38 year old with chronic back pain and lower extremity spasm

Donald Imwalle
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
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.
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
References


