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Comparison decompression by duraplasty or cerebellar tonsillectomy for Chiari malformation-I complicated with syringomyelia

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ABSTRACT

Objective: The current study aimed to assess the two surgical procedures of posterior fossa decompression (PDF) in treating Chiari malformation type I (CM-1) complicated by syringomyelia (SM), and to evaluate the postoperative complications, surgical effects and prognosis.

Patients and methods: A retrospective study was performed on 115 adult CM-I patients undergoing surgical treatment from November 2013 to November 2016 in a single comprehensive hospital. These patients underwent the surgical procedure of either posterior fossa decompression with duraplasty (PFDD) or posterior fossa decompression combined with the resection of tonsils (PFDRT) by five experienced neurosurgeon in a single center. The clinical outcomes of these two surgical procedures were evaluated through comparing the clinical data before and 6 months after the operation.

Results: A total of 115 patients, including 35 men and 80 women with the mean age of 43.4 ± 10.1 years (range, 16–60 years), were enrolled in the current study. 37 out of the 115 patients underwent PFDD, while the remaining 78 received PFDRT according to the surgical assessment. The operation time in PFDRT group (159.3 ± 40.0 min) was higher than that in PFDD group (134.1 ± 30.8 min) ($P < 0.05$). Besides, 20 cases in PFDRT group (20/78) developed postoperative dizziness and headache, and such incidence was higher than that in PFDD group (3/37) ($P < 0.05$). After 6 months of follow-up, a total of 69 patients (88.4%) in PFDRT group had alleviated symptoms, while 31 (83.8%) patients in PFDD group had improved symptoms. Altogether, the SM cavity was reduced in 54 patients (69.2%) in PFDRT group and 29 (78.4%) in PFDD group, respectively, after the operation. No statistical differences in symptom improvement and cavity reduction rate could be witnessed between the two groups.

Conclusion: Our study suggests that both PFDRT and PFDD can achieve comparable short-term clinical outcomes for adult CM-I patients. Surgical treatment is considered to be a reliable choice for the treatment of adult CM-I patients. Typically, PFDRT may lead to a higher risk of aseptic inflammatory complication. The precise surgical procedure should be selected based on detailed conditions of patients.

1. Introduction

Chiari malformation (CM) was first described in 1891 to be a caudal displacement of the cerebellar tonsils through the foramen magnum [1]. Hans Chiari classified these malformations into three types including Chiari malformations I, II,and III, and added the Chiari IV malformation after four years. Researchers have been encouraged to propose new classifications to encompass the variants (e.g., Chiari 0, Chiari1.5, and Chiari 3.5 malformations) [2]. In all types CM-I is the most common one in the clinic. About 60%–80% CM-I patients are accompanying with syringomyelia (SM) [3]. Upon definition, the cerebellar tonsils are at least 5 mm below the foramen magnum level. Cerebellar tonsils herniation may lead to distinct clinical symptoms, including pain, altered sensation, weakness, dysphagia, sleep apnea, sensory deficit, extremity weakness, and atrophy.

Magnetic resonance imaging (MRI) has been used to diagnose SM and explore its pathophysiology. However, its etiology and pathogenesis have not yet been fully illustrated. Gardner [4] postulated that obstructed cerebrospinal fluid (CSF) outflow from the cisterna magna into the spinal subarachnoid space would lead to hydrodynamic shocks of the CSF systolic wave from the fifth ventricle to the central canal walls, thus resulting in canal expansion and formation of a syringomyelic cavity. Oldfield [5] found that the progression of syringomyelia associated with Chiari I is produced by the action of the

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cerebellar tonsils, which partially occlude the subarachnoid space at the foramen magnum and act as a piston on the partially enclosed spinal subarachnoid space. He hypothesized that enlarged cervical subarachnoid pressure waves result in the migration of fluid through the perivascular spaces of the spinal cord, forming a syrinx. Moreover, Nishikawa et al. [6] had measured the posterior fossa parameters, and revealed that an underdeveloped occipital bone would induce overcrowding in the posterior cranial fossa, which contained the normally developed hindbrain in adult-type CM.

The main purpose of surgical treatment is to restore CSF circulation in craniovertebral junction. Many different treatment modalities have been studied and proposed at present, however, no consensus has been reached so far on the surgical treatment of Chiari malformation type I (CM-I) [7]. Notably, foramen magnum decompression is a well-established treatment for CM-I. However, it remains controversial on whether posterior fossa decompression combined with resection tonsils (PFDRT) or posterior fossa decompression with duraplasty (PFDD) is superior. In this study, we had compared the surgical results of posterior fossa decompression, aiming to assess the therapeutic outcomes and to answer some questions.

2. Material and methods

2.1. General information

A total of 115 patients, including 35 males and 80 females, were enrolled in this study. 78 of these 115 patients underwent PFDRT while the remaining 37 received PFDD from November 2013 to November 2016 in the department of the First Affiliated Hospital of Zhengzhou University. The diagnosis of CM I-SM in all patients was confirmed by MRI, CT and X-rays of the cervical spine. Concretely, the surgical indications were radiologic diagnosis of CM and SM, as well as the presence of progressive disabling symptoms. Patients with other types of CM, spinal cord cavity caused by spinal trauma, tuberculosis, tumor, and tethered cord were excluded. In the meantime, patients with atlantoaxial dislocation, as well as Goel [8] A type skull base depression which basilar invagination combined with fixed atlantoaxial dislocation, the odontoid tip falls into the foramen magnum. They required internal fixation were also ruled out from the current study.

The course of disease in patients ranged from 2 weeks to 20 years, with an average of 47.8 months. At the same time, the age of patients ranged from 10 to 60 years, with an average of 43.4 years.

2.2. Clinical symptoms

Symptoms related to damage in the central canal of spinal cord were found in 115 patients. Additionally, separable sensory disorders in limbs or bodies, as well as muscle atrophy in hands or upper extremities were found in 49 patients. Symptoms of nerve root irritation were found in 58 patients, including pain, as well as burning sensations in neck, shoulders, back, or upper extremities. Meanwhile, symptoms related to damage of lower cranial nerves and cerebellum were also found, including unsteady gait in 25 and dysphagia in 4 patients, respectively. Besides, symptoms related to damage of pyramidal tract could be observed in 9 patients, including increase in muscle tone, hyperreflexia, and muscle weakness. Additionally, symptoms associated with increased intracranial pressure were discovered in 24 patients, including headache, vomiting, and papilledema.

2.3. Imaging examination

All cases were diagnosed based on MRI in the craniospinal junction and cervical spine. As could be found on MRI, the cerebellar tonsils had protruded through the foramen magnum by more than 5 mm in all patients. Additionally, all patients underwent MR scanning of brain and the entire spinal cord using the 1.5 T MR imaging system (SIEMENS)

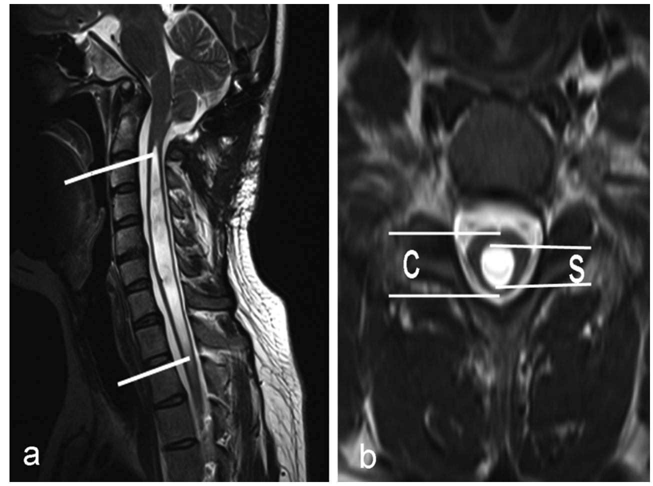

Fig. 1. Indexes used to describe the feature of syrinx (A patient diagnosis of Chiari malformation MR also showed brain stem herniation). (a) Syrinx length was measured by vertebral segments spanned by the syrinx. (b) The diameter of the syrinx and spinal cord wear measured where the syrinx was the widest.

described on the midline transect/sagittal T1/T2-weighted images. The degree of cerebellar tonsillar descent configuration and length of syrinx. The maximal anteroposterior diameters of the cervical and thoratic syrinx, as well as the spinal cord were measured, and the maximal syrinx/cord ratio was calculated (Fig. 1). Typically, a 20% [9] decrease in syrinx size represented a significant radiographic improvement. Such values were independently assessed by two investigators, including a certified radiologist with the standard software system used at our hospital. At the same time, the depth of tonsillar herniation ranged from 5 to 20 mm, with an average of 8.6 ± 4.1 mm. SM was restricted to the cervical spinal cord in 26 patients, in the cervical and thoracic spinal cord in 85 patients, and in the whole spinal cord in 4 patients. The ratio of SM diameter to spinal cord was less than 1/3 in 6 patients, 1/3- 2/3 in 46 patients, and more than 2/3 in 63 patients. A total of 30 patients had complications of basilar invagination, 27 of occipitalization, 34 of scoliosis, and 10 of Charcot's joints.

2.4. Surgical approaches

The surgical procedure PFDRT or PFDD was chosen by five experienced neurosurgeon in a single center on the basis of comprehensive assessment of preoperative patients and intraoperative conditions (If the fluctuation of the dura mater is not obvious when removed after removing the bone plate, internal decompression is performed. All patients were under general anesthesia placed in the prone position. Their heads were fixed and necks were bent slightly forward. The skin, subcutaneous tissues, and occipital and paraspinal muscle were cut through with a midline incision extending from the occipital protuberance to the C2 spinous process.

The incision exposed the edge of the occipital bone, atlantoaxial posterior arch, spinous process, and lamina. The inferior part of the occipitalbone was removed made a bone window (approximately 3 cm × 4 cm).The posterior arch of C1, and the tip of the spinous process of C2 were removed to achieve a bony decompression.

After decompression, a thick fasciculation-like tissue that compressed the dura was observed. The thick tissue was removed, and the dura was incised carefully under a microscope in a Y-pattern. After opening of the dura, dissection of arachnoid scars, coagulation or subpial resection of the cerebellar tonsils to facilitate normal passage of CSF out of the IVth ventricle, opening of the foramen of Magendie. After rinse the cavity carefully and suture the dura step-by-step.

The patients in PFDD group was characterized by a dural grafting performed with artificial dura without subdural incision of arachnoid

Table 1
Neurological Scoring System.

Score	Pain	Sensory Dist., Dysesthesias	Motor Weakness	Gait Ataxia
5	None	Normal	Full power	Normal
4	Slight no medication	Present not significant	Movement against resistance	Unsteady no aid
3	Good control with medication	Significant function not restricted	Movement against gravity	Mobile with aid
2	Insufficient control with medication	Some restriction of function	Movement without gravity	Few steps with aid
1	Severe despite medication	Severe restriction of function	Contraction without movement	Standing with aid
0	Pain	Incapacitated function	Plegia	Plegia

membrane. Then suture the outer layers step-by-step to achieve anatomical reduction. Finally, incisions were carefully sutured, and wearing a neck collar 3 months.

2.5. Preoperative and postoperative evaluation indexes

To evaluate the therapeutic outcomes, neurological and medical imaging examinations were performed. The general patient data and specific features before surgery and 6 months after surgery were recorded, respectively. Meanwhile, the clinical outcomes were evaluated using a scoring system, including gait, movement deficit, sensory changes and pain [10] (Table 1). The signs and symptoms were rated as "improved", "unchanged", and "deteriorated".

MRI examination was performed preoperatively and during the follow-up visit. The syrinx cavity solution was recorded (Figs. 2 and 3). Meanwhile, the postoperative state of syringomyelia cavity was also recorded. Postoperative state of patients was classified as 'stabilized', 'improved' or 'worsened', as suggested in the previous studies [11].

2.6. Statistical analysis

All statistical analyses were performed using the statistical software (SPSS Version 17.0, SPSS). The continuous variables were expressed as means ± standard deviation. Continuous data were analyzed using student's *t*-test and Wilcoxon rank sum test. Intergroup comparisons were analyzed using Chi-squre test or Fisher exact test for categorical data. A difference of $P < 0.05$ was deemed as statistically significant.

3. Result

3.1. Demographic information

Table 2 represented the general demographic information of patients enrolled in this study. The average age of patients was 43.4 ± 10.8 years, with an average clinical history of 47.8 months. There were 35 males and 80 females, resulting in a male-to-female ratio of 1:2.3. The mean tonsil descent degree was 8.6 ± 4.1 mm, and the mean involved SM cavity segments was 9.6 ± 4.6 mm. 11 patients in PFDD group and 19 patients in PFDRT group combine basilar invagination or occipitalization. According to the medical records, the mean length of stay was 19.7 days. Differences in the above characteristics were not significant between the PFDRT and PFDD groups ($p < 0.05$).

3.2. Clinical results

In this study, the clinical results 6 months after surgery were evaluated through reexamination visits. Almost all studies on decompression for CM-1 had reported improvements in the same order of magnitude regardless of the different ways to handle dura, arachnoid, or tonsils. The neurological scores in (Table 3) suggested the improvements of different symptoms after surgery. Typically, the most profound postoperative improvement could be observed in occipital pain, while scores in other items tended to improve only slightly, indicating limited postoperative symptom improvement [12–15]. The overall clinical symptoms had been improved, but patients still suffered from residual symptoms. Differences in all items between two groups after operation were not statistically significant (Z=-1.408,P = 0.159 > 0.05)

Overall 86.9% patients considered that their conditions were improved 6 months after surgery, whereas 10.4% reported no change and 2.7% reported worsened conditions (Table 4). Statistical analysis revealed no significant differences in the outcomes (effective, unchanged, and invalid) between 2 groups (χ^2 = 0.160, P = 0.557 > 0.05).

3.3. Radiological results

After decompression in patients with an associated syrinx, the sryinx resolution rate in patients could be compared. All patients underwent an MRI examination, and the syrinx location was recorded. The rate of postoperative syrinx regression was 69.2% in PFDRT group and 78.3% in PFDD group (Table5). No statistically significant difference was found in the surgical results between PFDD and PFDRT groups according to the statistical analyses (χ^2 = 1.046, P =0.376 > 0.05). Radiologically, the syrinx size was not correlated with the extent of clinical results. Radiological improvement in terms of syrinx size could

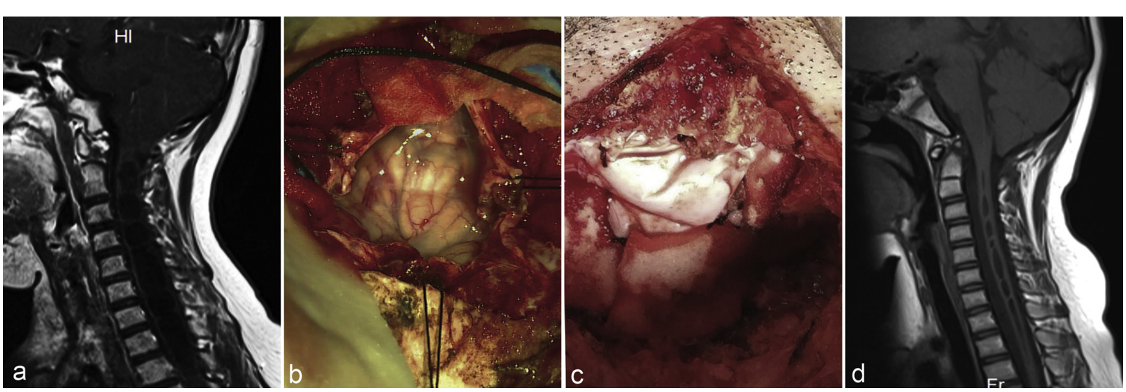

Fig. 2. A 34-year-old CM-I male patient with a 1-year history of right upper limb weakness. (a) Preoperative T1-weight image showed CM-I with a syrinx from C2 to T9. (b) After Y-pattern cutting of the dura mater, the arachnoid swelling above the cerebellar tonsils removed. (c) The artificial dura mater was carefully sutured, expanded and decompressed. (d) Follow-up MRI examination 0.5 year after surgery showed a wide cisterna magna and obvious regression of syrinx. The symptom of weakness was alleviated.

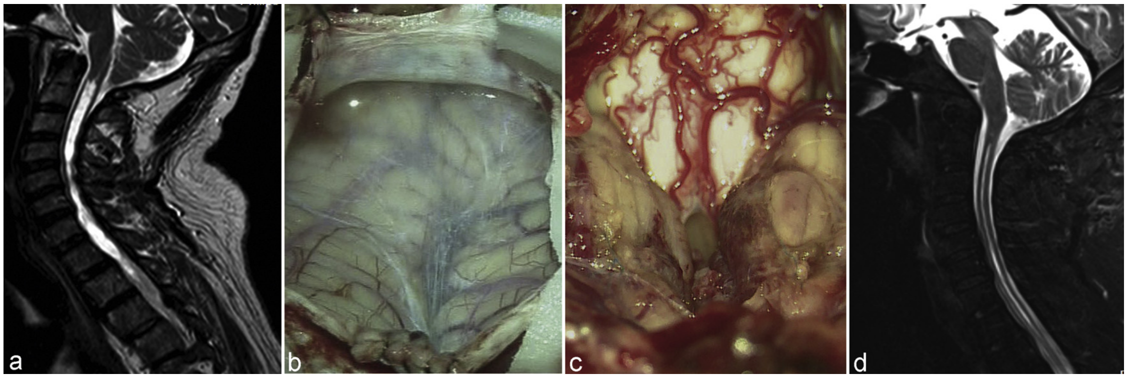

Fig. 3. A 43-year-old female patient with a 2-year history of occipital pain. (a) T2-weighted MR scan of cranio cervical junction revealed a classical CM-1 with a syrinx from C2 toT6. (b) After dura opening, no apparent arachnoid pathology was found. (c) Subarachnoid resection of bilateral tonsils and inspection of Magendie foramen. (d) Follow-up MRI examination 0.5 years after surgery revealed a wide cisterna magna with obvious resolution of syrinx. The symptom of occipital pain was vanished.

Table 2
Demographic and clinical information of patients.

	Total (n = 115)	PFDD group (n = 37)	PFDRT group (n = 78)	*P* value
Gender				
Male	35	14	21	0.180
Female	80	23	57	
Mean age (year)	43.37 ± 10.80	42.92 ± 11.66	43.59 ± 10.44	0.757
Symptom duration (Months)	47.76 ± 69.09	45.06 ± 64.71	49.03 ± 71.43	0.771
Hospital stay (days)	19.72 ± 6.07	19.86 ± 6.52	19.65 ± 5.88	0.868
Head-neck pain	62	17	45	0.238
Numbness of limbs	42	18	24	0.063
Posterior cranial nerve disorder	5	1	4	0.915[a]
Motor weakness	24	6	18	0.398
Gait ataxia	7	3	4	0.808[a]
Muscle atrophy	15	4	11	0.624
Pathological reflex	12	3	9	0.814[a]
Headache and dizziness	24	6	18	0.398
Degree of tonsil descent(mm)	8.61 ± 4.06	9.15 ± 3.94	8.36 ± 4.12	0.334
Location of syrinx				
Cervical	26	7	19	0.515
Cervicothoracic	85	28	57	0.767
Cervicothoracolumbar	4	2	2	0.816
Cavity segment	9.57 ± 4.57	10.13 ± 4.42	9.17 ± 4.16	0.579
Preoperative s (mm)	6.04 ± 2.77	5.37 ± 2.49	6.11 ± 2.81	0.674
Preoperative c(mm)	9.25 ± 2.26	9.23 ± 1.93	9.25 ± 2.31	0.984
Postoperative s(mm)	3.27 ± 1.96	3.18 ± 1.93	3.48 ± 2.25	0.311
Postoperative c(mm)	7.02 ± 2.03	7.38 ± 1.42	6.98 ± 2.09	0.598
Preoperative S/C	0.60 ± 0.19	0.56 ± 0.15	0.61 ± 0.19	0.071
Postoperative S/C	0.46 ± 0.19	0.45 ± 0.20	0.46 ± 0.16	0.333
Hydrocephalus	3	1	2	1.000[b]
Scoliosis	2	0	2	1.000[b]
Deformity of foramen magnum	30	11	19	0.084
Operation time(min)	150.40 ± 38.10	134.1 ± 30.76	159.32 ± 40.06	0.012
Blood loss volume(ml)	142.75 ± 107.36	143.96 ± 94.72	137.69 ± 102.01	0.759
Postoperative hospital Stay (days)	13.82 ± 6.06	13.67 ± 7.62	14.05 ± 8.93	0.758

[a] Represents the continuous correction chi square test.
[b] Indicated by exact probability test.

be achieved earlier in patients undergoing a syringosubarachnoid shunting, which did not match with the clinical recovery [7,14]. Shunting surgery was not used in all patients in this study. MR also show that 4 cases in PFDRT group and 3 cases in PFDD group cisterna magna reconstruction not obvious than before and continued crowding. The rest of the patients occipital cistern had been dissected obviously.

3.4. Complications

Complications could be divided into operation-related neurological complications and general postoperative complications. (Table 6) suggested that fever, dizziness and headache were caused by aseptic meningitis, which were the most frequently occurring procedure-related complications. Fever was diagnosed in the presence of body temperature of 38–39 degrees lasting for more than one week, as well as by a lumbar puncture negative bacterial culture for three times. Fever was

Table 3
Neurological Scores 6 month postoperative as defined in Table 1.

Score	Pain	Sensory dist dysesthesias	Motor deakness	Gait ataxia
Preoperatively PFDRT	3.47 ± 0.85	2.90 ± 0.89	3.62 ± 1.12	3.88 ± 0.12
6 month after PFDRT	4.64 ± 0.73*	3.72 ± 0.92*	3.92 ± 1.32*	4.75 ± 0.45*
Preoperatively PFDD.	3.64 ± 0.63	2.58 ± 0.90	3.00 ± 1.60	3.86 ± 1.38
6 month after PFDRT	4.71 ± 0.47*	3.26 ± 1.24*	3.13 ± 1.73*	4.86 ± 1.38*

"*" Comparison with pre-operation $p < 0.05$.

Table 4
Clinical changes after operation.

Symptoms(%)	PFDRT	PFDD
Improve	69(88.4)	31(83.8)
Unchanged	7(9.0)	5(13.5)
Deteriorate	2(2.6)	1(2.7)

Table 5
Post-operative syrinx.

Syrinx, n (%)	PFDRT	PFDD	Total
Worse	2(2.6)	1(2.7)	3
Stable	22(28.2)	7(18.9)	29
Improved	54(69.2)	29(78.4)	91

Table 6
The summary of the complications of surgery.

Complications, n(%)	PFDRT (n = 78)	PFDD (n = 37)	χ^2	P value
Fever	11(14.1)	8(21.6)	1.029	0.420
Headache and dizziness	20(25.6)	3(3.8)	4.822	0.028
Subcutaneous effusion	3(3.8)	1(1.2)	—	1.000*
Wound infection	3(3.8)	2(2.6)	—	1.000*
Total complications	37(47.4)	13(35.1)	1.545	0.234

"*"Extract probability method P value; "—"Represents no P value.

completely resolved in all cases after hormone therapy (dexamethasone 4 mg intravenously every 6 h) and symptomatic treatment. 4 cases had subcutaneous hydrops, with no obvious discomfort, and were thereby no given any special treatment. The symptom of headache was improved after symptomatic treatment. As could be seen, the incidence of dizziness and headache in PFDRT group was higher than that in PFDD group ($P = 0.028 < 0.05$). No significant differences were observed in overall complications between the two groups ($P > 0.05$).

3.5. Correlated analysis

Reduction in cavity volume by 20% after operation was regarded as good outcome for cavity recovery. Regression analysis revealed that only the preoperative cavity of > 30% was positively correlated with the improvement rate (Fig. 4, P = 0.001). In the meantime, age, sex, cranifacial junction of malformation, symptom duration, cerebellar tonsillar descent and operation were not correlated with improvement in the patient's cavity.

Besides, correlation analyses were conducted to assess the associations between syrinx size and the potential predictors (including age, sex, cerebellar tonsillar descent, and symptom duration, and cranifacial junction of malformation) (Tables 7 and 8), but no correlation was found between these potential predictors and syrinx size.

Correlation analysis was also conducted on the degree of cerebellar tonsillar hernia, and it was found that age was associated with the extent of cerebellar tonsillar hernia (r=-3.29, p = 0.03), but no correlation was found in other factors.

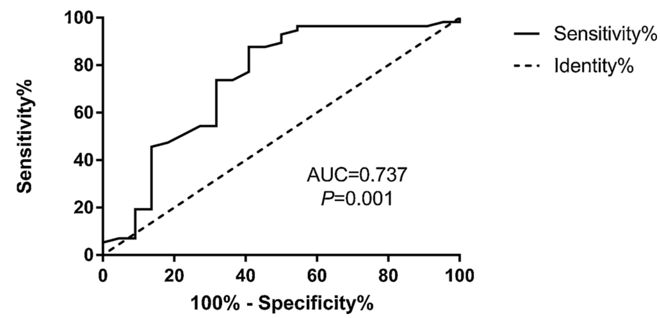

Fig. 4. The ROC curve of syringomyelia volume > 30% predicted the reduction in postoperative syrinx by > 20% AUC (Area Under Curve) = 0.737 p = 0.001.

Table 7
Factors of syringomyelia volume > 30%.

Factors	OR	*P* value
Age	1.01	0.665
Sex	3.02	0.082
Cranifacial junction of malformation	0.29	0.044
Duration of symptom	0.44	0.996
Cerebellar tonsillar discent	0.96	0.828

Table 8
Factors of syrinx reduced > 20%.

Factors	OR	*P* value
Age	0.82	0.993
Sex	0.17	0.362
Cranifacial junction of malformation	3.38	0.112
Duration of symptom	0.99	0.434
Cerebellar tonsillar discent	0.95	0.556
Syringomyelia volume	19.54	0.001
Surgical procedure	0.49	0.475

4. Disscussion

4.1. Pathology and pathogenesis

CM-1 is defined as the herniation of cerebellar tonsils below the foramen magnum level to the upper cervical, which is commonly associated with syringomyelia as a result of deranged CSF dynamics at the craniovertebral junction and upper cervical canal [14].

Although the mechanism for cerebellar tonsillar ectopia is not clear, congenital hindbrain herniation makes up the majority of these congenital malformations [16]. Several studies have shown that CM-1 is mainly related to a small posterior fossa volume, as well as the possible genetic disposition for an unknown percentage of patients [14,17].

Typically, additional anomalies of the joints and discs in craniocervical junction, assimilations of atlas to the occiput, basilar invagination, and Klippel-Feil syndromes are the features frequently found in preoperative imaging. As such a diverse range of associated anomalies exists, it has become difficult to identify one single theory that can effectively explain the occurrence of both hindbrain herniation and these associated anomalies. A number of theories have been proposed with the purpose of accounting for the embryology and pathogenesis of the Chiari I malformations and their associated complications and clinical syndromes. and outline their validity and relevance to our contemporary understanding of these anomalies [18]. Abnormal embryonic development is the most accepted theory. The embryology of the posterior cranial fossa is complex, which relies on a unique timing of various neurovascular and bony elements [19]. Derailment of the developmental processes of the hindbrain can lead to a wide range of malformations such as the Chiari malformations [20]. Moreover, pain is the most common preoperative symptom, including occipital pain, neck pain, back pain, and upper limb pain. Patients will also have mild functional impairments, which may affect their activities of daily living. In this study, the Neurological Scoring System is employed to evaluate the neurological functions in patients.

4.2. Radiology

MRI is used as a basic tool for investigation in this study. A tonsillar herniation of more than 5 mm is widely considered as a pathological sign of CM-I in adults [21]. Cardiac gated cine MRI and intraoperative ultrasound are very helpful in doubtful cases, which can demonstrate a CSF flow obstruction pattern as an indicator of a clinically relevant herniation [22]. Likewise, neurophysiological examinations and neurological evaluation have been proposed with an aim to search for evidence of medulla oblongata or spinal cord compression. Such disease is associated with diversified clinical manifestations and the diameter of syrinx is changed. Therefore, it is important to identify the main symptoms matching with their radiological changes and assess their dynamics. In our study, postoperative MRI is used to evaluate syrinx 6 months after suboccipital decompression. Such a period is long enough to evaluate the clinical symptoms and radiological changes for assessing the therapeutic outcomes after restoration of CSF circulation at craniovertebral level. In addition, CT and X-ray are also used in identifying these bony anomalies. A small posterior fossa volume, additional anomalies, assimilations of atlas to occiput, and basilar invagination, should be detected in preoperative imaging, since they may indicate craniospinal instability.

4.3. Syringomyelia

Most previous studies have reported the rate of syrinx reduction ranging from 52% to 69% [17,23]. It is also observed that, the syringomyelic cyst may not be changed or reduced regardless of the adequate decompression performed. This may be related to the fact that prolonged compression will lead to pathological changes in nerve tissue and decreases in spinal cord elasticity. Syrinx reduction tends to develop more slowly than clinical improvements [23,24]. Preoperative clinical symptoms are not correlated with the presence or extent of an associated syrinx. Nevertheless, the postoperative course of syringomyelia is not correlated with the clinical response to operation [24]. Sustained postoperative decrease indicates an adequate decompression and sufficient CSF flow at the foramen magnum.

4.4. Surgery

Surgical management is the only effective method for the treatment of CM I-SM with definite symptoms [25]. Indications of surgical treatment for CM I-SM are rapid progression signs and symptoms induced by impairments in the nervous system. Early stage surgery can relieve cerebellar tonsillar compression, and reduce the liquid flowing from SM to brain stem and spinal cord, thereby controlling the progressive nerve damage and promoting disease rehabilitation.

Bony decompression is not sufficient enough for mitigating patient symptoms caused by dural or arachnoidal adhesion or a neural abnormality at the foramen magnum level. Different combinations of surgical techniques have been evolved to improve symptoms and signs, to decrease the cavity size, and to reduce the incidence rates of adverse events and complications. Some experts have advocated duraplasty [7,24] since dural enlargement repair has been performed to enlarge and expand the volume of posterior fossa, and thus decompression is sufficient. General patients will be operated with bony decompression, with the formed bone window of approximately 3 cm × 4 cm. Such technique is shown to achieve a good decompression effect, which can maintain the posterior cranial fossa support for cerebellum and dura mater, thus avoiding further herniation of posterior fossa contents caused by the large bone window. However, dura mater opening and reconstruction can lead to CSF leakage, intracranial infection and other complications. Meanwhile, local adhesion and scar formation may result in shrunk and oppressed posterior skull.

Long-term [7,10] cerebellar tonsillectomy has been considered as a classic surgical method for treating CM. Specifically, it can effectively reduce posterior fossa compression and enlarge the cisterna magna to restore the CSF circulation. With the development of imaging diagnostic technology, tools which identification of craniocervical junction anatomy and CSF dynamics with real-time images may be used in selecting candidates for PFD with bone removal alone.

4.5. Result discussion

A recent literature review points out that bony decompression combined with duraplasty or bony decompression with the resection of tonsils will lead to a statistically significant higher rate of improvements in symptoms and signs than bony decompression alone without duroplasty or shunting, but no statistical differences can be detected between these two groups [26]. In our study, 86.9% patients have reported their clinical conditions as improved 6 months after the operation, with the most significant postoperative improvement in occipital pain. Neck pain can respond best to surgery due to the relief of craniospinal pressure. Besides, motor dysfunction in the lower extremities that caused by pressure in the corticospinal tracts, is also favorably improved after surgery. In comparison, dysesthetic pain and sensory deficits have poorer outcomes, and amyotrophy in the upper extremities has the poorest response, suggesting the irreversible destruction of anterior horn cells by the syrinx [27].

Most patients can attain a satisfied outcome. However, symptoms in some patients have not been improved, especially for those related to central spinal cord canal damage. Such findings show that our surgical treatment can only prevent disease progression but cannot reverse spinal cord damage. Moreover, cerebellar tonsillar ischemia and cyst can often be seen in CM- 1 patients [28]. However, it remains unclear on whether cell apoptosis exists in delayed cervical spinal lesions, as well as the relationships between relieved symptoms and cell regeneration, which needs to be explored in future research.

4.6. Surgical morbidity and complications

Specifically, the incidence of aseptic meningitis is the only difference between the two procedures, which may be related to the fact that PFDRT procedure is more complicated than PFD. Notably, keeping the subarachnoid space clear from any contamination with blood is mandatory to limit the formation of arachnoid scar postoperatively. The related symptoms can be cured after symptomatic treatments such as delayed onset and delayed ambulation. Moreover, postoperative incisional infection can be mainly due to the improper operation of dressing postoperatively.

CSF leakage is a common complication often induced due to the small laceration and tear in duraplasty. A tight running suture following duraplasty is as important as a tight closure of the muscular layer.

Hydrocephalus is another postoperative complication, which occurs in 3.0% decompression cases in this series. Dubey et al. [29] had analyzed 500 operation-related complications after posterior fossa surgery, determined a rate of postoperative hydrocephalus at 4.6%. Most authors believe that such finding is related to the formation of subdural hygromas in the posterior fossa. Whether surgical treatment is always required in postoperative ventricular dilatation should be decided based on the clinical course. Most related symptoms will be relieved after ventriculoperitoneal shunting

Several factors, such as sex, peroperative size and SM size, will not as the predictors of better postoperative outcomes. Regression analysis reveals that only the preoperative cavity of > 30% is positively correlated with the improvement rate. Patients with smaller holes may not be able to recover easily, which can be attrirbuted to the chronic cavity formation in the cavity of spinal cord.

5. Conclusion

The current longitudinal prospective study suggests that both PFDRT and PFDD can achieve comparable short-term clinical outcomes for adult CM-I patients with syringomyelia. The symptoms in patients are gradually relieved and the spinal cavity is decreased after surgery. Typically, PFDRT may lead to higher complication rates of dizziness and headache, as well as longer operation time and higher risk of operation. Specifically, the preoperative syrinx cavity of > 30% is an indicator of improvement rate and reduction in cavity volume by 20%. It is advisable that surgical procedure should be selected based on concrete patient condition and the operating skills of surgeon.

Conflict of interest

The authors declare that they have no conflict of interest.

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