Complications Related to Specific Diagnoses Syndromes, Dysplasia

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Overview

- Common Themes
- Marfan Syndrome
- NF1
- Skeletal Dysplasia
- NM
- Complications

Approach to Syndromes

-and systemic diagnoses

- Idiopathic Deformity:
  - Established patterns
  - Proven techniques
- Syndromes have opposite properties
  - Unique problems
  - Step back
- Get help!!

Resources

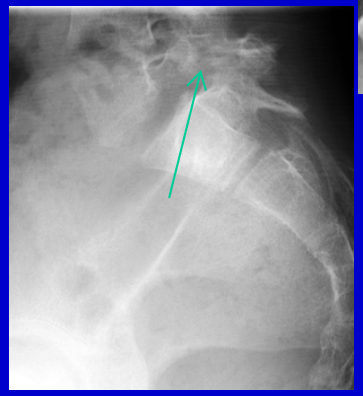
- OMIM (Online Mendelian Inheritance in Man)
  - NLM feature
  - 18,000 entries
  - Searchable by feature (i.e., pectus) + combinations (pectus AND scoliosis AND vertical talus)

- Pub Med: Search individual diagnosis if known

- Your friendly local geneticist
  - Dx & Management

Common Themes: Examine Entire Spine (C,T,L)

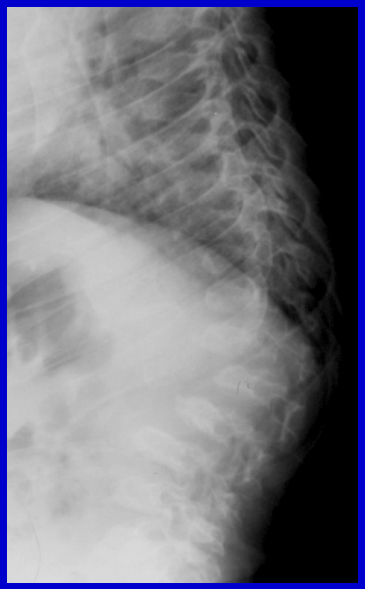
- Categories of Spinal Problems
  - Instability (C1-2, T12-L1)
  - Deformity
    - Kyphosis (C, T, L)
    - Scoliosis
  - Stenosis



- Apply to Cervical, thoracic, lumbar

Common Themes - General

- Casting, Bracing rarely helps deformity
  - Marfan syndrome 17% efficacy
- Exceptions
- Use only in small, flexible curves
  - If not a burden



Common Spinal Themes

- Failure of fixation
- Adding-on

Common Themes- preop

- Consider Spine Deformity in context of total Disability
  - Hurler syndrome: life expectancy < 20 yrs
  - Dysraphism: Infection, pressure sores, neuro deficit question benefit
  - Help FAMILY make cost-benefit analysis
    - Each family sees it differently

Common Themes- preop MRI, CT

- Usually high yield in syndromes
  - Dural ectasia
  - Cord size, location
  - Stenosis
- When?
  - Preop or if findings dictate

Common Themes
-Medical Comorbidities

- Pulmonary
  - Sleep apnea, obstruction
  - Pulmonary consult
  - Sleep study
- Cardiac problems in Morquio, Marfan, OI, others
  - Echo
  - Coumadin, other meds

Common Themes

- ICU if any question
- Involve pediatrician, hospitalist
  - From the start
- Longer hospital stays
- May need trach, GT

Medical Co-morbidities

- More frequent missed lengthenings
- Schedule yearly ?

Common Themes- technical

Narrow Pedicles

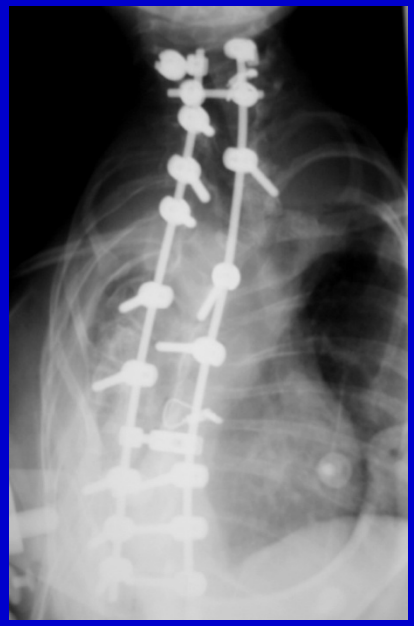
- Obtain good imaging in advance
- Coned AP plain films
- CT scans
- Traction films

Difficult pedicles

- Fix more levels
- Outside-in technique?
- Range of sizes
  - cervical 3.5, 4.0mm with transition rods
  - “Pediatric” 4.35, 4.5, 5mm

Hooks!

- Use when screws fail
- Also use rib as anchor (Skaggs)



Common Themes: technical

-Bone density in syndromes

- Decreased in Marfan, neuromuscular, OI
- Sclerotic/brittle in NF1

Marfan Infantile Kyphoscoliosis

- Large curves at early age
- Most Double major
- 1/3 have significant kyphosis

Marfan Syndrome

- Narrow Pedicles
- Expanded dura
- Osteopenia

Early Growing Strategies

- Luque Trolley
  - Spontaneous fusions
  - Crankshaft
- Moe technique
  - Hook cut-out
  - Junctional kyphosis

Modern Marfan GR Cohort

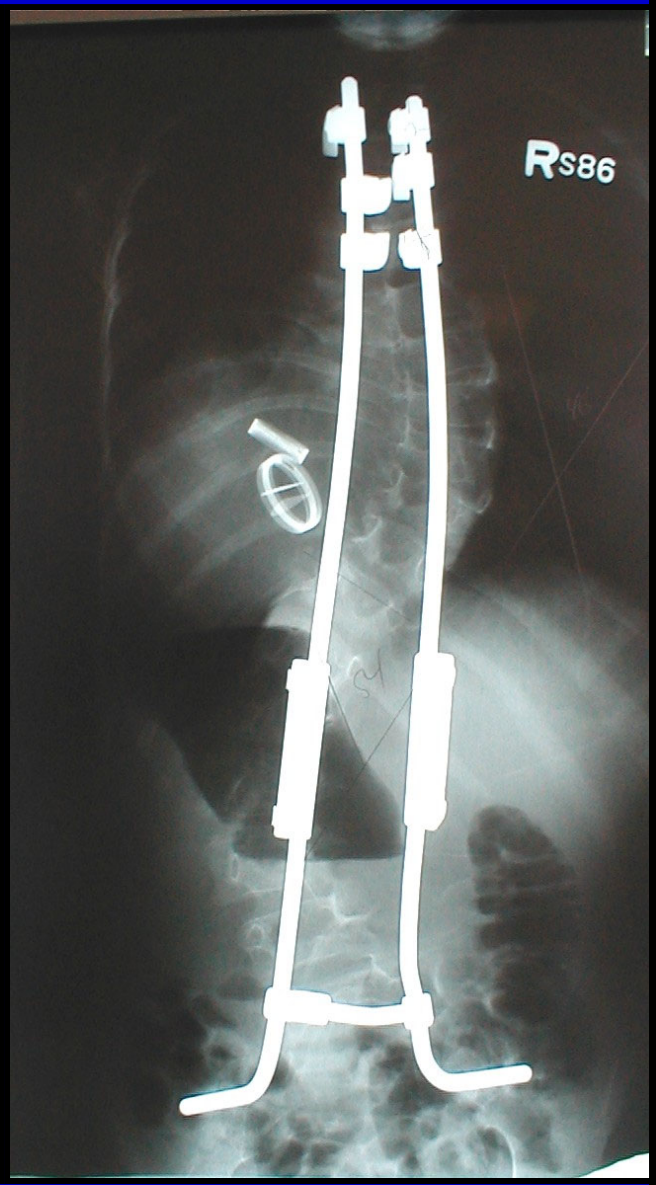
- 9 patients
  - Dx < 3 y.o.
  - Surgery 4.5 yrs (2.5-8)
- Mean curve preop: 80^{o}
- Kyphosis preop
  - 4 with excessive kyphosis, mean 78^{o}
  - 5 with normal kyphosis, mean 22^{o}

Techniques

- 2 techniques:
  - TL/L kyphosis: rod to pelvis
  - Normal kyphosis- typical growing rod

Final Fusion

- 3 patients
  - 46 months growing time
  - 9.5 years old
- Mean length T1-S1= 10.2 cm prior to fusion
- Mean Curve = 25^{o}
  - 73% correction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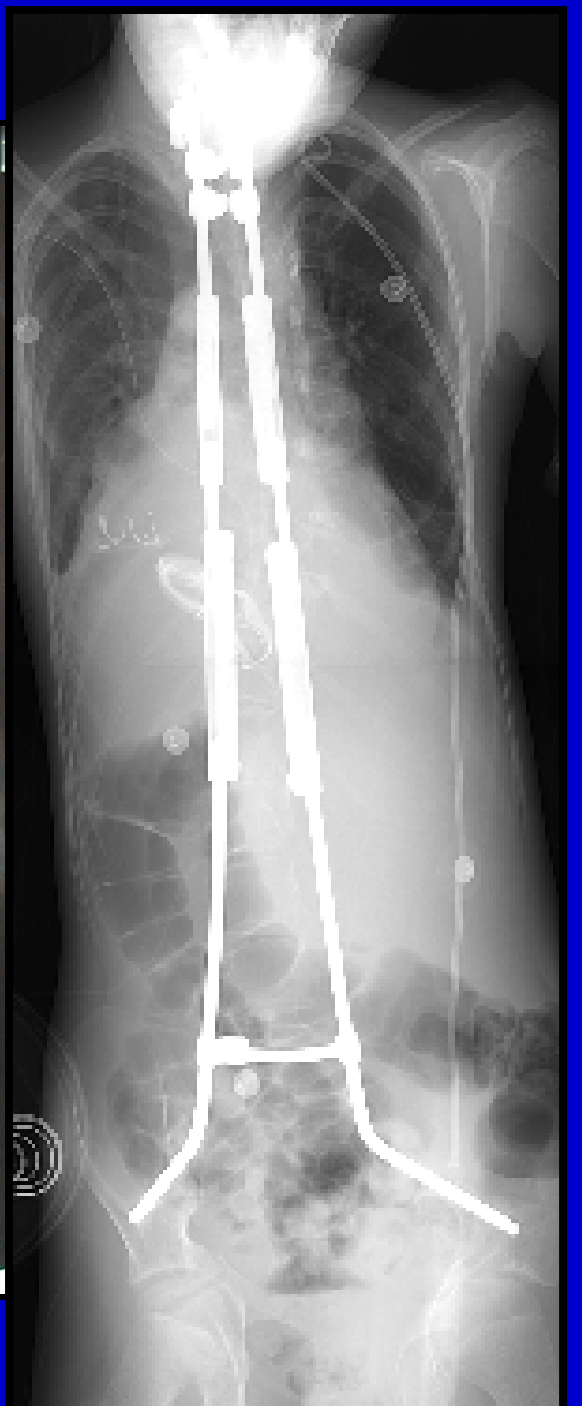
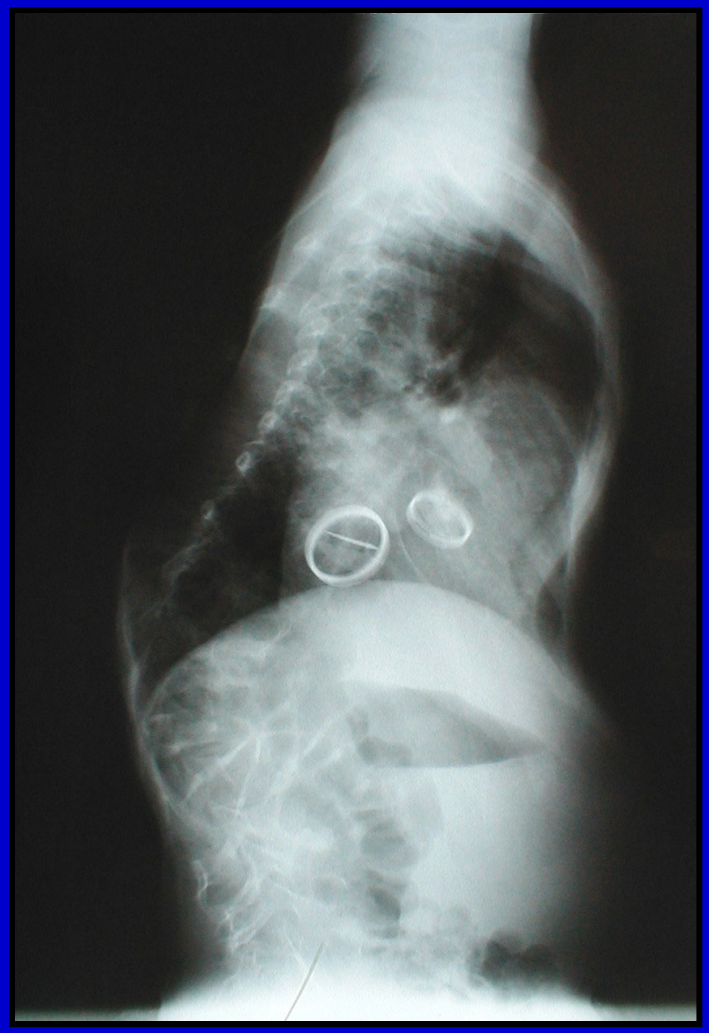
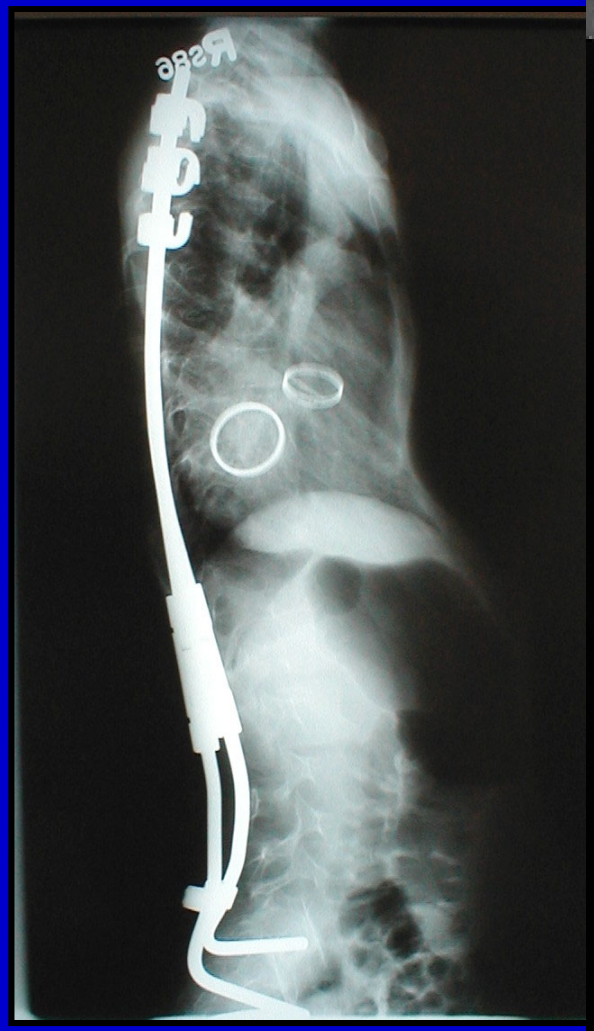
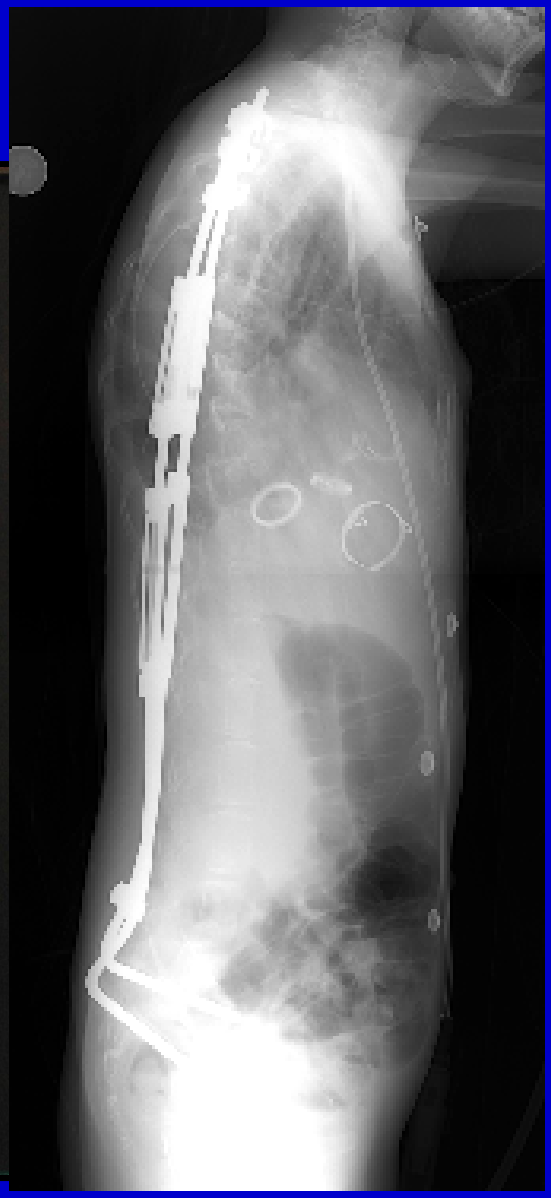


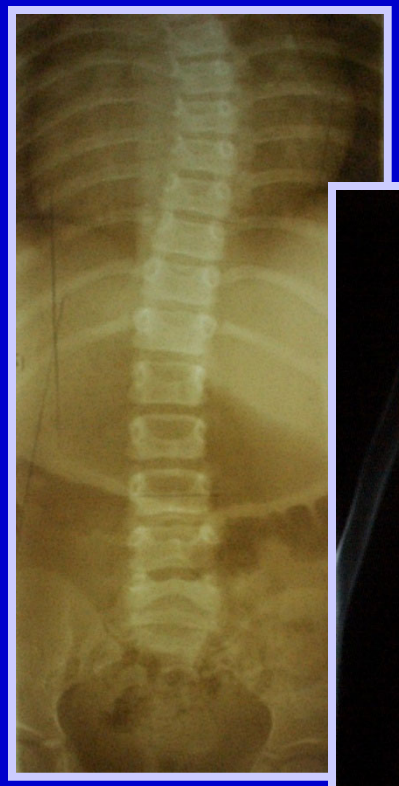




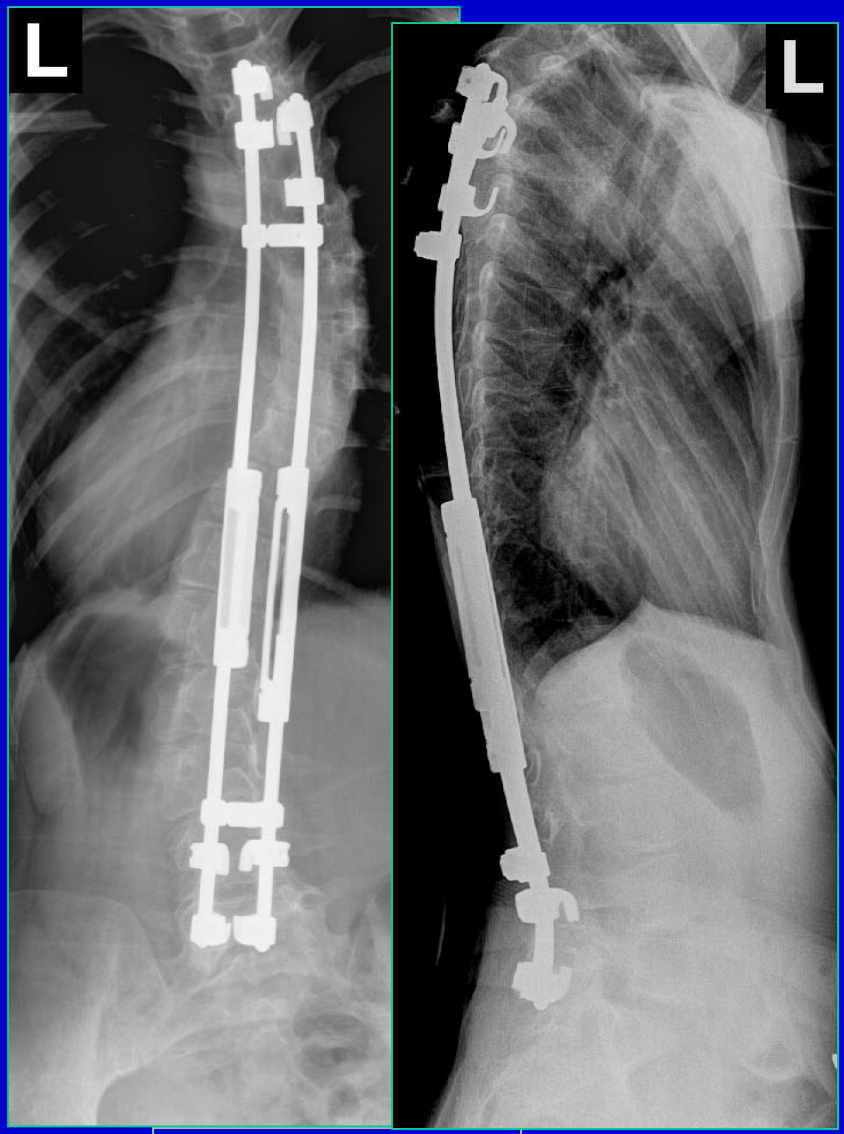


3+11 Years old with Marfan Syndrome

Initial



Pre-op



5 yr f/u

Complications

- 1 junctional kyphosis
- 1 pelvic rod backout (revised)
- 2 rod fractures
- 2 intra-operative dural leak (none postop)
- 1 died unrelated causes 3 months postop

CSF Leak

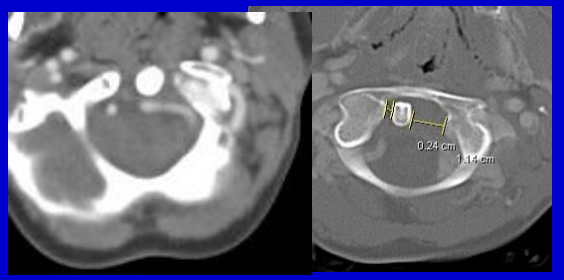
- OR Table in Trendelenburg
- Extreme care in bony dissection
  - May be completely eroded
- Rx:
  - Repair
  - Rest
  - Wrap

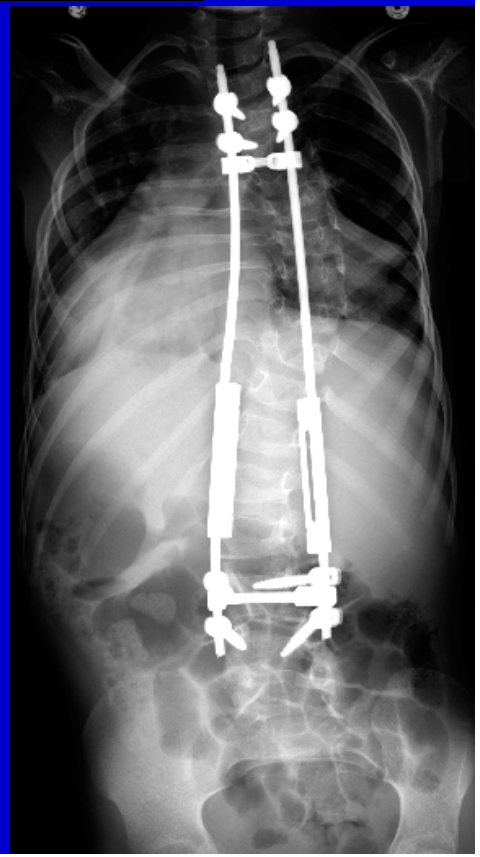
“New” Syndrome: Loeys-Dietz (LDS)

- TGF beta receptor abnormality
- Arterial tortuosity and aneurysms
- Hypertelorism
- Cleft palate and uvula

LDS: Orthopaedics

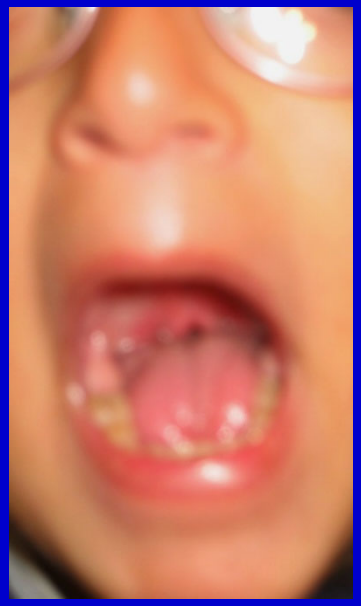
- Cervical spina bifida/ instability
- Scoliosis
- Clubfoot
- Ligamentous laxity





Loeys-Dietz Syndrome

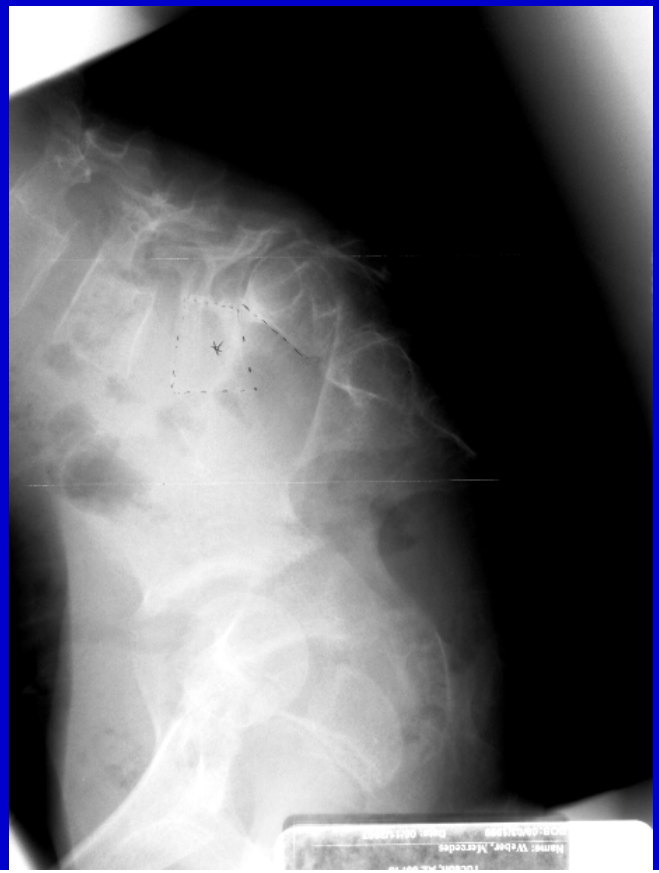
- Aneurysms treated aggressively
- Prediction: It will be frequently recognized
- Clubfoot, scoliosis, arachnodactyly: Open the mouth!



Loeys-Dietz Syndrome

- Some similarities to Marfan syndrome
- Scoliosis
- -listhesis

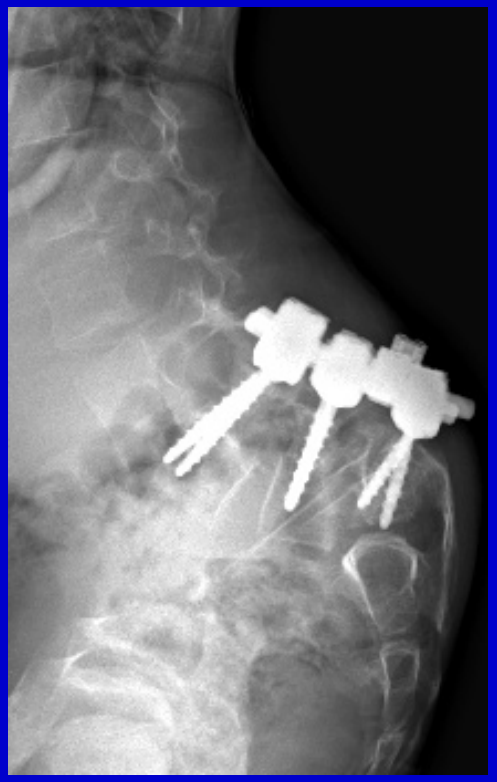
- But also Cervical deformity

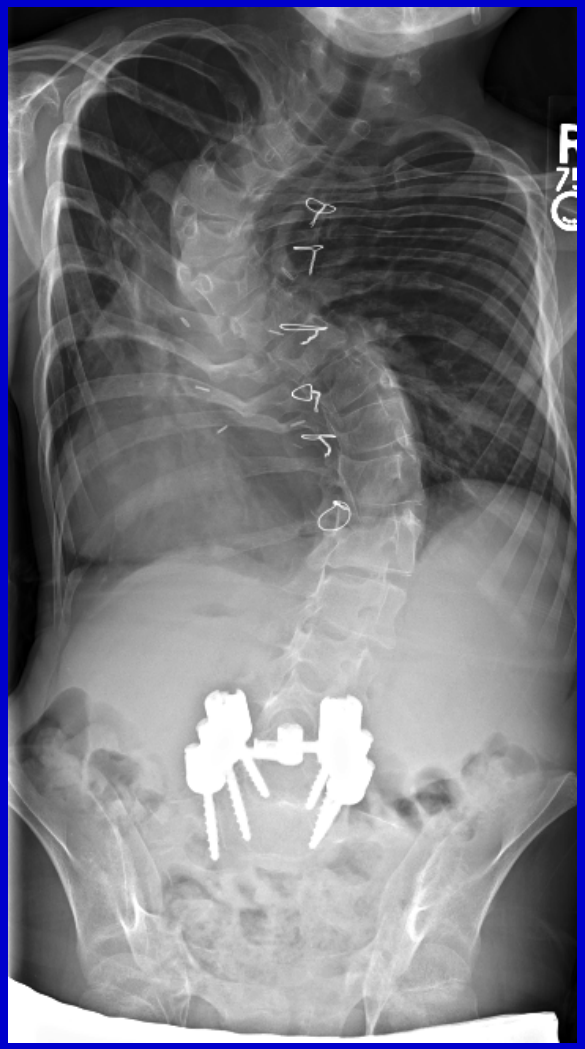


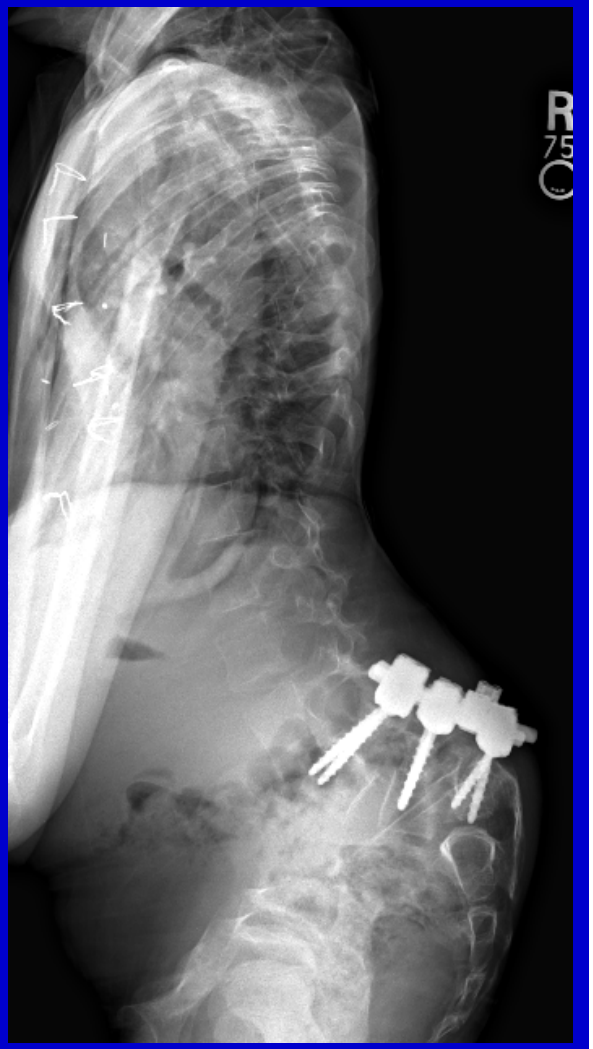
Loeys-Dietz Syndrome

- Lumbar screws plowed

9 yo
LDS
spondy
+ scoli

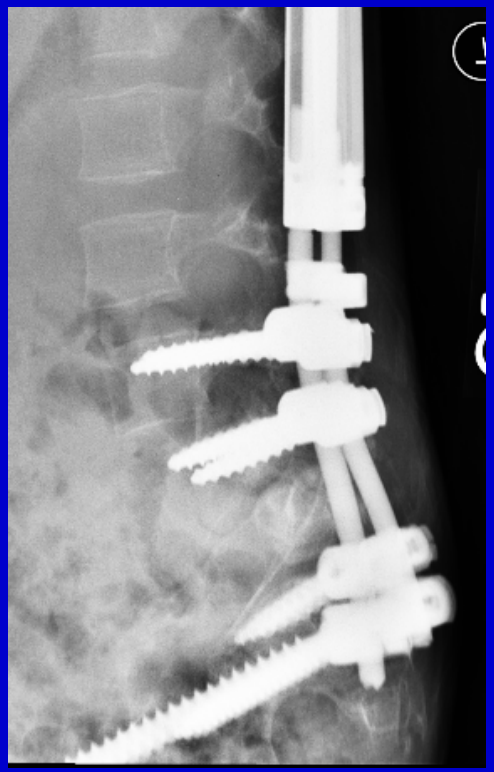


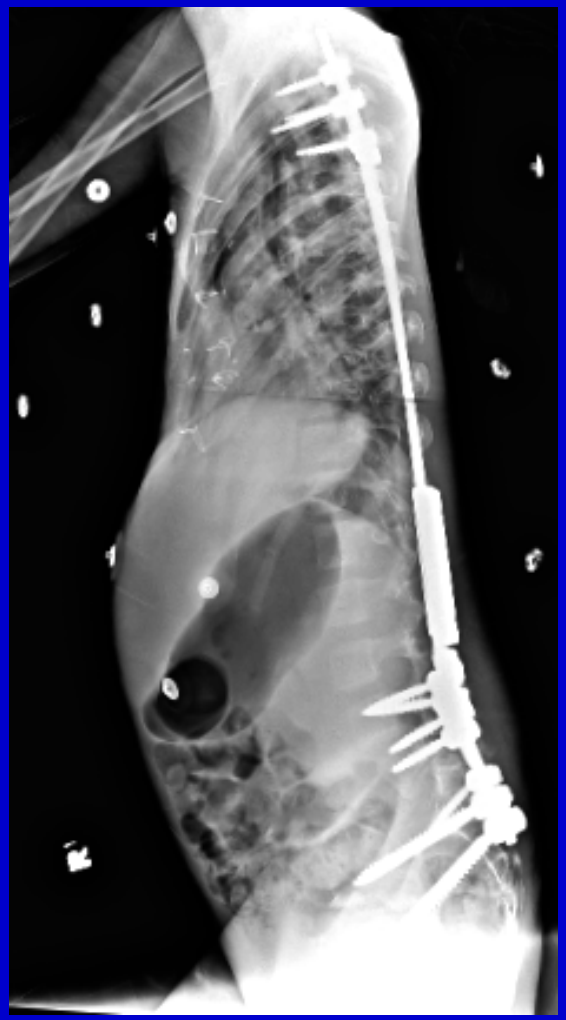


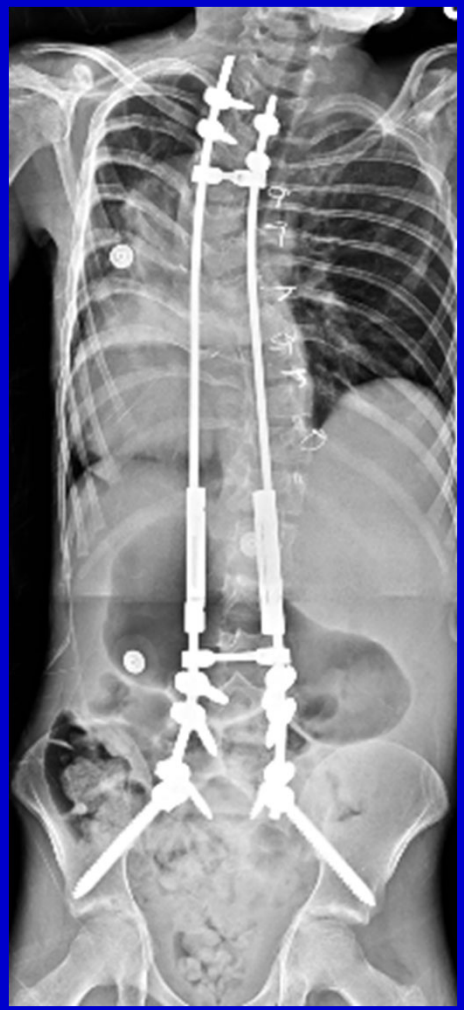


LDS

- Revision

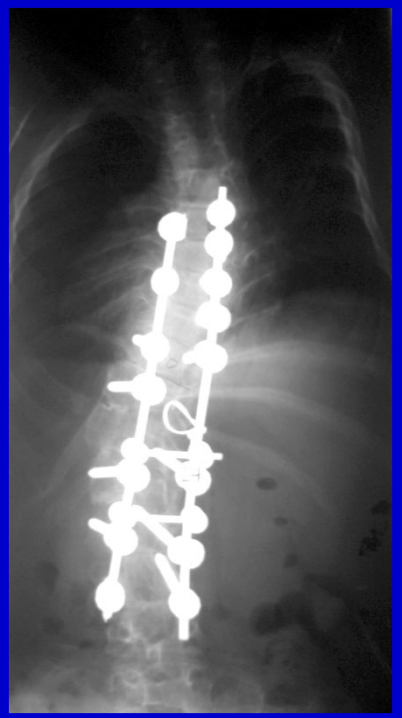






Skeletal dysplasia
-Diastrophic

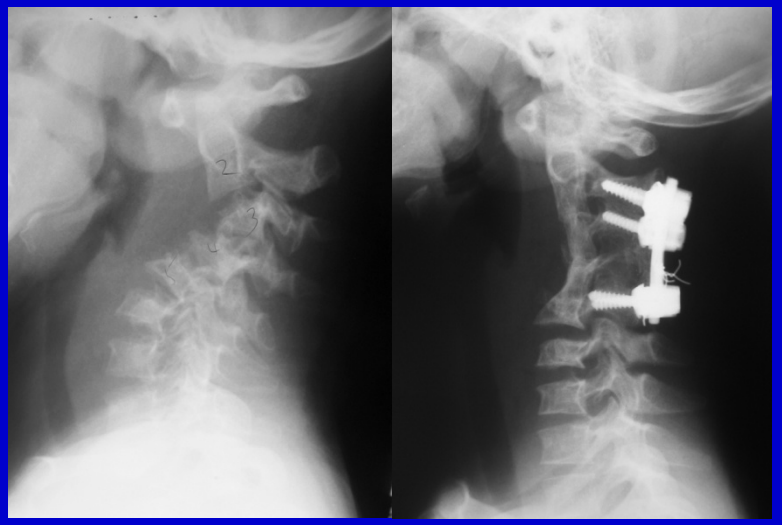
- Growing rods to provide initial correction
- Complication: Short fusion adds on



Skeletal dysplasia

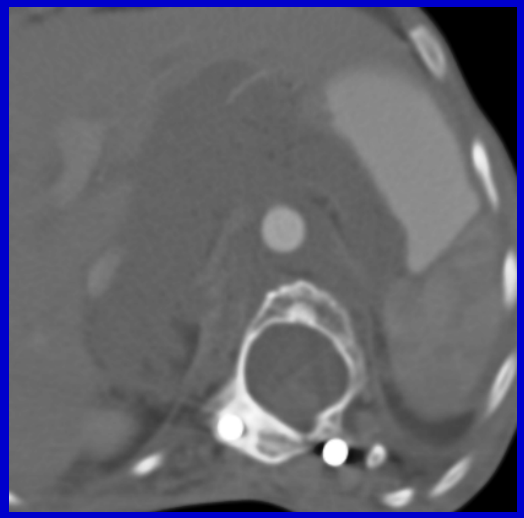
- Too-short fusion adds on

NF1



- Problems:
- Cervical deformity

- Focal bone dysplasia
  - Poor pedicles
  - Poor laminae
- CSF leak
- Neuro risks

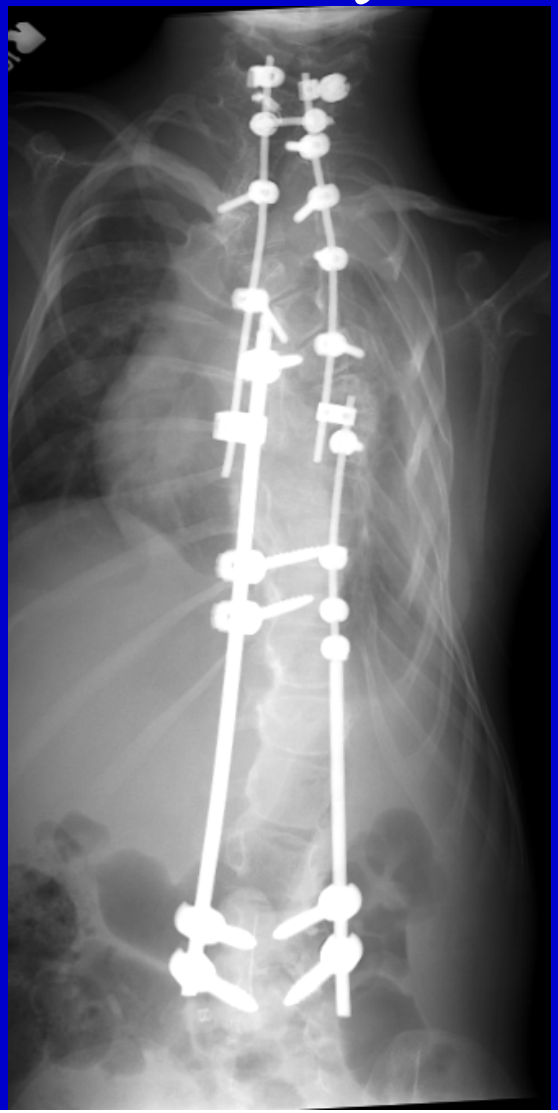


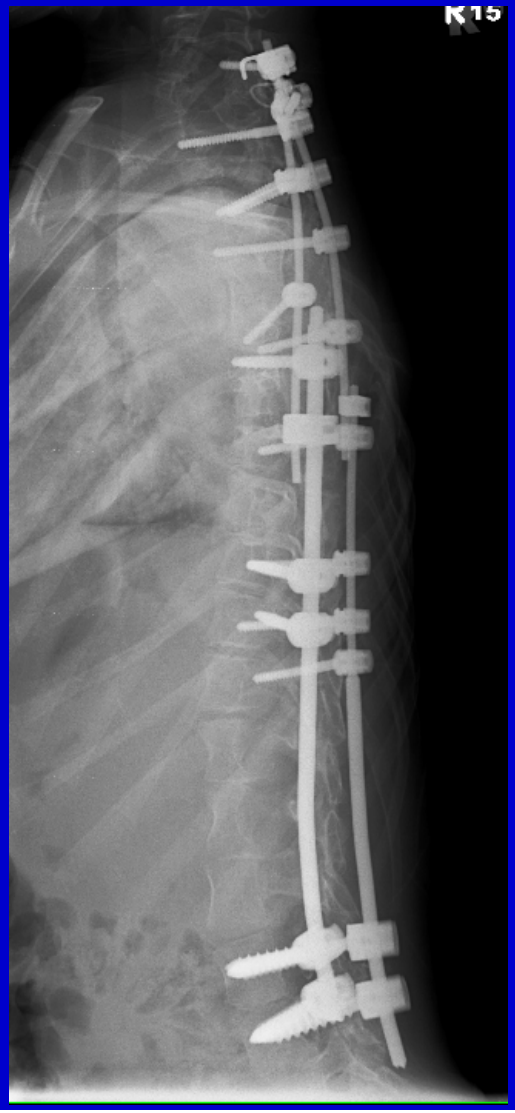
NF1

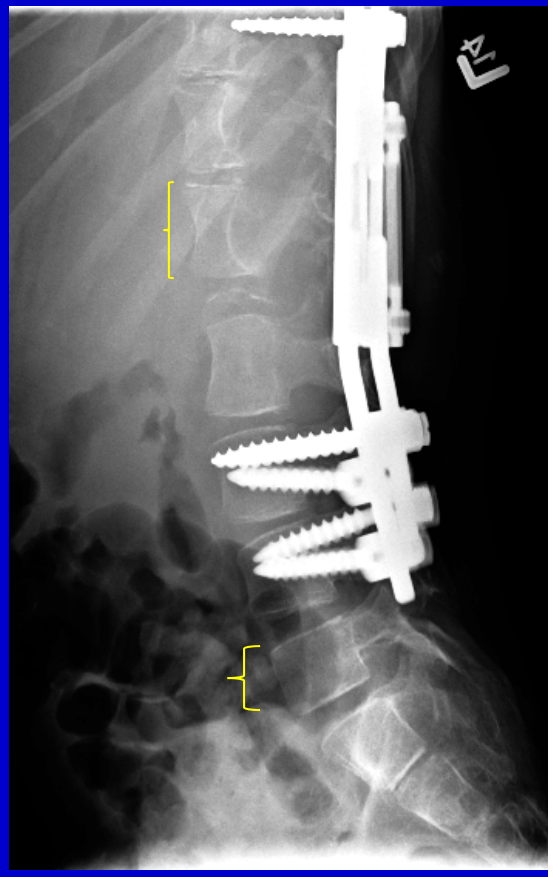
- 5 y.o. Failed VEPTR
  - 90° kyphosis & scoliosis

NF1

- Yearly distraction and osteotomies







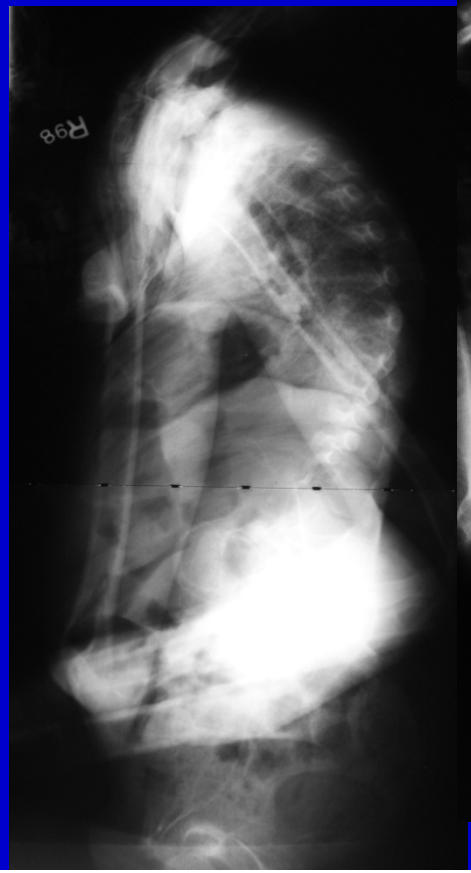
Growing Rods in Neuromuscular Disorders

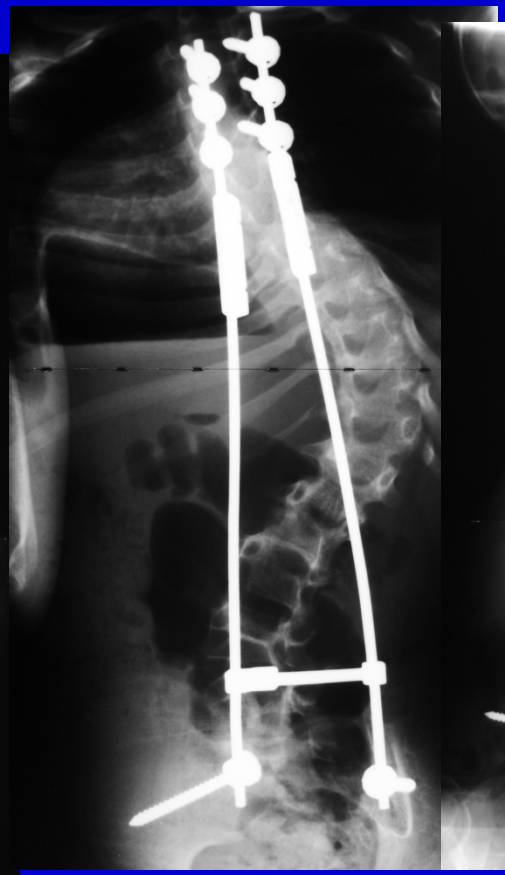
Infection

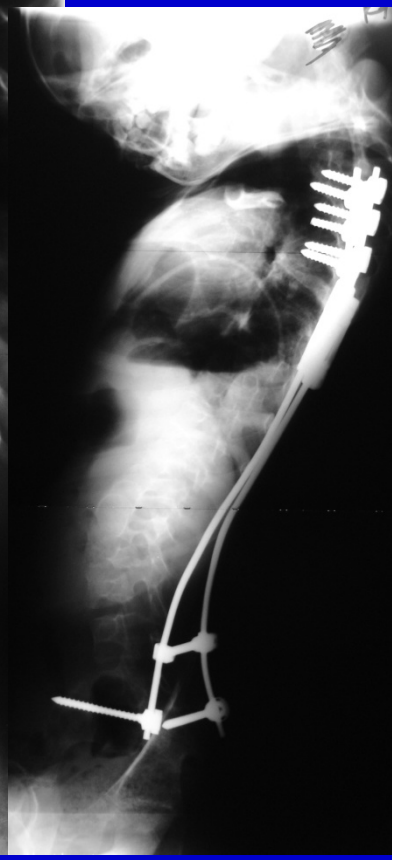
- Increased risk in NM patients
  - Especially CP, MM
  - Anecdotally less in others: SMA

Infection

- Increased in CP, MM
- Small 9 yo w. CP





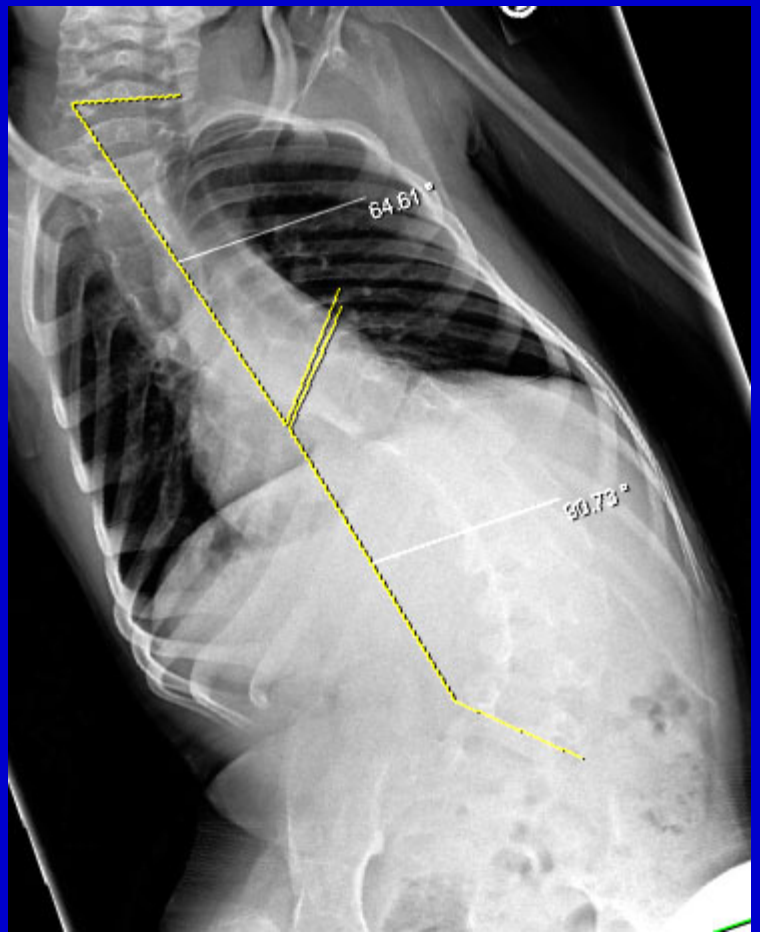


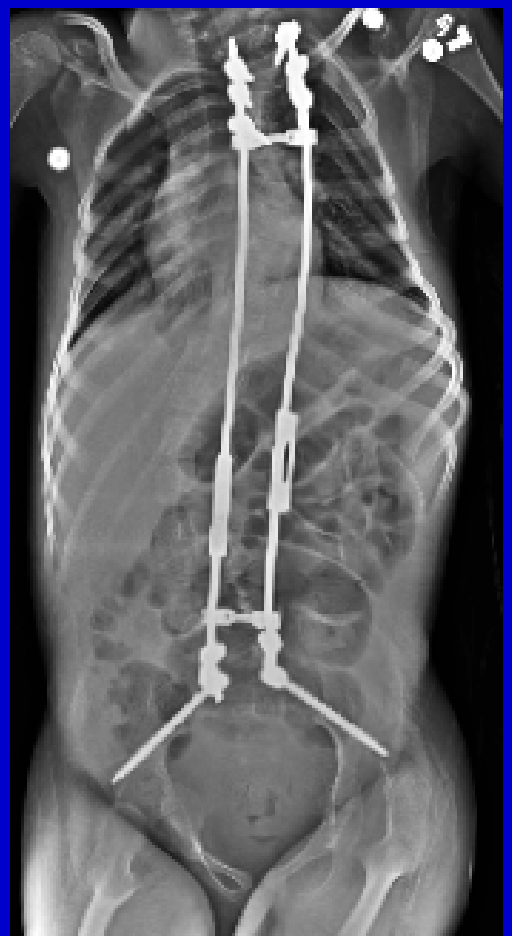
Neuromuscular patients

- Indications for pelvic fixation:
  - severe pelvic obliquity
  - distal deformity (coronal or sagittal)
  - lack of alternative anchor sites
- Rod Breakage
  - not significantly different from the rate for dual growing rods as a whole ($p=0.05$).

Pelvic Fixation

- Personal preference:
  - Iliac screws + S1 screws





Conclusions
Systemic Disorders have

- Increased rate of:
  - Failure of fixation
  - Adding on
  - Infection, Medical complications
- Increased burden of aftercare
- Cost benefit analyses differ for each syndrome

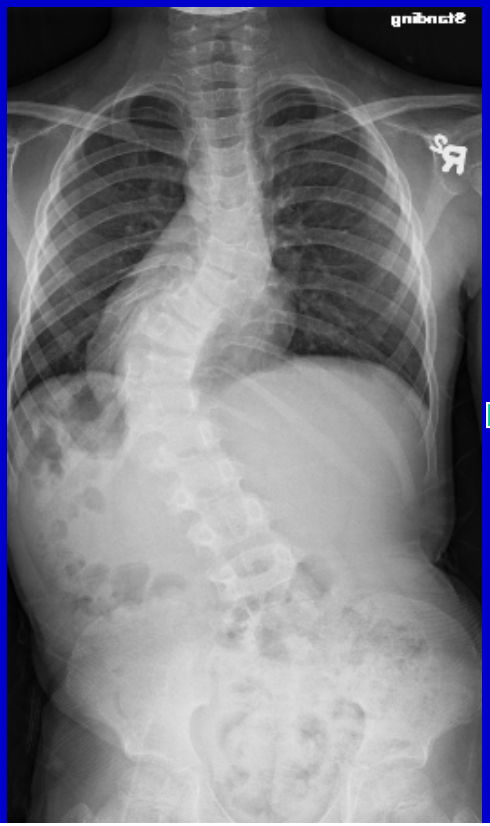
Thank You

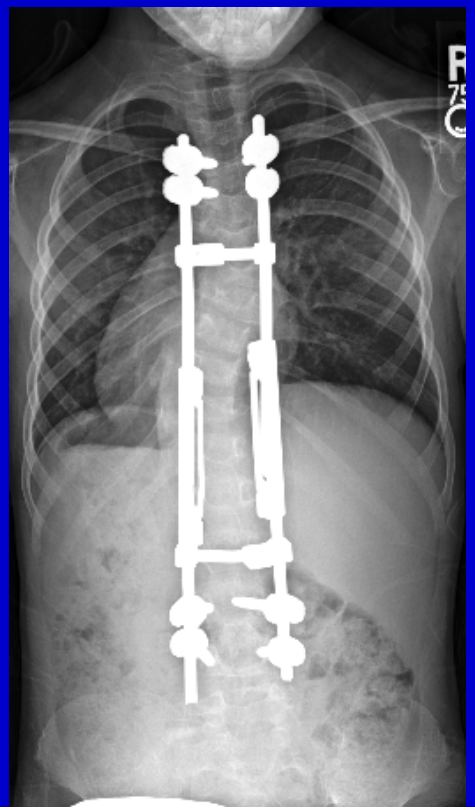
Resources

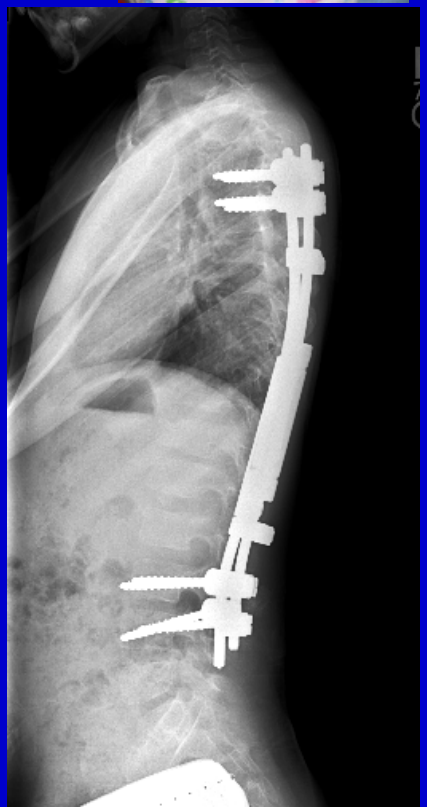
- Patients / Parents / support groups
- Internet has made them “experts”
  - Depth exceeds perspective
- Welcome their contributions
  - Orthopedists too often inhibited by syndromes
- Establish links to trusted “experts”
  - “What other specialists are you seeing?”

Skeletal Dysplasia

- 10 y.o. with SED tarda
  - Dad did not want to lose height
  - Definitive fusion a good alternative







Classification

- Bone
- Connective tissue
- Neurological
- Mixed

- Marfan syndrome
- Heart transplant age 2
- Now 3.5 and stable
- Top Hooks cut out; revised