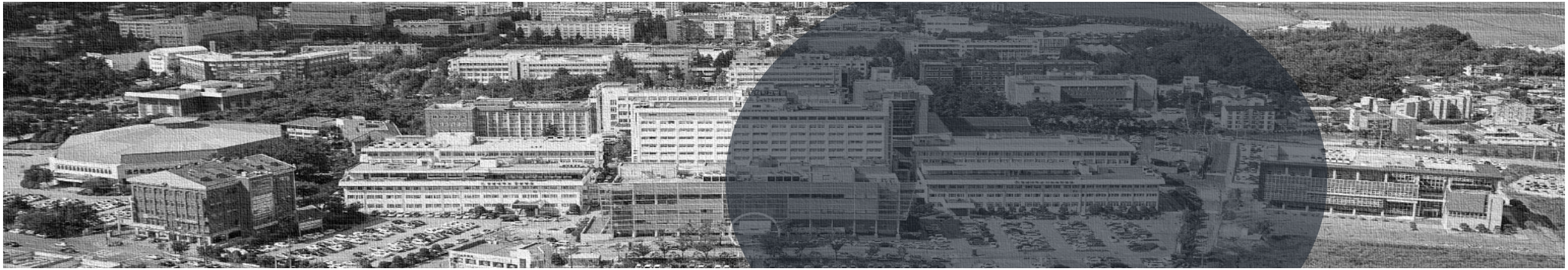
Central cord syndrome & Syringomyelia

원광대학교병원 재활의학과

주민철

목차 Content



Introduction

- Central cord syndrome, first described in 1954, is the **most common form** of incomplete spinal cord injury (SCI) and has an annual incidence of approximately 11,000 cases in the United States.

- It leads to motor deficits that are **more pronounced in the upper extremities** compared to the lower extremities, as well as **bladder dysfunction (retention) with sacral sparing**.
  - ✓ Because of its unique clinical presentation, central cord syndrome is also described in the differential diagnosis of "**man in a barrel**" syndrome.

Michael et al. J Emerg Med. 2015 Mar;48(3):333-4

Etiology

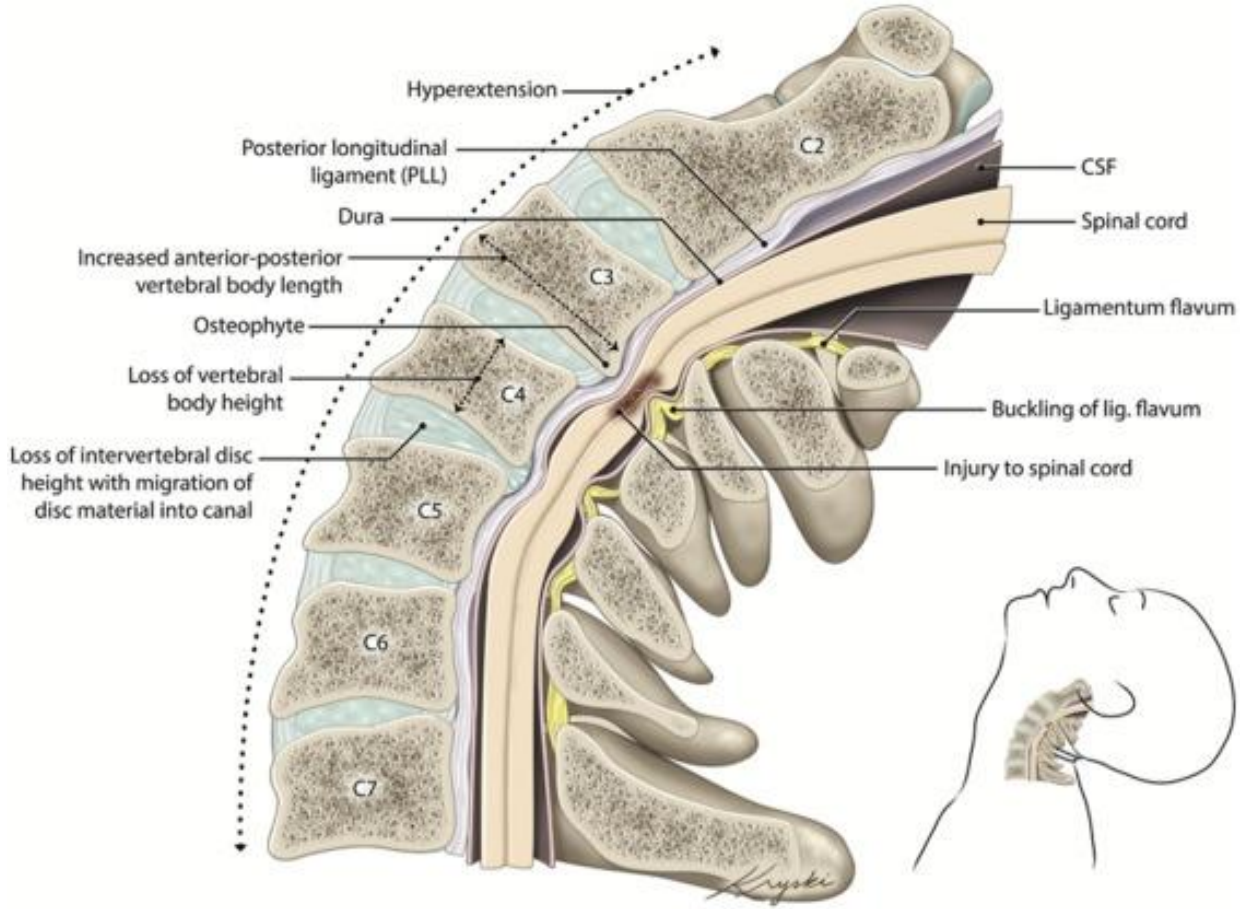
- Central cord syndrome is an incomplete cord syndrome that most commonly results following a **hyperextension injury** of the cervical spine leading to spinal cord impingement.

- Central cord syndrome usually occurs in people with **existing arthritis** changes in the bones of the **neck**.
  - In such situations, the canal through which the spinal cord travels can be narrow,
  - There is usually no obvious break or fracture in the bones of the neck and spine may be stable.

Vamsi et al. Radiographics. 2018 Jul-Aug;38(4):1201-1222.

Pathophysiology

- Central cord syndrome occurs most commonly in those who suffer a **hyperextension** injury.
  - This usually happens with a forward fall while striking the chin and having the neck extend backward at the time of the fall.
  - This leads to the posterior cord being either compressed or irritated by the posterior ligamentum flavum or anterior cord compression from underlying spondylosis.
  - So that if the neck is **forcefully extended (head tilted back)**, the spinal cord can be squeezed.



Jetan et al. J Neurosurg Spine. 2018 Dec 20;30(1):1-18.

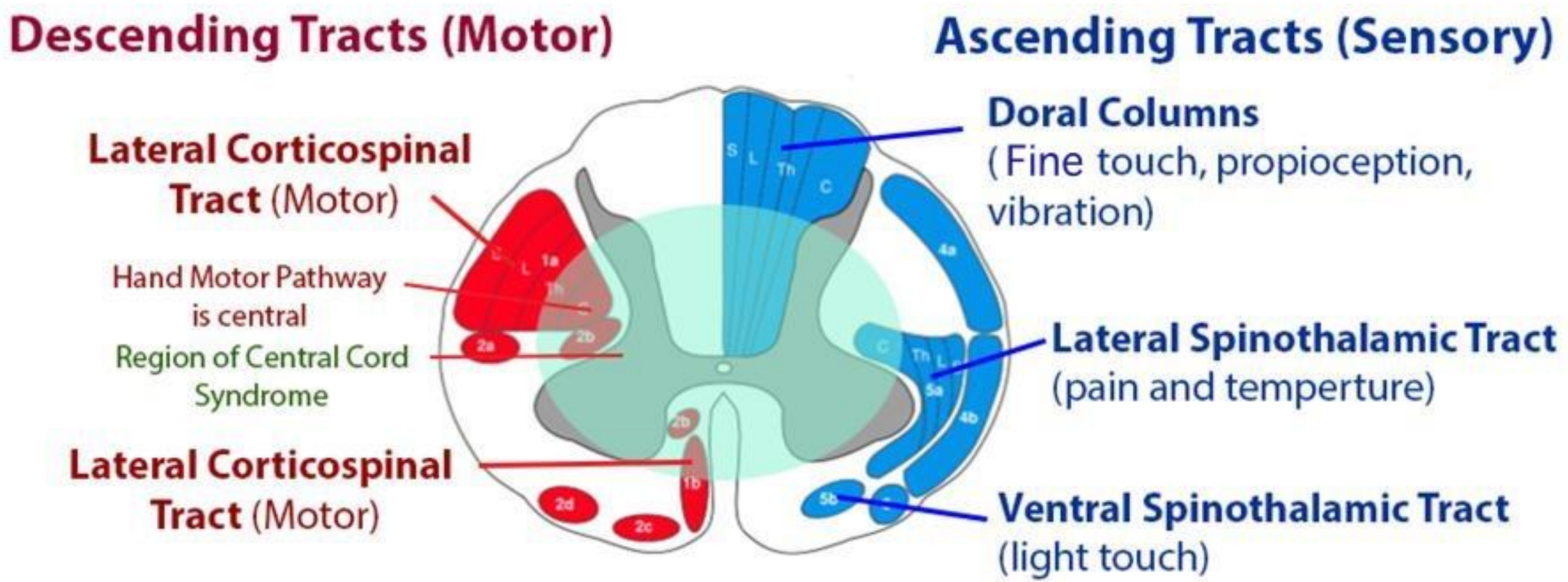
Pathophysiology

- These two contusion-type injuries to the cord can lead to clinical symptoms secondary to

1. **Edema** of the cord at the site of injury.
2. **Bleeding** into the cord at the injury site, which understandably has a **worse prognosis**
   - Usually, young patients often suffer from a higher velocity of trauma, leading to cervical spine subluxation or fracture that leads to the above-described compression, contusion, and bleeding.

Marjolaine et al. Ann Phys Rehabil Med. 2019 Nov;62(6):426-430.

Relevant anatomy



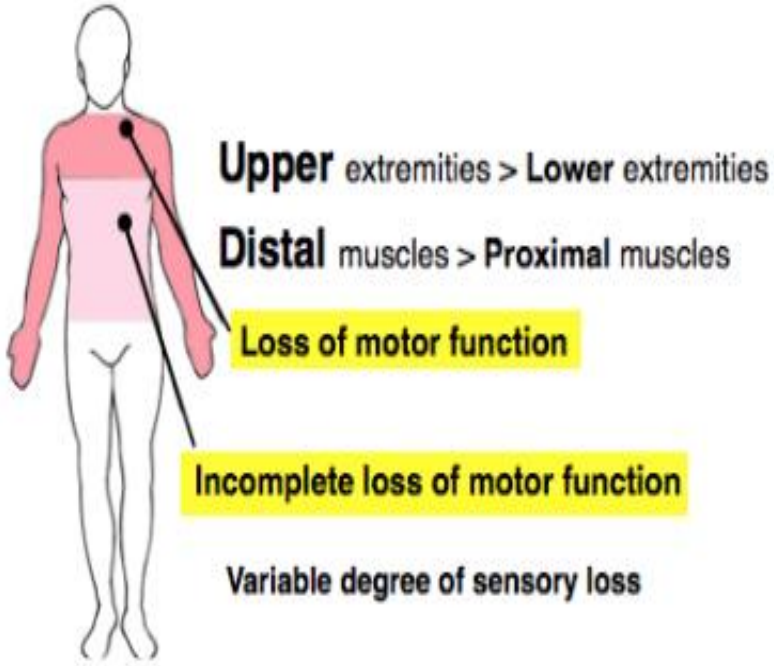
Sohaib et al. Clin Spine Surg. 2018 Dec;31(10):407-412.
Srikanth et al. Global Spine J. 2019 May;9(1 Suppl):89S-97S

Clinical manifestations

- On examination, patients will have more significant strength impairments in the upper extremities (especially the hands) compared to the lower extremities.

- Patients often complain of sensory deficits below the level of injury, but this is variable.
  - Pain and temperature sensations are typically affected, but the sensation of light touch can also be impaired.
  - The most common sensory deficits are in a "cape-like" distribution across their upper back and down their posterior upper extremities.

- The sacral sensation is usually preserved, but the clinician should assess the rectal tone to evaluate the severity of the compression.



Emilio Sacco. Urologia. 2012;79(1):24-35.

Diagnosis – Clinical

- The clinical evaluation of central cord syndrome mirrors other spinal cord injuries.

- The gold standard examination tool for any spinal cord injury is the ISNCSCI.
  - Pouw et al. proposed that **a difference of 10** between upper and lower limb motor scores on the International Standards for Neurological Classification of SCI (ISNCSCI) may be a more objective diagnostic criterion.

- Identifying the neurological level of injury is paramount as it can help determine prognosis and guide therapeutic strategies and functional rehabilitation goals.

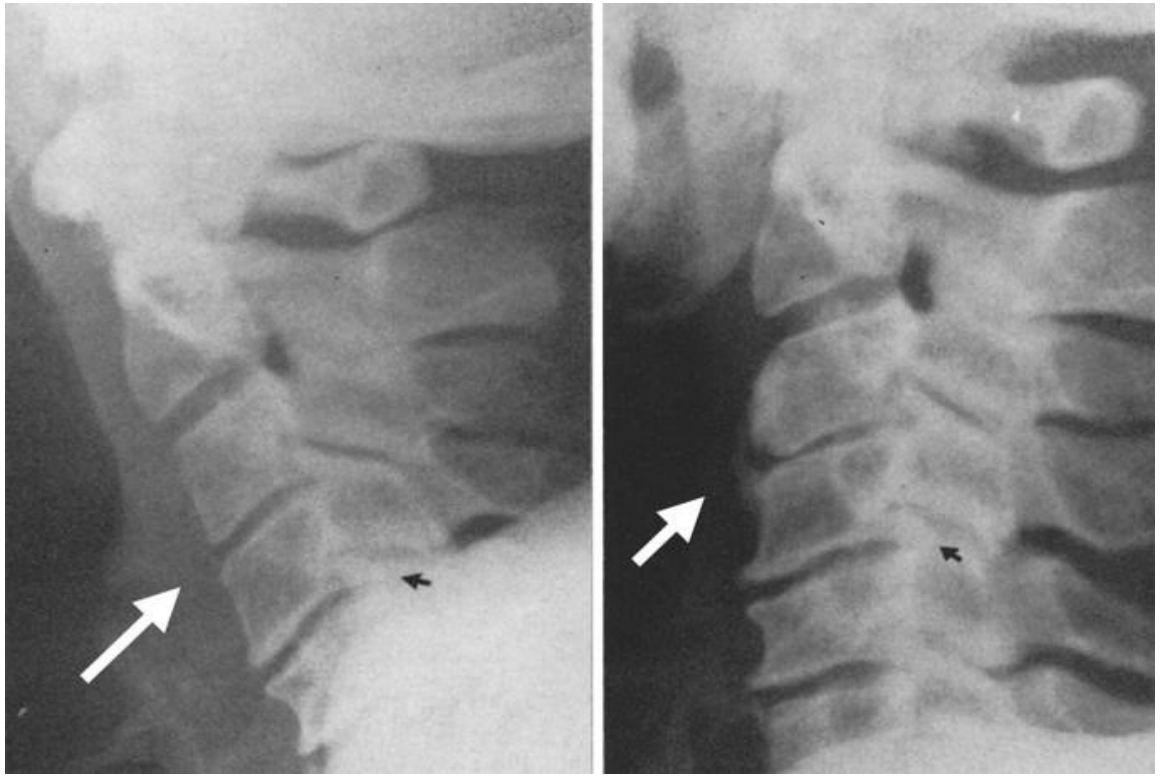
MH Pouw et al. Spinal Cord. 2010 Sep;48(9):652-6

Diagnosis – Radiologic

- Evaluation of a patient with suspected central cord syndrome includes :
  - Plain cervical spine X-rays, including supervised flexion and extension views
  - Cervical computed tomography (CT) scan
  - Cervical magnetic resonance imaging (MRI)

Diagnosis – Radiologic

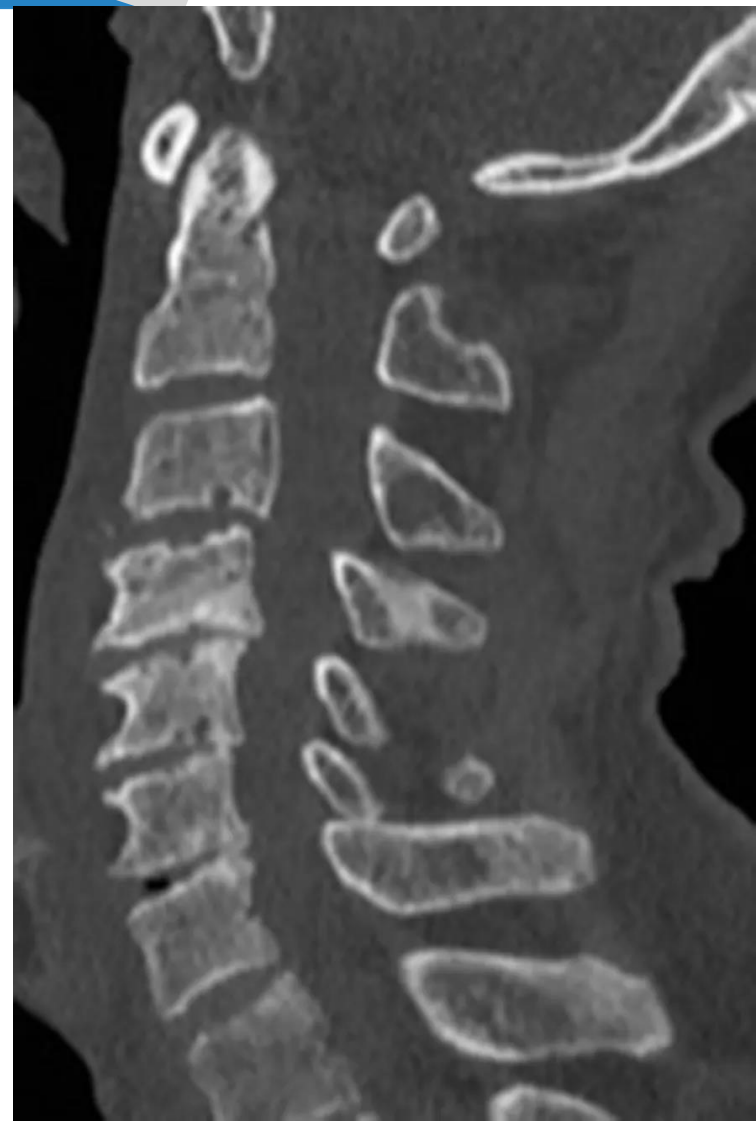
- Evaluation of a patient with suspected central cord syndrome includes :
  1. Plain cervical spine X-rays, including supervised flexion and extension views
  - X-rays of the spine delineate **fractures** and **dislocations**, as well as the degree and extent of **arthritis** type changes.
  - Flexion/extension views (moving the neck forwards and backwards) assist in the evaluation of **spinal stability**.



Krappinger, D et al. Eur Spine J 28, 434–441 (2019)

Diagnosis – Radiologic

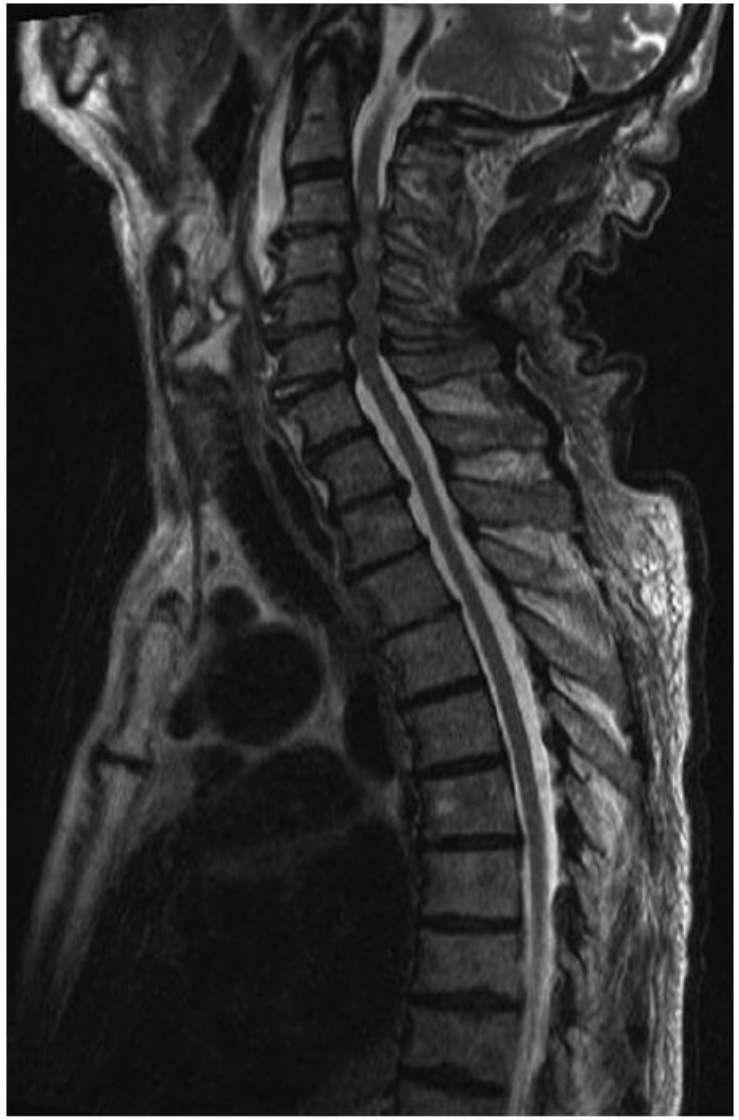
- Evaluation of a patient with suspected central cord syndrome includes :
  2. Cervical computed tomography (CT) scan
  - A computer-enhanced X-ray imaging device that shows **bony detail** superior to any other imaging device.
  - It also shows shape and **size of the spinal canal**, its contents and the **structures around it**.
  - It is usually performed prior to MRI scanning.



Krappinger, D et al. Eur Spine J 28, 434–441 (2019)

Diagnosis – Radiologic

- Evaluation of a patient with suspected central cord syndrome includes :
  3. Cervical magnetic resonance imaging (MRI)
  - A diagnostic test that produces three-dimensional images of body structures using powerful magnets and computer technology;
    - ✓ can show direct evidence of spinal cord compression from bone, disc or hematoma.
  - MRI can also show ligamentous and soft tissue injuries that might be missed by other imaging tests.



Bizhan Aarabi et al. Neurosurg Focus 25 (5):E9, 2008

Prognosis

- The natural history of the entity has an inherent potential for recovery even without surgery.
- The prognosis of central cord syndrome is variable, but most patients have a neurological recovery to some extent.
- Young trauma patients and those who seek immediate medical attention have better chances of neurological recovery.
- Prognostic factors include
  - Age / Severity of the initial neurologic deficit / Initial MRI findings.
- Roth et al. found age to be the most important prognostic indicator,
  - as patients **younger than 50** have more **favorable outcomes**.

Srikanth et al. Global Spine J. 2019 May;9(1 Suppl):89S-97S

Prognosis

- Patients with central cord syndrome recover substantial neurological function after the injury; their capacity to walk recovers in most cases.
  - However, some neurological deficits remain.

- Improvement usually occurs in an **ascending fashion**, with motor leg function recovering first, followed by bladder control, then proximal arms.
  - The hand function appears to return last.

- Abnormal MRI signals can help predict the likelihood of neurological recovery that may occur later in the course of recovery.

James G et al. Cureus. 2019 Jul 30;11(7):e5279.
Cody D. Et al. World Neurosurg. 2019 Dec;132:202-207.

Prognosis

- Aito et al. performed a retrospective analysis on 82 central cord syndrome patients and found that

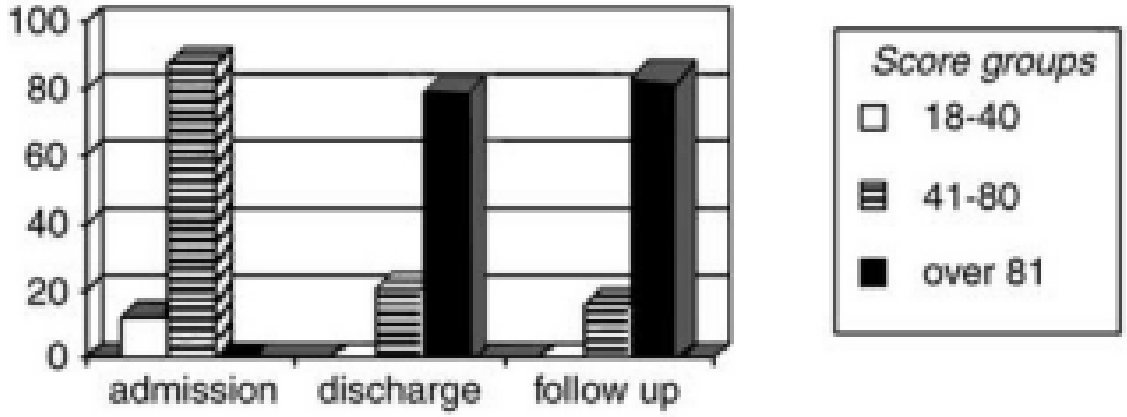


Figure 3 Distribution of FIM scores on admission, discharge and follow-up

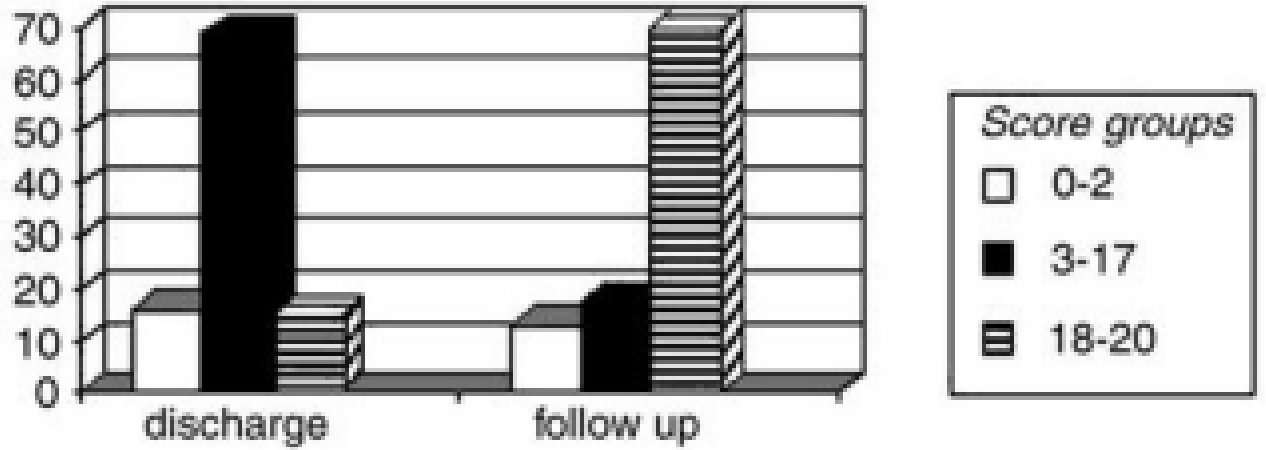


Figure 4 Distribution of WISCI scores at discharge and follow-up

C	37 (45.0%)	11 (13.4%)	11 (13.5%)
D	31 (38.0%)	70 (85.5%)	62 (75.5%)
E	0	1 (1.0%)	9 (11.0%)

Aito et al. Spinal Cord. 2007 Apr;45(4):292-7.

Prognosis

- Aito et al. performed a retrospective analysis on 82 central cord syndrome patients and found that
  - 47% of patients had persistent neuropathic pain
    - ✓ Patients at age 50 or more at the time of the lesion had more frequent pain than patients under 50 years of age.
  - 66% reported spasticity at follow-up (at least 1.5 years)
    - ✓ Of these patients, 72% claimed their spasticity to be a serious problem in their activities of daily living.
  - Achieving spontaneous voiding
    - ✓ 55% on discharge to 68% at follow up

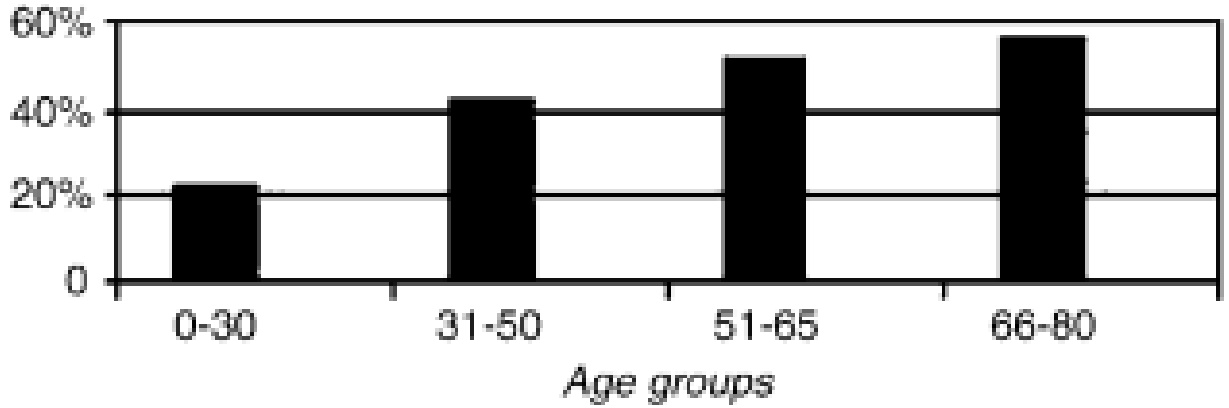


Figure 5 Percentage of patients suffering from neuropathic pain at follow-up, in different age groups

Aito et al. Spinal Cord. 2007 Apr;45(4):292-7.

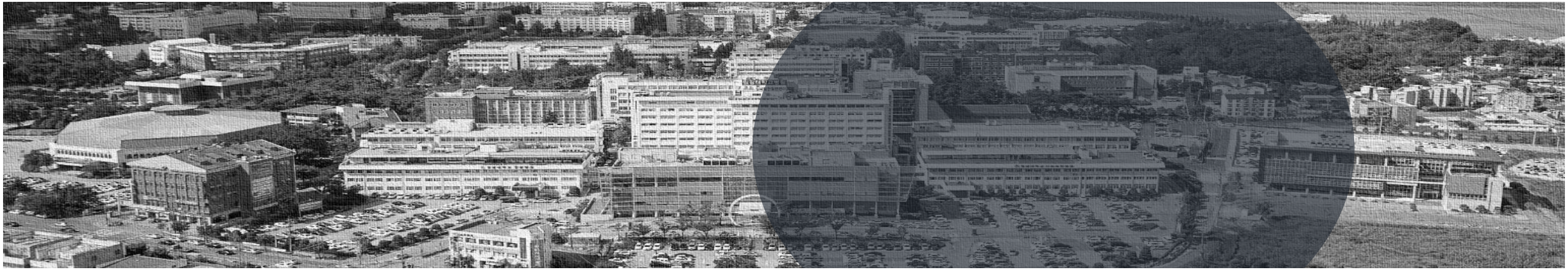
Prognosis

- Recovery generally plateaus at two years post-injury.
  - Approximately 86% of patients will recover the ability to ambulate
  - 80% of patients will have recovered functional independence

- At three years post-injury, patients should recover at least 90% of their motor score regardless of surgical versus conservative management;
  - however, patients who underwent surgical intervention had greater functional scores and were more likely to attain their pre-injury mobility status.

Nathaniel P. Brooks. Neurosurg Clin N Am. 2017 Jan;28(1):41-47.
Amina et al. J Spinal Cord Med. 2018 Jul;41(4):490-495.

목차 Content



Introduction

- Syringomyelia literally means "**cavi**
  is typically a progressive chronic co
- In the absence of trauma this can re
  (pyogenic or tuberculous), or hemo
- Intramedullary tumors can produce
  extension of their cystic componen
  hemangioblastoma) or by cord exp
  movement in the spinal theca.

HINDBRAIN RELATED
Cerebellar tonsillar ectopias:
- Chiari 0: Low position but not herniated, with typical Chiari symptoms
- Chiari 1: Cerebellar tonsillar descent only
- Chiari 1.5: Tonsillar herniation +some medullary descent*
- Chiari 2: Cerebellar+medullary descent, low inion, small posterior fossa†

Arachnoid fibrosis for example, birth injury
Other for example, Paget's disease, rickets

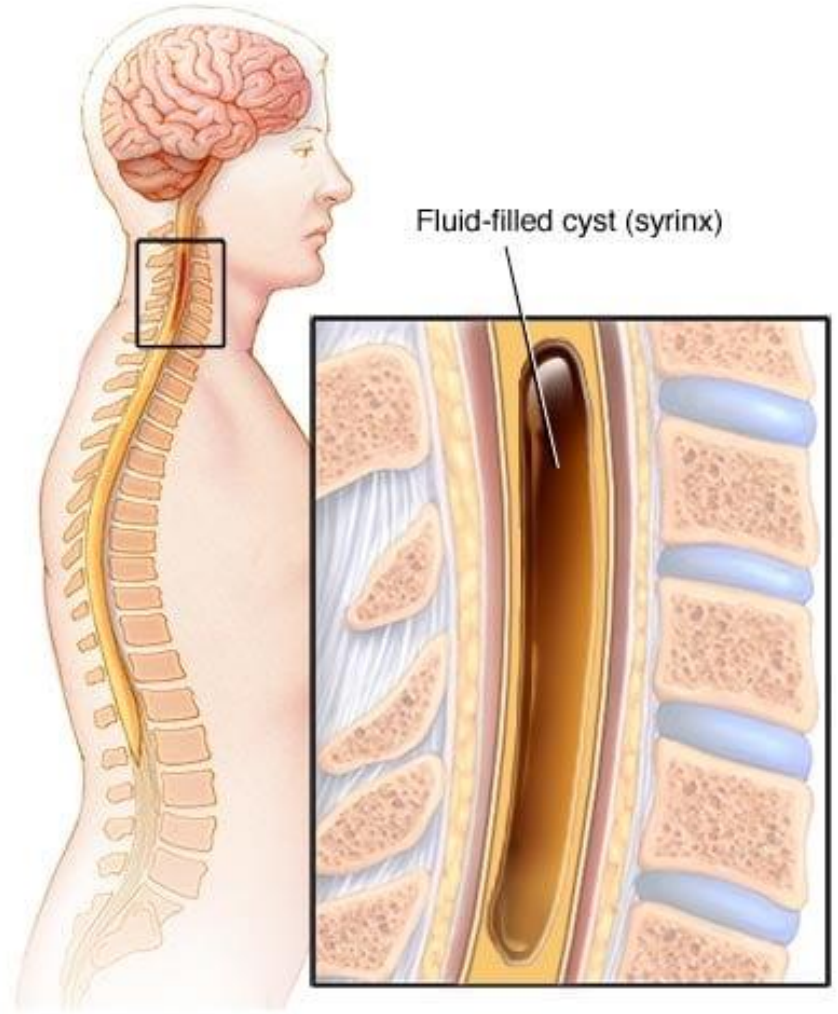
SPINAL PATHOLOGIES
Post-haemorrhagic adhesions:
- Spinal injury
- Subarachnoid haemorrhage

Post-infective adhesions:
- Pyogenic,
- Tuberculous

Arachnoid cysts and webs
Tumour related for example, haemangioblastoma

IDIOPATHIC
Embryological remnants: normal calibre central canal
Dilated central canals: as assessed on axial images
Fully formed cavities: open cisterna magna: no Valsalva/ hindbrain symptoms
Glio-ependymal cysts: if located in conus then possibly related to cord tethering

*May be associated with a low-lying, tethered conus
†Accompanying spinal dysraphism and, frequently, hydrocephalus



Flint G. Pract Neurol 2021;21:403–411.

Introduction

- The most common cause of progressive myelopathy after an SCI is posttraumatic syringomyelia (PTS).

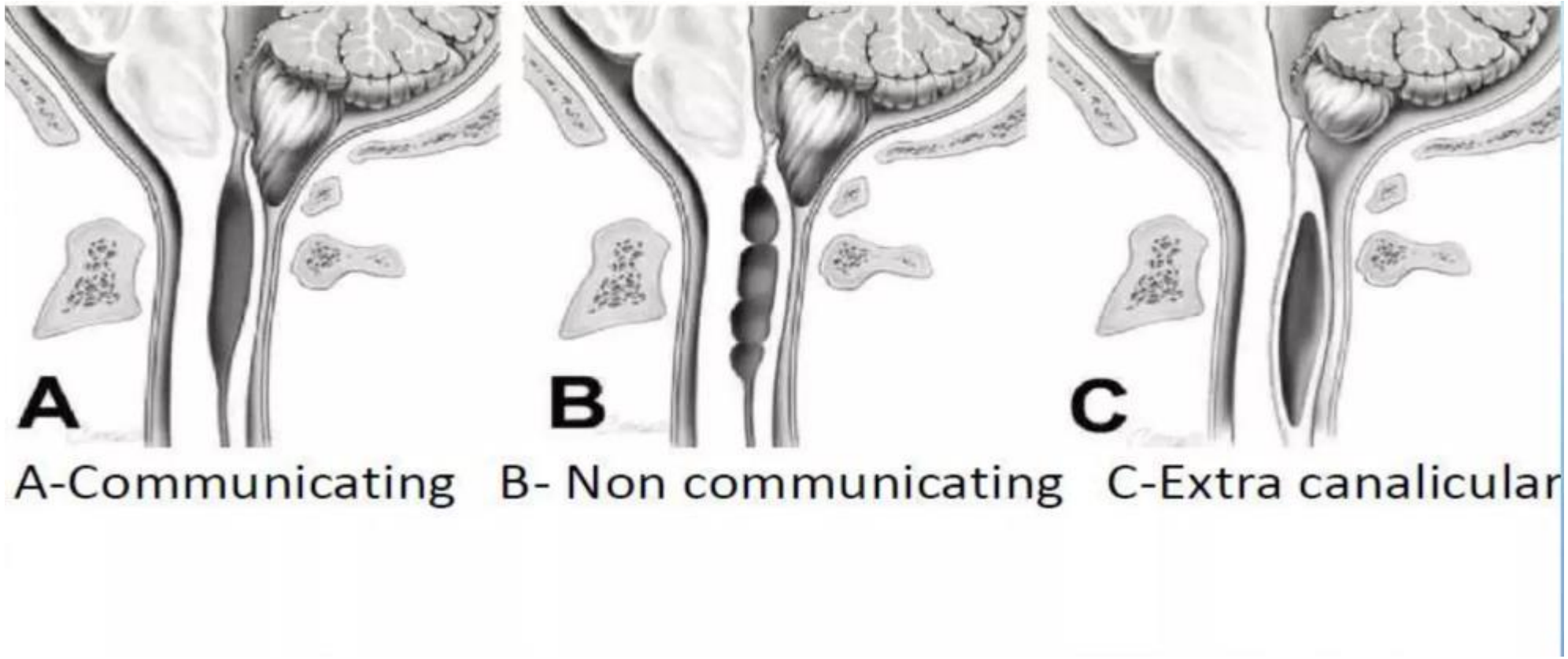
- Although it is common to find MRI evidence of a cyst within the spinal cord at the level of the injury,
  - ✓ only 5% of all people with SCI develop PTS.

- PTS may develop at any time, from 2 months to decades postinjury.
  - ✓ PTS usually presents at 5 to 15 years postinjury and is more common and may occur earlier in persons with neurologically complete SCI (AIS A), in cervical followed by thoracic compared with lumbar SCI, and in those injured after age greater than 30.

- It becomes a problem when the cyst expands longitudinally and damages the cord,
  - ➢ causing clinical symptoms such as pain, sensory loss, weakness, altered muscle tone, and a variety of autonomic symptoms.

Pathogenesis

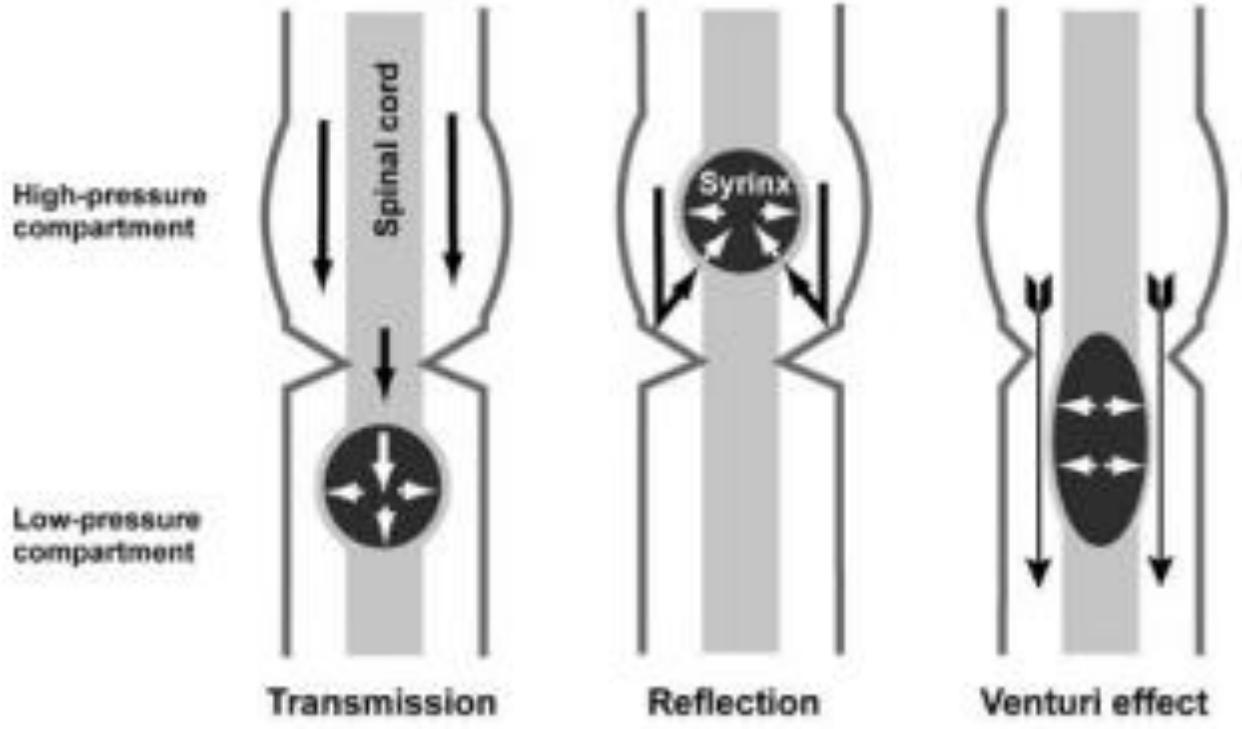
- Depending on the connection with fourth ventricle



Milhorat et al.

Pathogenesis

- The exact cause of PTS is unknown, but it might be **related to obstruction of the normal flow of CSF**
  - ✓ because of scarring or canal narrowing.
  - ➢ This leads to abnormal increases in CSF pressure during coughing and straining, ultimately causing a longitudinal dissection of the cord.



Clare et al. J Vet Intern Med 2006;20:469–479

Pathogenesis

- The cavity begins at the level of the cord injury in the gray matter between the dorsal horns and posterior columns.

- The cyst may extend rostrally and/or caudally, compressing the cord, by dissecting through the intermediate gray matter, and may **result from increases in subarachnoid fluid pressure due to intraabdominal or intrathoracic pressure increases**
  - ✓ i.e., cough, sneeze, straining or Valsalva, weight lifting, forward-lean pressure release, and quad coughing

Clinical manifestations

- The clinical symptoms of PTS and their progression vary greatly.

- It remains **impossible to predict** who might be at greatest risk.
  - It is therefore sensible to offer spinal cord injury patients life-long follow-up, with interval imaging should there be any change in their symptoms or signs

- Early signs and symptoms of PTS are often nonspecific and variable;
  - ✓some patients with a markedly elongated syrinx may even have minimal symptoms.

- The most common presenting symptom is **aching, burning pain**
  - that **increases with cough, sneeze, and straining**,
  - usually located at the site of the original injury or may radiate to the neck or upper limbs.

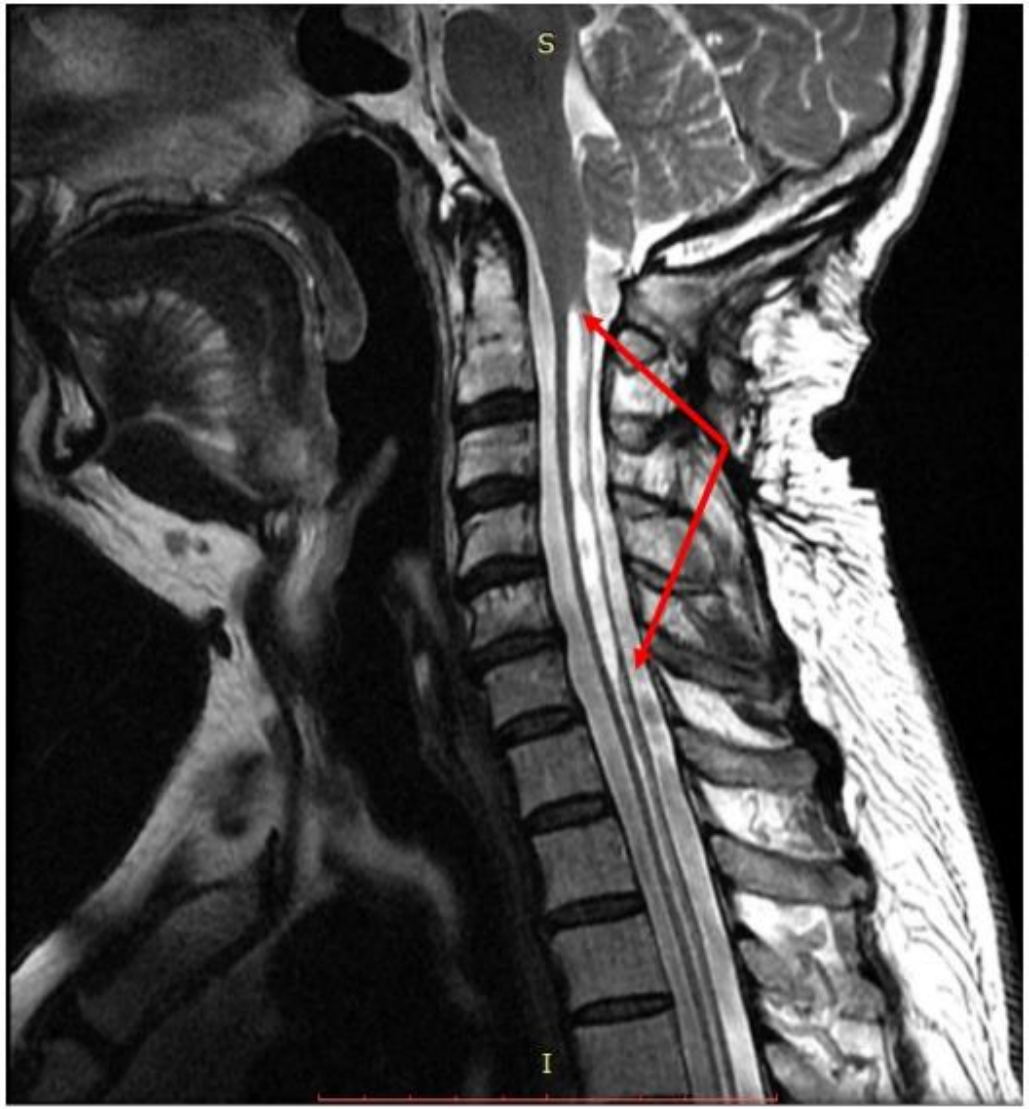
Flint G. Pract Neurol 2021;21:403–411.

Clinical manifestations

- The earliest sign is an ascending loss of deep tendon reflexes.
  - An ascending sensory level is common, typically with a dissociated loss of pain and temperature sensation, but intact touch, position, and vibration sense.
- Loss of pain sensation can lead to a Charcot joint.
- Weakness occurs, but rarely in isolation.
- Additional findings may include spasticity changes, hyperhidrosis, AD, fatigue, bladder changes, worsening OH, scoliosis, respiratory changes, and a myriad of other symptoms.

Diagnosis

- The diagnosis of PTS is confirmed by MRI or CT myelography with delayed images
  - ✓ MRI with gadolinium is the gold standard for diagnosing PTS.
- A syrinx may spontaneously resolve, progress, and then plateau or progress continuously.
- Neurologic monitoring is essential, and serial MRI imaging may be helpful.



Lodhi et al. Cureus 9(11): e1852

Treatment

- If no neurologic decline is noted on regular follow-up examinations, the treatment can be symptomatic.

- Conservative treatment includes
  - ✓ Pain control, activity restrictions, maintaining head of bed elevation greater than 20 degrees, and providing rehabilitation interventions as needed (i.e., functional training and adaptive equipment).

- **Activity restrictions** include avoiding maneuvers that increase intrathoracic/abdominal pressure,
  - ✓ High-force exercise,
  - ✓ Valsalva, Credé, and
  - ✓ Quad coughing with direct compression over the IVC, and
  - ✓ Avoiding anterior weight reliefs and weight lifting, especially if these activities could raise venous and CSF pressure and exacerbate symptoms.

Treatment

- Surgical treatment is usually indicated
  - ✓ if there is an ongoing neurologic (motor) decline or severe intractable pain.

- Surgical treatments include
  - ✓ Shunting (syringo-subarachnoid, syringo-pleural, or syringoperitoneal),
  - ✓ Reconstructing the subarachnoid space with dissection of arachnoiditis/meningeal scarring and duraplasty, and
  - ✓ Cordectomy.

- Surgery yields improved strength and improved pain control in some but not all, whereas sensory recovery is not usually as favorable.

Prognosis

- When treatment is successful, strength may increase and pain and spasticity may diminish.

- Reduction of syrinx size on postoperative MRI usually predicts a good surgical result;
  - ✓ however, complete resolution of the syrinx is not necessary for a good clinical outcome.

Prognosis

Table 3 Binary logistic regression results concerning the effects of age, injury level, injury severity and syrinx location on early syrinx formation, syrinx length and cranial syrinx

Predictors	*Ea*
	P-value
Age, years[a]	**0.001**
12–30	R
31–45	**0.004**
46–63	**0.003**
Injury level	0.7
Injury severity	**0.035**
Syrinx location	0.9

Table 4 Occurrence of ea

Table 1 Patien

Categories			
Age, years			
12–30	1/3.4%	24/82.8%	4/13.8%
31–45			
46–63			
Injury severity			
Complete			
Incomplete			

Table 5 Time from spinal cord injury to syrinx formation for different age, injury severity, injury level and syrinx location groups

	Time to syrinx formation (years)			*P-value*
	Median	*95% CI*	n	
Age, years				
12–30	16.7	15.2–19.8	88	Reference
31–45	5.1	2.2–14.2	28	**<0.001**
46–63	3.5	0.9–17.2	13	**0.002**
Injury severity				
Complete	13.5	9.5–16.0	103	**0.027**
Incomplete	19.2	13.6–21.5	25	
Injury level				
Tetraplegic	15.8	9.5–19.6	52	0.5
Paraplegic	14.2	10.0–16.7	77	
Syrinx location				
Cranial	13.6	5.5–19.2	35	0.6
Not cranial	15.0	10.6–17.7	89	

Cranial syrinx progression		
lue	*Exp(B)*	*95% CI*
5		
	Reference category	
5	1.4	0.5–3.7
	1.9	0.5–7.4
	Not evaluated	
l	0.4	0.1–1.3
	Not evaluated	

	Total
;)	*(n/%)*
%	91/100%
5%	32/100%
3%	15/100%
%	109/100%
	29/100%

Total
81/58.7%
57/41.3%
138/100%

J Krebs et al. Spinal Cord (2016) 54, 463–466

Prognosis: Surgical approach vs Conservative Tx

Acta Neurochirurgica (2020) 162:2541–2556
https://doi.org/10.1007/s00701-020-04529-w

REVIEW ARTICLE - SPINE - OTHER

Treatment of posttraumatic syringomyelia: evidence from a systematic review

Andrea Kleindienst[1,2] · Francisco Marin Laut[3] · Verena Roeckelein[2] · Michael Buchfelder[2] · Frank Dodoo-Schittko[4]

Andrea et al. Acta Neurochirurgica (2020) 162:2541–2556

Prognosis: Surgical approach vs Conservative Tx

- The outcome of conservative and surgical treatment is **not directly comparable**
  - because of the exclusively observational study design with the subsequent selection bias and cross-over.

- While a satisfying outcome defined as either an **improved or stable situation is identical**
  : Conservative 85% vs Surgery 88%
- The deterioration rate; 15.5% in Conservative vs 9.1% with surgery
  - But, surgery accompanied by a 0.33% surgery-related mortality and 23% complications.

Andrea et al. Acta Neurochirurgica (2020) 162:2541–2556

감사합니다

濟生醫世 ; 의술로써 병든 세상을 구제한다

건강 사회를 선도하는 맑고 밝고 훈훈한 원광대학교병원

원광대학교병원
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