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Surgical Outcome after Posterior Fossa Decompression with and without Duraplasty in Adult Chiari Malformation Type-I

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Abstract

Background Data: Adult Chiari malformation is a heterogeneous group of conditions, with the underlying commonality of disruption of normal CSF flow through the foramen magnum. Some cases are congenital, but others are acquired. The optimal surgical treatment of adult Chiari malformation type-I is unclear.

Purpose: To evaluate the operative and postoperative results of extradural and intradural approaches in the treatment of Chiari malformation type-I.

Study Design: A descriptive retrospective clinical case study.

Patients and Methods: Twenty patients underwent surgery for adult Chiari malformation type-I. They were divided into two groups; posterior fossa decompression group and posterior fossa decompression with duroplasty group. Each group included 10 patients. They were operated between 2008 and 2015. Participants were evaluated pre-operatively and post-operatively every three months. Operative time, hospital stay and complications were assessed. The clinical outcomes were compared between the two groups using The Chicago Chiari Outcome Scale.

Results: No statistically significant differences were found between the decompression and duroplasty groups with regard to demographics, preoperative symptoms, radiographic characteristics, and clinical outcomes.

However, the operative time, hospital stay and aseptic meningitis were higher in the duroplasty group.

Conclusion: The clinical outcome of posterior fossa decompression is nearly identical to that of posterior fossa decompression with duroplasty in adult patients with Chiari type-I; however, the operative time, hospital stay and complication rate is higher in duroplasty group. (2016ESJ113)

Keywords: Chiari malformation type-I, duroplasty, posterior fossa decompression.

Introduction

Chiari malformation refers to the downward displacement of the cerebellar tonsil through the foramen magnum into the upper part of the spinal canal.[8,20] No consensus exists regarding the etiology of Chiari malformations. Among the subtypes of Chiari malformation, Chiari malformation type-I (CM-I) is identified most commonly in adulthood and is defined by the degree cerebellar tonsillar extension below the basion opisthion line on sagittal and coronal magnetic resonance images.[8,20]

Despite the fact that many surgical modalities for Chiari malformation have been used in clinical practice, controversy still exists. Two main types of surgical modalities have been advocated for the treatment of Chiari malformation type-I. One type is posterior fossa decompression alone (PFD) or posterior fossa decompression with duroplasty (PFDD) and the other is reduction of the syrinx cavity using different types of shunt procedures.[11,18] Posterior Fossa decompression still remains the primary surgical technique for the treatment of Chiari malformation type-I because the shunt technique produces a risk of iatrogenic spinal cord injury.[3,5,11,13]

Extensive work has been performed concerning PFD and PFDD.[2,6,11,12,16,27] However, whether duroplasty is performed during posterior fossa decompression remains controversial. Most studies in this area have been conducted in pediatric patients.

In order to identify the different surgical outcome between PFDD and PFD in adult patients, we retrospectively studied the clinical data of twenty Chiari malformation type-I patients who had undergone operations from 2008 to 2015

Patients and Methods

This study was designed as a descriptive retrospective clinical case study. Between February 2008 and July 2015, at Suez Canal University Hospitals (Ismailia, Egypt) a total of twenty consecutive patients were included. The patients were categorized into two groups; group 1: Patients operated by posterior fossa decompression (PFD) (Ten patients), group 2: Patients operated by posterior fossa decompression and duroplasty (PFDD) (Ten patients). Inclusion criteria include all patients above 18 years with a preoperative magnetic resonance imaging study confirming Chiari malformation type-I with or without syringomyelia. Exclusion criteria include patients with other types of Chiari malformations, or with a history of severe diseases such as coronary artery atherosclerosis and hepatosclerosis. All patients underwent either PFD or PFDD, and their medical records and radiographic characteristics including MRI and computed tomography were reviewed and compared.

The details reviewed included sex, age, symptom duration, operative time, duration of the hospital stay, and preoperative symptoms. The preoperative symptoms were divided into three main categories according to the presence or absence of symptoms and signs specific to Chiari syndrome on the Chicago Chiari Outcome

Scale[1] as follows: pain symptoms, nonpain symptoms, and functionality conditions. (Table 1) Pain symptoms included headache, neck and back pain, and upper extremity pain. Nonpain symptoms included sensory loss, numbness and tingling, muscle weakness, extremity paresthesia, dysphagia, dizziness, ataxia, and others.

An initial MRI examination with contrast was performed. Chiari malformation type-I was stratified into 3 subgroups according to the cerebellar tonsillar descent (CTD)[1,4,26] as follows: grade 1, the tonsil descended more than 5mm below the foramen magnum but did not reach the C1 arch; grade 2, the tonsil reached the C1 arch; and grade 3, the tonsil descended over the C1 arch. The location of the syrinx was classified as none, cervical, or beyond cervical.

Surgical Procedure:

The specific surgical procedure (PFD or PFDD) was chosen by each surgeon on the basis of training and personal preference. No surgeon performed both procedures in this series. All patients were administered general anesthesia and placed in the prone position with slight flexion of the neck using 3-point Mayfield fixation. The skin, subcutaneous tissues, and occipital and Para spinal muscle were cut through with a midline incision extending from the occipital protuberance to the C2 spinous process. In some duroplasty cases, the occipital fascia was left intact for duraplasty use. The incision exposed the edge of the occipital bone, atlantoaxial posterior arch, spinous process, and lamina. The inferior part of the occipital bone, the posterior lamina of C1, and the tip of the spinous process of C2 were removed depending to the level of tonsillar descent to achieve a bony decompression. In the duroplasty group, after decompression, a thick fasciculation-like tissue that compressed the dura was observed. The thick fasciculation-like tissue was removed, and the dura was incised carefully through the midline under a microscope. After opening the dura, the lower pole of the cerebellar tonsil and the cervical spinal cord were exposed. In some cases, the arachnoid had scarring and adhesions and required a sharp dissection. Then, dural grafting was performed with the occipital fascia or artificial dura. Finally, the outer layers were sutured step-by-step to achieve anatomical reduction.

Follow-up and Outcome:

In both groups of patients, duration of surgery, blood loss, and the duration of inpatient treatment were recorded. Intraoperative and perioperative major and minor complications were assessed. Participants were evaluated post-operatively every 3 months. The outcomes were categorized as early (first 3 months) and late (at the end of the first year). The general postoperative outcomes were evaluated based on the Chicago Chiari Outcome Scale. In addition, the general postoperative outcomes were evaluated based on the following criteria: excellent results, improvement of the neurological deficit; good result, cessation of progression of the neurological deficit; and poor result, further deterioration of neurological function.[23] MRI study was performed during the follow-up consultation. The observation of the syrinx cavity resolution was recorded. (Figure 1)

Results

A total of twenty patients (ten in each group) were included in this study. The demographic data of the two study groups are presented in (Table 2) and showed that the two groups of patients were fairly homogeneous and comparable. Seven males and thirteen females were included. All patients were more than 18 years old. The mean age in the PFD group was

32.3±9.5 years in comparison to 34.2±9.7 years in the PFDD group. On average, patients had preoperative symptoms duration for 60 months that ranged from 10 to 108 months.

Chiari malformation type-I patients presented with different symptoms. The symptoms were classified into three main groups: pain symptoms, non-pain symptoms, and functionality. Among all the symptoms, the three most common symptoms included the following nonpain symptoms: sensory loss (70%), tingling and numbness (60%), and muscle weakness (60%). Neck pain and back pain was the most common pain symptoms (40%), and 55% of patients had a mild impairment in the functionality according to the Chicago Chiari Outcome Scale[1] (Table 1). No significant difference was observed in preoperative symptoms between the PFD group and the PFDD group.

All of the patients underwent an MRI examination, and the cerebellar tonsil descent (CTD) and location of the syrinx were recorded. No significant difference was found between the groups in the CTD and location of the syrinx according to the statistical analyses (Table 3).

In regard to the operative findings in the two groups, the decompression group in comparison to the duroplasty group showed less intraoperative blood loss (250 ± 60 ml in PFD versus 442 ± 80 ml in PFDD), shorter operative time (95±20 min in PFD versus 130±50 min in PFDD) and hospital stay (8.1 day in PFD versus 13.2 day in PFDD) and this was statistically significant (P value < 0.05). (Table 2)

In this study, the patients' clinical outcomes were recorded at two time points: short-term (after 3 month of follow-up) and long term (after 1 year of follow-up). The average Chicago Chiari Outcome Scale in the duroplasty group was 9.7 in comparison to 9.4 in the decompression group after 3 month follow up. At the end of the first year the average Chicago Chiari Outcome Scale was 10.4 in duroplasty group in comparison to 10.1 in the decompression group. The results showed that there was no significant difference in the outcome results (excellent, good, and poor) at the short-term and at the long-term follow-up between the two groups (Table 4). The case with CSF fistula in the decompression group was due to tight adhesions at the level of foramen magnum with CSF tear during bony decompression. There were two cases that underwent reoperations in the decompression group due to recurrence of symptoms after 2 and 3 years respectively.

The most serious complication was aseptic meningitis and was reported in two patients (2/20, 10 %), and both patients were reported in the PFDD group. Criteria of aseptic meningitis were positive CSF studies on lumbar puncture with negative culture. Patient with aseptic meningitis presented with fever, headache, and was response to steroids.[16] There was no difference in other complications such as wound infections, CSF fistulas, and subcutaneous hydrops between the two groups (Table 5).

Table 1. Chicago Chiari Outcome Scale[1]

Chicago Chiari Outcome Scale				
Pain	**Non-pain**	**Functionality**	**Complications**	**Total Score**
1: Worse	1: Worse	1: Unable to attend	1: Persistent complication, poorly controlled	4: Incapacitated outcome
2: Unchanged and refractory to medication	2: Unchanged or improved but impaired	2: Moderate impairment (< 50% attendance)	2: Persistent complications well controlled	8: Impaired outcome
3: Improved or controlled with medication	3: Improved and unimpaired	3: Mild impairment (> 50% attendance)	3: Transient complication	12: Functional outcome
4: Resolved	4: Resolved	4: Fully functional	4: Uncomplicated course	16: Excellent outcome

Table 2. Preoperative and Operative Data of the Study Groups

Parameters	PFD	PFDD	Total
Gender: Male/Female	3/7	4/6	7/13
Age/years	32.3±9.5	34.2±9.7	33.8±9.6
Symptoms Durations/months	56.4	63.9	60.4
Hospital stay/days	8.1	13.2	11.5
Blood Loss/ml	250±60 (170-460)	442±80 (300-750)	345±40 (170–750)
Operative time/minutes	95±20 (70- 115)	130±50 (100-180)	115±40 (70–180)

Table 3. Radiographic Characteristics of the Study Groups

Preoperative MRI		PFD	PFDD	Total
Tonsillar descent	Grade 1	2	3	5
	Grade 2	2	2	4
	Grade 3	6	5	11
Syrinx location	None	1	1	2
	Cervical	3	5	8
	Beyond Cervical	6	4	10

Table 4. Follow-up and Outcome

Outcomes		PFD	PFDD	Total
Short term	Excellent result	6	8	14
	Good result	2	1	3
	Poor result	2	1	3
Long term	Excellent result	5	8	13
	Good result	3	1	4
	Poor result	2	1	3

Table 5. Complications Reported in Study Groups

Complications	PFD	PFDD	Total
Reoperation	2	0	2
Wound infection	1	1	2
Aseptic meningitis	0	2	2
CSF fistula	1	1	2
Subcutaneous hydrops	1	2	3

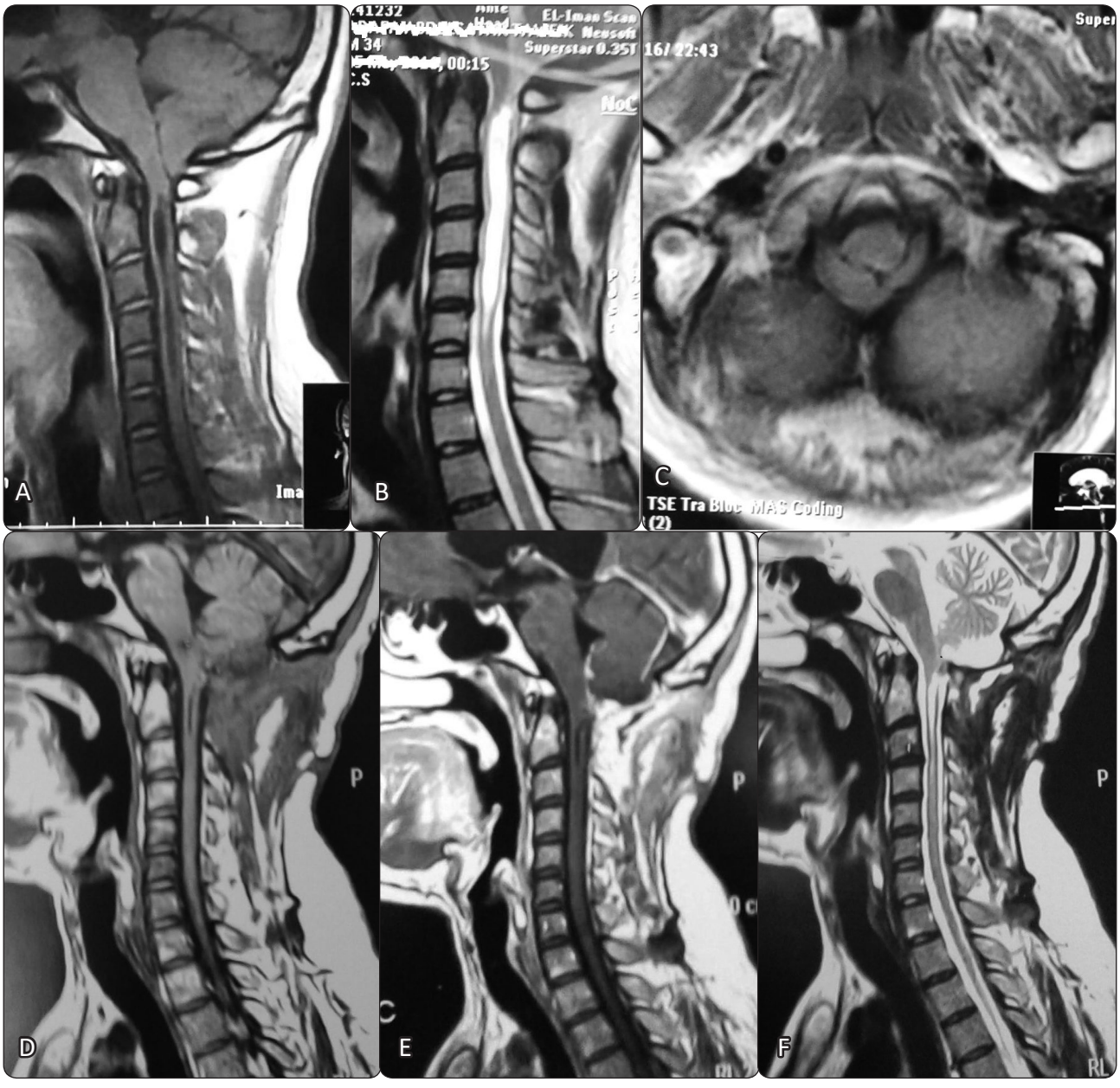

Figure 1. Preoperative MRI craniocervical junction A: sagittal T1, B: Sagittal T2, and C: Axial T1 images in a 31 years old male showing cerebellar tonsillar herniation, narrow posterior fossa, and cervical syringomyelia. Postoperative MRI cranio-cervical junction of D: Sagittal T1, E: Sagittal post-contrast T1, and Sagittal T2 images F: of same patient 1 year after operation (Duroplasty procedure) showing roomy posterior fossa with absent cerebellar tonsillar herniation, and marked decrease of the cervical syringomyelia.

Discussion

Symptoms of Chiari malformation type-I vary in different patients. According to previous studies,[2,8,11,20] the most common symptom is pain, including occipital pain, neck pain, back pain, and upper limb pain. Other clinical manifestations include sensory loss, numbness and tingling, muscle weakness, and ataxia. In this study, the preoperative symptoms were classified into pain symptoms, nonpain symptoms, and functionality based on the Chicago Chiari Outcome Scale.[1] It is noteworthy that the most frequent symptom type in our study was nonpain symptoms, which is not consistent with previous studies. A total of 55% of the patients had mild impairment in functionality, which may affect their daily life. Functionality should be given more attention when evaluating clinical outcomes after surgery in adults. Klekamp[14] analyzed a series of 371 Chiari malformation type-I cases and concluded that children exhibited higher neurological scores than adults. This may be explained by the postnatal growth of the cerebellum.[15] The cerebellum reaches the adult volume in the 2nd year of life after starting with only 15%of its adult volume at birth.[14,15]

Although Chiari malformation type-I can be diagnosed through a variety of imaging modalities, MRI is considered the gold standard diagnostic tool. MRI can be used to evaluate CSF flow, which is an important predictor of clinical outcomes.[20] Apart from the phase-contrast MRI, diffusion tensor imaging is used to evaluate the integrity of the brainstem and cerebellar white matter tracts in Chiari malformation type-I patients. Recently, Ucar et al,[24] demonstrated a new useful sign for Chiari malformation type-I, namely the tonsillar blackout sign on 3-dimensional-SPACE. This sign is particularly useful for distinguishing between symptomatic and asymptomatic Chiari malformation-I patients and patients who are likely to benefit from decompressive surgery.[24] Additional anomalies such as basilar invaginations and assimilations of the atlas to the occiput may also be seen in Chiari malformation-I patients. Other types of imaging tools, such as computed tomography and X-ray, are more useful in identifying these bony anomalies.

Is there any relationship between the clinical manifestations and the severity of Chiari malformation type-I? In a study by Wuet al,[25] the severity of the clinical symptoms did not correlate with the degree of cerebellar tonsillar herniation. However, Greenberg et al,[10] developed a preoperative Chiari Severity Index that integrates the clinical and neuroimaging characteristics. This is a novel tool that predicts patient-defined improvement following Chiari malformation type-I surgery, aids in preoperative counseling, and stratifies patients in comparative effectiveness trials.[10]

Currently, no general consensus exists for incorporating duraplasty in the surgical treatment of Chiari malformation type-I.[2,6,7,9,11,12,16,17,21,22,26] Some authors have advocated posterior fossa decompression is sufficiently effective, whereas others have suggested adding duraplasty. Some authors have concluded that the surgical outcomes between decompression and duroplasty are not significantly different, but the complication rate in duroplasty is higher. The results of this study are consistent with this conclusion. The short-term and long-term follow-up outcomes were similar. The only difference between the procedures was the occurrence of aseptic meningitis is higher in duroplasty group. This difference may be related to the fact that duroplasty has more steps requires opening the dura and suturing the dura with different types of materials. This

destroys the integrity of the original dura and increases the risk of CSF-related complications. Patients undergoing decompression may have preoperative complaints that recur during the postoperative period and subsequently need to undergo duroplasty operations.[11]

Shweikeh et al,[22] evaluated 1593 patients who underwent posterior fossa decompression and 1056 patients who underwent duroplasty and compared the complications and hospital charges in a large national study. The patients who underwent duroplasty experienced more reoperations (2.1% vs. 0.7%), more procedure-related complications (2.3% Vs 0.8%), a longer length of hospital stay (4.4 days 3.8 days), and higher hospital charges (USD 35.321 Vs 31.483). Thus, the authors concluded that duroplasty is associated with significantly more complications and immediate reoperations. Posterior fossa decompression was shown to be more economical by requiring fewer hospital resources. Overall, decompression is more favorable for Chiari malformation type-I. However, according to Mc Girt et al,[19] in children with displacement of the tonsils below the inferior border of the arch of the atlas, ultrasonography-indicated osseous decompression alone was associated with a 2-fold risk of symptom recurrence compared to decompression with duraplasty. Duraplasty may be warranted in cases of tonsillar herniation that extends below C1 lamina regardless of the intraoperative ultrasonography findings.

The present study has all of the limitations of any retrospective study design. The results of this study should be interpreted with caution. A prospective multicenter study with a large and equal number of patients in both groups might provide sufficient data for an adequate comparison of these two techniques to better define the indications and benefits.

Conclusion

The clinical outcome of posterior fossa decompression is nearly identical to that of posterior fossa decompression duroplasty in adult patients; however, the operative time, hospital stay and complication rate is higher for posterior fossa decompression duroplasty.

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الملخص العربي

النتائج السريريه لتوسيع الحق الخلفى للمخ بدون وبمصاحبة ترقيع الام الجافيه فى البالغين المصابين بمتلازمة كيارى ١

البيانات الخلفية: متلازمة كيارى ١ فى البالغين عباره عن مجموعه متنوعه من الصور المرضيه تشترك فى خلل سريان السائل النخاعى حول الثقب الكبير بقاع الجمجمه. بعض الحالات تكون خلقيه و البعض الاخر يكون مكتسب. العلاج الجراحى الامثل لهذه المتلازمه غير واضح.

الغرض: توضيح النتائج الجراحيه و المقارنه بين توسيع الحق الخلفى للمخ بدون و بمصاحبة ترقيع الام الجافيه فى البالغين المصابين بمتلازمة كيارى ١

تصميم الدراسة: دراسه لحالات اكلينيكيه على ٢٠ مريض بالغ يعانون من متلازمة كيارى ١.

المرضي و الطرق: تم اجراء الجراحات من ٢٠٠٨ الى ٢٠١٥ . تم متابعه الاعراض و العلامات و ملاحظه النتائج الاكلينيكيه. تم تقسيم المرضى على مجموعتين . المجموعه الاولى تم توسيع عظمى فقط للحق الخلفى للمخ فى ١٠ مرضى و المجموعه الثانيه تم اضافة ترقيع للام الجافيه للاجراء السابق فى ١٠ مرضى.

النتائج: اوضحت النتائج تقارب المجموعنين من المرضى من حيث الخصائص الديموجرافيه و الاعراض و خصائص الاشعات و النتائج السريريه. تميزة مجموعه ترقيع الام الجافيه بطول فتره الاقامه بالمستشفى و حدوث التهاب سحائى غير ميكروبى فى حالتين.

الاستنتاج: يتضح من هذه الدراسه ان النتائج السريريه لتوسيع الحق الخلفى للمخ متقاربه مع توسيع الحق الخلفى مصحوبا بترقيع الام الجافيه فى البالغين المصابين بمتلازمه كيارى ١. زمن الجراحه و الاقامه بالمستشفى والمضاعفات الجراحيه اكثر فى حالات توسيع الحق الخلفى للمخ مع ترقيع الام الجافيه.