Management of Spinal Deformities in Spinal Muscular Atrophy

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Make today a breakthrough.

Disclosure

- Consultant MediCrea Spine
- Consultant OrthoPediatrics 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.

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- 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.

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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

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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.

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v.sta tx

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.

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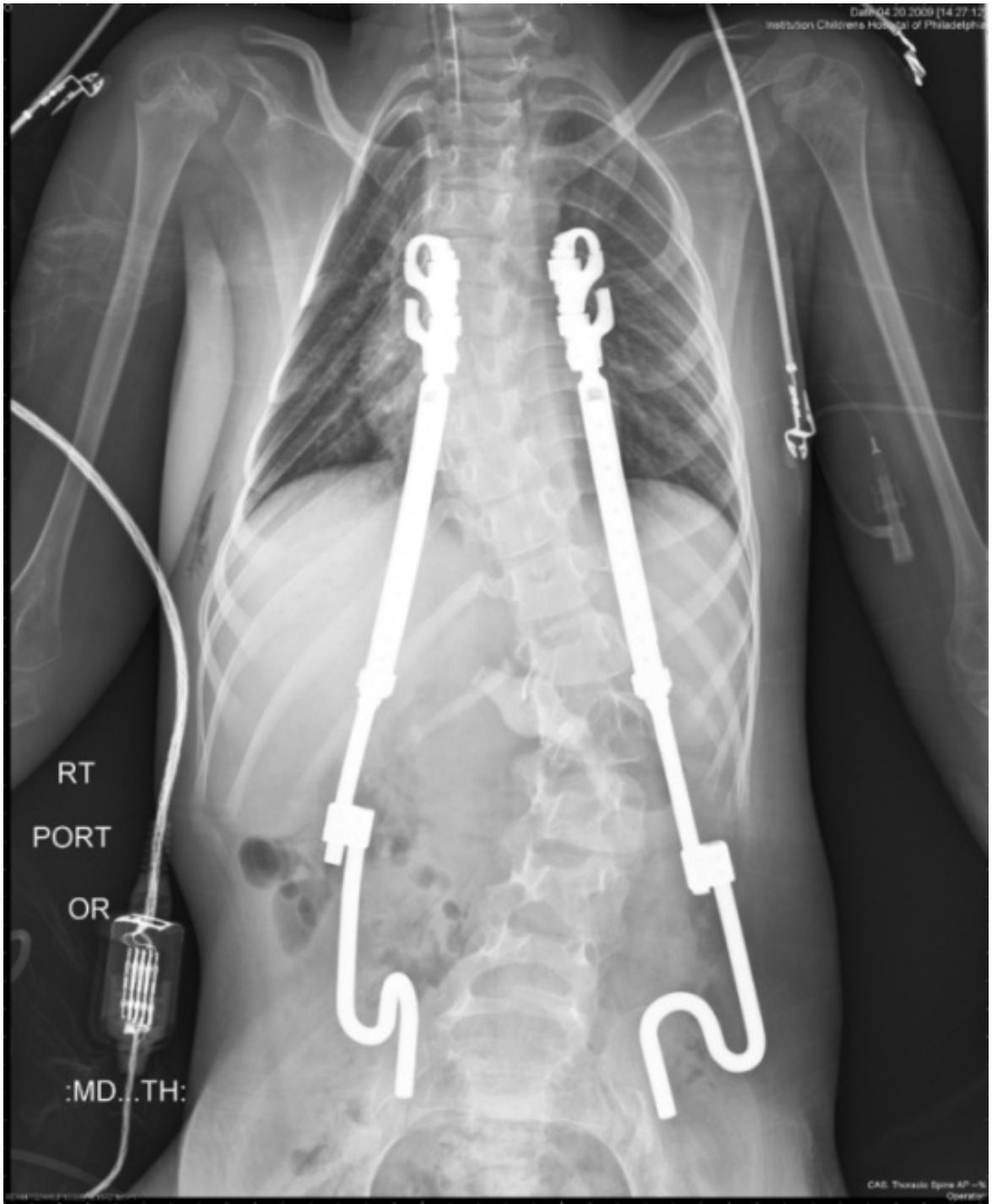
RT
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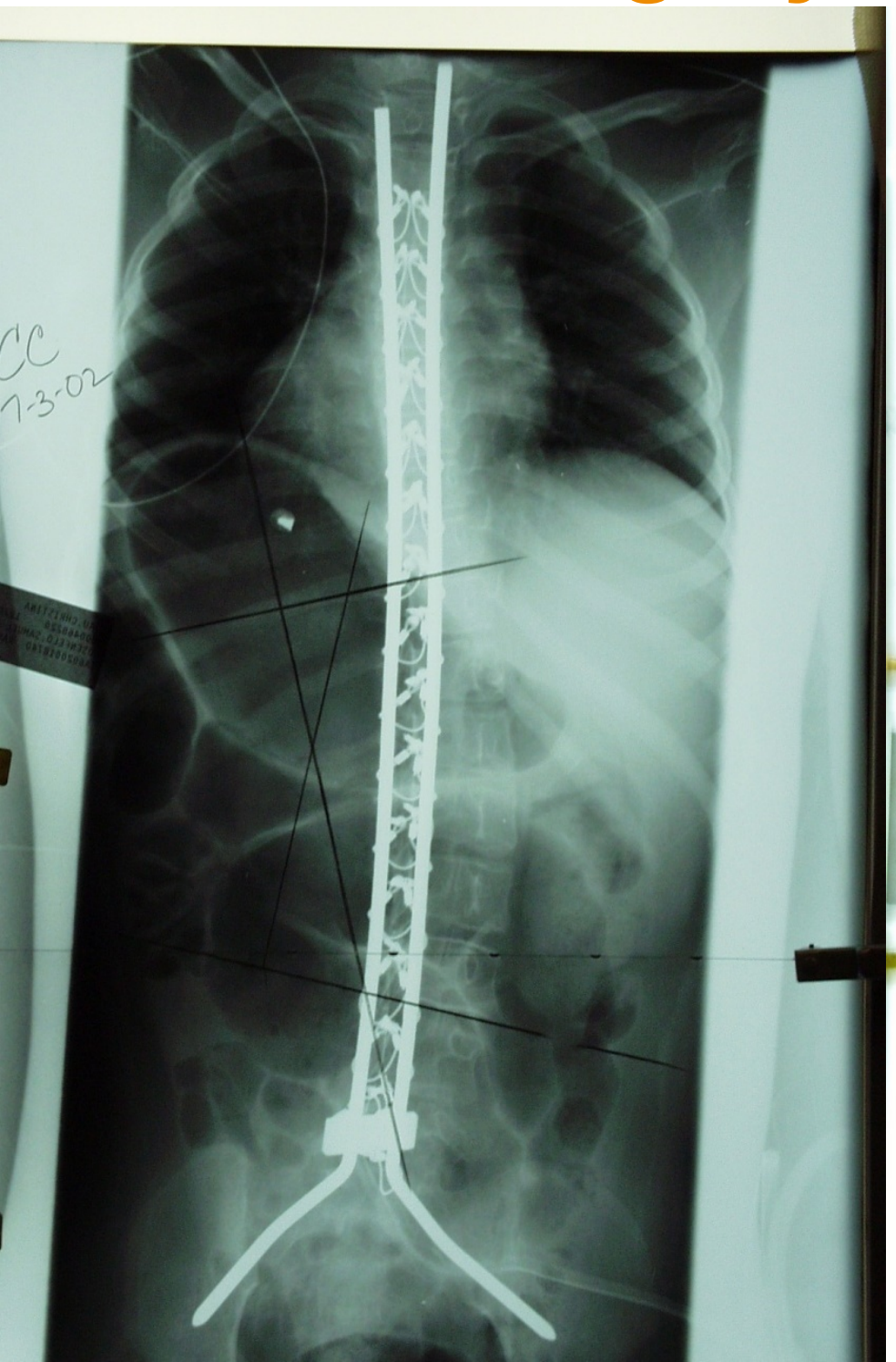
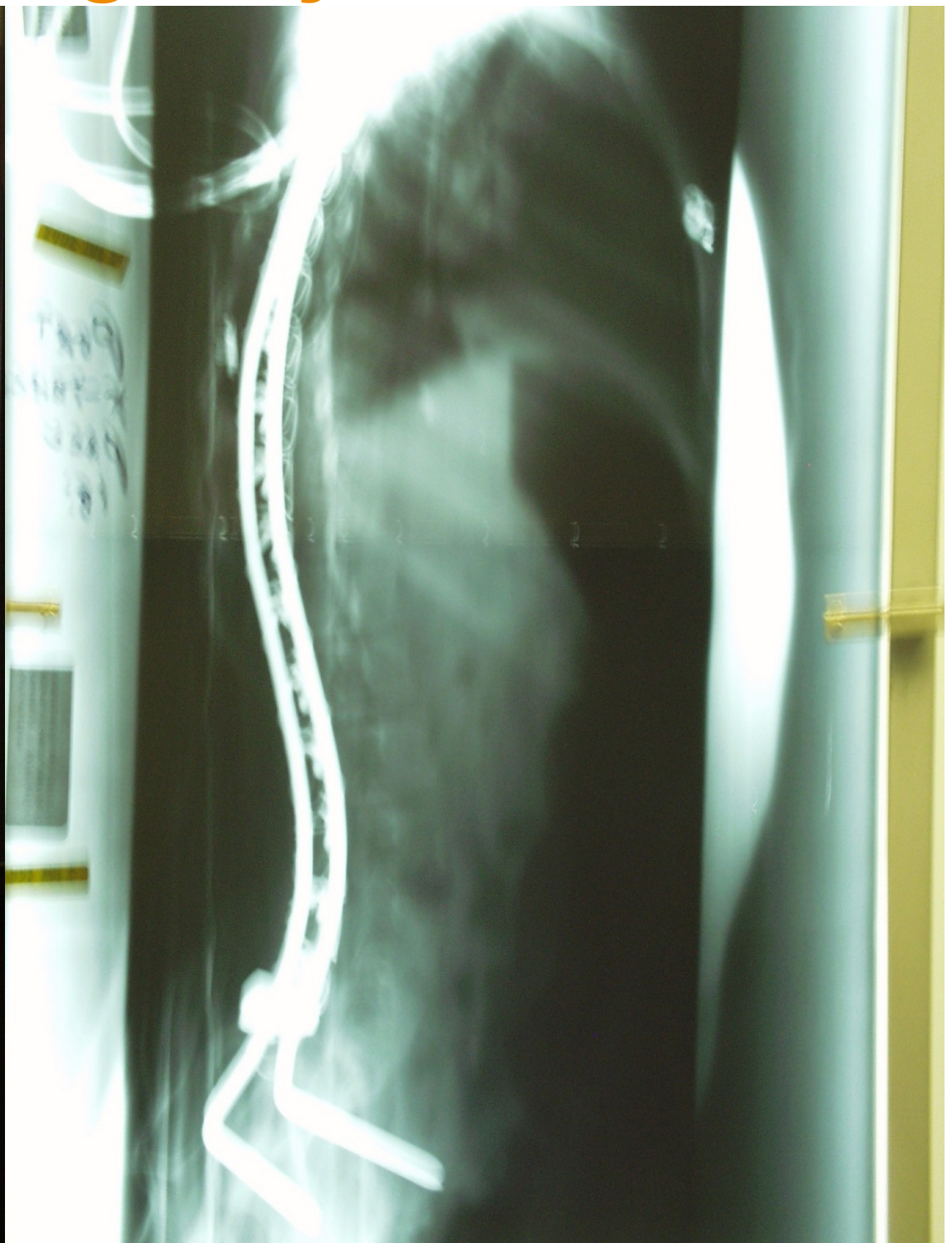
MAGEC Rod

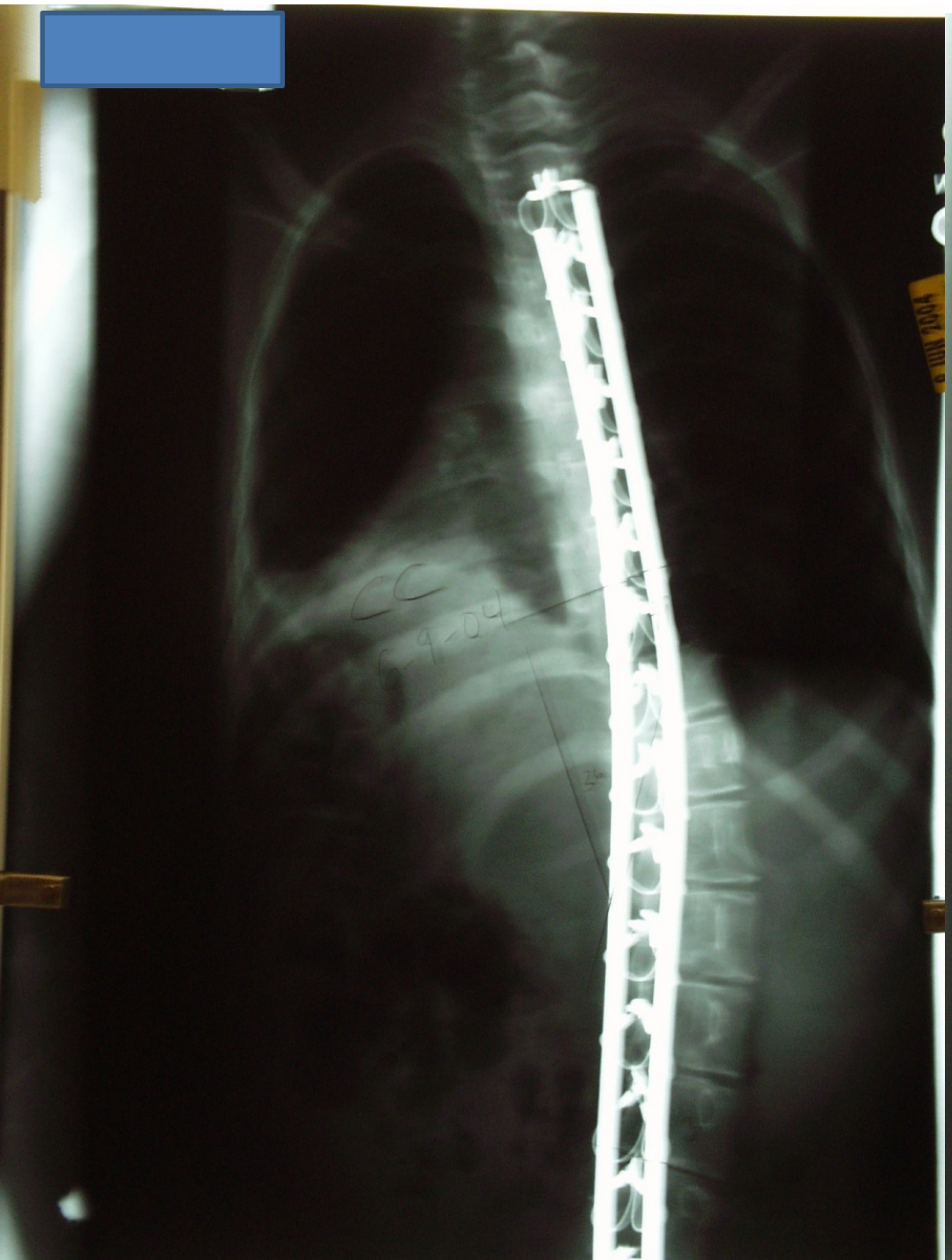
MAGEC Rod

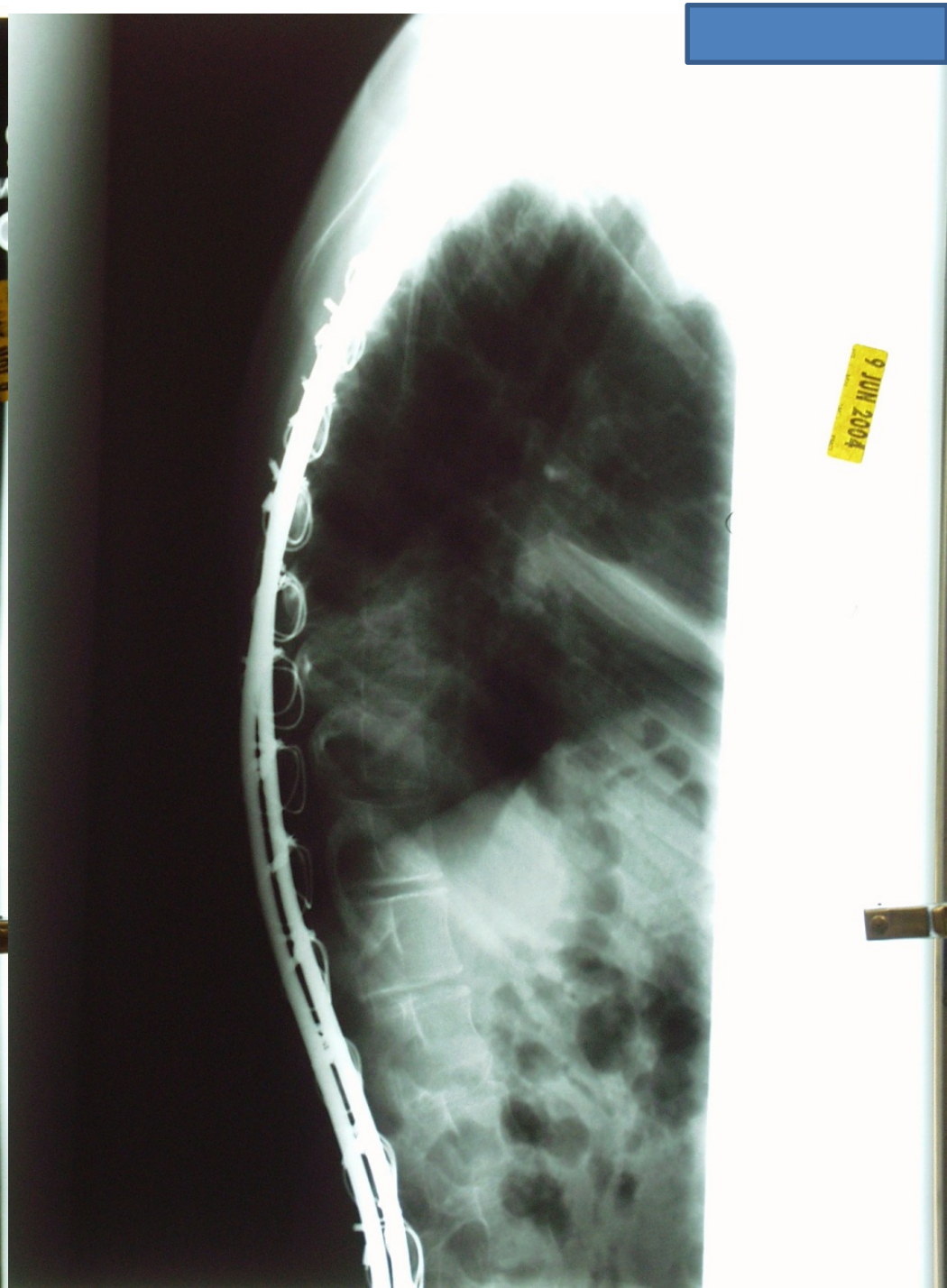
Surgery age 9 years



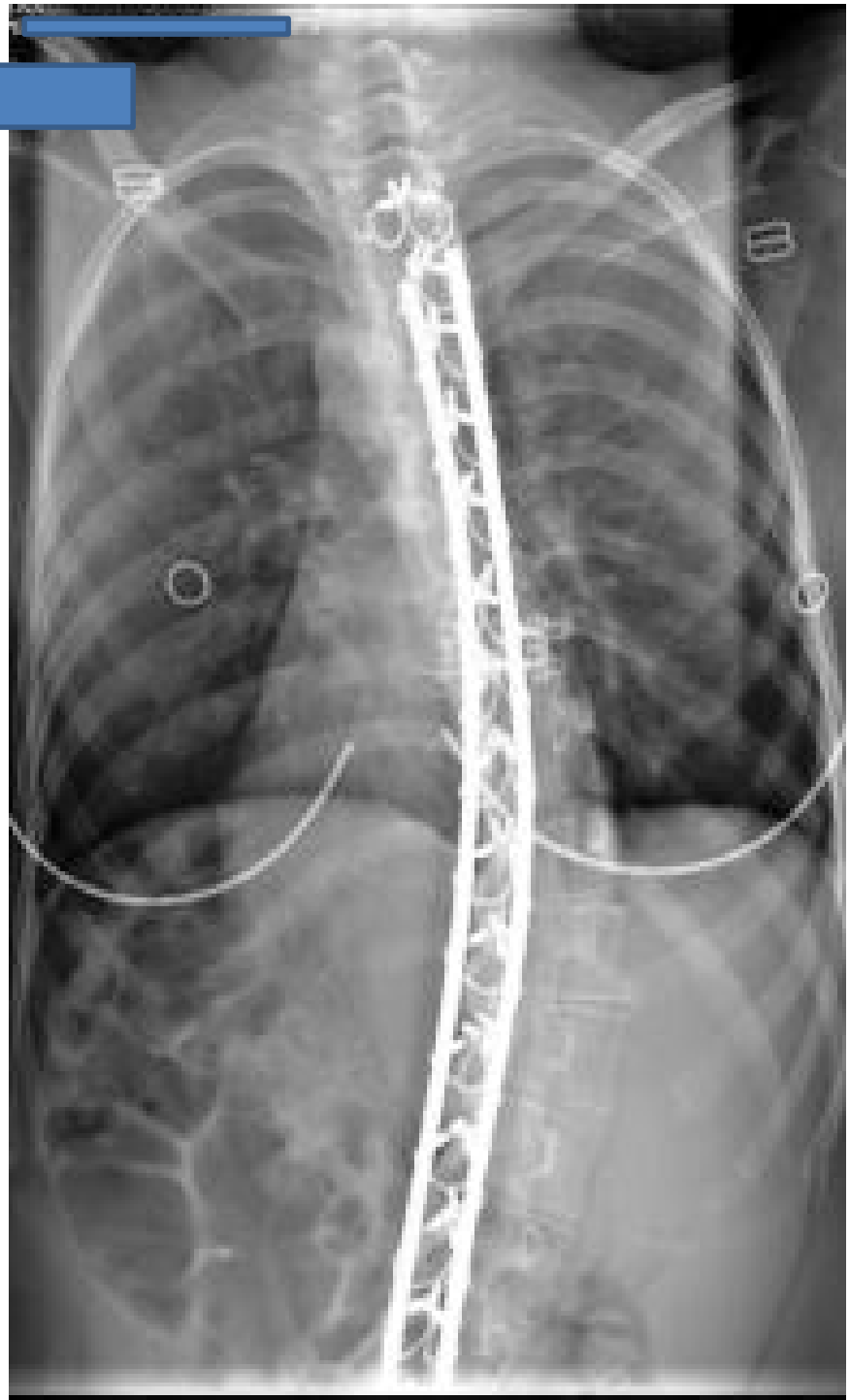


2 years post op

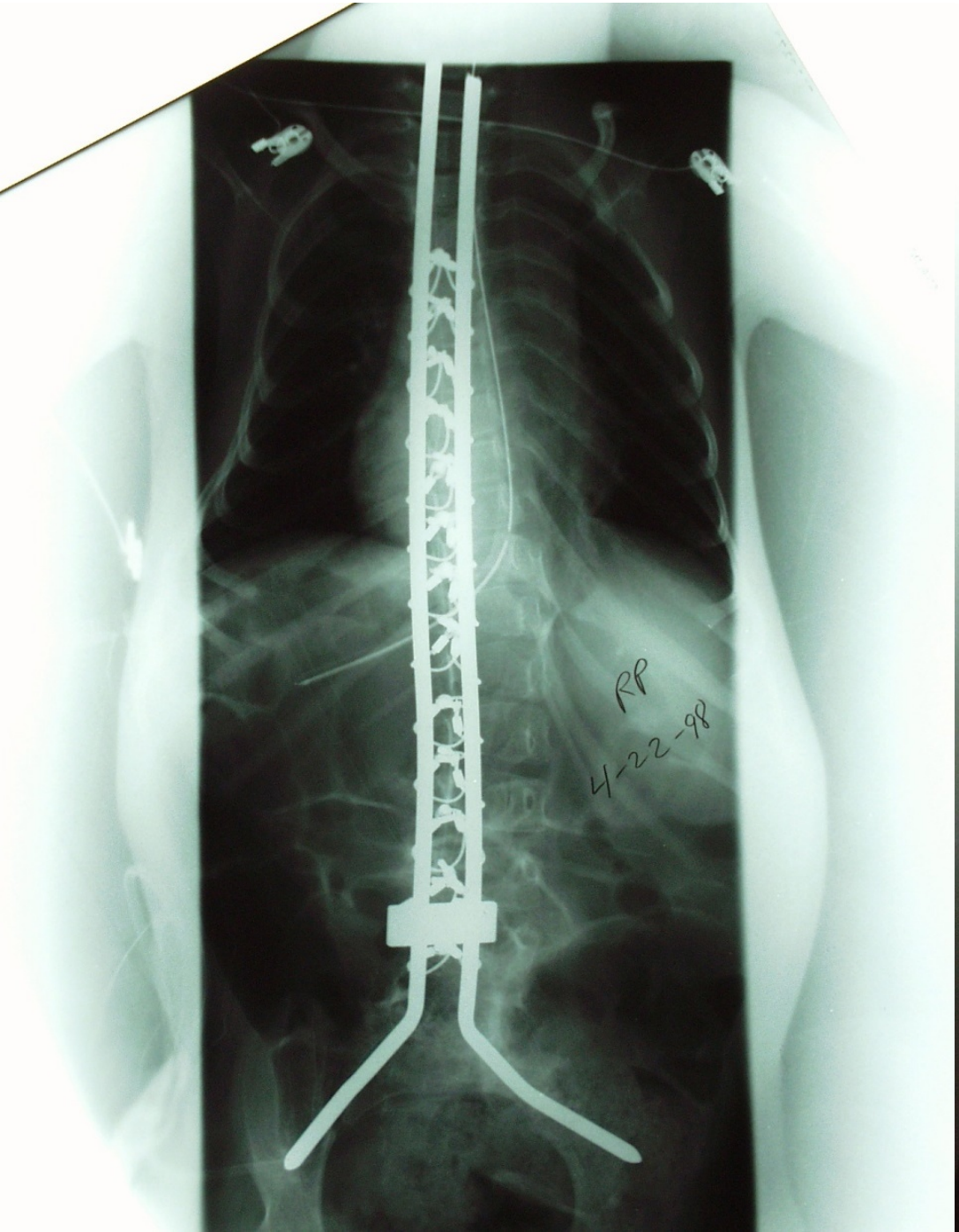




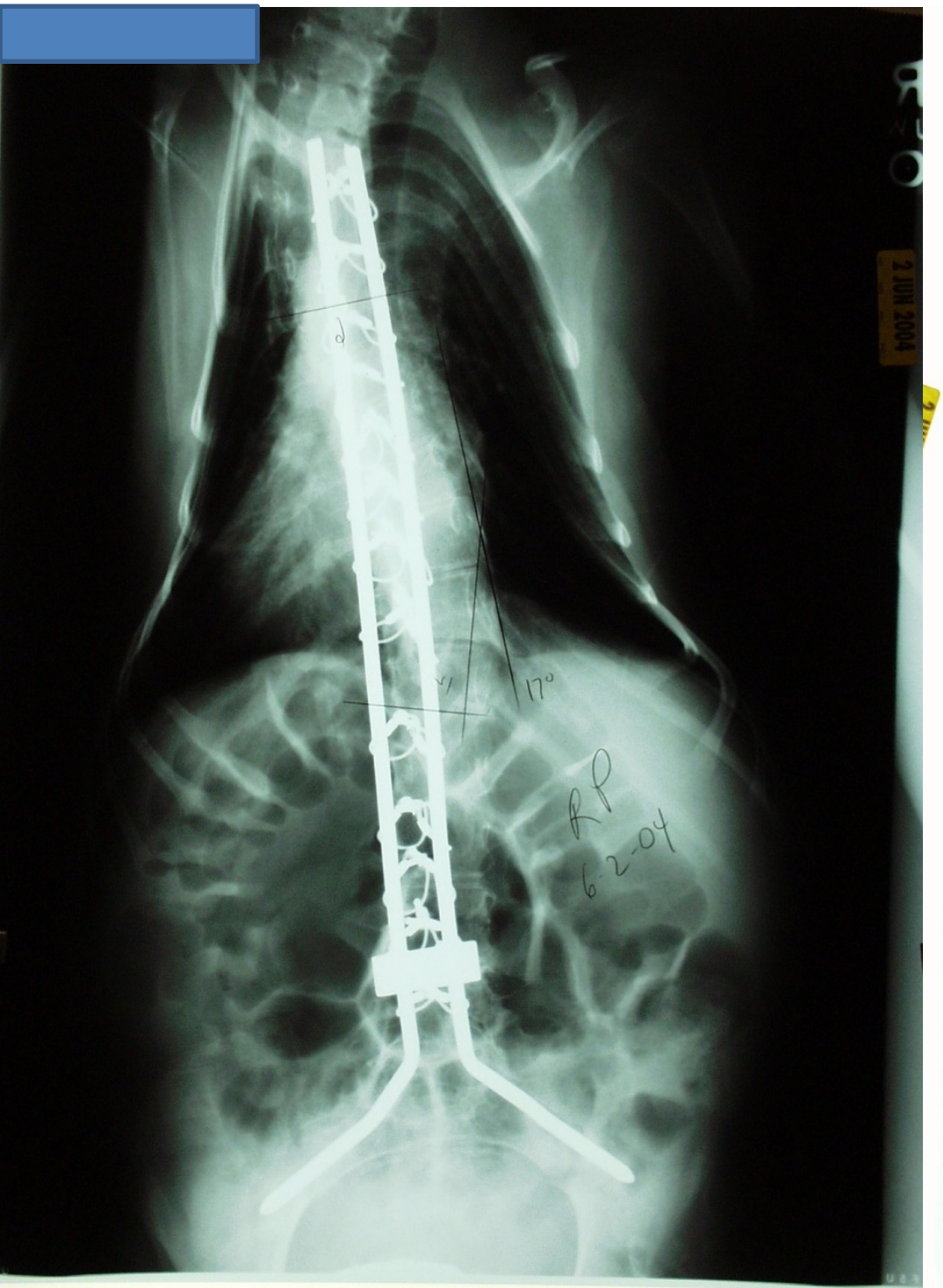
9 years post op

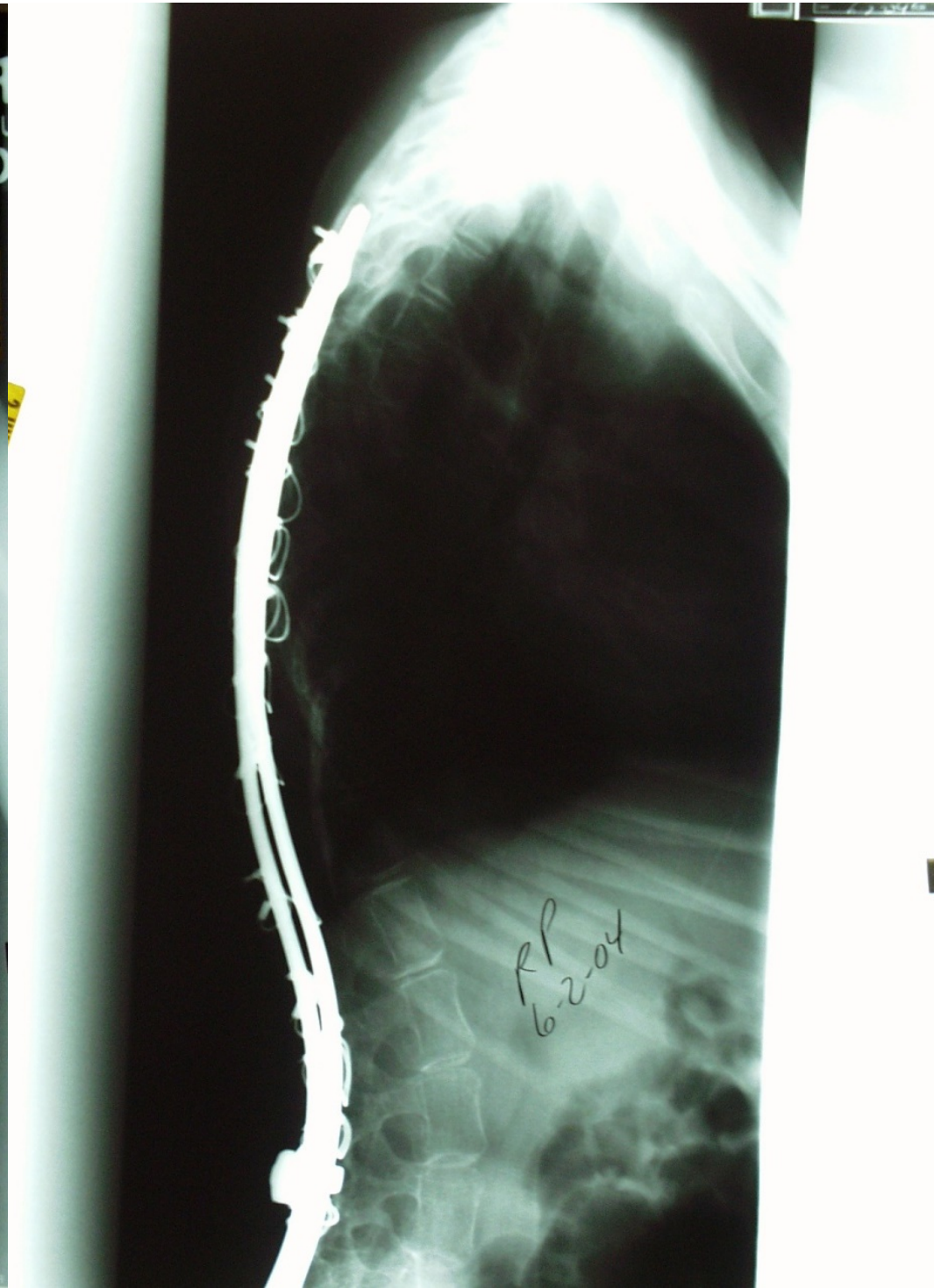


Surgery age 4 years



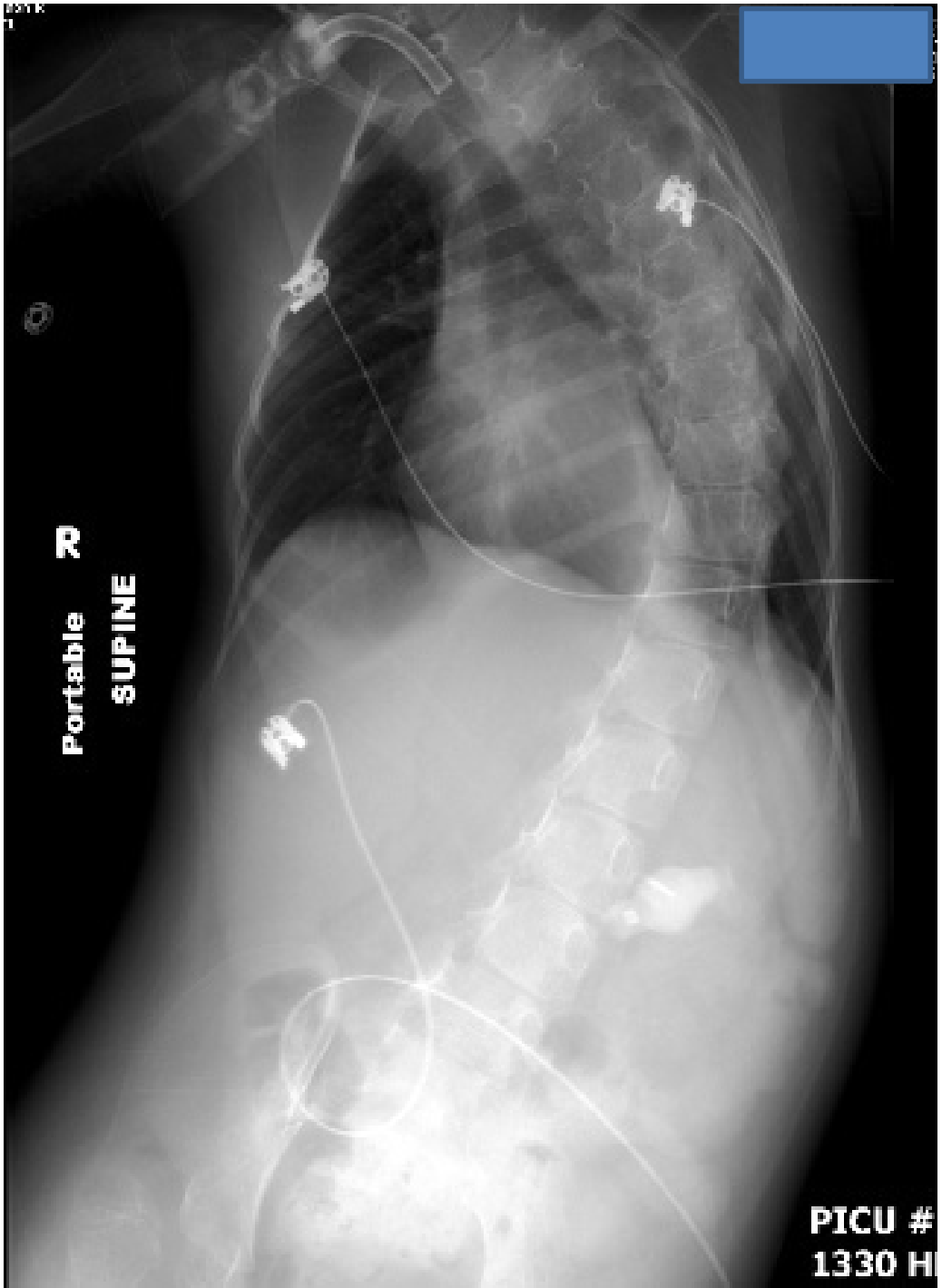
6 years post op

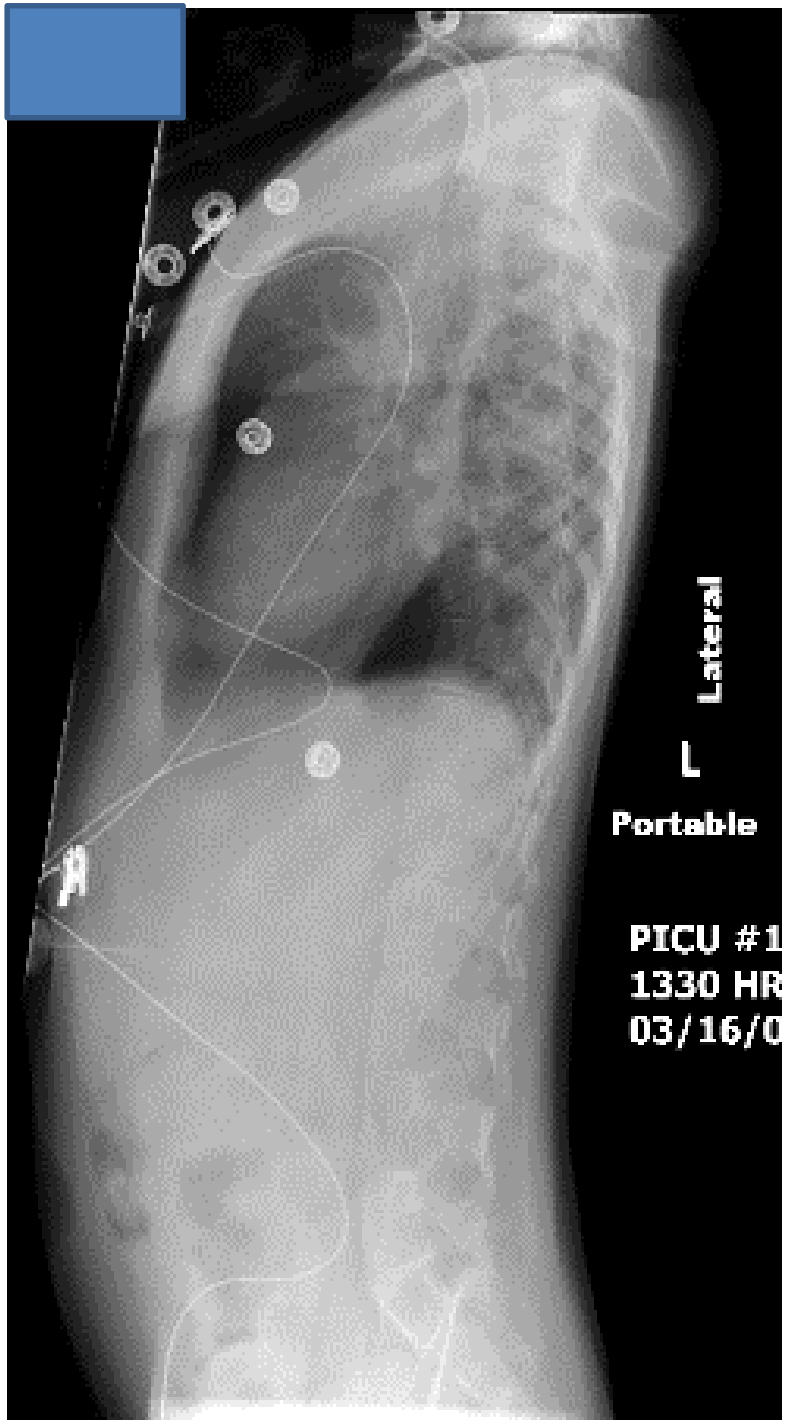


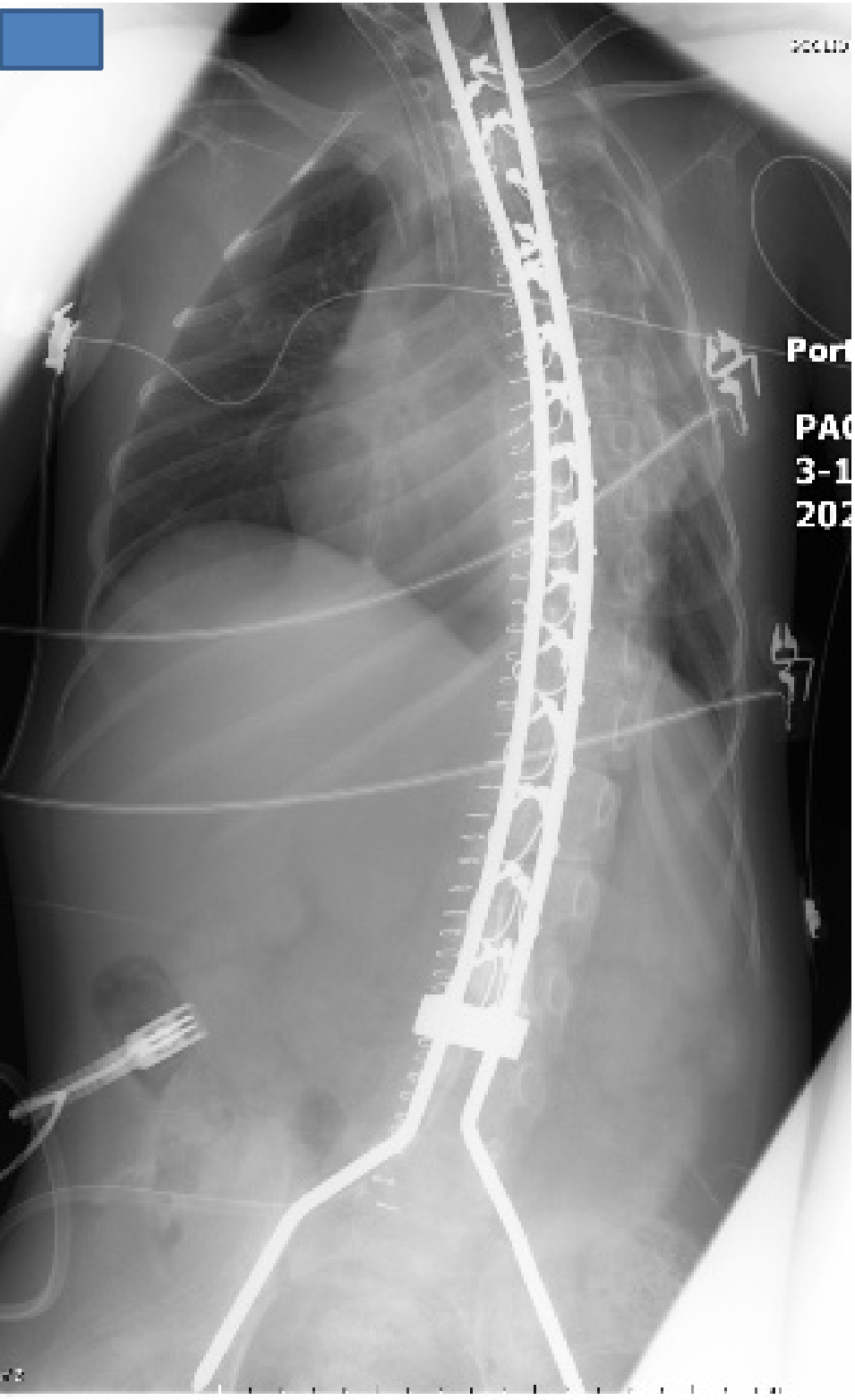


13 years post op

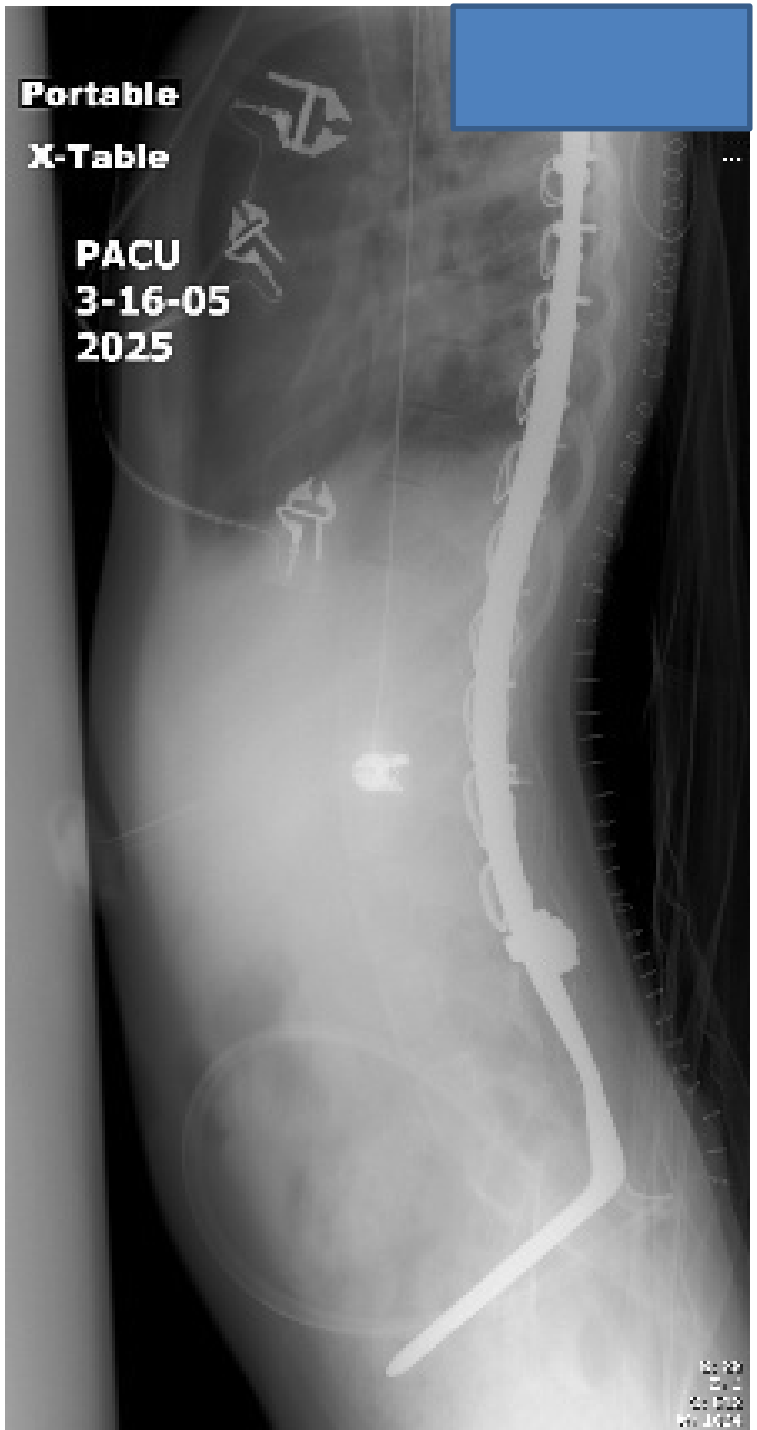
Surgery age 7 years



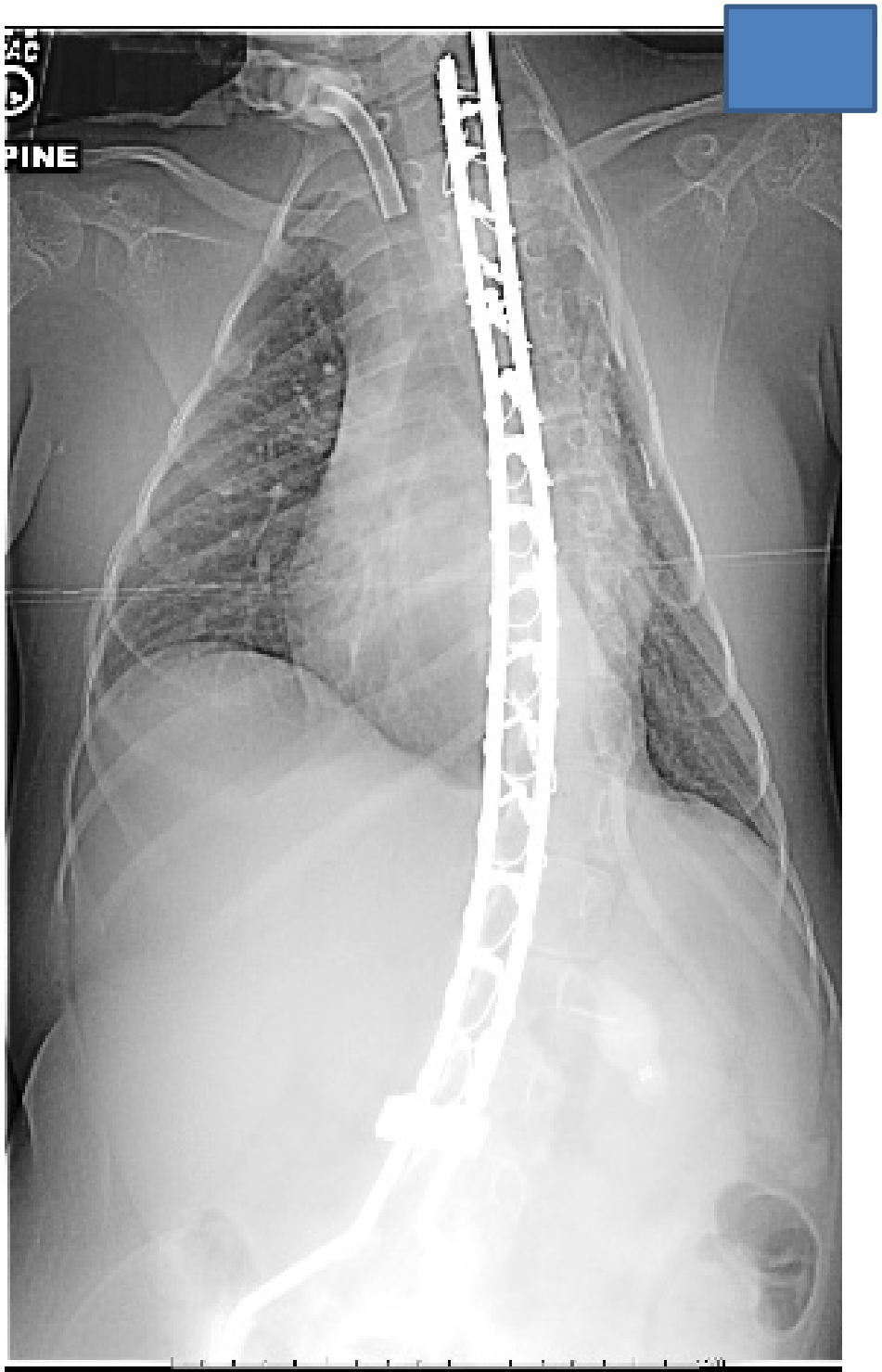


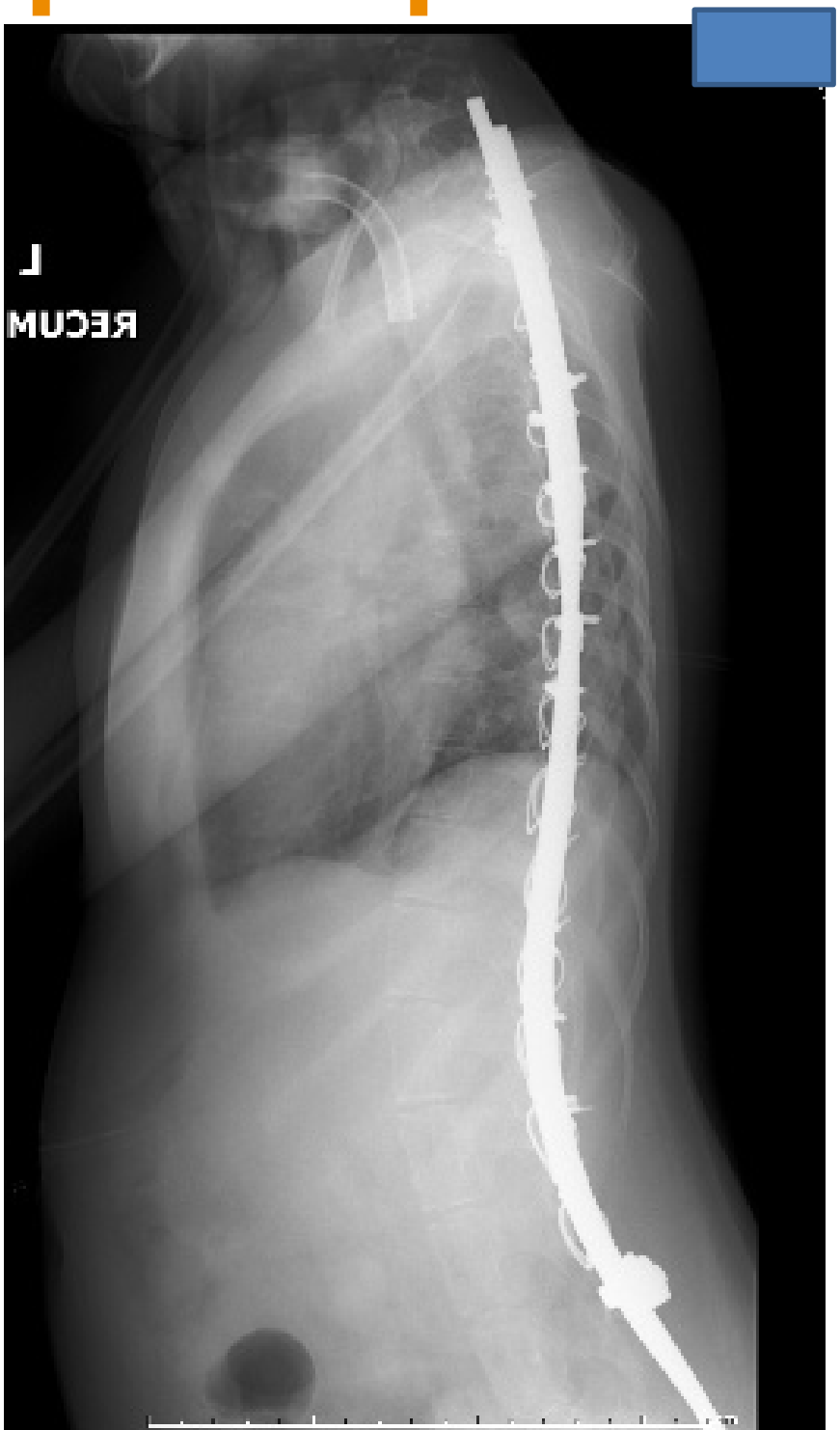


Post op

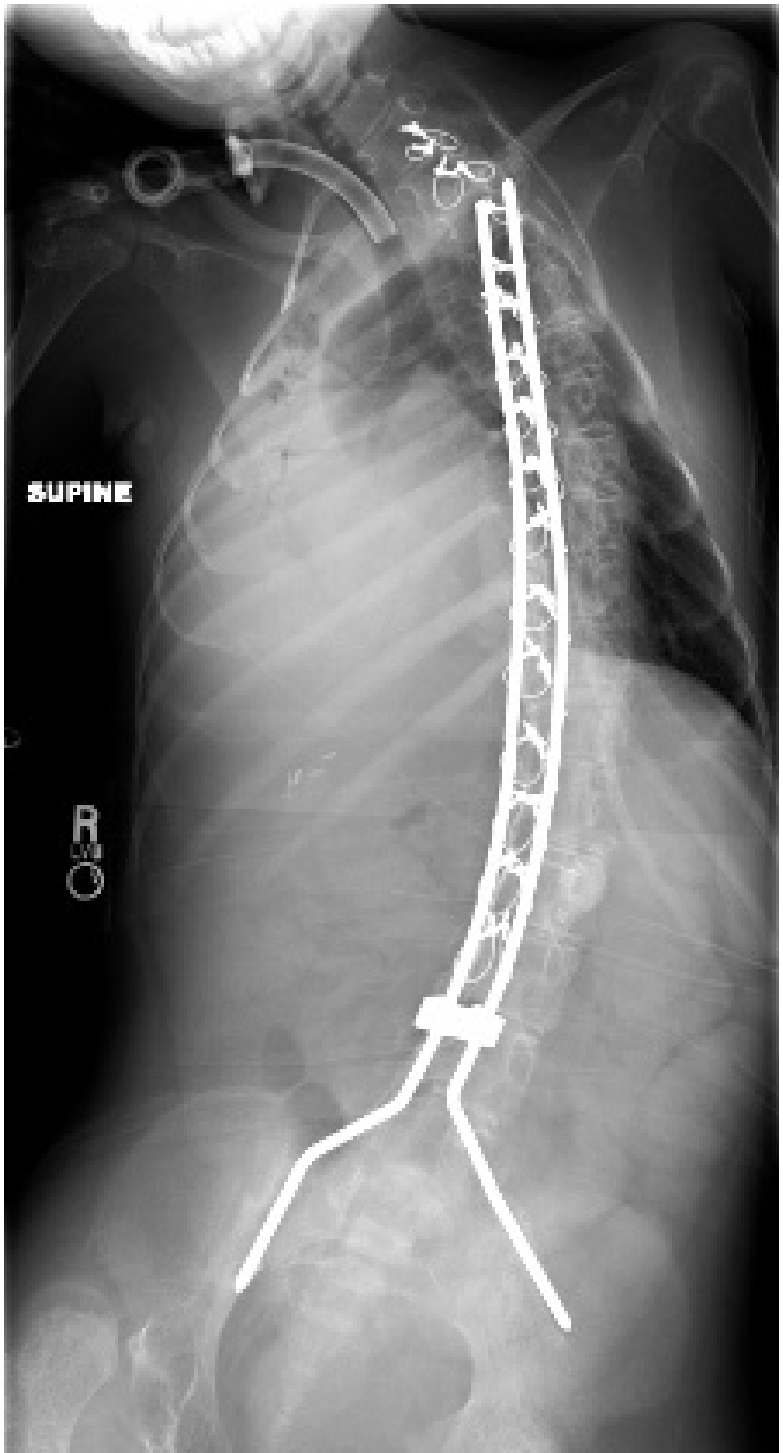


7 months post op





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.

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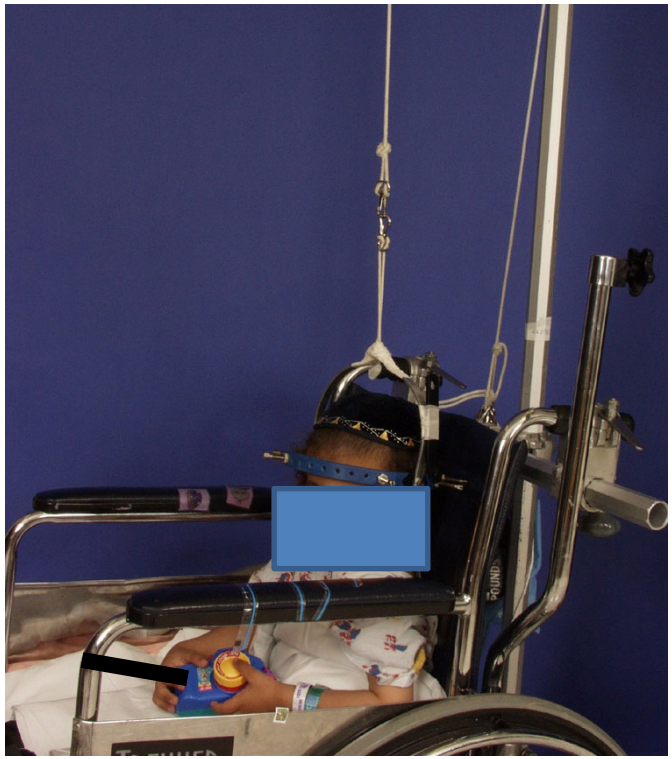
Halo Traction

- 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

Halo Traction



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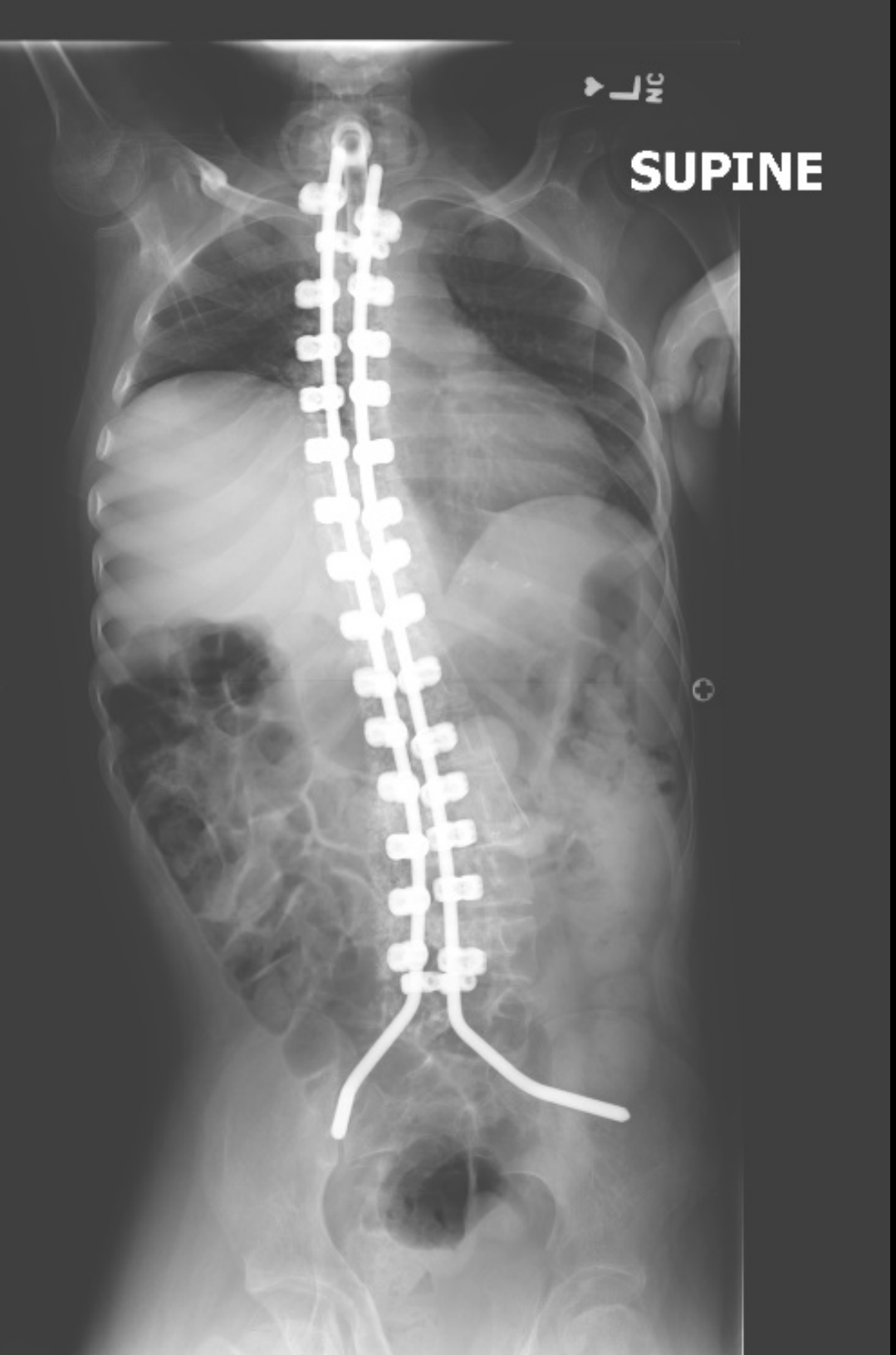
SUPINE


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NC/TW
SUPIN

Intrathecal Administration of Medication with Spinal Deformities

- Guided placement of intrathecal catheter: ultrasound, fluoroscopy, interventional radiologist
- Procedural sedation, cumulative effects of anesthesia
- Laminotomy, laminectomy to facilitate guided placement of needle, catheter
- Indwelling catheter, reservoir, infusion pump to deliver medications without repetitive dural puncture
- Planning long term intrathecal access at the time of spinal reconstructive procedures

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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

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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

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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

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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

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