## When to Operate on a Child with Spinal Deformity

# Understanding the Differences Between Deformities

#### S. Matt Hollenbeck 2/22/19



## Deformity

- a condition in which part of the body does not have the normal or expected shape
- Synonym
  - Flaw
  - Blemish
  - Distortion
  - Disfigurement
  - Defect



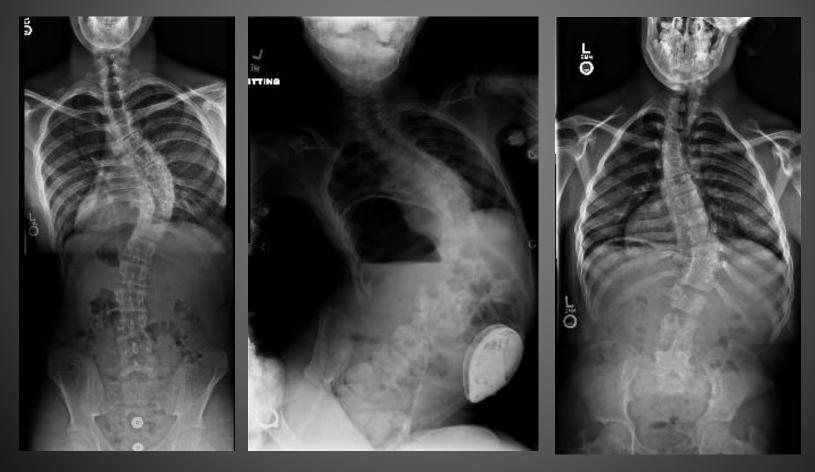
## Spinal Deformity

- Scoliosis
  - Idiopathic
    - Adolescent
    - Juvenile
    - Infantile
    - Early Onset
  - Neuromuscular
  - Congenital
  - Syndromic

- Kyphosis
  - Scheuermann's
  - Post Traumatic
  - latrogenic
  - Post Surgical
  - Congenital
  - Neuromuscular

#### Are these curves not the same?

• 15 yo M 11 yo M w/CP 14 yo M



- Adolescent Idiopathic
  - Spinal Instrumentation & Fusion around 50° in the thoracic spine
  - Consider 45° in the lumbar spine
  - Possibly as low as 40° in the lumbar spine with a large trunk shift (clinical deformity)
  - If no intervention then progression of the deformity into adulthood (1° per year) (Weinstein)
  - Can lead to Pain, worsening deformity, & back, lung, and heart problems

- Adolescent Idiopathic Scoliosis
  - Cor Pulmonale & R Heart Failure (>90-100° curve)
  - Pulmonary function is limited in severe scoliosis (>70°)
    - Chest wall deformity can cause restrictive lung disease
    - Thoracic Lordosis decreases lung volumes
  - Higher rate of back pain in adults with AIS vs.
     controls (65% vs. 35%) at 50 year f/u (Weinstein)
  - Psychosocial Impact
    - Lower Marriage rates, Self Image issues, Parental unacceptance

#### AIS

#### • 19 yo F



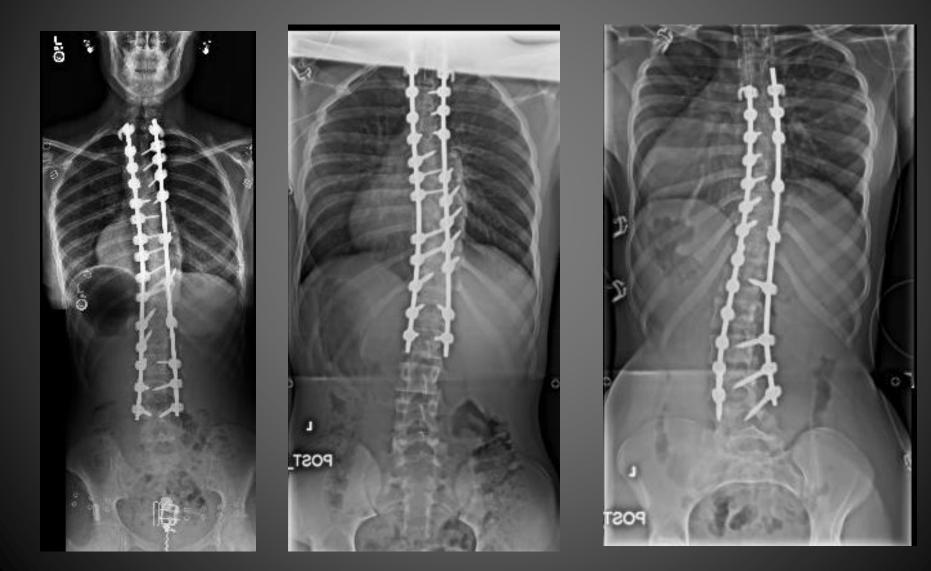
#### 15 yo male



#### 15 yo F



### AIS



#### Adult Sequela of AIS

#### 32 yo Female





#### Adult Sequelae of AIS

• 40 yo Female



#### Adult Sequelae of AIS

40 yo F
 – Post op

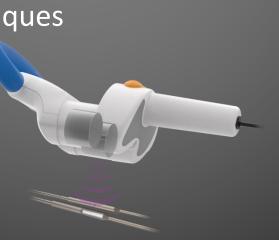


### **Scoliosis Surgical Intervention**

- Juvenile/Infantile Idiopathic
  - Indications much more variable
  - Goals:
    - Control the spinal deformity
    - Allow for continued growth
    - Delay spinal fusion
  - Interventions variable
    - Based on curve size
    - Growth remaining
    - Progression of the curve

### **Scoliosis Surgical Intervention**

- Juvenile/Infantile Idiopathic
  - Intervention Options
    - Bracing
    - Risser Casting
    - Growing Rods
    - Magnetic Rods
    - Schilla Rod Techniques
    - VCR





- Juvenile/Infantile Idiopathic
  - Progressive
  - Conservative intervention failure (brace/Risser)
  - Moderate to Large Size Curve
    - Growth preserving



#### • Juvenile/Infantile Idiopathic 1+10 yo 3+2 yo





#### 3+7 yo



• Juvenile/Infantile Idiopathic





• Juvenile/Infantile Idiopathic

5+0 yo



5+6 yo



- Adult Sequela of Juvenile Idiopathic Scoliosis
  - 49 yo Female
  - Diagnosed at 6 yo
  - Brace until Mature
  - Curve was 50°/50°



### **Scoliosis Surgical Intervention**

- Neuromuscular
  - Goals
    - Sitting or Standing Balance (Coronal and Sagittal)
    - Encourage Independence
    - Assist mobility (gait & transfers)
    - Prevent Progression
    - Maximize Cardiopulmonary Function
    - Improve Hygiene
    - Improve patient/caregiver quality of life

- Neuromuscular
  - Older literature states similar indications as AIS
    - >50° Curve
  - No new literature regarding this subject
    - But risk of progression is high due to NM condition
  - Proactive vs. Reactive Approach
    - Risk of Progression (>50°)
    - After patient/caregiver have significant challenges

#### The Pros and Cons of Operating Early Versus Late in the Progression of Cerebral Palsy Scoliosis

Steven M. Hollenbeck, Burt Yaszay, Paul David Sponseller, Carrie E. Bartley, Suken A. Shah, Jahangir Asghar, Mark F. Abel, Firoz Miyanji, Peter O. Newton

- Neuromuscular Proactive vs. Reactive
  - Being proactive (< 70°) has no advantages
    - No decrease in risks
    - No improvement in quality measures
  - Being Reactive (> 90°)
    - Increased risk of infection
    - Increased blood loss
    - Increased need for anterior/posterior surgery
    - Increased operative time
    - Trend towards lower quality of life

Ideally surgery recommended on curves < 90°</li>

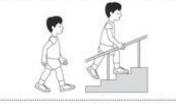
• Neuromuscular

GMFCS
 Classification
 System

#### GMFCS for children aged 6-12 years: Descriptors and illustrations

#### GMFCS Level I





#### including running and jumping, but speed, balance and coordination are impaired.

#### GMFCS Level II

Children walk indoors and outdoors and climb stairs holding onto a railing but experience limitations walking on uneven surfaces and inclines and walking in crowds or confined spaces and with long distances.

Children walk indoors and outdoors and climb stairs without limitation. Children perform gross motor skills

#### **GMFCS** Level III

Children walk indoors or outdoors on a level surface with an assistive mobility device and may climb stairs holding onto a railing. Children may use wheelchair mobility when traveling for long distances or outdoors on uneven terrain.

#### GMFCS Level IV

Children use methods of mobility that usually require adult assistance. They may continue to walk for short distances with physical assistance at home but rely more on wheeled mobility (pushed by an adult or operate a powered chair) outdoors, at school and in the community.

#### GMFCS Level V

Physical impairment restricts voluntary control of movement and the ability to maintain antigravity head and trunk postures. All areas of motor function are limited. Children have no means of independent mobility and are transported by an adult.

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Neuromuscular - 15 yo M GMFCS Level 4 CP

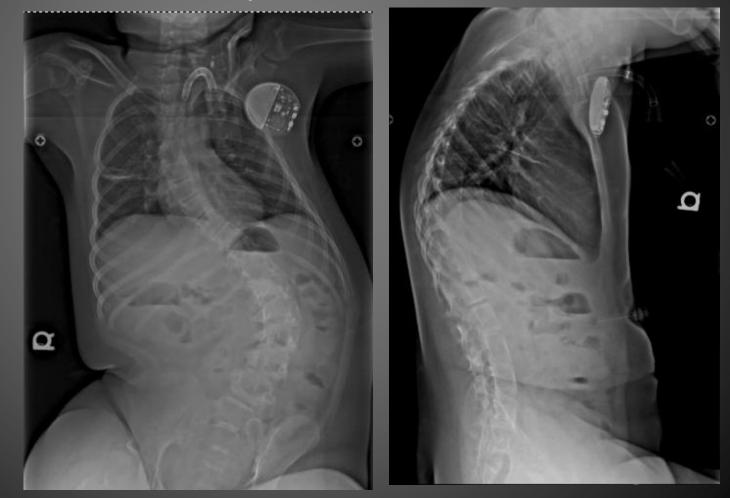




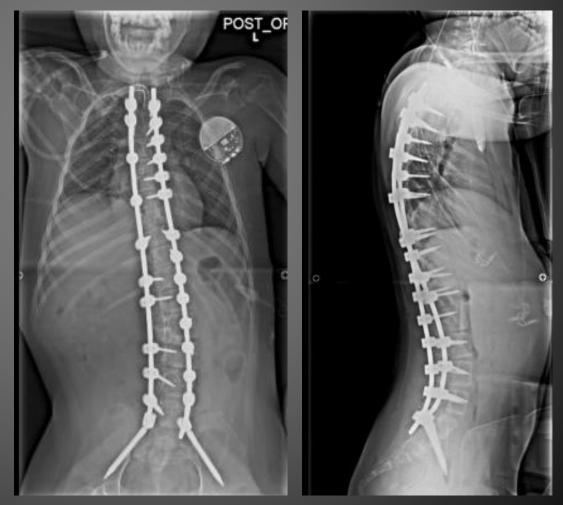




Neuromuscular – 13 yo M GMFCS Level 5 CP



Neuromuscular – 13 yo M GMFCS Level 5

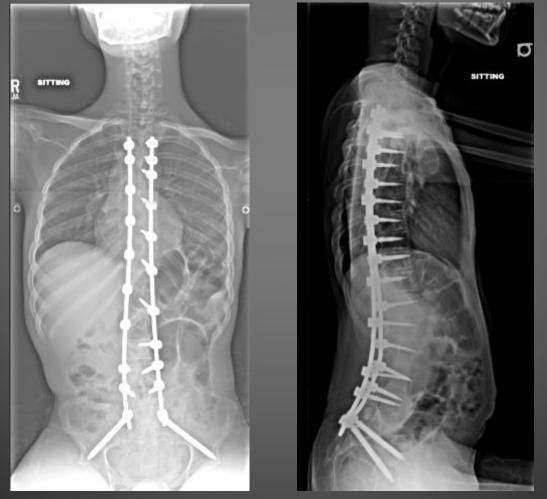


Neuromuscular – 14 yo M w/ Duchenne MD





Neuromuscular – 14 yo M w/ Duchenne MD

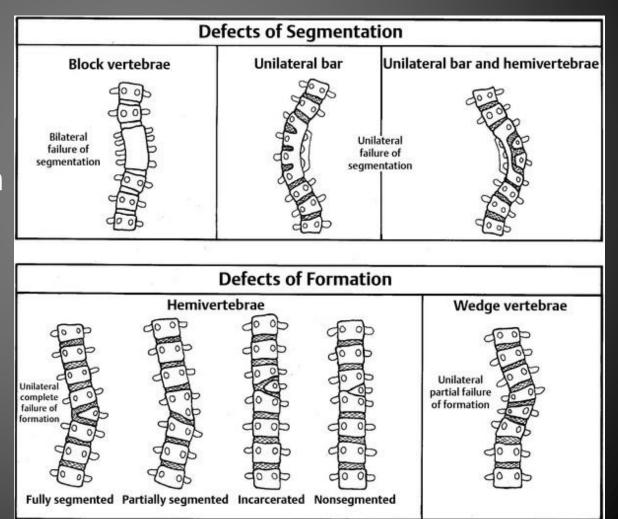


### **Scoliosis Surgical Intervention**

Congenital

Failure of
 Segmentation

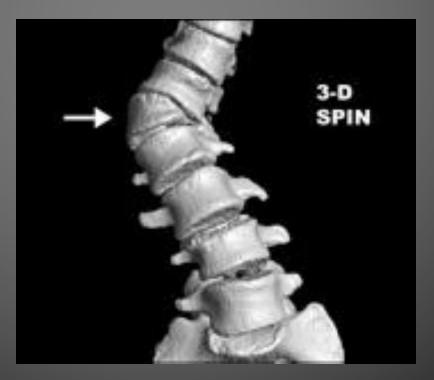
Failure ofFormation



#### Congenital

- Hemivertebrae & Contralateral Bar Formation
- Significant Progression
  - Segmental Hemivertebrae –usual progress
    - Age 3-8 yo
    - Posterior Approach
    - Localized Fusion
  - Unilateral Bar usual progress
    - Localized fusion
- Neurologic Deficit
- Declining Respiratory Status

- Congenital
  - 3D CT Scan is helpful for surgical intervention
    - Especially with Vertebrecomy



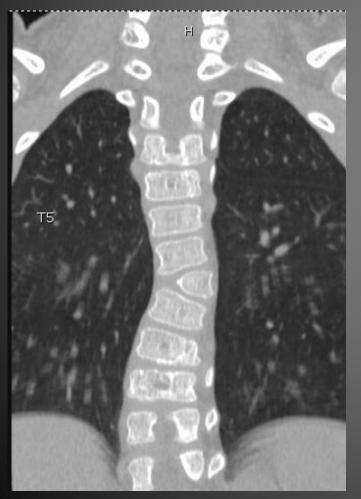
• Congenital – 6 yo female with hemivertebrae





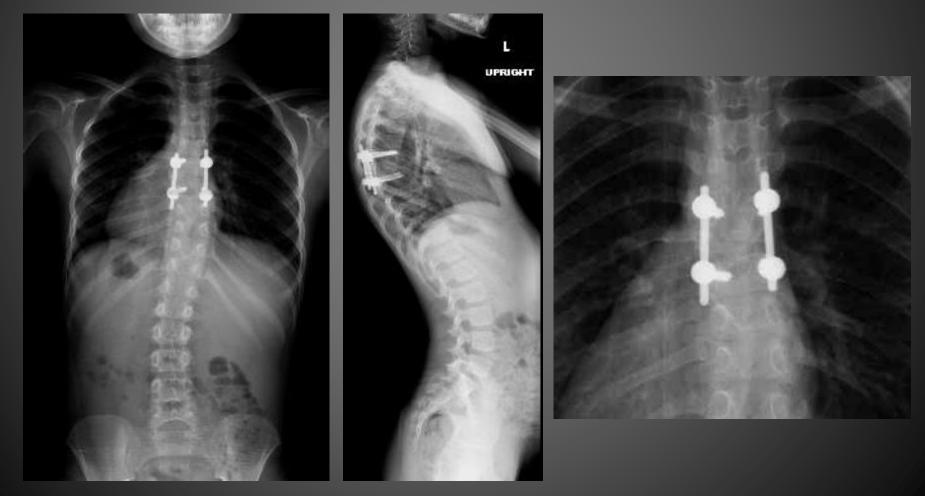


Congenital – 6 yo female with hemivertebrae





Congenital – 6 yo F w/ Hemivertebrae Excision

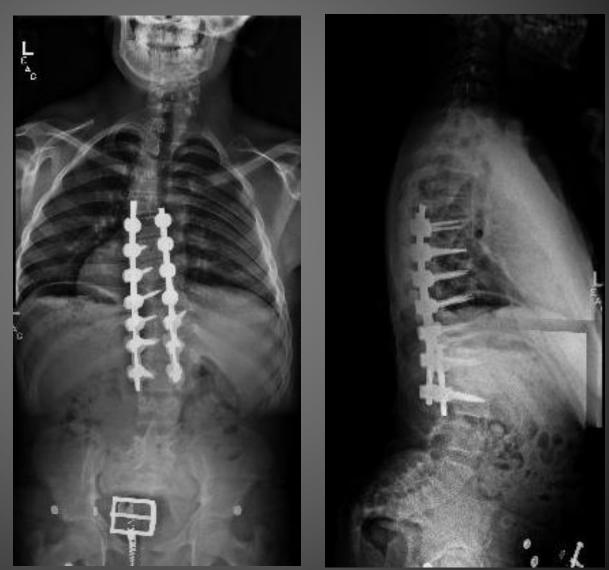


Congenital – 11.5 yo M w/ Congenital Bar





Congenital



# **Scoliosis Surgical Intervention**

#### • <u>Syndromic</u>

- Prader Willi
- MarFan
- Neurofibromatosis
- Angelman
- Down Syndrome
- Prune Belly
- Ehlers Danlos

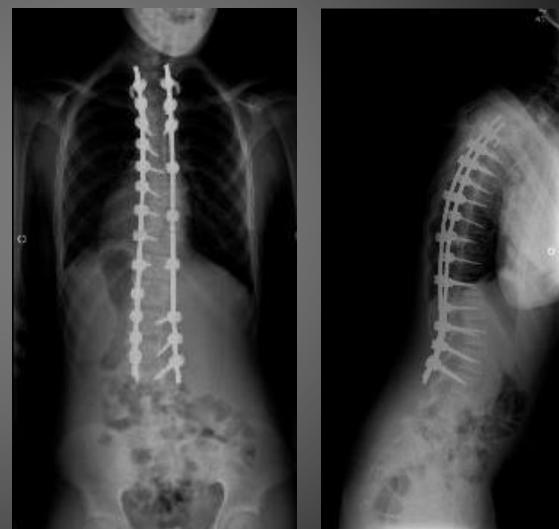
- OI
- Muscular Dystrophy
- Noonan
- VATER/VACTRL
- Rett
- Osteochondrodystrphy
  - Dwarfism

- Variable Progression
- No set indication different than other scoliosis except DMD
  - Multiple approaches depending on deformity & progression
- DMD
  - 20-30° in nonambulatory patients
  - Wait until 40° if resonding well to corticosteroids
  - Operate before pulmonary function decline

 12.5 yo M w/ Down Syndrome & h/o Cong. Heart Defect s/p repair as infant



• Down Syndrome



#### • 5.5 yo F w/ h/o clubfoot & torticollis



# Neurofibromatosis Type 1

Syndromic – NF1
– 6.5 yo F



• NF1 - 7yo F



• NF1 - 9.5 yo F



# When do you operate on a child with Scoliosis?

- Depends on ...
  - Type of Scoliosis
  - Progression
  - Pt. Age
  - Pt. Maturity
  - Curve Size
  - Neurologic Status
  - Functional Impairment



# Thank You

• Questions

