SCOLIOSIS: what is new and true?
Natural history, screening/evaluation and treatment

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Swedish Pediatric Specialty Update
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OBJECTIVES

• 1) review of entity adolescent idiopathic scoliosis
• 2) review the natural history of treated and untreated disease
• 3) review screening recommendations and delineating patients who warrant referral
• 4) review of treatment modalities
Scoliosis

• **Definition:** a lateral spinal curvature measuring at least **10 degrees** on x-ray

• Complex **3 dimensional** spinal deformity
Adolescent Idiopathic Scoliosis

- AIS: Scoliosis most often develops and progresses during the most rapid times of growth, typically between ages 10 - 15 years.

- NOT infantile or juvenile scoliosis which have different natural history profiles

- NOT congenital spinal deformities, neuromuscular scoliosis or spinal dysraphism
Adolescent Idiopathic Scoliosis

• Classic Teaching
  – Small curves are common
  – Progression occurs during most rapid times of growth
  – Female predominance
  – Big curves get bigger
  – Etiology unclear: familial predisposition, and?
  – Idiopathic scoliosis not typically painful
Adolescent Idiopathic Scoliosis

- **Mild** scoliosis curves are **common**, affecting 2-3% of the adolescent population, males and females equally

- Less than 0.1% of adolescents have curves greater than **40 degrees**.
  - Ratio of **females** to males **10:1**

<table>
<thead>
<tr>
<th>Cobb Angle</th>
<th>Female:Male</th>
<th>Prevalence</th>
</tr>
</thead>
<tbody>
<tr>
<td>&gt;10</td>
<td>1.4-2 :1</td>
<td>2 - 3 %</td>
</tr>
<tr>
<td>&gt;20</td>
<td>5:1</td>
<td>.3 - .5 %</td>
</tr>
<tr>
<td>&gt;30</td>
<td>10:1</td>
<td>.1 - .3 %</td>
</tr>
<tr>
<td>&gt;40</td>
<td>10:1</td>
<td>&lt;0.1%</td>
</tr>
</tbody>
</table>

Adolescent Idiopathic Scoliosis

- **RISK FACTORS FOR CURVE PROGRESSION**
  - Female gender
  - Skeletal immaturity
    » If curve greater than 30 degrees before peak height velocity, strong likelihood of progressing to surgical magnitude  
      Sanders at al  
      JBJS 2008
  - Curve magnitude
    » Big curves get bigger
  - Curve location
    » Thoracic > double major > lumbar
      » Thoracic curves and double major curve have progression rate 25-30% compared to single lumbar or thoracolumbar curves which progressed 10-15%
      Soucacos et al  
      Eur Spine J 1998

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Table 1. Incidence of Progression as Related to the Magnitude of the Curve and the Risser Sign

<table>
<thead>
<tr>
<th>Risser sign</th>
<th>5- to 19-degree curves</th>
<th>20- to 29-degree curves</th>
</tr>
</thead>
<tbody>
<tr>
<td>Grade 0 or 1</td>
<td>22</td>
<td>68</td>
</tr>
<tr>
<td>2, 3, or 4</td>
<td>1.6</td>
<td>23</td>
</tr>
</tbody>
</table>

Adolescent Idiopathic Scoliosis

- **How do we measure skeletal maturity in relation to scoliosis**
- **Risser stage: 0-5** (pelvis image on scoliosis xray)
- **Sanders Stage 1-8** (on hand xray)

<table>
<thead>
<tr>
<th>Sanders Stage</th>
<th>Radiographic Features</th>
<th>Risser Stage</th>
<th>Radiographic Features</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. Juvenile slow</td>
<td>Digital epiphyses are not covered</td>
<td>0</td>
<td>No ossification of the epiphysis</td>
</tr>
<tr>
<td>2. Preadolescent slow</td>
<td>All digital epiphyses are covered</td>
<td>0</td>
<td>No ossification of the apophysis</td>
</tr>
<tr>
<td>3. Adolescent rapid (early)</td>
<td>Most digits are cupped. 2nd-5th MC epiphyses are wider than metaphyses</td>
<td>0</td>
<td>Triradiate cartilage open (peak height velocity)</td>
</tr>
<tr>
<td>4. Adolescent rapid (late)</td>
<td>Any of distal phalangeal physes are clearly beginning to close</td>
<td>0</td>
<td>Triradiate cartilage remains (open growth plates in the long bones)</td>
</tr>
<tr>
<td>5. Adolescent steady (early)</td>
<td>All distal phalangeal physes are closed, while others are open</td>
<td>0</td>
<td>Triradiate cartilage closed (menarche in female patients)</td>
</tr>
<tr>
<td>6. Adolescent steady (late)</td>
<td>Middle or proximal phalangeal physes are closing</td>
<td>$\geq 1$</td>
<td>Ossification of the iliac apophysis ranges from 25% to 75%</td>
</tr>
<tr>
<td>7. Early mature</td>
<td>Only distal radial phys is open. MC physeal scars may be present</td>
<td>4</td>
<td>100% ossification of the iliac wing, with no fusion to iliac crest</td>
</tr>
<tr>
<td>8. Mature</td>
<td>Distal radial phys is completely closed</td>
<td>5</td>
<td>Fusion of the iliac apophysis to the iliac crest (cessation of growth)</td>
</tr>
</tbody>
</table>

MC indicates metacarpal.
Adolescent Idiopathic Scoliosis

- Risk of Curve Progression

If curve is > 30 degrees before peak height velocity, there is strong likelihood of curve progressing to surgical range.
## Adolescent Idiopathic Scoliosis

### Age at Detection

<table>
<thead>
<tr>
<th>Curve Magnitude</th>
<th>10-12</th>
<th>13-15</th>
<th>16 year</th>
</tr>
</thead>
<tbody>
<tr>
<td>&lt;19 degrees</td>
<td>25%</td>
<td>10%</td>
<td>0%</td>
</tr>
<tr>
<td>20-29 degrees</td>
<td>60%</td>
<td>40%</td>
<td>10%</td>
</tr>
<tr>
<td>30-39 degrees</td>
<td>90%</td>
<td>70%</td>
<td>30%</td>
</tr>
<tr>
<td>&gt;40 degrees</td>
<td>100%</td>
<td>90%</td>
<td>70%</td>
</tr>
</tbody>
</table>

- Weinstein JPO, 2019
- 50 yr follow up of untreated cohort of AIS patients
AIS: ETIOLOGY

- **ETIOLOGY** unclear, but thought to be attributable to:
  - genetic factors
    - linkage of scoliosis and sites on chromosomes 6,10,12, and 18
  - nutrition
    - Low Vit D levels associated with increased risk of curve progression
  - early exposure to toxins
    - Methyl donors, polyphenols, zinc, selenium, and Vit A may trigger methylation of DNA
  - hormonal dysregulation
    - **Melatonin** levels decrease significantly with increased curve progression. **Melatonin** receptor polymorphisms seem to have a role in development of AIS.
AIS: ETIOLOGY

• ETIOLOGY:

  – GENETIC FACTORS

    • Danish Twin Registry study found that monozygotic twins had a higher concordance rate (.4) than dizygotic twins (0.05)
      » Simony et al. Spine 2016

    – Study of family pedigrees of 131 patients with AIS found familial scoliosis connection in 127, and suggested that 1 or 2 majors genes likely responsible. The genes under investigation encode for extracellular matrix components and hormone receptors.
AIS: PAIN

- Adolescent **idiopathic** scoliosis is typically **NOT** painful
  
  - If a scoliosis patient has more than mild back discomfort, a thorough evaluation is warranted.
    - In one study, 23% of pts (560/2442) with presumed AIS presented with back pain at time of diagnosis. 9% of this cohort (48/560) were found to have an underlying associated condition such as spondylolisthesis, syringomyelia, tethered cord, disc herniation or tumor.
      » Ramirez et al. JBJS 1997
      » The prevalence of back pain in children who have idiopathic scoliosis
Adolescent Idiopathic Scoliosis

- Clinical evaluation
Adolescent Idiopathic Scoliosis

CLINICAL EVALUATION

- ADAM’S forward bend test
- “Inclinometer/level” can be helpful
  - Tool or app
  - Measures the angle of trunk rotation

- XRAYS appropriate for curves measuring 7 degrees or more with level/inclinometer or in setting of curve that is progressing
Adolescent Idiopathic Scoliosis

• **Clinical Evaluation**
  
  – Presentation of Adolescent Idiopathic Scoliosis: the Bigger the Kid, the Bigger the Curve
    » Goodbody et al JPO 2017

  – 150 pts, 50 each normal wt, overweight and obese
  – Average curve at presentation
    • NI wt: 18.1 degrees, over wt: 23.9 degrees, obese: 24.5 degrees.

  – Overweight and obese patients with AIS present at significantly larger curve magnitudes and significantly higher degrees of skeletal maturity.
  – More over, these patients were significantly more likely to present with very large curves and all patients presenting with surgical range curve were either overweight or obese

  – CONSIDER OBTAINING XRAYS WHEN INCLINOMETER READING 5 DEGREES IN OVERWEIGHT PATIENTS
Adolescent Idiopathic Scoliosis

- Position Statement on Screening for AIS
- US Prevention Services Taskforce in 2004 concluded that the current evidence is insufficient to assess the balance of benefits and harms of screening for children and adolescents aged 10-18
- AAOS, POSNA, SRS, and AAP believe that recent high quality studies demonstrate that non operative interventions such as bracing and scoliosis specific exercises can decrease the likelihood of curve progression to the point of requiring surgical treatment and thus have generated following best screening recommendation

- FEMALES should be screened TWICE, once at age 10 and again at 12.
- MALES should be screened ONCE, at age 13 or 14.
AIS: Xray Evaluation

• **RADIOLOGIC EVALUATION**
  - PA/lateral scoliosis xrays
    - **EOS** low dose xrays
      - 50-80% less radiation compared to traditional xrays
        - Only available on one machine at Laurelhurst campus SCH.
        - Need baseline routine xrays to look for fine detail
  - Referral to orthopedics
    - Curves **15-20 degrees** or more in individuals less than 12 years old
    - Greater than or equal to **30 degrees** any age.
AIS: Natural History

- **Natural History of Adolescent Idiopathic Scoliosis**
  - MORTALITY
  - CARDIOPULMONARY FUNCTION
  - BACK PAIN
  - CURVE PROGRESSION
  - QUALITY OF LIFE

- **Mortality**
  - 2X mortality rate
  - 60-80% patients died of cardiopulmonary disease
    - Nachemson, 1968
    - Nilsonne, 1968
    - Ascani, 1986

- **MORTALITY FOR PATIENTS WITH ADOLESCENT IDIOPATHIC SCOLIOSIS IS EQUAL TO THE GENERAL POPULATION**
  - Peterson, 1992
  - Weinstein, 2003
AIS: Natural History

• **Natural History of Adolescent Idiopathic Scoliosis**
  - MORTALITY: equal to the general population
  - CARDIOPULMONARY FUNCTION
  - BACK PAIN
  - CURVE PROGRESSION
  - QUALITY OF LIFE

• **Cardiopulmonary Function**
  - The larger the curve, the less lung volume and chest wall compliance
  - Patients with curves > 50 degrees have reduced vital capacity and more shortness of breath, but no apparent increase in respiratory disability

  - Cardiopulmonary death risk may be increased in patients with thoracic curves >100 degrees

  - SEVERE CARDIOPULMONARY COMPROMISE IS EXCEEDINGLY RARE IN AIS

  
  Weinstein JAMA 2003
  Ponseti JBJS 1950
AIS: Natural History

- **Natural History of Adolescent Idiopathic Scoliosis**
  - MORTALITY: equal to the general population
  - CARDIOPULMONARY FUNCTION: no apparent increase in respiratory disability
  - BACK PAIN
  - CURVE PROGRESSION
  - QUALITY OF LIFE

- **Back Pain in AIS**
  - 40-90% prevalence of back pain with variable severity

- 61% of scoliosis patients vs 35% of (adult) controls with reported back pain
- Pain was similar in frequency, intensity and duration when compared to controls
- Radiographic arthritis did not correlate with back pain
  - Weinstein, JAMA 2004
- AIS patients will likely have more back pain but no greater disability from back pain.
AIS: Natural History

• **Natural History of Adolescent Idiopathic Scoliosis**
  – MORTALITY: equal to the general population
  – CARDIOPULMONARY FUNCTION: no apparent increase in respiratory disability
  – BACK PAIN: 2x normal, mild
  – CURVE PROGRESSION
  – QUALITY OF LIFE

• **Curve Progression**

• **Before skeletal maturity**
  – Gender female > male
  – Curve magnitude
  – Remaining skeletal growth: Risser sign, menarchal status, bone age/Sanders, triradiate status

• **After skeletal maturity**
  – Curves less than 30 degrees do not progress
  – Curves > 50 degrees in thoracic spine will progress 1 degree/year
  – Curves >40 degrees in the lumbar spine with progress 1 degree/year
AIS: Natural History

- **Natural History of Adolescent Idiopathic Scoliosis**
  - MORTALITY: equal to the general population
  - CARDIOPULMONARY FUNCTION: no apparent increase in respiratory disability
  - BACK PAIN: 2x normal, mild
  - CURVE PROGRESSION: 1 degree/year
  - QUALITY OF LIFE

- **QUALITY OF LIFE as adult**
  - Most studies show equal marriage rate at 82-98%
  - Scoliosis patients equivalent to controls in DEPRESSION INDEX
  - Older patients with untreated AIS are much less satisfied than controls with body image
  - 1/3 feel that curvature has restricted them in some way (purchasing clothes, physical ability, self consciousness)
  - Some think they are less healthy and restricted in physical and social activities with “real psychological disturbances”
AIS: Natural History

- Natural History of Adolescent Idiopathic Scoliosis
  - MORTALITY: equal to the general population
  - CARDIOPULMONARY FUNCTION: no apparent increase in respiratory disability
  - BACK PAIN: 2x normal, mild
  - CURVE PROGRESSION: 1 degree/year
  - QUALITY OF LIFE: “cosmetic” concerns
AIS: Treatment

- **TREATMENT:** classic teaching
  - Curves 30 degrees or less: **OBSERVATION**
  - Curves 30 degrees- (45) 50 degrees: **BRACE**
  - Curves > 50 degrees in skeletally immature or > 60 degrees in the skeletally mature: **SURGICAL STABILIZATION**
AIS Treatment: BRACING
AIS Treatment: BRACING

Rigo Style Brace: Axially Asymmetric

TLSO - Rotation Control
AIS Treatment: BRACING

- Bracing has been shown to **slow** or **prevent** the progression of spinal deformity **during growth**
  - A brace WILL NOT produce lasting correction
  - Prevention of curve progression in the growing individual may prevent need for surgery

- Bracing does not make the curve go away
AIS Treatment: BRACING

• Who needs to be braced?
• SRS Bracing Criteria
  – Scoliosis curves greater than 30 degrees
  – OR greater than 25 degrees with history of documented progression

• Patients must have growth remaining
  – Within 1 year of menarche
  – Risser 2 or less
AIS Treatment: BRACING

• Once initiated, bracing is worn until skeletal maturity
  
  • 18-24 months post menarchal, Risser IV and no change in height for 6 months.

• Brace wear is often 12-36 months
AIS Treatment: Bracing

- New data supporting efficacy of bracing
- **BrAIST study  NEJM 2013**
  - Treatment success as defined by preventing curve progression to surgical range/ 50 degree curve
    - 72% success with bracing
    - 48% in non braced group
  - Positive association between hours of brace wear and rate of treatment success.
  - Of those patients who were treatment successes, 13 hours of brace wear/day resulted in 90-93% success.
  - 0-6 hr/day: no benefit/same as observation

- Number needed to treat 3
AIS Treatment: BRACING

• **Compliance is challenging**
  – On average, patients wore braces 65% of prescribed amount of time
  – Only 15% of patients demonstrated a highly compliant (>90%) brace wear routine.
    • Patients on parttime (16 hr/day) regimen actually demonstrated worse compliance (58%) compared to those prescribed full time wear (71%)
    DiRaimond et al JPO 1988

• **Bracing is not effective for everyone**
  – Less effective for older patients, males, heavier patients
AIS: Treatment

• Brace Treatment Summary
  – Brace TLSO is an effective treatment for adolescent idiopathic scoliosis
  – Bracing can only slow or prevent progression
  – Brace candidates
    • Must have growth remaining
    • Curves between 20-45 degrees
  – Wear schedule matters
    • 13 or more hours/day is most effective
  – Brace is better than observation along in preventing progression and avoid surgery
  – 3D bracing showing promise; more study needed.
AIS: Treatment

- Physiotherapeutic Scoliosis Specific Exercises
  - Schroth PT
  - SEAS: Scientific Exercise Approach to Scoliosis
    - Alignment training process
    - Develop deep postural muscles to improve scoliotic alignment
    - Addresses all 3 dimensions of the POSTURAL (flexible) component of the structural scoliosis
- Ideally to prevent or slow progression of AIS
  - Prior to bracing to potentially avoid need for brace
    - Small curve 20-25 degrees in skeletally immature < Sanders 6
  - With bracing to improve compliance with bracing and maintain core strength during bracing
AIS: Treatment

- **Physiotherapeutic Scoliosis Specific Exercises**

- **Effectiveness of Schroth exercises during bracing in adolescent idiopathic scoliosis: results from preliminary study**  
  Kwan et al  Scoliosis and Spine Disorders, 2017
  
  - Compared bracing alone vs brace + PSSE
  - Rates of progression were > 2x higher in the brace alone group
  - 50% progression vs 21% progression with added PSSE

- **Schroth compliance study**  
  Vitale et al 2017
  
  - 85 patients
  - Compliance with home exercise program = > 80 min/week after >10 hrs of training
  - Non compliant patients had more curve progression
AIS: Treatment

• Physiotherapeutic Scoliosis Specific Exercises
  – Recent systematic reviews are split on recommendations for PSSE
  – Few studies
  – Most studies are small, often fewer than 50 pts

– Barriers
  • Access: not many trained providers
  • Cost:
    – not covered by all insurance plans
    – Requires purchase of equipment and room to install it
  • Compliance: needs right patient and family
AIS: Treatment

• Vitamin D and calcium in AIS
  – Does Vit D and Ca+ supplementation affect curves in AIS?
    » Lam et al, 2018
  – Randomized, double blind, placebo controlled study over 2 years
  – AIS patients 11-14yrs Tanner <IV
  – Femoral neck bone mineral density Z scores <0
  – Cobb angles > 15 degrees
  – Study design split all eligible girls with AIS into 3 groups
    • Group 1: Placebo
    • Group 2 low dose 400 IU Vitamin D and 600 mg Ca
    • Group 3 High dose 800 IU Vitamin D and 600 mg Ca
  – The lowest rate of curve progression was seen in the high dose group
    • Group 1 46.7% curve progression
    • Group 2 24.4% curve progression
    • Group 3 21.7% curve progression.
AIS Treatment: Surgery

• Posterior Spinal Fusion: gold standard
  • **Indications:**
    – large, progressive curves
      » > 40-45 degrees in growing individuals
      » > 50-60 degrees in skeletally mature patients
  • **Goals**
    – Prevent long term progression in adulthood
    – Achieve maximum deformity correction
    – Maintain optimal coronal and sagittal balance
AIS: Treatment

• Outcome measures
• Orthopedist
  – Spinal alignment
  – Xray measurements

• Patient
  – Asymmetries of shoulders, ribs, flank and breasts
AIS: Treatment

Long term outcome of Surgery for AIS

Long Term Clinical Outcomes of Surgery for Adolescent Idiopathic Scoliosis 21 to 41 Years Later

Akazawa et al, SPINE 2012

256 patients treated surgically between 1968 and 1988

- Control group 130 healthy volunteers; no scoliosis, no spine surgery

Based on SRS 22 responses, when compared to control group

- AIS patients had significantly decreased FUNCTION (AIS 4.3 +/- 0.6, CTR 4.7 +/- 0.5, P < 0.01)
- AIS patients had decreased SELF IMAGE (AIS 3.0 +/- 0.8, CTR 3.7 +/- 0.5, P < 0.01)
- Both groups were similar with respect to PAIN (AIS 4.3 +/- 0.6, CTR 4.2 +/- 0.5, P = 0.14)
- Both groups were similar with respect to MENTAL HEALTH (AIS 3.9 +/- 0.9, CTR 3.7 +/- 0.7, P = 0.14)

Mean follow up was 31.5 years. Generation implants

Although scores for function and self image were significantly lower in AIS patients, the clinical significance of these differences is unknown

Better post operative outcomes expected with 3 dimensional “modern” constructs used today.
AIS: Treatment

• Spinal Fusion
  – Complications
    • Neurologic injury 1:1000
    • Pseudarthrosis 1-2%
    • Infection 1-2%
    • Flat back syndrome
    • Crankshaft
    • SMA syndrome
  – Residual asymmetry of flank, breast, torso, shoulders
AIS: Treatment

- Future of AIS Treatment
  - **Fusionless procedures**
    - Guided growth vertebral body stapling
    - Anterior tethering
  - These methods control the patients remaining spinal growth to achieve curve correction.
  - Avoid the morbidity associated with spinal fusion.
AIS: Treatment

• **Operative vs Non operative outcomes**
  
  – SRS-22r Scores in Nonoperated Adolescent Idiopathic Scoliosis Patients With Curves Greater Than Forty Degrees
    » Ward et al SPINE 2017
    • 190 patients, >18 yrs of age, with curves > 40 degrees who elected to forgo surgery
    • 166 patients who underwent surgery
    • No statistically significant differences were found between group on the PAIN, FUNCTION, or MENTAL HEALTH domains of the SRS 22r. Differences in favor of the operative cohort were found for SELF IMAGE, SATISFACTION and TOTAL SCORE.

**Conclusion**
• No meaningful clinically significant differences in SRS 22r scores at an average of 8 year follow up.
• These data in conjunction with an absence of long term evidence of serious medical consequences with non surgical management of curve >40 degrees should encourage surgeons to reevaluate the benefits of routine surgical care.
AIS Summary

• AIS is a 3 dimensional deformity of the spine
• Most patients with small curves will never require active treatment
• Young, skeletally immature girls are most at risk for curve progression, and they require close monitoring.
• Scoliosis screening recommendations
  – FEMALES should be screened TWICE, once at age 10 and again at 12.
  – MALES should be screened ONCE, at age 13 or 14.
  – Xrays should be obtained for scoliometer measurements of 7 degrees or greater. (5 degrees or greater in the obese patient)
  – Referral to orthopedics for curves 15-20 degrees in patients 12 years or less
  – Referral to orthopedics for patients of any age with curves greater than 30 degrees.
AIS Summary

• Bracing is an effective in preventing curve progression and it’s effect is dose dependent

• Scoliosis specific physical therapy exercises can improve the flexible component of scoliosis. There is evidence that they may reduce the rate of progression and augment the effectiveness of bracing.

• Surgical fusion is an option for those curve that become severe. Posterior spinal fusion is the current gold standard.

• In the future, treatment may allow scoliosis correction without fusion and its associated morbidity
THANK YOU FOR YOUR ATTENTION