

The Spectrum of Early Onset Spinal Deformity in Skeletal Dysplasia

ICEOS 2016
Utrecht, Holland



Suken A. Shah MD

William G. Mackenzie MD

Wilmington DE USA

Nemours Children's Health System

Spine Problems in Skeletal Dysplasia

Instability - O-C 1, C 1-2, Sub -axial

Stenosis – Cervical, thoracic or lumbar

Deformity – Scoliosis/Kyphosis/Lordosis



Clarify diagnosis prior to treatment as the natural history and associated problems vary with dysplasia type

Scoliosis

- Spondyloepiphyseal dysplasia
- Diastrophic dysplasia
- Chondrodysplasia punctata
- Metatropic dysplasia
- Camptomelic dysplasia
- Pseudoachondroplasia



Diastrophic Dysplasia

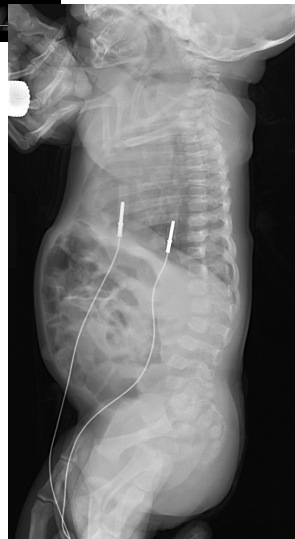
- Scoliosis
 - Remes et al Spine 2001
- Incidence ~ 75%
- Onset
 - Most common 2nd decade
 - < 3y – more progressive and severe
- Bracing (? Useful)
- Spinal stenosis common
- Poor spine flexibility
 - Vertebral deformity, facet and disc degeneration



Diastrophic Dysplasia



4 months



1.5 years



Initially braced then serial “Mehta” casting





**Diastrophic
Dysplasia – 3 m**

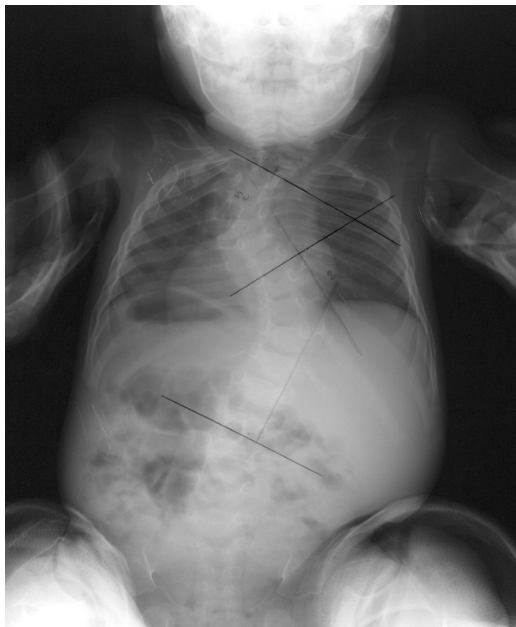


Serial “Mehta” Cast

Pulmonary Problems in Skeletal dysplasia

- Casting for Early Onset Scoliosis
 - Causes restrictive lung disease
 - Peak inspiratory pressures increased an average of 108% with cast application prior to belly window cutout
 - Most pressures returned to within 10% baseline with the cutout
- Impulse Oscillometry and Thoracoabdominal Motion Analysis
 - Pulmonary function evaluation possible in very young
 - Chest resistance and compliance can be determined
 - Abnormal thoraco/abdominal motion can be measured

Diastrophic Dysplasia



19 months

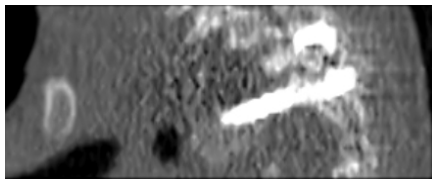
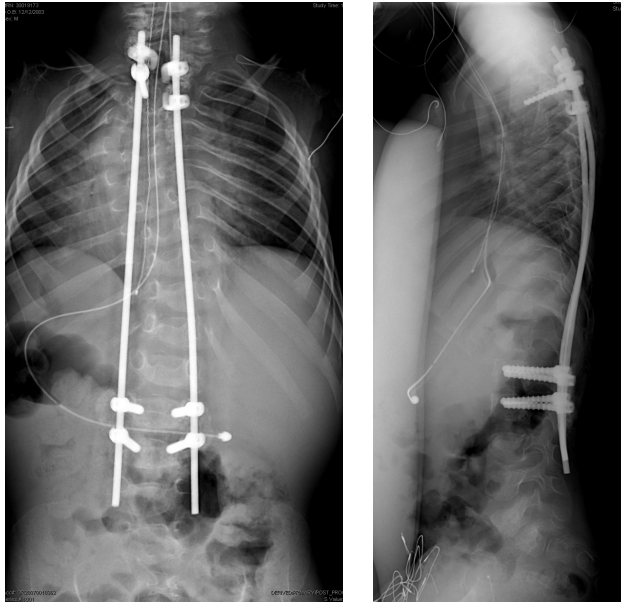


2 years 9 months

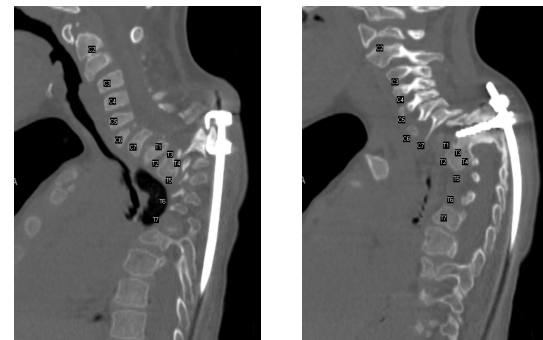
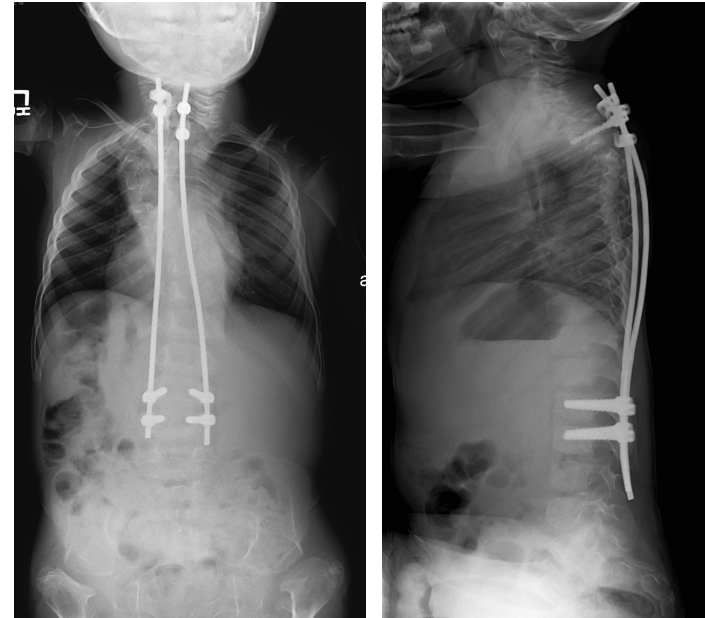


Diastrophic Dysplasia

Growing Rod

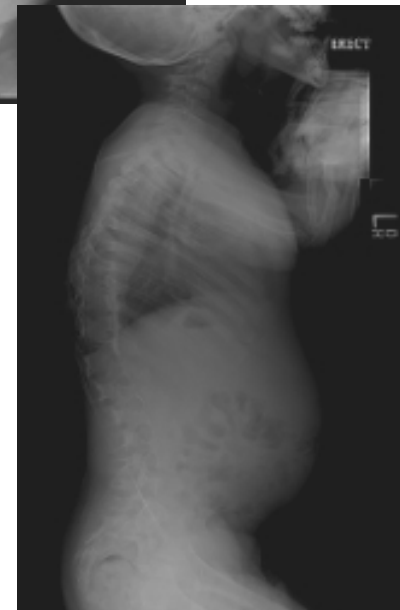


3 yo



5 yo

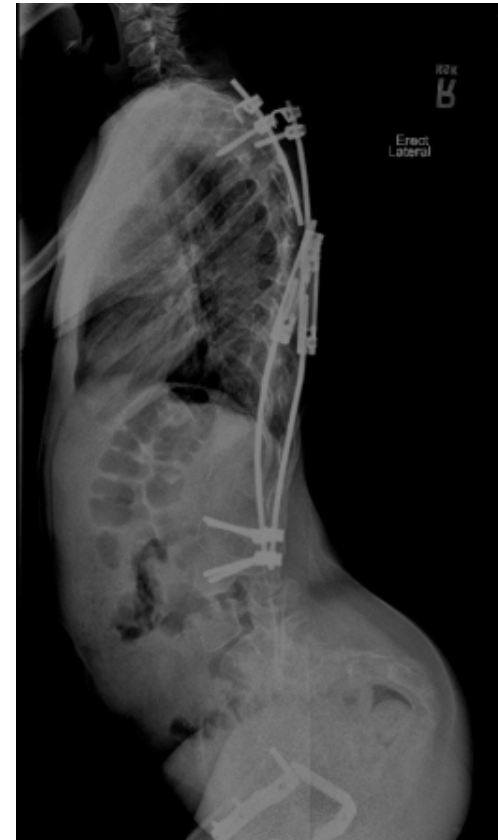
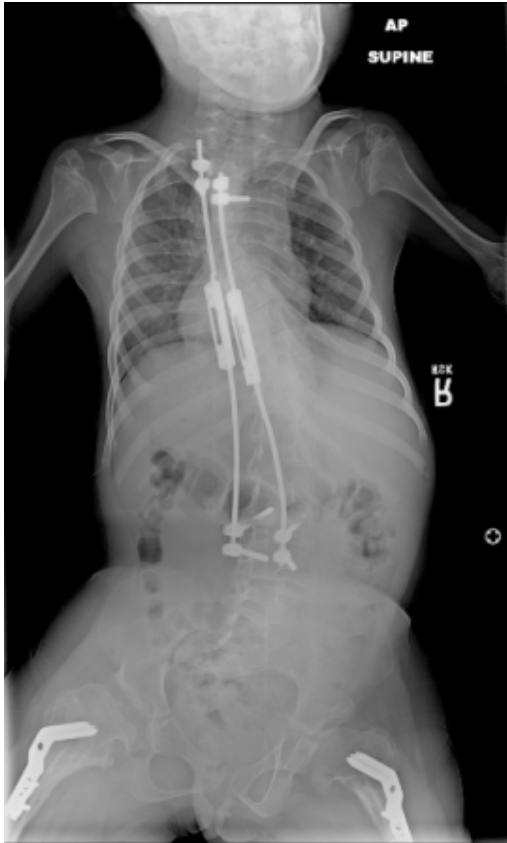
Diastrophic Dysplasia 4 years



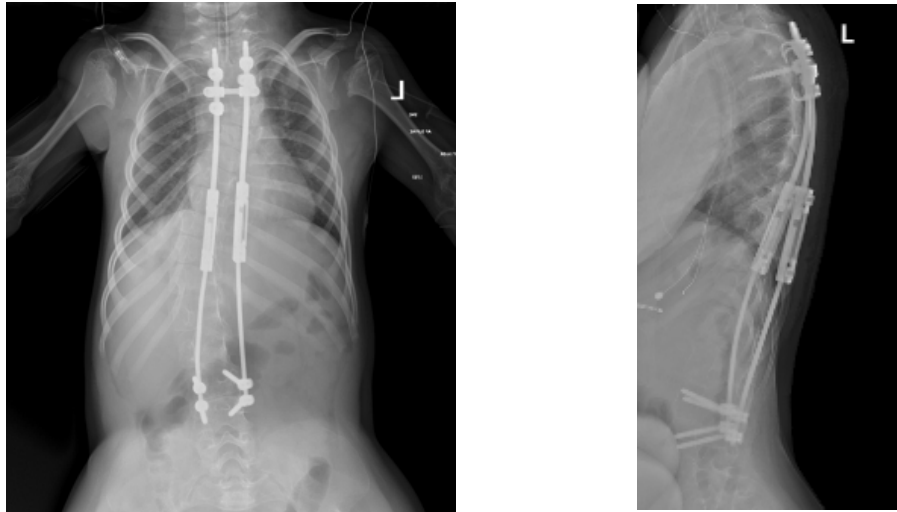
Spinal Growing Rod System - 4 years old



One revision of thoracic fixation and then rod fracture at 9 years with loss of upper thoracic fixation



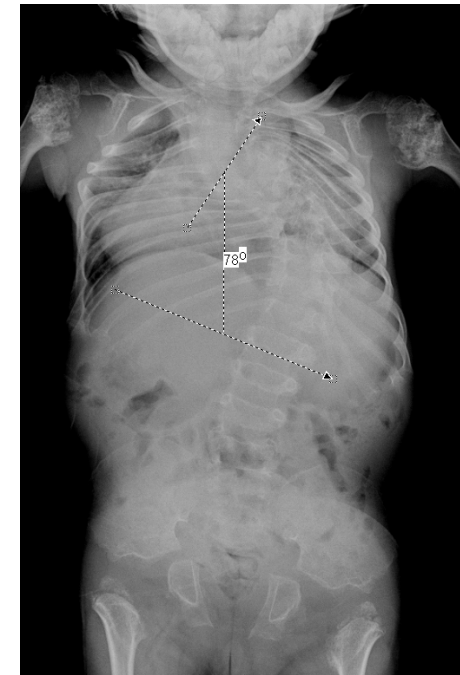
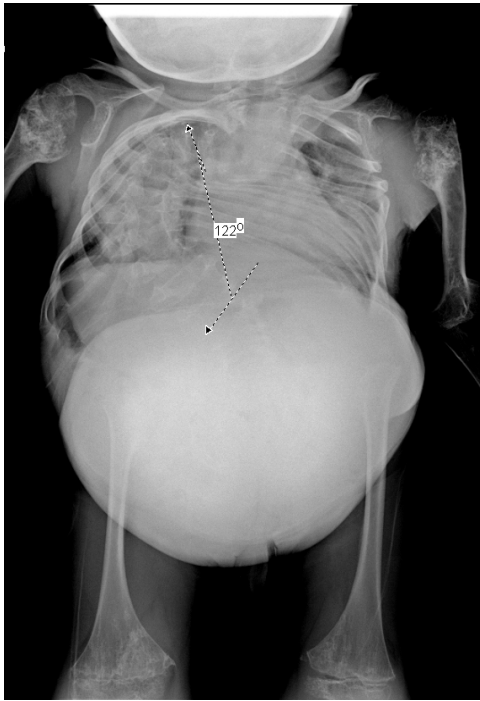
Replaced Rods and revised fixation and now 10 years old



■ Results of Growing Rod Systems

- Bess et al *JBJS Am* 2088, Watanabe et al *Spine* 2013, Kabirian et al *JBJS Am* 2014, Noordeen *Spine* 2011
- Infection
- Revision
 - Fixation failure, rod breakage, length
- Psychological
- MAGEC- early but promising- Akbarnia *Spine* 2013

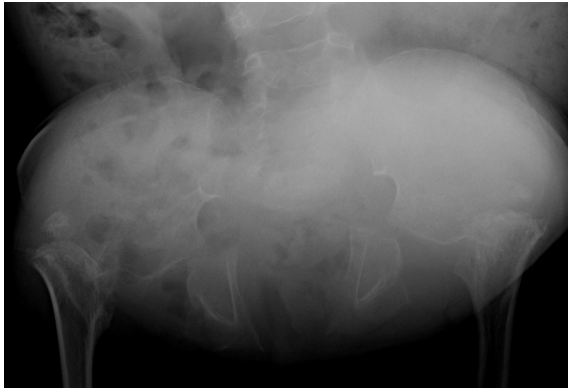
SEMD - Kyphoscoliosis



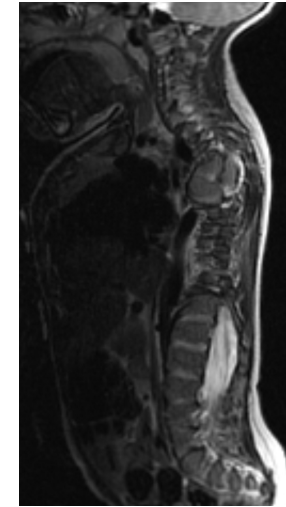
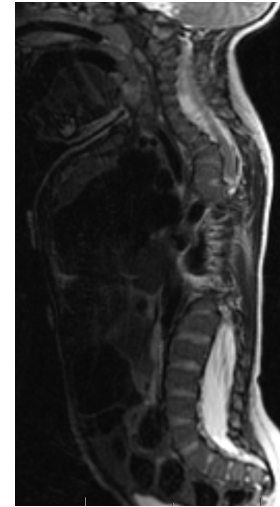
Traction

- 7yo boy
 - Untreated severe kyphoscoliosis with lordosis assoc with coxa vara
 - Neurological exam normal
 - Restrictive lung disease

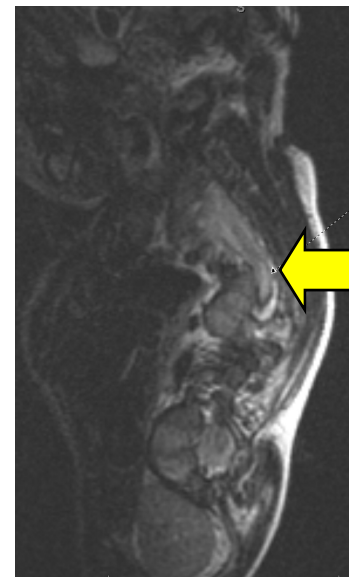
Anterior Spinal Cord Injury



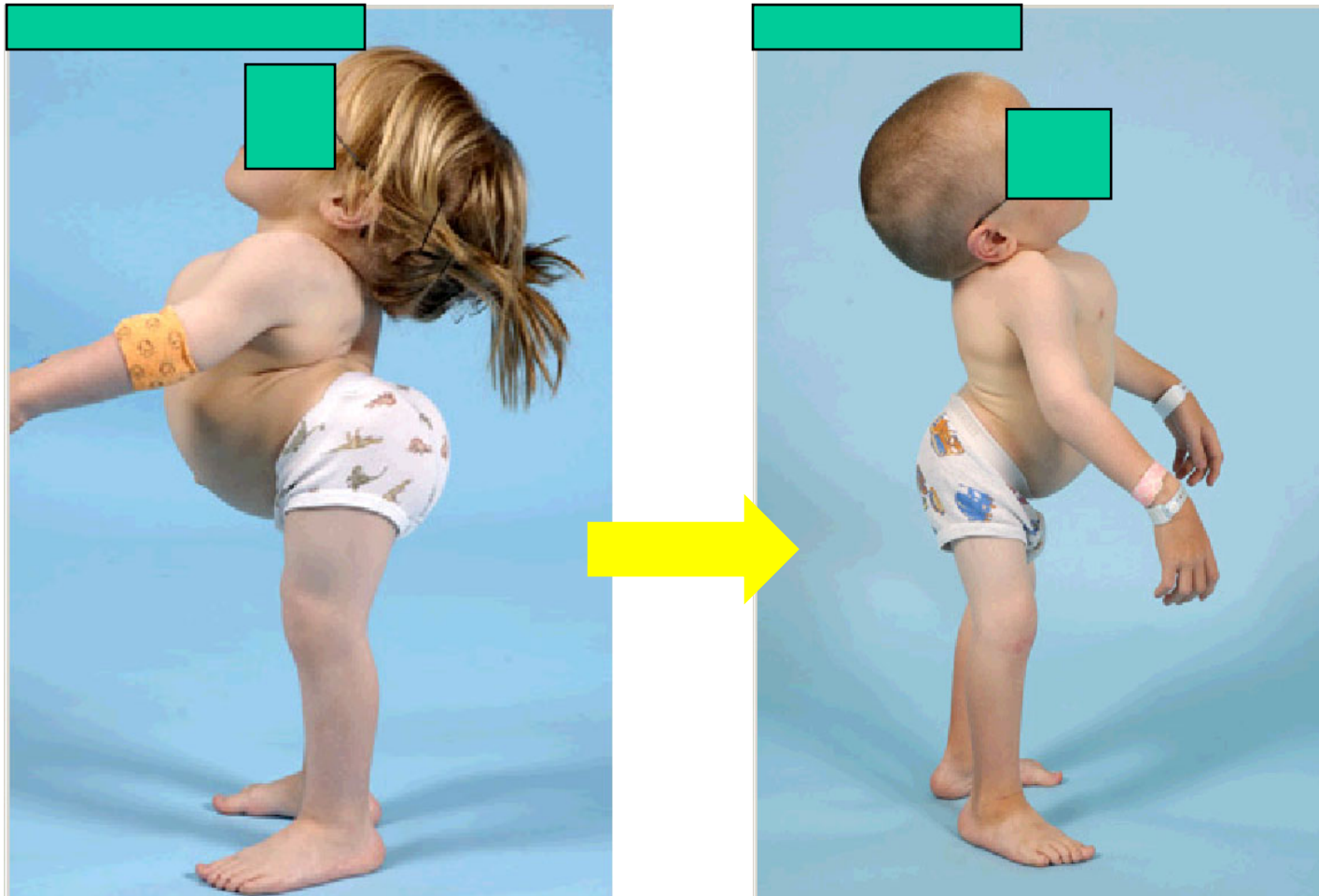
~3 hrs, 200 cc blood loss, no movement of legs post op with good sensation



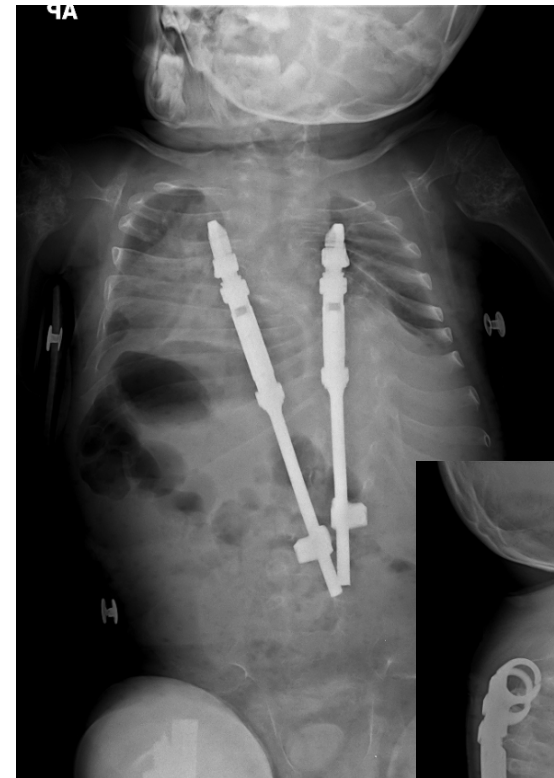
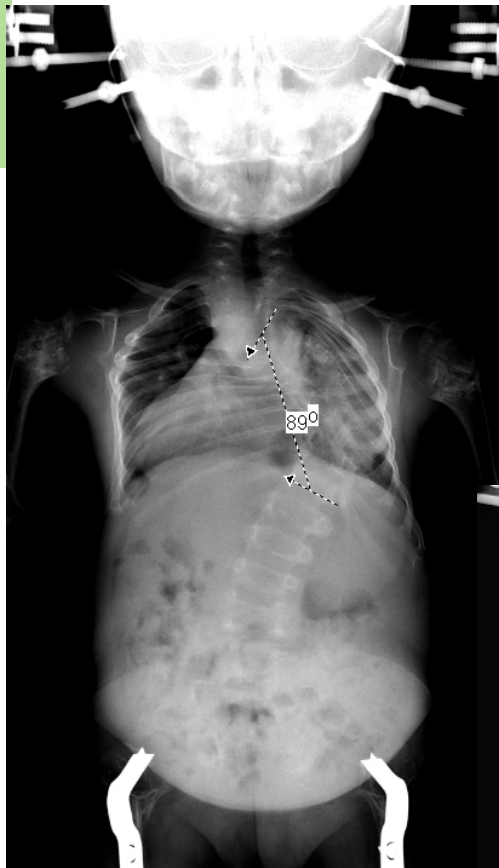
Preop MRI



Postop MRI



**Difference between these pictures is ~ 6 months,
a haircut and correction of coxa vara**

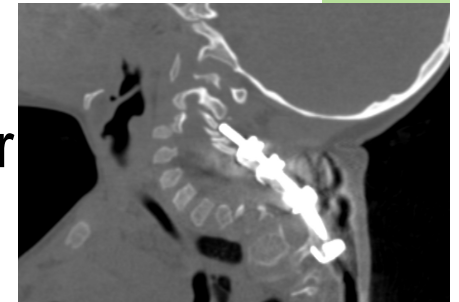
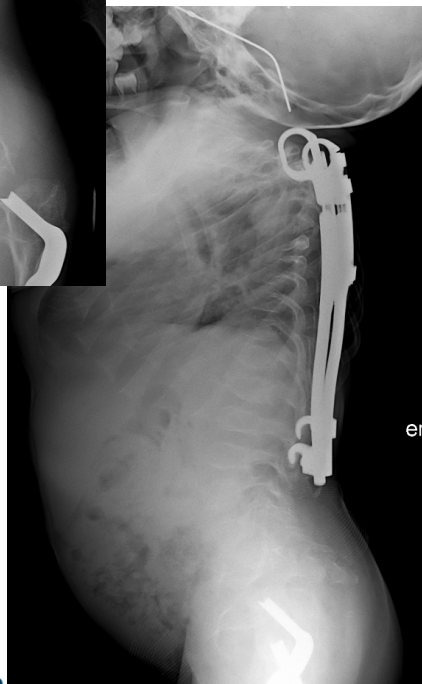


8y

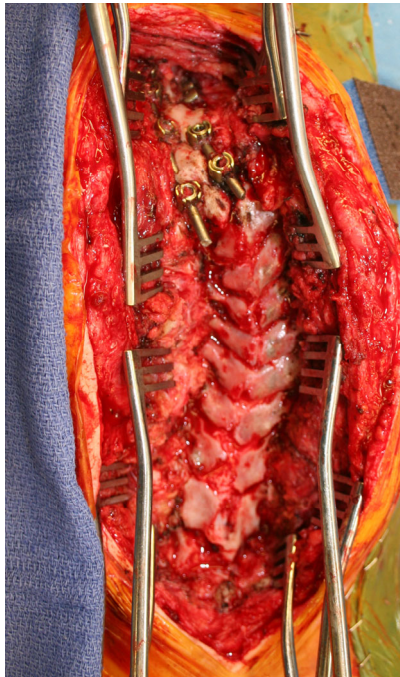
**Neurological
recovery**

**Halo Traction 6 m
later**

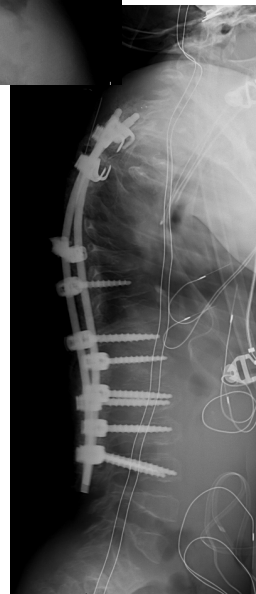
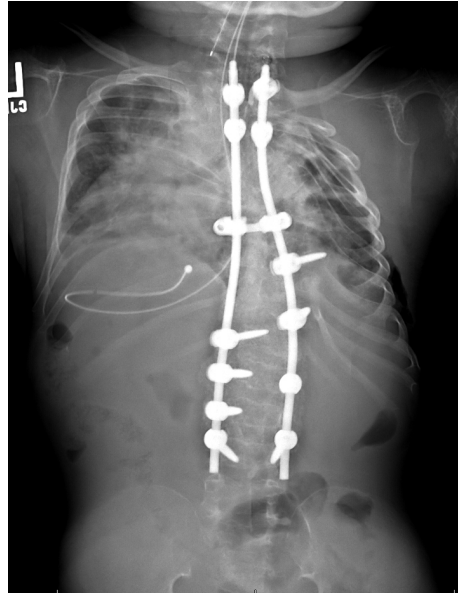
- 11yo
- Progressive kyphosis at upper end of VEPTR
- PSF and instrumentation



Final PSF and Instrumentation- 12y



No spontaneous fusion



Metatropic Dysplasia Kyphoscoliosis - 17 months



Metatropic Dysplasia

8 years old



Stiff chest,
Restrictive lung
disease



Anterior Thoracoscopic Release with Halo traction (bed, sitting and standing)



Post op



Metatrophic Dysplasia, Kyphoscoliosis

- 7-year-old boy with metatrophic dysplasia, kyphoscoliosis, restrictive lung disease and an anomalous pulmonary artery.
- Presented with a progressive kyphoscoliosis with the kyphosis measuring 105 degrees preoperatively
- Anterior thoracoscopic release of the apex of his kyphosis and was placed in halo traction for 4
- He demonstrated remarkable improvement and on the standing and extension x-rays there was excellent correction
- Neurological examination remained normal

Pre-Op



Traction



Initial Post-Op



1st lengthening-9/2015

Left side 5.5 mm

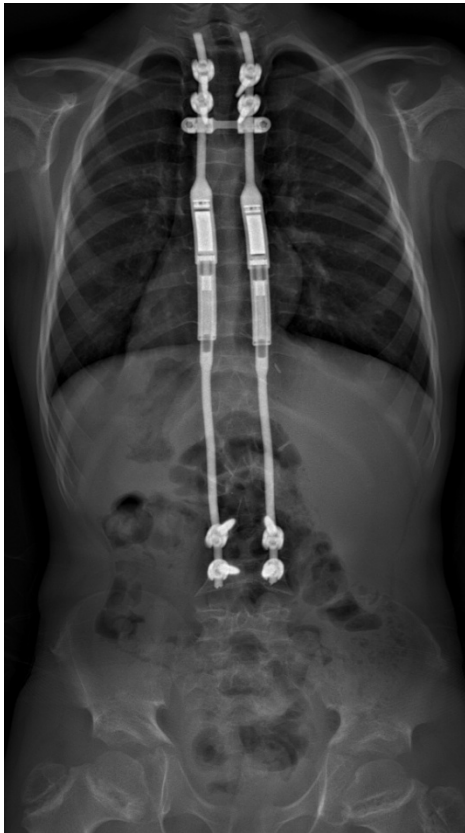


Right side 5.5mm



2nd Lengthening-1/2016

Left side 2.8 mm 3 rounds



2.5 mm 3 rounds



3rd Lengthening-4/2016

Left side 2.0mm 3 rounds

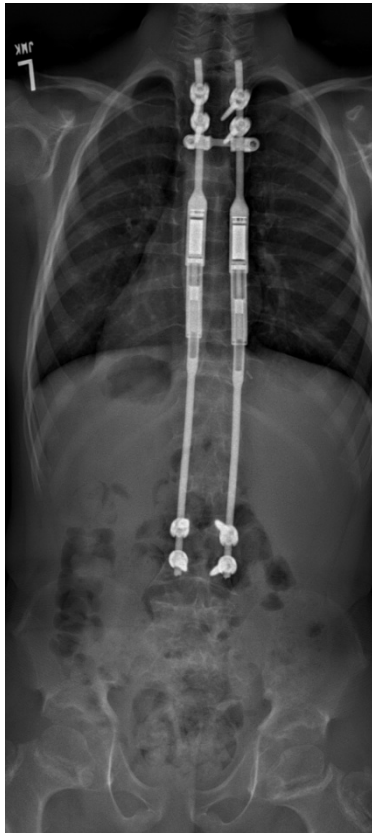


Right side 2.5mm 3 rounds



4th Lengthening-7/2016

Left side 3.0mm 2 rounds

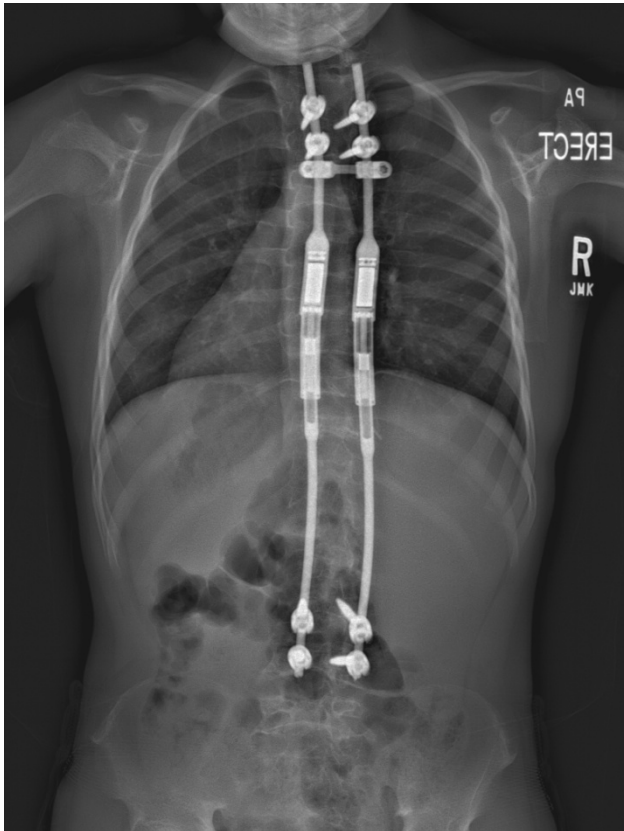


Right side 3.0mm 1 round



5th Lengthening-11/2016

Left side .5mm- 3 rounds(one of the rounds with 1 compression/1 distraction)



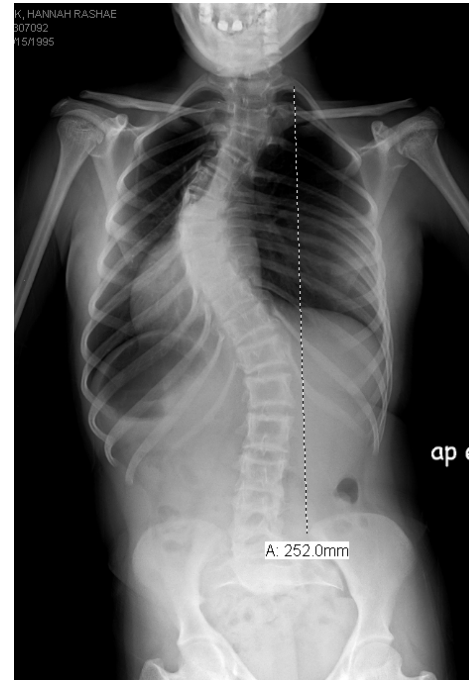
Right side 2.0 3 rounds



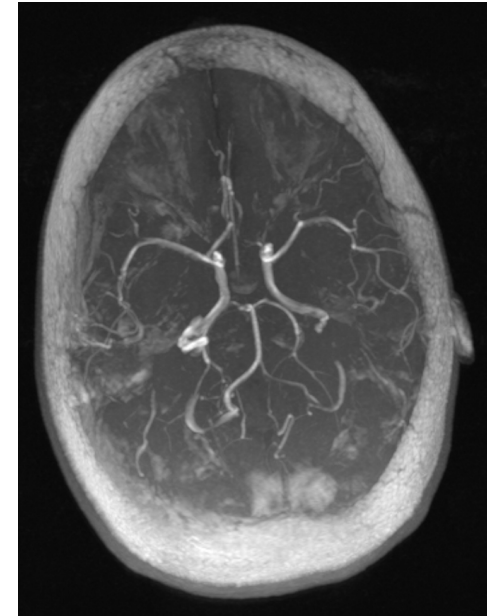
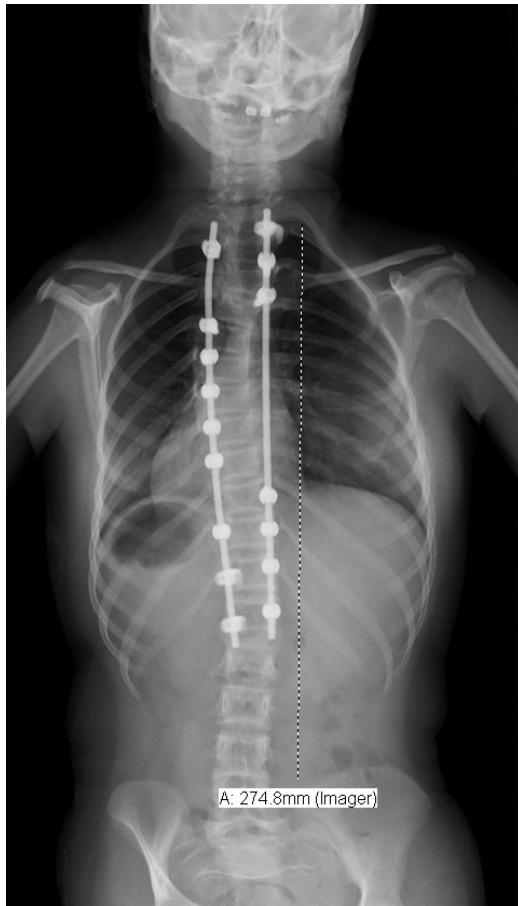
SMD



Primordial Dysplasia



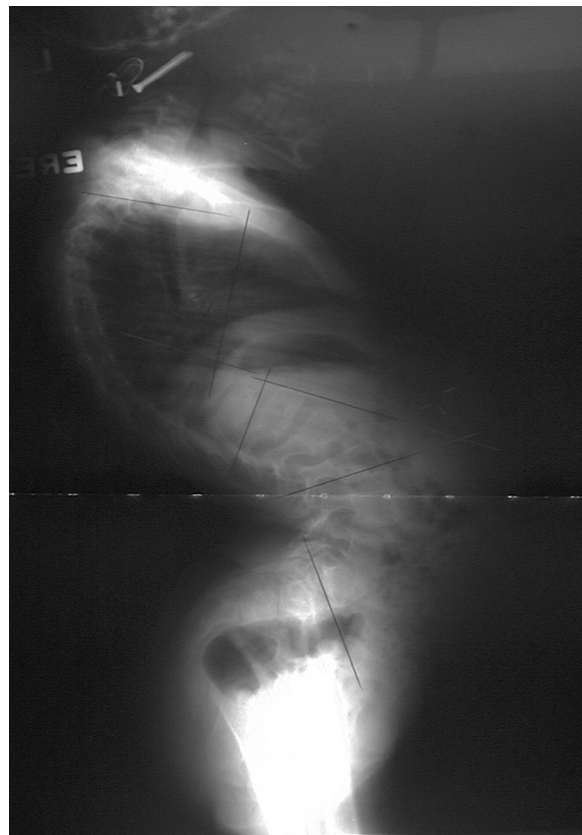
- **Majewski Osteodysplastic Primordial Dysplasia**
 - **Type II**
- 12yo
- 3'3", 28 lb
- T1-L5 - 25 cm
- SMALL endotracheal tube



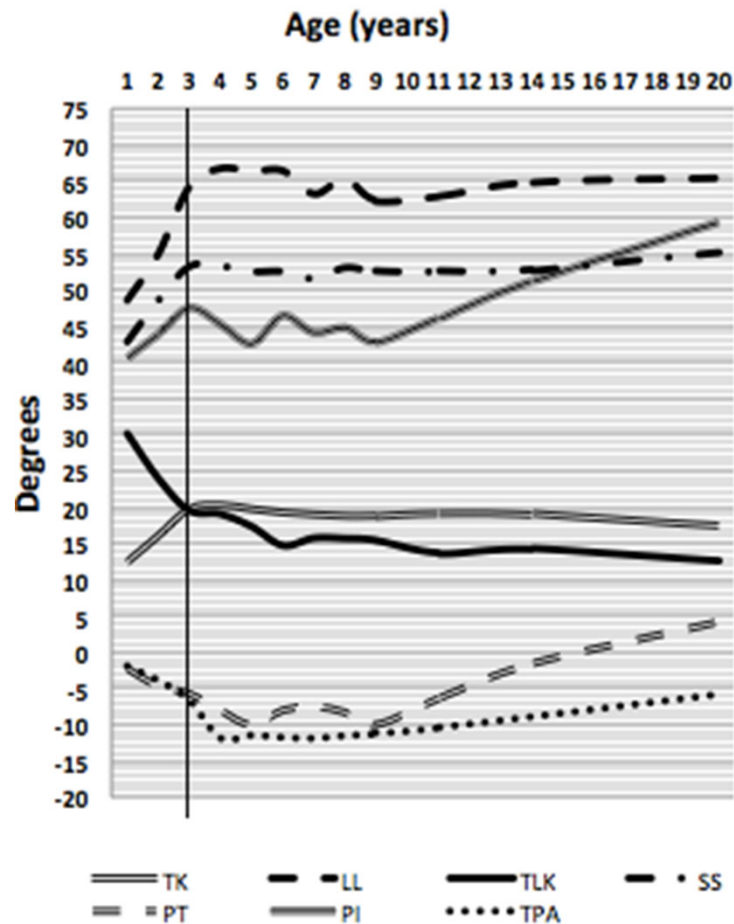
- 3.5 mm titanium instrumentation
- These children can have severe vascular disease
 - Moya moya

Relationship of Spine and Limb Alignment

- Sagittal spine malalignment (thoracic kyphosis, lumbar lordosis) often associated with flexion contractures in the lower extremities

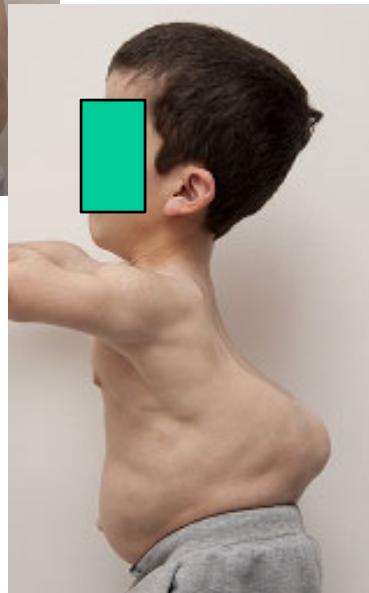


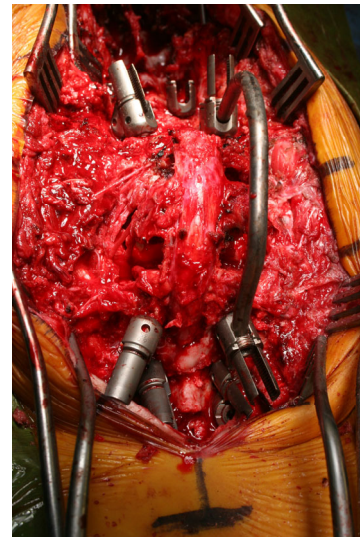
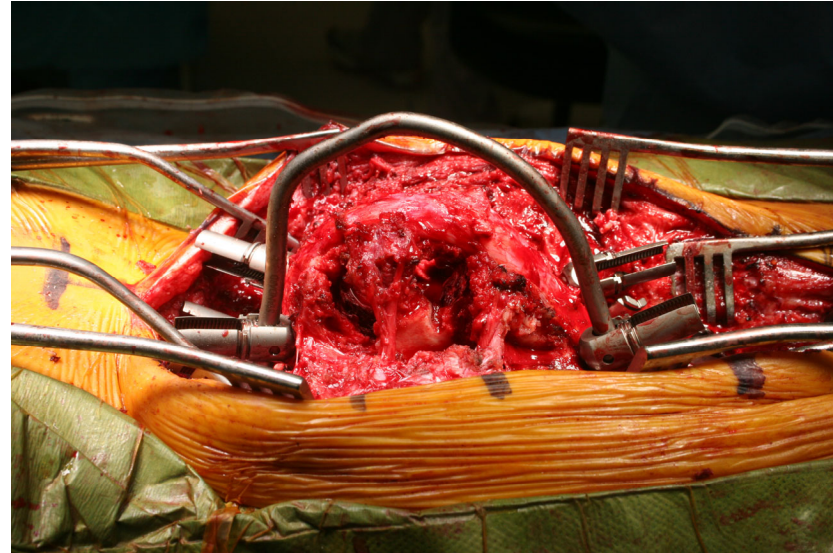
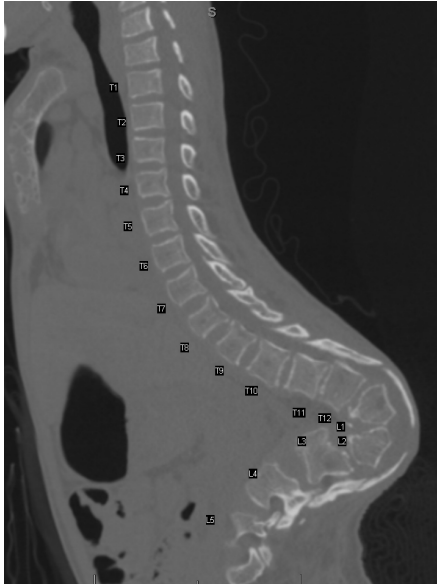
Relationship of Spine and Limb Alignment

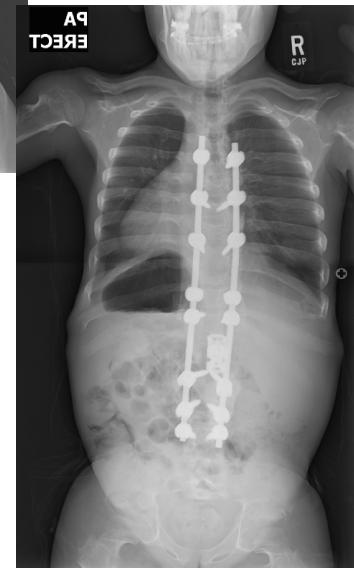
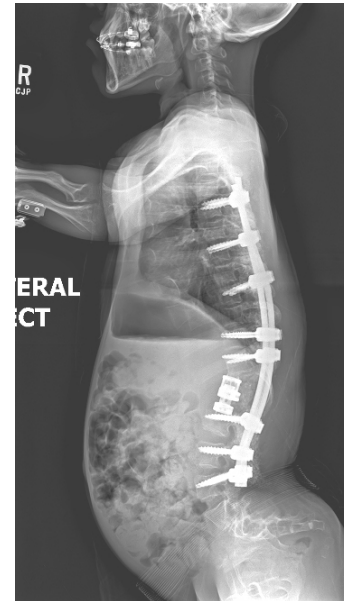


Achondroplasia

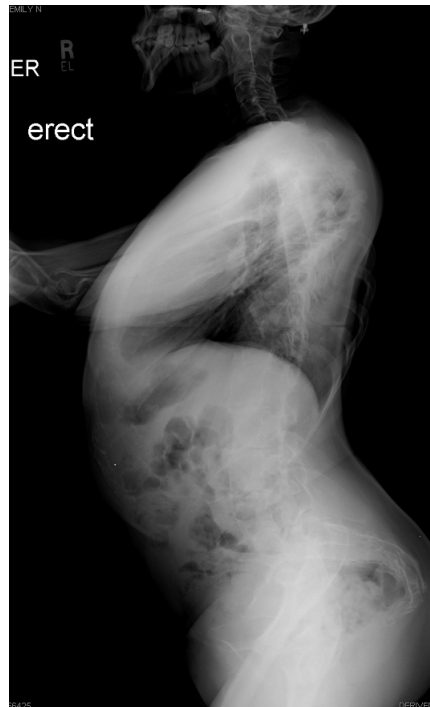
16y- previous ASF/PSF with pseudarthrosis, progression and symptomatic spinal stenosis





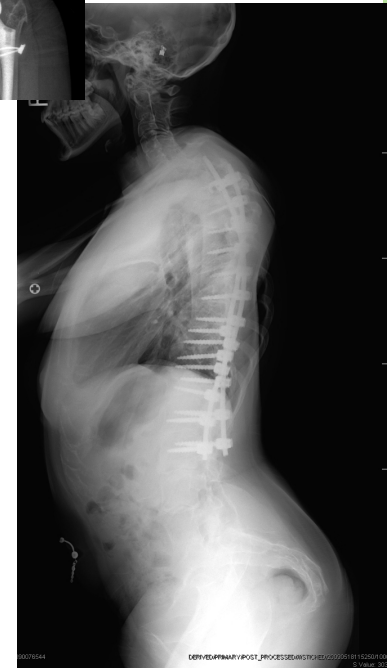
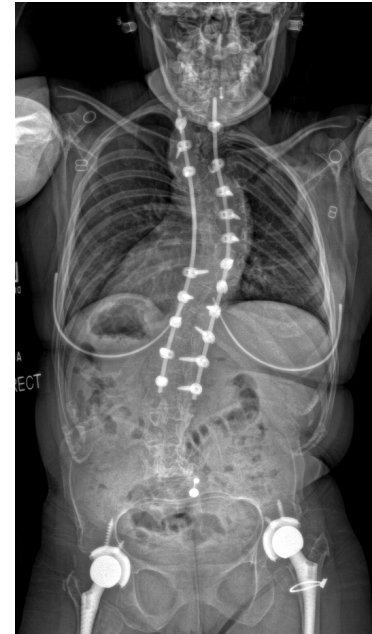


Diastrophic Dysplasia



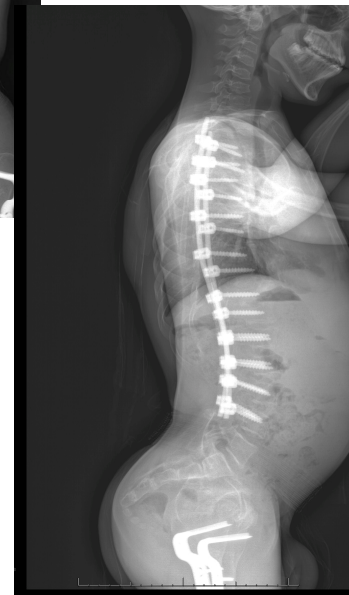
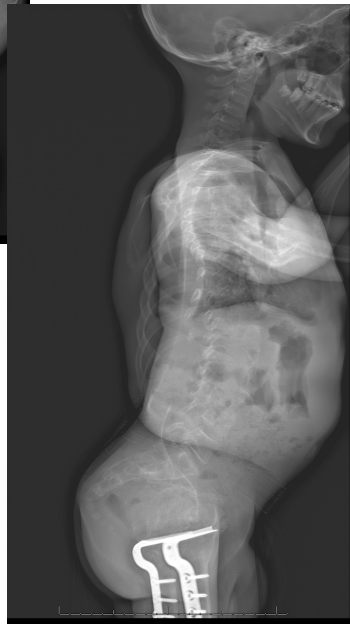
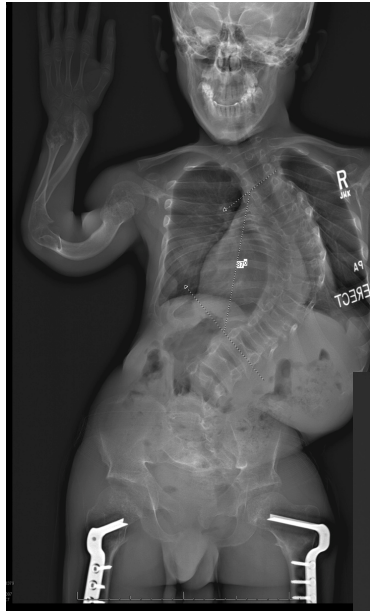
18 y with progressive scoliosis
and normal neurological exam





Spinal Stenosis common

SEDCC – 16y



Conclusions

- If the diagnosis is known, natural history is predictable
- Preserve thoracolumbar growth and avoid aggravating restrictive lung disease
- Beware kyphosis