Results of Surgical Treatment for Symptomatic Chiari I Malformation in Adults

Erişkin Semptomatik Chiari Tip I Malformasyonlarında Cerrahi Tedavi Sonuçları

ZEKİ ŞEKERCİ, NEZİH ORAL, NURİ ARDA, UYGUR ER

Ankara Social Security Hospital, Department of Neurosurgery, Ankara, Türkiye

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Abstract: This study presents the results of epiarachnoidal posterior fossa decompression for 20 adults who had symptomatic Chiari I malformation with syringomyelia, and discusses the efficacy of this technique.

Key words: Chiari I Malformation, syringomyelia, posterior fossa decompression

Özet: Bu çalışmada epi anahnoid posterior fossa dekompresyonu yapılmış 20 syringomyelisi olan semptomath chiari Tip I malformasyonlu hastanın sonuçları sunulmuş ve bu tekniğin etkinliği tartışılmıştır.

Anahtar Sözcükler: Chiari I malformasyonu, siyringomyeli, posterior fossa dekompresyonu

INTRODUCTION

Chiari I malformations are characterized by caudal descent of the cerebellar tonsils to below the foramen magnum. In 50-76% of patients, the malformation is associated with hydromyelic cavitation of the spinal cord and/or medulla oblongata (1,9). The use of magnetic resonance imaging (MRI) techniques facilitates accurate preoperative diagnosis and postoperative follow-up of patients with Chiari I malformations and associated anomalies. MRI also provides detailed anatomic information in these patients (7). Although various surgical methods have been proposed to treat Chiari I malformation with syringomyelia, to date there is no preferred surgical approach (8). All of the surgical options remain controversial, and data is lacking with regard to results for these methods (2,7,8). Posterior fossa decompression leaving the arachnoid layer intact appears to be more effective than other methods of treating Chiari I malformation with syringomyelia. Oldfield and coworkers operated on seven patients with Chiari I malformations and syringomyelia, and achieved bone and dural decompression of the foramen magnum without entering the subarachnoidal space. They used ultrasound intraoperatively and dynamic MRI techniques postoperatively to demonstrate flow increasing in the CSF channels and the unobstructed transmission of systolic waves across the foramen magnum. Syringomyelia resolved within 1 to 6 months of surgery in all of their patients (5).

MATERIALS AND METHODS

Between 1992 and 1996, 20 adults with symptomatic Chiari I malformation underwent surgical treatment at our neurosurgery department. We retrospectively analyzed the management of and outcomes for these individuals. The patients were 21 to 57 years old, with a mean age of 44 years. The study group included 11 females and 9 males. Presurgery

MRI of the cranium, the craniocervical junction, and the entire spinal cord was done in all cases. Repeat MRI was done at months 3 and 24 postsurgery, and was compared with preoperative images to assess syrinx diameter, CSF flow at the craniovertebral junction, and hydrocephalus. Preoperative neurological examinations and plain x-rays of all patients were done at our clinic, and results were kept on file.

The duration of symptoms ranged from 1 to 72 months, with an average of 50 months. Follow-up ranged from 3 to 36 months. All the patients had cervical, bulbar, and/or thoracic syringomyelic cavitations (Figures 1a,2a,3a,4a). Hydrocephalus was identified in two patients, scoliosis in four, the Klippel-Feil sign in one patient, a temporal arachnoid cyst in one patient, and odontoid compression of the anterior cervical cord in one patient (Table I). The most frequent complaint was pain (Table II). Ninety percent of patients suffered from pain in the occipitocervical region, occasionally with radiation to the neck and arm following coughing or valsalva maneuver. The patients with hydrocephalus also reported having severe headaches. Sensory loss was the second most common symptom, seen in 70% of the patients. Weakness, disequilibrium, vertigo, difficulty swallowing, diplopia, tinnitus, and vomiting were the other complaints (Table II). Ataxia was the most common preoperative sign (30% of patients), and other signs included paralysis of various cranial nerves, reflex abnormalities, papillary edema, nystagmus, and skeletal abnormalities (Tables II,III and). Six patients had central cord-like syndrome with upper extremity weakness and decreased deep tendon reflexes (Table III). Dissociative sensory loss was identified in 25% of the patients (Table III).

Table I: Associated anomalies in the 20 Chiari Type I patients studied.

Anomalies	No. of patients (n=20)	%
Syringomyelia*	20	100
Scoliosis	4	20
Hydrocephalus	2	10
Klippel-Feil sign	2	10
Odontoid compression	1	5
Intracranial arachnoid cyst	1	5

*Syrinx located in cervical and/or bulbus region in 14 cases, in cervicothoracic region in 6 cases

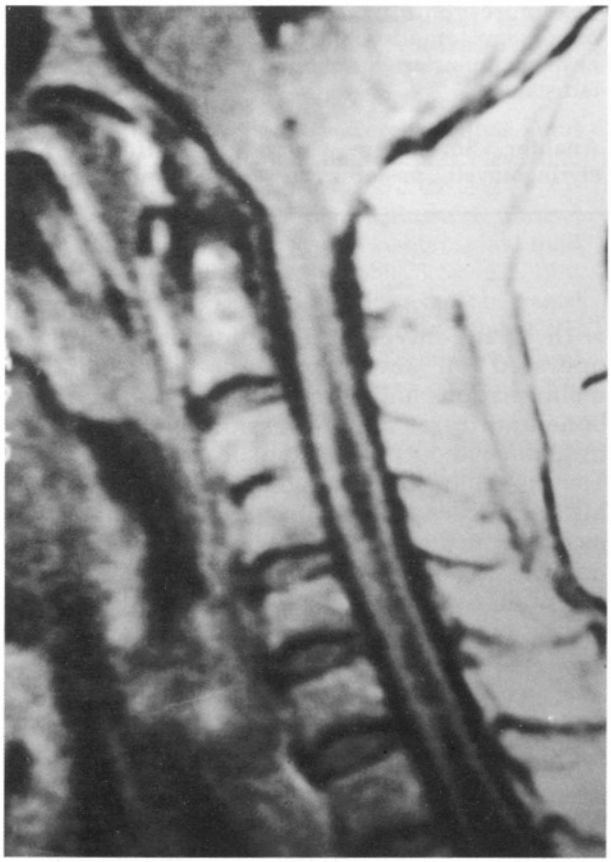

Figure 1a: Preoperative MRI image of a Chiari I malformation and syringomyelia.

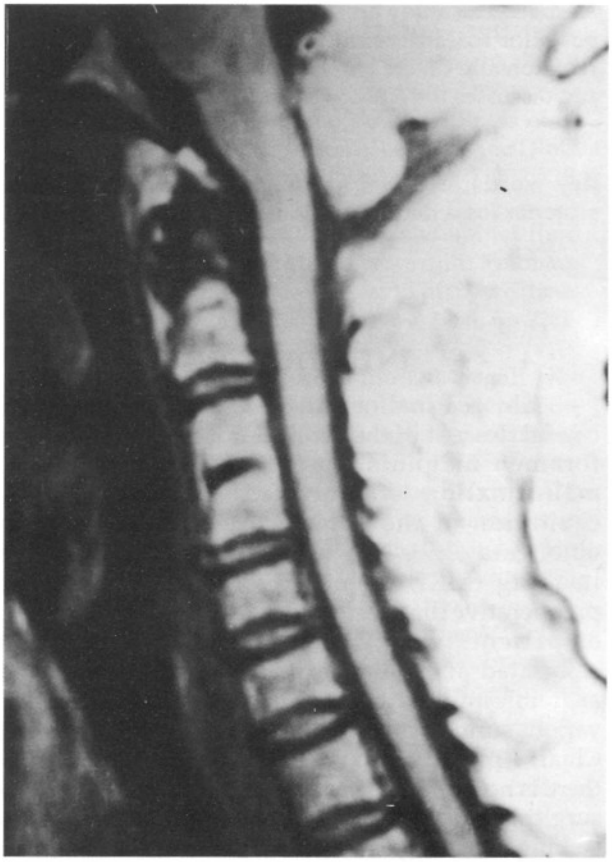

Figure 1b: Postoperative image of the same patient showing resolution of the syrinx following posterior fossa decompression.

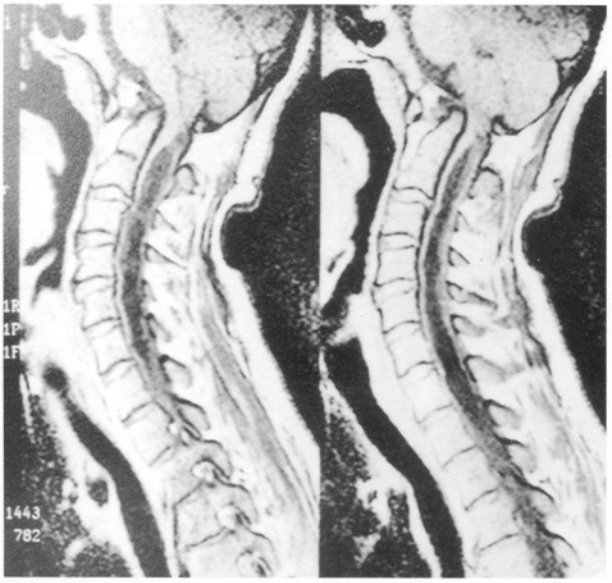

Figure 2a: Large syringomyelia cavity extended to bulbus is observed in preoperative MRI of a patient with a Chıarı type I malformation. Compression of the subarachnoid space anterior to the brainstem is also observed.

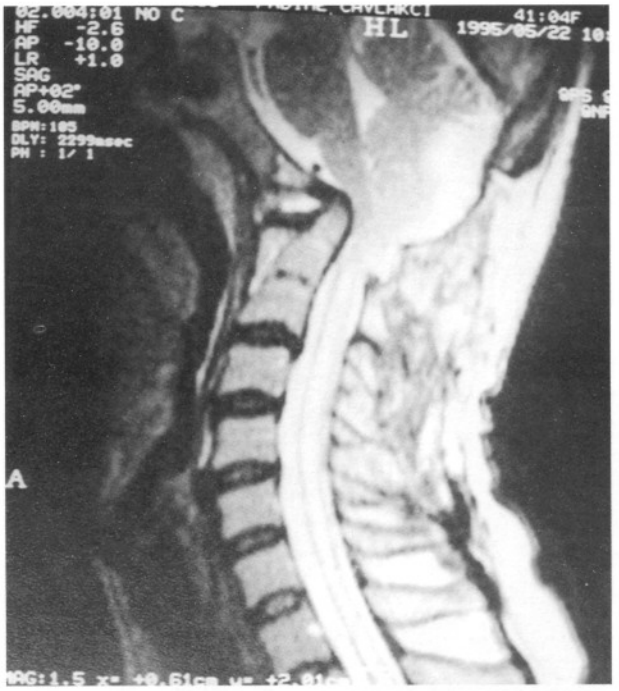

Figure 2b: Enlargement of the subarachnoid space anterior to the brainstem following posterior fossa decompression is observed postsurgery MRI of the the same patient.

OPERATIONS

Posterior fossa decompression for freeing CSF flow by removing extradural adhesions or bands without entering the subarachnoid space, and duraplasty were performed in 17 of the 20 patients. The procedure involved total laminectomy at C1 and C2, partıal craniektomi of the posterior part of the foramen magnum, removing of the extrdural bands encountered and opening the dura in vertical linear fashion and removing any subdural adhesions and bands without opening arachnoıd layer. Fascia lata was the only material used for duraplasty. In the two patients that had hydrocephalus, a vetriculoperitoneal shunt procedure was done. One other patient underwent a two-step operation in which the odontoid process was resected transorally following posterior fossa decompression (Table V).

Table II. Preoperative symptoms of 20 Chiari Type I cases

Symptoms	No. of patients (n=20)	%
Pain	18	90
Headache	11	
Neck pain	4	
Arm pain	2	
Back pain	1	
Weakness	13	65
Upper extrem	8	
Lower extremity	1	
Hemiparesia	2	
Quadriparesia	2	
Sensory loss	14	70
Ataxia	7	35
Vertigo	3	15
Dysphagia	3	15
Diplopia	2	10
Tinnitus	2	10
Comiting	2	10

Table III. Preoperative deficits.

Deficits	Bilateral upper	Bilateral lower	Right/Left side
Motor			
Wakness*	62	2	2 / 4
Atrophy	1	-	- -
Hyperreflexia	1	7	2 / 2
Hyporeflexia	8	1	- / 2
Sensory**			
Spinothalamic	11	2	- / -
Dorsal column	3	2	- / -

* Central cord-like syndrome was identified in 30% of patients.

** Dissociative sensory loss was identified in 25% of patients.

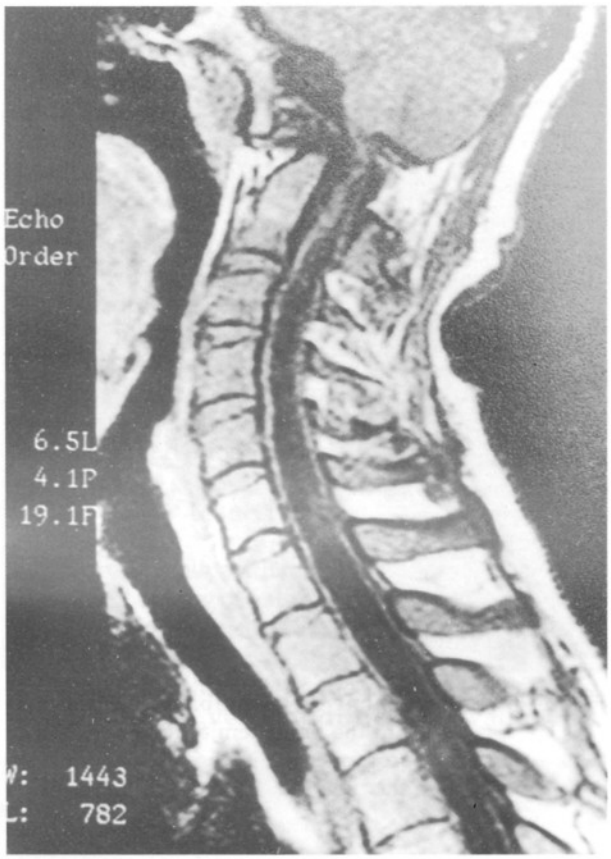

Figure 3a: Preoperative image of a Chiari I malformation and large defects in a patient with syringomyelia and syringobulbi.

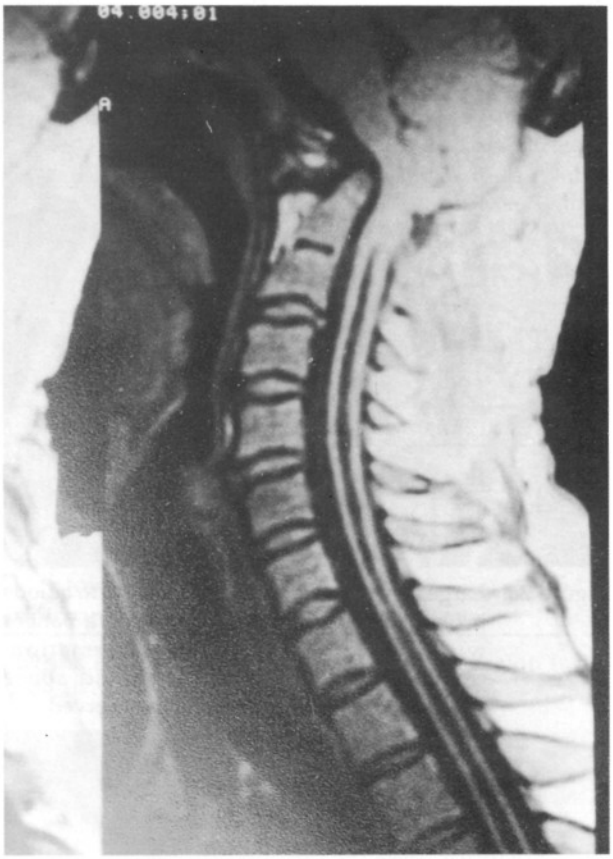

Figure 3b: Decreased syrinx diameter after posterior fossa decompression in the same patient.

RESULTS

The surgical outcomes for our patients are summarized in (Table VI). Seventy percent of the patients improved, and 20% remained unchanged from their preoperative status. The condition of two patients deteriorated after surgery. In particular, posterior fossa decompression led to significant reversal of gait disturbances, but there was no or little clinical change with regard to sensory loss. Pain was the complaint that resolved most rapidly, and the second fastest improvement was noted in weakness.

Repeat craniocervical MRI results were done 3 and 24 months after surgery are summarized in (Table VII). After surgery, syringomyelia resolved completely in two patients (Figures 1b and 4b) and regressed in 13 patients (Figure 3b). Enlargement of the subaracnoidal space anterior to the brainstem was observed in 11 cases (Figure 2b). The two patients with hydrocephalus improved such that there was no evidence of this condition at their 1-month postsurgery MRI examination. Patients with large syringomyelia and syringobulbi defects showed less improvement than those with small defects, which partially resolved.

Table IV: Preoperative signs.

Signs	No. of patients (n=20)	%
Ataxia	6	30
Cranial nerve palsy	4	20
Nystagmus	4	20
Gag reflex abnormality	4	20
Scoliosis	4	20
Papillary edema	2	10

Table V: Operative procedures.

Procedure	No. of patients (n=20)	%
PFD*+C**1,2 laminectomy+duraplasty	17	85
Ventriculoperitoneal shunt placement	2	10
PFD+C**1,2 laminectomy+duraplasty+ transoral odontoidectomy	1	5

*PFD: posterior fossa decompression
**C: cervical

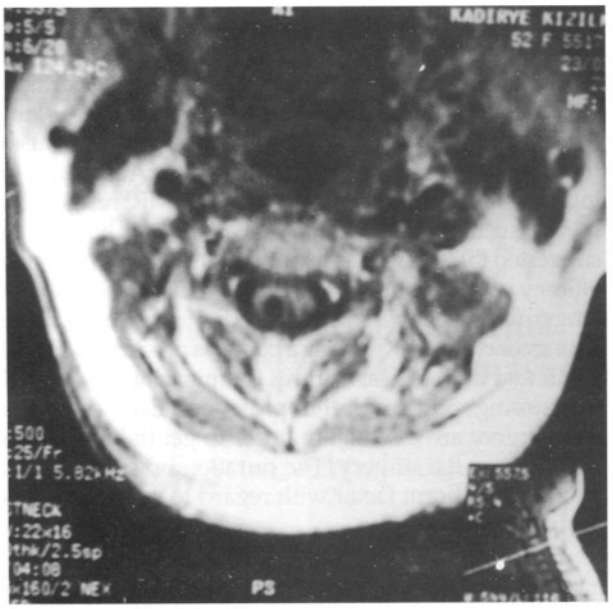

Figure 4a: A syrinx is seen in the axial plane.

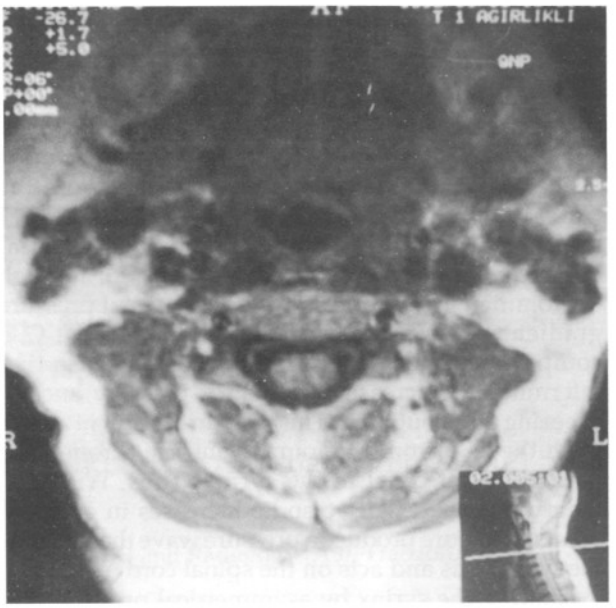

Figure 4b: Resolution of the same syrinx after surgery.

DISCUSSION

Controversy about the origin, persistence, and progression of syringomyelia has prompted the development of many surgical procedures (3). Gardner and colleagues postulated that the cause of syringomyelia associated with Chiari I malformation is delayed and incomplete embryonic opening of the outlet of the fourth ventricle, which would lead to retained communication between the fourth ventricle and the central canal (3). According to Gardner's hydrodynamic theory, the "water hammer effect" of CSF causes the development and progression of cord cavitations (3). Based on this hypothesis, Gardner advocated relief of the obstruction of the normal CSF passage route into the central canal by plugging the obex with muscle (3). Some authors have supported this hypothesis, but others have found fault with the theory. The arguments against Gardner's scheme are as follows:

1. In most patients, there is no communication between the fourth ventricle and the syrinx.

2. Most patients have patent fourth ventricle foramina.

3. Only occasionally is obstruction of normal CSF

Table VI: Surgical outcomes.

Procedure	improved	no progression	deterioration
PFD	11	4	2
PFD+odontoidectomy	1	-	-
Ventriculoperitoneal shunt placement	2	-	-
Total	14	4	2

Table VII: Control MRI findings.

Procedure	results				
	syrinx diameter			enlargement of subarachnoid space anterior to brainstem	hydrocephalus regression
	resolved	regression	same		
PFD	1	12	4	10	-
PFD+odontoidectomy	-	-	1	1	-
Ventriculoperitoneal Shunt	-	1	1	-	2
Total	1	13	6	11	2

flow from the fourth ventricle sufficient to produce hydrocephalus (4).

Williams proposed the "cranial-spinal dissociation theory," which is based on obstruction of CSF flow from the cranial to the spinal subarachnoid space at the foramen magnum, resulting from the Valsalva maneuver in patients with Chiari I malformation and syringomyelia (3,6). Partial obstruction of the caudal movement of the CSF through the foramen magnum results in a significant pressure gradient between the intracranial and spinal CSF compartments during routine daily activities that intermittently increase intrathoracic pressure, such as sneezing and coughing. This pressure gradient acts to push the CSF from the fourth ventricle through the patent central canal to the syrinx. Thus, Williams' theory proposes that episodic increases in epidural venous pressure produce a pressure wave that ascends the spinal axis and acts on the spinal cord externally, expanding the syrinx by asymmetrical pressure and propelling the syringomyelia fluid up and down the central canal (the "slosh effect"). Immediately following the elevation of intrathoracic pressure, the prolonged elevation of intracranial pressure over spinal intrathechal pressure forces fluid from the fourth ventricle down the central canal into the syrinx, producing "communicating syringomyelia." However, the following summarizes the opposition to William's theory:

1. In most patients, there is no communication between the ventricle and the syrinx.

2. In some patients with Chiari I malformation and syringomyelia, there is no blockage at the foramen magnum to caudal transmission of CSF pressure associated with the Valsalva maneuver.

3. Resolution of syringomyelia occurs after treatment limited to shunting the CSF from the lumbar subarachnoid space to the abdomen, even though this does not eliminate any blockage of CSF flow at the level of the foramen magnum.

Oldfield and colleagues have suggested a new theory to explain the formation and progression of syringomyelia associated with Chiari I malformation and occlusion of the subarachnoid space at the foramen magnum. They demonstrated the efficacy of posterior fossa decompression without entering the subarachnoid space in resolving syrinx formation in seven patients (5), and supported their thesis with dynamic MRI and intraoperative ultrasonography. According to this theory, the brain expands as it fills with blood during systole, imparting a systolic pressure wave to the intracranial CSF that is accommodated in normal subjects by sudden shift of CSF from the basal cisterns to the upper portion of the spinal canal. When this rapid shift through the foramen magnum is obstructed, the cerebellar tonsils, which plug the subarachnoid spaces posteriorly, move downward with each systolic pulse, acting as a piston on the partially isolated spinal CSF, which then acts on the surface of the spinal cord. They hold that this pulsating pressure causes the progression of syringomyelia (3,5). According to intraoperative ultrasound and dynamic MR[1] results, this theory seems more acceptable others that have been suggested (5). The results in our patients also support the efficacy of this method, but studies of larger series with longer follow-up periods are needed in order to make strong conclusions. Knowing a patient's presurgery clincal status and radiological features is important with regard to assessing post surgical improvment. Early diagnosis and surgery are essential in order to get the maximum benefit from the surgery. The duration of symptoms is another significant factor with regard to surgical results and expectations.

Correspondence: Zeki Şekerci, M.D.,
39. Sokak Doruk apt. 20/7
Birlik Mahallesi, Çankaya
06610 Ankara, Türkiye

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