Efficacy and Safety of Duraplasty in Chiari Malformation Type-I Patients Associated With Syringomyelia

Momen M Almamoun, MD., Walid K Abouzeid, MD.
Neurosurgery Department, Faculty of Medicine, Sohag University hospital, Sohag, Egypt.

Abstract

Background Data: Chiari malformation type-I (CM-I) is a challenging subject to wrap our hands around table. Chiari symptoms often range from, unexplained, and/or occipital Valsalva type headache, chronic fatigue syndrome, to lower cranial nerve abnormalities, or brain stem compression, till severe neurological insult which augmented by syringomyelia, or syringobulbia. Exact diagnostic and prognostic tools carry a great controversy which ranged from simple MRI study to MR imaging–based CSF velocity measurements, morphological, dynamic craniocervical junction assessments, subarachnoid pressure recordings, and compliance calculations were compared before and after surgical treatment.

Purpose: This study aimed to estimate the efficacy and safely of duraplasty in CM-I patients associated with syringomyelia.

Study Design: A retrospective descriptive clinical case study.

Patients and Methods: The study was conducted on 23 consecutive adults patients with CM-I associated with syringomyelia. They underwent surgical treatments at Sohag University Hospital from February 2012 to May 2015.

Results: The current study was applied to 23 patients; 10 males (43.5%) and 13 females (56.5%), aged 18-64 with a mean age of 41 years. The duration of symptoms before presentation varied from 1 month to 20 years. Clinical outcome was classified according to Glasgow outcome scale, 21 patients (91.3%) were graded V, 2 patients (8.7%) were grade IV, and no patient graded I. Radiologically MRI craniocervical junction suggested that decompression of the posterior fossa was achieved in all patients. Post-operative reports
showed that no deterioration occurred in any of our patients. Improvement occurred and increased gradually on post-operative period.

**Conclusion:** Craniocervical decompression with duraplasty is effective in treating patients with Chiari Malformation type-I with syringomyelia. Both clinical and radiologic improvement was documented on the follow up period. (2015ESJ099)

**Keywords:** Chiari malformation, Duraplasty, Craniocervical, Syringomyelia, syringobulbia

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### Introduction

Why the Chiari malformation type-I (CM-I) is a challenging subject to wrap our hands around table? First, the true incidence among population, vary from 0.35 to 1.85% with a mean 0.75% of the population, and this anomaly is associated with syringomyelia in 45%–70% of patients.\(^{15}\) Second, lack of agreement as regards the constitutional criteria, with many authors accepting 5 mm of tonsillar descent as the minimum criteria, but others suggesting that 0–2 mm, or less.\(^3\) Third, Chiari symptoms often range from, unexplained, and/or occipital valsalva type headache, chronic fatigue syndrome, to lower cranial nerve abnormalities, or brain stem compression, till severe neurological insult which augmented by syringomyelia, or syringobulbia.\(^{15}\) Fourth, exact diagnostic and prognostic tools carry a great controversy which ranged from simple MRI study to MR imaging–based CSF velocity measurements, morphological, dynamic craniocervical junction assessments, subarachnoid pressure recordings, and compliance calculations were compared before and after surgical treatment.\(^{15}\)

Magnetic resonance imaging has revolutionized early detection, and provided a greater understanding of the pathology, genesis, and manifestations of CM-I, and has also transformed outcome studies.\(^3\) Finally, what is the appropriate treatment for Chiari malformation when surgery is indicated? Single bone-only decompression? Should the dura be opened, or closed, with or without duraplasty?

Use a combination of different surgical approaches based on presentation such as arachnoid adhesolysis, and 4th ventricular exploration with or without obex plugging or stenting of the ventricle, resection of the tonsils, and various shunting procedures for the syrinx itself, and or craniocervical junction fixation.\(^{12}\)

There is little in the literature to guide ideal algorithm for management.

This study aimed to estimate the efficacy and safely of duraplasty in CM-I patients associated with syringomyelia in Sohag University Hospital.

### Patients and Methods

This retrospective descriptive study was conducted on 23 consecutive adults patients with CM-I associated with syringomyelia. They underwent surgical treatments at Sohag University Hospital from February 2012 to May 2015.

Inclusion criteria were applied to patients with type-I Chiari malformation with syringomyelia, but patients with type-II Chiari malformation and those with type-I without syringomyelia were excluded.

All patients were clinically assessed before treatment by history taking, general and neurological examination. The diagnosis was documented by standard MRI craniocervical junction. In addition, data were collected in standardized patients’ sheets.

Patients who fulfilled our inclusion criteria were categorized according to: (A) Age group, (B) Clinical manifestation, (C) The duration of symptoms before presentation, (D) Presence of syringobulbia, (E) The level of tonsillar descent, (F) Syrinx size preoperative measured at the widest point by MRI using computer program measurements, (G) Postoperative change of syrinx size, (H) Recovery period.
**Operative technique:**
The main surgical procedure consisted of a craniocervical decompression by removing the lower part of the posterior fossa, opening the foramen magnum and removing C1 and C2 posterior arch. Duraplasty was done to all patients using fascia lata graft with water tight closure preceded by adhesolysis of the cerebellar tonsils and plugging of the obex without a need for shunt tube insertion in the syrinx.

**Post-operative follow up**
Patients were on a follow up schedule in the outpatient clinic for one-month intervals after the operation for 6 months. Then it was every year. Follow up MRI craniocervical junction was done for all patients after three months and one year of surgery for at least 2 years up to 6 years.

**Outcome**
Glasgow outcome scale was used to assess the patients during the follow-up period, as follows: (V) Good recovery, resumption of normal life, (IV) Moderate disability, disable but independent, (III) Severe disability, dependent for daily support, (II) Vegetative state, unresponsive and speechless, (I) Death. In addition, syrinx diameter was measured in the follow-up period by MRI.

**Results**
The current study included 23 patients; 10 males (43.5%) and 13 females (56.5%). The age ranged from 18-64 with a mean 41 years (Table 1). The duration of symptoms before presentation varied from 1 months up to 20 years (Table 2). The patients suffered mainly from neck pain, sensory disturbance, and Intrinsic hand weakness. In addition, there was lower limb heaviness in 9 patients (39%), gait disturbance in 12 patients (52%), stridor in 2 patients (8.6%) and bulbar nerves palsy in 8 patients (34.7%), and globally severe combined cranial and spinal manifestations was reported in 21 cases (91.3%) (Table 3). Eighteen patients (78.3%) were associated with syringomyelia alone, and 5 (21.7%) had additional syringobulbia.

According to MRI findings, the level of tonsillar descent was from the level of foramen magnum to C1 in 4 patients (17.3%), at C1-C2 level in 16 cases (69.5%), and below C2 level in 3 patients (13.2%). The syrinx size was less than 5mm in four patients (17.3%), 5-10mm in eleven patients (47.8%), and more than 10mm in eight patients (34.7%). According to postoperative change of syrinx size, MRI was done after 3 months, illustrating a marked improvement in the size of the syrinx in those patients with syrinx less than 5mm to became normal. For those with (5-10mm) syrinx size become less than 5 mm, and for those with more than 10mm size, it became (5-10mm). All 5 cases with syringobulbia completely resolved and recovered (Table 4).

The duration of recovery of clinical symptoms ranged from one day to one year, with nearly half of the cases recovered in less than one month (Table 5). Clinical outcome was classified according to Glasgow outcome scale, 21 patients (91.3%) were graded V, but 2 patients only (8.7%) were grade IV, and no patient graded I.

Radiologically MRI craniocervical junction suggested that decompression of the posterior fossa was achieved in all patients (Figure 1, 2). Post operatively illustrated that no deterioration occurred in any of them. Improvement occurred and increased gradually on post-operative period.

**Surgical Complications:**
In three patients, subcutaneous CSF collection occurred after removing the drain it managed conservatively with diuretics and carbonic anhydrase inhibitors. One patient required to lay prone for 3 days and this was resolved. There was no need for any other surgical interference or lumber puncture and drain. In addition, no obvious or serious complication related to our procedure was encountered.
### Table 1. Age distribution in our patients (N=23)

<table>
<thead>
<tr>
<th>Age group</th>
<th>No</th>
<th>%</th>
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<tbody>
<tr>
<td>15 years-</td>
<td>4</td>
<td>17.4%</td>
</tr>
<tr>
<td>25 years-</td>
<td>12</td>
<td>52.1%</td>
</tr>
<tr>
<td>35 years-</td>
<td>3</td>
<td>13%</td>
</tr>
<tr>
<td>45 years-</td>
<td>2</td>
<td>8.7%</td>
</tr>
<tr>
<td>≥55 years-</td>
<td>2</td>
<td>8.7%</td>
</tr>
</tbody>
</table>

### Table 2. Duration of symptoms in our patients (N=23)

<table>
<thead>
<tr>
<th>Duration</th>
<th>No</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td>1 month-</td>
<td>9</td>
<td>39.1%</td>
</tr>
<tr>
<td>3 months-</td>
<td>6</td>
<td>26%</td>
</tr>
<tr>
<td>1 year-</td>
<td>2</td>
<td>8.7%</td>
</tr>
<tr>
<td>3 years-</td>
<td>4</td>
<td>17.4%</td>
</tr>
<tr>
<td>≥ 5 years</td>
<td>2</td>
<td>8.7%</td>
</tr>
</tbody>
</table>

### Table 3. Clinical presentation of our patients (N=23)

<table>
<thead>
<tr>
<th>Clinical presentation</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td>Headache and neck pain</td>
<td>100%</td>
</tr>
<tr>
<td>Sensory dysesthesia/numbness</td>
<td>100%</td>
</tr>
<tr>
<td>Intrinsic hand weakness</td>
<td>100%</td>
</tr>
<tr>
<td>Gait problem</td>
<td>52%</td>
</tr>
<tr>
<td>Lower extremity heaviness</td>
<td>39%</td>
</tr>
<tr>
<td>Cranial nerve dysfunction</td>
<td>34%</td>
</tr>
<tr>
<td>Stridor</td>
<td>8.6%</td>
</tr>
</tbody>
</table>

### Table 4. Change in syrinx size according to MRI after surgery in our patients (N=23)

<table>
<thead>
<tr>
<th>Syrinx size</th>
<th>Preoperative</th>
<th>Postoperative</th>
</tr>
</thead>
<tbody>
<tr>
<td>Less than 5 mm</td>
<td>4</td>
<td>Normal</td>
</tr>
<tr>
<td>5-10 mm</td>
<td>11</td>
<td>Less than 5 mm</td>
</tr>
<tr>
<td>More than 10 mm</td>
<td>8</td>
<td>5-10 mm</td>
</tr>
</tbody>
</table>

### Table 5. Recovery duration of clinical symptoms in our patients (N=23)

<table>
<thead>
<tr>
<th>Duration</th>
<th>No</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td>&lt; 1 month</td>
<td>11</td>
<td>47.8%</td>
</tr>
<tr>
<td>1 month-</td>
<td>8</td>
<td>34.7%</td>
</tr>
<tr>
<td>3 months</td>
<td>3</td>
<td>13%</td>
</tr>
<tr>
<td>6 months-</td>
<td>1</td>
<td>4.3%</td>
</tr>
<tr>
<td>12 months-</td>
<td>0</td>
<td>0%</td>
</tr>
</tbody>
</table>
Figure 1. Female patient 29 years old present with progressive weakness of the small muscles of the hands with paresthesia both upper limbs and spastic gait with frequent falls, on examination hand grip power was G2, with wasting of the hand muscles, dissociated sensory loss with hypertonia and hyper reflexia both lower limbs. Surgery was done clinical improvement occurs in the upper and lower limbs, power become G4a and improved gait and sensation. Preoperative (A) T2, (B) T1 MRI Showing tonsillar herniation below the foramen magnum and syrinx extending down to the upper dorsal spine. (C) postoperative T2 MRI showing complete foramen magnum decompression and resolving syrinx. Intraoperative images (D) showing intradural exposure and (E) showing duraplasty and onlay fat graft.
Figure 2. Male patient 24 years old presented with stridor, difficulty in swallowing spastic gait, on examination there were lost gag and palatal reflexes, with hypertonia and hyperreflexia both lower limbs and right hemihyposthesia. Surgery was done to him and on follow up improved bulbar symptoms and gait. Preoperative MRI (A) sagittal T2 showing showing tonsillar herniation between C1 and C2 segments with syrinx extending down to the upper dorsal spine, (B) axial T1 showing markedly distended and thinned out spinal cord. Six months postoperative MRI T2 (C) sagittal and (D) axial images showing adequate foramen magnum decompression and restoration of the CSF flow around the upper cervical spinal cord.
Discussion

This retrospective study describes the clinical and radiological outcomes of 23 consecutive patients with syringomyelia associated with CM-I who underwent surgery at Sohag University Hospital from February 2012 to May 2015. Overall, results confirm that posterior fossa decompression gives high rates of clinical and radiological improvement and a low complication rate. These results do not differ from those reported in other studies.\(^6\),\(^9\),\(^{13}\) All patients clinically improved but with varying degrees during the follow up period. Symptoms duration and age were correlated with the postoperative clinical and radiological improvement\(^2\). This was approved in our series.

Almost 70% of the patients aged below 35 years and those who revealed short term duration not more than 1 year (56.2%) revealed postoperative clinical and radiological improvement in (82.6%), and (93.5%), respectively within 3 months postoperative. Milhorat, et al,\(^{13}\) reported that the mean age at onset of syringomyelia in CM-I was 25 years, with women representing (75%) of the patients. Alfeiri et al,\(^2\) found that the population’s mean age was 45.9±13.7 years (18–77 years), with females representing (58.8%) of the patients. In a study covering 177 patients, Batzdorf, et al,\(^4\) reported that male patients were 45 (25.4%) and female ones were 132 (74.5%). They aged 5-78, with a mean of 37.92 years. This study included 10 males patients and 13 female, aged 18-69 and a mean age of (43.5 years), denoting that syringomyelia with CM-I is more common in females and young than males and middle-aged adults.

Alfeiri et al,\(^2\) reported that because clinical signs and symptoms are complex and vary from one patient to another; they can be simplified into cranial (due to brainstem compression or hydrocephalus) and spinal (caused by the syrinx). In addition, that their patients mainly had signs or symptoms related to the syrinx, but only 22.9% of them experienced cranial symptoms. A combination of spinal and cranial symptoms was assessed in 16 individuals. Furthermore, clinical spinal disturbances included: sensory loss, dysesthetic pain, segmental dissociated anesthesia, limb weakness, spasticity, amyotrophy, and sphincter deficiency. Cranial symptoms of brainstem compression illustrated cranial nerve neuropathies, uncontrollable hypertension, and tinnitus.

Baisden\(^3\) reported that CMI patients were often young adults with a multiplicity of vague complaints, such as: headache, neck pain, generalized discomfort with nausea, vomiting, dizziness, fluctuating hearing loss, visual disturbances, paresthesias, weakness, fatigue, and gait difficulties. Physical examination might vary from one office visit to another. Although it sometimes reveals no focal abnormalities, it may demonstrate nystagmus, cerebellar signs, or frank myelopathy. Deng et al,\(^5\) reported that they believe that the dominant side of tonsillar herniation determined syrinx deviation, which in turn determined clinical presentations.

In our series, it is found that neck pain, intrinsic hand weakness, and sensory disturbance appeared in all patients and that 5 patients suffered from lower limb heaviness. In addition, gait disturbance was manifested in 8 patients and bulbar nerves palsy in 4 patients of them stridor was found in 3 patients. The physical signs were a motor weakness in patients, sensory affection in the form of dissociated sensory loss in patients, hemihyposthesia in patients, bulbar palsy with the loss of gag palatal reflexes in the patients.

Aghakhani et al,\(^1\) believed that many authors performed different procedures (Posterior fossa decompression and shunting), and that their results could not be easily distinguished. Most importantly, there were no clear or reproducible clinical scores to analyze patients’ outcome. Ellenbogen et al,\(^7\) claimed that successful treatment required reestablishing CSF pathways by a surgical procedure.
appropriate for the presumed origin and nature of the obstruction. Baisden reported that the bony extent of the cranial decompression, the need for duraplasty, the choice for the Dural substitute material and/or Dural sealant used, and the need for an expandable cranioplasty remained controversial. However, shunting of the obex and primary shunting for syringomyelia associated with CMI became disapproved. He concluded that the primary pathology in CMI was attributed to CSF flow obstruction, rather than the absolute location of tonsillar descent below the foramen magnum. Therefore, this surgery basically aimed at restoring normal CSF flow dynamics. Heiss et al. concluded that persistent blockage of the CSF pathways at the foramen magnum caused increased pulsation of the cerebellar tonsils, acting on a partially enclosed cervical subarachnoid space to create elevated cervical CSF pressure waves. It, in turn, affected the external surface of the spinal cord to force CSF into the spinal cord through the Virchow-Robin spaces and to propel the syrinx fluid caudally, leading to syrinx progression. A surgical procedure of reestablishing the CSF pathways at the foramen magnum reversed this pathophysiological mechanism and resolved syringomyelia. Heiss et al. also found that a spinal subarachnoid block increased spinal subarachnoid pulse pressure above the block. This produced a differential pressure across the obstructed segment of the SAS, which resulted in syrinx formation and progression.

Alfieri et al. reported that surgical decompression with durotomy, arachnoid opening, tonsillar shrinkage, and re-creation of the cisterna magna was a safe and effective procedure. Prognosis was excellent, with global clinical and radiological improvement in more than 90% and 80% of patients, respectively. Definite clinical predictors of poor clinical and radiological prognosis—namely, age at the time of surgery and symptom duration—were identified. Sindou and Gimbert reported that patients undergoing foramen magnum decompression with incision in the outer layer of dura or complete dural opening followed by duraplasty were significantly better (P <0.05) than those patients having foramen magnum decompression with dural and arachnoid opening but without duraplasty.

From a completely different point of view, Goel claimed that Given our experience, we conclude that CM, with or without basilar invagination, was associated with instability at the atlantoaxial joint, even if such instability was not clinically manifested or demonstrated on radiological imaging. He adopted stabilization of the atlantoaxial joint as the treatment. Foramen magnum bone or dural decompression was unnecessary. In addition, both syringomyelia and CM were secondary natural events related to long-standing atlantoaxial dislocation and there was no need for direct surgical manipulation.

Kennedy et al. reported, in their series of outcomes after posterior fossa decompression (PFD) without dural opening, that 40 out of 57 patients (70%) demonstrated radiographic improvement, 13 patients (23%) remained stable, and (4) patients (7%) developed increased syrinx size or a new one. Furthermore, 8 patients (20%) with a syrinx underwent reoperation for persistent or recurrent symptoms or progression of associated scoliosis. They recommend applying PFD without opening the dura for most symptomatic patients with CM-I. However, it had rapidly progressive neurological deficits, rapidly progressive scoliosis with the syrinx, and craniovertebral instability requiring fusion. Based on these results, dural opening surgery if preoperative MRI suggests that partial C-2 laminectomy was required to achieve adequate decompression of the tonsils.

In this study surgery was performed to all patients with craniocervical decompression removing the lower part of the posterior fossa, opening the foramen magnum, and removing the posterior arch of C1and C2. Duraplasty was also done to all patients using fascia lata graft with water tight closure preceded by adhesolysis.
of the cerebellar tonsils and inspection of the obex. During the follow up period, all patients improved and there were 21 patients with GO scored V and 2 scored IV and no patient deteriorated. Only 3 patients experienced post-operative CSF collection which was resolved with conservative measures and did not require any further procedures.

We observed that the higher the level of tonsillar descent was the higher clinical symptoms, the extent of syrinx and showing the syringobulbia were. The removal of C1 and C2 rapidly improved the symptoms of both syringomyelia and syringobulbia against atlantoaxial instability. This also enforced the theory of restoring of CSF pathway, and duraplasty essentially to create more space and arachnoid adhesolysis, allowing the tonsil to restore its original level.

**Conclusion**

Craniocervical decompression with duraplasty is effective in treating patients with Chiari Malformation type-I with syringomyelia. Both clinical and radiologic improvement was documented on the follow up period.

**Limitations**

The current study is not a comparative study to the syrinx’s shunting. In addition, a small number of patients and a longer period of follow up is needed to detect a late possibility of syringomyelia recurrence, with no dynamic plane for atlanto-axial segment.

**References**


Address reprint request to: Momen M Almamoun, MD.
Neurosurgery Department, Sohag University, Sohaj, Egypt.
Email: momen_67@yahoo.com
فعالية وسلامة ترقيع الأم الجافية في علاج تشوه شياري النوع الأول المرتبط بتكهف النخاع

استنتاج: عمليه إزالة الضغط عن المنطقة الدماغية العنقية مع تركيع الأم الجافية طريقة مؤثرة في علاج مرضى تشوه شياري النوع الأول المصاحب بتكهف النخاع وذلك دون الحاجة لوضع أنبوب تحويلي في التكهف النخاعي. تم توثيق التحسن خلال فترة المتابعة على مستوى الأكلينيكي والأشعة.