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Review Article

Scoliosis in adult Type 1 Chiari malformation with syringomyelia patients: from pathogenesis to treatment

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Abstract: The pathogenesis of type I Chiari malformation (CIM) is complex and remains unclear. The theory of posterior cranial fossa incompatibility has gained widespread acceptance in recent years. In the patients with CIM combined with syringomyelia, scoliosis is a common occurrence, with severe cases often leading to complications that necessitate surgical intervention. Scoliosis of various etiologies manifests as a three-dimensional structural spinal deformity. The etiology of scoliosis secondary to Chiari malformation is well-defined, and its imaging characteristics resemble those of idiopathic scoliosis. Vertebral rotation plays a critical role in the onset and progression of the deformity. However, spinal orthopaedic surgery in these patients is more challenging and hazardous due to the Chiari malformation and associated spinal cord abnormalities. Accurate preoperative assessment of scoliosis, Chiari malformation, and syringomyelia (SM), along with strategic planning of surgical approaches and fusion segment selection, is essential for improving patient outcomes and prognosis. In adults, CIM presents with more severe clinical manifestations compared to children, mainly including brainstem and cranial nerve compression, spinal-related syndromes, cerebellar injury syndromes, and hydrocephalus. Consequently, scoliosis deformities are often overlooked. Currently, there are limited reports on the efficacy of spinal correction surgery in adult patients with scoliosis associated with adult Type 1 Chiari malformation with syringomyelia (CIM-SM). Additionally, it remains uncertain whether and when syringomyelia should be addressed in these patients. This study aims to review previous research and explore whether intervention for Chiari malformation and syringomyelia should precede spinal orthopaedic surgery, as well as to evaluate the efficacy of spinal correction procedures.

Keywords: Type I Chiari malformation, syringomyelia, scoliosis, spinal orthopaedic surgery

Introduction

Type 1 Chiari malformation (CIM) is the most common type of Chiari malformation (CM) and is characterized by the presence of longitudinally oriented cavities, typically located in the cervical region of the spinal cord, often accompanied by spinal deformities [1-3]. It is reported that approximately 40% to 60% of CIM patients present with syringomyelia (SM) and varying degrees of spinal deformity [4, 5]. The typical clinical manifestations of Chiari malformation and syringomyelia include symptoms resulting from nerve damage. However, in many cases, scoliosis is initial presenting feature, leading to the neurological symptoms being overlooked or misdiagnosed as idiopathic scoliosis [6-8].

Clinically, 75% of scoliosis cases are idiopathic and of unknown etiology, with more than 30% of these patients requiring surgical intervention [9, 10]. While the exact relationship between Chiari malformation, syringomyelia, and scoliosis remains uncertain [11], it is clear that scoliosis is a clinical manifestation of both CIM [4, 5, 12]. In patients with CIM who develop severe or progressively worsening scoliosis despite bracing, spinal fusion becomes a necessary intervention to correct the deformity or halt its progression [7, 8, 13].

With advancements in internal fixation materials and fusion technology, increasing emphasis has been placed on achieving optimal scoliosis correction while preserving as much spinal mobility as possible and minimizing postoperative

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coronal and sagittal plane imbalances [8, 14, 15]. However, scoliosis correction surgery in the presence of Chiari malformation and syringomyelia is considered a high-risk neurosurgical procedure.

In 2015, Jakub et al. [15] compared the safety and outcomes of spinal deformity surgery in patients with CIM-associated scoliosis and a matched cohort of adolescent idiopathic scoliosis (AIS) patients. They found that while patients with CIM-SM achieved comparable deformity correction and prognostic scores, they also experienced higher rates of challenging neurological monitoring and surgery-related neurological complications. Notably, the study focused on adolescents, leaving it unclear whether similar complications occur in adults.

The effectiveness of posterior correction in treating CIM-SM remains unclear, prompting ongoing clinical research to evaluate its efficacy and potential complications. This study aims to review existing literature and analyze the outcomes and risks associated with posterior orthopedic techniques in CIM-SM patients.

The natural history of CIM-SM

Pathogenesis and epidemiology

Chiari malformation is characterized by the herniation of the cerebellar tonsil into the upper cervical spine through the foramen magnum [16]. It may be associated with pontine and bulbar extension, distortion, lower herniation or posterior cranial dysplasia, with type I being the most common subtype [8, 17]. The pathogenesis of CIM is complex and remains unclear.

The theory of posterior cranial fossa incompatibility: The theory of posterior cranial fossa incompatibility has gained significant recognition in recent years [18-20]. This theory posits that congenital dysplasia of the occipital protosegment results in a narrow posterior cranial fossa or the overdevelopment of its contents, causing overcrowding. This overcrowding leads to herniation of the cerebellar tonsils downward into the cervical canal or upward into the tentorium of the cerebellum, potentially resulting in syringomyelia and hydrocephalus [20-22].

Parameters such as the slope length and inclination angle of posterior fossa are widely used to evaluate its volume. These two parameters generally increase with age and stabilize by the twenties [23]. Huang et al. found that the severity of cerebellar subtonsillar herniation in CIM patients correlated with slope inclination angle, with smaller angles associated with more severe herniation [24]. Subsequent studies by Yang et al. confirmed this correlation, noting a link between slope dysplasia, syringomyelia formation and the severity of cerebellar subtonsillar hernia [22]. Adult CIM patients exhibit significant posterior cranial fossa dysplasia, with volumes markedly smaller than those of healthy adults, independent of gender.

Widespread fluid dynamic change: The 4D Flow technique, initially developed for cardiovascular applications, has shown promise in evaluating complex hemodynamics and cerebrospinal fluid (CSF) flow patterns in vivo. It has been utilized to analyze differences in CSF velocity between healthy controls and patients with and without Chiari malformation [25, 26]. Using 4D magnetic resonance flow imaging, researchers have observed widespread fluid dynamic changes at the craniocervical junction in patients with CIM-SM. These findings suggest that syringomyelia may results from abnormal CSF circulation at the foramen magnum caused by subtonsillar cerebellar herniation [27-29].

Pahlavian et al. emonstrated the reliability of 4D Flow for evaluating cervical CSF dynamics by validating its accuracy against in vitro models and numerical simulations [28]. Similarly, Williams et al. conducted a meta-analysis using in vitro models of type I Chiari malformation, confirming the consistency and repeatability of 4D flow in quantifying CSF peak velocity. These studies underscore the potential of 4D Flow as a diagnostic tool for quantifying CSF dynamics in Chiari malformation patients [27].

Muscle imbalance: Chiari deformity accompanied by syringomyelia disrupts fetal vertebral development and alters spinal pressure, leading to secondary postural reflex disorders and scoliosis [20, 30]. Additionally, in patients with CIM-SM, asymmetric damage to motor neurons in affected spinal cord segments can cause denervation of the trunk paravertebral muscles, resulting in muscle imbalances. Improper pulling forces from the paravertebral muscles on the spine lead to scoliosis [2, 31].

Table 1. Diagnosis methods of Chiari 1 malformation

MRI of the Spine	Tonsillar descent ≥5 mm below the foramen magnum is the conventional diagnostic threshold, but patients may have symptomatic tonsillar herniation with more modest descent.
	The measurement of posterior cranial fossa volume by MRI is helpful for the diagnosis of Chiari I malformation.
PCMR	Phase-contrast MR (PCMR) can be used in patients with syringomyelia to assess changes in cerebrospinal fluid circulation.
DTI	Diffusion tensor imaging (DTI) assesses damage to the intramedullary fiber bundle microstructure.
X-ray/CT	X-ray and CT can identify the complicated scoliosis and the changes of cervical canal and posterior fossa bone structure.
Clinical symptoms	The clinical symptoms of adult patients with CIM-SM are mainly neurological symptoms, including sensory disorders, motor disorders and autonomic nervous dysfunction. Neurological damage in patients with CIM is often insidious in onset and slow in progression, and is usually caused by atrophy and weakness of small hand muscles or sensory disorders.
Blood tests and Lumbar puncture	Blood tests are not helpful. Lumbar puncture is contraindicated as it may exacerbate the pressure differential between the intracranial and intraspinal compartments and cause further tonsillar herniation.

Note: PCMR - Phase-contrast MR; DTI - Diffusion tensor imaging; CIM-SM - Type 1 Chiari malformation with syringomyelia.

Adult CIM is predominantly characterized by tonsillar herniation and the absence of the cisterna occipitalis, with syringomyelia occurring in 50% to 75% of cases [8]. Neurological symptoms typically emerge in patients aged 20-30 years, often overshadowing spinal scoliosis, delaying its diagnosis and treatment. Consequently, orthopedic surgery for these patients is significantly more complex than that for those with idiopathic scoliosis.

Imaging examination and diagnosis

Occipitocervical MRI is pivotal for the diagnosis of CIM. However, a full spinal cord MRI scan is often necessary to identify additional spinal cord developmental abnormalities [32-34]. The T2-weighted sequence of MRI provides clear visualization of the neuroanatomical structures and precise locations of critical bone markers, enabling accurate morphological measurements while assessing lesions.

Most diagnostic criteria in current literature define cerebellar subtonsillar herniation as a descent of more than 5 mm below the foramen magnum plane [6, 35]. However, some experts suggest that a descent greater than 3 mm or beyond the line connecting the end of the clivus and the posterior margin of the foramen magnum is sufficient for diagnosis [36]. In recent years, some scholars have pointed out that relying solely on the degree of subtonsillar herniation is inadequate. Measuring posterior cranial fossa volume via MRI has proven valuable in diagnosing Chiari I malformation [34, 36].

For patients with syringomyelia, phase-contrast MRI (PCMR) is effective in evaluating CSF circulation changes, while diffusion tensor imaging (DTI) is useful for assessing microstructural damage to intramedullary fiber bundles. X-ray and CT imaging can provide insights into complex scoliosis and structural changes in the cervical canal and posterior fossa bone structure.

Clinical symptoms are also crucial for diagnosing CIM-SM. In adults, neurological symptoms dominate, including sensory disorders, motor impairments and autonomic dysfunction. Neurological damage in patients with CIM is often insidious in onset with slow progression, and frequently manifests as atrophy and weakness in small hand muscles or sensory deficits.

Table 1 summarizes the diagnosis methods of Chiari 1 malformation.

We believe that there is no absolute standard for diagnosing CIM-SM based on the degree of cerebellar subtonsillar herniation. Diagnosis should be made based on a combination of clinical symptoms, evidence of pathological processes such as syringomyelia, and MRI findings.

Should Chiari malformation or syringomyelia be treated before spinal orthopaedic surgery?

The abnormal hydrodynamics of CSF circulation at the occipitocervical junction plays an important role in the pathogenesis of CIM. Posterior fossa decompression (PFD), the standard surgical approach for CIM, restores normal CSF circulation at the occipitocervical junction and forms the pathophysiological basis for syringomyelia improvement [37]. Studies have reported that syringomyelia significantly improves in 90% of patients with six months of PFD after operation, and syringomyelia in about 40% of patients basically disappeared, and in some cases, progression halted entirely [38, 39]. Neurological symptom improvement rates following PFD exceed 94% [40]. However, irreversible spinal cord damage caused by syringomyelia can lead to inconsistent neurological outcomes post-surgery.

Surgical procedures for PFD: After routine disinfection, a single drape was laid and fixed to the skin with incision film. Tissue layers along the incision line were infiltrated with 0.25% to 0.5% procaine. The skin and subcutaneous tissues were incised midline, with the pericranium being incised midline above the external occipital protuberance and bypassed on both sides. Below the external occipital protuberance, the incision was made strictly along the midline ligament, reaching the occipital bone, the posterior tubercle of the atlas, and the spinous process of the axis. After incision and dissection of the tissues on the posterior tubercle of the atlas, the pericranium was incised transversely along the surface of the posterior arch on both sides and dissected with a dissector. The muscles on the spinous process of the axis and the lamina on both sides were dissected outwards. During the dissection process, electrocoagulation was used to control muscle bleeding, and bone wax was applied to stop bleeding from the emissary veins on both sides of the midline below the protuberance. An automatic retractor was used to open the incision. Then, a craniotomy was performed through a bone window, starting with drilling a hole on one side of the occipital squamous portion. After drilling through the skull, a rongeur was used to gradually remove the occipital bone. The dura mater was incised in a flap-like manner and flipped towards the transverse sinus, with an additional midline incision made below. The dura mater and upper cervical spinal dura mater were fully incised to facilitate decompression.

A long-standing debate exists regarding whether syringomyelia should be treated neurosurgically before scoliosis correction [38]. Mackinnon et al. first reported in 1969 that syringomyelia increases the risk of scoliosis correction surgery [41]. Subsequently, the case report of Nordwall and Phillips et al. also confirmed this point, suggesting that cavity drainage before spinal correction improves neurological function, reduces surgical risks, and prevents scoliosis progression to a certain extent in patients with immature bones [42, 43]. Sengupta et al. later recommended early hindbrain decompression for young patients under 10 years old with scoliosis and syringomyelia [44].

Current consensus holds that surgery should be performed only in symptomatic patients, with preventive surgery unsuitable for those with mild imaging abnormalities [45, 46]. PFD is prioritized in patients with syringomyelia, as it addresses the primary cause of symptoms and can halt scoliosis progression by restoring CSF circulation and intracranial pressure balance [38]. Early intervention is widely advocated to prevent neurological deterioration and assist in scoliosis correction.

Neurological symptoms, signs and their improvement after PFD are considered indicators of surgical success and predictors of scoliosis progression [46, 47]. Ono et al. found that patients with severe preoperative neurological damage were more likely to experience scoliosis progression post-PFD [8]. Attenello et al. reported that unresolved syringomyelia after PFD correlated with poor scoliosis prognosis [48]. However, Flynn et al. and Sengupta et al. suggested no significant correlation between neurological symptoms and scoliosis outcomes post-PFD [44, 49], highlighting ongoing controversies

Evidence indicates that PFD can improve scoliosis prognosis while treating neurological deficits. Factors such as age, preoperative scoliosis status, neurological symptoms, and syringomyelia condition can predict outcomes. However, most studies are based on limited clinical data, underscoring the need for large-scale, multicenter, prospective research.

Table 2. Treatment methods of CIM-SM patients

PFD	PFD is only suitable for patients with symptoms, and preventive surgery is unsuitable for patients with imaging abnormalities that not obvious. The purpose of the operation is to restore the cerebrospinal fluid circulation in the foramen magnum by decompressing the suboccipital bone and craniocervical junction, rebuild the intracranial and subarachnoid space pressure balance, reduce the pressure of the brain stem, reduce or eliminate the syringomyelia, and relieve clinical symptoms.
Spinal orthopaedic surgery (Posterior pedicle screw internal fixation system)	For scoliosis secondary to CIM-SM, patients are usually advised to undergo corrective surgery when significant curvature progression is observed. Posterior pedicle screw internal fixation system has been widely used in the correction of scoliosis in CIM-SM patients to obtain better coronal and sagittal correction results.

Note: PFD - Posterior fossa decompression.

While PFD can yield significant benefits, potential complications include CSF leakage, suppurative meningitis, acute intracranial hypertension, wound infections, and bleeding. Thorough preoperative evaluation is essential to minimize risks and optimize outcomes.

Table 2 summarizes the treatment methods of CIM-SM patients.

Application of spinal orthopaedic surgery in adult patients with CIM-SM

Type 1 CIM-SM is often accompanied by various neurological complications not typically seen in adult idiopathic scoliosis. Advances in magnetic resonance imaging (MRI) have enabled clearer differentiation between CIM-SM and adult idiopathic scoliosis [4, 7, 14]. The characteristics of scoliosis associated with CIM-SM include thoracic hyperkyphosis, early-onset, left- apex curves, and rapid progression, distinguishing it significantly from idiopathic scoliosis. Corrective surgery is generally recommended for patients with CIM-SM when significant curvature progression is observed [45].

In recent years, posterior pedicle screw internal fixation system has been widely employed to achieve better coronal and sagittal correction in CIM-SM patients [7, 46, 47]. However, the safety of scoliosis correction and posterior spinal fusion in these patients remains controversial, and the efficacy of such procedures is not well-documented.

A study by Ndiaye et al. demonstrated that optimal craniocervical osteo-dural decompression and duraplasty yielded early and sustainable clinical improvements in symptomatic CIM patients [50]. Another study by Shi et al. reported that somatosensory evoked potentials (SEPs) and motor evoked potentials (MEPs) in asymptomatic CIM-SM patients were comparable to those in idiopathic scoliosis patients [51]. However, CMS patients with syringomyelia exhibited lower SEPs amplitude and more severe spinal deformity, even after prior PFD surgery.

The presence of Chiari deformity and syringomyelia increases the risks associated with scoliosis correction, as downward displacement of the medullary spinal cord into the foramen magnum can lead to serious neurological complications during surgery [5, 7, 47]. For CIM-SM patients with neurological symptoms, scoliosis assessment should be a key component of preoperative evaluation. Accurate preoperative diagnosis and tailored surgical strategies for CIM-SM can enhance scoliosis correction outcomes and reduce neurological complications.

A recent study reported that primary posterior spinal orthopaedic surgery may be a viable option for CIM-SM scoliosis patients without significant neurological impairment [13]. In such cases, it is possible to achieve safe and effective scoliosis correction without prior neurosurgical decompression, and syringomyelia often stabilizes or improves postoperatively.

In general, current consensus indicates that the fusion strategy of CIM-SM with scoliosis is broadly similar to that of idiopathic scoliosis. For thoracic curvature, posterior spinal fusion with pedicle screws can yield comparable clinical and imaging outcomes. However, the presence of syringomyelia, necessitates a cautious surgical approach to minimize spinal cord tension and avoid excessive traction, which could result in severe neurological complications. In cases of extremely rigid CIM-SM with scoliosis,

treatment options include one-stage nail release, Halo femoral supracondylar traction, second-stage posterior spinal orthosis, or one-stage posterior three-column orthopedic osteotomy (**Table 2**).

Conclusion

Posterior fusion for the treatment of Chiari malformation with syringomyelia and spinal deformity has shown satisfactory correction outcomes, comparable to those achieved in adult idiopathic scoliosis. However, CIM-SM patients exhibit higher rates of neurological complications related to spinal fusion. During the perioperative period, close monitoring of neurological symptoms is essential. A limitation of this review is that it primarily discusses mainstream treatment strategies and their reported outcomes without providing direct evidence of clinical efficacy. Further studies, including large-scale and multicenter clinical trials, are needed to validate the safety and effectiveness of surgical strategies for CIM-SM scoliosis.

Disclosure of conflict of interest

None.

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