Os Odontoideum in Children: Treatment Outcomes and Neurologic Risk Factors

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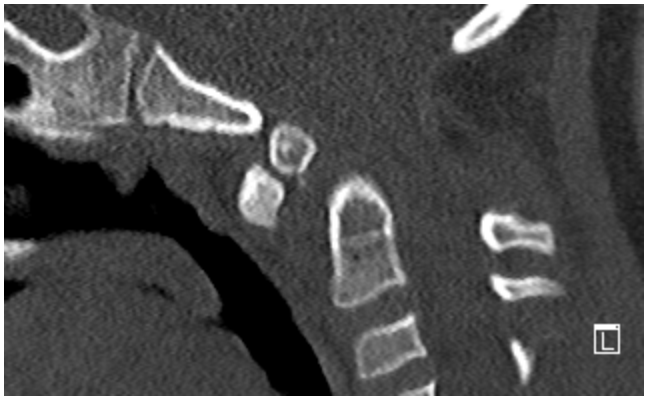
Os Odontoideum

- 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
- 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
- Risk factors for neurologic deficits, indications for conservative and surgical treatment has remained unclear in children



Orthotopic Os odontoideum

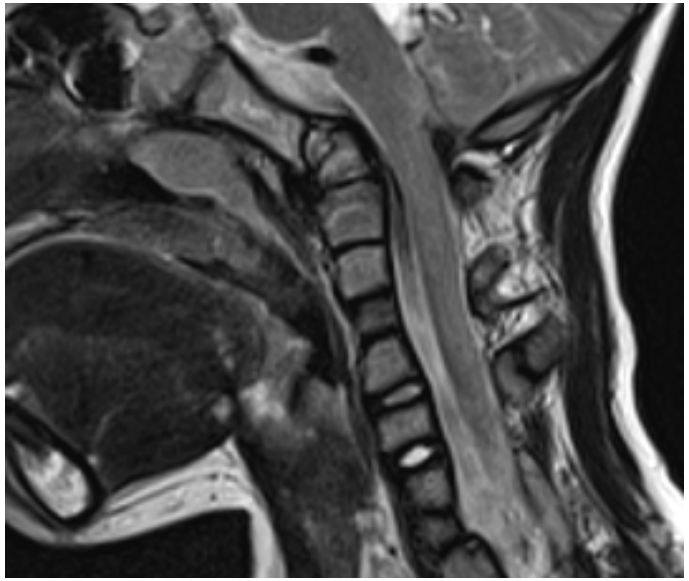
Dystopic Variant

Materials and Methods

- 102 Children with os odontoideum identified from 12 academic Children's Hospitals in Scandinavia and United States between 2000 and 2017 with minimum 2-yr FU
- 31 underwent conservative treatment
- 71 underwent instrumented spinal fusion
  - 50 C1-C2 and 21 occipitocervical arthrodesis
  - 70 posterior, 1 combined approach (transoral odontoidectomy)
  - Mean (range) age at surgery 11.0 (1.5-18) years
- In the surgical cohort 34 idiopathic, 37 with associated syndrome
  - Down syndrome 17
  - Other chromosomal 6

Conservative Treatment

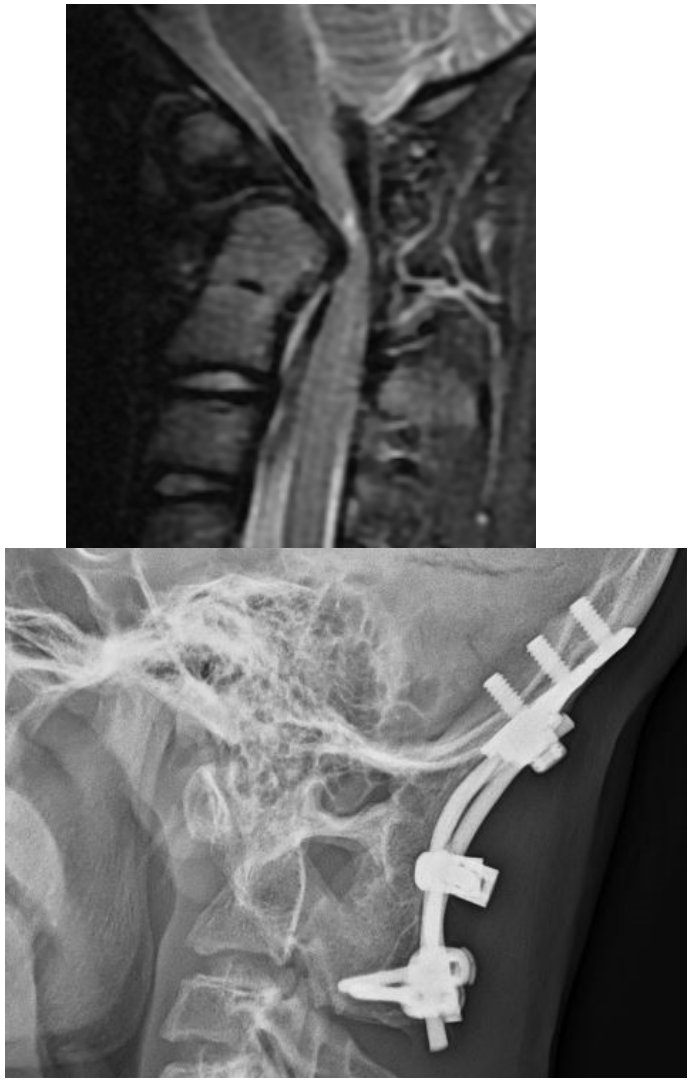
- Indications for conservative treatment:
  - 1) Atlantoaxial distance (AAD) ≤ 5 mm, and/or
  - 2) No neurologic deficit.
- Mean age 7.0 years (range 0.2-16.6 yrs)
- Observation only (n=21)
- Activity restriction (n=9)
- Short immobilization with a rigid collar (n=1) or halo body jacket (n=1) after transient neurologic deficit
- One patient treated conservatively as they refused spinal fusion
- Minimum 1 series of flexion-extension radiographs during follow-up (Mean 3.8 years, range, 2.0 to 11.9 years)



5-yr-old boy with os odontoideum

Surgical Methods

- Indications for arthrodesis:
  - 1) Atlantoaxial distance (AAD) > 5 mm, and/or
  - 2) Limited space available for the cord (SAC) ≤13 mm on maximal flexion or extension radiographs or MRI, irrespective of the age of the child.
- Harms (C1 lateral mass screw & C2 pedicle screw) (n=15)
- Abumi (Occiput plate and C2 pedicle screws) (n=12)
- C1 lateral mass and C2 laminar screws (n=7)
- 19 (27%) children had posterior spinal cord decompression
- One (1%) anterior odontoidectomy
- 32 (45%) Halo, 39 (55%) rigid collar postop (Mean FU 3.5 years, range 2.0-10.6 yrs)

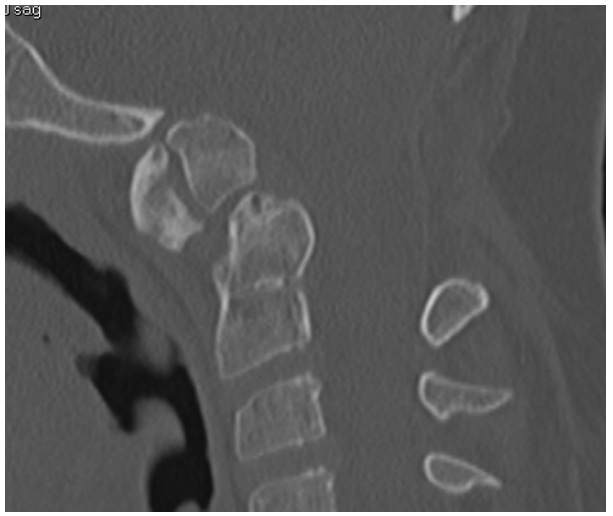


Anterior odontoidectomy

Presenting Symptoms

	Conservative	Surgical
Incidental	24 (77%)	27 (30%)
Neuro deficit	2 (6%)	20 (28%)
Neck pain	3 (10%)	16 (23%)
Trauma	2 (6%)	8 (11%)

P<0.05



12-yr-old boy with ataxic gait

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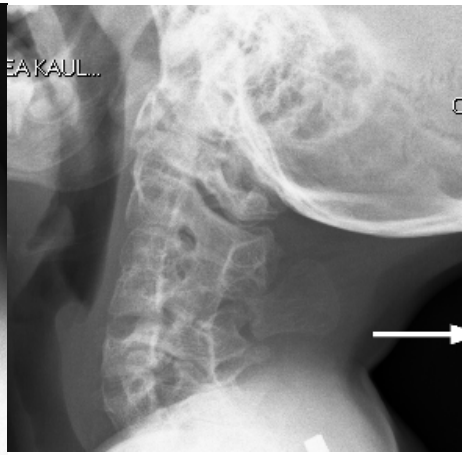
Outcomes of Conservative Treatment

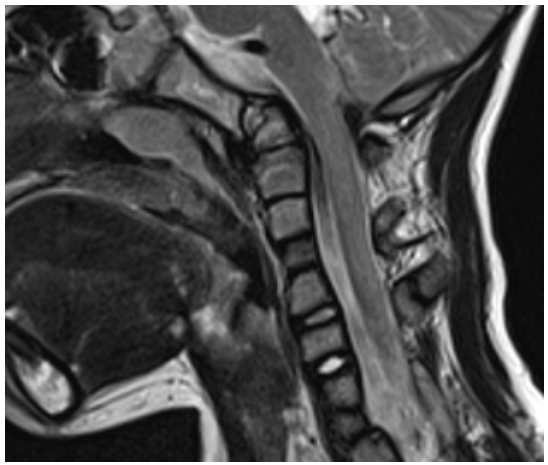
- 27 (87%) children fulfilling criteria for conservative treatment remained asymptomatic during FU
- One 4-yr-old with tetraparesis refused surgery and had persistent deficit
- One 16-yr-old boy with instability (8 mm AAD) declined surgery but remained asymptomatic (6-yr FU)
- 2 children underwent cervical spine arthrodesis after initial conservative treatment
  - One developed instability
  - One developed high signal change in the spinal cord

Asymptomatic & No instability during FU

Dg: 5-yr

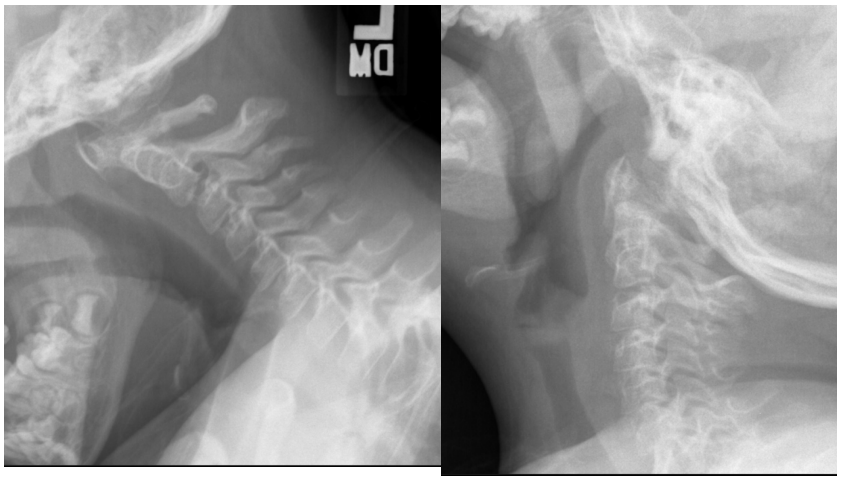


Flexion-extension radiographs at 5-yr FU

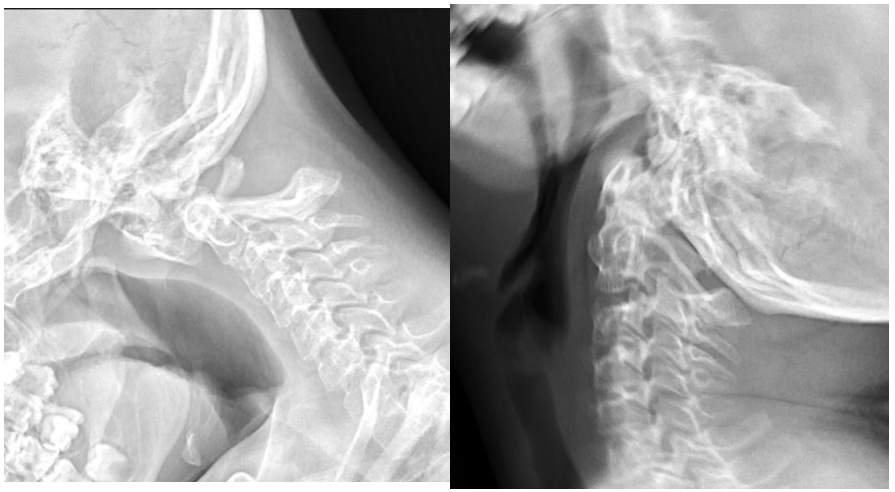


10-yr FU

Down Child Developed Instability



Flexion-extension radiographs at 4-yr
AAD < 4 mm



Flex-ext radiographs at 9-yr
AAD 8 mm, SAC 11 mm

Outcomes of Spinal Fusion

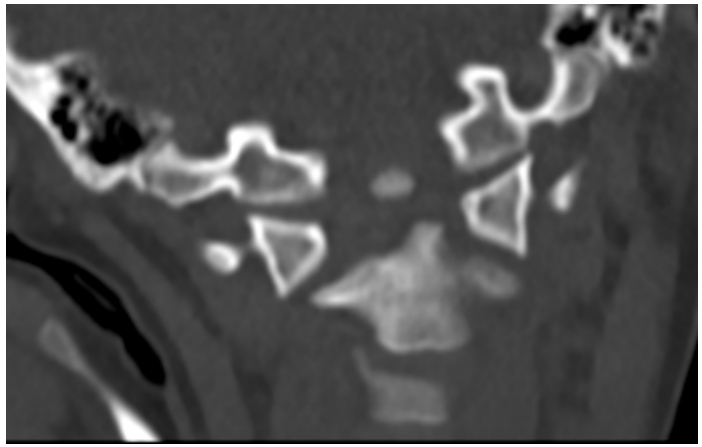
- 68 (96%) had spinal fusion at FFU
- Neck pain
  - 34 (48%) preoperatively
  - 10 (14%) at FFU
- Neurologic improvement (JOA, $p<0.05$)
  - Upper extremity 3.4→3.6
  - Lower extremity 3.3→3.7
- 21 (30%) had a complication
  - 12 non-union
  - 4 new neurological deficit
  - 2 CSF leak
  - 2 symptomatic instrumentation requiring removal
  - 1 vertebral artery lesion

Risk of Re-operation

- 9 (13%) at least one re-operation
- 12 re-operations in total
  - 9 for non-union
  - 2 symptomatic instrumentation requiring removal
  - 1 CSF leak

Risk Factors for Complications

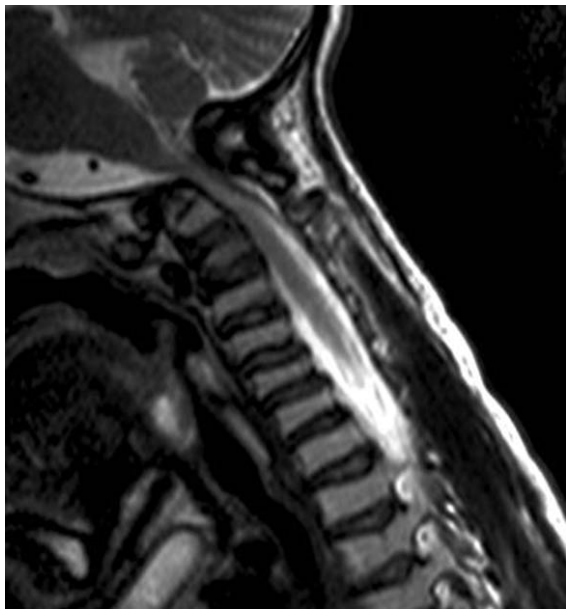
- Age <10 years at the time of surgery reduced complications (22% vs. 34%, p=0.031)
- Down sdr had higher risk of complications (53% vs. 22%, p=0.016)
- All new neurologic deficits in the dystopic subgroup (p=0.052)
- Rigid instrumentation group had less non-union than non-rigid (9/64 vs. 3/7, p=0.054)



5-yr-old girl with Down sdr

Risk factors for Neuro Deficits

- 30 children out of whole cohort 102 children (non-op & surgical) presented with neurologic deficit
- 28/30 of them showed radiographic risk factors either AAD > 5mm or SAC ≤13 mm vs. 2/30 did not (RR 7.8, 95%CI 2.0-31)



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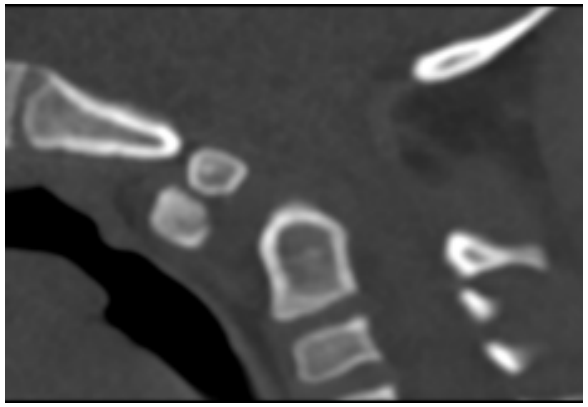
Conclusions

- Atlantoaxial distance > 5mm or Space available for cord ≤ 13mm increase the risk of neuro deficits by 8-fold in children with os odontoideum
- Non-operative treatment provides good outcomes in children with normal neurology and stable atlantoaxial joint.
- Neck pain and neurological deficits improve with arthrodesis w/o decompression
- Risk of complications 30%, re-operation 17%, non-union 17%
- Dystopic variant increases risk of neuro deficits and Down sdr all complications
- Rigid fixation improves fusion rates.



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