Chapter 10
The Management of Idiopathic and Refractory Syringomyelia

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10.1 Introduction

Tsz-Lu asked to the Master: "As the prince of Wei, sir, has been waiting for you to act for him in his government, what is it your intention to take in hand first?", and the master answered: "One thing of necessity, the rectification of terms" [1]. This sentence from the Analects of Confucius seems perfect to introduce this very controversial topic.

Syringomyelia is a term used to describe a fluid-filled cavity within the spinal cord. This disorder does not usually constitute a disease in its own right, but it is rather a sign of another underlying condition which typically involves an obstruction of the cerebrospinal fluid (CSF) pathways caused by pathologies such as Chiari 1 malformation (CM1), a cranio-cervical junction (CCJ) anomaly, a spinal cord tumor, a spinal degenerative pathology, a posttraumatic, posthemorrhagic or postinfective arachnoiditis, a spinal tethered cord, or a craniosynostosis [2–19].

However, not every fluid-filled cavity in the spinal cord deserves the diagnosis of syringomyelia. For example, cystic spinal cord tumors, glioependymal cysts, myelomalacias, and central cord cavitation (sometimes called hydromyelia) should be distinguished from the syringomyelia [7, 9].

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C. Di Rocco (ed.), *Advances and Technical Standards in Neurosurgery*, Advances and Technical Standards in Neurosurgery 45,
https://doi.org/10.1007/978-3-030-99166-1_10

When the cause of the syringomyelia is identified and correctly treated, the syringomyelia usually regresses and the symptoms improve or do not deteriorate. If the underlying condition is unknown, the syringomyelia is classified as idiopathic.

Until the recent introduction of the magnetic resonance imaging (MRI) as routine diagnostic tool, syringomyelia was diagnosed as idiopathic in the majority of the cases [20]. Nowadays, employing high resolution MRI and a specific sequence such as the cardiac-gated cine-MRI to study spinal CSF flow, areas of flow obstruction, and turbulence which may correspond to circumscribed arachnoid pathologies can be identified [21].

Nonetheless, in clinical practice, nearly 40% of patients presenting with syringomyelia will be still diagnosed as having an idiopathic syringomyelia (IS) [21]. In the pediatric population, about 50% of children diagnosed with IS present with a concomitant scoliosis [22]. Some authors have reported cases of IS with neurological symptoms and signs consistent with CM-1 [6], but without cerebellar tonsillar descent where the foramen magnum decompression improved patients' clinical symptoms and syringomyelia radiological appearance; hence they suggested to define the condition as Chiari 0 malformation [10, 22].

The topic is controversial, vast, and confused. This is mainly due to the heterogeneity of the underlying conditions, the interchanged terminology of syringomyelia with hydromyelia often used in the publications, the different methods employed to determine when the condition is idiopathic, the treatment options proposed, and the outcome measures.

Even when the syringomyelia has a defined cause and the main condition is treated, like in patients with CM-1, the syringomyelia may fail to resolve, can deteriorate or recur after an initial shrinkage in up to 66% of the cases [20, 23, 24]. We define such syringomyelia as refractory syringomyelia.

This chapter will first try to clarify when a syringomyelia can be truly defined as idiopathic and subsequently it will focus on the management of syringomyelia refractory to previous surgery, proposing an algorithm of treatment for both these challenging clinic-radiological conditions. In the final part, a description of the neurosurgical armamentarium available to manage the idiopathic and refractory syringomyelia is described.

10.2 Idiopathic Syringomyelia

Considerable debate exists in the literature regarding the terms utilized to define this condition.

Holly and Batzdorf argued that a slit-like syrinx is not a true syringomyelia and is also different from a pre-syrinx state [25]. However, in the literature, there is no uniformity regarding this opinion. For example, Roser et al. consider hydromyelia only a dilated central canal and affirm that IS presents with different clinical and radiological signs [12]. The 34 patients with hydromyelia in their study had no neurological deficits and presented with pain that could be radicular, burning, or

musculoskeletal. They further distinguish hydromyelia as a congenital condition and differentiated it from IS [26].

Batzdorf makes a clear distinction between syringomyelia and a persistent central canal. The latter is characterized by linear or fusiform appearance on sagittal MRI scans, is between 2 and 4 mm in maximal width, and almost perfectly round, centrally placed in the cord on axial MRI scans obtained through these areas [27].

Analyzing the available literature, it is really challenging to differentiate between slit-like syringomyelia, hydromyelia, and IS and understand if they are different entities or simply a continuum on a spectrum. A deep pathophysiological review is beyond the scope of this chapter, and it would be enough to mention scenarios such as the case of a patient who had undergone shock wave lithotripsy for renal calculi and developed arachnoid scarring 7 years later near the level of the affected kidney [21], or the cases of a slit-like syringomyelia which represent the late phase of a spontaneous resolution of a preexistent bigger syrinx [15] to demonstrate the complexity of this condition.

A particularly important first distinction which should and is possible to be made is between a central canal dilation and a syringomyelia (Fig. 10.1a, b).

We have found six useful criteria to distinguish between a central canal dilatation and a syringomyelia which can assist the clinician in distinguishing between these two entities.

The central canal dilatation usually presents the following features: (1) lower cervical and/or mid-thoracic cord location, (2) no space-occupying effect, (3) no change or regression on follow-up MRIs, (4) spindle or slit like shape suggestive of lower intracavitary pressure, (5) no flow signal inside the dilatation and no obstruction of the CSF flow around it on the cine-MRI, (6) no correlation between location of the cord dilatation and patient's clinical symptoms [25, 26, 28]. The management

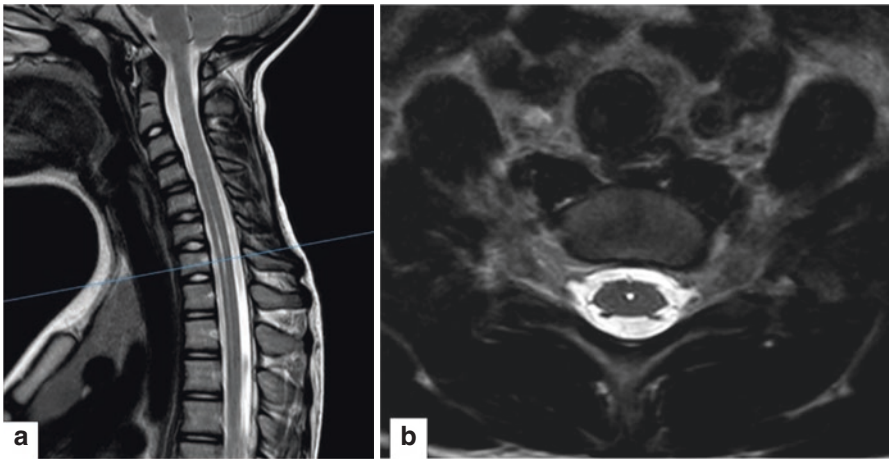

Fig. 10.1 MRI sagittal (**a**) and axial (**b**) T2 sequences of a typical central canal dilation

of central cord dilatation is nonsurgical; patients should be reassured about the diagnosis and ensure that a "label" of syringomyelia is not attached to their notes.

Once this first distinction has been made, in order to succeed in the treatment of a patient with a "true" syringomyelia it is essential to try our best efforts to disclose the underlying causative mechanism. This can be done in our opinion in the majority of cases where there are enough health care resources.

In a patient presenting with a syringomyelia, the following diagnostic algorithm should be followed

1. MRI of the head and the entire spine, including gadolinium enhancement.
2. Cardiac-gated phase-contrast CSF flow studies of the cranio-cervical junction and the cervical, thoracic, and lumbar spine in the median sagittal plane for the visualization of craniocaudal CSF flow. Cardiac-gated phase-contrast CSF flow studies are also known as fast imaging employing steady state acquisition (FIESTA), a technique in which the MRI signal acquisition is synchronized to the cardiac and/or respiratory cycle. This is a noninvasive investigation which has been proved to be more reliable compared to the invasive conventional myelography [21].
3. Serum and CSF samples' analysis to rule out the presence of demyelinating diseases, immunological disorders, vasculitis, infection, and vitamin deficits.

Once the available technology and laboratory tests have been employed without disclosing the cause of the syringomyelia, this can be defined as IS and managed according to the algorithm proposed (Fig. 10.2). In these cases of IS, usually the commonest cause remains a thoracic arachnopathy [7–9, 29, 30].

At this point, it is crucial to distinguish between asymptomatic and symptomatic IS patients. The former should be always managed conservatively and followed up clinically with or without radiological investigations in selected cases. For instance, in case of an oval-shaped three-level cervical syringomyelia, previous reports of isolated cervical IS hiding low-grade tumors (especially ependymomas non-enhancing after contrast MRI) which then progressed into a high-grade lesion should always serve as lesson, and long-term clinical and MRI follow-up are recommended for these syringes [31]. Nonspecific and manageable pain, sensory impairment, and a relatively stable small syrinx are often managed conservatively [21]. There is evidence to prove that children with IS remain asymptomatic, stable, or improve in over 90% of cases and, radiologically, the majority of their syringes (87.5%) remain stable or shrank over time, with no clear correlation between changes in size and changes in symptoms [32]. These patients should be managed nonsurgically with parental reassurance about the condition.

The situation becomes a challenge when these patients become symptomatic or present with neurological deterioration. In these cases, the severity and the progression of the symptoms should guide the treatment. Surgical exploration is recommended in patients with IS and progressive neurological deficits [12].

Several procedures have been proposed to treat the IS ranging from the insertion of shunt into the cavity to drain it into the subarachnoid space, pleura, or peritoneum till the direct arachnidolysis and duroplasty [33].

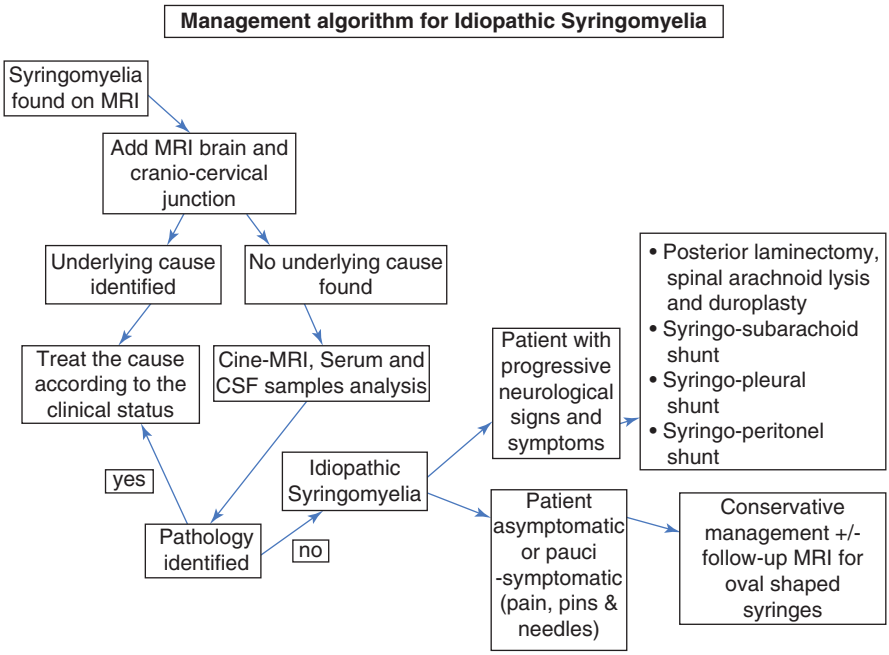

Fig. 10.2 Management algorithm in patients with idiopathic syringomyelia (IS)

Reaffirming that syringomyelia is always an epiphenomenon of an underlying pathology, any surgical intervention should always be directed to resolve the underlying causative mechanism. In presence of a true IS, after a rigorous workup, this usually means unblocking CSF pathway obstruction in the thoracic spinal canal, reserving shunting techniques to nonresponsive cases.

In the authors' experience, when the radiological workout hasn't been helpful in disclosing a precise location of the CSF obstruction, a "rule of thumb" is to perform a posterior decompression and arachnoidolysis at the spinal level (usually thoracic) where a turbulence inside the syringomyelia is visible on the MRI T2 sagittal sequences. An example of this scenario is depicted in Fig. 10.3a, b.

Posterior arachnolysis and duroplasty have shown better long-term outcomes compared to syrinx-shunting procedures in the treatment of an arachnoid pathology which as we have explained above is the cause of the IS in the majority of cases [7].

10.3 Refractory Syringomyelia

We define refractory a syringomyelia that remains persistent on the postoperative follow-up MRI scans despite surgical treatment or recurs after an initial response to the surgical management.

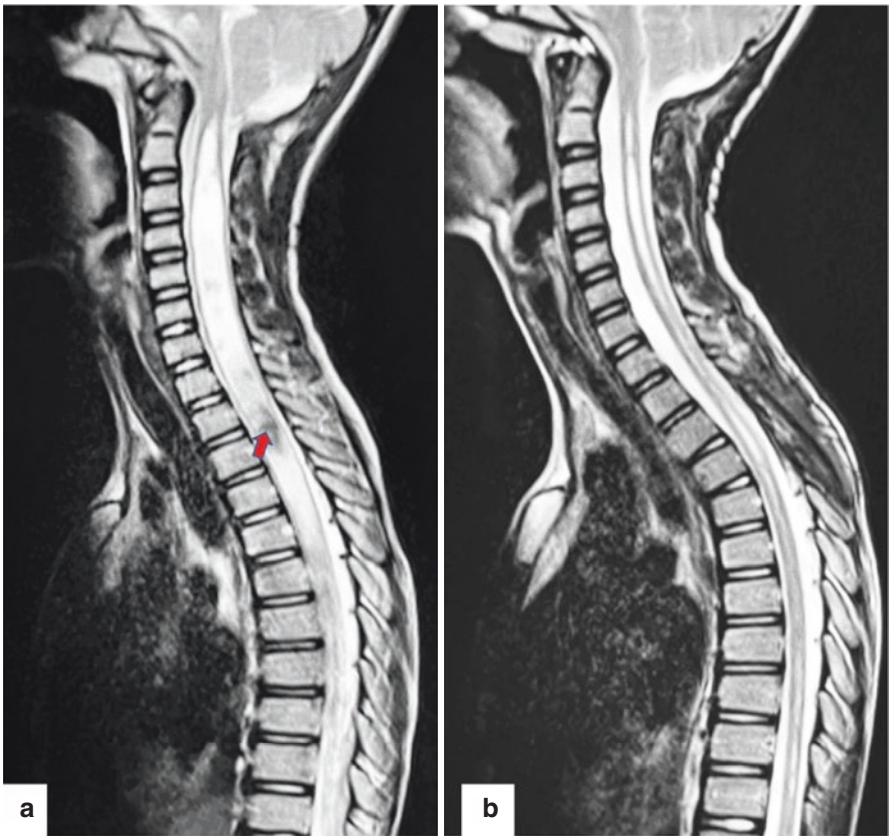

Fig. 10.3 10 years old boy with IS where a full laboratory and radiological workout failed to disclose the underlying cause of the syringomyelia or a precise location of the CSF obstruction. The patient was initially followed up, but after 1 year he experienced a clinical and radiological progression of the syrinx; hence a posterior decompression and arachnoidolysis was performed at the spinal level T3 where a turbulence on the MRI was visible inside the syringomyelia. (**a**) Preoperative MRI sagittal-T2 sequence showing the holocord syrinx and the turbulence inside it (red arrow); (**b**) postoperative MRI sagittal-T2 sequence showing the syrinx decompression 3 months after the T3–4 laminectomy and posterior arachnoidolysis

The reasons for persistent syringomyelia are dependent on various factors. The primary etiology that contributes to the syringomyelia falls into one of these: i.e., Chiari Malformation 1 (CM1), cranio-cervical junction (CCJ) anomalies, spinal dysraphisms, spinal tumor, spinal degenerative diseases, posttraumatic, posthemorrhagic, and postinfective arachnoiditis, hydrocephalus, and craniosynostosis. Depending on the initial causative mechanism of the syringomyelia, the initial surgical technique employed, and the clinical symptomatology of the patient when the refractory syringomyelia is diagnosed, the management will vary.

10.3.1 Chiari 1 Malformation (CM-1)

In CM-1 patients, improvement of syringomyelia post-foramen magnum decompression (FMD) usually occurs within 3 months and continues thereafter, with most patients experiencing syrinx reduction in the first postoperative year [16]. Nonetheless, persistent, progressive, or recurrent syringomyelia following FMD for CM1 is described in up to 66% of the cases in long-term follow-ups [7, 8].

In case of FMD for CM1, the CSF flow through foramen magnum is hindered by intrinsic and extrinsic factors. Intrinsic factors are usually secondary to the obstruction of the foramen of Magendie due to tonsils overlying the foramen of Magendie or posterior inferior cerebellar artery (PICA) and/or arachnoid veils obstructing it [34]. On the other hand, the extrinsic factors include pseudomeningocele formation with adhesions in the foramen magnum and inter-tonsillar adhesions, primary insufficient decompression of the foramen magnum resulting in persistent tonsillar herniation impacting the foramen magnum or causing brainstem compression, bone regrowth at the edges of the previous craniectomy or complete reclosure of the craniectomy (e.g., in infants or young children), and scar formation (and arachnoiditis) after a sufficient suboccipital decompression [35]. According to Tosi et al. in case of small or incomplete decompression or reclosure of the craniectomy, an enlargement of the previous decompression reduced the syrinx size in 15 of 16 patients [16].

In case of refractory syringomyelia post CM-1 where a satisfactory decompression has been performed, the first thing to consider is if any arachnoid veil has been possibly left behind or if a postsurgical scar has formed and is now the causative mechanism sustaining the syringomyelia. The latter is usually the case when the syringomyelia recurs after an initial shrinkage. In these circumstances, a reexploration of the previous cranio-cervical decompression with arachnoid lysis and enlarged duroplasty is recommended [13, 36, 37]. If an enlarged watertight duroplasty was not performed during the first operation, we strongly recommend, in case of RS, to reexplore the previous decompression and perform it. In the authors' experience, this is an effective remedy to resolve the syringomyelia [38, 39]. An example of a similar scenarios is shown in Fig. 10.4a–f. We have previously described our surgical technique regarding performing an enlarged watertight duroplasty [38].

If the cranio-cervical decompression seems appropriate on post-op MRI and cine-MRI study and/or the cranio-cervical junction has already been reexplored with no success and the patient is symptomatic or with progressive neurological symptoms, a direct exploration of the arachnoid surrounding the syrinx, a syrinx fenestration, and/or syringo-shunting technique can be considered.

Posterior arachnolysis and duroplasty, in general, have a better long-term outcome compared to syrinx-shunting procedures in the treatment of a localized arachnoid pathology [7–9, 30]. Nevertheless, it should be acknowledged that it is not often possible to predict the level of CSF blockage on the basis of the site, size, and shape of syringomyelia [21].

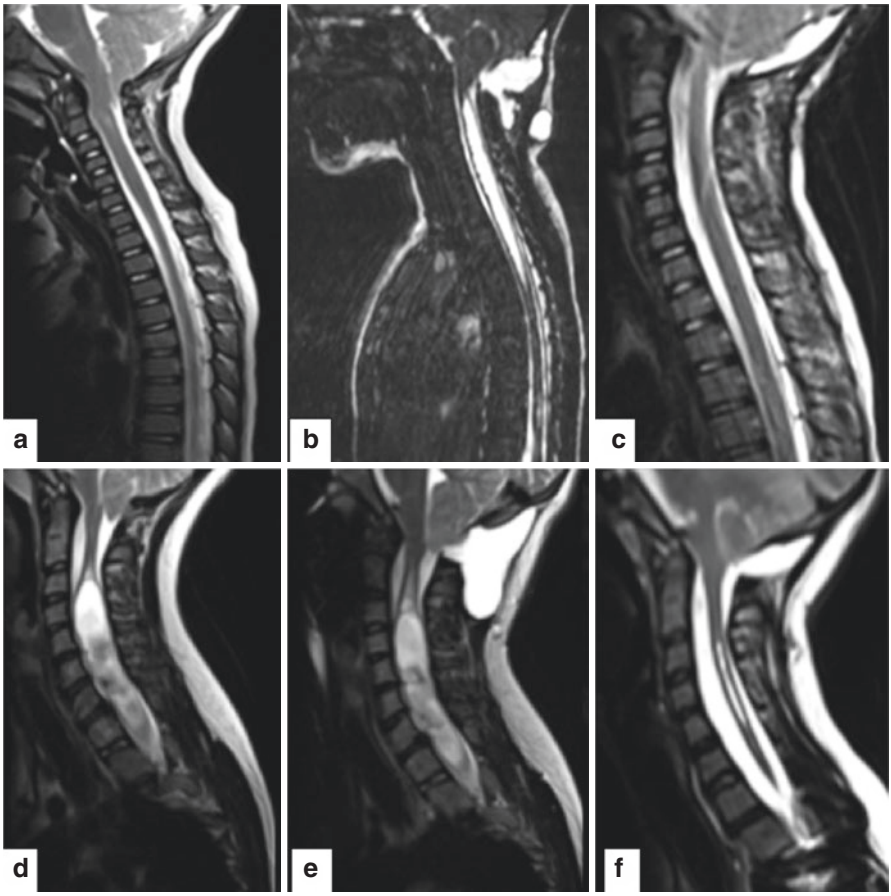

Fig. 10.4 (**a**) Preoperative MRI sagittal T2 sequence of a patient with CM-1 undergone foramen magnum decompression and durotomy without duroplasty; (**b**) postoperative MRI sagittal T2 2 months after the initial operation showing a large pseudomeningocele and a de-novo syringomyelia; (**c**) MRI sagittal T2 sequence of the same patient 3 months after reexploration of the FMD, arachnoidolysis, and enlarged watertight duroplasty showing a complete resolution of the syringomyelia; (**d**) preoperative MRI sagittal T2 sequence of another patient with CM-1 and preoperative cervico-thoracic syringomylia undergone foramen magnum decompression and durotomy without duroplasty; (**e**) postoperative MRI sagittal T2 3 months after the initial operation showing a large pseudomeningocele and a further upward enlargement of the previous syringomyelia; (**f**) MRI sagittal T2 sequence of the same patient 6 months after reexploration of the FMD, arachnoidolysis, and enlarged watertight duroplasty showing a remarkable improvement of the syringomyelia

In terms of shunting options, such as syringo-subarachnoid shunt (SSS), syringo-pleural shunt, and syringo-peritoneal shunt [11, 33, 36–42], Soleman et al. have shown that SSS has a favorable clinical and radiological outcome [43] and in our opinion should be preferred to the other shunting techniques.

10.3.2 Cranio-Cervical Junction (CVJ) Anomalies

The pathophysiology of syringomyelia combined with CVJ anomalies is only partially understood and its treatment is still a matter of debate [18, 44–46]. The most common form of cranio-cervical pathology associated to syringomyelia is foramen magnum arachnoiditis [8] which can usually be treated successfully with posterior fossa decompression, arachnoidolysis, and expansile duroplasty. The second most common cause is basilar invagination/atlanto-axial dislocation (BA/AAD). In this case, several theories have been proposed to explain the pathogenesis of syringomyelia [5] which remains still unclear.

The main factors thought to be responsible are (1) ventral compression by dislocated odontoid process, (2) dorsal compression due to a small posterior fossa, and (3) concomitant Klippel–Feil syndrome and occipitalization of atlas [47, 48].

Several surgical techniques have been proposed to treat BA/AAD [44, 46], but still disagreement exists on what represents the best way to manage the condition [46]. In the last 20 years, there has been a shift toward posterior approaches (including direct posterior reduction and fixation—C0–C2, C0–C3 or C1–C2—plus or less decompression), leaving the ventral transoral odentoidectomy (which nowadays can be also performed endoscopically) only to cases of unsolved ventral compressions without clinical improvement [45].

Untreated CVJ anomalies are usually one of the causes of persistent syringomyelia in patients who previously underwent foramen magnum decompression (FMD) for CM-1 and the CVJ pathology was missed or left untreated. Addressing the cranio-cervical instability is of paramount importance to manage the syringomyelia in these patients.

10.3.3 Spinal Dysraphisms

Syringomyelia is reported in the various types of spinal dysraphism ranging from 21 to 67% of the patients [6, 49]; myelomeningocele and split cord malformation are the most common disorders associated with syringomyelia [50]. Several hypotheses exist regarding the pathophysiology of this syringomyelia.

Greiz has speculated that tensile radial stress on the spinal cord may cause syrinx in spinal dysraphisms, as the transient lower pressure of the cord parenchyma may draw in interstitial fluid causing enlargement of the syrinx [5]. Lee et al. showed how producing epidural compression using kaolin material in a rat model caused a syrinx cranially to the compression in the animals, speculating that compression of the cord related to spinal dysraphism with cord tethering is sufficient to cause the syringomyelia [50].

The presence of syrinx in an asymptomatic patient with spinal dysraphism does not represent a reason per se to recommend a surgical intervention, especially if the size of the syrinx is not extensive [51].

An increase in size of the syringomyelia, instead, could represent a progression of the cord tethering and usually warrants a surgical untethering [52]. The syrinx status is also an important factor to know during the postoperative follow-up. Although an improvement of the syrinx in the postoperative period is a sign that untethering was successful, an unchanged syrinx does not represent a failure of the treatment as several reports have shown that symptomatic improvement is frequently seen in patients in whom the syrinx was unchanged [51].

On the other hand, a syrinx deterioration is an important radiological sign of retethering (Fig. 10.5a–d) and sometimes can be useful to anticipate symptomatic deterioration in the patient [50].

There has been debate in the past regarding whether direct drainage of the syrinx was needed in these cases [53]; however, the recent evidence suggests that the best strategy is, as in all cases of syringomyelia, to treat the main cause and hence perform a spinal cord re-untethering under intraoperative neuromonitoring (IOM) [14, 54].

Finally, worthy of note is a worsening syringomyelia in a patient with myelomeningocele and a ventricular shunt due to hydrocephalus. Shunt malfunction should always be ruled out as it is the commonest cause of the increase in size of the syringomyelia (Fig. 10.6a–d).

10.3.4 Spinal Tumors

Intradural extramedullary tumors (such as meningiomas, schwannomas), intradural arachnoid cysts, or most commonly, intramedullary tumors can present with syringomyelia [55–57]. This association is known at least since 1875 [58], and in a postmortem study of 209 patients with an intramedullary tumor, a syringomyelia cavity was found in 31% of the cases [57]. Ependymomas, angioblastomas, and astrocytomas are the commonest to show an association with syringomyelia [8].

However, the pathogenesis of this kind of syringomyelia is still debated; several theories have been proposed which can be summarized in five main groups: (1) the syringomyelia cavity is part of the tumor, (2) the syrinx arises from the stasis of tissue fluid resulting from occlusion of the normal drainage pathways, (3) spread of edema and spontaneous autolysis of the tumor itself or hemorrhage from the tumor into the cord, (4) altered perimedullary CSF pathway from the extrinsic compression of the tumor resulting in the intramedullary cavity, and (5) exudation caused by disruption of the blood–brain barrier [59]. In the majority of cases, a combination of the above mechanisms is likely the driver for the syringomyelia formation, according to the location and the etiology of the tumor. For instance, intramedullary ependymomas present with syringomyelia in half of the cases [60].

Surgical excision of the tumor usually improves or resolves the syringomyelia and a residual syringomyelia after tumor resection does not usually require any additional surgical treatment. Occasionally, the syringomyelia can deteriorate, in case of incomplete tumor resection and regrowth, following adjuvant treatment,

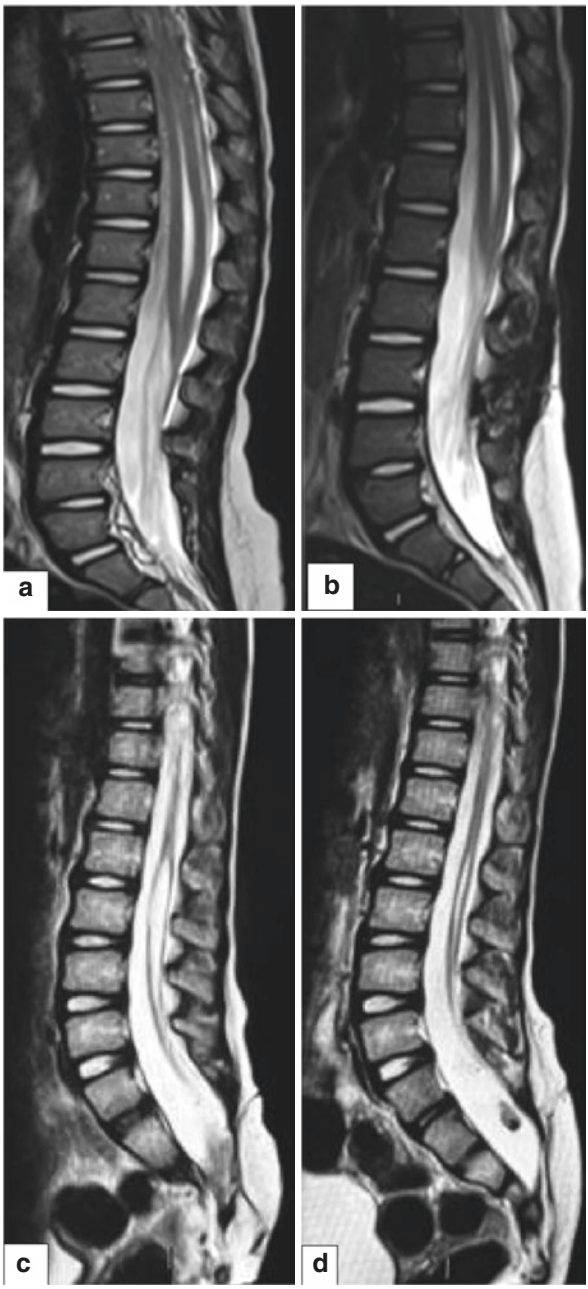

Fig. 10.5 (**a**) MRI sagittal T2 sequence showing a re-tethering of a previously (7 years before) untethered fatty filum with syringomyelia; (**b**) MRI sagittal T2 sequence of the same patient 6 months after re-untethering of the spinal cord showing improvement of the syringomyelia; (**c**) MRI sagittal T2 sequence of a 5-year-old girl with spinal lipoma previously partially untethered (4 years before) showing a de novo syringomyelia onset as first sign of cord re-tethering; (**d**) MRI sagittal T2 sequence 6 months after re-untethering and lipoma resection under IOM showing remarkable improvement of the syringomyelia

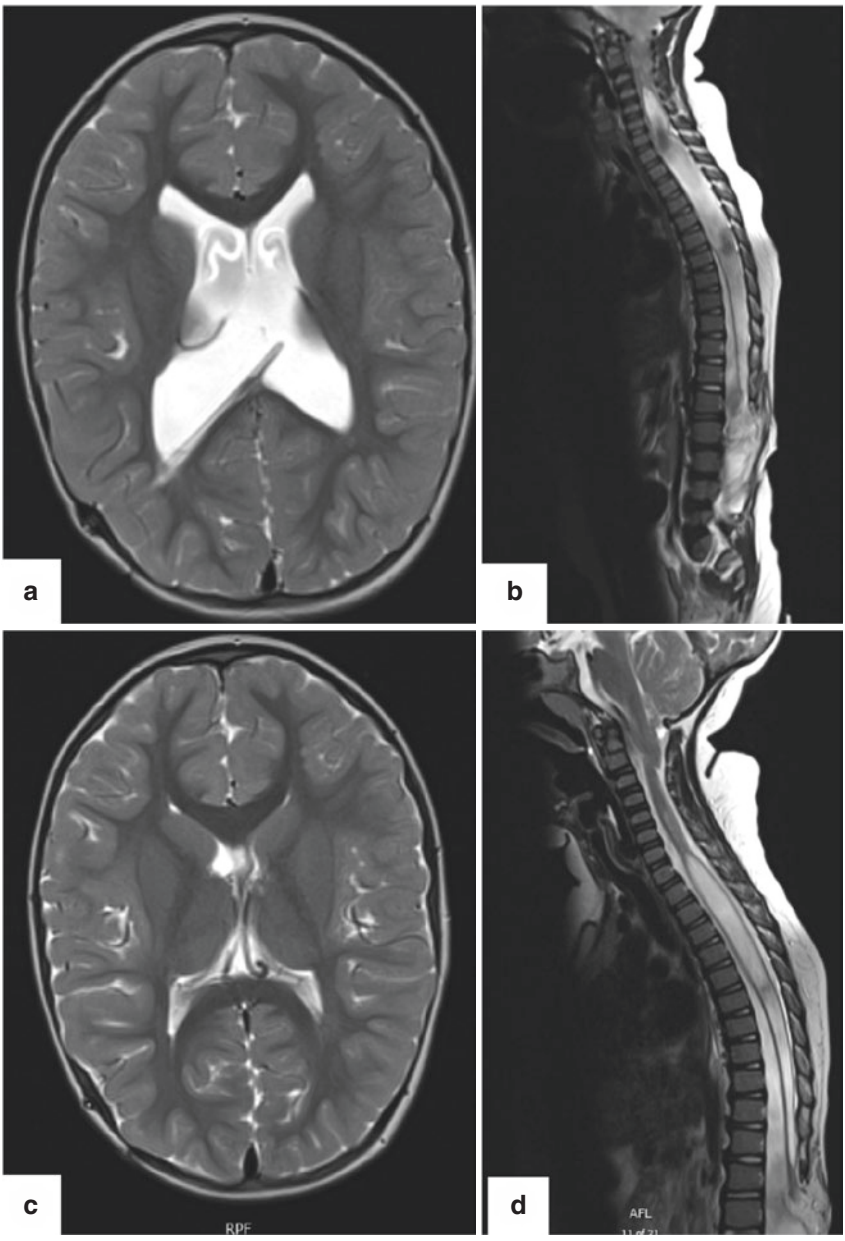

Fig. 10.6 (**a**) MRI head axial T2 sequence of a patient with spina bifida, Chiari 2 malformation, and ventricular enlargement secondary to VP shunt malfunction; (**b**) MRI whole spine sagittal T2 sequence of the same patient showing a holocord syringomyelia at the time of the VP shunt malfunction; (**c**) MRI head axial T2 sequence 3 weeks after shunt revision showing reduced ventricles size; and (**d**) MRI spine sagittal T2 sequence showing reduction in size of the previous holocord syringomyelia

including chemotherapy and radiotherapy, or when there is a disseminated spinal tumor (tumoral leptomeningitis) not amenable of surgical treatment. In these cases, if the syringomyelia is the cause of a severe progressive neurological deterioration, and according to the oncological prognosis, direct fenestration of the syrinx or shunting of the syrinx is recommended.

The management is purely on a case-by-case basis and preferably performed in centers that have the appropriate expertise with facility for long-term oncological follow-up.

10.3.5 Trauma/Hemorrhage/Infection

We have complied this in one group as the etiology of RS in this cohort is primarily due to arachnoidal adhesions. The adhesions may sometimes be very extensive posing a challenge in management.

Phase-contrast cine MRI may accurately localize subarachnoid space obstruction and demonstrate normalization of CSF flow after surgery, and it may also be used to confirm spinal cord tethering and communication of spinal cord cysts with the subarachnoid space [4]. Surgical intervention should be considered only in patients with progressive debilitating neurological deficits. A number of surgical strategies have been suggested in the literature including cyst aspiration, fenestration and shunt placement, and intradural exploration and duraplasty. Many recommend intradural exploration, lysis of subarachnoid adhesions, and duraplasty as the preferred primary treatment options for this kind of syringomyelia in case of neurological deterioration. The goal of this intervention is to create a channel for CSF circulation extrinsic to the spinal cord itself. By creating this pathway, CSF is no longer forced into the parenchyma of the cord, thereby allowing the syrinx to collapse. In cases where this technique is not possible or it has failed, shunting of the syrinx may be considered. Syringo-subarachnoid, syringo-peritoneal, and syringopleural shunts have been described. In all cases, septations in the syrinx need to be lysed to allow an optimal decompression. Syrinx shunts, however, have a propensity to fail, which can limit their usefulness. Cyst aspiration alone is a palliative procedure, and as such, has limited usefulness in this patient population [3].

In posttraumatic syringomyelia, reconstruction of the subarachnoid space by arachnoidolysis and untethering the cord usually allows to improve or stabilize the vast majority of patients [2].

The adhesions can be quite diffuse in case of infection and the primary aim should be based on management of infection as initial step followed by surgery. SSS can be resorted to if arachnoidolysis fails. There can be associated hydrocephalus post-initial surgery and this may contribute to CSF malabsorption. In such cases, endoscopic third ventriculostomy or ventriculoperitoneal shunt can be utilized to manage the hydrocephalus and this may help the resolution of the syringomyelia.

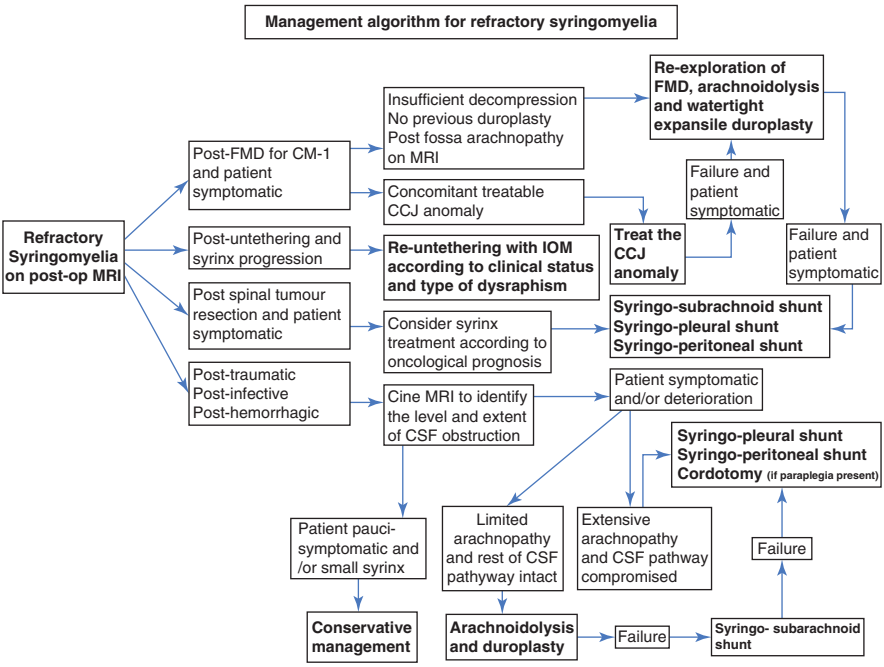

Fig. 10.7 Management algorithm in patients with refractory syringomyelia (IS)

Arachnoidolysis, untethering, and duroplasty provide good long-term results for focal arachnopathies. Nonetheless, for extensive pathologies with a history of subarachnoid hemorrhage or meningitis, treatment remains a major challenge [8].

Shunt placement should be reserved to these cases of diffuse arachnoiditis or with recurrences on the basis of their short-term effect and the possibility to reoperate in case of failure [29, 30]. Overall, improvement after shunt placement, regardless of modality and circumstances (syringo-subarachnoid, syringo-peritoneal or syringo-pleural shunts, primary operation or recurrence), accounts for 60% of cases with a recurrence rate of 50% [30].

Patients with a complete spinal cord lesion can be treated with cordectomy which has proved to be a very effective form of treatment for syringomyelia [9]. However, accepting this kind of operation is not easy for patients and the psychological burden should be considered when proposing such option.

An algorithm for the management of recurrent syringomyelia is outlined in Fig. 10.7.

10.4 Surgical Techniques

We describe below the neurosurgical armamentarium employed in patients with IS and RS. The most common surgical procedures are described below:

10.4.1 Reexploration of Posterior Fossa, Arachnoidolysis, and Enlarged Duroplasty

Under general anesthesia, the patient is positioned prone on Mayfield 3 pins headrest. Previous midline scar is explored under microscopic guidance to carefully dissect the soft tissue layers and reach the dura. Subsequently, the dura is carefully opened to inspect the foramen magnum and to explore for patency of the foramen of Magendie to ensure the presence of CSF egress. Tonsillar coagulation may be necessary to accomplish this during the primary procedure. In case of adhesions, careful arachnoidolysis is performed. As previously discussed, careful consideration should be given to possible obstruction of the Foramen of Magendie due to tonsils overlying the Foramen of Magendie, posterior inferior cerebellar artery (PICA) obstructing the foramen of Magendie, veil obstructing the Foramen of Magendie, and pseudomeningocele formation with adhesions [34, 61]. Expansile duroplasty should be performed following reexploration due to high chance of adhesions due to bleeding and by the procedure per se. McGirt et al. showed in their series that by performing duraplasty during posterior fossa decompression decreased the likelihood of reoperation; 72% of their patients underwent duraplasty and 70% had improvement in their syrinx [62]. We have successfully used Duraguard (bovine pericardium dura) in these cases to perform an enlarged watertight duroplasty with running prolene 5/0 or 6/0 [38, 39].

10.4.2 Spinal Laminectomy, Arachnoidolysis, and Duroplasty

Decompression procedures of the spinal subarachnoid space may have beneficial effects unless the patient shows longitudinally extensive arachnoiditis (such as in case of meningitis).

Under general anesthesia and with the use of intraoperative neuromonitoring (IOM) [54], the patient is placed prone and a laminectomy is performed at the levels involved with arachnoid scarring (previously visualized on FIESTA MRI). We suggest the use of intraoperative ultrasound to identify the ideal place where to start the dura opening under the microscope; this helps in avoiding direct injury to the spinal cord which in some cases can be directly adherent to the overlying dura. Once the dura is opened and separated from the arachnoid, the arachnoid and the scar are dissected away from the pial surface of the cord using sharp dissection in order to untether the spinal cord and reestablish free CSF flow.

Following these manoeuvres, the normal pulsations of the spinal cord, which are usually attenuated or absent at the level of the scar, reappear and occasionally the syrinx visibly collapses [29, 30]. With extensive arachnoid scarring, the arachnoid appears thicker, denser, and whiter and, occasionally vascularized obscuring the underlying spinal cord. In these cases, to prevent any spinal cord injury, the arachnoid dissection should be limited to the dorsal surface of the spinal cord. The

duroplasty can be performed either with autologous fascia lata or lyophilized dura [8, 9]. The authors have successfully used bovine pericardium dura (Duraguard) in these cases.

10.4.3 Syringo-Subarachnoid Shunt (SSS)

Among the shunting technique, SSS is usually the first choice when there is normal CSF flow in the subarachnoid space; it should not be used in cases of severe arachnoiditis.

Under general anesthesia, the patient is positioned prone. IOM is recommended for this procedure as this can alert the surgeon to potential intraoperative threats to the functional integrity of the spinal cord [23, 54]. After skin disinfection and draping, a midline skin incision is completed over the previously identified spinal levels. The muscle fascia is opened, and the muscles are dissected exposing the desired spinal processes and laminae. A standard laminectomy of one or two laminae is performed. Under microscopic magnification, the dura is incised in the midline and suspended on each side. We recommend the use of a fine ultrasound probe prior to opening the arachnoid and performing the myelotomy to confirm the most dilated location of the syrinx [14] and decide where to define the subarachnoid pocket (rostral or caudal in line with the syrinx). An appropriate length of patent subarachnoid space is needed to allow the inline placement of the catheter. The arachnoid is opened and a 5–6 cm lumboperitoneal shunt (Medtronic, Minneapolis, MN, USA) is inserted into the caudal or cranial subarachnoid space for 3–4 cm distally, while the proximal side of the shunt catheter is put between the dural retraction sutures in the meantime the myelotomy is performed [24, 63].

It is crucial to ensure that the tip is actually subarachnoid (and not just subdural), otherwise it will not function. A midline myelotomy is usually performed. Sometimes it might be difficult to define the midline (posterior median sulcus), especially if the cord is rotated, or if significantly compressed by the syrinx. In these cases, the small pial arteries folding medially towards the central canal should be sought and recognized, since they point out towards the posterior median sulcus. Another tip to identify the midline is to expose the nerve roots bilaterally and the posterior midline sulcus is situated equidistant from both nerve roots. Alternatively, if the syringomyelia is bulging laterally, the myelotomy can be safely performed in the dorsal entry zone (DREZ) between the lateral and posterior columns because this is the thinnest part especially when there is already an upper extremity proprioceptive deficit caused from the syrinx [33].

If the midline is defined, the superficial pial vessels are cauterized and the pia is incised using a diamond knife. With the help of a non-tooth forceps, the cord is split until the syrinx is reached. When the cord is under high pressure, the opening of the syrinx is completed before inserting the tip of the shunt into the subarachnoid space, to avoid a possible injury to the cord tissue [63]. Following syrinx decompression,

the proximal tip of the shunt is inserted into the syrinx cavity for 2–3 cm cranially or caudally. A 6–0 prolene suture is used to fix the shunt to the arachnoid to avoid dislocation. The dura is closed in a watertight fashion using a running suture 5–0 prolene. In case of laminoplasty, the laminae are fixed using plates and screws. The wound is closed in layers.

10.4.4 Syringo-Pleural Shunt (SPS)

Syringo-pleural shunting is safe and a straightforward technique with good results both clinically and radiologically. The result is to be expected within the first few weeks and may be long lasting [64]. Under general anesthesia, the patient is positioned prone. A laminectomy or a laminotomy of 1–2 laminae is performed, preferably below the T1 level. The best level of shunt insertion remains controversial: the advantage to place it under the T1 level is the avoidance of neurological injury of the upper extremities; however, it should be considered that the dorsal columns of the thoracic cord are extremely fragile, and the "non-gentle" manipulation may have a higher chance of causing neurological symptoms of the lower extremities [64, 65].

Different types of shunt can be used such as the 1-piece Spetzler lumboperitoneal shunt (Integra Neurosciences, Plainsboro, NJ), with a 0.7-mm inner diameter, 1.5-mm outer diameter, and 31.5-inch length; the lumbar end of this catheter, with multiple perforations, is placed inside the syrinx, and the opposite end is positioned in the pleural space; lumboperitoneal shunt catheter (Medtronic, Minneapolis, MN, USA) or a T- or Y-shaped catheter or antibiotic impregnated catheter can also be utilized for the SPS [42, 64–66]. A distal slit valve to avoid over drainage of the syrinx into the pleural cavity should be considered.

The patient is placed in the prone position and after radiographic confirmation, a midline mid-thoracic incision is made. A single-level laminectomy followed by durotomy is performed. Syrinx location and spinal cord thickness are verified with the help of intraoperative ultrasound. A standard dorsal midline or dorsal root entry zone (DREZ) myelotomy is performed, and the catheter is inserted, under microscopic magnification, and directed superiorly for approximately 4–5 cm through a small midline myelotomy. The use of IOM is recommended to perform this surgery. Somatosensory potential (SSEPs) is often lost during a midline myelotomy; if motor-evoked potentials (MEPs) drop significantly or disappear and/or do not recover after a short period of time, the catheter should be repositioned [54]. The catheter is sutured to the dura and tunnelled toward the separate paramedian incision, previously made at the level of the fifth intercostal space 5 on the right side. This incision is taken down to the superior margin of the sixth rib, and the parietal pleura is entered via a stab incision just over the superior margin of the rib. The distal end of the catheter is inserted into the pleural space. Two anchoring sutures are placed to the paraspinal and intercostal muscles to prevent catheter migration [34].

10.4.5 Syringo-Peritoneal Shunt (SPRS)

The syringo-peritoneal shunt has undergone several modifications over the past five decades [41, 67]. The skin over the spine, neck, chest, and abdomen is draped as for a ventriculoperitoneal shunt. Under general anesthesia, the patient is placed in a lateral position with the head in a Mayfield 3 pins headrest. A single-level laminectomy and a midline durotomy are performed. The operating microscope is used to perform a small myelotomy into the thinnest portion of the spinal cord, usually the dorsal root entry zone. The use of IOM is recommended also for this operation and the considerations about IOM features and catheter repositioning are the same as described above for the SPS.

The flexible T-tube arms shunt with multiple drainage holes is used as this can be cut to the desired length and inserted into the syrinx. The shunt is brought out through the spinal cord and dura at a right angle and is secured with the suture tab. A metal step-up connector is used to adapt the SPRS to a standard peritoneal shunt tubing. Standard techniques are used to insert a low-pressure peritoneal catheter into the abdominal cavity. Similar to the SPS, adding a distal slit valve is beneficial to prevent over drainage. The shunt is then inserted into the peritoneum through a mini-laparotomy, with a peritoneal trocar, or laparoscopically assisted [10, 41].

10.5 Conclusions

The most important step in treating a syringomyelia is to differentiate it from other clinico-radiological conditions that should not be termed as syringomyelia.

In presence of a syringomyelia, it is of paramount importance to utilize appropriate diagnostic facilities available to identify the underlying cause, considering that this entity represents always an epiphenomenon of another pathology. If our best efforts to identify the cause have failed, then such syringomyelia should be deemed “idiopathic”. Most patients with this condition can be managed conservatively with clinical and radiological follow-up if asymptomatic or pauci-symptomatic. Rarely patients can present with deteriorating neurological signs and symptoms; in these circumstances, surgery should be warranted. Several techniques can be offered; however, the favored first line option is either a posterior thoracic laminectomy and arachnoidolysis or a syringo-subarachnoid shunt.

Refractory syringomyelia is a multifactorial condition, and understanding the pathophysiology of the persistent, progressing, or recurrent syrinx is crucial in order to offer the patient the right treatment [68].

In patients with refractory syringomyelia secondary to CM-1 treated in the first instance with foramen magnum decompression, reexploration of the foramen should be considered the initial option and enlarged watertight duroplasty should be performed if not performed during the first operation. A cranio-cervical instability should also be treated when present.

RS secondary to arachnoidal adhesions (posttraumatic, postinfective, or post-hemorrhagic arachnopathies) is the most difficult etiology to successfully treat. However, the best option is always to try to resolve the arachnopathy with arachnoidolysis, marsupialization of the subarachnoid space, and duroplasty before offering any kind of shunting diversion. If the above fails, among the different shunting techniques, SSS should be preferred over other shunting options when feasible.

Finally, it is worth stressing how a multidisciplinary approach is indicated in patients with refractory syringomyelia as there are several aspects of their care that need to be considered including pain management, neuropathic bladder and/or bowel, orthopedic issues, psychological support, and neurorehabilitation along with physiotherapy and occupational therapy.

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