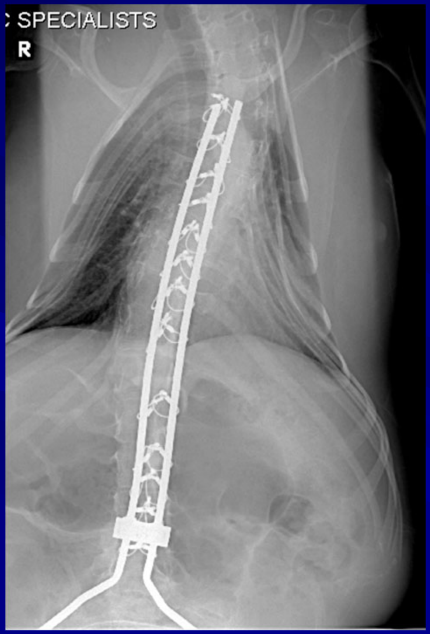
Growing Segmental Spinal Instrumentation Without Fusion in Spinal Muscular Atrophy



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ICEOS November 18-19, 2011

APOS
Adult & Pediatric
Orthopaedic Specialists

Disclosure

- Consultant,   Zimmer Spine
- I have no potential conflicts with this presentation

Background

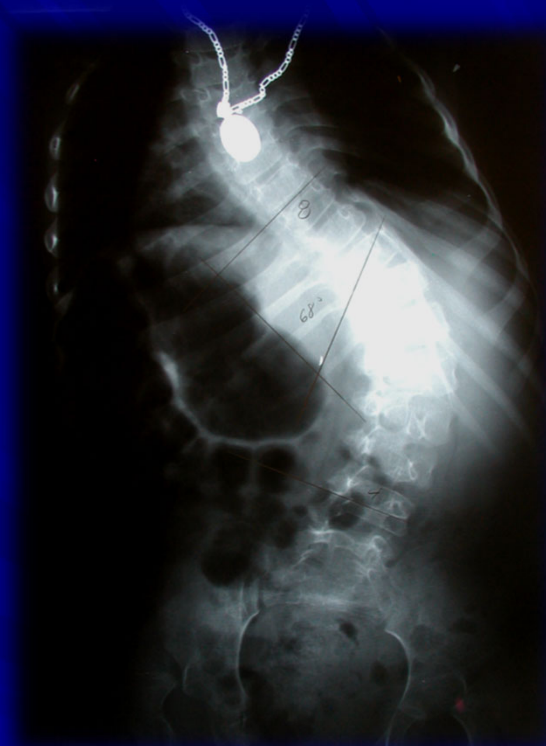
- Spinal muscular atrophy (SMA) is a group of disorders inherited in an autosomal recessive pattern, mapped via linkage studies to chromosome 5q.[1]
- The disorder is secondary to degeneration of the spinal cord anterior horn cells, leading to symmetrical muscle weakness and atrophy of the trunk and proximal musculature of the shoulder and hip girdle.[2]

1 .Brzustowicz, L.M., et al., *Genetic mapping of chronic childhood-onset spinal muscular atrophy to chromosome 5q11.2-13.3.* Nature, 1990. 344(6266): p. 540-1.

2. Thompson, G.H., Berenson,F.R., *Other Neuromuscular Disorders*, in *Lovell and Winter's Pediatric Orthopaedics*, R.T. Morrissy, Weinstein, Stuart L., Editor. 2006, Lippincott Williams and Wilkins. p. 669-673.

Background

- Spinal deformity is commonly associated with SMA.
  - neuromuscular type in which the pelvic obliquity is proportional to the severity of the curve
  - early onset
  - progresses rapidly prior to the onset of puberty
- Respiratory impairment is common in SMA.
  - restrictive lung disease advances simultaneously with the progression of the spinal curvature.



Literature Review

- Chandran, S., et al Early treatment of scoliosis with growing rods in children with severe spinal muscular atrophy: a preliminary report, J Pediatr Orthop. 2011 Jun; 31 (4): 450-4

- McElroy, M.J., et al Growing rods for scoliosis in spinal muscular atrophy: structural effects, complications, and hospital stays, Spine, 2011 Jul 15; 36 (16): 1305-11

- Improvement of trunk height, space available for lung ratio
- Good sagittal and coronal correction maintained
- Multiple surgeries with good safety profile

Purpose

To report the long term follow up of a series of patients with spinal muscular atrophy whose neuromuscular scoliosis was treated surgically by growing posterior segmental spinal instrumentation without fusion with only ONE surgical intervention

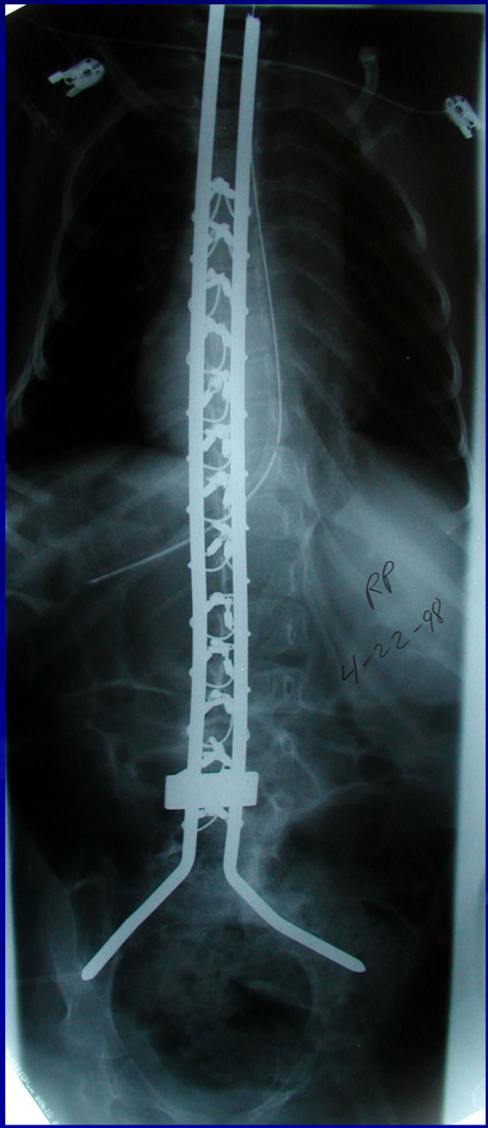
Methods

- Retrospective chart and radiographic review of a single-center series of non-ambulatory SMA patients

Inclusion Criteria:

- Spinal Muscular Atrophy type I or II
- Curve progression despite bracing
  - Curve Magnitude > 40˚
- Growing posterior spinal instrumentation without fusion
- Minimum 6-year follow-up

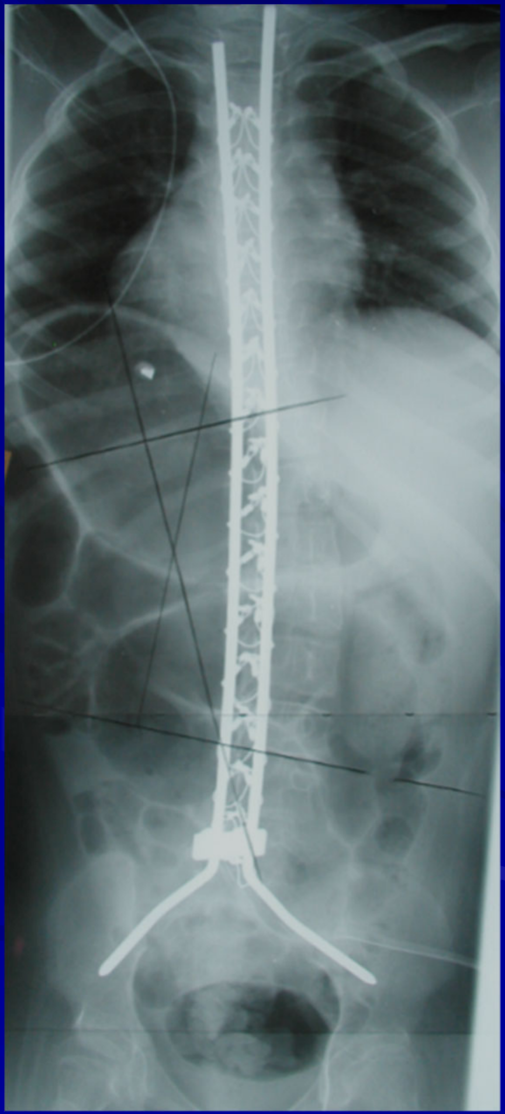
Surgical Technique



- A standard posterior approach
- Subperiosteal dissection of the lamina was carried laterally to the facet joints which were preserved.
- Sublaminar cables were passed at each level
- Two paired 3/16th inch unit rods were cut and contoured for maximal curve correction, normal sagittal alignment, and secured in the illium

Surgical Technique

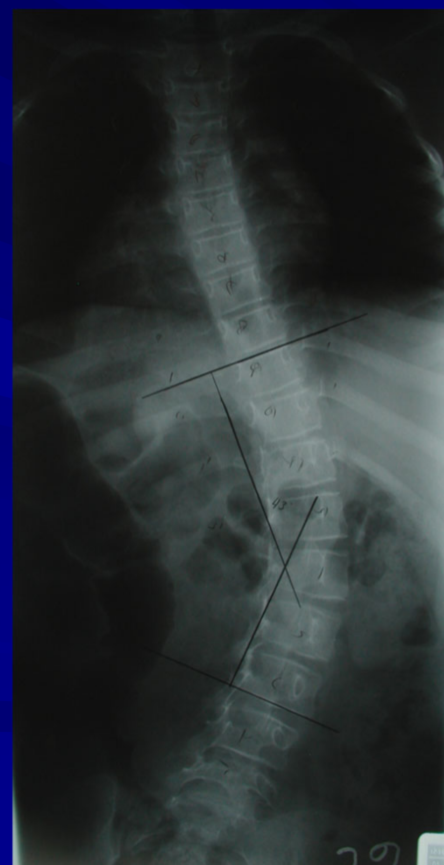
- A crosslink was placed at the caudal end of the construct
- The cephalad rod ends were left long
  - To Allow spinal growth along the construct, and
  - Obviate the need for further lengthening surgeries
- Postoperative bracing was not utilized



Methods

Serial radiographs were followed for:

1. curve progression
2. spinal growth
3. implant stability



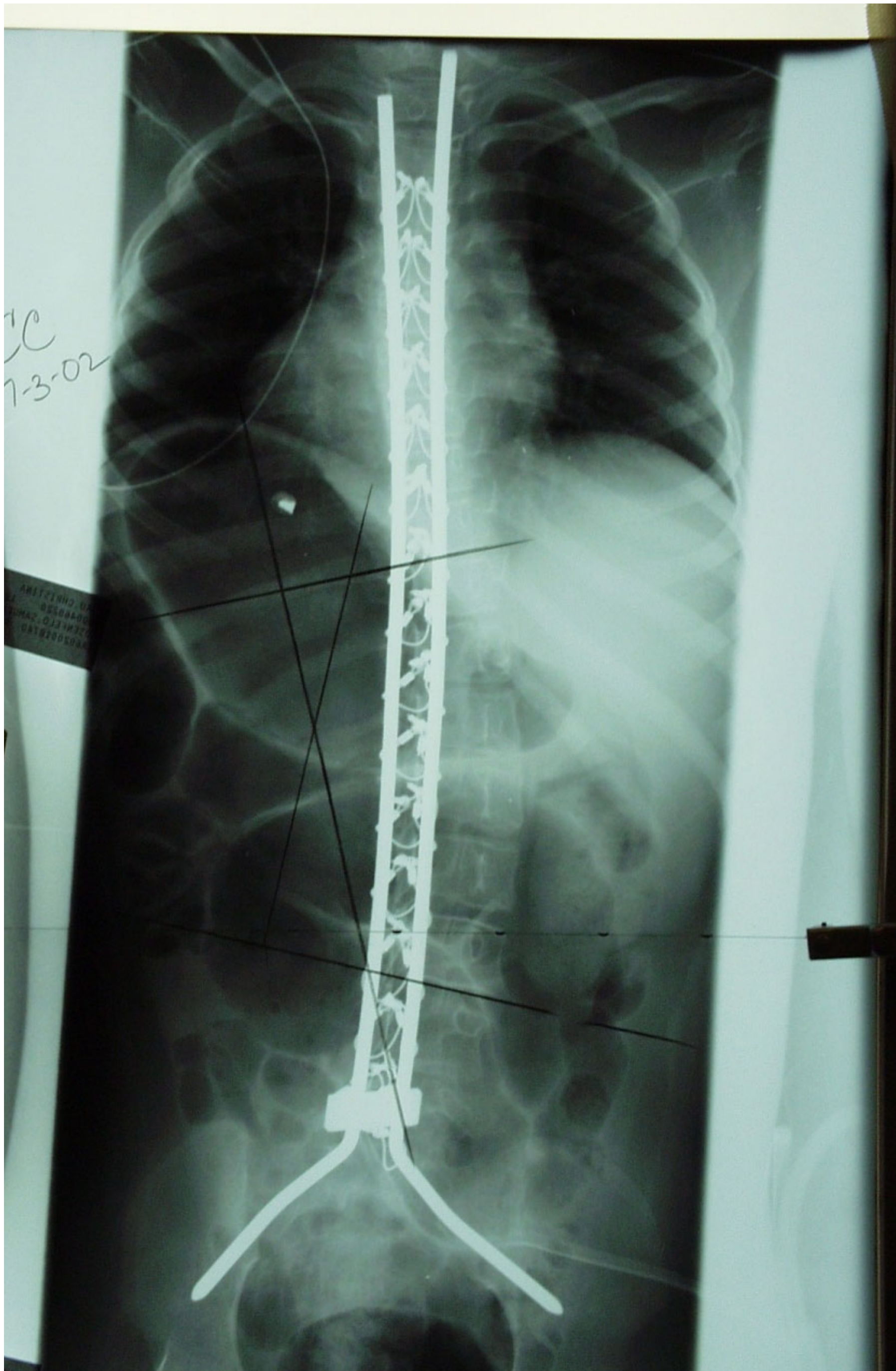
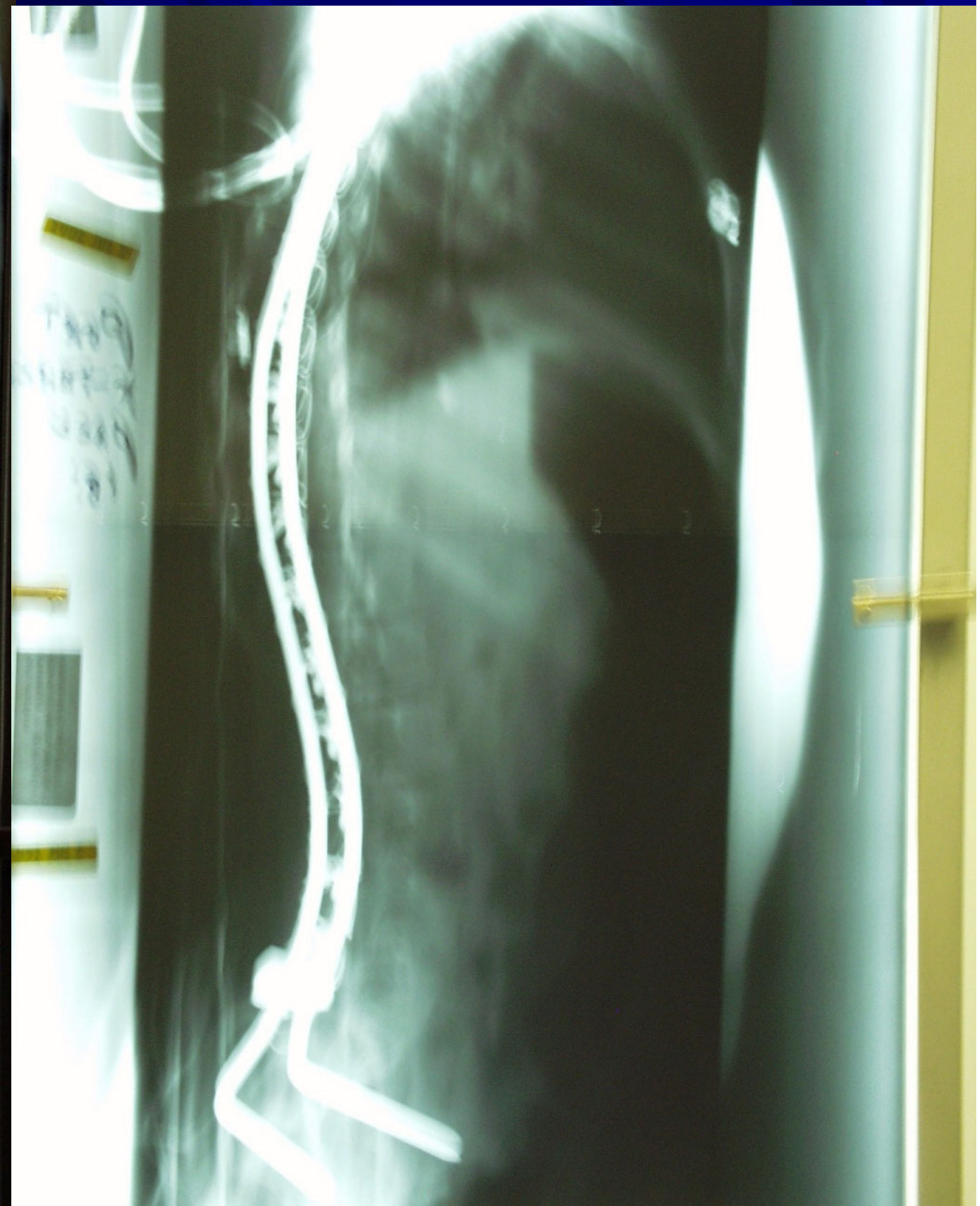
Results

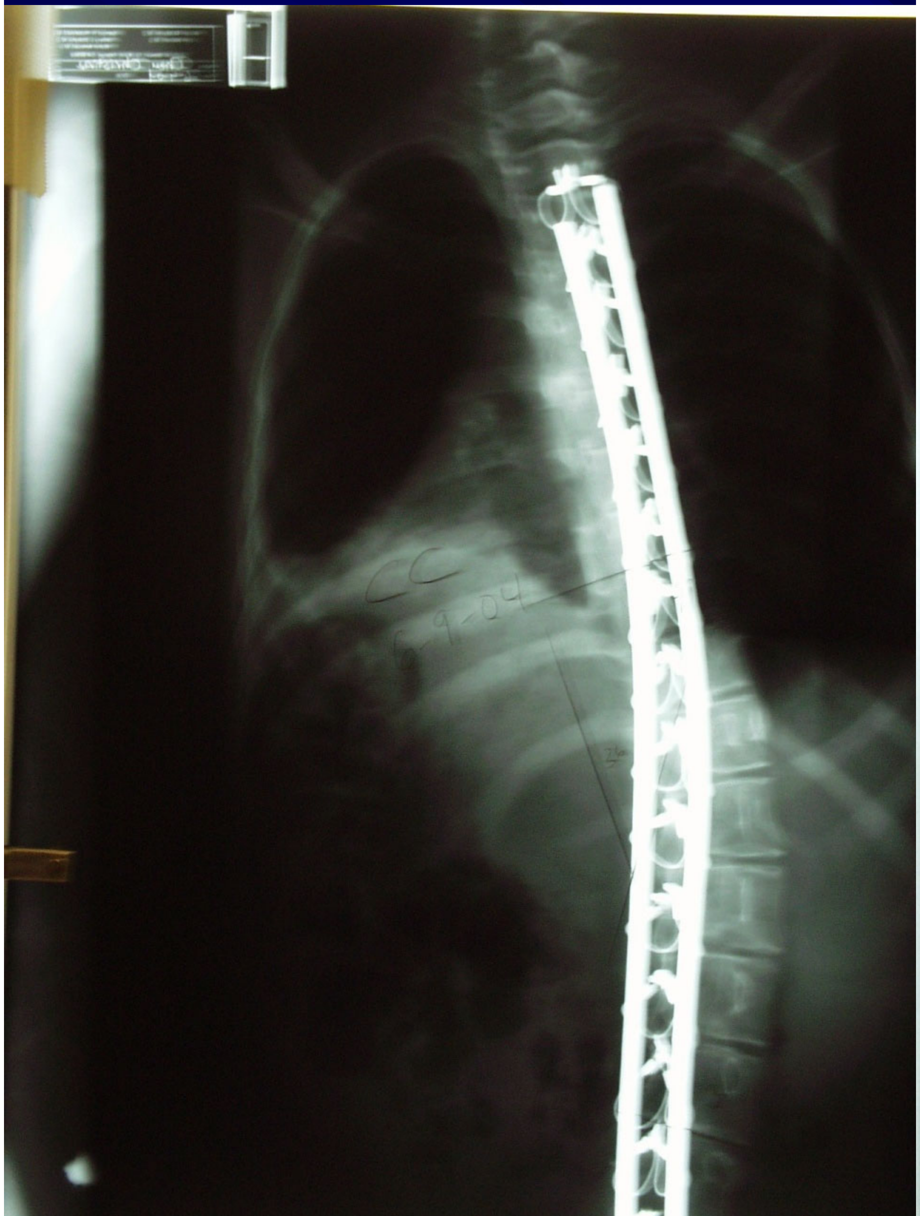
Demographics	Age	Follow-up (years)	Cobb Angle (pre-op)	Cobb Angle (last exam)	Junctional Kyphosis	Total Growth (cm/year)	Risser Stage (last exam)	Complications
♀ SMA-2	18+6	13	44°	30°	12°	7.9 cm	V	None
♀ SMA-2	19+11	9	48°	38°	16°	10 cm	V	None
♂ SMA-2	15+11	9	68°	55°	45°	12 cm	III	None
♀ SMA-1	15+6	8	50°	42°	2°	5.8 cm	I	Revision of pelvic fixation
♂ SMA-1	13+8	6	76°	35°	17°	4.6 cm	0	None
Mean Values	16	10	57°	40°	18°	1.4	III	-
(Range)	(9-19)	(6-13)	(44-76°)	(30-55°)	(2-45°)	-	-	-

Results

❖ Patients on average achieved 1.4cm/year of spinal growth measured from T1-S1 on sequential radiographs

Spinal Growth	cm/yr
Patient A	0.8
Patient B	2.2
Patient C	1.4
Patient D	0.8
Patient E	2
Mean	**1.4**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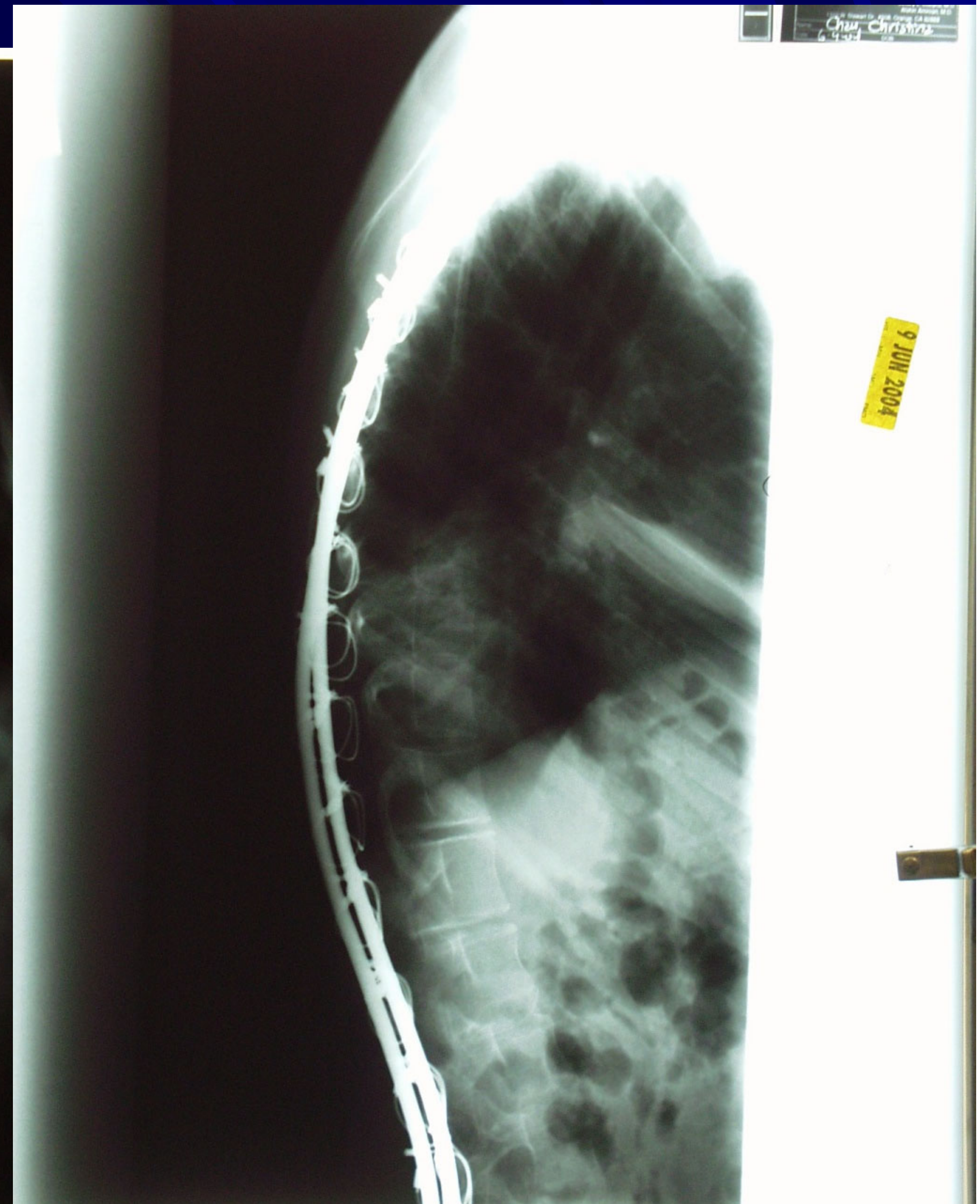
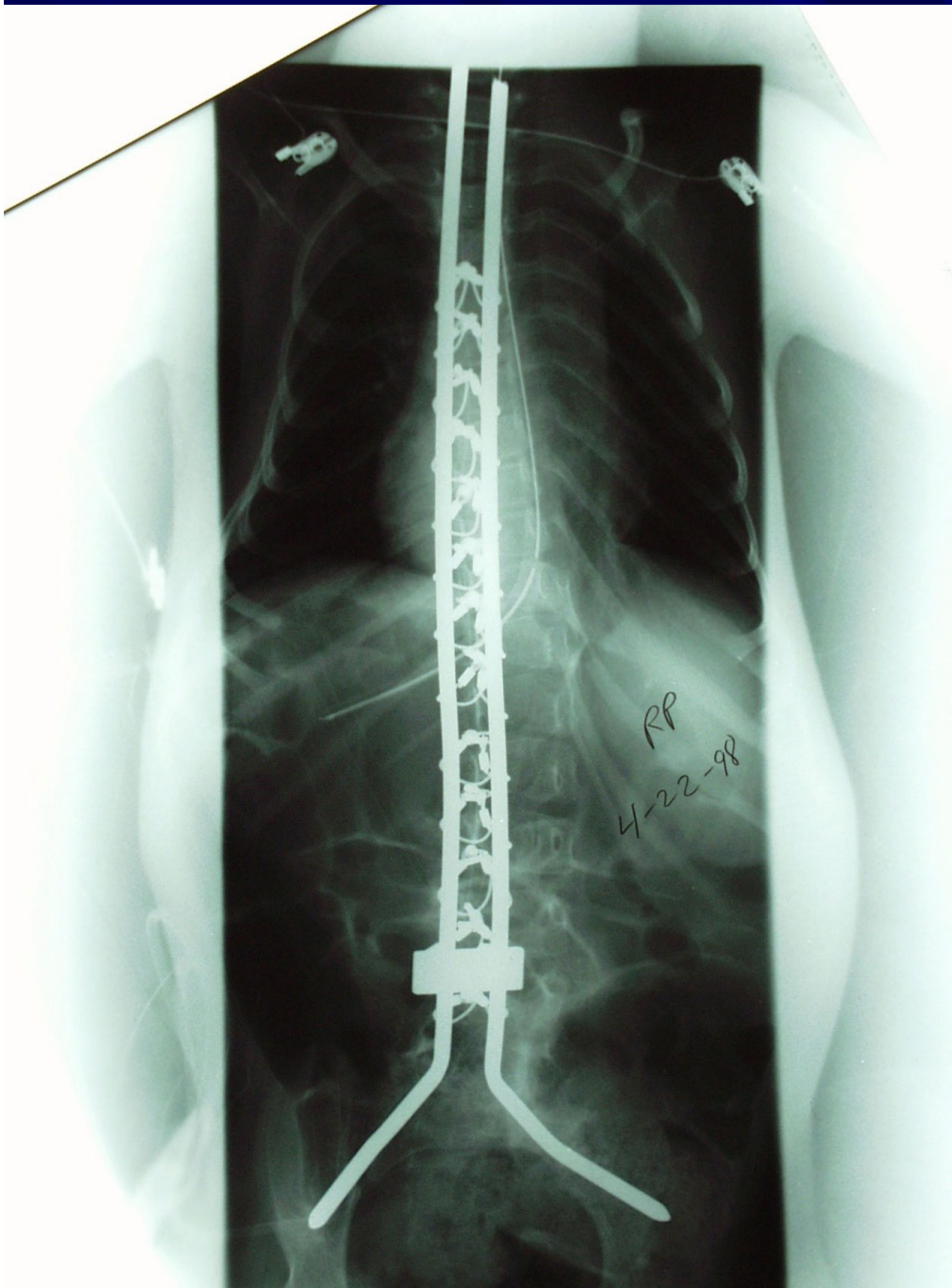

9 JUN 2004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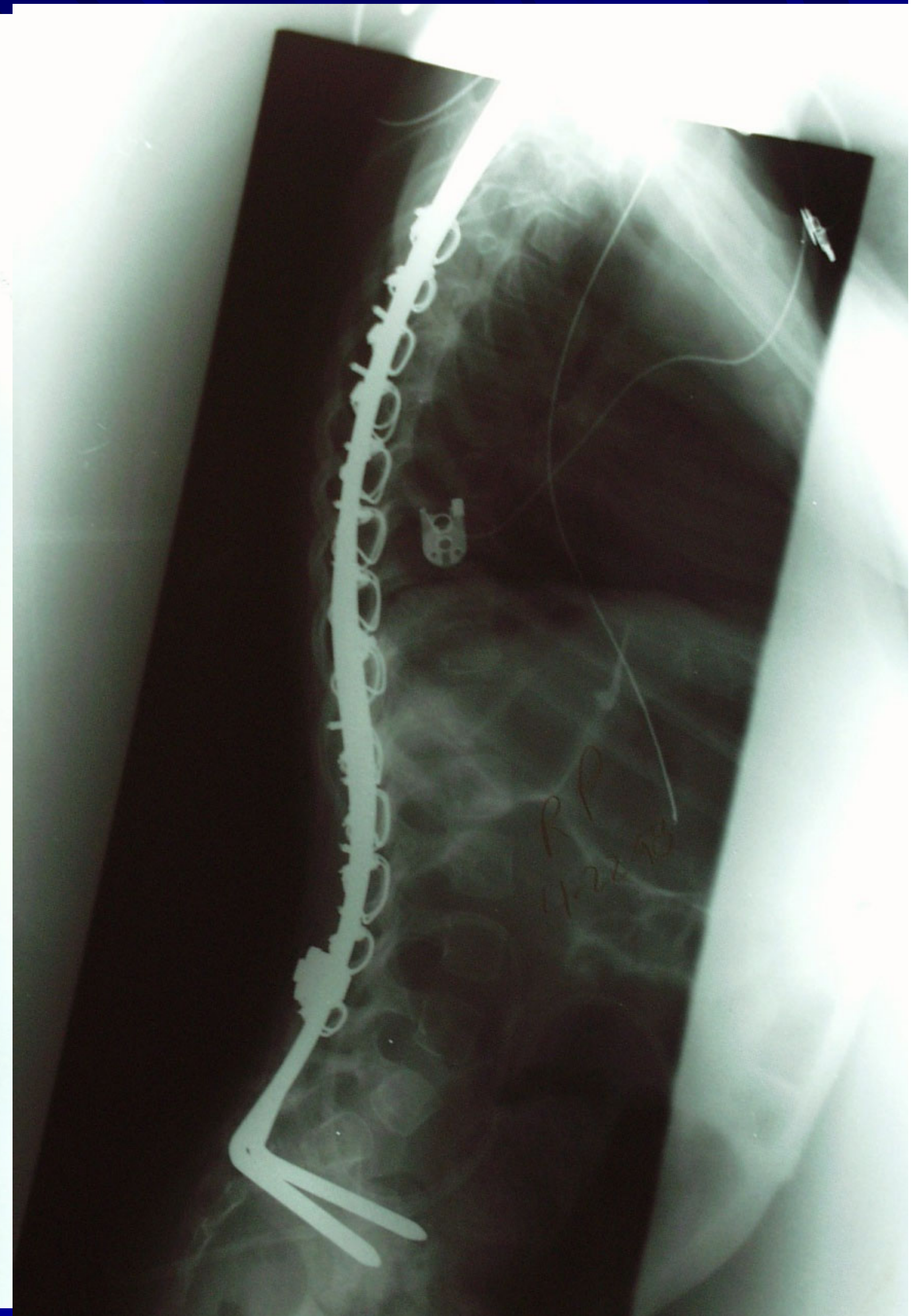
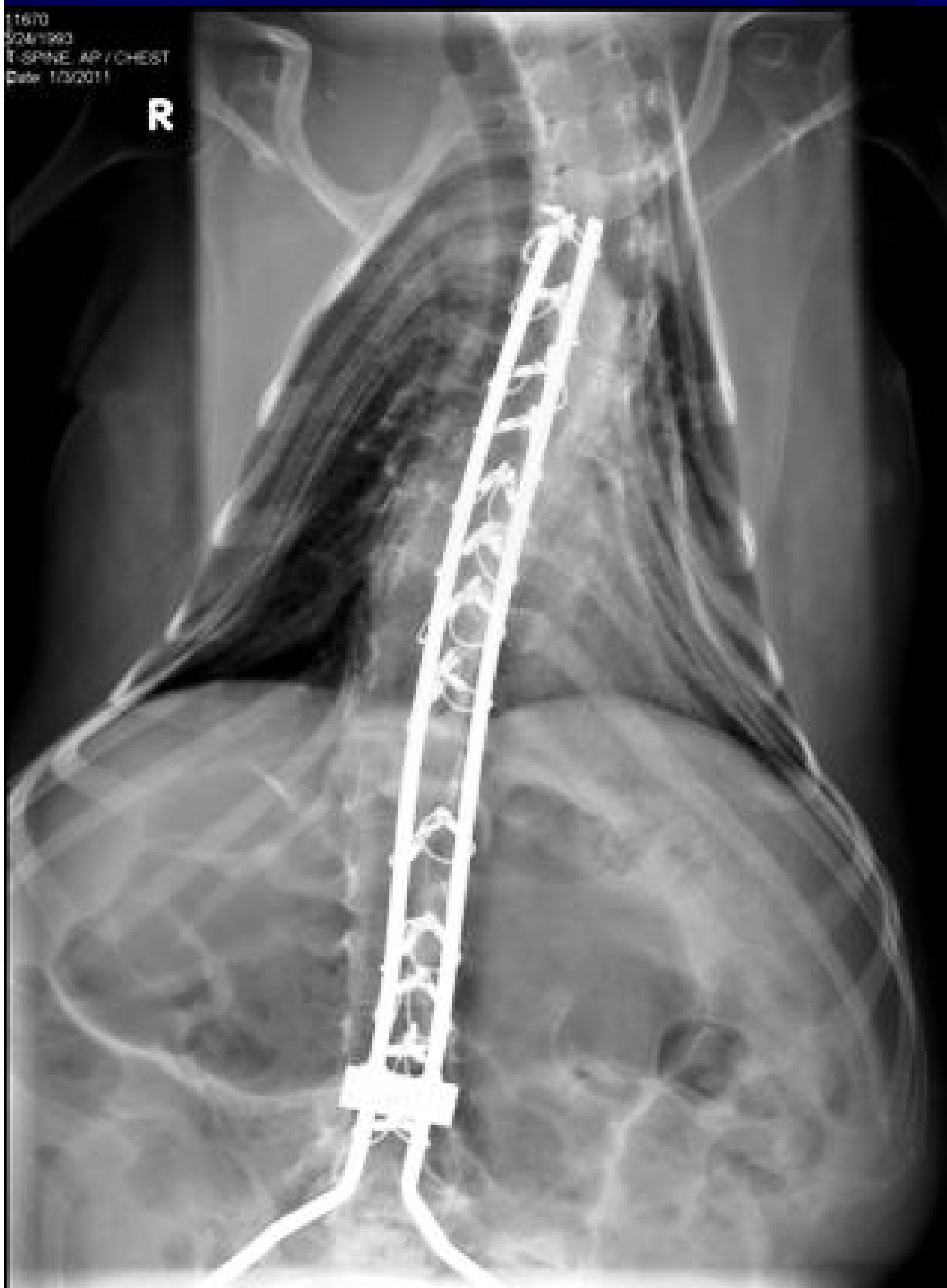



17°
RP
6-2-04


2 JUN 2004
RP
6-2-04


11670
5/24/1993
T-SPINE, AP / CHEST
Date: 1/3/2011
R


ND PEDIATRIC ORTHO
70
/1993
MBAR, LATERAL / PELVIS
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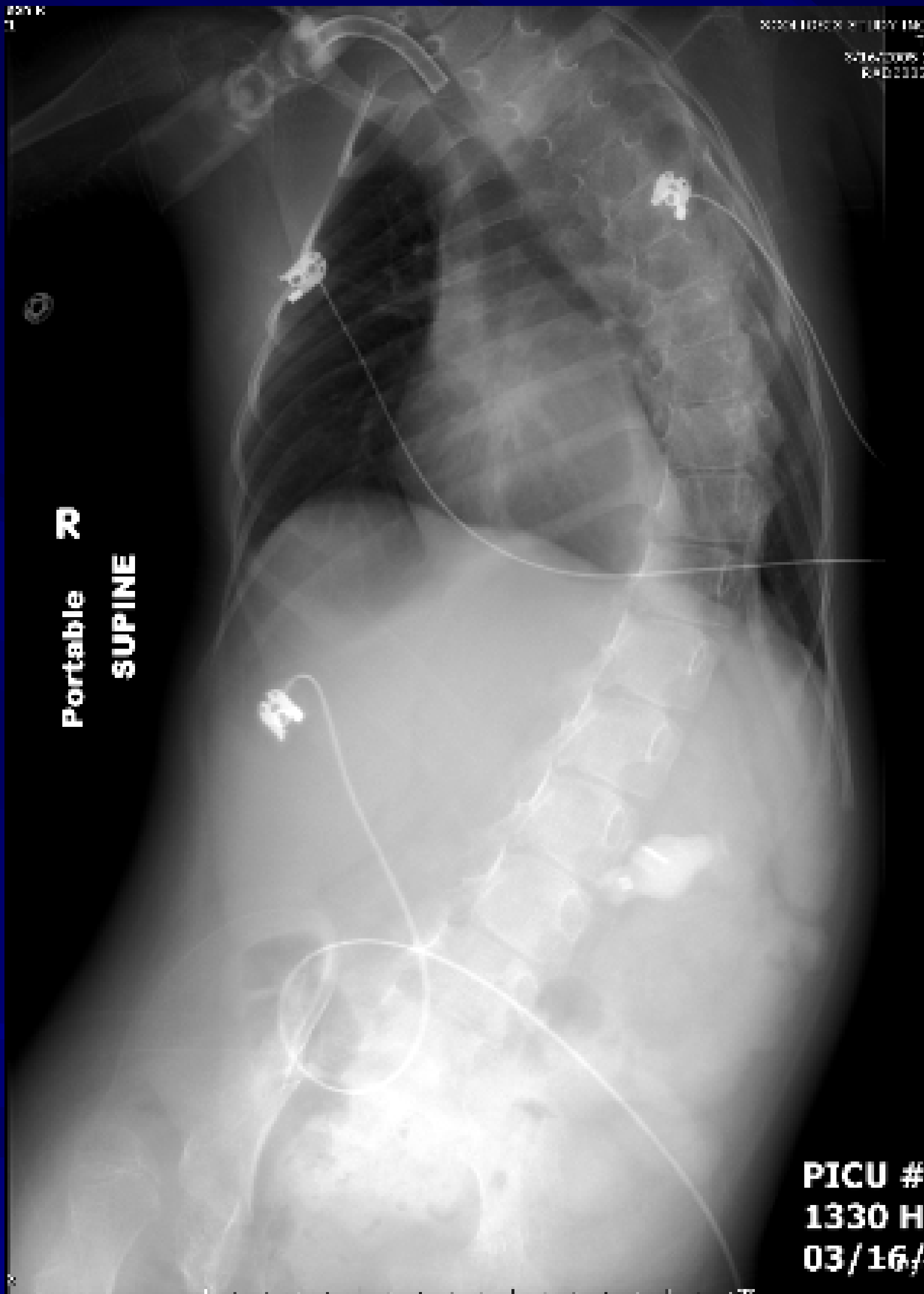
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Portable
SUPINE
PICU #
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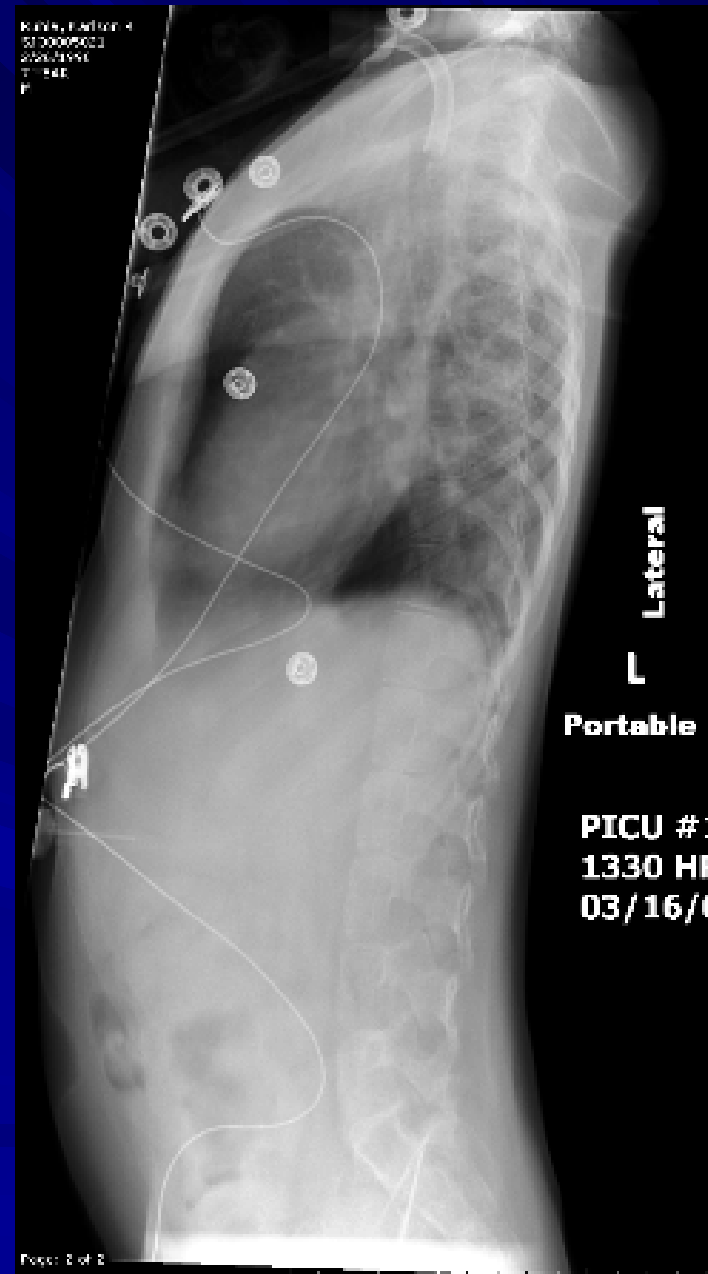
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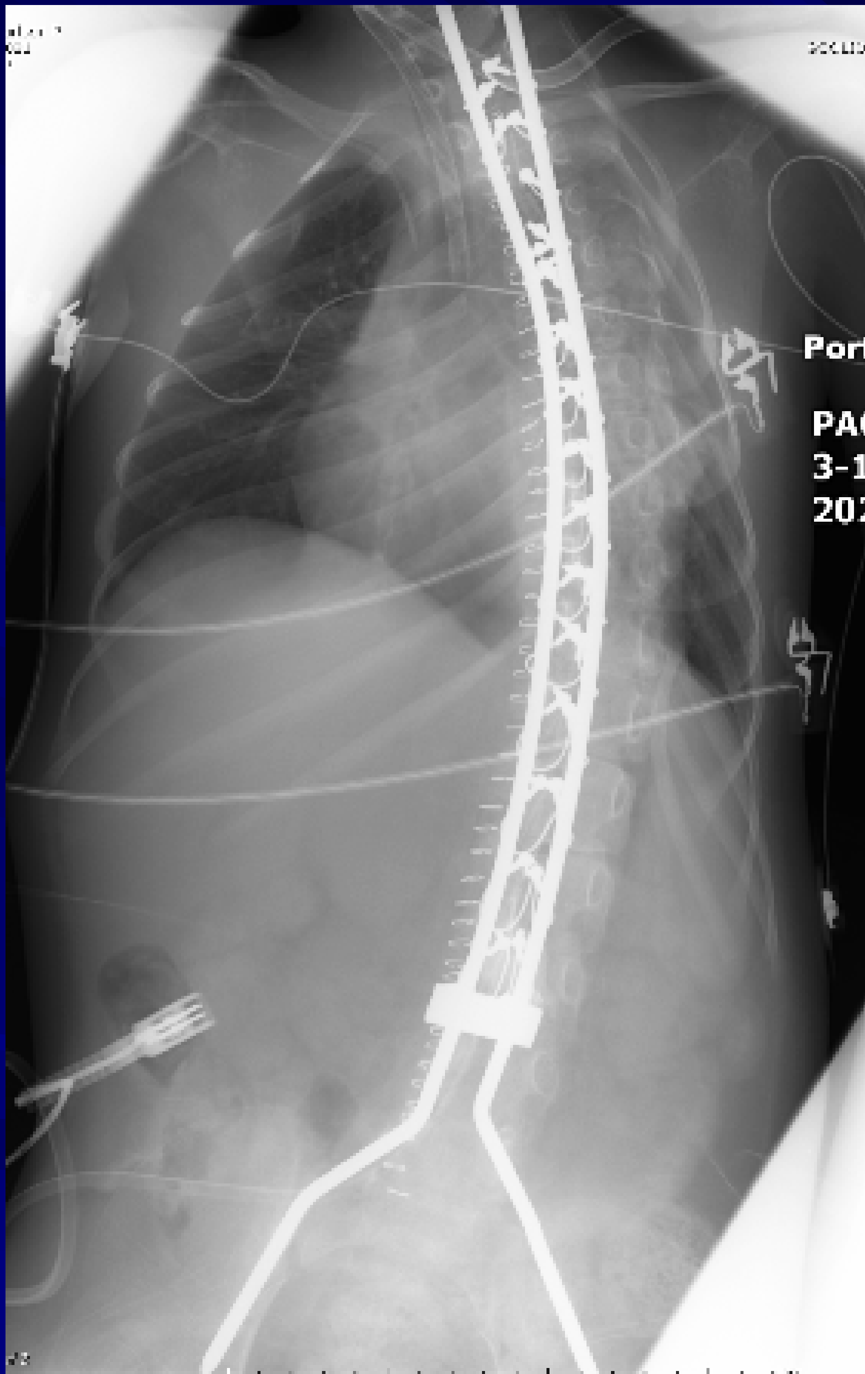

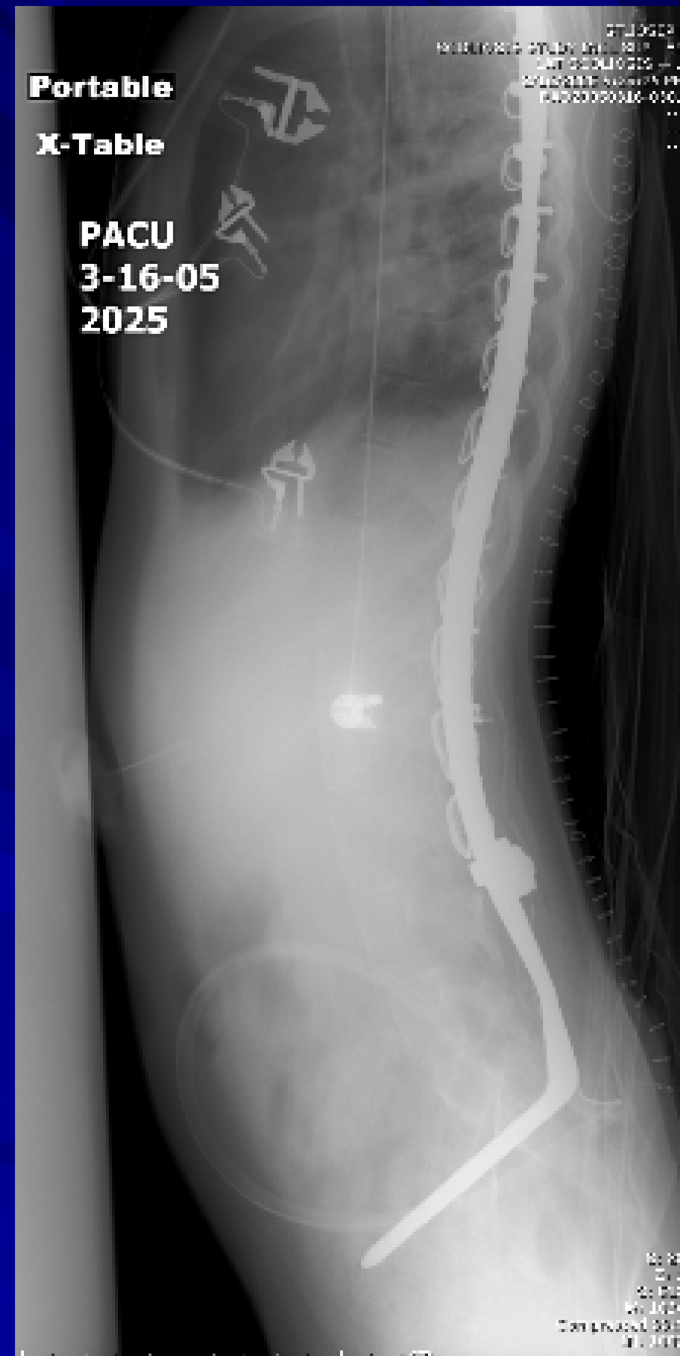
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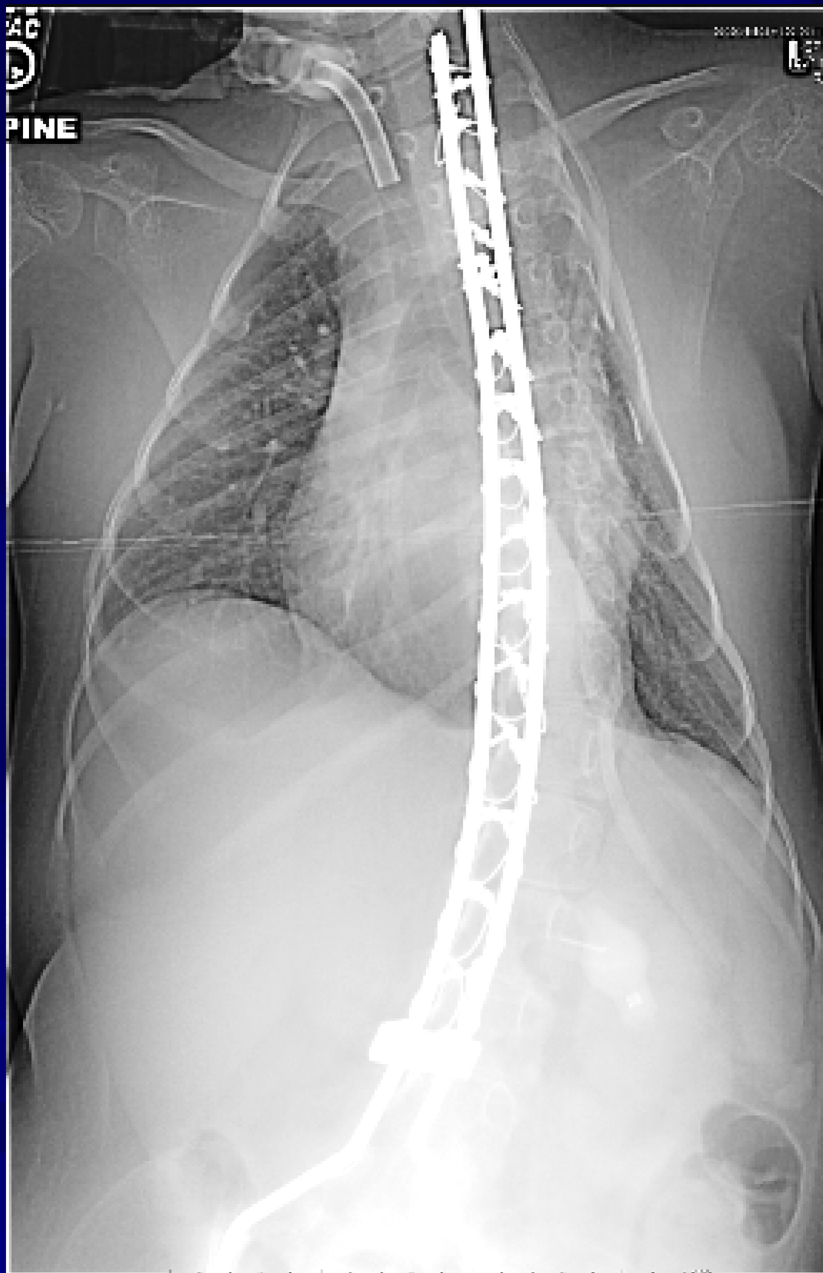
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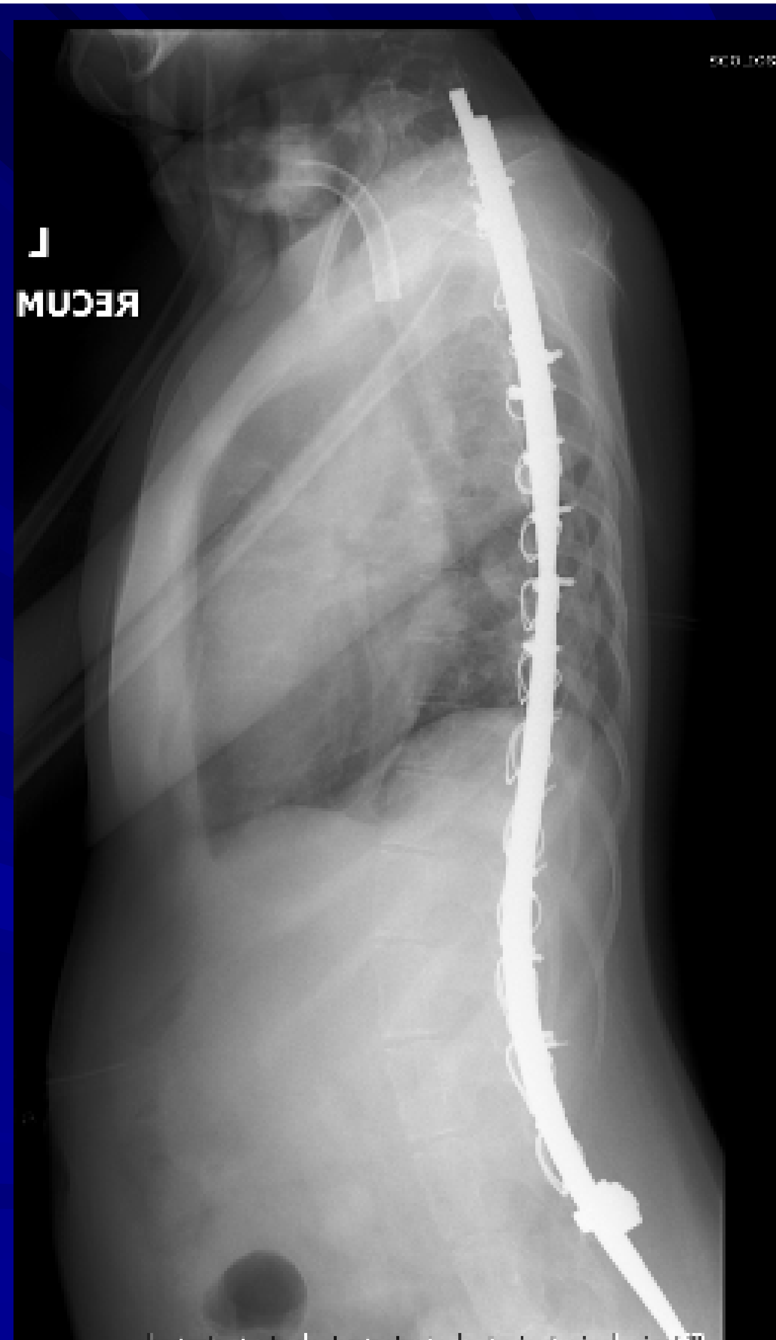

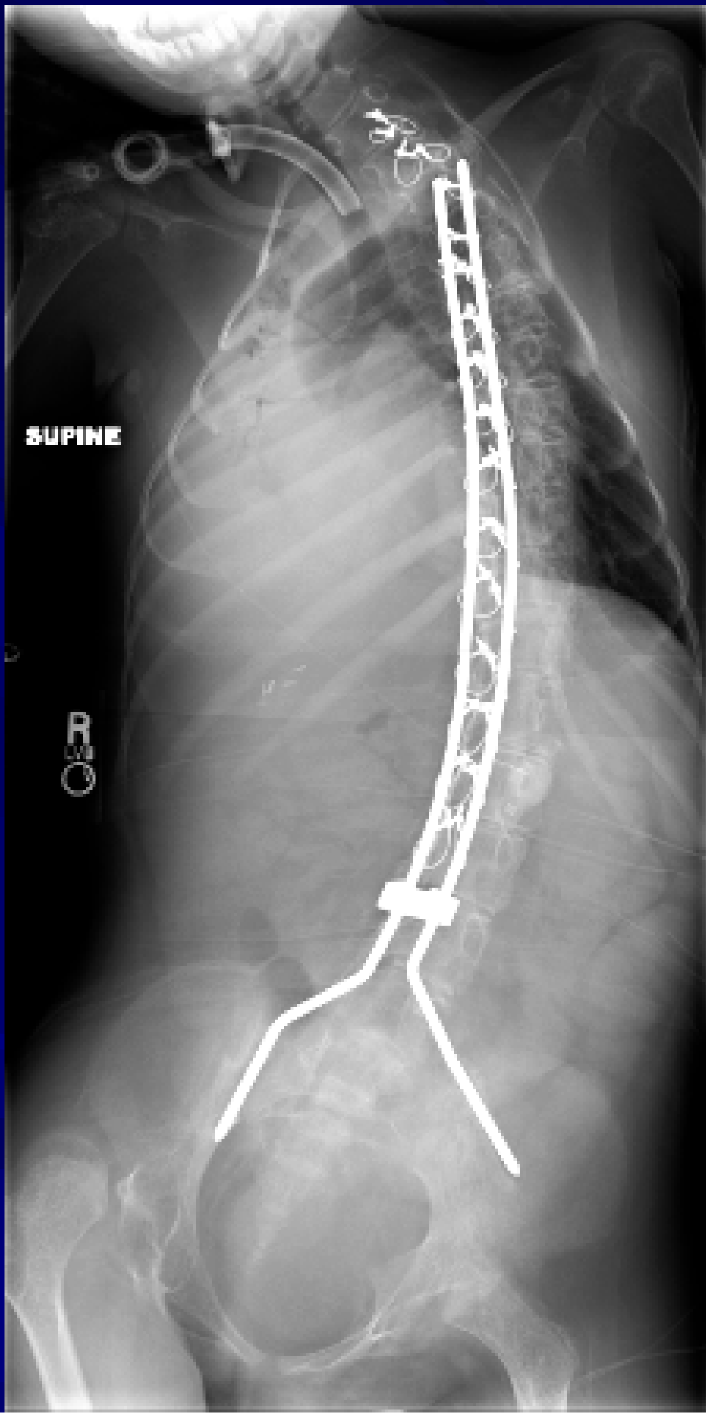
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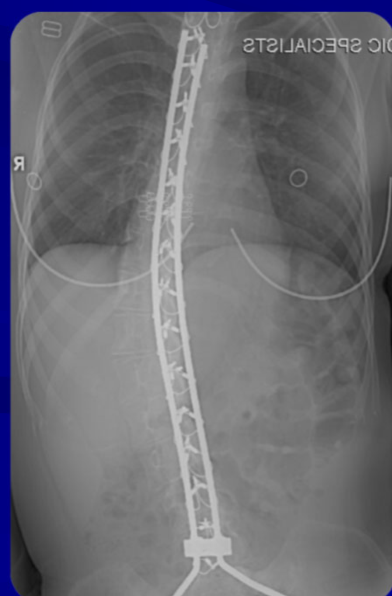
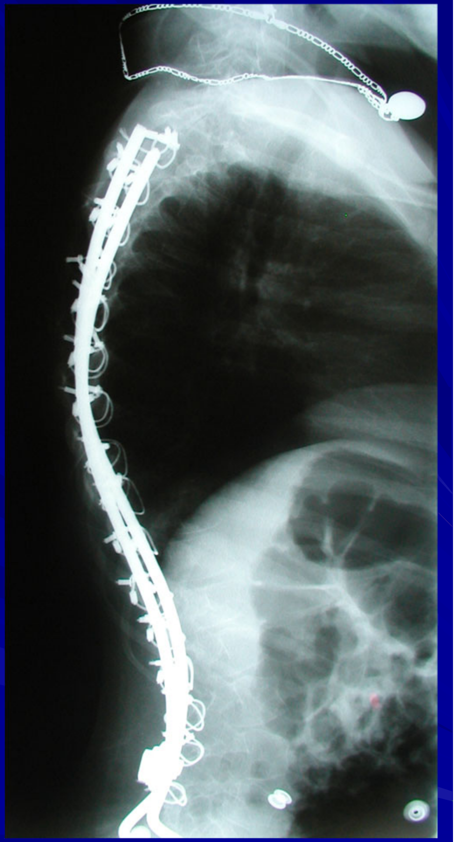
Take Home Points

- Growing Segmental Spinal Instrumentation without fusion is a well-tolerated & effective surgical technique that avoids the morbidity associated with multiple surgeries in this very fragile patient subset



Take Home Points

- ❖ Allows spinal growth without multiple lengthening procedures
- ❖ No observed spontaneous fusion
- ❖ Controls Sagittal & Coronal Alignment
  - – Despite the spine growing past the implant, significant post-op proximal junctional kyphosis was not observed
  - – Stability during growth and beyond without the need for definitive fusion



Additional Applications of this Technique

- Nemaline rod myopathy
- Congenital muscular dystrophy
- Rett syndrome
- Mitochondrial myopathy