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Management of Chiari malformations: opinions from different centers—a review

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Abstract

Purpose Surgical decision-making in Chiari malformation type I (CM-I) patients tends to depend on the presence of neurological signs and symptoms, syringomyelia, and/or scoliosis, but significant variability exists from center to center. Here, we review the symptoms of CM-I in children and provide an overview of the differences in opinion regarding surgical indications, preferred surgical techniques, and measures of outcome.

Methods A review of the literature was performed to identify publications relevant to the surgical management of pediatric CM-I patients.

Results Most surgeons agree that asymptomatic patients without syringomyelia should not undergo prophylactic surgery, while symptoms of brainstem compression and/or lower cranial nerve dysfunction warrant surgery. Patients between these extremes, however, remain controversial, as does selection of the most appropriate surgical technique.

Conclusions The optimal surgical procedure for children with CM-I remains a point of contention, and widespread variability exists between and within centers.

Keywords Chiari · Duraplasty · Syringomyelia · Scoliosis

Introduction

Chiari malformations comprise a spectrum of hindbrain anomalies. The most common subtype, Chiari malformation type I (CM-I), features herniation of the cerebellar tonsils through the foramen magnum, resulting in alterations of cerebrospinal fluid (CSF) flow. Affected patients have high rates of concomitant hydrocephalus, syringomyelia, and/or scoliosis [1, 2]. CM-I may arise as an acquired condition, in which case management is straightforward and consists of treating the underlying pathology. Congenital CM-I, however, is significantly more controversial and has generated vigorous discussion regarding its optimal management. Here, we review the symptoms of CM-I in children and provide an overview of the differences in opinion among different centers regarding surgical indications, preferred surgical techniques, and measures of outcome.

Typical symptoms

Children are frequently diagnosed with CM-I during a headache workup, though the CM-I is often an unrelated, incidental finding. The headache that is associated with CM-I has several characteristic features: it is located in the occipital region, is of short duration, and is exacerbated or reproduced by a Valsalva maneuver. Other presenting symptoms may include neck, shoulder, and back pain, motor and sensory changes in the extremities, and/or difficulty with balance or coordination. Toe-walking, though typically considered a sign of tethered cord syndrome, may also be seen in the setting of CM-I. Additionally, lower cranial nerve deficits resulting in central sleep apnea and/or dysphagia may be prominent

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features, especially in infants [[3, 4]. Such symptoms may be severe on presentation, particularly when the diagnosis is delayed, and in extreme cases, patients have been referred with gastrostomy tubes already in place. We have also seen teenagers and young adults present with uncontrollable hypertension or even dysautonomia, which has completely resolved following posterior fossa decompression. The neurosurgeon should keep an open mind about atypical presentations related to CM-I while recognizing that the more atypical the clinical presentation, the less predictable the outcome from surgery.

Surgical indications

A series of publications spanning from 1991 to 2018 summarize the results of surveys administered to members of the pediatric section of the American Association of Neurological Surgeons (AANS), the American Society of Pediatric Neurosurgeons (ASPN), and the International Society of Pediatric Neurosurgeons (ISPN) [5–9]. These surveys offer some insight into the ways that different centers approach patients with CM-I, and how this has evolved over time.

Symptomatic patients

There is widespread consensus that surgical intervention is appropriate for severely symptomatic CM-I patients, particularly those with brainstem and/or lower cranial nerve dysfunction [5]. The AANS survey published in 2000 found that the vast majority of respondents recommended surgery for patients with motor or sensory loss, with slightly fewer (but still over 70%) of the respondents favoring surgery for patients with progressive pain [6]. "Minimally symptomatic" patients, such as those with isolated suboccipital headaches, are more controversial. Over time, however, surgical intervention has been increasingly embraced—46% of respondents recommended surgery in the ISPN survey published in 2004, compared with 63% in 2018 [7, 9].

Asymptomatic patients

Asymptomatic patients represent another area of consensus. With increasing numbers of patients undergoing magnetic resonance imaging, incidental CM-I is being diagnosed more frequently in asymptomatic patients, with rates ranging from 0.4 to 2.4% [10–12]. Few pediatric neurosurgeons (fewer than 10% of respondents in most surveys) recommended prophylactic surgical decompression for asymptomatic CM-I patients without syringomyelia [5–7, 9]. However, natural history studies have indicated that approximately 5–10% of asymptomatic patients ultimately become symptomatic and require surgical intervention [10, 13–15]. Therefore, regular clinical follow-up is encouraged, though routine follow-up imaging remains controversial [15].

Syringomyelia

Syringomyelia occurs in approximately 50–75% of patients with CM-I. Although prophylactic surgery is discouraged in asymptomatic CM-I patients without syringomyelia, surgical decision-making becomes more complicated when a syrinx is present. Interestingly, there has been a shift over time. Earlier surveys found that posterior fossa decompression as a primary treatment for syringomyelia was controversial, and few pediatric neurosurgeons recommended surgery for asymptomatic patients unless the syrinx demonstrated radiological progression [5, 6]. More recent surveys, however, have shown that the size of the syrinx makes a difference—although a minority recommended surgery for an asymptomatic patient with a 2-mm thoracic syrinx, ~ 75% of respondents would operate on an asymptomatic patient with an 8-mm syrinx, and this number increases to ~ 90% if the patient is minimally symptomatic [7, 9].

The surgery of choice seems to vary by region. Surveys of American pediatric neurosurgeons have shown an increasing preference for posterior fossa decompression as a primary treatment for syringomyelia in CM-I patients [8, 9]. Even among patients with persistent symptoms attributable to syringomyelia, some surgeons have advocated a second posterior fossa exploration and decompression, with an emphasis on removing occlusive arachnoid veils [16]. On the other hand, international surveys have demonstrated a preference for syringo-subarachnoid and syringo-pleural shunts, particularly for patients with persistent or progressive syringomyelia following a posterior fossa decompression [7].

Scoliosis

Scoliosis, defined as a coronal Cobb angle of greater than 10°, is identified in 25–50% of CM-I patients, and the majority of cases have an associated syrinx [16, 17]. In patients with Chiari-related scoliosis, a posterior fossa decompression may lead to stabilization and potentially improvement of the scoliosis, with the severity of the curvature being a significant predictor of outcome—patients with a curve of 40° or less are most likely to respond [16, 18, 19]. Additional intervention for scoliosis, such as bracing or surgical correction, should be delayed until after treatment of the CM-I. Survey data have indicated that the vast majority of respondents would perform a suboccipital decompression in a patient with CM-I, a substantial syrinx, a normal-level conus, and progressive scoliosis. Patients without syringomyelia and those with a low-lying conus are considered more controversial [7, 9].

Occipitocervical fusion

A subset of CM-I patients have additional radiological findings that include medullary kinking, retroflexion of the odontoid, an abnormal clival-cervical angle, assimilation of the atlas, and/or basilar invagination [20]. These have been described as "complex Chiari malformations," and unique treatment algorithms have been proposed for such patients, depending on (1) the degree of odontoid retroflexion as measured by the pBC2 distance (the maximum perpendicular distance from the posterior-superior aspect of the odontoid process to the line joining the basion to the posterior-inferior aspect of the C2 vertebral body) and (2) the clival-cervical angle. The combination of odontoid retroflexion (pBC2 distance > 9 mm) and a clival-cervical angle less than 125° is highly predictive of progressive craniocervical kyphosis and the need for occipitocervical fusion [20, 21]. Of note, patients with connective tissue disorders such as Ehlers Danlos Syndrome and Down Syndrome may be at risk for craniocervical instability both before and after posterior fossa decompression.

Surgical techniques

The surgical management of children with CM-I should have several goals, including to (1) relieve brainstem and cervical spine compression, (2) restore the flow of CSF at the foramen magnum, (3) alleviate the cranio-spinal pressure differential, (4) improve symptoms, (5) stabilize or improve syringomyelia, when present, and (6) stabilize or improve scoliosis, when present. At a minimum, surgery tends to involve a bony posterior fossa decompression (PFD) but beyond this, there are numerous variations and permutations that have been endorsed by different groups. The amount of suboccipital bony removal varies and is often accompanied by a C1 laminectomy. The dura may be left intact, scored, or opened, and some surgeons utilize intraoperative ultrasound or MRI to determine whether or not to open the dura. If opened, the arachnoid may be left intact, or an intradural dissection may be performed with lysis of arachnoid adhesions. Fourth ventricular stenting is performed by some, though plugging of the obex has fallen out of favor. The cerebellar tonsils are sometimes coagulated or resected. Ultimately, the dura may be left open, or a posterior fossa decompression and duraplasty (PFDD) may be performed using various autograft and allograft materials, including bovine, cadaveric, synthetic, pericranial, or fascia lata grafts.

The optimal surgical procedure for children with CM-I remains a point of contention, and widespread variability exists between and within centers. Surveys of members of the pediatric section of the AANS as well as members of the ASPN found that the majority of respondents favored opening of the dura, though the degree of intradural dissection was controversial. If syringomyelia was present, then respondents were slightly more likely to recommend intradural dissection [6, 8]. Interestingly, international surveys of ISPN members have indicated that although most respondents still favor opening the dura, bone-only decompressions have increased in popularity, from 1% of respondents in the early 2000s to ~ 16% more recently [7, 9]. In the experience of the senior author, bone-only decompression may be an effective option particularly for patients with concomitant genetic syndromes that result in abnormal bony development and growth at the craniocervical junction, including achondroplasia, Paget's disease, Goldenhaar syndrome, and syndromic craniosynostosis [22].

Despite the variability in surgical management, supportive data have been primarily limited to case series [23]. Two recent meta-analyses have compared PFD with PFDD in pediatric patients, with one finding no statistically significant difference between the two, and the other finding only a mild benefit of PFDD over PFD (88.1% vs. 72.3%, respectively, $p = 0.009$), with respect to rates of clinical improvement [24, 25]. Both procedures resulted in similar rates of improvement of syringomyelia and/or scoliosis, while PFDD was associated with a higher rate of CSF-related complications. The first prospective, randomized study comparing PFD and PFDD is currently underway (NCT02669836), organized by the Park-Reeve Syringomyelia Research Consortium. Subjects must be less than 21 years of age, have a Chiari 1 malformation with more than 5 mm of tonsillar herniation, and have a syrinx between 3 and 6 mm in length. Outcomes related to complications, syrinx size, and quality of life metrics will be analyzed and will provide a foundation for evidence-based surgical decision-making in the future.

Assessing outcomes

The success of surgical intervention is determined by assessing both clinical and radiological outcomes. Clinical variables include the presence of suboccipital headaches and/or neurological deficits. A review of all prior surgical series of CM-I found that 80% of the series reported subjective postoperative neurological outcomes—improvement or resolution, no change, and worsening of the preoperative neurological status [23]. Improvement or resolution of the patient's neurological status was noted in 84% of pediatric patients, while improvement or resolution of headache was noted in 88% of patients. Headaches improved more frequently in pediatric patients compared with adults. Radiological outcomes typically focus on syringomyelia. Following surgery, syringomyelia improved in 79% of the pediatric patients in the literature [23].

In a recent study examining the time to resolution of symptoms following PFDD in children with CM-I, 57% of patients with Valsalva-related headaches had resolution of their symptoms by the time they were discharged from the hospital, while all of the patients in the series were headache-free by 6 months postoperatively. Additionally, among patients with a syrinx, the syrinx was either "resolved" or "improved" in 65% of patients 3 months postoperatively, and in 79% of patients by 6 months [26].

Due to the inherent subjectivity in assessing clinical outcomes following Chiari surgery, the Chicago Chiari Outcome Scale (CCOS) was developed so that outcomes could be compared more effectively [27]. The scale quantifies postoperative outcomes in 4 categories: pain, non-pain symptoms, functionality, and complications. The score ranges from 4 to 16, with higher scores denoting more favorable outcomes. Although the CCOS has some limitations and refinements have been suggested, it has been externally validated in a pediatric neurosurgical population and may prove useful in comparing outcomes across studies in the future [28].

Conclusions

Despite decades of experience, the surgical management of children with CM-I remains highly controversial, with significant variability from center to center. At either end of the spectrum, most surgeons agree—namely, that asymptomatic patients without syringomyelia should not undergo prophylactic surgery, while symptoms of brainstem compression and/or lower cranial nerve dysfunction warrant surgery. Patients between these extremes, however, remain controversial, as does selection of the most appropriate surgical technique. There is increasing recognition that prospective, high-quality data will be necessary in order to resolve these differences; the ongoing randomized trial comparing PFD and PFDD is a good first step toward achieving this goal.

Compliance with ethical standards

Conflict of interest On behalf of all authors, the corresponding author states that there is no conflict of interest.

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References

1. Cesmebasi A, Loukas M, Hogan E, Kralovic S, Tubbs RS, Cohen-Gadol AA (2015) The Chiari malformations: a review with emphasis on anatomical traits. Clin Anat 28:184–194
2. Hankinson TC, Klimo P Jr, Feldstein NA, Anderson RC, Brockmeyer D (2007) Chiari malformations, syringohydromyelia and scoliosis. Neurosurg Clin N Am 18:549–568
3. Tubbs RS, Lyerly MJ, Loukas M, Shoja MM, Oakes WJ (2007) The pediatric Chiari I malformation: a review. Childs Nerv Syst 23: 1239–1250
4. Albert GW, Menezes AH, Hansen DR, Greenlee JD, Weinstein SL (2010) Chiari malformation type I in children younger than age 6 years: presentation and surgical outcome. J Neurosurg Pediatr 5: 554–561
5. Haines SJ, Berger M (1991) Current treatment of Chiari malformations types I and II: a survey of the pediatric section of the American Association of Neurological Surgeons. Neurosurgery 28:353–357
6. Haroun RI, Guarnieri M, Meadow JJ, Kraut M, Carson BS (2000) Current opinions for the treatment of syringomyelia and Chiari malformations: survey of the pediatric section of the American Association of Neurological Surgeons. Pediatr Neurosurg 33:311–317
7. Schijman E, Steinbok P (2004) International survey on the management of Chiari I malformation and syringomyelia. Childs Nerv Syst 20:341–348
8. Rocque BG, George TM, Kestle J, Iskandar BJ (2011) Treatment practices for Chiari malformation type I with syringomyelia: results of a survey of the American Society of Pediatric Neurosurgeons. J Neurosurg Pediatr 8:430–437
9. Singhal A, Cheong A, Steinbok P (2018) International survey on the management of Chiari 1 malformation and syringomyelia: evolving worldwide opinions. Childs Nerv Syst 34:1177–1182
10. Strahle J, Muraszko KM, Kapurch J, Bapuraj JR, Garton HJ, Maher CO (2011) Natural history of Chiari malformation type I following decision for conservative treatment. J Neurosurg Pediatr 8:214–221
11. Aitken LA, Lindan CE, Sidney S, Gupta N, Barkovich AJ, Sorel M, Wu YW (2009) Chiari type I malformation in a pediatric population. Pediatr Neurol 40:449–454
12. Meadows J, Kraut M, Guarnieri M, Haroun RI, Carson BS (2000) Asymptomatic Chiari type I malformations identified on magnetic resonance imaging. J Neurosurg 92:920–926
13. Benglis D Jr, Covington D, Bhatia R, Bhatia S, Elhammady MS, Ragheb J, Morrison G, Sandberg DI (2011) Outcomes in pediatric patients with Chiari malformation type I followed up without surgery. J Neurosurg Pediatr 7:375–379
14. Novegno F, Caldarelli M, Massa A, Chieffo D, Massimi L, Pettorini B, Tamburrini G, Di Rocco C (2008) The natural history of the Chiari type I anomaly. J Neurosurg Pediatr 2:179–187
15. Whitson WJ, Lane JR, Bauer DF, Durham SR (2015) A prospective natural history study of nonoperatively managed Chiari I malformation: does follow-up MRI surveillance alter surgical decision making? J Neurosurg Pediatr 16:159–166
16. Tubbs RS, Beckman J, Naftel RP, Chern JJ, Wellons JC 3rd, Rozzelle CJ, Blount JP, Oakes WJ (2011) Institutional experience with 500 cases of surgically treated pediatric Chiari malformation type I. J Neurosurg Pediatr 7:248–256
17. Tubbs RS, Doyle S, Conklin M, Oakes WJ (2006) Scoliosis in a child with Chiari I malformation and the absence of syringomyelia: case report and a review of the literature. Childs Nerv Syst 22: 1351–1354
18. Krieger MD, Falkinstein Y, Bowen IE, Tolo VT, McComb JG (2011) Scoliosis and Chiari malformation type I in children. J Neurosurg Pediatr 7:25–29
19. Hwang SW, Samdani AF, Jea A, Raval A, Gaughan JP, Betz RR, Cahill PJ (2012) Outcomes of Chiari I-associated scoliosis after intervention: a meta-analysis of the pediatric literature. Childs Nerv Syst 28:1213–1219
20. Brockmeyer DL, Spader HS (2015) Complex Chiari malformations in children: diagnosis and management. Neurosurg Clin N Am 26: 555–560
21. Bollo RJ, Riva-Cambrin J, Brockmeyer MM, Brockmeyer DL (2012) Complex Chiari malformations in children: an analysis of

preoperative risk factors for occipitocervical fusion. J Neurosurg Pediatr 10:134–141
22. Speer MC, Enterline DS, Mehltretter L, Hammock P, Joseph J, Dickerson M, Ellenbogen RG, Milhorat TH, Hauser MA, George TM (2003) Review article: Chiari type I malformation with or without syringomyelia: prevalence and genetics. J Genet Couns 12: 297–311
23. Arnautovic A, Splavski B, Boop FA, Arnautovic KI (2015) Pediatric and adult Chiari malformation type I surgical series 1965-2013: a review of demographics, operative treatment, and outcomes. J Neurosurg Pediatr 15:161–177
24. Durham SR, Fjeld-Olenec K (2008) Comparison of posterior fossa decompression with and without duraplasty for the surgical treatment of Chiari malformation type I in pediatric patients: a meta-analysis. J Neurosurg Pediatr 2:42–49
25. Lu VM, Phan K, Crowley SP, Daniels DJ (2017) The addition of duraplasty to posterior fossa decompression in the surgical treatment of pediatric Chiari malformation type I: a systematic review and meta-analysis of surgical and performance outcomes. J Neurosurg Pediatr 20:439–449
26. Hidalgo ET, Dastagirzada Y, Orillac C, Kvint S, North E, Bledea R, McQuinn MW, Redel-Traub G, Rodriguez C, Wisoff JH (2018) Time to resolution of symptoms after suboccipital decompression with duraplasty in children with Chiari malformation type I. World Neurosurg 117:e544–e551
27. Aliaga L, Hekman KE, Yassari R, Straus D, Luther G, Chen J, Sampat A, Frim D (2012) A novel scoring system for assessing Chiari malformation type I treatment outcomes. Neurosurgery 70: 656–664; discussion 664-655
28. Yarbrough CK, Greenberg JK, Smyth MD, Leonard JR, Park TS, Limbrick DD Jr (2014) External validation of the Chicago Chiari Outcome Scale. J Neurosurg Pediatr 13:679–684