Treatment of Holocord Syringomyelia–Chiari Complex by Posterior Fossa Decompression and a Syringosubarachnoid Shunt in a Single-Stage Single Approach

Giovanni Raffa, Stefano Maria Priola, Rosaria Viola Abbritti, Antonino Scibilia, Lucia Merlo, and Antonino Germanò

Abstract *Background:* Posterior fossa decompression with expansive duraplasty is the first-line surgical approach for the treatment of symptomatic syringomyelia associated with Chiari malformation. Despite good decompression, the clinical failure rate is reported to be up to 26%. A syringosubarachnoid (S-S) shunt may be used as a secondary option.

Methods: In this paper we describe a single-institution experience of three cases of holocord syringomyelia–Chiari complex treated with foramen magnum decompression, expansive duraplasty and an S-S shunt carried out in a single-stage single approach. Following a standard suboccipital craniectomy, patients were submitted to syrinx fenestration and simultaneous insertion of an S-S shunt through a 1-mm posterior midline myelotomy at the C2 level prior to expansive dural reconstruction.

Results: Postoperative imaging showed immediate reduction of the holocord cavities. Preoperative neurological deficits rapidly improved significantly and were stabilized at follow-up.

Conclusion: In our experience the positioning of the shunt catheter at a high level of the spinal cord (C2) did not add a significant risk of morbidity and obviated the need for a second operation and/or a separate incision in cases of clinical failure. This technique avoided the risk associated with a second surgery and its morbidity, and allowed prompt clinical recovery.

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Introduction

Since the first description of syringomyelia–Chiari complex [1], several surgical procedures have been proposed. So far, however, none of these has been able to completely resolve the disease and be proposed as a universally recognized standard [2–11]. In the last decade, surgical strategies have been focused on splitting the outer leaf of the dura mater without duraplasty [12], widening or decompression of the foramen magnum by dural microincisions alone [13] or by excision of the external dura with delamination and widening of the internal layer through longitudinal incisions [14] and, finally, posterior fossa reconstruction with duraplasty [15].

Nevertheless, a syringosubarachnoid (S-S) shunt typically placed at the largest level of the syrinx has also been suggested in patients with a large-sized cavity and central cord syndrome [3, 4, 16, 17].

Despite these numerous treatments aimed at direct and/or indirect resolution of the proposed different pathophysiological mechanisms of this complex disease, in some cases it is still possible to have an unsatisfactory clinical and/or radiological postoperative outcome after posterior fossa decompression (PFD). The current literature reports a clinical failure rate of up to 26% [18] and a radiological failure rate of up to 55% in these cases [19–21]. Therefore, treatment of syringomyelia still requires new surgical strategies to improve the prognosis in these subsets of patients.

PFD and S-S shunting are usually performed separately in different types of patients [3, 4, 16, 17, 22], eventually combined in the same patient but usually separated into two different surgical sessions [4, 9, 17, 23] or performed in a two-stage approach [16, 24].

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The aim of this study is to describe our experience with three cases of holocord syringomyelia associated with Chiari malformation submitted to standard PFD with expansive duraplasty and, simultaneously, placement of a C2 S-S shunt in a single-stage single approach.

Material and Methods

Patient Population

Between September 2010 and October 2011, three patients (two male and one female), ranging in age from 18 to 62 years (mean 46.7 years), were diagnosed with holocord syringomyelia–Chiari complex by magnetic resonance imaging (MRI). We collected and reviewed information on all nosological, preoperative and postoperative clinical symptoms and neurological signs, neuroradiological findings, and length of follow-up for each patient, as reported in Table 1. All patients signed informed consent for surgery and for scientific use of their clinical data.

The patients experienced progressive symptoms over a period ranging from 3 weeks to 2 years before diagnosis, which included a common history of dysesthesia in the lower extremities and extending to the upper ones, unsteady gait, and impairment of fine hand movements. The neurological examination on admission revealed spastic paraparesis with increased reflexes and weakness of the forearm muscles and grasp, with hypesthesia in all cases, increased muscle tone in the upper extremities in two patients and hand deformity (*main en griffe*) in one.

We semiquantitatively assessed the severity of cervical myelopathy by using the modified Japanese Orthopaedic Association (mJOA) scale [25]. The mean preoperative mJOA score was 9.4 (range 9–10).

MRI of the spinal cord showed caudal dislocation of the cerebellar tonsils into the cervical canal below the foramen magnum, ranging from 10 to 17.2 mm (mean 14.1 mm) (Table 1), associated with a large holocord syringomyelia with thinned spinal cord tissue and an obliterated spinal subarachnoid space extending from C2 to T11 level (Fig. 1a). The preoperative mean diameter of the cavity, calculated at the largest level on the axial T2 sequence, was 12.2 mm (range 8.4–18.1 mm) (Fig. 1b; Table 1). The syrinx/canal index proposed by Hida and Iwasaki [3] exceeded 90% in all cases (range 91.3–97.5%) (Table 2).

Table 1 Summary of nosological, neuroradiological and clinical data

Case no.	Patient age (years); sex	Time to diagnosis (months)	Syrinx location	Cerebellar tonsil caudal dislocation (mm)	Main diameter of syrinx (mm)		Symptoms and signs[a]		Follow-up duration (months)
					Preoperative	Postoperative	Preoperative	Postoperative	
#1	18; male	12	C2–T11	17.2	18.1	6.2	*mJOA score 10* I: 2 (right hand *en griffe*) II: 2 III: 1/1/1 (>right side) IV: 3	*mJOA score 16* I: 3 (right hand *en griffe*) II: 4 III: 2/2/2 IV: 3	60
#2	62; male	24	C2–T7	10	8.4	2.8	*mJOA score 9* I: 2 II: 2 III: 1/0/1 IV: 3 Dysphagia and odynophagia	*mJOA score 16* I: 4 II: 3 III: 2/2/2 IV: 3	58
#3	60; female	15	C2–T5	15	10	2.5	*mJOA score 9* I: 2 II: 2 III: 1/1/0 IV: 3	*mJOA score 17* I: 4 II: 4 III: 2/2/2 IV: 3	48

[a]Symptoms and signs according to the modified Japanese Orthopaedic Association (mJOA) scale

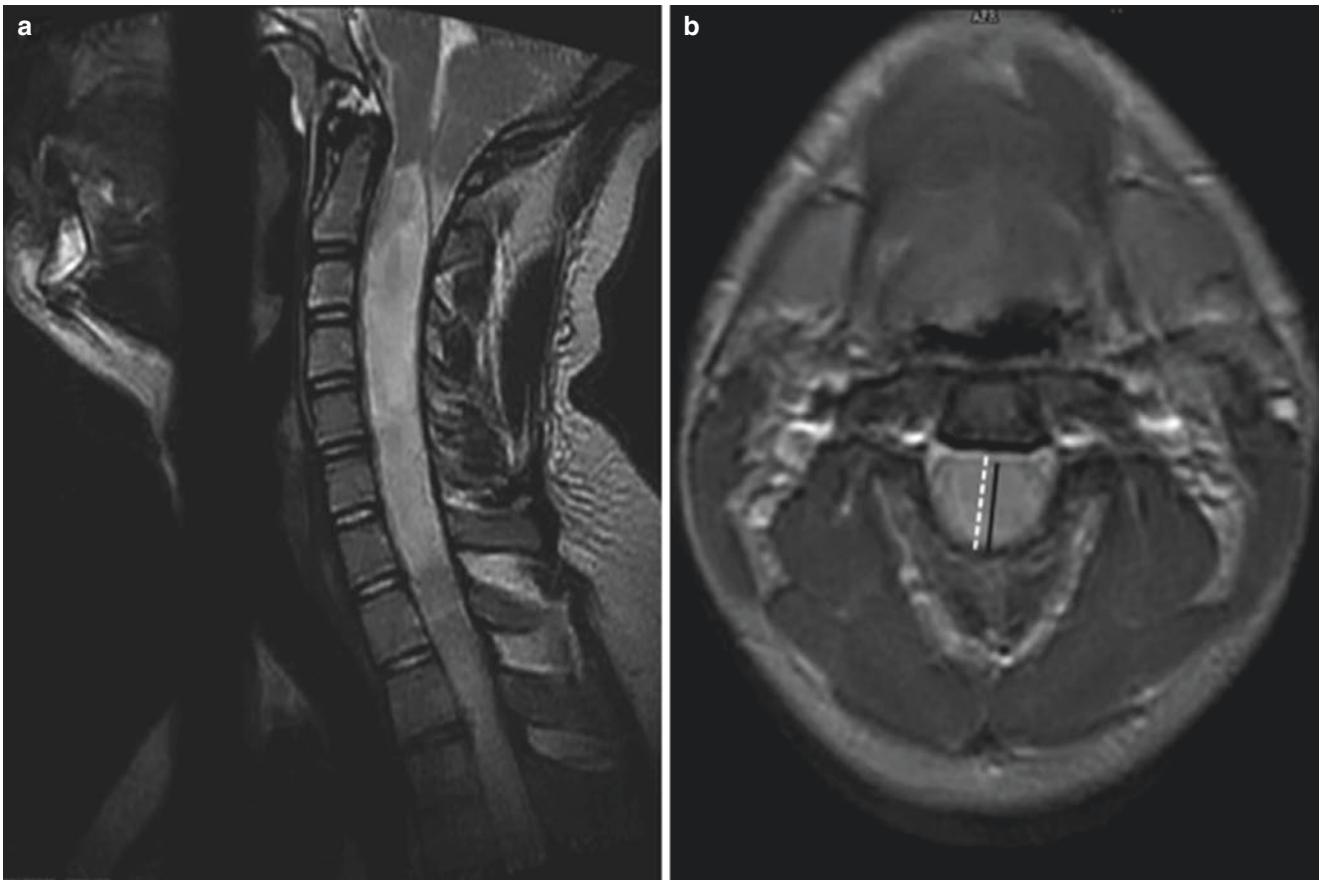

Fig. 1 Preoperative magnetic resonance imaging (MRI) of case #1. (**a**) Sagittal T2-weighted study demonstrating Chiari malformation type I and a large holocord syringomyelia extending from C2 to T11. (**b**) Axial T2-weighted image; the syrinx/canal index at the C2 level exceeds 90%. (The *dashed line* indicates the maximum diameter of the spinal canal; the *solid line* shows the diameter of the syrinx cavity)

Table 2 Pre- and postoperative syrinx/canal indexes [9]

	Syrinx/canal index (%)	
Case no.	Preoperative	Postoperative
#1	97.5	31
#2	91.3	59.7
#3	92.5	42.3

Operative Procedure

Patients were submitted to standard PFD. The operation was carried out in the prone position, with a midline skin incision extending from the external occipital protuberance to the spinous process of C3. A suboccipital craniectomy (3 × 3 cm) with opening of the foramen magnum, removal of the posterior arch of C1 and laminectomy of C2 was performed. The dura mater at the cervico-occipital junction was exposed and an operating microscope was brought in. The dura mater was carefully opened in a Y shape and the arachnoid membrane was incised, showing migration of the cerebellar tonsils into the spinal canal.

At this point, a 1-mm-long posterior C2 myelotomy was performed on the midline, targeted to the syrinx, in a relatively avascular area (Fig. 2a). With use of microscissors, the cavity was fenestrated, obtaining immediate outflow of cerebrospinal fluid (CSF) under pressure. Thereafter, an antibiotic-impregnated silicone catheter with multiple fenestrations at its end [26] (Codman® Bactiseal® EVD Catheter Set; Codman & Shurtleff, Inc., Raynham, MA, USA) was inserted (Fig. 2b). The catheter was placed in a cephalic direction to maintain the physiological CSF outflow, with the caudal part positioned in the subarachnoid space at the C3 level. The distal lateral catheter's outlets were left outside the cavity to facilitate free CSF subarachnoid drainage. The catheter was left in place without any securing [27] to avoid iatrogenic spinal cord fixation. Finally, an expansive reconstructive posterior fossa duraplasty was performed with a dural substitute graft (Gore Preclude® PDX; WL Gore & Associates, Inc., Flagstaff, AZ, USA).

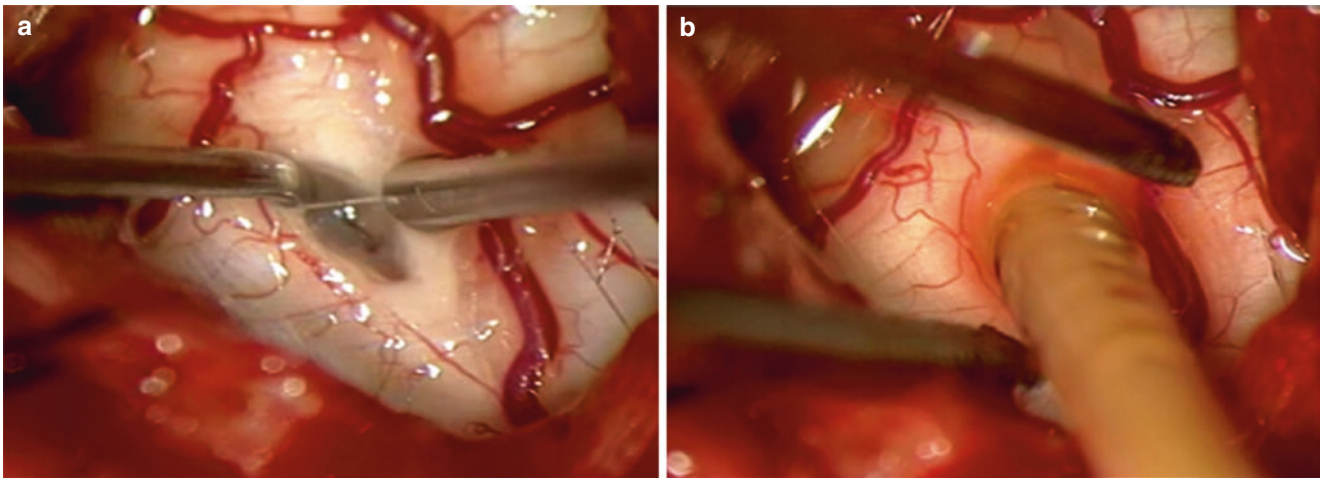

Fig. 2 Intraoperative pictures of case #1. (**a**) Posterior 1-mm midline myelotomy performed in a relatively avascular area at the C2 level following posterior fossa decompression. (**b**) Insertion of a syringosubarachnoid shunt into the syrinx cavity through the myelotomy

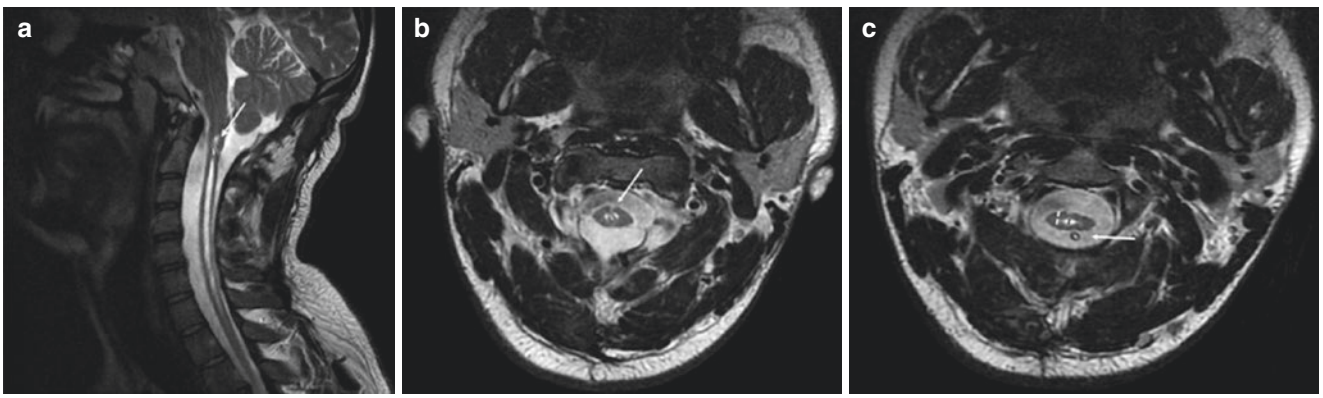

Fig. 3 Postoperative magnetic resonance imaging (MRI) of case #1. (**a**) Sagittal T2-weighted study demonstrating the catheter inside the syrinx (*arrow*) and significant reduction of the holocord cavity. (**b**, **c**) Axial T2-weighted images showing the catheter within the cavity (*arrow*), exiting from the spinal cord at the C2 level and lying freely in the subarachnoid space, and confirming the reduction of the syrinx/canal index (*dashed line*)

Results

Clinical Course and Outcome

The patients had a fast postoperative recovery. Improvement of the preoperative leg weakness and hypesthesia was immediately evident 1 day after surgery. At a mean follow-up of 55 months after surgery (range 48–60 months), all patients had achieved satisfactory recovery of their neurological symptoms. At follow-up the mean mJOA score was 16.4 (range 16–17). In detail, motor weakness had improved by two points in two cases and by one point in the remaining patient, whose right hand was still *en griffe*. Sensitive deficits showed a three-point improvement in the mJOA score in one case and four points in the other two. No bladder dysfunction was present preoperatively or postoperatively in any of the patients. In case #2, the patient experienced dysphagia and odynophagia, but they rapidly resolved in the postoperative period. The patients remained neurologically stable at the follow-up evaluations up to 60 months postoperatively (Table 1).

MRI performed in the postoperative period demonstrated optimal PFD together with a significant reduction in the holocord cavity (Fig. 3a). Axial T2 MRI sequences showed that the maximum diameter of the syrinx cavity at the C2 level was reduced by 75–66.6% (mean 69.1%) to 3.8 mm (range 2.5–6.2 mm) (Fig. 3b, c). Therefore, the syrinx/canal index was reduced to 31% (range 31–59.7%) (Table 2). No surgical complications or damage of the spinal cord at the myelotomy entry point were observed.

Discussion

In this paper we report a retrospective analysis of a single-centre experience of three patients affected by holocord syringomyelia and Chiari malformation treated at our insti-

tution with combined PFD, expansive duraplasty and simultaneous insertion of a C2 S-S shunt. All surgical procedures were carried out in a single-stage single approach. This strategy was performed with the aim of combining the two most widely used surgeries for management of this disease complex. The combination of these techniques allowed effective and less invasive treatment with prompt clinical and radiological recovery.

In recent years we have observed a growing body of literature focused on analysis of the pathophysiological mechanisms of syringomyelia–Chiari complex, suggesting different surgical procedures [1]. PFD is one of the most widely used surgical procedures for treatment of this disease complex. Nevertheless, there is still a significant proportion of patients who fail to improve, either clinically or radiologically, after PFD.

Over the years, several surgical strategies have been proposed to improve the clinical and neuroradiological results of PFD: removal [12] or widening by transverse microincisions [13] of the outer layer of the dura mater; opening of both dural layers, leaving the arachnoid intact [14]; and posterior fossa reconstruction with expansive dural grafting [15].

Although none of these approaches is free of complications [12, 28], PFD with its variants remains the most widely performed surgical approach for syringomyelia–Chiari complex. Therefore, the positioning of an S-S shunt is still considered a secondary procedure that should be performed in the event of failure of the former procedure [29].

There is evidence that in some cases, patients with small syringes could experience progression despite craniovertebral decompression and could require a second surgery for the syrinx shunt [16]. Conversely, in some cases the syrinx resolves after shunting alone, even if PFD is not performed and the CSF flow obstruction at the level of the foramen magnum has not been removed [10]. Subsequently, to ensure postoperative relief of symptoms, PFD and S-S shunting are often both necessary to achieve a satisfactory outcome in many cases.

In the literature, various types of shunts have been described, including S-S [3, 4, 7, 9], syringoperitoneal [5], syringopleural [11] and thecoperitoneal [10] shunting. These procedures are associated with a good postoperative outcome, at least in the early postoperative period. Nevertheless, there are some reports of delayed complications and disadvantages, such as shunt malfunction, slippage and cord injury [4, 30]. Moreover, it has been reported that many patients treated using shunt procedures alone have required a subsequent revision or PFD [29].

In the light of such evidence, PFD and S-S shunts should be considered two complementary procedures, acting on two different pathophysiological mechanisms: compression at the craniovertebral junction and alteration of the normal CSF circulation. Actually, they are usually performed separately in different subset of patients [3, 4, 16, 17, 22]. In many cases, they are combined but done during two separate surgical sessions [4, 9, 17, 23]. In a few cases, they are performed during the same operation [8] or in a two-stage approach [16, 24]. The shunt is usually inserted at the largest level of the syrinx [3, 4, 9, 16, 17, 23, 24, 30, 31], usually at the lower cervical or midthoracic level. The choice of the catheter insertion point usually corresponds to the largest portion of the syrinx, where its insertion is considered to ensure better decompression. This has been reported even for multilocular syringes [9]. Nevertheless, in the literature there is also a lack of consensus regarding myelotomy in terms of its exact location (dorsal root entry [3, 4] versus midline entry [16]) and its length (1 cm [8] versus 2 mm [17]), and the direction of the shunt (craniocaudal [17] versus caudocranial [31]).

Considering the conflicting literature reports, it is actually difficult to prove the usefulness of any therapeutic procedure in decreasing intramedullary tension. Currently there are no objective criteria and no standard radiological parameters to suggest which technique will better resolve a syrinx when it is associated with Chiari malformation. The clinical and radiological recurrence rate of syringes following PFD with duraplasty alone in patients with syringomyelia–Chiari complex has been reported to be up to 26% [18], with a long-term radiological failure rate ranging between 6% [18] and 55% [19–21].

In the light of this evidence, there is still a need for definition of an effective strategy for treatment of syringomyelia–Chiari complex, aimed at improving the postoperative outcome. In the literature, there are only a few reports describing the combination of PFD with an S-S shunt, or insertion of an S-S shunt following failure of initial PFD [8, 9, 16, 22, 24]. In those studies it was concluded that the combination of PFD and an S-S shunt was superior to PFD or shunt placement alone: resolution of the syrinx was achieved in a higher percentage of patients, without recurrences [8, 9, 16, 22, 24]. In the present study, we documented that simultaneous PFD and S-S shunt insertion could be performed in a single-stage approach and was associated with satisfactory short- and long-term postoperative outcomes. Moreover, the strength of this strategy is that it could avoid the need for subsequent surgeries after treatment failure in cases in which PFD or an S-S shunt alone is not sufficient to provide satisfactory relief from symptoms.

Conclusion

In this paper we have presented a different surgical strategy to treat holocord syringomyelia–Chiari complex in a single-stage procedure. The positioning of the shunt catheter at a high level of the spinal cord (C2) did not add a significant

risk of morbidity. This technique, which links two already standardized procedures in a single-stage single approach, avoided the risk associated with a second surgery and its morbidity, and allowed prompt clinical recovery.

We recognize that a retrospective single-institution experience does not allow us to draw definitive conclusions. However, our report supports the existing evidence that the combination of both techniques performed in the same surgical approach is safe, is feasible and contributes to achieving faster recovery times without adding a significant risk of morbidity related to both the site of catheter insertion and a second surgery, in comparison with a standard two-stage approach.

Further studies are required to evaluate the long-term efficacy of this strategy.

Compliance with Ethical Standards No financial support was received for this work.

Competing Interests The authors declare that they have no competing interests.

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