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Surgical Treatment of Chiari I Malformation with Ventricular Dilation

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Abstract

Ventricular dilation affects 7% to 10% of patients with Chiari type I malformation (CIM), but the choice of surgical treatment is controversial. To study the surgical approaches for treating CIM with ventricular dilation and to evaluate the efficacy of posterior fossa decompression (PFD), clinical and imaging data of 38 adult patients who received surgical correction performed at the authors' department from 2004 to 2011 were reviewed. Ventricular dilation was defined as Evans' index > 0.30. Preoperative fundus examinations were done on all patients and no papilledema was found. Surgical procedures included a sub-occipital decompression and a C1 laminectomy, followed by a duraplasty with an autologous graft. Evans' index was measured before and after surgery respectively, and a paired samples t-test was performed to examine the difference. Factors predicting outcomes were investigated using logistic regression analysis. Follow-up was done to all patients with an average duration of 43 months. All postoperative magnetic resonance (MR) images showed a relief of cervicomedullary compression and recreation of the cisterna magna. Symptoms improved in 33 patients (86.8%), remained stable in 5 (13.2%), and no patient deteriorated. No significant change in ventricular size was observed after surgery (P = 0.257). Regression analysis showed duration of illness had a significant effect on clinical outcome (P = 0.034, OR = 12.5, 95% CI: 1.214, 128.661). Our study suggests that the intracranial pressure (ICP) of patients with CIM and ventricular dilation is usually normal. PFD with duraplasty is an effective and safe treatment for CIM with ventricular dilation. Treatment of ventricular dilation is unnecessary before PFD as long as there is no persistent headache, vomiting, and papilledema.

Key words: Chiari malformation, ventricular dilation, posterior fossa decompression

Introduction

Among patients with Chiari type I malformation (CIM), 7% to 10% present with associated ventricular dilation.[7,10,12] The pathogenesis of this association is still a matter of debate. Most authors believe ventricular dilation is the result of obstruction of the foramen magnum, and it is a hypertensive hydrocephalus.[13,15] Some insist the ventricular dilation should be treated before posterior fossa decompression (PFD), otherwise acute ptosis of the cerebellar tonsil could be induced[1,14,18]; or a pure PFD is insufficient to relieve the symptoms and a ventriculo-peritoneal (V-P) shunt needs to be added. However, our clinical findings differ from those described above. In this article, we report a series of 38 patients with CIM and ventricular dilation who underwent PFD without preoperative V-P shunt. The clinical presentation, surgical approaches, and outcomes are reviewed and discussed.

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Materials and Methods

I. Clinical presentation

From September 2004 to October 2011, 38 patients with CIM and ventricular dilation were surgically treated in our department (15 males, 23 females; average age, 40 years; range, 16 years to 54 years). Clinical symptoms and signs were retrospectively reviewed. Follow-up was done to all patients with an average duration of 43 months (range, 8 months to 93 months).

II. Fundus examination

Fundus examinations were performed on all patients preoperatively.

III. Neuroradiological assessment

All patients were studied with cranial-cervical magnetic resonance imaging (MRI) and brain MRI or brain computed tomography (CT) scans before and after surgery (Fig. 1A–D). The diagnosis of CIM was defined as the descent of cerebellar tonsils 5 mm or greater below the foramen magnum on MRI.[11] Ventricular enlargement was assessed with Evans' index, the maximum width of the frontal horns divided by the maximum width of the inner skull at the same level (Fig. 1B, D). Ventricular dilation was defined as Evans' index > 0.30,[2,8] and all 38 patients met this criterion. Evans' index was measured again postoperatively (1 month to 41 months after surgery). Moreover, each patient underwent evaluation to determine the extent of tonsillar herniation, the degree of caudal descent of the brainstem, the tentorial angle (Fig. 1A-a), the clival angle (Fig. 1A-b), the brainstem-spinal cord angle, the inclination of the odontoid process (Fig. 1A-c), and any associated cranio-vertebral anomalies. The angulation of the odontoid process was measured by drawing one horizontal line through a midpoint of the synchondrosis between the base and apex of the odontoid process and a second line through the midpoint of the synchondrosis and apex of the odontoid process on a midsagittal MR imaging (Fig. 1A-c).[9] The clival angle is the angle between the clivus and the formen magnum (the MacRae line). The brainstem-spinal cord angle is the angle between the axis of brainstem and the axis of cervical spinal cord.

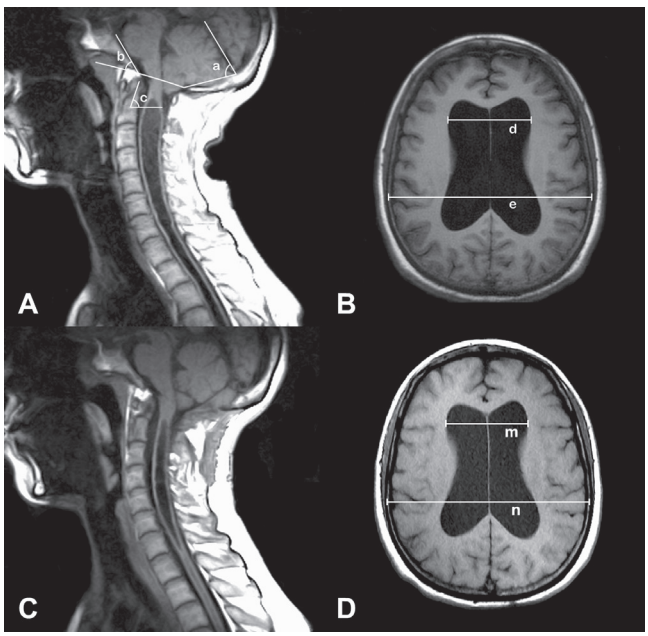

Fig. 1 A: Preoperative cervical T_1-weighted sagittal magnetic resonance (MR) image revealing tonsillar herniation with a holocord syrinx, and showing the measurement of the tentorial angle (a), the clival angle (b), and the angulation of the odontoid process (c). B: Preoperative brain T_1-weighted axial MR image showing ventricular dilation and the measurement of Evans' index (d/e). C: Postoperative cervical T_1-weighted sagittal MR image showing reduction in syrinx size. D: Postoperative brain T_1-weighted axial MR image showing the measurement of postoperative Evans' index (m/n) and demonstrating no obvious change of ventricular size after surgery. (A–D were taken from the same patient, case number 28; C and D were taken 6 months after surgery).

IV. Surgical approach

PFD with duraplasty was performed on all the 38 patients (without V-P shunt). After patients were left-lateral positioned, a middle linear skin incision was made from 1 cm below the inion to the spinous process of C4 and an autologous graft (2 × 2 cm) was resected from the fascia and reserved. The musculo-fascial plane was opened and then the occipital squama, the posterior arch of the atlas, and the spinous process of C-2 were exposed. A middle sub-occipital craniectomy (diameter, 2.5–3 cm) and a C-1 laminectomy (1.5–2 cm) were performed. The thick and constraining dural band found at the occipital-cervical junction was removed. The dura mater was opened with the arachnoid intact, and then the dura mater was grafted with the autologous graft.

V. Statistical evaluation

All the statistical analyses were performed with PASW Statistics 18.0.0 software. The pre- and postoperative Evans' indexes were compared using a paired-samples t test. Logistic regression analysis was done to investigate risk factors on clinical outcome, with each continuous variable divided into two groups by the mean, and odds ratio with 95% confidence interval (CI) was presented. A probability value < 0.05 was considered as statistically significant.

Results

The clinical symptoms and signs were respectively summarized in Figs. 2 and 3. The dominant symptom was headache, which was usually triggered by valsalva maneuver, coughing, and changes of body position. The most frequent sign was sensory disturbance,

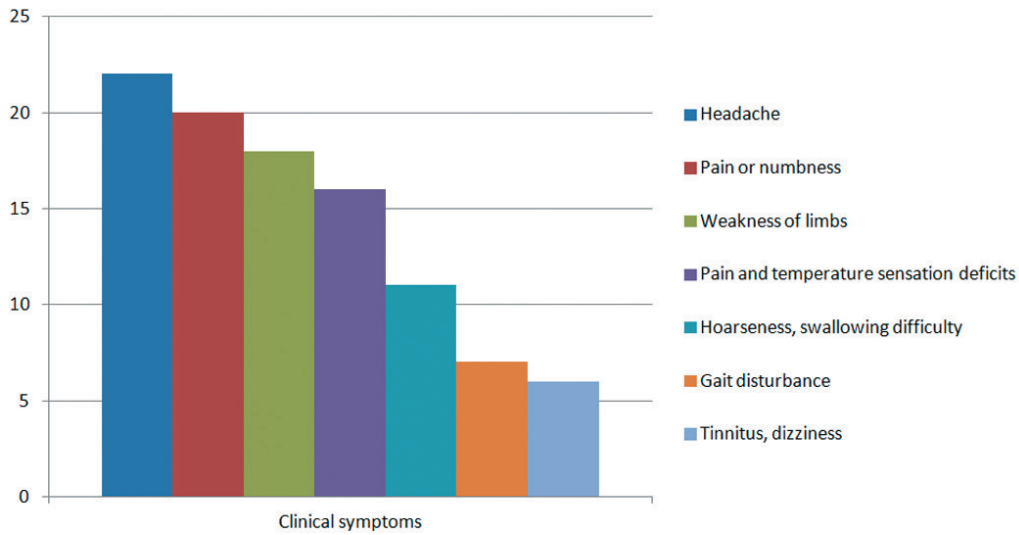

Fig. 2 Summary of clinical symptoms.

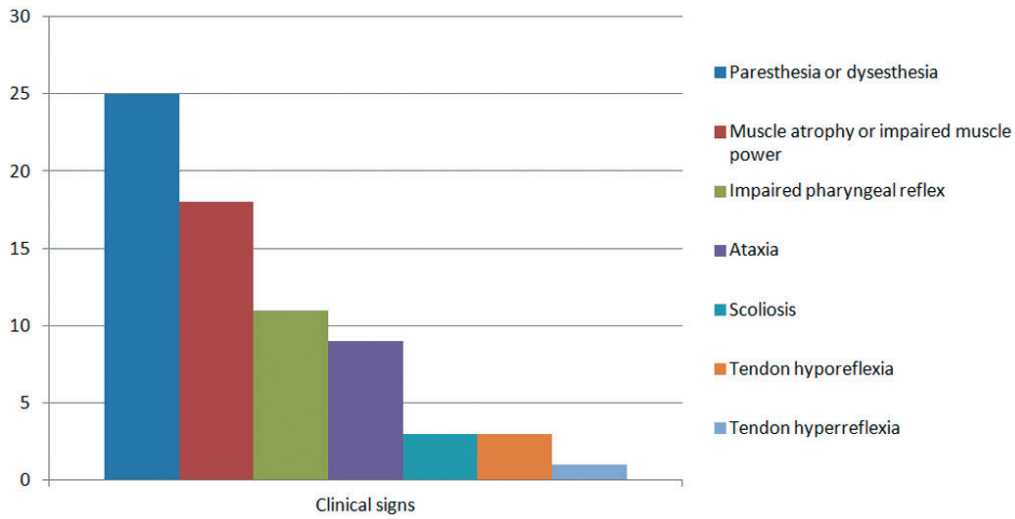

Fig. 3 Summary of clinical signs.

followed by impaired muscle power. Papilledema was found in none of the patients. Other clinical data and detailed imaging characteristics are summarized in Table 1. The duration of symptoms ranged from 1 year to 20 years, with a mean of 5 years. No periventricular edema was observed on preoperative brain MRI or CT images. Neither prominent digital marking nor thinning of skull nor decalcification of dorsum sellae was found on plain craniograms. The length of tonsillar herniation ranged from 5 mm to 16 mm (mean 9.1 ± 3.3 mm), with the tentorial angle from 55° to 120° (mean 88.4° ± 16.0°), the clival angle from 27° to 59° (mean 43° ± 8.1°), the brainstem-spinal cord angle from 143° to 176° (mean 162.3° ± 8.5°), and the angulation of the odontoid process from 60° to 86° (mean 77.3° ± 5.2°). The mean preoperative Evans' index was 0.36 ± 0.04 (range, 0.30 to 0.45). Caudal descent of the brainstem was detected in 11 patients, and the distance of the obex beneath the foramen magnum ranged from 3 mm to 10 mm (mean 6.8 ± 3.3 mm). In one patient (case 4) we found assimilation of the atlas to the occipital bone and a Klippel-Feil anomaly. In another patient (case 35) we observed both basilar invagination and a Klippel-Feil anomaly. Scoliosis was found in 3 patients (case 3, 24, and 29). In 31 patients (81.6%) syringomyelia was observed: 6 with a cervical syrinx, 16 with a cervicothoracic syrinx, and 9 with a holocord syrinx. No other associated cranio-vertebral anomalies were found, such as short clivus and instability of cranio-verterbal junction.

No surgical mortality or severe surgical complications occurred. Recovery of tonsillar pulsation after duraplasty was detected in all patients during surgery. All postoperative MRIs revealed relief of compression on the brainstem and recreation of

Table 1 Summary of clinical and imaging data

Case number	Sex/ Age	Duration of illness (years)	Length of tonsillar herniation (mm)	Angulation of odontoid process (degrees)	Tentorial angle (degrees)	Clival angle (degrees)	Brainstem-spinal cord angle (degrees)	SM	Outcome	Evans' index	
										(Pre-op)	(Post-op)
1	M/24	10	15.9	84	113	51	170	C/T	S	0.37	0.38
2	M/29	2	10.5	78	83	57	160	Holo	I	0.39	0.40
3	M/39	1	7.3	82	55	43	170	C/T	I	0.41	0.40
4	M/53	15	13.8	86	101	55	176	C/T	S	0.36	0.36
5	M/16	2	11.2	78	80	31	162	C/T	I	0.36	0.35
6	M/27	1	8.2	83	120	38	161	C/T	I	0.36	0.37
7	M/40	5	6.0	81	74	59	148	C/T	S	0.37	0.36
8	M/53	1	5.0	78	92	35	172	C	I	0.39	0.39
9	M/26	1	9.8	75	69	34	172	None	I	0.41	0.40
10	M/27	3	12.0	83	96	36	156	None	I	0.39	0.41
11	M/37	6	10.0	84	88	48	165	Holo	I	0.45	0.46
12	M/42	1	7.0	71	105	52	161	C	I	0.40	0.36
13	M/30	1	8.0	73	102	47	162	C/T	I	0.33	0.33
14	M/39	3	6.0	77	85	52	166	C/T	I	0.32	0.32
15	M/55	20	10.3	76	82	53	171	C/T	I	0.36	0.36
16	F/42	2	15.0	73	73	32	145	None	I	0.35	0.38
17	F/39	4	6.7	76	82	44	169	Holo	I	0.30	0.32
18	F/28	1	12.0	79	58	44	170	C/T	I	0.35	0.36
19	F/35	3	12.6	75	72	49	169	Holo	S	0.36	0.35
20	F/45	8	12.0	80	96	43	167	C/T	I	0.34	0.33
21	F/54	14	9.6	81	108	46	159	C/T	I	0.35	0.36
22	F/43	17	5.0	77	91	51	158	C	S	0.30	0.30
23	F/47	1	13.5	72	109	39	170	Holo	I	0.36	0.33
24	F/39	2	11.0	79	68	45	176	C/T	I	0.37	0.35
25	F/41	3	7.8	73	102	41	157	C/T	I	0.33	0.32
26	F/37	2	12.0	70	82	40	153	Holo	I	0.30	0.31
27	F/50	4	5.0	74	85	27	151	C/T	I	0.36	0.36
28	F/53	4	6.5	84	104	54	168	Holo	I	0.41	0.41
29	F/40	3	7.1	80	87	51	143	C	I	0.36	0.35
30	F/24	7	6.5	73	90	34	153	Holo	I	0.43	0.40
31	F/56	1	8.0	76	116	40	165	C/T	I	0.34	0.33
32	F/38	2	5.5	73	83	34	150	C	I	0.36	0.37
33	F/56	19	10.3	78	109	38	160	None	I	0.43	0.42
34	F/43	6	5.0	83	80	41	169	None	I	0.31	0.31
35	F/55	5	7.7	60	74	44	161	None	I	0.31	0.30
36	F/27	3	5.0	71	100	35	153	C	I	0.35	0.34
37	F/54	12	16.0	80	71	55	167	Holo	I	0.33	0.32
38	F/35	1	7.0	82	75	45	165	None	I	0.41	0.41

C: cervical, C/T: cervicothoracic, F: female, Holo: holocord, I: improvement, M: male, S: stability, SM: syringomyelia.

the cisterna magna. Symptoms improved in 33 patients (86.8%), remained stable in 5 (13.2%), and no patient deteriorated after surgery. Ventricle size remained stable in all patients and no significant change was observed ($t = 1.152$, $P = 0.257$). So far, no patient had required a V-P shunt to treat ventricular dilation. Out of the 31 patients with syringomyelia, 8 patients showed obvious reduction in syrinx size, and a slight reduction was observed in the others.

Logistic regression analysis showed duration of illness had a significant effect on clinical outcome ($P = 0.034$). Patients with duration of less than 5 years were almost 12 times more likely

than those with duration of 5 years or more to improve or to become asymptomatic (OR = 12.5, 95% CI: 1.214, 128.661). However, age at surgery (P = 0.261), syrinx (P = 0.139), length of tonsillar herniation (P = 0.955), brainstem herniation (P = 0.746), Evans' index (P = 0.989), angulation of the odontoid process (P = 0.622), the tentorial angle (P = 0.730), the brainstem-spinal cord angle (P = 0.828), and the clival angle (P = 0.081) had no significant effect.

Discussion

It is believed that there are two kinds of CIM, congenital and acquired.[3,4] In congenital CIM, tonsillar herniation is the primary change and it leads to ventricular dilation. Concerning the pathogenesis of congenital CIM, recent morphometric studies have revealed that tonsillar herniation is resulted from overcrowding due to small posterior cranial fossa, which may be caused by underdevelopment of the mesodermal occipital somite.[12,19] The herniated tonsils are responsible for the obstructed CSF flow at the level of the foramen magnum and supratentorial ventricles are enlarged because of the increased intracranial pressure (ICP). In acquired CIM, supratentorial hypertensive hydrocephalus is the primary change and it causes tonsillar herniation, exerting pressure from the above.[5] Patients always suffered from hydrocephalus for a long time previously, which could be caused by intracranial infection, subarachnoid hemorrhage, or traumatic brain injury. The long-standing hypertensive hydrocephalus leads to tonsillar herniation.

All cases presented in this report are congenital CIM complicated with ventricular dilation. History of intracranial lesions, meningitis, subarachnoid hemorrhage, head trauma, or lumbar puncture were absent in all patients.

Most authors believe the ventricular dilation in congenital CIM is a hypertensive hydrocephalus secondary to obstruction of the foramen magnum. However, from our results, a number of observations can be made. First, although headache was the most common complaint,[6] persistent headache and projectile vomiting were really rare. Twenty-two out of 38 cases suffered from headache, but they were all sub-occipital pain and none of them had a history of projectile vomiting. We speculate that headache is due to the compression on cervical nerves, rather than increased ICP. Second, there was no papilledema in any patient of this series, including the 22 cases with headache. Third, no periventricular edema was observed on brain MRI or CT images. Neither prominent digital marking nor thinning of skull nor decalcification of dorsum sellae was found on plain craniograms. Fourth, obstruction of the CSF flow around the foramen magna was relieved and symptoms were improved in most cases after surgery but ventricle sizes were not reduced. This conflicts the theory that it is an obstructive and hypertensive hydrocephalus.

Besides, prior to this series of patients, ventricular puncture and external ventricular drainage were performed under a local anesthesia on four patients before PFD (these were not included in this series). ICP was measured immediately the puncture succeeded, and intermittent measurements were done subsequently in a conscious condition in all patients. All results were within normal range. Symptoms were not improved in any of the four patients, and no change of ventricular size was observed on brain CT images after 2 days of drainage. PFD was subsequently performed and all patients' symptoms were significantly improved. Moreover, it was also reported that ICP monitoring revealed normal results in these patients.[17]

These findings suggest that the ICP of patients with congenital CIM and ventricular dilation is usually normal, rather than hypertensive. Therefore, the treatment of ventricular dilation was unnecessary before PFD as long as there is no persistent headache, vomiting, and papilledema. On the contrary, the ICP of patients with primary hydrocephalus and acquired CM are always high and the ventricular dilation should be managed first. Thus, it is very important to identify whether the patient has a history of long-standing hydrocephalus, intracranial infection, subarachnoid hemorrhage, and traumatic brain injury. Moreover, we recommend that fundus examination and brain CT or MRI should be the routine examinations for CIM patients.

In this series, symptoms were improved in 33 (86.8%) out of the 38 patients. However, in the 31 patients with syringomyelia, only 8 patients showed obvious reduction in syrinx size after surgery. In our opinion, the objective of the surgical management is to improve clinical presentations, rather than radiographic improvement. And it is reported that there is no significant correlation between the reduction of syrinx size on MRI and the degree of clinical improvement.[16] In clinical practice, we have even found deterioration of symptoms in some patients whose syrinx was rapidly reduced after surgery. On the other hand, excessive decompression and opening of the arachnoid should be avoided in these patients to prevent the sudden decline of the cerebellar tonsil. In our experience, a micro-bone window PFD (diameter, 2.5–3 cm) with duraplasty

is enough to relieve symptoms in most patients but may not enough to significantly reduce the syrinx in a short time after surgery.

Besides, risk factors were investigated and duration of illness was significantly correlated with the clinical outcome. This finding may suggest that surgery should be performed early in patients with CIM and ventricular dilation.

In conclusion, we speculate the ICP of patients with CIM and ventricular dilation is usually normal, and our current study confirmed the effectiveness and safety of PFD with duraplasty for the management of this condition. A micro-bone window PFD with duraplasty is enough to improve symptoms and to prevent the progression of the disease in most cases and a V-P shunt is usually unnecessary, unless persistent headache, vomiting or papilledema are present. Duration of illness is a predictor of poor clinical prognosis.

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