Pediatric Spine Disorders

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Disclosure

• No conflict of interest
• Majority of the pictures were taken from Barkovich, Pediatric neuroimaging books

Introduction

• Understanding of the embryonic development of the spine is essential to understanding congenital/pediatric spine disorders
• Knowledge of embryology and the anatomy allows for proper identification of the spine condition
• Development of the vertebral columns and spinal cord are from two different originating cells and processes.
Development of the Spinal Cord and Nerves

- 3rd week of gestation
  - The ectoderm overlying the notochord thickens and forms the neural plate – folds at the lateral edge – becomes the neural folds and when it meets in the midline becomes the neural tube
  - Neural tube begins in the cervical region and proceeds cephalically and caudally
- Neural tube development
  - Neural plate development – 18 day
  - Cranial closure – 24 day (upper spine)
  - Caudal closure – 26 day (lower spine)

Formation of the vertebral column

Stage 1: Formation of mesenchymal vertebral column
- Migration of sclerotomes
- Differentiating into cephalic and caudal parts
- Development of intervertebral discs
- Development of the body of the vertebrae
  - Notochord degenerates and disappears when surrounded by the vertebral body
- Development of the neural arch

Formation of Vertebral body

- Stage 2: Formation of cartilaginous column
- Process of chondrification
- Stages of ossification of vertebral column
  - Primary ossification center
  - Secondary ossification center
Spondylolisthesis

- Common cause of back pain in children
- Most common location
  - Lower lumbar spine
- Etiology
  - Dysplastic
    - Abnormal development of posterior elements, intact pars interarticularis
  - Developmental
    - No history of repetitive trauma, hereditary
  - Traumatic
    - Acute vs. chronic (repetitive) stress
  - Pathologic
    - Insufficiency of posterior elements from underlying bone pathology
  - Other: Post surgical, neuropathic, degenerative
- Associated abnormalities: Scoliosis may develop

Spondylolisthesis

- CT scan better than MRI unless neurologic symptoms are present

STIR image – L5 on S1
Bilobed shaped

Congenital Spinal Stenosis

- Mostly in teenagers
- Male > Female
- May be asymptomatic and seen as incidental finding
- Most common symptom: depends on location
  - Cervical: (absolute AP diameter < 14 mm)
    - Radiating pain or numbness
    - If cord involvement – myelopathy
  - Lumbar: (absolute AP diameter < 15 mm)
    - Back pain + bladder/bowel
    - Neurogenic claudication
    - Cauda equina syndrome
Congenital Spinal Stenosis

- Short, thick pedicles
- Laterally directed laminae
- MR Findings:
  - T1: short AP Diameter
  - +/- hypointense facet
- Etiology:
  - Idiopathic
  - Achondroplasia
  - MPS
- DDx: Acquired SS
  - Normal pedicle length
MPS

Achondroplasia

Posterior element incomplete closure (spina bifida occulta)

- Failure of lamina/spinous process fusion
- No underlying neural or dural component
- Incidental finding on children imaged for back pain
- Most common location: LS junction
Posterior element incomplete closure

Imaging findings

- **T1WI**
  - Normal conus position, filum thickness
  - No abnormal neural, dural, or lipomatous tissue within posterior osseous defect
- **T2WI**: Same as T1WI
- **CT Findings**
  - Bone CT: Unfused spinous process/lamina approximate at midline
  Posterior elements do not fuse in midline; margins are rounded, well-corticated, and may overlap

Posterior element incomplete closure

Differential Diagnosis

- **Normal Progression of Ossification**
  - Normal L5 laminae may remain unfused until 5-6 years of age
- **Closed Spinal Dysraphism (CSD)**
  - Lipoma, tethered cord, lipomyelomeningocele, dorsal meningocele, dorsal dermal sinus
  - Search for clinical tethering, cutaneous stigmata
- **Surgical Laminectomy Defect**
  - Surgical history present; look for incision scar, denervation of paraspinal muscles, laminectomy defect

Vertebral segmentation failure

- Segmentation anomalies from aberrant vertebral column formation
- Most common: transitional vertebrae at TL and LS transition
- Genetics: Deranged Pax-1 gene expression
  - Abnormal notochord signaling
- Associated with syndromes
  - KFS
  - Spondylothoracic dysplasia
- Differential Diagnosis:
  - Chronic JRA
  - Chronic discitis
  - Ankylosing Spondylitis
Vertebral segmentation failure

• Symptoms:
  – Asymptomatic
  – Kyphoscoliosis
  – Myelopathy
  – Limb or organ involvement in syndromic cases
  – Respiratory failure
  – Neuroaxis abnormalities (40%)
• Severe cases in infancy or childhood

Vertebral segmentation failure

• MR Findings
• T1WI
  – Deformed, fused vertebral bodies and posterior elements
  – Normal marrow signal intensity
  – +/- Scoliosis
• T2WI
  – Findings similar to T1WI
  – +/- Scoliosis, cord compression

Coronal T2WI MR (L2-4) shows that 2 hemivertebrae (arrows) cause the abnormal curvature.
Failure of vertebral formation

- Majority asymptomatic or found during scoliosis evaluation
- Can be seen in syndromic cases
  - Detected in infancy
- Scoliosis is progressive
- Imaging appearance of vertebral formation failure determined by deficient vertebral body portion
  - Lateral formation failure – hemivertebrae
  - Anterior formation failure – sharply angulated kyphosis
  - Posterior formation failure (rare) – hyperlordotic curve
- Differential Diagnosis: Vertebral fracture, Inherited spinal dysplasias

Dorsal Dermal Sinus

- No gender predilection
- Midline or rarely paramedian dimple or pinpoint ostium ± pigmented patch, hairy nevus, or cutaneous hemangiom
- Differentiate from simple sacral dimple (< 2.5 cm from anus, extends inferiorly toward coccyx) and pilonidal sinus (low ostium, does not enter spine)
- High suspicion if dimple above intergluteal fold

Dorsal Dermal Sinus

- Decrease curvilinear tract through subcutaneous fat
- Dural tenting at dural penetration
- Location: LS most common
- May develop abscess or arachnoiditis
- May be associated with dermoid, epidermoid + tethering
Partial vertebral duplication

- Produces one or more supranumerary vertebra
  - Missing pedicle on hypoplastic side
- Location: thoracolumbar > cervical
- Differential Diagnosis:
  - Hemivertebra (single pedicle, ossification center fail to form)
  - Butterfly vertebra (2 pedicle, ossification center fail to unite)
  - Vertebral fracture (2 pedicle /level)
  - Inherited spinal dysplasias (MPS, Achondroplasia)

Dural Dysplasia

- Patulous dural sac with posterior vertebral scalloping
  - Primary meningeal dysplasia/weakness
  - Smooth C on imaging
- Symptoms:
  - Back pain – most common
  - Headache, incontinence and pelvic symptoms may occur
  - Seen in Marfan syndrome, NF1, EDS, Homocystinuria
- Differential Diagnosis:
  - Congenital Vertebral Dysplasia (MPS, OI, Achondroplasia)
  - Spinal Tumor or Syrinx (astrocytoma, ependymoma)
Dural dysplasia

- **T1WI**
  - Posterior vertebral scalloping, expansion of osseous spinal canal, patulous dural sac, +/- kyphoscoliosis
  - +/- Pedicular thinning, lateral meningocele(s)
- **T2WI**
  - Similar findings to T1WI
  - Best evaluates position of neural elements relative to dural ectasia
- **MRA:** +/- Arterial dissection or aneurysm (Marfan, Ehlers-Danlos syndrome)

3 Categories that produce vertebral scalloping

- Dural ectasia
- Increased intraspinal pressure
- Congenital vertebral dysplasia

Important for treatment, genetics and prognosis

Ventriculus Terminalis

- AKA 5th ventricle
- Cystic dilatation of distal central spinal cord canal
- No cord signal abnormality or enhancement
- Size: 2-4 mm (transverse); rarely exceeds 2 cm in length
- Conus terminates at normal level (T12 → L2)
Ventriculus Terminalis

- Incidental finding on imaging performed for unrelated indications
- Asymptomatic or nonspecific neurological symptoms
- Onset: < 5 years old, all ages
- Forms during embryogenesis (9th week) via canalization and retrogressive differentiation of caudal spinal cord
- Represents point of union between central canal portion formed by neurulation and portion formed by caudal cell mass canalization
- Usually regresses in size during 1st weeks after birth; persistence leads to identification in children or adults
- Differential Diagnosis:
  - Transient Dilatation of the Central Canal
  - Hydrosyringomyelia
  - Cystic Spinal Cord Neoplasm
  - Myelomalaecia

Ventriculus Terminalis

- T1WI
  - Hypointense fluid in dilated central canal of distal spinal cord
  - Cauda TERMINALIS terminates at normal level
  - No filum terminale thickening or lipoma

- T2WI
  - Hypointense (CSF intensity) fluid in dilated central canal of distal spinal cord
  - No septations within dilated central canal

- T1WI C+:
  - No nodular or ring enhancement

- 2.6% of normal children (under 5 years) have a visible ventriculus terminalis on MR