Syringomyelia

W. Peter Vandertop[1]

[1] Neurosurgical Center Amsterdam, VU University Medical Center, Amsterdam, The Netherlands

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Address for correspondence W. Peter Vandertop, MD, PhD, VU University Medical Center, De Boelelaan 1117, 2F-020, PO Box 7057, 1007 MB Amsterdam, The Netherlands (e-mail: wp.vandertop@vumc.nl).

Abstract

Syringomyelia is an etiologically diverse affliction caused by disturbance of normal cerebrospinal fluid flow dynamics. An evaluation by comprehensive imaging, using advanced three-dimensional constructive interference in steady state and four-dimensional phase contrast imaging techniques, should be focused on finding the causative lesion because this determines which surgical strategy could be employed. This review discusses the pathogenesis and various etiologies of syringomyelia, and focuses on the specific surgical approach for each, mentioning aspects of surgical complications and prognosis. Particular attention is given to the "persistent central canal," a normal anatomical variant of the persistent fetal configuration of the central canal, as this is increasingly seen nowadays on routine magnetic resonance imaging.

Keywords
- syringomyelia
- spinal cord diseases
- neural tube defects
- malformation
- Arnold-Chiari

> Syrinx (Gr. *Συρινξ*) was a Naiad fresh-water nymph who lived on the cold mountainsides of Arcadia. One day she was amourously pursued by the Greek god Pan, and fleeing from her chaser, she was barred by the river Ladon. She asked for assistance from her Sorores Liquidae (Watery Sisters), who transformed her into a hollow water reed. When the god's frustrated breath blew over the river, it made a haunting sound. Pan cut the reeds to fashion the first set of pan pipes, which were thenceforth known as syrinx.

Ovid, Metamorphoses 1. 689 ff (trans. Melville) (Roman epic C1st BC to C1st AD)

Introduction

Syringomyelia literally means "cavity within the spinal cord" and is typically a progressive chronic condition. The syrinx is either a fluid-filled, gliosis-lined cavity within the spinal cord parenchyma or it is a focal dilatation of the central canal, in which case it is called hydromyelia.[1] In everyday clinical practice, the term syringomyelia is commonly used for both conditions. Most lesions are located between C2 and T9, but they can descend down to the conus medullaris, or extend upward into the brain stem (i.e., syringobulbia). The estimated prevalence varies widely from 1.94 per 100,000 in Japan to 8.4 per 100,000 in Western countries and the cause for this wide variety is unknown.[2,3]

In children, syringomyelia usually occurs in the setting of congenital anomalies, most commonly the Chiari-I malformation and tethered cord, but it can also develop years after meningitis and spinal trauma or with intramedullary and extramedullary tumors. Several clinical aspects relevant for the diagnosis and treatment are reviewed.

Pathogenesis

Congenital syringomyelia is seen as an abnormality of primary neurulation and can be distinguished into two forms.[4] The embryonic form consists of an abnormal dilatation of the central canal of the spinal cord, with a thin and elongated roof plate, and lack, or near-absence, of mesenchymal tissue between the spinal cord and the surface ectoderm, causing a gap in the posterior vertebral arches—an open neural tube defect. The fetal form on the contrary does not affect mesenchymal tissue development and is therefore associated with an intact vertebral column—a closed neural tube defect.

Acquired syringomyelia is thought to be caused by disturbance in normal cerebrospinal fluid (CSF) circulation.

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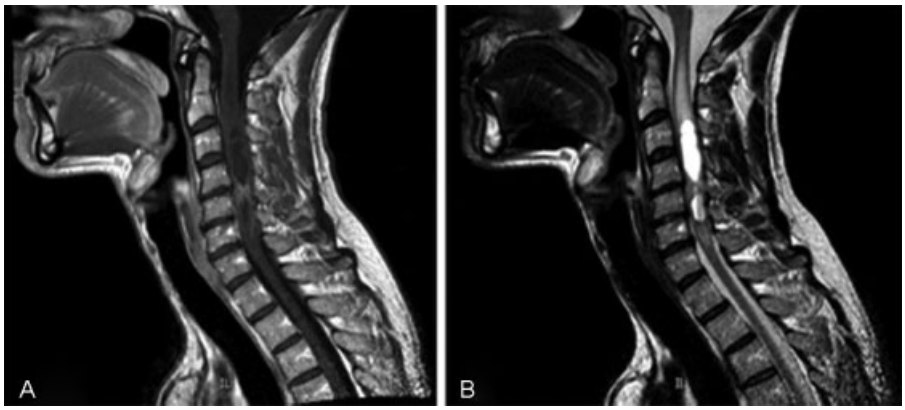

Fig. 1 Spinal cord syrinx, edema, and tumor cysts in spinal cord ependymoma. (A) Sagittal T1-weighted image after intravenous gadolinium administration shows a multiloculated lesion, with irregular enhancement in the spinal cord from C2 to C6. (B) Sagittal T2-weighted image shows the cystic portion of the tumor better with spinal cord hyperintensity indicative of spinal cord edema extending cranially to the medulla oblongata.

In Chiari-I malformation, this disturbance is caused by the downward herniation of the cerebellar tonsils, obstructing normal CSF flow from the cranial to spinal compartment during the normal cardiac cycle or normal Valsalva maneuvers, such as coughing, laughing, etc. These abnormal pressure gradients eventually lead to accumulation of fluid in either the central canal or the spinal cord parenchyma. The same mechanism is thought to be responsible for syringomyelia after infection (e.g., meningitis), inflammation (e.g., sarcoidosis and multiple sclerosis), trauma (adhesive arachnoiditis), and extramedullary lesions (e.g., tumors and arachnoid cysts). Syringomyelia in association with intramedullary tumors may have a similar pathogenesis, but it can also be the result of tumor activity per se, as seen in ependymomas and hemangioblastomas (►**Figs. 1** and **2**).

New formation of a syrinx, or enlargement of an already present syrinx, in a child with a tethered spinal cord can also be caused by an accompanying Chiari-II malformation, causing likewise obstruction of normal CSF flow dynamics as in Chiari-I malformations. Mechanical effects of spinal cord tethering or metabolic and ischemic changes in the distal spinal cord have also been suggested as contributing factors in syrinx formation in these patients.[5]

Etiology

The etiology of syringomyelia can be deduced from its presumed pathogenesis such as, a congenital malformation, any acquired lesion, or disease, causing disturbance of normal CSF flow dynamics.

The most common congenital causes are neural tube defects (myelomeningocele and tethered cord syndrome) and Chiari malformations. Magnetic resonance imaging (MRI) reveals syringomyelia in 30 to 50% of these patients, ranging from localized, segmental cavities to severe holocord cavities.[6] Most syringes are present since birth, and others

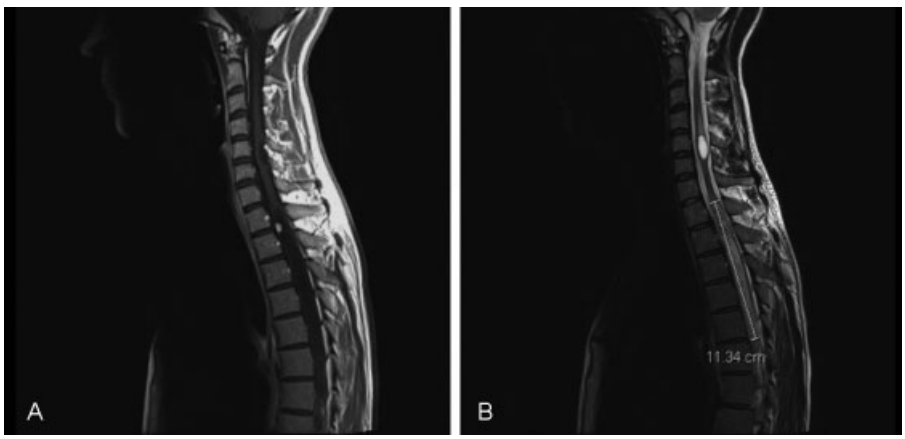

Fig. 2 Extensive syringomyelia with hemangioblastomas in von Hippel–Lindau disease. (A) Sagittal T1-weighted image after intravenous gadolinium administration shows a syrinx behind C5 with a very small nodular enhancing lesion located dorsally and cranially in the cyst wall and an extensive thoracic syringomyelia caused by a very small enhancing nodule behind T1–T2 located ventrally in the cyst. (B) Sagittal T2-weighted image shows the cervical syrinx more clearly with rostral extension of hyperintensity, indicative of spinal cord edema, and the extent of the thoracic syrinx (11.34 cm).

develop (or enlarge) during life. Other congenital spinal anomalies, such as, Klippel–Feil syndrome, that cause spinal canal narrowing may also lead to CSF flow disturbances with resulting syringomyelia.[7]

Acquired causes of syringomyelia are plentiful and include the following:

- Hydrocephalus, leading to pegging of the cerebellar tonsils, and therefore mimicking Chiari-I malformation.
- Postinfectious, after any kind of meningitis, as this invariably leads to arachnoid scarring.
- Postinflammatory after, for example, transverse myelitis, sarcoidosis, and multiple sclerosis.[8–10]
- Posttraumatic, including postsurgical, arachnoid scarring.
- Spinal cord tumors, especially ependymomas and hemangioblastomas.
- Extramedullary tumors and arachnoid cysts.
- Spinal canal stenosis, especially cervical, usually seen only in adults.

Symptoms

A syrinx can be asymptomatic and then it is only discovered on incidental spinal cord imaging. In symptomatic patients with syringomyelia, bilateral motor and sensory symptoms and signs that do not involve the head or face can be seen as in any other spinal cord lesion.

Patients often present with pain, weakness, and stiffness in the back, shoulders, and extremities. Loss of the ability to feel extremes of hot or cold, especially in the hands, is frequently seen, with a cape-like loss of pain and temperature sensation along the back and arms, caused by damage to the centrally crossing fibers of the spinothalamic tract. Symptoms of autonomic bladder and bowel dysfunction are very uncommon until end-stage spinal cord dysfunction. The course of the symptoms often fluctuates in severity and duration, and each patient experiences a different combination of these symptoms, typically varying depending on the extent and the location of the syrinx within the spinal cord.

The clinical picture in patients with a neural tube defect is somewhat different. Usually they present with new neurological deficits in the lower extremities, bowel, and bladder dysfunction (often asymptomatic and only found on urological dynamic examinations), or pain, and this actually cannot be distinguished from clinical deterioration caused by the tethered cord itself, rather than by the syringomyelia. However, scoliosis seems to be a prominent feature in those patients who have a terminal syrinx, independent of the size of the syrinx.[11] Syringomyelia extending upward into the cervical spinal cord can present with hand weakness and sensory symptoms, and this can be difficult to distinguish from symptomatic Chiari-II malformation, which is invariably present in these patients. Fortunately, the surgical therapy is the same for both.

In closed neural tube defects, the syrinx is usually located immediately rostral to the tethering lipoma or bony spur and remains localized to the lower thoracic and lumbar levels. In meningomyelocele patients, the syrinx can be quite extensive and reach up to the cervical spinal cord.[12]

Imaging

Visualization of intramedullary cavities or perimedullary CSF flow disturbances used to be very difficult. Invasive contrast myelography was required and the images obtained were quite "crude". This was improved upon by computed tomographic (CT) myelography, until the advent of MRI, which can easily identify the location and extent of any intramedullary cavity. To detect, sometimes subtle, extramedullary arachnoid adhesions or membranes, advanced imaging techniques, such as, three-dimensional (3D) constructive interference in steady state (CISS) sequences, can be very helpful and often indispensable. Using electrocardiographically gated flow-sensitive techniques, such as four-dimensional (4D) phase contrast (PC) MRI, CSF flow dynamics can be studied to distinguish physiological from complex pathological flow patterns at the craniocervical junction and cervical and thoracic spine, and to detect cystic spaces not communicating with normal CSF spaces.[13–15]

For the complete work-up of a patient with syringomyelia, one should keep in mind that all possible causes should be eliminated, so MRI of the lumbar-sacral spine to rule out a tethered cord is mandatory, as is an MR scan of the head and craniocervical junction to rule out hydrocephalus and Chiari malformation. The administration of contrast medium is essential to diagnose any associated conditions, such as, intramedullary and extramedullary tumors.

Differential Diagnosis

When evaluating a patient with syringomyelia it is important to first identify the etiology, as any therapeutic strategy will most certainly focus on a causative lesion rather than on the syrinx itself. This means that hydrocephalus, or shunt malfunction, should be ruled out by MR scanning of the head, and tethered cord should be excluded by MR scanning of the lumbosacral spine, as should any other lesion as discussed in the "etiology" section. In the absence of a causative lesion, there are not many alternatives. However, nowadays widespread MR availability and pre-emptive screening of asymptomatic individuals is increasingly leading to incidental MR findings of intramedullary cavities in all forms, shapes and sizes, and in a significant percentage of these patients no causative lesion or mechanism can be found, even with all advanced imaging techniques. In these cases, a diagnosis of "idiopathic localized hydromyelia" is sometimes made, but as this condition probably represents persistence into adult life of a fetal configuration of the central canal of the spinal cord, it is probably wiser to refer to this condition as the "persistent central canal."[16,17] The central canal of the spinal cord is present at birth and usually becomes progressively obliterated, but we now know that it can persist partially or completely. To make a diagnosis of "persistent central canal," there should not be any factors predisposing to syringomyelia and the syrinx should be located at the junction of the ventral one-third and

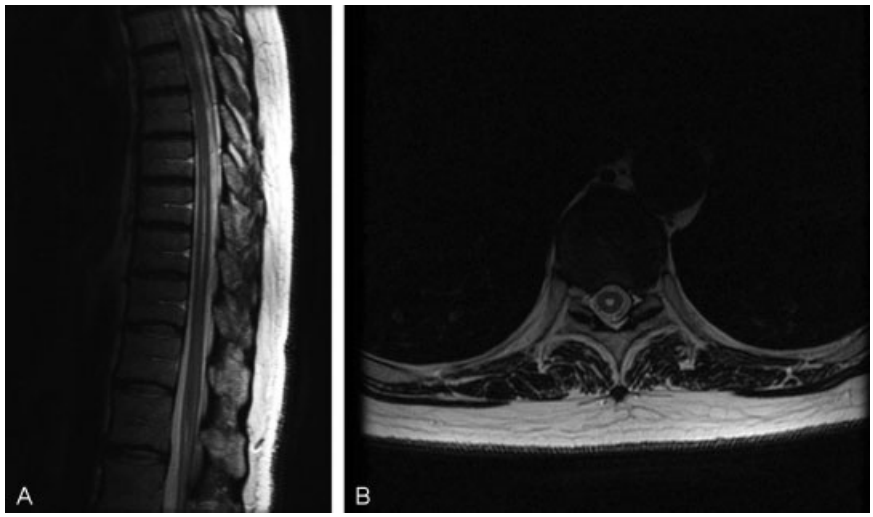

Fig. 3 Thoracic persistent central canal. (A) Sagittal T2-weighted image of a small, thoracic syrinx, filiform in shape. (B) Axial T2-weighted image showing the cavity at the junction of the ventral one-third and dorsal two-third of the spinal cord.

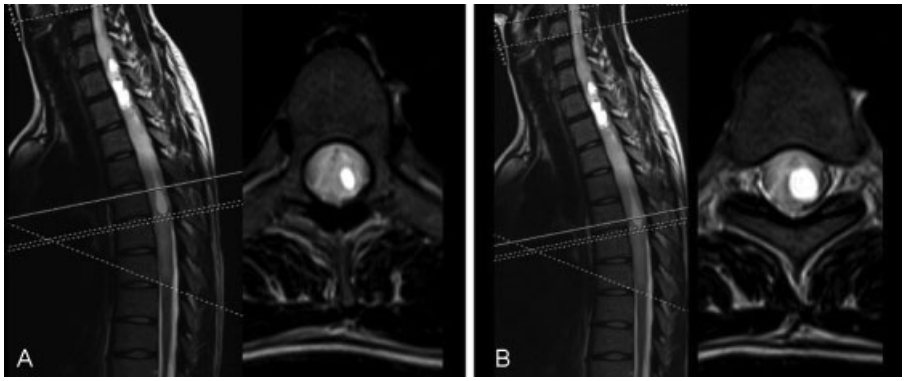

Fig. 4 Eccentrically located cavity in the spinal cord parenchyma: a "true" syrinx. (A) Sagittal T2-weighted magnetic resonance image of intramedullary spinal cord hemangioblastoma from C7 to T2 with extensive rostral and caudal spinal cord hyperintensity indicative of spinal cord edema and teardrop-like eccentric cavity in the spinal cord at the cranial level of T5 (axial T2-weighted image). (B) As in ►**Fig. 4A**, at the caudal level of T5.

dorsal two-third of the spinal cord and filiform in shape (►**Fig. 3**) and not eccentrically located (►**Fig. 4**).

Therapy

As the persistent central canal is considered a normal anatomical variant, this requires no treatment. (There is no etiological cause to be "removed or corrected" and the cavity is usually so small that surgical treatment is not even possible.) Large clinical series have not yet been reported, but initial findings show that these cavities do not change in size in time.[16,17] Therefore as it is a normal anatomical variant, they can be considered an incidental finding, and likewise, follow-up imaging is not warranted. The exact size and extent of these persistent canals is not (yet) known, as increasingly sophisticated MR techniques are enabling an increased visualization of cavities that were not visible before.

In asymptomatic syringomyelia, an expectant policy seems justified, although sometimes this can prove quite difficult especially in those cases where a large syrinx extends all the way up to cervical spinal cord and the brain stem. Nevertheless, it should be noted that spontaneous resolution of syringomyelia has been observed, albeit infrequently.

Nonsurgical treatment of symptomatic syringomyelia focuses either on management of pain and (painful) paresthesias with analgesics, antidepressants, antiepileptic drugs, or gamma-aminobutyric acid-analogs, or on maintaining functional ability and quality of life with physical medicine and rehabilitation.

In patients with neural tube defects, syringomyelia, and symptoms, such as, progressive scoliosis, urological problems, pain, and motor or sensory defects, shunt malfunction should be excluded first, even if this entails empirical shunt revision. If this is not successful, untethering of the cord is usually considered before specific surgical treatment of the syringomyelia. In many cases, untethering leads to improvement in the neurologic deficits. However, one should be aware that a syrinx can appear larger on follow-up MRI after successful untethering.[18]

Patients with tonsillar herniation of less than 5 mm but *normal* CSF flow dynamics on 4D-PC MRI and only mild

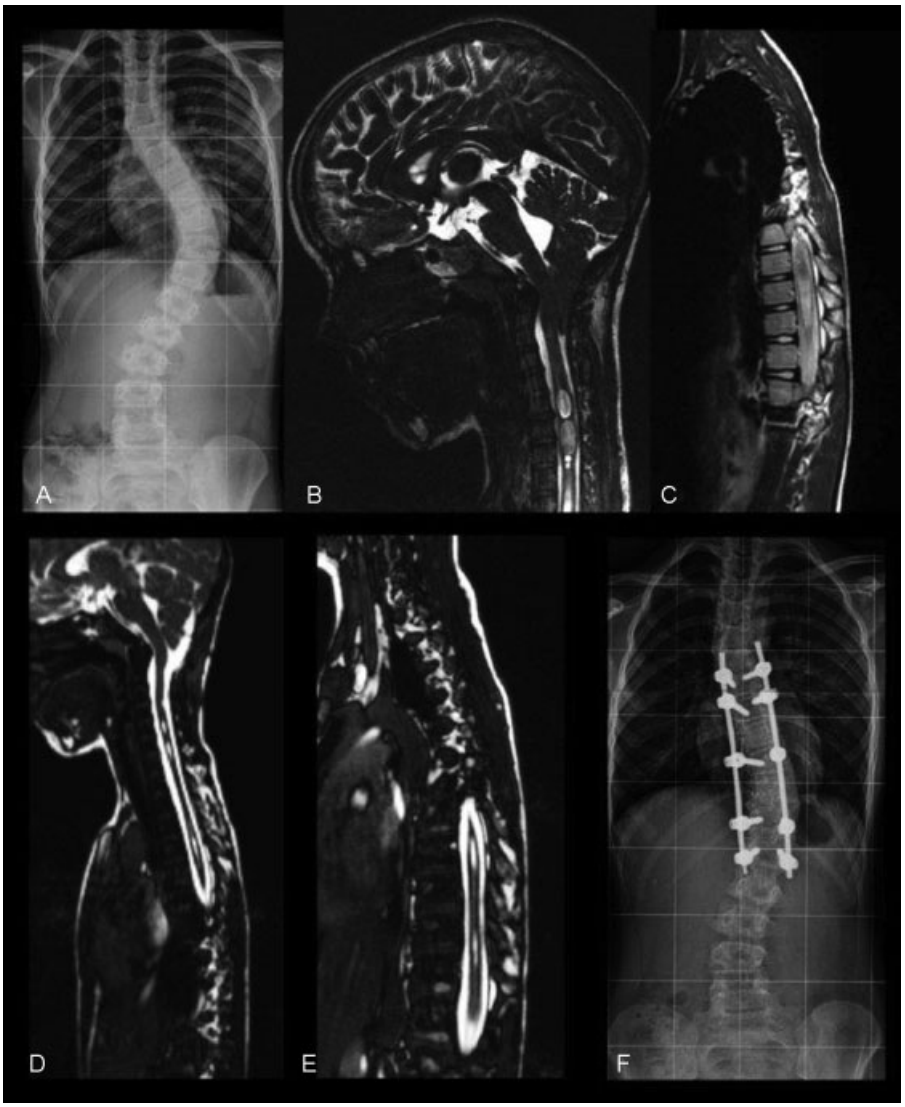

Fig. 5 Syringomyelia, Chiari-I malformation, scoliosis. A 7-year-old girl with headaches, dysesthesia right arm. An analysis for progressive scoliosis revealed Chiari-I malformation with holocord syringomyelia. (A) Preoperative plain X-ray of the total spine showing a severe, left convex scoliosis. (B) Preoperative sagittal three-dimensional constructive interference in steady state magnetic resonance image showing Chiari-I malformation with multiloculated, cervical syringomyelia. Normal ventricles. (C) Preoperative sagittal T2-weighted magnetic resonance image showing caudal part of holocord syrinx. (Because of the severe scoliosis the syringomyelia can only be shown in consecutive panels.) (D–E) Postoperative three-dimensional constructive interference in steady state magnetic resonance images showing adequate craniocervical decompression with collapse of holocord syrinx. (F) Spine fixation was performed later as a separate procedure.

symptoms suggestive of symptomatic syringomyelia are probably best treated conservatively as decompressive surgery in these patients probably will not meaningfully change an already normal CSF flow.

In all other cases, the goal of surgery is restoration of normal CSF flow dynamics. The surgical strategy then varies according to the etiological cause of the syringomyelia. In Chiari-I malformation, a craniocervical decompression will be performed, often combined with an augmentation duraplasty to restore normal craniospinal CSF flow (►**Fig. 5**). Normal fourth ventricle CSF outflow must also be restored, if necessary by placing a fourth ventricular-spinal or ventriculopleural catheter (►**Fig. 6**). In Chiari-II malformation, the foramen magnum is typically enlarged and decompression usually consists of the removal of cervical laminae until the level of the normal spinal cord (depending on the extent of tonsillar descent), section of constricting fibrous bands (usually at the C0–C1 level), and again, normal fourth ventricle CSF outflow must be restored, if necessary, by placing a fourth ventricular-spinal catheter. In the case of spinal canal stenosis, a laminectomy without opening of the dura will suffice to restore normal CSF flow dynamics and subsequent syrinx resolution. Syringomyelia caused by intramedullary or extramedullary tumors or arachnoid cysts is treated by resection of the tumors or fenestration or marsupialization of the cysts.

The most difficult form of syringomyelia to treat surgically is that caused by arachnoid scarring, whether it is caused by infection, inflammation, or trauma. Localized, constrictive bands of fibrosis can be quite safely sectioned (i.e., adhesiolysis), with restoration of CSF flow. But in widespread

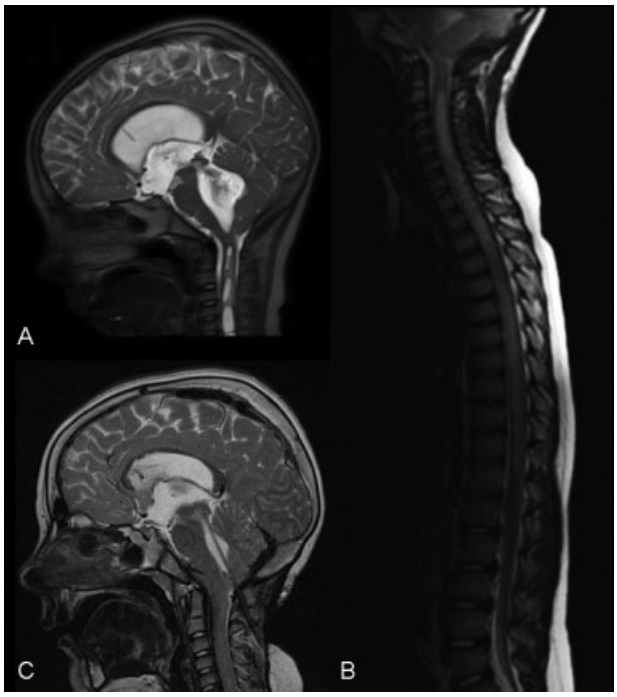

Fig. 6 Holocord syringomyelia and enlarged fourth ventricle. A 6-year-old girl who presented with headaches and a pain in the neck. Previously she had been treated for a meningitis and subdural empyema with eventually a hydrocephalus which was subsequently treated by ventriculo-peritoneal shunting. A holocord syringomyelia was found caused by a progressively enlarged fourth ventricle because of fourth ventricle outlet obstruction of the foramen of Magendie and both foramina of Luschka. (A) Preoperative sagittal T2-weighted magnetic resonance image showing the tip of the ventriculoperitoneal shunt, an enlarged fourth ventricle and syringomyelia extending down to T12. (B) Preoperative sagittal T2-weighted magnetic resonance image showing holocord syrinx from C2 to T12. (C) Postoperative sagittal T2-weighted magnetic resonance image with shunt in fourth ventricle (to pleural cavity), normalization of the fourth ventricle and resolution of the cervical syrinx.

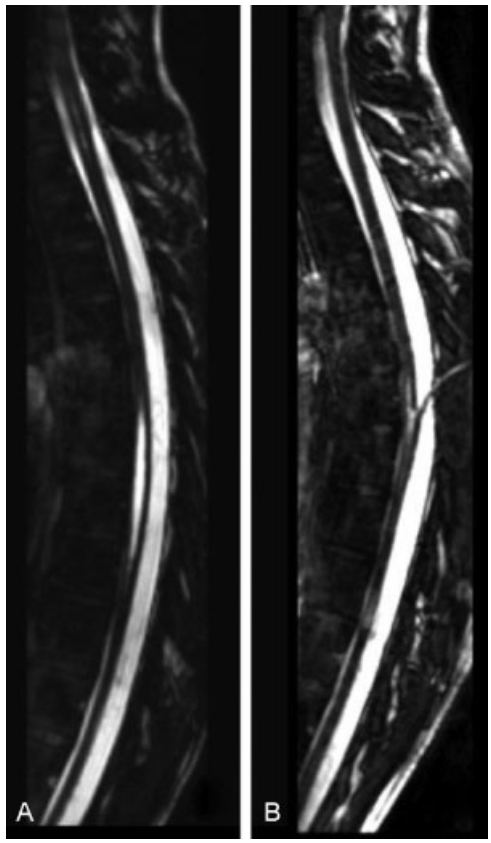

Fig. 7 Cervicothoracic syringomyelia for which no cause could be found. A 13-year-old girl with slowly progressive pain in back, shoulders, and knees. Now and then she drops on the floor because of weakness in both the legs. (A) Preoperative sagittal T2-weighted magnetic resonance image showing a cervicothoracic syrinx which is maximally dilated from T5 to T9. (B) Postoperative sagittal three-dimensional constructive interference in steady state magnetic resonance image showing syringo (pleural) shunt and collapse of the syrinx.

arachnoiditis, the dura, arachnoid, and pia matter can all be completely opaque white and adherent to one another, making it virtually impossible during surgery to distinguish normal anatomical structures, even when using the operating microscope. In these cases, adhesiolysis is not possible and the only remaining option then is direct drainage of the syrinx.

There are three options for direct drainage of a syrinx: syringoarachnoid, syringoperitoneal, and syringopleural. Syringoarachnoid drainage is effective in localized arachnoid scarring, it more or less restores normal CSF dynamics as the CSF is absorbed through normal pathways and obviates the need to open the peritoneum or the pleura, but it is also fraught with shunt obstruction caused by new arachnoid scarring. Syringoperitoneal drainage is analogous to ventriculoperitoneal drainage but, in everyday practice, not very practical, as virtually all patients with syringomyelia are operated on in a prone position, making the abdomen not very accessible. It is reserved for those patients in whom the pleural cavity is not an option. Syringopleural drainage has several advantages: the CSF is drained away from the arachnoid space, which is often compromised by the extensive scarring causing the syrinx in the first place; the pleural cavity is readily accessible in the prone position using a small incision below the right scapula; and the negative intrapleural pressure gently "sucks out" the CSF from the syrinx (►**Fig. 7**).

Complications of Surgery

Besides general complications in neurosurgery such as anesthetic and general complications, infections, and postoperative hematomas, the most prominent problems in syringomyelia surgery are scar formation around the spinal cord and any implanted shunts, wound leakage of CSF, and shunt obstruction. For direct drainage of a syrinx, a micromyelotomy is required. In paraplegic or tetraplegic patients, this is performed below the functional level so no additional neurological deficit will be made. In intact patients, the micromyelotomy will be placed at the level where the posterior part of the spinal cord is thinnest to minimize any additional sensory disturbances from posterior column damage.

Prognosis

Syringomyelia is a chronic, often slowly, progressive disease, with remissions, and exacerbations. Invariably, progressive loss of function will be incapacitating and often this necessitates surgical intervention. Although the progression of neurologic deficits usually stabilizes after intervention, and patients sometimes improve, they often remain symptomatic. Especially, a debilitating central pain syndrome may prove to be quite therapy-resistant, despite adequate surgical therapy and collapse of the syrinx on imaging.

Conclusion

Fluid-filled cavities within the spinal cord can be found on routine MR investigation in asymptomatic subjects, probably represent persistence into adult life of a fetal configuration of the central canal of the spinal cord, and should be regarded as a variant of normal anatomy—the persistent central canal.

In symptomatic patients with syringomyelia (a gliosis-lined cavity within the spinal cord parenchyma), or hydromyelia (a focal dilatation of the central canal), an extensive work-up should be done to find the etiology causing a disturbance in normal CSF dynamics. In shunted patients, a shunt dysfunction should be ruled out first. Treatment should primarily be directed at restoring the normal CSF dynamics rather than at the syrinx itself. Neurologic deficits usually stabilize after surgical intervention, although patients often remain symptomatic. Pain can be a persisting problem.

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