

# Syringomyelia

Synonyms: *SYRINGOMYELUS*, *HYDROSYRINGOMYELIA*, *SYRINGOHYDROMYELIA*, *MYELOSYRINGOSIS*, *MORVAN'S DISEASE*

Last updated: September 5, 2017

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Posttraumatic syrinx → see p. TrS5 >>	

## 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

- 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.
- Syringomyelia may also be late\* consequence of **spinal trauma**:
  - a) ex vacuo after absorption of *intramedullary hematoma*
  - b) flexion-extension injury
  - c) *cord ischemia* due to hypotension
  - d) birth trauma

\*up to 20 years after trauma
- Tumors** - syrinx may cap rostral or caudal pole of intramedullary (rarely, extramedullary) tumor (e.g. low grade astrocytomas).
- 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

- 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.
- 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**

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

## CLINICAL FEATURES

Manifests in 3-4<sup>th</sup> decade (mean age at onset ≈ 30 years) - chronic, slowly progressive **CENTRAL CORD syndrome**. see p. Spin1 >>

- 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 >>

- 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 C<sub>2</sub>

- **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/mm<sup>3</sup>, mild elevation of CSF protein).

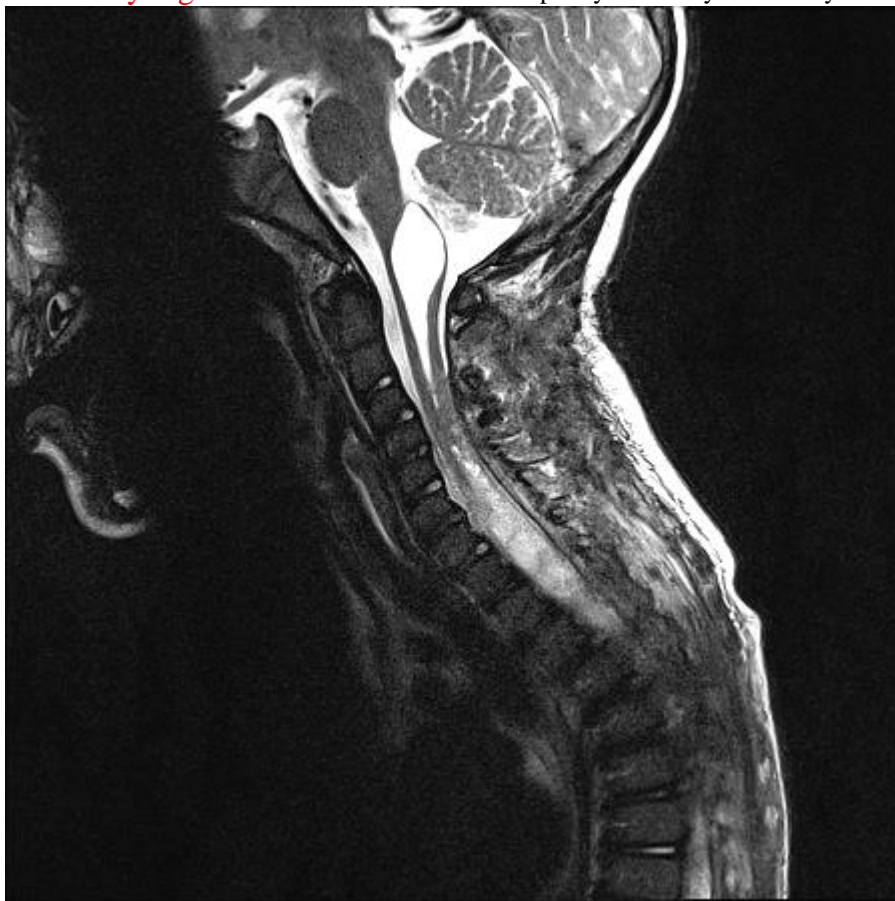
**MRI** - diagnostic procedure of choice! (syringomyelia, and its cause, are shown well)

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:



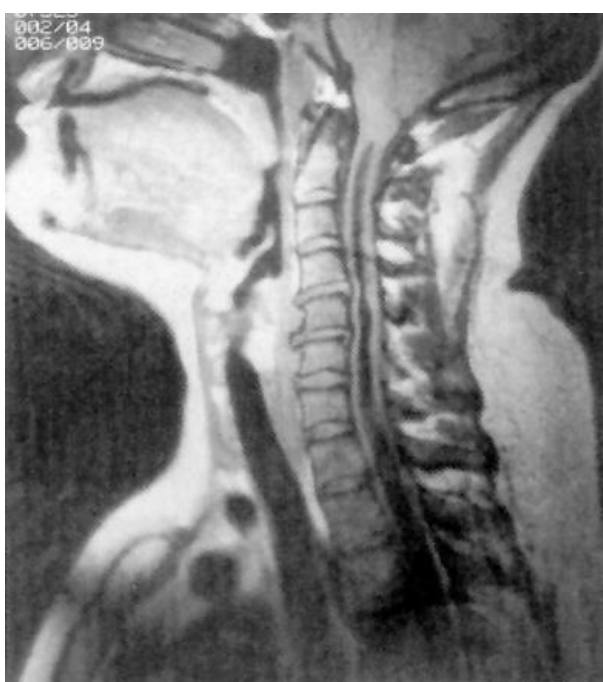
Source of picture: Viktoras Palys, MD >>

**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):





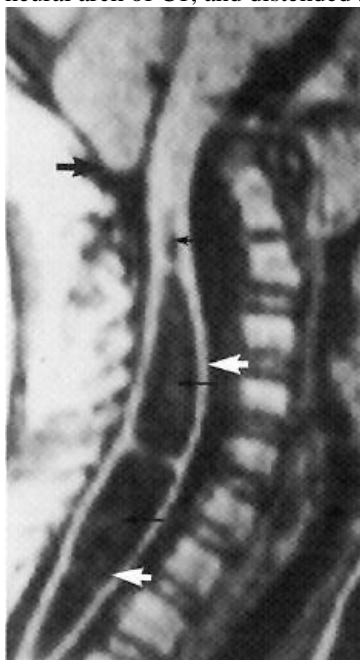
**MRI (syringomyelia + Chiari malformation):**

**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.



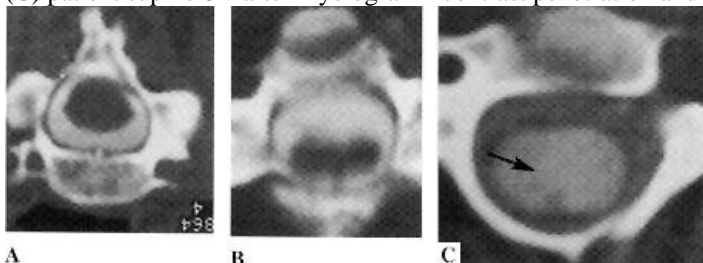
**T1-MRI (syringohydromyelia + Chiari I malformation):** cerebellar tonsils (upper thick black arrow) lie below neural arch of C1, and distended syrinx (*white and black arrows*) extends from C2 to T2:



**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*).



The myelographic appearances of communicating syringomyelia in association with a Chiari type I anomaly. The anteroposterior C1 (on the left) shows marked widening of the upper cervical cord, while the lateral supine view demonstrates the ectopic cerebellar tonsils (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.

A. First choice - **correction of abnormal CSF dynamics**:

- **tumor**: *mass removal* → syrinx resolution.
- **Arnold-Chiari malformation**:
  - a) hydrocephalus is present → *ventriculoperitoneal shunting*.
  - b) no hydrocephalus (or ineffective ventriculoperitoneal shunt) → *posterior fossa decompression*, ± simultaneous syrinx shunting.

B. Second choice - **drainage of syrinx cavity** when etiology cannot be easily addressed (e.g. in **arachnoiditis**, **post-traumatic** syrinx): see p. Op230 >>

- a) **temporary** – *percutaneous needle aspiration* (can be repeated) or *open syringotomy*.
- b) **permanent** – various *shunts* (*syringopleural, syringoperitoneal, syringo-subarachnoid*).

**BIBLIOGRAPHY** for ch. “Spinal Disorders” → follow this [LINK >>](#)