



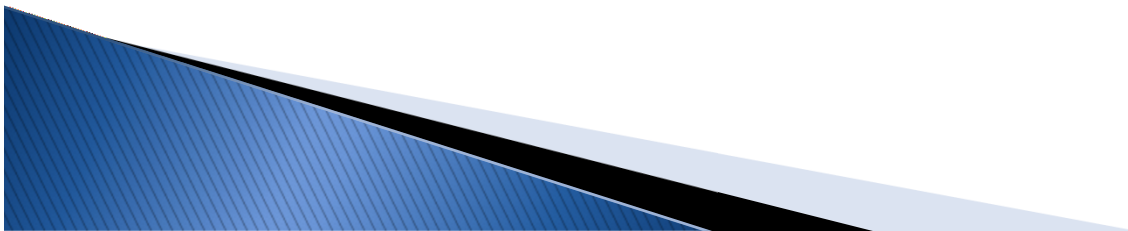
Case Presentation

International Congress of Early Onset Scoliosis
San Diego, California
November 2013

Ron El-Hawary, MD, MSc, FRCS(C)
Halifax, Nova Scotia, Canada

History

- ▶ 6 year old male with achondroplasia
- ▶ Previous C1–3 laminoplasties
- ▶ VP shunt
- ▶ Positive Babinski bilateral



Physical Examination



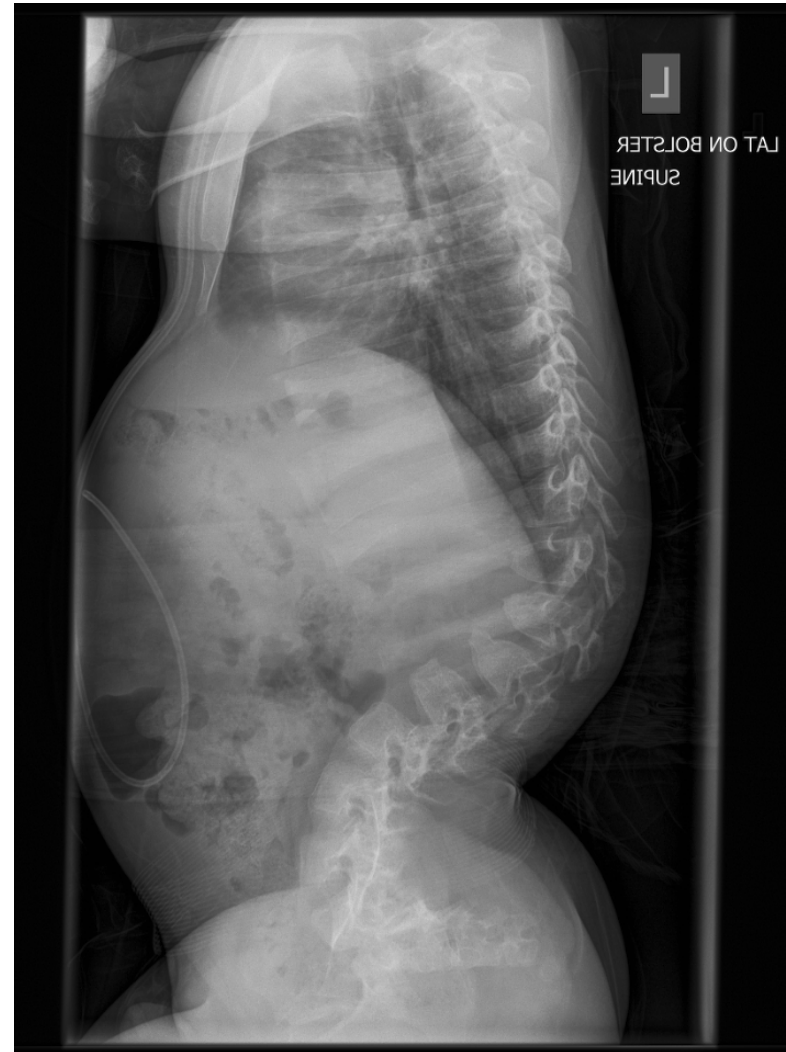
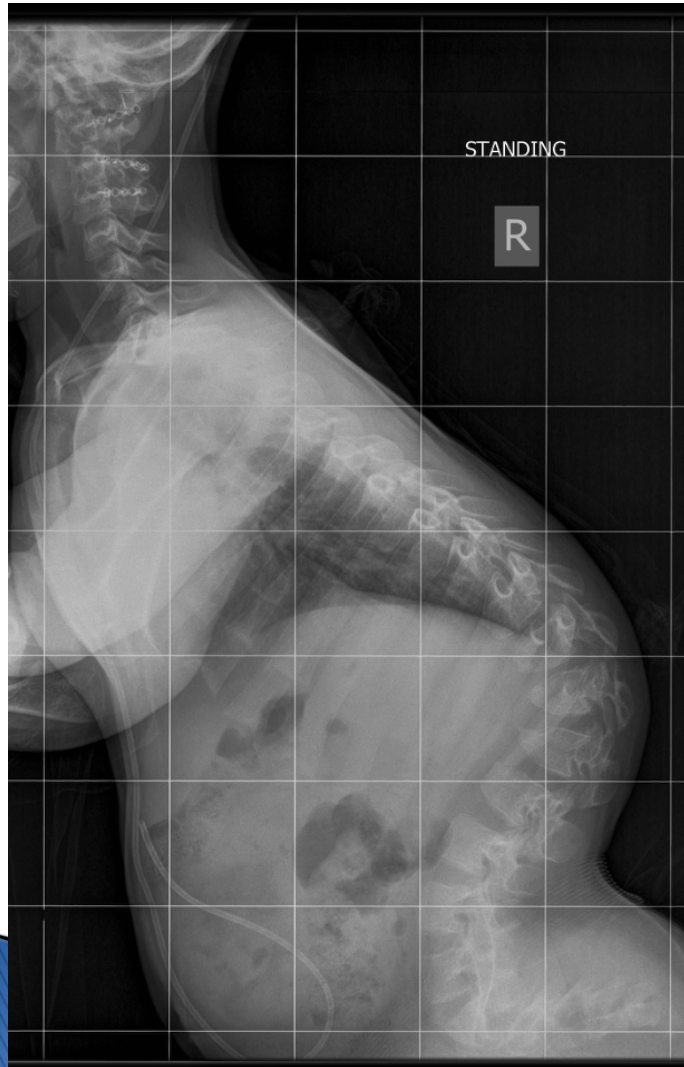
Flexion – Extension C-Spine



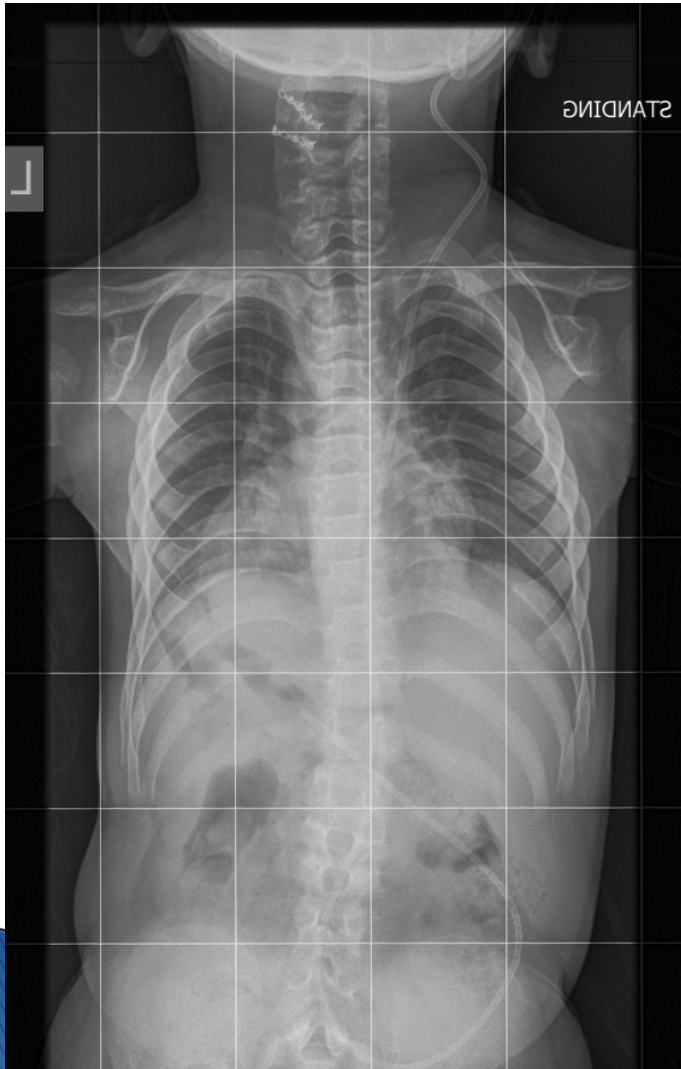
Scoliosis Radiographs



Lateral Flexibility Film



What to do?



Spinal Arthrodesis With Instrumentation for Thoracolumbar Kyphosis in Pediatric Achondroplasia

Michael C. Ain, MD, and James A. Browne, BA

- ▶ PSFI between 1998 and 2001 on 12 patients
- ▶ Mean age = 12 years

Spinal Arthrodesis With Instrumentation for Thoracolumbar Kyphosis in Pediatric Achondroplasia

Michael C. Ain, MD, and James A. Browne, BA

- ▶ ASF / PSFI (n=5 patients)
- ▶ PSFI (n=7 patients)

Indications for spinal fusion in the pediatric achondroplast with thoracolumbar kyphosis

Concomitant laminectomy, regardless of curve magnitude

Curve $> 50^\circ$ at age 4 or older

Indications for an anterior/posterior procedure

For corpectomy to relieve anterior impingement

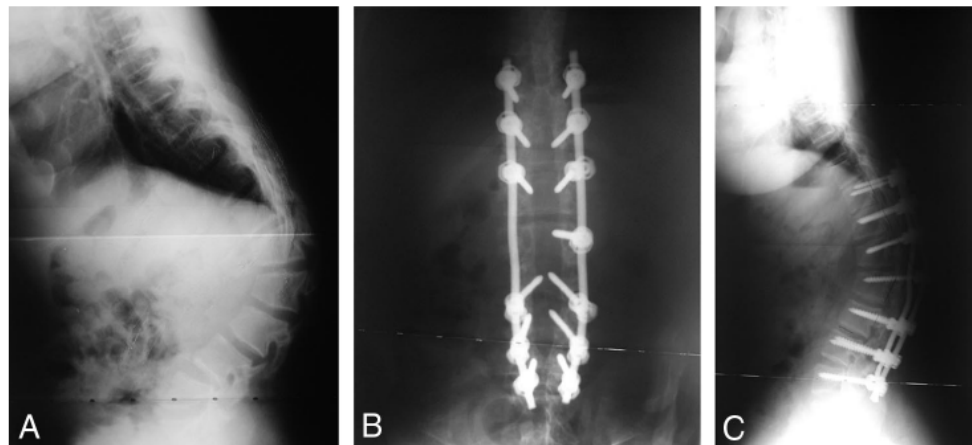
Kyphosis $> 50^\circ$ on preoperative hyperextension lateral radiographs

Small pedicle size thought to be inadequate for screw placement

Spinal Arthrodesis With Instrumentation for Thoracolumbar Kyphosis in Pediatric Achondroplasia

Michael C. Ain, MD, and James A. Browne, BA

- ▶ Average thoracolumbar kyphotic deformity of 64° (range, 43°–88°).
- ▶ Mean improvement in kyphotic deformity was 50%.



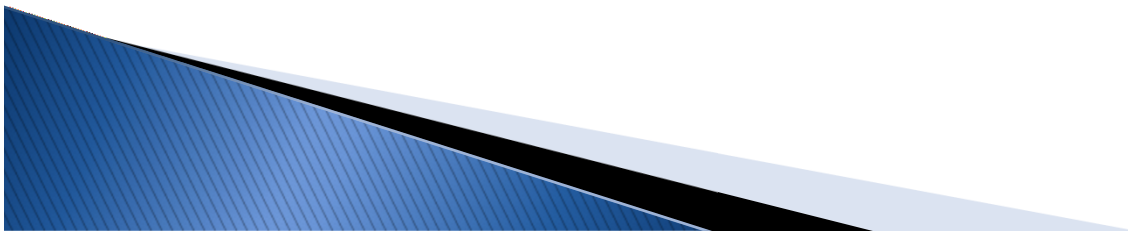
Spinal Arthrodesis With Instrumentation for Thoracolumbar Kyphosis in Pediatric Achondroplasia

Michael C. Ain, MD, and James A. Browne, BA

- ▶ Successful fusion was obtained in all patients.
- ▶ No intra or post-op neurologic deterioration
- ▶ Complications included
 - 3 instrumentation fractures (2 patients)
 - 1 dural leak.

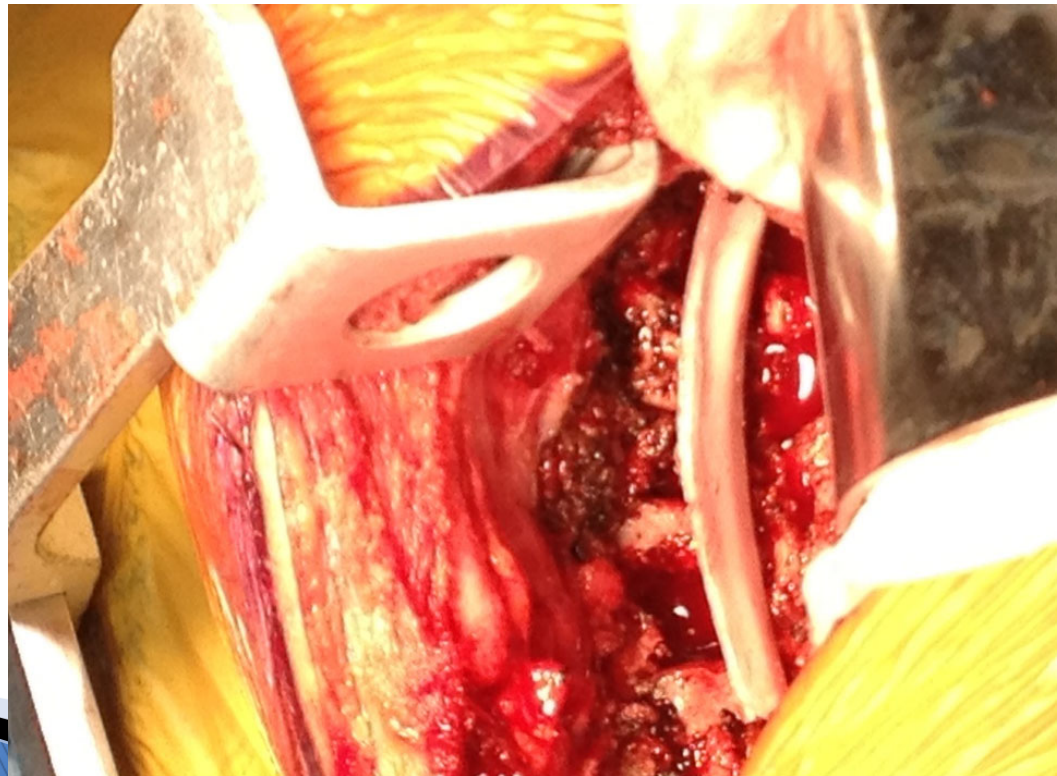
Surgery – Plan

- ▶ Single Stage – in situ
- ▶ Anterior Spinal fusion T11–L2 with Strut Graft
- ▶ L1–T12 laminectomies
- ▶ Posterior Spinal Fusion and Instrumentation T9–L3
- ▶ No attempt at reduction



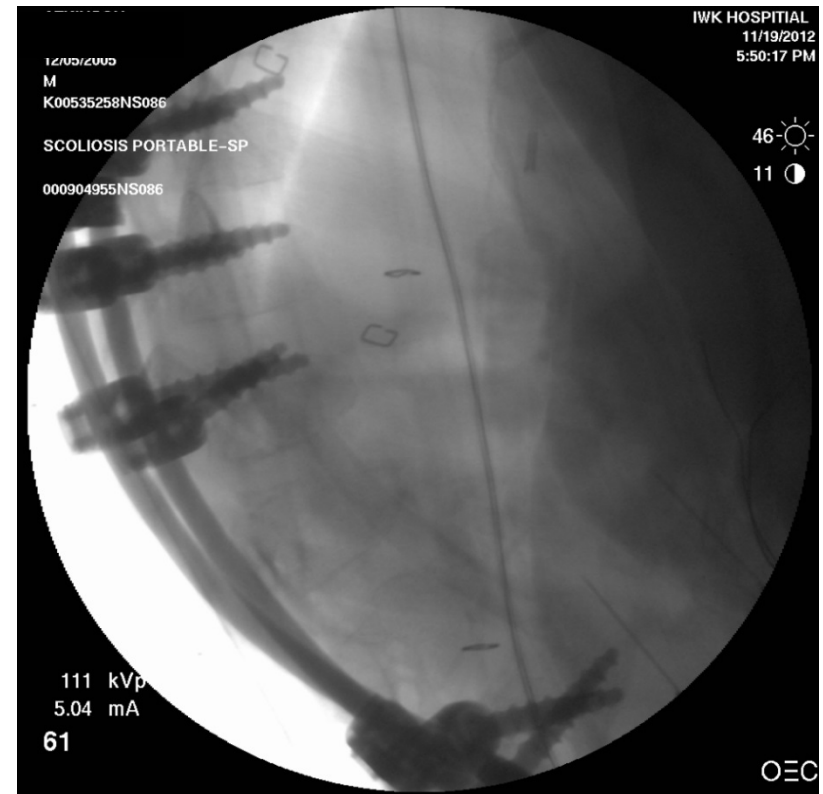
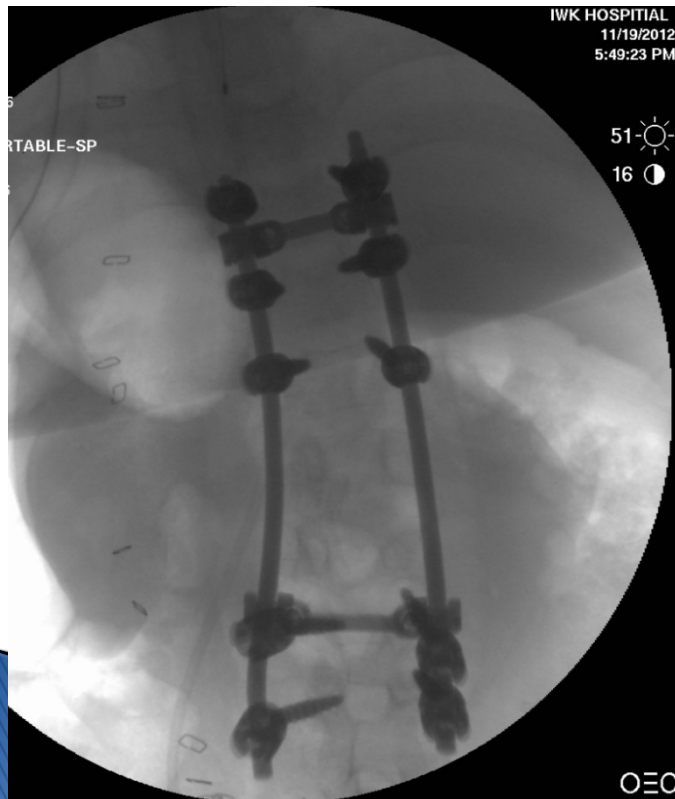
Surgery

- ▶ Anterior spinal fusion T11-L2
 - Left thoracotomy
 - 10th rib strut graft



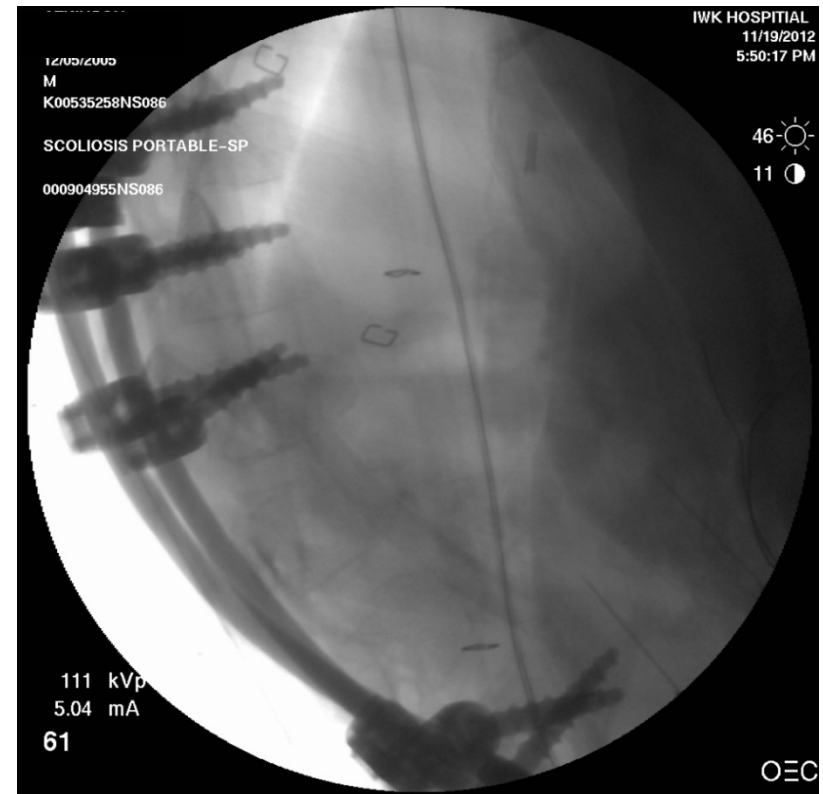
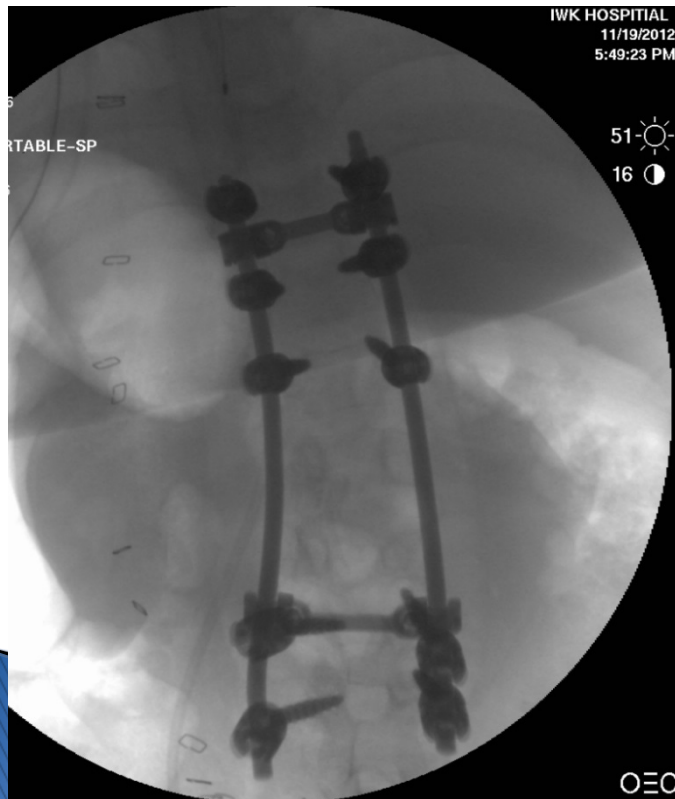
Surgery

- ▶ L1-T12 laminectomies
- ▶ PSFI T9-L3



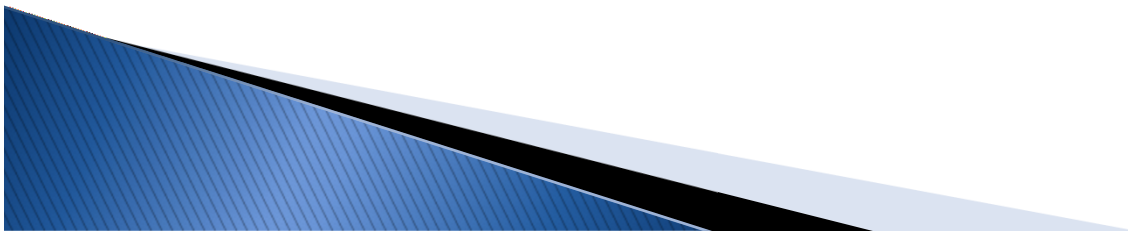
Surgery

- ▶ Lost MEPs after crosslinks tightened.
- ▶ What to do?



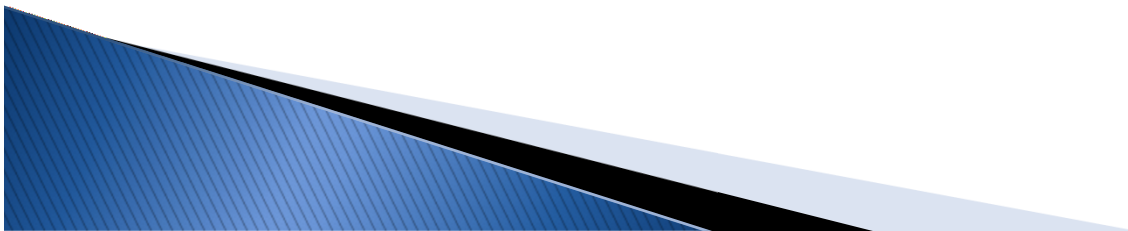
Surgery

- ▶ Removed crosslinks
- ▶ Increased BP
- ▶ Transfused
- ▶ Methylprednisolone (30 mg/kg bolus)



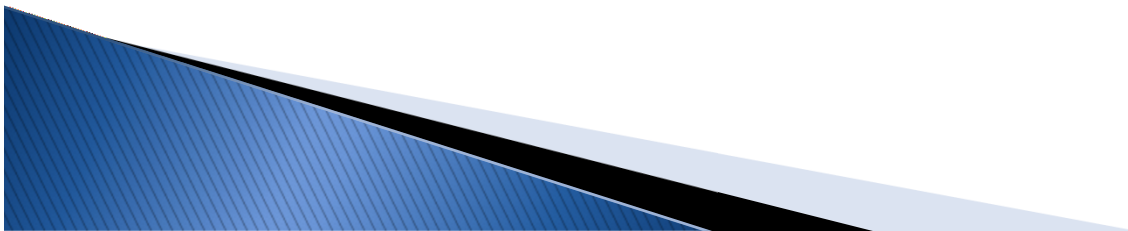
Surgery

- ▶ Wake up test – no lower extremity movement
- ▶ Removal posterior rods (screws left in place)
- ▶ Improvement in MEPs within 10 minutes



Post Op

- ▶ Neuro intact in PACU
- ▶ PICU for monitoring
- ▶ Dopamine / Norepi gtt for MAP > 80 for 48 hr
- ▶ Methylprednisolone 5.4mg/kg for 48 hr
- ▶ Hgb > 100

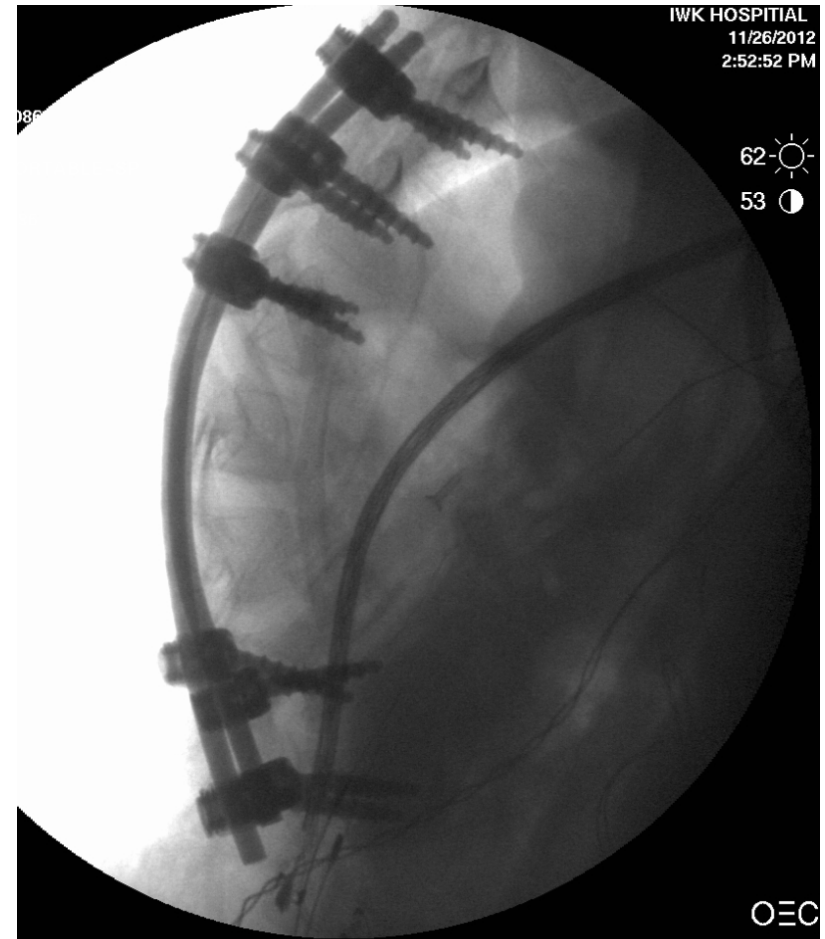
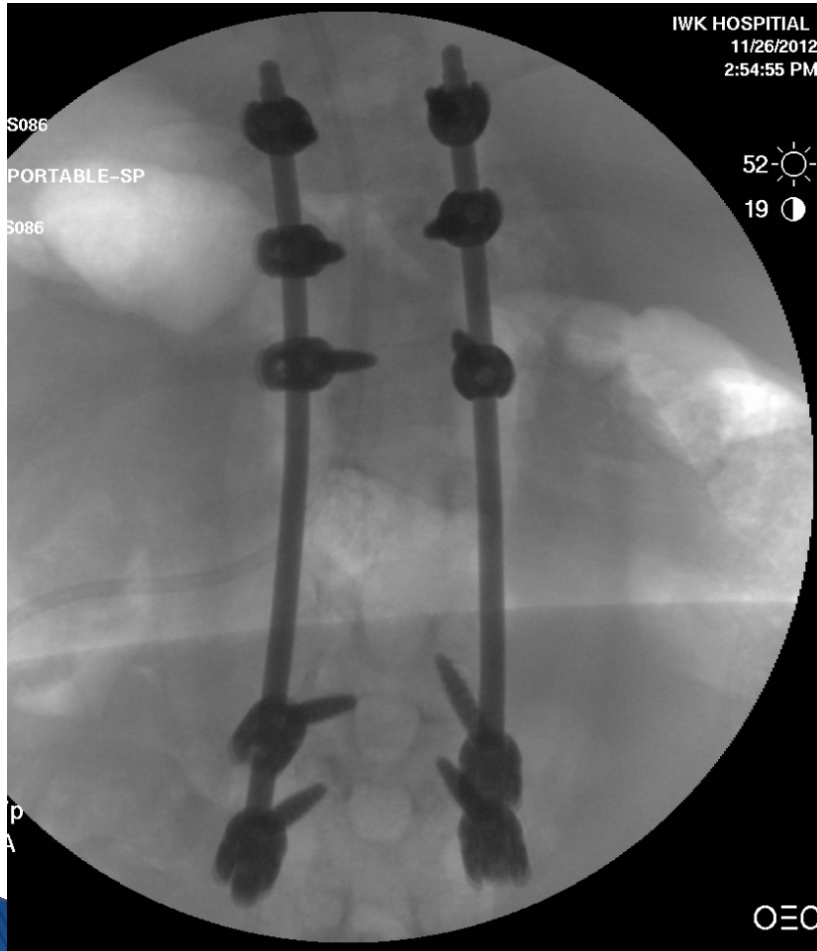


Post Op

- ▶ Potential for return to OR at later date for rod insertion?

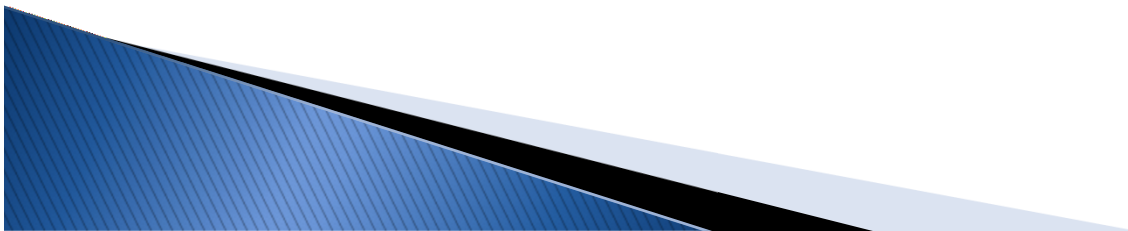


O.R. #2 – One week later



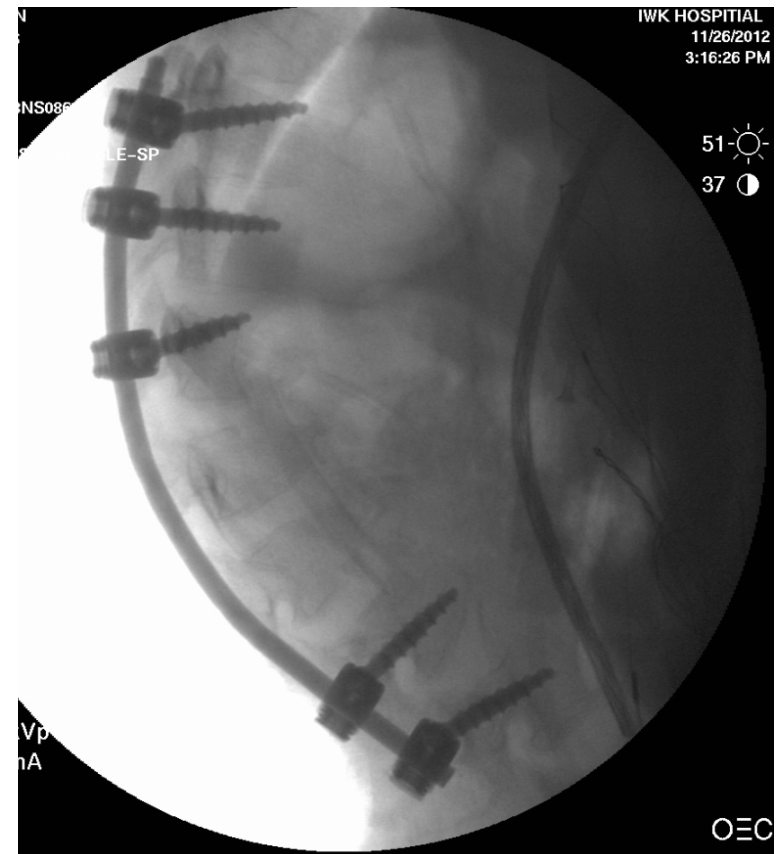
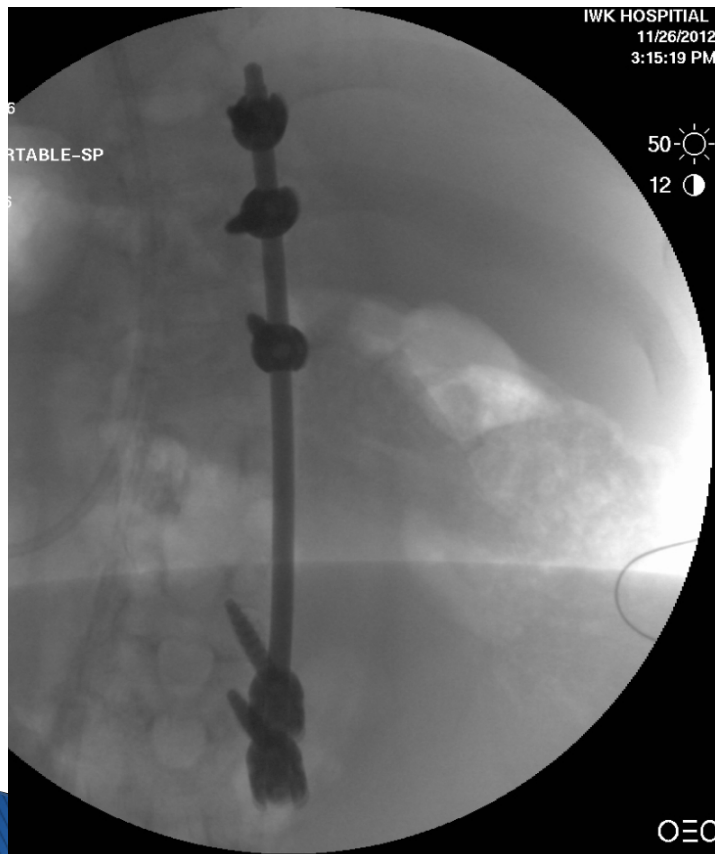
O.R. #2 – One week later

- ▶ “The left lower extremity TcMEPs were lost several minutes following placement of the second rod.”



O.R. #2 – One week later

- ▶ Removed left rod and left-sided screws

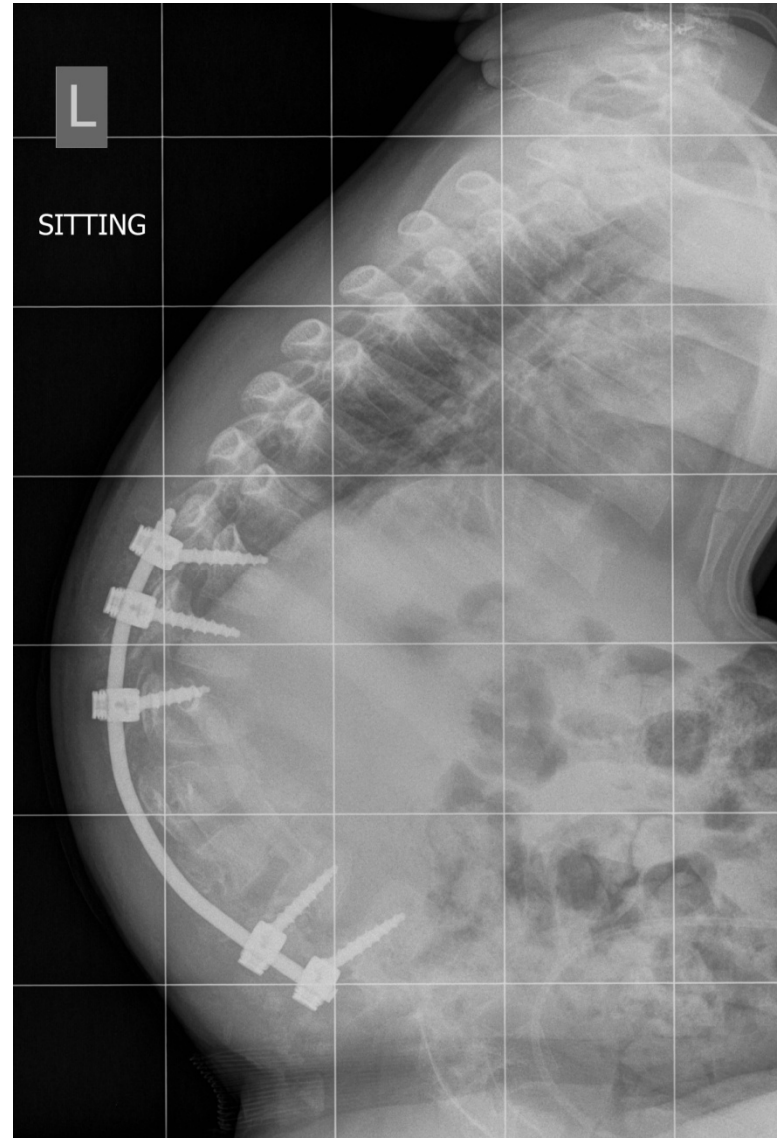


O.R. #2 – One week later

- ▶ “The TcMEPs returned to baseline values within minutes after removal of the rod on the left side. The decision was made to leave the left rod out and the left-sided pedicle screws were also removed.”
- ▶ “The lower extremity TcMEPs remained robust and at baseline values bilaterally through skin closure.”



Post-Op #2 - One Week...



Post-Op #2 - 2 Months



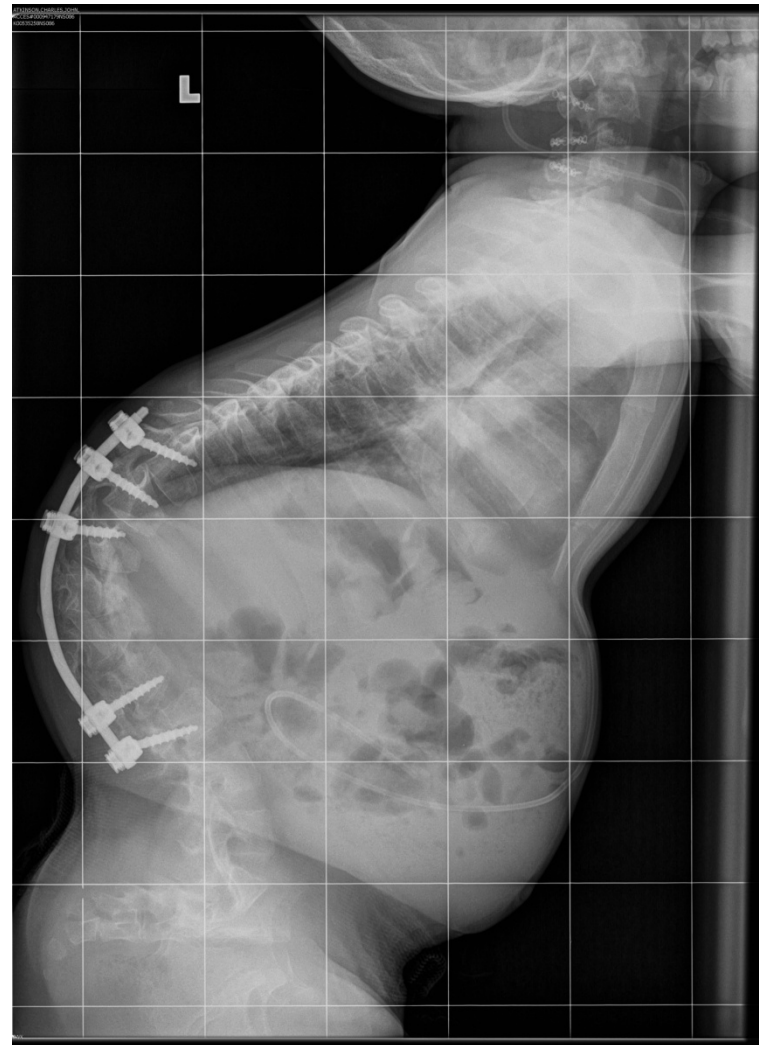
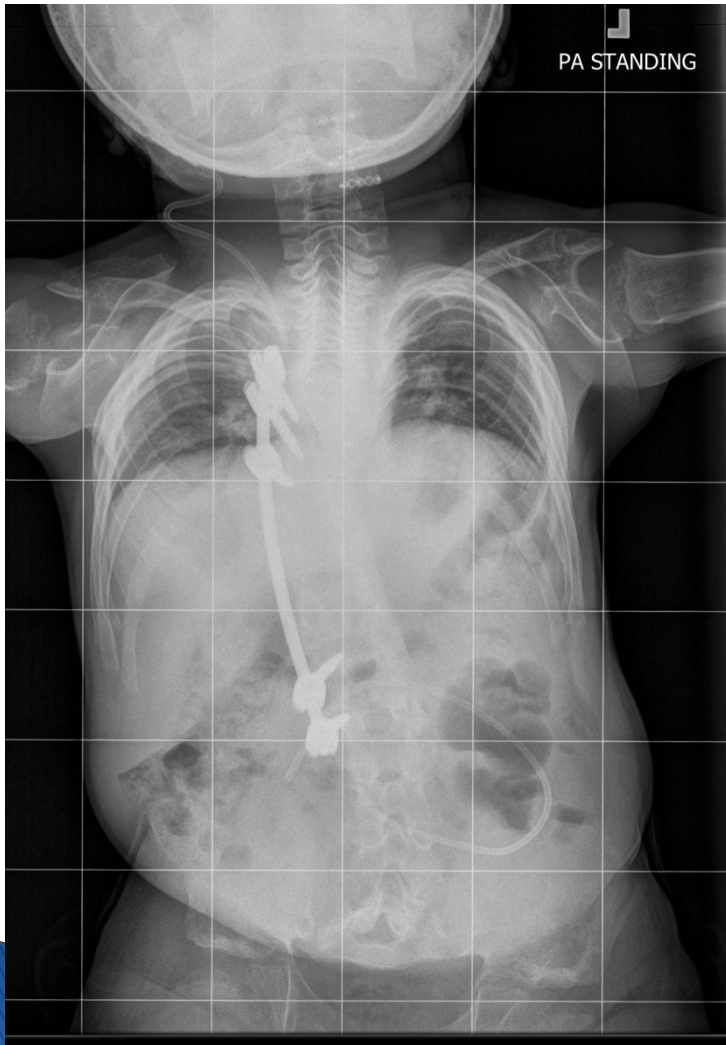
Post-Op #2 - 2 Months



Post-Op #2 - 2 Months



10 Months



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22.68

K00535258NS086

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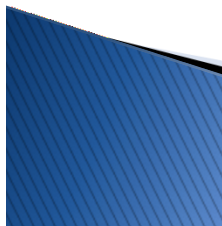
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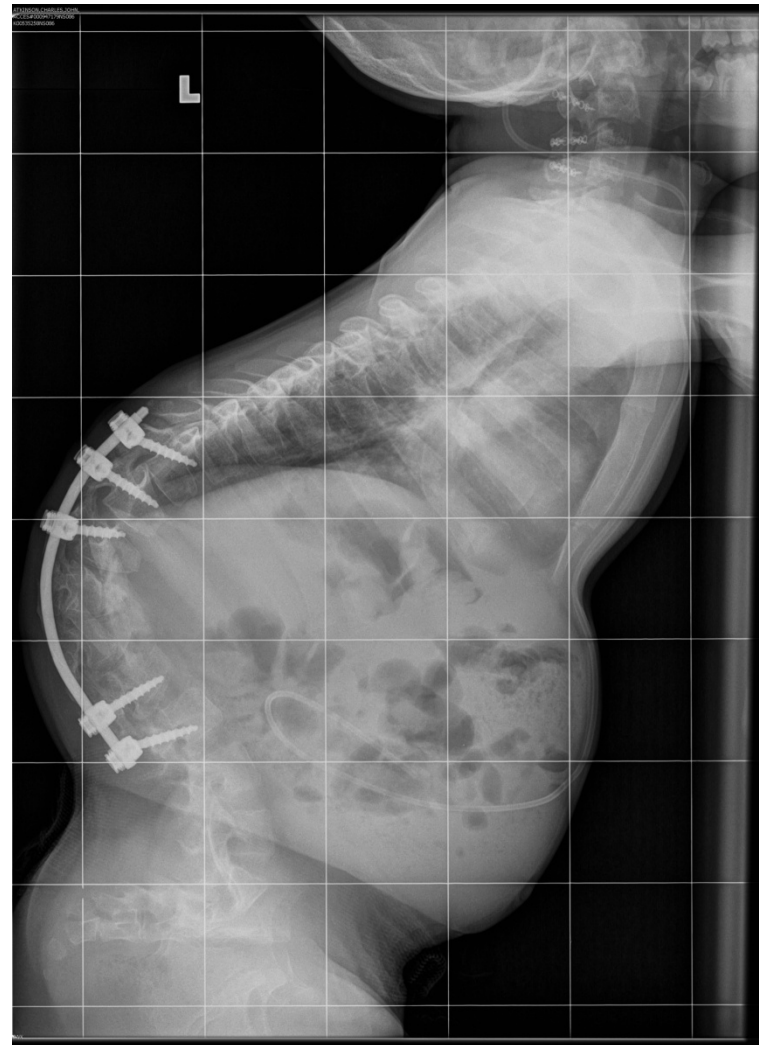
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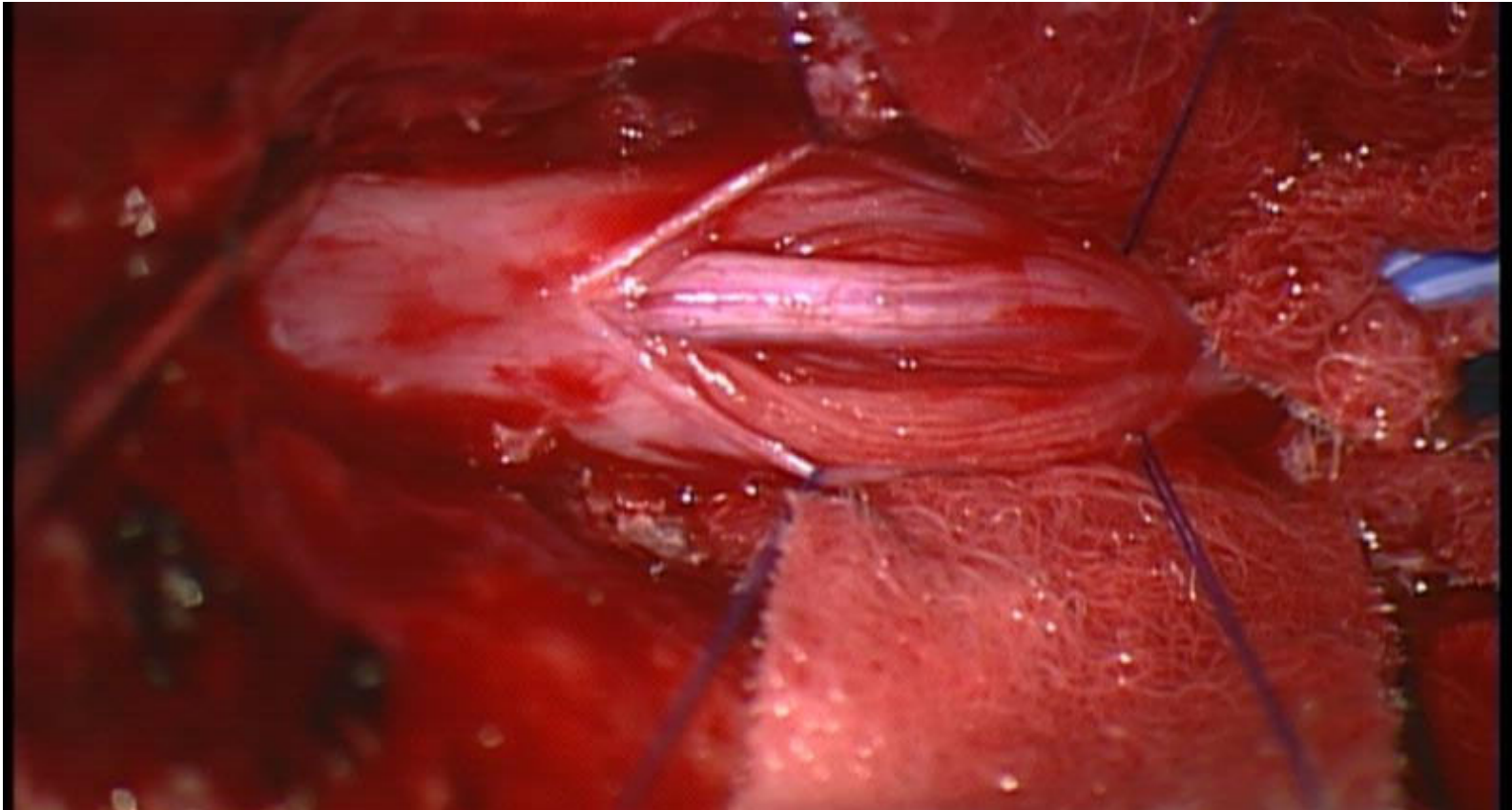
IWK HEALTH CENTRE

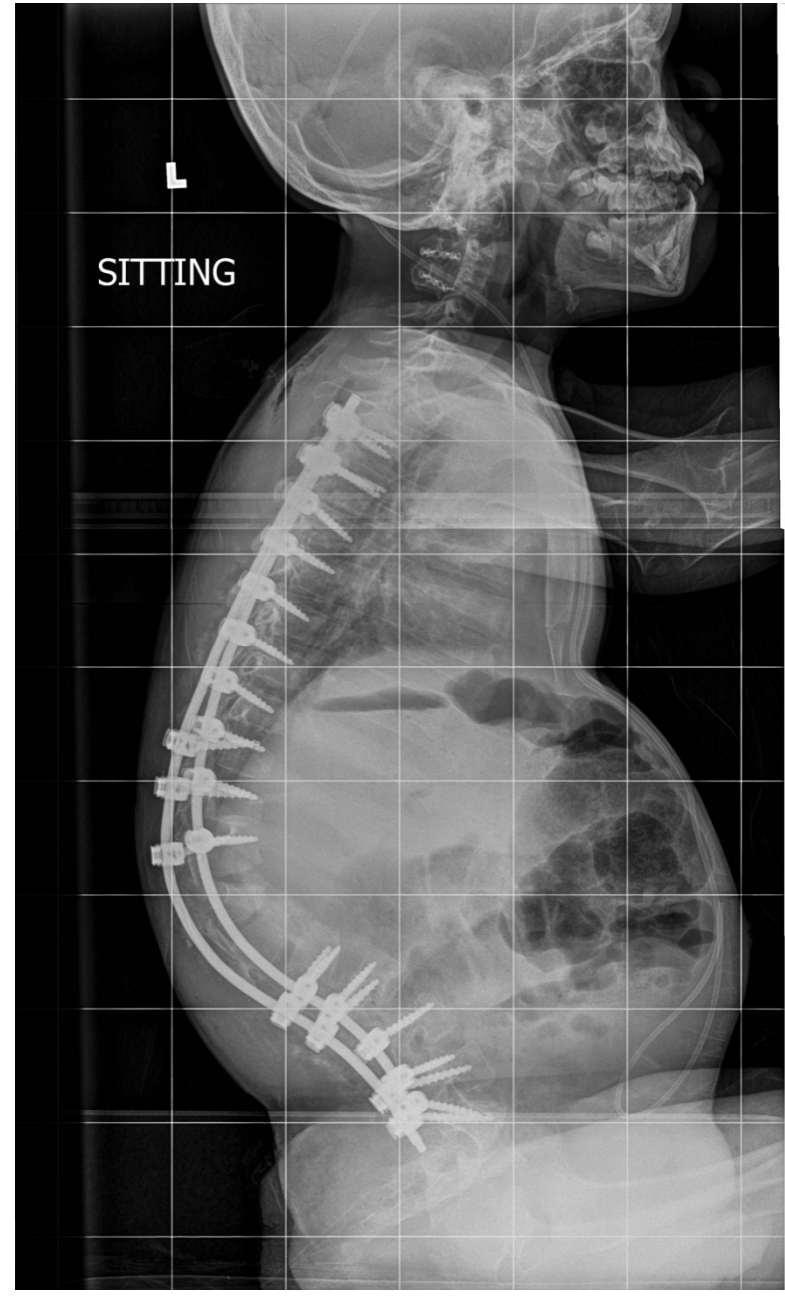
IRP

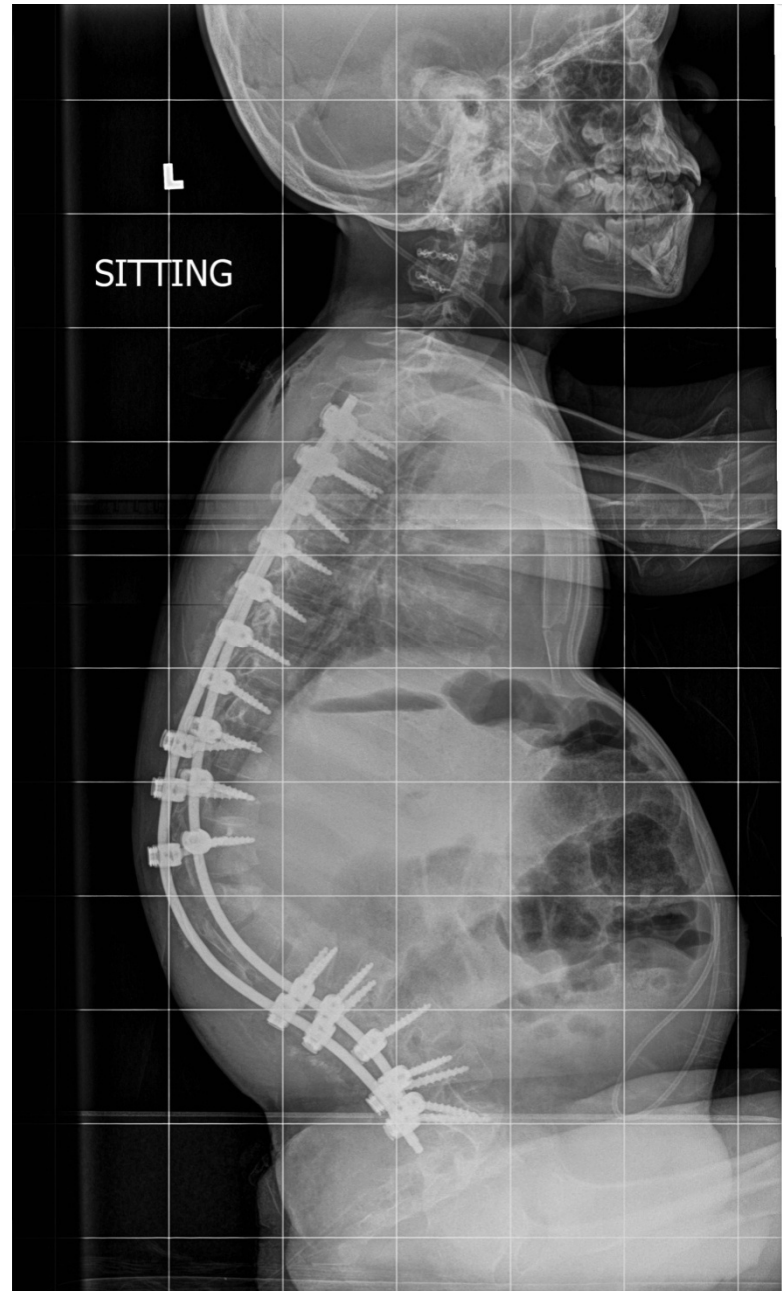
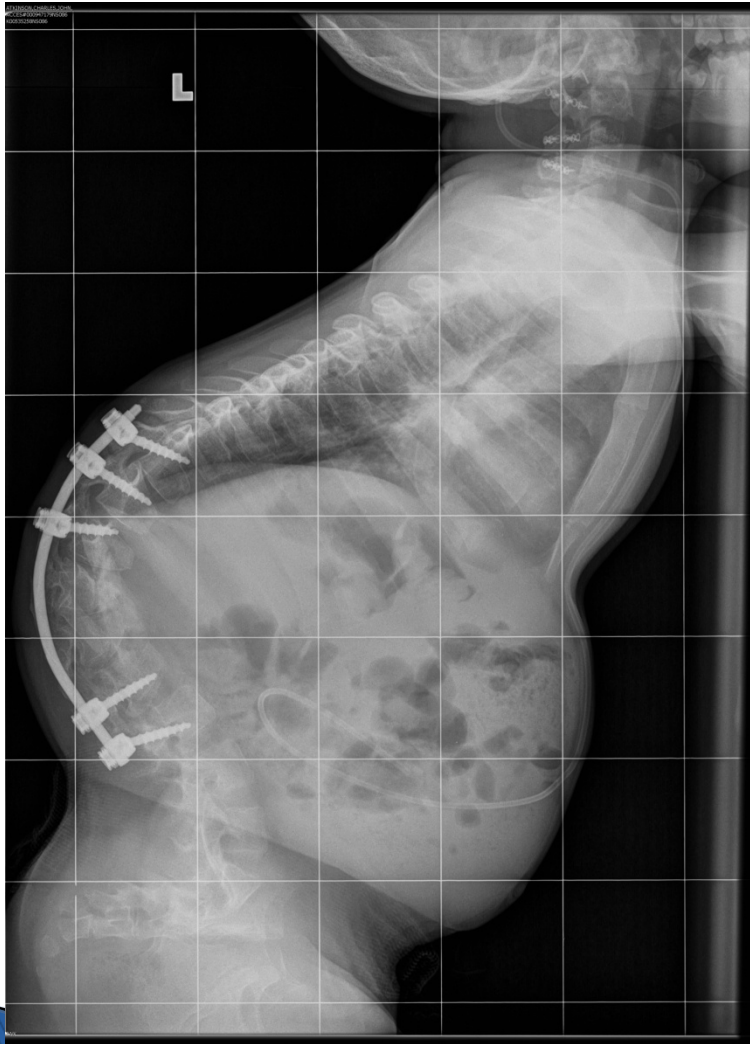


10 Months – What to do?



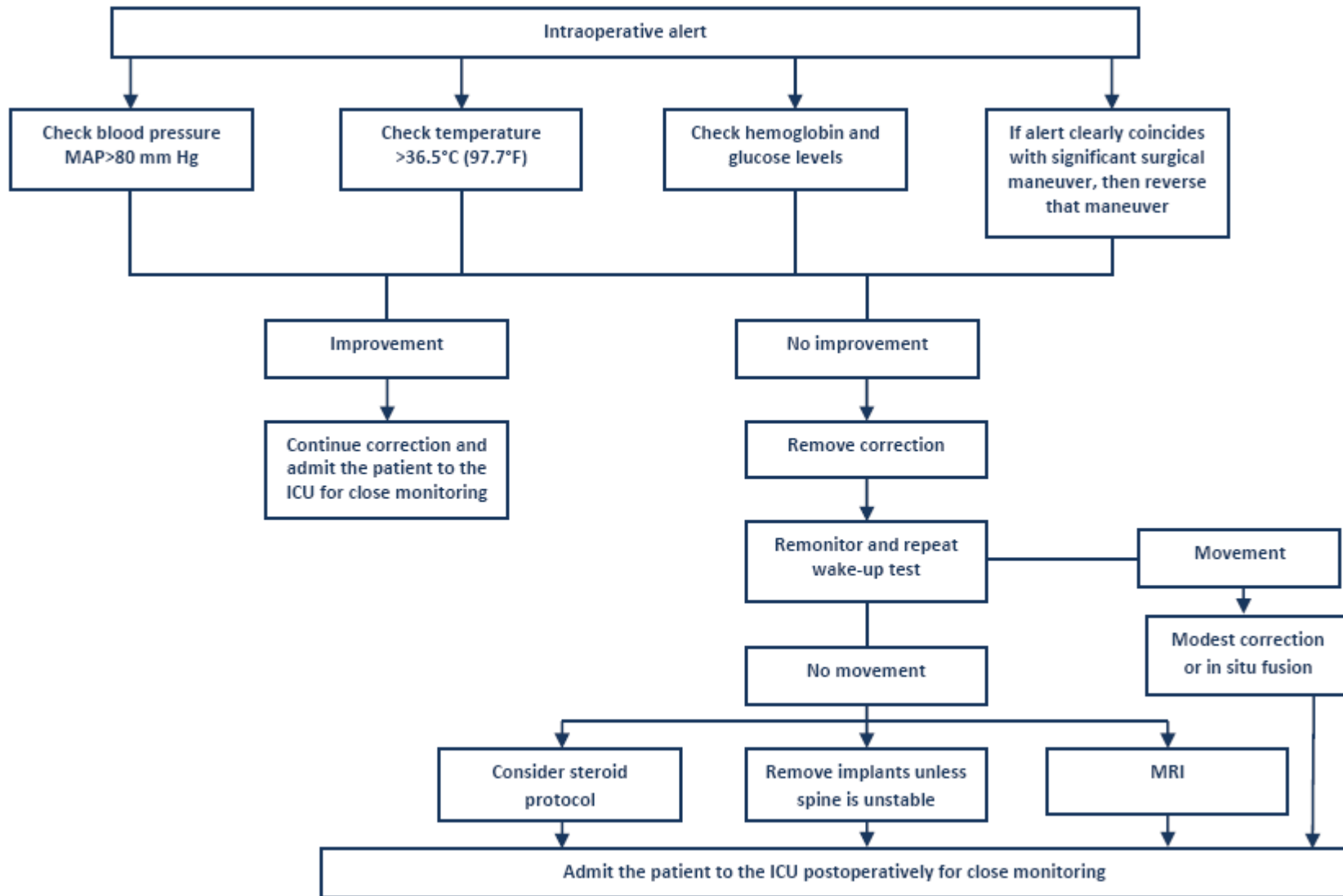






Thank You





Achondroplasia

- ▶ Most common skeletal dysplasia (1:30,000)
- ▶ FGR-3 mutation (AD, sporadic (80%), increased paternal age) – underdevelopment and shortening of long bones formed by endochondral ossification
- ▶ Rhizomelic short stature, frontal bossing, midface hypoplasia, foramen magnum stenosis (brainstem compression, apnea, sudden death)
- ▶ MSK: radial head dislocation, trident hand, genu varum, thoracolumbar kyphosis, spinal stenosis (short, thickened pedicles with narrowing of interpedicular distance)
- ▶ X-ray: squared iliac wings, rhizomelic shortening and flared metaphyses, inverted V shaped distal femoral physis, equal length metacarpals / metatarsals

