

Original Research Article

Comparative study of duraplasty and non duraplasty in Chiari 1 malformation with syringomyelia our institute experience

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ABSTRACT

Background: The prevalence of Chiari malformation, defined as tonsillar herniations of 3 to 5 mm or greater, is estimated to be in the range of one per 1000 to one per 5000 individuals. The objective was to study the clinical presentation and outcome of ACM syrinx after foramen magnum decompression, C1 posterior arch removal duraplasty and without duraplasty.

Methods: The study included 75 cases admitted with ACM with syrinx in neurosurgery ward in King George hospital, Visakhapatnam, Andhra Pradesh, India, during a period of five years.

Results: Clinical improvement correlated strongly with enlargement of the subarachnoid cisterns and also correlated with reduction in size of the syrinx cavities. Postoperatively all the patients had decreased neck pain and two third of the patients had improvement in nystagmus, headache and dissociative anaesthesia in few cases.

Conclusions: In the available literature, the treatment options offered for ACM syrinx are foramen magnum decompression and C1 posterior arch removal, release of compression bands, which were followed in our institute also. We have compared pre-and post of MRI. Clinical improvement has been observed in 2 thirds of patient after a period of 2 year period follow up.

Keywords: Chiari type 1, Duraplasty, Suboccipital craniotomy, Syringomyelia

INTRODUCTION

Chiari malformations (CMs) constitute a group of different clinic pathological entities with varying etiology, pathophysiology, and clinical features. They represent varying degrees of hindbrain herniation through the foramen magnum. Professor Hans Chiari (1851-1916) developed a four-tier scheme for classifying these entities.^{1,2} His initial description was based on the findings of more than 40 autopsies, which he had performed while he was working as a pathologist in Prague^{1,2}

The prevalence of Chiari I malformation, defined as tonsillar herniations of 3 to 5 mm or greater, is estimated

to be in the range of one per 1000 to one per 5000 individuals. The incidence of symptomatic Chiari is less but unknown worldwide. The prevalence of non-post traumatic syringomyelia is 8.4 cases/1,00,000 population between 20-50 years. Syringomyelia is associated with Chiari type-1 malformation. Although many individuals with CM 1 are asymptomatic, the malformation can cause headaches, ocular disturbances, otoneurological disturbances, lower cranial nerve signs, cerebellar ataxia or spasticity.^{3,4} The condition is being diagnosed more frequently in adult patients due to the widespread use of MRI, particularly in the management of patients with headache. Surgical therapy for these entities has evolved over the years, but there are considerable variations in the treatment of adult patients with CM with and without

syringomyelia. Basic to all the procedures is enlargement of the foramen magnum with the intent of establishing relatively unimpeded flow of CSF from the cranial cavity to the spinal subarachnoid space.⁵⁻⁹

In the present study, we review our experience with the treatment of CM1 in patients who underwent decompression with or without duraplasty and the correlation of clinical and imaging outcomes after 2 year of follow up.

Aim of the study was to study the clinical patterns of the disease, to compare the outcome of surgery both with and without duroplasty, regression of the syrinx after surgery with post-operative MRI.

METHODS

The present study is both prospective and retrospective study to evaluate the clinical presentation of Chiari 1 malformation with syringomyelia of 75 patients who underwent posterior fossa decompression with and without duraplasty from 2011 to 2016.

Inclusion criteria

- Chiari malformation type I ≥ 5 mm tonsillar ectopic.
- Syrinx between 3 mm and 6 mm.
- MRI of the brain and cervical and thoracic spine are required prior to surgery.

Exclusion criteria

- Neuro-imaging demonstrating basilar invagination.
- Clival canal angle $< 120^\circ$ (signs of severe craniovertebral junction disease).
- Chiari Malformation I + syringomyelia secondary to other pathology (e.g. a tumor and trauma).
- Hydrocephalus patients.

Their histories were obtained from clinical charts, and the symptoms and their duration were determined. Preoperative magnetic resonance imaging scans were examined for the presence of syringomyelia.

Table 1: Clinical presentation.

Variable	Number (%)
Hypoesthesia	51 (68)
Head and neck pain	48 (64)
Hyperreflexia	27 (36)
Motor deficits	27 (36)
Temperature discrimination	21 (28)
Wasting of hand muscles	21 (28)
Extremity pain	15 (20)
Cerebellar sign	12 (16)
Cranial nerve sign	9 (16)
Drop attack	3 (4)

The diagnosis of Chiari malformation and syringomyelia was made exclusively by means of MRI in this series. The diameter of the syringomyelic cavity was measured in relation to the diameter of the spinal cord, as proposed by Fujii et al.¹⁰

The indications for surgery included but were not limited to progressive or disabling symptoms, such as headache or tussive headache; drop attacks; neck, arm, or back pain; swallowing difficulties; upper-extremity numbness or tingling. Clinical symptoms in present study (Table 1). Postoperative clinical improvement was assessed from clinical notes and generally reflected subjective reports of improvement in symptoms, return to work and decreases in pain. Radiological improvement was defined as any demonstrable decrease in maximum syrinx diameter, as seen on postoperative MRI scans.

The specific surgical procedure, i.e. non-duraplasty (without durotomy) or duraplasty, was chosen by each surgeon on the basis of training and personal preference. All patients underwent decompressive suboccipital craniectomy extending at least 2 cm above the foramen magnum, with bilateral removal of the C1 laminae. 36 of the patients then underwent removal of all dural scarring or bands on the outside of the dura, as described by Isu et al.¹¹ 39 patients underwent bone removal and dural grafting using the G graft. Follow-up was performed postoperatively at six, twelve and 24 months. Postoperative improvement or worsening of symptoms was determined and noted in the chart. For patients with postoperative MRI scans, the change in the size of the syrinx cavity was classified as improved (decreased maximum diameter) (Figure 2), unchanged or increased. This study was approved by the local ethics committee.

RESULTS

The patient population ranged in age from 15 to 55 years (mean 35 years) (Table 2) and included 33 men and 42 women (Table 3). The follow-up period ranged from six months to two years. Foramen magnum decompression with duroplasty was done in 39 patients and without duroplasty in 36 patients (Table 4). The selection of the cases for duroplasty was absent of the dural pulsation and greater than 8mm tonsillar decent.

Table 2: Age distribution.

Age	Number of patients	Percent
15-25	16	21.33
25-35	26	34.66
35-45	15	20.00
45-55	18	24.00

Hypoesthesia (68%) and sub occipital pain (64%) was the common symptoms in the study (Table 1). Improvement of clinical outcome same or improved both for duroplasty and non duroplasty was compared. Regression of syrinx in 33 patients after duraplasty is seen and in non

duroplasty (Figure 1) group only 12 cases have shown regression of the syrinx.

Table 3: Sex distribution.

Sex	Number of patients	Percent
Male	30	40
Female	45	60

Table 4: Type of procedure.

Type of procedure	Number
Non duroplasty	36
Duroplasty	39

Of the 36 patients who did not undergo duroplasty, 24 showed improvement postoperatively. Improvement of the clinical symptoms after duroplasty was seen in 33 patients and 6 patients did not show any clinical improvement. All of the patients after surgery postoperative MRI was done in the follow up period at 6 month 12 months and 24 months (Figure 2).

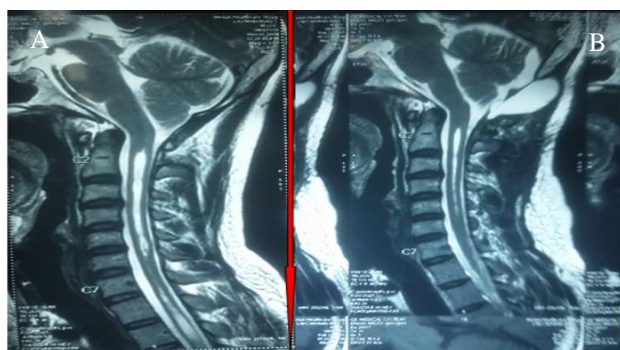


Figure 1: Regression of syrinx without duroplasty. A: Before surgery, B: After 24 months.

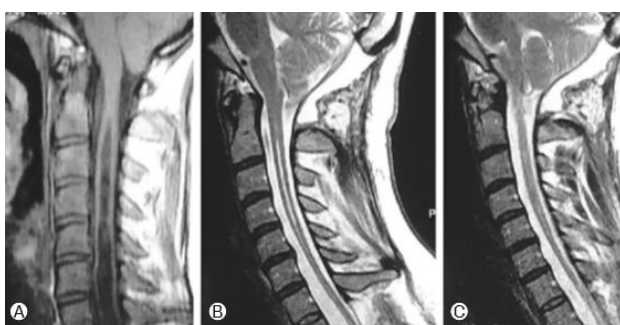


Figure 2: Regression of syrinx with duroplasty. A: 6 months, B: 12 months, C: 24 months.

When the dura was opened, the surgical complications included 12 cases of cerebrospinal fluid (CSF) leaks associated with aseptic meningitis in one patient; one case of subgaleal CSF or seroma collection that subsequently resolved with conservative treatment only; one case of superficial wound infections. When the dura was not opened, the only complication was a superficial

wound infection that resolved. Reoperation was done because of the persistent clinical symptoms and CSF leak.

DISCUSSION

Chiari type 1 malformations occur in the region where the brain and the spinal cord join. In this disorder, the portions of the brain called the cerebellum and/or brainstem lie lower than usual. Often, a portion of the cerebellum called the cerebellar tonsils protrudes out of the base of the skull into the spinal canal. This protrusion causes pressure in the brain, thus contributing towards the symptoms that such individuals experience. The cause of CM1 is not known. Some CM1 cases are believed to be present at birth.

The posterior fossa is smaller and shallower in patients with Chiari malformation, compared with that in normal individuals. Therefore, enlargement of the foramen magnum or creation of a cisterna magna is one goal of surgery. CM-1 includes a group of entities of congenital or acquired aetiology that have in common the descent of the cerebellar tonsils into the cervical spinal canal. It is associated with syringomyelia in 30%-70% of cases. Conversely, 90% of syringomyelia cases are associated with CM-1, in which the obstruction of CSF at the level of the foramen magnum alters CSF dynamics and results in syringomyelia.

Several mechanisms for the pathogenesis of syringomyelia have been proposed. According to the theory of Gardner and Angel, the obstruction of CSF flow at the craniocervical junction causes CSF to enter the cervical central canal.¹² However, no such communication exists in the majority of these patients and therefore this theory cannot explain the formation of the cavity.^{13,14} Oldfield et al proposed that CSF enters the spinal cord directly via the perivascular spaces, or so-called Virchow-Robin spaces, by means of arterial pulsations.¹³ This is consistent with studies that show movement of water-soluble contrast media from the subarachnoid space to the cavity. Aboulker considered that an increase in venous pressure in the epidural veins was the reason for the increase in fluid in the spinal cord, with subsequent hydromyelia. Recently, Stoodley et al showed that, under normal conditions in a sheep model, CSF flows rapidly from the subarachnoid space to the perivascular spaces, and that this flow is dependent on arterial pulsations.¹⁵ 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 hydromyelia in Chiari malformation is still controversial, there is general agreement on the importance of decompressing the craniocervical junction in treatments for Chiari malformation and hydromyelia. Suboccipital decompression, with or 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 duraplasty, syringosubarachnoid shunting or obex plugging. If the purpose of surgery for CM1 is enlargement of the cisterna magna, thereby allowing improved CSF flow, adequate bone removal must be obtained in all patients.¹⁶ Current study includes a comparison of two surgical techniques, FMD alone (non-duraplasty) and FMD with duraplasty (Table 5).

Table 5: Comparison of nonduroplasty and duroplasty.

Variable	Non duraplasty	Duraoplasty
Syrinx		
Same	24 (66.7)	6 (15.4)
Regressed	12 (33.03)	33 (84.6)
Clinical outcome		
Same	24 (66.4)	6 (15.4)
Improved	12 (33.3)	33 (84.6)
Complication	3 (8.3)	12 (30.8)
Reoperation	5 (41.6)	3 (7.6)

The average age of onset of symptoms compared with other studies (Table 6). The rate of clinical improvement

was significantly higher with duraplasty (84.6%) than without (33.3%, p<0.05). The rate of postoperative syrinx regression was significantly higher with duraplasty (84.6%) than without (33.3%, p<0.05).

Table 6: Age of onset compared to other studies.

Study	Range	Average
Present study	15-45	30
Banerji and Millar ²¹		
With Syringomyelia		40.4
Without Syringomyelia		29.2
Saez et al ²²	13-68	38
Levy et al ²³	12-73	41
Dyste et al ²⁴	1-57	29
Pillary et al ²⁵	18-57	38.5
Verasriet et al ²⁶	-	34
Bindal et al ²⁷	18-66	45

The rate of patients who remained clinically unchanged was 15.4% with duraplasty and 66.7% without duroplasty, revealing that clinical improvement relies on syrinx regression.

Table 7: Comparison with other studies.

Authors, year	Patients	Pathology	Surgical approach	Outcome	Morbidity mortality complications
Krieger et al ¹⁷	31	CM-1±HSM*	Occipital craniectomy, CL + duraplasty	Syrinx: 88% improvement, 3 patients also required a shunt	26% headaches 16% nausea No mortality
Alden et al ¹⁸	21	CM-1±HSM*	Suboccipital craniectomy + CL ± (i) durotomy + duraplasty:4, (ii) cerebellar tonsillectomy + adhesiolysis + duraplasty: (17)	67% symptom resolution, 29% improvement, 4% no improvement	N/A
Parker et al ¹⁷ , 2011	114	CM-1	Occipital craniectomy, CL, duraplasty ± tissue sealant	Re-operations	Graft/ sealant failures, Cumulative complications: 21%
Mottolese et al ¹⁹	82	CM-1	Occipital craniectomy, CL, duraplasty ± tissue sealant	Re-operations	Graft/ sealant failures, Cumulative complications: 21%
Sindou et al ²⁰	44	CM-1±HSM	Cranio-cervical decompression + far lateral foramen magnum opening + duraplasty + arachnoid preservation.	Improvement of syrinx in 60%, stabilization of syrinx in 40%	4.5% CSF leak, 2.3% laryngeal edema, 2.3% pneumonia, 11.4% wound infection, no mortality
Present study	75	CM-1+HSM	Suboccipital craniectomy Duroplasty (39) Nonduroplasty (36)	Syrinx 84% imorovement, 12% in non duroplasty	8.3% csf leak 5% seroma collection 3% infection

* HSM: hydrosyringomyelia.

If the size of the syrinx regressed than the clinical improvement was also noted in the patients whose syrinx size is regressed. These findings show that FMD and duraplasty is superior to FMD without duraplasty in CM-1 cases associated with syringomyelia. The syrinx size remained unchanged in 15.4% of patients who underwent duraplasty and 66.7% of patients who did not undergo duraplasty.

The patient with no change in the syrinx cavity size showed no change in the CSF space behind the cerebellum. It is also possible that this patient harboured or developed arachnoid scarring or subarachnoid adhesions. 30 patients had persistent syringomyelia, with 6 of these being in the duraplasty group, who subsequently improved clinically. However, none of the patients with persistent SM who were in the non-duraplasty group (n=24) improved clinically, and were evaluated as the same clinical condition as seen preoperatively, nevertheless none of them worsened. Postoperative infection was seen in 3 cases in the nonduraplasty group, and 12 cases were seen in the duraplasty group. The average age of onset of symptoms is compared with other studies (Table 6).

The clinical conditions of the patients having persistent syringomyelia remained unchanged (no improvement), however the clinical conditions of the patients whose syringomyelia regressed, achieved clinical improvement. These data also enable us to conclude that in CM-1 associated syringomyelia, FMD with duraplasty is superior to FMD alone, with respect to clinical improvement. Erdogan et al reported that the symptoms and signs had been resolved in 83% of their patients following FMD without duraplasty, however the rate of syrinx regression was only 28%. Present study also shown the similar results compared with the other study in improvement of the symptoms. Comparison of present study with the others studies the complication rate is slightly higher with duraplasty than without duraplasty.

Parker et al reported a 21.1% complication rate in their retrospective study including 114 patients in the duraplasty group. In their series, the most frequent complications were aseptic meningitis, symptomatic pseudomeningocele and CSF fistula. Klekamp reported a 21.8% complication rate in their retrospective study including 359 patients in the duraplasty group. Arruda et al reported 23% of pseudomeningocele and 6.6% of meningitis in their series including 60 patients in the duraplasty group.

In current study, the overall complication rate was 20%, which is in line with the current literature. The postoperative complication rate is 8.3% without duraplasty and 30.8% with duraplasty, which is consistent with the literature. There was significant improvement in duraplasty group in terms of size of the syrinx, sub occipital pain, dissociative sensory loss.

The morbidity and outcome of the study has been compared with other studies (Table 7). FMD with duraplasty is superior to non-duraplasty (FMD only) in CM-1 associated with SM, despite a slightly higher complication rate. It therefore seems that duraplasty provides a better chance of enlarging the size of the cisterna magna and improvement in the clinical symptoms as well as regression of the syringomyelia. It is difficult to draw conclusions from a series of limited size; however, there is a suggestion that patients with syringomyelia may have a higher likelihood of improvement after undergoing duraplasty. Nevertheless, some patients showed a decrease in syringomyelia, with an improvement in symptoms, through bone removal alone. This improvement was associated with an increase in the size of the cisterna magna, presumably allowing improved CSF flow and thereby leading to resolution of the syringomyelia. It therefore seems that duraplasty provides a better chance of enlarging the size of the cisterna magna.

CONCLUSION

Performing duraplasty to treat Chiari I malformation may lead to a greater decrease in concurrent syringomyelia. However, a subset of patients whose syringomyelia will decrease through bone removal alone still exists. Further studies are needed in order to better characterize these patients and to determine which patients with Chiari I malformation are better served with bone decompression alone, and which patients will require duraplasty to resolve their syringomyelia.

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Conflict of interest: None declared

Ethical approval: The study was approved by the Institutional Ethics Committee

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