Review of Scoliosis in Children

Thomas L Pommering, DO, FAAFP
Division Chief of Sports Medicine, Nationwide Children’s Hospital
Assistant Professor, Department of Family Medicine, OUHCOM
Associate Professor, Departments of Family Medicine and Pediatrics
The Ohio State University College of Medicine
Outline

- Definitions
- Classification
- Epidemiology
- Natural History
- Screening/Genetic Testing
- Radiographic Evaluation
- Clinical Evaluation
- Mgmt/Surveillence
- Indications for Referral
- Surgery
Is this scoliosis?

Curve measures 9 degrees
Definition

- **Scoliosis**
  - 3-D deformity of the spine
    - Lateral curvature (coronal plane)
  - Defined by **Cobb angle**
    - > 10 degrees
  - < 10 degrees = **Spinal Asymmetry**
Screening Recommendations

- **USPSTF** and **AAFP** recommend against routine screening of asymptomatic adolescents for AIS
- Canadian Task Force on the Periodic Health Examination – insufficient evidence
- **AAOS, Scoliosis Research Society, POSNA, AAP**
  - Girls ages 10-12 yrs, boys ages 13-14 yrs
- Bright Futures guidelines
  - Children and adolescents ≥ 8 yrs
Classification of Scoliosis

- **Neuromuscular**
  - Due to underlying neurologic or MSK problems
    - CP, myelomeningocele, muscular dystrophy
  - Result of muscle imbalance and lack of trunk control
- **Congenital**
  - Vertebral asymmetry from congenital anomaly
    - Hemivertebrae, failure of segmentation
  - Manifests before adolescence
- **Syndromic**
  - Genetic disorders
    - Marfan syndrome, OI, neurofibromatosis
  - May be first manifestation of underlying disease
Idiopathic Scoliosis

- **Idiopathic Scoliosis – 3 categories**
  - Infantile – ages 0-3 years
  - Juvenile – ages 4-9 years
  - Adolescent – ages 10 years and older

- **Adolescent idiopathic scoliosis (AIS)** – accounts for up to 85% of idiopathic cases
Pathophysiology of AIS

Fig. 38-8 Possible interrelationships of various factors that have been shown to have possible role in cause of idiopathic scoliosis. (From Lowe TG, Edgar M, Margulies JY, et al: Etiology of idiopathic scoliosis: current trends in research. J Bone Joint Surg 82A:1157, 2000.)
Radiographic Evaluation (2V)

• **Standing PA and Lateral** views of spine (C7 to sacrum, including iliac crests to get a Risser score)

• **Curve description:**
  – Direction of curve (L vs R) defined by **convexity**
  – Apical vertebrae – most deviated and rotated from midline
  – Typical AIS curve is **right thoracic, left lumbar**
Describing the curve in Osteopathic Terms

Naming by Side

Named according to the Convexity

Dextroscoliosis-curve that is sidebent left, scoliosis/convexity is to the right.

Levoscoliosis-curve that is sidebent right, scoliosis/convexity is to the left.
Lateral views – Spinal or Sagittal Balance

• Spinal imbalance can be related to:
  – pain
  – functional disability (ie: impaired PFTs)
  – Cosmetic appearance/self image

• Thoracic hyperkyphosis may indicate loss of neural axis
Types of Curves

- thoracic curve
- thoracolumbar curve
- lumbar curve
- double major curve
COBB ANGLE
Double Curve
 PLEASE ALIGN UPPER SIDE OF IPHONE TO UPPER VERTEBRA AND TOUCH SCREEN. 28°
“Therefore, we conclude that the iPhone is a clinically equivalent measuring tool to the traditional protractor”
Comparison between Oxford Cobbmeter and digital Cobbmeter for measurement of Cobb angle in adolescent idiopathic scoliosis

Yasser Allam¹ · Tarek El-Fiky¹ · Mahmoud Yasser Farghally¹ · Sameh Al-Sabagh¹ · Ahmed Ezzat Slam¹,²

Fig. 2 Smart phone running Tiltmeter application to select most inclined upper and lower endplates

Fig. 3 Smart phone running Cobbmeter application to measure Cobb angle between most inclined upper and lower endplates
Risser Staging
AIS Prevalence

- 10% of patients with AIS require treatment
- If both parents have AIS = their children are 50x more likely to require scoliosis treatment
- Higher concordance in monozygotic twins

Table 38-2 • Adolescent Idiopathic Scoliosis Prevalence

<table>
<thead>
<tr>
<th>Cobb Angle (degrees)</th>
<th>Female:Male</th>
<th>Prevalence (%)</th>
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</thead>
<tbody>
<tr>
<td>&gt;10</td>
<td>1.4-2:1</td>
<td>2-3</td>
</tr>
<tr>
<td>&gt;20</td>
<td>5.4:1</td>
<td>0.3-0.5</td>
</tr>
<tr>
<td>&gt;30</td>
<td>10:1</td>
<td>0.1-0.3</td>
</tr>
<tr>
<td>&gt;40</td>
<td></td>
<td>&lt;0.1</td>
</tr>
</tbody>
</table>

Natural History of AIS

Curve Stabilization vs Progression

• Before skeletal maturity
  – Progression occurs during rapid adolescent growth spurt (peak height velocity)
  – Younger you are, more likely to progress
• After skeletal maturity
  – Curves $\leq 30^\circ$ typically do not progress
  – Curves $>40^\circ$ have a higher risk of progression
  – Curves $>50^\circ$ progress $1^\circ$ per year after skeletal maturity into adulthood

Risk Factors for Progression

<table>
<thead>
<tr>
<th>Risk Factor</th>
<th>Likelihood Factor</th>
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<tbody>
<tr>
<td>Female gender</td>
<td>(3-10x)</td>
</tr>
<tr>
<td>Premenarchal status</td>
<td></td>
</tr>
<tr>
<td>Risser 0-1</td>
<td></td>
</tr>
<tr>
<td>Double curve (3x)</td>
<td></td>
</tr>
<tr>
<td>Thoracic curve (3x)</td>
<td></td>
</tr>
<tr>
<td>Higher curve severity (&gt;20°)</td>
<td></td>
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</tbody>
</table>
Curve Progression depends on:

1. Size of curve
2. Location of curve
3. Physiologic Age of Child
**Natural History**

- **Size of curve:**
  - larger curves progress to greater degree than smaller curves

- **Location of Curve:**
  - thoracic and double primary curves progress more than single lumbar or thoracolumbar curves

- **Physiologic age** (based on menarche & Risser status)
  - Risser stage 0-1: curves between 20-29 deg have > 65% risk of progression
  - Risser grade 2-4: curves between 20-29 deg have > 20% risk of progression
Table 2. Probabilities of Progression
Risser grade versus curve magnitude at detection

<table>
<thead>
<tr>
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<tbody>
<tr>
<td>Grade</td>
<td>5-19°</td>
<td>20-29°</td>
</tr>
<tr>
<td>0-1</td>
<td>22%</td>
<td>68%</td>
</tr>
<tr>
<td>2-4</td>
<td>1.6%</td>
<td>23%</td>
</tr>
</tbody>
</table>
Curve Progression in Mature Patients

- Patients less than 30 degrees do not progress regardless of curve pattern.

- Curve greater than 50 degrees progress at about 1 degree/year.
Chance of Progression in an IMMATURE Patient with Scoliosis

- Less than 30 degree curve = 20%
- 30-50 degrees = 60%
- Greater than 50 degrees = 90%
Clinical Evaluation - History

- Focused on determining etiology, risk for progression
- When was deformity noted, by whom?
- Rate of progression?
- Pain? (↑ likelihood of non-idiopathic etiology)
- Symptoms suggesting neuromuscular etiology? (muscle weakness, bowel/bladder problems, headache, neck pain)
- Signs of puberty and has pubertal growth spurt begun? (menarche, shaving in males)
- Family history of scoliosis?
Clinical Evaluation - Inspection

- Inspection
  - Shoulder/scapular height
  - Waistline symmetry
  - Distance of arms from trunk
  - Head centered over sacrum
  - Direction of curve (L vs R)
Describing the curve “Osteopathically”

Naming by Side

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Abdominal Reflex: The ONLY Thoracic-mediated reflex

- Tethered Cord
- Diastematomyelia
- Chiari Malformation
- Syrinx
Skin Exam

- Café-au-lait spots, axillary freckling (NF)
- Vascular, hypo/hyperpigmented lesions, hair patch over spine (spinal dysraphism)
- Dimpling in lumbosacral area (intraspinal tumor)
Adam’s Forward Bend Test

• Demonstrates rotational component
• Measure asymmetry with scoliometer
• Screening test performed in schools
  • Sensitivity 73-100%
  • Specificity 60-99%

FIGURE 1. Adams forward-bending test
Scoliometer

• Measures trunk rotation NOT Cobb

• Trunk rotation < 7° (scoliometer) – follow clinically q6 months

• Trunk rotation ≥ 7° – obtain x-rays
Treatment of Scoliosis

• Goal – Cobb angle $\leq 40^\circ$ at skeletal maturity
  – Curves $> 50^\circ$ may progress $1^\circ$ / year
• Treatment options
  – Observation
  – Bracing
  – Surgery
• Depends on degree of curve and further growth potential
  • Growth potential – age, menarchal status, skeletal maturity, Risser stage
    – Girls – 2 yrs post-menarchal, Risser 4
    – Boys – daily shaving, Risser 5
• PT, chiropractic treatment, electrical stimulation, biofeedback not effective
## Surveillance Strategy

<table>
<thead>
<tr>
<th>Curve (°)</th>
<th>Risser stage</th>
<th>X-ray frequency</th>
<th>Treatment</th>
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<tbody>
<tr>
<td>10-19</td>
<td>0-1, 2-4</td>
<td>Q6 months</td>
<td>Observation</td>
</tr>
<tr>
<td>20-29</td>
<td>0-1</td>
<td>Q3-4 months</td>
<td>Brace if progression, &gt;25°</td>
</tr>
<tr>
<td>20-29</td>
<td>2-4</td>
<td>Q6 months</td>
<td>Observation, consider bracing</td>
</tr>
<tr>
<td>30-40</td>
<td>0-2</td>
<td>Q4 months</td>
<td>Brace</td>
</tr>
<tr>
<td>40-50</td>
<td>0-2</td>
<td>Q4 months</td>
<td>Brace, consider surgery</td>
</tr>
<tr>
<td>&gt;50</td>
<td>0-2</td>
<td>Q6 months</td>
<td>Surgery</td>
</tr>
</tbody>
</table>
When to Refer?

Congenital, Infantile, and Juvenile curves should be referred always.

Adolescent scoliosis is up to the comfort of the primary care physician caring for the patient. Should always be referred if curve is > 20 degrees and skeletally immature.

Mature patients (>18 months postmenarchal and Risser IV or V with curves under 30 degrees do not need referral-low risk progression).
Advances in Imaging

- 3D spine imaging
- 8 x less radiation than traditional PA films
EOS Imaging
Be wary of curves that don’t follow the rules

- Left thoracic, right lumbar
- Rapid Progression (greater than 1 degree/month)
- Curve >20 degrees and < 9 years old
- Progressive curve in a boy
- Pain
- MRI if anything unusual
Left thoracic scoliosis

- Left thoracic curvatures are uncommon (1-2% of curves)
  - associated conditions: (33 %)
    - occult syrinx;
    - treatment of the scoliosis without recognition of syringomyelia and Chiari malformation can lead to paraplegia
      - Arnold-Chiari
      - spinal cord tumor
      - neuromuscular disorder
Duration of Treatment

- **Risser ≥ 3** – follow at least 1 year past skeletal maturity with x-rays every 6-9 months
- **Skeletally mature patients with Cobb angle <20°**
  - Reassurance, discharge from clinic
- **Skeletally mature patients with Cobb angle 30-40°**
  - Standing PA x-rays yearly for 2-3 years after skeletal maturity, then every 5 years throughout life***
Bracing

- Doesn’t correct curve already present at diagnosis
- Goal: prevent curve progression, ↓ need for surgery
- Indications:
  - Risser 0-2, Cobb angle 30-40°
  - Skeletally immature with progression $\geq 5^\circ$ during any 6-8 month observation period
  - Apex at or below T7
- Contraindications
  - Skeletal maturity (Risser 4-5)
  - Cobb angle $\geq 50^\circ$
Bracing was somewhat controversial...until

- BrAIST
- Multicenter, RCT, 242 pts
- Better Tx success for braced (72%) vs. control (48%)
- Study was stopped early
- Tx success = curve remained < 50 deg by skeletal maturity
- Bracing decreased progression of high risk curves (ie: surgery)
- + brace-dose relationship (>13 hr/d)
Final Protocol

- Population
- Treatment assignment
- Treatment
- Observation periods
- Data collection
- Exit from Study

Enrollment April 2007 to March 2011
25 institutions, US and Canada
Bracing

- TLSO (Boston brace)
- Charleston, Providence brace (night)
- CTLSO (Milwaukee brace)
  - Thoracic curve apex T8 or above
  - Double thoracic curves
  - Right thoracic/left thoracolumbar pattern
Growing Rods

- Useful in younger, skeletally immature
- Typically does not eliminate need for definitive spinal fusion
- Distractible spinal implant lengthened to accommodate spinal growth
- Traditional – surgical lengthening q6 months
- MCGR – magnetically controlled, outpt procedure

Figure 1: A single magnetically controlled growing rod fixed to a spine model. Cervical vertebrae at top, sacrum at bottom. Arrow shows enlarged portion containing the distraction mechanism.
Growing Rods

Figure 3: Radiographs of patient 2 (dual magnetically controlled growing rod)
Posteroanterior (A, C, E) and lateral (B, D, F) views of preoperative spine (A, B); immediately postoperative spine (C, D), and spine at latest follow-up (E, F). For all posteroanterior radiographs (A, C, E), the right-hand side of the patient is on the left-hand side of the image.
Return to Activity after Surgery

• Full participation in most sports once bone fusion achieved
  – 9-12 months
  – Collision sports (football, hockey, rugby) may be restricted
Return to Activity after Surgery

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  – 9-12 months
  – Collision sports (football, hockey, rugby) may be restricted
Take Home Points

• Get Radiographs if scoliometer > 7 deg
• Girls more likely to progress
• Good skin, back and neurologic exam is important
• Significant Pain is NOT typical in AIS
• Closely monitor:
  – Curves < 10-20 deg in skeletally immature pts
• Beware of:
  – Left sided thoracic curves
  – Left thoracic / Right lumbar curves
  – Rapidly progressing curves
Consider Referral for:

- Cobb angle 20-29 in skeletally immature premenarchal girl, boy 12-14 yrs
- Cobb angle >30° (any patient)
- Cobb angle progression >5°
- Findings suggestive of non-idiopathic scoliosis
Bibliography

- Scherl SA. Treatment and prognosis of adolescent idiopathic scoliosis. UpToDate, accessed August 14, 2016.
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