Adolescent Idiopathic Scoliosis - Vertebral Body Tethering

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Disclosure

- Orthopediatrics
  - Consultant, SAB, Royalties
- Biomarin
  - Consultant, PI

Disclosures regarding VBT

- Required to inform our patients/physicians:
  - Experimental procedure
  - Implants are not yet approved by the FDA – safe/effective for tethering
- No prospective clinical investigation
  - Approval by the FDA => prospective clinical study as an investigational device exemption (IDE)
Overview

- Scoliosis
  - Lateral (coronal) deviation of the spine
    - True scoliosis ≥10 degrees (Cobb angle)
    - <10 degrees: spinal asymmetry
  - Three dimensional deformity:
    - Sagital (hypokyphosis)
    - Axial rotation

Overview of Scoliosis

- Scoliosis: categories
  - Idiopathic (EOS, AIS)
  - Neuromuscular (CP, SMA, SCI)
  - Syndrome related (NF, Marfan)
  - Congenital
- Idiopathic scoliosis
  - Exact cause unknown
  - Largest subset of patients with structural scoliosis: 80%

Overview

- Idiopathic scoliosis: age of onset important
  - Significant differences in natural history and treatments
  - Early onset scoliosis
    - Infantile (0-3 years) => .5%
    - Juvenile (4-10 years) => 10.5%
    - Adolescent (11-17 years) => 89% of AIS cases
Majority of AIS curves do not require treatment

- 2% - 3% for curves >10°
- 0.3% for curves >25° (need a brace)
- 0.1% for curves >50° (need surgery)

Epidemiology

- Sex prevalence in idiopathic scoliosis
  - Female:Male
  - 6-10 deg – 1:1
  - 11-20 deg – 1:1
  - > 21 deg not requiring treatment – 5:1
  - Scoliotic curves requiring intervention – 7:1
  - Curve progression more common in females

Fraternal Twins

- 57 degrees
- 30 degrees
**Etiology**

*Genetic component* but most likely multifactorial

- Many other factors implicated but not proven:
  - Melatonin deficiency
  - Growth hormone
  - Asymmetric spinal growth
  - Neurologic dysfunction
  - Vestibulo-ocular system dysfunction
  - Asymmetric response to vibratory stimuli

Adolescent idiopathic scoliosis. Monograph Series 26

**Vertebral Body Growth**

- Anterior spinal column overgrowth
  - Increase in growth of anterior spine compared with posterior elements
  - Spine rotates to maintain sagittal balance
  - Buckles in response to extra column length
  - Accounts for all three planes of deformity - hypokyphosis

**Genetics**

- Genetic testing for IS
  - Scoliscore AIS prognostic test
    - DNA based saliva test
    - 53 genetic markers said to be associated with curve progression
    - 1-200: < 50 = low risk > 180 = high risk
    - Reliability questioned
      - Launched: 12/08
      - Dr. James Ogilvie – chief investigator

- Specific genes continue to be elusive
Pathophysiology

- **Coronal plane deviation**
- **Axial plane**
  - Spinous processes rotate toward the concavity of curvature
- **Sagittal plane**
  - Hypokyphosis

Pathophysiology

- Compression and distraction forces induce bony remodeling: Hueter-Volkmann principle
  - **Convex**
    - Vertebral body longer
    - Laminae broader and widely separated
  - **Concave**
    - Vertebral body shorter
    - Laminae narrow and close
- Pedicles on concave side shorter, more narrow transverse width

Fusionless Strategies

- **Guided growth techniques** have been used reliably in limb deformity
- The spine is actively growing during the natural history of AIS
- Could this concept of **guided growth** be applied to gradually correct an AIS curve over time?
Natural History

- Agabegi et al., JAAOS 2015
  - Review of literature on natural history studies of AIS
  - Rarely, progression to curves > 100 degrees
  - When this does occur, can lead to cardiopulmonary issues
  - Iowa studies from 1981-83: 1 degree per year progression of curves that reach 50 degrees at skeletal maturity
  - 30 degree curves rarely progress past skeletal maturity
  - Back pain is common in adulthood; thoracolumbar and lumbar curves
  - Cosmetic concerns are significantly increased as an adult
  - No increase in mortality and patients function well in adulthood

Lung Function

- Pehrsson et al., Thorax 1991 examined 24 patients with unfused IS
  - 20 yr follow up, strongest predictors of respiratory failure were low vital capacity and scoliosis curve greater than 110 degrees
  - Respiratory failure may develop even without progression of the scoliosis curve due to underlying disease

Pulmonary Function

- Thoracic curves:
  - Curves > 60° => 68% TLC (usually asymptomatic)
  - Curves > 100° => 50% TLC (symptomatic)

- No correlation between:
  - Thoracolumbar / Lumbar curves and pulmonary function
Natural History

- Before skeletal maturity
  - Growth potential, curve magnitude, curve type
  - Curves > 25° before maturity tend to progress

- After skeletal maturity
  - Curves ≥ 50° after maturity tend to progress ~ 1°/year into adulthood

Screening

- AIS Screening
  - Many patients evaluated for AIS will be referred from school screening programs or pediatricians
  - Controversy regarding screenings

- USPSTF (2004)
  - "The U.S. Preventive Services Task Force (USPSTF) recommends against the routine screening of asymptomatic adolescents for idiopathic scoliosis. D recommendation."

Screening – in response to USPSTF

- AAOS, SRS, POSNA, and AAP believe that screening examinations for spine deformity should be part of the medical visit for females at age 10 and 12 years, and males once at age 13 or 14 years."
Screening

- School/pediatrician screening
  - Scolimeter quantifies/establishes a threshold for referral
    - Measures in degrees of deviation from horizontal
    - Relies on axial rotational of scoliotic spine
    - 7 degrees is the recommended threshold for referral (Bunnell 1993)
      - 5 degrees → 100% sensitive; 47% specific
      - 7 degrees → 83% sensitive; 86% specific
    - Referral Rate: 3-30%

Screening

- 8°

Diagnosis

- AIS is a diagnosis of exclusion
  - Should pay careful attention to detail in history, physical examination, and radiographs
  - Don’t ignore the red flags
History

- Pain??
  - Localize and quantify – constant, well-localized back pain

- Neurologic Symptoms
  - Weakness / clumsiness; incontinence

- Growth history:
  - Onset of menses (girls)
  - Axillary and facial hair (boys)
  - Peak height velocity occurs approx 6-12 months prior to menses or axillary/facial hair in boys

- Family history:
  - Idiopathic scoliosis, syndromes or other conditions

Back Pain and Scoliosis

- Present 23% at time of initial evaluation*
  - 9% will have an identifiable cause*

- Identifiable causes:
  - Spondylolysis
  - Scheuermann’s disease
  - Disk, syrinx, or tumor

- Extensive *initial* work up not indicated!!!

*Ramirez et al. JBJS 1997; 79: 364

Clinical Findings

Thoracic cage asymmetry

- Concave: ribs rotate anteriorly (chest wall prominence/breast asymmetry)
- Convex: ribs rotate posteriorly (rib hump)
- Upper thoracic curve: left trapezial fullness
Clinical Findings
- Shoulder height
- Trunk Shift
- Rotation
- Rib or scapula prominence
- Sagittal deformity

Red Flags
- Don’t ignore the red flags
- Physical Examination
  - Foot deformity (unilateral)
  - Tight hamstrings
  - Abnormal curvature (left thoracic)
  - Abnormal reflexes

Radiographic Evaluation
- Imaging (PA/lateral)
  - Upper left thoracic curve
  - Right (main) thoracic curve
  - Lower left lumbar/thoraco-lumbar curve
  - Pedicle asymmetry
  - Greatest deformity at curve apex
  - Trunk shift with relatively well maintained coronal balance
  - Hypokyphosis/apical lordosis
King Classification

- King classification (1983) described 5 curves
- Provided recommendations for correction
- Newer implants = improved ability to correct triplanar deformity
- Revealed shortcomings in King classification

Lenke Classification

- Lenke et al. JBJS 2001
  - Addresses multiple planes (coronal, sagittal)
  - 6 main Lenke curve types
    - Lumbar (A, B, C) and sagittal (C, N, +) modifiers
    - 42 possible curve types
  - Used reliably for preoperative planning
    - "structural" versus "nonstructural" curves
    - What levels should be instrumented
    - How much correction to achieve

Surveillance

- Long-term exposure to radiation (repeated XR) has deleterious effects in this population
  - Elevated breast cancer risk (Hoffman 1998, Ronckers 2000) => .22%
  - Elevated leukemia risk (Bone 2000)
  - Some risk addressed by PA projections and lead shielding
Surveillance

- EOS imaging
  - Simultaneous 2D standing images and creates a 3D bone rendering of patient's spine
  - Pre and post-operative analysis
  - 50-fold reduction radiation exposure for pediatric scoliosis patients

Treatment

- Treatment options
  - Observation
  - Non-surgical treatment: bracing
  - Surgical treatment: fusion vs VBT

- Before recommending a treatment
  - Curve magnitude?
  - Growth potential (risk of progression)?
  - Curve flexibility?

Are we skeletally mature??

- Risser Sign
- Tanner Stage
- Peak height velocity
- Triradiate cartilage closure
- Menarchal age
- Sanders stages
Growth Potential

- How much growth remains?
- Pre or post-menarchal?
- Triradiate cartilage closed?
- Reached peak height velocity?

Risser, CORR 1958
Based on ossification of iliac apophysis
Ossifies lateral to medial
Risser 0 (no ossification) and Risser 1 most at risk for curve progression

Growth Potential

- Sanders staging

Sanders et al., JBJS 2008
Cohort of 22 girls with AIS
Serial spine radiographs analyzed together with bone age (AP left hand) radiographs
Developed skeletal maturity scoring system comprised of 8 stages based on AP left hand
Generated from this a table for risk stratification of AIS curves
**Growth Potential**

- **Quantifying risk of progression**

<table>
<thead>
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<th>Table 1: Logit proportion of the probability of Type I and Type II curve progression to greater than a 90° threshold</th>
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- **Sanders Stage**

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- **Curve Magnitude**

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Growth Potential

- Identifies higher risk for progression

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Future research needed into correlation of Risser and Sanders classifications: we use both.

Treatment

- Treatment outline
  - based on growth potential and curve magnitude

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Bracing

- Various brace types named after location of origin
  - Milwaukee, Boston (most common), Wilmington, Charleston (nighttime brace)
Bracing

Effects of Bracing in Adolescents with Idiopathic Scoliosis
Stuart L. Weinstein, M.D., Lori A. Dolan, Ph.D., James G. Wright, M.D., M.P.H., and Matthew B. Dobbs, M.D.

- Weinstein et al. NEJM 2013
  - Multicenter randomized controlled trial (terminated early)
  - Primary outcome curve progression to ≥50 at skeletal maturity (thelum)
  - 2 treatment groups: 116 randomized to bracing versus observation, 126 allowed to choose treatment (preference cohort)
  - Randomized cohort: 75% rate of bracing success versus 42% success in observation group
  - Both groups: 72% success of bracing versus 48% with observation (1.93)
  - Clear dose response relationship (temperature sensors)
    - 0-6 hours per day: 41% success rate
    - ≥12.9 hours per day: 90-93% success rate

Surgery

- When a curve progresses despite bracing or when a child presents with an operative curve, ≥45° and growth remaining or ≥50° at skeletal maturity, surgery recommended.

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What type of surgery?

- Curve magnitude, skeletal maturity (Sanders), flexibility
Increasingly AIS surgery performed through **posterior approach** with pedicle screws (hooks or wires) and rods.

- Pedicle screw makes fusion constructs significantly stronger than prior hook and wire constructs (3 column fixation).
- Pedicle screws do not involve placing instrumentation into the central canal.
- Ponte osteotomies can achieve effective correction of the AIS curve.
- Strength may prevent development of crankshaft phenomenon.

Axial plane correction

- Pedicle screws facilitate axial correction through various system specific instrumentation.
- Powerful coronal correction can also be achieved utilizing the strength of the pedicle screw and intra-operative reduction devices.

15 y/o male – Pre-op
Managing Expectations

- Posterior Spinal Fusion:
  - T12 or L1: High percentage have a full return to sports
  - L2 or L3: Less likely to have a full return to sports
  - L4: Unlikely to return to sports - flexibility

Fusionless Strategies

- The spine actively grows during the natural history of AIS
- Guided growth techniques have been used reliably in limb deformity
- Could the concept of guided growth be used to gradually correct an AIS curve over time?
Fusionless Strategies

Vertebral Body Stapling
- Nitinol Memory alloy staple
  - Nickel, Titanium, Javel ordnance
  - 50% Nickel, 50% Titanium
  - Chilled: straight
  - Body temperature: curved inward
- Early studies demonstrate equivalency when compared to bracing effectiveness in preventing curve progression (75%)
- Recent studies demonstrate progression despite VBS
  - Issue: cannot adjust once implanted => correction is obtained at the time of surgery.

Vertebral Body Tethering
- Apply pusher to 2nd screw
- Reduce curve with pusher
- Tension at the bottom
- Apply set screw through pusher
- Torque down
- Release tensioner
- Repeat steps (A-E) one screw closer to tensioner
VBT for Growth Modulation:

- **WHO** is the intended treatment group?
  - Defining the clinical problem.
- **What** is the mechanism of action?
  - Understanding the Heuter-Volkman principle.
- **When** should it be done?
  - Predicting models of growth.
- **Where** should it be done?
  - Choose the appropriate levels.
- **How** should it be done?
  - Technical considerations.

Indications for VBT

- Age >9 yrs old
  - Youngest patient is 10.5 yrs old
  - >2 yrs growth remaining
  - Consider up to Sanders 5
- Thoracic AIS curve
  - Consider Lenke 5 curves
- Cobb: 45-65 degrees
  - Largest curve 70 degrees
  - Flexible curves < 30 deg
- Instrumented levels: T5 – L1

- Cobb angle
- Skeletal Maturity
- Curve Flexibility

- Vertebra body angling
- Vertebra body tethering
- Incomplete posterior fusion
- Full posterior fusion
Anterior vertebral body tethering

- Newton et al. JBJS 2008
- Porcine model and mechanical tether across 4 thoracic vertebrae
- Tethering was able to induce deformity in the coronal and sagittal planes
- Crawford and Lenke JBJS 2010
- Case report demonstrating efficacy of vertebral tethering in correction of idiopathic type curves

Most recent 2014 (Spine) and 2015 (European Spine Journal) articles demonstrating the safety and potential efficacy of tethering
**Anesthesia Considerations**

- Double-lumen endotracheal intubation for facilitation of intra-operative exposure and tethering
  - 28 french – 50 kg pt
  - 26 french – 40 kg pt
- Smaller pt – appropriate sized endotracheal tube w/ bronchial blocker

**Patient Positioning**

- Patient positioned on padded towel rolls to optimize curve correction prior to surgical start. Confirmation via fluoroscopy.

**Tethering**

- Head
The Procedure
Segmental takedown

- Ligation of neurovascular bundle at each level for vertebral body tethering

The Procedure
Screw Insertion

- Placement of staple
  - Superior, inferior, posterior tine (rib head)
  - Confirm adequate position with fluoroscopy on PA and lateral

Tethering

- Top loading screws placed under fluoroscopic guidance through thoracoscopic portals
A tether is placed in the screws and tensioned along its length.

A certain degree of curve correction can be obtained on the table.

The tether is secured to its screw with a set screw.

The Procedure

Tensioning the tether

- Feed the cord thru the remaining screws
- Attach tensioner to inferior screw and remove slack

Vertebral Body Tethering

- Apply pusher to 2nd screw
- Reduce curve with pusher
- Tension at the bottom
- Apply set screw through pusher
- Torque down
- Release tensioner
- Repeat steps (A-E) one screw closer to tensioner
Case Report

- 12 year old female
- 47 degree right thoracic curve, 38 degree left lumbar curve

Case Report

- Risser 0
- Sanders 2-3

Case Report

- Traction and bending films
04/27/2017

Case Report

- Post-operatively

18 degree thoracic curve; 20 degree lumbar curve

Tethering

- Benefits
  - Safe procedure
  - Recovery can be more rapid
  - Theoretically preserves motion segments (lumbar spine)

- Risks
  - Narrower indications
  - Flexible curve with sufficient growth remaining
  - Over-correction is possible

- Studies are needed to evaluate the long-term outcomes of tethering
15 year old – Before

15 year old – Tethering

54º thoracic, 46º lumbar
24º thoracic, 13º lumbar

15 year old – After

Before

Tethering

After
MIZZOU Experience

39 VBT patients:
- Thoracoscopic T5 – L1
- Lenke 5 TL curves
- Started in late 2013
- Large learning curve:
  - Surgical technique
  - Decision making: PSF vs VBT
- Advanced our techniques:
  - Avoid taking down the diaphragm, spare segmental vessels, pleural closure endoscopically