The craniocervical junction; anatomy, variants, trauma and pathologies.

Poster No.: P-0090
Congress: ESSR 2013
Type: Scientific Exhibit
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Keywords: Anatomy, Musculoskeletal spine, Musculoskeletal joint, CT, MR, Conventional radiography, eLearning, Education, Congenital, Trauma, Neoplasia
DOI: 10.1594/essr2013/P-0090

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Purpose

The Craniocervical junction (CCJ) is a collective term used to describe the articulation of the skull and the cervical spine. This includes the occipito-atlantalandatlanto-axial articulations.

Despite being commonly imaged, the intricacy of the anatomical structures in this region poses challenges to interpreting the imaging findings. Symptomatic cases often have neck pain. Myelopathy, syringomyelia, cerebellar symptoms, cranAial nerve or spinal cord deficits and verteobasilar ischemiamay result from CCJ pathology. Differentiating clinically significant pathologies from normal variants is crucial for guiding further medical and surgical management.

We describe the anatomy of this complex region(1), with demonstration of the imaging features of normal variants, common patterns of trauma as well as various pathology that could affect the CCJ.
Methods and Materials

We describe the anatomy of this region on cross-sectional imaging and review the plain radiographic, CT and MRI imaging for a large gamut of normal variants, anomalies and conditions affecting the CSJ with illustration of cases from our institution. This includes fusion & segmentation anomalies, Chiari malformations, syndromes including Klippel-Feil, accessory ossicles, trauma, arthropathy, infection, osteoporotic basilar invagination, benign tumours as well as primary and secondary malignant bone tumours.

Treatment includes medical management, rehabilitation, internal or external stabilisation or excision and reconstructive surgery.

This is an educational exhibit. No scientific data will be presented.
Results

Introduction:

The CCJ is an anatomical region that is formed by complex articulations involving the occipital condyles and the first two vertebra. These bony structures encase and therefore protect the occipital lobes of the brain, brain stem and the proximal segment of the cervical spinal cord and surrounding CSF spaces. Numerous ligaments stabilise this framework whilst maintaining flexibility of movements at the articulating joints.

Anatomy:

Synovial joints at the suboccipital and atlanto axial articulations. (Images 1-9 Plain radiographs and MRI with annotations as attached image).

Two main stabilising ligaments are:

-The transverse ligament:
The largest, strongest, and thickest craniocervical ligament (mean height/thickness 6-7 mm) (3). The transverse ligament runs posterior to the odontoid process of C-2 and attaches to the lateral tubercles of the atlas bilaterally. Part of the the cruciform ligaments. The transverse ligament maintains stability at the CCJ by locking the odontoid process anteriorly against the posterior aspect of the anterior arch of C-1, and it divides the ring of the atlas into 2 compartments: the anterior compartment houses the odontoid process, and the posterior compartment contains primarily the spinal cord and spinal accessory nerves.

-The alar ligament:
The alar ligament attaches the axis to the base of the skull, from the lateral aspects of the odontoid process, to the anterolateral part of the foramen magnum and/or on the medial aspect of the occipital condyles. (4, 5, 6).

Other ligaments and membranes exist which reinforce stabilisation whilst maintaining adequate flexibility for optimal mobilisation across the CCJ, including the anterior longitudinal ligament, the anterior atlanto-axial and atlanto-occipital ligaments, and the tectorial membrane.
Normal measurements have been standardised at the craniocervical junction and are crucial when evaluating the CCJ and assessing for upper cervical spine injury (7).

**Anatomic Location & Dimensions:**

Basion-dens interval <12 mm

Basion-posterior axial line interval <12 mm posterior to dens, <4 mm anterior to dens

Prevertebral soft tissues <6 mm at C2, flat or concave

Anterior atlanto-dens interval <2 mm

Lateral atlanto-dens interval <2-3-mm side-to-side difference

Atlanto-occipital articulation 1-2 mm

Atlantoaxial articulation 2-3 mm

A variety of lines and standardised measurements are applied to the CCJ to assess for normal alignment and detect disruption of these normal relations (1&8). Sagittal T2WI MRI study of the cervical spine is the recommended sequence to demonstrate the normal alignment and craniometry of the CCJ- (Images 4-9: small field of view to demonstrate the CCJ). The most widely used craniometric lines include (8):

A. Mc Rae’s line: From basion to opisthion. The tip of the odontoid process should always lie below this line in normal individuals.

B. Wackenheim’s clivus baseline/basilar line: drawn parallel to the posterior clivus and extrapolated distally into the spinal canal, where it should touch the odontoid process.

C. The clivus-canal angle/craniovertebral angle: This is formed by the intersection of the basilar line with a line drawn along the posterior cortex of the axis and the odontoid process. The normal range is 150 degrees in flexion and 180 degrees in extension. Ventral spinal cord compression is likely to occur when the angle is less than 150 degrees.

D. The power ratio: the ratio of the distance from the basion to the posterior arch of atlas to the distance between the opisthion and the anterior arch of atlas. This should always be less than one in normal CCJs.

E. The basion-dens interval (< 12mm).
F. The basion-posterior axial line interval: This varies from 4mm anterior to basion to 12mm posterior to basion.

G. The pre-dental space: In infants and young persons this measures 3-5mm, whilst in adolescents and adults this should measures <3mm. The posterior-dental space is normally at least 15mm in children and 19mm in adults and should never be less than 13mm.

We now discuss various pathology of the CCJ with demonstration of cases imaged in our institution,. These include:

-Congenital and normal variants:

• Normal Variants and unfused physis: Accessory ossicles eg Os odontoideum ( image 10) and Ossiculum terminale (image 11&12).

• Congenital basilar invagination: associated with anomalies of the posterior cranial fossa and a short vertical clivus. The causes include basioccipital hypoplasia, occipital condyle hypoplasia, atlanto-occipital assimilation and various neural abnormalities including Chiari malformations ( images 13-18), and may also occur as a result of the bone softening seen in osteogenesis imperfecta ( Image 19&20), muchopolysaccharidosis such as Morquios ( Image 21) and Hurlers’ syndromes as well as metabolic conditions including rickets and hyperparathyroidism. In all cases the craniovertebral angle becomes abnormally acute ( <90 degrees) with possible compression at the CCJ and syringomyelia.

• Atlantoaxial assimilation: Segementation anomalies (image 22 -24).

• Anomalies of the atlas ring: partial or complete aplasia or hypoplasia and fusion anomalies and rotatory subluxation (image 25-28).

• Odontoid process anomalies: such as aplasia/ hypoplasia.

Other miscellaneous conditions associated with CCJ anomalies; including Down’s syndrome, Achondroplasia , Retts (image 29-30 ) and Klippel Feil syndromes (image 31 ) as well as rare conditions such as spondyloepiphyseal dysplasia (image 32-35).

CCJ abnormalities in Down’s syndrome: Variousligamentousand bony abnormalities may occur. These includeatlanto-axial
subluxation, odontoid hypoplasia, os odontoidium and rotatory atlanto-axial subluxation. Hypoplasia of the posterior arch of the atlas is reported in 26% of cases and may be a cause of CCJ spinal cord compression.

CCJ abnormalities in Achondroplasia: A congenital disorder which is the commonest form of short limb dwarfism. CCJ disorders include skull base stenosis, short vertical clivus, segmentation & fusion anomalies, odontoid hypoplasia, atlanto-axial instability, fibrous union of the posterior arch of atlas to the occiput and basilar invagination in more than 50% of cases.

-Trauma:
- Atlanto-occipital dissociation
- Ligamentous avulsion injuries (Image 36).
- Atlas fracture (Image 37-44).
- Atlantoaxial rotatory instability
- Odontoid peg fractures (Image 45-51).
- Fractures of the axis including Hangman's fractures (Image 52-58).

-Infection: (Image 59-61):

CCJ infection is relatively rare. Infection may be blood born, secondary to direct spread, post traumatic or iatrogenic (following surgery or percutaneous interventions). The commonest forms are pyogenic osteomyelitis or craniocervical TB.

-Pyogenic osteomyelitis: very rare at the CCJ. The most reported cases are staphylococcus aureus infection involving C2 vertebra (10). Rare cases of septic arthritis of the atlanto-axial joint have also been reported (11).

Grisel Syndrome: A condition where inflammatory disorders of the upper neck may result in secondary transverse atlantal ligament insufficiency, possibly due to hyperaemia and decalcification of the anterior arch of the atlas. This condition- again uncommon- is seen in children who present with atlanto-axial instability following upper respiratory tract infection. This may rarely be seen in adults (12).
Craniocervical TB: Cervical TB accounts for 10% of all cases of spinal TB, but is very rarely isolated to the CCJ (1% of cases). However, it is a relatively common cause of CCJ instability and cervico-medullary compression in the developing world. Pathologically, three stages of craniocervical TB have been described: Stage 1: Intact ligaments, minimal bone destruction and no signs of instability; Stage 2: atlanto-axial subluxation with minimal bone destruction; Stage 3: Advanced bone destruction and complete obliteration of the anterior arch of C1, often with associated occipitocervical instability.

Arthropathy: (Image 62-75):

The commonest types of arthropathy affecting the cervical spine and CCJ include:

- Degenerative OA

- Rheumatoid arthritis (CCJ and atlanto-axial involvement in 50% of RA patients)

- Calcium pyrophosphate deposition arthropathy (CPPD)

- Amyloid arthropathy (a recognised cause for pseudo-tumours of the CCJ in long term renal dialysis patients)

Tumours:

We present several benign, primary and secondary tumours affecting the CCJ. (Images 76-88).
Fig. 1: Image 1: Lateral plain radiograph of the cervical spine demonstrating normal alignment. The red circle is referred to as the Harris' ring at the base of C2. A broken ring indicates a C2 fracture.

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Fig. 2: Image 2: Lateral plain radiograph of the cervical spine demonstrating normal alignment. Three lines are used to assess for sagittal alignment of the CCJ, the anterior two lines correspond to the anterior and posterior longitudinal ligaments (Red and Blue). The third line assesses for the alignment of the posterior column structures, predominantly the spinous processes (yellow and green). Normally the tip of the C1 spinous process does not articulate with the occiput, with a gap seen on neutral, flexion and extension views (green line).

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Fig. 3: Normal relationships within the craniocervical junction. Drawings illustrate the basion-dens interval (red), the posterior axial line, which is drawn along the posterior cortex of the axis (green), and the basion-posterior axial line interval (c). Horizontal line in c (yellow) represents the range of acceptable basion positions.

Fig. 4: Image 4-9: Sagittal T2WI MRI study of the cervical spine demonstrating normal alignment and craniometry of the CCJ- (small field of view to demonstrate the CCJ).

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Fig. 8: Image 4- 9: Sagittal T2WI MRI study of the cervical spine demonstrating normal alignment and craniometry of the CCJ- (small field of view to demonstrate the CCJ).

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Fig. 7: Image 4- 9: Sagittal T2WI MRI study of the cervical spine demonstrating normal alignment and craniometry of the CCJ- (small field of view to demonstrate the CCJ).

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Fig. 6: Image 4-9: Sagittal T2WI MRI study of the cervical spine demonstrating normal alignment and craniometry of the CCJ- (small field of view to demonstrate the CCJ).

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Fig. 5: Image 4-9: Sagittal T2WI MRI study of the cervical spine demonstrating normal alignment and craniometry of the CCJ- (small field of view to demonstrate the CCJ).

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Fig. 9: Image 4-9: Sagittal T2WI MRI study of the cervical spine demonstrating normal alignment and craniometry of the CCJ- (small field of view to demonstrate the CCJ).

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Fig. 10: Os odontoideum. A bony ossicle cephalad and separate from the axis body in the location of the odontoid process. Considered by some scholars to be traumatic in origin through developmental/embryonic fracture of the dens causing an apical fragment to be pulled proximally by the alar ligament whilst the caudal portion resorbs, leading to the appearance of ossicle. Associated with ligamentous abnormalities and instability of atlanto-axial joint and cervical cord injury as well as rounding and hypertrophy of the anterior arch of the atlas and hypoplasia of the remaining dens with a wide separating gap.

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**Fig. 19:** Image 19 & 20: Osteogenesis imperfecta causing bone softening and secondary cranial settling, with reduced AP dimensions of the foramen magnum and proximal spinal cord compression. Courtesy of radiopaedia (http://radiopaedia.org/cases/osteogenesis-imperfecta-spine).

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Fig. 18: Image 18: T2WI sagittal MRI study showing Chiari malformation type 3 with occipital encephalocele. This is an extremely rare condition which is associated with agenesis / hypoplasia of the corpus callosum and syringomyelia of the cervical spinal cord. Courtesy of http://radiopaedia.org/articles/chiari-iii-malformation

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**Fig. 17:** Image 15-17: Chiari malformation type 2 and an intramedullary syrinx.

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Fig. 16: Image 15-17: Chiari malformation type 2 and an intramedullary syrinx.

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**Fig. 15:** Image 15-17: Chiari malformation type 2 and an intramedullary syrinx.

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**Fig. 14**: Image 13 & 14: Chiari 1 malformation in a scoliosis patient. Sagittal T2 weighted scan of the cervical spine, demonstrating low lying cerebellar tonsils displaced into the upper cervical canal through the foramen magnum (arrow).

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**Fig. 12:** Image 11 & 12: Ossiculum terminale: 2y oss centre at tip of peg. Appears at 3 years and fuses at 12. Present in 26% of normals aged 5-11 years of age.

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Fig. 32: Images 32 - 35 : Spondyloepiphyseal dysplasia. This presents with variable effects. Tarda may go unnoticed until it presents as a short adolescent with premature OA. AP images of the pelvis and knees, demonstrating generalised spondyloepiphyseal dysplasia. Sagittal T1 weighted image of the cervicothoracic spine, showing reduced vertebral body height with increased AP dimensions, an incompletely ossified dens, atlanto-axial instability and spinal canal narrowing at C2, in the same patient.

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Fig. 35: Images 32 - 35: Spondyloepiphyseal dysplasia. This presents with variable effects. Tarda may go unnoticed until it presents as a short adolescent with premature OA. AP images of the pelvis and knees, demonstrating generalised spondyloepiphyseal dysplasia. Sagittal T1 weighted image of the cervicothoracic spine, showing reduced vertebral body height with increased AP dimensions, an incompletely ossified dens, atlanto-axial instability and spinal canal narrowing at C2, in the same patient.

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Fig. 36: Image 36: Sagittal T2 weighted image, showing increased atlanto-axial joint space after trauma, with no associated fracture, indicating disruption of the atlanto-axial ligamentous complex.

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Fig. 37: (Image 37-39): Open mouth peg view, lateral cervical radiograph and an axial CT image showing a showing unilateral fracture of the anterior arch of C1 ring C1 Jefferson’s . This is an extension injury(9). The atlanto-axial joint space appears satisfactory. Note the incidental posterior fusion defect in the posterior arch of C1 on the axial CT image, allowing the presence of a single fracture in the C1 ring.

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Fig. 31: Images 31: Klippel Feil syndrome. Congenital bony fusion of the cervical spine with basilar invagination and odontoid abnormalities. Those particularly at risk include patients with odontoid hypoplasia and C2-3 cervical bony fusion. Both atlanto-axial and occipitocervical instability is demonstrated on dynamic imaging with flexion extension plain radiographs or MRI imaging. Sagittal T1 weighted image, showing fused cervical vertebrae (white arrows), and a partial residual intervertebral disc (black arrowhead).

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Fig. 30: Image 29 & 30: AP and lateral scoliosis views, following surgical treatment of scoliosis in a patient with Retts Syndrome, a neurodevelopmental disorder of the grey matter of the brain, associated with microcephaly, scoliosis, spina bifida, basilar invagination and segmentation anomalies.

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**Fig. 21:** Image 21: Morquio's syndrome. Abnormalities include odontoid hypoplasia, atlanto-axial instability, dural sac stenosis and subluxation of C3/4. Clinically patients often present with gradual myelopathy. Lateral view of the cervical spine, demonstrating anterior vertebral body hypoplasia or beaking (white arrow), atlanto-axial space widening and odontoid hypoplasia (black arrow). Anterior vertebral body beaking is also evident in the thoracic spine.

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Fig. 22: Images 22-24: CT of the cervical spine and coronal T1WI MRI study showing segmentation anomalies, with left hemivertebra at C3 level causing a secondary cervical scoliosis and rotatory instability of the cervical spine

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**Fig. 25:** Image 25-28: Axial CT images, a 3D CT reformat, and T2FSWI MRI image in a patient with a congenital predisposition to rotatory instability of the CCJ. No ligamentous injury was present on MRI.

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**Fig. 40:** (Images 40 & 41): bilateral complete fracture of the anterior and posterior arches of the C1 vertebra. Jefferson's fracture. Note the incongruity and step between the lateral masses of C1 and C2 on the peg view (red arrow), and the fracture involving the C1 spinous process on the lateral view (white arrow).

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**Fig. 61:** Image 59-61: Sagittal PDFSWI, T2WI and T1 post contrast MRI showing C2 osteomyelitis with dens erosion and destruction, joint effusion and fluid along the posterior longitudinal ligament.

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Fig. 53: Image 52 & 53: Open mouth peg view and lateral C-spine radiographs of an unstable type 3 odontoid peg fracture (arrows) Images courtesy of radiology assistant website.

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**Fig. 54:** Image 54: Classification of Hangman's fracture (Courtesy of radiology assistant website). Fracture classification is based on the degree of translation and angulation between C2 & C3.

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**Fig. 55:** Image 55: The CT-images illustrate the fracture-lines of the hangman's fracture. They run through the pars interarticularis resulting in a traumatic spondylolysis (images and text courtesy of radiology assistant website)

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**Fig. 56**: Image 56-58: Sagittal CT scan, Sagittal T1WI and T2WI MRI study. Note the preserved bone marrow signal on T1WI and T2WI MRI. The odontoid process has corticated bony margins in keeping with an old healed C2 fracture. Note the secondary degenerative change, most severe at C2/3.

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**Fig. 51:** Image 50 & 51: 3D and coronal reformats of a type 3 odontoid peg fracture, extending into the lateral masses of C2. (images courtesy of radiology assistant website)

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Fig. 42: Image 42 & 43: Axial and coronal CT images, showing a fractured C1 ring (arrows), with mild atlanto-axial rotatory subluxation.

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**Fig. 43:** Image 42 & 43: Axial and coronal CT images, showing a fractured C1 ring (arrows), with mild atlanto-axial rotatory subluxation.

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Fig. 44: Image 44: Isolated posterior arch fracture of C1 spinous process

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**Fig. 45:** Image 45 & 46: Axial and sagittal images through the atlanto-axial joint, showing a healing type 2 odontoid peg fracture (white arrow), and calcification of the transverse ligament (black arrow)

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Fig. 47: Image 47-49: Sagittal CT reformats, showing angulation and waisting of the odontoid peg in different patients, following healing of previous odontoid peg fractures

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Fig. 48: Image 47-49: Sagittal CT reformats, showing angulation and waisting of the odontoid peg in different patients, following healing of previous odontoid peg fractures

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Fig. 62: Images 62: RA with Cranial settling and C2 vertical instability and atlanto-axial effusion seen posterior to the dens.

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**Fig. 75:** Image 74 & 75: Axial and coronal CT images of a patient with severe degenerative changes at C1-C2, which have resulted in new bone formation arching around the tip of the dens (arrows)

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**Fig. 63:** Image 63& 64: Sagittal CT and coronal CT angiogram images in a patient with rheumatoid arthritis, showing reduced clivus - dens distance (white arrow). Also note the lack of left vertebral artery opacification (black arrow).

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Fig. 65: Image 65: Lateral plain radiograph in a patient with rheumatoid arthritis and cranial settling. The clivus - dens distance is not well-visualised, but the spinous process of C1 is articulating with the occiput.

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Fig. 66: Image 66-68: Sagittal and axial CT images of a patient with rheumatoid arthritis, showing vertical instability and increased atlanto-axial joint space. Note the tapered appearances of the tip of the dens, secondary to erosive changes

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Fig. 68: Image 66-68: Sagittal and axial CT images of a patient with rheumatoid arthritis, showing vertical instability and increased atlanto-axial joint space. Note the tapered appearances of the tip of the dens, secondary to erosive changes.

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Fig. 69: Lateral radiograph in a patient with rheumatoid arthritis, demonstrating increased atlanto-axial joint space, and erosive changes at the dens.

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**Fig. 70:** Image 70 & 71: Lateral flexion and extension views in a patient with rheumatoid arthritis, showing cranial settling, erosive changes at the dens and atlanto-axial joint instability

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Fig. 71: Image 70 & 71: Lateral flexion and extension views in a patient with rheumatoid arthritis, showing cranial settling, erosive changes at the dens and atlanto-axial joint instability

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Fig. 72: Image 72 & 73: Sagittal and axial T2 weighted images, showing the tip of the dens articulating with the clivus (white arrow), and a C1-C2 joint effusion (red arrows). Minor erosive changes are present at the posterior aspect of the dens.

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Fig. 73: Image 72 & 73: Sagittal and axial T2 weighted images, showing the tip of the dens articulating with the clivus (white arrow), and a C1-C2 joint effusion (red arrows). Minor erosive changes are present at the posterior aspect of the dens.

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**Fig. 76:** Case 1: Images 76-78: Axial CT, axial T2 weighted image and sagittal T1 weighted image of a C2 spinous process aneurysmal bone cyst, demonstrating an expansile lesion, containing fluid-levels

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**Fig. 79:** Case 2: Images 79 & 80: Axial CT showing diffuse calcification within the left C1 chordoma (arrows)

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Fig. 80: Case 2: Images 79 & 80: Axial CT showing diffuse calcification within the left C1 chordoma (arrows)

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Fig. 81: Images 81 & 82: Sagittal CT image showing the calcified C1 chordoma (arrows) extending into the left C2 neuroforamen and lateral spinal canal

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**Fig. 84:** Images 83 & 84: Sagittal T2 and T1 weighted images of the cervical spine, demonstrating the soft tissue mass of the C1 chordoma (arrows)

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**Fig. 85:** Case 3: Images 85 & 86: Axial CT scan (post IV contrast) and axial T2 weighted image, demonstrating a destructive enhancing circumferential soft tissue mass at C1-C2 (arrow)

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Fig. 86: Case 3: Images 85 & 86: Axial CT scan (post IV contrast) and axial T2 weighted image, demonstrating a destructive enhancing circumferential soft tissue mass at C1-C2 (arrow)

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Fig. 88: Images 87 & 88: Lateral radiograph and sagittal CT scan showing destruction of the C1 spinous process and anterior arch of C1, secondary to renal cell metastases (arrows)

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Conclusion

Understanding the CJS appearances on imaging is crucial in accurate diagnosis and management of patients.

Misdiagnosis can lead to serious complications and potentially life threatening outcomes.

This review on high quality multimodality imaging of the CCJ, of a wide range of cases with detailed description of findings and differential diagnosis, should increase the reader's awareness of potential findings at the CCJ and boost their confidence in identifying and precisely diagnosing abnormal imaging appearances.
References


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Acknowledgment: The authors would like to express their gratitude to Dr Paul O'Donnell for his contribution of several images for this scientific poster.