniocervical instability. Additionally, most of the current studies on surgical strategies in CMI are single-center, retrospective, and unstandardized, making it challenging to statistically compare the clinical efficacy and potential risks of different surgical approaches. Therefore, future research should focus on large-scale, multicenter, prospective, and standardized randomized-controlled trials to compare different surgical approaches and facilitate the standardization of CMI surgical treatment.

Conclusions

CMI is a complex condition characterized by the downward herniation of cerebellar tonsils, leading to the impairment of CSF flow. Its varied clinical manifestations in adults, along with a lack of consensus on standard surgical approaches, highlight the need for individualized surgical strategies. Surgical approaches such as PFD, dura splitting, and PFDD are the primary options. Among these, PFDD seems to hold the middle ground due to its combination of higher clinical efficacy and manageable complications. However, the decision-making regarding the three techniques, with their different intradural procedures, should be based on symptom severity, radiological findings, associated abnormalities, and patient-specific factors. Future studies should also investigate the clinical efficacy of atlantoaxial fixation, syrinx shunting, and endoscopic PFD.

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Conflict of interest

One of the authors, YCZ, has been an executive associate editor of *Neurosurgical Subspecialties* since July 2024. The authors have no other conflict of interest to note.

Author contributions

Conceptualization, resources, and writing-original draft preparation (LW, DL, YZ, HZ); methodology, image processing, and literature summarization (LW, DL); writing-review and editing (NT, HW, YZ); project administration and supervision (YZ, HZ). All authors have made a significant contribution to this study and have approved the final manuscript.

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