



Syndromic Spine Deformity

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Alfred I. duPont
Hospital for Children

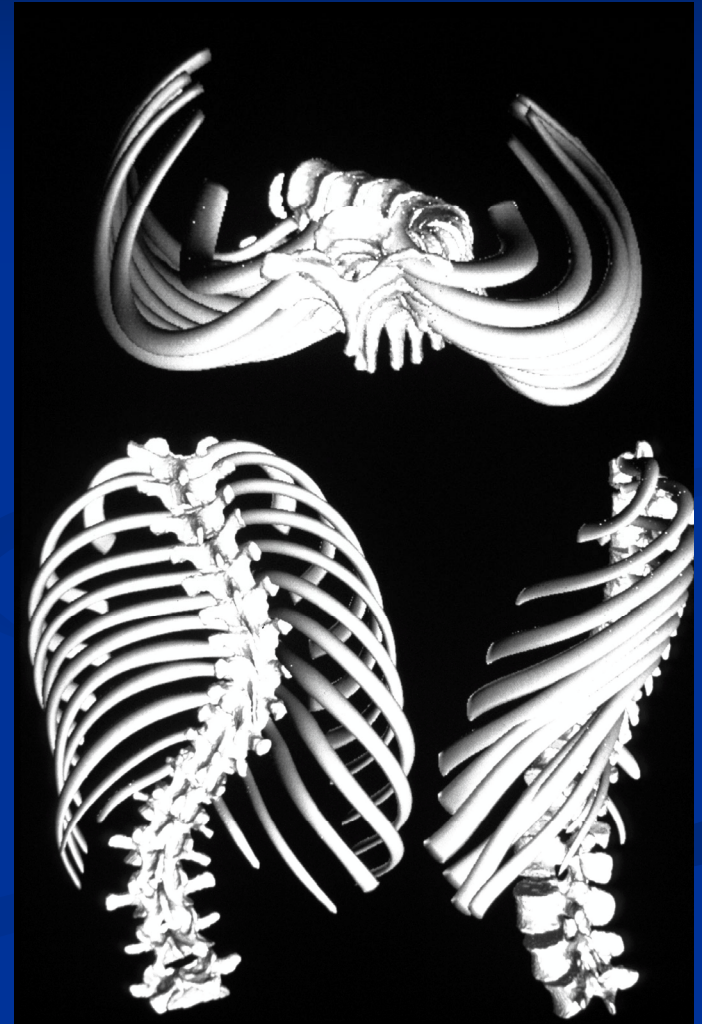
Nemours
Children's Clinic

My Disclosures

- DePuy Spine, Inc.
 - Research support, Consultant, Royalties
- K Spine, Inc.
 - Consultant
- Axial Biotech, Inc.
 - Research support

Idiopathic Scoliosis

- Anterior overgrowth vs Posterior tether
- Buckling of column
- Crankshaft-like effect w/ rotation
- Heuter-Volkman changes



Syndromic Scoliosis

- Osteoporotic bone
- Stiff curves
- Congenital elements
- Braces don't work
- Bleeds more
- Poor signals
- Medically complex
- Pulmonary disease / TIS
- Frail
- Nutritionally depleted

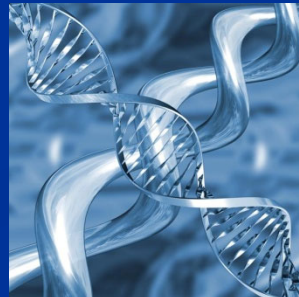


Systematic Approach

- Establish diagnosis of specific syndrome and become familiar with it
 - Specific co-morbidities, risks, predilections
 - Orthopaedic and other medical manifestations of syndrome
- Dysmorphic child with “undiagnosed syndrome” – consider re-evaluation by geneticist
 - Impossible to make specific preparations

Syndromic Early Onset Scoliosis

- Marfan
 - Gene encoding fibrillin-1 (15)
 - Autosomal dominant
 - 25% incidence of spont mutations
- Larsen
 - AD, some sporadic
- Arthrogryposis
 - Heterogeneous, sporadic
- Neurofibromatosis - 1
- Goldenhaar
- Jeune

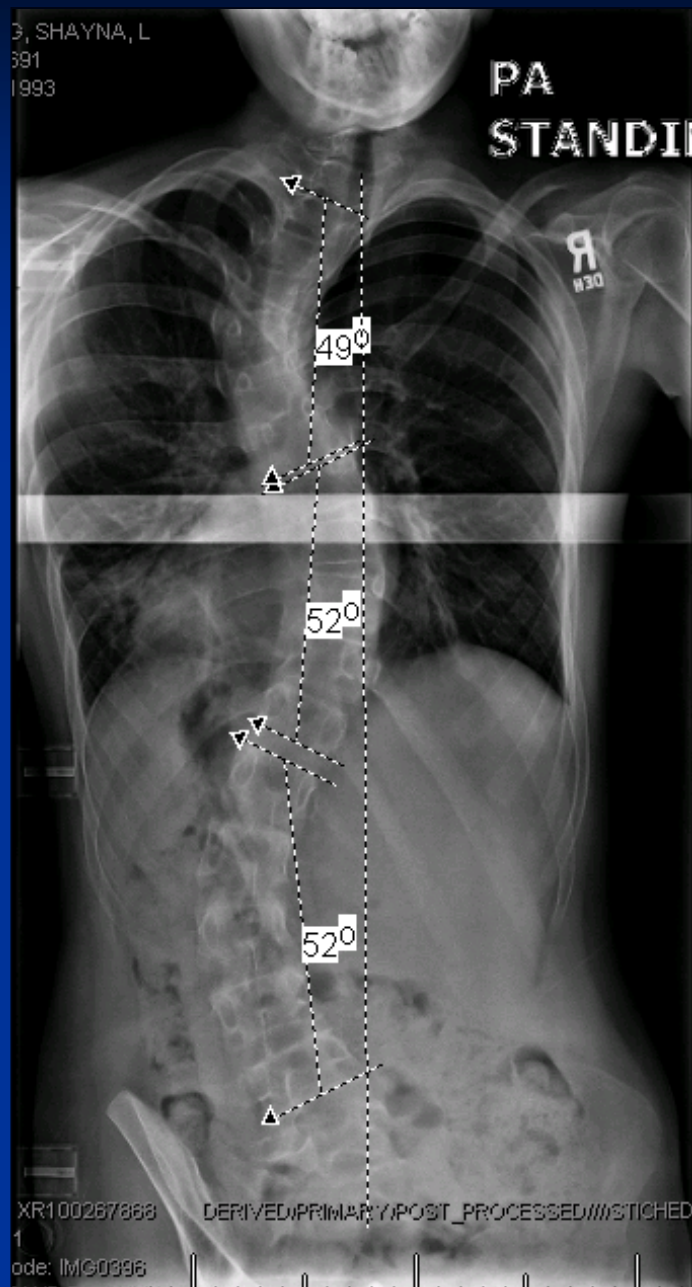


Systematic Approach

- Preoperative workup
 - PA, lat, bending X-rays
 - MRI of the entire spine (and head when indicated)
 - CT of the spine and chest
 - Congenital anomalies – hemivertebra, bars, occult bifidas,
 - 3D reconstructions
 - Lung volumes, rib anomalies, TIS
 - Examine neck, hips, knees, feet
- Consultations from medical specialists – should be a partnership

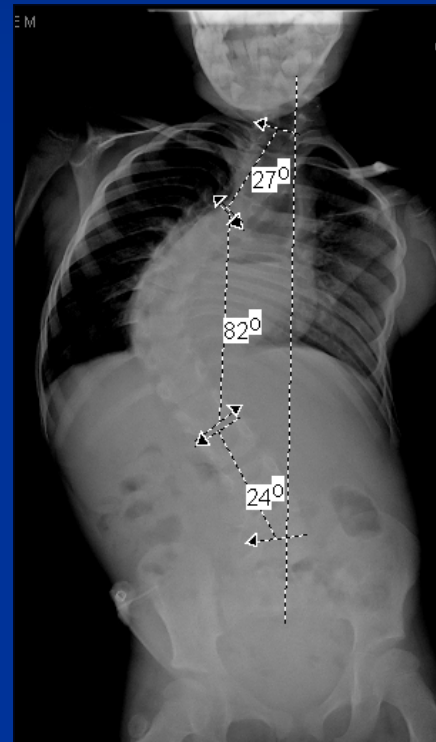
Systematic Approach

- Family Conference – discuss expectations, plan and complications
 - ...and document it!
- Operating Room
 - Assemble your “A” team
 - Prepare for excessive bleeding – IV access, blood products, warmers, antifibrinolytics
 - Instrumentation concerns – inventory, special implants, poor bone bail outs (old school)
 - Monitoring

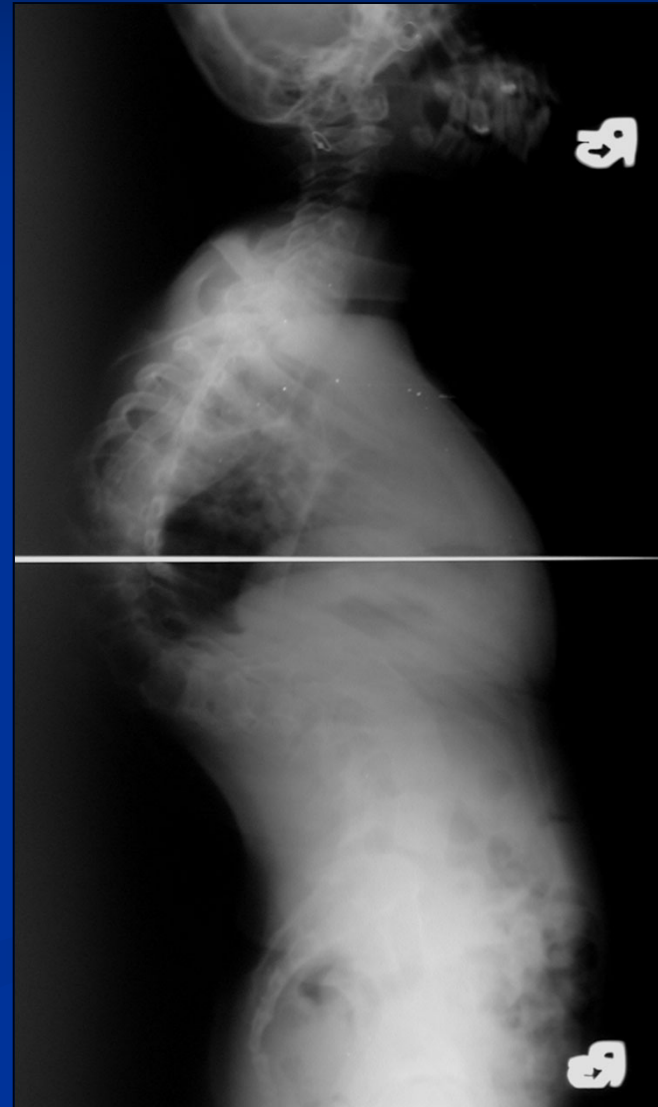


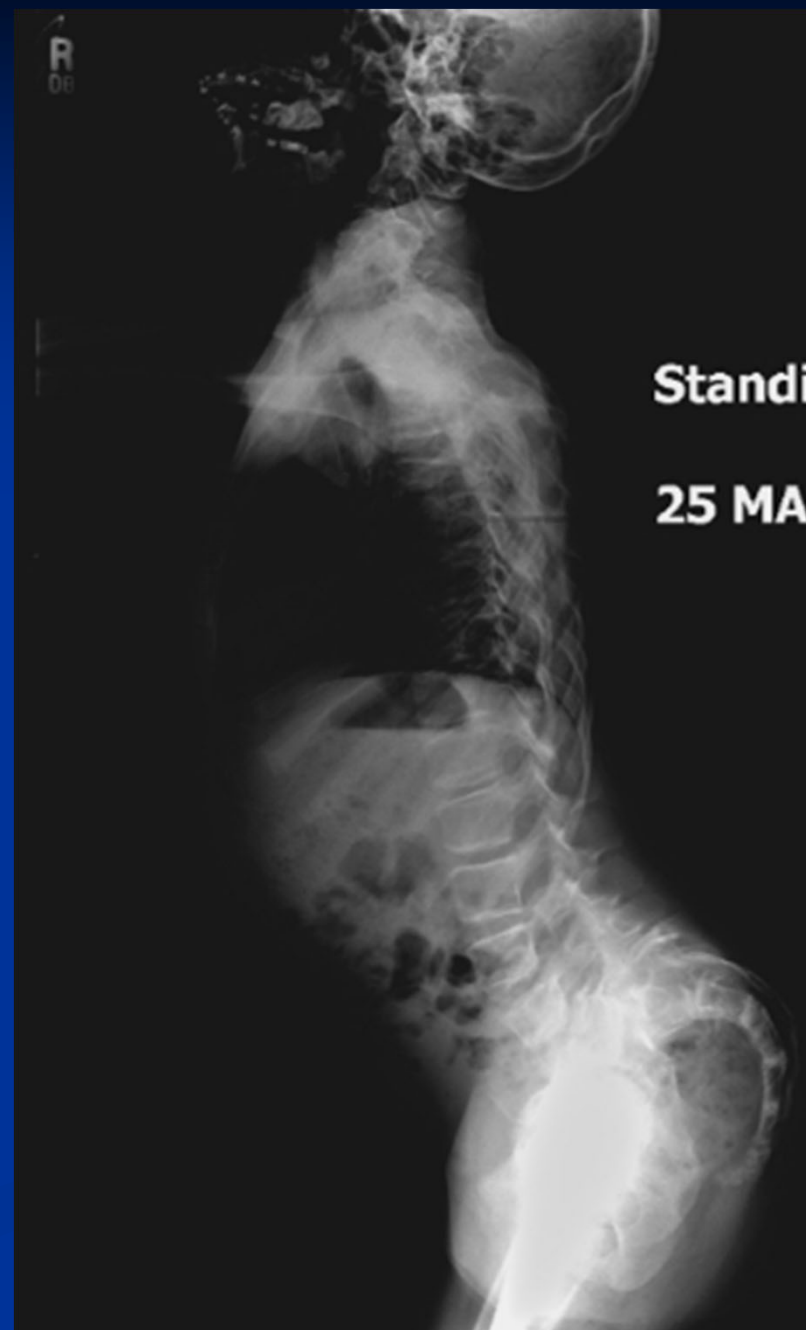
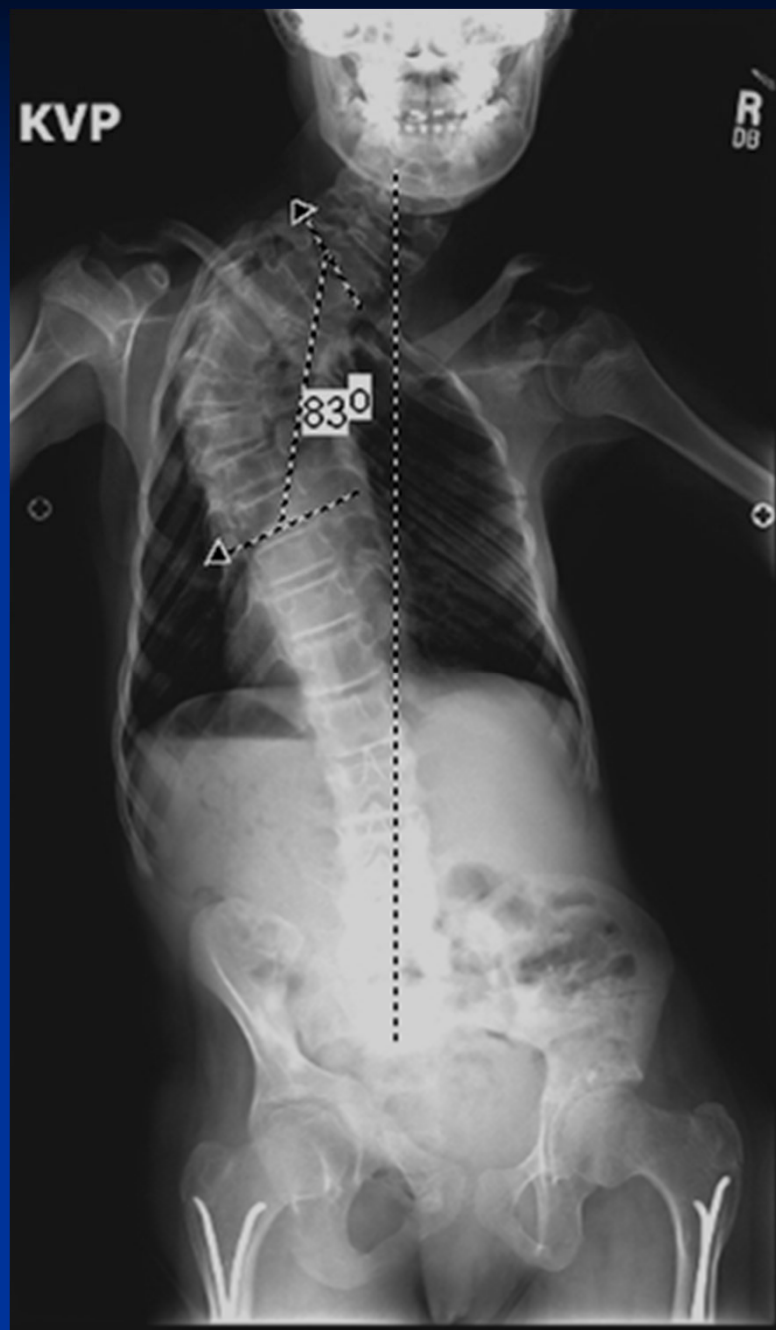
Classification

- Bone Dysplasia
- Connective Tissue
- Neurological
- Mixed



Severe Kyphoscoliosis

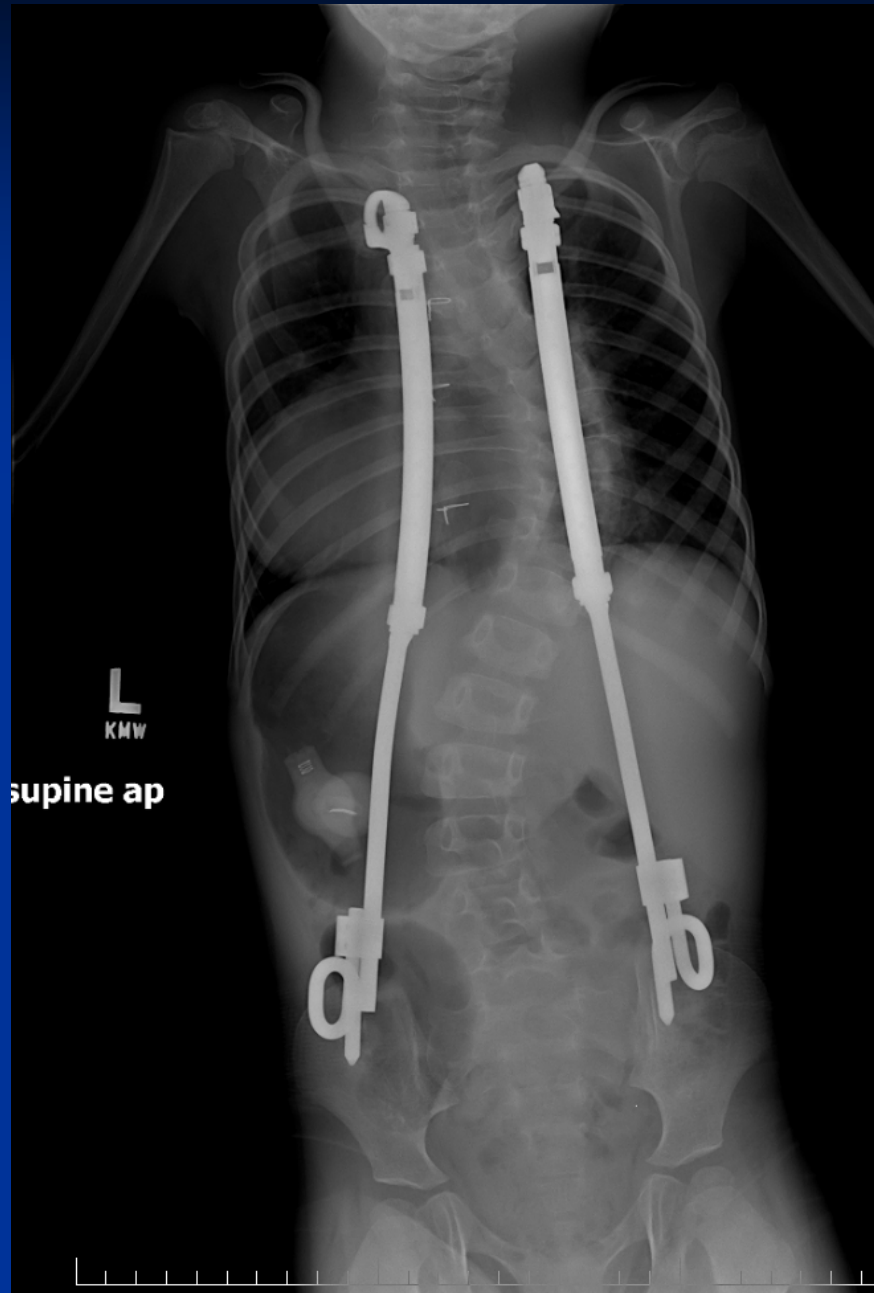






REED, NICHOLAS M
MRN: 10490390
D.O.B: 5/13/2004
Sex: M

Study Date: 12/20/2006
Study Time: 10:21:36



Acq#: DP00050055904
Series #: 1002
Procedure Code: 72069E
Spine Scoliosis PABLAT-Erect(72069E)

DERIVED: PRIMARY POST-PROCESSED
S Value: 226.00
Zoom: 33.9%
VIEW: 926 / 266



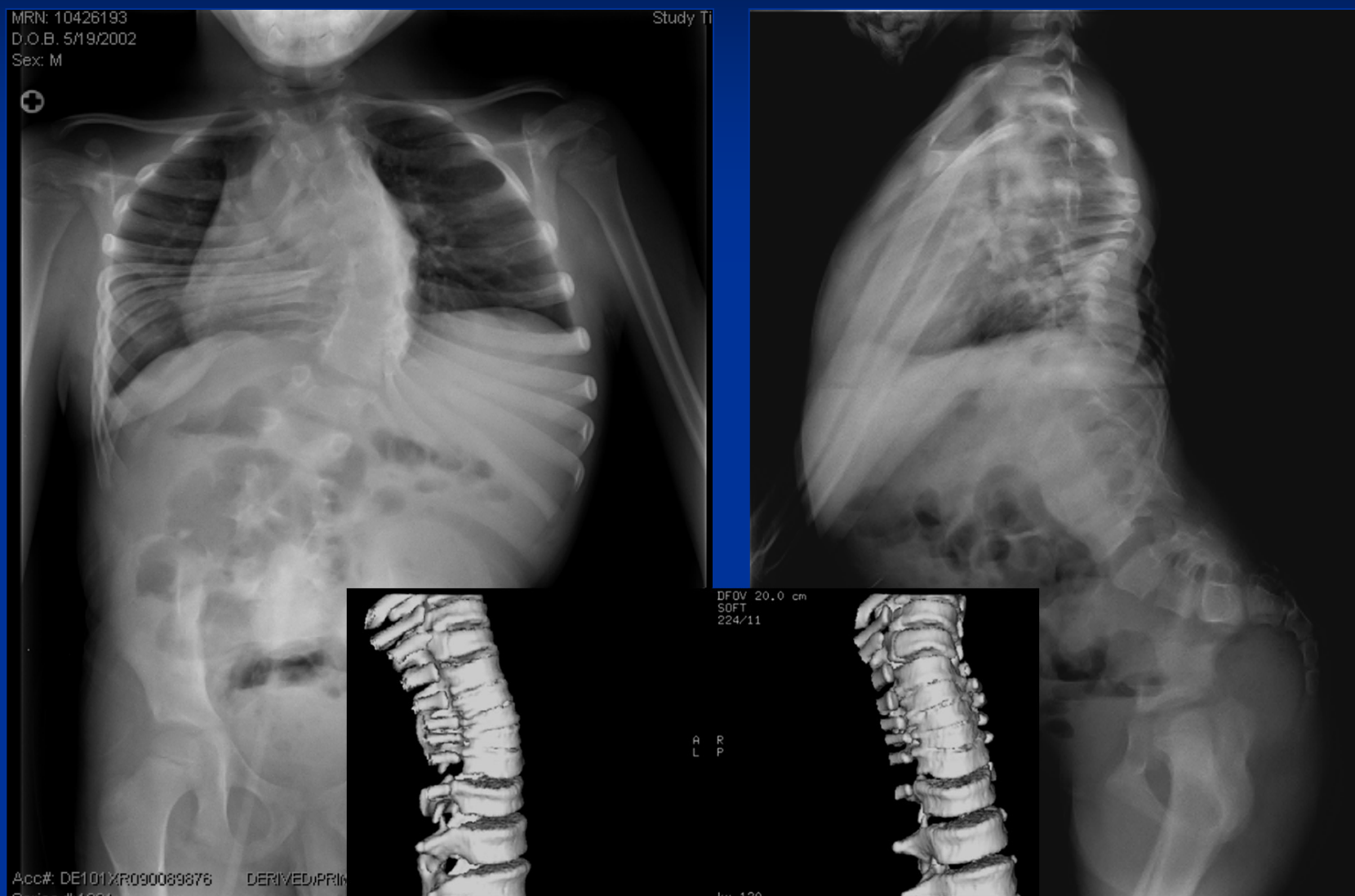
Syndromes associated with small thorax and stiff curves

- Arthrogryposis*
- Camptomelic dysplasia
- Cerebro-costo-mandibular *
- Chondroectodermal dysplasia
- Cleido-cranial dysostosis*
- Infantile hypophosphatasia*
- Jarcho-Levin
- Jeune thoracic dysplasia
- Metatropic dysplasia *
- Trisomy 8, 9 or mosaics
- Achondroplasia
- Diastrophic dysplasia *
- Osteogenesis imperfecta
- Oto-palatal-digital
- Pseudoachondroplasia
- Chondrometaphyseal dysplasia

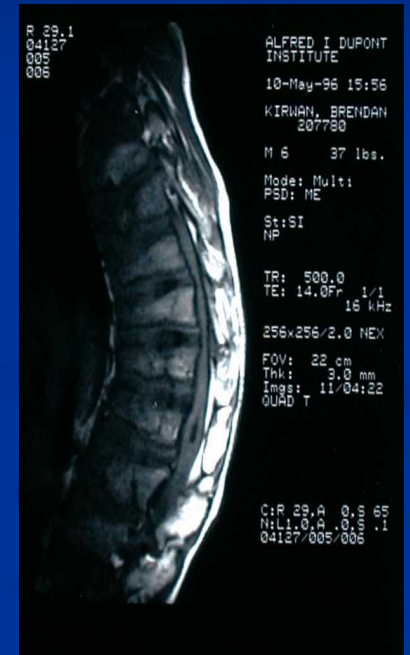
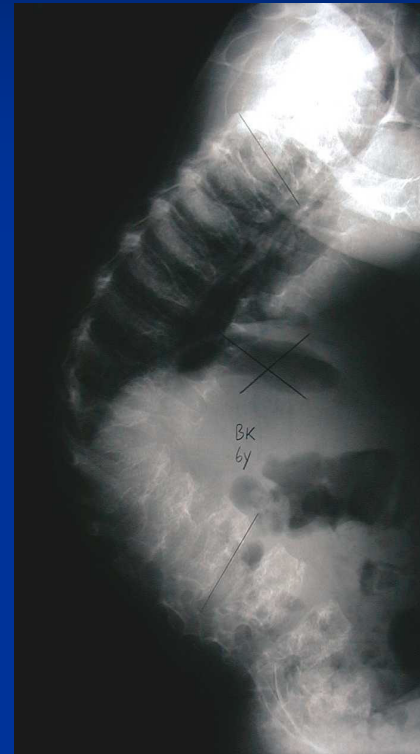
* - scoliosis frequently associated

Courtesy of Charles Johnston MD

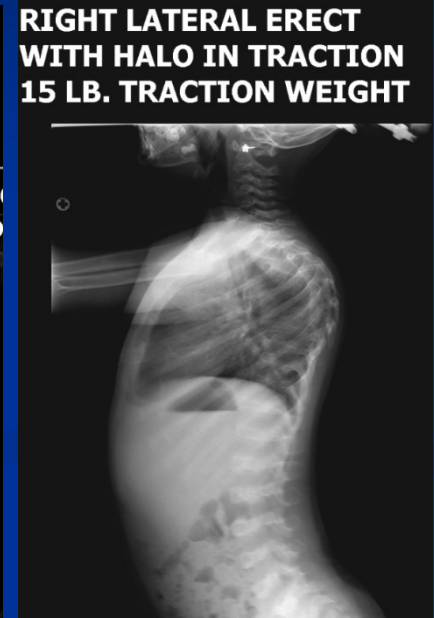
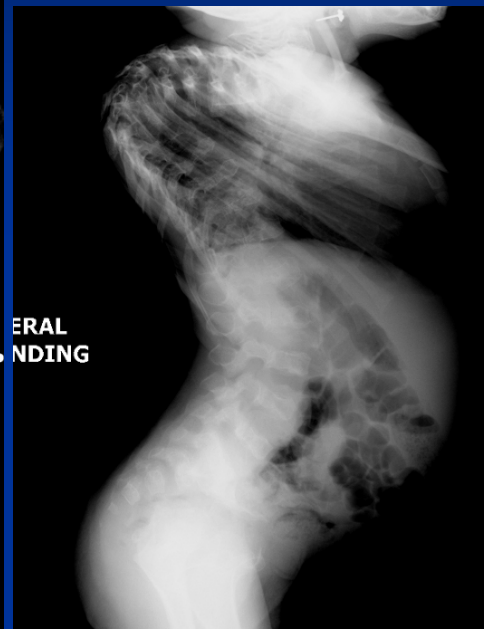
Congenital Scoliosis & Arthrogryposis



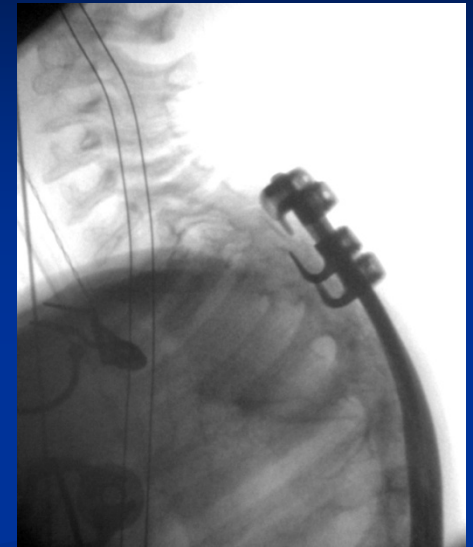
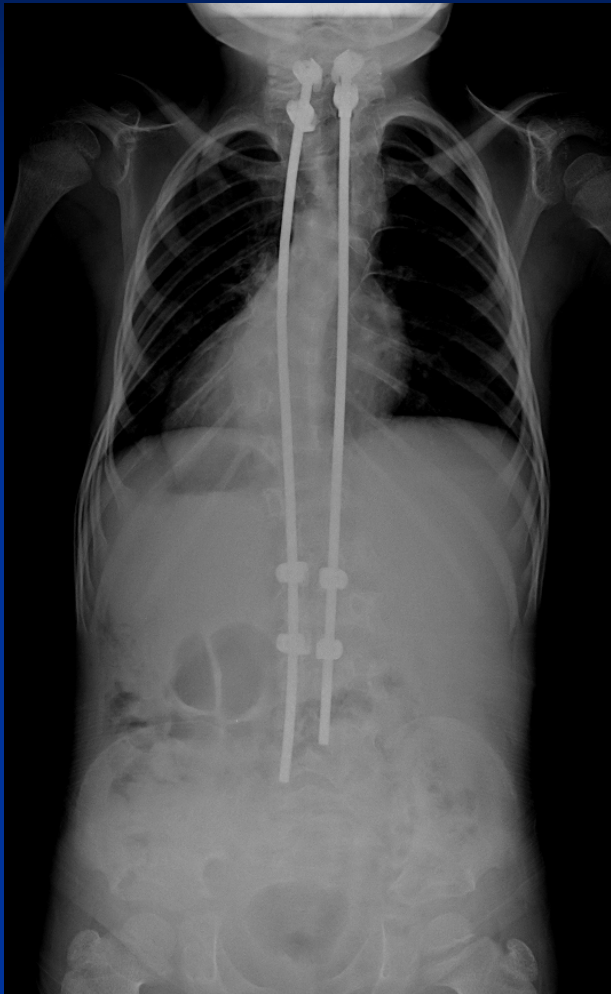
Metatropic Dysplasia



Growing Spine in Syndromic Deformity

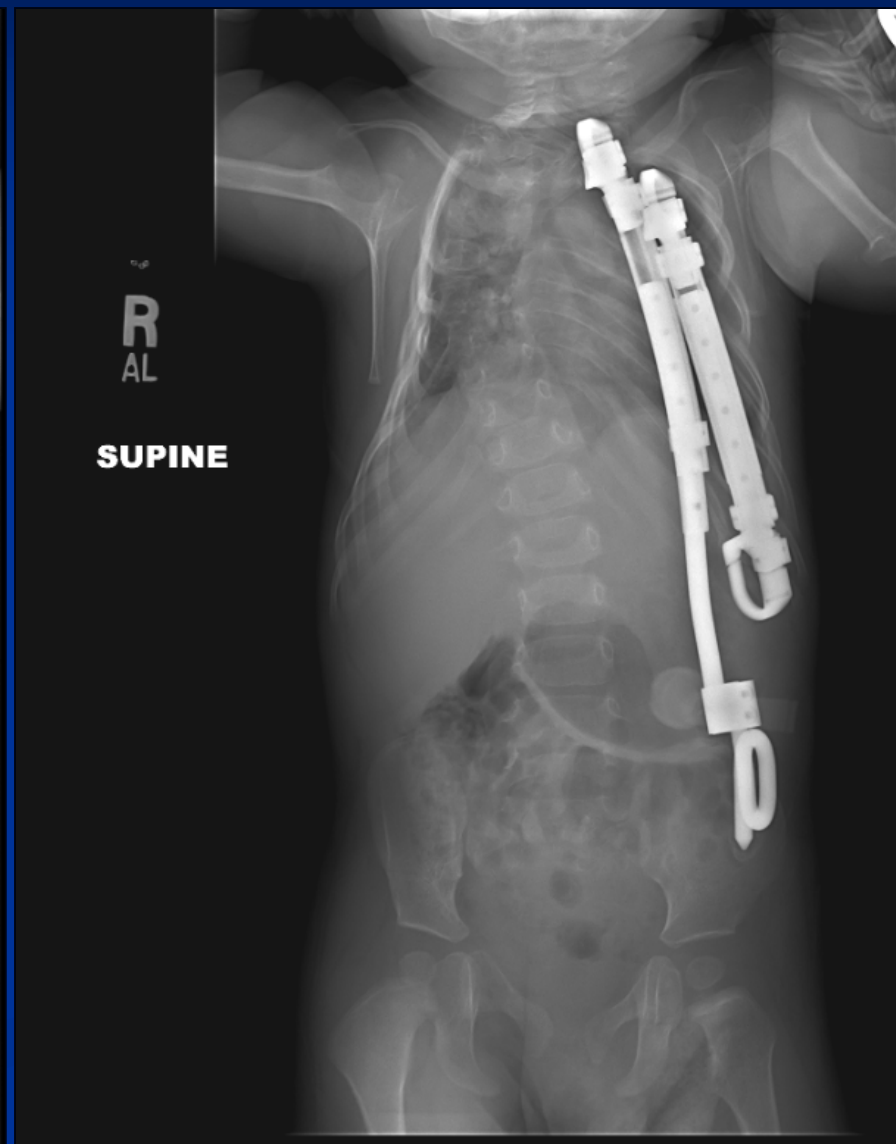


- SEMD- 2 1/2 yo
- Severe kyphoscoliosis
 - Previous posterior fusion at apex w/o immobilization ~ 1 yo
- Halo traction X 6 weeks



Different anchor choices have their merits/disadvantages

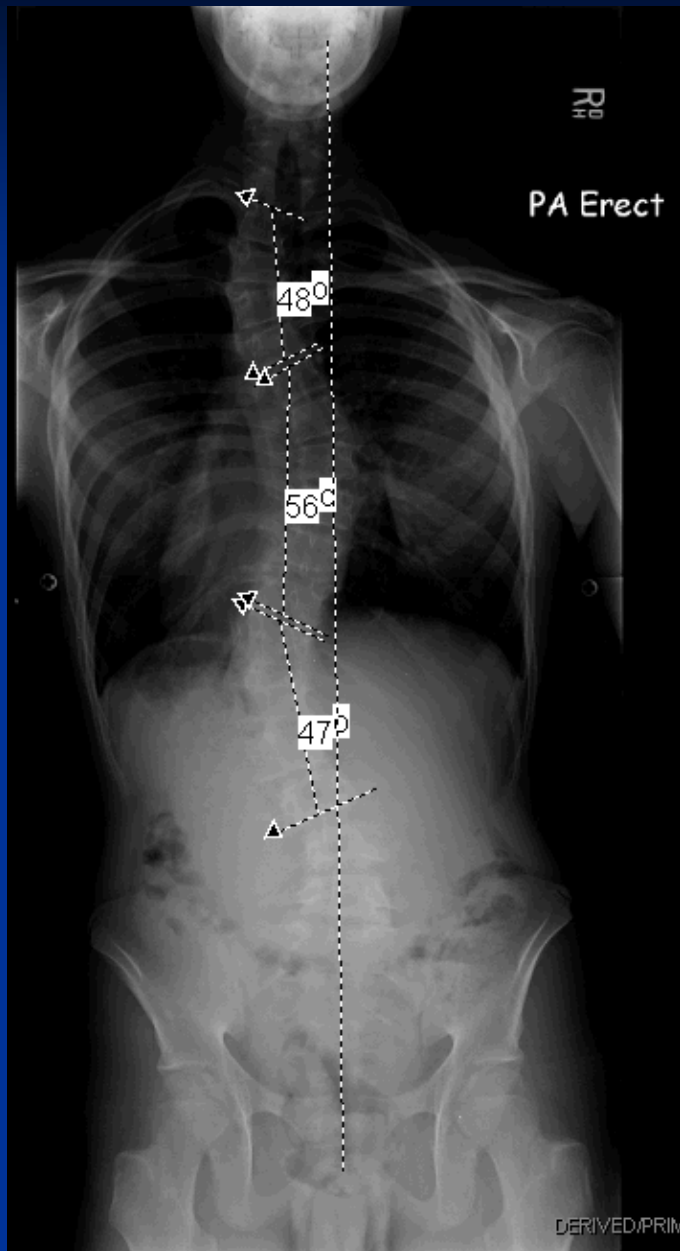
TIS

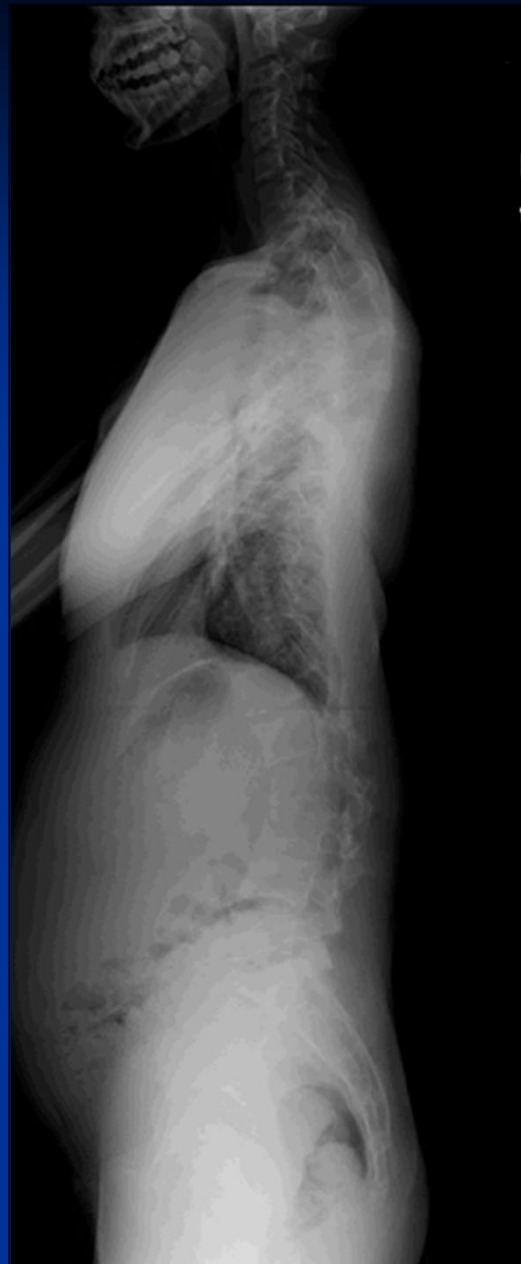
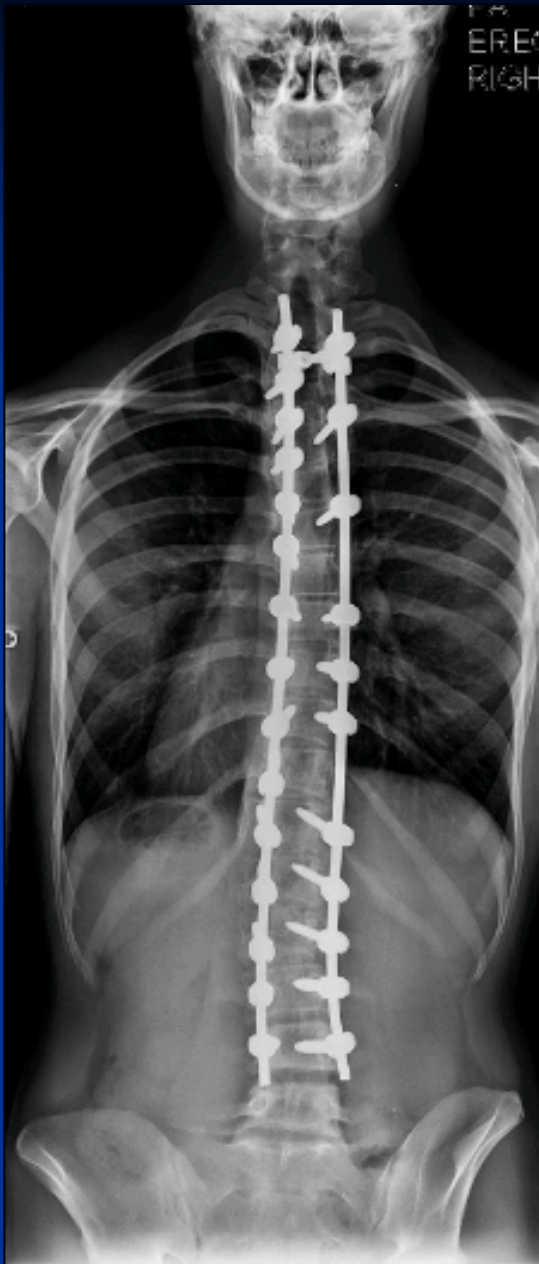


Syndromes associated with sagittal plane reversal

- Abnormal ligamentous laxity, hypotonia
- Possible vascular fragility
- Marfan
- Beals
- Ehlers-Danlos
- Larsen (no vascular fragility)

Courtesy of Charles Johnston MD





CTD : “Can’t go wrong if you fuse long”

Syndromes associated with osteopenia + poor bone healing

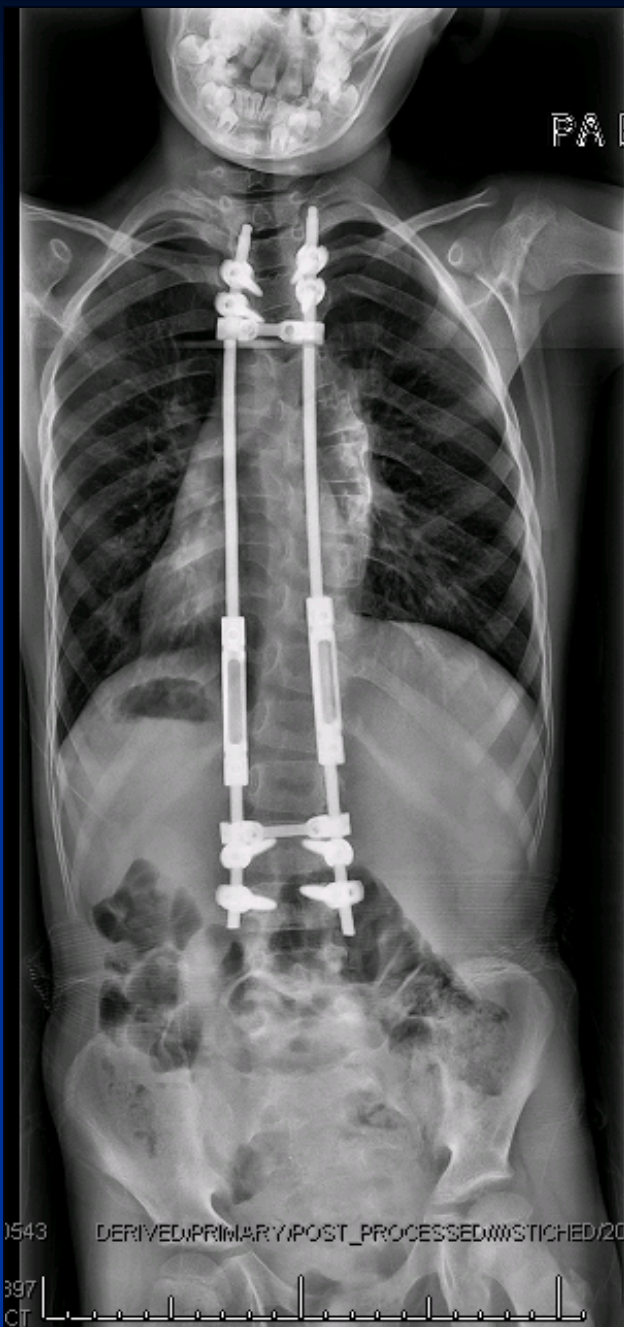
- Neurofibromatosis – dysplastic
- Syndromes in category II
- Cerebro-costo-mandibular
- Chondrodysplasia punctata
- Osteogenesis imperfecta

Courtesy of Charles Johnston MD

NF-1



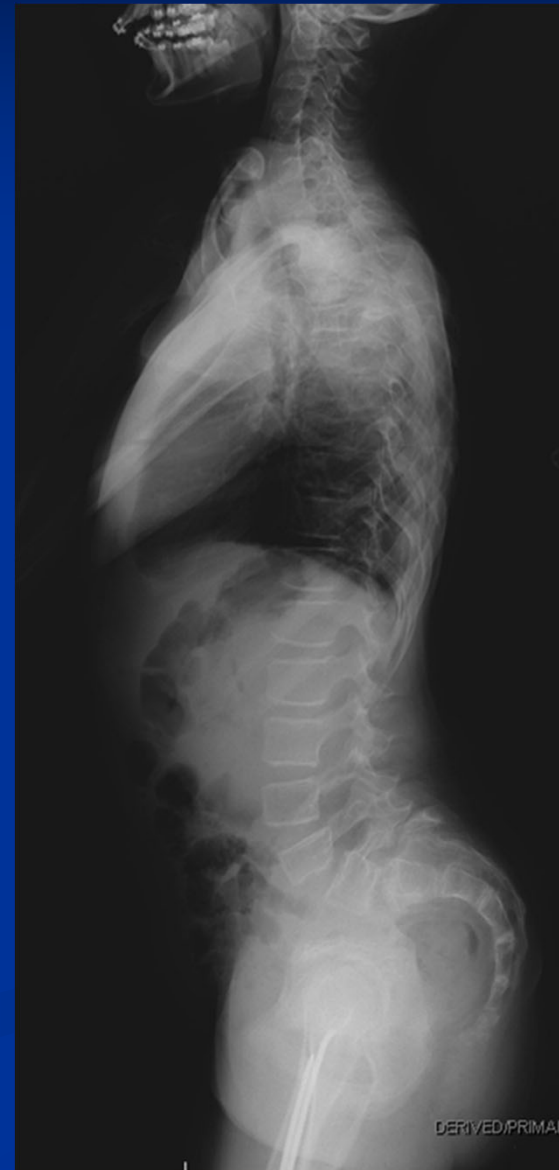
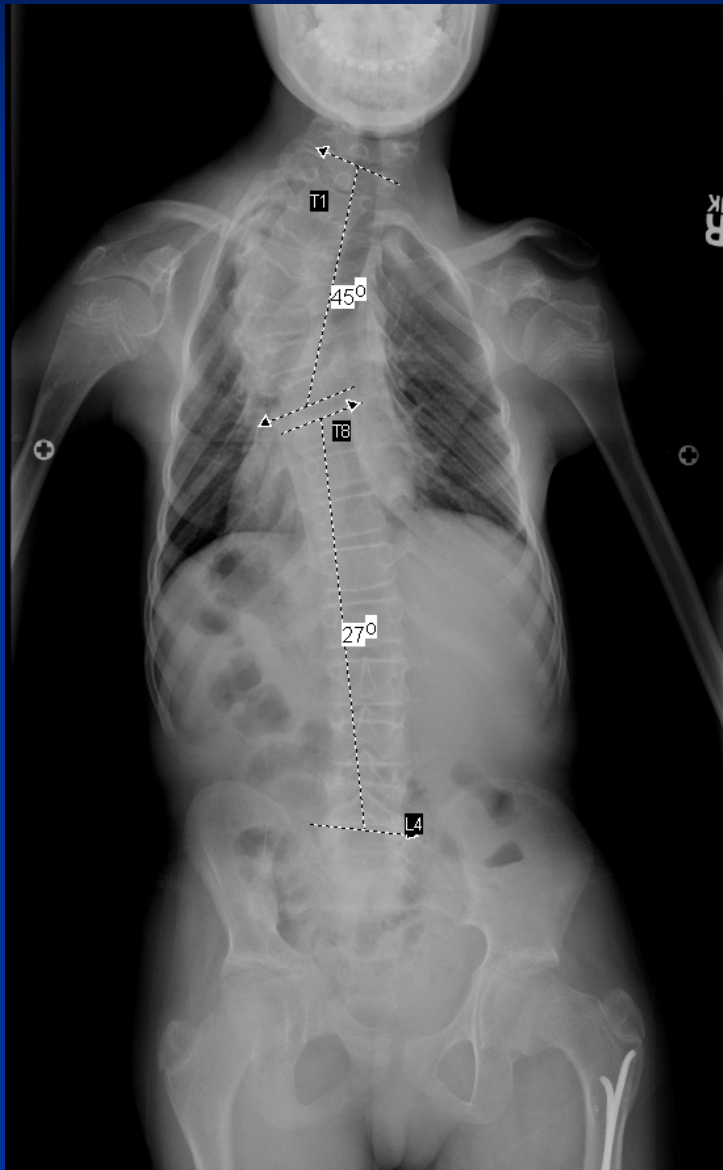


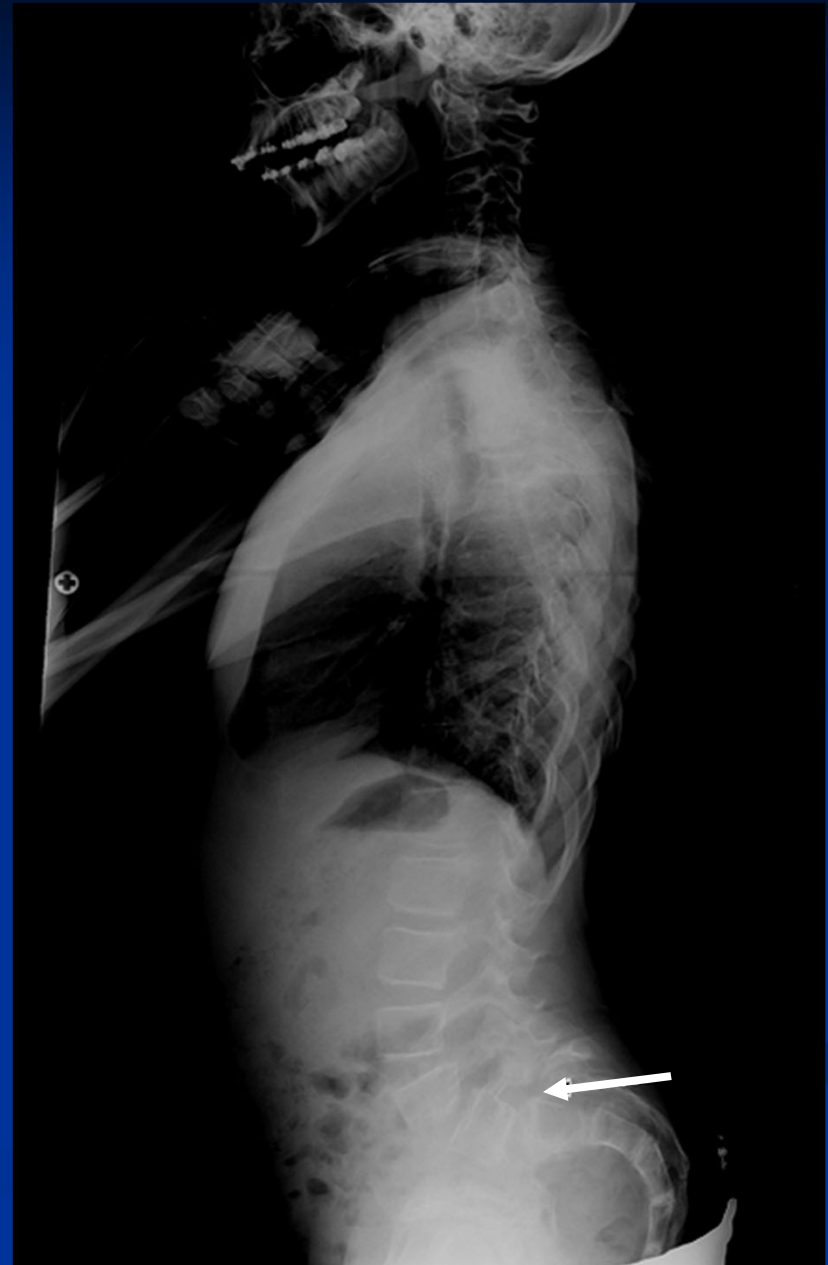
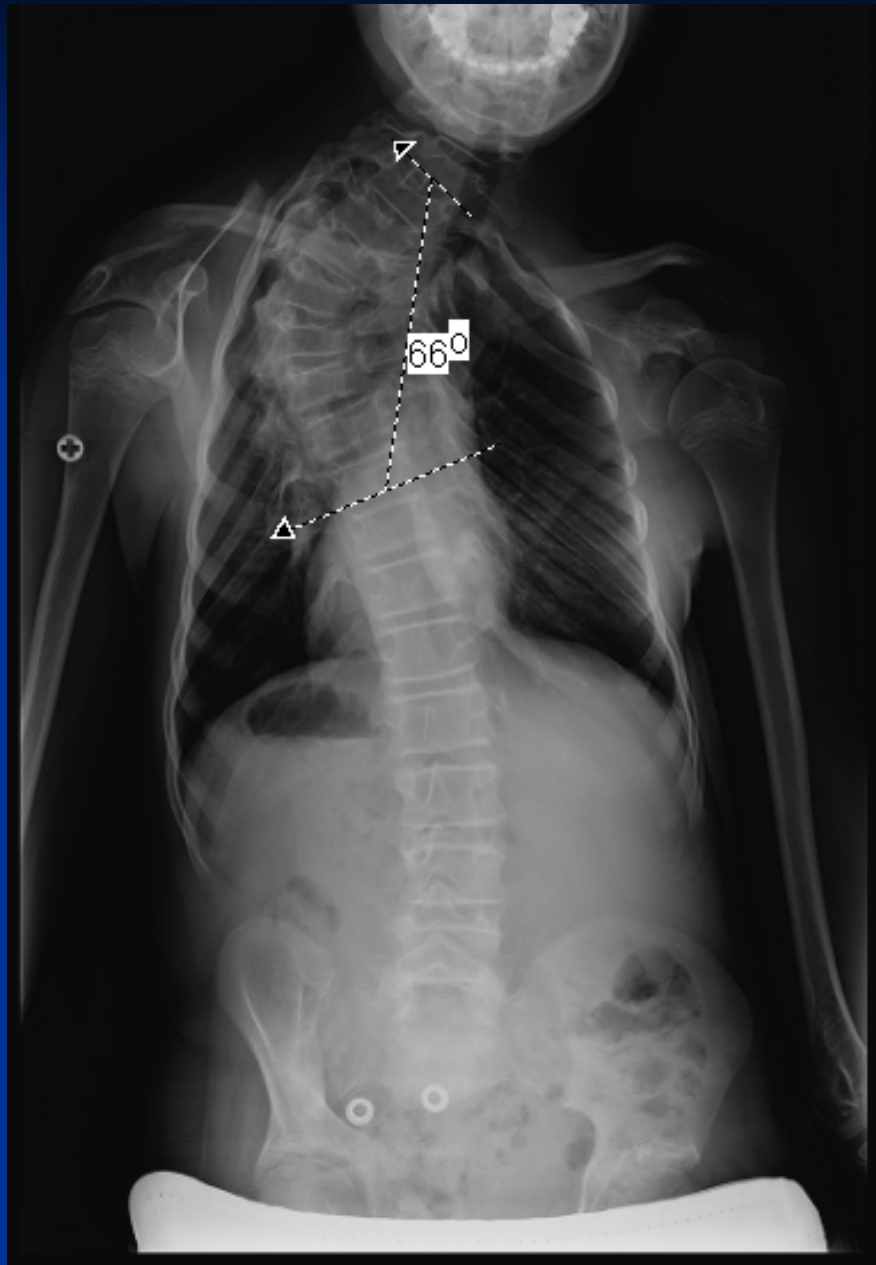


NF-1



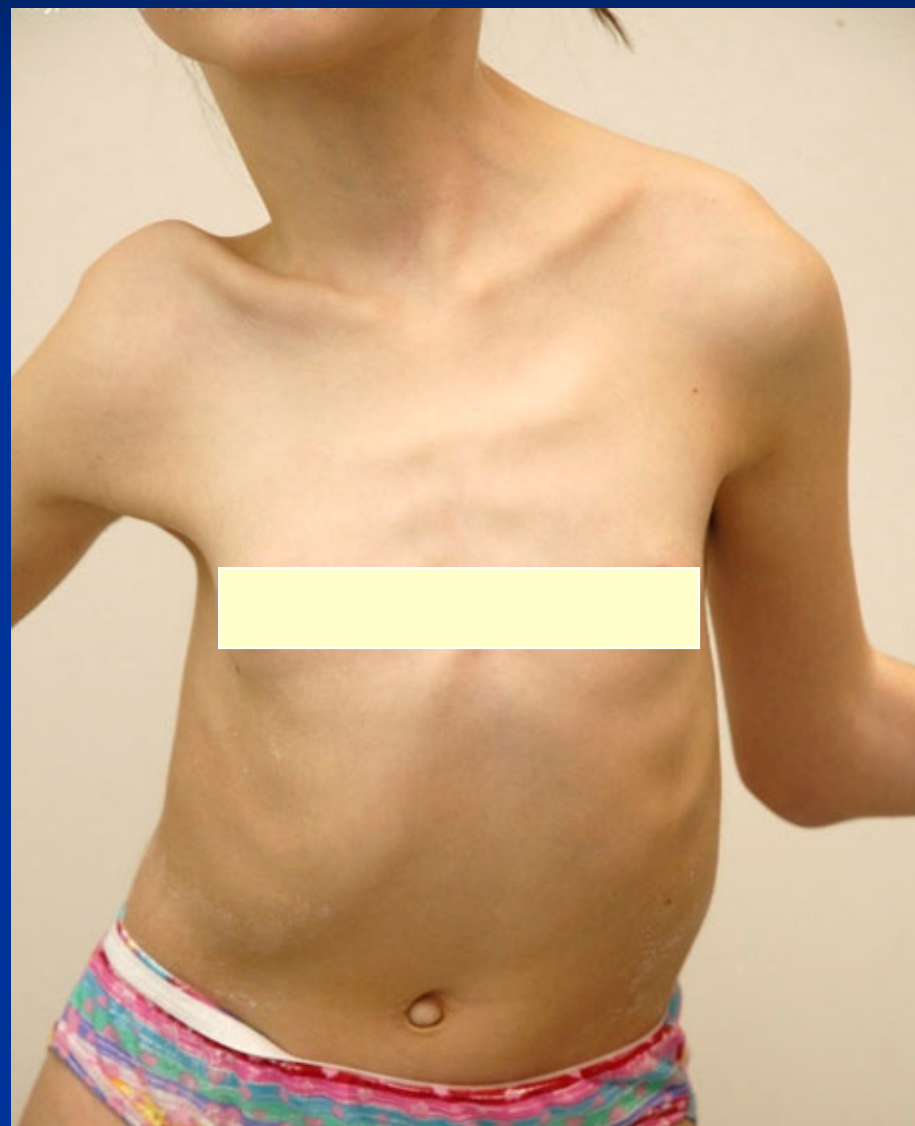
13 yo female Bruck's syndrome





- Very stiff deformity
- Scheduled for surgery
- Pamidronate therapy
- PFT's: FVC 45% predicted
- Femur fracture : elastic nails

Clinical Photos



Clinical Photos



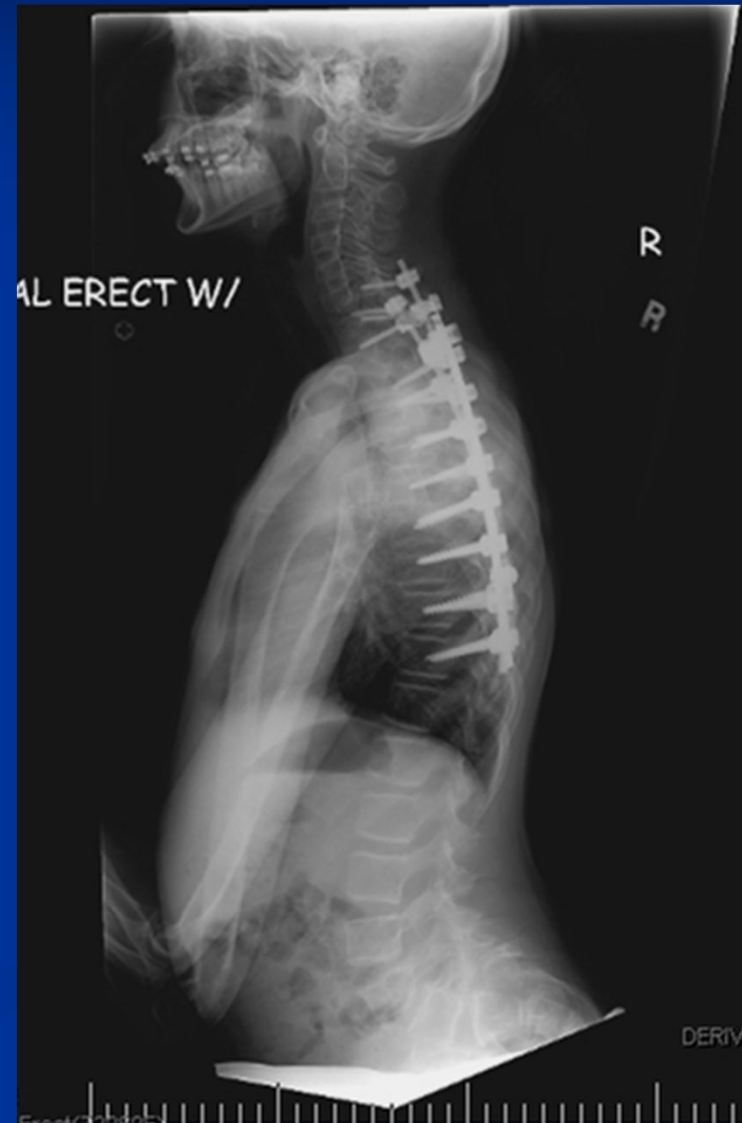
Traction film after posterior release

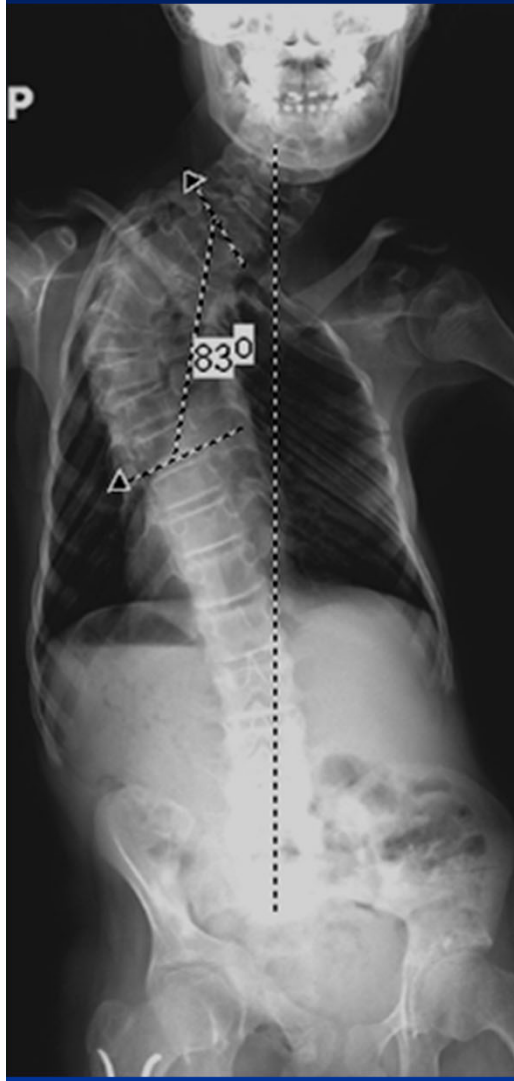


Halo gravity traction



Post op



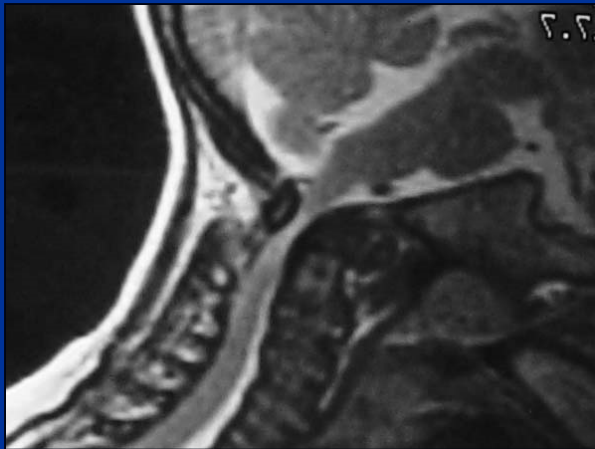
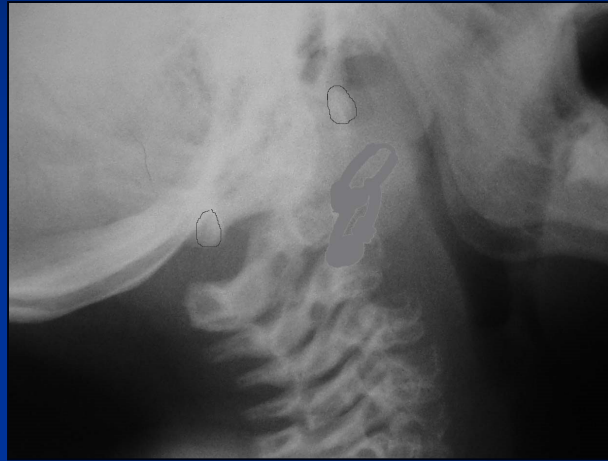
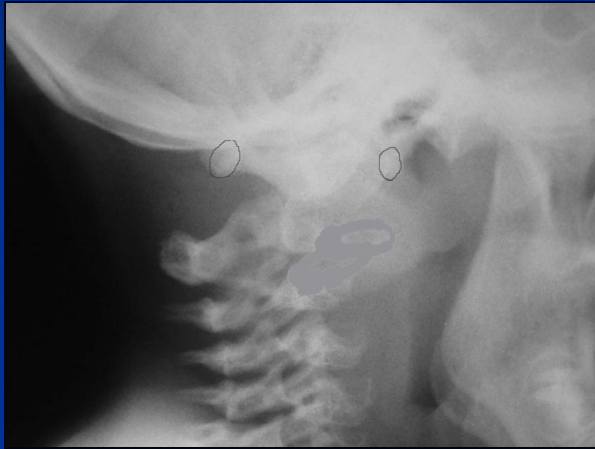


Syndromes associated with cervical instability/stenosis

- Achondroplasia
- Spondylo-epiphyseal dysplasias
- MPS storage diseases
- Jeune
- Larsen
- Diastrophic dysplasia (cervical kyphosis may resolve)
- Chondrodysplasia punctata
- VATER/ Klippel-Feil

C 1-2 Instability- Morquio Syndrome

Irreducible subluxation, Decompression and O-C2 fusion



Courtesy of Will Mackenzie MD

Cervical Kyphosis

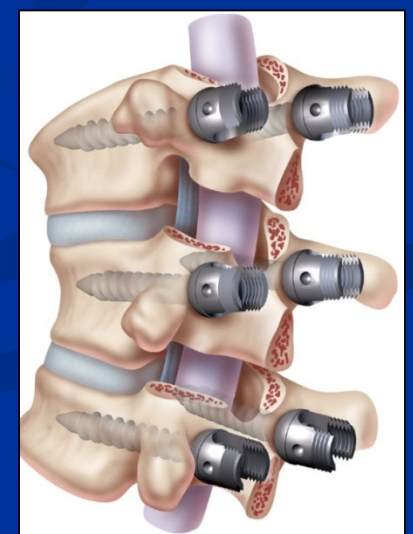
Decompression/Fusion



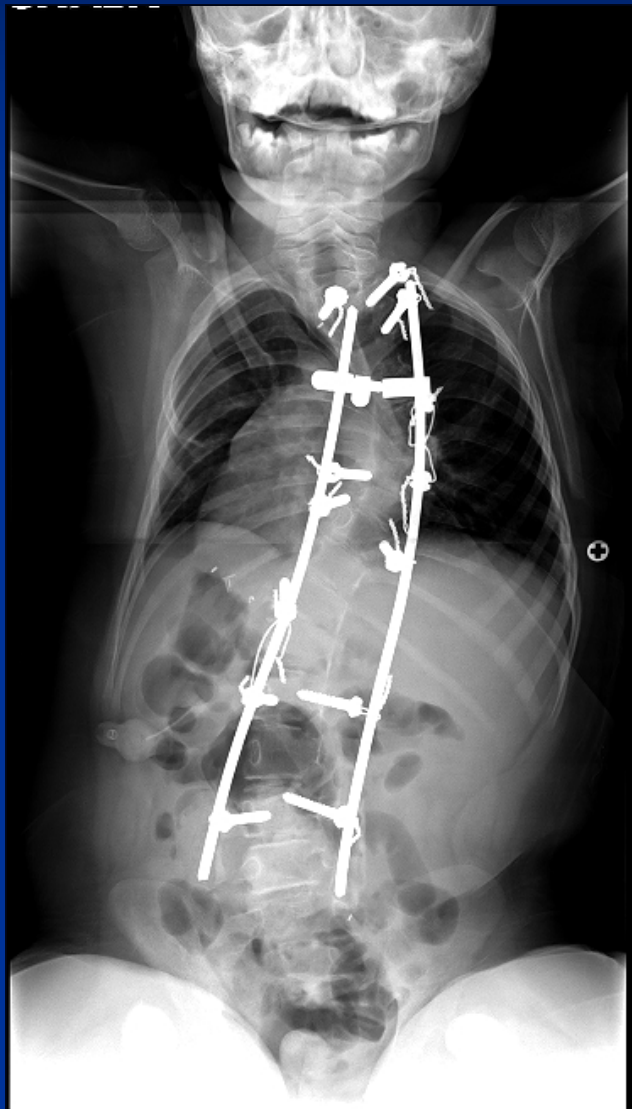
The Ponte Osteotomy

Correction of Junctional Issues

- Sagittal plane correction over a few levels
- Much smaller version of a PSO
- Proximal junctional kyphosis
- Distal junctional kyphosis



9 yo ♀ Trisomy 9



9 yo ♀ Trisomy 9



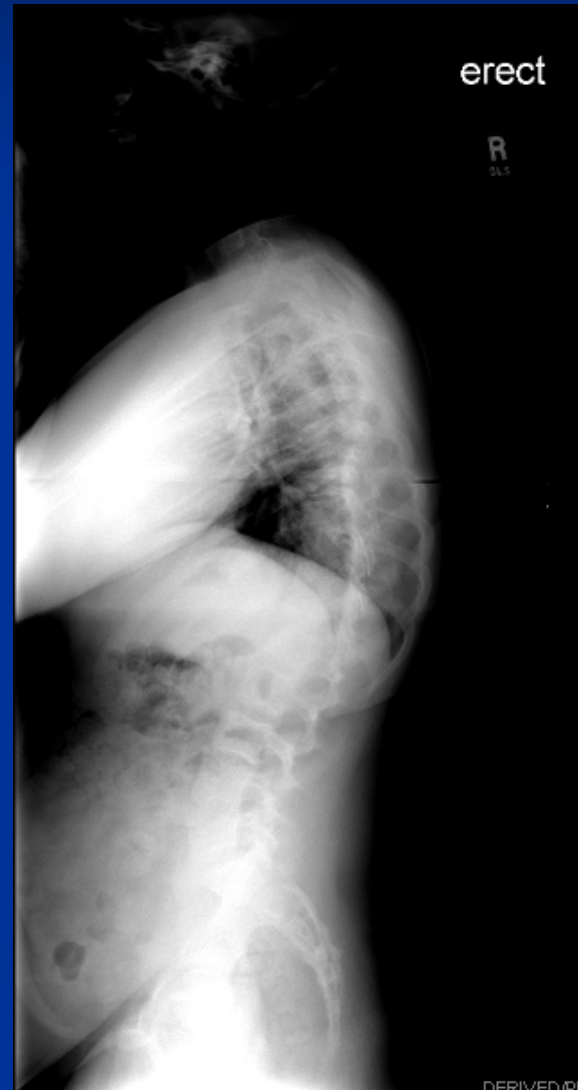
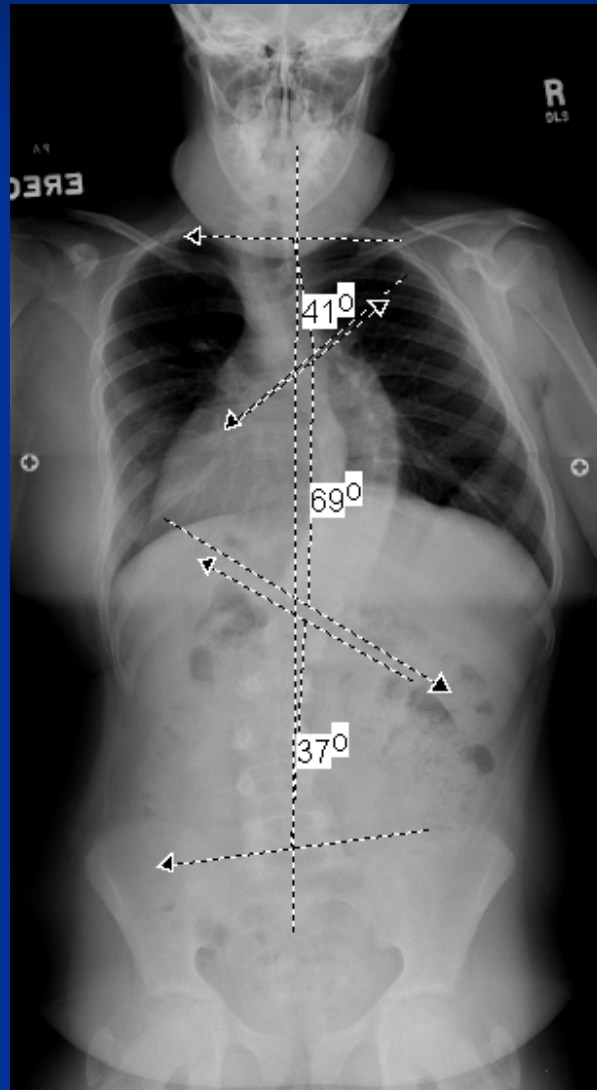
9 yo ♀ Trisomy 9



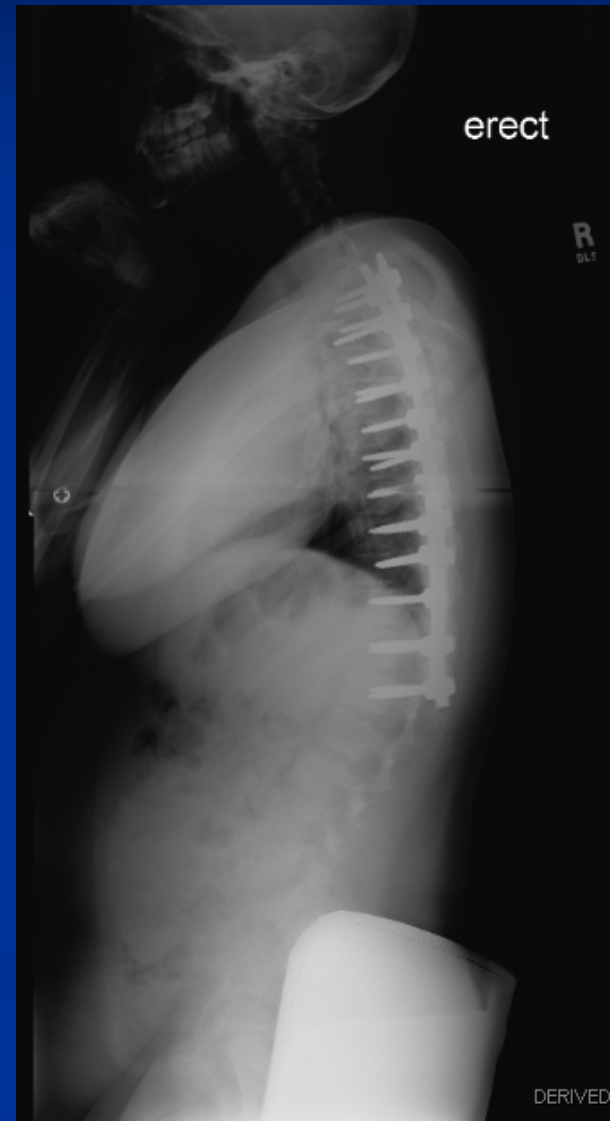
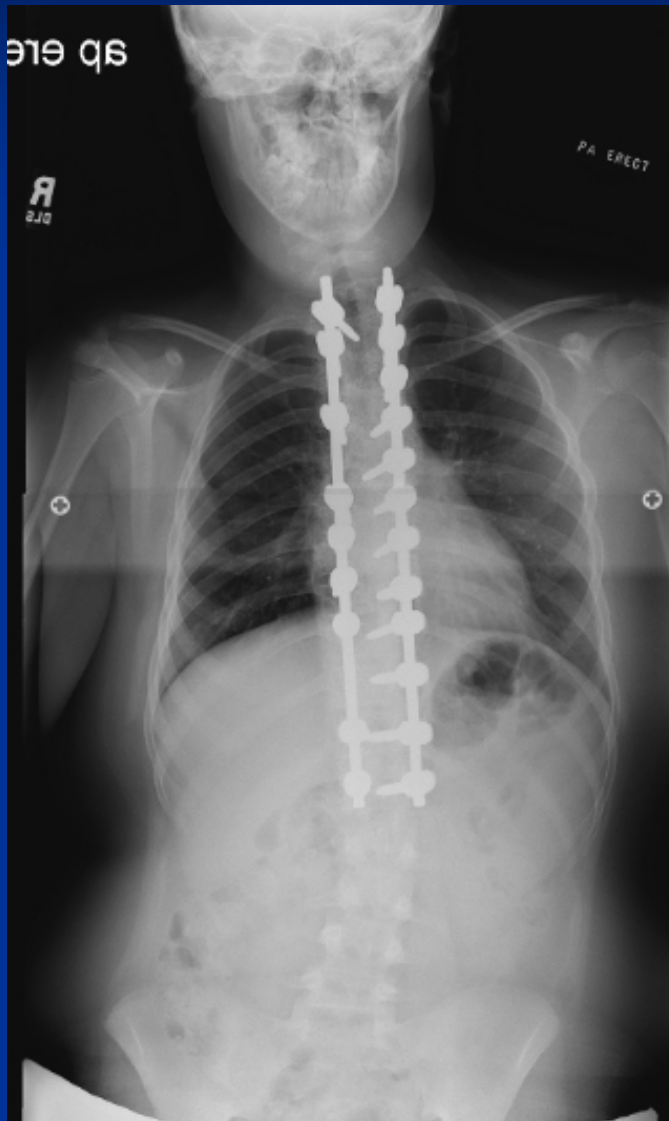
12 mo sitting films



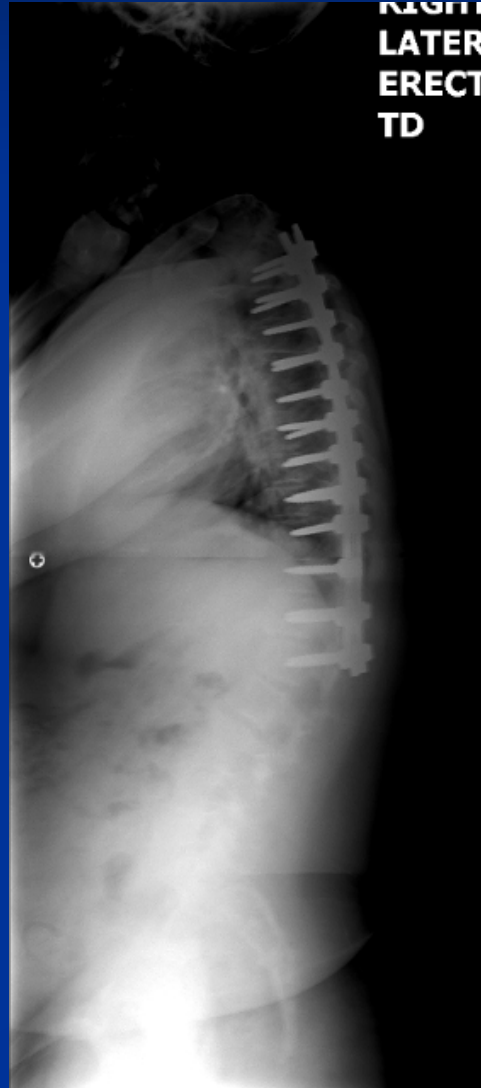
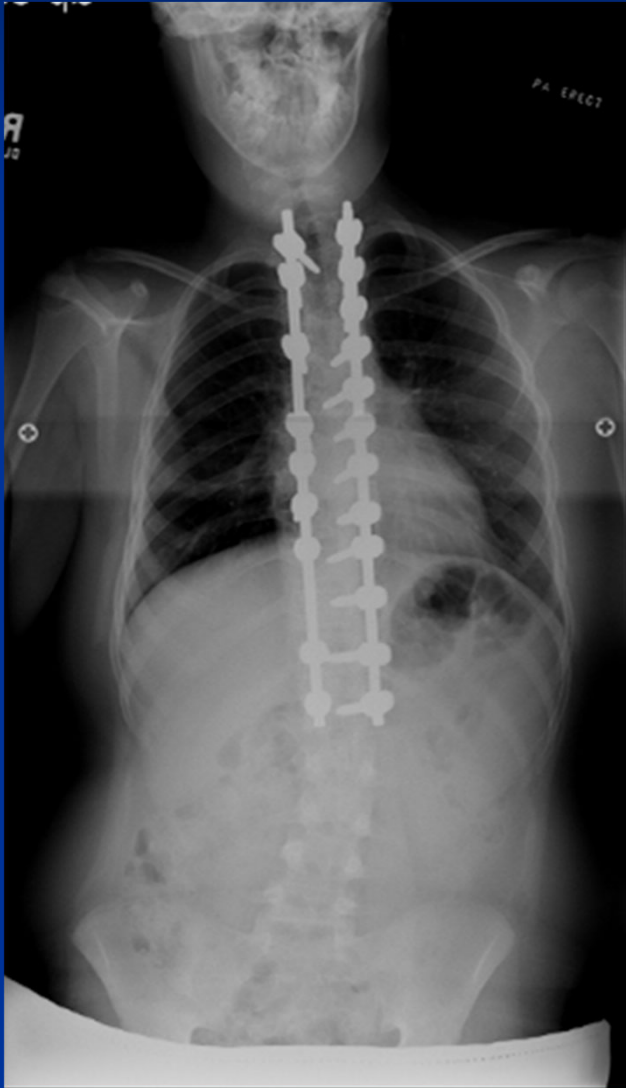
15 yo ♂ AIS Lenke 2BN



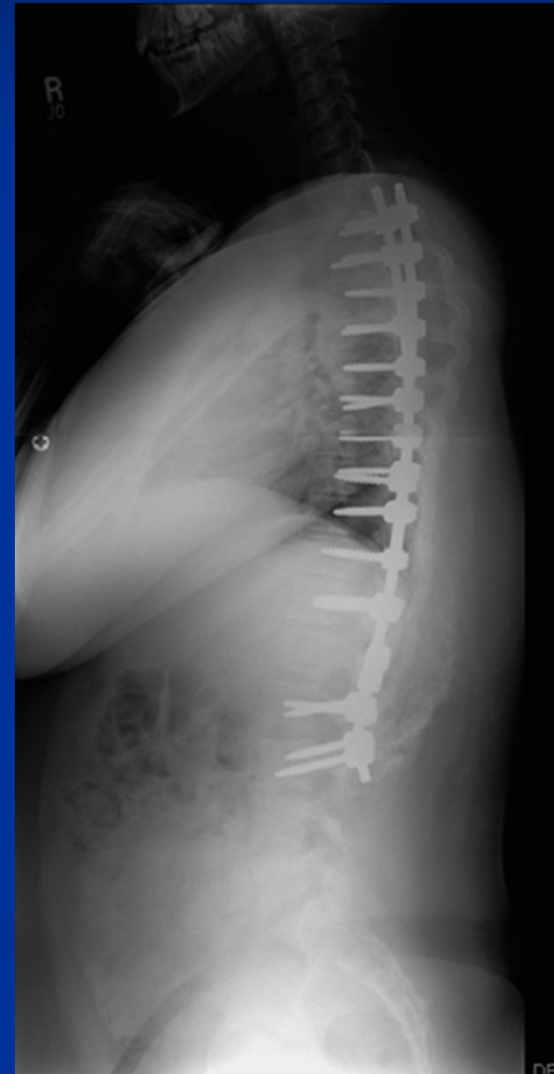
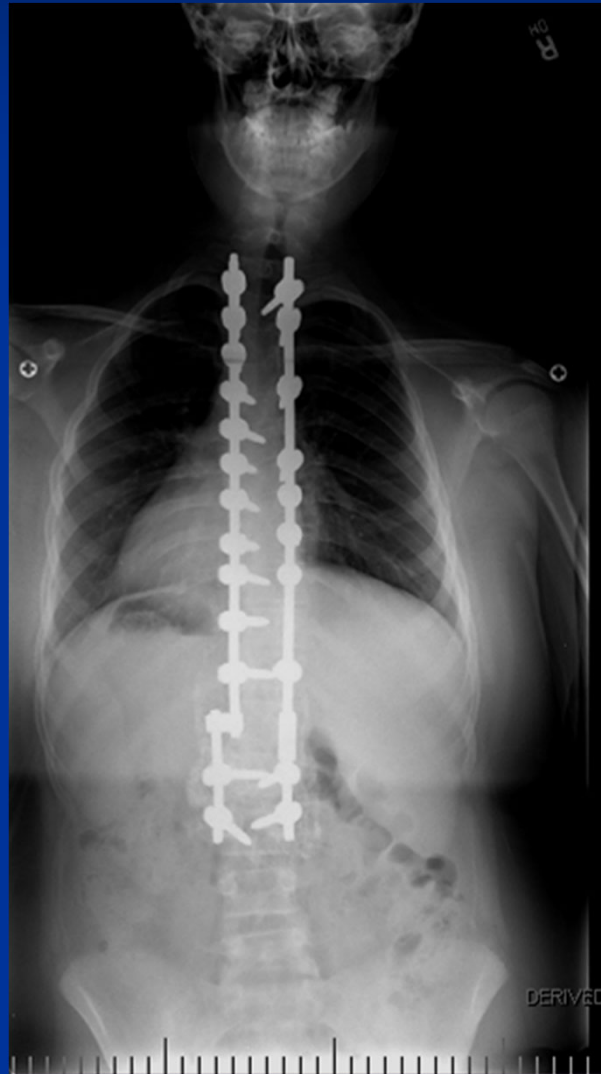
6 mo postop



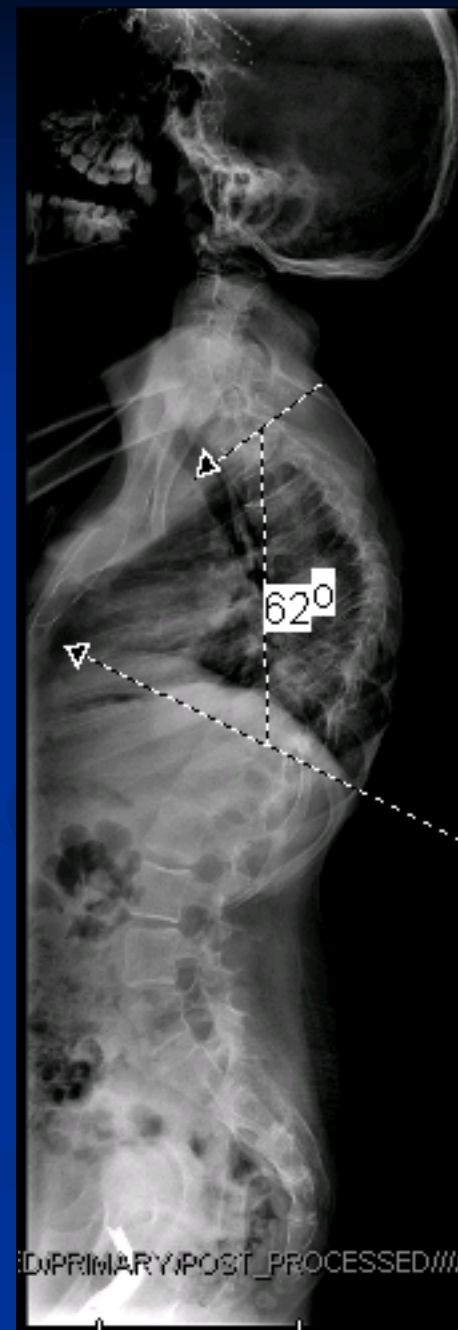
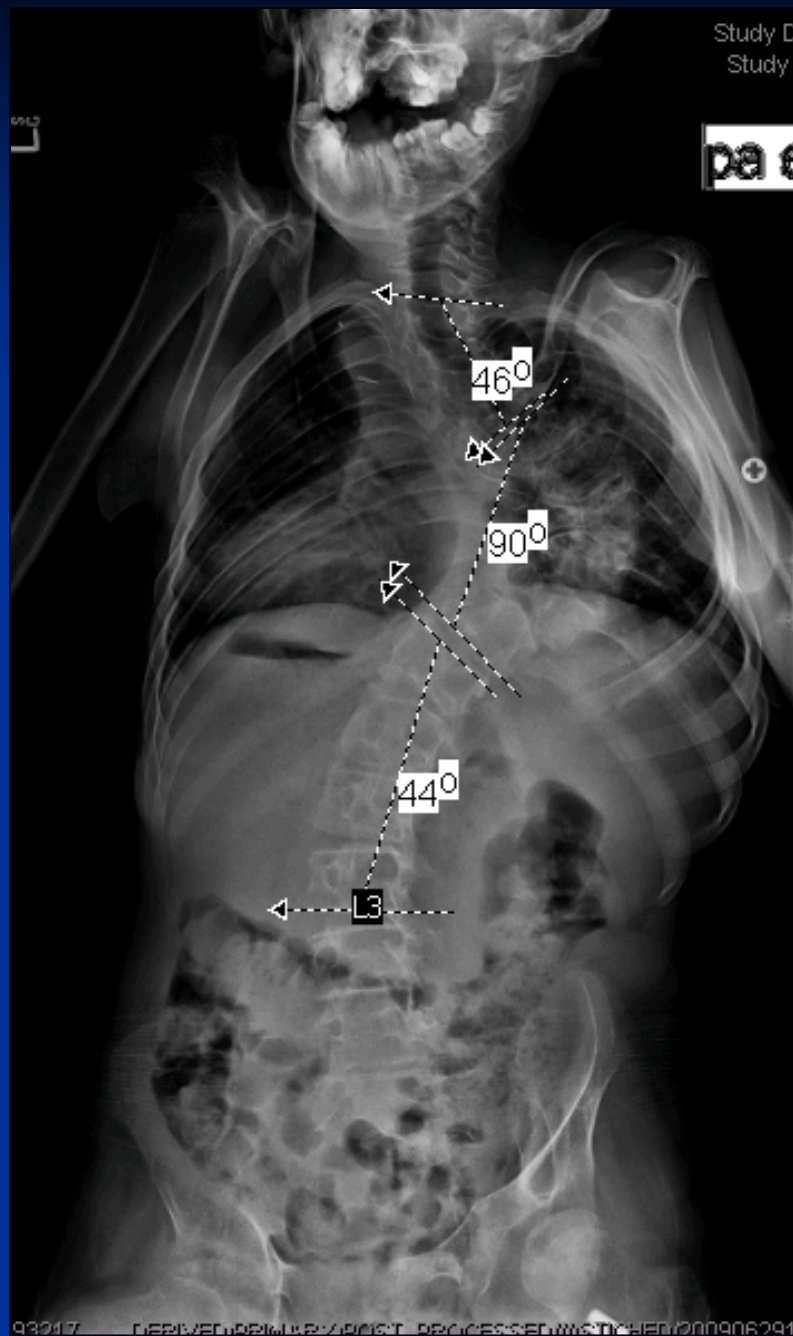
18 mo postop – progressive DJK



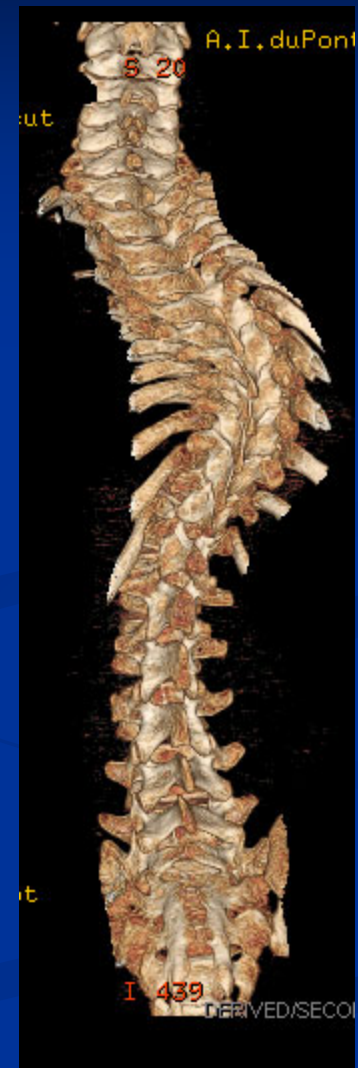
Sagittal Plane Reconstruction





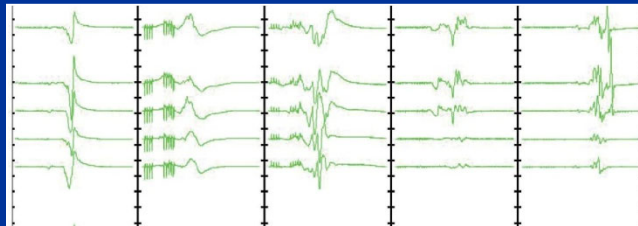


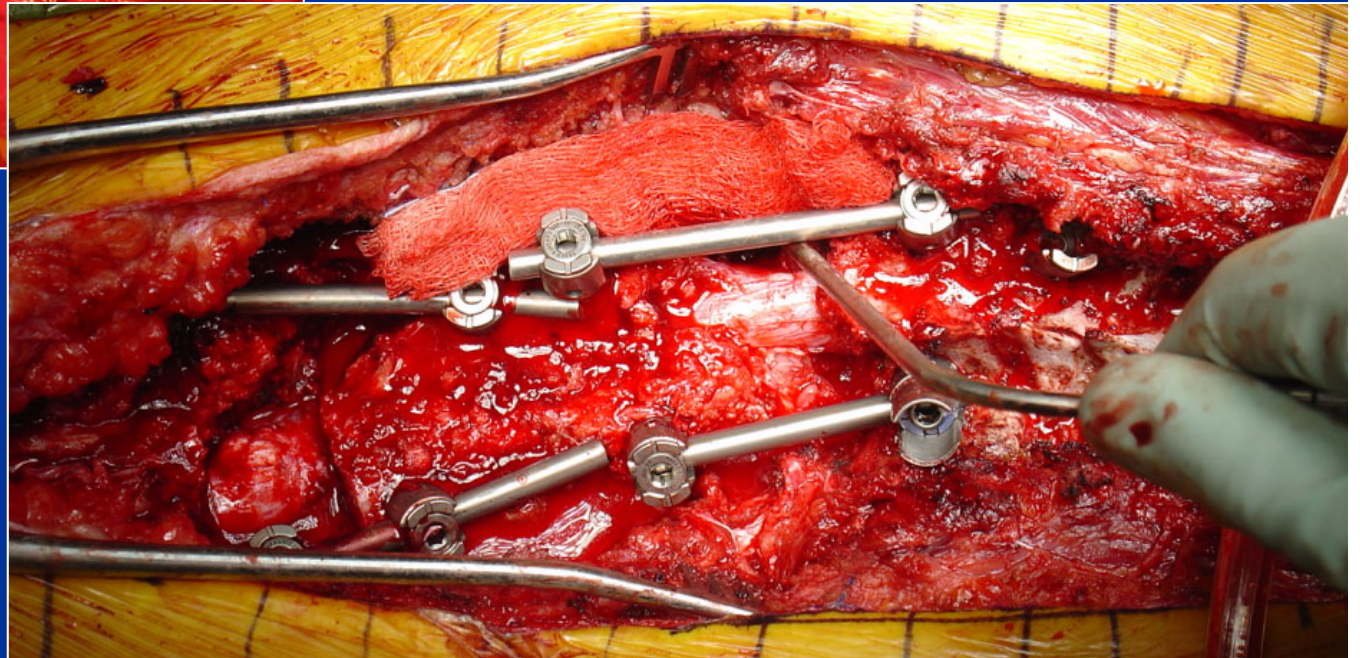
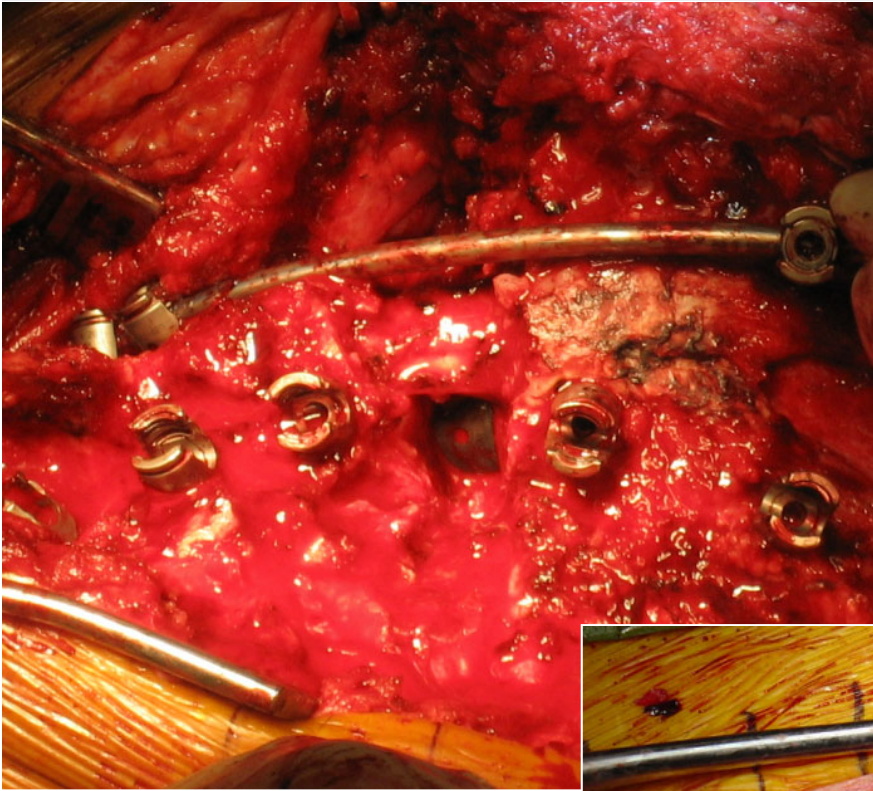




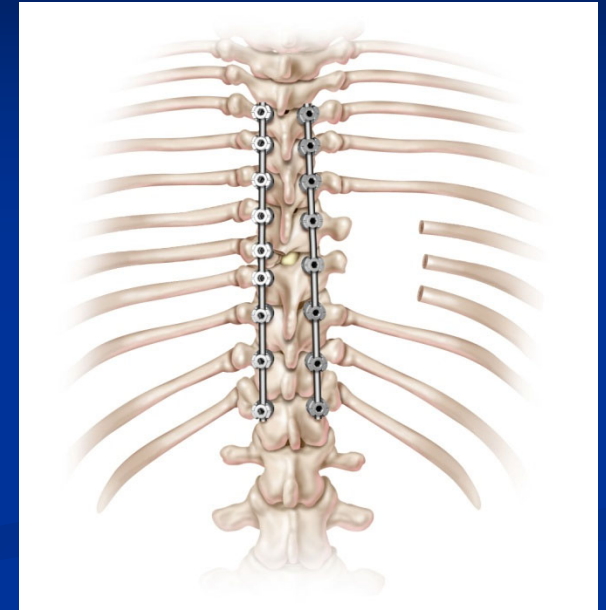
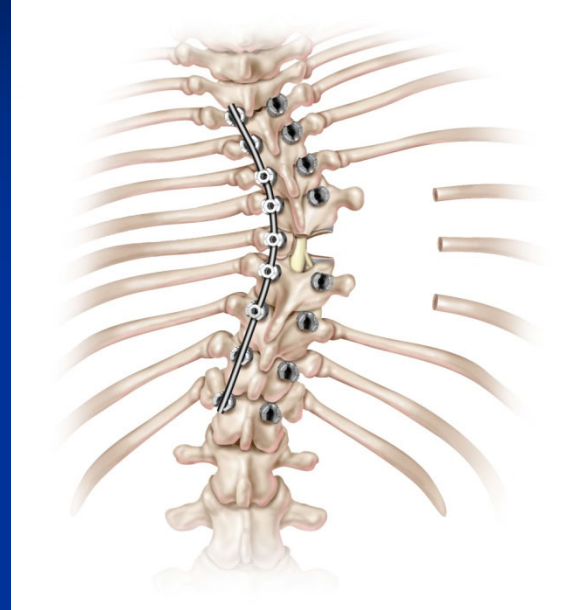
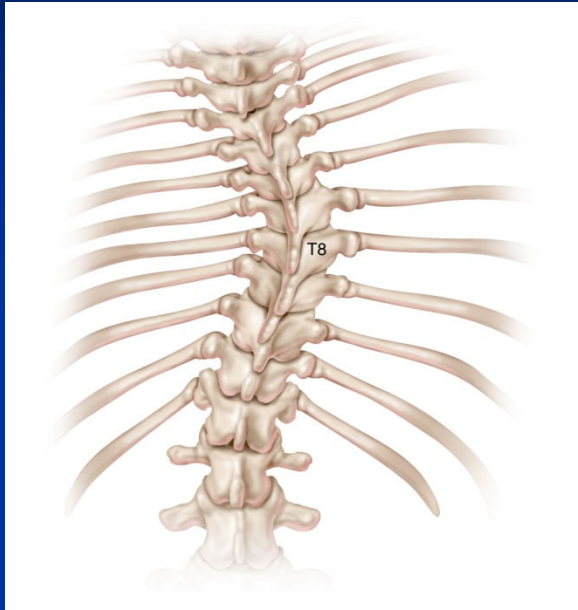
Spinal Osteotomy / Vertebrectomy

- Posterior Vertebral Column Resection
 - Avoids anterior procedure, less OR time
 - Kyphoscoliosis – deformity is presented posteriorly
 - Shortens spine, slow correction
 - Control vertebral column with pedicle screws and working rods (avoid translation/subluxation)
 - Anteriorly positioned cage when necessary
 - Good TcMEP monitoring mandatory





Asymmetric Thoracic VCR for Deformity



Theory: Leaving the concave pedicle with a screw provides a fulcrum for closure of VCR with large coronal deformity, preventing instability during surgery and excessive potential shortening of canal. Obviates need for cage



Summary

- Complication rate is high
- Careful preop workup: studies, consultants
- Be prepared for challenges in the OR
 - Bleeding, stiff curve, osteopenia
 - Antifibrinolytics, make spine flexible, secure fixation
- Growth sparing techniques useful in young pts
- Stormy postoperative course
- Rewarding

There was a little syndromic girl who had a little curve
Right in the middle of her back;
When she was good, she was very, very good,
And when she was bad... she was horrid.

Thank You



Nemours

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