Cervical Instability in the Ligamentously Lax Child

Jonathan H Phillips MD

Orlando

ARNOLD PALMER HOSPITAL
For Children
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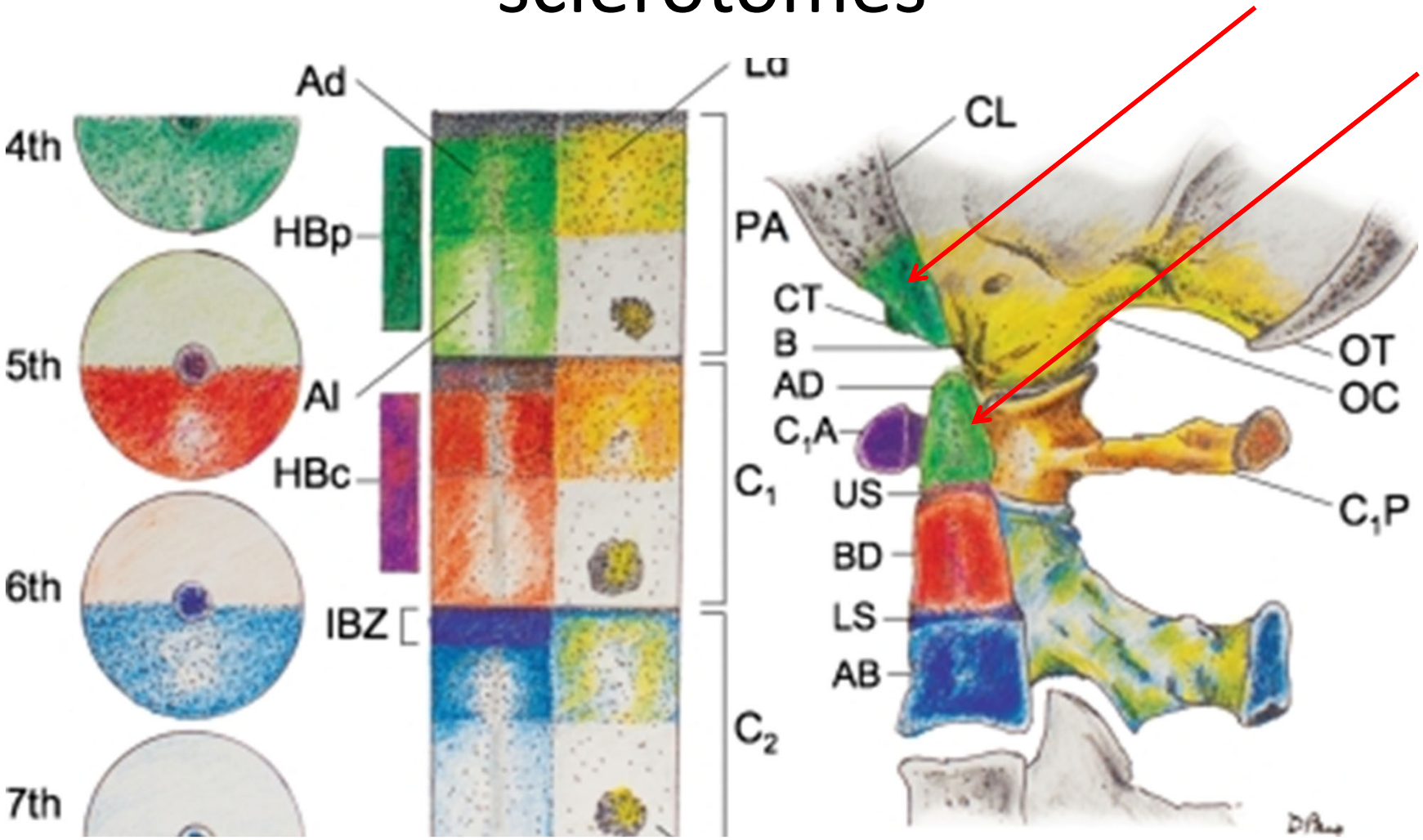
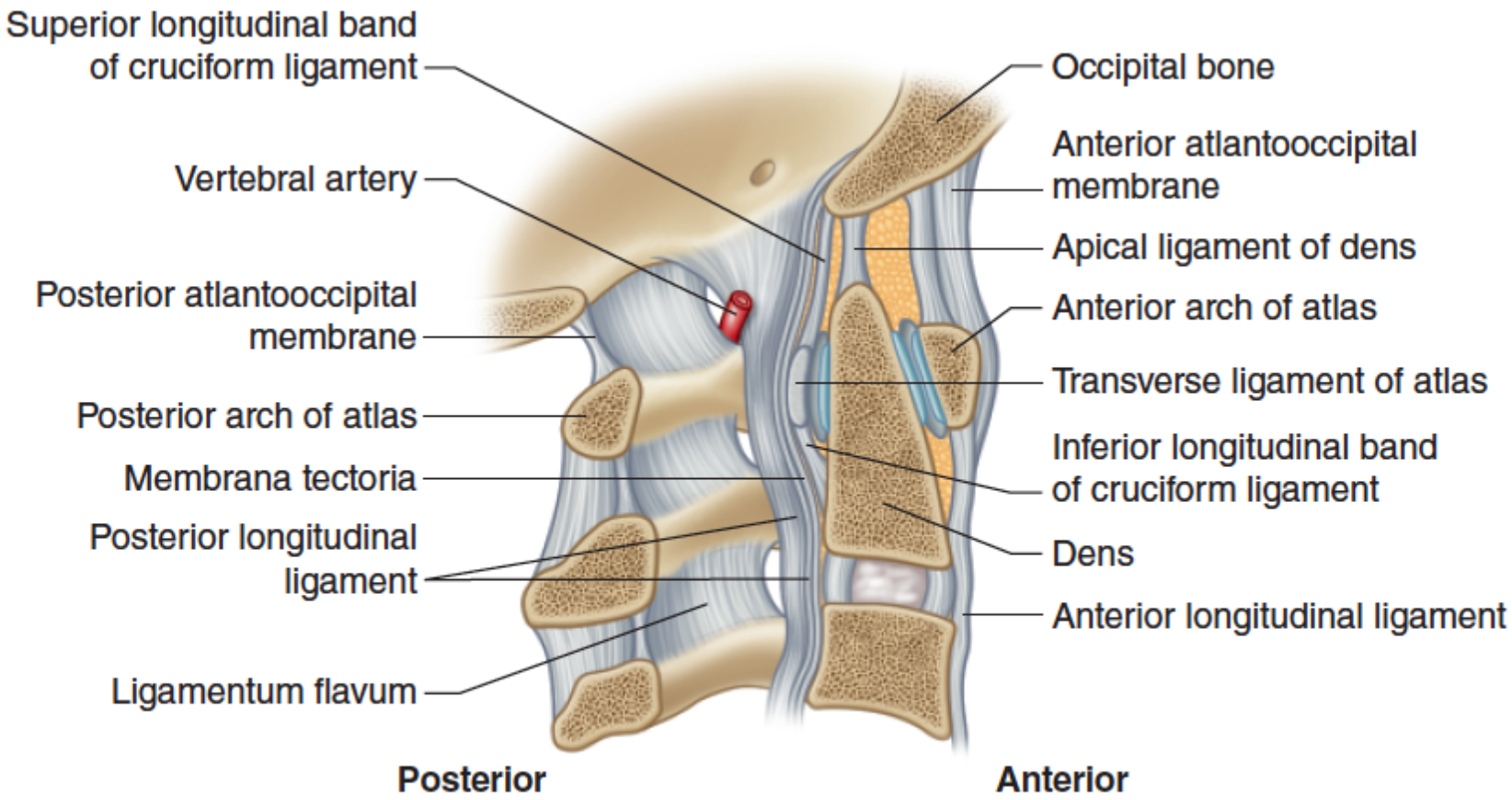
Anatomy and Embryology

- Fundamental to understanding pathology and treatment of the area
- Highly atypical compared with the rest of vertebral anatomy (particularly C1 and C2)
- Variations in the anatomy very common so each individual case has to be studied with advanced imaging

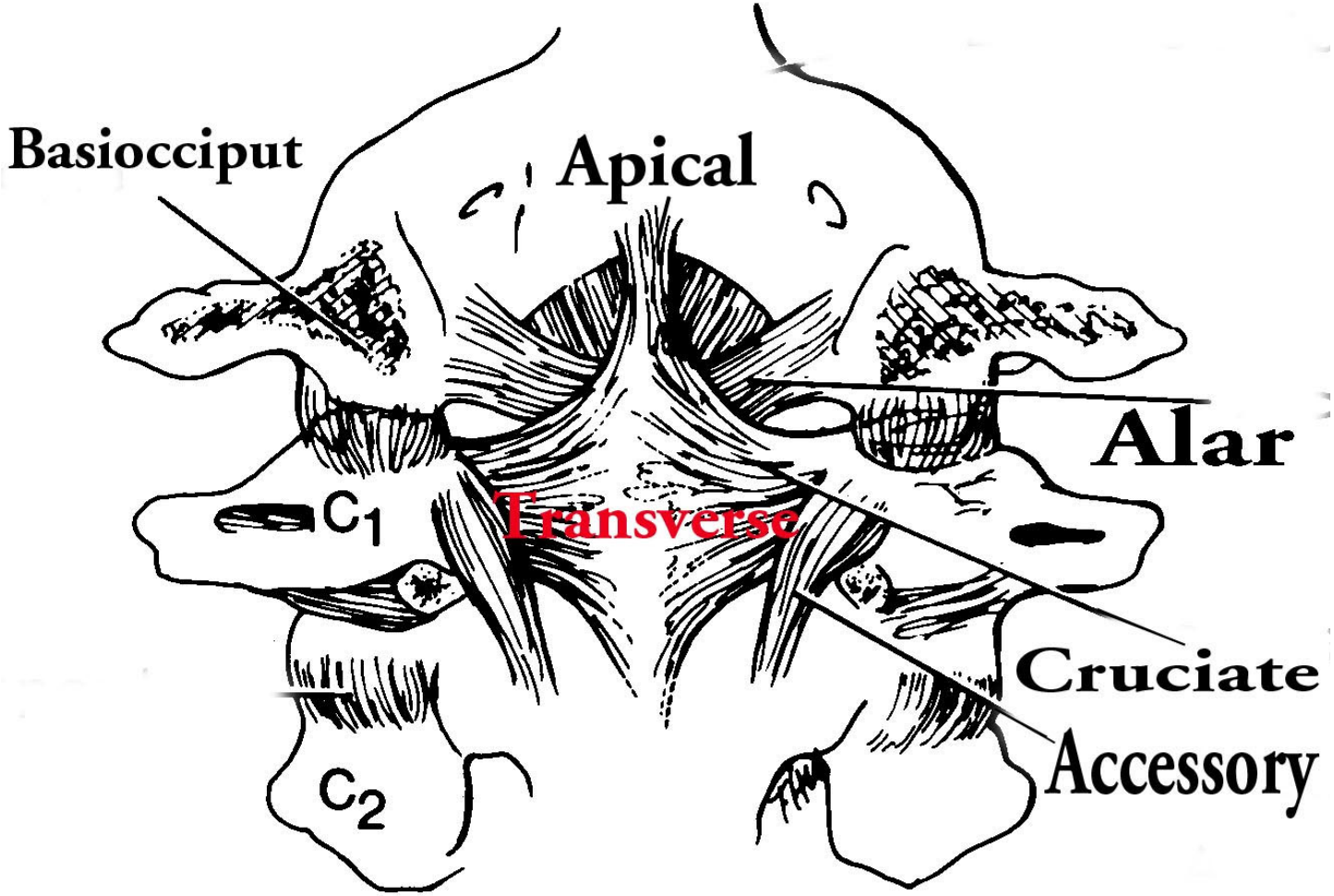
- The upper four sclerotomes form the basiocccciput but also borrow from somite five which is a cervical one,
- Thus the intimate relationship of the atlas to the skull base embryologically.
- Sclerotome five (a cervical one) forms both the posterior arch of the atlas and occipital condyles.

Somites and their resegmented sclerotomes




Superior longitudinal band of cruciform ligament
Vertebral artery
Posterior atlantooccipital membrane
Posterior arch of atlas
Membrana tectoria
Posterior longitudinal ligament
Ligamentum flavum
Occipital bone
Anterior atlantooccipital membrane
Apical ligament of dens
Anterior arch of atlas
Transverse ligament of atlas
Inferior longitudinal band of cruciform ligament
Dens
Anterior longitudinal ligament
Posterior
Anterior

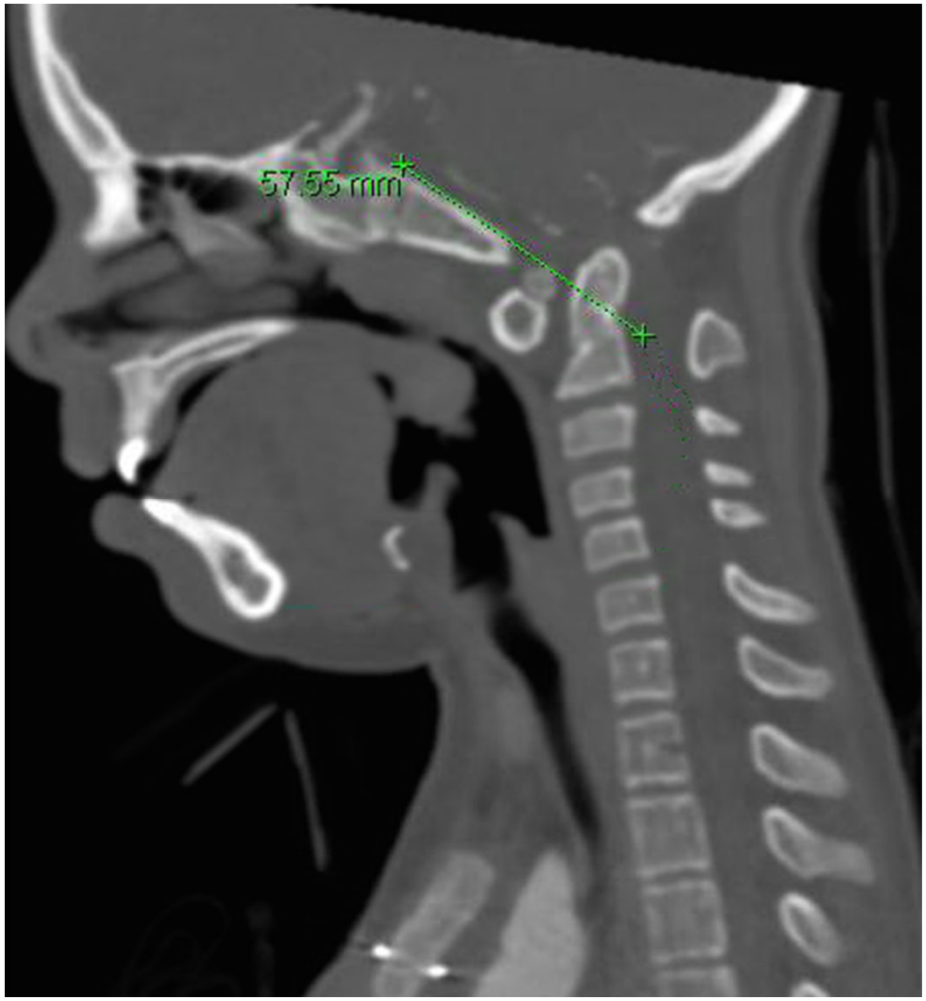
Atlanto Axial Ligaments



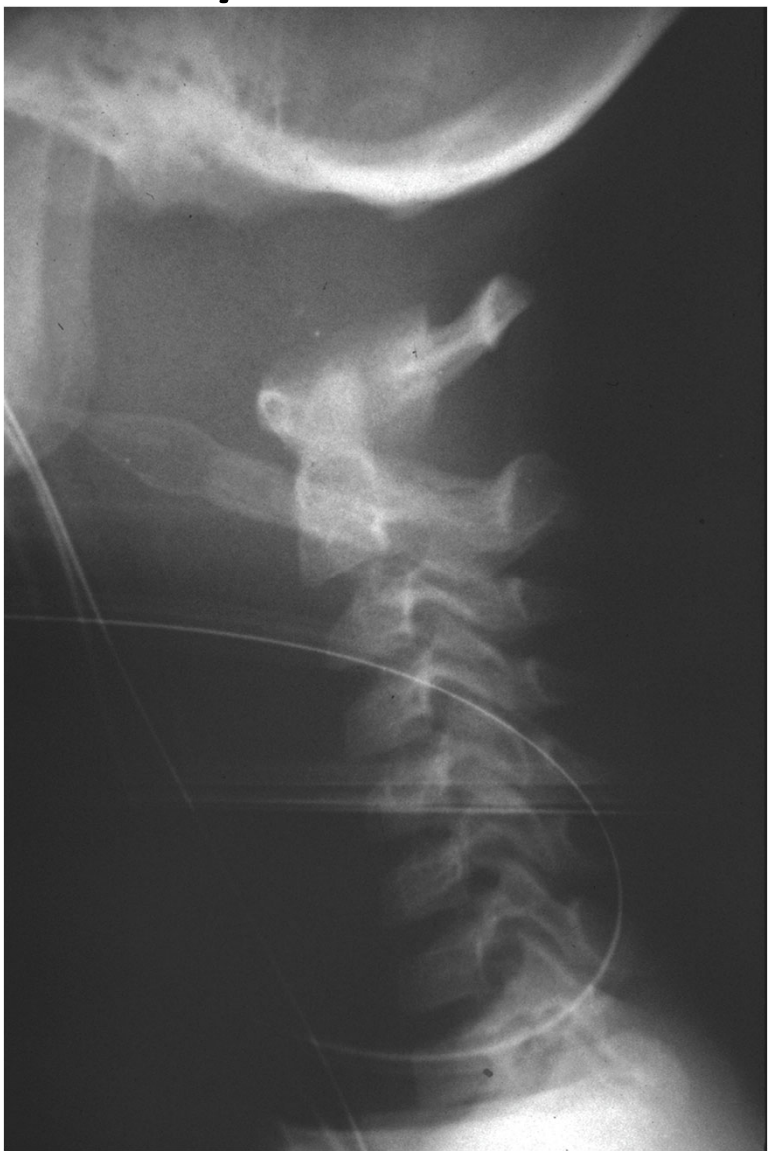
Ligamentous Laxity

- Can be Acute or Chronic
- Can be congenital or acquired
- Can be traumatically generated
- Can be all of the above

Down Syn. (Syndromic/genetic and slow evolving)



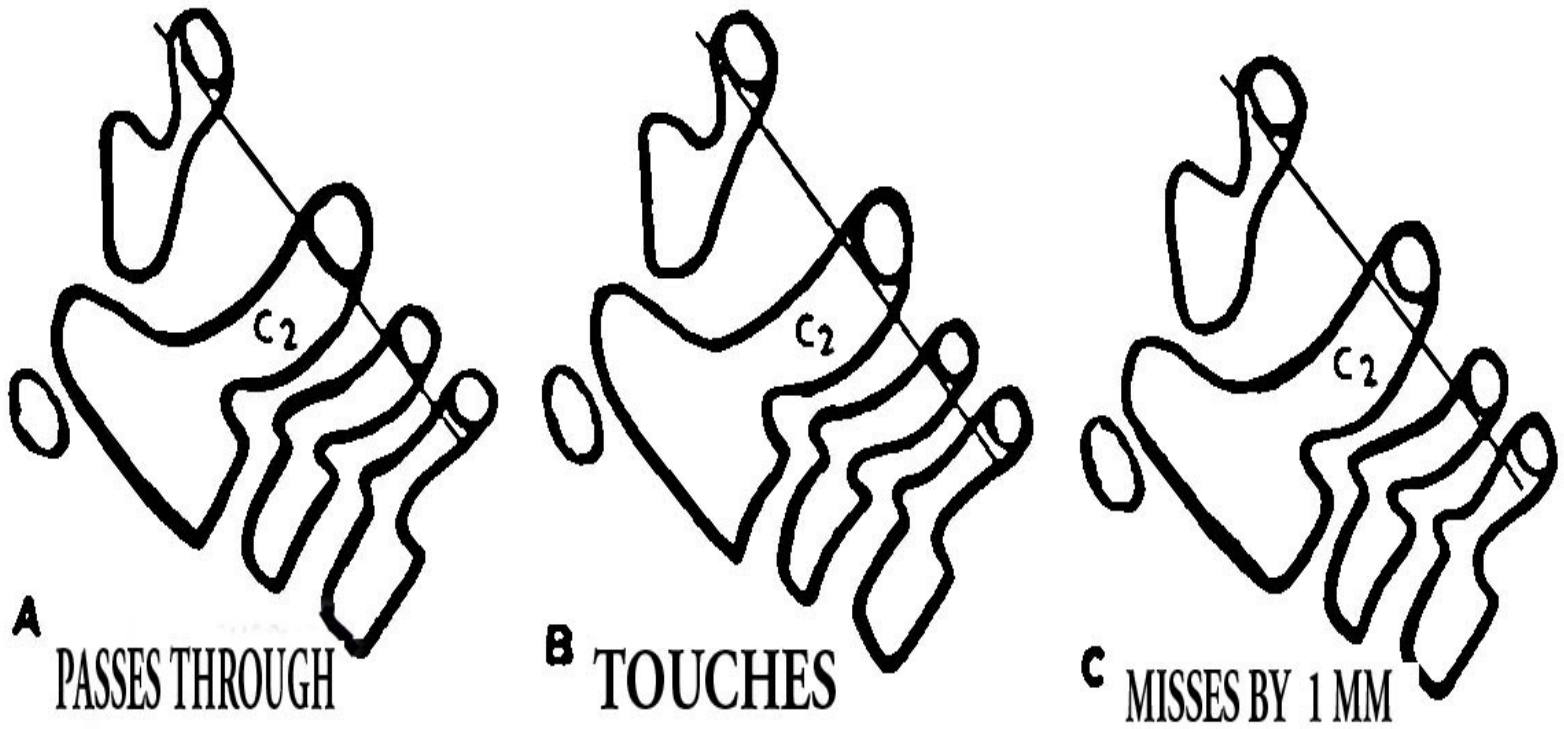
Atlanto Occipital Dissociation
(Traumatic / instantaneous)



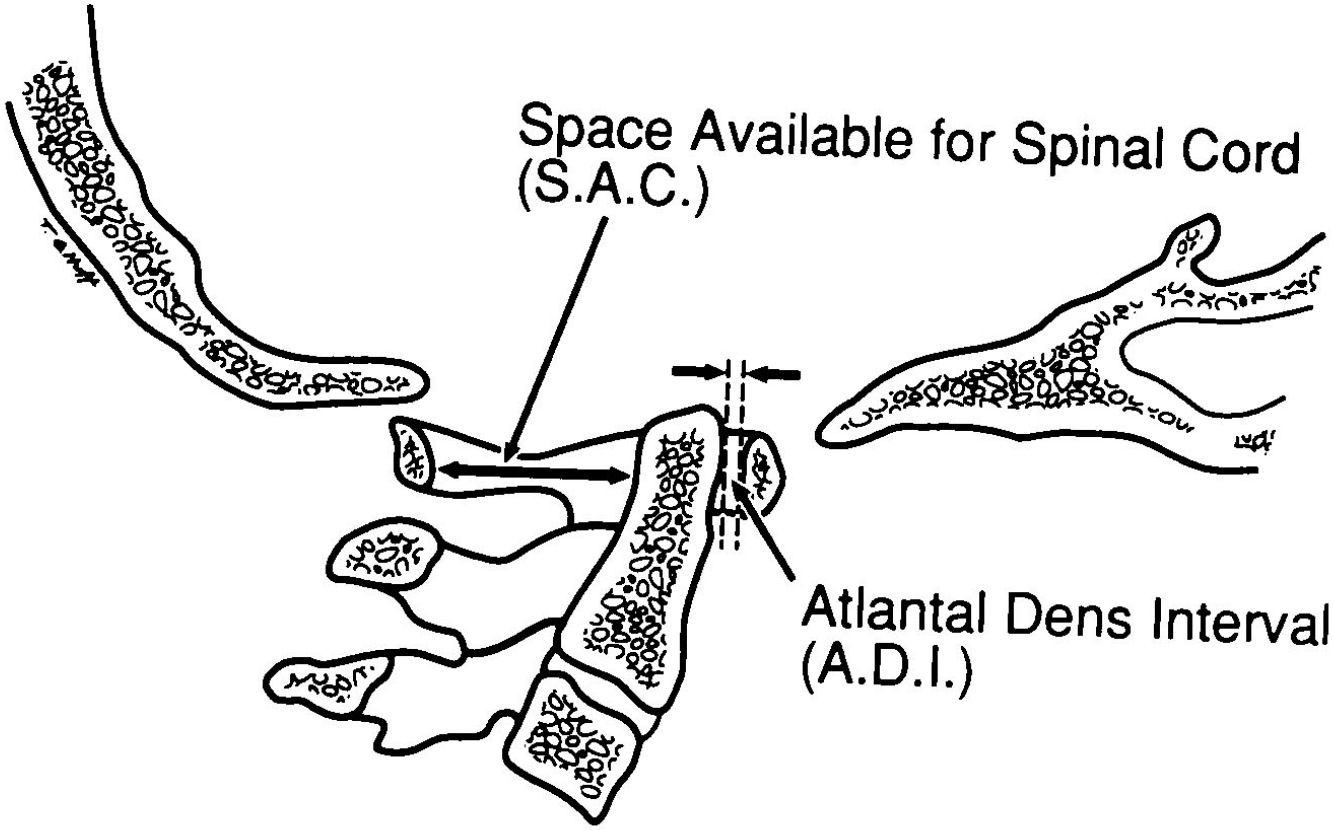
Diagnosis

- Traditionally by neutral and flexion / extension lateral radiographs
- Huge limitations at O C1 and often difficult at C1/2
- Dev. Delay and anatomical variations confuse the picture
- Positioning for radiographs is challenging

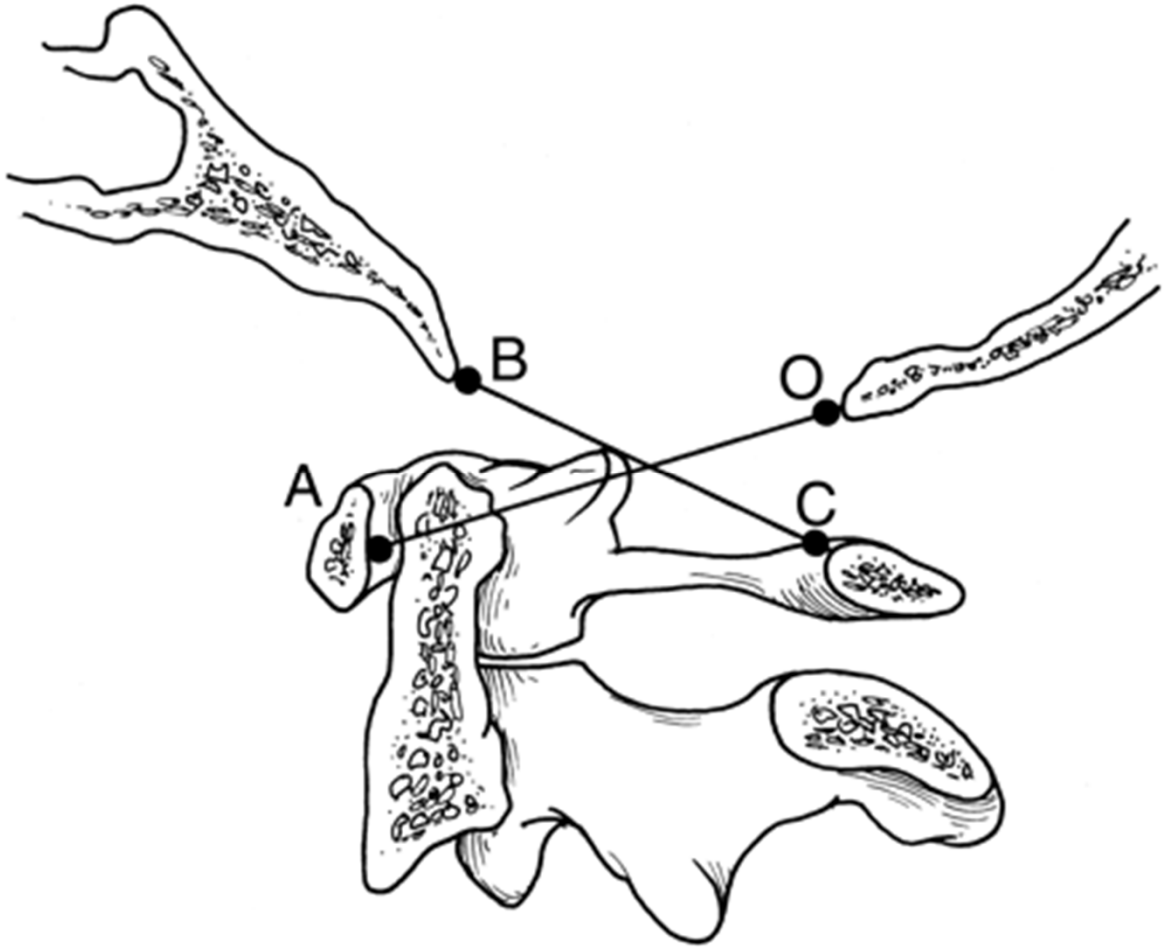
Swischuk's Line



Atlas - Dens interval



Power's Ratio (should be about 1.0)



Et al.

- Wackenheim
- Chamberlain
- McRae
- McGregor
- Welcher…….

Reproducibility in the Measurement of Atlanto-occipital Instability in Children With Down Syndrome

Karol, Lori A. MD[*]; Sheffield, Eugene G. MD[†]; Crawford, Kevin MD[*]; Moody, Marilyn K. MD[†]; Browne, Richard H. PhD[*]

Abstract

Study Design: This study compared the reproducibility of three techniques used to measure translation between the occiput and C1 in children with Down syndrome.

Objectives: Intraobserver and interobserver variability were computed to determine if there is a reliable way to measure occiput-C1 instability.

Summary of Background Data: No studies have been performed comparing measurement techniques in children with Down syndrome.

Methods: Powers ratios, basion-axial intervals, and translational anteroposterior motion as described by Wiesel and Rothman were calculated for 60 pairs of flexion-extension lateral cervical radiographs from children with Down syndrome. Calculations were made on two occasions by four reviewers.

Results: Mean differences between measurements for two observers were 0.14 for the Powers ratio, 1.3 mm for the Wiesel technique, and 1.8 mm for the basion-axial interval. The 95th percentiles of the differences between measurements for two observers were 0.38 for Powers ratios, 3.5 mm for the Wiesel technique, and 5.3 mm for the basion-axial interval.

Conclusions: Measurement of atlanto-occipital translation by any of these methods is not reproducible. Although the technique by Wiesel and Rothman is the easiest to apply, confirmation of instability with magnetic resonance imaging should guide management.

Spine 1996

Congenital Instability

- Down syndrome is the prototypical condition
- Commonest chromosomal disorder
- Multiple areas of joint laxity
- Patello femoral, hip, Atlanto axial

Congenital Instability

- Also cave: EDS, Marfan, Loez Dietz
- Bone Dysplasias
- Larsen

Down Syn. AAP Update 2011

- Children with Down syndrome are at increased risk of atlantoaxial instability. However, not until age 3 years will they have adequate vertebral mineralization and epiphyseal development for accurate radiographic evaluation of the cervical spine. Plain radiographs do not predict well which children are at increased risk of developing spine problems. ***Therefore, routine radiologic evaluation of the cervical spine in asymptomatic children no longer is recommended***.

Down Syndrome

- C1 /2 instability 10 – 30%
- O/C instability less well known
- Probably the two coexist more than we know.
- High complication rate in Down Fusions

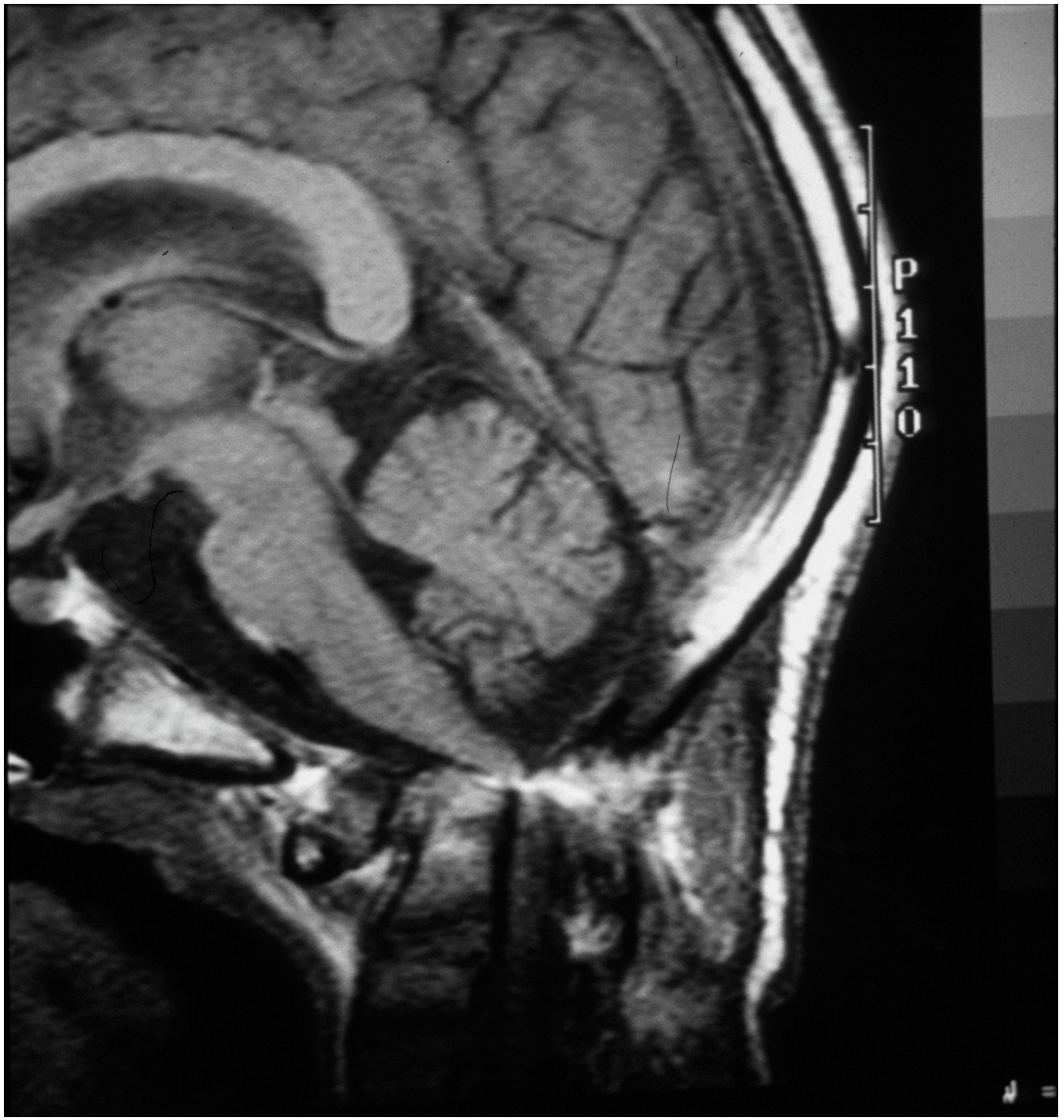
15 year Male, DS

- Quadriparetic but still just about feeding himself
- Frequent Nocturnal apnoea
- Two previous failed C1/2 fusions

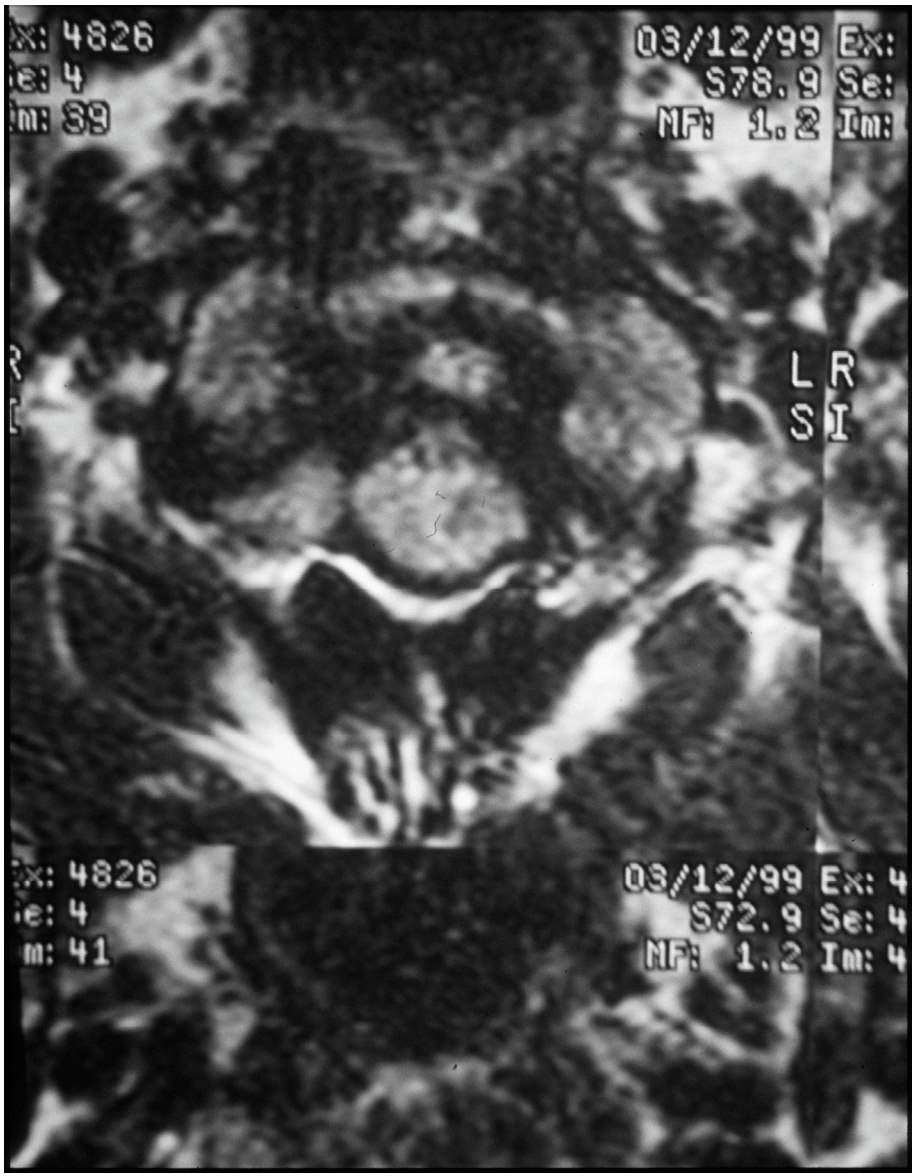
Down Syndrome

Down Syndrome

Down Syndrome



Down Syndrome

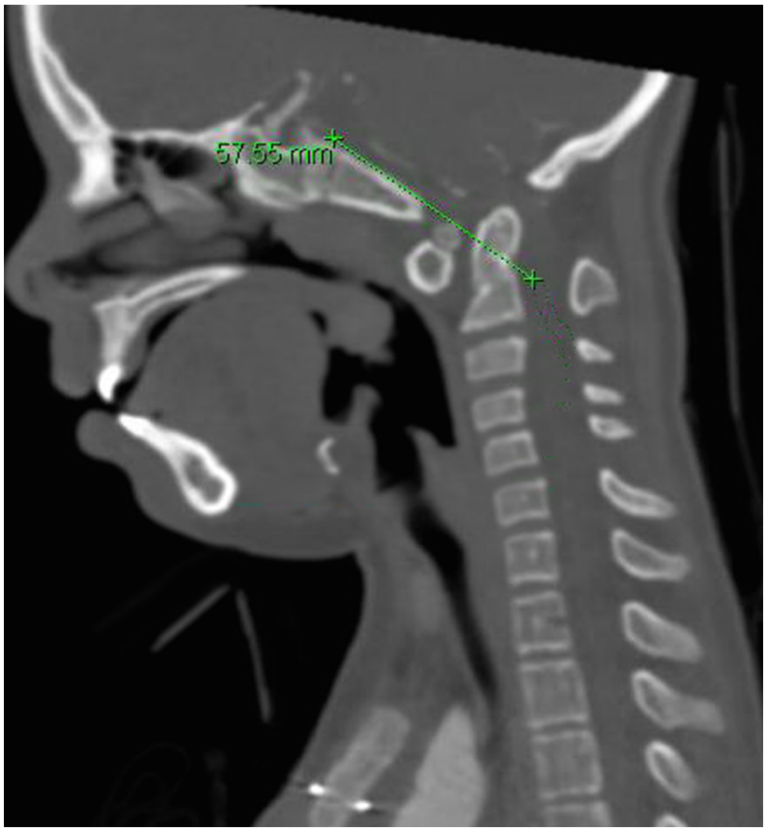


15 yr male DS

- “mother” refused further surgery
- Ethics committee supported JHP and neurosurgeon
- Then committee did 180 and supported “mother”
- Child died about a year later

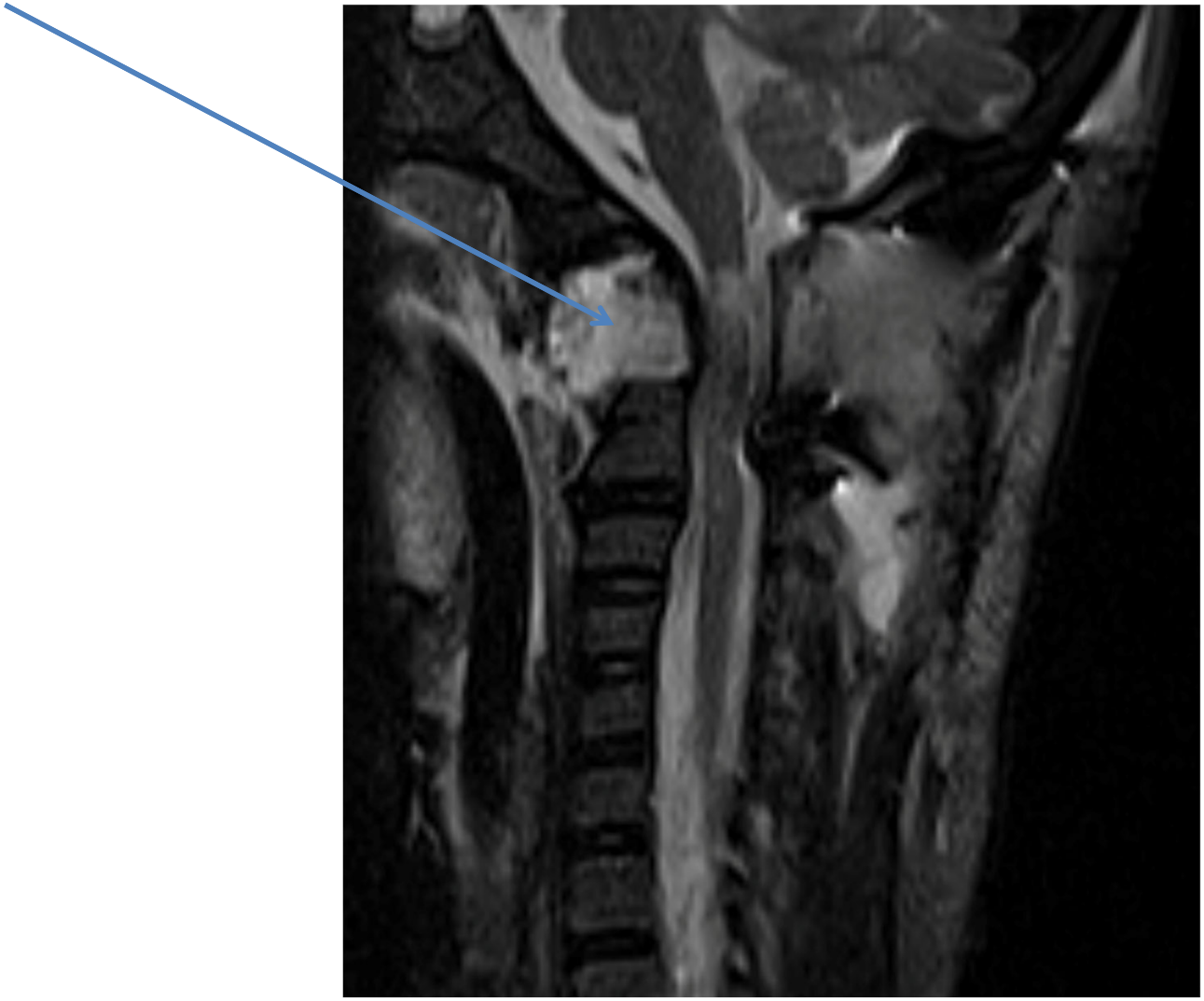
12 year female DS

- Quadriparetic after hip recon surgery
- Posterior occiput to C2 decomp / fusion

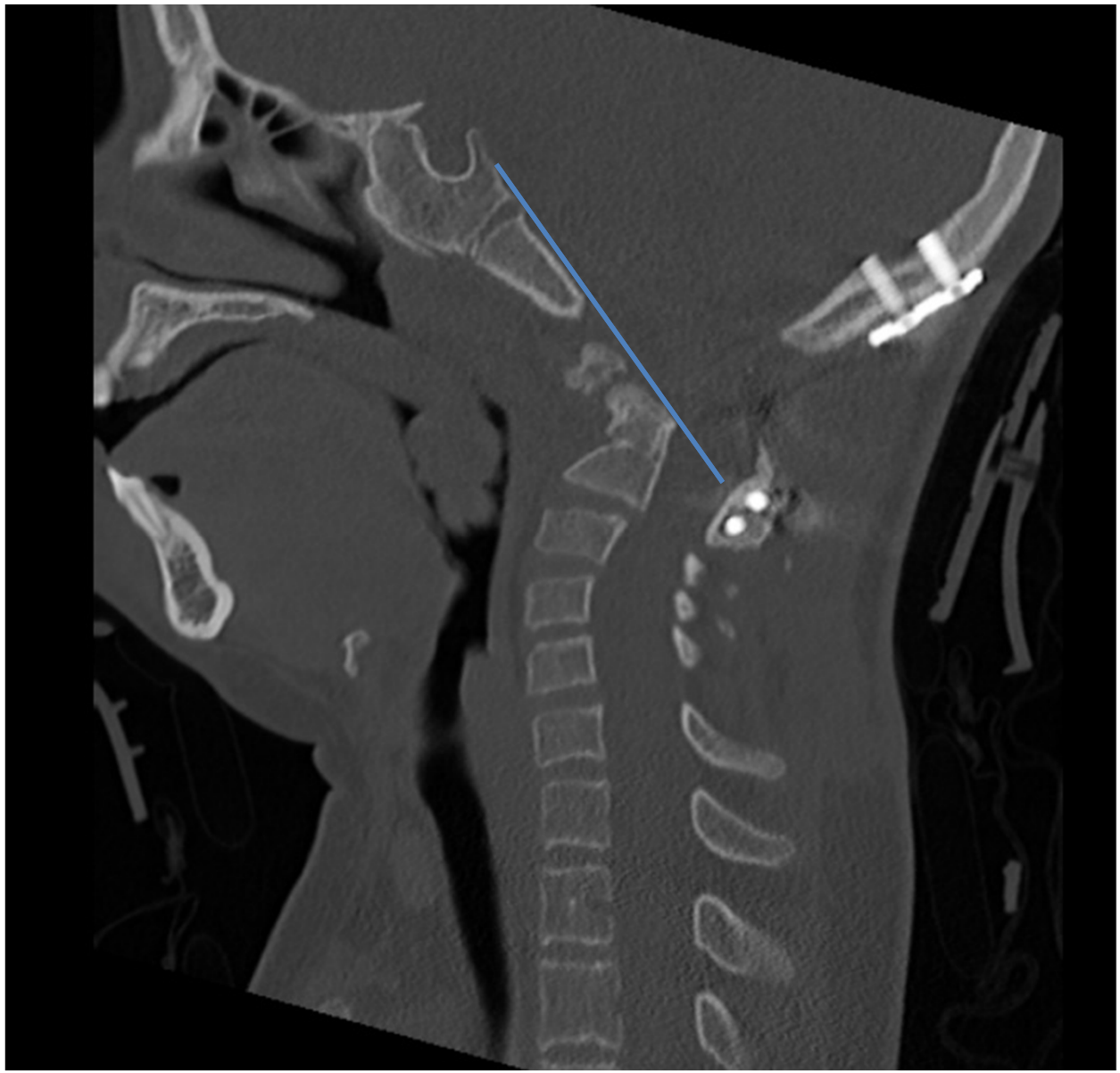


Postop still tight at dens

- Anterior transoral decompression



Final CT, Good clinical recovery



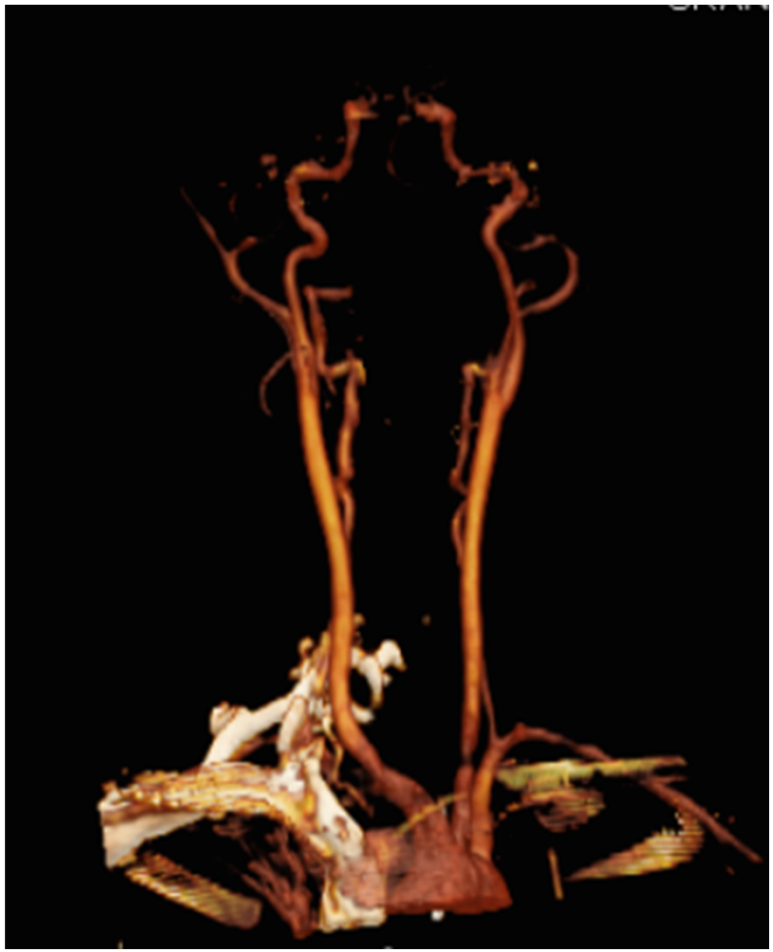
Diagnosis

- Advanced dynamic imaging is often necessary to confirm (or refute) OC instability
- Flex ex CT can be done almost as quickly as radiograph
- Flex ex MRI takes much longer and radiologists may be very wary.

Advanced Imaging

- Take neutral CT /MRI first and proceed with flex ex if further info needed.
- Beware excessive movement if there's any significant stenosis on static neutral studies
- If surgery is anticipated, use some IV dye on the CT study to get a concomitant CT angiogram of the vertebral arteries

Cervical CT Angiogram



Diagnosis

- Careful History and Physical
- Story of altered gait, drooling, snoring head hanging / neck pain
- Loss of fine motor function
- Sphincter disturbance
- Spasticity and or weakness
- Neck posturing

Treatment options

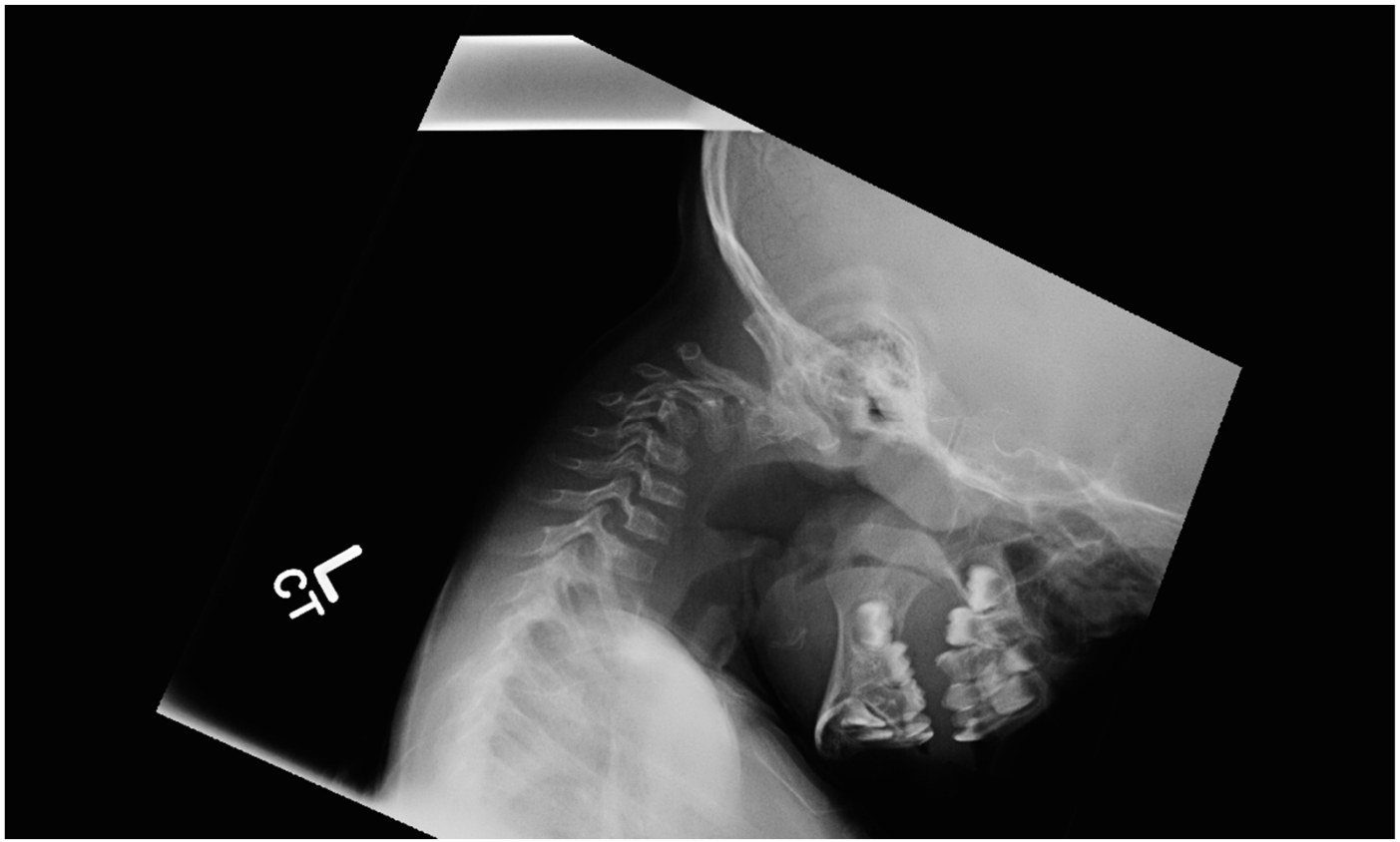
- In absence of symptoms and signs observation may be warranted with strict restriction of impact activities
- Neurological signs and or gross instability requires surgery
- Isolated C1/2 instability can be fused at this level only
- Don’t hesitate to extend fusion to the skull

Treatment Options

- Modern modular instruments are a great help
- Must know old school techniques as well
- Must be handy with a halo

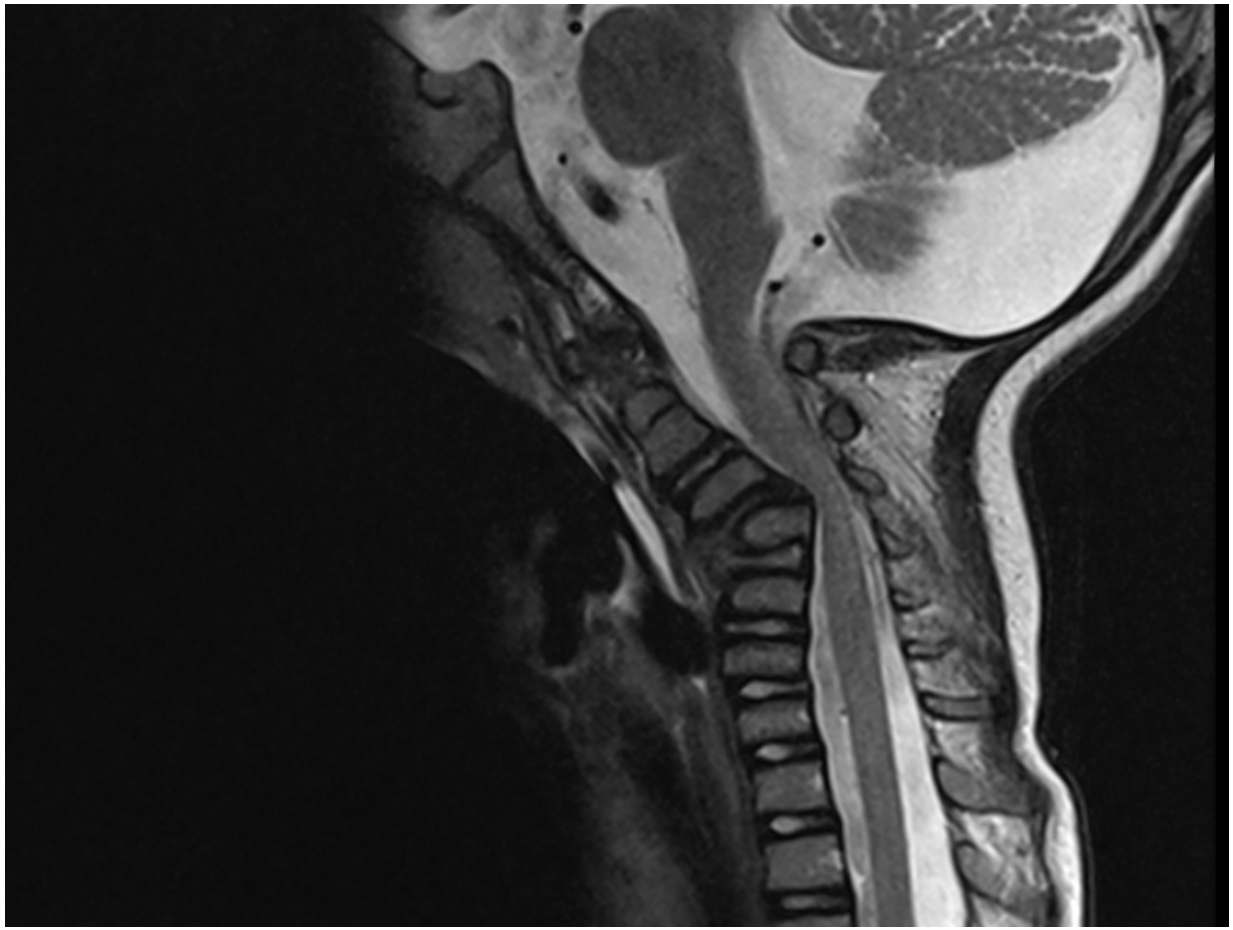
Other ligamentous conditions?

- Its not just Down syndrome! Not just O/C1/C2
- 3 yr old with OI



3 yr old OI

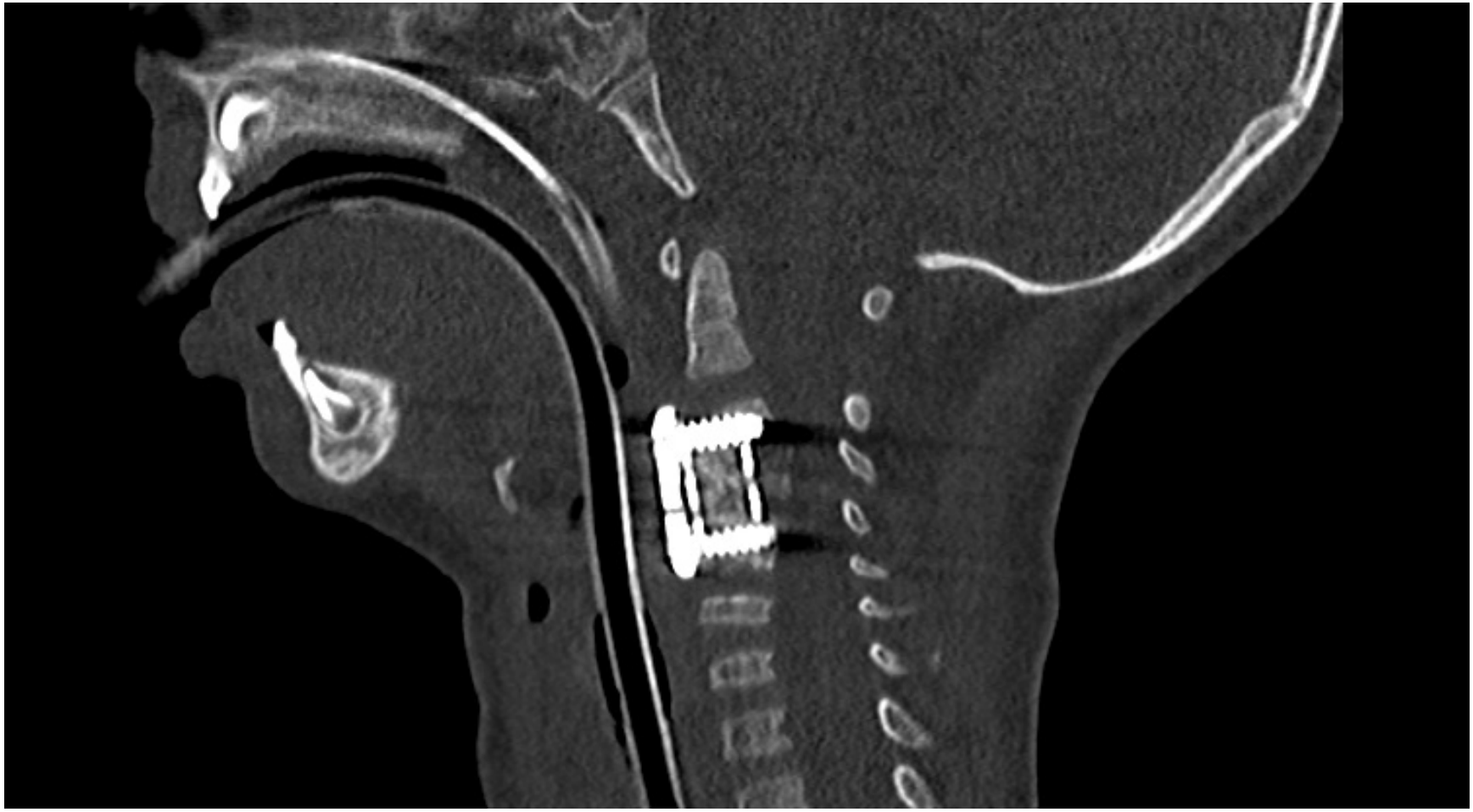
- Snoring, drooling, sleep apnoea, dysphagia
- Falling episodes



3 yr old OI

- Halo placement (ten pins at 2 pounds)

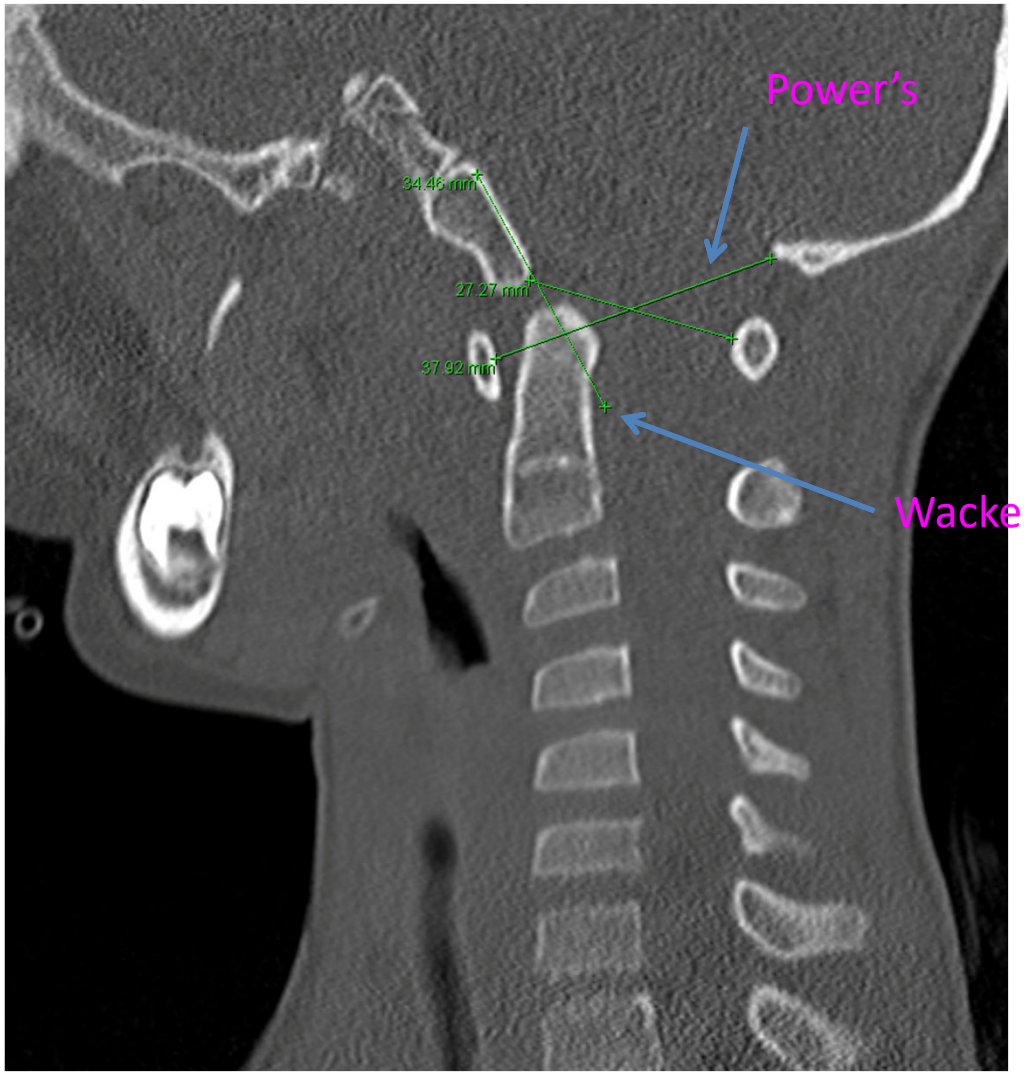
Anterior C4 corpectomy



How about Trauma?

- All young kids are ***relatively*** lax ligamentously
- Modulus of elasticity of C Spine in general is much greater than adults who tend to have more bony fractures

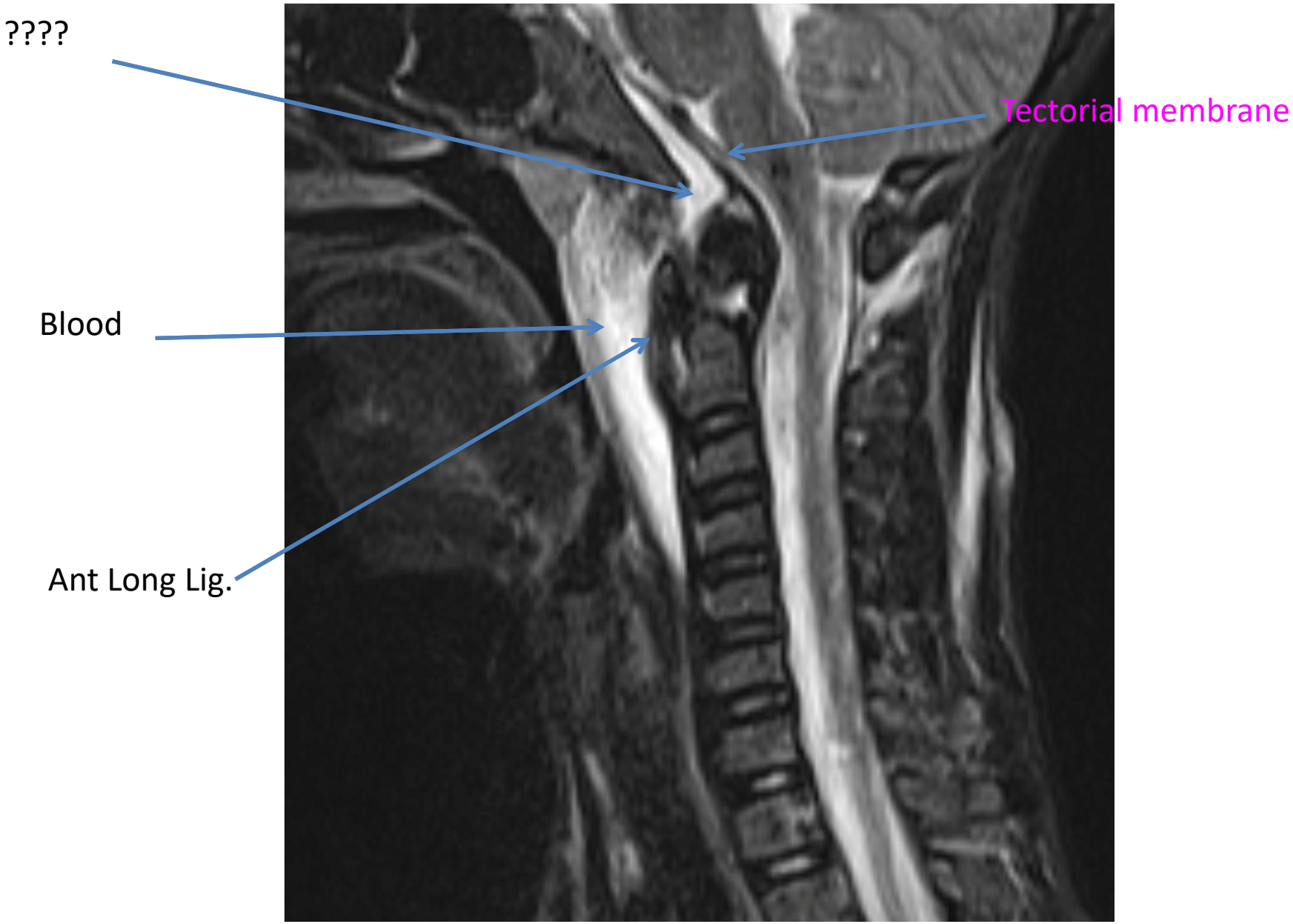
Nine yr female, MVC



CT Read:

IMPRESSION:
1. No CT evidence of cervical spine traumatic osseous abnormality.
2. However blood products in the right atlantoaxial and occipital axial intervals may suggest ligamentous injury at the craniocervical junction, correlation with MRI advised.
3. Positional related versus fixed head deviation to the right requiring clinical correlation.
4. Non displaced right posterior first rib fracture.

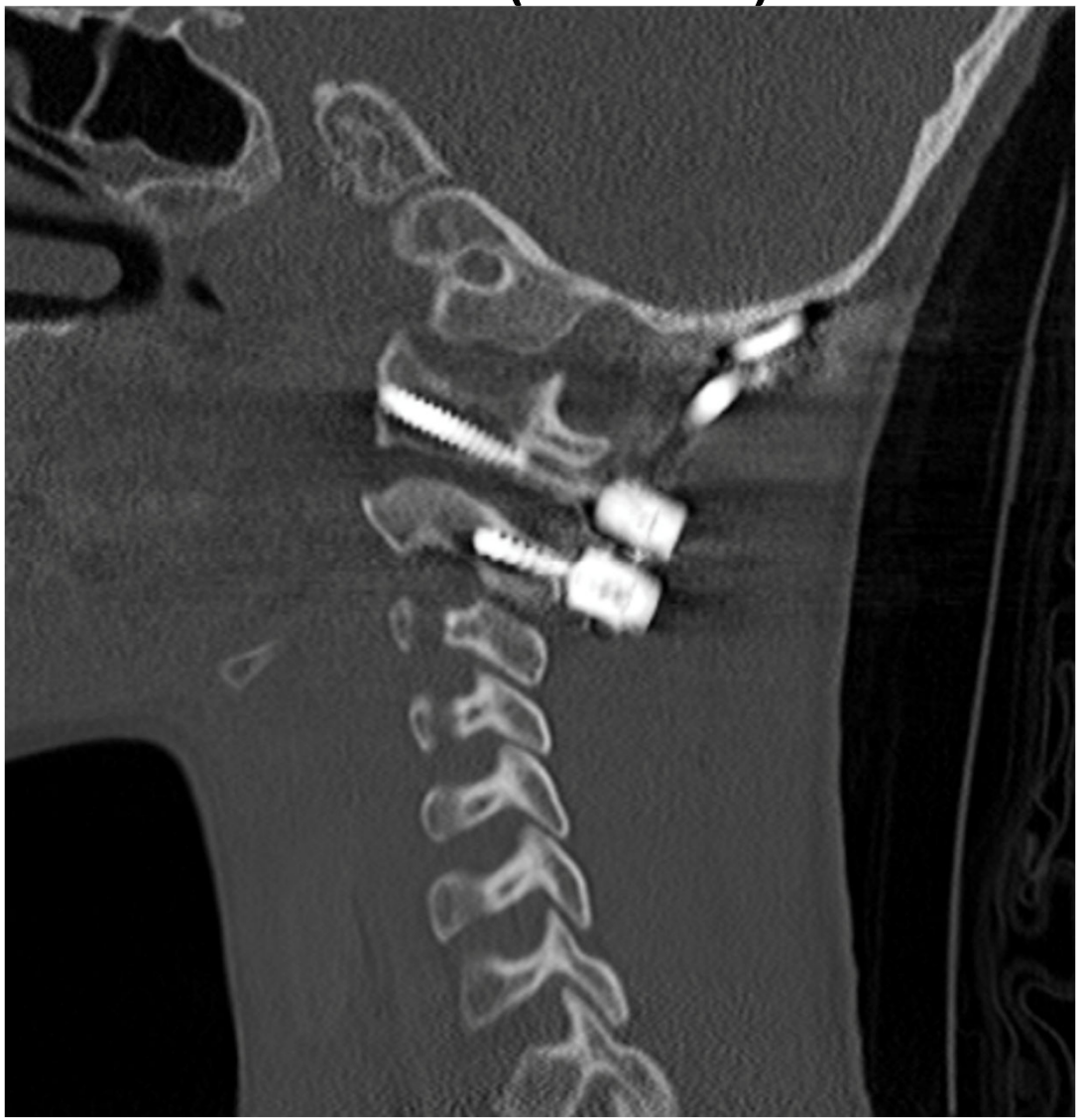
MRI



Diagnosis:

- Atlanto Occiptal dissociation
- Misnomer should be atlanto cervical dissociation because dens is dissociated also
- 5 yr old brother died in same accident, autopsy showed AOD
- 6 yr old brother had AOD also, mild, treated ‘conservatively’ in hard collar

O-C1/2 Fusion
Skull plate, C1 lateral mass screws, C2 pars screws (Harms)



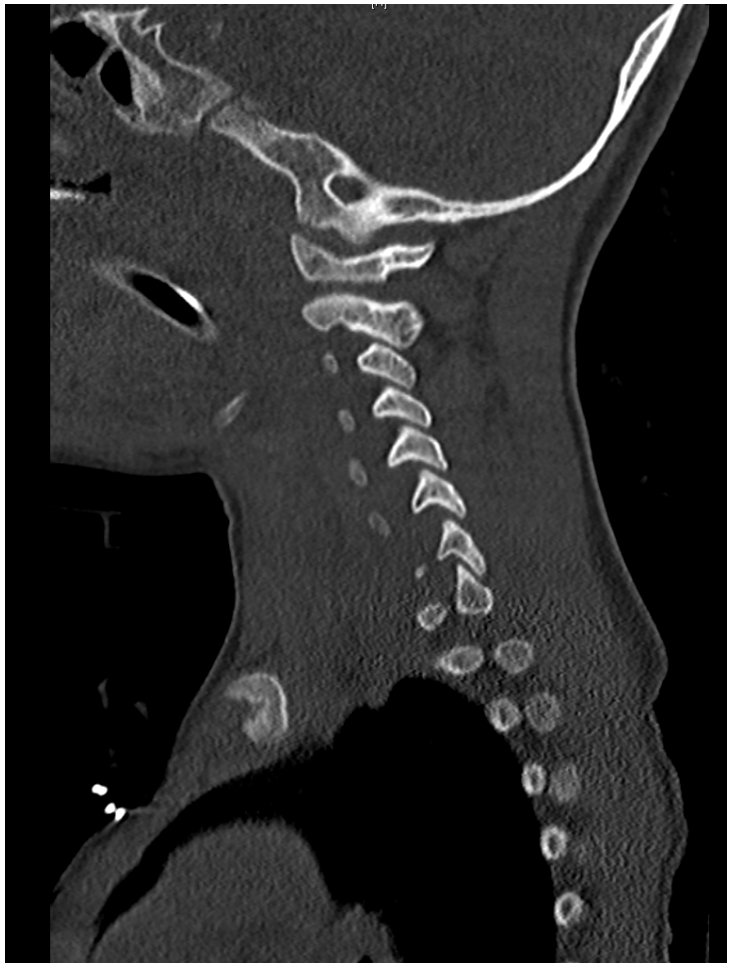
Postop R.O.M.

Not just at Skull base /C1/2

- Sub axial ligamnetous injuries much rare in small kids but do happen

5 yr female, ejected from car.
Possibly restrained?

Emergent Lap for small bowel perf. Quadriplegic / vented



Case courtesy of Jose Hererra-Soto

MRI C Spine

MRI T Spine

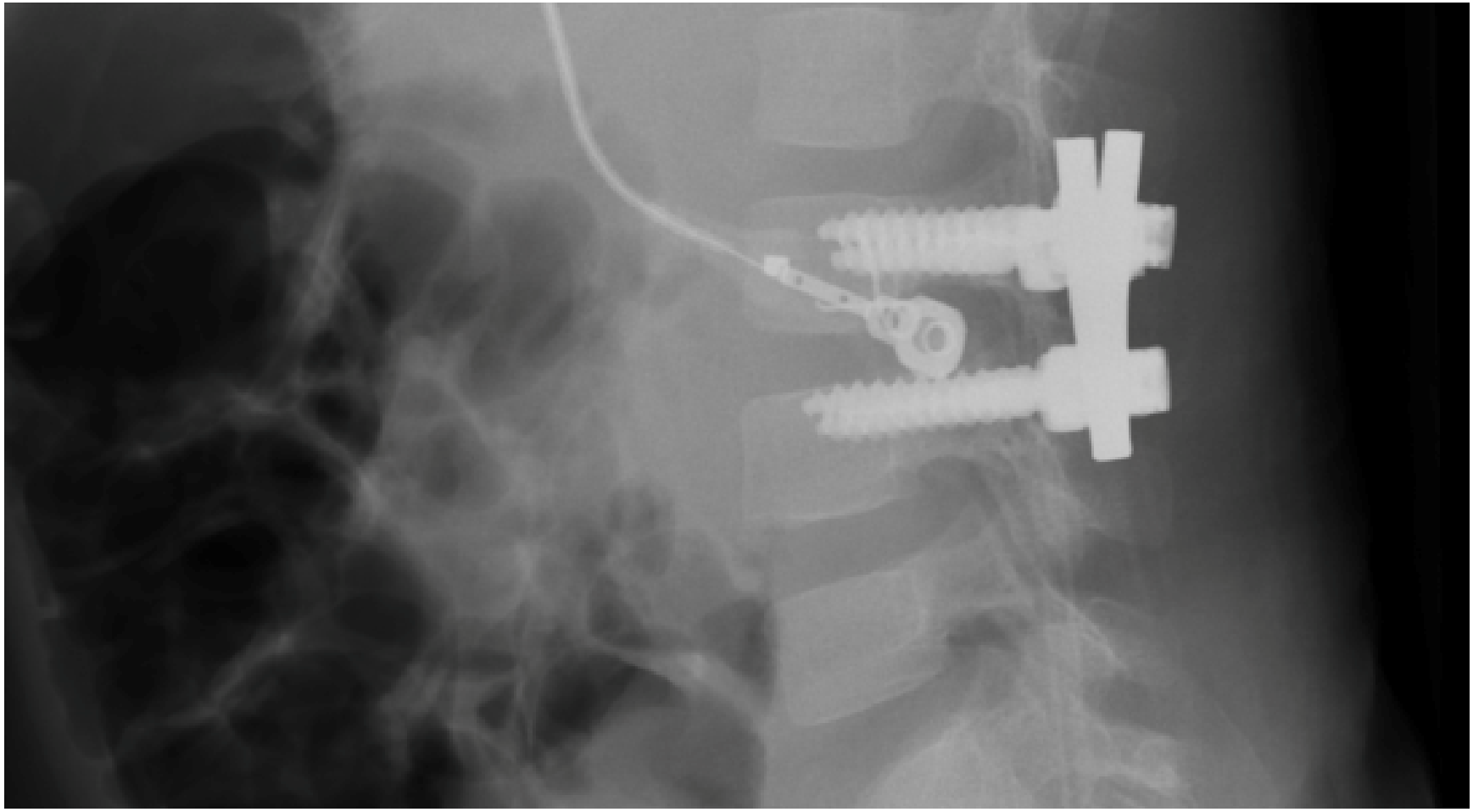
Imaging Summary at this stage:

- Chance fracture (soft tissue) L3/4
- C5/6/7 posterior ligamentous disruption
- C5/6 cord contusion
- T9/10 cord contusion

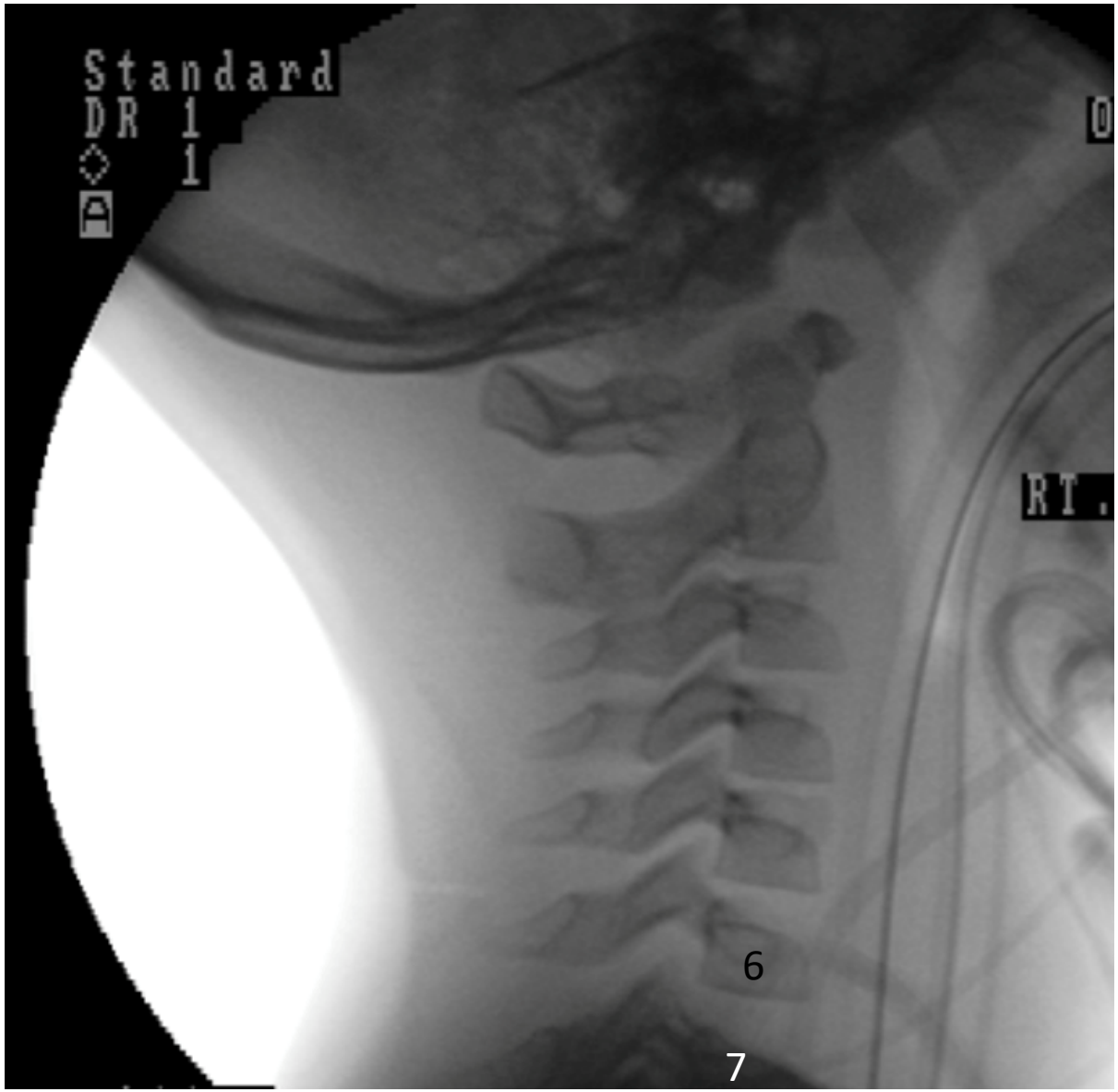
What to do?

- This is in effect a SCIWORA and Chance in the same patient
- Two level pathology
- Quadriplegic and vent dependen

Fixed Lumbar spine while exposing Cspine, no dural tear.



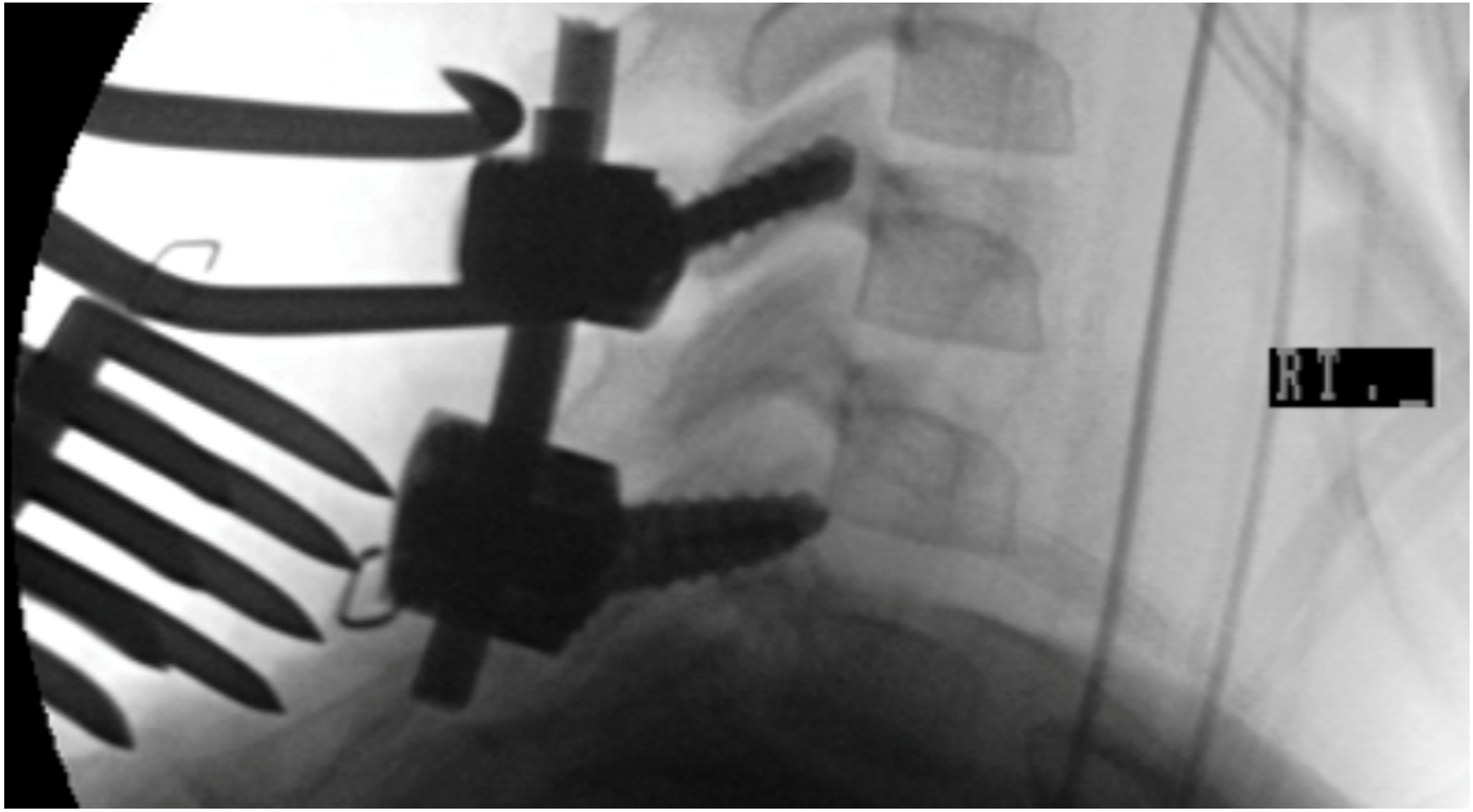
Halo and vest / Mayfield for C Spine



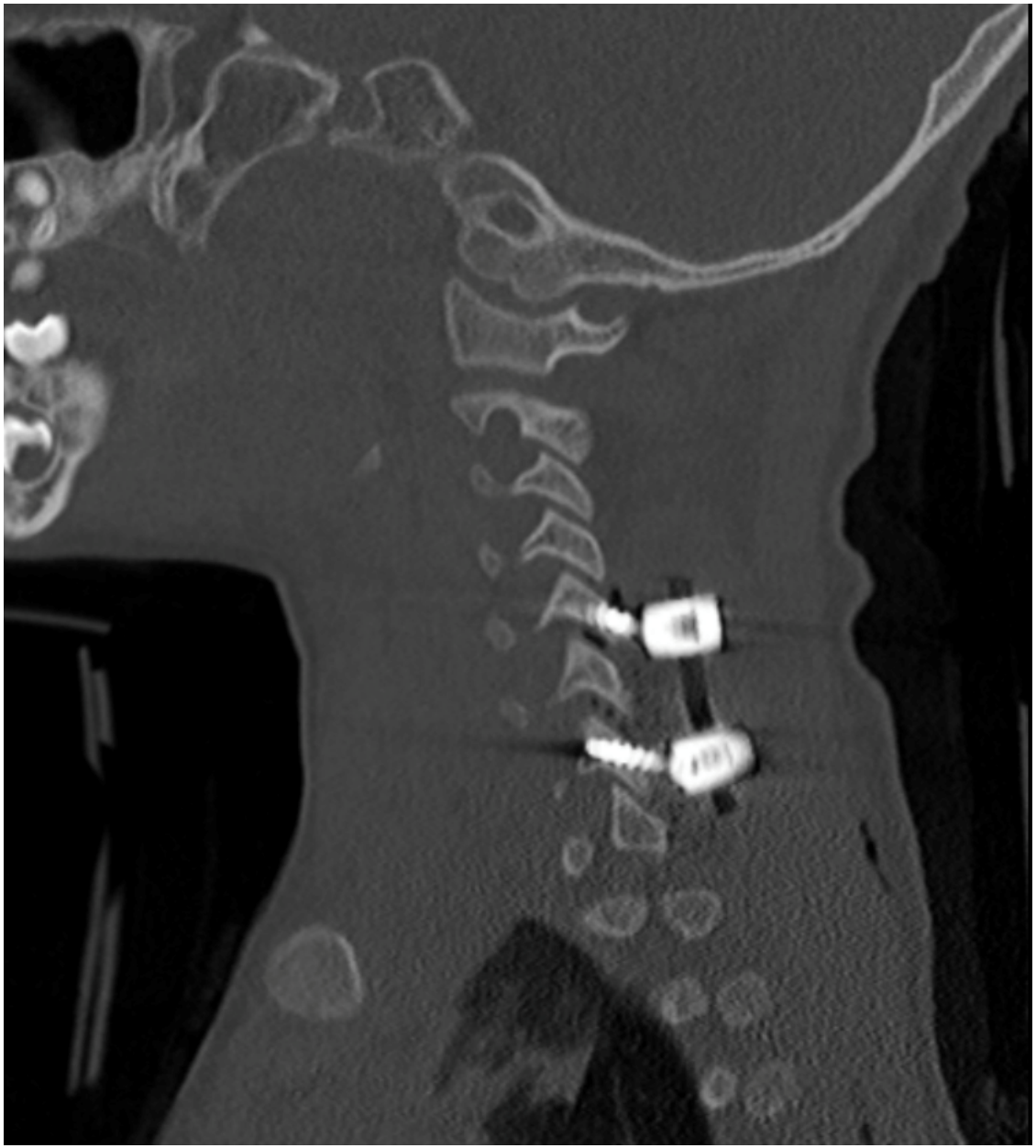
6

7

Capusles at C5/6/7 torn, no supra or interspinous ligaments left



Postop CT



Summary

- Ligamentous laxity has many aetiologies
- Look for multiple levels of involvement
- Know your anatomy well
- It’s not just about Down, not just at O / C1/2
- Know various techniques for surgical stabilisation including halo vest
- Use advanced imaging liberally in preop planning