Syringomyelia — Anatomy and Lesions of the Spinal Cord

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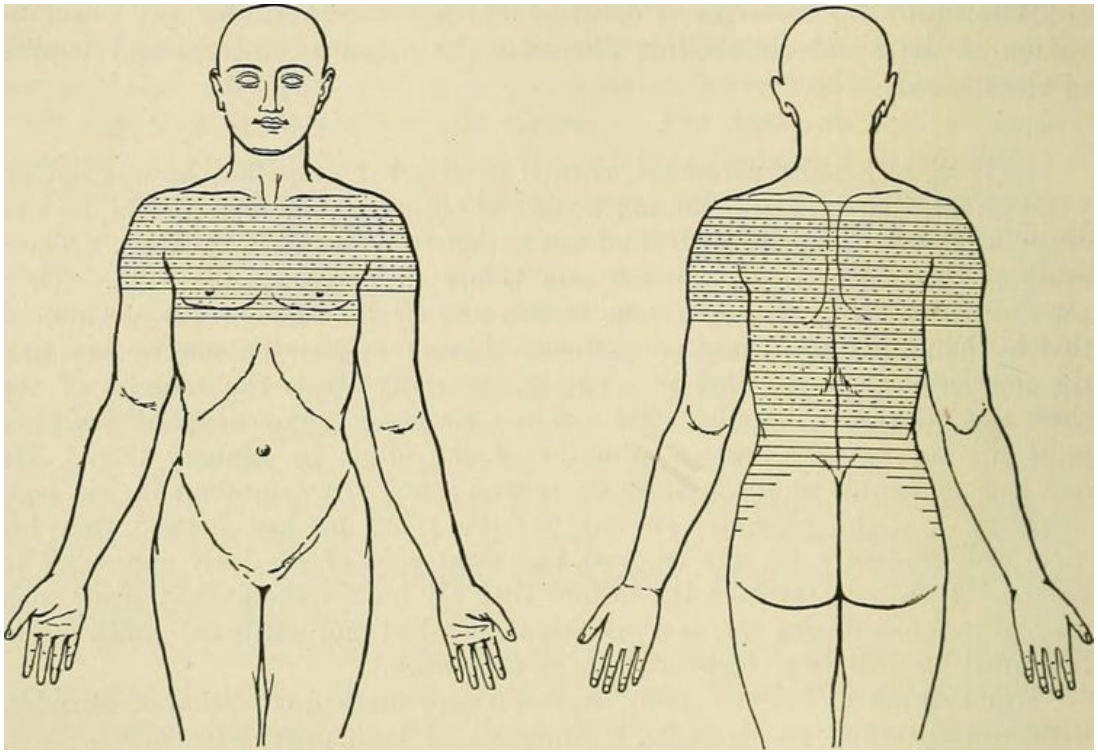
Syringomyelia is characterized by progressive abnormal cerebrospinal fluid (CSF) accumulation in the spinal cord. It causes dissociated suspended anesthesia that affects pain and temperature sensation, predominantly in the upper limbs. Etiology is unclear, although a number of theories have been proposed. The accepted consensus, however, is that syringomyelia is a manifestation of an underlying pathology rather than a disease itself. Treatment of the primary cause often leads to regression of the syrinx.



Definition

The term “syringomyelia” was introduced by Ollivier d’Angers in 1827 to describe cystic cavitations of the spinal cord. However, not every cystic formation of the spinal cord should be described as a syrinx. Syringomyelia is a progressive accumulation of fluid inside the spinal cord. This fluid may be localized inside the parenchyma or the central canal.

Other types of cystic formation of the spinal cord include:

- **Hydromyelia:** syrinx that is confined to the central canal
- **Holo-syrinx:** fluid-filled cavity extending through the entire spinal cord

- **Syringobulbia:** the presence of syrinx in the brainstem

Epidemiology and Pathogenesis

The exact incidence of syringomyelia is unknown, as many patients are **asymptomatic**.

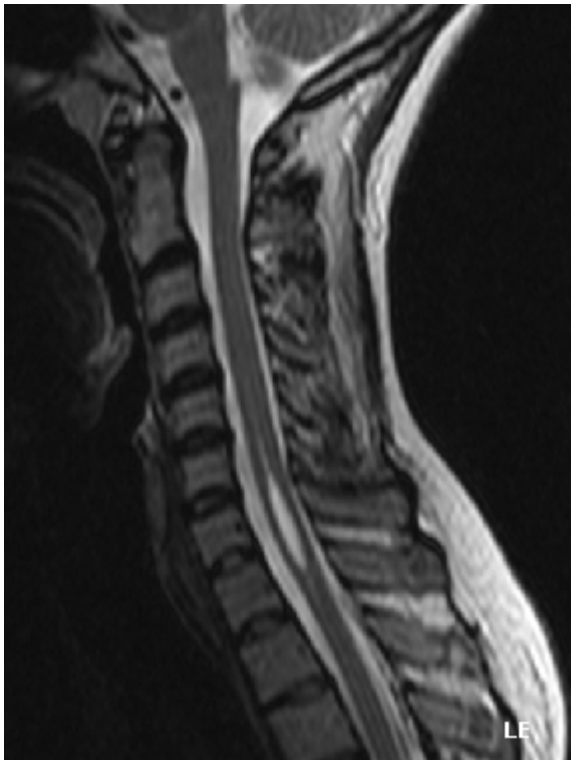


Image: “T2-weighted MR image of a syrinx located at C6-C7 in the cervical spine” by Cyborg Ninja. License: Public Domain

As of now, no single unifying pathophysiological theory unequivocally explains syringomyelia. The following few facts are, however, universally accepted:

- Almost any pathology in the spinal canal or the craniocervical junction can potentially lead to the syrinx.
- The syrinx is usually a response of the spinal cord to 3 common abnormal phenomena: disturbance of cerebrospinal fluid (CSF) flow, spinal cord tethering, or an intramedullary tumor.

Among the many theories proposed to explain syringomyelia, a few prominent ones are as follows:

Theory	Mechanism of syrinx formation
Chiari et al.	Persistence of embryological state
Gardner et al.	Hydrodynamic theory-arterial pulsations lead to cerebrospinal fluid (CSF) accumulation.
Williams et al.	Valsalva maneuvers lead to abnormal CSF accumulation
Oldfield et al.	Systolic pressure changes lead to CSF accumulation.
Milhorat et al.	Production of CSF in segments
Ball and Dyan	Dissection of fluid through Virchow-Robin spaces
Aboulker et al.	Fluid entry along nerve roots

Classification

Syringomyelia is a pathologic response of the spinal cord. It can be primary (congenital) or secondary (acquired). The most common example of primary congenital idiopathic

syringomyelia is Arnold-Chiari malformation (ACM). There are 4 types of ACM:

- **Type I**: syringomyelia with tonsillar herniation greater than 5 mm
- **Type II**: syringomyelia with neural tube defects with hydrocephalus
- **Type III**: occipital encephalocele
- **Type IV**: cerebellar hypoplasia

Acquired syringomyelia is secondary to a known primary cause. The different causes of secondary syringomyelia include:

- Post-traumatic (most common)
- Post-infective (post-tubercular, post-arachnoiditis)
- Secondary to a tumor in the spinal cord or, sometimes, due to intracranial tumor
- Spinal cord tethering and scarring
- Irritation by contrast agents such as metrizamide, which is used in myelography
- Degenerative disc disease (rare)
- After any spinal intervention (secondary to fibrosis and tethering)

Symptoms

As noted, many patients are asymptomatic or minimally symptomatic. For those patients who are symptomatic, the most common symptoms include:

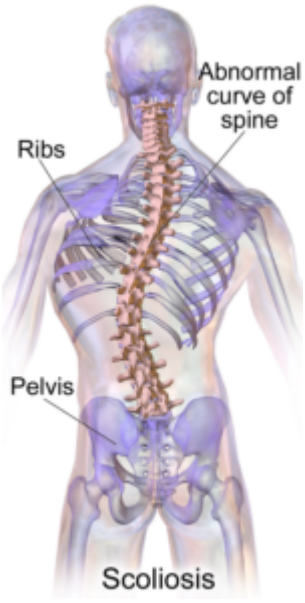


Image: "Scoliosis" by Blausen.com staff. Blausen gallery 2014. License: CC BY 3.0

- Dissociated anesthesia due to the involvement of crossing spinothalamic fibers in the anterior white commissure
- The loss of pain and temperature sensations; posterior column sensations are largely preserved, however. Dysesthesias, paraesthesias, pain, tingling, and numbness are the most common presenting features.
- Suspended anesthesia or preferential involvement of upper limbs in a cape-like distribution due to medially located cervical fibers
- Early involvement of bowel/bladder

- Local destruction of anterior horn cells at the site of the syrinx leading to a lower motor neuron-type of weakness in the involved segments. This symptom is characterized by wasting, weakness, and diminished reflexes.
- Disruption of the local microvascular system leading to ischemia at the watershed levels of the spinal cord. This occurs in the C8–T1 region, with borderline blood supply, and commonly manifests as gross wasting of the small muscles of the hand.
- Further expansion of the syrinx causing the involvement of corticospinal tracts, descending sympathetics, and, lastly, the posterior column.
- Patchy dysesthesias because of asymmetric expansion of the syrinx
- Charcot joints (neuropathic arthropathy) in the shoulder joints due to loss of sensory fibers to the joint
- Scoliosis
- Abnormal abdominal reflexes in children

The table below outlines the major tracts involved and their manifestations:

Tract	Description
Anterior spinothalamic tract decussation in the gray commissure	Loss of pain and temperature sensation in a predominant capelike distribution (involving bilateral upper limb and the neck)
Spinobulbospinal and reticulospinal pathways	Early involvement of bowel and bladder-upper motor neuron type (urgency, frequency, and nocturia)
Late involvement of corticospinal tract	Late motor weakness
Local anterior horn cells at the site of the syrinx	Local lower motor neuron type of weakness
Sparing of posterior columns	Sparing of vibration sense, position sense, and proprioception
Ischemia of the watershed zone at C8–T1	Gross wasting of small muscles of the hand

Progression

If untreated, syringomyelia can spread rapidly. Long-term prognosis is often a culmination of the natural course of the primary pathology.

Diagnosis and Differential Diagnosis

The first symptoms of syringomyelia are related to the primary underlying local pathology. Hence, a detailed, accurate history can often point to the specific investigations needed to reach a diagnosis.

Imaging studies are often the first investigations performed. Magnetic resonance imaging (MRI) of the spine (plain with contrast) is the most common and useful initial investigation. It helps to rule out cystic lesions of the spinal cord and any associated spinal tumors. Lepto-meningeal enhancement can pinpoint infection. Arachnoid scarring can sometimes be seen.

Cardiac gated Cine-MRI flow study is the most sensitive tool currently available to document CSF flow disturbances, as well as CSF flow around the foramen magnum and around potential obstructions. It is also a key way to compare CSF flow changes post-operatively and to document objective improvement in the same.

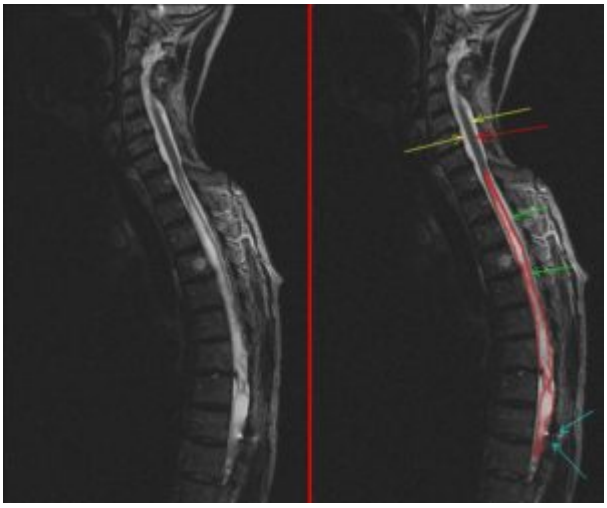


Image: “MRI image of the sagital thoracic spine with syringomyelia” by Lucien Monfils. License: CC BY-SA 3.0

Differential diagnosis

- Intramedullary tumor
- Intramedullary cysts
- Arachnoid cysts
- Myelomalacia
- Glioependymal cysts
- Dilatations of the central canal

Therapy

The fact that each syrinx is a manifestation of underlying pathology has significant repercussions for treatment. Treatment of the primary disease is the gold standard. The presence of syrinx should lead to a detailed and meticulous work-up to find the primary pathology. **Not every syrinx requires treatment, however.** Reversal of the inciting factor leads to regression of the syrinx.

The first causes to look for are intramedullary spinal tumors and spinal dysraphisms leading to tethered cord. Next in line are the causes of disturbed CSF flow, the most common being cranio-cervical junction disorders, Arnold-Chiari malformations, and arachnoid scarring. Treatment is then tailored to the causal factor.

The role of surgery can be summarized in the following manner:

- In patients with intramedullary tumors, the tumor should be removed.
- In patients with an ACM type I malformation, decompression of the foramen magnum is required.
- In patients with arachnoid scarring related to trauma, subarachnoid hemorrhages, infections, or other causes, the arachnoid scar should be resected and CSF flow established.
- In patients with spinal stenosis, the spinal canal must be decompressed.
- In patients with a tethered cord, the tethering must be released.
- In some patients, progressive syringomyelia or re-tethering secondary to multiple spinal surgeries requires direct treatment of the syrinx. A theco-peritoneal shunt is valuable in these patients. Caution should be exercised, as

“once a shunt, always a shunt.” As well, shunt surgery has a high rate of complications. Programmable shunts may improve these rates in the future.

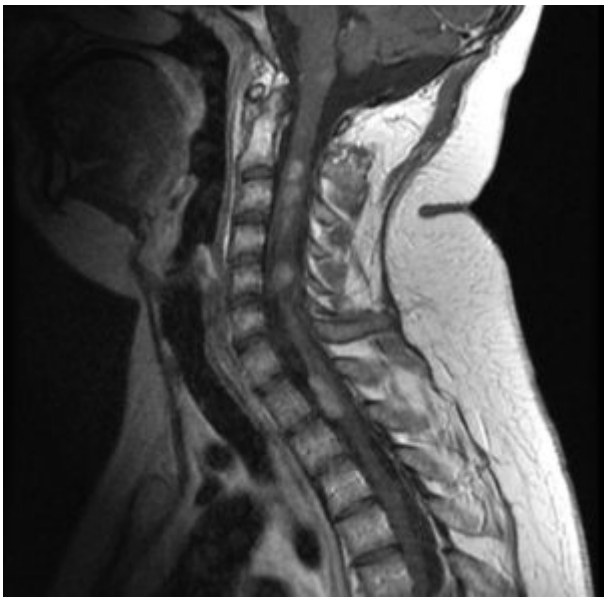


Image: “Multiple ependymomas in a patient with NF2, MRI” by RadsWiki. License: CC BY-SA 3.0

The benefits of other shunt surgeries such as **syringo-peritoneal** and **syringo-subarachnoid shunts** do not clearly outweigh the risks, and failures are not uncommon. Although a number of materials have been used to shunt fluid, the search for the best shunt type, material, and valve is ongoing.

Complications

The progressive nature of syringomyelia necessitates treatment. The complication rate for surgery is about 21%. The most common complication is **CSF fistula formation**. Other potential complications include:

- Hemorrhage
- Arachnoid scarring
- Re-tethering
- Infection
- Re-accumulation of syrinx
- Spinal cord contusion/edema/injury
- Nerve root injury/damage
- Shunt-related complications
- Added postoperative neurological deficits due to the treatment of the primary pathology

Summary

Syringomyelia is not a disease in its own right, but a manifestation of underlying pathology. Symptoms include patchy, dissociated suspended anesthesia and early bowel/bladder involvement with late motor affection and relative preservation of posterior column sensations. Targeted treatment of the primary cause can lead to regression of the syrinx.

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Notes