Os Odontoideum

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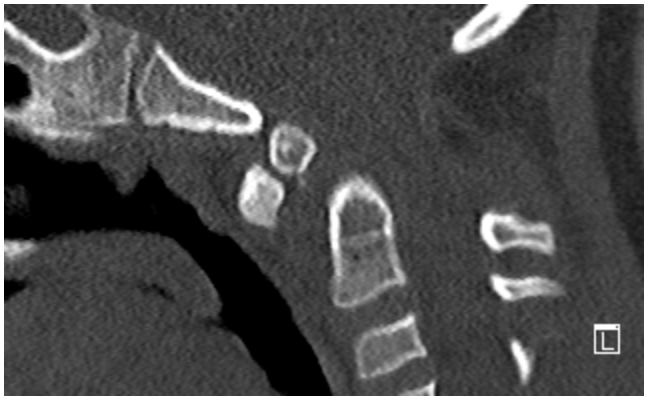
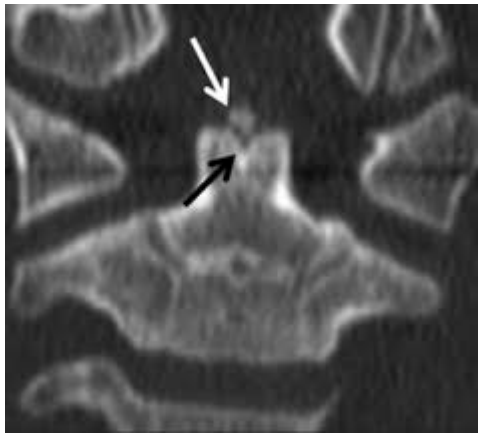
Os Odontoideum

- 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
  - Neck or occipital pain most common presenting symptom
  - Neurologic deficits: Brainstem or spinal cord compression
  - Incidental finding
- 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
  - History of trauma common
  - Down syndrome, skeletal dysplasia



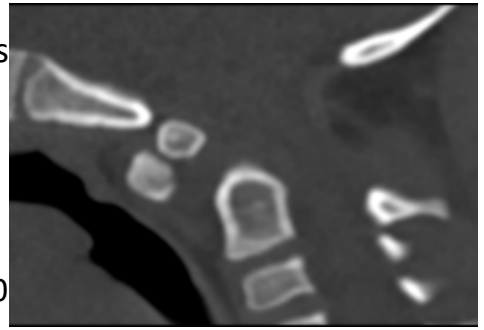
Os odontoideum with C1/C2 instability



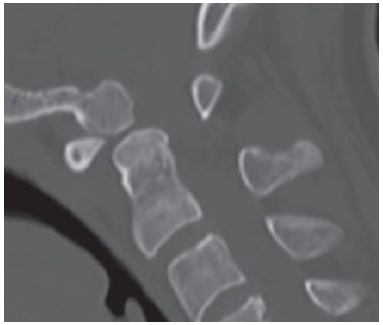
Ossiculum terminale

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 associated or 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
  - Anterior, posterior or combined
  - Measured betweenposterior border of anterior arch (C1) vs. Posterior border of body of axis
- Canal encroachment
  - Bony
  - Reactive synovitis



Orthotopic

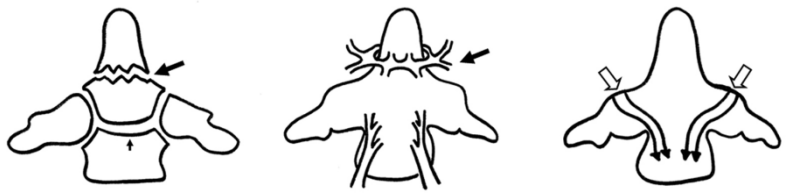


Dystopic

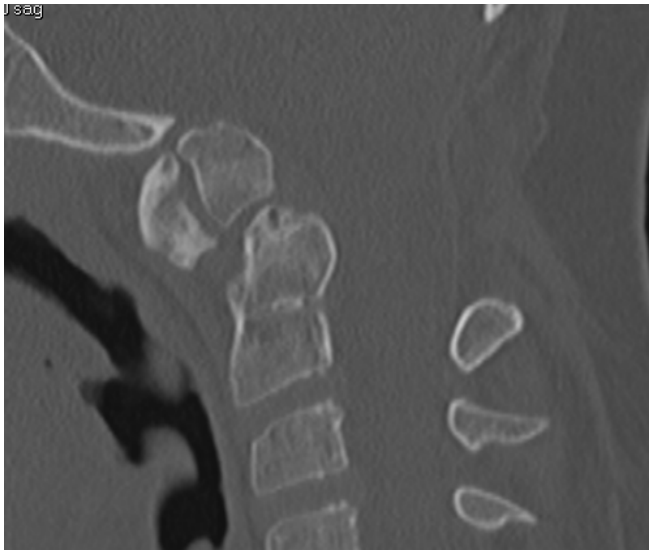
Reactive synovitis causing compression

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
  - Skeletal dysplasias
  - Associated with other congenital anomalies: 10% out of 279 (Zhao et al. Neurosurgery 2015)
- Traumatic
  - History of trauma common in os odontoideum: 40% out of 279 (Zhao et al. Neurosurgery 2015)
- Etiology varies (Sankar et al. Spine 2006)
  - Some patients traumatic background others congenital



Arvin et al. Neurosurgery 2010



Chromosomal aberrations

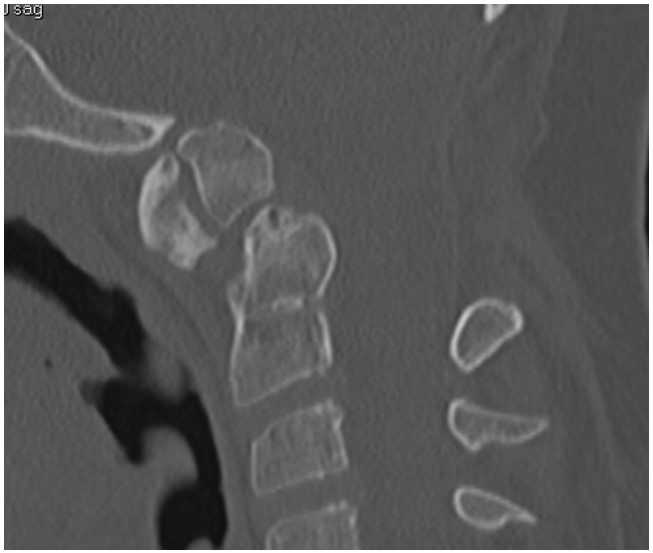
- Down's syndrome (trisomy 21): 1:733 live births
  - Craniocervical instability: 8-63%
- Skeletal dysplasias
  - Spondyloepiphyseal dysplasia, OI
- C0/C1 instability (dysplasia), C1/C2 instability, os odontoideum
- Role of asymptomatic instability/screening unclear in patients with Down's syndrome
  - 40 catastrophic cord injuries reported Down's syndrome
  - No child had neurologic deficit, 3 adults showed minor neuro deficits (22q11.2 Deletion syndrome)

Spondyloepiphyseal dysplasia

Down sdr

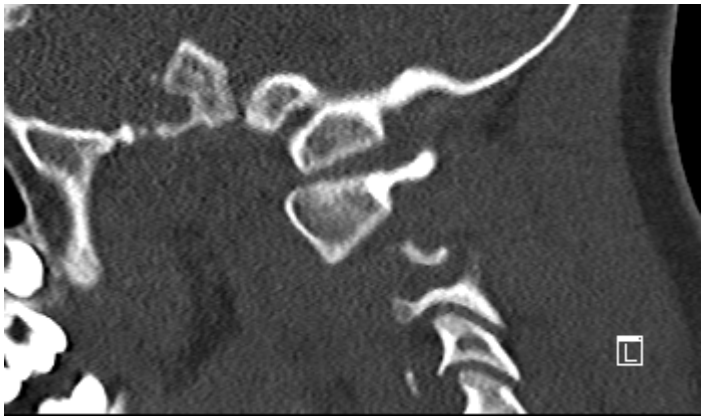
Conservative treatment

- 20 patients treated conservatively (Spierings and Braakman JBJS Br 1982)
  - 15 patients without neurologic deficits, FU 6.5 years, none developed neurologic deficits
  - Including 8 patients with C1/C2 instability (>7mm)
  - 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!

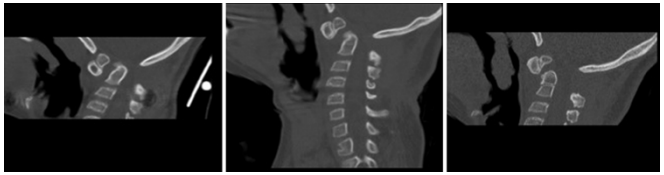


Preoperative considerations

- 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 w/o halo vest (Eö-Barr et al. 2016)
- Fixation points
  - Occiput
  - C1 posterior elements, lateral mass
  - C2 posterior elements, pedicle
  - Transarticular screws

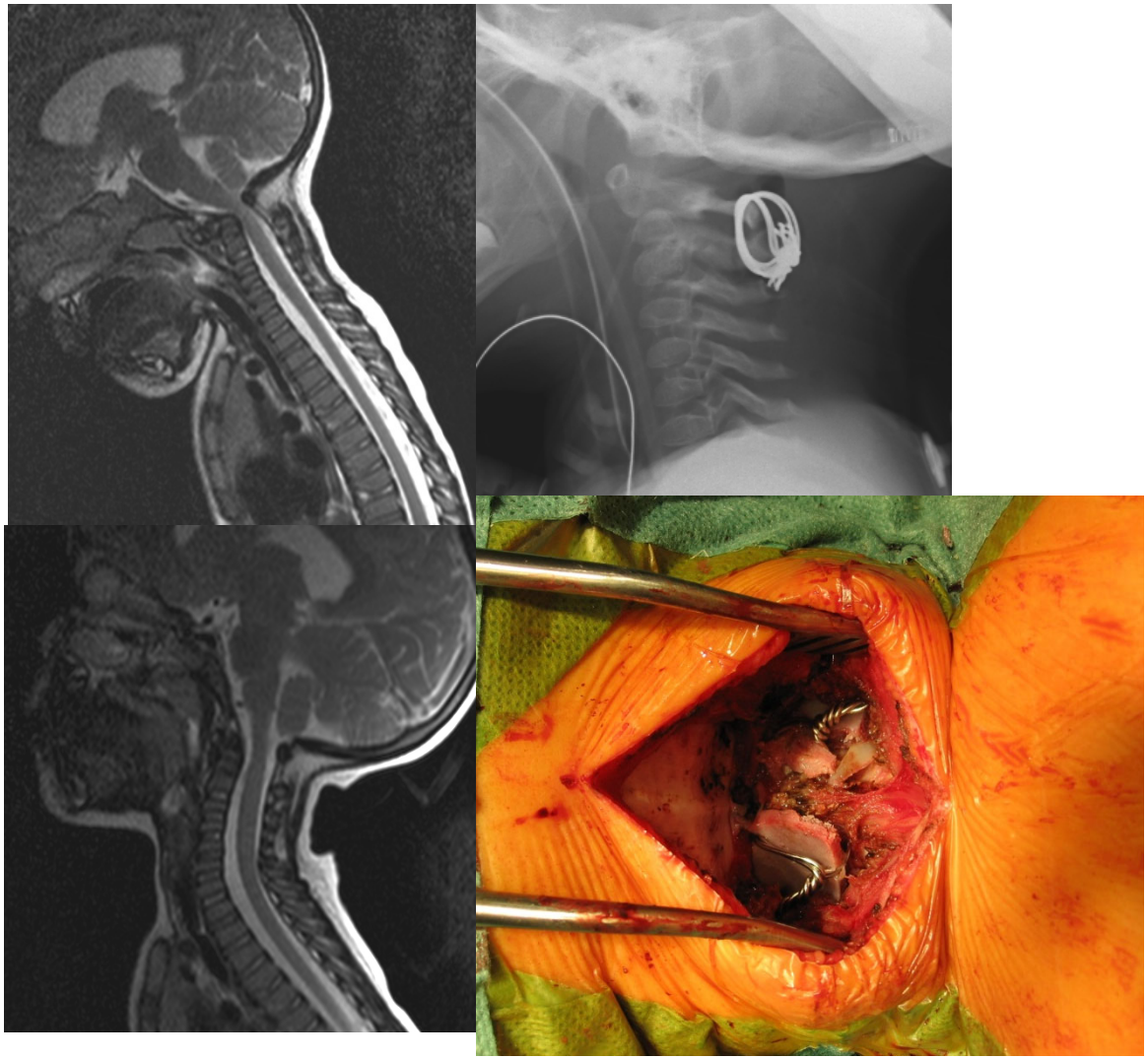


Dysplastic C0/C1 joints



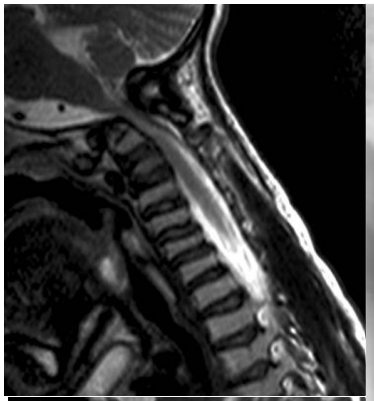
Effect of preop traction on alignment (El-Barr et al. J Neurosurg Ped 2016)

Brooks-Jenkins wiring



- Brooks-Jenkins and Jenkins JBJS 1978
  - Flexion prevented by wiring
  - Extension prevented by bone graft
  - 3 children with os odontoideum achieved fusion with Minerva cast
- Requires normal arches of C1 and C2
- 11 children with os odontoideum (Smith et al. Spine 1991)
  - 18% non-union
  - 1 child with cord injury with sublaminar wire passage

Case of non-union



1-yr pop

4-yr pop

Broken cable

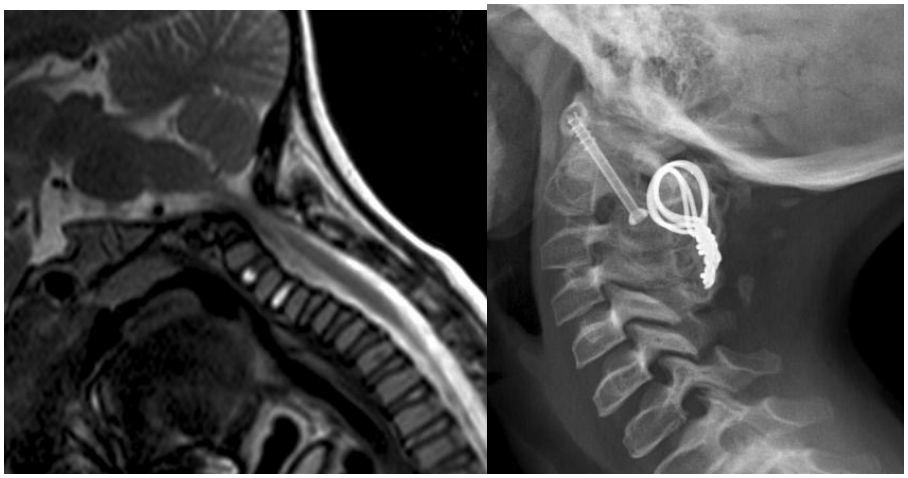
5-yr-old SED boy with os odontoideum

AAD 9mm, SAC 8mm

C1 laminectomy, C0-C2 with Codman cables.

Symptomatic non-union

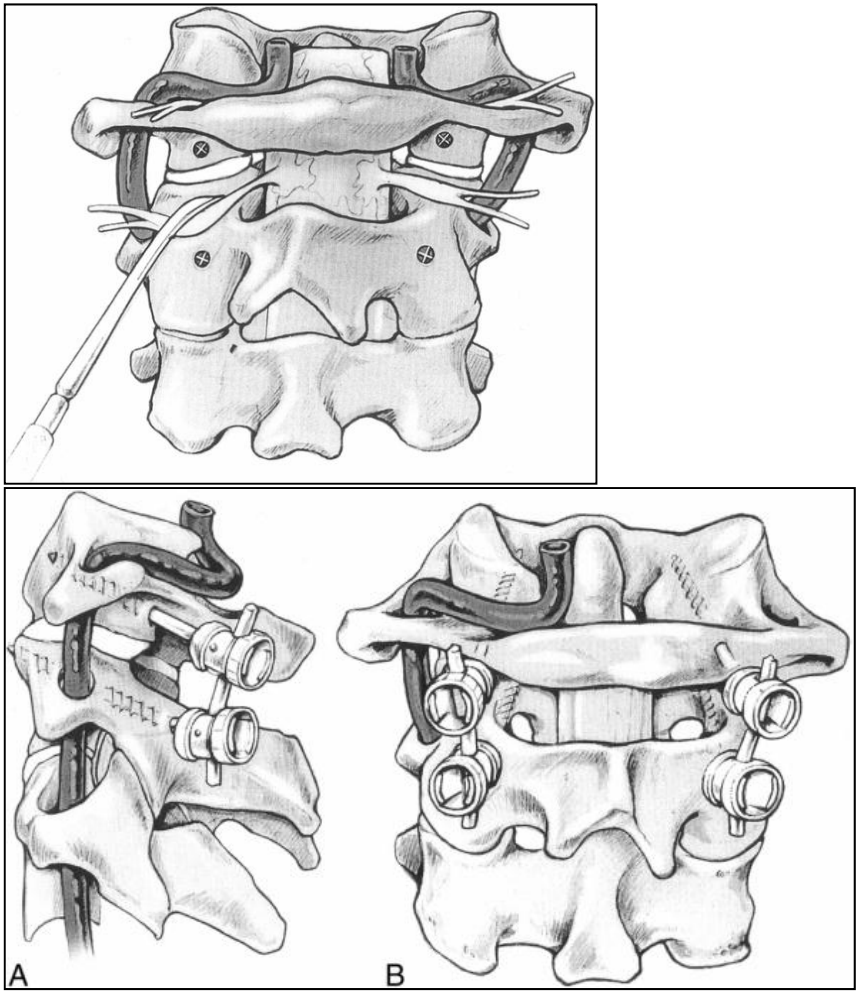
Transarticular screws



- 121 patients (9 with os odontoideum) operated using transarticular screws (Dickman and Sonntag Neurosurgery 1998)
  - 98% fusion rate
- 25% of children present with C2 anomaly preventing transarticular screw pathway, risk of vertebral artery injury in children 3% (2/67) (Gluf and Brockmayer. J Neurosurg Spine 2005).
- Requires alignment before screw insertion.
- Biomechanical stability of single transarticular screw & wiring almost the same as 2 TA screws (Naderi et al. Spine 1998)
- 38 Os odontoideum patients, mean age 39 years (Zhang et al. J Neurosurg 2015)
  - No vertebral artery lesions

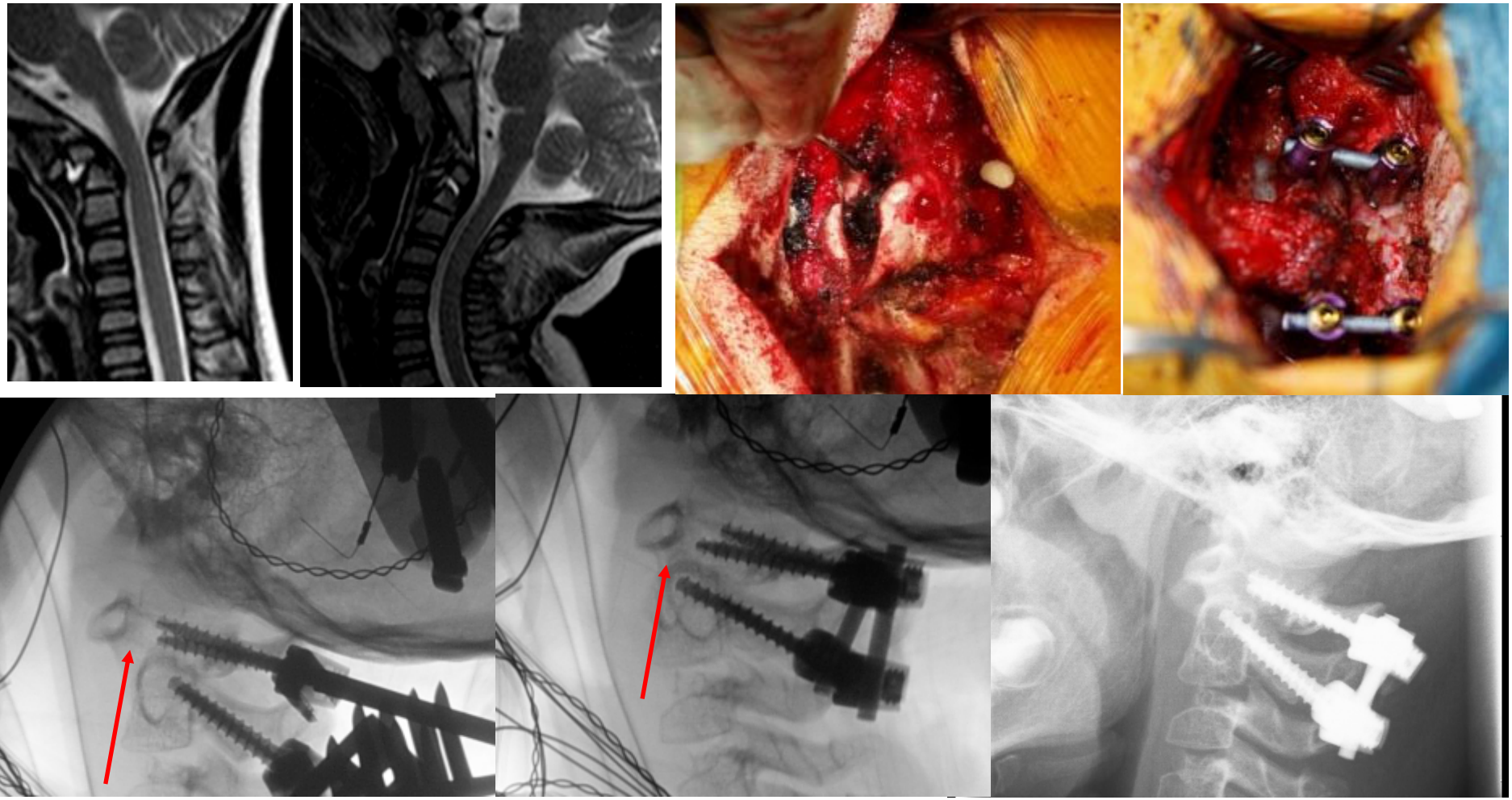
Harms' technique

- C1 mass lateral & C2 pedicle screws (Melcher&Harms, Spine 2001)
- C2 nerve root can be sacrified, typically relatively thick venous plexus over C1 lateral mass
- In adults C1 pedicle screw (if lamina >4 mm) less bleeding, shorter OR time, less C2 irritation as compared with C1 lateral mass (RCT; Yan et al. BMC Musculoskel Dis 2016)
- 202 Os odontoideum patients fused using Harm's technique (Zhao et al. Neurosurgery 2015)
  - Mean age at surgery 38.6 years
  - Whole series of 279 pts included 9 (3.2%) non-unions, 4 infections, 1 csf leak

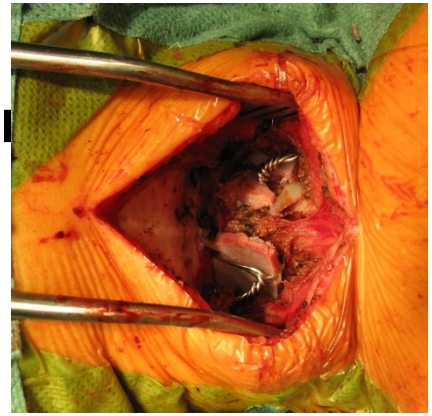


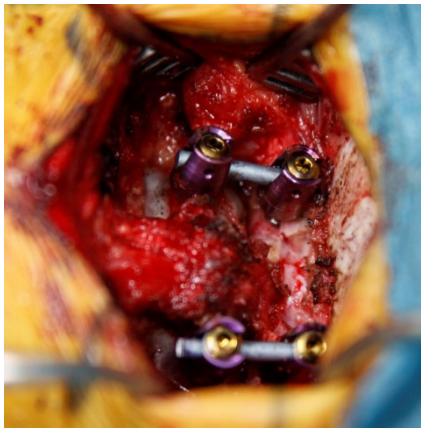
Melcher and Harms, Spine 2001

Harms technique (C1 lateral mass, C2 pedicle)



Complications of wiring vs. rigid

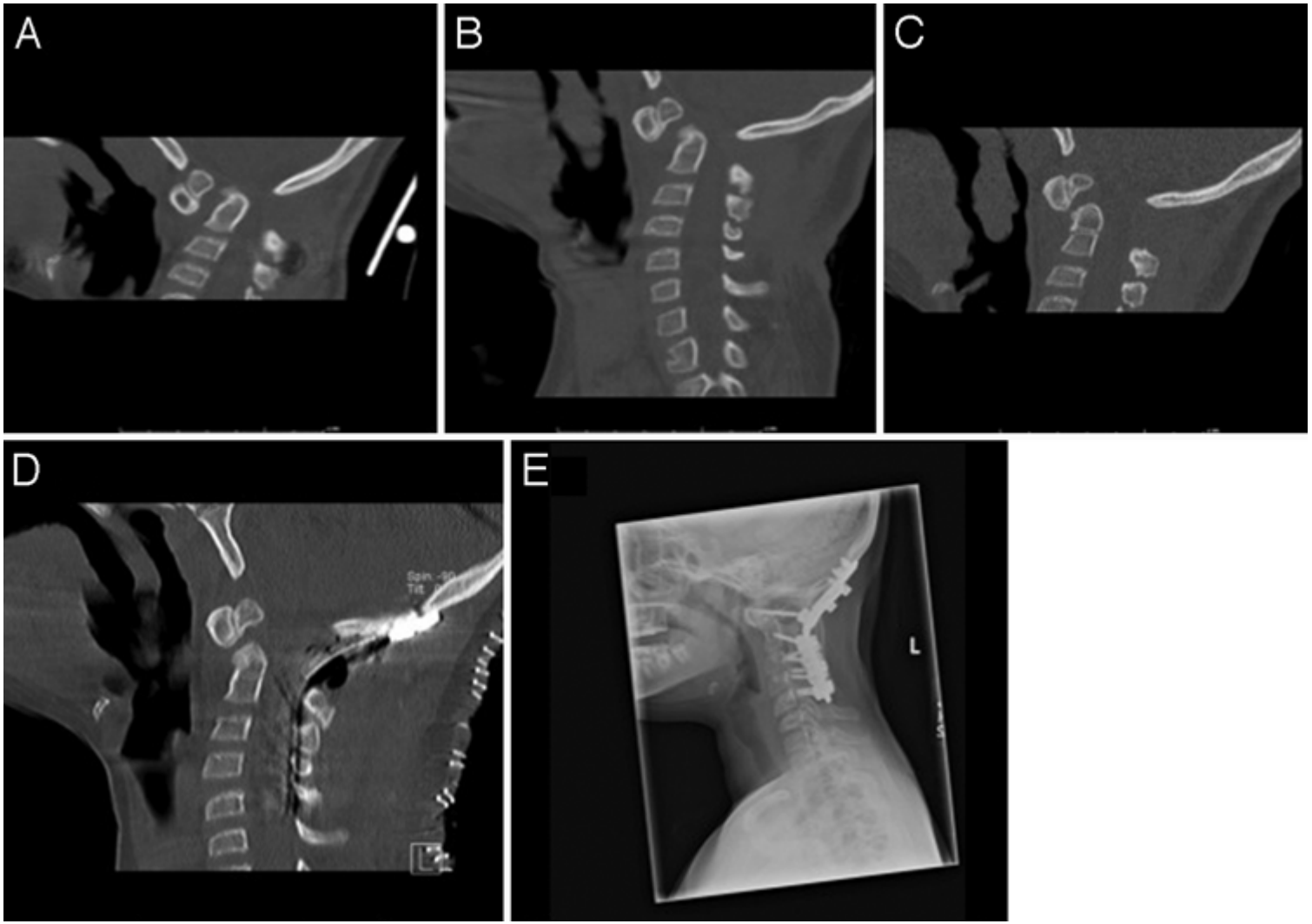




28 children with skeletal dysplasia and C1/C2 instability
14 operated with non-rigid, 14 with rigid fixation (Helenius et al. JBJS 2015)

	Non-rigid	Rigid	
Any complication	9 (7)	1 (3)	p=0.031
Non-union	6	0	p=0.0057
CSF leak	1	1	
V. arterial bleed	0	2	
Autograft dislocated	1	0	

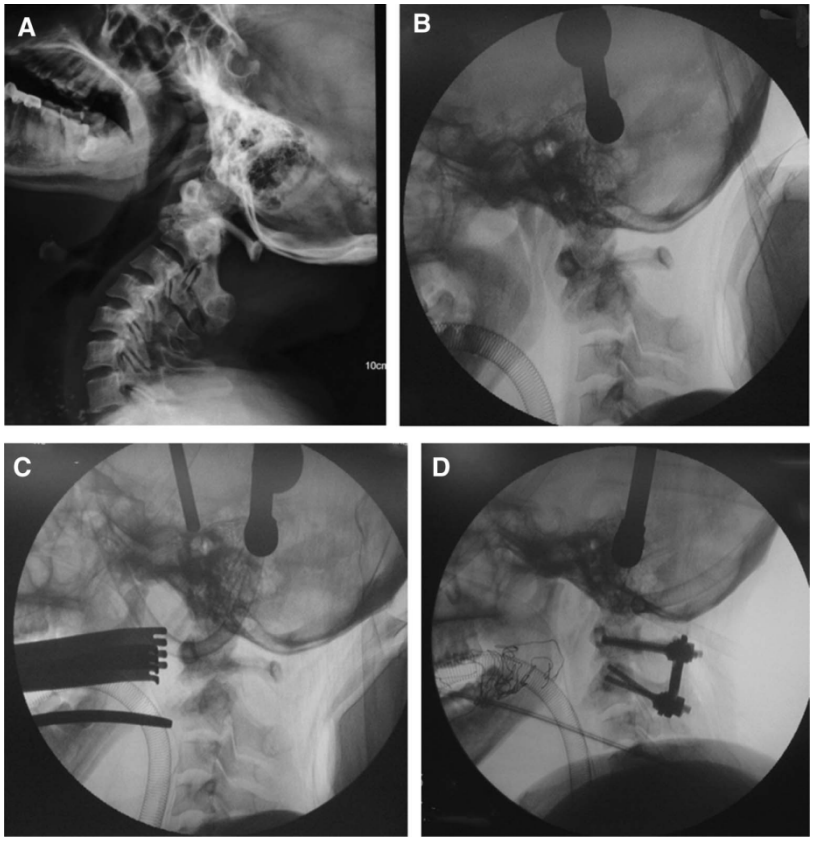
Preoperative halo traction for fixed ventra compression



Abd-El-Barr et al. J Neurosurg Ped 2016

Transoral release or decompression

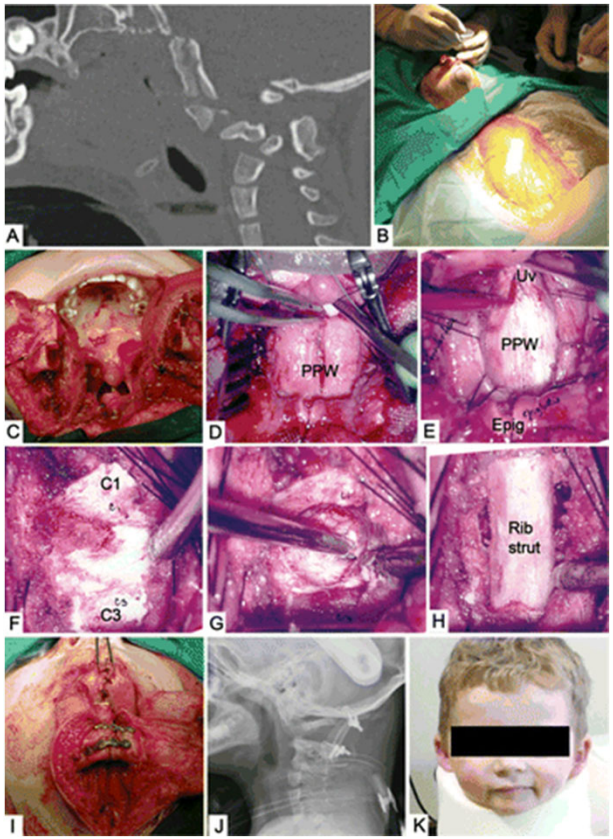
- Transoral procedures decreased (Dlouhy et al. Neurosurg Focus 2015)
  - Traction, rigid internal fixation with distraction
- Transoral approach
  - In small children ability to open mouth may limit use
  - Release or odontoidectomy
  - Transnasal may carry less risks, but requires endoscopic approach (Level of decompression vs. Hard palatinum)
  - Extended transmadibular approach
- Dystopic subtype may increase the risk of irreducible C1/C2 dislocation
- 14% (38/279) required transoral release (Zhao et al. Neurosurgery 2015)
  - Age of the patients or type of Os Odontoideum needing anterior approach not reported



Zhao et al. Neurosurgery 2015

Outcomes of transoral decompression

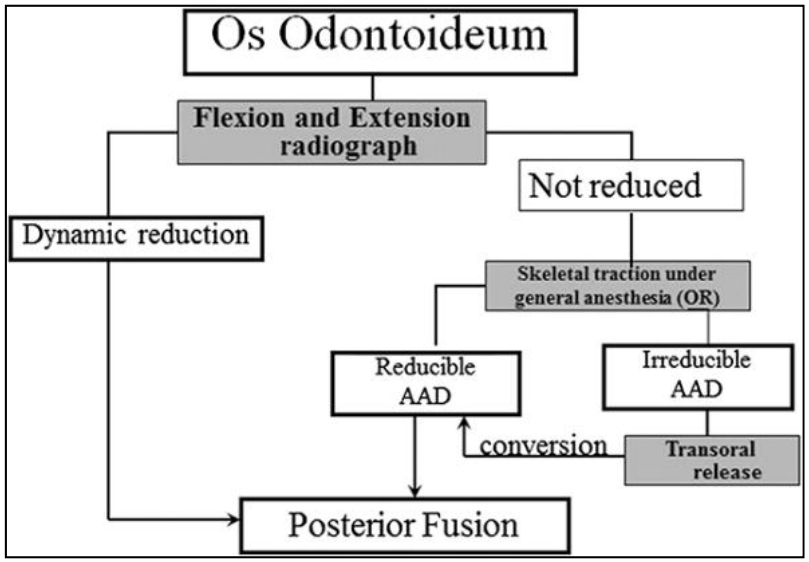
- 280 children undergoing transoral approach (Menezes et al. Childs Nerv System 2008)
  - CSF leak or meningitis 0%
  - Wound dehiscence 0.7%
  - Velopharyngeal insufficiency 1.8%
- 411 transoral approaches (Choi and Crockhard, Neurosurgery 2013)
  - Wound infection 1.1%
  - Dysphagia 3.3%
  - CSF leak 1.1%
  - Velopharyngeal insufficiency 14% (associated with soft palate split)
- Endoscopic endonasal approach (Ponce-Gomez et al. Neurosurgery Focus 2014)
  - Less need for soft palate division, tongue and oropharyngeal swelling
  - May lead to earlier extubation and decreased risks of velopharyngeal insufficiency



Dlouhy et al. Neurosurg Focus 2015

Treatment algorithm

- Most children can be treated with posterior approach
- Preoperative traction with or without vest (Abd-El-Barr et al. J Neurosurg Ped 2016)
- Intraoperative traction trial
- Dystopic subtype may increase the risk of irreducible C1/C2 dislocation
- 14% (38/279) required transoral release (Zhao et al. Neurosurgery 2015)
  - Mostly adult patients?



Zhao et al. Neurosurgery 2015

Author's current practice

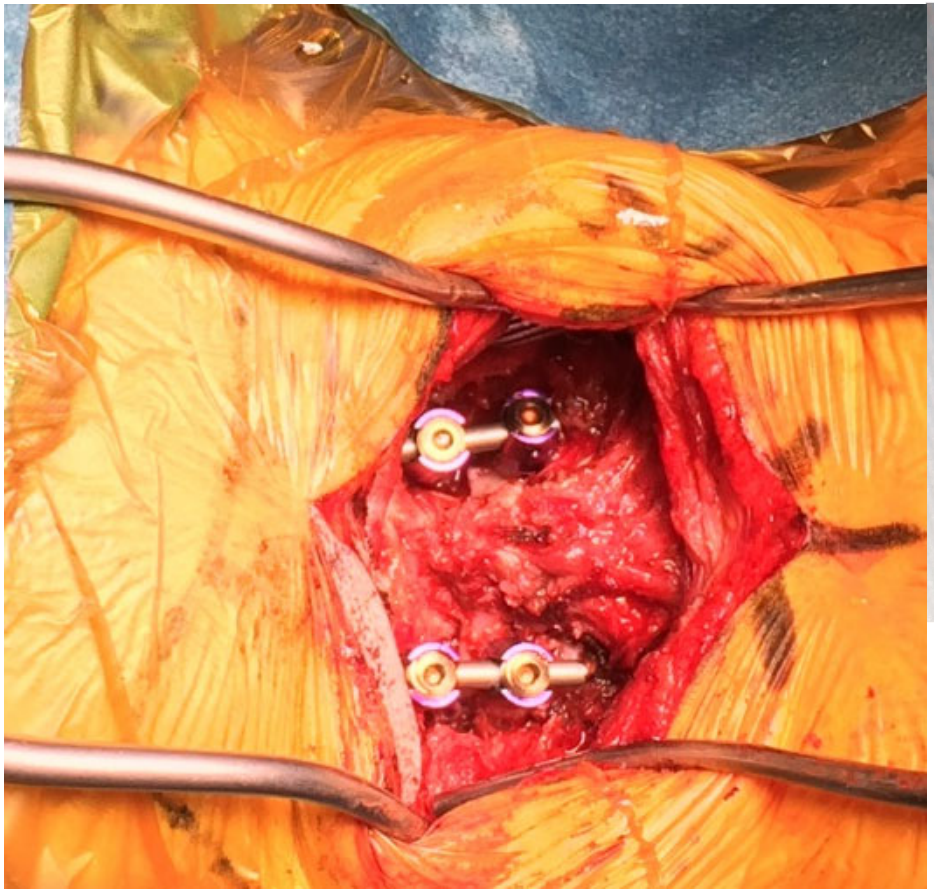
- Preoperative evaluation
  - MR: signal changes, cord morphology, anatomy of arteries (dominant vertebral artery?)
- Intraoperative traction
  - Halo + 1-2 kg traction
- Harms technique
  - C1 navigated + exposure of lateral mass (C2 sacrifice)
  - C2 pars/pedicle free hand
  - Intraoperative O-arm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

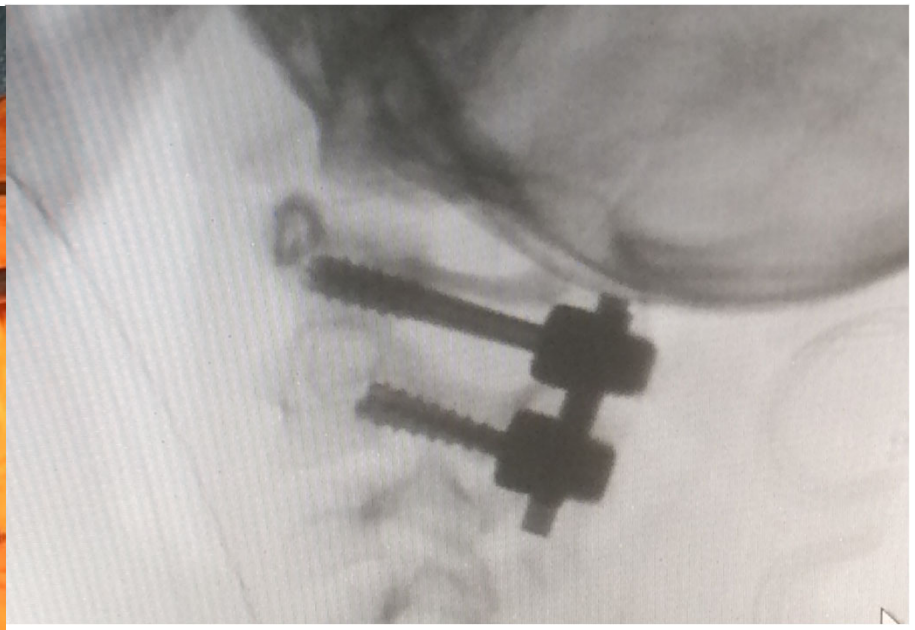
5-yr-old girl, head tilt, neck pain

Intraoperative traction

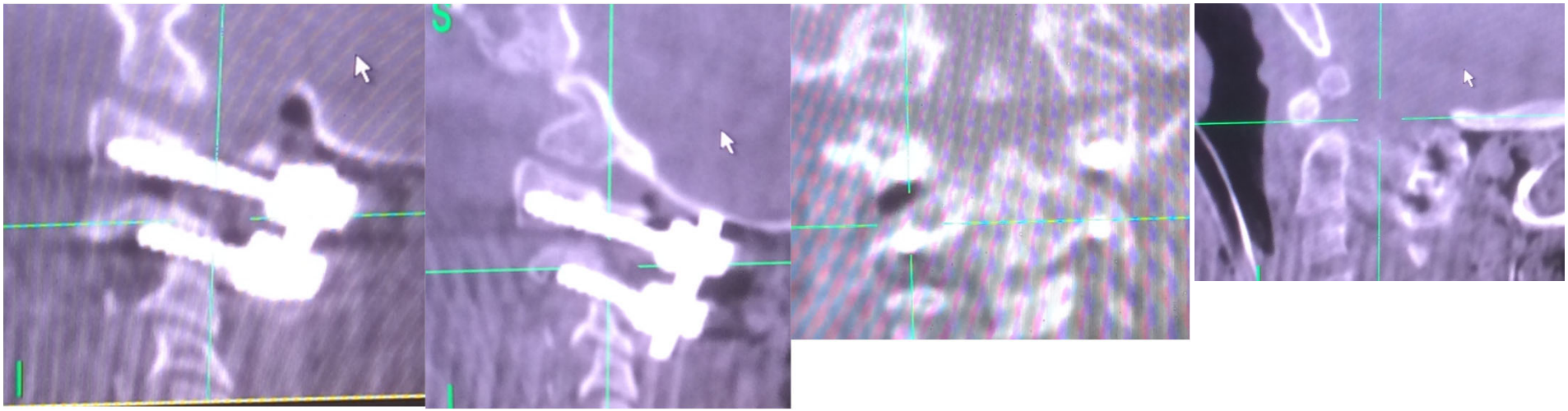
Make sure cervical alignment is acceptable with fluoroscopy

Implant placement





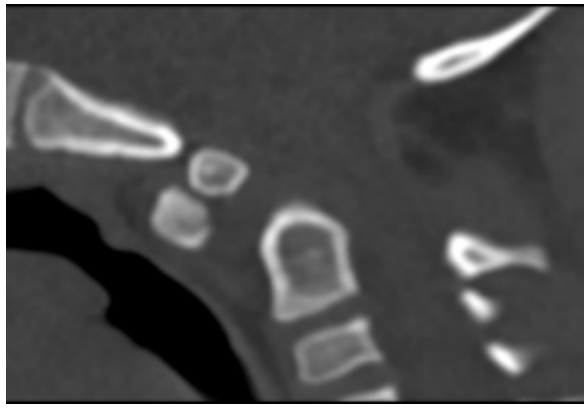
Intraoperative O-arm



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

Conclusions

- Role of conservative treatment unclear
- Indications for spinal fusion
- All posterior for children w/o preoperative / intraoptraction
- Indication and need for anterior release / decompression unclear in children
- Ongoing study on Os Odontoideum by the Pediatric Cervical Spine Study Group
  - Idiopathic vs. non-idiopathic children
  - Conservative treatment
  - Operative treatment



Interested in study?
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