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The Evaluation of Surgical Treatment Options in the Chiari Malformation Type I

Chiari Malformasyonu Tip 1'in Tedavi Seçeneklerinin Karşılaştırılması

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ABSTRACT

AIM: There have been several treatment modalities to reduce the volume of the syringomyelic cavity and the pressure on the brainstem in Chiari Malformation Type I (CM-I). Foramen magnum decompression with and without duroplasty were compared in this retrospective study.

MATERIAL and METHODS: From 2003 to 2006, 27 patients suffering from CM-I were operated on at our institute. The following were measured: the ratio of the syringomyelic cavity to the spinal cord; pre-operative tonsillar herniation from the foramen magnum; pre- and postoperative tonsillo-dural distance; and spino-posterior fossa dural angle.

RESULTS: 83.3 % of the patients in the non-duroplasty and 73.3% of the patients in the duroplasty group were symptom free. The ratio of syrinx regression was 28±10% in the non-duroplasty and 36±33% in the duroplasty group. The tonsillo-dural distance was 3.1±1.8 mm in the non-duroplasty and 4.6±2.1 mm in the duroplasty group (p>0.05). The spino-posterior fossa dural angle was 133.6±9.44º preoperatively and 136.7 ± 9.78º postoperatively in the non-duroplasty (p=0.376); 123.7±11.7º preoperatively and 129.8±11.1º postoperatively in the duroplasty group (p=0.885); no significant difference was found postoperatively (p=0.55, z=1.92), respectively. One patient was re-operated in the non-duroplasty group and thereafter duroplasty was performed.

CONCLUSION: Almost the same clinical outcomes can be achieved with and without duroplasty. There might be an option to perform duroplasty if simple procedure fails.

KEYWORDS: Arnold-Chiari Malformation, Therapeutics, Combined Modality Therapy

ÖZ

AMAÇ: Chiari Tip I Malformasyonlu hastalarda sirengomyelik kavitenin hacmini ve beyinsapı basısını azaltmak amacıyla birçok tedavi yöntemi geliştirilmiştir. Bu çalışmada, basit foramen magnum dekompresyonuyla, dura yamasının da yapıldığı dekompresyon yöntemi karşılaştırıldı.

YÖNTEM ve GEREÇ: 2003-2006 yılları arasında 27 hasta kliniğimizde tedavi edildi. Sirengomyelik kavitenin medullaya oranı, ameliyat öncesi tonsil fıtıklaşmasının uzunluğu, ameliyat öncesi ve sonrası tonsillo-dural mesafe ve spinal aks ile arka çukur arasındaki açı ölçüldü.

BULGULAR: Dura yaması yapılmayan hastaların %83,3'ü yapılanların %73,3'ünde operasyondan sonra belirtiler tamamen kayboldu. Sirenksteki küçülme oranı; dura yaması yapılan grupta %36±33 ve yapılmayan grupta %28±10 idi. Tonsillo-dural mesafe dura yaması yapılan grupta 4,6±2,1mm, yapılmayan grupta 3,1±1,8mm idi (p>0.05). Spinal aks ile arka çukur arasındaki açı dura yaması yapılan grupta ameliyat öncesi 123,7±11,7º ve ameliyat sonrası 129,8±11,1º (p=0,885); dura yaması yapılmayan grupta ameliyat öncesi 133.6±9.44º ve ameliyat sonrası 136.7±9.78º (p=0.376) idi ve ameliyat sonrası gruplar arasında belirgin bir fark saptanmadı (p=0.55, z=1.92). Dura yaması yapılan 3 hastada BOS fistülü gelişti ve belirtileri kötüleşen dura yaması yapılmayan bir hastaya dura yaması yapıldı ve hastada iyileşme gözlendi.

SONUÇ: Dura yaması yapılan hastalarla, yapılmayan hastalar karşılaştırıldığında hemen hemen benzer sonuçlar elde edilmektedir. Basit yöntemlerle başarı sağlanamadığı durumlarda her zaman daha agresif yöntem uygulama seçeneği mevcuttur.

ANAHTAR SÖZCÜKLER: Arnold Chiari Malformasyonu, Tedavi, Kombine tedavi

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INTRODUCTION

Hans Chiari's initial work on what would become known as Chiari Malformation (CM) was published in Deutsche Medizinische Wochenschrift in 1891 and entitled "Concerning alterations in the cerebellum resulting from cerebral hydrocephalus". The first type he described, which came to be known as Chiari Malformation Type I (CM-I), was characterized by "elongation of the tonsils and medial divisions of the inferior lobules of the cerebellum into cone shaped projections which accompany the medulla oblongata into the spinal canal". These features were demonstrated in a "relatively large percentage of cases of chronic congenital hydrocephalus but never without hydrocephalus or in cases of acute or later-developing hydrocephalus. The elongated portions of the cerebellum can show normal structure, fibrosis or softening and extend nearly to the top of the atlas, however in many cases to the undersurface of the axis" (2). There have been many studies on the pathogenesis of this malformation (1,3,10,32). Nishikawa et al (32) revealed after measurement of posterior fossa parameters that an underdeveloped occipital bone, possibly due to underdevelopment of the occipital somite originating from the paraxial mesoderm, induces overcrowding in the posterior cranial fossa, which contains the normally developed hindbrain in adult-type CM. Many treatment options were described for this malformation (29) in order to reduce the craniospinal pressure on the craniovertebral junction, to create a subarachnoidal space, and to reduce the volume of syringomyelic cavity and the pressure to the brainstem but the optimum approach is still unclear. On the other hand, Krieger et al. (26) evaluated the effectiveness of durotomy without duroplasty and reported effective treatment outcomes with rare occurrence of postoperative complications. We compared FMD with and without duroplasty to verify the effectiveness of foramen magnum decompression (FMD) without duroplasty in this retrospective study.

MATERIAL and METHODS

A total of 27 patients suffering from CM-I were operated on at our institute from 2003 to 2006. The histories were obtained from clinic charts, and the symptoms and their duration were determined. The diagnosis of CM-I and syringomyelia was demonstrated by magnetic resonance imaging (MRI). We did not detect any hydrocephalus in the neuroradiological examinations of our patient population. Indications for surgery were progressive/disabling symptoms as shown in Table I, Table II. The presence of a syrinx was also an indication for surgery when coupled with the aforementioned symptoms. Somatosensory evoked potential (SSEP) was performed pre-operatively. Follow-up MRI, SSEP, and detailed neurological examinations were performed in the third and twelfth postoperative months (Figure 1, 2). Clinical improvement was assessed from detailed clinical examinations and changes in SSEP examinations. The ratio of the syringomyelic cavity to spinal cord, the spinal-posterior fossa dural angle, the pre-operative tonsillar herniation from the foramen magnum and the pre- and postoperative tonsillo-dural distance were measured on the mid-sagittal plain of the craniocervical MRI (Figure 3, 4, 5). Syrinx improvement was defined as a decrease in syrinx cavity/spinal cord diameter ratio (Figure 3).

The patients were subdivided according to the use of duroplasty. Duroplasty was performed on 15 male patients aging 25.86±13. They were suffering

Table I: Symptoms of the Patients

Symptoms	Duroplasty group (%)	Non-duroplasty group (%)
Suboccipital headache	15(100)	12(100)
Neck and back pain	13(87)	11(92)
General imbalance	12(80)	12(100)
Tinnitus	10(67)	10(83)
Shoulder and arm pain	9(60)	10(83)
Visual phenomenon (stars in the vision)	8(53)	5(42)
Vertigo	7(47)	5(42)
Photophobia	5(33)	4(33)
Visual loss	4(27)	3(25)
Drop attack	4(27)	4(33)
Pressure in ear	3(20)	2(17)
Swelling disturbance	2(13)	3(25)

Table II: Neurological Signs of the Patients

Signs	Duroplasty group (%)	Non-duroplasty group (%)
No neurological findings	6 (40)	4 (33)
Epicritic paresthesia	6 (40)	5 (42)
Pathological reflex	3 (20)	2 (17)
Decreased touch sense	2 (13)	2 (17)
Paresis	2 (13)	2 (17)
Cerebellar signs	2 (13)	1 (8)

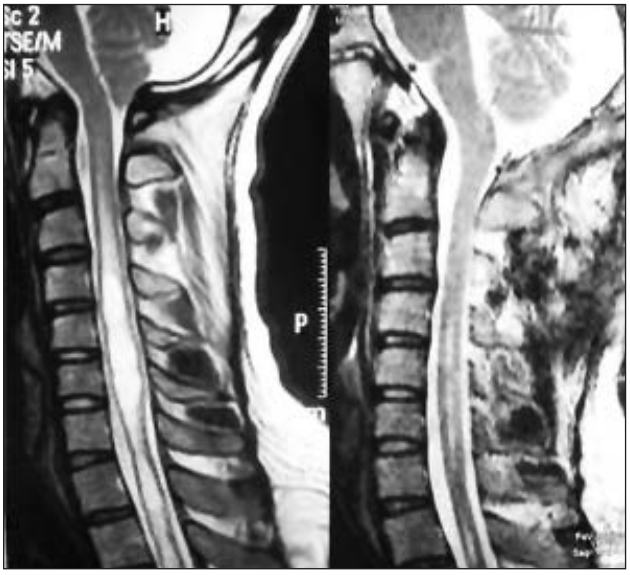

Figure 1: *Pre- (left) and post- (right) operative MRI of a patient from duroplasty group that was shown to have adequate FMD with syrinx shrinkage.*

from headache (100 %), neck and back pain (87%), ataxia (80%) and tinnitus (67%). No neurological pathology was detected in 40% of the patients and they were operated due to bothersome symptoms of CM-I. Epicritic paresthesia was found in 40% of the patients and paresis in 13% neurologically. Platybasia was diagnosed in two patients. Only FMD was performed to 9 male and 3 female patients aging 31.58±11.4. All patients were suffering from headache and ataxia, 92 % from neck and back pain and 83% from tinnitus. No neurological findings were examined in 33 % of the patients and 42 % of them had epicritic paresthesia. The patients without any neurological deficits were operated due to bothersome symptoms of CM-I. Platybasia was diagnosed in one patient. The symptoms and signs

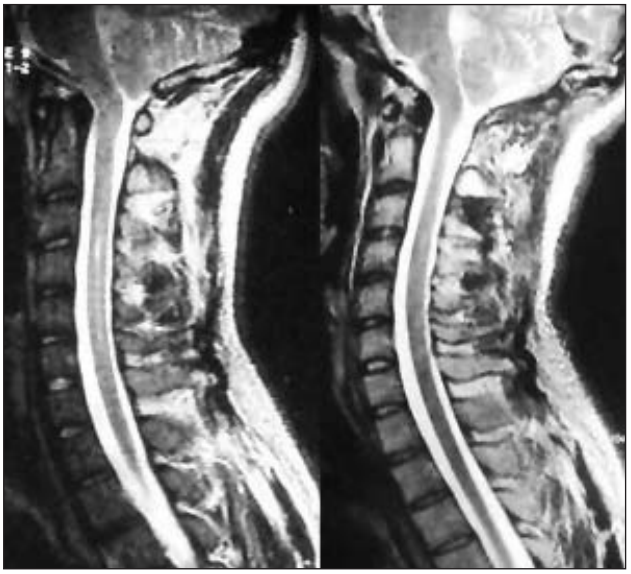

Figure 2: *Pre- (left) and post- (right) operative MRI of a patient from non-duroplasty group that was shown to have adequate FMD with syrinx shrinkage. The tonsillo-dural distance was shorter than duroplasty group, but spino-posterior fossa dural angle was wider than pre-op MRI.*

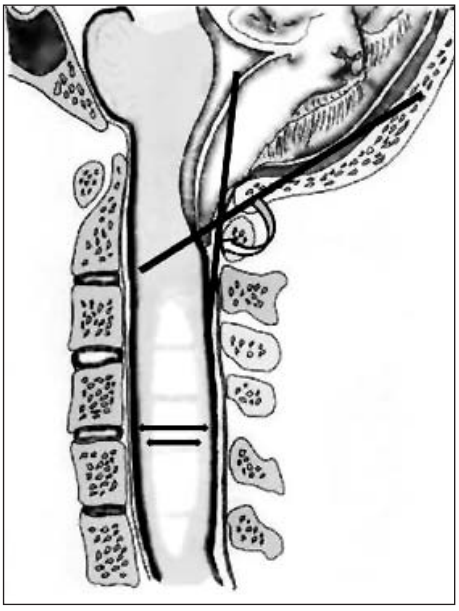

Figure 3: *The measurement technique of medullo-syringomyelic cavity diameter ratio from the widest part (black arrows), spino-posterior fossa dural angle (curved arrow)*

of the patients are shown in Table I and Table II. Pathological neural transmission was detected in five patients' SSEP examination pre-operatively. There were only three female patients with CM-I since our department mainly provides medical service to military personnel and their relatives.

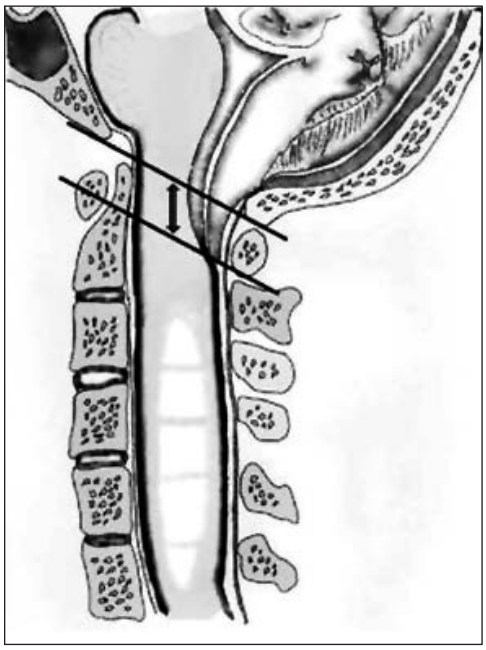

Figure 4: *The measurement technique of tonsillar herniation (black arrow)*

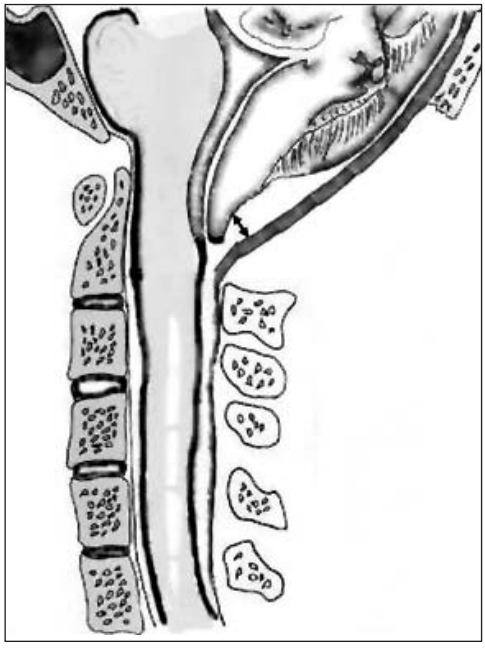

Figure 5: *The measurement technique of tonsillo-dural distance from the beginning of the tonsil (black arrow)*

Operative procedure

The specific surgical procedure (non-duroplasty / duroplasty) was chosen randomly. All surgeries were performed with the patient in the prone position. All patients underwent a decompressive suboccipital craniectomy extending at least 3 cm above the foramen magnum with a width of 3 cm. C1 laminectomy was performed totally. Atlantooccipital ligament and dural scarring or bands on the outside of the dura was removed. After that level, homeostasis and closure of the layers was performed in the non-duroplasty group. In the duroplasty group, the dura was opened in "Y" shape and dural grafting with cadaveric dura was followed by homeostasis and closure of the layers. A thick arachnoid layer was opened and thick arachnoid bands between the tonsils were removed microsurgically in only 5 patients. Passage of cerebrospinal fluid (CSF) was achieved.

Statistical Analysis

Standard supplemental statistical methods (mean, standard deviation and percentage) were used to evaluate the results of this study. The normality of the range of the parametric variables was evaluated graphically and with the Shapiro-Wilk test. Numbers and percentages were used to show the qualitative variables. Student's t test was used to correlate quantitative variables in respect to gender and treatment groups. Kruskal-Wallis one-way analysis of variance was used to compare pre-operative and postoperative syrinx ratio, tonsillar herniation, tonsillo-dural distance and spino-posterior fossa dural angles in respect to age groups. The Wilcoxon test was used on the matched samples to compare the pre- and postoperative variables. MS-Excel and SPSS for Windows Version 13.0 (SPSS INC. Chicago, IL, USA) were used for statistical analyses and calculation. $P<0.05$ was accepted as statistically significant in every conclusion.

RESULTS

On follow-up examination, 83.3% of the patients in the non-duroplasty and 73.3% in the duroplasty group were free of symptoms. The symptoms did not change in 13.3% of the duroplasty and 8.3% of the non-duroplasty group; and got worse 13.3% of the duroplasty and 8.3% of the non-duroplasty group. None of the patients but one in the non-duroplasty group had pathological findings on follow-up SSEP examination. Three patients showed decrease and two showed mild increase in spino-posterior fossa dural angle and one remained unchanged. When the groups were compared with respect to age groups, no statistically significance was found between the pre-operative and

postoperative syrinx ratio, tonsillar herniation, tonsillo-dural distance and spino-posterior fossa dural angles. Syrinx ratio showed regression in all but one patient at various degrees (Table V). Tonsillar herniation was 13.4±5.5mm in the non-duroplasty and 10±4.3mm in the duroplasty group (p = 0.330). Tonsillo-dural distance was 3.1±1.8mm in the non-duroplasty and 4.6±2.1mm in the duroplasty group postoperatively (p=0.507). When the tonsillo-dural distance was compared globally, a significant change was found postoperatively in both groups (p<0.005, z=4.47). The spino-posterior fossa dural angle was 133.6±9.44º preoperatively and 136.7±9.78º postoperatively in the non-duroplasty and 123.7±11.7º preoperatively and 129.8±11.1º postoperatively in the duroplasty group. No expansion was acquired in the spinal-posterior fossa dural angle in 26.6% of the duroplasty and 33.3% of the non-duroplasty group (Table V). When we compared the angles of the groups postoperatively, no difference was found (p=0.55, z=1.92). When we compared the symptoms (p=0.675) and signs (p=0.601) of each group, no significant difference was found postoperatively. All patients but one in duroplasty group showed regression of their syrinx ratio. The ratio of syrinx regression was 28±10% in the non-duroplasty and 36±33% in the duroplasty group. When the groups were compared postoperatively, no statistical significance was found (p>0,05). When the syrinx ratio was compared globally, a significant change was found postoperatively in both groups (p<0.05, z=3.30). One patient from the non-duroplasty group with narrowed spino-posterior fossa dural angle had progression of her symptoms and was re-operated with duroplasty. His symptoms were resolved.

Complications

One patient was re-operated due to open CSF fistula. Continued spinal drainage was used in two patients due to a closed subgaleal CSF fistula. These complications were seen in the duroplasty group.

DISCUSSION

CM-I is characterized by an abnormal position of the cerebellar tonsils, which herniate outside the cranial cavity into the upper cervical canal, associated with an obliteration of the subarachnoid

Table III: Pre-op and Post-op Measurement of the Patients from Non-Duroplasty Group

Age, sex	Syrinx ratio (pre-op) (%)	Syrinx ratio (post-op) (%)	Tonsillar Herniation (pre-op) (mm)	Angle (pre-op) (º)	Angle (post-op) (º)	Tonsillo-dural distance (pre-op) (mm)	Tonsillo-dural distance (post-op) (mm)	SEPP (pre-op)	SEPP (post-op)
21,m	-	-	14	140	147	0	2	-	-
22,m	-	-	19	127	140	0	7	-	-
22,m	-	-	11	130	155	2	5	-	-
28,m	77	46	14	132	148	1	4	-	-
25,m	40	32	5	152	131	1	5	-	-
42,f	33	27	12	130	135	2	3	-	-
56,f	-	-	11	135	132	1	3	+	-
23,m	-	-	24	118	118	0	2	-	-
52,m	-	-	5	144	136	0	1	+	-
38,f	50	33	17	142	132	0	1	+	+
21,m	-	-	11	125	137	1	2	-	-
29,f	-	-	18	128	130	2	3	-	-

SEPP: (+) having pathological finding; (-) no findings

Table IV: Pre-op and Post-op Measurement of the Patients from Duroplasty Group

Age, sex	Syrinx ratio (pre-op) (%)	Syrinx ratio (post-op) (%)	Tonsillar Herniatio (pre-op) (mm)	Angle (pre-op) (°)	Angle (post-op) (°)	Tonsillo-dural distance (pre-op) (mm)	Tonsillo-dural distance (post-op) (mm)	SEPP (pre-op)	SEPP (post-op)
28,m	-	-	12	140	151	1	4	+	-
24,m	-	-	10	128	133	2	7	-	-
27,m	70	60	8	115	123	1	7	-	-
21,m	86	72	4	113	134	0	4	-	-
21,m	82	80	15	133	124	2	7	-	-
30,m	33	30	7	105	112	0	3	-	-
25,m	40	30	11	130	144	0	4	-	-
21,m	83	33	22	127	135	0	9	-	-
21,m	50	30	10	112	128	0	3	+	-
20,m	60	0	7	140	120	0	5	-	-
67,m	-	-	8	127	120	1	3	-	-
21,m	40	40	6	126	134	0	2	-	-
21,m	75	13	10	102	120	0	5	-	-
21,m	70	33	8	130	123	1	1	-	-
20,m	-	-	12	128	146	1	5	-	-

Table V: The Distribution and the Variation of the Parameters According to Treatment Modality (**n:** patients' number, $p<0.05$ was significant when the groups were compared.)

Tonsillo-dural distance (mm)	**0-2**	**3-5**	**6-8**	**≥ 9**	**unchanged**	**reduced**	***p***
Duroplasty group (n)	2	10	1	1	1	-	>0.05
Non-duroplasty group (n)	8	3	1	-	-	-	
Angle (°)	**0-5 ↑**	**6-14 ↑**	**≥ 15 ↑**	**0-5 ↓**	**6-14 ↓**	**≥ 15 ↓**	***p***
Duroplasty group (n)	1	6	4	-	3	1	0.55
Non-duroplasty group (n)	3	3	2	1	2	1	
Tonsillar herniation (mm)	**< 5**	**6-10**	**11-15**	**16-20**	**>20**	***p***	
Duroplasty group (n)	1	9	4	1	-	0.658	
Non-duroplasty group (n)	-	2	6	3	1		
Syrinx reduction (%)	**0-10**	**11-20**	**21-30**	**31-40**	**41-50**	**>50**	***p***
Duroplasty group (n)	5	1	1	1	1	2	<0.05
Non-duroplasty group (n)	2	1	-	1	-	-	

spaces at the level of foramen magnum. Bony anomalies are seen in about one quarter of all patients with CM-I and include atlantooccipital assimilation, platybasia, basilar invagination, and fused cervical vertebrae. These are thought to be caused by an underdeveloped occipital bone, possibly due to underdevelopment of the occipital somite (4,6). Platybasia was diagnosed in three of our patients. Additionally, the displacement of one or both cerebellar tonsils 5 mm and more below the basion–opisthion line represents a basic feature for the neuroimaging diagnosis of CM-I in patients. Five to ten mm of displacement is considered a borderline malformation that becomes a pathological entity in the presence of associated anomalies like syringomyelia. Several mechanisms for the pathogenesis of syringomyelia have been proposed. According to the theories of Gardner and Angel (13) and Williams (39), the obstruction of CSF flow at the craniocervical junction causes CSF to enter the cervical central canal. However, such a communication does not exist in the majority of the patients (24,31) and does not explain the formation of the cavity. On the other hand, Oldfield et al. (34) suggested that CSF enters the spinal cord directly via the perivascular spaces, or so-called Virchow-Robin spaces, by arterial pulsations. Furthermore, Stoodley et al. (36) showed that CSF flows rapidly from the subarachnoid space to the perivascular spaces and this flow is dependent on arterial pulsations under normal conditions in a sheep model. With any obstruction to the flow of CSF in the central canal, such as at the craniocervical junction, a cavity can form that could subsequently enlarge.

Although the exact mechanism for the formation of the syringomyelia in CM-I is still controversial, there is general agreement on the importance of decompressing the craniocervical junction in the treatment. FMD, with/without duroplasty, serves to directly relieve the bony compression at the craniocervical junction. However, most authors differ on the usefulness and safety of additional procedures, such as duroplasty, syringosubarachnoid shunting, or obex plugging. The approaches preferred by American Pediatric Neurosurgeons are; only osseous decompression (20%); osseous decompression with dural grafting (30%); osseous decompression with dural grafting and intradural dissection of adhesions (25%); and osseous decompression with dural grafting, intradural dissection, and tonsillar manipulation and resection (30%) (18). Only 9% recommended performing decompressive surgery in asymptomatic patients. Furthermore, the decision of whether or not to operate is more difficult in these patients than in those with clear symptoms because of the lack of information about the natural course of this disease. We follow both asymptomatic and slightly symptomatic patients with CM-I with periodic clinical and radiological examinations in our daily practice, as suggested in the literature (8,27,33).

We compared two methods and discussed the results in the current study. The patients without any neurological deficit were operated in our series due to disturbing symptoms, which decreased the life-quality of the patients. While 83% of the patients were symptom-free with FMD in the duroplasty group, 73% of the patients were symptom-free in the FMD group but the difference was not statistically significant ($p=0.557$).

The authors found an increase in collagen fibers, hyalinous nodules, and calcification in the dural band of patients with CM-1 compared with healthy controls and advocated simple FMD (30). Adequate but not extensive osseous decompression should be performed due to the risk of cerebellar subsidence and recurrence of symptoms. Tokuno et al. (37) attempted to address the small posterior fossa by conducting an expansive suboccipital cranioplasty. We recommend performing a limited suboccipital craniectomy to enlarge the foramen magnum and allow for both decompression and maintenance of the posterior fossa neural elements and protect the patient from a high complication rate ($p<0.0001$). Dural elastance will be enough to build a cisterna magna in the postoperative three months if adequate bony decompression was performed in the surgery. Furthermore, Krieger et al. (26) recommend durotomy without duroplasty and have reported surgery-related outcome and complications comparable with those achieved by authors who advocate duroplasty, plugging of the obex, and tonsillar resection. Other authors have emphasized that duroplasty is essential for the prevention of scar formation and recurrent symptoms (9,10,12,15,28,29,34).

Debate still exists as to whether arachnoid dissection should be performed once the dura is opened. One benefit of arachnoid dissection, in the

setting of CM-I, is that it allows the surgeon to release adhesions that could potentially contribute to obstruction of CSF flow from the fourth ventricle to the spinal canal. We recommend opening the dura if the surgeon has any suspicion about maintaining the CSF flow in posterior fossa with the limited operation technique (thick fibrotic dura without any pulsation in non-duroplasty). If the arachnoid layer is too thick, and does not allow the surgeon to see the CSF flow, it must be opened, arachnoid bands must be cleaned and the CSF flow between the tonsils must be visualized. The arachnoid layer needed to be opened to visualize and maintain CSF flow in five of our patients. These patients were symptom-free in the follow-up examination but one was re-operated due to CSF fistula. In most papers in which outcome is discussed, the authors report an 80 to 90% rate of good outcome, with either resolution of the symptoms or cessation of progression. The most common complications include postoperative hemorrhage, CSF leakage, aseptic/ bacterial meningitis, and recurrence. Other reported complications include cervical instability and cerebellar subsidence. One patient was re-operated due to open CSF fistula; continued spinal drainage was used in two patients due to closed subgaleal CSF fistula in our series. One patient from the non-duroplasty group was re-operated due to progression of his symptom and duroplasty was performed. Inadequate dural band dissection was seen and dura was opened to perform duroplasty. Arachnoid bands blocking the CSF passage were seen and dissected. The symptoms of the patient resolved in the postoperative third month examination. Hida et al. (19) compared the results of two major surgical procedures - FMD and placement of a syringosubarachnoid shunt - used for the treatment of CM-I with associated syringomyelia in a mixed pediatric and adult population. Thirty-three patients (with a small syrinx) underwent FMD and 37 (with a large syrinx) underwent syringe-subarachnoid shunt treatment. Concerning the results obtained in the FMD group, postoperative MRI demonstrated a reduction in the syrinx size in 94% of the cases and clinical improvement was reported in 82%; a repeated operation was performed in three of 33 patients. Two cases of postsurgical meningitis and one case of postoperative kyphosis were observed.

Dyste and Menezes (10,11) reported 11 pediatric patients treated with FMD using dissection of the arachnoid, insertion of a dural graft, and placement of a fourth ventricle–subarachnoid shunt. Two patients also underwent a transoral odontoidectomy procedure. The condition of more than 87% of these patients improved significantly. Follow-up MRI performed in this group of patients' revealed collapse of the hydromyelic cavity. No operative complications or deaths were reported. Park et al. (35) reported on 68 pediatric patients, all of whom underwent FMD, C-l laminectomy, and dural graft placement. Cerebellar tonsil resection was performed in 40 of them. Forty patients had syringomyelia; in 32 of these patients the operation included the placement of a fourth ventricle–cervical subarachnoid space shunt, and 23 also underwent plugging of the obex. Within the first postoperative month all patients exhibited improvement in their symptoms, and 93% had improvement in their signs. Postoperative MRI showed a decrease in the syringomyelia in 93.3% of the cases. The authors believed that bone and dural decompression maximized the restoration of CSF circulation at the level of the foramen magnum. Tubbs et al. (38) reported on 130 pediatric patients who underwent suboccipital craniectomy, C-l laminectomy (one patient also underwent a C-2 laminectomy), and duroplasty in all but one of the cases. Stents extending from the fourth ventricle to the cervical subarachnoid space were placed in 26 patients. Seventy-five patients presented with syringomyelia, 22 of whom had unilateral tonsillar coagulation. Eight (36.3%) of the 22 underwent repeat surgery for persisting syringomyelia. Only one patient was treated with syringopleural shunt placement. Postoperatively two patients developed extraaxial subdural collections and acute hydrocephalus; one patient developed acute life-threatening signs of brainstem compression. Improvement in the clinical symptoms was observed in 83% of the cases. There has been a recent resurgence in the initial use of a syringosubarachnoid shunt in the treatment of patients with CM-I and syringomyelia (16,19-21). Although these reports suggest that this therapy leads to an increased rate of recovery and improved resolution of the syrinx, it must be noted that the underlying pathophysiology is not addressed. This approach is not recommended in the setting of

symptoms not related to spinal dysfunction. If sufficient decompression of posterior fossa could be established, surgeons must patiently wait for the regression of the syrinx. All but one patient showed regression by their syrinx ratio in our study. Extended surgical approaches like tonsillar coagulation can cause granulation tissue and arachnoid band regeneration that obstructs CSF flow despite adequate decompression.

MRI performed three months after the operation showed a decrease in syrinx size in fourteen of the fifteen patients, whereas a similar reduction was reported after a few months in the other patient. Genitori et al. (14) evaluated 26 pediatric patients with symptomatic CM-I, of whom 10 had syringomyelia; all were treated with simple bone decompression without dural opening. Improvement in or resolutions of clinical symptoms was recorded in almost all the children (97.2%), with disappearance of the syringomyelic cavity in 80% of the cases. Our data seem to confirm the hypothesis that performing the simple and limited FMD, often in combination with a C-1 laminectomy, in patients with CM-I can achieve comparable results with those obtained with more invasive surgical methods. Indeed, the percentage of good results in our series is similar to that reported in the literature when more aggressive surgical procedures were used (19,34,38).

Concerning the aspect of syringomyelic cavity reduction, the percentage of success is evidently lower than that reported when traditional procedures are performed but, in our opinion, even a moderate or no change on postoperative neuroimages may be regarded as a worthwhile result when accompanied by clinical improvement or complete recovery. Twenty-five (89.3%) of the 27 patients in our study, 15 of whom had syringomyelia, exhibited complete resolution or significant improvement of their preoperative neurological deficits, whereas in only 3 (10.7%) (two from the duroplasty and one from the non-duroplasty group) the condition remained unchanged or worsened. It should be noted that at least one of these negative results could be explained on the basis of rapid bone regrowth or granulation tissue or arachnoid band occurrence rather than an inappropriate surgical technique. In fact, repeating the bone decompression in association with dural delamination or releasing these arachnoid bands could achieve complete restoration of clinical wellness. One patient from the non-duroplasty group was re-operated due to progression of his symptoms and duroplasty was performed. Inadequate dural band dissection was observed and the dura was opened to perform duroplasty. Arachnoid bands blocking CSF passage were observed and dissected. The symptoms of the patient had resolved in the postoperative third month follow-up.

The low morbidity rate associated with the simplified approach contrasts with a significant incidence of postoperative complications reported in the literature in the case of "traditional" treatment (3,17,22). There was no morbidity related to the non-duroplasty procedure; no CSF leakage was observed in patients undergoing simple decompression. The duration of hospital stay was significantly shorter with the simplified procedure (mean 5.4 days) than in those undergoing traditional procedures (14.2 days). In the current study, clinical improvement was assessed by pre- and postoperative SSEP changes due to the sensitivity of SSEP in the evaluation of CM-I (5). In the non-duroplasty group, 10 patients (83.3%) exhibited a significant clinical improvement, whereas only a mild improvement in one (8.3%) and worsening in one (8.3%) were noted. Only one patient in the non-duroplasty group had pathological findings on follow-up SSEP examination. The correlation between clinical improvement and SSEP changes were prominent in the non-duroplasty group in our series. Thus, SSEP monitoring may be considered a valuable instrument for the evaluation of brain stem or upper cervical cord functional abnormalities in patients with CM-I. Regarding the duroplasty group, eight (73.3%) showed a significant improvement or complete recovery of their clinical symptoms, two (13.3%) a mild improvement and two (13.3%) worsening.

In a small subset of patients, a Chiari malformation can present with signs of raised intracranial pressure due to obstruction of cerebrospinal fluid flow or with raised intracranial pressure as the primary pathological driving force resulting in tonsillar herniation (23). We did not find any hydrocephalus clinically or in neuroradiological examinations in our patient population. The management of this entity is still controversial. Some authors suggest that CSF shunting is the best

approach due to hydrocephalus. On the other hand, acquired Chiari I malformation may develop as an intriguing late complication of CSF shunt surgery (7). In the recent literature due to the surgical inventions (e.g., endoscopic), some authors suggested that even in the CM-I-syringomyelia complex with normal or small ventricles, patients presenting with isolated signs and symptoms of raised intracranial pressure alone can be effectively managed with an electromagnetic-guided stereotactic endoscopic third ventriculostomy (23). Fundamentally, the understanding of the underlying pathogenetic mechanisms of CM-I with hydrocephalus may provide the best management, whether conservative or surgical.

The limitation of this study is the shortcoming evaluation of CSF flow study in our patient population. Basically, this study focused on the therapeutic effect of FMD performed by posterior approach without duroplasty in the comparison with duroplasty group. In the recent literature, some authors suggested that CSF flow measurements might give important information regarding the prognosis and follow-up of the patients with CM-I (25).

In contrast with the good clinical outcome, the neurological improvement was less rewarding, as the anatomical features of CM-I remained almost unchanged except spino-posterior fossa dural angle in most cases. The tonsillo-dural distance was minimally affected by the limited suboccipital craniectomy. However, minimal enlargement of the subarachnoid spaces at the craniocervical junction resulting from bone decompression was sufficient to relieve the impact on the nervous structures and to improve CSF circulation. The tonsillar herniation improved in a few cases, a finding that could support the hypothesis of arachnoid scarring as the main factor responsible for maintaining the abnormal position of cerebellar tonsils. These considerations raise the issue of the tonsil position being a useful criterion to evaluate the results of treatment of CM-I (24). A good outcome can be acquired if the expansion of spino-posterior fossa dural angle (133.6±9.44^{o} preoperatively and 136.7±9.78^{o} postoperatively) and tonsillo-dural distance (0.7±0.7mm preoperatively and 3.96±2.1mm postoperatively) can be achieved with the chosen approach.

CONCLUSION

Almost the same clinical outcomes can be achieved with PDF with and/or without duroplasty. The little difference should not let the surgeon to tend to invasive approach due to higher complication rates. There is always an option to perform duroplasty if simple procedure fails. The surgeons must wait at least three months for an extensive approach to allow cisterna magna formation by means of dural elastance. Narrowed spino-posterior fossa dural angle was measured in the patients with unfavorable outcomes. The narrowing of this angle shows inadequate decompression of posterior fossa and can be used on follow-up examination.

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