

Cervical Spine Challenges in Children with Neurofibromatosis type 1

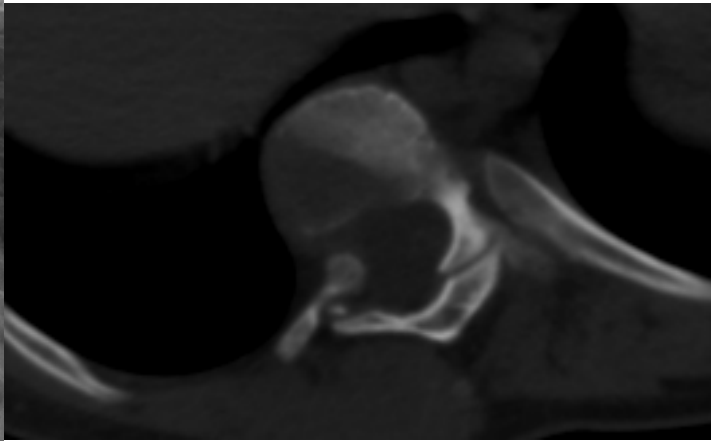
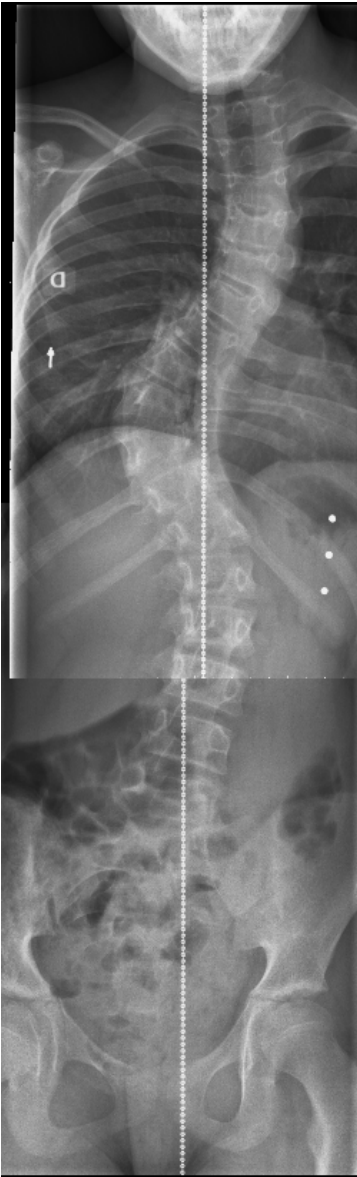
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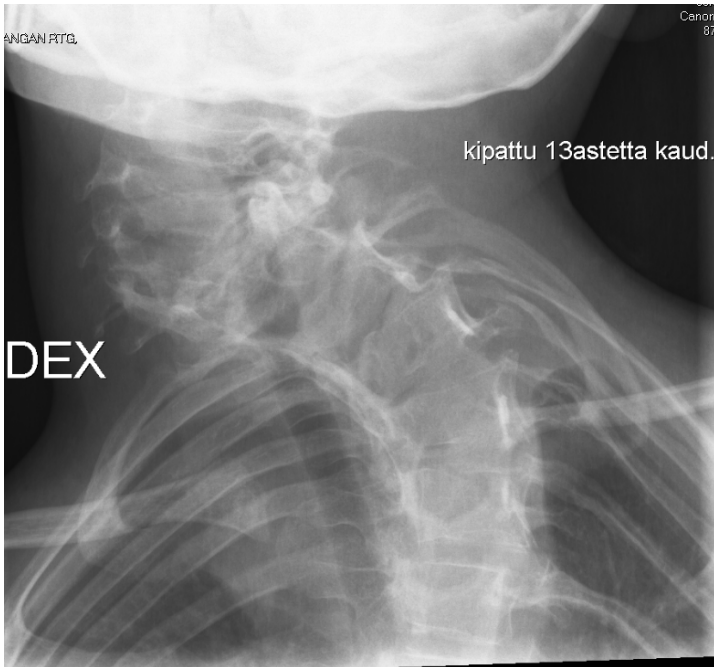


Dysplastic scoliosis



- Short angular deformities with 4-6 vertebrae
- Rapid progression of the deformities
- Vertebral dysplasia, scalloping
- Neurofibroma mass inside the canal
- Rib lesions, rib intrusion into spinal canal
- Early Idiopathic-like scoliosis may progress into dysplastic (70%)

Cervicothoracic scoliosis



- Dysplastic form
- Short angular deformities
- Rib lesions, rib intrusion into spinal canal
- Intraspinal neurofibroma mass
- Enlarged neural foramina.
- Dural ectasia and elongation of pedicles make fixation points difficult

Cervical kyphosis

- Cervical kyphosis is sometimes seen in NF 1 (Yong-Hing et al. JBJS 1979)
- Etiology:
 - Dysplastic bone, postlaminectomy
- High risk of non-union (Graig & Govender JBJS Br 1992)
- Vertebral dysplasia, dural ectasia makes fixation points unreliable
- Untreated cases poor natural history with high risk of neurologic deficits
- Existing literature limited



12-yr-old boy with postlaminectomy kyphosis

Spontaneous Cervical Spine Dislocation

- One previous case report exists (Goffin and Grob Spine 1999)
- Non-traumatic
- Dural ectasia may provide protection for spinal cord
- This 9-yr-old boy presented with cervicothoracic deformity and neck pain
- Neurologically intact



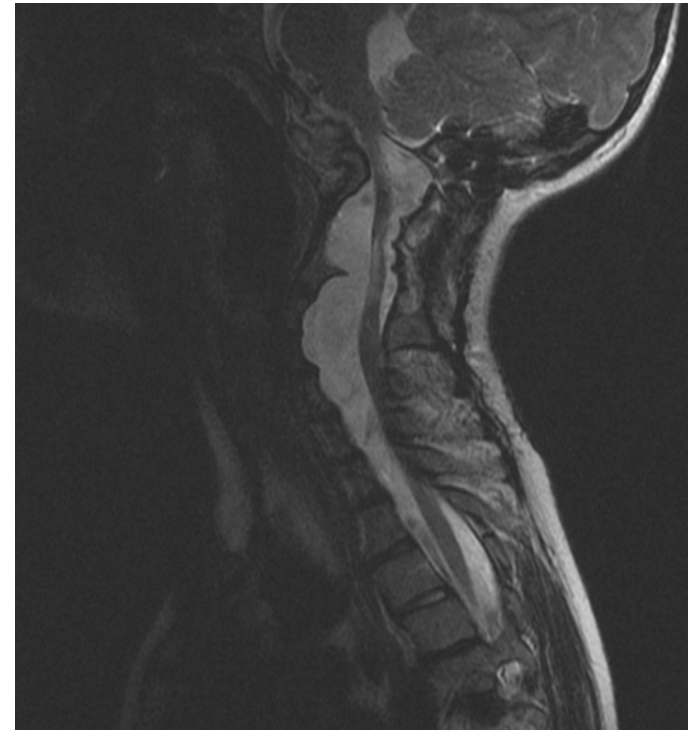
Atlantoaxial Instability

- Few case reports available in children with NF-1.
- Neurofibroma between C1 arch and dens (Toyohiko et al. J Neurosurg 1983)
- Neurologic compromise possible
- Coexistence with basilar impressions and bilateral vertebral arteriovenous fistulae possible (Maheshwari et al. Austr Radiol 2002).
- In 2 children all posterior surgery with C1 laminectomy and occipitocervical fusion provided good neurologic recovery (Toyohiko et al. J Neurosurg 1983).



Vertebral dysplasia and Dural ectasia

- Vertebral dysplasia
 - Thinning of AP diameter of vertebral bodies
 - Elongation of vertebral pedicles
 - Standard anterior cervical plate screws too long
 - Use of pedicle screws (C2, C7) difficult
 - Careful posterior approach
- Widened central spinal canal, enlarged neural foramina
 - Dural lesions during surgery
 - Posterior vertebral column resection becomes difficult



Retrospective multicenter study

N=22

Age at surgery

11.0 yrs

Range

2-19.9

Follow-up, yrs

4.1

Range

2.0-10.1

Preop Neurologic deficit

5 (23%)

Neck pain

13 (59%)

Head tilt/torticollis

10 (45%)

Scoliosis

11 (50%)



Effect of preoperative halo traction



34% correction of kyphosis with traction (N=9)
No neural deficits due to traction

Anteroposterior spinal fusion with C6 corpectomy



Note continued dysplasia and erosion

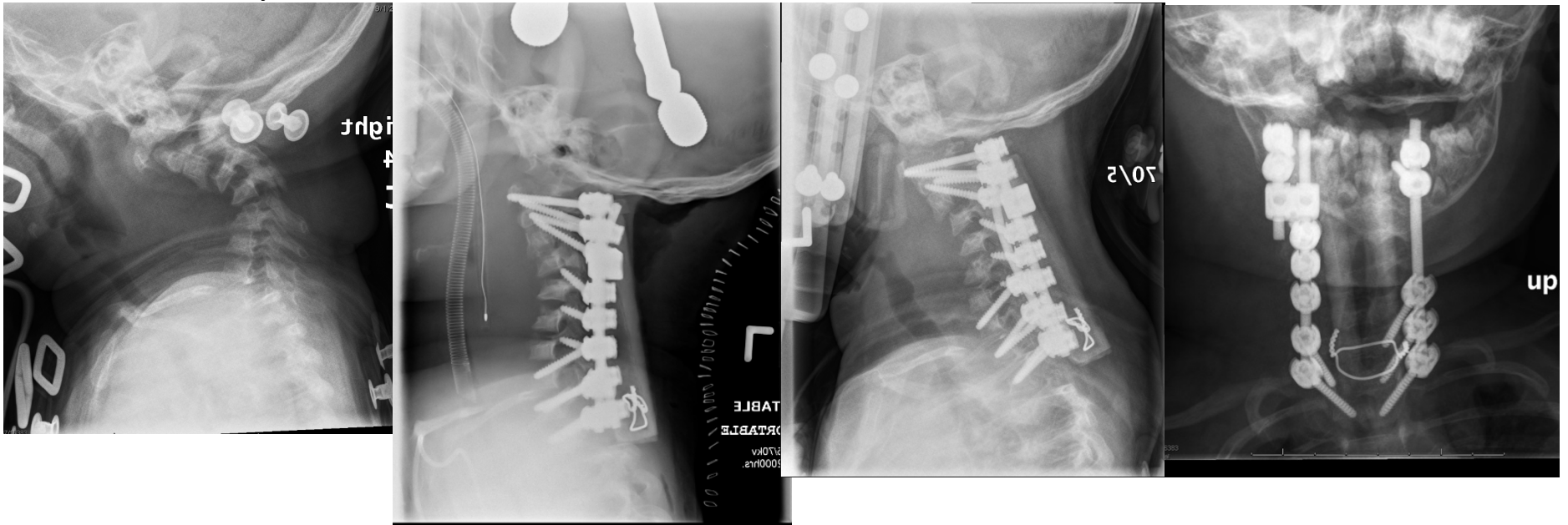
Average correction of cervical kyphosis from 66 deg to 19 deg (68%),
N=22

Postlaminectomy kyphosis at early age

Pre-op in brace
1.7 yrs, C1-C4
laminectomy

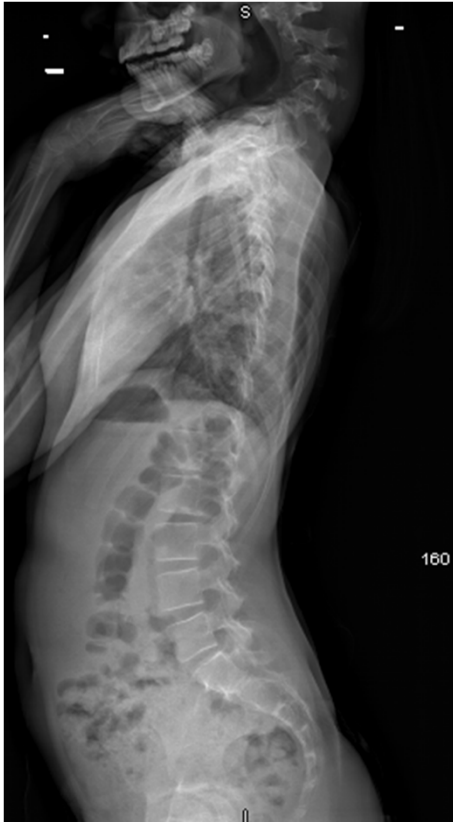
Immediate post-op

3,5 year follow-up



Courtesy of Dr. Firoz Miyanji, BC Children's, Seattle

Cervicothoracic scoliosis and cervical kyphosis



Courtesy of Dr. Joshua Pahys, Shiner's Hospital of Philadelphia, PA

10 days of halo traction with 25 pounds



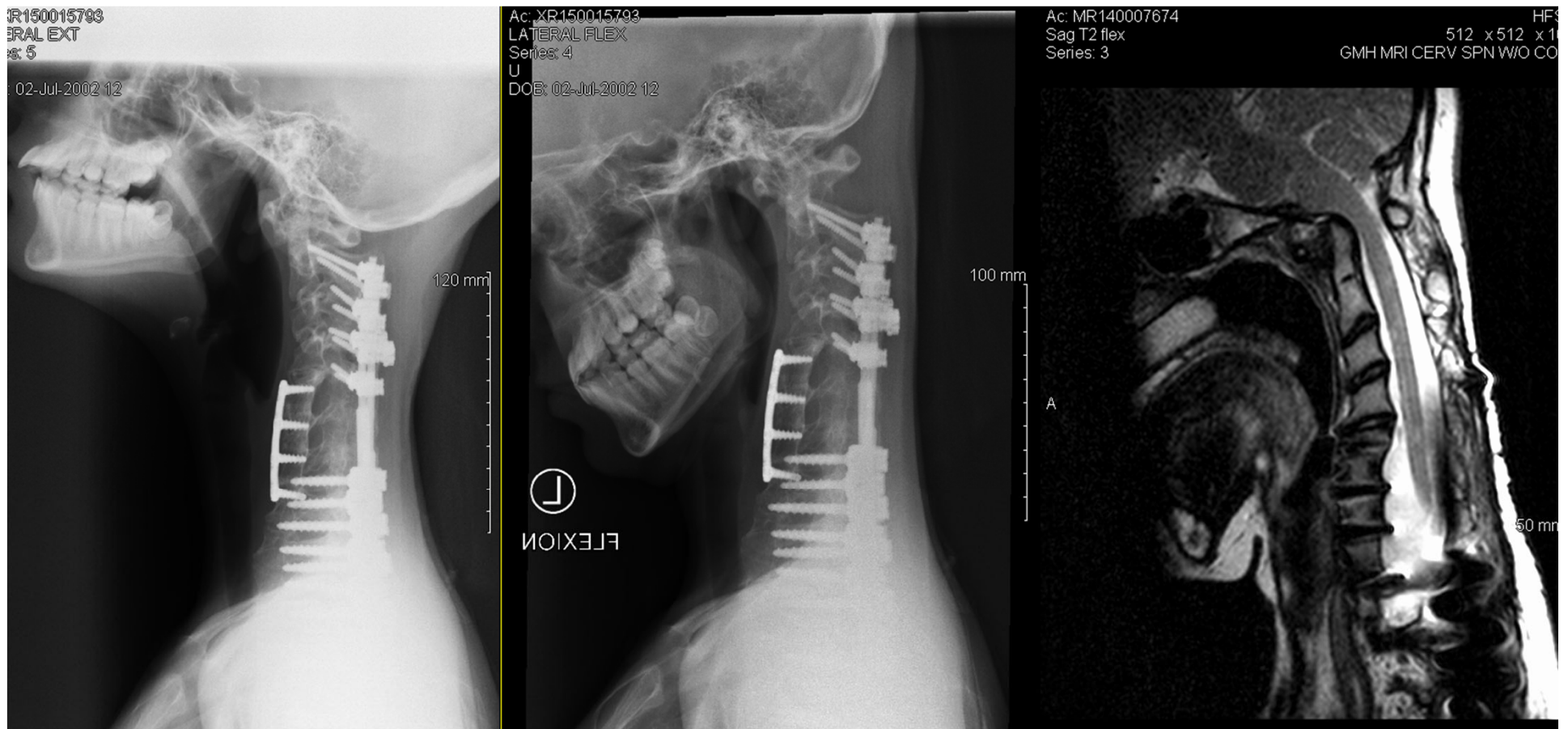
Courtesy of Dr. Joshua Pahys, Shiner's Hospital of Philadelphia, PA

6 mos postop



Courtesy of Dr. Joshua Pahys, Shiner's Hospital of Philadelphia, PA

18mos postop; progressive C1/2 instability



Courtesy of Dr. Joshua Pahys, Shiner's Hospital of Philadelphia, PA

2 yrs postop from original Ant/Post PSF
8 months s/p revision fusion to Occiput

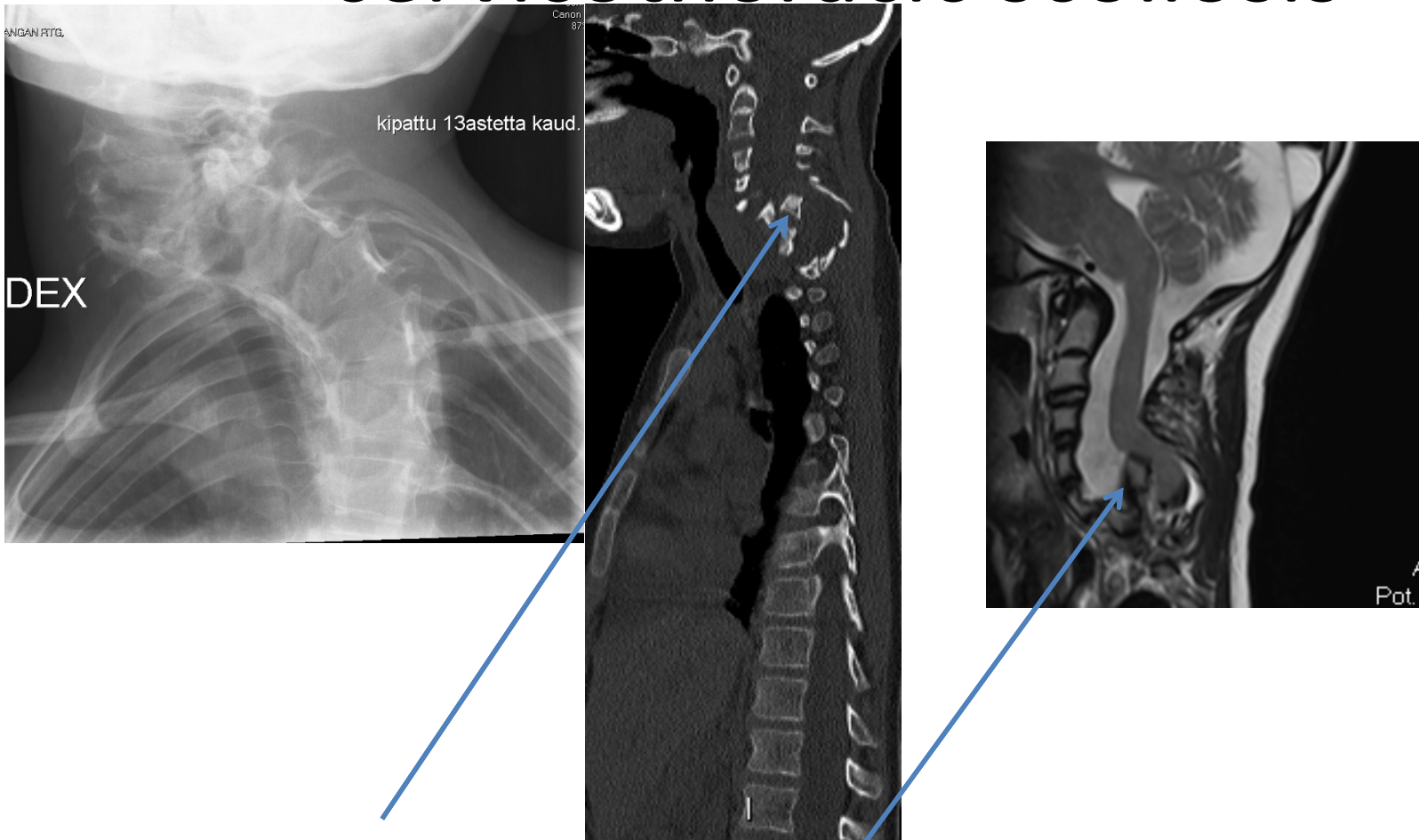


2yrs Postop after revision



Courtesy of Dr. Joshua Pahys, Shiner's Hospital of Philadelphia, PA

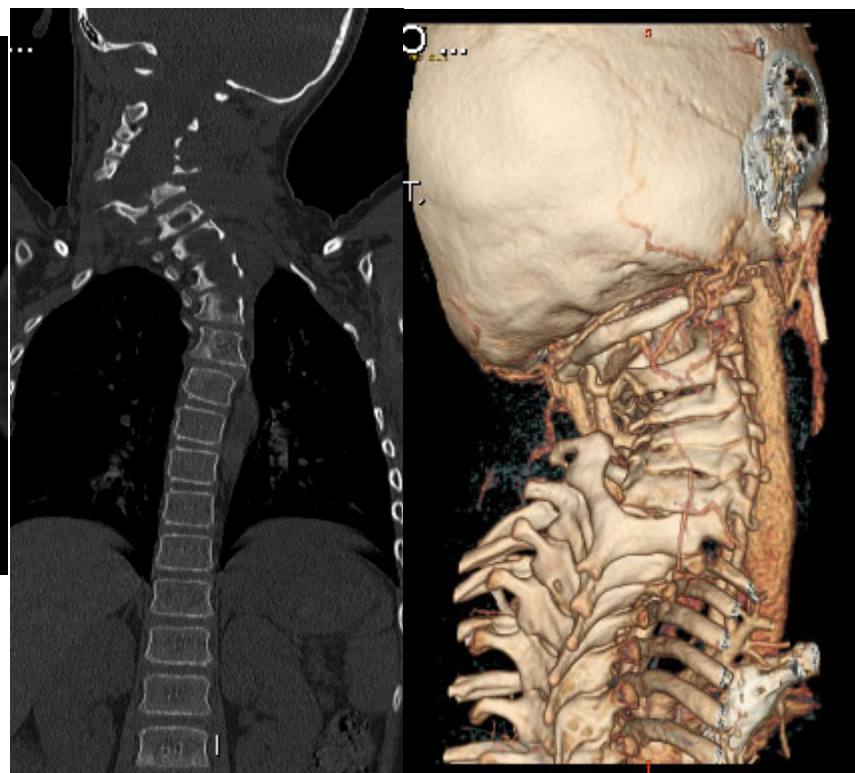
Spontaneous dislocation with cervicothoracic scoliosis



CT

C6

C7

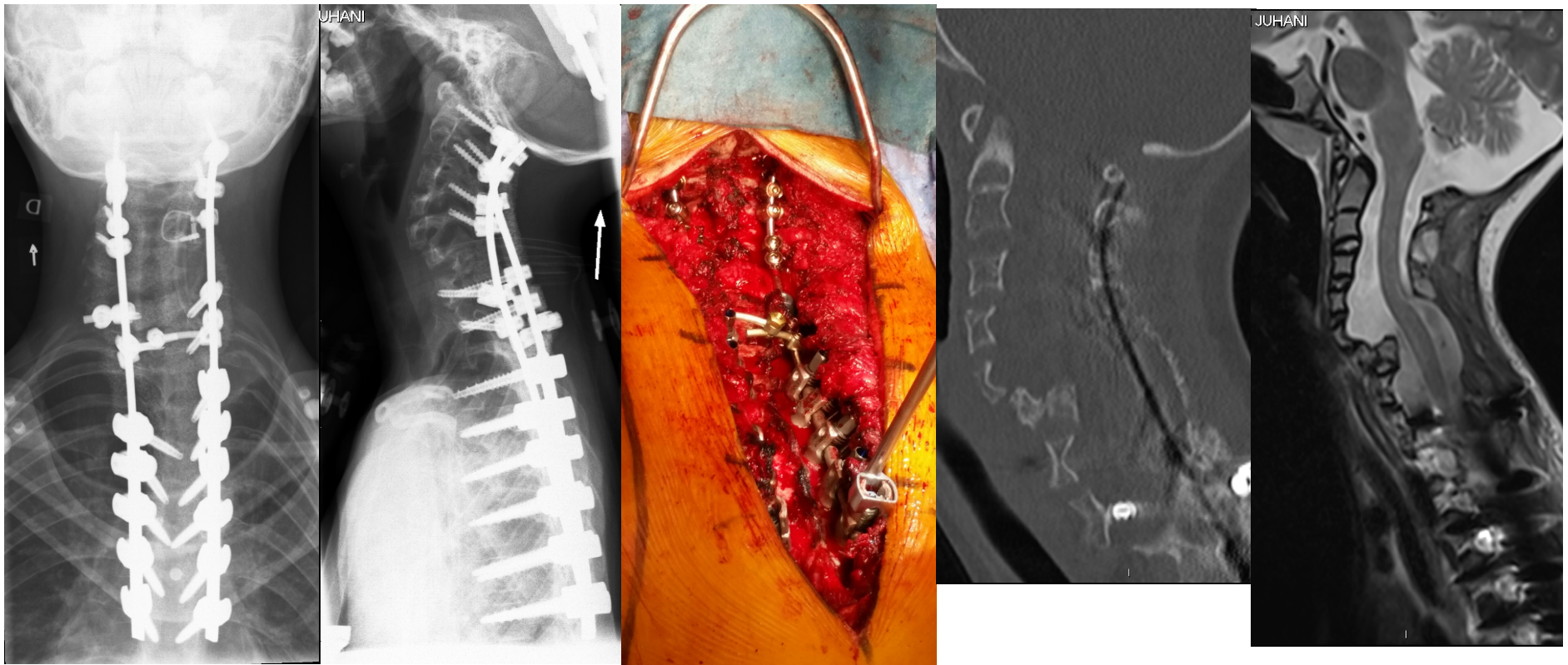


Traction 10 weeks with 8,5 kg



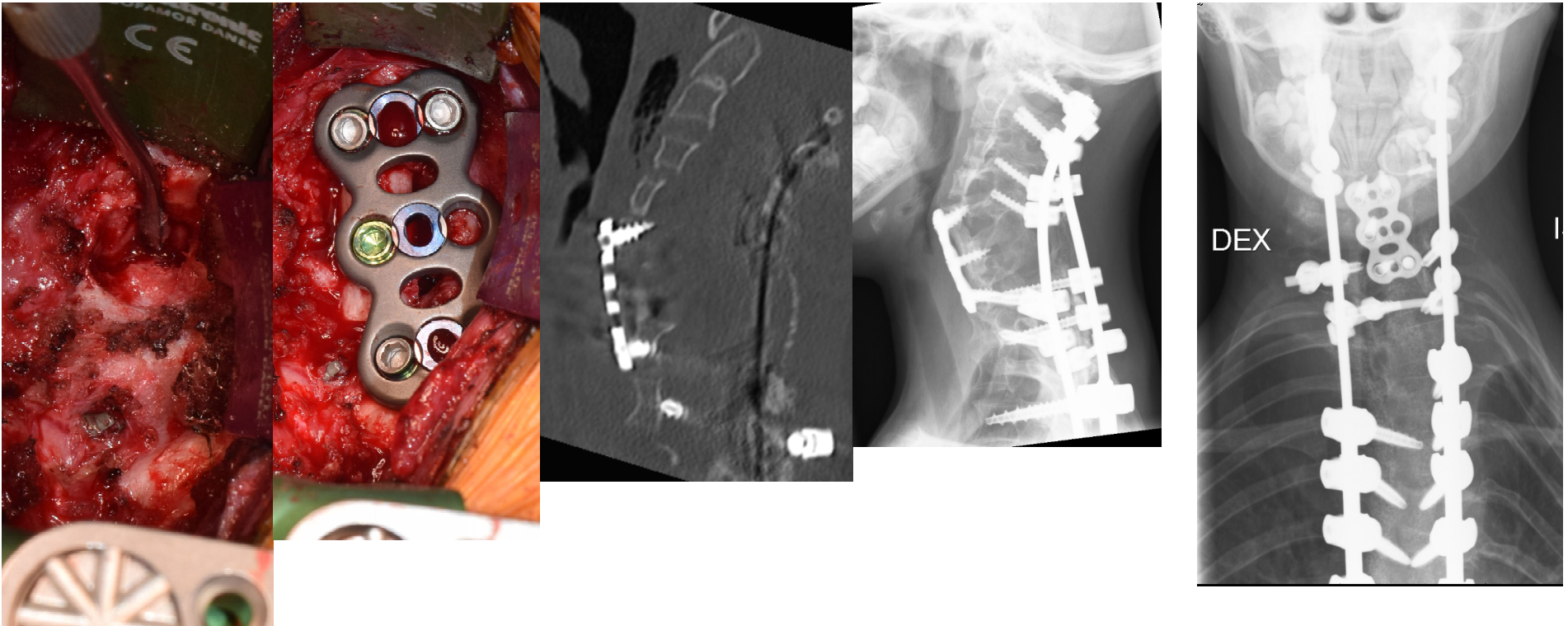
Dislocation did not reduce

Posterior C1-T9 with decompression

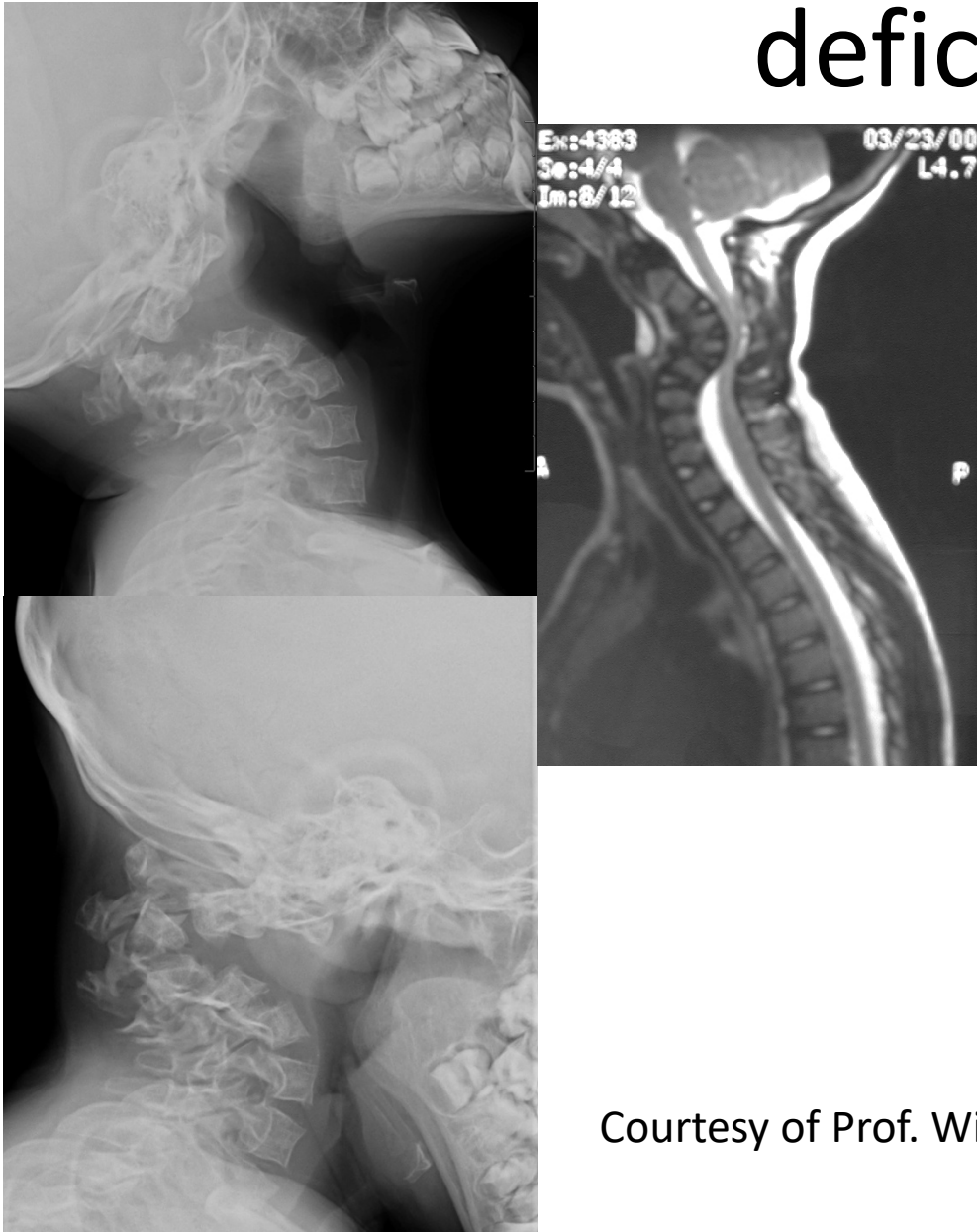


Cervical spine aligned, but spine remained dislocated. Risk of non-union?

Staged anterior corpectomy C6&7



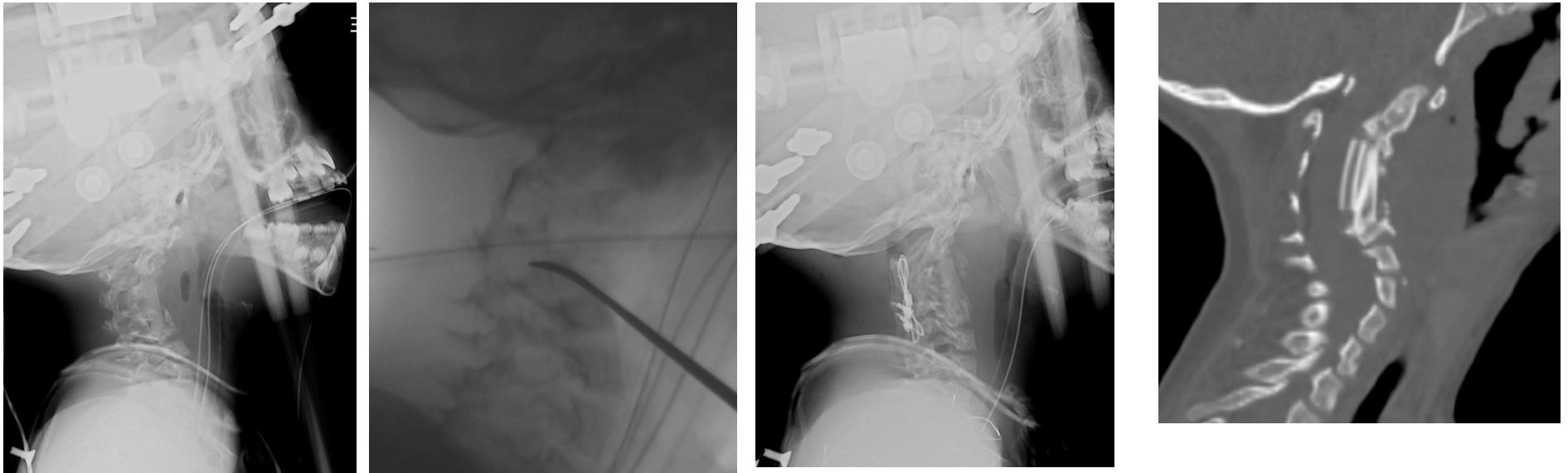
Dysplastic kyphosis with neurologic deficit



- NF-1
- 8 yo boy
- Fell on playground , c/o tingling and x-rays done

Courtesy of Prof. William Mackenzie, Wilmington

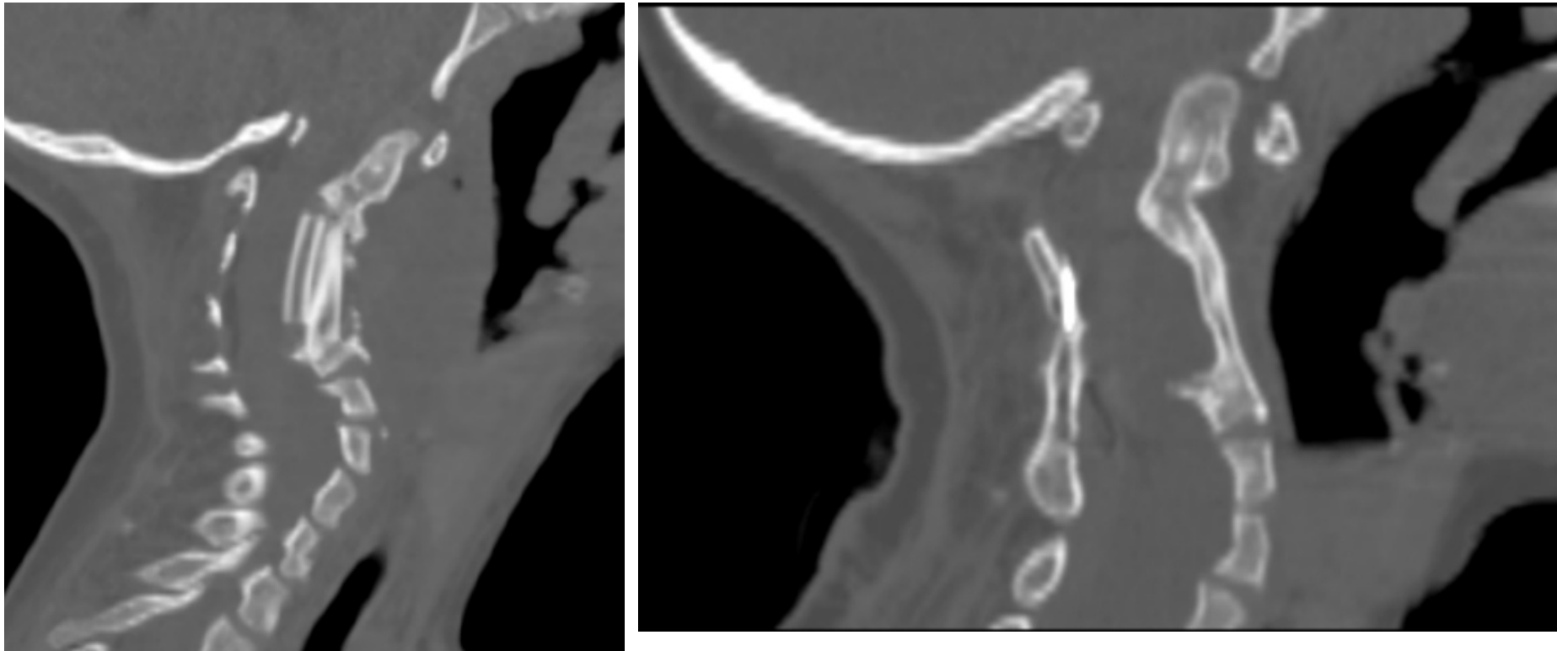
Stabilization with halo and anterior spinal cord decompression



C5 nerve root injury during osteotomy closure, recovery 3/5
Graft dislocation

Courtesy of Prof. William Mackenzie, A. I. Dupont Hospital for Children, Wilmington

Vertebra erosion continues (5 yr FU)

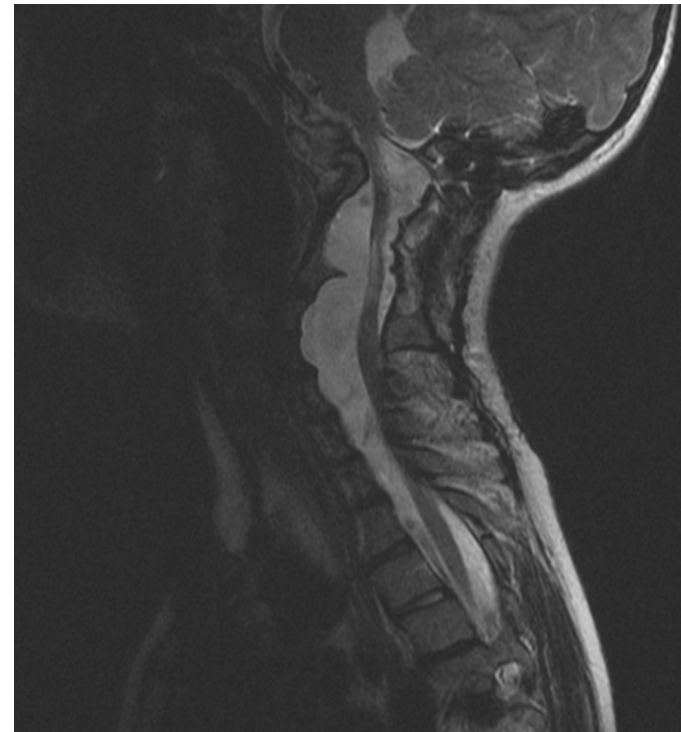
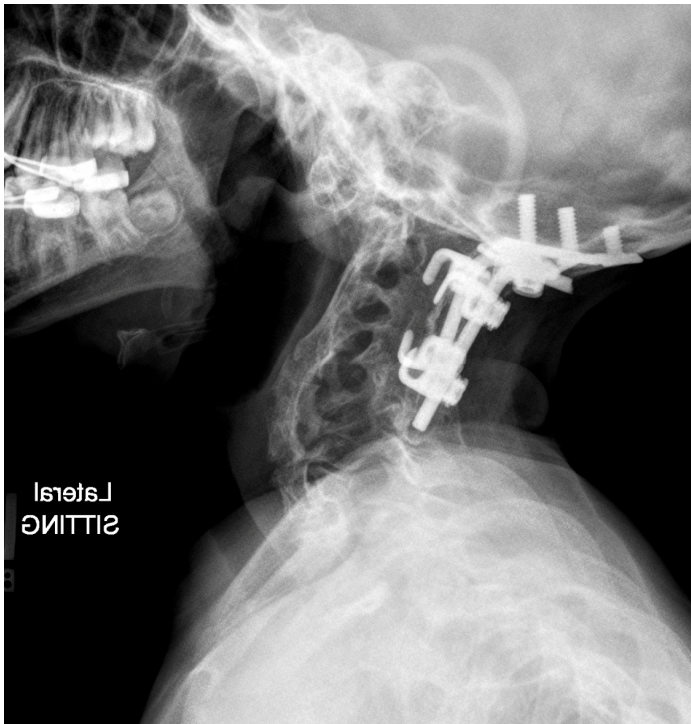


Risk of non-union after combined approach 0/13

New neurologic deficit: 5/22 (23%)

Junctional kyphosis observed: 8/22 (36%)

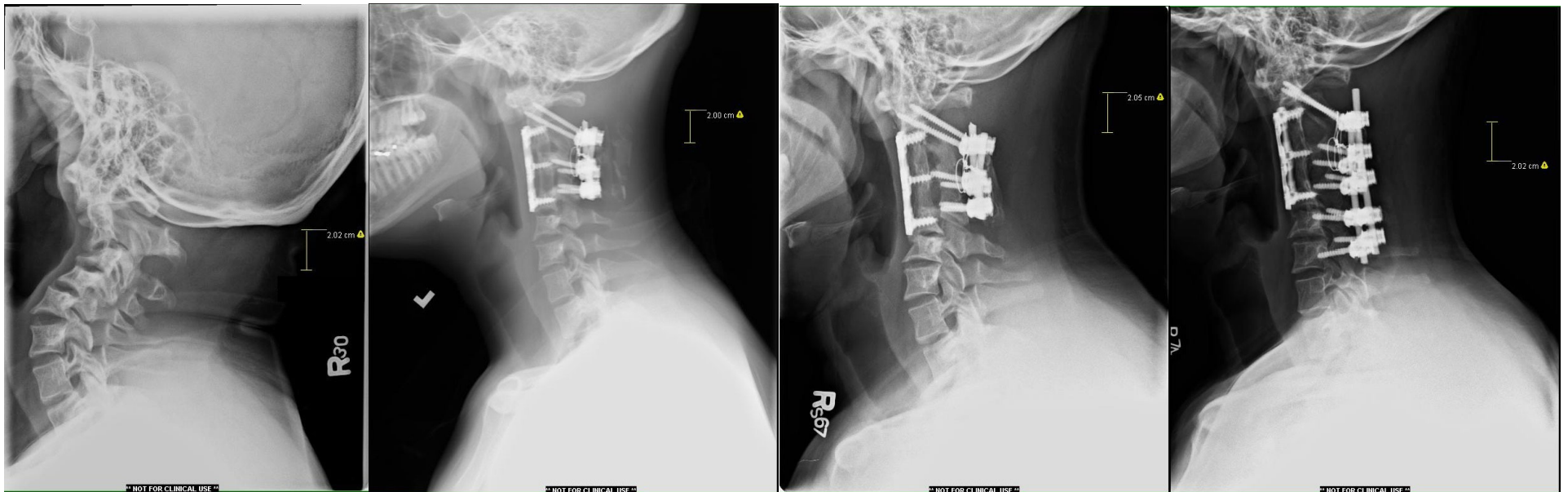
Seven years after initial surgery



- Normal neurological exam except for left deltoid weakness 3/5 (occurred after 1st surgery)

Courtesy of Prof. William Mackenzie, A. I. Dupont Hospital for Children, Wilmington

Length of spinal fusion and risk of junctional issues



Junctional kyphosis more in children with 5 or less levels fused ($p=0.054$).

Courtesy of Prof. Paul Sponseller, Johns University, Baltimore

Conclusions

- Continued vertebral erosion observed.
- Spinal union obtained with anteroposterior approach
- High rate of complications especially C5 nerve root deficits
- Junctional issues common
- Dural ectasia makes fixation points difficult
- Can we prevent these issues with instrumentation / laminoplasty at the time of neurosurgical tumor excisions



Special thanks: Paul Sponseller, William Mackenzie, Thierry Odent, John Dormans, Jahangir Asghar, Karl Rathjen, Joshua Pahys, Firoz Miyanji, Daniel Hedequist, Jonathan Phillips
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