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Posterior fossa decompression and duraplasty with and without tonsillar resection for the treatment of adult Chiari malformation type I and syringomyelia

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Abstract

Background: The current surgical management of adult Chiari malformation type I (CM-I) with associated syringomyelia remains controversial. The objective of this study was to explore posterior fossa decompression and duraplasty (PFDD) with and without tonsillar resection in adult patients with CM-I and syringomyelia.

Methods: A total of 116 adult patients suffering from both CM-I and syringomyelia who were scheduled to undergo surgical decompression at our institution between 2012 and 2020 were randomly divided into 2 groups: the PFDD group (n = 64) underwent PFDD without tonsillar resection, while the PFDD-T group (n = 52) underwent PFDD with tonsillar resection. The primary outcome was improvement or resolution of the syrinx. The secondary outcome was an improvement in clinical outcome based on Chicago Chiari Outcome Scale (CCOS) scores. All participants were followed-up to 1-year postoperatively.

Results: The proportions of patients who had >20% improvement in syrinx size were 60.9% and 78.8% in the PFDD and PFDD-T groups, respectively (P = .038). The improvement in clinical outcome based on CCOS scores was significantly different in the 2 groups (P = .004). The functionality sub-score was significantly different between the 2 groups (P = .027), but there were no significant differences in the pain symptoms, non-pain symptoms, and complications sub-scores. The total CCOS scores were higher in the PFDD-T group than in the PFDD group (P = .037).

Conclusion: This study determined the role of tonsillar resection in achieving obvious syrinx improvement following PFDD-T. PFDD with tonsillar resection seems to be a safe and effective surgical option to treat adult CM-I patients with syringomyelia.

Abbreviations: CCOS = chicago Chiari outcome scale, CM-I = Chiari malformation type I, CSF = cerebrospinal fluid, PFDD = posterior fossa decompression and duraplasty.

Keywords: Chiari malformation decompression, Chicago Chiari outcome scale, syrinx improvement, tonsillar resection

1. Introduction

Chiari malformation type I (CM-I) consists of posterior fossa anomalies that generally share the feature of cerebellar tonsillar descent through the foramen magnum.[1] Syringomyelia is a fluid-filled cavity within the spinal cord that occurs secondary to a variety of etiologies, including tumor, trauma, arachnoiditis, or, as in the present study, CM-I.[2]

Typically, the surgical treatment for CM-I includes enlargement of the posterior cranial fossa.[1,3] The decision to treat CM-I surgically is based on the severity and progression of signs and symptoms, as well as magnetic resonance imaging and cerebrospinal fluid (CSF) flow findings. Nevertheless, there is considerable controversy around which surgical technique should be used.[4,5] While some specialists advocate bone-only craniocervical decompression, others recommend posterior fossa decompression and duraplasty (PFDD), and still others recommend bone decompression and duraplasty with tonsillar resection.[3] As a result, a rigorous comparison of the outcomes of various surgical techniques remains unavailable, as most studies to date have examined small, retrospective cohorts.[6,7]

The goal of this trial was to compare 2 standard decompression procedures used to treat CM-I, by conducting a prospective,

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Written informed consent was obtained from all patients.

The authors have no conflicts of interest to disclose.

The datasets generated during and/or analyzed during the current study are available from the corresponding author on reasonable request.

This study was approved by the Ethics Committee of the Cangzhou Central Hospital.

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surgical cohort comparison study of patients with CM-I and syringomyelia: PFDD without tonsillar resection and PFDD with tonsillar resection (PFDD-T).

2. Methods

This was a prospective cohort study for patients with CM-I and syringomyelia that compared the outcomes of PFDD with and without tonsillar resection. The study was conducted between April 2012 and May 2020 in accordance with the Declaration of Helsinki and was approved by the Ethics Committee of Cangzhou Central Hospital.

2.1. Participants

Inclusion criteria included: aged more than 18 years; a CM-I with cerebellar tonsil position at least 5 mm below the foramen magnum; and presence of syringomyelia. Exclusion criteria included one or more of the following preexisting conditions: hydrocephalus; tumor; brain trauma; stroke; meningitis; abscess; other congenital anomalies of the brain or skull base; vertebral anomalies; degenerative or demyelinating disease; and prior posterior fossa surgery. Syringomyelia is a chronic progressive disease of the spinal cord, which is characterized by abnormal fluid-filled cavities and glial (non-nerve cell) proliferation in the spinal cord, often occurring in the cervical spinal cord. Informed written consent to participate in the study was obtained from each patient prior to surgery.

2.2. Randomized controlled trial

Randomized controlled trial is a means of testing the effect of a therapy or drug in medical and health services, especially used in medicine, pharmacy, and nursing research.[8] The design of randomized controlled trials follows 3 basic principles, namely, the randomization of research subjects, the setting of a control group, and the use of blinding. Through randomized allocation, the qualified research objects were divided into experimental group and control group, make the test factors in as much as possible to keep balance between groups, and then accept the corresponding experiment, 1 group gives to assess test interventions, another group to control intervention treatment, follow-up period, is the end of the 2 groups, scientifically evaluate some measures of clinical effect.[9]

2.3. Sample size calculation

$$n = \frac{Z^2\sigma^2}{d^2},$$

where Z is the confidence interval, n is the sample size, d is the sampling error range, and σ is the standard deviation, which is generally taken as 0.5.

2.4. Procedures

All patients were administered general anesthesia. A midline incision was made, extending from the occipital protuberance to the C2 spinous process. Then, suboccipital craniectomy, C1 and, occasionally, C2 laminectomy were performed to achieve a bony decompression. After opening the dura in a Y-shaped fashion, the lower pole of the cerebellar tonsil and the brain pulsate could be seen. Then, the occipital fascia was sutured for dural grafting to achieve duraplasty. The difference was that the PFDD group received no manipulation of the cerebellar tonsils, while the PFDD-T group received tonsillar resection. Finally, the outer layers were sutured step-by-step to achieve anatomical reduction.

2.5. Follow-up and data collection

Standardized forms were used to collect data relating to patient demographics, radiological assessments, surgical details, and clinical follow-ups. Data collected included pre- and post-operative signs and symptoms, pre- and post-operative magnetic resonance imaging scans of the brain and entire spine, details of the surgical intervention, and details of any postoperative complications. The comparison between the 2 groups was made based on the outcomes of the total number of patients seen at 1-year follow-up visits through scheduled outpatient interviews.

The primary outcome was the improvement or resolution of the syrinx, defined as >20% improvement in length, maximal cross-sectional diameter, or both. The secondary outcome was the improvement in clinical outcomes based on patients' Chicago Chiari Outcome Scale (CCOS) scores. The clinical outcomes were evaluated based on CCOS scores that included categories for pain symptoms, non-pain symptoms, functionality, and complications.[10] Component scores from 1 to 4 were summed for a total score of between 4 and 16: score categories of 4 to 8, 9 to 12, and 13 to 16 were indicative of patients whose outcomes were poor, good, or excellent from baseline, respectively. The CCOS assesses the outcomes of various symptoms that patients may have in a more comprehensive way. CCOS was then applied to the patients and the predictors of prognosis were analyzed in more detail using CCOS.[11]

2.6. Statistical analysis

For quantitative variables, data are expressed as the mean (SD), and normality was tested with using the Shapiro–Wilk tests. When obeying a normally distribution, data were analyzed using the Student *t*-tests. For categorical variables, data are expressed as number (percentage) and were analyzed using the chi-square tests or Fisher exact tests based on expected cell counts, with the exception of ordered multi-categorical variables, which were analyzed using the Mann-Whitney *U* tests. A 2-sided *P* value < .05 was considered statistically significant. Statistical analysis was performed using IBM SPSS Statistics 21.0 (IBM, Armonk, NY).

3. Results

A total of 140 patients were recruited, with 116 included in the analysis (Fig. 1). Table 1 shows the baseline demographic and clinical data by assigned group. There were no statistically significant differences between the 2 groups in any variables at baseline.

The primary outcome chosen when planning this study was >20% syrinx improvement. The proportions of patients who had >20% improvement in syrinx size were 60.9% and 78.8% in the PFDD and PFDD-T groups, respectively (P = .038). The proportions of patients who showed any improvement in syrinx size were 68.8% and 88.5% in the PFDD and PFDD-T groups, respectively (P = .011; Table 2).

For the secondary outcome of clinical outcome improvement based on CCOS scores, there were significant differences between the 2 groups (P = .004; Table 2). The patients' CCOS scores are shown in Table 3. The functionality sub-score was significantly different between the 2 groups (P = .027) but there were no significant differences between pain symptoms, non-pain symptoms, and complications sub-scores. The total CCOS scores were higher in the PFDD-T group than in the PFDD group (P = .037).

One patient in the PFDD-T group experienced CSF leak, while 2 patients in the PFDD group experienced CSF leaks. There were no wound problems, pseudomeningoceles, or neurological deficits. No patient underwent repeat surgery within the 1-year follow-up period. There were no differences in complications between the 2 groups.

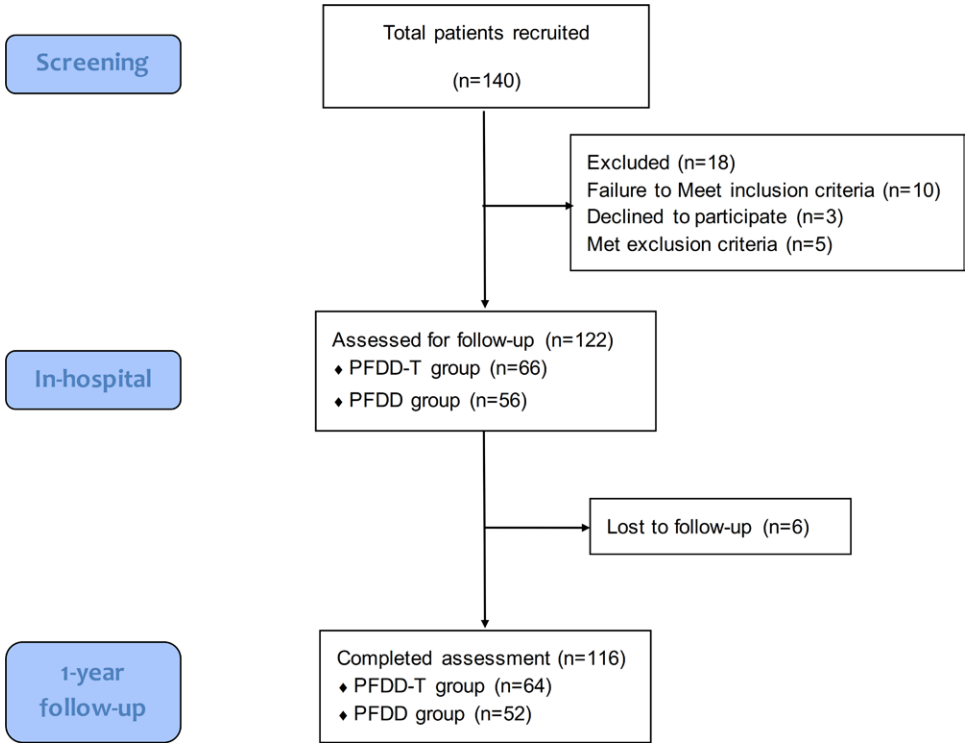

Figure 1. A total of 140 patients were recruited, with 116 included in the analysis.

Table 1

Baseline demographic and clinical characteristics of study participants.

Characteristic	PFDD group (n = 64)	PFDD-T group (n = 52)	*P* value
Age (yr.)	41 ± 8.9	42 ± 8.4	.876
Gender (male/female)	27/37	20/32	.684
Symptom duration (yr.)	4.9 ± 2.4	5.3 ± 2.8	.491
Measurements on MRI			
Tonsillar descent (mm)	9.1 ± 3.4	9.7 ± 2.9	.752
Syrinx length (mm)	129 ± 55	134 ± 58	.288
Maximal diameter of syrinx (mm)	8.2 ± 3.6	8.6 ± 3.9	.344

Data are presented as mean ± SD or number.

MRI = magnetic resonance imaging, PFDD = posterior fossa decompression and duraplasty, PFDD-T = PFDD with tonsillar resection.

Table 2

Syrinx and clinical improvement by the 1-year follow-up.

Outcomes	PFDD group (n = 64)	PFDD-T group (n = 52)	*P* value
Syrinx improvement			
>20% improvement	39 (60.9%)	41 (78.8%)	.038
Any improvement	44 (68.8%)	46 (88.5%)	.011
CCOS			
Excellent	40	45	.004
Good	22	6	
Poor	2	1	

Data are presented as number (percentage).

CCOS = Chicago Chiari Outcome Scale, PFDD = posterior fossa decompression and duraplasty, PFDD-T = PFDD with tonsillar resection.

4. Discussion

This prospective cohort study evaluated the outcomes of PFDD with and without tonsillar resection for the treatment of adult patients with CM-I and syringomyelia. The results confirmed the effectiveness and safety of PFDD-T, in terms of comparable neurological and syringomyelia outcomes.

CM-I is a congenital disorder that is usually associated with syringomyelia.[4] There are many theories about the pathogenesis of CM-I but none of them can completely explain the disease. CM-I is the result of abnormal embryonic development, resulting in the formation of a small posterior fossa.[2] Therefore, adequate posterior fossa decompression has been

Table 3

Component and total Chicago Chiari Outcome Scale scores by the 1-year follow-up.

Categories	PFDD group (n = 64)	PFDD-T group (n = 52)	*P* value
Pain score	3.2 ± 0.78	3.3 ± 0.84	.303
Non-pain score	3.3 ± 0.81	3.4 ± 0.87	.106
Functionality score	3.5 ± 0.77	3.8 ± 0.88	.027
Complications score	3.5 ± 0.78	3.6 ± 0.85	.146
Total scores	13.5 ± 1.99	14.2 ± 2.08	.037

Data are presented as mean ± SD.

PFDD = posterior fossa decompression and duraplasty, PFDD-T = PFDD with tonsillar resection.

widely advocated for reducing syrinx progression; however, the persistence of syringomyelia is generally considered to be a treatment failure. Adults have more symptoms than children, and a longer clinical presentation, which may suggest a poorer prognosis. Radiographic improvements, as defined by a reduction in syrinx diameter, are reported to appear 10 months after patients experience clinical improvement, and some clinicians see such improvements as an absolute way to define a successful procedure.[12,13] The improvement or resolution of the syrinx was selected here as the primary outcome measure to evaluate the effectiveness of CM-I decompression procedures. Clinical improvement following surgery in patients with CM-I is one of the most important outcome measures. The CCOS was also used in our study to clearly quantify the clinical outcomes.

The present study did not address the technique of bone-only posterior fossa decompression, which has become more popular in the past several years. Patients with syringomyelia are more likely to improve following PFDD compared with patients who receive posterior fossa decompression, and this may be related to better reconstruction of the flow dynamics at the craniocervical junction.[14] However, PFDD is also associated with a higher rate of overall complications. Advocates of tonsillar resection consider it a safe procedure that ensures optimal expansion of the foramen magnum and patency of the CSF pathways, with low rates of complications.[15–17] Moreover, tonsillectomy has been used to mitigate tissue volume mismatch, especially in pediatric cases, where the advantage of amelioration of the risk of syrinx collapse and spine deformity were observed.[18] Our study confirmed that syrinx and clinical improvements by the time of 1-year follow-up were improved in the PFDD-T group compared with the PFDD group. It is presumed that because chronic compression renders the tonsils atrophic and gliotic, removing them adds no neurological injury.[15,17] Consistent with this finding, we also found that there was no statistically significant difference between the PFDD and PFDD-T groups in terms of clinical complications. Although cerebellar tonsillar resection is not associated with discernible neurological deficits, it is likely that all brain regions exhibit distinct neurological functions. The cerebellum contains nearly 80% of all intracranial neurons, which implies that it is likely to play a vital functional role, possibly as a coordinating center.[19] Future advances in diagnostic technology may allow subtle neurological sequelae of tonsillar injury to be detected.

There are several limitations to our study. Although the sample size in the current study was larger than in most previous studies, the conclusion was drawn from a limited sample size. The experience from a single medical center may limit the universality of our findings.

Our study showed there was a significant impact on imaging and clinical 1-year outcomes when cerebellar tonsillar resection was added to PFDD in the treatment of CM-I with syringomyelia. This study highlighted the role of tonsillar resection in achieving obvious syrinx improvement following PFDD-T.

Author contributions

Conceptualization: Ming Yang.
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