Management of Spinal Deformities in Spinal Muscular Atrophy

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Disclosure

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

- Chest wall deformities and scoliosis contribute to restrictive pulmonary disease.
- Pulmonary complications cause morbidity and mortality.
- Weak intercostal muscles and unopposed diaphragmatic function may result in the bell shaped chest (parasol rib deformity).
- Symptoms include poor management of airway secretions, hypoventilation during sleep, poor chest wall development, recurrent pulmonary infections, skin pressure areas, back and buttock pain.

- Scoliosis occurs in greater than 50% of patients with SMA 1 and 2.
- Non-ambulatory patients are at greater risk for scoliosis.
- Pelvic obliquity and kyphosis are often associated with this spinal deformity.
- Because of the progression of the scoliosis and pulmonary compromise, early intervention is important.

What are the goals of treatment?

- Improve sitting balance/tolerance
- Decrease likelihood of decubiti, aspiration
- Relief of pain in hips and back
- Decrease need for assistance
- Eliminate use upper extremities for support
- Facilitate positioning/transfers
- Improve pulmonary function or pulmonary growth

Nonsurgical Management

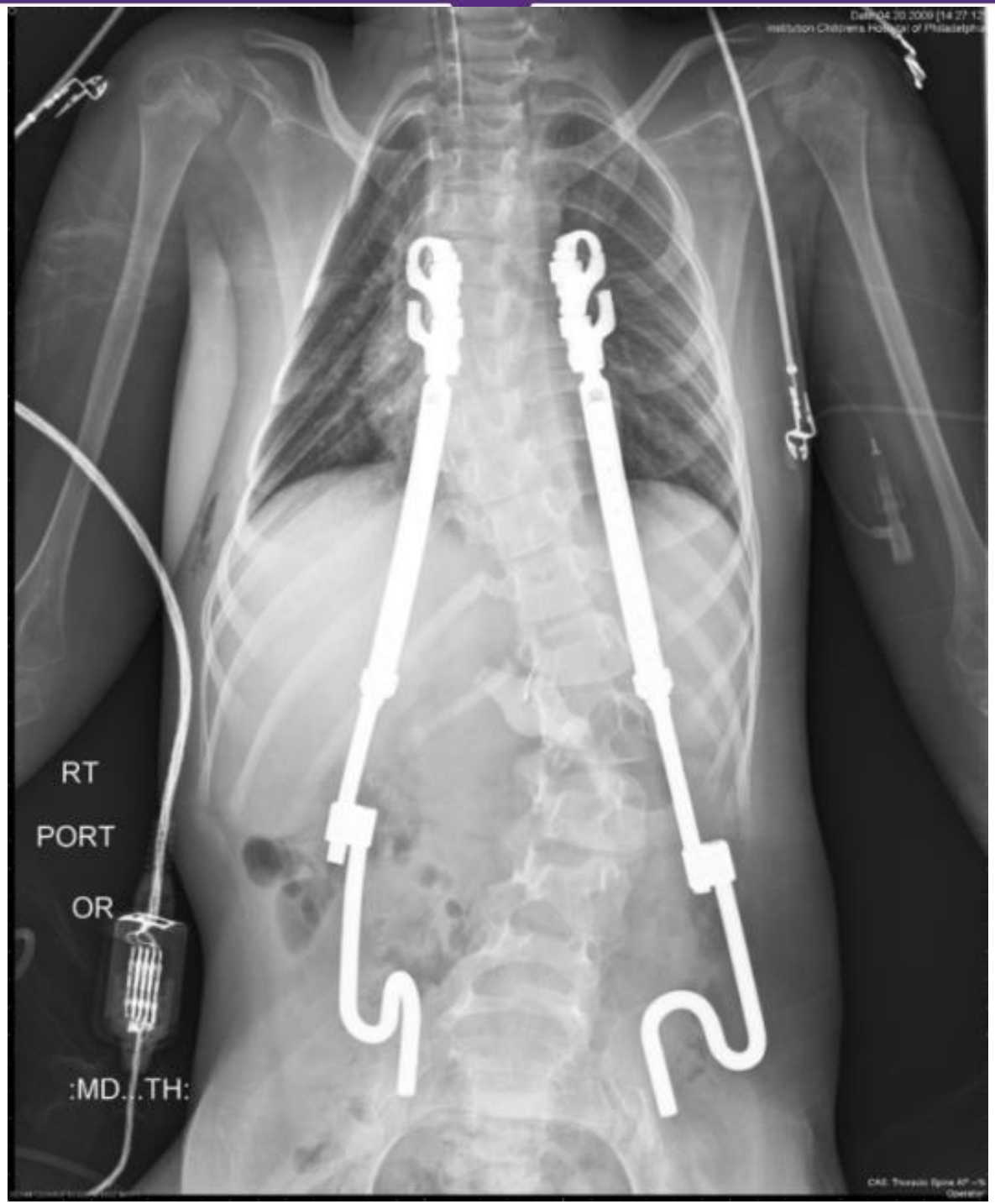
- Careful observation for mild deformity.
- Orthotic management (avoid further constriction of thorax leading to impaired pulmonary function)
- Wheelchair seating systems to maintain sitting posture and accommodate pelvic obliquity.
- Orthoses may slow scoliosis progression; however, discontinue if there is progressive spinal deformity.


vstatx

Surgical Management in Skeletal immaturity (<10 years of age)

- Growing rod constructs without arthrodesis
- Distraction based systems:
- Vertical Expandable Prosthetic Titanium Rib (VEPTR)
- MAGEC Rods
- Guided growth systems:
- Luque trolley
- Shilla

Complications: infection, anchor displacement, laminar fracture, implant prominence, rod failure, premature arthrodesis, multiple surgical procedures.

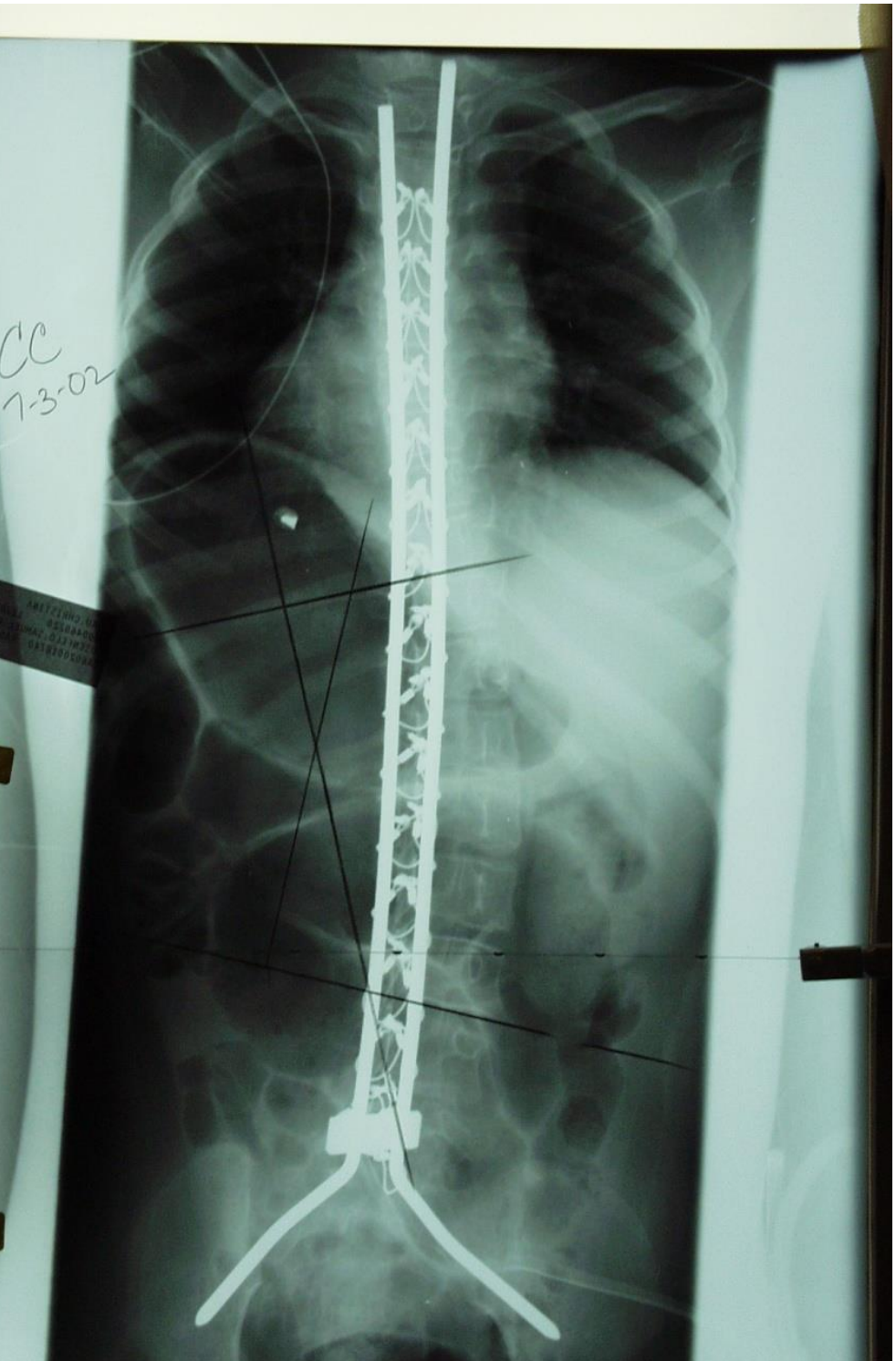

RT
PORT
OR
:MD..TH:

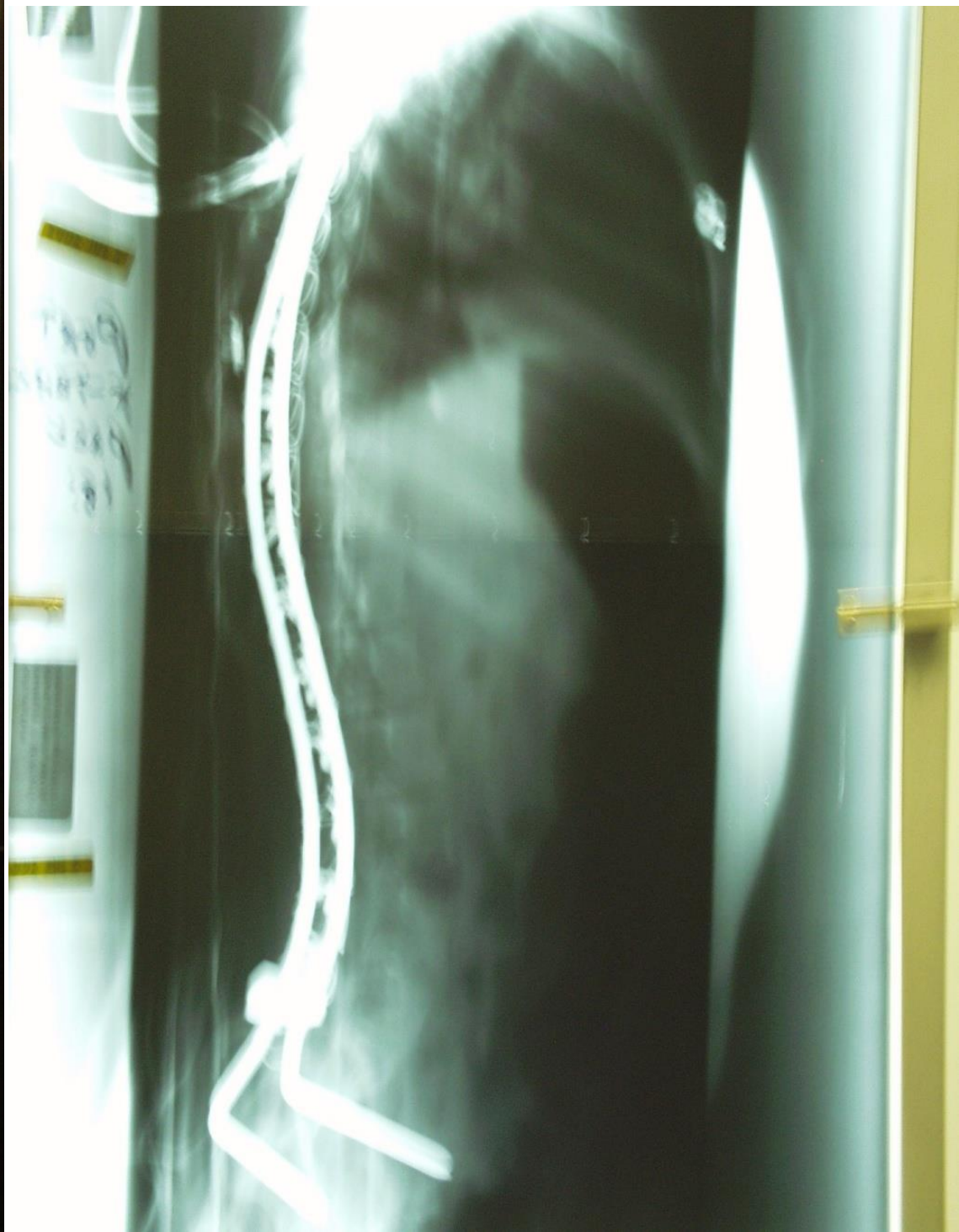
MAGEC Rod


MAGNET
DO NOT BEND
DO NOT BEND
MAGNET
MAGNET
DO NOT RESTERILIZE
MAGNET

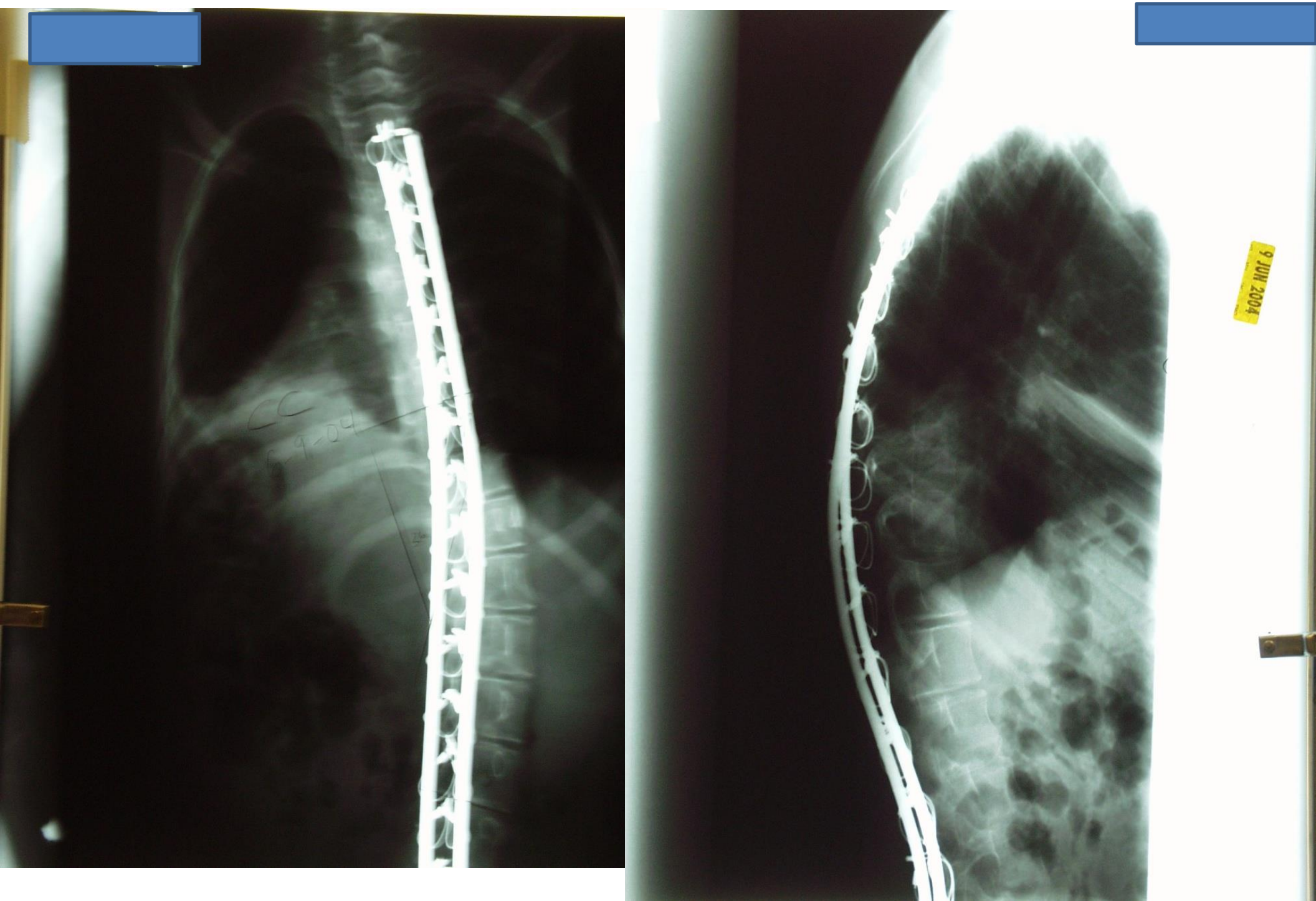
MAGEC Rod

Surgery age 9 years

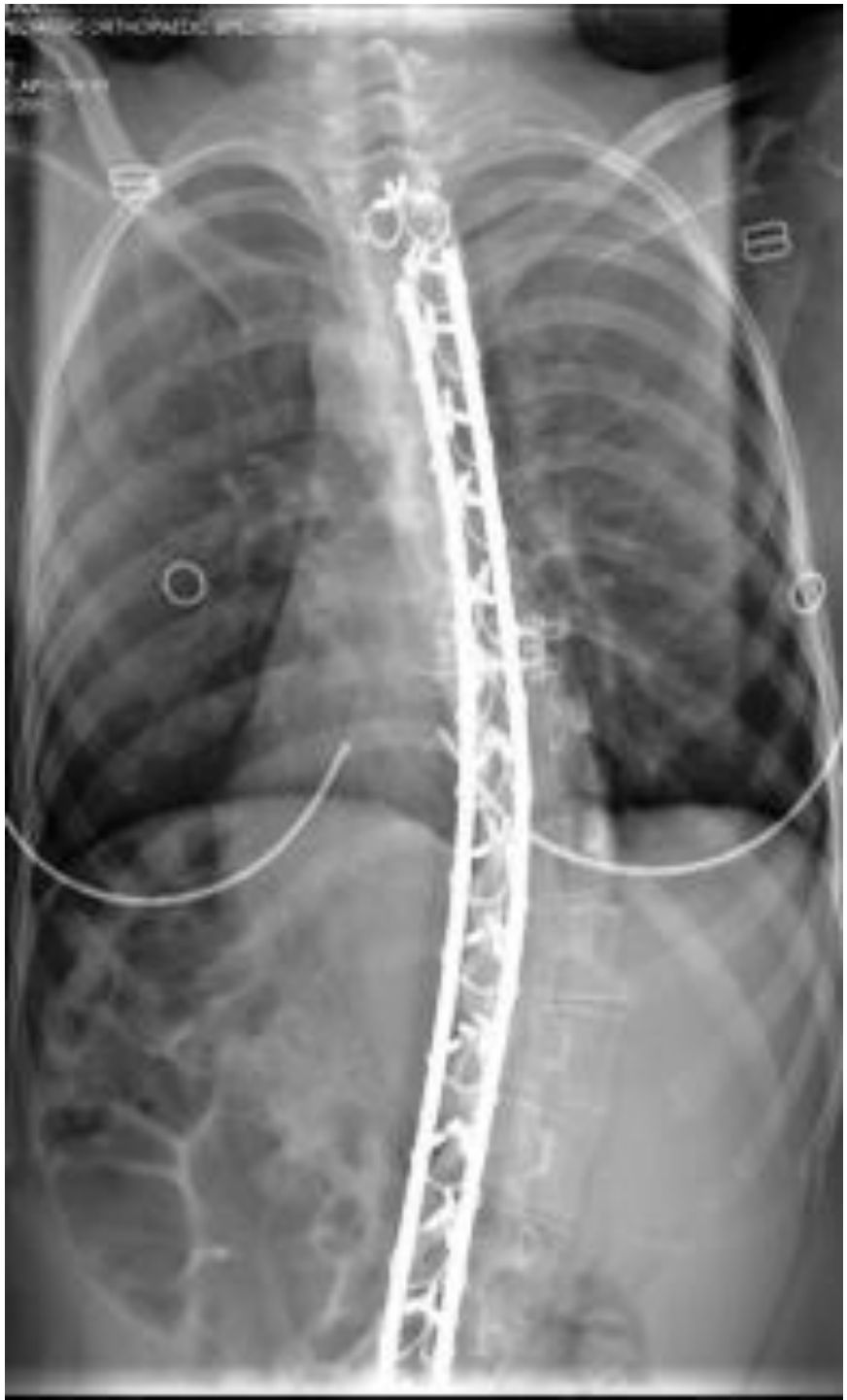




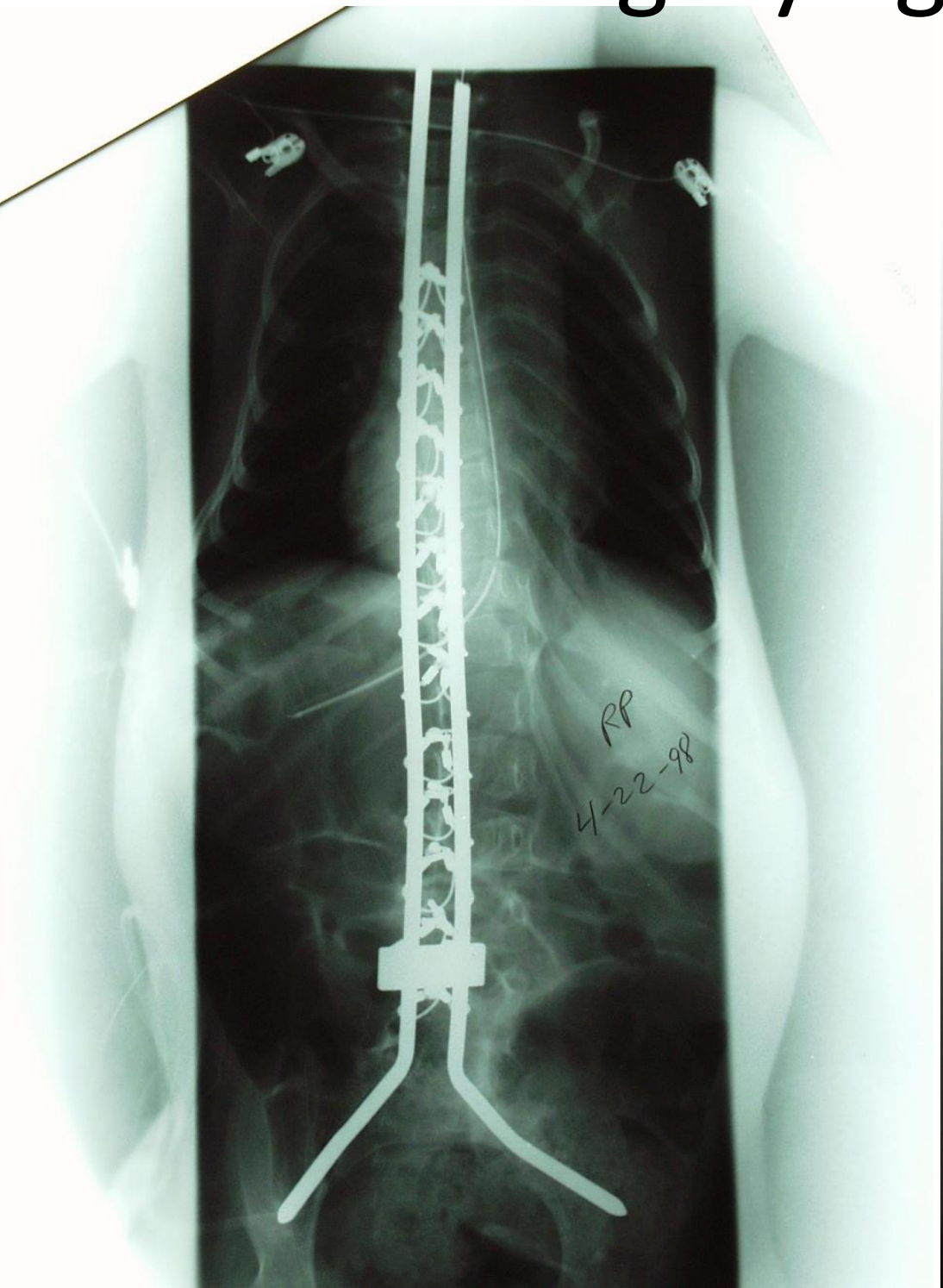
2 years post op

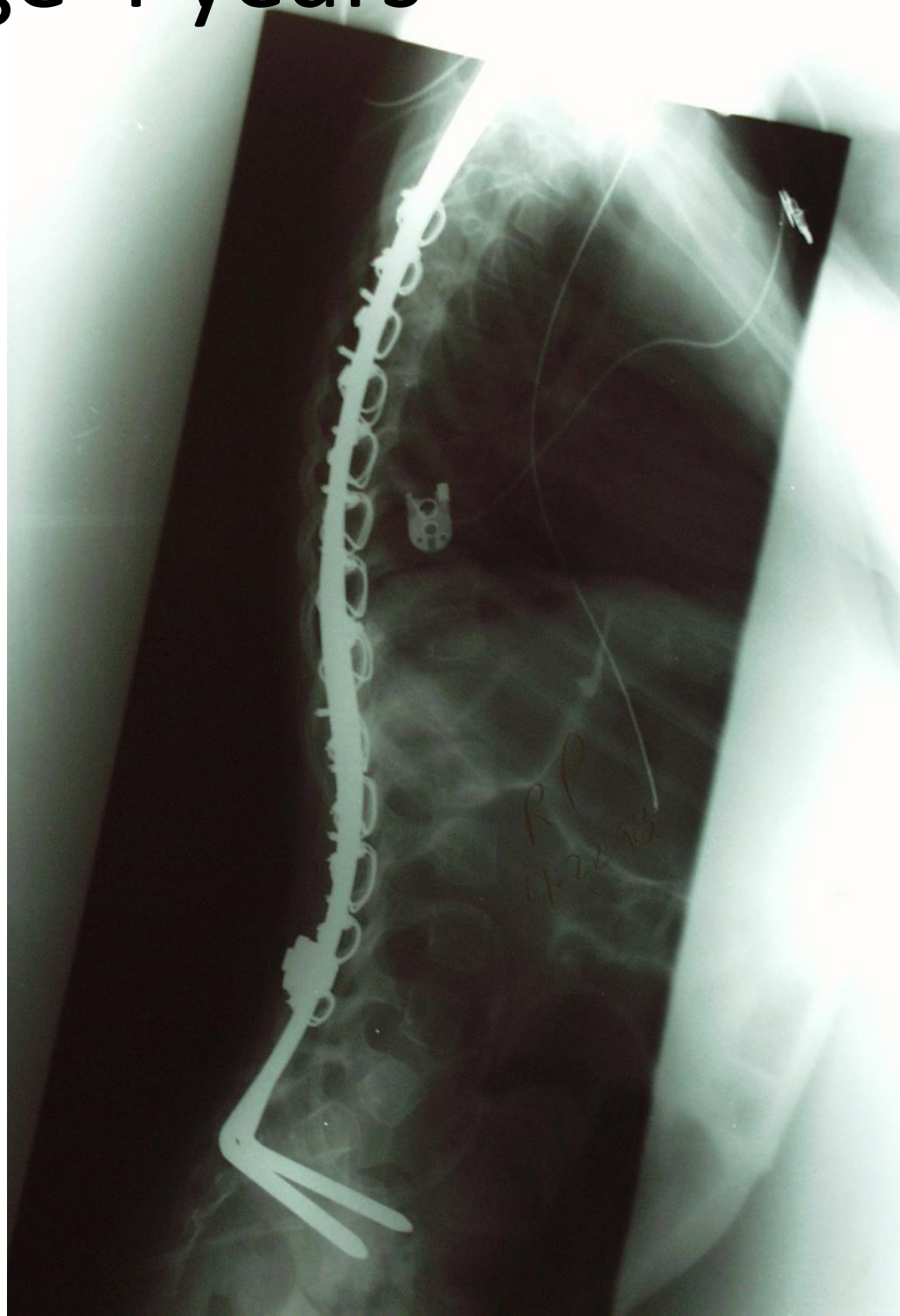


9 years post op

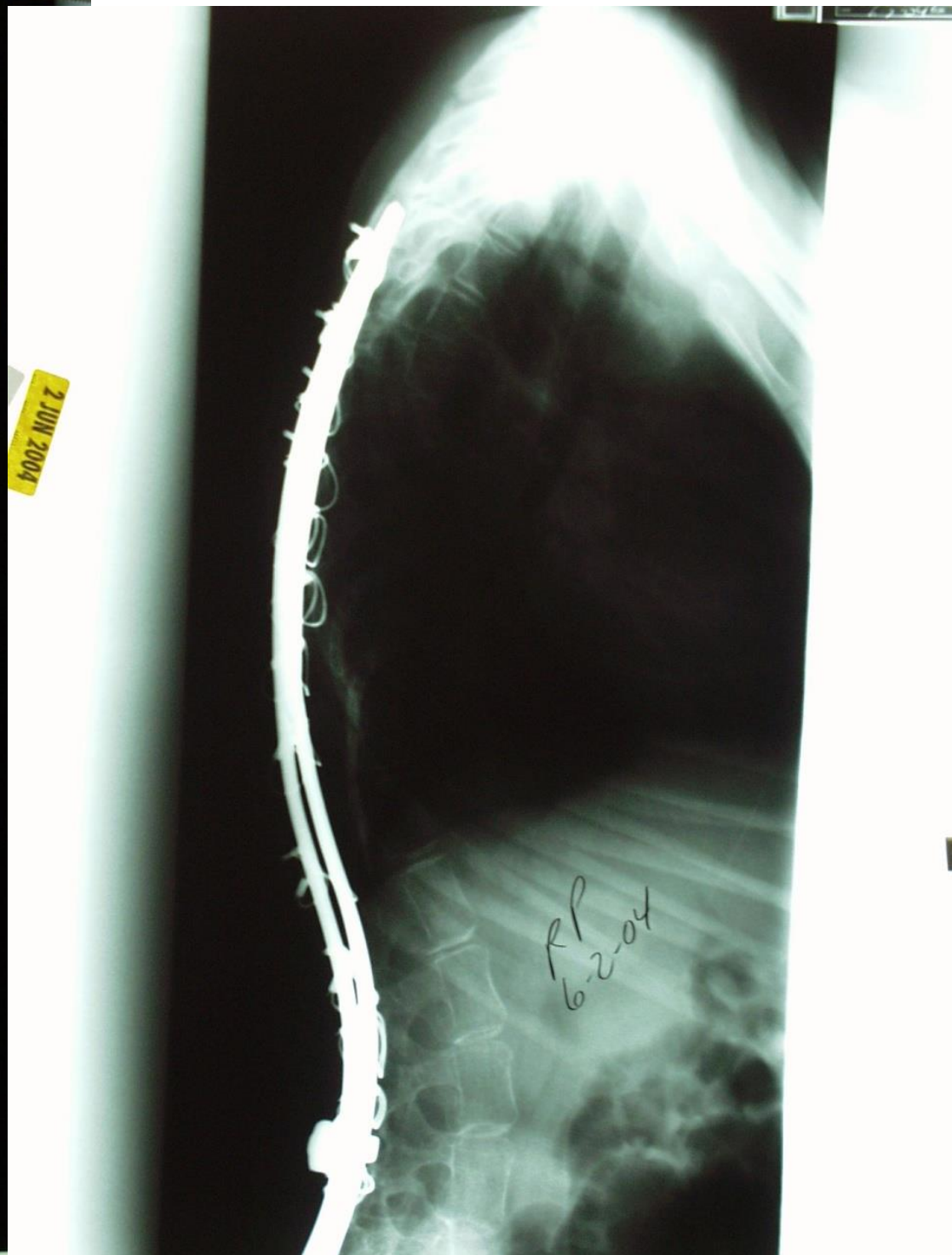


Surgery age 4 years



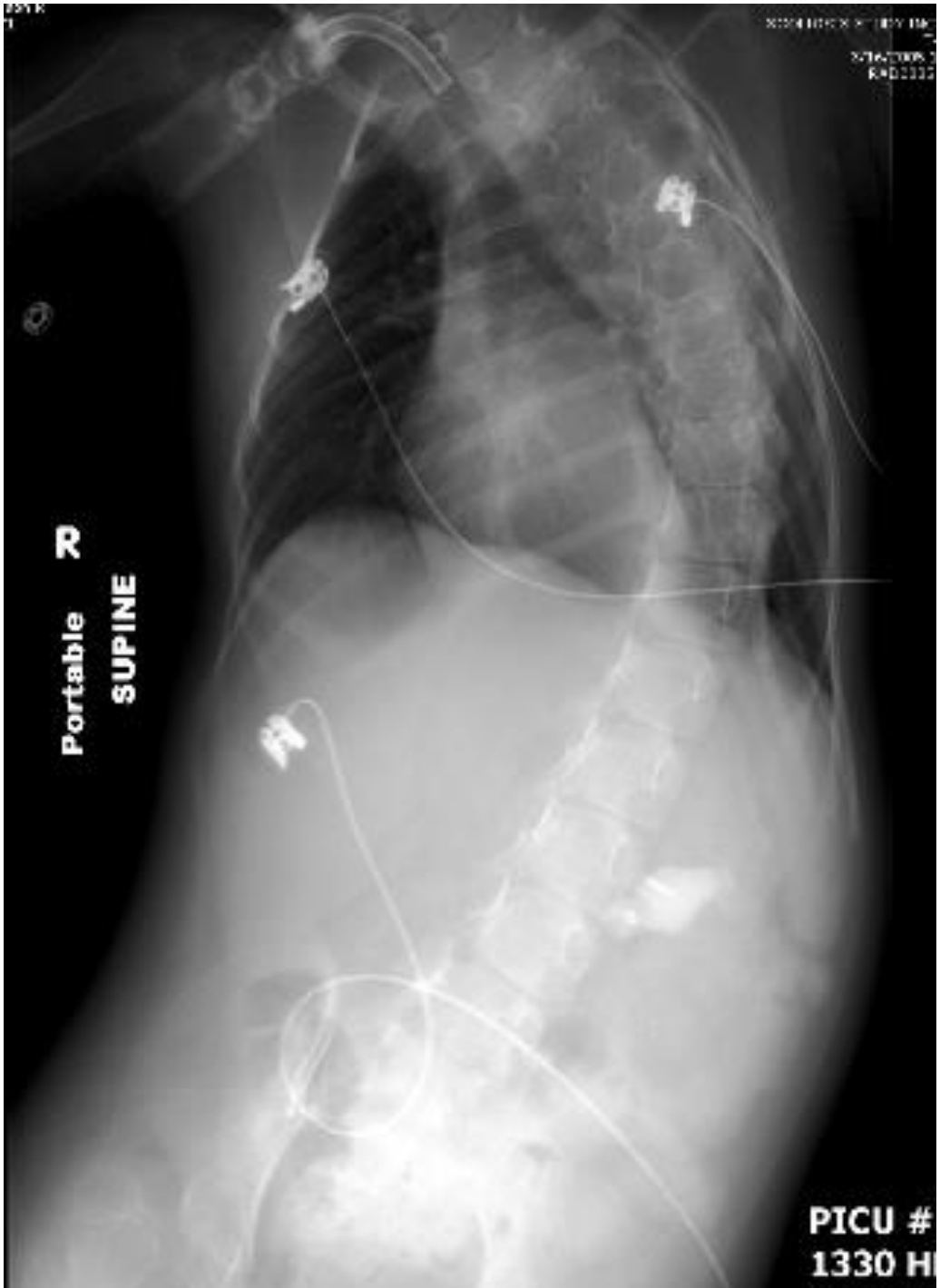


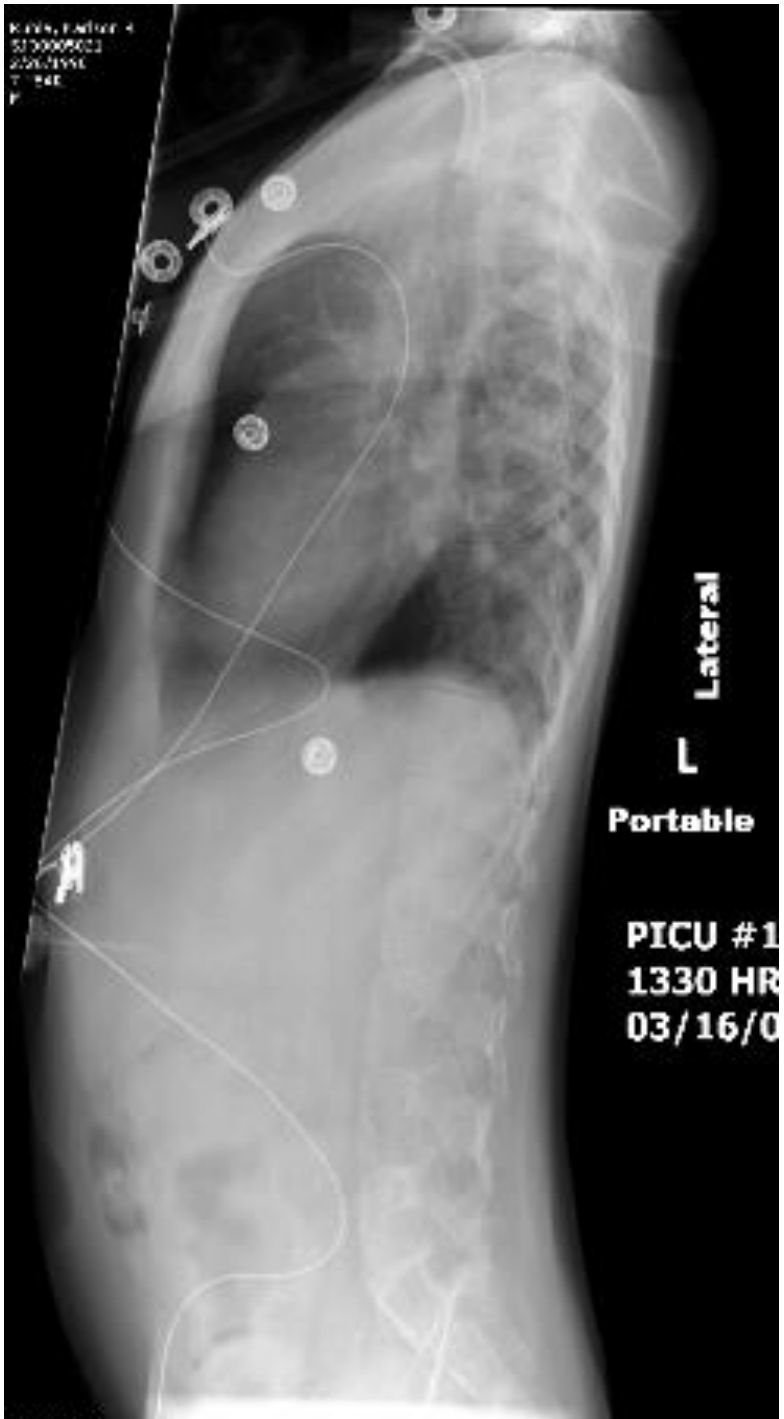
6 years post op



13 years post op

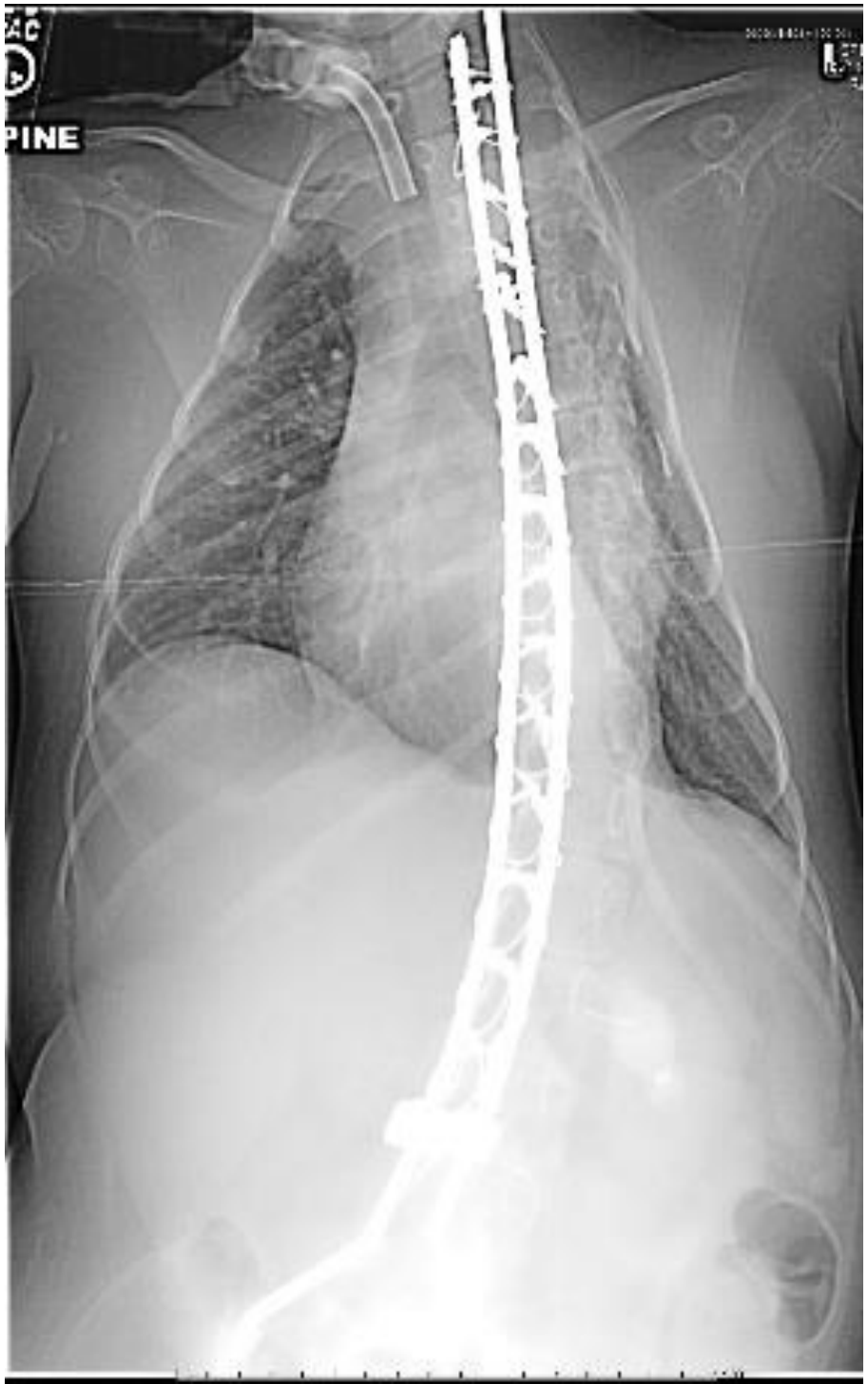
Surgery age 7 years



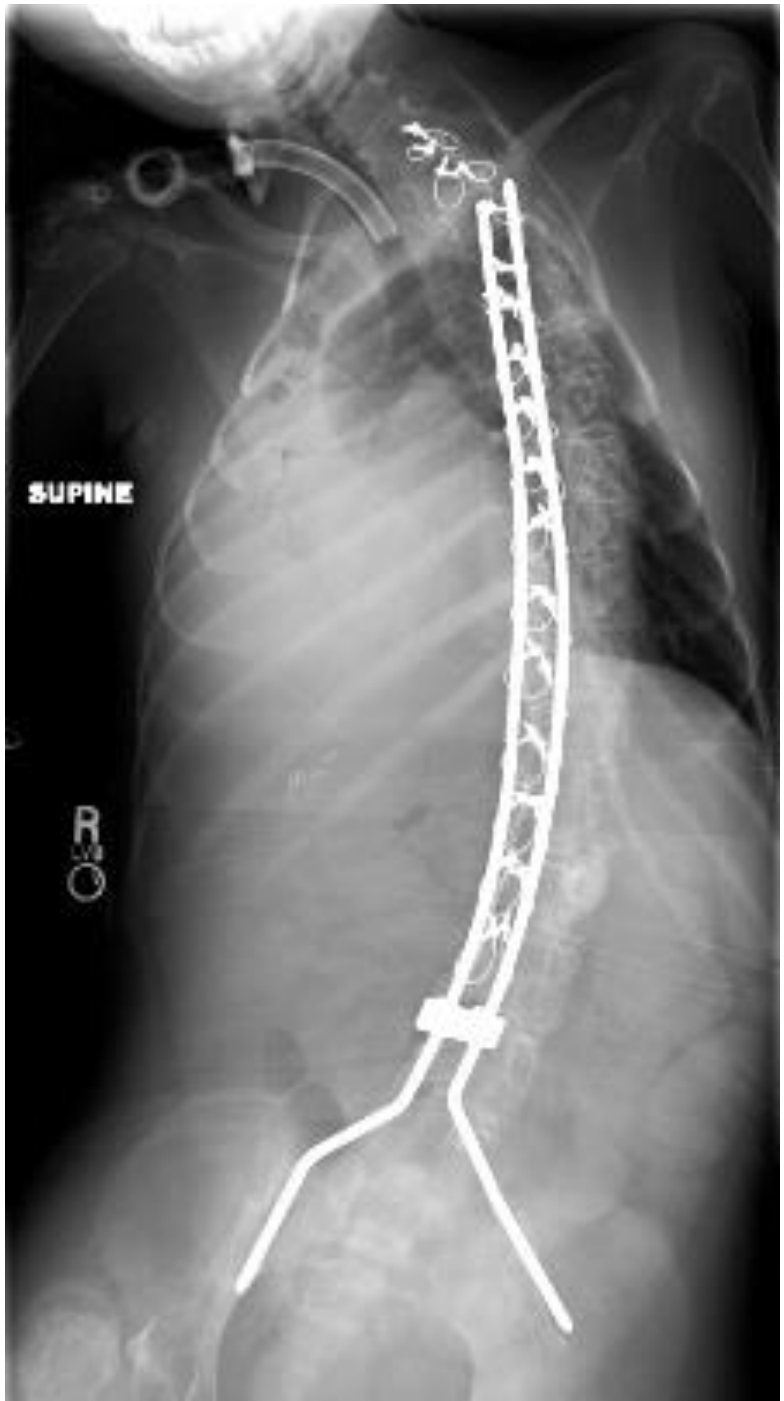


Post op

7 months post op



6 years post op



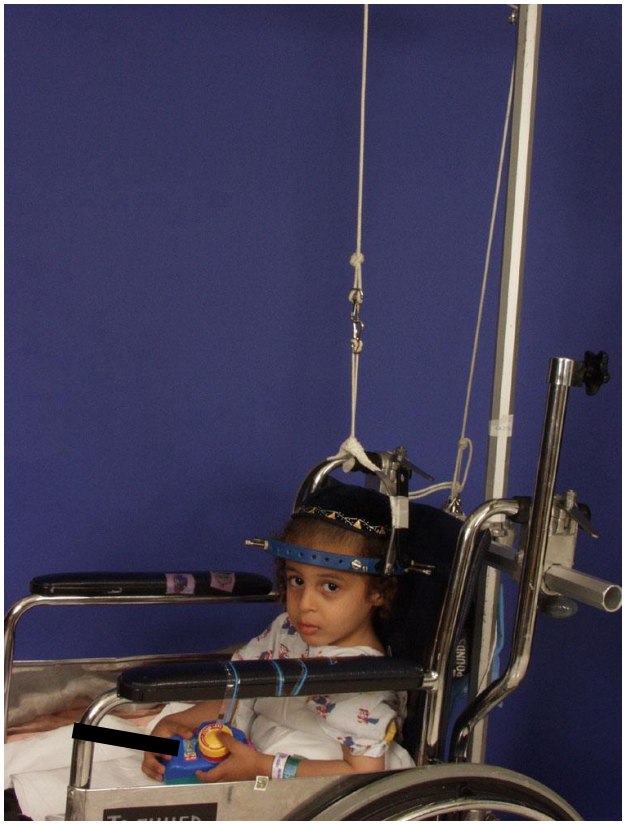
Surgical Management in Skeletal Maturity (>10 years of age)

- Posterior spinal arthrodesis, osteotomies to correct deformity, with segmental spinal instrumentation, pelvic fixation, and autologous / allograft bone graft.

- Complications: pseudoarthrosis, infection, functional deterioration, blood loss / transfusion, implant failure, thromboembolic phenomenon.

Halo Traction

- Large rigid curves where spinal balance cannot be safely obtained via Anterior + Posterior procedure
- Halo-pelvic, Halo-femoral, Halo-gravity
  - ✓Keep head/trunk elevated, sit up
- Traction applied before or between staged anterior and posterior procedure
- Must be able to tolerate traction
  - ✓Normal Cervical spine – no instability
  - ✓Monitor neuro status every shift
    - cranial n (esp Abducens), cervical chain



Efficacy of Perioperative halo-gravity traction in the tratment of severe scoliosis in children. Sink, et al. JPO 2001

Perioperative halo-gravity traction in the treatment of severe scoliosis and kyphosis. Lenke, et al. SPINE 2005

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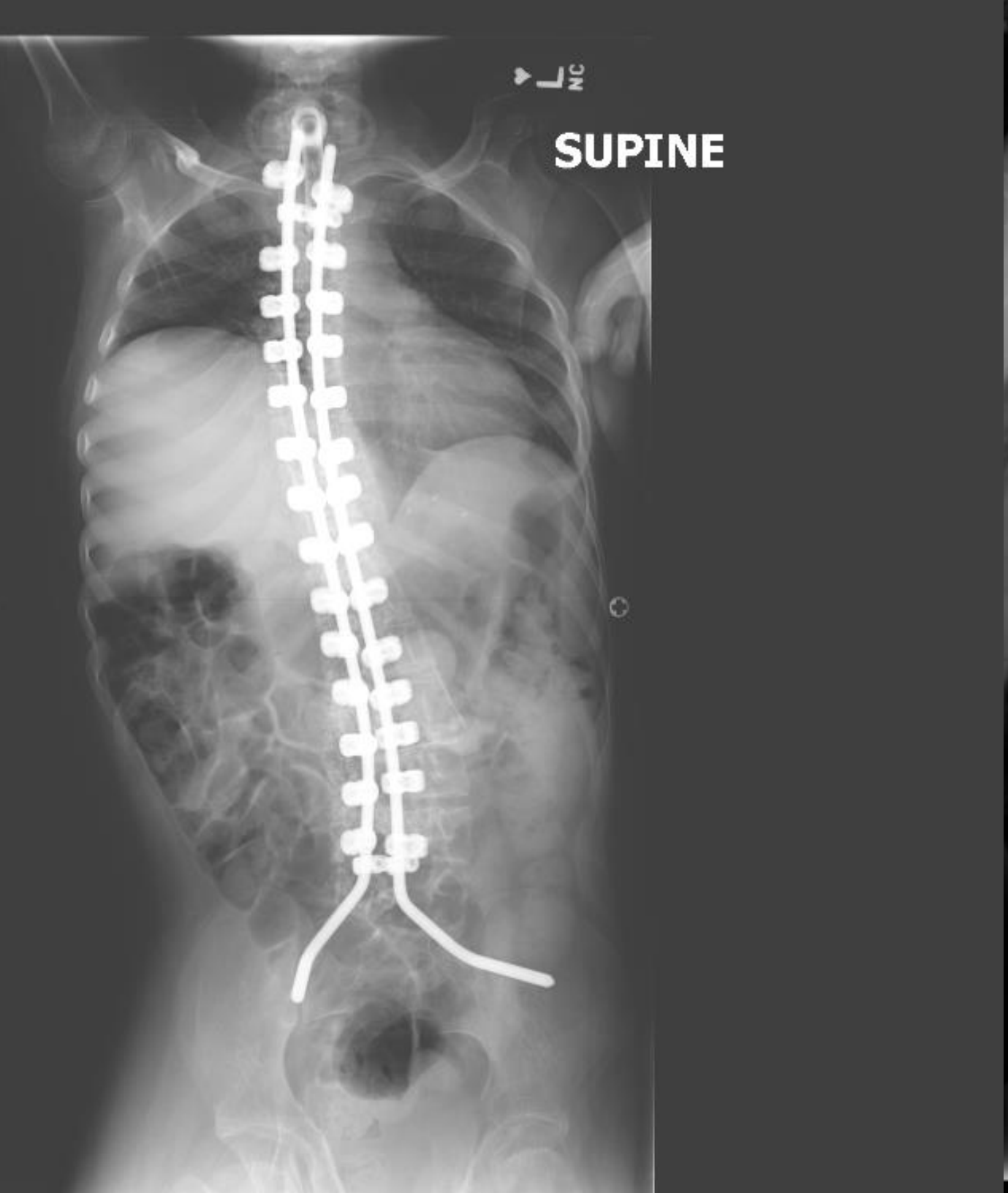
SUPINE


LEFT
NC/TW
SUPIN

Safe Surgery

- Pre-op pulmonary / cardiology evaluation
- Total intravenous anesthetic technique
- Potassium supplementation
- Replace blood loss
- Cell-Saver
- Aminocaproic acid / Tranexamic acid
- Thromboembolic prophylaxis
- Steroid prep
- Malignant Hyperthermia Precautions

Spinal Cord Monitoring

- Somatosensory evoked potentials
- Motor evoked potentials
- EMG

Autologous Blood Tranfusion

- **Pre donation**
- **Cell-saver**
- **Constavac reinfusion**

Pulmonary management / intervention

- Volume recruitment
- Ventilators
- Tracheostomy
- Mechanical insufflator / exsufflator
- Mucus mobilization devices
- Pneumococcal, influenza immunizations

Cardiac Management

- Evaluation: ECG, ECHO, Holter
- Intervention: angiotensin-converting-enzyme inhibitor (ACE inhibitor) i.e. enalapril
- Beta-blockers (carvedilol)

Gastroenterology / Nutrition

- Swallowing evaluation
- Diet control
- Supplementation
- Gastrostomy
- Pharmacologic
- Constipation management
- GERD management

Dietary supplements

- Calcium citrate (better absorbed than Calcium carbonate) age 5 to 10
  up to 600 mg./day age 11 to adult
  more than 1300 mg./day (in divided dosage)
- Vitamin D3 (better absorbed than D2) age 5 to 10 at least 800 I.U./day age 11 to adult over 5000 I.U./day

Wheelchair Indications

- Prevent muscle fatigue
- Appropriate seating system
- Part-time use for long distance mobility; encourage short distance ambulation and transfers

Wheelchair Specifications

- Rigid seat and back
- Jay or Roho seating systems
- Appropriate trunk support, head control
- Tilt-in-space >> reclining
- Power assist modifications / controls
- Accommodate ventilatory support and growth adjustments

SMA Conference Survey

Please complete your conference survey at this link:

https://www.surveymonkey.com/s/2016AnnualSMAConference

Or fill out the paper survey in your conference folder.

- **All participants who complete a survey by 10:30 am on Sunday June 19th, will have their name entered into a raffle for a brand new iPad!**
- **The winners will be drawn and announced on Sunday, June 19th at the Closing General Session/It's a Wonderful Life.**

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