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Pediatric & Adolescent Scoliosis

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Children's Hospital Colorado
Here, it's different.™

Objectives

1. Participants will learn about the different types of scoliosis
2. Participants will learn about growth-sparing options to manage scoliosis
3. Participants will learn about spine fusion surgery and post-operative considerations

What is Scoliosis?

- 3 dimensional deformity
- Frontal plane
  - Lateral spinal curve or Cobb angle 10º or more on X ray
- Transverse plane
  - Axial rotation/angle of trunk rotation 5º or more with Scoliometer
- Sagittal plane
  - Altered contour; hypokyphosis
- 2-3% of children meet diagnostic criteria
- Further named for the etiology that caused the spinal deformity

Mo & Cunningham 3

Why is it important to monitor scoliosis?

- Thoracic Insufficiency Syndrome:
- When the size and shape of the thorax is not able to support normal respiration/air exchange

- Efficient respiration:
- Age appropriate thorax volume
- Rib symmetry and movement
- Diaphragm anchored at base of chest

- Respiration in scoliosis:
- 3D spine and rib position asymmetric
- Decreased rib mobility, lung volume, and respiratory muscle function

Srs.org, Mayer et al.
Image: Science photo library 4

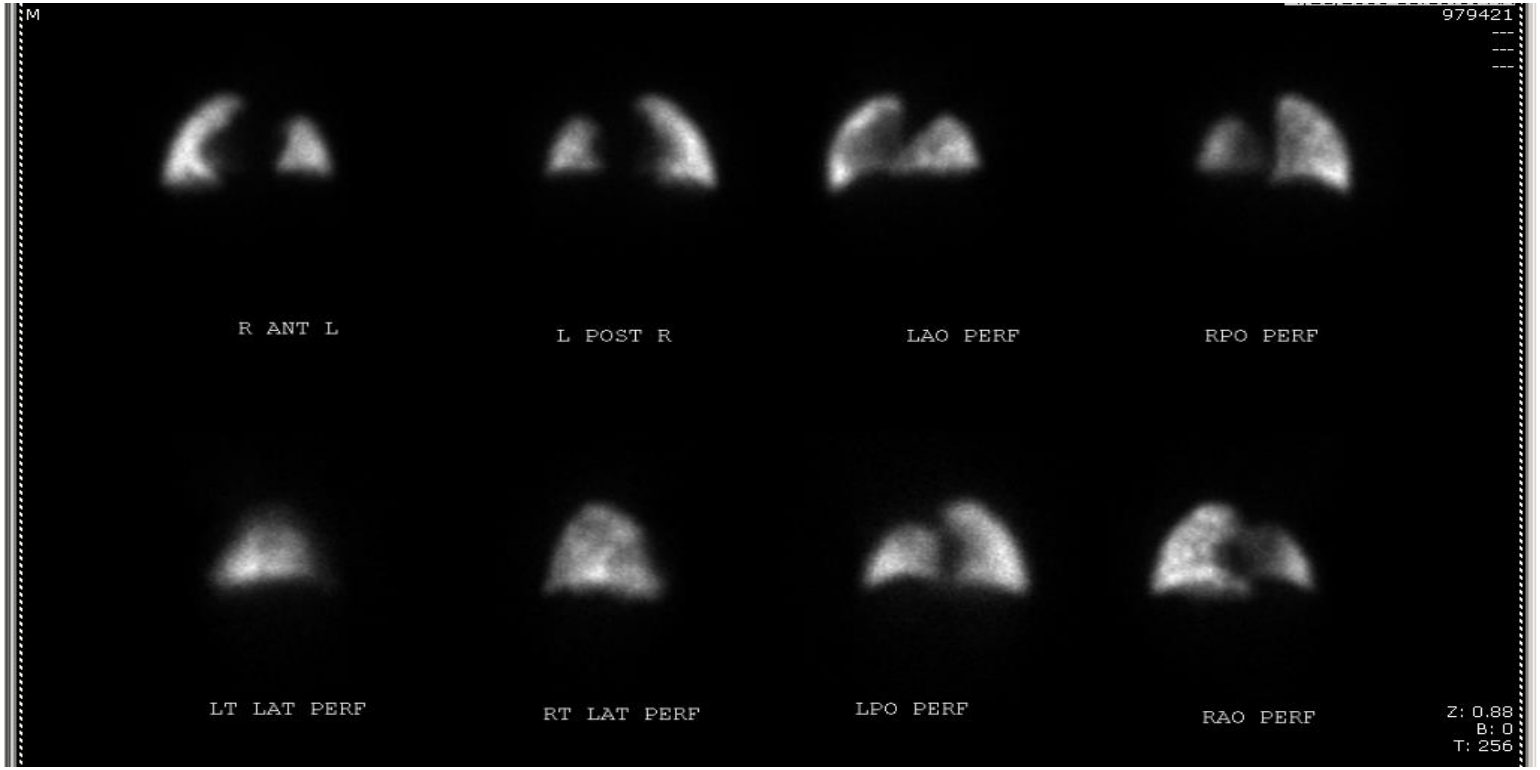
Why is it important to monitor scoliosis?

- Progressive curves can limit lung volume
- Spine growth is most rapid in first 5 years
- Alveolar development greatest in first 8 years
- Need ≥22cm thoracic height

- Consider trunk height *and* lung development
- Relationship between T1-T12 height and FVC

Karst, image from Science Photo Library 5

Thoracic Insufficiency Syndrome



Types of Scoliosis

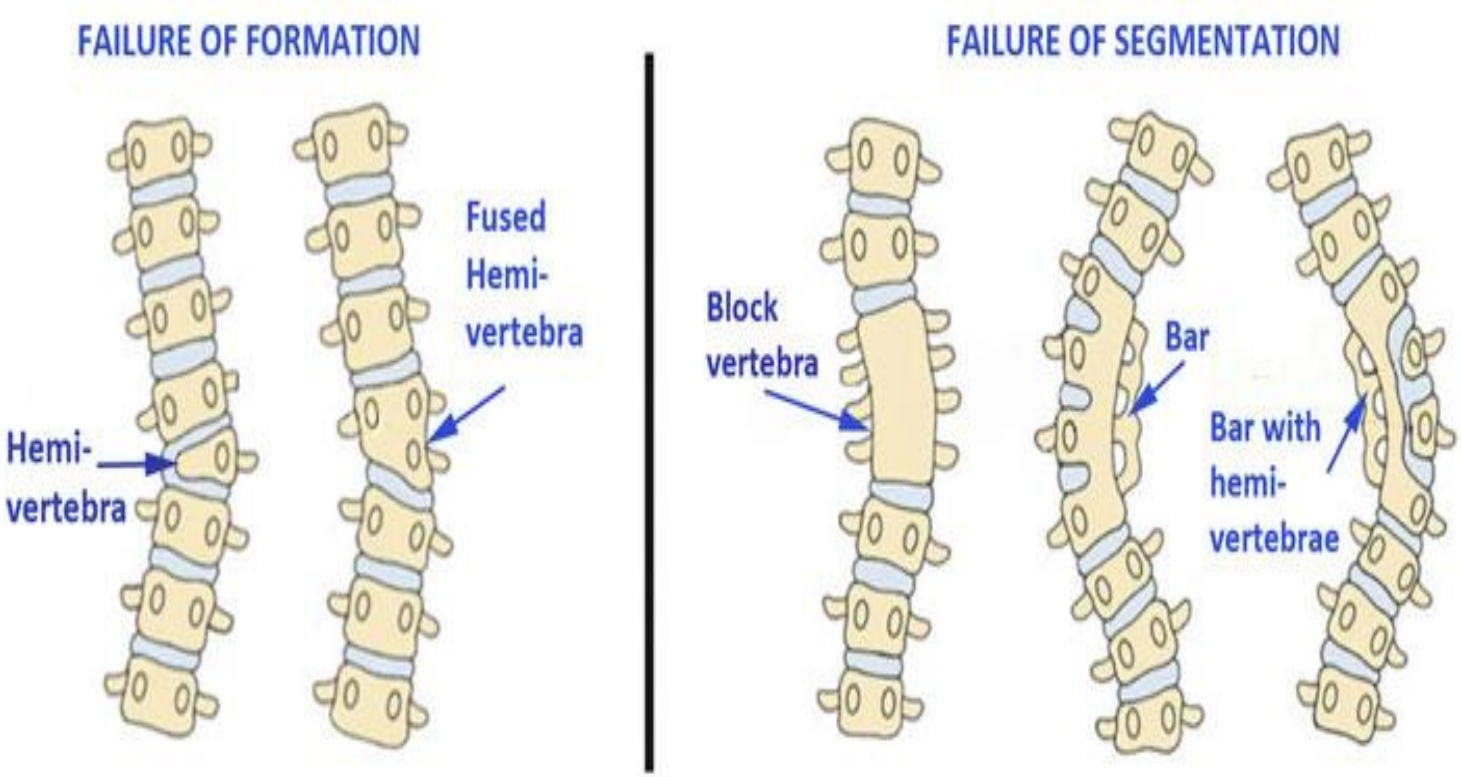
1. Congenital Scoliosis
2. Neuromuscular Scoliosis
3. Early Onset Scoliosis
4. Adolescent Idiopathic Scoliosis

Congenital Scoliosis

Congenital Scoliosis

- Due to bony abnormalities
- Failure of Formation
  - Hemivertebrae
  - Fused hemivertebrae
- Failure of Segmentation
  - Block vertebra
  - Bar
  - Bar with hemivertebrae

Congenital Scoliosis



Goldstein et al. Kumar et al.
Radiopedia.org, srs.org 10

Congenital Scoliosis

- Genetic studies have not identified any inherited risk
- Prognosis
  - risk of curve progression depends on type of congenital deformity
  - mild curve at skeletal maturity= low risk of progression or pain in adulthood

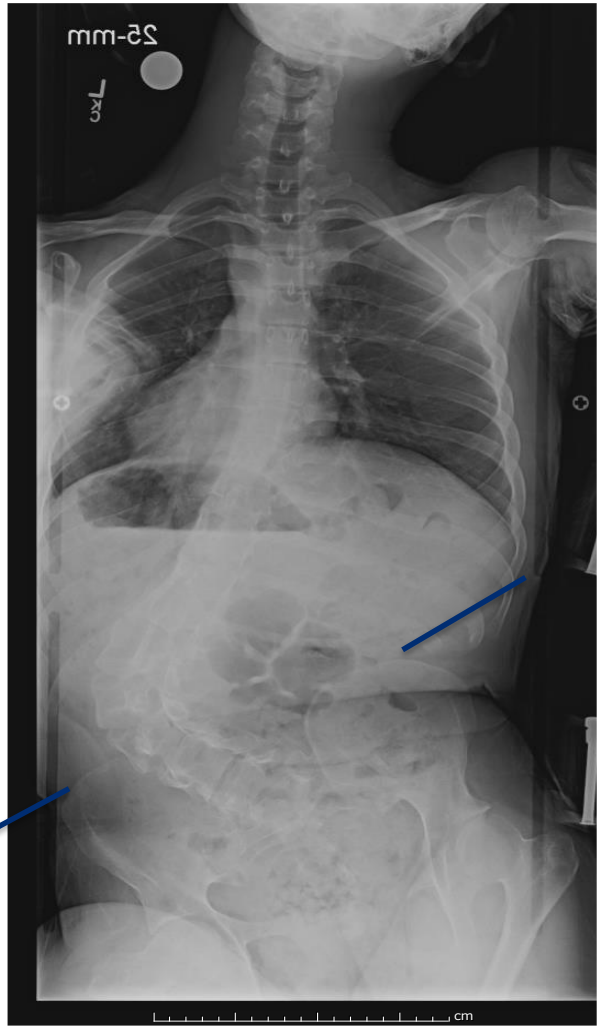
Congenital Scoliosis

- Treatment
  - Serial observation of curve and associated conditions
  - Bracing and/or Casting
  - Physical Therapy
  - Surgery

Neuromuscular Scoliosis

Neuromuscular Scoliosis

- An irregular spinal curvature caused by disorders of the brain, spinal cord, and muscular system srs.org
- Imbalance of trunk/spine muscles, poor muscle control, spasticity Chua et al, Choudhury et al
- Thoracic and lumbar spine plus pelvic obliquity
- More severe and progressive, particularly in patients who are non-ambulatory Allam & Schwabe
- Curves are usually not associated with pain srs.org



Neuromuscular Scoliosis:

Associated Diagnoses and Incidence

- Cerebral palsy
  - 2 limb involvement 25%
  - 4 limb involvement 80%
- Spinal muscle atrophy (SMA) 67%
- Chiari malformation, syrinx
- Spinal cord tumors
- Spinal cord injury <10 years 100%
- Myopathic disorders
  - Duchenne muscular dystrophy 90%
  - Spina Bifida
- Connective tissue disorders
  - Marfan Syndrome
  - Elhers Danlos Syndrome
- Genetic conditions
  - Friedreich ataxia (spinocerebellar degeneration) 80%
  - Noonan
  - Neurofibromatosis
  - Osteochondrodystrophy

Neuromuscular Scoliosis

- **Multidisciplinary team:**
- Pediatrician
- Orthopedics
- Pulmonary
- Neurology
- Gastroenterology
- Nutrition
- Nursing
- DME provider
- Parents/caregivers

- **Treatment:**
- Observation
- Bracing ?
- Wheelchair modifications
- Physical therapy
- Surgery

Srs.org, Choudhury et al, Halawi et al,
photo Elise Benefield CHCO 16

Early Onset Scoliosis

Early Onset Scoliosis

- Cobb angle 10º or more (frontal plane)
- Angle of trunk rotation 5º or more with Scoliometer (transverse plane)
- Hypokyphosis thoracic spine (sagittal plane)
- Adams Forward Bend Test
- Diagnosed before age 10, *and* not due to congenital or neuromuscular etiologies
- Infantile (0-3 yo)
  +Juvenile (4-10 yo)
  =Early Onset Scoliosis

Early Onset Scoliosis (Infantile)

- Infantile Scoliosis (0-3y)
- 1% of all idiopathic scoliosis in children
- Boys 60%; Girls 40%
- Intra uterine molding vs post delivery theories
- 90% resolve without treatment
- X ray to assess risk of progression
  - Cobb angle (curve)
  - Mehta Angle (rib vertebral angle difference)
  - Angle of trunk rotation

Early Onset Scoliosis (Juvenile)

- Juvenile Scoliosis (4-10y)
- 10-15% of patients with idiopathic scoliosis

Early Onset Scoliosis

- Goals:
- minimize curve progression
- maximize lung function and motion of chest and spine
- consider overall development

- Treatment:
- Observation X rays every 4-6 months
- Cast or Brace
- Distraction based (Halo)
- Magnetic growing rods
- Definitive fusion

srs.org. Image: Elise Benefield 21

Adolescent Idiopathic Scoliosis

Adolescent Idiopathic Scoliosis

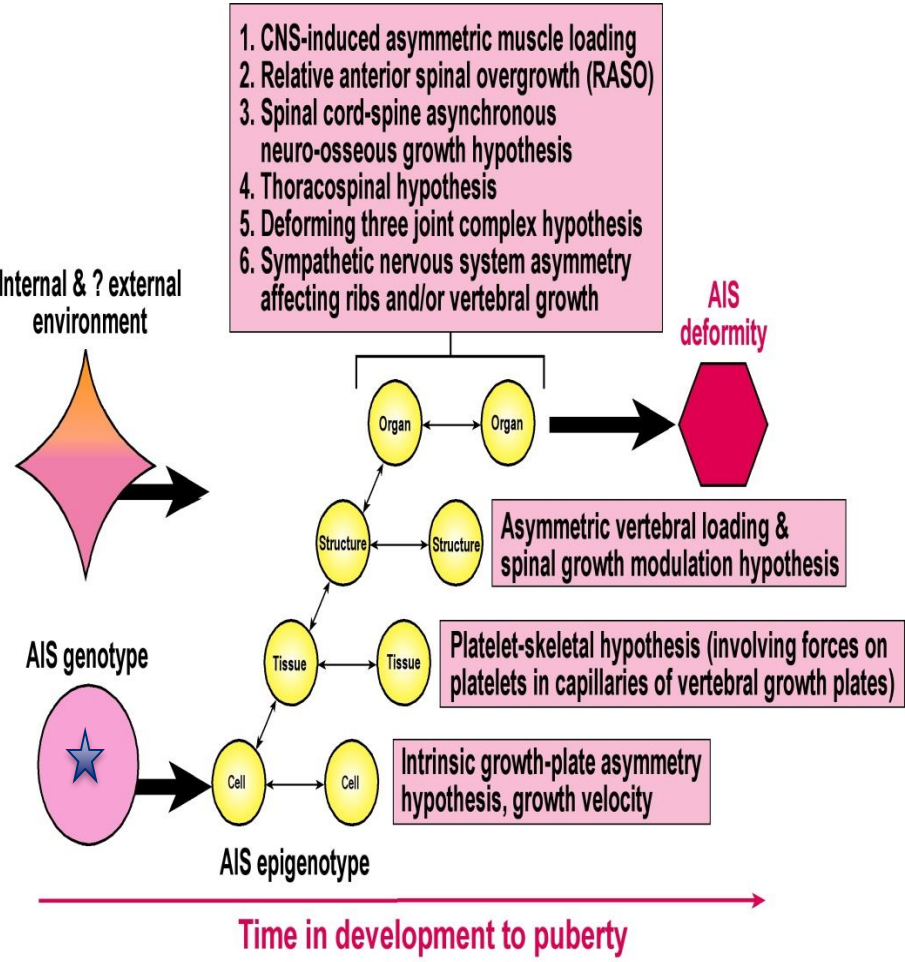
- Cobb angle 10º or more (frontal plane)
- Scoliometer 5º or more angle of trunk rotation (transverse plane)
- Hypokyphosis thoracic spine (sagittal plane)
- Adams Forward Bend Test
  - Left to right asymmetry
  - Spinous processes not in line
  - Loss of normal arc of flexion
- Identified during pre/pubertal growth spurt, age 10-18

Adolescent Idiopathic Scoliosis

- Idiopathic = No known cause
- Diagnosis of exclusion, first rule out other etiologies
- Intermittent back pain?
- 1.5% of all teens
- Tall, slim, active, **girls** & boys
- Curves 10-30º 1.4:1
- Curves over 30º 10:1
- Most patients only need periodic X rays and orthopedic follow ups
- Attention to curve type

AIS is Multifactorial

- **Genetic basis**, influenced by
  - Hormones & metabolic dysfunction
  - Spine growth & development
  - Biomechanical factors
    - Geometric & mechanical torsion
    - Mechanics of upright spine
    - Left to right asymmetry
  - Aerobic exercise capacity limitations & generalized muscle dysfunction
  - Vestibular system?



Genetics

- 53 genetic markers are correlated with scoliosis Mo & Cunningham
- In women with curves >15°, the incidence of scoliosis in their daughters is 27% Harrington
- Identical twins 73% Carr
- Chromosomes 6, 9, 16, 17, 18 and the secondary regions in 1, 3, 5, 7, 8, 11, 12, 19 Miller, Gurnett

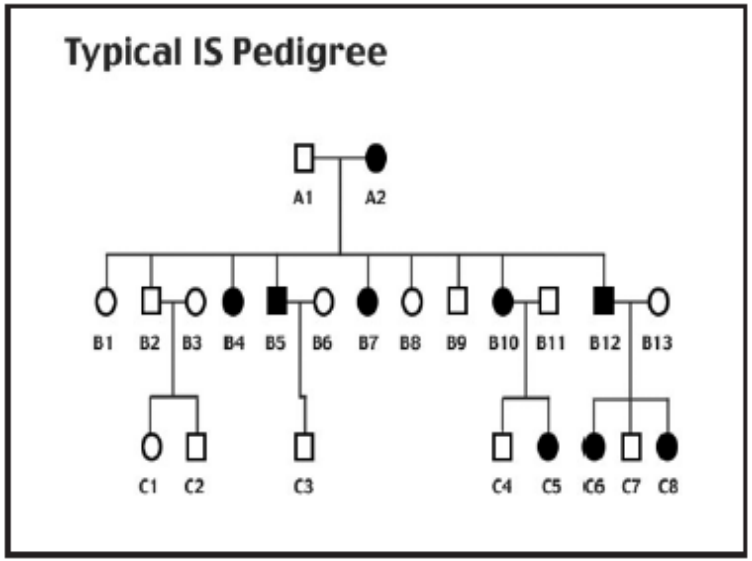
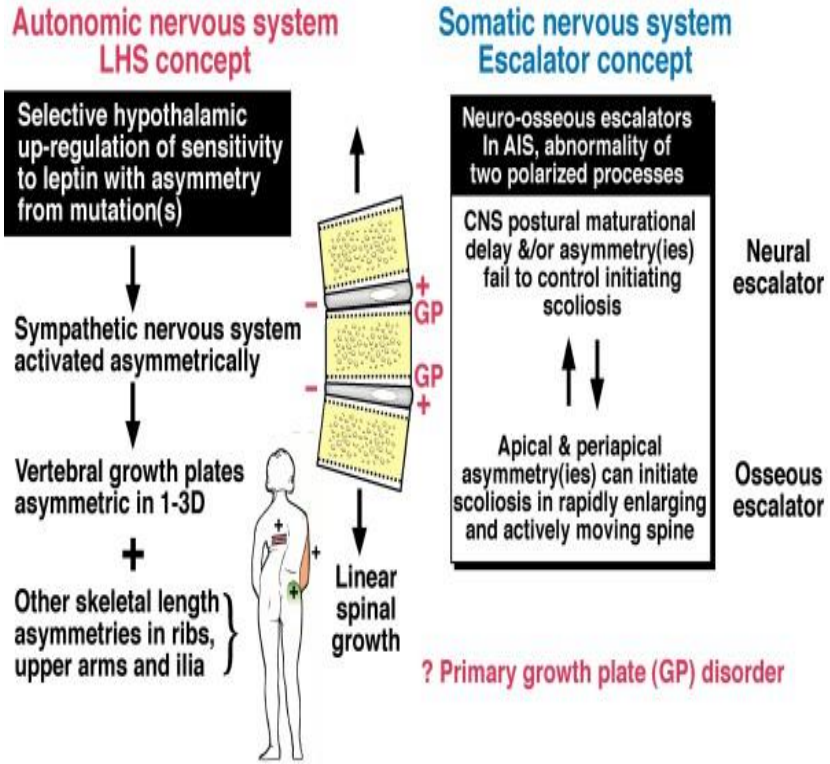


Figure 3. The genetic map of a family with extensive scoliosis. People with scoliosis are shaded. Females are represented by circles, males by squares.

Mo & Cunningham, Harrington, Carr, Miller et al, Gurnett et al, Wang et al. Image from scoliosis.org.

Hormones and metabolic factors

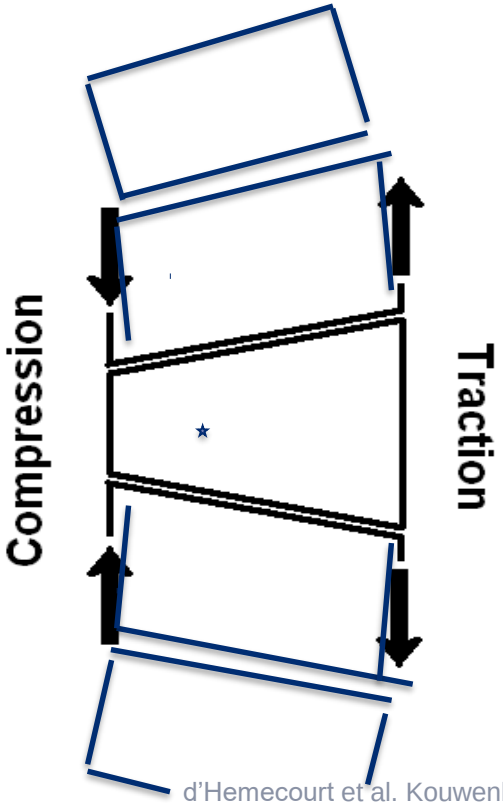
- Leptin-Hypothalamic-Sympathetic nervous system, LHS concept
  Burwell et al 2008
- Somatic nervous system Escalator concept
- Hypoestrogen
- Decreased bone density and muscle mass
- Low nocturnal melatonin, platelets
- Inflammation Kouwenhoven & Castelein. Martinex-Liorens et al



Burwell et al, Kouwenhoven & Castelein. Martniex-Liorens et al

Spine Growth and Development

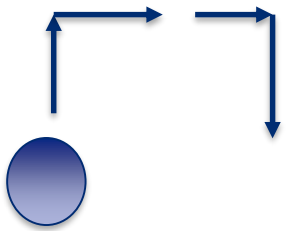
- Asymmetric closure of lateral ossification center on concave side
- Hueter Volkmann Law
  - Compressive forces at epiphyseal plates reduce growth on concave side
  - Distractive forces accelerate growth on convex side
- Relative anterior spinal overgrowth (RASO)= hypokyphosis of thoracic spine



d'Hemecourt et al. Kouwenhoven & Castelein. Burwell et al.
Image adapted from ScientificSpine.com

Geometric and Mechanical Torsion

- Apical vertebra moves ventral lateral and lateral dorsal
- Mechanics of flexion change, resulting in torsion

- anterior/ventral
- Left  Right
- dorsal/posterior

- Twist a towel, then flex

Intervertebral Disc

- Nucleus pulposis shifts to convex side of curve; greatest wedging at apex

Upright spine mechanics

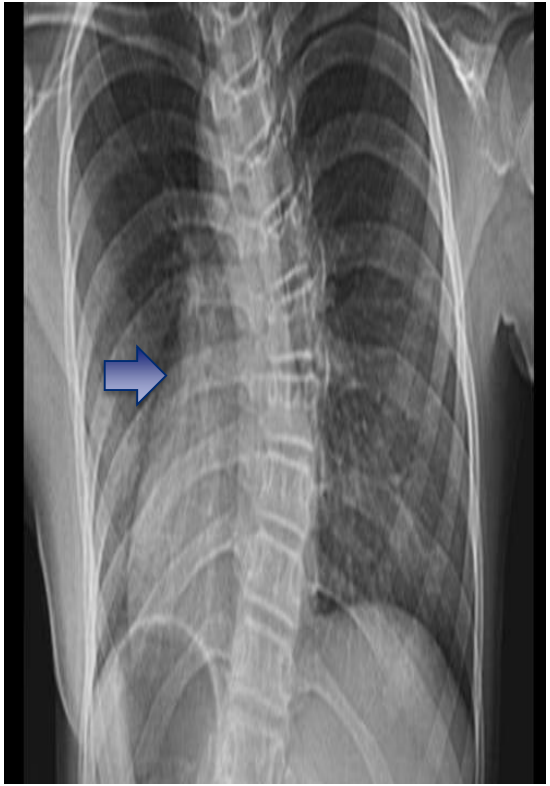
- Axial loads and Shear loads; Gravity, muscle pull

Left to Right Asymmetry

- Location of thoracic aorta and vertebral column

Paravertebral Muscles

- Asymmetry of paraspinal muscle strength; multifidi predominantly type 1 (slow twitch) fibers in convexity of curve



Kouwenhoven & Castelein, Wang et al

Aerobic Exercise Capacity Limitations & Generalized Muscle Dysfunction

- **Functional aerobic exercise capacity limitations**
- Noted non athletes
- No direct link between severity of spinal deformity and lung function

- **Generalized muscle dysfunction**
- Contributes to reduced aerobic capacity when Cobb angle >40º
  - spirometry for lung function
  - max inspiratory & expiratory pressures (MIP, MEP)
- Limb muscle strength
  - bilateral hand grip
  - isometric quads (QMVC)

$$\text{Progression Factor} = \frac{\text{Cobb Angle} - (3 \times \text{Risser sign})}{\text{Chronological age}}$$

Risk of curve progression in AIS

- Pubertal growth spurt is time of greatest risk of curve progression

- Prognosis for curve progression after skeletal maturity:
- 0-30 degrees: minimal risk
- 30-50 degrees: gray area
- 50 degrees or greater: very likely to progress, up to 1 degree/year

Management of Scoliosis

Management of Scoliosis

- Growth Sparing
- Casting
- Bracing
- Physical Therapy
- Halo Traction
- Magnetic Growing Rods
- Vertebral Body Tethering (VBT)

- Goal: avoid or delay spine fusion (trunk height, lung development)

- Success depends on the etiology of the deformity and patient compliance

- Growth Arresting
- Definitive Spine Fusion

- Goal: stabilization of severe or progressive deformities

Growth Sparing Management of Scoliosis

Observation

- Observation at regular intervals
- All patients who have scoliosis
- Frequency depends on
  - Type of scoliosis
  - Risk of progression
    - Degree of curve
    - Skeletal maturity

Mo & Cunningham36

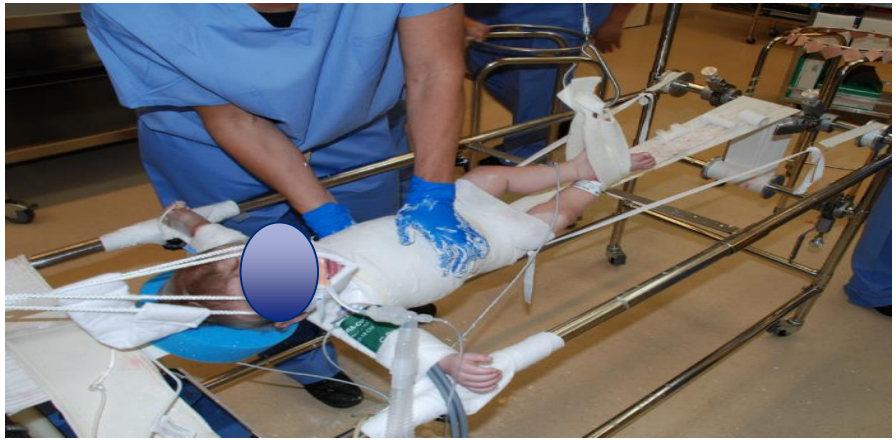
Casting

Casting

- Congenital
- Early Onset

photos from Elise Benefield RN

Casting



Photos from Elise Benefield RN 39

Casting

Photos Elise Benefield RN 40

Casting


outside film labeled
incorrectly, this is left
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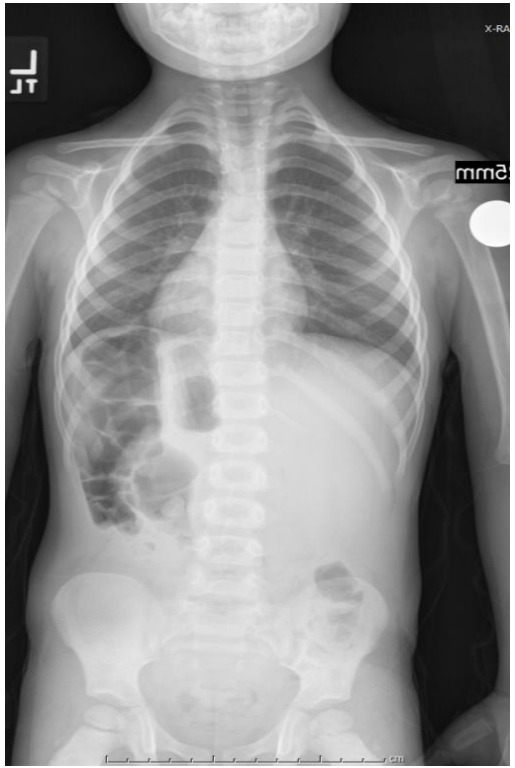


Bracing

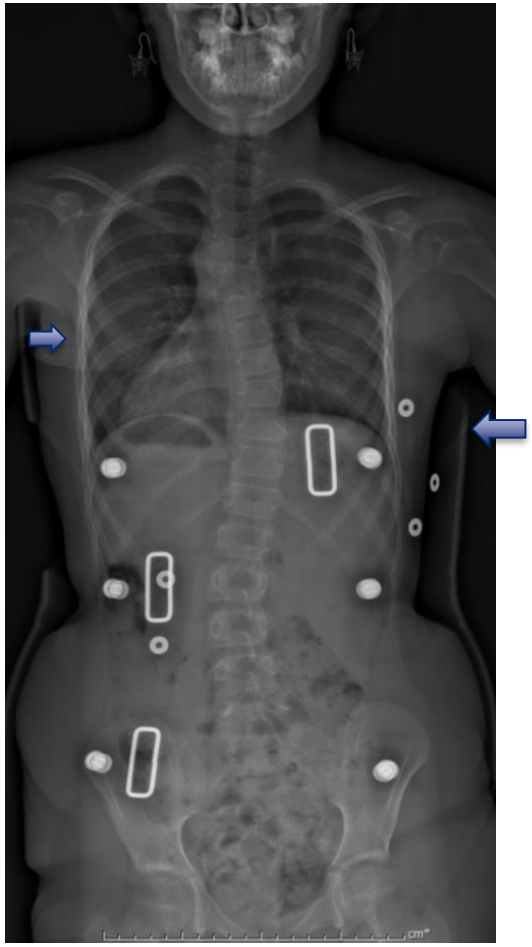
Bracing

- Curves 20-40 degrees in kids who are still growing
- Adolescent Idiopathic
- Early Onset
- Congenital
- Neuromuscular (rarely)

- Bracing aims to halt or minimize curve progression
- MUST be worn as recommended

Image of Boston Brace 43

Bracing

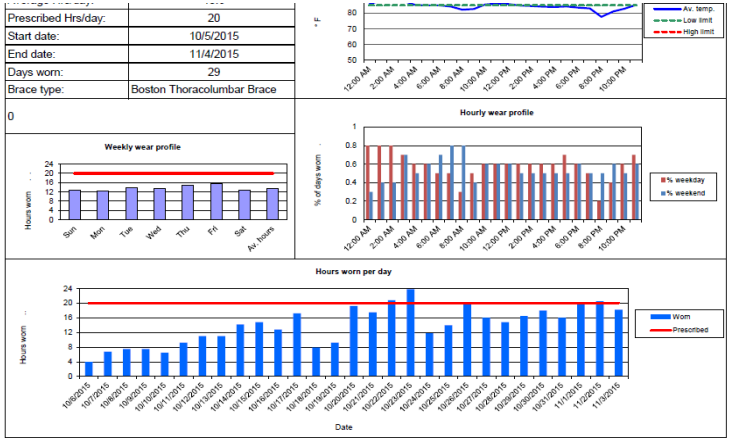


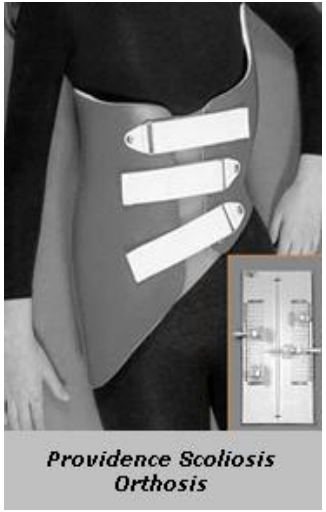
Images from Elise Benefield44

Bracing:

Adolescent Idiopathic Scoliosis

- Full Time Rigid Brace
  - 18-22 hours per day
  - Boston/Boston 3D
  - Rigo-Cheneau
  - Wood Cheneau Rigo
  - Scoli Brace
- Night Time Rigid Brace
  - Providence
  - Charleston Bending Brace
- Soft Brace
  - Spine Cor



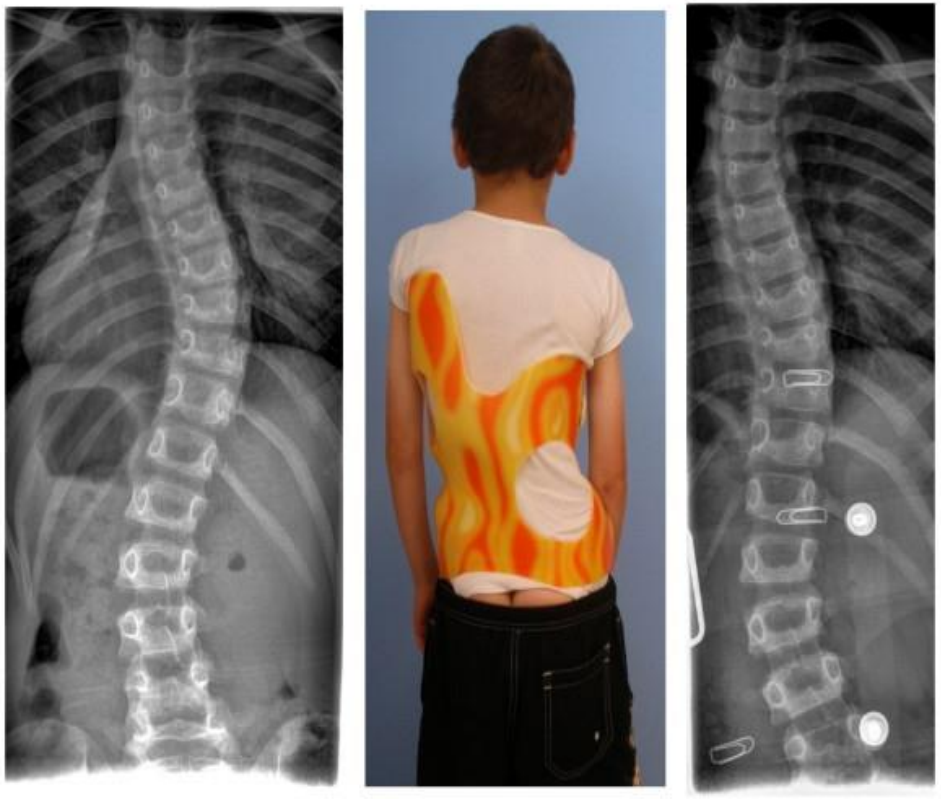


Yrjonen et al. Providence 45

Bracing:

Early Onset Scoliosis

- Often used after casting
- Aim to halt or minimize curve progression
- Aim to delay surgery until patient is older and taller



Srs.org, image from ResearchGate.net 46

Bracing:

Congenital Scoliosis

- Rarely effective for primary curve or rigid curve due to bony abnormality
- May help a compensatory curve or flexible curve
- Aim to slow down the curve progression
- Aim to delay surgery until patient is older and taller

srs.org, image from Pinterest 47

Bracing:

Neuromuscular Scoliosis

- Small curves may be braced, but most curves caused by neuromuscular conditions do **not** benefit from bracing Chua et al 2017
- Wheelchair modifications to improve sitting function for patients who do not ambulate Chua et al 2017
- Bracing may help improve function (sitting posture) Halawi et al 2015 but can have negative effect on respiratory function Mullender et al
- Boston Brace and sitting using pressure-mapping system during 10 day wearing time Bloomkvist et al 2018
  - Sitting function 73/105, 5 deteriorated
  - Symmetry 44/86, 3 deteriorated
  - Stability 20/40, 7 deteriorated

Physical Therapy

Physical Therapy:

Congenital Scoliosis

- Range of Motion: cervical spine, trunk, hips, knees, ankles, UEs, LEs
- Strength: trunk, core, shoulder girdle, pelvis, LEs
- Breathing
- Gross motor skills: midline, symmetry of motor skills, equal weight bearing, transitions over each side, bimanual skills, symmetric
- Recommendations based on clinical practice

youtube photo: Balanced By Beth

Physical Therapy:

Neuromuscular Scoliosis

- PT does not directly impact neuromuscular scoliosis
- Therefore, PT to address
  - Range of motion
  - Stretching
  - Submaximal (moderate) resistance/ weight lifting
  - Aerobic exercise
  - Ambulation
  - Functional use of arms and legs
  - Posture muscles
  - Respiratory exercises/ breathing muscles

Physical Therapy:

Early Onset Scoliosis

- Manipulation, PT, and exercise have **not** been shown to influence spinal deformity srs.org
- Goals in conservative management of EOS:
  - minimize curve progression
  - **maximize lung function and motion of chest and spine***
  - **consider overall development***

- ***Physical therapy fits here**
- Range of motion/flexibility
  - Spine
  - Rib cage and breathing
  - Upper and lower extremities
- Strength
  - Trunk/core
  - Shoulder girdle
  - Pelvis and lower extremities
- Symmetry of gross motor skills
  - Weight bearing
  - Transitions
  - Bimanual and midline skills

- Recommendations based on clinical practice

Physical Therapy:

Adolescent Idiopathic Scoliosis

- Bracing plus Scoliosis Specific Exercises:
- “Together can reduce progression of scoliosis curves and angle of trunk rotation, while improving aesthetics and health related quality of life”
- Helps avoid loss of correction after brace wearing completed

SOSORT 2016

0-20 | 20-40 | above 40

		Low		Moderate		Severe	
		Min	*Max*	*Min*	*Max*	*Min*	*Max*
Infantile		Obs3	Obs3	Obs3	TTRB	TTRB	Su
Juvenile		Obs3	PSSE	PSSE	FTRB	HTRB	Su
Adolescent	Risser 0	Obs6	SSB	HTRB	FTRB	TTRB	Su
	Risser 1	Obs6	SSB	PSSE	FTRB	FTRB	Su
	Risser 2	Obs6	SSB	PSSE	FTRB	FTRB	Su
	Risser 3	Obs6	SSB	PSSE	FTRB	FTRB	Su
	Risser 4	Obs12	SIR	PSSE	FTRB	FTRB	Su
Adult up to 25 y		Nothing	PSSE	Obs12	SIR	Obs6	Su
Adult	No Pain	Nothing	PSSE	PSSE	SIR	Obs12	HTRB
	Pain	PSSE	SSB	PSSE	HTRB	PSSE	Su

Halo Traction

Halo Traction Indications

- Severe deformity of spine and chest wall
- Decreased pulmonary function due to size of chest cavity
- Large, stiff curves
- Used before surgery or before or between bracing or casts
  - Congenital
  - Early Onset
  - Adolescent Idiopathic

Halo Traction

- Distraction based, growth-friendly ‘surgery’
- Halo Ring with 4 to 12 pins
- Pulley system attached to walker or wheelchair
- Amount of weight and traction depends on child’s weight
- 4-8 weeks, 8-12 hrs/day when awake
- Standing or walking 4 hrs/day
- Completed with support of family at home

Halo Traction Goals

- Gentle pull on muscles and joints
- Decrease stiffness of curve, helps with surgical outcomes
- Decrease pressure on heart and lungs
- Decrease energy expenditure
- Breathe easier
- Eat more
- Gain weight
- Decreases back pain

Texas Scottish Rite Hospital for Children.
Photo courtesy of Elise Benefield, CHCO 58

Magnetic Growing Rods

Magnetic Growing Rods

- Surgical rod placement single or **dual**
- Adjusted with external device in office
- Fewer surgeries
- Cost savings
- Improvement in Cobb angle and pulmonary function

- Indications
  - After age 3 or 4 due to rod size
  - Primarily Thoracic curves
  - Early Onset, Neuromuscular or Congenital scoliosis

Johar and Nemade, Lonner
Image: Ridderbusch et al, 60

Magnetic Growing Rods

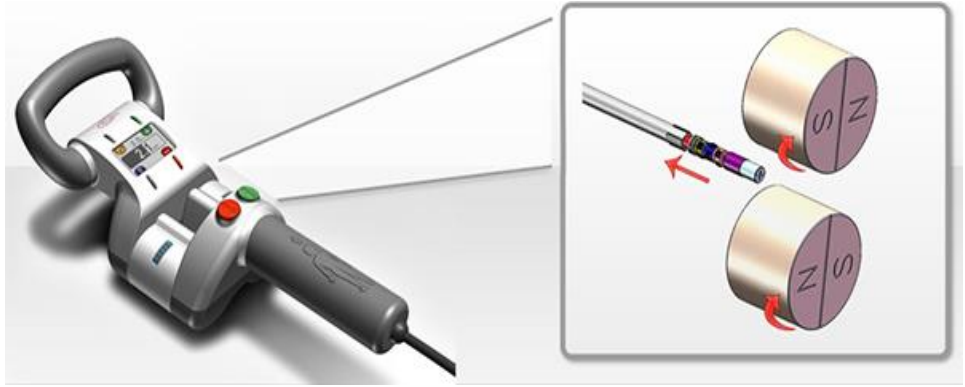
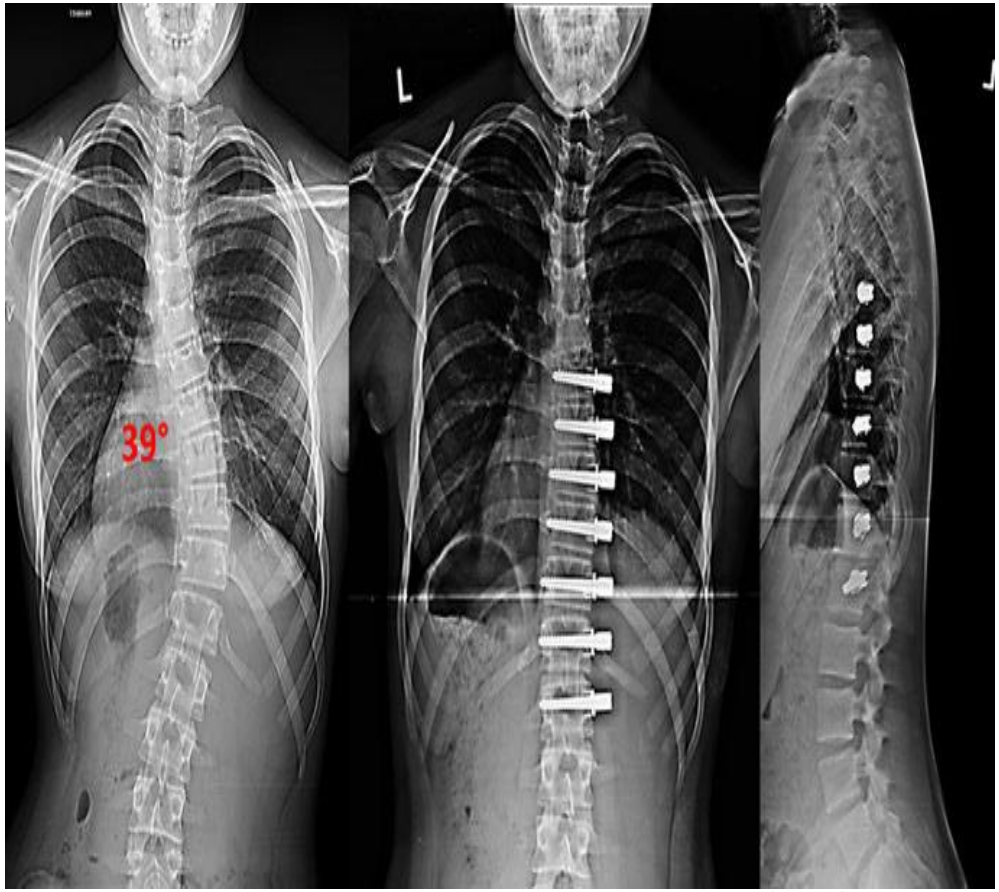


Image 1: medgadget.com, Image 2: Vanderbilt University Medical Center 61

Vertebral Body Tethering

Vertebral Body Tethering (VBT)

- Screws in affected vertebrae on convex side
- Flexible tether on convex side
- Gradual curve correction as patient grows
- Preserves flexibility
- Candidates have specific curve magnitudes and open growth plates



Srs.org, Lonner SpineUniverse.com,
Images from Mario Morgado Mt. Sinai NY 63

VBT

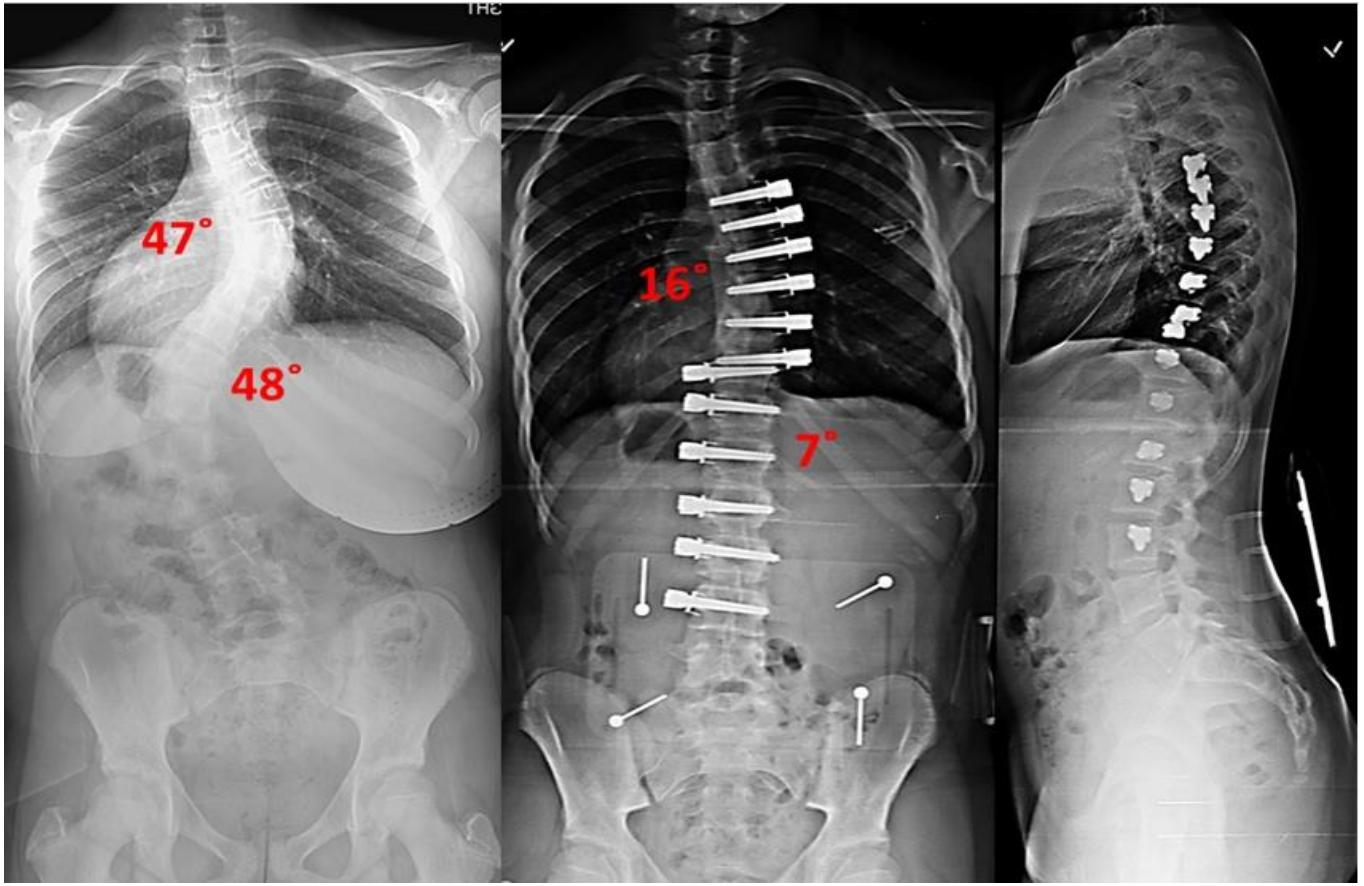


Photo source: SpineUniverse.com Baron S Lonner MD

Growth Arresting: Spine Fusion

Spine Fusion Goals

- Prevent curve progression or decrease magnitude of curve
- Improve sitting balance and tolerance in patients who do not ambulate
- Reduce any pain
- Improve lung function
- Improve aesthetics of back

Srs.org, images Children's Hospital Colorado

Spine Fusion

AIS

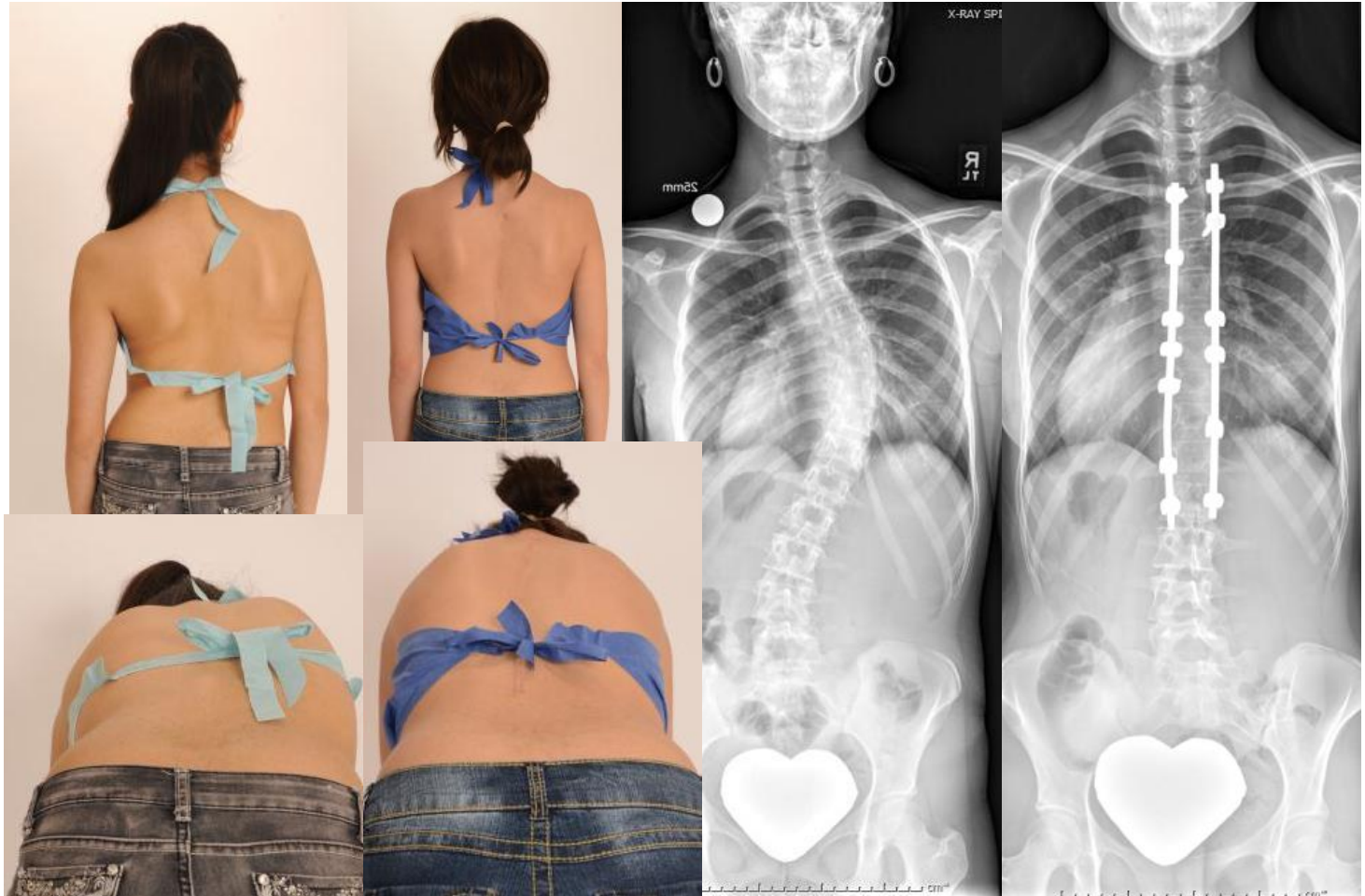
- Pre op class for patient and parent
- X rays, including side bending x rays to determine flexibility of curve
- Spare lumbar when possible

Image from Srs.org 67

Spine Fusion:

AIS

- 2-3 days in hospital
- Activity
  - Log roll, sit edge of bed, up to chair, walk to bathroom, walk hallway, stairs
- Breathe- incentive spirometer
- Communicate needs
- Return to school 2-3 weeks
- Light backpack recommended
- No precautions once incision healed
- Return to sports/activities as tolerated


X-RAY SPI
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Spine Fusion:

Neuromuscular Scoliosis

‘High Risk’ Care Pathway for Spine Surgery

- Multidisciplinary team evaluates patient medically
- Determine risks
- Assess family’s resources and ability to care for patient once discharged
- Patient, parent/caregiver, and surgeon **all** come to a consensus on whether surgical management is the best option

- Sleep Study
- Pulmonary
- Nutrition
- Cardiology if indicated
- High Risk Clinic: RN, PT, Social Work, Nutrition, Respiratory
- Discharge Planning meeting
- Ethics if requested
- Other consults as needed

Spine Fusion:

Neuromuscular Scoliosis

High Risk Pathway Considerations:

- Age, ambulation status, underlying diagnosis
- Consider risk factors or possible medical complications
- Quality of life
  - Degree or progression of spinal deformity
  - Ability to sit in wheelchair
  - Pain
  - Increased difficulty with daily cares

Goals of fusion surgery:

- Prevent curve progression
- Improve lung function
- Increase sitting balance/ tolerance if non ambulatory
- Decrease pain

Srs.org, CHCO Spine Team, Photos: Elise Benefield71

17 year old male with CP, GMFCS level 5

Photos: Elise Benefield 72

Post Operative Care Path

- Ventilator up to 24 hours, sedated, pain management
- Then BiPap for additional support
- Fluids & Nutrition
- Labs
- Incision/dressing
- Neurovascular checks
- Pain management after extubated
- Activity
  - log roll every 2 hours with nursing
  - head of bed to 90 degrees
  - dangle edge of bed after extubated
  - up to chair within 18 hours of extubation
  - PT train family on mobility or transfers if non ambulatory

Spine Fusion-High Risk Pathway

References

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Thank You!

Questions?