30 Syringohydromyelia

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INTRODUCTION

The term *syringomyelia* was introduced by the French pathologist and clinician Charles-Prosper Ollivier d'Angers in 1827 to described cystic cavities in the spinal cord.[1] More specifically, *hydromyelia* refers to dilatation of the central canal lined by ependymal cells and *syringomyelia* refers to a cystic cord cavity separate from the central canal not lined by ependymal cells. This distinction has little practical importance as both entities appear similar on imaging and there can be disruption of the ependymal lining as hydromyelia enlarges.[2]

The central canal can be thought of as a cul-de-sac where the only site of macroscopic communication with the subarachnoid space occurs at the obex. The central canal is lined by a single layer of columnar ependymal cells and appears oval in the axial plane (Fig. 30.1). It is positioned slightly ventral of midline in the cervical and thoracic cord, at midline in the lumbar cord, and slightly dorsal of midline at the conus.[3] Animal models suggest that cerebrospinal fluid (CSF) flows in a rostral direction within the central canal.[4] However, dilation caudal to sites of obstruction is often not observed and sites of obstruction have been identified at autopsy in patients without syringohydromyelia.[5] Consequently,

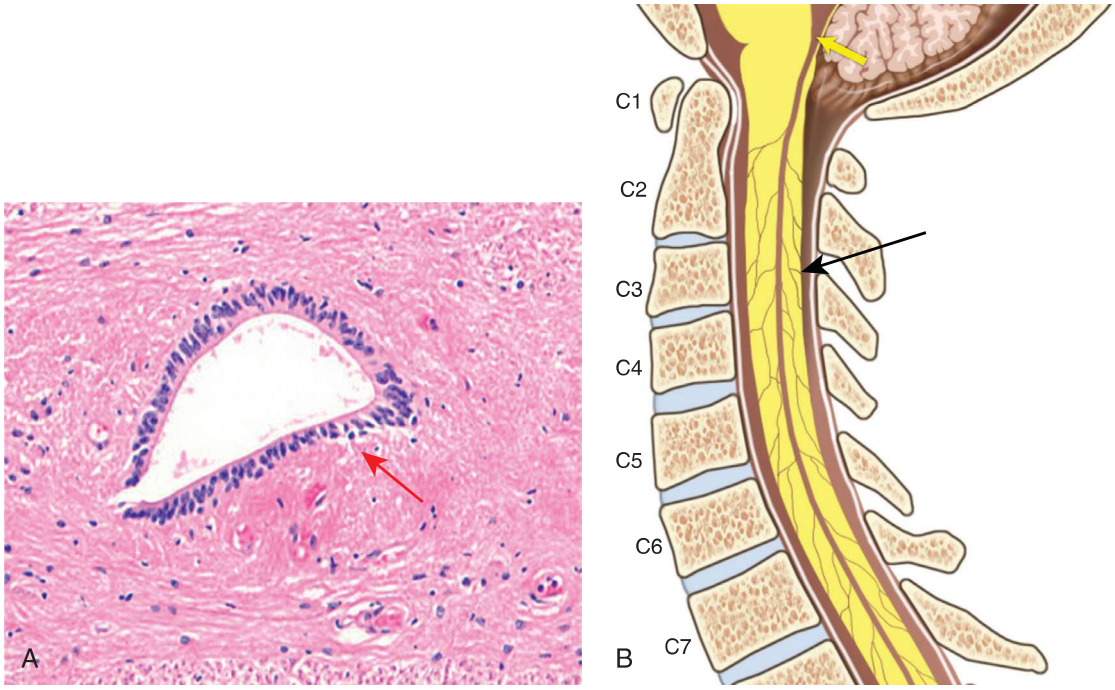

Figure 30.1. Anatomy of the central canal. (A) Hematoxylin and eosin staining of the normal central canal in the axial plane demonstrates the oval shape of the canal, which is lined by a single layer of columnar ependymal cells *(red arrow)*. (B) Sagittal image of the central canal demonstrates its macroscopic communication with the subarachnoid space at the obex *(yellow arrow)*, with theoretical microscopic channels positioned along the length of the cord also providing communication with the subarachnoid space *(black arrow)*. *([A] From Saker E, Henry BM, Tomaszewski KA, et al. The human central canal of the spinal cord: a comprehensive review of its anatomy, embryology, molecular development, variants, and pathology.* Cureus. *2016;8[12]:e927.)*

there is speculation about the existence of microscopic channels positioned along the length of the cord providing communication between the central canal and subarachnoid space that likely need to be obstructed in order for syringohydromyelia to develop.[2]

Syringohydromyelia can be classified into the following four categories based on etiology: hindbrain-related, posttraumatic/inflammatory, tumor-associated, and idiopathic. The incidence of syringohydromyelia ranges from 65% to 80% in Chiari I malformation and 35% in Chiari II malformation.[6] The incidence of posttraumatic syringohydromyelia is 22%, with the interval between injury and diagnosis ranging from 2 months to 34 years.[7,8] Postinflammatory syringohydromyelia has an incidence of less than 1% and is secondary to arachnoiditis or myelitis.[9] The incidence of syringohydromyelia is 4.5% and 16% in patients with multiple sclerosis and neuromyelitis optica, respectively.[10,11] The incidence of tumor-associated syringohydromyelia is 45%, highest with hemangioblastoma and ependymoma, and most commonly located superior to the tumor.[12] Idiopathic syringohydromyelia has a 12% to 28% incidence, often no imaging or clinical progression, and may represent embryologic persistence of the central canal.[1,13,14]

Syringohydromyelia characteristically presents with a dissociative sensory deficit of asymmetric loss of pain and temperature sensation in a cape-like distribution with preservation of vibratory and proprioception.[15] Muscle atrophy, loss of deep reflexes, and scoliosis can be seen.[1] Treatment is aimed at addressing the underlying cause and most commonly involves correcting osseous deformities or lysing adhesions. Catheter drainage is reserved for cases when normal CSF flow dynamics can no longer be restored.

IMAGING: OVERVIEW

Optimal management of syringohydromyelia depends on a timely and accurate diagnosis that requires an understanding of the disease's imaging spectrum.

Syringohydromyelia associated with a Chiari I malformation most commonly involves the lower cervical and upper thoracic cord, demonstrates a multilobulated or haustral contour, and is preceded by a segment of normal cord (Fig. 30.2). Correlation between syringohydromyelia and the degree of tonsillar herniation remains controversial.[16] Syringohydromyelia associated with a Chiari II malformation develops around 4 to 7 years of age, typically after the repair of the myelomeningocele; it often remains stable.[17,18]

Posttraumatic/inflammatory syringohydromyelia is often delayed in its development and extends above and below the level of pathology (Fig. 30.3). An important distinction is to be made with posttraumatic cysts resulting from cord damage that are located at the level of pathology and appear rounded without cord expansion.[1] One large study of tumor-associated syringohydromyelia demonstrated 49% to be positioned above the tumor (Fig. 30.4), 40% both above and below the tumor, and 11% below the tumor.[12]

Idiopathic syringohydromyelia can be conceptually divided into localized and extended forms. Localized syringohydromyelia is less than three vertebral body heights in length and tends to present with milder symptoms and may represent benign enlargement of the central canal. Any central canal diameter greater than 3 mm should be considered abnormal, whereas central canal diameters up to 2 mm within a normal-appearing cord should not be considered abnormal.[2] Extended syringohydromyelia are three or more vertebral body heights in length. A small posterior fossa and increased peak CSF velocities have been reported in idiopathic syrinx.[19,20]

PATHOPHYSIOLOGY

Early theories held that syringohydromyelia was congenital or neoplastic in origin.[21] Modern theories are based on CSF dynamics.

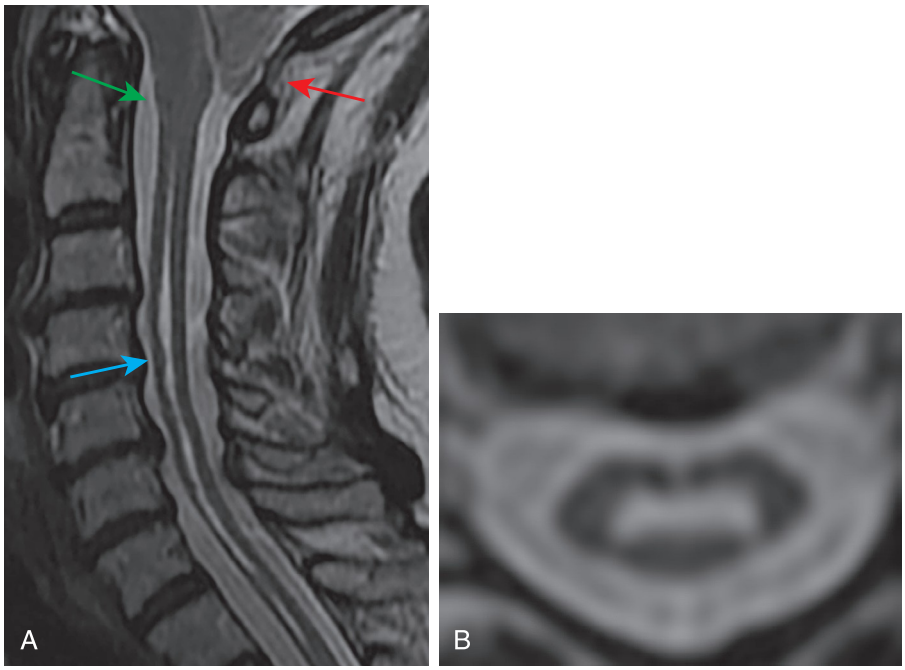

Figure 30.2. Case example of Chiari I syringohydromyelia involving a 67-year-old female who presented with acute shooting pain radiating from her neck into the right parietal region and a chronic low-grade headache. Sagittal T2 (A) and axial T2 (B) images demonstrate a Chiari I malformation (A, *red arrow*) and syringohydromyelia with the characteristic multilobulated or haustral contour *(blue arrow)* that is preceded by a segment of normal cord (A, *green arrow*).

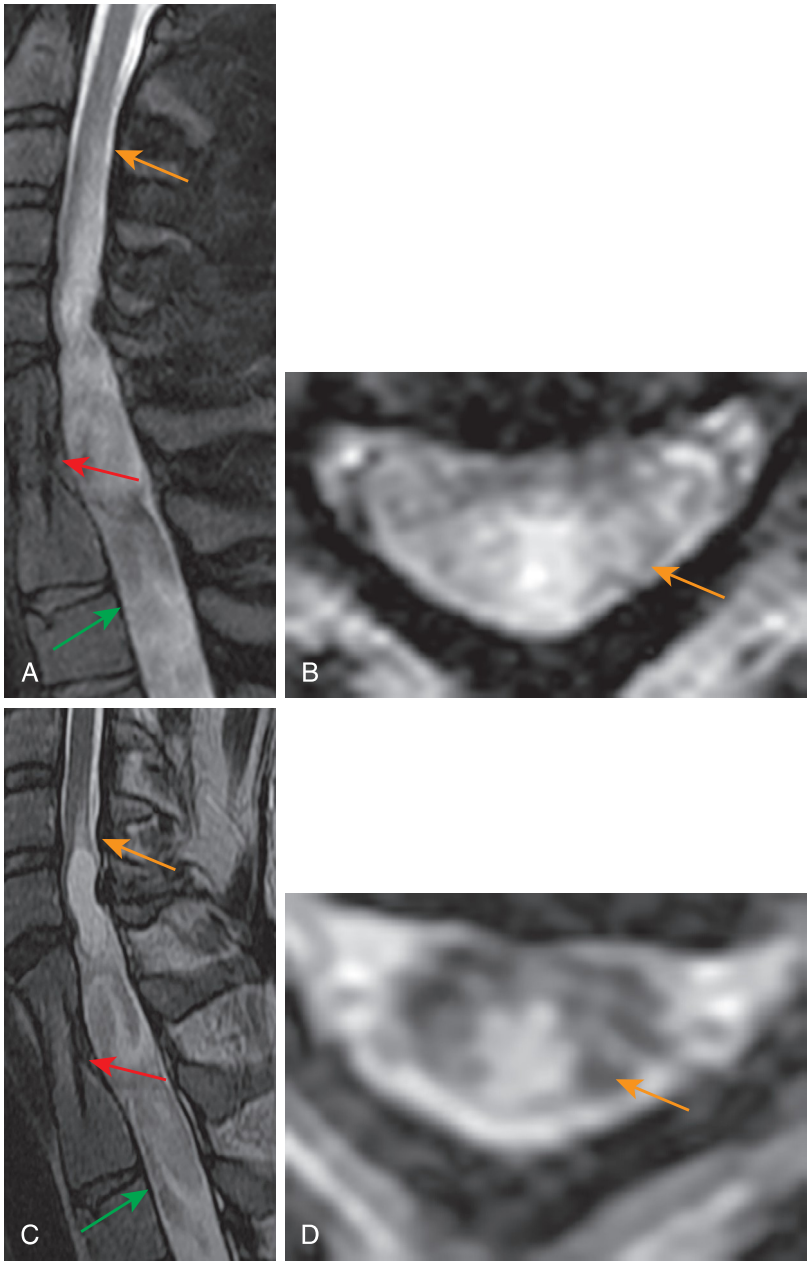

Figure 30.3. Case example of an enlarging posttraumatic syringohydromyelia involving a 44-year-old male with a history of traumatic spinal cord injury. Sagittal T2 (A) and axial T2 (B) images at presentation and sagittal T2 (C) and axial T2 (D) images from 10 years prior demonstrate posttraumatic and postoperative findings *(red arrows)* involving the cervical spine with a large syringohydromyelia characteristically extending above and below the site of injury with cord expansion. Note the dramatic cerebrospinal fluid pulsation artifact *(green arrows)*, likely contributing to the expansion of syringohydromyelia over the 10-year span between the two studies *(orange arrows)*.

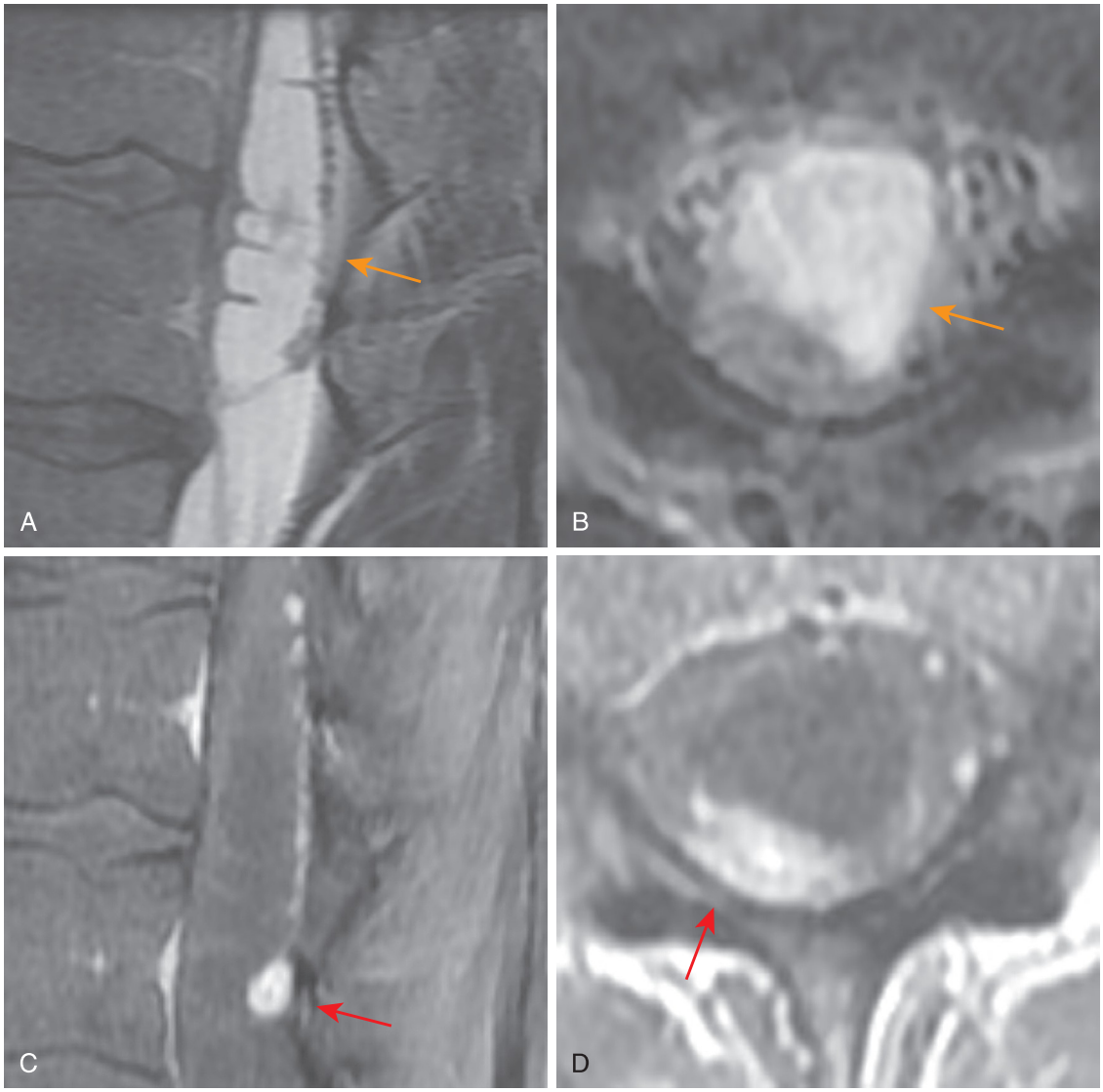

Figure 30.4. Case example of tumor-associated syringohydromyelia involving a 44-year-old male with a history of Von-Hippel Lindau disease. Sagittal T2 (A), axial T2 (B), sagittal T1 postcontrast (C), and axial T1 postcontrast (D) images demonstrate a hemangioblastoma at the L1 level (C and D; *red arrows*) and associated syringohydromyelia (A and B; *orange arrows*) positioned above the tumor.

A hydrodynamic theory for hindbrain-associated syringohydromyelia published in 1958 claims that outflow obstruction at the level of the fourth ventricle forces CSF into the cord leading to syringohydromyelia formation.[22] Skeptics believe that this would instead result in hydrocephalus. A suck-and-slosh theory published in 1980 claims that a pressure gradient across the foramen magnum in Chiari I patients during Valsalva maneuvers creates a "suck" force, driving CSF into the cord and leading to syringohydromyelia formation. CSF movement within the syringohydromyelia creates a "slosh" force, leading to syringohydromyelia enlargement.[23] Skeptics point to a lack of an identifiable communication between the syringohydromyelia and the fourth ventricle. A theory published in 1994 claims that the piston-like downward movement of the cerebellar tonsils during systole creates pressure waves that force CSF into the cord leading to syringohydromyelia formation.[24]

One of the most intriguing aspects of syringohydromyelia pathophysiology is why some enlarge and others remain unchanged. In our experience, posttraumatic syringohydromyelia may enlarge. A recent study evaluating CSF flow dynamics in patients with posttraumatic syringohydromyelia found peak CSF flow to occur earlier in the cardiac cycle compared with controls.[25] The authors speculate that this difference in timing may result in peak CSF pressure occurring before peak arterial pressure, thereby enhancing CSF flow through microscopic channels or even through macroscopic areas of communication at sites of myelomalacia into the cord leading to syringohydromyelia formation. Syringohydromyelia enlargement may then result from internal CSF flow dynamics. A recent cine magnetic resonance imaging (MRI) study demonstrated that during systole there is a collapsing force generated by downward displacement of the cerebellar tonsils on the upper portion of a syrinx forcing CSF inferiorly. During diastole, this pressure gradient is reversed with recoil of the distended lower portion of the syrinx pushing CSF superiorly and reexpanding its upper portion.[26]

DIFFERENTIAL DIAGNOSIS

The major differential diagnosis for syringomyelia includes ventriculus terminalis (Fig. 30.5), a cystic spinal cord tumor, and myelomalacia. Ventriculus terminalis represents a cystic

Figure 30.5. Case example of ventriculus terminalis involving a 59-year-old male with left leg radiculopathy. Sagittal T2 (A), axial T2 (B), sagittal T1 postcontrast (C), and axial T1 postcontrast (D) images demonstrate cystic dilation of the distal central canal (A and B; *red arrows*) without enhancement and normal termination of the conus at T12-L1.

dilation of the distal central canal. The intracystic fluid follows CSF signal intensity on all pulse sequences. Importantly, the walls are smooth without internal septa or enhancement. Additionally, the conus terminates at a normal level without gliosis or myelomalacia. A cystic spinal cord tumor is expansile, with surrounding T2 hyperintensity and enhancement. Myelomalacia demonstrates volume loss and T2 hyperintensity consistent with gliosis.

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