



Opinion for different centers: surgical experience with Chiari malformation type I in children at Xuanwu Hospital, China

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Abstract

Purpose To evaluate the presenting symptoms, surgical strategy, and outcome in children with Chiari malformation type I (CM-I).

Methods The medical records of children who were diagnosed with CM-I under 14 years old and subsequently underwent surgery for CM-I between 2014 and 2018 were reviewed. The medical records for presentation, radiological image, surgical intervention, and outcome were evaluated.

Results Twelve children with CM-I and syringomyelia were included. All of the children were symptomatic. The most common presenting symptom was weakness, followed by scoliosis. All of them underwent posterior fossa decompression with/without duraplasty. Relief of preoperative pathologies and syringomyelia was experienced by all of them.

Conclusions The presenting symptoms of CM-I in children may be neurological deficits and scoliosis, which have a relationship with syringomyelia. Early recognition and a tailored operative procedure of CM-I in children could lead to good outcomes. Additional therapies for syringomyelia and scoliosis could be avoided.

Keywords Chiari malformation · Syringomyelia · Scoliosis · Posterior fossa decompression · Duraplasty

Introduction

Chiari malformations represent varying degrees of hindbrain herniation through the foramen magnum. Chiari malformation type I (CM-I), described in 1891, is characterized by ectopia or herniation of the cerebellar tonsils through the foramen magnum [1, 2]. It is increasingly recognized in children [2–10]. Patients may be asymptomatic or may have a variety of neurologic symptoms, including headache with a Valsalva-induced nature, dysarthria, dysphagia, vertigo, and ataxia. Furthermore, CM-I may lead to the development of syringomyelia, which can lead to additional neurologic deficits [2–4, 11–17]. Scoliosis is also frequently associated with CM-I [18–20]. Surgical intervention can improve existing symptoms, as well as prevent further neurologic deterioration from syringomyelia and benefit patients with CM-I [2, 3, 6, 8, 9, 12, 19, 20].

What symptoms are considered typical

Presenting symptoms

The classic clinical symptom seen in young adults with Chiari I consists of occipital pain precipitated by cough or Valsalva maneuver [11]. Other neurologic symptoms that have been described in children with Chiari I include ataxia, sensory and motor deficits, and lower cranial nerve abnormalities such as apnea and hoarseness [1–4, 6, 8, 11, 13, 21–23]. Compared with adult series, in which only 14–30% of patients are asymptomatic, as many as 37–57% of children are symptom-free at the time of initial Chiari I diagnosis [15, 24, 25]. In our study, the most common presenting symptoms were weakness, followed by scoliosis and sensory change (Table 1). Besides, all the children exhibited syringomyelia.

Displacement of one or both cerebellar tonsils 5 mm or more below the basion-opisthion line was seen in all children. Of the 12 patients, 6 (50%) had tonsils that extended more than 10 mm below the foramen magnum. Both blunt and pointed configuration were also observed. The degree of tonsillar hernia in children with pointed tonsils was greater than that of blunt hernia. The level of syrinx in children with pointed tonsils was also longer than that of blunt hernia.

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Table 1 Patient demographics, presenting symptoms, radiological features, operation features, and postoperative course of 12 children who underwent surgery for CM-I

Variable	All patients (<i>n</i> = 12)
Demographics	
Mean age (years)	8.7 + 3.1 (3–13)
Males	6 (50%)
Duration of symptoms/signs (months)	8.9 + 9.4 (0.5–24)
Presenting signs/symptoms*	
Weakness	5 (42%)
Scoliosis	4 (33%)
Sensory change	3 (25%)
Headache	3 (25%)
Ataxia	2 (17%)
Oropharyngeal symptoms	2 (17%)
Vertigo	1 (8%)
Preoperative radiological features	
Tonsillar hernia (mm)	10.8 + 5.6 (5–23)
Abnormally pointed tonsils	8
Tonsillar hernia in children with pointed tonsils (mm)	13.1 + 5.3 (8–23)
Tonsillar hernia in children with blunt tonsils (mm)	6.3 + 2.5 (5–10)
Retrocerebellar CSF compression	4
Syrinx	12
Level of syrinx	8.7 + 5.0 (2–18)
Level of syrinx in children with pointed tonsils	10.6 + 4.7 (4–18)
Level of syrinx in children with blunt tonsils	4.8 + 3.1 (2–9)
Scoliosis	4
Operation features	
PFD	2
PFDD	7
PFDD + TR	3
Operation time (min)	109.6 + 32.2 (60–151)
Blood loss (ml)	31.7 + 22.3 (20–100)
Postoperative course	
Follow-up (months)	32.4 + 19.9 (3–63)
Patients with improved syringomyelia	12
Patients with improved symptoms	12
Patients with complications	0
Symptom recurrence	0

*Many children exhibited more than one sign or symptom; hence, the total does not equal to 12

PFD posterior fossa bone decompression and partial thickness durotomy, *PFDD* posterior fossa decompression with duraplasty, *PFDD with TR* posterior fossa decompression with duraplasty and tonsil resection

Furthermore, retrocerebellar cerebrospinal fluid compression was always seen in children with abnormally pointed tonsils (Table 1). These clues may indicate that a pointed

configuration would be a more severe pathological condition compared with blunt tonsil.

Syringomyelia

Syringomyelia occurs in 50 to 76% of patients with Chiari I malformations, associating with a stepwise worsening of symptoms as the cavity enlarges or moves [2, 26]. The clinical features of syringomyelia include muscular atrophy and weakness in the upper limbs, absence of deep tendon reflexes in the upper limbs, and unilateral or bilateral sensory change [20, 27, 28]. Compared with the adult group, the children showed a lower incidence of sensory change and pain [20]. In our study, all of the children were observed with syringomyelia at the time of CM-I diagnosis. Of these children, 42% showed weakness, 25% scoliosis, and 25% sensory change.

Scoliosis

Children with CM-I and syringomyelia have a 76–82% chance of having scoliosis [18, 20, 29]. As might be expected, more children had syringomyelia than scoliosis, as it has been postulated that syringomyelia causes scoliosis due to damage sustained by the motor nuclei of the ventral horn [9, 30]. Scoliosis was not a rare finding in our series as the initial sign in children (33%). Furthermore, the children with scoliosis also have syringomyelia.

Criteria for surgical indications

Typically, CM-I in children shows a favorable natural outcome and conservative management should be considered first for asymptomatic cases [10]. In symptomatic children with CM-I, posterior fossa decompression may be indicated, whether or not syringomyelia is present [1, 2, 8, 31–33]. However, due to the variability of headache severity and the subjective quality of pain perception, mild headaches or headaches that are clearly migrainous in origin should be primarily managed medically [1, 31]. Scoliosis and symptomatic syringomyelia were also the surgical indications [8, 20, 31]. However, opinions were mixed on how to manage individuals with asymptomatic syringomyelia, as neurosurgeons perceive that syringomyelia may progress over years while some studies suggest a benign natural history [2, 8, 31, 34, 35]. We decided to perform surgery in children who were clearly symptomatic. As for children presenting with headache only, other causes of headache should be precluded. For children presenting with asymptomatic syringomyelia, we prefer a close follow-up until there is image evidence of syrinx progression or symptoms observed.

Type of surgery commonly used

The definitive treatment for symptomatic patients with CM-I remains to be the posterior fossa decompression, which can provide an improvement in symptoms and syringomyelia [2, 8, 9, 36, 37]. Proponents of posterior fossa decompression without duraplasty suggest it could achieve a satisfactory decompression with a shorter procedure time, reduced hospital stay, and reduced complication rate [37–39]. Advocates of posterior fossa decompression with duraplasty argue that a fundamental requirement of the surgery is the creation of a capacious CSF cistern at the foramen magnum and that this is better achieved with dura opening [36, 40, 41]. Some authors have also advocated tailored operative procedure by pre-operative evaluation and the use of intraoperative ultrasound [9, 42, 43].

We prefer tailored operative procedure to achieve decompression and reduce the syrinx with minimal procedure. The primary surgical treatment for each of the patients in the present series was posterior fossa decompression and partial thickness durotomy with removal of the posterior arch of C-1. Then, the need for duraplasty with autograft was evaluated intraoperatively with ultrasound according to the CSF flow and tonsil pulsation. If the dura was opened, an intradural exploration will be performed and an arachnoid web or veil impairing flow through the foramen of Magendie will be

dissected. The cerebellar tonsils would also be manipulated to make sure these will not occupy the fourth ventricle. If the cerebellar tonsils could not be accommodated in the fourth ventricle, tonsil resection or shrinkage would be done.

When the result is considered good

In children who are clearly symptomatic, studies indicate that surgical treatment of CM-I often results in either improvement or stabilization in symptoms [1, 2, 4, 8, 18, 26, 36, 44]. A complete resolution or substantial improvement in the size of syrinx cavities following posterior fossa decompression for CM-I has been reported to be 50–100% [4, 20, 21, 23, 45–48]. Children with scoliosis of more than 40° were less likely to improve with posterior fossa decompression even when there was a decrease in the size of the syrinx [8]. Thus, a scoliosis correction surgery is mandatory for them. Meanwhile, children with syringomyelia and CM-I with pre-operative Cobb angles of less than 40° will have an improvement or stabilization in scoliosis after surgery treatment of CM-I and syringomyelia [2, 8, 20, 49]. The improvement in symptom or syrinx was considered a good outcome in our series. Speaking of scoliosis, an improvement or at least no progression in the Cobb angle was expected in children whose Cobb angle was less than 35° before surgical treatment.

Fig. 1 Sagittal T2-weighted MR images obtained in a male patient who presented at 5 years of age. **a** Preoperative image demonstrating a CM-I, syrinx cavity, and basilar impression. **b** Postoperative image obtained at 3 months after surgery demonstrating a marked improvement in the size of the syrinx cavity. Note the posterior fossa decompression, C-1 laminectomy, and tonsillar resection performed at surgery. The basilar impression was also improved



Personal series

Material and methods

A total of 297 patients diagnosed with CM-I underwent posterior fossa decompression at Xuanwu Hospital of Capital Medical University between January 2014 and December 2018. A retrospective study was conducted on patients who were under 14 years old. Tailored posterior fossa decompression was performed in this group. All the children underwent magnetic resonance imaging (MRI) preoperative and at last follow-up. Standing anteroposterior and lateral 36-in. films were also obtained to evaluate scoliosis. Clinical features were also reviewed. Statistical analysis was calculated using the paired *t* test; a *p* value ≤ 0.05 was considered statistically significant.

Results

Twelve patients younger than 14 years at presentation who were admitted to our neurosurgery clinic underwent surgery for CM-I. Patient demographics, presenting signs/symptoms, preoperative radiological features, operation features, and postoperative course are summarized in Table 1.

The degree of tonsillar ectopia in children with Chiari I ranged from 5 to 23 mm (average 11.0 ± 5.6 mm). The degree of tonsillar hernia in children with pointed tonsils was greater than that in blunt hernia (13.1 ± 5.3 mm vs 6.8 ± 3.5 mm, $p = 0.04$). Syringomyelia was present in all of the children at the time of CM-I diagnosis. All syrinxes involved the cervical spinal cord, and 7 of the 12 extended into the thoracic cord. The level of syrinx in children with pointed tonsils was longer than that in blunt hernia (10.6 ± 4.7 vs 4.8 ± 3.1 , $p = 0.05$). The width of the syrinxes ranged from 4 to 13 mm (9.1 ± 2.8 mm).

All of the children had posterior fossa decompression surgery by means of the following: posterior fossa bone decompression and dural delamination (PFD) or posterior fossa decompression with duraplasty (PFDD) or posterior fossa decompression with duraplasty and tonsil resection (PFDD with TR). The improvement in preoperative symptoms and syringomyelia was seen in all of the children (Fig. 1). No complication occurred in the entire group. Recurrence of the symptoms and syringomyelia was seen in none of the children.

Four children presented with scoliosis. For all children, scoliosis affected the thoracic spine only. One of these children (Cobb angle greater than 35°) underwent surgery correction (growing rod) for scoliosis following posterior fossa decompression. The other 3 children (Cobb angle less than 35°) who underwent non-operative treatment following posterior fossa decompression had an improvement in their scoliosis until the end of the study period.

Conclusions

The CM-I in children is a complex anatomical and clinical challenge. It is usually associated with syringomyelia. The presenting symptoms in children could also have a close relationship with associated syringomyelia. However, with a tailored operative procedure, the radiological and clinical outcomes of symptomatic children could be favorable.

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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