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HOW I DO IT - PEDIATRIC NEUROSURGERY

Syringo-subarachnoid shunt: how I do it

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Abstract

Background Syringo-subarachnoid shunt (SSS) is a valid method for the treatment of syringomyelia persisting after foramen magnum decompression (FMD) for Chiari I malformation.

Method We give a brief overview on indication and outcome of SSS, followed by a detailed description of the surgical anatomy, and of the microsurgical technique. In particular, we highlight some key points for complication avoidance.

Conclusion SSS is a valid option to treat syringomyelia, since in experienced hands, the outcome is good in most patients, including those with holocord syringomyelia. Careful understanding of anatomy and spinal cord physiology is required to minimize complications.

Keywords Syringo-subarachnoid shunt · Syringomyelia · Chiari I malformation · Spinal cord · Microsurgery · Pediatric neurosurgery · Spinal electrophysiological monitoring

Relevant surgical anatomy

A cross section of the spinal cord (SC) reveals the white matter, consisting of ascending and descending tracts, arranged around the gray matter, consisting of neuronal bodies, unmyelinated motor-neuron fibers, and interneurons. At the posterior and anterior midline, the median posterior sulcus and median anterior fissure, respectively, divide the two sides of the SC. At the region of the posterior median sulcus, small pial arteries fold medially and inward towards the central canal, aiding the surgeon in identifying the midline of the SC. The posterior column of the SC consists of ascending tracts only (fasciculus gracilis and cuneatus) carrying sensory information, while the dorsolateral columns consist of ascending and descending tracts carrying motor and sensory information. Generally, in the posterior part of the gray commissure, interneurons are not found and therefore the blunt opening of this region is a safe passage for reaching the central canal or the syringomyelia cavity.

The SC is supplied with blood mainly through the anterior spinal artery running along the median anterior fissure and the two posterior spinal arteries running laterally along the posterolateral sulcus.

Description of the technique

Under general anesthesia, the patient is settled in prone position and is connected to intraoperative neuromonitoring (motor- and somatosensory-evoked potentials (MEP, SSEP)). To avoid interference with the electrophysiological recordings, inhalation anesthetics should be avoided. IV antibiotics are administered perioperatively. If necessary (especially for limited syrinxes in the thoracic or cervical region) fluoroscopic confirmation of the optimal vertebral level is made, while the shunt is placed at the level with the largest syringomyelia cavity diameter seen on magnetic resonance imaging, preferably below the T1 level, to reduce the risk of injuring the cervical gray matter. After skin disinfection and draping the patient, a midline skin incision is completed (to expose a height of 2 laminas). The muscle fascia is opened and the muscles are

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dissected subperiosteally exposing the desired spinal process and laminae. A standard laminectomy (or laminoplasty) of one or two laminae is performed. Intraoperative ultrasound is used to verify the syringomyelia cavity before dural opening. Under microscopic magnification, the dura is incised in the midline. The arachnoidea is opened and a 6-cm-long 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 "parked" between the dural retraction sutures (Fig. 1). 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 guide the surgeon towards the posterior median sulcus (Fig. 1). Another technique for identifying the midline is by exposing the bilateral nerve roots. The posterior midline sulcus will be at equal distance from both nerve roots. In addition, stimulation of the posterior column using a neurostimulation forceps can be used to better identify the midline. As an alternative, if the syringomyelia is bulging laterally, it can be approached through the dorsal root entry zone (DREZ) (Fig. 2). Therefore, the intraoperative inspection of the cord itself, its midline, whether the syrinx is visible through some part of the cord (e.g., DREZ), is an essential and vital part of the operation. Once the midline is defined, the superficial, small pial vessels are cauterized. The pia is incised using a diamond knife, while small bleedings on the surface can be cauterized using low current. Using a plated bayonet, the cord is bluntly split open until the syrinx is reached and opened (Fig. 1). When the cord seems to be under significant pressure, the opening of the syrinx is completed before inserting the shunt into the subarachnoid space, to avoid the risk of injuring cord tissue. Following syrinx decompression, the proximal tip of the shunt is inserted into the syrinx cavity for 2–3 cm cranially or caudally (Fig. 1). A 6-0 prolene suture is used to fix the shunt to the arachnoidea, to avoid dislocation (Fig. 1). The dura is closed in a watertight fashion using a running suture 5-0 prolene suture. In case of laminoplasty, the laminae are fixed using plates and screws, while in small children, resorbable plates and screws or sutures can be used. The wound is closed in layers in orderly fashion.

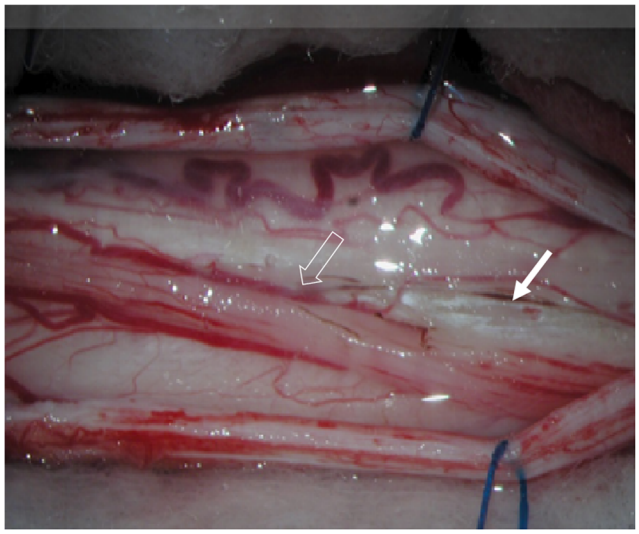

Fig. 2 Intraoperative image showing the insertion of an SSS at the DREZ. Full arrow SSS catheter, empty arrow DREZ

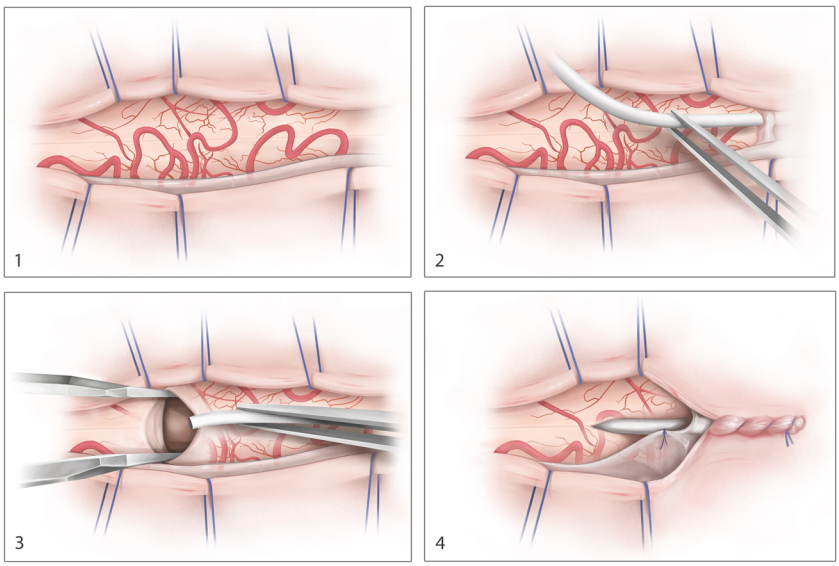

Fig. 1 Illustration of the main surgical steps of SSS. **a** Overview of the SC after dura opening. Small pial arteries fold medially and inward towards the posterior median sulcus aiding the surgeon in identifying the midline. Care should be taken not to open the arachnoid layer during dura opening. **b** After opening the arachnoid layer in the cranial part, the shunt catheter is inserted gently into the subarachnoid space. **c** Using a blunt platelet bayonet, the posterior midline is dissected gently and bluntly until the syrinx cavity is opened. Thereafter, the rostral end of the shunt catheter is inserted into the syrinx cavity. **d** The shunt catheter is sutured to the arachnoid, to avoid shunt dislocation. Watertight dural closure using a running suture

Indications

The main indication for SSS is persisting, progressing, recurring, or new syringomyelia in patients with Chiari I syringomyelia complex who underwent FMD [4]. However, hydrocephalus, regrowth of the posterior fossa bone (especially in young children), tethered cord syndrome, spinal cord tumors, and spinal instability should be ruled out. Even in the absence of symptoms, patients with syringomyelia progression or with very ballooned syringomyelia showing advanced SC compression should be treated, as the functional reserve of the spinal cord is limited, and once symptoms occur, they may be irreversible.

Limitations

Although described by some authors [3], SSS for posttraumatic, postinfectious, or arachnoiditis-associated syringomyelia may be of limited value, as adhesions within the subarachnoid space may limit catheter insertion and CSF flow.

How to avoid complications

Beyond the abovementioned technical nuances, we stress two specific issues: neuromonitoring (MEP, SSEP) are indispensable for SSS surgery and should be routinely used [2]. During the midline myelotomy, SSEPs are often lost and recover gradually. However, if any intraoperative changes in neuromonitoring signals persist and do not recover after a short period of time, the catheter must be repositioned.

The platelet bayonet introduced by Dr. Fred Epstein is an excellent tool for splitting the cord minimizing tissue injury [1].

Specific perioperative considerations

A urine catheter is kept for several days, until opium treatment is stopped. All patients are treated with a patient-controlled analgesia (PCA) device for several days, while toddlers and small children receive continuous opium drip. A soft collar is used for cervical procedures, reducing local pain.

Children or patients with upper cervical syringomyelia are admitted postoperatively to the intensive care unit for the first 24–48 h. Mobilization is commenced on the second postoperative day, while low molecular–weighted heparin for thrombosis prophylaxis is administered in teenagers and adults until full mobilization is achieved. Commonly, patients are discharged on the seventh postoperative day. Follow-up MRI is concluded 3–4 months after surgery and thereafter annually.

Specific information to give the patient about surgery and potential risks

Patients should be informed about the risk of temporary neurological deficits, mostly hypesthesia and gait disturbance, occurring in roughly 33% of the cases [4], and the risk of CSF leak. Urinary and anal sphincter dysfunction, lower cranial nerve palsy, the need for tracheostomy, and the need for spinal fusion due to instability are considered rare complications. In some cases, neurorehabilitation is recommended. It is important to acknowledge that the main goal of surgery is preventing neurological deterioration, although in most cases, some improvement in preoperative neurological function is seen [4]. In addition, the patients should be informed that most postoperative neurological deficits are reversible; however, recovery might take time. Lastly, malfunction or dislocation of the shunt catheter may rarely occur, leading to syringomyelia recurrence and necessitating repeated surgery.

Compliance with ethical standards

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

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Key points

- The main indication of SSS is persistent, progressing, recurrent, or newly developed syringomyelia after FMD decompression for Chiari I malformation.

- Before indicating SSS surgery, hydrocephalus, regrowth of the posterior fossa bone (especially in young children), scoliosis, tethered cord syndrome, spinal cord tumors, and spinal instability should be ruled out.

- Microsurgical technique, accompanied by neuromonitoring are imperative as they help preserve neurological function.

- The identification of the posterior midline (posterior median sulcus) of the SC is essential, in order to avoid damage to the ascending columns of the posterior SC.

- Small pial arteries fold medially towards the posterior median sulcus, aiding the surgeon in identifying the midline.

- The syrinx is best reached through a blunt dissection of the SC posterior midline using a plated bayonet, so that the surrounding neuronal structures are minimally affected.

- As an alternative, if the syringomyelia is bulging laterally, it can be approached through the DREZ.

- The shunt catheter is inserted 2–3 cm into the syrinx cavity and subarachnoid space rostrally and cranially, respectively.

- The shunt catheter has to be long enough (at least 4 cm) and must be sutured to the arachnoid, to lower the risk of shunt dislocation.

- The goal of surgery is to release pressure from the SC and prevent further neurological deterioration.