Persistent Terminal Ventricle

- Ventriculus Terminalis
- Incomplete regression of TV of $2^\circ$ neurulation, continuity with central canal $\rightarrow$ small cavity
- PTV vs terminal myelocystocele (severe manifestation from inability of CSF to escape)
Persistent Terminal Ventricle

- Common, transient finding until 5 yrs
- Identical to CSF, in conus or conus-filum transition
- Anatomic variant, usually incidental but > 4-5mm can be associated with pain and neurologic symptoms
Terminal Ventricle
Fibrolipomatous Infiltration of Filum

- Anomaly of 2° neurulation, from totipotential caudal cell mass
- Axial T1 most sensitive
  - Small amount of fat anatomical variant vs any fat or thickening (>2mm) = tether
Fatty Filum and Ventriculis Terminalis
Incidental Fatty Filum
Terminal Myelocystocele

- Large, skin covered lumbosacral mass
- Dilatation of terminal ventricle
- High assoc with caudal cell mass anomalies (GU, lower GI, abd wall)
- Cerebellar herniation late due to lesser CSF sump effect?
- Incontinent, extremely poor LE function
- Neural elements attenuated
Terminal Myelocystocele
Myelocele (Myeloschisis)

- Placode of OSD in plane with back
- Less common, embryologically similar
- Clinical signs & function similar
Lateral Thoracic and Lumbar Myeloceles
Meningocele

- Meningeal-lined CSF sac protruding through defect
- Cord does not enter sac, may be assoc with hypertrophy of filum or cord tethering
- ? From CSF pulsations, Weakened or abnormal dura?
- Lateral assoc with NF I, also post-trauma, connective tissue disorders
Dorsal meningocele
Thoracic Meningocele
Dural Ectasia, Meningocele associated with NF-1
Spinal Lipoma

- Premature disjunction of cutaneous ectoderm from neuroectoderm allows mesenchyme to contact inner portion of neural tube.
- Mesenchyme intended for other cell types regresses into fat when exposed to CSF
- As tube tries to close, mesenchyme → fat, may interferes with neurulation
- Lipomas contain ectodermal, mesodermal, endodermal elements = teratomas
  - Grows in proportion to overall adipose
Intraspinal Lipoma

- Mesodermal cells contact primitive ependyma
- LS, any level
- Usually intradural, rarely entirely intramedullary
Spinal Lipomas
Epidermoid / Dermoid

- Trapped epithelial mesenchymal tissue
- Developmental
- Post procedure
- May grow over time
- Related to teratomas sometimes
Epidermoid dermoid
Dermal Sinus → Proteus Infection of Dermoid, EDA
Spinal Teratoma
Lipomyelocele (with dermal sinus)
Placode within canal
Caudal Regression Syndrome

- Spectrum from coccygeal/LS hypogenesis → sirenomelia
- Infants of diabetic mothers, 1 in 7500 live births
- Assoc with:
  - OEIS (omphalocele, exstrophy, imperforate anus, spinal anomalies)
  - VACTERL (vertebral, renal, cardiac, limb anomalies with anorectal atresia & TEF)
  - Curarino triad (sacral hypogenesis, anorectal malformations, presacral teratoma or meningocoele)
Caudal Regression: Sirenomelia
Sirenomelia
Caudal Regression Syndrome

- Level determines type & severity
  
  **Type I:** above S1, even mid-thoracic, cord terminates high, blunted tip & deformation of cauda common, ant & post separation of roots

  **Type II:** Below S2, less severe, distal-most conus absent, tethered by tight filum, lipoma, or CSD with subcutaneous mass

  **Mild CRS:** only tip of conus absent, cord not tethered, may be missed
Type I CRS
CRS with fibrolipoma
Fibrolipoma, Tether, Sacral Hypoplasia – CRS Type I
Terminal Myelocystocele and Sacral Hypoplasia – CRS Type I
Segmental Spinal Dysgenesis

- Focal segment of lumbar or thoracic spine agenetic-markedly hypogenetic, cord segmentally disrupted, distal canal unaffected
- Distal cord large, focal kyphosis → early presentation
- Anomalous lower extremities, incontinence

- Frequency of CRS (11:1), indicates higher degree of susceptibility of the caudal cell mass to derangement
Segmental dysgenesis
Duplication Anomalies

- Diastematomyelia
- Diplomyelia
- Trimyelia
Split Cord Malformation

- Diastematomyelia (splitting) & diplomyelia (duplication), not radiologically distinguishable
- Type I: less common, osseous septum dividing two dural tubes & hemicords, assoc vertebral anomalies
- Type II: more common, hemicords in single dural tube ± fibrous septum
- Each hemicord contains a central canal, dorsal horn & ventral horn, each → 1 nerve root
- Can be asymmetrical, smaller easily missed
- Diplomyelia – two complete separate cords
Split Cord Malformations

- Septum causes tethering, ?SCM around CT junction under-recognized
- Despite lack of signs and sx, ~75% abnormal voiding on formal testing
- Cutaneous stigmata (Type I), F > M
- Presentation (any age): scoliosis, tethering, unilateral lower extremity weakness, wasting
Type I Diastematomyelia
Type I Diastematomyelia
Diastematomyelia Type 1
Type I Diastematomyelia
Diastem Type 1 with bony spur
Type II Diastematomyelia
Diastematomyelia Type 2
Trimyelia – fatal anomaly
Neurenteric Cyst

- Partial regression of neurenteric canal (of Kovalesky)
- Lined by secretory epithelium, contents similar to CSF or proteinaceous
- Intraspinal, ventral to thoracic cord, anywhere
- Associated with dysplastic vertebrae more than GI, respiratory anomalies
- Presents late teens, compressive signs & symptoms
Neurenteric Cyst and Canal
Neuroenteric Cyst
Neuroenteric Cyst
Neuroenteric Cyst
Dorsal Enteric Fistula

- Complex anomaly – split notochord
- Persistence of neurenteric Canal of Kovalesky connects neural tube and archenteron.
- Fistula from bowel to dorsal skin
- Extends through spine and canal, usually lumbar
- High assoc with other CNS & non-CNS anomalies (renal, GI, CDH, pulmonary)
Confused Sclerotomte Syndrome

- Transitional anatomy - Hox
- Rib and Vertebral Fusion anomalies, Klippel-Feil
- Sprengels Deformity
- Congenital Short Pedicles
Sclerotomes Split!
Sprengels Deformity

- Congenital High Scapula, Otto Sprengel 1891
- 75% female, Left > Right
- Assoc with other adjacent bony anomalies, Klippel Feil
- Omovertebral bone may connect scapula and lower cervical vertebra
CSS – Sprengels, Klippel Feil
Congenital Short Pedicles

- Very common, not well defined
- Greater than 2/1 AP vertebral body to pedicle?
- Less than 1 cm pedicle
- Most Neurorads use the eyeball test
Congenital Short Pedicles
CSS – Multiple fatal anomalies
CSS – with craniorachischisis
Klippel Feil Syndrome
C1 Anomaly
Conjoined Nerve Root

- Two nerve sets; same root sleeve
- Most common nerve root anomaly
- Uncommonly associated with bony anomalies
- Usually asymptomatic but may induce diagnostic confusion and subsequent therapeutic misadventure.
- 6-8% incidence in some studies, if so underdiagnosed
Conjoined Nerve Root
Spinal anomalies arise from diverse and complex etiologies and exhibit complex and overlapping appearances.

There will come a time when our understanding of the underlying mechanisms of both normal and abnormal spinal cord development will make these differences much more coherent than they are now.