Pediatric Chiari malformation type I: Long-term outcomes following small-bone-window posterior fossa decompression with autologous-fascia duraplasty

HAI LIU*, CHENLONG YANG*, JUN YANG and YULUN XU

Department of Neurosurgery, China National Clinical Research Center for Neurological Diseases, Beijing Tiantan Hospital, Capital Medical University, Beijing 100050, P.R. China

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Abstract. Chiari malformation type I (CM-I) is a common hindbrain disorder that is associated with deformity and elongation of the cerebellar tonsils. Although CM-I occurs in both pediatric and adult patients, its prevalence, clinical features and management in the pediatric population are not well defined. The current study evaluated a consecutive case series of 92 children (38 females and 54 males) who were diagnosed with congenital CM-I. All patients underwent small-bone-window posterior fossa decompression with autologous-fascia duraplasty. Clinical and radiological features were analyzed and long-term follow-up data were recorded. Risk factors associated with clinical outcomes were investigated using comprehensive statistical methods. Out of the 92 children, 11 (12.0%) were asymptomatic. Associated ventricular dilation was observed in 24 children (26.1%) and concomitant syringomyelia was observed in 72 children (78.3%). A total of 44 children (47.8%) showed scoliosis on plain films. Follow-up data (mean duration, 88.6 months) were available for all patients. Syringomyelia was absent or markedly reduced in 56 patients (77.8%). Symptoms were alleviated in 66 patients, remained unchanged in 12 patients and progressed in 3 patients. Statistical analysis indicated that the cerebellar tonsillar descent (CTD) grade, basilar invagination and platybasia influenced the clinical outcome ($P<0.05$). In conclusion, early recognition and surgical treatment of CM-I in pediatric patients can lead to good outcomes. The current results suggested that small-bone-window posterior fossa decompression with autologous-fascia duraplasty was an effective safe treatment option with a low complication rate. High CTD grade, basilar invagination and platybasia were indicated to be predictors of poor clinical prognosis.

Correspondence to: Professor Yulun Xu, Department of Neurosurgery, China National Clinical Research Center for Neurological Diseases, Beijing Tiantan Hospital, Capital Medical University, 6 Tiantan Xili, Beijing 100050, P.R. China
E-mail: xuhuxi@sina.com

*Contributed equally

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Introduction

Chiari malformation (CM) is a hindbrain disorder that is associated with deformity and elongation of the cerebellar tonsils. It is specifically characterized by the descent of the cerebellar tonsils >5 mm below the foramen magnum into the spinal canal (1,2). CM can be divided into four types, among which CM type I (CM-I) is the most common. Although CM-I occurs in pediatric and adult patients, the prevalence of CM-I is not well defined (2). CM-I often becomes clinically apparent when the patient is aged 20-39. Thus, in the past, CM-I had been diagnosed primarily during adolescence or adulthood (2,3). With the advent of magnetic resonance imaging (MRI), however, the number of pediatric patients diagnosed with CM-I is increasing (4). Previous studies have described the clinical features of CM-I in pediatric populations (2,5,6).

CM-I is usually associated with ventricular dilation, syringomyelia and scoliosis, but the relevant pathogenesis is not clearly understood. Moreover, the incidence of these conditions co-presenting with CM-I has varied considerably in previous studies (7-10).

Management of CM-I and concomitant syringomyelia and scoliosis in children remains controversial. Some authors have recommended syringoperitoneal shunting for treating syringomyelia (11,12). The treatment of scoliosis consists of serial observation, bracing and/or corrective spinal surgery. It is yet to be established whether aggressive surgical treatment is necessary, and the ideal therapeutic option is undetermined (13). It is widely accepted, however, that posterior fossa decompression should be included in the treatment protocol for CM-I as it can lead to both clinical and radiological improvement (8,14,15). The use of specific surgical procedures and related issues, however, remain controversial; these include the use of craniectomy, the size of the bone window, whether the arachnoid should be opened and whether a cerebellar tonsil should be manipulated (14,15).

The current study evaluated a consecutive case series of 92 children who were diagnosed with CM-I. Their clinical manifestations, concomitant ventricular dilation, syringomyelia, basilar invagination, platybasia and scoliosis were analyzed. The study also evaluated the surgical outcomes of small-bone-window posterior fossa decompression (SPFD) with autologous-fascia duraplasty (AFD) in the treatment of pediatric CM-I.

Patients and methods

Patient recruitment. A search of medical records was conducted to identify pediatric patients (aged 0-18 years at their initial presentation to the neurosurgery clinic) with CM-I who had been operated on in the Department of Neurosurgery at Beijing Tiantan Hospital, Capital Medical University (Beijing, China). The cohort included 92 children (female:male ratio, 1.00:1.42; mean age ± standard deviation, 10.0±4.5) with congenital CM-I who were treated with SPFD + AFD between January 2001 and January 2015. CM-I was diagnosed according to the following criteria (1): i) Herniation of tonsils ≥5 mm was below the plane of the foramen magnum; or ii) tonsillar herniation of 3-5 mm accompanied by other CM-I features, such as syringomyelia. Exclusion criteria were as follows: i) Occipitocervical instability; ii) congenital spinal bifida; iii) diastematomyelia; iv) concurrent intraspinal tumors or myelitis; v) acquired CM-I as a complication of cerebrospinal fluid (CSF) diversion procedures or chronic CSF leakage; and vi) incomplete data. The study protocol was approved by the Institutional Review Board and Ethics Committee of Beijing Tiantan Hospital, Capital Medical University.

Clinical presentation. Subjective clinical symptoms and physical examinations were documented. Symptoms recorded included headache, foramen magnum nerve compression symptoms (occipitocervical headaches, neck or shoulder pain), sensory disturbance, motor dysfunction (weakness or muscle atrophy), lower cranial nerve dysfunction (dysphagia, hoarseness or coughing), cerebellar syndrome (truncal and appendicular ataxia) and scoliosis. Some patients also had bradycardia. Following the exclusion of endocrine dysfunction and organic heart disease by comprehensive evaluations, including laboratory, electrophysiological and radiological examinations, these symptoms were suspected to be associated with CM-I or with concomitant syringomyelia.

Imaging. Preoperative plain radiographs and perioperative MRI scans of the cervical spine were available for all patients. The descending distance between the inferior pole of the cerebellar tonsil and the level of the foramen magnum was measured on sagittal T1-weighted images in a picture archiving and communication system workstation (OsiriX software; version 6.0; Pixmeo SARL, Bernex, Switzerland). The degree of cerebellar descent was classified into three groups according to the cerebellar tonsillar descent (CTD) grading system (16): Grade I, the tonsil descends over the foramen magnum but does not reach the C1 arch; Grade II, the tonsil descends to the C1 arch level; and Grade III, the tonsil descends below the C1 arch. The location of the syringomyelia was determined on sagittal MRI images, and its size was determined by measuring the longitudinal and transverse diameters.

The data for concomitant scoliosis were collected from standing posteroanterior radiographs (Fig. 1), and coronal curvature measurements were performed according to the Cobb method (17). Scoliosis was defined as a Cobb's angle of ≥10˚. Curve severity was defined as follows: mild, 10-25˚; moderate, 26-40˚; severe, ≥41˚. In cases of S-shaped scoliosis, the major curve was selected for analysis and the minor curves were considered compensatory.

Other associated abnormalities were also recorded, such as ventricular dilation, platybasia and basilar invagination. Ventricular dilation was defined by an Evans' index of >0.30 (18).

Surgical approach. Surgery was performed on all children with a combined strategy that included SPFD and AFD. After patients were positioned left-laterally, a posterior midline skin incision was made from 1 cm below the inion to the spinous process of C4. An autologous graft (2x2 cm) was resected from the fascia and reserved. The SPFD procedure, which included a small-bone-window suboccipital craniectomy (diameter, 2.0-2.5 cm) and C-1 laminectomy (1.5 cm), was performed. The thick constraining dural band found at the occipitocervical junction was resected. When the dura mater was opened, the surgeon aimed to keep the arachnoid intact. The dura mater was subsequently grafted with the autologous graft to enlarge the cisterna magna. Six children presented with progressive hydrocephalus and severe intracranial hypertension, and an emergency ventriculoperitoneal shunt was implanted in these cases (Fig. 2).

Follow-up. Follow-up data for all children were available in their records. The data had been obtained during individual office visits or telephone interviews, with a mean follow-up time of 88.6 months (standard deviation, 46.2; range, 10-166 months). Clinical outcomes were qualitatively categorized as follows: i) Improved, the patients experienced partial or complete relief of their chief complaints; or ii) not improved, no notable change in their chief complaints, or deterioration of their clinical status. Furthermore, the clinical outcomes were quantitatively evaluated according to the Chicago Chiari Outcome Scale (CCOS; Table I) (19).

Statistical analysis. SPSS 22.0 software (IBM SPSS, Armonk, NY, USA) was used for statistical analyses. Chi-squared test, continuity correction test or Fisher's exact probability test were used to screen the potential risk factors, including age at onset, age at diagnosis, sex, duration of symptoms, CTD grades, concomitant syringomyelia, hydrocephalus, basilar invagination, platybasia and scoliosis. Logistic regression analysis was performed to identify the risk factors for clinical outcomes (binary-classification qualitative data). Furthermore, to verify the association between potential risk factors and clinical outcomes, CCOS scores (continuous variables) were compared using one-way analysis of variance (ANOVA) with a Bonferroni post hoc test test for multi-group comparisons and Mann-Whitney U test for two-group comparison. $P \leq 0.05$ was considered to indicate a statistically significant significance.

Table I. Chicago Chiari outcome scale.

	Score			
Characteristic	1	2	3	4
Pain	Worse	Unchanged and refractory to medication	Improved or controlled with medication	Resolved
Non-pain	Worse	Unchanged or improved but impaired	Improved and unimpaired	Resolved
Functionality	Unable to attend	Moderate impairment (<50% attendance)	Mild impairment (>50% attendance)	Fully functional
Complication	Persistent complication, poorly controlled	Persistent complication, well-controlled	Transient complication	Uncomplicated course

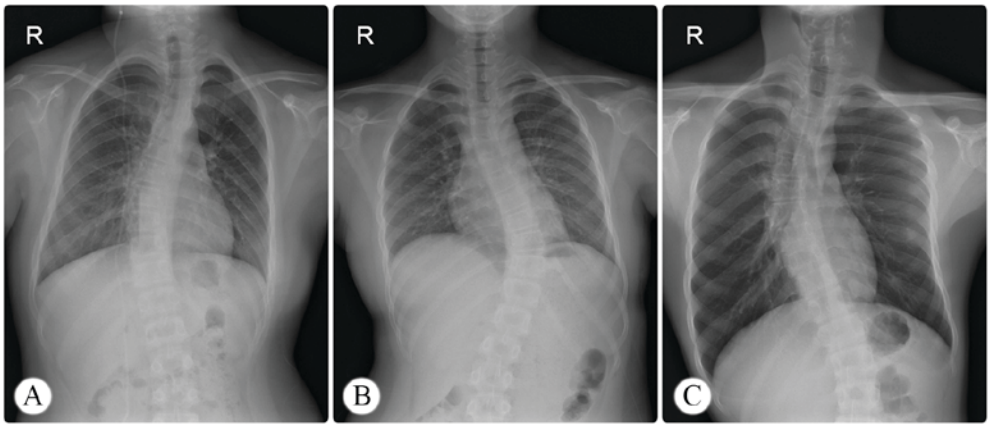

Figure 1. Plain radiographs of 3 pediatric patients with scoliosis. (A) Right thoracic curve and ventriculoperitoneal shunt. Cobb's angle is 32˚. (B) Left thoracic curve, with a Cobb's angle of 40˚. (C) Right thoracic curve, with a Cobb's angle of 47˚. R, right-hand side of the patient.

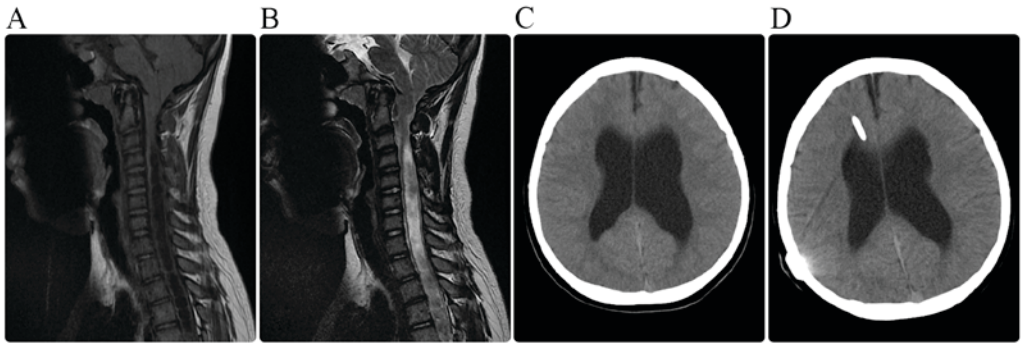

Figure 2. Preoperative sagittal MRI scans of a female patient. (A) T1-weighted and (B) T2-weighted MRI scans indicate tonsillar herniation and a cervicothoracic syringomyelia. This patient's scoliosis is presented in Fig. 1A. (C) On admission, a head computed tomography scan indicated hydrocephalus. (D) An emergency ventriculoperitoneal shunt was implanted. MRI, magnetic resonance imaging.

Results

Patient population and clinical characteristics. Over the 14-year span of this study, 98 pediatric patients were diagnosed with CM-I and underwent SPFD + AFD. Six patients were lost to follow-up and excluded. This case series consisted of 38 females and 54 males, with a female:male ratio of 1.00:1.42. The average age at the time of surgery was 10.0±4.5 years (range, 1-18 years). In total, 11 (12.0%) children were asymptomatic. In the symptomatic group (n=81), the duration of symptoms preceding the initial diagnosis ranged from 2 weeks to 4 years (mean, 16.8 months). A total of 59 children presented with headache (72.8% of all symptomatic children), 17 with foramen magnum nerve compression symptoms (21.0%), 29 with sensory disturbance (35.8%), 36 with motor dysfunction (44.4%), 7 with lower cranial nerve dysfunction (8.6%) and 16 with cerebellar syndrome (19.8%). In addition, the results of electrocardiography, which is performed routinely upon

Table II. Characteristics of coexisting scoliosis.

Characteristic	Number	Percentage (%)
Sex		
Male	25	56.8
Female	19	43.2
Curve type		
C-shaped thoracic scoliosis	35	79.5
S-shaped thoracic scoliosis	8	18.2
S-shaped thoracolumbar scoliosis	1	2.3
Curve orientation		
Convex to the left	20	45.5
Convex to the right	24	54.5
Severity		
Mild (10-25˚)	26	59.1
Moderate (26-40˚)	12	27.3
Severe (>41˚)	6	13.6
Total	44	100

admission, indicated that 4 patients had atrioventricular block (3 patients with second-degree and 1 patient with third-degree block), with no relevant underlying disease detected. None of the children complained of bowel or bladder dysfunction.

Preoperative MRI. The distance of tonsillar herniation ranged from 6.0 to 18.4 mm (mean, 11.2±3.2 mm). On the preoperative MRI, platybasia was observed in 5 children (5.4%) and basilar invagination in 12 children (13.0%). Associated ventricular dilation was observed in 24 children (26.1%). Concomitant syringomyelia was observed in 72 children (78.3%): Cervical syringomyelia (n=4; 5.6% of concomitant syringomyelia cases), cervicothoracic syringomyelia (n=67; 93.0%) and holocord syringomyelia (n=1; 1.4%).

Scoliosis evaluation. A total of 44 children (47.8%; 19 females and 25 males) showed scoliosis on plain films: 35 children had C-shaped thoracic scoliosis in a single curve; 8 children had S-shaped thoracic scoliosis with two adjacent curves; and 1 child had S-shaped thoracolumbar scoliosis. According to the severity classification, 26 children were diagnosed with mild scoliosis, 12 with moderate scoliosis and 6 with severe scoliosis. Further analysis indicated that all children with scoliosis had concomitant syringomyelia. In total, 36 (81.8%) of the children with scoliosis also had a unilateral sensory disturbance and/or motor disturbance or unilateral muscle atrophy. These data are summarized in Table II.

Complications and follow-up. There were no deaths in this series. Central nervous system infection occurred postoperatively in four children. Antibiotics were prescribed and the infections were effectively treated. Suboccipital hydrops was noted in two children (Fig. 3). During the follow-up period,

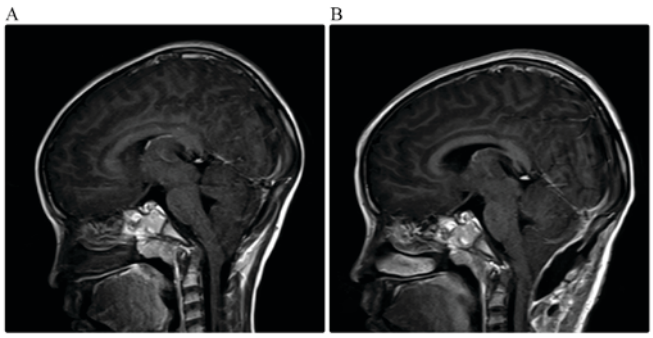

Figure 3. Sagittal T1-weighted MRI scans of a male patient. (A) A preoperative MRI scan indicates tonsillar herniation. During the follow-up period, the patient's symptoms were unchanged. (B) A follow-up MRI scan indicates suboccipital hydrops. MRI, magnetic resonance imaging.

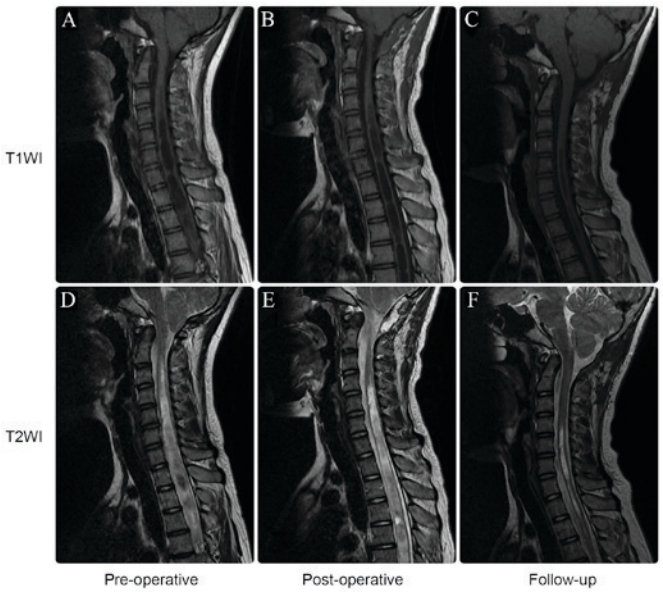

Figure 4. Sagittal MRI scans of a female patient. Preoperative sagittal (A) T1-weighted and (B) T2-weighted MRI scans reveal tonsillar herniation and cervicothoracic syringomyelia. The patient's scoliosis is presented in Fig. 1B. (C and D) Immediately postoperative MRI scans indicate reconstruction of the cisterna magna and an upward shift of the tonsils, but no marked change in the syringomyelia. (E and F) Follow-up MRI scans at 6 months after small-bone-window posterior fossa decompression with autologous-fascia duraplasty indicate a marked reduction of the syringomyelia. MRI, magnetic resonance imaging; T1WI, T1-weighted image; T2WI, T2-weighted image.

symptoms were alleviated in 66 patients (81.5% of all the symptomatic children), remained unchanged in 12 patients (14.8%) and progressed in 3 patients (3.7%). Preoperatively, 72 patients had concomitant syringomyelia. According to follow-up MRI scans, syringomyelia was absent or markedly reduced in 56 of these patients (77.8%; Fig. 4). Analysis of these results using Chi-squared tests indicated that CTD grades and the incidence of basilar invagination (Fig. 5) and platybasia were associated with clinical outcomes ($P<0.05$); the statistical data are summarized in Table III. Logistic regression analysis indicated that a high CTD grade [P=0.016; odds ratio (OR)=3.675; 95% confidence interval (CI), 1.138-8.527], basilar invagination (P=0.008; OR=3.489; 95% CI, 1.048-10.754) or platybasia (P=0.002; OR=3.981;

Table III. Results of Chi-squared tests.

Characteristic	Chi-square value	P-value
Age at onset (<10 vs. >10 years)	0.305	0.581
Age at diagnosis (<10 vs. >10 years)	0.007	0.932
Sex (male vs. female)	0.268	0.605
Duration (<16.8 vs. >16.8 months)	1.522	0.217
CTD (Grade I-III)	14.224	0.001
Syringomyelia (present vs. absent)	0.683[a]	0.408[a]
Hydrocephalus (present vs. absent)	1.658[a]	0.198[a]
Basilar invagination (present vs. absent)	11.864[a]	0.001[a]
Platybasia (present vs. absent)	-	0.004[b]
Scoliosis (present vs. absent)	1.897	0.168

[a]Continuity correction. [b]Fisher's exact probability. CTD, cerebellar tonsillar descent.

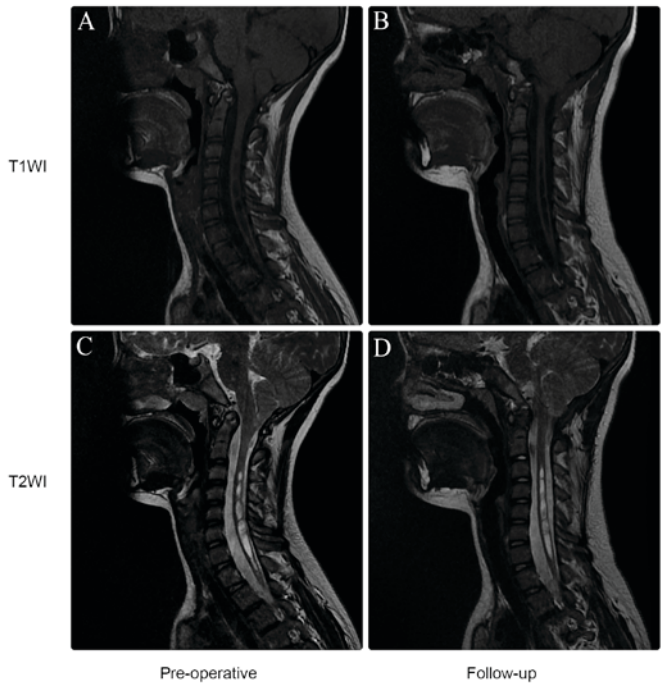

Figure 5. Sagittal MRI scans of a male patient. Preoperative sagittal (A) T1-weighted and (B) T2-weighted MRI scans reveal tonsillar herniation, basilar invagination and cervical syringomyelia. The patient's scoliosis is presented in Fig. 1C. (C and D) MRI scans at 28 months after small-bone-window posterior fossa decompression with autologous-fascia duraplasty show an upward shift of the tonsils and slight reduction of syringomyelia.

95% CI, 1.654-9.386) significantly increased the likelihood of poor clinical outcomes. A comparison of CCOS scores by ANOVA also indicated that CTD grade, and basilar invagination and platybasia by Mann-Whitney U tests are predictors of poor clinical prognosis (P<0.05; Table IV).

Discussion

CM-I is a craniocervical junction disorder characterized by a cerebellar tonsil descending below the foramen magnum into the spinal canal. This was first described by Hans Chiari in 1891 (1). The incidence of CM-I has been reported to be 0.5-3.5% in the general population (20). A previous study reported a slight female predominance (female/male ratio 1.3:1.0) (20), although this is inconsistent with the findings in the current study (female/male ratio 1.00:1.52). CM-I is generally considered a congenital neurological condition, although in recent years its acquired form has been identified as a complication of CSF diversion procedures or chronic CSF leakage (21,22).

CM-I is thought to be a multifactorial condition, although the pathogenesis is still undetermined. A well-accepted hypothesis is that the hindbrain tissues are dislocated into the spinal canal because of an overcrowded posterior cranial fossa, due to underdevelopment of the mesodermal occipital somite (23). Previous morphometric results have supported this postulation. The posterior fossa volume in CM-I patients was reported to be smaller compared with the control group, although the difference was not statistically significant (23,24). In addition, some familial cases suggest a genetic component (25).

The incidence of syringomyelia in CM-I patients has ranged widely in previous reports, from 30 to 70% (20). The exact prevalence of syringomyelia in pediatric patients remains unclear. In the current study, a high syringomyelia occurrence rate (78.3%) was observed in CM-I patients. There are several hypotheses concerning the development of syringomyelia in the CM-I context, including the 'water-hammer' mechanism proposed by Gardner (26) and the 'pressure dissociation' mechanism proposed by Williams (27). However, most authors agree that partial obstruction in the foramen magnum area, which blocks the normal circulation of CSF, is the pacing factor in the development of syringomyelia (28).

Diagnosis of CM-I depends on MRI (29). For a long time, the diagnostic criteria have been under debate. Barkovich *et al* (30) proposed that 3 mm below the foramen magnum was the lowest extent of tonsillar descent in normal patients, and defined CM-I as >3 mm of tonsillar descent. Others have proposed that 5 mm of tonsillar ectopia should be adopted as the cutoff for diagnosing CM-I (2). In addition, Mikulis *et al* (31) reported that age affects the normal position of cerebellar tonsils and proposed age-based diagnostic criteria. These were as follows: In patients aged 0-9, the cutoff distance for tonsillar descent should be 6 mm; in patients aged 10-29, it should be 5 mm; in patients aged 30-79 years it should be 4 mm; and at >79 years of age, it should be 3 mm (31). Considering the fact that patients with lesser degrees of tonsillar ectopia (3-5 mm) may also develop classic neurological symptoms and syringomyelia that are amenable to neurosurgical intervention, the current study adopted the more flexible comprehensive diagnostic criteria proposed by Tubbs *et al* (1).

Patients with CM-I can be asymptomatic or can present with a variety of signs and symptoms ranging from headache to severe myelopathy and brain stem compression (29). With the increasing availability of diagnostic MRI, more asymptomatic patients are being identified (4). In the adult population, 15-30% of CM-I patients are asymptomatic (20). In the current study, 11 (12.0%) asymptomatic children were identified. Innate bias, however, is inevitable in an in-hospital cohort. Sleep apnea and feeding problems are reported to be more common in pediatric patients, although other clinical manifestations appear to be similar in different age groups (6). As previously reported, the

Table IV. Results of statistical analyses.

	CTD (Grade I-III)		Basilar invagination (present vs. absent)		Platybasia (present vs. absent)	
Clinical CCOS assessment	F value	P-value	Z value	P-value	Z value	P-value
Composite CCOS score	3.580	0.032	-4.927	0.000	-3.601	0.000
Pain subscore	0.139	0.871	-6.215	0.000	-3.996	0.000
Non-pain subscore	3.459	0.036	-3.629	0.000	-3.451	0.001
Functionality subscore	3.942	0.023	-3.578	0.000	-4.125	0.000
Complications subscore	0.984	0.378	-1.177	0.239	-1.573	0.116

CTD, cerebellar tonsil descent; CCOS, Chicago Chiari outcome scale.

most common presenting symptom of CM-I is headache (4), which is consistent with the current findings. In infants and children who are unable to communicate verbally, headaches may manifest as crying and irritability. Other common symptoms include non-radicular pain in the shoulder, back and extremities, motor and sensory disturbances, clumsiness, ataxia and lower cranial nerve dysfunction (29). In the current cohort, two patients with atrioventricular block were also identified.

There is no effective non-surgical strategy for treating patients with CM-I (12). Posterior fossa decompression, with or without dural opening, and with or without arachnoid opening or dissection, is most commonly used for the surgical treatment of CM-I and can change its clinical course (13,32). However, the size of the bone window required for posterior fossa decompression is still under debate (33,34). Furthermore, it has not yet been established whether a difference exists in the operative techniques used for pediatric and adult patients. Large-bone-window posterior fossa decompression effectively enlarges the posterior cranial fossa and therefore relieves symptoms (35). However, there may be a higher incidence of complications, such as CSF leak, pseudocysts, meningitis and hydrocephalus (35). Furthermore, excessive removal of the squamous part of the occipital bone could lead to downward and backward displacement of the cerebellum and brain stem, leading to long-term complications. According to a previous study, small-bone-window craniectomy is enough to achieve favorable clinical outcomes, and the incidence of complications is reduced (35). The procedure described in the aforementioned study relieves the compression of the occipitocervical bones and preserves enough bone to support the cerebellum and brain stem. In the current study, all patients underwent a small-bone-window approach, with which high rates of clinical (81.5%) and radiological (77.8%) improvement were achieved.

Whether it is necessary to open the arachnoid space is another focus of controversy. Some authors assert that CSF-related complications, such as CSF leak or meningitis, are directly associated with opening the arachnoid (36). A previous study has indicated that the incidence of CSF leaks is not higher in the arachnoid-opening group, provided that watertight duraplasty is performed (37). Furthermore, reconstruction of the cisterna magna and tonsillar manipulation could contribute to a better prognosis (37). It is the current authors' opinion that arachnoid opening is unnecessary in most patients; SPFD and duraplasty without arachnoid incision is sufficient to relieve compression and alleviate clinical symptoms. It should be noted that the incidence of CSF-related complications was low in the pediatric cohort of the current study.

The prevalence of scoliosis in the general pediatric population is 2-4%, while in children with CM-I the prevalence of scoliosis is >4-fold increase (9,38). Progressive scoliosis is a relatively common manifestation of CM-I when there is coexistent syringomyelia (38). In the current study, the incidence of scoliosis was markedly higher (47.8%) in CM-I patients compared with the reported prevalence in the general pediatric population. Nokes *et al* (39) identified 3 patients with CM-I and scoliosis, but without syringomyelia. In the current cohort, all scoliotic children had coexistent syringomyelia. Considering the relatively mild severity of pediatric scoliosis, spinal orthopedic surgery was not recommended for the majority of these patients. It was considered that brace treatment could effectively reverse scoliotic progression. Eule *et al* (40) conducted a 20-year review of surgical and nonsurgical treatment in a pediatric cohort with CM-I associated with syringomyelia and scoliosis. It was found that early decompression of CM-I helped stabilize or alleviate the associated scoliosis, avoiding the requirement for orthopedic spinal surgery.

The current study had several limitations. Preoperatively, the scoliosis evaluations were based only on standing posteroanterior radiographs. The absence of side-bending radiographs limited a three-dimensional assessment. During the follow-up period, only MRI scans were performed. Plain radiography was not performed in all of the patients. Therefore, the present study did not track the progression of scoliosis. Furthermore, for ethical reasons, there were no control groups undergoing large-bone-window posterior fossa decompression or arachnoid opening in the current study.

In conclusion, early recognition and surgical treatment of pediatric CM-I leads to good outcomes in the majority of patients. SPFD with duraplasty was demonstrated to be an effective, safe treatment option with a low complication rate. High CTD grade, basilar invagination and platybasia were indicated to be predictors of poor clinical prognosis.

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