

Bridging defects-Radio-anatomical review of cranio-vertebral junction anomalies

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Learning objectives

- To discuss the normal and the variant anatomy involving the CVJ region and the various craniometric measurements used for the radiologic assessment of region.
- To discuss various congenital and acquired anomalies involving CVJ.
- Radiological workup of the craniovertebral anomalies.

Background

Anatomy of CVJ:

Articular anatomy: upper surfaces of C1 lateral masses are cup like or concave which fit in to the ball and socket configuration, united by articular capsules surrounding the atlanto-occipital joint & by anterior & posterior atlanto-occipital membranes.

Atlas and axis- four synovial joints between atlas and axis

- two median- front and back of dens (pivot variety)
- two lateral-between opposing articular facets(plane variety), rotation is up to 90⁰ and approximately half occurs at A-A joint

Ligamentous anatomy: principal stabilising ligaments of C1(figure.1)

1.Transverse atlantal ligament

2.Alar ligaments

Secondary stabilising ligaments

1.Apical ligaments

2.Anterior and posterior A-O membranes

3.Tectorial membrane

4.Ligamentum flavum

5.Capsular ligaments

Craniometric landmarks

(figure.2 and3)

Craniometric measurements	Anatomic landmarks	Normal values and clinical implications

Chamberlain line	Line joining basion to opisthion	Dens projecting >6mm above it indicates basilar invagination
McRae line	Line joining anterior and posterior margins of foramen magnum	Odontoid tip lying above this line is indicative of type A basilar invagination
Weckenheim clivus base line	Line drawn along clivus and extending into upper cervical canal, it should be tangent to odontoid	If it intersects the body of dens anterior craniocervical dislocation is present and vice versa
Clivus canal angle	The angle formed by the weckenheim line and a line	Normal is 150-180 if <150 ventral cord compression may occur

	constructed along the posterior surface of the axis body and odontoid process	
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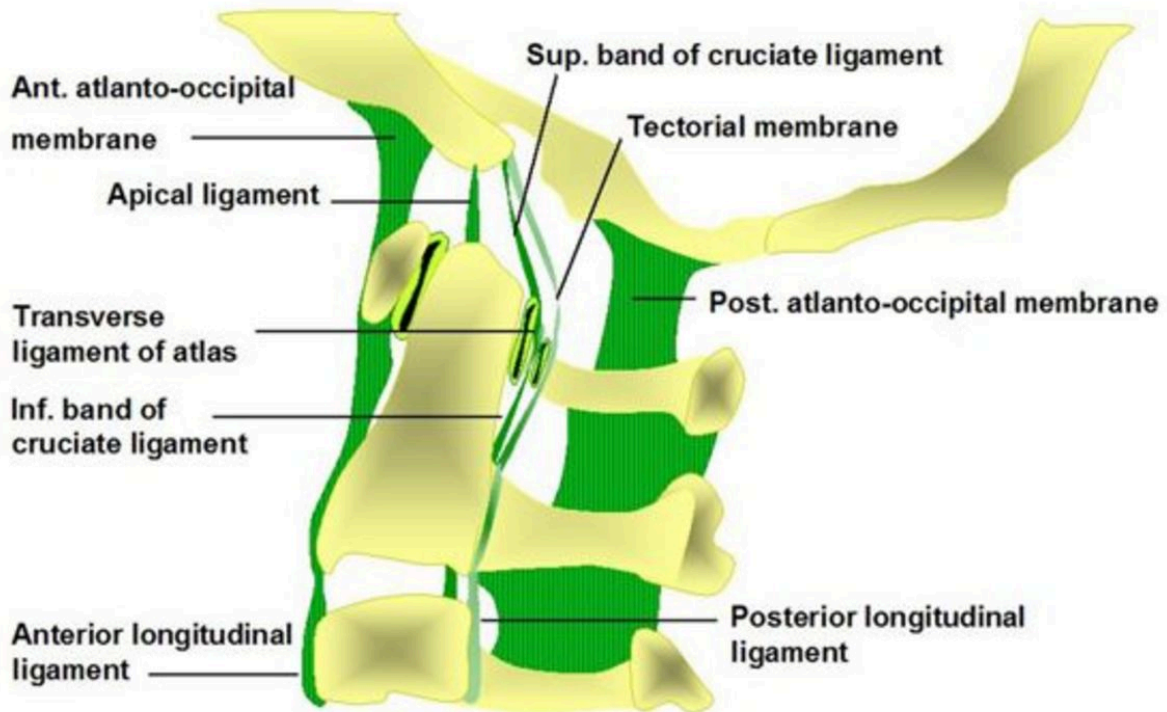


Fig. 1: Normal anatomy and ligaments around CVJ

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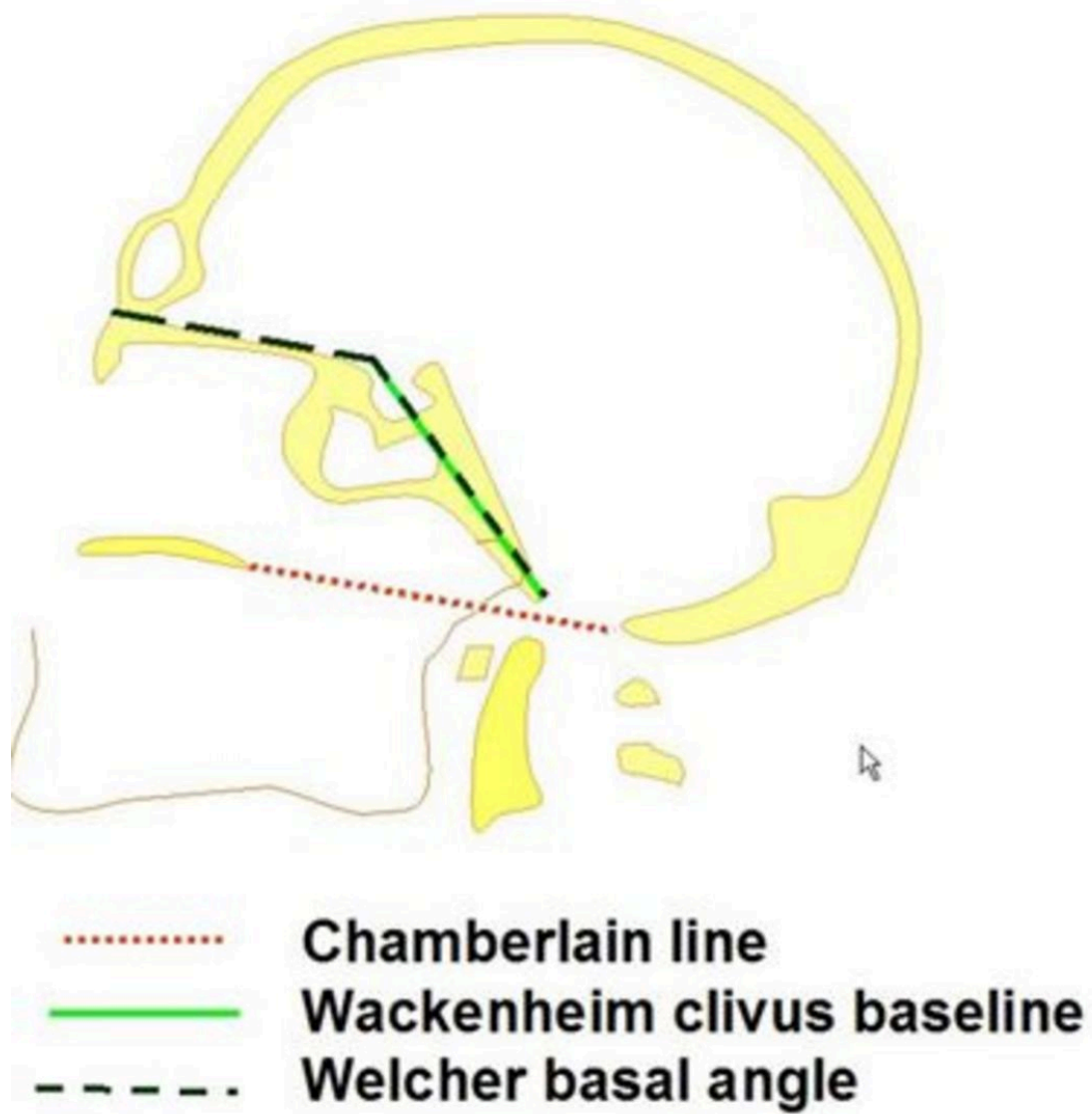


Fig. 2: Craniometry graphical representation

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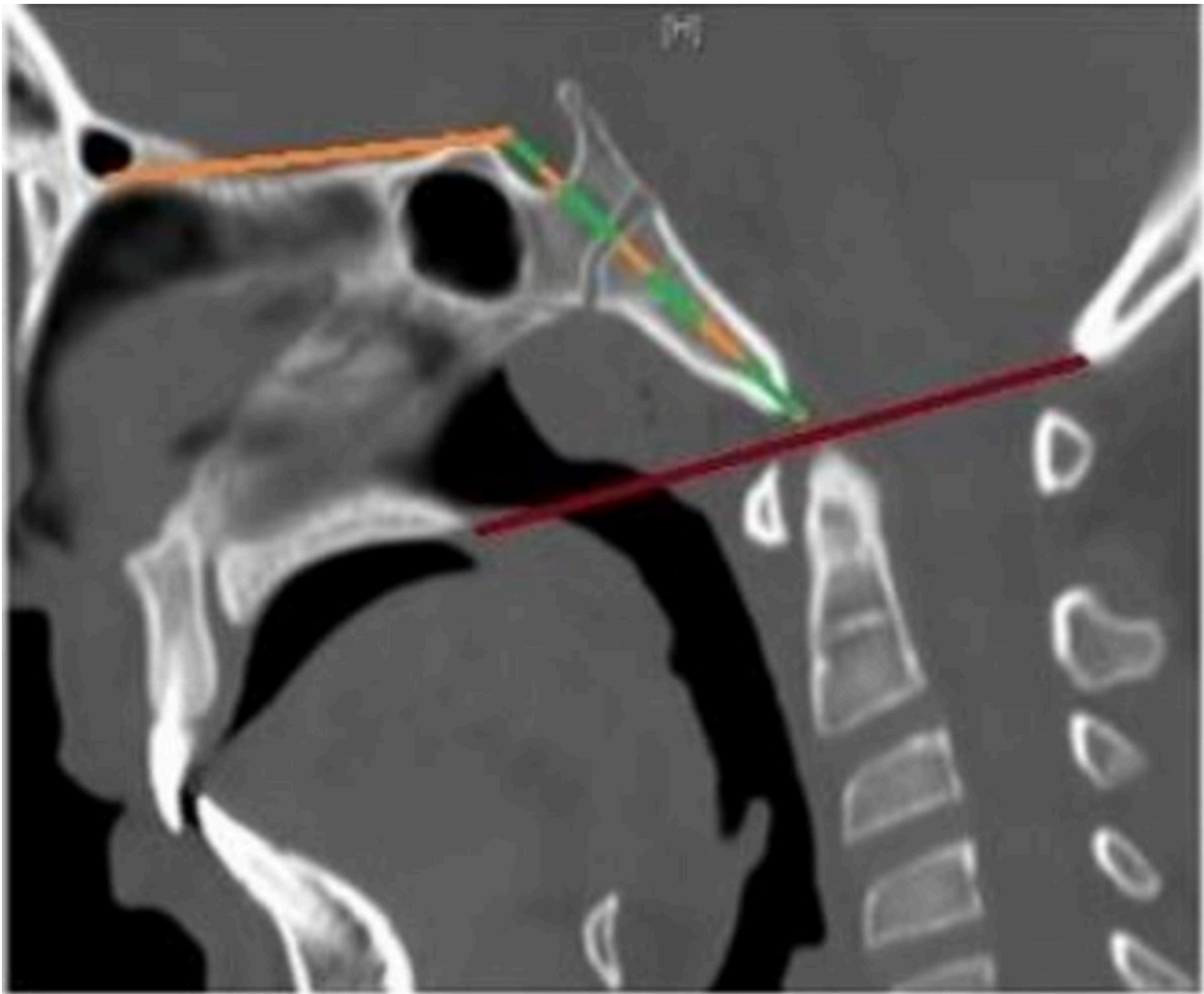


Fig. 3: CT saggital section depicting the various lines used in the assesing the craniovertebral junction anatomy. welcher basal angle(orange) chamberlain angle(red) wackenheim clivus baseline(green)

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Findings and procedure details

Evaluation and assessment of CVJ anomalies is challenging and coming to accurate diagnosis plays a crucial role in the management. Congenital, developmental and acquired anomalies cause atlanto-axial instability or it can affect the vertebra basilar vascular system.

The various causes are broadly classified into

1. congenital

Malformation of the occipital bone

- Clivus segmentation
- Remnants around foramen magnum
- Atlas variants
- Dens segmentation anomalies
- Basilar invagination
- Condylar hypoplasia
- Assimilation of atlas

As Malformations of atlas

- Assimilation of atlas
- Atlanto axial fusion
- Aplasia of atlas arch

Malformation of axis

- Irregular atlanto axial segmentation
- Dens dysplasia
- Ossicular terminalis persistent
- Os odontoidium
- Hypoplasia/ aplasia

Segmentation failure of C2-C3

2 Acquired

Abnormalities at foramen magnum

- Secondary basilar invagination (pagets disease, rheumatoid arthritis)
- Foraminal stenosis(achondroplasia)

Atlanto axial instability

- Errors of metabolism
- Downs syndrome
- Infections (Grisel's syndrome)
- Inflammatory (rheumatoid arthritis)
- Traumatic
- Tumours - neurofibromatosis
- Miscellaneous-syringomyelia, fetal warfarin syndrome, contradi's syndrome

Congenital CVJ anomalies

1.ATLANTOOCIPITAL ASSIMILATION

Failure of segmentation between 4th occipital sclerotome and first spinal sclerotome. Assimilation can be partial or complete. Atlanto-occipital assimilation almost invariably results in basilar invagination. It is also found associated with C2-C3 fusion and atlantoaxial subluxation (figure.4)

2.PLATYBASIA

Is the flattening of skull base with narrowing/ decreased clival angle and increased basal angle.

It Shows a bow string deformity and commonly associated with basilar invagination (figure.6)

3.BASILAR INVAGINATION

Primary developmental defect implying prolapse of the vertebral column into skull at the base. Characterised by abnormally high vertebral column and often found associated with blocked vertebrae and defects of fusion of atlas and occipitalization. (figure.7)

Basilar impression is an acquired form of basilar invagination, secondary to softening of skull (due to conditions like hyperparathyroidism, pagets disease, osteogenesis imperfecta, rickets, hurler's syndrome). Basilar invagination should be suspected when

lateral atlantoaxial articulation cannot be visualised in open mouthed projection radiograph.

4. Basiocciput hypoplasia

Characterised by the shortening of clivus and violation of chabers line. often associated with spondyloepiphyseal and spondylometaphyseal dysplasias (figure.9)

5. Os odontoideum

Commonly encountered congenital anomaly of odontoid.

Anatomical variant of C2 (figure.10)

Associated with atlanto axial instability.

- Morquio syndrome
- Multiple epiphyseal dysplasia

6. Ossiculum terminale

Appears as secondary ossification of dens at the age of 3 to 6 yrs and usually fuses by 12 years.

Failure to fuse results in persistent ossiculum terminale. (figure.11)

Bergmann ossicle- Failure of fusion of apical segment with base of dens.

7. Klippel Feil syndrome

Defined by segmentation failure at one or multiple levels of cervical spine with or without segmentation anomalies of thoracic and lumbar vertebrae.

3-subtypes

Type 1

Extensive fusion of most of the cervical and upper thoracic spine (figure.12)

Type 2

Fusion at 1 or 2 levels and may include atlanto occipital fusion (mc)

Type 3

Comprised of type 1 or type 2 with coexistent lower thoracic or lumbar fusion.

8. Chiari Malformation

Congenital hind brain malformation where cerebellar tonsils are displaced below the level of foramen magnum. (figure.13)

Low lying cerebellar tonsils with extension 5mm below the foramen magnum (type 1)

Associated basioccipital hypoplasia, atlanto-occipital assimilation, platybasia.

Acquired anomalies.

1. Grisel syndrome

Spontaneous subluxation of atlanto axial joint as a result of spread of infection from parapharyngeal space.

Subluxation is due to metastatic inflammation causing ligamentous stretching and subluxation, muscle spasm, regional hyperemia with decalcification of ligamentous structures.

2. Downs syndrome

Most common CVJ anomaly that occurs in children with downs syndrome is atlantoaxial instability and is also often found associated with myelopathy and spinal cord injury. In children with downs syndrome growth of the atlas is slower than that in healthy children. Hypoplasia of the posterior arch of atlas is also seen.

3. Rheumatoid arthritis

Most common inflammatory disease involving the spine and has more predilection for involving craniocervical spine. Three major manifestations of rheumatoid arthritis in cervical spine are basilar invagination, atlantoaxial instability, and subaxial subluxations. (figure.14)

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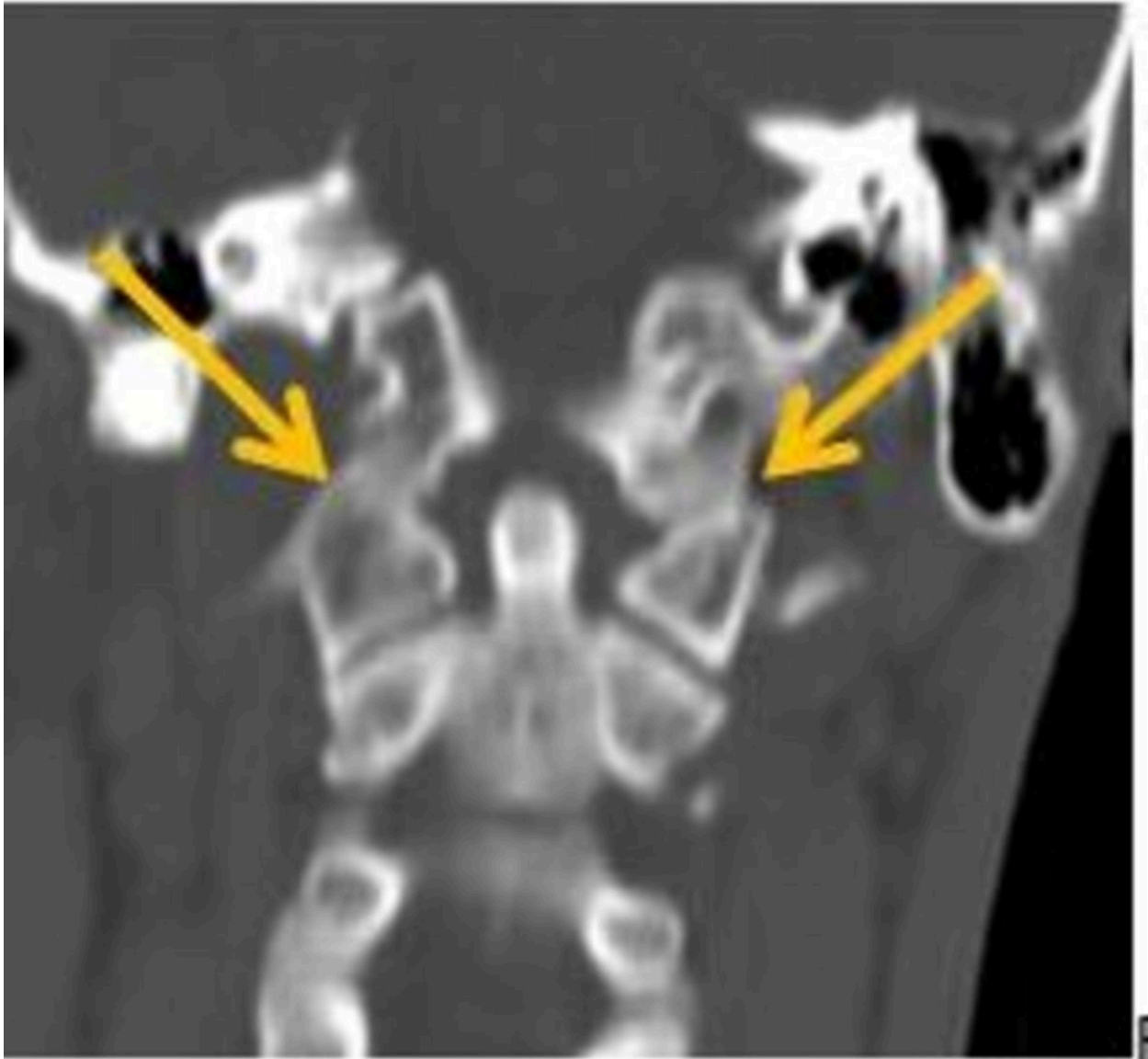


Fig. 4: CT coronal section showing complete atalantooccipital assimilation on right side and incomplete atlanto-occipital assimilation on left side

© • Smoker WRK Craniovertebral junction: normal anatomy, craniometry, and congenital anomalies. Radiographic 1994; 14:2555-7.

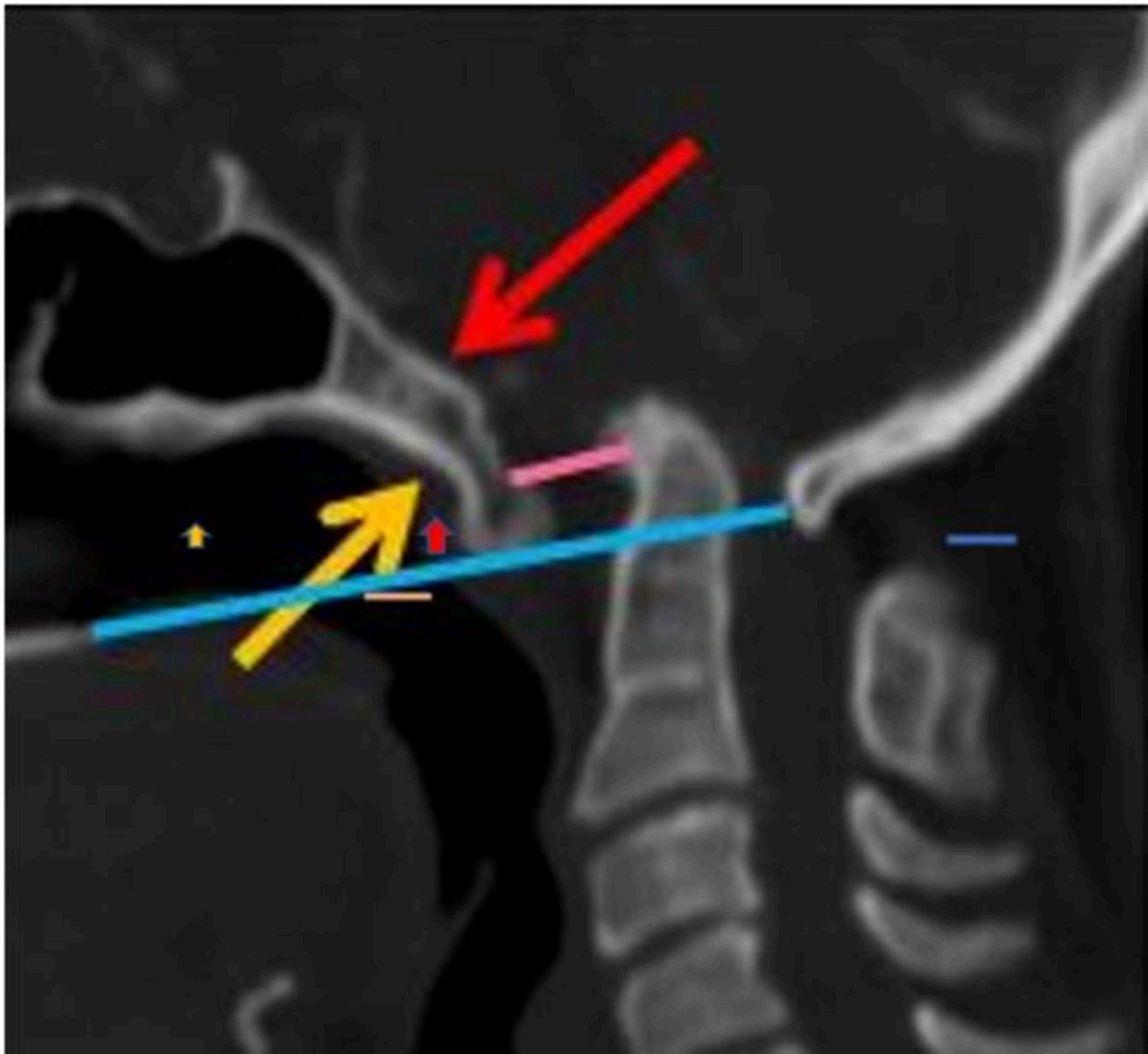


Fig. 5: CT saggital section showing complete atlanto-occipital assimilation(yellow), short clivus(red), voilation of chamberlains line(blue)-basilar invagination and atlantoaxial dislocation(orange).

© • Wendy etal, Craniovertebral junction:Normal craniometry and congenital anomalies; Radiographics:1994:14:225-277

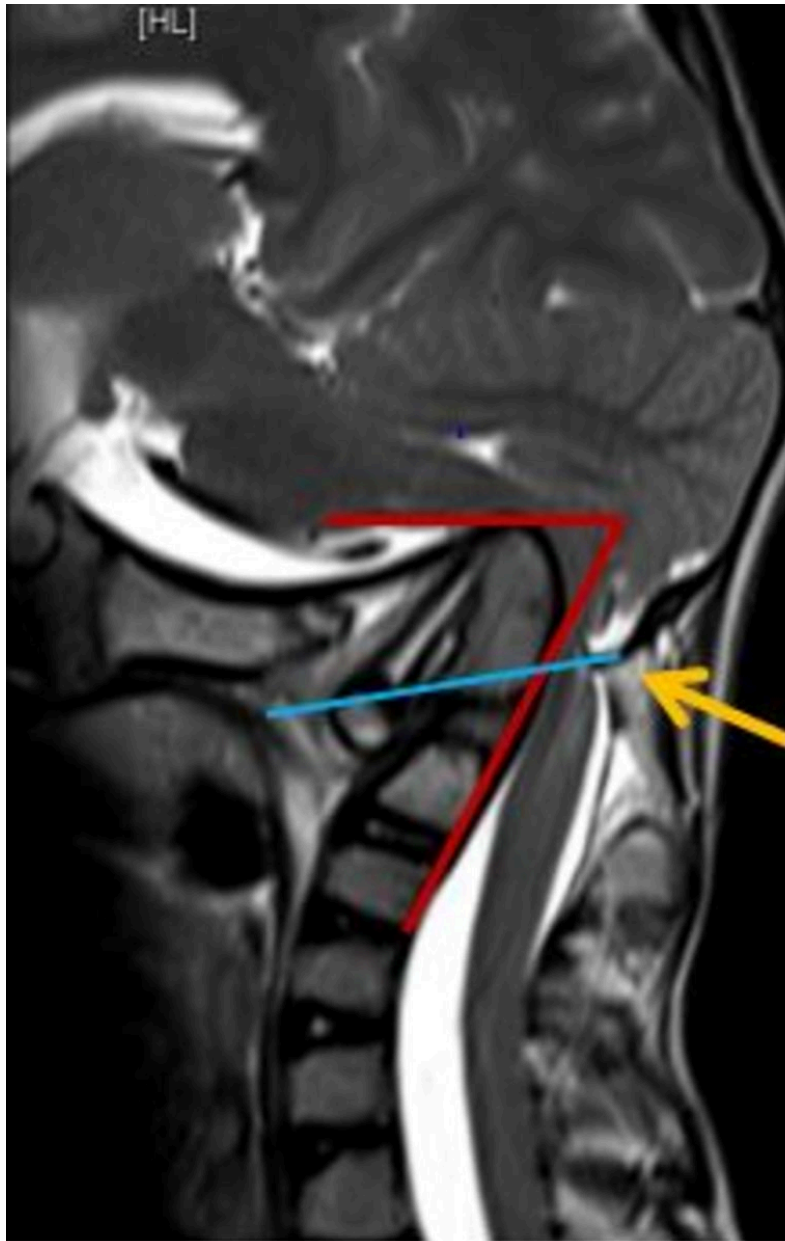


Fig. 6: MRI sagittal section showing decreased clivus canal angle(red) , violation of Chamberlain's line (blue) and acute angulation compression of cervicomedullary junction(yellow).

© • Krakener, J.MRI assessment of normal ligamentous structures in the craniovertebral junction *Neuroradiology*. 2001 Dec;43(12):1089-97

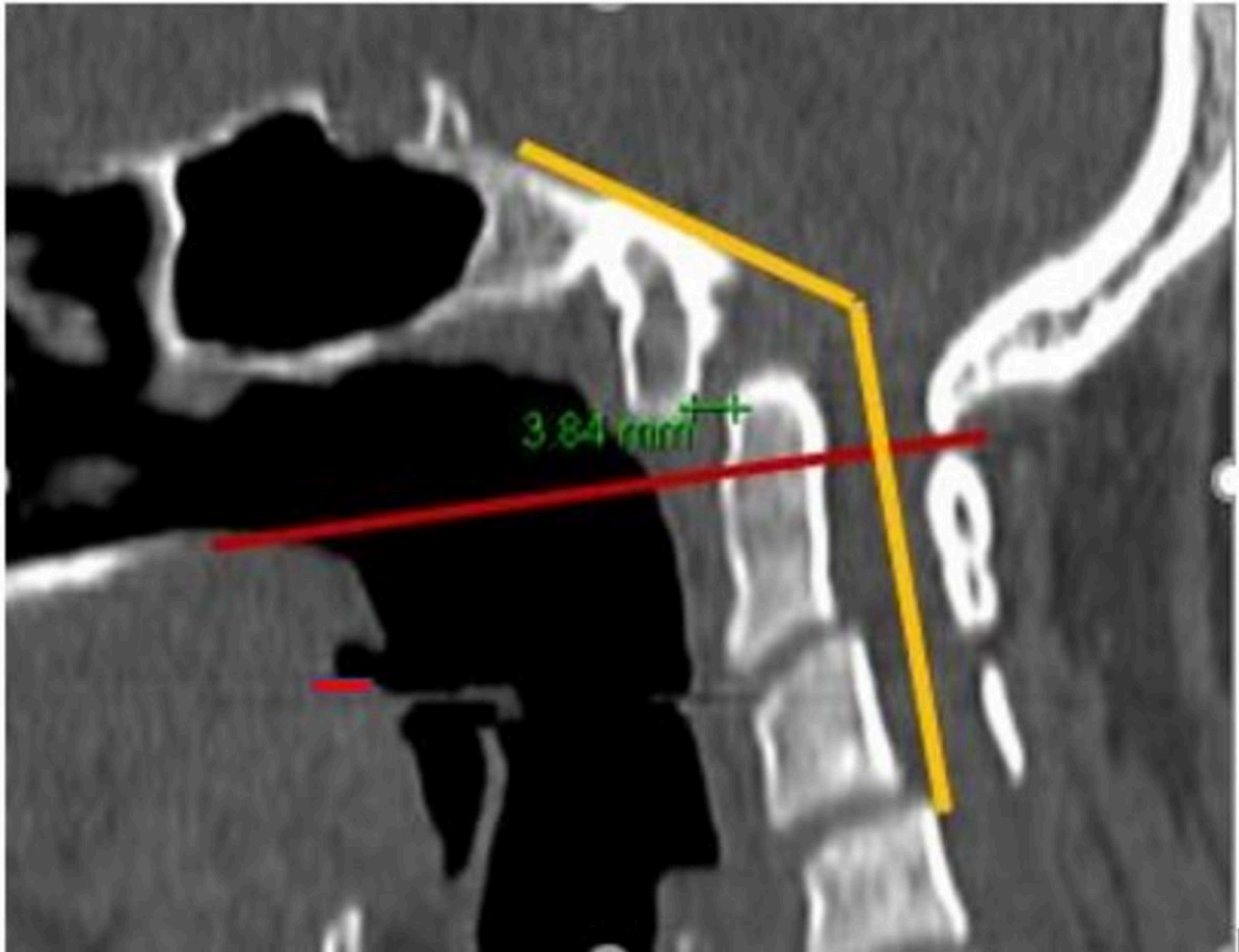


Fig. 7: CT saggital section showing voilation of chamberlain's line(red) and atlantoaxial dislocation(atlantodens interval -3.8mm).Dens projecting 6mm above chamberlains line is diagnostic of basilar invagination

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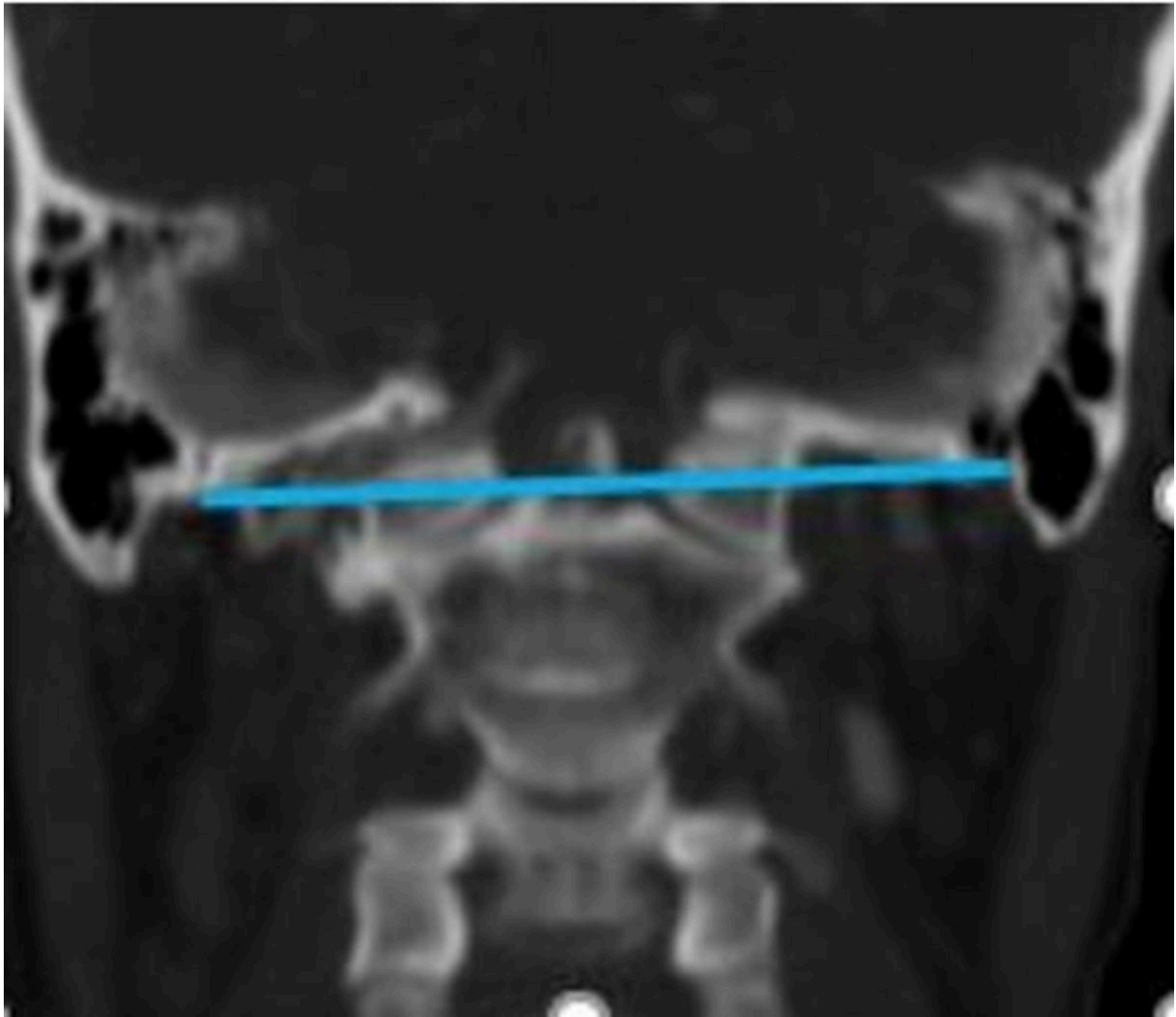


Fig. 8: CT coronal section showing violation of Mc Gregors line in basilar invagination.

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Radiographics:1994:14:225-277

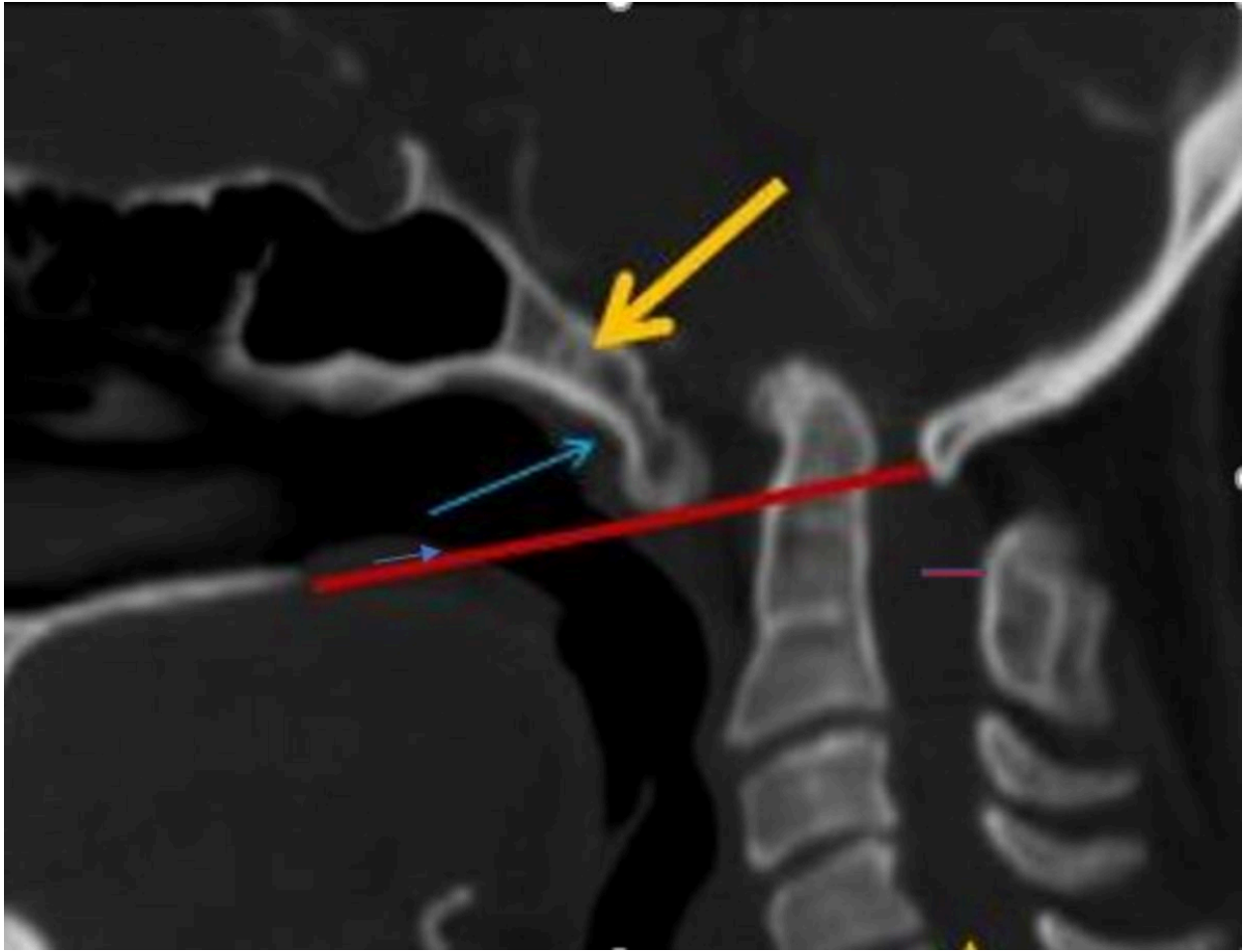


Fig. 9: CT sagittal section depicting short clivus(yellow), atlantooccipital assimilation(blue) and disturbed chamberlain's line(red) suggestive of basiocciput hypoplasia.

© Goel A,Basilar invagination,Chiari malformation,syringomyelia:a review,Neurology India, 2009(3):235-246



Fig. 10: MRI T1W saggital section showing os odontoideum(yellow)

© • Tassanawipas et al, magnetic resonance imaging study of the craniocervical junction, J Orth surg, 2005:13(3):228-231

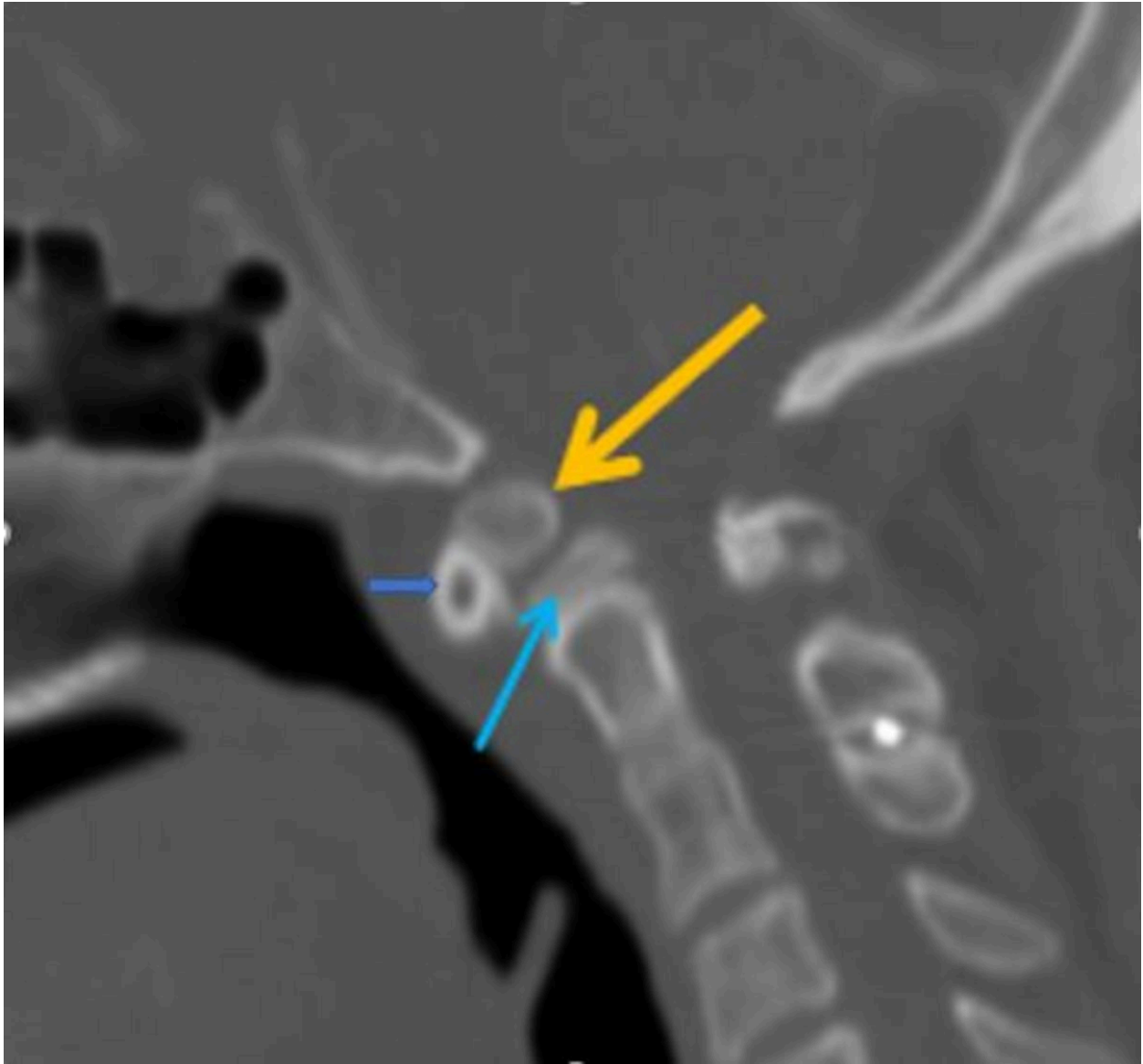


Fig. 11: CT sagittal section showing os odontoideum(yellow) with ossiculum terminale(blue)

© • Crockhard, H.A.Craniovertebral junction anomalies in inherited disorders: part of the syndrome or caused by the disorder? Eur J Pediatr. 1995 Jul;154(7):504-12. Review

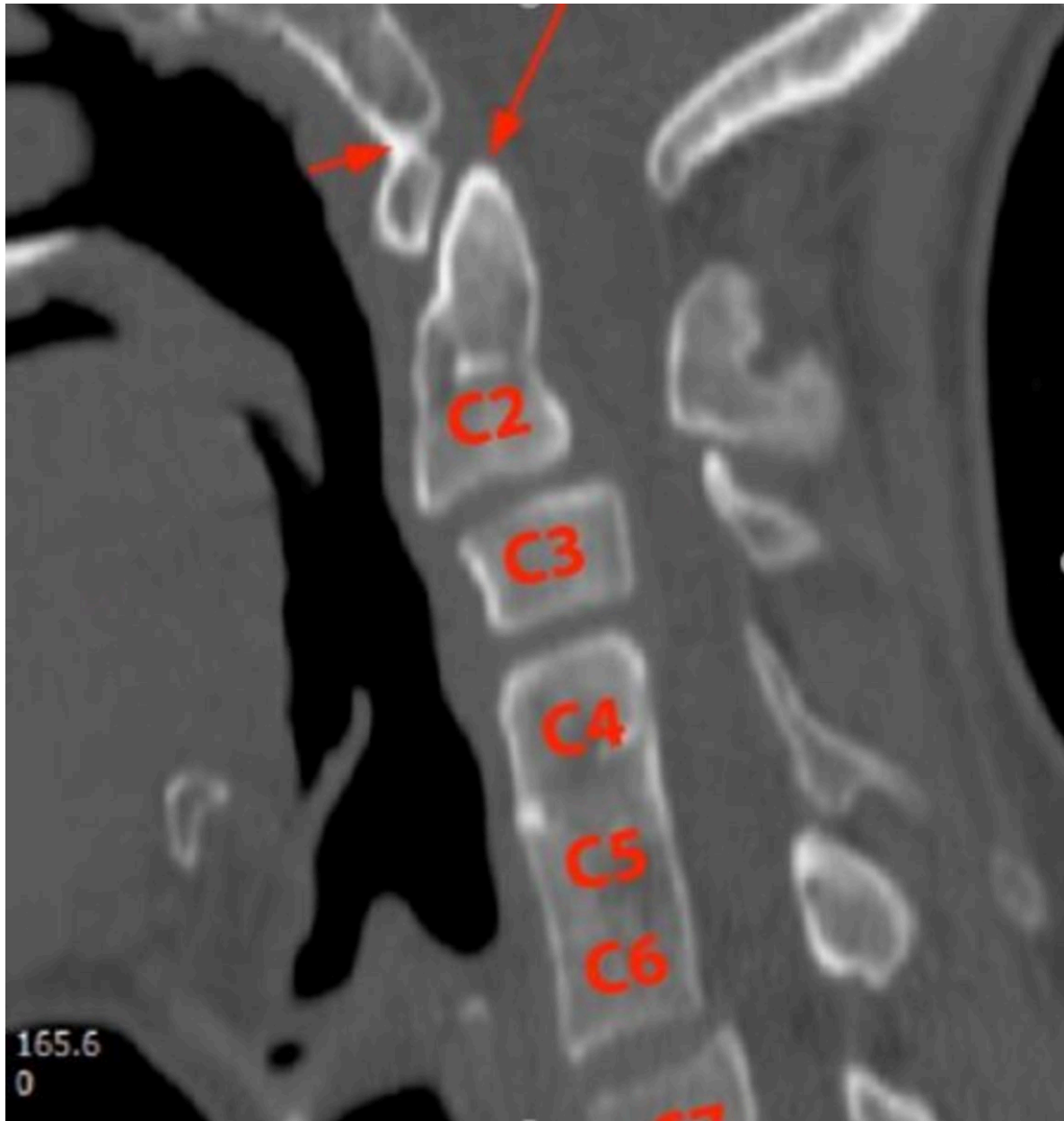


Fig. 12: CT sagittal reconstructed image showing multilevel fusions with atlantooccipital assimilation (arrows), fused vertebrae from C4-C6.-representing type 1

© • Clarke RA, Kearsley JH, Walsh DA. Patterned expression in familial Klippel-Feil syndrome. *Teratology*, 1996;53:152-7

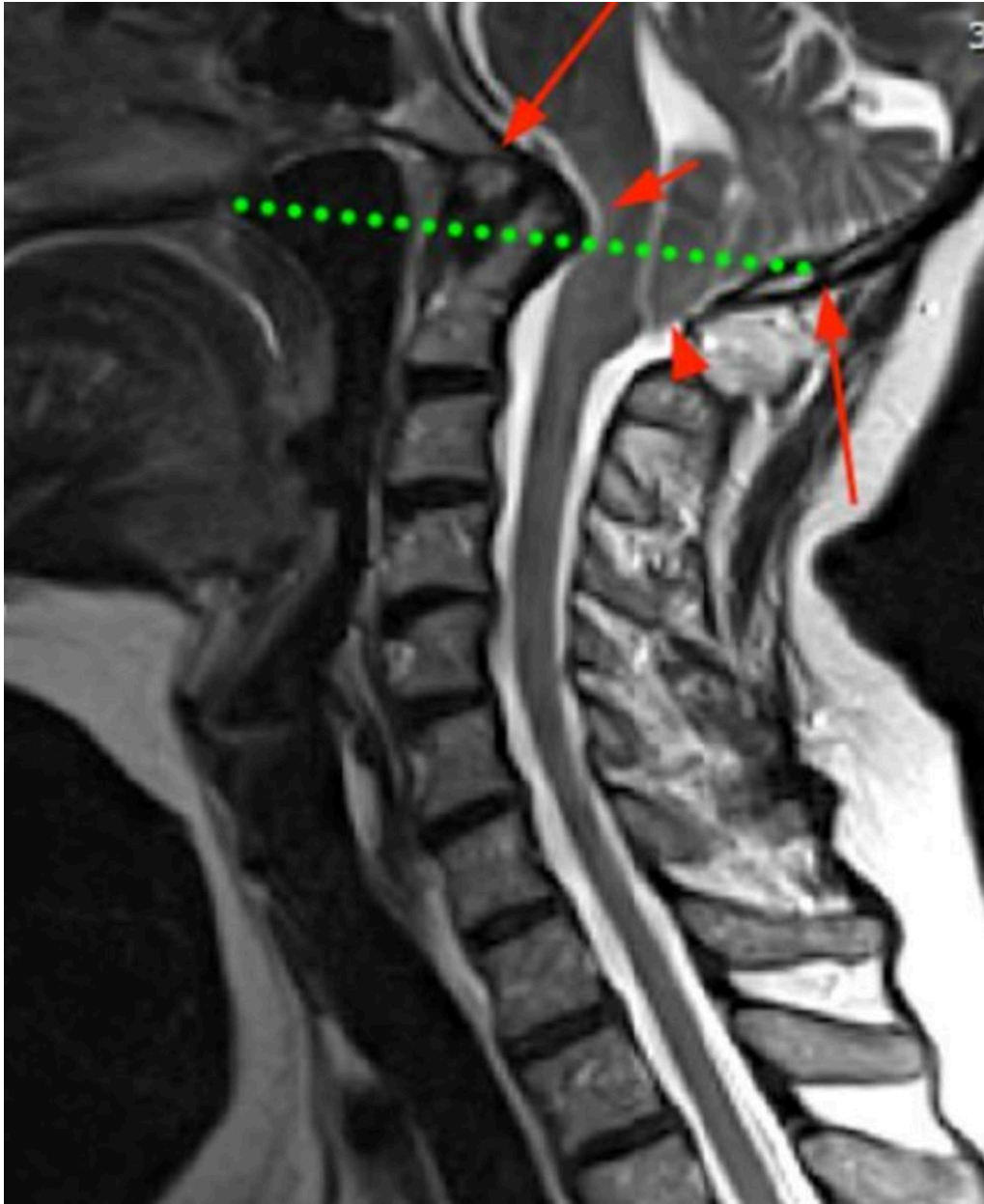


Fig. 13: MRI congenital fusion of the occiput and c1 is present (long arrows) , known as atlantooccipital assimilation. The tip of the dens violates chamberlain line , showing basilar invagination with associated brain herniation (type 1 chiari).

© • Goel A,Basilar invagination,Chiari malformation,syringomyelia:a review,Neurology India, 2009(3):235-246

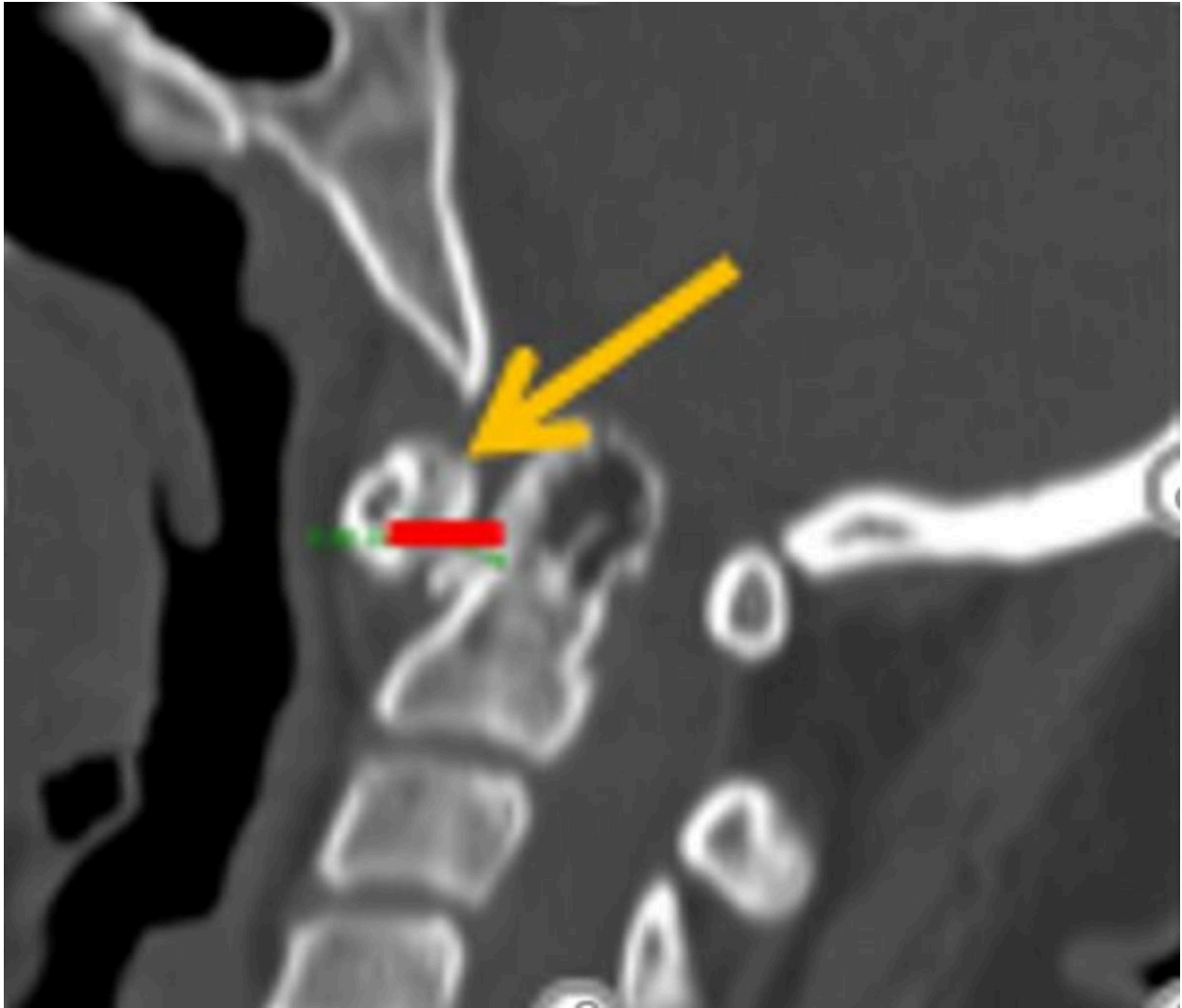


Fig. 14: sagittal ct sections showing basilar impression with sclerosis of atlantoaxial joint and atlantoaxial

© • Wollowick AI, et.al, Rheumatoid arthritis in the cervical spine: what you need to know. The American Journal of Orthopedics, 2007: p.401-406

Conclusion

The CVJ is a common site for anatomic variants and acquired pathology.

Understanding of the important land marks and accurate assessment of the lines and angles is crucial in the evaluation of craniovertebral junction anomalies

Knowledge of the normal anatomy as well as the spectrum of bony variants, acquired anomalies of this region is vital for the diagnosis and correct image interpretation and ulterior management.

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