Analysis of Clinical and Radiographic Outcomes of the Angle between Clivus and Supraocciput in Patients with Chiari's Malformation Type I Following Surgical Decompression

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Abstract

Background The objective of this study was to investigate whether different angles between the clivus and supraocciput (C–SO angle) in patients with Chiari's malformation type I (CMI) after posterior fossa decompression lead to different clinical and radiographic outcomes.

Methods A total of 73 patients who underwent surgical decompression at our institution between 2010 and 2016 were retrospectively divided into two groups: group A, with an angle less than 96 degrees, and group B, with an angle ≥ 96 degrees. Patient clinical outcomes were determined using the Chicago Chiari Outcome Scale (CCOS), and radiographic outcomes were determined by changes in the syrinx size before and after surgery. Direct comparisons were made between the two groups.

Results There were no statistically significant differences between the two groups regarding demographics, preoperative symptoms or radiographic characteristics ($p > 0.05$), except for the cerebellar tonsillar descent ($p \leq 0.001$). The mean overall CCOS score was 13.11 ± 1.99 . The total CCOS score was significantly different between the two patient groups ($p < 0.05$). Moreover, group A demonstrated significantly better postoperative improvements than group B in clinical outcome measures based on the CCOS system ($p = 0.035$). For radiographic outcomes, the change in the syrinx between the two groups was also significantly different, with a value $p = 0.024$.

Conclusions A direct comparison between the two groups revealed that a smaller C–OS angle (< 96 degrees) was related to unsatisfactory clinical and radiographic outcomes. Thus, the C–OS angle may emerge as a predictor of clinical or radiographic outcomes following surgical decompression in patients with CMI.

Keywords

- Chiari's malformation type I
- c–os angle
- syringomyelia
- Chicago Chiari outcome scale

Introduction

Chiari's malformation type I (CMI) is characterized by caudal displacement of the cerebellar tonsils through the foramen

magnum into the upper cervical canal which is associated with syringomyelia.^{1,2} So far, no correlation has been identified between the extent of tonsillar herniation and the severity of clinical symptoms.³ Most people believe that the

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pathogenesis of CMI is the result of hindbrain herniation caused by overcrowding of the posterior fossa.⁴ Thus, surgical decompression is the most commonly suitable approach for CMI which can lead to both clinical and radiologic improvement. Surgical approaches are mainly divided into posterior fossa decompression with or without duraplasty (PFDD or PFD, respectively). There has always been controversy over the merits of both approaches. Lin et al performed a meta-analysis to evaluate the advantages of the two strategies and believed that PFDD can be an optimal surgical method due to its higher clinical improvement and lower recurrence rate and that PFD can be a preferred choice because of its similar clinical improvement and lower costs in patients without syringomyelia.⁵ However, unfavorable outcomes have been reported in approximately 30% of patients, perhaps suggesting that not everyone is suitable for surgical decompression.^{6,7}

Most studies have indeed proven that the posterior cranial fossa (PCF) in patients with CMI is smaller than in normal individuals as measured by morphometric analysis of magnetic resonance images (MRI). Moreover, Basaran et al found in their analysis that the radiological features of the posterior fossa were different among the subtypes of CM.⁸ Among Basaran et al's measurements, we are interested in the angle between the clivus and supra-occiput (C–SO angle) because it determines whether the shape of the posterior fossa is similar to a funnel. In particular, we would like to know whether the C–SO angle is related to surgical outcomes. To identify the reason for unfavorable outcomes in some patients, we retrospectively studied clinical data from 73 CMI patients who had undergone decompression operations from 2010 to 2016.

Materials and Methods

Patients and Data Collection

This retrospective study included patients with CMI who received decompression surgery between 2010 and 2016 at the Neurosurgery Department of Peking Union Medical College Hospital (PUMCH). Informed consent was obtained from each patient before surgery. We excluded patients with other types of Chiari's malformations, spinal deformities, craniofacial junction malformation (CJM), scoliosis, atlantooccipital fusion, atlantoaxial dislocation, basilar impression, and so on. The inclusion criteria were (1) age above 18 years; (2) CMI with syringomyelia on MRI examination; and (3) absence of other bony malformation, CJM, skull or spinal trauma, tumor, or surgery. A total of 73 cases met the inclusion criteria and were reviewed in this study. All patients underwent PFDD and their medical records and radiographic characteristics including MRI were reviewed and compared. This study protocol was approved by the institutional review boards at PUMCH and Peking Union Medical College.

We collected each patient's data including age at surgery, gender, symptom duration, and preoperative symptoms. Preoperative symptoms were divided into three main categories according to the Chicago Chiari Outcome Scale (CCOS)

as follows: pain symptoms, nonpain symptoms, and functionality conditions.⁹ Pain symptoms included neck and back pain, headache, and upper extremity pain. It is noteworthy that we also included nonpain symptoms, such as sensory symptoms, motor symptoms, cranial nerve symptoms, and cerebellar-related symptoms. Sensory symptoms included sensory loss, numbness, and paresthesia; motor symptoms included muscle weakness, atrophy, and tremor; cranial nerve symptoms included dysphagia, hoarseness, and bucking; and cerebellar-related symptoms included dizziness, ataxia, and tinnitus. Preoperative functionality conditions were classified as nonfunctional, moderately impaired, mildly impaired, and fully functional. This functionality mainly affected daily life, including combing the hair and holding objects, buckling, and lifting the arms.

All patients underwent preoperative MRI where the following parameters were recorded: the cerebellar tonsillar descent (CTD), location of the syrinx and the C–SO angle (►Fig. 1). According to Basaran et al's research, the mean C–SO angle is 96.317 ± 10.674 degrees for CMI patients.⁸ Thus, patients can be grouped based on their C–SO angle. Patients were divided into two groups: group A, with an angle less than 96 degrees, and group B, with an angle ≥ 96 degrees (►Fig. 2). There were 35 subjects in group A (24 females, 11 males) and 38 in group B (28 females, 10 males).

Surgical Procedure

PFDD was performed on all patients. All patients were administered general anesthesia. A midline incision was made extending from the occipital protuberance to the C2 spinous process which exposed the edge of the occipital bone, atlantoaxial posterior arch, lamina, and spinous process. Then, we removed the inferior part of the occipital bone, the posterior lamina of C1, and the tip of the C2 spinous process to achieve a bony decompression (approximately 2×3 cm). After opening the dura in a Y-shaped fashion, we could see the lower pole of the cerebellar tonsil and the brain pulsate. Then, the occipital fascia was sutured for dural grafting. Finally, the outer layers were sutured step-by-step to achieve anatomical reduction.

Follow-up

We assessed clinical outcomes at 1-year follow-up visits through outpatient clinics or telephone interviews. All patients underwent follow-up MRI. The radiographic outcomes were evaluated through the changes in syrinx size before and after surgery. The syrinx changes were assessed by measuring the maximal anteroposterior diameter of the syrinx and the syrinx length, and an improvement was defined as a $> 20\%$ decrease in the maximal syrinx diameter or syrinx length on follow-up MRI.^{10,11}

The clinical outcomes were evaluated based on CCOS scoring which included categories for pain symptoms, nonpain symptoms, functionality, and complications. Component scores from 1 to 4 were summed for a total score from 4 to 16: score categories of 4–8, 9–12, and 13–16 were indicative of patients whose outcomes were poor, good, or excellent from baseline, respectively. Furthermore,

Fig. 1 The angle between clivus and supraocciput (C–So angle) is constructed by drawing a line along the clivus and another line along the supraocciput.

Fig. 2 The example of group A patient and group B patient. (A) showed a patient's C–So angle is 105 degrees which is an example of group B. (B) showed a patient's C–So angle is 85 degrees which is an example of group A.

significant complications were recorded which included chemical meningitis, cerebrospinal fluid (CSF) leak, wound infection, and deep vein thrombosis. The CCOS scores and syrinx measurement were evaluated by two surgeons.

Statistical Analysis

All statistical analyses were performed using statistical software (SPSS Version 22.0). Comparisons between the two groups were analyzed using χ^2 or Fisher's exact test for categorical data and two independent sample Student's *t*-tests for continuous data. The Wilcoxon's signed rank test was used for the analysis between the outcomes and the C–So angle. *P*-Values < 0.05 were considered statistically significant.

Results

Preoperative Characteristics

This study included 73 patients with CMI at the PUMCH from 2010 to 2016. This cohort comprised 52 females (71.23%) and 21 males (28.77%), with a mean age of 40.91 ± 11.40 years at surgery. ▶Table 1 shows the patient demographic information. No significant differences were noted between the two patient groups in terms of age, gender, or symptom duration. The symptoms were classified into three main groups: pain symptoms, nonpain symptoms, and functionality conditions. The three most common symptoms were sensory loss (69.86%), numbness (64.38%), and muscle weakness (53.42%). No significant difference was observed in the preoperative symptoms experienced by each patient group ($p > 0.05$). The above results can be seen in ▶Table 2. In addition, ▶Table 3 summarizes the radiographic characteristics. A significant difference was found in cerebellar tonsillar descent (CTD) and C–So angle between the two groups. No significant difference was found in the location of the syrinx according to our statistical analyses ($p > 0.05$).

Table 1 Comparison of basic demographics between the two groups

Demographics	Group A (< 96 degrees), n = 35	Group B (≥96 degrees), n = 38	All patients, n = 73	p-Value
Sex				
Male	11	10	21	0.630 ^a
Female	24	28	52	
Symptom duration (y)	5.48 ± 7.14	4.37 ± 5.88	4.86 ± 6.50	0.066 ^b
Mean age (y)	36.94 ± 8.96	44.13 ± 9.67	40.91 ± 11.40	0.084 ^b

^aIndicates the χ^2 or Fisher's exact test.^bIndicates the two independent samples Student *t*-test.

Note: Values are means ± standard deviations.

Table 2 Summary of the preoperative symptoms

Presenting symptoms		Group A (< 96 degrees), n = 35	Group B (≥96 degrees), n = 38	All patients, n = 73	p-Value ^a
Pain symptoms					
Headache		5	9	14	0.308
Neck and back pain		15	20	35	0.404
Upper extremity pain		7	16	23	0.042
Nonpain symptoms					
Sensory symptoms	Sensory loss	24	27	51	0.817
	Numbness	23	24	47	0.749
	paresthesia	2	1	3	1.000
Motor symptoms	Muscle weakness	22	17	39	0.121
	Muscle atrophy	10	8	18	0.248
	Muscle tremor	1	0	1	0.967
Cranial nerve symptoms	Dysphagia	0	1	1	1.000
	Hoarseness	2	1	3	0.942
	Bucking	3	2	5	0.924
Cerebellar related symptoms	Dizziness	5	9	14	0.308
	Ataxia	9	6	14	0.294
	Tinnitus	0	1	1	1.000
Functionality conditions					
Unable to attend		0	0	0	0.801
Moderate impairment (< 50%)		6	4	10	
Mild impairment (> 50%)		3	6	9	
Fully functional		25	29	54	
Complications		12	8	20	0.205

^ap-Value is used the χ^2 or Fisher's exact test.

Table 3 Radiographic characteristics of the two groups

Preoperative MRI	Group A (< 96 degrees), <i>n</i> = 35	Group B (≥96 degrees), <i>n</i> = 38	<i>p</i> -Value
Cerebellar tonsillar descent	10.65 ± 4.09	11.20 ± 7.61	< 0.001 ^a
Location of the syrinx			
Cervical	10	12	0.780 ^b
Beyond cervical	25	26	
C–SO angle	86.06 ± 6.73	110.16 ± 10.15	< 0.001 ^a

Abbreviations: C–So angle, clivus–supraocciput angle; MRI, magnetic resonance imaging.

^aIndicates the two independent samples Student's *t*-test.

^bIndicates the χ^2 or Fisher's exact test.

Note: values are means ± standard deviations.

Table 4 Component and the total Chicago Chiari outcome scale scores

Component	Group A (< 96 degrees), <i>n</i> = 35	Group B (≥96 degrees), <i>n</i> = 38	All patients, <i>n</i> = 73	<i>p</i> -Value
Pain symptoms score	3.23 ± 0.83	3.16 ± 0.78	3.22 ± 0.84	0.572
Nonpain symptoms score	2.83 ± 0.84	3.08 ± 0.66	2.93 ± 0.76	0.051
Functionality score	3.49 ± 0.87	3.66 ± 0.66	3.57 ± 0.77	0.037
Complications score	3.29 ± 0.85	3.45 ± 0.68	3.35 ± 0.78	0.043
Total score	12.80 ± 2.07	13.45 ± 1.80	13.11 ± 1.99	< 0.001

Note: values are means ± standard deviations. *p*-Value is used the two independent samples Student's *t*-test.

Complications

No patient had a reoperation. The most common complication was chemical meningitis which often manifested as fever and headache that responded to steroids. The overall rate of complications in group A was 34.29% (12/35) compared with 21.05% (8/38) in group B. Other rare complications included CSF leak, wound infection, and subcutaneous hydrops. There were no differences in complications between the two groups (►Table 2).

Outcomes

The average follow-up time of clinical and radiographic outcome for each of the group was 1-year through outpatient clinics and MRI. Clinical outcome scores based on CCOS criteria are summarized in ►Table 4 for each group. The mean overall CCOS score was 13.11 ± 1.99. The mean score of group B was 13.45 ± 1.80 which showed clinical improvement and a clear benefit following surgical decompression. However, the mean score of group A was < 13. Nonpain symptoms, functionality, and complications were statistically significantly different between the two groups but not pain symptoms. Moreover, clinical outcomes were significantly different between the two groups (*p* < 0.05). The Wilcoxon's signed rank test analysis of the two groups showed a significant difference in the outcome results (excellent, good, and poor), with a value *p* = 0.035 as shown in ►Table 5. Moreover, as for radiographic outcomes, the change of syrinx between the two groups was also significantly different, with a value *p* = 0.024 (►Table 6). Thus, we can conclude that C–SO angle may influence the outcome of decompression surgery.

Discussion

CMI is a congenital disorder, usually associated with syringomyelia. There are many theories about the pathogenesis of CM but none of them explain the disease completely.

Most scholars hold the view that CMI is the result of abnormal embryonic development, resulting in the formation of a small posterior fossa.^{12,13} Therefore, adequate posterior fossa decompression has been widely advocated for reducing syrinx progression but persistence of syringomyelia is generally considered a treatment failure, and the highest reported incidence of persistent syringomyelia was 40%.^{14–16} To explore the reasons for treatment failure, the present study compared the surgical efficacy of Chiari's decompression between patients with a C–OS angle above and below 96 degrees. This is the first study comparing such imaging measures to clinical and radiographic outcomes.

Table 5 Clinical outcome measures between the two groups

Outcomes	Group A (< 96 degrees), <i>n</i> = 35	Group B (≥96 degrees), <i>n</i> = 38	<i>p</i> -Value
Excellent results	21	31	0.035
Good results	12	7	
Poor results	2	0	

Note: *p*-value is used the Wilcoxon's signed rank test. *p*-Value (0.035) < 0.05, which is statistically significant.

Table 6 The change of syrinx between the two groups

The change of syrinx	Group A (< 96 degrees) $n = 35$	Group B (≥ 96 degrees) $n = 38$	p -Value
Improvement	12	24	0.024
Unimprovement	23	14	

Note: p -value is used the χ^2 or Fisher's exact test.

The C-OS angle between the clivus and supraocciput was first discussed by Basaran et al.⁸ They found that the C-OS angle was narrower than in the control group for all patients. Moreover, they demonstrated that an increase in the angle was a risk factor for syringomyelia formation. However, the mechanism behind this is not understood. Thus, by comparing different C-OS angles, this study aimed to determine whether the angle can affect posterior fossa decompression.

Our study compared the clinical and radiographic outcomes analyzed by C-OS angle of 73 CMI patients who underwent PFDD. The mean age of the patients with CMI was 40.91 ± 11.40 years, and the female/male ratio was 2.48. The most frequent symptoms were nonpain symptoms, including sensory loss and numbness. The outcome measurements were based on the CCOS score and the changes in syrinx size. In this study, there was a statistically significant difference with regards to the CCOS scores, including the nonpain, functionality and complication scores, between our two CMI patient groups ($\rightarrow$ Table 4). There was a statistically significant difference between clinical outcomes (evaluated as excellent, good, or poor) and radiographic outcomes when comparing the two groups ($\rightarrow$ Tables 5 and 6). Our study is the first to publish this kind of comparison.

Based on our data analysis, we concluded that the C-OS angle may affect the outcomes of posterior fossa decompression. But why? We speculated that the C-OS angle may affect the morphology of the posterior fossa and the outlet of the skull base. A smaller C-OS angle may have a smaller outlet of the basis cranii due to its funnel-like shape. Therefore, more cerebellar tonsils will be herniated, as shown on our preoperative MRI ($\rightarrow$ Table 3). Performing PFDD for such patients will not solve these problems and cerebellar tonsils will continue to herniate. Dufton et al proved that a wider foramen magnum diameter is associated with a greater tonsillar descent.¹⁷ Thus, the decompression effect may not be ideal. In addition, when there is a larger C-OS angle, the cerebellar tonsils may happen to herniate the outlet and caused syringomyelia because of piston-like pressure in the lower hernia body and downward CSF pressure waves.¹⁸ Therefore, PFDD will reduce the syrinx size. However, there is no sufficient theoretical basis for the above discussion, and thus, further investigations need to be conducted. Based on our analysis, we suggested no surgery at all for the C-OS < 96 degrees. Further researches are needed to determine the optimal treatment or some other operation.

Our study raises the question of whether surgery is suitable for all patients. Operations on patients with a

smaller C-OS angle need to be carefully considered. Based on our findings that a smaller C-OS angle (< 96 degrees) is related to a relatively low CCOS score and a slow change in the syrinx, it is appropriate to apply the C-OS angle as a novel prognostic indicator and recommend that patients with a smaller C-OS angle have a poor prognosis after PFDD.

Limitations of our study should also be noted. First, this is a retrospective study that has all of the limitations of such a design. Second, the imaging measured data were limited in scope, and we need to measure other morphometric features of the posterior cranial fossa to investigate whether such a difference can influence treatment outcomes. Third, we only know that a smaller C-OS angle was correlated with syrinx severity and low CCOS score. However, the reason why a smaller C-OS angle leads to unsatisfactory clinical results is worth exploring further. Finally, in this study, we only included 73 patients. Larger cohorts and more detailed subgroups should be included in our following studies to explore this potential relationship. Further study is needed to determine the optimal extent of bony decompression.

Conclusion

Different C-OS angles were identified in CMI patients with the presence of a syrinx in this study. The C-OS angle was related to clinical improvement outcomes based on CCOS scoring and radiographic improvement of decreasing the syrinx. Based on our data, a smaller C-OS angle (< 96 degrees) was related to unsatisfactory clinical and radiographic outcomes. Thus, assessment of the C-OS angle before operation is helpful in the formulation of surgical strategies and prediction of postoperative improvement of CSF circulation and syrinx outcomes.

Therefore, more trials are needed to corroborate these findings.

Author Contributions

Authors Y.L. and X.W. designed the research; authors X.W., J.G., S.H., and Z.L. performed the research; authors Y.L. and J.G. collected the data; X.W. analyzed and prepared the manuscript; besides, all authors reviewed and commented on the manuscript.

Ethics Approval and Consent to Participate

Not applicable.

Consent for Publication

The manuscript has not been published previously, in any language, in whole or in part, and is not currently under consideration elsewhere.

Competing Interests

The authors declare that they have no competing interests.

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Conflict of Interest

None declared.

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