Syringomyelia

Synonyms: ***syringomyelus, hydrosyringomyelia, syringohydromyelia, myelosyringosis, Morvan's disease***

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**Posttraumatic syrinx** → see [p. TrS5 >>](HTTP://WWW.NEUROSURGERYRESIDENT.NET/TrS.%20Spinal%20trauma/TrS5.%20Spinal%20Trauma.pdf#POSTTRAUMATIC_SYRINX)

Epidemiology

* exact incidence unknown but it is rare.
* more frequent in men.

Pathology

- longitudinal (tubular) cavitation in central spinal cord.

**Hydromyelia** - *simple cystic expansion* of central canal of cord.

**Syrinx** - pathologic *tube-shaped cavity* in CNS parenchyma (outside central canal).

* syrinx is *lined* by dense, gliogenous tissue and *surrounded* by dense glial fibril wall.
* syrinx may communicate with central canal (*fluid within syrinx* is similar to, if not identical with CSF).
* most common starting location - *base of posterior horn in cervical cord*.
  + syrinx may be limited to cervical cord or may extend length of neuraxis.
* ***transverse diameter*** varies from segment to segment (usually maximal in cervical and lumbosacral enlargements).
* syrinx slowly enlarges (due to hydrodynamic forces) to involve much of both gray and white matter (at times, only narrow rim of parenchyma is left).
* if syrinx is long, it may have septations but usually (but not always) all cavities communicate.

Etiology

1. **Congenital**

* familial cases have been described.
* rarely occurs in isolation; usually associated with other anomalies:

1. ***Arnold-Chiari malformation*** (60-90%)!!!
2. platybasia
3. atresia of Magendie foramen
4. Dandy-Walker syndrome
5. vascular malformations.
6. Syringomyelia may also be late\* consequence of **spinal trauma**:

\*up to 20 years after trauma

* 1. ex vacuo after absorption of ***intramedullary hematoma***
  2. flexion-extension injury
  3. *cord ischemia* due to hypotension
  4. birth trauma

1. **Tumors** - syrinx may cap rostral or caudal pole of intramedullary (rarely, extramedullary) tumor (e.g. low grade astrocytomas).
2. Chronic adhesive **arachnoiditis** (tuberculosis, post-traumatic, post-surgery) → impaired CSF circulation.

Pathogenesis

**Communicating**\* **syringomyelia (s. hydromyelia)** – most cases of syringomyelia!

\*syrinx is part of ventricular system

1. **hydrodynamic Gardner's theory**: *occluded exit foramina of fourth ventricle* (e.g. by developmental defect in rhombic roof) + CSF pulsations directed downward → dilatation of central canal ± hydrocephalus.
2. **Williams** **theory**: *ball-valve effect of cerebellar tonsils* in Arnold-Chiari malformation (central canal spinal constriction at level of foramen magnum) → during Valsalva maneuver CSF may pass caudad through narrowed canal → syrinx formation; foramen magnum decompression usually results in syrinx resolution.

**Noncommunicating syringomyelia**

1. extension of CSF under pressure along ***Virchow-Robin spaces***
2. cystic degeneration of ***intramedullary tumor***
3. ischemia in anterior spinal ***artery insufficiency***.
4. resorption of ***intramedullary hematoma*** (hematomyelia)
5. ***cord contusion / compression*** → microcystic cavitation.
6. ***obliterated*** original communication with ventricular system.

Clinical Features

Manifests in 3-4th decade (mean age at onset ≈ 30 years) - chronic, slowly progressive **central cord syndrome**. [see p. Spin1 >>](http://www.neurosurgeryresident.net/Spin.%20Spinal%20Disorders\Spin1.%20GENERAL%20-%20Spinal%20Syndromes.pdf#Central_Cord_Syndrome)

* usually located in ***cervical ÷ upper thoracic*** segments (rarely, syrinx develops in lumbar cord either in association with or independent of cervical syrinx).

Damage to decussating spinothalamic fibers → segmental loss of pain and temperature sensations (**analgesia with thermoanesthesia**):

* bilateral, frequently asymmetrical.
* over shoulders (*“cape” distribution*) or across shoulders and upper torso, front and back in shawl-like distribution (*en cuirasse*).
* deep and aching **chronic pain** (≈ 30%) in impaired segments.
* complications - painless hand ulcers, burns, and whitlows.

Damage to anterior horns → segmental LMN signs (**areflexia**, **weakness**, **atrophy**, **fasciculations**):

* intrinsic hand muscles are often affected first → striking early hand atrophy - ***claw-hand deformity*** (*main en griffe*).
* ascends to forearms, and ultimately affects shoulder girdle.
* complications:
  1. thoracic ***scoliosis*** (thoracic LMN innervating paraspinal musculature).
  2. ***neurogenic arthropathies*** (Charcot joints) in shoulder, elbow, wrist.

Classic vignette: cervical syrinx → Charcot shoulder

As syrinx size increases, other spinal structures can be involved. [see p. Spin1 >>](http://www.neurosurgeryresident.net/Spin.%20Spinal%20Disorders\Spin1.%20GENERAL%20-%20Spinal%20Syndromes.pdf#Central_Cord_Syndrome)

1. *dorsal columns*
2. *corticospinal pathways* → spastic leg weakness.
3. *intermediolateral columns*; e.g. hands may develop remarkable subcutaneous edema and hyperhidrosis (*main succulente*).

* spontaneous arrests for several years are not uncommon.
* violent cough or sneeze may produce hemorrhage into syrinx.

Syringobulbia

Syrinx may expand into ***brainstem*** - into medulla oblongata or even into pons:

N.B. only 10% syringes extend above C2

* **CN9-12** may be involved (**bulbar palsy**), usually asymmetrically.
* nuclear involvement of **CN5** → facial pain and thermal hypesthesia in *onion skin pattern*.
* nystagmus is not rare.
* rarely, syrinx extends into centrum semiovale (syringocephalus).

Diagnosis

**CSF** - few abnormalities (CSF pressure may be elevated, cell count rarely > 10/mm3, mild elevation of CSF protein).

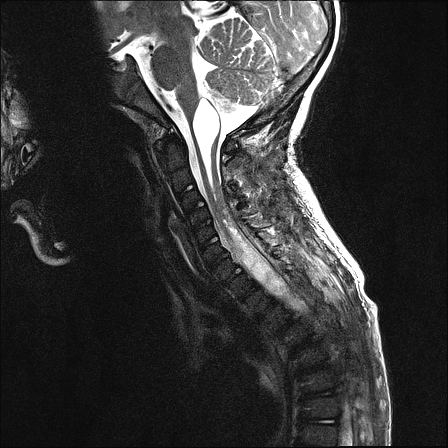
**MRI** - diagnostic procedure of choice! (syringomyelia, and its cause, are shown well)

N.B. no correlation between clinical severity and size of syrinx relative to remaining cord substance!

* signal intensity of cyst ≈ CSF.
* cyst margins often irregular (may demonstrate periodic folds or septations - result from turbulent flow within cavity).
* further MRI evaluation:
  1. brain (hydrocephalus?)
  2. craniovertebral junction (Arnold-Chiari malformation?).

N.B. if syringomyelia occurs without *Arnold-Chiari malformation* or *prior spinal cord injury* → *complete spinal MRI with gadolinium* (to rule out intramedullary tumor).

**MRI-T2**: syringobulbia due to cervicothoracic pilocytic astrocytoma in 4 yo female:

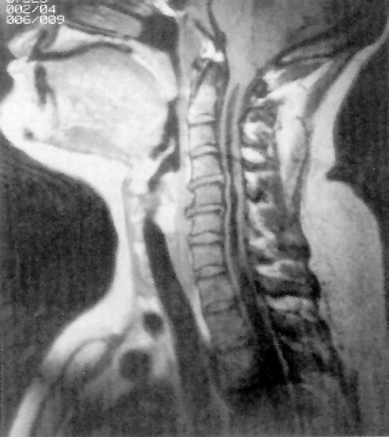


[Source of picture: Viktoras Palys, MD >>](mailto:vpalys@vcu.edu)

**MRI-T1**: large syrinx + associated Arnold-Chiari malformation (cerebellar tonsillar herniation below foramen magnum):



**MRI**: dilated central spinal canal + pointed cerebellar tonsils (Chiari I malformation):

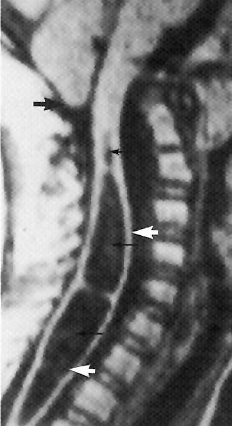


**MRI** (syringomyelia + Chiari malformation):  
**A.** MRI-T1: descent of cerebellar tonsils and vermis below level of foramen magnum (*black arrows*); CSF collection dilates central canal (*white arrows*).

**B.** MRI-T2: high-signal-intensity syringomyelia (*white arrows*) expanding cervical cord with signal intensity equivalent to CSF.



**T1-MRI** (syringohydromyelia + Chiari I malformation): cerebellar tonsils (upper thick black arrow) lie below neural arch of C1, and distended syrinx (*white and black arrows*) extends from C2 to T2:



###### Myelography followed by delayed CT.

* spinal cord is:

enlarged (fusiform expansion) in 80%;

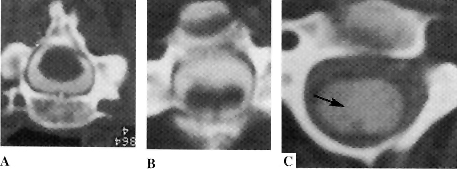
normal in 10%;

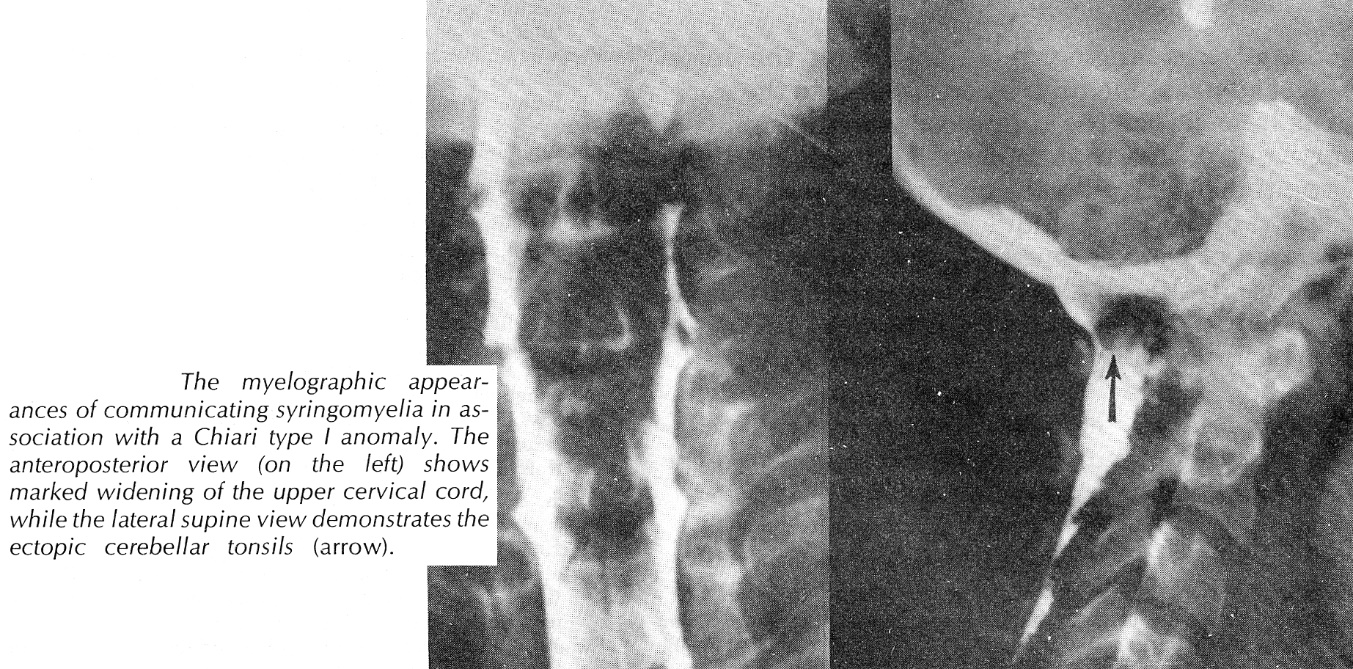
small (flattened in sagittal plane) in 10%.

* rotating patient from prone to supine position → *considerable changes in cord size and shape*.
* contrast medium enters all intramedullary cavities (most likely 6–12 h after myelography).

Patient prone (A) and supine (B) - fluctuation in cyst size (low radiodensity) with posture.

(C) patient supine 8 h after myelogram - contrast penetration and retention in syrinx cavity (*arrow*).





Differential diagnosis

- all easily excluded with MRI:

1. intramedullary tumors.
2. ALS
3. MS
4. cervical spondylosis
5. anomalies of craniovertebral junction

Treatment

Incidental syrinx (i.e. asymptomatic and no neurologic deficit) with no identified etiology → **observation** with MRI at 2-3 year intervals (if the size remains stable).

Symptomatic / enlarging syrinx → surgery.

* 1. First choice - **correction of abnormal CSF dynamics**:
     + - tumor: ***mass removal*** → syrinx resolution.
       - Arnold-Chiari malformation:
         1. hydrocephalus is present → ***ventriculoperitoneal shunting***.
         2. no hydrocephalus (or ineffective ventriculoperitoneal shunt) → ***posterior fossa decompression***, ± simultaneous syrinx shunting.
  2. Second choice - **drainage of syrinx cavity** when etiology cannot be easily addressed (e.g. in arachnoiditis, post-traumatic syrinx): [see p. Op230 >>](HTTP://WWW.NEUROSURGERYRESIDENT.NET/Op.%20Operative%20Techniques/200-299.%20Spine/Op230.%20Syringomyelia%20(techniques).pdf)
     1. **temporary** – ***percutaneous needle aspiration*** (can be repeated) or ***open syringotomy***.
     2. **permanent** – various ***shunts*** (***syringopleural, syringoperitoneal, syringo-subarachnoid***).

Bibliography for ch. “Spinal Disorders” → follow this [link >>](http://www.neurosurgeryresident.net/Spin.%20Spinal%20Disorders\Spin.%20Bibliography.pdf)

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