

Syringomyelia

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

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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).

- syrx is lined by dense, gliogenous tissue and surrounded by dense glial fibril wall.
- syrx may communicate with central canal (fluid within syrx is similar to, if not identical with CSF).
- most common starting location - base of posterior horn in cervical cord.
 - syrx 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).
- syrx slowly enlarges (due to hydrodynamic forces) to involve much of both gray and white matter (at times, only narrow rim of parenchyma is left).

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** (e.g. birth 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.
- Tumors** - syrx 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. hydromelia) – 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 → syrx formation; foramen magnum decompression usually results in syrx 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 3-4th decade (mean age at onset ≈ 30 years) - chronic, slowly progressive **CENTRAL CORD syndrome**. see p. Spin1 >>

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

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.

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



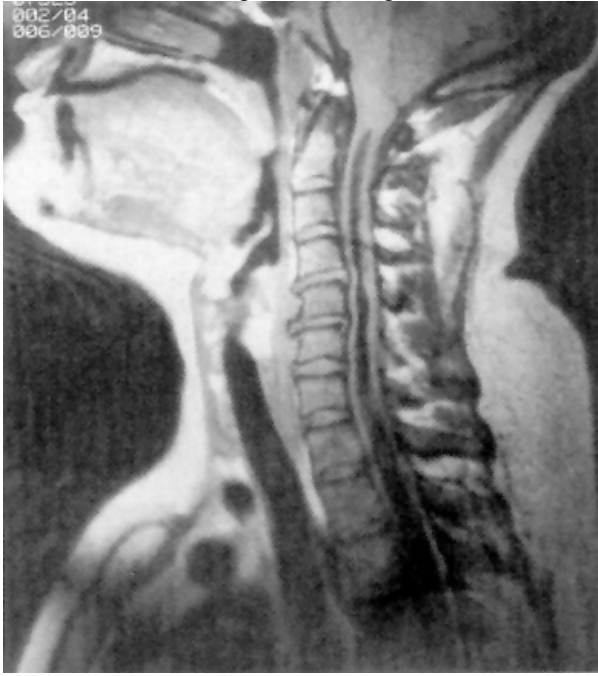
MRI (syringomyelia + Chiari malformation):

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

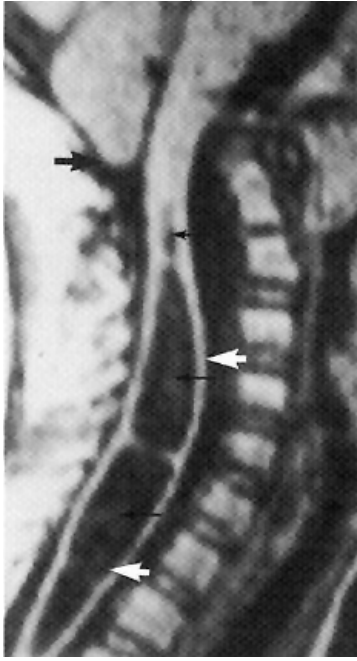
B. MRI-T2: high-signal-intensity syringomyelia (*white arrows*) expanding cervical cord with signal intensity equivalent to CSF.



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



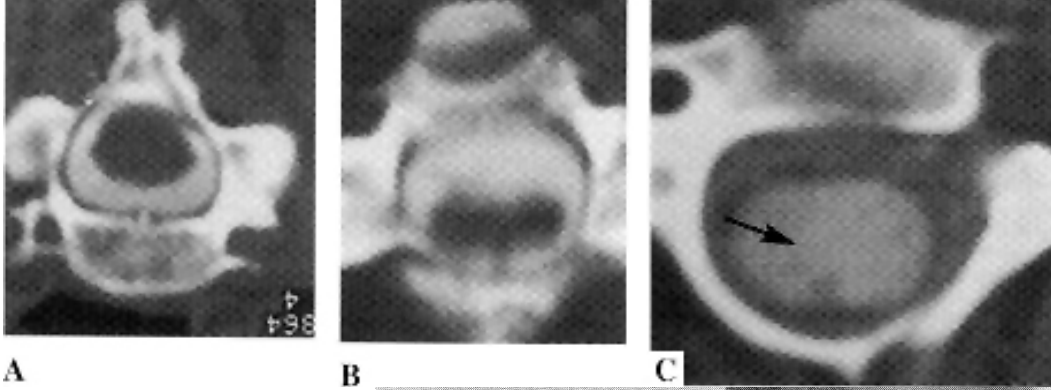
T1-MRI (syringohydromyelia + Chiari I malformation): cerebellar tonsils (upper thick black arrow) lie below neural arch of C1, and distended syrinx (white and black arrows) extends from C2 to T2:



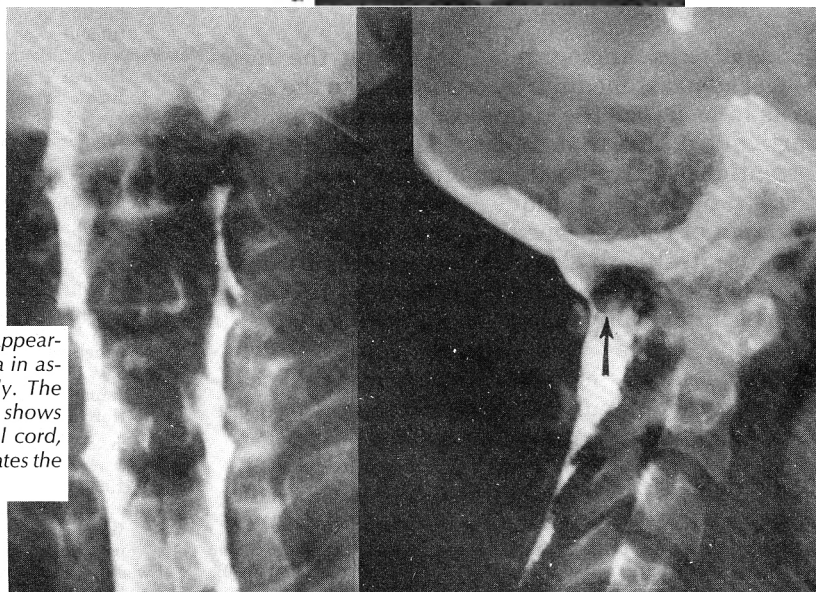
Myelography followed by delayed CT.

- 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).



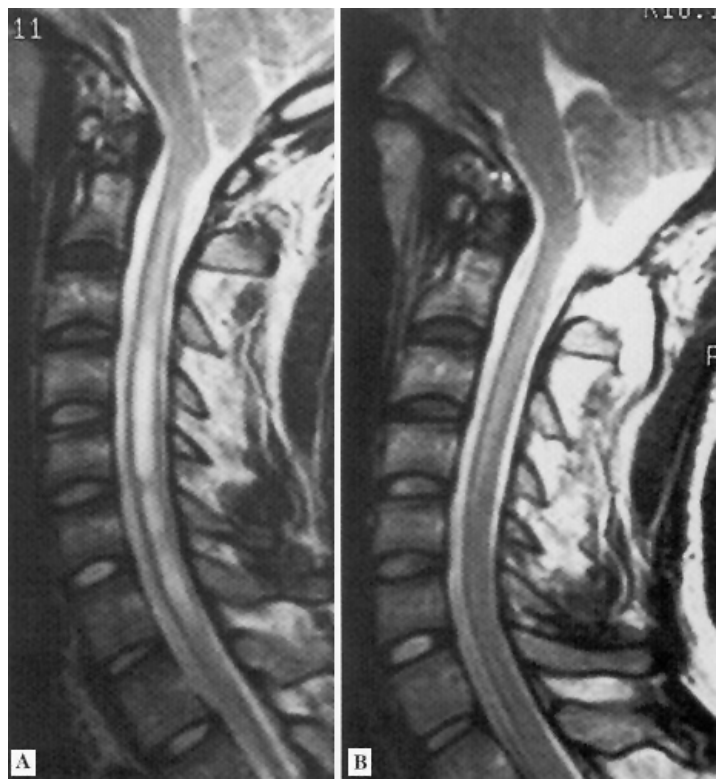
DIFFERENTIAL DIAGNOSIS

- all easily excluded with MRI:
 - 1) intramedullary tumors.
 - 2) ALS
 - 3) MS
 - 4) cervical spondylosis
 - 5) anomalies of craniovertebral junction

TREATMENT

- A. **Drainage of syrinx cavity** (e.g. in **arachnoiditis**, **post-traumatic** syrinx):
 - a) **temporary** - *percutaneous needle aspiration* or *open syringotomy*.
 - b) **permanent** - *syringopleural* or *syringoperitoneal shunt*: insert small Silastic tube (through laminectomy) directly into syrinx cavity; other catheter end → pleural or peritoneal space (continuous drainage into cavity of lower pressure).
- B. **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.

MRI-T2 (syringomyelia):
(A) mild cerebellar ectopia and syrinx (white) in cervical spinal cord.
(B) same case 1 year post foramen magnum decompression - syrinx has partially collapsed.



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