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ORIGINAL ARTICLE

Treatment failure of syringomyelia associated with Chiari I malformation following foramen magnum decompression: how should we proceed?

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Abstract

The preferred treatment of patients with persistent, recurrent, or progressive syringomyelia after foramen magnum decompression (FMD) for Chiari I (CMI)-associated syringomyelia is controversial, and may include redo FMD, stabilization, or shunting procedures (such as syringopleural or syringo-subarachnoid shunts). We describe our experience in treating these patients and discuss the treatment modalities for these patients. We retrospectively collected data of CMI patients with persistent, recurrent, or progressive syringomyelia after FMD. In addition to baseline characteristics, surgical treatments and neurological and radiological outcomes were assessed. Further, we assessed through uni- and multivariate analyses possible technical, surgical, and radiological factors which might lead to failed FMD. Between 1998 and 2017, 48 consecutive patients (35 females (73%), average age 16.8 ± 11.5 years) underwent FMD for a syringomyelia-Chiari complex. Twenty-four patients (50%) underwent surgical treatment for a persistent ($n = 10$), progressive ($n = 12$), or recurrent ($n = 2$) syringomyelia 21.4 ± 27.9 months (median 14.6 months, range 12 days–134.9 months) after FMD. Of all analyzed factors, only extradural FMD was significantly associated with lower failure rates. Two patients (8%) underwent redo FMD, 18 (75%) underwent 19 syringo-subarachnoid-shunts, and 4 (17%) had 6 cranial CSF diversion procedures. The overall follow-up time was 40.1 ± 47.4 months (median 25 months, range 3–230 months). Based on our results, 50% of the patients undergoing FMD for syringomyelia-Chiari complex may require further surgical treatment due to persistent, progressive, or recurrent syringomyelia. Treatment should be tailored to the suspected underlying pathology. A subgroup of patients may be managed conservatively; however, these patients need close clinical and radiological follow-ups. The technical aspects of FMD in CMI-syrinx complex should be the focus of larger studies, as an effort to improve failure rates.

Keywords Foramen magnum decompression · Chiari I malformation · Syringomyelia · Spinal cord · Hydrocephalus · Syringo-subarachnoid shunt

Introduction

Syringomyelia accompanies Chiari I malformation (CMI) in 35–75% of the cases [2, 7, 27]. Foramen magnum decompression (FMD) remains the recommended primary surgical treatment, although some authors advocate upfront shunting of the syringomyelia [11, 13]. Persistent, progressive, or recurrent syringomyelia following FMD is described in up to 66% of the cases [2, 5, 7, 27]. To date, no consensus exists regarding the indications and the treatment modality for persistent, progressive, or recurrent syringomyelia after FMD. Surgical options include redo FMD or shunting procedure (syringopleural or syringo-subarachnoid shunt) [2, 7, 10, 17, 27, 28]. If hydrocephalus is apparent, ventricular shunting or endoscopic third ventriculostomy (ETV) is advised [9, 10, 19]. Recently, atlanto-axial fusion was proposed as an additional treatment option [8].

The aim of the current study is to report the rate of failed FMD for the treatment of syringomyelia-Chiari complex, and report our experience treating these patients.

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Methods

The study protocol was approved by the local ethics committee (Institutional Review Board/Ethics (Helsinki) Committee Tel Aviv Sourasky Medical Center), while informed consent was waived by the ethics committee due to the retrospective nature of the study. The manuscript was designed according to the STROBE guidelines. Inclusion criteria were patients of any age, who underwent FMD for syringomyelia-Chiari complex. We included only patients with Chiari Type I malformation, since none of the patients presented with Chiari Type 1.5, while patients with Chiari type 2 were excluded from this study. We initially identified 54 consecutive patients with a syringomyelia-Chiari complex undergoing FMD surgery at our department between 1998 and 2017. Six patients underwent FMD concurrently with syringo-subarachnoid shunt (SSS) and were therefore excluded from this study, resulting in a study population of 48 patients. Failed FMD was defined as repeat surgery do to new or progressive neurological symptoms (unrelated to the syringomyelia size), extensive persistent syringomyelia with cord compression (which most probably will lead to neurological symptoms), or syringomyelia progressing over time, even if no or only minimal symptoms are apparent. We defined the FMD to be sufficient if no regrowth of the bone (especially in patients where FMD was done at a very young age) was seen, no clear fourth ventricular outlet obstruction (FVOO) was apparent, and sufficient CSF was surrounding the brainstem on postoperative MRI also compared to the preoperative images. All pre- and postoperative MRIs were discussed with a pediatric neuroradiologist. Patient data were retrospectively collected through medical and radiological records. All patients were followed clinically and radiologically with cranio-spinal magnetic resonance imaging (MRI) (mean 40.1 months ± 47.4 months). Since 2003, FMD and SSS procedures were all done using intraoperative monitoring (IOM) measuring the somatosensory-evoked potentials (SSEP) and transcranial electric motor-evoked potentials (MEP) [3, 23]. Our surgical technique for SSS has been published recently and is therefore not emphasized on within this manuscript [29]. Variables included in the univariate and multivariate analyses as potential risk factors leading to failure of FMD were age, gender, occurrence of scoliosis, hydrocephalus, or basilar invagination, complications (chemical or bacterial meningitis, CSF leak, pseudomeningocele, and intracranial bleed), FMD size, intra-/extradural FMD, number of cervical laminas resected, resection/shrinkage of the cerebellar tonsils, type of dural graft used, syringomyelia size and length on preoperative MRI, tonsillar herniation (in mm), and sufficient CSF space assessed by an independent pediatric neuroradiologist on postoperative MRI. For the assessment of possible instability, the MRI before FMD was assessed for the clivoaxial angle (CXA) and the ventral brainstem compression (pB-C2) according to Bollo et al. [4].

Statistics

All statistical analyses were done using SPSS (Statistics Version 22.0, IBM Corp, 2013, Armonk, NY, USA). Contingency tests were done using Fisher's exact test. For non-parametric tests, the Mann-Whitney *U* test was used. For the analysis of possible risk factors leading to persistent, progressive, or recurrent syringomyelia, univariate analysis was undertaken. Values showing a p value under 0.1 were inserted into a multivariate logistic regression model for further analysis. A p value of < 0.05 was considered significant.

Results

Patient population

Of the 48 consecutive patients included, 35 patients (73%) were female, 79% were pediatric patients (under the age of 18 years), with an average age of 16.8 years (± 11.5 years, median 14 years, range 2–51 years). The patients' characteristics and clinical data are presented in Table 1. Patients were followed up clinically and by MRI for a mean duration of 40.1 months (± 47.4 months, median 25 months, range 3–230 months).

FMD for syringomyelia-Chiari complex

Out of the 48 patients included, extradural FMD with outer dural layer longitudinal cuts was performed in 10 patients (21%), and intradural FMD was performed in 38 patients (79%). Indications for intra- or extradural FMD were surgeon's preference. In all patients undergoing intradural FMD, duraplasty using an artificial graft was inserted (Table 2). The graft was sutured in watertight fashion using a non-resorbable, running suture. The graft material and suture technique were the same and standardized for all patients undergoing intradural FMD. The distribution of craniotomy size, the number of laminas resected, and tonsil shrinkage/resection are listed in Table 2. In one case, FMD was done concurrently with an occipital EVD due to hydrocephalus. Postoperatively, the EVD was closed, and then subsequently removed. Mean hospitalization time after FMD was 7.2 days (± 7.0 days, median 6 days, range 3–45 days). Surgical morbidity occurred in six patients (13%); details are presented in Table 2. Five complications were FMD-associated, while in one patient the complication was due to the concomitant EVD insertion (local intraparenchymal bleed). Potential losses during intraoperative monitoring did not occur in any of the cases.

Rate of FMD failure

During the follow-up period in 12 patients (25%), the syringomyelia progressed; in 19 patients (40%), it remained

Table 1 Patient characteristics and clinical data for 48 patients undergoing foramen magnum decompression for syringomyelia-Chiari complex

Variable	*n* (%)
Age (years ± SD)	16.8 (± 11.5)
Sex (female)	35 (72.9)
Hydrocephalus at presentation	1 (1.9)[×]
Symptoms at presentation	48 (92.3)
Headache	16 (33.3)
Cranial nerve deficits	6 (12.5)
Dysesthesia	7 (14.6)
Hypesthesia	4 (8.3)
Obstructive sleep apnea	4 (8.3)
Ataxia	3 (6.3)
Motor deficits	3 (6.3)
Pyramidal signs	3 (6.3)
Torticollis	2 (4.2)
Vomiting	1 (2.1)
Vertigo	1 (2.1)
Papilledema	1 (2.1)
None	4 (8.3)
Comorbidities	
Ehlers-Danlos syndrome	1
Atrioventricular block (grade I)	1
Atrioventricular block (grade III)	1
S/P ventriculoperitoneal shunt	1*
Guillain Barré syndrome	1
Pylomyxoid astrocytoma	1[#]
Down syndrome	1
Premature birth	2[+]
Hypothyroidism	1
Dyslipidemia	1
Crouzon syndrome	2[^]
Migraines	1
Factor XI deficiency	1
S/P microvascular decompression (Janetta)	1[~]
Obesity	3

[×] Patient underwent concomitant FMD with EVD placement

*Underwent VP shunt before FMD due to hydrocephalus and syringomyelia. Bowel perforation after VP shunt and surgical repair. Suffered in addition a subdural hematoma due to over drainage, which was surgically evacuated

[#] Underlying pilomyxoid astrocytoma treated by subtotal resection, radio- and chemotherapy, post radiation developed Moya-Moya, post chemotherapy developed glomerulonephritis

[^] One of the Crouzon patients suffered a basal encephalocele which was treated endoscopically, underwent Le-Fort distraction and VPS due to hydrocephalus before FMD treatment

[+] One of the prematurely born patients, suffered IVH grade I, hydrocephalus which was treated with a VPS, Choanal atresia, suspected Marshal Smith syndrome, chronic lung disease (O_2 dependent)

[~] Suffered from hemifacial spasm which was treated by microvascular decompression (Janetta)

Table 2 The distribution of craniotomy size, the amount of laminas resected, tonsil shrinkage/resection, extradural or intradural procedure, and complications in 48 patients with syringomyelia-Chiari complex undergoing foramen magnum decompression (FMD)

Variable	*n* (%)
Craniotomy size	
2.5 × 2.5 cm	2 (4.2)
2 × 3 cm	5 (10.4)
2.5 × 3 cm	1 (2.1)
3 × 3 cm	38 (79.2)
3 × 4 cm	2 (4.2)
Laminectomy	
C1	33 (68.8)
C1 and partial C2	3 (5.7)
C1–2[”]	11 (22.9)
C1–3[”]	1 (1.9)
Extradural FMD	10 (20.8)
Intradural FMD	38 (79.2)
Tonsil shrinkage	21 (43.8)
Tonsil resection	9 (18.75)
Dural graft	38 (79.2)
Graft material	
Bovine pericardium graft	33 (86.8)
Duragen®	3 (7.9)
Fascia	1 (2.6)
Gortex	1 (2.6)
Complications	6 (11.5)
Chemical meningitis	2 (4.2)
Bacterial meningitis	1 (2.1)[α]
CSF leak	2 (4.1)
Clinically significant pseudomeningocele	1 (2.1)
Hydrocephalus	1 (2.1)[α]
Cranial nerve palsy	1 (2.1)[Ω]
Bleed	1 (2.1)*

[”] C2 and C3 laminectomies were performed in cases where the tonsils were displaced downwards up to these levels showing local pressure on magnetic resonance imaging

[α] One month after FMD wound revision due to CSF leak, bacterial meningitis, received antibiotics, developed hydrocephalus, which was treated with a ventriculoperitoneal shunt (VPS). Due to abdominal injury after VPS, laparoscopic repair was done the same day

[Ω] Bilateral vocal cord palsy, vent dependent, underwent tracheostomy. With intensive rehabilitation treatment, complete recovery after a couple of months

*Patient underwent concomitant FMD with EVD placement and suffered from an asymptomatic intraparenchymal bleed at the EVD insertion site

unchanged; in eight patients (17%), it improved but did not resolve completely; in seven patients (15%), the syringomyelia resolved completely; and in two patients (4%), the syringomyelia initially resolved completely but recurred after a period of time. Radiological measurements of syrinx size

and length and the geometry of the syrinx before and after FMD are presented in Table 3.

Twenty-four (50%) patients underwent, after a mean of 21.4 months (± 27.9 months, median 14.6 months, range 12 days–134.9 months), additional surgery due to failed FMD. All 12 patients (44%) with progressive syringomyelia, seven of whom displayed new ($n = 3$), persistent ($n = 3$), or progressive ($n = 1$) symptoms and five patients without symptoms. Nine (33%) of the 19 patients with stable syringomyelia that displayed new ($n = 1$), persistent ($n = 2$), persistent and new ($n = 3$), or progressive ($n = 3$) symptoms. One (4%) of the eight patients with persisting but slight improvement of the syringomyelia showing new symptoms, and both patients (7%) with recurrent syringomyelia who both showed new symptoms. Before undergoing FMD, the syrinx size and length in the failed FMD group (61.9 ± 41.1 cm^3 and 10.6 ± 5.9 levels, respectively) were comparable to the conservatively treated group (49.0 ± 61.9 cm^3 and 9.9 ± 5.9 levels, respectively; $p = 0.3$ and $p = 0.85$, respectively). After undergoing FMD, the syrinx size was significantly larger in the failed FMD group (79.4 ± 38.0 cm^3 and 31.2 ± 29.2 cm^3; $p < 0.001$), while the syrinx length remained comparable in both groups (11.3 levels ± 5.3 levels and 8.9 ± 6.2 levels; $p = 0.19$). A preoperative holocord syrinx was seen in two patients (8%) of the failed FMD group and in three patients (13%) of the conservative treatment group ($p = 1$), respectively.

Of the 24 patients who underwent additional surgery, 18 patients (75%) underwent 19 SSS procedures, four patients (17%) had six CSF diversion procedures (EVD, ETV, or shunting), and two patients (8%) underwent redo FMD, of which one also underwent an endoscopic third ventriculostomy (ETV) and a ventriculoperitoneal shunt (VPS) at a later stage (Table 3). Within the follow-up time of the study, none of the patients undergoing SSS needed a redo FMD at a later time point and vice versa. The median follow-up time after the second surgery was 15.8 months (mean 29.1 ± 36.7 months).

Of the other 24 patients who were followed conservatively after FMD (mean 129.6 ± 116.0 months), six patients (24%) showed persisting and unchanged syringomyelia, nine patients (36%) showed persisting and improving syringomyelia, and nine patients (36%) had complete resolution of the syringomyelia (Table 3). During the follow-up time after FMD surgery, all patients were asymptomatic, or showed minimal symptoms (e.g., headaches) which were clearly not associated with the persistent syringomyelia.

Of all analyzed factors in univariate analysis, extradural FMD was associated with lower failure rates ($p < 0.01$) and suffering a postoperative complication rate was associated with higher failure rates ($p = 0.02$). However, when analyzing each complication separately, no correlation with FMD failure was found. In multivariate regression analysis, only extradural FMD (OR 24.3, 95% CI: 1.9, 316.9; $p = 0.015$) correlated significantly with lower FMD failure rates. All other variables showed no significant correlation with FMD failure. The CXA and pB-C2 measurements did not show a significant difference ($p = 0.33$ and $p = 0.22$, respectively) between the failed FMD group and the conservative treatment group (Table 3).

Redo FMD

Redo FMD was undertaken in two cases (4%) (one 2 months and the other 2.5 years after the first surgery) due to persistent tonsillar herniation leading to brainstem compression, impaction of the foramen magnum, and failure of syringomyelia improvement in both cases. In one patient, ventricular enlargement and a big pseudomeningocele were apparent. In both cases, the primary FMD procedure was intradural, with opening of the arachnoid. Both patients underwent duraplasty with an artificial dura patch. Both patients suffered from typical Chiari-related headaches. Intraoperative findings were local mechanical compression of the tonsils by an inward fold in the dura patch in one patient, and tonsil herniation and brainstem compression in the second case. In both patients, the tonsils were coagulated or partially resected, arachnoidal webs opened if apparent, and duraplasty completed using an artificial dura patch. Surgical morbidity is described in Table 3. Overall, on follow-up MRI, shrinkage of the syringomyelia was seen in one patient, and complete syringomyelia regression in the other.

SSS

SSS was the treatment of choice in 18 patients (38%) with persistent or progressing syringomyelia after FMD. Of these, 13 patients (72%) showed neurological symptoms, while 5 patients (28%) were asymptomatic and underwent surgery due to clear progression of the syrinx. One patient underwent two SSS procedures due to recurrence of her syringomyelia. The syringomyelia was approached through cervical ($n = 6$, 32%), thoracic ($n = 11$, 58%), and cervico-thoracic ($n = 2$, 11%) laminectomy or laminoplasty. In the thoracic region, we opted to perform a laminectomy over a laminoplasty, believing that a limited, one- or two-level thoracic laminectomy is very rarely associated with secondary spinal deformity. In the cervical region, laminoplasty was done whenever possible. The syringomyelia was opened through a myelotomy at the dorsal root entry zone (DREZ) in four patients (21%), and at the midline in 15 patients (79%). In one patient, the procedure was aborted, since post infectious arachnoiditis due to meningitis after the primary FMD procedure precluded us from inserting the catheter into the subarachnoid space. However, in this case, the syringomyelia was drained intraoperatively.

Table 3 Natural history of 48 patients undergoing foramen magnum decompression (FMD) for syringomyelia-Chiari complex, including distribution of treatment, complications of treatment, and follow-up time/time to treatment

Variable	*n* (%)
Syringomyelia status before FMD (*n* = 48)	
Holocord syrinx	5 (10.4)
Focal syrinx	43 (89.6)
Syrinx diameter (mean ± SD, cm^2)	55.3 (± 39.6)
Syrinx length (mean ± SD, vertebra levels)	10.2 (± 5.8)
Radiological measurements of possible instability	
Failed FMD group (*n* = 24)	
CXA (mean ± SD)	148.8 (± 9.1)
pB-C2 (mean ± SD, mm)	7.9 (± 1.5)
Conservative group (*n* = 24)	
CXA (mean ± SD)	145.9 (± 9.4)
pB-C2 (mean ± SD, mm)	8.3 (± 1.3)
Syringomyelia status after FMD (*n* = 48)	
Progressing	12 (25.0)
Persistent unchanged	19 (39.6)
Persistent improved[+]	8 (16.7)
Recurred	2 (4.2)
Resolved[+]	7 (14.5)
Holocord syrinx	5 (10.4)
Focal syrinx	43 (89.6)
Syrinx diameter (mean ± SD)	56.4 (± 41.6)
Syrinx length (mean ± SD)	10.1 (± 5.8)
Treatment (*n* = 48)	
Conservative/follow-up	24 (50.0)
Syringo-subarachnoid shunt (SSS)	18 (37.5)°
CSF diversion procedure	4 (8.3)
Redo FMD	2 (4.2)[∞]
Symptoms after FMD leading to surgical treatment (*n* = 24)[β]	
Headache	7 (29.2)
Ataxia	5 (20.8)
Dysesthesia	5 (20.8)
Hypesthesia	4 (16.6)
Motor deficit	4 (16.6)
Pyramidal signs	3 (12.5)
Vomiting	2 (8.3)
Hydrocephalus	2 (8.3)
Cranial nerve palsy	1 (4.2)
Obstructive sleep apnea syndrome	1 (4.2)
Pseudomeningocele	1 (4.2)
Torticollis	1 (4.2)
Urinary incontinence	1 (4.2)
Cerebrospinal fluid (CSF) leak	1 (4.2)
None (progressing syringomyelia)	5 (20.8)
Complications after SSS (*n* = 19)	5 (26.3)
Kyphosis treated with spinal fusion	1 (5.3)[”]
Wound dehiscence	2 (10.5)*
Urine incontinence	1 (5.3)[#]
Cranial nerve palsy	1 (5.3)[#]
Redo SSS	1 (5.3)[Δ]
Complications after redo FMD (*n* = 2)	1 (50.0)
Fourth ventricular outlet obstruction (FVOO)	1 (50.0)[×]
Complications after CSF diversion procedure (*n* = 6)	1 (16.7)
Cerebellar ptosis	1 (16.7)
Neurological outcome after surgical treatment (*n* = 24)	
Improved	14 (58.4) [0/12/2][^]
Stable	8 (33.3) [2/4/2][^]
Worse	2 (8.3) [0/2/0][^]
Radiological outcome after surgical treatment (*n* = 24)	
Progressive	0 (0)
Persistent unchanged	2 (8.3) [0/1/1][^]
Improved	17 (70.8) [1/15/1][^]
Resolved	5 (20.9) [1/2/2][^]
Mean time to syringomyelia treatment	Months (± SD)
Overall (*n* = 24)	21.4 (± 27.9)
SSS	26.3 (± 30.0)
Redo FMD	16.4 (± 15.1)
CSF diversion	2.0 (± 1.2)
Follow-up time for conservatively treated patients (*n* = 24)	129.6 (± 116.0)

[+] Improved syrinx is defined as syrinx with smaller diameter and/or shorter length on postoperative imaging when compared to preoperative imaging. Resolved syrinx is defined as a syrinx which is not appreciated on postoperative imaging

°19 SSS procedures in 18 patients

[∞] 4 patients underwent 6 CSF diversion procedures

[β] These symptoms developed mostly over time after the FMD procedure

[”] Kyphosis occurred in 3 patients, while only one required fusion

*One patient required revision surgery, one patient treated with antibiotics only

[Δ] After the first SSS, clinical improvement, however, syringomyelia progressed with minor swallowing problems so additional SSS at a different site than the first one was indicated

[#] Clinical deterioration 4 years after SSS due to progressive chronic cord atrophy

[×] Due to FVOO/HCP, underwent ETV after 4 months. HCP persisted, so a VPS was installed after 7 months

[^] Redo FMD/SSS/CSF diversion

Neurological improvement was seen in 12 patients (63%), while five patients (26%) remained stable, and two patients (11%) deteriorated. Surgical morbidity is described in Table 3, while no major postoperative complications occurred; in particular, no catheter dislocation, meningitis, or mortality was observed. Potential losses during intraoperative monitoring

did not occur in any of the cases. On follow-up, the syringomyelia size improved in 16 patients (84%), showed complete remission in two patients (8%), and remained unchanged in one patient (4%) (where the SSS procedure was aborted).

Cerebrospinal fluid diversion procedures

Four patients (8%) with progressive or persistent syringomyelia following FMD underwent six cerebrospinal fluid (CSF) diversion procedures. One patient underwent a cystoperitoneal shunt due to pseudomeningocele, which was thereafter connected to a VPS due to development of hydrocephalus at a later stage. In one patient, an EVD was inserted concomitant to the FMD procedure, which was removed postoperatively. However, 2 months later, a VPS was installed due to recurrent hydrocephalus. One patient underwent VPS due to post FMD hydrocephalus and one patient underwent an ETV due to postoperative FVOO.

Neurological condition after surgery improved in two patients (50%), of which the syringomyelia resolved completely in one patient and remained unchanged in the other. In the other two patients, neurological condition remained stable after surgery, while in one patient the syringomyelia resolved completely and in the other significant shrinkage was seen. One patient, with complete syringomyelia resolvement and improved neurology, developed cerebral ptosis, causing persistent headaches and neck pain; however, further surgical treatment was declined by the patient.

Discussion

Failure of FMD—our philosophy and suggestions

In this study, we retrospectively reviewed 48 patients undergoing FMD for syringomyelia-Chiari complex, emphasizing on the failure rate of FMD for the treatment of CMI-syringomyelia complex. In addition, the different treatment modalities for persistent, progressive, or recurrent syringomyelia after FMD are described. Many studies exist on the efficacy of FMD for syringomyelia treatment, on the factors which might influence the success rate of FMD, and on the success rate of different FMD techniques. However, literature on the different treatment modalities for failed FMD and when failed FMD should be treated is sparse [2, 25, 27, 29]. When FMD seems to fail, alternative causes such as hydrocephalus, insufficient FMD, spinal instability, and others should be excluded and treated. Once other causes have been eliminated, and a sufficient FMD is radiologically documented, the indication for direct syringomyelia treatment at our institution is one of the following: new or progressive neurological symptoms (unrelated to the syringomyelia size), extensive syringomyelia with cord compression (which most probably will lead to neurological symptoms), or syringomyelia progresses over time, even if no or only minimal symptoms are apparent. Our protocol in these cases is to offer the patient an SSS, since the outcome has been proven favorable in our experience [29]. However, clear guidelines and robust data supporting this routine do not exist.

The main limitation of our data consists in its retrospective and descriptive nature, with no direct comparison between the different treatment modalities. The primary FMD procedures completed at our institution are heterogeneous, including both intra- and extradural FMD, which might influence the failure rate of FMD. In addition, the fact that our work combines the results of adult and pediatric patients might also somewhat limit our conclusions. Although our study includes a rather small sample size and therefore might be underpowered, it is to our knowledge the first and largest series describing and analyzing the treatment options of failed FMD and its failure rate. In our series, the failure rate of FMD for syringomyelia treatment was 50%, falling within the range described in the literature (25–50%), although it was still higher than in most published series [2, 10, 31, 34]. A possible explanation might be the fact that we feel comfortable and confident to treat syringomyelia [29]. Nevertheless, we still treat asymptomatic patients or patients with mild symptoms with residual or slightly progressing syringomyelia, conservatively at first. Our routine is to follow these patients closely, both neurologically and radiologically. Only if a clear progression is seen, either in syringomyelia size or in the patient's neurological condition, do we advocate surgical treatment. In some cases, the progression or recurrence occurs more than 5 years after the initial FMD surgery (Table 4). This highlights the fact that patients undergoing FMD for syringomyelia-Chiari complex should routinely be followed for a long period of time. In addition, in some cases, an improvement of the syringomyelia is seen initially after FMD, while on further follow-up a new recurrence or progression is seen (and vice versa). Therefore, even when an improvement of the syringomyelia is apparent following FMD, the patients should be followed regularly for many years. On the other hand, if a progression of the syringomyelia with no clinical symptoms is visible on the first postoperative MRI, immediate intervention is not always indicated, and the patient can be followed closely, as shown in Table 4. A flow chart of the management of syringomyelia-Chiari complex patients with persistent, progressive, or recurrent syringomyelia after FMD is presented in Fig. 1.

There is no clear proven correlation between the patients' age, syringomyelia location, type, size, surgical method of FMD (intra- or extradural FMD, tonsillar resection/shrinkage, etc.), and syringomyelia improvement after FMD [16, 22]. Most studies analyzing these factors are retrospective in nature, consist of very small patient cohorts, and show contradictory results [16, 20, 22, 25]. In our series, the failure rate was rather high when compared to other series. We therefore

Table 4 Selected cases with late progression or improvement of the syringomyelia during follow-up

No.	Treatment group	Initial MRI after FMD	Follow-up MRI 1	Follow-up MRI 2	Clinical symptoms	Plan/treatment
1	Conservative	At 6 months persistent unchanged syringomyelia	At 18 months syringomyelia resolved		None	Continue follow-up
2	Conservative	At 4 months progression of syringomyelia	At 23 months significant improvement of syringomyelia		None	Continue follow-up
3	Conservative	At 6 months progression of syringomyelia	At 67 months* persistent unchanged syringomyelia		None	Continue follow-up
4	Conservative	At 2 months progression of syringomyelia	At 11 months improvement of syringomyelia		None	Continue follow-up
5	SSS	At 10 months improvement of syringomyelia	At 25 months progression of syringomyelia		Ataxia and pyramidal signs	SSS due to progressing and symptomatic syringomyelia
6	SSS	At 4 months improvement of syringomyelia	At 17 months progression of syringomyelia		None	SSS due to significant syringomyelia progression
7	SSS	At 2 months progression of syringomyelia	At 4 months improvement of syringomyelia	At 10 months progression of syringomyelia	None	SSS due to significant syringomyelia progression
8	SSS	At 1 month complete resolution of syringomyelia	At 4 months recurrence of syringomyelia	At 10 months further progression of syringomyelia	Ataxia, motor deficit, and hypoesthesia of fingers	SSS due to progressing and symptomatic syringomyelia
9	SSS	At 1 month complete resolution of syringomyelia	At 6 months recurrence of syringomyelia	At 8 months further progression of syringomyelia	Dysesthesia and hypoesthesia of extremities	SSS due to progressing and symptomatic syringomyelia
10	SSS	Until the 61st month persistent unchanged syringomyelia"	At 61 months progression of syringomyelia		None	SSS due to significant syringomyelia progression

No., number; *FMD*, foramen magnum decompression; *MRI*, magnetic resonance imaging; *SSS*, syringo-subarachnoid shunt

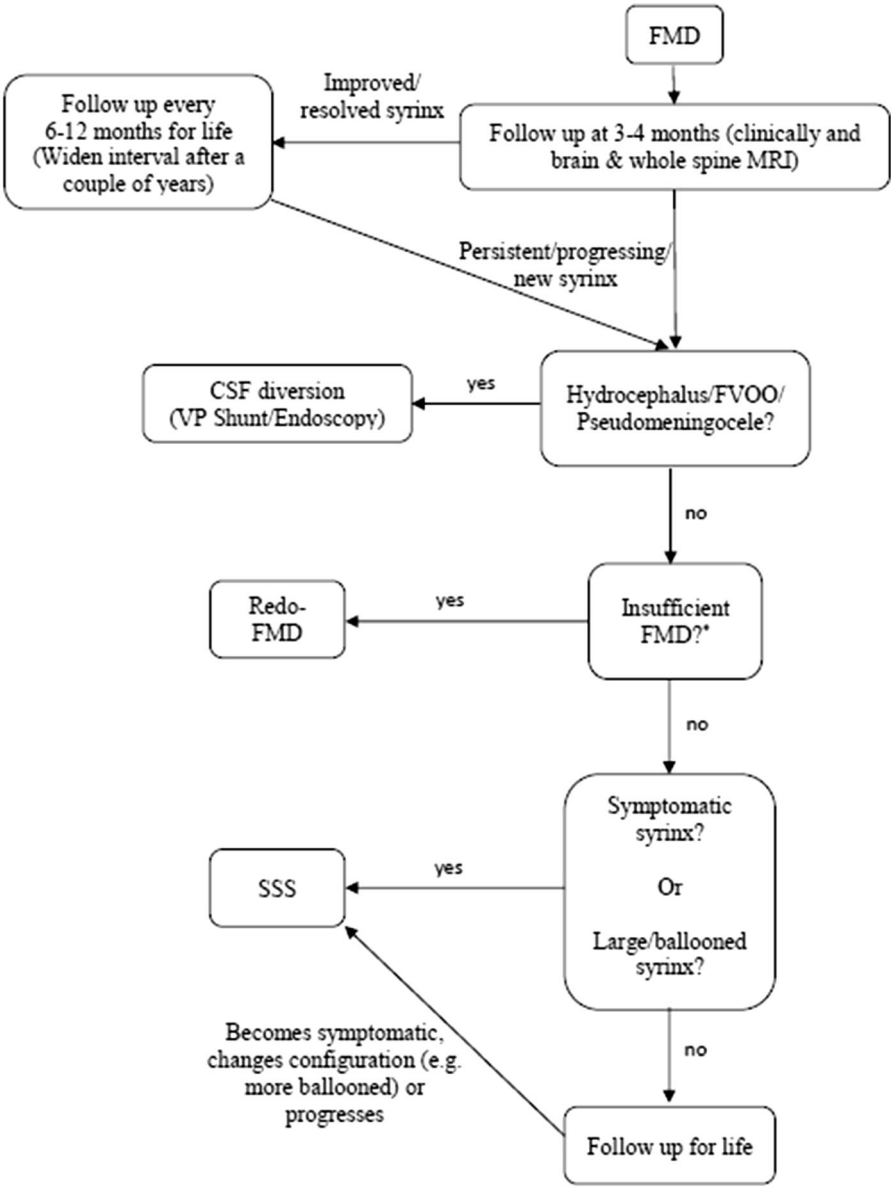
*Underwent 6 clinical and radiological (MRI) follow-ups up to this point

"Underwent 5 clinical and radiological (MRI) follow-ups up to this point

analyzed many technical, surgical, and radiological aspects of our cohort in order to identify factors that might have led to this rather high rate of failure. Of all analyzed factors, none showed a correlation with syringomyelia persistence, progression, or recurrence after FMD, except for the extradural FMD procedure, which showed lower failure rates in multivariate logistic regression analysis (1 failure vs 23 failures in extradural vs intradural FMD). This data however must be analyzed with caution, since our study focus was not to analyze the risk factors leading to failure of FMD, surgical technique was randomly chosen, and since our cohort is rather small and contradictory results have been published in more robust studies with larger patient cohorts [16]. Thus, we cannot assess with certainty the reason for the rather high failure rate of FMD in our cohort. The different surgical techniques and aspects of FMD in CMI-syrinx complex should therefore be the focus of larger studies, as an effort to improve failure rates.

Surgical management of persistent, recurrent, progressing, or new syringomyelia after FMD

FMD failure may occur due to pathologies such as hydrocephalus, cerebellar ptosis, tethered cord, brain tumor, and spinal instability (especially occipito-cervical instability), which must be ruled out through cranial and spinal MRI, and if present, treated accordingly [21, 27, 29]. On the other hand, persistent syringomyelia can be associated with insufficient decompression of the foramen magnum, showing persistent tonsillar herniation impacting the foramen magnum or causing brainstem compression, bone regrowth at the edges of the craniectomy, or complete reclosure of the craniectomy (e.g., in infants or young children), or even occur after a sufficient suboccipital decompression [1, 17, 31]. In these cases, the treatment options include redo FMD with a broader opening of the bone and/or dura expansion (e.g., using a graft), with/without tonsillar shrinking or resection, adhesiolysis at



FMD: foramen magnum decompression; CSF: cerebro-spinal fluid; SSS: syringo-subarachnoid shunt; MRI: magnetic resonance imaging; FVOO: forth ventricle outlet obstruction; VP: ventriculo-peritoneal
*Assessed and defined based on postoperative imaging (CT/MRI)

Fig. 1 The suggested flow chart for the management of patients with persistent, progressive, or recurrent syringomyelia after FMD for syringomyelia-Chiari complex. FMD foramen magnum decompression, CSF cerebrospinal fluid, SSS syringo-subarachnoid shunt, MRI magnetic resonance imaging, FVOO fourth ventricle outlet obstruction, VP ventriculoperitoneal. Asterisk indicates assessed and defined based on postoperative imaging (CT/MRI)

the region of the foramen magnum, syringomyelia shunting procedures (syringopleural shunt, syringo-peritoneal shunt, and SSS), terminal ventriculostomy, or close clinical and radiological follow-ups [2, 7, 10, 17, 27, 28, 32]. Some authors advocate surgical exploration in cases of failed FMD, which is certainly a valid option [2, 14, 25, 27, 31]. None of these studies emphasized the technical aspects or the clinical and radiological outcomes of redo FMD surgery; therefore, firm conclusions cannot be drawn. One could argue that if an extradural approach was initially chosen, a redo FMD with opening of the dura and reduction of the tonsils could be undertaken; however, no clear evidence exists proving the superiority of such an approach [19, 20, 22, 30, 31]. In our series, 80% of the failed patients initially underwent intradural FMD, reinforcing the theory that the syringomyelia may remain persistent irrespective of the surgical FMD technique [12, 16]. Cine flow MRI is discussed controversially in the literature for the assessment of CSF obstruction in Chiari I before and after FMD [6, 15, 24]. Some authors discuss its value to tailor surgical procedures for Chiari I patients, while others recommend it as a postoperative tool for the assessment after performing FMD. In our clinic, due to unavailability, we

have no experience with cine flow MRI for the assessment of insufficient FMD. Therefore, our decision making and analysis of FMD sufficiency is based on standard MRI images.

Very few publications describe and analyze the value and outcome of shunting procedures (e.g. SSS, syringopleural, or syringo-peritoneal shunts) for syringomyelia associated with failed FMD; although a survey published by Schijman et al. showed that in the case of persisting or progressive syringomyelia after FMD, the majority would shunt the syringomyelia [26]. Our group recently published a series of 21 patients undergoing SSS for persistent or progressive syringomyelia after FMD, showing significant clinical and radiological improvements [29]. However, complication rates of up to 16% have been described, with shunt blockage occurring in 5%, on long-term follow-up after syringomyelia shunting [28]. Still, in both our current and previous study [29], complication rates were lower and no major complications or mortality occurred [29]. Postoperative kyphosis at the laminectomy site may occur following syringomyelia shunting procedures, leading to chronic back pain. In our series, two of the three patients that developed kyphosis were managed conservatively, and one required spinal fusion. Further series evaluating the different syringomyelia shunting procedures for the treatment of syringomyelia associated with failed FMD are needed.

Hydrocephalus and/or pseudomeningocele resulting in cerebrospinal fluid (CSF) leak with or without wound or CSF infections is known to occur after FMD [17, 33]. The pathophysiology of hydrocephalus after FMD seems multifactorial. Reports analyzing the underlying reasons for developing hydrocephalus after FMD are rare [33]. To our knowledge, studies looking at the correlation between persistent syringomyelia after FMD and hydrocephalus, and the management of these patients, do not exist. Some reports have shown that in patients presenting with a CMI syringomyelia complex and hydrocephalus the treatment of the hydrocephalus improves the syringomyelia [9, 18]. The traditional causes of postoperative hydrocephalus, such as FVOO and malabsorption of CSF, seem to apply also after FMD. In the case of FVOO, either ETV or VPS provides an adequate treatment. Hydrocephalus secondary to malabsorption is best treated by VPS shunt. If a persistent pseudomeningocele is apparent, one should always rule out hydrocephalus as the cause, especially if a CSF leak occurs. Persisting pseudomeningocele can be treated either with a cystoperitoneal shunt or a VPS. Our results reinforce these recommendations, since all 4 patients with hydrocephalus showed regression or stabilization of the syringomyelia once the hydrocephalus was addressed.

Atlanto-occipital instability is discussed as an additional factor which might influence the outcome of syringomyelia [4, 8]. Basilar invagination, CM 1.5, and CXA under 125° are presumed to be radiological factors significantly increasing the risk of occipito-cervical fusion [4]. Based on our results, basilar invagination and the CXA showed no correlation with FMD failure, while none of the included patients showed CM 1.5. Therefore, it does not seem that in our series atlanto-cervical instability played a role in persisting or progressing syrinx.

Conclusion

Based on our results, FMD failure occurs in about 50% of syringomyelia-Chiari complex patients, indicating a need for subsequent surgical treatment. The management of these patients depends on the suspected underlying pathology. The favored treatment for hydrocephalus occurring after FMD is CSF diversion. Persistent tonsilar herniation after FMD, leading to brainstem compression or impaction of the foramen magnum and failure of syringomyelia improvement, is probably best treated with redo FMD. In all other cases, SSS is a valid treatment option and shows good outcome in experienced hands. Asymptomatic or mildly symptomatic patients with persistent or mildly progressing syringomyelia can be treated conservatively; however, close clinical and radiological follow-ups are warranted. The different surgical techniques of FMD in CMI-syrinx complex should be reevaluated in larger cohorts as an effort to improve failure rates.

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Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

Ethical approval The study was approved by the local ethics committee.

Informed consent Informed consent was waived by the ethics committee due to the retrospective nature of the study.

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