Timing of Surgery for Cervical Spine Anomalies

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Disclosures

- IH: Consultant and Grants to Institution from Medtronic

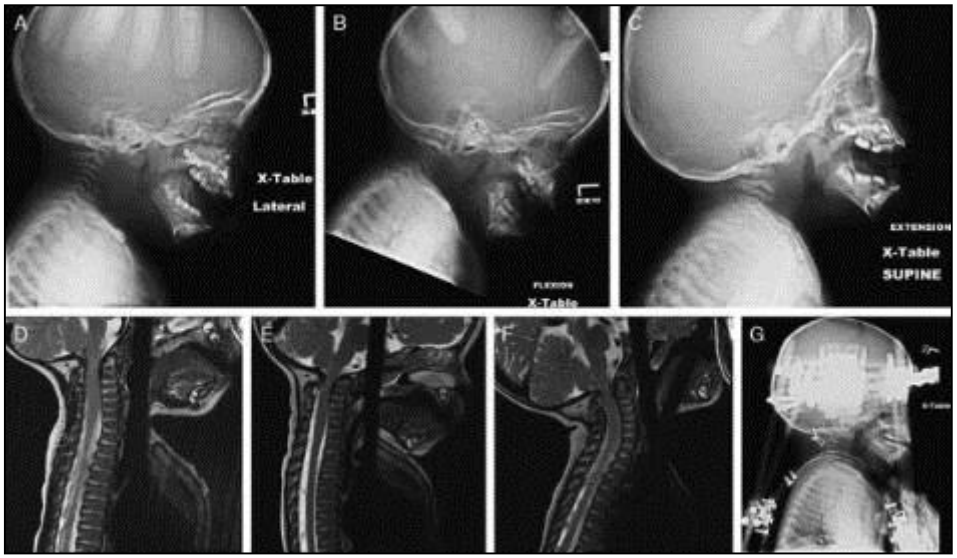
Pathology of Cervical Spine

- Instability
  - Occipitocervical (Down)
  - Atlantoaxial (Os odontoideum)
  - Subaxial (Klippel-Feil, Diastrophic dysplasia, Larsen syndrome)
- Deformity
  - Cervical kyphosis (Congenital, Neurofibromatosis)
- Spinal stenosis
  - Achondroplasia

Cervical Spine Instability

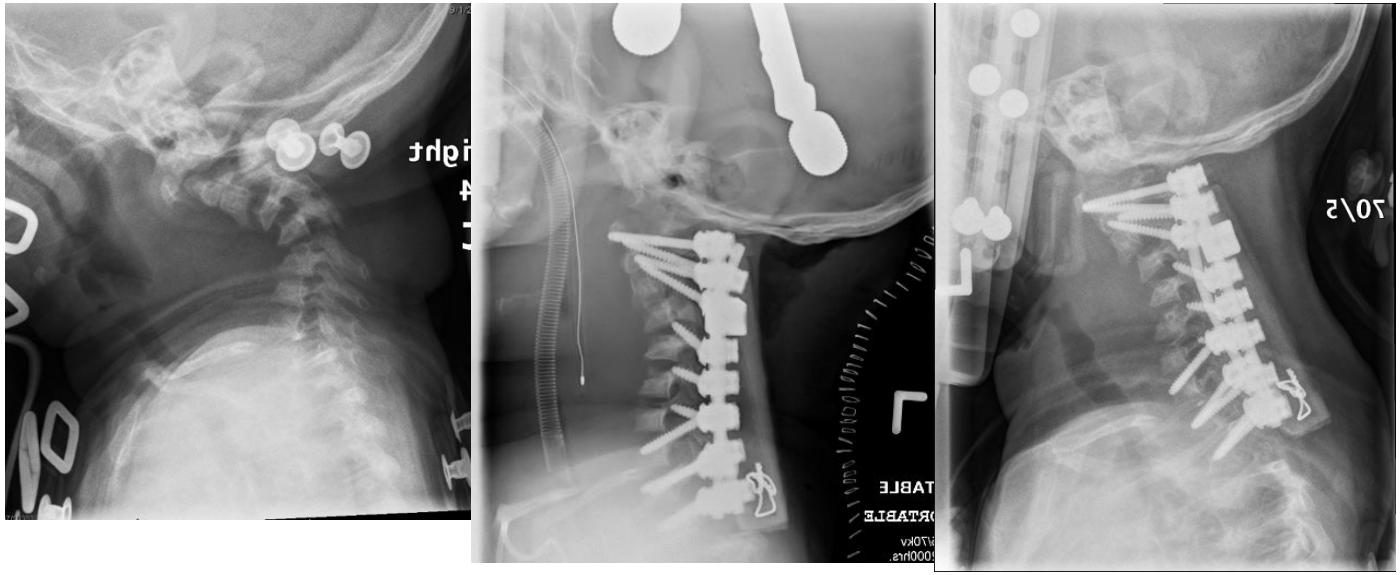
- Occipitocervical instability (C0/C1)
  - Basilar invagination: Tip of dens above level of foramen magnum (McRae line) (Chamberlain WE. Yale J Biol Med 1939;11:487-96)
  - Instability (Down, 22q11.2 Deletion syndrome): >1 mm of translation at the occipitoatlantal articulation (measured from the anterior surface of the occipital condyles to the posterior surface of the anterior arch of the atlas) (Wiesel SW. Spine 1979;4:187-91.)
- Atlantoaxial instability (C1/C2)
  - Atlantoaxial distance ≥5 mm (Locke GR, AJR 1966;97:135-40)
  - Space available for cord (SAC) < 13mm (Spierings and Braakman, JBJS Br 1982;64-B:422-8)
  - Os odontoideum, skeletal dysplasias such as spondyloepiphyseal dysplasia
- Subaxial instability
  - Vertebral body hypoplasia (congenital, diastrophic dysplasia)
  - Klippel-Feil with block vertebrae
  - Vertebral dysplasia and erosion (Neurofibromatosis)
  - Postlaminectomy (without instrumentation)

Flexion-Extension MR images



- Bony landmarks often difficult skeletal dysplasia
- Flexion-extension MR images diagnostic and safe (MacKenzie et al. JPO 2013)
- Protocol in children skeletal dysplasia
  - General anaesthesia or sedation. Radiologist and anesthesiologist available
  - T1+T2 sagittal, axial in neutral position (external auditory meatus at the level of shoulders)
  - If no cord compression / airway considerations continue with dynamic MRI
  - Flexion: Padding under occiput
  - Padding under shoulders
- Findings
  - Space available for cord, critical value less than 13 mm
  - Cord encroachment, signal changes
  - Soft tissue mass not shown in plain filmas or CT scan (Morquio) (Dede et al. JBJS 2013)

C Spine Instrumentation before 2 yrs

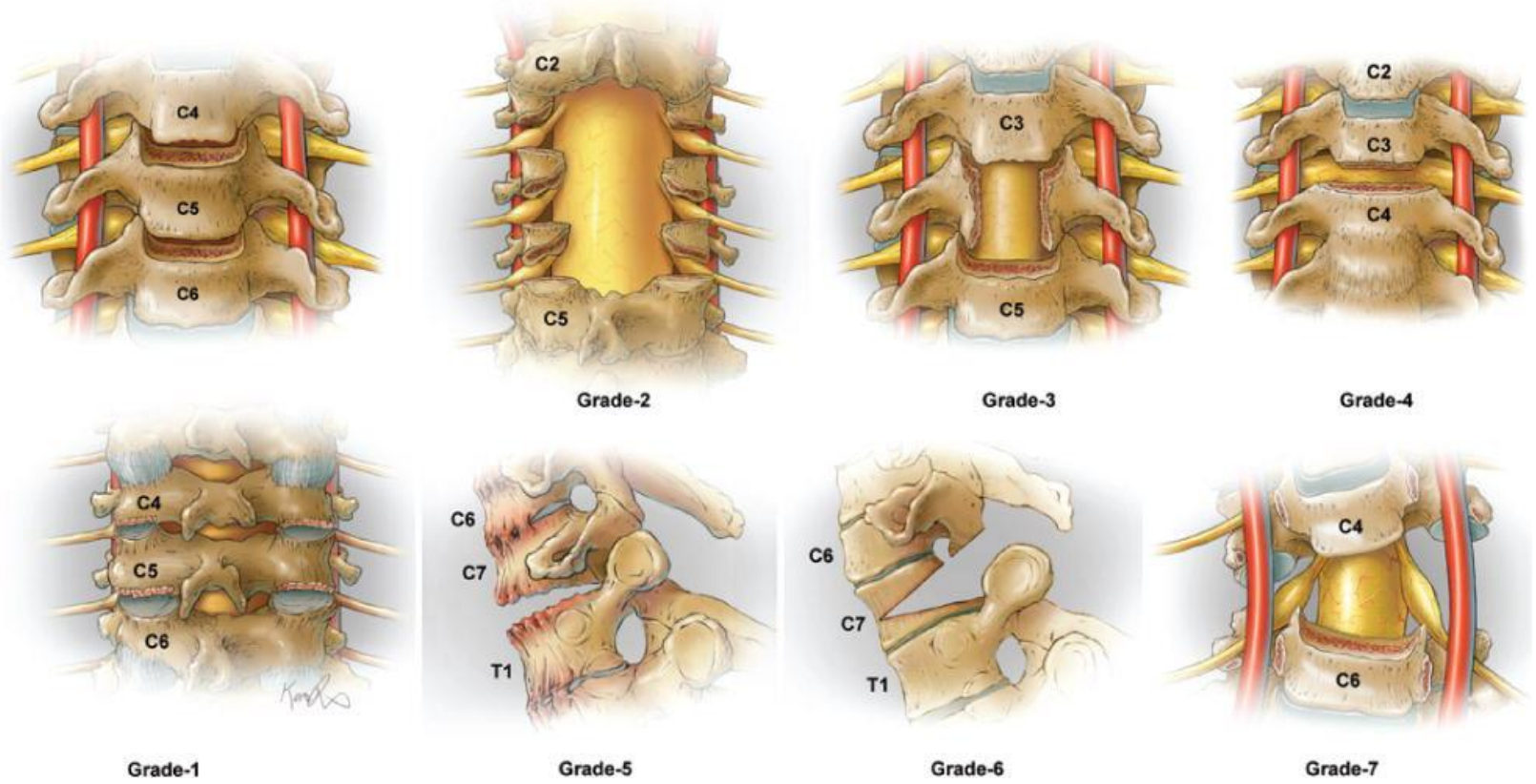


1.7-yr-old with NF1
Courtesy of
Dr. Firoz Miyangi

Cervical kyphosis in NF1
(Helenius I et al. JBJS 2016)

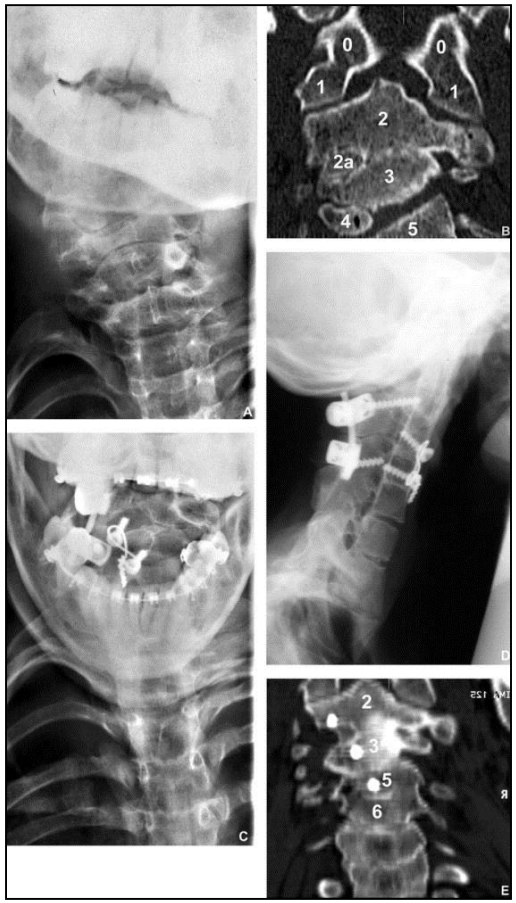
- Anatomic dimensions on CT more important than age (Lastikka et al. JCO 2017, in press)
- Occipitocervical fusion before the age of 2 years (Hale et al. Child Nerv Syst 2017;33:1253-69)
  - 10 infants with traumatic C0/C1 instability or C2 fx
  - 10 infants with congenital instability (3 with Down sdr, 3 with skeletal dysplasia, 4 with congenital anomaly
  - Complications: transversal sinus injury (1), worsening neurology (2), all fused
  - C1 lateral mass, C2 pedicle or translaminar screws can often be used; strut graft useful (rib, iliac)

Osteotomy classification of Cervical Spine



Ames CP et al. J Neurosurg Spine 2013;19:269-78.

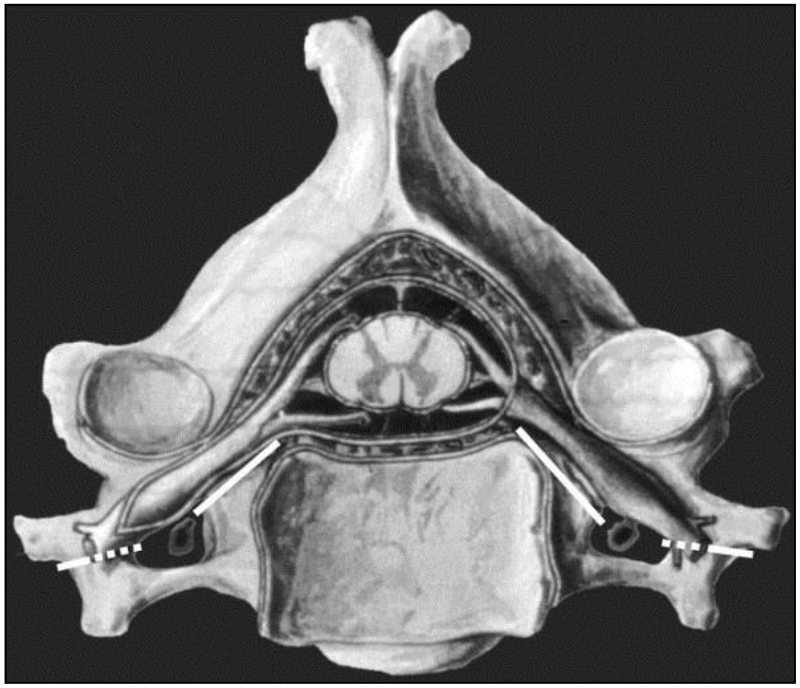
Hemivertebra Resection (C Spine)



Hemivertebra Resection in the Cervical Spine.
Ruf M et al. Spine. 30(4):380-385, February 15, 2005.

- Traditional treatment for C spine hemi: Posterior in situ with/o instrumentation (Hensinger RN. Clin Orthop 1991)
- 1st description of cervical hemi resection by Deburge and Briard JBJS 1981 (C7, four staged surgery)
- Above C7 (in the vertebral artery area) described by Harms and Ruf (Spine 2005)
  - Combined posteroanterior or posteroanteroposterior approach
  - Three children, one developed C5 nerve palsy
- Harms and Ruf advocated early hemi resection to prevent compensatory curves
- Indications
  - Head tilt, prevention of compensatory curves if fully segmented hemi

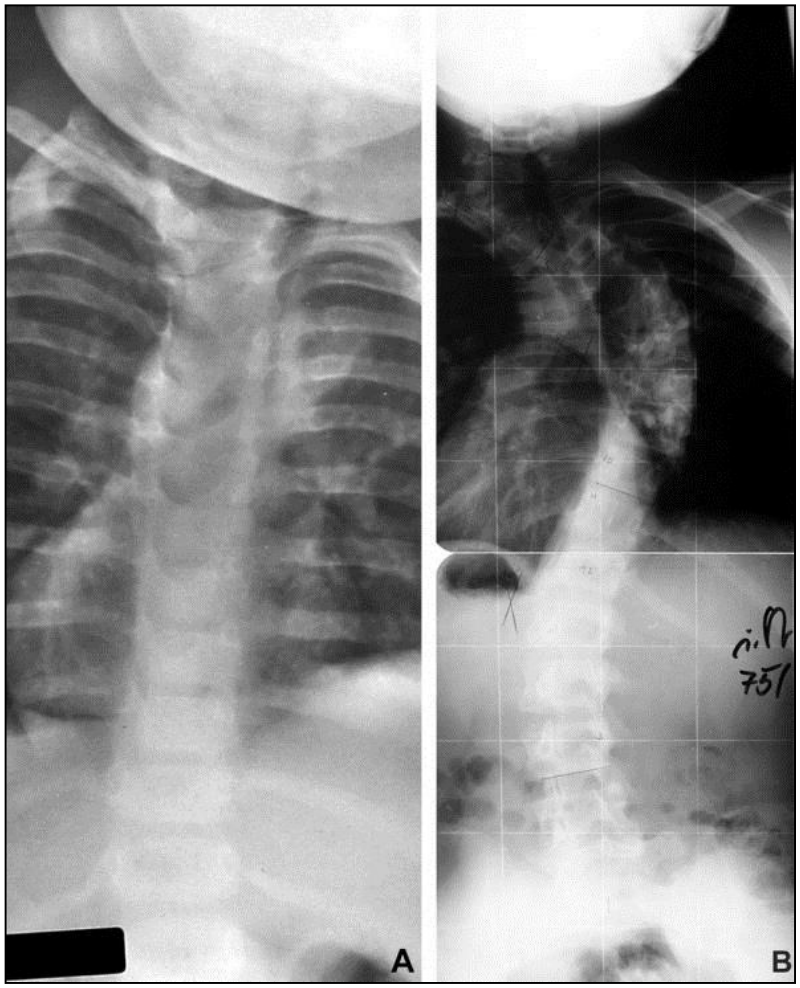
Technical aspects of Hemivertebra Resection



Hemivertebra Resection in the Cervical Spine.
Ruf M et al. Spine. 30(4):380-385, February 15, 2005.

- Posterior exposure
  - Hemilamina excision with facet joint above and below
  - Nerve root exposure, divisions of nerve root – vertebral artery connections
  - Resection of posterior wall for transverse foramen
- Anterior exposure
  - Disc above and below excised
  - Hemicorpus resected piecemiel
  - Transverse process resected with anterior wall of transverse foramen and exposure of vertebral artery
  - Anterior closure of resection with plate, if necessary posterior instrumentation with C2 pedicle and lateral mass screw

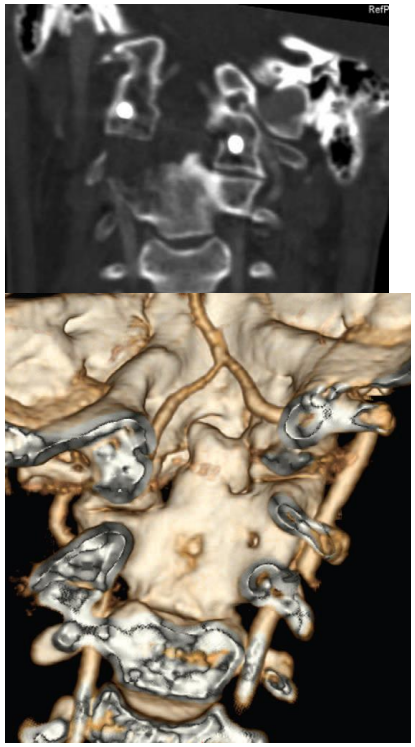
Timing and Effects of C Spine Deformity



Natural course of a cervical congenital scoliosis in a boy with hemivertebrae C3a and C7a at the left side. At the age of 2 years, the thoracic spine was completely straight (A). At the age of 14 years, there was a compensatory, now structural curve of 79[degrees] at the thoracic spine (B). Ruf et al. Spine 2005

- Harms and Ruf advocated early hemi resection to prevent compensatory curves, age ranged 4.4 to 14.9 in three patients (Spine 2005).
- Anatomic dimensions need to be satisfactory for implant placement (Lastikka et al. JCO 2017)

Craniocervical Junction Hemi



- Described on adult patient (Ruf et al. Spine 2015)
- Visualization of basilar & vertebral arteries preoperatively
- Transoral resection + posterior compression

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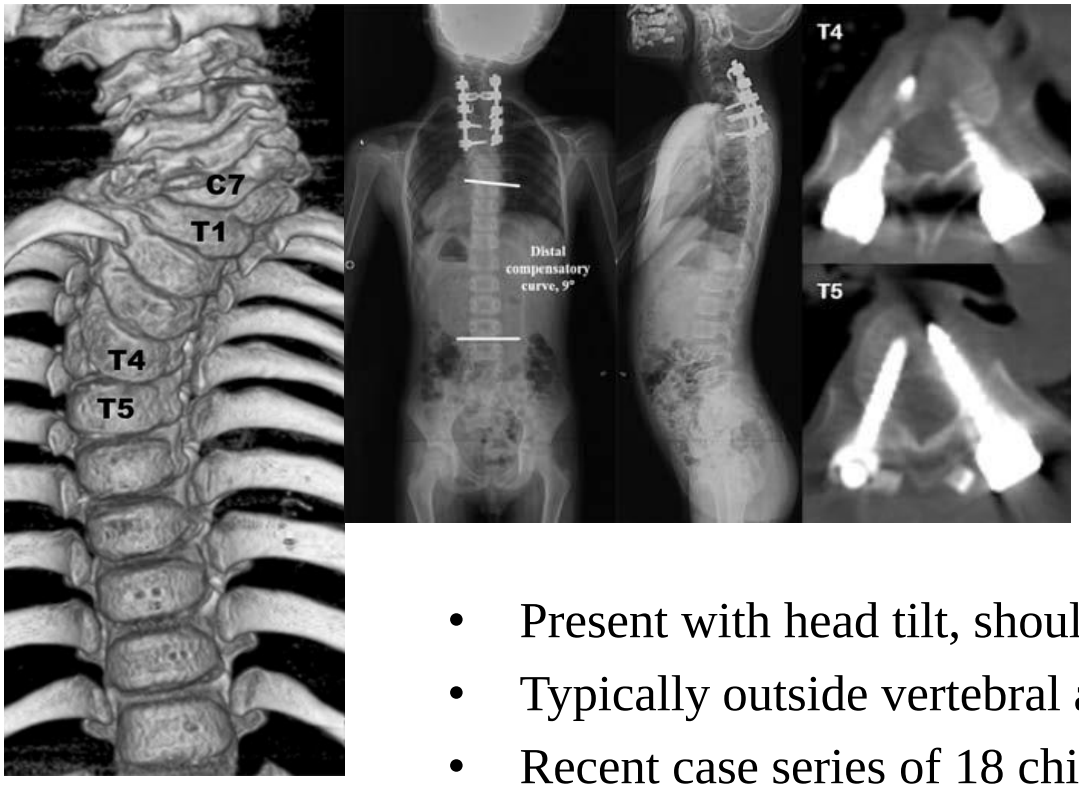
Cervicothoracic Juntion Hemivertebra



- Present with head tilt, shoulder imbalance
- Typically outside vertebral artery area
- Recent case series of 18 children (aged 5.5 to 15.2 yrs) with hemis between C7 and T4 (Chen et al. Spine 2017, Jul 11, epub)
  - 58% scoliosis correction with hemivertebrectomy
  - Neck tilt: 20˚ → 11 ˚
  - One Horner sdr, one radicular pain
  - 20% pedicle screw malposition risk

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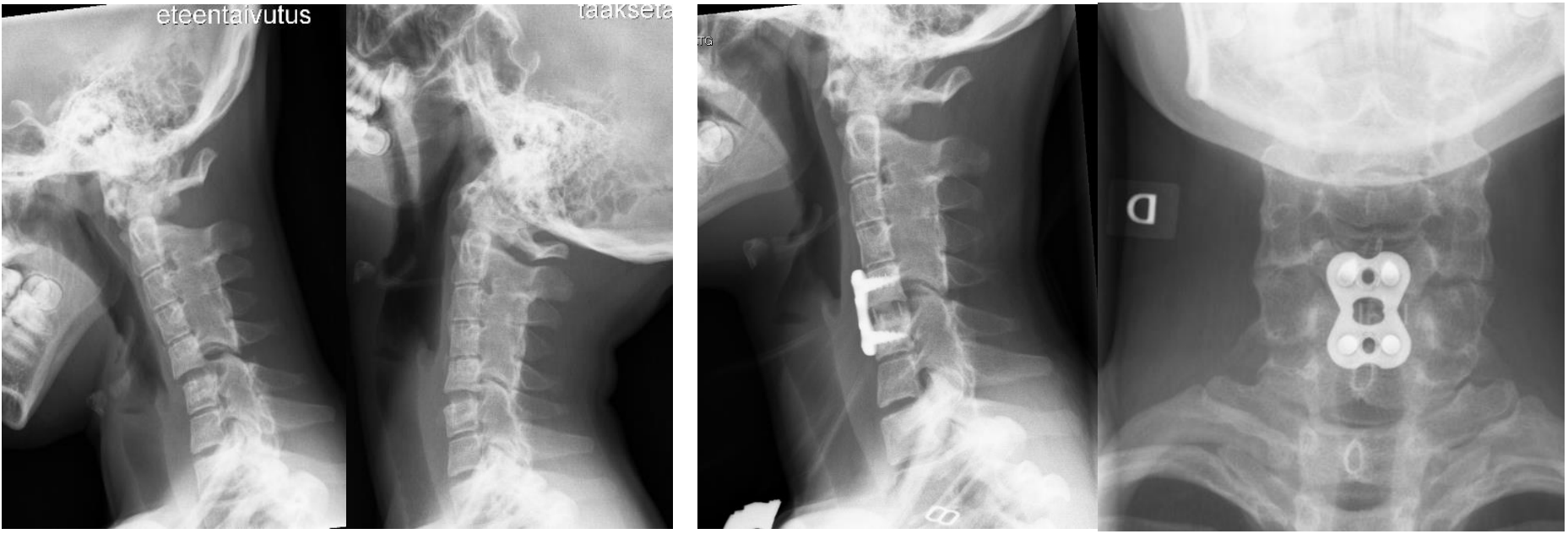
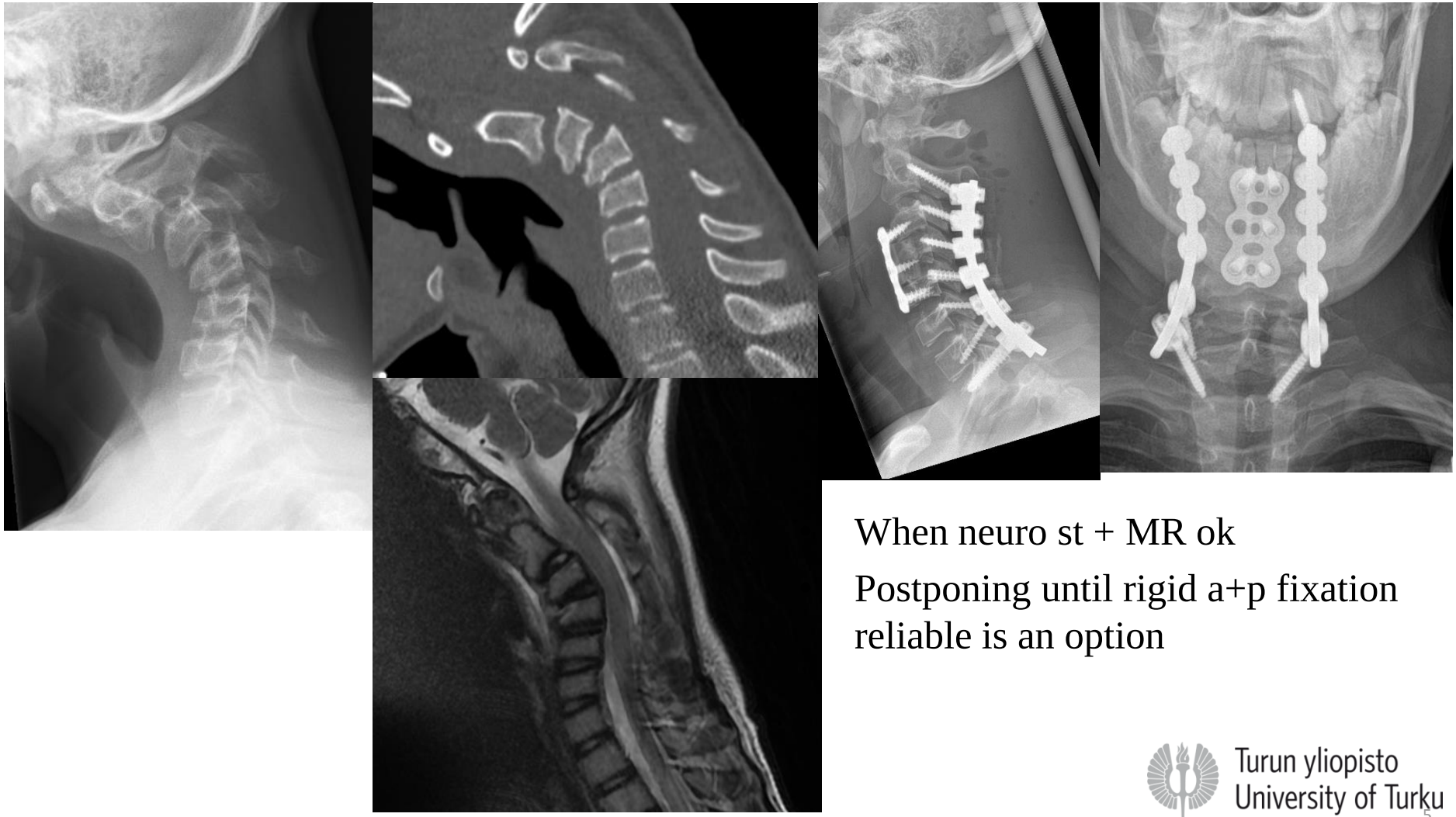
Klippel-Feil Instability



- Instability presents typically at adult age
- Contact sports denied
- Single level ACIF vs. posterior based instrumentation

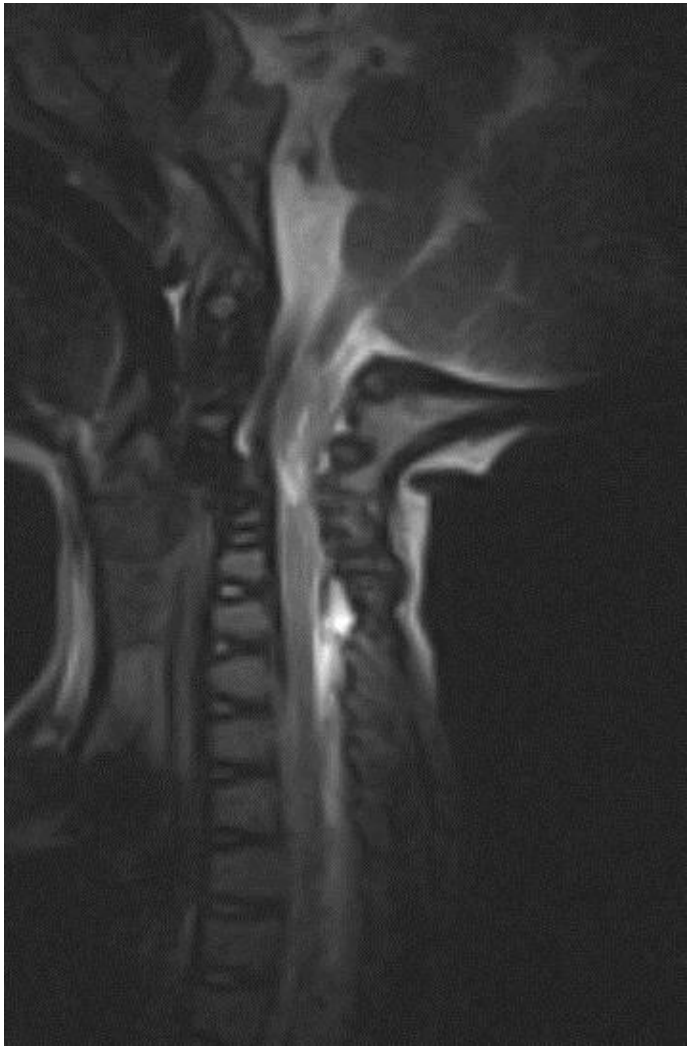
Correction of Congenital Kyphosis >10yrs



When neuro st + MR ok

Postponing until rigid a+p fixation reliable is an option

Chondrodysplasia punctata



5-yr-old boy

Mid-cervical kyphosis and stenosis

Associated myelopathy

Custom-made collar since 1-yr-old

Neurosurgical plan: Anterior decompression via C4 corpectomy and plate with postop Halo immobilization

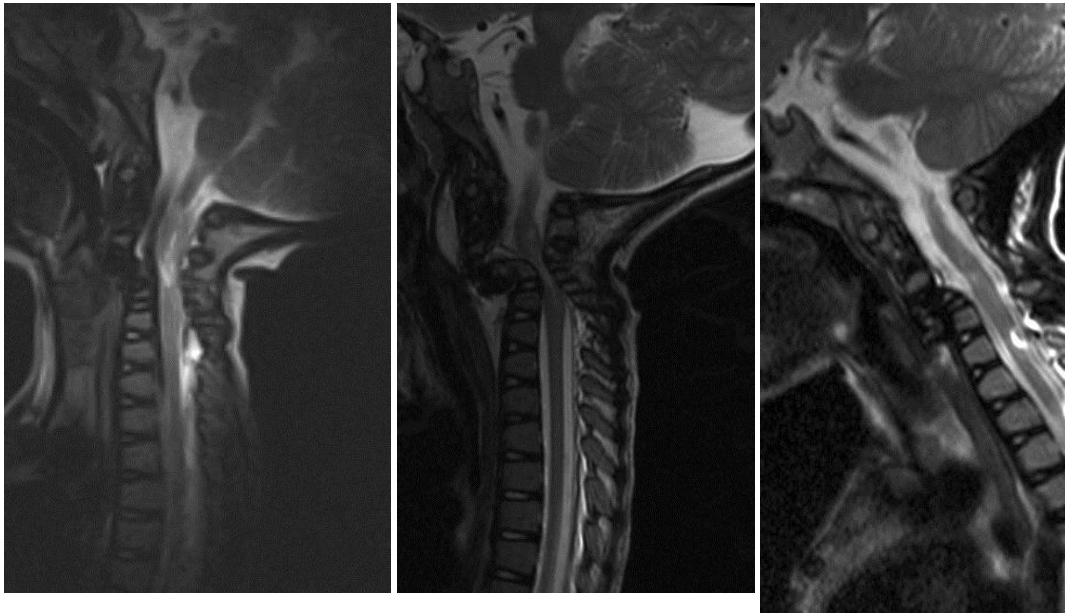
Immediate Postoperative MRI

Anterior corpectomy + biodegradable plate

No suitable halo body jacket available

Fixation failed and resulted into full tetraplegia + respiratory arrest (T2 sag MRI)

Posterior Decompression & C0-T2 Instr



Regained standing and eating but remains tetraparetic

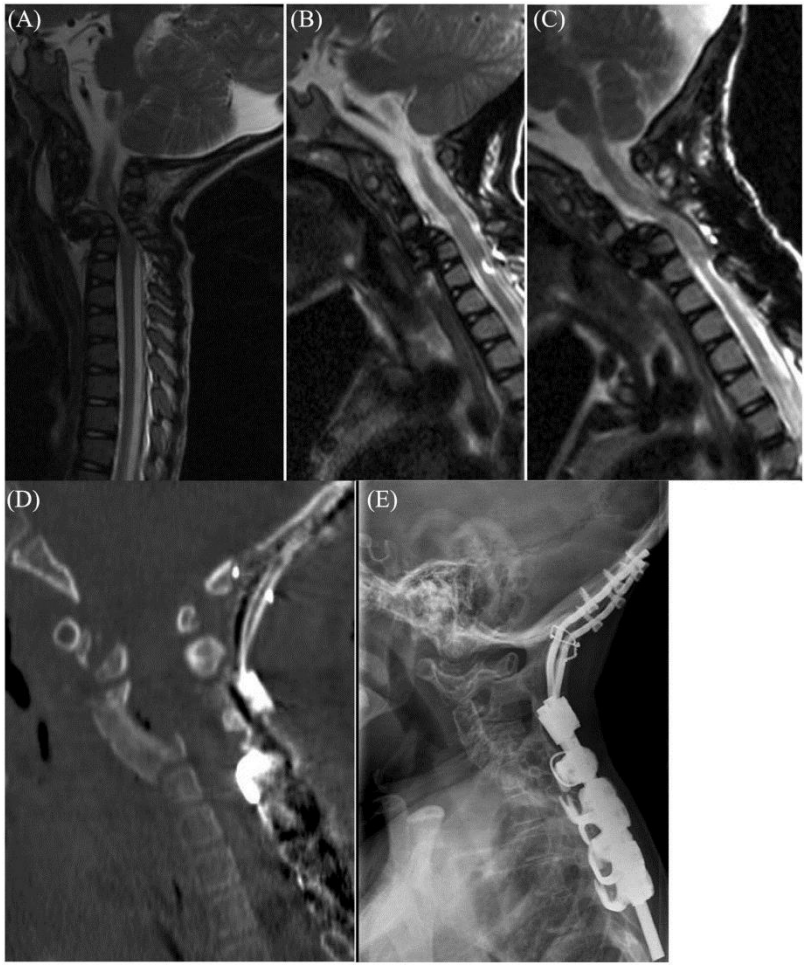
Posterior decompression and C0-T4 fix



Non-union at 1.5 years postop, no anterior column support?

Combined approach revision with anterior support

At 4-yr Follow-Up Union

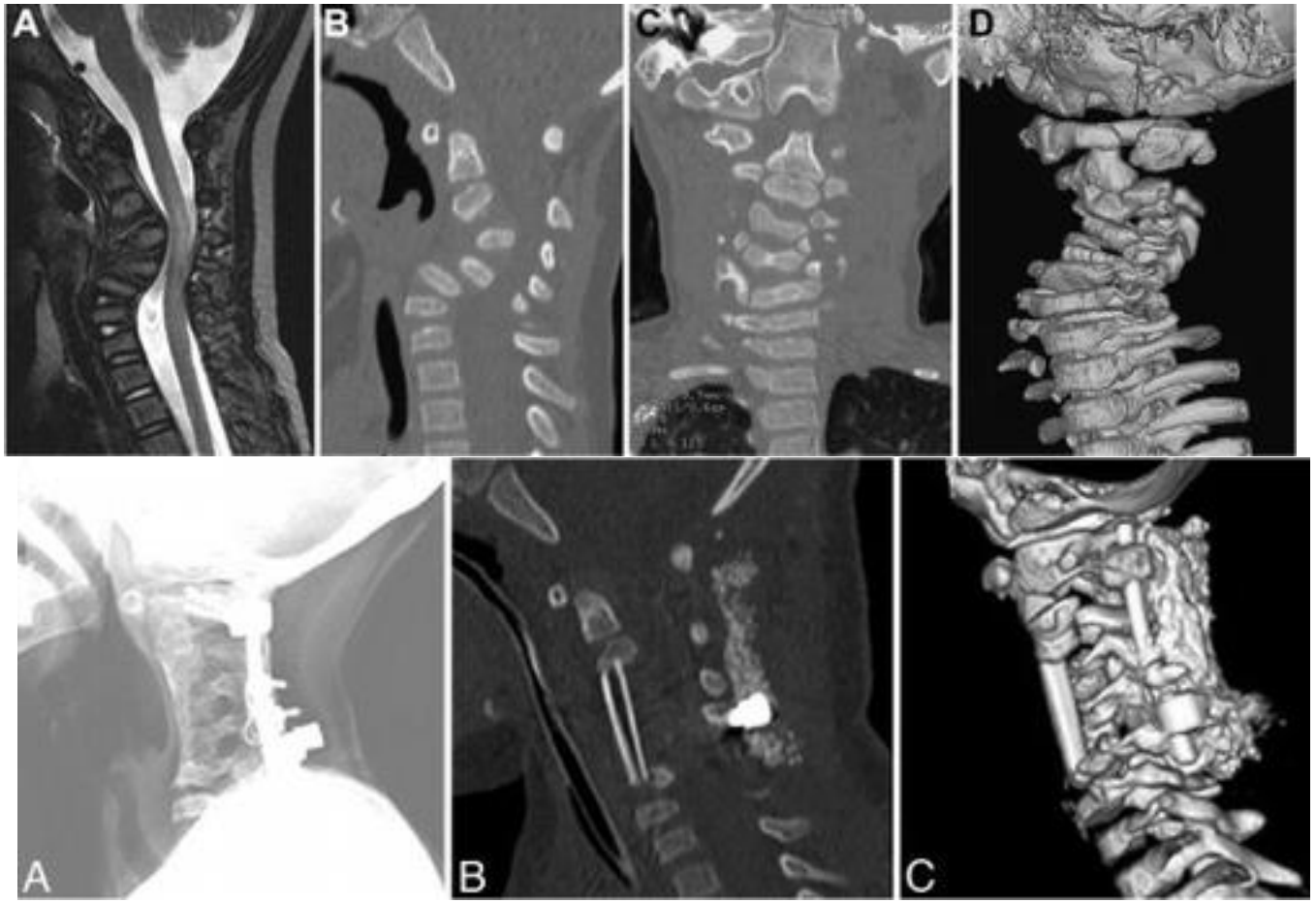


N. Pakkasjärvi; M. Mattila; V. Remes; I. Helenius; *Scandinavian Journal of Surgery* 2013;102:189-196.

Larsen’s syndrome

- Connective tissue disorder
- Congenital joint dislocations
- Progressive cervical kyphosis and risk of neuro deficit with minor trauma (Johnston et al. JBJS 1996)
- Anterior and posterior elements may become detached (Katz et al. JPO 2005)
- Guidelines missing for treatment
  - Combined approach preferred (Madera et al. J Neurosurg Ped 2008)

Case from Literature



Madera M et al. J Neurosurg Ped 2008 Apr;1:320-4

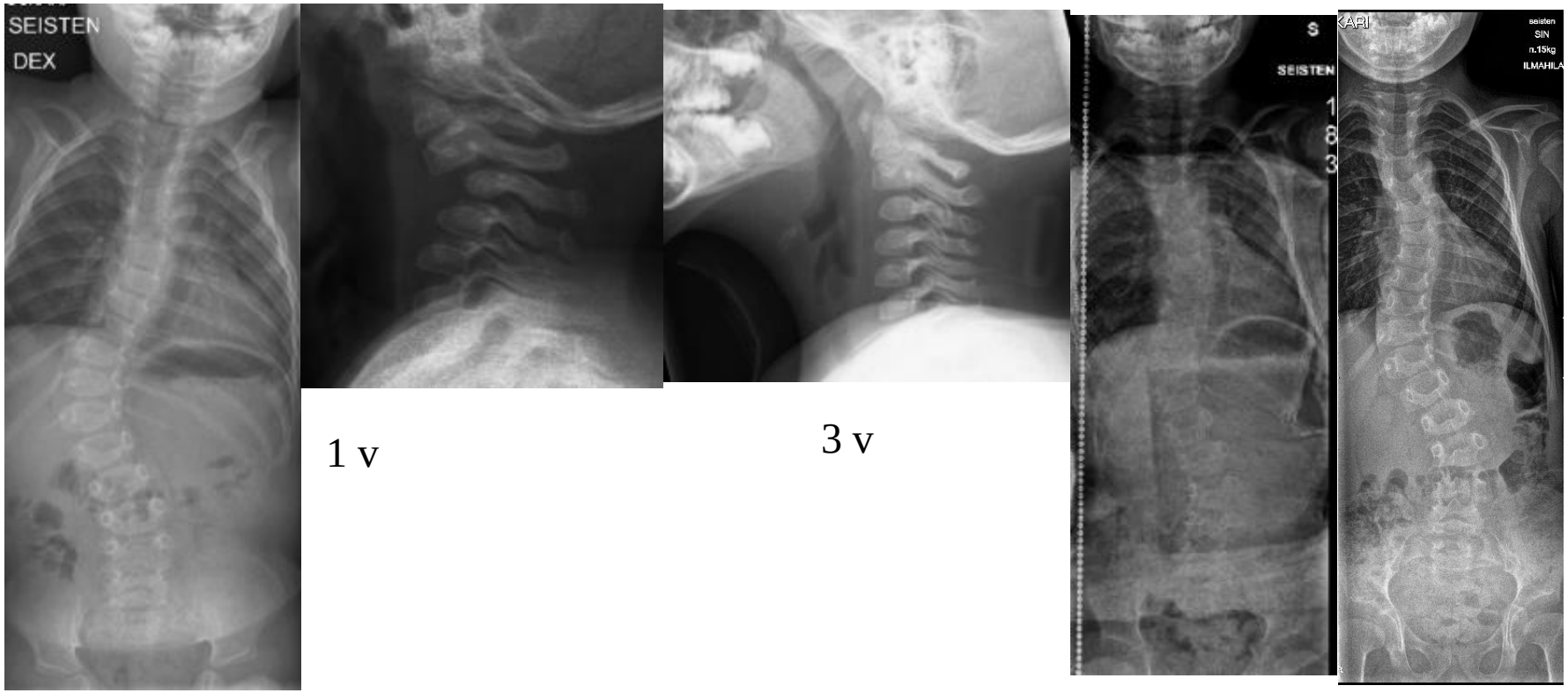
Spontaneous Correction in Diastrophic Dysplasia

- Kyphosis <60º → resolving
  (Remes et al., Spine 1999)
- Close follow-up 4 mths
  - Custom made collar 4-6kk->
- >60 º AP fusion
- Fiberoscopic intubation for all procedures (tracheomalacia)

1-yr-old

4-yr-old

Implications in Early onset scoliosis treatment



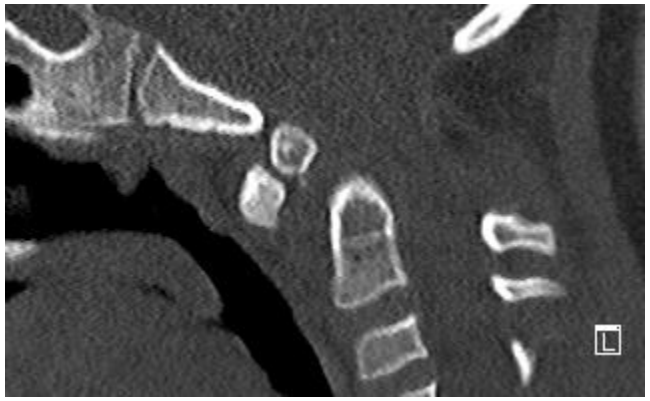
1 v

3 v

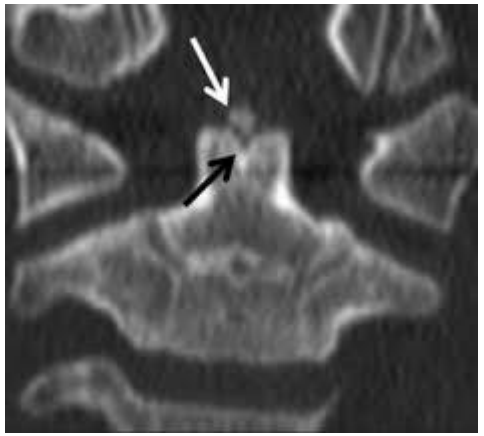
EOS typically highly progressive in Diastrophic Dysplasia (Remes et al. Spine 2002)
Cervical kyphosis prevents Glisson traction during casting of scoliosis – Use underarm traction instead. Cervical kyphosis resolves by continued growth if less than 60 deg (Remes et al. Spine 1999)

Os Odontoideum

- Os odontoideum
  - Bone (os), tooth (odontoideum), latin (Giacomini, Gior R Acad Med Torino 1886)
  - Lack of continuity between the odontoid process and the body of C2 (axis)
  - An independent ossicle with smooth cortical margins separated from a shorthened axis
  - Ossiculum terminale=non-union of secondary ossification center, not associated with C1/C2 instability
- Clinical Presentation
  - Asymptomatic
  - Neurologic deficits: Brainstem or spinal cord compression
- Two main anatomical types
  - Orthotopic: Ossicle associated with C1 anterior arch
  - Dystopic: Ossicle migrated towards clivus, functionally fused to the basion
- Idiopathic and associated with syndromes
  - Down syndrome, skeletal dysplasia
- Atlantoaxial instability (C1/C2)
  - Atlantoaxial distance (AAD) ≥5 mm (Locke GR, AJR 1966;97:135-40)
  - Anterior, posterior or combined
  - Measured betweenposterior border of anterior arch (C1) vs. Posterior border of body of axis



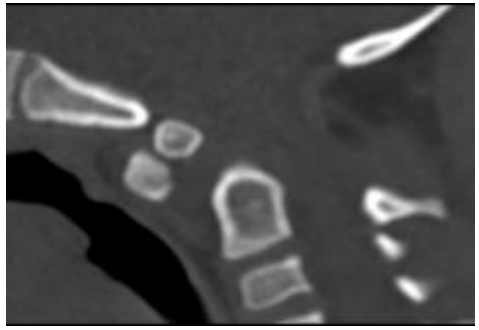
Os odontoideum with C1/C2 instability



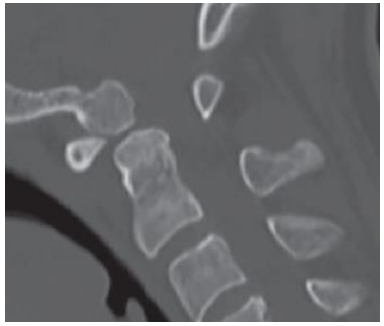
Ossiculum terminale

Radiographic findings

- Lack of continuity between the odontoid process and the body of C2 (axis)
  - An independent ossicle with smooth cortical margins separated from a shorthened axis
  - Enlarged anterior arch of C1
  - Jigsaw sign = articulation between anterior arch and the os
- Two main anatomical types (Fielding et al. JBJS 1980)
  - Orthotopic: Ossicle associated with C1 anterior arch
  - Dystopic: Ossicle fused to the basion
- Atlantoaxial instability (C1/C2)
  - Atlantoaxial distance (AAD) ≥5 mm (Locke GR, AJR 1966;97:135-40)
  - Space available for cord (SAC) <13mm
- Canal encroachment
  - Bony
  - Reactive synovitis



Orthotopic



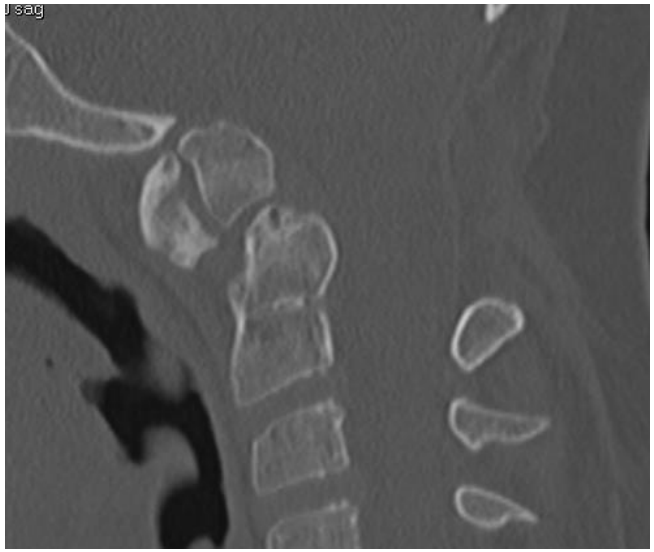
Dystopic

Reactive synovitis causing compression

Etiology

- Embryology
  - 4th occipital sclerotome (proatlas) → apical cap of dens and apical ligament
  - 1st spinal sclerotome (C1) → rest of dens
  - 2nd spinal sclerotome (C2) → axis body & arches
- Congenital
  - Fusion failure between the dens and body of atlas
- Traumatic
  - History of trauma common in os odontoideum
- Etiology varies (Sankar et al. Spine 2006)
- Atlantoaxial instability (C1/C2)
  - Atlantoaxial distance (AAD) ≥5 mm (Locke GR, AJR 1966;97:135-40)
  - Space available for cord (SAC) <13mm
- Canal encroachment
  - Bony
  - Reactive synovitis

Arvin et al. Neurosurgery 2010



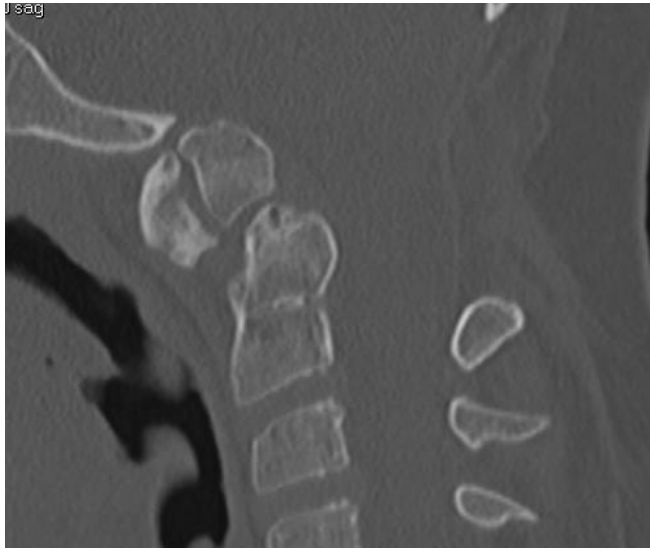
Conservative treatment

- 20 patients treated conservatively (Spierings and Braakman JBJS Br 1982)
  - 15 patients without neurologic deficits, FU 6.5 years, none developed neurologic deficits
  - 4 patients with transient cord signs, no deterioration

- Minor trauma associated with
  - Sudden death (Michaels et al JBJS 1969; Dempster et al. Am J Forensic Med 1990)
  - Quadriplegia, serious neurologic deficits, worsening of neurology (Clements et al. Injury 1995; Choi et al Ped Radiology 2005; Klimo et al JNS 2008)
  - Spinal cord atrophy (Fielding et al. JBJS 1980)
  - Cerebellar infarction (Sasaki et al. Spine 2000)

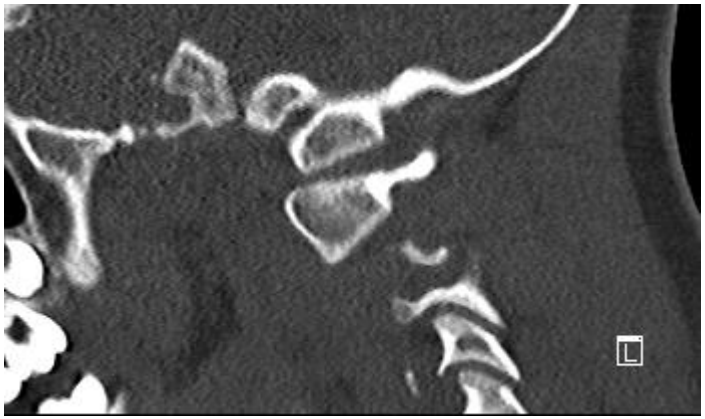
- Minimum requirements for conservative treatment
  - Normal cord morphology
  - Minimum SAC >13 mm
  - 10% risk of myelopathy with SAC<13mm (Spiering and Braakman JBJS Br 1982)

- Conservative treatment includes
  - Stable, yearly flexion-extension radiographs
  - MR images every 5 years to prevent signal changes
  - No contact sports
  - Requires further investigations!

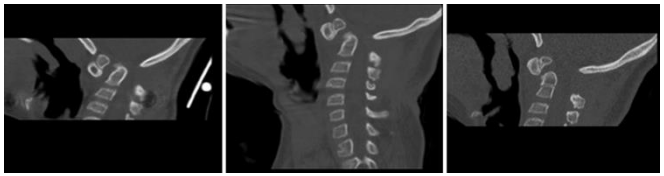


Preoperative considerations

- C1/C2 instability
  - AAD>4 mm, SAC<13mm
  - 10% risk of myelopathy with SAC<13mm (Fieding JBJS Br 1982)
- C1-C2 vs. C0-C2 spinal fusion
  - Down patients with dysplastic C0/C1 joints?
- Preoperative imaging
  - MR angiography: Vertebral arteries
  - CT: Bifid C1 posterior elements
  - Use of intraoperative navigation
- Preoperative traction (Hedequist et al. 2016)
- Fixation points
  - Occiput
  - C1 posterior elements, lateral mass
  - C2 posterior elements, pedicle
  - Transarticular screws

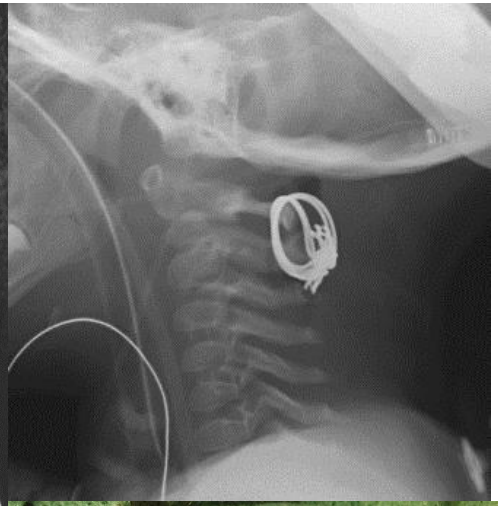


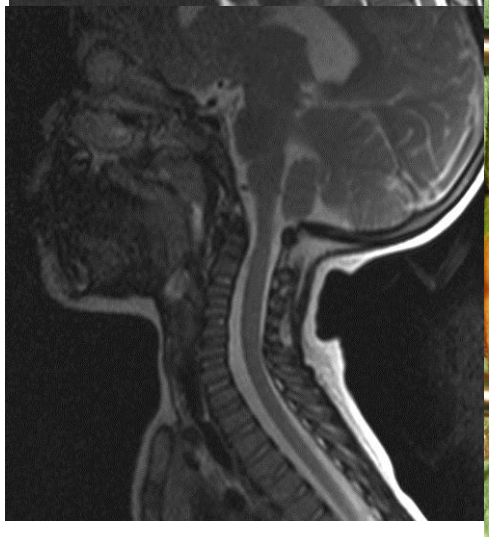
Dysplastic C0/C1 joints

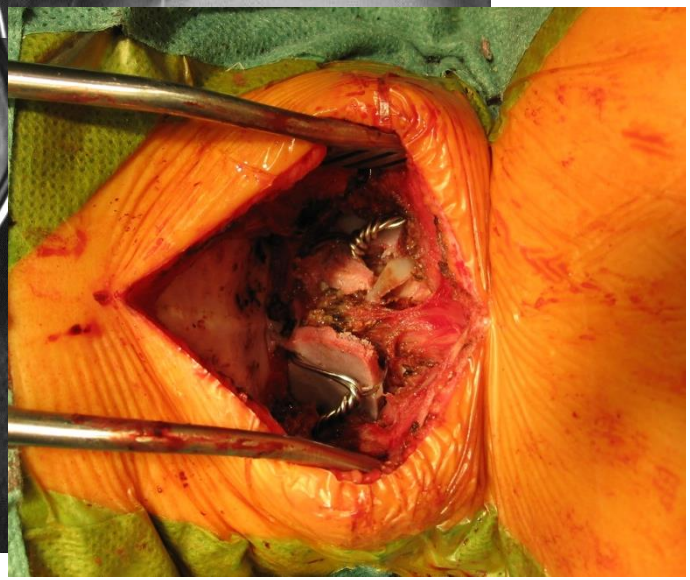


Effect of preop traction on alignment (El-Barr et al. J Neurosurg Ped 2016)

Brooks-Jenkins wiring

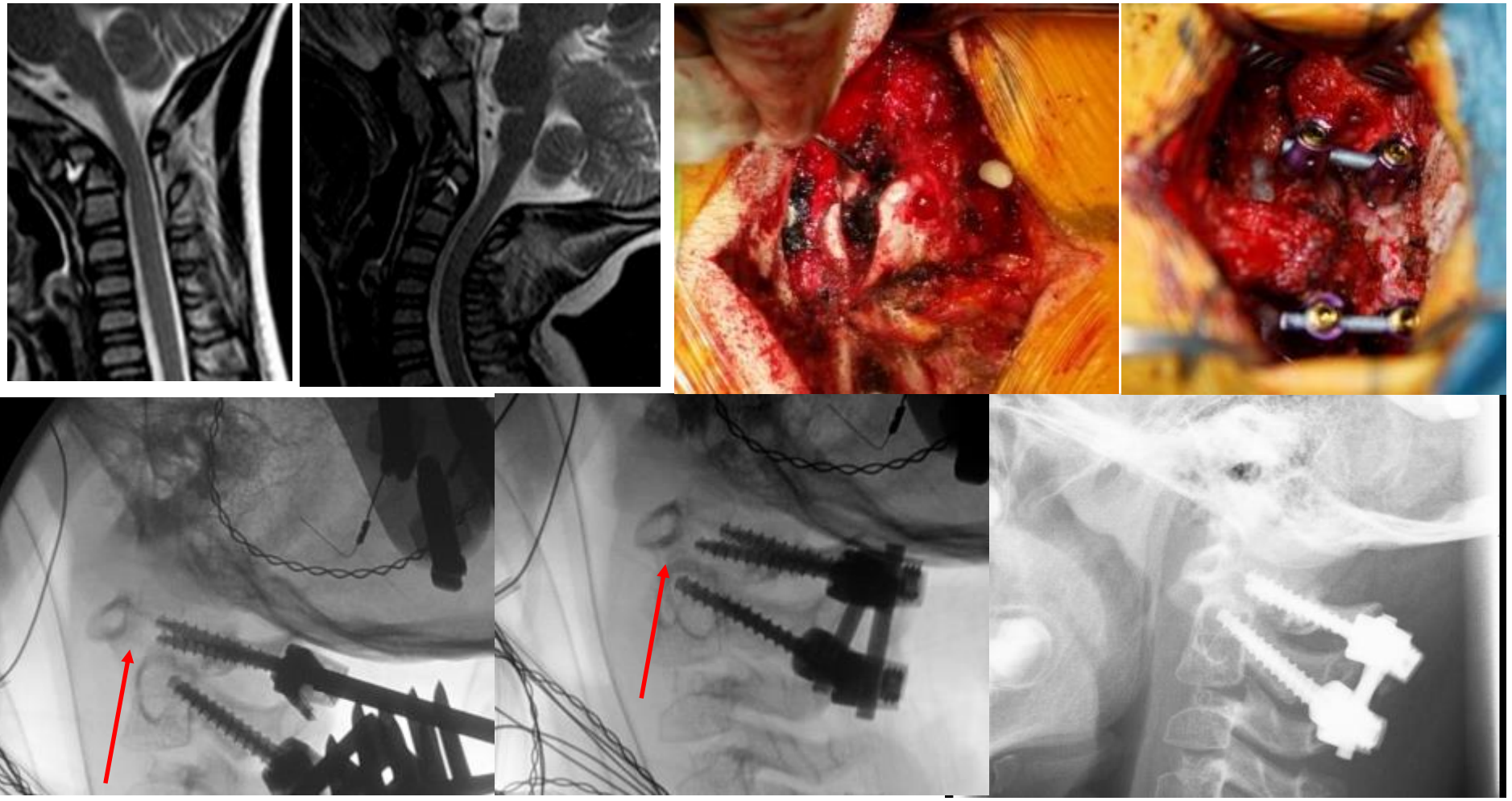






- Spondyloepiphyseal dysplasia
- 4-yr-old girl
- Odontoid hypoplasia
  - AAD 8mm or SAC 9mm

Harms technique (C1 lateral mass, C2 pedicle)



Rigid fixation improves fusion rates

- 28 children with skeletal dysplasia and C1/C2 instability (Helenius ym. JBJS 2015)
- 14 non-rigid (mostly wiring), 14 rigid (Harms or hooks)
- Risk of non-union higher in non-rigid 6/14 vs. 0/14 (p=0.0057)
- Two vertebral artery lesions with attempted C2 pedicle screw placement

Author's current practice

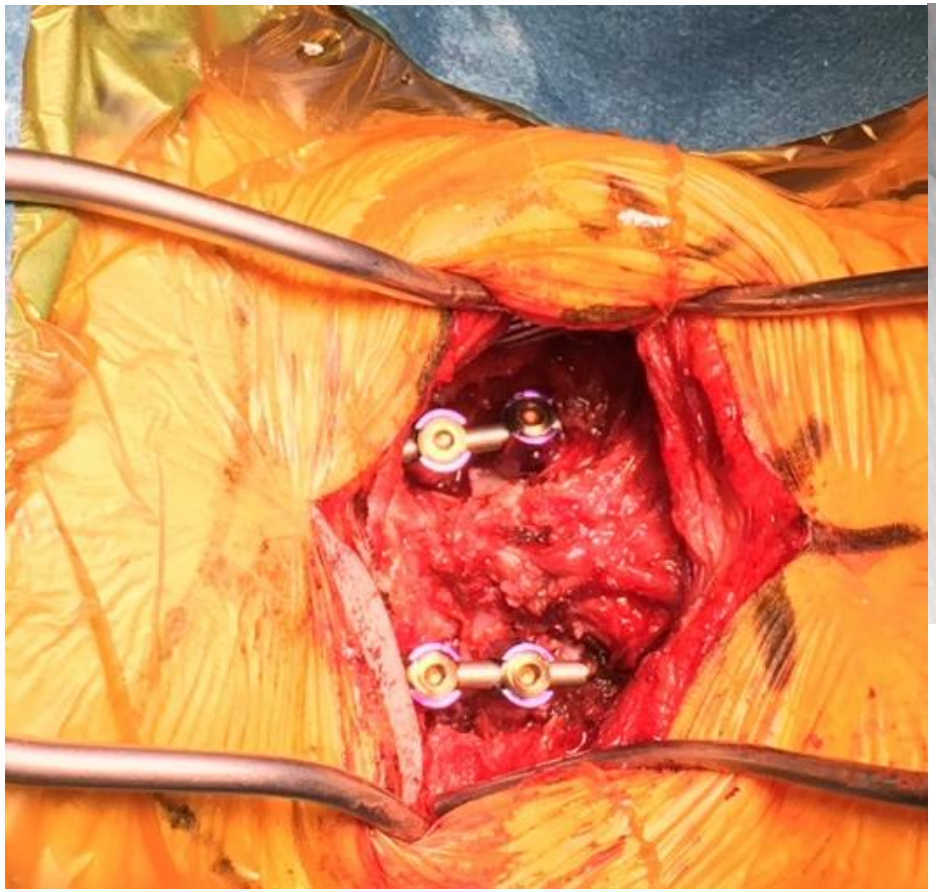
- Preoperative evaluation
  - MR: signal changes, cord morphology, anatomy of arteries (dominant vertebral artery?)
- Intraoperative traction
  - Halo + 1-2 kg traction
- Harms technique
  - C1 navigated + exposure of lateral mass (C2 sacrifice)
  - C2 free hand
  - Intraoperative O-arm to confirm reduction, implant placement
  - Avoid C0/C1 fusion
- Halo body jacket 2-3 months
- Custom made collar 3 months
- Contact sports not allowed

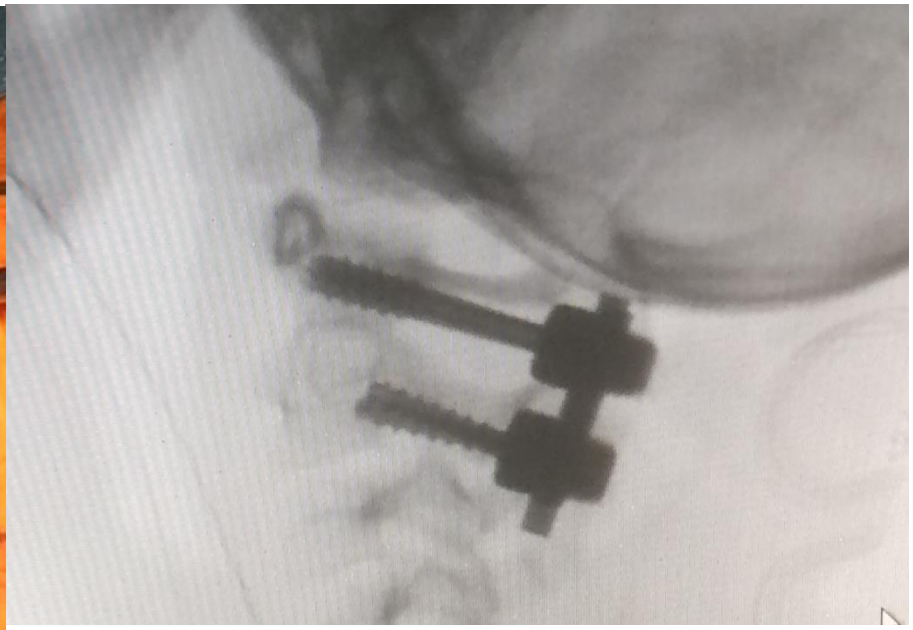
5-yr-old girl, head tilt, neck pain

Intraoperative traction

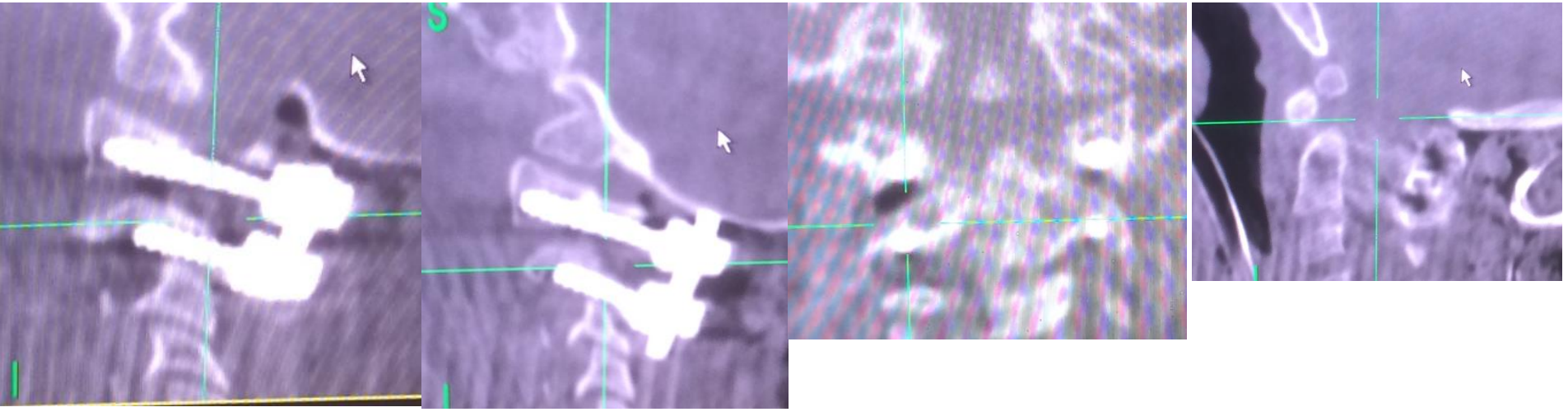
Make sure cervical alignment is acceptable with fluoroscopy

Implant placement





Intraoperative O-arm



Implant placement
Space available for cord
Alignment of os odontoideum of axis, avoid over/undercorrection

Conclusions

- Early surgery may prevent compensatory curves
- Anatomical dimensions need to be evaluated preoperatively
- If neurological status OK, postpone surgery until school age to allow stable, segmental, rigid fixation
- Ongoing study on Os Odontoideum by the Pediatric Cervical Spine Study Group
  - Idiopathic vs. non-idiopathic children
  - Conservative treatment
  - Operative treatment

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or jonathan.phillips@orlandohealth.com