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Technical Notes

Foramen magnum decompression with cervical syringotomy for Chiari malformation type I with syringomyelia – A useful adjunct in selected cases

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ABSTRACT

Background: Persistent or worsening syringomyelia after foramen magnum decompression (FMD) for Chiari I malformation (CIM) can be challenging to manage. We present a previously unpublished surgical technique of FMD with concomitant cervical syringotomy in selected patients.

Methods: A retrospective analysis of prospectively collected data was carried out. Patients who underwent FMD and expansion duraplasty (FMDD) with concomitant syringotomy were collected.

Results: Three patients with CIM with high cervical syringomyelia who underwent FMDD with concurrent syringotomy were identified. All cases had an idiopathic CIM. Improvement in clinical symptoms was noticed in all patients. Early postoperative imaging (within 6 weeks–4 months) showed syrinx transverse diameter reduction in the range of 85–100%. There were no postoperative complications.

Conclusion: FMDD with concurrent high cervical syringotomy through a standard approach in selected cases of CIM with high cervical syringes achieves clinical improvement without additional complications.

Keywords: Chiari malformation, Foramen magnum decompression, Syringomyelia, Syringotomy

INTRODUCTION

The surgical practice to manage hindbrain herniation has evolved over the past several decades. Chiari I malformation (CIM), first described in 1891, involves caudal descent of the cerebellar tonsils through the foramen magnum. The association between CIM and syringomyelia is seen in 35–75% of the cases.[1] Syrinx can present with sensory disturbances, bulbar symptoms, and spinal deformity.

While most neurosurgeons agree that foramen magnum decompression (FMD) is the mainstay of surgical treatment, there is considerable diversity of opinion regarding the extent of surgery with a wide range of permutations of operative technique.[7] Whichever technique is utilized, the persistence or progression of syrinx after FMD is reported in various series in the approximate range of 30–40%.[1,7] Cases with an increase in syrinx size and worsening of the neurological symptoms after FMD have been described.[2,7,15] It has been reported that a large holocord syrinx

induces a component of spinal cord injury, making the persistence of syrinx after FMD an undesirable outcome of surgery for syringomyelia.[23]

FMD with expansion duraplasty (FMDD) on its own successfully addresses cerebrospinal fluid (CSF) flow dynamics at the craniocervical junction thereby affecting the syrinx filling mechanism concurrently. Although other techniques have also been utilized to try to improve the rapidity of decompressing the syrinx, such as concurrent FMD with syringo-subarachnoid shunt described by Sgouros and Williams.[17] However, it is accepted that this has significant limitations in that syrinx shunts have a short half-life, require a separate exposure, and are fraught with many complications.[18]

We, therefore, considered whether, in a highly selected group of patients with CIM and high, tense cervical and medullary syringomyelia, the incorporation of syringotomy during FMDD could achieve rapid decompression and drainage of syrinx without any added risks and complications.

MATERIALS AND METHODS

We present three patients with CIM with extensive syringomyelia with the upper part of the syrinx reaching up to the cervicomedullary junction, who underwent FMDD with concurrent syringotomy. All cases had an idiopathic CIM with no associated craniovertebral junction anomaly/instability or hydrocephalus. The decision for concurrent syringotomy for all patients was taken preoperatively based on the location of the syrinx and after review of imaging confirmed an extensive, tense syrinx accessible through our routine surgical exposure for FMD (suboccipital craniectomy and C1 posterior arch laminectomy). All patients included in this study were consented preoperatively regarding the syringotomy.

The predominant outcome measures were improvement in symptomatology and improvement in syrinx status on magnetic resonance imaging (MRI). The presence or absence of surgical complications was also noted. The radiological features of the syringes were assessed using comparative measurements. A collapse meant that the diameter of the syrinx had become zero. This could involve a segment of the syrinx (segmental collapse) or the whole syrinx (total collapse). In syringes or segments without collapse, we simply measured the percentage change in the maximum transverse diameter of the syrinx.

The Institutional Review Board/Ethics Committee approval and patient consent were not sought, as this was a retrospective study.

Surgical technique

First, we describe our standard technique for conducting FMD with duraplasty (FMDD). Intraoperative neurophysiological monitoring is used in all cases with transcranial motor evoked potentials (MEPs) and somatosensory evoked potentials (SSEPs). The midline skin incision was followed by the musculofascial flap elevated in a T-shaped fashion and subperiosteal dissection to expose the subocciput, foramen magnum, and posterior arch of C1. Suboccipital craniectomy at the foramen magnum (3 cm width × 2 cm height) is performed, followed by C1 posterior arch laminectomy. Extradural constrictive bands including the posterior atlanto-occipital membrane are excised followed by a Y-shape dural opening and the tonsils are visualized.

Depending on the extent of tonsillar herniation, under magnification, the cerebellar tonsils are reduced with bipolar coagulation and expose the Obex. The Obex and foramen of Magendie are explored to treat any obstruction to CSF outflow from the fourth ventricle [Figure 1a].

Intraoperative ultrasound is used to confirm the extent of the syrinx cavity on the upper cervical cord. Often, the midline median sulcus is visible together with an expanded cord. Neurophysiological monitoring and mapping can be used to identify the median sulcus with a phase reversal technique, although this has not been reliable in our short experience, owing most likely to the displacement of fibers due to the expansive syrinx cavity. We have found anatomical markers an excellent and reproducible indicator in particular pial arteries that can be seen dipping inward at the median sulcus on the dorsal spinal cord surface [Figure 1b].

A syringotomy is performed using a sharp ophthalmic/diamond blade and expanded in a craniocaudal fashion using a microdissector [Figure 1c]. Often, a gush of syrinx fluid under pressure is observed and microsuction tip is utilized to drain the cavity. Monitoring of MEPs and SSEPs has always confirmed stability during this. The cervical spinal cord always appears significantly lax following syrinx drainage. We do not leave any direct syrinx shunts nor do we place any pial sutures to close the syringotomy as it is an extremely small opening [Figure 1d]. We believe that syringotomy allows rapid decompression and drainage of the syrinx to decompress the cord as quickly as possible, with the filling mechanism being disabled by addressing the CSF outflow obstruction at the craniovertebral junction achieved through the FMD. A duraplasty is performed utilizing a bovine pericardial graft augmented with fibrin sealant on the lay. Layered closure is performed.

RESULTS

Case 1

This is an 11-year-old girl with CIM and an extensive holocord syrinx [Figures 2a and b]. She presented with sensory dysesthesias, headaches, and mild scoliotic deformity. On examination, she manifested poor balance and coordination

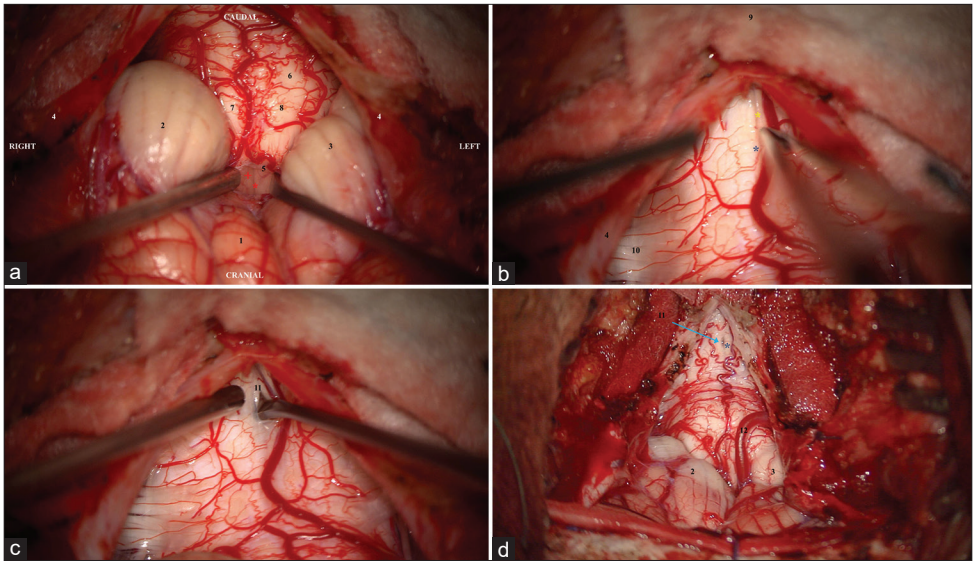

Figure 1: Intraoperative microscopic photographs following foramen magnum decompression, C1 laminectomy, and y-shaped dural and arachnoid opening to demonstrate sequence of steps for syringotomy. (a) (1) Inferior vermis, (2) right and (3) left cerebellar tonsils coagulated to superiorly retract, (4) right and left dural flaps reflected, (5) arachnoid veil obstructing 4th ventricular outflow through foramen of Magendie opened exposing Obex with (Red*) median sulcus and (Red +) base of trigones, (6) expanded and tense cervical cord with (7) right and left, and (8) dorsal columns. (b) (4) Right dural flap, (9) spinous process of C2 not exposed is covered by cottonoid, and (10) right dorsal C2 nerve rootlets from the dorsal root entry zone. (Yellow*) Median sulcus of the spinal cord demonstrated. Phase reversal has not been reliable in identifying the midline likely due to fiber stretch due to the expansile syrinx. Careful exploration can demonstrate midline pial vessels dipping into the median sulcus (Blue*). (c) (11) An ophthalmic/diamond blade can be used to make the initial syringotomy which is widened with a micro-dissector through the sulcus. With the syrinx being tense and bulging, only a small opening is needed with rapid drainage of fluid and decompression of the intramedullary cord. The cavity can be further drained using a micro-suction tip. (d) (Blue*) Midline pial vessels dipping into the median sulcus (11 with Blue arrow) The small syringotomy can be visualized with the spinal cord significantly less tense and expanded compared to previously (12). The left posterior inferior cerebellar artery is observed as are the (2 and 3) coagulated cerebellar tonsils.

with regard to gait. She underwent FMDD with concurrent syringotomy with no postoperative complications.

Tonsillar ascent and syrinx reduction were visible on her first postoperative MRI at 6 weeks. There was a segmental collapse of the syrinx with an 85% change noted in maximum transverse diameter (from 1.66 cm preoperatively to 0.25 cm postoperatively) [Figures 2c and d]. Over a follow-up period of 36 months, her headaches and sensory symptoms got better, and her spinal curve improved.

Case 2

This is a 12-year-old girl with CIM and a cervicothoracic syrinx [Figures 2e and f]. She presented with headaches and difficulties with balance. She underwent FMDD with concurrent syringotomy with no postoperative complications.

Her postoperative MRI 4 months after surgery demonstrated ascent of tonsils, a total collapse of the syrinx, and thus the complete resolution of syrinx from a maximum preoperative transverse diameter of 1.01 cm [Figures 2g and h]. Over a follow-up period of 18 months, she remained headache-free and showed improvement in balance, with stable radiology.

Case 3

This is an 8-year-old boy with CIM and holocord syrinx [Figures 2i and j]. He presented with a 3 months history of suboccipital headaches and progressive scoliotic deformity over a duration of 4 years. There were few recent episodes of urinary incontinence despite being toilet trained. He underwent FMDD with concurrent syringotomy with no postoperative complications.

A total collapse of the syrinx was demonstrated on the first postoperative MRI done 2 months after surgery with a 100% reduction from the preoperative maximum transverse diameter of 1.52 cm [Figures 2k and l]. Over a clinical follow-up period of 24 months, he was headache-free with

improvement in his urinary symptoms and stable spinal curve.

Table 1 summarizes the patient characteristics and outcomes in our short series.

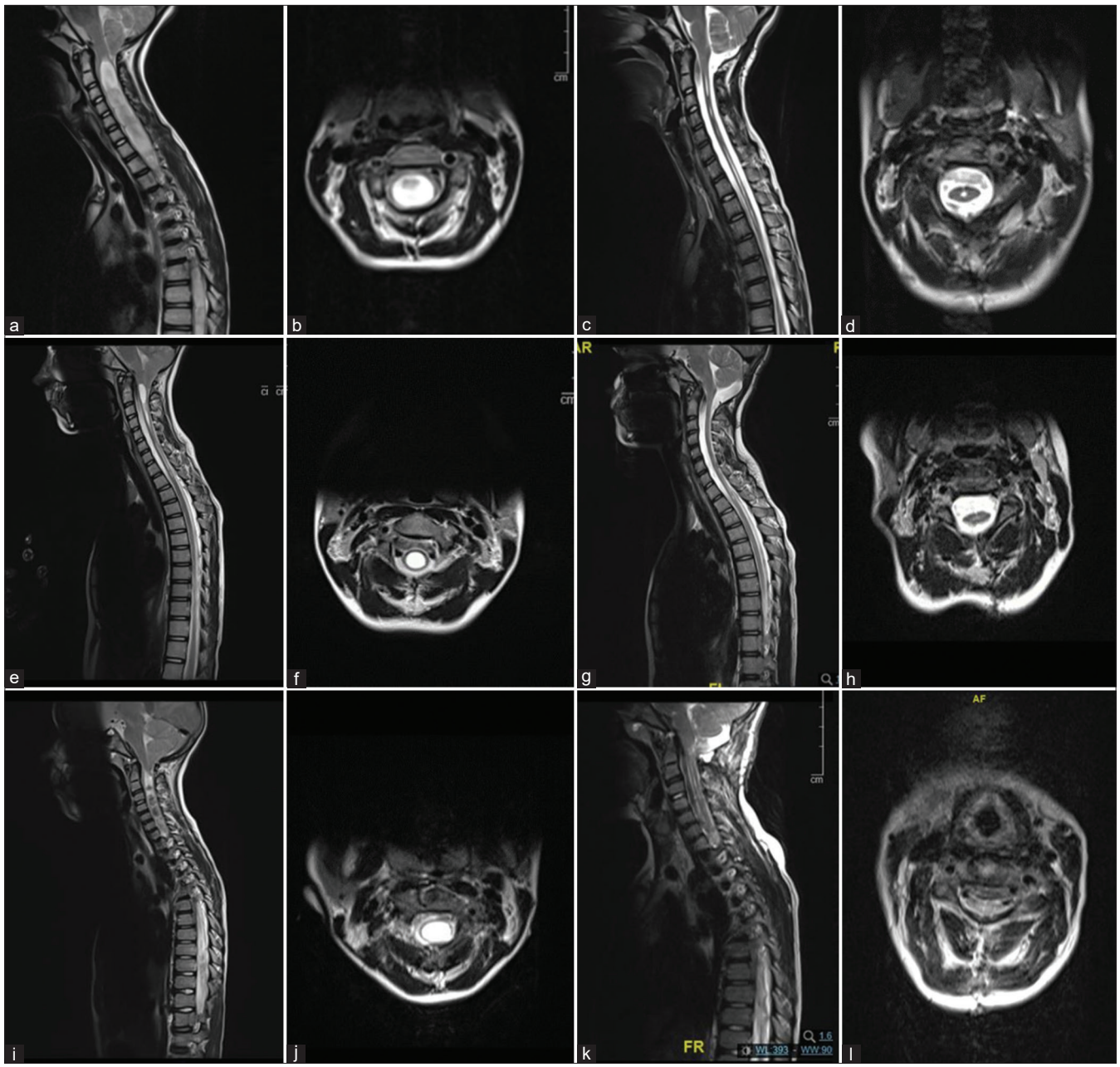

Figure 2: Pre- and post-operative magnetic resonance imaging T2-weighted images demonstrating impact on syrinx cavity in our three patients. (a and b) Case 1 preoperative sagittal and axial imaging with extensive multiseptated holocord syrinx and evidence of turbulent flow within the syrinx cavity as evidenced by the hypo-intense signal changes within. (c and d) Case 1 postoperative sagittal and axial imaging at 1½ month follow-up confirming significant reduction in all septated cavities. (e and f) Case 2 preoperative sagittal and axial imaging with cervicothoracic syrinx cavity and a thin rim of neural tissue. (g and h) Case 2 postoperative sagittal imaging at 4-month follow-up demonstrating significant resolution. (i and j) Case 3 preoperative sagittal and axial imaging with extensive holocord syrinx and minimal neural tissue overlying the dorsal aspect. (k and l) Case 3 postoperative sagittal imaging at 2-month follow-up demonstrating significant resolution of syrinx.

Table 1: Outcomes for patients treated with foramen magnum decompression with duraplasty and syringotomy.

Case no., sex and age at surgery	Preoperative symptoms	Extent of syrinx	Symptom resolution	Complications	Reduction in syrinx size	Timing of postoperative MRI
Case number 1, Female, 11 years	Sensory Headache Scoliosis	Holocord upper extent at C1	Resolution of headaches. Sensory symptoms and scoliosis improved.	Nil	Preoperative 1.66 cm → Postoperative 0.25 cm 85% reduction	6 weeks
Case number 2, Female, 12 years	Vestibular/Balance Radiological syrinx progression	Cervicothoracic upper extent at C1	Improvement in coordination.	Nil	Preoperative 1.01 cm → Postoperative no syrinx 100% reduction	4 months
Case number 3, Male, 8 years	Headache Scoliosis Some episodes of urinary incontinence	Holocord upper extent at C1	Resolution of headache. Scoliosis stabilized. Improvement in urinary symptoms.	Nil	Preoperative 1.52 cm → Postoperative no syrinx 100% reduction	2 months

MRI: Magnetic resonance imaging, F: Female, M: Male

DISCUSSION

FMD is the most common primary surgical procedure used to treat CIM with syringomyelia, the intent being to restore CSF flow dynamics across the foramen magnum thus reversing the pathophysiologic mechanisms leading to syrinx formation. However, FMD is known to fail in improving syringomyelia in a significant number of postoperative cases of CIM, and the persistence of syrinx can have permanent sequelae.[5,16,24] In an outcome analysis of 49 children with CIM, it was found that hindbrain decompression failed to improve the syrinx in 42% of cases.[1] There are studies indicating that dysesthetic pain and sensory deficits have a poorer outcome, while amyotrophy in the upper extremities responds very badly to FMD, suggesting irreversible destruction of anterior horn cells by the syrinx. Thus, the persistence of syrinx is not a desirable outcome of surgery for syringomyelia.[10]

Surgical options to treat persistent/progressive syrinx after FMD include revision of the FMD and syrinx shunting procedures such as syringo-subarachnoid, syringopleural, and syringoperitoneal shunts.[3,4,12-14,19] Surgeries such as cord transection and terminal ventriculostomy were practiced in the past.[12,13] However, there is no clear consensus on which procedure is the most beneficial of the above.[13] Therefore, in CIM with syringomyelia, the role of a surgical procedure to correct the impaired CSF flow dynamics at the craniocervical junction with concurrent syrinx drainage to achieve rapid decompression merits consideration.

Soleman *et al*. has described concurrent FMD and syringo-subarachnoid shunt for CIM and syringomyelia.[18,19] However, syrinx shunting requires another incision and exposure depending on the level of the maximum syringomyelic cavity diameter. It has been reported that a sizable number of complications in terms of shunt obstruction/dislocation, tethering of the spinal cord by the shunt, and a low CSF pressure state may occur after shunt placement.[20-22] The relatively short functional half-life of these shunts in a pediatric population is also a deterrent.[6,8,9,11]

We present a short case series of three patients with CIM and concurrent syringomyelia who underwent FMDD with the addition of a syringotomy to achieve rapid decompression of radiologically expansile and tense syrinx cavities. These patients were selected for syringotomy based on the presence of:

- Syrinx extending to the high cervical spine, accessible through our standard exposure for FMD (i.e., following FMD and C1 laminectomy)
- Expansive syrinx cavity extending to the surface of the dorsal pial spinal cord surface and median sulcus and thereby directly accessible with minimal neural tissue transgression
- Availability of intraoperative neurophysiological monitoring.

Our aim in the presence of extensive, expansile syrinx cavities in particular with rostral extension toward the medulla was to achieve a rapid decompression of the syrinx with no need for any further incisions. There was clinical improvement in all cases. There were no significant postoperative complications. Radiological improvement was reported in all three cases, with the earliest improvement in the 6th postoperative week. The syrinx transverse diameter reduction was in the range of 85–100%. There was no syrinx recurrence in any of the

three cases at their last follow-up. In a study of 85 pediatric patients, the median time to >50% reduction in syrinx anteroposterior diameter was 8 months after FMD for CIM.[5] In a prospective study, Wetjen *et al*. reported a median time of 3.6 months to ≥50% syrinx regression after FMD.[22]

Limitations to our study include small numbers. Furthermore, only selected patients are eligible for consideration, and it is not suitable for patients in whom the syrinx is below C2 level and thus not accessible through standard exposure. We feel that our case series demonstrates that syringotomy can be safely performed in selected patients with early resolution of syrinx in all patients.

CONCLUSION

FMDD with concurrent syringotomy in selected cases of CIM with high cervical syringes achieves surgical objectives in a single exposure. This procedure results in clinical improvement with no additional complications. Prolonged follow-up with a larger patient cohort is required to better establish the long-term benefit of the procedure.

Declaration of patient consent

Patients' consent not required as patients' identities were not disclosed or compromised.

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Conflicts of interest

There are no conflicts of interest.

Use of artificial intelligence (AI)-assisted technology for manuscript preparation

The author(s) confirms that there was no use of artificial intelligence (AI)-assisted technology for assisting in the writing or editing of the manuscript and no images were manipulated using AI.

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