Congenital Spine Deformity

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Congenital Scoliosis
• 13 year old female
• Spinal curve
• No pain
• No neurological symptoms
• Menses for 1 year
• Healthy
• Spinal asymmetry
• No cutaneous manifestations
• Motor/Sensory examination normal
• Reflexes: Increased left achilles + clonus
Next Steps?
- X-ray
- MRI
  - Neurologic S & S
  - Foot Deformity
  - Pre-operatively
  - Butterfly vertebrae (cleft vertebrae)
    - Increased incidence of diastematomyelia
- Abdominal Ultrasound
- Pediatric Assessment
• Scoliosis due to abnormal vertebral development
• Genetics: most sporadic
• 20 % incidence of spinal dysraphism
  ➢ Tethered spinal cord
  ➢ Diastematomyelia
  ➢ Etc.
• 20 - 33 % incidence urinary tract anomalies
  ➢ Common undifferentiated mesenchyme
  ➢ Many have normal renal function
    • 6% life threatening
• 10 - 15 % CHD
- Classification
  - Segmentation
  - Formation
  - Mixed
### Defects of Segmentation

<table>
<thead>
<tr>
<th>Defect</th>
<th>Image Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>Block Vertebra</td>
<td>Bilateral failure of segmentation</td>
</tr>
<tr>
<td>Unilateral Bar</td>
<td>Unilateral failure of segmentation</td>
</tr>
<tr>
<td>Unilateral bar &amp; Hemivertebra</td>
<td>Unilateral failure of segmentation</td>
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</tbody>
</table>

### Defects of Formation

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<tbody>
<tr>
<td>Hemivertebra</td>
<td>Fully segmented</td>
</tr>
<tr>
<td>Semi-segmented</td>
<td>Semi-segmented</td>
</tr>
<tr>
<td>Incarcerated</td>
<td>Incarcerated</td>
</tr>
<tr>
<td>Nonsegmented</td>
<td>Nonsegmented</td>
</tr>
<tr>
<td>Wedge Vertebra</td>
<td>Unilateral partial failure of formation</td>
</tr>
</tbody>
</table>
• Natural History
  ➢ 75 – 90% Progressive

• Poor prognosis:
  ➢ Thoracic curve
  ➢ Unilateral unsegmented bar with ≥ 1 convex hemivertebrae
  ➢ Hemivertebrae at lumbosacral level
• Options?

➤ Orthotic Treatment
  • Poor response
    – Primary defect is in bone not soft tissue
  • Milwaukee brace preferred
    – underarm braces cause thoracic compression and reduce vital capacity?
  • Flexible or secondary curve treatment
  • No role in rigid deformity
• Surgical Treatment
  ➢ Indications:
   • Progressive curves not responding to orthosis
   • Certain anomalies?
  ➢ Early fusion may not stunt growth
• Posterior Fusion:
  ➢ Aim is curve stabilization
  ➢ Correction and immobilization with traction, cast or Milwaukee brace
  ➢ Instrumentation depending on age
• Combined Anterior and Posterior Fusion:
  ➢ Used for curves with poor prognosis
    • Good convex growth potential
    • Young age
  ➢ Pseudoarthrosis rate lower
  ➢ Correction and immobilization with traction, cast, brace, or instrumentation
• Convex Growth Arrest:
  ➢ Achieved by anterior and posterior convex fusion
    • anterior hemiepiphysodesis and posterior hemiarthrodesis
  ➢ Arrest convex growth, allow concave growth
  ➢ Indicated in cases with progresses scoliosis or severe scoliosis on presentation with single or adjacent convex hemivertebrae and chance for concave growth
• Hemivertebrae Excision:
  - Essentially anterior and posterior wedge osteotomy combined with correction and fusion
  - Used for rigid angulated scoliosis not amendable to other treatments
  - Usually lumbosacral due to lack of compensation
Congenital Kyphosis
• Kyphosis due to abnormal vertebral development
• Associated Anomalies:
  ➢ Intraspinal
  ➢ Cardiac and pulmonary
  ➢ Renal
  ➢ Auditory
  ➢ Klippel-Feil Syndrome
• Classification (Winter)
  ➢ Type I: Failure of Formation
  ➢ Type II: Failure of Segmentation
  ➢ Type II: Mixed
<table>
<thead>
<tr>
<th>DEFECTS OF VERTEBRAL-BODY SEGMENTATION</th>
<th>DEFECTS OF VERTEBRAL-BODY FORMATION</th>
<th>MIXED ANOMALIES</th>
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<tbody>
<tr>
<td>PARTIAL</td>
<td>ANTERIOR AND UNILATERAL APLASIA</td>
<td>ANTEROLATERAL BAR AND CONTRALATERAL QUADRANT VERTEBRA</td>
</tr>
<tr>
<td>ANTERIOR UNSEGMENTED BAR</td>
<td>POSTEROLATERAL QUADRANT VERTEBRA</td>
<td></td>
</tr>
<tr>
<td>COMPLETE</td>
<td>ANTERIOR APLASIA</td>
<td>ANTERIOR HYPOPLASIA</td>
</tr>
<tr>
<td>BLOCK VERTEBRA</td>
<td>POSTERIOR HEMIVERTEBRA</td>
<td>WEDGE VERTEBRA</td>
</tr>
<tr>
<td></td>
<td>BUTTERFLY VERTEBRA</td>
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</table>

This table illustrates various defects in vertebral body segmentation and formation, along with mixed anomalies.
- Natural History:
  - Type I: 7°/year
  - Increased incidence of neurologic involvement
  - Type II: 5°/year
• Orthotic Treatment
  ➢ Compensatory Curves

• Surgical Treatment
  ➢ Early
  ➢ Late
  ➢ Cord Compression
• Early Treatment of Mild Deformities
  ➢ < 55° and < 5 years = Posterior fusion

• Late Treatment of Moderate to Severe Deformities
  ➢ < 55° = Posterior fusion
  ➢ > 55° = Anterior and Posterior fusion
    • ± strut graft
    • ± instrumentation

• Late Treatment of Severe Deformities with Cord Compression
  ➢ Anterior Decompression
  ➢ Anterior strut graft
  ➢ Posterior fusion ± instrumentation
Ref: Lovell and Winter’s Pediatric Orthopaedics (5th edition)