



# Comparison of the Surgical Results for Foramen Magnum Decompression with and without Duraplasty in Chiari Malformation Type 1

## *Tip 1 Chiari Malformasyonlu Hastalarda Duraplastili ve Duraplastisiz Foramen Magnum Dekompresyonunun Cerrahi Sonuçlarının Karşılaştırılması*

Mehmet Sabri GURBUZ<sup>1</sup>, Numan KARAASLAN<sup>2</sup>, Tezcan CALISKAN<sup>3</sup>, Emre UNAL<sup>1</sup>, Mehmet Zafer BERKMAN<sup>4</sup>

<sup>1</sup>Agri State Hospital, Department of Neurosurgery, Agri, Turkey

<sup>2</sup>Namik Kemal University, School of Medicine, Department of Neurosurgery, Tekirdag, Turkey

<sup>3</sup>Gaziosmanpaşa University, School of Medicine, Department of Neurosurgery, Tokat, Turkey

<sup>4</sup>Acibadem Maslak Hospital, Department of Neurosurgery, Istanbul, Turkey

Corresponding Author: Mehmet Sabri GURBUZ / E-mail: mehmetSabrigurbuz@gmail.com

### ABSTRACT

**AIM:** The surgical results for foramen magnum decompression (FMD) with and without duraplasty in Chiari Malformation type 1 (CM-1) were compared retrospectively.

**MATERIAL and METHODS:** Thirty-nine cases of CM-1 with and without syringomyelia (SM) were included. There were 18 patients in the non-duraplasty and 21 in the duraplasty group. Syringomyelia, tonsillar herniation (TH), preoperative symptom duration, and postoperative SM size were compared.

**RESULTS:** No significant difference was found between improvement in the duraplasty group (81%) and the non-duraplasty group (61.1%). In cases whose symptom duration was 0–36 months, improvement in the duraplasty group (93%) was significantly better than in the non-duraplasty group (50%) ( $p<0.01$ ). The rate of syrinx regression was 92.3% in the duraplasty group and 12.5% in the non-duraplasty group ( $p<0.05$ ). In cases with SM, the improvement was 21.4% in the non-duraplasty group compared to 78.6% in the duraplasty group ( $p=0.056$ ). In cases with TH greater than 10 mm, the improvement was 66.7% in the non-duraplasty group, whereas all six cases (100%) in the duraplasty group had improved.

**CONCLUSION:** In SM associated cases, cases with TH greater than 10 mm, and whose symptom duration is less than 36 months, duraplasty is a more reliable choice despite a slightly higher rate of complications.

**KEYWORDS:** Chiari malformation type 1, Syringomyelia, Duraplasty, Surgical results

### ÖZ

**AMAÇ:** Bu retrospektif çalışmada tip 1 Chiari Malformasyonlu (CM-1) hastalarda duraplastili ve duraplastisiz foramen magnum dekompresyonunun cerrahi sonuçları karşılaştırılmıştır.

**YÖNTEM ve GEREÇLER:** Sirengomyeli (SM) bulunan ve bulunmayan 39 CM-1 olgusu çalışmaya dahil edildi. duraplastisiz grupta 18, duraplastili grupta 21 olgu bulunmaktaydı. SM varlığı, tonsiler herniasyon (TH) miktarı, preoperatif semptom süresi ve postoperatif SM boyutu karşılaştırıldı.

**BULGULAR:** Duraplastili (%81) ve duraplastisiz grubun (%61,1) iyileşme oranları arasında anlamlı fark bulunmadı. Preoperatif semptom süresi 0-36 ay arasında olan olgularda; duraplastili grubun iyileşme oranı (%93) duraplastisiz gruba göre (%50) anlamlı oranda yüksekti ( $p<0.05$ ). Sirens boyutunda küçülme oranı duraplastili grupta % 92,3 iken duraplastisiz grupta % 12.5 idi ( $p<0.05$ ). SM bulunan olgularda; duraplastili grubun iyileşme oranı % 78,6, duraplastisiz grubun iyileşme oranı ise % 21,4 idi ( $p=0.056$ ). TH'un 10 mm'den fazla olduğu olgularda; duraplastisiz grubun iyileşme oranı % 66,7 iken, duraplastili 6 olgunun tamamı iyileşmişti (%100).

**SONUÇ:** SM'li olgularda, TH'un 10 mm'den fazla olduğu olgularda ve semptom süresi 36 aydan kısa olan olgularda duraplasti hafif yüksek komplikasyon oranı olmakla birlikte daha iyi bir seçenek olarak düşünülmüştür.

**ANAHTAR SÖZCÜKLER:** Tip 1 Chiari malformasyonu, Sirengomyeli, Duraplasti, Cerrahi sonuçlar

## INTRODUCTION

Chiari Malformation type 1 (CM-1) is defined as the downward displacement of the cerebellar tonsils and medial portions of the inferior cerebellar lobules through the foramen magnum into the upper cervical spinal canal. Obstruction of cerebrospinal fluid (CSF) at the level of the foramen magnum and thus altered CSF dynamics result in syringomyelia. CM-1 patients present with findings of either syringomyelia or brainstem compression (9, 25).

In 1883, John Cleland (29) described deformities of the brainstem, cerebellum, spina bifida and medullary cavitation. In 1891, Hans Chiari described three types of anomaly of the craniovertebral junction based on a few cases, and in 1895, described the three types and added a fourth type based on a higher number of cases (9, 29). In 1998, Iskandar et al. (12) described 'Chiari Zero malformation' to characterize a type of syringomyelia that exhibits classical CM-1 symptoms with little to no herniation. In 2010, Kim et al. described 'Chiari 1.5 malformation' as an advanced form of CM-1 (14).

Despite the fact that many theories have increased our knowledge about Chiari malformation (CM), the exact pathogenesis is still unclear (18). In 1943, Lichtenstein (16) was the first to mention the theory of a small posterior fossa and he pointed out that the reason for the downward displacement of the rhombencephalon is an underdeveloped posterior fossa. In 1978, Nyland and Krogness (23) reported a small posterior fossa in five cases of CM. The most accepted theory has been this 'theory of a small posterior fossa' (4, 20). After measurement of posterior fossa parameters, Nishikawa et al. revealed that an underdeveloped occipital bone induces overcrowding in the posterior cranial fossa, which contains the normally developed hindbrain in adult-type CM (22).

There is no consensus on the surgical treatment of CM-1 (25). Despite the fact that many different treatment modalities have been studied and proposed, there are two main modalities available. One of these is posterior fossa decompression with or without duraplasty to increase the posterior fossa volume and the other is different shunt procedures to reduce the cavity of syringomyelia (SM) (18). Some authors have advocated that posterior fossa decompression without duraplasty [suboccipital craniectomy, C1 (first cervical vertebra) laminectomy and C2 (second cervical vertebra) laminectomy when needed] is sufficient (9, 13, 26, 31), whereas others have advocated duraplasty (6–8, 25, 27). In this study, we compared the surgical results for posterior fossa decompression with and without duraplasty in the light of the current literature.

## MATERIAL and METHODS

From 2005 to 2012, 45 cases suffering from CM-1 with and without syringomyelia (SM) were operated at our institute. In total, 39 met our criteria and were analysed retrospectively. Cases were identified from the hospital medical archive from epidemiological information, clinical presentation, radiological findings, and the condition of the patients at discharge and at follow-up.

As in the literature, the following patients were excluded: those who were under 15 years of age, patients with other types of CM, patients with other associated malformations such as basilar invagination or other craniovertebral anomalies, and patients who had been treated with other methods such as syringosubarachnoid shunt (25). Indications for surgery were progressive disabling symptoms (Table I), in addition to radiological diagnosis. The first group was the non-duraplasty group (posterior fossa decompression only) consisting of 18 patients, and the second group was the duraplasty group (posterior fossa decompression and duraplasty) consisting of 21 patients. All patients underwent a decompressive suboccipital craniectomy extending at least 3 cm above the foramen magnum with a width of at least 4 cm, and total C1 laminectomy was performed as described in the literature (5, 9, 29, 31).

Yılmaz et al. (30) classified the amount of tonsillar herniation (TH) according to the spatial relationship between the herniated tonsils and the posterior arch of C1. As a simpler method, we classified the TH according to the length (mm) of tonsillar descent from the foramen magnum as either 5–10 mm or 10–20 mm. The preoperative symptom duration was divided into two groups as either 0–36 months or 36–120 months. The presence of SM and postoperative state of the syringomyelia cavity were taken into consideration. Postoperative states of the patients were classified as 'same and stabilized', 'improved' or 'worsened' as suggested in the literature (31).

## RESULTS

The ages of the patients ranged from 15 to 59 with a mean age of 36. The follow-up period was between 5 and 75 months with a mean of 43 months; 33.3% (n=13) of the patients were male and 66.7% (n=26) were female. Preoperative symptom duration was 0–36 months (n=25) in 64.1% and 36–120 months (n=14) in 35.9% of the patients. Preoperative

**Table I:** Preoperative Signs Symptoms

	n	%
Head-neck pain	29	74.4
Vertigo	1	2.6
Numbness	27	69.2
Drop attack	1	2.6
Gait disturbance	5	12.8
Imbalance	6	15.4
Cranial nerve problems	3	7.7
Cerebellar symptoms	6	15.4
Motor deficit	13	33.3
Extremity pain	5	12.8
Hearing abnormality	1	2.6
Impairment of temperature sensation	11	28.2
Hyperreflexia	10	25.6
Pathological reflex	9	23.1

symptoms were the same as in the literature (Table I). TH was 5–10 mm in 61.5% (n=24) and 10–20 mm in 38.5% (n=15) of the patients. SM was associated with 53.8% (n=21) of the patients.

In our study, foramen magnum decompression (FMD) alone was performed in 18 patients (non-duraplasty group), and FMD with duraplasty was performed in 21 patients (duraplasty group). No statistically significant difference was found between the surgical results for FMD with and without duraplasty. The rate of improvement was 37.5% in patients whose syrinx cavity size did not decrease and 84.6% in patients whose syrinx cavity size decreased postoperatively. The rate of postoperative syrinx regression was significantly higher in the duraplasty group (92.3%) than in the non-

duraplasty group (12.5%) ( $p < 0.01$ ) (Table II). Improvement was seen in 11 out of 13 (84.6%) cases with SM, and 3 out of 8 (37.5%) cases without SM (Table III). The rate of patients who remained unchanged was 15.4% in the duraplasty group and 62.5% in the non-duraplasty group.

Among the patients whose preoperative symptom duration was 36–120 months, no significant difference was found between the rate of improvement in the duraplasty and non-duraplasty groups. However, among the cases whose preoperative symptom duration was 0–36 months, the rate of improvement was 93% in the duraplasty group and 50% in the non-duraplasty group ( $p < 0.05$ ) (Table III).

Among the patients whose TH was 5–10 mm, no significant difference was found between the rate of improvement in

**Table II:** The Relationship Between the Postoperative Syrinx Size and the Surgical Technique

		Surgical technique		p
		Non-duraplasty	Duraplasty	
		n (%)	n (%)	
		<b>(n=8)</b>	<b>(n=13)</b>	
Postoperative syrinx size	Unchanged	7 (87.5%)	1 (7.7%)	<b>0.001**</b>
	Regressed	1 (12.5%)	12 (92.3%)	

Fisher's Exact Test, \*\* $p < 0.01$

**Table III:** The Relationship Between the Surgical Technique and Presence of SM, Preoperative Symptom Duration and the Amount of Tonsillar Herniation

		Surgical technique		p
		Non-duraplasty	Duraplasty	
		n (%)	n (%)	
<b>Not associated with SM</b>		<b>(n=10)</b>	<b>(n=8)</b>	
Surgical result	Unchanged	2 (20.0%)	2 (25.0%)	1.000
	Improved	8 (80.0%)	6 (75.0%)	
<b>Associated with SM</b>		<b>(n=8)</b>	<b>(n=13)</b>	
Surgical result	Unchanged	5 (62.5%)	2 (15.4%)	0.056
	Improved	3 (37.5%)	11 (84.6%)	
<b>0-36 months of symptom duration</b>		<b>(n=10)</b>	<b>(n=15)</b>	
Surgical result	Unchanged	5 (50.0%)	1 (6.7%)	<b>0.023*</b>
	Improved	5 (50.0%)	14 (93.3%)	
<b>36-120 months of symptom duration</b>		<b>(n=8)</b>	<b>(n=6)</b>	
Surgical result	Unchanged	2 (25.0%)	3 (50.0%)	0.580
	Improved	6 (75.0%)	3 (50.0%)	
<b>Tonsillar herniation 5-10 mm</b>		<b>(n=9)</b>	<b>(n=15)</b>	
Surgical result	Unchanged	4 (44.4%)	4 (26.7%)	0.412
	Improved	5 (55.6%)	11 (73.3%)	
<b>Tonsillar herniation of 10-20 mm</b>		<b>(n=9)</b>	<b>(n=6)</b>	
Surgical result	Unchanged	3 (33.3%)	0 (0.0%)	0.229
	Improved	6 (66.7%)	6 (100.0%)	

Fisher's Exact Test, \* $p < 0.05$

the duraplasty and non-duraplasty groups. However, among the cases whose TH was 10–20 mm, the rate of improvement was 66.7% in the non-duraplasty group, whereas all six cases (100%) in the duraplasty group improved (Table III).

Postoperative infection was seen in 5.6% of the non-duraplasty group and 19% of the duraplasty group. The rate of reoperation was 14.3% in the duraplasty group and 22.2% in the non-duraplasty group. Two cases of postoperative CSF fistula and one case of pseudomeningocele occurred in the duraplasty group. The overall complication rate was 5.6% in the non-duraplasty group and 28.6% in the duraplasty group which is slightly higher but not statistically significant.

### DISCUSSION

CM-1 constitutes a group of entities of congenital or acquired aetiology that have, in common, descent of the cerebellar tonsils into the cervical spinal canal (3,25). It is associated with syringomyelia in 30–70% of cases (1,10). Diagnosis of CM-1 is made based on both clinical and radiological findings (29). Clinical signs result either from compression of neural structures at the craniovertebral junction or from the cavity of SM (25,28).

Preoperative symptom duration is invariably long in CM-1 since it is a slowly progressive disease (7,8). Mean preoperative symptom duration was 7.6 years in the series studied by Matsumoto and Symon (19), 6.2 years in the series reported by Arruda et al. (1) and 5.4 years in the series studied by Dyste and Menezes (8). The preoperative symptom duration was 0–36 months in 64.1% of our patients and 36–120 months in 35.9% of our patients. In the series reported by Ramnarayan et al., when the duration of illness was less than 5 years, the rate of improvement increased (25). However, there are no data available in the literature mentioning the relationship between preoperative symptom duration and the success of the surgical technique in CM-1. In our series, among the patients whose preoperative symptom duration was 0–36 months, the rate of improvement was 93% in the duraplasty group whereas it was 50% in the non-duraplasty group, which is statistically significant ( $p < 0.05$ ). No significant difference was found between the duraplasty and non-duraplasty groups among the patients with preoperative symptom duration of 36–120 months.

Despite the fact that many different treatment modalities have been studied, two types of surgical procedure have generally been proposed. One of these is posterior fossa decompression with or without duraplasty to increase the posterior fossa volume, and the other is to use different types of shunt procedure to reduce the size of the syrinx cavity (18). Some authors (9, 13, 26, 31) have advocated that posterior fossa decompression without duraplasty is sufficiently effective, whereas others (6–8, 25, 27) have suggested adding duraplasty. Since shunt procedures have a potential risk of iatrogenic spinal cord injury, posterior fossa decompression with or without duraplasty remains the primary surgical technique whereas shunt surgeries are accepted as complementary surgical procedures (5, 6, 11).

James and Brant suggested FMD without durotomy as an effective treatment modality in children and teenagers (13). Erdogan et al. reported that the symptoms and signs had resolved in 83% of their patients after FMD without duraplasty but the rate of syrinx regression was only 28% (9). In another study including FMD without durotomy, 'very good' results were achieved in 66% of patients with 'good' results in 26% (31). Romero et al. reported that four out of six cases undergoing FMD without duraplasty had improved (26). In the series reported by Yilmaz et al, 24 out of 58 patients underwent FMD without duraplasty and clinical improvement was seen in 79.1% of the patients (30). Several groups have suggested that duraplasty is absolutely necessary and without it, tissue scarring and recurrence rates increase (6–8). Sindou et al. suggested opening of the dura with preservation of the arachnoid membrane and adding duraplasty, and reported favourable results (27). Improvement was seen in 20 out of 23 patients in the series reported by Ramnarayan et al. (25). In that series, 8 out of 11 patients benefited from posterior fossa decompression but the syrinx cavity regressed in only 50% of the patients. On the other hand, in the duraplasty group, 20 out of 23 patients improved and the syrinx cavity regressed in all of them (25). Klekamp reported postoperative improvement after 3 months as 82% of 359 patients having undergone posterior fossa decompression, arachnoid dissection, and an alloplastic duraplasty (15). In the duraplasty group in our study, 81% of the patients improved and 19% remained unchanged and stabilized, and none of them worsened. In the non-duraplasty group, 11 of 18 patients improved (61%) and the remaining seven patients (39%) remained unchanged or stabilized and none of them worsened. Taking all cases into consideration, no statistically significant difference was found between the surgical results for FMD with and without duraplasty.

Romero et al. reported that four out of six patients improved in the non-duraplasty group and nine out of 10 patients improved in the duraplasty group concluding that duraplasty is effective in cases with associated syringomyelia (26). Matsumoto and Symon found no significant difference between regression rates for the syrinx cavity in their duraplasty and non-duraplasty groups (19). Munshi et al. pointed out that surgical results are significantly worse with the non-duraplasty technique (21). Romero and Pereira reported that the syrinx regressed in all of five patients with syringomyelia who underwent duraplasty concluding that duraplasty is more effective in cases with associated syringomyelia (26). In our series, the syrinx cavity regressed in 92.3% of the duraplasty group compared to 12.5% in the non-duraplasty group which is statistically significant ( $p < 0.01$ ). On the other hand, the rate of improvement was 37.5% in patients whose syrinx remained unchanged and 84.6% in patients whose syrinx regressed ( $p = 0.056$ ). In cases with associated SM, the rate of improvement was 21.4% in the non-duraplasty group compared to 78.6% in the duraplasty group ( $p = 0.056$ ). In cases without SM, no difference was found between the duraplasty and non-duraplasty groups. In cases with SM,

15.4% remained unchanged in the duraplasty group whereas 62.5% remained unchanged in the non-duraplasty group.

Yilmaz et al. classified cerebellar tonsillar descent (CTD) as grade I (the tonsil descended more than 5 mm below the foramen magnum but did not reach the C1 arch), grade II (the tonsil reached the C1 arch) and grade III (the tonsil descended below the C1 arch) (30). There were no statistically significant differences between preoperative and postoperative size of the syrinx cavity and rate of improvement in the duraplasty and non-duraplasty groups in CTD grade I and II, but in CTD grade III, the decrease in size of the syrinx cavity and clinical improvement were statistically significantly better in the duraplasty group. In our study, there was no statistically significant difference between the duraplasty and non-duraplasty groups with TH of 5–10 mm, however with TH of 10–20 mm, the rate of improvement was 66.7% (6 out of 9 patients) in the non-duraplasty group compared to 100% (all of six cases) improvement in the duraplasty group ( $p > 0.05$ ).

The complication rate was slightly higher in the duraplasty group than in the non-duraplasty group (9). Parker et al. reported a 21.1% complication rate in their retrospective study including 114 patients in the duraplasty group, and in their series, the most frequent complications were aseptic meningitis, symptomatic pseudomeningocele and CSF fistula (24). Klekamp reported a 21.8% complication rate in a retrospective study including 359 patients in the duraplasty group (15). In the series reported by Hida et al., postoperative meningitis was seen in two and kyphosis in one out of 33 patients in the non-duraplasty group (11). Arruda et al. reported 23% pseudomeningocele and 6.6% meningitis in their series including 60 patients in the duraplasty group (1). In our study, the rate of complication was 20.5% which is in line with the literature. CSF fistula was seen in 5.1% of patients, meningitis in 12.8% of patients and pseudomeningocele in 2.6% of patients. The postoperative complication rate was 5.6% in the non-duraplasty group and 28.6% in the duraplasty group which is consistent with the literature. There were no statistically significant differences between the reoperation rates in the two groups. Seven cases needed reoperation, four of which were from the non-duraplasty group and three from the duraplasty group. In three patients in the non-duraplasty group, preoperative complaints recurred in the postoperative period and additional duraplasty operations were carried out. In one patient, the syrinx size increased again 3 months after surgery and a syringopleural shunt operation was performed. In the duraplasty group, two patients were operated for enlargement of the duraplasty and one patient had an additional C2 laminectomy.

The mortality rate was 3.1% in the series reported by Matsumoto and Symon (19) and 1.3% in the series reported by Logue and Edwards (17). Bindal et al. and Arruda et al. reported no mortality in their series (1, 2). No mortality was seen in our study.

## CONCLUSIONS

No statistically significant difference was found between the surgical results for FMD with or without duraplasty. However, in cases with associated SM, whose TH was greater than 10 mm and whose symptom duration was less than 36 months, duraplasty may be considered to be a more reliable choice despite a slightly higher rate of complications. However, these conclusions need to be verified in studies with larger series.

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