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Technical Note

# Surgical treatment of idiopathic syringomyelia: Silastic wedge syringosubarachnoid shunting technique

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## Abstract

**Background:** The underlying pathophysiology leading to syringomyelia is elusive with multiple flow-related theories constituting our current limited understanding of the disease process. Syringomyelia is associated with pathologies related to the disturbance of cerebral spinal fluid flow found in conditions such as Chiari I malformations, spinal malignancy, spinal cord tethering, trauma, or arachnoid adhesions. Our aim is to describe a unique surgical shunting technique used to treat refractory cases of idiopathic syringomyelia.

**Methods:** Five patients, aged 22-50, presented with progressive neurologic symptoms associated with an idiopathic syrinx. All underwent decompressive laminectomy surgery with syringosubarachnoid shunting using the silastic wedge technique.

**Results:** In five cases of idiopathic syringomyelia, clinical and radiographic follow up ranges from 3 to 36 months. Three patients have radiographic and clinical follow up greater than 24 months. All patients improved clinically and their symptoms have been stable.

**Conclusions:** Shunting procedures for the syringomyelia disease spectrum have been criticized due to the inconsistent long-term outcomes. This surgical technique used to treat symptomatic idiopathic syringomyelia has been devised based on our intraoperative experience, surgical outcomes, and evaluation of the literature. The purpose of the wedges is to preserve patency of the communication between the syrinx cavity and the expanded subarachnoid space by preventing healing of the myelotomy edges and by maintaining an artificial conduit between the syrinx cavity and the subarachnoid space. Although short-term results are promising, continued long-term follow up is needed to determine the ultimate success of the silastic wedge shunting procedure.

**Key Words:** Idiopathic syringomyelia, silastic wedge technique, syringosubarachnoid shunt

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## INTRODUCTION

Surgical intervention for syringomyelia is considered when conservative medical treatment fails and neurological symptoms progress.[5-9] The underlying pathophysiology leading to syringomyelia is elusive with multiple flow-related theories constituting our current limited understanding of the disease process.[4,5,12] Syringomyelia is associated with pathologies related to the disturbance of cerebral spinal fluid flow found in conditions such as Chiari I malformations, spinal malignancy, spinal cord tethering, trauma, or arachnoid adhesions.[5,6] In these conditions, surgical intervention involves directly removing the source causing the syrinx formation. Malignancy resection, decompressive, and untethering procedures with adhesion removal and duraplasty have been successful in removing offending pathologies creating the syringomyelia condition.[1,3,11] In cases where these methods have failed, the use of syrinx shunting, whether syringoperitoneal, syringopleural, or syringosubarachnoid, has been advocated as a procedure of last resort. Many shunting procedures have been described and the results have been erratic. Complications include fibrosis with subsequent obstruction, shunt migration, and shunt infections, which minimize the success of this surgical treatment.[10,11] Data from long-term studies on shunting procedures for syringomyelia suggest 12-53% of patients improve, 10-56% unchanged, and 12-32% regress.[2,11]

Idiopathic syringomyelia is an entity not associated with any of the previously mentioned conditions.[2] With no overt etiology, surgical decision making and treatment can be challenging. This report describes a unique surgical shunting technique used to treat refractory cases of idiopathic syringomyelia.

## MATERIALS AND METHODS

Institutional review board approval was obtained and all patients with refractory idiopathic syringomyelia who underwent a syringosubarachnoid shunt using the silastic wedge technique at a single institution were analyzed.

### Patients

Five patients, aged 22-50, presented with progressive neurologic symptoms associated with an idiopathic syrinx [Table 1]. One patient (Case 1) had a history of multiple sclerosis with new onset left sided chest pain in a band-like distribution and exacerbation of her gait abnormalities and urinary retention. There was no proceeding traumatic event prior to the onset of symptoms. Two patients (Cases 2 and 3) presented with intractable neck pain and headaches following motor vehicle accidents. They were conservatively treated without relief of their symptoms. One patient (Cases 4) presented with intractable thoracic radiculopathy and no evidence of trauma preceding the symptoms.

### Technique

A decompressive laminectomy extending rostral and caudal to the syrinx cavity is performed. The dura is opened and reflected laterally and secured with 4-0 sutures. Arachnoid inspection is undertaken with assistance of an operating microscope to identify any possible constricting/flow diverting lesions [Figure 1]. Intraoperative ultrasound is employed to identify the syrinx cavity and a midline myelotomy is performed [Figure 2]. Once the syrinx cavity has been entered, two silastic wedges are contoured from a 0.13 mm silastic sheet to fit the myelotomy dimensions [Figure 3]. The two wedges placed side-by-side and sutured to the pia using 9-0 prolene [Figure 4]. If adhesions are encountered, a duraplasty with Gortex is performed. Otherwise closure is performed in a typical layered fashion.

## RESULTS

In the five cases of idiopathic syringomyelia, clinical and magnetic resonance imaging (MRI) follow-up

**Table 1: Patient clinical and operative data**

| Case no | Age (years.), sex | Symptoms and signs | Imaging | Treatment |
|---|---|---|---|---|
| 1 | 35, female | New onset burning thorax pain with bilateral LE weakness | T4-T6 syrinx 3 mm diameter, 52 mm length | T5-T7 laminectomy with syringosubarachnoid shunt |
| 2 | 38, female | Intractable neck pain and headaches | C5-C6 syrinx 4 mm diameter, 29 mm length | C4-C7 posterior cervical decompression with syringosubarachnoid shunt |
| 3 | 50, female | Progressively worsening right arm pain, burning pain in the shoulders and neck, intractable headaches | C6-T1 syrinx 4.5 mm diameter, 40 mm in length | C5-T1 posterior cervical decompression with syringosubarachnoid shunt |
| 4 | 30, female | Progressively worsening thoracic pain | T4-T5 syrinx 3.5 mm diameter, 40 mm in length | T4-T5 laminectomy with syringosubarachnoid shunt |
| 5 | 37, male | Intractable thoracic radicular pain | N/A | T4-T6 laminectomy with syringosubarachnoid shunt |
| 6 | 22, female | Intractable thoracic pain with LE paresthesias and weakness | T9 syrinx, 10 mm diameter, 16 mm in length | T9-T10 laminectomy with syringosubarachnoid shunt |

ranged from 3 to 36 months. Three patients have MRI and clinical follow up greater than 24 months. All patients improved clinically and symptoms have been stable. Cases 4 and 5 have limited follow up radiographic data. Case 4 has improved clinically but refuses follow up imaging. All shunted syrinx cavities have shown a decrease in size [Table 1] and have been stable from 3 to 36 months [Figures 5-8] in terms of MRI images.

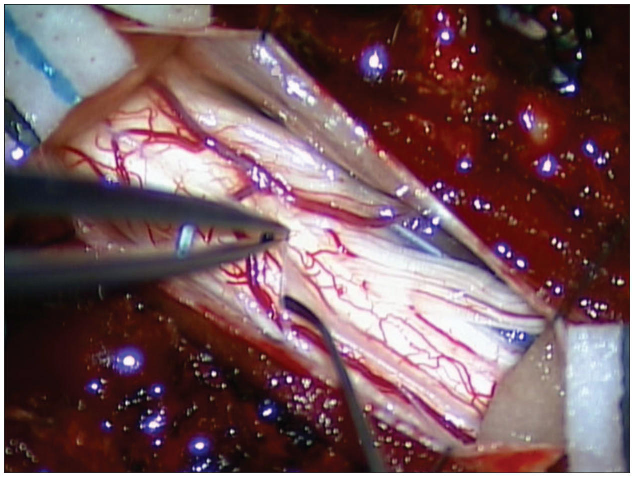

**Figure 1: Opening the arachnoid membrane**

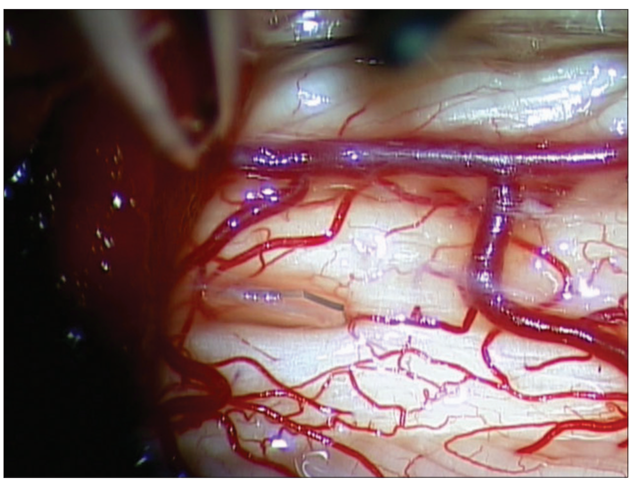

**Figure 3: Initial placement of contoured silastic wedge**

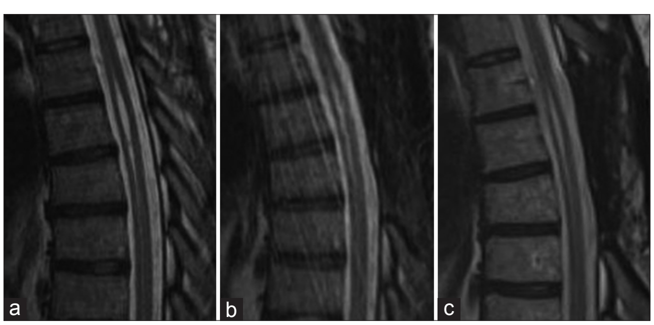


**Figure 5: Case 1. (a) preoperative syrinx state. (b) 12 months postoperative. (c) 24 months postoperative**

## DISCUSSION

Shunting procedures for the syringomyelia disease spectrum have been criticized due to the inconsistent long-term outcomes. This is largely the result of small volume flow at a very low-pressure profile leading to occlusion or malfunction of the shunts. This surgical technique used to treat symptomatic idiopathic syringomyelia has been devised based on our intraoperative experience, surgical outcomes, and evaluation of the literature. The purpose of the wedges is to preserve patency of the communication between the syrinx cavity and the expanded subarachnoid space

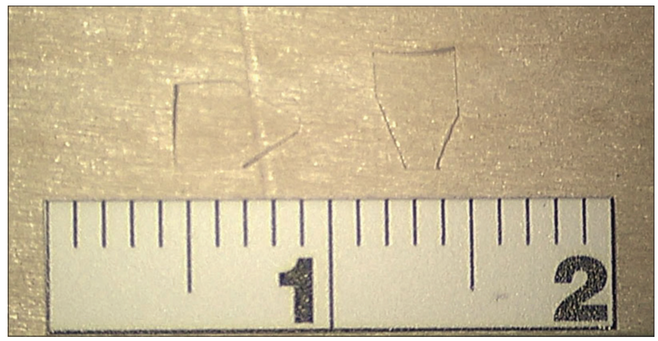


**Figure 2: Contoured silastic wedges**

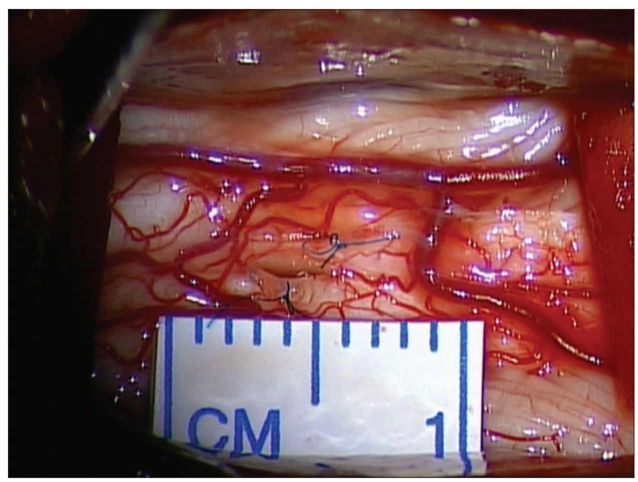


**Figure 4: Final position of silastic wedges**

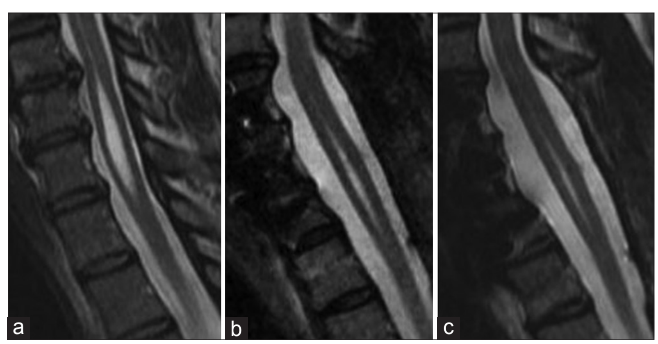


**Figure 6: Case 2. (a) preoperative syrinx state. (b) 12 months post perative. (c) 24 months postoperative**

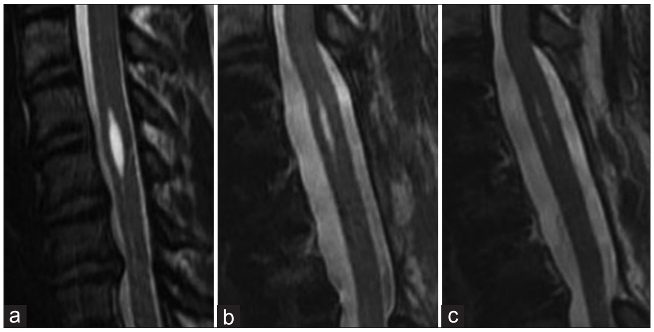

**Figure 7: Case 3. (a) preoperative syrinx state. (b) 12 months postoperative. (c) 36 months postoperative**

by preventing healing of the myelotomy edges and by maintaining an artificial conduit between the syrinx cavity and the subarachnoid space. The goal is to enable small volume flow more consistently. The silastic wedges allow for the passage of cerebral spinal fluid through capillary action under very small pressures without collapse. This technique allows for insertion of two wedges. Capillary action will allow for the fluid to travel along the space between the two wedges. The benefit to this method is that even after the syrinx cavity is drained and pressures within the cavity diminish, the patency of the conduit and the mechanism of the capillary action will still be maintained. For traditional shunt procedures with diversionary tubing, a higher pressure must be maintained to achieve continued flow. As fluid is drained, the pressure decreases and the cavity collapses around the perforations in the shunt tubing. Flow through the tube becomes stagnant and the susceptibility to obstruction, a well-recognized complication with shunting procedures, increases. With the silastic wedges, a potential space is created between the two surfaces allowing a constant channel for fluid to pass under changes in pressure and cavity size. The low profile of the silastic wedges within the subarachnoid space may also help reduce subarachnoid adhesions that have previously been reported in cases using tubing to shunt the syrinx cavity. When securing the traditional tubing into place, the round opening of the tubing makes it difficult to be sutured to the pia. The flexible silastic wedge allows for a simple stitch that can be placed under direct microscopic visualization to avoid damage to any surface vessels. The triangular shape of the wedge allows for direct visualization and safe placement into the syrinx cavity without blind advancement.

## CONCLUSION

The inconsistent results that are reported with shunt tubing leads us to question whether drainage will continue once the initial drainage is achieved. Because a consistently high pressure would be required to maintain flow, there is most likely cerebral spinal fluid egress around the shunt tubing into the subarachnoid space rather than drainage through the tubing. We have found that the silastic wedges provide a similar conduit requiring less pressure to drain fluid from the cavity. The shape and pliability of the wedges lend to a one-step, safe insertion that does not require blind advancement or manipulation into the syrinx cavity. The wedge will also be able to maintain small volume flow more consistently even under a low-pressure profile. Although short-term results are promising, continued long-term follow up is needed to determine the ultimate success of the silastic wedge shunting procedure.

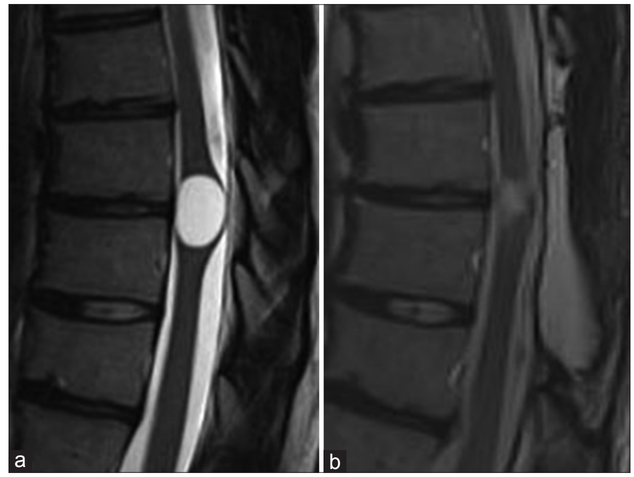

**Figure 8: Case 4. (a) preoperative syrinx state. (b) 3 months postoperative**

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## Commentary

Johann Conrad Brunner, an anatomist and a physician, has been recognized as the first scientist reporting syringomyelia, in the year 1688, in western literature.[2] A lesser known fact is that the first description of the disorder can actually be found 15 centuries before that in an ancient Chinese manual, The Yellow Emperor's Inner Canon.

In modern neurosurgery, syringomyelia shunts are considered a procedure of last resort, to be considered whenever the cause of the syrinx cannot be identified (idiopathic cases) or the cause of the syrinx has been refractory to former surgical treatment.

Syringomyelia cavities are identified in up to 50% of patients affected by Chiari I malformation. Tumors, vascular malformations, inflammatory disorders, infections, traumatic sequelae, vascular ictal events, and congenital anomalies are the main other causes of syringomyelia.

The advancement of diagnostic techniques and the increased base of knowledge pertaining to syringomyelia have contributed to the progressive decline in the quota of true idiopathic cases.

Regional cerebrospinal fluid (CSF) blockage and CSF pressure differentials have been recognized as key factors in the pathophysiology of syringomyelia.[3] The understanding of the CSF dynamics in each individual case of idiopathic syringomyelia is the key in identifying the proper surgical treatment.

In general, the mere presence of a syrinx is not an automatic indication for surgery, unless there is evidence of progressive enlargement on serial magnetic resonance imaging (MRI), or there is a large size on the very first MRI, with the girth of the lesion being a more important factor than its length. Lesser syringomyelia cavities can become surgically relevant when they cause severe symptoms or focal neurological deficits. In the latter scenario, a syrinx bleb would burst outside of the limits of the central canal, cutting through the spinal cord parenchyma, thus producing the focal deficit.

Whenever the surrounding subarachnoid spaces have stagnating CSF flow and elevated CSF pressure, any attempt to shunt CSF from the syrinx to such space is going to encounter limited or no success. In these cases, a syringopleural or syringoperitoneal shunt would be preferable, given the intermittent negative pressures in the end compartments. Syrinx cavities located in the lower segments of the spinal cord are often first treated with a terminal ventriculostomy, before entertaining the spinal cord manipulations linked to syrinx shunting.[6]

A 50% failure rate at the 5-year postoperative interval has been described as a late complication of syringomyelia shunting.[1]

In order to minimize the occurrence of iatrogenic deficits, the surgical access to the syrinx generally occurs through the midline or (less frequently) at the dorsal root entry zone. Large syringomyelia cavities often deform the standard anatomical landmarks and the perception of where the anatomical midline should be. Other times, the presence of sizeable vessels on the posterior cord surface can force the surgeon to veer away from an ideal entry point to avoid a vascular insult. In specific situations, the surgeon can intentionally decide on a parasagittal entry point, whenever a syrinx has expanded posteriorly and off the midline, already causing a deep sensory deficit in the posterior columns.

The access to the syrinx cavity can be facilitated by the use of intraoperative ultrasound. Neurophysiological intraoperative monitoring also provides important feedback during these delicate maneuvers.

Syrinx shunting can be performed to the peritoneal cavity, pleural cavity, or the subarachnoid space. A rare variant of syrinx shunting has the distal catheter ending in the cisterna magna.[4]

Syringo-subarachnoid shunting techniques are favored by some surgeons to avoid the risk of CSF leakage at the dural exit point of the distal catheter. This problem can be easily solved by reinforcing the exit point with a small piece of muscle held in place by cross stitching.

Syringo-subarachnoid shunting can be performed by inserting a catheter or by creating an ostomy through the cord tissue surrounding the syrinx, with the latter modality being complicated by a higher occlusion rate.

Myringotomy (aka tympanostomy) tubes have been used with success in syringostomy surgeries in the past to maintain the ostomy open.[5]

The above article describes an original variation of the same technique using wedges of silastic anchored to the pia. We are interested to hear from the authors about the patency rate of this approach at the 5-year term.

In conclusion, during the last decades, syringomyelia shunting has become progressively less used by neurosurgeons due to its complication rate and long-term patency problems. In the few instances in which its use is contemplated, individual anatomical and pathophysiological details should be taken into consideration to choose the most effective surgical strategy.

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