Paediatric C-spine

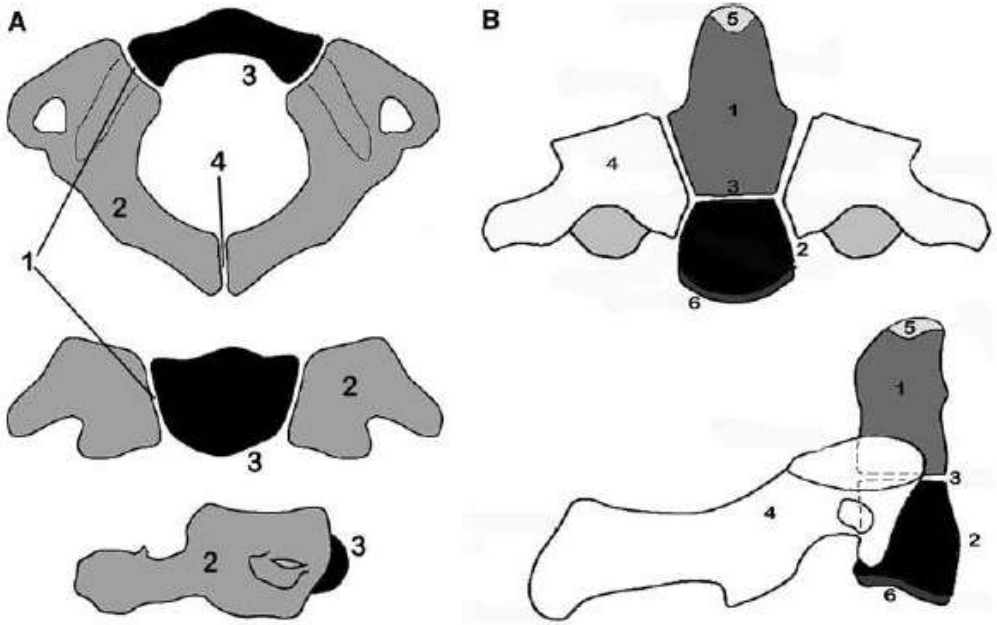
EMBROYOLOGY

ATLAS

- 3 ossification centers
- Internal diameter - 6 to 7 yrs of age.
- Further growth- periosteal appositional growth.
- **Surgical fusion should not be performed** before this age because of the potential for later cervical stenosis

AXIS

- 4 ossification centers
- **Both the posterior and the anterior synchondroses are closed by 6 years of age**

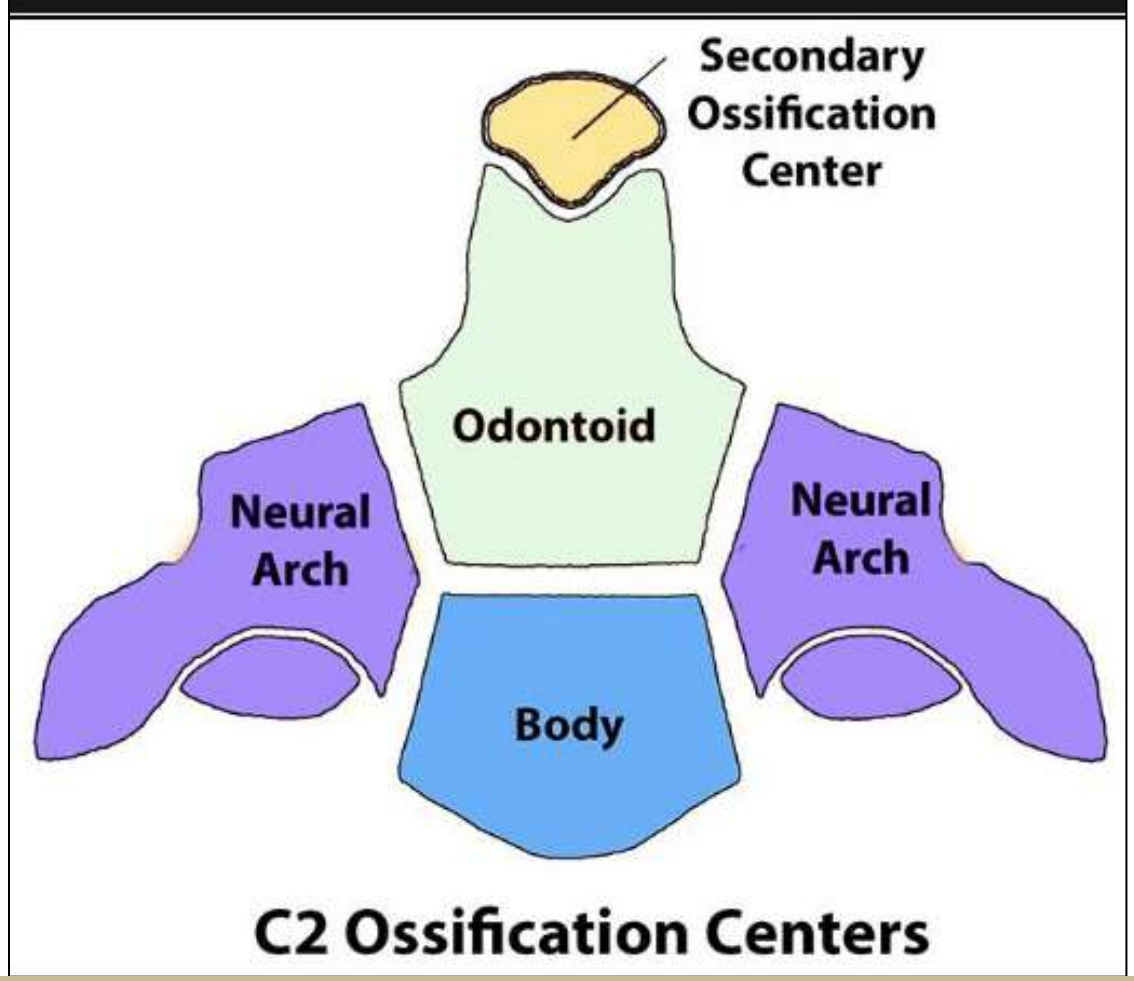


ATLAS

- Ossification is present only in the two neural arches at birth.
- Body 6 months and 2 years
- By 4 to 6 years posterior synchondrosis fuses >> anterior

AXIS

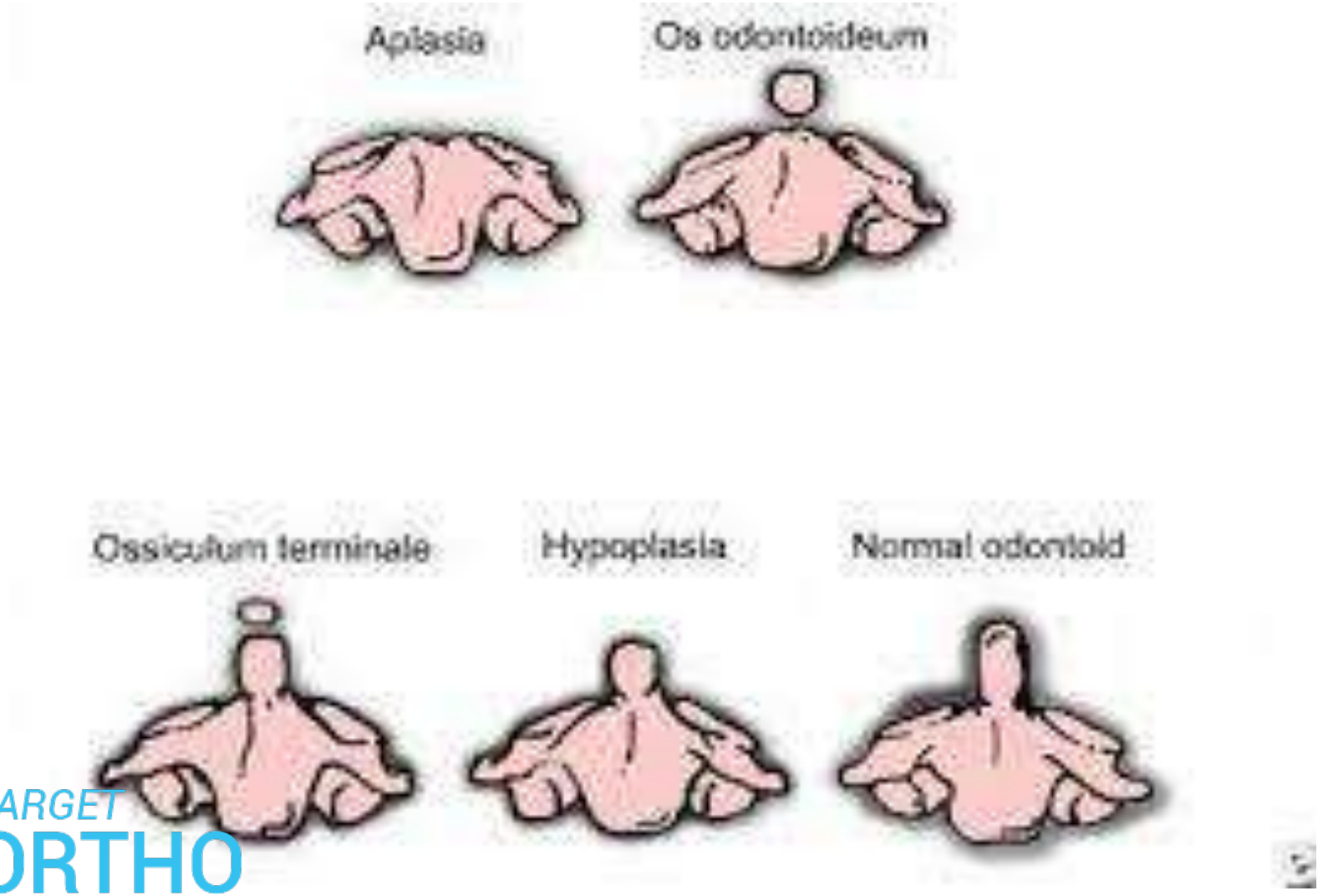
- between 5 and 8 years of age, tip of dens develops an ossification center, becoming the **ossiculum terminale**.
- The ossiculum terminale fuses to the remainder of the odontoid between 10 and 13 years of age

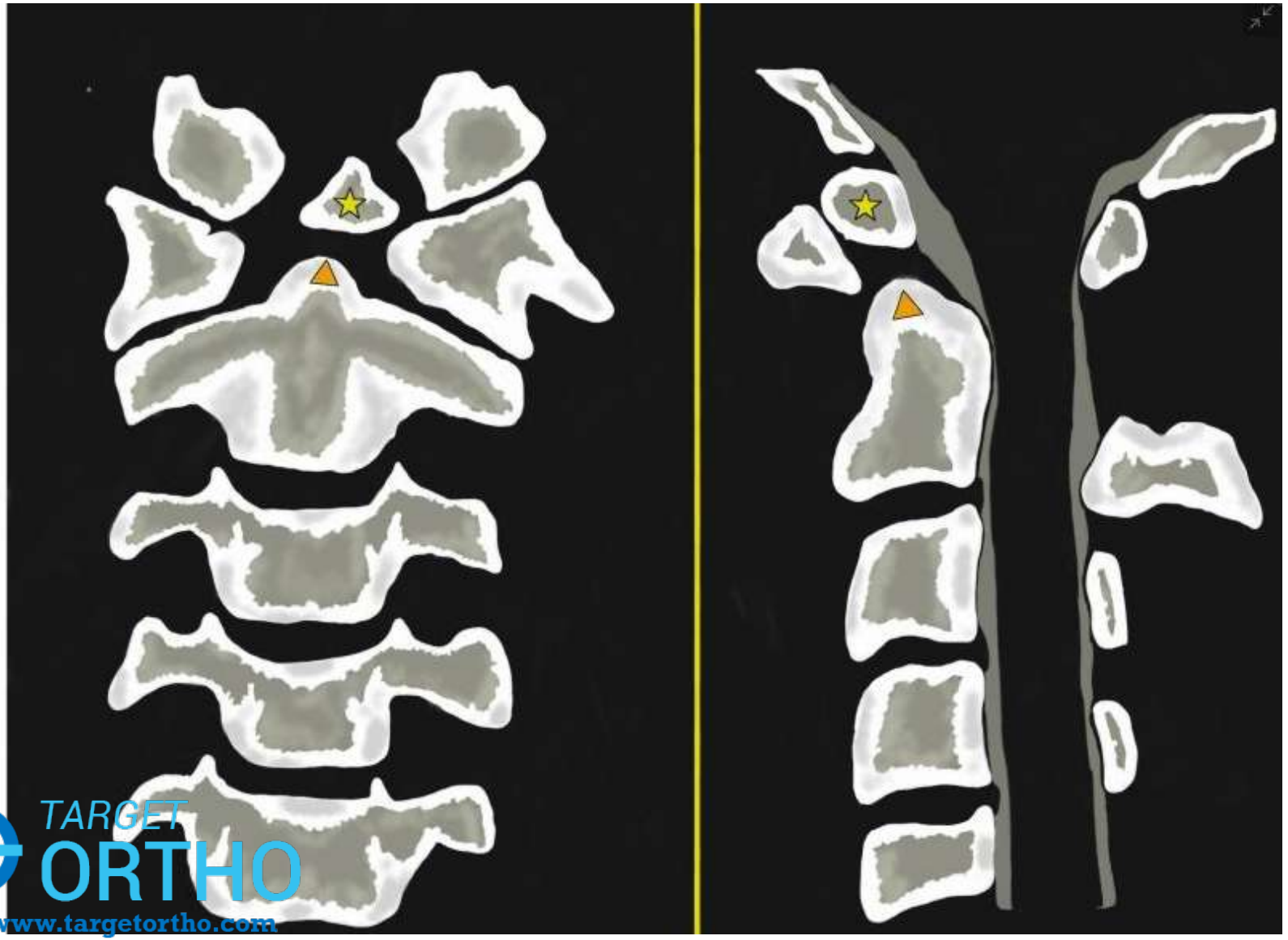


Failed fusion of odontoid and axis vertebral body

1. an incomplete migration of the axis centrum,
2. failure of segmentation
3. nontraumatic avascular necrosis

Anomalies of Odontoid



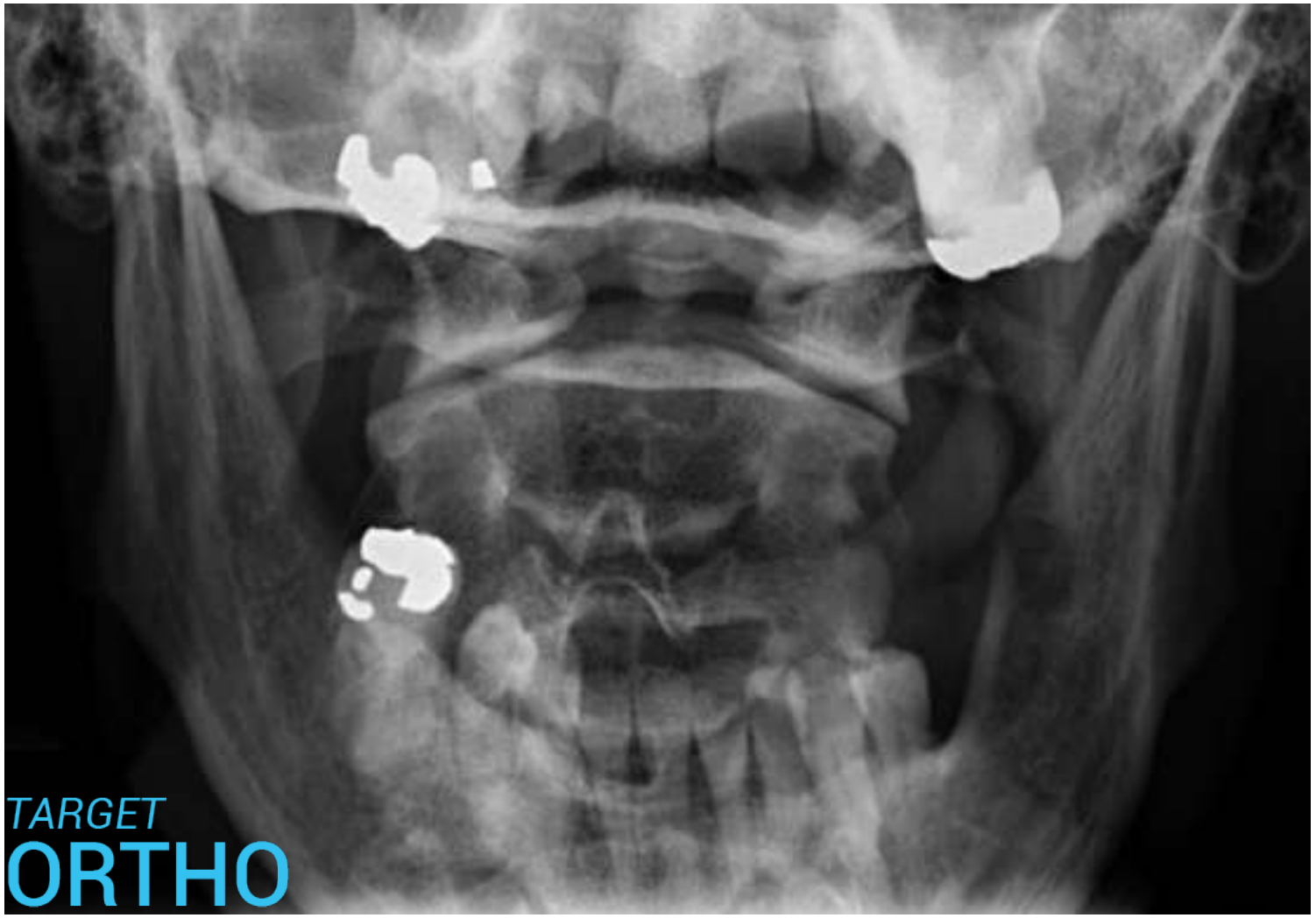


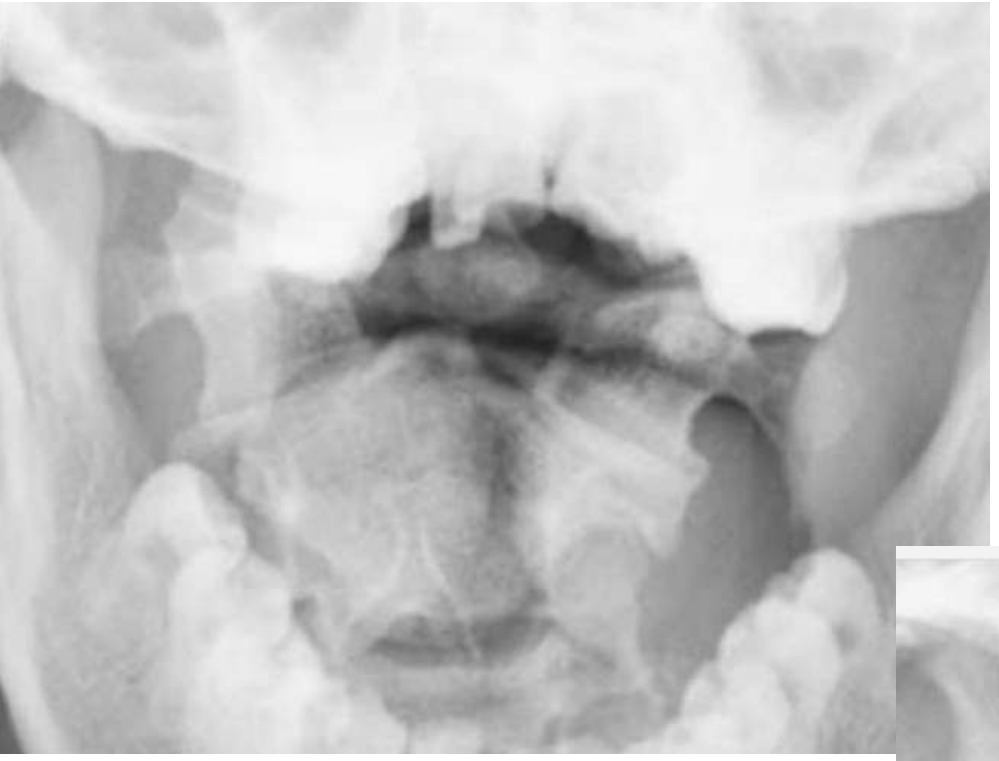
Os odantoidium

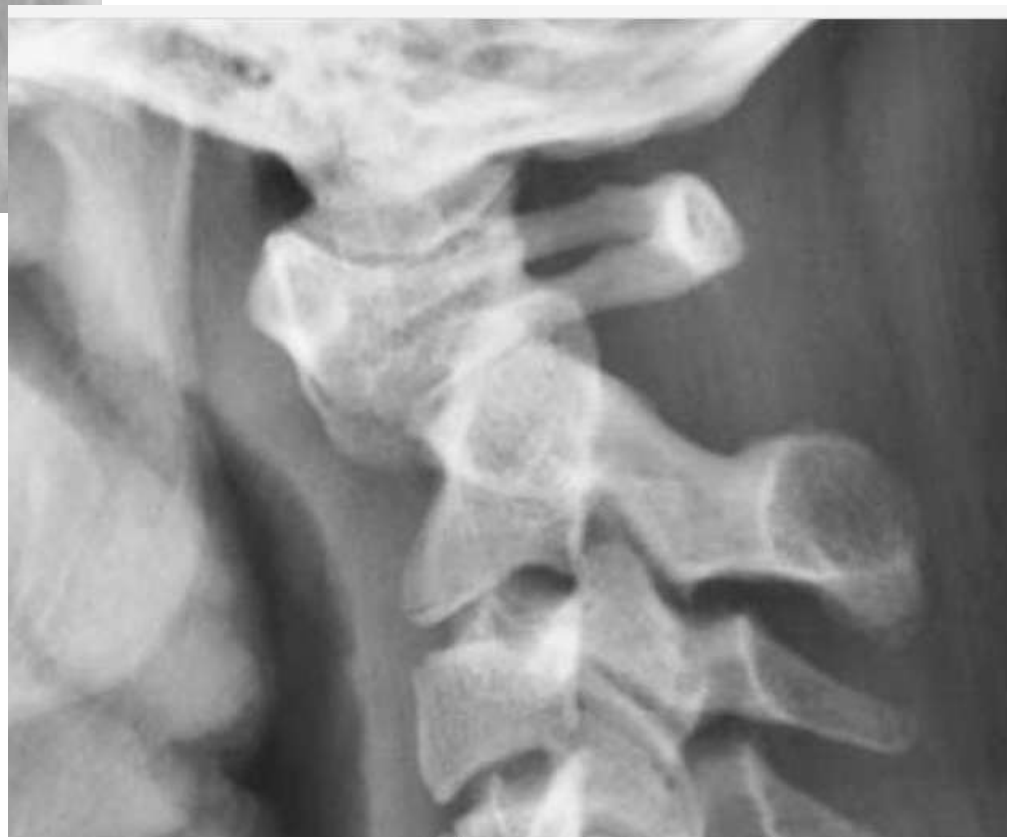
- Age of diagnosis- 18yrs – 30 yrs
- Asso. With Downs , Morquio, KF syndrome.
- **Presentation**- Occipito cervical pain

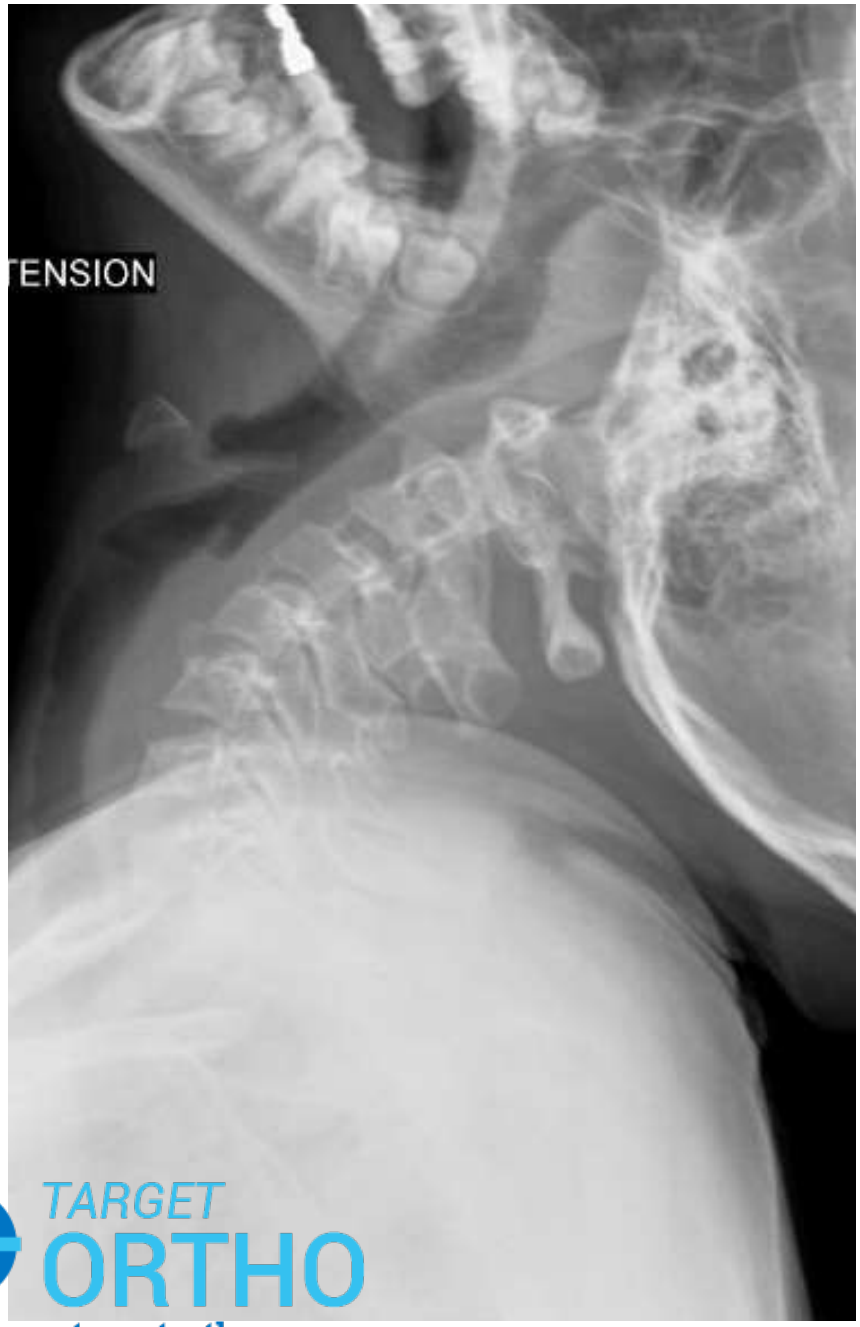
  Myelopathy – cord compression

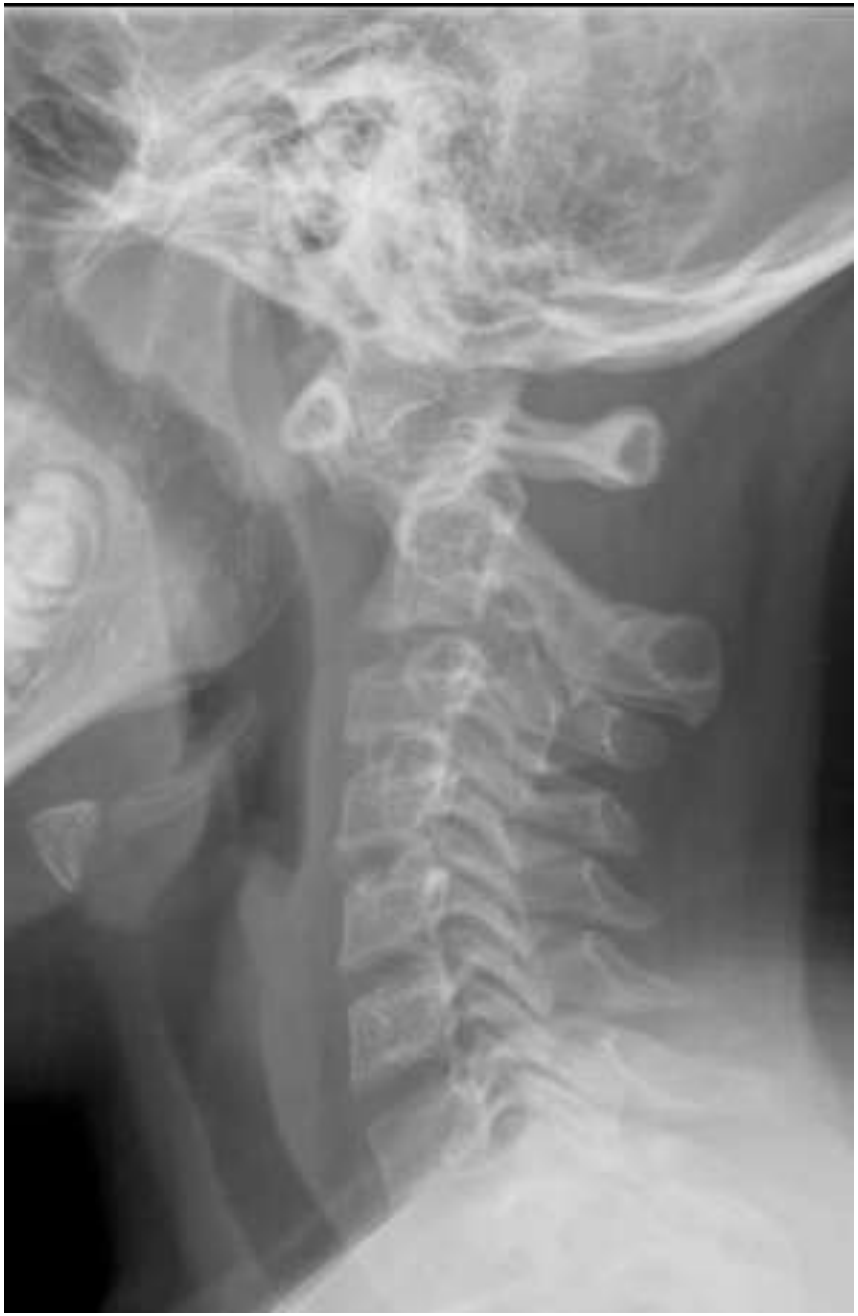
- Asymptomatic/ incidental finding- observation
- Occipito cervical pain- in instability is seen – operate otherwise conservative Tt.
- Cervical myelopathy- reduction & fixation

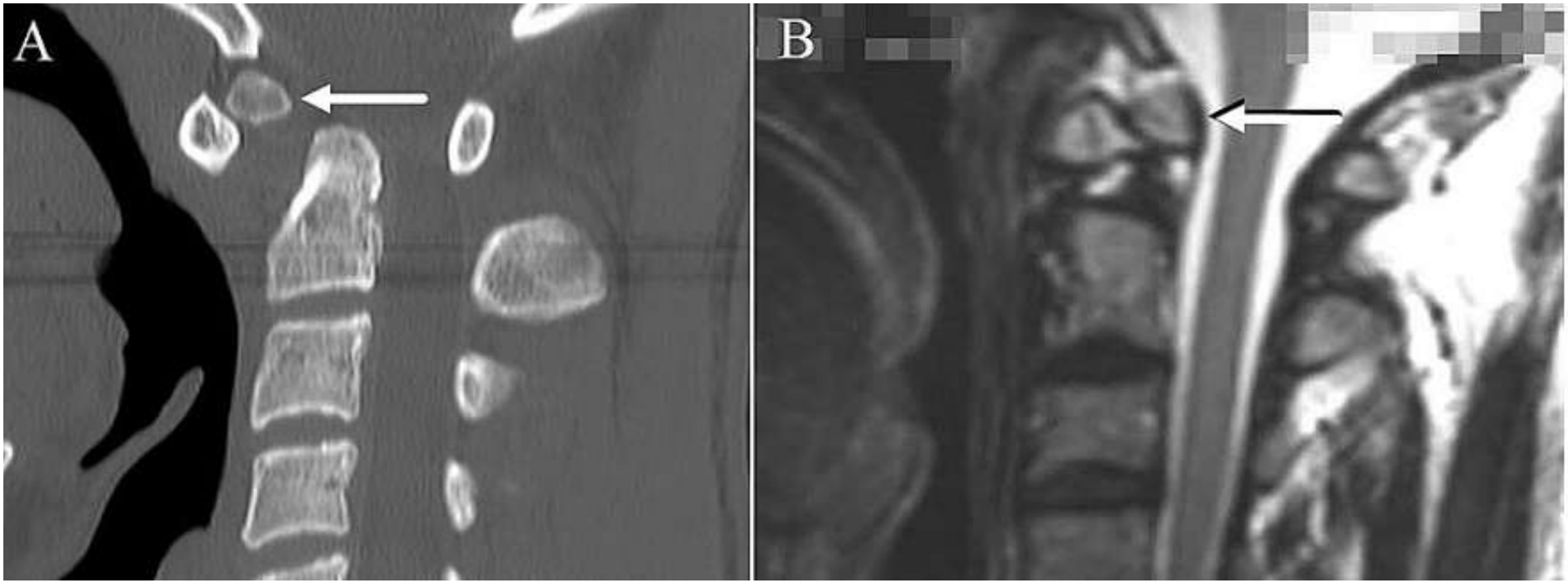




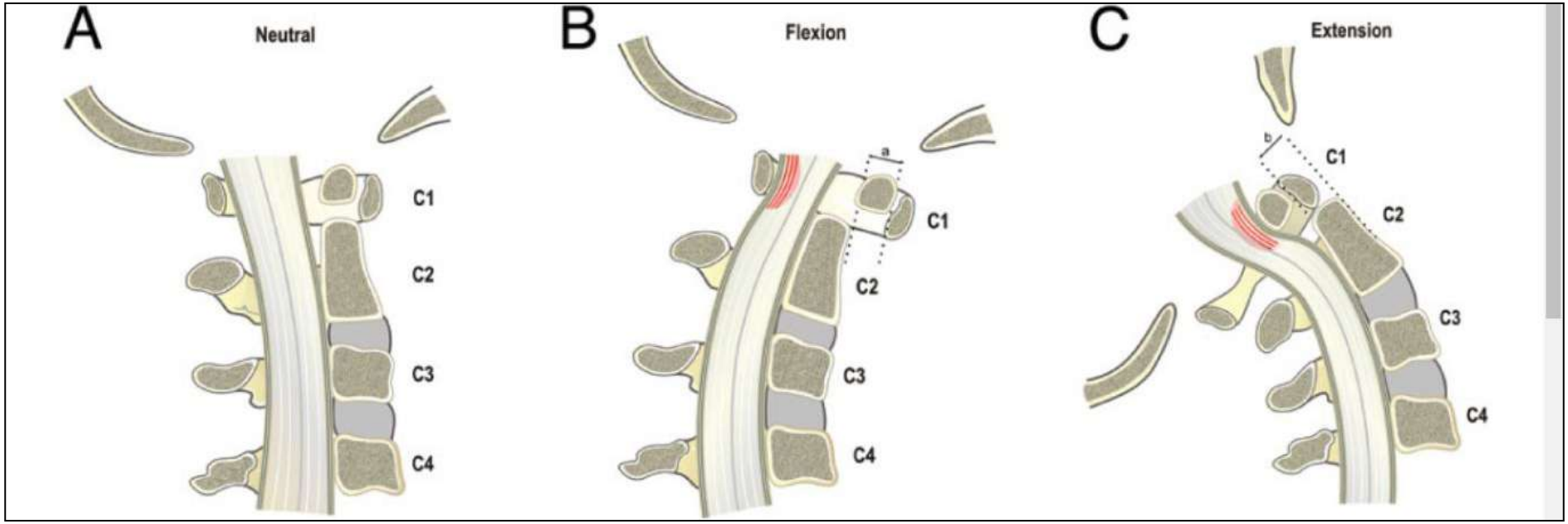


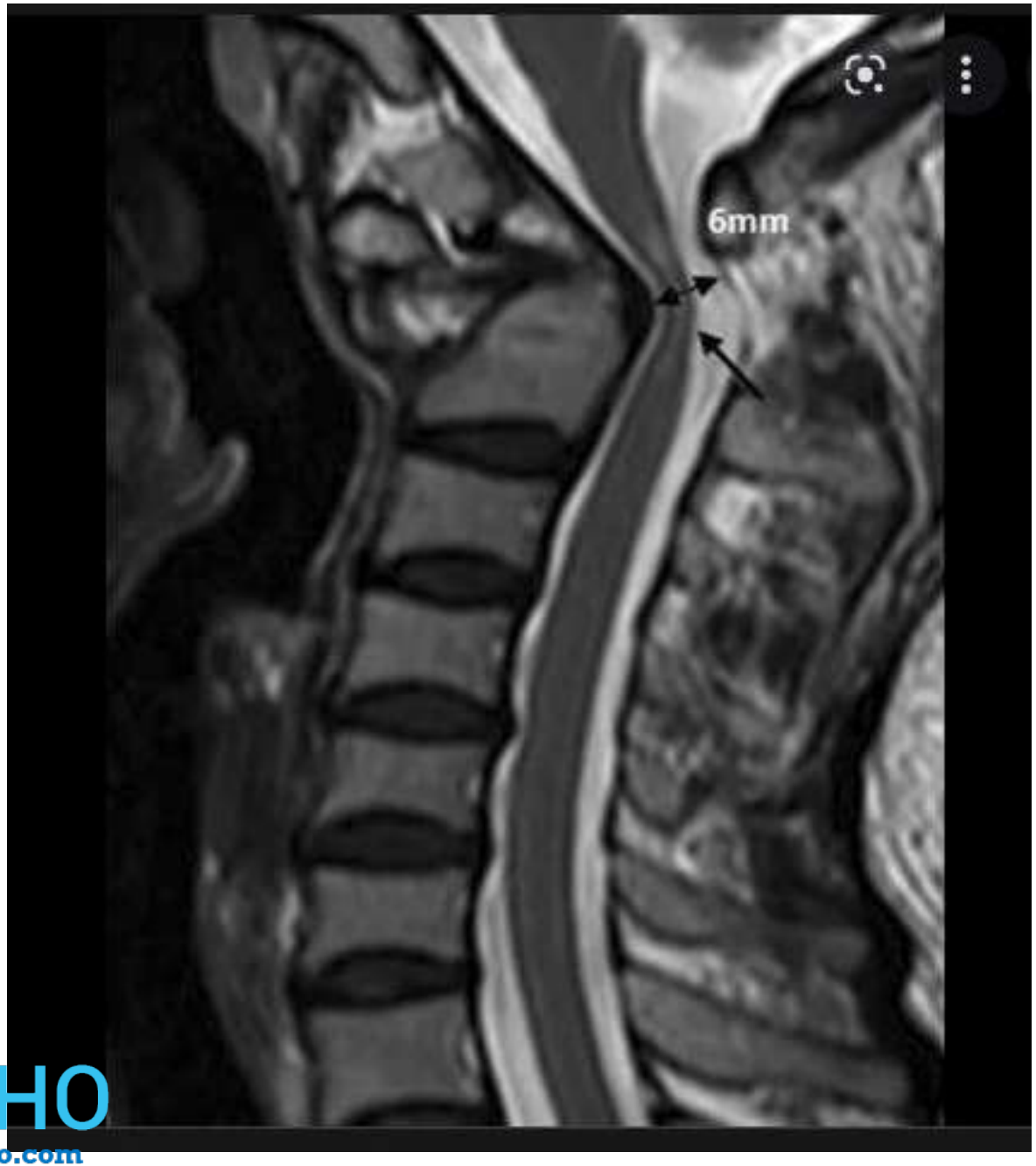


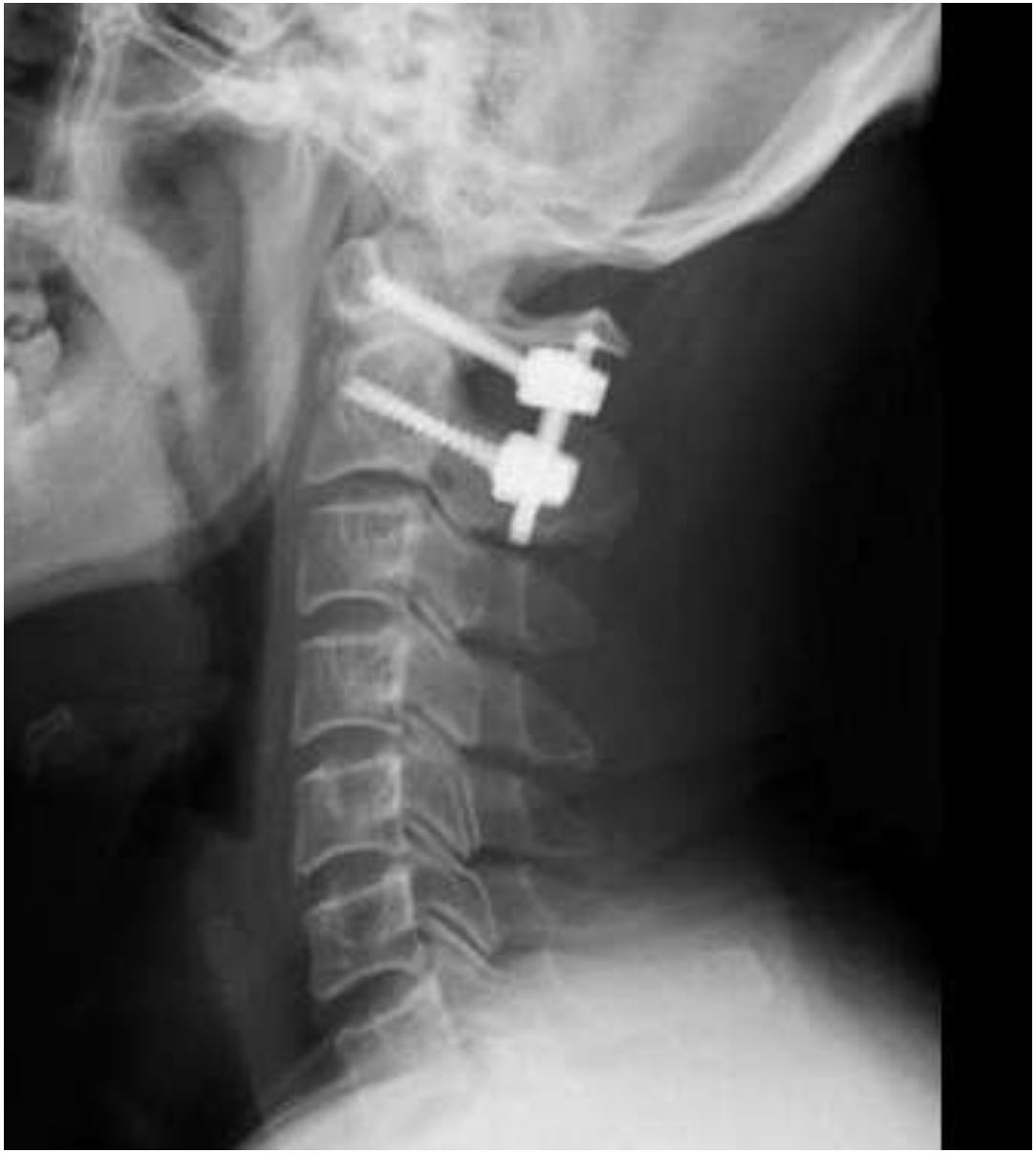




Os odantoidium- dynamic instability







BASILAR IMPRESSION

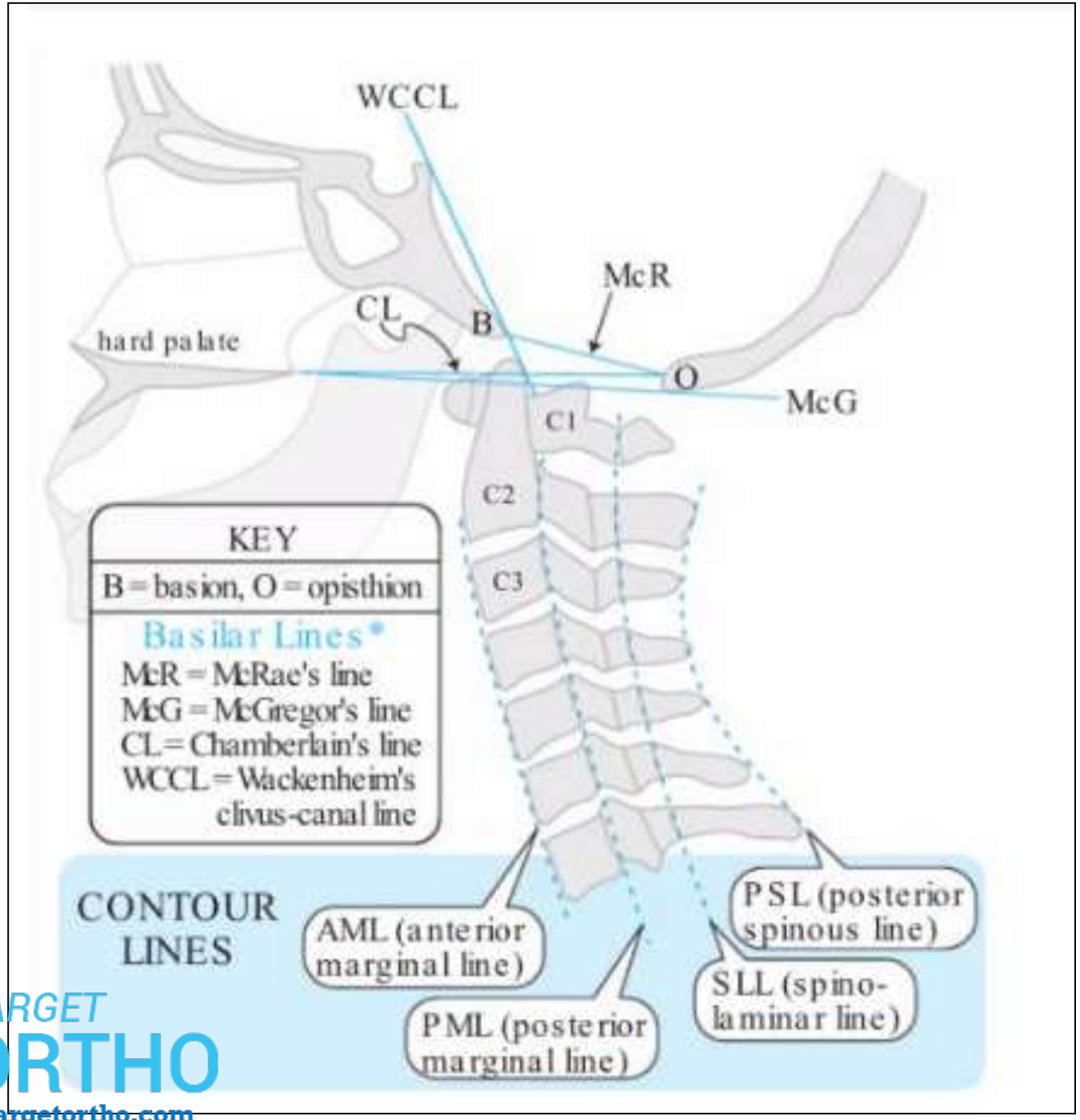
- Indentation of the skull floor by the upper cervical spine.
- The tip of the dens is more cephalad--protrudes into the opening of the foramen magnum.
- Neurologic damage from direct injury, vascular compromise, or alterations in cerebrospinal fluid flow.

PRIMARY

- **Congenital abnormality**
- often associated with other vertebral defects
  - Klippel-Feil syndrome
  - odontoid abnormalities
  - atlantooccipital fusion
  - atlas hypoplasia

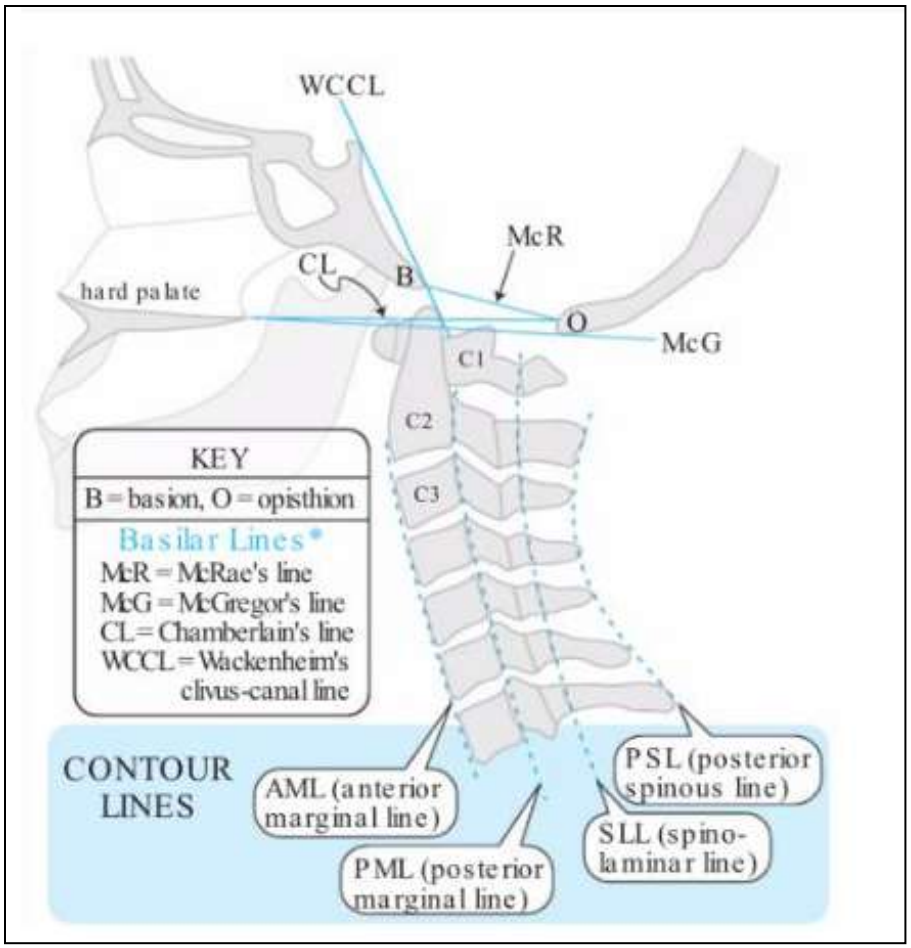
SECONDARY

- **Developmental condition**
- **Softening of the osseous structures at the base of the skull**
- Paget disease
- renal osteodystrophy
- rickets and osteomalacia
- bone dysplasias and osteogenesis imperfecta ,
- achondroplasia
- hypochondroplasia
- neurofibromatosis
- Rheumatoid arthritis and ankylosing spondylitis



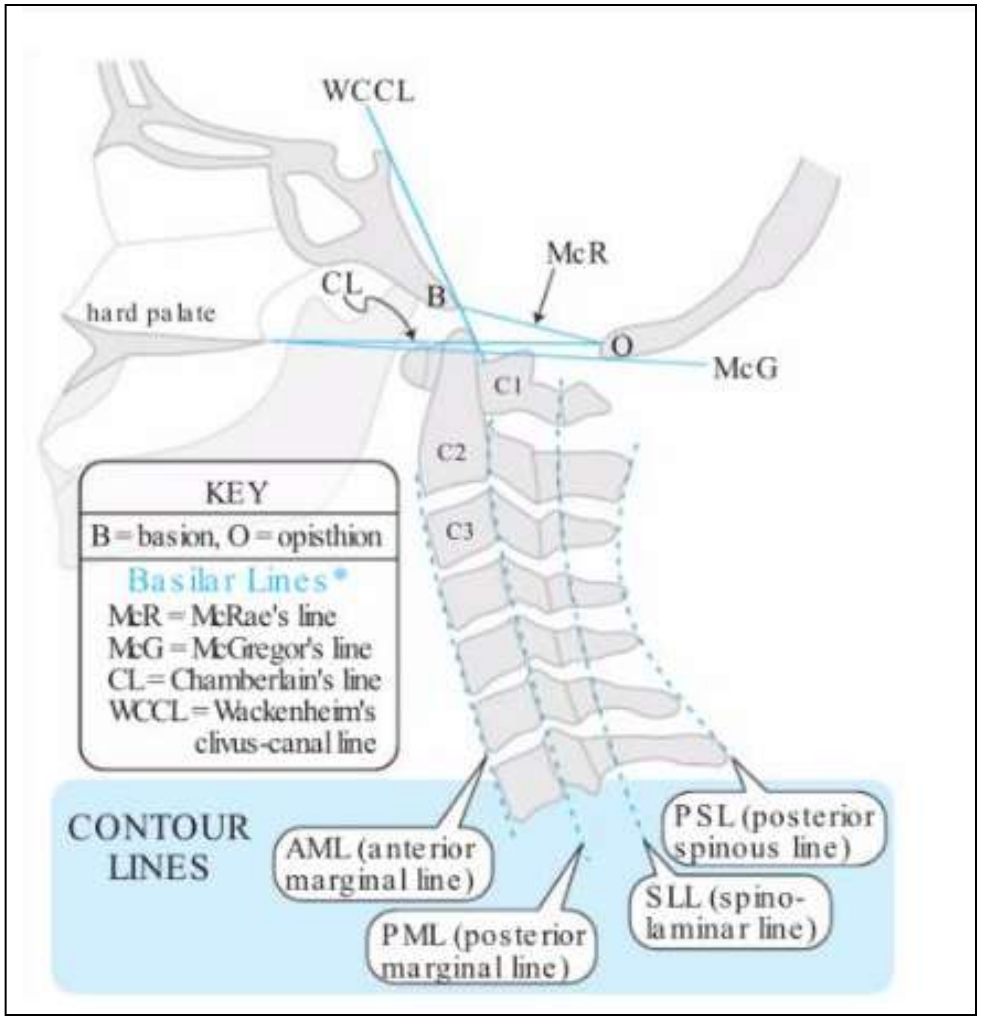
- **Chamberlain line** - back of hard palate with the opisthion(posterior margin of foramen magnum) on a lateral view of the craniocervical junction.

- Abnormal if tip of the odantoid is more than 3mm above this line

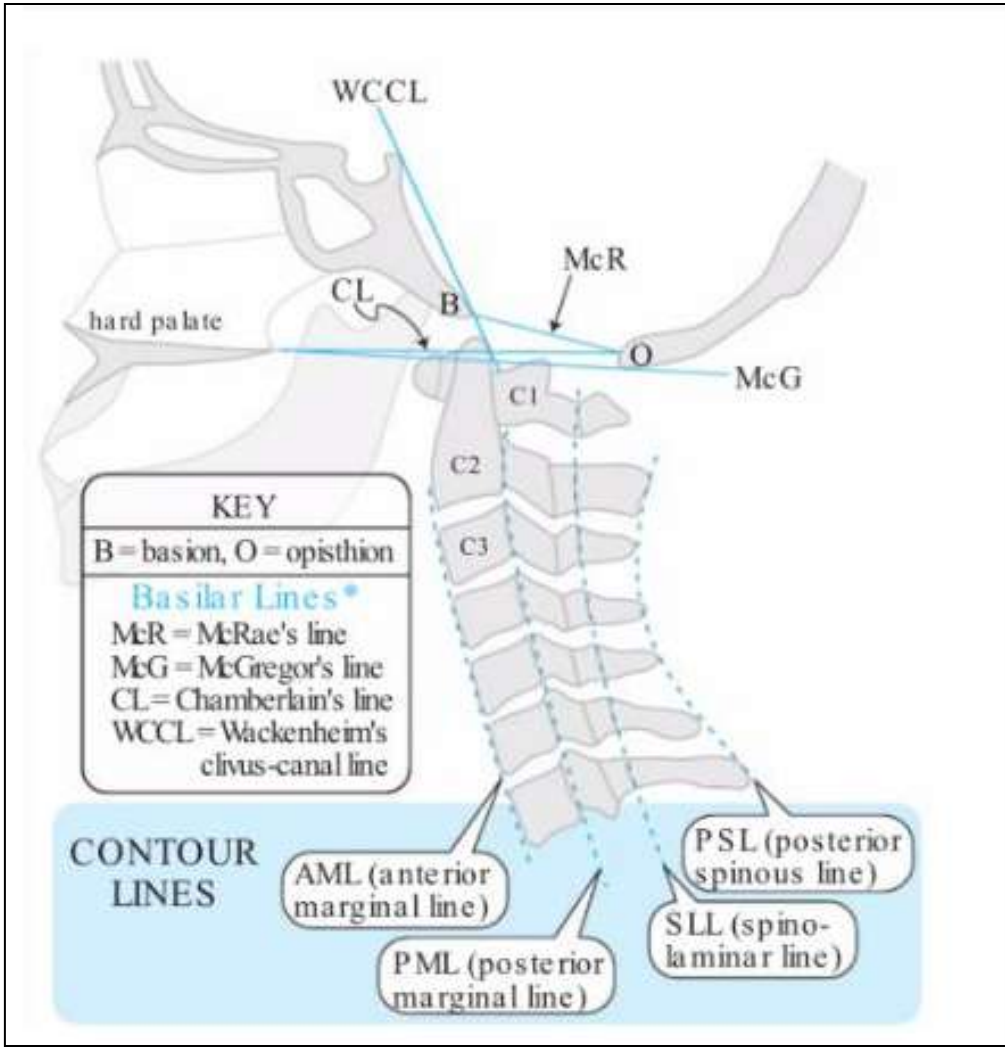


- **Mcgregor line**
- Superior margin of hard palate to inferior margin of occiput

- Odantoid tip more than 4.5mm above the line is abnormal.



- **Mc Rae line-** opening of foramen magnum
- Odantoid tip should be below this line



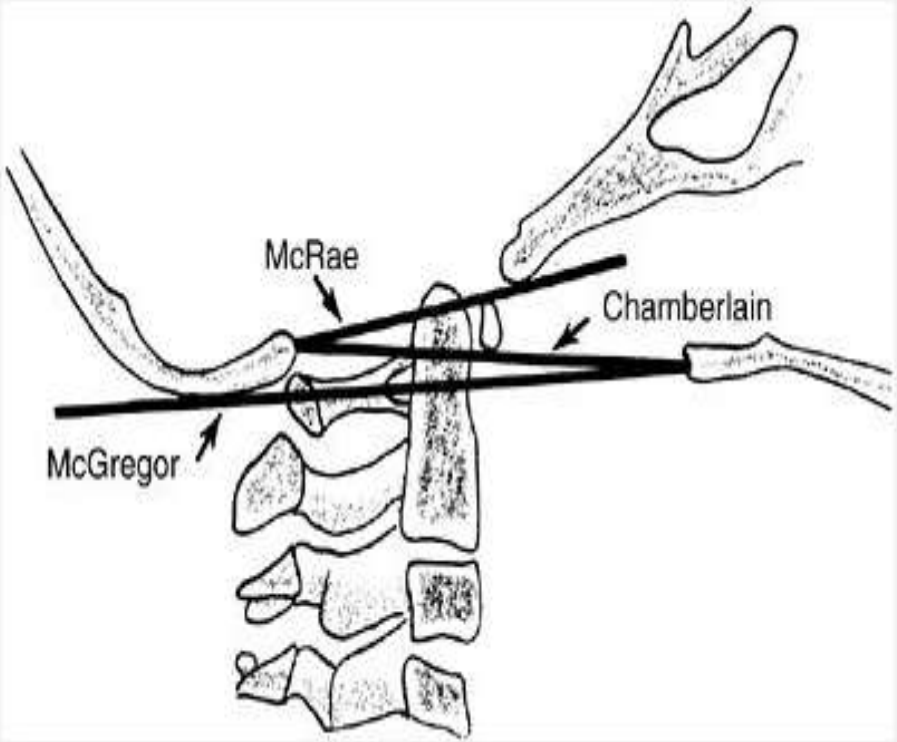
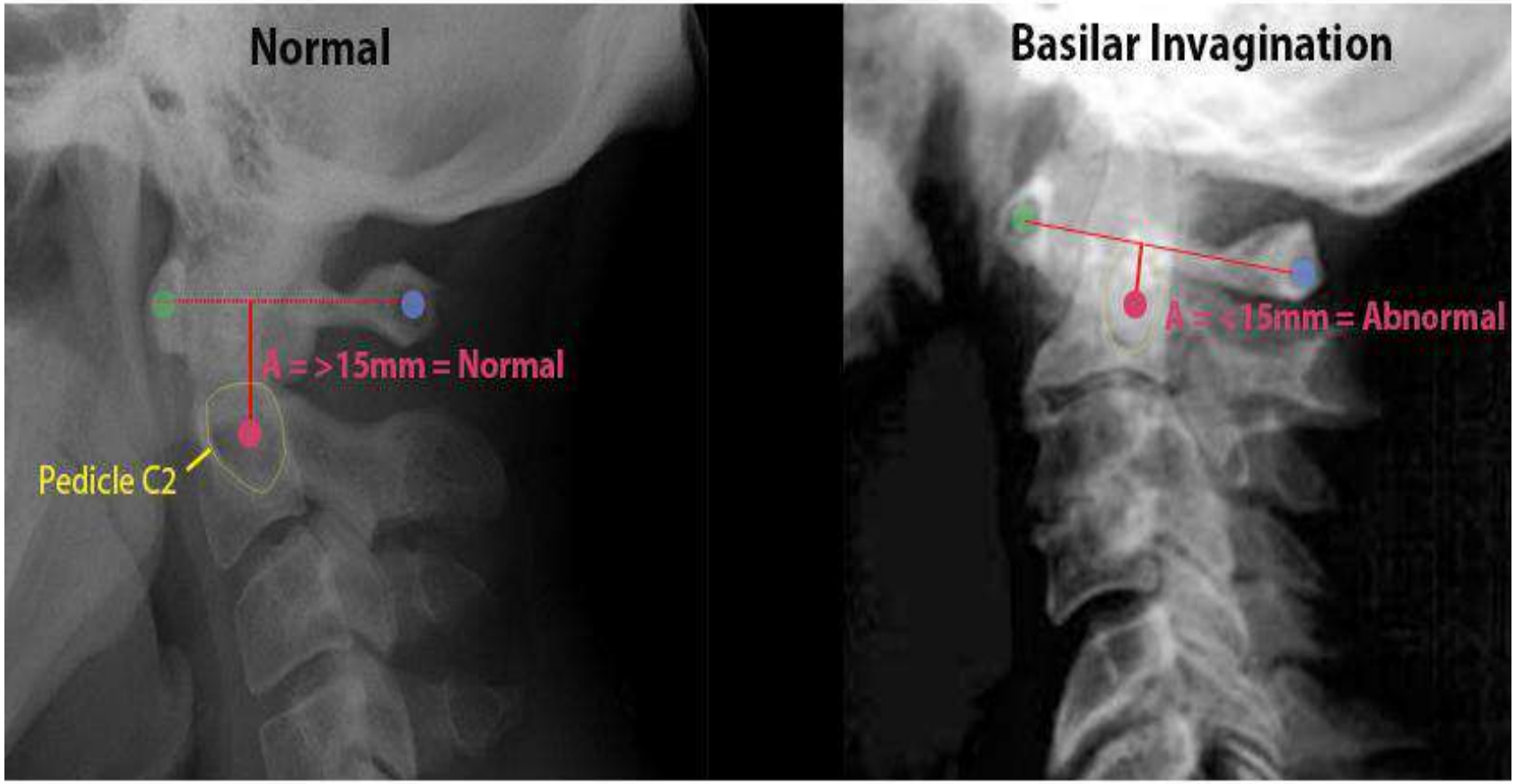
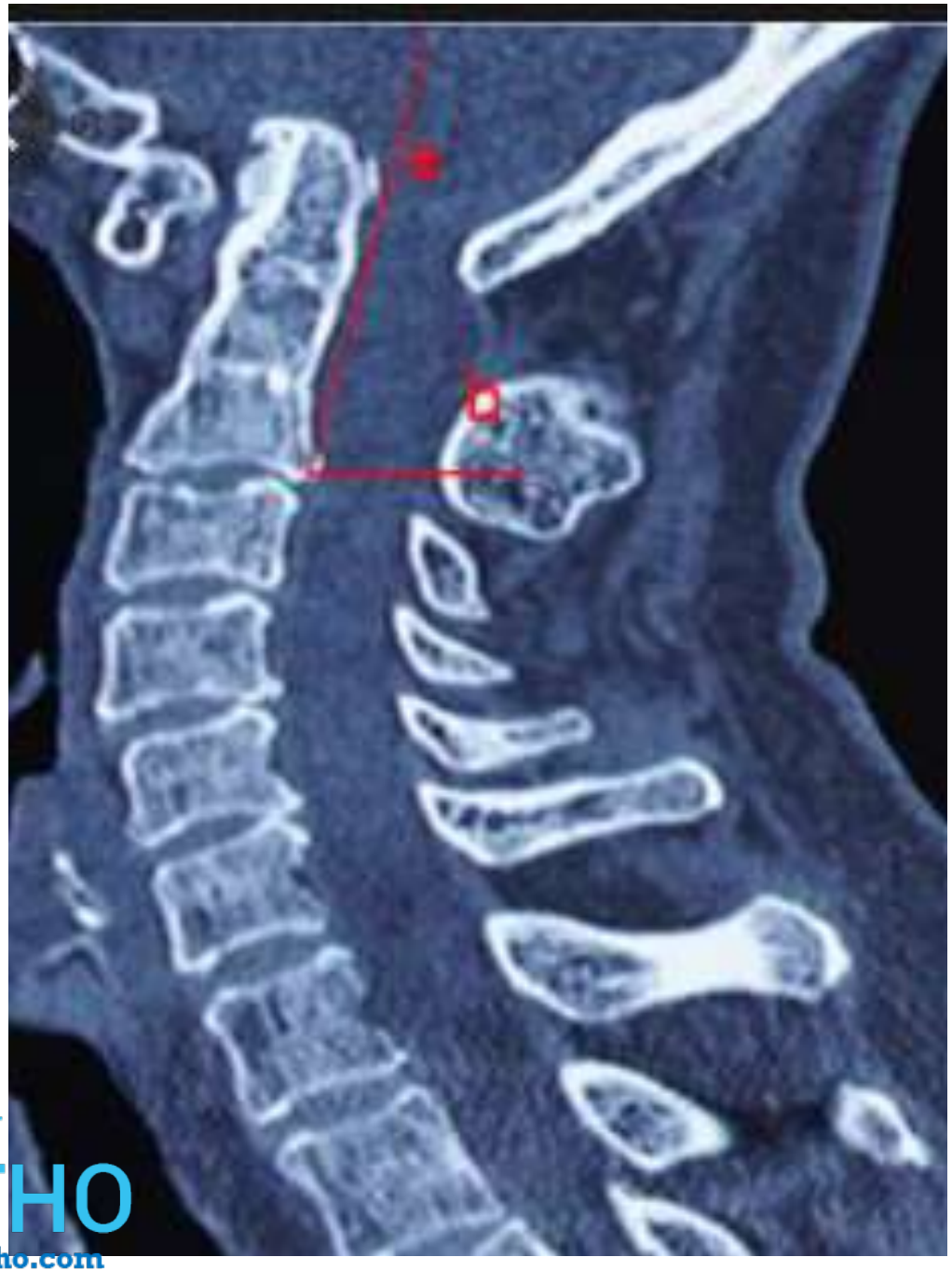


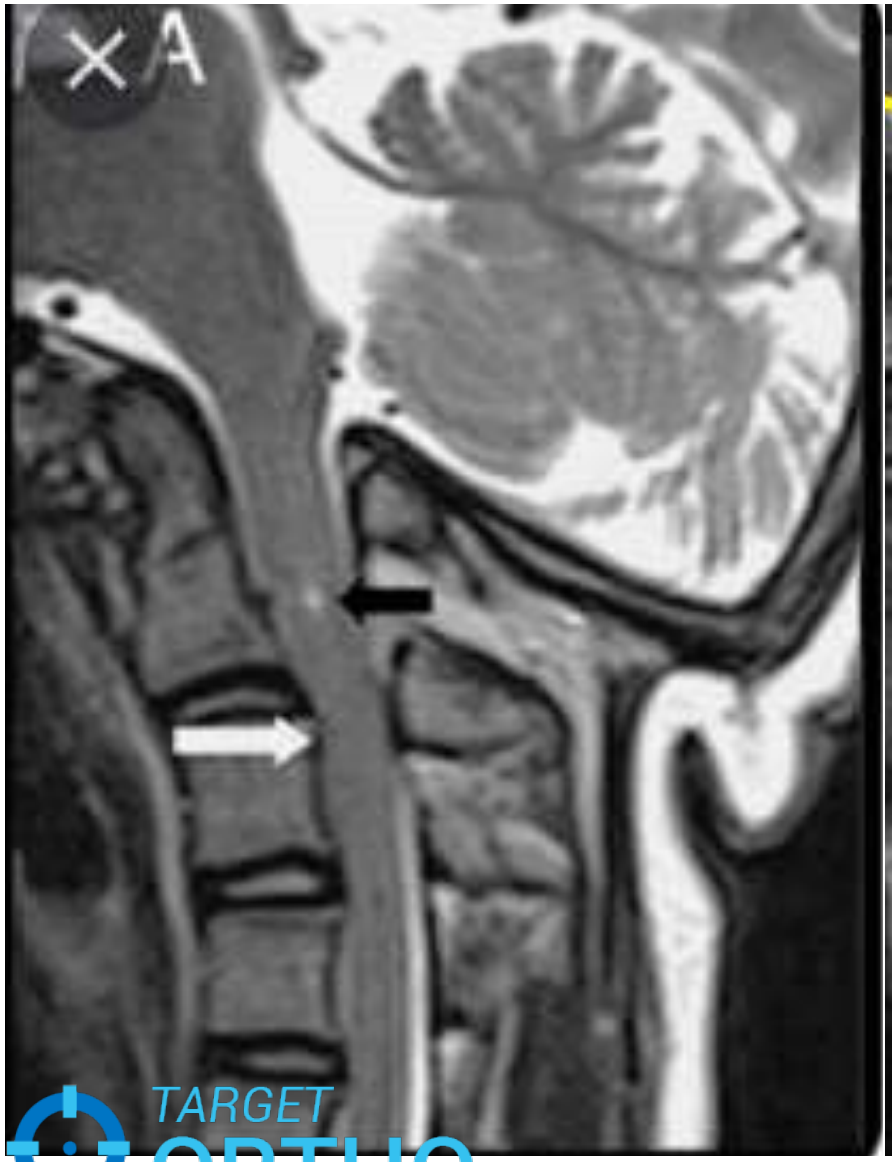
Figure 22.7 The landmarks used on a lateral radiograph of the skull and upper cervical spine used to assess basilar impression. McRae's line defines the opening of the foramen magnum. Chamberlain's line is drawn from the posterior lip of the foramen magnum to the dorsal margin of the hard palate. McGregor's line is drawn from the upper surface of the posterior edge of the hard palate to the most caudal point of the occipital curve of the skull. McGregor's line is the best for screening because of the clarity of the radiographic landmarks in children of all ages.

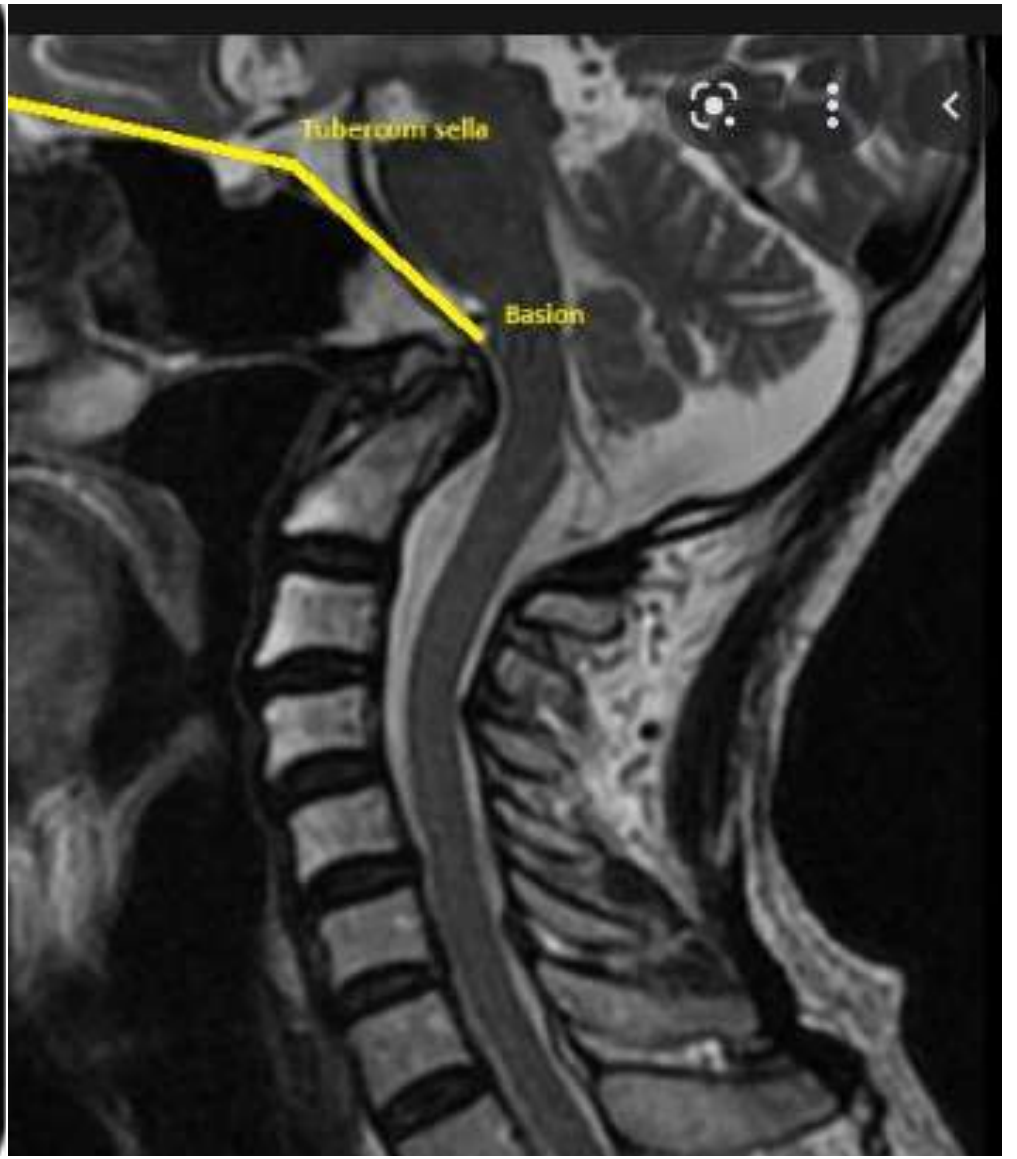
Ranawat C1-2 Index (A)



The Ranawat index is used to assess vertical subluxation (VS) by detecting settling of C1 on C2. The measurement is made from the center of the pedicles of C2 to a line connecting the midpoint of the anterior and posterior arches of C1. Normal values are 15 mm or greater for men and 13 mm or greater for women. Anything less than this confirms VS.







Clinical features

- Short neck (**apparent shortening)**
- Asymmetry of the skull and face
- Painful cervical motion
- Torticollis can also occur.
- Neurologic signs and symptoms are often present (motor weakness, limb paresthesias)- develops at 2nd /3rd decade.
- PYRAMIDAL ISOLATED

- Associated with Arnold-Chiari malformations - the neurologic involvement is usually **cerebellar**, and symptoms include motor incoordination with ataxia, dizziness, and nystagmus.

- Ataxia is a very common finding in children with basilar impression.

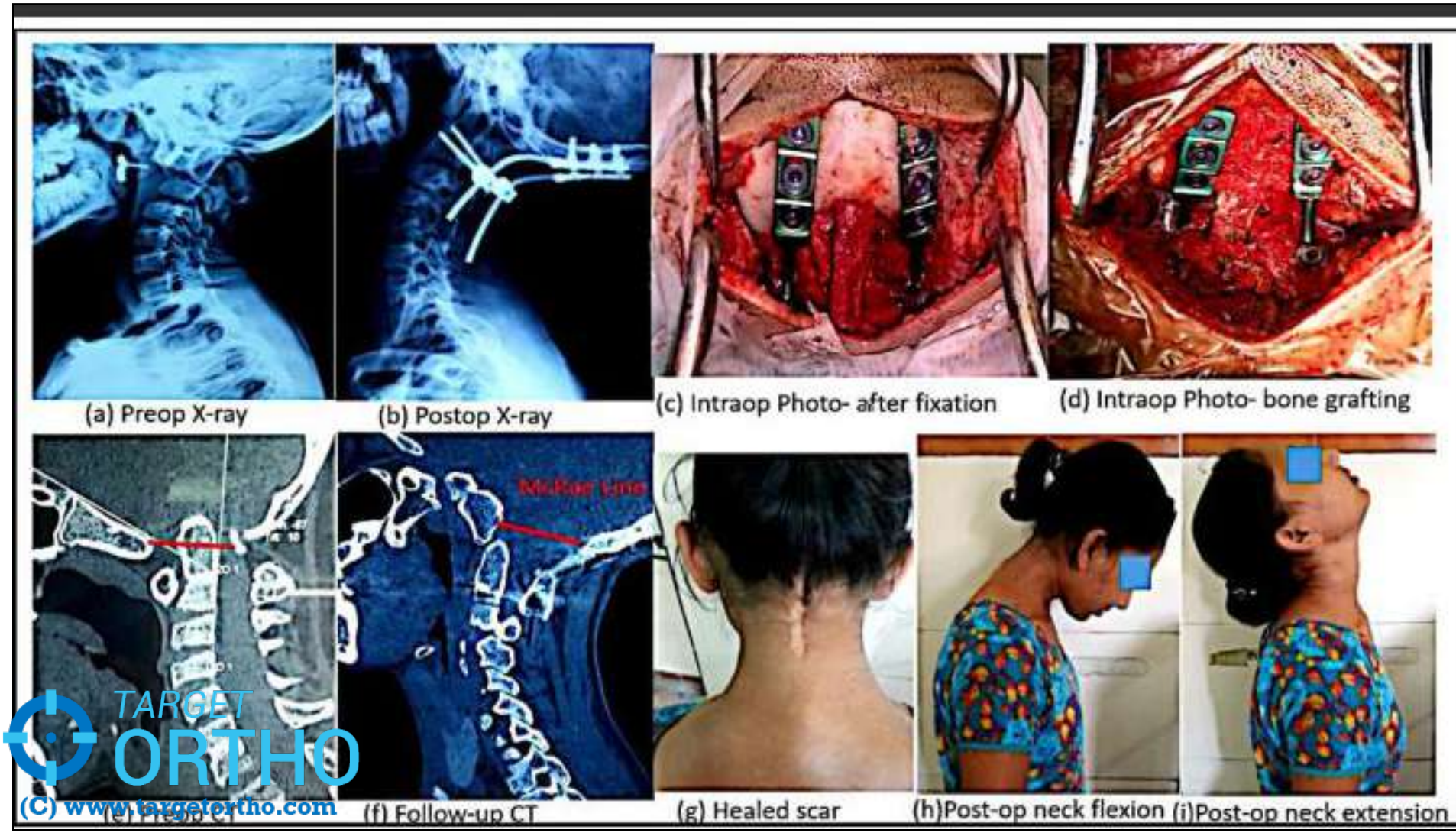
- Hydrocephalus(Obstruction)
- Impingement of lower cranial nerves (5,9,10,12)
- Vertebral artery compression- VBI

Treatment

- **Primary treatment** is surgical.
- If hydrocephalus is present- first to be addressed with VP shunt

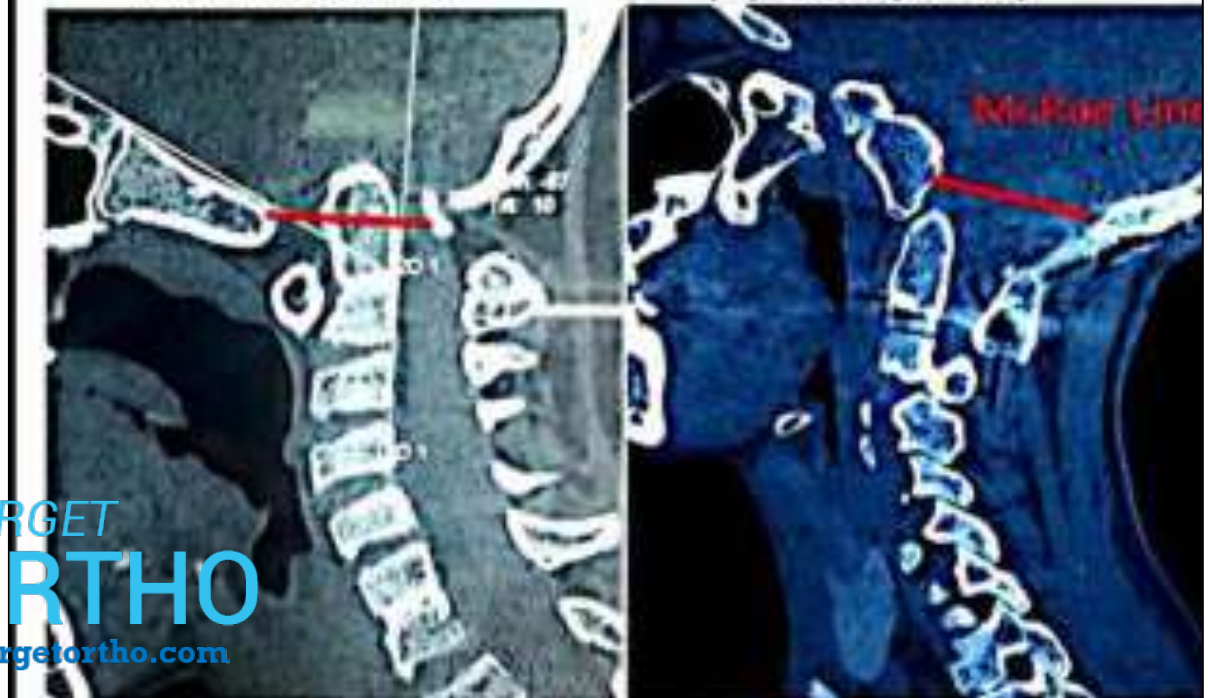
- **If the Oc region can be realigned** with traction- posterior decompression and occipito cervical instrumented fusion.

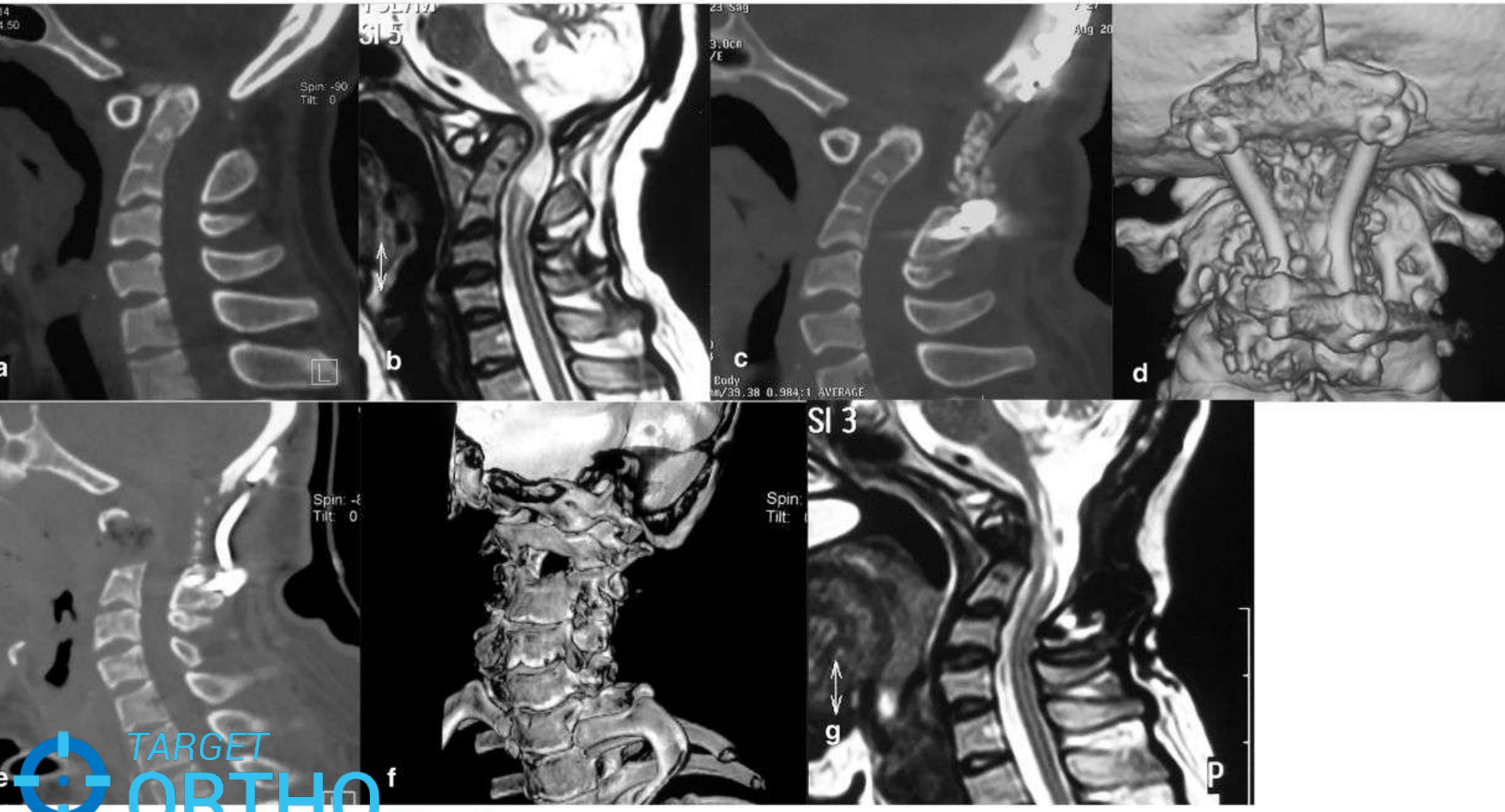
- **If not realigned-** the initial posterior surgery shoulkd be followed by anterior odantoidectomy and decompression.



(a) Preop X-ray

(b) Postop X-ray





ATLANTOOCCIPITAL FUSION

- Anterior arch of C1 is commonly assimilated to the occiput.
- **Hypoplastic ring posteriorly as well as condylar hypoplasia** --
  primary basilar impression
- The odontoid directed more posteriorly than normal
- Asso. with other anomalies: congenital fusion of C2 and C3.

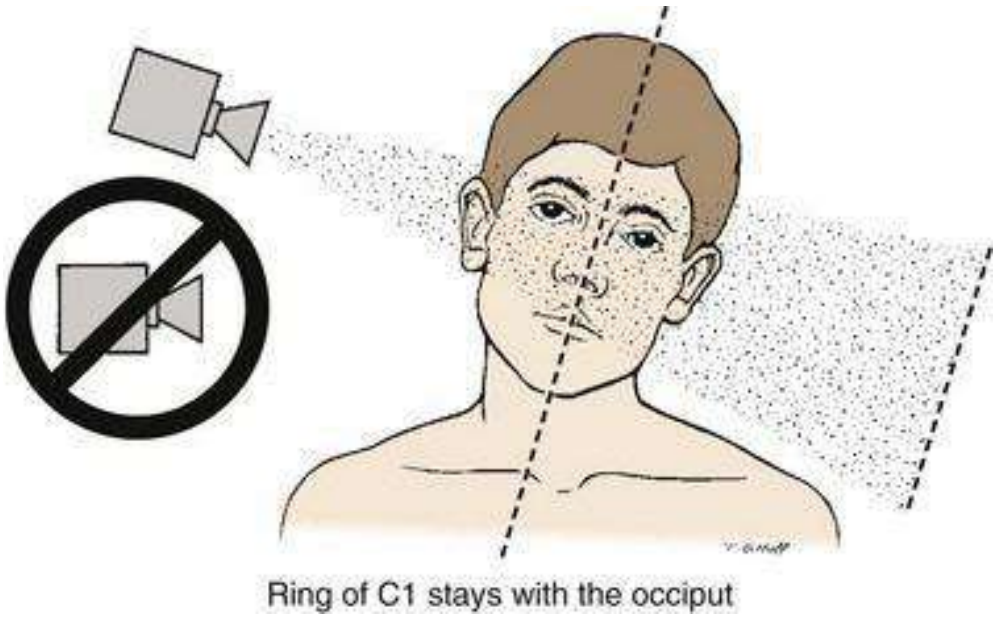
Clinical features

- Short, broad necks
- Restricted neck motion
- Low hairline
- High scapula and
- Torticollis
- Hemifacial microsomia
- Pagiocephaly.

- CT scan(**head-wag autotomography**).

- This technique involves side-to-side rotation of the child's head while a slow anteroposterior radiographic exposure of the upper cervical spine is performed.

- improved visualization of the occiputC1-C2 complex



Flexion-extension MRI

- Compression of the brain stem or upper cervical cord anteriorly occurs because of the backward-projecting odontoid.

- Compression from the posterior lip of the foramen magnum disturb the **posterior columns**

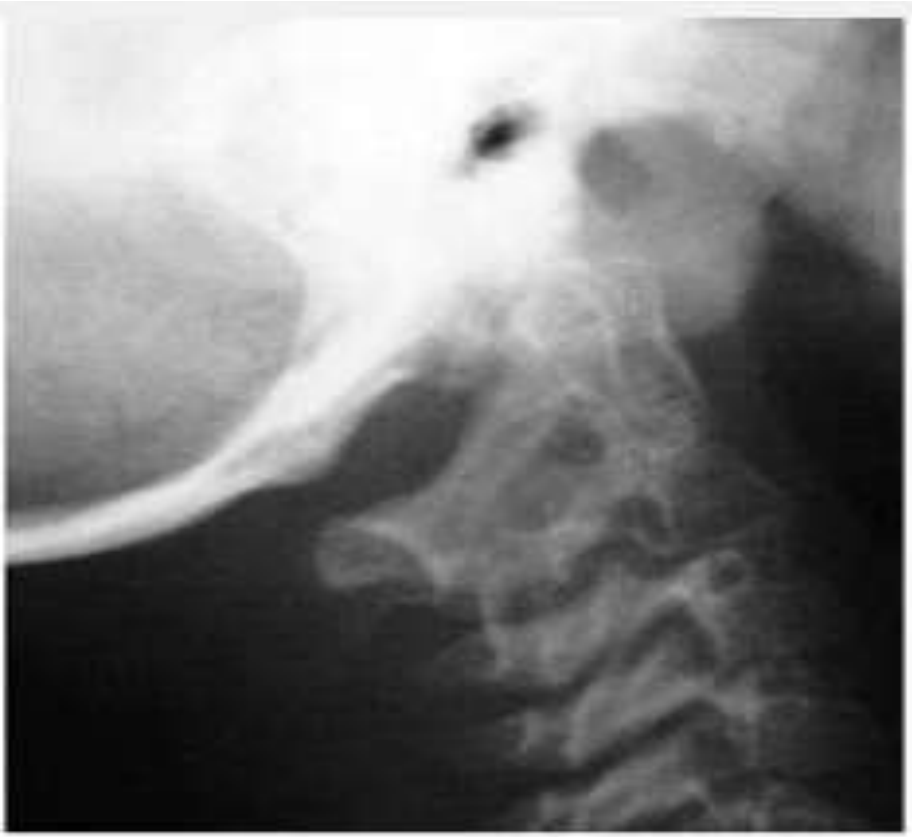
TREATMENT

C1-C2 instability are present

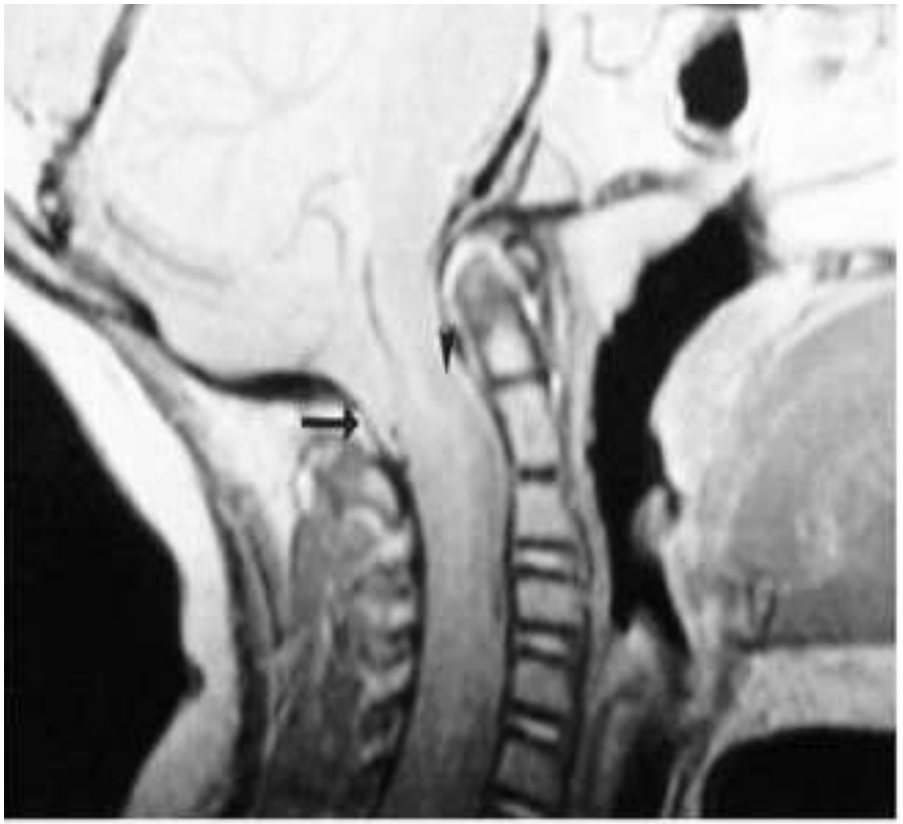
- Posterior C1-C2 fusion
- Preliminary traction
- Posterior signs and symptoms may be an indication for posterior decompression depending on the evidence of dural or osseous compression.

C1-C2 insatbility absent

- only compressive pathology, posterior fusion not recommanded
- if decompression (whether anterior or posterior) could lead to a destabilized spine, then concomitant posterior fusion should be considered



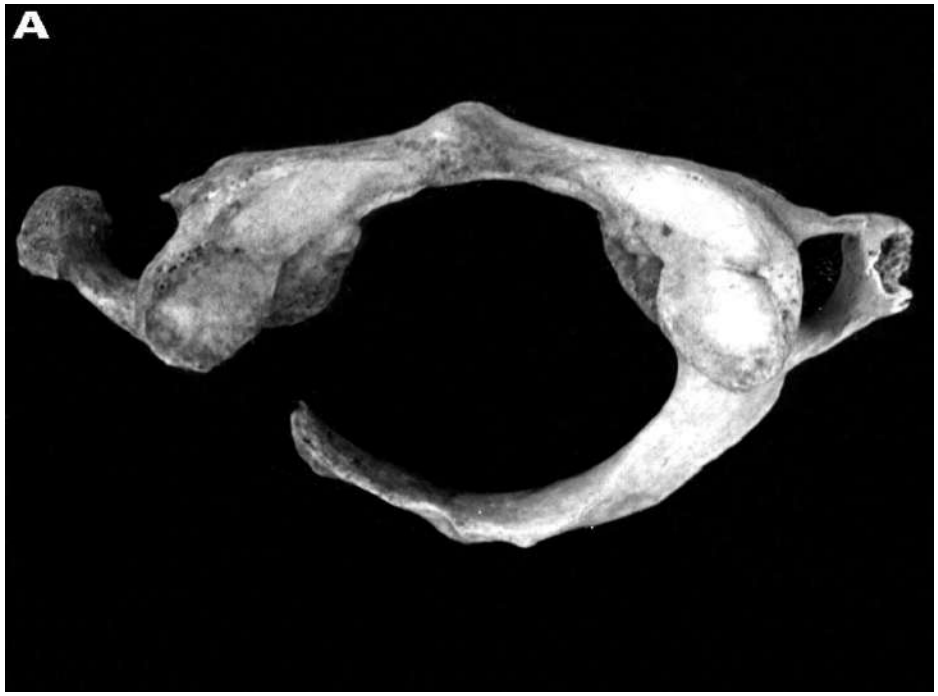
Plain lateral radiograph shows fusion of C2 and C3 and absence of the ring of C1 with occipitalization

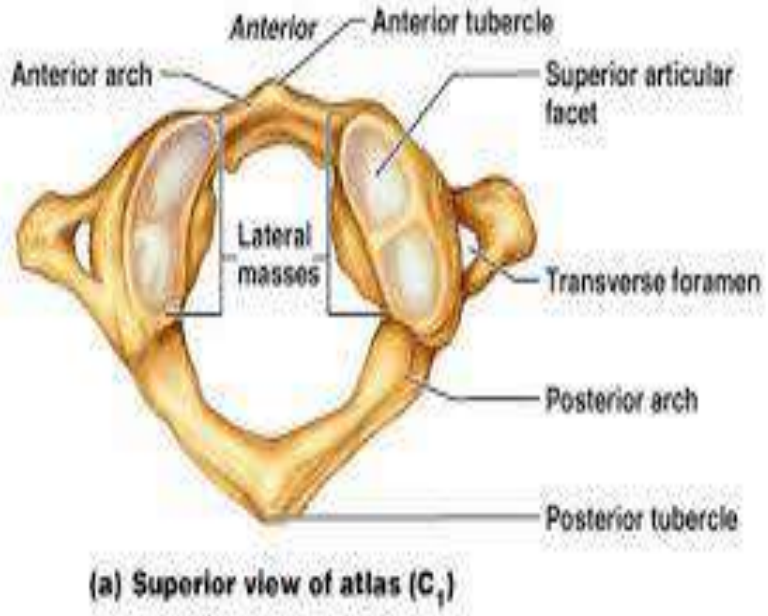


Magnetic resonance image (MRI) shows an Arnold-Chiari malformation, with herniation of the cerebellar tonsils into the foramen magnum (arrow).

UNILATERAL ABSENCE OF C1

- Present at birth OR when torticollis develops.
- A lateral translation of the head on the trunk, with variable degrees of lateral tilt and rotation
- The sternocleidomastoid muscle is not tight - **regional aplasia** of the muscles.
- Neck flexibility is variable and decreases with age.
- Painless.
- Neurologic signs (e.g., headache, vertigo, myelopathy) are present





(a) Superior view of atlas (C_1)

Doubousset classification

- Type I isolated hemiatlas

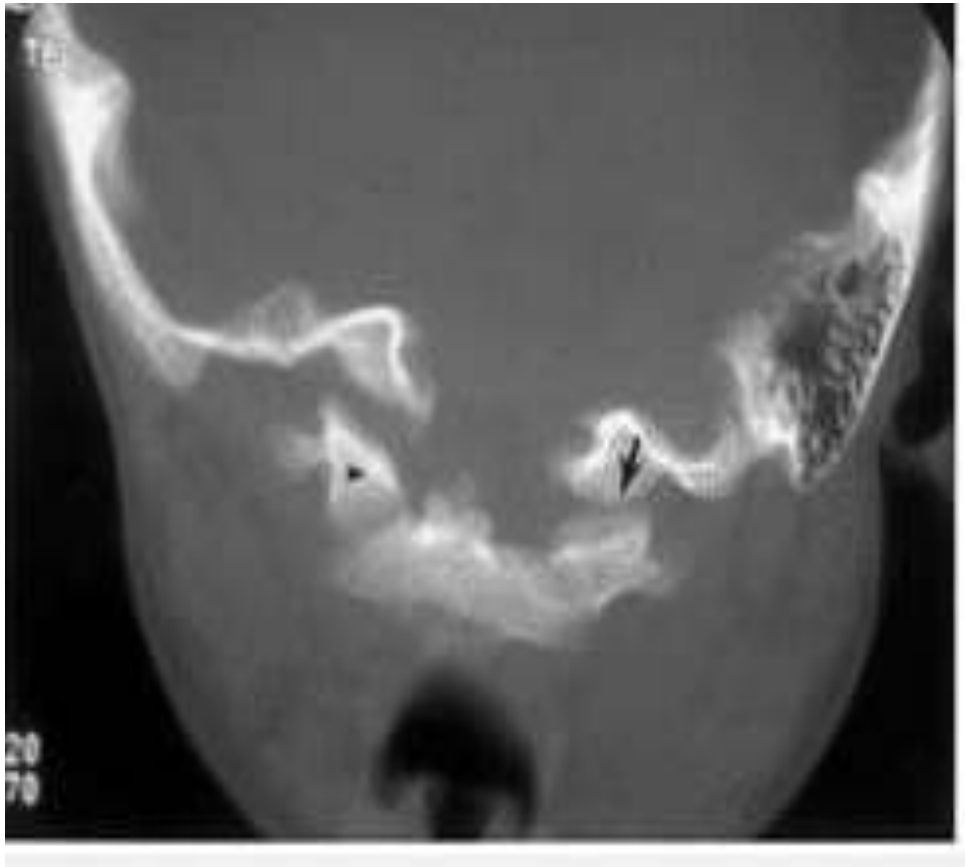
- Type II **partial or complete aplasia of one hemiatlas**

  other associated anomalies of the cervical spine
  e.g., fusion of C3-C4 and congenital bars

- Type III **partial or complete atlantooccipital fusion and symmetric or asymmetric hemiatlas** aplasia

  with or without anomalies of the odontoid and the lower cervical vertebrae

- Anteroposterior and lateral radiographs rarely give the diagnosis
- open-mouth odontoid view may suggest
- Tomograms or CT scans usually are needed in order to see the anomaly (**hypoplasia of the lateral mass to a complete hemiatlas)** with rotational instability and basilar impression.
- Occasionally the atlas is occipitalized.
- Angiography **arterial anomalies**
- MRI stenosis of the foramen magnum/Arnold-Chiari malformation



CT scan absent lateral mass of C1 (arrow) with a normal right lateral mass (arrowhead). This represents a Doubousset type II, C1 unilateral absence

Treatment

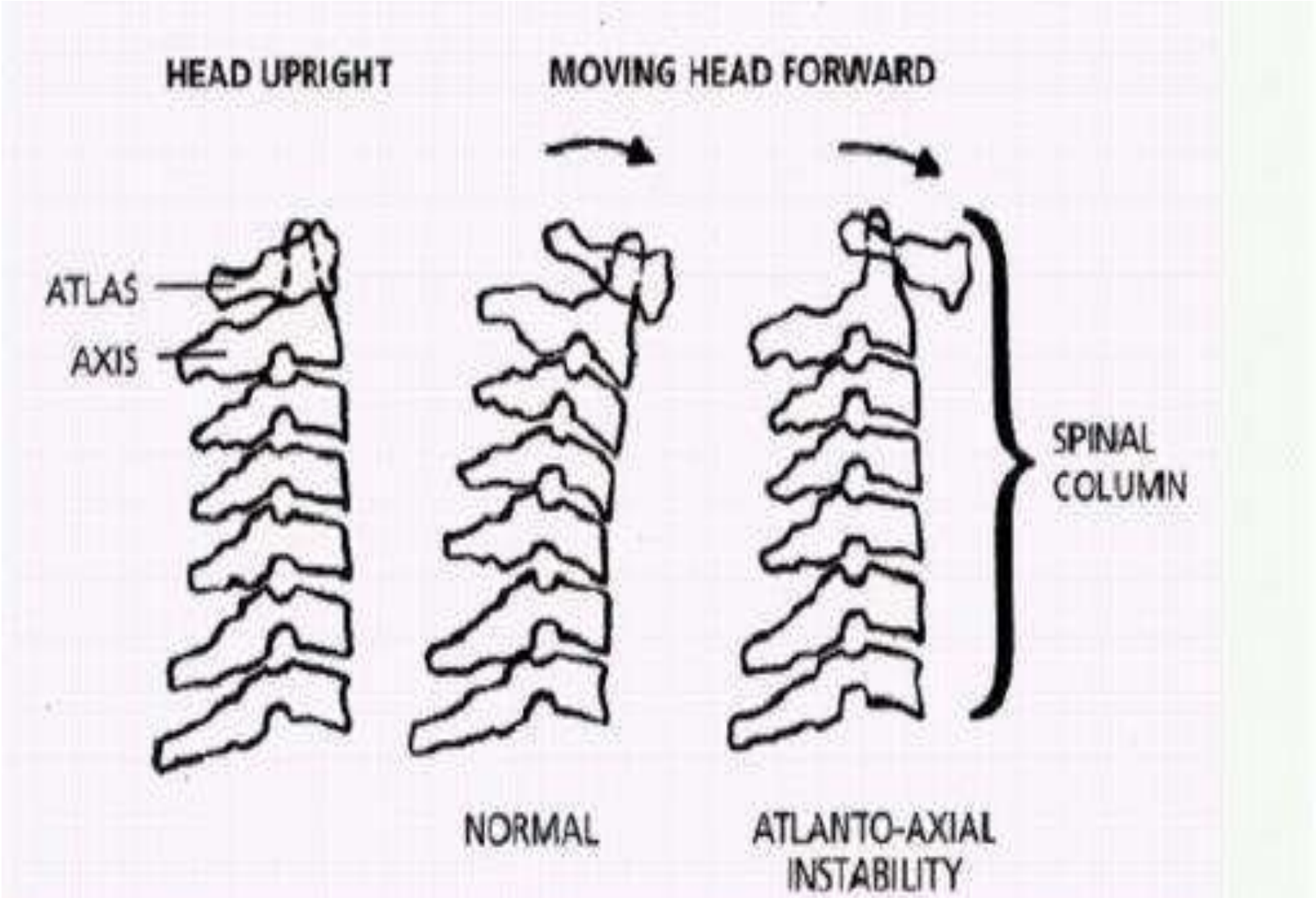
- Observation for progression
- Bracing does not halt progression of the deformity.
- Surgical intervention □severe deformities.
- A preoperative halo is used for gradual traction correction **over 6 to 8 days.**

- A posterior fusion from the occiput to C2 or C3

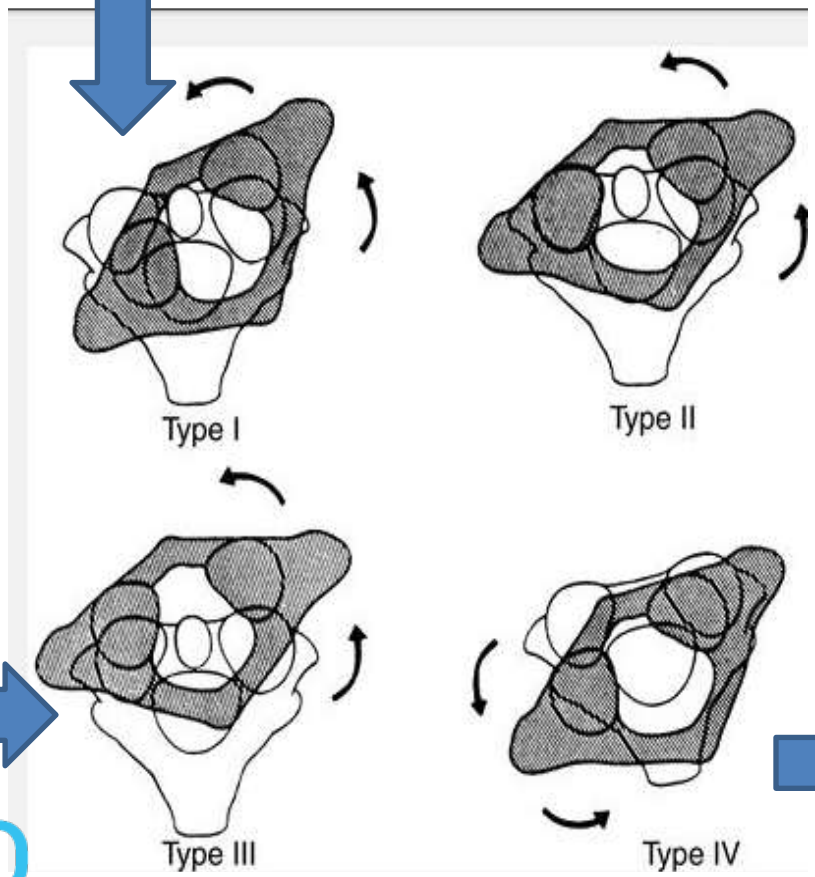
- The ideal age for posterior fusion is between **5 and 8 years**, corresponding to the age at which the canal size reaches adult proportions

Atlantoaxial Rotary Displacement

- **Most common** causes of childhood torticollis.
- Rotary displacements-- characteristically a pediatric problem--may occur in adults.
- If the deformity persists, the children present with a resistant and unresolving torticollis that is best termed **atlantoaxial rotary fixation or fixed atlantoaxial displacement**.
- Complete atlantoaxial rotary dislocation has rarely been reported in surviving patients



simple rotary displacement without an anterior shift (most common pediatric type –resolves



rotary displacement with an anterior shift of 5 mm or less, (more dangerous

rotary displacement with an anterior shift greater than 5 mm

rotary displacement with a posterior shift

- X ray the position of C1 and C2 in a child with subluxation appears to be the same as that in a normal child whose head is rotated.

- Open-mouth views are difficult to obtain and interpret, and the lack of co operation.

- The facet joints may be obscured because of apparent overlapping.

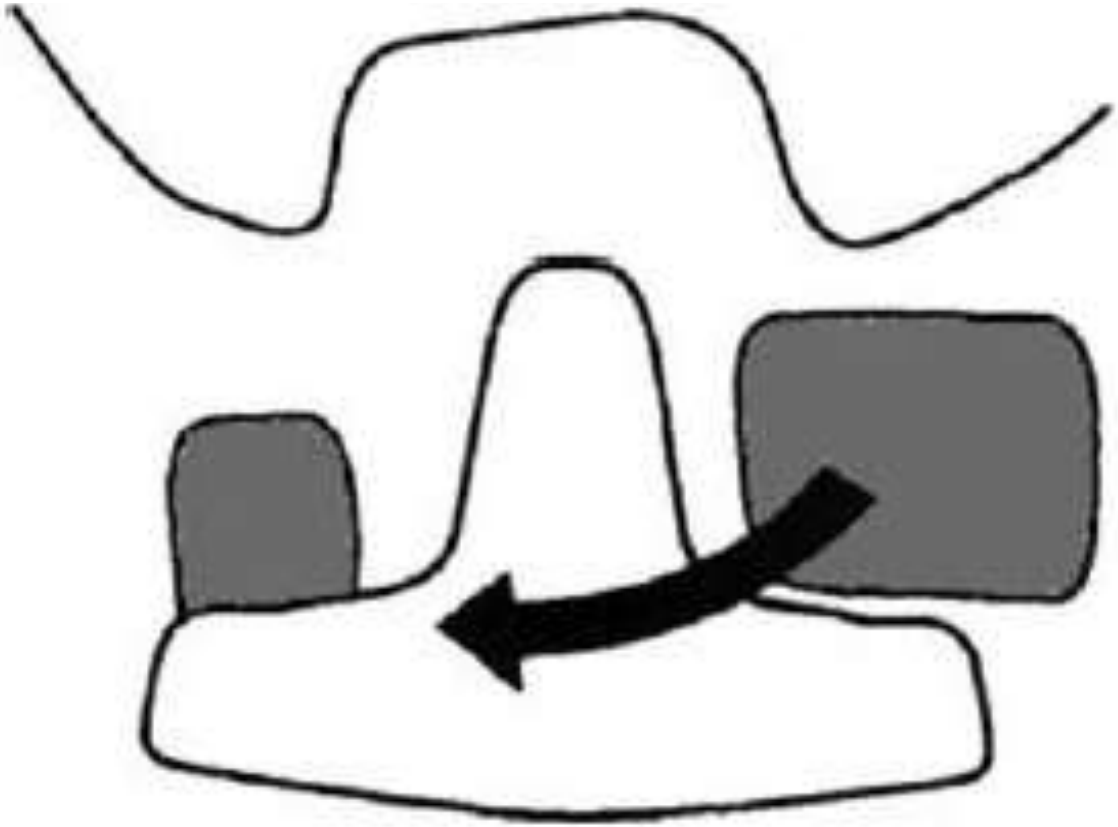
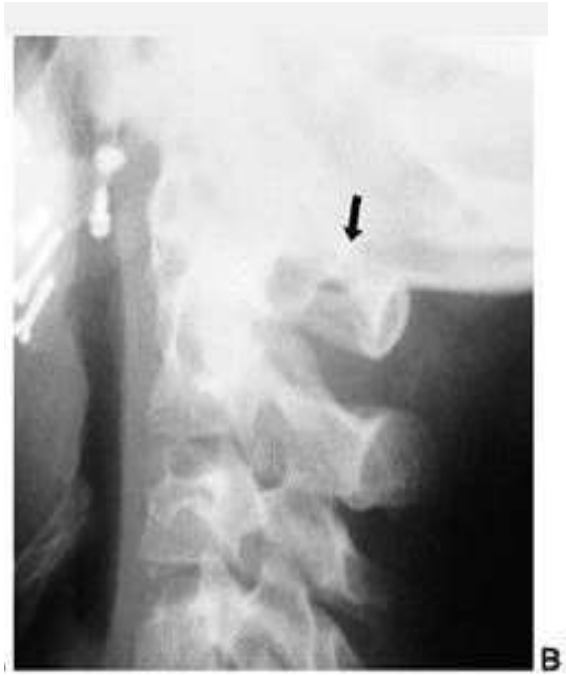
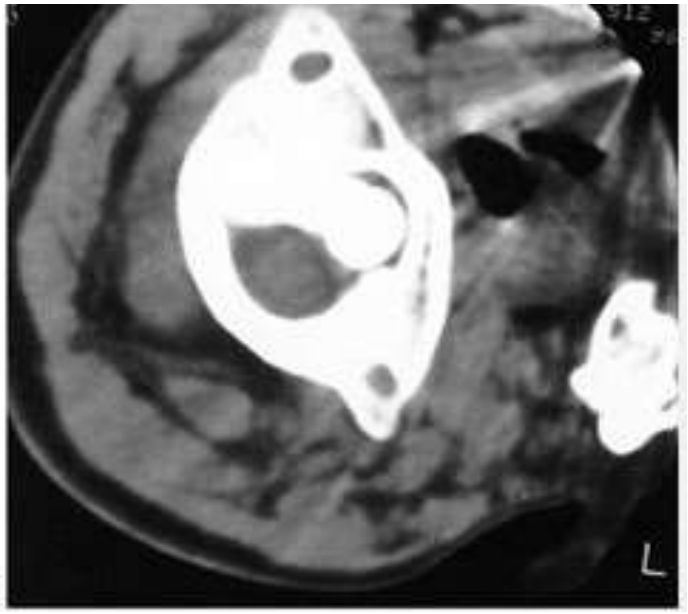
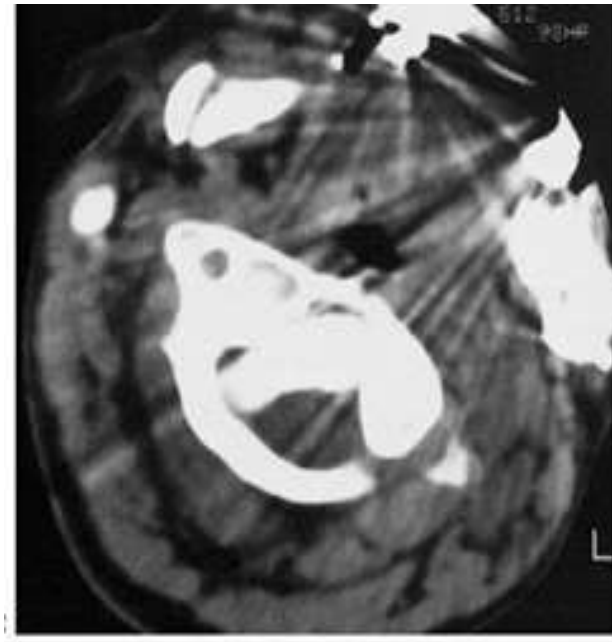


Fig. 7 Schematic anteroposterior view revealing the relationship of the lateral masses of C1 to the odontoid with fixed atlantoaxial displacement. The lateral mass rotating anteriorly appears broader, more cephalad and closer to the midline odontoid process (medial offset) than the lateral mass rotating posteriorly, which appears narrower and further from the odontoid (lateral offset)







The lateral cervical spinal radiograph. The posterior arches fail to superimpose because of the head tilt

Dynamic computerized tomography (CT) scans in fixed atlantoaxial rotary displacement, with the head **maximally rotated to the left** The ring of C1 is still in the exact relation to the odontoid indicating a fixed displacement

Her head **maximally rotated to the right**, in this case, does not reach the midline.

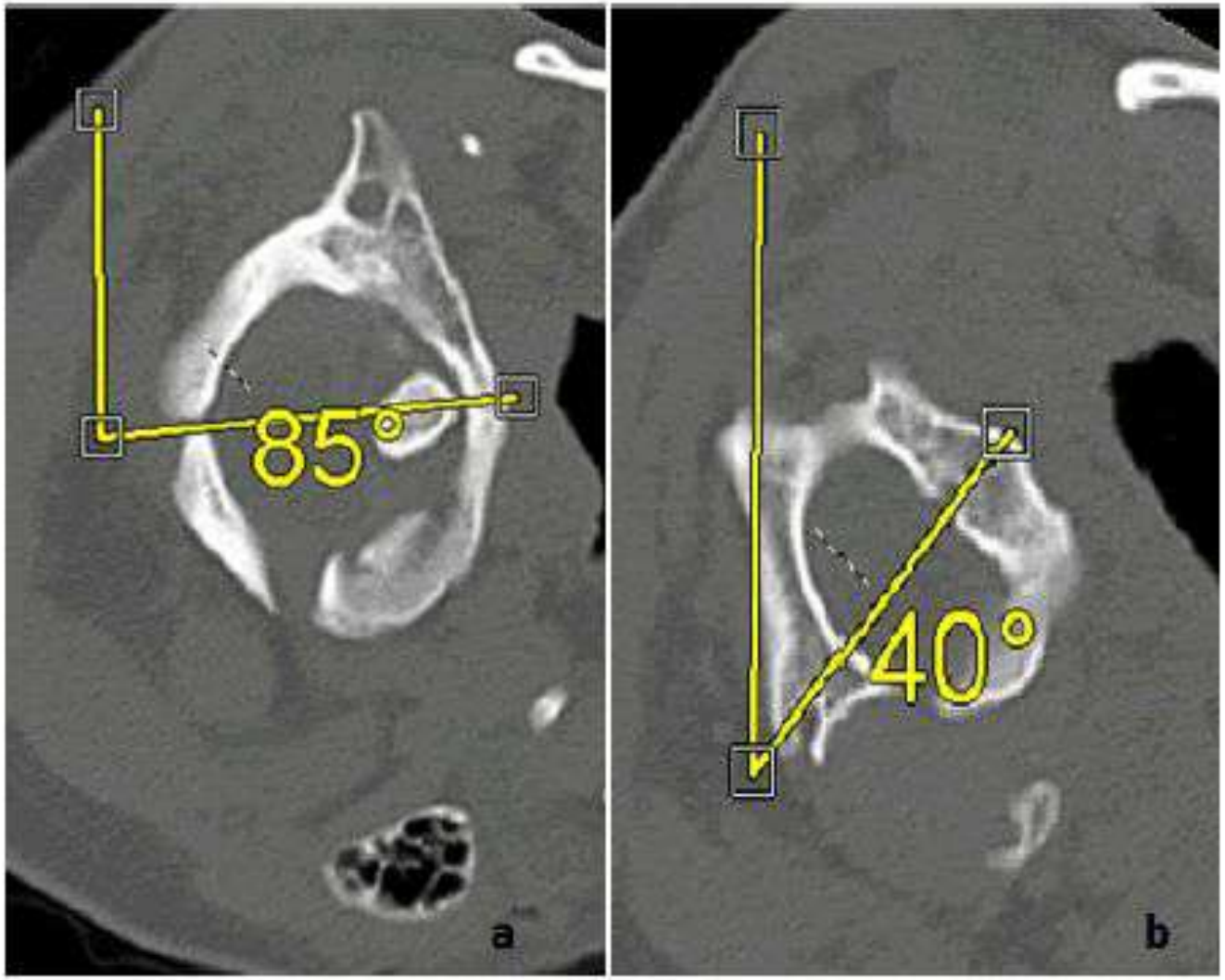


Figure 2: Excessive (>41°) rotation between C1 and C2. Axial CT views through (a) C1 and (b) C2 on full neck rotation demonstrate that the angle between C1 and C2 is (85°-40°) = 45°, and therefore unstable.

GRISEL SYNDROME

- Spontaneous atlantoaxial subluxation with inflammation of adjacent neck tissues.

- Children after upper respiratory tract infections .
- frequently febrile

- Direct connection exists between the pharyngovertebral veins and the periodontal venous plexus and suboccipital epidural sinuses □ hematogenous transport of peripharyngeal septic exudates to the upper cervical spine

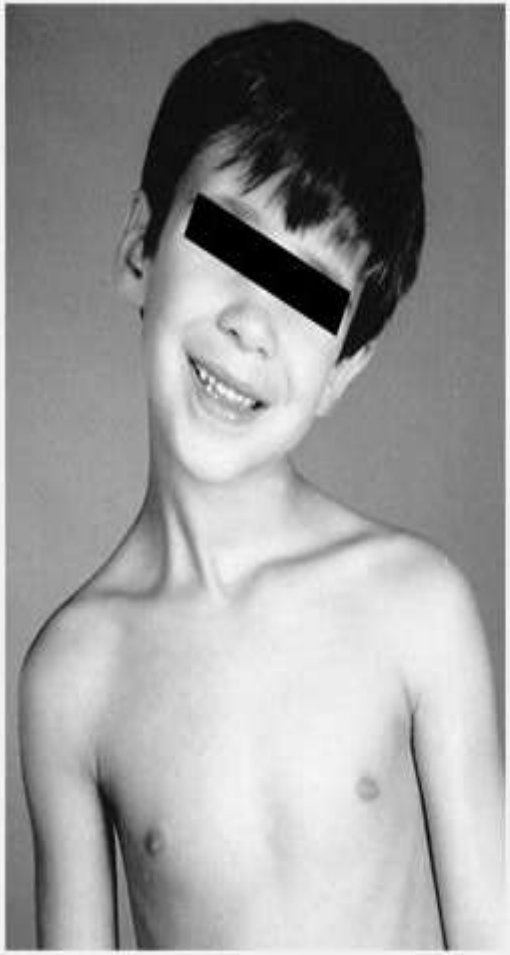


Figure 22.13 A 5-year-old boy developed an atlantoaxial rotary subluxation after an upper respiratory viral infection (Grisel syndrome). It rapidly resolved after treatment with a soft collar and mild doses of diazepam.

Treatment

- Most resolve spontaneously.
- The pain subsides and the torticollis becomes fixed.

- **less than 1 week's** duration immobilization in a soft cervical collar and rest for approximately 1 week.

- **more than 1 week but less than 1 month** immediately for cervical traction, relaxants, and analgesics.
  - If no anterior displacement is noted after reduction, cervical support should be continued only so long as symptoms persist.
  - If there is anterior displacement, immobilization should be continued for 6 weeks to allow ligamentous healing to occur.

- **more than 1 month**, cervical traction (usually halo skeletal) can be tried for up to 3 weeks, but the prognosis is guarded.

two groups:

A, those whose rotary subluxation can be reduced with halo traction but, resubluxate when the immobilization is stopped

B, those whose subluxation cannot be reduced and is fixed.

- When the deformity is fixed, especially when anterior C1 displacement is present, the transverse atlantal ligament **posterior C1-C2 fusion should be performed**

- Indications for fusion
  - Neurologic involvement
  - Anterior displacement
  - Failure to achieve and maintain correction
  - Deformity that has been present for **more than 3 months**
  - Recurrence of deformity following an adequate trial of conservative management (**at least 6 weeks of immobilization after reduction**).

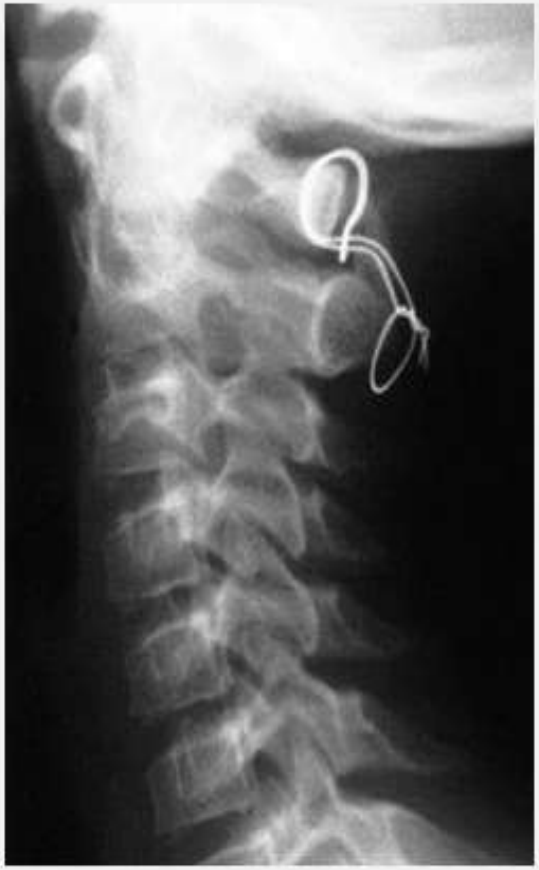


Figure 22.14 The child in Figure 22.11 had a fixed deformity that occurred 6 months earlier, immediately after reconstructive maxillofacial surgery for Goldenhar syndrome. It did not respond to traction, including halo traction. She underwent a posterior C1-C2 (Gallie-type) fusion. A solid fusion was present 9 months later; clinically, the patient achieved 80 degrees of rotation to the left and 45 degrees of rotation to the right.

Treatment Plan for Rotatory Subluxation (Phillips and Hensinger)

Present <1 week: immobilization in soft collar, analgesics, bed rest for 1 week; if no spontaneous reduction: hospitalization, traction

Present >1 week but <1 month: hospitalization, cervical traction (head-halter), cervical collar 4-6 weeks

Present >1 month: hospitalization, cervical traction (skeletal), cervical collar 4-6 weeks

Nonoperative treatment is used only if radiograph shows no significant anterior displacement or instability

CONGENITAL TORTICOLIS

- Congenital muscular torticollis, or congenital wry neck
- Common cause of torticollis infant and young child
- contracture of the sternocleidomastoid muscle, with the head involved side and the chin opposite shoulder.
- Cause unknown.

- Because of the birth history, one theory is that a compartment syndrome occurs due to soft tissue compression of the neck at the time of delivery

- utero crowding

- histopathologic evidence of denervation and reinnervation

- venous occlusion of the sternocleidomastoid muscle .
  - This occlusion may result in a compartment syndrome, as manifested by edema, degeneration of muscle fibers, and muscle fibrosis.

CAUSES

- Sternocleidomastoid tumor (43%)
- muscular torticollis (31%)
- postural torticollis (22%)

- Depend on the age of the child (first 6 to 8 weeks of life).
- during the first 4 weeks of life, a mass or tumor may be palpable in the neck (nontender, soft)maximum size within the first 4 weeks of life then gradually regresses.

- After 4 to 6 months of life the contracture and the torticollis are the only clinical findings.

INVESTIGATIONS

- C SPINE xray normal, aside from the head tilt and rotation
- MRI demonstrate abnormal signals in the sternocleidomastoid muscle, but no discrete masses within the muscle.
- The muscle diameter is two to four times greater than that of the contralateral muscle.
- In older patients the signals are consistent with atrophy and fibrosis, similar to those encountered in compartment syndromes of the leg and forearm

Treatment

- Initially consists of conservative measures.
- stretching exercises alone (simple muscle torticollis sternocleidomastoid tumor)
- The extent of sternocleidomastoid fibrosis on ultrasound examination is also predictive of the need for surgery.

- TIMING of SX after 1year. Best (1-4)

- The child's neck and anatomic structures are larger, making surgery easier.

- Established facial deformity or a limitation of more than 30 degrees of motion usually precludes a good result, and surgery is required to prevent further facial flattening and further cosmetic deterioration.

- Surgical treatments include a unipolar release at the sternoclavicular or mastoid pole

- bipolar release, middle third transection, and even complete resection.

- Bipolar release combined with a Z-plasty of the sternal attachment yielded 92% satisfactory results in one series, whereas only 15% satisfactory results were obtained with other procedures.

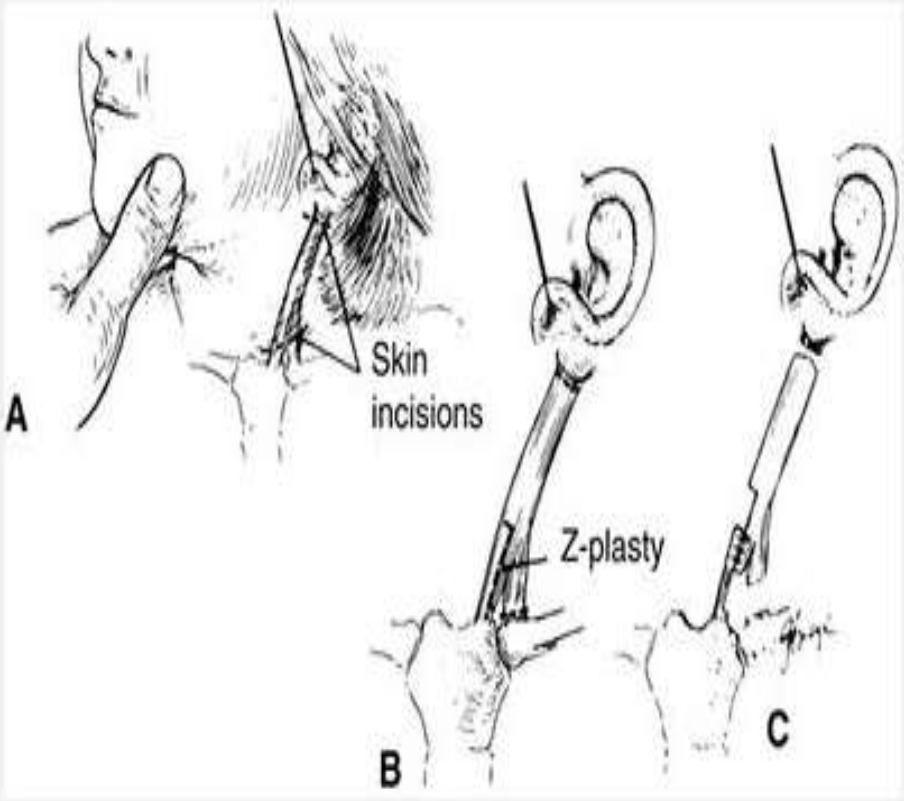
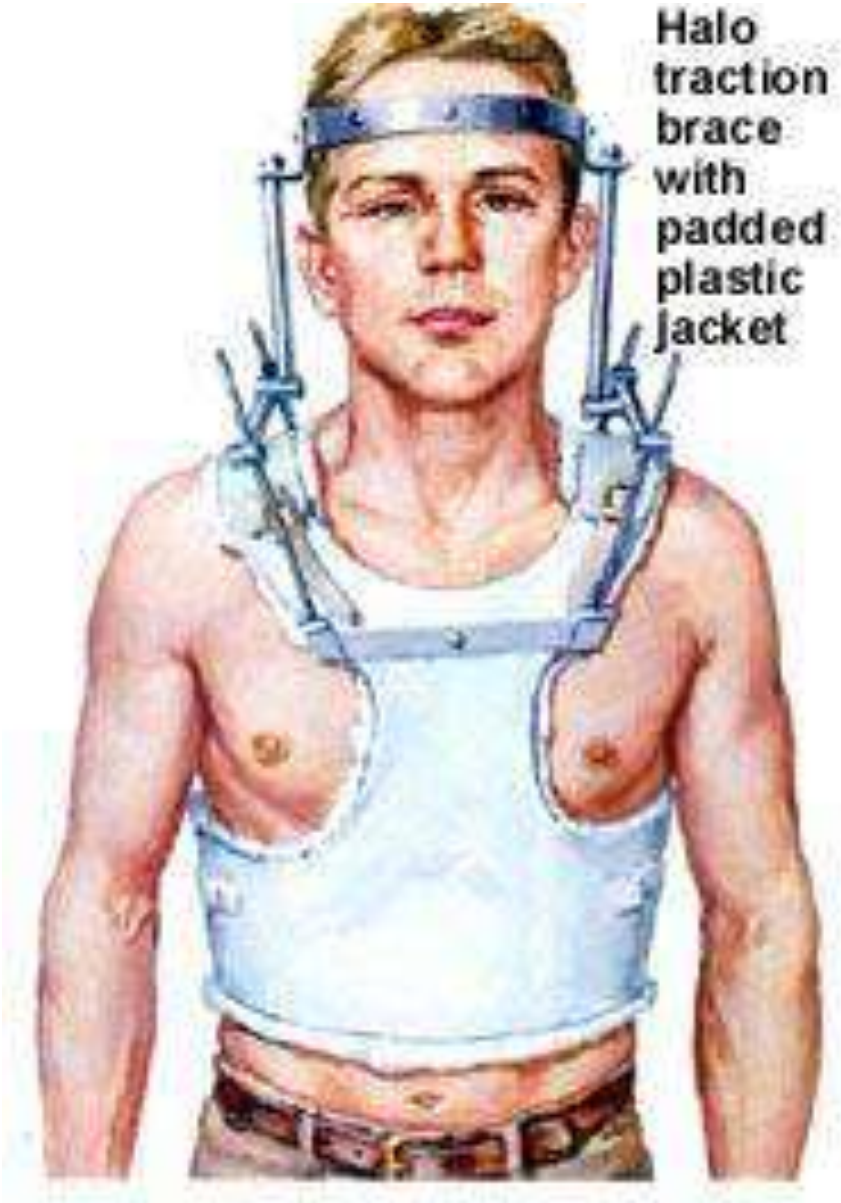


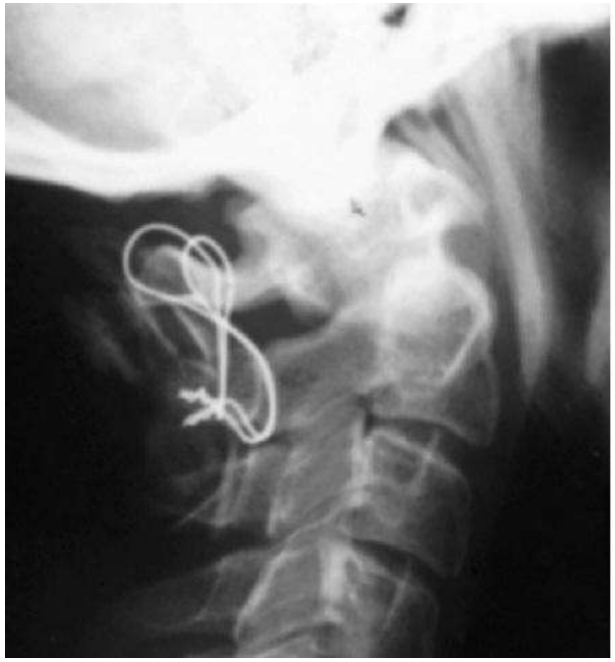
Figure 22.15 The Z-plasty procedure for torticollis. **A:** The location of the skin incisions. **B:** The clavicular and mastoid attachments of the sternocleidomastoid muscle are cut, and a Z-plasty is performed. Note that the medial aspect of the sternal attachment is preserved. **C:** The completed procedure after release of the proximal muscle insertion. (From Ferkel RD, Westin GW, Dawson EG, et al. Muscular torticollis. A modified surgical approach. *J Bone Joint Surg Am* 1983;65-A:894â€"900, with permission.)

Fixation methods for pediatric C1-C2

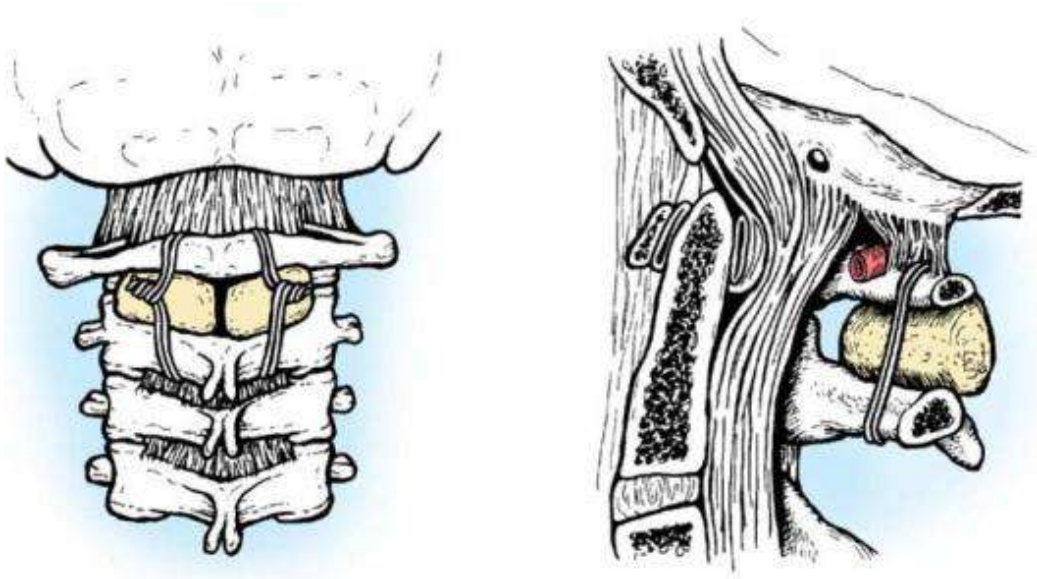
- Atlas reaches - Internal diameter - 6 to 7 yrs of age.
- Further growth- periosteal appositional growth.
- **Surgical fusion should not be performed** before this age because of the potential for later cervical stenosis



Traditional methods of fusion



Gallie

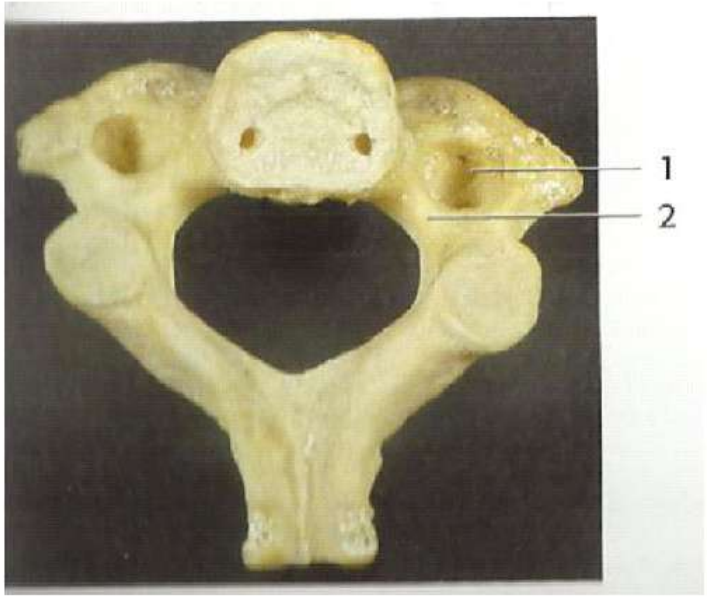


Brooks and Jenkins

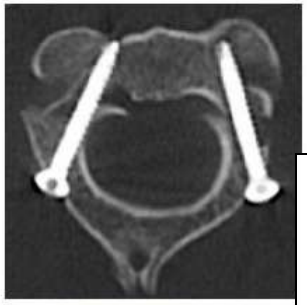
- Sublaminar wires
- Less rigid
- Lamina is the anchor
- Needs post op bracing

Anchorage Points

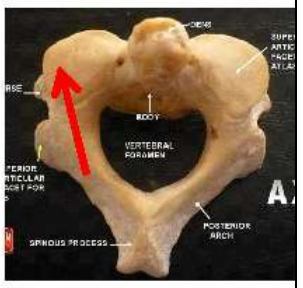
Axis



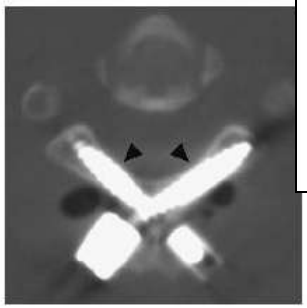
- Pedicle

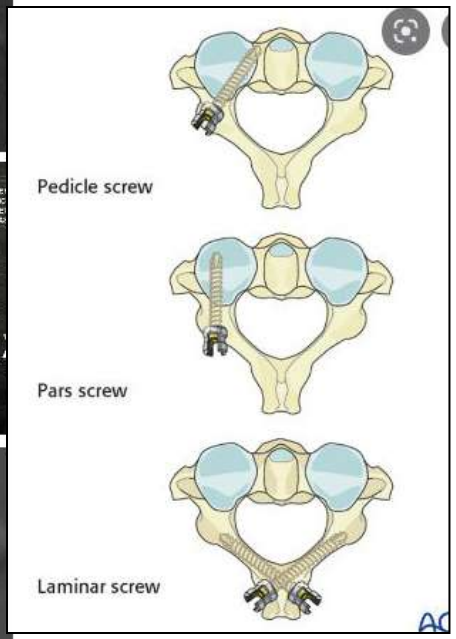


- Pars

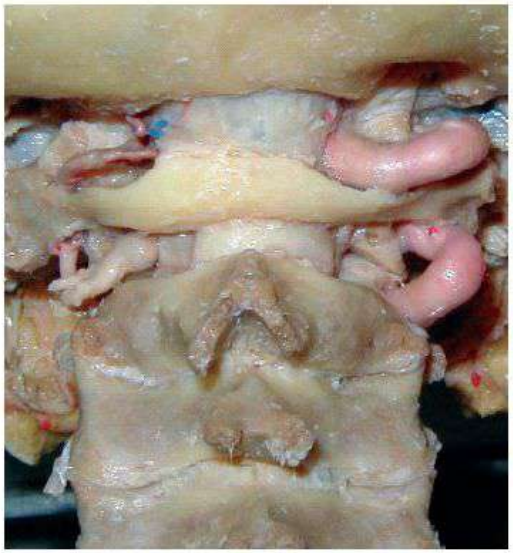


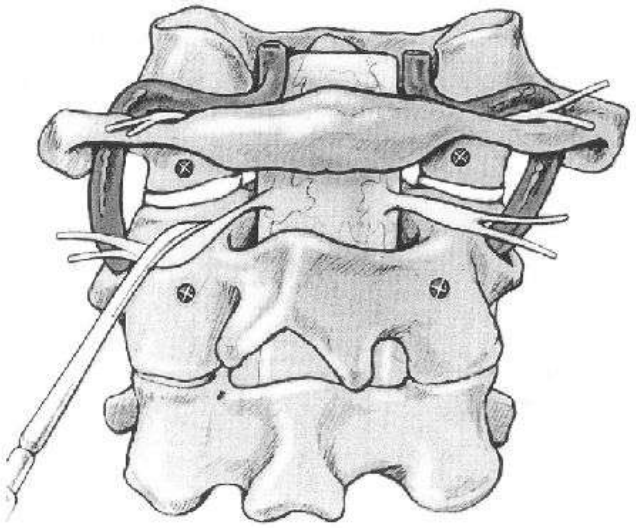
- Translaminar

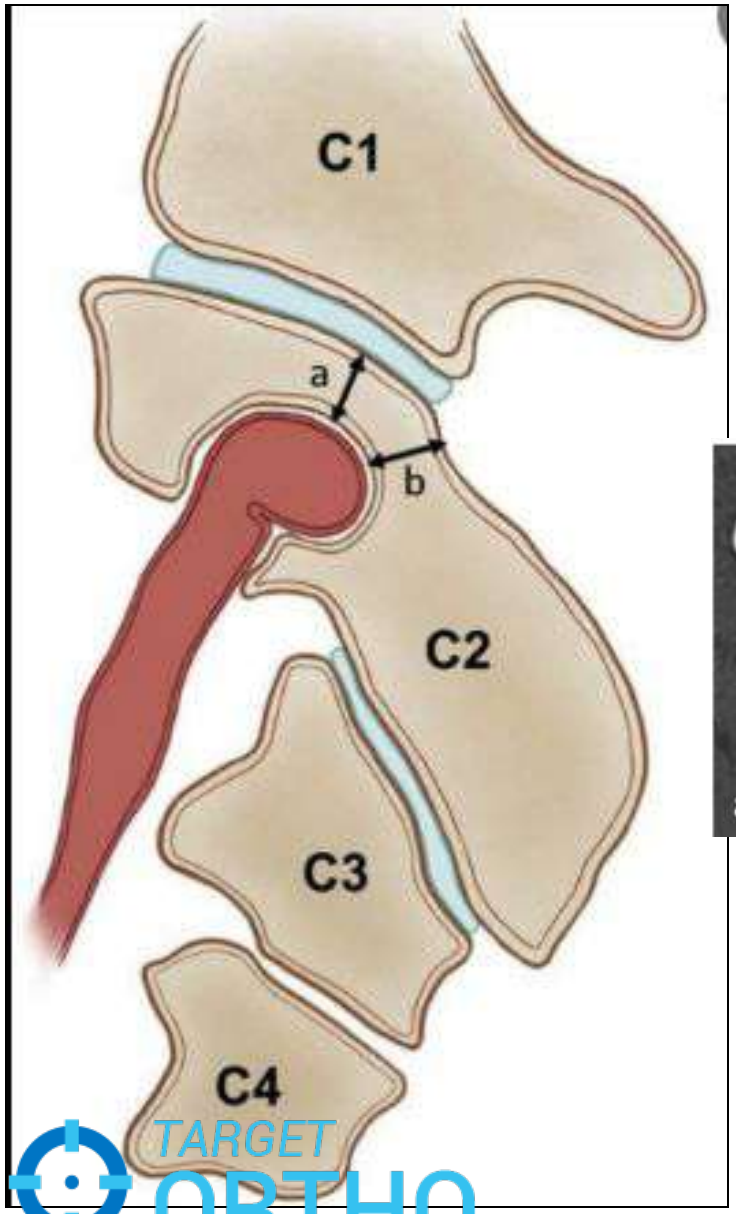


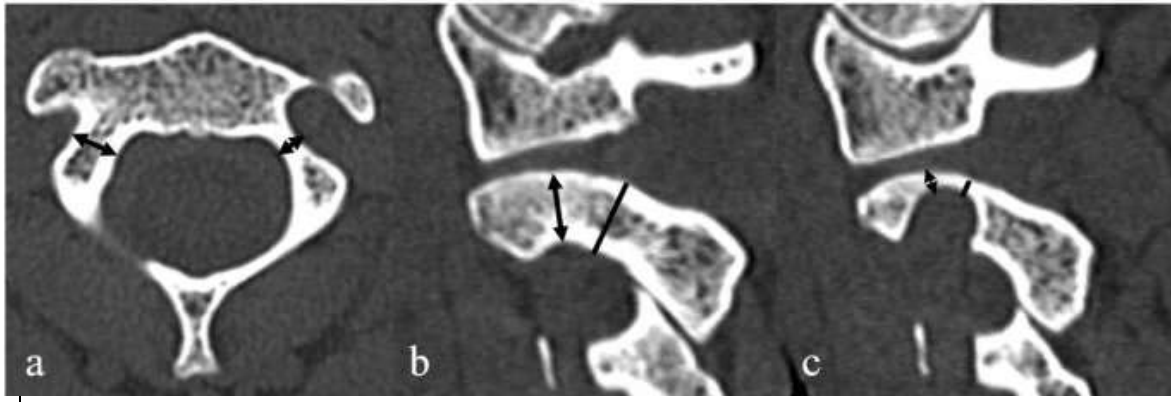


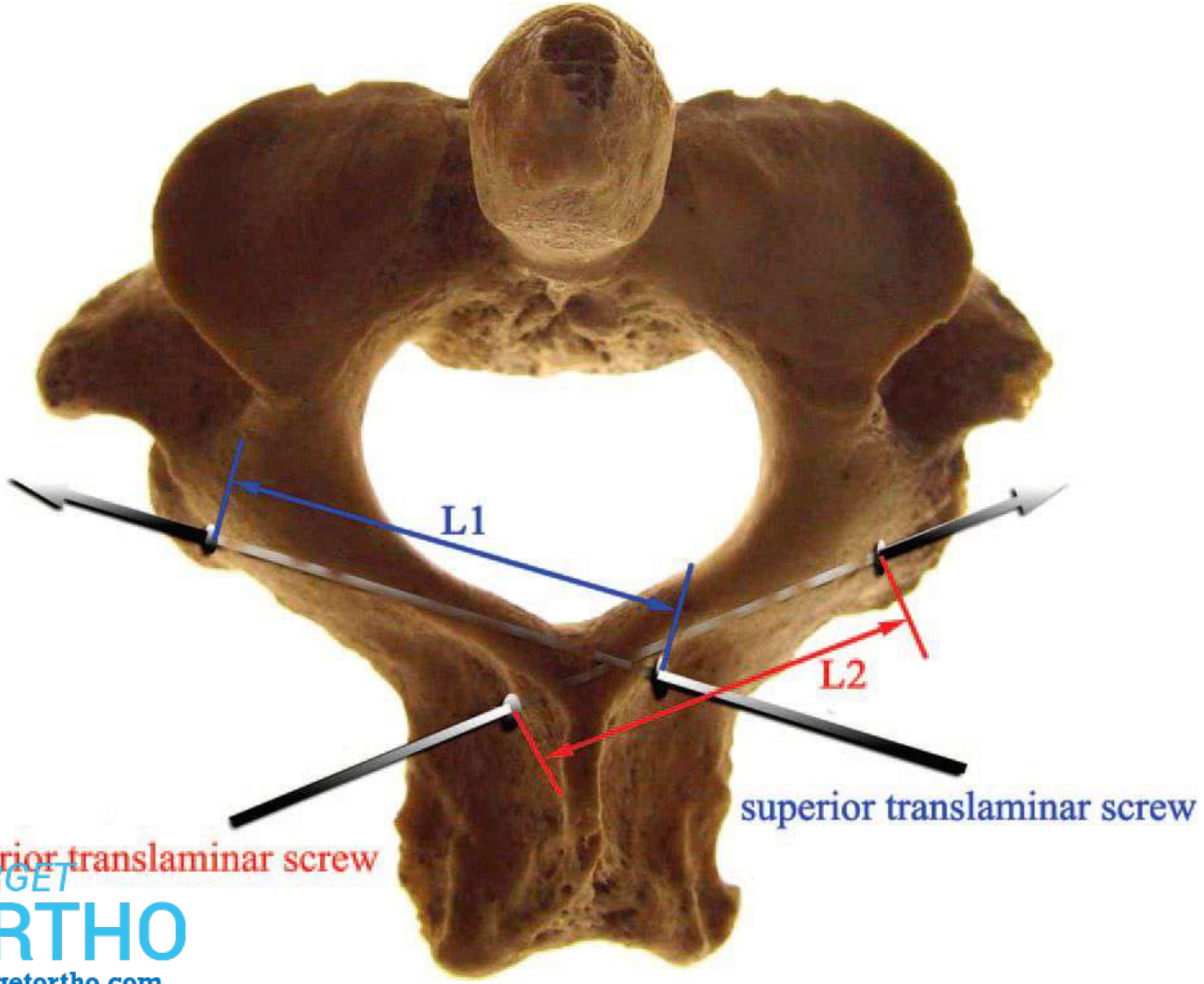
Vertebral artery
C1,2 nerve roots and ganglia

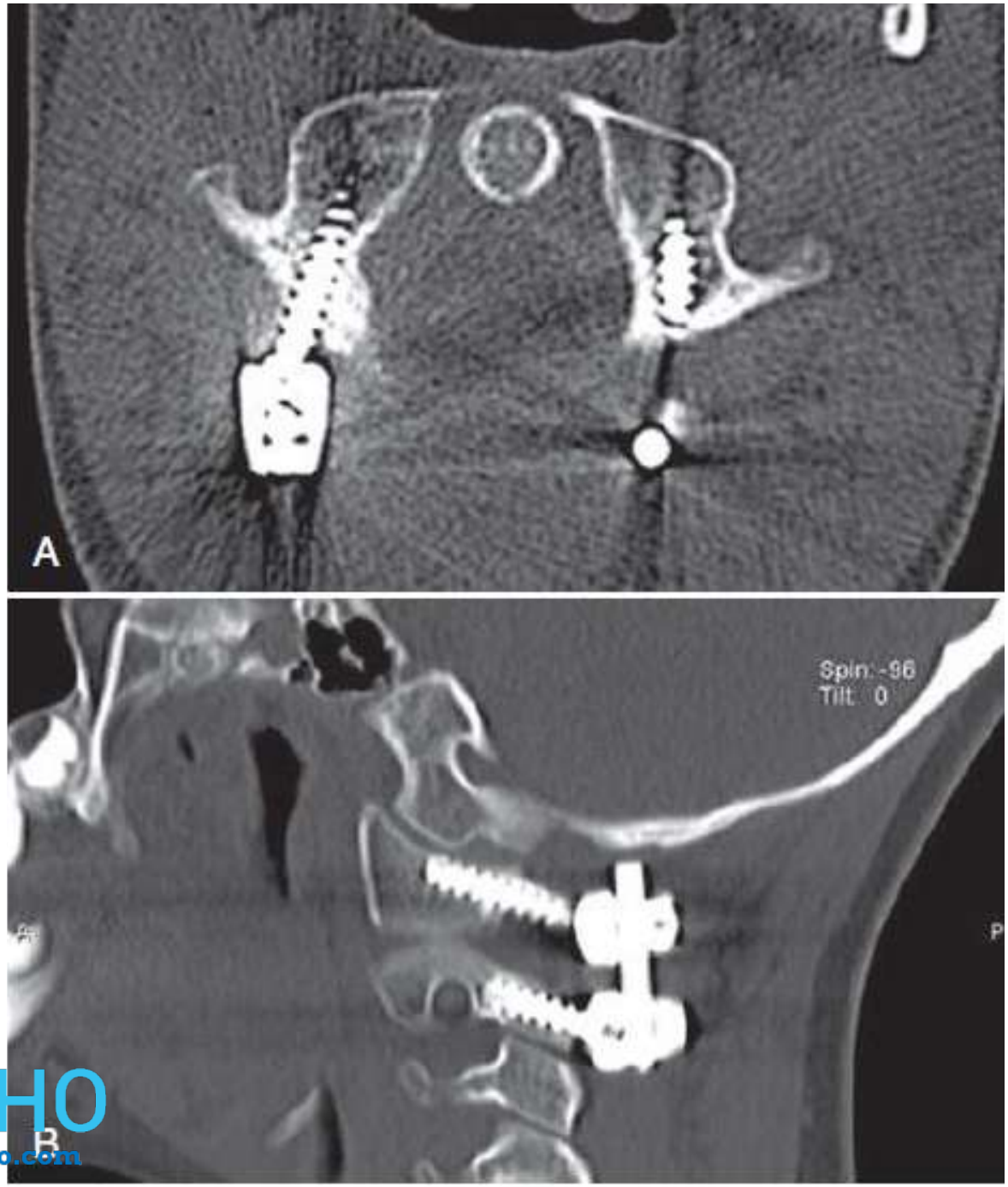




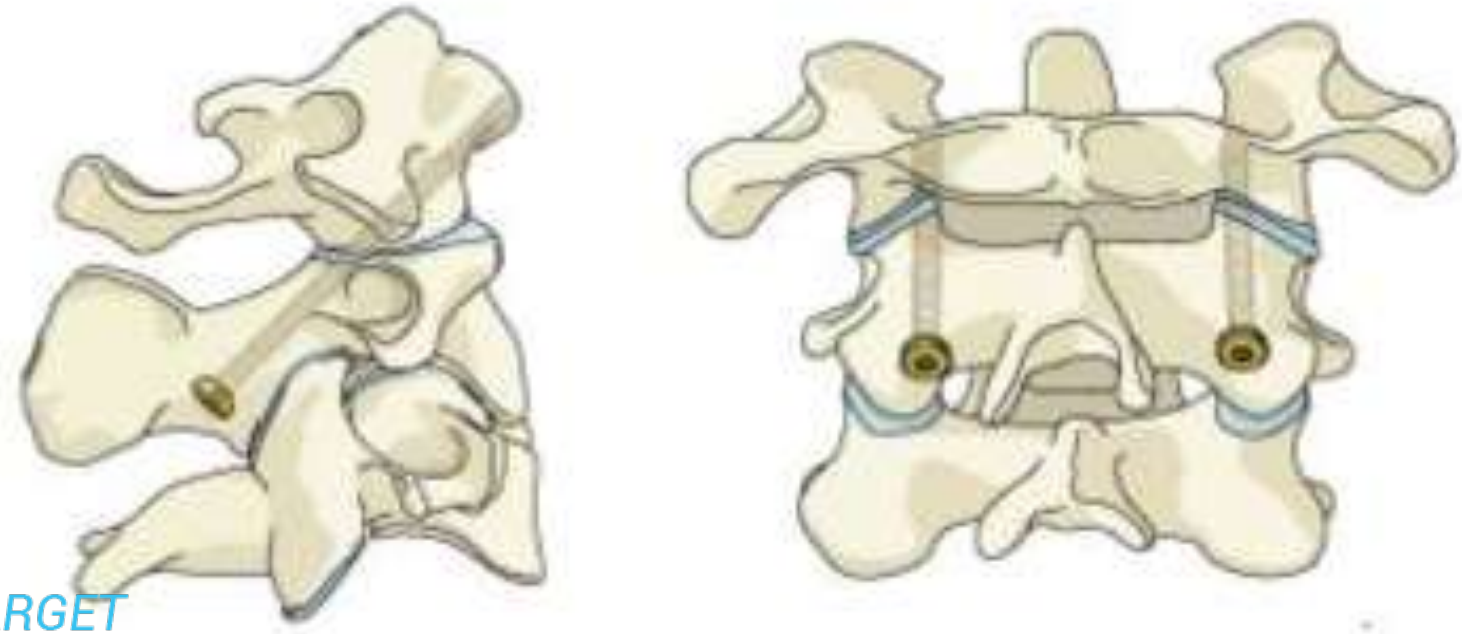


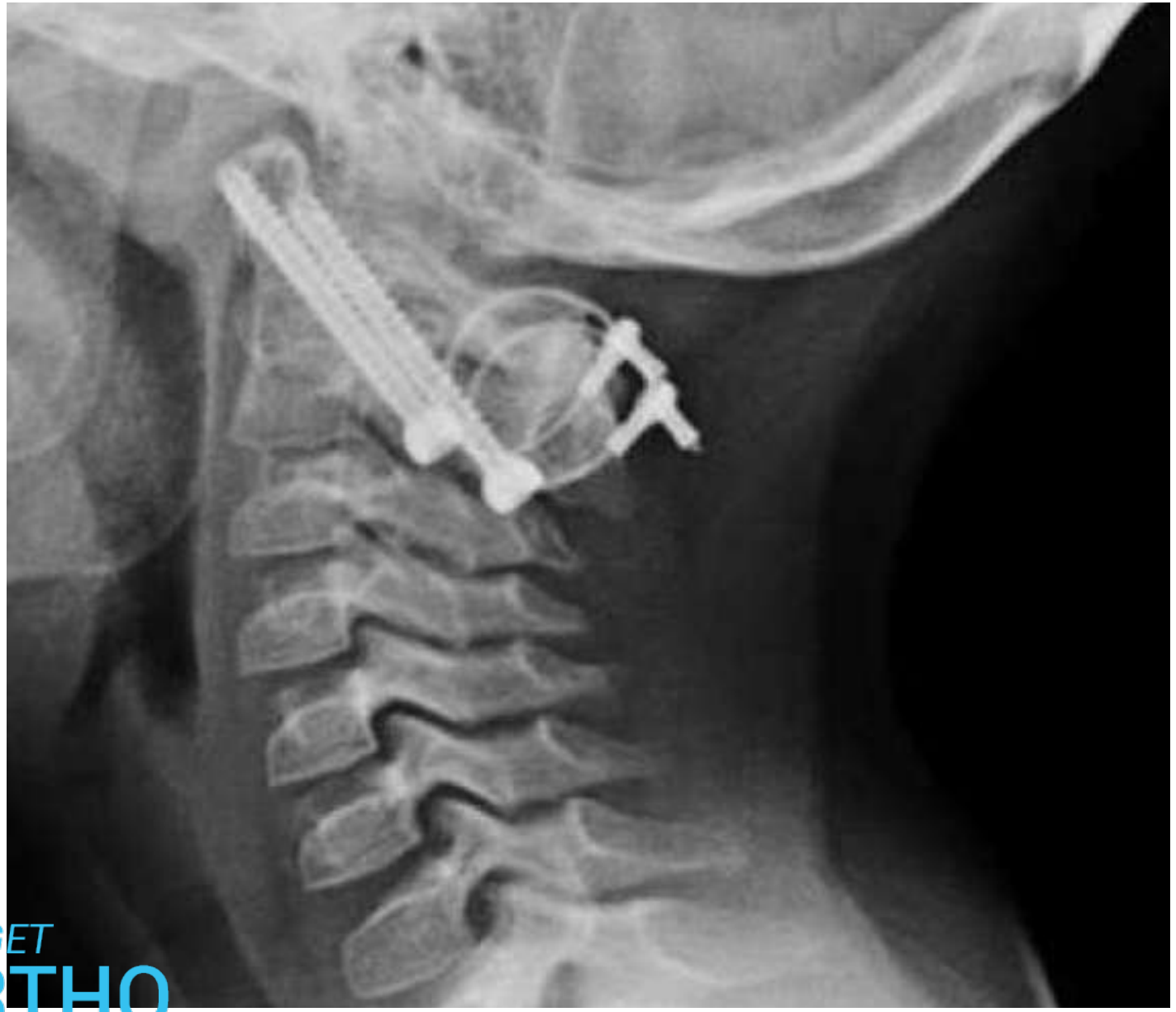






Magerl





THANK YOU