Syringomyelia

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

Last updated: April 22, 2019

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

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.

2. Syringomyelia may also be late* consequence of **spinal trauma**:

*up to 20 years after trauma

   a) ex vacuo after absorption of ***intramedullary hematoma***
   b) flexion-extension injury
   c) *cord ischemia* due to hypotension
   d) birth trauma

3. **Tumors** - syrinx may cap rostral or caudal pole of intramedullary (rarely, extramedullary) tumor (e.g. low grade astrocytomas).

4. 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

A) **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.
B) **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

A) extension of CSF under pressure along ***Virchow-Robin spaces***
B) cystic degeneration of ***intramedullary tumor***
C) ischemia in anterior spinal ***artery insufficiency***.
D) resorption of ***intramedullary hematoma*** (hematomyelia)
E) ***cord contusion / compression*** → microcystic cavitation.
F) ***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 >>

- 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_2

- **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^3, mild elevation of CSF protein).

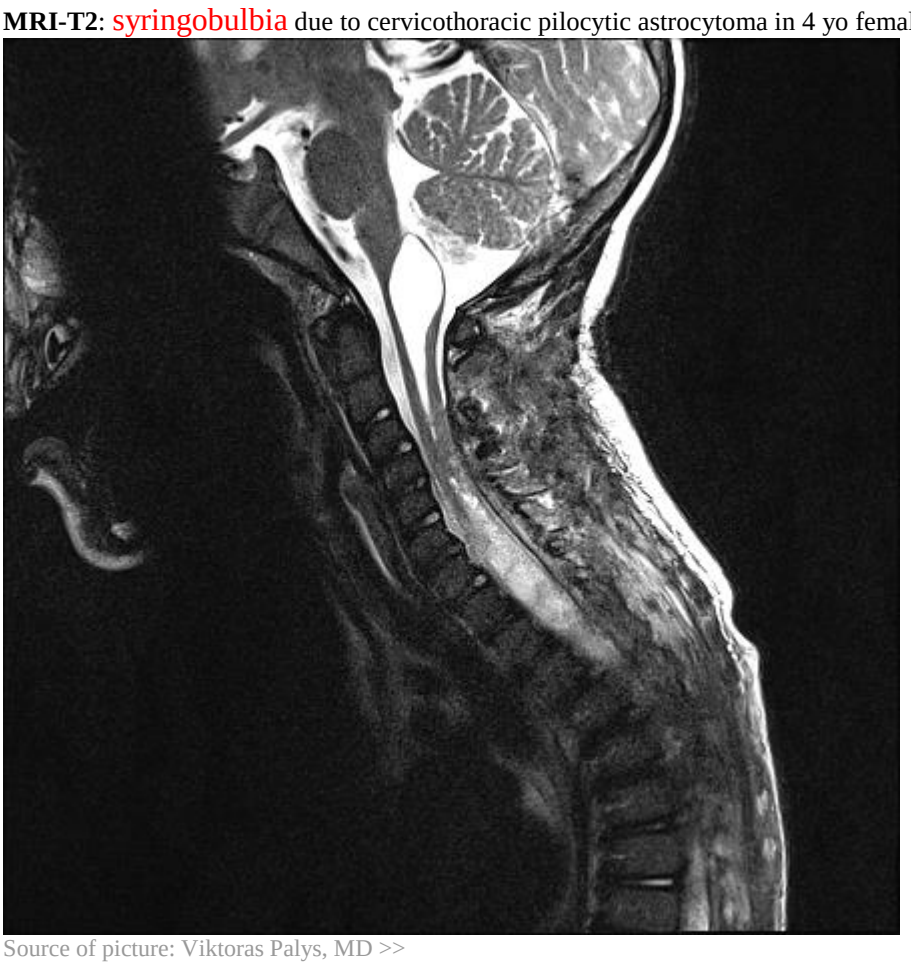
MRI - diagnostic procedure of choice! (syringomyelia, and its cause, are shown well)

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?).

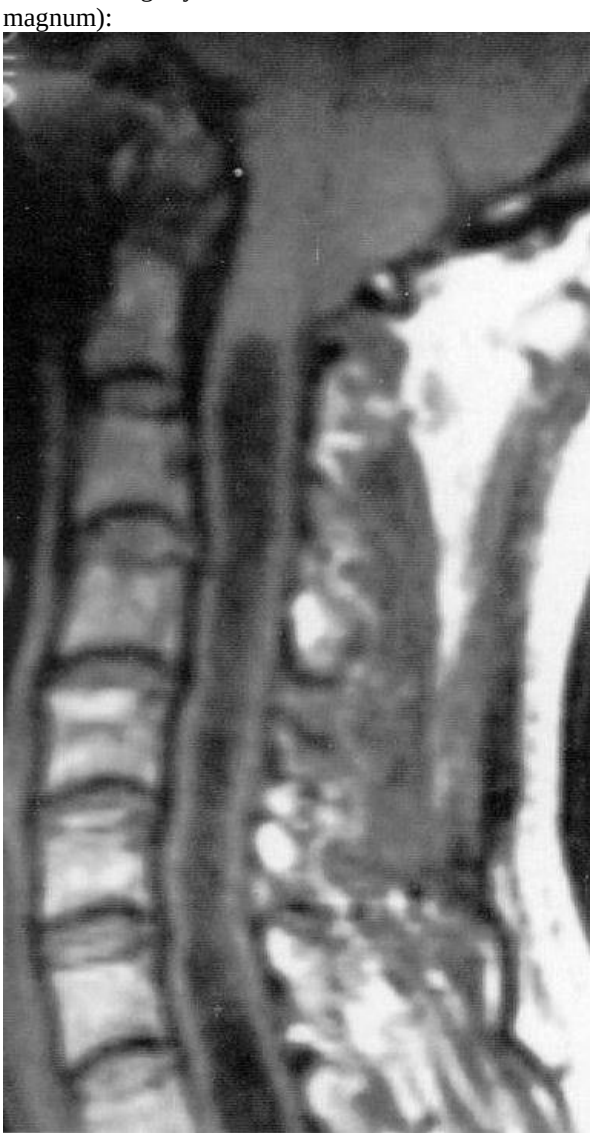
N.B. if syringomyelia occurs without *Arnold-Chiari malformation* or *prior spinal cord injury* → *complete spinal MRI with gadolinium* (to rule out intramedullary tumor).

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

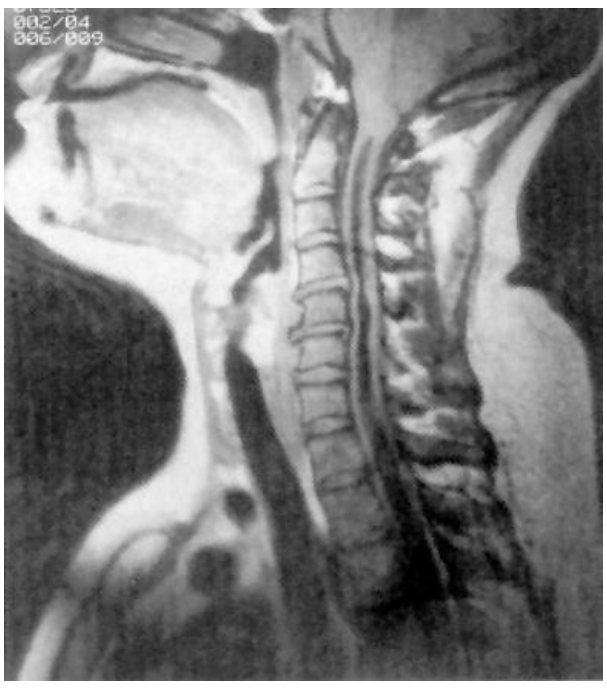


Source of picture: Viktoras Palys, MD >>

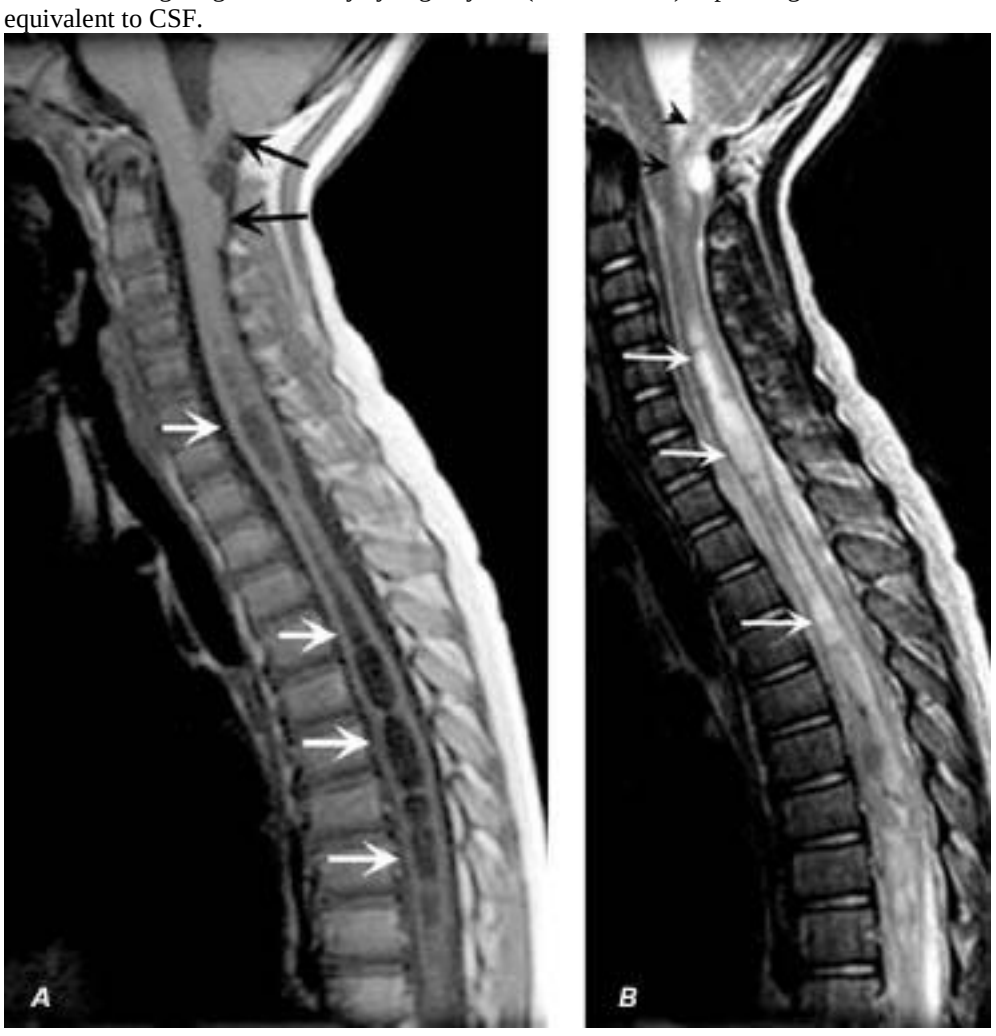
MRI-T1: large syrinx + associated Arnold-Chiari malformation (cerebellar tonsillar herniation below foramen magnum):



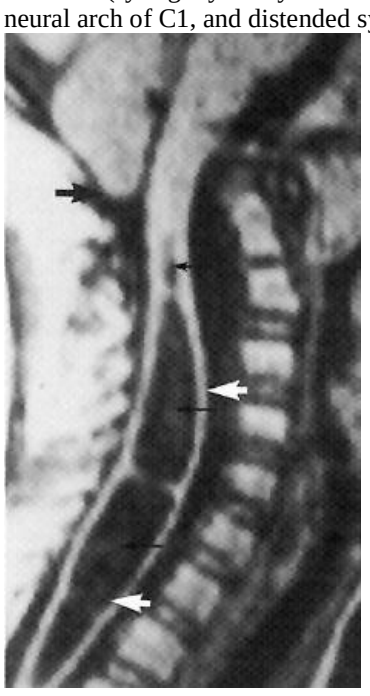
MRI: dilated central spinal canal + pointed cerebellar tonsils (Chiari I malformation):



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.



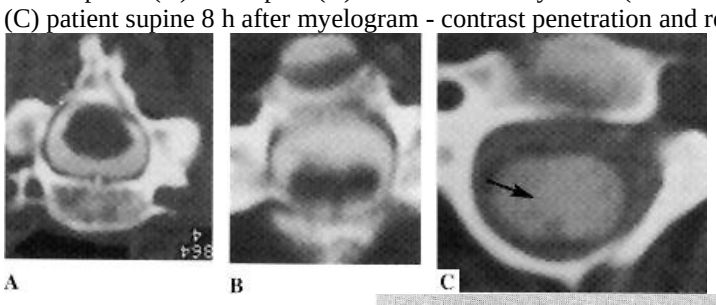
T1-MRI (syringohydromyelia + Chiari I malformation): cerebellar tonsils (upper thick black arrow) lie below neural arch of C1, and distended syrinx (*white and black arrows*) extends from C2 to T2:

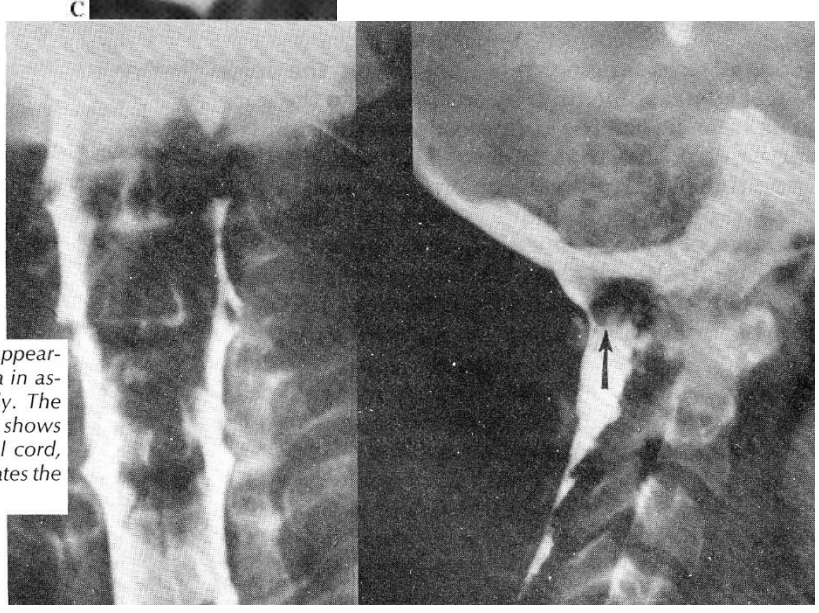


Myelography followed by delayed CT.

- 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 view (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 >>