

# 38 year old with chronic back pain and lower extremity spasm

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Spinal Dysraphism (manifest as spina bifida, tethered cord, and diastematomyelia)

- Broad term for a group of anomalies of dorsal malformations of the embryo
- Result of abnormal notochord development
  - Bunches of genes: Hox, GLUT1, HK1, LEP...
  - But strangely there's not much evidence of a congenital link to explain majority of cases



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### Spinal Dysraphism

### • Open

- Meningoceles
- Myelomeningocele
- Closed
  - Thickened filum terminale
  - Dorsal dermal sinuses
  - Neurenteric cyst
  - Diastematomyelia

### Diastematomyelia

- Longitudinal split in the spinal cord
- AKA split cord malformation
- Two types:
  - Type I: Duplicated dural sac
  - Type II: Single dural sac containing both hemicords

### Diagnosis

- X-ray
  - multilevel spina bifida
  - widening of the interpedicular distance
  - AP narrowing of vertebral bodies
- CT
  - All the above, plus delineation of the bony septum
- MRI
  - Modality of choice
  - Demonstrates the split cord, hydromelia, plus any of the associated anomalies
- Ultrasound
  - Antenatal ultrasound can detect

## Type I

- Clinically more severe, earlier presentation
- Duplicated dural sac
- Midline spur, either osseous or osteocartilaginous
- Vertebral anomalies
- Skin lesions
- Scoliosis and tethered cord syndrome



- 14 yo girl
- Numbness and tingling in legs
- Back pain
- Weakness
- Constipation

Aly, N, Towbin, A, Towbin, R.





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## Type II

- Milder form, even asymptomatic
- Single dural sac
- Sometimes hydromelia
- Spina bifida maybe present, but other vertebral anomalies much less common

### Signs and Symptoms

- Asymptomatic at birth
- Gradually...
  - Bowel and bladder dysfunction
  - Motor and sensory dysfunction
  - Progressive pain
- Can be associated with visceral malformations, skin abnormalities

### Treatment

- Conservative, if patient is asymptomatic
- If symptomatic:
  - Surgical decompression
  - Removal of bony spur and reconstruct dura around the separated cords
  - Release of tethered cord

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