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HOW I DO IT

Syringopleural shunt for refractory syringomyelia: how I do it

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Abstract

Background Surgical treatment of syringomyelia isdirected at the reconstruction of the subarachnoid space and restoration normal cerebrospinal fluid flow. Direct intervention on the syrinx is a rescue procedure and should be offered to patients with refractory syringomyelia.

Methods We provide an overview on indications and technique of syringopleural shunt (SPS). The procedure involves the connection of syrinx with the pleural space using a lumboperitoneal shunt. The occurrence of a negative pressure inside the pleural compartment offers an appropriate gradient for drainage from the syrinx.

Conclusions The SPS allows for a safe and effective treatment of persistent syringomyelia when management of the underlying cause does not yield substantial improvement.

Keywords Chiari malformation · Syringomyelia · Syringopleural shunt

Introduction

Syringomyelia is characterized by the occurrence of cystic cavities inside the spinal cord and can lead progressive injury of the spinal tracts by hydrostatic pressure as the syrinx continues to expand. Syringomyelia is the result of an alteration in physiologic cerebrospinal fluid (CSF) flow dynamics generally induced by Chiari type I malformation (CMI), spinal trauma, craniovertebral junction malformations, arachnoid web, and hydrocephalus. When the etiological treatment directed to restore normal CSF flow fails, directed fluid diversion of the syrinx is indicated [2, 7]. In this scenario, syringopleural shunt (SPS) allows a safe and effective rescue treatment for refractory syringomyelia [1, 3, 7, 9].

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Relevant surgical anatomy

The spinal cord presents a deep anterior median fissure and shallow posterior median and dorsolateral sulci which demarcate the posterior funiculi (or dorsal columns) carrying ipsilateral epicritic sensory information [4]. The dorsolateral columns consist of ascending and descending fibers carrying motor and sensory information. The dorsal rootlets enter the cord in the dorsolateral sulcus. The posterior median septum consists of a thin sheet of pia/arachnoid extending from the median posterior sulcus (PMS) toward the gray commissure of the spinal cord [8]. The anterior spinal artery runs into the anterior median sulcus and gives rise several sulcal arteries and lateral branches. The posterior spinal arteries run on the posterolateral surface of the spinal cord and provide circumferential branches to the pial plexus. The anterior and posterior sulcal veins drains directly into the anterior and posterior spinal veins, respectively. The posterior spinal vein generally runs over the PMS. In these situations, delicate dissection and lateral vein mobilization are required to expose the PMS. In presence of syringomyelia, the spinal cord is enlarged, and the PMS flattened. The microsurgical dissection at the PMS allows safe entry into the syrinx after few millimeters of myelotomy.

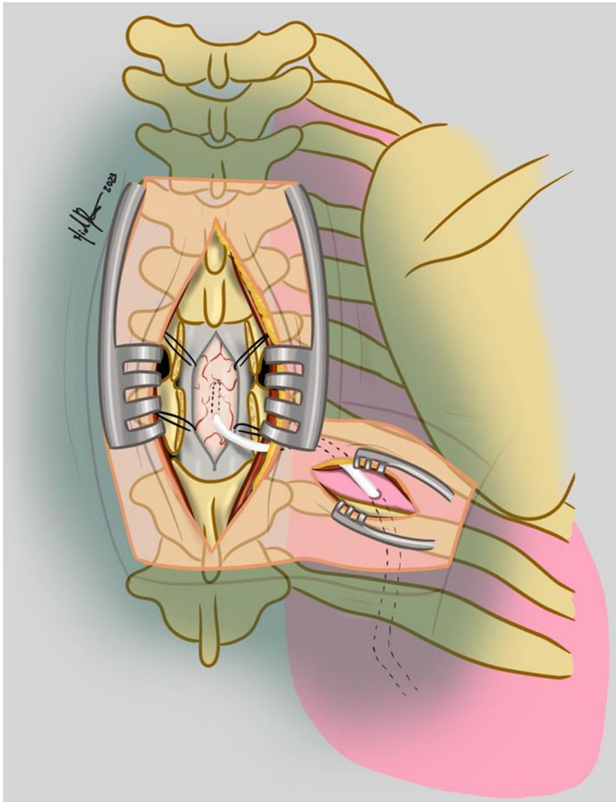

Fig. 1 Original illustration depicting the syringopleural shunt coursing from a thoracic syrinx to the pleural space

Description of the technique

SPS is performed using the one-piece lumboperitoneal shunt (Medtronic, Inc.) with a 0.7-mm inner diameter, 1.5-mm outer diameter, and 87-cm length. The end of the device with multiple perforations is placed inside the syrinx and the opposite end shortened to the appropriate length is placed in the pleural space for 8 to 10 cm (Fig. 1). Short-acting muscle relaxants are used to facilitate intubation and then stopped to allow the use of motor evoked potential (MEP) during the procedure. Under general anesthesia, the patient is placed prone on chest rolls. When the incision site is at the cervical level or cervico-thoracic junction, the head is secured into a Mayfield head clamp and the neck slightly flexed forward. Before the beginning of the procedure, sensory-evoked potentials (SSEPs) and MEPs are recorded. The center of the incision is chosen by analyzing the widest and most caudal portion of the syrinx as demonstrated by the MRI. Fluoroscopic confirmation of the vertebral level with a marker is made. After skin incision and subperiosteal dissection, a standard single-level laminectomy at the thoracic level or two-level laminectomy at the cervical level is made. Under microscopic magnification, the dura is opened with a no. 11 blade and retracted with retention sutures. After arachnoid opening, careful inspection of the enlarged spinal cord allows selection of an area with a paucity of blood vessels along the midline. The spinal cord is incised

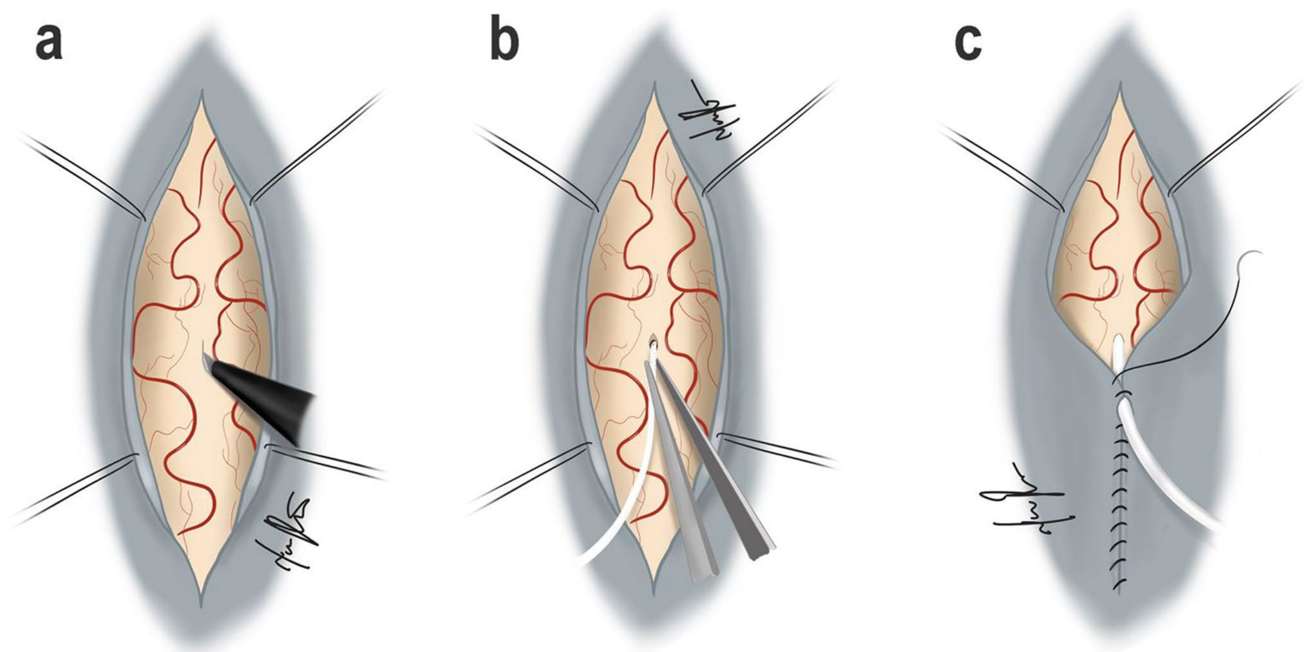

Fig. 2 Original illustrations depicting the syrinx cannulation through keyhole myelotomy. **a** After dural opening, a small midline myelotomy is performed under magnification using a beaver scalpel at a length of 2 to 3 mm and the syringomyelia is entered. **b** The catheter is inserted and directed cranially for at least 3 cm. **c** The dura is sutured with a 5–0 non absorbable suture and the catheter is tunneled to a separate thoracic paramedian incision

with a beaver scalpel at a length of 2 to 3 mm (Fig. 2a). The myelotomy is completed with blunt dissection using a number 7 Rhoton dissector and with bipolar forceps's tips until the syrinx is opened. The catheter is carefully inserted and cranially directed for at least 3 cm (Fig. 2b). The holes of catheter should line only in the syringomyelia cavity to avoid the transmission of pleural negative pressure to the subarachnoid space with resultant low-pressure headache. A 5-0 non absorbable suture is used to close the dura in a watertight fashion (Fig. 2c) and the catheter is tunneled to a separate paramedian incision, which is made at the eighth or ninth intercostal space, below the tip of the scapula which is at T7. Latissimus dorsi and intercostal muscles are incised with a diathermy needle and subperiosteal dissection of the superior margin of the chosen rib is performed to expose the parietal pleura. The parietal pleura is entered via a stab incision and the distal end of the catheter is passed into the pleural space and secured with a purse-string suture. Anchoring suture collars are fastened to the muscles to prevent catheter dislocation.

Indications

SPS is indicated when the etiological treatment of symptomatic syringomyelia directed to restore normal CSF flow fails. The main indication is refractory syringomyelia in patients with CMI who underwent foramen magnum decompression (Fig. 3) [1, 2, 6]. Additional indications are as follows: (1) Post-traumatic syringomyelia with persistent

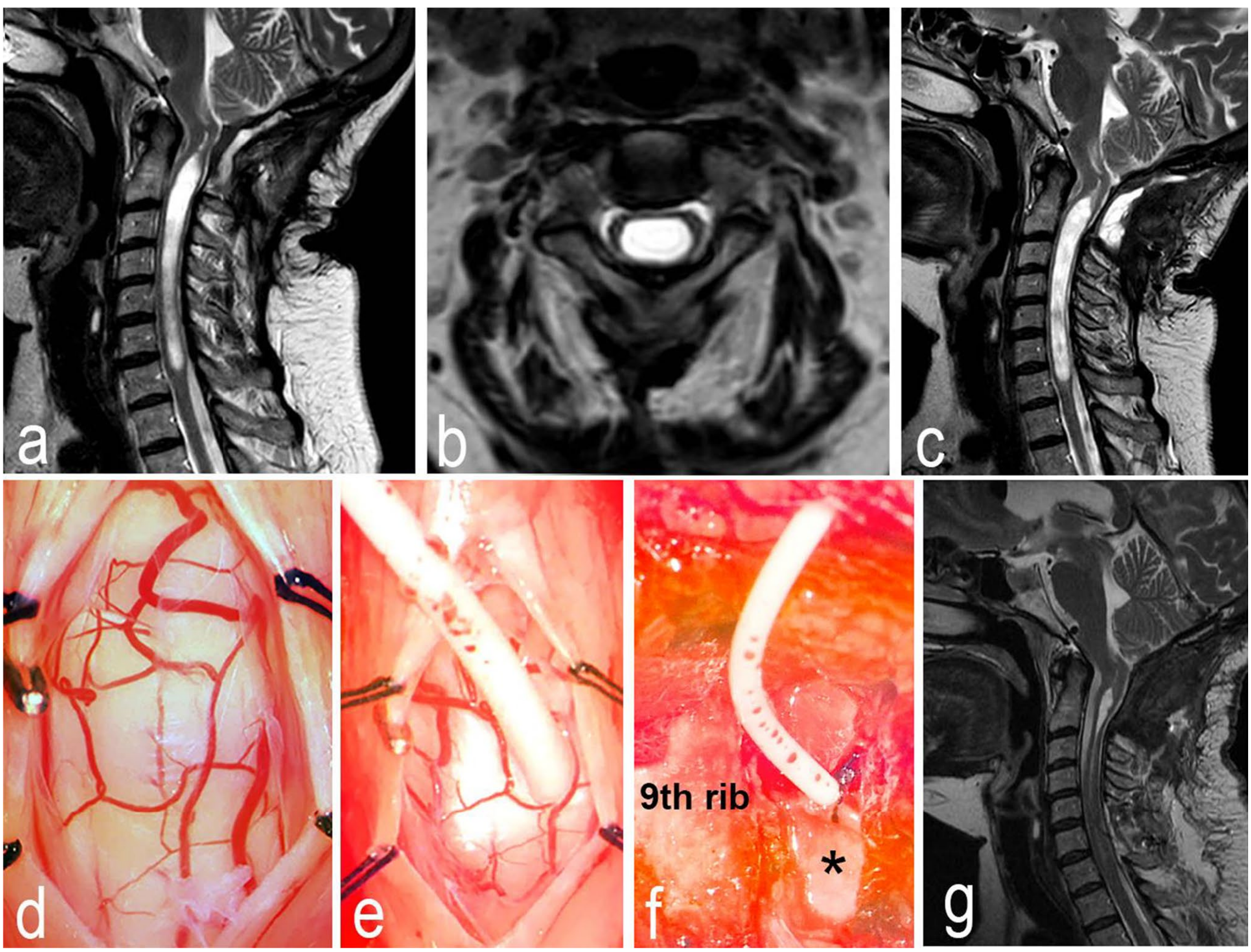

Fig. 3 Persistent cervical syringomyelia requiring syringo-pleural shunt (SPS) in a 57-year-old woman with Chiari type I malformation who previously had foramen magnum decompression, tonsillar coagulation, and duraplasty (FMDD) at another institution. **a**, **b** Sagittal and axial T2-weghted MRI scans demonstrating persistent cervical syringomyelia after FMDD. **c** Sagittal T2-weighted MRI scan obtained after revision of FMDD and adhesiolysis of the arachnoidshowing syringomyelia persistence. **d**, **e** Intraoperative views of SPS. After C5-C6 laminectomy and dural opening, a keyhole myelotomy was performed and the syrinx cannulated. The distal end of the catheter was inserted into the pleural space through the eighth intercostal space (**f**) (*asterisk*: parietal pleura). **g** Postoperative sagittal T2-weighted image demonstrating syrinx shrinkage

canal compromise and/or arachnoid scarring after recalibration of the canal, adhesiolysis of the arachnoid and dural reconstruction. (2) Persistent syringomyelia in patients with craniovertebral junction (CVJ) malformations after CVJ decompression and fusion [1, 5].

Limitations

Prerequisites for SPS are the absence of significant pleural disease or reduced respiratory capacity. Large pleural effusions are exceptional and require shunt revision to syringoperitoneal shunt.

How to avoid complications

Neurologicaldeterioration after SPS occurs in roughly 10% of patients [7]. Intraoperative neurophysiological monitoring using MEP and SSEPs is essential during myelotomy. When SSEPs are lost, the procedure should temporarily have halted allowing SSEPs recover gradually. The pleural entry site should be located below the scapula tip which corresponds to D7 to minimize the occurrence of dislodgment of the catheter.

Specific perioperative considerations

Low molecular weight heparin is administered to all patients as prophylaxis of postoperative thromboembolism. Patients are mobilized within 24 h and a chest x-ray is performed on the first postoperative day to rule out a significant pleural effusion. Patients are discharged on day 4–6 post-operatively and follow-up MRI is performed 8 weeks after surgery.

Specific information to give to the patient about the surgery and potential risk

Patients should be informed that SPS is a rescue procedure and is proposed as last resort for refractory syringomyelia after failure of causal surgical approaches [1]. Neurological complications can occur, which include truncal numbness and pain, and are generally temporary. The occurrence of low-pressure headache can require shunt revision to syringoperitoneal shunt. Clinical improvement after SPS occurs roughly in 70% of patients and shrinkage of syringomyelia is detected in most of the cases [1, 7]. In addition, patients should be aware that shunt dislocation or infection can occur, causing syringomyelia recurrence and requiring shunt revision.

Supplementary Information The online version contains supplementary material available at https://doi.org/10.1007/s00701-023-05654-y.

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Code availability Not applicable.

Declarations

Ethical approval Not applicable

Consent to participate Not required

Consent for publication Not required

Conflict of interest The authors declare no competing interests.

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Key points

1. SPS is indicated in refractory syringomyelia after failure of surgical procedures that address the underlying cause.
2. Syringomyelia persistence after foramen magnum decompression for CMI is the main indication for SPS.
3. The laminectomy site is centered at the widest and most caudal portion of the syrinx as demonstrated by the MRI.
4. Intraoperative neurophysiological monitoring with MEP and SSEPs is required to minimize postoperative neurological complications.
5. Myelotomy should not exceed 3 mm and is performed in an area with a paucity of blood vessels along the midline.
6. The holes of proximal tip catheter should line within syringomyelia cavity to avoid syrinx persistence and low-pressure headache.
7. The paramedian thoracic incision is performed below the tip of the scapula to avoid dislodgment of the catheter.
8. The distal end of the catheter should be shortened to the appropriate length to avoid pleuritic chest pain.
9. Postoperative pleural effusion is a common finding at the postoperative chest X-ray and is generally self-limiting.
10. Postoperative shrinkage of the syrinx is observed at follow-up MRI in the majority of patients.