Indications for Surgery and Surgical Options in Chiari Malformation

WFNS Spine Committee Recommendations

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Study Design. A systematic literature review and consensus using Delphi method.

Objectives. This review aims to create recommendations on the surgical indications and approaches to treat Chiari malformation (CM) with or without syringomyelia.

Summary of Background Data. Despite the growing body of knowledge on CM, there are diverse and sometimes contradicting perspectives about surgical indications and procedures in both pediatric and adult populations.

Methods. The authors reviewed the literature on CM published from 2011 to 2022. Two consensus conferences were organized by WFNS Spine Committee. The first one was held in Sao Paulo, Brazil on August 2022, and the second one was held in Porto, Portugal on December 2022. Using the Delphi method, a panel of expert spine surgeons and members of the WFNS Spine Committee examined the strength of the literature, elaborated and voted statements about the surgical management of CM.

Results. We present 11 consensus statements on the surgical management of CM. Surgery is recommended for patients who have symptoms or if an MRI shows progression in asymptomatic patients. In pediatrics, osteoligamentous decompression only is indicated, whereas adults can have foramen magnum decompression with duroplasty, which is usually sufficient to control the associated syringomyelia. Syrinx drainage is the last option. Arachnoid opening can be performed in patients who have previously failed surgery or if arachnoid morphological anomalies are identified during the initial procedure. Tonsillar shrinkage provides somewhat better clinical efficacy than decompression alone, but at a larger risk of complications. Only patients with concurrent basilar invagination and atlanto-axial instability are advised to undergo atlanto-axial fixation alone.

Conclusions. The consensus statements created by a collaborative work provide useful information for surgeons treating CM worldwide to achieve better surgical outcomes and avoid complications.

Key Words: Chiari malformation, syringomyelia, foramen magnum decompression, cranio-vertebral junction stabilization

(*Spine* 2025;50:760–766)

Received for publication November 10, 2024; accepted January 30, 2025.

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The authors report no conflicts of interest.

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DOI: 10.1097/BRS.0000000000005288

In 1891, Hans Chiari described a group of congenital hindbrain anomalies, eventually named after him as Arnold-Chiari malformation.[1] He classified these malformations into three types (Chiari malformations I, II, and III) and 4 years later added the Chiari IV malformation.[2] The most recent and controversial definitions "CM1.5" and "CM0" were included in the Chiari classification during the last two decades as distinct entities.[3–5] Chiari malformation type I (CM1) is a common and often debilitating neurological disease. Efforts to improve the treatment of CM are often impeded by inconsistent and limited methods of evaluating clinical outcomes. To understand current approaches and lay a foundation for future research, some authors have conducted a review of

the methods used in original published research articles, concluding that, in many clinical studies examined, the outcomes in patients treated for CM are evaluated according to methods inconsistent and frequently not comparable.[6] Despite the growth of the current literature, mainly provided by the neurosurgical community, compared with the less experienced orthopedic one (although as well involved in craniocervical junction (CCJ) bone-related procedures) on Chiari, neurosurgical randomized controlled studies on significant case series to drive guidelines are missing both in the pediatric and adult populations. Because of several and sometimes contradictory opinions about surgical indications and techniques related to Chiari malformation, two consensus meetings were held by the World Federation of Neurosurgical Societies (WFNS) Spine Committee. Despite the differences in opinions even among the experts, these conferences aimed to reach a consensus majority guideline statement both regarding the indications for surgery and the surgical options that are available. In this paper, we report the most relevant recommendations about indications for surgery and surgical options that emerged from those meetings. Consensus guidelines are needed for the ideal management of ACM. We believe to have utilized a well-designed approach to formulate these consensus guidelines through a reputable forum. The categories made as symptomatic, asymptomatic, adults or children, with or without syrinx/Basilar Invagination/Atlanto-Axial Dislocation (BI/AAD) are precisely depicted. Furthermore, the type of treatments included all existing forms and presentation as well as the voting results depict the strength of the given statement for the reader to decide in their clinical practice.

METHODS

We reviewed the literature between 2011 and 2022 using a search with keywords "Arnold-Chiari malformation OR Chiari malformation OR Chiari type 1 OR Chiari syndrome OR Chiari OR Syringomyelia"; there were 58 results in PubMed, 122 results in Scopus and 101 results in Web of Science. Retrospective studies, clinical trials, meta-analyses, and practice guidelines were considered eligible for inclusion as described in Figure 1. We removed non-English language papers, case reports, and low-quality case series. Then, we analyzed 22 papers for this review.

Up-to-date information on Chiari malformation surgical indications and techniques was reviewed to reach an agreement in two consensus meetings of the WFNS Spine Committee. The first meeting was conducted in Sao Paulo, Brazil in August 2022. The second meeting was held in Porto, Portugal in December 2022. Both meetings aimed to analyze a preformulated questionnaire through a preliminary literature review. On the basis of the evidence from the literature review, statements based on the current evidence levels to generate recommendations through a comprehensive voting session. A total of eleven statements on CM and Syr were created and voted by spine experts and members of the WFNS Spine Committee. Voting was done using google voting through cell phones anonymously. We utilized the Delphi method to administer the questionnaire to preserve a high degree of validity.[7] To generate a consensus, the levels of agreement or disagreement on each item were voted independently in a blind manner through a Likert-type scale from 1 to 5 (1 = strongly disagree, 2 = disagree, 3 = somewhat agree, 4 = agree, 5 = strongly agree). No ethical approval was required for the present study as it only requested the opinions of clinicians, and no patient-specific data was involved.

RESULTS

We tried to find answers to the questions below:

What are the indications of surgery for Chiari malformation (CM)?

When and how to perform an osteoligamentous decompression alone for CM?

When and how to perform extra-arachnoid approach, dural opening and grafting for CM?

When and how to perform cerebellar tonsil resection and arachnoid dissection for CM?

Is there an indication of posterior C1 to C2 fusion for CM?

What are the indications of surgery for CM and associated syringomyelia?

Results were presented as a percentage of respondents who scored each item as 1 or 2 (disagreement) or as 3, 4, or 5 (agreement). The consensus was achieved when the sum for disagreement or agreement was $\geq 66\%$ as shown in Table 1.

The recommendations after two consensus meeting and voting results of the second consensus meeting are below under the title "Recommendations".

DISCUSSION

Despite the remarkable effort made over the past decades, the ideal surgical management of CM is still a matter of debate, raising more questions than answers. Not surprisingly, despite a plethora of papers and clinical recommendations "flourished" in recent years, there is a paucity of "well-proven" therapeutic guidelines when clinicians face the patient harboring tonsillar herniation in the setting of multiple objective and subjective complaints.[8–21] Controversy exists in every management aspect, including indications, timing, and types of surgery.

Indications of Surgery for CM

Regarding the indication for surgery in symptomatic CM1-Syr complex and abstaining from operating in asymptomatic isolated CM1, a total agreement was reached. In isolated CM1, surgery is indicated in the case of typical headache together with auditory/cerebellar/bulbar/ spinal signs at neurological examination, outlining the symptomatic CM1 or "Chiari syndrome". In asymptomatic patients, surgery may be an option, if MRI shows progression. Neurophysiological studies may help in the diagnostic phase both in CM1-A and in CM1-B or asymptomatic or mild CM1 patients because neurophysiological abnormalities

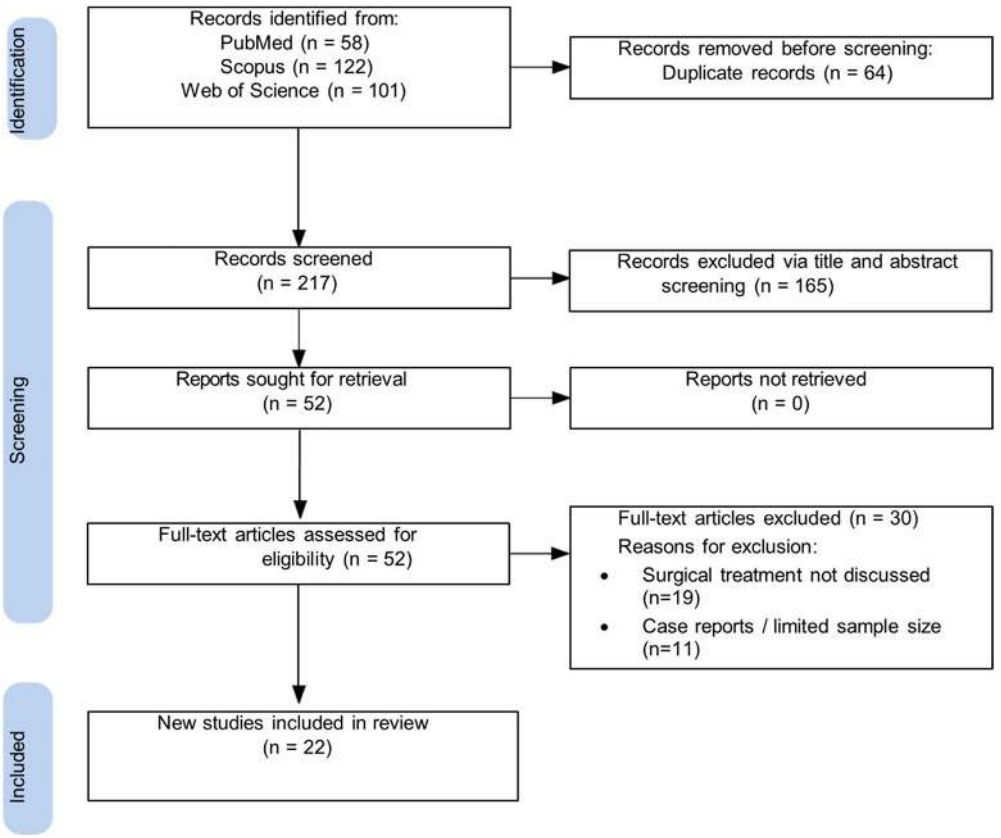

Figure 1. Flowchart detailing the search for pertinent literature regarding surgical indications and approaches in Chiari malformation patients.

could help in establishing objective evidence of subclinical dysfunctions that may indicate a need for surgery or to follow-up further progression.

Indications of Osteoligamentous Decompression Alone for CM

Several proposed techniques for posterior fossa decompression include osteoligamentous decompression of the posterior fossa/C1 to subsequent intradural exploration for tonsillar manipulation and direct examination of CSF flow at the fourth ventricular outflow.

Regarding the optimal entity of decompression, osteoligamentous decompression alone is recommended in the pediatric population, patients presenting with suboccipital headache only, not associated syringomyelia. The failure rate of posterior fossa osteoligamentous decompression alone is higher in the syringomyelia adult population. In this case, the extent of the bony decompression of the posterior fossa should be wide on the foramen, always including C1 laminectomy and never extended to C2 for the risk of CVJ instability.

Indications of Extra-Arachnoid Approach, Dural Opening, and Grafting for CM

Recently, the Congress of Neurological Surgeons performed a systematic review and evidence-based guidelines for patients with Chiari Malformation.[21] Concerning surgical recommendations, in patients with symptomatic CM1 malformation (with or without syrinx), either posterior fossa decompression or posterior fossa decompression with duraplasty may be utilized as a first-line treatment to improve preoperative symptoms (Class III C). When syrinx is present, surgeons may resect or reduce cerebellar tonsil tissue to improve syrinx and/or symptoms (Class III C).

Indications of Cerebellar Tonsil Resection and Arachnoid Dissection for CM

The role of intradural exploration in the treatment of CM1, particularly when associated with syringomyelia, remains controversial. Advocates for arachnoid dissection suggest that intradural disease, including arachnoid veils, webs, and medialized tonsils, may affect CSF flow through the foramen of Magendie. These advocates suggest that this partial or complete occlusion of CSF flow may result in syringomyelia formation because the presence of arachnoid disease is related to the presence of syringomyelia.

A meta-analysis[22] showed that, in adults, PFDD with arachnoid preservation has been observed to result in an increased rate of clinical improvement compared with other techniques and results in a lower incidence of total complications, CSF-related complications, and reoperation compared with PFDD with arachnoid dissection.

TABLE 1. Summary of WFNS Spine Committee's Recommendations on the Indications and Type of Surgery Indicated for Patients With Chiari Malformation With or Without Associated Syringomyelia

Indications for surgery		
1	In presence of CM type 1 with characteristic symptoms, surgery is indicated. In asymptomatic patients, surgery may be an option if MRI shows progression	Agreement: 90%
Osteoligamentous decompression alone		
2	On the basis of short-term outcome, the osteoligamentous decompression alone is recommended in: pediatric population, patients presenting with suboccipital headache only, not associated syringomyelia	Agreement: 80%
3	The failure rate of osteoligamentous decompression alone is higher in: syringomyelia, adult population.	Agreement: 80%
Extra-arachnoid approach, dural opening, and grafting		
4	Symptomatic patients with CM type 1 with/without syrinx may have foramen magnum decompression with duroplasty	Agreement: 60%
5	Although available recent literature shows no significant advantage of subarachnoid dissection over extra-arachnoid approach, arachnoid opening could be done in patients with failed previous surgery	Agreement: 90%
6	For patients with CM, the extra-arachnoid approach is a safe procedure showing globally fewer complications than the subarachnoid dissection in short-term follow-up.	Agreement: 80%
Tonsil resection and arachnoid dissection		
7	Arachnoid membrane may be opened when intraoperatively arachnoid morphological changes are seen.	Agreement: 80%
8	In the short-term follow-up, tonsillar shrinking has slightly better clinical efficacy when compared with decompression alone; however, it may lead to a higher rate of complications.	Agreement: 90%
Posterior C1–C2 fusion		
9	Atlanto-axial fixation alone is recommended to treat CM1 patients without BI and AAI	Agreement: 30%
Associated syringomyelia		
10	The available scientific data suggest that syringomyelia is a manifestation of CSF obstruction at cranio-vertebral junction and does not represent a separate entity in CM type 1.	Agreement: 90%
11	Decompression of CVJ in CM in the form of either PFD or PFDD is sufficient enough to control the associated syringomyelia in most of the cases. In case of previously failed surgery, symptomatic patients with enlarging syrinx re-exploration or duroplasty may be considered. Syrinx drainage is the last option.	Agreement: 90%

AAI indicates atlanto-axial instability; BI, basilar invagination; CM, Chiari malformation; PFD, bony decompression with band excision; PFDD, bony decompression with duroplasty.

Arachnoid preservation reduces the risk of complications and maintains the quality of posterior fossa decompression and should be considered a preferable treatment option in adult patients with CM1.

Although available recent literature shows no significant advantage of subarachnoid dissection over the extra-arachnoid approach, arachnoid opening could be done in patients with failed previous surgery. This statement reached an almost unanimous consensus (an overall 90% agreement).

Another statement reaching more than 90% overall positive consensus is that in the short-term follow-up, tonsillar shrinking has slightly better clinical efficacy when compared with decompression alone. However, it may lead to higher complications. Actually, a serious concern in the surgical treatment of CM1 is the risk of CSF-related complications, including CSF leak and pseudo-meningocele, which overall may occur in up to 21.8% and are directly related to the incision of the inner dural layer.[23,24] The risk of CSF-related complications can be reduced by using an arachnoid-preserving technique, providing a watertight seal, and preventing leakage.[25,26]

BI is defined as the invagination of the high cervical spine into the cranial base. CM can be associated with a certain rise of the dens, even though the degree of invagination is not sufficiently substantial to meet the diagnostic criterion of BI.[27,28] An "accordion" phenomenon has been demonstrated in a CM patient as a reappearance of the dens after its surgical removal and of the CM.[20] Moreover, most atlanto-axial dislocation cases present with superior dislocation of the dens.

Indication of Posterior C1 to C2 Fusion for CM

In 2015, Goel published his theory of atlanto-axial instability as the cause of CM[29] Goel claims that CM with or without syringomyelia is caused by atlanto-axial dislocation, regardless of the presence or absence of basilar invagination, considering basilar invagination, CM and syringomyelia as a continuum of the same pathological phenomenon that originates from atlanto-axial instability. Accordingly, he performed atlanto-axial fixation in cases in which there was CM with or without syringomyelia and with or without basilar invagination. The main stone explanation of the Goel's Theory is: "The tonsillar part of the cerebellum herniates into the spinal canal to provide a protective cushion for the craniocervical cord in an effort to protect the neural structures from getting pinched between the bones[29–31]

So in Goel's theory, the C1 -C2 fusion represents the pathophysiological surgical interpretation of the assumed basic dynamic mechanism of Chiari "Formation". Again: "simultaneously, "water" or "neuroaqua" delivers its motherly protective properties by increasing its presence inside (syringomyelia) and/or outside the spinal

cord (external syrinx) and inside (syringobulbia) and/or outside the brainstem and cerebellum (external syringobulbia)".[30,31]

Nevertheless, although it was felt that the need for atlanto-axial stabilization in all cases of CM deserves further studies and pathophysiological explanations. As not all Chiari patients present with C1 to C2 instability, a consensus on atlanto-axial fixation alone was not reached (overall 70% disagreement). Thus, atlanto-axial fixation alone is not recommended to treat CM1 patients without basilar invagination (BI) and atlanto-axial instability (AAI).

Indications of Surgery for CM and Associated Syringomyelia

The association between CM and syringomyelia is frequent, with a reported incidence ranging from 35% to 75% of the cases.[32,33] Symptomatic syrinx may present with sensory disturbances, bulbar symptoms, along with spinal deformity.

Although foramen magnum decompression and band excision (FMD + BE) is almost unanimously considered the mainstay of treatment, there is still debate regarding the extent of decompression needed. There is no clear consensus on which procedure is the most beneficial.[34] Whichever surgical technique is performed, persistence or progression of syrinx despite FMD is reported in more than 30% of cases, with neurological worsening in some cases.[35,36] FMD with expansion duraplasty (FMDD) may successfully reduce the gradient pressure at the cranio-vertebral junction, thus restoring an orthodromic cerebrospinal fluid (CSF) flow dynamics at the CVJ and reducing the syrinx filling mechanism concurrently.

Nonetheless, other techniques have also been proposed with the aim of getting rapid decompression of the syrinx, such as concomitant FMD with syringo-subarachnoid shunt.[35–38] In contrast, the limitations of this approach are well-known mainly for the high risk of complications.[39] Other surgical strategies reported for addressing persistent/progressive syrinx after FMD include syringopleural and syringoperitoneal shunts.[40–43] A large number of complications have been reported after syrinx shunting in terms of shunt obstruction/dislocation, tethering of the spinal cord by the shunt, and a low CSF pressure state may occur after shunt placement.[44–46]

Decompression of CVJ in CM in the form of either PFD (bony decompression with band excision) or PFDD (bony decompression with duraplasty) is sufficient to control the associated syringomyelia in most cases. In case of previously failed surgery, symptomatic patients with enlarging syrinx re-exploration or duraplasty may be considered. Syrinx drainage is the last option. This statement reached 90% agreement.

Recommendations

The final recommendations after the second meeting on indications of surgery and surgical options for Chiari malformation are as follows:

Indications of surgery for Chiari malformation

In the presence of CM type 1 with characteristic symptoms, surgery is indicated. In asymptomatic patients, surgery may be an option if MRI shows progression (90% consensus)

Osteoligamentous decompression alone for Chiari malformation

On the basis of short-term outcome, the osteoligamentous decompression alone is recommended in the pediatric population, patients presenting with suboccipital headache only, not associated syringomyelia (80% consensus)

The failure rate of osteoligamentous decompression alone is higher in the syringomyelia, and adult population. (80% consensus)

Extra-arachnoid approach, dural opening, and grafting for Chiari malformation

Symptomatic patients with CM type 1 with/without syrinx may have foramen magnum decompression with duraplasty (60% consensus)

Although available recent literature shows no significant advantage of subarachnoid dissection over the extra-arachnoid approach, arachnoid opening could be done in patients with failed previous surgery. (> 90% consensus)

For patients with CM, the extra-arachnoid approach is a safe procedure showing globally fewer complications than the subarachnoid dissection in short-term follow-up. (> 85% consensus)

Cerebellar tonsil resection and arachnoid dissection for Chiari malformation

Arachnoid membrane may be opened when arachnoid morphological changes are seen during surgery. (80% consensus)

In the short-term follow-up, tonsillar shrinking has slightly better clinical efficacy when compared with decompression alone. However, it may lead to higher complications. (> 90% consensus)

Posterior C1 to C2 fusion for Chiari malformation

Atlanto-axial fixation alone is recommended to treat CM1 patients with basilar invagination (BI) and atlanto-axial instability and not recommended for those who do not have basilar invagination or radiologically demonstrable atlanto-axial instability (AAI) (> 70% consensus)

Surgery for Chiari malformation and associated syringomyelia

The available scientific data suggest that syringomyelia is a manifestation of CSF obstruction at cranio-vertebral junction and does not represent a separate entity in CM type 1. (90% consensus)

Decompression of CVJ in CM in the form of FMD + BE or FMDD (bony decompression with duroplasty) is sufficient to control the associated syringomyelia in most cases. In case of previously failed surgery, symptomatic patients with enlarging syrinx re-exploration or duroplasty may be considered. Syrinx drainage is the last option. (90% consensus)

CONCLUSIONS

The present study is the result of a huge and cooperative work project of the WFNS Spine Committee regarding surgical indications and techniques related to CM1 with or without syringomyelia.

The final document, consisting of 11 statements,

provide both a practical information for surgeons dealing with this pathology, and a useful background for further international guidelines and management of CM patients.

➢ Key Points

- ❑ Surgery is recommended for CM type 1 patients who have characteristic symptoms. If an MRI shows progression in asymptomatic patients, surgery may be considered.
- ❑ In the pediatric, only osteoligamentous decompression is recommended. Adults with CM type 1 may have foramen magnum decompression with duroplasty, which is usually sufficient to control the concomitant syringomyelia. Syrinx drainage is the last option.
- ❑ Arachnoid opening can be performed in patients who have previously failed surgery or when arachnoid morphological abnormalities are discovered during the index surgery. Tonsillar shrinkage provides somewhat better clinical efficacy than decompression alone but at a larger risk of complications.
- ❑ Atlanto-axial fixation alone is recommended to treat CM1 patients with concomitant basilar invagination (BI) and atlanto-axial instability.

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