### Objectives
- Introduce key embryologic factors in congenital spinal lesion development
- Utilize a clinical-radiologic framework to categorize lesions
- Case based review of representative congenital spinal lesions
- Introduce key concepts of specific entities

### Primary Neurulation
- **Primary Neurulation**
  - Neural plate folds and fuses to form the neural tube
  - **Disjunction** - the separation of neuroectoderm from ectoderm to form the neural tube.

### Disjunction

#### Nondisjunction
- OSD
- Meningocele
- Myelomeningocele
- Dermal Sinus

#### Premature Disjunction
- Lipoma
- Lipomyelocele
- Lipomyelomeningocele
Secondary Neurulation

- Secondary Neurulation
- Development of neural structures caudal to the posterior neuropore
- Connects to the neural tube formed by primary neurulation

Clinical Radiologic Framework

- Caudal regression
- Thick filum, “TCS”
- Filar lipoma/fibrolipoma
- Dilated terminal ventricle
- Terminal myelocystocele
- Sacrococcygeal tumors

Case 1

- Abnormal 18 week screening ultrasound
- Referral with follow up ultrasound
- Subsequent fetal MRI for further characterization

Obstetric Ultrasound
Fetal MRI

Postnatal MRI

Myelomeningocele

Myelocele (myeloschisis)

Open Spinal Dysraphism

• Myelocoeles and Myelomeningocele
• Sequela of nondisjunction
• Link with maternal folate deficiency
• Rarely imaged postnatal
• Pre vs. Postnatal repair

Companion Case 1b

OSD Repair
OSD Repair
• Commonly have this “tethered” appearance at the OSD closure.
• Post operative neurologic deficit should be stable
• Change would be an indication for MRI
• Symptoms at 5-9 years, period of rapid growth

OSD Repair: What am I Looking For
• Tethered by scar?
  • Diagnosis of exclusion:
• Dural ring at the closure
• Mass: epi-/dermoid, arachnoid cyst
• Syringohydromyelia
• Vascular compromise
• Associated malformations
  • SCM, Hydro, Sinus tract

Framework Part 1
1. Open Spinal Dysraphism
  • Myelocoele
    – Hemimyelocoele in the setting of diastematomyelia
  • Myelomeningocele
    – Hemimyelomeningocoele in the setting of diastematomyelia

Case 2
• Infant with low back soft tissue mass
• Hairy tuft

DDX: Framework Part 2
1. Open Spinal Dysraphism
  • Myelocoele
  • Myelomeningocele
2. Closed Spinal Dysraphism (With Mass)
  • Meningocele
  • Lipomyelocoele
  • Lipomyelomeningocele
  • Myelocystocele
**Lipomyelomeningocele**

- Neural placode inserts into fatty mass
- Mass contiguous with subcutaneous
- Expansion of arachnoid space
- Displacement of placode beyond the spinal canal

**Lipomyelomeningocele**

- Sequela of premature disjunction
- Inner neural tube induces mesenchyme to fat
- Spinal cord always tethered to fatty mass
- Fatty mass may be large or nearly imperceptible

**Lipomyelocele**

- Sequela of premature disjunction
- Neural placode fat interface within the spinal canal
- Fat herniates through dorsal defect
- Cord is always tethered

**Companion Case 2a**

**Companion Case 2b**
Companion Case 2b

Dorsal Meningocele

- Presents as palpable mass or incidental
- Usually neurologically intact
- Cord may be tethered at the defect neck
- By definition does not contain neural elements
- Debated embryogenesis

Dorsal Meningocele

- Communicates with SA space and may change with valsalva
- Neck may be very small
- Surgery may be warranted to close dural defect
- Exclude neural elements by US/MRI

Framework Part 2

1. Open Spinal Dysraphism
2. Closed Spinal Dysraphism (With Mass)
   - Meningocele
   - Lipomyelocele
   - Lipomyelomeningocele
   - Myelocystocele (not included)

Case 3

- Lower extremity weakness in a child
- No cutaneous stigmata or mass
**DDX**

- Lipoma
- Filar fibrolipoma
- Dermoid/epidermoid

**Intradural Lipoma**

- Intradural, intimately involved with the cord
- Follows fat on all modalities and sequences
- Focal premature disjunction
- Normal induction of dura and posterior elements

**Companion Case 3a**

Extensive intradural lipoma

**Companion Case 3b**

Fibrolipoma of the Filum

**Filum Fibrolipoma**

- Often an incidental finding
- Hypodense, hyperintense
- If symptomatic or >2mm thick consider lipoma
- 4-6% of population
- Anomaly of secondary neurulation
- May be difficult to see on sagittal imaging, axial T1?

**Companion Case 3c**

- Clubfoot
- Findings on ultrasound prompted subsequent MRI
Companion Case 4c

Companion Case 4c

Tight Filum/"Tethered Cord"

Tight/Thick Filum

- Many lesions result in tethering of the cord
- Related to shortening and thickening of the filum
- Likely a consequence of dysfunctional caudal cell mass regression
- Presentation at birth may be as lower extremity deformities or scoliosis
- Presentation during somatic growth: weakness, urinary symptoms, abnormal gait, pain

Tight Filum

- Neurosurgical untethering
- Neurological defect should remain stable
- Recurrence prompts concern for retethering

Case 5

- Incidental spinal cord lesion
**DDX**
- Syrinx
- Cystic dilation of the terminal ventricle
- Cystic cord neoplasm
  - Astrocytoma
  - Ependymoma
  - Hemangioblastoma
- Cord insult resulting in myelomalacia

**Cystic Dilation of Terminal Ventricle**
- Focal expansion of terminal ventricle
- Usually 2-4mm
- Focal expansion localized to conus
- Difficult to distinguish from syrinx
- Exclude neoplasm with MRI

**Cystic Dilation of Terminal Ventricle**
- Typically incidental
- No treatment
- Option to follow with MRI to document stability
- Some authors argue to consider a normal variant
- Ependymal lined
- Symptomatic may fenestrate

**Case 6**
- Sinus tract on physical exam
- Asymptomatic

**Sinus tract w/ epidermoid**

Sag STIR  
Sag T1  
Sag T2  
Ax T2  
Sag DWI
**Dorsal Dermal Sinus**
- Focal nondisjunction
- Usually terminates at the conus when lumbar
- +/- Tethering
- +/- Mass
  - Dermoid/epidermoid
  - Lipoma
  - Arachnoiditis
- +/- Osseous defect

**Dorsal Dermal Sinus**
- Delineation of tract important for Sx planning
- As cord ascends pulls tract with it
- Usually presents as dimple
- If tethered, may present with neurologic symptoms

**Companion Case 7a**
Sinus tract w/ tethering

**Companion Case 7b**
Tract w/ Appendage

**Companion Case 7c**
Sacral/Coccygeal Dimple

**Sacral/Coccygeal Dimple**
- Low sacral dimple with fibrous tract to bone
- Usually within the gluteal cleft
- Usually worked up over undue concern
- Becomes less conspicuous with growth
- Limited imaging role
Framework Part 3

1. Open Spinal Dysraphism
2. Closed Spinal Dysraphism (With Mass)
3. Closed Spinal Dysraphism (Simple Dysraphic States)
   - Intradural Lipoma
   - Fibrolipoma
   - Tight filum
   - Persistent terminal ventricle
   - Dermal sinus

Case 8

- Child with scoliosis
- Now with "tethered cord" symptoms
- Urinary irregularities

Diastematomyelia Type 1

Diastematomyelia

- Type 1: two cords, arachnoid space, and thecal sac, osseous/fibrous bar
- Type 2: two cords share same arachnoid space and thecal sac, no osseous bar
- Resection of bar, untethering, correct scoliosis

Diastematomyelia Postoperative

- Split cord malformation
- Defect of notocord development
- High incidence of cutaneous stigmata
- High incidence of segmentation abnormality
Companion Case 8a

Diastematomyelia Type 2

Case 9

- Anal atresia
- Abnormality on screening obstetric ultrasound

Caudal Regression Type 1

Caudal Regression

- Spectrum ranging from complete absence to coccygeal truncation
- Spectrum of neurologically normal to severely impaired
- Associated with anal/rectal and genital urinary abnormalities
- Infants of diabetic mothers
- Complex embryology

Caudal Regression

- Type 1: Distal cord hypoplasia/truncation, more pronounced sacral abnormality
- Type 2: Tethered cord, less severe sacral abnormality, may be tethered by mass
- Complex neurosurgical and orthopedic management
Companion Case 10a

Caudal Regression Type 2

1. Open Spinal Dysraphism
2. Closed Spinal Dysraphism (With Mass)
3. Closed Spinal Dysraphism (No Mass)
4. Closed Spinal Dysraphism (Complex Dysraphic States)
   - Diastematomyelia
   - Caudal agenesis
   - Segmental spinal dysgenesis (not included)
   - Neurenteric cyst (not included)
   - Dorsal enteric fistula (not included)

References