SURGICAL TREATMENT OF SYRINGOMYELIA WITH CHIARI MALFORMATION

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ABSTRACT

The subject of surgical approach and aetiology of syringomyelia is controversial. In our retrospective study, done between 1992-1998, we evaluated the results of different operative techniques applied to twelve patients with syringomyelia and associated Chiari malformation. Twelve patients underwent a total of sixteen operative procedures. Foramen magnum decompression with dural plasty (fascia lata or freeze-dried cadaveric dura) was performed on six patients. The foramen magnum decompression was achieved via suboccipital craniectomy and C1 and C2 laminectomy. Postoperative follow-up periods averaged 20 months (3-38 months). Of the twelve patients who had had preoperative symptoms and neurologic and physical signs, seven had improved, two had partially improved, one had no change and two had worsened. In conclusion, the patients who had the foramen magnum decompression with dural plasty showed a more favourable prognosis than those who had undergone the foramen magnum decompression or/and syringosubarachnoid shunting procedures.

Key words: Syringomyelia, Foramen magnum decompression, Chiari malformation

ÖZET

SİRİNGOMYELİ İLE CHİARİ MALFORMASYONUNUN CERRAHİ TEDAVİSİ

Siringomyelinin cerrahi tedavisi ve etyolojisi halen tartışmalıdır. 1992 ile 1998 yıllarını kapsayan dönemle ilgili olarak yaptığımız çalışmada Chiari malformasyonu ile birlikte olan siringomyelili 12 hastada uygulanan farklı cerrahi tekniklerin sonuçları değerlendirilmiştir. 12 hastaya toplam 16 cerrahi girişim uygulanmıştır. Operasyon sonrası takip ortalama 20 aydır. Foramen magnum dekompresyonu ve duraplasti uygulanan altı hastanın dördünde klıniksel iyileşme, ikisinde ise kısmen iyileşme görülmüştür. Siringosubaraknoid sant uygulanan iki hastadan birinde iyileşme, diğerinde ise kötüleşme görülmüştür. Foramen magnum dekompresyonu ve siringosubaraknoid şant uygulanan üç hastada ise, ikisinde iyileşme birinde kötüleşme saptanmıştır. Sadece foramen magnum dekompresyonu uygulanan iki hastanın biri düzelmiş, diğeri ise kısmen iyileşmiştir. Operasyon öncesi dönemde semptom ve nörolojik fonksiyon kayıpları bulunan toplam 12 olgunun 7'si iyileşmiş, ikisi kısmen düzelmiş, birinde değişiklik olmazken ikisi kötüleşmiştir. Özetle, foramen magnum dekompresyonu ve duraplasti yöntemi, tek başına foramen magnum dekompresyonu ve/veya siringosubaraknoid şant yöntemlerine oranla daha etkili yöntemdir.

Anahtar Sözcükler : Siringomiyeli, Foramen magnum dekompresyonu, Chiari malformasyonu

INTRODUCTION

The term "syringomyelia" is used for the presence of cystic cavities or syrinx in the spinal cord, often associated with other pathologies such as Chiari type 1 malformation, trauma and tumour(3,15). Of these, Chiari 1 malformation is the most frequently encountered craniocervical junction deformity, characterised by a downward herniation of cerebellar tonsils through the foramen magnum without displacement of hindbrain (17). Two theories have been proposed. Gardner has proposed the "hydrodynamic theory", in which an extension of syringomyelia results from the "water-hammer" effect of pulsatile transmission of cerebrospinal fluid from the fourth ventricle to the cavity in the spinal cord(6). In an alternative theory proposed by Williams, the

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obstruction at the level of the foramen magnum leads to a dissociation of pressure between the cranial and spinal cerebrospinal fluid compartments. This causes the fluid to be sucked from the fourth ventricle into the central channel(29).

From the point of view of surgical approach to syringomyelia, there is no consensus among authors. Many surgical modalities have been applied, including shunting procedures(24,28) and foramen magnum decompression with or without closure of dura(11,19). Foramen magnum decompression with duraplasty has been widely accepted in the treatment of syringomyelia associated with Chiari 1 malformation (4,10,25).

PATIENTS and METHODS

Twelve patients with syringomyelia related with Chiari 1 malformation were operated on between 1992 and 1998 at the Neurosurgery Department of the Medical Faculty of Osmangazi University. 7 of the patients were male and 5 female. Their ages at operation ranged from 14 to 50 years (mean 36.1 years). Presenting symptoms were dysesthesia, weakness, occipital and cervical pain, difficulty in walking, back pain, dyspnea and pain in the arm. Neurologic and physical signs were dissociated sensory loss, motor weakness, pathologic reflex, atrophy, gait abnormality, bowel dysfunction and bladder dysfunction.

Table 1. The major symptoms and clinical signs of the patients

Symptoms		Signs	
Dysesthesia	10(83%)	Dissociated sensory loss	10(83%)
Occipital and cervical pain	6(50%)	Motor weakness	6(50%)
Weakness	6(50%)	Pathologic reflex	4(33%)
Difficulty in walking	5(41%)	Atrophy	4(33%)
Back pain	2(16%)	Gait abnormality	2(16%)
Pain in arm	2(16%)	Bowel dysfunction	1(8%)
Dyspnea	1(8%)	Bladder dysfunction	1(8%)

The most common symptoms were dysesthesia, pain and weakness, and the signs were dissociated sensory loss and motor weakness. Preoperative duration of symptoms and signs ranged from 18 months to 9 years (mean 49.7 months).

Properative imaging consisted of plain radiography of skull and spine (12 patients), cranial computed tomography (4 patients), spinal computed tomography (4 patients) and spinal magnetic resonance imaging (12 patients).

Twelve patients with syringomyelia associated with Chiari malformation underwent a total of sixteen operative procedures. Foramen magnum decompression with duraplasty was performed on six patients,

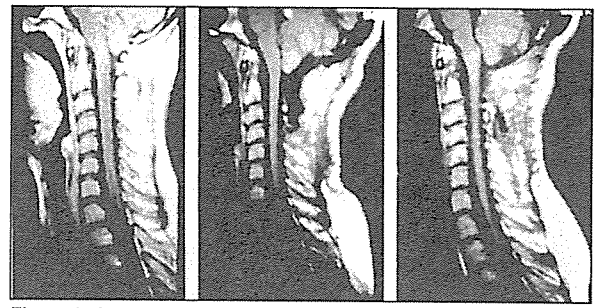


Figure 1a.b.c.
Preoperative (a), postoperative second month (b) and postoperative seventh month (c) magnetic resonance imaging of patient with syringomyelia treated with foramen magnum decompression and duraplasty

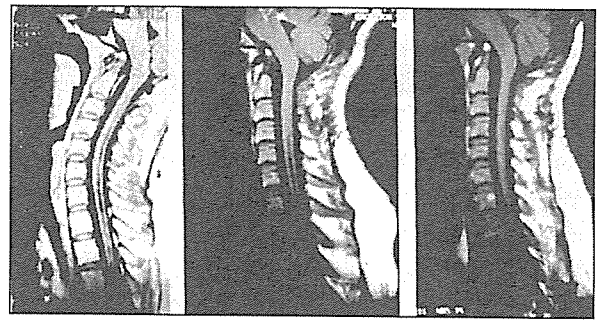


Figure 2a.b.c.
Preoperative (a), postoperative seventh month (b) and postoperative tenth month (c) magnetic resonance imaging of patient with syringomyelia treated with foramen magnum decompression and duraplasty

foramen magnum decompression (dura open) in two patients, syringosubarachnoid shunting and foramen magnum decompression in three patients, and syringosubarachnoid shunting by itself in two patients. Foramen magnum decompression was achieved via suboccipital craniectomy, cercival first and second vertebrae laminectomy. The dura mater was opened, with care taken to preserve the arachnoid membrane

Table 2. Clinical and radiological results of the patients

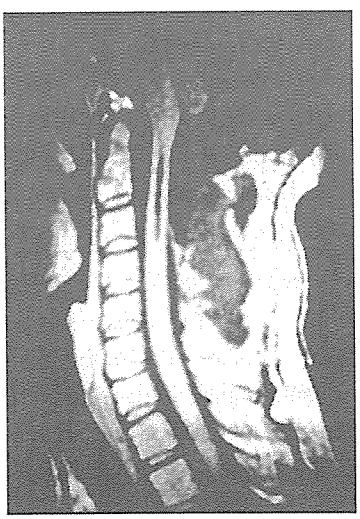
Patient	Age	Sex	Postop. cli.	Postop. rad.	Operation	Level
1	26	F	İmproved	İmproved	FMD and dura (f.lata)	C7 - T2
2	38	F	Par. impro	No change	FMD and dura (cadave.)	C1 - C5
3	47	M	İmproved	İmproved	FMD and dura (cadave.)	C2 - C5
4	26	F	İmproved	No change	FMD and dura (cadave.)	C3 - C7
5	14	F	İmproved	Par. impro	FMD and dura (f.lata)	C2 - C5
6	54	F	Par. impro	Par. impro	FMD and dura (cadave.)*	C2 - C7
7	31	M	İmproved	Par. impro	SSS and FMD	C2 - T1
8	31	M	Worsened	No change	SSS and FMD	C1 - C5
9	33	F	İmproved	No change	SSS and FMD	C2 - T2
10	50	M	İmproved	No change	FMD	C1 - C6
11	43	M	Worsened	No change	SSS	C6 - T5
12	41	F	No change	No change	SSS	C4 - T2

FMD : Foramen magnum decompression, Dura : Duraplasty, Cadave: Freeze - Dried cadaveric dura, F.lata: Fascia lata, Postop. Cli.: Post-operative clinical status, Postop. Rad.: Post-operative radiological evaluation. Two months later, duraplasty was performed due to CSF leakage.

intact. Fascia lata was used for duraplasty in two cases and freeze-dried cadaveric dura in four cases.

The postoperative follow-up period averaged 20 months (range 3-38 months). Of the twelve patients who had had preoperative symptoms and neurologic and physical signs, seven had improved, two had partially improved, one had no change and two had worsened. Mortality was zero in the pre-operative and early post-operative period. Syringosubarachnoid shunting was performed in five cases. Three of the five patients underwent a second operation (foramen magnum decompression). As shown in Table II, two of the three improved and one worsened. Foramen magnum decompression and duraplasty was performed in six cases. Four of these improved (66%) and two (33%) partially improved.

Postoperative radiological examination consisted of spinal X-ray and spinal magnetic resonance imaging. In patients on whom foramen magnum decompression was performed with duraplasty, magnetic resonance imaging studies showed the new cisterna magna formation. Of these patients two improved, two remained unchanged and two worsened radiologically. No correlation was observed between radiological and clinical improvements.



Complications: Subcutaneous cerebrospinal fluid accumulation was observed in 3 patients with foramen magnum decompression with duraplasty. (Figure 3)

Figure 3. Subcutaneous cerebrospinal fluid accumulation

In one of the two patients with foramen magnum decompression without duraplasty, there was severe subcutaneous cerebrospinal fluid accumulation and leakage that required repeated duraplasty.

DISCUSSION

Syringomyelia is cavitary enlargement of the spinal cord. It is usually characterized by slowly-progressing dissociate sensory deficit and associated with craniovertebral anomalies such as Chiari malformation. Chiari malformation 1 was found in 57 % of cases with syringomyelia(20). The other causes include basilar impression, basilar arachnoiditis, Dandy-Walker cysts, suboccipital encapholocele, foramen magnum cysts and foramen magnum tumours(16).

Abbe and Coley made the first surgical treatment (Syringostomy) on syringomyelia in 1891(3). Laminectomy was performed in the treatment of syringomyelia for a number of years(1,21). Surgical procedures such as syringostomy, terminal ventriculostomy and myelotomy have been tried in past years but the results of these procedures have been poor (2,13). There are several surgical techniques for syringomyelia now considered acceptable. These are percutaneous drainage(12), syringopleural shunting(28), syringosubarachnoid shunt(23), ventriculoatrial or ventriculoperitoneal shunting(18), cerebellar tonsil resection(9), foramen magnum decompression and obex plugging(6). The aim of treatment in patients with syringomyelia associated with Chiari malformation is relief of the syringomyelia and stabilisation/regression of symptoms and signs.

In 1976, Rhoton reported his experience of microsurgical exploration on 11 adults with Chiari malformation associated with hydromyelia, treated by suboccipital craniectomy, upper cervical laminectomy, creating an outlet from the fourth ventricle and opening the distended cord in the thinnest exposed area, usually along the dorsal root entry zone. Only one poor result had been seen (22).

Tator et al. (1982) advocate the syringosubarachnoid shunting procedure as a treatment of syringomyelia. They report a series of 20 patients treated by syringosubarachnoid shunting. 15 of the patients had idiopathic syringomyelia, four had post-traumatic syringomyelia and one, syringomyelia secondary to spinal arachnoiditis. An excellent or good long-term result was obtained in 15 patients (75%). They also note the role of microscope operating technique in this study(24).

Treating twenty-seven patients with traumatic syringomyelia, Vernon (1983) performed three types of operation: cord transsection, tube syringostomy to the subarachnoid space, and tube syringostomy to the peritoneal cavity. Fourteen patients improved after treatment, four patients showed no change and six patients deteriorated but improved after further operations. Three patients deteriorated but underwent no further operations. He emphasises the advantage of shunting procedures in cases of this type(29).

In 1983, Williams evaluated the terminal ventriculostomy performed on thirty-one patients with syringomyelia. Seventeen of these patients had had some previous surgical intervention. Eleven patients were in the process of deterioration at the time of the operation. Postoperative improvement was reported in twenty one patients. Sixteen of the patients who improved have since proceded to deteriorate. Thirteen patients sustained no improvement and some of these have continued to deteriorate(31).

Grant et al. (1987) report 9 patients with syringomyelia. They found that in patients with syringomyelia and a posterior fossa abnormality, the syrinx became smaller after foramen magnum decompression alone, without any plugging of the obex. Their observations support the view that foramen magnum obstruction is important in the pathogenesis of syringomyelia and an appropriate target for treatment. Neither before nor after operation was any communication between the 4^{th} ventricle and the syrinx observed(7,8).

In 1988, Filizzolo reported a review of the

treatment of thirty cases of syringomyelia over seven years, with the following results(5):

1) One of the six patients treated by terminal ventriculostomy showed an improvement, while five continued to deteriorate. Three of them needed a foramen magnum decompression and one, a syringoperitoneal shunt.

2) Of the twenty-seven patients treated by foramen magnum decompression 20 improved, 4 remained unchanged and 3 worsened.

In 1989, Matsumoto and Symon published their retrospective study of 98 patients with syringomyelia. They proposed craniovertebral decompression and duraplasty as the primary intervention of choice for all syringes associated with Chiari malformation, to regulate the abnormal hydrodynamic forces created by the malformation ultimately resulting in collapse of the syrinx. They advocated implanting a syringoperitoneal shunt as a primary operation only in cases where the syrinx is not associated with hind brain abnormality. In patients whose cysts and symptoms persisted following the primary intervention, they recommended syringoperitoneal shunting as a secondary procedure. They also point out the increased success of shunting procedures (syringoperitoneal and syringosubarachnoid) and the improved quality of shunts due to modern microsurgical techniques. Acceptable results were obtained in 60% of the cases (14).

In 1991, Vengsarkar reported three cases of syringomyelia associated with Chiari 1 malformation, where the clinical manifestation was myelopathy. All patients were successfully treated by percutaneous placement of the thecoperitoneal shunt. Two of these patients had undergone craniovertebral decompression in other centers. Post-operative MRI showed considerable shrinkage of the cysts corresponding with clinical improvement (27).

Van der Bergh (1991) presented one case where trauma had played a role in triggering classical syringomyelia, forming an intraparanchymatous cavity, extended in the cranial direction by the pulsating cerebrospinal fluid (26).

In recent years, there are two favoured surgical procedures for cases of syringomyelia raleted with Chiari malformation. First, shunting systems, especially syringosubarachnoid shunting, and second, foramen magnum decompression and duraplasty. At our clinic, we have been performing syringosubarachnoid shunting for treatment of syringomyelia for many years. An important complication of SSS has been damage to the spinal cord at the catheter entry foramen. In addition, the catheter can only work if the pressure in the syringes is higher than that in the subarachnoid space. In shunting procedures, the complication rate for the catheter (displacements and obstructions) was 12.8%(23). The results of our patients undergoing syringosubarachnoid shunting and/or foramen magnum decompression (dura open) were not favourable. On the other hand, improvement was observed in sixty-six percent of patients undergoing foramen magnum decompression and duraplasty. We conclude that the foramen magnum decompression and duraplasty procedure is more effective than syringosubarachnoid shunting in cases of syringomyelia related with Chiari malformation is controversial, to date, no surgical procedure has been acknowledged as the ideal one.

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REFERENCES

1. Ballantine HT, Ojemann RG, Drew JH: Syringohydromyelia. Progr Neurol Surg 4:27-245, 1971

2. Cahan LD, Bentson JR: Consideration in the diagnosis and treatment of syringomyelia and the Chiari malformation. J Neurosurgery 57:24-31, 1982

3. Donauer E: 100 years syrinx-surgery. Acta Neurochirur 123: 157-225, 1993

4. Erdinçler P, Canbaz B, Oğuz E, Sanus GZ, Dashti R, Özyurt E, Kuday C: Surgical management of hydrosyringomyelia related with Chiari 1 malformation. Turkish Neurosurgery 8:6-12, 1998

5. Filizzolo F, Versari P, D'Aliberti G, Arena O, Scotti G, Mariani C: Foramen magnum decompression versus terminal ventriculostomy for the treatment of syringomyelia. Acta Neurochir 93:96-99, 1988

6. Gardner WJ, Angel J: The mechanism of syringomyelia and its surgical corrections. Clin Neurosurg 6:131-140, 1959

7. Grant RH, Hadley DM, Macpherson P, Condon B, Patterson J, Bone I, Teasdale GN: Syringomyelia: Cyst measurement by magnetic resonance imaging and comparison with symptoms, signs and disability. J Neuro Neurosurg Psychiatry 50:1008-1014, 1987

8. Grant R, Hadley D.M, Lang D, Condon B, Johnson R, Bone I, Taesdale G.M: MRI measurement of syrinx size before and after operation. J Neuro Neurosurg Psychiatry 50:1685-1687, 1987

9. Guyotat J, Bret P, Mottolese C, Jouhanneau E, Abdurrahman M, Lapras C: Chiari 1 malformation with syringomyelia treated by decompression of the cranio-spinal junction and tonsillectomy. Apropos of 8 cases. Neurochirurgie 43(3):135-141, 1997

10. Imae S: Clinical evaluation on aetiology and surgical outcome in syringomyelia associated with Chiari type 1 malformation. No To Shinkei 49(12):1131-1138, 1997 (Abstract)

11. Klekamp J, Batzdorf U, Samii M, Bothe HW: The surgical Treatment of Chiari malformation. Acta Neurochir 138:788-801, 1996

12. Levy WJ, Rosenblatt S, Russell E: Percutaneous drainage and serial magnetic resonance imaging in the diagnosis of symptomatic posttraumatic syringomyelia: case report and review of the literature. Neurosurgery 29:429-434, 1991

13. Logue V, Edwards MR: Syringomyelia and its surgical treatment-an analysis of 75 patients. J Neurol Neurosurg Psychiatry 44:273-284, 1981

14. Matsumoto T, Symon L: Surgical management of syringomyelia-current results. Surg Neurol. 32:258-265, 1989

15. Milhorat TH, Capocelli AL, Anzel AP, Kotzen RM, Milhorat RH: Pathological basis of spinal cord cavitation in syringomyelia: analysis of 105 autopsy cases. J Neurosurg 82:802-812, 1995

16. Milhorat TH, Miller JI, Johnson WD, Adler DE, Heger BA: Anatomical basis of syringomyelia occurring with hindbrain lesions. Neurosurgery 32(5): 748-753, 1993

17. Nishikawa M, Sakamoto H, Hakuba A, Nakanishi N, Inoue Y: Pathogenesis of Chiari malformation: a morphometric study of the posterior cranial fossa. J Neurosurg 86: 40-47, 1997

18. Oi S, Kudo H, Yamada H: Hydromyelic hydrocephalus. Correlation of hydromyelia with various stages of hydrocephalus in postshunt isolated compartments. J Neurosurgery 74: 371-379, 1991

19. Oldfield EH, Muraszko K, Shawker TH, Patronas NJ: Pathophysiology of syringomyelia associated with Chiari malformation of the crebellar tonsils. J Neurosurgery 80:3-15, 1994

20. Pillay PK, Awad IA, Little JR, Hahn JF: Symptomatic chiari malformation in adults a new classification based on magnetic resonance imaging with clinical and prognostic significance. Neurosurgery 28:639-645,1991

21. Pitts FW, Groff RA: Syringomyelia: current states of surgical therapy. Surgery 56:806-809, 1964

22. Rhoton Albert L: Microsurgery of Arnold Chiari

malformation in adults with and without hydromyelia. J Neurosurgery 45: 473-483, 1976

23. Sgouros S, Williams B: A critical appraisal of drainage in syringomyelia. J Neurosurgery 82: 1-10, 1995

24. Tator CH, Meguro K, Rowed DW: Favourable results with syringosubarachnoid shunts for treatment of syringomyelia. J Neurosurgery 56:517-523, 1982

25. Vanalocha V, Saiz-Sapena N, Garcia-Casaola MC: Surgical technique for craniocervical decompression in syringomyelia ssociated with Chiari type 1 malformation. Acta Neurochirur 139(6): 529-539, 1997

26. Van der Berg R: Pathogenesis and treatment of delayed post-traumatic syringomyelia. Acta Neurochirur 110:82-86, 1991

27. Vengsarkar US, Panchal VG, Tripathi PD, Patkar SV, Agarwal A, Doshi PK, Kamat MM: Percutaneous thecoperitoneal shunt for syringomyelia. J Neurosurgery 74: 827-831, 1991

28. Vernet O, Farmer J, Montes JL: Comparison of syringopleural and syringosubarachnoid shunting in the treatment of syringomyelia in children. J Neurosurgery 84: 624-628, 1996

29. Vernon JD, Silver JR, Symon L: Post-traumatic syringomyelia. Paraplegia 21:37-46, 1983

30. Williams B: Simultaneous cerebral and spinal fluid pressure recordings. 2.cerebrospinal dissociation with lesions at the foramen magnum. Acta Neurochirur 59:123-142, 1981

31. Williams B, Fahy G: A critical appraisal of "Terminal ventriculostomy" for the treatment of syringomyelia. J Neurosurgery, 58: 188-197, 1983

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