16 Non-hindbrain-Related Syringomyelia

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The term syringomyelia was introduced by Ollivier D'Angers in 1827 [1] for cystic cavitations of the spinal cord. Syringomyelia describes a progressive accumulation of fluid inside the spinal cord. Up to this day, no pathophysiological concept for the development of syringomyelia is generally accepted [2]. However, with the advent of modern imaging techniques in the 1970s and 1980s, it became clear that a syrinx is always associated with other pathologies in the spinal canal or craniocervical junction. This observation has changed treatment concepts for these patients in a fundamental way. If the associated pathology can be treated successfully, no further measures for the syrinx are needed. It is now widely accepted that syringomyelia is related to intramedullary tumors or pathologies that cause a disturbance of cerebrospinal fluid (CSF) flow or spinal cord tethering [2, 3]. Table 16.1 gives an overview on the different pathologies related to syringomyelia in the author's series.

Currently, syringomyelia is considered as an accumulation of extracellular fluid of the spinal cord [3, 4]. In case of intramedullary tumors, it is generally believed that alterations of the blood-spinal cord barrier play a major role [5]. But this may not be the only mechanism. It is noteworthy that infiltrating intramedullary tumors rarely produce syringomyelia, whereas a syrinx is a common feature of displacing neoplasms [6]. More information is available on the effects of CSF flow obstructions on the spinal cord from animal [7] as well as computer models [8]. The subarachnoid space pressure is increased above the obstruction inducing changes of extracellular fluid distribution in the spinal cord [7], which may then lead to syringomyelia [3, 4]. Increased flow in the perivascular spaces has been implicated for this effect [3, 7, 9–12]. If flow capacities in the extracellular space are exceeded, there appears to be an evolution from spinal cord

Table 16.1 Pathologies associated with syringomyelia

Diagnosis	Total	Syringomyelia
Craniocervical junction	971	677
Chiari I	856	607 (70.9%)
Chiari II	54	28 (51.9%)
Foramen magnum arachnoiditis	34	34 (100%)
Posterior fossa tumors—Chiari I	13	3 (23.1%)
Posterior fossa arachnoid cysts—Chiari I	14	5 (35.7%)
Spinal canal	2897	1077
Posttraumatic syringomyelia	177	177
Non-traumatic arachnopathies	429	429
Intramedullary tumors	391	183 (46.8%)
Extramedullary tumors	971	117 (12.0%)
Extradural tumors	600	22 (3.7%)
Tethered cord syndromes	264	84 (31.8%)
Degenerative disc disease	65	65

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edema (i.e., the so-called presyrinx state) to syringomyelia [13]. Intramedullary neoplasms and cord tethering may alter extracellular fluid movements to similar effects. Once syringomyelia has developed, the increased intramedullary pressure [14] and fluid movements inside the syrinx [15, 16] may lead to spinal cord damage [17–19] and progressive neurological symptoms.

Diagnosis

In patients with Chiari malformations, CSF flow can be compromised by cerebellar tonsils filling the space of the cisterna magna, by arachnoid scarring in the foramen magnum area, and by obstruction of the foramen of Magendie. In posttraumatic syringomyelia, CSF flow obstruction may be caused by arachnoid scarring at the trauma level and narrowing of the spinal canal due to posttraumatic stenosis or kyphosis. Furthermore, posttraumatic cord tethering may contribute to syrinx development.

In the absence of a craniocervical malformation, an intramedullary tumor, a tethered cord syndrome, or a history of spinal trauma, syringomyelia is still considered idiopathic by many physicians. However, these patients have to be evaluated very carefully for radiological and clinical signs of arachnoid pathologies in the spinal canal causing CSF flow obstructions. The syrinx starts at the level of obstruction and expands from there. If the syrinx expands in a rostral direction, the obstruction will be found at the caudal end of the syrinx and vice versa. This also implies that the obstruction will most likely be found close to the largest diameter of the syrinx [2] (Fig. 16.1).

Due to the pulsatile movements of arachnoid septations, webs, or cysts, standard magnetic resonance imaging (MRI) may not always be able to demonstrate an arachnopathy directly. With a history of spinal meningitis or subarachnoid hemorrhage [20], the often quite extensive arachnopathy is rather easy to diagnose on MRI [21] (Fig. 16.2). Many arachnopathies, however, are quite discrete and extend over a few millimeters only. Cardiac gated cine MRI should be employed for such instances to study spinal CSF flow to identify areas of flow obstruction that may correspond to such circumscribed arachnoid pathologies [16, 21] (Fig. 16.1). Sometimes significant flow signals can also be detected in the syrinx itself. In such cases, the highest flow velocities in the syrinx can be expected adjacent to the arachnoid scarring (Fig. 16.1). The spinal cord should be studied with thin axial slices in T2 over the entire extent of the syrinx to search for areas of cord compression, displacement, or adhesion to the dura [21–23] (Figs. 16.1, 16.2, and 16.3). In the sagittal plane, the contour of the cord may appear distorted in areas of arachnoid scarring. Constructive interference in steady state (CISS) sequences can be used not only for the demonstration of the syrinx [24] but may also be helpful to detect arachnoid webs, scars, and cysts, because this technique is less susceptible to CSF flow artifacts [24]. Primary arachnopathies—i.e., unrelated to trauma or any other disease process—will almost always be found in the thoracic spine posterior to the spinal cord [21, 25]. Myelography and postmyelographic computed tomography (CT) are alternative methods to demonstrate arachnoid pathologies but have a lower sensitivity.

The sequence of events leading to a syrinx has implications not only for the neuroradiological appearance as just described but also for the evolution of clinical symptoms. The syrinx develops as a consequence of events that are set off by a pathology leading to CSF flow obstruction. Therefore, the first neurological symptoms in the patient's history are generally caused by this underlying pathology rather than the syrinx. In other words, a carefully taken clinical history can provide clues to the underlying pathology. If neurological signs spread to other parts of the body in an ascending pattern, the cause of the syrinx will be located at the lower pole of the syrinx and vice versa similar to the radiological evolution [2]. Apart from trauma, arachnoid scarring may be related to infection [26], hemorrhage [20], irritation by old contrast agents such as pantopaque [27], or surgery, to mention a few [21].

It is always puzzling that patients may harbor a huge syrinx and yet have just minor symptoms with exactly the opposite observation for some smaller syrinx cavities associated with major

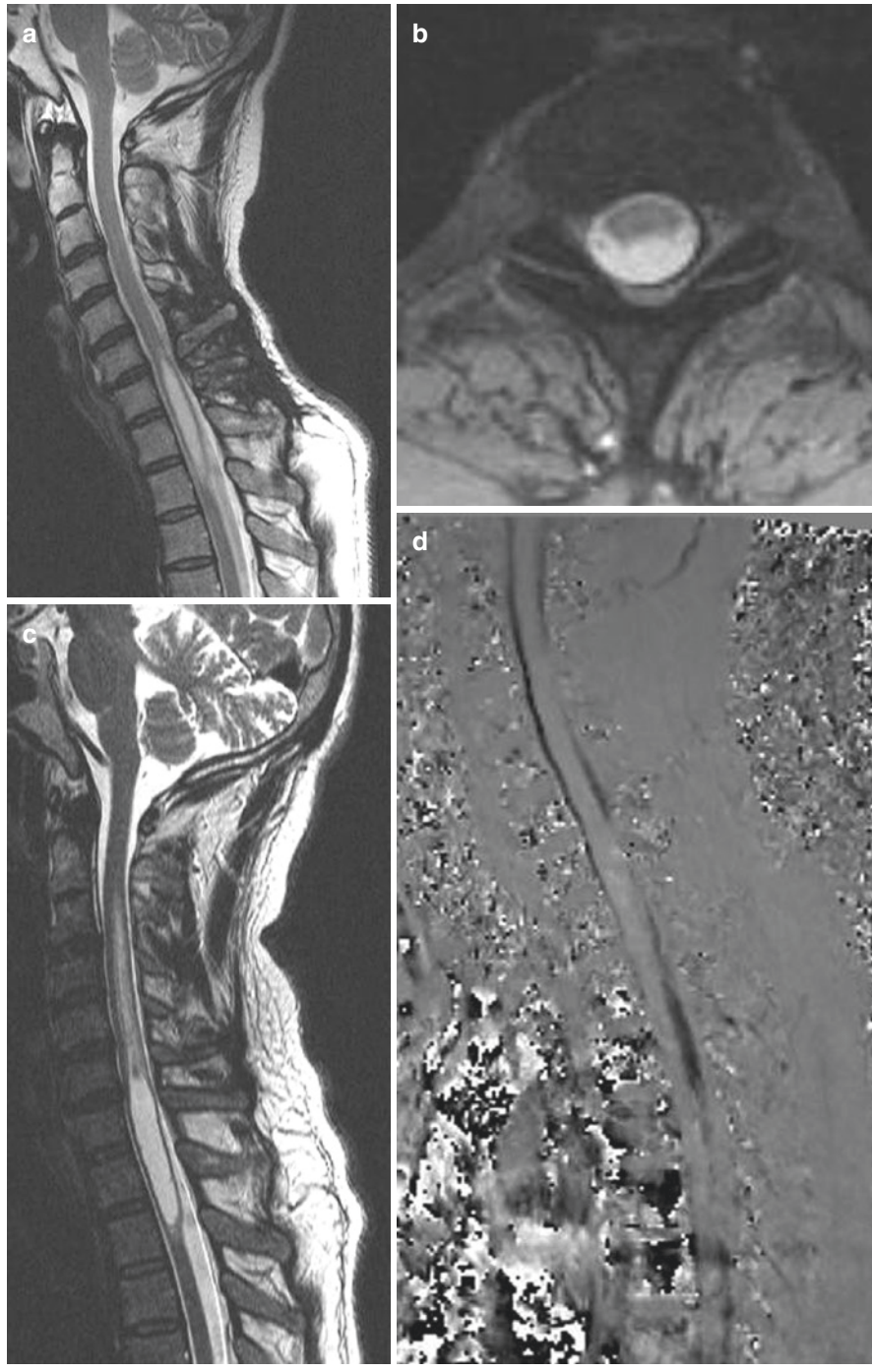
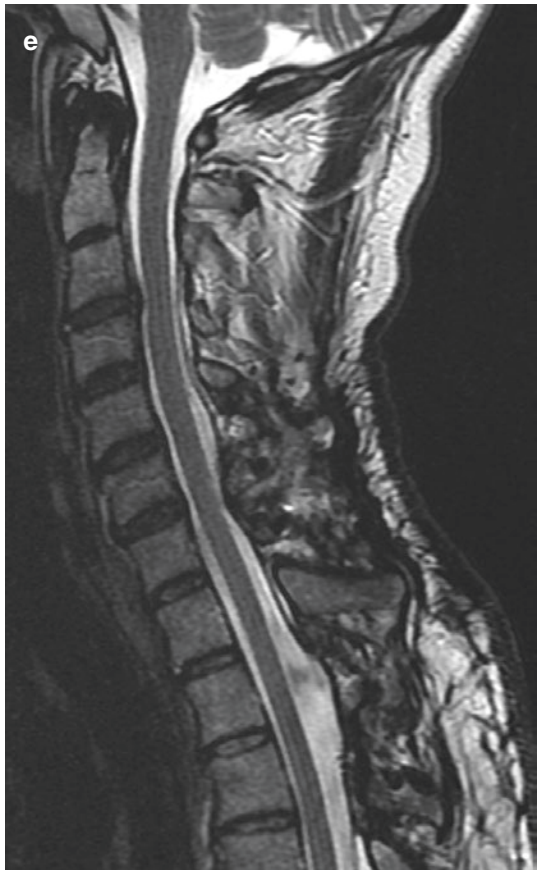


Fig. 16.1 (**a**) This sagittal T2-weighted MRI shows a syrinx extending from C6 to Th2 in a 43-year-old neuroradiologist with pain, sensory deficits, and dysesthesias in his left arm. Next to the lower pole, the spinal cord appears slightly indented. (**b**) The axial scan right below the syrinx demonstrates a slight posterior compression of the cord. (**c**) Two years later, the sagittal scan demonstrates the edema extending to C3 with increased diameter of the syrinx C6 to Th2. (**d**) The cine MRI shows a diminished CSF flow posteriorly across the syrinx and at Th2. (**e**) After decompression at Th2, the postoperative MRI demonstrates complete resolution of the syrinx and edema with no further compression of the cord in the axial scan (**f**). Postoperatively, the patient reported no change with 2 years of follow-up



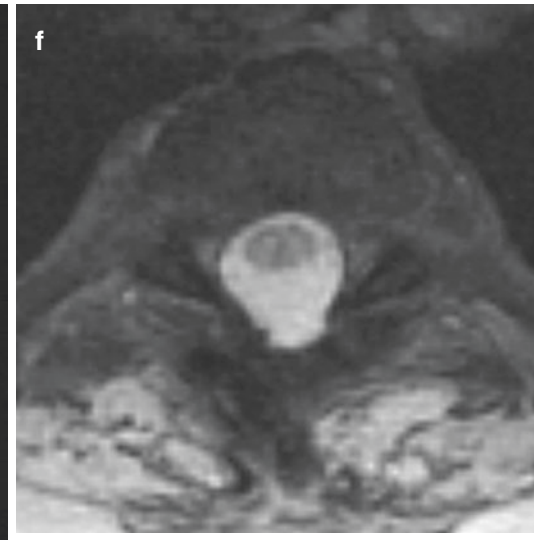


Fig. 16.1 (continued)

neurological deficits. One explanation for this paradox may be that a great deal of the clinical problems are related to the underlying disease process causing the syrinx rather than to the syrinx itself [2, 28]. This holds particularly for patients with extensive arachnopathies completely encircling the spinal cord after multiple surgeries, subarachnoid hemorrhages, or spinal meningitis [21, 25]. Similarly, a syrinx associated with a tethered cord or an intramedullary tumor will almost never become symptomatic as signs of cord compression and tethering dominate the clinical picture [2].

The classical symptoms of syringomyelia are a dissociated sensory loss with loss of sensation for temperature and pain but preserved sensation for light touch. Pain related to syringomyelia is either permanent or aggravated by maneuvers such as coughing and sneezing and perceived in dermatomes corresponding to the syrinx. Late symptoms of syringomyelia are muscle atrophies corresponding to damage of anterior horn cells or trophic changes leading to skin and joint damages, particularly in the shoulder and elbow [2, 28].

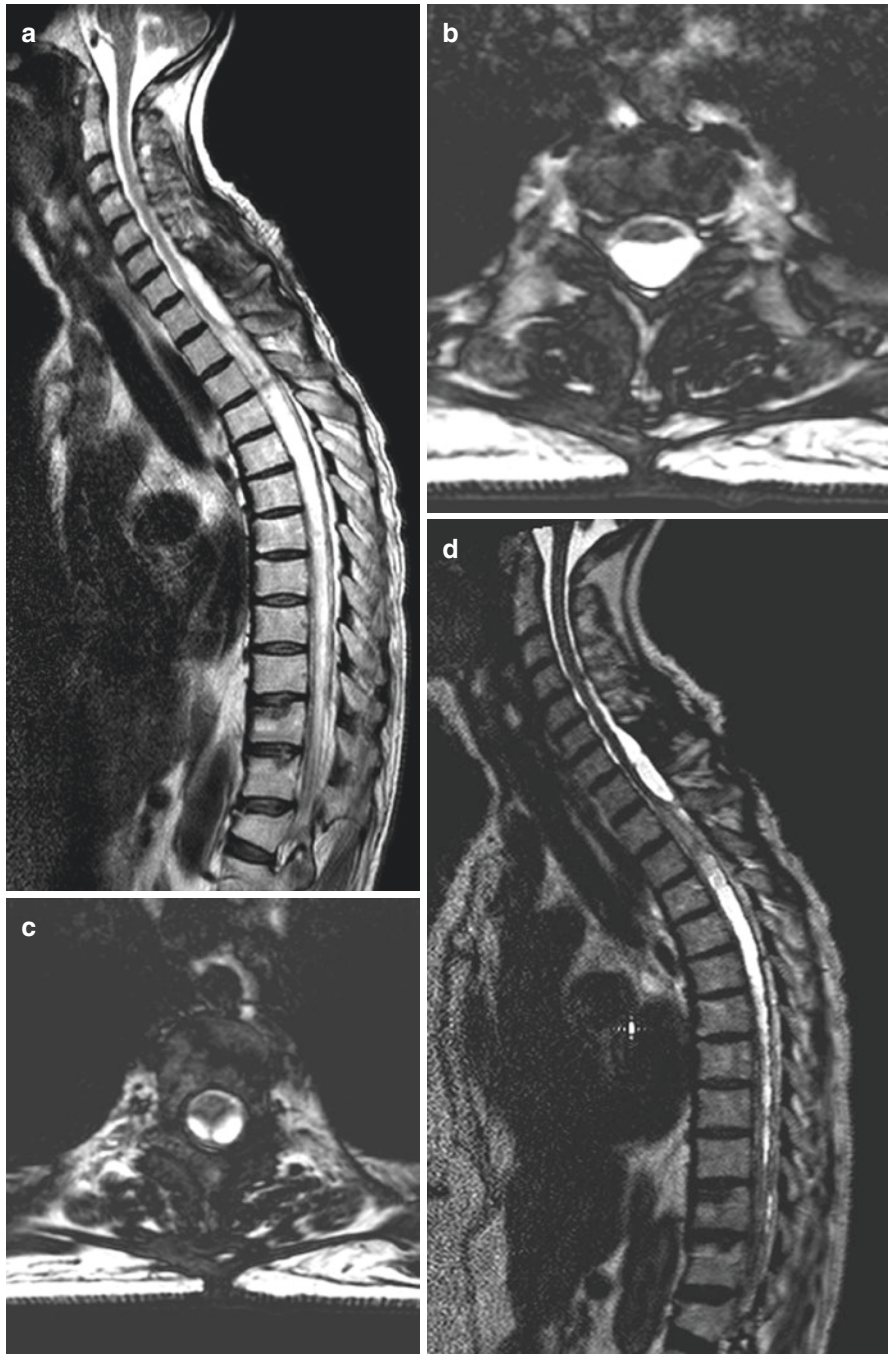
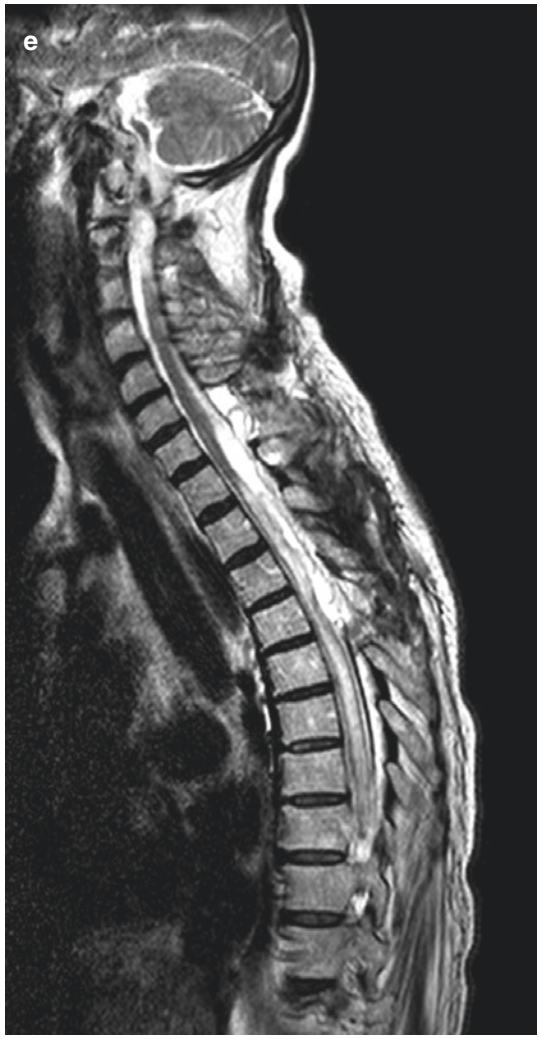


Fig. 16.2 (**a**) The sagittal T2-weighted MRI shows a syrinx Th2 to Th12 related to an extensive postmeningitic arachnopathy extending from C7 downward, causing compression of the cervical cord as demonstrated in the axial scan (**b**) in a 67-year-old woman with progressive paraparesis making her wheelchair dependent after unsuccessful fenestration of arachnoid septations in the cervicothoracic region in another institution. (**c**) The posterior median arachnoid septum appears thickened. (**d**) Thin sagittal slices in T2 demonstrate arachnoid septations, cysts, and adhesions between the cord and dura from C7 downward throughout the entire thoracic canal. The postoperative sagittal (**e**) and axial (**f**) MRIs show the decompression of the spinal cord after fenestration of arachnoid septations and cysts in the cervicothoracic area. The patient reported postoperative improvement of sensory functions, dysesthesias, pain, and motor weakness but was still left confined to a wheelchair



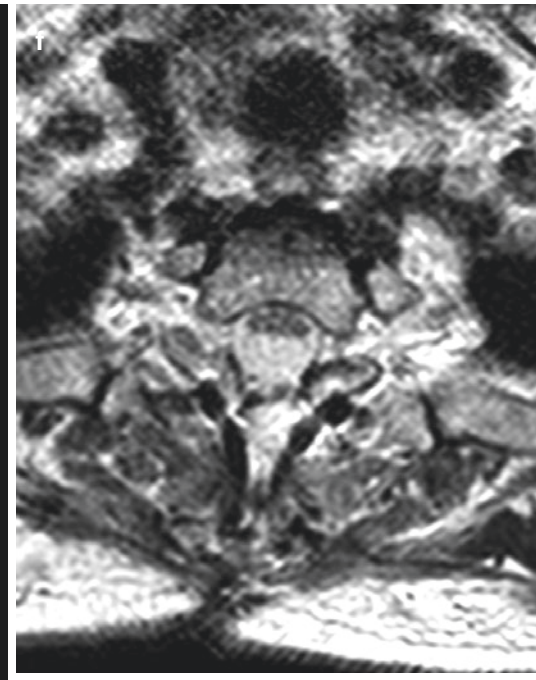


Fig. 16.2 (continued)

Management

About 71.1% of patients with a Chiari I malformation developed a syrinx in the author's series. No other pathology causes syringomyelia in such a high proportion (Table 16.1). For syringomyelia associated with intramedullary tumors and Chiari I malformation, the rates for postoperative syrinx resolution are above 80%, provided the tumor is removed and all components contributing to CSF flow obstruction in Chiari malformations have been surgically addressed, respectively.

Successful treatment of syringomyelia related to spinal rather than craniocervical CSF flow obstructions is much more challenging. The underlying causes are more difficult to identify and to deal with surgically. For spinal arachnopathies, surgery should be reserved for patients with progressive symptoms. Nevertheless, treating the cause of the syrinx with arachnolysis and duraplasty is rewarded by considerably better results compared to syrinx shunting procedures [25, 28–32].

Neuropathic pain and dysesthesias, particularly those of burning character, may be major

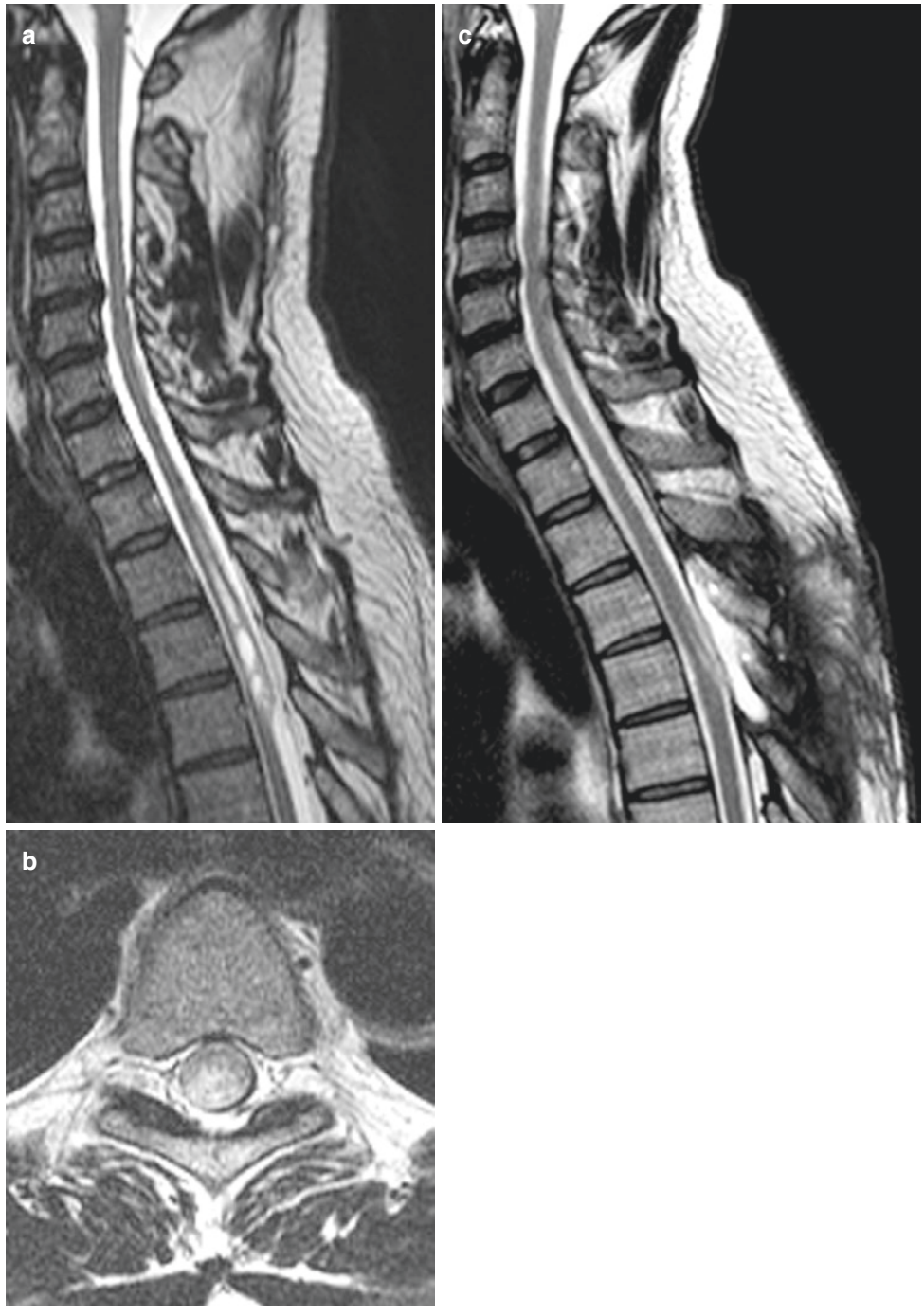


Fig. 16.3 (**a**) The sagittal T2-weighted MRI shows a posttraumatic syrinx Th2–Th4 in a 46-year-old man 16 months after suffering an incomplete cord injury at Th4 with sensory but no motor deficits. The cyst caliber is largest at the lower pole. (**b**) At Th4/Th5 the axial scan reveals an area of cord compression by a cystic posttraumatic arachnopathy at this level. (**c**) After decompression at Th3 and Th4, the postoperative MRI demonstrates a complete resolution of the syrinx. Postoperatively, symptoms remained unchanged for 14 months

clinical problems. Even though these may improve with successful treatment of the syrinx, this is never certain. Therefore, the decision for or against surgery should be based on the course of neurological signs and symptoms rather than pain syndromes alone.

In general, surgery can be recommended for patients with arachnoid scarring limited to about 2–3 spinal segments in the posterior section of the subarachnoid space [21, 25] (Figs. 16.1 and 16.3). All operations are performed in prone position. Laminotomies are recommended to reinsert the lamina at the end of the operation with titanium miniplates. After exposure of the dura, the extent of the arachnoid pathology can be visualized with ultrasound. The syrinx can be visualized. Pulsations of syrinx fluid and CSF may become visible. Sometimes, arachnoid septations can be seen. Most importantly, the safest spot for opening of the dura can be chosen with this technique. As contamination of the CSF with blood may cause inflammatory reactions of the arachnoid, great care is taken to achieve good hemostasis. For this purpose, the entire surgical field is covered by moist cottonoids, which keep soft tissues moist and soak up any minor bleeding. Then the dura is opened under the operating microscope in the midline without opening of the arachnoid. Once the dura is held open with sutures, the arachnoid pathology can be studied, and adequate exposure cranially and caudally is ensured in order to gain access to normal and unaffected subarachnoid space on either end. Obviously, any surgeon should be familiar with the normal anatomy of the spinal subarachnoid space [33]. The posterior subarachnoid space is divided in two halves by a posterior longitudinal arachnoid septum. This septum extends between the outer arachnoid layer and an intermediate layer on the cord surface. The insertion on the cord surface is related to the midline dorsal vein. Further strands of arachnoid may be encountered in the posterior and—to a lesser degree—lateral subarachnoid space. Another landmark is the dentate ligaments, which originate from the spinal cord pia mater, run between posterior and anterior nerve roots, and insert close to the dural nerve root sleeve. With a microdissector, arachnoid and dura can be separated from each other without any problem in areas without arachnoid scarring, i.e., at either end of the exposure. In the area of scarring, sharp dissection with microscissors is usually required to achieve this. At the level of CSF flow obstruction, the arachnoid may become densely adherent to the cord surface. With opening of the rostral and caudal subarachnoid space, CSF flushes into the surgical field, and often the cord, which was distended by the syrinx, starts to pulsate, and the syrinx may collapse at this point. The arachnoid scar can be resected layer by layer leaving a last sheath on the cord surface to avoid injury to the cord or surface vessels. This last layer resembles the intermediate arachnoidal layer mentioned previously. In this way, a free CSF passage in the posterior subarachnoid space can be created in every patient across the region of the arachnopathy. Dissection is then continued laterally on either side toward the dentate ligaments. This leads to complete untethering of the cord in the majority of cases. No dissection should be performed anteriorly of the dentate ligaments to avoid injuries to motor pathways and anterior spinal cord vessels. Closing the microsurgical part of the operation, an expansile duraplasty is inserted with a tight running suture and finally lifted up with tenting sutures on either side. To avoid scar formation and tethering between duraplasty and spinal cord, alloplastic material for duraplasty should be preferred, i.e., Gore-Tex® (W.L. Gore & Associates GmbH, Putzbrunn, Germany). Special attention is finally paid to a good, tight closure of the muscle layer to prevent any CSF from entering the epifascial space [25, 29]. In patients who have been operated before, as in patients with posttraumatic syringomyelia who underwent spinal instrumentation, for instance, a lumbar drain is placed prophylactically if the soft tissue appears scarred and sparsely vascularized.

Considerable experience is needed to be successful with this surgical technique. The more focused the surgery, the less scarring may result. If unnecessary steps are taken, such as a too extensive dura opening, or the surgical field is contaminated with considerable amounts of blood, postoperative scarring may counterbalance completely the effect of surgery. On the other hand, if the dura opening is not extensive enough to gain access to the normal subarachnoid space above and below the level of scarring, the

procedure is insufficient. As always, it is the right measure that counts and determines whether an operation will be successful.

For patients with more extensive arachnopathies after meningitis, multiple intradural surgeries, or spinal subarachnoid hemorrhage, surgery can rarely provide a sustained normal CSF passage [21, 25, 28]. Axial MRIs taken over the entire area of the arachnopathy may be evaluated in such instances for evidence of cord compression. Quite often, pouches and cysts have formed causing profound cord compression over a few spinal segments. Such compressions can be treated surgically by wide fenestration of the corresponding arachnoid membranes in individual cases (Fig. 16.2). Such an operation can improve neurological symptoms related to the cord compression for some time, but it will neither influence the syrinx nor the further neurological progress in the long term [21].

For patients, in whom the cause of syringomyelia is not amenable to surgery, thecoperitoneal shunts have been introduced, which drain CSF from the subarachnoid space above the level of obstruction to the peritoneal cavity [34–38]. For cavities extending into the cervical cord, ventriculoperitoneal shunts have been used for the same purpose [39, 40]. However, these shunts have their problems. There is little experience concerning the correct pressure settings other than to set them as low as possible avoiding signs of overdrainage or low intracranial pressure. Ten patients in the author's series were treated with low-pressure shunts of 4 cm H_2O opening pressure. In one patient, this low pressure was still not low enough, so that the valve was removed leaving the patient with a valveless drain. One patient developed a low-pressure syndrome including subdural effusions requiring surgery so that the shunt needed to be removed despite a good effect on the syrinx. Overall, about half of the patients benefitted from thecoperitoneal shunts for at least 4 years in the author's series (Fig. 16.4).

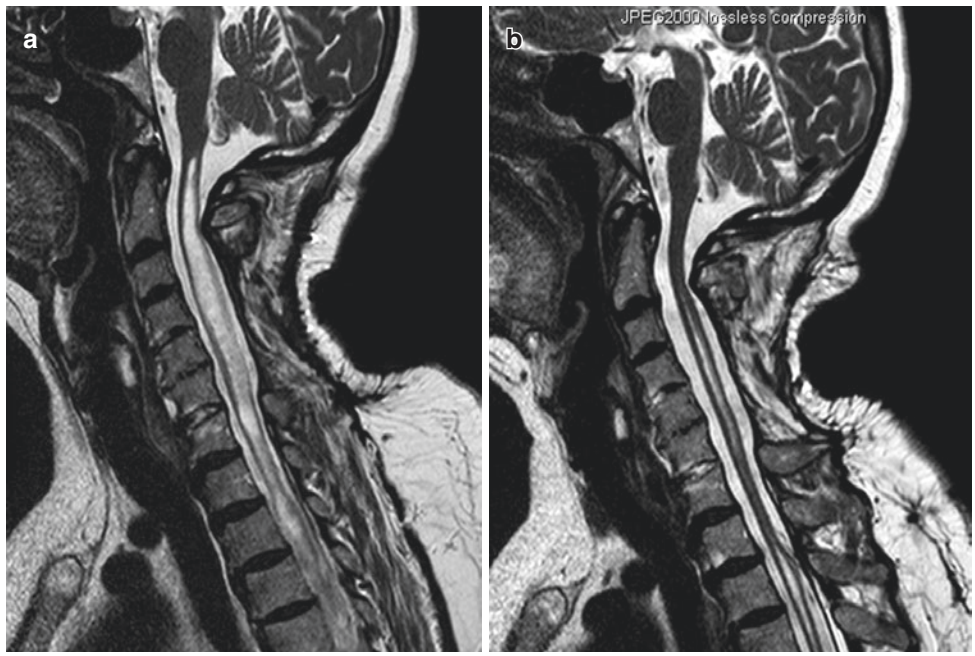


Fig. 16.4 (**a**) The sagittal T2-weighted MRI shows a posttraumatic syringomyelia extending from the injury level at Th12 up to the foramen magnum in a 61-year-old woman 40 years after the accident. In the previous 5 years, she underwent two attempts to improve CSF flow at the level of her incomplete cord lesion. Due to severe adhesions between the conus and dura, the posttraumatic tethering could not be completely resolved, and postoperative arachnoid scarring obstructed the subarachnoid space within weeks of these attempts. Furthermore, a posterior decompression and fusion of her cervical spine had been performed to eliminate any further cervical cord damage from her multilevel cervical stenosis. (**b**) Four years after placement of a thecoperitoneal shunt at the level of Th1/Th2, the syrinx is still considerably decreased in size with sustained resolution of neurological symptoms in her right arm and hand

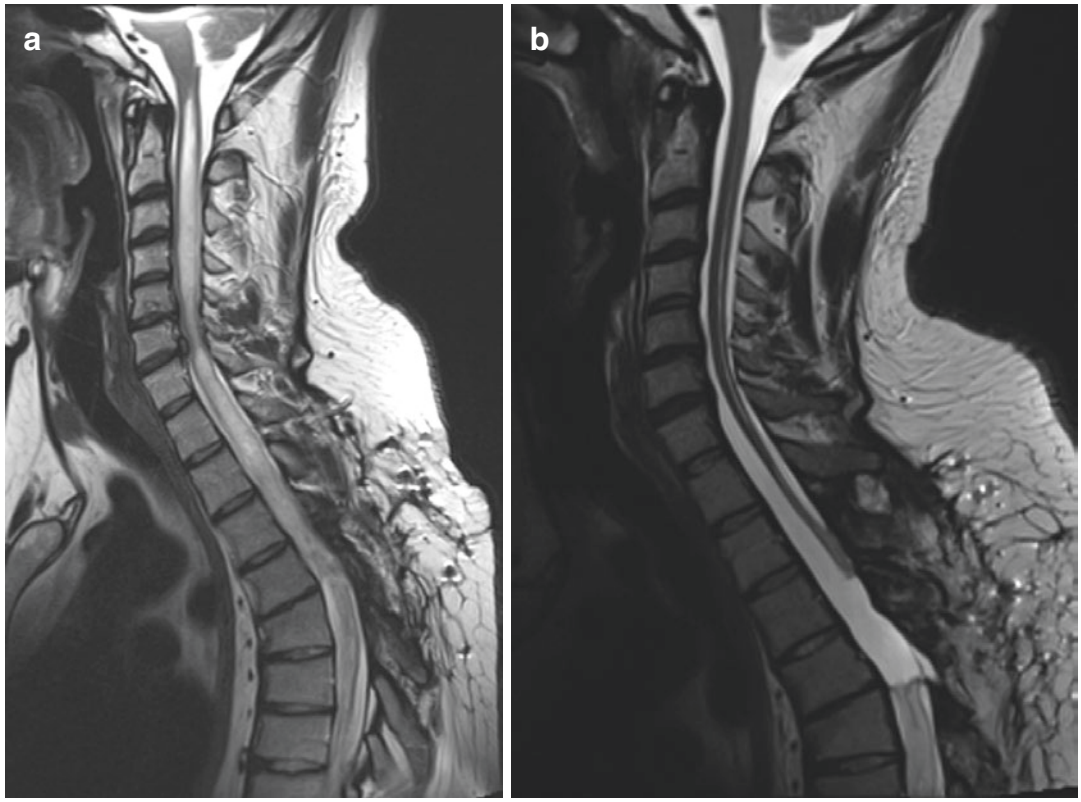


Fig. 16.5 (**a**) The sagittal T2-weighted MRI shows a posttraumatic syringomyelia C2 to Th6 14 years after a motorcycle accident resulting in complete paraplegia with an injury level at Th4/Th5. One year previously, an attempt to improve CSF flow at Th4/Th5 with untethering of the cord was undertaken, which led to a short-lived resolution of the syrinx. With reappearance of the syrinx, the patient requested a revision fearing permanent neurological deficits in his upper extremities. A cordectomy at Th4 was undertaken. (**b**) The postoperative MRI shows a complete resolution of the cervical syrinx and decrease in size below that level. The dissociated sensory loss in his right arm improved during a follow-up of 14 months

For patients with a complete cord lesion, cordectomy is a very effective form of treatment for syringomyelia [29, 41–46]. All 17 patients treated in this manner in the author's series improved neurologically with permanent resolution of the syrinx (Fig. 16.5). However, the psychological burden for a patient to accept this operation should not be underestimated. Most patients prefer to undergo a decompression first. After all, this operation does provide good results for the majority of patients [45]. Patients will accept a cordectomy, however, if the ascending neurology cannot be arrested by decompression or shunting procedures and the neurological progress threatens important functions such as respiratory or hand muscles.

Results

Concentrating on patients with syringomyelia related to spinal arachnopathies, 177 patients with posttraumatic arachnoid scarring and 429 patients with non-traumatic arachnopathies were encountered in the author's series. Reserving surgery for patients with progressive neurological symptoms and refusal of surgery by some patients led to 119 operations for 92 patients with post-

Table 16.2 Operations for patients with spinal arachnopathies

Type of surgery	Posttraumatic arachnopathies 92 patients	Non-traumatic arachnopathies 135 patients	All 227 patients
Arachnolysis + duraplasty	86	139	225
Cordectomy	16	1	17
Thecoperitoneal shunt	4	6	10
Ventral fusion	5	4	9
Posterior decompression	7	4	11
Opiate pump	1	–	1

traumatic and 154 operations for 135 patients with non-traumatic arachnopathies (Table 16.2). Overall, 225 decompressions aiming at improving CSF flow and decompressing the spinal cord by arachnolysis and duraplasty were performed, while 17 patients with complete paraplegias underwent cordectomies and 10 thecoperitoneal shunts were placed. One patient received an opiate pump for his neuropathic pain syndrome. Twenty operations dealt with degenerative diseases of the cervical spine.

Concentrating on the 225 decompressions with arachnolysis and duraplasty, complications were observed after 59 operations (26.0%); the commonest were urinary tract infections (8%), wound infections (6%), hemorrhages (5%), and CSF fistulas (3%). Permanent surgical morbidity defined as a permanent neurological worsening within 1 month after surgery occurred in 19 patients, i.e., 8.4%. A postoperative decrease of the syrinx was observed in 76%, and 21% showed no postoperative change, while 4% increased further despite surgery. After 3 months, 49% considered their condition improved, 42% as unchanged, and 9% as worsened. Looking at individual symptoms revealed postoperative improvements for sensory deficits and pain, whereas motor weakness, gait, and sphincter functions were left unchanged. Long-term results were determined with Kaplan-Meier statistics to determine the rates for progression-free survival after surgery. Overall, 63% of all patients undergoing a decompression with arachnolysis and duraplasty for traumatic or non-traumatic arachnopathies regardless of etiology remained in an unchanged or improved neurological status for at least 5 years after surgery. This rate was reduced to 48% after 10 years.

However, not all patients with spinal arachnopathies are good candidates for this type of surgery, while others respond particularly well. Looking at subgroups revealed good long-term results for patients with focal non-traumatic arachnopathies not exceeding two spinal segments operated first time and for posttraumatic patients who had not conceded a spinal cord injury with their accident (Table 16.3). For these subgroups, significantly higher progression-free survival rates for 10 years of 76% and 89%, respectively, were determined. Patients with an incomplete cord injury or those requiring a revision, on the other hand, were the most difficult to treat. The risk of further spinal cord damage in these patients is very real so that intraoperative compromises were often required, especially when trying to dissect dense adhesions between the spinal cord and dura. For patients with extensive arachnopathies after meningitis or intradural hemorrhages, the surgical concept of establishing a permanently improved CSF passage cannot be generally recommended. For such patients, a causative treatment leading to a better outcome compared to the natural history is not available. Symptomatic treatment with thecoperitoneal shunts offers better results in such patients although they address the syrinx only and leave impaired spinal cord blood flow due to the arachnoiditis untreated. The same applies to syrinx shunts, which, according to the author's experience, provide less favorable results compared to thecoperitoneal shunts.

Table 16.3 Progression-free survival for patients with spinal arachnopathies

Patient group	5 years	10 years	*P*
All arachnopathies	63%	48%	
Non-traumatic Focal—first surgery	86%	76%	0.001
Non-traumatic Focal—revision surgery	44%	–	
Non-traumatic Focal	80%	72%	<0.0001
Non-traumatic Extensive	29%	23%	
Non-traumatic All	63%	55%	
Posttraumatic No cord injury	89%	89%	0.03
Posttraumatic Incomplete cord injury	53%	19%	
Posttraumatic Complete cord injury	63%	46%	
Posttraumatic All	62%	36%	

Conclusion

The diagnosis of syringomyelia should be reserved for patients with a space-occupying intramedullary cyst of progressive character and differentiated from such entities such as a dilatation of the central canal or myelomalacia [2, 28, 47, 48]. Syringomyelia is not a disease in its own right but a manifestation of a disorder of the spinal canal or craniocervical junction that has either resulted in an obstruction of CSF flow or spinal cord tethering or is associated with an intramedullary tumor [2]. Management of patients with syringomyelia requires the correct diagnosis of the underlying disorder and the successful treatment of it. The long-term prognosis depends on the treatability of the underlying disorder. Whenever this can be achieved, no further surgical measures for the syrinx are required [2].

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